

# Medical guidance for DLA and AA decision makers

# (child cases): Staff guide

Medical guidance for DWP staff who make decisions on child cases for Disability Living Allowance and for Attendance Allowance.

This guide is sometimes referred to by staff as the “A to Z of medical conditions”.

Children’s A-Z of Medical conditions

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**A**

Look up	Condition
Acute Lymphoblastic Leukaemia - (ALL)	<a href="#">Acute Lymphoblastic Leukaemia - (ALL)</a>
Acute Myelogenous (Myeloid) Leukaemia - (AML)	Acute Myelogenous (Myeloid) Leukaemia - (AML)
ADD/ADHD (including hyperkinetic disorder/ syndrome)	Attention Deficit Hyperactivity Disorder
Allergies and Anaphylaxis:	
Allergy with a risk of anaphylaxis	<a href="#">Allergies and Anaphylaxis</a>
Allergy - risk of anaphylaxis unknown or not fully assessed	
Allergy - no risk of anaphylaxis	
Oral allergy syndrome	
Food intolerance	
Angioedema	
Hypersensitivity diseases – Other/type not known	
Anaemia – Sickle cell anaemia	<a href="#">Sickle cell anaemia</a>
Angioedema	<a href="#">Allergies and Anaphylaxis</a>
Anorexia Nervosa	<a href="#">Anorexia Nervosa</a>
Anxiety	<a href="#">Anxiety disorders</a>
Arthritis – Juvenile Idiopathic Arthritis	<a href="#">Juvenile Idiopathic Arthritis</a>
Asperger syndrome	Autism Spectrum Disorders

Asthma

Asthma

Ataxic type cerebral palsy

Cerebral palsy

Athetoid type cerebral palsy

Attachment disorder

Decision Makers are advised to discuss with the Department Medical Services if necessary

Attention Deficit Disorder (ADD)/ Attention Deficit Hyperactivity Disorder (ADHD)

Attention Deficit Hyperactivity Disorder

Autistic Spectrum Disorders:-

Autism Spectrum Disorders

Aspergers syndrome

Autism including: -

Pervasive Development Disorder (PDD)

Pathological Demand Avoidance syndrome (PDA)

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**B**



Look up	Condition
Batten disease	<u>Batten disease</u>
Becker type muscular dystrophy	<u>Muscular dystrophy</u>
Bedwetting (Enuresis)	<u>Nocturnal Enuresis</u>
Behavioural and Mental Disorders of children – Other	<a href="#">Learning Disabilities</a> and Decision Makers are advised to discuss with the Department Medical Services if necessary
Blindness	<u>Visual Impairment</u>
Blood clotting disorders:- Haemophilia A Haemophilia B (Christmas disease) Clotting disorder other / type not known	<u>Haemophilia</u>
Brain Tumours	<u>Brain tumour</u>

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**C**

Look up	Condition
Cerebral palsy:-	
Cerebral palsy – Athetoid	<a href="#"><u>Cerebral palsy</u></a>
Cerebral palsy – Ataxic	
Cerebral palsy – Hemiplegic	
Cerebral palsy – Diplegic	
Cerebral palsy – Quadriplegia	
Cerebral palsy – Other type not known	
Child development	Normal development and Disability in children
Choroiditis (Posterior uveitis)	<a href="#"><u>Visual impairment</u></a>
Christmas disease (Haemophilia B)	<a href="#"><u>Haemophilia</u></a>
Chronic suppurative otitis media	<a href="#"><u>Hearing impairment</u></a>
Cleft lip/palate	<a href="#"><u>Cleft lip/palate</u></a>
Clubfoot	<a href="#"><u>Clubfoot</u></a>
Coeliac disease	<a href="#"><u>Coeliac disease</u></a>
Conduct disorder (including oppositional defiant disorder)	<a href="#"><u>Attention Deficit Hyperactivity Disorder (ADHD)</u></a> / <a href="#"><u>Learning Disabilities</u></a> / Netdoctor(CD) / Netdoctor (ODD)  and Decision Makers are advised to discuss with the Department Medical Services if necessary
Conductive hearing loss:	

Otitis media with effusion (OME)

Hearing loss

Chronic suppurative otitis media

Conductive hearing loss due to trauma

Mastoiditis

Otitis externa – chronic

Otosclerosis

Conductive hearing loss - Other cause / cause unknown

Congenital Heart Disease (CHD)

[Congenital Heart Disease \(CHD\)](#)

Conjunctivae/ Corneal / Eyelids/ Lacrimal apparatus diseases:

Corneal ulceration

Visual impairment

Entropion

Herpes zoster – ophthalmic

Keratitis

Keratoconus

Orbital cellulitis

Ptosis

Scleritis

Cystic fibrosis

Cystic fibrosis

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**D**

Look up	Condition
Deficits in attention, motor control and perception (Damp)	<u><a href="#">DAMP</a></u>
Deafness	<u><a href="#">Hearing loss</a></u>
Depression	<u><a href="#">Depression</a></u>
Developmental Coordination disorder also known as Dyspraxia	<u><a href="#">Dyspraxia</a></u>
Diabetes Mellitus:	
Type 1 (insulin dependent)	<u><a href="#">Diabetes</a></u>
Type 2 (insulin and non – insulin dependent)	
Diabetes Mellitus – Category unknown	
Diplegia type cerebral palsy	<u><a href="#">Cerebral palsy</a></u>
Double vision (Diplopia)	<u><a href="#">Visual impairment</a></u>
Down syndrome	<u><a href="#">Down syndrome</a></u>
Duchenne type muscular dystrophy	<u><a href="#">Muscular dystrophy</a></u>
Dyspraxia also known as Developmental Coordination disorder	<u><a href="#">Dyspraxia</a></u>
Dystrophia myotonica (Myotonic Dystrophy)	<u><a href="#">Muscular dystrophy</a></u>

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**E**

Look up	Condition
Eczema – dermatitis type	<u>Eczema</u>
Enuresis (bedwetting)	<u>Nocturnal Enuresis</u>
Epilepsy:	
Generalised seizure (with status epilepticus in last 12 months)	<u>Epilepsy</u>
Absence seizure (Petit mal)	
Atonic seizure	
Clonic seizure	
Myoclonic seizure	
Tonic seizure	
Tonic-clonic seizure (Grand mal) primary or secondary	
Generalised seizure (without status epilepticus in last 12 months)	
Absence seizure (Petit mal)	
Atonic seizure	
Clonic seizure	
Myoclonic seizure	
Tonic seizure	
Tonic-clonic seizure (Grand mal) primary or secondary	

Partial seizure (with status epilepticus in last 12 months)

Complex partial seizure

Complex partial seizure evolving to generalised tonic-clonic seizure

Simple partial seizure

Partial seizure (without status epilepticus in last 12 months)

Complex partial seizure

Complex partial seizure evolving to generalised tonic-clonic seizure

Simple partial seizure

Seizure - unclassified

Non epileptic attack disorder (pseudo-seizure)

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## F

Look up	Condition
Facioscapulohumeral dystrophy	<a href="#">Muscular dystrophy</a>
Failure to thrive/faltering growth	<a href="#">Faltering growth</a>
Fragile X syndrome	<a href="#">Learning Disabilities</a>

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## G

Look up	Condition
Generalised epileptic seizure (with status epilepticus in last 12 months)	<u>Epilepsy</u>
Generalised epileptic seizure (without status epilepticus in last 12 months)	
Global Developmental Delay	<u>Learning Disabilities</u>
Gastro-oesophageal reflux disease (GORD)	<a href="#">Gastro-oesophageal reflux disease (GORD)</a>

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**H**

Look up	Condition
Haemolytic disorders:	
Sickle cell anaemia	<u>Sickle cell anaemia</u>
Haemolytic disorder – Other / type not known	
Haemophilia A	<u>Haemophilia</u>
Haemophilia B (Christmas disease)	
Hearing disorders	
Hearing loss conductive:	
Otitis media with effusion (OME)	<u>Hearing loss</u>
Chronic suppurative otitis media	
Conductive hearing loss due to trauma	
Mastoiditis	
Otitis externa – chronic	
Otosclerosis	
Conductive hearing loss - Other cause / cause unknown	
Hearing loss, Mixed:	
Hearing loss - mixed	<u>Hearing loss</u>
Hearing loss sensorineural:	



Congenital deafness / Pre lingual	<u>Hearing loss</u>
Labyrinthitis	
Meniere's disease	
Presbycusis	
Sensorineural hearing loss due to Trauma	
Sensorineural hearing loss - Other causes of / type not known	
Hearing & balance - Disease affecting hearing & balance – Other / type not known	<u>Hearing loss</u>
Hemiparesis type cerebral palsy	<u>Cerebral palsy</u>
Hip disorders:	
Congenital dislocation of hip	<u>Perthes disease</u>
Perthes disease	
Hydrocephalus	<u>Hydrocephalus</u>
Hyperkinetic disorder/syndrome (includes ADD/ADHD)	Attention Deficit Hyperactivity Disorder
Hypersensitivity diseases:	

Allergy with a risk of anaphylaxis	<u>Allergy and anaphylaxis</u>
Allergy - risk of anaphylaxis unknown or not fully assessed	
Allergy - no risk of anaphylaxis	
Oral allergy syndrome	
Food intolerance	
Angioedema	
Hypersensitivity diseases – Other/type not known	

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## I

Look up	Condition
Incontinence (not Enuresis/Bedwetting) – Other / type not known	<u>Nocturnal Enuresis</u>
Incontinence - stress	
Incontinence - urge	
Inflammatory arthritis - Juvenile Idiopathic Arthritis	<u>Juvenile Idiopathic Arthritis</u>
Iritis (Anterior uveitis)	<u>Visual Impairment</u>

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## J

Look up	Condition
Juvenile Idiopathic Arthritis	<u>Juvenile Idiopathic Arthritis</u>
Joint Hypermobility	<u>Joint Hypermobility</u>

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**L**

Look up	Condition
Labyrinthitis	<u>Hearing loss</u>
Learning disabilities (general):	
ADD / ADHD (including hyperkinetic disorder/syndrome)	Attention Deficit Hyperactivity Disorder
Down's syndrome	<u>Learning Disabilities /</u>  <a href="#">NHS Choices</a> and Decision Makers are advised to discuss with the Department Medical Services re needs and award duration
Fragile X syndrome	<u>Learning Disabilities</u>
Learning disability general – Other / type not known -	<u>Learning Disabilities</u> and Decision Makers are advised to discuss with the Department Medical Services re needs and award duration
Cri du chat syndrome	
Foetal Alcohol syndrome	
Global Development Delay	
Hurler's syndrome	

Klinefelter syndrome

Lesch-Nyan syndrome

Prader-Willi syndrome

Sturge-Weber syndrome

Tay-Sachs disease

[NHS Choices](#) and Decision Makers are advised to discuss with the Department Medical Services if necessary

Trisomy syndromes

[Learning Disabilities](#) and Decision Makers are advised to discuss with the Department Medical Services if necessary

Turner syndrome

[NHS Choices](#) and Decision Makers are advised to discuss with the Department Medical Services if necessary

Learning disorders (Specific):

Speech or language disorders e.g.

Dysphonia

Stammer / Stutter

[Speech or Language disorders](#) and Decision Makers are advised to discuss with the Department Medical Services re needs and award duration

Dyspraxia -

also known as Developmental coordination disorder

[NHS Choices](#) and/or [Speech or Language disorders](#) and Decision Makers are advised to discuss with the Department Medical Services re needs and award duration

Leukaemia lymphoblastic – acute (ALL)

[Acute Lymphoblastic Leukaemia - \(ALL\)](#)

Leukaemia myeloid – acute (AML)

Acute Myelogenous (Myeloid) Leukaemia - (AML)

Limb girdle muscular dystrophy

[Muscular dystrophy](#)

Long sightedness (Hypermetropia)

[Visual Impairment](#)

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**M**

Look up	Condition
Mastoiditis	<u>Hearing loss</u>
Meniere's disease	
Mental and Behavioural disorders of childhood – other/ type not known e.g.	<u>Learning Disabilities</u> and Decision Makers are advised to discuss with the Department Medical Services re needs and award duration
Foetal Anticonvulsant syndrome	
Attachment disorder	
Mixed hearing loss	<u>Hearing loss</u>
Muscular Dystrophy:	
Becker type muscular dystrophy	<u>Muscular dystrophy</u>
Duchenne muscular dystrophy	
Facioscapulohumeral dystrophy	
Limb girdle muscular dystrophy	
Muscular dystrophy – Other / type not known	
Myotonic Dystrophy (Dystrophia myotonica)	

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**N**

Look up	Condition
Neuroblastoma	<u>Neuroblastoma</u>
Nocturnal Enuresis	<u>Nocturnal Enuresis</u>
Non epileptic attack disorder (Pseudo-seizure)	<u>Epilepsy</u>
Normal development in children	Normal development and Disability in children
Nystagmus	<u>Visual Impairment</u> / <u>Nystagmus</u> and Decision Makers are advised to discuss with the Department Medical Services re needs and award duration

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## O

Look up	Condition
Oppositional defiant disorder	<u>Attention Deficit Hyperactivity Disorder (ADHD)</u> / <u>Learning Disabilities</u> / <u>Netdoctor(CD)</u> / <u>Netdoctor (ODD)</u>  and Decision Makers are advised to discuss with the Department Medical Services re needs and award duration
Oral allergy syndrome	<u>Allergy and anaphylaxis</u>
Otitis externa – chronic	<u>Hearing loss</u>
Otitis Media with effusion (OME)	
Otosclerosis	



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**P**

Look up	Condition
Pancreatic disease:	
Diabetes mellitus Type 1 (insulin dependent)	<a href="#"><u>Diabetes</u></a>
Diabetes mellitus Type 2 (insulin & non insulin dependent)	
Diabetes mellitus (category unknown)	
Papulosquamous and inflammatory rashes:	
Eczema – dermatitis type	<a href="#"><u>Eczema</u></a>
Urticaria	<a href="#"><u>Allergy and anaphylaxis</u></a>
Partial epileptic seizure (with status epilepticus in last 12 months)	<a href="#"><u>Epilepsy</u></a>
Partial epileptic seizure (without status epilepticus in last 12 months)	
Perthes disease	<a href="#"><u>Perthes disease</u></a>
Pervasive Development Disorder (PDD)	Autism spectrum disorders
Post-COVID Syndrome (Long COVID)	<a href="#"><u>Post COVID Syndrome (Long COVID)</u></a>
Prematurity	<a href="#"><u>Premature babies</u></a>
Presbycusis	<a href="#"><u>Hearing loss</u></a>

Pseudo-seizure (Non epileptic attack disorder)      [Epilepsy](#)

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## Q

Look up	Condition
Quadriplegia type cerebral palsy	<a href="#">Cerebral palsy</a>

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## S

Look up	Condition
Scleritis	<u>Visual Impairment</u>
Seizure (Epileptic) – unclassified	<u>Epilepsy</u>
Sensorineural hearing loss:	
Congenital deafness	<u>Hearing loss</u>
Labyrinthitis	
Meniere's disease	
Presbycusis	
Sensorineural hearing loss – due to trauma	
Causes of sensorineural hearing loss - Other / cause not known	
Sensorineural hearing loss – due to trauma	
Short-sightedness (Myopia)	<u>Visual Impairment</u>
Sickle cell anaemia	<u>Sickle cell anaemia</u>
Specific childhood learning disorders:	
Speech or language disorders – Other / type not known e.g.	<u>Speech or Language disorders</u>
Dysphonia	
Stammer / Stutter	
Dyspraxia - also known as Developmental coordination disorder	

Spina bifida

Spina bifida

Still's disease (Juvenile Idiopathic Arthritis)

Juvenile Idiopathic Arthritis

Strabismus (Squint)

Visual Impairment

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## U

Look up

Condition

Urinary incontinence:

Stress incontinence

Nocturnal Enuresis /

Urge incontinence

NHS Choices and Decision Makers are advised to discuss with the Department Medical Services re needs and award duration

Urinary overflow

Urinary incontinence (not Enuresis/Bedwetting)- Other / type not known

Urinary overflow

Urinary tract infection (UTI)

Urticaria

Allergy and anaphylaxis

Uveitis (Chorioretinal disorder):

Anterior Uveitis (iritis)

Visual Impairment and Decision Makers are advised to discuss with the Department Medical Services if necessary

Posterior Uveitis (choroiditis)

Uveitis (chorioretinal disorder) - Other / type not known

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v

Look up	Condition
Vertigo	<u>Hearing loss</u>
Visual disorders:	
Cataract	<u>Visual Impairment</u>
Diseases of conjunctiva, cornea, eyelids and lacrimal apparatus:	
Corneal ulceration	<u>Visual Impairment</u> and Decision Makers are advised to discuss with the Department Medical Services if necessary
Entropion	
Herpes zoster - ophthalmic	
Keratitis	
Keratoconus	
Orbital cellulitis	
Ptosis	
Scleritis	Conjunctiva, cornea, eyelids and lacrimal apparatus - Other diseases of / type not known
Diseases of the retina and optic nerve:	

Diabetic retinopathy	<a href="#">Visual Impairment</a> and Decision Makers are advised to discuss with the Department Medical Services if necessary
Hypertensive retinopathy	
Macular degeneration	
Optic atrophy	
Optic neuritis	
Retinal artery occlusion	
Retinal detachment	
Retinal vein occlusion	
Retinitis Pigmentosa	
Retinopathy - Other / type not known	
Retina and optic nerve - Other diseases of / type not known	
Disorders of eye movement:	
Nystagmus	<a href="#">Visual Impairment / Nystagmus</a> and Decision Makers are advised to discuss with the Department Medical Services if necessary
Strabismus (Squint)	
Eye movement - Other disorders of / type not known	
Refractive errors:	
Astigmatism	<a href="#">Visual Impairment / Nystagmus</a> and Decision Makers are advised to discuss with the Department Medical Services if necessary
Hypermetropia (long-sighted)	
Myopia (short-sighted)	
Presbyopia	
Refractive errors - Other / type not known	
Uveitis:	



Anterior Uveitis (iritis)	<a href="#">Visual Impairment</a> and Decision Makers are advised to discuss with the Department Medical Services if necessary
Posterior (choroiditis)	
Chorioretinal disorders - Other / type not known	
Visual field defects:	
Amblyopia	<a href="#">Visual Impairment</a> and Decision Makers are advised to discuss with the Department Medical Services if necessary
Cortical blindness	
Diplopia (double vision)	
Hemianopia	
Quadrantanopia	
Scotoma	
Tunnel vision	
Visual field defects - Other / type not known	
Vitreous disease:	
Posterior vitreous detachment	<a href="#">Visual Impairment</a> and Decision Makers are advised to discuss with the Department Medical Services if necessary
Vitreous haemorrhage	
Vitreous disease - Other / type not known	

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## W

Look up	Condition
Wilms tumour	<a href="#">Wilms tumour</a>

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## What you need to know about Acute Lymphoblastic Leukaemia

### What is Acute Lymphoblastic Leukaemia?

Rapid development of leukaemia symptoms is called 'acute' leukaemia. The type of leukaemia: 'lymphoblastic' or 'myeloid' depends on.....

- [Acute Lymphoblastic Leukaemia](#)

### What are the effects and signs?

Children with acute leukaemia feel generally unwell and often rapidly become really ill.....

- [The effects of Acute Lymphoblastic Leukaemia](#)

### How is it assessed?

Leukaemia is usually suspected from blood tests, particularly the full blood count (FBC).

Further tests are done to

- [Assessment of Acute Lymphoblastic Leukaemia](#)

### How is it treated and managed?

Symptoms of Acute Lymphoblastic Leukaemia develop quickly; a child is likely to become very ill with symptoms of.....

- [Treatment of Acute Lymphoblastic Leukaemia](#)

### What evidence is available?

All children will be managed at their local paediatric oncology centre. The GP will be able to confirm the diagnosis but .....

- [Evidence](#)

### How long will the needs last?

Treatment with combination chemotherapy lasts for just over 2 years for girls and just over 3 years for boys....

- [Prognosis and duration of the award](#)

## What is Leukaemia?

Leukaemia is a form of cancer affecting the white blood cells. Blood cells include white cells to fight infection, red cells to carry oxygen and platelets which help stop bruising and bleeding. Blood cells are made in the bone marrow and when mature they are released into the peripheral blood. However, in leukaemia, these early cells fail to mature and multiply in an uncontrolled way, spilling out into the circulation. They also crowd out normal cells in the bone marrow effectively preventing normal blood cell production. This causes the symptoms of leukaemia which are anaemia, reduced resistance to infection and abnormal bleeding. It can also cause bony pain. This process usually happens very quickly and symptoms develop rapidly over a few days or weeks. Rapid development of leukaemia symptoms is called 'acute' leukaemia. The type of leukaemia: 'lymphoblastic' or 'myeloid' depends on what type of abnormal cell is found on testing.

Without treatment acute leukaemia can cause death in a few weeks from bone marrow failure. Treatment takes from 9 months to 2-3 years to complete depending on the type of leukaemia.

There are two main types of acute leukaemia in children -:

- Acute lymphoblastic leukaemia (ALL)
- Acute myeloblastic (myeloid) leukaemia (AML)

ALL is the most common leukaemia accounting for 80% of cases, the remaining 20% being AML. ALL is also the most common childhood cancer. Children with Down syndrome are more likely to develop leukaemia. Other genetic conditions associated with leukaemia include neurofibromatosis, Shwachman Syndrome, Bloom Syndrome and ataxic telangiectasia.

### **Survival rates**

Survival rates have improved dramatically with modern treatment from a 5 year survival of less than 10% in the 1960s to more than 80% in the 1990s. In the induction period of treatment, the first 4 weeks, approximately 2% of children with ALL will die usually from infection and 2% will fail to go into remission and therefore have a poor prognosis. ALL is fatal without treatment. The treatment is intensive and toxic and lasts up to 3 years. The emotional strain on the child and their family is considerable.

### **What is the incidence of leukaemia?**

Around one third of all childhood cancers are leukaemias. Of these, 80% are the Acute Lymphoblastic (ALL) type. ALL is more common in areas of Britain with higher socio-economic status than average. Acute Myeloid

Leukaemia is much less common and chronic forms of myeloid leukaemia are very rare.

Note: - chronic myeloid leukaemia is not covered by this guidance.

Leukaemia is more common in boys than girls. There is a sharp peak in incidence at age 2-3 years. 400 to 500 children develop a type of leukaemia each year in the UK. More than half of these will be aged under 5 at the time of diagnosis.

### **What are the effects and signs?**

Children with acute leukaemia feel generally unwell and often rapidly become really ill. Parents may notice that their child looks pale or has

nosebleeds or bruises from unexplained or minor trauma. Common symptoms include -:

- Tiredness,
- Weakness,
- Weight loss,
- High temperatures that come and go,
- Frequent infections that do not get better as expected,
- Easy bruising,
- Nose bleeds or bleeding gums,
- Bone or joint pain,
- Blood in the urine or in stools,
- Skin rashes,
- Shortness of breath,
- Swollen glands; and
- Abdominal pain or discomfort.

Leukaemia may affect the brain causing cognitive defects, fits, difficulty moving limbs or personality changes. These symptoms, due to involvement of the brain, are more common in recurrent leukaemia than primary disease.

Indicators of severe functional restriction

- Failure to respond to induction therapy within 4 weeks of treatment – discuss with medical services
- Terminal illness – refer to medical services
- Failure to respond to rescue therapies– discuss with medical services
- Relapse while on therapy or within 6 months of completion of therapy
- Nasogastric feeding

- Undergoing stem cell transplant

### **How is it assessed?**

Leukaemia is usually suspected from blood tests, particularly the full blood count (FBC).

Further tests are done to confirm and type the leukaemia. These tests include -:

- Further blood tests including liver function tests (LFT) and urea & electrolytes (U+E) to check kidney and liver function.
- Bone marrow aspiration /biopsy.
- Further tests will be carried out on the blood or bone marrow to find out what type of leukaemia is present – this may be called ‘immunophenotyping’ in the medical evidence.
- Chromosomal analysis (cytogenetics) and molecular characterisation (see MRD below) of the cells will also be undertaken.
- Lumbar puncture to see if the brain or spinal cord is affected by leukaemia.
- Chest X-ray.
- Ultrasound scan of the abdomen to look at possible kidney involvement or abnormalities. An enlarged spleen, liver and lymph nodes are commonly seen.
- Minimal residual disease (MRD) – if the cells can be characterised at the molecular level, a very sensitive molecular technique can be used to look for (MRD) at different time points during therapy. If minimal residual disease is not detected after induction, patients do very well with only one intensification block and then maintenance therapy. If MRD is detected at this time point, then more intensive therapy is given before maintenance is started.

### **How is it treated and managed?**

#### **Overview**

Symptoms of ALL develop quickly; a child is likely to become very ill with symptoms of their leukaemia over a few days or weeks although some

children can appear deceptively well. All children will undergo assessment of their leukaemia as part of treatment planning. Children will have treatment planned depending on how responsive the leukaemia is likely to be to therapy and the risk of it coming back (relapse). Important parameters in this assessment are the age of the child, the white cell count at diagnosis and the chromosomal (cytogenetic) abnormalities in the leukaemic cells. Later modification of treatment depends on minimal residual disease (MRD) results and response to therapy within the first one to two weeks. Those children in the lowest risk group at diagnosis will be started on the least intensive protocol (regimen A in UK ALL2003). This group includes children aged 1-9 with a white cell count  $<50 \times 10^9/l$  with no adverse cytogenetics. Children in the intermediate risk group will be started on a more intensive protocol (regimen B in UK ALL2003). These include children aged  $\geq 1$  year with white cells counts  $\geq 50 \times 10^9/l$  and all children aged  $\geq 10$  years regardless of their white cell count but with no adverse cytogenetics. Some children will be in a group where the prognosis is known to be poor. This is often determined by the cytogenetics of the leukaemic cells but will include children who do not go into complete remission at the end of induction therapy. These form a high risk group and will be given the most intensive protocol (regimen C in UK ALL2003). A small proportion of these children will require a stem cell transplant. Therapy can be intensified after induction if the MRD results are positive. Children aged under 1 year require different therapy from older children and are treated on an infant protocol which is more intensive. They have a significantly worse prognosis. Most children will be treated in clinical trials. In a clinical trial current best treatment is usually compared to a new treatment that may be more effective at controlling the disease or less toxic in terms of acute or long term side effects. Being in a clinical trial has no effect on the care and mobility needs.

### **CNS directed therapy**

All children with ALL will have a diagnostic lumbar puncture to see if leukaemic cells are present in the spinal fluid surrounding the brain. All children receive treatment into the spinal fluid whether abnormal cells are present or not. Such treatment takes place at regular intervals during treatment of ALL and is called CNS (central nervous system) directed therapy. Leukaemic cells can hide out in the central nervous system during standard intravenous chemotherapy treatment and cause recurrent disease at a later date. It is this therapy and its effects on the brain and spinal cord that cause some of the most significant and enduring side effects of leukaemia treatment. If leukaemia cells are found in the spinal fluid, then high dose intravenous methotrexate or radiotherapy treatment is given to the brain. This causes significant additional short term and long term effects.

### **Phases of chemotherapy treatment**

The initial diagnosis and treatment, including chemotherapy treatment, is likely to be given as an in-patient in hospital, along with supportive care. After the first cycle of chemotherapy the leukaemia will be undetectable when

bone marrow slides are viewed down the microscope in >97% of children. This first cycle of chemotherapy to induce remission is often called 'induction' or 'remission induction' chemotherapy. Further treatment is then given predominantly as an out-patient. This will usually be in two phases -:

- 'Consolidation' or 'intensification' – a high dose treatment to stop the leukaemia coming straight back. CNS directed therapy is given during this phase.
- 'Maintenance' or 'continuation' - a long course of drugs which have a less intensive effect on the bone marrow to ensure the leukaemia does not relapse. CNS-directed therapy is given during this phase.

During the first admission to hospital for diagnosis and treatment an assessment of how the leukaemia has responded to therapy will be made. This is done morphologically i.e. looking down a microscope to determine 'blast percentage' on bone marrow aspirates 7 or 14 days after induction chemotherapy and using 'minimal residual disease' determination at the end of induction, day 28. These results are used to plan ongoing therapy which can be expected to last for 2 years for girls and 3 years for boys (in UK ALL 2003) for those children having chemotherapy as their main treatment. Some children will progress straight to bone marrow transplant or Peripheral Blood Stem Cell Transplant from a matched donor after their initial



chemotherapy treatment. All these children will come from the high risk group.

### **Induction chemotherapy**

Children will normally receive at least 3 separate drugs as part of their induction therapy. These are likely to be -:

- Vincristine (intravenous)
- Prednisolone/dexamethasone (steroid drug) (oral)
- L-asparaginase (intramuscular)

Children on the more intensive protocols (schedules B or C in UK ALL2003) will also receive -:

- daunorubicin (intravenous infusion).

The first part of induction chemotherapy is likely to be given in hospital. Additional therapy to the CNS will also be given. Usually:

- Intrathecal methotrexate

Combination drugs are sometimes given:

- Intrathecal methotrexate with cytarabine and hydrocortisone

5-20% of children with ALL will have cranial irradiation - radiotherapy treatment to the head. This treatment has significant acute and long term side effects.

### **Ongoing treatment after induction chemotherapy**

97% of children given induction chemotherapy will go in to remission within the first 4 weeks of treatment and move on to either -:

- consolidation chemotherapy including CNS-directed therapy followed by maintenance chemotherapy
- PBSCT or bone marrow transplant – less than 3 % of children. No further treatment is given after a transplant unless the patient relapses

For more information see -:

[Peripheral Blood Stem Cell Transplant \(PBSCT\) or Bone Marrow Transplant](#)

## **Consolidation chemotherapy**

Consolidation chemotherapy treatment is given over 6-10 months and begins once remission is achieved. Consolidation therapy is variable between children depending on treatment required but will normally consist of -:

A block or blocks of therapy which may include -:

- intravenous chemotherapy
- intrathecal chemotherapy
- oral chemotherapy
- intramuscular therapy

Intensity and number of blocks will depend on response to therapy assessed at end of induction. Children with CNS disease will receive cranial irradiation or if under the age of 2 years enhanced intrathecal therapy. Boys with testicular involvement will receive testicular irradiation.

Children will be treated predominantly as outpatients or on a day case basis during this phase. They may be attending for chemotherapy from daily to weekly during this period and may also attend for monitoring and management of side effects of chemotherapy such as immunosuppression and one

marrow suppression. Admission is frequently needed for treatment of febrile episodes, infection or blood product support.

### **Maintenance chemotherapy**

Most children will receive chemotherapy agents such as -:

- Daily oral Mercaptopurine
- Weekly oral Methotrexate
- Monthly intravenous vincristine and pulses of oral steroids (dexamethasone or prednisolone)
- Intrathecal chemotherapy with methotrexate every 12 weeks. Those who have had cranial irradiation receive no further intrathecal therapy

See: [Side effects of treatment](#)

**What evidence is available?**

<b>Sources of Evidence</b>	<b>Information Provided</b>	<b>Limitation of Evidence</b>
Claimant/carer	First source of information	May not be objective, does not have specialist knowledge
GPFR	The GP should be able to confirm the diagnosis and provide contact details for the paediatric oncologist or haematologist managing the child's leukaemia.	All children will be managed at their local paediatric oncology centre. The GP will be able to confirm the diagnosis but may not be able to give details of current or planned treatment or prognosis of disabling effects.

<p>Hospital FR</p>	<p>The best source of information will be the hospital based specialist nurse or the consultant haematologist. They will be able to provide details of current treatment and confirm disabling effects. They are the best source of information on duration of planned treatment and prognosis of disabling effects.</p> <p>The most important information to check will be:</p> <p>Confirm diagnosis</p> <p>Confirm treatment is being received e.g. chemotherapy</p> <p>Confirm severe disability due to specific side effects of therapy if claimed e.g. walking difficulties due to acute cerebellar syndrome or myelopathy (spinal cord damage due to drugs). Such damage may resolve, even when severe – always ask for prognosis or expected duration of side effects.</p>	<p>Details of planned treatment may change depending on response to treatment or the development of side effects, information provided may become quickly out of date.</p>
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### How long will the needs last?

Treatment with combination chemotherapy lasts for just over 2 years for girls and just over 3 years for boys. Recovery from the immunosuppressive effects of therapy usually takes 6 months to a year after stopping therapy. If needs related to treatment are identified awards of 3 years for girls and 4 years for boys would be appropriate. Complications such as avascular necrosis, learning difficulties and fits/seizures may cause continuing problems.

For those undergoing PBSCT/stem cell or bone marrow transplant the treatment period is shorter but the treatment is more intensive; recovery after transplant takes from 1-2 years. If needs are identified award duration

should coincide with recovery from treatment - 2 year awards are suggested.

<b>Treatment type</b>	<b>Award Period</b>
Needs related to combination chemotherapy	Boys - award for 4 years from start of treatment
	Girls - award for 3 years from start of treatment
Needs related to PBSCT/stem cell or bone marrow transplant treatment	Award for 2 years

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Side effects of treatment**

Intravenous and oral chemotherapy have well established short term side effects that include -:

- Fatigue
- Hair loss
- Nausea and vomiting
- Diarrhoea
- Weight gain

Steroid drugs - including prednisolone and dexamethasone

These drugs are used in short bursts during chemotherapy treatment. Side effects can include -:

- significant weight gain,
- osteoporosis leading to fractures of long bones or vertebrae
- diabetes mellitus

Rarer side effects include -:

- psychiatric problems ranging from depression and anxiety to full blown psychotic episodes.

- osteonecrosis particularly in hips and knees; patients may require joint replacement in the longer term

#### Biological therapy with Imatinib or Dasatinib

These drugs usually cause minimal side effects. Some children may experience loss of appetite, nausea or diarrhoea. Rare but serious adverse effects are liver toxicity and bone marrow suppression.

CNS-directed therapy in the form of intrathecal chemotherapy or cranial irradiation may have the following side effects -:

- Fits (5-10% of children having intrathecal chemotherapy will experience fits as a side effect)
- Myelopathy – damage to the spinal cord by drugs – numbness, tingling or difficulty moving are likely symptoms. Peripheral

neuropathy – numbness or tingling are common symptoms. This is usually partially or completely reversible over time.

- Very severe tiredness is seen with cranial irradiation and may be called 'somnolence syndrome' in the medical evidence- the child sleeps all the time
- Acute cerebellar syndrome – difficulty balancing when walking or difficulty walking at all are likely symptoms

CNS therapy may lead to ongoing neurological problems including learning difficulties that persist in to adult life which can be mild to severe.

Long term side effects of chemotherapy treatment

In the long term young people and children who have had intensive chemotherapy are at significant risk of long term side effects of treatment – these are summarised here -:

- Second cancers – this is 1 in 20 over the 15 to 20 years after treatment for ALL.
- Lung damage
- Heart disease
- Learning difficulties (related to CNS-directed therapy)
- Behavioural problems (related to CNS-directed therapy)

See: [Problems in adults who had cancer treatment as children](#)

## **Care and mobility considerations**

### **Care**

Children over 10 undergoing treatment are likely to have more care needs than younger children for two reasons. Firstly they are likely to have more intensive treatments, being in a higher risk group, because of their age. This means a greater proportion of children over 10 could be expected to have severe side effects related to therapy. Secondly children aged 10 and over are likely to be substantially self caring – any help with personal care in this age group is likely to be related to their condition rather than immaturity.

Children of all ages are likely to require extra emotional and psychological support and practical help from their parents related to both the disease and



its treatment side effects. In relation to treatment parents will need to spend time doing the following for any child undergoing leukaemia treatment -:

- Supporting their child through painful or distressing treatment
- Caring for a central venous access device. Most children have central lines for treatment and blood tests. Lines must be kept secured, clean and dry
- Ensuring oral drug treatments are taken as prescribed
- Monitoring their child for the side effects of treatment such as fever indicating possible infection or easy bruising. Such monitoring is continuous, signs of infection or bleeding require urgent hospital review and possible admission
- Protecting their child from infection during and after treatment,
- Encouraging their child to eat as taste and appetite are often affected, diarrhoea, constipation and vomiting or nausea may also occur. Eating is particularly difficult during periods of stomatitis (sore dry mouth)
- Some children require nasogastric feeding, often for 10-12 hours over night for which the parents will need special training
- Providing daily mouth care
- Providing an appropriate diet for the child during therapy
- Giving psychological and emotional support to their child (and any siblings) through their illness e.g. dealing with hair loss/time away from school/ being different from peers

Immunosuppression is continuous during treatment but there are additional periods when the white cell count is especially low. Avoidance of crowded places is advised which may include shops such as supermarkets.

Episodes of severe fatigue may endure for many months related to chemotherapy treatment and anaemia. Younger children will require help with all aspects of self care and dressing because of their age and to ensure their central line is not disturbed. Older children may require help due to severe

fatigue, dizziness or sleepiness related to CNS treatment as well as care of their central line.

Some children will require additional care because they have developed significant and sometimes severe side effects. Examples include -:

- Peripheral neuropathy – numbness or tingling are common symptoms and may make using the hands difficult due to numbness. The child may have difficulty walking if the lower limbs are involved
- Tiredness which may be severe. The ‘somnolence syndrome’ where the child sleeps much of the time is a well described side effect of cranial radiotherapy and affects the majority of children who undergo this treatment. Its onset is between 1-12 weeks after the cranial irradiation and usually resolves within 4 months
- Encephalopathy or fits: 5-10% of children having intrathecal chemotherapy will experience encephalopathy most commonly presenting as fits or a focal neurological deficit (stroke). The encephalopathy usually resolves within 48 hours but can have longer lasting effects. A persisting stroke may result in problems with strength and/or balance or if fits continue the child may need to take anti-epileptic drugs. The parent may need to

manage fits if they cannot be controlled with antiepileptic medication

- Acute cerebellar syndrome – difficulty balancing when walking or difficulty walking at all are likely symptoms. This condition tends to resolve in days or weeks at most; it is very rare
- CNS therapy may lead to ongoing neurological problems which may be severe. These include learning difficulties that can persist in to adult life
- Steroid treatment can result in -:
  1. diabetes which may need insulin
  2. avascular necrosis of large joints such as hip and knee can occur and may ultimately lead to joint replacement. In the meanwhile, pain can limit walking and movement
  3. Sleeping difficulties/insomnia
  4. psychosis including mood swings, depression and insomnia
  5. proximal muscle weakness making walking particularly up stairs difficult
  6. vincristine can result in neurological problems such as foot drop and weakness, again hampering mobility
- Regression to earlier stages of development can occur. Often young children go back to bed wetting, sometimes back to nappies. Wet nappies and bed need changing as soon as possible in order to prevent the chemotherapy toxins that are excreted in the body fluids from causing rashes and soreness.

Sickness and diarrhoea are a regular occurrence, as are night sweats all of this means change of bedding and night clothes.

### **Equipment Used (Access) for giving Chemotherapy**

It is essential that the following 'lines' & 'ports' etc and the sites of entry into the body are kept meticulously clean & hygienic in order to prevent infection & this is a time consuming task.

Name	Access	Area of the body
Hickman Line	Venous	This type of line is tunnelled under the skin of the chest wall and inserted into the large veins in the neck. The end of the line is outside the skin about half way down the front of the chest.
PICC (peripherally inserted central catheter)	Venous	This is a long line inserted into a vein in the arm and threaded up to the large veins in the neck. The end of the line is outside the skin near the crook of the elbow.
Portacath/implantable port/port	Venous	This is also inserted into the neck veins but the end of the line is under the skin, usually of the chest wall. A small lump will be palpable under the skin and this is the chamber into which, drugs can be injected.
Peritoneal port	peritoneal	These catheters are usually inserted surgically and have ports underneath the skin of the abdomen.
Omay reservoir	Spinal fluid	The reservoir is a chamber placed under the skin of the scalp, this communicates via a tube with the spinal fluid. This avoids repeated spinal tap procedures.

## Mobility

Severe fatigue related to either of the following may reduce the ability to walk normal distances and exercise tolerance is likely to be much reduced:-

- Chemotherapy treatment; or
- Anaemia.

Some treatments cause severe specific side effects that make walking very difficult or dangerous because of the risk of falls. Most of these effects resolve over time. Information on prognosis of disabling effects relating to treatment is best obtained from the treating paediatric oncologist or haematologist or the specialist nurse on the hospital team. Examples of effects are described above.

### **Peripheral Blood Stem Cell Transplant (PBSCT) or Bone Marrow Transplant**

A Peripheral Blood Stem Cell Transplant (PBSCT) or Bone Marrow Transplant is a way of transplanting a new immune system into a person's body. The immune system consists of stem cells that make all the different kinds of blood cells that perform the functions of the immune system, these are -:

- Red blood cells – these carry oxygen around the body
- White blood cells – these cells travel around the body destroying bacteria, viruses and abnormal cells in the body such as cancer cells
- Platelets – these cells help the blood to clot

A transplant involves destroying the person's own immune system and stem cells and replacing them with either -:

- Their own previously harvested stock of stem cells or bone marrow – this is called an **autologous** transplant because a person's own cells are used.
- Someone else's stem cells or bone marrow – this is called an **allogeneic** transplant because someone else's cells are used.

The donor usually has the same or similar genetic markers to the recipient. Checking for compatibility between donor and recipient is called '**matching**'. The other person can be a close relative such as a brother or sister – a '**sibling donor**' or someone who is unrelated – this is called a '**matched unrelated donor**'. The name of the procedure depends on how the stem

cells are collected from the donor although the principles of the procedures are the same and the effect on the recipient is the same:

### **Peripheral Blood Stem Cell Transplant (PBSCT)**

Peripheral blood stem cells are harvested from the blood. Growth factors are given to the donor to promote production of larger quantities than normal of stem cells for harvesting. These spill out into the peripheral blood from the bone marrow. The stem cells are filtered from the blood using a procedure similar to blood donation. No anaesthetic is required and the process takes several hours. The side effects for the recipient are the same whether bone marrow or peripheral blood stem cells are used.

### **Bone Marrow Transplant**

Bone marrow is harvested from bone marrow in the pelvis during a short operation. The side effects of bone marrow harvesting are minimal – for example there may be soreness around the pelvis for a week or so afterwards. The side effects for the recipient are similar whether bone marrow or peripheral blood stem cells are used.

### **How is a transplant given?**

In preparation for transplant a person's immune system will be wiped out by high dose chemotherapy and sometimes radiotherapy treatment. This often has the effect of killing off residual cancer cells and is the aim of the treatment. Once this treatment is complete the bone marrow or stem cells can be given via an intravenous drip. The stem cells do not start working straight away and can take up to a year to start working effectively and really protecting a person from infection.

The side effects of such high dose treatment are severe. A person is not able to produce their own red, white blood cells or platelets and the transplanted tissue is unable to do this for some time either. This means they need regular support with blood and platelet transfusions to prevent bleeding and control anaemia. Transfusions of white cells to control infection cannot be given and so despite antibiotics they are at very high risk of life threatening infection with normally innocuous bacteria or viruses. When a person is unable to produce their own white blood cells to fight infection this is called 'neutropenia'. The high risk period after transplant when a person

is very vulnerable to infection is called the '**neutropenic period**' – the neutropenic period is usually spent in hospital.

Once some improvement has occurred and the immune system has started to work to a degree they will be discharged from hospital. The person undergoing treatment is likely to feel extremely ill and weak and may sleep for most of the time. They will have many of the side effects of chemotherapy.

Often the first phase of treatment will be given in isolation in hospital – isolation means just that, a person is kept in one room with minimal visitors who all wear barrier clothing to prevent the transfer of infection. The psychological effects of isolation can be severe. The time spent in isolation in the recent past was 1-2 months. This time has been reduced with more of the recovery time spent at home. The same precautions may be necessary at home as would be taken in an isolation room in hospital. These may be troublesome for carers in terms of cleanliness of the home, providing safe food, preventing contact with potentially harmful everyday items and restricting access to people who are themselves unwell. In addition frequent blood tests are needed to monitor progress and this is likely to involve accompanying their family member to hospital and back.

### **Problems in adults who had cancer treatment as children**

People in this category have much longer to develop the long term or enduring side effects of chemo and radiotherapy. The oldest members of this group will have had their treatment in the 1970s. What will happen to them as they age is unknown. Some childhood survivors have already developed significant enduring problems because of their treatment, either during treatment or some years later. The number of adults 'at risk' in this category is set to rise.

Cancer therapy, in particular chemotherapy made great progress in the 1980s and 1990s and for the first time high rates of cure were achieved in some of the common childhood malignancies such as leukaemia and lymphoma. Over time treatment has been modified to become as effective as possible with as few side effects as possible. Significant long term side effects of treatment given in the past are increasingly being recognised. These side effects generally occur because of changes in normal tissue caused by the treatment, these changes take many years to cause symptoms or become apparent. The medical profession is still in the early days of recognising and researching these disorders.

Over time more members of this group can expect to either develop these problems or have them recognised. A breakdown of problems is provided by treatment. Effects tend to be greater when treatment of cancer began at a



young age (under 3) and when large doses of chemotherapy and radiotherapy were necessary. Common cancers in children include leukaemia, lymphoma, brain tumours, bone and soft tissue sarcomas.

Type of cancer treatment	Disabling effect
Cranial irradiation esp. if combined with intrathecal chemotherapy	Neurocognitive defects – reduced IQ, attention deficit, poorer motor/verbal skills, may be severe enough for a Statement of Education Needs (SEN), deafness, epilepsy.
Cranial irradiation, effects are worse if this was combined with abdominal radiation	Hormonal effects including growth impairment in childhood, hypothyroidism, increased risk of infertility, early menopause.
Cranial irradiation	Obesity and its disabling effects.
Chemotherapy or radiotherapy	Increased risk of infertility.
Chemotherapy esp. anthracycline doxorubicin	Heart problems including heart failure, myocardial infarction, rhythmias and sudden death at young age.
Radiotherapy to chest	Lung problems – breathlessness.
Steroids, methotrexate, inactivity due to illness	Osteopaenia/Osteoporosis.
Chemotherapy or radiotherapy	Second cancers especially brain tumour.
Radiotherapy to abdomen (bladder/bowel/liver)	Chronic diarrhoea, malabsorption, bladder problems, kidney problems including rarely kidney failure.

## What you need to know about Acute Myeloid Leukaemia

### **What is Acute Myeloid Leukaemia?**

Leukaemia is a form of cancer affecting the white blood cells.....

- [Acute Myeloid Leukaemia](#)

### **What are the effects and signs?**

Children with acute leukaemia feel generally unwell and often rapidly become really ill.....

- [Effects of Acute Myeloid Leukaemia](#)

### **How is it assessed?**

Leukaemia is usually suspected from blood tests, particularly the full blood count (FBC).

Further tests are done to.....

[Assessment of Acute Myeloid Leukaemia](#)

### **How is it treated and managed?**

Symptoms of Acute Myeloid Leukaemia develop quickly; a child is likely to become very ill with symptoms of.....

- [Treatment for Acute Myeloid Leukaemia](#)

### **What evidence is available?**

All children will be managed at their local paediatric oncology centre. The GP will be able to confirm the diagnosis but .....

- [Evidence](#)

### **How long will the needs last?**

2 year awards are recommended for all children diagnosed with AML...

- [Prognosis and duration of the award.](#)

## **How is it treated and managed?**

### **Overview**

Symptoms of AML develop quickly; a child is likely to become very ill with symptoms of their leukaemia over a few days or weeks although some children can appear deceptively well. All children will undergo assessment of their leukaemia as part of treatment planning and a course of induction

chemotherapy treatment. Having completed induction chemotherapy there are three treatment options for children with AML -:

- The majority of children will have induction chemotherapy followed by 3 courses of consolidation chemotherapy over a period of approximately 6 months. The total treatment and recovery period lasts up to 1 year or so
- A small number of children will have induction therapy followed by one course of consolidation therapy and then a bone marrow or peripheral blood stem cell transplant, very rarely a cord blood transplant, from a matched related or unrelated donor. Total period of treatment and recovery is 1 to 2 years
- Children with a specific form of AML called 'Acute Promyelocytic Leukaemia (APML)' will undergo induction chemotherapy followed by consolidation therapy followed by lower dose maintenance chemotherapy. Children in this group will undergo a prolonged period of treatment similar to children with ALL. Total period of treatment and recovery is 3 to 4 years

Most children will be treated in clinical trials. In a clinical trial current best treatment is usually compared to a new treatment that may be more effective at controlling the disease or less toxic in terms of acute or long term side effects. Being in a clinical trial has no effect on the care and mobility needs.

### **CNS directed therapy**

Children will have a lumbar puncture (LP) at diagnosis to see if they have central nervous system (CNS) disease. If the lumbar puncture is clear, then two treatments (intrathecal therapy) with 3 drugs are given via further LPs, one after each of the first two courses of chemotherapy, to prevent development of disease as leukaemic cells can hide out in the central nervous system during standard intravenous chemotherapy treatment and cause disease later.

Those who have CNS disease will have a minimum of 6 triple drug intrathecal treatments in the first 3 weeks. Monthly intrathecal therapy is then given until after the final course of chemotherapy is completed. Only children who fail to clear disease with intrathecal therapy are considered for cranial radiotherapy; this therapy results in some of the most significant and enduring side effects of leukaemia treatment. Children who have received both cranial irradiation and intrathecal chemotherapy at the youngest ages are likely to have the severest and most enduring side effects.

### **Phases of chemotherapy treatment**

The initial diagnosis and subsequent treatment is given as an in-patient in hospital, along with supportive care. The first phase of chemotherapy to

induce remission is often called 'induction' or 'remission induction' chemotherapy. In the current UK trial for AML (excluding APML), a risk assessment takes place after the first course of therapy and further therapy then depends on the results. Both morphological response (blast count in bone marrow) and molecular response (Minimal Residual Disease) may be assessed. Specific biological markers may also be sought and appropriate targeted (biological) therapy added where indicated. Overall, the next phase of treatment will be some combination of three further courses of chemotherapy unless the patient is in the high risk category when a single course of therapy followed by stem cell transplant is considered.

A small group of children with promyelocytic acute myeloid leukaemia (APML) will not have a stem cell transplant but will have a further prolonged course of 'maintenance' or 'continuation' chemotherapy - a long course of

lower dose drugs to ensure leukaemia remains in remission after consolidation treatment after initial remission induction.

### **Induction chemotherapy**

Children will normally receive a combination of drugs as part of their induction therapy; these will often be given daily over 10 days. The combination of drugs used is likely to include some of the following -:

- Cytarabine\*
- Daunorubicin\* or idarubicin
- prednisolone/dexamethasone (steroid drug)
- thioguanine
- etoposide\*
- Gemtuzumab\* (biological therapy)
- all-transretinoic acid (ATRA) for children with promyelocytic leukaemia only
- arsenic trioxide for children with promyelocytic leukaemia only (blood monitoring required)

\* Currently used in the UK AML 17 trial.

Induction chemotherapy is given in hospital as an intravenous infusion. Additional therapy to the CNS, usually intrathecal chemotherapy, will also be given as described above and will usually include -:

- Intrathecal methotrexate with cytarabine and hydrocortisone

The time from the start of a cycle of treatment to recovery is about 35 days. There may be as few as 6 days after recovery from one treatment cycle before the next is given. Around 90% of children given induction chemotherapy will go in to remission.

### **Consolidation chemotherapy**

All children will progress to consolidation chemotherapy treatment after going in to remission. This is likely to consist of at least 3 further courses of intensive chemotherapy using the same or similar drugs to those used during induction with or without biological therapies. Drugs used will depend on

type of leukaemia, response to induction therapy and the results of biological markers. The gaps between courses are usually no longer than 7 days.

### **Maintenance chemotherapy**

Maintenance chemotherapy is ongoing chemotherapy used for a prolonged period following successful consolidation chemotherapy. It is only used for children with Acute Promyelocytic Leukaemia. Most children will take oral chemotherapy agents such as -:

- ATRA
- Mercaptopurine (oral drug taken every day)
- Methotrexate (oral drug taken once a week)

Some children may continue to receive intrathecal chemotherapy with methotrexate during this period.

### **Treatment for recurrent AML**

AML may recur anytime after achievement of complete remission. It is most common for recurrence to occur in the first year after treatment (50 to 60%). Treatment of recurrence is again chemotherapy and possibly bone marrow

transplant or Peripheral Blood Stem Cell Transplant (PBSCT). Survival is significantly reduced in children that have recurrent AML.

see: [Peripheral Blood Stem Cell Transplant \(PBSCT\) or Bone Marrow transplant](#)

### **Side effects of treatment**

Intravenous chemotherapy has well established short term side effects that include -:

- Immunosuppression (lowered resistance to infection) is particularly severe in those undergoing AML treatment
- Fatigue
- Hair loss
- Nausea and vomiting
- Diarrhoea
- Weight loss

CNS directed therapy in the form of intrathecal chemotherapy has few side effects as it has been significantly reduced. However, those with CNS disease who require repeated intrathecal therapy have a higher incidence of side effects and those with additional radiotherapy the most severe effects. Side effects may include fits and later learning difficulties. Those who have undergone radiotherapy may suffer from the somnolence syndrome (severe tiredness, the child sleeps all the time) or the acute cerebellar syndrome. Combined intrathecal and CNS radiotherapy may lead to severe ongoing

neurological problems including learning difficulties that persist in to adult life.

### **Biological therapy**

The following drugs may be mentioned in the medical evidence. In general their side effects are minimal:

- gemtuzumab ozogamicin (GMTZ) - the most serious adverse effects are liver toxicity and bone marrow suppression
- CEP-701 – lestaurinib – biological therapy, side effects are diarrhoea and vomiting

### **Long term side effects of chemotherapy treatment**

Side effects following treatment of AML more than 10 years ago were reported in an American study:

- Growth abnormalities (mainly reduced height) 51%
- Neurocognitive abnormalities – 30% includes one of the following: academic difficulties, hearing loss, speech problems or epilepsy
- Endocrine abnormalities – e.g. hypothyroidism, growth hormone deficiency, delayed puberty, infertility
- Cataracts – 12%
- Cardiac abnormalities 8%
- Second cancers 2 -10%

See: [Problems in adults who had cancer treatment as children](#)

**How long will the needs last?**



Age at date of claim	Award Period
0 – 15	Award for 2 years, with a further 2 year award on renewal if treatment related needs are identified for children with promyelocytic AML or recurrent leukaemia.

2 year awards are recommended for all children diagnosed with AML. Treatment and recovery for those who successfully undergo combination chemotherapy is likely to take 1 year. For those undergoing PBSCT or bone marrow transplant after chemotherapy it is likely to take 1 to 2 years. Children with promyelocytic AML are likely to undergo treatment for 3 to 4 years but the majority is less intensive and on an out-patient basis.

Some children will endure further courses of treatment in addition to that described above because their leukaemia recurs. Children who will have a recurrence and children who will die from their leukaemia cannot be identified at the beginning of treatment. Of those who will die from their disease almost all will do so within 18 months of onset. Consequently 2 year awards are recommended for all children diagnosed with AML to minimise the risk of a renewal occurring during a child's terminal illness.

On expiry of the 2 year award, the majority of children who have successfully undergone treatment for AML will have recovered and any treatment related needs will no longer be present. The small group of children undergoing treatment for promyelocytic AML will be undergoing maintenance treatment and may have treatment related needs. Awards related to such treatment should be renewed to coincide with the end of treatment. In rare circumstances a child will be undergoing treatment for recurrent leukaemia – 2 year awards are recommended in these cases.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

## **Care and mobility considerations**

### **Care**

Older children undergoing treatment are likely to have more care needs than younger children. This is because older children are likely to be substantially self caring – any help with personal care in this age group is likely to be related to their condition rather than immaturity.

Children of all ages are likely to require extra emotional support and practical help from their parents related to both treatment and the disease. In

relation to treatment parents will need to spend time doing the following for any child undergoing leukaemia treatment:

- Supporting their child through painful or distressing treatment;
- Ensuring oral drug treatments are taken as prescribed, despite side effects
- Monitoring their child for the side effects of treatment – this includes monitoring for signs of infection and easy bruising
- Protecting their child from infection during periods of immunosuppression
- Encouraging their child to eat during periods of stomatitis (sore dry mouth) and providing mouth care
- Providing an appropriate diet for the child when immunosuppressed
- Emotionally supporting their child through their illness e.g. dealing with hair loss/time away from school/ being different from peers

During treatment their child is likely to have periods of being immunosuppressed and be unable to go out in public. Episodes of severe fatigue may endure for many months related to chemotherapy treatment and anaemia. Younger children will require help with all aspects of self care and dressing because of their age. Older children may also require such help due to severe fatigue. Those who have had intensive CNS therapy may also have problems with dizziness and sleepiness.

A very small number of children with CNS involvement will require additional care because they have developed severe side effects mostly related to CNS directed therapy. Examples include:

- Myelopathy – damage to the spinal cord by drugs – numbness, tingling or difficulty moving are likely symptoms. Problems may

include difficulty walking or using the hands for everyday activities. This is rare

- Peripheral neuropathy – numbness or tingling are common symptoms and may make using the hands difficult due to numbness. This is rare
- Very severe tiredness – may be called ‘somnolence syndrome’ in the medical evidence- the child sleeps all the time
- Acute cerebellar syndrome – difficulty balancing when walking or difficulty walking at all are likely symptoms. This is rare
- Fits (5-10% of children having intrathecal chemotherapy will experience fits as a side effect). Child may need to take anti-

epileptic drugs. The parent may need to manage fits if they cannot be controlled with antiepileptic medication

- CNS therapy may lead to severe ongoing neurological problems including learning difficulties that persist in to adult life

### **Equipment Used (Access) for giving Chemotherapy**

It is essential that the following 'lines' & 'ports' etc and the sites of entry into the body are kept meticulously clean & hygienic in order to prevent infection & this is a time consuming task.

Name	Access	Area of the body
Hickman Line	Venous	This type of line is tunneled under the skin of the chest wall and inserted into the large veins in the neck. The end of the line is outside the skin about half way down the front of the chest.
PICC (peripherally inserted central catheter)	Venous	This is a long line inserted into a vein in the arm and threaded up to the large veins in the neck. The end of the line is outside the skin near the crook of the elbow.
Portacath/implantable port/port	Venous	This is also inserted into the neck veins but the end of the line is under the skin, usually of the chest wall. A small lump will be palpable under the skin and this is the chamber into which, drugs can be injected.
Peritoneal port	peritoneal	These catheters are usually inserted surgically and have ports underneath the skin of the abdomen.
Omay reservoir	Spinal fluid	The reservoir is a chamber placed under the skin of the scalp, this communicates via a tube with the spinal fluid. This avoids repeated spinal tap procedures.

## Mobility

Severe fatigue related to either of the following general effects may reduce the ability to walk normal distances, exercise tolerance is likely to be much reduced in all children because of these:

- Chemotherapy treatment
- Anaemia

Some treatments cause severe specific side effects that make walking very difficult or dangerous because of the risk of falls. Most of these effects resolve over time. Information on prognosis of disabling effects relating to treatment are best obtained from the treating paediatric oncologist or haematologist or the specialist nurse on the hospital team.

Examples of effects include:

- Myelopathy – damage to the spinal cord by drugs – numbness, tingling or difficulty moving are likely symptoms. Problems may

include difficulty walking or using the hands for everyday activities.

- Peripheral neuropathy – numbness or tingling are common symptoms and may make walking difficult if the feet are affected by numbness.
- Acute cerebellar syndrome – difficulty balancing when walking or difficulty walking at all are likely symptoms. This condition tends to resolve in days or weeks at most.

## **What you need to know about allergy and anaphylaxis**

### **What is Allergy and anaphylaxis?**

Allergy is a common condition characterised by a 'hypersensitivity' reaction, or exaggerated sensitivity, to substances that are normally tolerated. ....

- [Allergy and anaphylaxis](#)

### **What are the effects and signs?**

Anaphylaxis is a severe, life-threatening, generalised or systemic hypersensitivity reaction .....

- [Effects of anaphylaxis](#)

### **How is it assessed?**

The main principle in diagnosis of allergy is that the clinical history is the most important factor and any test result should be interpreted in the light of this .....

- [Assessment of allergy](#)

### **How is it treated and managed?**

Care for children at risk of anaphylaxis should be provided as a comprehensive management package, containing the essential elements of avoidance advice, provision and training in the use of emergency medication and appropriate follow up.....

- [Treatment for anaphylaxis](#)

### **What evidence is available?**

An allergy specialist will be able to diagnose and define the risk of anaphylaxis for conditions where allergy tests would normally be negative .....

- [Evidence](#)

### **How long will the needs last?**

The risk of anaphylaxis continues for many years.....

- [Prognosis and duration of the award](#)

### **What is Allergy and anaphylaxis?**

The focus of this guidance is severe allergy; this is defined as allergy with a risk of anaphylaxis (an acute hypersensitivity reaction). Allergy is a common condition characterised by a 'hypersensitivity' reaction or exaggerated



sensitivity to substances that are normally tolerated. These substances are called allergens and examples include peanuts, milk and grass pollen. In allergy, the body has made an unwanted and harmful antibody called Immunoglobulin E or 'IgE'. The interaction between the allergen and IgE antibody leads to symptoms that we call allergic reactions. Reactions can be mild – causing eczema to flare up or can be severe - anaphylaxis. Anaphylaxis may be brought about by other mechanisms in some people when allergy is not the cause and IgE antibody is not involved.

The commonest cause of anaphylaxis in children is peanut allergy. The symptoms occur because of the child's immunological response to peanut protein. Common symptoms include swelling of the lips, throat and mouth within 30 minutes of eating peanuts. The most severe symptom of allergy is anaphylaxis. Definitions of anaphylaxis vary but one consistent feature is that it is a life threatening allergic reaction. Life is threatened by one or more of the following effects -:

- severe difficulty breathing due to throat swelling (laryngeal oedema)
- sudden severe asthma
- collapse due to low blood pressure

Any of these allergy related effects can reduce oxygen supply to vital organs and may result in death. The rate of death per anaphylactic episode in food allergy is 1 in 100 events. As a potentially fatal condition and one that rarely resolves, anaphylaxis can have a profound impact on quality of life, extending way beyond the acute phase of the illness. Although still often thought of mainly as an acute disorder, the long term burden of this diagnosis on the family and the impact on quality of life of carers should not be under-estimated.

Many other conditions may be claimed under allergy or labelled allergy. This guidance covers most of these.

Children with allergies frequently have more than one allergic condition. Eczema and asthma commonly accompany allergy with a risk of anaphylaxis. Separate guidance is provided on these conditions.

### **How does an allergy develop?**

In order to develop an allergy a person must become 'sensitised' to a specific allergen. Sensitisation involves exposure to an allergen and in most normal people nothing would happen. When an allergy develops, abnormal IgE specific to that allergen is made. Once the specific IgE is produced, that person is then 'sensitised' to the allergen and is ready to react to it next time they come in to contact with it. However, not everyone with IgE becomes clinically allergic and other factors are involved, which are poorly

understood. Common allergens include pollen, moulds, animals, dust mites and foods.

Allergies develop more commonly in children from families with a history of allergy in the parents or siblings. Certain allergies tend to develop commonly at particular ages. This process is known as the 'allergic march'. For example, eczema often begins in infancy followed by food allergy in toddlers. Hay fever usually begins towards the end of the first decade of life. Exceptions occur and 'adult-type' allergies (e.g. hay fever, tree nut or shellfish allergy) are increasingly diagnosed in young children.

- see: [Incidence and prevalence](#)

### **What are the effects and signs?**

What is anaphylaxis?

Types of allergies and allergens

### **What is anaphylaxis?**

Anaphylaxis is a severe, life-threatening, generalised or systemic hypersensitivity reaction that is characterised by rapidly developing life-threatening respiratory and/or circulation problems. It is usually associated with skin and mucosal (lip, mouth or gut) changes.

The symptoms of anaphylaxis, for example in peanut allergy, are caused by the effects of an inappropriate immunological response to peanut protein. Pre-formed anti-peanut IgE is present in the circulation, skin and elsewhere. The IgE recognises peanut protein and activates cells in the body. These cells release inflammatory chemicals such as histamine. These chemicals cause blood vessels to dilate and become more permeable to fluid. The effect of this is best seen in the skin, which becomes red or swollen or raised. The effect may be described by a parent as flushing or a rash. Anaphylaxis develops when swelling in the airway causing difficulty in breathing, or loss

of fluid from the circulation and dilation of blood vessels cause reduction in blood pressure and loss of consciousness.

### **Symptoms and signs of anaphylaxis**

Typical symptoms include onset of the following symptoms within 30min of exposure to an allergen -:

- A sense of impending doom
- Swelling inside the throat (laryngeal oedema)
- Wheezing and difficulty breathing
- Collapse
- Loss of consciousness
- Severe abdominal pain and diarrhoea

In children, anaphylaxis is most commonly characterised by respiratory difficulty rather than sudden collapse due to reduced blood pressure.

Other features of an allergic reaction may be present but do not in themselves indicate that anaphylaxis has occurred -:

- Swelling of the lips or mouth
- Nausea/vomiting
- Hives – a skin rash also known as urticaria (looks very similar to a stinging nettle rash) – may be all over the body as part of a generalised allergic reaction or be a local effect e.g. touching a peanut with the hand.
- Rhinitis – runny nose
- Conjunctivitis – red, inflamed itchy or watery eyes

Potentially life threatening symptoms may not be listed in the medical evidence but simply be referred to as anaphylaxis or an episode of anaphylaxis. Anaphylaxis is under and over diagnosed by non-specialists. If there is doubt about whether anaphylaxis has occurred or whether there is a risk of anaphylaxis – always rely on the opinion of the consultant allergist.

### **Anaphylaxis risk assessment**

Peanut allergy is the commonest cause of anaphylaxis in children and provides a useful model for risk assessment. The clinical severity of reactions to peanut varies from mild to severe (anaphylaxis) both between individuals

and within individuals over time. Further accidental reactions are usually of similar severity or less severe than the original reaction. Child reactions usually do not become more severe or more frequent, providing they receive appropriate advice. The severity of any reaction and whether the child develops anaphylaxis will depend on the dose of allergen, the route of exposure and the state of health of the child at the time. For example consuming a meal that contained peanut would usually cause a more severe reaction than skin contact. Uncontrolled asthma increases the risk of anaphylaxis.

There is a large number of children who have the potential to develop anaphylaxis, for example all those with peanut allergy, however in reality anaphylaxis rarely occurs. It can be difficult to identify those children at significant risk but anaphylaxis is most likely to occur amongst -:

Those who have previously had an anaphylactic reaction

Those with poorly controlled asthma (defined by a doctor)

Those who react to trace amounts of allergen

Anaphylaxis is the most severe type of allergic reaction covered in this guidance. Milder reactions via the same mechanism may occur, for example, the only allergic reactions experienced so far may have been lip swelling and hives/urticaria. Such reactions are not serious in themselves but they are a marker of allergy and may indicate a risk of anaphylaxis.

## **Types of allergies and allergens**

IgE mediated allergies are diverse, many are mild with few symptoms and others are severe. Examples of mild IgE mediated reactions include mild hay fever and mild asthma. An example of more severe allergy is the presence of nut allergy with a history of anaphylaxis. Children with more severe allergies often have several allergic conditions, for example, they will usually have a food allergy and eczema and asthma. This guidance is specifically designed to cover those children with severe allergies who have either had or are at risk of anaphylaxis. A small number of conditions can induce anaphylaxis independently of IgE, these are described below. Other conditions related to allergy such as eczema and asthma are covered elsewhere in guidance. It may be difficult to work out which cases are severely affected by allergy and which are not and which sections of guidance should be used. The link to 'Guidelines for which guidance to use' below will help you to decide to what category a case belongs and which guidance is most relevant. Several sections of guidance may need to be used to assess a case

with severe allergies because the allergy has caused a range of problems or effects, for example, eczema and asthma and a history of anaphylaxis.

- see: [Guidelines for which guidance to use](#)
- see: [Substances that cause allergy – ‘allergens’](#)
- see: [Care and mobility considerations](#)

### **How is it assessed?**

The main principle in diagnosis of allergy is that the clinical history is the most important factor and any test result should be interpreted in the light of this. Test results do not give any indication of the risk of anaphylaxis, they can only confirm or exclude the causal allergen. The opinion of the Consultant Allergist on the meaning of the test results and the risk of anaphylaxis should always be followed. There are two main investigations for IgE mediated allergy in children and these are skin prick testing and blood tests.

### **Skin prick testing**

In this test drops of diluted allergen are placed on to the arm and the skin pricked through the drop. This exposes the allergen to the immune system. A positive result is indicated by swelling of the skin around the skin prick. The area of swelling is called a wheal. Skin prick testing is a very good way of ruling out allergy as a cause for symptoms. Positive results are not always associated with allergic symptoms and the view of the allergist reporting the skin prick test result is the best guide to interpretation of the test results.

### **Blood tests**

The main test used is called ‘specific IgE’ this measures the amount of IgE that reacts with a particular allergen e.g. peanut. These tests are more commonly available, but are less accurate than skin prick tests.

### **Food challenge testing**

This test involves eating the suspect food in increasing amounts, usually to exclude an allergy. This may be recommended in circumstances where a food allergy is suggested by the history, but allergy tests are negative. Depending on medical judgement this may take place at home, under guidance from the doctor, or in hospital under direct supervision. It is commonly

used to demonstrate resolution of the common food allergies of infancy (e.g. egg and cow milk).

### **Alternative allergy testing**

Children at risk of anaphylaxis will have had allergy testing using one or a combination of the above tests to diagnose their allergy. The GP should be able to confirm the presence of allergy and how and by whom it was diagnosed.

A whole industry of private allergy tests has grown up. They can be bought at health shops and over the internet. Examples include Vega testing, kinesi-ology, hair analysis and alternative blood tests. None of these test results constitute evidence of allergy and any results of such tests should be ignored.

### **The role of the GP, paediatrician and allergist**

It is quite common for children even with severe or multiple allergies to visit the allergy specialist only once for diagnosis and receive all of their future care through the GP or paediatrician. Some allergy specialists will review the child regularly but infrequently, e.g. yearly.

### **How is it treated and managed?**

Care for children at risk of anaphylaxis should be provided as a comprehensive management package, containing the essential elements of avoidance advice, provision and training in the use of emergency medication and appropriate follow up. School and early years setting staff should receive avoidance advice and training in emergency medication. Evidence shows that provision of such a plan by an allergy specialist reduces the risk of accidental reactions.

### **Avoidance**

The main treatment of allergy with a risk of anaphylaxis is avoidance. This means total avoidance of the allergen. All other treatments are secondary to this and are designed to treat the child only when avoidance has failed.

Children and parents will be trained how to practise avoidance and this will be the main action that they will take to preserve the health of their child. It is not easy to avoid common allergens completely. Allergens may be hidden in foods that are not clearly labelled or foods may be contaminated with allergens during manufacture or packaging, again this will not be labelled.

Parents are likely to spend time contacting manufacturers about individual products to find out if they are safe. The amount of time and effort put into avoidance will depend on the perceived severity of each child's allergy.

Children with peanut and nut allergies are most likely to be affected by this as many of them react to small traces of nut allergen. Up to 50% of children with peanut allergy may have reactions to 3mg (1/50<sup>th</sup>) of a peanut, although the majority of reactions to such small doses are mild. This is clearly a very small amount, and too small to see or smell in food to be able to avoid it. A smaller number of children will react to even smaller traces of nut allergen, and may not have to eat it to react. For example nut proteins in the air of a bakery may be enough to cause a reaction. Nut proteins on a door handle when touched by a severely allergic person may be enough to cause a reaction, although this degree of sensitivity is extremely rare. Reactions to tiny traces of allergen that have not been eaten are most likely to cause mild, but not immediately life threatening reactions.

Avoidance requires constant vigilance and means simple activities like eating lunch at school or going to a friend's house require planning and may cause stress. The simplest solution is for the child to eat only safe food prepared at home in a nut free environment at all times. No matter how well educated and motivated parents and children are, reactions are likely to occur. Most accidental reactions to peanut occur at home or with the extended family, only 1 in 20 reactions happen at school. Children's independent activities are likely to be restricted because of their allergy, these include sleepovers and playing at friends' houses.

Social isolation can be a real problem for the child with allergies. This can range from 'being different' not being able to go on school trips without a parent, to eating lunch at a peanut free table. All of these issues are negative but necessary aspects of avoidance. They apply to children with any type of allergy where there is a risk of anaphylaxis. The problems are worse for children with more than two allergies. A proportion of children may be home schooled because of their allergy and more may feel very isolated despite attending school.

### **Treatment of asthma**

The risk of death during anaphylaxis increases in children who have asthma that is not well controlled. In these children, it is vital that asthma medication is given regularly as prescribed in addition to any other measures but mainly allergen avoidance that are taken.

### **Treatment of allergic reactions**

Each child at risk of anaphylaxis should have a written emergency treatment plan to follow when they have an allergic reaction. This should include provision of an intramuscular adrenaline autoinjector and oral antihistamines. School and Early Years Settings staff should also have a copy of

the treatment plan and know how to avoid the appropriate trigger and be trained in treatment of the allergic reaction.

## **Emergency Treatment**

The cornerstone of treatment of an episode of anaphylaxis is an injection of adrenaline given into a muscle. This is often given by a care-giver using an auto-injector device. Other medications such as antihistamines or corticosteroids may also be given by mouth, or injection. Less severe allergic reactions may be treated with oral antihistamines alone.

Once anaphylaxis has occurred then that child will remain at risk of further anaphylactic reactions for some considerable time, as the condition rarely resolves and some triggers are highly prevalent in the environment (e.g. children with milk induced anaphylaxis). Some children who have only ever had mild reactions may also be at risk of anaphylaxis because of the nature of the allergy and other co-factors (e.g. peanut allergy or a child with food allergy and poorly controlled asthma).

Prevention of further episodes of anaphylaxis is vital and is achieved by referral to a specialist allergy clinic. The triggers of anaphylaxis are ruled in or out using the clinical history, specialised testing and where relevant challenge testing. A risk assessment for future anaphylaxis is performed, detailed advice is given on allergen avoidance and treatment of further reactions. Training of carers and school staff in all these aspects is achieved by coordinating with trained community nurses, and other allergies (especially asthma) are treated. This approach has been shown to reduce the number and severity of further anaphylactic reactions.

## **Adrenaline autoinjector**

An intramuscular injection of adrenaline, given early during a reaction is the cornerstone of treatment. It can be referred to sometimes by the brand name (e.g. EpiPen, AnaPen or Twinject). It should be carried at all times. In practice for children with food allergies, the rule is that the device should be available whenever food is eaten, i.e. both in the home and in School and Early Years Settings.

The autoinjector is a disposable spring-loaded needle device. Different devices are activated using specific techniques and training should be provided. An autoinjector with 0.3mg adrenaline is provided for children >30kg and 0.15 for children <30kg. A further dose of 0.5mg is available for some injector types.

Adrenaline reverses the symptoms of an allergic reaction because it has the opposite effect on the body to the chemical mediators produced during a severe allergic reaction. So it raises blood pressure, reduces leakiness of small blood vessels so reducing or reversing allergic swelling. It relaxes



smooth muscles in the airways relieving asthma/ shortness of breath. Families should receive training on when to give adrenaline during a reaction, and how to do this.

Children with allergies should always go to hospital for further monitoring and possible treatment after they have been given adrenaline. A second injection of adrenaline may be given after 5 minutes, but this is rarely required. Other treatments that may be used include antihistamine or steroid tablets. Most children with anaphylaxis will not require admission to hospital but will be observed for 4-6 hours before being sent home.

Unusually, some children may have biphasic reactions. This means the reaction occurs in 2 phases, there is the initial reaction, which may resolve quite quickly and a further similar reaction within 24 hours, which may be more severe than the initial phase. Children who suffer anaphylaxis are also routinely given antihistamines and corticosteroid tablets, which may help to prevent a biphasic reaction.

### **Other treatments used in children at risk of anaphylaxis**

- Corticosteroids - e.g. soluble prednisolone tablets – short course used after anaphylactic reactions. Sometimes children with asthma may have a worsening of their asthma symptoms several hours after the allergic reaction, known as a 'late phase' or 'biphasic reaction'. Corticosteroids are given to prevent this from happening
- Antihistamines – e.g. chlorpheniramine syrup, may be prescribed to use in the event of an allergic reaction. This

medication helps to relieve mild symptoms such as itchiness hives and reduce flushing associated with an allergic reaction

- see: [Care and mobility considerations](#)
- see: [Types of food allergy](#)

**What evidence is available?**

<b>Sources of Evidence</b>	<b>Information Provided</b>	<b>Limitation of Evidence</b>
GPFR	Many children will have been seen only once or twice at the hospital for diagnosis of their allergy. The GP will provide day to day care and medical advice. The GP will have access to the hospital out patient letters and should be able to confirm whether the child has one or more allergies, what the child is allergic to and whether that child is at risk of anaphylaxis or has had an anaphylactic reaction	The GP may not provide details of how the allergy was diagnosed. However, if the GP confirms an allergy is present and is prescribing adrenaline autoinjectors, it is likely that there is a risk of anaphylaxis.
Hospital FR	The allergy specialist is likely to be able to confirm the presence of allergy because they will have access to the hospital records containing records of the allergy tests performed. They are likely to be able to confirm whether there is a risk of anaphylaxis. They are also likely to be able to give a reasonable description of the effects of the allergy on the child and their family even if they have not seen them for several years. An allergy specialist will be able to diagnose and define the risk of anaphylaxis for conditions where allergy tests would normally be negative e.g. chronic urticaria or food and exercise induced anaphylaxis.	If the allergy has got worse and is now causing multiple problems that are being managed by the GP the HFR is unlikely to reflect this.

**How long will the needs last?**

## Allergy and anaphylaxis Duration Guidance

Food Allergy with a risk of anaphylaxis where a moderate functional restriction is identified or a severe functional restriction is identified because the child has more than 2 specific food allergies

Food Allergy with a risk of anaphylaxis where a severe functional restriction is identified due to the co-existence of severe asthma, previous PICU admission or reactions to environmental food allergens.

Idiopathic (of no known cause) anaphylaxis

Latex allergy

Insect venom allergy bee sting/wasp sting

Pet dander

Drug allergy

Hay fever or allergic rhinitis

Oral allergy syndrome

This guidance covers -:

Allergy - with a known risk of anaphylaxis
Allergy - risk of anaphylaxis unknown or not fully assessed
Allergy - no risk of anaphylaxis
Oral allergy syndrome
Food intolerance
Angioedema
Immune system – Other disease of /type not known
Urticaria

**Food Allergy with a risk of anaphylaxis where a moderate functional restriction is identified or a severe functional restriction is identified because the child has more than 2 specific food allergies**

Age at date of claim	Award period
3 - 8	Award to age 8 (or for 1 year, whichever is the longer)

The risk of anaphylaxis continues for many years, often into adulthood. Children at risk of anaphylaxis aged 3 to 8 will require extra care and supervision to avoid anaphylaxis until they are 8. At age 8 they can be expected to avoid allergens and communicate reliably with adults about their allergy. Constant supervision or care from another person will probably not prevent reactions beyond this age even where multiple food allergies are present. The child could be expected to understand their condition, cooperate with avoidance measures and can always choose to be safe by not eating.

Time limited awards are recommended to age 8 in moderate functional restriction. Extension of care needs beyond age 8 will be unusual. Such needs are only likely when the child has developed more severe allergy and now has a severe functional restriction or because the child has learning or behavioural difficulties that prevent them from practicing allergen avoidance for themselves.

Time limited awards are recommended to age 8 in severe functional restriction caused by multiple food allergies. This is because the child can be expected to cooperate with avoidance measures taken by their carers and practice avoidance themselves by age 8. Extension of care needs beyond age 8 will be unusual. Such needs are only likely when the child has developed more severe allergy and now has a severe functional restriction for another reason other than multiple food allergies or because the child has learning or behavioural difficulties that prevent them from practicing allergen avoidance for themselves.

**Food Allergy with a risk of anaphylaxis where a severe functional restriction is identified due to the co-existence of severe asthma, previous PICU admission or reactions to environmental food allergens.**

Age at date of claim	Award period
3 - 11	Award to 12th birthday (or for 1 year, whichever is the longer)

Children who react to environmental contamination with anaphylaxis may require care or supervision beyond age eight. These are children with severe functional restrictions. How much care is required can be determined

from medical evidence from the treating specialist or the school. If continuing needs are identified, a renewal should be conducted at the 12th birthday. This is because at 12 many children who are at risk due to severe asthma will have much improved; those that have not improved could be expected to substantially manage their own asthma and their own EpiPen in most cases. Children who still have anaphylaxis related to environmental contamination may still require help or supervision beyond this age because they have behavioural or learning difficulties or may require someone to accompany them outside the home because of their allergy – there should be medical evidence available to confirm that this is necessary.

### **Idiopathic (of no known cause) anaphylaxis**

<b>Age at date of claim</b>	<b>Award period</b>
3 - 11	Award to 12 <sup>th</sup> birthday (or for 1 year, whichever is the longer)

Medical evidence from the treating hospital consultant will confirm the diagnosis, frequency and severity of anaphylaxis and care needs. Some children will be unable to use their adrenaline autoinjector in time because of the rapid onset of their reactions. Others will be able to do this for themselves from the age of 12. Duration of award should reflect the medical evidence from the treating doctor.

### **Latex allergy**

Severe latex allergy with anaphylaxis from environmental latex is now rare in children. Anaphylaxis due to latex allergy is most likely to occur, in the current generation of children, during hospital or dental treatment. The diagnosis and the severity should always be confirmed by seeking medical evidence from the treating consultant. Awards are recommended until age 12 and should be guided by the medical evidence from the treating doctor.

### **Insect venom allergy bee sting/wasp sting**

Although this type of allergy carries a risk of anaphylaxis there are no additional care or mobility needs associated with it. This is because contact with insects is not an unavoidable part of every day life. Carrying the adrenaline

autoinjector out of doors and providing emergency treatment in the unlikely event of an insect sting will not be onerous or time consuming.

### **Pet dander**

Anaphylaxis to pet dander is very rare. Pet dander can be avoided.

### **Drug allergy**

Some drugs cause allergic reactions ranging from a rash to anaphylaxis. Many more people think that they are allergic to penicillins than actually have the allergy. Assessment by an allergy specialist will clarify this. Although this type of allergy carries a risk of anaphylaxis there are no additional care or mobility needs associated with it. This is because the drug in question can be avoided. Carrying an adrenaline autoinjector is not necessary.

### **Hay fever or allergic rhinitis**

Hay fever is caused by an allergic reaction to airborne plant allergens such as grass or tree pollen. Symptoms include sore itchy, red watering eyes and similar nasal symptoms. These symptoms are annoying but there is no risk of anaphylaxis. Extra care and supervision is not required.

### **Oral allergy syndrome**

Oral allergy syndrome is a condition that only occurs in people with bad hay fever. It is a mild form of allergy. People with this condition develop allergic reactions to certain fruits during the hay fever season. The reaction to fruits includes lip swelling from eating the fruit or touching it to their lips. These symptoms are annoying but there is no risk of anaphylaxis. Extra care and supervision is not required.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Incidence/prevalence**

Overall, a quarter of the childhood population suffer from allergy of one type or another. The majority have mild symptoms such as mild hay fever. Up to

7% of all children have allergic reactions to foods but most reactions are mild. The majority of children will have begun to grow out of their food allergy before they start school. Egg (2.5% of the population), milk (2.5-3.8%) and peanut (2%) are the most common triggers of food allergy reactions in young children.

The prevalence of allergic disease, particularly the incidence of anaphylaxis has increased dramatically over the last 30 years. A UK study showed that rates of hospital admission for anaphylaxis increased from 6 admissions per year per million of population in 1990 to 41 admissions per year per million of population in 2000. There is evidence that rate of increase is tailing off and incidence of new allergy is stabilising at the new higher level. From population-based data, the incidence rate, where this is defined as the number of episodes of anaphylaxis that occur in a defined population over a given period of time, is estimated at between 80-210 episodes per million person-years. Incidence varies by age, gender, geography and socio-economic position.

The most common trigger for anaphylaxis in childhood is food (41-57%), followed by medicinal drugs (6%-34%). The most frequent food triggers in childhood are peanut and nuts, cows milk, hen eggs, fish and shellfish. Anaphylaxis due to other triggers such as insect venom or exercise is rare.

Around 1-2% of adults have diagnosed IgE mediated allergies that could result in anaphylaxis. Peanut, nut and fish allergies are the most common severe allergy in the adult population. A range of other allergies develop over time in older children and adults. Examples include allergies to fruits, drugs and insect venom. Consequently allergies in adults and older children are more diverse.

## **Care and mobility considerations**

### **Food Allergy**

'The acquisition, preparation and consumption of food are a fundamental and unavoidable part of life.' [Hourihane] Consequently this issue of allergy to food cannot be avoided or ignored but must be faced every time the child eats. It can significantly increase care needs of a child with a food allergy.

The effect of the allergy on care will depend on the age of the child and the severity and number of food allergies diagnosed.

### **Food allergy related care needs - children aged 3 to 8**

Children diagnosed with an allergy where there is no risk of anaphylaxis as assessed by the treating allergist will not require significantly more care. This is a mild condition.

For more information see: [Various scenarios](#)

Children who have been diagnosed as at risk of anaphylaxis will require extra care and supervision at all times until they are older than a child without allergies. All children require constant supervision until the age of 3 because of the risk of choking on food and non-food items. This supervision will continue for a child at risk as the parent will be monitoring for an additional hazard (food allergen) that the child has not yet learnt to avoid.

The child at risk will require extra supervision to prevent allergen exposure until she/he can reliably avoid known hazards or communicate hazards to people she/he does not know well. At age 5, normally developing children can communicate well with people they know and generally avoid hazards by themselves. At age eight, they can reliably communicate with people they do not know well. A child could be expected to take on avoiding allergens and communicating effectively about their allergy between the age of 4 and 8 years. Supervision is required during waking hours and it would be expected that the child's bedroom is kept free from potential hazards at night. It would also be expected that the home environment is generally kept allergen free but most accidental food allergy reactions occur at home. Depending on the severity of the allergy, the parent may not feel comfortable allowing the child to be supervised by people the child does not know well because of worries over communication related to avoidance of allergens or an allergic reaction. Children can communicate more reliably with strangers by age 8 years. Children with learning and behavioural difficulties may require supervision beyond this age. Such supervision of allergic children may seem excessive but as most fatal reactions occur outside the home and to teenagers and young adults, it may be that this extra care substantially prevents severe reactions at a young age. The diagnosis of an allergy will certainly cause anxiety and extra care and supervision will be needed to prevent harm to young children at risk.

### **Severity of the functional restriction**

For any child with a food allergy, avoidance of the allergen will be key to continuing health and this may be very easy to do and the consequences of failing to avoid the food may be mild. The main assessment will be whether the child has been diagnosed with a food allergy and whether the medical evidence supports a risk of anaphylaxis. If there is a risk of anaphylaxis, the



difficulty of avoiding the allergen should be assessed. Table 2 in the link below provides information about the risk of anaphylaxis associated with certain foods and with environmental contamination.

Supervision required will depend on what dose of allergen is required for anaphylaxis to occur and whether the child reacts to food allergens in the environment without eating the food. The vast majority of children will need to eat a food item containing the allergen to have anaphylaxis - even if only a trace of the allergen is present in the food. This means a child can keep themselves safe by not eating if necessary. This is a moderate condition.

For more information see: [Various scenarios](#)

A child who reacts to traces of allergen by skin contact, for example who reacts with anaphylaxis after grasping a door handle with traces of peanut protein on it will find it much harder to keep safe. This is a severe condition.

For more information see: [Various scenarios](#)

In these cases anaphylaxis may occur for no obvious reason but the assumption is the child reacted to traces of food allergen in the environment. Such severe reactions to environmental contamination with allergens are very rare and should be supported by clear medical evidence from an allergy specialist. It should be clear that anaphylaxis has actually occurred, and not for example just a generalised rash. Alternative diagnoses such as idiopathic or exercise induced anaphylaxis also need to be ruled out. Children with extreme sensitivity will require meticulous attention to the contents of their diet like other children at risk. However they are likely to require more care and attention from parents because they are more at risk outside the home. For example a parent may need to accompany them on school trips to provide supervision to ensure avoidance and treatment in case of a reaction. Such a child might be home schooled because of their allergy.

### **Typical food preparation practice**

Typically parents of children at risk of anaphylaxis will be very knowledgeable about their allergy, the foods that may contain the allergen, about food labelling regulations and food manufacturing methods. In order to avoid reactions families will often prepare most meals from scratch at home. This is the only way to ensure meals are not contaminated. Pre-prepared meals and sauces for home use or in catering often contain many different ingredients and unless clearly stated on the box there is always the possibility of contamination. A suitable item might list 'prepared in a nut free facility'. Such labels are much less common than 'may contain nuts'. A common problem when eating out for severely nut and fish allergic people is contamination of their food with traces of nut when their meal has been fried in oil

previously used to fry nuts or fish. Often commercial food outlets will say they cannot guarantee any of the food is safe.

Families with allergic children are therefore less likely to eat out and less likely to allow their children to visit other people's houses in the absence of a parent. In addition to keeping the home free of known allergenic foods and cooking from scratch, parents will often prepare safe food for their children to take with them when they visit friends and relatives or go to school. The time commitment per week is considerable. The time, care and attention paid to meals is fairly uniform across children at risk of anaphylaxis who have never reacted and children who have had severe reactions. Provision of safe food by parents is likely to continue until the child starts to take control of their own diet. At age 8 the child can be relied upon to understand the need to eat only safe food even when the parent is not there to remind them. If the child is allergic to multiple allergens time and care will be the same but providing a nutritious diet will be more challenging with limited ingredients.

### **Care required related to medication**

Children at risk will be expected to follow the rule of eating only safe foods and only eating food when an adrenaline autoinjector is available. The parent or child will need to carry this with them at all times along with any other medication such as oral antihistamines. Carrying the adrenaline autoinjector is only a small part of the main care required. The main care is supervision

to maintain allergen avoidance until the child is able to take on this role themselves.

### **Types of food allergy**

Name of food	Features of allergy & risk of anaphylaxis	Do young children tend to grow out of this allergy?	How common is anaphylaxis to environmental contamination?
Peanuts	Likely to be a risk of anaphylaxis. Can be very severe particularly in older children who may react to minute traces of nuts from cross contamination of food during preparation. About 30% of peanut allergic children are also allergic to tree nuts	No – only up to 5% of children grow out of this allergy. Young children with mild allergy are most likely to resolve.	It is very rare for children to react to contact with peanuts (including airborne nut proteins). This should be confirmed in the medical evidence.
Tree nuts	Likely to be a risk of anaphylaxis in most cases. Tree nuts include Brazil, hazelnut, cashew, almond, walnut, pecan and macadamia. The symptoms are the same as for peanut allergy and there is a risk of anaphylaxis. Can be very severe particularly in older children who may react to minute traces of nuts from cross contamination of food during preparation. A person with tree nut or nut allergy may also be allergic to peanuts.	No – only those with the mildest allergies will grow out of it.	It is very rare for children to react to contact with tree nuts (including airborne nut proteins). This should be confirmed in the medical evidence.

Egg	<p>Likely to be a risk of anaphylaxis in a minority of cases. Very common in young children – the majority have mild reactions with hives and facial swelling when lightly cooked egg is eaten. The majority grow out of this by age 5 years. As their allergy resolves, they can tolerate baked egg products first, followed by lightly cooked egg. A small number have persistent allergy to cooked egg as well as raw egg, Reacting to cooked egg severely limits their food options. Reactions in a small number may be severe involving collapse and difficulty breathing, some may be severely affected by small amounts of contamination in foods.</p>	<p>Yes, most cases will be mild and will have started to resolve by age 3 years. Children who react severely are less likely to grow out of it.</p> <p>Children with persistent egg allergy tend to have more severe reactions on re-exposure.</p>	<p>A small number of children will react to contact with eggs with anaphylaxis, this is also very rare. These children are unlikely to grow out of their egg allergy. This should be confirmed in the medical evidence.</p>
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<p>Cow's milk (Dairy)</p>	<p>Likely to be a risk of anaphylaxis in rare cases. Is very common in young children – commonly causes mild reactions involving hives and lip or facial swelling when milk or milk containing products are ingested. The majority will grow out of this by age 3. A small number have severe allergy and may be severely affected by tiny amounts in contaminated foods.</p>	<p>Yes, most cases will be mild and will have started to resolve by age 3 years. Children who react severely are less likely to grow out of it.</p> <p>Children with persistent milk allergy tend to have more severe reactions on re-exposure.</p>	<p>A small number of children will react to contact with milk proteins with anaphylaxis. Milk proteins are ubiquitous in environment where milk is regularly consumed – both inside and outside the home These children are unlikely to grow out of their allergy. This should be confirmed in the medical evidence.</p>
<p>Soy</p>	<p>Common in young children and may co-exist with milk allergy in the same child. Usually causes food intolerance or mild allergic reactions when soy or soy containing products are ingested. The majority will grow out of this by age 3. Anaphylaxis caused by soy protein is extremely rare in the UK.</p>	<p>Yes, in most cases children will grow out of soy allergy.</p>	<p>No</p>
<p>Sesame</p>	<p>May cause anaphylaxis and be difficult to avoid</p>	<p>Rarely resolves.</p>	<p>Rare.</p>
<p>Wheat</p>	<p>Is uncommon in young children – it can cause swelling and hives, or more commonly intolerance type reactions. Anaphylaxis due to wheat is rare.</p>	<p>Yes.</p>	<p>No</p>

Fish	Likely to be a risk of anaphylaxis in most cases. May cause severe reactions. Cross contamination of fish products is common so may be advised to avoid fish and shellfish even if not allergic to shellfish. May have severe reactions to traces of fish allergens for example eating food cooked in oil previously used to fry fish.	No. Rare in young children. Tends to develop in teenagers and young adults and it does not resolve.	Fish is relatively easy to avoid as its presence in food is usually obvious.
Shellfish	Likely to be a risk of anaphylaxis in most cases. May cause severe reactions. Cross contamination of fish products is common so may be advised to avoid fish and shellfish even if not allergic to fish. May have severe reactions to traces of fish allergens for example eating food cooked in oil previously used to fry shellfish.	No. Rare in young children. Tends to develop in teenagers and young adults and it does not resolve.	Shellfish is relatively easy to avoid, as its presence in food is usually obvious.
Fruit	<p>Most often causes mild reactions with lip swelling or mouth itching as part of 'oral allergy syndrome' (OAS), OAS does not lead to anaphylaxis.</p> <p>Emerging severe allergy to fruit, especially kiwi with anaphylaxis</p>	<p>Usually long lived allergy, cooked fruit tolerated in OAS</p> <p>Severe fruit allergy rarely resolves.</p>	No even in severe allergy. Reactions to environmental allergens are likely to be mild e.g. facial swelling or hives.

Pulses – peas, beans and lentils	Can cause spectrum of reactions from mild to anaphylaxis. Avoidance can com- promise diet in vege- tarians.	Rarely resolves.	No even in severe allergy, reactions to environmental aller- gens are likely to be mild e.g. facial swell- ing or hives.
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### **Guidelines for which guidance to use**



Condition claimed	Effects of condition	Which guidance
Allergy to a food	Reacts to specific foods in minutes to hours.	Check medical evidence for diagnostic information and use this guidance if allergy diagnosed with specific IgE blood test or skin prick test.
Urticaria	<p>May be related to an allergy or may not.</p> <p>Do not use this guidance if medical evidence does not confirm allergy</p>	Check medical evidence for diagnostic information and use this guidance if allergy diagnosed with specific IgE blood test or skin prick test. Most cases of urticaria will not be due to underlying allergy and will be labelled 'chronic urticaria'.
Angioedema	<p>May be related to an allergy or may not. Do not use this guidance if medical evidence does not confirm allergy.</p> <p>Usually not allergy if diagnostic information is 'angioedema'</p>	Check medical evidence for diagnostic information and use this guidance if allergy diagnosed with specific IgE blood test or skin prick test. Most cases will be a non allergic condition due to inherited enzyme deficiency (hereditary angioedema) causing spontaneous swelling and abdominal pain. Refer to medical services.
Anaphylaxis		Use this guidance.
Hay fever or allergic rhinitis	Rhinitis and conjunctivitis may be associated with asthma.	If asthma is present use the <a href="#">asthma</a> guidance. Use this guidance for hay fever.

Oral allergy syndrome	A condition associated with hay fever – allergic swelling of the lips often to fruits during the hay fever season.	Use this guidance.
Insect venom allergy bee sting/wasp sting		Use this guidance.
Drug allergy		Use this guidance.
Food intolerance	Symptoms develop hours to days after eating the food.	Use this guidance.
Asthma	Wheezing and difficulty breathing due to an allergy	Use the <a href="#">asthma</a> guidance and check medical evidence for diagnostic information. Use this guidance if allergy diagnosed with specific IgE blood test or skin prick test and there is a risk of anaphylaxis.
Eczema	Dry, itchy, inflamed skin	Use the <a href="#">eczema</a> guidance for information on eczema care in all cases. Only use this guidance if the claimed allergy causes other effects including a risk of anaphylaxis in addition to eczema. Check medical evidence for diagnostic information and use this guidance if allergy diagnosed with specific IgE blood test or skin prick test.

## Substances that cause allergy – ‘allergens’

### Food Allergy

Food allergy is common in very young children with 1 in 20 toddlers affected. The incidence in older children is much lower because children tend

to grow out of their allergies. It is not known why food allergies develop but they occur more commonly in children with eczema or a family history of allergy. Children are most commonly allergic to a small group of foods, these account for 90% of food allergies in children. Allergy to other foods is less common, but the pattern is changing with allergies that were rare a few years ago becoming more common (e.g. kiwi and pulses).

The majority of children with food allergy have mild reactions; symptoms include lip swelling and urticaria. Even though the reactions are not life threatening they are distressing and food allergy may also significantly exacerbate eczema. Eczema is very common in food allergy and usually appears before the food allergy becomes apparent.

Food allergy has a significant impact on children and families for several reasons. There is a continuous fear of suffering allergic reactions. It becomes harder to provide a safe and nutritious diet for the family once a food allergy is diagnosed. Shopping becomes more time consuming, as all labels have to be checked for allergens and labels that inform of the risk of cross-contamination of purchased foods are confusing. Cross contamination means a food contains traces of an allergen, such as nuts, despite not containing nuts as an ingredient because the factory makes or packages nut products and nut proteins could contaminate the product. Similarly eating out at other people's houses or in restaurants becomes a source of stress and requires planning. As a consequence the quality of life of affected families is adversely affected. The effect on activities will depend partly on whether the child reacts to minute traces of a food making reactions much more difficult to avoid and whether they have more than two food allergies. More than two food allergies significantly increase the difficulty of providing a safe and nutritious diet. It increases the risk of reactions and research shows it has a much greater impact on life than one or two food allergies alone. Nuts and peanuts count as one allergy in this context.

Table 2 in the link below provides information about the risk of anaphylaxis associated with certain foods and with environmental contamination.

For more information see: [Types of food allergy](#)

### **Insect venom allergy bee sting/wasp sting**

Anaphylaxis after insect stings does occur but is rare in childhood. Desensitisation therapy for this type of allergy is available from allergy specialists, but is rarely required. This type of treatment can either prevent further reactions after an insect sting or reduce the severity of any reaction experienced. Children who have large local reactions after an insect sting are not

at significant risk of future anaphylaxis. An allergy specialist will be able to provide a risk assessment.

### **Drug allergy**

Medicinal drugs may cause allergic reactions, most commonly these are mild rashes and anaphylaxis occurs rarely. Allergy to penicillin drugs is commonly claimed, but when investigated by a specialist, allergy is often not present.

### **Pet dander**

Allergy to cats and dogs may cause worsening asthma control over time, which may be resolved by re-housing the pet. Rarely, a child may suffer a severe allergic reaction when visiting another house with a pet but once the diagnosis is made, this circumstance should be straightforward to avoid. Repeated severe allergic reactions may occur rarely in circumstances where avoidance is not possible (e.g. when a parent's career depends on animal contact). In this case extra care is required by the parent to change clothes and shower on arrival to the home.

### **Latex allergy**

This condition is now rare in children due to the increasing use of non-latex products in healthcare. Affected children most commonly develop mild reactions with urticaria and swelling after contact and anaphylaxis is extremely rare. Latex allergy is often blamed when children have otherwise unexplained reactions, usually because there has been a suspicion of latex contact from the environment. However, allergy testing should be performed as this association is often shown not to be genuine. The child should be assessed by an allergy specialist before this diagnosis is accepted.

### **Hay fever or allergic rhinitis**

Rhinitis (commonly known as Hay fever) is usually caused by an allergic reaction in the airway and eyes to airborne plant allergens such as grass or tree pollen. This causes seasonal symptoms with worsening in the Spring and/or Summer. Allergic rhinitis can also be stimulated by 'perennial' allergens such as house dust mite or pet dander and in this case the symptoms are present all year round. Non-allergic rhinitis is also common and in this case there is no allergen to blame but the symptoms are identical. Symptoms include sore itchy, red watering eyes, nasal itching, running and blockage. These can be particularly troublesome, resulting in poor sleep, lethargy, missed school days and reduced examination performance. Hay fever

itself is not life threatening but it is often associated with asthma. Allergic rhinitis may worsen asthma control and indirectly contribute to a severe allergic reaction. Allergic rhinitis in the Spring, stimulated by tree pollen is also part of the 'oral allergy syndrome' (see below).

### **Oral allergy syndrome**

Oral allergy syndrome is a condition that occurs in people with allergic rhinitis to tree pollen (causing spring hay fever). People with this condition develop allergic reactions to certain fruits. Symptoms include lip swelling after eating the fruit, but cooked fruit is usually tolerated. In oral allergy syndrome reactions are usually limited to lip swelling and progression to difficulty breathing or collapse is uncommon. However, a related allergy to fruits is increasingly recognised in childhood where reactions can be severe, characterised by throat tightening and respiratory difficulty (e.g. to kiwi or peach) and there is a risk of anaphylaxis, although usually the trigger is easy to avoid.

### **Angioedema**

Angioedema is a name used for swelling of deep tissues anywhere on the body, but it most commonly affects the face, the hands and the genitals. Swelling of these areas is not associated with redness like the more superficial swelling seen with hives/urticaria, which also involves swelling of the skin. In practice, the term angioedema is mainly used to describe dramatic swelling of the face. Angioedema is one symptom of allergy but does occur in other conditions where allergy testing may be negative.

Hereditary angioedema is a life-long condition caused by an inherited deficiency of the enzyme 'C1 esterase inhibitor'. There is usually a family history of an affected member and the disease usually presents in the second decade of life with spontaneous angioedema or swelling (urticaria does not occur, as a rule). Swelling can appear spontaneously or after injury or surgery (e.g. tooth extractions). Angioedema can occur anywhere but may be life threatening if it affects the upper airway. Swelling in the bowel wall can cause repeated episodes of severe abdominal pain. Allergy tests are negative in this condition; diagnosis is by a specialised test for the defective enzyme in the blood, which is either reduced or poorly functioning. Treatment of acute episodes is with infusions of fresh frozen plasma or concentrated enzyme replacement.

### **Urticaria**

Urticaria is the name given to the itchy raised skin rash otherwise known as hives, and is often described by patients as similar to a nettle sting rash. Urticaria has many causes. It is a common symptom of food allergy or may occasionally be caused by contact with grass in pollen-allergic children. Allergy tests will usually be informative. There is a distinct condition known as

chronic urticaria and angioedema in which the child suffers repeated episodes of rash and swelling which appears identical to an allergic reaction (each episode typically lasts longer than 24hrs) but there is no allergic cause. It usually arises in a child with an insignificant history of allergy. Sometimes patients identify a physical trigger such as infection, heat, cold, sweating or exercise. Allergy tests in this condition are usually negative and so the diagnosis relies on exclusion of other allergies by a specialist.

### **Food intolerance**

Food intolerance is any reaction to a food not related to allergy. Food intolerance may be caused by absence of an enzyme needed to digest the food – e.g. lactase deficiency means the enzyme lactase used to digest lactose, a milk protein, is absent and drinking milk causes diarrhoea. It may also be caused by a substance present in several foods for example sulphites in wine might exacerbate asthma in susceptible people. There are many different types of food intolerance, which may cause significant and disabling symptoms when the foods are eaten but reactions to traces of foods and immediately life-threatening reactions to such traces do not occur.

### **Asthma**

Asthma is often caused by allergy in children. You should consult the guidance on asthma if asthma alone is claimed. If asthma is the only symptom of the allergy and there is no risk of anaphylaxis just use the asthma guidance to assess the case.

### **Eczema**

Eczema is sometimes caused by allergy in children. If eczema is a symptom of the allergy and there is no risk of anaphylaxis just use the eczema guidance to assess the case.

### **Food and exercise induced anaphylaxis**

In this uncommon condition, a food which is normally tolerated (usually wheat or shellfish) can cause anaphylaxis if the patient exercises within four hours after ingestion of the food. The exercise is usually out of the ordinary but may in fact be relatively mild. Avoidance of exercise for four hours after

eating is the only way to prevent reactions. This condition usually requires allergy specialist diagnosis.

### **Idiopathic anaphylaxis**

Idiopathic (of no known cause) anaphylaxis is a rare condition where the child undergoes episodes of spontaneous anaphylaxis - there is no allergic trigger. This is a frightening condition as there are no clear preceding triggers, which the patient can avoid. The diagnosis is by exclusion of allergic and physical triggers and should be made by an allergy specialist.

### **Food protein induced enterocolitis syndrome (FPIES)**

This rare syndrome usually begins in infancy. Affected children develop severe diarrhoea after eating any food protein, as a consequence they rapidly lose fluid from their circulation and present with shock. The appearance can be of anaphylaxis although the mechanism is unknown. Allergy tests are usually negative. Great care is required to avoid sources of food protein in the diet. The condition can resolve in early childhood.

### **Other conditions that may be claimed under allergy and anaphylaxis**

#### **Angioedema**

Angioedema is a name used for swelling of the deep tissues of the face, the hands and the genitals. It can be one of the signs that there is a risk of anaphylaxis. There are other causes of this condition. If a risk of anaphylaxis due to allergy is identified please follow the appropriate guidance on the allergy concerned, e.g. food allergy. Hereditary angioedema can cause sudden life-threatening reactions either spontaneously or after trauma or surgery – refer to medical services.

#### **Urticaria**

Urticaria is the name given to the itchy raised skin rash otherwise known as hives. Nettle rash is an example of urticaria or hives. It can be one of the signs that there is a risk of anaphylaxis. There are other causes of this condition. If a risk of anaphylaxis due to allergy is identified please follow the appropriate guidance on the allergy concerned, e.g. food allergy.

#### **Food intolerance**

Food intolerance is any reaction to a food not related to allergy. This condition is not associated with a risk of anaphylaxis or with reactions to traces of

food. Consequently although care will be taken with diet by the parents the supervision required to prevent anaphylaxis in young children is not necessary and older children can equally avoid the foods to which they are intolerant.

### **Asthma**

Asthma is most often caused by allergy in children. You should consult the guidance on asthma if asthma alone is claimed. If a risk of anaphylaxis due to allergy is identified please follow the appropriate guidance on the allergy concerned, e.g. food allergy in addition to the asthma guidance, care needs may be additive.

### **Eczema**

Eczema is sometimes caused by allergy in children. If eczema is a symptom of the allergy and there is no risk of anaphylaxis just use the eczema guidance to assess the case. If a risk of anaphylaxis due to allergy is identified



please follow the appropriate guidance on the allergy concerned, e.g. food allergy in addition to the eczema guidance, care needs may be additive.

You must take all information into account when considering the duration of care and mobility needs. You must base the duration of care and mobility needs on the particular circumstances of the child.

<b>Mild functional restriction</b>	<b>Moderate functional restriction</b>	<b>Severe functional restriction</b>
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<p>A 7 year old boy has hay fever and during the hay fever season takes regular antihistamines and steroid nose drops to control his itchy watery eyes and runny nose. Last Spring his lips swelled up dramatically when he ate an apple. Because of this his parents and GP were worried that he might develop anaphylaxis. He was prescribed an EpiPen, advised not to eat apples and referred to the Consultant Allergist for tests. The Consultant diagnosed hay fever and oral allergy syndrome that explained the lip swelling from eating apples.</p> <p>He is not at risk of anaphylaxis because he has oral allergy syndrome although he does have a food allergy to apples, he does not need to carry an EpiPen.</p>	<p>A 3 year old girl developed lip swelling and hives every time she ate dairy products from the age of 9 months. Each episode of lip swelling and hives was followed by a flare up of her otherwise mild eczema. Her mother cut out dairy from her diet. When she was 12 months old she had a similar reaction to peanut butter. The GP prescribed an EpiPen and advised her to completely avoid milk and peanuts and referred her to a specialist. She had skin prick testing by the specialist when she was 18 months old and skin prick tests to cow's milk and peanut were positive. She had anaphylactic shock after eating a pie when she was 2. Her face swelled up, she was blotchy and sweaty and 'not all there'. Her mother used the EpiPen and by the time she got to A&amp;E 15 minutes later she had almost recovered. She was observed for 4 hours and sent home with tablets. She has not had anaphylaxis since. She attends nursery for half days only because of her allergies and the nursery staff have had EpiPen training to accommodate her. Her parents and grandparents are the only people who cook for her. She will be</p>	<p>A 5 year old girl developed lip swelling and hives when she ate cow's milk yoghurt at age 9 months. Her brother has a food allergy so her mother knew she should stop giving her dairy and she substituted soy products. The little girl reacted similarly to soy products and goats milk. Each episode of lip swelling and hives was followed by a flare up of her otherwise mild eczema. She then had anaphylactic shock after eating home made cod and potato when she was 12 months old. She was treated in A&amp;E and admitted over night. 2 weeks later, she had skin prick tests that showed she was allergic to cow's milk, goat's milk, soy, lentils and fish. She has had anaphylactic shock approximately once a year since then. She was diagnosed with asthma 6 months ago and was admitted to PICU 3 months ago with anaphylaxis after eating potato chips fried in oil that had been previously used to fry fish.</p> <p>This little girl has a severe functional restriction because she has 3 or more specific food allergies. In most</p>
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	<p>having a food challenge test soon because her skin prick tests show that she is growing out of her cow's milk allergy.</p> <p>She remains at risk of anaphylaxis from her peanut allergy. She is in the moderate category because she is at risk of anaphylaxis due to food allergy but she does not have severe asthma and has not been admitted to PICU.</p>	<p>cases the award duration for a child like this should be to age 8 but she has been admitted to PICU and will need more time to learn how to manage her medication as well as avoidance so her award duration should be to age 12.</p>
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What you need to know about Anorexia Nervosa

**What is Anorexia Nervosa?**

Anorexia nervosa is one of the eating disorders. It is usually characterised by a fear of...

[What is Anorexia](#)

**What are the effects and signs?**

Although this condition begins with dieting that initially appears to be within normal limits, it soon .....

[Effects of Anorexia](#)

**How is it assessed?**

Children and adolescents with suspected anorexia nervosa will usually present to ....

[Assessment of Anorexia](#)

**How is it treated and managed?**

The majority of children and adolescents with anorexia nervosa will be treated on an....

**[Treatment of Anorexia](#)****What evidence is available?**

Physical examination will identify physical effects of low weight. For moderate to severe cases it ...

**[Evidence](#)****How long will the needs last?****[Prognosis and duration of award](#)**

What is Anorexia?

Anorexia nervosa is one of the eating disorders. It is usually characterised by a fear of fatness and weight gain, body image distortion and a strong desire to be thin, which drives food restriction resulting in low weight. This can be expressed verbally or non-verbally through a child/young person's behaviour.

Children and young people with anorexia nervosa are typically preoccupied with their weight, food and eating. Typical behaviours could include calorie counting, body checking and frequent weighing. In order to achieve a low weight, they either restrict the amount of food they eat, that is, typically eating only small amounts of high calorie and fat foods or they may cut down their intake of healthy foods. This is known as the restrictive subtype.

Others can resort to other weight restricting methods to achieve low weight including over exercising, purging behaviours (such as forcing themselves to vomit or taking laxatives, diuretics or diet pills). Amongst children and adolescents, the characteristic loss of weight may not show. The inadequate dietary intake or weight controlling strategies may result in failure to gain weight and grow normally, so the young person falls off the growth centiles. As weight falls, in young women there is resultant cessation of menstruation

or for pre-pubertal girls puberty may be delayed so normal menstruation does not start.

Many children and young people with anorexia nervosa see themselves as overweight even though their actual body weight or Body Mass Index (BMI) is low. If asked, they will deny that they have a problem with their weight.

Body Mass Index (BMI) is a calculation based on the weight and height of an individual as follows:

- divide weight in kilograms (kg) by height in metres (m)
- divide the answer from the above calculation by height again

It has to be remembered that as children and adolescents grow their BMI increases. As children get older and grow, they increase in height and weight, and also their shape changes, reflected in increasing BMI with age. This means that the criteria that are used for adults to identify healthy BMI cannot be used with this younger population. Their BMI has to be looked at on a BMI chart to see how it differs from the healthy BMI in a typically developing same age and sex peer.

The BMI centiles relates to the percentage of the population corrected for age and gender at each BMI.

The cause of anorexia nervosa is not currently known, but most of the causation (70%) is explained by genetic factors, indicating it runs in the family and an increase in risk in first degree relatives occurs. It is known that environmental factors, particularly the circumstances of the individual, also play a part in the development of this disorder.

#### Incidence and Prevalence

The onset of the disease can occur from the age of 8 and upwards, though the peak incidence of cases occurs between the ages of 15 and 19 for girls

and 10-14 years for boys. Anorexia nervosa is much more common amongst girls with a ratio of one male case for every 10-15 female.

The prevalence of anorexia nervosa (including mild forms) is approximately 1% amongst adolescent girls, though disordered eating occurs at higher prevalence.

## Body mass index (BMI)

The BMI centile is a simple and reliable indicator of thinness and fatness in childhood. Where severe overweight or underweight is a concern or where there is a need for monitoring over time, BMI can be calculated and plotted on this chart.

It is important also to plot the height and weight separately on the main 2-18 chart. There is also a BMI centile look-up on the standard 2-18 chart for less complex cases.

BMI is calculated by dividing weight (in kg) by the square of height (in metres, for example, 1.32 m, not centimetres, for example, 132 cm).

A simple way to do this on a calculator or mobile phone is:

1. Enter the weight
2. Divide by height
3. Divide the result by height

The result can then be plotted on the chart below.

## Overweight and obesity

A BMI above the 91st centile suggests overweight. A child above the 98th centile is very overweight (clinically obese) while a BMI above the 99.6th centile is severely obese. In addition to the usual nine centile lines, the BMI

chart displays high lines at +3, +3.33, +3.66 and + 4 SD, which can be used to monitor the progress of children in overweight treatment programmes.

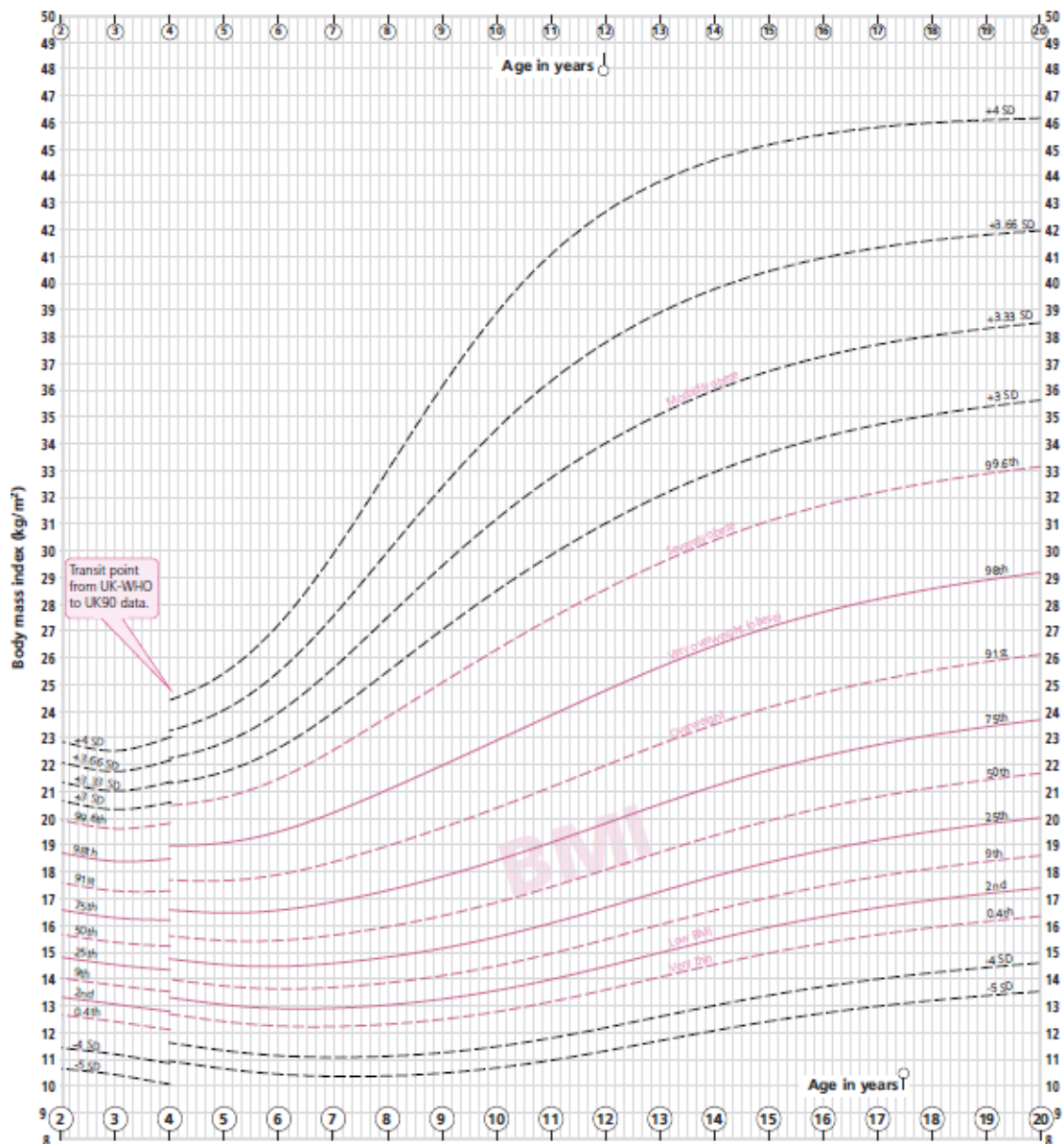
### Thinness

A BMI below the 2nd centile is unusual and may reflect undernutrition, but may simply reflect a small build. The chart also displays low lines at -4 and - 5 SD for those who are severely underweight. Children whose BMI lies





Females 2-20 years of age



What are the Effects and Signs?

[Levels of anorexia](#)

Psychological changes associated with anorexia nervosa

Physical changes associated with anorexia nervosa

Hormonal changes in anorexia nervosa

[Associated conditions](#)

[Long term effects](#)

Although this condition begins with dieting that initially appears to be within normal limits, it soon escalates to more alarming levels. At this point the dieting is often associated with other behaviours such as social isolation,

withdrawal and obsessional behaviours. This can have a detrimental effect on schoolwork.

#### Levels of anorexia

Although there are strict definitions for each level of anorexia, the level of severity may be increased to reflect clinical symptoms, the degree of functional disability and the need for supervision.

#### Mild Anorexia

Percentage median BMI greater than 85 % (approximately above 9<sup>th</sup> BMI centile).

#### Moderate Anorexia

Percentage median BMI 80-85% (approximately 9<sup>th</sup> – 2<sup>nd</sup> BMI centile).

#### Severe Anorexia

Percentage median BMI 70-80% (approximately between 2<sup>nd</sup> – 0.4<sup>th</sup> BMI centile).

#### Extreme Anorexia

Percentage median BMI less than 70% (approximately below 0.4<sup>th</sup> BMI centile).

Psychological changes associated with anorexia nervosa:

- Irritability
- Indecisiveness
- Poor concentration
- Confusion
- Depressed mood
- Anxiety
- Hyperactivity
- Insomnia
- Perfectionism
- Obsessive behaviour
- Social withdrawal

Physical changes associated with anorexia nervosa:

- Emaciation (severe weight loss and loss of muscle and body fat)
- Slow heartbeat and pulse (can be associated with fainting)
- Low blood pressure
- Dizziness
- Bloating
- Constipation
- Swelling of feet and hands
- Dry skin
- Fine facial and body hair
- Some loss of head hair
- Cold feet and hands
- Pale skin
- Cessation of menstruation or failure to start menstruating in young women or general delayed pubertal development
- Changes in thyroid, hormone, kidney or liver blood tests
- Low white blood cells
- Changes in heart rhythm

Hormonal changes in anorexia nervosa

Blood tests performed on those with anorexia nervosa show that most of the body hormones are reduced.

Low thyroid level hormone causes slow heart rate and dry skin. The low thyroid level is as a result of the low weight and starvation and not the cause. Reduced levels of thyroid hormone reduce the metabolic rate of the person

with anorexia nervosa. This can be seen as a method of conserving limited energy supplies.

Abnormal oestrogen and progesterone levels in young women cause problems with menstrual periods. This usually occurs when the body stores of energy reach a low level.

When weight increases and is maintained within the normal range, hormonal levels usually return to normal. As a result periods will usually recommence.

#### Associated conditions

- Mood Disorders including Depression (present in up to 60% of cases)
- Anxiety disorder
- Obsessive Compulsive Disorder (OCD)
- Body Dysmorphic Disorder (BDD)
- Social interaction difficulties
- Long term effects
- Bone fractures and osteoporosis
- Infertility
- Damaged tooth enamel from vomiting episodes
- Retarded growth due to imbalances in growth hormones from starvation

How is it assessed?

The diagnostic criteria for anorexia nervosa are:

1. Significantly low or falling BMI, for example, percentage median BMI less than 90 % or lower and maintained over 3 months (BMI criteria are not fixed in children in view of developmental considerations)
2. Weight loss is self-induced by avoidance of fattening foods, vomiting, exercising and purging
3. Body image is distorted with a dread of being fat
4. Disturbance of normal hormone levels. This includes delayed/absent menstruation in adolescent girls and a lack of sex drive or potency in males

## 5. In pre-pubertal children there is delay in growth

Children and adolescents with suspected anorexia nervosa will usually present to their GP in the first instance. From there, they will be referred to the local Child and Adolescent Mental Health Services (CAMHS) or to a specialist Eating Disorders Clinic. Specialist Eating Disorder clinics will involve the work of paediatricians, psychiatrists, psychologists and specialist dieticians.

The diagnosis itself is based on a history taken from the child along with corroborative information from relatives and friends. A physical examination including height, weight, pulse and blood pressure, peripheral circulation (coldness of extremities) is carried out to measure the level of loss of weight, muscle and body fat. The changes in weight and height and BMI, from previous measures where available, should be plotted on the appropriate growth charts to help identify the onset of the disorder and severity.

Blood tests including, thyroid function, full blood count, kidney and liver function tests and a hormone profile are done. An electrocardiogram is indicated for the more severely affected and for those with abnormal electrolytes (especially potassium).

How is it treated and managed?

[Psychological therapy](#)

[Medication](#)

[Treatment outcomes](#)

The majority of children and adolescents with anorexia nervosa will be treated on an outpatient basis. Only the most severely affected children are admitted for periods of inpatient therapy.

Psychological therapy

Family Interventions

This form of therapy is crucial to the success of any treatment plan. In the initial stages most families will need help with reinforcing boundaries around



eating and exercise to help prevent any further weight loss. Subsequently, any issues which existed before the anorexia nervosa can be dealt with.

### Individual therapy

Cognitive behavioural therapy, interpersonal therapy and individual psychotherapy can all be used. It is however, better for a child to have a therapist they can engage with rather than to be too restrictive about which therapy should be used. Goals in individual supportive psychotherapy are:

- Acceptance of a menu plan
- Acceptance of regular adjustment of a menu plan
- Monitoring of weight and behaviour
- Monitoring of moods, thoughts and feelings
- Challenging unhelpful beliefs about food and nutrition
- Counselling about eating and potentially dangerous methods of losing weight
- Involving the family
- Find other ways for dealing with unpleasant feelings and stress
- Dietetic Input

In severe cases it is important to have the input of a dietician to help the damage that can occur when re-feeding very emaciated children. This re-feeding syndrome can lead to damage to the heart and even death.

In other cases the involvement of the dietician helps reinforce healthy eating and helps to reassure family members that their child is receiving a balanced diet.

### Re-feeding

This is the main way in which a child or young person with anorexia nervosa will gain weight. A menu plan that includes a balanced diet that also includes feared high energy foods in moderation is used. The main aim is for weight gain 300g to 500g a week in outpatients and 1kg to 1.5kg for the more severely under nourished (that is, those usually in hospital).

Weighing is usually carried out by clinic and hospital staff and certainly not done by the patient themselves.

Supervision is needed as some children with anorexia nervosa will attempt to cheat when being weighed or may attempt to cheat on their eating plan.

One method, for example, of cheating to increase weight includes drinking large amounts of water the night before being weighed.

Of those most severely affected, there will be some with anorexia nervosa who are unable to recognise that they are ill and may refuse treatment.

### Medication

There is little evidence to support the use of medication in treating the core condition of anorexia nervosa. Antidepressants like Fluoxetine and Sertraline can be useful in treating associated anxiety or mood disorders, for example, Anxiety disorders.

For severely affected cases and those who are difficult to treat antipsychotic drugs such as olanzapine may be used in combination with other approaches.

Vitamin, mineral and food supplements are useful in the early stages of treatment and will usually be overseen by a dietician. Thiamine, Vitamin B complex should be offered in the first 10-14 days in those who are malnourished. Those at high risk of re-feeding syndrome could be considered for phosphate supplementation after discussion with a consultant psychiatrist.

### Treatment outcomes

The prognosis of anorexia in childhood and adolescence is very variable and there is little research data on long term follow up of this category of patient. Treatment of anorexia nervosa can take between 6 months and 5 years depending on the underlying severity of the condition. The information available indicates:

- approximately 50% of cases will completely recover
- 30 to 40% will recover sufficiently to lead a normal life, but with full recovery occurring after a few years. They may though continue to

have intrusive thoughts about food or have some food avoidance behaviours

- the remaining 10% will go on to develop a chronic long term eating disorder that continues into adulthood.

What evidence is available?

Sources of evidence	Information Provided	Limitation of Evidence
Parent/Carer	First source of information and associated evidence.	Does not have specialist knowledge.
GP	Clinical features, treatment and some information about disability and needs.	May be the only source of information if no other professional involvement.
Children's & Adolescent's Mental Health Services (CAMHS)	Clinical features, treatment and information about disability and needs.	Some information regarding symptoms and disabling effects may be based on what the parent/carer has told the relevant professional. Physical examination will identify physical effects of low weight. For moderate to severe cases it is expected CAMHS will have obtained corroborative reports, for example, school reports.
Eating Disorder Clinic	Clinical features, treatment and information about disability and needs.	Some information regarding symptoms and disabling effects may be based on what the parent/carer has told the relevant professional. Physical examination will identify physical effects of low weight. For moderate to severe cases it is expected CAMHS will have obtained corroborative reports, for example, school reports.

How long will the needs last?

The following table is to be followed if an award is made. It is not a statement of intended awards for individual categories.

• Age at date of claim	• Length of Award		
	• Mild	• Moderate	• Severe (with or without psychotic symptoms)
Aged 6-9	3 years	3 years	3 years
Aged 10-13	3 years	3 years or to age 16 (whichever is greater)	3 years or to age 16 (whichever is greater)
Aged 14-16	to age 16	to age 16	to age 16

#### Example

An 8-year-old child with mild Anorexia nervosa is given an award until they are 11. They must then be re-assessed and any changes in care or mobility needs reviewed in light of any additional therapy they have received.

You must take all information into account when considering the duration of care and mobility needs. You must base the duration of care and mobility needs on the particular circumstances of the child.

#### What you need to know about Anxiety Disorders

<p><b>What are Anxiety disorders?</b> The term anxiety disorder is a general one which covers a number of diagnoses where..... <a href="#">What is Anxiety</a></p>
<p><b>What are the effects and signs?</b> Generalised Anxiety Disorder can cause a change in the child's behaviour and due to a change in..... <a href="#">Effects of Anxiety</a></p>
<p><b>How is it assessed?</b> Children with symptoms of an anxiety disorder will usually see their GP initially..... <a href="#">Assessment of Anxiety</a></p>

**How is it treated and managed?**

Cognitive Behavioural Therapy is the main therapy for all types of anxiety in children. Medication is.....

[Treatment for Anxiety](#)

**What evidence is available?**

Information regarding symptoms and disabling effects may be based on what the.....

[Evidence](#)

**How long will the needs last?**

Symptoms of anxiety are present for more than 50% of the time, but.....

[Prognosis and duration of the award](#)

What are the Anxiety disorders?

The term anxiety disorder is a general one which covers a number of diagnoses where excessive fear and worry are triggered by internal or external events. This can interfere with normal daily activity and can lead to impairment of functioning.

The anxiety disorders are:

Generalised Anxiety Disorders

- Generalised anxiety disorder (GAD)
- Panic disorder

Situational Anxiety Disorders

- Separation anxiety
- Social anxiety
- Selective mutism

Specific phobias

- Agoraphobia

The anxiety disorders have a large overlap. 90% of children with generalised anxiety disorder will have the presence of another anxiety disorder. Of those children with separation anxiety, 60% will also have social phobia or GAD.

**Generalised anxiety disorders****General anxiety disorder (GAD)**

The main feature of this condition is the presence of a generalised, persistent and impairing anxiety not related to any particular circumstance.

90% of those children diagnosed with general anxiety disorder will also have evidence of another anxiety disorder.

Incidence in adolescents (12-18) is 2-5%. Incidence is very low in children.

### **Panic disorder**

This condition is marked by recurrent episodes of severe unpredictable anxiety (panic attacks). Often these attacks are accompanied by physical symptoms: shortness of breath, fast heart rate, sweating, depersonalisation (the child has the feeling that they're observing themselves from outside their body or having a sense that things around them aren't real, or both) and derealisation (an alteration in the child's perception of the environment such that things that are ordinarily familiar seem strange, unreal or two-dimensional).

Incidence in children and adolescents is 2-5%.

### **Situational Anxiety disorders**

#### **Separation anxiety**

This is when an individual experiences excessive anxiety regarding separation from home or from people to whom the individual has a strong emotional attachment.

Incidence is 3-5%. Onset is in late teens.

#### **Social anxiety**

This condition is characterised by significant anxiety in social situations despite the desire for social interaction. The anxiety can impair functioning in all situations.

Incidence is 1-7% in children and adolescents. Onset usually in childhood.

#### **Selective mutism**

Children will demonstrate normal language skills in some situations but will freeze and be unable to talk in others. The child will speak at home and familiar surroundings provided they know whom they are speaking to. As a rule they will be unable to speak to strangers or at school.

Incidence is 0.1%. Onset is in pre-school years.

#### **Specific phobias**

Excessive fears of clearly circumscribed objects or situations that provoke an immediate anxiety symptom.

Incidence is 5%. Typical onset is in later teens.

#### **Agoraphobia**

A cluster of phobias relating to two or more of: crowds, public places, public transport leaving home or travelling alone.

To be diagnosed with agoraphobia, the person will need to be exhibiting avoidance behaviours. These avoidances occur out of a fear of experiencing

a panic attack or anxiety-related symptoms in a situation from which it would be difficult to flee or no help would be available.

Incidence of 2.5% amongst adolescents. Onset in mid teens.

### **Prevalence**

The overall prevalence of anxiety disorders is 5-10%. The male to female ratio is equal for ages 8-11. During adolescence this changes and anxiety become twice as common amongst females.

What are the effects and signs?

Generalised anxiety disorders

Generalised Anxiety Disorder (GAD)

Psychological symptoms

GAD can cause a change in the child's behaviour and due to a change in the way they think and feel about things result in symptoms such as:

- restlessness
- a sense of dread
- feeling constantly "on edge"
- difficulty concentrating
- irritability

These symptoms may cause the child to withdraw from social contact (seeing family and friends) to avoid feelings of worry and dread.

The child may find going to school difficult and stressful and may take time off sick. These actions can make the child worry even more and increase their lack of self-esteem.

Physical symptoms of GAD

GAD can also have a number of physical symptoms, including:

- dizziness
- tiredness
- a noticeably strong, fast or irregular heartbeat (palpitations)
- muscle aches and tension
- trembling or shaking
- dry mouth

### **Panic Disorder**

Symptoms of panic disorder are physical and occur in unpredictable attacks.

These are:

- depersonalisation
- derealisation
- shortness of breath
- fast heart rate



- sweating
- dizziness

a feeling of impending doom or that the person is going to die.

## Situational Anxiety Disorders

### **Separation anxiety**

Some of the most common behaviours include:

- clinging to parents
- extreme and severe crying
- refusal to do things that require separation
- physical illness, such as headaches or vomiting
- violent, emotional temper tantrums
- refusal to go to school
- poor school performance

### **Social anxiety**

Symptoms of social anxiety disorder can include:

- Intense anxiety in social situations
- Avoidance of social situations
- Physical symptoms of anxiety, including confusion, pounding heart, sweating, shaking, blushing, muscle tension, upset stomach, and diarrhoea

### **Selective mutism**

The child will speak at home and familiar surroundings provided they know whom they are speaking to. As a rule they will be unable to speak to strangers or at school.

In addition they may also have:

- anxiety disorder (a social phobia)
- excessive shyness
- fear of social embarrassment
- social isolation and withdrawal

### **Specific phobias**

The symptoms of specific phobias are the same as for panic attacks and panic disorder.

### **Agoraphobia**

The physical symptoms of agoraphobia are the same as for panic disorder and panic attacks.

In addition, the child will exhibit avoidance behaviours, that is, avoiding situations that could lead to panic attacks such as crowded places, public transport and queues. They may also be unable to leave the house for long periods of time or display the need to be with someone that they trust when going anywhere or they may try to avoid being far away from home.

### **How is it assessed?**

Children with symptoms of an anxiety disorder will usually see their GP initially. The diagnosis of anxiety disorder is a clinical one and is based on symptoms alone. The GP will usually refer cases to the local CAMHS where the child will be assessed by a variety of professionals, which may include a psychiatrist, psychologist and a community psychiatric nurse (CPN) and others.

### **Generalised anxiety disorder (GAD)**

Diagnostic criteria are:

1. A period of at least six months with prominent tension, worry and feelings of apprehension, about everyday events and problems.
2. At least four symptoms out of a large list of items must be present, of which at least one from the following main symptoms must be present.
  - Palpitations
  - Sweating
  - Trembling or shaking
  - Dry mouth

### **Panic disorder**

The diagnostic criteria for panic disorder are:

A. Both 1 and 2 (a to c) below

1. Recurrent unexpected panic attacks

More than one of the three situations below has been present for more than a month following at least one panic attack -:

2. Persistent concern about having the attacks

a. Worry about the implications of the attack or its consequences (for example, having a heart attack)

b. A significant change in behaviour related to the attacks.

c. The panic attacks are not due to the direct physiological effects of a drug of abuse, a medication or general medical condition (over active thyroid).

The panic attacks are not better accounted for by another mental condition.

### **Social anxiety**

The diagnostic criteria for social anxiety are:

A. A marked and persistent fear of more than one social situation in which the person is exposed to unfamiliar people or to scrutiny by others. This occurs in peer settings and not just in interactions with adults.

B. Exposure to the feared social situation almost invariably provokes anxiety. In addition to panic attacks the anxiety may be expressed by crying.

C. Tantrums, freezing or shrinking from social situations.

D. The feared social situation is avoided or endured with intense anxiety.

E. The avoidance, anxious anticipation, or distress in the feared social situation interferes significantly with the person's normal routine. Duration lasts more than 6 months.

F. The fear or avoidance is not due to medication, medical or mental health conditions or diagnoses.

### **Selective mutism**

The diagnostic criteria for selective mutism are:

A. Consistent failure to speak in specific social situations (in which there is an expectation for speaking, for example, at school) despite speaking in other situations.

B. The disturbance interferes with educational or occupational achievement or with social communication.

C. The duration of the disturbance is at least 1 month (not limited to the first month of school).

D. The failure to speak is not due to a lack of knowledge of, or comfort with, the spoken language required in the social situation.

E. The disturbance is not better accounted for by a Communication Disorder (for example, stuttering) and does not occur exclusively during the course of a Pervasive Developmental Disorder, Schizophrenia or other Psychotic Disorder.

### **Specific phobias**

The diagnostic criteria for specific phobias are:

A. Fear or anxiety about a specific object or situation. The phobic object or situation almost always provokes immediate fear or anxiety.

B. The phobic object or situation is actively avoided or endured with intense fear or anxiety.

C. The fear or anxiety is out of proportion to the actual danger posed by the specific object or situation and to the sociocultural context.

D. The fear, anxiety, or avoidance is persistent, typically lasting for 6 months or more.

E. The fear, anxiety, or avoidance causes clinically significant distress or impairment in social, occupational, or other important areas of functioning.

F. The disturbance is not better explained by symptoms of another mental disorder.

### **Agoraphobia**

A person who has agoraphobia disorder experiences significant and persistent fear when in the presence of, or anticipating the presence of, at least two situations. These situations may include crowds, public places, public transportation, being outside of the home and in open spaces. To meet the diagnostic criteria, when in these situations, the person must engage in avoidance behaviours to avoid the fear and/or a related panic attack.

The symptoms for all ages must have a duration of at least 6 months.

The anxiety, panic attack or phobic avoidance associated with the specific situation is not better accounted for by another mental disorder.

### **School Performance/Attendance**

According to the office of national statistics, teachers consider that 40% of children with an anxiety disorder to be behind on their school work. 30 to 40% will have special educational needs but the numbers who have a statement of

educational needs is the same as that for children who do not have an anxiety disorder.

How is it treated and managed?

### **Cognitive Behavioural Therapy (CBT)**

CBT is the main therapy for all types of anxiety in children. Medication is usually used only in those children that do not respond to one or more courses of CBT.

CBT is a 'talking therapy' that can help the child manage their problems by changing the way they think and behave. It is most commonly used to treat anxiety and depression but can be useful for other mental and physical health problems.

CBT is very effective. The numbers needed to treat (NNT) in studies is between 3 and 4. This means that for every 3-4 children or adolescents treated, one child will return to a normal level of functioning. The benefits of CBT are usually maintained long term with 85% of all children being free of a diagnosis of an anxiety disorder at 3 years.

For treatment with CBT, children rate themselves as either much or very much improved in 60% of cases after 3 months.

### **Medication**

This is reserved for when there is no response to CBT/Behavioural therapy or the child is reluctant to undergo CBT.

### **Selective serotonin re-uptake inhibitors (SSRIs)**

SSRIs are safe and effective in treating all forms of anxiety in children and adolescents. The numbers needed to treat are 3-4. That is, for every 3-4 children or adolescents treated one will go back to normal. The others treated will experience a significant improvement in symptoms but may still have some anxiety. Current guidelines recommend the use of Fluoxetine first line then Sertraline if the first drug is not tolerated. 60% of children will rate themselves as much or very much improved after 12 weeks.

**Fluoxetine:** This is an antidepressant drug, which increases the level of serotonin in the brain.

**Dose:** Start dose is half adult dosage for adolescents (aged 12-18) and weight based for children (aged 5-11). Start dose is 10mg once daily. Increased to 20mg once daily after 7 to 14 days.

Side effects:

- insomnia
- headache
- fatigue
- indigestion
- dizziness

This is a well tolerated drug and most side effects will settle within the first 10-14 days of taking.

**Sertraline:** This is another drug which increases serotonin in the brain. This drug is not licensed for use in children so will only be prescribed for severe depression (usually with suicidal behaviour) after a fair trial of fluoxetine coupled with psychological therapy has failed to show any improvement.

**Dose:** 25mg for 14 to 28 days. Increased to 50mg after 2-4 weeks.

Side effects:

- insomnia
- dizziness
- tiredness
- headache
- nausea
- diarrhoea

Side effects usually settle over the first month of treatment for the majority of people. Well tolerated.

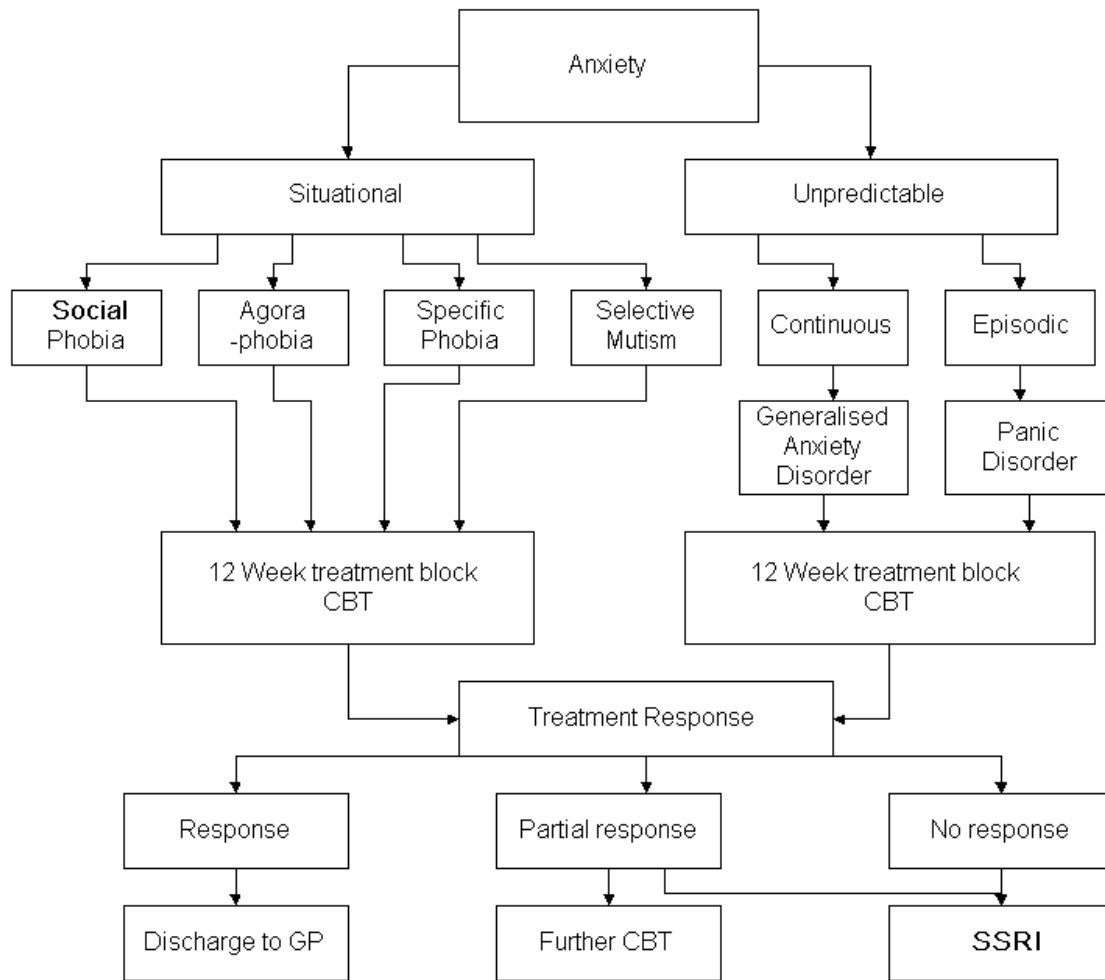
### **Combination of an SSRI and CBT**

The numbers needed to treat (NNT) are 1.7 for combination therapy. In combination treatment with an SSRI and CBT studies have shown that 81% of children rate themselves as much or very much improved after treatment.

What evidence is available?

Sources of evidence	Information provided	Limitation of evidence
Parent/Carer	First source of information and associated evidence.	Does not have specialist knowledge.
Multidisciplinary Team (Psychiatrist, Psychologist, Community psychiatric nurse )	Clinical features, treatment and information about disability and needs.	Information regarding symptoms and disabling effects may be based on what the parent / carer has told the relevant professional.
General Practitioner	Clinical features, treatment and some information about anxiety and needs.  May be the only source of information if no other professional involvement.	Does not have specialist knowledge.  Unlikely to have detailed or recent information about resulting disability or needs.
School (Teacher or SENCO if allocated)	Able to detail any additional support and needs required during the day.	Does not have specialist knowledge.

## Anxiety disorders - severity



How long will the needs last?

The following table is to be followed if an award is made. It is not a statement of intended awards for individual categories.

Age at date of claim	Length of award		
	Mild	Moderate	Severe
5-16 first award*	Award for 18 months	Award for 18 months	Award for 3 years

**\*Note:** For subsequent awards, if needs persist, award for 3 years.

You must take all information into account when considering the duration of care and mobility needs. You must base the duration of care and mobility needs on the particular circumstances of the child.

Severity of Anxiety disorder

**Mild**

Symptoms of anxiety are present for more than 50% of the time but, not every day. There is only slight impairment to the child's family life, social and school life.

**Moderate**

Symptoms present for more than 6 months for more than 50% of the time. There will be restrictions to family life because of the symptoms and the child will be missing social activities and school. The physical symptoms of anxiety may be leading to significant school absence.

**Severe**

Symptoms present nearly all the time for more than 6 months. This leads to a consistent pattern of impairment of the child's family life, social life and school attendance. The physical symptoms of anxiety may be leading to medical and Emergency Department consultations.

You must take all information into account when considering the duration of care and mobility needs. You must base the duration of care and mobility needs on the particular circumstances of the child.



## What you need to know about Asthma

### What is Asthma?

Asthma is a lung condition that affects breathing .....

- [Asthma](#)

### What are the effects and signs?

There is no gold standard definition of asthma and diagnosis is based on symptoms and response to treatment .....

- [Effects of Asthma](#)

### How is it assessed?

Most diagnoses of asthma will be based on history of symptoms .....

- [Assessment of Asthma](#)

### How is it treated and managed?

The following drugs are used in mild to moderate asthma .....

- [Treatment for Asthma](#)

### What evidence is available?

The best source of information on children with more severe asthma will be the asthma specialist nurse or the consultant paediatrician .....

- [Evidence](#)

### How long will the needs last?

The majority of children have mild asthma and will have no care needs.....

- [Prognosis and duration of the award](#)

## What is Asthma?

Asthma is a lung condition that affects breathing. Unlike other lung conditions the effect on breathing is intermittent. Typically, there will be episodes of asthma attacks with normal function in-between. During episodes of asthma the airways become irritated and inflamed. Airways become narrower and may produce extra mucus (the thick, slippery secretion of mucous membranes). This makes it more difficult for air to pass through the

airways increasing the effort of breathing. Narrowed inflamed airways give rise to the symptoms of asthma which are -:

- Coughing
- Wheezing
- Shortness of breath or difficult breathing
- A tight feeling in the chest

The commonest cause of asthma episodes in children is upper respiratory tract infections (URTIs) – these may occur 6-8 times each year in a normal healthy child with asthma. Asthma may also be triggered by allergens in children with allergies. Common allergies associated with asthma include house dust mite, cat and pollen allergy. Non-specific triggers such as dusts, fumes, chemicals and smoke may trigger asthma whether allergy is present or not. Exercise may trigger asthma in some children particularly outside in cold air. Once asthma is triggered symptoms may last for a few minutes to hours. In more severe asthma, symptoms may last for up to a week. Treatment of asthma is designed to eliminate symptoms for most of the time. During exacerbations of asthma treatment will often need to be increased to gain control. The majority of children with asthma lead normal healthy lives, controlling their asthma with effective medication.

### **Incidence/Prevalence**

1 in 8 children in the UK have symptoms of asthma. 1 in 10 children will visit their GP about asthma symptoms. The majority have mild symptoms; mainly wheeze during upper respiratory tract infections. Many will not require asthma treatment except when they have a cold. For those who have symptoms more frequently, regular treatment will be required – the majority of children on regular treatment for asthma will have well controlled asthma and will live a normal life. A small number of children with asthma will have more severe disease; 2-5% of children with asthma will fall into this

category. They are likely to need close supervision and regular treatment with multiple drugs to control their asthma.

What are the Key Facts for Asthma?



[What conditions does this guidance cover?](#)

[Pen Pictures](#)

Key Facts:

1. What level of the asthma treatment guidelines does the child's prescribed medication relate?
2. [Has the child been prescribed 5 or more courses of oral steroids in the past 12 months?](#)
3. [Is the child currently prescribed any of the drugs listed?](#)
4. [Has the child been admitted to the paediatric intensive care unit \(PICU\) with asthma in the last 12 months?](#)
5. [Is the child's under a hospital specialist \(consultant or nurse specialist\)?](#)

What conditions does this guidance cover?

This guidance covers the condition  
Asthma – **Disability Code T17.**

**DM Note** - In asthma, it is important to know whether the child is under 5 years old or 5 years old and over.

Key Facts

1. What level of the asthma treatment guidelines does the child's prescribed medication relate?

**DM Note** - "The British Guidelines on Asthma Management" (BGAM – produced by the British Thoracic Society) include steps/stages of the management of asthma, each step/stage increasing with severity of the illness, in line with additional medication or increased dosages. Patients may start at any of the levels. Regular review is recommended so that, with improvement, a step down may be taken.

The relevant step of the asthma treatment guideline will have to be identified from the medication prescribed. It is not information that the parent / carer or Health Care Professional will be able to provide.

**Under 5s and 5's and over** - if medication relates to later steps of either [Asthma treatment guideline table](#), this is an indication of a severe condition.

**Under 5s and 5s and over** - if medication relates to early steps of either Asthma treatment guideline table, a mild or moderate condition is likely.

The best sources of evidence to determine this information are a copy of the current prescription list, the GP or the hospital consultant.

2. Has the child been prescribed 5 or more courses of oral steroids in the past 12 months?

More than five short courses of oral steroids in the past 12 months is an indication of a poorly controlled or severe condition.

The best source of evidence for this information is the parent / carer (for a copy of the current prescription list), the GP or hospital consultant.

3. Is the child currently prescribed any of the drugs listed?

Link to the drugs used in [severe asthma](#) page.

The drugs in the link above are only prescribed in cases of severe asthma.

The best source of evidence for this information is a copy of the current prescription list.

4. Has the child been admitted to the paediatric intensive care unit (PICU) with asthma in the last 12 months?

A very small number of children require 24 hour supervision for much of the time; there are very few children that would fall into this category because of asthma. Such children are likely to have been admitted to Paediatric Intensive Care (PICU) for their asthma in the last year.

The best source of evidence for this information is the parent / carer, hospital letters may be available.

5. Is the child's medical management consultant led?

Children with a severe functional restriction will invariably be under the care of a hospital paediatric clinic. It should be noted that some children with a mild or moderate functional restriction may also be receiving hospital care, especially if the child is under 5.

The best sources of evidence for this information are the parent / carer, the GP or the hospital consultant.

The Key Facts proforma is a word based electronic form, which allows you to record and print the answers to the Key Facts – [Click here to complete the Key Facts Proforma.](#)

Pen Pictures

Children under 5 Mild to Moderate functional restriction	Children under 5 Severe functional restriction	Children aged 5 and over Mild to Moderate functional restriction	Children aged 5 and over Severe functional restriction
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<p>Children under 5 with mild to moderate asthma may or may not be on preventative treatment with low dose steroid inhalers and leukotriene inhibitor. They will need to take 'preventer' steroid inhaler <b>twice a day</b> probably using a spacer device and a mask or a mouthpiece. Use of these devices <b>does not indicate severe asthma or care needs</b>. The parent or carer will need to carry reliever inhaler for asthma symptoms. Parents will need to monitor asthma control and step up treatment when necessary. They are likely to spend <b>5-10 minutes per day, morning and evening</b>, giving asthma 'preventer' inhalers if used.</p>	<p>Severe asthma is rare in children under 5 (though under-5s may have viral-induced wheeze which can be severe enough to need intensive care). Affected children are likely to be on later steps of the SIGN guidelines treatment protocol. Care will relate to <b>complexity of medication given and the need to intervene quickly during asthma exacerbations</b>. Care will be more difficult if the child is not able to comply with treatment</p>	<p>Children with mild to moderate asthma are unlikely to have significant care needs. This includes 95-98% of all children with asthma. They are likely to be on early <b>steps</b> of the asthma treatment guideline. They may or may not be on preventative treatment with low dose steroid inhalers and leukotriene inhibitor. They will need to take 'preventer' steroid inhaler <b>twice a day</b> probably using a spacer device. Using a spacer device with a mask or a mouthpiece <b>does not indicate severe asthma or care needs</b>. The delivery systems used for asthma drugs are simple and straightforward to use. It is the total daily dose of steroids and the number of hospital attendances that give a much better indicator of asthma severity. Parent/carer or child will need to carry a reliever</p>	<p>Children under 12 with severe asthma are likely to have care needs in excess of a healthy child. This includes 2-5% of children with asthma. The child is likely to be on later <b>steps</b> of the asthma guideline treatment protocol. They are likely to have multiple regular drugs to take every day and likely to be on a high dose steroid inhaler (see treatment section to assess dose prescribed). They may be extremely unwell during exacerbations resulting in regular attendances at A+E, admissions to hospital and <b>&gt;5 short courses of oral steroid per year. More than 5 A+E attendances per year is severe</b>. They are likely to have been admitted to hospital at least once in the previous year with asthma. For these children use of regular 'preventer'</p>
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		<p>inhaler for asthma symptoms. The parent will need to monitor asthma control and step up treatment when necessary or take the child to A+E up to 3 times a year. They may spend up to <b>5-10 minutes per day morning and evening</b> giving asthma 'preventer' inhalers if used, <b>the child is likely to be able to take medication themselves</b> but may need prompting or supervision by a parent to make sure they do it every day.</p>	<p>treatment is vital to prevent exacerbations. The child will be under the care of a Consultant Paediatrician or Consultant Respiratory Paediatrician for their asthma. They are likely to be on the final <b>steps</b> of the guideline <b>OR</b> be on middle steps of the guideline but with asthma not being controlled as indicated by having also had <b>5 courses of oral steroids or 5 A+E attendances</b>.</p> <p>DM Note:</p> <p>In terms of 'Key Facts', parents / carers of children on middle steps of treatment with uncontrolled asthma must be able to answer 'yes' to Q5 <b>and</b> 'yes' to at least one of Q2, Q3 or Q4 to be in the group of children considered to may well need considerable time &amp; attention related to treatment or symptoms.</p>
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			<p>A very small number of children require 24 hour supervision for much of the time; there are very few children that would fall into this category because of asthma. Such children are <b>likely to have been admitted to Paediatric Intensive Care (PICU) in the previous year.</b> Such children are likely to have such unstable asthma that they are on a subcutaneous terbutaline pump 24 hours a day, oral steroids or other immunomodulating drugs such as azathioprine or omalizumab.</p>
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### **What are the effects and signs?**

There is no gold standard definition of asthma and diagnosis is based on symptoms and response to treatment. Typically, asthma symptoms will occur intermittently -:

- Wheezing especially at night and early in the morning

- Coughing at night
- Chest tightness
- Difficulty breathing

These symptoms are likely to get worse or only be present when the child has a cold. For the purposes of this guidance, mild to moderate asthma will be discussed together and severe asthma will be described and considered separately. Children with asthma under 5 years of age will be described separately from children aged 5 and over.

Asthma is a condition that can be controlled with medication, using the least amount of medication to control symptoms is the aim of treatment. This reduces the risk of side effects. Beneficial effects of treatment always outweigh side effects.

The management of asthma is a step wise process. Each step up in treatment is designed to gain better control of asthma symptoms and enable a normal life with normal exercise tolerance and undisturbed sleep. Clear guidelines on asthma have been produced by the Scottish Intercollegiate Guideline Network (SIGN). These are widely used across primary and secondary care in the UK. The steps are described in detail in this section, as treatment used will give a clear indication of what step of treatment a child is on, how severe their asthma is and whether there are likely to be any resulting care needs. The step or stage of asthma according to these guidelines may also be given in any accompanying medical evidence.

### **Other respiratory conditions claimed as 'Asthma'**

Asthma is a very common respiratory condition; any condition affecting the lungs is likely to cause difficulty breathing and coughing and at first may appear to be asthma. Children who fail to improve with asthma treatment or who appear to have more severe or worsening asthma are likely to have further investigations for an underlying lung condition. The underlying condition may be identified after a period of asthma treatment. Examples of such conditions include [cystic fibrosis](#), Goodpasture's syndrome, bronchiectasis

and many others. If another respiratory condition is identified, asthma guidance is not appropriate.

Other conditions which increase care needs related to asthma i.e. care needs present in mild to moderate asthma can be -:

- Any diagnosed behavioural condition which makes compliance with treatment on a daily basis difficult or prolonged.
- Severe allergy with history of anaphylactic shock (severe reaction) – particularly if allergen can be inhaled and is a common environmental contaminant
- When asthma is one of a number of conditions which in combination require extra care and attention on a daily basis

### **How is it assessed?**

Most diagnoses of asthma will be based on history of symptoms including -:

- Wheezing – musical sounds heard in the chest during asthma exacerbations
- Coughing – especially at night and in the morning
- Chest tightness
- Difficulty breathing or shortness of breath

Examination findings are likely to include evidence of wheezing on auscultation during exacerbations, wheezing may be absent between attacks.

### **Peak Flow Rate Monitoring (PEFR monitoring)**

In children over 5 Peak Flow Rate (PEFR) Monitoring is likely to be done. A Peak Flow is measured using a Peak Flow Meter. This is a simple device which the child blows into as hard as possible, the device measures maximum 'puff' in litres per minute (L/min). The value can be compared to the normal value expected given the height of the child or the most recent personal best PEFR. PEFRs may be given as a ratio in medical evidence; a ratio of 1 is normal and could imply that the asthma is well controlled or that there is no asthma. Often PEFR will be measured and charted for some days or weeks to assess whether there is an asthmatic pattern and whether this improves with asthma treatment. Typical asthma patterns include variation in peak flow measurements – peak flow in people without asthma is relatively constant, variation of up to 5% is normal, variation of more than 15% indicates asthma. A typical pattern of variability is 'diurnal variation' – this means the peak flow measurement varies through the day, being typically lower in morning and is a hallmark of asthma. Another hallmark of asthma is

'reversibility'. This means that a low peak flow can be significantly improved by using a beta-agonist inhaler. A person without asthma would only show minimal increase of peak flow after using one. The Peak Flow meter can be used at home to chart PEF in severe asthma to enable treatment adjustments. It is likely to be used for a few weeks at a time in mild to moderate asthma.

In the majority of children with mild to moderate asthma PEF monitoring and response to treatment will be the only investigations used.

In children who fail to respond to asthma treatment or who appear to have severe asthma requiring intensive treatment further investigations may be performed.

### **Pulmonary Function Tests/Spirometry**

This test involves blowing into a spirometry machine which measures the following -:

- Forced Vital Capacity (FVC) – volume of air that can be breathed out
- Forced Expiratory Volume in 1 second (FEV1) – volume of air that can be breathed out in 1 second

The expected peak flow and spirometry readings for a child will vary depending on age and height. If an interpretation of the readings is not given in medical evidence but the readings are provided, seek medical advice for interpretation. In asthma the readings will show an 'obstructive defect' – this means the test shows that breathing out is restricted compared to normal and is characteristic of asthma. In a case of severe asthma, these readings would be expected to be abnormal with an FEV1 of less than 80% of normal, in-between asthma exacerbations.

### **Exercise testing**

This involves the child exercising and then having peak flow monitoring or spirometry. Children with severe asthma are more likely to have a positive exercise test. Having a positive exercise test can mean that asthma is not well controlled on current treatment.

### **Allergy testing**

Children with severe asthma that is hard to control are likely to have allergy testing to assess whether asthma is due to avoidable inhalant allergens.

This is usually done by skin prick testing or blood tests. Very rarely, food allergy or food preservatives such as sulphites may worsen asthma control and avoiding foods to which the child is allergic on testing or that contain preservatives can improve asthma control.

### **Chest X-ray**

These are generally normal in asthma but are used to rule out other causes of respiratory illness.

### **Tests for rhinitis**

This is tested for using a Rhinitis Assessment Questionnaire and clinical examination. Treating allergic rhinitis (inflammation of the mucous membrane of the nose) in addition to asthma when it is present can significantly improve asthma control. CT is not often used in children.

All these tests will clarify whether asthma is the condition causing breathing difficulties. Where asthma is confirmed, further testing will guide therapy. For example, allergy testing may reveal inhalant allergies which can be avoided to improve asthma control and enable treatment of allergic rhinitis, which can improve asthma control.

### **How is it treated and managed?**

#### **Mild, moderate and severe Asthma treatments**

The following drugs are used in mild to moderate asthma and are effective in the vast majority of children. Children with severe asthma are likely to also take many of these drugs and still have poor control. The section on

drugs used in severe asthma lists the drugs that may be taken in addition to the ones listed below.

### **Short acting beta agonist inhalers (Reliever Inhalers)**

These drugs are used in mild, moderate and severe asthma.

Salbutamol (Ventolin) dose - 100-200mcg (2-10 puffs) up to every 4 hours. These can be given via

- Metered dose inhaler (MDI)
- Breath actuated inhalers e.g. 'easibreathe'
- MDI Inhaler and spacer device
- Nebuliser
- dry powder inhaler (DPI) – ventodisks using 'accuhaler'

Other trade names for salbutamol include Maxivent, Salamol, Airolin, and Airomir.

Terbutaline (bricanyl) can be given via

- Metered dose inhaler (MDI)
- Inhaler and spacer device
- Nebuliser – trade name for nebuliser doses is Respules
- dry powder inhaler (DPI)– Turbohaler

### **Steroid inhalers (Preventer inhalers)**

There are several different types of steroid inhaler and doses are not equivalent between different types. A table of doses categorised as high and low dose steroid inhaler use by age is provided. Steroid inhalers are usually taken twice a day every day to control inflammation in the lungs and reduce the risk of uncontrolled asthma. The use of steroid inhalers in mild to moderate asthma enables a normal active life with minimal asthma related interruption to activities including sleep and exercise as long as the inhaler is taken regularly. Steroid inhalers can be given using a standard metered dose inhaler or other methods including dry powder formulation, breath actuated inhaler or nebuliser. The methods used will reflect the age of the child and ability to use an inhaler rather than the severity of the asthma. The prescribed dose of steroid inhaler used will give a good indication of

severity of asthma as high dose steroid inhalers are avoided as much as possible in children because of potential effects on growth of the child.

Steroid inhalers dosing information: - To use the tables (see link at bottom of page), work out which steroid drug the inhaler contains and then work out the dose of steroid inhaler used -:

E.g. Clenil 100 (100mcg) is prescribed

Dosing instruction are 2 puffs twice a day = 4 puffs in total

Each dose = 2 puffs times 100mcg = 200mcg twice daily

Total daily dose is 400mcg daily. N

Note the total daily dose is given in the SIGN treatment guidelines pages.

### **Sodium Cromoglicate (Preventer inhalers)**

These drugs should always be considered equivalent to low dose steroid inhalers and are used in mild to moderate asthma. They are taken twice daily like steroid inhalers and dose may be increased during asthma exacerbations. Trade names include Intal and Tilade. These are rarely used.

**Initial 'add on' therapy These treatments are used in addition to regular 'preventer' steroid inhalers and use of reliever inhalers to improve asthma control and reduce exacerbations of asthma.**

### **Leukotriene inhibitors**

These are tablet treatments taken to improve asthma control. The usual dose is 1 tablet per day. Drug names include Montelukast (Singulair) and Zafirlukast (Accolate) - side effects are relatively common and include sleep disturbance and behavioural issues. This tablet treatment is used without 'preventer' inhalers to treat rhinitis (inflammation of the mucous membranes of the nose), which may or may not accompany asthma. This drug is starting (in 2009) to be used earlier instead of low dose steroid inhalers to treat asthma, especially in younger children. Use of these drugs does not necessarily indicate moderate asthma and these drugs are more likely to be

trialed in young children as some parents can find it easier to give this medication than use inhalers.

### **Long acting beta agonist inhalers (LABA)**

These drugs are inhalers containing long acting beta agonist drugs similar to the beta agonist drugs used in reliever inhalers like Salbutamol (Ventolin). These drugs have a longer lasting airway opening effect and are taken twice daily with steroid inhalers to improve asthma control. Names of drugs include Salmeterol (Serevent) and Formoterol. Use of these drugs indicates at least moderate asthma. It is important to note that some inhalers have a combination of drugs – for example seretide is a combination of an inhaled steroid and a LABA – as this aids compliance of treatment. This is an important note as number of drugs, rather than number of inhalers, is one of the markers of severity of disease.

### **Theophylline (Nuelin SA, Uniphyllin Continus)**

This is a tablet treatment for asthma. Doses are given twice a day and blood levels of the drug need to be monitored. Use of these drugs indicates at least moderate asthma. These drugs may also be used intravenously in the treatment of acute asthma.

### **Short courses of oral steroids for acute asthma exacerbations**

Short courses of high dose oral steroid (up to 40mg prednisolone per day) for 3-5 days are commonly given to children with acute asthma and are not an indication that asthma is severe. However, 5 or more short courses in one year is an indicator of poorly controlled asthma.

**Note:** The Alphabetical List of Medication and the Steroid Inhaler Doses by age tables below should be used together with the information provided in



the appropriate Sign treatment guidelines table for the child's age. See the links below:

[Alphabetical list of inhaler medication by Non-propriety name \(includes links to Steroid Inhaler doses by age\)](#)

[Alphabetical list of inhaler medication by Brand name \(includes links to Steroid Inhaler doses by age\)](#)

[SIGN treatment guidelines for under 5s](#)

[SIGN treatment guidelines for 5s and over](#)

[Drug treatments only for severe asthma](#)

[Inhalers & other drug delivery devices](#)

[Non-drug treatments](#)

**What evidence is available?**

<b>Sources of Evidence</b>	<b>Information Provided</b>	<b>Limitation of Evidence</b>
Parent / Carer	First source of information about details of GP and Consultant appointments and prescription of medication.	Does not have specialist knowledge.

<p>GPFR</p>	<p>Children with asthma should be identified by the primary care trust. The <a href="#">Quality outcome Framework (QOF)</a> for Primary Care, details minimum data that should be collected about a person with asthma. For those children with asthma managed in primary care the following information is likely to be collected annually -:</p> <ul style="list-style-type: none"> <li>• Information on whether asthma is well controlled may include scoring from 'Asthma Control Questionnaire' or 'Asthma Control Test'.</li> <li>• Lung function – either PEFR reading or spirometry.</li> <li>• Information on exacerbations of asthma including number of courses of oral steroids and time off school</li> <li>• Inhaler technique</li> <li>• Compliance with treatment (checked by reviewing whether repeat prescriptions have been requested)</li> <li>• Reliance on bronchodilators (checking repeat prescription for beta agonist inhaler)</li> <li>• Possession of and use of self management plan/personal action plan for asthma.</li> </ul> <p>The majority of children managed in primary care will have this information collected about their asthma. Those being managed in secondary care will be identified as having asthma and receive their medication via the GP. Confirmation of diagnosis and prescription information will be available. Details of asthma control should be available from the treating hospital. The children's asthma specialist nurse or paediatrician will be the best source of information on asthma control in these cases.</p>	<p>More detailed information will be available for children with mild asthma who are managed in primary care. The GP may only be able to confirm medication prescribed for children with more severe asthma managed in secondary care, this may be sufficient evidence for children over 5.</p>
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HFR	The best source of information on children with more severe asthma will be the asthma specialist nurse or the consultant paediatrician. They will be able to provide written confirmation of specific advice on supervision given to parents by the hospital and provide up to date details of current medication and asthma control.	N/A
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### How long will the needs last?

#### Asthma Duration Guidance

Age at date of claim	Award to the following age (Under consultant /specialist – on medication regime)	Award to the following age (On terbutaline Pump treatment)
1 - 3	5	5
4	5 (or 1 year award whichever is the greater)	5 (or 1 year award whichever is the greater)
5 - 10	12	14
11	12 ( or 1 year award whichever is the greater)	14
12	N / A	14
13	N / A	14 (or 1 year award whichever is the greater)

#### Under 5

Care needs may be present under age 5 because of the time taken to administer asthma treatment in this age group. This will depend less on the

severity of the asthma than the behaviour of the child and the difficulty of administering daily treatment to control the disease.

By 5 years of age, child behaviour and ability to co-operate with asthma treatment is much improved. Care needs may be substantially reduced.

At age 5 a renewal with fresh medical evidence will be needed.

## **5 and over**

The majority of children have mild asthma and will have no care needs. Children aged over 5 are likely to have care needs when asthma is severe and difficult to control. These children constitute 2-5% of children with asthma in the UK. Such children will always be managed in secondary care by either a Consultant Paediatrician or a Consultant Respiratory Paediatrician in a regional centre.

They are likely to have multiple courses of oral steroids each year to control asthma exacerbations and be on multiple regular medications in-between. Ensuring multiple drugs are available and taken several times a day, recognising when asthma is uncontrolled and seeking medical help is the care required. The medication will need to be stepped up and down to control exacerbations. Drugs used are likely to include some of the following -:

- High dose inhaled steroids,
- LABA
- leukotriene inhibitors (tablets)
- theophylline
- methotrexate
- omalizumab
- subcutaneous terbutaline infusion
- daily oral steroids
- 

Monitoring will be regular and frequent, with the need to interpret Peak Flow Monitoring charts to modify medication regimes to avoid severe exacerbations of asthma and hospital admission. Exacerbations are likely with every cold and children with severe asthma have these 6-8 times a year. Such children will be able to manage their own complex monitoring/medication needs from the 12<sup>th</sup> birthday. Those on terbutaline subcutaneous infusion

pumps or other drug infusions for asthma control would be expected to manage this from the 14<sup>th</sup> birthday.

Children with needs identified aged 5 and over are likely to require care until age 12, by which time they will have been taught how to manage their asthma themselves. The exception to this is children using subcutaneous pump treatment. They are likely to require extra care until age 14, at which age they can be expected to manage their treatment themselves.

Children with other conditions in addition to asthma such as learning difficulty or behavioural disorder may not be able to manage their treatment at these ages without help and supervision, duration guidance here does not apply to them, assess each case individually.

Poorly controlled or difficult to treat (previously known as **Brittle or very unstable asthma**)

A very small number of children (less than 1000 in the UK) will have been advised not to go anywhere without adult supervision or supervision from a child older than 14 because of their propensity to have life threatening asthma attacks within a few minutes of inhaling asthma triggers or allergens.

These children may need help until age 14. From that age onwards they can be expected to recognise an asthma attack and act accordingly to get help. If the child cannot do this, then they will continue to need help. Confirmation of such attacks should be obtained from the Clinical Nurse Specialist

or treating Paediatrician and duration of award details discussed with Medical Services.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Alphabetical List of Inhaler medication by non-propriety name**

Non-proprietary name	Brand name	Medication type	Steroid dose information
Beclometasone	Asmabec clickhaler 50mcg per MDI	Corticosteroid - PRE- VENTER	Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a>
	Asmabec clickhaler 100mcg per MDI		
	Asmabec clickhaler 250mcg per MDI		
	Beclometasone Easyhaler check dose per metered dose inhalation (MDI)		
	Beclazone Easi-breathe 50mcg per metered dose inhalation (MDI)		
	Beclazone Easi-breathe 100mcg per MDI		
	Beclazone Easi-breathe 250 mcg per MDI		
	Becodisks 100mcg per metered dose inhalation (MDI)		
	Becodisks 200mcg per MDI		
	Becodisks 400mcg per MDI		
	Clenil modulite 50mcg per metered dose inhalation (MDI)		
	Clenil modulite 100 mcg per MDI		
	Clenil modulite 200mcg per MDI		



	Pulvinal Beclometasone Dipropionate check dose per MDI		
	Qvar 50 contains 50mcg per metered dose inhalation (MDI)	Corticosteroid - PRE-VENTER	Standard or Low dose Child 12-18 years 50-200 micrograms twice daily
	Qvar 100 contains 100mcg beclometasone per MDI	Corticosteroid - PRE-VENTER	High dose Child 12-18 years 200-500 micrograms twice daily
<b>Non-proprietary name</b>	<b>Brand name</b>	<b>Medication type</b>	<b>Steroid dose information</b>
Budesonide	Easyhaler budesonide 100 contains 100mcg per metered dose inhalation (MDI)	Corticosteroid - PRE-VENTER	<p>Note: - may be given once daily. Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a></p>
	Easyhaler budesonide 200 contains 200mcg per MDI		
	Easyhaler budesonide 400 contains 400mcg per MDI		
	Novolizer contains 200mcg per metered dose inhalation (MDI)		
	Pulmicort contains 100mcg per MDI		

	Pulmicort turbohaler contains 100mcg per MDI	Corticosteroid - PRE-VENTER	dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a>
<b>Non-proprietary name</b>	<b>Brand name</b>	<b>Medication type</b>	<b>Steroid dose information</b>
Ciclesonide	Alvesco 80 mcg per MDI	Corticosteroid - PRE-VENTER	Taken once daily. Children aged 12-18 years. Low or standard dose.
Fluticasone	Flixotide Accuhaler 50mcg per metered dose inhalation (MDI)	Corticosteroid - PRE-VENTER	Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose
	Flixotide Accuhaler 100mcg per metered dose inhalation (MDI)		
	Flixotide Accuhaler 250mcg per metered dose inhalation (MDI)		
	Flixotide Accuhaler 500mcg per metered dose inhalation (MDI)		
	Flixotide Diskhaler 100mcg per MDI		
	Flixotide Diskhaler 250mcg per MDI		
	Flixotide Diskhaler 500mcg per MDI		
	Flixotide Evohaler 50mcg per MDI		

	Flixotide Evohaler 125mcg per MDI		by age – see <a href="#">Steroid Inhaler doses by age</a>
	Flixotide Evohaler 250mcg per MDI		
Formoterol	Atimos modulite	Long-acting Beta 2 Ago- nist - RE- LIEVER	N/A
	Foradil		
	Formoterol Easyhaler		
	Oxis turbohaler		
Formoterol and Budesonide (steroid)	Symbicort 100/6 Contains 100mcg budesonide per dose	Combination Long-Acting Beta 2 Ago- nist and ster- oid inhaler - RELIEVER and PRE- VENTER	Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a>
	Symbicort 200/6 Con- tains 200mcg budesonide per dose		
	Symbicort 400/12 Contains 400mcg budesonide per dose		
Ipratropium bro- mide	Atrovent	Short acting Anticholiner- gic - RE- LIEVER	N/A
Mometasone	Asmanex twisthaler	Corticosteroid - PRE- VENTER	Note: - may be taken once daily. Stand- ard or low dose Child 12- 18 years 200 micrograms twice daily High dose Child 12-18 years up to 400 mi- crograms twice daily

Nedocromil sodium	Tilade CFC free inhaler	Sodium Cromoglicate - PRE-VENTER	Equivalent to low dose steroid inhaler
Salbutamol	Airolin	Short acting Beta 2 Agonist - RELIEVER	N/A
	Airomir		
	Asmasal Clickhaler		
	Easyhaler Salbutamol		
	Maxivent		
	Pulvinal Salbutamol		
	Salamol Easi-breathe		
	Salbulin Novolizer		
	Salbutamol		
	Ventmax SR		
	Ventolin Evohaler		
Salmeterol	Serevent Evohaler	Long-acting Beta 2 Agonist - RELIEVER	N/A
	Serevent Accuhaler		
	Serevent Diskhaler		
	Seretide 50 Evohaler Contains 50mcg Fluticasone per dose	Combination Long-Acting Beta 2 Agonist and steroid inhaler - RELIEVER	Check number of puffs per dose and number of
	Seretide 100 Accuhaler Contains 100mcg Fluticasone per dose		
	Seretide 125 Evohaler Contains 125mcg Fluticasone per dose		

Salmeterol and Fluticasone (steroid)	Seretide 250 Accuhaler Contains 250mcg Fluticasone per dose	and PRE-VENTER	times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a>
	Seretide 500 Accuhaler Contains 500mcg Fluticasone per dose		
Sodium cromoglicate	Intal CFC free inhaler	Sodium Cromoglicate - PRE-VENTER	Equivalent to low dose steroid inhaler
Terbutaline	Bricanyl turbohaler	Short acting Beta 2 Agonist - RELIEVER	N/A

[Asthma – How is it treated and managed](#)

[SIGN treatment guidelines for under 5s](#)

[SIGN treatment guidelines for 5s and over](#)

[Drug treatments only for severe asthma](#)

[Inhalers & other drug delivery devices](#)

[Non-drug treatments](#)

**Alphabetical List of Inhaler medication by Brand Name**

Brand name	Medication type	Steroid dose information	Non-proprietary name
Airolin	Short acting Beta 2 Agonist - RELIEVER	N/A	Salbutamol
Airomir			
Alvesco 80 mcg per MDI	Corticosteroid - PREVENTER	Taken once daily. Children aged 12-18 years. Low or standard dose.	Ciclesonide
Asmabec click-haler 100mcg per MDI		Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a>	Beclometa-sone
Asmabec click-haler 250mcg per MDI		Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a>	Beclometa-sone
Asmabec click-haler 50mcg per MDI		Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a>	Beclometa-sone

Asmanex twisthaler		Note: - may be taken once daily. Standard or low dose Child 12-18 years 200 micrograms twice daily High dose Child 12-18 years up to 400 micrograms twice daily	Mometasone
Asmasal Clickhaler	Short acting Beta 2 Agonist - RELIEVER	N/A	Salbutamol
Atimos modulite	Long-acting Beta 2 Agonist - RELIEVER	N/A	Formoterol
Atrovent	Short acting Anticholinergic - RELIEVER	N/A	Ipratropium bromide
Beclametasone Easyhaler check dose per metered dose inhalation (MDI)	Corticosteroid - PREVENTER	Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a>	Beclometasone
Beclazone Easi-breathe 100mcg per MDI			Beclometasone
Beclazone Easi-breathe 250 mcg per MDI			Beclometasone
Beclazone Easi-breathe 50mcg per metered dose inhalation (MDI)			Beclometasone
Becodisks 100mcg per metered dose inhalation (MDI)			Beclometasone

Becodisks 200mcg per MDI			Beclometa- sone
Becodisks 400mcg per MDI			Beclometa- sone
Bricanyl turbo- haler	Short acting Beta 2 Ag- onist - RELIEVER	N/A	Terbutaline
Clenil modulite 100 mcg per MDI	Corticosteroid - PRE- VENTER	Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid In- haler doses by age</a>	Beclometa- sone
Clenil modulite 200mcg per MDI			
Clenil modulite 50mcg per me- tered dose in- halation (MDI)			
Easyhaler budesonide 100 contains 100mcg per metered dose inhalation (MDI)	Corticosteroid - PRE- VENTER	Note: - may be given once daily. Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid In- haler doses by age</a>	Budesonide
Easyhaler budesonide 200 contains 200mcg per MDI			
Easyhaler budesonide 400 contains 400mcg per MDI			
Easyhaler Sal- butamol	Short acting Beta 2 Ag- onist - RELIEVER	N/A	Salbutamol



Flixotide Ac-cuhaler 100mcg per metered dose inhalation (MDI)	Corticosteroid - PRE-VENTER	Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a>	Fluticasone
Flixotide Ac-cuhaler 250mcg per metered dose inhalation (MDI)			
Flixotide Ac-cuhaler 500mcg per metered dose inhalation (MDI)			
Flixotide Ac-cuhaler 50mcg per metered dose inhalation (MDI)			
Flixotide Diskhaler 100mcg per MDI			
Flixotide Diskhaler 250mcg per MDI			
Flixotide Diskhaler 500mcg per MDI			
Flixotide Evo-haler 125mcg per MDI			
Flixotide Evo-haler 250mcg per MDI			

Flixotide Evo- haler 50mcg per MDI			
Foradil	Long-acting Beta 2 Ago- nist - RELIEVER	N/A	Formoterol
Formoterol Easyhaler			
Intal CFC free inhaler	Sodium Cromoglicate - PREVENTER	Equivalent to low dose steroid in- haler	Sodium cromoglicate
Maxivent	Short acting Beta 2 Ag- onist - RELIEVER	N/A	Salbutamol
Novolizer con- tains 200mcg per metered dose inhalation (MDI)	Corticosteroid - PRE- VENTER	Note: - may be given once daily. Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid In- haler doses by age</a>	Budesonide
Oxis turbohaler	Long-acting Beta 2 Ago- nist - RELIEVER	N/A	Formoterol
Pulmicort con- tains 100mcg per MDI		Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid In- haler doses by age</a>	Budesonide

Pulmicort turbohaler contains 100mcg per MDI		Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a>	Budesonide
Pulvinal Beclometasone Dipropionate check dose per MDI	Corticosteroid - PREVENTER	Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a>	Beclometasone
Pulvinal Salbutamol	Short acting Beta 2 Agonist - RELIEVER	N/A	Salbutamol
Qvar 100 contains 100mcg beclometasone per MDI	Corticosteroid - PREVENTER	High dose Child 12-18 years 200-500 micrograms twice daily	Beclometasone
Qvar 50 contains 50mcg per metered dose inhalation (MDI)		Standard or Low dose Child 12-18 years 50-200 micrograms twice daily	
Salamol Easi-breathe	Short acting Beta 2 Agonist - RELIEVER	N/A	Salbutamol
Salbulin Novolizer			
Salbutamol			

Seretide 100 Accuhaler Con- tains 100mcg Fluticasone per dose	Combination Long-Act- ing Beta 2 Agonist and steroid inhaler -RE- LIEVER and PRE- VENTER	Check number of puffs per dose and number of times daily against age. To work out low, standard or high dose by age – see <a href="#">Steroid In- haler doses by age</a>	Salmeterol and Fluticasone (steroid)
Seretide 125 Evohaler Con- tains 125mcg Fluticasone per dose			
Seretide 250 Accuhaler Con- tains 250mcg Fluticasone per dose			
Seretide 50 Evohaler Con- tains 50mcg Fluticasone per dose			
Seretide 500 Accuhaler Con- tains 500mcg Fluticasone per dose			
Serevent Ac- cuhaler	Long-acting Beta 2 Ago- nist - RELIEVER	N/A	Salmeterol
Serevent Diskhaler			
Serevent Evo- haler			
Symbicort 100/6 Contains 100mcg budesonide per dose	Combination Long-Act- ing Beta 2 Agonist and steroid inhaler - RE- LIEVER and PRE- VENTER	Check number of puffs per dose and number of times daily against age. To	Formoterol and Budesonide (steroid)

Symbicort 200/6 Contains 200mcg budesonide per dose	work out low, standard or high dose by age – see <a href="#">Steroid Inhaler doses by age</a>		
Symbicort 400/12 Contains 400mcg budesonide per dose			
Tilade CFC free inhaler	Sodium Cromoglicate - PREVENTER	Equivalent to low dose steroid inhaler	Nedocromil sodium
Ventmax SR	Short acting Beta 2 Agonist - RELIEVER	N/A	Salbutamol
Ventolin Evo-haler			

### **SIGN treatment guidelines for under 5s**

Children are unlikely to be diagnosed with asthma under the age of one. This is because many wheezy babies will grow out of their condition and not go on to develop chronic asthma. Children aged over 1 with persistent cough or wheeze may be diagnosed with and treated for asthma. The

following are steps in the treatment guidelines for children under 5 with asthma.

Note: The table below should be used together with the age and dosage information in the Treatment page. See the link at the bottom of the page.

<b>SIGN Asthma treatment guidelines for children aged under 5</b>	
<b>Mild to moderate asthma</b>	
Initial	Mild intermittent asthma
	Inhaled short-acting Beta agonist as required – ‘reliever’
If symptoms are uncontrolled	Regular ‘preventer’ therapy
	Inhaled short-acting Beta agonist inhaler as required and one of the following: <ul style="list-style-type: none"> <li>• Low or standard dose steroid inhaler twice daily</li> </ul> OR <ul style="list-style-type: none"> <li>• Leukotriene receptor antagonist</li> <li>•</li> </ul>
If symptoms are uncontrolled	Initial ‘add on’ therapy - medication is given in addition to regular preventer and reliever inhaler from previous step
	Inhaled short-acting Beta agonist as required <ul style="list-style-type: none"> <li>• Low or standard dose steroid inhaler twice daily</li> </ul> AND <ul style="list-style-type: none"> <li>• Leukotriene receptor antagonist</li> </ul>
<b>Severe asthma</b>	
If symptoms are uncontrolled	Persistent poor control
	Increase steroid inhaler dose (up to 400 mcg) beclomethasone

## **Mild to moderate Asthma**

Symptoms of mild to moderate asthma are likely to include -:

- Coughing especially at night
- Wheezy sounds from the chest
- Shortness of breath and rapid breathing.

The symptoms may be reported by either the child or the parent, depending on the age of the child. Intermittent use of bronchodilator beta agonist inhalers may control symptoms sufficiently, if these have to be used more than twice a week to control symptoms or if there are exercise induced asthmatic symptoms or trips to Accident & Emergency with asthma then a low dose steroid inhaler may be used as 'preventer' treatment. The inhalers are likely to be given using either a 'spacer device' or more rarely a nebuliser. These are used to enable a child to take the drug without having to learn an inhaler technique like an adult or an older child. They are not an indicator of severe asthma or care needs. Asthma inhalers require co-ordination of both breathing in and pressing the inhaler to release the inhaler spray. This is difficult for small children; the use of spacers or nebulisers does not indicate severe asthma. Upper respiratory tract infections (URTI) such as coughs and colds are common in young children – they may have 6-8 URIs each year, children with asthma are likely to cough more and wheeze during these infections unlike children without asthma. Children with asthma that is not controlled with the above measures are likely to have further tests and be referred to a paediatrician.

## **Severe Asthma**

Severe asthma in young children is rare but is likely to result in frequent attendance at the GP and more than 3 attendances per year to Accident & Emergency with difficulty breathing due to acute asthma. Children with severe asthma are likely to have been admitted to hospital at least once in the previous year and to be under the care of a consultant paediatrician for their asthma. Severe asthma in young children is rare. These children are likely to still have poor control of their asthma despite all add on therapies. These children are likely to be wheezy most of the time and to wake at night with frequent coughing. Frequent stepping up of treatment to control acute asthma especially with inter-current upper respiratory tract infection will be required. In younger children with severe asthma feeding may be difficult because of breathlessness and in older children exercise tolerance is likely to be limited by breathlessness. Those on high dose steroid inhalers are likely to have the most severe asthma. Missing doses of 'preventer' treatment in these children is likely to result in episodes of severe asthma and



ensuring that medication is available and taken as prescribed will be particularly important in this group.

see: [How is it treated & managed](#)

### **SIGN treatment guidelines for 5s and over**

In most children who have childhood asthma, asthma develops before the age of five years. However, many children under five who are wheezy will grow out of it. In addition, children aged 5 and over can understand that they have asthma and comply with treatment to prevent symptoms. Treatments for the under 5s may be difficult because of the age-related behaviour of the child; the over 5s will be able to comply with treatment. Wheeze

in many children under five will have resolved and no excess care needs will be present.

Note: The table below should be used together with the age and dosage information in the Treatment page. See the link at the bottom of the page.

<b>SIGN Asthma treatment guidelines for children aged 5 and over</b>	
<b>Mild to moderate asthma</b>	
initial	Mild intermittent asthma
	Inhaled short-acting Beta agonist as required - 'reliever' therapy
If symptoms are uncontrolled	Regular 'preventer' therapy & 'reliever' therapy
	Short-acting Beta agonist inhaler as required and Low dose steroid inhaler twice daily
If symptoms are uncontrolled	Initial 'add on' therapy - medication is given in addition to regular preventer and reliever inhaler from previous step
	Inhaled short-acting Beta agonist as required and Low dose steroid inhaler twice daily AND at least one of the following: <ul style="list-style-type: none"> <li>• Long-acting beta agonist inhaler</li> <li>• Increase steroid inhaler from low dose to standard dose</li> <li>• Leukotriene receptor antagonist</li> <li>• SR theophylline</li> </ul>
<b>Severe asthma</b>	
If symptoms are uncontrolled	Persistent poor control

	<ul style="list-style-type: none"> <li>• Increase steroid inhaler dose to high dose (800 mcg beclomethasone/day under 12, up to 200 mcg beclomethasone/day over 12)</li> </ul> <p>Plus continue other treatments from previous step:</p> <ul style="list-style-type: none"> <li>• Inhaled short-acting Beta agonist as required</li> <li>• Long-acting beta agonist inhaler</li> <li>• Leukotriene receptor antagonist</li> <li>• SR theophylline</li> </ul>
If symptoms are uncontrolled	<p>Continuous or frequent use of oral steroids in addition to treatments listed in previous step.</p> <ul style="list-style-type: none"> <li>• Other systemic drug treatments for severe asthma as listed in treatment section</li> </ul>

### **Mild to moderate asthma**

Children will be on early steps of the SIGN treatment guidelines. They may use bronchodilators (drugs that open the airways in spasm) for symptoms several times a week. They may have symptoms such as coughing at night and attend Accident and Emergency up to 3 times a year with asthma symptoms. They may have lost up to 10% of days off school with asthma. They may need to step up treatment regularly because of upper respiratory tract infection to control asthma. They may take up to 3 courses of oral steroids per year for asthma exacerbations. They will not be on long term high dose inhaled steroids. These children are likely to be well when not experiencing asthma exacerbations, able to exercise normally and sleep through

the night. They may be under the care of the GP in primary care or under a hospital paediatrician.

### **Severe Asthma**

Indicators of severe functional restriction

Indicators of severe functional restriction can only be applied to children aged 5 and over and are -:

- On later steps of the treatment guideline.
- >5 short courses oral steroids in one year.
- On any of the drugs listed under drug treatment only used in severe asthma.
- Required IV treatment as an emergency for an asthma attack
- Admission to paediatric intensive care (PICU) with asthma in the last year.
- Poorly controlled asthma and previous episodes of anaphylaxis.

These children will be under the care of a Consultant Paediatrician. They will have been identified in the medical evidence as having 'severe' or 'unstable' asthma. They are likely to be on later steps of the SIGN guidelines asthma treatment. They are likely to have symptoms of asthma most of the time and will often be wheezy during the day and cough at night. They are likely to have had significant time off school (more than 10% of days lost). They are likely to have had more than 3 admissions to hospital in the previous year or more than 5 Accident and Emergency attendances with asthma and more than 5 courses of oral steroids. This group of children will have had further investigations for their asthma and may do daily PEFr measurements and recording to guide asthma treatment.

Children with hard to control asthma as described above may be controlled by high dose inhaled steroids (see steroid inhaler dose information by age in treatment section) or other treatments such as long-term low dose oral steroids. Either of these treatments requires daily input and close supervision from parents to ensure medication is taken correctly in addition to asthma monitoring even though asthma itself may be better controlled with high dose treatment. Children on a terbutaline subcutaneous pump are likely to have very severe asthma. Children with hard to control asthma on multiple medications are likely to continue to lose time from school during exacerbations and require extra care on school trips to ensure medication is

taken as prescribed. Parents may be advised to check on children during the night when they have upper respiratory tract infections.

A very small number of children require 24-hour supervision for much of the time; there are very few children that would fall into this category because of asthma. Such children are likely to have been admitted to Paediatric Intensive Care (PICU) in the previous year. Such children are likely to have such unstable asthma that they are on a subcutaneous terbutaline pump 24 hours a day, oral steroids or other immunomodulating drugs such as azathioprine or ciclosporin.

### **Poorly controlled or difficult to treat (previously known as Brittle or very unstable asthma)**

A very small number of children will require supervision out of doors because of rapid deterioration of asthma. They are likely to be on later steps of the asthma guidelines and take high doses of medication for asthma control. These children can develop life threatening attacks of asthma within a few minutes of inhaling a trigger or on exercise. Parents will have been advised by the Consultant Paediatrician that the child should not go off alone on a bike or go swimming without supervision from another child of at least 14 or an adult. Written confirmation of such advice will always be available from the Clinical Nurse Specialist or treating Paediatrician.

see: [How is it treated & managed](#)

### **Drug treatments only for severe asthma**

Children with severe asthma are likely to take multiple drugs (mentioned elsewhere in the Asthma treatment guidance), in addition to one or several of the drugs in this section. Children taking drugs in this section will always be classified in the severe asthma category. It is usually vital that the drugs in this section are taken regularly, as prescribed.

These drugs are given in tablet form and will be taken regularly once or twice daily. Side effects such as kidney failure do occur and are serious. Such treatment is likely to be given at a specialist centre with these drugs being carefully monitored by a Consultant Paediatrician.

#### Monoclonal Antibody treatment (Omalizumab and mepolizumab)

These treatments are given every few weeks as an injection in a hospital setting. Omalizumab combines with Immunoglobulin E (IgE), the antibody which is responsible for recognising and binding with allergens and causing allergic symptoms from asthma to anaphylaxis (severe allergic reaction). Mepolizumab binds to Interleukin-5 to decrease the allergic response. Regular treatment with Omalizumab/mepolizumab can improve asthma control

in children over 12 who have uncontrolled asthma despite maximal treatment. The use of these treatments is growing.

### **Regular oral steroids**

The main drug used is prednisolone. In severe asthma, oral steroids are given every day e.g. prednisolone 5mg once a day. The lowest dose that controls asthma is used. Steroids are used like this only as a last resort because side effects can be serious. The most common side effect is weight gain. Other effects include diabetes, osteoporosis and growth retardation. Stopping regular steroids abruptly can also cause adrenal problems and therefore this needs careful supervision.

### **Terbutaline subcutaneous continuous infusion**

This treatment is used for severe asthma which is uncontrolled with maximal therapy including high dose inhaled steroids and/or oral steroids. Children using a terbutaline pump are likely to be on multiple other medications as well as the terbutaline subcutaneous pump. The drug is administered using a syringe and pump device approximately the size of a mobile telephone. The drug is given as a 24-hour infusion through a needle placed under the skin and the dose is adjusted depending on asthma control. The device and care needs associated with it are the same as looking after an insulin pump for a child with difficult to control diabetes.

### **Theophylline**

Theophylline is a smooth muscle relaxant used to open the airways. It is used in the emergency setting and can be used daily in hard-to-treat asthma.

see: [How is it treated & managed](#)

### **Inhalers and other drug delivery devices**

#### **Metered Dose Inhaler**

Different devices are used in asthma – the aim of the device is to deliver the drug to the lungs as efficiently as possible. An adult or older child can use a standard inhaler, sometimes called a metered dose inhaler. This delivers a puff or spray of medication when pressed. Breathing in and pressing the inhaler have to be co-ordinated so the spray is breathed into the lungs rather than landing in the mouth. A demonstration can be viewed here -:

[http://www.asthma.org.uk/using\\_your\\_inhaler.html](http://www.asthma.org.uk/using_your_inhaler.html). (Link provided with kind permission of Asthma UK).

### **Spacer Devices, masks and mouthpieces**

In young children, metered dose inhalers are still used but with a spacer device to overcome the need to co-ordinate breathing in and pressing the inhaler at the same time. The child breathes normally in and out through a mask or mouthpiece into the spacer device. The reservoir of air inside the spacer contains a puff of medication that is breathed in and delivered to the lungs. In the under 5s, the parent will usually need to hold the spacer and actuate or press the inhaler to release a spray of medication. A demonstration can be viewed here -: [http://www.asthma.org.uk/using\\_your\\_inhaler.html](http://www.asthma.org.uk/using_your_inhaler.html). (Link provided with kind permission of Asthma UK).

### **Dry powder inhalers and breath actuated inhalers**

Other types of inhalers include dry powder inhalers which deliver a spray of powder rather than aerosol. Some of these are breath actuated – this means that breathing in actuates the dose of medication and co-ordination is not required. Children can usually use these with minimal supervision from age 8. A demonstration can be viewed here: [http://www.asthma.org.uk/using\\_your\\_inhaler.html](http://www.asthma.org.uk/using_your_inhaler.html). (Link provided with kind permission of Asthma UK).

All of the devices are simple and straightforward to use and take a few minutes on each occasion.

### **Nebulisers**

Nebulisers are most commonly used in the treatment of acute asthma. Some patients with severe asthma may be given a nebuliser to use in asthma attacks whilst waiting for medical help. It is rare for families to be given nebulisers to deliver daily treatment, although possible in young children who struggle to use inhalers even with a spacer. Some parents of children with asthma buy their own nebuliser even when this has not been recommended by their doctor.

Nebulisers are more time consuming. Nebuliser drugs are supplied in packets of individual doses. Each individual dose may need to be mixed with nebuliser solution for use. Whether mixing is required or not the individual dose has to be decanted into the reservoir of the nebuliser and the machine switched on. The dose is delivered by mask over around 15 minutes. The parent will need to supervise to ensure that the whole dose is delivered, and



that the child keeps the mask on throughout the period. The nebuliser machine itself needs to be carefully maintained and serviced. Check that a nebuliser is being used by looking for evidence of prescription of drugs for

use with the nebuliser. These include nebuliser or respirator solution and Salbutamol nebules or equivalent.

If a nebuliser is being used to deliver low dose steroids it is usually because the child is very un-cooperative with standard treatment or for some other reason often related to disability. It is a marker of significant care needs.

There is only one type of nebulised steroid available called 'pulmicort respules'.

### **Non-drug treatments**

There are many non-drug treatments, a lot of which are subject to ongoing research, therefore only those where there is evidence of beneficial effect are listed here.

<b>Prevention of Asthma</b>	
Avoidance of tobacco smoke	Parents should be advised of the many adverse effects smoking has on their children including increased wheezing in infancy and persistent asthma in later childhood.
<b>Dietary manipulation</b>	
Weight reduction	Weight reduction is recommended in obese patients with asthma to promote general health and improve asthma control.
<b>Secondary prevention of Asthma</b>	
House dust mites	In committed families, multiple measures to reduce house dust mite may help.
Smoking	Parents should be advised about the dangers to their children of smoking and offered appropriate support to stop smoking.
Smoking	Children who smoke should be advised about the dangers of smoking and offered appropriate support to stop smoking.
Immunotherapy	May be beneficial where a significant allergen cannot be avoided.
<b>Complementary and alternative medicine</b>	
Buteyko technique	A breathing technique that can be learned and may help some people control symptoms of asthma.
Family therapy	In difficult childhood asthma, there may be a role for family therapy in addition to drug treatment.

[How is it treated & managed](#)

## Steroid Inhaler doses by age

### Beclometasone Dosage Details

<b>Beclometasone Steroid Inhaler - dose by age</b>
Beclometasone – standard or low dose by age
Child under 2 years 5 micrograms twice daily
Child 2-5 years 100 micrograms twice daily
Child 5-12 years 100-200 micrograms twice daily
Child 12-18 years 100-400 micrograms twice daily
Beclometasone – high dose by age
Child under 2 years 100 micrograms twice daily
Child 2-5 years 400 micrograms twice daily
Child 5-12 years up to 400 micrograms twice daily
Child 12-18 years 400-1000 micrograms twice daily

[How is it treated & managed](#)

### Budesonide Dosage Details

<b>Budesonide Steroid Inhaler - dose by age</b>
Budesonide – standard or low dose by age
Child under 2 years 50-100 micrograms twice daily (rarely used in this age group)
Child 2-5 years 100-200 micrograms twice daily (rarely used in this age group)
Child 5-12 years 100-200 micrograms twice daily
Child 12-18 years 100-400 micrograms twice daily
Budesonide – high dose by age
Child under 2 years up to 200 micrograms twice daily (rarely used in this age group)
Child 2-5 years up to 400 micrograms twice daily (rarely used in this age group)
Child 5-12 years up to 400 micrograms twice daily
Child 12-18 years 400-1000 micrograms twice daily

[How is it treated & managed](#)

### **Fluticasone Dosage Details**

<b>Fluticasone Steroid Inhaler - dose by age</b>
Fluticasone – standard or low dose by age
Child 4-12 years 50-100 micrograms twice daily
Child 12-18 years 50-200 micrograms twice daily
Fluticasone – high dose by age
Child 4-12 years 100-200 micrograms twice daily
Child 12-18 years 200-500 micrograms twice daily

[How is it treated & managed](#)

### Fluticasone Propionate Dosage Details

Fluticasone Propionate Steroid Inhaler - dose by age
Fluticasone Propionate standard or low dose by age
Child 4-12 years 50-100 micrograms twice daily
Child 12-18 years 50-200 micrograms twice daily
Fluticasone Propionate high dose by age
Child 4-12 years 100-200 micrograms twice daily
Child 12-18 years 200-500 micrograms twice daily

Combination inhalers (steroid plus long-acting beta agonist)

**Seretide** – fluticasone and salmeterol

**Relvar ellipta** – fluticasone and vilanterol. Note this inhaler delivers the steroid as small particles. This means that 100mcg of fluticasone delivered from this inhaler is equivalent to 250mcg delivered from a standard inhaler

**Fostair** – beclomethasone and formoterol. Note that this inhaler delivers the steroid as small particles. This means that 100mcg of beclomethasone delivered from this inhaler is the equivalent to 250mcg from a standard inhaler.

### What you need to know about Attention Deficit Hyperactivity Disorder (ADHD) / Attention Deficit Disorder (ADD)?

## **What is Attention Deficit Hyperactivity Disorder (ADHD) / Attention Deficit Disorder (ADD)?**

ADHD / ADD is a behavioural disorder characterised by inattention, over activity and impulsivity...

### [Attention Deficit Hyperactivity Disorder \(ADHD\)](#)

## **What are the effects and signs?**

The core symptoms of ADHD / ADD are:-

Inattention: Easily distractible, forgetful and difficulty sustaining tasks such as play, learning and work .....

- [Effects of ADHD](#)

## **How is it assessed?**

According to the latest NICE guidelines, the diagnosis of ADHD / ADD should only be given by accredited Child and Adolescent Psychiatrists .....

- [Assessment of ADHD](#)

## **How is it treated and managed?**

Children with ADHD / ADD require integrated care that addresses a wide range of personal, social and educational needs .....

- [Treatment for ADHD](#)

## **What evidence is available?**

A Psychiatrist report - crucial in those under 6yrs old. Gives a profile of significant behavioural issues, medication and most up to date treatment and its effect.....

- [Evidence](#)

## **How long will the needs last?**

For claims made between the ages of 10-15 yrs old, it is reasonable to award for 2 years....

- [Prognosis and duration of the award](#)

## **What is Attention Deficit Hyperactivity Disorder (ADHD) / Attention Deficit Disorder (ADD)?**

ADHD or Attention Deficit/Hyperactivity Disorder is a neurodevelopmental disorder. Neurodevelopmental disorders are impairments of the growth and development of the brain or central nervous system. They are persistent through that person's life. The onset of ADHD is in childhood and there are persistent patterns of severe inattention, hyperactivity and/or impulsivity. These behaviours have to exist in a number of settings, for example, at home, socially, at school, are excessive and not appropriate for that child's age.

These behaviours can lead to impairment in social, academic and (later) occupational functioning.

Children with untreated ADHD are 100 times more likely to be excluded from school. In addition, the Youth Crime Action Plan showed that ADHD was one of the main risk factors for criminal offending during childhood.

Treatments that are available for children with ADHD can substantially improve their behaviours and academic outcomes. Response rates for therapy are high. Diagnosis is therefore important as there are effective and well tolerated treatments for ADHD.

Children with ADHD have the same range of intelligence as children without the condition. Early low scoring on IQ testing usually improves with treatment. The low scoring is as a result of the ADHD affecting the child's ability to perform in testing rather than representing a genuinely low IQ. However, ADHD can also present in children with a learning disability.

Attention Deficit Disorder (ADD) is an older term used to describe the presence of more severe difficulties with inattention and milder symptoms of impulsivity/hyperactivity. Hyperkinetic Disorder (HKD) is used to describe more severe difficulties with hyperactive/impulsive behaviour and milder ones with inattention.

What causes ADHD?

Currently, it is believed that ADHD is caused by the interaction between abnormalities in different genes and variable environmental risk factors. Risk factors for developing ADHD include adverse maternal mental health during pregnancy, drug and alcohol exposure during pregnancy and illness of the baby during delivery and shortly after birth. The interaction between abnormal genes and environmental risk factors leads to abnormal function over multiple brain pathways and systems. There is no single cause for the condition and so no single treatment.

What is the prevalence of ADHD?

Prevalence is the proportion of a population found to have a condition.

The overall prevalence in the UK amongst all children is 5.3%. For children under 12 the figure is 6.5% and for Adolescents it is 2.7%. This difference is



explainable by the tendency for this condition to improve with and without therapy over a lifetime. These figures are for all children diagnosed with

ADHD whether or not they are on treatment. The condition affects more boys than girls with a ratio of between 6 and 9 to 1.

### **What are the symptoms of ADHD?**

The presentation of these children is highly variable. Some may have only very minor symptoms while others may have severe impairment

The core symptoms are:

#### Inattention

- Fails to give attention to details or makes careless mistakes in schoolwork.
- Difficulty sustaining attention in tasks or play.
- Does not seem to listen when spoken to directly
- Does not follow through on instructions and fails to complete tasks/schoolwork
- Has difficulty in organising tasks and activities
- Easily distracted by external irrelevant stimuli
- Forgetful in daily tasks.

#### Hyperactivity

- Fidgets or taps feet/hands. Squirms in seat.
- Leave seat in situations in which remaining seated is expected
- Runs about or climbs in situations where it is inappropriate
- On the go or seems driven by a motor.
- Talks excessively

#### Impulsivity

- Has difficulty waiting his or her turn
- Interrupts or intrudes on others
- Blurts out answers before questions have been completed

- Talks excessively without appropriate response to social constraints
- Does not foresee the consequences of an action ('acts before he/she thinks')

### **How is ADHD diagnosed?**

ADHD diagnosis is based on interpreting data gathered from multiple sources. These sources are the child's parents, teachers and the child him/herself. The information is gathered in questionnaire format as well as on face-to-face assessments. The diagnosis of ADHD is made in a secondary or tertiary health care setting. This may be within a multidisciplinary team, typically consisting of Child and Adolescent Mental Health services (CAMHS) - usually headed by a consultant psychiatrist but also consisting of social workers, clinical psychologists and psychiatric nurses, paediatricians and clinical/educational psychologists.

Alternatively, the child may be assessed and diagnosed within a uni-disciplinary service, for example, a Community Paediatric clinic. There is no single service model and arrangements vary from area to area. If a diagnosis of ADHD is stated on a claim it would be expected that the child will have been seen within a consultant-led team and details of the method of assessment used should be available, often in the consultant's letter.

If such evidence is lacking and there appears to be doubt about the validity of the diagnosis, the claim should be discussed with an HCP.

Diagnosis of ADHD requires that there be a combination of overactive, poorly modulated behaviour with marked inattention and lack of persistent task involvement. There must be no other identified cause which can account for this behaviour.

### **Core features required for ADHD diagnosis**

Exclude diagnosis of anxiety disorders, affective mood disorders, pervasive developmental disorders and schizophrenia:

- Symptoms present since before the age of 7
- Several symptoms are present in more than one setting such as home and school
- At least 6 of the inattention symptoms are present for more than 6 months or at least six of the hyperactive/impulsive symptoms are present for at least 6 months

**ADHD should be considered in all age groups, with symptom criteria ad-justed for age-appropriate changes in behaviour. However,**

**although with hindsight, traits suggestive of ADHD can be present in very young children, it is unusual for a confirmed medical diagnosis to be made in children under 5.**

### **How is it treated and managed?**

Treatment falls in to two main categories:

#### Behavioural Therapies

These are aimed at the child, their family and also at schools/teachers.

- Behavioural Parent Training
- Teacher Training and behavioural classroom management strategies
- Social skills Training
- Cognitive Behavioural Therapy (CBT). This can be offered to adolescents who are motivated to improve their own symptoms.

A combination of several of these is often required. This is called a Multi-modal Psychosocial Intervention.

Studies of behavioural interventions versus treatment with medication for ADHD have shown that, although the effect on a child's main symptoms is less in the first few months, the lasting effect on behaviour is similar in the long run.

#### Drug Therapy

All drug therapies for ADHD can have significant side effects and therefore all children being administered these medications will be under follow up with a specialist service. Look for evidence of letters from these services detailing the therapy and its beneficial effects as well as any potential side effects that they may be suffering from. If the drug is having no effect on behaviour it won't continue to be prescribed but the benefits of drug treatment will vary and be greater in some children than in others.

#### Treatment Recommendations for Different Patient Groups

##### Pre-school age children

Principal therapy is Parent Training and Education Programme either as a group or on an individual basis. Children of this age would usually be referred to a Tertiary Specialist Unit if their ADHD was severe enough to require medication. This would be the case if they had severe ADHD or if they had failed behavioural therapies for moderate ADHD. Tertiary specialist teams would be required because of the length of time that they would

potentially need to take the medication coupled with the potential for suppressing their growth over this time period.

### School Age Children

- Mild ADHD - These children will be managed with group-based Parental Training Programmes. These programmes involve identifying specific problem situations and specific behavioural problems. The parents can then be taught how best to manage these with improved communication, specific reward systems (tokens for good behaviour) and how to manage bad behaviours with appropriate mild negative consequences and response cost systems (removing tokens from the jar). Responses to behavioural interventions will be monitored by the diagnosing specialist. Further training for parents can be recommended if it is felt necessary.
- Moderate ADHD - Principal treatment is with Group Based Parental Training and Teacher Education programmes. Responses to behavioural interventions will be monitored by the diagnosing specialist. Further training for parents or teachers can be recommended if it is felt necessary. Teachers can be taught how to manage pupils with ADHD in the classroom. Additional support can be arranged via school psychologists to help devise individual learning programmes. Teaching assistants can be used to help with more one-to-one teaching if required. If these are ineffective or interventions are refused by the child, then medication may be considered.
- Severe ADHD - If severe impairments are present, then medication is the first line choice for treatment. Medication can be considered first line for severe ADHD cases but usually in conjunction with behavioural education programmes for parents, teachers and the children themselves.

### ADHD Subtypes

ADHD has been subdivided into different symptomatic subgroups. These subtypes will influence the symptoms that are commonest at presentation. However, overall functional ability is more affected by the severity of the symptoms rather than the individual subtype presentation. All subtypes have similarly good responses to both types of therapy.

### Medication

#### Methylphenidate (Concerta, Equasym, Ritalin, Medikinet)

This is a drug, which acts on the brain to increase levels of neurotransmitters. It is felt that increasing the levels of these neurotransmitters is responsible for the effectiveness of the drug on ADHD behavioural patterns. Treatment effect includes increased concentration, reduced levels of disruptive

behaviours, reduced aggression and improved relationships with parents, teachers and peers.

- Concerta XL: dosage range 18mg to 54mg once daily
- Equasym XL: dosage range 10mg to 60mg once daily
- Ritalin: dosage range 5mg to 30mg twice daily. Maximum daily dose is 60mg.
- Medikinet XL: dosage range 18mg to 54mg once daily

#### Undesirable Side Effects

- Very Common (1-10% of children will experience a degree of)
  - Nervousness/Insomnia

#### Common

- Anorexia and accompanying weight loss
- Mild growth retardation
- Depression and Anxiety
- Tiredness
- Abdominal Pains and Diarrhoea

#### Lisdexamfetamine (Elvanse)

The drug acts by increasing noradrenaline and dopamine within the brain. These are both neurotransmitters. Treatment effect includes increased

concentration, reduced levels of disruptive behaviours, reduced aggression and improved relationships with parents, teachers and peers.

- Dosage range 20-70mg once daily

#### Undesirable Side Effects

Very common (more than 10% will experience a degree of)

- Decreased Appetite
- Weight Loss
- Insomnia

Common (1-10% may experience a degree of)

- Unstable mood
- Aggression
- Somnolence (strong desire to fall asleep)
- Abdominal pain and Diarrhoea
- Nausea and Vomiting

#### Atomoxetine (Strattera)

Noradrenaline is a neurotransmitter (chemical messenger that allows brain cells to communicate) and this medication allows existing levels in the brain

to last longer before being broken down. The higher-than-normal levels of Noradrenaline are felt to be the reason why children's behavior improves.

- Dosage range: 10-80mg once daily

#### Undesirable/Side effects

- Very Common (greater than 10% will experience a degree of)
  - Appetite reduction
  - Tiredness
  - Abdominal Pain
  - Nausea
- Common (1-10% will experience a degree of)
  - Anorexia (loss of appetite) and Weight Loss
  - Irritability
  - Insomnia

#### Guanfacine (Intuniv)

Like atomoxetine, this is a non-stimulant drug. It is of proven benefit in treating the symptoms of ADHD though there is uncertainty about exactly how it works.

- Dosage range: 1 – 7 mg daily

#### Undesirable side effects

##### Common or very common

- Anxiety
- Appetite reduction
- Heart rhythm disturbances
- Constipation
- Diarrhoea
- Dizziness



- Drowsiness
- Dry mouth
- Mood alteration
- Nausea or vomiting
- Skin reactions
- Sleep disturbance
- Weight gain

### Treatment Effects

Medication for ADHD is very effective and works quickly. It is prescribed for moderate and severe ADHD only. Studies have shown that effects will often

be noticed after one week with full treatment effects appearing after 6-8 weeks of treatment.

For every 5 children treated with medication, one child will improve from severe/moderate ADHD to being in the mild category. Of the other 4 children, 3 will experience a significant reduction in symptoms.

The number of children that respond to their first medication is 70%. If a second drug is tried after the first one fails or is not tolerated, then the numbers of children that improve behaviours is 95%.

The behavioural effects are:

- Improved concentration
- Improved social interactions
- Reduced aggression
- Reduced hyperactivity and impulsivity

Children taking their medication are rated as more co-operative, better fun to be with and more likely to be a best friend by their peers.

**What evidence is available?**

Sources of evidence	Information Provided	Limitation of Evidence
Claimant/Carer	First source of information and associated evidence. SEN statement/Education Healthcare plan or (in Scotland a Co-ordinated Support Plan - CSP) or Individual Education Plan (IEP).	Does not have specialist knowledge.
Multidisciplinary Team (community/hospital paediatrician, consultant psychiatrist, specialist nurse, Associated health specialists for example, occupational therapist, speech and language therapist, physiotherapist)	<p>Clinical features, treatment and information about disability and needs.</p> <p>Also request a copy of a completed standardised assessment (adaptive behaviour scale, Connors rating scales, child and adolescent psychiatric assessment).</p>	Information regarding symptoms and disabling effects may be based on what the parent/carer has told the relevant professional.
<p>Special Educational needs teacher/co-ordinator</p> <p>School Report</p>	<p>Resulting disability and needs.</p> <p>Differentiate between support for learning and supervision for behaviour (latter more important for DLA).</p>	<p>May not have information about symptoms, investigations and treatment but this may vary depending on the type of school the child is attending.</p> <p>It is unusual in children with ADHD, who are receiving treatment, for behaviour at school to be significantly different from that at home.</p>

	<p>Clinical features, treatment and some information about disability and needs.</p> <p>May be the only source of information if no other professionals involved.</p>	<p>Does not have specialist knowledge. May not have the most recent information about disability and needs.</p>
	<p>Resulting disability and needs. Request copies of previous assessments if available.</p>	<p>Unlikely to have information about symptoms, investigations and treatment.</p>
	<p>Educational features, educational intervention required and information about disability and needs.</p> <p>Request copy of SEN statement/Education Healthcare plan or (in Scotland a Co-ordinated Support Plan - CSP) or any other educational involvement.</p>	<p>May not have up to date information if assessments are not recent. Dependant on type of school claimant attending.</p>

### **How long will the needs last?**

Children who are awaiting formal diagnosis or are undergoing assessments for behavioural problems will generally receive a diagnosis within 6 to 12 months of referral.

Awards for as yet undiagnosed behavioural disorders should be discussed with the HCP. The HCP can review the attached evidence and advise whether a disorder is likely to be present and whether it is responsible for needs claimed.

Regardless of severity, ADHD responds to therapy in most cases. Medication should have beneficial effects within 6-8 weeks of starting treatments. Onset for purely behavioural therapy would expect to be longer but certainly

some improvement should have been detected within 3 months, or the child would have been switched to medication.

The following table is to be followed if an award is made. It is not a statement of intended awards for individual categories

Age at date of claim	Length of Award
Age 18 months to 9 years	Two years if new diagnosis, or to age 12
Age 10 to 11 years	Two years
Age 10 to 15 years	Two years if new diagnosis, or to age 16
* Recent diagnosis made (up to 12 months before the claim is made)	If an award is made it should be made for two years

\* Recent diagnosis: (up to 12 months before the claim is made)

In some instances, claims may be received shortly after a diagnosis has been made and if so, will often be before or shortly after medication has been started. The treatment effect of medication and behavioural therapies for ADHD can be quite marked both in the short and long term, with onset of action within a couple of months. As a result, if an award is made based on current needs these may change once therapy is commenced. To reflect this, it is advised that awards in this instance should be for two years. The claimant's needs can then be re-assessed once treatment has been stabilised.

#### **Night needs:**

Night needs in ADHD are likely to be limited to intervention being needed to stabilise behaviour.

**Note:** When severe learning difficulties or overt psychiatric conditions are present, ADHD / ADD appear to be less important in affecting the needs and the severity.

**Note:** In cases of undiagnosed ADHD or a young child under school age where it may be difficult to identify the level of functional severity, the

Decision Maker should consider entitlement and award duration taking account of the Departments Medical Services advice as required.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Commonly Associated Co-morbid Conditions**

Anxiety	Dyspraxia
Autism Spectrum Disorders (ASD)	Tic disorders
Depression	Tourette syndrome
Dyslexia	

### **Oppositional Defiant Disorder**

A pattern of angry/irritable mood, argumentative/defiant behaviour, or vindictiveness lasting at least 6 months as evidenced by at least four symptoms from any of the following categories and exhibited during interaction with at

least one individual who is not a sibling. For further information - see: [Net-doctor](#).

#### Angry/Irritable Mood

- Often loses temper
- Is often touchy or easily annoyed
- Is often angry and resentful

#### Argumentative/Defiant Behaviour

- Often argues with authority figures or for children and adolescents with adults
- Often actively defies or refuses to comply with requests from authority figures or with rules
- Often deliberately annoys others
- Often blames others for his or her mistakes or misbehavior

#### Vindictiveness

- Has been spiteful or vindictive at least twice within the past 6 months

#### Severity

Mild: Symptoms are confined to only one setting (for example at home, at school, at work, with peers).

Moderate: Some symptoms are present in at least two settings.

Severe: Some symptoms are present in three or more settings.

#### Sleep Disturbance

##### Sleep problems in 1–4-year-olds

Sleep problems are common in this age group. Common problems include the child does not go to sleep when put to bed (bedtime resistance) or wakes in the night for attention (night waking). This is usually related to parent behaviour around the child's sleep routine at an early age. Half of children in this age group wake at least once during the night. Frequent waking (> 3 times a night every night) peaks in infancy (10% of all infants) and steadily declines in the preschool years. Waking more than three times a

night is becoming unusual and can be a marker of an underlying neurodevelopmental problem.

Bedtime resistance is common and can be considered normal. [OBJ]

#### Normal sleep in 5–10-year-olds

At age five 11 to 12 hour nighttime sleeps are average; normal sleep duration varies from 9.9 to 12.7 hours. This continues until the age of about nine when the need for sleep reduces slightly and the bedtime can be a bit later. By the age of eleven average sleep duration has dropped to 9 hours 49 minutes with a normal range of 8.6 to 11 hours.

No age at which a child can be expected to settle themselves back to sleep can be given. The age that this is achieved will depend on the personality of the child and the parenting given. Some children can settle themselves from 8-9 months of age. Many can do this by five years old but many healthy 5-year-old children cannot. Self-settling or self-soothing can be taught, and this is the basis of many behavioural interventions.

#### Sleep problems in 5–10-year-olds

Sleep problems reduce once the child has started primary school. 78% of five-and-a-half-year-olds sleep through and night waking becomes progressively less common with age, 87% of nine-and-a-half-year-olds usually sleep through.

There are a number of reasons that a child might be taking a long time to fall asleep. Sleep latency (time to fall asleep) longer than 1hr, on 5 out of 7 nights each week is unusual however, it is greatly influenced by the home life and parenting that the children receive. Medical advice should be sought if behavioural interventions do not help.

By five years of age waking more than three times a night is very unusual (<1%) and can be a marker of an underlying neurodevelopmental problem.

Sleep continuity is another way to conceptualise sleep quality and a child who does not achieve 7 hours continuous sleep on 5 out of 7 nights each week is unusual and suggests medical advice should be sought.

The ALSPAC study on children with ADHD showed the attention deficit hyperactivity disorder group had slightly more night-waking (three or more times a night) at every age, becoming most significant from about 5 years.

Waking at night three or more times every night occurred in 3% of children with ADHD at five years old and in 6% of children at eleven years old. This



compares with less than 1% of children without ADHD waking frequently (3 or more times every night) at age five.

#### Normal sleep in 11–16-year-olds

In the teenage years, the bedtime moves later and instead of having problems with the children waking at night, the children are very hard to wake in the morning. British data shows that at age eleven, average sleep duration is 9 hours 49 minutes with a normal range of 8.6 to 11 hours. There's no detailed British data on sleep after age eleven but from other studies we can generalise that on average nighttime sleep duration falls by about 30 minutes between eleven and sixteen. Duration of normal sleep varies from about 8 to 11 hours per night in Swedish sixteen-year-olds. Night waking on all or most nights is not very common at age 12 (5%) and age sixteen (2%). Sometimes waking at night is common - 20% at age twelve and age sixteen (Swedish data).

#### Sleep problems in 11 - 16-year-olds

##### Night waking

At age eleven children are developing independence in all areas including sleep. This means children having more of a say on their bedtime and often waking themselves up to go to school. At this age night waking requiring settling is not normal, we would expect a child to be able to go back to sleep by themselves if they woke at night. Children may need attention at night if they wake and misbehave, this is unusual.

Waking most nights is unusual by age sixteen (2%). Frequent waking (3 or more times) will be much rarer than that.

An intervention for this would be supervision and discipline for the child who wakes at night and misbehaves because they cannot sleep e.g. plays games and wakes other members of the household or leaves the house in the middle of the night. This type of behaviour is most likely to be seen in children with a severe functional restriction and a severe behavioural disorder such as severe ADHD or oppositional defiant disorder. They may or may not be attending mainstream school. There will be evidence from the school

or community paediatrician confirming behaviour and sleep is severely affected.

## Melatonin treatment of sleep disorders

Use of melatonin indicates that the child's parents have consulted a specialist about their child's sleep problems, and they are receiving treatment.

## Living with ADHD – Example Profiles

### Mild ADHD

### Moderate ADHD

### Severe ADHD

#### Mild ADHD

These children have few, if any, symptoms in excess of those that are required to make the diagnosis. These symptoms result in no more than minor impairments in social functioning. For example, these children may have increased aggression and impulsiveness that makes them less popular than their peers. They may also struggle at times with attention in class and at home, though not more so than other children within their class.

Children with mild ADHD will be managed by parents and teachers who have undergone appropriate psycho-behavioural training. As a result, they will have little difference in mobility or care when compared to children of a similar age.

#### Example:

J is 8 years old and lives with his mother and two sisters. At a recent parent evening his mother learned that at times J struggle to focus on his schoolwork and occasionally he turns in homework of an unacceptable standard. He struggles with joining in with other children in the playground and has been involved in arguments during break. He only has a few friends.

At home J has difficulty in doing as he is told. He tends to fidget and frequently get up and down when watching films or at mealtimes.

J saw his GP and was referred on to the local Neurodevelopmental Disorder Clinic. After full assessment, his consultant paediatrician diagnosed Mild ADHD. His parents attended a behavioural intervention course. He was moved classes at school to a teacher who had been trained in dealing with children with ADHD and he was monitored from clinic for a year. In this time his behaviour at home and school improved so he was discharged.

Two years later J is still experiencing periods of inattentiveness in class but, his schoolwork and performance has improved, and he is getting on with his classmates much better. At home he still tends to be disorganised, but his

concentration has improved to such an extent that he is now able to concentrate for the length of a film that he is interested in watching.

Clearly J has mild symptoms that are managed and controlled with simple measures. He does not require additional supervision or guidance when mobilising. He requires supervision during the day, but this is not in excess of other children in his class. There are no nighttime needs as he sleeps well and goes to bed when requested.

He has no statement of special educational needs or individual education plan.

### Moderate ADHD

Those in the moderate category do not by definition have severe impairments or behavioural problems.

This is the difficult 'middle ground'. Children will have symptoms in excess of the diagnostic criteria, enough to cause some impairment at school, home and in social situations but not enough that they will have any noted severe impairment. Most of these children will be managed with behavioural therapy.

Children with moderate ADHD may require additional supervision during the day or at night. Children who suffer from insomnia prior to being treated with medication find that their nighttime symptoms improve. At school they may require educational support. This will be usually provided by a teacher who is trained to deal with children who have ADHD, backed up by input from educational psychologists who are usually provided by the local education authority. One to one tuition or learning support may be provided by volunteers, teaching assistants or by the teachers themselves. This is more likely to be the case if not on medication. The more markedly affected children in this group, however, will have been started on medication or have been included in multimodal behavioural interventions. Both of these treatments have established beneficial effects on behaviour. Both behavioural interventions and medication have high response rates (70%) and will experience a marked improvement in attention, social impairment, aggression and impulsiveness.

### Example:

Child G is 10yrs old. He lives with both parents and a younger brother. He is currently in year 6 and due to move up to the senior school in the next year. Recently it has becoming apparent that he has been struggling with his schoolwork and has fallen behind in several subjects. There has been difficulty in some of his peer groups due to arguments in the playground and G is finding it difficult to keep friends.

G's parents have noticed that he has become more and more disorganised over the last year or so. He has lost items of school equipment on a number of occasions and is often late leaving the house for school or social engagements due to the length of time it takes him to get ready. He is restless and

finds it difficult to concentrate at times, even when it is something that he enjoys.

G was referred to his local hospital where he was assessed by a paediatrician and a clinical psychologist. Moderate ADHD was diagnosed. Initially G was referred for CBT but struggled with appointment times and concentration during the session. In addition, his parents were unable to cope with their psycho-behavioural training. After 3 months and a detailed discussion with parents, G was started on Methylphenidate.

G was given support during his school day. He was placed in a form class with a teacher who has had some training in managing pupils with ADHD. A teaching volunteer was used to help with one-to-one learning for four hours per week. An individualized education programme was used to help G to catch up with his subjects. He has a statement of special educational needs. He initially required some additional supervision at break times to reduce the risk of arguments and to help to encourage him to socialize more. Input from educational psychologists at school helped support the teachers and teaching assistants so that they could, in turn, support G.

G was assessed after a further 3 months and he had substantially improved. His initial nausea and dizziness on the medication had settled and his behaviours had improved. At school he was concentrating much better. He had started to make up lost ground in his academic year and the playground arguments had dropped back to a similar frequency to his friends.

At home he has improved his organisational skills and concentration.

On his 12-month review G has now caught up in his schoolwork. His homework, though untidy, is of an acceptable standard. He is now more accepted by his peers and has one or two close friends. His medication is at a stable dose and the clinic will continue to review him on a 6 monthly basis.

At initial assessment G required additional help and supervision when at home. He required this during the day in order to get out of the house on time. In addition to this he needed supervision at school that would have been considered normal for a child several years younger than he was at the time. At night he required no supervision as he still went to bed before his parents. Out and about he required supervision to remain focussed enough to get to destinations. This was required for some of the time but was not most of the time and, was not largely in excess of that required for child of the same age.

After treatment his day care needs reduced down due to improvements in his behaviour. He requires little more supervision and guidance than would be expected for a boy of his age.

### Severe ADHD

Severe is reserved for cases with many symptoms in excess of those required for the diagnosis. One or two of the core symptoms can be present to an especially severe degree. The impairment in social and educational functioning is significant. These children will be under regular follow up with

a Consultant Paediatrician or Psychiatrist. They will be receiving treatment, either behavioural therapies or medication.

These children will be receiving treatment from specialist teams. Treatments, both behaviour and medication based, have enduring positive effects on behaviour. Those being treated with medication for more than 6 weeks should show marked improvements in behaviour at school and at home. After 6 weeks the medication effects are prolonged, so evening exacerbations are unlikely to occur.

Those who do not respond well to treatments will have documentary supportive evidence of this from CAHMS or Paediatricians. In addition, schools will be able to supply information about how the child's behaviour impacts on their educational and care needs during school hours.

It would be expected that children on therapy for ADHD would have greatly reduced care needs. Medication for ADHD can have significant short and long term effects on a child health and so is stopped if there is no improvement in behaviour or if side effects become significant.

Example:

D is a 13yr old girl. She is in year eight and has a maths and reading aged 1-2 years below her biological age. She is struggling with most of her subjects and is currently at risk of needing to retake her current academic year. Her teachers describe her as disruptive in class, difficult to control and she struggles to pay attention during activities.

At home she is rebellious, refuses to obey her curfews, going to bed late at night, frequently waking and failing to rise for school in the morning.

D is referred than assessed by a paediatric neurologist and a consultant paediatric psychiatrist at her local ADHD clinic. She is diagnosed with severe ADHD and offered, after discussion with her parents, Methylphenidate.

Initially tolerating the medication, she developed severe insomnia, and the decision was made to withdraw the medication. The next step is Atomoxetine which she tolerates much better. After 2 months there has been a marked improvement in her symptoms. Her attention has improved and as a result her schoolwork is of a more acceptable standard. She is less argumentative and has started to develop friendships with a couple of children in her age group. At home, although she still goes to bed late, she is able to stay asleep once she is in bed.

She was assessed as having special educational needs and an individual action plan was put in place. At school she was put in the class of a teacher who has been trained to teach children with ADHD. A personalised learning plan was put in place and a teaching assistant was employed to help with one-to-one learning. She is supervised at break times to help her to socialise better and to prevent disputes with other children. One evening per week she receives extra tuition at school from her form teacher. Her parents have monthly meetings with her teacher to keep up to date on her studies

and to help maintain consistencies between home and school environments for behaviour and learning.

After 12 months D has integrated much more into school life. With the additional help of a teaching assistant, she has caught up enough to remain in her current academic year. Her scores for reading and maths are age appropriate. At home her behaviour has improved, and she has started to go to bed at a more reasonable time.

She continues under specialist follow up every 6 months. Once her behaviour improves, she is enrolled in cognitive behaviour therapy (CBT) and her parents are signed up for a Psycho-education training course.

Before treatment D requires additional supervision and care during the day at home due to lack of concentration and poor time keeping. At school she requires more structured one to one time than would be expected to help her maintain her current academic level. She goes to bed late and after her parents. She is up and down during the night and requires repeated prompting to get up. When mobilising about she requires supervision to get to school on time and is picked up again after school to prevent her wandering and taking too long to return home.

After therapy she is getting on at school but still required one to one work to help her catch up and to keep on task. The help she is receiving is for a

large portion of the day and in excess of that which would be expected given her age.

Her sleeping pattern has improved so she would no longer require attention at night. She is no longer taken to and from school.

## **What you need to know about Autism**

### **What are Autism spectrum disorders (ASD)?**

Autism spectrum disorders (ASD) are neuro-developmental disorders (impairment of the growth and development of the brain or central nervous system), which arise in birth or in very early life.....

- [Autism spectrum disorders \(ASD\)](#)

### **What are the effects and signs?**

A key feature of the deficit apparent in people with Autism spectrum disorders (ASD) is their inability to understand the thoughts of other people and themselves.....

- [Effects of ASD](#)

### **How is it assessed?**

There has been substantial research into the causes of Autism spectrum disorders (ASD).....

- [Assessment of ASD](#)

### **How is it treated and managed?**

The evidence so far indicates that early social and communication based intervention is effective....

- [Treatment for ASD](#)

### **What evidence is available?**

An Occupational therapist gives a profile of significant sensory issues.....

- [Evidence](#)

### **How long will the needs last?**

Most children with Autism Spectrum Disorders (ASD) have difficulties that are life-long. ...

- [Prognosis and duration of the award](#)

## **What are the Autism Spectrum Disorders (ASD)?**

The Autistic Spectrum Disorders (ASD) are conditions that are due to impairments of the growth and development of the brain or central nervous system. They are characterised by impaired social interaction and communication, as well as stereotyped behaviour and interests. They have an



onset typically before 3 years of age but may not be identified until later. Those children who are on the spectrum may have a wide variation in disability and learning ability. There may also be the presence of additional disabilities.

Autistic spectrum encompasses all types of autism.

Normal children develop an inbuilt interest in the sight and sound of other human beings. Initially this is mainly of the mother or main care giver, though over time they develop communication and social skills that allow them to interact with others and to begin imaginative play. Playing games of this kind depends on the child developing the knowledge that other people have thoughts and feelings of their own. Play of this sort allows them to improve their knowledge and understanding of other people further. These skills are necessary for them to learn how to integrate into social life. This ability to understand that other people have their own thoughts and feelings is absent in children with ASD.

Autistic children do develop an interest in other people. This is variable and largely depends on the severity of their autism.

Differences between girls and boys

Studies looking into the difference between girls and boys with autism have found parents and peers see significant differences between the two. Parents report a higher prevalence of masking (see next paragraph) in girls, and peers were slower to identify autism in diagnosed girls than diagnosed boys when they first met and spoke to them. This difference is seen less when professionals assess girls and boys which may be because the diagnostic criteria is the same for girls and boys, but how the individual manages their autistic symptoms, and presents themselves in the world varies.

Masking is the social phenomenon where people with autism learn superficial skills to fit in, but it is more of an imitation of peers to compensate for difficulties than an understanding of the complex social construct we live in. It is felt that girls may use this technique more than boys which may result in girls being older at diagnosis and also have fewer problems in school. It is possible that this masking may be inherently stressful for the individual which results in a release of emotion when they are out of the environment in which they are masking. This may lead to meltdowns following school. Some parents may refer to this as the 'coke-bottle effect' where a child becomes increasingly anxious all day then 'explodes' when they get home.

In claims we usually look for consistency of needs across different environments, but a child who masks their problems may not show this. Note that not all children with autism will mask, and most children with autism who have needs will show consistent needs across school and home, but it is important to consider if masking may be a factor, such as in those where

needs are claimed at home in those that have few/no problems in school then meltdown immediately and significantly afterwards.

## PDA

Pathological demand avoidance (PDA) is something that professionals have had some debate over. It is not accepted as a separate diagnosis or sub-type of autism by the NHS, and it is not described in internationally recognised classification manuals for illness. However, it is seen by many mainstream autism assessors and autism charities (such as the National Autistic Society) as a recognisable profile of behaviours in some children with autism, and parents may put it as their child's diagnosis rather than autism. The main characteristic of these children's behaviour is to avoid everyday demands and expectations to an extreme extent. They may even avoid or self-sabotage things they really want to do as even their own wants are perceived as demands. It's usually an anxiety-based condition. Often, they are using their controlling behaviours, for example controlling what their parent does, to control severe anxiety. Of course, a child cannot have complete control over their environment and adults around them and when they perceive they have lost control; extreme emotion, aggression, socially maladaptive behaviours and meltdowns occur. These children often find school very difficult to cope with and may struggle even in specialist settings designed for children with autism. A flexible and non-confrontational set of strategies is needed for children with this type of profile.

## High functioning autism

Traditionally those functioning at the higher end of the autistic spectrum have been referred to as having Asperger syndrome. This terminology is likely to continue to appear on claims as well as in reports submitted in evidence for some time to come. However, it has been internationally agreed within the scientific community that this term should no longer be used. Diagnostically, children with high functioning autism by have no delay in onset of speech and are of high, normal or near normal intelligence. Over time, they are more likely to lead independent lives.

## What causes ASD?

Despite the convincing genetic basis for ASD, a definite cause remains unknown in most individual cases. The majority of ASD are probably caused by complex interaction between multiple genetic abnormalities and environmental exposures/risk factors. For example, Intra-uterine brain infections also increase the risk of ASDs (rubella, cytomegalovirus).

Perinatal and postnatal traumatic brain injury, cerebrovascular lesions, severe encephalopathy, and brain infections may lead to the development of

autistic symptoms. Autism can be part of other genetic conditions such as Smith Magenis syndrome.

What is the prevalence of the Autistic Spectrum Disorders?

The prevalence amongst the UK population as a whole has been quoted as 1-2 per 1000. Amongst children of all ages this equates to 1 in 100 meeting diagnostic criteria for ASD. The ratio of males to females is 4:1. However there is an ongoing debate as to whether the condition is being diagnosed more frequently. Many large-scale studies date back to the first decade of this century. Statistical information from the Department of Health in Northern Ireland gives the prevalence of autism among school children in 2017-18 as approximately 3%.

### **What are the effects and signs?**

Impairments in social interaction and social communication

Using Speech

Understanding Speech

Understanding non-verbal communication

Impairment of Imagination

Repetitive Behaviours

Sensory Issues

Sleep Disturbance

Associated Impairments

Using Speech

Speech development is frequently delayed in autism. Despite intervention, 10% of autistic children will not develop any speech for the whole of their lives. Children on the autistic spectrum who have speech, do not tend to communicate much when compared to children of a similar age. They often use phrases they have heard to communicate such as "Do you want some juice?" rather than "Can I have a drink?". Sentence structure tends to be abnormal with linking words not used "Can go shops" instead of "Can we go to

the shops in the car?" Even when present, linking words can be used incorrectly "Put cup in the table".

## Understanding Speech

Their understanding of others' speech can be as varied, although often their understanding of speech is better than their use of speech. A major characteristic of autistic children, no matter how good their language skills are, is their literal interpretation of language. For example: "It's raining cats and dogs." The autistic child is likely to struggle with this phrase. This can have implications for their understanding in school or in other environments, as we often use abstract phrases to communicate.

Their understanding of language does improve over time, but outcomes depend on the child's Intelligence level and the support that they receive. Children with high functioning autism will often develop normal language understanding by adulthood.

## Understanding non-verbal communication

Autistic children also struggle to understand other people's use of gestures, facial expressions and bodily movements. This ability is known as non-verbal communication. Autistic children have difficulty understanding non-verbal communication. They do not use as many gestures or facial expressions and tend to not use eye contact to communicate. As a result, children with autism can be mistaken for being rude or inattentive. Improvements over time occur more commonly with children on the higher end of the autistic spectrum.

Children on the autistic spectrum have difficulty making friends and understanding friendships. They prefer to play with younger or older children whom they can direct to follow their own agenda. They can have difficulty with empathy and can be aggressive towards their peers. In comparison they may be very passive, not understand social norms and more likely to be bullied.

## Impairment of Imagination

Autistic children can find imaginative play difficult and do not do much role play or play pretend play games in the same way as other children. Their play is often limited in scope and tends to be repetitive in nature. Toys are often handled purely for physical sensation or repetition. They can also have strong obsessions, especially children at the higher end of the spectrum. These obsessions tend to go through phases and can switch from one to another.

## Repetitive Behaviours

Simple repetitive actions include staring at lights, watching things spinning, touching surfaces, twisting and turning hands. Occasionally this can take the form of head banging, rocking, hitting or scratching. These forms of

behaviour tend to happen in the youngest of children with autism and usually become less as the child grows into adolescence. These behaviours can often occur in response to feelings of agitation or anxiety but can also be used as a soothing or stress relieving act. A small percentage of the most profoundly affected persist in these habits into adult life.

More complex repetitive behaviours would include watching the same segment of a DVD over and over or listening to the same song continuously.

In higher functioning children, the repetitive behaviour can develop into a specialist interest.

### Sensory Issues

Children on the autistic spectrum can have a number of sensory issues that can impact on their ability to participate in school or home life. Common hypersensitivities include sensitivity to loud noises such as hand driers, hoovers, loud busy environments such as supermarkets and school assembly. Other common sensitivities include certain clothing, smells and bright lights.

### Sleep Disturbance

Sleep disturbances are common in autism. The prevalence of disordered sleep is approximately 66% with the greatest problems occurring in the younger autistic children, in those with sensory issues and those with epilepsy.

#### Pre-School Age children:

Sleep problems are not uncommon in neurotypical children in this age group. Common problems include difficulty falling asleep when put to bed (bedtime resistance) or frequently waking in the night and demanding attention (night waking). The child's sleep pattern is influenced by how the parent manages the child's sleep routine at an early age. Half of children in this age group wake at least once during the night. Frequent waking (> 3 times a night every night) peaks in infancy (10% of all infants) and steadily declines in the preschool years. Sleep problems can be more severe in the more severe forms of autism.

Autistic children have been seen to need less sleep when compared with other children.

#### School Age children:

Many studies show that children with autism have a higher rate of nearly all sleep disorders. This includes problems settling, staying asleep and a shorter sleep duration. Sleep problems are common in children with autism and normal intelligence and even more common in children with autism and learning difficulties.

Children with autism sleep less and wake more at night than children without autism. Differences in sleep duration and night waking often emerge at between 18 and 30 months of age. By age five 7% of children with autism

are waking 3 or more times at night compared to less than 1% of neurotypical children. At age nine and a half 6.5% are waking 3 or more times at night compared to 0.5% of children without autism. Their sleep can also be affected due to feelings of anxiety.

Interventions for sleep problems:

Two types of interventions are usually tried:

- Settling to help the child go back to sleep
- Supervision of behaviour

Settling is only likely to be required in severely disabled children e.g. developmental disorders such as severe autism with learning disabilities. Such children are unlikely to be able to attend a mainstream school.

Supervision, having a good routine and having clear boundaries are often needed for children with sleep difficulties and their inappropriate nighttime activities such as playing games, waking other members of the household or leaving the house in the middle of the night. This type of behaviour may be due to a child's autism, but it can also be seen in non-autistic children for other reasons such as: poor sleep routine and parental control, other neurodevelopmental disorders such as ADHD or a chaotic home environment.

Use of Melatonin indicates the child's parents have consulted a specialist about their child's sleep problems and they are receiving treatment. Melatonin treatment may or may not be effective, you should contact the specialist for the latest information on the child's sleep habits.

### Associated Impairments

Intellectual disability (IQ below 70) coexists in approximately 50% of children and young people with autism.

Around 70% of people with autism also meet diagnostic criteria for at least one other (often unrecognised) psychiatric disorder that further impairs psychosocial functioning, for example, attention deficit hyperactivity disorder

(ADHD) or anxiety disorders. Half of these will have at least two mental health disorders.

Looking at the whole range of Autistic Spectrum, 20-25% of children have additional learning disabilities (dyspraxia, dyslexia, dyscalculia).

Epilepsy - 20-25% of autistic children will go on to develop seizures, usually as adolescents.

Anxiety Disorders - About 40% of children with ASD had at least one comorbid diagnosed anxiety disorder.

Depression - 10-20% of children on the Autistic Spectrum have symptoms of a depressive disorder.

Sleep - 40-86% of autistic children will have some additional difficulties with sleep, settling or frequent waking.

- [Asperger's Syndrome](#)
- [Living with Autism](#)

### **How is it assessed?**

ASD is diagnosed by multi-disciplinary teams set up specifically for this purpose. These teams will consist of Consultants specialising in Psychiatry and Paediatrics plus input from psychologists. In each area a multidisciplinary group (the autism team) will be set up. The core membership will include a: paediatrician and/or child and adolescent psychiatrist speech and language therapist clinical and/or educational psychologist.

The autism team should either include or have regular access to the following professionals if they are not already in the team: A paediatrician or

paediatric neurologist, child and adolescent psychiatrist, educational psychologist, clinical psychologist and an occupational therapist.

Diagnostic assessments will include:

- A full physical examination
- A full medical history
- A developmental history from the child's parents
- Direct assessment of the child's social and communication skills
- Information regarding the performance of the child at school from Teachers.

Autism Teams do not rely on any particular autism-specific diagnostic tool alone to diagnose autism.

### Diagnostic Criteria

A total of six (or more) items from (A), (B) and (C) comprising all three from (A), at least two from (B) and to include (C). (D) and (E) must also be satisfied.

- A.** Persistent deficits in social communication and social interaction across multiple contexts, as manifested by the following
  1. Abnormal social approach and failure of normal back-and-forth conversation; to reduced sharing of interests, emotions, or affect; to failure to initiate or respond to social interactions
  2. Poorly integrated verbal and nonverbal communication; from abnormalities in eye contact and body language or deficits in understanding and use of gestures; to a total lack of facial expressions and nonverbal communication
  3. Difficulties adjusting behaviour to suit various social contexts; from difficulties in sharing imaginative play or in making friends; to absence of interest in peers
- B.** Restricted, repetitive patterns of behaviour, interests, or activities, as manifested by at least two of the following
  1. Stereotyped or repetitive motor movements, use of objects, or speech (e.g., simple motor stereotypies, lining up toys or flipping objects, idiosyncratic phrases)
  2. Insistence on sameness, inflexible adherence to routines, or ritualised patterns or verbal nonverbal behaviour (for example, extreme distress at small changes, difficulties with transitions, rigid



thinking patterns, greeting rituals, need to take same route or eat food every day)

3. Highly restricted, fixated interests that are abnormal in intensity or focus (for example strong attachment to or preoccupation with unusual objects)
  4. Apparent indifference to pain/temperature, adverse response to specific sounds or textures, excessive smelling or touching of objects, visual fascination with lights or movement.
- C.** Symptoms must be present in the early developmental period. Before age 3 but age is flexible. Can be up to age 8.
- D.** Symptoms cause clinically significant impairment in social, occupational, or other important areas of current functioning.
- E.** These disturbances are not better explained by intellectual disability (intellectual developmental disorder) or global developmental delay.

### Severity levels

DSM-5 lists three levels of severity in each of the two domains ('social communication' and 'restrictive, repetitive behaviours').

The levels are:

- Level 1 - requiring support
- Level 2 - requiring substantial support
- Level 3 - requiring very substantial support

The complexity of the presentation of autism spectrum disorders means that the severity of impairment can only be assessed on an individual basis. Severity levels may vary by context and also fluctuate over time.

### Severe Autism IQ less than 70

- Severe deficits in verbal and non-verbal social communication skills.

For example, a child who has very little intelligible speech and who rarely initiates interaction with others. They mostly make unusual interactions that meet their needs only and respond to only very direct social approaches.

- Inflexibility of behaviour, extreme difficulty coping with change. They often have repetitive behaviours or do repetitive movements and have marked difficulty functioning in all spheres

For example, the child spends the majority of his time engaged in stereotyped behaviours to the exclusion of interacting with others. This child will

require 1 to 1 supervision, and this will usually take place in a special school or unit.

#### Moderate Autism IQ 71-84

- Marked deficits in verbal and non-verbal social communication skills; social impairments apparent even with supports in place; limited initiation of social interactions; and reduced or abnormal responses to social overtures from others

For example, a child who speaks in simple sentences, whose interaction is limited to narrow special interests and has markedly odd nonverbal communication.

- Inflexibility of behaviour; difficulty coping with change or other restricted/repetitive behaviours appear frequently enough to be obvious to the casual observer and interfere with functioning in a variety of contexts. Distress and/or difficulty changing focus or action. For example, the child may talk excessively about a specific topic which creates social difficulties with school peers and family members. Additional school support required, usually in a school for children with special needs. Low classroom numbers with additional teaching support required.

#### Mild Autism IQ greater than 85

- A child who is able to speak in full sentences and engages in communication but whose to-and-fro conversation with others fails; whose attempts to make friends are odd and typically unsuccessful; difficulty switching between activities; problems of organisation and planning hamper independence

For example, they may have difficulty shifting between tasks, which interfere with his ability to follow a school schedule.

- Additional educational support in mainstream school; a classroom assistant will be required for some extra help; a tailored educational plan to assist with schoolwork; requires assistance with socialisation

This help can be provided by teachers, assistants and educational psychologists.

### **How is it treated and managed?**

There are no medications that will treat or cure children on the Autistic Spectrum.

Treatment for ASD is tailored depending on the severity of the problems and level of need. Behavioural interventions coupled with treatments of any co-morbid illnesses are the best forms of therapy. Medication is only used as a

last resort for severe behavioural or sleep problems or any additional anxiety and depression.

There are many claims of a 'cure' for autism, all of which are without foundation.

Behavioural therapy includes specific social-communication interventions for the core features of autism in children and young people that includes play-based strategies with parents, carers and teachers to increase joint attention, engagement and reciprocal communication in the child or young person.

**Some children with autism who have very aggressive behaviour may be treated with antipsychotics for challenging behaviours. This type of prescription is initiated by specialists as these drugs are not currently**

**licensed for this type of problem. Such a prescription would indicate a substantial functional restriction.**

**What evidence is available?**

Sources of evidence	Information Provided	Limitation of Evidence
Parent / carer	First source of information and associated evidence for example a SEN statement/Education Health and Care plan or (in Scotland a Co-ordinated Support Plan - CSP) or an Individual Education Plan (IEP).	Does not have specialist knowledge.
<p>Multidisciplinary community team (Community / Hospital Paediatrician, Consultant psychiatrist, Consultant psychologist, Associated Health Specialists E.g. Occupational Therapist, speech and Language Therapist, Physiotherapist)</p> <p>(if child is under their care).</p>	<p>Clinical features, treatment and information about disability and needs.</p> <p>Also request a copy of a completed standardised assessment (e.g. the Adaptive Behaviour Scale) and a copy of a SEN statement/Education Health and Care plan or (in Scotland a Co-ordinated Support Plan - CSP).</p>	Information regarding symptoms and disabling effects may be based on what the parent / carer has told the relevant professional.
<p>Special Needs teacher/coordinator</p> <p>(if child has one).</p>	Resulting disability or needs.	May not have information about symptoms, investigations and treatment but this may vary depending on the type of school the child is attending.

GP.	<p>Clinical features, treatment and some information about disability and needs.</p> <p>May be the only source of information if no other professional involvement.</p>	<p>Does not have specialist knowledge.</p> <p>Unlikely to have detailed or recent information about resulting disability or needs.</p>
Educational psychologist (if child has seen one).	<p>Educational features, educational intervention required and information about disability and needs.</p> <p>Also request a copy of a completed standardised assessment (e.g. the Adaptive Behaviour Scale) and a copy of a SEN statement/Education Health and Care plan or (in Scotland a Co-ordinated Support Plan - CSP).</p>	<p>May not have up to date information if assessment not recent but this may vary depending on the type of school the child is attending.</p>

### How long will the needs last?

This guidance covers: -

Autism including Pervasive Development disorder (PDD) or Pathological Demand Avoidance syndrome (PDA)
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High functioning autism (formerly Asperger's Syndrome)
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What is the long term prognosis of ASD?

The long-term prognosis for ASD depends on the severity of the condition and any associated reduction in intellectual ability.

In severe infantile autism:

- 20% of children with severe infantile autism begin to improve between the ages 4 and 6 years and are able to attend ordinary schools and have ordinary employment.
- 20% of children with this condition can live at home but attend special schools and are not able to have ordinary employment.

- About 60% of children show little improvement and are unable to lead an independent life, most requiring long- term residential care.
- 10% of children with autism remain non-verbal throughout life

At the other end of the spectrum. Children with high IQ and milder symptoms will usually adapt their behaviour and appear normal in later life. As adults they are more likely than the general population to suffer with episodes of treatable depression, anxiety or obsessive-compulsive disorder.

The following table is to be followed if an award is made. It is not a statement of intended awards for individual categories

Age at date of claim	Length of Award		
	Mild	Moderate	Severe
18mths - 4 years	Award to age 7	Award to age 12	Award to age 12
5 to 11 years	Award to age 12 or for two years (whichever is greater)	Award to age 12 or for two years (whichever is greater)	Award to age 12 or for two years (whichever is greater)
10-12 years	Award to age 16	Award to age 16	Award to age 16

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### High functioning autism

This term is used to describe people who are assessed as functioning at the higher end of the autistic spectrum. They were formerly diagnosed as having Asperger's syndrome, which was regarded as a subtype of autism. However current thinking is that autism is one condition with variable severity (the autistic spectrum). Partly in recognition of this it has been agreed that the term Asperger's syndrome should no longer be used. Nevertheless, it is likely that the old terminology will continue to appear on claims and in documents provided in evidence for some time to come. Children with a diagnosis of high functioning autism will often have no delay in onset of speech and have high, normal or near normal intelligence. Although language is

preserved, non-verbal communication abnormalities are similar to those of other people on the spectrum. These children show diminished non-verbal communication behaviours such as facial expression, posture, eye contact and gesture. They can demonstrate social insensitivity or even apparent indifference, can have limited capacity for spontaneous social interactions and a limited number of intense and highly focused interests. It occurs more often in boys, the estimated male-to-female ratio is approximately 4:1.

Children with high functioning autism may be taught specific social guidelines but the underlying social impairment is believed to be life-long. The comparative preservation of function relative to classical autism may give



the impression of lack of needs, however the social impairment may be quite debilitating.

Co-morbid conditions

Depression

20% of children with high functioning autism suffer with an episode of depression in their lifetime. It is more common from adolescence onwards.

Anxiety

30% of children who have high functioning autism will also suffer from an anxiety disorder.

Epilepsy

20-25% of children with high functioning autism will develop Epilepsy

Children with high functioning autism are more likely to lead independent lives in adulthood.

## **Living with Autism – Example Profiles**

[Mild Autism](#)

[Moderate Autism](#)

[Severe Autism](#)

[Care and Mobility Considerations](#)

Mild Autism

A is 9 years old. He has language development along with reading and maths skills 3 years above his biological age. Although not good at team sports A is an excellent swimmer, competing for his local club.

He is very keen on Star Wars and is able to recite the script for episode 4 verbatim. He will watch it at home whenever he has any free time.

A has a few friends and manages to interact well one on one but struggles with the rules of play when in a group. He is easily led and has been in trouble a couple of times after doing something he was told to be one of the older boys at school.

He suffers from full bed wetting two nights a week at night, but this is slowly improving with time. Although he struggles to sleep at times he will drop off before his parents, provided an upstairs light is left on.

A has mild autism and his intellectual functioning and social problems would probably put him at the upper end of the autistic spectrum (high

functioning). He will be unlikely to qualify for an award and as he gets older the apparent nature of his problems should lessen as he develops coping strategies.

At school A's teachers are aware of his diagnosis. He is in a normal form class. He requires no additional help with his schoolwork. At break times he is more supervised to help encourage better socialisation.

#### Moderate Autism

R is 5 and lives with his parents and older brother. He started using single words at age 18 months but still doesn't use 2 words together. He stopped using words he had previously learnt between 18 and 24 months but has now regained most of these words.

He also seems uninterested in engaging with other children. He occasionally engages with his parents but less than they think he should. He doesn't tend to look at them much and he has difficulty maintaining eye contact with them. When he wants something, he pulls them to where the object is and screams; he doesn't point like other children.

His parents have also noticed that he does not play in the same way as other children of his age; he tends to line toys up, or plays with certain aspects of them, such as the car doors. He doesn't use the toys in the

imaginative way that other children do. When his toys are moved, he becomes very upset.

He tends to become distressed when he thinks there is change around the house. In contrast, he is not concerned when either of his parents leaves the house.

He tends to flap his hands at times and his parents report him staring at the ceiling lights for 10 to 20 minutes at a time. He is a fussy eater and hates being messy.

He has an Education, Health and Care Plan needs and goes to a local mainstream school with additional support/resource for children with autistic spectrum disorders.

### Severe Autism

J is 14 years old. He has very little speech. He can say parts of words, but these are really only intelligible to his mother and his small team of carers.

He sleeps for about 2hrs at a time and will try and run up and down his bedroom on an invisible track. When not doing this, he orders then re-orders his CD collection.

He also rocks backwards and forwards in a chair often in a trance like state.

J rarely makes eye contact, though sometimes he will put his face close to other people's making them feel uncomfortable.

When in public places he will run away from his mother. He has attempted to drink red hot tea straight down without pause.

He has associated learning difficulties, and his intelligence score is that of a 6-year-old on standard scales.

J's Statement of Special Educational Needs has recently been converted to an Education, Health and Care Plan. He attends a special school for children with severe autism. There are eight children in each class with two teaching assistants and a teacher. He receives 5 hours of speech and language therapy per week.

J clearly has significant problems and will qualify for both components of DLA. Given the degree of his problems it is unlikely that he will improve

substantially, and his current award could be extended up to the day before his 17<sup>th</sup> birthday.

### Care and Mobility Considerations

Requirements for care will vary widely amongst children with ASD.

A proportion of those children most severely affected (60%) are unlikely to improve significantly over their lifetime.

Those with high functioning autism (formerly known as Asperger's syndrome) would be expected to require less input and show the most improvement over time.

Autistic Spectrum Disorders are, by their nature, difficult to diagnose. Current NICE guidelines recommend that autism is diagnosed by specialist teams using interviews and assessments of the child, parent and teachers. Any claimant that has not been diagnosed in this way should have their case referred to an HCP. The HCP can then review any evidence and make an appropriate recommendation.

In cases where a diagnosis has been provided by a private paediatrician or private child psychiatrist then this diagnosis may be accepted provided that the diagnosis has been made with reference to DSM5 or ICD10 criteria. Autistic children with needs will, however, also have been made known to NHS services in order to access additional help for the child at home and in a school environment. Evidence should be available to support this in the form of a SEN statement/EHCP, school letters and GPs letters. An autistic child with no additional needs will not need to seek this help.

In situations where there is doubt then an opinion can be requested of the HCP.

## **Batten disease**

Batten disease is one of a small group of rare, progressive and fatal diseases of the nervous system. It is inherited and typically begins in childhood. There are some forms of Batten disease that begin very early in a child's life, within the first 6 months, but in general early symptoms can appear from the age of 2 onwards.

Over a short period of time the child experiences worsening cognitive impairment, seizures and progressive loss of motor skills and vision. Over the next few years children will become blind and immobile and require help with all aspects of daily living. Depending on the type of Batten disease,

death occurs from age 6 onwards, but affected children may die suddenly and sooner from seemingly trivial infections e.g. respiratory infections.

As this is a progressive disease, the level of disability does not improve.

A new drug treatment, Cerliponase Alfa (Brineura), which aims to slow down the progression of symptoms in Batten disease has recently become available. The early results are said to be promising but the long-term efficacy and safety are unknown. Treatment with Cerliponase Alfa is currently very

costly. At present, the National Institute for Health and Care Excellence (NICE) does not recommend its use within the NHS but is still considering it.

<b>Impairment</b>	<b>Award Period</b>	<b>Code</b>
Batten disease	Award to age 16	G99

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

## **What you need to know about Brain tumours**

<p><b>What is a Brain tumour?</b></p> <p>A brain tumour may arise from the brain itself or from .....</p> <ul style="list-style-type: none"> <li>• <a href="#">Brain tumours</a></li> </ul>
<p><b>What are the effects and signs?</b></p> <p>Symptoms of brain tumours are often initially non-specific and .....</p> <ul style="list-style-type: none"> <li>• <a href="#">Effects of a brain tumour</a></li> </ul>
<p><b>How is it assessed?</b></p> <p>Investigations used will depend on the cause of the problem and.....</p> <ul style="list-style-type: none"> <li>• <a href="#">Assessment of brain tumours</a></li> </ul>
<p><b>How is it treated and managed?</b></p> <p>Most children will have surgery for their brain tumour as this .....</p> <ul style="list-style-type: none"> <li>• <a href="#">Treatment of brain tumours</a></li> </ul>
<p><b>What evidence is available?</b></p> <p>A Hospital FR is the best source of evidence .....</p> <ul style="list-style-type: none"> <li>• <a href="#">Evidence</a></li> </ul>
<p><b>How long will the needs last?</b></p> <ul style="list-style-type: none"> <li>• <a href="#">Prognosis and duration of the award.</a></li> </ul>

### **What is a brain tumour?**

A brain tumour may arise from the brain itself or from the membranes surrounding the brain. There are over 100 different types of brain tumour, and they can be named from the originating type of brain cell or from the location of the tumour or both. The prognosis is very variable between differing tumour types. In children more than 80% of brain tumours are one of the

following types: Astrocytomas, Primitive Neuroectodermal tumours (PNET) or Ependymomas.

The symptoms of brain tumours in children depend on 3 main factors:

- age
- tumour location; and
- absence/presence of raised intracranial pressure

In this guidance symptoms are split into general symptoms that any child with a brain tumour may have and symptoms related to the tumour's location in the brain.

Whether a tumour can be cured will depend on how aggressive the tumour is (cell type and grade - see link at bottom of page), surgical accessibility and what treatment options are possible (determined by age of child and location of tumour). Generally high-grade tumours have a worse prognosis than low grade ones but some high-grade lesions have an excellent chance of cure (e.g. germinoma) and some low grade/ benign tumours may be life threatening and may cause significant disability.

This guidance will describe disabling effects that may arise as a result of the direct effect from the tumour in different parts of the brain and secondary effects as a result of morbidity/toxicity from the different treatment methods used (surgery, radiotherapy and chemotherapy). A list of brain tumours and their likely outcome in terms of survival is provided, although this can only provide a guide due to the complexity of prognostic factors (see link at bottom of page). Specific disabling effects and outcome of treatment can be anticipated if the tumour type, location and treatments used are known. Other effects such as fatigue and somnolence (drowsiness) may be more general and non-specific.

Medical evidence is likely to give the name of the tumour concerned and describe the location, treatment outline and resulting disabilities. Brain tumours very rarely spread to other organs unlike other cancers; however, some tumour types (e.g. PNET, ependymoma) can spread within the central nervous system to other parts of the brain or spine. This means that children with primary CNS or brain tumours have a different pattern of disabling effects compared to children with other kinds of cancer. When a brain tumour progresses, it gives rise to new symptoms/ signs and resulting disability by virtue of local extension, metastatic spread to other areas of the brain and possibly causing raised intracranial pressure.

### **What is the incidence/prevalence?**

Brain and spinal cord tumours are the second most common form of cancer in childhood after leukaemia. These make up 20-25% of cancers in children



and 350 to 400 new cases are diagnosed each year in Great Britain in those under 16 years of age. 5-year survival for this group as a whole is over 60%; however, this figure hides the wide variability in cure rates with some tumour types having a poor prognosis. Over half of survivors have significant and enduring neurological, developmental and or growth problems related to their disease or its treatment.

see: [World Health Organisation \(WHO\) classification of brain tumours](#)

### **What are the effects and signs?**

Symptoms of brain tumours are often initially non-specific and can start intermittently before becoming more persistent and worsening in severity. Only a third of children are diagnosed with their condition within a month of their first symptom. The commonest symptoms in children over 2 years are:

- Headache, usually frequent, recurring and gradually worsening (approx 33%)
- Nausea/vomiting (approx 32%)
- Ataxia/poor balance and clumsiness when walking (approx 27%)
- Fits (approx 13%)
- Squint/double vision (approx 7%)
- Behavioural changes (approx 7%)

Children under 2 tend to present with even more non-specific symptoms such as 'irritability' or 'failure to thrive'. Their most common presenting symptoms are:

- Macrocephaly -large head size for age (up to 40%).
- Nausea/Vomiting (approx 30%).
- Irritability (approx 24%).
- Lethargy (approx 21%).
- Head tilt (approx 7%).
- Developmental delay (approx 5%).

Brain tumours may cause raised intracranial pressure – usually as a result of a blockage in the circulation of the fluid that surrounds the brain and spinal cord (cerebrospinal fluid or 'CSF'). This is termed 'hydrocephalus' and is

sometimes referred to in lay terms as 'water on the brain' and presents with a particular pattern of symptoms including:

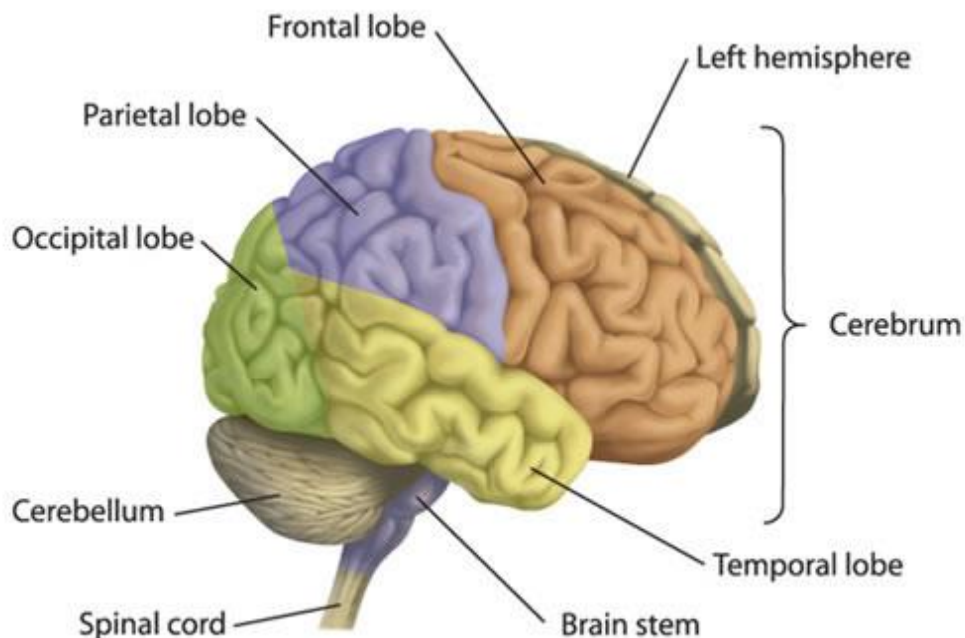
- Headache – may be severe, classically worse in the morning and on coughing or sneezing
- Nausea and vomiting
- Drowsiness - usually a late sign/symptom
- Blurred vision and blindness – raised pressure causes damage to the back of the eye. When there is swelling at the back of the eye this can be seen when the eye is examined with an ophthalmoscope - the characteristic appearance is called 'papilloedema'
- In young children the bones of the skull are soft and pliable – their head circumference may expand to accommodate increased amounts of cerebrospinal fluid (CSF). An enlarged head is called 'macrocephaly'

Without adequate treatment raised intracranial pressure causes death within a short period.

In addition to general symptoms there may be other symptoms that relate to where the tumour is in the brain. The following list gives an indication of symptoms and signs by location and is not exclusive or specific for any given child.

The brain is divided into lobes as illustrated below. The diagram shows the right side of the brain seen from the side. Note the names and locations of the different lobes. This picture has been reproduced from the website:

<http://sciencealive.wikispaces.com/Human+Brain> under [Creative Commons Attribution Share-Alike 2.5 License](#)



Childhood brain tumours are often grouped by the region of the brain they arise from. This includes supratentorial tumours; frontal, parietal, temporal, occipital lobe tumours and those in midline structures such as the pituitary gland, optic pathway, basal ganglia and thalamus. Infratentorial tumours or 'posterior fossa' tumours are more common in children than adults and include those arising from the cerebellum, IV Ventricle and brain stem.

### **Symptoms of brain tumours affecting the frontal lobe**

The frontal lobes determine personality and contain the area called the motor cortex that controls movement of the muscles of the body (the motor cortex on the right half of the brain controls the left side of the body and vice versa). The frontal lobes are thought to be the area of the brain most

involved in conscious thinking. Symptoms of tumours affecting the frontal lobe can include:

- Personality change
- Disinhibition – loss of inhibition leading to offensive behaviour which is out of character for that child e.g. swearing, rudeness
- Irritability
- Aggression
- Apathy – loss of interest in life
- Difficulty planning or organising
- Weakness of one side of the face or body
- Problems walking
- Difficulty speaking

### **Symptoms of brain tumours affecting the parietal lobe**

The parietal lobe contains areas responsible for the sensation of touch and association, this area enables fine judgement of sensation such as texture, weight, size. Symptoms of tumours in this area include:

- Loss of sensation in part of the body.
- Sensory or motor neglect – e.g. a child with right sided sensory neglect will not respond to a sound from the right, gesturing by someone standing on their right or a touch to the right side of the body. They will respond normally to these stimuli on the left side.
- Difficulty speaking or understanding speech.
- Problems with reading and/or writing.

### **Symptoms of brain tumours affecting the temporal lobe**

The temporal lobe has many functions including processing of audio and visual information, comprehension and memory of verbal information. Symptoms of tumours in this area include:

- Fits – these may be called ‘temporal lobe epilepsy’ this type of epilepsy is often associated with strange feelings, smells or

déjà vu sensations which accompany or precede a fit (often termed an 'aura')

- Short term memory problems
- Inability to recall words

### **Symptoms of brain tumours affecting the occipital lobe**

This area is responsible for vision; symptoms include problems with or loss of vision on one side. Blindness that is caused by damage to the visual area of the brain is called 'central' or 'cortical' blindness.

### **Symptoms of brain tumours affecting the cerebellum**

The cerebellum helps to coordinate balance, symptoms of tumours affecting the cerebellum include:

- Problems with balance and coordination.
- Intention tremor.
- Squint/ double vision.
- Abnormal eye movements – 'nystagmus'.
- Nausea and dizziness.
- Raised intracranial pressure.

### **Symptoms of brain tumours affecting the pituitary gland**

The pituitary gland is a small gland arising from the bottom of the hypothalamus at the base of the brain. This pea sized gland sits inside a bony cavity known as the sella turcica close to the optic chiasm (crossover of the optic nerves). It produces many hormones that control and regulate body processes such as growth, puberty, adrenal and thyroid functions. Symptoms of these tumours may arise as a result of abnormal hormone production – too much in 'functioning' tumours or too little in 'non-functioning' tumours. They may also compress important parts of the brain located next to the pituitary gland (e.g. optic nerves or hypothalamus). In children, tumours arising from either the pituitary or nearby structures are often termed supra or parasellar tumours and include; craniopharyngioma, germinomas and

astrocytomas of the optic chiasm or hypothalamus and present with similar symptoms and signs as discussed below.

Symptoms may include:

- Arrested growth
- Excessive growth
- Precocious puberty
- Delayed puberty
- Menstrual irregularity in girls
- Weight gain (morbid obesity)
- Failure to thrive
- Mood swings (behavioural change)
- High blood pressure
- Diabetes
- Loss or restriction of visual fields and acuity
- Excessive thirst and urine production (diabetes insipidus)
- Infertility in the longer term
- Hypothyroidism (lethargy, cold intolerance, weight gain)
- Hypoadrenalism (underactivity of the adrenal glands)
- Raised intracranial pressure

see: [Care and mobility considerations](#)

see: [Indicators of severe functional restriction](#)

### **How is it assessed?**

When the brain is affected by a tumour it functions abnormally and causes symptoms as described. Signs of damage can often be found on neurological examination. This is a detailed examination of nerve function in the body and can sometimes give a picture of likely pathology. Investigations used

will depend on the cause of the problem and can include either alone or in combination:

- Computed Tomography (CT) scan
- Magnetic Resonance Imaging (MRI) scan
- Blood tests – include tests to measure hormone function if a pituitary tumour is suspected and tumour markers to test for germ cell tumours
- Lumbar puncture - assesses Cytology (appearance of cells)
- Electroencephalography (EEG) - recording of electrical activity from the scalp to investigate seizure disorders (fits)
- Ophthalmology tests - acuity, colour, visual fields
- Audiology tests - assess hearing

Biopsy – this will be performed under general anaesthetic and may be ‘open’ under direct vision by the neurosurgeon usually as part of therapeutic resection (removal of tumour) or ‘stereotactically’ by using a needle directed under MRI or CT guidance. For midline tumours the surgeon may use a Neuroendoscope – a thin flexible tube with a camera which can navigate through and examine the cerebrospinal fluid (CSF) spaces in and around the brain.

see: [Medical terms relevant to disability assessment](#)

see: [Medical terms describing cognitive problems](#)

### **How is it treated and managed?**

Most children will have surgery for their brain tumour as this is the most effective treatment to relieve selected symptoms at short notice and is a significant prognostic factor in the management of several tumours. Surgery, with the aim to remove the entire visible tumour may not be possible due to the location and proximity to functionally important or vital areas of the brain (e.g. pons - responsible for breathing or motor strip in the fronto-parietal lobes of dominant hemisphere). In these cases, either a biopsy or partial ‘debulking’ will be performed and adjuvant (follow-up) therapy with either radiotherapy, chemotherapy or a combination of both may be used. For some tumour types (e.g. low-grade astrocytoma or other benign tumours) complete surgical resection is the only treatment necessary. However, for malignant brain tumours (e.g. medulloblastoma, malignant glioma) and some lower grade tumours (e.g. ependymoma) adjuvant therapy will be

recommended for the majority of cases even if a complete resection is achieved because of concerns about residual microscopic disease.

In young children (under 3 years of age) the management depends not only on the tumour type, location and stage but also possible late effects of treatment, particularly radiotherapy induced effects on brain development. The immature developing brain is very sensitive to radiotherapy and this can result in significant and severe ongoing learning difficulties, growth problems and endocrine deficits. The younger the child is, the worse the outcome. The functional effects are manifested over a prolonged period (years) as the child's ability to learn is reduced compared to their peers, which means that the gap in their abilities compared to the norm widens over time. This means that children under 3 who have received radiotherapy have a significantly lower IQ after treatment and frequently have impaired fine motor, visuo-motor and visuo-spatial skills and memory difficulties. This is not restricted to those below 3 years of age, but it is more pronounced in the younger age group. Over time radiotherapy treated children are likely to require special help at school because of their cognitive impairments. Young children, therefore, often have more chemotherapy-based adjuvant therapy to either, avoid, delay or reduce the need for radiotherapy.

## **Surgery**

### **Craniotomy**

This is the main operation used for most brain tumours - a large incision is made, and a flap of bone lifted from the skull to gain access to the brain underneath. It is performed under general anaesthesia in children. Initial recovery takes place in hospital with the hospital stay ranging from 5-14 days. The scars tend to be behind the hairline and do not affect appearance in the



longer term. Return to normal function after this type of surgery depends on several factors:

- Level of function before surgery
- Whether further damage was caused by surgery
- Whether neurorehabilitation (medical processes to aid recovery from nervous system injury and to compensate for functional alterations) is required
- Whether further treatment such as radiotherapy or chemotherapy is required

Common problems after surgery include:

- Headaches – these normally resolve within two weeks but may persist for months
- Lack of concentration and memory problems – can gradually improve over several months
- Tiredness – likely to be severe for the first 2 weeks and then improve but does not always completely resolve and is made worse by adjuvant treatment

A child who was well prior to their surgery and who does not need any neurorehabilitation, or any further adjuvant treatment may be well enough to return to school 6 -12 weeks after surgery.

A child with a neurological deficit such as weakness, clumsiness, memory problems or difficulty with speech should have access to neurorehabilitation. For mild problems this can be offered as an outpatient with regular therapy sessions including Physiotherapy, Occupational therapy and Speech and Language therapy. This may be at a local hospital or via community teams and therapists. For more severe deficits this may involve a period as an inpatient on a neurorehabilitation ward.

Neurological deficits are likely to improve quickly for a few months and then improve much more slowly, usually improving for up to 2 years. This can vary and can be assessed by therapists who can monitor patients' recovery at key stages of their pathway and offer appropriate intervention. Needs will decrease as improvement occurs. Some neurological deficits will not improve with rehabilitation and for some children a neurological deficit may persist. Part of the rehabilitation process will include learning compensatory and coping skills to overcome disabling effects of both physical and cognitive impairment and to optimise function. This may include the use of

adaptive devices or equipment to enable activities of daily living. A stable condition is likely to be reached in two years.

Other types of surgery:

Shunts - thin flexible plastic tubes are implanted into the body to redirect cerebrospinal fluid (CSF) when a blockage has occurred – they relieve raised intracranial pressure and its symptoms. Shunts may be temporary or permanent. Recovery from this type of surgery takes up to a few weeks; it is not associated with any permanent neurological deficits but can be complicated by shunt blockage and infection requiring further treatment and possible revision of the original shunt.

Neuroendoscopy - this technique uses an endoscope to operate through, this means only one or two small 'burr' holes have to be made in the skull and recovery is quicker. Unfortunately, at the moment, it can only be used for a small proportion of brain tumours. A 'third ventriculostomy' is an endoscopic operation to relieve raised intracranial pressure avoiding a plastic shunt.

Transphenoidal surgery - this means an operation performed via the nose so there are no external scars. This type of surgery is virtually exclusively used for pituitary gland tumours as the pituitary gland is located behind the nose. There are no external scars after this type of surgery – recovery time is up to 6 weeks.

Radiosurgery - this is not surgery at all but highly focused and targeted radiotherapy treatment.

## **Radiotherapy**

Radiotherapy is a commonly used treatment for brain tumours, it is used as:

- primary treatment for tumours that cannot be removed by surgery
- adjuvant treatment for tumours that have been either completely removed or 'debulked' by surgery either after a time of observation (less aggressive tumours) or at first diagnosis of a malignant tumour, or for treatment of recurrent disease.

Radiotherapy treatments are usually given daily (Monday to Friday) over 6 weeks. Side effects occur during treatment but are not usually severe. In young children (less than 5 years old) daily general anaesthesia may be needed to deliver the radiotherapy. Long term side effects are significant in

children especially if they are under 3. If possible, radiotherapy treatment is delayed until the child is older.

Short term side effects include:

- Tiredness and sleepiness – if severe, towards the end of treatment the child having radiotherapy may do little other than sleep. In medical evidence this may be referred to as ‘somnolence syndrome’. This can occur after the radiotherapy has been completed and persist up to 12 weeks after treatment has finished but usually improves without intervention.
- Sickness – may require treatment with anti-emetic drugs.
- Worsening neurological deficit – problems caused by the tumour, such as clumsiness or speech difficulty, may get worse during treatment because of swelling within the treatment area. Steroid drugs are often used to treat this. Deficits associated with swelling should improve once treatment is complete.
- Hair loss and skin changes – hair in the treated area is likely to fall out and this may be permanent depending on dose and the individual. There may be skin changes similar to mild sunburn - the skin may become very sore, red and dry.
- Disturbed hearing – hearing may be affected during treatment due to local inflammation, which may persist if the cochlea is within the radiotherapy field.
- Difficulty concentrating and remembering – in children under 7 the IQ is reduced after radiotherapy treatment, the younger the child the greater the reduction. This is rare in the short-term setting and usually manifests 6 months or later after treatment.

In the long-term mild-moderate problems with concentration and memory are common in children who have had radiotherapy treatment to the head (depends on age, dose and volume of brain irradiated), they are not necessarily disabling but they may mean that a child does not fulfil their intellectual potential.

Conformal radiotherapy is a technique for giving radiotherapy that reduces the dose of radiotherapy to normal tissues so reducing associated damage and long-term side effects. It not used very often in children.

### **Proton therapy**

Proton treatment is a highly specialised means of delivering radiotherapy dose to a target volume. The particular characteristic of proton beams means that comparatively little radiation is given to normal tissues. This is especially useful for children with certain types of brain and spinal cord tumours. This is a new technique and long-term outcomes are still limited. There are currently no Proton facilities in the UK capable of treating brain tumours. In May 2007, the final report of the National Radiotherapy Advisory

Group (NRAG) contained a recommendation that the Department of Health facilitated the setting up of panel to review high priority clinical cases on behalf of Primary Care Trusts (PCTs). Since April 2008, suitable cases have been funded for treatment abroad by a service nationally commissioned by the National Commissioning Group (NCG.) A Proton Therapy Clinical Reference Panel (PTCRP) has been set up, which manages the referral pathway and has the necessary knowledge of proton therapy to approve cases for consideration for treatment abroad. Local clinicians following discussions with patients and their families may choose to refer to the panel for consideration of Proton therapy rather than conventional radiotherapy.

For further details of see - [Proton Beam Therapy](#)

### **Steroid drugs**

Steroid drugs are commonly used in short courses to reduce brain swelling related to the tumour itself or its treatment, particularly with surgery or radiotherapy – both of which cause tissue inflammation and swelling. Prolonged use of steroid drugs will have side effects; they cause weight gain, osteoporosis, muscle weakness, behavioural changes and growth retardation.

### **Anticonvulsants**

Epileptic fits are common symptoms of brain tumours, but they may also occur as a result of treatment such as radiotherapy or surgery. These drugs are commonly used prophylactically to prevent fits in children undergoing treatment whether they have had a fit because of their tumour or not. Children who have had fits may need to take anticonvulsants permanently to control epilepsy. Disabling effects of epilepsy relate to frequency and type of fits – refer to [epilepsy](#) guidance.

### **Chemotherapy**

Chemotherapy, unlike in adults with CNS tumours, may be the primary treatment of choice for some patients e.g. unresectable low-grade astrocytoma (glioma- NB. this term is often used if no histological diagnosis), and malignant tumours in young children. It is also frequently used in combination with radiotherapy.

Chemotherapy can occasionally be delivered directly into the tumour by placing a drug impregnated implant or wafer into the tumour bed at the time of surgery. This has only been used in clinical trials in 2010. Conventionally it is given in cycles using oral or intravenous combinations of chemotherapy

drugs. Most chemotherapy for brain tumours is given either intravenously or by mouth (orally). Drugs likely to be used include:

- Cisplatin.
- Carboplatin.
- Cyclophosphamide.
- Etoposide.
- Vincristine.
- Methotrexate.
- Temozolomide.
- Nitrosoureas.

In young children to avoid radiotherapy or in poor prognostic tumours 'high dose chemotherapy' may also be used. Trials are underway testing the outcome of high dose chemotherapy with stem cell transplant in children under 4 and for high risk PNET (Primitive Neuroectodermal Tumour). This

treatment involves high dose chemotherapy followed by peripheral blood stem cell transplant (PBSCT).

**What evidence is available?**

Sources of Evidence	Information Provided	Limitation of Evidence
Claimant/carer	First source of information	May not be objective, does not have specialist knowledge.
GPFR	<p>The GP will be able to confirm that a brain tumour has been diagnosed.</p> <p>Even once intensive treatment is complete all children are followed up in a 'late effects' clinic. The child may be seeing other specialists about their problems; the GP should be able to provide a broad overview of all the issues affecting the child but may not see the child themselves.</p>	<p>At the time of diagnosis, the GP may not be able to provide a histological diagnosis or details of the proposed treatment particularly the duration of treatment and recovery which may be several years in the under 3s. Following treatment, the GP may not see the child regularly as they will continue hospital follow up in the late effects clinic.</p>

<p>Hospital FR – best source of evidence</p> <ul style="list-style-type: none"> <li>• Clinical Nurse Specialist</li> <li>• Treating Consultant</li> <li>• Paediatric Oncologist</li> </ul>	<p>During diagnosis and the initial stages of treatment, the hospital consultant and the specialist nurse will be able to provide the most up to date information on diagnosis, treatment plan, current disabling effects and prognosis of disabling effects.</p> <p>Once treatment is complete all surviving children will be followed up in a 'late effects' clinic. The consultant or other specialist from this clinic is likely to be able to provide the most complete and up to date assessment of residual disabling effects such as cognitive impairment or learning difficulties.</p>	<p>Once the child has finished active treatment the original specialist may not be up to date with the child's progress. The late effects clinic will be the most appropriate source of evidence if for example; the child is diagnosed with learning difficulties and provided with a SEN after radiotherapy to the head.</p>
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**How long will the needs last?**

Primary brain tumours

Recurrent brain tumours

Types of brain tumour & prognosis information

This guidance covers -:



Tumours – benign – other / type not known
Brain and spinal cord – cancer of

### **Primary brain tumours**

There are around 100 different types of brain tumour and prognosis is highly variable between them. For this reason, guidance on award duration is provided by tumour diagnosis in the table below. It is appropriate to use this guidance for children over 3, for children under 3 years treatment may be prolonged because a long course of chemotherapy is being given to delay radiotherapy treatment. In cases where needs are present related to disease or its treatment in the under 3s it may be most appropriate to make the initial award duration until the 4<sup>th</sup> birthday to give them time to recover from their prolonged treatment. Medical evidence will often include a proposed plan of treatment on which award duration can be based.

In order to use the table, the correct histological diagnosis must be known. This information is most likely to be available from the Clinical Nurse Specialist, treating neurosurgeon, treating neuro-oncologist or the GP. Accurate information on impairment is most likely to be available from the Consultant or Clinical Nurse Specialist.

For many tumours, awards are recommended for two years in the presence of neurological deficits if needs are identified. This is because function may improve significantly over time especially with neurorehabilitation. For example, a child may learn how to walk again, or their personality may substantially return to normal. Review at two years to assess residual impairment once neurorehabilitation is complete. Once recovery is complete

needs may be absent or reduced. In cases where disease progresses despite treatment needs are likely to increase.

### **Recurrent brain tumours**

If needs are identified indefinite awards are recommended.

### **Types of brain tumour & prognosis information**

Name of brain tumour	Features
<p><b>Astrocytic tumours</b></p> <p>There are many types of astrocytoma.</p>	
<p>Pilocytic astrocytoma (WHO grade 1) and subtypes:</p> <ul style="list-style-type: none"> <li>• Pilomyxoid astrocytoma (WHO grade 2)</li> <li>• Pleomorphic xanthoastrocytoma (WHO grade 2)</li> <li>• Subependymal giant cell astrocytoma (WHO grade 1)</li> </ul>	<p>This is a slow growing type of tumour that has a good prognosis; it is often curable. The main treatment is surgery very occasionally followed by radiation therapy. Typically occur in the optic nerve, optic chiasm/hypothalamus, thalamus, basal ganglia, cerebral hemispheres, brain stem, spinal cord. 80% of astrocytomas occurring in the cerebellum are this type and are usually curable by surgery – may also be called Gilles Type A in the medical evidence. If needs are identified, a 2-year award is recommended. Recovery is expected in the typical case. Note that visual field defects are not likely to improve. Enduring side effects related to radiotherapy treatment in rare cases where this has been used. This is the commonest type of brain tumour in children and 10-year survival is around 90%.</p>

<p>Diffuse astrocytoma/low-grade diffuse astrocytoma (WHO grade 2) including 3 different subtypes:</p> <ul style="list-style-type: none"> <li>• Gemistocytic astrocytoma</li> <li>• Protoplasmic astrocytoma</li> <li>• Fibrillary astrocytoma</li> </ul>	<p>These are slow growing tumours that have a good prognosis. Survival at 4 years is 90%. Older children do better than younger children. The main treatment is surgery occasionally followed by radiation therapy. If needs are identified a 2-year award is recommended. If needs persist on renewal, an indefinite award is recommended. Diffuse tumours Gilles Type B astrocytomas have a worse prognosis. Recovery is expected in the typical case. Note that visual field defects are not likely to improve. Enduring side effects related to radiotherapy treatment in rare cases where this has been used.</p>
<p>Anaplastic or malignant astrocytoma (WHO grade 3)</p>	<p>These tumours have a poor prognosis with survival at best 40% and on average 19% at 5 years. The main treatment is surgery followed by radiation therapy. Younger children are likely to have chemotherapy. Children whose initial treatment successfully removes the entire tumour are most likely to do well (40% 5-year survival); although removal of the entire tumour with clear margins is unusual. If needs are identified, a 2-year award is recommended. If needs persist on renewal, an indefinite award is recommended.</p>

<p>Glioblastoma/ malignant glioma/glioblastoma multiforme (WHO grade 4)</p>	<p>These tumours have a poor prognosis with survival at best 40% and on average 19% at 5 years. The main treatment is surgery followed by radiation therapy. Younger children are likely to have chemotherapy. Children whose initial treatment successfully removes the entire tumour are most likely to do well (40% 5-year survival); although removal of the entire tumour with clear margins is unusual. If needs are identified, a 2-year award is recommended. If needs persist on renewal, an indefinite award is recommended.</p>
<p>Pleomorphic xanthoastrocytoma (WHO grade 2)</p>	<p>Is very rare it usually affects young adults. Recurrence free survival rates of 71% at 5 years and 61% at 10 years are reported adults. If needs are identified, a 2-year award is recommended.</p> <p>Recovery can be expected in the typical case.</p>

<p>Subependymal giant cell astrocytoma (SEGA) (WHO grade 1)</p>	<p>Very slow growing treatable tumour that mainly affects children with tuberous sclerosis (a rare genetic disorder). If needs are identified, a 2-year award is recommended.</p> <p>Recovery is expected in the typical case, however there may be ongoing needs unrelated to the brain tumour in this group.</p>
<p><b>Brain Stem Gliomas</b> – the brain stem is the most primitive part of the brain it consists of the following structures: the pons, the midbrain, the tectum, the medulla.</p>	
<p>Diffuse Intrinsic Pontine Glioma</p>	<p>Surgical removal is not usually possible; radiotherapy to the head is the main treatment. Survival is poor at less than 10% at 18 months from diagnosis. If needs are identified, a 5-year award is recommended.</p>
<p>Focal or low-grade brain stem gliomas, this group includes pilocytic astrocytomas.</p>	<p>Surgery is the main treatment of these tumours. Long term survival is seen even if residual tumour is not removed. A shunt may be required to treat hydrocephalus. Tumours that progress may be treated with conformal radiotherapy or chemotherapy or both. Prognosis is good. If needs are identified, a 2-year award is recommended.</p>

<p>Neurofibromatosis and brain tumours associated with it</p>	<p>Children with neurofibromatosis type 1 commonly develop slow growing brain tumours that may not require treatment or cause symptoms. Consider in each case whether there are disabling effects of the tumour and what treatment, if any, is being used.</p>
<p><b>Embryonal and Primitive Neuroectodermal tumours (PNETs)</b></p>	
<p>Medulloblastoma subtypes:</p> <ul style="list-style-type: none"> <li>• Desmoplastic medulloblastoma</li> <li>• Large cell medulloblastoma</li> <li>• Medullomyoblastoma</li> <li>• Melanocytic medulloblastoma</li> </ul>	<p>Arise in the posterior fossa. May spread via the cerebrospinal fluid into the spinal cord. Children under 3 are in the high-risk group. The main treatment is surgery; enduring neurological defects after surgery are common: mutism, suprabulbar palsy (difficulty feeding and speaking) ataxia (difficulty walking). Surgery is followed by radiotherapy with or without chemotherapy. If needs are identified, a 5-year award is recommended.</p> <p>5-year survival ranges from 85% in standard risk children to 30% for high-risk children. High dose chemotherapy and stem cell transplant may be used in this group.</p>

<p>Supratentorial primitive neuroectodermal tumours includes:</p> <ul style="list-style-type: none"> <li>• Cerebral neuroblastoma</li> <li>• Ganglioneuroblastoma</li> <li>• Pineoblastoma</li> <li>• Pineal parenchymal tumour</li> </ul>	<p>Arise in the cerebrum and suprasellar region. Surgery is the main treatment but because of the location of these tumours, complete removal is rarely achieved. Radiotherapy treatment with or without chemotherapy is given after surgery. 5-year survival is 40 to 50%. High dose chemotherapy and stem cell transplant may be used in this group. 2-year awards are recommended in children aged 3 and over.</p>
<p>Medulloepithelioma and Ependymoblastoma</p>	<p>These are rare occurring most frequently in infants and younger children. 5-year survival is 0 to 30%. If needs are identified, a 5-year award is recommended.</p>
<p>Atypical Teratoid/Rhabdoid tumour (WHO grade 4)</p>	<p>Most common in children under 3, presents with hydrocephalus or regression of motor skills/ataxia. Treatment includes surgery, chemotherapy and often radiotherapy. Median survival is 12 months and 2-year survival is 14% in children who receive chemotherapy alone. Older children who receive radiotherapy median survival around 4 years. If needs are identified, a 5-year award is recommended.</p>



<p>CNS germ cell tumours includes:</p> <ul style="list-style-type: none"> <li>• Germinomas</li> <li>• Embryonal yolk sac tumours</li> <li>• Choriocarcinomas</li> <li>• Immature teratomas</li> <li>• Mature teratomas</li> <li>• Teratomas with malignant transformation</li> <li>• Mixed germ cell tumours</li> <li>• Nongerminomatous germ cell tumours</li> </ul>	<p>Primary treatment of germinomas is radiotherapy, this is usually curative. Other types are treated with chemotherapy. No survival data. If needs are identified, a 2-year award is recommended. If needs persist on renewal, an indefinite award is recommended.</p>
<p><b>Oligodendroglial tumours:</b> there are two types of this tumour see below to be treated with radiotherapy or chemotherapy and survival is good if response to treatment is complete.</p>	
<p>Oligodendroglioma (low grade, WHO grade 2))</p>	<p>Very rare in children. Good prognosis if completely excised with surgery. If needs are identified, a 2-year award is recommended. If needs persist on renewal, an indefinite award is recommended.</p>
<p>Anaplastic oligodendroglioma (WHO grade 3).</p>	<p>Poorly differentiated tumour with a worse outcome than low grade oligodendroglioma. If needs are identified, a 2-year award is recommended. If needs persist on renewal, an indefinite award is recommended.</p>
<p><b>Mixed gliomas</b></p>	

Oligoastrocytoma (low grade, WHO grade 2)	Usually occurs in the frontal or temporal lobe. If needs are identified, a 2-year award is recommended. If needs persist on renewal, an indefinite award is recommended.
Anaplastic Oligoastrocytoma (WHO grade 3)	These are high grade tumours with outcomes similar to other malignant astrocytomas. If needs are identified, a 2-year award is recommended. If needs persist on renewal, an indefinite award is recommended.
<b>Ependymoma</b> – subtypes account for 9% of childhood brain tumours	
Myxopapillary ependymoma (WHO grade 1)	This tumour commonly affects the lower end of the spinal cord – survival is excellent. Recovery is expected in the typical case. Surgery is the main treatment with local radiotherapy performed afterwards. If needs are identified, a 2-year award is recommended. If needs persist on renewal, an indefinite award is recommended.
Subependymoma (WHO grade 1)	Commonly arises in the tissue lining the ventricles of the brain and may present with hydrocephalus. If needs are identified, a 2-year award is recommended.

<p>Ependymoma (WHO grade 2)</p> <p>Variants of this tumour are called:</p> <p>Cellular</p> <p>Papillary</p> <p>Tancytic</p> <p>Clear cell</p> <p>mixed</p>	<p>5-year survival is 78%. It is difficult to completely remove these tumours surgically (25%) radiotherapy is likely to be used in every case. Some children will also have chemotherapy. Can occur in the spine and cause paraplegia that is permanent. Recovery from initial treatment is expected in the typical case. If needs are identified, a 2-year award is recommended. If needs persist on renewal, an indefinite award is recommended.</p>
<p>Anaplastic ependymoma also called malignant ependymoma (WHO grade 3)</p>	<p>No survival data. Survival is worst with this type of ependymoma. Treatment is surgery and radiotherapy. If needs are identified, a 2-year award is recommended. If needs persist on renewal, an indefinite award is recommended.</p>
<p><b>Meningioma</b></p>	
<p>Meningioma – these can be benign or highly malignant. Benign tumours will be WHO grade 1 and malignant tumours will be grade 4.</p>	<p>Account for 1-2% of brain tumours in children. In all cases outcome will depend on whether complete surgical removal is achieved. Radiotherapy may be used; chemotherapy treatment may be used. If needs are identified, a 2-year award is recommended. If needs persist on renewal, an indefinite award is recommended.</p>
<p><b>Pituitary tumours</b></p>	

Benign adenomas of the pituitary gland	Around 80% of this group experience resolution of symptoms with treatment. Needs unlikely, recovery expected. Good long-term prognosis. Rarely visual field problems will persist after treatment. May require hormone replacement.
Carcinomas of the pituitary gland.	Median survival of around 2 years. If needs are identified, a 5-year award is recommended.

<p>Craniopharyngoma</p> <p>Subtypes called:</p> <p>Adamantinomous</p> <p>Squamous papillary</p>	<p>Rare – account for 6% of childhood brain tumours. These arise near the pituitary gland in the optic chiasm. Hormonal problems such as restricted growth or visual field defects are common. Hydrocephalus may occur. Long term survival is good at 79%. Main treatment is surgery. Almost all children will require lifelong pituitary hormone replacement therapy. Enduring complications of surgery include obesity, mood change, blindness, epilepsy, difficulty moving the eyes. Radiotherapy can be used following minimal surgery to decompress optic nerves; this can also lead to blindness, loss of pituitary function and difficulty with eye movements. Although these tumours are benign, complications of treatment are lifelong. If needs are identified, a 5-year award is recommended.</p>
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All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Peripheral Blood Stem Cell Transplant (PBSCT) or Bone Marrow Transplant**

A Peripheral Blood Stem Cell Transplant (PBSCT) or Bone Marrow Transplant is a way of transplanting a new immune system into a person's body.

The immune system consists of stem cells that make all the different kinds of blood cells that perform the functions of the immune system, these are -:

- Red blood cells – these carry oxygen around the body
- White blood cells – these cells travel around the body destroying bacteria, viruses and abnormal cells in the body such as cancer cells
- Platelets – these cells help the blood to clot

A transplant involves destroying the persons own immune system and stem cells and replacing them with either -:

- Their own previously harvested stock of stem cells or bone marrow – this is called an **autologous** transplant because a person's own cells are used.
- Someone else's stem cells or bone marrow – this is called an **allogeneic** transplant because someone else's cells are used.

The donor usually has the same or similar genetic markers to the recipient. Checking for compatibility between donor and recipient is called '**matching**'. The other person can be a close relative such as a brother or sister – a '**sibling donor**' or someone who is unrelated – this is called a '**matched unrelated donor**'. The name of the procedure depends on how the stem cells are collected from the donor although the principles of the procedures are the same and the effect on the recipient is the same:

### **Peripheral Blood Stem Cell Transplant (PBSCT)**

Peripheral blood stem cells are harvested from the blood. Growth factors are given to the donor to promote production of larger quantities than normal of stem cells for harvesting. These spill out into the peripheral blood from the bone marrow. The stem cells are filtered from the blood using a procedure similar to blood donation. No anaesthetic is required, and the process takes several hours. The side effects for the recipient are the same whether bone marrow or peripheral blood stem cells are used.

### **Bone Marrow Transplant**

Bone marrow is harvested from bone marrow in the pelvis during a short operation. The side effects of bone marrow harvesting are minimal – for example there may be soreness around the pelvis for a week or so afterwards.

The side effects for the recipient are similar whether bone marrow or peripheral blood stem cells are used.

### **How is a transplant given?**

In preparation for transplant a person's immune system will be wiped out by high dose chemotherapy and sometimes radiotherapy treatment. This often has the effect of killing off residual cancer cells and is the aim of the treatment. Once this treatment is complete the bone marrow or stem cells can be given via an intravenous drip. The stem cells do not start working straight away and can take up to a year to start working effectively and really protecting a person from infection.

The side effects of such high dose treatment are severe. A person is not able to produce their own red, white blood cells or platelets and the transplanted tissue is unable to do this for some time either. This means they need regular support with blood and platelet transfusions to prevent bleeding and control anaemia. Transfusions of white cells to control infection cannot be given and so despite antibiotics they are at very high risk of life-threatening infection with normally innocuous bacteria or viruses. When a person is unable to produce their own white blood cells to fight infection this is called 'neutropenia'. The high-risk period after transplant when a person is very vulnerable to infection is called the '**neutropenic period**' – the neutropenic period is usually spent in hospital.

Once some improvement has occurred and the immune system has started to work to a degree they will be discharged from hospital. The person undergoing treatment is likely to feel extremely ill and weak and may sleep for most of the time. They will have many of the side effects of chemotherapy.

Often the first phase of treatment will be given in isolation in hospital – isolation means just that; a person is kept in one room with minimal visitors who all wear barrier clothing to prevent the transfer of infection. The psychological effects of isolation can be severe. The time spent in isolation in the recent past was 1-2 months. This time has been reduced with more of the recovery time spent at home. The same precautions may be necessary at home as would be taken in an isolation room in hospital. These may be troublesome for carers in terms of cleanliness of the home, providing safe food, preventing contact with potentially harmful everyday items and restricting access to people who are themselves unwell. In addition, frequent blood tests are needed to monitor progress, and this is likely to involve accompanying their family member to hospital and back.

### **Side effects of chemotherapy**

Side effects of chemotherapy are well known and are mostly related to the effect of chemotherapy drugs on normal dividing cells. Some cells in the body divide more rapidly than others because they have to constantly

replace themselves for the body to function normally. Blood cells are a good example of this. The bone marrow produces white blood cells, which fight infection. These cells are produced by stem cells, which divide frequently, and white blood cells only live for a short time, they can easily fall to low levels during chemotherapy treatment. This is important because when the white blood cells are low this allows the possibility of overwhelming and sometimes fatal infection. Having a low white count is called 'neutropenia'.

The cells, which line the bowel also rapidly divide and are sloughed off by the passage of food and liquid. When these are not replaced quickly enough there is ulceration of the mouth, throat and bowel. Ulceration lower down in the intestines can lead to bleeding, nausea, abdominal pain and diarrhoea. It is these effects on normal cells, which limit the amount of chemotherapy that can be given.

### **Main short term side effects during Chemotherapy**

- Fatigue - is the main physical side effect of treatment with chemotherapy. In the days after treatment, it is usual for the patient to feel like they are developing a flu-like illness. The acute effects will resolve but feelings of tiredness may persist throughout the treatment cycle and can be very debilitating
- Hair loss - is common and may cause significant distress
- Nausea and Vomiting - the majority of chemotherapeutic drugs cause nausea and vomiting of varying degrees. The treatment of this has dramatically improved in recent years and it can usually be effectively managed with anti-emetic drugs
- Psychological effects - because the physical symptoms are better controlled, the emotional side effects of having cancer and receiving chemotherapy have become more important. It is impossible to separate out the side effects of the treatment and the side effects of having cancer. General tiredness, the strain of being diagnosed with cancer, undergoing tiring treatment and the burdens this imposes on the family are all important. Worries about the future are common

Another common cause of anxiety about chemotherapy in the past was fear of needles and the discomfort of treatment; this has been addressed by the



use of local anaesthetic creams before putting needles in and to an extent by the increased use of indwelling ports and catheters.

### **Other commonly reported short term side effects**

- Giddiness on standing up
- Diarrhoea
- Weight gain
- Shortness of breath

Short term side effects resolve once treatment is complete.

### **Long-term disabling effects of chemotherapy treatment**

Peripheral neuropathy (neuritis, plexopathy) - is a toxic effect on nerves, which prevents nerves from working properly.

- Motor nerves – these control movement of muscles; damage may lead to clumsiness or in severe cases paralysis of muscles supplied by the affected nerves. Some recovery may occur once treatment is stopped but the changes are usually slow to improve or permanent
- Sensory nerves – these nerves enable the sensation of touch, damage to them results in numb areas or areas of pins and needles. In some cases, pain fibres are affected, and this can lead to pain syndromes

Abnormal sensations can make simple activities like making a cup of tea, fastening a button or walking difficult, impossible or painful and distressing.

- Breathlessness - has many causes including lung problems and heart failure, which may have been caused by

chemotherapy or radiotherapy. Lung damage or heart problems caused by certain drugs may be irreversible or progressive

- Leukaemia - is more common in children who have had chemotherapy or radiotherapy treatment
- Infertility – chemotherapy and radiotherapy can cause infertility

These side effects are rare.

### **Biological therapy**

There are multiple options most of which will be used in the context of clinical trials. They include monoclonal antibodies (e.g. Avastin™), dendritic cell vaccination, tyrosine kinase inhibitors e.g. Glivec (Imatinib), gene therapy and oncolytic viruses. Side effects of these are generally milder compared to the effects of conventional chemotherapy but targeted therapies affecting developmental pathways may possibly result in side effects that could only be seen in children e.g. growth effects.

### **Treatment of recurrent brain tumours**

Often recurrent brain tumours cannot be cured but they can be controlled for a period of time, depending on the tumour type and location of the recurrence. Children may be offered additional chemotherapy, (radiotherapy if not already received), enrolment onto a clinical trial for a new treatment or symptomatic palliative care.

### **Problems in adults who had cancer treatment as children**

People in this category have much longer to develop the long term or enduring side effects of chemo and radiotherapy. The oldest members of this group will have had their treatment in the 1970s. What will happen to them as they age is unknown. Some childhood survivors have already developed significant enduring problems because of their treatment, either during treatment or some years later. The number of adults 'at risk' in this category is set to rise.

Cancer therapy, in particular chemotherapy made great progress in the 1980s and 1990s and for the first-time high rates of cure were achieved in some of the common childhood malignancies such as leukaemia and lymphoma. Over time treatment has been modified to become as effective as possible with as few side effects as possible. Significant long term side effects of treatment given in the past are increasingly being recognised. These side effects generally occur because of changes in normal tissue

caused by the treatment, these changes take many years to cause symptoms or become apparent. The medical profession is still in the early days of recognising and researching these disorders.

Over time more members of this group can expect to either develop these problems or have them recognised. A breakdown of problems is provided by treatment. Effects tend to be greater when treatment of cancer began at a young age (under 3) and when large doses of chemotherapy and

radiotherapy were necessary. Common cancers in children include leukaemia, lymphoma, brain tumours, bone and soft tissue sarcomas.

Type of cancer treatment	Disabling effect
Cranial irradiation esp. if combined with intrathecal chemotherapy	Neurocognitive defects – reduced IQ, attention deficit, poorer motor/verbal skills, may be severe enough for a Statement of Education Needs (SEN), deafness, epilepsy.
Cranial irradiation, effects are worse if this was combined with abdominal radiation	Hormonal effects including growth impairment in childhood, hypothyroidism, increased risk of infertility, early menopause.
Cranial irradiation	Obesity and its disabling effects.
Chemotherapy or radiotherapy	Increased risk of infertility.
Chemotherapy esp. anthracycline doxorubicin	Heart problems including heart failure, myocardial infarction, rhythmias and sudden death at young age.
Radiotherapy to chest	Lung problems – breathlessness.
Steroids, methotrexate, inactivity due to illness	Osteopaenia/Osteoporosis.
Chemotherapy or radiotherapy	Second cancers especially brain tumour.
Radiotherapy to abdomen (bladder/bowel/liver)	Chronic diarrhoea, malabsorption, bladder problems, kidney problems including rarely kidney failure.

## Care and mobility considerations

### Primary tumours

Needs may arise from either the:

- Effect of the tumour
- Effects of treatment

Needs are likely to arise because of neurological impairments caused or exacerbated by either the tumour or its treatment. Most primary tumours will be treated with surgery or surgery followed by radiotherapy in older children or surgery followed by chemotherapy and then by radiotherapy some years later in children under 3. When radiotherapy cannot be delayed in the under 3s the side effects of the treatment may be significant and enduring. The primary tumour may be completely or partially removed. Most children will have had a craniotomy – the recovery from this type of surgery is described under treatment. In an uncomplicated case, where there was no or minimal neurological impairment before surgery, recovery would be expected to take up to 3 months.

If a neurological impairment is present on diagnosis, the impairment may get temporarily worse because of the treatment. Recovery may take much longer than 3 months. Some improvement in impairment is likely with recovery and will be aided by neurorehabilitation (medical processes to aid recovery from nervous system injury and to compensate for functional alterations).

## **Care**

Care needs may arise due to physical or behavioural neurological impairments.

Physical problems may include problems with limb or trunk movement ranging from clumsiness/unsteady balance to paralysis. There may be paralysis or loss of sensation on one side of the body (hemiplegia), similar to a person who has had a stroke. If the upper limbs are affected, help may be required with activities of daily living. Severe problems with balance and weakness are likely to make aspects of self-care, particularly dressing, difficult. Preparing or carrying hot food or drinks with balance problems is potentially dangerous. Sudden onset of visual impairment, in addition to the other symptoms of a brain tumour are, likely to create or exacerbate care needs. Fits are a common symptom and supervision may be required until fits can be controlled with appropriate medical treatment.

Behavioural problems may include reduced or absent sense of danger as well as inappropriate or distressing behaviour. Patients often lack motivation and planning strategies for daily activities. Short term memory loss is a frequent feature of brain tumours and their treatment. When behavioural problems are present regular supervision will be necessary. Symptoms may get worse during treatment but may improve afterwards over several months.

Improvement may continue gradually over several years but usually plateaus after maximal rehabilitation.

## **Mobility**

Mobility may be affected in several ways by neurological impairments:

- Hemiplegia - loss of movement / clumsiness to either side of the body
- Altered sensation to either side of the body
- Perceptual neglect of one side of the body
- Difficulties with balance
- Poor concentration
- Central sensory deficits such as blindness or visual field defects and hearing problems
- Behavioural problems

Children with weakness, sensory problems and balance problems may have difficulty walking. They may require assistance or assistive devices and/or equipment to enable safe mobility and independence. Severe problems with balance may also make walking difficult or dangerous even though they have normal strength and movement in their legs. Children with sensory problems may require guidance and supervision if their deficit is severe.

Children with behavioural or cognitive problems may require guidance and supervision because of one of the following:

- Loss of awareness of danger
- Memory loss
- Inappropriate behaviour

Children with difficult to control or uncontrolled epilepsy may require guidance and supervision in both the home and unfamiliar places. 30% of people with brain tumours do not achieve complete control of their epilepsy.

Symptoms may get worse during treatment and can improve afterwards over several months. Improvement may continue gradually over several

years but not always completely resolve, particularly memory loss, which can actually continuously worsen following completion of primary treatment.

### **Recurrent brain tumours**

Brain tumours usually recur because they are either highly malignant, or they were in an inaccessible area of the brain and could not be completely removed. Further treatment is likely to be able to control symptoms and slow further progression down; but impairments are less likely to improve. If needs are identified because of neurological or cognitive impairment in recurrent brain tumour, indefinite awards are recommended.

### **Indicators of severe functional restriction**

Enduring disabling side effects are particularly common in:

- Children aged under 3, who have had radiotherapy to the head. Disability may become more, not less apparent as they get older
- Craniopharyngioma treatment is commonly associated with significant enduring disabling effects
- Spinal cord tumours

5-year awards are recommended if the following tumour types are diagnosed:

- Diffuse Intrinsic Pontine Glioma
- Medulloepithelioma
- Ependyoblastoma
- Atypical Teratoid/Rhabdoid tumour

### **World Health Organisation (WHO) classification of brain tumours**

Unlike other cancers where grade can vary depending on how aggressive cancer cells look under the microscope, most brain tumours can be given a grade based on the diagnosis. So, for example, Pilocytic astrocytoma is always a grade I and a Glioblastoma is always grade IV. The exception to this rule is Meningiomas – they can be grade I, II or III. The grade of tumour correlates with tumour behaviour. Grade I tumours are more benign in their

nature and can on occasion be cured by surgery; grade IV tumours are most commonly highly malignant and fatal.

Grade I – includes tumours that grow slowly and are often removable and curable with surgery.

Grade II – includes tumours that tend to spread and recur more after treatment than grade I tumours. Most of these tumours will in due course transform into high grade brain tumours with a poor prognosis.

Grade III – these tumours behave quite aggressively and tend to spread into surrounding brain tissue quite easily.

Grade IV – these are the most aggressive malignant tumours; they often progress rapidly and have a poor prognosis.

### **Medical terms relevant to disability assessment**

Neurological deficit – an abnormal physical finding such as paralysis or loss of sensation.

Hemiparesis – paralysis of one side of the body e.g. an arm and a leg on the same side. Dense hemiparesis or hemiplegia means no movement at all on the affected side - such a person would definitely not be able to walk unaided.

Dysarthria – a muscle/ nerve weakness causing difficulty speaking or making themselves understood – slurred or slow speech.

Dysphasia – a complex disorder of language processing; both understanding, and expression can be affected differently in the same person; language breakdown can be at any level; may include difficulties with reading, writing and numeracy.

Aphasia – an inability to understand or to speak at all.

Cranial nerve palsy – paralysis of one of the 12 cranial nerves that arise directly from the brain and control many motor, sensory and autonomic functions such as; vision (optic nerve), eye movements including the pupil, facial



movements and sensation (facial and trigeminal nerves), swallow, taste and speech.

## **Medical terms describing cognitive problems**

Common difficulties include problems with:

### **Attention**

Allows information into the brain and underlies all other cognitive processes. The pre-requisites are alertness and arousal. The problems observed are that information is not properly processed by the brain resulting in reduced understanding or misuse of information. The person may be impulsive, show perseveration, lack of insight, denial or be too concrete or literal. They might have difficulty comprehending, be withdrawn, frustrated or aggressive.

Assessing for attention problems

The occupational therapist can use standardised assessments such as Test of Everyday Attention or Digit Span, a rating scale questionnaire and functional observations of everyday activities such as making a hot drink, dressing self.

### **Executive functions**

Allow us to accomplish goal directed behaviour, to co-ordinate and control our thinking and behaviour, to self-monitor, show initiation, ability to plan and to set goals and process information. The problems observed include difficulty planning and executing tasks, difficulty initiating appropriate behaviour, difficulty with problem-solving and inability to identify strengths / weaknesses.

Assessing Executive Functions

The occupational therapist can use any functional activity or may use a standardised test such as 'Behavioural Assessment of dysexecutive Syndrome' (BADs), to identify areas which are often difficult to pick up in everyday conversation.

### **Information Processing**

This means the ability to make sense of the world by dealing effectively and efficiently with sensory and other information that constantly enters the system. It can occur at conscious or automatic level. Three components underlie conscious processing, these include speed (the amount of information a person can attend to within a given time), capacity (the amount of

information at any given time) and control (the ability to guide selective process by directing & organising whatever processing capacity he has.) Problems observed include difficulty following instructions, “delayed reaction” to instructions and overloading with information.

### Assessing for Information Processing Problems

The occupational therapist can use standardised assessments including speed reading of word lists and questionnaires.

### Memory

This includes the ability to keep things in the mind. It is a process involving attention (attending to information), encoding (registration of information), storage (stored in long term memory), consolidation (practising rehearsing information) and recall (retrieving information stored in long term memory). Two most functionally important aspects of memory are episodic memory (every day) and prospective memory (act in future).

### Assessing for Memory Problems

The occupational therapist uses questionnaires e.g. the Rivermead Behavioural Memory Test (RBMT) and digit span etc.

### **Visual Processing**

This includes acquisition of visual information and appropriate use and manipulation of that information. It includes oculomotor skill (ability to move eyes). Visual fields (the area that the eye can see at any one time (160-180 degrees), visual acuity (sharpness of eyesight), visual attention (ability to attend to critical features visually or staring), scanning (ability to record all details of a scene in an organised way), pattern recognition (features of an object e.g. shape, colour, etc), visual memory (visually process information,

store it and recall it) and visual cognition (mental manipulation of visual information).

## What you need to know about Cerebral Palsy

### What is Cerebral Palsy?

Cerebral Palsy (CP) is essentially a movement disorder. The term covers a group of conditions caused by injury to or abnormal development in the parts of the brain that control. ....

- [Cerebral Palsy](#)

### What are the effects and signs?

Children with CP may have a number of musculo-skeletal abnormalities that are often quite debilitating and .....

- [Effects of Cerebral Palsy](#)

### How is it assessed?

The brain lesions of Cerebral Palsy (CP) occur from the foetal or neonatal period to up to age 2 years.

- [Assessment of Cerebral Palsy](#)

### How is it treated and managed?

Cerebral Palsy (CP) cannot be cured but management aims to use appropriate combinations of interventions to

- [Treatment for Cerebral Palsy](#)

### What evidence is available?

Speech / language therapists give a profile of function in terms of language and communication skills, feeding skills and aspiration risks.....

- [Evidence](#)

### How long will the needs last?

- [Prognosis and duration of the award](#)

## What is Cerebral Palsy?

Cerebral Palsy (CP) is essentially a movement disorder. The term covers a group of conditions caused by injury to or abnormal development in the parts of the brain that control body movement and muscle coordination. The brain lesion often occurs during foetal development, but may also take place during, or shortly after birth; or during infancy up to 2 yrs old. The brain abnormalities do not get worse; however, the effects may change over time e.g. mobility may improve initially but then deteriorates later as the child gets heavier with development of contractures (permanent shortening of a muscle or tendon making it hard to stretch the area and preventing normal movement.)

### **Current official definition (2006)**

A major international consensus conference was held in 2006 when the definition and classification of CP was re-examined and agreed by experts. It is useful to note that from this conference the new definition of cerebral palsy is as follows -:

- (CP) describes a group of permanent disorders of the development of movement and posture, causing activity limitation, that are attributed to non-progressive disturbances that occurred in the developing foetal or infant brain.
- The motor disorders of cerebral palsy are often accompanied by disturbances of sensation, perception, cognition, communication, and behaviour, by epilepsy and by secondary musculo-skeletal problems.

### **Incidence & Prevalence**

(CP) is the most common physical disability in childhood. In the Western world, the condition occurs with a frequency of 2-2.5 of every 1000 live born children, but this is higher in premature and twin births. However, the greater majority of children with cerebral palsy are born at full term, as the

actual number of premature births is small in comparison with full term births.

see: [Causes](#)

### **What are the effects and signs?**

There are two main groups of problems in children with Cerebral Palsy (CP)  
-:

#### **Medical problems**

- Musculo-skeletal problems (further impairing mobility and posture)
- Oromotor difficulties (leading to feeding and nutritional problems and speech problems)
- Epilepsy

#### **Functional problems as a result of the medical impairments**

- Mobility problems
- Speech problems
- Vision and hearing problems
- Impaired cognitive functions
- Incontinence

### **Musculoskeletal Problems**

Children with CP may have a number of musculo-skeletal abnormalities that are often quite debilitating and cause further deterioration to their gross motor functioning -:

- Difficulty walking and maintaining balance resulting from spasticity and contractures
- Difficulty with fine motor tasks such as writing or doing up buttons
- Arm/leg weakness or both
- Scoliosis (abnormal curvature of the spine) is more common in patients with CP

- Various abnormalities of the foot and ankle such as flat feet or equinus (deformity of the ankle which results in child toe-walking)
- Degenerative hip disease and hip dislocation are common complications during the adolescent growth spurt, particularly in children with athetoid Cerebral Palsy. This may result in pain when sitting and standing

Note: Children that are able to walk by age 2 years are likely to continue to be able to walk. Children who do not sit by age 4 years don't usually ever walk.

### **Oromotor difficulties**

When the muscles of the face and throat are affected by CP there are multiple effects. Even in the absence of a learning difficulty, the child may have a significant communication problem due to problems pronouncing words clearly. They often also have difficulty with controlling secretions resulting in excessive drooling as well as problems swallowing. This may cause significant feeding difficulties for children with CP (feeding can take a long time) and has implications for nutrition.

Poor nutrition and dental problems may result from feeding and swallowing difficulties. Furthermore, trouble with swallowing makes these children prone to choking on secretions from the mouth and stomach (aspiration). Aspiration may injure the lungs causing difficulty breathing and if it occurs repeatedly can permanently damage the lungs. In the most extreme situation, aspiration can cause sudden death in a young infant with CP.

### **Epilepsy**

Epileptic seizures occur frequently in children with cerebral palsy (CP). The incidence is related to the severity of the brain damage. It is higher in those children with quadriplegia, lower in those with hemiplegia, and much lower in children with diplegia and the athetoid form of CP.

The first epileptic seizures typically are seen during infancy. The seizure disorder is the consequence of the brain abnormalities associated with the CP. Clinical studies indicate that early seizures are associated with more cognitive deficiencies.

Practically every type of epileptic seizure has been described in individuals with CP. Generalized tonic and tonic-clonic seizures and partial complex seizures are most frequent. Some syndromes, such as infantile spasms and

[Lennox-Gastaut syndrome](#) are particularly frequent in children with CP. See [Epilepsy](#) guidance.

## **Hearing and Vision**

Some CP patients have hearing problems, particularly those children who had kernicterus (severe neonatal jaundice). Visual impairment, especially strabismus ("cross-eye") is very common in CP. Other visual problems result from incomplete development of the retina of the eye or from injury to the area of the brain that interprets the signals from the eye, this may result in cataracts and other abnormalities. See [Hearing Impairment](#) guidance.

## **Incontinence**

Children with CP may have poor bladder function caused by impaired control of bladder muscles or a small irritable bladder. This often results in repeated infections and incontinence.

## **Cognitive/Psychological/Behavioural**

Learning difficulties occurs more frequently in persons with CP, the overall rate is between 30-50%. One difficulty in assessing cognitive ability is to evaluate verbal skills and may result in the underestimation of cognitive abilities in CP. There is also increased occurrence of depression, ADHD and autistic spectrum disorders.

see: [Classification & types](#)

## **How is it assessed?**

The brain lesions of Cerebral Palsy (CP) occur from the foetal or neonatal period to up to age 2 years. Although the injury to the brain occurs prior to this, a definitive diagnosis of CP may not be made until after that time. This may be because the clinical picture may not be clear for some time and potentially allows exclusion of progressive diseases. In addition, some children

have been diagnosed with CP at an early age only to have the symptoms resolve later.

The diagnosis of cerebral palsy is made on the basis of:

- History:

History of a complicated delivery with clear evidence of birth injury or of low-birth weight increases the probability of CP.

- Examination:

A complete neurological examination assessing muscle tone, posture, reflexes and motor milestones are crucial to making a definitive diagnosis of CP and also indicate the type of CP.

- Exclusion of other disorders:

It is important to differentiate between cerebral palsy and other neurological disorders such as tumours, degenerative brain disorders and diseases of the spinal cord and neuromuscular disorders.

A Magnetic Resonance Imaging (MRI) examination can often assist as it shows the brain structure with any abnormalities. Additional specialised investigations may be carried out if there is suspicion of specific conditions. Nerve conduction studies are helpful when a muscle or nerve disorder is suspected.

Blood tests may for instance determine abnormal thyroid function that can cause abnormalities in muscle tone or deep tendon reflexes or to movement disorders. Chromosome analysis can rule out a genetic syndrome. Biochemical tests are useful to screen for hereditary metabolic conditions that result in accumulation of poisonous chemicals to the brain.

Additional testing may be necessary to evaluate associated conditions. An Electroencephalogram (EEG) is important to evaluate seizure disorders if they are present.

### **How is it treated and managed?**

Cerebral Palsy (CP) cannot be cured but management aims to use appropriate combinations of interventions to promote function, to prevent



secondary impairments and attempt to increase a child's developmental capabilities.

Its management is multidisciplinary with physical, occupational and nutritional therapy to maximise rehabilitative efforts.

### **Medical treatment**

Oral medications such as baclofen and diazepam are used to reduce spasticity and other medications such as trihexyphenidyl can be used to treat dystonia. Tizanidine (zanaflex) as well as dantrolene (dantrium) are sometimes used to help spasticity, however they are less commonly used due to their side effects.

More recently, botulinum toxin injections such as Botox have been used to reduce the stiffness in specific muscles.

Seizure disorders are managed with anticonvulsant medications. See [Epilepsy](#) guidance.

### **Surgical Treatment**

Surgical management may be helpful to treat contractures, deformities or joint dislocations e.g. tendon lengthening, tendon transfer, bone surgery, hip and spinal surgery.

Selective Dorsal (or Posterior) Rhizotomy is a surgical procedure used to treat spasticity in the lower limbs. It involves cutting some of the nerves in the spine to release the spasticity. Other recent advances with proven benefits are intrathecal baclofen (for spasticity) and deep brain stimulation (for dystonia). Stem cell treatment trials have been conducted but the benefits are uncertain at present.

### **Diet**

Problems with the muscle tone of the tongue and throat may cause limitations in the texture of food and liquid. Some children can only manage pureed food and thickened fluids. Bite size food may cause choking and thin fluids may be aspirated. In severely affected children, feeding may be possible only by a tube directly into the stomach (gastrostomy) or into the small intestine (jejunostomy). Those children with quadriplegia and those who are tube fed are at increased risk of aspiration. A change in solid and liquid diet consistency, teaching techniques to protect the airway, and proper

positioning during and after meals decrease the occurrence of aspiration to the lungs.

### **Walking & self-care activities**

Regular physiotherapy and occupational therapy are crucial. Physiotherapy helps to develop stronger muscles and work on skills such as walking, sitting, and maintaining balance. Occupational therapy can help a child develop fine motor skills such as dressing, feeding, writing, and other daily living tasks.

Various mobility aids may be helpful to assist with walking. Braces may be used to hold the foot in place when the child stands or walks. Some children do require wheelchairs or other special chairs for ambulation. Some chairs also support the entire body in remaining upright. The independent use of these aids by a child is largely dependent on their level of upper limb as well as cognitive function.

Casting and splinting can improve the range of motion of a joint and decrease tone. This can be done on a contracted joint to provide a slow, progressive stretch.

### **Speech and language Problems**

Speaking may be difficult due to problems with the muscle tone of the tongue and throat. Speech therapy can help improve swallowing and communication. Some children also learn to use signing to communicate.

### **Assistive technology (to manage communication problems) includes -:**

- Communication boards have pictures, symbols, letters, or words attached. The child communicates by pointing to or gazing at the pictures or symbols
- Devices such as voice synthesizers enable the child to "talk" with others
- Computer technology such as electronic toys with special switches, computer programs operated by simple switch pads or keyboard adaptations

### **What evidence is available?**

It is important to determine from the customer who regularly manages their condition. It is also important to determine what kind of information you need. This enables contact with the appropriate healthcare professional is

contacted.

<b>Sources of evidence</b>	<b>Information Provided</b>	<b>Limitation of Evidence</b>
Parent / carer	First source of information and associated evidence.	Does not have specialist knowledge.
Neurologist / Paediatrician	Gives an overall profile of significant issues, medication and most up to date treatment and its effect. Nutritional status. Epilepsy control. Respiratory status.	It is important to determine from who is the individual that is regularly managing the condition. This enables contact with the appropriate healthcare professional.
Physiotherapist / Occupational Therapist	Provide details of movement difficulties such as gait and balance problems and ability to carry out tasks such as self-care. Likely to have knowledge of specialised equipment and aids being used.	Same as above
Orthopaedics	Details of outcome and any future surgical procedures to manage hip dislocation, scoliosis, spasticity and release of contractures etc.	Same as above
Speech / language Therapist	Give a profile of function in terms of language and communication skills, feeding skills and aspiration risks.	Same as above
School teacher, Educational Psychologist	Details of learning difficulties. Account of daily functioning and activities in school, including mobility, self-care skills, communication and behavioural issues.	Same as above

### **How long will the needs last?**

This guidance covers -:

Cerebral palsy - quadriplegia
Spastic diplegia
Cerebral palsy - athetoid
Cerebral palsy - causing hemiparesis
Cerebral palsy - ataxic
Cerebral palsy - Other / type not known

- Children with severe forms of CP may have a significantly reduced life span. Those with milder forms of CP have a life expectancy close to the general population, although still somewhat reduced
- Reduced life span is related to the severity of CP and associated medical complications, especially the respiratory ones. In quadriplegic patients for instance the likelihood of

epilepsy, respiratory and other associated conditions is greater than in those with diplegia or hemiplegia

- Increased tone and spasticity may improve or resolve over time in patients with CP, but may not make significant difference to their overall level of function and independence

The overall picture of cerebral palsy looks like -:

- Approximately 25%: have mild involvement with minimal or no functional limitation in ambulation, self-care, and other activities
- Approximately 50% are moderately impaired to the extent that complete independence is unlikely, but function is satisfactory
- About 25% are severely disabled and require extensive care and do not walk. In these cases, the level of functional restriction is unlikely to/will not improve
- 

Age at date of claim	Level of functional restriction		
	Mild	Moderate	Severe
1-5 years	Award to age 8	Award to age 12	Award to age 12
6-10 years	Award to age age12	Award to age age12	Award to age age16
11-16 years	Award to age16 (or for 1 year whichever is the longer)	Award to age 16 (or for 1 year whichever is the longer)	Award to age16 (or for 1 year whichever is the longer)

Regardless of the type of cerebral palsy, those who have significant excess needs have different manifestations with time and this is reflected in the guidance by the needs being considered at different ages. Some with cerebral palsy adapt but a large proportion of others have lifelong significant disabilities. Although there are multiple types of treatments available, such as medication, surgical procedures, Botox injections etc. these are unlikely to

significantly alter functionality and therefore unlikely to alter care/mobility needs.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

## **Causes**

The brain abnormalities in Cerebral Palsy (CP) are associated with a variety of prenatal (before birth), perinatal (during birth), or postnatal (after birth)

events. CP is mostly due to factors occurring before birth, however prenatal diagnosis is not available and often a cause cannot be found.

### **Prenatal risk factors (44%):**

Genetic conditions (chromosomal disorders, hereditary spastic paraplegia), foetal malformations, intrauterine infections, prenatal strokes, maternal conditions e.g. diabetes, thyroid abnormalities, mercury exposure and maternal iodine deficiency.

### **Perinatal risk factors (27%):**

Birth asphyxia (deprived of oxygen), prematurity, maternal pre-eclampsia (severe pregnancy induced hypertension), complications of labour and delivery, perinatal infections e.g. Group B Streptococcus infection, kernicterus (severe neonatal jaundice, sometimes due to blood group incompatibility).

### **Postnatal risk factors (5%):**

Infections in early infancy (meningitis, encephalitis), bleeding in the brain, head injury e.g. as a result of child abuse.

In 24% of people with cerebral palsy, the cause is not obvious. This percentage may drop with the advances in diagnostic facilities.

## **Classification and types**

### **Current official classification of cerebral palsy**

The previous way of describing Cerebral Palsy (CP) was felt to be insufficiently accurate and is to be replaced by a new way of classifying CP (from

the consensus conference). This is comprised of a description of four main dimensions -:

- Motor abnormalities
- Accompanying impairments
- Anatomical and neuro-imaging findings
- Causation and timing

This is relatively new and not all professionals have switched to this new way of thinking as yet. Therefore, most reports to date will still define and classify cerebral palsy as described below.

### **Classification according to limbs affected**

- Monoplegia - only one limb is affected
- Hemiplegia – 20 - 30%; one side of the body is affected and usually affects the upper and lower limbs
- Diplegia – 30 - 40%; impairment primarily of the legs (often with some involvement of arms to lesser extents)
- Quadriplegia (tetraplegia) – 10 -15%; all four limbs are affected, and the trunk is often involved

### **Classification according to movement disorder**

Spastic Cerebral Palsy:

- Most common type (70- 80% of cases), as a result of impairment to the nervous pathway known as the pyramidal tract, which include the surface of the brain ('motor cortex') where voluntary movements are initiated, and the nerve fibres that carry the signals to the spinal cord
- Spastic muscles are tight and stiff and have increased resistance to being stretched. The stiffness affects certain patterns of muscles, causing an imbalance so the child's limbs

can be pulled into certain positions which then can be difficult to move out of

- Spasticity may be mild and affect only a few movements, or severe and affect the whole body. The amount of spasticity usually increases over time

#### Dyskinetic Cerebral Palsy: (Dystonic and Athetoid)

- Accounts for 10-15% of CP cases. As a result of impairment to the basal ganglia in the brain, this fine-tunes signals to the muscles.
- Affected children have variable muscle tone i.e. the tension in the muscles can change between low and high and can occur suddenly and be from one extreme to the other.
- It can cause children to become stuck in a position or can make it difficult for them to be still. The fluctuations often occur as a result of stimulation e.g. excitement, fear or effort. This often appears as posturing patterns e.g. neck arching back, arms stretching and turning inwards.
- Children with athetoid cerebral palsy have many involuntary movements and some are constantly in motion.
- They often have speech difficulties.

#### Ataxic Cerebral Palsy:

- Occurs in <5% of cases, due to impairment to the cerebellum, the brain's major centre for balance and coordination.
- Affected children have jerky, uncoordinated movements caused by a disturbed sense of balance and depth perception. They usually have poor muscle tone, a staggering walk and unsteady hands.

#### Mixed Cerebral Palsy:

- Most children with cerebral palsy have a mixture of spasticity and dystonia
- Some will have a predominant spastic picture and others will have a predominant dystonic pattern.
- In some children, there is predominant underlying weakness as the main cause of functional impairment. Treatment that



modifies spasticity ('stiffness') or dystonia ('posturing') has limited effects in improving function.

The classifications of movement disorder and number of limbs involved are usually combined. For example, spastic diplegia refers to spastic type CP that affects primarily the legs.

## Cleft lip/palate

What is a Cleft lip/palate?

The palate is the roof of the mouth and is divided into the soft palate (at the back) and the hard palate (at the front). During development in the womb, two seams (just left and just right of the mid-line) close up like zips to form the upper lip and palate. That is why the upper lip has its 'double-peak' shape. When this seam closure is incomplete, the baby is born with a gap or 'cleft' of either the palate (back of the seam), or lip (front of the seam) or both. Sometimes, the term 'oro-facial cleft' is used to encompass all of these variations. The proportion of babies born with oro-facial clefts (incidence) varies by country and race but is about 1 in 700 in the UK.

Like all faults of embryo formation, babies born with oro-facial clefts are more likely than normal to have problems with their genetic blueprint and more likely to have other embryo formation (dysmorphic) problems.

This means that people with oro-facial clefts may have problems of four kinds:

1. Physical consequences of the cleft – for example speech, swallowing, middle-ear ventilation problems and dental problems
2. Social and psychological consequences of the cleft – the burden of 'looking different'
3. Treatment burden (see below)
4. A higher risk of associated medical problems

## Cleft lip

This represents 30% of occurrences of oro-facial cleft.

Cleft lip is formed in the top of the lip as either a small gap or an indentation in the lip (partial or incomplete cleft) or it continues into the nose (complete

cleft). Cleft lip can occur as a one sided (unilateral) or two-sided (bilateral) feature. The palate is not affected.

Within the first 2–3 months after birth, surgery is performed to close the cleft lip.

#### Cleft palate

This represents 25% of occurrences.

Cleft palate is a condition in which the two plates of the skull that form the hard palate (roof of the mouth) are not completely joined. The soft palate is affected in these cases cleft as well.

Often a cleft palate is temporarily covered by a palatal obturator (a prosthetic device made to fit the roof of the mouth covering the gap).

Cleft palate can also be corrected by surgery, usually performed between 6 and 12 months. Approximately 20–25% only require one palatal operation to achieve a seal good enough for speech and swallowing. Unfortunately, the rest require combinations of surgical methods and repeated operations as the child grows. One of the innovations of cleft lip and cleft palate repair is the Latham appliance. The Latham is surgically inserted by use of pins during the child's 4th or 5th month. After it is in place, the doctor or parents turn a screw daily to bring the cleft together to assist with future lip and/or palate repair.

#### Cleft lip and palate

This represents 45% of occurrences.

This is a combination of cleft lip and palate and its management is surgical closure as outlined in the previous sections.

All oro-facial cleft conditions are treatable. However, the kind of treatment depends on the type and severity of the cleft. All children in the UK are monitored by a specialist cleft palate team or craniofacial team throughout young adulthood. Care can be life-long.

- Many babies with oro-facial clefts are diagnosed before birth by ultrasound and the specialist team is involved from delivery, so claims may be made early
- Treatment burden and the impact of swallowing problems may well result in significant increases in the burden of care beyond that of an unaffected newborn from the very start of life
- Associated conditions cited in claims may include Pierre-Robin Syndrome (also known as Pierre-Robin Sequence) which also affects the jaw, Stickler's Syndrome (which affects the jaw and the eyes), 22q11 deletion (also known as Velo-Cardio-Facial syndrome) and DiGeorge Syndrome, which can have very wide-spread physical and intellectual effects and other, rarer genetic syndromes. Alternatively

congenital (inborn) structural heart problems or eye problems may be mentioned without a syndrome name

- Recurring middle ear infections and psycho-social problems will start to become relevant to claims in older pre-school and school age children.
- Speech development problems can be very severe indeed because of the unique combination of hearing impairment and problems with articulation and sound formation because of the palate

## Prognosis

### Cleft lip

For isolated cleft lip, outcomes from surgery are usually good but, it can be difficult to assess this until significant speech has developed. Awards for the 0-4 age group should be until the age of 5, or 2 years, whichever is longest.

Subsequent awards will be dependent on any surgical interventions. An award of 2 years followed by subsequent review for evidence is advised.

### Cleft palate or Cleft lip and palate

Surgical correction of these cases is more complicated and review interventions occur more commonly than in isolated cleft lip.

The following table is to be followed if an award is made. It is not a statement of intended awards for individual categories.

Age at date of claim	Mild	Moderate	Severe
0-4	To age 5 (or 2 years whichever is greater)	To age 5 (or 2 years whichever is greater)	To age 5 (or 2 years whichever is greater)
5-9	2 years	2 years	5 years
10-12	2 years	2 years	5 years (or until aged 16 whichever is shorter)
13-15	Until age 16	Until age 16	Until age 16

## Clubfoot

### Description

Clubfoot (talipes equinovarus) is a common disorder, where one or both feet are turned downwards and inwards at the ankle so that the sole of the foot

may appear to face backwards or even upside down. The tendons on the lower leg are shortened with a tight Achilles tendon.

It is usually picked up on antenatal scans or immediately after birth. The condition is painless, but the foot cannot be moved into a normal walking position. Both feet are affected in approximately half of cases.

## Treatment

Treatment for club foot usually starts within a week or two of the baby being born. A technique known as the 'Ponseti method' is the main treatment for club foot nowadays, which involves gently manipulating the baby's foot into a better position, then putting it into a cast. This is repeated every week for about 5 to 8 weeks.

After the last cast comes off, most babies need a minor operation to loosen the tendon at the back of their ankle (Achilles tendon). This is done using a local anaesthetic. It helps to release their foot into a more natural position.

The baby will need to wear special boots attached to each other with a bar to prevent the club foot returning. They'll only need to wear these full-time for the first 3 months, then overnight until they're 4 or 5 years old.

## Boots and Bars

When a child wears boots and bars for clubfoot, they are still able to move the legs, but they can only move together. While it might take them a little longer, children wearing a brace can learn how to sit, crawl and walk just like other children their age.



Image kindly provided by C-Pro Direct

During the first days of using the device, the parents/guardians are encouraged to move the bar gently up and down so the child sees how the legs

can move and kick together. They are also encouraged to push and pull the brace, so the child learns how to bend his or her knees while in the brace.

The majority of the children learn how to use the device properly in the first few days of use.

Attention at night is unlikely to be needed.

## What you need to know about Coeliac disease

### What is Coeliac disease?

Coeliac disease is an inflammatory condition of the small intestine .....

- [Coeliac disease](#)

### What are the effects and signs?

Older children are more likely to present subtle gastrointestinal symptoms such as abdominal pain, intermittent diarrhoea / constipation and slowing of growth .....

- [Effects of Coeliac disease](#)

### How is it assessed?

Blood tests for abnormal antibodies are used to identify which children need further testing for coeliac disease .....

- [Assessment of Coeliac disease](#)

### How is it treated and managed?

The only current treatment of coeliac disease is avoidance of gluten. The treatment is called a gluten free diet. ....

- [Treatment for Coeliac disease](#)

### What evidence is available?

The GP will be able to confirm the diagnosis and will have records of gluten free items prescribed .....

- [Evidence](#)

### How long will the needs last?

- [Prognosis and duration of the award](#)

## **What is Coeliac disease?**

Coeliac disease is an inflammatory condition of the small intestine. Another name for coeliac disease is gluten sensitive enteropathy. The condition is caused by an abnormal immune response to dietary protein called gluten found in wheat, barley and rye. The abnormal response causes inflammation of the mucosa of the small intestine and flattening of the villous processes – these are the finger like projections of intestinal mucosa, which serve to increase the surface area and absorptive capacity of the small bowel. The condition causes symptoms which may be gut related or not. If it is not treated complications related to malabsorption of nutrients may occur e.g. inadequate uptake of vitamins and minerals from the diet.

The treatment of this condition is a 'gluten free diet'. If the diet is adhered to symptoms resolve completely and complications of the condition are minimal. Even though a person becomes completely well on a gluten free diet they still have coeliac disease, which is a lifelong condition.

## **Incidence/prevalence?**

Coeliac disease is most common in white European populations and runs in families. The condition occurs in genetically predisposed people. The risk of a person developing coeliac disease at some time in their life is 1 in 100 in the UK but in those with an affected close relative, such as a parent, or brother or sister, it is 1 in 10. The disease typically presents in childhood, but also presents in adults.

The onset of the disease is related to timing and amount of exposure to gluten in the diet. Since the 1970s weaning guidelines have postponed introduction of gluten-containing products from 3 months of age to at least 6 months of age and this may have reduced the incidence of coeliac disease in early childhood. The incidence of coeliac disease is 1 in 2500 to 1 in 3000 children per year with a median age of onset of 7 years.

It is estimated that only up to 20% of children in the UK who have coeliac disease have actually been given a formal diagnosis.

## **What are the effects and signs?**

The classic presentation of coeliac disease in infancy is less common than it was. Affected babies typically developed symptoms after weaning, when cereals containing gluten were introduced to the diet. They exhibited 'faltering growth' and were miserable, refused to eat and did not gain weight.

Typically, they had a distended abdomen, muscle wasting and offensive diarrhoea.

Older children are more likely to present subtle gastrointestinal symptoms such as abdominal pain, intermittent diarrhoea / constipation and slowing of growth.

In summary, the symptoms of coeliac disease are variable and may be non-specific. They may or may not include gastrointestinal symptoms. In almost all cases, symptoms resolve with a gluten free diet.

The following symptoms may occur -:

- Chronic or intermittent diarrhoea
- Failure to thrive or faltering growth in children
- Persistent and unexplained gastrointestinal symptoms including nausea and vomiting
- Prolonged fatigue
- Recurrent abdominal pain, cramping or distension
- Sudden or unexpected weight loss
- Unexplained anaemia

The condition may have no symptoms but be identified because children with the following conditions have been screened with an antibody test -:

- Autoimmune thyroid disease
- Dermatitis herpetiformis (a skin rash)
- Irritable bowel syndrome
- Type 1 diabetes mellitus
- A close relative, such as a parent, or brother or sister, with coeliac disease
- Down's syndrome
- Turners' syndrome

In children, the complications of untreated coeliac disease in addition to any symptoms it causes include the following -:

- Growth failure (small for age)



- Delayed puberty
- Dental problems
- rickets
- osteoporosis
- see: [Care and mobility considerations](#)

## How is it assessed?

### Blood tests

Blood tests for abnormal antibodies are used to identify which children need further testing for coeliac disease. Antibody blood tests may also be used at follow up visits to check whether the gluten free diet is being followed – the antibody tests become negative once gluten is no longer eaten.

The following blood tests are used -:

- tTGA testing – this measures IgA tissue transglutaminase antibody and is the main test used currently. This test may be falsely negative if there is IgA deficiency
- EMA test – IgA endomysial antibody (EMA) test is used if the tTGA test is not clearly positive or negative – the result will be called ‘equivocal’ in medical evidence

If either of these tests is positive, referral for intestinal biopsy will be made. If IgA deficiency is present, other antibody tests will be performed instead -:

- IgA levels to quantify IgA deficiency
- IgG tissue transglutaminase antibody
- IgG endomysial antibody (EMA) test

### Intestinal Biopsy

This test involves a gastroscopy and biopsy of the duodenum, the first part of the small intestine. It is a definitive test for coeliac disease as long as sufficient gluten is being eaten as part of the diet. The biopsy and blood tests will be normal in a person with coeliac disease if they are sticking to a

gluten free diet. A repeat biopsy to check whether the gut lining has healed is not necessary in most cases.

### **How is it treated and managed?**

The only current treatment of coeliac disease is avoidance of gluten. The treatment is called a gluten free diet. The consequences of eating gluten may be abdominal pain and diarrhoea or recurrence of other symptoms or no symptoms at all; often it will depend on whether there were symptoms to start with. The symptoms are not life threatening but may be unpleasant. Children who are diagnosed on screening and have an abnormal biopsy but who do not have symptoms on eating gluten, may find it more difficult to follow the diet. The consequences of continuing to eat gluten in this circumstance are not known but are thought to be the same as for any child with an abnormal biopsy.

### **What is a gluten free diet?**

A gluten free diet is essentially a diet free from wheat, barley and rye, all of which are frequently included in processed foods. This means that durum

wheat, semolina, barley and rye cannot be eaten. Typically, the following cannot be eaten because they contain gluten -:

- Bread
- Pasta
- Cereals
- Biscuits and crackers
- Cakes and pastries
- Pies
- Gravies and sauces
- Oats (some people with coeliac will have to avoid oats in addition to wheat)

Gluten free versions of the above foods are available. They can be expensive and so foods and gluten free flour for home baking may be provided on prescription.

The following foods are naturally gluten free and can be eaten freely -:

- Milk, cheese and butter
- Fruit
- Vegetables
- Meat and fish (not breaded or marinated)
- Potatoes
- Rice
- Gluten free flours and grains including rice flour, corn flour, potato flour

### **Other treatments**

Vitamin supplementation may be needed for the first few months to make up for deficiencies that developed before diagnosis. Examples of vitamins and mineral supplements include -:

- Calcium

- Iron
- Folate

**Treatment for dermatitis herpetiformis (a skin manifestation of coeliac disease)**

Drug treatment in the form of dapsone tablets taken twice daily may be prescribed for this condition. It does resolve on a gluten free diet but dapsone treatment helps to speed up the process.

- See: [Care and mobility considerations](#)

**What evidence is available?**

Sources of Evidence	Information Provided	Limitation of Evidence
Parent/Carer	First source of information and associated evidence.	Does not have specialist knowledge.
GPFR	The GP will be able to confirm the diagnosis and will have records of gluten free items prescribed.	The child is likely to have coeliac disease follow up at the hospital. The GP may not know about ongoing problems with the disease or dietary treatment.
Hospital FR	The specialist consultant is likely to be able to confirm the diagnosis and will be able to give a reasonable description of the effects of the condition on the child and their family.	If the problems relate to following the gluten free diet, the specialist may not have information about this. The hospital dietician seeing the family will be a better source of evidence in such cases.

**How long will the needs last?**

Age at date of claim	Award period
0 - 11	Award to age 12 (or for 1 year, whichever is the longer)

If care needs related to supervision of dietary intake are identified, entitlement should be considered in the context of why those specific care needs are present and whether those needs are substantially in excess. Duration of award should also be considered having regard to whether improvement of any other condition present, such as learning, or a behavioural disorder is expected.

## Background

The child will need to remain on a gluten free diet for life. Otherwise, healthy children will learn how to do this for themselves in childhood becoming proficient at maintaining their safe diet by age 11 to 12. This is the age at which they will be expected to make unsupervised consistently correct food choices at lunch times. Children diagnosed in early childhood may be proficient at managing safe food choices from age 8. Recently diagnosed older children take 3-6 months to learn how to manage their gluten free diet. Otherwise, healthy children with coeliac disease can cooperate with the diet their parents provide for them. Additional thought is required to provide a healthy diet for affected children, but the effort is not substantially more than providing a healthy diet for a child without coeliac disease.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

## Care and mobility considerations

Children with coeliac disease can remain completely well on a gluten free diet. Parents will need to provide this for their children on a daily basis until the child can manage their own dietary intake. The consequences of eating gluten are not life threatening in children with coeliac disease; however, the consequences of continuing to eat gluten even if it does not cause symptoms are significant. They include an increased risk of other autoimmune diseases (diabetes, thyroid disease), osteoporosis, and very rarely, development of small bowel lymphoma. There is no treatment to reduce symptoms having eaten a foodstuff containing gluten.

## Care needs

Children diagnosed with coeliac disease will require more care and attention to their diet than an unaffected child. This is because they are unable to

eat a common component of the diet, namely gluten. Parents will need to provide completely gluten free meals for their children and will need to ensure that their food does not become contaminated with gluten during food preparation or storage. For example, gluten free bread should not be toasted in a toaster used for normal bread; similarly separate butter should be used so that butter does not act as a transfer medium for gluten containing crumbs to the gluten free bread. Children will eat a diet of naturally gluten free foods and some gluten free substitutes for wheat-based products.

Children are not at risk from gluten in the environment or from touching or using wheat-based products providing that they do not put them in their mouths or eat them.

### **Typical food preparation practice**

Typically, parents of children with coeliac disease will be very knowledgeable about their condition and foods that may contain gluten.

In order to avoid feeding their child gluten, families will often prepare most meals from scratch at home. This is the only way to ensure meals are not contaminated. Pre-prepared meals and sauces for home use or in catering often contain many different ingredients and unless clearly stated on the box there is always the possibility of contamination. A suitable item might list 'gluten free' or 'suitable for a gluten free diet' on the label. The Coeliac Society provides updated lists of safe foods including gluten free substitutes, recipes and tips for managing the diet. This type of information makes managing a gluten free diet much easier than it otherwise would be.

A common problem when eating out is contamination of cooking oil for example fish and chip shops may fry chips separately having previously used the oil to fry battered fish. So, although the chips should be safe, they are contaminated with gluten from wheat flour batter. Similarly, other bought products may be unexpectedly contaminated e.g. gluten free chickpea flour may be contaminated with wheat gluten during the milling process at the mill by wheat flour in the machinery or in the air. Very small amounts of contamination may make a food unsuitable. For example, the international standard for gluten free food which means that a food can be labelled gluten free is that the food contains less than 20 parts per million of gluten. Such food is safe for anyone with coeliac disease. 'Very low gluten' foods can be labelled as such if they contain less than 100 parts per million of gluten – this may be too much for some people with coeliac disease. Parents

will provide supervision to prevent their child from eating gluten and provide gluten free food until they can do this for themselves.

## What you need to know about Congenital Heart Disease (CHD)

What is Congenital Heart Disease?

Congenital Heart Disease is a problem with the structure of the heart that has been .....

[Congenital Heart Disease](#)

What are the effects and signs?

In Infants, this may present as poor feeding during which the baby may become tired. Fast breathing and .....

[Effects of Congenital Heart Disease](#)

How is it assessed?

50-60% of babies with congenital heart disease will have been diagnosed within the first months .....

[Assessment of Congenital Heart Disease](#)

How is it treated and managed?

Treatments for congenital heart disease are variable and depend on the individual lesion type and .....

[Treatment for Congenital Heart Disease](#)

What evidence is available?

Details of which investigation results can be used to confirm disability affecting walking and .....

[Evidence](#)

How long will the needs last?

[Prognosis and duration of the award](#)

## What is Congenital Heart Disease (CHD)?

Congenital Heart Disease is a problem with the structure of the heart that has been present since birth. The signs and symptoms can vary from condition to condition and can vary from none to life threatening. Usually, they are dependent on the severity of the underlying cardiac defect.

The incidence of CHD is 5 to 8 per 1,000 live births.

Congenital cardiac defects have a wide variety of severity with only 2-3% being symptomatic as infants. By one month of age, 50-60% of cases will have already been diagnosed.

## The Normal Heart

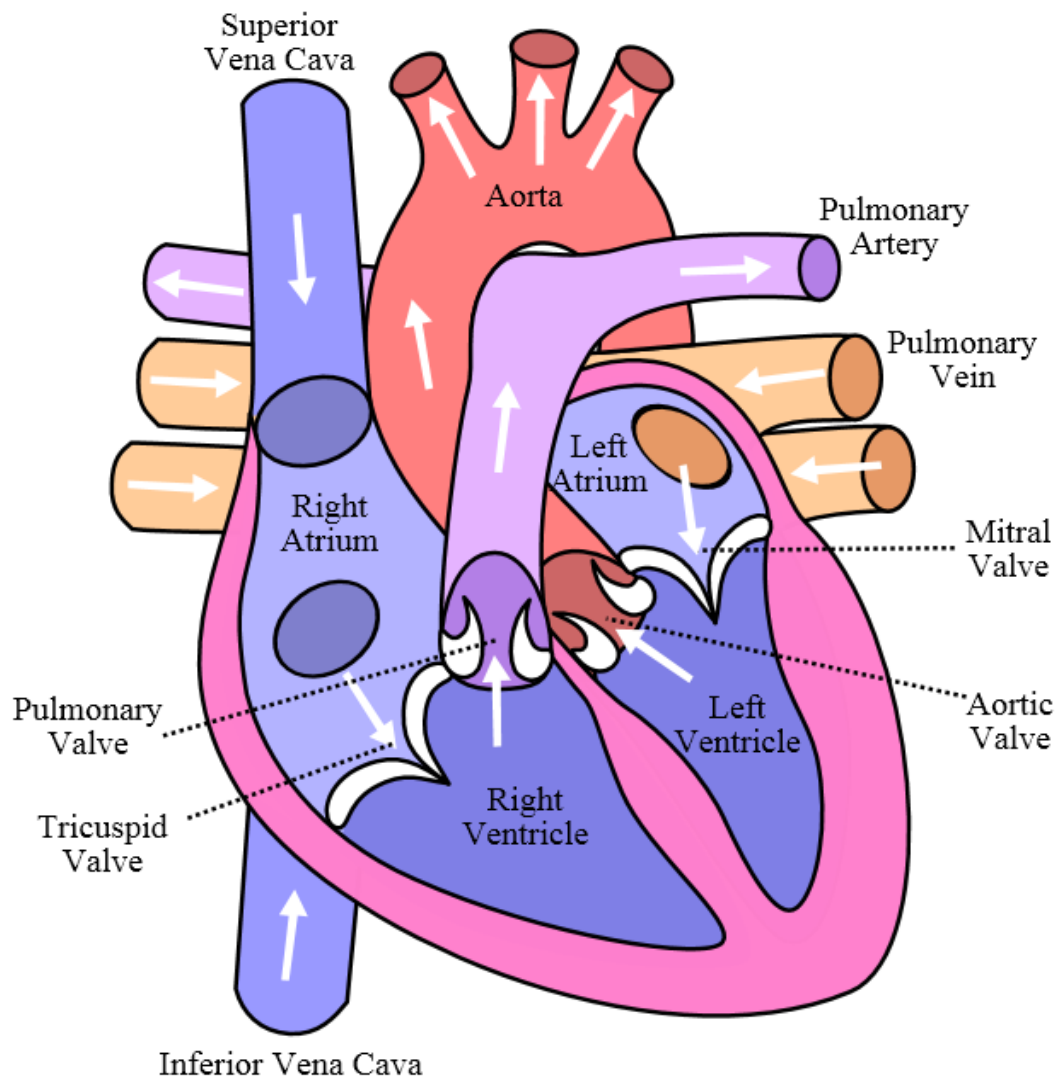
The heart is basically a pump for the body's transport system; the hollow blood vessels are the delivery routes. Using blood as the transport medium, the heart continually propels oxygen, nutrients, waste and many other substances into the interconnecting blood vessels that move to and from the cells of the body.

The heart comprises four chambers: the right atrium, right ventricle, left atrium and left ventricle. The right side of the heart receives blood from the body and pumps it to the lungs to be filled with oxygen. The left side of the heart receives oxygen rich blood from the lungs and pumps it around the rest of the body. Between the atrium and ventricle on each side and between the ventricles and the pulmonary artery/aorta (outflow tracts) there



are valves. The valves are important in ensuring blood flows in the correct direction.

The heart is enclosed in a double-walled sac called the pericardium. The pericardium anchors the heart to surrounding structures, provides lubrication for contraction and acts as a barrier to infection.



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What are the effects and signs?

Congestive Heart failure

In Infants, this may present as poor feeding during which the baby may become tired. Fast breathing and a fast heart rate may also be present as well

as cough and sweating. The most common symptom that can occur is faltering growth.

In children, heart failure causes difficulty in breathing and depending on the severity, can be at rest or on exertion. Symptoms of breathlessness in heart failure can be variable.

Other symptoms and signs present will be a fast heart rate, sweating, pallor, poor feeding, liver and heart enlargement.

### Cyanosis

Cyanosis is the appearance of a blue or purplish discolouration of the skin or mucous membranes that is due to low levels of blood oxygen levels within the circulation.

### Faltering growth

This symptom is defined as the failure to maintain a normal growth rate such that the child or baby's growth falls significantly behind that of the normal population – See: Faltering growth guidance.

### Heart murmur

A heart murmur is a term used to describe sound from turbulent flow of blood through the heart valves or the outflow of blood through an abnormal connection from one heart chamber to another.

Heart murmurs in the majority of children, however, are innocent. In these cases, there is no underlying structural abnormality, but the murmur is simply heard as a result of noisy flow of normal valves.

### How is it assessed?

50-60% of babies with congenital heart disease will have been diagnosed within the first months of life and 80-90% within the first year. In many cases a heart defect may be suspected from the routine ultrasound scan carried out in pregnancy. When that happens more detailed scanning, known as foetal echocardiography, can be performed to try to confirm the exact diagnosis. However, it may not be possible to do this until after birth. Detailed scanning is also offered in pregnancies where there is thought to be an increased risk of the baby having congenital heart disease. The diagnosis of CHD is usually made in hospital by consultants in foetal medicine or

consultant paediatric cardiologists after multiple investigations, the most important of which is an echocardiogram.

Some children will have undergone an invasive test called a heart catheter. This is usually performed by a specialist prior to undergoing any planned surgical interventions.

The infant will usually have been referred to a specialist paediatric cardiologist at a specialist hospital. If surgery is required, this will be carried out by a consultant paediatric cardiothoracic surgeon.

How is it treated and managed?

## Introduction

The majority of children with congenital heart disease will have minor lesions or completely repaired congenital heart disease. These children will have no symptoms or minimal symptoms and are likely to be on 1-3 yearly follow-up. There are certain diagnostic subgroups which represent the more severe end of the spectrum of congenital heart disease and these children are more likely to have functional restriction, these children are most likely to be managed primarily by surgery. These groups include:

- **Children with a single ventricle circulation.** Diagnoses include hypoplastic left heart syndrome, hypoplastic right heart syndrome, tricuspid atresia, double inlet left ventricle, mitral atresia. The initial palliation (operation to control symptoms) is usually undertaken in the first few days or weeks of life (Norwood procedure, BT shunt, ductal stent, pulmonary artery band). Babies and infants will be cyanosed (blue) at rest and will usually be part of a care of the '**high risk infant programme**' at the congenital cardiac centre. These babies and infants will often need considerable additional care in terms of saturation monitoring, the need to ensure weight gain and early presentation to secondary or tertiary care in the event of a deterioration.

**High risk infant programmes** involve frequent clinical monitoring of the child by the parent carer at home – See: Congenital Heart Assessment Tool2 (CHAT2). This form is an example of a tool used by parents to check their child at least daily so that they can intervene by attending hospital as soon as any deterioration in clinical condition occurs. The frequency of checks and the tools the parent uses will vary between hospitals and between children. The parent/carer will be able to explain what they are doing.

- The 2<sup>nd</sup> palliation (operation to control symptoms, cavopulmonary anastomosis/Glenn procedure) is usually undertaken between 3 months and a year of age. Children will remain cyanosed, though

with improvement in their exercise capacity. As these children become older, they will again become more cyanosed and are then considered for the final stage of the palliation (total cavopulmonary anastomosis/Fontan procedure). This is usually carried out between 2 and 6 years of age. Following the total cavopulmonary anastomosis symptoms usually improve again. These children will often be anticoagulated, and symptoms are highly variable between children and include degrees of exercise limitation and other symptoms such as nocturnal leg pain. As they become older symptoms often get worse and a proportion of these children/young adults are considered for heart transplantation. Life expectancy is significantly reduced but unknown with modern treatment. Some children may spend many years feeling well with reduced exercise tolerance before they deteriorate. Others may remain unwell for long periods and have only short periods of feeling better and having fewer symptoms.

- **Children with palliation prior to complete repair of congenital heart disease.** Some children, for example, those with pulmonary atresia or Tetralogy of Fallot, will undergo initial palliation in a similar way to the 1<sup>st</sup> stage of the single ventricle circulation above. During this time, they will need considerable additional care. Once the complete repair has been undertaken and the child has recovered from that surgery there is usually a little functional restriction. Less commonly a complete repair cannot be carried out and there will be functional limitation.
- **Children with heart failure or pulmonary hypertension.** Children may develop heart failure from several different diagnostic categories of congenital heart disease as well as cardiomyopathy. Some of these, particularly those on the heart transplant waiting list will have severe functional restriction. Children with pulmonary hypertension can have functional limitation and often need strict attention, for example being given regular medication which in severe cases can include a continuous infusion.

## Heart Transplant

Some children will deteriorate to the point of being listed for heart transplant. Their condition is likely to be dramatically better, once they have had their transplant, in terms of their symptoms that related to the heart failure (breathlessness, failure to gain weight, inability to exercise). Following transplant, they may start to grow and be able to run about and join in at school. There is an ongoing need for some additional care related to taking drugs to prevent rejection and control other aspects of health care following heart transplant including risk of infections. Life expectancy following

transplant is reduced, although in the UK more than 80% of children are alive 10 years from heart transplant.

#### Change of circumstances after surgery and due to deterioration

Although many children who have had surgery for their congenital heart disease will be well with minimal symptoms, there will be some, particularly those with complex disease requiring multiple palliative procedures who may never be particularly well. These children may require considerable care even after surgery to improve their condition and when they are at their best.

#### Non-surgical treatments for congenital heart disease

##### Anticoagulant drugs

These drugs are used to prevent blood clots forming in the circulation and are commonly required in congenital heart disease because artificial valves or implants have been used, artificial valves and implants predispose to blood clots. Clots can cause pulmonary embolism, chronic lung damage and predispose to sepsis around artificial heart valves causing serious illness and sometimes necessitating their removal. The effect of the anticoagulant is to make the blood less likely to clot, so preventing clots, which is helpful, but a side effect is an increased risk of prolonged bleeding if the child cuts themselves. Sometimes it can be difficult to control the anticoagulation, so the risk of bleeding is significant. The child will require attention around taking their anticoagulants, blood test monitoring and sometimes extra supervision to prevent injury or treat injury quickly if the coagulation is not well controlled. The most common anticoagulant in children is warfarin although direct acting oral anticoagulants (DOACS) are being increasingly used in older children/ young adults. DOACS include apixaban, dabigatran, edoxaban, rivaroxaban. The blood test most often used for anticoagulant monitoring with warfarin is called the 'INR' or 'INR monitoring'.

##### Beta blockers

These drugs block beta receptors principally within the heart itself. The stimulation of these receptors that occurs when the heart is under stress causes the heart to beat harder and faster. Although this a normal and useful function in the normal functioning heart: it can cause symptoms, signs

and accompanying disability in some children with congenital heart disease. The use of beta blockers can help reduce this.

#### Side Effects

- Tiredness (1-10% of patients)
- Dizziness (1-10% of patients)
- Headache (1-10% of patients)
- Sleep disturbance (1-10% of patients. Treatable with melatonin)

Commonly used beta blockers:

- Atenolol
- Carvedilol

#### Diuretics

Diuretics are also known as water tablets as they are principally used when a patient has heart failure and is retaining too much water. They work by encouraging the kidney to excrete larger volumes of water. This reduces the overload on the heart and as a result, improves the child's symptoms.

#### Side Effects

The main side effect of diuretics is passing large amounts of urine. Daily dosing is optimised to reduce the unnecessary burden of this during the day; however, these drugs can be taken up to three times each day.

Diuretic medication

- Furosemide
- Spironolactone

Regular blood test for kidney function and body salts such as sodium and potassium need to be monitored to make sure that the levels of these substances do not cause healthcare needs.

#### Ace Inhibitors/Angiotensin receptor blockers

These two types of medication work by reducing the effect on the kidney of the renin-angiotensin system. Activation of the renin-angiotensin system causes the kidney to retain too much water and it also makes the arteries in the body tighten up. Both of these effects worsen heart failure by

overloading the heart. Taking either of these drugs helps to block the effects of this chemical cascade and so improves the symptoms of heart failure.

#### Side effects

- Cough
- Rash

#### Ace Inhibitors

- Ramipril
- Lisinopril
- Enalapril

Kidney function and blood tests are required whilst treatment is initiated and after dosage changes.

#### Digoxin

Digoxin is a drug that helps improve the pumping ability of the heart muscle. The improvement in heart function helps to control the associated symptoms of reduced exercise tolerance and breathlessness.

#### Side effects

- Kidney damage
- Anorexia
- Nausea vomiting
- Diarrhoea
- Blurred vision

#### Monitoring

High levels of digoxin can accumulate within the body if kidney function worsens. Regular blood tests for digoxin levels and kidney function are required.



What evidence is available?

Sources of evidence	Information provided	Limitation of evidence
Parent / Carer	First source of information.	May not be objective about child's health needs. Will not have up to date information about the treatment of the claimant's medical condition.
General Practitioner	Copies of latest clinic letters should be available from the GP record.	The GP may only be able to confirm diagnosis and medication prescribed for children with congenital heart disease as they usually receive their care via paediatric cardiologists/cardiothoracic surgeons.
Consultant cardiologist/cardiothoracic surgeon	The best source of up-to-date information of ongoing treatment and investigations.	Details of which investigation results can be used to confirm disability affecting walking and care needs typically associated with various treatments are in the guidance. The medical evidence may contain a lot of complex information that needs sifting through.
School	May be able to supply information about any additional help that a claimant receives at school.	Unable to supply information about specific health needs or treatments.

How long will the needs last?

The following table is to be followed if an award is made. It is not a statement of intended awards for individual categories.

If transformative surgery **is planned**, make the award to 1 year post surgery to allow for physical recovery and child/family adaptation to the new level of needs.

Age at date of claim	Length of Award		
	Mild	Moderate	Severe
0-4 years	Award for 2 years	Award for 5 years	Award for 5 years if treatable condition. Award to age 16 if no transformative treatment planned
5-9 years	Award for 2 years	Award for 5 years	Award for 5 years if treatable condition. Award to age 16 if no transformative treatment planned.
10-14 years	Award for 2 years	Award for 5 years or to age 16 (depending on age at date of claim)	Award for 5 years or to age 16 (depending on date of claim) if treatable condition. Award to age 16 if no transformative treatment planned.
15 years	Award to age 16	Award to age 16	Award to age 16

You must take all information into account when considering the duration of care and mobility needs. You must base the duration of care and mobility needs on the particular circumstances of the child.

Congenital Heart Assessment Tool 2 (CHAT 2)

**Congenital Heart Assessment Tool 2 (CHAT2) Parent Version**

<b>Behaviour</b>	<input type="checkbox"/> Behaves normally e.g. content, smiles <input type="checkbox"/> Stays awake or awakens quickly (as normal) <input type="checkbox"/> Normal crying, easily soothed e.g. by feeding, comfort	<input type="checkbox"/> Quieter than usual <input type="checkbox"/> Sleeping more than normal or less easy to settle <input type="checkbox"/> Less responsive during normal activity <input type="checkbox"/> Slightly irritable, unable to comfort	<input type="checkbox"/> Not responding to usual interactions <input type="checkbox"/> Does not wake or if roused does not stay awake <input type="checkbox"/> Weak or floppy <input type="checkbox"/> High-pitched or continuous cry or no crying at all
<b>Skin Colour Warmth</b>	<input type="checkbox"/> Your baby's usual colour of skin, tongue, especially lips and nails <input type="checkbox"/> Your baby's usual warmth of hands and feet. <input type="checkbox"/> Soft spot feels normal. <input type="checkbox"/> Normal body temperature between 36.5 – 37.5 °C	<input type="checkbox"/> Blue tinge around mouth or finger tips, paler than usual, or skin is mottled <input type="checkbox"/> Sweaty skin or cooler hands and feet than usual <input type="checkbox"/> 'Soft spot' feels different to normal <input type="checkbox"/> Temperature more than 37.5 – 38 °C or below 36 °C	<input type="checkbox"/> Very mottled, pale, blue or grey skin <input type="checkbox"/> Very cold, sweaty or clammy <input type="checkbox"/> Very puffy eyes, hands, feet or tummy <input type="checkbox"/> Very sunken 'soft spot' <input type="checkbox"/> Temperature more than 38°C
<b>Breathing</b>	<input type="checkbox"/> Usual breathing rate and usual effort for your baby (ease of breathing) <input type="checkbox"/> No cold symptoms, such as runny nose	<input type="checkbox"/> More breathless, faster or slower breathing rate <input type="checkbox"/> Has cold symptoms, runny nose, cough, noisier breathing than normal <input type="checkbox"/> Sucking in skin below ribs or in the lower front neck area, more than normal <input type="checkbox"/> Nasal flaring (nostrils become wider as baby breathes)	<input type="checkbox"/> Not breathing <input type="checkbox"/> Struggling to breathe or exhausted <input type="checkbox"/> Cannot cry <input type="checkbox"/> Very noisy breathing, grunting or gasping <input type="checkbox"/> Severe 'sucking in skin' below ribs or lower front neck area, head is bobbing
<b>Oxygen saturation</b>	<input type="checkbox"/> Oxygen saturation range normal for your baby, if measured by you at home or by your community team <input type="checkbox"/> Add individual details .....	<input type="checkbox"/> Oxygen saturation above or below the range for your baby on two separate readings, repeated 30 minutes apart	<input type="checkbox"/> If oxygen saturations constantly above or below the normal range for your baby <input type="checkbox"/> Or you are unable to obtain an oxygen saturation reading
<b>Feeding Nappies</b>	<input type="checkbox"/> Baby feeding normally <input type="checkbox"/> Regularly wet and dirty nappies <input type="checkbox"/> Wakes for feeds and is hungry <input type="checkbox"/> No vomits, unless small possets (milk coming back up after feed) are normal <input type="checkbox"/> Weight increasing as expected. (Add individual ..... )	<input type="checkbox"/> Any diarrhoea or vomiting, including vomit after medications or not keeping feeds down <input type="checkbox"/> Struggling to feed, taking longer to feed, breathless on feeding, or sweaty during feeding <input type="checkbox"/> Nappies not wet or dirty (change from usual nappy change times) <input type="checkbox"/> Has lost weight or weight unchanged or more weight increase than expected	<input type="checkbox"/> Vomiting or diarrhoea much more than normal (or twice in a row if this is not normal) <input type="checkbox"/> Cannot feed <input type="checkbox"/> Dry nappies for 4 hours or more <input type="checkbox"/> Blood in nappy
<b>Parent response</b>	<input type="checkbox"/> Parents/ carers happy with their baby	<input type="checkbox"/> Parents / carer concerned about their baby <input type="checkbox"/> There may be no visible changes in the baby condition	<input type="checkbox"/> Extreme worry - Baby very unwell/not breathing <input type="checkbox"/> Parents Start Basic Life Support assessment

Carry on normal care

**Contact for Advice:**  
 Cardiac Ward (evening/night/weekend/bank holidays)  
 Tel: .....  
 Cardiac Nurse Specialist Team (9-5pm)  
 Tel: .....

If you see any of these signs  
 ring 999  
**STRAIGHT AWAY**

280617/CHAT2/Parents version/V7/KG/AD

## Types of Congenital Heart Disease

## Introduction

### Acyanotic (Pink)

Ventricular septal defect (VSD)

Atrial septal defect

Ostium primum defect (also called partial Atrio-ventricular septal defect)

Ostium secundum defect

Sinus venosus defects

Atrio-Ventricular Septal Defect (AVSD)

Patent ductus arteriosus (PDA)

Pulmonary Stenosis (PS)

Coarctation of the aorta

Aortic stenosis (AS)

### Cyanotic (Blue)

Tetralogy of Fallot

Transposition of the Great Arteries

Hypoplastic Left Heart Syndrome

Hypoplastic Right Heart Syndrome

Total Anomalous Pulmonary Venous Drainage

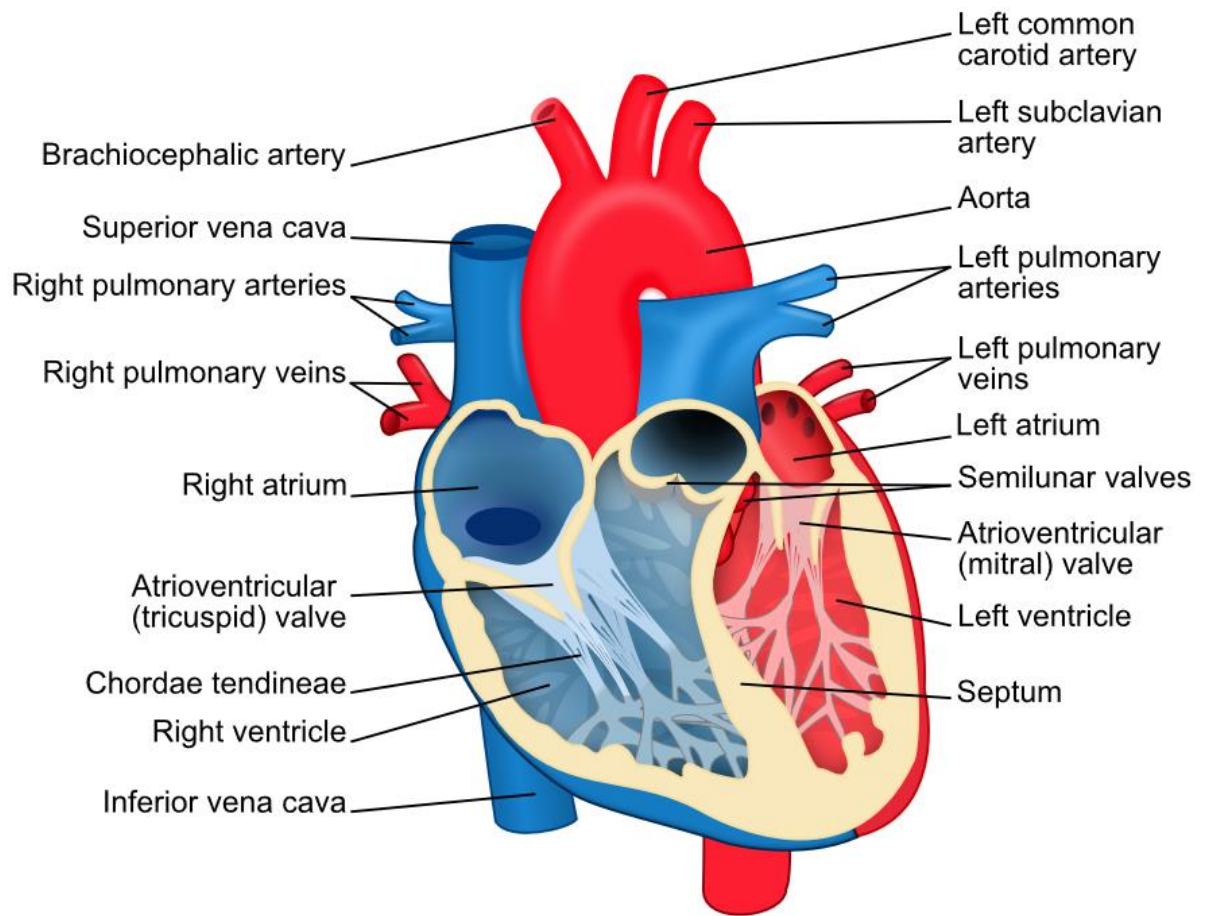
Mixtures of cardiac abnormalities/other cardiac Abnormalities

Endocarditis

## Introduction

Congenital heart disease can involve a problem with the great vessels, septum (walls) between chambers, heart valves or the muscle itself.

## Anatomy of a normal heart



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Congenital heart disease can be classified as either:

- 'acyanotic' (pink), the patient appears pink because their blood is oxygenated in the normal range; or
- 'cyanotic' (blue), the patient appears blue due to low oxygen saturation of the blood in the mucus membranes and skin

80% of congenital cardiac defects consist of eight conditions.

Acyanotic (pink)

Ventricular septal defect (25-30%)

Patent ductus arteriosus (8-10%)

Pulmonary stenosis (6-8%)

Atrial septal defect (6-7%)

Coarctation of the aorta (6-7%)

Aortic stenosis (5-6%)

Ventricular septal defect (VSD)

This is caused by a hole in the ventricular septum (the wall) between the two ventricles.

A VSD is a hole occurring anywhere in the ventricular septum. They can be considered according to size and position of the lesion. Blood flows through the VSD from the left side of the heart into the right side of the heart.

The prognosis of the VSD depends on the size of the defect, location within the septum and whether there is more than one defect present.

The size of the defect is usually described as small, moderate, or large. Small defects cause no symptoms during infancy or childhood and often close spontaneously. Moderate and large defects are less likely to close spontaneously, resulting in excessive blood flow to the lungs, high pulmonary artery pressures, extra work for the heart, and congestive heart failure.

Larger defects may present with symptoms of heart failure (earlier than moderate sized VSD's), breathlessness, feeding difficulties, faltering growth or recurrent chest infections. Children with more significant defects may require medication with diuretics, ACE inhibitors and extra nutritional supplements. These treatments control the symptoms and allow the child to grow bigger prior to surgical VSD closure.

Surgical repair is usually carried out in the first few months of life though occasionally is carried out in older children. The surgery is to reverse the

symptoms of excess pulmonary blood flow and to prevent a very serious long term complication called pulmonary vascular disease.

Larger defects will be repaired surgically. The vast majority of defects repaired surgically will have an excellent result. They have a good long-term prognosis. Occasionally in <1% of cases a pacemaker is required after surgery.

#### Atrial septal defect

An atrial septal defect is a hole occurring anywhere in the atrial septum. They can be considered according to size and position of the lesion. Blood flows through the defect from the left side of the heart into the right side of the heart. Many children with ASDs have no symptoms. If symptomatic then you would expect a reduced exercise tolerance with easy fatigue and more frequent chest infections. The timing of closure of ASDs depends upon the type of ASD and child's symptoms and is usually carried out between 1 and 7 years of age. ASD closure is occasionally carried out in babies less than a year of age if there are significant symptoms.

#### Ostium primum defect (also called partial Atrio-ventricular septal defect)

Primum ASDs (pAVSD) occur low down in the atrial septum and involve valve structure between the atria and the ventricles (AV valves). This can result in deformed or incompetent (leaky) AV valves. Most small defects are asymptomatic (without symptoms). Larger defects can present with recurrent chest infections or heart failure. The timing of surgical correction depends on the clinical progress of the child and the amount of leak on the AV valves.

#### Ostium secundum defect

The secundum ASD is a defect in the centre of the atrial septum and is the most common type of ASD. These are often asymptomatic and discovered incidentally. Complications don't usually present unless untreated into adulthood. Closure by trans-catheter occlusion device or open-heart surgery is recommended where spontaneous closure doesn't occur and the hole is significantly large.

#### Sinus venosus defects

Sinus Venosus ASDs occur high or occasionally low in the atrial septum adjacent to the vena cava. These can be associated with abnormal drainage

of pulmonary veins into the vena cava. Sinus Venosus are corrected in childhood to prevent heart failure in adulthood.

### Atrio-Ventricular Septal Defect (AVSD)

An AVSD = ASD + VSD + Abnormal AV Valves. These are complex lesions and are commonly seen in Down syndrome. There is significantly increased blood flow from the left side of the heart to the right side through both the holes. Infants present with breathlessness, heart failure and usually faltering growth. Diuretics, ACE inhibitors and nutritional support are often required prior to surgery usually between 3-9 months of age.

### Patent ductus arteriosus (PDA)

When a foetus is in the womb the lungs are not in use. The oxygen supply is derived directly from the placenta. Blood bypasses the lungs directly into the left-sided heart circuit via a special duct. This is called the ductus arteriosus. Normally this duct closes after birth. When it fails to close it is abnormal and referred to as a patent ductus arteriosus.

Children with small defects will usually have no symptoms and a normal lifespan.

Treatment is a simple operation or transcatheter closure, which will show immediate improvement in symptoms. Once closed, children have little or no symptoms and should lead a normal life.

### Pulmonary Stenosis (PS)

This abnormality accounts for 7-10% of all congenital cardiac defects.

The pulmonary valve sits between the right ventricle and the main artery carrying deoxygenated blood to the lungs. A stenosis of a valve means it is narrowed and the degree of narrowing can vary from child to child. Most children with PS are asymptomatic but the narrowing can put strain on the right ventricle leading to progressive hypertrophy (muscular enlargement) of the right ventricle and a reduced exercise tolerance. Treatment is warranted where the pressure across the valve reaches a certain level. This is usually



by trans-catheter balloon dilatation although open surgical valvuloplasty may be required.

Mild and moderate cases and cases following balloon or surgical valvotomy will usually have no symptoms and their growth and development should be within normal limits. Moderate stenosis can worsen as the child gets older.

Severe pulmonary stenosis can present in the new-born period with a duct-dependent circulation. As the ductus arteriosus closes there is no longer a route for blood to flow to the lungs to be oxygenated and the neonate becomes very sick and cyanosed. Urgent intervention is required in these cases. The pulmonary valve in these cases may be left with a degree of leak (pulmonary regurgitation) after the valve stretching procedure.

### Coarctation of the aorta

The aorta is the large artery that carries oxygen rich blood from the left ventricle around the body. The word coarctation means narrowing.

Coarctation that is noticed after infancy is not usually associated with significant symptoms. Some children or adolescents may complain of pain in the legs after exercise, but in the majority of cases, even with a severe narrowing, the children are asymptomatic.

Neonates or infants with severe coarctation, are usually diagnosed as a result of heart failure. Coarctation will require intervention. In the neonatal period, this is most commonly done by a surgical repair and in older children, either by balloon dilatation, stenting or surgical repair. They all require life-long follow up as recoarctation or aneurysm formation may occur.

### Aortic stenosis

The aortic valve sits between the left ventricle and the aorta. Oxygen rich blood is pumped out of the high pressure left ventricle, through the aortic valve and then to the brain and the rest of the body. Aortic stenosis means that the valve is too tight, and the resulting blockage can cause a variable obstruction to blood flowing out of the left ventricle and around the body. This can lead to a build-up of pressure within the left ventricle and failure of the left ventricle and then the heart.

Most children with aortic stenosis are asymptomatic, but the narrowing can put strain on the left ventricle leading to progressive hypertrophy (muscular

enlargement) of the left ventricle, a reduced exercise tolerance, syncope (fainting) or chest pain on exertion. Regular clinical and echocardiographic assessments are required to decide when intervention is required. Treatment is warranted where the pressure across the valve reaches a certain level. This may be by trans-catheter balloon dilatation or open valvuloplasty surgery. Surgery is generally delayed to an older age where possible.

Critical aortic stenosis can present in the neonatal period with heart failure and duct dependent systemic circulation. As the ductus arteriosus closes there is no longer a route for the oxygenated blood to flow from the heart to the body and the neonate becomes very sick with circulatory collapse.

Cyanotic (Blue)

Tetralogy of Fallot (6-7%)

Transposition of great arteries (5-6%)

Tetralogy of Fallot

There are four parts to these defects. The main two are a large ventricular septal defect and a narrowing of the blood vessel leading from the heart to the lungs. The right sided ventricular muscle thickens and the main blood vessel from the left side of the heart to the body connects to both ventricles because of the large ventricular septal defect.

Presentation and symptoms depend on the degree of right heart outflow obstruction. When this is severe deoxygenated blood passes from the right heart through the VSD into the aorta causing cyanosis.

Children with this heart abnormality will have surgical correction performed. The timing of this is dependent on the precise anatomy and on the child's symptoms. Children with severe cyanosis may undergo a stent of the right heart narrowing or BT (Blalock-Taussing) shunt prior to the complete repair.

Surgical repair is required and is a complex open-heart procedure. Surgery improves the heart dramatically and many children will be asymptomatic or have slightly reduced exercise tolerance as their only symptom. However residual problems can remain after surgery which can become more serious over time. They will be carefully followed up to detect these early. Problems include residual pulmonary valve narrowing causing an increase in the right heart pressures and the leak on the pulmonary valve often created as part of the surgical repair can become more severe and cause the right heart to

enlarge. Arrhythmias may develop. Any of these problems may cause symptoms and require drug and or surgical treatment.

### Transposition of the Great Arteries

In this condition the main arteries are in a swapped position resulting in two parallel circulations, where oxygenated blood is pumped to and from the lungs and deoxygenated blood is pumped to and from the body. This is not compatible with life unless there is a connection such as a patent ductus arteriosus, ventricular septal defect or atrial septal defect between the right and left side of the heart. As the duct closes, babies can become very sick and cyanosed and require urgent treatment with a prostaglandin infusion to keep the duct open. This is followed by a catheter procedure called balloon atrial septostomy and then finally definitive surgical correction called an 'Arterial Switch' in the first few weeks of life. Further balloon procedures may be required to the branch pulmonary arteries in later life.

Most children have an excellent result from the procedure and are asymptomatic although clinical manifestations occur related to any residual or progressive lesions. This is another condition where symptoms may reoccur later in life and require further treatment. Careful follow up is required.

### Hypoplastic Left Heart Syndrome

The aorta and left ventricle are under-developed leading to restricted or reduced flow of oxygenated blood around the body. Depending on the severity of the defect, babies have a variable degree of cyanosis at birth and will normally die within days without surgery. Reported survival in some series at 5 years of age is around 58%.

A full correction is not achievable, and a series of staged surgeries are performed whereby the developed side of the heart is utilised as a single ventricle system to pump blood around the body. Blood flow is directed from the vena cava (the main large vein that brings blood into the heart from the body) to the pulmonary arteries and on to the lungs bypassing the heart. In Hypoplastic Left Heart this first stage operation is called a Norwood procedure. After this, a second stage operation around 6-12 months called a Glenn shunt is done whereby the superior vena cava is joined to the pulmonary artery (the main artery from the heart to the lungs). Children are still cyanosed after the second stage. The third stage, from 3 years onwards, called the Fontan is where the inferior vena cava is joined to the pulmonary

artery thereby creating a single ventricle pump but is also a correction of cyanotic condition.

Heart transplantation is likely to be required when the Fontan procedure starts to fail.

There will be periods when these children are relatively well with reduced exercise tolerance and periods where they will be becoming symptomatic again as they await further surgery. Overall life expectancy with modern treatment is unknown but much reduced.

### Hypoplastic Right Heart Syndrome

The right atrium and ventricle are underdeveloped leading to reduced flow of blood through the lungs. The presentation will depend on the precise anatomy with some becoming blue or cyanosed as a baby requiring early surgery (shunt or ductal stent) shortly after birth, others having a more balanced circulation and others requiring a pulmonary artery band to limit excess blood flow to the lungs. Following the initial surgery, a Glenn shunt is carried out at around 3 months- 12 months of age followed a few years later by a Fontan. The outcome following Fontan is similar to hypoplastic left heart syndrome. Overall life expectancy with modern treatment is unknown although best estimates suggest that 75% are still alive 25 years after Fontan completion.

Children with a Fontan circulation following surgery for hypoplastic left heart or hypoplastic right heart syndrome often have reduced exercise tolerance

### Total Anomalous Pulmonary Venous Drainage

Normally pulmonary veins return oxygen rich blood from the lungs directly into the left side of the heart from where it is pumped to the brain and around the rest of the body. In totally anomalous pulmonary venous drainage (TAPVD), the pulmonary veins communicate with the systemic

circulation. This mixes oxygen rich blood with the deoxygenated blood returning to the heart after it has passed through the body.

The reduced level of oxygen rich blood causes the symptoms associated with this condition: breathlessness, feeding difficulties.

The timing of surgical correction depends on where the anomalous veins drain to and if the veins are obstructed. Surgical correction of obstructed TAPCD is an emergency while those with unobstructed TAPVD are operated on in the first few weeks to months of life. Most do very well with near normal exercise tolerance and are very unlikely to require more surgery. Occasional patients develop narrowing of the pulmonary veins after surgery and become symptomatic and require further intervention.

Mixtures of cardiac abnormalities and associations with non-cardiac abnormalities

Only the most common cardiac problems have been described here, there will be many children whose condition is not covered in the guidance.

Cardiac abnormalities often occur in association with each other (for example ventricular septal defect with coarctation of the aorta) and the various combinations of abnormality complicate each other, and the treatment required.

There are many fewer common forms of congenital heart disease with variable symptoms. In addition, congenital heart disease often co-exists with other abnormalities (for example syndromes, gastrointestinal abnormalities, renal disease, neurological abnormalities) which need to be taken into account as part of any assessment of disability and needs associated. In these instances, the HCP can be contacted for advice.

Endocarditis

This is a bacterial infection on the lining of the heart and heart valves. Many of those with congenital heart disease are at increased risk of developing endocarditis. Those at particular risk are those with an artificial valve, cyanosed patients and those who have had a previous episode of endocarditis. Precautions against this happening may need to be taken when a child with a heart defect has a procedure that releases bacteria into the blood

stream. This includes some dental work or urological procedures, for example, having a catheter inserted.

The current guidelines recommend that those at high risk of endocarditis should be given antibiotic prophylaxis to cover some dental and urological procedures.

There are no care needs associated with this risk.

### Causes and associations of Congenital Heart Disease

The majority of congenital heart disease has no identified cause. It is, instead felt to be due to the interaction of multiple genetic abnormalities as well as environmental exposures of the child during pregnancy. It is estimated that at least 15% of cases of congenital heart disease are caused by genetic factors.

There are some specific genetic conditions that have an association with congenital heart disease.

#### Down Syndrome

See: Down syndrome for guidance.

#### DiGeorge/22q 11 deletion syndrome

A genetic syndrome with a range of features including characteristic facial features including cleft lip and palate, developmental delay, learning and behaviour problems, feeding difficulties, hypoparathyroidism, immune system abnormalities and congenital heart disease. Presentation can be at any age depending on the severity of the condition. The most common congenital heart defects are tetralogy of Fallot, pulmonary atresia with ventricular septal defect, truncus arteriosus, interrupted aortic arch and isolated ventricular septal defect. Congenital heart disease surgery is undertaken in the usual

way although these children may have additional needs in addition to those related to the CHD.

## Turners Syndrome

This is a congenital condition that is caused by abnormal sex chromosomes.

Approximately one third of girls with Turner Syndrome will have an associated congenital heart defect. The two most common are aortic stenosis or coarctation of the aorta.

Intelligence is normal and life expectancy is near normal.

## Maternal exposures during Pregnancy

### Medication

Phenytoin

Lithium

Warfarin

Thalidomide (Celgene)

Isotretinoin (Roaccutane)

This may lead to structural or electrical disturbances of the heart in some children exposed in utero.

### Alcohol

Commonly ventricular or atrial septal defects.

### Infections

Rubella (associated with patent ductus, ventricular septal defect and coarctation of the aorta)

### Maternal Illness

- Systemic Lupus Erythematosus
- Diabetes

## What you need to know about Cystic fibrosis

### What is Cystic fibrosis?

Cystic fibrosis is the commonest life limiting inherited disease in the UK.

- [Cystic fibrosis](#)

### What are the effects and signs?

The main effects are on the lung, but other organs are often affected as well. ....

- [Effects of Cystic fibrosis](#)

### How is it assessed?

All newborns are screened for cystic fibrosis using a heel prick blood test when they are a few days old .....

- [Assessment of Cystic fibrosis](#)

### How is it treated and managed?

Children will begin their treatment as soon as they are diagnosed .....

- [Treatment for Cystic fibrosis](#)

### What evidence is available?

The best source of evidence will be the specialist nurse or consultant paediatrician and the cystic fibrosis care centre .....

- [Evidence](#)

### How long will the needs last?

- [Prognosis and duration of the award.](#)

## What is Cystic Fibrosis (CF)?

Cystic fibrosis, often abbreviated to CF, is the commonest life limiting inherited disease in the UK. The condition affects the production of mucus in all organs of the body that secrete mucus. Mucus is thicker and stickier than normal. The abnormal mucus primarily affects the function of the lungs, the state of the lungs determines disability and survival in most cases.



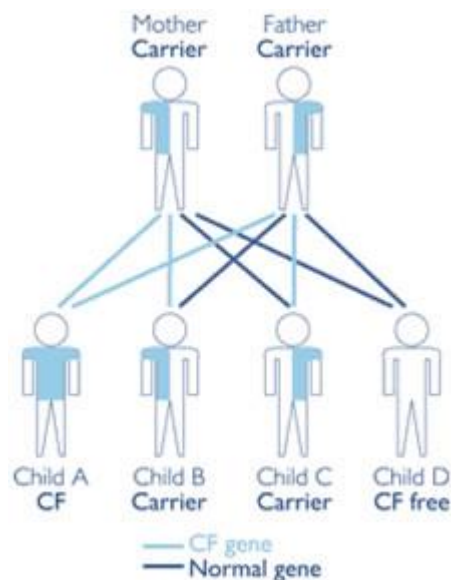
Respiratory failure due to progressive lung disease is the usual cause of death in cystic fibrosis.

The gut is the second most common organ to be significantly affected. The effects on the gut are life limiting and the second most common cause of disabling effects and care that is required in this condition.

Cystic fibrosis is a recessively inherited genetic condition. Genes consist of two pairs of alleles; one allele is inherited from each parent. A dominant gene is always expressed, and a recessive gene is only expressed if both halves of the pair are recessive.

The gene affected in cystic fibrosis encodes for a protein called Cystic Fibrosis Transmembrane Conductance Regulator (CFTR) that controls the movement of salt across membranes in the body. In the UK about 1 in 25 people carry an abnormal recessive form of this gene. They are not affected by cystic fibrosis because the other dominant gene is expressed. The abnormal trait is expressed only if both copies are abnormal. If 2 carriers have a baby together their child can inherit the faulty copy of the gene from each parent. If the child has two abnormal copies of the gene and no normal copy to compensate, he/she will have cystic fibrosis. Each child of parents who are carriers has a 1 in 4 chance of cystic fibrosis.

Image reproduced with kind permission of the Cystic Fibrosis Trust illustrates this:



### **What is the incidence and prevalence of cystic fibrosis?**

Cystic fibrosis is the commonest life limiting inherited disease in the UK. The incidence is 1 in every 2500 live births, 5 babies are born each week

with cystic fibrosis. There are 2 million carriers of the cystic fibrosis gene in the UK and more than 8000 people with cystic fibrosis.

With improved health care the demographic profile of cystic fibrosis is changing. In 2017 60% of people in the UK who have a diagnosis of CF were aged over 16. Life expectancy in the condition continues to rise, with death rates in childhood being very low (4 deaths in children under 14 in 2012). The current quoted life expectancy figure for people with cystic fibrosis is 47 years but is expected to continue increasing. Research shows that the treatment burden in people with CF tends to increase as they get older.

### **What are the effects and signs?**

Effects and signs of cystic fibrosis relate to the effects of abnormal mucus production and the effects this has on organs over time. The main effects are on the lung, but other organs are often affected as well. This section of the guidance describes each organ system separately; in every case the lungs will be affected, for some this will be the only or most significant problem. Most children will have lung and gut related problems, and some will have more than this. Older children because they have had the condition for longer are more likely to have severe disabling effects related to their condition than younger children.

### **Lungs and respiratory system**

The lungs are the main organ affected in every case. The state of the lungs will determine how well the child can function and usually be the main contributor to care or mobility problems. In the past children would be tested for and diagnosed with cystic fibrosis in their first years, usually already having developed the chronic respiratory symptoms associated with the condition:

- Cough in an infant under 4 weeks of age
- Wheezing as a baby or toddler
- Recurrent respiratory or chest infections
- Chest infections with unusual organisms
- Breathlessness

UK wide screening has been introduced for newborn babies. Affected babies will now receive prophylactic treatment from birth. Lung damage that would have occurred in the past, prior to diagnosis, hopefully can be minimised by early interventions to clear mucus and prevent repeated chest infections. Such treatment may delay colonisation with organisms such as pseudomonas that are associated with lung damage.

Over time, despite treatment, repeated chest infections occur, and the lungs are damaged by repeated cycles of infection and inflammation. Bronchiectasis (abnormal dilated airways with pooling of mucus) occurs. At some point mucus becomes chronically colonised with bacterial organisms the

most common of which is called *Pseudomonas aeruginosa*. Once this occurs lung function tends to deteriorate more rapidly. Many of the treatments in the treatment section are designed to postpone this event for as long as possible.

The following symptoms are likely:

- Chronic cough, may produce a lot of sputum even when well
- Repeated chest infections
- Repeated chest infections with *pseudomonas*
- Wheezing
- Reduced exercise tolerance
- Loss of appetite and weight loss

Children with cystic fibrosis will have their lung function monitored regularly. Some children may remain relatively well with near normal exercise tolerance whilst others deteriorate, having frequent chest infections, admissions to hospital and progressive lung damage. Once enough lung function is lost oxygen demands of the body cannot be met. At first this may mean the child cannot run about, it may progress to getting out of breath walking slowly, to being unable to walk at all and getting breathless from the oxygen demands of getting dressed or eating a meal. Once a child has reached this point they may be referred to in the medical evidence as having 'end stage lung disease' or 'respiratory failure', such children are likely to be on home oxygen and may be on a waiting list for lung transplant.

## **Nose**

The normal nose produces mucus which works to keep the nose clean and healthy. In cystic fibrosis the mucus is abnormally thick and sticky just like it is elsewhere in the body. Recurrent and chronic sinusitis is common. Nasal polyps commonly occur probably as a result of repeated infections and may need to be removed surgically. The main reasons for surgical removal are the polyps can become very large, affecting growth and development of the face or blocking the nose. Very rarely nasal polyps may erode through the sinus wall.

## **Gut and digestive organs**

Mucus and other secretions are produced throughout the normal digestive tract by the intestines and the digestive organs. These secretions enable

enzymatic digestion of food and absorption of nutrients from the diet. Cystic fibrosis has the following effects on each organ:

### Intestines

The intestines normally secrete a lot of fluid in the upper end and this aids passage of intestinal contents on their journey. At the lower end the gut re-absorbs most of this fluid. In cystic fibrosis fluid secretion is much reduced and gut contents may remain relatively solid, sometimes blocking the bowel completely. When this occurs in newborns it is called 'meconium ileus', it is one of the ways cystic fibrosis was diagnosed at birth in the past (up to 10% affected). Meconium is the medical term for the contents of the bowel at birth which is normally evacuated within a few hours of being born. In meconium ileus the bowels are not evacuated, and the baby is ill with signs of bowel obstruction. Treatment is given in hospital either in the form of enemas or surgery, there are usually no long-term disabling effects.

Recurrent incomplete blockages due to abnormally solid bowel contents may occur for the same reasons later in life and may be called 'distal intestinal obstruction syndrome' or 'meconium equivalent syndrome'. The symptoms are colicky abdominal pain, abdominal bloating and vomiting and failure to open the bowels. It can often be prevented from recurring by attention to the diet and treated by the use of hypertonic enemas when it occurs. Children with thick intestinal contents, prone to sludging and blocking the bowel, are more prone to developing acute bowel obstruction due to any cause and may require emergency surgical management. Complications of cystic fibrosis due to blockage of the intestines are likely to require hospital care and may require long term medical treatment in the form of laxatives and mucolytic agents.

### Pancreas

The pancreas is a gland that secretes acid-neutralising bicarbonate and enzymes into the upper end of the small intestine as acidic stomach contents arrive. The pancreatic digestive enzymes cannot work in an acidic environment. Unfortunately, in cystic fibrosis bicarbonate secretion is reduced and enzymes work less efficiently. Thick mucus may block pancreatic ducts leading to inflammation and pancreatitis damaging the enzyme producing capacity of the pancreas. This leads to the characteristic cystic and fibrosed (scarred) appearance of the pancreas seen in cystic fibrosis that gave the condition its name. In most it is severe and pancreatic insufficiency occurs. This is a clinical syndrome of inadequate production of digestive enzymes and inadequate absorption of calories and nutrients from the diet that

causes growth retardation and vitamin deficiencies even though the diet is adequate. The symptoms of this are:

- Poor weight gain (failure to thrive)
- Frequent greasy offensive stools (steatorrhoea)
- Flatulence
- Colicky abdominal pain after feeding
- Vitamin deficiencies – especially fat-soluble vitamins A, D, E and K

This complication may be managed by pancreatic enzyme supplements taken by mouth and may require medication to reduce the acidity of intestinal contents. In severe cases particularly when the appetite is reduced through chronic illness artificial feeding may be required.

### Cystic Fibrosis Related Diabetes

The pancreas is also an endocrine gland that produces insulin. This function of the pancreas can fail. The loss of insulin producing capacity is usually very gradual in cystic fibrosis and this type of diabetes is called Cystic Fibrosis Related Diabetes (CFRD). There is a long period during which insulin continues to be produced but levels are not quite enough to prevent hyperglycaemia after meals. Affected children are often screened for diabetes with an oral glucose tolerance test yearly once they are 12. About 50% of people with cystic fibrosis will become diabetic by the age of 30. Some children may require an insulin injection just to cover meals; others will require a more standard regimen because they produce no insulin. Blood sugar management may be complicated by artificial feeding, for example overnight gastrostomy feeding. The principles of managing blood sugar are the same in children with cystic fibrosis as they are for children with type 1 diabetes. However dietary advice given to children with CF will differ from that given to children with type 1 diabetes in view of the need for a high calorie diet in CF.

### Liver

The liver has an important role in digestion through the production of secretions into the upper part of the small bowel and it is affected by cystic fibrosis. These secretions are called bile, bile carries breakdown products for excretion and helps to neutralise stomach acid along with pancreatic secretions. The effect is that small bile ducts in the liver that secrete bile may become blocked by thick secretions; this can lead to cirrhosis of the liver. Cirrhosis tends not to cause symptoms until it is at an advanced stage and the liver is failing. Cirrhosis has serious complications including portal hypertension (high blood pressure in the veins draining the gut) and oesophageal varices (dilated veins at the lower end of the oesophagus that can bleed

profusely). Symptoms of advanced cirrhosis are those of bleeding from the gut and liver failure and include lethargy, jaundice, ascites and confusion.

Because the bile is thicker than normal, gallstones are more common and occur at a younger age in cystic fibrosis. Gallstones can usually be removed surgically.

see: [Effects on the lungs and gut](#)

see: [Other physical effects](#)

see: [Care and mobility considerations](#)

### **How is it assessed?**

It is helpful to consider investigations in two ways, those that confirm the diagnosis is cystic fibrosis and those that give information about how the disease is affecting the child.

### **Diagnostic tests**

#### **Screening at birth**

All newborns are screened for cystic fibrosis using a heel prick blood test when they are a few days old. This is sometimes called the Guthrie test. The levels of a protein called immunoreactive trypsinogen (IRT) are abnormally high in babies with cystic fibrosis. The blood is then tested for the commonest CF gene mutations. If 1 or 2 gene mutations are identified and/or if the IRT is high on 2 occasions the diagnosis of CF is suspected (positive screening test). Babies with a positive test will go on to have the sweat test or repeat genetic testing to confirm the diagnosis.

#### **Sweat test**

This test measures the amount of salt in sweat on the skin and is a definitive test for cystic fibrosis.

#### **Genetic testing**

There are more than 1000 different gene defects known to occur in cystic fibrosis. Genetic testing only tests for the commonest of these so, used as a test by itself genetic testing is not a foolproof method of diagnosing cystic

fibrosis. It is most commonly used to enable a family to identify the cystic fibrosis genes in their family and to plan future pregnancies.

Other hospital investigations that may be performed at diagnosis include:

- Bronchoscopy – with biopsy and bronchoalveolar lavage (fluid is squirted into small areas of the lung and recollected for analysis) - to document appearance of airways, assess level of inflammation and detect occult infection (by analysing the lavage fluid). Only used in some centres
- Overnight pH probe – a probe is placed in the throat and stomach overnight to assess for gastro-oesophageal reflux. Persistently refluxing stomach contents can be inhaled and adversely affect the lungs. Only used in some centres
- Other tests may be carried out such as stool elastase to monitor pancreatic function, respiratory samples, chest X-ray

### **Investigations to monitor progress of disease that can be used to confirm disabling effects or care needs**

Children with cystic fibrosis will receive routine monitoring every 1-2 months at a CF clinic as well as having an annual review. They will be seen at least annually by the Regional CF centre team, either at their own clinic or in the CF centre. They may also need urgent review in between routine visits. This section describes the data that is typically collected at the annual review and how it can be used to confirm needs in DLA assessment. The most important information will be that relating to lung function because lung function closely correlates with disability and survival in cystic fibrosis.

### **Information collected at annual review about lung function including test result and their meaning:**

- Record of chest symptoms – e.g. cough, sputum, exercise tolerance, number of exacerbations in the year
- Usual microbiology - organisms found growing in sputum through the year, chronic colonisation with some organisms indicates greater probability of disability or care needs because the organism accelerates lung damage, the main organism is *Pseudomonas aeruginosa*. Colonisation with *Mycobacterium abscessus* is associated with a bad prognosis and significantly increases the burden of care
- Lung function tests – also called spirometry measures FVC (forced vital capacity – maximum volume of air that can be exhaled in a breath), FEV1 (forced expiratory volume in 1 second) amount of air breathed out during first second of forceful

breathing out and a number of other values. FEV1 is the most important. The expected or normal FEV1 for a person can be calculated using their age height and sex and the result expressed as a percentage, for example normal lungs will equal or exceed expected performance and the FEV1 will be 80-100% or more. In children with cystic fibrosis FEV1 can be reduced. Children less than 6-7 years of age may not produce consistent interpretable results. FEV1 values are recorded at every clinic review, when seen in between appointments, before during and after starting new treatments e.g. before and



after intra-venous antibiotics. Results correlate closely with respiratory disability affecting walking as below:

Table - FEV1 measurement and likely disability affecting walking

<b>FEV1 percentage of predicted</b>	<b>Severity</b>	<b>Functional restriction affecting walking</b>
More than 100% or 81-100%	Normal function	Normal walking ability
60-80%	Mild	<ul style="list-style-type: none"> <li>• Not breathless walking on the level</li> <li>• Can walk up 1 flight of stairs without breathlessness</li> <li>• Little or no breathlessness</li> </ul>
40-59%	Moderate	<ul style="list-style-type: none"> <li>• Unable to keep up with others when walking on the level</li> <li>• Breathless climbing 1 flight of stairs</li> <li>• Daily symptoms breathlessness</li> <li>• May be incapacitated by chest infections</li> </ul>
Less than 40%	Very severe	<ul style="list-style-type: none"> <li>• Continual symptoms that interfere with normal activities</li> <li>• Severe limitation of walking ability may be bed bound or house-bound</li> <li>• Unable to climb 1 flight of stairs</li> <li>• Breathless on minimal exertion</li> </ul>

- Pulse oximetry/Oxygen saturation (SpO<sub>2</sub>) – tested using an oxygen probe on the finger, normal reading 97 to 100%. Home oxygen may be used if readings are repeatedly below 90%. Less than 85% is considered severe. Abnormal overnight oxygen saturation monitoring will indicate worsening respiratory function. Other symptoms of respiratory failure include waking with a headache /increased tiredness
- Plethysmography – the child is placed inside a small chamber the size of a telephone booth and breathes in. Changes in pressure inside the sealed booth are measured and from this the Functional Residual Capacity (FRC) and total lung capacity (TC) of the lungs can be measured. These results cannot be used to determine the presence of respiratory disability and is rarely used
- Chest X-ray score results may be used to monitor the changes in X-ray appearances seen over time in cystic fibrosis. This information is provided to aid understanding of the medical evidence that may be supplied, X-ray score results should not be used to assess disability in DLA even though abnormal looking X-rays are associated with disability
- Northern Score – appearance of chest X-rays is scored between 0 (normal looking) and 4 (very severe changes associated with cystic fibrosis). Severity of X-ray changes correlate with FEV<sub>1</sub>
- Brasfield Score – appearance of chest X-ray is scored between 25 (normal looking) and 5 (very severe changes associated with cystic fibrosis), this score of between 5 and 25 can be used to complete the X-ray findings column in the Crispin-Norman score – appearance of chest X-ray is scored between 3 (normal looking) and 22 (very severe changes of CF). It requires a lateral chest X-ray to be performed
- CT or MRI scan of the chest – although not performed at annual review routinely, if done may give information on the degree of lung damage
- Physiotherapy review – techniques used by the child at home are reviewed for effectiveness. Training or modification of home regimen may be advised at the clinic. May be some information from the physiotherapist on the amount of exercise

the child is doing and their exercise tolerance. Information on effectiveness of inhaled medication may be in this section

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### **Information collected at annual review about gut function and nutritional status:**

- Dietary assessment by dietician – likely to include assessment of nutritional needs and advice on supplements required or artificial feeding if required.
- Height, weight and growth velocity
- Body Mass Index (BMI)
- Growth charts are used to assess whether nutrition is adequate and whether nutritional support is required

### **Other complications**

- A range of blood tests will be checked at the annual review.
- Oral glucose tolerance tests may be carried out for older children to identify the early stages of cystic fibrosis related diabetes (CFRD)
- continuous glucose monitoring gives more accurate picture of blood sugar variations during normal activities
- Bone scan (DEXA) – to identify osteoporosis (thinning of bones) this may develop because of malnutrition and or use of steroids

The main investigations will be those monitoring lung function. Progression of disease may be scored using various scoring systems, for example to monitor chest X-ray changes. It is expected that all children will have a minimum dataset collected by their Regional CF centre including scoring

information. This information can be used to confirm disability where care needs are claimed.

### **Indicators for severe functional restriction**

Any one of the following:

- On lung or liver transplant list
- Terminally ill
- On home oxygen or non-invasive ventilation at night or day and night
- FEV1 <40% of expected
- On artificial feeding regimen, e.g. feeding gastrostomy tube
- Requirement for frequent intravenous antibiotics –more than every 3 months

### **How is it treated and managed?**

#### **Lungs and respiratory system**

“The presence of pathogenic organisms in the lower respiratory tract sets up a vicious cycle of infection, inflammation and lung damage which leads to bronchiectasis and ultimately respiratory failure and death. Early treatment of infection is crucial in delaying or halting the cycle” (CF Trust 2009).

The first part of this section describes typical treatment that all children will require in relation to their lungs. It covers twice daily physiotherapy, nebulised treatments, management of exacerbations due to frequent colds experienced throughout childhood. Physiotherapy with or without nebulisers is the least care a child with uncomplicated cystic fibrosis and without pancreatic insufficiency will require. Most children have pancreatic insufficiency as well as their chest disease.

The second part covers the treatment of various additional lung related problems a child may develop as their lung damage progresses such as colonisation of sputum with specific bacteria and the treatment required for these. Children with cystic fibrosis may have asthma or additional problems with their lungs termed ‘small airways disease’. The treatments for these problems will be in addition to routine chest care described in the first

section and may add significantly to the time taken for chest treatments every day. See:

- [Physiotherapy](#)
- [Nebulised treatments](#)
- [Non-nebulised antibiotic treatment](#)
- [Treatments for additional lung related problems](#)
- **[Gut and digestive organs](#)**
- [Pancreatic insufficiency and nutritional support](#)
- [Distal Intestinal Obstructive Syndrome](#)
- [Cystic Fibrosis Related Diabetes \(CFRD\)](#)
- [Liver](#)
- [Care and mobility considerations](#)

**What evidence is available?**

Sources of Evidence	Information Provided	Limitation of Evidence
Claimant/carer	First source of information	May not be objective, does not have specialist knowledge
GPFR	<p>The diagnosis will be known to the child's GP, but they will rarely see them. Shared care arrangements are between the local and regional hospital rather than the GP. The community nurse may see the child regularly. The GP will be able to provide information such as lung function test results only if the hospital has provided those results to the GP. The Cystic Fibrosis Centre where the child attends for their annual review will always be the best source of further information.</p>	<p>The GP may only be able to confirm medication prescribed for children with cystic fibrosis as they usually receive their care via the cystic fibrosis care centre.</p>

Hospital FR	<p>The best source of information on children with cystic fibrosis will be the specialist nurse or the consultant paediatrician at the cystic fibrosis care centre. Each child will have their own cystic fibrosis consultant and nurse. They will be able to provide recent lung function test results and details of medication and physiotherapy required. Records of the annual review summary or findings will be available for every child. This information is ideal objective medical information that can be used to determine what care needs are present in the individual child.</p>	<p>Details of which investigation results can be used to confirm disability affecting walking and care needs typically associated with various treatments are in the guidance. The medical evidence may contain a lot of complex information that needs sifting through.</p>
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### How long will the needs last?

Age at date of claim	Award Period
0 – 13	Award to age 14 (or for 1 year, whichever is the longer) if functional restriction is mild or moderate and needs are identified.
14 - 16	Award to age 16 (or for 1 year, whichever is the longer) - if functional restriction is mild or moderate and needs are identified.
0 - 16	Indefinite award – if severe functional restriction is identified.

Children with cystic fibrosis may develop care needs at any age. Some children with a milder form of cystic fibrosis will remain relatively well on their minimal physiotherapy regimen and pancreatic replacement through

childhood. These children will grow and develop well, maintain reasonable school attendance and gradually learn to manage their condition independently. They may or may not have care needs substantially in excess of a normal child. Children in this mild group will, on most occasions be able to manage their treatment by themselves from the age of 14. If needs are present, awards are recommended to age 14 in the mild group.

A proportion of children will show declining lung function and will require longer sessions of physiotherapy twice daily. Children in this moderate group are likely to be on multiple medications including time consuming nebuliser treatments spaced out around their physiotherapy. These children will have care needs in excess whether a parent or carer carries out the physiotherapy for them or they do it for themselves under supervision. Children will be expected to take an active part in their therapy gradually learning to do it for themselves. After age 14, constant supervision during therapy is not required for most children and they could be expected to do it themselves. Some prompting and encouragement may be required, and the parent/carer is likely to help more during exacerbations when the child is unwell. Children aged over 14 in general who are physically able will want to manage the majority of their own treatment if they are well enough. The exceptions to this will be children with learning difficulties or diagnosed behavioural disorders. If needs are present, awards are recommended to age 14 in the moderate group.

Other children will deteriorate significantly in the teenage years and become more reliant on their parents due to severe functional restriction. Indefinite awards are recommended in the severe functional restriction group.

A small number of children will be severely affected by cystic fibrosis and develop respiratory failure in childhood. These children are likely to have mobility needs and be severely affected by fatigue, making it impossible for them to carry out all of their treatments for themselves reliably every day. These children have a severe functional restriction and may be terminally ill and/or on the lung transplant list. Indefinite awards are recommended for these children.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

## **Cystic fibrosis – effects on the lungs and gut**

### **Lungs**

In normal lungs mucus is produced and coats the airways. The cells lining the airways have small hair like projections from their surfaces called cilia that beat together to move the mucus up from the bottom of the lungs to the mouth. This mucus carries any dust, particles or bacteria that have been breathed into the throat where they can be coughed out or swallowed. This is called the mucociliary escalator and is one of the major ways the lungs



keep themselves clean and free from infective agents. The normal nose also functions like this.

In cystic fibrosis the mucus is too thick for the mucociliary escalator to function, and the mucus blocks off some of the small airways, this is called mucus plugging. This means that instead of moving bacteria out of the lungs the mucus acts as a reservoir within the lungs where bacteria can multiply and cause infection and inflammation. In cystic fibrosis the lungs are normal at birth but repeated cycles of infection and inflammation damage them leading to damage and dilatation of the airways. This is called bronchiectasis. Mucus collects in these abnormal wide areas and further infections result. Mucus in the lungs may become colonised with unusual bacteria that do not cause infections in normal lungs but can in lungs damaged in cystic fibrosis. The nasal passages are also affected and repeated infections and nasal polyps may occur. Much of the extra care and attention in cystic fibrosis is devoted to managing the abnormal mucus thereby preventing and

treating infections. The following terms may be used to describe the effects of cystic fibrosis on the lungs in the medical evidence:

- Bronchiectasis – some of the airways are abnormally dilated (wide), mucus can pool in these areas and act as a reservoir for infection
- Bronchitis – inflammation or infection of the large airways – the bronchi
- Bronchiolitis – inflammation or infection of small airways – the bronchioles
- Pneumonia – acute infection of the lungs
- Atelectasis – the alveoli or ‘air sacs’ are collapsed or filled with fluid
- Haemoptysis – coughing up blood
- Pneumothorax – collapsed lung
- Cor pulmonale – heart failure due to severe lung disease
- Respiratory failure – lungs unable to meet the oxygen demands of the body
- Mucoid impaction of bronchi – plugging of large airways by mucus
- Allergic bronchopulmonary aspergillosis – allergic response to fungal infection of the lungs with toxin producing highly allergenic fungi

## **Gut**

The main effect on the gut is again caused by abnormally thick sticky mucus production but this time affecting the pancreas. The pancreas is an exocrine gland that produces digestive enzymes that breakdown food into absorbable nutrients. These enzymes are vital. In cystic fibrosis thick mucus blocks off pancreatic ducts preventing secretion of enzymes. Blockage of ducts causes inflammation in the pancreas which leads to loss of productive capacity. This lack of digestive enzymes is called pancreatic insufficiency. Without enzymes malabsorption particularly of fats and fat soluble vitamins occurs. This leads to malnutrition even though the diet is adequate. Poor growth and poor weight gain is seen, and vitamin deficiencies develop. Vitamins A, D, E and K vital for healthy bones, blood clotting and immune

defences are affected. In a small number of children with cystic fibrosis the pancreas is not affected.

The pancreas is an endocrine as well as an exocrine gland. Its endocrine function is to produce the hormone insulin that controls blood sugar. Lack of insulin is the cause of diabetes which develops commonly in people with cystic fibrosis.

## **Summary of main effects**

In summary children with cystic fibrosis have normal lung function at birth. This deteriorates over time because of the effects of abnormal mucus production and the tissue damage caused by repeated infections and chronic inflammatory processes particularly in the lungs, and sometimes other organs. Most children have pancreatic insufficiency from birth and the remainder can develop it with time. The treatment of cystic fibrosis is mainly directed towards correction of organ dysfunction and relieving symptoms that result from the disease.

Children with cystic fibrosis vary in terms of the severity of their condition e.g. a few children do not develop pancreatic insufficiency and some children maintain good lung function through childhood. Others are severely affected developing respiratory failure and requiring lung transplant before their 16th birthday. Screening of all newborns is in place throughout the UK with the aim of diagnosing children before lung damage occurs and preserving lung function for as long as possible in affected children.

Cystic fibrosis causes many other problems in the body including liver disease and cystic fibrosis related diabetes. These may add to care needs related to lung and gut complications of cystic fibrosis. As a life-threatening condition diagnosed in early childhood with a life expectancy of 47 years at birth (2017 figure) the condition, even if well controlled, is likely to have significant effects on the child and their family.

## **Other physical effects**

### **Heat stroke and salt depletion**

Sweat is much saltier than normal in cystic fibrosis because the ability to move salt and water across membranes is impaired. This means in hot conditions a child cannot conserve salt and water and they may quickly become salt depleted and dehydrated.

## **Fertility**

Most males with cystic fibrosis are infertile because the vas deferens, a tube that connects the sperm production area in the testicle to the penis is blocked. They can still conceive using assisted reproduction techniques to overcome this. Most females with cystic fibrosis have near normal fertility although they may have difficulty conceiving because of thick cervical

mucus or the disruptive effects of chronic illness especially malnutrition, if present, on ovulation.

Summary diagram effects of cystic fibrosis on the body

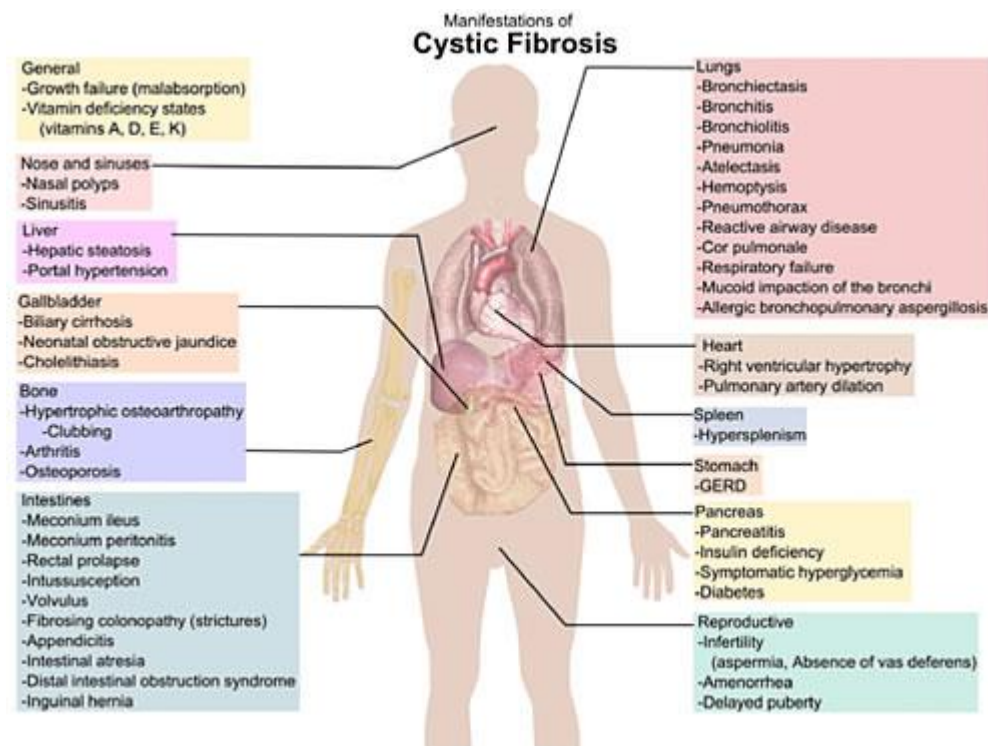


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## Psychological effects

Cystic fibrosis is a genetic disorder that damages many of the body's organs and shortens life expectancy. Currently there is no cure. Consequently, children and their families may need help to adjust psychologically and emotionally. Recent studies indicate that cystic fibrosis patients generally lead active age-appropriate social lives, and that good information and support can reduce negative effects on families.

Treatment regimens can be time consuming, tiring and unpleasant or boring. Some young people may become depressed, and this can affect their ability to learn how to manage their condition for themselves.

Rebellion in the form of non-adherence to treatment for short periods in adolescence is common. Examples of rebellion include not taking vitamins,

treatments or pancreatic enzymes or not doing chest physiotherapy. This is a common phase in growing up with cystic fibrosis for many children. Parents are advised to support and encourage but not to intervene or hover over their children, who are learning to take responsibility for their own medical management and learning the consequences of their actions. More rarely some children with cystic fibrosis display typical rebellions in relation to their cystic fibrosis treatment and have more severe behavioural problems, for example self-harming and using drugs or alcohol. Intervention will be required in these cases. Good adherence is associated with better outcomes. Poor adherence is associated with adolescence and depression.

Young people carrying out much of their routine treatment for themselves will require support from their parents in a variety of ways including prompting, being there, stepping in to look after them during periods of illness, troubleshooting by liaising with primary and secondary care services, e.g. incorrect prescriptions or arranging admission to hospital.

## **Care and mobility considerations**

### **Care**

Care needs in children with cystic fibrosis relate to the amount and complexity of medical management of their condition. Older children are likely to require more complex and time-consuming treatments as lung damage progresses due to repeated infections. Older children may carry out much of their care themselves but may require prompting and close supervision. Care of their chronic illness together with other life changes i.e. adolescence, school changes, examinations can be over whelming for the young person. Close supervision of treatment by a parent may extend beyond 16.

For all children the main component of care will be related to chest disease. Care needs relate to twice daily physiotherapy, inhaled, nebulised and oral drug treatments. Most children will have pancreatic insufficiency requiring enzyme replacement therapy with meals and snacks and some may have additional care needs related to artificial feeding. Some children may have needs related to both chest disease, pancreatic insufficiency and other complications of cystic fibrosis such as cystic fibrosis related diabetes. Chest disease and dietary management are the most time-consuming aspects of care and should be the focus of needs assessment.

The extra care required can make it difficult to get out of the house in time to attend groups and parents of non-CF children do not all ways understand because people with CF usually look well. This can leave a family feeling isolated.

## Lung and respiratory system

Mildly affected babies and very young children are unlikely to have significant care needs in excess of a normal child although these babies do require extra care. They will be on prophylactic oral antibiotics and need chest physiotherapy.

Some babies may need changes of oral antibiotics, pancreatic enzyme supplements and high calorie supplementary feeding to keep them well. The aim is to closely monitor and respond promptly to slight changes in their baby's condition in order to keep their baby's lungs free from changes due to infection. Such changes include development of a cough, changes in appetite, changes in bowel movements and lethargy. The parent carer will have been taught how to assess their baby's chest and to how to carry out physiotherapy at diagnosis.

Care needs often develop over time as a result of repeated chest infections. The main component of care in the early years is physiotherapy. Children also need appropriate antibiotic prophylaxis, salt supplements, vitamin A, D & E, plus a second antibiotic as soon as there is a slight change in symptoms. Parents need to make sure all medicines are taken. This is difficult when babies dislike the taste and spit them out.

Initially they have 2 weekly appointments, then monthly for the 1<sup>st</sup> year and then every 2 months and also in between when necessary. They may travel a long way to a regional centre for some of their care.

Days out need to be planned to ensure there is time for physiotherapy and all of the medications. Ensuring enzymes and appropriate feeds/snack are available. Everything is more time consuming and fiddly away from home.

When going on holiday an adequate supply of everything needs to be ordered in advance, there needs to be thought as to where holidays are taken, adequate travel insurance for example.

Parents are advised to avoid people with colds/coughs/runny noses this may mean cancelling family events.

The role of physiotherapy is to remove mucus from the chest; the amount of mucus produced is variable between children. Babies and young children usually do physiotherapy every day from diagnosis, rarely once a day usually twice daily. This takes around 20mins each time if they don't object and wriggle too much. Physiotherapy is usually done about an hour before feeds to lessen the possibility of vomiting.

For school age children the minimum is around 15 minutes twice a day, duration and frequency are variable depending upon the mood of the child and how well they are, if they are coughing, have thick secretions or are wheezy it will take longer. Specific physiotherapy techniques and the use of positive

expiratory pressure (PEP) devices do not indicate prolonged physiotherapy is required. As lung function deteriorates physiotherapy takes longer. Care needs are likely if nebulised drug treatments are used. These have to be spaced out around the physiotherapy. Use of these drugs indicates that lung complications have occurred such as:

- Colonisation with bacteria such as *Pseudomonas aeruginosa*
- Difficulties expectorating sputum
- FEV1 is 70% or less of normal or evidence of small airways obstruction

These problems often mean that more than 15 minutes of physiotherapy is prescribed. It is convenient to set aside a time twice a day to do everything at once particularly if several drugs have to be given spaced around physiotherapy. Some nebulised drugs must be given at least an hour before physiotherapy, some just before and some are best given when the lungs are clear. The nebulised drug/ physiotherapy routine is a significant time commitment each day and may take up to an hour and a half. Children are encouraged to learn about and do their own physiotherapy and nebulisers under close supervision from the age of 8. The need for prompting and supervision reduces over the years and children with mild disease could be expected to manage their nebulisers and physiotherapy substantially themselves from the age of 14. Some young people require close supervision beyond age 16 and some will require supervision and help because of the severity of their condition. Even when close supervision is not required parents are usually still involved and provide encouragement and prompting.

Note that nebulised drugs cannot be mixed together, and they may not be given at the same time. Depending on the brand of nebuliser and mood of the child each nebulised drug may take 3-4 minutes or 15 minutes to give twice a day for example:

- rhDNase Pulmozyme) –at least 1 hour prior to physiotherapy
- Bronchodilator (Salbutamol) – 10-15 minutes prior
- Hypertonic saline – immediately prior
- Steroids and antibiotics - after physiotherapy

Children will most likely also take regular oral antibiotics to prevent infection, these do not add significantly to care needs. During exacerbations children may take intravenous antibiotic treatment, this is used intermittently for 2-week courses and therefore does not add to care needs on an average day. If long term antibiotic treatment is required these will be given as nebulisers or orally. The exception to this is in end stage disease, IV antibiotics may be given at home for a 3-month course. This adds significantly to care

needs as doses are typically 3 times a day and must be given at regular intervals e.g. 8 hourly at 0800, 1600 and 0000.

The parent /carer will monitor their child carefully for small changes in secretions, cough and general health in order to catch any exacerbations early. Care needs increase during chest infections in all children whether mildly or severely affected.

### Asthma and severe small airways disease

Mild asthma is common and does not significantly increase care needs because prophylactic treatment takes minimal time to administer but nevertheless these are additional treatments to an already busy treatment regimen. Asthma on step 4 or step 5 of the asthma guideline or a diagnosis of severe small airways disease are likely to significantly increase care needs because the time spent on physiotherapy increases and medication needs are complex. The use of nebulised treatments significantly adds to the time commitment.

### Home oxygen

Home oxygen is given at night via concentrator. Home oxygen does not indicate night needs as the parent does not need to stay awake to monitor oxygen levels. However, it does indicate severe chest disease. Such children are likely to have a time-consuming regimen to follow during the day including more than 15 minutes of physiotherapy and time-consuming nebulised drugs. Even if they are over 16 fatigue and breathlessness may mean that they need help from a parent/carer with all aspects of this. Children in the home oxygen group are likely to have mobility needs.

### Lung transplant

Lung transplant is not a cure for cystic fibrosis. However, if transplant is successful walking ability, fatigue and breathlessness can be expected to significantly improve for some years. After successful transplant care in terms of physiotherapy and medication is required but often a young person will be able to do the majority, if not all of this for themselves. Reassessment of needs after lung transplant should be delayed until after rehabilitation. It takes time to recover from surgery, settle down on anti-rejection drugs and build physical stamina. At 3-6 months post-transplant longer term prognosis of disability should be clear although there will be a subsequent gradual decline in lung function due to chronic rejection.

### Gut and digestive organs

#### Babies

Enzyme replacement is required for nearly all babies with CF, it is fiddly to mix the enzymes with a spoonful of milk and give it to the baby at the beginning of a feed. It is difficult to get enzymes levels right as it depends on the individual baby and how much milk they take. It is more difficult for breast feeding babies as the amount of milk consumed cannot be measured. This means malabsorption is not always well controlled. Breast or bottle-fed



babies struggling to gain weight may need additional standard or nutritionally enhanced formula milk after feeds.

Some babies also need Gaviscon after feeds if vomiting frequently. Stools need to be monitored for frequency, colour, odour and consistency to assess the correct dose of enzyme replacement. Increased amounts of vomiting and loose stools mean more time is spent cleaning and washing these babies and their clothes.

## Children

90% of children have pancreatic insufficiency which is treated with pancreatic enzyme replacement therapy. Enzymes are taken whenever food is eaten and this can prolong mealtimes. Salt tablets, vitamin supplements and sometimes dietary supplements are also taken daily by mouth. Enzyme doses have to be adjusted depending upon the fat content of the meal. It is important to get the doses as correct as possible to ensure maximum utilisation of calories and nutrients, which maximises growth and protects against infection. Too much Creon can result in abdominal pain, constipation and an obstruction; too little Creon results in increased bowel action, abdominal pain and weight loss.

Nutrition is very important; children should have 3 meals and 1-3 snacks daily together with their enzyme replacement. Good nutrition improves life expectancy and quality of life.

It is very difficult for some children to achieve this, and very often fortified drinks are included to ensure sufficient calories are eaten each day. Daily encouragement to eat enough may be hard work and an endless battle.

## Artificial feeding

Care needs in excess are likely when artificial feeding is used. Typically, feeds are run overnight though a gastrostomy or PEG feeding tube. The tube and tube site needs to be kept clean and some tubes need turning every day. This takes a few minutes and can be done at bath time. The tube may need to be dressed during the day if the port site is infected.

The feeds are set up at bedtime, some feeds require mixing and preparing and others are ready made and can simply be attached to run through the feed pump. Monitoring during the night is not required and older children e.g. 8 and over should be able to take the feed pump to the bathroom if they go to the toilet during the night. Parents will need to disconnect the feeding tube in the morning and make sure the tube is clean and securely closed for the day.

Children over 14 will be able to set up and disconnect their feeds themselves and care for the tube site. Children over 14 and young adults with

poor lung function are likely to require help with some aspects of artificial feeding due to fatigue, e.g. mixing feeds.

Feeding pumps are very sensitive and alarm during the night if child turns over a lot in their sleep. This may well result in extra time getting the child back to sleep by the parent/carer.

### Cystic Fibrosis Related Diabetes

Separate guidance on diabetes and insulin treatment is provided. The child with cystic fibrosis will have multiple things to think about, understand and take responsibility for as they learn to manage their condition. In addition to monitoring blood sugar and calculating insulin requirements they will also need to manage their pancreatic enzyme replacement therapy at mealtimes and remember to take vitamins, antibiotics and possibly dietary supplements. Artificial overnight feeding may make their diabetes more complicated to manage. Care needs are additive.

### Liver disease

Liver disease does not give rise to disabling effects until liver failure has occurred. Liver failure causes fatigue. Walking ability and ability to manage physiotherapy may be reduced.

### **Mobility**

Mobility needs only arise when severe lung damage and respiratory failure are present. This means that the lungs cannot meet the oxygen demands of the body. FEV1 will be 40% or less and the child may be on the waiting list for a lung transplant.

### **Physiotherapy**

The main problem for the lungs is the presence of increased amounts of sticky mucus that are prone to becoming colonised with bacteria. Although the lungs are normal at birth repeated infections and inflammation in the airways damages the lungs over time. The role of physiotherapy is to remove as much of this mucus as possible either through mechanical means (physiotherapy techniques) or in combination with nebulised treatments to loosen mucus enabling easy expectoration (coughing up phlegm). Techniques used by the child at home are reviewed at each clinic visit. The parents/patient's techniques are assessed, advice is given on exercise, advice on pelvic floor

management and incontinence management is also given, this becomes a problem due to frequent episodes of coughing.

## **Babies**

Many babies appear completely well with no chest symptoms, others have problems from the beginning. In either case the first things parents learn is how to assess the chest and how to carry out physiotherapy. Most babies will require physiotherapy twice every day, some only when they have a cold or a cough.

### **What physiotherapy techniques can be used in babies?**

1. Postural drainage – uses the force of gravity to drain secretions from the chest. The baby is placed in different positions to help secretions drain from different areas of the chest. Usually there are 4 positions which the baby is placed in for a few minutes at a time. This can be done on a pillow on a parent's lap and takes about 15 minutes. More frequent or longer treatments are given when the baby is unwell. All babies are treated flat; 'tipping' techniques are no longer used
2. Percussion or chest clapping is often combined with postural drainage positions. The baby's chest is clapped repeatedly with a cupped hand. The baby remains lightly clothed. They usually enjoy the attention, and it is not painful
3. Positive Expiratory Pressure (PEP) mask. A face mask is used
4. Assisted autogenic drainage – uses deep and shallow breathing in sequences so aid movement of mucus from the small airways to the large
5. Some babies may be on nebulised drug treatments

## **Toddlers**

Children aged 2 to 3 years can co-operate with and learn techniques for themselves gradually taking a more active role in their own physiotherapy. For example, they can learn huffing and play blowing games e.g. blowing bubbles under water (bubble PEP). Most children will require two 15-minute sessions of physiotherapy, and time taken will vary depending on mood and co-cooperativeness of the child, whether they cough up secretions or vomit

with coughing. Physiotherapy may seem like an endless battle for some at this age. Some children will have begun nebulised treatments at this age.

## **Older children**

Older children can co-operate with physiotherapy and will gradually learn to manage their therapy for themselves. Typically, children are encouraged to do their treatment independently with help and supervision from the parent carer in the same room from age 8. Taking responsibility for their own treatment and ensuring it is done is the next milestone and this might be achieved by age 14. For example, a teenager may carry out all of their own physiotherapy except for when they are unwell with an exacerbation. In practice teenagers doing their own treatments without supervision tend to cut corners. Parents may take no active part in treatments at this age but perform an important role prompting and supervising the majority of treatments. A child's ability to manage their physiotherapy will depend on their maturity and their state of health.

The following physiotherapy techniques are commonly used:

- Postural drainage – positioning the body to take advantage of gravity to clear secretions
- Percussion or chest clapping – clapping the chest with a cupped hand to loosen secretions often combined with postural drainage
- Chest shaking/vibrations – shaking or vibrating the chest wall whilst breathing out
- Huff – important airway clearance technique – forced breath out with mouth open as if steaming up a mirror with a breath
- Autogenic drainage – a series of breathing exercises that increase flow of air to move secretions and improve ventilation. There are 3 phases of breathing:
  - Mobilising phase
  - Collecting phase
  - Clearing phase – huff
- High Frequency Chest Wall Oscillation (HFCWO)/vest therapy – an inflatable jacket is worn, this is pressurised and vibrates, so loosening secretions. This is used rarely but may be useful

in patients who have problems with adherence or who have significant disease, as an adjunct to other techniques

- Positive Expiratory Pressure (PEP) – applies back pressure to the airways to prevent them from collapsing when they are empty when breathing out. This can help move secretions and open the airways. There are various small devices that can be used to provide positive pressure. 8 -12 breaths are taken using the device followed by huffs to clear secretions. This is repeated until the chest feels clear. They are called PEP devices
- Oscillating positive expiratory pressure (PEP) devices: These are also small non-electrical devices that contain moving parts that can transmit vibration to the airways and aid in loosening secretions. The devices include:

The flutter

The Acapella

The Cornet

### **Exercise**

Vigorous exercise improves lung function and children will be encouraged to do some every day.

### **Nebulised treatments**

A nebuliser/compressor delivers a mist of drug solution that is inhaled directly into the lungs. It is important that nebulisers are serviced regularly and kept clean so that they deliver drugs as prescribed. Nebulised drugs will usually be planned around physiotherapy sessions. It is convenient to set aside a time in the morning and evening of each day to do everything at once particularly if several drugs have to be given twice a day and physiotherapy has to be done twice a day as well. Some nebulised drugs must be given at least an hour before physiotherapy, some just before and some are best given when the lungs are clear.

Note that nebulised drugs cannot be mixed together, and they may not be given at the same time. Depending on the brand of nebuliser each

nebulised drug may take 3-4 minutes or 15 minutes to give, and the drugs must be spaced out around the physiotherapy twice a day for example:

- rhDNase (Pulmozyme) –at least 1 hour prior to physiotherapy
- Bronchodilator (Salbutamol) – 10-15 minutes prior
- Hypertonic saline – immediately prior
- Steroids and antibiotics - after physiotherapy

Nebulised drugs tend to be given to school age children for whom the minimum recommended daily amount of physiotherapy is 10-15 minutes twice daily. Nebulised treatments are often twice daily also. This makes the nebulised drug/ physiotherapy routine a significant time commitment each day.

Nebulised drugs are given to:

Help clear sputum from the lungs, usually by diluting it to make it easier to cough up.

Antibiotics to prevent or treat infection with organisms that cause infection and lung damage in children with cystic fibrosis.

### **Nebulised treatments to aid clearance of sputum**

- Pulmozyme (rhDNase) is a synthetic enzyme which aims to make sputum thinner making it easier to cough up. It tends to be used when the FEV1 is 70% or less than normal and the response to it is variable. It may be used in children with milder diseases if there is a need for intravenous antibiotics and evidence of small airways disease on lung function. May be used daily or on alternate days and must be taken at least an hour before physiotherapy
- Hypertonic saline is a concentrated salt solution that can be used to encourage coughing up of sputum. For example, in a child who has a rattly cough who produces no sputum. It is given just before physiotherapy, up to twice daily
- Bronchodilator used to counteract bronchoconstricting effect of other nebulised drugs (e.g. salbutamol). This is a bronchodilator used in asthma, it may be used in children without asthma who have cystic fibrosis to counteract the bronchoconstricting

effect of hypertonic saline or other nebulised drug treatments that are being used

### **Nebulised antibiotics**

Are mainly used to treat colonisation with different organisms to prevent episodes of active infection (exacerbations) and reduce long term lung damage due to these. Some typical combinations are listed below -:

- Gentamicin and Colistin (antibiotics, gentamicin and polymixin E)
- Double dose Colistin and Gentamicin (antibiotics, polymixin E and gentamicin)
- Nebulised TOBI (tobramycin an antibiotic) or
- Colistin (polymixin E an antibiotic) or
- Amphotericin (nebulised antifungal used to treat Aspergillosis)
- Amikacin (nebulised antibiotic used for mycobacterium infections)

### **Non-nebulised antibiotic treatment**

#### **Oral antibiotic treatment – prophylaxis**

“All young children with CF identified by newborn screening or diagnosed clinically should be started on anti-staphylococcal antibiotic prophylaxis with Flucloxacillin until 3 years of age” (CF Trust 2009)

Children of any age may receive long term oral antibiotics to prevent infection with common bacteria. Typical drugs include:

- Augmentin duo (syrup) – twice daily
- Augmentin (tablets) – twice daily
- Cotrimoxazole tab/syrup twice daily
- Doxycycline tab once daily
- Flucloxacillin (tablet/syrup) –twice daily
- Azithromycin (tablet) – once daily or three times per week

#### **Oral antibiotics - treatment**

Slight changes in symptoms are treated with additional oral antibiotics. Typical symptoms include:

- Increased / change in / productive cough
- Changes in the appearance / volume of sputum
- Decreased exercise tolerance
- Loss of appetite
- Absence from school/work
- New signs on chest auscultation
- New changes on chest x-ray
- Fever
- Fall in lung function

### **Intravenous antibiotics**

The decision to commence intravenous antibiotics is made jointly with patient/parent/clinician once oral treatments have failed to bring about an improvement. Persisting low grade symptoms such as a cough alone is an indication for IVAB (CF Trust 2009).

There is a low threshold to commence IV treatment. The timing of the intravenous antibiotics can be influenced by life events (exams, holidays), frequency and timing of previous antibiotics, lung function, microbiology results, nutritional state, microbiology results, etc.

Courses of antibiotics are usually started in hospital but if frequent courses are needed children may prefer to have this treatment at home if it is the only reason they are staying in hospital. Lines used are temporary in the hand or arm (including long lines) or permanently indwelling e.g. portacath and courses generally last 2 weeks. Long term daily IV antibiotics are rarely used. Parents are taught how to care for lines, prepare and administer the treatments using Aseptic Non-Touch Technique (ANTT). In some areas disposable infusion systems are used, they are light, do not require batteries and can be worn under clothes. Time taken to set up and connect one of these would be up to 15 minutes per dose. In other areas the parent has to mix the antibiotics and set up the infusion themselves. Typically, the decision to give a course of IV treatment is decided, a date agreed, prescription taken to pharmacy, treatment dispensed, and the parents usually give the



first dose in the department where the line has been inserted. The process takes a whole day if episode is unplanned. Once home the parents have to store all of the treatment and equipment safely, prepare 2 antibiotics to administer three times daily usually at 0800, 1600 and 0000 or times around these to fit in with their other commitments. Each administration can take 45-60 minutes. Time taken depends on equipment provided, type of venous access and experience of the parent/carer. Portacaths require monthly flushing using aseptic non touch techniques whether antibiotic treatment is being given or not.

## **Treatments for additional lung related problems**

### **Coughs and colds**

These are caused by viruses, but antibiotics are always given in cystic fibrosis to counteract the increased risk of secondary bacterial infection. Doses of prophylactic drugs are doubled, or another antibiotic is prescribed such as:

- Augmentin if not taking already
- Azithromycin
- Ciprofloxacin

### **Asthma**

Asthma is common in children with cystic fibrosis, as it is in children without cystic fibrosis. Treatment of asthma follows the British Thoracic Society guidelines. Consult the asthma guidance for details of asthma care requirements if the child has asthma in addition to cystic fibrosis. Dry powder

inhalers are preferred for steroid inhalers and use of spacer devices is encouraged.

### **Severe Small Airways disease/Fixed Airways Obstruction or Bronchiolitis Obliterans**

The terms listed above such as 'severe small airways disease' are used to describe a small number of children with the following ongoing symptoms:

- Little if any sputum production despite large amounts on the chest
- Wheezing
- Tight chest
- Severe obstructive pattern on spirometry testing
- Minimal bronchiectasis on CT scan chest
- Often IgE blood test >500iu/l

It is not known whether these children have a severe form of asthma or something else such as Allergic Bronchopulmonary Aspergillosis. Children

are likely to be fully investigated with bronchoscopy and CT scan. Possible treatments include some of the following:

- Long-acting beta agonists (LABA) e.g. salmeterol or formoterol inhaler
- Symbicort – combination of steroid and formoterol (LABA) inhaler
- High dose steroid inhaler
- Salbutamol or other reliever for symptoms - up to 10 puffs for symptoms
- Montelukast – leukotriene antagonist tablet
- Prednisolone – 2mg per kg body weight per day for 2-3 weeks followed by dose reduction
- IV immunoglobulin – this treatment is given in hospital once a month, there are no known long term side effects although during infusion severe headache and flushing may rarely occur.
- Azithromycin – oral antibiotic treatment
- Subcutaneous terbutaline pump – see asthma guidance for information about this. It is rarely used.
- Methotrexate – this is an immunosuppressant drug given once a week. Blood monitoring is required weekly.
- Itraconazole or Voriconazole – oral antifungal treatment
- IV liposomal amphotericin (Ambisome) – intravenous antifungal treatment given daily over 4-6 weeks.

### **Allergic Bronchopulmonary Aspergillosis (ABPA) or Aspergillosis**

Aspergillus is a fungus that can grow at body temperature in the lungs. The fungus produces toxic and allergenic products. It has the potential to cause serious lung damage with 0.6 to 11% of children with cystic fibrosis affected. The mechanism of lung damage is severe allergic inflammation caused by the presence of Aspergillus and its products in the lungs. The inflammatory response related to the allergy to fungal products is much greater than what

would be seen if the lungs were simply infected with the fungus. This is called Allergic Bronchopulmonary Aspergillosis (ABPA) or Aspergillosis.

Symptoms of aspergillosis include:

- Increased wheezing
- High temperature
- Tiredness
- Thick sputum with brown or black casts

It is treated with:

- Oral prednisolone (steroid) tablets
- Intravenous methylprednisolone
- Antifungal drugs such as Itraconazole or Voriconazole which are taken by mouth and or Amphotericin (nebulised antifungal)
- rhDNAse has also been used

See also the treatment of [severe small airways disease](#) some of which is caused by aspergillosis.

### **Unusual organisms that colonise sputum**

Children will have their sputum cultured regularly at each contact or if there is a change in symptoms to see if organisms are present. This may be referred to in the evidence as positive cultures or microbiology results. Terms used:

- Colonisation – the organism is present in sputum and is not causing symptoms. Colonisation is often treated with antibiotics to prevent infection
- Infection – loose term, may be used interchangeably with colonisation – i.e. the organism is present in the sputum (colonisation) or may be used to indicate that the organism is causing chest symptoms, and the child is unwell

Often both colonisation and infection will require treatment, some of the organisms, once present, indicate ongoing risk of infection with them and life-long antibiotics are prescribed. Often these are oral or nebulised and they can easily be given at home. Use of daily oral and or nebulised antibiotics reduces the risk of acute infections and the need for repeated course of IV antibiotics. IV antibiotics tend to be given as a short course, usually 2 weeks. The treatment can be commenced in hospital for the first couple of days and completed at home; some have the first dose in hospital once the

line is inserted and the remainder done at home. This all depends on their presentation/wellbeing/ parents' willingness and ability to take on the responsibility. Prolonged IV antibiotic treatment at home is very unusual unless there is severe functional restriction or infection with *Mycobacterium abscessus*

Below is a list of organisms that may be found on sputum culture, their significance and their likely treatments:

*Staphylococcus aureus* - Common organism – treated by increasing dose of oral antibiotics. May require intravenous antibiotics

*Haemophilus influenzae* (H influenzae or Hib) - Common organism – treated by increasing dose of oral antibiotics. May require intravenous antibiotics

*Pseudomonas aeruginosa* (*Pseudomonas* or *P aeruginosa*)

Children may be chronically colonised or infected with this. Presence of this organism accelerates lung damage, and it is always aggressively treated in an attempt to eradicate it. If treatment is successful and sputum is clear 're-

eradication' may be tried if it comes back, this means a further course of treatment.

Treatments include:

- Oral ciprofloxacin and
- Nebulised TOBI (tobramycin an antibiotic), or
- Nebulised Colistin (polymixin E an antibiotic), or
- Nebulised Gentamicin and Colistin (antibiotics, gentamicin and polymixin E), or
- 2-week course of intravenous antibiotics

If pseudomonas is persistently present in sputum lifelong nebulised antibiotics are prescribed, these are:

- Gentamicin and Colistin (antibiotics, gentamicin and polymixin E), or
- Double dose Colistin

### **Multi-resistant gram-negative organisms**

Long term oral antibiotics are used to treat these. IV antibiotics may be used.

### **Stenotrophomonas maltophilia**

May not be treated because it is expected to clear by itself or oral antibiotics if child is unwell.

### **Burkholderia cepacia (cepacia)**

This is a potentially life-threatening organism and will be aggressively treated with oral or IV antibiotics possibly at home and for extended periods. Children with this organism cannot be seen in the usual CF clinic and must maintain strict isolation from others with cystic fibrosis if admitted to hospital. This may have psychological implications for the child and their family.

### **Non-tuberculous mycobacteria (NTM)**

These may or may not be treated. Mycobacterium abscessus, one type of NTM has a particularly bad prognosis and will require long term treatment

with 3 or 4 antibiotics over at least a 12–18-month period. Nebulised amikacin or meropenem may be used.

## **Candida**

Candida is the fungus that causes thrush; this can affect the mouth and is treated if it is causing symptoms.

MRSA (Methicillin resistant staphylococcus aureas)

May or may not be treated with oral and or IV antibiotics. But patients harbouring MRSA are seen separately or at the end of a CF clinic and must maintain strict isolation from others with cystic fibrosis if admitted to hospital. This may have psychological implications for the child and their family.

## **Haemoptysis (coughing up blood)**

Small amounts of haemoptysis are common in chronic infective lung conditions. Large bleeds may occur in cystic fibrosis, these will always be treated in hospital. Children prone to recurrent large bleeds may be treated with oral tranexamic acid (a drug that improves blood clotting) and in very severe cases embolisation (occlusion of the bleeding blood vessel) or surgical removal of the affected part of the lung may be carried out.

## **Pneumothorax**

This is an acute condition where air from the lung escapes into the space between the lung and the chest wall and the lung collapses. A Pneumothorax is a marker for severe lung disease in cystic fibrosis. Surgery may be used to prevent recurrence of Pneumothorax, but this type of surgery can make subsequent lung transplant more complicated.

## **Nasal polyps**

These may be annoying and block the nose but in most cases, there are no symptoms. Nasal polyps can impact significantly on quality of life in some

patients. Rhinitis (inflammation in the nose) that leads to polyps can be treated with -:

- Steroid nasal spray or drops e.g. Flixonase or Nasonex
- Surgery – polyps can be removed but they often recur
- Oral steroids may be used in severe cases

### **Sinusitis**

Almost all children with cystic fibrosis have sinusitis but only about 1% have symptoms. Chronic sinusitis is associated with polyps. Treatments include:

- Long term oral antibiotics e.g. for 3-6 weeks
- Surgery – drainage procedures may rarely be used to prevent the build of mucus in the sinuses

### **Home oxygen**

Initially this is almost always used only at night. It is given when overnight oxygen monitoring shows that oxygen saturation levels are less than 90% for more than 5% of the time. There may be associated symptoms including waking with a headache and tiredness. Oxygen cylinders are not used, oxygen concentrators are used which concentrate oxygen from room air so multiple deliveries of oxygen tanks to the home are not required. The concentrator will be maintained under contract during this period. Portable oxygen is used to enable some activities of daily living.

### **Lung transplant**

Obviously, lung transplant is only carried out in end stage respiratory failure. Only a small number of children reach this stage in childhood. Only 6 children received lung transplants in the year to March 2009 and not all of these will have been for cystic fibrosis. Not every child on the transplant list gets a transplant and having new lungs is not a cure for the chronic chest problems associated with cystic fibrosis. After lung transplant many of the problems associated with impaired oxygenation such as fatigue, inability to walk and weight loss will improve dramatically. Children will be able to walk and run around; will feel better and be able to go back to school. However pancreatic insufficiency continues, and enzymes and vitamins need to be taken. Any other non-lung complications will remain the same – e.g. liver disease, diabetes. Diabetes can get worse after transplant. The new lungs will function very well at first but because of chronic colonisation of the sinuses with unusual and drug resistant bacteria infection and deterioration of the new lungs often occurs. A chronic process called obliterative



bronchiolitis occurs which results in a progressive decline in lung function. The regimen of physiotherapy often needs to continue and there may be gradual deterioration of the new lungs despite this. In addition, immunosuppressant drugs to suppress transplant rejection need to be taken every day, these have side effects of their own. One report of survival after lung transplant reported:

- 1 year survival 86%
- 3-year survival 65%
- 5-year survival 50%

Lung transplant is not a cure for cystic fibrosis but can significantly improve mobility needs.

### **Pancreatic insufficiency and nutritional support**

90% of children with cystic fibrosis have pancreatic insufficiency, this means they are unable to extract enough nutrition from their diet without medical help. The level of intervention required is based on assessment of weight for height as a percentage of normal. This table shows what type of nutritional support is required by weight/height assessment, details of treatments

are listed below: Children who are well nourished have better outcomes and better lung function during childhood.

Nutritional state	Children 5-18 years	Medical intervention required	What is involved?
	Weight/height ratio expressed as a percentage of normal		
Normal	Wt/ht is 90 to 110% of normal	Preventative counselling	High calorie, high fat diet.  Pancreatic Enzyme Replacement Therapy (PERT).  Vitamin tablets.  Salt supplement (if necessary).
Underweight	Wt/ht is 85 to 89% of normal  or  weight loss over 4-6 months  or  plateau in weight over 6 months	Dietary supplements.  Consider other investigations glucose monitoring, tighter monitoring of chest symptoms, infection related anorexia, gastro-oesophageal reflux  Maximise enzyme Therapy may require antacid treatment i.e. omeprazole  Test stool for microscopy/faecal fat analysis.  Consider depression and feeding behaviour problems.	High calorie, high fat diet.  Pancreatic Enzyme Replacement Therapy (PERT).  Vitamin tablets.  Salt supplement (if necessary).  Dietary supplements.  Food diary.  Extra snacks.

Very underweight	Supplements tried and Wt/ht is. less than 85% of normal or weight loss by 2 centiles.	Aggressive nutritional support.  As above if not already done.	High calorie, high fat diet.  Pancreatic Enzyme Replacement Therapy (PERT).  Vitamin tablets.  Salt supplement (if necessary).  Dietary supplements.  Artificial feeding with gastrostomy (PEG) tube which involves daily care of PEG site, flushing tube before and after giving the feed, administering feed often through the night using a feeding pump.  Pumps have sensitive alarms therefore often disturbed night sleep.  Often trial of nasogastric feeding (NG) before PEG fitted, which involves all of the above plus testing for PH of stomach contents before feeding.
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### **Preventative counselling - high fat, high calorie healthy diet**

Typical energy needs for a child with cystic fibrosis are higher than for a normal child, in fact up to 150% of normal. This means for every 2 meals a healthy child needs a child with cystic fibrosis needs 3 to maintain normal growth and development. A healthy diet for a child with cystic fibrosis would be unhealthy for a normal child. Children with cystic fibrosis should not eat diet foods, should eat full fat versions of milk and dairy products. Ordinary meals may need to be bulked up with fat and calories by adding extra butter or fats. Homemade milkshakes and high calorie meals and snacks including fast food can be very helpful. As this type of diet is very different to the notion of a healthy diet careful education of the parent/carer and child are required. It is also important to ensure the child is having the recommended

intake of dietary calcium. 90% of children have pancreatic insufficiency and will require the following supplements or treatments:

### **Pancreatic Enzyme Replacement Therapy (PERT)**

Pancreatic enzymes are not produced and so must be taken by mouth every time fat containing foods or drinks are consumed. They should be swallowed prior to the meal and cannot be sprinkled on food. The capsules are large and can either be swallowed whole or opened and the granules mixed with the first mouthful of food. A further dose might be needed during a meal. If not enough PERT is taken symptoms of fat malabsorption including bloating, flatulence and diarrhoea will be present. Most children will take Creon 10,000 capsules and typical doses are listed below: (Approximately one 10,000 unit capsule per Kg of body weight per day but this varies greatly between patients). Babies and toddlers can take creon micro (5,000 units per scoop). This is usually administered on a teaspoon with a drop of milk or puree food.

<b>Age of child</b>	<b>Typical dose of Creon (but variation between patients)</b>
Babies	½ Creon capsule per feed (1/2 Creon per 6-8 g of fat)
Toddlers	2 Creon with meals, 1 or 2 with snacks
Pre-school	2-3 Creon with meals, 1-2 with snacks
School age	4-6 Creon with meals, 2-3 with snacks
Adolescents	5-8 Creon with meals, 2-3 with snacks

### **Brand names of PERT**

Pancreatin is a mixture of enzymes including lipase, protease and amylase used in pancreatic insufficiency. There are several brands that provide pancreatin in different strengths. Any of them may be used although Creon is prescribed most commonly. Brand names include:

- Creon Micro 5000 per scoop Creon 10,000, Creon 25,000, Creon 40,000
- Nutrizym 10, Nutrizym 22
- Pancrex, Pancrex V
- Pancrease HL

### **Salt tablets and vitamins**

Salt depletion may be treated with salt supplements, brand name: Slow Sodium. Chronic salt deficiency may be severe in children with cystic fibrosis and can be a cause of failure to grow – this is called ‘Pseudo-Bartter Syndrome’.

Vitamin supplements (A, D, E, K) – children are likely to take multivitamins such as Abidec or Dalivit that contain vitamins A to D and separate preparations of vitamins E and K (menadiol).

### **Dietary Supplements**

The principle of supplements is to provide concentrated calories in a palatable form. There are many flavours and presentations:

- High energy milk-based sip feeds, e.g. Paediasure Plus, Fortini, Frebini Energy,
- High energy fruit juice-based feeds, e.g. Enlive Plus, Forti-juice, Provide Xtra
- High energy infant milks, e.g. SMA High Energy, Infatrini
- Powdered energy sources, e.g. Maxijul and Caloreen (carbs), Calogen and Liquigen (fats) and a mixture of fats and carbs – Duocal

Pancreatic enzymes need to be taken with most but not all of these. Powdered energy sources require mixing before use and the others can be drunk straight from the carton with a straw. Babies can be fed with a bottle. Children with loss of appetite who struggle to eat meals are likely to struggle with supplements as well.

### **Artificial feeding with gastrostomy (PEG) tube or button gastrostomy**

Children who fail to gain weight despite supplements or who are very underweight will have artificial feeding. Getting their weight up reduces the number of chest exacerbations and improves their ability to recover from these. It can improve well-being and energy levels and may be used over the short

or longer term. A Percutaneous-Endoscopic- Gastrostomy (PEG) tube is an indwelling thin plastic tube from the abdominal wall to the stomach. This is connected to a feed pump overnight and improves calorie intake without reducing appetite for food during the day which can be problem with supplements. Enzymes need to be taken beforehand but children should not be woken during the night to take a further dose of enzymes. Some feeds do not need to be taken with enzymes. Brand names of feeds include:

- Infant feeds – SMA high energy, Infatrini, Neocate, Pregestimil (enzymes not required)
- Older children – Paediasure Plus, Nutrini, Tetrini, Frebini, Emogen (enzymes not required)

### **Distal Intestinal Obstructive Syndrome**

Bowel contents are more solid than is normal because of abnormal mucus production and other factors such as dehydration. Bowel contents sludge up and partially block the terminal ileum causing abdominal pain. This is diagnosed and may require treatment in hospital. Children with recurrent episodes may take medication to prevent recurrence such as:

- Lactulose (laxative)
- Ranitidine (antacid)
- Omeprazole (antacid)
- Fluimucil (N-acetylcysteine)

Constipation is also common for the same reason and may be treated with oral laxatives such as:

- Lactulose
- Movicol

### **Cystic Fibrosis Related Diabetes (CFRD)**

The loss of insulin producing capacity is usually very gradual in cystic fibrosis and this type of diabetes is called Cystic Fibrosis Related Diabetes (CFRD). There is a long period during which insulin continues to be produced but levels are not quite enough to prevent hyperglycaemia after meals. Children are often screened for diabetes with an oral glucose tolerance test yearly once they are 12. Continuous glucose monitoring (for 6 days) is being used more frequently and provides a more detailed report recording blood sugars every 5 minutes, a food diary has to be recorded and

4 blood sugars during the period the monitor is on. About 50% of people with cystic fibrosis will be diabetic by the age of 30. Some children will require an insulin injection just to cover meals; others will require a more standard regimen because they produce no insulin. Blood sugar management may be complicated by artificial feeding, for example overnight gastrostomy feeding.

The principles of managing blood sugar are the same in children with cystic fibrosis as they are for children with type 1 diabetes. An injection regimen or an insulin pump may be used.

Children with cystic fibrosis will not follow a typical diabetic diet they will still be encouraged to eat as much as they can of a high fat high calorie diet. The only change might be to avoid high sugar snacks and drinks between meals and to have them with meals instead.

## **Liver**

Cystic fibrosis related liver disease is common (abnormal biochemical tests and ultrasound scans), but it rarely causes symptoms. Once cirrhosis is present but not causing symptoms treatment may be given to prevent further cirrhotic change, this includes:

- Ursodeoxycholic acid – this drug increases bile flow but can cause diarrhoea as side effect
- Vitamin K

Advanced liver cirrhosis may cause the following complications:

- Jaundice – a yellow discoloration of the skin and eyes that develops because the liver cannot excrete waste products
- Varices – swollen veins, most often found at the bottom of the oesophagus that bleed easily and may cause acute haematemesis (vomiting blood) or melaena (altered blood in the stool).
- Ascites – abdominal swelling due to leakage of fluid into the abdominal cavity always a sign of advanced liver disease
- Encephalopathy – confusion and neurological signs always a sign of advanced liver disease

The effects of advanced liver disease can be managed for a short time but the only hope of return to normal function is liver transplant. It is unlikely that advanced liver disease with ascites and encephalopathy will be



managed at home unless the child cannot or will not have a liver transplant or is terminally ill.

DAMP (Deficits in attention, motor control and perception)

This is an umbrella term used to describe children with ADHD and Dyspraxia. The main disabling impairment is ADHD, use F95 to code the main impairment, use the ADHD guidance to assess the case and determine award duration. You should use F04 to code for Dyspraxia as a 'Secondary'

disability unless you think they have another impairment which is more disabling, for example, Conduct disorder or Autism.

How long will the needs last?

Use the ADHD guidance

Impairment	Award Period
ADHD	As ADHD guidance
Dyspraxia	Seek medical advice on prognosis and award duration if needs claimed.

What you need to know about Depression, suicide and self harm in children and young people

What is Depression?

Depression is a mood disorder in which the main symptoms are sadness, marked .....

### Depression

What are the effects and signs?

In addition to the sad mood, loss of interest in things and .....

### Effects of Depression

How is it assessed?

Depression is a clinical diagnosis so, as such, there are no physical diagnostic tests .....

### Assessment of Depression

How is it treated and managed?

The treatment for a child's depression depends on .....

### Treatment for Depression

What evidence is available?

Symptoms of the depression and any self-harm risk may not be in evidence or be observed in .....

### Evidence

How long will the needs last?

Prognosis and duration of the award

What is Depression?

Depression is a mood disorder in which the main symptoms are:

- sadness
- marked loss of interests or pleasure

At least one of these needs to be present for most days, most of the time for at least two weeks. This must represent a change from a person's baseline mood.

The presence of associated symptoms helps to define the severity of the depression. These are:

- disturbed sleep
- decreased or increased appetite
- fatigue or loss of energy
- agitation or slowing of movements
- poor concentration or indecisiveness
- feelings of worthlessness or excessive guilt
- low self confidence
- suicidal thoughts or acts

Symptoms should be present for at least two weeks and every symptom should be present for most of the day.

Depression is a common disorder with 20% of all of us experiencing an episode of depression at some time in our life.

### **Prevalence**

Depression has a prevalence of 1% to 2% amongst children (aged 5-11) and 6-8% in adolescents (aged 11-18). In children there is a 50:50 split in boys and girls. During adolescents this changes with girls twice as likely as boys to suffer with the condition.

### **What are the effects and signs?**

In addition to the sad mood, loss of interest in things and loss of pleasure/enjoyment there are a number of associated symptoms. These are the most common:

#### **Disturbed sleep**

This can present as either too much sleep (hypersomnia) or insufficient sleep (insomnia). Insomnia is difficulty in initiating or maintaining sleep coupled with early morning awakening.

Sleep disturbance is increased amongst children with depression. Overall incidence of sleep disturbance in depression is 40%.

Amongst children who have severe depression, sleep disturbance is present in up to 70% of cases.

### Decreased or increased appetite

Children with depression can either overeat for comfort or have little appetite and eat little. Eating less food will reduce a child's energy level. This may result in reduced concentration and school performance.

### Fatigue or loss of energy

Children with depression may suffer from fatigue or lethargy. This is a state of tiredness, weariness, fatigue, or lack of energy.

### Agitation or slowing of movements

Slowing of movements is manifested by difficulty in performing physical task for example particularly those requiring sudden changes.

Agitation is an inner restlessness or tension that can be associated with increased movements, for example, leg shaking, being fidgety, pacing or hand wringing.

### Poor concentration or indecisiveness

This can affect the ability of a child to perform academically as well as socially. It is characterised by a slowing of thought processes and an inability to make decisions.

### Feelings of worthlessness or excessive guilt

Feelings of worthlessness or guilt are the first stages of thought leading to suicidal ideation, for example, a child feeling they are worthless can lead to them thinking that as they have no worth they have no right to exist and should die.

### Low self confidence

Low self-confidence can lead to reduced social contact and isolation. In addition it may lead to reduced school performance.

### Self harm or suicidal thoughts or acts

Self-harm, also known as self-injury, is the act of deliberately harming your own body such as cutting or burning yourself. It is often not meant as a

suicide attempt but instead it is used as a way of coping with emotional distress and may be an indicator of underlying depression and anxiety.

Suicidal thoughts concern the possibility of ending their own life and suicidal attempts are the physical actions related to these thoughts.

14% of children with depression admit to thinking about or attempting suicide or self-harm during the last 6 months. This figure rises to 28% when looking at children aged 12 to 18.

#### School performance

10 % of children with depression will have special educational needs (SEN). This is comparable with the rest of the population of children who have a 14% chance of qualifying for SEN.

#### Co-Morbid illnesses

Anxiety: 25% of children with depression will also suffer with generalised anxiety disorder. A further 25% will suffer with social phobia or separation anxiety.

Conduct Disorders: 33% of children with depression will also have a conduct disorder.

Autistic spectrum Disorder: 2% of children suffering with depression will also have autistic spectrum disorder. This represents 20% of all children with autism.

ADHD: 20% of children with depression will have symptoms consistent with ADHD.

#### How is it assessed?

Depression is a clinical diagnosis so as such, there are no physical diagnostic tests. The assessment of children diagnosed with depression will, in many cases, initially be performed by the child's GP.

Children with mild depression may be observed closely for a period of 2-3 months. If no improvements have been observed, then they will be referred on to CAMHS (Child and Adolescent Mental Health Services).

Children with moderate or severe depression will be referred on to CAMHS. CAMHS consist of psychiatrists, psychologists, nurses and other therapists. They are usually community based and take direct referrals from the child's GP but sometimes, they also take referrals from school and directly from the child's parents or from the young people themselves.

CAMHS will interview the child and parents and may observe them and their interactions as a family unit. They may perform behavioural questionnaires such as a Mood and Feelings Questionnaire and will commonly do a Child and Adolescent Psychiatric Assessment. With the family's permission, CAMHS will usually contact the child's school to get additional information

about how the child is performing academically and how they are interacting during their normal school day.

### Severity of Depression

The severity of depression depends on the symptom severity and on the impairment caused to the young person's everyday life. Severity is not measured by counting the number of symptoms. A child/young person may be

severely impaired while only displaying a small number of symptoms in some cases.

Every symptom should be present for most of each day:

- sadness
- marked loss of interests or pleasure
- disturbed sleep
- decreased or increased appetite
- fatigue or loss of energy
- agitation or slowing of movements
- poor concentration or indecisiveness
- feelings of worthlessness or excessive guilt
- low self confidence.
- suicidal thoughts or acts

Not Depressed

Fewer than four symptoms likely to be present. No obvious impairment.

Mild Depression

Four symptoms present. The child will continue to function but, with some difficulty in some areas of their life.

Moderate Depression

5-6 symptoms present. The child with moderate depression experiences considerable difficulty in functioning at school, home and with peers.

Severe Depression

7 or more symptoms present. This can be with or without prominent and distressing psychotic symptoms. Their functioning is significantly affected in all areas of life.

Psychotic symptoms: When these are present this is sometimes referred to as psychotic depression.

This indicates the prominent and distressing presence of at least two of the following for one month: delusions, hallucinations or disorganised speech.



Delusions are thoughts or beliefs that are unlikely to be true, for example, they may become convinced they're to blame for something or that they've committed a crime.

Hallucinations are when a person hears (and in some cases feels, smells, sees or tastes) voices and sounds or sees people or objects that aren't there. A common hallucination is hearing voices.

How is it treated and managed?

The treatment for a child's depression depends on their age and also the severity of their depression.

### Mild Depression

For children or young people with mild depression an initial re-assessment can be arranged for 4 weeks time. This would allow a period of watchful waiting. If the child still has mild depression then they can be referred on to CAHMS for consideration of psychological or behavioural therapy.

### Moderate Depression

Children or young people with moderate depression will be managed by their local CAMHS service.

Initially they will be treated with psychological therapy (Cognitive Behavioural Therapy mainly but, family therapy or psychotherapy may also be used). If after 3 months there is no improvement then combined therapy can be considered with medication and psychological therapy. If the child does not engage or struggles with psychological therapies then medication alone is usually recommended.

### Severe Depression

Children with severe depression are treated with psychological therapies with medication added if there has been no response after some 4-6 sessions of this therapy. Medication is used alone in children who do not engage with psychological therapies.

### Types of psychological therapies

Cognitive behavioural Therapy (CBT). A type of psychotherapy in which negative patterns of thought about the self and the world are challenged in order to alter unwanted behaviour patterns or treat mood disorders such as depression.

Interpersonal therapy is a brief, present-focused psychotherapy that centers on resolving interpersonal problems and decreasing symptoms.

Family therapy is a branch of psychotherapy that works with families to nurture change and development. It tends to view change in terms of the

systems of interaction between family members. It emphasises family relationships as an important factor in psychological health.

Psychotherapy involves talking to a trained therapist, either one-to-one, in a group or with your family. It allows you to look deeper into your problems and worries, and deal with troublesome habits and a wide range of mental disorders.

## Medication

Medication for children or adolescents is prescribed and monitored by CAMHS. Children under 12 will only be prescribed medication for their depression after a multi disciplinary team assessment of their depression and previous treatment.

Fluoxetine: This is a Selective Serotonin Reuptake Inhibitor (SSRI) antidepressant drug which increases the level of serotonin in the brain.

Dose: Start dose is half adult dosage for adolescents (aged 12-18) and weight based for children (aged 5-11). Start dose 10mg once daily. Increased to 20mg once daily after 7 to 14 days.

Side effects:

- Insomnia
- Headache
- Fatigue
- Indigestion
- Dizziness

This is a well tolerated drug and most side effects will settle within the first 10-14 days of taking.

Sertraline: This is another drug which increases serotonin in the brain. This drug is not licensed for use in children so will only be prescribed for severe

depression (usually with suicidal behaviour) after a fair trial of fluoxetine coupled with psychological therapy has failed to show any improvement.

Dose: 25mg for 14 to 28 days. Increased to 50mg after 2-4 weeks.

Side effects:

- Insomnia
- Dizziness
- Tiredness
- Headache
- Nausea
- Diarrhoea

Side effects usually settle over the first month of treatment for the majority of people. Well tolerated. In some children SSRI medication may lead to an increase in suicidal symptoms. Children taking this type of medication need close monitoring.

Treatment Effects

Fluoxetine is an effective and well tolerated treatment for depression in children and adolescents. The treatment benefit improves the older a child is. For all child age groups, if nine children are treated with antidepressants then one child will have their mood return to normal. This number drops to 4 for adolescents but is larger at 21 for children under the age of 11.

Cognitive Behavioural Therapy (CBT) is well tolerated and effective. For every 10 adolescents treated with a CBT, 4 children will improve to normal

mood. This effect is usually still in place for, at least, the following 12 months.

Most episodes of depression in children and adolescents last less than nine months. After one year, 70 to 80 per cent of children will have recovered but 1 in 10 remains persistently depressed and recurrences are common.

What evidence is available?

Sources of evidence	Information Provided	Limitation of Evidence
Parent / Carer	First source of information and associated evidence.	Does not have specialist knowledge.
<b>General Practitioner</b>	Clinical features, treatment and some information about anxiety and needs.  May be the only source of information if no other professional involvement.	Does not have specialist knowledge.  Unlikely to have detailed or recent information about resulting disability or needs.
<b>CAMHS</b>	Clinical features, treatment and information about disability and needs. A Common Assessment Framework (CAF). Suicide/self harm care plans.	Information regarding symptoms and disabling effects may primarily be based on what the parent / carer has told the relevant professional.
<b>School</b>	Information about the child's social interactions and educational performance will help. Recent changes in educational and social performance are especially relevant.	Symptoms of the depression and any self harm risk may not be in evidence or be observed in the school setting.

Social worker (if child has one) / Local Authority	Information for social care needs will be available for some cases. They may have information on previous assessments (including educational).	May not have information about symptoms, investigations and treatment.
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How long will the needs last?

If needs awarded, the following is a suggested guide only. Except in rare cases, children or young people with mild depression are unlikely to have needs in excess of a child without depression.

In some cases, either a child's depressive illness lasts longer than anticipated or they develop a recurrence of this mood disorder within 12 months of the previous one. If this is the case, an award on renewal should be considered with discussion of the case with an HCP.

Approximately 90% of adolescent depressive episodes remit 1-2 years after onset.

The following table is to be followed if an award is made. It is not a statement of intended awards for individual categories.

Age at date of claim	Length of award		
	Mild	Moderate	Severe (with or without psychotic symptoms)
5-16 first award*	Award for 18 months	Award for 18 months	Award for 2 years

**\*Note:** For subsequent awards, if needs persist, award for 3 years.

You must take all information into account when considering the duration of care and mobility needs. You must base the duration of care and mobility needs on the particular circumstances of the child.

Suicide and Self harm

**Definitions:**

- **Suicide** – most contemporary definitions of suicide rely on 2 elements: the outcome (death) and a prerequisite (intention or wish to die) – such

as the World Health Organisation definition (1998) 'For the act of killing oneself to class as suicide, it must be deliberately initiated and performed by the person concerned in the full knowledge, or expectation, of its fatal outcome'

- **Attempted suicide** (previously 'parasuicide') – this term is usually used to denote someone who has failed to die as a result of suicide – it again implies an intent but has a non-fatal outcome
- **Self harm** – the act of injuring oneself on purpose rather than by accident. Some people have differentiated fatal versus nonfatal self harm. It may be by self poisoning, hitting, cutting or burning, pulling hair or picking skin or by self strangulation
- **Suicidal thoughts** – Suicidal thoughts are thoughts a person has about wanting to end their life to be free from unbearable emotional or physical pain. These might be general thoughts about not wanting to be alive, or specific thoughts about how to end one's life. Suicidal thoughts are common and not all of those who have suicidal thoughts will attempt suicide, indeed most people who have suicidal thoughts are able to keep themselves safe

## Introduction

Suicide is a leading cause of death in older adolescents. Families managing children with suicidal thoughts and behaviours will feel under considerable pressure to reduce the risk of harm occurring. Although suicidal thoughts are common in adolescence they should not be dismissed without being carefully examined.

For many children the thoughts will be fleeting, associated with a crisis, and no self-harm or suicide attempt will occur. For other young people there will be clear indicators of risk of suicide or self harm which the family may be managing by providing extra care, supervision and support until their mental health improves. In some families the young person may be experiencing neglect or abuse.

Suicide is the second most common cause of death for young people and yet it is preventable. The UK has one of the highest rates of self-harm in Europe (at 400 episodes per 100 000 population).

Self-harm is common. A survey of young people aged 15–16 years estimated that more than 10% of girls and more than 3% of boys had self-harmed in the previous year. In 2018-19 over 20 000 9-17 year olds attended A+E due to self-harm. The true number of episodes of self-harm will be higher as not all self harm needs hospital treatment, though all self-harm is significant mentally. Self harm increases the risk that the person will eventually die by suicide by between 50 to 100-fold above that for the rest of the population. Self harm is always a signal of distress, and it is important to avoid using stigmatising and invalidating terms like attention-seeking and manipulation.

## Suicide

Amongst adolescents, suicidal thoughts are common. The World Health Organisation (WHO) estimates that as many as 50% of adolescents will have

suicidal thoughts at some point during their adolescence. 17% of them will have persistent suicidal thoughts. The rates for completed suicide are:

- 1-2 per 100,000 children aged 10-14
- 4-8 per 100,000 children aged 15-19
- Male to female: 4:1

The most important risk factors for children who complete suicide are psychiatric illness, previous suicide attempt, family factors including parental mental illness, being abused (including online and exploitation), disorders of gender identity, bullying, academic pressures and, easy access to the means of committing suicide. The role of social media in those who are already vulnerable may be important.

Clearly these factors around the child, their family and their environment are complex and interact with one another.

What is key in terms of assessment is to identify those children at high risk who are receiving additional care and supervision to prevent suicide. For example, a depressed child whose parent has committed suicide who is expressing the wish to join their parent is likely to be at high risk and to require support and supervision and careful management of their environment to minimise suicide attempts.

## **Self harm**

This is an act with a non-fatal outcome. It is defined as the intentional, direct injuring of body tissue. It can be done with or without suicidal intent. However, self-harm is commonly done without any suicidal intent. It is usually done in one of the following ways:

- ingesting medication or a substance (in excess of any prescribed dose)
- swallowing non ingestible items (batteries or razor blades)
- self-inflicted wounds (such as cutting or burning)
- alcohol or drug abuse

About 25% of young people self-harm on one occasion, most commonly by self-cutting. Recurring self-harming is less common, with 9.5% of young people self-harming on more than four occasions.

It is important to note that the level of harm does not correlate with the seriousness of the problem. A child who makes superficial scratches which do not require medical attention may have significant mental health problems and require significant care.

A child who frequently self-harms may need constant supervision, at home and throughout school including lunch breaks, and intensive therapy. They may struggle to leave the house and get to school or other activities. Red flags include the constant carrying of items used to self-harm (note these may

be everyday objects such as a pen lid rather than a blade to cut), frequent expressions of worthlessness and helplessness, social isolation, behavioural changes and sudden changes in school engagement and attainment.

### **Suicide/Self harm care plan**

Care plans are put in place, usually by CAMHS, after a suicide attempt or episode of self-harm. They are designed to help:

- identify and deal with the stresses or disorders, such as depression, which may be at the root of the suicide attempt or self-harm episode
- prevent repetition or escalation of self-harm
- reduce or stop self-harm
- reduce or stop other risk related behaviour
- improve social, school or occupational functioning
- improve quality of life

They will usually include some form of suicide risk/self-harm management.

The needs of these children with suicidal thoughts and those who deliberately self-harm are likely to be increased. They may require ongoing therapy from professionals, but even those that do not will require increased monitoring from parents to assess and mitigate their risk of further harm. The care plan will be a very helpful piece of evidence when considering the additional care needs.

Parents will usually be asked to limit access to anything the child may use to self-harm, for example, blades, locking away medications. Extra care needs will vary from child to child and the presence of the extra care may need to be continual. For example, a child may not have self harmed for some time because their school attendance has been reduced to mornings only to decrease anxiety but if this was stopped, they would be at high risk of self harming behaviour again. Children/adolescents may need more supervision than those of the same age to spot problems and avoid self-harming behaviour.

Children who have problems in school are likely to have a plan in place to reduce risk. An example is being allowed time out from lessons when they need with a prearranged signal to the teacher. Other examples range from children who use breathing techniques within the classroom to reduce stress to those who are home schooled.

### **Risk management plans**

A risk management plan should be a clearly identifiable part of the care plan and will usually involve the child's family. It should:

- address each of the long-term and more immediate risks identified in the self-harm assessment, address the specific factors (psychological, pharmacological, social and relational) identified in the assessment as associated with increased risk, with the agreed aim of, reducing the risk of repetition of self-harm and/or the risk of suicide



- include a crisis plan outlining self-management strategies and how to access services
- Treat any identified, related psychiatric disorder such as depression

### **Example One**

E is a 13 year old girl. She was admitted under paediatrics after an overdose of 8 paracetamol tablets. She took them after an argument with her father about her new boyfriend. Her father refused to let her have any contact with him outside of school hours. She remembers being angry with her dad and storming upstairs. The tablets were in the bathroom cabinet and she took them with some water.

At the time of taking the tablets she was angry, sick of arguing with her parents and wished that she was dead so that they would stop. Shortly after taking the tablets she was worried about what might happen. She was scared that her dad might be angry so phoned her mum at work who, in turn, called for an ambulance.

E had a psychiatric assessment that revealed no underlying depressive illness.

At the hospital E expressed no suicidal intent. She was happy that her parents were not angry and about how much they seemed to care about her. On questioning E said that she had started to argue more with her dad since he and her mother had started sleeping in separate bedrooms. Her dad would get very angry, shout a lot and on one occasion had pushed her over.

E's father was seen separately to go through the pushing incident. It was clear that he had been having problems with his mood for some time and that these had started to deteriorate since his wife moved into the spare room. He agreed to make an appointment with his GP to take things forward. Both parents and E agreed to attend family therapy sessions.

As there were some child safeguarding issues, a safeguarding referral was made and a discussion was held with a social worker. At this meeting it was decided not to put E on the child protection register as there was an appropriate plan in place to meet the family's needs.

E was discharged from hospital but followed up by the CAMHS team. As she was felt to be at no further risk from self-harm, she was discharged after a couple of appointments.

Although E has had a significant event she is not at risk in the longer term and is unlikely to meet the prospective test in DLA.

### **Example Two**

B is a 15 year old boy. He has had problems with anxiety since he was 12. He finds it difficult to cope at school and when he feels out of control. When

he feels overly anxious, B cuts his arms. He has used a razor in the past but his parents now limit access to blades.

He is under CAMHS who see B regularly and are teaching him techniques to decrease his anxiety which have reduced the number of occasions B cuts himself but it still happens when B is feeling very anxious. B's parents have had input to help them learn when B is showing signs of increased anxiety. They work hard to make sure the home is a relaxed space for B by not placing too many demands on B at once and are undergoing family therapy.

He has a plan in place at school so he can spend time with a teaching assistant on the days he is finding the lunch break stressful. This works well and has improved B's school attendance.

B has a moderate functional restriction. His parents and school provide additional care throughout the day and as a result of the care he receives his condition is under control.

### **Example Three**

CP is a 16-year-old girl who lives in a residential setting after several failed foster placements. She was abused in her parental home when young and was recently abused again. She has a diagnosis of autism and has had disordered eating in the past and has made previous suicide attempts. She recently made 'Facebook' posts saying she was worthless and used other social media to say goodbye to friends.

CP is at high risk of suicide. She has a care plan involving constant supervision. Measures are in place to maintain her dignity and show her that her supervision is not punitive, as well as trying to support and develop CP in her skills and strengths. Inpatient treatment has been considered for her, and remains under active consideration.

She is on the child protection register and has a social worker allocated to her. She has been prescribed anti-depressants and is undergoing therapy. Her team, involving her social worker, psychiatrist, carers and therapists have regular meetings about and with CP.

CP has a severe functional restriction. She has constant supervision in place in all settings and remains at risk of suicide.

### **Example Four**

DT is a 14 year old boy who lives with his family. He experienced bullying at school from a group of other pupils. They would pick on him and humiliate him because of his small stature and sent him threatening text messages. His parents became concerned as he seemed to be lacking in confidence and increasingly withdrawn.

He often complained of headaches or stomach pains and had time off school as a result. His parents knew nothing about the bullying until he began refusing to go to school and they found that he had been searching websites about committing suicide. They took him to the GP who referred him urgently to Child and Adolescent Mental Health Services (CAMHS) and the school was subsequently informed. As a precaution, his parents needed to monitor DT carefully and make the house safe by removing access to knives and other potentially lethal hazards. He was provided with counselling and changes were made at school, including a change of form and a mentor.

DT has a severe functional restriction. He has a significant history of suicidal intent with planning due to bullying. He is at risk of being bullied again and responding in the same way. His parents have to constantly monitor him and keep the home environment free from anything he could use to commit suicide.

## Living with Depression - Example Profiles

### Mild Depression

R is a 10 year old boy he has been struggling with behaviour at school and home for the last 3 months. His parents had a meeting at school as his school work has been deteriorating for the last month. In addition to this, he has looked sad and been withdrawn most of the time. He has been struggling to settle at night and has been leaving his meals half finished and saying that he isn't hungry. His GP saw him and he admitted to feeling fed up and lonely at school due to bullying by a couple of the other pupils. At school he has been described as overactive and he is behind in his learning when compared to his peers.

The GP recommends that his parents take action at school and opts for a period of watchful waiting. Despite this R's mood doesn't improve and the GP refers him to CAHMS. He is assessed by CAMHS and they find that there are background problems in family interactions coupled with a recent deterioration in this is identified as of relevance to R's problems. CAHMS decide to offer family therapy. After interventions at school and family therapy R's mood begins to improve with a corresponding improvement in school work, appetite and sleep pattern. After 6 months he is discharged back to his GP and his mood is still stable at 12 months.

### Moderate Depression

D is a 15yr old boy who lives with his parents. He has had persistently low mood for about six months and has been seeing his GP for 2 months. He is also anxious, has become irritable and argumentative and has been falling out with his family and friends over simple things. He has been falling behind in his school work as he has been struggling to concentrate and has been lacking in motivation.

His GP referred him to CAMHS as he has been describing intermittent thoughts that he would be better off dead. He hasn't had any plans to harm

himself. D and his parents were assessed by CAMHS and, after being given reading material, D was referred for a course of CBT.

Subsequently D's mood improved significantly over his 12 week treatment. The therapist kept D under review for 6 months when he was discharged because he remained symptoms free.

### Severe Depression

J is a 12 year old girl who lives with her mother and older brother. Her mother suffers with depression and her parents separated 6 months ago. She presented to her GP after concerns were expressed about her performance at school. Although initially settling at senior school for the last 6 months her academic performance has begun to slide. She isn't engaging in class, her homework is of an unacceptable standard and she spends most of her dinner and break time alone. When she was spoken to by her teacher she admitted to feeling sad, lonely and expressed thoughts about self harm and suicide.

Her GP assessed her and referred her urgently to CAMHS as in addition to expressing ambiguity about living, she reported hearing blaming voices several times per day. A CAMHS psychiatrist assessed J and found her to be markedly slowed down, expressing persistent feelings of loss of enjoyment and energy, fatigue, feelings of guilt and self-blame. She also said that she thought that she would be better off dead. After discussion with her and her parents, the psychiatrist commenced her on Fluoxetine. In addition to this she was referred for a 12 week course of CBT. She was seen by CAMHS weekly for the first 6 weeks with her Fluoxetine dose increased to 20mg after 2 weeks.

After 12 weeks J's hallucinations had stopped and her mood had improved. Her school work, although not as good as previous was improving. CAMHS kept J under follow up for a total of 12 months and at this point she was back to normal and her Fluoxetine was being gradually tapered off. At 2 years her mood continued to be good and her medication had been discontinued.

## What you need to know about Diabetes

### What is Diabetes?

Diabetes mellitus is a condition caused by a lack of the hormone insulin in the body or resistance to its effects .....

- [Diabetes](#)

### What are the effects and signs?

Type 1 diabetes tends to develop quickly over days or a couple of weeks. The symptoms are those of hyperglycaemia (high blood sugar levels).....

- [Effects of Diabetes](#)

### How is it assessed?

Blood sugar levels are normally controlled within strict limits by the body's production of insulin .....

- [Assessment of Diabetes](#)

### How is it treated and managed?

Type 1 diabetes will always need insulin treatment. Type 2 diabetes will sometimes require this treatment .....

- [Treatment for Diabetes](#)

### What evidence is available?

The vast majority of children will receive repeat prescriptions for insulin through their GP .....

- [Evidence](#)

### How long will the needs last?

- [Prognosis and duration of the award](#)

## What is Diabetes?

Diabetes mellitus is a condition caused by a lack of the hormone insulin in the body or resistance to its effects. The insulin acts in the body to control levels of glucose, protein and fat in the blood. Insulin is produced when a rise in blood glucose is detected, and acts on cells to make them take up glucose from the blood, so reducing blood glucose levels. The cells cannot function without this influx of glucose, which is insulin dependent. Insulin

acts in balance with other hormones to keep glucose levels steady between meals. When blood glucose levels drop too low hypoglycaemia occurs. When blood glucose levels are too high, the term used is hyperglycaemia. Both states cause unpleasant symptoms and can become life threatening if untreated. Hyperglycaemia (due to lack of insulin or inadequate doses of insulin) is the hallmark of diabetes mellitus. The treatment is insulin to reduce blood glucose levels to normal limits. Hypoglycaemia can occur as a result of either too high a dose of insulin, or of inadequate food intake during treatment with insulin.

There are many types of diabetes, but the two most common are Type 1 and Type 2.

Type 1 diabetes is the most common type in children, affecting 96% of all children and young adults with diabetes in the UK – around 31,500. In this condition, Type 1, the pancreas fails and stops making the hormone Insulin. The peak age of diagnosis of childhood Type 1 diabetes is 9-13. The symptoms of Type 1 diabetes are produced by hyperglycaemia, which develops quickly over days or a couple of weeks (see below). This rapid onset is more pronounced in children and young people, meaning that failure to diagnose the condition can result in the death of a child.

In Type 2 diabetes, which affects fewer than 2% of children and young people, the pancreas cannot make enough insulin or the insulin it makes

doesn't work properly. The onset is more gradual and the symptoms similar, but less dramatic.

### **What are the effects and signs?**

Common symptoms of Type 1 diabetes include:

- Increased thirst
- Passing water frequently – Known as 'polyuria'
- Fatigue
- Weight loss
- General itching and thrush infections

Less common symptoms include:

- Cramps
- Constipation
- Blurred vision
- Recurrent skin infections and urinary tract infections

Once the condition is diagnosed and treated with insulin, these symptoms will resolve unless the blood glucose is poorly controlled.

### **Overview of treatment and complications of diabetes**

Diabetes is a life long condition that requires daily management. Adherence to tight blood sugar monitoring and control is fundamental in preventing later-life complications in the form of visual impairment, kidney disease and arterial disease. It is recommended by NICE that children and young people are prompted and supervised to maintain a target level of blood glucose as near normal as possible, that is, in people with diabetes: before meals 4-7 mmol/l and 2 hours after meals 5-9 mmol/l.

As in all life-long conditions, an important aim is to enable the young person ultimately to manage his/her own condition. At initial diagnosis many children are able to administer their own insulin injections but they will require adult support in calculating the correct insulin doses and the carbohydrate content of meals. Adult support (parent or other suitably trained person) would also be required to recognise and deal with abnormal blood glucose readings. The NICE guidance supports this aim of self management (by

school leaving age) and a number of courses/programmes exist to promote the transition to complete self management.

### **Short term complications**

**Hyperglycaemia:** if blood glucose is not managed effectively and glucose levels are too high, symptoms such as mood changes, tiredness, excessive thirst and urination may develop once again. If hyperglycaemia is uncontrolled and blood glucose continue to rise, there is a risk of diabetic ketoacidosis (DKA), which is life threatening and will almost always require hospitalisation, often in intensive care units.

**Hypoglycaemia:** if blood glucose is not managed effectively and glucose levels are too low, there may be symptoms including dizziness, sweating, trembling, confusion, weakness, nausea and ultimately, if not recognised and treated seizures and coma (and death). Achieving a good, near normal blood glucose level in children with both Type 1 and Type 2 diabetes is mainly dependent on the balance between insulin, food intake and physical exercise and requires careful management that is not always easy to achieve. Blood glucose levels are also affected by changes in diet, changes in weather, minor/major illness, growth and puberty.

### **Long term complications**

A high proportion of people who have had diabetes for many years will develop complications. These are caused by damage to the microvascular and macrovascular systems due to excessive blood glucose levels over time and include an increased risk of cardiovascular disease, sight loss, amputations and kidney disease. Children and young people with diabetes will have regular screening for signs of diabetic eye disease (retinopathy), foot problems and kidney disease from the age of 12 years.

To consider H/R Mobility Severely Visually Impaired (SVI) criteria, see - [Visual Impairment Deeming Provisions](#)

### **How is it assessed?**

#### **Blood tests**

Blood sugar levels are normally controlled within strict limits by the body's production of insulin. Abnormally high blood glucose levels are the hallmark of diabetes. Blood sugar can be measured using a pin prick of blood and a glucose meter or via a routine lab blood test for blood glucose.

#### **Urinalysis**

Glucose is not present in the urine under normal conditions, the presence of glucose in the urine usually indicates diabetes – this can be confirmed by blood glucose monitoring.

#### **Glucose tolerance test**

In cases of type 2 diabetes where there is doubt over the diagnosis of diabetes, a glucose tolerance test can be performed. This test measures blood glucose after a test dose of glucose solution. In a normal person insulin will



be produced to limit the rise in blood glucose. In someone with diabetes the 'glucose tolerance curve' (a graph produced of blood sugar levels after a test dose of glucose) will be abnormal. If there is evidence of poor blood sugar control that is not abnormal enough to be diabetes this may be called 'impaired glucose tolerance' or pre-diabetes or metabolic syndrome.

### **How is it treated and managed?**

Children with Type 1 diabetes have extra care needs relating to their diabetes, due to the intensive management of Type 1 that is now recommended within the revised NICE guidelines. This tight control may be achieved by intensive insulin management (multiple daily injections or insulin pump therapy) from diagnosis, accompanied by carbohydrate counting.

Insulin is a hormone given by injection that reduces blood glucose. Regular administration of insulin will be required to control blood glucose through the day. Monitoring blood glucose, calculating the carbohydrate content of food, working out the insulin dose required, and administering insulin by injection or using an insulin pump is the treatment required. Insulin treatment may be managed in two ways by:

- Regular insulin injections through the day.
- Use of an insulin pump. An insulin pump is the size of a mobile telephone and is worn around the waist under clothes. The pump is attached via a tube to a plastic cannula under the skin and gives a background dose of insulin to control blood glucose. Booster doses can be programmed into the machine at mealtimes based on carbohydrates to be eaten and most recent blood glucose level.

Both methods require regular monitoring of blood glucose through the day and knowledge of carbohydrate content of foods to calculate dose of insulin required. It is important that if a dose of insulin has been calculated to cover a meal that the entire meal is then eaten, supervision at meal times may be required particularly for younger children. The aim of treatment is to keep blood glucose readings within normal limits, as if the body was producing its own insulin to control blood glucose. Good control of blood glucose reduces the risk of long term complications of diabetes and short-term problems with control, namely symptoms of hyperglycaemia and hypoglycaemia. There are different types of insulin available that have short, medium and long durations of action.

### **Insulin Injection Regimen**

Examples include:

- The most commonly used regimen is multiple injections - this will usually consist of short acting insulin at mealtimes and a longer acting insulin in the morning and/or evening. At least 4 insulin injections per day are needed, often more. Multiple

blood glucose measurements are made to calculate insulin dose required (between 4 and 10 blood glucose measurements per day). This regimen involves more monitoring and injections but allows flexibility in terms of meal times. Children are most likely to use a pen type device for administering insulin.

- Twice daily injections – 12 hours apart – rigid eating pattern required – not very often used in current treatment regimens
- Three times daily injections – rigid eating pattern required – not often used

Children may be able to do injections themselves from age 8 but will need prompting and close supervision to ensure correct doses are drawn up and injection sites are rotated. If they are newly diagnosed they are likely to need help for a few months at least. Even those who have had diabetes for a while may find the pens difficult to use – they are not designed for children's hands.

### **Insulin Pump**

The insulin pump has tended to be used in children whose diabetes has been hard to control on an injection regimen but is now commonly offered to all children who are clinically suitable. The pump is worn 24 hours a day and the insertion site is moved around to avoid skin complications. Managing diabetes with an insulin pump is more complex than using an injection regimen. Children may be able to do some of this themselves, depending on age but will need prompting and close supervision.

### **Blood glucose monitoring**

Blood glucose levels vary substantially during the day and between days. The HbA1C is a formal blood test measured in a laboratory to give an indication of blood glucose control over a 3-month period. The NICE recommendation is that an HbA1c target level of 48 mmol/mol (6.5%) or lower is ideal to reduce the risk of long-term complications. This is a challenging target and many children do not achieve it.

It is necessary to check glucose at home to guide day to day insulin management. Children should be aiming for levels of 4-7mmol/l before meals and 5-9mmmol/l after meals. Children and/or carers can be

trained to look for peaks and troughs of blood glucose and to avoid these by adjusting insulin dose or carbohydrate intake.

There are three ways to check blood glucose:

### **Fingertip testing/capillary blood glucose checking.**

The fingertip is pricked to obtain a drop of blood, and this is used on a simple palm sized blood glucose monitoring device. Children/young people with Type 1 diabetes and their family members or carers are advised to routinely perform 7 – 10 blood glucose checks per day and will sometimes be required to check at night. It is necessary to do this before every meal and snack, before and after exercise, before the child's bedtime and at the parents' bedtime when the child is young. With teenagers, parents may do this first thing in the morning on non-school days while the young person is asleep.

### **Flash glucose monitoring/ intermittently scanned continuous glucose monitoring.**

The child/young wears a small monitoring device (sensor) on their arm/stomach which takes readings of the glucose level in the fluids between the cells (Interstitial glucose). A device is then passed over this sensor to get a glucose reading. The device must be changed after 14 days or if it falls off. This medical device is attached to the child 24/7.

### **Continuous blood glucose monitoring/ real-time continuous glucose monitoring.**

As with flash glucose monitoring, a device is worn 24/7 to measure glucose, but this transmits glucose levels to linked devices, such as a smartphone, automatically. Some devices may give a warning when the glucose level is very high or very low. Devices may be programmed to link up directly to the specialist team looking after the child to aid decisions about insulin regimes. Follow-on Apps can also be used by parents and school staff. This could be used as a source of information when needed and may be particularly helpful when determining night needs for children/young people with diabetes.

It is possible to link some continuous glucose monitors to insulin pumps so that the pump alters the amount of insulin being given depending on the glucose reading. This does not replace the need to carbohydrate count and enter this information into the insulin pump.

The type of blood glucose monitoring a child has depends on personal preference and local availability of options, though it is very rare in children/young people to use only fingertip checking. NICE guidelines (2022) recommend continuous blood glucose monitoring in children and young people with diabetes, unless there is a clear preference for flash

monitoring.

Children who use flash or continuous blood glucose devices to monitor their glucose level will also need to do some fingertip checking to ensure the device is accurate and as a back-up if the device stops working.

### **Night checking**

Night checking, if used, is likely to be once or twice a week, and takes a few minutes. If more extensive night checking is claimed this should always be supported by medical evidence. However, for very young children, or when a child is ill, this is expected.

Please note that for those using continuous glucose monitoring, alarms alert the child or the parents if the levels are going too low or too high. When this occurs action must be taken. With Flash glucose monitoring, information about glucose levels overnight are obtained when the sensor is scanned on waking. This information may indicate that adjustments are needed or confirm safe levels overnight. If parents/guardians send in information on blood glucose levels downloaded from their device with their claim, please consider referring to medical services for interpretation.

Care needs overnight will relate to the parent/guardian needing to check with fingertip monitoring, flash sensor monitoring, action required due to alarms, adjusting of insulin administration for high glucose levels, or treating low sugar levels.

### **Self-checking**

Children/young people may be able to do the blood glucose checking themselves from diagnosis, depending on age, but will need prompting and close supervision. They are unlikely to manage any night time checks themselves. They may not be able to manage the equipment that is not designed for children's small hands. They will also need guidance to interpret results and calculate the next insulin dose. This can be difficult for a child/young person, so they will need the support of an adult to do this until they are mature enough (see previous re preparation for the child's self-management).

### **Ketone testing**

Children and young people with type 1 diabetes, and their family members or carers are also told to check for ketones if they are ill or have hyperglycaemia. Ketones can only be checked by capillary blood (fingertip check). If the child / young person has ketones they will need to give extra insulin to treat this and likely will need to speak to their diabetes clinical team for advice to manage ketones. The presence of ketones can

lead to life threatening Diabetic Ketoacidosis (DKA) if it is not treated properly.

### **Useful websites:**

Glucose monitor manufacturer websites:

<https://www.dexcom.com/en-gb>

<https://www.freestyle.abbott/uk-en/home.html>

Insulin pump manufacturer website:

<https://www.medtronic-diabetes.com/en-gb/insulin-pump-therapy/minimed-780g-system> insulin pump system manufacturer website

Diabetes UK page on flash and continuous glucose monitoring.

<https://www.diabetes.org.uk/guide-to-diabetes/diabetes-technology/flash-glucose-monitors-and-continuous-glucose-monitors>

## **Optimising glucose control**

### **Hyperglycaemia**

If blood glucose (BG) is raised, it can be reduced by increasing insulin dose and increasing fluids. If it is getting dangerously high, children may need medical help and advice or admission to hospital to correct it. Insulin requirements increase during infections, times of stress and growth spurts for example and learning how to manage this independently takes time and experience.

### **Hypoglycaemia**

Blood glucose levels may decrease below normal and cause symptoms of nausea, dizziness, blurred vision, disorientation, confusion and unconsciousness or seizure. Children should carry dextrose tablets or 'Glucogel' or other sources of glucose to take if this occurs. Some children do not develop warning signs of hypoglycaemia and this is a hazard as loss of consciousness may be the first sign of it. Such children can carry an emergency injection kit containing a hormone called glucagon, which raises blood glucose when injected. A non-medical person can be trained to do

this. Warning signs can be restored by improving diabetes control and minimising episodes of hypoglycaemia.

The child may need to eat a more complex form of carbohydrate after the initial hypoglycaemia has resolved, for example fruit or a sandwich, especially if they were exercising.

- [Other factors affecting care needs](#)

**What evidence is available?**

Sources of Evidence	Information Provided	Limitation of Evidence
Parent / Carer	First source of information and associated evidence.	Does not have specialist knowledge.
GPFR	<p>Children with diabetes should be identified by the primary care trust. The vast majority of children will receive repeat prescriptions for insulin through their GP. Confirmation of the diagnosis of diabetes and the prescription of insulin to the child is all that is required in the typical case.</p> <p>Where extra difficulties are claimed such as behavioural difficulties making treatment more challenging, evidence of such difficulties in addition to confirmation of diabetes will need to be obtained from the GP.</p>	Not likely to be able to provide evidence on difficulties with blood sugar control if this being managed by the diabetic specialist nurse from the hospital.
HFR – best source of evidence Clinical Nurse Specialist or Consultant Paediatrician	Where extra difficulties are claimed such as behavioural difficulties making treatment more challenging, evidence of such difficulties in addition to confirmation of diabetes will need to be obtained from the GP, Consultant Paediatrician or Children’s Diabetes Clinical Nurse Specialist. If extra difficulties are related to problems with diabetes control the clinical nurse specialist will be the best source of information.	N/A
Orthoptist	Assessment of vision (visual acuity and fields)	May not have information about symptoms, signs, investigations other than assessment of vision, treatment/management, and unlikely to have information about resulting disability or needs

Optometrist	Symptoms, signs, investigations including assessment of vision, treatment/management, and likely to have information about resulting disability, needs and provision of low vision aids.	None
If considering entitlement to H/R Mobility component under the Severely Visually Impaired (SVI) provisions, the following evidence source must be used:		
Consultant Ophthalmologist	Symptoms, signs, investigations including assessment of vision, treatment/management, and likely to have information about resulting disability or needs	None

[List of NHS hospitals with ophthalmology departments](#)

**How long will the needs last?**

This guidance covers:-

Diabetes mellitus Type 1	Disability Code S11
Diabetes mellitus Type 2	Disability Code S12
Diabetes mellitus (type not known)	Disability Code S13

<b>Age at date of claim</b>	<b>Injected insulin treatment – award as follows</b>	<b>Insulin pump treatment - award as follows</b>
1-16	Award to age 16	Award to age 16

**Night needs**

Night needs are not dependent on age, for example, they might become relevant for the first time in relation to an award for a teenager. It may be that



a child who previously did not satisfy the conditions for an award based on night needs now meets the conditions.

Age at date of claim – night needs	Injected insulin treatment – award as follows	Insulin pump treatment - award as follows
N/A	Award for 2 years	Award for 2 years

Diabetes with visual complications – Retinopathy. For information on H/R Mob SVI deeming provisions see [Visual Impairment Deeming Provisions](#).

The important indicator in terms of care needs in diabetes is whether treatment with insulin is required. Children with Type 1 diabetes will always need insulin treatment and sometimes those with Type 2 diabetes will also require this treatment. Children with type 2 diabetes are unlikely to have significant care needs unless they are receiving insulin treatment.

#### Insulin injection treatment

A child having insulin injection treatment is likely to require attention with treatment at home and at school until they leave school at the end of year 11. The latest NICE guidance recommends that children require supervision and support at school until this age. Supervision and support at school is not provided beyond year 11, NICE recommends that children can substantially manage their condition themselves at this stage. The amount of support a parent provides will vary between children and over time and even though a child may be substantially managing their condition themselves the parent may still take a keen interest.

#### Exceptions

Care needs related to insulin treatment may be prolonged beyond the ages given above where there is any diagnosed learning, behavioural,

developmental, or mental health condition, which affects the ability to independently manage treatment on a daily basis at the usual age expected.

Written evidence of a health condition and care needs arising must be provided to support a claim in a young adult over the age of 16.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Other factors affecting care needs of children with insulin dependent diabetes**

Any condition which makes controlling dietary intake and delivering timely insulin treatment more difficult will increase care needs:

- Any diagnosed behavioural condition or developmental condition which makes compliance with treatment on a daily basis difficult or prolonged will increase time spent providing treatment.
- Any diagnosed behavioural condition or developmental condition that impairs the ability to learn how to manage treatment independently at the age expected will prolong care needs beyond the age expected.
- When diabetes is one of a number of conditions which in combination require extra care and attention on a daily basis.

Diabetes may be caused by use of oral steroid drugs, these are necessary in a number of childhood conditions including kidney disease, childhood cancer and asthma. Diabetes and insulin treatment will add an extra dimension of care required to the parents of affected children.

Children who develop the longer-term complications of diabetes such as kidney failure and visual problems may have care needs related to these in addition to those of unaffected children.

### **Care needs of a typical diabetic child on insulin treatment**

Insulin can be given intermittently through the day as individual injections or by means of a pump which the child wears on their person and adjusts whenever food is eaten. Whether a pump is used, or injections are used the principles of managing blood sugar levels with insulin treatment are the

same. Insulin is administered to control blood sugar each time food is eaten.

The steps involved in the administration of insulin include:

- Monitoring blood glucose
- Assessing carbohydrate content of meals
- Calculating dose of insulin required based on carbohydrate content of food to be eaten and blood sugar level
- Safe storage and use of insulin and medical equipment
- Preparation of appropriate amount of insulin for injection
- Injection of insulin
- Safe disposal of needles

It will take some time for a child to learn how to carry out these steps with guidance and supervision from parents. The child is likely to administer their own injections from diagnosis but prompting, supervision and guidance will be required for all of the above steps for some time depending on the age of the child. Delivering treatment as prescribed every day is necessary to avoid hyperglycaemia and hypoglycaemia.

Adjusting to a diagnosis of diabetes and incorporating blood sugar management into daily life is a significant challenge for any child. Gaining experience of insulin dose adjustment for different types of food and during inter-current illness takes time. Significant parental help, guidance, prompting and supervision of treatment and emotional support accounts for the extra care required.

### **Type 2 diabetes**

Children with type 2 diabetes are unlikely to have significant care needs unless they are receiving insulin treatment. If insulin treatment is being used, follow the above section on 'care needs of a typical diabetic child receiving insulin treatment'.

Children with type 2 diabetes may be following a calorie restricted diet if they are overweight. They may require encouragement to take physical exercise. They may require supervision for a few minutes each day whilst taking their tablet treatment. Attention to diet and physical exercise is a normal part of parenting.

### **Tablet treatments for type 2 diabetes**

These children are likely to be overweight and weight reduction can improve insulin sensitivity. Blood sugar levels can be improved by weight loss, increased physical activity and careful diet. Where diet, exercise and weight loss fail to improve blood sugar control tablet treatments that reduce blood

sugar can be tried. It should be used in addition to those measures. The main drug used is Metformin, it is a tablet taken 1-3 times daily. Blood sugar

monitoring on this treatment is required but does not have to be done on a daily basis.

This drug can be combined with an insulin injection regime, usually one or two injections per day. Blood glucose monitoring is required but does not have to be done on a daily basis.

Other tablet treatments rarely used include glibenclamide, gliclazide and tolbutamide.

What you need to know about Down syndrome

### **What is Down syndrome?**

Down syndrome is a genetic condition which causes learning disabilities and

..... [Down syndrome](#)

### **What are the effects and signs?**

There are over 120 different features of Down syndrome ..... [Effects of Down syndrome](#), [Living with Down syndrome](#)

- 

### **How is it assessed?**

Down syndrome is usually diagnosed at or shortly after birth.....

- [Assessment of Down syndrome](#)

### **How is it treated and managed?**

Children with Down syndrome will be ..... [Treatment of Down syndrome](#)

### **What evidence is available?**

- First source of information and associated evidence .....
- [Evidence](#)

### **How long will the needs last?**

- [Prognosis and Duration of award](#)

What is Down syndrome?

Down syndrome is a genetic condition which causes learning disabilities and characteristic physical features. Down syndrome is congenital. This means that it has been present from birth and is as a result of abnormal development of the foetus. As many as 120 features have been described in Down syndrome. Many children have no more than 6 or 7 of these. A degree of intellectual disability is the only feature that is present in all children.

What causes Down syndrome?

Down syndrome is due to having three copies of chromosome number 21. This additional copy causes excessive amounts of certain proteins to be formed in the foetus which interferes with the normal foetal development. This is called Trisomy 21 and represents 95% of children with Down syndrome.

Translocation. This occurs in 4% of the children with Down syndrome and is due to an extra part of Chromosome 21 being present. These children have the same symptoms and presentation as Trisomy 21.

Mosaic Down syndrome. This affects 1% of children with Down syndrome. It is caused when only a proportion of the body's cells contain the extra copy of chromosome 21. Mosaic children will be less markedly affected as some

of their cells will have been functioning normally. Their development and function are closer to the rest of the population.

What is the prevalence of Down syndrome?

- For every 1,000 babies born, one will have Down syndrome
- About 750 babies with Down syndrome are born in the UK each year
- Down syndrome affects people of all ages, races, religious and economic situations
- There are approximately 40,000 people with Down syndrome living in the UK

The average life expectancy for a baby with Down syndrome is around 60 years.

What are the effects and signs?

Appearance

Low muscle tone

Chest

Visual problems

Hearing problems

Hypothyroidism

Learning disability

Delayed development

Congenital heart disease

Leukaemia

Obstructive sleep apnoea

Craniovertebral instability

Coeliac disease

Diabetes

## Arthritis

There are over 120 different features of Down syndrome. No one child with Down syndrome will have all 120. The features listed below are the more common ones seen.

### Appearance

Children who have Down syndrome have a characteristic facial appearance with rounded face, slightly flattened head and up slanting eyes. Their mouth cavity is slightly smaller than average, and the tongue slightly larger. They are shorter than average and have a slower rate of growth. Despite this it can be noted that children with Down syndrome will usually have normal or high BMI after infancy.

### Low muscle tone

Due to this, babies with Down syndrome are floppy and may have feeding problems. They may need to be fed via a tube in the stomach.

### Chest

Frequent chest infections are also common in the first few years. The reason for this is not completely understood and there are likely to be several contributory factors.

### Visual problems

60% of children with Down syndrome have problems with their vision.

Cataracts: These are usually small flakes at the edge of the lens, which usually do not interfere with vision. Rarely, cataracts are denser and



located in an area that causes problems with vision. These are removed surgically.

Refractive errors: This means short or long sighted plus astigmatism. These difficulties are corrected by wearing prescription glasses.

Squint: This is where both eyes cannot look at an object at the same time. Management is by usage of a patch or glasses and a simple corrective operation in more severe cases.

Keratoconus: The front of the cornea becomes cone shaped. Treatment is with special types of contact lens. Lens transplants may be needed but this will be when they are an adult.

Problem	Percentage
Cataract	15%
Squint	20%
Refractive errors	80%
Keratoconus	10%

### Hearing problems

Over 50% of people with Down's syndrome have hearing impairment, which may be mild, moderate, severe or profound. If undetected it is likely to be a significant cause of preventable additional learning delay. Lifelong audiological surveillance is essential for all.

Conductive deafness: This is when sound cannot pass efficiently through the outer and middle ear to the inner ear (cochlea and auditory nerve). The most common type of conductive deafness in childhood, and in those with Down's syndrome, is caused by 'glue ear' (usually caused by middle ear infections).

Glue ear: This is a build-up of sticky fluid in the middle ear. For the ears to work properly, the middle ear needs to be kept full of air. About 60% to 70% of children with Down syndrome have a conductive deafness caused by glue ear.

This condition can be managed with the insertion of small tubes into the ear drum to allow the fluid to drain away (grommets). There is a high risk of recurrence so the usual recommendation is to treat with hearing aids.

Sensori-neural deafness: This is caused by a fault in the inner ear or the auditory nerve (the nerve that carries the electrical signals from the cochlea to the brain). Sensori-neural deafness is permanent. A very small number of children with Down syndrome are born with this type of deafness and some

develop sensori-neural deafness as they get older, particularly through adolescence and into adult life. About 10% to 15% of children with Down syndrome have sensori-neural deafness.

These children will be managed with hearing aids. The more severely affected may need to be taught sign language to help communicate.

### Hypothyroidism

At all ages thyroid disorder (usually hypothyroidism) occurs more frequently in people with Down's syndrome than in the general population. Around 10% of the school age population have hypothyroidism. The prevalence increases with age. If undiagnosed, thyroid disorder constitutes a significant cause of preventable additional disability. It is diagnosed with blood tests

and treated with replacement thyroid hormone tablets once daily. Once their blood tests are stable, children will only need a blood test every 12 months.

If the thyroid gland is underactive (hypothyroidism), the person is unable to make enough thyroxine and can become tired, overweight and generally sluggish with slow physical and mental reactions.

### Learning disability

The average IQ of a child with Down syndrome is 50. Around 10% of children with Down syndrome have an additional diagnosis of an autistic spectrum disorder.

Degree of learning disability	IQ range	Percentage of children with Down Syndrome
Borderline	70-80	1
mild	50-69	39.4
moderate	35-49	29.8
severe	20-34	20.5
profound	Less than 20	9.3

### Delayed development

Children with Down syndrome develop slower than normal children.

	Down Syndrome		Normal	
	Average Age attained	Age range	Average Age Attained	Age range
Sits Alone	11 months	6-30 months	6 months	5-9 months
Crawls	12 months	8-22 months	9 months	6-12 months
Stands	20 months	1-3.25 years	11 months	8-17 months
Walks alone	2 years	1-4 years	14 months	9-18 months
First word	23 months	1-4 years	12 months	8-23 months
Two word phrase	3 years	2-7.5 years	2 years	15-32 months
Responsive smile	3 months	1.5-5 months	1.5 months	1-3 months
Finger feeds	18 months	10-24 months	10 months	7-14 months
Drinks from cup	23 months	12-32 months	13 months	9-17 months
Uses spoon	29 months	13-39 months	14 months	12-20 months
Bowels Controlled	3.75 years	2-7 years	19 months	16-42 months
Dresses Self	7.25 years	3.5- 8.5 years	4 years	3.25-5 years

This chart is to be used as a guide only. It is useful as a comparison between an average non disabled child and an average child with Down syndrome.

### Congenital heart disease

Also - See: [Congenital Heart Disease \(CHD\)](#) for more details.

Forty to sixty percent of children with Down syndrome will have congenital heart disease. This is disease of the heart that has been present since birth.

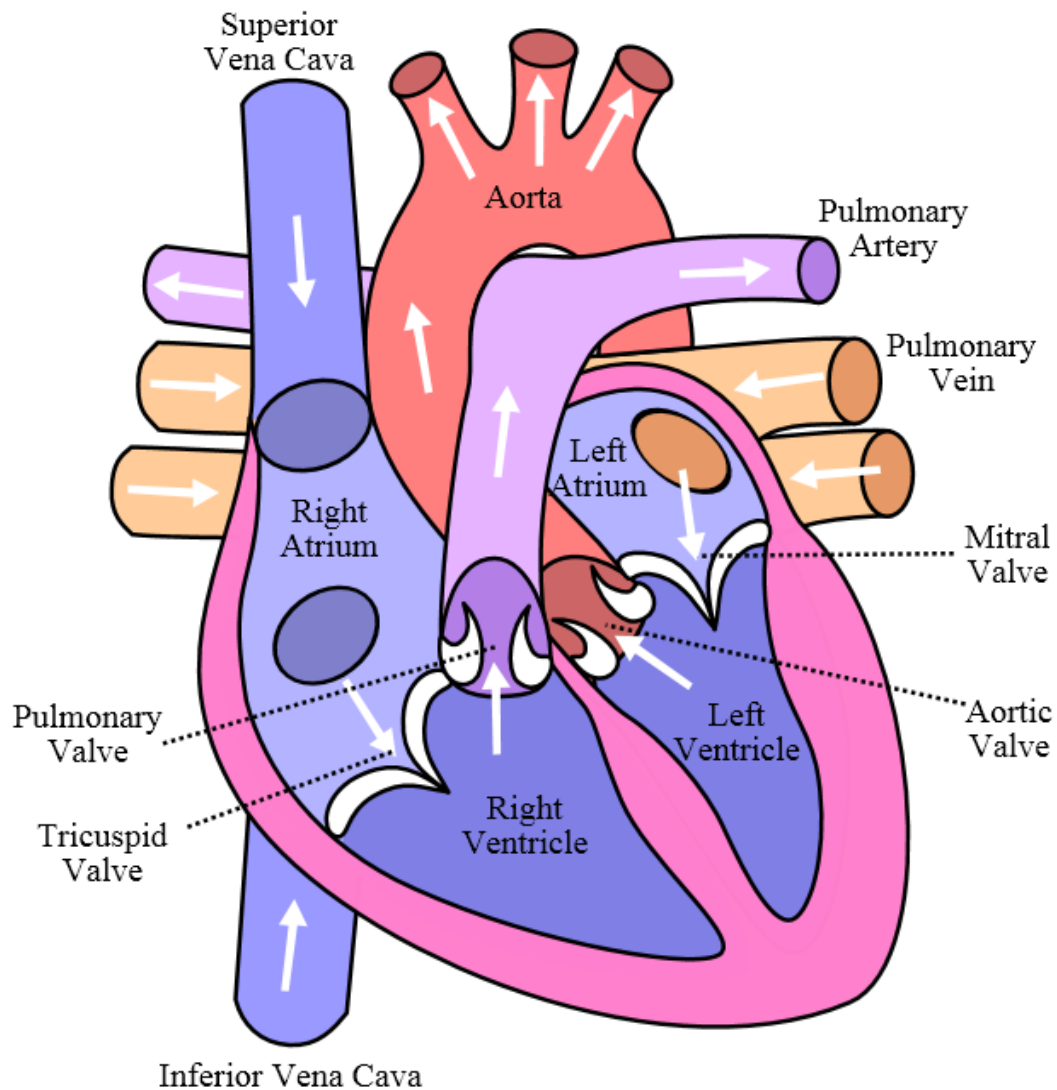
Mixed Atrioventricular septal defect	35%
Atrial septal defect	30%
Ventricular septal defect	24%
Patent ductus arteriosus	7%
Tetralogy of Fallot	4%

Congenital Heart Disease is a problem with the structure of the heart that has been present since birth. The signs and symptoms can vary from condition to condition and can vary from none to life threatening. Usually they are dependent on the severity of the underlying cardiac defect.

The heart is a muscular pump that consists of two pumping circuits and moves blood around the body. Each circuit has one atria sitting on top of its ventricle. Blood flows into the atria before passing into the ventricle. The ventricle is the main pumping chamber for each circuit. The right side of the heart pumps blood to the lungs and the left side pumps it around the body. At the exit of each heart chamber is a heart valve. These valves help regulate the blood flow between the chambers and also the flow out of the ventricle and around the blood circuits. The two sides of the heart are joined together down the middle normally blood travels separately through the two sides. Where the two sides join together is called the septum. Between the

atria it is called the atrial septum and between the ventricles it is called the ventricular septum.

Figure 1- The heart



By Own work, CC BY-SA 3.0, <https://commons.wikimedia.org/w/index.php?curid=830253>

**Atrioventricular septal defect (AVSD):** This is the most common congenital cardiac defect in people with Down's syndrome. An atrioventricular septal defect is when the septum or partition between the two ventricles (lower chambers) of the heart and between the two atria (upper chambers) of the heart does not develop properly and a hole is present. This allows mixing of

blood from the left to the right side of the heart causing increased blood flow to the lungs and complications.

Atrial septal defect: This is the most common heart defect found in children with Down syndrome. There two main types:

- Ostium primum defect - Many children with this type of defect have no symptoms. If symptomatic then you would expect a reduced exercise tolerance with easy fatigue. Babies may require feeding with higher calorie feeds. More frequent chest infections may occur. This would present as mild failure to thrive in babies and reluctance to play in older children. Surgical closure is the definitive treatment and is usually carried out as a baby.
- Ostium secundum defect - Usually diagnosed whilst the baby is in mother's uterus, or it is an incidental finding in a child with no symptoms. When symptoms are present then there will be a subtle failure to thrive in an infant or a slightly reduced exercise tolerance in older children. Surgical closure is recommended between the ages of two and five. Surgery has a low complication rate and has a good clinical effect, restoring babies' growth and improving a child's exercise tolerance.

Ventricular septal defect: This is caused by a hole in the ventricular septum allowing these two chambers to directly communicate.

Small ventricular septal defects close spontaneously usually by the age of 1-2. Small defects are usually asymptomatic with no restrictions on physical activities and surgery is not normally recommended.

Larger or symptomatic defects will be repaired surgically with excellent results. They have a good long-term prognosis. Children in this category may present as babies with failure to thrive or as children with breathlessness, reduced exercise tolerance and more frequent chest infections. These problems are corrected by surgery.

Patent ductus arteriosus: When a foetus is in the womb the lungs are not in use. The oxygen supply is derived directly from the placenta. Blood bypasses the lungs directly into the left-sided heart circuit via a special duct. This is called the ductus arteriosus. Normally, this duct closes shortly after birth but if it fails to close, it is abnormal and referred to as a patent ductus arteriosus.

Children with small defects will usually have no symptoms and a normal lifespan.

Large defects will cause breathlessness, reduced exercise tolerance and failure to thrive. Treatment is a simple operation that will show immediate

improvement in symptoms. Even children with a closed large defect have a good prognosis.

**Tetralogy of Fallot:** There are four parts to these defects. The main two are a large ventricular septal defect and a narrowing of the blood vessel leading from the heart to the lungs. In addition, the right sided ventricular muscle thickens and the main blood vessel from the left side of the heart to the body connects to both ventricles because of the large ventricular septal defect.

As a result of this blood from both sides of the heart mixes before being ejected down both heart circuits. Most of this blood goes around the body because of the narrowing in the blood vessel going to the lungs (causing a blockage to flow). Blood doesn't get enough oxygen, so the child becomes blue (cyanosed).

Children with this heart abnormality become symptomatic by the age of 12 months. They are usually breathless, rest frequently and fail to thrive.

Surgical repair is required and is a complex open-heart procedure. This improves the heart dramatically but will leave them with a slightly reduced exercise tolerance. Follow up is usually long term to monitor their health and growth.

Whilst waiting for surgery children will require iron supplements and additional medication like propranolol (Beta blocker) or digoxin to help control symptoms.

**Mixtures of cardiac abnormalities/other cardiac abnormalities**

Children with Down syndrome may have more than one cardiac abnormality.

In addition, they might have a rare form of congenital heart disease. In these instances, the HCP can be contacted for advice.

**Endocarditis:** This is a bacterial infection on the lining of the heart and heart valves. Precautions against this happening are usually taken when a child with a heart defect has a procedure that releases bacteria in to the blood stream. This includes dental (extractions or drilling, not dental examination or cleaning) work or urological procedures (having a catheter inserted). This is usually done by giving a dose of antibiotics one hour before any risk



prone procedures. There are no disabling features unless the child develops endocarditis which may be life threatening.

Children with isolated atrial septal defects, fully repaired ventricular septal defects or closed patent ductus lesions do not require preventative antibiotics.

### Leukaemia

This is a cancer of the blood cells. The incidence of Leukaemia for children with Down syndrome is 1% - See: Acute Lymphoblastic Leukaemia – (ALL) / Acute Myelogenous (Myeloid) Leukaemia (AML).

Obstructive sleep apnoea also known as sleep related breathing disorder

This condition occurs in 60% of children with Down syndrome.

Obstructive sleep apnoea in children with Down syndrome is characterised by partial obstruction of the upper airways, for example by enlarged tonsils and/or adenoids. The cause of the obstruction is usually treated through surgery and this tends to be very successful. However some children will continue to have significant problems even after surgery and may need non-invasive ventilation at night.

This condition is unlikely to cause night waking but the parent may be woken by noise and observe the child's breathing. There is no need for watching over and there are no night needs associated with it.

Children with significant obstructive sleep apnoea may have frequent drops in their blood oxygen level throughout the night, resulting in regular "arousals." Although they do not wake up fully this situation can lead to their being sleepy during the day, or underperforming at school or having other symptoms such as headaches.

### Craniovertebral instability

Problems can develop if a vertebra slips too far and puts pressure on the nerves in the spinal cord. This can be a gradual process or it can happen if a person is jolted suddenly such as when a whiplash injury occurs. In people with Down's syndrome, the most common place for this slippage to happen is at the first and second vertebrae of the neck (known as 'atlanto-axial instability'). Similar problems can also occur between the base of the skull and the atlas vertebrae (known as 'atlanto-occipital instability') although this isn't as common as atlanto-axial instability. These two conditions are collectively known as craniovertebral instability.

### Coeliac disease

People with Down syndrome have an increased incidence of coeliac disease. Rates vary in different geographical locations. Coeliac disease is an intolerance to the gluten contained in wheat based foods and results in a range of gastro intestinal symptoms and other symptoms such as fatigue. In children it may cause faltering growth. The treatment is the same as for

people who do not have Down syndrome and consists of excluding all gluten containing foods from the diet.

## Diabetes

Type 1 diabetes is estimated to be at least 4 times commoner in children with Down syndrome than in the general population and usually presents at an earlier age (often before the age of 2). Treatment is the same as for other children with diabetes but may be more challenging to implement because of the child's young age and associated learning disability.

## Arthritis

Inflammatory arthritis is thought to be at least 3 times commoner in children with Down syndrome than in the general child population. There is often a delay in diagnosis because the symptoms, e.g. difficulty/reluctance in walking, may be wrongly attributed to the Down syndrome. The treatment is the same as for other children with juvenile idiopathic arthritis.

How is it assessed?

## Diagnosis

Down syndrome is usually diagnosed at or shortly after birth. In most cases, the doctor will be certain of the diagnosis from the child's appearance alone. The diagnosis will be confirmed by an analysis of the baby's chromosomes. Some children will have been diagnosed before birth as a result of screening for Down syndrome.

Once the diagnosis has been made babies with Down syndrome should have some additional medical investigations, for example, an echocardiogram, to detect any associated problems that might be present.

How is it treated and managed?

Children with Down syndrome will often be regularly monitored by a Paediatrician to ensure that all associated health conditions can be diagnosed and treated and learning needs supported. This varies from area to area. In some places a child who has no additional medical needs may no longer be receiving specialist follow up once they start school.

Most children with Down syndrome are receiving support from therapists. They are also very likely to have additional hearing and/or eyesight problems, which will be monitored and treated by the relevant specialist health teams.

Children with Down syndrome who have one or more of the additional medical conditions that can be associated with the syndrome, e.g. congenital

heart disease, will be under the care of the relevant specialist team and will be receiving the standard treatment for that condition.

What evidence is available?

Sources of evidence	Information Provided	Limitation of Evidence
Claimant / Carer	First source of information and associated evidence, for example Statement of SEN/Education Health and Care Plan or Co-ordinated Support Plan (CSP) or an Individual Education Plan (IEP).	Does not have specialist knowledge.
Multidisciplinary community team (Community / Hospital Paediatrician, Consultant psychiatrist, Consultant psychologist, specialist nurse – learning disability, Associated Health Specialists e.g. Occupational Therapist, speech and Language Therapist, Physiotherapist)  (if child is under their care)	Clinical features, treatment and information about disability and needs.  Also request a copy of a completed standardised assessment (for example the Adaptive Behaviour Scale) and a copy of a Statement of SEN/Education Health and Care Plan or Co-ordinated Support Plan (CSP).	Information regarding symptoms and disabling effects may be based on what the parent / carer has told the relevant professional.
Special Needs teacher/coordinator  (if child has one)	Resulting disability or needs.	May not have information about medical symptoms, investigations and treatment but this may vary depending on the type of school the child is attending.

<p>Educational psychologist (if child has seen one)</p>	<p>Educational features, educational intervention required and information about disability and needs.  Also request a copy of a completed standardised assessment (for example the Adaptive Behaviour Scale) and a copy of a Statement of SEN/Education Health and Care plan or Co-ordinated Support Plan (CSP) if available.</p>	<p>May not have up to date information if assessment not recent but this may vary depending on the type of school the child is attending.</p>
<p>Social worker/local authority (if child has one)</p>	<p>Resulting disability or needs. Also request copies of previous assessments including educational assessments if available.</p>	<p>May not have information about symptoms, investigations and treatment.</p>
<p>GP</p>	<p>Clinical features, treatment and some information about disability and needs.  May be the only source of information if no other professional involvement.</p>	<p>Does not have specialist knowledge.  May not have detailed or recent information about resulting disability or needs..</p>

How long will the needs last?

This guidance covers:

<p>Impairment</p>
<p>Down/Down's syndrome</p>

The table below is to be used as a guide to length of award once this decision has been made. Please do not use it as a means of deciding if an award is warranted.

There will be multiple factors influencing needs for a child with Down syndrome. For the sake of a 'broad brush' approach, the single most important

factor that will delineate needs is the level of learning disability that the child has.

If other physical factors complicate a case or indicate a more severe level of functional restriction than would be expected with the learning disability alone, then advice from an HCP is advised.

The following table is to be followed if an award is made. It is not a statement of intended awards for individual categories

Age at date of claim	Length of Award			
	Mild Learning Disability	Moderate Learning Disability	Severe Learning Disability	Profound Learning Disability
1-4 years	Until age 7	Until age 7	Until age 16	Until age 16
5-9 years	Until age 12 or for two years (whichever is greater)	Until age 12 or for two years (whichever is greater)	Until age 16	Until age 16
10-15 years	Until age 16	Until age 16	Until age 16	Until age 16

### Living with Down syndrome - Example Profiles

#### Example 1:

N is a 5-year-old boy attending mainstream school. He has an IQ of 68. He has a mild general learning disability. He has no dedicated additional teaching hours. He has an individual education plan (IEP). Recently at school he has started to struggle due to problems concentrating. His mother has noticed that this is worse when he hasn't slept well. He requires occasional help with his concentration and there is a teaching assistant in his class of 28 who helps him out when he requires it.

N was born with a VSD which initially led to him having frequent chest infections. This has closed spontaneously and causes no health needs.

He goes to bed at around 9pm wakes 2-3 times during the night and finally wakes at around 5am. One or two nights per week he sleeps less and then his daytime behaviour is much worse. His parents usually go to bed at

11pm. He is dry during the day but has episodes of full bed wetting at least 4 times per week.

#### Example 2:

B is an 8-year-old child with Down syndrome. She lives with her two sisters and both parents. She has some problems with her speech and has speech and language therapy 3 hours per week at school. She has an IQ of 50 and attends a mainstream school but has an Education, Health and Care Plan (EHCP). The class has a teaching assistant who gives B one to one support for 2 hours per day. Every 12 months there is a formal review of B's progress so that future goals and education can be adjusted or updated. The class has two teaching assistants.

Her parents collect her from her classroom at the end of the day.

Her sleep pattern is well regulated. She is dependent on her mother for a large portion of her activities of daily living as she struggles with fine motor co-ordination.

B has obstructive sleep apnoea and uses a CPAP machine at night. Once it is set up he usually settles within an hour or so and requires no further supervision.

#### Example 3:

J is 14 years old lives with her parents and younger sister, and attends a special needs school. She has an EHCP. J has an IQ of 58. She is in a class of 6 pupils with one teacher and 3 teaching assistants. She receives 2 hours of OT per week and 2 hours of speech and language therapy. She requires supervision at break times to help her socialise with the other children at the school. Her EHCP is reviewed every year to make sure that the goals and provision laid down in it remain appropriate to J's needs.

She had an atrial septal defect repaired when she was 12 months old. Her growth and exercise tolerance are as would be expected for a child her age who has Down syndrome.

She takes time to settle in the evenings, often staying awake until 9 or 10pm but, once asleep she usually requires no additional settling and wakes around 6am.

#### Example 4:

R is 10 years old. He lives with his parents and two younger brothers at home. He attends a special school and has a Statement of Special Educational Needs with an individual education plan. He is in a class of 12

children, one teacher and three teaching assistants. He gets 2 hours of OT and 1 hour of speech and language therapy each week. He has an IQ of 44.

His parents have a social worker allocated to help co-ordinate with school about his physical and educational needs. He has a weekly visit at home from the social worker.

At age 3, R had a Fallots tetralogy repaired. He has done well but despite this he gets breathless with exertion and requires frequent rests when playing out doors. He attends regular follow up appointments at the hospital for this and needs to be given regular medication to help with his symptoms.

Example 5:

F is 9 years old. She has an IQ of 30. She has severe learning disabilities. She lives in a residential unit for children with learning disabilities. She communicates using basic sign language and limited speech due to hearing loss. She has severe dyspraxia so requires help with dressing and feeding.

She is in a class of 6 children with one teacher and two teaching assistants. She requires one to one teaching, speech and language therapy for 3 hours per week and Occupational therapy for 3 hours per week. In addition she has a Statement of Special Educational Needs. The plan allows for an educational psychologist to attend monthly meetings to discuss her educational needs.

F has problems sleeping and will wake frequently during the night, requiring staff to attend to settle her back to sleep again.

Dyspraxia or Developmental Coordination Disorder (DCD)

What is dyspraxia?

Dyspraxia is one of the specific developmental disorders.

Specific developmental disorders have been defined as a severe and persistent dysfunction in a specific developmental domain. They result in skills being significantly below the expected level for a child's chronological age and cannot be explained by any neurological disorder or specific adverse psychosocial or family circumstances. Other conditions on this class include ADHD, the autistic spectrum disorders, dyslexia, dysgraphia and dyscalculia.

Dyspraxia affects the planning and sequencing of gross and fine movements (motor dyspraxia) and the planning and sequencing of events (non-



motor dyspraxia). Developmental verbal dyspraxia (DVD) probably stems from both motor and non-motor planning difficulties.

Dyspraxia is only diagnosed where the movement problems are not explained by more fundamental neurological conditions such as cerebral palsy or muscular dystrophy.

It affects up to 5-6 % of schoolchildren depending on the threshold for diagnosis.

What are the effects and signs?

Like all specific developmental disorders it has all the following features, but different children are affected to different degrees:

- **Spectral:** both healthy and diagnosed individuals vary in their executive motor control, which means that everyone has a unique place in a spectrum of ability and the borderline between health and illness is not black and white.
- **Overlapping:** children with dyspraxia commonly have features of several other specific developmental disorders – either ‘full blown’ or in a form too mild to result in a separate diagnosis.
- **Associations:** children with any brain injury can be affected in more than one way so dyspraxia is associated with epilepsy, cerebral palsy, cognitive disability and so on. All of these conditions are more common in children who were born prematurely or have suffered brain trauma of any kind.
- **Sensory processing problems:** children with dyspraxia also frequently struggle to prioritise and organise sensory information, which can result in poor sensory prioritisation (not paying attention to the right things) and sensory overload (paying attention to the wrong things).
- Short term memory problems
- Hypotonia: low resting body tone

Various areas of development can be affected by dyspraxia and these will persist into adulthood. Often, various coping strategies are developed, which can be enhanced through occupational therapy, psychomotor therapy, physiotherapy, speech therapy or psychological training.

Resulting disability

Motor skills

The most obvious disability (and the one used for diagnosis) is that activity dependent on movement planning can be affected. Hypotonia can make these difficulties worse. Late development of walking, frequent unexplained

tripping, failure to attain smooth, efficient running patterns or to learn cycling, failure to acquire or to perfect fine-motor skills such as machine operation and handwriting and so on are typical.

### Personal organisation

A number of factors contribute to frequent failure in basic activities of daily living. These include poor executive oversight of activities (keeping the important things foremost and the trivial things at the back of the mind), poor short-term memory, being overwhelmed by unimportant sensory information and the additional effort required to carry out motor tasks. Children typically cause their parents and teachers frustration by failing to achieve basic tasks such as brushing their teeth or dressing. This is especially the case with multi-stage tasks or tasks requiring prioritisation.

### Communication

Developmental verbal dyspraxia is also probably multifactorial, key elements include:

- difficulties controlling the speech organs.
- difficulties making speech sounds
- difficulty sequencing sounds within a word
- forming words into sentences
- difficulty controlling breathing, suppressing salivation and phonation when talking or singing with lyrics
- slow language development

### Psycho-social

Children and adults with dyspraxia have a lifelong experience of frequently meeting with failure and the frustration of others. This takes a toll on their wellbeing and they are at higher risk of financial failure, depression, anxiety and suicide than the general population.

### How is it assessed?

Assessments for dyspraxia typically require a developmental history, detailing ages at which significant developmental milestones, such as crawling and walking occurred. Motor skills screening includes activities designed to indicate developmental coordination disorder, including balance, physical sequencing, touch sensitivity and variations on walking activities. The other purpose of the examination is to exclude a more fundamental cause of the motor difficulties such as cerebral palsy or muscular dystrophy.

A baseline motor assessment establishes the starting point for developmental intervention programs. Comparing children to normal rates of

development may help to establish areas of significant difficulty. Currently there is no single gold standard assessment test.

The diagnosis of dyspraxia in a child is usually made following assessments by a paediatrician and an occupational therapist.

### Treatment

There is no cure for dyspraxia and the condition is life long. Treatment aims to teach child strategies for managing its effects. Over time most children with dyspraxia do learn to cope with their condition better and are less severely affected by it.

Treatment approaches may involve specific motor skills programmes, for example to help improve a child's balance or handwriting or the teaching of strategies to assist with planning, e.g. using checklists. Children may benefit from the use of equipment, e.g. a laptop at school for those with severe

hand writing difficulties, or specialised cutlery. Older children with dyspraxia may be allowed additional time in formal school exams

How long will the needs last?

If a child's needs for this condition mean that they qualify for an award, then an advised duration would be for 2 years in the first instance.

If longer awards are considered then the advice of an HCP should be sought.

## What you need to know about Eczema

### What is Eczema?

Eczema is a dry itchy skin condition. Boys and girls are equally affected  
.....

- [Eczema](#)

### What are the effects and signs?

The hallmark of eczema is that the skin is itchy. Affected areas may look dry, flaky or red.....

- [Effects of Eczema](#)

### How is it assessed?

A clinician can diagnose eczema and assess its severity by looking at it.  
.....

- [Assessment of Eczema](#)

### How is it treated and managed?

The main treatment of eczema is emollient cream .....

- [Treatment for Eczema](#)

### What evidence is available?

The best source of information on children with more severe eczema will be the eczema specialist nurse or the consultant dermatologist or paediatrician.

- [Evidence](#)

### How long will the needs last?

- [Prognosis and duration of the award](#)

## What is Eczema?

Eczema is a dry itchy skin condition. Boys and girls are equally affected and the condition often runs in families. The condition typically starts in the first few months of life and usually before the age of 3. The skin becomes dry and flaky and is likely to be itchy. The skin barrier does not function normally and this makes the skin susceptible to otherwise harmless trigger factors, including irritants and allergens which make the eczema worse. Eczema is a relapsing and remitting condition through early childhood. Most children grow out of it. Typical features of eczema include -:

- Red skin
- Dry skin
- Itchy skin
- Small watery blisters on the skin – these can weep, particularly when on the hands and feet
- Scaly thickened patches of skin, especially areas of the skin which are scratched regularly – this may be termed 'lichenification' in the medical evidence

Scratching skin that is itchy leads to abrasion of the skin, abraded areas feel itchier as they heal – this is the itch scratch cycle. Children with itchy skin may not sleep or feed well. Areas of flaky, dry or weeping skin may become infected with bacterial organisms or blistered when involved in herpes

virus infection – both of these are acute complications of eczema and require urgent medical treatment.

Children in the toddler age group are most likely to have eczema. The areas of skin most commonly affected include -:

- Backs of knees
- Front of elbows
- Chest
- Face
- Neck

The majority of cases will be mild and children will grow out of their condition around the time they begin primary school.

Eczema is a chronic condition that can be controlled with treatment. For the majority of children eczema is well controlled with treatment, with occasional flare ups. Treatment needs to be stepped up during flare ups and stepped down when the skin is under control. This applies to any severity of eczema; baseline treatment is used when skin is good and step up treatment used when eczema flares up. Treatment is likely to be more time consuming in a severe case than a mild case. A small number of children will have moderate or severe eczema. They are less likely to grow out of their eczema completely but it is likely to improve over the years. Small numbers of children will have eczema that persists into adulthood. The majority of children with eczema lead normal healthy lives, controlling their eczema with effective medication. The time taken by parents and carers to provide care to control the skin condition is the basis of care needs in this condition.

### **Incidence/Prevalence**

1 in 5 children under 3 in the UK have symptoms of eczema. The majority of children (over 80%) have mild eczema. 15% of children with eczema will have moderate eczema and 5% of the children with eczema will have severe eczema.

Eczema follows a typical pattern in most children. The vast majority of children with eczema will develop the condition under the age of 3 years. It is most likely to be severe in this age group. Children will then grow out of their eczema, with the mildest cases resolving earliest. 50% of cases will have resolved by age 5. Children with moderate to severe eczema are likely to take longer to grow out of their eczema completely, however the eczema often improves over the years; a child with severe eczema at age 3 may have moved into the moderate category by age 6. Children with the most

severe eczema are likely to continue to have eczema to some degree. Eczema often improves substantially at puberty.

### **What are the effects and signs?**

Diagnosis of eczema is based on examination of the skin by a clinician. The criteria listed here are those commonly used to diagnose atopic eczema in children. The hallmark of eczema is that the skin is itchy. Affected areas may look dry, flaky or red. There may be scratch marks or skin thickening.

Eczema may be limited to small areas of skin or affect most of the body. The following features are commonly present:

- Previous history of eczematous rash in the skin creases e.g. bend of elbow, behind knees
- Personal or family history of [asthma](#) or hay fever
- Tendency towards dry skin
- Flexural eczema (affecting backs of knees, front of elbows and inner side of wrist) - currently present or in the past
- Onset of skin condition under age 2 years

Other features which may be present -:

- Skin condition aggravated by contact with cats or dogs
- Skin condition aggravated by exposure to irritants such as soap and household cleaning chemicals
- Disturbed sleep due to itching and scratching
- Impact on quality of life
- Effect on schoolwork or family
- Bullying

Other conditions which increase care needs related to eczema i.e. care needs present in mild to moderate eczema or beyond age 10:

- Any diagnosed behavioural condition which makes compliance with treatment on a daily basis difficult or prolonged.
  - When eczema is one of a number of conditions which in combination require extra care and attention on a daily basis.
  - Where there is a physical disability affecting upper limbs and application of emollients cannot be done independently.
- 
- [Terms used to describe eczema](#)
  - [Assessing Needs](#)
  - [Indicators of a severe functional restriction](#)



## **How is it assessed?**

Usually no investigations are performed to diagnose eczema. The condition affects the skin and has a typical appearance. A clinician can diagnose eczema and assess its severity by looking at it. Most children are managed in primary care by the GP. Some children with more severe eczema need specialist advice or treatment and these children will be referred to a dermatologist. Reasons for referral include:

- Severe eczema not responding to treatment
- Infected eczema that has not responded to treatment
- Diagnosis is uncertain
- Uncontrolled eczema as assessed by child or carer
- Eczema on the face that has not responded to treatment
- An allergic cause for eczema is suspected e.g. contact allergy or food allergy
- Eczema is causing significant social or psychological problems for the child or family
- Repeated or severe infection of eczematous areas

A specialist referral may be required in cases where eczema is controlled with treatment but -:

- Impact on quality of life or psychosocial well-being has not improved (psychological advice)
- Failure to grow as expected

## **Allergy testing**

Where contact or food allergy is suspected tests can be performed. 10% of children with eczema have a food allergy, eating the food makes their eczema significantly worse and removing the food from the diet often results in dramatic improvement of the eczema. The commonest food allergens responsible for this are called the 'big eight', they are: peanuts and tree nuts, eggs, cow's milk, wheat, fish and soya. Children are likely to grow out of

most of these allergies except nut and fish allergy which are likely to persist into adult life. Allergy tests may include the following -:

Blood tests:

Specific Immunoglobulin E (IgE) allergy to foods e.g. the big eight and any other suspected foods.

Total IgE – if the serum IgE level is very high, allergy may be present and skin prick testing is usually done as a further test.

Skin tests -:

Skin prick tests – are carried out and read on one clinic visit – the skin is pricked through a drop of liquid containing the allergen, swelling of the pricked area indicates allergy. Skin prick tests are used to identify food and inhalant allergies.

Skin patch tests – small amounts of suspected allergen are applied to the skin and protected by a dressing. The dressing is removed after 48 hours and evidence of irritation or inflammation is assessed. Patch testing is used to identify contact allergy to common chemicals and preservatives.

High street and internet allergy testing results do not constitute evidence of allergy.

### **Assessment of clinical severity**

There are clear guidelines on assessment of eczema and categorising it into the mild, moderate or severe category. The guidelines include an assessment of the effect of eczema on quality of life as well as assessment of the skin condition. Severity of the eczema as recorded in the medical evidence will be an important indicator of care required. It will be based on

both an assessment of the skin and the effect it is having on the child and their family.

Quality of life assessment -:

- No effect - no impact
- Mild - little impact on everyday activities, sleep and psychosocial well-being
- Moderate - moderate impact on everyday activities and psychosocial well-being and frequently disturbed sleep
- Severe - severe limitation of everyday activities and psychosocial activities, nightly loss of sleep

Assessment of skin:

- Clear - normal skin
- Mild - areas of dry skin, infrequent itching with or without small areas of redness
- Moderate - areas of dry skin, frequent itching and redness with or without excoriation and localised skin thickening
- Severe - widespread areas of dry skin, incessant itching and redness with or without excoriation, extensive skin thickening, bleeding, oozing, cracking and altered skin pigmentation

### **How is it treated and managed?**

The main treatment of eczema is emollient cream. These are non-irritant moisturising creams that hydrate and soothe dry, itchy skin. Use of emollients alone will often be enough to control eczema most of the time. Emollients may be used once a day – typically after washing or bathing to retain moisture in the skin, or they may be used four times a day in severe eczema. Stepping up emollients to several times a day will be the main step up treatment in eczema. Emollients are applied thickly and smoothed onto the skin rather than rubbed in. Use of soap is avoided in eczema and either emollient cream is used for washing and cleaning the skin or emollient bath

additives are used. These can be bought over the counter or prescribed. This is a list of commonly used emollients but there are many others:

<b>Names of emollient creams and ointments</b>	
Aqueous cream BP	Diprobase
Cetraben	Drapolene
Hydrous Ointment BP	E45
Liquid and White or Yellow Soft Paraffin Ointment	Oilatum Epimax
White Soft Paraffin BP	Ultrabase
Yellow Soft Paraffin BP	Epaderm
Aveeno	
<b>Names of emollient bath additives</b>	
Alpha Keri bath	Oilatum Junior Emollient bath additive
Aveeno Bath Oil	E45 emollient bath oil
Balneum bath oil	

### Bandages or wraps

These are applied after emollients usually at night to improve hydration of the skin and reduce itching. Bandages may be dry or wet or medicated. Bandages are a time consuming treatment. Eczema needs to be kept very clean to prevent secondary infection, so a bath is needed every day. A typical routine for a child with eczema having bandage treatment would include washing in a bath using an emollient cream or emollient bath additive. Patting the skin dry followed by the application of emollient cream to retain hydration of the skin. Waiting half an hour and then applying treatment creams. Only then can the bandage or wrap be applied. The bandage may be wet or dry or medicated or may be an elasticated stockinette. The whole process from bathing to bandaging and being ready for bed may take around 90 minutes to complete. The following morning the bandages are removed. This may take time and patience particularly if the eczema is weepy and the bandages have become stuck to areas of the skin, another bath

may be required to soak some bits of the bandage off. Once the bandages are removed further emollient and treatment creams need to be applied before the child can be dressed for the day. Typically, the morning routine will take around 30 minutes. Children having bandage treatment on a daily basis rather than just during flares are likely to require repeated application of

emollient cream during the day every day; the child will have to be undressed on each occasion that this is applied.

Treatments used in mild eczema

- Emollients – usually up to twice daily
- Mild potency topical corticosteroids up to twice daily

During flare ups treatment may be stepped up -:

- Emollients more than twice daily
- Moderate potency topical corticosteroids up to twice daily
- Tacrolimus or Pimecrolimus ointment
- Bandages

Treatments used in moderate eczema

- Emollients up to 4 times daily
- Moderate potency topical corticosteroids
- Tacrolimus or Pimecrolimus ointment
- Bandages

During flare ups treatment may be stepped up:

- Potent topical corticosteroids
- Systemic therapy (the use of oral drug treatments to control a condition)

Treatments used in severe eczema

- Emollients up to 4 times daily
- Potent topical corticosteroids
- Tacrolimus or Pimecrolimus ointment
- Bandages
- Phototherapy (treatment involving exposure of the skin to light)
- Systemic therapy

[Corticosteroid creams](#)

[Other therapies and treatments](#)

**What evidence is available?**

Source of Evidence	Information Provided	Limitation of Evidence
Parent / Carer	First source of information.	Does not have specialist knowledge.
GPFR	<p>Most children receive their treatment and follow up in Primary Care. Only those children with the most severe eczema for example on systemic therapy are likely to be managed in Secondary Care. For the majority of children all treatment information will be available from the GP. Those children with severe eczema are likely to receive some medication from the GP and the severity of their eczema can often be confirmed with this information alone.</p> <p>Information of particular value will include -:</p> <ul style="list-style-type: none"> <li>• Reviewing whether repeat prescriptions have been requested – ongoing evidence of use of emollients (200-500g) weekly is good evidence of active eczema treatment.</li> <li>• Information on severity of eczema and current treatment from most recent clinical review (within last 12 months)</li> </ul>	<p>More detailed information will be available for children with mild eczema who are managed in primary care. The GP may only be able to confirm medication prescribed for children with more severe eczema managed in secondary care, this is likely to be sufficient evidence. More information is likely to be required for children with moderate eczema aged 6 and over as they may or may not have care needs.</p>



HFR	<p>The best source of information on children with more severe eczema will be the eczema specialist nurse or the consultant dermatologist or paediatrician. They will be able to provide written confirmation of specific advice on treatment given to parents by the hospital and provide up to date details of current medication and eczema control. Particularly useful information will include -:</p> <ul style="list-style-type: none"> <li>• Confirmation of advice given on number of times per day emollients should be applied.</li> <li>• Whether bandaging is used.</li> <li>• Frequency and severity of eczema flare-ups.</li> </ul>	N/A
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### How long will the needs last?

Age at date of claim	Award Period
Under 1	Award to age 3
1 - 5	Award to age 6 (or for 1 year whichever is the longer)
6 - 9	Award to age 10 (or for 1 year whichever is the longer)
10 - 14	2 year award
15	Award to age 16 or for 1 year whichever is the longer

What is prognosis and duration for children aged under 1?

Children awarded care on the basis of eczema when they are aged under 1 may have substantially improved by age 3. A significant proportion of such children would be expected to have improved over the period and may now have clear skin or mild eczema. Those who still have care needs should be renewed until the 6<sup>th</sup> birthday. The 6<sup>th</sup> birthday is chosen because eczema often flares up badly when children start school around the 5<sup>th</sup> birthday.

What is prognosis and duration for children aged 1 to 5?

Children aged 1 and over awarded care on the basis of eczema should be awarded to the 6<sup>th</sup> birthday. A significant proportion of such children would be expected to have improved over the period and may at age 6 have clear skin or mild eczema.

What is the prognosis and duration for children aged 6 and over?

Children with moderate eczema at age 6 are likely to be in the improving group, care needs are not expected to persist beyond age 10. Renewal at age 10 is suggested.

Children in the severe category are more likely to have ongoing eczema at age 10 but many of them will have substantially improved. Those who have not improved are likely to be able to manage the majority of their treatment themselves including use of bath time treatments and application of emollients although help with application of emollients, to hard to reach areas may still be required.

Children on systemic therapy (the use of oral drug treatments) will require supervision to ensure that they take their medication at the prescribed dose on a regular basis. They will often need frequent clinical review and blood test monitoring. These children are likely to be those with the most severe eczema. Children are generally able to manage this type of treatment themselves at 12 and children receiving care awards on the basis of eczema aged 10 and over should be renewed every 2 years with fresh medical evidence. The majority of children who still have eczema beyond middle childhood can be expected to improve significantly at puberty.

Children up to 16 years of age

Children with additional conditions such as a learning difficulty or a behavioural disorder, may not be able to manage their treatment at the above ages without help and supervision.

If other particular circumstances are stated to be present preventing self medication, each case must be assessed individually. Confirmation of such

circumstances should be obtained from the treating Paediatrician and duration of award details discussed with Medical Services.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Terms used to describe Eczema**

Different names may be used for eczema depending on specific features of the skin condition. All of these terms are used to describe forms of eczema

and are covered by the blanket term 'Eczema' used in this guidance. These names and some terms are explained or defined below.

Eczema	An umbrella term for a group of skin conditions where the skin is dry itchy, irritated or inflamed.
Atopic eczema	The commonest type of eczema - the condition runs in families.
Atopic dermatitis	Another name for Atopic eczema.
Flexural eczema	Eczema on backs of knees, front of elbows and inner side of wrist.
Infantile eczema	Eczema that begins in infancy.
Seborrheic eczema or dermatitis	This is when the rash is greasy rather than dry. It usually begins on the scalp in babies and can spread to other areas - often it resolves by age 1.
Cradle cap	Seborrheic eczema affecting the scalp in babies – common in babies under age 1.
Allergic contact eczema	Eczema that develops due to contact with an allergen in a person with an allergy e.g. eczema in a cat allergic person after stroking a cat.
Irritant contact eczema	Eczema that develops after skin contact with irritant substances such as house hold cleaners – no allergic mechanism is involved.
Discoid eczema	Round scaly patches of eczema commonly on the arms and/or legs.
Pompholyx	A type of eczema that affects the palms of the hands and sometimes the soles of the feet. It causes small blisters which resemble frogspawn.
Eczema herpeticum	An acute condition – large areas of damaged skin become infected with herpes virus. The child is likely to be very unwell and hospital treatment is required.
Cellulitis	An acute infection with bacteria of the deeper layers of the skin. Requires treatment with antibiotics.

### **Eczema triggers and their avoidance**

Children with eczema have dry skin that is easily irritated by soaps and shampoos, household cleaners and clothes containing wool. 90% of children with eczema are allergic to house dust mites. Minimising exposure to these in the home is very difficult because contact with a small amount of allergen sufficient to exacerbate eczema is hard to prevent. Around 10% of children have a food allergy; avoidance of the relevant food can significantly improve the eczema.

Molluscum contagiosum is a common viral infection of the skin in small children. The condition causes crops of small wart-like bumps to appear on the skin. It is generally harmless but itching and scratching of these areas in children with eczema can lead to broken skin, areas of cellulitis and patches of difficult to control eczema. Molluscum can exacerbate eczema for several years.

Triggers may significantly aggravate eczema. Some such as house dust mites (HDM) will be difficult to avoid. Others such as household cleaners and woollen garments are easy to avoid. Parents are likely to put the most effort into avoiding food allergy triggers. This is because it is an effective way of minimising eczema in a child with a food allergy and because they will have received medical advice that it is important to do this.

### **Assessing needs in relation to severity of Eczema**

The main evidence to assess severity of eczema will be the label given to the eczema in the medical evidence e.g. 'moderate' eczema. This will give a good indication of what treatment is being given and this can be confirmed using the medical evidence.

Treatment changes depending on severity of eczema at most recent clinical review. Eczema may become more severe over time, for example moving from mild eczema into the moderate category. Alternatively it may remain mild, moving into the moderate category only during flare ups. When reviewing medical evidence consider what category the child's eczema is in, the majority of the time. Children in the moderate category may move into the severe category during flare ups. Flare ups may be infrequent – the child will be in the moderate category for care needs most of the time. Flare ups may be present for 2 weeks out of every 4 – the child will have eczema that is labelled 'moderate' at the moment but have care needs of a child with severe eczema most of the time. The category used for care needs assessment should be the category the child is in for at least half of the time.

### **Children aged 5 and under**

#### **Mild Eczema**

No care needs are anticipated for children with mild eczema. Children with mild eczema will require daily application of emollients and use of steroid creams or other treatments during flare-ups. Bath time emollients or soap substitutes may be used. Emollients won't need to be applied more than

twice a day for the majority of the time and this can be done when the child is being dressed or undressed at the beginning and end of the day.

### **Moderate and Severe Eczema**

Children aged 5 and under with moderate or severe eczema are likely to have care needs. Care is mainly related to providing daily treatment. The mainstay of treatment is the application of emollients to the skin. In a severe case these may need to be applied 4-6 times daily to the whole body. The child will need to be undressed on each occasion, this can be done on each occasion the nappy is changed in young children, but this adds considerably to the time taken to change a nappy and means significant amounts of emollients or other creams will need to be transported wherever the child goes. It may be difficult to find a childcare provider for such children.

Wet wraps or bandages may need to be used at night, this prolongs bath time and preparation for bed significantly. In the morning removal of bandages can be prolonged and difficult, especially if the eczema is weeping and areas of bandage have become stuck to the eczema overnight. These two treatments make the most significant contribution to the extra time taken to care for a child with eczema.

In addition to these treatments other treatments may be required and these include:

- Wound care.
- Use of bath time treatments instead of soap and shampoo to promote skin hydration.
- Use of targeted steroid creams to worst affected areas, different potencies of cream may be used for different areas of the body.
- Antihistamine drugs at night to aid sleep and reduce itching.
- Systemic treatment with immunomodulating drugs such as ciclosporin or steroids.
- Children who are unable to sleep properly because of itchy skin or who look different to other children may develop behavioural problems requiring extra care or supervision from parents in addition to physical care provided.
- Children with eczema due to food allergy are likely to require extra supervision from parents or carers in the pre-school years particularly to prevent ingestion of the foods. It may be difficult to find a childcare provider for such children.

### **Children aged 6 and over**

Children with severe eczema will have care needs and children with moderate eczema with certain features are also likely to have care needs. In both

moderate and severe cases, care is mainly related to providing daily treatment. The mainstay of treatment is the application of emollients to the skin. In a severe case these may need to be applied 4-6 times daily to the whole body. On each occasion, the child will need to be undressed and provision may need to be made to provide treatment during the school day. The child may do this themselves under supervision at school or the parent may come in to school to apply emollients during the day. This is because schools cannot provide a suitable person to apply emollients to children during the school day. Similar arrangements will be made for swimming. Emollients need to be applied both before and after swimming and either the parent will come in to do this or school staff will supervise the child doing this for themselves.

Out of school, it may be difficult to find a childcare provider for such children. Wet wraps or bandages may need to be used at night prolonging bath time and preparation for bed significantly. In the morning, removal of bandages can be prolonged and difficult especially if the eczema is weeping and areas of skin have become stuck to the eczema overnight. These two treatments, application of emollients 4 or more times a day and bandaging make the most significant contribution to the extra time taken to care for a child with eczema.

In addition to these treatments other treatments may be required and these include:

- Wound care.
- Use of bath time treatments instead of soap and shampoo to promote skin hydration.
- Use of targeted steroid creams to worst affected areas, different potencies of cream may be used for different areas of the body.
- Antihistamine drugs at night to aid sleep and reduce itching.
- Systemic treatment with immunomodulating drugs such as ciclosporin or steroids.
- Children who are unable to sleep properly because of itchy skin or who look different to other children may develop behavioural problems requiring extra care or supervision from parents, teachers or carers in addition to physical care provided.

Identifying children with moderate eczema who have care needs

Many children with moderate eczema will not have care needs. A child with moderate eczema with any one of the following features identified from the

medical evidence is likely to have care needs related to their eczema management:

- 1 admission to hospital in the previous year with eczema.
- 2 or more hospital out-patient appointments in the previous 12 months.
- 4 or more GP attendances over previous 12 months with problems related to their eczema.
- a recent prescription (within 6 months) or a repeat prescription for antihistamines at night.

### **Indicators of severe functional restriction**

The indicators of severe functional restriction for children are applied as follows; children with severe but well managed eczema may be very well and minimally restricted by their condition. This may be achieved by several hours per day of eczema related skin care by parents or carers. Indicators of severe functional restriction in eczema refer to evidence that significant care is being given. Children may still have limiting eczema despite receiving excellent and time consuming care. Any one of the following features indicates significant care is likely to be being given to the child with eczema:

Children aged 5 and under:

- Systemic therapy with any of the drugs listed in the '[Other therapies & treatments](#)' section
- Use of bandages or wraps
- Written evidence of advice from eczema specialist nurse or dermatologist that emollients should be used 4 or more times daily on an on-going basis to control eczema
- Referral to paediatric dermatologist within last 12 months
- Wound care management plan in place

Children aged 6 and over:

- Systemic therapy with any of the drugs listed in the '[Other therapies & treatments](#)' section

Children aged 6 and over with severe eczema are likely to be receiving significant eczema related skin care. Most children with moderate eczema will be reasonably well controlled and require a lot less care than a child with severe eczema. However there will be some children who appear to be in



the moderate category who do require significant care. These can be identified by looking for evidence of the following in the medical evidence:

- Use of bandages or wraps.
- Under care of dermatologist with a hospital out-patient attendance twice in the last 12 months – children of this age with moderate eczema would not normally see a hospital dermatologist as in most cases eczema is controlled and improving. Those who are seeing a hospital dermatologist are likely to have difficulty to control eczema or eczema that is getting worse.
- One admission to hospital in previous 12 months with eczema – children of this age are very unlikely to be admitted to hospital unless their eczema is hard to control or getting worse.
- Prescription of night time antihistamine tablets on an ongoing basis – this identifies a small proportion of children with very disturbed sleep, such children are likely to have eczema at the more severe end of the moderate category and require more care. The antihistamines, if taken, should reduce care required at night.
- More than 4 consultations with the GP about eczema in the last 12 months – children of this age will usually have well controlled eczema that is improving, more than 4 consultations with the GP about the eczema in one year implies that the eczema is hard to control or is significantly affecting the child e.g. behavioural problems. Significant care is likely to be being given in these cases.
- Wound care management plan in place – a wound care management plan is a nursing care plan relating to the care of a wound. It means that the child's skin has broken down into a wound or open sore. The plan is likely to include details of frequency of wound cleaning and dressing, dressing prescription details and brief notes on the appearance and progress of wound healing.

### **Corticosteroid creams**

The treatment of eczema is a step-wise process. Other treatments are add-on treatments to emollients. Treatment is stepped up and down depending

on the severity of the eczema. All other treatments are used in addition to emollients and include -:

### **Mild potency topical corticosteroid creams**

These are used in small amounts as a spot treatment for a limited period on more active patches of eczema. Mild potency is commonly used intermittently in mild eczema.

<b>Mild potency topical corticosteroid creams</b>
Hydrocortisone 0.1-2.5% - Dioderm, Efcortelan, Mildison
Mild with antimicrobials – Canesten HC, Daktacort, Fucidin H, Econocort, Nystaform HC, Timodine, Vioform-Hydrocortisone.

### **Moderate potency topical corticosteroid creams**

These are used in small amounts as a spot treatment for limited period (3-5 days) on more severe patches of eczema. They are likely to be used in moderate or severe eczema.

<b>Moderate potency topical corticosteroid creams</b>
Betnovate-RD, Eumovate, Haelan, Modrasone, Synalar 1 in 4 dilution, Synalar 1 in 10 dilution, Ultralanum Plain,
Moderate with antimicrobials – Trimovate

### **Potent and Very potent topical corticosteroid creams**

These tend only to be used in severe eczema.

<b>Potent topical corticosteroid creams</b>
Betamethasone valerate 0.1% - Betacap, Bettamousse, Betnovate, Diprosone.
Beclometasone dipropionate – Propaderm.
Hydrocortisone butyrate – Locoid, Locoid crelo, Metosyn, Nerisone, Synalar
Others – Metosyn, Halciderm topical, Elocon, Cutivate
Potent with antimicrobials – Aureocort, Betnovate-C, Betnovate-N, FuciBET, Locoid C, Lotriderm, Synalar C, Synalar N, Tri-Adcortyl, Acorvio Plus
Potent with salicylic acid – Diprosalic

<b>Very potent topical corticosteroid creams</b>
Dermovate, Nerisone Forte

**Topical calcineurin inhibitor cream or ointment**

These are used in small amounts as a spot treatment for a limited period on more severe patches of eczema. Tacrolimus ointment (protopic) is the name of one sort of commonly used topical calcineurin inhibitor.

Pimecrolimus cream (Elidel) is an alternative. It tends to be used when steroid creams cannot be used.

**Other therapies and treatments**

**Phototherapy**

There are two types of phototherapy, both involving exposure of the skin to light for a short period, usually on an outpatient basis. UVB light therapy involves exposing the skin to UVB light and PUVA involves exposing the skin to UVA light after taking a drug called psoralen. The side effects of these treatments include sun burn type immediate reactions, potential long term side effects include premature skin aging and skin cancers. Because of concerns about long term side effects it is only likely to be used in children with severe eczema.

**Systemic therapy**

This means the use of oral drug treatments to control eczema. These include oral steroids and immunomodulating drugs such as ciclosporin and mycophenolate mofetil. These drugs work by damping down the immune system and so improving eczema. Steroid drugs do this very effectively but

their use is limited by their side effects which in children includes growth retardation. Often other immunomodulating drugs are used to control eczema and limit the amount of steroid drug a child is exposed to. These too can have serious side effects and may require regular monitoring for toxicity. All of these drugs impair the immune response to infection. Systemic therapy is reserved for the most serious cases of eczema.

<b>Drugs used for systemic therapy of eczema</b>		
<b>Name of drug</b>	<b>Type of drug</b>	<b>Side effects and monitoring</b>
Steroids e.g. Prednisolone	Immunosuppressant	Usually used in short courses to minimise side effects, which include osteoporosis, high blood pressure, diabetes, adrenal suppression, psychiatric problems and impaired growth.
Ciclosporin	Immunomodulating drug	May cause kidney damage – regular blood monitoring is required.
Azathioprine	Immunomodulating drug	May cause bone marrow suppression – regular blood monitoring required. May cause impairment of liver function and jaundice.
Mycophenolate mofetil	Immunomodulating drug	May cause bone marrow suppression – regular blood monitoring required. Increased risk of skin cancer.

### **Other treatments used**

There are other treatments used for eczema that are not part of the step treatment process that may be used at any stage -:

#### **Antihistamines**

These are oral drug treatments that can reduce itching particularly when itching is severe and where there is associated urticaria (a rash often caused by contact with something to which a person is allergic). There are two types of antihistamines used in eczema. Sedating antihistamines promote sleep in addition to reducing itch and can be used at night to aid

sleep. Non-sedating antihistamines can be used during the day the day to reduce itch.

<b>Names of Antihistamine medication</b>	
Non-Sedating	Cetirizine, Desloratidine, Fexofenadine, Levocetirizine, Loratidine, Mizolastine
Sedating	Alimemazine, Chlorpheniramine (Piriton), Hydroxyzine, Promethazine

## **What you need to know about Epilepsy**

### **What is Epilepsy?**

Epilepsy is a disorder of the brain in which clusters of nerve cells (or neurones) .....

- [Epilepsy](#)

### **What are the effects and signs?**

An epileptic seizure is a brief episode of symptoms and signs caused by .....

- [read more about the effects and signs](#)

### **How is it assessed?**

The diagnosis of epilepsy is almost entirely based on a history of what has happened and any eyewitness accounts.....

- [Assessment of Epilepsy](#)

### **How is it treated and managed?**

Epilepsy is eminently treatable with medication and the aim of treatment is to totally suppress seizures.....

- [Treatment for Epilepsy](#)

### **What evidence is available?**

Evidence from the paediatric neurologist, epileptic nurse or GP gives a profile of the seizures i.e. type, frequency, injuries sustained.....

- [Evidence](#)

### **How long will the needs last?**

- [Prognosis and duration of the award](#)

### **What is Epilepsy?**

Epilepsy is a disorder of the brain in which clusters of nerve cells (or neurones) send off abnormal electrical signals.

Normally, neurones send electrical signals that act to produce thoughts, feelings, sensations and actions. When a seizure occurs, the normal pattern of brain activity is disturbed causing strange movements, sensations,

emotions and behaviour. Convulsions may occur with muscle spasms and loss of consciousness.

Epilepsy is not caused by mental illness or learning difficulties although it is much more common in individuals with learning difficulties (at least 25% have epilepsy).

Having a seizure does not necessarily mean that a child has epilepsy. Only when a child has 2 or more seizures are they considered to have epilepsy. Adolescents may grow out of epilepsy and some patients (of any age) may

go many years without a seizure; 60% of patients may discontinue medication.

### **Incidence/Prevalence**

- Epileptic seizures, including febrile convulsions occur in 3-5% of children
- Epilepsy starts in childhood in 60% of cases and most of the clinically significant aspects of the disease occur during childhood
- Many children who experience a first seizure may never experience a second seizure. However, a seizure may be the initial presentation of a more serious medical condition

In one study from Finland, the point prevalence of active epilepsy was found to be 3.93 per 1000 in children aged 0 -15 years.

### **What are the effects and signs?**

An epileptic seizure is a brief episode of symptoms and signs caused by the response to an abnormal electrical discharge in the brain.

These symptoms may include disturbances of consciousness, behaviour, emotion, movement or sensation. Older words for seizures include convulsions, fits, attacks and turns.

Epilepsy is the tendency to have recurrent seizures even if a long interval separates attacks.

Typically, a seizure lasts from a few seconds to a few minutes. Following a seizure the individual normally recovers completely. Seizures may be stereotyped for a given patient.

The time taken for complete recovery from an attack may vary from a few minutes to several hours, days, or even a week. There is a wide range of effects, some of which are minor.

- [Classification of epilepsy](#)
- [Generalised Seizures](#)
- [Partial Seizures](#)
- [Non epileptic seizures](#)

### **How is it assessed?**

The diagnosis of epilepsy is almost entirely based on a history of what has happened and any eyewitness accounts. An Electroencephalogram (EEG) and other scans can help but do not in themselves give a diagnosis. In ninety percent of cases, the EEG is normal because seizures are



intermittent and sometimes hospitalisation is necessary for monitoring and observation. An accurate description of seizure might give clues as to whether the seizure was a partial seizure with secondary generalization versus a primary generalized seizure.

When evaluating a child who has experienced a first seizure, the clinician needs to address the following -:

- An identifiable aetiology (cause) to the seizure
- The most appropriate therapy
- The prognosis

### **Differential diagnosis**

Many disorders can mimic seizures in children and should be considered in the differential diagnosis of first seizure in a child. The most common non-epileptic disorders include the following -:

- Syncope or breath-holding spells
- Migraine
- Benign paroxysmal vertigo
- Staring spells
- Movement disorders including tics, benign myoclonus, and dyskinesias
- Sleep disorders such as night terrors
- Febrile seizures - These are convulsions brought on by a high fever, typically above 38.5° C in the absence of central nervous system infection and affect 2-5% of children in the first 6 years of life with peak incidence between 18 months and 4 years. 3-6% of patients with febrile seizures will develop afebrile seizures or epilepsy. Children prone to febrile seizures are not considered to have epilepsy, (since epilepsy is characterized by recurrent seizures that are not triggered by fever.)

If the description is consistent with a seizure a number of tests may be important in helping to confirm the diagnosis of epilepsy, including the following -:

### **Electroencephalogram (EEG)**

This test records electrical activity in the brain. Some types of seizures produce characteristic EEG patterns but a normal recording does not rule out epilepsy. Not all abnormalities detected by an EEG are related to epilepsy. If

a characteristic seizure pattern is detected then further investigation may not be necessary.

N.B. It is important to note that some patients with unequivocal epilepsy will have persistently normal or non-epileptic EEG's.

### **Brain Scan (usually MRI or CT)**

This is useful to identify any structural abnormality of the brain.

A Magnetic Resonance Image (MRI) scan is more sensitive and specific than Computerised Tomography (CT) for detecting small brain lesions and abnormalities, which may be a relevant cause of epilepsy).

### **Blood tests**

These would be necessary to rule out any underlying pathology and to identify other possible causes of the seizure or event, though any metabolic disturbance is likely to have already been recognized. Blood levels of medications are also monitored.

### **Video-telemetry**

Video-telemetry combines continuous EEG and video recording and is valuable in the specialist assessment of difficult cases of episodes of disturbed consciousness.

It is possible for all of these tests to be negative and yet for the diagnosis of epilepsy to be established.

- [‘Status epilepticus’ & serial epilepsy](#)
- [Epileptic syndromes](#)
- [Other conditions & epilepsy](#)

### **How is it treated and managed?**

#### **Medication**

Epilepsy is eminently treatable with medication and the aim of treatment is to totally suppress seizures with the lowest possible dose of a single Anti-Epileptic Drug (AED). It is worth being aware that control of seizures is

affected by failure to comply with prescribed treatment and a surprisingly large number of individuals do not accurately follow prescribed treatment.

In general, the outlook is better than many people realise, with 80% being well controlled with no or few seizures. Complete suppression of seizures can be achieved in about 70% of those developing epilepsy.

The type of AED prescribed will depend upon the following factors:

- Type of epilepsy
- Age
- Drugs taken for other conditions
- Possible AED side-effects

Monitoring of blood levels may be appropriate with the use of some AED's, and in some situations, such as the suspected toxic effects of the medication or suspected non-compliance.

The following table lists the current recommended anti-epileptic drugs (AEDs) for different seizure types.

The individual should always be started on a single drug and if the epilepsy is more difficult to control, more drugs will be added.

<b>Seizure Type</b>	<b>First-line Drugs</b>	<b>Second-line Drugs</b>	<b>Other Drugs that may be considered</b>	<b>Drugs to be avoided (may worsen seizures)</b>
Generalised tonic-clonic	Carbamazepine Lamotrigine Sodium Valproate Topiramate	Clobazam Levetiracetam Oxcarbazepine	Acetazolamide Clonazepam Phenobarbital Phenytoin Primidone	Tiagabine Vigabatrin
Absence	Ethosuximide Lamotrigine Sodium Valproate	Clobazam Clonazepam Topiramate		Carbamazepine Gabapentin Oxcarbazepine Tiagabine Vigabatrin
Myoclonic	Sodium Valproate (Topiramate)	Clobazam Clonazepam Lamotrigine Levetiracetam Piracetam Topiramate		Carbamazepine Gabapentin Oxcarbazepine Tiagabine Vigabatrin
Tonic	Lamotrigine Sodium Valproate	Clobazam Clonazepam Levetiracetam Topiramate	Acetazolamide Phenobarbital Phenytoin Primidone	Carbamazepine Oxcarbazepine

Atonic	Lamotrigine Sodium Valproate	Clobazam Clonazepam Levetiracetam Topiramate	Acetazolamide Phenobarbital Primidone	Carbamazepine Oxcarbazepine Phenytoin
Focal with/without secondary generalisation	Carbamazepine Lamotrigine Oxcarbazepine Sodium Valproate Topiramate	Clobazam Gabapentin Levetiracetam Phenytoin Tiagabine	Acetazolamide Clonazepam Phenobarbital Primidone	

### Diazepam Use

Rectal diazepam may be used in the home management of prolonged or repetitive seizures. The clinical effect from rectal diazepam occurs rapidly, within a few minutes. It should be used with caution as it can cause, although rarely, respiratory depression (slower breathing or even brief cessation of breathing) especially in children. It is usually not necessary for children with mild or well-controlled forms of epilepsy. More recently however, buccal midazolam (the medication is placed against the sides of the gums and cheek) is being used instead of rectal diazepam. Buccal midazolam is easier to administer, with similar effectiveness and a better safety margin.

### Withdrawing medication

The reduction and withdrawal of medication is a complex decision usually negotiated over a period of time, between the epilepsy specialist and

patient. The risks include increased risk of seizures. This has implications for driving, and the use of machinery.

If an individual has been seizure free for 2 years, then consideration may be given to the withdrawal of medication in certain types of epilepsy.

Generally, medication is continued until at least a 2 to 3 year period free of seizures is established. Any drug withdrawal should be gradual, (i.e. over a period of 3 to 6 months).

Factors to take into account include:

- Fear and risks of long term unwanted effects of Anti-Epileptic Drug (AED) treatment

AED withdrawal is associated with an increased risk of seizure recurrence.

### **Other therapies**

#### Surgery

This is an option in a very small number of cases. It may be considered where a specific structural abnormality of the brain can be identified, such as an area of scar tissue, and this is in an area suitable for surgery, and where medication has proven unsuccessful. The need for surgery suggests likely poor control of the condition and therefore increased risk and the need for supervision or attention to avoid or reduce that risk.

#### Devices

A battery-powered device to stimulate the vagus nerve can be fitted under the skin like a pacemaker to send electrical signals to the brain via the vagus nerve in the lower back. This can help reduce seizure frequency. It is of most use where seizures are not well controlled by Anti-Epileptic Drugs (AEDs).

#### Diet

A "Ketogenic diet" which is rich in fats and low in carbohydrate can reduce seizure frequency in some children where AEDs have been ineffective. This therapy is mainly for children and young people. The need for a specialised

diet suggests likely poor control of the condition and therefore increased risk and the need for supervision or attention to avoid or reduce that risk.

### Lifestyle Issues

Getting adequate sleep, eating regularly, and the avoidance of stress and alcohol (in older children) are sensible lifestyle modifications. Compliance with treatment is also a very important factor.

- [Living with Epilepsy](#)

### What evidence is available?

Sources of evidence	Information Provided	Limitation of Evidence
Parent / Carer	First source of information and associated evidence e.g. basic information about medical management and appropriate Key Fact information.	Does not have specialist knowledge.
GPFR or Paediatrician / Paediatric Neurologist or Epileptic / Paediatric Nurse	Give a profile of the seizures: type, frequency, injuries sustained; day and / or night; episodes of status; warning; nature of post-ictal phase.  Also provide information on most up to date treatment and its effect and any further investigation or treatment (e.g. surgery) planned.	It is important to determine from the parent / carer who is the individual that is regularly managing their child's condition. This enables contact with the appropriate healthcare professional.

### Role of the specialist nurse

Those patients in tertiary care centres are likely to have access to a specialist epilepsy nurse (others are likely to be under a paediatric nurse). The role

of the specialist nurse in epilepsy management is diverse. Their role comprises mainly of:

- Improving seizure management: determining patterns of seizure activity, medication adjustment (in conjunction with the consultant) and treatment plans
- Education - general education of patients and families about epilepsy, promote compliance with medication, explain the potential for sudden unexplained death in epilepsy (SUDEP) and take measures to avoid or resolve misconceptions, agree emergency intervention plans and liaise with school (determine if support or training needed)
- Liaison with consultant – to communicate problems with seizure control and highlight to the consultant those children eligible for surgical consideration
- Other - to provide vital emotional support to patients and their families to cope with epilepsy

### How long will the needs last?

This guidance covers:

Generalised seizures (without status epilepticus in last 12 months)
Generalised seizures (with status epilepticus in last 12 months)
Partial seizures (without status epilepticus in last 12 months)
Partial seizures (with status epilepticus in last 12 months)
Seizures – unclassified
Non epileptic Attack disorder (pseudoseizures)

Age at date of claim	Onset less than 2 years	Onset more than 2 years
1 - 15	Award for 1 year	Award for 5 years

Note: In cases of epilepsy resulting from underlying brain damage / trauma or long-standing (i.e. of 5 or more years duration) poorly controlled epilepsy,



needs are unlikely to reduce despite medication and an award to age 16 is recommended.

Age at date of claim	Episode of Status Epilepticus
1 - 15	Award for 1 year

## Background

Epilepsy is a condition in which a person has recurrent seizures. Despite there being several different types of seizures, once treatment has been instituted for any type of seizure, the majority of children with epilepsy will improve significantly within 2 years. For those who continue with poorly controlled seizures, further treatment may also bring about seizure control, although this may be more difficult in this group.

A significant proportion of those claiming within the first 2 years of being diagnosed with epilepsy will improve after 1 year therefore a renewal at this stage is appropriate. The longer the duration of poorly controlled epilepsy, the less likely that it will be controlled. Therefore in claims from those who have had epilepsy for longer than 2 years, a renewal after 5 years is appropriate in the first instance, but after this, no further change is likely.

For many children with epilepsy the outlook is very positive. Factors, which may have some bearing, are the type of epilepsy they have and whether they have any additional health problems. Some children have severe forms of epilepsy with seizures that are difficult to control, such as in West's syndrome.

Giving a definitive prognosis after a single seizure is difficult, but some general rules do apply, based on epidemiological data.

Children who have a single, short, generalized seizure along with normal neurological development and normal findings on neurological examination have a 24% risk of having another seizure within 1 year and a 36% chance of having a second seizure within 3 years. The risks in those children with developmental problems or other neurological problems are significantly increased.

If a child has a second unprovoked seizure, the risk for further seizures is greater than 50%, even among children without other risk factors. Identifying the seizure as part of a syndrome has additional predictive value. For example, patients with benign epilepsy with Rolandic foci are likely to go

into remission; however, patients with juvenile myoclonic epilepsy are likely to have lifelong seizure recurrence.

### **Life expectancy**

Many children with epilepsy will have the same average life expectancy as children without epilepsy while others will have a shorter life expectancy. This can be due to an underlying cause of their epilepsy e.g. metabolic disorders, tumours etc. Additionally, life expectancy may be reduced due to

accidents occurring because of seizures e.g. having a seizure in the bath or near water.

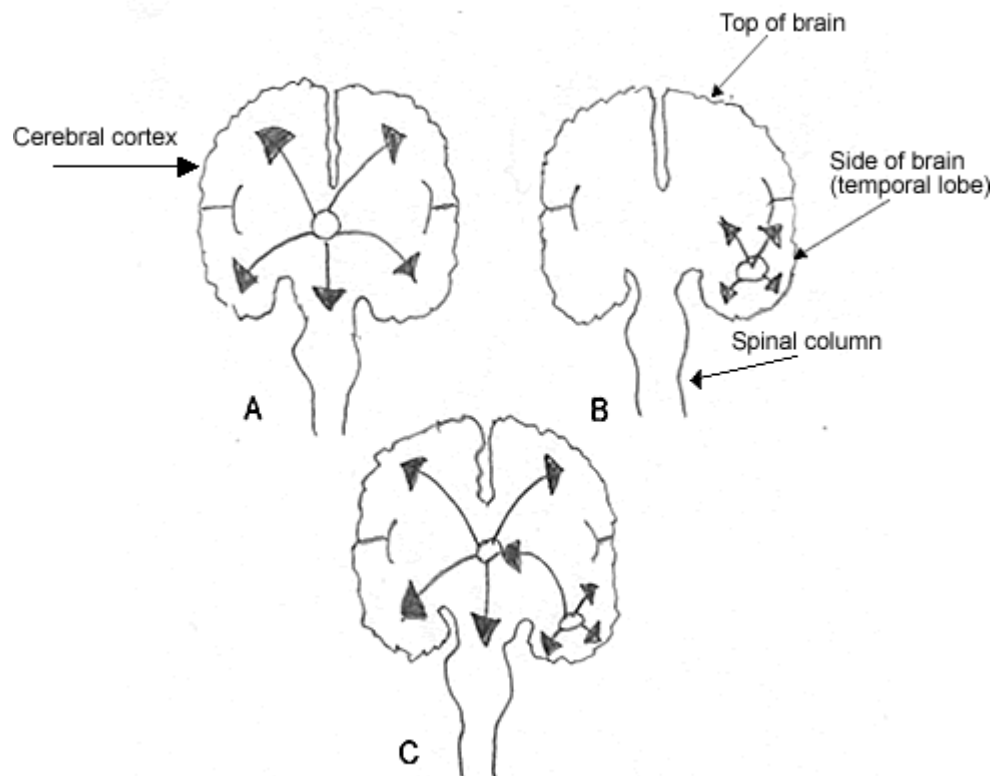
All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

## Classification of Epilepsy

### The ILE (1989) Classification of Epilepsy (different types of seizures)

The International Classification of seizures divides them into two major categories, partial and generalized seizures. However there are also several different types of seizures in each of these categories.

### Seizure Diagram



A. Primary Generalized Seizure

B. Partial Seizure

C. Partial Seizure with Secondary Generalization

Seizure Category	6 main seizure types
Generalised Seizures	<ul style="list-style-type: none"> <li>• Tonic-clonic (Grand mal) primary or secondary types</li> <li>• Absence (Petit mal)</li> <li>• Myoclonic</li> <li>• Tonic</li> <li>• Clonic</li> <li>• Atonic</li> </ul>
	<b>3 main seizure types</b>
Partial Seizures	<ul style="list-style-type: none"> <li>• Simple partial</li> <li>• Complex partial</li> <li>• Complex partial evolving to generalised tonic-clonic</li> </ul>

### Generalised Seizures

This describes an event where the abnormal electrical activity affects all or most of the brain from the outset. Consequently, the symptoms tend to be general and involve most of the body.

There are 6 types of generalized seizures:

- Tonic-clonic (previously called grand mal or generalized convulsion). These may be either primary or secondary in nature.
- Absence seizures (previously called petit mal)
- Myoclonic seizures or Juvenile myoclonic seizures
- Tonic seizures
- Clonic seizures
- Atonic seizures

### Tonic-Clonic seizures

This type of seizure is the commonest seizure event accounting for about 60% of all types. The seizure event may be preceded by a prodromal period, (period of precursor symptoms) which may manifest as a change in mood or behaviour and during which, the patient feels different, lasting hours or days. This is not part of the seizure itself.

They may or may not then experience an aura or vague warning, which is itself, part of the seizure and precedes the other manifestations. It may be a strange feeling in the stomach or a strange smell and can take many forms. In such cases the individual would normally be able to take steps to avoid danger.

However, the description of an aura is not commonly obtained in tonic-clonic seizures. This may be due to memory loss (retrograde amnesia) for events immediately preceding the seizure. Following the aura, if present, the child then goes rigid due to intense sustained muscular contractions and becomes unconscious. This is the tonic phase.

During this phase air is forced out of the lungs causing the individual to cry out or make a strange noise. The bladder also contracts and urinary and faecal incontinence may occur. The jaw muscles contract and the teeth clench together often with biting of the inside of the lips and the tongue. If the child is standing they may fall heavily to the ground and may sustain serious injury.

During the tonic phase breathing ceases and the child may begin to turn a blue colour (central cyanosis).

After a few moments the rigidity is periodically relaxed and the clonic phase begins. This phase is characterized by rhythmic jerking of the muscles causing the whole body to shake (convulse) uncontrollably. Frothing at the mouth often occurs. Breathing returns in the clonic phase but is erratic. This phase may last for a few seconds to several minutes. On average the entire event will last about 3-5 minutes.

The child will then gradually regain consciousness but is often dazed and confused (the post epileptic or post-ictal state). The child gradually regains consciousness but will be confused and disorientated for at least half an hour after regaining consciousness and may not recover full memory function for some time (hours).

They may also complain of a headache and feel terrible with the desire to lie down and sleep. In some cases the child may remain unresponsive following the seizure. In other cases they may become aggressive with

disturbed behaviour. This post-ictal state may persist for several hours, or up to days following the seizure.

Afterwards, muscles are often sore and aching due to the intense contractions and the tongue, which has often been severely bitten, may be sore.

Tonic-clonic seizures may be subdivided into 2 main types:

- **Primary Generalised Epilepsy** - This type of epilepsy invariably begins in childhood or adolescence. There is no structural abnormality of the brain to be found on investigation to account for the seizures and a genetic predisposition is likely. Prognosis is generally good with treatment.
- **Secondary Generalised Epilepsy** - This may arise from a partial seizure, which spreads out of its localized area to involve the entire brain, or it may be secondary to other causes such as drugs, metabolic disorders, head injury, etc. Such seizures are more difficult to control.

### **Absence Seizures**

Absence seizures are another type of generalised epilepsy that invariably begins in childhood. There is no convulsion with this form of seizure. Activity ceases, and there is a brief (seconds only) loss of awareness. The child may stare and there may be some twitching of the eyelids. A few muscle jerks may occur. The child does not fall over. The attack is normally over in a few seconds, when normal activity is resumed. There is no post-ictal state.

Although attacks are brief, they can be frequent and may occur hundreds of times a day in young children interfering with behaviour and school performance.

### **Myoclonic Seizures**

This is another type of generalized seizure. It is also known as juvenile myoclonic epilepsy. Myoclonic seizures describe isolated muscle jerking. This type of epilepsy begins in adolescence and is characterised by sudden muscular contractions usually on awakening. These normally affect one or both arms. There may be a brief period of loss of consciousness.

Patients with myoclonic seizures can get worse if put on the wrong medication, (such as carbamazepine). The prognosis is excellent with this form of epilepsy, but over 95% of cases will relapse if medication is stopped. A related syndrome is that of tonic-clonic seizures on awakening.

### **Tonic Seizures**

Tonic seizures cause a brief loss of consciousness. The body becomes rigid and the person falls to the ground. There is no subsequent convulsion or

clonic phase and the child regains consciousness. Injury may be sustained during the fall.

### **Clonic Seizures**

Clonic seizures cause loss of consciousness with rhythmic muscular jerking. There is no preceding stiffness or rigidity.

### **Atonic Seizures**

Atonic seizures cause a brief loss of consciousness with sudden loss of muscle tone so that the child falls to the ground.

### **Partial Seizures**

This describes a localised or focal seizure, which starts in and normally stays in one part of the brain. There are 3 types of partial seizures -:

- Simple partial
- Complex partial
- Complex partial evolving to generalised tonic-clonic seizures

#### Simple partial seizure

(with no loss of consciousness previously called Jacksonian seizures).

In this type of seizure the abnormal electrical impulse originates in the motor cortex of the brain, which controls muscular movements.

Typically, jerking movements begin at the angle of the mouth, or in the thumb and index finger. The jerking may spread gradually to involve the whole limb on the affected side. This spread is called the march of the seizure. Attacks may last from a few seconds to several hours.

If the seizure is prolonged and affects the entire limb then weakness or paralysis of the affected limb (s) (Todd's Paralysis) may be present for several hours after the seizure ceases.

There is no loss or impairment of consciousness with simple partial seizures, and the child is generally aware throughout the entire event.

Complex partial seizure (with variable impairment or loss of consciousness, previously often called Temporal lobe Epilepsy).

In Complex partial seizures, the abnormal electrical impulse commonly originates in the temporal lobe of the brain, but may start in any part.

Depending on the part of the brain affected the child may behave in a strange manner for a few seconds or minutes. They may fiddle with an object or pluck at their clothing, mumble incoherently or exhibit automatisms

such as blinking, twitching, mouth movements or walking in a circle. (These are non-convulsive attacks and sometimes it may not be obvious to the medical staff or observer that this person is fitting).

Individuals may also continue activities started before the seizure began, such as washing dishes, but these activities are continued in a repetitive un-productive manner.

Such seizures may last from a few seconds to several minutes. Consciousness is normally impaired or lost, and there may be no recollection of the event.

The child may feel dazed and muddled for a short while following the attack but does not normally fall or collapse to the ground.

Some patients with complex partial seizures experience an aura – unusual sensations that warn of an impending attack. Such sensations may on occasion mimic psychiatric conditions. Déjà-vu, which is a subjective feeling that an experience which is occurring for the first time has been experienced before, may occur.

Complex partial seizure (evolving to generalised tonic-clonic seizures).

In a small proportion of individuals, the partial seizure spreads out of its localised area to involve the rest of the brain. A generalised convulsion (tonic-clonic) then occurs with complete loss of consciousness. This is termed Secondary Generalised Seizure (figure C of the seizure diagram on the Classification of Epilepsy page refers).

### **Non-epileptic seizures**

Any seizure is a cause of concern, but having a seizure by itself does not mean that a child has epilepsy. The following are examples of seizures that may not be associated with epilepsy.

- First seizures
- Febrile seizures (febrile convulsions)

Febrile seizures are convulsions brought on by a fever in infants or small children. During a febrile seizure, a child often loses consciousness and shakes, moving limbs on both sides of the body. Less commonly, the child becomes rigid or has twitches in only a portion of the body, such as an arm or a leg, or on the right or the left side only. Most febrile seizures last a



minute or two; some can be as brief as a few seconds whilst others last for more than 15 minutes.

Children prone to febrile seizures are not considered to have epilepsy, since epilepsy is characterized by recurrent seizures that are not triggered by fever.

- Non-epileptic events (Non-Epileptic Attack Disorder)

Sometimes young children and adolescents appear to have seizures, even though their brains show no seizure activity. This type of phenomenon has various names, including Non-Epileptic Attack Disorder [NEAD], psychogenic seizures, and pseudo-epileptic seizures. These terms essentially mean something that looks like a seizure but is not.

The difference between epileptic seizures and NEAD is that epileptic seizures are accompanied by a change in brain activity, whereas NEAD is not.

NEAD may indicate dependence, a need for attention, avoidance of stressful situations, or specific psychiatric conditions, including personality disorders. Some young children and adolescents with epilepsy have NEAD in addition to their epileptic seizures. Others who have NEAD do not have epilepsy at all. NEAD cannot be treated in the same way as epileptic seizures. In fact, anti-epileptic treatment will not have an effect on NEAD. Instead, they are often treated by mental health specialists and with other medication but not Anti-Epileptic Drugs (AED's).

Other non-epileptic events may be caused by narcolepsy (excessive daytime sleepiness, involuntary daytime sleep episodes and sudden loss of muscle tone), Tourette's Syndrome (a neurological disease of unknown cause that presents with multiple tics [uncontrolled behaviour], snorting, sniffing, involuntary vocalisations and explosive utterance of obscenities), cardiac arrhythmia (abnormalities of heart rate or rhythm), and other medical conditions with symptoms that resemble seizures. Because symptoms of these disorders can look very much like epileptic seizures, they are often mistaken for epilepsy. There may be alteration in behaviour, some movement of limbs, and loss of consciousness. Distinguishing between true epileptic seizures and non-epileptic events can be very difficult and requires a good history of the seizures, and eyewitness accounts, thorough medical assessment, careful monitoring, and knowledgeable health professionals.

Video-telemetry and EEG enable a diagnosis of non- epileptic events to be made.

- Eclamptic seizures (seizures which only occur in pregnancy, or soon after delivery).
- Fainting and syncope. A faint is distinguishable from a seizure in not having the following features: aura, tongue- biting, cyanosis, subsequent confusion, amnesia, or headache.

## **Status epilepticus and serial epilepsy**

### **Status epilepticus**

Status epilepticus occurs most commonly in tonic-clonic seizures and is a serious form of epilepsy in which there is persistent epileptic activity for more than 30 minutes, or in which there are successive seizures without recovery of consciousness in between.

‘Status epilepticus’ may be divided into two groups:

- Those where the ‘status’ was precipitated by an avoidable cause e.g. sudden withdrawal of anticonvulsant medication or alcohol withdrawal in some adolescents. This may be termed as ‘explained status epilepticus’
- Those where there is no identifiable cause, i.e. the child may have poorly controlled epilepsy. This may be termed as ‘unexplained status epilepticus’

‘Status epilepticus’ is the most serious event children with epilepsy are likely to encounter and has a significant mortality rate. The risk of recurrence of status epilepticus will depend on whether there are any avoidable precipitating factors e.g. sudden withdrawal of alcohol or medication (explained) or not (unexplained).

If no cause is found, then recurrence cannot be excluded. The risk of recurrence decreases significantly with the passage of time and is minimal after 12 months from the date of the last episode.

### **Serial Epilepsy**

Serial epileptic attacks (also known as “closely spaced seizures”, or “cluster attacks”) should be distinguished from ‘Status epilepticus’.

In serial epilepsy although attacks are very frequent, the person recovers consciousness in between attacks. It is not associated with the same risks

as 'Status epilepticus', in which the attacks occur in such rapid succession that recovery of consciousness between the episodes does not occur.

## **Epileptic syndromes**

An epileptic syndrome is a group of signs and symptoms that share a common pathogenesis, prognosis and response to treatment.

The classification of epileptic syndromes comprises 2 major categories:

- localization-related syndromes
- generalized-onset syndromes

Clinicians ideally classify their patients' seizures by using the classification for seizure types and make a syndrome diagnosis if possible.

Clinical features and prognosis depend a lot on the specific epileptic syndrome but it is not always possible to make a syndrome diagnosis. More often, there is only a description of the type of seizures.

## **Examples of Epileptic Syndromes in children**

Benign epilepsy with Rolandic foci

Onset is between 2 and 12 years and is maximal between 7 and 10 years and stops by 13 years. Attacks are clonic, partially sensori-motor affecting the face, tongue and pharyngeal (of the pharynx) muscles, hand and arm and they occur particularly on waking.

Juvenile myoclonic epilepsy

Juvenile myoclonic epilepsy (JME) occurs in the teen years. In JME, individuals have generalized motor seizures, myoclonic jerks (suddenly drop things) and staring spells that occur upon awakening.

Infantile spasms (West's syndrome)

Infantile spasms, or West's syndrome, consist of 'runs' of tonic spasms occurring every 5-10 seconds. Attacks start in the first year with a peak onset at around 4 months, the outcome being worse with early onset. The commonest spasms are flexion (the bending of limbs). Parents may notice loss

of visual and social interaction rather than spasms. West's syndrome is symptomatic of many underlying brain pathologies.

A grossly disorganised epileptic electroencephalogram (hypsarrhythmia) is characteristic but not always seen. The mainstays of treatment are corticosteroids and vigabatrin.

A death rate of about 20% and the occurrence of cerebral palsy in 30-50% and cognitive disability in up to 85% should prompt early determination and energetic treatment.

### Lennox-Gastaut syndrome

In the Lennox-Gastaut syndrome, multiple seizures occur in the first eight years of life. The seizures include tonic attacks (particularly in sleep), atypical (not conforming to the usual type) absence seizures, atonic and myoclonic attacks and episodes of non-convulsive 'Status epilepticus' with long periods of impairment of motor and cognitive function. Half of the children affected have primary developmental delay and all have learning problems after five or more years of the condition. Only up to 10% have a reasonable outcome.

Seizures are often resistant to anti-epileptic drugs, but benzodiazepines, sodium valproate, lamotrigine, corticosteroids and a ketogenic diet (containing minimal amounts of protein and carbohydrate) are regularly used. Occasionally, surgical treatment can be offered for instance to reduce severe drop attacks (that cause sudden falls and head injury).

### Sudden unexplained death in epilepsy (SUDEP)

This is a mysterious, rare condition in which individuals with epilepsy die without a clearly defined cause. By definition, (1) death is sudden and unexpected, (2) a clear cause of death is absent, and (3) the individual had epilepsy. They are otherwise in a reasonable state of health at the time of death. Although seizures are suspected to have occurred prior to death, there is no evidence of seizure as the direct cause of death.

Recent studies have suggested that a combination of impaired breathing (apnoea), increased fluid in the lungs (impairing the exchange of oxygen and carbon dioxide) and being face down on the bed all combine to cause death due to impaired respiration.

The risk of SUDEP for someone with epilepsy is about 1 in 3000 per year. This risk is multiplied in those who have frequent seizures and take large doses of many anti-epileptic drugs (AEDs). A major risk factor for SUDEP

appears to relate to the severity of the epilepsy, as SUDEP is more common in those with:

- frequent convulsive seizures
- early age of onset of epilepsy
- long duration of epilepsy
- higher number of anti-epileptic medications and at high doses
- frequent medication changes

Steps to minimise the chances of SUDEP

Ensure compliance with medication

Supervision of individuals with a high likelihood of tonic-clonic seizures in sleep

Basic first aid should be provided during a seizure, including rolling the individual onto one side, checking respiration and avoiding putting any object in their mouth.

A family member or carer of children with uncontrolled convulsive seizures should learn cardiopulmonary resuscitation.

### **Other medical conditions with epilepsy as a significant feature**

#### **Landau-Kleffner syndrome**

This syndrome typically begins with partial or generalised seizures (including atypical absence seizures) and language comprehension and speech are lost after two or more years of normal development. The range of impairments includes behavioural disorders and global cognitive, motor and social regression with many autistic features. Anti-epileptic drugs (AEDs) often have no effect on the cognitive deficit but high doses of sodium valproate, often in combination with lamotrigine, sometimes help.

#### **[Tuberous sclerosis](#) – NHS Choices**

Approximately 80% of children with tuberous sclerosis (TS) have some form of epileptic seizures and mental retardation is seen in 60%. The epileptic disorder is often severe and resistant to the treatments available, with a very low remission rate. Various epileptic disorders occur in children with TS, but infantile spasms are the most common presentation. As these

children age, the epileptic disorder may change; in some children, Lennox-Gastaut syndrome emerges.

Surgery with removal of the epileptogenic tuber should be considered if there is poor response to treatment. The surgical outcomes vary but there is marked improvement in a substantial number of children.

### Autism

Approximately 20-30% of children and adolescents with autism develop some form of epileptic disorder. The seizures are observed more frequently in patients with more severe cognitive impairment.

### Rett syndrome – Medterms

Rett syndrome is a major cause of severe mental retardation associated with seizures in girls. It is characterized by a progressive mental and growth impairment that starts in infancy.

They develop an autism-like syndrome with stereotyped movements, some of which, especially those in the hands, are considered very typical of the disease. Epileptic seizures are seen in 25-30% of cases, mostly generalized and complex partial.

### Cerebral palsy

Epileptic seizures occur in 25-50% of children with cerebral palsy (CP). The incidence is related to the severity of the brain damage. It is higher in those children with quadriplegia, lower in those with congenital hemiplegia and much lower in children with diplegia and the dyskinetic form of CP.

The first epileptic seizures typically are seen during infancy. The seizure disorder is the consequence of the brain abnormalities associated with the CP. Clinical studies indicate that early seizures are associated with more cognitive deficiencies.

Practically every type of epileptic seizure has been described in individuals with CP. Generalized tonic and tonic-clonic seizures and partial complex seizures are most frequent. Some syndromes, such as infantile spasms and Lennox-Gastaut syndrome are particularly frequent in children with CP.

### **Living with epilepsy**

Most children with epilepsy attend mainstream schools. Many children with epilepsy have no other major condition and the seizures are well controlled.

Some children with epilepsy have other major conditions (such as learning difficulties) and may require special schooling. Depression is more common

in adolescence when the young person may become very frustrated, have low self-esteem and have fears of losing control.

Direct supervision may be required to manage safety issues indoors/outdoors.

Children with poorly controlled seizures should not engage in activities that are potentially harmful. They should not be allowed to take unsupervised baths (because of the risk of drowning). Supervised swimming, bike riding (helmeted) and playing video games are considered by most neurologists to be safe activities.

#### Aids / adaptations

Non-slip rugs and high balcony railings are two simple adaptations. A bed alarm can detect convulsive seizures occurring during sleep, some models also monitor the heart rate and breathing patterns. A pressure mat alerts when a person lands on the mat e.g. falling out of bed or wandering in a seizure. Special breathable pillows are available to minimise the risk of suffocation. In the bathroom, a walk-in shower with a non-slip floor or mat and a shower seat may minimise the risk of injury.

These types of measures indicate that the family is taking additional steps when the child's seizure control is poor and hence at increased risk.

#### Nocturnal (night-time) seizures

Some seizures occur at night and for some children, seizures occur only at night.

Once a child is in bed, they are not at risk of falling and injuring themselves. The danger of choking or being suffocated by a pillow is extremely small and special pillows are available on the market to reduce this possibility to a minimum. Alternatively, the individual may choose not to use a pillow.

Parents may choose to use a mattress alarm that detects when a child has a convulsive seizure and triggers the alarm. Note - these aids may be used by parents / carers to create a safer environment for their child and so are not reliable indicators of overall severity of functional restriction. There are also other adaptations which could be made to the room such as the use of a low bed.

With some types of epilepsy, confused or automatic behaviour may lead to the danger of wandering or other behaviour, which may lead to harm. In

these circumstances, it would be sensible for appropriate precautions to be taken, such as having another person in the house and minimizing hazards.

It should be sufficient that someone is present in the house with the child, as there would not usually be a need for another person to remain awake observing the child in case they had a fit.

### Faltering growth

Faltering growth (previously called failure to thrive) is failure to maintain a normal rate of growth compared to other children. It is normally used in reference to babies and young children.

Faltering growth is best described as failure to advance along a centile on a growth chart. Thus, a child whose weight is on the 3rd centile, and always has been, is growing normally. A child who is now on the 3rd centile, after having been on the 50<sup>th</sup> centile, has faltering growth.

In 95% of cases there is no underlying disease causing the failure to thrive.

Current evidence suggests that weight faltering in infancy does have an effect on long term growth and may have a small effect on cognition.

### Award instructions

Where faltering growth is due to a diagnosed, underlying disease then coding and award duration should be based on the underlying disease.

If the sole problem is faltering growth see below.

Impairment	Award Period
Faltering growth	Seek medical advice on prognosis and award duration if needs claimed.

### Gastro-oesophageal reflux disease (GORD)

#### What is GORD?

Gastro means stomach and oesophagus means gullet. Gastro oesophageal reflux is the backflow of stomach contents into the gullet.

Everyone experiences some reflux some of the time. This is especially the case following large quantities of food (especially 'rich' or spicy food) and when lying horizontal. All babies experience reflux because they drink around 1/6 of their body weight in milk each day and are frequently



horizontal. For most babies this manifests as regurgitation or possetting of milk into the mouth.

GORD is the diagnostic term used when reflux causes symptoms severe enough to merit medical treatment. Medically, judging whether reflux is 'normal' or is the cause of symptoms can be difficult.

There is a wide spectrum of disease in GORD with different children being affected in different patterns and to different degrees. A child diagnosed with GORD might display any or several of the following symptoms:

- regurgitation or persistent vomiting - generally possetting
- faltering growth
- back arching/irritability/persistent crying
- feeding refusal or established oral aversion
- symptoms of oesophageal acid damage including haematemesis (vomiting of blood), dysphagia (inability to swallow) or odynophagia (painful swallowing)
- anaemia due to oesophageal blood loss
- symptoms from respiratory acid overflow including wheezing, stridor, cough, hoarseness, laryngospasm (spasm of the vocal cords) and apnoea (temporary cessation of breathing)
- stomach content aspiration (food being breathed into the respiratory tree)
- dental erosion
- sleeping difficulties

#### Impact on daily living

Infants (under 1 year old): The vast majority of babies with reflux or GORD requiring treatment do not require significantly more care than another child of their age (though they can be exhausting to care for). However, there is a small group with severe disease where either, faltering growth, established oral aversion or respiratory complications like apnoea or aspiration require substantial additional care. This may include hospital admissions, specialist

feeding groups and specialist devices such as naso-gastric (nose to stomach) tubes, monitoring or even surgery.

**Children with disabilities:** A substantial number of children with neurodevelopmental disability such as cerebral palsy or neuromuscular disease also experience GORD. This can be the result of neurological swallowing difficulties and spinal deformities, which interfere with the normal function of the oesophagus. In these children, the additional burden arising from feeding problems, feed-related pain or hospitalisations due to respiratory complications such as aspiration can have a major bearing on the level of care required.

**School-aged children without disability:** It is unusual for this group to require substantial medical input due to GORD, but it is being increasingly recognised as a contributory factor in asthma and other respiratory problems so may be mentioned on a claim relating to respiratory problems. In this circumstance, the impact on daily living is a direct result of the severity of the respiratory problem.

#### Treatment and management

Typical treatments are listed below in an escalating fashion to help judge how much treatment the health professionals involved feel is justified or what level of treatment has, thus far, been reached.

#### Positioning

- primarily used for infants and disabled children who are dependent on a carer for their posture; strategies include staying upright after a feed, raising the head of a bed or cot, avoiding seating positions that compress the abdomen and lying prone (face down) or on the side when supervised or awake.

#### Dietary measures

- eliminating exacerbating dietary components such as rich or spicy food or dairy products in children with allergy or intolerance
- frequent small feeds or meals
- thickened feeds, typically powder thickeners (such as Gaviscon or Carobel), specialised self-thickening milks or the addition of rice or corn

#### Pharmacological

- antacids (for example, Gaviscon). These work by neutralising the acid in the stomach contents, which both reduces acid-promoted reflux and reduces the acidic consequences of reflux. When added to milk feeds, most are also thickeners

- H2 antagonists (for example, Ranitidine) which reduce gastric acid production
- proton pump inhibitors (PPIs) for example, Omeprazole, which also reduces gastric acid production
- prokinetic agents (for example, Domperidone and the antibiotic Erythromycin) which speed up gastric emptying (following further recent evidence from the medicines safety regulator these are now used very rarely)

### Surgery

Fundoplication is major surgery to strengthen the muscular ring (sphincter) that prevents reflux and is usually only considered where all other measures have been implemented but symptoms are still serious.

How long will the needs last?

Usually, this condition resolves spontaneously during the first year of life. In a small proportion of individuals with this condition there may be the development of ulceration in the lower end of the oesophagus due to repeated reflux of the acid stomach liquid. This may cause repeated episodes of

haematemesis (vomiting of blood) and rarely leads to the development of an oesophageal stricture (permanent critical narrowing).

For the most part this is a self-limiting condition that responds well to medication and usually improves as a child grows.

If the condition and the evidence is consistent with an award, then it is reasonable to award for a duration of 2 years in the first instance. Further awards should be reviewed based on subsequent evidence submitted.

## Gender and gender dysphoria

This is a short note on gender rather than full guidance

### Terminology

Gender dysphoria - the distress felt by those whose gender identity and gender assigned at birth are not the same

Transgender (or trans) person – an individual whose identified gender is not the same as the gender they were assigned at birth

Cisgender – an individual who identifies with the gender assigned to them at birth

Non-binary – an individual who does not identify with conventional 'maleness' or 'femaleness.' A non-binary person may have no gender, 2 or more genders or move between genders

Trans woman – an individual who identifies as female having been assigned the male gender at birth

Trans man – an individual who identifies as male having been assigned the female gender at birth

Transitioning – the process an individual takes to live as the gender they identify with – this may be social (changing name, changing pronouns, dressing as the preferred gender), medical, surgical, or any combination of these. Some individuals may transition only in some environments (eg at home but not at work) or only to some people (eg to a particular group of friends but not family)

Personal pronouns (she/he/they etc)– different individuals have different preferences. Some individuals may wish to use binary pronouns related to the gender they identify with, whilst others prefer to adopt non-binary pronouns (they and them). There is no 'right', it is important to use what the individual wants, and respect their reasons for doing so. Both cisgender and transgender individuals may choose to use non-binary pronouns.

People experiencing a mismatch between their gender assigned at birth and their gender identity may be anxious and depressed and may self-harm.

There is guidance on these topics which should be followed. They commonly experience bullying from others which has its own mental health problems. Some individuals also express their distress at this mismatch through externalised behaviours such as aggression towards others. An individual may also experience bodily-dissatisfaction and they may have additional care needs around washing due to the mismatch between their body

and their gender identity meaning they find it difficult to look at or touch their body.

## What you need to know about Haemophilia

### What is haemophilia?

Haemophilia is a bleeding disorder in which the blood fails to clot normally .....

- [Haemophilia](#)

### What are the effects and signs?

Children with haemophilia experience bleeding into the tissues of the body after a trivial injury or sometimes spontaneously. ....

- [Effects of haemophilia](#)

### How is it assessed?

Plasma levels of individual clotting factors can be measured with simple blood tests.....

- [Assessment of haemophilia](#)

### How is it treated and managed?

There is no cure for haemophilia and the need for monitoring and treatment is lifelong.....

- [Treatment of haemophilia](#)

### What evidence is available?

Children with haemophilia are likely to receive all their haemophilia care through the Haemophilia Comprehensive Care Centre.....

- [Evidence](#)

### How long will the needs last?

- [Prognosis and duration of the award](#)

## What is Haemophilia?

Haemophilia is a genetic bleeding disorder in which the blood fails to clot normally. Blood clotting is the process by which protein factors and platelets activate one another to form a plug at a site of bleeding such as a torn

blood vessel to prevent further loss of blood from the site. Over time the initial plug forms into a scab and healing of the torn vessel occurs. Clots are formed by the activation in the blood of special proteins called clotting factors, these factors activate one another in a sequential manner or 'cascade' that results in a blood clot over a few minutes, and this stops bleeding. When a clotting factor is missing, only present in low amounts or in an ineffective form the cascade cannot complete. An ineffective clot is formed so bleeding continues.

This bleeding most frequently occurs internally.

### **Types of haemophilia**

There are two types -:

- Haemophilia A – this is the most common type of haemophilia. In this condition clotting factor VIII is abnormally low or absent.
- Haemophilia B – this is the least common type of haemophilia. In this condition clotting factor IX is abnormally low or absent. Also called Christmas disease.

These conditions are both sex-linked recessive conditions. This means that the abnormal gene is carried on the X chromosome and the condition affects boys. The X chromosome is a long chromosome that pairs with another X chromosome in women (XX) and with a very short Y chromosome in men (XY). The Y chromosome does not carry the genes for clotting factors. Women have two copies of the X chromosome carrying genes for clotting factors VIII and IX. In women with the haemophilia gene on one X chromosome the unaffected X chromosome ensures enough clotting factor is produced. In boys with the affected X chromosome there is no second X chromosome to ensure clotting factor production and they have haemophilia. Boys with the affected chromosome will be affected by haemophilia, girls with the affected X chromosome will not have haemophilia but they will be

carriers and their male children have a 50% chance of being affected by haemophilia.

### **What is the incidence/prevalence?**

The prevalence of haemophilia in the UK is:

- Haemophilia A: 1 in 5000 to 1 in 10 000 males
- Haemophilia B: 1 in 35 000 to 1 in 50 000 males

Around 9500 boys and men in the UK have haemophilia.

Bleeding disorders caused by blood clotting factor deficiencies

- [Cause of disability](#)
- [Bleeding disorders caused by blood clotting factor deficiencies](#)

### **What are the effects and signs?**

#### **Haemophilia A and B**

Children with haemophilia experience bleeding into the tissues of the body after a trivial injury or sometimes spontaneously. The bleeding may occur into joints, muscles, abdomen or the brain. The probability of bleeding and the risk of a severe bleed depend on how little normal clotting factor is produced. This is measured in international units per decilitre (iu/dl) with a normal range of 50-150 iu/dl (for children under 6 months old the normal ranges for Factor IX are lower). A person with haemophilia will produce less than this, the lower the value the higher the risk of bleeding. This can be used to categorise haemophilia into severe (<1iu/dl), moderate (1-5iu/dl) and mild (5-50iu/dl). Sometimes these values are expressed as a percentage of normal clotting factor so severe (<1iu/dl) may be expressed as less than 1% of normal clotting factor levels.

Moderate haemophilia (1-5iu/dl) may be expressed as 1-5% of normal clotting factor levels. Mild haemophilia (5-50iu/dl) may be expressed as 5-25% of normal clotting factor levels. These categories refer to the amount of clotting factor produced and not the disability experienced. Disability will depend on how many severe bleeds there have been and whether these have resulted in long term damage.

#### **Inhibitors**

Inhibitors can sometimes be a troublesome complication in the treatment of haemophilia. They can range from being mild to severe. They occur when



the immune systems of people being treated with blood clotting agents, such as nonacog alfan, start to regard the clotting agents as foreign objects.

The immune system will start to create antibodies to block the effects of the clotting agent. These antibodies are known as inhibitors. These inhibitors can make the medication that is used to treat haemophilia less effective, which means that it is more difficult to prevent and control the symptoms of bleeding.

An estimated 15-25% of people who receive treatment for haemophilia A develop inhibitors.

An estimated 1.5-3% of people who receive treatment for haemophilia B develop inhibitors.

Severe haemophilia means less than <1iu/dl of Factor VIII or Factor IX are found in the blood (less than 1% of normal clotting factor levels)

Boys with severe haemophilia experience serious bleeding episodes. The condition may present soon after birth for example with bleeding into the scalp of a newly delivered infant. Bleeding occurs after mild or trivial injury and sometimes spontaneously. Bleeding into joints and muscles is a common problem. The joints most commonly affected are the knee, elbow, ankles, shoulder, wrist and hip. Repeated bleeds into joints leads to long term damage in the form of severe arthritis and sometimes deformity and reduced range of joint movement. The joint damage rather than the bleeding episodes themselves are the cause of disability. Since the late 1990s prophylactic (preventative) treatment has been introduced to prevent such bleeds and boys with severe haemophilia born since then may escape serious joint damage. Anyone born prior to the 1990s (or outside the UK in a place where treatment is not available) with severe haemophilia is likely to have some joint damage. Many will have multiple affected joints and enduring disability related to this, despite now using prophylactic treatment to prevent further bleeds and joint damage.

Bleeding into tissue causes haematoma formation, a haematoma is a collection of blood. Haematomas can damage surrounding tissue through pressure effects; the brain is especially at risk as it is housed in a non-expandable space inside the skull. Haemorrhage into the brain may be life threatening and can cause long term disability, an intracranial bleed may cause the following enduring effects:

- cerebral palsy
- neurological deficits
- epilepsy

Haematomas elsewhere in the body can cause muscle damage or peripheral nerve damage. Bleeding into large spaces such as the abdomen can

result in extensive blood loss from the circulation, such bleeds are life threatening.

Frequency of bleeding episodes varies between individuals. Without prophylactic treatment some boys experience bleeds several times a week in others they may occur only a few times a year.

Moderate haemophilia - Factor VIII or Factor IX 1-5iu/dl (1-5% of normal clotting factor levels)

Bleeding tends to occur after trauma or injury, which can be minor. Spontaneous bleeding does not usually occur. If not treated episodes of bleeding may cause joint or neurological damage as described above in severe haemophilia.

Mild haemophilia – Factor VIII or Factor IX 5-50iu/dl (5-25% of normal clotting factor levels)

Abnormal bleeding tends only to reveal itself after significant injury, surgery or tooth extraction. Treatment of clotting factor deficiency is only required at these times. Life is normal otherwise and joint related or neurological disability from bleeds does not occur.

### **Female haemophilia carriers**

Abnormal bleeding may occur after surgery or tooth extraction as some carriers will have low levels of clotting factor. The levels are very variable and may be under 20iu/dl. The carrier state is unlikely to be recognised unless there is a bleeding event that prompts investigation or a family history leading to genetic testing. Life is normal and there is no associated disability.

### [Care and mobility considerations](#)

#### **How is it assessed?**

- Plasma levels of individual clotting factors can be measured with simple blood tests.
- The haemophilia carrier state can be identified by genetic testing in women and prenatal diagnosis is possible in families

who know they have a gene mutation. Knowing a child has haemophilia before birth can prevent birth trauma.

- Inhibitors are screened for regularly.

The presence of an inhibitor is abnormal.

The amount of inhibitor is measured in Bethesda units.

### **How is it treated and managed?**

There is no cure for haemophilia and the need for monitoring and treatment is lifelong.

The aim of treatment in haemophilia is to control or prevent abnormal internal bleeding so preventing the joint and neurological impairments and disability associated with the condition. Boys with haemophilia born from the late 1990's onwards are most likely to have benefited from modern treatment regimes and very few will have any significant impairment related to bleeding complications. The treatment of clotting disorders is replacement of missing clotting factors in the blood to restore normal clotting and prevent, reduce or minimise internal bleeding.

Children with a bleeding disorder of any severity should have access to a Haemophilia Comprehensive Care Centre (CCC). Any boy who requires regular treatment or who has joint damage is likely to receive all of their

haemophilia care via the CCC. There are various aspects to care required and these are co-ordinated through the centres. Examples of care include:

- Management of bleeding tendency and clotting factor replacement
- Management of complications of bleeds
- Specialist physiotherapy assessment and treatment
- Psychological and social care
- Genetic counselling

The aims of management are:

- control bleeding tendency
- treat bleeding episodes
- prevent joint damage and neurological complications
- manage joint damage
- enable normal growth and development during childhood and support normal life in adulthood

### **Clotting Factor replacement therapy**

The cornerstone of treatment is the replacement of clotting factors to enable normal blood clotting and to prevent complications of bleeds. If frequent treatment is required it is likely to be administered at home with help, support and guidance from the centre. The clotting factor used will vary depending on which factor is deficient:

- Haemophilia A – clotting factor VIII
- Haemophilia B – clotting factor IX

There are two different regimens used and these are:

- Prophylactic (preventative) clotting factor therapy
- “on demand” therapy - clotting factor replacement as needed to treat bleeding

Choice of regimen will depend on frequency of bleeding without treatment and levels of clotting factors.

Prophylactic (preventative) clotting factor treatment was introduced throughout the UK in the late 1990's; it has revolutionised care for those with severe haemophilia. The aim of treatment is to prevent or minimise bleeding episodes thereby preventing joint damage, which is the most common

cause of disability in the condition. It is most likely to be used in children with very low levels of clotting factors (<1iu/dl).

Treatment involves regular intravenous injection of clotting factors 2 to 4 times a week. Dose adjustments are made and extra doses given depending on activities, Bleeding tendency and clotting factor levels are monitored. Children will require help with all aspects of this until they are able to manage it substantially for themselves.

Children with severe haemophilia who have had prophylactic treatment since early childhood can achieve adulthood with minimal or no joint damage. The advantage of maintaining normal joints through prophylactic treatment is that normal healthy joints are less likely to bleed spontaneously and although careful management of the bleeding tendency is required a normal life with no or minimal disability can be achieved. Despite prophylactic treatment the risk of bleeding persists, as low levels of factors will be present just before the next injection. Some joint damage may occur because of this. In those with poor veins a portacath (or rarely indwelling Hickman line) for administering clotting factors may be used. Children born before this treatment was introduced or outside the UK, or in whom treatment started late are likely to have joint problems or other difficulties related to bleeds that happened before they started using prophylactic clotting factor replacement.

#### Obtaining and storing clotting factors

Clotting factors are home delivered throughout the UK. They are delivered in powder form and need to be mixed with a carrier for injection just prior to use. Supplies should be stored in the fridge but can be kept outside the fridge for up to 3 months making them suitable for carrying around to use at the onset of bleeding. Immediate treatment with clotting factors prevents large and damaging bleeds into joints so minimising damage.

#### Clotting factor replacement 'on demand' therapy

Children with less severe haemophilia or other clotting factor deficiency are likely to use clotting factor replacement only when needed. They will need to administer the clotting factors as described above. Treatment is likely to be intermittent i.e. much less often than in prophylactic treatment. Again the aim is to administer clotting factors as soon as a bleed begins, to minimise long term damage to joints. Children will require help with all aspects of their treatment until they can do it for themselves.

Children with mild haemophilia may need clotting factor replacement after trauma or to cover them for surgery or tooth extraction. There is only

occasional need for children like this to learn how to administer their own clotting factors or to keep supplies at home.

### **Drugs used in haemophilia and clotting factor deficiency**

Children with low levels of clotting factors can use drug treatments to control bleeding. There are two drugs that may be used at home to control bleeding, these are:

- Tranexamic acid – an oral tablet or syrup or mouthwash
- Desmopressin (DDAVP) – this drug can be given as an injection or as a nasal spray

### **Treatment of acute bleeding episodes**

Minor cuts and bruises are easily managed at home with pressure, first aid and administration of clotting factors if necessary. There are no long term consequences from these injuries.

Anything more than minor trauma may require hospital attendance to check for internal bleeding e.g. banging head on cupboard door. This is particularly important for those with moderate and severe clotting factor deficiency.

The most common problem in terms of bleeding, for someone with severe clotting factor deficiency, will be bleeding into joints. Obviously as joint damage is the most common cause of mobility problems for them, minimising harm from bleeding into joints is a very important. Most children will learn how to identify bleeding at a very early stage. Early clotting factor treatment can then be administered. This prevents the large joint haematomas that were so characteristic of this condition, from forming. Because bleeding is stopped at an early stage splintage and joint aspiration are rarely required and parents can manage bleeds at home. Home management consists of:

- Administration of clotting factor
- Rest
- Ice
- Elevation
- Wrapping of affected limb.

Gentle mobilisation can begin the following day. Recovery from a bleed like this will take 2-3 days during which a child will be mobile around the home.

Children with inhibitor may not be able to manage their bleeding at home and are at risk from large haematomas – they may need to attend hospital

and are likely to be managed with splintage in addition to clotting factor treatment. Recovery from an acute bleed in this situation is likely to take two weeks.

### **Treatment of neurological events**

The principles of treatment are to stop bleeding by replacing clotting factors and prevent or reduce permanent disability due to brain damage. Such treatment will take place in hospital and may include neurosurgery.

### **Age and treatment and disabling effects**

Children are most likely to have no or minimal joint damage because they have received prophylactic clotting factor treatment from a young age thereby reducing bleeds and long-term damage. Prophylactic therapy was not used throughout the UK until the late 1990s. Children born before then may have disabling problems related to joint damage. Children with

haemophilia born since then are unlikely to have significant joint damage unless they have an inhibitor.

- [Complications of clotting factor replacement therapy](#)
- [Care and mobility considerations](#)

**What evidence is available?**

Sources of Evidence	Information Provided	Limitation of Evidence
Parent/Carer	First source of information and associated evidence.	Does not have specialist knowledge.
GPFR	The GP will be able to confirm the diagnosis.	Children with haemophilia are likely to receive all their haemophilia care through the Haemophilia Comprehensive Care Centre and will only see the GP about non-haemophilia issues.
Hospital FR – Haemophilia Comprehensive Care Centre - Clinical Nurse Specialist or Consultant Haematologist.	If medical evidence relating to haemophilia and its treatment are required, this is the best source of evidence.	Able to provide complete details on type and severity of haemophilia, treatment required frequency of home treatment and extent of disabling effects. Likely to be able to provide information on all aspects of a child's health in children who require clotting factor therapy or who have complications of bleeding. Will be able to provide detailed information on presence of inhibitor and any disabling effects relating to neurological or joint complications from previous bleeds.

**How long will the needs last?**



This guidance covers -:

Haemophilia A
Haemophilia B
Clotting disorder - Other / type not known

“On demand” therapy

Age at date of claim	Award period
0-13	Limited award until 14 <sup>th</sup> birthday (if needs are identified)

These children will require the parent to remain in a state of preparedness to treat bleeding episodes and administer clotting factors until the child can do this for themselves. On demand clotting factor therapy is likely to be required once a month or less. Children will be able to manage bleeding episodes and the administration of clotting factors substantially by themselves from age 12 to 14.

It is very unlikely that children using ‘on demand’ therapy for their haemophilia will have any neurological or joint problems or ongoing disability related to these.

Routine prophylactic (preventative) treatment of Severe haemophilia – less than 1iu/dl of Factor VIII or Factor IX (less than 1% of normal clotting factor levels)

Age when routine prophylactic treatment starts/started	Award period
0-13	Limited award until 14 <sup>th</sup> birthday.

Children are likely to require help with routine treatment until age 12 to 14 and so awards related to routine treatment are recommended to the 14<sup>th</sup> birthday. This applies to children starting prophylactic treatment for the first

time and those children who have completed inhibitor treatment who will continue with routine prophylaxis.

Needs associated with treatment may persist beyond this age in children with problems related to joint damage or neurological deficit affecting the upper limbs because they have difficulties with manual dexterity.

Children with learning difficulties or behavioural problems may also require help with treatment beyond this age.

Treatment for inhibitor

<b>Age when treatment for inhibitor starts/started</b>	<b>Award period</b>
0-14	Award for 2 years to coincide with duration of treatment for inhibitor. Children aged under 13 years are likely to move from inhibitor treatment to prophylactic treatment for their severe haemophilia and will continue to require help and supervision with this treatment.

Children affected by severe haemophilia with inhibitor require intensive treatment for 18 months to 2 years (sometimes longer) at home. Bleeding episodes are likely to be more difficult to control and short hospital admissions may be frequent. Children under 4 who develop an inhibitor may require extra supervision for bleeding episodes compared to children without inhibitor. All children will require extra help and support from their parents during the inhibitor treatment phase when they are receiving daily intravenous infusions of clotting factor. Parents are likely to prepare and administer the clotting factor for children under 8.

Most children aged 8-14 are likely to need help with some aspects of their treatment. Some children may be able to self medicate but they will still require support and supervision from their parents to ensure clotting factor is administered hygienically, safely and in the right dose. Some children may continue to have inhibitor treatment for more than two years and continue to have needs associated with intensive treatment. However, awards are generally recommended for all children to coincide with the end of the majority

of them receiving treatment for inhibitor as treatment related needs reduce at this point.

Enduring disability related to bleeding episodes

Age at date of claim	Award period
0-14 – with enduring effects related to multiple joint damage or stable neurological deficits	Indefinite award.
0-14 – with enduring effects related to a single joint	Award until joint surgery is planned (if known) or until age 21 if no surgery date known.
0-14 – with enduring effects related to recent onset neurological deficit (within last 18 months)	2 year award.

All children of any age who have had inhibitor may have enduring effects of uncontrolled bleeding including neurological deficits or joint damage. If these effects are claimed and related to multiple joint damage or stable neurological deficits indefinite awards are recommended.

If a single joint is the cause of reduced mobility or difficulties with self care or self treatment, improvement is expected as joint replacement is likely to be offered. A renewal should be conducted following joint surgery (if date known) or at age 21, if no information about joint surgery has been received.

If recent onset of neurological deficit (onset within the last 18 months) is claimed, a 2 year award with renewal is recommended as improvement is expected.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Cause of disability**

For most people with haemophilia small cuts and bruises to the skin will cause more prolonged bleeding than normal but will be manageable by applying pressure to the area. The real risk is internal bleeding into joints, muscles and the brain. Internal bleeding like this is the cause of permanent disability in clotting disorders. In untreated haemophilia bleeding into joints causes scarring and arthritis in the joint at an early age, repeated bleeding

into the same joint exacerbates the problem. Once a large bleed into a joint has occurred the joint becomes inflamed and new blood vessels appear in the joint, these are more likely to bleed than normal blood vessels in a normal joint. With further bleeding more inflammation develops. Eventually the joint becomes very scarred. Scar tissue may fix a joint in one position or significantly reduce the range of movement of a joint. A fixed scarred joint is likely to be termed a 'fixed flexion deformity' in medical evidence. It is the pain associated with inflammation that limits mobility in acutely affected joints. Once scarring and fixing occurs or when range of movement is significantly affected, movement, for example gait when walking becomes abnormal. This places abnormal strain on other joints and they are prone to further bleeds and damage. The commonly affected joints are the large joints and these are the:

- Knees
- Ankles
- Hips
- Elbows
- Shoulders

Bleeding into the brain may cause permanent neurological problems e.g. hemiplegia, epilepsy. In infancy bleeding into the brain may cause cerebral palsy, learning difficulties or epilepsy. A bleed in to the brain that causes a permanent impairment, such as cerebral palsy, during infancy may be the first indication that a child has haemophilia. These impairments are the complications of uncontrolled bleeding.

## **Complications of clotting factor replacement therapy**

### **Development of Inhibitor**

Inhibitors are antibodies to clotting factors VIII and IX. They are made by the body's immune system and attack and destroy the factor VIII and IX proteins in clotting factor concentrates, making treatment ineffective. It is most common in children with severe Haemophilia A. Around 30% of boys with haemophilia A develop inhibitors and around 4% of boys with haemophilia B develop inhibitors. Boys are regularly screened for the presence of inhibitor using a blood test. Treatment is available and the aim of treatment is to stop the body producing the inhibitor. Treatment involves intensive high dose treatment with the clotting factor concerned and sometimes immunomodulating drugs. This therapy regime may involve the use of daily or twice daily injections of factor concentrate for up to a year or even longer in some cases. The inhibitor cannot always be treated and in these cases factor concentrate does not work. Other treatments can be given that help to stop bleeding but they are less effective. Progressive joint damage is likely and

disability is more likely to be present in a child with an inhibitor than in a child without.

### **Blood borne diseases**

No-one with haemophilia in the UK has been infected with Hepatitis B, C or HIV via blood products since 1986. There is a theoretical risk of infection with variant Creutzfeld–Jacob disease (variant CJD) but to date there have been no cases.

### **Care and mobility considerations**

#### **Haemophilia and inhibitor**

Likely treatment related to care

Inhibitors can sometimes be a troublesome complication in the treatment of haemophilia. They can range from being mild to severe and occur when the immune systems of people being treated with blood clotting agents, such as nonacog alfan, start to regard the clotting agents as foreign objects.

The immune system will start to create antibodies to block the effects of the clotting agent. These antibodies are known as inhibitors. If inhibitors are present, the replacement of clotting factors to treat bleeding using factors that bypass the missing one will be less effective and such treatment may need to be given in hospital. Bleeds into joints may require splinting. Recovery from individual episodes is likely to take up to 2 weeks. The parent will need to be available to take the child to hospital and support the child during admissions, which may be frequent and prolonged in addition to administering treatment at home in between.

Treatment related care at home of a typical child with haemophilia and inhibitor

Such children are likely to have an indwelling catheter or line for administration of treatment. This will be either a Hickman line or a portacath. Both lines need to be monitored for infection and the Hickman line will need cleaning and dressing on a daily basis. Parents will be trained to manage the line and administer treatment. Children with inhibitor are likely to be

prescribed daily intravenous infusions of clotting factor. Steps involved in administering this include:

- Storing clotting factors in a clean and appropriately cool environment
- Ensuring the area where treatment is administered at home is clean and hygienic
- Topical anaesthetic cream needs to be applied 30-45 minutes prior
- Preparing powdered clotting factor concentrate with a carrier liquid for administration
- Drawing up the mixed clotting factor in a syringe and checking the dose is correct.
- Partially undressing the child
- Cleaning the Hickman line or skin overlying the portacath.
- Administering the clotting factor through the portacath or Hickman line using aseptic technique to prevent infection.
- Making sure the Hickman line is securely closed and protected or making sure the portacath site is clean and there is no bleeding
- Dressing the child

Intravenous clotting factors are likely to be administered once a day and would be expected to take around half to one hour of the parent's time. The child may be prescribed other drugs in tablet form including immunosuppressant drugs – it is vital that these are taken at specific times of day and the parent will need to ensure that this is done and the drugs are securely stored the rest of the time.

#### Monitoring of a typical child with haemophilia and inhibitor

In addition to administering treatment on a daily basis parents will need to monitor their children for bleeding episodes. This is because children with inhibitor may have uncontrolled bleeding episodes because the inhibitor prevents clotting factor from working. Parents will need to monitor the amount of activity a child does as strenuous activity may increase the risk of a bleed.

Parents will seek to protect their child from injury and monitor their child for bleeding after minor injury. This is because there is an increased risk of significant bleeding after common minor injuries such as a bang to the head. There will be an extra dimension to supervision of children with haemophilia and inhibitor until the child can reliably communicate with others and avoid

injury. Older children with inhibitor do not require extra supervision compared to children without inhibitor because they can communicate the first signs of bleeding to their parents and will be aware of their bleeding tendency.

No extra care or monitoring is required at night.

Over some years the child will gradually take over aspects of their treatment. This will involve at least once daily doses of clotting factor and possibly the use of immunosuppressant drugs. If this is not effective the inhibitor will still be present afterwards.

### Mobility

Because treatment of bleeds is less effective multiple large joints may be affected by arthritis from childhood onwards. If there is joint damage mobility is likely to be restricted because of pain related to arthritis in the hips, knees and ankles. Joint replacement of individual joints will relieve pain from arthritis and prevent further bleeding into that joint but will not improve mobility or range of movement. This is because other joints are affected. Joint replacement is not likely to be performed until the child has stopped growing. If flexion deformity of the knee or hip is present than mobility is especially likely to be reduced, a flexion deformity effectively shortens the affected leg and affects gait. In someone with multiple damaged joints this places further strain on other joints and increases the risk of bleeding when walking.

**Severe haemophilia – less than <1iu/dl of Factor VIII or Factor IX are found in the blood (less than 1% of normal clotting factor levels)**

Treatment related care at home of a typical child with haemophilia on prophylactic (preventative) clotting factor treatment

Likely to be on prophylactic (preventative) clotting factor treatment at home. Clotting factor is likely to be administered 2 to 4 times a week. Such children are likely to have an indwelling catheter or line for administration of treatment. This will be either a Hickman line or a portacath. Either line needs to be monitored for infection and the Hickman line will need cleaning

and dressing on a daily basis. Parents will be trained to manage the line and administer treatment. Steps involved in administering treatment include:

- Storing clotting factors in a clean and appropriately cool environment
- Ensuring the area where treatment is administered at home is clean and hygienic
- Topical anaesthetic cream needs to be applied 30-45 minutes prior
- Preparing powdered clotting factor concentrate with a carrier liquid for administration
- Drawing up the mixed factor in a syringe and checking the dose is correct.
- Partially undressing the child
- Cleaning the Hickman line or skin overlying the portacath.
- Administering the clotting factor through the portacath or Hickman line using aseptic technique to prevent infection.
- Making sure the Hickman line is securely closed and protected or making sure the portacath site is clean and there is no bleeding
- Dressing the child

Administering the treatment is likely to take half to one hour of the parent's time two or four times a week. Extra doses may occasionally be required.

Monitoring of a typical child with haemophilia on prophylactic clotting factor treatment

In addition to administering treatment parents will need to be aware just prior to the next dose of clotting factor that the child will be at increased risk of bleeding – this is because clotting factors are used up and are low before the next treatment. This means a child will require slightly more supervision from their parents during early childhood. For example a minor bang to the head may cause intracranial bleeding in such a child if it occurs when clotting factors are low. Early treatment with clotting factor will stop bleeding and prevent long term disability, early treatment relies on bleeding being recognised and young children will rely on their parent to do this for them.



Children aged 4 and over may be able to report back to their parents after a minor injury or at the first signs of bleeding.

No extra care or monitoring is required at night.

Over some years the child will gradually take over aspects of their care until they can manage it independently. It is very unlikely that any child will have significant upper limb problems that prevent them from doing this. The exception to this will be children who have had inhibitor previously who may have had many uncontrolled bleeding episodes – see guidance under haemophilia and inhibitor for guidance about this.

### Mobility

Mobility will depend on whether there is joint damage to the lower limbs. It is most likely that children receiving prophylactic treatment will have normal or near normal joints and joint function. Children who have had inhibitor in the past may have developed joint or neurological damage at that time. Children with enduring disability related to bleeding episodes for guidance on those cases.

### **Moderate haemophilia - Factor VIII or Factor IX 1-5 iu/dl (1-5% of normal clotting factor levels)**

Likely treatment related to care

Likely to need infusion of clotting factors on demand. Generally, this is likely to be once a month or less. Treatment may be given at home by the parent or child, or in hospital. Whether the parent or child administers treatment at home is likely to depend on how frequently clotting factors are required. The child is unlikely to have an indwelling catheter and so venous access will need to be gained at home using a cannula or butterfly (a type of small cannula that is very easy to use for temporary venous access). All other steps

involved in administering clotting factors will be the same as described under prophylactic (preventative) treatment. These are:

- Storing clotting factors in a clean and appropriately cool environment
- Ensuring the area where treatment is administered at home is clean and hygienic
- Preparing powdered clotting factor concentrate with a carrier liquid for administration
- Drawing up the mixed factor in a syringe and checking the dose is correct.
- Gaining temporary venous access by putting a butterfly cannula into a small vein on the arm or hand.
- Administering the clotting factor in a hygienic fashion
- Removing the butterfly cannula and stopping any bleeding appropriately

Local anaesthetic ('magic cream') may need to be applied 30-45 minutes before the butterfly cannula is put in. This will extend the time taken by parents to administer treatment. The cream needs to be applied to the child. It takes 30-45 minutes to numb the skin before a butterfly can be put in. The clotting factor will then need to be prepared and administered. This may take up to 1½ hours in total.

Treatment is likely to be given only when bleeding occurs so the parent will need to provide appropriate care for the bleeding episode as well – see ['home treatment of bleeding episodes'](#). Although parents will spend less time administering treatment because it is required less frequently the child will still require extra supervision if under 4 to monitor for minor injuries and identify abnormal bleeding early. The parent will need to be in a state of preparedness to provide treatment at short notice and this may be more difficult for a family than having a child on prophylactic treatment – where both parents and other carers have plenty of opportunities to practice administering treatment at predetermined times. It is very unlikely that such children

will have developed complications of uncontrolled bleeding. No extra care or monitoring is required during the night.

**Mild haemophilia – Factor VIII or Factor IX 5-50iu/dl (5-25% of normal clotting factor levels)**

Likely treatment related to care

Clotting factor replacement is only required after significant injury or trauma. It is not required frequently enough to require home supplies and self treatment.

**Mobility**

Immediate treatment of abnormal bleeding episodes with appropriate clotting factors is the norm and enduring disability related to neurological or joint damage in children is not expected in the typical case.

**Care**

Treatment of bleeding, should it occur, will be given in hospital and is only likely to occur with significant trauma. No extra care is anticipated in children of any age.

**Female haemophilia carriers**

No care or mobility needs anticipated.

**Bleeding disorders caused by blood clotting factor deficiencies**

**What is a bleeding disorder?**

Bleeding disorders are those in which the blood fails to clot normally. Blood clotting is the process by which protein factors and platelets activate one another to form a plug at a site of bleeding such as a torn blood vessel to prevent further loss of blood from the site. Over time the initial plug forms into a scab and healing of the torn vessel occurs. Clots are formed by the activation in the blood of special proteins called clotting factors, these factors activate one another in a sequential manner or cascade that results in a blood clot over a few minutes and this stops bleeding. When a clotting factor is missing, only present in low amounts or in an ineffective form the cascade cannot complete. No clot is formed so bleeding continues.

**Names of clotting factor deficiencies**

The most common disease caused by clotting factor deficiency is haemophilia due to deficiency of blood clotting factor VIII. There are other much

rarer clotting factor deficiencies, the absences of which cause bleeding disorders. The rarer types are named for the factor that is missing:

- Factor II deficiency
- Factor V deficiency
- Factor VII deficiency
- Factor X deficiency
- Factor XIII deficiency

### **Treatment of clotting factor deficiency**

The treatment of these rarer clotting factor deficiencies is the same as the treatment of severe haemophilia – clotting factor less than 1% of normal levels. This usually means prophylactic (preventative) treatment with clotting factors several times a week.

Obviously the factor used will be the one that is missing. Factors V, VII and XIII can be manufactured and are called recombinant factors. Factors II and X cannot be manufactured and are extracted and concentrated from human

blood. There is currently no known risk of virus or disease transmission from these plasma products as manufactured today.

## What you need to know about Hearing Loss

### What is Hearing Loss?

The term “hearing loss” encompasses the following terms -:

Hearing impairment, deafness, deafened, partially deaf, hard of hearing  
.....

- [Hearing loss](#)

### What are the effects and signs?

A child with mild deafness will hear conversation one-to-one in quiet background conditions .....

- [Effects of hearing loss](#)

### How is it assessed?

All babies in England are offered a hearing screen routinely within a few weeks of birth as part of the Newborn Hearing Screening Programme (NHSP).....

- [Assessment of hearing loss](#)

### How is it treated and managed?

Bone-conduction or surgically implanted Bone Anchored Hearing Aids can be used in specific circumstances .....

- [Treatment for hearing loss](#)

### What evidence is available?

A speech and language therapist may provide information about symptoms, investigations, treatment and resulting disability or needs .....

- [Evidence](#)

### How long will the needs last?

- [Prognosis and duration of the award](#)

## What is Hearing Loss?

The term “hearing loss” encompasses the following terms -:

Hearing impairment, deafness, deafened, partially deaf, hard of hearing.

Levels of deafness can be divided into different categories using the following definitions.

<b>Severity of deafness</b>	<b>Range of hearing loss in decibels (dBHL)</b>
Mild deafness	A child with mild deafness has 21 to 40 decibels of hearing loss (dBHL) on average in their better ear.
Moderate deafness	A child with moderate deafness has 41 to 70 decibels of hearing loss (dBHL) on average in their better ear.
Severe deafness	A child with severe deafness has 71 to 95 decibels of hearing loss (dBHL) on average in their better ear.
Profound deafness	A child with profound deafness has more than 95 decibels of hearing loss (dBHL) on average in their better ear.

Hearing loss can also be subdivided into:

Conductive hearing loss - resulting from interference with the passage of sound through the external and middle ear to the inner ear.

Sensorineural hearing loss - resulting from disorders of the cochlea

(in the inner ear) or the auditory nerve; it is called sensory where the problem is in the inner ear or the cochlea and neural where the problem is in the

auditory or cochlear nerve – this results problems in loudness and speech discrimination.

Pre-lingual deafness means that the child was born deaf or became deaf before the development of language skills.

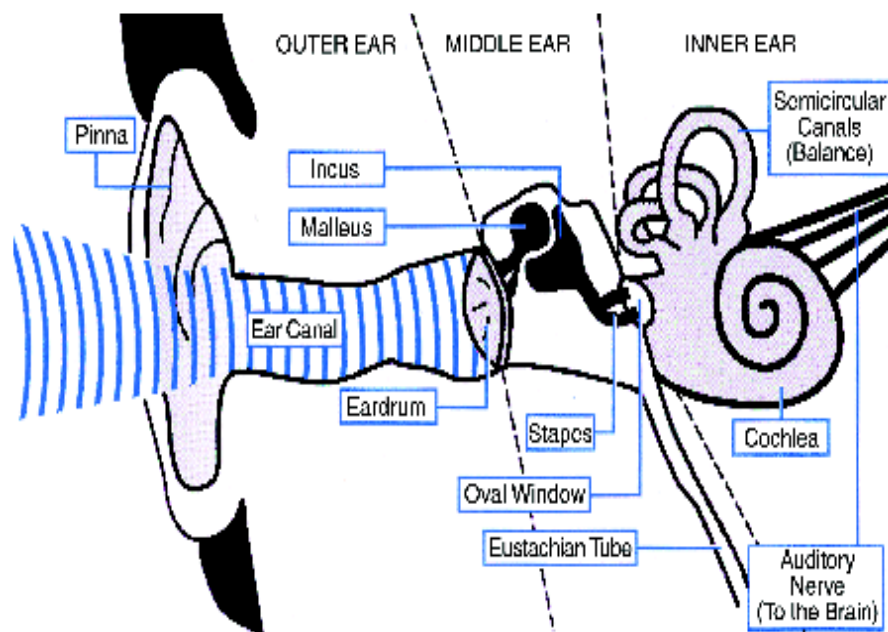
Post-lingual deafness means that the child had already acquired speech when the deafness was diagnosed.

Auditory neuropathy spectrum disorder (ANSD) is a condition characterised by a problem in the conduction of the sound energy through the auditory nerve and its formations up to its central connections in the brain – this may

result in completely normal hearing or varying degrees of hearing loss and processing problems.

Auditory processing disorder (APD) is a condition characterised by an intact hearing at the level of the ear but problems in processing the incoming signal from the ear in the hearing centres in the brain.

Figure 1 - Anatomy of the ear



Anatomically, the ear is divided into three parts, the outer, middle and inner ear.

The outer ear consists of the trumpet shaped auricle or pinna and the external auditory meatus (outer ear canal), which together funnels sound to the middle ear. The middle ear is separated from the outer ear by the eardrum or tympanic membrane. It is an air filled cavity in the temporal bone of the skull, connected to the throat through the Eustachian tube. The three auditory ossicles, malleus, incus and stapes are small bones that provide a mechanical link between the tympanic membrane and inner ear. The inner ear is divided into the organ of balance (vestibular labyrinth) and the organ of hearing (the cochlea). This contains the Organ of Corti and its thousands of



sensory hair nerve cells. The nerve of hearing or auditory nerve passes to the brain from the inner ear.

### **Incidence/Prevalence**

There are no comprehensive national statistics on the number of children and young people in the UK with hearing loss. The best estimate on the total number of children and young people aged 0 – 25 in the UK with permanent bilateral (affecting both ears) hearing loss (greater than 40dBHL) is 34,800. It is estimated that 1 in 1000 babies born in the U.K. has a severe to profound hearing loss at birth. About 50-90 percent of permanent childhood hearing impairment may develop after birth.

Thousands more children will experience mild to moderate deafness. A child with this level of deafness can hear some sounds, so it may be harder to recognise that they have some level of hearing loss.

Around 10% of all babies who fail the newborn hearing screening may have ANSD. Around 8% of school going children may have some form of APD.

- [How you hear](#)

### **What are the effects and signs?**

#### Mild deafness

Within this range a hearing aid may be required depending upon the frequencies affected by the hearing loss and whether the child has a dual sensory disability. They will be unlikely to require communication aids, or use manual communication (BSL) unless they have a co-existing condition (e.g. Down's syndrome). A child with mild deafness will hear conversation one-to-one in quiet background conditions. They might not hear when sounds are presented from over a distance (e.g. they might be unable to hear the teacher from the back of a classroom or someone calling across an outdoor space).

There might be disproportionate central processing of sounds due to a mild hearing loss and these children usually may slip grades in academia or manifest changed behaviour from the social and communication angle.

#### Hearing loss in one ear

Unilateral hearing loss (hearing impairment of one ear only), regardless of how severe the level of hearing loss is, should not normally affect the ability to perform normal day to day activities appropriate to the child's age. However, these children may have an inability to hear in the presence of

background noise which may be the class room or able to localise the source of a sound which may be a safety issue for example during crossing a road.

Again, in some children; there might be a disproportionate central processing which intrudes in their behaviour and academia and also language development.

#### Moderate deafness

A child with moderate deafness finds it difficult to follow speech without a hearing aid or other technology to amplify sound. The child may lip read and therefore finds it harder to follow speech when not one-to-one even with hearing aids in.

They are likely to gain benefit from amplification either from a hearing aid or by external devices. They may rely on a combination of amplified sound and lipreading. They will usually have speech that can be understood by strangers and will be able to hear and understand a normal voice at 1 metre with appropriate amplification. Background noise will have a notable affect in understanding speech.

#### Severe and profound deafness

A child with severe or profound deafness will probably lip read and may use sign language. When hearing aids are not powerful enough for the child they may be fitted with a cochlear implant (a specialist hearing device surgically implanted into the inner ear).

A few children are deaf because they were born without a cochlea (inner ear) or hearing nerve. In these cases the child will have no hearing at all and will not benefit from amplification devices. These children will communicate using sign language. They may be eligible for a brainstem implant, but this is rarely carried out. Speech may be affected such that they may not be understood clearly by strangers.

Hearing aids and cochlear implants do not restore hearing to normal levels. They use microphones which have a limited pick up range and may amplify extraneous background noise as well as speech. Deaf children may use a

variety of other equipment to complement their prescription hearing aids or cochlear implants e.g. text phone, loop systems etc.

Indicators of severe functional restriction

Children with a severe functional restriction are likely to:

- Have Hearing loss of 71dB or more with or without hearing aids/cochlear implant
- Rely on text rather than speech when using phones
- Use manual communication if the onset of hearing loss was before the development of language skills (prelingual deafness)
- Be eligible for a cochlear implant
- Be unable to hear and understand a raised voice at 1 metre with or without hearing aids/cochlear implant
- Have speech affected such that they may not be understood clearly by strangers
- Have a medical condition making use of hearing aid or cochlear implant impossible (e.g. absent or severely deformed cochlea or hearing nerve, Auditory Neuropathy Spectrum Disorder)

Vestibular hypofunction

The majority of children with profound hearing loss have vestibular hypofunction (inadequate function of the body's balance system). Some children with severe hearing loss and occasionally children with moderate hearing loss also have vestibular hypofunction.

About a third of children with any degree of hearing loss may have vestibular hypofunction (inadequate function of the body's balance system).

Vestibular hypofunction may cause early delay in gross motor development though children tend to catch up, usually by age 5. They have a delay in

walking, the mean age for walking being 18 months compared with 12 months for a normal child.

It affects the ability of children to right themselves if they are off balance and as a result they are more likely to fall. They may have difficulty walking over rough terrain or in low light conditions.

[Causes of hearing loss](#)

[Other types of deafness](#)

[Deeming Provisions](#)

### **How is it assessed?**

All babies in England are offered a hearing screen routinely within a few weeks of birth as part of the Newborn Hearing Screening Programme (NHSP). For more details about this - see [NHSP](#).

Automated Otoacoustic Emission (OAE) screening test

This is the initial test and works on the principle that a healthy cochlea will produce a faint response when stimulated with sound. A small earpiece (containing a speaker and microphone) is placed in the child's ear. A clicking sound is played and if the cochlea is working properly, the earpiece will pick up the response. This is recorded on a computer.

Automated Auditory Brainstem Response (AABR) screening test

Children who fail the OEA (about 15%) have a second test, the Automated Auditory Brainstem Response (AABR) screening test. This test measures whether sound is being sent from the cochlea and through the auditory nerve to the brain. The audiologist places three small sensors and a set of headphones on the child's head and in the automated test the results are interpreted by a computer.

Diagnostic Auditory Brainstem Response test (ABR)

Children who fail the AABR are referred for further investigations which are usually carried out at the local audiology clinic. This usually involves a

diagnostic ABR where different levels of sound are used and the results are interpreted by a clinician.

If the diagnostic ABR shows a unilateral hearing loss the child is usually monitored. If the diagnostic ABR shows a bilateral permanent hearing loss, the child will be fitted with hearing aids.

### Behavioural tests

Older children may have their hearing assessed by behavioural tests. These tests use toys and play as part of the assessment and involve the child listening for a variety of sounds as part of the assessment.

### Cochlear microphonics

Children who exhibit Auditory neuropathy spectrum disorder (ANSD) in the screening process undergo this special test to confirm or exclude ANSD.

### Visual response audiometry (VRA)

Visual response audiometry (VRA) is suitable for children from six months to about two and a half years. Using a machine called an audiometer, sounds of different frequencies and loudness are played through a speaker. When the child hears the sound, they will turn their head when a visual reward is “activated” such as a toy lighting up or a puppet. The test can be conducted through small earphones if the audiologist wants information about each ear separately.

### Pure tone audiometry

This is used from about the age of three. Younger children are shown how to move a toy (for example, putting a peg into a board) each time they hear a sound. Older children are asked to respond to sounds by saying yes or pressing a button.

An audiometer generates pure tone signals of frequency 125Hz, 250Hz, 500Hz, 1,2, 4 and 8kHz at variable intensities ranging from –10 dB to +120 dB usually in steps of 5dB.

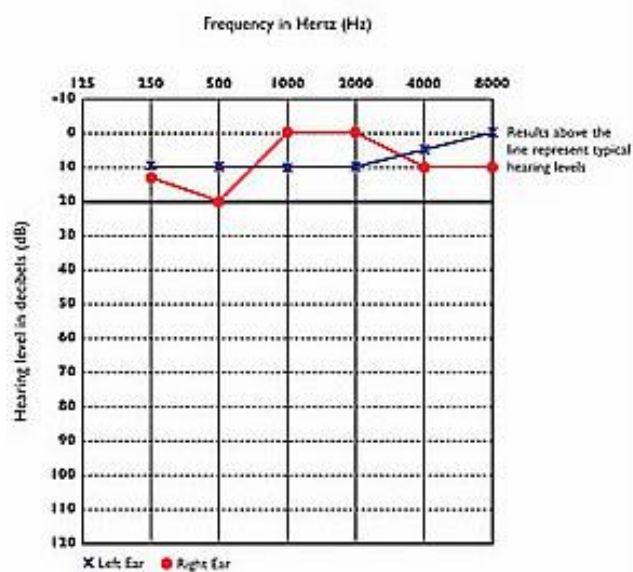
Signals of decreasing intensity at each frequency are presented to the child tested, from a level the child can hear to find the point at which they fail to hear. The hearing threshold levels are usually plotted on a graph or audiogram, with sound intensity (dB) on the y (vertical) axis and the frequency (Hz) along the x (horizontal) axis. Standard symbols are used to denote

right (o) and left (x) ears for air conduction and right ([ or Δ) or left (] or Δ) ears for bone conduction.

The hearing threshold is defined as the quietest sound heard by the child when being tested. A normally hearing child would expect to have a threshold of 20dB or better, and this represents no hearing loss on the audiogram.

It should be noted that audiometry is a subjective test of hearing.

### Normal audiogram



Air conduction assesses the function of both the conduction (outer and middle ear) and sensorineural (cochlea and auditory nerve) components of the ear. To measure air conduction (AC), the sounds come through either headphones, earphones placed inside the child's ear or sometimes through a speaker (when the test is known as soundfield audiometry) and the signal passes by air conduction through the outer and middle ear. It is then transmitted to the inner ear, auditory nerve and auditory cortex of the brain.

Bone conduction (BC) assesses the function of the cochlea and auditory nerve. To measure bone conduction, the signal stimulates the cochlea directly by the application of the vibratory stimulus to the skull.

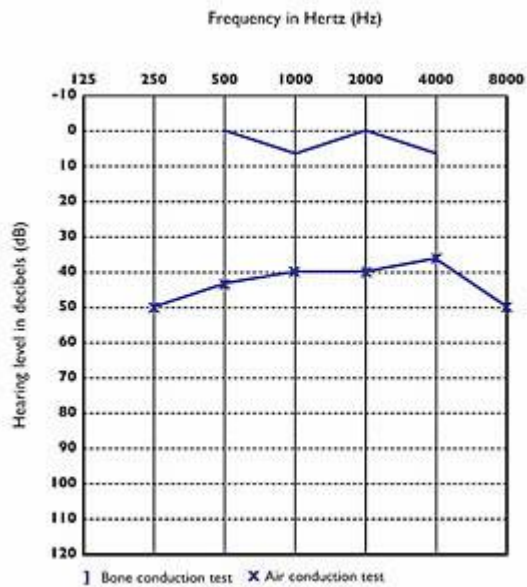
Using these two measures the type of hearing loss can be classified into conductive, sensorineural or mixed type.

### Conductive hearing loss

If there is a problem in the external or middle ear (conductive hearing loss) the AC threshold will be higher than the BC threshold because the child will

hear better by bone than air conduction. This is called the air bone gap (ABG). The greater the air conduction loss, the greater will be the ABG.

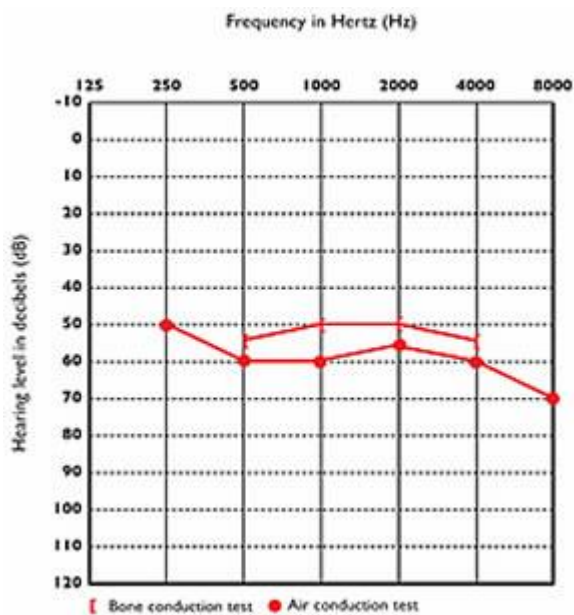
Audiogram showing conductive hearing loss in the left ear



Sensorineural hearing loss

If there is a problem with the cochlea or the auditory nerve, the AC and BC thresholds will be the same.

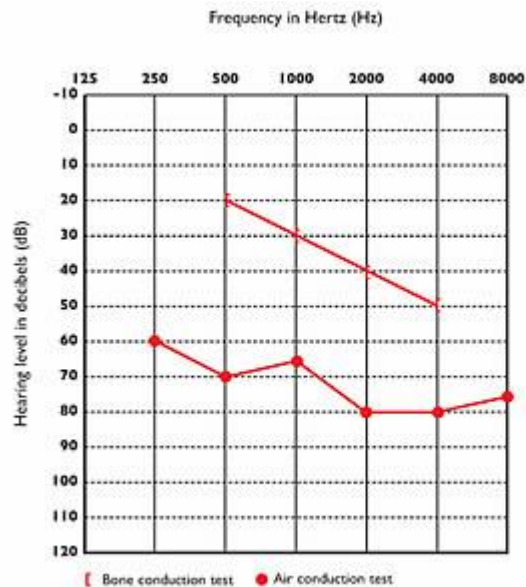
Audiogram showing sensorineural hearing loss in the right ear



## Mixed hearing loss

Mixed hearing loss is a reduction in hearing of both AC and BC.

Audiogram showing mixed hearing loss in the right ear



## Speech discrimination tests

Speech discrimination tests check the child's ability to hear words at different listening levels. The tester asks the child to identify toys or pictures, or to copy words spoken by themselves or from a recording. From this the tester can assess the quietest level at which the child can correctly identify the words used. The test can also be used to assess lip reading and signing skills.

## Tympanometry

Tympanometry is not a test of hearing. It is used to check how well the moving parts of the middle ear are working. A small earpiece is held in the ear canal. A pump causes the pressure in the ear canal to change. The eardrum should move freely in and out with the change in pressure. The earpiece measures this by checking the sound reflected by the eardrum. If the eardrum is not moving freely, there is likely to be some fluid or another problem with the middle ear. A build up of fluid is usually due to glue ear.

## Advanced diagnostic testing

These include otoacoustic emissions and auditory brain stem responses for diagnostic purposes in any hearing loss. For ANSD, intact cochlear function needs to be demonstrated with abnormal brain stem responses. For



Auditory processing disorder (APD), there is a full battery of diagnostic behavioural tests.

### **How is it treated and managed?**

#### Conductive hearing loss

This group of hearing loss may be treated either with hearing aids or with surgery or in milder cases, no treatment is necessary. Bone-conduction or surgically implanted Bone Anchored Hearing Aids can be used in specific circumstances. Sometimes surgical interventions may be offered as treatment for glue ear (insertion of grommets), otosclerosis (stapedectomy) and perforated tympanic membrane (myringoplasty or tympanoplasty). Middle ear implants may be used for some mixed and conductive losses in patients where conventional hearing aids cannot be used.

#### Sensorineural hearing loss

This is usually managed by the use of hearing aids. Surgically implanted devices such as Cochlear Implants or Middle Ear Implants may be used in specific circumstances. Middle ear implants may be used for some losses in patients where conventional hearing aids cannot be used.

### **Aids for Improving Hearing**

#### Hearing aids

Hearing aids are devices that amplify sounds. They can be used in both conductive and sensorineural hearing loss. Their use usually improves a child's hearing to the level of the speech frequencies so that they are able to hear conversation one-to-one and in small groups. Hearing aids do not return hearing to the normal ranges and additional tactics may be needed in noisy situations (e.g. lipreading) and when listening at a distance. Children who are severely to profoundly deaf bilaterally who get limited or no benefit from conventional hearing aids may be fitted with a cochlear implant. Hearing aids last 5 years on average, and in the UK all children obtain them free from the NHS. Rarely, they have privately purchased hearing aids from a registered Hearing Aid Dispenser.

There are occasions when a hearing aid may not be worn, for example when the child is in the bath or when swimming.

The hearing aid consists of:

- A microphone to pick up sound signals
- A battery powered amplifier to amplify sound signals
- A receiver to deliver the amplified sound to the ear canal

Digital aids are programmable to individual hearing requirements. The settings can be controlled to match the hearing loss frequency only, thereby limiting amplification of background noises. The benefit is limited in real life

situations. Many digital hearing aids are designed to reduce steady kinds of background noise, such as the rumble of traffic or the whirr of a fan. This can make listening more comfortable, but it does not necessarily enable the user to pick out a single voice from everything else going on, especially when several people are talking at once. Directional microphone systems amplify sounds that come from in front of the person more than sounds to the side or behind them. This makes it easier for the person to focus on what they want to listen to in a noisy place but means they will not hear a call from behind. The user can switch between directional and all-round sound, depending on what they need to hear at the time. Some digital aids will detect where the noise is coming from and automatically adjust to reduce the noise selectively. However, a hearing aid cannot know what the person wants to listen to. Directional hearing remains a problem for many hearing aid users.

They are customised to the person and are normally available in the “behind the ear” style, on the NHS.

Hearing aids require reasonable care and cleaning and are powered by dry batteries which need replacing from time to time. There are a number of precautions that parents may take to prevent children getting hold of batteries, such as child proof battery locks that can be fitted by an audiologist.

Children under 3 years - Hearing aids are not “baby safe” and there is a risk of choking on the soft components, such as tubing and ear moulds. Therefore, supervision will be needed for children under 3 years who wear a hearing aid and carers are advised to remove the hearing aid(s) if the child is to be left alone even for a brief period of time.

Children between the ages of 3 and 8 years - If a child has a history of pulling out or switching their hearing aid(s) off, carers are advised to be vigilant and check the hearing aid(s) at least every 30 minutes during the day.

At nighttime - Carers are advised to take hearing aid(s) out at night. Occasionally a child becomes distressed when the hearing aid is removed and,

in this case, carers are advised to wait until the child falls asleep before removing the aid(s).

## Cochlear Implants

Cochlear Implants are surgically - implanted devices, which directly stimulate the auditory nerves in the cochlea.

Children with severe and profound sensorineural hearing loss who have not benefited from a trial of hearing aids are usually referred to a specialist cochlear implant team for assessment.

There are a number of factors that are taken into consideration before a child is offered a cochlear implant. These include:

- Hearing. Is the hearing loss severe to profound and are hearing aids insufficient?
- Age. In children who are born deaf, the acquisition of speech and language is related to the age at which cochlear implantation takes place. The best results are obtained in children who receive a cochlear implant before the age of two years. In one study, 90% of children who received a cochlear implant below the age of 2 years entered mainstream education; 60% for implantation between age 2 and 4 years; and 20% for those after the age of 4. The maximum age for implantation is considered to be around the age of five years for those born deaf although those with acquired and progressive deafness can be implanted later in life
- Stable family support is required because of the need for cooperation of the child's carers in a long term rehabilitation programme
- Absence of medical contraindications to surgery such as chronic ear infections or abnormal inner ear anatomy

After implantation, children who've never heard require a few years of rehabilitation to teach them to listen to and make sense of the new sounds and to optimally tune the device. Most children are eventually able to hear conversation without lip reading and use spoken language for everyday communication.

## Bone conduction hearing aids

Bone conduction hearing aids may be given to children who have a conductive deafness. The hearing aid vibrates in response to sounds going into the

microphone and the vibrations are transmitted to the cochlea through the bones in the skull.

### Bone anchored hearing aids

Bone Anchored Hearing Aids are surgically implanted devices that transmit sound directly to the inner ear through the bones of the skull and can be suitable for children with a permanent conductive hearing loss who have already tried a bone conduction aid. They tend to be more comfortable and give a better sound quality than bone conduction aids.

### Middle Ear Implants

A middle ear implant (MEI) is essentially a hearing aid, but one in which the receiver or the entire hearing aid is surgically implanted into the middle ear. Unlike cochlear implants, MEIs are designed for people with significant residual hearing. They have a number of advantages over conventional hearing aids. These include elimination of acoustic feedback, avoidance of the

need to insert an earpiece in the ear canal and improved cosmetic appearance. MEIs are currently rarely used in children.

Click on the links for details of -:

[Communication methods](#)

[Communication support](#)

[Environmental aids](#)

**What evidence is available?**

<b>Sources of evidence</b>	<b>Information Provided</b>	<b>Limitation of Evidence</b>
Claimant / Carer	First source of information and associated evidence.	Does not have specialist knowledge.
Teacher of deaf children  Note: This is the preferred source of evidence if available.	A factual report should be sent to the Teacher of the Deaf employed by the relevant local authority specialist education support service, or if the child attends a special school for deaf children, a factual report should be sent to the Teacher of the Deaf employed by the school.	May not have information about symptoms, investigations and treatment.
Audiologist, ENT or audio-vestibular physician	Symptoms, investigations and treatment/management, and may have information about resulting disability or needs.	May not have information about resulting disability or needs.
Social worker	Resulting disability or needs.	May not have information about symptoms, investigations and treatment.
GP	Symptoms, investigations and treatment/management.	Does not have specialist knowledge.
Speech and language therapist	Symptoms, investigations, treatment and resulting disability or needs.	N/A.
Orthoptist	Assessment of vision (visual acuity and fields)	May not have information about symptoms, signs, investigations other than assessment of vision, treatment/management, and unlikely to have information about resulting disability or needs

Optometrist	Symptoms, signs, investigations including assessment of vision, treatment/management, and likely to have information about resulting disability, needs and provision of low vision aids.	None
If considering entitlement to H/R Mobility component under the Severely Visually Impaired (SVI) provisions, the following evidence source must be used:		
Consultant Ophthalmologist	Symptoms, signs, investigations including assessment of vision, treatment/management, and likely to have information about resulting disability or needs	None

[List of NHS hospitals with ophthalmology departments](#)

### How long will the needs last?

Details of each age group depending on the date of onset of the condition

Otitis Media with effusion (OME) - previously known as Chronic Secretory Otitis Media

Other hearing conditions

The most common hearing conditions listed in the table below are covered by this duration guidance.

<b>Impairment</b>
Conductive hearing loss due to Trauma
Other causes of conductive hearing loss / type not known
Deafness – congenital / Pre lingual
Sensorineural hearing loss due to Trauma
Sensorineural hearing loss - Other causes of / type not known
Hearing loss - mixed
Disease affecting hearing & balance - Other/ type not known

Also consult the Visual Impairment guidance for award duration advice.

**Details of each age group depending on the date of onset of the condition**

Date of onset less than 2 years

For a child with hearing loss of less than 2 years the response to treatment is likely to be clear for the majority within 2 years.

<b>Age at date of claim</b>	<b>Award Period</b>
Up to 1	Award to age 3
1 -15	2 year award

Date of onset more than 2 years

A child with hearing loss of more than 2 years is likely to be provided with aids and adaptations and adapt to their hearing loss with increasing age. At age 16 it should be clear whether the child has fully adapted or not.



Age at date of claim	Award Period
1	Not applicable
2 – 7 cochlear implant surgery done	2 years (minimum) from date of cochlear implant
2 – 7 cochlear implant surgery not done	Award to age 8 (or for 1 year whichever is the longer)
8 – 13 (cochlear implant surgery not done/done but of limited success)	To age 16
14 -15 (cochlear implant surgery not done/done but of limited success)	2 year award

#### Reason for Renewal

The majority of children who are considered for cochlear implantation will have had this performed by the age of 8. After cochlear implantation, children require a few years of rehabilitation to teach them to listen to and make sense of the new sounds and to optimally tune the device. Most children are eventually able to hear conversation without lip reading and use spoken language for everyday communication. Therefore an award for a minimum of 2 years should be considered after cochlear implantation.

#### **Otitis Media with effusion (OME) - previously known as Chronic Secretory Otitis Media**

The majority of children with OME (glue ear) will improve without the need for treatment or medical intervention and almost all children grow out of the condition. OME can be relieved with a surgical procedure to drain the fluid and insert temporary 'grommets' in the eardrum. However, a smaller proportion suffer with persistent long term glue ear and may be unsuitable for surgery. Some of these children use hearing aids. Such cases should be discussed with Medical Services.

#### **Other hearing conditions**

With the exception of Chronic Suppurative Otitis Media the conditions below are uncommon and for this reason there is no clinical information about them in this guidance but they may have care / mobility needs depending

on the degree of hearing loss. Medical Services advice should be obtained in these cases.

<b>Impairment</b>
Otitis externa - chronic
Chronic Suppurative Otitis Media
Mastoiditis
Otosclerosis

Hearing impairment with visual impairment. You may need to consider whether H/R Mob Severely Visually Impaired (SVI) or blind/deaf deeming provisions are satisfied. See [Visual Impairment Deeming Provisions](#)

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **How you hear**

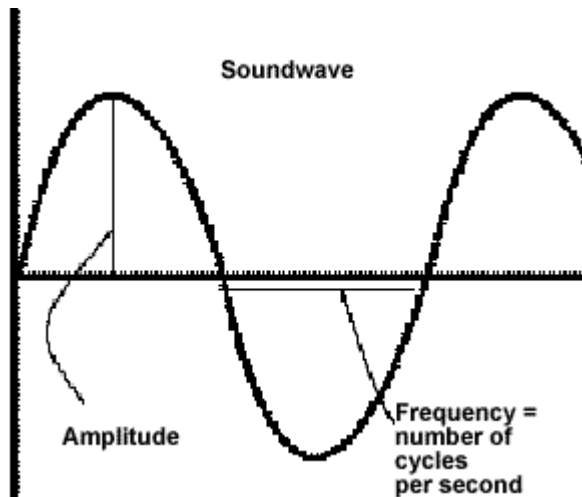
Sound waves in the air are collected by the auricle and passed down the external auditory canal to the eardrum making it vibrate. These vibrations are then transmitted across the middle ear by the ossicles into the fluids of the inner ear cochlea. This moves the outer and the inner hair cells, which in turn activate the auditory nerve. The nerve impulses are transmitted to

the brain along the auditory nerve. The brain processes the sounds and the final perception occurs in the frontal lobe of the brain.

## Sound

Sound waves are characterised by their frequency (pitch) and amplitude (loudness or intensity).

Figure 2 - Sound wave



Higher frequencies are perceived as high-pitched sounds and vice versa for lower frequencies. Loud sounds have high amplitude and quiet sounds have low amplitude.

Frequency is measured in Hertz (Hz) and amplitude is measured in decibels (dB).

Approximate loudness of common sounds	In dBs
Whisper	30dB
Normal conversation	Up to 60dB
Loud shout at 1 metre	85dB
Lawnmower	90dB
Pneumatic drill	100dB
Gunshot	140dB

It should be noted that, although a loud shout has a level of about 85 dB at a distance of one metre, the maximum shout, which can be sustained for more than a few words, is about 90 dB.

Human ears detect sounds in the frequency range of approximately 20Hz to 20kHz but most information in speech is contained in the frequency range of approximately 500Hz to 4kHz.

### **Causes of hearing loss**

#### **Conductive hearing loss**

Conductive hearing loss can be congenital or acquired. Congenital conditions include abnormalities of the pinna, outer ear canal, tympanic membrane and auditory ossicles. Acquired conditions causing conductive hearing loss include infections in the outer ear canal, wax, perforation of the eardrum and glue ear.

Glue ear is the commonest cause of temporary mild to moderate hearing loss in children. It is presumed due to obstruction of the eustachian tube which runs from the middle ear to the back of the throat, resulting in fluid accumulation in the middle ear. Children under five are the largest group affected, but it can persist into adolescence. It can result in delayed speech development and can affect children's behaviour and their educational progress.

Changes in behaviour, becoming tired and frustrated, lack of concentration, preferring to play alone and not responding when called may indicate glue ear. These signs can often be mistaken for stubbornness, rudeness and

being naughty. As a result many children with glue ear are misunderstood or labelled as “difficult”.

A prolonged period of time with reduced hearing can affect the way in which a child’s speech develops. For example, parts of words may not be pronounced clearly. However, in the longer term, speech is not affected in this condition. Children with glue ear may also fall behind at school and become disruptive if they do not have extra support.

Middle ear infection is common in infants and children but can affect people of any age. Acute otitis media is one of the most common illnesses of childhood and is often recurrent particularly up to age 7 years. Eighty to ninety percent of cases of otitis media occur in children under the age of 5 years. Early acute otitis media may predispose a child to develop chronic suppurative otitis media. Other factors like abnormal eustachian tube dysfunction as seen in children with cleft palate and Down’s syndrome, patulous Eustachian tube and systemic immune deficiency are associated with chronic otitis media.

### Sensorineural hearing loss

Sensorineural hearing loss can be congenital or acquired. Congenital conditions include:

- Infections. The most common cause of congenital sensorineural hearing loss is cytomegalovirus (CMV) infection during pregnancy and the resulting hearing loss can be progressive. Much less common congenital infective causes of sensorineural hearing loss include syphilis and toxoplasmosis during pregnancy. Congenital CMV, toxoplasmosis and syphilis may present with delayed onset sensorineural hearing loss months to years after birth. Rubella, once the most common infective cause of congenital sensorineural hearing loss is now uncommon in the UK as a result of effective vaccination programmes
- Difficulties encountered around the time of birth e.g. prematurity, oxygen deprivation, severe jaundice etc can all cause hearing loss and especially auditory neuropathy spectrum disorder
- Auditory Neuropathy Spectrum Disorder (ANSD) can be caused by genetic factors, prematurity, severe jaundice at birth. ANSD causes mild to profound deafness with an additional distortion of sound that makes understanding speech very difficult (over and above what would be expected from the level of hearing loss alone). In severe cases children cannot use hearing aids which amplify the distortion further and may use sign language as their main communication mode. A cochlear implant is an option for some children with ANSD
- Genetic conditions may be a stand alone hearing loss or part of a syndrome where other organs in addition to the ear are

affected. Mutation in the Connexin 26 gene is the commonest non syndromic genetic cause of hearing loss

The most frequently encountered genetic syndromes are:

Usher syndrome is a condition in which there is an association of deafness and retinitis pigmentosa. Problems with balance are common. The degree of hearing loss in Usher syndrome varies from congenital and total to middle aged onset and partial. Usher syndrome may account for up to 6% of people with congenital hearing loss. There are three types of Usher syndrome, type 1, 2 and 3. Types 1 and 2 are the most common types. The features of the different types are shown in the table below.

	<b>Type 1</b>	<b>Type 2</b>	<b>Type 3</b>
Hearing	Profound bilateral hearing loss from birth. May obtain little or no benefit from hearing aids. May use a cochlear implant.	Moderate to severe hearing loss from birth. Most can obtain benefit from hearing aids.	Normal hearing at birth. Progressive loss in childhood or early teens. Usually require hearing aids by mid to late adulthood.
Vision	Decreased night vision before age 10 with rapid progression to complete blindness.	Decreased night vision late childhood or teens with slower progression than type 1.	Blind spots appear by late teens to early adulthood. Usually blind by mid adulthood.
Balance	Problems from birth. Delay in sitting and late walking (18 months).	Normal balance.	Balance usually normal or near normal but problems can develop later in life.

Pendred syndrome is a condition which causes hearing loss, and it may also affect the thyroid gland and the vestibular system in the inner ear resulting in an impairment of balance. Hearing loss may start at any time from birth to the age of three. It is progressive but occurs in stages so that after a sudden decrease in hearing the person's hearing will nearly return to its previous level. Hearing loss is bilateral but often greater in one ear than the other. Some people eventually become totally deaf. About sixty percent of people will develop a swelling of the thyroid gland in the neck (goitre) during

their lifetime. Most people do not have balance problems. Some babies may have a delay in walking independently.

Waardenburg syndrome (WS) is a condition that causes varying degrees of hearing loss and associated skin and hair pigmentation. There are at least four types of Waardenburg syndrome. The most common are types 1 and 2. Type 1 is associated with an unusually wide space between the corners of the eyes. Twenty percent of people with type 1 have hearing loss. People with type 2 do not have a wide space between the inner corners of their eyes, but have many other WS characteristics. About 50 percent of people with type 2 have hearing loss.

- Late onset causes include genetic conditions presenting late, anatomical abnormalities of the inner ear (the so called third window syndromes), or other acquired causes for example infections (usually meningitis), trauma, and ototoxic medications (e.g. certain antibiotics, chemotherapy)

### **Other types of deafness in addition to bilateral, permanent deafness**

#### Unilateral deafness

Unilateral deafness affects only one ear. The deafness can range from mild to a complete loss of hearing (profound). Unilateral deafness particularly affects the child's ability to work out which direction sounds are coming from (localisation) and to hear speech clearly in background noise. The majority of children with unilateral deafness manage well in a hearing environment, however there are situations where they may find it difficult to hear well enough, particularly in classrooms and when outside. Children with unilateral deafness have particular safety considerations e.g. when crossing the road. There are approximately 23,300 – 27,000 children aged 0 -18 years with a unilateral or mild hearing loss in the UK.

#### Temporary deafness

The most common cause of temporary deafness is a middle ear condition known as otitis media with effusion (OME) or "glue ear". This is caused by a build up of fluid in the middle ear that prevents sounds passing to the inner ear efficiently. An estimated 1 million 0-8 year olds experience temporary deafness caused by this condition. That is, one in five children in the UK. The majority of children with glue ear will improve without the need for treatment or intervention and almost all children grow out of the condition. OME can be relieved with a surgical procedure to drain the fluid and insert temporary 'grommets' in the eardrum. However, a smaller proportion of children experience persistent long term glue ear and may be unsuitable for surgery. Some of these children use hearing aids. Particular groups of children may

suffer with glue ear for many years e.g. children with [Down syndrome](#) and children with cleft palate.

Other causes of temporary deafness include external ear infections; build up of wax and very rarely, the self limiting condition where there is a build up of fluid in the inner ear.

### **Newborn Hearing Screening Programme (NHSP)**

About one to two babies per 1,000 births are born with a hearing loss that will affect their social and language development. Early screening means that if a baby has a hearing loss the parents can receive the information and support they need at an early stage.

The Newborn Hearing Screening Programme (NHSP) was commissioned by the Department of Health to implement newborn hearing screening throughout England; from March 2006 all eligible babies born in England have been offered the screen.

Newborn hearing screening might take place in a variety of settings, including hospital, the baby's home, clinic or surgery. It involves the use of specific equipment and liaison with parents, health professionals and other staff, in particular the midwifery and health visiting teams. The hearing screen is offered either in hospital prior to discharge home (75% of NHSP screening sites) or by the health visitor at about 10 days of age (25% of NHSP screening sites). Screening would usually take place while the baby is settled or in natural sleep.

Mothers are given the National Screening Booklet, by community teams, during the antenatal period. Please contact us if the screening booklets do not give you enough information.

### **Screening methods**

Two screening methods are used:

#### **Automated Oto-Acoustic Emissions (AOAEs)**

A small, soft-tipped earpiece is placed in the baby's outer ear and soft clicking sounds are played. When an ear receives sound, the cochlea produces a sound in response. These responses can be picked up by the tiny microphone earpiece.

#### **Automated Auditory Brainstem Responses (AABRs)**

Specially designed baby headphones deliver quiet clicking sounds. Responses from the baby's hearing nerve can be picked up via three small sensors that are placed on the baby's head and neck.

Babies referred from the hearing screening programme should be seen for a full audiological assessment within four weeks of the screen's completion. If they are under 40 week's gestation we try to delay the diagnostic



assessment until this time, but would liaise with the parents to try to avoid too much anxiety.

## **Communication methods**

Deaf children often use a combination of communication approaches depending on the level of hearing that they have, the situation they are in and the skills of the person they are communicating with.

Various communication options include -:

### **Auditory – oral approaches**

Auditory-oral approaches aim to develop speaking and listening skills in deaf children. They emphasise the use of hearing aids, radio aids and cochlear implants to make the best use of any hearing a deaf child has (their 'residual hearing'). Most auditory-oral approaches will also use lipreading to help the child's understanding. Auditory-oral approaches do not use sign language or fingerspelling to support the understanding of spoken language.

#### **Lipreading**

Lipreading is the ability to read words from the lip patterns of the person speaking. Deaf children will naturally try to lipread when they are communicating.

It is difficult to measure how much of a conversation a deaf person understands just by relying on lipreading, as lip patterns vary from person to person. It is estimated that about 30% to 40% of speech sounds can be lipread under the best conditions. There are many things that can make lipreading difficult, for example:

- Beards and moustaches
- Talking while eating
- Covering your mouth while talking
- Poor lighting

When children are still building up an understanding of the language they may find it difficult to lipread words they are not familiar with. It also relies on a speaker having a clear lip pattern. An adult, who has a good understanding of the language being spoken, may understand more.

Lipreading can be used with other communication approaches such as fingerspelling and gestures. A child will also watch the facial expression and

body language of the speaker to get more clues. The combination of these things makes it possible to understand most of the conversation.

### **Sign bilingualism**

Sign bilingualism uses sign language as the child's first language. The spoken language of the family is learned as a second language. This can be taught through speech, writing or a sign-support system. The aim of using sign bilingualism is to allow the child to communicate in a way that doesn't depend on their hearing. British Sign Language (BSL) is a visual language that uses handshapes, facial expressions, gestures and body language to communicate. It has a structure and grammar different from that of written and spoken English. Because BSL is a totally visual language, being deaf does not affect a child's ability to learn the language. When a child has become confident in BSL, they can use this as a way to learn English. BSL will also give them access to other deaf people in the community.

It is estimated that over 70,000 people use BSL as their first or preferred language. In March 2003, the Government officially recognised BSL as a language.

### Fingerspelling

Fingerspelling supports sign language. It uses the hands to spell out English words. Each letter of the alphabet is indicated by using the fingers and palm of the hand. It is used for spelling names, places and for words that don't have an established BSL sign. For more details see [Fingerspelling](#).

### Sign Supported English

Sign Supported English (SSE) uses signs taken from British Sign Language. Signs are used in the same order as English words, but not every word that is spoken would be signed. Many hearing parents find this an easier way to become familiar with sign language as it means that they can use signs with their own language. As it uses the same signs as BSL, it can

be helpful if the child wants to develop BSL skills at a later stage. SSE is not a language in its own right. For more details see [Sign Supported English](#).

### Signed English

Signed English (SE) uses signs to represent English exactly by using a sign for every spoken word. It uses BSL signs, fingerspelling, and specifically developed signs to represent important grammatical information.

Signed English is not a language like BSL, but it has been designed as a teaching tool to be used at the same time as spoken English. Its aim is to develop reading and writing skills.

### Cued Speech

Cued Speech is a simple sound-based system that uses eight hand shapes in four different positions (cues), together with natural mouth movement of speech. Some words which sound different to hearing people can look very similar when they are lipread by deaf people (for example, 'pat' and 'bat'). Cued Speech is visual and the cues are placed near the mouth. This helps

to make every sound and word clear to a deaf child. It can be used together with sign language or to complement speech.

### **Total communication**

Total communication is based on using a combination of methods at the same time to communicate with a deaf child. The idea is to communicate and teach vocabulary and language in any way that works.

The child and their family are encouraged to use:

- A sign language system based on the English language, such as Sign Supported English (SSE); fingerspelling
- Natural gestures
- Lipreading
- Body language
- Speech and
- Hearing aids, cochlear implants and radio aids

### **Communication methods for deaf children with extra needs**

Children who have another physical or learning disability may have difficulty using some of the methods detailed above. The methods listed below are commonly used with children with extra needs.

#### Signalong

Signalong is a form of Sign Supported English. It is a relatively new signing system devised by professionals for children (and adults) who have language difficulties associated with learning disabilities and autism.

The signs are mostly based on BSL and are used in the same order as spoken English. It can be used with other languages too. Signalong is intended to support speech and is sometimes used with deaf children who have not developed speech but use some gestures. For more details see [Signalong](#).

#### Makaton

Makaton is a language programme that uses signs from British Sign Language together with unique Makaton symbols to provide basic communication to develop language and teach literacy skills. Grammatical signs are taken from signed English. Makaton is not a language but was designed as a tool for teaching children with severe communication and learning disabilities.

Makaton is made up of a main vocabulary of 450 concepts. It also has a larger resource vocabulary of approximately 7,000 concepts (for example

for animals, food, growth and development and many others). Concepts are visual images that are illustrated with signs and symbols. For more details see [Makaton](#).

Deafblind children

There are different communication approaches that a deafblind child may learn. These include British Sign Language (if a child has Usher's syndrome, for example, they might use visual frame BSL signing or hands on BSL signing to understand what someone is saying to them). They may also use Makaton, Signed English, Sign Supported English or one of the auditory-oral approaches. Deafblind children may also use the manual alphabet or a symbol system.

There are also written communication methods such as Braille or Moon, which use raised dots to indicate letters.

### **Communication support**

People who provide communication support (human aids to communication) include:-

#### **British Sign Language interpreters**

Sign language interpreting is a highly skilled profession that involves working in a variety of environments and situations, such as in schools, colleges, courts and theatres.

#### **Lipspeakers**

A lipspeaker is a person trained to accurately convey information from a speaker to a deaf lipreader using silent speech, clear lip patterns, facial expressions and gestures. They can be useful to deaf young people who use speech rather than sign to communicate.

Lipspeakers are often helpful in environments where there might be too much background noise, or perhaps where the person speaking does not have clear lip patterns. Deaf young people use lipspeakers in further or higher education, at job interviews or at meetings. Lipspeakers also work in the same settings as sign language interpreters.

#### **Communication support workers**

Communication support workers (CSWs) support deaf children and young people in schools and colleges, working closely with other professionals such as teachers and interpreters. A CSW is trained in communication

skills, and also in teaching methods and deaf-related issues. They work with children and young people with a range of communication needs.

### **Communicator guides and deafblind interpreters**

Communicator guides work with deaf-blind young people. They are trained in communication skills, the DeafBlind Manual alphabet, and have specialist skills in guiding deaf-blind people.

Deafblind interpreters interpret speech for deaf-blind young people, and also relay supporting information such as other people's reactions to what has been said, and people's movements around the room.

### **Notetakers**

Many deaf students in further and higher education have the support of a notetaker, as it is important that they have full notes, especially for revision purposes. Notetakers are expected to be skilled in taking notes, in handwritten English, to be directly passed onto students. They must also be aware of deaf issues.

Some notetakers also use laptop computers. The notes are stored on a disk to be given to the student at the end of the lecture.

### **Speech-to-text reporters**

A speech-to-text reporter provides a computerised word-for-word record of what is spoken. Text is viewed instantly on a monitor, laptop screen or on a projector screen. This is often used in meetings or at conferences.

Colleges can receive additional funding to meet the additional learning needs of deaf or hearing impaired students which may include providing interpreters or notetakers.

Disabled Students' Allowances can help pay the extra costs a student may incur to study a course of higher education, as a direct result of a disability. The allowances can help pay the cost of a non-medical personal helper such as an interpreter or notetaker.

### **Other professional support**

Teacher of deaf children

It is important that deaf children are taught by a person who understands their needs, regardless of their level of deafness. This makes sure that deaf children are given the same educational opportunities as hearing children of the same age.

A teacher who specialises in working with deaf children is commonly known as a teacher of the deaf. Teachers of the deaf require specialised skills.

They help deaf children to develop their language skills and they also play an important role in developing a deaf child's general education.

A teacher of the deaf works in a variety of different settings within the educational system. The four main areas in which a teacher of the deaf may work are a mainstream school; a special unit within a mainstream school; working in the home with the family to help develop the child's communication skills; or in a school for deaf children.

#### Speech and language therapist

A speech and language therapist works with children and adults who have difficulties producing and using speech sounds, difficulties using and understanding language, fluency (stammering) or problems with their voice. They may also work with people who have eating and swallowing difficulties.

The speech and language therapist assesses the nature of the client's problem and then provides appropriate treatment, advice and support. Speech and language therapy helps people to use their residual speech to the best of their ability. They also work with children who use sign language, to help them develop their auditory skills and speech.

Most speech and language therapists work in NHS hospitals or clinics, but some work in independent practice. Others may be employed by education authorities to work in schools with deaf children.

#### Educational Audiologist

Educational Audiologists work closely with deaf children, teachers and families and give advice about using hearing aids and about equipment and

services for deaf people. They may assess children's hearing and communication needs and fit hearing aids.

### **Environmental aids**

Technology has facilitated the development of "environmental aids" which help hearing impaired children to be safe and more independent. Below are some of the more important examples.

#### Alerting

- Alarm clocks. Children are woken up by clocks with flashing lights and / or vibrating pads under their pillow
- Doorbell alerting devices. Doorbells can be bought which are very loud, or they can be connected to the house lighting system, which is activated on and off, when the doorbell rings
- Smoke alarms and burglar alarms can also be connected to strobe flashing lights or vibrating pads, which are kept under the pillow
- Pagers can be connected by radio signals to telephone, doorbell, smoke alarms, baby alarm, burglar alarm etc; the pager usually vibrates and symbols or texts on the pager can indicate what is happening

#### Telephones

- Telephones can be adapted to have flashing lights, on ringing, a different ring tone frequency, or can be connected to house lights or a lamp (to flash) if needed. Telephones are available that amplify the sound and have volume and tone controls
- Mobile telephones can be set to vibrate or flash and communication in both directions may be by text messaging
- Loops or links to mobiles enable hearing aid users to use some models of mobile phones
- Videophones are used through the standard telephone network and each caller can see the other on a small video screen. These are rarely used at the present time because of problems with speed of transmission
- Text telephones (for severe or profound hearing loss). These are known as TDD (Telephone Devices for the Deaf) and are small keyboard telephones with a display screen, showing incoming and outgoing text. A text telephone is usually used at each end of the line, but in the situation where a text phone is not at the other end, a confidential relay system called



Typetalk is used – (a human operator converting text to talk and vice versa)

## Listening

- Subtitling and signing on TV. Subtitles (which can be turned on or off) are available for an increasing number of BBC TV programmes. The percentage is lower for other channels, and considerably so for cable and satellite channels. It is common for digital television and DVD's to contain subtitles that can be turned on and off now.
- Loop systems consist of a microphone, to pick up the sound source, a small amplifier and loop cable running around the edge of the room which sets up a magnetic field. This is picked up by the hearing aid when it is switched to the "T" or "MT" programme. Loop systems can be used at home and also in public places, such as banks, churches, cinemas and theatres.
- Hearing aids can also be used with a telephone, using the T programme, if the telephone is hearing aid compatible.
- FM Systems are commonly used by deaf children in schools and in other situations to enable them to hear the sound source directly through the hearing aid. The teacher wears a microphone and their voice is carried via radio waves to a small receiver plugged into the bottom of the hearing aid. The microphone can also be used plugged into any equipment that has a standard headphone socket such as computer, TV, MP3 player etc.

The following are not commonly used with children but may be available:

- Infra- red listening aids. These are an alternative to the magnetic loop systems. The sound is transmitted to an infrared transmitter which is beamed to a receiver unit, which could be in a neck loop (an alternative to looping the whole room) or headphones.
- Personal listening aids. A microphone is put on the TV or stereo or given to someone else to speak into and the output is then fed into a neck loop or headphones.
- Conversational listening aids are portable, small listening aids for conversations.

## **Sign Supported English (SSE)**

Sign language is not the only way deaf people can use gestures or make signs with their hands to communicate. Sign language - whichever one - is usually preferential to other forms of gesture-based communication systems

because it is a language as a whole. It has grammar, structure, syntax and rules. However, for a variety of reasons people may not want to or indeed be able to learn a whole language based around signing. In this case, other forms of manually coded language come in. The most popular of these in the UK at least is Sign Supported English.

### **What is Sign Supported English?**

As the name suggests, sign supported English relates to the English language. British Sign Language does not. In a sign language - British or otherwise, the structure of the language is unique to that language. It bears no resemblance to the main spoken language in the country in which it is used. Sign supported English, however, takes the signs from British sign language and uses them in the order that the words would be spoken in English. This means that a working knowledge of the signs for different words is needed in order to understand and use sign supported English, but the more complex grammar is not. This method of signing is also sometimes referred to as conceptually accurate signed language.

### **Who Can Use Sign Supported English?**

Anyone who knows plenty of signs can use sign supported English, so as long as they have a good vocabulary, they will be able to manage it. The more signs known the better, but people who only know a limited number of signs can often manage by signing the main words in a sentence and mouthing the others clearly. This of course, relies on the person being spoken to being able to lip read.

### **Other Similar Languages**

Systems of communication such as sign supported English cannot truly be called languages and so are known as manually coded English. There are many different variants throughout the English-speaking world. The one thing they all have in common is that they represent English using signs, rather than having their own language system. But in the USA and other countries, the relationship between various sign languages and American English gets complicated and boundaries blurred. For the purposes of speakers in the UK, sign supported English is the main variant.

Sign supported English may not be as satisfactory a language in its own right as British Sign Language but there can be no doubt that it fills a much-needed gap. For people who struggle with language learning, sign supported English removes this need and all the person must learn is new vocabulary. If the person you are signing to does not know English, it can be difficult to make them understand and so sign language would be better.

However, if they are familiar with English then sign supported English should be understood.

## **List of NHS hospitals with Ophthalmology departments**

### **England**

<b>Hospital Name</b>	<b>Hospital Address</b>
Addenbrooke's Hospital	Hills Road, Cambridge, Cambridgeshire, CB2 0QQ
Airedale General Hospital	Skipton Road, Steeton, Keighley, West Yorkshire, BD20 6TD
Alder Hey Hospital	Eaton Road, West Derby, Liverpool, Merseyside, L12 2AP
Alexandra Hospital	Woodrow Drive, Redditch, Worcestershire, B98 7UB
Altrincham General Hospital	Market Street, Altrincham, Cheshire, WA14 1PE
Andover War Memorial Hospital	Charlton Road, Andover, Hampshire, SP10 3LB
Arrowe Park Hospital	Arrowe Park Road, Upton, Wirral, Merseyside, CH49 5PE
Ashford Hospital	London Road, Ashford, Middlesex, TW15 3AA
Axminster Hospital	Chard Street, Axminster, Devon, EX13 5DU
Barnsley Hospital	Gawber Road, Barnsley, S75 2EP
Basingstoke and North Hampshire Hospital	Aldermaston Road, Basingstoke, Hampshire, RG24 9NA
Bassetlaw Hospital	Kilton Hill, Worksop, Nottinghamshire, S81 0BD
Bedford Hospital	South Wing, Kempston Road, Bedford, Bedfordshire, MK42 9DJ
Berkeley Hospital	Marybrook Street, Berkeley, Gloucestershire, GL13 9BL
Bexhill Hospital	Holliers Hill, Bexhill-on-Sea, East Sussex, TN40 2DZ
Bideford Hospital	Abbotsham Road, Bideford, Devon, EX39 3AG

Birch Hill Hospital	Union Road, Rochdale, Lancashire, OL12 9QB
Birmingham Midland Eye Centre (Bmec)	Dudley Road, Birmingham, West Midlands, B18 7QH
Bishop Auckland Hospital	Cockton Hill Road, Bishop Auckland, County Durham, DL14 6AD
Blackpool Victoria Hospital	Whinney Heys Road, Blackpool, Lancashire, FY3 8NR
Blandford Community Hospital	Milldown Road, Blandford Forum, Dorset, DT11 7DD
Bradford Royal Infirmary	Duckworth Lane, Bradford, West Yorkshire, BD9 6RJ
Bridlington and District Hospital	Bessingby Road, Bridlington, North Humberside, YO16 4QP
Bridport Community Hospital	Hospital Lane, Bridport, Dorset, DT6 5DR
Bristol Eye Hospital	Lower Maudlin Street, Bristol, Avon, BS1 2LX
Broomfield Hospital	Court Road, Chelmsford, Essex, CM1 7ET
Buckland Hospital	Coombe Valley Road, Dover, Kent, CT17 0HD
Burnley General Hospital	Casterton Avenue, Burnley, Lancashire, BB10 2PQ
Calderdale Royal Hospital	Salterhebble, Salterhebble, Halifax, West Yorkshire, HX3 0PW
Cannock Chase Hospital	Brunswick Road, Cannock, Staffordshire, WS11 5XY
Chase Farm Hospital	The Ridgeway, Enfield, Middlesex, EN2 8JL
Chelsea and Westminster Hospital	369 Fulham Road, London, Greater London, SW10 9NH
Cheltenham General Hospital	Sandford Road, Cheltenham, Gloucestershire, GL53 7AN

Chesterfield Royal Hospital	Calow, Chesterfield, Derbyshire, S44 5BL
Chippenham Hospital	Rowden Hill, Chippenham, Wiltshire, SN15 2AJ
Chorley and South Ribble Hospital	Preston Road, Chorley, Lancashire, PR7 1PP
Christchurch Hospital	Fairmile Road, Christchurch, Dorset, BH23 2JX
Cirencester Hospital	Tetbury Road, Cirencester, Gloucestershire, GL7 1UY
Clayton Hospital	Northgate, Wakefield, West Yorkshire, WF1 3JS
Conquest Hospital	The Ridge, St. Leonards-on-Sea, East Sussex, TN37 7RD
Countess Of Chester Hospital	The Countess Of Chester Health Park, Chester, Cheshire, CH2 1UL
County Hospital Louth	High Holme Road, Louth, Lincolnshire, LN11 0EU
Crawley Hospital	West Green Drive, Crawley, West Sussex, RH11 7DH
Cromer and District Hospital	Mill Road, Cromer, Norfolk, NR27 0BQ
Cumberland Infirmary	Newtown Road, Carlisle, Cumbria, CA2 7HY
Darent Valley Hospital	Darent Wood Road, Darent Wood Road, Dartford, Kent, DA2 8DA
Darlington Memorial Hospital	Hollyhurst Road, Darlington, County Durham, DL3 6HX
Delancey Hospital	Charlton Lane, Cheltenham, Gloucestershire, GL53 9DT
Derriford Hospital	Derriford Road, Crownhill, Plymouth, Devon, PL6 8DH
Dewsbury and District Hospital	Halifax Road, Dewsbury, West Yorkshire, WF13 4HS

Diana, Princess Of Wales Hospital	Scarcho Road, Grimsby, North East Lincolnshire, DN33 2BA
Doncaster Royal Infirmary	Armthorpe Road, Doncaster, South Yorkshire, DN2 5LT
Dorset County Hospital	Williams Avenue, Dorchester, Dorset, DT1 2JY
Ealing Hospital	Uxbridge Road, Southall, Middlesex, UB1 3HW
East Surrey Hospital	Canada Avenue, Redhill, Surrey, RH1 5RH
Eastbourne District General Hospital	Kings Drive, Eastbourne, East Sussex, BN21 2UD
Edenbridge War Memorial Hospital	Mill Hill, Edenbridge, Kent, TN8 5DA
Edgware Community Hospital	Burnt Oak Broadway, Edgware, Middlesex, HA8 0AD
Ellesmere Port Hospital	114 Chester Road, Whitby, Ellesmere Port, Merseyside, CH65 6SG
Epsom Hospital	Dorking Road, Epsom, Surrey, KT18 7EG
Essex County Hospital	Lexden Road, Colchester, Essex, CO3 3NB
Fairfield General Hospital	Rochdale Old Road, Bury, Lancashire, BL9 7TD
Fairford Hospital	The Croft, Fairford, Gloucestershire, GL7 4BB
Farnham Hospital	Hale Road, Farnham, Surrey, GU9 9QL
Finchley Memorial Hospital	Granville Road, Greater London, N12 0JE
Friarage Hospital	Northallerton, North Yorkshire, DL6 1JG
Frimley Park Hospital	Portsmouth Road, Frimley, Surrey, GU16 7UJ
Frome Victoria Hospital	Park Road, Frome, Somerset, BA11 1EY

Furness General Hospital	Dalton Lane, Barrow-In-Furness, Cumbria, LA14 4LF
George Eliot Hospital	College Street, Nuneaton, Warwickshire, CV10 7DJ
Gloucestershire Royal Hospital	Great Western Road, Gloucester, Gloucestershire, GL1 3NN
Good Hope Hospital	Rectory Road, Sutton Coldfield, West Midlands, B75 7RR
Goole and District Hospital	Woodland Avenue, Goole, North Humberside, DN14 6RX
Gosport War Memorial Hospital	Bury Road, Gosport, Hampshire, PO12 3PW
Grantham and District Hospital	101 Manthorpe Road, Manthorpe Road, Grantham, Lincolnshire, NG31 8DG
Halton General Hospital	Halton, Hospital Way, Runcorn, Cheshire, WA7 2DA
Haltwhistle War Memorial Hospital	Westgate, Haltwhistle, Northumberland, NE49 9AJ
Harrogate District Hospital	Lancaster Park Road, Harrogate, North Yorkshire, HG2 7SX
Heartlands Hospital	Bordesley Green East, Birmingham, West Midlands, B9 5SS
Hertford County Hospital	North Road, Hertford, Hertfordshire, SG14 1LP
Herts and Essex Community Hospital	Haymeads Lane, Bishop's Stortford, Hertfordshire, CM23 5JH
Hillingdon Hospital	Pield Heath Road, Uxbridge, Middlesex, UB8 3NN
Hinchingbrooke Hospital	Hinchingbrooke Park, Hinchingbrooke, Huntingdon, Cambridgeshire, PE29 6NT
Holsworthy Hospital	Dobles Lane, Holsworthy, Devon, EX22 6JQ



Homerton University Hospital	Homerton Row, London, Greater London, E9 6SR
Horsham Hospital	Hurst Road, Horsham, West Sussex, RH12 2DR
Horton General Hospital	Oxford Rd, Banbury, Oxfordshire, OX16 9AL
Hospital Of St Cross	Barby Rd, Rugby, Warwickshire, CV22 5PX
Huddersfield Royal Infirmary	Acre Street, Huddersfield, West Yorkshire, HD3 3EA
Hull Royal Infirmary	Anlaby Road, Anlaby Road, Hull, East Yorkshire, HU3 2JZ
Ilfracombe Hospital	St. Brannocks Park Road, Ilfracombe, Devon, EX34 8JF
James Paget University Hospital	Lowestoft Road, Gorleston, Great Yarmouth, Norfolk, NR31 6LA
John Coupland Hospital	Ropery Road, Gainsborough, Lincolnshire, DN21 2TJ
John Radcliffe Hospital	Headley Way, Headington, Oxford, Oxfordshire, OX3 9DU
<b>Hospital Name</b>	<b>Hospital Address</b>
Kent and Canterbury Hospital	Ethelbert Road, Canterbury, Kent, CT1 3NG
Kettering General Hospital	Rothwell Rd, Kettering, Northamptonshire, NN16 8UZ
Kidderminster General Hospital	Bewdley Road, Kidderminster, Worcestershire, DY11 6RJ
King Edward Vii Hospital	St Leonards Road, Windsor, Berkshire, SL4 3DP
King George Hospital	Barley Lane, Ilford, Essex, IG3 8YB
King's College Hospital (Denmark Hill)	Denmark Hill, London, Greater London, SE5 9RS

King's Mill Hospital	Mansfield Road, Sutton-In-Ashfield, Nottinghamshire, NG17 4JL
Kingston Hospital	Galsworthy Road, Kingston Upon Thames, Surrey, KT2 7QB
Leicester Royal Infirmary	Infirmery Square, Leicester, Leicestershire, LE1 5WW
Leigh Infirmary	The Avenue, Leigh, Lancashire, WN7 1HS
Leighton Hospital	Middlewich Road, Crewe, Cheshire, CW1 4QJ
Lincoln County Hospital	Greetwell Road, Lincoln, Lincolnshire, LN2 5QY
Lister Hospital	Coreys Mill Lane, Stevenage, Hertfordshire, SG1 4AB
Luton and Dunstable Hospital	Lewsey Road, Luton, Bedfordshire, LU4 0DZ
Lydney and District Hospital	Lydney, Gloucestershire, GL15 5JF
Maidstone Hospital	Hermitage Lane, Maidstone, Kent, ME16 9QQ
Malton Community Hospital	Malton, North Yorkshire, YO17 7NG
Manchester Royal Eye Hospital	Nelson Street, Manchester, Greater Manchester, M13 9WH
Manor Hospital	Moat Road, Walsall, West Midlands, WS2 9PS
Mary Hewetson Cottage Hospital (Keswick)	Crosthwaite Road, Keswick, Cumbria, CA12 5PH
Mayday University Hospital	530 London Road, Thornton Heath, Surrey, CR7 7YE
Medway Maritime Hospital	Windmill Road, Gillingham, Kent, ME7 5NY
Mile End Hospital	Bancroft Road, London, Greater London, E1 4DG

Millom Hospital	Lapstone Road, Millom, Cumbria, LA18 4BY
Milton Keynes Hospital	Standing Way, Eaglestone, Milton Keynes, Buckinghamshire, MK6 5LD
Montagu Hospital	Adwick Road, Mexborough, South Yorkshire, S64 0AZ
Moore Cottage Hospital	Moore Road, Bourton-on-The-Water, Cheltenham, Gloucestershire, GL54 2AZ
Moorfields Eye Hospital	162 City Road, London, Greater London, EC1V 2PD
Moreton-In-Marsh Hospital	Moreton-In-Marsh, Gloucestershire, GL56 0BS
Mount Vernon Hospital	Rickmansworth Road, Northwood, Middlesex, HA6 2RN
Musgrove Park Hospital	Musgrove Park, Parkfield Drive, Taunton, Somerset, TA1 5DA
New Cross Hospital	Wolverhampton Road, Heath Town, Wolverhampton, West Midlands, WV10 0QP
Newark Hospital	Boundary Road, Newark, Nottinghamshire, NG24 4DE
Newham General Hospital	Glen Road, Plaistow, London, E13 8SL
Norfolk and Norwich University Hospital	Colney Lane, Norwich, Norfolk, NR4 7UY
North Devon District Hospital	Raleigh Park, Barnstaple, Devon, EX31 4JB
North Middlesex University Hospital	Sterling Way, Greater London, N18 1QX
Northampton General Hospital	Cliftonville, Northampton, Northamptonshire, NN1 5BD
Northwick Park Hospital	Watford Road, Harrow, Middlesex, HA1 3UJ

Nottingham University Hospital - Queen's Medical Centre Campus	Derby Road, Nottingham, Nottinghamshire, NG7 2UH
Nuffield Diagnostic Centre	Cottingham Rd, Corby, Northamptonshire, NN17 2UW
Ormskirk and District General Hospital	Wigan Road, Ormskirk, Lancashire, L39 2AZ
Palmer Community Hospital	Wear Street, Jarrow, Tyne and Wear, NE32 3UX
Paulton Memorial Hospital	Salisbury Road, Paulton, Bristol, Avon, BS39 7SB
Pembury Hospital	Tonbridge Road, Pembury, Tunbridge Wells, Kent, TN2 4QJ
Penrith Hospital	Bridge Lane, Penrith, Cumbria, CA11 8HX
Peterborough District Hospital	Thorpe Road, Peterborough, Cambridgeshire, PE3 6DA
Pilgrim Hospital	Sibsey Road, Boston, Lincolnshire, PE21 9QS
Pinderfields General Hospital	Aberford Road, Wakefield, West Yorkshire, WF1 4DG
Pontefract General Infirmary	Friarwood Lane, Pontefract, West Yorkshire, WF8 1PL
Poole General Hospital	Longfleet Road, Poole, Dorset, BH15 2JB
Potters Bar Community Hospital	Barnet Road, Potters Bar, Hertfordshire, EN6 2RY
Princess Alexandra Hospital	Hamstel Road, Harlow, Essex, CM20 1QX
Princess Royal Hospital	Lewes Road, Haywards Heath, West Sussex, RH16 4EX
Princess Royal University Hospital	Farnborough Common, Orpington, Kent, BR6 8ND
Purley War Memorial Hospital	856 Brighton Road, Purley, Surrey, CR8 2YL

Queen Alexandra Hospital	Southwick Hill Road, Cosham, Portsmouth, Hampshire, PO6 3LY
Queen Elizabeth II Hospital	Howlands, Welwyn Garden City, Hertfordshire, AL7 4HQ
Queen Elizabeth The Queen Mother Hospital	St Peters Road, Margate, Kent, CT9 4AN
Queen Mary's Hospital Sidcup	Frognal Avenue, Sidcup, Kent, DA14 6LT
Queen Victoria Hospital	Thornton Rd, Morecambe, Lancashire, LA4 5NN
Queen Victoria Hospital (East Grinstead)	Holtye Road, East Grinstead, West Sussex, RH19 3DZ
Queen's Hospital	Rom Valley Way, Romford, Essex, RM7 0AG
Queen's Hospital, Burton Upon Trent	Belvedere Road, Burton -on- Trent, Staffordshire, DE13 0RB
Ripon and District Community Hospital	Firby Lane, Ripon, North Yorkshire, HG4 2PR
Rossendale Hospital	Haslingden Road, Rossendale, Lancashire, BB4 6NE
Rotherham Hospital	Moorgate Road, Rotherham, S60 2UD
Rowley Regis Hospital	Moor Lane, Rowley Regis, West Midlands, B65 8DA
Royal Albert Edward Infirmary	Wigan Lane, Wigan, Lancashire, WN1 2NN
Royal Berkshire Hospital	London Road, Reading, Berkshire, RG1 5AN
Royal Blackburn Hospital	Haslingden Road, Blackburn, Lancashire, BB2 3HH
Royal Bolton Hospital	Minerva Road, Farnworth, Bolton, Lancashire, BL4 0JR
Royal Bournemouth General Hospital	Castle Lane East, Bournemouth, Dorset, BH7 7DW

Royal Cornwall Hospital (Treliske)	Treliske, Truro, Cornwall, TR1 3LJ
Royal Derby Hospital	Uttoxeter Road, Derby, Derbyshire, DE22 3NE
Royal Devon and Exeter Hospital (Wonford)	Barrack Road, Exeter, Devon, EX2 5DW
Royal Eye Infirmary	Apsley Road, Plymouth, Devon, PL4 6PL
Royal Free Hospital	Pond Street, Greater London, NW3 2QG
Royal Hallamshire Hospital	Glossop Road, Sheffield, South Yorkshire, S10 2JF
Royal Hampshire County Hospital	Romsey Road, Winchester, Hampshire, SO22 5DG
Royal Lancaster Infirmary	Ashton Road, Lancaster, Lancashire, LA1 4RP
Royal London Homeopathic Hospital	60 Great Ormond Street, London, Greater London, WC1N 3HR
Royal Oldham Hospital	Rochdale Road, Oldham, Lancashire, OL1 2JH
Royal Preston Hospital	Sharoe Green Lane North, Fulwood, Preston, Lancashire, PR2 9HT
Royal Shrewsbury Hospital	Mytton Oak Road, Shrewsbury, Shropshire, SY3 8XQ
Royal Surrey County Hospital	Egerton Road, Guildford, Surrey, GU2 7XX
Royal United Hospital	Combe Park, Bath, Avon, BA1 3NG
Royal Victoria Hospital (Folkestone)	Radnor Park Avenue, Folkestone, Kent, CT19 5BN
Russells Hall Hospital	Pensnett Road, Dudley, West Midlands, DY1 2HQ
<b>Hospital Name</b>	<b>Hospital Address</b>

Saffron Walden Community Hospital	Radwinter Road, Saffron Walden, Essex, CB11 3HY
Salford Royal	Stott Lane, Salford, M6 8HD
Salisbury District Hospital	Odstock Road, Salisbury, Wiltshire, SP2 8BJ
Sandwell General Hospital	Lyndon, West Bromwich, West Midlands, B71 4HJ
Savernake Hospital	General Hospital, Marlborough, Wiltshire, SN8 3HL
Scarborough General Hospital	Woodlands Drive, Scarborough, North Yorkshire, YO12 6QL
Scunthorpe General Hospital	Cliff Gardens, Scunthorpe, North East Lincolnshire, DN15 7BH
Selby and District War Memorial Hospital	Doncaster Road, Selby, North Yorkshire, YO8 9BX
Selly Oak Hospital	Raddlebarn Road, Selly Oak, Birmingham, B29 6JD
Sevenoaks Hospital	Hospital Road, Sevenoaks, Kent, TN13 3PG
Sheffield Children's Hospital	Western Bank, Sheffield, South Yorkshire, S10 2TH
Shepton Mallet Community Hospital	Old Wells Road, Shepton Mallet, Somerset, BA4 4PG
Sittingbourne Memorial Hospital	Bell Road, Sittingbourne, Kent, ME10 4DT
Skegness and District General Hospital	Dorothy Avenue, Skegness, Lincolnshire, PE25 2BS
Skipton General Hospital	Keighley Road, Skipton, North Yorkshire, BD23 2RJ
Solihull Hospital	Lode Lane, Solihull, West Midlands, B91 2JL
South Molton Hospital	Widgery Drive, South Molton, Devon, EX36 4DP

South Tyneside District Hospital	Harton Lane, South Shields, Tyne and Wear, NE34 0PL
Southampton General Hospital	Tremona Road, Southampton, Hampshire, SO16 6YD
Southend Hospital	Prittlewell Chase, Westcliff-on-Sea, Essex, SS0 0RY
Southport and Formby District General Hospital	Town Lane, Southport, Merseyside, PR8 6PN
St Albans City Hospital	Waverley Road, St. Albans, Hertfordshire, AL3 5PN
St Ann's Hospital	St. Ann's Road, London, Greater London, N15 3TH
St Bartholomew's Hospital	West Smithfield, London, Greater London, EC1A 7BE
St George's Hospital	Blackshaw Road, London, Greater London, SW17 0QT
St Helens Hospital	Marshall's Cross Road, St. Helens, Merseyside, WA9 3DA
St James's University Hospital	Beckett Street, Leeds, West Yorkshire, LS9 7TF
St Peter's Hospital	Guildford Road, Chertsey, Surrey, KT16 0PZ
St Peter's Hospital	Spital Road, Maldon, Essex, CM9 6EG
St Thomas' Hospital	Westminster Bridge Road, London, Greater London, SE1 7EH
St. Margaret's Hospital	The Plain, Epping, Essex, CM16 6TN
Stafford Hospital	Weston Road, Stafford, Staffordshire, ST16 3SA
Stamford and Rutland Hospital	Ryhall Road, Stamford, Lincolnshire, PE9 1UA
Stepping Hill Hospital	Poplar Grove, Stockport, Cheshire, SK2 7JE



Stoke Mandeville Hospital	Mandeville Road, Aylesbury, Buckinghamshire, HP21 8AL
Stratford Hospital	Arden Street, Stratford-upon-Avon, Warwickshire, CV37 6NX
Stretford Memorial Hospital	226 Seymour Grove, Stretford, Manchester, Greater Manchester, M16 0DU
Stroud General Hospital	Trinity Road, Stroud, Gloucestershire, GL5 2HY
Sunderland Eye Infirmary	Queen Alexandra Road, Sunderland, Tyne and Wear, SR2 9HP
Sussex Eye Hospital	Eastern Road, Brighton, East Sussex, BN2 5BF
Sutton Hospital	Cotswold Road, Sutton, Surrey, SM2 5NF
Tameside General Hospital	Fountain St, Ashton Under Lyne, Lancashire, OL6 9RW
Tewkesbury General Hospital	Barton Road, Tewkesbury, Gloucestershire, GL20 5QN
The County Hospital	The County Hospital, Union Walk, Hereford, Herefordshire, HR1 2ER
The Great Western Hospital	Marlborough Road, Marlborough Road, Swindon, Wiltshire, SN3 6BB
The Ipswich Hospital	Heath Road, Ipswich, Suffolk, IP4 5PD
The James Cook University Hospital	Marton Road, Middlesbrough, Cleveland, TS4 3BW
The Johnson Community Hospital	Spalding Road, Pinchbeck, Spalding, Lincolnshire, PE11 3PB
The Princess Royal Hospital	Apley Castle, Grainger Drive, Telford, Shropshire, TF1 6TF
The Queen Elizabeth Hospital	Gayton Road, King's Lynn, Norfolk, PE30 4ET
The Royal Liverpool University Hospital	Prescot Street, Liverpool, Merseyside, L7 8XP

The Royal London Hospital	Whitechapel, London, Greater London, E1 1BB
The Royal Marsden Hospital (London)	Fulham Road, London, Greater London, SW3 6JJ
The Royal Victoria Infirmary	Queen Victoria Road, Newcastle Upon Tyne, Tyne and Wear, NE1 4LP
The Whittington Hospital	Magdala Avenue, London, Greater London, N19 5NF
Tiverton and District Hospital	Kennedy Way, Tiverton, Devon, EX16 6NT
Torbay Hospital	Newton Road, Torquay, Devon, TQ2 7AA
Torrington Hospital	Calf Street, Torrington, Devon, EX38 7BJ
Trowbridge Hospital	Adcroft Street, Trowbridge, Wiltshire, BA14 8PH
University College Hospital	235 Euston Road, London, Greater London, NW1 2BU
University Hospital (Coventry)	Clifford Bridge Road, Coventry, West Midlands, CV2 2DX
University Hospital Of North Durham	North Road, Durham, County Durham, DH1 5TW
University Hospital Of North Staffs	Hartshill Road, Stoke-on-Trent, Staffordshire, ST4 7PA
University Hospital Of North Tees	Hardwick Road, Stockton-on-Tees, Cleveland, TS19 8PE
Upney Lane Centre	132 Upney Lane, Barking, Essex, IG11 9LX
Victoria Hospital (Deal)	London Road, Deal, Kent, CT14 9UA
Victoria Hospital (Sidmouth)	Sidmouth, Devon, EX10 8EW
Victoria Infirmary (Northwich)	Winnington Hill, Northwich, Cheshire, CW8 1AW
Walton Hospital	Rice Lane, Liverpool, Merseyside, L9 1AE

Warrington Hospital	Lovely Lane, Warrington, Cheshire, WA5 1QG
Warwick Hospital	Lakin Road, Warwick, Warwickshire, CV34 5BW
Watford General Hospital	60 Vicarage Road, Watford, Hertfordshire, WD18 0HB
Watford General Hospital	Vicarage Road, Watford, Hertfordshire, WD18 0HB
West Berkshire Community Hospital	London Road, Benham Hill, Thatcham, Berkshire, RG18 3AS
West Cornwall Hospital (Penzance)	St Clare Street, Penzance, Cornwall, TR18 2PF
West Cumberland Hospital	Homewood, Hensingham, Whitehaven, Cumbria, CA28 8JG
West Suffolk Hospital	Hardwick Lane, Bury St Edmunds, Suffolk, IP33 2QZ
Westmorland General Hospital	Burton Rd, Kendal, Cumbria, LA9 7RG
Weston General Hospital	Grange Road, Uphill, Weston-Super-Mare, BS23 4TQ
Wharfedale Hospital	Newall Carr Road, Otley, West Yorkshire, LS21 2LY
Whipps Cross University Hospital	Whipps Cross Road, London, Greater London, E11 1NR
Whitby Community Hospital	Springhill, Whitby, North Yorkshire, YO21 1EE
Whitstable and Tankerton Hospital	174-176 Northwood Road, Whitstable, Kent, CT5 2HN
Whitworth Hospital	330 Bakewell Road, Darley Dale, Matlock, Derbyshire, DE4 2JD
William Harvey Hospital (Ashford)	Kennington Road, Willesborough, Ashford, Kent, TN24 0LZ

Worcestershire Royal Hospital	Charles Hastings Way, Worcester, Worcestershire, WR5 1DD
Workington Community Hospital	Park Lane, Workington, Cumbria, CA14 2RW
Wycombe Hospital	Queen Alexandra Road, High Wycombe, Buckinghamshire, HP11 2TT
Yeatman Hospital	Yeatman Hospital, Hospital Lane, Sher- borne, Dorset, DT9 3JU
Yeovil District Hospital	Higher Kingston, Yeovil, Somerset, BA21 4AT
York Hospital	Wigginton Road, York, North Yorkshire, YO31 8HE

## Scotland

<b>Hospital Name</b>	<b>Hospital Address</b>
Abbey Carrick Glen Hospital	Dalmellington Road, Ayr, KA6 6PG
Aberdeen City Hospital	Urquhart Road, Aberdeen, AB24 5AU
Aberdeen Royal Infirmary	Foresterhill, Aberdeen, AB25 2ZN
The Ayr Hospital	Dalmellington Road, Ayr, KA6 6DX
Balfour Hospital	New Scapa Road, Kirkwall, Orkney, KW15 1BH
Borders General Hospital	Melrose, TD6 9BS
City Health Clinic	9 Earl Grey Street, Edinburgh, EH3 9BN
Crosshouse Hospital	Crosshouse, Kilmarnock, KA2 0BE
Dr Gray's Hospital	Pluscarden Road, Elgin, IV30 1SN
Dumfries and Galloway Royal Infirmary	Bankend Road, Dumfries, DG1 4AP
Dalrymple Hospital	Stranraer, DG9 7DQ
Falkirk and District Royal Infirmary	Major's Loan, Falkirk, FK1 5QE
Gartnavel General Hospital	1053 Great Western Road, Glasgow, G12 0YN
Golden Jubilee National Hospital	Beardmore Street, Clydebank, Glasgow, G81 4HX
Glasgow Royal Infirmary	84 Castle Street, Glasgow, G4 0SF
Hairmyres Hospital	Eaglesham Road, East Kilbride, G75 8RG
Hay Lodge Hospital	Neidpath Road, Peebles, EH45 8JG
Inverclyde Royal Hospital	Larkfield Road, Greenock, PA16 0XN
Jubilee Hospital	Bleachfield Street, Huntly, AB54 8EX
Monklands Hospital	Monkscourt Avenue, Airdrie, ML6 0JS
Montrose Royal Infirmary	Montrose, DD10 8AJ
The New Victoria Hospital	Grange Road, Glasgow, G42 9LF
Ninewells Hospital	Dundee, DD1 9SY

Peterhead Community Hospital	21 Links Terrace, Peterhead, AB42 2XB
Princess Alexandra Eye Pavilion	Chalmers Street, Edinburgh, EH3 9HA
Perth Royal Infirmary	Perth, PH1 1NX
Queen Margaret Hospital	Whitefield Road, Dunfermline, KY12 0SU
Raigmore Hospital	Old Perth Road, Inverness, IV2 3UJ
Royal Aberdeen Children's Hospital	Westburn Road, Foresterhill, Aberdeen, AB25 2ZG
Royal Hospital for Sick Children	Yorkhill, Glasgow, G3 8SJ
Royal Alexandra Hospital	Corsebar Road, Paisley, PA2 9PN
Royal Hospital for Sick Children	9 Sciennes Road, Edinburgh, EH9 1LF
Stirling Royal Infirmary	Livilands, Stirling, FK8 2AU
Southern General Hospital	Glasgow, G51 4TF
Spire Murrayfield Hospital	122 Corstophine Road, Edinburgh, EH12 6UD
St John's Hospital at Howden	Howden, Livingston, EH54 6PP
Stobhill ACH Hospital	133 Balornock Road, Glasgow, G21 3UW
Turner Memorial Hospital	Turner Street, Keith, AB55 5DJ
Uist and Barra Hospital	Balivanich, Benbecula, HS7 5LA
Victoria Hospital	Hayfield Road, Kirkcaldy, KY2 5AH
Western Isles Hospital	Macaulay Road, Stornoway, Isle of Lewis, HS1 2AF
Western Infirmary	Dumbarton Road, Glasgow, G11 6NT
Wishaw General Hospital	50 Netherton Street, Wishaw, ML2 0DP

## Wales

<b>Hospital Name</b>	<b>Hospital Address</b>
Bridgend Clinic, Princes of Wales Hospital	Coity Road, Bridgend, CF31 1RQ
Bro Ddyfi Community Hospital	Machynlleth, SY20 8AD
Bronglais General Hospital	Aberystwyth, SY23 1ER
Bryn Beryl Hospital	Pwllheli, LL53 6TT
Colwyn Bay Community Hospital	Hesketh Road, Colwyn Bay, LL29 8AY
Dolgellua & Barmouth District Hospital	Dolgellau, LL40 1NP
Ffestiniog Memorial Hospital	Blaenau, Ffestiniog, LL41 3DW
Glan Clwyd Hospital	Bodelwyddan, Rhyl, LL18 5UJ
H M Stanley Hospital	St Asaph, LL17 0RS
Llandrindod Wells Hospital	Temple Street, Llandrindon, LD1 5HF
Llandudno General Hospital	Llandudno, LL30 1LB
Longfield Medical Consulting Rooms	196 Stow Hill, Newport, Gwent, NP20 4HB
MDA Clinic	65 Cardiff Road, Llandaff, Cardiff, CF5 2AA
Neath Port Talbot Hospital	Baglan Way, Port Talbot, SA12 7BX
Nevill Hall Hospital	Brecon Road, Abergavenny, NP7 7EG
Prince Charles General Hospital	Merthyr Tydfil, CF47 9DT
Prince Philip Hospital	Bryngynmawr, Dafen, Llanelli, SA14 8QF
Royal Glamorgan Hospital	Ynysmaerdy, Llantrisant, CF72 8XR
Royal Gwent Hospital	Cardiff Road, Newport, Gwent, NP20 2UB
Sancta Maria Hospital	Ffynone Road, Swansea, SA1 6DF
Singleton Hospital	Sketty Lane, Swansea, SA2 8QA
South Pembrokeshire Hospital	Fort Road, Pembroke Dock, SA72 6SX

Spire Cardiff Hospital	Croescadarn Road, Pentwyn, Cardiff, CF23 8XL
Spire Yale Hospital	Wrexham Technology Park, Croesnewydd Road, Wrexham, LL13 7YP
St Asaph Consulting Rooms, Yale Hospital	St Asaph rooms, 88 Bowen Court, St Asaph, LL17 0JE
St Joseph's Hospital	Harding Avenue, Malpas, Newport, Gwent, NP20 6ZE
St Tysilio Medical Centre	Llanfairpwll, Anglesey, LL61 5YR
Tenby Cottage Hospital	Church Park, Tenby, SA70 7EE
Twywn Hospital	Bryn Hyfryd Road, Twywn, LL36 9HH
University Hospital of Wales	Heath Park, Cardiff, CF14 4XW
Vale Healthcare Cardiff By Clinic	Dunleavy Drive, Cardiff Bay, Cardiff, CF11 0SN
Victoria War Memorial Hospital	Salop Road, Welshpool, SY21 7DU
West Wales General Hospital	Glangwili, Carmarthen, SA31 2AF
Withybush General Hospital	Fishguard Road, Haverfordwest, SA61 2PZ
Wrexham Maelor Hospital	Croesnewydd Road, Wrexham, LL13 7TD
Ysbyty Gwynedd	Penrhosgarnedd, Bangor, Gwynedd, LL57 2PW
Ysbyty Penrhos Stanley	Holyhead, LL65 2PR
Ystrad Mynach Hospital	Caerphilly Road, Ystrad Mynach, Hengoed, CF82 7XU
Ystradgynlais Community Hospital	Glanrhyd Road, Ystradgynlais, Swansea, SA9 1AE

## What you need to know about Hydrocephalus



### **What is Hydrocephalus?**

Hydrocephalus is excessive accumulation of cerebrospinal fluid (CSF) in the brain .....

- [Hydrocephalus](#)

### **What are the effects and signs?**

The clinical features vary depending upon the age of the child.....

- [Effects of Hydrocephalus](#)

### **How is it assessed?**

Diagnosis of congenital hydrocephalus may be made by routine prenatal ultrasound .....

- [Assessment of Hydrocephalus](#)

### **How is it treated and managed?**

Treatment depends upon the underlying cause of the hydrocephalus and in infancy, the weight of the child .....

- [Treatment for Hydrocephalus](#)

### **What evidence is available?**

The Occupational therapist / Physiotherapist will be able to provide details of symptoms, investigations and treatment / management.....

- [Evidence](#)

### **How long will the needs last?**

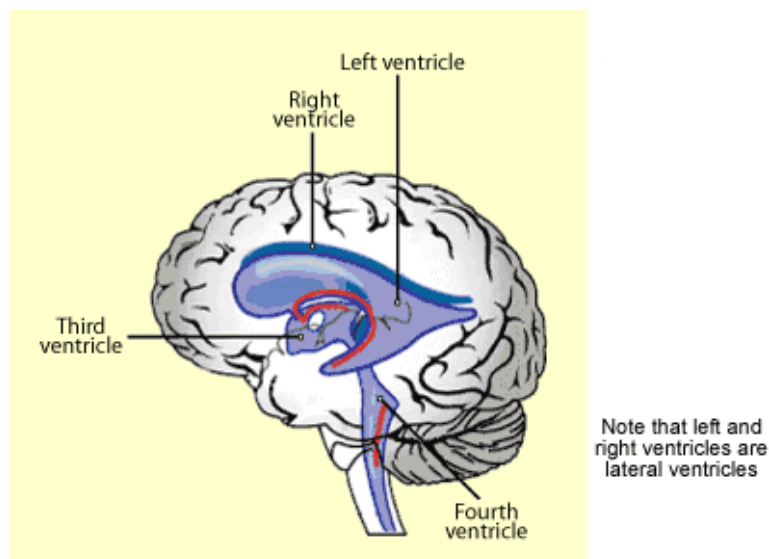
- [Prognosis and duration of the award](#)

### **What is Hydrocephalus?**

Hydrocephalus is excessive accumulation of cerebrospinal fluid (CSF) in the brain.

Cerebrospinal fluid is mainly produced in chambers (ventricles) in the brain. It flows through the ventricular system of the brain, bathes the surface of

the brain and spinal cord and is subsequently absorbed into the blood stream.



Cerebrospinal fluid has a number of functions that include support and protection of the brain and delivery of nutrients and removal of waste from the brain.

Hydrocephalus results if there is any interruption to the normal flow of CSF. The resulting dilatation (expansion) of the ventricles may lead to compression and damage of brain tissue.

Hydrocephalus can be non – communicating (obstructive) or communicating (non – obstructive). Non – communicating hydrocephalus results from an obstruction within the ventricular system. Communicating hydrocephalus results from a failure of absorption of CSF back into the blood stream. However, there are some causes of hydrocephalus that do not fit precisely into these categories.

#### Incidence and Prevalence

Hydrocephalus occurs in approximately 1 in 1000 live births worldwide. Significant variations are seen between different countries with the highest

incidence occurring in middle and low income countries. In high income countries such as the UK, the incidence is of the order of 0.8 per 1000 live births.

- [Causes](#)

### **What are the effects and signs?**

The clinical features vary depending upon the age of the child and the severity of the hydrocephalus.

In infants, the initial symptoms and signs include an unusually large head or increasing head circumference, a bulging fontanelle (a gap in the bones of the skull which usually fuse together during infancy) poor feeding, vomiting, drowsiness, irritability and fits. A late sign is downward deviation of the eyes known as “sunsetting.”

In older children, the initial symptoms and signs include headache, nausea and vomiting, lethargy, drowsiness, irritability, changes in personality and cognition including memory loss, blurred and double vision, problems with balance, coordination and gait, urinary incontinence, developmental delay and precocious (early) puberty.

Long term problems associated with hydrocephalus include -:

- [Learning disability](#) (low IQ <70)
- Cognitive problems including difficulties with learning, memory and executive function (the ability to organise, set priorities, manage time and make decisions)
- [Speech and language problems](#)
- Behavioural problems
- [Autism](#)
- [ADHD](#)
- [Cerebral palsy](#)
- Other motor deficits (delay or deficiency in attaining motor milestones), for example difficulty walking and problems with gross and fine motor control and balance. Motor deficits may worsen after a certain age due to factors such as the effect of growth and ageing and the effect on the musculoskeletal

system. The age at which this deterioration occurs is very variable.

- [Epilepsy](#)
- [Visual problems](#) and squint
- [Hearing problems](#)
- Shunt related problems

Mechanical - either obstruction or fracture/destruction of the shunt tube

Infection - usually occurring within six months of insertion

Over drainage - which can result in sub-dural haematomas (blood clots under the skull overlying the brain due to tearing of blood vessels) or collapse of the ventricles (slit ventricles) resulting in obstruction

“Shuntalgia” - discomfort at the shunt valve or along the route of the catheter in the neck.

Accumulation of cerebrospinal fluid (CSF) at the site of drainage, for example abdominal swelling or hydroceles (fluid surrounding the testicles usually only in infants less than 12 months of age) in ventriculoperitoneal shunts.

### **Children with myelomeningocele and hydrocephalus**

Children with myelomeningocele and hydrocephalus have similar clinical features to those with hydrocephalus alone. However, children with myelomeningocele and hydrocephalus are less likely to have learning disability and epilepsy and are extremely unlikely to have cerebral palsy. Children with myelomeningocele are more likely to have a severe functional loss with regard to mobility and physical dependence and are likely to have a problem with social integration as well.

### **Factors associated with more severe disability include -:**

- Gestational age  
Term infants are those born after 36 weeks of gestation (pregnancy). Preterm infants are those born between 32 and 36 weeks of gestation. Very pre-term infants are those born before 32 weeks of gestation

There is a correlation between lower gestational age at birth and the number of neuro-impairments (learning disability, cerebral palsy or epilepsy)

present. There is also a correlation with poor motor function and higher total handicap scores.

- **Cause of hydrocephalus**  
Associated impairments are more likely in children in whom the cause of their hydrocephalus is perinatal or postnatal rather than prenatal. Children who develop hydrocephalus after intraventricular haemorrhage or infection are much more likely to develop cognitive problems than those who have a prenatal cause for their hydrocephalus
- **Number of shunt failures**  
There is a correlation between the number of shunt failures and poor fine motor control and the number of neuro-impairments
- **Ventricular size at follow up**  
Additional neuro-impairments, especially learning disability, are more likely in children who have enlarged or collapsed (slit) ventricles at follow up. Slit ventricle syndrome usually only occurs if the shunt has been in place for more than 5 years

Note: Being born very preterm and hydrocephalus that is obvious at birth are the factors most likely to predict a poor outcome.

### **How is it assessed?**

Diagnosis of congenital hydrocephalus may be made by routine prenatal ultrasound. After birth, clinical suspicion may be confirmed by CT scan, MRI scan or ultrasound. CT scan, MRI scan or ultrasound may be used to follow progression once the diagnosis has been made.

### **How is it treated and managed?**

Treatment depends upon the underlying cause of the hydrocephalus and in infancy, the weight of the child.

Treatment of the hydrocephalus is the first priority. However, very often, treatment of the underlying cause must be carried out at the same time or very soon afterwards.

Most children with hydrocephalus are treated by surgical insertion of a shunt to divert cerebrospinal fluid (CSF) from the lateral ventricle to another part of the body, where it can be absorbed thereby reducing pressure in the brain. This procedure is performed in the operating room by a neurosurgeon. A flap is cut in the scalp and a small hole is drilled in the skull. A small silastic catheter is passed into a ventricle of the brain. A one way valve which controls flow of CSF is attached to the catheter. Another catheter is attached to the valve and tunnelled under the skin, behind the ear, down the

neck and chest and usually into the peritoneal (abdominal) cavity (ventriculo-peritoneal shunt). Less often used alternative sites to the peritoneal cavity include the right atrium of the heart via neck veins or the pleural space surrounding the lungs.

Some children can be treated by an alternative procedure called endoscopic third ventriculostomy (ETV) in which a neuroendoscope is used to make a small hole in the floor of the third ventricle so that CSF can drain towards the re-absorption sites of the brain.

Another procedure that can be performed is External Ventricular Drainage (EVD), which is the temporary drainage of CSF from the lateral ventricles to a closed collection system outside the body.

For children less than 2kg in weight, shunting tends to be deferred because of the high rate of shunt failure, usually secondary to infection. Temporary procedures, such as serial lumbar punctures, serial ventricular taps, EVD or an Ommaya reservoir can be used until the child is old enough for a permanent procedure to be carried out.

### **Shunt care**

Parents and older children need to be taught about the signs and symptoms of shunt failure. They will be provided with printed information about the shunt and a management plan about who to contact and when to go to hospital. Children with shunts should be encouraged to live as normal a life as possible. However, there are some special precautions that apply. These include avoiding some contact sports like boxing that may cause injury to the shunt valve or head injury and avoiding wearing bags on the side of the body where the shunt tubing passes down the side of the neck.

If a child has a programmable shunt, the shunt can be adversely affected by magnetic fields. Parents and children are advised not to place or play with magnets near the shunt and the shunt may require reprogramming before the child undergoes certain procedures, for example MRI scanning.

### **Shunt related problems**

These include -:

- Mechanical, either obstruction or fracture/destruction of the shunt tube
- Infection which usually occurs within six months of insertion
- Acute over drainage which can result in sub-dural haematomas (blood clots under the skull overlying the brain due to tearing of blood vessels) or chronic over drainage which can

result in collapse of the ventricles (slit ventricles) resulting in obstruction

- “Shuntalgia” which is discomfort at the shunt valve or along the route of the catheter in the neck

Accumulation of CSF at the site of deposition, for example abdominal swelling or hydroceles (fluid surrounding the testicles) in ventriculoperitoneal shunts

### **Shunt revision**

About 6 of every 10 children require shunt revision or replacement at some time in their lives because of shunt failure. Shunt failure is most likely in the

first year after insertion. Causes of shunt failure include obstruction, infection and disconnection, breakage or displacement of components.

### **Aids and adaptations**

There are no aids or adaptations specific to the management of hydrocephalus. However, various aids and adaptations may be used for the associated disabling conditions.

### **What evidence is available?**

<b>Sources of evidence</b>	<b>Information Provided</b>	<b>Limitation of Evidence</b>
Parent / carer	First source of information and associated evidence.	Does not have specialist knowledge.
Paediatrician / neurosurgeon / specialist nurse	Symptoms, investigations and treatment / management, and likely to have information about resulting disability or needs.	None.
Occupational therapist / Physiotherapist	Symptoms, investigations and treatment / management, and likely to have information about resulting disability or needs.	None.
GP	Symptoms, investigations and treatment / management.	Does not have specialist knowledge.  Unlikely to have information about resulting disability or needs.

### **How long will the needs last?**



<b>Age at date of claim</b>	<b>Award Period</b>
1 - 7	Award to age 8 (or for 1 year, whichever is the longer)
8 - 15	Award to age 16 (or for 1 year, whichever is the longer)

Renewal at age 8 is suggested as the disability associated with hydrocephalus may improve as a consequence of surgical, medical, physiotherapy, occupational therapy, psychosocial and educational interventions.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Causes of hydrocephalus**

The causes of hydrocephalus may be classified according to whether they are -:

#### **Prenatal**

This means the cause occurred before the onset of labour. The most common causes are malformations. These include:

**Chiari malformations** (formerly called Arnold – Chiari malformations) which are problems with the base of the skull and the hole in the base of the skull (foramen magnum) that the spinal cord exits through into the neck. There are two types, which occur when the cerebellum (or hind brain which is responsible for coordination and balance) and brain stem are forced through the foramen magnum. Type 2 is associated with spina bifida. Chiari malformations are associated with hydrocephalus because there may be obstruction to the flow of cerebrospinal fluid (CSF) by the malformation. Symptoms from the malformation can present in childhood but most often present in adult life.

Depending upon the age of the child these may include characteristic headaches (worse on coughing, straining etc), sensory disturbance, visual disturbance with double vision or nystagmus (uncontrollable jerking movements of the eyes), swallowing problems, alteration in speech, poor coordination on walking, choking, breath holding attacks, brief recurrent respiratory arrests (apnoeic episodes), and opisthotonos (holding the head forced backwards).

**Dandy Walker syndrome** is a rare condition in which there is an abnormality of the fourth ventricle and the CSF spaces around the cerebellum and under-development of the cerebellum. About 90% of affected children have hydrocephalus. About 70% of affected children have associated

abnormalities of the central nervous system (CNS). For example, 25% have underdevelopment of the corpus callosum (nerve tracts that connect the two sides of the brain). About one third of children have non – CNS malformations, for example, cardiac abnormalities. Children with Dandy Walker syndrome usually present with developmental delay and signs of hydrocephalus. Difficulty with balance, increased muscular tone and poor motor control are common. Associated problems may include respiratory failure due to poor respiratory control, swallowing problems, [epilepsy](#), [hearing](#) or visual problems (nystagmus and tracking problems), and [learning disability](#) in a significant proportion of children.

### **Aqueduct stenosis**

Narrowing of the cerebral aqueduct.

### **Chromosome abnormalities (Genetic conditions)**

Examples are [Down syndrome](#) and achondroplasia - like (restricted growth) syndromes.

### **Perinatal**

This means the cause occurred between the onset of labour and the 28<sup>th</sup> day of life. The most common cause is intraventricular haemorrhage which

is bleeding into or around the ventricles of the brain. Infection is also a common cause.

Intraventricular haemorrhage is more common in low birth weight, preterm and very preterm infants.

### **Postneonatal**

This means the cause occurred after the 28<sup>th</sup> day of life. The most common causes are -:

- Idiopathic (i.e. there is no obvious cause)
- Infection - most commonly as a result of meningitis
- Haemorrhage - bleeding
- Tumours - especially those affecting the back of the brain, are a rare cause

### Joint Hypermobility and the Joint Hypermobility Syndrome

What is Joint Hypermobility?

In simple terms a hypermobile joint is a joint, which moves through a wider range than usual (like 'double-jointedness'). However, there are two important things to clarify in any claim.

- **Adjective or diagnosis:** lots of people have 'looser than average' joints (half the population by definition). Any of these people may use the adjective 'hypermobile'. However, only a very small proportion will experience symptoms as a result. This minority has a diagnosis – Joint Hypermobility Syndrome (JHS)
- **Hypermobility or hypotonia:** just as hypermobility affects joint stability and posture so also does muscle tone and strength. Low muscle tone (hypotonia) and diseases causing muscular weakness both reduce joint stability and cause problems with joint posture, but they are different from non-muscular hypermobility and should be dealt with separately

A small proportion of people with hypermobility, and particularly with more extreme hypermobility and/or symptoms, have an underlying medical condition.

This means that joint hypermobility may affect a claimant in one or more of three ways:

- Symptoms and consequent limitations
- Treatment burden for the primary condition or its complications
- Medical problems linked to an underlying cause

#### Underlying conditions & associations

There are key underlying conditions affecting the strength of the collagen fibres that make up joint-stabilising ligaments and tissues. These include Ehlers-Danlos Syndrome (EDS), Marfan Syndrome (which also causes tallness and eye and heart problems) and Osteogenesis Imperfecta (causing bone weakness).

Hypermobility in children with developmental delay, cognitive disability or cerebral palsy probably arises from hypotonia and is not covered by this guidance.

#### Effects and signs

Muscle pains or strains.

Muscles in children with Joint Hypermobility Syndrome have to work harder as the joints are too flexible. This can cause pains and strains – commonly after exercise.

#### Joint stiffness

This can occur if joints are injured or overused. Typical symptoms include swelling and reduced movement.

#### Postural pain

Some of the joints that carry a significant postural load can become painful with no signs of swelling, inflammation or injury. This may result from abnormal postural positions or loads. Most typically this affects the feet, ankles, back and neck.

#### Joint dislocation or subluxation (partial dislocation)

Hypermobility joints are more likely to get injured if they're overstretched. Sometimes the joint can dislocate – this is most common in the shoulder or

the kneecap. Sometimes the soft tissues in and around joints (cartilage, tendons, ligaments) can tear.

How is it assessed?

Most children will have been to see their general practitioner. Those with significant symptoms will usually have been referred to see a Paediatrician or Paediatric Orthopaedic Surgeon.

Few children have a full Beighton assessment (see below), but any formal assessment deals with the same two issues – evaluating the extent of hypermobility and evaluating symptoms. Clearly it's possible to have extreme hypermobility with no symptoms or symptoms with modest hypermobility, but, in general, symptoms become more likely with greater degrees of hypermobility.

The Beighton assessment

The Beighton score is measured by adding 1 point for each of the following:

- placing flat hands on the floor with straight legs
- left knee bending backward
- right knee bending backward
- left elbow bending backward
- right elbow bending backward
- left thumb touching the forearm
- right thumb touching the forearm
- left little finger bending backward past 90 degrees
- right little finger bending backward past 90 degrees

Some older children will have their diagnosis confirmed by the Brighton criteria. The Brighton criteria integrate the Beighton score with symptoms into a diagnostic tool. This tool has only been validated for children over the age of 16.

The Brighton criteria do not replace the Beighton score but instead use the previous score in conjunction with other symptoms and criteria. JHS is diagnosed in the presence of either two major criteria, one major and two minor criteria or four minor criteria. The criteria are:

**Major criteria:**

- A Beighton score of 4/9 or more (either current or historic)
- Arthralgia for more than three months in four or more joints

#### Minor criteria:

- A Beighton score of 1, 2 or 3/9 (0, 1, 2 or 3 if aged 50+)
- Arthralgia (> 3 months) in one to three joints or back pain (> 3 months), spondylosis, spondylolysis/spondylolisthesis
- Dislocation/subluxation (partial dislocation) in more than one joint or in one joint on more than one occasion
- Soft tissue rheumatism > 3 lesions (for example, epicondylitis, tenosynovitis and bursitis)
- Tall and slim. Arm span/height ratio greater than 1.03. Upper body to lower body segment ratio less than 0.89, arachnodactyly (fingers or toes are long and thin when compared to the palm or sole of the foot), positive Steinberg thumb (when folding the thumb across the palm it extends past the end of the palm) /Walker wrist signs (when grasping the wrist of the opposite hand, the thumb and index finger overlap each other)
- Abnormal skin: striae (scar like lesions), hyperextensibility (ability to extend a joint beyond the normal range) , thin skin, papyraceous (paper like) scarring
- Eye signs: drooping eyelids, myopia (short-sightedness) or anti-mongoloid palpebral slant (angle of the line where closed eyelids meet)
- Varicose veins, hernia or uterine/rectal prolapse

#### Treatment and management

##### Physical Therapy and Exercise

Research has shown the benefit of exercise and physiotherapy. In most cases claimants can ease their symptoms by doing gentle exercises to

strengthen and condition the muscles around the hypermobile joints. This is only effective if these strengthening exercises are done often and regularly.

Physiotherapists will be able to advise on suitable exercises and monitor any improvements in symptoms. For some people gentle stretching seems to be of additional benefit.

### Medication

Non-opiate analgesics drugs such as Paracetamol.

Non-steroidal anti-inflammatory drugs such as Ibuprofen.

### Surgery

This usually only recommended for repeated dislocations of a joint or if there is a tear of a ligament or tendon.

### Sources of Evidence

Source of evidence	evi-	Information Pro-	Limitation of Evi-
General Practitioner		Up to date assessment of symptoms and any specialist treatment	Unable to supply information about day-to-day function
School/teacher		Reports detailing what, if any, help is required during the normal school day	Will be unable to supply information about current treatment
Orthopaedic Surgeon / Paediatrician		Information about investigations or planned treatments	May be unable to supply assessment about day-to-day function
Physiotherapist		Information about functional level and response to any treatments	May be unable to help with objective assessment of day-to-day functional level.

### Severity indicators guide

Mild functional restriction	under GP and physiotherapy; receiving medication to control symptoms but will be taking as and when needed
Moderate Functional restriction	may have seen an orthopaedic surgeon but not under active follow up; taking regular medication; under the care of the GP
Severe functional restriction	may have had surgery; under follow up with an orthopaedic surgeon; receiving physiotherapy and occupational therapy

Most children with joint hypermobility syndrome will improve slowly over time and it is expected that the majority will be better by the time they reach their late teens.

Those children under their GP and physiotherapy only, are unlikely to have significant functional impairment and as a result are unlikely to have significant needs.

Children most severely affected would be expected to be under follow up with an orthopaedic surgeon or paediatrician. In addition, they may be having support from physiotherapy and occupational therapy.

How long will the needs last?

Award length will depend upon the severity of associated disability. Treatments for joint hypermobility syndrome are effective and most children should expect symptoms to improve with therapy.

An award is suggested for 2 years if the child is under active follow up with an Orthopaedic surgeon.

An award is suggested for 12 months for children who are under their GP only, with or without physiotherapy.

## **What you need to know about Juvenile Idiopathic Arthritis?**



### **What is Juvenile Idiopathic Arthritis?**

Juvenile Idiopathic Arthritis (JIA) is defined as persistent arthritis in 1 or more joints for at least three months and occurs in children under 16 years.

- [Juvenile Idiopathic Arthritis](#)

### **What are the effects and signs?**

There are 3 major subtypes of JIA as based on the symptoms at disease onset .....

- [Effects of Juvenile Idiopathic Arthritis](#)

### **How is it assessed?**

Arthritis is usually suspected from symptoms of constant joint pain or swelling.

- [Assessment of Juvenile Idiopathic Arthritis](#)
- 
- Linked pages: [Severity indicators](#)

### **How is it treated and managed?**

The goals of treatment are the control of inflammation, pain relief, prevention or control of joint damage .....

- [Treatment for Juvenile Idiopathic Arthritis](#)

### **What evidence is available?**

Juvenile arthritis is often managed by a multidisciplinary team.

- [Evidence](#)

### **How long will the needs last?**

- [Prognosis and duration of the award](#)

### **What is Juvenile Idiopathic Arthritis?**

Juvenile Idiopathic Arthritis (JIA) is defined as persistent arthritis in 1 or more joints for at least three months and occurs in children under 16 years. It is the most common form of arthritis in children and is one of the most common chronic diseases of childhood. It is a group of disorders that present as chronic joint inflammation. It is not clear what causes JIA, but it is

an autoimmune disorder, i.e. the immune system, which normally fights infection, attacks the body's own tissues (in this case joints).

The most common symptoms of JIA are joint swelling, pain and stiffness. JIA can affect any joint and, in some cases, internal organs as well. The symptoms are usually worse in the morning or after a rest period. The condition may start with a high fever and skin rash and lymph node swelling; or more slowly with a parent noticing a child becoming increasingly clumsy.

Most children with JIA have times when the symptoms get better or go away (remission) and other times when they get worse (flare).

Terms: Various terms are/have been used to describe JIA; these include juvenile rheumatoid arthritis, juvenile chronic arthritis & Still's disease. However, juvenile idiopathic arthritis is the preferred term.

[Other forms of arthritis affecting children](#)

**What are the effects and signs?**

There are 3 major subtypes of JIA as based on the symptoms at disease onset.

Type	Symptoms	Additional Characteristics
Pauciarticular (50%)  Sub-type - extending pauciarticular	Fewer than 5 joints involved during the first 6 months.  New joints affected slowly over a number of years.	Usually affects large joints (e.g. knee) in early childhood.  Affects girls more often.  High incidence of chronic uveitis (inflammation of the middle layer of the eye).  This subtype has poor prognosis.
Polyarticular (30-40%)	5 or more joints involved during the first 6 months.	Both large and small joints.  Can also affect the lower jaw in later childhood.  Affects girls more often.  Resembles adult rheumatoid arthritis.
Systemic (10-20%)	Fever, light salmon-coloured rash.	Enlarged liver and spleen.  Anaemia.  Lung and cardiac involvement.  Affects boys and girls equally.

- [Complications](#)

### **How is it assessed?**

Arthritis is usually suspected from symptoms of constant joint pain or swelling. There is no test for JIA, the diagnosis being made by 'pattern' recognition preferably by a specialist. In order to be sure that it is JIA, clinicians rely on:

- A complete history of symptoms (must be present for 3 months or more)
- Physical examination (to look for signs of joint inflammation, rashes, nodules, signs of internal organ inflammation and/or eye problems)
- Family history
- Blood tests:

To detect signs of inflammation.

To look for the antinuclear antigen (ANA), which is commonly found in systemic lupus and which, is also associated with uveitis in pauciarticular JIA.

- X rays, which are used sparingly and only when there is doubt about the diagnosis or when surgery is being considered

### [Severity indicators](#)

### **How is it treated and managed?**

The effective management of JIA requires early diagnosis and a multidisciplinary team approach as well as education of the patient and family. The goals of treatment are the control of inflammation, pain relief, prevention or control of joint damage and to maximise function. For children with severe arthritis or a rapid progression of symptoms, the use of two more modalities (methods of treatment) appears to alter the disease course.

### **Pharmacological management**

#### **1. NSAIDs**

Non-steroidal anti-inflammatory drugs (NSAIDs) are used to help control pain and inflammation. Common NSAIDs include ibuprofen, naproxen, and indomethacin.

Side effects of concern are gastric irritation, liver and kidney toxicity or other evidence of drug sensitivity that require stopping the use of these drugs.

However, children are much less likely to suffer such side effects than adults.

## **2. Analgesics**

Analgesics like paracetamol (Panadol) and tramadol provide pain relief.

## **3. DMARDs**

Disease modifying anti-rheumatic drugs (DMARDs), reduce the activity of JIA. These drugs are often used in combination with NSAIDs. They require frequent monitoring blood tests. Some of these medications are listed below:

Methotrexate is the most commonly prescribed DMARD and has made a significant difference in the management of JIA. It can help the arthritis, as well as the systemic illness including control of uveitis. It starts to work quickly in most cases but can take up to 4 weeks to take effect. It is given weekly either orally as a liquid or in pill form, or, more usually, by injection. Regular laboratory monitoring is important as it can affect the blood count as well as causing nausea, mouth ulcers, and more rarely liver and lung problems, although it is usually tolerated very well by children.

Other DMARDs used in JIA are Hydroxychloroquine (Plaquenil) Sulfasalazine (Salazopyrin); Gold injections (Myocrisin); Penicillamine; Azathioprine (Imuran); Cyclophosphamide and Cyclosporin A.

## **4. Biological Agents**

Biological agents are a newer class of medications made of synthetic proteins that block the inflammatory proteins in patients with arthritis. These drugs include:

- Etanercept (Enbrel) - a self-injectable drug given twice weekly.
- Infliximab (Remicade) IV - infusion every two months.
- Adalimumab (Humira) - a self-injectable drug.

## **5. Steroids**

Steroid treatment is reserved to treat more severe features of illness. They may also be injected into joint space to treat a single troublesome joint. Steroid eye drops are used to treat Uveitis. Although effective in decreasing inflammation, they have significant long-term side effects when taken orally.

Prolonged steroid use may cause growth problems, weak bones and decreased resistance to infections.

The usual preparation used is prednisolone.

## **Non- Pharmacological management**

### **Splints**

Splints help keep joints in the correct position and relieve pain. They may be used for a variety of joints, from the knee to the wrist and fingers. Often worn at night, they may be used to treat or prevent contractures. The child is usually under the care of a specialist occupational therapist or physiotherapist.

### **Exercise**

Therapeutic exercise can improve joint flexibility and build muscles. It helps to keep joints mobile; keep muscles strong; regain lost motion or strength in a joint or muscle. Crucially, it makes everyday activities like walking, eating and dressing easier. A therapist instructs on how to perform these exercises at home. Hydrotherapy may be of benefit to strengthen muscles and improve range of motion of joints.

### **Eye Care**

Frequent eye examinations in at-risk children can identify inflammatory problems early and reduce the potential for serious eye complications. Eye inflammation may occur even when the joint disease is inactive, therefore periodic examination is essential.

### **Dental Care**

Problems with jaw movement can make brushing and flossing their teeth difficult. Various toothbrush handles, electric toothbrushes and rinses can be used to maintain healthy teeth and gums.

### **Surgery**

Surgery is used as a last resort in children that have responded poorly to medication and other measures. 'Release' operations may be needed to loosen tight muscles and tendons and increase range of movement. In order to fix deformed bones, an operation called an osteotomy may need to be performed to reset the bones. Total hip and knee replacements may

relieve pain and restore function in a functionally disabled child with debilitating disease.

**What evidence is available?**

Sources of evidence	Information Provided	Limitation of Evidence
Parent / carer	First source of information and associated evidence.	Does not have specialist knowledge.
<p>Juvenile arthritis is often managed by a multidisciplinary team.</p> <p>It is important to determine the healthcare professional that is regularly managing the child's condition. This enables contact with the person most likely to give the most up to date and most useful information</p>	If medical evidence relating to JIA and its treatment are required this is the best source of evidence.	N/A
<p>The team may include:</p> <ul style="list-style-type: none"> <li>Paediatric rheumatologist</li> </ul>	Provides an overall profile of significant issues, medication and most up to date treatment and its effect.	N/A
<ul style="list-style-type: none"> <li>Occupational therapist</li> </ul>	Can provide valuable information regarding function and interventions such as splints/ aids	May be unable to provide information on overall management and control of the condition.
<ul style="list-style-type: none"> <li>Physiotherapist</li> </ul>	Can provide information about therapeutic exercise and function.	May be unable to provide information on overall management and control of the condition.



<ul style="list-style-type: none"> <li>• Specialist nurse</li> </ul>	Works closely with the rheumatologist and can provide information about overall control, drug treatment, blood tests etc.	N/A
<ul style="list-style-type: none"> <li>• Ophthalmologist</li> </ul>	For those with visual impairment, can provide information to indicate level of visual impairment	May be unable to provide information on overall management and control of the condition.
<ul style="list-style-type: none"> <li>• Orthopaedic surgeon</li> <li>•</li> </ul>	Provide details of outcome and any future surgical procedures, as well as information on joint function.	May be unable to provide information on overall management and control of the condition.

### How long will the needs last?

This guidance covers -:

Juvenile Idiopathic Arthritis (also known as juvenile rheumatoid arthritis, juvenile chronic arthritis & Still's disease)

The increase in effective drugs and the change towards earlier, aggressive therapy has changed outcomes. With proper therapy, the majority of

children will improve over time. There is a minority of children however that go on to have problems with active disease through to adulthood.

Most children will have little or no disability and may experience temporary difficulties in activity during a flare up but after appropriate treatment return to full function.

A few children experience moderate disability and fewer still have severe problems.

JIA has a very variable course of disease, and it is certainly difficult to predict long term outcome accurately. There are some factors, however associated with poorer long-term outcomes and some of these are:

- Extending pauciarticular JIA
- Systemic JIA
- Prolonged active disease (more than 4 years)

Duration of functional restrictions for those with significant disability will depend on the length of time the child has had JIA.

<b>Age at date of claim</b>	<b>Date of Onset</b>	<b>Award Period</b>
1-15	Less than 4 years or child currently on a waiting list for joint replacement surgery	2 year award
	More than 4 years	Indefinite award

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Other forms of arthritis affecting children**

There are several other forms of arthritis that can affect children and these are distinct from JIA, although in the early stages are indistinguishable from JIA. Spondyloarthropathy is a family of related diseases causing a form of arthritis. It includes ankylosing spondylitis, psoriatic arthritis, spondyloarthropathy associated with inflammatory bowel disease, reactive arthritis,

and Behcet's syndrome. They are distinct from JIA in that the optimal treatment, monitoring and outcome are different.

Adult-type rheumatoid arthritis can also develop in early teenagers. This will need to be treated aggressively and this type of arthritis is life-long.

## **Complications**

JIA can produce serious complications in more severe cases.

### **Joints**

JIA primarily affects the joints and may cause significant damage. Long term disease can gradually result in deformity, wasting of muscles, tight ligaments, and contractures (permanent shortening of a muscle or tendon making it hard to stretch the area and preventing normal movement.) It can cause bones and joints to grow unevenly causing one limb to be longer than the other.

### **Eyes**

JIA can cause Uveitis (eye inflammation), which often has no symptoms in the initial stages. This complication is seen most frequently in pauciarticular disease, especially in the extending subset. It is associated with the presence of antinuclear antigen (ANA) in the blood tests. Regular monitoring is done to detect early disease in 'at risk' children. If not detected and treated, it may lead to scarring of the lens and permanent visual damage (even blindness). Treatment with steroid eye drops often prevents progression of disease.

### **Jaw**

Slower growth may occur in the jaw due to arthritis in the jawbone (at the temporomandibular joint - TMJ) causing pain and discomfort with chewing as well as headaches. It may also affect dental care and eating habits.

### **Other effects**

Some patients develop inflammation of the lungs (pleuritis) or around the heart (pericarditis) with occasional fluid accumulation around the lungs (pleural effusion) or heart (pericardial effusion). Also, inflammation rapidly breaking down red blood cells may lead to anaemia.

### **Severity indicators**

The improvement in management has made a tremendous difference in terms of function for children with JIA; the majority of children are well controlled. The main determination when considering function is deciding if the condition is "well controlled" or "poorly controlled". If a child has poorly controlled JIA, they are much more likely to have significant functional impairment. Included below are potentially useful characteristics of children with JIA that indicate poor control and therefore functional problems to a level that is likely to give rise to care and mobility needs. However, consideration

should be given to children with long term disease that may be well controlled now but that has left them with permanent joint deformities.

Indicators of poor control:

1.) **Active symptoms** persistent morning stiffness; joint swelling, pain and muscle wasting. These symptoms should be present in a major joint/s: neck, wrist, fingers, hips, knees, ankles.

If objective information is available on the degree of loss of range of motion of the joint, a reduction of joint range of motion of more than 30% would indicate that a significant disability is likely.

2.) Sub-type of arthritis:

The worst severity is seen in Systemic Onset JIA, Extended Oligo-articular JIA and Poly-articular JIA, all of which may also cause significant disability because of the greater number of joints involved and the fact that these diseases tend to get worse over time if medical control is difficult.

Those children who are Rheumatoid factor positive (if that information is available) tend to have more aggressive disease.

3.) **Disease modifying anti-rheumatic drugs (DMARD):** persisting symptoms despite being on a DMARD such as Methotrexate.

4.) **Duration of diagnosis:** Children with longer duration of disease have a higher risk of developing physical disability. Those children with ongoing

symptoms for more than 6 months are more likely to have significantly reduced function.

5.) Awaiting Joint replacement surgery: Rare in children and used only as a last resort; indicative of significant functional impairment (usually hip or knee).

6.) Presence of a prescribed mobility aid: Children with significant functional restriction of lower limb joints may be prescribed an aid such as a wheelchair.

7.) Systemic JIA: Involvement of eyes, lungs, heart

- Eye (ophthalmologist report to indicate level of visual impairment);
- Heart/ lung (specialist report required to determine if significant disability - look for pulmonary and cardiac function tests)

Please note the following:

Physiotherapy is not a distinguishing criterion for JIA since all children with JIA should have a physiotherapy/home exercise plan.

These characteristics are a guide and should be considered when determining where a child is on the severity spectrum. Therefore, these characteristics should not be used in a prescriptive way as usually, no single characteristic tells specifically the level of severity. Additionally, for each level of severity indicated, every single characteristic may not be present; however

medical evidence/advice should clarify where a child may be on the spectrum.

## What you need to know about Learning Disabilities

### What is Learning Disability?

Learning disability includes the presence of a significantly reduced ability to understand new or complex information .....

- [Learning Disabilities](#)

### What are the effects and signs?

The most obvious manifestation of learning disability is low performance in all intellectual tasks .....

- [Effects of learning disabilities](#)

### How is it assessed?

Learning disability may be identified after birth by hospital doctors .....

- [Assessment of learning disabilities](#)

### How is it treated and managed?

Treatment for children involves, Education, Leisure and recreation, Psychological therapies .....

- [Treatment for learning disabilities](#)

### What evidence is available?

A Multidisciplinary community team may provide information about disability and needs, clinical features, treatment .....

- [Evidence](#)

### How long will the needs last?

- [Prognosis and duration of the award](#)

## What is a learning disability?

Difference between a learning disability and a learning difficulty

Prevalence of learning disabilities

Clinical presentations of learning disabilities

Profound learning disability

Severe learning disability

Moderate learning disability

Mild learning disability

Causes of learning disability

Premature babies

A learning disability is defined by 3 core criteria:

1. Lower intellectual ability (usually an IQ of less than 70).
2. Significant impairment of an ability to engage in social interactions, interpersonal relationships, and activities of independent living or, significant impairment in how well a person handles common demands in life and how independent they are compared to others of a similar age and background.
3. Onset in childhood.

Having a lower than average IQ does not automatically mean that an individual will have a lower than average level of everyday functioning.

The degree of assistance required may vary in terms of intensity (e.g. physical or verbal prompting) and frequency (e.g. daily or less often than daily), but the

required assistance should always be outside the range of that expected within the individual's particular culture/community.

Difference between a learning disability and a learning difficulty

Many people with learning disability prefer to use the term learning difficulty. This can cause confusion as the terms then become interchangeable.

A learning difficulty is a condition which creates an obstacle to a specific form of learning, but does not affect the overall IQ of an individual. For example, Down syndrome is classed as a learning disability because the

child's learning is impaired in all areas, whereas dyslexia is classed as a learning difficulty because learning is impaired in a specific area.

In terms of health and social care it is the underlying level of functional restriction that is important and not the specific term.

#### Prevalence of learning disabilities

It has been estimated that 1,519,000 people in England (2% of the population) have a learning disability.

Currently there are 286,000 children who have a learning disability.

#### Clinical presentations of learning disabilities

Having a learning disability means that people find it harder to learn life skills. The problems experienced vary from person to person and can include; communication, managing money, reading, writing, or personal care. Some people are born with a disability, whereas others may develop one as a result of an accident or illness in childhood.

Functional effects differ hugely. Someone with mild disabilities may be able to live independently with minimal support, whereas someone with severe and profound disabilities may require 24-hour care, and help with performing most daily living skills.

A learning disability is defined by the Department of Health as a “significantly reduced ability to understand new or complex information, to learn new skills (impaired intelligence), with a reduced ability to cope independently (impaired social functioning), which started before adulthood” (Department of Health, 2001).

Sometimes, the term 'Global Developmental Delay' (GDD) is used to describe a learning disability. GDD is usually used to describe failure to reach developmental milestones between birth and the age of 18, for example learning to communicate, processing information, remembering things and organising their thoughts.

#### Profound learning disability

- 1% of children with a learning disability
- IQ less than 25

Most children in this group need support for complex health needs involving hearing, vision, mobility and learning as well as often having other problems such as epilepsy and autism. They may require extensive support for all their personal needs such as feeding, toileting and bathing. They may have considerable difficulty communicating and often have no or very limited



speech and characteristically have very limited understanding. In addition, some people may need support with behaviour that is seen as challenging.

#### Severe learning disability

- 7% of children with a learning disability
- IQ 25-39

These children often use basic words and gestures to communicate their needs. Many need a high level of support with everyday activities, but they may be able to look after some if not all of their own personal care needs. Some people may have additional medical needs and some need support with mobility issues.

Most are educated in a specialist school. Some will be maintained within a mainstream school with a statement of special educational needs or in a specialist unit attached to a mainstream school.

#### Moderate Learning Disability

- 12% of children with a learning disability
- IQ 40-54

These children are likely to have some language skills that mean they can communicate about their day to day needs and wishes. Some people may need more support caring for themselves, but many will be able to carry out day to day tasks.

Children with moderate learning disabilities are more likely to need more input in educational settings. They can be taught in mainstream school with additional support and a statement of special educational needs. More often they will be in a special school designed to help teach activities of daily living and education aimed at their developmental level rather than age.

#### Mild learning disability

- 80% of children with a learning disability
- IQ 55-69

They are usually able to hold a conversation and communicate most of their needs and wishes. They may need some support to understand abstract or complex ideas. People are often independent in caring for themselves and doing many everyday tasks. They usually have some basic reading and writing skills. People with a mild learning disability quite often go

undiagnosed. They may still however, need support with tasks such as budgeting or completing forms.

Education will usually be in mainstream schools. Sometimes this will be with an individual education plan.

Children with mild learning disabilities have varying delay in their developmental milestones. Their milestones can vary from normal to a global mild delay in all areas.

### Causes of learning disability

Learning disabilities are caused by something affecting the development of the brain. This may occur before birth (prenatally), during birth, or in early childhood.

Learning disabilities can be caused by any one of a variety of factors, or by a combination. Sometimes the specific cause is not known. Possible causes include the following:

- An inherited condition, meaning that certain genes passed from the parents affected the brain development.
- Chromosome abnormalities such as Down syndrome
- Complications during birth resulting in a lack of oxygen to the brain
- A very premature birth
- Mother's illness during pregnancy
- Excessive drinking during pregnancy
- A debilitating illness affecting brain development
- Injury to the brain during childhood, for example a road traffic accident or child abuse
- Contact with damaging material (like radiation)
- Neglect, and/or a lack of mental stimulation early in life.

Some people with learning disabilities have additional physical disabilities and/or sensory impairments.

### Premature babies

Premature babies are ones born before 37 weeks' gestation.

There are 770,000 births every year and 7.1% of these will be premature. That is 54,000 preterm babies every year.

Weeks' Gestation	Percentage
32-36	84%
28-31	11%
24-27	5%

The earlier the child is born premature the more likely that child will have learning disabilities when older.

For babies born before 27 weeks' gestation:

Learning disability	Percentage
Borderline	23%
Mild	34%
Moderate	21%
Severe or profound	22%

Premature birth is often associated with other problems, including socio-economic difficulties, and it is difficult to work out which of the various factors contributes most to the baby's eventual developmental outcome.

### **What are the effects and signs?**

#### **Sleep Problems in children with learning disabilities**

Learning disabilities are probably linearly associated with sleep difficulties. That is to say, that the more severe the underlying learning disability the more severe the sleep problems will be. There are a number of reasons for this, and these often need considerable medical involvement. A number of learning disabilities with underlying known genetic causes have their own

specific sleep disorders, here is a short list of the most common of these, for rarer disorders seek medical advice:

- Down syndrome - 50% have obstructive sleep apnoea
- Smith-Magenis Syndrome - Severe sleep problems; often 'day night reversal' and awake most of night. Also, often self-injury
- Angelman Syndrome - Severe learning difficulties and often epilepsy but also independently severe sleep difficulties
- Cri du Chat - Severe learning difficulties and sleep problems
- Sotos Syndrome - Challenging behaviours and sleep problems
- Williams Syndrome - Overactive, impulsive problems and sleep difficulties
- Pierre -Robin Syndrome and cleft palate - often sleep related breathing (obstructive sleep apnoea) problems

In children with learning disabilities nighttime settling problems were reported for 41 per cent of children aged 4-12 in special schools compared with 27 per cent of children in mainstream schools; figures for night waking in the same circumstances were 45 per cent compared with 13 per cent. Figures for children with severe learning disability are particularly high: for example, problems in settling at night can be present in over 80 per cent of children aged up to 11 years and 77 per cent of 12 to 16 years.

### Challenging behaviour

What are challenging behaviours? A person with challenging behaviour may have the following: have temper tantrums, (more than would be expected for their age), be un-cooperative, make unreasonable demands, injure themselves, be extremely noisy, shout at others, display physical aggression towards others, for example, kicking, biting, slapping or damage property.

Between 5-15% of people with learning disabilities show behaviours which present a significant challenge for those caring for them. Challenging behaviour is much more likely in severe and moderate learning disabilities.

Many forms of challenging behaviour are thought of as responses to challenging situations, in that they serve as a method of communication with the

people with whom they interact (for example, stopping unwanted attention, attracting attention or attempting to explain they are experiencing pain).

### Treatment for Challenging behaviour

Gaining a good understanding of the reasons for this behaviour is extremely important:

- Treating the cause
- Dealing with social and environmental issues
- Anger management
- Establishing clear guidelines for communication with the person

Interventions used are appropriate to the child's level of learning disability and medication can be tried if all the above measures fail.

### Professional help and support

- Psychologists can help to identify the cause for this behaviour and/or suggest ways of dealing with it
- Psychiatrists and paediatricians may prescribe medication, which can be used if behavioural methods do not help
- Community nurses can support with physical and mental health needs. Outreach nurses will observe the behaviours and suggest ways of dealing with it
- Occupational Therapists may be able to help in suggesting activities which will help in reducing these behaviours
- Paediatricians are usually involved to assess the underlying development and provide an overview
- Speech & Language Therapists can help in dealing with any communication problems
- Social workers will help to make sure that the person is placed in an appropriate care setting.

### [Associated impairments](#)

### [Specific Syndromes](#)

### **How is it assessed?**

In children consultant paediatricians usually assess their development under the age of 5 years but, when they are older, assessment of learning is

done by clinical or educational psychologists. The consultant will also look for any potential biological cause, for example Down syndrome.

Intellectual ability is assessed using psychometric testing. Commonly used tests include WISC (Wechsler Intelligence Scale) for children over the age of 6 years and WPPSI (Wechsler Preschool and Primary Scale of Intelligence) which is designed to be used between 2 years 7 months and 7 years.

The age at which they are diagnosed depends on the cause and also the severity of the disability. Mild learning disabilities may only be diagnosed once a child reaches secondary school.

Those with moderate to severe disability will undergo regular review in order to monitor their progress from both a physical and behavioural point of view.

A further assessment will occur at school leaving age to clarify any on-going needs.

### **How is it treated and managed?**

#### Education

Children with a learning disability have Special Educational Needs (SEN), that is, need additional support at school. Current educational thinking stresses the importance of inclusion which suggests that children with learning disability should be educated as far as possible within mainstream schools.

The procedures for dealing with children with SEN are laid down in the Special Educational Needs Code of Practice. Local Education Authorities are obliged to follow the guidance in the code.

In a proportion of cases, when the needs of the child are not being met in mainstream school, a statutory assessment is carried out by an educational psychologist. This usually results in a Statement of Special Educational Needs (currently being replaced by Educational and Healthcare Plans). The Statement/EHCP specifies the needs of the child, the help required to meet the needs and recommends schools, either special or mainstream suitable for the child.

The majority of children with SEN have a mild learning disability and attend mainstream schools.

A minority of children with SEN attend special school, that is, one that is just for children with a statement of SEN/EHCP and is likely to cater for children

with moderate and severe learning disability – see: Special Educational Needs.

#### Leisure and recreation

Children with learning disability should be given the same opportunities to engage in social and sporting activities as children without learning disability. They may need additional support to access these opportunities.

#### Psychological therapies

A range of psychological therapies can be adapted for use in children with learning disability.

#### Drug treatment

Psychotropic medication may be used in addition to social, environmental and behavioural strategies in managing severe behavioural disorders.

#### Family support

The birth of a child with learning disability puts great strain on most families. The parents need to adapt to their changed situation, and they may grieve for the loss of their anticipated healthy child. The additional physical and financial consequences of caring for a child with additional needs may put

strain on all family relationships particularly on relationships between parents. It can take considerable time for families to adjust.

### What evidence is available?

Sources of evidence	Information Provided	Limitation of Evidence
Parent / Carer	First source of information and associated evidence for example Statement of SEN/Education Healthcare Plan or Co-ordinated Support Plan (CSP) or an Individual Education Plan (IEP) etc.	Does not have specialist knowledge.
Multidisciplinary community team (Community / Hospital Paediatrician, Consultant psychiatrist, Consultant psychologist, specialist nurse – learning disability, Associated Health Specialists E.g. Occupational Therapist, speech and Language Therapist, Physiotherapist)  (if child is under their care)	Clinical features, treatment and information about disability and needs.  Also request a copy of a completed standardised assessment (e.g. the Adaptive Behaviour Scale) and a copy of a Statement of Special Educational Needs/Education Healthcare Plan if available.	Information regarding symptoms and disabling effects may be based on what the parent / carer has told the relevant professional.
Special Needs teacher/coordinator  (if child has one)	Resulting disability or needs.	May not have information about symptoms, investigations and treatment but this may vary depending on the type of school the child is attending.

### How long will the needs last?

This guidance refers to a number of different conditions and syndromes; some may cause both mental and physical disability. This guidance covers



the mental impairment aspect of the overall disability only. Refer to the appropriate guidance where there is also any physical impairment.

This guidance covers:-

<b>Impairment</b>
Fragile X syndrome
Learning disability other / type not known

The following table is to be followed if an award is made. It is not a statement of intended awards for individual categories.

Age at Date of Claim	Length of Award			
	Mild	Moderate	Severe	Profound
1-4 years	To age 5 or for two years (whichever is greater)	To age 5 or for two years (whichever is greater)	Award to age16	Award to age16
5-9 years	To age12	To age12	Award to age16	Award to age16
10-15 years	To age16	Award to age16	Award to age16	Award to age16

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

## **Specific syndromes**

[Specific Syndromes](#)

[Other Syndromes](#)

## **Specific Syndromes**

See: Down/Down's syndrome guidance.

### Fragile X syndrome

Fragile X syndrome is the second most common genetic condition associated with learning disability after Down's syndrome.

Approximately 1 in 3500 – 8900 males and 1 in 4000 females have the full mutation. Males with Fragile X syndrome tend to be more severely affected and generally have moderate to severe learning disability, while females will generally have an IQ that falls within the normal to mild learning disability range.

Fragile X syndrome is associated with a typical appearance that includes an elongated face, long and prominent ears, a large head circumference and enlarged testicles. It is also associated with physical problems such as mitral valve prolapse, joint laxity, spinal scoliosis and flat feet.

Features of Fragile X syndrome include abnormalities of speech and language, problems with attention and concentration, difficulty adjusting to change (particularly environmental change), sensory sensitivities and mood instability.

Boys tend to have more behavioural problems than girls and tend to be shy and socially withdrawn. Approximately 1/3 of adolescent males with Fragile

X syndrome may show angry and aggressive behaviour. Girls often suffer from anxiety and depression.

Care and mobility needs vary according to the severity of the condition and the associated learning disability.

### Autistic Spectrum Disorder

It is common to have a dual diagnosis of Learning Disability and Autistic Spectrum Disorder.

### **Other Syndromes**

With the exception of cerebral palsy the following syndromes are uncommon but may be associated with learning disability. Please note that this list is not exhaustive.

Angelman syndrome	<a href="#">Phenylketonuria</a>
<a href="#">Cerebral palsy</a> -: Cerebral palsy – ataxic Cerebral palsy – athetoid Cerebral palsy - causing hemiparesis Spastic diplegia Cerebral palsy - Other / type not known	<a href="#">Prader – Willi syndrome</a>
<a href="#">Cytomegalovirus</a>	<a href="#">Rett syndrome</a>
<a href="#">Foetal Alcohol syndrome</a>	<a href="#">Sturge – Weber syndrome</a>
<a href="#">Homocystinuria</a>	<a href="#">Tay – Sachs disease</a>
<a href="#">Hydrocephalus</a>	<a href="#">Triple X</a>
<a href="#">Hurlers syndrome</a>	<a href="#">Trisomy 13 Patau's syndrome</a>
<a href="#">Klinefelter syndrome</a>	<a href="#">Trisomy 18 Edwards syndrome</a>
Lesch – Nyan syndrome	<a href="#">Tuberous sclerosis</a>
<a href="#">Neurofibromatosis</a>	<a href="#">Turner's syndrome</a>
<a href="#">Cri du Chat syndrome</a>	

### Associated impairments

Mild learning disability is often an isolated disability; severe learning disability is often associated with other impairments that increase the level of disability. The frequency of these impairments increases as the level of learning disability increases. Associated physical impairments include -:

- Cerebral palsy is a general term used to refer to a set of [neurological](#) conditions, which affect a child's movement and coordination. Affected children have loss of voluntary movement,

motor difficulties affecting walking and muscular tension/rigidity. They may have feeding problems and failure to thrive. Cerebral palsy is caused by damage to the brain, which normally occurs before, during, or soon after birth. It affects approximately 1 in 400 children, of whom 60% have an associated learning disability.

Sensory impairments, of which visual impairments, especially squint and refractive errors are the most common, occur in more than half of children with severe learning disability and twenty five percent of children with mild learning disability. Hearing impairment is also common.

Epilepsy occurs in about 20% of children with learning disability. Children with learning disability often have the same types of epilepsy that occur in the general population. However, severe and mixed epilepsy syndromes are more common. Some specific medical syndromes, known to be associated with learning disability, have particular types of seizure disorders within some of these syndromes.

Motor impairments include spasticity, ataxia (unsteadiness) and abnormal movements.

Abnormal movements including head banging and rocking are common in children with severe learning disability. Although they may be due to motor pathology, in the majority they are due to behavioural problems.

Continence difficulties may well be experienced.

Living with a Learning Disability – Example Profiles

How does a learning disability affect a person's life?

[Mild Learning Disability](#)

[Moderate Learning Disability](#)

[Severe Learning Disability](#)

[Profound Learning Disability](#)

How does a learning disability affect a person's life?

People with learning disabilities do not learn certain skills as quickly as other people and may therefore need extra help in certain aspects of their lives. The specific skills in question will depend upon the type of disability. People with mild learning disabilities may live alone, travel independently, and work. They may not require any support from their local authority or may just need support in managing their finances. Other people may require more regular support to ensure their safety and health on a daily basis. Those with more severe or complex needs may need extensive, hour-

to-hour help in performing basic skills such as eating, dressing and washing.

With the right support people can live full and meaningful lives. However, if this support is not provided, they may face problems in gaining independence or a home of their own, in accessing leisure and recreation activities, and/or in developing friendships and relationships.

Children and young people with learning disability often exhibit poor judgment and are more vulnerable to influence by others. They therefore need greater levels of supervision as they may be vulnerable to involvement in criminal activities. They are also more likely to be the victim of bullying.

#### Mild Learning Disability

##### Example:

E is a thirteen-year-old with a mild learning disability. She lives at home with her family and four siblings.

She attends mainstream school with some additional support. She has 3 hours per week of one-to-one help with reading and writing with her form tutor. There is a learning support worker who helps out with E in her classes.

She requires help socialising and is supervised during break times. She walks home from school with her friends. There are no incidents of behaviour that would be considered to be challenging.

#### Moderate Learning Disability

##### Example:

P is a 12-year-old boy who lives with his parents and attends a special needs school. He has a moderate learning disability and severe dyspraxia. P has good verbal communication although he often struggles to co-ordinate his breathing and speaking.

He is struggling with daily tasks such as washing, dressing, and schoolwork and playing with his peers. He also demonstrates difficult behaviour in that he will shout and swear at his parents. This behaviour has often been the result of frustration if he was asked to do something that he has struggled with.

At home he has temper tantrums, usually when tired or struggling to understand what he has been asked to do. When he has been most upset, he will bang his head against the wall or floor. As he has grown older, this self-harming behaviour has improved but has been replaced with hitting out at his sister or he will resort to throwing things.

The school that he attends has class sizes of 8 pupils with one teacher and two teaching assistants. He has an Individual Education Plan (IEP),

receives speech and language therapy for 5 hours per week and occupational therapy for 3 hours per week.

He is handed directly over to his parents by his teacher.

### Severe Learning Disability

#### Example:

J is 8 years old and lives with his brother and both parents. He is in a special school for children with learning disabilities. He was diagnosed with severe learning disability and speech impairment (he was unable to clearly articulate words and only spoke in one or two-word phrases in English).

J is in a class of seven pupils and four support staff in addition to a teacher. He receives Occupational Therapy 5 hours per week and Speech therapy for 5 hours per week.

J will get frustrated if he does not understand the task he has been set in school. This is worse when he is tired and hasn't slept well the night before. On some occasions he has hit one of the other children in his class. When he was first at the school, he used to take the glasses of another pupil and throw them across the classroom.

For respite he attends a residential unit for one weekend a month and for 14 days during the summer.

He is driven to and from school and dropped with his teacher.

### Profound Learning Disability

#### Example:

R is 10. She was diagnosed with Global developmental delay aged 18 months. Her diagnosis was switched to profound learning disability aged 8. She lives in a residential nursing unit. She is able to communicate non-

verbally only and is fed by nursing staff. R requires two people to help wash and dress her.

She will become agitated and shout if she is hungry or in pain.

She is mobile with the use of a wheelchair but requires someone to push it.

It is likely that R will remain in residential care for her entire life.

## **What you need to know about Muscular Dystrophy**



### **What is Muscular Dystrophy?**

The main symptom of muscular dystrophy is muscle weakness. Muscular dystrophy is not a single disease.....

- [Muscular Dystrophy](#)

### **What are the effects and signs?**

Duchenne Muscular Dystrophy

is a condition caused by an X-linked recessive condition affecting the gene for the muscle protein dystrophin. ....

- [Effects of Muscular Dystrophy](#)

### **How is it assessed?**

The diagnosis of these conditions is made clinically. The exact subtype or name of the condition will be determined by investigations. ....

- [Assessment of Muscular Dystrophy](#)

### **How is it treated and managed?**

The aim of treatment in these early years is to prolong walking ability for as long as possible. ...

- [Treatment of Muscular Dystrophy](#)

### **What evidence is available?**

The diagnosis will be the most important piece of information in the medical evidence particularly in the rarer types, it is recommended that this information comes from the treating consultant.

- [Evidence](#)

### **How long will the needs last?**

- [Prognosis and duration of the award](#)

### **What is Muscular Dystrophy?**

The main symptom of muscular dystrophy is muscle weakness. Muscular dystrophy is not a single disease. These diseases are caused by an abnormal or absent protein in muscle cells. This means that the cells and therefore the whole muscle cannot shorten or contract normally. This is experienced as progressive muscle weakness. The type of muscular dystrophy can be diagnosed by examining a muscle biopsy specimen and identifying

which muscle protein is either lacking or present in lower than normal quantities.

The commonest types of muscular dystrophy are caused by deficiency or absence of the muscle protein called dystrophin. These types of muscular dystrophy cause progressive muscle weakness. They are called Duchenne and Becker type Muscular Dystrophy. Duchenne is a severe condition; Becker is usually a mild condition.

A good example of muscular dystrophy is Duchenne Muscular Dystrophy. Muscle weakness causes progressive disability from early childhood. This is because the protein dystrophin is missing from muscle cells. In this severe form of muscular dystrophy, all the muscles of the body are affected and the symptoms are muscle weakness that gets worse over time. Mobility is affected first as the skeletal muscles become too weak for walking. As other muscles including the heart muscle and respiratory muscles become weaker, respiratory failure and heart failure due to cardiomyopathy (the term for a heart muscle disorder) develop. There is no cure for muscular dystrophy; management involves controlling the effects of muscle weakness to maintain life and quality of life for as long as possible. Such severe forms of muscular dystrophy are fatal. The emotional effects on a family of diagnosis in early childhood of a progressive severely disabling and eventually fatal disease are significant.

In Becker Muscular Dystrophy the dystrophin protein is present but in less than normal amounts and the affected person may develop some symptoms in childhood but muscle weakness usually becomes a problem only in adulthood.

Muscle proteins other than dystrophin are absent or reduced in the rarer types. Some forms of muscular dystrophy are so rare or so variable in their effects that they are not covered in this guidance – seek medical advice. There are many different types, some of which are quite mild and compatible with a normal life. All of them are inherited and in many a definitive diagnosis can be made on genetic testing or muscle biopsy. The disabling effects of most of these conditions and their prognosis are well described. In this guidance the clinical features and care and mobility considerations of each form of muscular dystrophy will be described separately because the disabling effects and the prognosis of each type are specific. The investigations however, will be described together. It is recommended that the investigations section is used to confirm the diagnosis claimed, if necessary, and then use the appropriate section for clinical features and care and mob. There are some very rare forms of muscular dystrophy which are not

included in this guidance because their effects are not well described or very variable – seek medical advice.

**What are the different types of Muscular Dystrophy?**

<b>Type of muscular dystrophy</b>	<b>Severity of condition</b>
Duchenne Muscular Dystrophy	severe
Becker Muscular Dystrophy	mild
<b>Rarer types of muscular dystrophy</b>	<b>Severity of condition</b>
Limb girdle muscular dystrophy	<p>Many different types of varying severity:</p> <p>Adult onset LGMD types 1A,1D, 1E, 1F</p> <p>Childhood onset LGMD types 1B, 1C, 2A, 2B, 2C, 2D, 2E,2F, 2G,2H,2I, 2J</p>
Facioscapulohumeral muscular dystrophy	<p>infantile onset – severe</p> <p>childhood onset – moderate or severe</p> <p>adult onset - mild</p>
Emery-Dreifuss Muscular Dystrophy	<p>infantile onset –severe</p> <p>childhood onset – moderate or severe</p> <p>adult onset – affects heart</p>
Congenital Muscular dystrophy (CMD) with protein laminin-A2 (LAMA2) deficiency – the condition is also known as MDC1A, classic CMD or merosin deficient CMD	Usually severe, rarely mild
Ullrich Congenital Dystrophy	severe
Walker-Warburg Syndrome	severe

## **Incidence & Prevalence**

### Duchenne / Becker

The incidence of Duchenne Muscular Dystrophy is 1 in 3500 live male births. The prevalence is around 24 per million of the population in Britain. Becker Muscular Dystrophy is rarer, but the prevalence of this condition is the same, at around 24 per million of the population, this is because the life expectancy is longer in this condition.

### Other types

Most of the other subtypes of Muscular Dystrophy are very rare; indeed, some are so rare that that only a few cases have been described.

## **What are the effects and signs of the different types of Muscular Dystrophy?**

### **Duchenne Muscular Dystrophy**

This condition is caused by an X-linked recessive condition affecting the gene for the muscle protein dystrophin. This means that the abnormal gene is carried on the X chromosome. The Y chromosome, which partners with the X chromosome in boys is short and does not carry a normal copy of the dystrophin gene. This means that the condition almost always only occurs in boys.

Girls with the affected gene will have a normal gene on their other X-chromosome and will nearly always be unaffected by the abnormal gene. 2% of girls will have muscular dystrophy symptoms and they are called manifesting carriers of Duchenne Muscular Dystrophy. Symptoms are likely to be very mild and are often similar to those of Becker Muscular Dystrophy. Very rarely, a female carrier may have symptoms equivalent to the full syndrome of Duchenne Muscular Dystrophy as seen in affected boys.

The clinical features and presentation of Duchenne Muscular Dystrophy in boys is very characteristic. A common sign is big 'hypertrophied' calf muscles. The mean age at diagnosis is 4 years and 10 months but there will

have been early signs of the condition within the first three years of life. These are:

- Delayed motor milestones with half of affected children not walking by 18 months
- Delayed speech

Once walking is established children are:

- Unable to run like other children
- Struggle going up stairs
- Walk with a waddling gait when trying to walk fast
- Unable to jump up and down
- Get up off the floor by using their hands to climb up their legs to reach standing position – this is called ‘Gower’s manoeuvre’
- Learning and behavioural difficulties in up to 30%

Even with treatment and support to delay loss of walking, the mean age when independent walking is lost is 9 years. The condition is always severe but there is a spectrum of symptoms with some children losing mobility very early and others remaining mobile for longer. Almost all children will have lost the ability to walk by age 12 –this is called ‘loss of ambulation’ in the medical evidence. A small number of children will still be able to walk at 12 but will be expected to lose walking ability by age 16 – these may be called ‘intermediate’ cases or ‘Duchenne outliers’ in the evidence. It is not possible, in early childhood, to say which children will be in this category so intermediate or Duchenne outliers can only be identified by their ability to walk at age 12.

Following loss of independent walking, some children will undergo rehabilitation to enable them to walk a minimal distance with walking aids. This will enable them to take a few steps for exercise but will not constitute a useful walking ability. They will be unable to use stairs wearing aids and will need help getting from sitting to standing to walk wearing the aids. They require supervision when wearing the aids because of the risk of falls and physical help to put them on. Maintaining minimal mobility in aids like this can help to

maintain respiratory function for longer by preventing scoliosis (a deformity of the back that develops once children are confined to a wheelchair).

Eventually, even limited mobility with aids is lost and most children will be confined permanently to a wheelchair at 12 years. In the teenage years, loss of muscle tone around the spine leads to progressive and severe scoliosis sufficient to restrict breathing – this can be treated by spinal surgery. Weakness of the respiratory muscles leads to gradual decrease of Forced Vital Capacity (FVC) and they are at risk of severe chest infections and ultimately respiratory failure. Once wheelchair dependent, FVC falls gradually and sleep studies carried out once every 6 months or so are used to identify when respiratory support at night should be introduced. Night time respiratory support can prolong survival by several years. The heart muscle is also affected by the condition, when the heart is significantly affected spinal surgery cannot be performed and prognosis is much worse because lung capacity cannot be protected from increasing scoliosis. Nutritional problems are common and there is more information about this in the 'Treatment' section. Intelligence is at the low end of the normal range and up to 30% have learning difficulties. Children may be educated in mainstream or special schools. Death usually occurs in the 20s, though some individuals on respiratory and other supportive treatments are now surviving into their 30s.

Indicators of severe functional restriction -:

- Use of long callipers or ankle-knee-foot orthoses for mobility
- Virtually full time wheelchair user
- Age 12 and over
- Use of night time respiratory support
- Scoliosis or previous spinal surgery
- Gastrostomy feeding

[Disabling effects of Duchenne MD related to age](#)

### **Becker Muscular Dystrophy**

This condition is caused by an X-linked recessive condition affecting the gene for the muscle protein dystrophin. This means that the abnormal gene is carried on the X chromosome. The Y chromosome, which partners with the X chromosome in boys is short and does not carry a normal copy of the dystrophin gene. This means that the condition occurs only in boys. Girls

with the affected gene will have a normal gene on their other X-chromosome and will not be affected by the condition.

The condition has been described as a slow motion version of Duchenne Muscular dystrophy – the symptoms are similar but the onset is later and the disease takes many years to progress. The symptoms are muscle weakness but this is much milder than in Duchenne Muscular Dystrophy and walking ability is often maintained beyond age 50. Childhood is relatively normal. The onset of symptoms and the rate at which they develop is very variable. Life expectancy is normal in all but the most severe cases. The mean age of onset is 11 years. A proportion will have had delayed motor milestones in early childhood. Typical presenting symptoms are:

- Difficulty keeping up with other children when running around at school
- Difficulty climbing high steps and climbing up hills
- Muscle pains after exercise

These are the only symptoms likely to be experienced during childhood in the typical case and mobility is maintained into adulthood. Scoliosis does not usually occur as a result of this condition. These boys are at risk of cardiomyopathy and treatment may be required for this if it occurs in childhood.

A proportion of boys with Becker Muscular Dystrophy are more severely affected and present in childhood with muscle cramps and reduced walking speed. Ankle contractures (shortening) can occur and requires stretching exercises and physiotherapy. Reduced walking speed is rarely that severe but these severely affected children may require a wheelchair or scooter to get around –evidence of loss of walking ability in Becker Muscular Dystrophy must always be received from the treating Consultant Neuromuscular Diseases Specialist.

Intelligence is usually in the normal range although a minority of boys have variable learning and behavioural difficulties. They are educated in mainstream schools with no difficulties and can enter most fields of employment



on leaving school. However less physical jobs are recommended as physical strength is likely to deteriorate with age.

Indicators of severe functional restriction -:

- Severe cardiomyopathy

### **Limb Girdle Muscular Dystrophy**

There are many different types of this condition. The name of the condition describes its effects – muscle weakness affecting the large muscles around the pelvis and shoulder. In most types of this condition, problems with walking due to weakness of the muscles around the pelvis affecting the legs will be the main symptom. The general rule with these is that those that present in infancy are severe, those that present in childhood are usually moderate and those that present in adulthood are mild. Adult onset types include Types 1A, 1D, 1E, 1F – no disabling effects are anticipated during childhood.

Those that may occur during childhood are briefly described below:

#### Childhood onset

LGMD type 1B – weakness of the legs begins in early childhood and is followed some years later by weakness of the arms. Walking speed and distance may be reduced; the condition slowly gets worse until walking ability is lost. Cardiac involvement is common and a pacemaker is often inserted to prevent sudden cardiac death.

LGMD type 1C – weakness of the legs and arms occurs in early childhood and slowly gets worse. Muscle pain is a feature of this condition; contractures particularly of the hips and ankles are also common.

LGMD type 2A – causes weakness of the arms and legs with the first symptoms noted on average at 9 years of age. Muscle wasting may occur in childhood. This is a mild condition with no care or mobility needs anticipated during childhood. Muscle wasting commonly occurs later on. Loss of ambulation in this condition occurs on average at around 38 years of age.

LGMD type 2B – this type causes weakness of the legs much more than the arms and symptoms are first noted during childhood. This is a mild

condition and no care or mobility needs would be anticipated during childhood.

LGMD types 2C, 2D, 2E AND 2F – these types may be severe with walking ability lost by age 10; alternatively walking ability may be maintained until late adulthood. Cardiomyopathy is common. As with all rare types, evidence from the treating neuromuscular specialist especially on prognosis of the individual child's condition will be important.

LGMD type 2G – onset may be in early childhood or the teenage years. Foot-drop is a common problem and can be corrected by a walking splint. Young children will need help with this. The condition is slowly progressive and cardiomyopathy may occur. Needs are not anticipated except in young children who need help with splints or who have severe cardiomyopathy.

LGMD type 2H – onset in teenage or adult years. The symptom is muscle weakness but no needs are anticipated in the typical case.

LGMD type 2I and 2J– onset from childhood to adult – the progression of weakness is very variable– seek evidence on disabling effects from the neuromuscular specialist.

### **Facioscapulohumeral muscular dystrophy**

Facioscapulohumeral muscular dystrophy is named for the muscle groups it affects. These are the muscles of the face, shoulder and upper arm. The severity of the condition is related to age of onset, with the most severe cases developing symptoms in early childhood – severe cases with onset during childhood may be termed 'infantile' facioscapulohumeral muscular dystrophy in the medical evidence. The mild 'adult onset' form may be diagnosed from age 13 onwards. It will be important to confirm which type has been diagnosed and what the prognosis of the condition is from the treating specialist. The clinical features are as follows:

- Childhood onset or Infantile Facioscapulohumeral muscular dystrophy is a severe condition that occurs in early childhood. It presents with obvious facial weakness and progressive weakness of the muscles around the shoulders and pelvis. May be associated with deafness and abnormalities of the retina at the back of the eye. It is progressive and leads to loss of independent mobility in childhood, scoliosis, respiratory and

nutritional problems as in Duchenne Muscular Dystrophy. Unlike Duchenne Muscular Dystrophy, cardiomyopathy is rare.

- Facioscapulohumeral muscular dystrophy - symptoms of weakness in the affected muscles first occur in the teenage years or early twenties and may be asymmetrical. Symptoms relate to weakness around the shoulders include difficulty reaching for objects on high shelves, changing light bulbs or climbing ropes. Foot drop may also occur.

### **Emery-Dreifuss Muscular Dystrophy**

The general rule with this type of muscular dystrophy is that those that present in infancy are severe, those that present in childhood are usually moderate and those that present in adulthood are mild. Children diagnosed in infancy are likely to have a severe condition and may never be able to walk or manage activities of daily living for themselves.

Those with onset during childhood may lose walking ability before they reach 16, others may never lose walking ability. In all cases information on the diagnosis and prognosis of the condition should be requested from the treating neuromuscular specialist. A feature of this condition is involvement of the heart. Heart failure and arrhythmias are the most common causes of death. Serious heart problems particularly arrhythmias may develop by the 30s in an otherwise mild case. Fitting a pace maker can be life saving. Survival depends on appropriate management of the heart condition.

Contractures (shortening) of tendons are common and these reduce joint movement. The joints most commonly affected are the elbow, ankle and back of the neck. These contractures can be treated surgically to maintain mobility and range of movement.

### **Congenital Muscular dystrophy (CMD) with protein laminin-A2 (LAMA2) deficiency – the condition is also known as MDC1A, Classic CMD or Merosin Deficient CMD**

This is the most common form of congenital muscular dystrophy accounting for around 40% of congenital muscular dystrophy cases. As the name

suggests this condition causes symptoms early in life and is often diagnosed before the age of one. The symptoms include any or all the following:

- Floppiness (called hypotonia)
- feeding problems
- Joint contractures especially of the knees hips or ankles
- Delayed motor milestones
- Failure to thrive
- Repeated chest infections due to inadequate ventilation of the lungs
- Learning difficulties
- Epilepsy in up to 30%

The diagnosis is made based on the clinical features, muscle biopsy and genetic testing. The majority of these children are very disabled by their muscle weakness and though most will learn to sit up they will rarely be able to stand or walk. Children with rare subtypes of this condition will learn to walk and self care. Evidence from the treating neuromuscular specialist

on diagnosis and the prognosis of the child's condition will be very important.

### **Ullrich Congenital Muscular Dystrophy**

As the name suggests this condition causes symptoms early in life and is often diagnosed before the age of one. The symptoms include any or all the following:

- Floppiness (called hypotonia) in babies
- Joint laxity of the wrists and ankles
- Contractures (tendon shortening) of the hips/knees/shoulders
- Spinal deformity
- Breathing difficulties and repeated chest infections related to weak respiratory muscles in childhood
- Joint contractures especially of the knees hips or ankles
- Abnormal facial features

The diagnosis is made based on the clinical features, muscle biopsy and genetic testing. Intelligence is normal in these children. The majority are very disabled by their muscle weakness; most of them will never walk due

to muscle weakness and those that do tend to lose walking ability within 2-10 years of learning to walk.

### **Walker-Warburg Syndrome (a form of congenital muscular dystrophy)**

This condition has all the features of the other types of congenital muscular dystrophy:

- Floppiness (called hypotonia)
- Sometimes feeding problems
- Joint contractures especially of the knees hips or ankles
- Delayed motor milestones
- Failure to thrive
- Repeated chest infections

and

- Low intelligence
- Abnormal development of the eyes such as micro-ophthalmia and visual impairment
- May have hydrocephalus and may need a ventriculo-peritoneal shunt

### **Indicators of severe functional restriction in rare types of muscular dystrophy – applies to all types**

Diagnosis with:

- a form of congenital muscular dystrophy before the age of 1
- Walker-Warburg Syndrome
- Respiratory insufficiency requiring nighttime respiratory support
- Gastrostomy feeding
- Severe contractures / scoliosis with sitting or postural difficulties
- Severe cardiac involvement

### [Care and mobility considerations](#)

**How is it assessed?**

The diagnosis of these conditions is made clinically. The exact subtype or name of the condition will be determined by investigations. Investigations are used to confirm the diagnosis and give some idea of the prognosis. The tests carried out to confirm the diagnosis in these conditions are:

- Serum creatine kinase – this test measures the level of the muscle enzyme creatine kinase in blood. It is raised in most forms of muscular dystrophy. It does not confirm the diagnosis.
- Genetic testing – in Duchenne Muscular Dystrophy 60-80% of cases on dystrophin gene analysis will have an abnormality of the dystrophin gene and this will confirm the diagnosis. If the genetic test is normal this does not exclude the diagnosis, this is because the tests we have available are not sensitive enough to pick up small abnormalities of the dystrophin gene. In the other types, an abnormality may or may not be found. In rare types of muscular dystrophy genetic testing can confirm the presence of suspected genetic abnormalities and confirm the diagnosis in some cases.
- Muscle biopsy – the muscle biopsy will confirm the presence of muscular dystrophy in all cases. In most cases it will confirm the type as well. In rare types special staining techniques will be used to determine which muscle proteins are abnormal. This will often enable a precise diagnosis of the type of muscular dystrophy the child has, giving useful information on prognosis of the condition.

### **How is it treated and managed?**

#### **Duchenne Muscular Dystrophy**

##### Physiotherapy and ankle splints

The diagnosis is likely to be made in early childhood at 4-5 years of age. At this stage the child will be mobile with an abnormal gait and unable to run and jump because of muscle weakness. They are more likely to fall over or be knocked over by other children because they do not have normal strength to maintain posture and balance. The aim of treatment in these early years is to prolong walking ability for as long as possible. Treatment will consist of physiotherapy exercises focussing on stretching exercises of the hips and ankles to promote normal gait -these children tend to walk on their toes. Achilles tendon contracture (shortening) is common. To prevent this, tendons are stretched using daily exercises and ankle splints may worn

on the lower legs to stretch the Achilles tendon at night. Splints are not worn during the day at this stage.

### Steroid drug treatment

Drug treatment between ages 5 and 10 may aid muscle function and delay confinement to a wheelchair. The main drug used is corticosteroids (usually prednisolone), which may be given for a prolonged period. These improve muscle strength over a six-month period, but trials are still ongoing to determine the effect on walking ability. Side effects of treatment include weight gain, behavioural problems and changes in appearance. Obesity will make both mobilising in walking aids more difficult for the child and care more difficult once the child is confined to a wheelchair.

### Rehabilitation of walking ability for exercise when independent walking ability is lost

Eventually the progressive muscle weakness results in the child being unable to walk at all. This is called 'loss of ambulation'. Treatment at this stage may involve rehabilitation and possibly surgery to enable the child to walk a minimal distance with aids. This is likely to consist of the following:

- Fitting of long leg callipers/ knee-ankle-foot orthosis (KAFO)
- Child learning to walk in the callipers
- Surgery to lengthen the Achilles tendon may be required to enable fitting of callipers
- Training the parent to manage the callipers

Useful walking ability is rarely achieved but it does provide exercise for the child and improves prognosis. Walking in the KAF splints or callipers needs to be supervised because of the risk of falls. At first the child will be able to walk short distances in the walking aids but over time this distance will reduce as muscle weakness gets worse. If a 'functional walking distance' is mentioned in the medical evidence this means a distance of 10 metres. Many children will be using the aids mainly for standing and taking a few paces, they are unable to walk a useful distance. Wearing the aids and spending time standing may delay the onset of scoliosis.

### After independent mobility is lost

Inevitably there will come a time when a child is unable to stand even with the support of orthoses/KAFO splints/callipers and at this time they will be



confined to a wheelchair. This usually occurs at any age between 8 and 12. The aim of treatment at this stage is to preserve respiratory function to prolong survival. There are two aspects to loss of respiratory capacity in Duchenne Muscular Dystrophy. These are:

- The development of severe scoliosis (curvature of the spine), this restricts chest movement as well as making sitting in the wheelchair more difficult.
- The effect of gradually weakening respiratory muscles.

Respiratory function is monitored by measuring Forced Vital Capacity (FVC) this falls once independent mobility is lost. FVC falls gradually and boys become at serious risk of chest infection and ultimately respiratory failure. The underlying problem is that breathing is shallow because muscles are weak and this leads to low oxygen levels in the blood (hypoxia). Sleep studies will show if blood oxygen levels are becoming low at night. If this develops, night-time respiratory support will be provided. This delivers breaths via a snugly fitting face or nose mask. It is non-invasive and is not worn during the day. This type of support reduces the frequency of chest infections and improves survival. Later on in the condition, as muscle weakness progresses, the mask may be used during chest infections or routinely during the day.

### Surgery for scoliosis

Surgery is performed before severe curvature develops. During the operation the vertebrae of the spine are fused together with metalwork – this is called spinal fusion. The whole length of the spine is fused and this restricts twisting, bending and reaching ability because the spine is made rigid. The procedure prevents further curvature from developing and preserves upright posture, which is important functionally for wheelchair users as well as preserving lung function. Recovery from surgery takes around 3 months. The surgery may make some activities with the upper limbs such as eating and personal care more difficult. In particular eating becomes more difficult as they cannot bend down to the plate and must reach further. If spinal surgery cannot be performed perhaps because of cardiomyopathy making anaesthesia and surgery too risky, the prognosis for survival is poor. Surgery is always more effective than conservative management of the spinal curvature with braces or supports. The priorities for physiotherapy are postural support and containment of contractures. Some boys may wear a spinal jacket

and this is as limiting as spinal surgery in terms of movement. The carer must put on and remove the jacket for them each day. It is not worn in bed.

## Cardiomyopathy

The heart is composed of muscle, when the muscle fibres become weak heart failure develops. This can be controlled with drugs for a time. The onset of heart failure in this condition is usually the early to mid teens. Drug treatment can prevent the onset of symptoms of heart failure and prolong survival. Drugs used include:

- Angiotensin Converting Enzyme (ACE) Inhibitor
- Beta-blockers
- Diuretic tablets to treat heart failure e.g. frusemide

## Nutritional Problems

If steroid drug therapy is used to preserve mobility obesity may result. Obesity will make mobility more difficult for the child and is likely to persist into the period when mobility is lost. If the child is very heavy helping with care, especially transferring from bed to wheelchair and to the bath or shower for washing may be very difficult and a hoist may be required.

Once the child is in the wheelchair the reverse problem is encountered and weight loss is common, nutritional supplements may be necessary. Eating becomes more effortful over time and they develop swallowing problems, which can lead to choking episodes. Choking risks and difficulties with swallowing may mean that they physically cannot consume enough calories to support themselves and extra nutritional support is required. This commonly includes gastrostomy feeding.

## [Disabling effects of Duchenne MD related to age](#)

### **Becker Muscular Dystrophy**

Often no treatment is required during childhood for Becker Muscular dystrophy. Physiotherapy, splints and rarely oral steroid treatment may be used. If cramps of the legs after exercise are a particular problem, which they may be in boys in the teenage years, then intermittent compression therapy boots can be worn for half an hour at a time to relieve them (trade name for these boots is 'Flotron'). Often cramps are related to exercise or walking

and over time teenagers will learn what their walking threshold is and can avoid episodes of painful cramping by staying under it.

### [Care and mobility considerations](#)

**What evidence is available?**

Sources of Evidence	Information Provided	Limitation of Evidence
Parent / carer	First source of information and associated evidence.	Does not have specialist knowledge.
GPFR	<p>The GP may be able to confirm the diagnosis and may have access to records of home care provided.</p> <p>Becker Muscular Dystrophy - The GP will be able to confirm the diagnosis and prescription of relevant drugs</p>	The GP may not have details of treatment received and may not see the child regularly about their condition. If it is one of the rarer conditions the GP may not be able to provide any useful information.

<p>Hospital FR – best source of evidence - Hospital Neuromuscular Disease Specialist who will be able to confirm the diagnosis.</p>	<p>The diagnosis will be the most important piece of information in the medical evidence particularly in the rarer types, it is recommended that this information comes from the treating consultant who will be able to give the most accurate prognosis and information on disabling effects.</p> <p>Becker Muscular Dystrophy - Evidence is only likely to be required when cardiomyopathy or limited mobility is claimed. Information including evidence of cardiac function should be available from the neuromuscular disease specialist or cardiologist. Evidence of reduced mobility should always be obtained from the Consultant Neuromuscular disease specialist.</p>	<p>When contacting a specialist try to use the specialist co-ordinating care rather than those dealing with individual muscular dystrophy related problems such as respiratory and heart failure. This will ensure that a more complete picture of disability and related needs is provided. In all cases this will be the neuromuscular disease specialist.</p>
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**How long will the needs last?**

This guidance covers -:

Dystrophia Myotonica (Myotonic Dystrophy)
Duchenne Muscular Dystrophy
Becker Type Muscular Dystrophy
Limb Girdle Type Muscular Dystrophy
Facioscapulohumeral Muscular Dystrophy
Muscular Dystrophy Other/Type not known -: e.g.
Congenital Muscular dystrophy (CMD) with protein laminin-A2 (LAMA2) deficiency/ MDC1A/Classic CMD/Merosin Deficient CMD
Emery-Dreifuss Muscular Dystrophy
Ullrich Congenital Muscular Dystrophy
Walker-Warburg Syndrome

## **Dystrophia Myotonica (Myotonic Dystrophy)**

### **Severe functional restriction**

The most severe form of Myotonic Dystrophy is usually present from birth (congenital) or diagnosed whilst the child is still an infant and it is reasonable to expect claims to be made shortly after diagnosis. In these cases, the level of functional restriction is unlikely to/will not improve.

Age at date of claim	Award Period
Likely to be up to 1 year old	Indefinite award

### **Moderate/Mild functional restriction**

Milder forms of Myotonic Dystrophy occur but are usually diagnosed in older children. In these cases, needs may improve (particularly any care needs present) as the child gets older and is able to achieve more independence. The clinical presentation and therefore needs, in older children can be highly variable.

Age at date of claim	Award Period
1-11	To age 12 or for 2 years (whichever is longer)
12-15	To age 16

### **Duchenne Muscular Dystrophy**

There is no treatment that prevents worsening muscular weakness. Both care and mobility needs increase over time. Indefinite awards are recommended in all cases.

Note: All of these children will eventually become wheelchair reliant and totally dependent on others for self-care.

Age at date of claim	Award Period
0 -15	Indefinite award

### **Becker Muscular Dystrophy**

No needs are anticipated in the typical case during childhood.

Any award for care needs related to therapy should be reviewed at age 12 – the age when most children could be expected substantially to manage their own therapy. Children aged 12 and over will be able to manage their therapy for themselves unless learning difficulties or other problems prevent them from doing so. Indefinite awards are recommended for mobility problems.

Age at date of claim	Award Period
0 -11 – care needs related to therapy	Award until age 12 (or for 1 year, whichever is longer)

### **Limb Girdle Muscular Dystrophy**

If needs are identified solely related to therapy – for example putting on splints or having physiotherapy then review these awards at age as this is

the age when a child could be expected substantially to manage their own therapy.

If care needs or mobility problems related to severe muscle weakness are identified, indefinite awards are recommended. Children aged 12 and over will be able to manage their therapy for themselves unless learning difficulties or other problems prevent them from doing so.

Age at date of claim	Award Period
0 -11 – care needs related solely to therapy	Award until age 12 (or for 1 year, whichever is longer)
0 -15 – needs related to severe muscle weakness	Indefinite award

### **Facioscapulohumeral muscular dystrophy**

- Childhood onset or Infantile Facioscapulohumeral muscular dystrophy - Indefinite awards are recommended if needs are identified. Needs are likely to increase with time.

Age at date of claim	Award Period
0 -15	Indefinite award

- Mild form or adult onset of Facioscapulohumeral muscular dystrophy - no care or mobility needs would be anticipated during childhood.

### **Emery-Dreifuss Muscular Dystrophy**

If needs are identified solely related to therapy – for example putting on splints or having physiotherapy then review these awards at age 12 as this is the age when a child could be expected substantially to manage their own therapy. Children aged 12 and over will be able to manage their



therapy for themselves unless learning difficulties or other problems prevent them from doing so.

If care needs or mobility problems related to severe muscle weakness are identified, indefinite awards are recommended.

Age at date of claim	Award Period
0 -11 – care needs related solely to therapy	Award until age 12 (or for 1 year, whichever is longer)
0 -15 – needs related to severe muscle weakness	Indefinite award

**Congenital Muscular dystrophy (CMD) with protein laminin-A2 (LAMA2) deficiency – the condition is also known as MDC1A, classic CMD or merosin deficient CMD**

If care needs are identified indefinite awards are recommended. These children are very unlikely to be able to walk at 3 years old. Mobility awards should be reviewed at 7 as useful walking ability may be achieved between the ages of 3 and 7. Indefinite awards are recommended if walking ability is lost at any age or if good walking is not achieved by age 7 i.e. the child is only walking a few steps. No improvement is expected.

Age at date of claim	Award Period
0 -6	Award until age 7 (or for 1 year, whichever is longer)
7 -15	Indefinite award

**Ullrich Congenital Muscular Dystrophy**

If care needs are identified indefinite awards are recommended. These children are very unlikely to be able to walk at 3 years old. Mobility awards should be reviewed at 7 as useful walking ability may be achieved between the ages of 3 and 7. Indefinite awards are recommended if walking ability is

lost at any age or if good walking is not achieved by age 7 i.e. the child is only walking a few steps. No improvement is expected.

Age at date of claim	Award Period
0 -6	Award until age 7 (or for 1 year, whichever is longer)
7 -15	Indefinite award

### **Walker-Warburg Syndrome**

If care needs are identified indefinite awards are recommended. These children are very unlikely to be able to walk at 3 years old. Mobility awards should be reviewed at 7 as useful walking ability may be achieved between the ages of 3 and 7. Indefinite awards are recommended if walking ability is lost at any age or if good walking is not achieved by age 7 i.e. the child is only walking a few steps. No improvement is expected.

Age at date of claim	Award Period
0 -6	Award until age 7 (or for 1 year, whichever is longer)
7 -15	Indefinite award

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Care and mobility considerations**

#### **Duchenne Muscular Dystrophy**

Needs are summarised and described in the text and disabling effects of the Duchene Muscular Dystrophy table (see the link at the end of this section).

This can be used to identify what stage the Duchenne Muscular Dystrophy has reached and the typical needs present at this stage.

### **From diagnosis at age 4- 6 years**

#### Care

In the first few years after diagnosis extra care will be required. The parent or carer will need to help with physiotherapy stretching exercises each day. Most children need to do these exercises and typically they take 20 minutes each day to do them. Over time physical help with some aspects of mobility may be required due to muscle weakness and may include helping the child to climb into a bath if climbing is required or help or support climbing up stairs. These children tend to struggle on the stairs and getting up from the floor. They fall down more than other children and are more easily knocked over by other children in the playground – this is because they do not have the strength in their postural muscles to regain their balance.

If Achilles tendon contractures develop, they will need to apply splints to be worn over night. These are not walking splints and parents may need to help their child to the toilet during the night. The speed and distance the child can walk will reduce over time and they are likely to start using a wheelchair some of the time.

This age group may start steroid treatment to prolong walking ability. The steroids are taken in high doses and may cause behavioural problems. A side effect of steroid treatment is ravenous hunger and children will need to be restrained from eating to avoid significant obesity. Managing the diet to avoid obesity particularly if there are behavioural problems, enhanced or caused by steroid use, may be a significant aspect of care given in some families.

At some stage when the child is aged between 8 and 11 parents may need to help them rehabilitate and learn to walk again wearing long leg callipers (KAFO) as walking aids. This is because their child cannot support their

own weight to walk around and walking distance has reduced to a few steps.

## **Rehabilitation of walking ability when independent walking ability is lost age 8-12**

### **Mobility**

Fitting the child with callipers or knee-ankle-foot orthoses (KAFO) may involve surgery to release the Achilles tendon contractures (shortening) and a process of learning to walk again. With the walking aids useful walking ability will rarely be achieved and children will only be able to mobilise short distances. They will need help rising to standing in the aids and negotiating steps is not possible. They will require help to put the aids on and with transferring at other times. They will use a wheelchair some of the time and will need help with all transfers. They should be considered as virtually unable to walk once these walking aids are necessary. Some children will not try callipers or walking aids and will go straight into a wheelchair. Over time, walking with the aids will become more and more difficult and a wheelchair will be used more frequently. An electric wheelchair is the best choice because it offers the capacity for moving around independently. The weak upper limb muscles make an ordinary wheelchair unsuitable. Confinement to the wheelchair almost always occurs by the age of 12.

### **Care**

Care will be required related to the mobility aids, transferring whenever that is required and helping with therapy. Any walking aids or splints will need to be put on in the morning and taken off for washing, going to bed at night and with activities such as swimming. Help with therapy exercises to prevent contractures (shortening) may still be required on a daily basis. Mobilising around the bathroom to wash will not be possible independently and the parent or carer will have to help with this. Children are likely to need help with putting on trousers and shoes but they may still be able to put on their upper garments.

Parents may report difficulties with the layout of the home particularly if they are using a stair lift and problems with transferring their child and their wheelchair to their car to go out. If obesity related to steroid treatment is present this will be more difficult and adaptations to the home such as a hoist are more likely to be required. Upper limb mobility is maintained but the arms are weak and some help with washing and drying may be required at this stage.

A proportion of children may need turning at night from this stage because they are unable to roll over in bed or adjust their position. At this stage they

will also require assistance at night to pass urine. This is because they haven't the physical strength to manoeuvre a urine bottle. Continence aids such as catheters are rarely used by children with muscular dystrophy.

### **After independent mobility is lost age 8-12**

#### Care

Weakness affects all muscles including the upper limbs. Help will be needed with all transfers because of the effect of weakness of the upper limbs. Parents are likely to have a hoist at home to assist with this. Assistance will be required with transferring, dressing and some aspects of washing and dressing.

Boys will require help at night whenever they want to pass urine because upper limbs are weak and they are unable to lift a full urine bottle. They are likely to require help turning in bed at night and readjusting their position to get comfortable because of muscular weakness.

Spinal surgery may be performed at this time. There may be a step change increase in care needs at this point because the fused spine limits the range of movement of the upper limbs – for example reaching to wash and to get dressed will be limited.

Once the Forced Vital Capacity (FVC) is reduced, night time respiratory support will be used. This is a form of non-invasive ventilation using a tight fitting face or nose mask. The parent or carer will have to fix this on at night and will need to be available to make adjustments through the night if necessary. Some individuals may have ventilation via tracheostomy at a later stage when ventilation is required for night time and the majority of the daytime. It is common for weight loss to occur in the later stages of the condition and supplemental feeding may be required. Eating becomes more effortful over time and they develop swallowing problems that can lead to choking episodes. Choking risks and difficulties with swallowing may mean that they physically cannot consume enough calories to support themselves and extra nutritional support is required. This commonly includes gastrostomy feeding. Care required includes cleaning and monitoring the gastrostomy site on a daily basis. Attaching the pump feed to the gastrostomy tube

as necessary for feeding and keeping the gastrostomy tube flushed and clear between feeds.

See table - [Disabling effects of Duchenne Muscular Dystrophy related to age](#)

### **Becker Muscular Dystrophy**

This condition is less likely to be diagnosed in early childhood unless it is known to run in the family or is severe. There are unlikely to be any symptoms beyond cramps and limb pains in the early years. Motor milestones may be delayed in some children. The principle effects of the condition will be difficulty running, climbing stairs and getting up hills as easily or as quickly as other children, because of relative muscle weakness. There is likely to be some limitation of long walking distance. Postural problems do not develop, and normal walking ability is maintained throughout childhood in all but the most severe cases. Regular exercise is encouraged as this can increase muscle strength in the early years. A common problem for these boys in the teenage years is muscle cramps after exercise and this may be relieved by intermittent compression therapy boots (flotrons). These are pain relieving and can be worn as required. A teenager could be expected to decide when to use them and to put them on himself.

Reduced walking ability may be present in a severe case and walking aids, typically a scooter, for longer distances may be used.

### **Limb Girdle Muscular Dystrophy**

Needs are very variable ranging from children with severe disability who were diagnosed in infancy who will never walk or independently self care due to muscle weakness, to children with no or minimal disability who have a normal childhood and no care or mobility needs.

### **Facioscapulohumeral muscular dystrophy**

Childhood onset or Infantile Facioscapulohumeral muscular dystrophy is a severe progressive condition with care and mobility needs very similar to those seen in Duchenne Muscular Dystrophy with the exception that cardiomyopathy is rare in this condition. Care and mobility needs related to profound and progressive muscle weakness seen in this condition are

described in detail in the [Duchenne Muscular dystrophy Care and mobility section](#).

Adult onset or mild form of Facioscapulohumeral muscular dystrophy - no care or mobility needs would be anticipated during childhood.

### **Emery-Dreifuss Muscular Dystrophy**

Needs are very variable ranging from children with severe disability who were diagnosed in infancy who will never walk or independently self-care due to muscle weakness, to children with no or minimal disability who have a normal childhood and no care or mobility needs.

Congenital Muscular dystrophy (CMD) with protein laminin-A2 (LAMA2) deficiency – the condition is also known as MDC1A, classic CMD or merosin deficient CMD

Care required from diagnosis until age 5 is likely to consist of extra care and attention with feeding to overcome any feeding or swallowing problems and may include anything from special feeding techniques to gastrostomy feeding. Physiotherapy exercises involving muscle stretching to maintain or encourage normal posture. Splints or braces may be used, and the parent will have to fit these on a daily basis.

Less severely affected children may learn to walk but may not achieve normal walking ability, for example they may be standing and taking a few steps but not walking a useful distance so may be in the virtually unable to walk category. Children that learn to walk a reasonable distance and maintain walking ability are less likely to develop scoliosis and respiratory complications and would have a moderate rather than a severe condition. These children are still likely to require significant care in the form of physiotherapy exercises and help with braces or splints. Children who do not learn to walk or at least stand by around 7 are likely to require help with all aspects of care and may develop scoliosis requiring surgical treatment, respiratory failure requiring ventilation at night or cardiomyopathy.

### **Ullrich Congenital Muscular Dystrophy**

Care required from diagnosis until age 5 is likely to consist of extra care and attention to prevent contractures (shortening) and support normal development despite muscle weakness. Motor development is likely to be severely

delayed. Nighttime respiratory support may be required as early as the 1<sup>st</sup> decade of life.

Physiotherapy exercises involving muscle stretching to maintain or encourage normal posture will be done daily. Splints or braces may be used, and the parent will have to fit these on a daily basis.

Most children will not learn to walk. Less severely affected children may learn to walk a useful distance but will never achieve normal walking ability and require extra care related to their reduced walking distance and difficulties climbing stairs. Walking ability will be lost 2-10 years later and once lost, considerable care and help with activities of daily living is likely including:

- night time respiratory support (always required before the age of 20)
- turning in bed
- swallowing and feeding difficulties (gastrostomy feeding may be used)

Spinal surgery may be required – these children are likely to have care needs broadly similar to children with Duchenne muscular dystrophy.

### **Walker- Warburg Syndrome (a form of congenital muscular dystrophy)**

Care required from diagnosis until age 5 is likely to consist of extra care and attention to feeding to overcome any feeding or swallowing problems and may include anything from special feeding techniques to gastrostomy feeding. Care may be required related to visual impairment and learning disabilities. Physiotherapy exercises involving muscle stretching are necessary to maintain or encourage normal posture. Splints or braces may be used and the parent will have to fit these on a daily basis. Less severely affected children may learn to walk but may not achieve normal walking ability, for example they may be standing and taking a few steps but not walking a useful distance so may be in the virtually unable to walk category.

Children who learn to walk a reasonable distance and maintain walking ability are less likely to develop scoliosis and respiratory complications. These children are still likely to require significant care in the form of physiotherapy exercises and help with braces or splints. They are likely to require extra



supervision related to their learning disability or are likely to require extra help and supervision if they have visual impairment.

Children who do not learn to walk or at least stand by around 7 are likely to require help with all aspects of care and may develop scoliosis requiring

surgical treatment, respiratory failure requiring ventilation at night or cardiomyopathy.

### **Disabling effects of Duchenne Muscular Dystrophy related to age**

Age range	Medical and developmental problems	Care Issues	Mobility Issues
Early years	Delayed motor milestones Speech delay common Learning difficulties common		N/A
Age 4-6 around the time of diagnosis	Unable to run and jump	<ul style="list-style-type: none"> <li>• Daily physiotherapy</li> <li>• Application of night time splints</li> <li>• Non-walking splints at night</li> </ul>	N/A

<p>Years after diagnosis – age 4-12 depending on severity</p>	<p>Increasing mobility problems due to muscle weakness:</p> <ul style="list-style-type: none"> <li>• Falls</li> <li>• Difficulty getting off the floor</li> <li>• Struggling with the stairs</li> <li>• Walking slower</li> <li>• Difficulties climbing in and out of the bath</li> <li>• Reduced walking distance</li> </ul> <p>Also:</p> <ul style="list-style-type: none"> <li>• Risk of injury due to falls</li> <li>• Risk of joint contractures</li> <li>• Steroid drug side effects</li> </ul>	<ul style="list-style-type: none"> <li>• Daily physiotherapy</li> <li>• Application of nighttime splints</li> <li>• Non-walking splints at night</li> <li>• Help with climbing into bath/upstairs/lifting from floor</li> <li>• Wheelchair for long distances</li> <li>• Behaviour management (steroid side effects)</li> <li>• Weight management (steroid side effect)</li> <li>• Extra care after surgery (if required) for joint contractures</li> </ul>	<p>Walking difficulties present</p>
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<p>Increasing mobility problems age 4 - 12 depending on severity</p>	<p>Increasing mobility problems due to muscle weakness:</p> <ul style="list-style-type: none"> <li>• Unable to climb up/downstairs</li> <li>• Unable to get up from floor</li> <li>• Difficulty washing/mobilising round bathroom</li> <li>• Uses wheelchair for longer distances and at school</li> <li>• Walking short distances</li> </ul>	<ul style="list-style-type: none"> <li>• Daily physiotherapy</li> <li>• Application of nighttime splints</li> <li>• Non-walking splints at night</li> <li>• Help with mobilising and washing</li> <li>• Wheelchair daily</li> <li>• Behaviour management (steroid side effects)</li> <li>• Weight management (steroid side effect)</li> <li>• Extra care after surgery (if required) for joint contractures</li> </ul>	<p>Walking difficulties present</p>
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<p>Increasing mobility problems age 8 - 12 depending on severity</p>	<p>No outdoor mobility – uses wheelchair</p> <ul style="list-style-type: none"> <li>• Unable to climb up/downstairs</li> <li>• Unable to get up from floor</li> <li>• Difficulty washing/mobilising round bathroom</li> <li>• Walking functional distance indoors</li> </ul>	<ul style="list-style-type: none"> <li>• Daily physiotherapy</li> <li>• Application of night time splints</li> <li>• Non-walking splints at night</li> <li>• Daytime callipers for exercise</li> <li>• Help with mobilising and washing</li> <li>• Behaviour management (steroid side effects)</li> <li>• Weight management (steroid side effect)</li> <li>• Extra care after surgery (if required) e.g. scoliosis</li> </ul>	<p>Increasing mobility problems present</p>
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<p>95% by age 12 – fully wheelchair dependent</p>	<p>Uses wheelchair indoors and outdoors</p>	<ul style="list-style-type: none"> <li>• Daytime callipers for exercise – help to put on and supervision – risk of falls</li> <li>• Help with transferring</li> <li>• Help with all aspects of washing – likely to have a hoist</li> <li>• Likely to need turning at night</li> <li>• Help with toileting day and night</li> </ul>	<p>Likely inability to walk</p>
<p>Age 12 and over</p>	<p>Uses wheelchair indoors and outdoors</p> <p>Increasing weakness of upper limbs and restricted reach due to spinal surgery or spinal jacket</p>	<ul style="list-style-type: none"> <li>• Help with transferring</li> <li>• Help with all aspects of washing – likely to have a hoist</li> <li>• Likely to need turning at night</li> <li>• Help with toileting day and night</li> </ul>	<p>Inability to walk</p>

<p>Age 12 and over increasing weakness of upper limbs and respiratory muscles</p>	<p>Uses wheelchair indoors and outdoors</p> <p>Increasing weakness of upper limbs and restricted reach due to spinal surgery or spinal jacket</p> <p>Respiratory muscle weakness leading to low blood oxygen levels at night and repeated chest infections</p>	<ul style="list-style-type: none"> <li>• Help with transferring</li> <li>• Help with all aspects of washing – likely to have a hoist</li> <li>• Turning at night</li> <li>• Help with toileting day and night</li> <li>• Help with eating due to restricted reach and lifting or swallowing difficulties</li> <li>• Gastrostomy feeding</li> <li>• Night time respiratory support</li> </ul>	<p>Inability to walk</p>
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## What you need to know about Neuroblastoma



<p><b>What is Neuroblastoma?</b></p> <p>Neuroblastoma is a type of cancer that particularly affects children under 5 years old.....</p> <ul style="list-style-type: none"> <li>• <a href="#">Neuroblastoma</a></li> </ul>
<p><b>What are the effects and signs?</b></p> <p>Symptoms of neuroblastoma will depend on where the tumour is located.....</p> <ul style="list-style-type: none"> <li>• <a href="#">Effects of Neuroblastoma</a></li> </ul>
<p><b>How is it assessed?</b></p> <p>Investigations are likely to include blood tests, bone marrow biopsy.....</p> <ul style="list-style-type: none"> <li>• <a href="#">Assessment of Neuroblastoma</a></li> </ul>
<p><b>How is it treated and managed?</b></p> <p>The low risk group of children consists of children with surgically removable tumours.....</p> <ul style="list-style-type: none"> <li>• <a href="#">Treatment for Neuroblastoma</a></li> </ul>
<p><b>What evidence is available?</b></p> <p>The best source of information will be the hospital based specialist nurse or the consultant oncologist. ....</p> <ul style="list-style-type: none"> <li>• <a href="#">Evidence</a></li> </ul>
<p><b>How long will the needs last?</b></p> <ul style="list-style-type: none"> <li>• <a href="#">Prognosis and duration of the award</a></li> </ul>

## What is Neuroblastoma?

Neuroblastoma is a type of cancer that particularly affects children under 5 years old. It is the second most common 'solid' tumour in this age group after brain tumours and the most common cause of death from cancer in this age group. Neuroblastomas develop from cells called pluripotent stem cells that migrate within the developing foetus to form the sympathetic nervous system. They develop in areas of the body where sympathetic nerves are found with the commonest site being the abdomen, as it has two structures,

which contain a lot of sympathetic nerves. Abdominal neuroblastomas most commonly occur in:

- Adrenal glands - a pair of glands, one on top of each kidney – 40%
- Para-spinal sympathetic chain – a chain of sympathetic nerve bodies running down the back wall of the abdominal cavity – 25%. Para-spinal tumours grow near and may affect the spinal cord

Many other sites in the body contain sympathetic nerves and these can give rise to neuroblastoma tumours. These sites include:

- Thorax (chest cavity) – 15%
- Pelvis – 5%
- Neck – 3%
- Other sites – 12 %

Neuroblastomas usually produce neurotransmitter substances (also known as catecholamines) in greater than normal quantities. The breakdown products of these can be measured in the urine and is one way of testing for neuroblastoma. In the majority of cases of neuroblastoma, the cause is unknown. In very rare cases, other family members have been affected by neuroblastoma and these families have a genetic condition predisposing children to develop neuroblastoma.

Children may not have symptoms from their tumour or they may simply feel generally unwell. The parent may notice a lump or swelling, abdominal swelling is a common reason for the child to have been taken to the doctor in neuroblastoma.

Age, stage and biological features of the tumour strongly influence survival. Children under 18 months with early stage disease do best. Children with low risk and intermediate risk neuroblastoma have an excellent prognosis and outcome, with 3 year survival rates of 88% to 96% reported in one study. Children with high risk neuroblastoma do not do well.

There are several staging systems for this type of tumour. The stage and other information about the child and the tumour are used to stratify individual children into very low, low, intermediate or high risk groups. The risk classification is used as the basis for treatment decisions. For example, a child in the low risk group may have their neuroblastoma removed surgically and recover completely within 3 months developing no care or mobility needs. A child in the intermediate group may need surgery followed by 4-8 cycles of chemotherapy with treatment related needs throughout the period, making a complete recovery. A child in the high risk group may undergo

high dose chemotherapy and a stem cell transplant taking two years to recover. Stratification in to risk groups is complex and based on multiple factors.

Most recent data from the National Registry of Childhood Tumours shows 5 year survival rates for all risk groups together are 67%, being much improved from 18% survival rate reported for the period 1962-66. There is no screening programme for neuroblastoma in the UK.

### **What is the incidence/prevalence?**

Around 80 children develop neuroblastoma in the UK each year. 40% of these children will be under 1 year, 35% aged 1 to 2 years and 25% over 2 years when diagnosed. Neuroblastoma is very rare beyond age 10 years.

### **What are the effects and signs?**

Symptoms of neuroblastoma will depend on where the tumour is located. Symptoms may not be present at all, or they may be vague. Vague symptoms reported in neuroblastoma include tiredness, loss of appetite, fevers and non-specific pain. Symptoms related directly to the tumour will depend on where it is. Abdominal tumours may cause:

- Abdominal swelling
- Abdominal pain or discomfort
- Spinal cord compression, causing leg weakness

Neuroblastomas in the chest may cause:

- Breathing difficulties
- Chest pain
- Secondary chest infection
- Spinal cord compression, causing leg weakness

Tumours in the neck may cause:

- Swelling in the neck
- Difficulty breathing
- Spinal cord compression causing arm and leg weakness

Neuroblastoma can invade the bone marrow and cause bone marrow failure - this means that the bone marrow cannot do its job of producing enough

red blood cells, white blood cells or platelets to replenish the blood. Symptoms include:

- Anaemia due to low red cell count
- Increased number or severity of infections due to reduced white cell production
- Easy bruising or bleeding due to reduced platelets

Neuroblastoma often spreads to bones (as well as bone marrow) and causes symptoms of:

- Bone pain
- Limp
- Lump on bone
- Eye swelling if bones around eyes involved

Children with neuroblastoma can have high blood pressure at diagnosis because:

- The tumour causes pressure on the kidney
- Production of catecholamines by the neuroblastoma

Para-spinal neuroblastomas may grow into the spinal cord and cause compression of the spinal cord, which can cause difficulty walking and bladder and bowel problems. These tumours often need to be treated urgently with chemotherapy or surgery to try and prevent progression of symptoms.

Recovery of spinal cord function is variable, but generally children recover better if treatment begins soon after symptoms first develop.

Indicators of severe functional restriction

- Para-spinal tumour with neurological symptoms or signs - walking and continence may be affected
- The child is in the high-risk group
- undergoing stem cell transplant
- Gastrostomy feeding
- [General care & mobility considerations](#)

### **How is it assessed?**

Investigations are likely to include the following:

Blood tests

- Full blood count (FBC).
- Liver function.
- Urea and electrolytes.
- Calcium.
- Lactate dehydrogenase (LDH).
- Serum Ferritin.

Bone marrow biopsy (to confirm or exclude bone marrow involvement)

Urine tests

- Spot urine collection for catecholamines and its metabolites (e.g. vanillyl mandelic acid or VMA) as these are usually produced by neuroblastoma cells and can be measured in the urine

Imaging studies are very important and include:

- CT scan
- MRI scan
- Metaiodobenzylguanidine (MIBG) scan – MIBG is injected into the blood stream, is picked up by neuroblastoma cells and can

show up the location of the primary tumour and any secondary sites

Tumour biopsy is always performed unless the tumour can easily be removed, in which case initial resection of the tumour is performed. Tumour material is examined for genetic abnormalities; these can help in treatment planning by putting children in higher or lower risk groups. The genetic tests most commonly used are:

- MYCN amplification status – There are 2 copies of the MYC gene in normal cells, but in neuroblastoma cells there can be multiple copies of the MYC gene. This is termed MYCN amplification and is detectable in tumour material. MYCN amplification only occurs in children with high risk disease
- Ploidy – this term describes the number of chromosomes in the neuroblastoma cell nucleus. Diploid is normal and means there are two sets of chromosomes in each cell. Each chromosome is paired with another, making 46 chromosomes in 23 pairs. Triploid means there are three of each chromosome and hyperdiploidy means there are multiple sets of chromosomes in each cell. Hyperdiploid tumours are associated with a better prognosis

Appearance of cells under the microscope – histology – ‘Shimada classification’ - is an assessment of the appearance of the neuroblastoma cells under the microscope. The appearance of the cells in conjunction with the child’s age correlates with outcome; some tumours look more malignant than others. Shimada classification divides children in to two groups: favourable and unfavourable and this classification based on appearance correlates with prognosis

- [Staging of disease](#)
- [Risk factors](#)

## **How is it treated & managed?**

### **Overview**

Treatment of children with Neuroblastoma will be at regional paediatric oncology principal treatment centres that may be some distance from the child’s home. Treatment will be based on the risk group the child is in. Types and duration of treatments described in this guidance are presented in the same format. If medical evidence does not say what risk group the child is in, follow the high risk guidance for children undergoing stem cell transplant, the intermediate risk guidance for children undergoing

chemotherapy but no stem cell transplant and low risk for children who appear to be having surgery as their only treatment.

### **Low risk group treatment strategy**

The low risk group of children consists of children with surgically removable tumours and most children with stage 4S or MS disease, note that 4S and MS refer to the same stage, it depends which staging system is reported in the medical evidence.

#### Children with surgically resectable tumours

These children are likely to undergo surgical removal of their tumour. Recovery from surgery will depend on the site of the tumour. Recovery in the typical case will be well within 6 months. In most cases adjuvant therapy (add-on treatment) is not required. Even if the disease recurs and chemotherapy is required to treat recurrence, survival is around 95%. Note that a small number of children with surgically resectable small tumours will be in the high risk group because they have amplified MYC – refer to the high risk section for details.

#### Babies with 4S or MS disease

Babies in this category tend to experience spontaneous regression; this means the tumour goes away without treatment. Many of these children will have no or minimal symptoms. Occasionally, the site of the tumour can cause difficulties, for example if the liver becomes very large due to tumour infiltration and impedes breathing. Such babies will require chemotherapy treatment. A typical treatment regimen would include 2-4 courses of intravenous chemotherapy given at 3 weekly intervals and each course would take 3-5 days to administer. Survival in babies who require treatment is around 80%. Treatment and recovery from treatment occur within 6 months and recovery with long term survival is the norm.

### **Intermediate risk group treatment strategy**

Children in this group are likely to have stage 3 or 4 disease (by INSS) or stage L2 or M by INRGSS, this means that complete surgical removal is not possible. Treatment plans for these children begin with chemotherapy to reduce the size of the tumour, followed by surgery to remove as much of the tumour as possible. Some children then go on to have adjuvant chemotherapy. A minority have radiotherapy treatment to the tumour bed. It is not clear in this group of children how much chemotherapy is required and children may be entered in to clinical trials designed to answer this question. Children are likely to receive 2 to 6 cycles of chemotherapy over 6 to 18 weeks

respectively. Chemotherapy is likely to consist of moderate doses of the following drugs:

- Carboplatin
- Cyclophosphamide
- Doxorubicin
- Etoposide
- Vincristine

Rarely children may have radiotherapy to the affected organs. This is usually given once a day for 14 days under general anaesthetic.

Children under 1 have a greater than 80% cure rate in this group; older children have a 50 to 70% cure rate.

It is clear that duration of treatment and recovery are variable within the intermediate risk group. Some children will undergo surgery and complete chemotherapy within 4 months; others may complete treatment in 7-8 months and spend longer recovering having had more intensive more prolonged treatment. Total duration of treatment and recovery may be up to a year in this group.

### **High risk group treatment strategy**

This group of children have MYCN amplification and/or metastatic disease. There is often widespread disease, affecting a number of organs. This group are likely to have larger tumours that are difficult or impossible to remove completely with surgery. They will undergo intensive treatment usually in the following order:

- Intensive chemotherapy
- Surgery
- High dose chemotherapy and stem cell transplant.
- Radiotherapy to the tumour site
- Retinoic Acid and Immunotherapy

Total duration of treatment is variable depending on amount and type of chemotherapy treatment used. Treatment takes a minimum of 12 months, but could take up to 18 months with 6-12 months of recovery time



afterwards. More details about the treatments used in this group are provided below:

### Induction Chemotherapy

Children are likely to begin treatment with high dose chemotherapy. The chemotherapy is given every 10 days for 8 cycles over 10 - 12 weeks with assessment of response during and at the end of treatment. A combination of the following drugs is likely to be used:

- Vincristine
- Cyclophosphamide
- Cisplatin
- Etoposide
- Carboplatin

Significant side effects are likely. All chemotherapy will be given in hospital as intravenous infusions, with most cycles requiring at least one overnight stay. Children may be receiving daily subcutaneous injections of G-CSF, which is likely to be given at home by the parent/carer.

### Further Chemotherapy

If children do not show sufficient response to this initial induction treatment then they will receive further chemotherapy with the following drugs:

- Vincristine
- Doxorubicin
- Topotecan

### Surgery

Surgery is likely to follow induction chemotherapy, although it may be delayed for further chemotherapy (see above) if response is slow. Neuroblastoma can occur in various sites, although the abdomen is the most common. The aim of surgery is for complete tumour resection whenever possible. This can mean complex and extensive operations in some cases. Post-operative hospital stay is variable 1-3 weeks.

### Stem cell transplant

Children will undergo autologous stem cell transplant, which means the patient's own cells are used. This is usually undertaken after surgery. Stem cell transplant involves high dose myeloablative chemotherapy to kill off the

bone marrow and neuroblastoma cells remaining in the body. The risks and side effects of this treatment are significant.

### [Peripheral Blood Stem Cell Transplant \(PBSCT\) or Bone Marrow Transplant](#)

#### Radiotherapy

Children usually have external beam radiotherapy after stem cell transplant. Radiotherapy is given daily for 14 days and patients usually require general anaesthetics on a daily basis for this part of the treatment.

#### Therapy after stem cell transplant

Children are likely to continue oral drug therapy for 6 months following their transplant. This consists of a tablet, 13-cis-retinoic acid (Accutane), taken twice daily for 14 days in 28 day cycles. Treatment with this drug improves survival in this group. It is a Vitamin A derivative that may cause liver enzyme dysfunction, muscle cramps, dry skin or neurological symptoms. It does not inhibit recovery from chemotherapy in terms of hair regrowth or return of energy.

Most patients will also receive immunotherapy at the same time as 13-cis-retinoic acid and this is given as part of the main European High Risk Neuroblastoma clinical trial. There are 2 components to the immunotherapy:

- Anti-GD2 – this is a neuroblastoma antibody given for 5 days by intra-venous infusion as an in-patient. Five courses are given at 28 day intervals and therefore this treatment finishes one month prior to completing the 13-cis-retinoic acid treatment. This treatment causes significant pain and patients need to be on intra-venous morphine treatment at the same time
- Interleukin-2 (Aldesleukin) - only 50% of patients receiving the anti-GD2 will also receive IL-2 and this is decided in a random fashion as part of the clinical trial protocol. This treatment involves a daily subcutaneous injection for 14 days every month and this will mean a daily visit to hospital. The treatment can cause significant fever. Five courses of the treatment are given and it will be complete at the same time as the anti-GD2
- 

#### Relapsed or refractory group treatment strategy

In a number of children the condition will relapse or will not respond completely to treatment. This is an extremely difficult group to treat and the outlook for these children is poor. Recently, however, a number of new

treatments have gone into development and are undergoing trials. Children receiving any of these are likely to be having frequent treatment and monitoring in a specialist centre. It would be expected that confirmatory

evidence would be available from the treating consultant or an oncology specialist nurse.

[Side effects of treatment](#)

[Side effects of chemotherapy](#)

[Problems in adults who had cancer treatment as children](#)

**What evidence is available?**

<b>Sources of Evidence</b>	<b>Information Provided</b>	<b>Limitation of Evidence</b>
Claimant/carer	First source of information	May not be objective, does not have specialist knowledge
GPFR	The GP should be able to confirm the diagnosis and provide contact details for the paediatric oncologist managing the child's neuroblastoma.	All children will be managed at their local paediatric oncology centre. The GP will be able to confirm the diagnosis but may not be able to give details of current or planned treatment or prognosis of disabling effects.

<p>Hospital FR</p>	<p>The best source of information will be the hospital based specialist nurse or the consultant oncologist. They will be able to provide details of the risk group the child is in and current and planned treatment and confirm disabling effects. They are the best source of information on duration of planned treatment and prognosis of disabling effects.</p> <p>The most important information to check will be:</p> <p>Confirm diagnosis</p> <p>Confirm treatment is being received e.g. some type of chemotherapy. Some children will not need treatment.</p> <p>Confirm severe disability due to specific side effects of therapy if claimed e.g. rare disabling side effects of chemotherapy or effects of disease such as walking difficulties due to paraspinal neuroblastoma. Always ask for prognosis of side effects.</p>	<p>Details of planned treatment may change depending on response to treatment or the development of side effects, information provided may become quickly out of date.</p>
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### How long will the needs last?

#### Low risk group

Needs are not anticipated in this group. Full recovery is expected even if recurrent disease occurs. The exception to this is children with neurological symptoms who have difficulty walking or continence problems. Two year awards are recommended for these complications of the neuroblastoma. At

two years if medical evidence shows mobility or continence problems persist, an indefinite award is recommended.

### **Intermediate Risk Group**

This group may undergo chemotherapy and surgery and may have additional care needs related to the treatment and its side effects for 4 to 8 months. This will depend on the duration and type of treatment planned. If needs related to treatment are identified, a 1 year award is recommended. If needs related to neurological problems are claimed, a two year award is recommended in relation to these. At two years if medical evidence shows mobility or continence problems persist, an indefinite award is recommended. In the majority of cases a full recovery is expected.

### **High risk group and children who plan to have stem cell transplant**

Children in the high risk group will undergo intensive treatment including surgery, radiotherapy, chemotherapy, high dose chemotherapy and stem cell transplant and finally 13-cis-retinoic acid and immunotherapy. Children in this group are likely to develop care needs related to administering treatment and coping with side effects. In particular they will require care and help to cope with the physical side effects of chemotherapy and to prevent infection during periods of immunosuppression.

Children in this group are at increased risk of recurrent disease and may undergo further treatment including chemotherapy for recurrence when this occurs.

A 4 year award is recommended for children in this group. At 4 years, prognosis becomes clear, children who will recover will have done so and any enduring disabling effects of the disease or treatment will be stable. If needs persist at 4 years of age, an indefinite award is recommended in this group.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **General care and mobility considerations**

#### **Care**

Some children with neuroblastoma will develop care needs due to the nature and duration of their treatment. For example children with low risk and some with intermediate risk disease could be expected to make a full recovery within 6 months. Other children undergoing chemotherapy for

intermediate risk neuroblastoma may have care needs related to the duration of their chemotherapy regimen. Children undergoing treatment for high risk neuroblastoma are likely to have significant care needs. Older children undergoing treatment are likely to have more care needs than younger children for two reasons -:

- Firstly they are likely to have more toxic treatments, being in the high risk group because of their age. This means a greater proportion of children over 18 months could be expected to have severe side effects related to therapy because they are having more intensive treatment.
- Secondly children aged 5 and above are likely to be substantially self caring - any help with feeding, toileting and mobility in this age group is likely to be related to their condition rather than immaturity.

Children may be immunosuppressed and unable to attend school for prolonged periods requiring care and protection from infection at home. Pre-school children, who normally attend nursery, will need to be cared for in a safe environment rather than nursery to reduce risk of infection.

Children of all ages in any risk group are likely to require extra emotional support, and practical help from their parents related to both treatment and the disease. In relation to treatment parents will need to spend time doing the following for any child undergoing chemotherapy treatment -:

- Supporting their child through painful or distressing treatment.
- Pain management - either chronic, disease related or acute, treatment related.
- Ensuring any oral drug treatments are taken as prescribed, despite side effects.
- Caring for a central venous access device/central line - in most cases a 'Hickman' type line – the site must be kept clean and



the line regularly flushed, care must be taken not to dislodge the line when washing and dressing.

- Monitoring their child for the side effects of treatment – this includes close monitoring for signs of infection and easy bruising or bleeding.
- Protecting their child from infection during periods of immunosuppression
- Encouraging their child to eat during periods of stomatitis (sore dry mouth) and providing mouth care.
- Providing an appropriate diet for the child when immunosuppressed.
- Many children will require supplemental or complete enteral feeding during their treatment; this may be either via a nasogastric tube, or a gastrostomy.
- Some parents will give subcutaneous drug treatments to the child.
- Emotionally supporting their child through their illness e.g. dealing with hair loss/time away from school/ being different to peers.

Following stem cell transplant their child is likely to have periods of being immunosuppressed and be unable to go out in public. Episodes of severe fatigue may endure for many months related to chemotherapy treatment and anaemia. Younger children will require help with all aspects of self care and dressing because of their age. Older children may also require such help due to debilitation and severe fatigue.

Some children will require additional care -:

- Peripheral neuropathy related to chemotherapy (particularly vincristine). This can cause 'foot drop', which can limit mobility or numbness or tingling, which may make using the hands

difficult due to numbness. This is rare but may affect personal care, toileting ability, feeding and dressing.

- Para-spinal neuroblastoma that has caused neurological problems in the legs and affected continence.
- Terminal illness.

For some children treatment will not be successful, and they will require palliative and subsequent end of life care

### **Mobility**

Severe fatigue related to chemotherapy treatment and anaemia may affect walking for periods during treatment but this is not a continual effect over 6 months or more. Children with para-spinal tumours may have neurological problems making walking difficult or impossible due to paralysis. Some children will recover walking ability and others will not. If the condition is reported as permanent by the treating oncologist indefinite awards in respect of mobility are recommended. In other cases where ability to recover is unknown review after 2 years, 2 years is recommended as significant recovery, including learning to walk again, may occur. If walking is not achieved in the first two years indefinite awards are recommended.

A small number of children may also experience peripheral neuropathy as a side effect of chemotherapy which can also affect walking. Full recovery is expected in most of these cases, the best source of evidence on mobility problems and ability to recover is the treating oncologist or specialist nurse.

### **Staging of disease**

There are two main staging systems for neuroblastoma -:

- International Neuroblastoma Staging system (INSS), which is a similar staging system to that used in other cancers, with stages 1-4 (and then a special 4S).
- International Neuroblastoma Risk Group Staging System (INRGSS) is a more recently introduced staging system that describes whether the patient has a tumour which can be removed by surgery only (L1) or whether the patient needs chemotherapy initially (L2), as well as having more distant disease (M or MS).

The table below shows details of how patients are staged within the two systems. The two staging systems are different and there is now a gradual move from the old INSS system to the newer INRGSS. INSS stage was previously assigned after surgery but the newer INRGSS tries to determine on CT and MRI appearances which tumours should be resected and which

should be biopsied based on surgical risk factors. Therefore stage 1 and 2 tumours by INSS do not necessarily correlate directly with L1 tumours on INRGSS.

Staging terms used in the INSS and INGRSS staging system classifications for risk stratification in neuroblastoma

INSS Stage	INRGSS Stage
<p>Stages 1 and 2</p> <p>Stage 1: The tumour is localised and can be completely removed with surgery.</p> <p>Stage 2: The tumour has not spread and is on only one side of the body but cannot be completely removed by surgery because of its size or position.</p> <p>Stage 2A: No lymph node spread. Stage 2B: Lymph node spread is present.</p>	<p>Stage L1</p> <p>L1: Surgical removal looks straightforward.</p>
<p>Stage 3: The tumour has spread to nearby organs close to where it started. Lymph nodes may or may not be involved.</p>	<p>Stage L2: Surgery is possible but difficult i.e. there are 'surgical risk factors' present.</p>
<p>Stage 4: The cancer has spread to other parts of the body that are some distance from the primary tumour</p>	<p>Stage M: The cancer has spread to other parts of the body that are some distance from the primary tumour</p>
<p>Stage 4S: This is a special case and has a better outlook than other stages. It means the child is under 12 months old and has neuroblastoma that has spread to the liver or skin but not the bones. Less than 10% of the bone marrow cells are cancer cells. Some of these children may not need any treatment because the cancer can disappear or 'regress' on its own.</p>	<p>Stage MS: This is a special case and has a better outlook than other stages. It means the child is under 12 months old and has neuroblastoma that has spread to the liver or skin but not the bones. Less than 10% of the bone marrow cells are cancer cells. Some of these children may not need any treatment because the cancer can disappear or 'regress' on its own.</p>

### Risk factors

Risk group classification in neuroblastoma is an important step that determines treatment and prognosis.

There is now an International Neuroblastoma Risk Group Classification System that combines 7 risk factors to place patients into low, intermediate or high risk groups. Treatment is planned based on risk group. In the medical evidence, the risk group may be clear, or the risk group may be inferred from the treatment the child is having. Information about the factors used to place children in risk groups is provided. Information is provided to aid understanding of terms used in medical evidence and should not be used to classify individual cases in to risk groups.

The risk factors used to determine the appropriate risk group are -:

- Stage of disease - as defined by the INRGSS, generally those with L1 have a good prognosis and those with metastatic disease (M) have a poorer prognosis. There is a special group of those under the age of one with low level metastatic disease (MS) who have a good prognosis. The stages may be described by the older INSS as stage 1-4 or 4S.
- Age of the child - age is a factor that affects outcome, younger children tend to do better no matter what treatment they have

and older children tend to do worse no matter what treatment they have.

- Features of the tumour -:

Genetic markers - present in the tumour including MYCN amplification, ploidy and 11q deletion.

Histological category - there are various sub-types of neuroblastoma which have particular histological appearances and this can be relevant to prognosis.

Grade of tumour - Neuroblastomas are graded on the basis of "how malignant" the cells appear.

The table below shows how these known risk factors can be used to assign all patients as low, intermediate or high risk. There is also a very low risk group.

The table also shows how the risk factors act with one another using INRGSS to classify children into risk groups using various factors.

Note: In the table below:

The acronym (GN) refers to Ganglioneuroma

The acronym (GNB) refers to Ganglioneuroblastoma

The term (MYCN) refers to genetic abnormality of the tumour cells

The term (11q aberration) refers to (genetic abnormality of the tumour cells)

<b>INRGSS stage</b>	<b>Age</b>	<b>Histologic category</b>	<b>Grade of tumour</b>	<b>MYCN</b>	<b>11q</b>	<b>Ploidy</b>
L1/L2	N/A	GN maturing; GNB intermixed	N/A	N/A	N/A	N/A
L1	Less than 18 months	Any except GN maturing and GNB intermixed	N/A	No amplification	N/A	N/A
L1	Less than 18 months	Any except GN maturing and GNB intermixed	N/A	Amplification	N/A	N/A
L2	Less than 18 months	Any except GN maturing and GNB intermixed	N/A	No amplification	No	N/A
L2	Less than 18 months	Any except GN maturing and GNB intermixed	N/A	No amplification	Yes	N/A
L2	Over 18 months	GNB nodular; neuroblastoma	Differentiating	No amplification	No	N/A
L2	Over 18 months	GNB nodular; neuroblastoma	Differentiating	No amplification	Yes	N/A
L2	Over 18 months	GNB nodular; neuroblastoma	Poorly differentiated or undifferentiated	N/A	N/A	N/A
L2	Over 18 months	GNB nodular; neuroblastoma	N/A	Amplification	N/A	N/A
M	Less than 18 months	N/A	N/A	No amplification	N/A	Hyperdiploid
M	Less than 12 months	N/A	N/A	No amplification	N/A	Diploid

M	12 to 18 months	N/A	N/A	No amplification	N/A	Diploid
M	Less than 18 months	N/A	N/A	Amplification	N/A	N/A
M	Over 18 months	N/A	N/A	N/A	N/A	N/A
MS	N/A	N/A	N/A	N/A	No	N/A
MS	N/A	N/A	N/A	No amplification	Yes	N/A
MS	Over 18 months	N/A	N/A	Amplification	N/A	N/A

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### **Peripheral Blood Stem Cell Transplant (PBSCT) or Bone Marrow Transplant**

A Peripheral Blood Stem Cell Transplant (PBSCT) or Bone Marrow Transplant is a way of transplanting a new immune system into a person's body.



The immune system consists of stem cells that make all the different kinds of blood cells that perform the functions of the immune system, these are -:

- Red blood cells – these carry oxygen around the body
- White blood cells – these cells travel around the body destroying bacteria, viruses and abnormal cells in the body such as cancer cells
- Platelets – these cells help the blood to clot

A transplant involves destroying the person's own immune system and stem cells and replacing them with either -:

- Their own previously harvested stock of stem cells or bone marrow – this is called an **autologous** transplant because a person's own cells are used.
- Someone else's stem cells or bone marrow – this is called an **allogeneic** transplant because someone else's cells are used.

The donor usually has the same or similar genetic markers to the recipient. Checking for compatibility between donor and recipient is called '**matching**'. The other person can be a close relative such as a brother or sister – a '**sibling donor**' or someone who is unrelated – this is called a '**matched unrelated donor**'. The name of the procedure depends on how the stem cells are collected from the donor although the principles of the procedures are the same and the effect on the recipient is the same:

### **Peripheral Blood Stem Cell Transplant (PBSCT)**

Peripheral blood stem cells are harvested from the blood. Growth factors are given to the donor to promote production of larger quantities than normal of stem cells for harvesting. These spill out into the peripheral blood from the bone marrow. The stem cells are filtered from the blood using a procedure similar to blood donation. No anaesthetic is required and the process takes several hours. The side effects for the recipient are the same whether bone marrow or peripheral blood stem cells are used.

### **Bone Marrow Transplant**

Bone marrow is harvested from bone marrow in the pelvis during a short operation. The side effects of bone marrow harvesting are minimal – for example there may be soreness around the pelvis for a week or so afterwards.

The side effects for the recipient are similar whether bone marrow or peripheral blood stem cells are used.

### **How is a transplant given?**

In preparation for transplant a person's immune system will be wiped out by high dose chemotherapy and sometimes radiotherapy treatment. This often has the effect of killing off residual cancer cells and is the aim of the treatment. Once this treatment is complete the bone marrow or stem cells can be given via an intravenous drip. The stem cells do not start working straight away and can take up to a year to start working effectively and really protecting a person from infection.

The side effects of such high dose treatment are severe. A person is not able to produce their own red, white blood cells or platelets and the transplanted tissue is unable to do this for some time either. This means they need regular support with blood and platelet transfusions to prevent bleeding and control anaemia. Transfusions of white cells to control infection cannot be given and so despite antibiotics they are at very high risk of life threatening infection with normally innocuous bacteria or viruses. When a person is unable to produce their own white blood cells to fight infection this is called 'neutropenia'. The high risk period after transplant when a person is very vulnerable to infection is called the '**neutropenic period**' – the neutropenic period is usually spent in hospital.

Once some improvement has occurred and the immune system has started to work to a degree they will be discharged from hospital. The person undergoing treatment is likely to feel extremely ill and weak and may sleep for most of the time. They will have many of the side effects of chemotherapy.

Often the first phase of treatment will be given in isolation in hospital – isolation means just that, a person is kept in one room with minimal visitors who all wear barrier clothing to prevent the transfer of infection. The psychological effects of isolation can be severe. The time spent in isolation in the recent past was 1-2 months. This time has been reduced with more of the recovery time spent at home. The same precautions may be necessary at home as would be taken in an isolation room in hospital. These may be troublesome for carers in terms of cleanliness of the home, providing safe food, preventing contact with potentially harmful everyday items and restricting access to people who are themselves unwell. In addition frequent

blood tests are needed to monitor progress and this is likely to involve accompanying their family member to hospital and back.

### **Side effects of treatment**

#### **Intermediate and high risk groups**

Late side effects after treatment

Children surviving longer than 5 years are still at risk of recurrent disease as late recurrences do occur. They may experience ongoing side effects of cancer treatment into adult life because of the intensive treatment they have had. A number of treated children may have the following on-going problems after their cancer treatment -:

- Heart problems.
- Kidney dysfunction or failure.
- Hearing problems.
- Fertility problems.
- Reduced bone growth (if radiotherapy has been used).
- Increased risks of other cancers.
- Residual neurological defects if neuroblastoma was para-spinal, including weakness in the legs, limping, paralysis or

bladder and bowel problems, including incontinence due to nerve compression.

### **High risk group**

This group undergo intensive and prolonged therapy. During chemotherapy and stem cell transplant typical side effects are -:

- Nausea.
- Diarrhoea.
- Stomatitis (inflammation of the mucous lining of any of the structures in the mouth) and mucositis (any inflammation of a mucous membrane).
- Weight loss and nutritional insufficiency.
- Hair loss.
- Increased risk of infection, which can be life threatening.
- Bruising and bleeding requiring blood product support.
- Extreme tiredness and debilitation.

There will be periods of immunosuppression where the child is at high risk of infection and will be unable to go out in public or see other children. Treatment and full recovery including return to school full time in this group is likely to take up to 1 to 2 years. 5 year survival in this group is 30 to 50%, this means despite having undergone intensive treatment many children will experience recurrent disease and the majority of these recurrences and deaths occur in the 4 years following diagnosis.

### [Side effects of chemotherapy](#)

### [Problems in adults who had cancer treatment as children](#)

### **Side effects of Chemotherapy**

Side effects of chemotherapy are well known and are mostly related to the effect of chemotherapy drugs on normal dividing cells. Some cells in the body divide more rapidly than others because they have to constantly replace themselves for the body to function normally. Blood cells are a good example of this. The bone marrow produces white blood cells, which fight infection. These cells are produced by stem cells, which divide frequently and white blood cells only live for a short time, they can easily fall to low

levels during chemotherapy treatment. This is important because when the white blood cells are low this allows the possibility of overwhelming and sometimes fatal infection. Having a low white count is called 'neutropenia'.

The cells, which line the bowel also rapidly divide and are sloughed off by the passage of food and liquid. When these are not replaced quickly enough there is ulceration of the mouth, throat and bowel. Ulceration lower down in the intestines can lead to bleeding, nausea, abdominal pain and diarrhoea. It is these effects on normal cells, which limit the amount of chemotherapy that can be given.

### **Main short term side effects during Chemotherapy**

- Fatigue - is the main physical side effect of treatment with chemotherapy. In the days after treatment it is usual for the patient to feel like they are developing a flu-like illness. The acute effects will resolve but feelings of tiredness may persist throughout the treatment cycle and can be very debilitating.
- Hair loss - is common and may cause significant distress.
- Nausea and Vomiting - the majority of chemotherapeutic drugs cause nausea and vomiting of varying degrees. The treatment of this has dramatically improved in recent years and it can usually be effectively managed with anti-emetic drugs.
- Psychological effects - because the physical symptoms are better controlled, the emotional side effects of having cancer and receiving chemotherapy have become more important. It is impossible to separate out the side effects of the treatment and the side effects of having cancer. General tiredness, the strain of being diagnosed with cancer, undergoing tiring treatment and the burdens this imposes on the family are all important. Worries about the future are common.

Another common cause of anxiety about chemotherapy in the past was fear of needles and the discomfort of treatment; this has been addressed by the

use of local anaesthetic creams before putting needles in and to an extent by the increased use of indwelling ports and catheters.

Other commonly reported short term side effects

- Giddiness on standing up
- Diarrhoea
- Weight gain
- Shortness of breath

Short term side effects resolve once treatment is complete.

### **Long term disabling effects of chemotherapy treatment**

Peripheral neuropathy (neuritis, plexopathy) - is a toxic effect on nerves, which prevents nerves from working properly.

- Motor nerves – these control movement of muscles, damage may lead to clumsiness or in severe cases paralysis of muscles supplied by the affected nerves. Some recovery may occur once treatment is stopped but the changes are usually slow to improve or permanent.
- Sensory nerves – these nerves enable the sensation of touch, damage to them results in numb areas or areas of pins and needles. In some cases pain fibres are affected and this can lead to pain syndromes.

Abnormal sensations can make simple activities like making a cup of tea, fastening a button or walking difficult, impossible or painful and distressing.

- Breathlessness - has many causes including lung problems and heart failure, which may have been caused by the

chemotherapy or radiotherapy. Lung damage or heart problems caused by certain drugs may be irreversible or progressive.

- Leukaemia - is more common in children who have had chemotherapy or radiotherapy treatment.
- Infertility – chemotherapy and radiotherapy can cause infertility.

These side effects are rare.

### **Problems in adults who had cancer treatment as children**

People in this category have much longer to develop the long term or enduring side effects of chemo and radiotherapy. The oldest members of this group will have had their treatment in the 1970s. What will happen to them as they age is unknown. Some childhood survivors have already developed significant enduring problems because of their treatment, either during treatment or some years later. The number of adults 'at risk' in this category is set to rise.

Cancer therapy, in particular chemotherapy made great progress in the 1980s and 1990s and for the first-time high rates of cure were achieved in some of the common childhood malignancies such as leukaemia and lymphoma. Over time treatment has been modified to become as effective as possible with as few side effects as possible. Significant long term side effects of treatment given in the past are increasingly being recognised. These side effects generally occur because of changes in normal tissue caused by the treatment, these changes take many years to cause symptoms or become apparent. The medical profession is still in the early days of recognising and researching these disorders.

Over time more members of this group can expect to either develop these problems or have them recognised. A breakdown of problems is provided by treatment. Effects tend to be greater when treatment of cancer began at a young age (under 3) and when large doses of chemotherapy and

radiotherapy were necessary. Common cancers in children include leukaemia, lymphoma, brain tumours, bone and soft tissue sarcomas.

Type of cancer treatment	Disabling effect
Cranial irradiation esp. if combined with intrathecal chemotherapy	Neurocognitive defects – reduced IQ, attention deficit, poorer motor/verbal skills, may be severe enough for a Statement of Education Needs (SEN), deafness, epilepsy.
Cranial irradiation, effects are worse if this was combined with abdominal radiation	Hormonal effects including growth impairment in childhood, hypothyroidism, increased risk of infertility, early menopause.
Cranial irradiation	Obesity and its disabling effects.
Chemotherapy or radiotherapy	Increased risk of infertility.
Chemotherapy esp. anthracycline doxorubicin	Heart problems including heart failure, myocardial infarction, arrhythmias and sudden death at young age.
Radiotherapy to chest	Lung problems – breathlessness.
Steroids, methotrexate, inactivity due to illness	Osteopaenia/Osteoporosis.
Chemotherapy or radiotherapy	Second cancers especially brain tumour.
Radiotherapy to abdomen (bladder/bowel/liver)	Chronic diarrhoea, malabsorption, bladder problems, kidney problems including rarely kidney failure.

## What you need to know about Nocturnal Enuresis



### **What is Nocturnal Enuresis?**

Nocturnal enuresis is the medical term for bedwetting. The term enuresis is used to describe children who do not have voluntary control over their bladder.

- [Nocturnal Enuresis](#)

### **What are the effects and signs?**

Children are usually dry at night by 5 years. Because achieving dryness is variable and many children who are bedwetting at 5 and 6 will master control without medical intervention, diagnosis of nocturnal enuresis is not made until age 7

- [Effects of Nocturnal Enuresis](#)

### **How is it assessed?**

A careful history and physical examination will exclude most physical causes of enuresis such as .....

- [Assessment of Nocturnal Enuresis](#)

### **How is it treated and managed?**

The majority of children aged 6 and under would be expected to become continent at night in the near future.

- [Treatment of Nocturnal Enuresis](#)

### **What evidence is available?**

If the child is undergoing active management under the enuresis clinic the clinical specialist nurse is likely to be the best source

- [Evidence](#)

### **How long will the needs last?**

- [Prognosis and duration of the award](#)

### **What is Nocturnal Enuresis?**

Nocturnal enuresis is the medical term for bedwetting. The term enuresis is used to describe children who do not have voluntary control over their bladder in the absence of any anatomical abnormality or health condition.

Enuresis is not used to describe children who have an underlying condition affecting their urinary tract – the term urinary incontinence is used.

### Incidence & Prevalence

The age when dryness at night is achieved is very variable and difficult to measure – this is because the definition of bedwetting is variable. For example, enuresis might be defined, as wetting the bed at least twice a week in some studies and numbers will be small. In others the definition of enuresis may be that the child is not reliably dry and children who occasionally wet the bed will be included in the enuresis group - numbers will be higher. The DSM V defines enuresis as bed wetting at least twice a week during the previous 3 months. The condition is more common in boys. The prevalence of nocturnal enuresis decreases with age, but some children do not achieve night-time continence by adulthood. Around 0.5% of adults aged 18-64 years have nocturnal enuresis.

The following rates of enuresis by age are commonly quoted:

#### Rates of nocturnal enuresis by age

<b>Age of child</b>	<b>Prevalence of nocturnal enuresis</b>
5 years	15%
8 years	7%
10 years	5%
15 years	2%
Adults 18-64 years	0.5%

### What are the effects and signs?

Children are usually dry at night by 5 years. Because achieving dryness is variable and many children who are bedwetting at 5 and 6 will master control without medical intervention, diagnosis of nocturnal enuresis is not made until age 7. Age 7 is the earliest age that referral to enuresis clinic for specialist help can be made.

Enuresis is normal in young babies. Children learn to control their bladder and bowels during toilet training when they are 2 or 3 years old. Toilet training is a stepwise process that involves recognising the physical signals that the bladder or bowel is full and needs to be emptied. Once children can recognise these signs they can learn how to hold on or delay emptying them. Children are taught the cultural norms of how and where to go to the toilet during this period. Toilet training ends when a child routinely goes to the toilet appropriately in private. Faecal continence is usually learnt first followed

by urinary continence. Daytime control is usually learnt before nighttime control. There will be a period during which a child is dry during the day but wears nappies at night. The age when dryness at night is achieved is variable and difficult to measure. The definition of bedwetting is variable, for example enuresis might be defined as wetting the bed at least twice a week in some studies and numbers will be small. In others the definition may be that the child is not reliably dry, consequently the enuresis group will include children who only occasionally wet the bed.

Nocturnal enuresis may be primary or secondary:

- Primary nocturnal enuresis – means the child has never been satisfactorily dry at night – includes around 80% of children with nocturnal enuresis
  - Secondary nocturnal enuresis – means the child has had a period of at least 6 months of being dry at night – includes around 20% of children with nocturnal enuresis. This type of enuresis may be caused by a medical condition, for example urinary tract infection or by emotional upset in which case it may be considered a form of behavioural regression.
- [The bladder & bladder function](#)

### **How is it assessed?**

A careful history and physical examination will exclude most physical causes of enuresis such as [spina bifida](#), kidney disease and neurological problems

Investigations will include -:

- Urinalysis to exclude infection and diabetes

and may include -:

- Ultrasound of kidneys, ureters and bladder (US KUB) may be carried out to confirm that the urinary tract is normal

A careful and complete history of enuresis will be taken on the first visit to the enuresis clinic during which the following information is likely to have been recorded:

- Toileting history to distinguish primary from secondary enuresis
- Frequency of wet nights, fluid intake (day and night) or symptoms such as frequency, dribbling and the presence of constipation. Wetting 1-2 times a week may be described as mild,

wetting 3-6 times a week may be described moderate and wetting every night may be described as severe

- whether there is a family history of nocturnal enuresis
- Identify predisposing environmental factors such as stress and emotional disturbances (particularly important in secondary nocturnal enuresis)

The physical examination and investigations should be normal. If they are not, a diagnosis of nocturnal enuresis cannot be made.

- [Risk factors and causes](#)

### **How is it treated and managed?**

#### **Children aged 6 and under**

The majority of children aged 6 and under would be expected to become continent at night in the near future. Consequently, in these children it is better not to aggressively treat this problem but use a benign strategy of star charts and praise or small rewards for dry nights. Lifting at night may also be tried to reduce the frequency of wet nights. Often this is all that is necessary for the child to become reliably dry. Children who reach seven

and are still not dry at night despite this strategy will usually be offered treatment.

### **Treatment of children aged 7 and over**

Children may be treated with one or a combination of the following therapies. A common combination is the 'three system approach'. This is detailed after the individual therapies have been described.

#### **Behavioural Therapy**

This means building good routines and behaviours around going to the toilet to empty bladder and bowels. It involves encouraging and supporting a child to do the following:

- Begin the day by voiding in the toilet,
- Encourage the child not to hold on to urine unless they are doing bladder training,
- Encourage voiding at least once every 2 hours - enough to avoid urgency and incontinence,
- Ensure access to the toilet is easy at school (parents may have contacted the school),
- Encouraging the child to drink liberal amounts of water during the morning and early afternoon,
- Minimising intake of fluids after dinner,
- Encourage daily bowel movement, preferably before school.

#### **Lifting at night**

This involves making sure the child empties their bladder in the toilet at a point in the night just before they would normally wet the bed. There are two ways of doing it:

- Waking the child so they go the toilet – the child will often stay awake after this.
- Lifting the sleeping child and putting them on the toilet without waking them – this is more popular because the child can be returned back to bed without waking up.

Either of these approaches reduces the number of wet nights and may be all that is required in young children. Lifting is usually done once at around the time wetting normally occurs and before the parents go to bed. It is not expected that parents would be doing this more than once or getting up

especially to do lifting. Not waking the child takes much less time, as a woken child may require settling again.

### **Alarm Therapy**

This may be in the form of a pad placed under the sheets or attached to the underclothes, which sounds an alarm as soon as it becomes slightly wet. It is designed to sound just as the child begins to pass urine. The alarm wakes the child up so that they can finish emptying their bladder in the toilet. Parents are involved in this treatment, as they must ensure the child wakes and goes to the toilet each time the alarm sounds. The treatment is ineffective if the child switches off the alarm and goes back to sleep.

Usually the alarm will sound before the parents go to bed but they may also need to get up during the night if it goes off later or a second time. Substantial improvement is seen quickly with this treatment and it would not be expected to continue for more than 6 months, with the child remaining dry every night for the final few months of treatment.

This treatment involves time and effort in terms of making sure the alarm and pad are set and in the right place every night, listening for the alarm and attending to the child when it sounds. Attention during the night is only required for the first few months of treatment, the child is expected to be dry for the final few months if the treatment is working. The system will need to be checked before the child goes to bed – this takes a few minutes as part of the bed time routine.

The treatment is highly effective with a positive effect being noticed within the first month in those cases where it will be successful. Around 2/3 of children using this method will become dry at night. A reduction in wet nights will be seen in the first month and the alarm continues to be used until a pattern of consecutive dry nights has been maintained for some months. On average alarm therapy needs to be used for 3-6 months. This treatment is likely to be most effective in those children with delayed bladder maturation. If there has been no improvement after 1 month with the alarm treatment it can be stopped as it is unlikely to be effective.

### **Drug treatment**

#### **Desmopressin**

Treatment with drugs is used on an occasional basis to help children stay dry when it is particularly important to do so. Desmopressin is the drug most commonly used, it mimics the action of a natural hormone in the body called Anti Diuretic hormone (ADH). It effectively stops the kidneys from excreting water so if combined with a large fluid intake can cause a serious side effect called 'water intoxication'. Symptoms of this include headache, nausea and vomiting.

It is sometimes used on an ongoing basis and is effective in around half of children for the duration of therapy – enuresis reoccurs when it is stopped. The drug can be taken as a tablet or melt in the mouth preparation (note

spray no longer used in children). This treatment is most effective in the group of children with relative lack of ADH at night. Medication is taken only once a day at bed time and takes a few minutes.

Note the different way this drug is used when part of the three system approach.

#### Oxybutynin and tolterodine

These drugs are anti-cholinergic drugs. They increase bladder capacity by reducing over activity of the detrusor (bladder) muscle. They theoretically increase bladder volume. Side effects are the 'anti-cholinergic' side effects and these include constipation, dry mouth, blurred vision and flushing.

They are only used when children have not responded to non-drug therapies and may be used in conjunction with desmopressin. This treatment tends to be used on an ongoing basis. Enuresis tends to recur when treatment is stopped.

Medication is taken only once a day at bed time and takes a few minutes.

Note the different way this drug is used when part of the three system approach.

#### Imipramine and other tricyclic drugs

These drugs are used only when all other therapies including other drugs have failed. They stop bedwetting in around 20% of treated children. Symptoms recur when treatment is stopped.

Tablets are taken once a day at bed time. Side effects may be severe and include mood changes, nausea, sleep problems and heart problems. It is easy to overdose on small amounts of these tablets so they must be stored securely in the home and the parent must supervise the child taking the

tablets. Supervising the child taking the tablet and securing the drugs again takes a few minutes each evening.

### **Three System Approach**

This approach combines alarm therapy with drug treatment, the three components are -:

- Desmopressin a drug to reduce urine production correcting deficient anti-diuretic hormone release.
- Bladder relaxing drugs such as oxybutynin to prevent detrusor (bladder) muscle overactivity.
- Alarm therapy

These treatments may be used in a step-wise fashion or together and may be combined with behavioural therapies detailed above or bladder training. Bladder training involves holding on to a full bladder for as long as possible, at least once a day, to increase bladder volume. This can promote an increase in bladder capacity in children with enuresis who often have a small bladder capacity for their age.

Parents will need to supervise their children to make sure that they take medication as prescribed and carry out any behaviour modifications recommended. They will also need to attend to their child's alarm therapy as detailed under alarm therapy. Around 70% of children would be expected to be dry at night after a few months of this treatment, therefore additional care needs related to enuresis and its treatment would not be expected to last more than 6 months in the typical case.

In rare cases nighttime attention related to treatment on an ongoing basis will be needed and corroborating evidence from the enuresis clinic should be obtained to confirm this.

Note: Treatment / therapy outcomes are generally very successful within the first few weeks or months of treatment. It is important therefore that the



likely duration of care needs is fully considered in deciding whether entitlement is appropriate.

### What evidence is available?

Sources of evidence	Information Provided	Limitation of Evidence
Parent / carer	First source of information and associated evidence.	Does not have specialist knowledge.
GPFR	The GP will be able to confirm the history of enuresis and will have details of referral to enuresis clinic and current treatment.	The frequency of enuresis and the type of treatment will have the greatest effect on care needs in this condition. The GP is most likely to have a complete history of the condition and be aware of current management including frequency of enuresis.
Consultant Paediatrician / Clinical Nurse Specialist	If the child is undergoing active management under the enuresis clinic the clinical specialist nurse is likely to be the best source of current information on treatment, progress and frequency of enuresis.	If the child has only been seen once for assessment, the hospital is likely to have little information on the child's current care needs or treatment. However, they are likely to have taken a detailed history of the enuresis at the time of the first appointment.

### How long will the needs last?

Age at date of claim	Award Period
7 - 9	Award to age 10 (or for 1 year whichever is longer)
10 - 15	Award for 1 year

Almost all children with nocturnal enuresis can expect to achieve nocturnal continence eventually. Those with a family history of late nocturnal continence, those with behavioural disorders and those with developmental delay will take longer. Boys take longer than girls do. There is a high spontaneous resolution rate of nocturnal enuresis. Many children overcome incontinence naturally, without treatment, as they grow older. The number of cases of incontinence goes down by 15% for each year after the age of 5.

This means approximately half of cases will have resolved in 3 years. If care needs are identified awards should be made based on the underlying condition -:

- In most cases there will be no underlying condition. In otherwise healthy children awards should be made until age 10, the age at which half of affected children could be expected to have become dry at night and the age at which a significant proportion of children who still have enuresis could be expected to manage their bedclothes during the night themselves. On review, the presence and frequency of ongoing enuresis should be confirmed with medical evidence. An assessment of whether the child is receiving active treatment, involving the parents should be made. An assessment of whether the child requires help with cleaning themselves or their room up at night should also be made
- At age 10-11, children are encouraged to do their own cleaning up during the night and the majority will do this
- If enuresis is related to an underlying medical condition awards should be based on how long this condition is expected to last or until surgery to achieve continence is carried out
- Children with underlying conditions such as learning difficulties, developmental delay and autistic spectrum disorder are much less likely to stop bedwetting or learn how to manage it themselves during the night. Refer to separate guidance where these conditions are present

Note: Treatment / therapy outcomes are generally very successful within the first few weeks or months of treatment. It is important therefore that the

likely duration of care needs is fully considered in deciding whether entitlement is appropriate.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **The bladder and bladder function**

The bladder is a distensible and contractile muscular bag lying in the lower part of the pelvic cavity. It acts as a storage receptacle for urine before its intermittent discharge. It receives a continuous inflow of urine from the kidneys via the two ureters (one from each kidney).

Bladder function:

- The muscle layer around the bladder is called the detrusor muscle – this is the muscle that contracts to empty the bladder
- In the neonatal period micturition (passing urine) occurs in the region of 20 times a day due to uncontrolled detrusor muscle contractions in a small capacity bladder
- Bladder capacity increases during childhood from 60-70 ml in the neonatal period to 120-140ml at 4 years of age
- Bladder awareness or sensation of bladder fullness develops in the first few years of life
- Approximately 30% of 2-year-olds, 70% of 3-year-olds and 100% of 4-year-olds are able to indicate bladder awareness. This is obviously not the same as having control of bladder function (micturition)
- Urine is stored in the bladder, which stretches like a balloon as it fills up. When it stretches to a certain point the nerves in the

bladder will send a message to the brain saying that it needs to be emptied

- Children acquire the ability to suppress detrusor contractions voluntarily and co-ordinate sphincter control and detrusor function
- Daytime bladder control is achieved in most children by 4 years of age, while night bladder control is generally achieved by 7 years of age (93%)
- Referral to a nocturnal enuresis clinic is not accepted before the age of 7 years

### **Risk factors and causes**

Primary Nocturnal Enuresis:

- A family history of nocturnal enuresis in close relatives. Epidemiological studies show that if one parent has a history of nocturnal enuresis the children are 5-7 times more likely to have the disorder
- Delayed bladder maturation. The high spontaneous resolution rate of nocturnal enuresis implies a role for delayed maturation of a normal development process. Many children overcome enuresis naturally – without treatment as they grow older. The number of cases of enuresis goes down by 15% for each year after the age of 5 years
- Small capacity bladder. Children with nocturnal enuresis have been noted to have a smaller bladder capacity than age matched children who do not have nocturnal enuresis
- Decreased nocturnal anti-diuretic hormone (ADH) secretion and polyuria. Some children with nocturnal enuresis have decreased nocturnal secretion of ADH and increased urine production at night. ADH is a hormone produced in the posterior pituitary gland of the brain. It stimulates renal (kidney) retention of water. Decreased nocturnal ADH secretion would mean the kidneys retained less water and so more urine was produced (polyuria). Since ADH secretion is thought to increase with bladder distension, small bladder capacity may contribute to the decreased nocturnal ADH secretion
- Sleep disorders are associated with nocturnal enuresis
- Bladder instability. Some children with nocturnal enuresis have detrusor (bladder muscle) instability. This means the bladder involuntarily empties partly or fully before the child is fully aware of the need to pass urine or to find a toilet. This is often

accompanied by a small volume bladder and frequently symptoms of daytime urgency, frequency or daytime enuresis

#### Secondary Nocturnal Enuresis:

- An extremely important cause of secondary enuresis is emotional upset e.g. parental separation or illness, bullying at school or sexual abuse etc
- Other causes of secondary enuresis include cystitis (urinary tract infection) and constipation, both of which reduce bladder

capacity, diabetes (if not treated), which increases the volume of urine produced and some drugs

- 

## **What you need to know about Normal development & disability in children**

## **What is normal development & disability in children?**

In order to understand disability in children and where the consequent needs differ.....

- [Normal development & disability in children](#)

## **Care involving technical procedures**

The care of some infants with disabilities involves the use of technical procedures such that.

- [Technical procedures](#)

## **Care needs in infants**

Healthy infants require a great deal of attention in connection with their bodily functions. They must be fed....

- [Care needs in infants](#)

## **Care needs in older infants & young children**

As the healthy infant gets older, the emphasis shifts from attention to supervision. Feeds become.....

- [care needs in older infants & young children](#)

## **Care needs in older children & adolescents**

The variety and level of care needs and mobility requirements in older children and adolescents with disabilities are.....

- [care needs in older children & adolescents](#)

## **'Difficult' children**

Some healthy children are described by their parents as 'difficult' because they require more attention or supervision than.....

- [Difficult children](#)

## **What evidence is available?**

If the child is undergoing active management under the enuresis clinic the clinical specialist nurse is likely to be the best source

- [Evidence](#)

## How long will the needs last?

- [Prognosis and duration of the award](#)

## What is normal development and disability in children?

In order to understand disability in children and where the consequent needs differ significantly from those of a non-disabled child, it is necessary to have an understanding of the normal development process. The sequence of development is normally the same for all children e.g. they sit before they can walk etc, but there are some recognised variations on the commonest pattern. For example, up to 10% do not crawl before they walk but "bottom-shuffle", creep, roll or just stand and walk. This may occur in those children who have an inherited pattern of low muscle tone and there is usually a history of affected relatives. Most children walk (even if it's only a few steps) by the age of 2 years. The median age (i.e. the most commonly encountered age in years) of walking in shufflers, creepers and rollers is several months later than for crawlers and a few are still not walking by the age of 2. Eventually however, they function normally with walking established by the age of 3 years in the majority of these children. Those who just stand and walk also have low muscle tone and a similar family history but walk a month or two earlier than the crawler.

Development may be divided into four broad categories:

- Vision and manipulation
- Hearing and speech
- Gross motor skills
- Social behaviour

Disability or disease in a child has a great impact on parents and the immediate family. Chronic illness or disability in the infant or young child may produce considerable additional care needs - usually provided by the parents themselves. Increasing numbers of children receive high dependency care provided at home over long periods.

The attention which is given, particularly to infants and very young children with disabilities, may **differ in kind** from that given to healthy children of the same age; but this may not mean that the amount of attention given is in excess of that usually required by a healthy child of the same age. Many healthy children waken at night for a variety of reasons and require attention. Likewise young children who are not disabled require care in relation to bodily functions such as eating, washing, dressing, undressing, and using



the toilet. Some children, however, may not be receiving the attention they need as a result of their disabilities.

Assessment of care needs is also influenced by the fact that children develop both physically and mentally. This may result in decreased care needs; on the other hand, some care needs may increase. Physical development of the upper limbs in a child with defective lower limbs may enable them to move independently with mechanical aids where these are used. Increasing maturity may lead some children with chronic illness or disabilities (e.g. the child with diabetes mellitus, cystic fibrosis or arthritis etc) to assume responsibility for the care of their condition and so require less supervision. Training received may also have an effect, notably with blind and deaf children. On the other hand, physical development may increase the burden of disablement: a child with a learning disability may require more rather than less supervision as they get older and become more mobile. Adolescents with disabilities will also have to cope with care and/or mobility needs against a background of changing patterns in body functions, social attitudes and sometimes non-conforming and "rebellious" behaviour commonly encountered at this time.

### **Care involving technical procedures**

The care of some babies and children with disabilities involves the use of technical procedures such that the attention or supervision/watching-over required from birth may be greatly in excess of that required by a healthy baby or child. These include:

- Regular mechanical suction because the child has a tracheostomy (an opening made through the neck into the trachea to aid breathing) or other upper airway problem
- Tube feeding into the stomach/upper bowel or a vein
- Regular use of oxygen in order to survive. This includes babies or children with broncho-pulmonary dysplasia (impairment of normal lung development and impaired lung function) as a result of very premature birth
- Care following surgical procedures whereby a segment of the stomach or bowel is opened up onto the abdominal wall for feeding or for the elimination of waste: gastrostomy (the stomach has an opening onto the abdominal wall to assist in feeding by tube); ileostomy; jejunostomy; colostomy (all these are connections between a particular part of the bowel and the abdominal wall. They are usually constructed to form an exit from

the intestine when part of it is blocked or has been destroyed by disease)

- Care following a nephrostomy (a connection between the urinary tract and the abdominal wall, constructed to form an exit for the passage of urine)

### Prematurity in babies

## **Care needs of older infants/young children**

### **The non-disabled infant**

As the healthy infant gets older, the emphasis shifts from attention to supervision. Feeds become less frequent; winding is no longer necessary, and the child begins to feed themselves. However, from the age of about six months the development of investigative skills together with increasing mobility puts the healthy child at risk of danger; the level of supervision required to avoid danger is considerable.

### **The infant with disabilities**

At this stage (often between 9 and 15 months), the gap between the care needs of a healthy child and a child with disabilities may have widened to the extent that the needs of the child with disability are significantly in excess. These may include continued attention to bodily functions no longer required by the healthy child and more attention than that needed by the healthy child for the development of new skills such as crawling, standing and walking. The age at which the need for attention of the child with disability becomes greater than that of the healthy child cannot be defined precisely and judgement will depend on the evidence available in the individual case.

### **Disabilities posing substantial needs**

There will be some children with disability with needs persisting or first manifesting at a level in excess of the norm at this age, for example:

- Children with brittle bones (osteogenesis imperfecta), haemophilia and other severe bleeding disorders in who bumps and falls are associated with the risk of fractures or haemorrhage
- Mobile children with hearing and/or visual problems who cannot respond to a warning shout or see a potential danger, which a healthy child would avoid
- Children with cerebral palsy whose mobility is impeded and whose risk of postural deformity is reduced by frequent changes in position by parents
- Children with a severe learning disability who eat undesirable substances (pica) or exhibit self-mutilation behaviour. A child

with severe learning disabilities may also require substantially more stimulation to maximise potential

- Children in whom developmental delay may first become evident because of a need to continue a level of attention appropriate for a much younger baby.

### **Night needs in infants and young children**

Healthy children **under the age of two years** normally require a considerable amount of attention, both in frequency and duration, during the night hours for feeding, changing or "settling" - the latter especially during teething. Specific, regular attention at night in excess of normal levels may be required by some children with disabilities whose medical condition calls for parental intervention in the form of turning, nebulizer or oxygen therapy, suction, intubation and care during fits etc.

If precautions are taken at night (such as the child being safely placed in a cot with sides and bumpers if required and used) there may be few conditions requiring watching-over in the absence of attention needs, which are substantially in excess of those needed by a child of comparable age.

However, the need for watching over in excess of normal levels will depend on the evidence available in an individual case. Notably, children with severe learning difficulties may have an abnormal tendency to develop a persistent habit of night wakening. In such cases, attention from parents may be required more than once a night and may last one hour or more.

### **Care needs of infants (children under 1 year old)**

#### **The non-disabled infant**

An infant for the purposes of this text is taken to be a child aged less than one year old. Healthy infants require a great deal of attention in connection with their bodily functions. They must be fed, winded, changed and bathed frequently. In addition, if emotional development is to proceed normally, an infant must be handled, cuddled, talked to and played with regularly. Furthermore, during the times when the infant is sleeping, periodic checks are made to ensure that all is well.

#### **The infant with disabilities**

Because of the amount of care and supervision/watching over required by a healthy infant, the amount required by an infant with disabilities may not usually be much greater than that needed by a healthy child. The kind of attention given may differ however, for example, instead of being handled in an ordinary manner, the infant with disabilities may need more specific stimulation or formal passive movements of the limbs in the form of

physiotherapy but the amount of care or supervision/watching-over may not be greater than that given to a healthy infant.

### **Disabilities posing very substantial needs**

Infants with certain disabilities will require considerable amounts of stimulation, care or supervision in addition to the normal care routine. These disabilities include:

- Infants with frequent loss of consciousness usually associated with severe fits secondary to birth asphyxia or rare forms of congenital metabolic disease
- Infants with severe impairment of vision and/or hearing. (Unless there is reason to suspect that a baby may be born with hearing impairment and has been checked with special techniques, it is unlikely that hearing loss will be picked up until the child is several months old)
- Infants with severe multiple disabilities
- Other categories of infants with disabilities may well require extra care such as infants with renal failure, cystic fibrosis, asthma, cerebral palsy and those survivors of extremely pre-term birth. See [Prematurity in babies](#)
- Infants with severe feeding problems, which are due to physical reasons such as oro-facial malformations (e.g. cleft palate) or cerebral palsy
- Some infants with developmental delay/learning disabilities require prolonged periods to take adequate amounts of each feed. Some children with Down syndrome may fall into this category.

### **Care needs of older children & adolescents**

The variety and level of care needs and mobility requirements in older children and adolescents with disabilities are dependent not only on chronological age but also on a number of other complex and interrelated factors, which arise not only from the disabilities themselves but from consideration of the circumstances operating in the individual child/adolescent.

#### "Difficult" children

Some healthy children are described by their parents as 'difficult' because they require more attention or supervision than other children of their age. However, the increased needs here may not necessarily arise from severe physical or mental disability. It is however important to determine that children with disruptive behaviour at home have been assessed properly to ensure there is not a physical, intellectual or other reason for their behavioural

problems. Refer to CCM [ADHD/ADD](#) guidance and [Learning Disability](#) guidance for further information.

### **What evidence is available?**

A report from the GP or paediatrician may help in determining the level of disability and the likely duration of care needs. By the age of six, the child may have been in some form of education for a year and assessment of potential will have been made. At this time, a copy of a statement of Special Education Needs (SEN), a copy of an Individual Education Plan (IEP) or a school report may also help in determining the level of any continuing care

needs and their likely duration. SEN statements are now being replaced by Education, Health and Care Plans.

Most Child Development Centres provide parents with a report on the child's assessment, which may be a useful source of additional information should this be required.

How long will the needs last?

It isn't possible to give generalisations on the duration of needs. This will depend entirely on the particular disability or disabilities for which the child has care and/or mobility needs.

**What you need to know about Perthes disease**

### **What is Perthes disease?**

Perthes disease is a condition that affects the hip joint in children. One, or more rarely, both joints are involved. The condition causes pain in the hip and...

- [Perthes disease](#)

### **What are the effects and signs?**

Perthes disease causes pain in the hip and a limp. The child may show signs of limping and may complain of mild pain.....

- [Effects of Perthes disease](#)

### **How is it assessed?**

The hip joint is a deep structure, so investigations are required for diagnosis. The following tests may be used .....

- [Assessment of Perthes disease](#)

### **How is it treated and managed?**

Treatment of the disease depends on the clinical picture at presentation. Young patients under age 4 sometimes.....

- [Treatment of Perthes disease](#)

### **What evidence is available?**

The best source of information about surgery, braces and plaster casts will be the Consultant Orthopaedic Surgeon who .....

- [Evidence](#)

### **How long will the needs last?**

- [Prognosis and duration of the award](#)

### **What is Perthes disease?**

Perthes disease is a condition that affects the hip joint in children. One, or more rarely, both joints are involved. The condition causes pain in the hip and typically presents with a limp. The hip joint is a ball and socket type joint, the femoral head is the ball, and the pelvis forms the socket around it.

The normal hip joint is both strong and flexible with a wide range of movement.

The underlying problem in this condition is disruption of the blood supply to the femoral head – the ball of the socket. Areas of the femoral head lose their blood supply and these areas of bone die. The medical term for this

process is avascular necrosis. The process of bone death and subsequent healing causes inflammation in the joint. Active inflammation is painful, and walking exacerbates this.

Picture shows Perthes disease affecting the left hip – Note the increased density, broadening and flattening of the bone in the femoral head on the affected left side while the bone in the unaffected right side remains tall, smooth and rounded.

Reproduced with the kind permission of Mr Colin E Bruce. Alder Hey Children's Hospital, Liverpool, UK.



The dead areas of bone heal over time. In a mild case a small area of bone is affected, and it heals over time by itself with little change to the shape of the femoral head. In a more severe case larger areas of the femoral head are affected, and the bone can become soft. Ordinary weight bearing can lead to crushing of areas of the femoral head. Without treatment the healing



process leads to flattening and deformity of the femoral head. This means the ball and socket joint does not work smoothly and the range of movement is reduced. Although the bone heals over several years the deformed flattened femoral head leads to early onset arthritis of the affected hip.

The aim of treatment is to prevent deformity from developing whilst the femoral head heals. Treatment will vary depending on how much of the femoral head is involved. Activities that could damage the softened bone such as running and jumping on hard surfaces may be restricted. In mild cases bed rest and crutches may be needed during periods when the hip is too painful to allow full weight bearing. In more severe cases surgical treatment, immobilisation of the joint in plaster or wearing a brace may be necessary.

This condition has a variable effect on mobility, typically for 2-4 years until healing has occurred. Mobility is likely to be maintained in mild cases that are being treated with physiotherapy, painkillers and x-ray monitoring. Those undergoing active treatment with surgery, immobilisation in plaster or

bracing are most likely to be unable to walk for a prolonged period. Once the condition has resolved mobility returns to normal and joint mobility

usually returns to give a functional range of movement even if the hip joint is abnormal. In the longer-term arthritis of the hip can develop at a young age, typically around 40 years.

### **Incidence/prevalence**

Perthes Disease (also called Legg-Calve-Perthes Disease) is a disease of childhood. It affects children aged between 3 and 10 years, with a peak between 5 and 7 years. It is four times more common in boys than girls. In

most cases one hip is affected, in one in ten cases both hips are affected. It affects 10 -15 per 100 000 each year.

### **What are the effects and signs?**

Perthes disease causes pain in the hip and a limp. The child may show signs of limping and may complain of mild pain. The symptoms may occur

intermittently over weeks or months. Resting often relieves the pain. Affected children are often very active and athletic. Examination of the hip is likely to be painful and show reduced range of movement of the joint.

The prognosis and likely treatment will depend on two main factors -:

- The age of the child
- Extent of the disease – see [grading of severity](#)

### **How is it assessed?**

The hip joint is a deep structure, so investigations are required for diagnosis. The following tests may be used -:

- X-ray – plain x-rays are likely to show changes in the femoral head. The child will usually have a series of x-rays over the course of treatment.
- Ultrasound of the hip joint – this test can reveal whether there is fluid in the hip joint but cannot determine the cause.
- A Magnetic Resonance Imaging (MRI) scan is not usually necessary. It is used if the diagnosis is not clear and can show the earliest changes associated with the disease before they are visible on X-ray. MRI may be used to plan surgical treatment.

### **Age at diagnosis**

Young children aged 2-6 have a better prognosis. This is because young children have many years of growth left – they have the maximum capacity

for re-growth and re-modelling of the femoral head back to a normal shape once healing has occurred. Older children do not have years of growth left and deformity of the femoral head is more likely to persist. For this reason, older children are more likely to have active treatment in the form of surgery and bracing to prevent deformity from developing whilst healing occurs.

### **How is it treated and managed?**

Treatment of the disease depends on the clinical picture at presentation. Young patients under age 4 sometimes maintain a good range of movement without intervention and do well with conservative management. Children who lose range of movement are often treated surgically.

Maintaining independent mobility is one of the aims of treatment. Patients who lose range of movement are likely to have a period of immobility during the course of management when they are often kept in a hip spica cast for a period of 6-8 weeks. After removal of the cast there is a variable period of rehabilitation lasting from 3-6 months when mobility gradually improves in most but not all patients.

The goals of treatment are to -:

- reduce pain and maintain or restore hip mobility
- prevent deformity or restore normal spherical shape to the femoral head

Treatment used to achieve these goals will depend on the age of the child and the extent of disease. It will also depend on the preference and experience of the treating doctor. There is currently no agreed best or standard treatment for Perthes disease and various treatments have been, and are used including physiotherapy, bracing and surgical methods. More recent studies currently suggest that non-surgical treatment using physiotherapy or bracing is of little benefit. There is evidence that some patients will benefit from surgical treatment (femoral osteotomy /pelvic osteotomy) while others will not. Determination of whether surgical treatment is appropriate is best left to specialists in children's orthopaedics.

- Physiotherapy and monitoring – many children will have conservative treatment consisting of physiotherapy and exercises done at home and regular clinical and x-ray monitoring of disease until healing occurs.
- Plaster cast or a brace – includes use of braces and short periods in plaster. The aim of the brace or plaster is to hold the femoral head in the best position for healing inside the hip socket, the hip socket acting as a mould around the femoral head.
- Surgical treatment – surgical operations on the hip are designed to ensure that the femoral head heals inside the socket of the hip joint and can include operations on the hip bone to

ensure the femoral head sits more snugly in the socket whilst it heals, or operations on the pelvis to ensure the socket fits more snugly around the femoral head – or a combination of both. In this case the hip joint socket acts as a mould to the femoral head helping it to heal into the best possible shape to enable normal function in the long term.

### **Physiotherapy and monitoring**

More than half of children will have this treatment. They are most likely to be children aged 5 and under at diagnosis or older children with very limited disease. These children are likely to have recurrent hip pain over the 2-4 year healing period but maintain most of their normal activities including walking for the majority of the time. Activities such as cycling and swimming are encouraged. During periods where the hip is very painful bed rest for a few days and painkillers may be advised. Prolonged bed rest or use of crutches is no longer advised. They may use crutches for a few days intermittently when weight bearing is painful. Although they will be able to walk the majority of the time they will sometimes be limited by hip pain and may for example sit out towards the end of PE and not be able to run about as much as other children because of hip pain or stiffness. Parents will be advised to give them painkillers such as ibuprofen and paracetamol to relieve pain and they may take one of these prophylactically (preventatively) prior to physical activity such as PE. The progress of their disease will be monitored by regular x-rays and observation. Physiotherapy continues as long as healing is progressing well. During this period daily physiotherapy exercise will need to be done at home to maintain mobility and muscle tone round the hip joint. In younger children the parent will need to help with and supervise these exercises. Older children may need prompting or reminding to do their physiotherapy exercises. These children may move on to have more active treatment such as surgery or bracing if healing does not progress as expected.

### **Plaster cast or a brace**

This type of treatment is more likely to be used in children over 5 and those who have more severe disease. The aim of the treatment is to hold the femoral head inside the hip joint to encourage it to heal in a less flattened more functional rounded shape. The best position for the joint is 'abduction'. In practice keeping one leg in abduction with a brace or cast is very difficult so both legs will often be braced to hold the legs apart and the hip joint in 'abduction'. Whether a brace or serial plaster casts are used or a combination of both, treatment may last for 18-24 months. Because of the prolonged effect on mobility of bracing or serial plaster casts these treatments are used much less often in this country than they were even 10 years ago. There will be a period of daily physiotherapy and further monitoring once the cast or brace comes off.

### **Surgical treatment**

#### **Osteotomy**

Surgery on the hip is used to improve the position of the femoral head in the socket of the hip joint. It is usually used when the assessment shows that

results with conservative treatment are likely to be poor. Generally, these will be children who have very limited movement of the hip, extensive disease on x-ray or who are 6 years old or older who have relatively little time for skeletal remodelling after healing.

The position of the femoral head is changed using surgery so that it is more completely contained in the hip joint. There are many different types of operations that can be performed and more than one operation may be performed at the same time to improve the alignment.

Operations involve cutting the bone in two and plating and screwing or screwing the two pieces back at a different angle to improve alignment. This type of surgery where bones are cut is called an 'osteotomy'. The metal plates or screws are used to hold the bone in place in the correct position whilst it heals. The screw and plates are called 'metalwork' and may need to be removed at a later date. The osteotomy may be performed on the femur - a femoral osteotomy or on the pelvis - a pelvic osteotomy. Or a combination of osteotomies may be done, e.g. triple osteotomy.

Recovery from these operations is similar and walking ability is almost always regained within 6 months of surgery. Recovery from surgery takes some months -:

- Recovery in hospital for up to one week.
- Up to 8 weeks in plaster after the operation - a hip spica may be used - the child will be unable to walk whilst this is in place.
- The child may not be in plaster whilst healing occurs but they are likely to be non-weight-bearing or partial weight bearing for a period after surgery, in either case they will be mobilising on crutches. An osteotomy is a deliberate break in a bone and takes time to heal like any other fracture

Bony healing may take up to 12 weeks. Once this has occurred the child will be walking normally for short distances without crutches but they will still have Perthes Disease. They will need to continue with their physiotherapy exercises and will still have symptoms of hip pain from time to time. Mobility is usually good within a few months of weight bearing being allowed. Following surgery they will be closely monitored until healing of the femoral head has occurred. Further surgery or treatment may be required.

### Arthrodiastasis

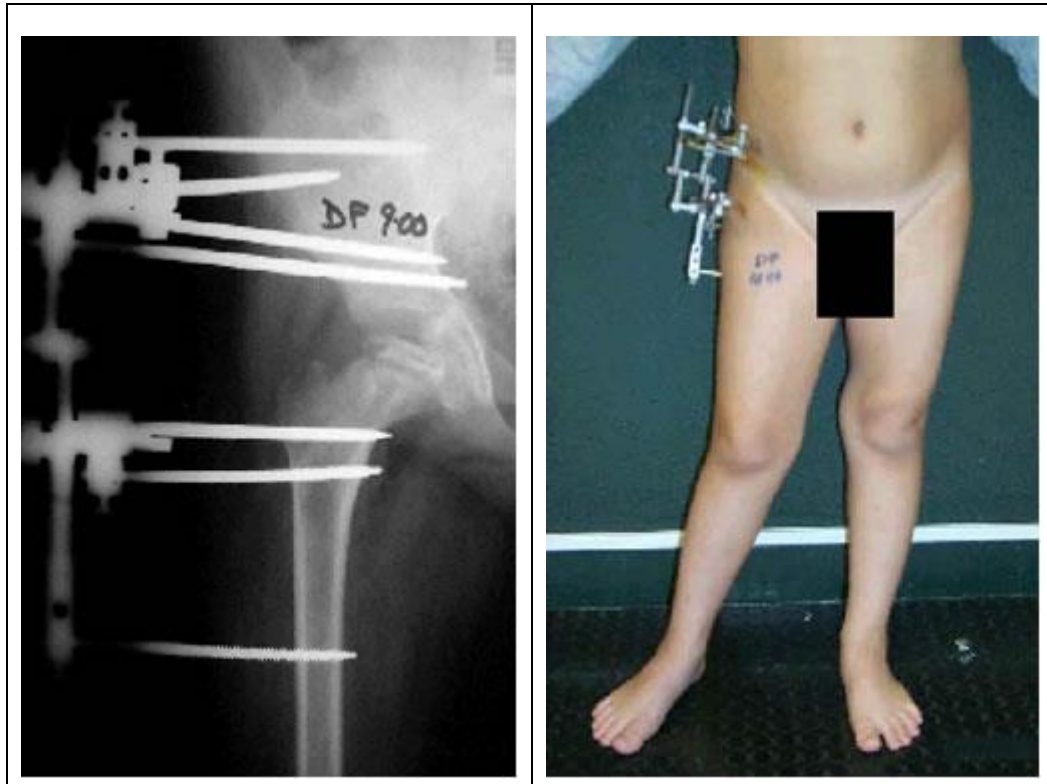
This is a surgical treatment using an external fixator. An external fixator is a metal frame that is used to support a joint. Metal pins are drilled in to bones above and below the joint and a very strong metal frame is constructed between the pins. The frame can hold the joint apart and this is effectively what the treatment is. The bones are not cut and there is no 'fracture' healing time. The metal frame holds the hip bone and the pelvis apart taking the weight that would normally go through the hip into the femoral head. By taking the weight from the femoral head it is hoped that this treatment will

result in less femoral head collapse. The frame is fixed under an anaesthetic and worn for around 2 months, then it is removed and a small brace is worn for two months. The child is able to walk with these devices but may

needcrutches. Normal walking ability is regained within 6 months of the start of treatment in most cases.

Picture. This boy is weight bearing in an external fixator. His x-ray shows he has Perthes disease of the right hip and an external fixator in place.

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- [Examples of plaster casts](#)
- [Indicators of severe functional restriction](#)
- [Other treatments](#)

**What evidence is available?**

Sources of evidence	Information Provided	Limitation of Evidence
Parent / carer	First source of information and associated evidence.	Does not have specialist knowledge.
GPFR	The GP will be able to confirm that a child has seen specialist about their hip.	The treatment of this condition is likely to be the most important factor affecting mobility and care needs in these children. The GP may not have details of treatment received and may not have seen the child since their referral.
Consultant Orthopaedic Surgeon / Clinical Nurse Specialist	The best source of information about surgery, braces and plaster casts will be the Consultant Orthopaedic Surgeon who will be able to provide details of the treatment received and how mobility has been affected. If the community paediatric nurse is involved, they will be able to provide details of the treatment received and may be able to provide some information on practical problems for the family including mobility problems and any help given with dressing, toileting and therapy.	N/A
Physiotherapist	If the main focus of the claim is for help with therapy the physiotherapist should be contacted to provide details of frequency of treatment required – is it daily and how long does the therapy take on each occasion.	N/A

**How long will the needs last?**



## Physiotherapy and monitoring

Physiotherapy is an important aspect of treatment, and the majority of the physiotherapy exercises will be done either with a parent or carer or under supervision. Such children will require help with their therapy on a daily basis. Typically exercises take around 15 minutes and are done twice daily.

Children will not require any extra help with care or mobility for the majority of the time. They will sometimes rest in bed or be mobile on crutches when the hip is particularly painful. They will require extra emotional help and support from their parents to deal with a painful condition especially if it restricts their lifestyle. These children are often athletic and in all but the mildest cases sporting activities are likely to be significantly affected.

What is the prognosis and duration?

If care needs such as daily help with therapy are identified these can be expected to last for 2-4 years. A 2-year award is recommended to check that treatment including daily therapy by the parents is ongoing.

<b>Age at date of claim</b>	<b>Award Period</b>
Age 0 - 8	Award for 2 years

## Plaster cast or a brace

### Broomstick plaster

Serial casts are rarely used in the UK. In these cases care and mobility needs related to the casts may be prolonged by more than 6 months. A child in broomstick plasters will be unable to walk normally. If they can weight bear to mobilise they will need crutches to get around. Mobilising with crutches in this type of plaster is tiring and the child may require a wheelchair for longer journeys and at school. Seating is difficult and adapted seating to accommodate the legs may be needed at school and at home too. Getting on and off a toilet may be difficult, if help is needed the parent is likely to accompany the child to school to provide such help. At home the parent is likely to provide help with both washing and dressing, the child will not be able to manage their shoes and socks and may need help with other aspects of washing and dressing related to their casts. Once the cast comes off the child is likely to resume walking normally very

quickly. However additional care may be provided by parents if daily physiotherapy exercises are prescribed.

What is the prognosis and duration?

Duration of mobility and care needs will depend on whether broomstick plasters are being used as a one off treatment of the hip for 6-8 weeks only or as treatment over a period with serial casts. Serial casts would be unusual and should be confirmed in the medical evidence. It is recommended that awards related to casts be given until the projected end of treatment, if the date is not known then review after one year. Care needs may continue beyond the period in plaster if daily physiotherapy is prescribed. Children of 12 years of age can be expected to take responsibility for and do their own physiotherapy.

Age at date of claim	Award Period
0 – 11 - Cast being worn	Award until projected end of treatment or award for 1 year if projected end of treatment date is not known
0 – 11 - Ongoing therapy once cast is removed	Award for 1 year in cases where continuing physiotherapy is required

Hip spica

A hip spica is only likely to be used for 6-8 weeks either as sole treatment of the hip or to protect the hip after surgery. A child in a hip spica will be unable to walk normally.

If they can partially weight bear to mobilise they will need crutches to get around. The child may require a wheelchair for longer journeys and at school. Seating is difficult and adapted seating to accommodate the leg may be needed at school and at home. As one hip is unable to bend getting on to and off the toilet and balancing on it is likely to be difficult. If help is needed with toileting the parent is likely to accompany the child to school to provide such help. At home the parent is likely to provide help with both washing and dressing, the child will not be able to manage their shoes and socks and may need help with other aspects of washing and dressing related to their cast. Once the cast comes off the child is likely to resume walking normally very quickly. Once the spica is removed the child still has Perthes disease, the treatment simply puts the hip joint in a better position for healing – healing must still occur. Daily physiotherapy exercises may be required and parents will have to help with or supervise these.

What is the prognosis and duration?

Time in plaster is likely to be short and normal walking should be resumed within 6 months of starting the treatment. If care needs such as daily help

with therapy once the cast is removed are identified these can be expected to last for 2-4 years. A 2 year award is recommended to check that treatment including daily therapy by the parents is ongoing. Children of 12 years of age can be expected to take responsibility for and do their own physiotherapy.

<b>Age at date of claim</b>	<b>Award Period</b>
Age 0 - 7	Award for 2 years
Age 8 – 9	Award for 2 years
Age 10 - 11	Award to age 12 (or for 1 year whichever is the longer)

### Orthotic brace

The child may or may not be able to mobilise by walking or running in the brace. If children can get around in a brace they will. They may be dependent on crutches or sticks for mobility. They will be able to get on and off the toilet by themselves and are unlikely to require help with this during the school day unless they are unable to walk.

The brace can be removed so the child can wash and a parent or carer will have been taught how to do this and will supervise the child during washing and bathing and reapply the brace afterwards. The parent will need to remove and refit the brace whenever the child gets changed, washes or goes swimming and supervise the child the whole time the brace is off, effectively either helping with or carefully supervising all washing and dressing. This treatment is rarely used in the UK and should be confirmed in the medical evidence.

What is the prognosis and duration?

Care will be required during the whole period in the brace. The brace will need to be taken off and put on again morning and evening and if the child goes swimming. Help may be required to putting clothes on and taking them off over the brace. If the child cannot walk in the brace mobility will be severely affected. Most children can walk in a brace and the majority use crutches to do this. Awards related to care and mobility in the brace should last until the brace is no longer worn. Typically the brace will be worn for 1-2 years. Once the brace is removed daily physiotherapy may be required, an

award for ongoing physiotherapy once the brace is removed should be renewed after one year.

Children of 12 years of age can be expected to take responsibility for and do their own physiotherapy.

<b>Age at date of claim</b>	<b>Award Period</b>
0 – 11 - Brace being worn	Award for 1-2 years depending on how long the brace is to be worn
0 – 11 - Ongoing therapy once brace is removed	Award for 1 year in cases where continuing physiotherapy is required after removal of the brace

## **Surgical treatment**

### Osteotomy

Clearly, children who have had surgery and are non-weight bearing on crutches or in a hip spica whilst bone healing occurs, will not be able to walk for a period. The majority of such children should be walking normally within 6 months. Children who are still unable to walk at 6 months are likely to be those with the most severe disease, bilateral disease or complications of surgery. The majority of these will resume walking within a further 6 months to a year.

It is likely that these children will have ongoing needs for physiotherapy at home when they have recovered from surgery. It is likely that children will have extra care needs related to reduction in mobility after surgery for several months, but resume walking within 6 months of the start of treatment. Care needs related to ongoing physiotherapy once normal walking is resumed could be expected to last for up to 2 years.

What is the prognosis and duration?

Care needs relate firstly to surgery and then to ongoing physiotherapy at home. Walking difficulties are only likely to last longer than 6 months in cases where complications of hip surgery have developed or where the second hip is operated shortly after the first. A 1 year award is recommended.

Children of 12 years of age can be expected to take responsibility for and do their own physiotherapy.

<b>Age at date of claim</b>	<b>Award Period</b>
0 – 11	Award for 1 year
Age 0 – 11- Ongoing therapy once out of cast & weight bearing	In cases where continuing physiotherapy is required award for 1 year

### Arthrodiastasis

The external fixator is likely to be in place for 2 months. Parents will need to follow a wound care plan to care for the pins during that period. They will also need to help with washing and dressing whilst the pins are in place. Once the pins are removed there may be an ongoing need for daily physiotherapy.

What is the prognosis and duration?

It is unlikely that wound care will be needed for more than 2 months and mobility is maintained through the treatment. There may be care needs related to physiotherapy following on from arthrodiastasis treatment. 2 year awards are recommended for care needs related to ongoing therapy.

Children of 12 years of age can be expected to take responsibility for and do their own physiotherapy.

<b>Age at date of claim</b>	<b>Award Period</b>
Age 0 - 9 - ongoing therapy	Award for 2 years
Age 10 – 11 (and over) - ongoing therapy	Award to age 12 (or for 1 year whichever is the longer)

Note: Mobility needs are unusual with modern treatment. Normal walking ability resumes once child is out of plaster or brace. Daily physiotherapy treatment usually continues for a period after this.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Severity of disease**

It is unusual for the whole of the femoral head to be involved in Perthes Disease. Several grading systems have been developed to measure the extent of involvement. The extent of disease is graded, based on the appearance of plain x-rays of the hip joint. Severity of disease and age will dictate treatment.

### **Grading of severity based on x-ray appearance of affected hip**

In simple terms the greater the proportion of the femoral head involved the more likely deformity is to develop. These children may be described as being in the 'head at risk' category in the medical evidence. This means they are at risk of developing deformity of the femoral head. Because of this risk

these children are most likely to have surgical treatment or other treatments that significantly limit their mobility.

The following grading systems may be used to describe severity in the medical evidence -:

- Catteral Classification – x-ray appearance is graded from I to IV (I being mild and IV being severe).
- Salter-Thomson Classification – simplifies the Catteral classification into two groups -:

Group A includes Catteral I and II and means less than 50% of the ball is involved.

Group B includes Catteral III and IV and means more than 50% of the ball is involved. If less than 50% of the ball is involved the prognosis is good and if more than 50% is involved the prognosis is potentially poor.

- Herring Classification - measures part of the femur on the X-ray called the lateral pillar. There are three grades, grade A, B and C with grade A being the most normal looking x-ray appearance with the best prognosis and group C being the most abnormal with a worse prognosis.

## **Examples of plaster casts**

### **Broomstick plaster**

Both legs are encased in plaster and a bar is fixed between the 2 legs to form the shape of an 'A'. Both legs are held apart keeping the hip joints in abduction – the best position for healing. The casts cannot be taken off at all. The casts are removed at review appointments every few weeks when the joint is assessed; the cast is reapplied if necessary before the child goes home. After some months plaster may be replaced by an orthotic brace designed to do the same job. This treatment is rarely used in the UK because of its effects on mobility.

#### **Effect on mobility and care needs**

A child in broomstick plasters will be unable to walk normally. If they can weight bear to mobilise they will need crutches to get around. Mobilising with crutches in this type of plaster is tiring and the child may require a wheelchair for longer journeys and at school. Seating is difficult and adapted seating to accommodate the legs may be needed at school and at home too. Getting on and off a toilet may be difficult, if help is needed the parent is likely to accompany the child to school to provide such help. At home the parent is likely to provide help with both washing and dressing,

the child will not be able to manage their shoes and socks and may need help with other aspects of washing and dressing related to their casts.

### **Hip spica**

A plaster cast is applied around the lower body and the affected leg, holding the hip joint in abduction (movement of the limbs away from the body). This type of plaster may be used for 4-8 weeks after surgery or whilst a brace is being custom made. It is not likely to be used for longer than this. Most children will not be able to walk or weight bear in the hip spica.

### **Orthotic brace**

Braces are custom-made and fitted to the individual child. If used they will be worn for at least 12 months. Most braces used in this condition will consist of a waist belt attached to cuffs designed to fit round the thighs and hold the legs apart. Pictured below is an example of the Scottish Rite Brace - most braces used in Perthes disease are based on this design.

Braces are designed to be worn all of the time but are designed to be taken off for washing and bathing and for swimming which is encouraged. Braces are individually designed and may be more or less restrictive depending on the extent of disease. It will be more difficult for a child to walk and run around in their brace if both hips are affected and they may require crutches. Most children will be able to walk in their brace but this will be slow and awkward even with the aid of crutches. This treatment is rarely used in the UK because of its effect on mobility.

#### **Effect on mobility and care needs**

The child will be able to walk awkwardly and slowly in the brace, most children will do this with the aid of crutches. They will be able to get on and off the toilet by themselves and are unlikely to require help with this during the school day unless they are unable to walk.

The brace can be removed so the child can wash and a parent or carer will have been taught how to do this and will supervise the child during washing and bathing and reapply the brace afterwards. The parent will need to remove and refit the brace whenever the child gets changed, washes or goes swimming and supervise the child the whole time the brace is off effectively either helping with or carefully supervising all washing and dressing. No other help with therapy is required during the period in the brace this is



because physiotherapy is not routinely used at the same time. Treatment time in the brace is normally at least 12 months.

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### **Other treatments**

Examination under anaesthesia (EUA) – because the hip is painful, it may not be possible to examine it properly with the child awake. An examination under anaesthetic is a short anaesthetic to enable assessment of the hip. There are no disabling effects.

Botox treatment – botulinum toxin injections may be injected in to muscles around the hip to reduce spasm of the muscle. The paralysing effect of botulinum toxin effectively weakens the muscle and can relieve pain and improve range of movement around the hip. The effects wear off after a few months.

Tenotomy - a tenotomy is a small operation on a muscle near the hip joint. It is done when the muscle becomes abnormally short due to prolonged

limping. The operation may be followed by 6-8 weeks in a hip spica to ensure the cut muscle does not heal short again.

Bed rest – used to be used for many months at a time. Bed rest is only recommended for a few days at most.

Traction – again used to be for many months in the past – now may be used for a few days to relieve pain.

Drug treatment -:

- Ibuprofen – is a pain killer commonly prescribed to children with Perthes disease.
- Paracetamol (Calpol) – is a painkiller commonly prescribed to children with Perthes Disease.

### **Indicators of severe functional restriction**

Any child undergoing active treatment for their disease may have mobility or care needs. These may or may not persist for more than 6 months. Children with bilateral disease where both hips are affected are more likely to have care or mobility needs persisting beyond 6 months. This is because the disease often occurs sequentially i.e. the second hip is affected some time later and the total duration of treatment is longer.

Any of the following treatments may affect care and mobility needs -:

- Broomstick plaster – children will be unable to walk in these. May also require help with mobilising to the toilet and dressing.
- Hip Spica – child will be unable to walk in this. May also require help with mobilising to the toilet and dressing.
- Surgery – osteotomy – there will be a period after surgery when the child will be unable to weight bear on the operated hip, younger children are likely to spend the healing period in a hip spica and older children will mobilise on crutches. Normal walking would be expected by the majority of children at six months after surgery.
- Arthrodiastasis – children with an external fixator will be mobile. Their parent or carer will need to clean the pin sites daily and monitor them for signs of infection. Children will need help and supervision with washing and dressing.
- Orthotic brace – all children with a brace will require help with washing and dressing as the parent will need to put on and

take off the brace for them whenever necessary. The child may or not be able to walk independently in the brace.

## Long-term effects of Coronavirus 19 in children (Post Covid-19 Syndrome-Long Covid)

### **Background**

Young people are presenting to child DLA with long term symptoms following infection with Coronavirus 19. Although the majority of children recover from Coronavirus 19 with no ongoing effects, some do not. The most severely affected children may claim child DLA.

Post COVID 19 syndrome is defined as signs and symptoms of COVID that persist for more than 12 weeks. Long COVID is defined by NICE as COVID symptoms that have persisted for 4 weeks or longer.

Some children have now been ill with long COVID since 2020. This guidance note provides a short update on the condition and advice on how to manage award reviews. This information is based on the medical literature, a review of child DLA cases and personal communications with Department of Health and Social Care and a clinician treating young people with long COVID. The understanding of the nature of the condition and best management continue to develop.

### **Long Covid**

Most children claiming DLA following COVID have 'long COVID' – they have had Coronavirus 19 and either remained unwell or become unwell shortly afterwards. Fatigue, headache and brain fog are the most common symptoms in all children with long COVID, but in child DLA claimants fatigue and sleep problems are more disabling. Children are often sleeping so many hours a day they are unable to attend school at all, or only manage a few hours a week, either in person or online. Pain and dizziness, which are common in our child DLA population, are rarer in children with long COVID as a whole. Many children with long COVID, on DLA, have significant mobility problems. Children experience a wide range of symptoms, only the main ones are mentioned here. These children may or may not have been in hospital with their initial illness.

### **PIMS**

A smaller number present with ongoing disability related to PIMS (paediatric multisystem inflammatory syndrome also known as PIMS-TS and MIS-C). This is a rare serious illness where inflammation occurs throughout the body around 6 weeks after the initial Coronavirus 19 infection. The inflammatory process is harmful to healthy tissues, for

example causing respiratory failure in the acute phase. It can also cause long term damage, for example to the heart. These children will have been in hospital undergoing treatment for the condition. Some of them will have been treated on ITU and may have ongoing health complications related to their treatment. They may be very debilitated by having had a serious illness. The evidence base shows whilst most make a full recovery, those that have not yet recovered tend to have ongoing fatigue and emotional problems. Some of them may have ongoing neurological problems or other rare complications of the condition or its treatment.

### **Care and mobility needs**

In our child DLA sample the children were mostly over 10 when they became ill and were off school with their illness, having been fit and well up to that point. Most of them were sleeping so many hours a day that they could not attend school, some managed a few hours a week. They were at home, needing help all day, with meals being prepared for them at lunchtime, some needed help with washing and dressing. Some, who were able to engage with education, were prompted or helped to manage school work, with the parent communicating with school and taking them to school, for example, for an hour, as part of a phased return. Many were unable to engage with learning at all, even online. Some of the children needed prompting or help with many activities of daily living due to severe fatigue.

When assessing needs consider the age of the child and what they would be doing if they were well. Most children over age 10 would be at school during the day, independent in washing, bathing, getting snacks, arranging their own social life and going out, including to school. Children with long COVID tended to be stuck at home almost all the time, sleeping a lot and requiring physical and emotional support from their parents far in excess of children without fatigue and hypersomnia. Many of the children have significant emotional problems due to issues created by long COVID. Some of the children had significant mobility issues and were wheelchair users. Consider mobility carefully in these children, if they go out, consider how they travel and why.

### **Prognosis**

Long COVID is not one condition, different subtypes with different clusters of symptoms are emerging. In time, there may be specific management or treatment for some symptom clusters. Symptoms may fluctuate over time and new symptoms may develop as others recede.

NICE guidance advises referring children with significant symptoms following COVID 19 at 4 weeks. Severely affected children claiming child DLA should have been referred to a paediatrician or to a post-COVID hub. Research into recovery from long COVID is being published all the

time, it is thought that recovery and the nature of long COVID may differ depending on the strain of COVID a person had. Our current understanding is that for a population of children who have had COVID and continue to have significant symptoms at 4 weeks many of these will recover and become well by 26 weeks. On average those who are still unwell at 26 weeks will continue to improve but much more slowly. A proportion of them will remain extremely unwell for some years and we don't yet understand their long term prognosis.

Recovery is defined in terms of fewer symptoms of COVID but this might not translate into reduced functional restriction in child DLA. Children who have recovered from their long COVID may not get back to how they were before, and recovery may mean adapting to managing with less energy and ongoing physical symptoms.

### **Award duration guidance**

#### **All ages - children with symptoms for less than 6 months**

Calculate the month of onset of their coronavirus infection, if the child is within 6 months of the onset of their coronavirus infection/long COVID then make an award for 12 months. This is because the prognosis is very uncertain at this stage and significant recovery may occur in the first 26 weeks following onset of the illness

#### **Children aged 12 and over with symptoms for more than 6 months**

Children who remain unwell with little improvement in the level of disability experienced at 6 months are likely to remain unwell for some time, this could apply to new claims or renewals.

Children who remain unwell with little or no improvement - award to age 16.

Children who show significant improvement, for example, starting to attend school or other activities part time for some months **without relapse of symptoms**, consider a shorter fixed term award based on individual medical advice. For example, the child has been unwell for 9 months but has improved significantly in the last 4 months – medical advice might be that continued improvement is likely and a 1 or 2 year award, depending on current level of disability and rate of progress

would be appropriate.

### **Children aged under 12 with symptoms for more than 6 months**

Children who remain unwell with little or no improvement in the level of disability experienced at 6 months are likely to remain unwell for some time, this could apply to new claims or renewals.

Children who remain unwell with little or no improvement - consider awarding to age 12 or for a 5 year period to give time for recovery. Do not make a short award to a milestone age. Consider taking individual medical advice on prognosis.

Children who show significant improvement, for example starting to attend school or other activities for some months part time **without relapse of symptoms**, consider a shorter fixed term award based on individual medical advice. For example, the child has been unwell for 9 months but has improved significantly in the last 4 months – medical advice might be that continued improvement is likely and a 1 or 2 year award, depending on current level of disability and rate of progress would be appropriate.

## **Premature babies**

### **What is prematurity?**

Premature (pre-term) babies are babies born before 37 weeks from the 1<sup>st</sup> day of the mothers last menstrual period (37 weeks of gestation).

There are 770,000 births every year in the UK and 7.1% of these will be premature. That is 54,000 premature babies every year. Most will be only

slightly premature, but an increasing number of moderately or extremely premature babies now survive.

weeks of gestation	percentage
32-37	84%
28-32	11%
24-28	5%

Why does it happen?

50% of premature births have no identifiable cause. Identifiable causes include:

- Multiple pregnancy (for example, twins or triplets)
- early rupture of membranes around the baby (waters breaking)
- preeclampsia (high blood pressure in the mother due to pregnancy which can lead to seizures)
- chronic medical illness of the mother
- intra uterine infection (within the womb, membranes and waters)
- abnormal anatomy of the placenta or uterus
- 

### **Effects and signs**

Very broadly, the difficulties arise because the early baby is not yet ready to function independently out of the uterus. In particular he or she is unready to keep warm, maintain hydration, swallow and digest food, fight infections, breathe, and maintain blood circulation. The earlier the birth, the more under-developed these functions are.

This can have two results: the baby may die even with the best medical support, or the baby may survive with medical support but carry damage from hypothermia, dehydration, undernourishment, infection, lack of

oxygen, or poor circulation or bleeding. One of the most frequently damaged organs is the brain.

## Assessment

Children with prematurity will be seen, assessed and followed up by consultant paediatricians in either the hospital or community setting. The individual follow up, therapy and assessments will be as variable as the condition itself.

See the treatment and management section for further explanation of assessment and therapeutic terms.

## Treatment and management

### Babies born 32-37 weeks

With incubation to keep them warm, and naso-gastric (nose to stomach) tube feeding, most babies in this group survive without long-term harm. Many of them are ready to go home much earlier than they were due to be born because they can mature rapidly. However, every baby is unique and it is possible to suffer bleeding into the brain, serious initial breathing problems or serious infections even at this level of maturity.

### Babies born 28-32 weeks

These babies require the same support as above but, in addition, many of them have immature lungs. Until recently most of them required ventilation after birth. However, if there is time to give steroids to the baby's mother shortly before birth, even babies in this group can be born ready to breathe independently. Most will require fluid and nutrition by vein because their



intestines are too unready. They are more vulnerable to infection. Few will leave hospital more than a week or two before they were due to be born.

### Babies born before 28 weeks

With every week under 28 weeks of gestation, the likelihood of a fatal problem increases. The four problems that most often prevent survival (even with all the measures described above are:

- lungs that are too immature to provide enough oxygen to the baby even with breathing support
- a heart that is too weak to maintain circulation even with support
- an intestine so fragile that it breaks down when food is given, and
- an immune system so weak that it cannot fight off even an ordinary infection

Between the lower limit of viability – roughly 23 weeks gestation – and the 28<sup>th</sup> week of gestation there is an appreciable mortality and morbidity. Thus, depending on the centre, a 26 week baby has a 50% chance of dying (mortality), with a high rate of associated illness (morbidity). All babies born before 28 weeks are destined to spend a matter of months on a special care baby unit.

### Explanation of medical terms

#### Keeping warm

Most babies that are under 2 Kg in weight will be unable to maintain their core temperature in a room environment. In addition to warm rooms and layers and hats, they may be placed in an incubator, which is simply a closed transparent box with a heater and access ports.

#### Maintaining hydration

Premature babies have thin moist skin and lose more fluid (especially below 28 weeks gestation). Most cannot feed themselves to keep up so depend on careful monitoring and replacement of fluid by the medical and nursing teams.

#### Swallowing and digesting food

Few babies below 34 weeks have developed a safe swallowing mechanism – so they must be fed by bypassing the mouth using either a naso-gastric,

oro-gastric, or gastrostomy tube. These reach the stomach through the nose, the mouth, or directly through the abdominal wall respectively.

Even less mature babies may have intestines that are unready to cope with milk even after bypassing the swallow. This is especially true for babies whose growth has been restricted in the uterus so that they are born small even for their gestation (intra-uterine growth restriction resulting in a baby who is small for gestational age). These babies are cared for by introducing premature formula milks or breast milk very slowly and incrementally and by providing additional fluid by vein (IV fluids through a cannula, an umbilical line or a long line). If progress is going to be especially slow then specialised nutrition is given by vein (total parenteral nutrition).

If the load of food is too great, or the baby becomes less resilient due to other changes, the intestine can break down (necrotising enterocolitis).

Even when they are more mature and fully fed, a greater proportion of premature babies suffer with [gastro-oesophageal reflux](#), which may cause symptoms.

#### Fighting infections

Premature babies have underdeveloped natural barriers to infection such as skin, tears, saliva and gastric acid. Their biological immune system (white blood cells and antibodies) is not yet mature. Furthermore, their infection risk is increased by many of the treatments they are given including cannulas, umbilical lines, long lines, catheters (bladder tubes), and endo-tracheal (breathing) tubes.

As a result they are very vulnerable to infections – usually with quite ordinary skin bugs that wouldn't cause harm in mature babies (staphylococcus) but sometimes with unusual bugs or even fungi (candida). Terms used for different sorts of infections include sepsis, septicaemia, line infections, meningitis, pneumonia, urinary tract infections and systemic candidiasis.

#### Breathing and oxygenation

The brain centres that drive and control breathing are not fully mature before gestation is complete. Under 32 weeks of gestation this causes quite predictable apnoeas (breathing gaps of over 10-20 seconds). However,

more mature babies can suffer apnoeas when anything disturbs their general health such as tiredness, infection or gastro-oesophageal reflux.

In order to avoid using a breathing machine, nurses will physically stimulate premature babies suffering with apnoea or provide brief periods of breathing support with a manual device (known as bagging). Caffeine is prescribed as a breathing stimulant.

Where the lungs themselves have not matured sufficient to transfer enough oxygen to the baby's blood stream, the baby can die from or be harmed by hypoxia (not enough oxygen in the blood stream). The following measures are used escalating from one to the next according to the severity of the problem: supplemental oxygen (typically by filling the incubator), pressurised oxygen normally given by making a seal through the nose leaving the mouth free (continuous positive airways pressure), full breathing pressure cycles (ventilation) usually given through a breathing tube into the windpipe (endo-tracheal tube), vibrating high-pressure ventilation (oscillation). Increasingly, there are special modes of ventilation that combine features of each of the above. Very rarely a baby will be given oxygenation by 'bypass' (extra-corporeal membrane oxygenation).

Finally, an artificial or specially produced form of lung-lining fluid (surfactant) can be given down an endo-tracheal tube resulting in quite rapid improvements in a baby's lung function where the baby has not produced the fluid for themself.

### Maintaining circulation

Coldness and dehydration can both result in poor peripheral circulation or even low blood pressure (hypotension) which, in turn can cause damage to the organs not getting enough blood and oxygen. Good basic attention to warming and hydration is important. Sometime a baby will require rapid infusions of a larger volume of fluid (boluses) or the infusion of special medications which increase the speed or power of heart contractions (inotropes).

In a baby with sepsis or necrotising entero-colitis or an extremely premature baby with limited heart muscle, even these measures may not be sufficient to maintain circulation. This results in hypotension and acidosis (a build up of lactic and carbonic acid in the blood) and death or brain damage may be the result.

Circulation may be further compromised by bleeding. The linings of the blood vessels are immature and the blood clotting system is not fully developed so that babies bleed more easily into their lungs, their intestines, their skin and their organs. Bleeding into the brain is a particularly common

problem because it can be triggered by changes in cerebral circulation arising from fluctuations in oxygen and acid levels in the bloodstream. The two commonest causes of circulatory damage to the brain are bleeding into the fluid spaces (intra-ventricular haemorrhage) and poor circulation into the surrounding brain leading to cell death (peri-ventricular leukomalacia).

Finally, the circulatory changes that normally take place after birth to switch off lung bypass and start lung circulation can be delayed or disordered in prematurity. This can lead to a proportion of blood shunting across on a pathway that bypasses the lungs through a duct (patent ductus arteriosus – See: Congenital Heart Disease) or even the persistence of the fully foetal circulation pattern. In these conditions medicines are used to help trigger the natural circulatory changes. Occasionally an operation is used to close a patent ductus arteriosus.

### Continuing medical problems after hospital discharge

All of the following are more likely in a more premature baby or who becomes more unwell whilst in hospital. Of babies born before 27 weeks, 21% will have some degree of learning disability and a further 26% will have a lower than normal IQ but above the cut off for learning disability.

#### Harm to the brain

- Cerebral Palsy
- Learning Disability - see guidance
- Vision or hearing impairment
- Developmental delay
- Poor school performance
- Behavioural/psychological problems such as ADHD
- Epilepsy

#### Harm to the lungs

The fine structure of the lungs is damaged by the combined effects of developing outside the uterus, chemical damage from rich oxygen, and physical trauma from ventilation. The commonest form of harm is called chronic lung disease or broncho-pulmonary dysplasia.

Children with this kind of damage have limited reserve capacity in their oxygenation, and poor lung function which means that they may become

hypoxic, or wheezy, or both with very mild illnesses such as a cold and may require medication and more frequent admissions to hospital.

A small number of children with chronic lung disease are sufficiently well to leave hospital before their lungs are good enough to breathe room air. They may go home needing supplemental oxygen provided from oxygen cylinders, or an oxygen concentrator either all the time or during the night.

In both cases the lung disease is not static and tends to improve very slowly with time.

Other harm

Other long term consequences of prematurity and its treatments include:

- Ear, nose and throat damage due to artificial ventilation
- Skin damage due to infusions that leak into the skin (extravasation injuries)
- Eye damage due to enriched oxygen (retinopathy of prematurity)
- Feeding delay or aversion sometimes necessitating naso-gastric tubes or gastrostomies
- Post operative stomas due to necrotising entero-colitis
- Sleeping difficulties

Note: Almost all of the medical conditions and treatments above are frequently referred to by abbreviations using the first letter of each word.

For example, continuous positive airways pressure is CPAP, endo-tracheal tube is ET tube and so on.

#### Sources of evidence

A report from the hospital or GP may help in determining the level of disability and the likely duration of care needs.

Children in schools may have an SEN/IEP in place, so letters from school, teacher, SENCO or Social Worker (if allocated) may be a useful source of evidence.

Most Child Development Centres provide parents with a report on the child's assessment, which may be a useful source of additional information should this be required.

There may be multiple sources of evidence depending on what, if any, long term complications the child is suffering from.

How long will the needs last?

Most pre-term babies eventually catch up with other children with regard to growth and development during childhood. Survival has improved dramatically, and the focus of management is now to reduce long-term disabilities, especially brain injury. Some of the complications related to prematurity may not be apparent until years after the birth. As previously indicated, cerebral palsy, learning difficulties, disorders of psychological development and behaviour, hearing and visual problems may result from prematurity. High risk infants are followed up closely in outpatient clinic and some are screened for potential problems such as hearing difficulties. Studies have shown that the risks of disabilities are higher in the smallest and most

immature babies and from statistics this represents the smallest number of babies born prematurely.

It is not possible to give generalisations on the duration of needs. This will depend entirely on the particular disability or disabilities for which the child has care and/or mobility needs.

## What you need to know about Sickle cell anaemia

### What is Sickle cell anaemia?

Sickle cell disorders are caused by inherited abnormalities of the blood protein haemoglobin A (Hb A).....

- [Sickle cell anaemia](#)

### What are the effects and signs?

The symptoms and course of sickle cell disease is very variable between individual children.....

- [Effects of Sickle cell anaemia](#)

### How is it assessed?

Diagnosis is usually made in the first year of life by blood tests. Most children are now picked up by the neonatal screening programme.....

- [Assessment of Sickle cell anaemia](#)

### How is it treated and managed?

Children require monitoring and treatment throughout their lives to improve the effects of crises and complications.....

- [Treatment of Sickle cell anaemia](#)

### What evidence is available?

Children who are severely affected by sickle cell disease are likely to receive all of their routine health care .....

- [Evidence](#)

### How long will the needs last?

- [Prognosis and duration of the award](#)

## **What is Sickle Cell Anaemia?**

Sickle cell disorders are caused by inherited abnormalities of the blood protein haemoglobin A (Hb A). Haemoglobin is found in red blood cells. It carries oxygen molecules from the lungs to the rest of the body. In sickle cell anaemia the haemoglobin A molecule is abnormal and the abnormal variant is called haemoglobin 'S' or Hb S and this affects the ability of red cells to travel through the circulation picking up and releasing oxygen. New red blood cells are a normal disc shape; because the sickle haemoglobin protein does not work properly they become deformed. The deformed red cells are sickle shaped when examined under the microscope hence the name 'sickle cell anaemia'.

Abnormally shaped sickle cells are removed from the circulation quickly as they do not function properly once damaged. Normal red blood cells last for about 120 days in the circulation, in sickle cell anaemia, the cells are damaged quite quickly and the average life span of red blood cells is much lower than normal. The reduced life span of red blood cells in sickle cell anaemia leads to two effects: one is anaemia – the haemoglobin count is lower than normal and the other is increased production of cells. High rates of production are needed to try to catch up with destruction of abnormal cells. This can lead to iron deficiency because iron is required by the body to make haemoglobin.

The sickle cells have two main effects -:

The red cells are more fragile than normal and tend to disintegrate more easily than normal cells. This is particularly true if oxygen levels become abnormally low. When the red cells disintegrate in the circulation they literally break apart releasing their contents into the blood plasma (called haemolysis). When many cells do this at once it is called a haemolytic crisis and can be life threatening.

- The second problem relates to the shape of the sickle cells. Once a cell has 'sickled', it is less elastic and is unable to squeeze through small spaces into the capillary beds where oxygen is exchanged. The rigid sickle cells tend to get stuck in very small vessels and block them off. This cuts off the oxygen supply to capillary beds and causes cell death in the affected areas. Over time as more small areas infarct (die due to lack of oxygen) measurable loss of function develops. Examples of organs typically affected by this process include the bones, nervous system, lungs and kidneys

### **Sickle cell disease is an inherited or genetic disorder**

The production of haemoglobin in the body is controlled by genes inherited from both parents. In sickle cell disorders an abnormal gene causes the production of abnormal 'sickle' haemoglobin in red blood cells. It is a



recessive condition – this means that a person must inherit one abnormal gene from each parent to have the condition. If they inherit one normal gene and one abnormal gene the normal gene will enable them to produce enough normal haemoglobin not to have the condition – this is called having sickle cell trait and means the person is carrier of the sickle cell gene. Under very low oxygen conditions a person with sickle cell trait may develop sickling of their red cells. Such conditions include diving underwater at depth, flying at high altitude and general anaesthesia. Precautions against 'sickling' include supplemental oxygen under these conditions – life is otherwise normal with no disabling effects.

If a person inherits the abnormal gene from both parents all their red cells contain the abnormal haemoglobin, and they are described as having sickle cell anaemia. This is because their red cells will have a strong tendency to 'sickle' and this is likely to cause painful tissue damage and anaemia.

### **What are the other types of sickle cell disorder?**

There are other types of abnormal inherited haemoglobin such as haemoglobin C (Hb C) which has a generally milder course and Hb D Punjab and Hb O Arab, which are more severe. Sickle cell genes can also be inherited with beta thalassaemia. Sickle Beta Thalassaemia is a sickle cell anaemia syndrome and children with this condition may be White or Asian, they will suffer from either severe sickle cell disease or a milder form with less frequent crisis depending on the severity of the beta Thalassaemia gene they have inherited. The type will often be clear from the medical evidence provided, for example -:

- sickle beta zero Thalassaemia (severe)
- sickle beta plus Thalassaemia (milder disease)

### **Incidence & Prevalence**

In the UK sickle cell disorders are most commonly found in people of African and Caribbean descent. They are also seen in people from the eastern Mediterranean, the Middle East and India. It is estimated that there are around 10-15000 adults and children with sickle cell disorders in the UK at the present time. About 140 babies with sickle cell anaemia are born in the UK each year.

### **What are the effects and signs?**

The symptoms and course of sickle cell disease is very variable between individual children. In some, the abnormality of the blood has few adverse effects, and the diagnosis is made by chance during routine blood tests. Other children have mild to moderate anaemia (a low red cell count) but function normally most of the time. Others experience frequent painful crises, have debilitating anaemia and develop serious complications.

Sickle cell disease causes episodes of acute illness called crises that will usually be managed in hospital. Over time the cumulative effects of crises

lead to chronic disability because of the tissue damage and loss of organ function that they cause.

In this guidance the acute and chronic effects will be considered separately.

### **Acute Effects**

Sickle cell crises cause episodes of pain affecting the abdomen, chest, back and limbs accompanied by fever, vomiting and malaise. The pain may be severe, the child is unwell, they may have jaundice and are confined to bed for a few days. During the crisis a child will become more anaemic than usual. Some children may develop severe anaemia of such rapid onset that an emergency blood transfusion is needed.

Children with sickle cell anaemia are prone to infections, and crises may be precipitated by a minor infection such as a cold. Infections may rapidly become severe leading to pneumonia, meningitis, osteomyelitis (infectious inflammatory disease of bone) or septicaemia (generalised blood poisoning). These infections may be life threatening.

Sudden and severe damage to the lungs may occur during a crisis and the body is unable to receive sufficient oxygen. This is known as an acute chest syndrome and presents with fever, chest pain and shortness of breath. Acute chest syndrome is a major cause of sudden death, especially in children.

The nervous system may be affected during a crisis leading to seizures, bleeding into the brain (cerebral haemorrhage) or stroke. Children are at highest risk of stroke, around 8-10% of children with sickle cell will have had a stroke. High risk children can be identified using trans-cranial Doppler screening (ultrasound of the blood vessels in the brain). Those at highest risk are put on a blood transfusion programme and this reduces the risk of a stroke happening. Those on a regular transfusion programme will develop the problems of iron overload if not treated preventively with iron chelation therapy. In children who have had a stroke the effects are devastating with both physical and mental disability. Children who have had a stroke and are now “physically recovered” are often left with severe neuro-cognitive defects and have learning disabilities.

Sickle retinopathy (a visual impairment) is caused by the development of new fragile blood vessels in the retina, these can result in retinal bleeds which can present as floaters in the eye or in more severe cases as a retinal detachment due to a severe bleed. This can result in significant damage to the vision and in some cases blindness. The smaller bleeds can be managed with laser therapy to the retina, but the retinal detachment cannot be corrected.

### **Between Crises**

In many children the symptoms of the crisis resolve with appropriate treatment and the child returns to normal after seven to ten days. Some children are persistently anaemic between crises with haemoglobin levels between

50 – 70% of normal. (Normal haemoglobin level 11.4 – 15.5 g/dl). In some children this will cause few symptoms because their body has adapted to functioning with low haemoglobin. In others it will cause fatigue and shortness of breath on exertion. Over the years children who have frequent crises, and those with more severe anaemia, will experience poorer health overall. These children are most likely to develop long-term disabling complications.

### **Long-term complications**

- Arthritis - The head of the femur (thigh bone), that forms part of the hip joint, may be severely damaged (avascular necrosis) during a sickle crisis. The hip joint becomes very painful on movement and ultimately severe arthritis of the hip develops. Joint movement will become very restricted and walking is limited. Avascular necrosis may also occur in the shoulder joint causing pain, deformity and restricted upper limb function. Severe arthritis of the hip or shoulder joint occurs at a relatively young age, and the joint requires artificial replacement. This will be seen in adolescent children and is unusual in younger children
- Risk of infection - Abnormalities of the spleen occur. One of the roles of the spleen is to remove red cells, which have a normal life span of about 120 days, from the circulation. In sickle cell anaemia excessive numbers of 'sickled' cells accumulate in the spleen causing enlargement of the organ in childhood. Ultimately the normal structure of the spleen is affected, shrinking in size and its functions become impaired. The normal spleen plays a role in protecting the body against infection. As a result the child with sickle cell anaemia is more prone to infections and more likely to develop serious infections. The commonest cause of death in children below the age of 5 years is infection.
- Breathing difficulties - Repeated damage to the lungs leads to chronic pulmonary hypertension, a condition that causes progressive shortness of breath and may lead to heart failure. This condition takes time to develop and is more common in adolescents than young children.
- Kidney failure - Over the years, permanent damage to the blood supply of the kidneys causes impaired renal function that may lead to renal failure.
- Cognitive impairment - Related to 'silent' infarcts (resulting from obstruction of circulation) this is present in 20% of adults with sickle cell. This will result in difficulty in understanding or

undertaking specific tasks and can result in relatively simple things like counting or reading becoming difficult to do.

- Blindness - May result from bilateral sickle retinopathy.
- Problems of iron overload - Related to regular transfusion treatment.
  
- [Care & mobility needs](#)
- [Indicators of severe functional restriction](#)

### **How is it assessed?**

Diagnosis is usually made in the first year of life by blood tests. Most children are now picked up by the neonatal screening programme.

### **Screening for sickle cell disease**

In communities in the UK where there is a high incidence of sickle cell disorders screening tests are available to identify the condition. Screening is offered to people where there is a family history of sickle cell, to women of reproductive age, to couples planning a pregnancy, to people having general anaesthetics and at antenatal clinics. Education and genetic counselling are offered to people and families at risk.

Screening of new-born babies is undertaken to enable early diagnosis of sickle cell anaemia in affected infants.

### **How is it treated and managed?**

Children require monitoring and treatment throughout their lives to control the effects of crises and reduce complications of the disease. Parents will need to know about and recognise the signs of sickle cell crisis and be

prepared to seek immediate medical treatment. They may need to manage minor crises at home.

### **All children will require the following long-term treatments**

A number of measures are undertaken to maintain health and to try to avoid crises. All children will require -:

- Balanced diet, adequate fluid intake and avoidance of cold,
- Regular folic acid supplements to replace loss by a process called haemolysis,
- Continuous oral penicillin or amoxycillin (twice daily) to reduce risk of infection from age 3 months onwards for life,
- Prompt treatment of infections,
- Full courses of vaccinations and flu vaccination.

Children on a programme of blood transfusions will need additional treatment at home. The parent/carer may need to administer iron chelation therapy (a treatment using a substance that is able to bind with excess iron in the body and enable it to be excreted) at home for those children receiving regular blood transfusions.

### **Some children may require the following long term treatments**

- Hydroxycarbamide (hydroxyurea), a drug that affects red cell production may be used to reduce the risk of crises in some children. This is often used in children especially if they have recurrent and severe crises especially acute chest syndrome
- Analgesic drugs e.g. paracetamol, co-codamol, dihydro-codeine, tramadol and anti-inflammatory drugs are prescribed for bone pain and arthritis. Severely arthritic joints may be suitable for artificial joint replacement.
- Treatment of kidney failure including renal transplantation may be necessary. See adult Kidney Disorders guidance.
- Bone marrow transplantation has the potential to cure the condition. However it has a relatively high mortality of 5 – 10% and is only suitable for some people.

### **Treatment of sickle cell crisis**

All but the mildest crises will be treated in hospital. During a crisis a child will need adequate pain relief, to be well hydrated, to keep warm and to rest for a few days. Some will require strong opiates such as morphine or

diamorphine for pain relief and these drugs may need to be given by injection. Moderately potent analgesics such as tramadol, dihydrocodeine or co-codamol are used. Intravenous fluids and drugs to control vomiting may be necessary.

Painful crises usually last a few days, and as the acute symptoms settle down children will be encouraged to mobilize and get back to normal activities. Children are likely to receive their acute treatment as a hospital in-patient with further recovery taking place at home. Complete resolution of symptoms is likely to take seven to ten days. There are a small group of children who are very severely affected and these may take longer to recover from crises, for example spending 3-4 weeks in hospital on each occasion.

Some children may be able to manage less severe crises at home with care from parents and advice and/or home nursing from specialist sickle cell teams. Others may routinely be admitted to hospital for a few days. Indications for admission include severe pain, oxygen administration, intravenous fluids and treatment of infections such as pneumonia or septicaemia (generalised blood poisoning). Hospital admission is also needed for severe anaemia requiring blood transfusion.

### **Some children will require iron chelation therapy at home**

Regular blood transfusions (transfusion therapy) are used for children at risk of severe complications such as stroke. The complication of repeated blood transfusions is iron overload, which can be life threatening if it is not treated.

Iron overload is treated with a chelating agent. It is time consuming and has unpleasant side effects. It is likely to be given at home by the parents with outreach support from the treating specialist team.

- Chelation therapy may be given as an infusion using Desferrioxamine subcutaneously or intravenously. Depending on the severity of iron overload the infusion can be given over 12 hours 5-7 days a week or 24 hours a day every day. A common regimen is 12 hourly infusions 3-5 times a week at home. Constant treatment is likely to be given in hospital as part of the treatment of life threatening complications of iron overload such as heart failure. Complications of iron overload such as

heart failure can occur in sickle cell anaemia and although rare it is potentially fatal.

- An oral chelation agent is available – deferiprone – this is given three times a day 7 days a week. This treatment has severe side effects including -:

Neutropenia (reduced white blood cell count) - this can lead to life threatening septicaemia – weekly blood test monitoring is required during treatment.

- Arthropathy and joint destruction. Deferiprone is not licensed for use in sickle cell anaemia but some doctors use it when the alternative treatments have failed or are not available.
- Another oral chelation agent called Deferasirox is also available and this has different but equally severe side effects including -:

Kidney toxicity and reduced renal function in 30%

Liver toxicity in 2%

Severe diarrhoea

Chelation treatment of one sort or another is started usually within one to two years of starting regular transfusions. The amount of iron in the body is monitored regularly by special investigations. It is difficult to comply with these treatments because of side effects, the time-consuming nature of treatment and no immediate and obvious benefit from adhering to them

carefully. Later problems with iron overload increase mortality if the young person defaults from chelation therapy.

### What evidence is available?

Sources of evidence	Information Provided	Limitation of Evidence
Parent / carer	First source of information and associated evidence.	Does not have specialist knowledge.
GPFR	The GP will be able to confirm the diagnosis.	All treatment for both sickle cell related problems and general health care are likely to be received via the treating hospital – the GP may not have any details about the child’s condition or care required. The GP will be most likely to provide information about very mild cases not having routine hospital treatment.
Consultant Haematologist / Clinical Nurse Specialist	Children who are severely affected by sickle cell disease are likely to receive all of their routine health care and their sickle cell related care through the haematology unit. This will always be the best source of information on diagnosis and disabling effects.	In a mild case there may be little information available from the hospital. The haematologist will always be able to provide supporting evidence in cases where disability is claimed or the condition is severe.

### How long will the needs last?

This guidance covers:-

Sickle cell anaemia
Haemolytic disorder - Other / type not known

Awards related to therapy are recommended to last until therapy ends or a child could be expected to manage their own therapy. Most children should



be able to manage their own therapy and monitor their condition by age 12-14. Children on chelation therapy could be expected to manage this substantially themselves by age 14. Children on transfusion therapy undergoing chelation therapy at home are likely to remain on this treatment.

<b>Age at date of claim</b>	<b>Award Period</b>
0-14 – currently on iron chelation therapy	Award to age 14 (or for one year whichever is longer)
0-15 – not on iron chelation therapy but having frequent crises	Initial award for 5 years followed by further awards for 5 years or to age 16 (whichever is sooner)
0-15 – joint problems present	Award for 2 years or until joint surgery is performed (whichever is sooner)
0-15 – undergoing rehab / treatment due to stroke complications	Award for 2 years

Children not on chelation therapy but who require care from their parents because of frequent crises more than twice a month should be reviewed every 5 years as the frequency of crises can change over time.

Awards related to arthritis and avascular necrosis should be reviewed after joint surgery or after 2 years whichever is sooner as treatment to improve joint mobility is expected to be successful.

Children awarded entitlement after a stroke should be reviewed after 2 years as a substantial number could be expected to have improved after 2

years. If further medical evidence at this stage shows that they still have needs then indefinite awards are recommended.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

## **Care and mobility needs**

### **Care**

Early diagnosis and treatment, especially of infection, improves prognosis. Children born in the UK will usually be identified by screening at birth. This means that long term complications can be prevented enabling normal development for many.

The aim of treatment is for the child to lead a normal life, attend school and enter adulthood with few or no complications of the disease. Despite this, up to 8% of children have had strokes by the age of 14 years. A higher number of children are at risk of stroke and will be treated with regular blood transfusions to prevent it.

Ongoing care of children includes -:

- Monitoring the child's condition and seeking medical help when appropriate to do so.
- Keeping the child warm, well nourished and hydrated to reduce the frequency of crises
- Ensuring child takes medication such as folic acid and penicillin every day as prescribed
- Managing minor crises at home

The frequency of crises and whether they are managed in hospital or at home will be important information to request from the treating specialist. Frequent crises will require more care but supervision will not prevent a crisis from occurring.

Most children will be unwell and in bed for up to week after a crisis. There is likely to be a period of recovery at home after the crisis before the child returns to normal activities and school. Significant care on a daily basis is

likely to be given if crises occur more frequently than twice a month on average. Children with frequent crises are also more likely to have developed some of the long-term complications of the condition such as a stroke and epilepsy - see [Epilepsy](#) guidance. Children who have infrequent crises are likely to take over the majority of their care in the teenage years. Children who have frequent crises i.e. more than twice a month are likely to continue to require care in relation to their crises.

#### Care needs related to chelation therapy

Some children will be on transfusion therapy. This means they will have regular blood transfusions at the hospital to prevent the long term complications of Sickle Cell Disease. In order to prevent a life threatening condition called 'iron overload' they will need to have iron chelation therapy at home. The chelation treatment is unpleasant and a parent will need to administer the treatment or supervise administration of the treatment to make sure that it is taken as prescribed.

Chelation therapy may be given as an infusion using Desferrioxamine subcutaneously or intravenously. A common regimen is 12 hourly infusions 3-5 times a week.

Another chelation agent called Deferasirox is available in tablet form and this has different but equally severe side effects including -:

- Kidney toxicity and reduced renal function in 30%
- Liver toxicity in 2%
- Severe diarrhoea

An oral chelation agent is available called deferiprone. This is given three times a day 7 days a week in tablet form. This treatment has severe side effects including neutropenia (reduced white blood cell count). This can lead to life threatening septicaemia – weekly blood test monitoring is required during treatment.

Parental supervision of each dose will be required to ensure children comply with the treatment. Older children may be able to administer their own chelation therapy. Children around the age of 12 to 14 years of age may be expected to take their medication independently but many parents will

continue to supervise and monitor this either due to concerns about compliance or due to learning difficulties that their child might have.

Long term complications include -:

- Stroke - Children may have mobility difficulties related to spastic paralysis of one leg or difficulties with balance. Other problems that could be caused by a stroke include epilepsy, cognitive difficulties and behavioural problems all of which may mean more monitoring or care and attention or supervision is required. Cognitive problems after stroke are common in children with Sickle Cell but permanent mobility problems are much rarer.
- Kidney failure – end stage renal failure is likely to require renal dialysis and this may be carried out at home or at the hospital. CAPD (Chronic ambulatory Peritoneal Dialysis) is likely to be done overnight, most nights of the week, at home. The parent will need to set up the dialysis equipment and be available to check it or adjust it during the 8-12 hours it takes during the night. Haemodialysis is done three times a week and takes around 4-6 hours to complete; it can be done at home or in the hospital.
- Arthritis - Avascular necrosis of the shoulder or hip joints can cause severe arthritis – it would be unusual in childhood for this to lead to persistent severe disability as severe arthritis takes some years to develop after the avascular necrosis event. Joint replacement is an option for these children once they have finished growing so disability is not expected to be permanent. Severe arthritis of the hip may limit walking distance. Severe arthritis of the shoulder will restrict the range of movement around the shoulder and may make dressing, washing and styling hair more difficult as these activities involve lifting both arms over the head.

## **Mobility**

In the typical case, ability to walk long distances is either normal or reduced by anaemia and breathlessness. Very few children with sickle cell disease have difficulties walking short distances. The complications of the condition that may cause difficulty walking short distances include severe arthritis of the hip due to avascular necrosis and neurological damage following a

stroke. Recovery of walking ability is usual after a stroke in children and severe arthritis is treated with joint replacement in severe cases.

### **Indicators of severe functional restriction**

The following difficulties are suggestive of a child with a severe functional restriction -:

On iron chelation therapy

- Persistent neurological deficits following stroke or cerebral haemorrhage
- Seizures and epilepsy
- Long term lung damage, including chronic pulmonary hypertension, causing breathing difficulties
- Enlarged heart (cardiomyopathy) and heart failure
- kidney damage
- visual defects

## **What you need to know about Speech & Language disorders**

### **What are Speech and Language disorders?**

Communication depends on three things: senses to receive .....

[Speech & Language disorders](#)

### **What are the effects and signs?**

In these children, communication .....

[Effects of Speech & Language disorders](#)

### **How is it assessed?**

In infancy speech, language and communication needs .....

[Assessment of Speech & Language disorders](#)

### **How is it treated and managed?**

Following evaluation and diagnosis, the Speech and .....

[Treatment of Speech & Language disorders](#)

**What evidence is available?**

The role of the speech and language therapist is varied and includes .....

**Evidence**

How long will the needs last?

**Prognosis and duration of the award****Speech, language and communication needs**

Communication depends on three things:

- senses to receive communicated information
- language processing in the brain
- a verbal or non-verbal means to transmit information

An impairment in any of these three elements leads to speech, language and communication needs (SLCN). Needs that arise due to hearing impairment, or visual impairment, or both are addressed as part of the guidance on that particular sensory impairment and are not covered here.

Also, there is a small group of children who are able to communicate but who choose not to do so however, 'Elective/selective mutism' is not covered in this guidance.

**What are the effects and signs?****Language delay**

In these children, communication emerges with a normal pattern and sequence, but more slowly than average. By definition this is extremely common (50% of all children will be at least slightly slower than average) so the prevalence quoted depends on the degree of delay. It is more common in children who are offered less communication as they develop. This has an excellent prognosis from a language perspective, but can be an early marker of intellectual impairment – See: [Normal Paediatric Development Milestones](#) table for information on the usual timing of language development.

**Speech sound disorder**

This is a 'basket' category that includes a number of disorders affecting the production of speech sounds in the oropharynx (mouth and upper throat) but excluding problems with the larynx (voice box). Speech sound production can be impaired in dysarthria, dyspraxia, articulation disorder, and structural problems with the mouth, tongue and throat (oro-pharynx) –

See: Cleft lip and palate guidance for information on the most common structural oro-facial disorder.

## Language disorder

Communication emerges in an abnormal pattern or sequence. There may be anomalies in the development of one or more of phonology, syntax, semantics, word-finding, communication pragmatics, and verbal memory / learning (each of these has a technical specialist meaning). Language disorder shares many risk factors with the other 'specific learning disorders' such as ADHD & dyslexia; it is more common in boys, relatively deprived groups, children of parents with similar disorders, and following complications of pregnancy or birth. It is a lifelong problem where the aim of treatment is to optimise potential and limit disability.

## **Developmental language disorder**

Sometimes called 'specific language impairment', this term applies to those children with language disorder in the absence of an associated medical condition.

## **Language disorder associated with another medical condition**

This term is used when the language disorder is part of another medical condition. Almost any disorder affecting the brain can affect language processing – including cerebral palsy, acquired brain injury, neurodegenerative disorders and epilepsy. In addition, the Autistic Spectrum Disorders (ASD) are characterised by a specific pattern of language disorder where social communication – especially the semantic and pragmatic aspects of language are particularly affected (see below). See: Autistic Spectrum Disorders guidance for information on this group of conditions.

## Social (pragmatic) communication disorder

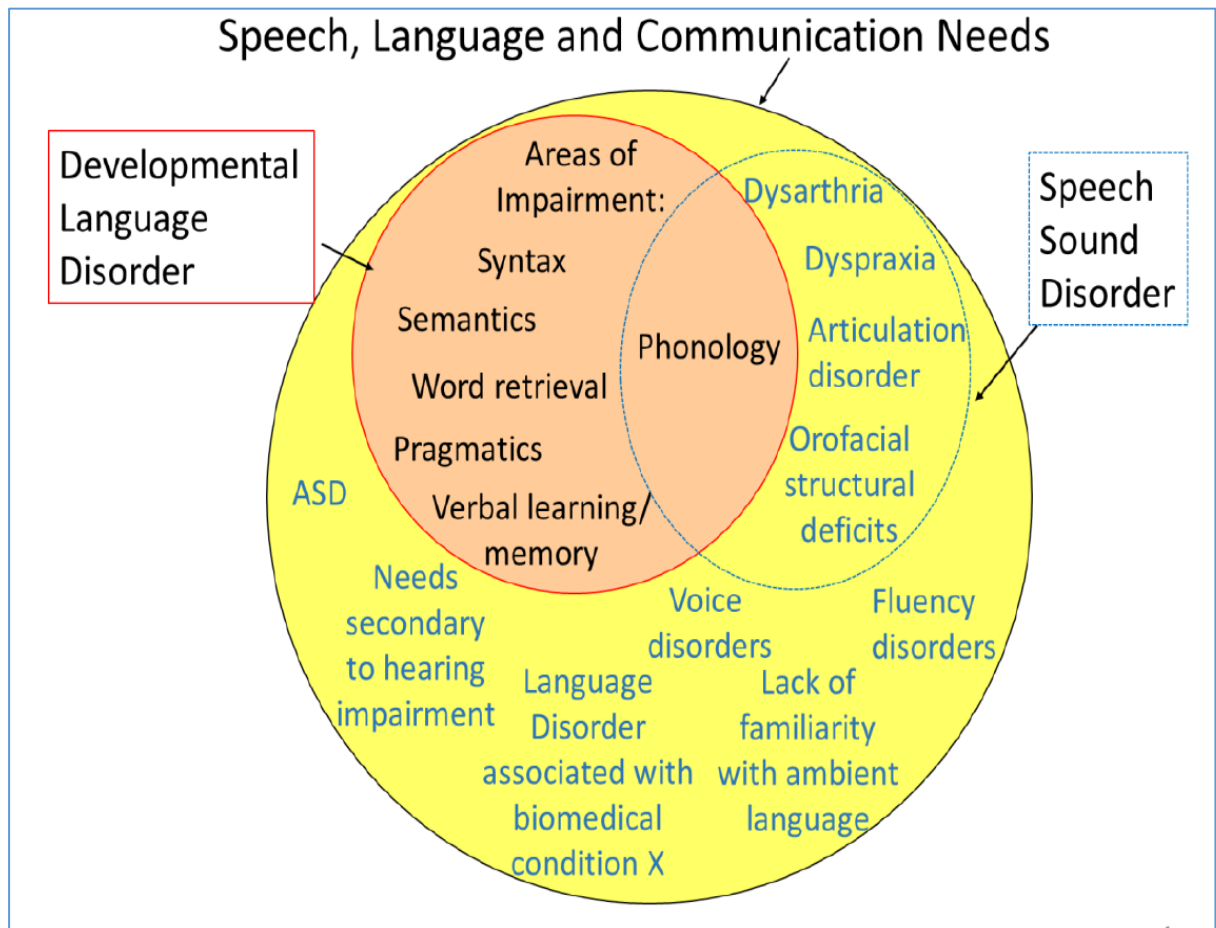
This is diagnosed in children who have the communication profile typical of a child with ASD, but do not have the other features of the condition. These include persistent difficulties in the social use of verbal and nonverbal communication, especially for social purposes. They show impairment in the ability to change communication to match context or the needs of the listener, and difficulties following rules for conversation and storytelling. They

also have difficulties understanding what is not explicitly stated and discerning the nonliteral or ambiguous meaning of language.

Childhood-onset fluency disorder (stuttering)

This is a disturbance in the normal time patterning and flow of speech and, characteristically, causes anxiety in the speaker.

Figure 1: a classification of speech, language and communication needs



Bishop DV, Snowling MJ, Thompson PA, Greenhalgh T, CATALISE-2 consortium (2016) CATALISE: a multinational and multidisciplinary Delphi consensus study of problems with language development. Phase 2. Terminology. PeerJ Preprints4:e2484v1 <https://doi.org/10.7287/peerj.preprints.2484v1>

### How is it assessed?

In infancy speech, language and communication needs (SLCN) may present as limited responses to sounds, while toddlers & pre-schoolers may have delays in producing first words and word combinations and difficulty interacting with peers. School age children may have difficulties following directions, attending and comprehending oral and written language, and problems producing narratives and using language appropriately in social contexts. Parents are often the first to notice. A timely diagnosis is important



because it is easier to learn language and communication skills before the age of five than later in life.

Primary health services or a paediatrician may be confident to make a provisional diagnosis of language delay. But if delay is substantial, or there is any suspicion of a speech sound disorder or a language disorder then the child will be referred for specialist speech and language assessment.

Evaluation procedures are usually performed by a speech and language therapist; however a wider team may be involved. Other members of the team might include educational psychologist, audiologists, paediatricians, occupational therapists, teachers, social workers and health visitors. This is especially the case if there is an associated medical condition.

Comprehensive assessments are needed to identify language strengths and areas of impairment and how this has affected communication, social participation and well-being.

A comprehensive assessment may involve:

- standardised tests (for example, Clinical Evaluation of Language Fundamentals [CELF], Renfrew & Reynell Developmental Language Scales or Test for Reception of Grammar [TROG])
- participation of the child, family and regular caregivers' informal measures such as observation, discussion, questionnaire and checklists
- account taken of a range of contexts: the classroom, playground and home as well as clinical setting – to ensure that children's functional communication ability is judged

### **How is it treated and managed?**

Following evaluation and diagnosis, the Speech and Language Therapist (SALT) usually draws up a treatment plan specific to the needs of the child. This may be linked to the child's Individual Education Plan (IEP) and outlines the goals/objectives of therapy and targets. These are usually set jointly with practitioners and/or parents/carers.

A therapy programme is delivered in an environment familiar to the child, for example, through parents (at home), nursery staff or teaching staff. This helps the child to develop their skills throughout the day. SLT s may also

provide direct individual or group therapy, but this is not necessarily the norm; it will depend on the child's needs.

In some cases, which the SALT assesses as mild, no specific programme may be set up, but the SALT will keep the child on the caseload in order to review his/her language development at intervals.

### Speech and Language Therapy

The wide variety of disorders requires a wide variety of approaches to address the nature of the problem.

### Alternative and Augmentative Communication

Augmentative and Alternative Communication (AAC) refers to any system of communication that is used in addition to or to replace speech in order to help children communicate. AAC aids cover a range of technologies, and the AAC method / system is tailored to the child's needs. These aids may improve social interaction and academic performance. They may also accelerate verbal language development:

- Non-assisted AAC uses the body to convey language (gestures, sign language).
- Assisted AAC requires tools/equipment (picture and symbol communication boards, electronic devices and such).

### Social Communication Disorder (SCD)

#### Diagnostic criteria

SCD causes persistent difficulties in the social use of verbal and nonverbal communication as indicated by all of the following:

- deficits in using communication for social purposes, such as greeting and sharing information, in a manner that is appropriate for social context
- impairment in the ability to change communication to match context or the needs of the listener, such as speaking differently in a classroom than on a playground, talking differently to a child than to an adult, and avoiding use of overly formal language
- difficulties following rules for conversation and storytelling, such as taking turns in conversation, rephrasing when misunderstood, and knowing how to use verbal and nonverbal signals to regulate interaction
- difficulties understanding what is not explicitly stated (for example, making inferences) and nonliteral or ambiguous meaning of language

(for example, idioms, humour, metaphors, multiple meanings that depend on the context for interpretation)

The onset of the symptoms is in the early developmental period (up to the age of 8). However, deficits may not become fully clear until social communication demands exceed limited capacities.

The deficits result in functional limitations in effective communication, social participation, social relationships, academic achievement, or occupational performance, individually or in combination.

The symptoms are not caused by another medical condition or to low ability in the areas of word structure and grammar. Also, the symptoms are not explained by medical conditions such as Autistic Spectrum Disorder, Learning Disabilities, Global Developmental Delay or another mental disorder.

Note: If any of the medical conditions stated in the paragraph above is diagnosed, any speech and language problems should be accepted as being due to that condition and the relevant disability code used. Do not consider a separate diagnosis of a Speech and Language Disorder and do not use the 'Speech or language disorders – Other/type not known' disability code F02.

**What evidence is available?**

Sources of evidence	Information Provided	Limitation of Evidence
Parent / Carer	First source of information and associated evidence.	Does not have specialist knowledge.
Speech and language therapist (SALT) Paediatrician Occupational Therapist	<p>The role of the speech and language therapist is varied and includes -:</p> <p>Assessment:</p> <p>Conducting evaluations of children's speech and language skills within the context of total development</p> <p>Administering standardised tests and scales and look for milestones in speech and language development</p> <p>Identifying communication problems and the best way to treat them</p> <p>Helping teachers and health care professionals identify children who are at risk</p> <p>Treatment:</p> <p>Providing therapy to children individually or in a group</p> <p>Training and advice for parents/carers and other service providers (health, social work and education)</p> <p>Provision of programmes of work and ways of supporting the child in different environments and by different people</p> <p>Assessment and provision of communication aids and resources</p> <p>Involvement with educational and transition planning</p>	It is important to determine who is regularly managing the child's condition to enable contact with the appropriate healthcare professional.

Educational Psychologist	May be a part of multidisciplinary team managing child's condition. Perform assessments as well as involved in delivering interventions.	Same as above
Audiologist	May be involved in cases where the language disorder is associated with a hearing problem.	Same as above
General Practitioner	May have been involved at initial referral stages; may be helpful in providing information in cases where the child is no longer under secondary care.	Same as above
School	Teachers may have been involved at initial referral stages; often collaboration with SALTs and teachers during assessment/intervention. Particularly helpful in providing information on ongoing academic/social impact or in cases where the child is no longer under secondary care.	Same as above

### **How long will the needs last?**

The duration of functional limitation for those with significant disability will depend on their age and the duration of therapeutic input.

The following table is to be followed if an award is made. It is not a statement of intended awards for individual categories.

Age at date of claim	Length of Award
0-4 years	Up to 5 <sup>th</sup> birthday (or 1 year award, whichever is greater)
5-8 years	Up to 9 <sup>th</sup> birthday (or 1 year award, whichever is greater)
9-11 years	Up to 12 <sup>th</sup> birthday (or 1 year award, whichever is greater)
12-16 years	2-year award (or up to 16 <sup>th</sup> birthday, whichever is greater)  Note: If entitlement is re-assessed at renewal when the child is 12 or older and eligibility continues award until age 16 (day before the 17 <sup>th</sup> birthday).

You must take all information into account when considering the duration of care and mobility needs. You must base the duration of care and mobility needs on the particular circumstances of the child.

#### Care and Mobility considerations

Care and mobility needs largely depend on the cause of the disorder. Whilst they may have ongoing difficulties, most children will have little or no disability (in terms of DLA threshold). The children most likely to qualify for an

award are those with an associated condition such as cerebral palsy or ASD.

Children who have a co-morbid condition such as autism, moderate to severe hearing impairment or a diagnosed behavioural problem are also more likely to have significant functional communication problems.

## What you need to know about Spina bifida

<p><b>What is Spina bifida?</b></p> <p>This is the most common neural tube defect (NTD) in which there is...</p> <ul style="list-style-type: none"><li>• <a href="#">Spina bifida</a></li></ul>
<p><b>What are the effects and signs?</b></p> <p>The primary neurological impairments in children with.....</p> <ul style="list-style-type: none"><li>• <a href="#">Effects of Spina bifida</a></li></ul>
<p><b>How is it assessed?</b></p> <p>Because of the complex effects of neural tube defects there are many potential investigations .....</p> <ul style="list-style-type: none"><li>• <a href="#">Assessment of Spina bifida</a></li></ul>
<p><b>How is it treated and managed?</b></p> <p>Surgical closure of the defect is undertaken shortly after birth to prevent .....</p> <ul style="list-style-type: none"><li>• <a href="#">Treatment of Spina bifida</a></li></ul>
<p><b>What evidence is available?</b></p> <p>The Paediatrician / Specialist nurse will be able to provide details of symptoms, investigations and treatment / management .....</p> <ul style="list-style-type: none"><li>• <a href="#">Evidence</a></li></ul>
<p><b>How long will the needs last?</b></p> <ul style="list-style-type: none"><li>• <a href="#">Prognosis and duration of the award</a></li></ul>

### What is Spina bifida?

#### Neural tube defects

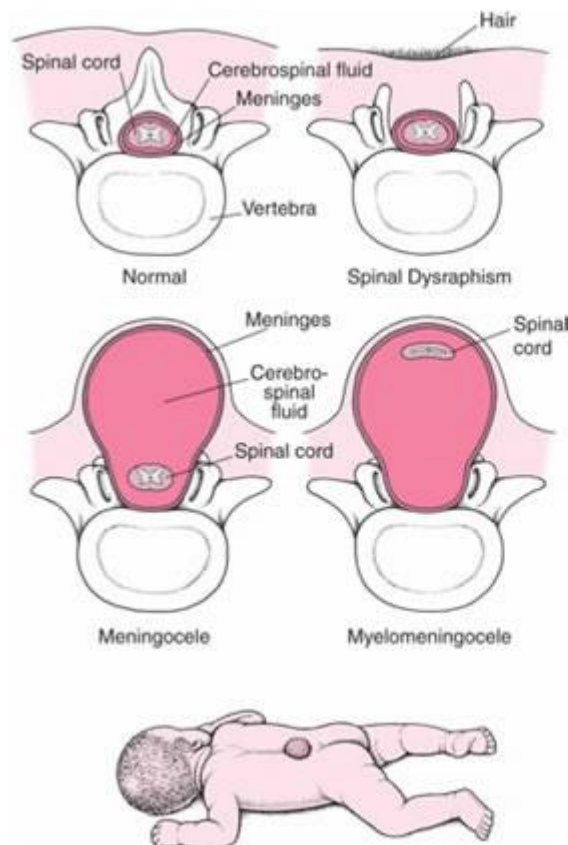
The neural tube is the structure in an embryo from which the brain and spine are formed. The neural tube normally closes spontaneously. If spontaneous closure does not occur, a neural tube defect results.



There are 3 major neural tube defects (NTDs) – spina bifida, encephalocele and anencephaly. This guidance covers only spina bifida.

## Spina bifida

This is the most common NTD in which there is abnormal development of the coverings of the spinal cord. There are several varieties of spina bifida.



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Spina bifida aperta (open neural tube defect)

## Myeloschisis / Rascischisis

This is malformation of the spinal cord with no normal spinal cord development below the lesion and no normal covering of the neural structures. This is a very significant abnormality akin to a complete spinal cord injury with

abnormalities of bladder, bowel and lower limb function. Some will have hydrocephalus and associated congenital brain abnormalities.

### **Open myelomeningocele**

The spinal cord is open to the surface (placode) but has some normal anatomical appearances below the lesion. However, in the majority of these lesions there is very poor function below the level of the lesion. As for Myelomeningocele / Spina bifida there is an association with hydrocephalus and congenital abnormalities of the brain.

### **Dermal sinus tract**

There is abnormal preservation of a tubular connection of skin tissue onto or into the spinal cord. They are commonly associated with dermoid cysts (a sac like growth that contains structures such as hair, fluid, teeth, or skin glands that can be found on or in the skin) and can result in severe inflammatory damage to the spinal cord and infection similar to meningitis. Spinal cord function may be normal but has the potential for late deterioration at any time in life.

Spina bifida occulta (closed neural tube defect)

### **Closed meningocele**

This is like an open myelomeningocele but in a closed meningocele the skin is closed over the surface.

### **Meningocele**

There is a cystic appearance on the surface of the spine with no spinal cord material in it, due to a cyst of the membranes lining the spinal cord and a defect in the coverings (bone, muscle and skin). Spinal cord function is usually minimally affected. These can have both normal skin cover and no skin

cover. They can both result in tethered cord syndrome, as can any of the spina bifida abnormalities.

### **Lipomyelomeningocele**

There is usually a connection of fatty material, blending into the subcutaneous fat through a defect in the dura (membrane covering the spinal cord), bone and muscle, attached to the spinal cord. Spinal cord function can vary from normal through to complete lack of function below the lesion.

### **Diastematomyelia**

There is a defect of the failure of the spinal canal to develop correctly with two canals and the cord split, usually asymmetrically, by a bone or cartilaginous spur or merely a dural fold. This tethers the cord causing traction injury that may be asymmetrical.

### **Fatty filum terminale**

There is abnormal fatty thickening and shortening of the filum terminale (a long slender strand of connective tissue extending from the end of the spinal cord to the termination of the spinal canal) that causes traction on the spinal cord that causes tethered cord syndrome. People with this may have normal function or have subtle abnormalities such as leg or back pain.

### **Anterior sacral meningocele**

There is dysraphism (defective closure of the neural tube) anteriorly in the sacrum with a significant defect causing mass effect on pelvic structures. When associated with a scimitar shaped sacrum and anorectal abnormalities, it is called Currarino triad.

There are many mixed up versions of these abnormalities that can cause all manner of problems.

### **Tethered cord syndrome**

Traction injury to the spinal cord, which is usually subtle and long standing in onset, with possible abnormalities in bladder, bowel and renal tract, lower

limb defects (leg length inequality, club foot, dislocated hip) and scoliosis, in addition to pain in the back and/or legs.

At least 50% of people with an anoctal abnormality have a tethered cord problem too. The majority of people with spina bifida have an additional congenital abnormality. Most will need significant surgical input over many years.

- [Associated Impairments](#)
- [Incidence and Prevalence](#)

### **What are the effects and signs?**

#### **Primary neurological impairments**

The primary neurological impairments in children with a meningomyelocele are paralysis, sensory loss and a Chiari II malformation (if present) with associated hydrocephalus.

#### **Paralysis and sensory loss**

The vertebral column consists of 7 cervical (C1 – C7), 12 thoracic (T1 - T12) and 5 lumbar (L1 - L5) vertebrae connected to the sacrum and coccyx

at the base of the spine. Spinal nerves leave the spinal cord and pass out from holes in the adjacent vertebrae.

Cervical nerves supply movement and feeling to the neck, arms and upper trunk. Thoracic nerves supply the trunk and abdomen. Lumbar and sacral nerves supply the legs, bladder, bowel and sexual organs.

The extent of paralysis and sensory loss depends upon the location of the spinal defect, as paralysis and sensory loss occur below that point.

<b>Level of defect</b>	<b>Degree of paralysis</b>
Thoracic (T1 - 12 ) or upper lumbar (L1 - 2) level	Paralysis affects the legs and variable weakness and sensory loss in the abdomen and lower body region.
L3 level	Children can usually flex their hips and extend their knees but their ankles and toes are paralysed.
L4 or L5 level	Children can usually flex their hips, extend their knees and flex their ankles but have weak or absent ankle extension, toe extension and hip extension.
Sacral level (low back/buttocks region)	Children usually only have mild weakness of their ankles or toes.

All children with meningomyelocele have sensory loss that is greater on the back of the legs than the front. Most affected children have sensory loss around the anus, genitalia and feet.

Weakness and sensory loss may be worse on one side of the body than the other.

Spina bifida is associated with upper limb dysfunction, for example hand weakness and poor fine motor control and coordination.

### **Chiari malformation and hydrocephalus**

Almost all children with a meningomyelocele above the sacral level have a Chiari II (used to be called Arnold Chiari) malformation of the brain. This is a downward displacement of the brainstem (the part of the brain that

connects the brain and the spinal cord) and cerebellum into the neck from the skull.

The Chiari II malformation is associated with symptoms caused by compression of the brainstem. Depending upon the age of the child these may include headaches, sensory disturbance, visual disturbance with double vision or nystagmus (uncontrollable jerking movements of the eyes), swallowing problems, alteration in speech, poor coordination on walking, choking, breath holding attacks, brief recurrent respiratory arrests (apnoeic episodes), and opisthotonos (holding the head forced backwards).

[Hydrocephalus](#) occurs in the majority of children with a Chiari II malformation.

(90% of children with T1 – T12 or L1, L2 defect; 80% if L3, L4 defect and 65% if L5 or sacral defect).

### **How is it assessed?**

Because of the complex effects of neural tube defects there are many potential investigations that can be carried out. The most common investigations include:

#### **Prenatal diagnosis**

Routine screening is undertaken for levels of alpha fetoprotein (AFP) in maternal serum between the 16<sup>th</sup> and 18<sup>th</sup> week of pregnancy. If the level of serum AFP is raised, an ultrasound scan of the baby's head and back is performed. If following ultrasound there is a strong suspicion of a neural tube defect (NTD), amniocentesis is performed and the levels of AFP and another substance called acetylcholinesterase (ACH) in amniotic fluid are measured. Chromosomal analysis of the amniotic fluid is also undertaken to exclude chromosomal syndromes that are associated with NTDs, such as [Down's syndrome](#). If the levels of AFP and ACH are raised and the ultrasound scan is abnormal, it is very likely that the baby will have a NTD. The results of these tests can be used to discuss the option of a therapeutic termination of pregnancy with the parents. If the parents decide not to have a termination of pregnancy, the results can be used to plan for the future

needs of the child, for example, delivery in a specialist neonatal unit with facilities for early closure of the defect.

A closed NTD may give normal enzyme levels and a normal ultrasound scan.

### **Mobility impairment**

Formal gait analysis using video cameras and sensors linked to computers may be used to decide on the best orthopaedic treatment, including bracing and surgery.

### **Cognitive impairment**

Because children with meningocele may have a number of cognitive impairments, formal neuropsychological testing can be useful to evaluate any problems and plan treatment.

### **Urinary dysfunction**

Beginning in early infancy, regular ultrasound of the bladder and kidneys should be undertaken to detect abnormalities. Bladder function can be assessed by performing urodynamic investigations, including cystometry in which fluid is injected into the bladder and the pressure is measured. Monitoring for urinary tract infections, which occur in at least half of children with meningocele, should also be undertaken.

### **Neurological deterioration**

Children who experience neurological deterioration should be investigated to identify any underlying cause.

### **Referral**

It is routine for all children with spina bifida to be referred to an orthopaedic surgeon, an endocrinologist and a urologist before discharge from hospital.

### **How is it treated and managed?**

#### **Bowel dysfunction**

There are several things that can be done to prevent constipation and soiling. Children should be given a high fibre diet to help regular bowel function. Between 2 ½ and 4 years of age children should be encouraged to sit on the potty after every meal. If bowel control is not achieved after several months, oral laxatives or regular rectal suppositories or enemas can be tried. If this is not successful continence may be achieved by using a surgical technique such as creating an opening in the abdominal wall to the colon to allow irrigation of the colon on a regular basis with an antegrade – or

forward flowing – colonic enema (ACE). Parents and children may require help with managing the stoma.

Biofeedback training may be used to achieve bowel control in a small number of older children with sacral meningomyeloceles. In this, a pressure sensor is placed in the rectum and connected to a visible pressure monitor.

### Chiari II malformation

If the child develops symptoms from the Chiari II malformation, treating any associated hydrocephalus may resolve the problem. If it does not, then a surgical operation is performed. This is usually an operation to decompress the cerebellum within the skull and restore the flow of CSF but may be an operation to untether the spinal cord at the level of the spina bifida allowing the tension in the cord to be released and the cerebellum to disimpact. Surgery usually stops progression of the symptoms, but improvement is not guaranteed. Some symptoms are more likely to improve with surgery than others – pain and headaches are most likely to improve, weakness and sensory problems are least likely to improve.

### Cognitive impairment

Learning disability is managed by a combination of educational provision, psychological and drug treatments.

### Epilepsy

Seizures usually respond well to anti-epileptic medication.

### Hydrocephalus

Treatment involves surgical insertion of a shunt which drains CSF from the lateral ventricle of the brain to another place in the body, most commonly



the abdomen (ventriculoperitoneal shunt). For further details see [Hydrocephalus](#) guidance.

### Interdisciplinary management

Because children with meningomyelocele have complex physical and emotional needs, management should be carried out by a multidisciplinary team. This team includes paediatric doctors (general physicians, community physicians, neurologists, neurosurgeons, orthopaedic and general surgeons, urologists and endocrinologists etc), specialist nurses, occupational therapists, physiotherapists, orthotists and teachers.

### Latex allergy

Early exposure to latex should be avoided if possible. Surgery should take place in a latex free environment. Latex free catheters and gloves should be used. Toys that contain a significant amount of latex, such as rubber balls and balloons, and dressings that contain latex, should be avoided.

### Musculoskeletal abnormalities

Physiotherapy and orthotics are used to treat both primary (from birth) and secondary musculoskeletal problems.

Club foot is usually treated by serial casting during the first 3 to 4 months of life followed by surgical correction between 4 months and 1 year of age. Other ankle and foot deformities may require surgical correction to allow the child to wear shoes. Hip deformities may be surgically corrected in order to improve mobility. Children should receive regular calcium and vitamin D and be encouraged to undertake weight bearing activities whenever possible in order to reduce the risk of osteoporotic fractures.

Scoliosis (curvature of the spine) may require orthotic support or surgical intervention depending upon the severity of the deformity. Kyphosis (hump back) may also require surgical intervention.

Some children have tethering of the spinal cord which may lead to rapidly progressive scoliosis. Surgery to untether the cord may halt or reverse progression of the scoliosis. It is important to identify tethering of the cord before carrying out spinal fusion, because it is more difficult to carry out surgery on the spinal cord following spinal fusion. If scoliosis is secondary to a

Chiari malformation, treatment of the Chiari malformation may lead to an improvement in the scoliosis.

### Pressure sores and ulcers

General preventative measures include avoiding tight fitting shoes or braces, avoid crawling on rough surfaces and regular checking of skin surfaces for abrasions and sores.

Pressure sores can be avoided by children in wheelchairs by adapting the seating system, for example by using special foam filled cushions. Children should be taught to perform regular pushups and to regularly change their position.

Established pressure sores are treated by alleviating the pressure on the sore and applying appropriate dressings. Infected pressure sores may require treatment with antibiotics. Children with large pressure sores that fail to heal may require admission to hospital for surgical debridement (removal of dead tissue) of the wound, skin grafting and intravenous antibiotics when necessary.

### Psychosocial problems

Children require support and encouragement from adults. They should be encouraged to manage their own condition whenever possible in order to achieve maximum independence. Depression should be treated as per normal.

### Surgical closure

Surgical closure of the defect is undertaken shortly after birth to prevent infection of the spinal cord and protect the spinal cord and nerves from subsequent physical injury. In addition, if the child has hydrocephalus, a shunting procedure is often required shortly afterwards to prevent cerebrospinal fluid (CSF) accumulating and causing progressive hydrocephalus.

### Urinary dysfunction

If cystometry demonstrates raised pressure in the bladder, this needs to be reduced. Treatment is usually by way of clean intermittent catheterisation (CIC) in which the parents are taught to insert a clean catheter into the

bladder at least four times a day to drain urine. This may be started soon after birth. Sometimes this is not successful and an operation called a vesicostomy is performed, in which an opening in the abdominal wall into the bladder is created so that urine can drain directly into the nappy. This is usually a temporary procedure and when the child is older the vesicostomy is closed and the child begins a CIC programme.

Confirmed urinary tract infections require treatment with appropriate antibiotics. If these occur frequently, long term preventative (prophylactic) antibiotics should be given either by mouth or instilled directly into the bladder through the catheter.

At age 3 to 4 the child and parents are taught CIC in order to try and achieve continence. Certain medications can be given by mouth or instilled into the bladder to improve the storage capacity of the bladder and help with continence. About 70% of children treated with CIC and medication achieve continence during primary school years.

If CIC and medication are not successful in achieving continence, there are a number of surgical procedures that can help.

Alternatively “volitional voiding” can be tried in which an artificial sphincter is placed around the urethra and is attached to a bulb which is placed under the skin in the genital area. When the bulb is squeezed the sphincter relaxes and urine is able to drain. This is however, only suitable for a small number of highly motivated people because if the person forgets to “void”

regularly, the bladder pressure rises and this risks subsequent kidney damage.

Visual impairment

Squint often requires surgical correction.

Weight and stature abnormalities

Children should be encouraged to be exercise regularly and eat a healthy diet.

### Aids and Adaptations

#### Urinary Catheters

#### **What evidence is available?**

<b>Sources of evidence</b>	<b>Information Provided</b>	<b>Limitation of Evidence</b>
Parent / carer	First source of information and associated evidence.	Does not have specialist knowledge.
Paediatrician / specialist nurse	Symptoms, investigations and treatment / management, and likely to have information about resulting disability or needs.	None.
Occupational therapist / Physiotherapist	Symptoms, investigations and treatment / management, and likely to have information about resulting disability or needs.	None.
GP	Symptoms, investigations and treatment / management.	Does not have specialist knowledge. Unlikely to have information about resulting disability or needs.

#### **How long will the needs last?**

Age at date of claim	Award Period
1 - 7	Award to age 8 (or for 1 year, whichever is the longer)
8 - 15	Award to age 16 (or for 1 year, whichever is the longer)

Renewal at age 8 is suggested as the disability associated with Spina bifida may improve as a consequence of surgical, medical, physiotherapy, occupational therapy, psychosocial and educational interventions.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

### **Associated impairments and medical complications**

#### Bowel dysfunction

Children with meningomyelocele are likely to have bowel problems secondary to uncoordinated bowel contractions, ineffective closure of the anal sphincter and loss of rectal sensation. This results in constipation and “overflow incontinence” and soiling.

#### Cognitive impairment

Children with a meningomyelocele can have a normal IQ and some have mild learning disability. A few have severe learning disability and this is usually due to shunt infection (if they have hydrocephalus) or prenatal hydrocephalus.

Even if they have a normal IQ, they tend to have impairments in perceptual skills, organisational abilities, attention span, speed of motor response, memory and hand function and many have a specific learning disability

such as dyslexia (difficulty with reading), dyscalculia (difficulty with maths) and dysgraphia (difficulty with writing).

They usually have more difficulty with maths than reading. They also often have difficulty with planning, initiating and sequencing tasks. About one third of children with meningomyelocele have ADHD.

### Epilepsy

About 15% of children with meningomyelocele develop epilepsy. Seizures are usually generalised tonic clonic and respond well to anti – epileptic medication. A blocked or infected shunt in children with hydrocephalus may result in a new type of seizure developing or an increase in seizure frequency.

### Latex allergy

More than half of children with open defects develop an allergy to latex. It is more common in children who have had frequent surgical procedures and the risk increases as the child gets older. It may be life threatening. Rarely children with latex allergy have sensitivities to certain foods such as bananas, water chestnuts, tomatoes and kiwi fruits.

### Mobility impairment

Meningomyeloceles at higher levels are associated with greater muscular weakness and greater mobility impairment. All children will have significant

delay in walking. Most children will not be able to walk independently after adolescence.

<b>Level of defect</b>	<b>Mobility status</b>
Thoracic (T1 - 12 ) or upper lumbar (L1 - 2) level	Unlikely to be able to walk but some may do so with considerable bracing and support. Most use a wheelchair within a few years of birth.
L3 – L4 level	Able to walk but often require bracing up to the hip and crutches. As they approach adolescence, they are likely to rely increasingly on a wheelchair. In adult life almost all use wheelchairs.
Low lumbar (L5) or sacral level	Usually learn to walk well by the age of 2 or 3 years with ankle bracing or no bracing at all. In adult life most able to walk independently but many elect to use a wheelchair if walking for considerable distances.

Other factors that affect mobility include cognitive function, complications including joint contractures and shunt infections, involvement of the parents and the treatment programme.

Children with high level defects are unlikely to walk, although their mobility is likely to be better if they have no cognitive impairment, undertake regular walking exercises and have parents who are committed to the exercises. Children with high level defects and significant cognitive impairment are unlikely to walk and should commence early wheelchair training.

#### Musculoskeletal abnormalities

Children with meningomyelocele have primary musculoskeletal problems that include muscle weakness, reduced joint mobility and leg movement and reduced sensation and awareness of the legs and severity of these depend upon the level of the lesion. As a consequence, they may have secondary problems which include joint stiffness, loss of muscle length, contractures

and deformities, that include spinal deformities. These problems may deteriorate with age.

Joint deformities include club foot (talipes equinovarus) and hip deformities, for example fixed flexion deformity or predisposition to dislocation of the hip.

Children with meningomyelocele are also prone to osteoporotic fractures because of loss of muscle strength and inactivity. Fractures may occur after orthopaedic surgery, especially after prolonged casting.

As people with spina bifida age, they may develop osteoarthritis of the hips and knees secondary to abnormal sensation and gait. Almost 90% of children with a meningomyelocele above the sacral level have a spinal deformity. These include scoliosis (curvature), kyphosis (hump) or kyphoscoliosis (combination of the two). They may be present before birth (congenital) or develop later in childhood (acquired). If untreated, spinal deformities may interfere with sitting, walking and possibly breathing. Spinal deformities may be painful, and the pain may interfere with function.

#### Neurological deterioration

A deterioration in strength and bowel or bladder function may be due to one of following reasons -:

- Malfunctioning or blocked ventricular shunt
- Tethered spinal cord
- Rarely syringomyelia (development of cysts in the centre of the spinal cord)

The Chiari II malformation may also become symptomatic.

If identified early, all of these problems may require treatment.

#### Pressure sores and ulcers

Children with meningomyelocele are prone to develop pressure sores (decubitus ulcers) on weight bearing surfaces, for example feet and buttocks, because of insensitivity to pain. These become more frequent during



adolescence. Preventative measures are most important together with early treatment. Failure to treat early may require prolonged hospitalisation.

### Psychosocial problems

Physical difficulties can lower the child's self esteem and impair the establishment of peer relationships. Depression is likely to be more common in children with meningomyelocele than the general population.

### Sleep problems

Children with myelomeningocele frequently have disordered sleeping. This may be due to factors such as pain, spasticity etc. It may cause affected children to be tired during the day, affecting their concentration and their ability to function at school.

### Urinary dysfunction

The nerves that control the bladder and rectum leave the spinal cord in the lower sacrum. For this reason virtually all children with meningomyelocele, (including those with sacral lesions and normal leg movement) have bladder and bowel dysfunction. They tend to be incontinent of urine and are also unable to completely empty the bladder which may predispose to infections of the bladder and kidneys and long term kidney damage.

Children with meningomyelocele have a higher incidence of malformations of the kidneys, such as horseshoe kidney and absent kidney than the general population.

### Visual impairment

About 20% of children with meningomyelocele have a squint (strabismus). Squint can also result from a rise in intracranial pressure caused by a

malfunctioning shunt. Visual impairment can also be caused by an abnormality of the visual gaze centre in the brain resulting in squint or nystagmus.

Weight, stature and endocrine abnormalities

Children with meningomyelocele are at increased risk for obesity (about two thirds are significantly overweight). They are also at increased risk for short stature, growth hormone deficiency and other endocrine abnormalities.

### **Incidence and Prevalence**

The incidence and prevalence of neural tube defects (NTDs) varies from country to country.

Spina bifida occulta is present in about 10% of the population.

Meningomyelocele occurs in approximately 60 per 100,000 births, encephalocele in approximately 10 per 100,000 births and anencephaly in approximately 20 per 1000,000 births. (US figures).

The prevalence of NTDs has fallen as a result of several factors that include better nutrition, prenatal screening and routine prescription of folic acid in pregnancy. In addition, there has been an increase in survival as a result in improvements in medical care, resulting in more adolescents and adults with meningomyelocele in the community.

The cause of NTDs is multifactorial and includes genetic and environmental influences. People with NTDs have an increased risk of having a child with an NTD. Couples who have had one child with a NTD, have a much higher risk of having a subsequently affected child. Environmental factors associated with an increased risk of NTDs include maternal exposure to certain drugs, for example antiepileptic drugs, high maternal alcohol consumption, maternal diabetes, maternal obesity, increasing maternal age and low socio-economic status.

Children with spina bifida have an increased risk of having anorectal malformations and sacral and genitourinary abnormalities.

There is a strong link between folic acid deficiency and NTDs. It has been proven that giving daily supplements of folic acid to the mother around the

time of conception and for the first 12 weeks of pregnancy substantially reduces the risk of having a child with an NTD.

### **Aids and adaptations**

Because of the complex effects of neural tube defects there are many aids and adaptations that can be used. The most commonly provided are:

#### **Bracing**

Lower limb bracing is often required to assist the child to walk. The type of bracing is usually determined by the level of the defect. Higher defects

require more extensive bracing. Bracing must be monitored to prevent skin breakdown over bony prominences.

### **Wheelchairs**

As most children with meningomyelocele cannot walk independently after adolescence, wheelchair provision should usually be considered by early adolescence.

Early wheelchair provision should be considered for children with a high level defect and learning disability as walking may not be considered a realistic goal.

### **Home environment aids**

Various modifications may be considered including the use of rails, wheelchair adaptations, bathroom and kitchen modifications etc.

### **Incontinence aids**

These include catheters and stomas.

### **Urinary catheters**

There are different types of urinary catheters.

### **Indwelling catheters**

These can be either urethral or suprapubic. The care of both of these is similar. Parents of young children need to:

- Make sure that the catheter is taped to the abdominal wall and replace the tape if it becomes loose or dirty
- Check the tubing regularly to prevent it becoming kinked
- Keep the collection bag below waist level but off the floor. Shoulder bags can be supplied
- Ensure that children do not ride bikes or straddle other toys that might result in the catheter becoming tangled
- Ensure that children avoid rough and tumble play which can cause damage to the catheter or operation site (suprapubic catheters only)
- Encourage the child to drink plenty of fluids to ensure a good flow of urine and prevent blockage and infection
- Empty the collection bag at least four times a day
- Change the collection bag once a week or if the bag becomes disconnected

- Contact the GP or hospital if the urine becomes infected (urine is darker, smells, or is cloudy or child develops a temperature), stops draining, leaks or if the catheter falls out.

### **Clean Intermittent Catheterisation (CIC)**

This involves insertion of a catheter through the urethra into the bladder about 4 to 6 times a day. Children should not go more than 8 hours without performing a CIC at night. Insertion should be carried out in clean conditions – thorough handwashing etc. Catheters are reused so they need to be cleaned immediately after insertion and sterilised once a day. Parents

should contact the GP or hospital if they have difficulty inserting the catheter, or if there are signs of urinary infection.

The age at which a child can independently perform CIC varies. The average age is about 8 years but can be as young as 5.

## What you need to know about Visual Impairment

### What is normal vision?

Normal vision occurs when there is no abnormality of structure or function of.....

- [Visual Impairment](#)

### What are the effects and signs?

There are many conditions that can affect the eye in children.....

- [Effects of Visual Impairment](#)

### How is it assessed?

Vision is assessed by testing visual acuity for both distance and near vision and the visual fields .....

- [Assessment of Visual Impairment](#)

### How is it treated and managed?

Local authorities employ specialist teachers and Rehabilitation Officers called ROVICs .....

- [Treatment of Visual Impairment](#)

### What evidence is available?

The Consultant Ophthalmologist will provide details of symptoms, signs .....

- [Evidence](#)

### How long will the needs last?

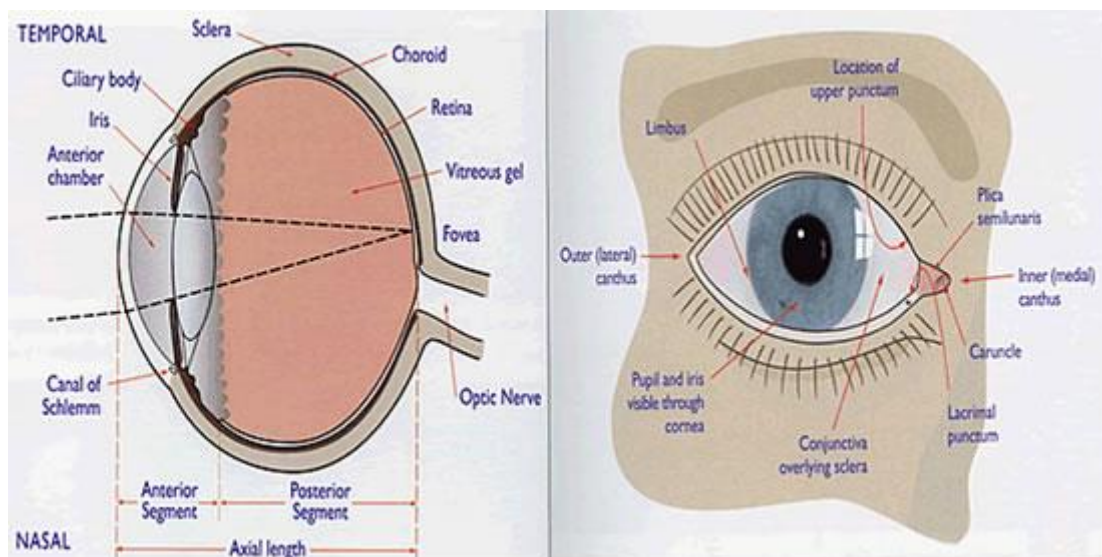
- [Prognosis and duration of the award](#)

## What is normal vision?

Normal vision occurs when there is no abnormality of structure or function of:

- the eye, the eye muscles or orbit
- the nerve pathways and blood supply to the eyes
- the structures in the orbit
- the optic nerves; and
- the subsequent pathways in the brain to the visual cortex in the back part of the brain and associated visual areas in the brain

## How do we see?



Light enters the eye through the cornea, at the front of the eye. This is a transparent dome, which has both protective and focussing properties. Light is refracted at the junction of air and the tear film on the surface of the cornea. The cornea is kept clear by the properties of the tear fluid and replenishing the tear film by the blink action of the eyelids.

It then travels through the pupil (the aperture in the centre of the iris, the coloured part of the eye), which varies in diameter, depending on the amount of light entering the eye. If the environment is bright, the pupil constricts and if it is dark, the pupil dilates. The lens sits behind the iris and it

again, refracts light, to focus it on the retina. The lens, controlled by a muscle, can change its shape to ensure the image of both near and distant objects are focused on the retina. The lens will become thicker to focus on near objects and thinner for looking into the distance.

The retina is composed of millions of light-sensitive cells (photo receptors) called rods and cones. Cones are vital for the clarity and sharpness of vision, as well as being sensitive to colour and are concentrated around the macular area where vision is at its sharpest. The rods are responsible for peripheral vision and night vision but are not colour sensitive. They are more numerous than cones and much more sensitive to light and form the large majority of the photoreceptors in the remaining retina, being grouped mainly in the periphery. They do not contribute to visual clarity as cones do.

Each eye records an image on the retina, converting the light energy from the image to nerve impulses through a series of chemical reactions in the cones and rods. As the two eyes are spaced apart on the face, the images are slightly dissimilar with the right eye recording more of the right and the left eye more of the left side of the image. The nerve impulses of each image travel through the optic nerve from the retina of each eye. Approximately half of the optic nerve fibres (the nasal or inner half) from each optic nerve cross over at the optic chiasm. The subsequent visual pathway travelling through the length of the brain has fibres from both eyes (outer half from the same eye and inner half from the other eye). The nerve impulses are relayed to the visual cortex also called the primary visual cortex in the occipital area (the back) of the brain. Here the two visual pathways relay the image from each eye and compare the slightly dissimilar images from each eye. After an initial analysis of the image, nerve impulses are sent to the secondary visual areas (associated visual areas) for a detailed analysis of



the image for all visual functions such as contrast, colour, motion, depth and shape, resulting in a final percept and recognition of the image.

### **Incidence and Prevalence**

In 2017, the number of children and young people in England registered as having a visual impairment was:

<b>Age</b>	<b>Severe sight impairment</b>	<b>Sight impairment</b>
0 – 4	615	555
5 – 17	4,495	5,390

Figures for the other nations of the UK are collected separately. The figures for England are the most recent.

### [Development of the eye](#)

### [Structure of the eye](#)

### **What are the effects and signs?**

There are many conditions that can affect the eye in children, and it is not possible to cover all of these in this guidance. The links below contain an explanation of the most common conditions within that category:

- [Amblyopia](#)
- [Developmental eye disorders](#)
- [Diseases of the anterior chamber](#)
- [Diseases of the conjunctiva & cornea](#)
- [Disorders of the lens](#)
- [Disorders of the vitreous & retina](#)
- [Eye movement disorders](#)
- [Eyelid disorders](#)
- [Inflammatory eye disorders](#)
- [Inherited retinal dystrophies](#)
- [Intraocular tumours](#)
- [Refractive errors](#)

- [Retinal detachment](#)
- [Visual impairment due to brain damage](#)
- [Visual pathway disorders](#)

If you encounter a condition that is not included in this guidance, you should seek advice from Medical Services.

- [Severity indicators](#)

### **How is it assessed?**

#### **Screening**

All new born babies are screened for congenital eye abnormalities by medical staff prior to discharge from hospital. The red reflex test (to identify any luminous red appearance seen on the retina) is mandatory.

Premature infants at risk of retinopathy of prematurity (a condition causing marked increase of the retinal blood vessels) are regularly screened for this by an ophthalmologist.

Routine checks of visual behaviour are carried out in the community at about 8 weeks of age.

Formal testing of visual acuity usually takes place at primary school entry. Some areas also offer this at nursery age and additional checks for junior school age children but this is not universal.

All children with Juvenile Immune Arthritis (Juvenile Idiopathic Arthritis (JIA)) should be screened for uveitis according to guidelines.

#### **Assessment of vision**

Best corrected vision is the vision in the better of the two eyes assessed using spectacles, contact lenses etc.

Vision is assessed by testing visual acuity for both distance and near vision and the visual fields. Binocular vision is vision using both eyes and

monocular vision is vision using one eye. The method of assessment depends upon the age of the child.

## **Visual acuity**

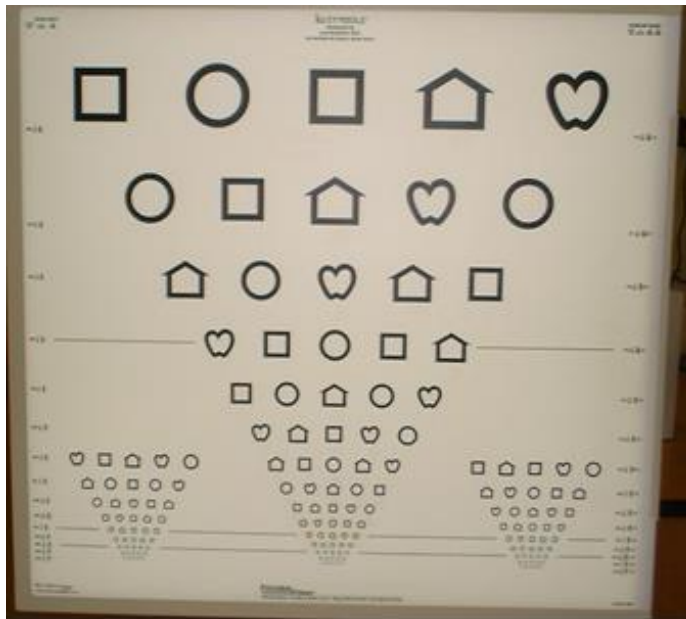
### **Over three years of age**

#### Lea Symbol Chart

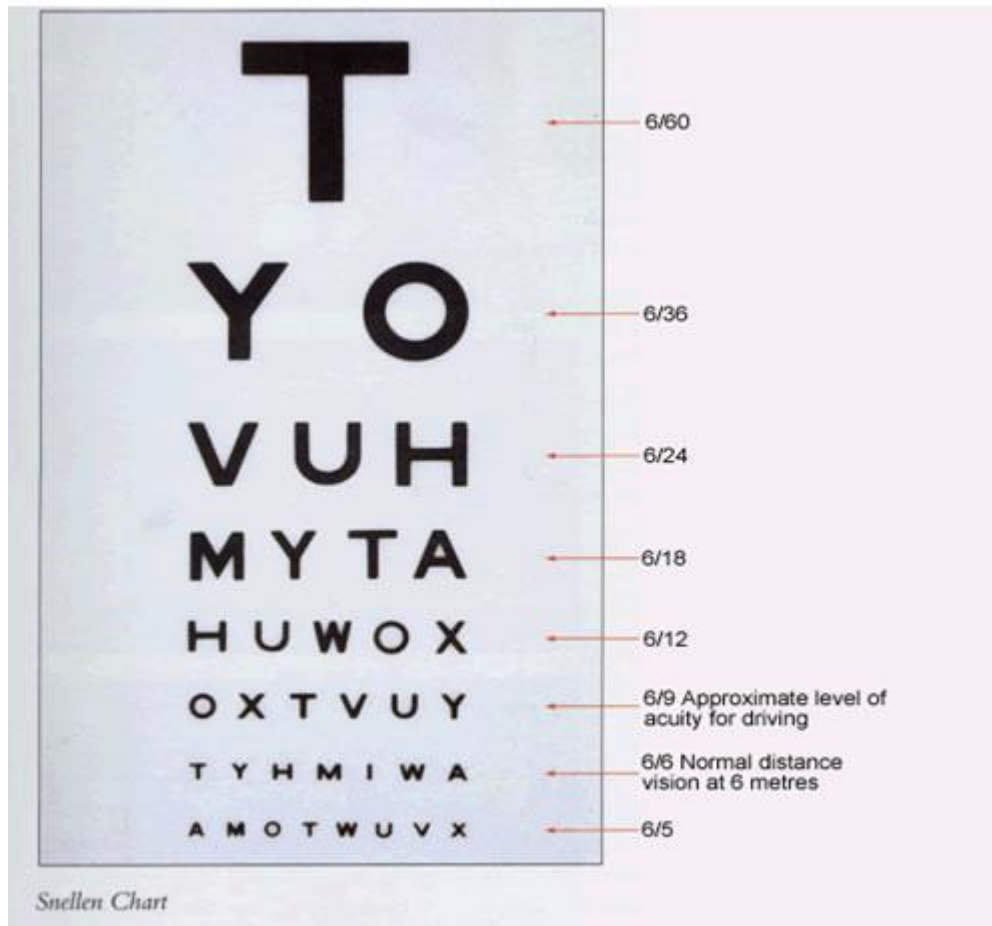
In children over the age of three years, distance visual acuity is usually checked using Lea Symbols. The test is back illuminated and carried out at 3 metres.

Four symbols are repeated and progression is in a logarithmic manner. The child is asked to identify or match the symbols. The central symbol of each line is identified till an incorrect answer is obtained. The child then must

identify four of the five symbols to accept the level of visual acuity. Numerical values denoting LogMar and Snellen are printed on the chart.



Snellen Chart



Snellen's chart with equivalent LogMar measurements

LogMar is a scale that expresses visual acuity as a decimal.

<b>Snellen 6 metres</b>	<b>Snellen 3 metres</b>	<b>LogMar</b>
6/60	3/30	1.0
6/48	3/24	0.9
6/38	3/19	0.8
6/30	3/15	0.7
6/24	3/12	0.6
6/19	3/9.5	0.5
6/15	3/7.5	0.4
6/12	3/6	0.3
6/9.5	3/4.8	0.2
6/7.5	3/3.8	0.1
6/6	3/3	0.0

The Snellen Visual Acuity Chart is designed to be read at a distance of 6 metres and is for older children. Its use is declining and being replaced by a LogMar letter chart.

Each row on the chart has a number, 60 on the top and 5 on the bottom. A child who can only read the top row has a visual acuity of 6/60 which means

that they can read at 6 metres what a normal sighted child could read at 60 metres. A child with normal vision would have a visual acuity of 6/6.

If the child cannot see the numbers on the chart, they are moved to 3 or 4 metres from the chart and tested. If this is not possible, counting fingers and hand movements (at 30cm) or light perception are recorded.

## **2 - 3 years of age**

In children over the age of two years who cannot read letters, vision can be formally tested using the Crowded Kay Picture Chart. The instructions are similar to the Lea Symbol Chart.



## **Under two years of age**

The vision of infants and preverbal children is assessed by:

- Asking the parents about visual behaviour,
- Observing whether the child can fixate and follow a small target (they should be able to do this from about two to three weeks after birth,
- Forced preferential looking (FPL) in which the child should look preferentially at a pattern stimulus rather than a plain one. A series of cards with a pattern (black and white stripes on one side of a plain card, i.e. gratings) are displayed sequentially through a puppet screen. The finest pattern that elicits preferential looking towards the stripes is taken as the vision. Norms for grating acuity have been established and at present is the

most reliable test within this age group. A difference between the two eyes is more reliable than actual grating acuity levels.

- 



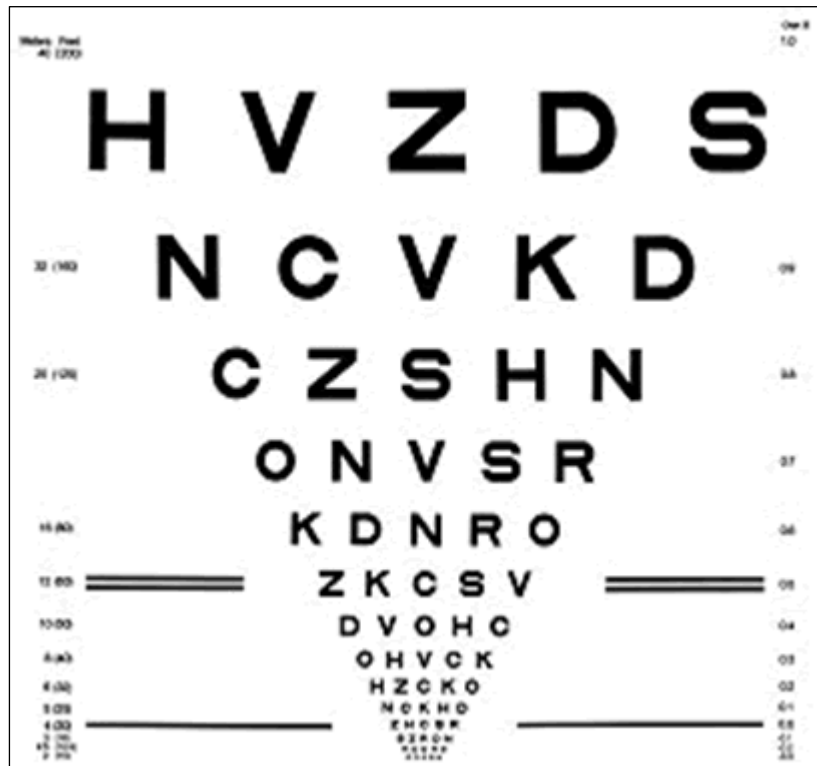
None of these can be precisely correlated with Snellen acuity.

### LogMar

LogMar is a scale that expresses visual acuity as a decimal. Visual Acuity Charts with LogMar scale are now commonly used in clinical practice. However, Snellen notation is still used for communicating visual acuity levels.



These are going to replace Snellen chart. The values for conversion to a Snellen notation are on the chart.



### Near visual acuity

This can be tested using the reduced Snellen tests read at 0.3 metres or the Maclure type test. Reliable picture or symbol tests for near vision are not



- Perimetry where a machine or computer is used. This provides an accurate assessment of visual field loss but can only be carried out accurately in children aged 7 or older. Perimetry may be kinetic where points of light are moved in until the child sees them or static where points of light are flashed onto a white screen and the child is asked to press a button in response. Assessment of the binocular visual field can be determined by either combining the results obtained from testing the visual field of each eye separately or by using a binocular test, for example, the Esterman binocular visual field test
- Detailed visual field assessment in routine clinical practice is only possible in children over 7 years of age

It would be reasonable to assume that central 10 degrees or less of a residual visual field would be considered as severe visual impairment. If the visual field is more than 10 degrees then it would depend on the pattern of the visual field defect. Inferior visual field defects, temporal visual field defects and involvement of the central visual field are all serious visual field defects.

### **Refraction testing**

The refraction test measures the extent of any refractive error present and allows for appropriate spectacles or contact lenses to be prescribed. In older children, subjective refraction may be performed where the child reads a chart with lenses of varying power in front of the eyes. In infants and younger children who cannot cooperate, cycloplegic refraction is carried out, when the pupils are dilated and the retina is viewed by an ophthalmoscope using lenses of varying power.

### **Electrophysiological investigations**

There are a number of investigations that can be carried out. Some of them may be used to confirm the diagnosis, but the Visual Evoked Potential (VEP) test can provide supplementary information about the likely degree of visual function.

In the VEP, electrodes are attached at the back of the head over the area of the visual cortex. The response to stimulation of the eyes by a flash of light

(in infants) or a patterned chart (in older children) is analysed by computer. This test provides an estimate of Snellen acuity.

The Electroretinogram (ERG) is obtained by electrodes on the cheek or lower eyelid or placed as a contact lens on the cornea. This tests the retina for retinal problems such as inherited retinal dystrophies.

There are other electrophysiological investigations, which are beyond the scope of this guidance.

Click on the links for details of:

[Deeming Provisions](#)

### **How is it treated and managed?**

Treatment varies depending upon the condition present and is specifically covered in the links listed on the [‘What are the effects & signs’](#) page of this guidance.

### **Services for visually impaired children**

If a child has severe visual impairment, the local authority is informed. Local authorities employ specialist teachers and Rehabilitation Officers called ROVICs (Rehabilitation Officers Visually Impaired Children) who provide

support to visually impaired children and their parents. Some Rehabilitation Officers work for voluntary organisations for the visually impaired.

Rehabilitation officers can provide support and training in:

- Crossing of roads, independent travel
- Cooking and other activities of daily living
- Use of magnifiers and low vision aids
- Modifications to the home to make them more accessible
- Advice to parents about support groups, societies, clubs, benefits, travel concessions, car badge schemes etc
- Counselling or referral to counselling services

Examples of voluntary organisations who work with visually impaired children include -:

- [Royal National Institute of Blind People \(RNIB\)](#)
- [National Blind Children's Society \(NBCS\)](#)
- [LOOK](#)
- [Sense](#) – the National Deaf/blind and Rubella Association

Many visually impaired children can be successfully integrated into mainstream schools, while special schools may be more helpful for children with additional learning difficulties.

[Aids & adaptations](#)

**What evidence is available?**

<b>Sources of evidence</b>	<b>Information Provided</b>	<b>Limitation of Evidence</b>
Claimant/carer	First source of information	May not be objective, does not have specialist knowledge
ROVIC (Rehabilitation Officers Visually Impaired Children)	Symptoms, signs, investigations including assessment of vision, treatment/management, and likely to have information about resulting disability or needs	None
Orthoptist	Assessment of vision (visual acuity and fields)	May not have information about symptoms, signs, investigations other than assessment of vision, treatment/management, and unlikely to have information about resulting disability or needs
GP	Symptoms, investigations and treatment/management	Unlikely to have information about resulting disability or needs

Optometrist	Symptoms, signs, investigations including assessment of vision, treatment/management, and likely to have information about resulting disability, needs and provision of low vision aids.  Does not have specialist knowledge	None
If considering entitlement to H/R Mobility component under the Severely Visually Impaired (SVI) provisions, the following evidence source must be used:		
Consultant Ophthalmologist	Symptoms, signs, investigations including assessment of vision, treatment/management, and likely to have information about resulting disability or needs	None

Note: If the Consultant Ophthalmologist doesn't have up to date information, consider arranging for an eyecare examination.

- [List of NHS hospitals with ophthalmology departments](#)

### How long will the needs last?

This guidance covers:-

Visual diseases
Deaf/blind deeming provision
Severely Visually Impaired (SVI) deeming provision

The need for attention is greatest in the early years but may diminish as the child matures. By the early teens the child with visual impairment may be a

competent touch reader and may have learned to cope with traffic and how to move around in familiar surroundings without danger.

<b>Age at date of claim</b>	<b>Award Period</b>
0 – 12	Care & L/R Mobility components – award to age 12 (or for 1 year whichever is greater)
13 - 15	Care & L/R Mobility components – award to age 16 (or for 1 year whichever is greater)

Deaf/Blind deeming provision

Also consult the Hearing Impairment guidance for award duration advice.

Severely Visually Impaired (SVI) deeming provision



<b>Entitlement to H/R Mobility under the SVI provisions is appropriate in the following circumstances -:</b>	
The child has been certified as severely visually impaired by a consultant Ophthalmologist AND their -:	
Visual acuity is less than (<) 3/60 – award to age 16 is appropriate	
; or	
Visual acuity is 3/60 or more but less than (<) 6/60 with a complete loss of peripheral visual field and a central visual field of no more than 10 degrees in total – award to age 16 is appropriate	
Note: Visual field assessment in routine clinical practice is only possible in children over 7 years of age. Therefore, it should be taken into account whether the consultant has indicated that the child requires a re-test when they are older to provide a more objective measurement of visual field.	
Age at date of claim 0 – 8	Care & Mobility components – award to the age at which a re-test is suggested by the consultant or to age 16 (if a re-test date is not known).
Note: Children under 8 who satisfy the H/R Mobility SVI provisions should be awarded to the age at which a re-test is suggested by the consultant or to age 16 (if a re-test date is not known).	
Age at date of claim 9 - 15	Care & Mobility components – award to age 16 is appropriate

**Note:** If visual field measurements are obtained from sources other than from the HFR, the need for & age of re-test may not be shown. In these circumstances, the duration of award needs to be discussed with Medical Services.

All information must be taken into account when considering the duration of care and mobility needs. The duration of care and mobility needs must be based on the particular circumstances of the child.

**Development of the eye**

In the human embryo, the eye starts developing at about 3 weeks of pregnancy. The eye is formed from multiple embryonic tissues interacting with each other in close connection - neural (relating to nervous system) tissue for the retina, which also gives rise to parts of the brain, ectodermal

(relating to the outermost cell layer) tissue for the lens and the eyelids which also gives rise to skin; mesodermal (relating to the middle cell layer) tissue for the iris, uvea (middle part of the eye), eye vasculature (the arrangement of blood vessels) and muscles of the eye. Genes such as PAX6 play an important part in controlling development. Neural cells form two spherical balls at the side of the head. These indent to form the optic cups. Specialised cell layers in the optic cups (neural and mesoderm) develop into the retina, choroid and uvea of the eye. The surface ectoderm is pinched off and incorporated in the optic cup to form the lens. At about 7 weeks of development, each optic cup fuses below from the centre going forwards and backwards to form ball shaped structures. The developing eye blood supply (the hyaloid vasculature) disappears just before birth to be replaced by a mature vascular system derived from blood vessels of the head and neck. As growth continues, the eyes move to the centre of the face. The eyelids fuse at the beginning of the second trimester (4<sup>th</sup> month) and reopen at the beginning of the 6<sup>th</sup> month (third trimester). Since the eye is formed from multiple tissues, disorders of the eye and brain (optic nerve hypoplasia and absence of optic chiasm), eye and skin (syndermatotic cataracts) are not uncommon.

Further development of the eye takes place during pregnancy with connections established between the eye and the primary visual cortex (the visual area) in the occipital lobe (one of the five lobes) of the brain. Further refinement in visual development takes place when the primary visual cortex forms connections with associated visual areas throughout the brain for detailed analysis of images. Abnormalities of focus (refractive error), alignment (squint) and obstruction to vision (cataract) will prevent normal visual maturation and cause poor vision (amblyopia).

During pregnancy eye growth can be disrupted due to various factors such as excessive intake of alcohol, drugs (foetal alcohol syndrome), infections (e.g. rubella causing cataract; toxocara (a genus of nematode parasites) or toxoplasma (a genus of intracellular parasites) causing inflammation and retinal scars). The newborn eye may show congenital abnormalities because of arrested embryonic development e.g. anophthalmos (absence of an eye), microphthalmos (an unusually small eye), coloboma (a cleft-like defect) & aniridia (almost complete absence of the iris), abnormal development such as congenital cataract and genetic dysfunction such as retinoblastoma (malignant tumour of the retina). In prematurely born infants, there may be persistence of embryonic tissues, which are programmed to disappear before birth such as nasolacrimal duct obstruction (causing tears to run down the affected side), persistent pupillary membrane (a middle cell layer attached to the rim or front of the iris), persistence of hyaloid vasculature (the developing eye blood supply) and primary hyperplastic posterior vitreous –PHPV (where the vitreous is replaced by a thick mass behind the lens). Finally, brain damage due factors such as lack of oxygen at birth can

lead to poor vision i.e. cerebral visual impairment (CVIU), the commonest cause of visual impairment in the developed countries.

### Visual development in children

All infants are born with an immature visual system. At birth, there is an established structural template from the eye to the brain with basic visual functions. Normal visual experience from very early life is essential for further visual development. Visual development is rapid in the first few months to approximately 18 months of life. There is further development at a slower pace for the next 5-7 years of life. Though each visual function (e.g. contrast, motion, shape recognition, colour & depth perception) has a separate developmental pattern, they are interlinked, with the development of one facilitating the development of the other. Visual development can be disrupted by abnormal visual experience. The severity of visual impairment is dependent on the age at which it occurs and the nature of the disease process. Early detection and appropriate treatment during the critical period of visual development often leads to a partial or complete recovery of disrupted visual development.

### Development of visual skills

Age	Visual skill
Within a few days of birth	Eyes have good optical clarity and can fixate briefly on a face. Red reflex (a luminous red appearance seen upon the retina) examination is normal.
3 months	Steady fixation and tracking of near object.
3 – 4 years	Many children can have vision formally tested by identifying pictures at a distance (e.g. crowded Kay Picture Test; Lea Symbol test; Glasgow Acuity Cards).
5 years and older	Standard Alphabet Test (LogMar Acuity Test).

### Structure of the eye

The eyeball itself is protected within the orbit (bony eye socket) by the eyelids, which blink automatically to spread a tear film over the surface of the eye and by the eyelashes, which sweep dust and particles away. It is also protected from trauma by the orbital bones.

The surface of the eyeball, that is, the clear cornea and the sclera (the tough, white outer coating of the eyeball) is protected in various ways. The cornea is very sensitive to touch and pain and the eye will reflexly blink if it is approached or touched. The sclera is protected by the conjunctiva, a thin membrane covering the sclera up to the corneal edge and is reflected back

onto the entire inner edge of the eyelids. Between the conjunctiva and sclera, the Tenon's Capsule (the connective tissue enveloping the posterior eyeball) prevents superficial infections (e.g. conjunctivitis) from spreading to the deeper structures.

Tears, which keep the eye moist, wash away particles and have antibacterial properties. The salty tear fluid is produced by the lacrimal glands, mucus-producing cells in the conjunctiva and small glands in the eyelids, which produce an oily substance that also add to the protective and lubricating nature of tears.

The eye itself is divided into:

- The anterior chamber - extending from the inside of the cornea, to the front surface of the iris. This is filled with a fluid called aqueous humour, as is the small space between the iris and the lens, known as the posterior chamber.
- The posterior segment - extending from the back of the lens to the retina, and is filled with vitreous humour, which is thick and jelly-like and keeps the eyeball firm.

### **Amblyopia (lazy eye)**

Up until about the age of eight, the visual system is immature and susceptible to amblyopia, in which a "healthy" eye does not see well because it is "turned off" by the brain. Under normal circumstances the brain receives input from both eyes and information from both is precisely integrated resulting in fine depth perception and correct alignment of the eyes. If poor information is sent to the brain from one eye because it is out of focus, usually because of a squint, refractive error or other problem (e.g. cataract), the brain chooses to use the other eye in preference. The other eye is ignored although it is anatomically normal and has the potential to see well. Early detection of amblyopia is essential to institute appropriate treatment and reverse the effects of amblyopia. As a principle the more severe the visual deprivation (e.g. cataract) the earlier it needs to be treated.

Mild to moderate amblyopia affects about 2% of the population. The risk of amblyopia is greatest in early childhood and reduces as the child gets older. With early recognition and treatment of the underlying cause visual loss can be minimised or even completely restored. The extent of the visual impairment from amblyopia ranges from mild to profound and depends upon the underlying cause, the age of onset and delay in identification and treatment.

Early treatment of amblyopia involves correcting the underlying cause, such as correcting a refractive error with spectacles, correcting a squint or surgically removing a cataract. The other key aspect of treatment is to force the brain to use the amblyopic eye by one of three techniques which are patching, pharmacological penalisation and optical penalisation. For a child with severe amblyopia the first line of treatment is patching, which involves

covering the good eye every day with an occlusive bandage patch until the vision in the amblyopic eye is optimised. Infants can only be patched for some hours whereas older children may be patched during all of their waking hours. All children must be followed up by an ophthalmologist, optometrist and orthoptist to determine the treatment required, treatment duration and to avoid over treatment of the good eye. Children with moderate amblyopia or those who refuse to use a patch can be treated by pharmacological penalisation with atropine eye drops which blur the good eye. Optical penalisation involves blurring the vision in the good eye by using a spectacle lens.

Prognosis: If detected early and treatment instituted with good compliance, the outlook is good.

### **Developmental eye disorders**

Deviations from normal development can lead to a wide variety of eye defects that range from anophthalmia (absent eye) to irregularly shaped pupils.

Developmental disorders of the eye can occur in isolation or as part of a syndrome.

Failure of fusion of the optic cup, usually inferiorly, may result in a coloboma, a cleft like defect in the eye. The effect of a coloboma varies depending upon the location and extent of the defect. For example, a coloboma in the pupil may only result in a cosmetic defect whereas a coloboma of the optic nerve and retina may cause severe visual impairment. A coloboma can be an isolated defect or may be part of a syndrome. Between 15 - 30% of children with small eyes (microphthalmia) and coloboma have the CHARGE association (coloboma of the eye, congenital heart disease, choanal atresia (a congenital blockage of the passageway between the nose and pharynx), retarded growth and development, genital abnormalities and ear malformations with or without hearing impairment).

### **Albinism**

This is a group of inherited disorders caused by a deficiency of the pigment melanin. It is characterised by -:

- A lack of pigmentation of the skin and hair
- Abnormalities of the eye due to absence of melanin which is normally found in the iris and retina

The eye abnormalities include nystagmus (jerking eye movements), reduced visual acuity, iris translucency, lack of pigmentation of the retina, underdevelopment of the fovea (central retina) and abnormalities of the visual pathways to the brain. Squint and refractive errors, especially astigmatism (irregular curving of the cornea), are common. Examination of parents, siblings and other family members, if possible, is essential to determine the inheritance pattern. Infants with albinism are slow to see and may appear to be blind immediately after birth, with irregular (roving) eye movements that

change to nystagmus in the first year. Visual acuity for distance vision is usually reduced to between 6/24 to 6/60. Near vision tends to be better and many young children can read small print unaided. Children may adopt a compensatory head posture in the form of a face turn and head tilt to dampen their nystagmus to obtain better vision. Parents may be reassured that vision usually gets better with time as there is delayed visual development and consequently nystagmus may also decrease with time.

Management involves correction of refractive errors, tinted lenses to reduce light sensitivity and low visual aids. Most children are able to be educated in a normal school environment. However, adequate support within the school should be available and tailored to the child. Surgery to improve the cosmetic appearance of squint or significant head posture may be considered. Referral of the family to a geneticist is recommended to help determine the possible risk to future siblings. It is also advisable to rule out other associations of albinism such as the rare bleeding disorders.

### **Congenital infections**

A number of maternal infections can affect the eye, in addition to the other clinical features seen in these infants. Infections that may affect the eye include rubella, toxoplasmosis, cytomegalovirus, chickenpox, herpes virus, syphilis and HIV.

These infections may affect many different parts of the eye. The degree of visual impairment depends upon the extent to which the various parts of the eye are affected and varies from child to child.

### **Specific conditions**

These include:

Abnormalities of the whole eye, that includes -:

- Anophthalmos when there is absence of the whole eye or the eye is replaced by a tiny cystic remnant. Treatment is by insertion of serial orbital prostheses to encourage orbital growth and later an artificial eye.
- Microphthalmos when the eye is smaller than usual. It may be associated with other abnormalities of the eye. The smaller the eye or the more severe the associated abnormalities of the eye, the worse the resulting visual impairment.

Abnormalities of the anterior segment, that includes:

- Abnormalities of the cornea, for example, megalocornea, when the diameter of the cornea is greater than normal, microcornea

when it smaller than normal or cornea plana when the cornea is flattened.

- Abnormalities of the iris, for example aniridia in which there is almost complete absence of the iris. In some children aniridia may be associated with a kidney tumour (Wilms tumour) and may need regular ultrasound scanning of the abdomen.

Abnormalities of the posterior segment, that includes -:

- Abnormalities of the vitreous, for example persistent hyperplastic primary vitreous when the vitreous is replaced by a thick mass behind the lens.
- Abnormalities of the retina, for example colobomas (cleft-like defects) of the retina.

Abnormalities of the optic nerve, for example, optic nerve hypoplasia, an uncommon condition where there is underdevelopment of the optic nerve. Bilateral cases may result in severe visual impairment and nystagmus (jerky eye movements) in infancy. Unilateral cases may be less severe and result in visual field defects and squint.

Prognosis: The above conditions are usually associated with moderate to severe visual impairment. Treatment, wherever possible, is essential to realise the child's visual potential. Long-term follow up is necessary.

## **Diseases of the anterior chamber**

### **Glaucoma**

If there is obstruction to the flow of aqueous humour the pressure in the anterior chamber will rise. This is called glaucoma, which can damage the optic nerve and subsequently affect visual fields and later vision if not effectively treated.

Although glaucoma in children is uncommon, it accounts for 2 - 15% of blindness in childhood.

Glaucoma may be primary in which there is a developmental abnormality of the drainage angle of the anterior chamber or secondary, due to a variety of other causes.

The commonest form of primary glaucoma is primary congenital glaucoma in which the drainage angle of the anterior chamber does not develop normally. It is usually bilateral but may be unilateral. Children usually present with classic symptoms of glaucoma, which include watering, sensitivity to light, an obviously enlarged eye (buphthalmos), one eye larger than the other and corneal swelling and clouding but in milder cases people may

comment on beautiful and large eyes of the child. All children will require surgery to open up the drainage angle. This is an ophthalmic emergency.

The other primary glaucoma is juvenile onset open angle glaucoma in which children present after the age of 3. These children tend to respond well to surgery to drain the anterior chamber.

There are numerous causes of secondary glaucoma that include developmental disorders of the eye, trauma, eye surgery, Neurofibromatosis (a genetic disease causing multiple soft tumours under the skin), Sturge Weber Syndrome (a congenital disease marked by a port-wine-coloured stain over the trigeminal nerve of the face), metabolic disorders, infections, inflammatory conditions, ocular tumours, chromosomal disorders and connective tissue disorders. Secondary glaucoma may also occur in children with congenital cataracts.

Glaucoma is diagnosed using a tonometer which tests the pressure in the eye. Tonometry may be performed whilst awake in babies but usually has to be carried out under anaesthetic in children up to about the age of 5 years of age.

The primary treatment of childhood glaucomas is surgery. However, medical treatment in the form of eye drops may be helpful to control intraocular pressure while waiting for surgery and may be required in the long term after surgery. Cyclo diode laser therapy and insertion of drainage devices are other methods of treatment.

Children need lifelong follow up because they can relapse for several decades after the initial treatment.

Amblyopia (a lazy eye) is a significant complication of childhood glaucoma and is important as a cause of poor vision in these children. Myopia or shortsightedness, squint and nystagmus (jerky eye movements) may be consequences of glaucoma.

Prognosis: The outlook is variable with the majority of children having moderate to severe visual impairment.

## **Diseases of the conjunctiva and cornea**

There are a large number of conditions that can affect the conjunctiva and cornea.

### **Corneal abnormalities**

These include:

- Corneal dystrophies in which there is opacification (clouding) of the cornea which may be associated with thickening, ulceration and cyst formation. There are a large number of corneal



dystrophies, which are usually inherited. Each type has different clinical features and prognosis

- Scarring due to forceps delivery during birth
- Keratoconus (conical distortion of the cornea) is usually bilateral but is unilateral (affecting one eye) in 10% of cases. It is more common in people with allergic conditions and is associated with other ocular and systemic disorders, for example congenital cataracts and Down syndrome. It results in astigmatism (blurred vision due to light not focused regularly onto the retina) and myopia (short sightedness). It usually presents between 10 and 20 years of age, but can be earlier. Progression of the disease is variable and asymmetrical (not even). It tends to worsen slowly for up to 10 years and then stop but there may be episodic deterioration over a longer period. Very rarely there may be sudden onset of visual deterioration caused by fluid accumulation in the cornea, which may result in corneal scarring. In the early stages, treatment of the refractive error may be achieved with spectacles but hard contact lenses are better at correcting the astigmatism (blurred vision due to light not focussed regularly onto the retina). Later on, surgery (corneal grafting) may be necessary with rejection being rare and the visual prognosis is usually good

**Prognosis:** Any corneal condition that leads to scarring will lead to visual impairment with central opacities causing severe visual impairment. Keratoconus has a good outlook with treatment by spectacles, contact lenses and if necessary a corneal transplant.

### **Infections**

There are a wide range of organisms that can cause infections of the conjunctiva and cornea. Some organisms, if inadequately treated can cause

inflammation and scarring of the cornea resulting in visual impairment. This is more common in developing countries. Such organisms include:

- Chlamydia trachomatis (a sexually transmitted micro-organism) producing trachoma (chronic inflammation of the mucous membranes of the eyes)
- Adenovirus infection (a type of virus that can cause conjunctivitis)
- Herpes simplex virus infection (an acute viral disease marked by groups of small blisters on the skin)
- Measles
- Gonorrhoea; and
- Certain bacteria, for example pseudomonas 9a type of bacteria)

Conjunctivitis beginning within the first month of life is called Ophthalmia Neonatorum and usually results from the eye being exposed to micro organisms in the birth canal during birth. The commonest cause in developed countries is chlamydial infection but other causes include herpes simplex and gonorrhoea. This is an ophthalmic emergency.

Treatment varies according to the infecting organism and the severity of the infection. Parents must also be seen and treated if necessary.

Prognosis: Most infections detected early and treated well have a good outlook. Severe infections, if undetected and untreated, can lead to corneal ulcer and impaired vision.

## **Disorders of the lens**

### **Cataract**

The major disorder affecting the lens is cataract, which is a defect in the clarity of the lens. Worldwide prevalence figures vary considerably. In the UK cataracts occur in about 1 in 250 infants, account for about 15% of blindness in children and about two thirds are bilateral. In most cases cataracts are present at birth and are identified by screening using a direct ophthalmoscope. Screening for the 'red reflex' (a luminous red appearance seen upon the retina during retinoscopy examination) is mandatory for all newborns and subsequently at checks in primary care.

Cataracts may be congenital (present at birth) or of childhood onset. Congenital cataracts may be unilateral or bilateral. Most unilateral congenital cataracts occur as an isolated abnormality in an otherwise normal child. Unilateral cataracts may be associated with a smaller eye and abnormalities of the vitreous. There are many possible causes of bilateral congenital

cataracts that include inherited (autosomal dominant), chromosomal abnormalities (the commonest is Down syndrome), intrauterine infections (Rubella, chickenpox, toxoplasmosis), metabolic disorders and genetically determined systemic syndromes, although in about 40% of cases no underlying cause can be found.

Childhood onset cataracts may not give rise to visual problems until later childhood. They may be either congenital cataracts that are mild in infancy but progress later or those that occur for the first time in later childhood, for example those associated with trauma, inflammatory conditions or resulting from treatments such as radiotherapy or oral steroid therapy. Childhood onset cataracts are usually easier to diagnose and treat than congenital cataracts and the prognosis for vision is better.

Treatment depends upon whether the cataract is bilateral or unilateral.

All infants with bilateral congenital cataract are referred to an ophthalmologist. The ophthalmologist initiates a referral to the paediatrician and geneticist, if necessary. Complete bilateral cataracts require early surgery within the first months of life. Some partial cataracts have a good visual prognosis and surgery can be postponed until later in childhood if vision deteriorates during the course of monitoring. If the degree of opacity (clouding) differs between the two eyes, occlusion therapy (patching of the better eye) may be required to prevent amblyopia (lazy eye).

Surgery involves removal of the cataractous lens. Visual rehabilitation for the loss of focussing mechanism of the lens (aphakia) usually involves correction by inserting a lens implant at the time of the operation. However, in children under the age of 2, correction is usually by use of contact lenses but may be by use of spectacles, but these are difficult to make and fit and may not be well tolerated in infants.

Amblyopia is a significant complication of cataract and is important as a cause of poor vision in these children. Vision can be significantly improved by occlusion of the better eye and correction of any refractive error.

If unilateral cataract is detected before eight weeks of age, good visual acuity can be achieved with prompt surgery. Amblyopia should be prevented by occlusion of the phakic eye (the eye which has not had the lens removed) and correction of any refractive error. Squint is common even with early treatment and glaucoma may occur as a late complication in up to 50% of children. If a dense unilateral cataract is identified after 4 months of age, amblyopia cannot be reversed, and surgery is not usually undertaken.

Prognosis: The outlook for congenital cataracts is improving though it is still common to have mild to moderate visual impairment. Unilateral cataract has a worse prognosis than bilateral cataract.

### **Lens subluxation (ectopia lentis)**

Lens subluxation (displacement of the lens within the pupillary space) is usually due to weakness of the suspensory ligaments that hold the lens in place. In most children it is bilateral and due to an underlying genetic

defect, for example Marfan syndrome (a connective tissue multisystemic disorder) or homocystinuria (a metabolic disorder) but may be secondary to trauma. Subluxation (incomplete or partial dislocation) may progress to dislocation of the lens into the anterior (front) or posterior (rear) chamber of the eye.

Lens subluxation may be asymptomatic (without symptoms) and only detected at screening examinations. Symptoms result from the lens becoming more spherical and unstable within the eye and include fluctuating myopia (short sightedness) and astigmatism. Squint and secondary amblyopia are common. An early sign is iridodonesis (tremulous iris) which may look like the eye is wobbling. Rarely the child may present with an acute painful red eye with reduced vision due to secondary glaucoma when the lens displaces and blocks the pupil.

All children with lens subluxation should be referred to an ophthalmologist and a paediatrician for investigation to exclude any underlying cause.

Most children with lens subluxation can be treated conservatively with correction of any refractive defect, treatment of amblyopia and regular monitoring of intraocular pressure for glaucoma.

Surgery may be indicated if it is not possible to maintain good visual acuity despite use of spectacles or contact lenses or if the lens dislocates regularly into the anterior chamber. Surgery involves removal of the lens and correction of the refractive error with spectacles or contact lenses.

Prognosis: The visual prognosis for children with lens subluxation is fair with most children having impaired vision (mild to severe) in one eye. Amblyopia related to high refractive errors is the major cause of visual impairment.

## **Disorders of the vitreous and retina**

### **Congenital abnormalities (present at birth)**

Minor abnormalities of the vitreous include vitreous cysts, which cause no visual impairment.

A more serious abnormality of the vitreous is persistent hyperplastic primary vitreous (PHPV), when the vitreous is replaced by a thick mass behind the lens). In this condition, which is unilateral, there is microphthalmia (an unusually small eye), an opacity (clouding) behind the lens and usually an associated cataract. Treatment varies according to the age of the child. If the abnormality is detected before 3 months of age surgery may be performed to remove the lens and vitreous with subsequent contact lens fitting and occlusion therapy (patching of the better eye) to prevent amblyopia (lazy eye), and in these cases good visual results may be obtained. If the abnormality is detected after 3-4 months surgery is not usually an option. With early surgery, prognosis can be improved but most children will have moderate to severe visual impairment in the affected eye. Occasional it is in both eyes.

Hamartoma, a benign tumour of the retina, usually presents in infancy or early childhood as either a squint or is identified at routine examination. The

vision in the affected eye is usually poor unless the macula is unaffected. They do not usually progress as the child ages.

Retinal colobomas (cleft like defects) vary in size and the degree of visual impairment usually depends on the extent of coexistent optic nerve involvement.

Retinal dysplasia is a condition where there is failure of retinal and vitreous development which results in bilateral retinal detachment which is present at birth. It is associated with a number of syndromes, the most common of which is Norrie's disease (causing learning disability and hearing loss). Affected children are also blind and have roving eye movements. Apart from surgery for any associated glaucoma, there is no specific treatment.

Angiomas (swelling of the blood vessels) of the retina may occur, usually in association with a syndrome called Von Hippel-Lindau disease (an hereditary disease causing vascular nodules in the retina). The angiomas vary in size and may present with blurred vision or be detected at routine screening. If treatment is required they usually respond well to laser therapy.

### **Acquired conditions**

#### **Retinopathy of prematurity**

Retinopathy of prematurity (ROP) is a condition caused by proliferation (marked increase) of the retinal blood vessels which occurs in very premature, low birth weight infants and is more common in infants who have received treatment with high concentrations of oxygen. In the majority, it is mild and undergoes spontaneous regression (reduction in symptoms) but in a small minority it may proceed to retinal detachment and blindness.

All infants at risk are screened by examination of the retina until about 36 weeks after birth.

Early stage disease (stage 1 and 2) usually regresses spontaneously but stage 3 disease requires treatment with laser to prevent progression. Stages 4 and 5 (subtotal and total retinal detachment) may be treated by surgical reattachment of the retina but the resulting vision is usually poor.

### **Eye movement disorders**

#### **Nystagmus**

Nystagmus is an involuntary spontaneous rhythmic oscillation (jerking) of one or both eyes. It is associated with poor visual acuity. The oscillations may be in the horizontal or vertical plane or may be rotary. Vertical nystagmus is usually a sign of significant brain disease and should be referred urgently. Onset may be at any age and is associated with a range of conditions varying from relatively benign to life threatening. Nystagmus that develops within 6 months of birth may be:

- Sensory defect, associated with a wide variety of sensory disorders and congenitally idiopathic (of no underlying cause). Visual acuity varies according to a number of factors, including

the level of sensory deficit, if present. There may be a gaze direction in which nystagmus is minimal called the “null zone” which may lead to the child developing an abnormal head posture. Treatment of the nystagmus involves correction of any refractive errors and prescription of appropriate low vision aids. The child, parents and teachers should be aware of the importance of the null zone. The child should be allowed to sit in the most appropriate part of the classroom to facilitate use of the null zone and allowed to use an abnormal head posture or adaptive head oscillations as required. Most children do not require surgery but it may be used to correct any associated squint or major abnormality of head posture

- Latent, which appears when one eye is covered, but may appear when both eyes are open if one eye has poor vision e.g. amblyopia (manifest latent nystagmus). It usually appears within the first year of life but may not be detected until much later. It is most commonly associated with early onset squint but may be associated with other conditions. True latent nystagmus is asymptomatic (without symptoms) and does not require treatment. Manifest latent nystagmus causes the child to hold the head in an abnormal position to ‘dampen’ the

nystagmus and therefore requires surgical treatment to reduce the nystagmus and abnormal head posture

- Neurological, secondary to an underlying neurological (affecting nerves) or neuromuscular (affecting nerves & muscles) disorder

## **Eyelid disorders**

### **Congenital disorders of the eyelids**

There are a number of congenital disorders (present at birth) of the eyelids that can cause keratitis (inflammation of the cornea) and scarring of the cornea due to abrasion of the cornea by the eyelashes. These include:

- Epiblepharon, in which there is a horizontal fold of skin that pushes up the lashes.
- Distichiasis, in which an additional row of lashes grow from the eyelid margins.
- Entropion, in which the eyelid margin turns inwards.
- Ectropion, in which the eyelid margin turns outwards.

If these conditions cause significant problems surgical correction may be performed.

### **Ptosis**

Ptosis (drooping of the eyelid) may be due to a number of causes but the most common is congenital ptosis, which is due to a weakness of the muscle that elevates the eyelid. It is usually unilateral but rarely may be bilateral, for example as part of the Ocular Fibrosis syndrome (where fibrous tissue replaces the usual, smooth tissue of the eye). Surgical treatment is the mainstay of treatment and is usually effective. It is important to check visual acuity prior to surgery in order to ensure that there is no amblyopia (lazy eye) that requires occlusion (eye patch) therapy.

### **Strabismus (squint)**

Squint is a condition in which the eyes are not aligned correctly due to an imbalance in the muscles that move the eyes. It is common, affects up to 5% of children and accounts for many of the children dealt with by

paediatric eye units. They usually appear in the first 3 years of life but can appear later.

Squints are classified according to the direction in which the eye turns -:

- Esotropia, in which the eye turns inwards
- Exotropia, in which the eye turns outwards
- Hypertropia, in which the eye turns upwards
- Hypotropia, in which the eye turns downwards

Esotropia and exotropia are more common than hypertropia and hypotropia.

Squints can either be constant (obvious at all times) or intermittent (apparent at certain times only).

In young children, a squint can result in amblyopia. Because the eyes are not aligned, the child may experience double vision. In order to prevent seeing double, the brain ignores the signals from the squinting eye (suppression) and only recognises signals from the normal eye. As the squinting eye is not being used the vision from that eye deteriorates and results in visual impairment. In older children, a squint may produce double vision but not amblyopia. This is because the brain has fully developed and it is not able to ignore signals from the squinting eye.

If it is identified that the vision in the squinting eye is poor, they may have to wear a patch over the normal eye (occlusion therapy) to encourage vision in the normal eye to develop.

### **Causes of squint**

Squints can either be:

- Congenital. Esotropia is more common than exotropia. Congenital Exotropia in the Caucasian population is uncommon and may signify significant pathology in the eye.
- Acquired:
  - Due to refractive error.
  - Other causes (less common) include trauma, viral infections such as measles, genetic conditions like Noonan syndrome (causing a webbed neck, ptosis and short stature) and



hydrocephalus (the abnormal expansion of ventricles within the brain).

### **Treatment of squint**

A squint should be treated as early as possible in order to prevent the development or progression of amblyopia in the affected eye.

Several types of treatment are available for a squint -:

- Glasses if the child is long sighted. This may correct the squint as well as the refractive error.
- Occlusion therapy to stop development or progression of amblyopia in that eye
- Botulinum toxin injection into the muscles of the affected eye to temporarily weaken the affected muscle allowing the eyes to realign themselves. This is usually done under general anaesthetic in children.
- Eye exercises, especially for an intermittent squint.
- Surgery. If non-surgical treatments are not effective, surgery is usually performed. Surgery should improve the alignment of the eyes and improve binocular vision. Surgery involves operating on the muscles of the eye to realign them. It is usually carried out as a day case under general anaesthetic. In the majority of cases surgery is highly effective. In some cases, however, binocular vision is not always fully restored. Further

surgery is sometimes required if the squint is large or if it recurs.

## **Inflammatory eye disorders**

### **Uveitis**

Uveitis is inflammation of the eye. It can be acute (sudden onset) or chronic (slow onset and persistent). There are four main types:

- Anterior uveitis, affecting the front of the eye (anterior chamber).
- Intermediate uveitis, affecting the middle of the eye (vitreous and peripheral retina).
- Posterior uveitis, affecting the back of the eye (choroid and retina).
- Panuveitis, affecting the whole eye

In children with Juvenile Immune Arthritis (Juvenile Idiopathic Arthritis (JIA) the uveitis may be silent (i.e. without symptoms or signs). A recommended national screening programme is in place for children with JIA.

Anterior uveitis and intermediate uveitis are the most common inflammatory disorders in children.

#### **Acute anterior uveitis**

This may be associated with a number of conditions including Ankylosing Spondylitis (an inflammatory condition affecting the spine), Reiter's disease (an inflammatory condition affecting the joints and urethra), Kawasaki disease (an inflammatory condition that may cause coronary artery aneurysms) and acute viral infections. It may be asymptomatic (without symptoms) or may present as a painful red eye. The mainstay of treatment is steroid eye drops and treatment of the underlying condition. The condition usually settles quickly and the prognosis for visual acuity is usually good.

#### **Intermediate uveitis**

This usually occurs in young adults but may occur in childhood. It may be associated with a number of conditions such as infections and the severity of the condition varies widely. It may be asymptomatic in the early stages, but the child usually presents with red, painful eyes, mild blurring of vision and sensitivity to light. Complications include cataract, vitreous haemorrhage and retinal detachment. Treatment usually involves use of steroid eye drops and other anti-inflammatory medication. The visual prognosis is usually fairly good.

#### **Chronic anterior uveitis**

This may be associated with a number of conditions including juvenile rheumatoid arthritis, infections and ulcerative colitis, but in the majority of children there is no obvious underlying cause. It may be unilateral or bilateral

and is frequently asymptomatic. However, it can cause secondary glaucoma or cataract formation. The mainstay of treatment is steroid eye drops and eye drops to dilate the pupil, monitoring and treatment of complications and management of the underlying condition but may involve the use of more potent anti-inflammatory drugs or systemic steroids. The prognosis for visual acuity depends upon the severity of the inflammation and duration of the uveitis and ranges from good to poor.

### **Posterior uveitis**

Posterior uveitis is usually caused by an underlying infection. The majority are due to toxoplasmosis and toxocariasis (parasitic infections transmitted by cats and dogs). There are a number of treatment options and visual prognosis varies.

### **Panuveitis**

Panuveitis may result from Behcet's disease (a multisystem disorder associated with oral and genital ulceration), endogenous endophthalmitis (infection spreads to the eye from somewhere else in the body via the bloodstream) and sympathetic ophthalmia (a bilateral panuveitis that follows a penetrating injury to one eye). Treatment varies according to the underlying cause and the visual prognosis varies widely.

A number of other conditions, for example retinoblastoma (the commonest intraocular tumour in children), leukaemia, retinal detachment and intraocular foreign body may be associated with intraocular inflammation.

Prognosis: The visual prognosis for these conditions also varies widely.

### **Inherited retinal dystrophies**

There are a large number of inherited retinal dystrophies, caused by damage to the light receptors (sensory nerve endings that respond to light) in the retina, which cause symptoms in childhood. Most are associated with a gradual deterioration in vision over time, but some are non progressive. They are all relatively uncommon. Some of the more common are described below.

#### **Non progressive dystrophies**

##### **Congenital stationary night blindness**

Congenital stationary night blindness (CSNB) – may be autosomal dominant, autosomal recessive or X linked. Children with the autosomal dominant type have poor night vision but vision is otherwise normal. Children

with the autosomal recessive and X linked type have moderately reduced visual acuity and mild colour vision disturbance.

### **Achromatopsia**

There are two main types. Children with the autosomal recessive type (rod monochromatism) in which there are no functioning cone receptors in the retina present in infancy with reduced vision, severe photophobia (intolerance to light) and nystagmus (jerking movements of the eyes). Visual acuity is usually about 6/60 and these children have no colour vision. Children with the less common X linked type (blue cone monochromatism) have blue cones in the retina. They have better visual acuity than children with the autosomal recessive type and have some colour vision.

### **Progressive dystrophies**

These are classified according to whether they affect the retina generally or the central retina.

#### **Generalised**

##### **Infantile rod cone dystrophy (Leber's amaurosis)**

Children with this condition present in infancy with severe visual impairment, nystagmus and roving eye movements. Most children are otherwise normal but some may have developmental delay and other neurological problems.

##### **Retinitis pigmentosa**

Retinitis pigmentosa may be autosomal dominant, autosomal recessive or X linked and is associated with characteristic pigmentation of the retina. The age of onset and prognosis is very variable. X-linked recessive disease tends to be of early onset and severe, autosomal dominant tends to be of later onset with a better visual prognosis and autosomal recessive is very variable. Retinitis pigmentosa is also associated with a variety of inherited genetic syndromes, for example Usher syndrome (associated deafness) and Refsum disease (associated unsteadiness, peripheral nerve damage and deafness). In the majority of these syndromes, treatment does not have any effect on the progression of the visual impairment. However, in Refsum disease, dietary modification may slow the deterioration in visual impairment.

Children with retinitis pigmentosa usually present with difficulties with night vision and peripheral field loss. There is usually subsequent progressive

visual field loss resulting in marked constriction of the visual field and there may be central field loss as well if the macula is affected.

Retinitis pigmentosa is associated with cataracts and oedema (swelling of the macula). Although there is no specific treatment for retinitis pigmentosa itself, vision may be improved by treating the cataracts or macular oedema.

### **Central retina**

#### **X linked juvenile retinoschisis**

This is the most common inherited retinal dystrophy affecting males and is due to splitting of the layers of the retina. It usually presents in childhood with reduced visual acuity. It is often discovered on routine testing, but may present with sudden visual loss secondary to vitreous haemorrhage (bleeding into the vitreous at the back of the eye). In the long term, vision is usually reasonably well preserved, but severe visual loss may occur if there is extensive involvement of the peripheral retina or if there is vitreous haemorrhage or retinal detachment.

#### **Stargardt disease**

Children with this condition have degeneration of the macula resulting in central visual field loss, although later in the disease they may have peripheral field loss as well. They usually develop symptoms during their school years but some may not present until early adult life. The prognosis is poor and most have a visual acuity of 6/60 or less by their early 20s.

#### **Vitelliform dystrophy (Best disease)**

This condition usually presents with mild visual loss in childhood or early adult life. The visual prognosis is generally good but there are some people who develop subretinal neovascularisation (new blood vessel formation in the retina) that may result in significant visual impairment.

#### **Progressive cone dystrophies**

There are many different types of varying severity. Children have early loss of colour vision, photophobia, reduced visual acuity, nystagmus in the early onset forms and most develop night blindness. Although there is variation in severity, overall the visual prognosis is poor.

### **Intraocular tumours**

These are rare in childhood. Benign tumours are more common and include hamartoma (a benign tumour of the retina) seen in conditions like Neurofibromatosis (a genetic disease causing multiple soft tumours under the skin). Malignant tumours include retinoblastoma and leukaemia involving the eye.

#### **Retinoblastoma**

Retinoblastoma is the commonest intraocular tumour in children but it is still very rare. About half of affected children develop the disease due to

inheritance of the retinoblastoma gene (RB1) and the tumours may be bilateral, but those who do not have the retinoblastoma gene have unilateral tumours. Most children present with a squint or an abnormal pupil reflex which is obvious to the parents. The red reflex test (a luminous red appearance seen upon the retina during retinoscopy examination) at birth and in subsequent early checks in primary care is mandatory for detection of retinoblastoma. Less commonly children present with a painful red eye or proptosis (protrusion of the eye). In most cases of unilateral retinoblastoma the tumour is advanced and enucleation (removal) of the eye is required. In children with bilateral retinoblastoma, one eye will usually have a large tumour that requires enucleation but the other usually has smaller tumours that may respond to other treatments, such as chemotherapy, radiotherapy, laser or cryotherapy, enucleation only being required if the tumour recurs afterwards.

### **Refractive errors**

Light entering the eye is focused (refracted) by the cornea and lens. Under normal circumstances the light rays are focused exactly on the retina resulting in a clear image. A refractive error occurs when the light is not focused accurately on the retina, resulting in a blurred image.

Accommodation is the process by which the eye changes optical power to maintain a clear image (focus) on an object as its distance changes. As an object moves closer, the ciliary (eyelid) muscles in the eye contract, the lens becomes fatter and its refracting power greater so that light rays are focused onto the retina. In addition, the eyes normally turn inwards (converge) when focusing on a near object. Accommodation and convergence are linked within the control centres in the brain and occur in relation to each other to maintain clarity of vision and appropriate focus.

### **Aphakia (absent lens)**

Aphakia is the condition where the lens has been removed because of a congenital (born with) or developmental cataract or the lens is lost through trauma or is absent. Children with this condition either need contact lenses, spectacles with a high power convex lens or the insertion of a replacement lens in the eye (intraocular lens).

### **Anisometropia**

In this condition one or both eyes have a significantly different error (e.g. high hypermetropia in one eye and a normal other eye). This may lead to asymmetric (unequal) development of vision with the visual cortex

'favouring' the normal eye or the eye with the least error and poor vision in the affected eye.

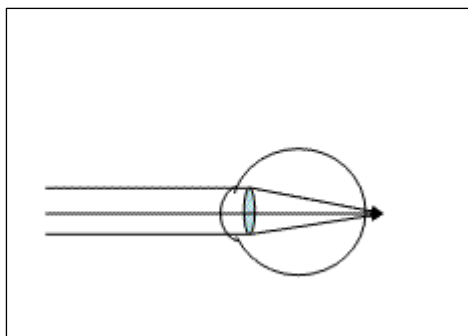
### **Astigmatism (blurred vision)**

Astigmatism occurs when the cornea is irregularly shaped so that light is not focused regularly onto the retina and vision is blurred at all distances. Cylindrical lenses are prescribed for constant use.

The child may have a combination of astigmatism with hypermetropia/myopia.

### **Hypermetropia (far sightedness)**

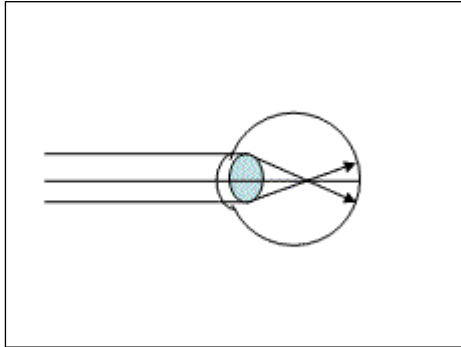
In hypermetropia, the eyeball is too short or the refractive power of the cornea and lens are too weak, and light from a near object is focused behind the retina. Children with hypermetropia have difficulty seeing near objects clearly, but if the refractive error is mild they can accommodate to focus light from near objects onto the retina. If correction is required, a convex lens which causes light to converge before hitting the cornea must be used. Again spectacles are usually prescribed for constant use.



### **Myopia (near or short sightedness)**

In myopia, the eyeball is too long or the refractive power of the cornea and lens are too great, and light from a distant object is focused in front of the retina. Myopic children can only see near objects clearly. To correct this, a

concave lens which causes light to diverge before hitting the cornea must be used. In children the spectacles are prescribed for constant use.



### **The correction of refractive errors**

Hypermetropia (a shorter eye) is the normal condition at birth and there is a high incidence of astigmatism. Subsequent eye growth normalises the refractive state of the eye. Therefore, spectacles are only prescribed if the error is beyond what would be expected and/or there is an associated eye condition such as squint (misalignment of eyes).

If the error is beyond the norm, correction may be achieved by using spectacles or contact lenses (hard or soft) or by surgery. "Laser Surgery" is at present not recommended for children in routine clinical practice.

Mild refractive errors may not need correction if the child is functioning well.

Severe refractive errors may require correction to improve vision and because they can cause amblyopia (lazy eye). In addition, if there is a significant difference in refractive error between the two eyes, this requires correction to avoid amblyopia developing in the eye that is poorly focused. In children with other disabilities, even small refractive errors may need to be corrected to optimise performance.

Spectacles are also used to treat squint, usually when the child has hypermetropia and the eyes turn in (esotropia) or less commonly if the child has myopia and the eyes turn out (exotropia). Spectacles are necessary and form an important part of the treatment if misaligned eyes are straight with spectacles. Surgery to correct misalignment is not then indicated. If the



squint is only partially corrected then surgery to align the eyes is undertaken to correct the degree of squint present with spectacles.

Spectacles can be prescribed for the youngest infant and for the child with multiple disabilities because the refractive error can be determined objectively using lenses of varying power.

Prognosis: If the refractive error is detected early and corrected with good compliance, the outlook is good.

### **Retinal detachment in childhood**

This is very uncommon in childhood and is usually secondary to trauma, surgery for congenital (present at birth) cataract or another ocular abnormality such as myopia (short sightedness), optic disc and retinal coloboma (cleft like defect), retinopathy of prematurity (a condition causing a marked increase of the retinal blood vessels), retinoschisis (splitting of the retina) and Marfan syndrome (a connective tissue multisystemic disorder). Treatment usually involves attempting to reattach the retina by surgery, laser or cryotherapy. The resulting visual acuity is very variable.

### **Visual impairment due to brain damage**

It is strongly recommended that all cases involving visual impairment due to brain damage are discussed with Medical Services.

For details of visual impairment due to brain damage see - [Scottish Sensory Centre/Strategies for dealing with visual problems due to cerebral visual impairment](#)

### **Visual pathway disorders**

#### **Acquired disorders of the optic nerve**

Papilloedema is swelling of the head of the optic nerve. It is usually due to raised intracranial (within the skull) pressure, secondary to, for example, brain tumours although other causes such as high blood pressure are possible. It does not usually cause significant visual problems and treatment is directed to the underlying cause. It can also be a sign of Idiopathic (Benign) Intracranial Hypertension (IIH).

Acquired optic atrophy is degeneration of the nerve fibres within the optic nerve. There are several possible underlying causes, including raised intracranial (within the skull) pressure (e.g. due to hydrocephalus, brain tumour), compression of the optic nerve, trauma, optic neuritis and metabolic disorders. It may cause reduced visual acuity and squint. Treatment is directed to the underlying cause.

Optic neuritis is an inflammation of the optic nerve. Unilateral (affecting one eye) optic neuritis is associated with virus infections and toxoplasmosis, bilateral (affecting both eyes) optic neuritis is usually of unknown cause but in teenagers, may be associated with multiple sclerosis. Optic neuritis usually presents with a sudden onset of eye pain and visual loss. It may be treated

with systemic steroids to speed up the resolution of symptoms. There is usually good recovery of visual acuity.

Optic nerve glioma is a tumour that affects the optic nerve. They are more common in children with Neurofibromatosis (a genetic disease causing multiple soft tumours under the skin) type 1 (NF1) but may occur sporadically in the absence of NF1. Children may present with a bulging eye (proptosis) or squint but may be asymptomatic (without symptoms) and visual acuity is usually well preserved. In children with NF1 they tend to be relatively benign and may regress spontaneously but in sporadic cases where there is rapid growth or vision is affected, they should be surgically removed.

Trauma from a head injury may result in damage to the optic nerve. Shaking in non-accidental injury may cause retinal haemorrhages, bleeding into the optic nerve and damage to the visual cortex, thereby affecting visual acuity.

Idiopathic or Benign Intracranial Hypertension is an incompletely understood medical condition in which the development of raised pressure within the skull causes symptoms such as headache, nausea and blurring of vision or double vision. The cause can often not be determined although the condition is seen more commonly in teenage girls and young women, many of whom are overweight or obese. It may also arise following the use of some medication (certain antibiotics, steroids, the oral contraceptive pill). There is no single treatment for the condition and the prognosis is variable, with some patients recovering completely and others experiencing on-going symptoms or periodic relapses. The term "benign" refers to the fact that the condition is not life threatening but many experts consider it to be misleading as the condition can have serious consequences for vision. A small proportion of people with IIH develop severe permanent loss of vision. Recent UK figures suggest this applies to up to 2% of cases but higher figures (up to 10%) have previously been quoted.

Disorders of the visual pathway affect the optic nerve and its communications with the area of the brain responsible for vision, the visual cortex. They result in visual loss but usually without other visual symptoms. Younger children rarely complain of unilateral loss of vision and bilateral visual loss may not be symptomatic until it is very advanced.

### **Congenital (present at birth) disorders of the optic nerve**

Optic nerve hypoplasia (ONH) is an uncommon condition where there is underdevelopment of the optic nerve. Bilateral cases may result in severe visual impairment and nystagmus (jerky eye movements) in infancy. Unilateral cases may be less severe and result in visual field defects and squint. ONH may be associated with brain abnormalities especially with defects in the pituitary gland, which may cause problems in growth. ONH may be caused by alcohol and drug abuse during pregnancy (Foetal Alcohol Syndrome).

Optic nerve coloboma (a cleft like defect) may affect the optic nerve alone or the iris and retina as well. The severity of visual loss is determined by whether the macula (central retina) is affected and by the severity of the optic nerve abnormality. It is associated with an increased risk of retinal

detachment. There is no specific treatment for the coloboma (cleft-like defect) but treatment of any refractive error and amblyopia (lazy eye) is required.

Optic disc dysplasia is a condition where there is abnormal development of the optic nerve. The child usually presents with a squint due to a refractive error and astigmatism. It is associated with an increased risk of retinal detachment. In most children, vision is good and management involves correcting any refractive error and treating amblyopia if present.

Congenital optic atrophy is a condition where the optic nerves are normal size but there has been degeneration of the nerve fibres within the optic nerve during pregnancy. Children with congenital (present at birth) optic atrophy usually have multiple neurological and developmental problems. The prognosis for visual acuity varies widely.

### **Disorders of the optic chiasm**

The optic chiasm is the place where the optic nerves meet and crossover behind the eyes. A variety of disorders may affect the optic chiasm, including developmental defects, tumours, inflammation and trauma. They may present in infancy as blindness, poor visual acuity and nystagmus or with progressive visual failure later in childhood. Visual acuity is often severely reduced at presentation and colour vision is also often affected. Treatment, if possible, is directed towards the underlying cause.

### **Disorders of the posterior visual pathways**

Disorders of the visual pathway after the optic chiasm, including the visual cortex are often associated with other developmental and neurological problems. They may have several causes that include developmental defects, trauma or tumours. Children may present in infancy with poor fixation, squint and abnormal eye movements. Visual function will depend upon the nature and extent of the underlying pathology. Children with disorders of the visual cortex often demonstrate improvement in visual function over a period of time.

Delayed visual maturation is a condition where an infant is noticed to have no eye contact or visual interest in the first six to eight weeks of life. No underlying cause for visual loss is found and spontaneous improvement in vision occurs at about twelve to fifteen weeks of age so that vision is eventually normal.

There is usually no specific treatment for children with visual pathways disorders. The extent of any visual deficiency may not be fully appreciated until the child starts school. Regular follow up is required, together with

correction of any refractive errors, provision of low visual aids, the management of amblyopia and surgical correction of unsightly squint.

### **Severity indicators**

#### **Mild Condition**

The following would normally be characteristic of a child with a mild condition.

Visual acuity of greater than ( $>$ ) 6/18 (20/60)

Reasonable visual fields

#### **Moderate condition**

The following would normally be characteristic of a child with a moderate condition.

Visual acuity of equal to ( $=$ ) or less than ( $<$ ) 6/18 (20/60) but greater than ( $>$ ) 6/36 (10/60)

Moderately reduced visual fields

#### **Severe condition**

The following would normally be characteristic of a child with a severe condition.

Visual acuity of equal to ( $=$ ) or less than ( $<$ ) 6/36 (10/60)

- Considerably reduced visual fields

#### **Higher rate mobility**

This applies to children who have been certified as severely visually impaired by a consultant Ophthalmologist AND their visual acuity is:

- Less than ( $<$ ) 3/60; or
- 3/60 or more but less than ( $<$ ) 6/60 with a complete loss of peripheral visual field and a central visual field of no more than 10 degrees in total?

Note: The presence of eye movement disorders such as nystagmus (jerky eye movements) may change the disability from mild to moderate or indeed severe. Similarly, poorly functioning children with intense photophobia (sensitivity to light), cerebral visual impairment (due to damage to the brain) with perceptual deficits (reduced ability to recognise and interpret objects) may

affect the level of disability independent of high contrast Snellen Visual Acuity.

### **Deeming Provisions**

Combination of blindness and hearing loss (Deaf-blind)

Deaf - blindness is defined by Sense (the National Deaf - blind and Rubella Association) as:

“A severe degree of combined visual and auditory impairment resulting in special needs in the areas of communication, access to information and mobility.”

100% disablement due to visual impairment

This is defined as being so blind as to be unable to perform any work for which eyesight is essential, the same criterion for being registered blind (see Registration of blindness/Partially sighted section). This equates to

visual acuity of 3/60 or less or visual acuity between 3/60 and 6/60 with visual field loss or visual acuity above 6/60 with very severe visual field loss.

80% disablement due to hearing impairment

The level of hearing loss must be 87dB or greater when aids are used, and it is usual to assess the degree of hearing loss by audiometry.

However, since audiograms are almost invariably performed without aids, it has been accepted that at this level of hearing loss, the use of a hearing aid is unlikely to provide significant improvement.

Medical Services advice should be sought in these cases.

Deaf – Blind Deeming Provision

Under the Deeming Provisions for DLA, a claimant can satisfy the conditions for the higher rate mobility component if -:

- they are both deaf and blind and
- as a result of the combined effects

they are unable to walk to their destination out of doors without the assistance of another person.

Legislation advises that the claimant must be 100% disabled due to blindness and 80% disabled due to deafness to be considered under the Deeming Provisions.

Higher Rate Mobility component Severely Visually Impaired (SVI) Higher Rate Mobility component Severely Visually Impaired (SVI) Deeming Provision

Under the Deeming Provisions for DLA, a child can satisfy the conditions for the higher rate mobility component if:

They have been certified as severely visually impaired by a consultant Ophthalmologist AND their visual acuity is:

Less than (<) 3/60 ;or

3/60 or more but less than (<) 6/60 with a complete loss of peripheral visual field and a central visual field of no more than 10 degrees in total?

If this is the case, the Severely Visually Impaired (SVI) deeming provision is satisfied.

[Registration of blindness/Partially sighted](#)

**Aids and adaptations**

There are many aids and adaptations that can be used by children with visual impairment. These include:

- Refractive errors may be corrected either by the use of spectacles, contact lenses or laser surgery
- Mobility and guidance – white canes and guide dogs
- Cooking – labelling and highlighting switches
- Magnifiers and low vision aids
- Modified telephones
- Talking books, talking newspapers
- Communication – Braille, Moon, deaf blind manual, modified computers, keyboard skills, use of cassette recorders etc

**List of NHS hospitals with Ophthalmology departments  
England**

<b>Hospital Name</b>	<b>Hospital Address</b>
Addenbrooke's Hospital	Hills Road, Cambridge, Cambridgeshire, CB2 0QQ
Airedale General Hospital	Skipton Road, Steeton, Keighley, West Yorkshire, BD20 6TD
Alder Hey Hospital	Eaton Road, West Derby, Liverpool, Merseyside, L12 2AP
Alexandra Hospital	Woodrow Drive, Redditch, Worcestershire, B98 7UB
Altrincham General Hospital	Market Street, Altrincham, Cheshire, WA14 1PE
Andover War Memorial Hospital	Charlton Road, Andover, Hampshire, SP10 3LB
Arrowe Park Hospital	Arrowe Park Road, Upton, Wirral, Merseyside, CH49 5PE
Ashford Hospital	London Road, Ashford, Middlesex, TW15 3AA
Axminster Hospital	Chard Street, Axminster, Devon, EX13 5DU
Barnsley Hospital	Gawber Road, Barnsley, S75 2EP
Basingstoke and North Hampshire Hospital	Aldermaston Road, Basingstoke, Hampshire, RG24 9NA
Bassetlaw Hospital	Kilton Hill, Worksop, Nottinghamshire, S81 0BD
Bedford Hospital	South Wing, Kempston Road, Bedford, Bedfordshire, MK42 9DJ
Berkeley Hospital	Marybrook Street, Berkeley, Gloucestershire, GL13 9BL
Bexhill Hospital	Holliers Hill, Bexhill-on-Sea, East Sussex, TN40 2DZ
Bideford Hospital	Abbotsham Road, Bideford, Devon, EX39 3AG



Birch Hill Hospital	Union Road, Rochdale, Lancashire, OL12 9QB
Birmingham Midland Eye Centre (Bmec)	Dudley Road, Birmingham, West Midlands, B18 7QH
Bishop Auckland Hospital	Cockton Hill Road, Bishop Auckland, County Durham, DL14 6AD
Blackpool Victoria Hospital	Whinney Heys Road, Blackpool, Lancashire, FY3 8NR
Blandford Community Hospital	Milldown Road, Blandford Forum, Dorset, DT11 7DD
Bradford Royal Infirmary	Duckworth Lane, Bradford, West Yorkshire, BD9 6RJ
Bridlington and District Hospital	Bessingby Road, Bridlington, North Humberside, YO16 4QP
Bridport Community Hospital	Hospital Lane, Bridport, Dorset, DT6 5DR
Bristol Eye Hospital	Lower Maudlin Street, Bristol, Avon, BS1 2LX
Broomfield Hospital	Court Road, Chelmsford, Essex, CM1 7ET
Buckland Hospital	Coombe Valley Road, Dover, Kent, CT17 0HD
Burnley General Hospital	Casterton Avenue, Burnley, Lancashire, BB10 2PQ
Calderdale Royal Hospital	Salterhebble, Salterhebble, Halifax, West Yorkshire, HX3 0PW
Cannock Chase Hospital	Brunswick Road, Cannock, Staffordshire, WS11 5XY
Chase Farm Hospital	The Ridgeway, Enfield, Middlesex, EN2 8JL
Chelsea and Westminster Hospital	369 Fulham Road, London, Greater London, SW10 9NH
Cheltenham General Hospital	Sandford Road, Cheltenham, Gloucestershire, GL53 7AN

Chesterfield Royal Hospital	Calow, Chesterfield, Derbyshire, S44 5BL
Chippenham Hospital	Rowden Hill, Chippenham, Wiltshire, SN15 2AJ
Chorley and South Ribble Hospital	Preston Road, Chorley, Lancashire, PR7 1PP
Christchurch Hospital	Fairmile Road, Christchurch, Dorset, BH23 2JX
Cirencester Hospital	Tetbury Road, Cirencester, Gloucestershire, GL7 1UY
Clayton Hospital	Northgate, Wakefield, West Yorkshire, WF1 3JS
Conquest Hospital	The Ridge, St. Leonards-on-Sea, East Sussex, TN37 7RD
Countess Of Chester Hospital	The Countess Of Chester Health Park, Chester, Cheshire, CH2 1UL
County Hospital Louth	High Holme Road, Louth, Lincolnshire, LN11 0EU
Crawley Hospital	West Green Drive, Crawley, West Sussex, RH11 7DH
Cromer and District Hospital	Mill Road, Cromer, Norfolk, NR27 0BQ
Cumberland Infirmary	Newtown Road, Carlisle, Cumbria, CA2 7HY
Darent Valley Hospital	Darent Wood Road, Darent Wood Road, Dartford, Kent, DA2 8DA
Darlington Memorial Hospital	Hollyhurst Road, Darlington, County Durham, DL3 6HX
Delancey Hospital	Charlton Lane, Cheltenham, Gloucestershire, GL53 9DT
Derriford Hospital	Derriford Road, Crownhill, Plymouth, Devon, PL6 8DH
Dewsbury and District Hospital	Halifax Road, Dewsbury, West Yorkshire, WF13 4HS

Diana, Princess Of Wales Hospital	Scarcho Road, Grimsby, North East Lincolnshire, DN33 2BA
Doncaster Royal Infirmary	Armthorpe Road, Doncaster, South Yorkshire, DN2 5LT
Dorset County Hospital	Williams Avenue, Dorchester, Dorset, DT1 2JY
Ealing Hospital	Uxbridge Road, Southall, Middlesex, UB1 3HW
East Surrey Hospital	Canada Avenue, Redhill, Surrey, RH1 5RH
Eastbourne District General Hospital	Kings Drive, Eastbourne, East Sussex, BN21 2UD
Edenbridge War Memorial Hospital	Mill Hill, Edenbridge, Kent, TN8 5DA
Edgware Community Hospital	Burnt Oak Broadway, Edgware, Middlesex, HA8 0AD
Ellesmere Port Hospital	114 Chester Road, Whitby, Ellesmere Port, Merseyside, CH65 6SG
Epsom Hospital	Dorking Road, Epsom, Surrey, KT18 7EG
Essex County Hospital	Lexden Road, Colchester, Essex, CO3 3NB
Fairfield General Hospital	Rochdale Old Road, Bury, Lancashire, BL9 7TD
Fairford Hospital	The Croft, Fairford, Gloucestershire, GL7 4BB
Farnham Hospital	Hale Road, Farnham, Surrey, GU9 9QL
Finchley Memorial Hospital	Granville Road, Greater London, N12 0JE
Friarage Hospital	Northallerton, North Yorkshire, DL6 1JG
Frimley Park Hospital	Portsmouth Road, Frimley, Surrey, GU16 7UJ
Frome Victoria Hospital	Park Road, Frome, Somerset, BA11 1EY

Furness General Hospital	Dalton Lane, Barrow-In-Furness, Cumbria, LA14 4LF
George Eliot Hospital	College Street, Nuneaton, Warwickshire, CV10 7DJ
Gloucestershire Royal Hospital	Great Western Road, Gloucester, Gloucestershire, GL1 3NN
Good Hope Hospital	Rectory Road, Sutton Coldfield, West Midlands, B75 7RR
Goole and District Hospital	Woodland Avenue, Goole, North Humberside, DN14 6RX
Gosport War Memorial Hospital	Bury Road, Gosport, Hampshire, PO12 3PW
Grantham and District Hospital	101 Manthorpe Road, Manthorpe Road, Grantham, Lincolnshire, NG31 8DG
Halton General Hospital	Halton, Hospital Way, Runcorn, Cheshire, WA7 2DA
Haltwhistle War Memorial Hospital	Westgate, Haltwhistle, Northumberland, NE49 9AJ
Harrogate District Hospital	Lancaster Park Road, Harrogate, North Yorkshire, HG2 7SX
Heartlands Hospital	Bordesley Green East, Birmingham, West Midlands, B9 5SS
Hertford County Hospital	North Road, Hertford, Hertfordshire, SG14 1LP
Herts and Essex Community Hospital	Haymeads Lane, Bishop's Stortford, Hertfordshire, CM23 5JH
Hillingdon Hospital	Pield Heath Road, Uxbridge, Middlesex, UB8 3NN
Hinchingbrooke Hospital	Hinchingbrooke Park, Hinchingbrooke, Huntingdon, Cambridgeshire, PE29 6NT
Holsworthy Hospital	Dobles Lane, Holsworthy, Devon, EX22 6JQ

Homerton University Hospital	Homerton Row, London, Greater London, E9 6SR
Horsham Hospital	Hurst Road, Horsham, West Sussex, RH12 2DR
Horton General Hospital	Oxford Rd, Banbury, Oxfordshire, OX16 9AL
Hospital Of St Cross	Barby Rd, Rugby, Warwickshire, CV22 5PX
Huddersfield Royal Infirmary	Acre Street, Huddersfield, West Yorkshire, HD3 3EA
Hull Royal Infirmary	Anlaby Road, Anlaby Road, Hull, East Yorkshire, HU3 2JZ
Ilfracombe Hospital	St. Brannocks Park Road, Ilfracombe, Devon, EX34 8JF
James Paget University Hospital	Lowestoft Road, Gorleston, Great Yarmouth, Norfolk, NR31 6LA
John Coupland Hospital	Ropery Road, Gainsborough, Lincolnshire, DN21 2TJ
John Radcliffe Hospital	Headley Way, Headington, Oxford, Oxfordshire, OX3 9DU
<b>Hospital Name</b>	<b>Hospital Address</b>
Kent and Canterbury Hospital	Ethelbert Road, Canterbury, Kent, CT1 3NG
Kettering General Hospital	Rothwell Rd, Kettering, Northamptonshire, NN16 8UZ
Kidderminster General Hospital	Bewdley Road, Kidderminster, Worcestershire, DY11 6RJ
King Edward Vii Hospital	St Leonards Road, Windsor, Berkshire, SL4 3DP
King George Hospital	Barley Lane, Ilford, Essex, IG3 8YB
King's College Hospital (Denmark Hill)	Denmark Hill, London, Greater London, SE5 9RS

King's Mill Hospital	Mansfield Road, Sutton-In-Ashfield, Nottinghamshire, NG17 4JL
Kingston Hospital	Galsworthy Road, Kingston Upon Thames, Surrey, KT2 7QB
Leicester Royal Infirmary	Infirmery Square, Leicester, Leicestershire, LE1 5WW
Leigh Infirmary	The Avenue, Leigh, Lancashire, WN7 1HS
Leighton Hospital	Middlewich Road, Crewe, Cheshire, CW1 4QJ
Lincoln County Hospital	Greetwell Road, Lincoln, Lincolnshire, LN2 5QY
Lister Hospital	Coreys Mill Lane, Stevenage, Hertfordshire, SG1 4AB
Luton and Dunstable Hospital	Lewsey Road, Luton, Bedfordshire, LU4 0DZ
Lydney and District Hospital	Lydney, Gloucestershire, GL15 5JF
Maidstone Hospital	Hermitage Lane, Maidstone, Kent, ME16 9QQ
Malton Community Hospital	Malton, North Yorkshire, YO17 7NG
Manchester Royal Eye Hospital	Nelson Street, Manchester, Greater Manchester, M13 9WH
Manor Hospital	Moat Road, Walsall, West Midlands, WS2 9PS
Mary Hewetson Cottage Hospital (Keswick)	Crosthwaite Road, Keswick, Cumbria, CA12 5PH
Mayday University Hospital	530 London Road, Thornton Heath, Surrey, CR7 7YE
Medway Maritime Hospital	Windmill Road, Gillingham, Kent, ME7 5NY
Mile End Hospital	Bancroft Road, London, Greater London, E1 4DG

Millom Hospital	Lapstone Road, Millom, Cumbria, LA18 4BY
Milton Keynes Hospital	Standing Way, Eaglestone, Milton Keynes, Buckinghamshire, MK6 5LD
Montagu Hospital	Adwick Road, Mexborough, South Yorkshire, S64 0AZ
Moore Cottage Hospital	Moore Road, Bourton-on-The-Water, Cheltenham, Gloucestershire, GL54 2AZ
Moorfields Eye Hospital	162 City Road, London, Greater London, EC1V 2PD
Moreton-In-Marsh Hospital	Moreton-In-Marsh, Gloucestershire, GL56 0BS
Mount Vernon Hospital	Rickmansworth Road, Northwood, Middlesex, HA6 2RN
Musgrove Park Hospital	Musgrove Park, Parkfield Drive, Taunton, Somerset, TA1 5DA
New Cross Hospital	Wolverhampton Road, Heath Town, Wolverhampton, West Midlands, WV10 0QP
Newark Hospital	Boundary Road, Newark, Nottinghamshire, NG24 4DE
Newham General Hospital	Glen Road, Plaistow, London, E13 8SL
Norfolk and Norwich University Hospital	Colney Lane, Norwich, Norfolk, NR4 7UY
North Devon District Hospital	Raleigh Park, Barnstaple, Devon, EX31 4JB
North Middlesex University Hospital	Sterling Way, Greater London, N18 1QX
Northampton General Hospital	Cliftonville, Northampton, Northamptonshire, NN1 5BD
Northwick Park Hospital	Watford Road, Harrow, Middlesex, HA1 3UJ
Nottingham University Hospital - Queen's Medical Centre Campus	Derby Road, Nottingham, Nottinghamshire, NG7 2UH

Nuffield Diagnostic Centre	Cottingham Rd, Corby, Northamptonshire, NN17 2UW
Ormskirk and District General Hospital	Wigan Road, Ormskirk, Lancashire, L39 2AZ
Palmer Community Hospital	Wear Street, Jarrow, Tyne and Wear, NE32 3UX
Paulton Memorial Hospital	Salisbury Road, Paulton, Bristol, Avon, BS39 7SB
Pembury Hospital	Tonbridge Road, Pembury, Tunbridge Wells, Kent, TN2 4QJ
Penrith Hospital	Bridge Lane, Penrith, Cumbria, CA11 8HX
Peterborough District Hospital	Thorpe Road, Peterborough, Cambridge-shire, PE3 6DA
Pilgrim Hospital	Sibsey Road, Boston, Lincolnshire, PE21 9QS
Pinderfields General Hospital	Aberford Road, Wakefield, West Yorkshire, WF1 4DG
Pontefract General Infirmary	Friarwood Lane, Pontefract, West Yorkshire, WF8 1PL
Poole General Hospital	Longfleet Road, Poole, Dorset, BH15 2JB
Potters Bar Community Hospital	Barnet Road, Potters Bar, Hertfordshire, EN6 2RY
Princess Alexandra Hospital	Hamstel Road, Harlow, Essex, CM20 1QX
Princess Royal Hospital	Lewes Road, Haywards Heath, West Sus-sex, RH16 4EX
Princess Royal University Hos-pital	Farnborough Common, Orpington, Kent, BR6 8ND
Purley War Memorial Hospital	856 Brighton Road, Purley, Surrey, CR8 2YL
Queen Alexandra Hospital	Southwick Hill Road, Cosham, Portsmouth, Hampshire, PO6 3LY



Queen Elizabeth I I Hospital	Howlands, Welwyn Garden City, Hertfordshire, AL7 4HQ
Queen Elizabeth The Queen Mother Hospital	St Peters Road, Margate, Kent, CT9 4AN
Queen Mary's Hospital Sidcup	Frognaal Avenue, Sidcup, Kent, DA14 6LT
Queen Victoria Hospital	Thornton Rd, Morecambe, Lancashire, LA4 5NN
Queen Victoria Hospital (East Grinstead)	Holtye Road, East Grinstead, West Sussex, RH19 3DZ
Queen's Hospital	Rom Valley Way, Romford, Essex, RM7 0AG
Queen's Hospital, Burton Upon Trent	Belvedere Road, Burton -on- Trent, Staffordshire, DE13 0RB
Ripon and District Community Hospital	Firby Lane, Ripon, North Yorkshire, HG4 2PR
Rossendale Hospital	Haslingden Road, Rossendale, Lancashire, BB4 6NE
Rotherham Hospital	Moorgate Road, Rotherham, S60 2UD
Rowley Regis Hospital	Moor Lane, Rowley Regis, West Midlands, B65 8DA
Royal Albert Edward Infirmary	Wigan Lane, Wigan, Lancashire, WN1 2NN
Royal Berkshire Hospital	London Road, Reading, Berkshire, RG1 5AN
Royal Blackburn Hospital	Haslingden Road, Blackburn, Lancashire, BB2 3HH
Royal Bolton Hospital	Minerva Road, Farnworth, Bolton, Lancashire, BL4 0JR
Royal Bournemouth General Hospital	Castle Lane East, Bournemouth, Dorset, BH7 7DW
Royal Cornwall Hospital (Treliske)	Treliske, Truro, Cornwall, TR1 3LJ

Royal Derby Hospital	Uttoxeter Road, Derby, Derbyshire, DE22 3NE
Royal Devon and Exeter Hospital (Wonford)	Barrack Road, Exeter, Devon, EX2 5DW
Royal Eye Infirmary	Apsley Road, Plymouth, Devon, PL4 6PL
Royal Free Hospital	Pond Street, Greater London, NW3 2QG
Royal Hallamshire Hospital	Glossop Road, Sheffield, South Yorkshire, S10 2JF
Royal Hampshire County Hospital	Romsey Road, Winchester, Hampshire, SO22 5DG
Royal Lancaster Infirmary	Ashton Road, Lancaster, Lancashire, LA1 4RP
Royal London Homeopathic Hospital	60 Great Ormond Street, London, Greater London, WC1N 3HR
Royal Oldham Hospital	Rochdale Road, Oldham, Lancashire, OL1 2JH
Royal Preston Hospital	Sharoe Green Lane North, Fulwood, Preston, Lancashire, PR2 9HT
Royal Shrewsbury Hospital	Mytton Oak Road, Shrewsbury, Shropshire, SY3 8XQ
Royal Surrey County Hospital	Egerton Road, Guildford, Surrey, GU2 7XX
Royal United Hospital	Combe Park, Bath, Avon, BA1 3NG
Royal Victoria Hospital (Folkestone)	Radnor Park Avenue, Folkestone, Kent, CT19 5BN
Russells Hall Hospital	Pensnett Road, Dudley, West Midlands, DY1 2HQ
<b>Hospital Name</b>	<b>Hospital Address</b>
Saffron Walden Community Hospital	Radwinter Road, Saffron Walden, Essex, CB11 3HY
Salford Royal	Stott Lane, Salford, M6 8HD

Salisbury District Hospital	Odstock Road, Salisbury, Wiltshire, SP2 8BJ
Sandwell General Hospital	Lyndon, West Bromwich, West Midlands, B71 4HJ
Savernake Hospital	General Hospital, Marlborough, Wiltshire, SN8 3HL
Scarborough General Hospital	Woodlands Drive, Scarborough, North Yorkshire, YO12 6QL
Scunthorpe General Hospital	Cliff Gardens, Scunthorpe, North East Lincolnshire, DN15 7BH
Selby and District War Memorial Hospital	Doncaster Road, Selby, North Yorkshire, YO8 9BX
Selly Oak Hospital	Raddlebarn Road, Selly Oak, Birmingham, B29 6JD
Sevenoaks Hospital	Hospital Road, Sevenoaks, Kent, TN13 3PG
Sheffield Children's Hospital	Western Bank, Sheffield, South Yorkshire, S10 2TH
Shepton Mallet Community Hospital	Old Wells Road, Shepton Mallet, Somerset, BA4 4PG
Sittingbourne Memorial Hospital	Bell Road, Sittingbourne, Kent, ME10 4DT
Skegness and District General Hospital	Dorothy Avenue, Skegness, Lincolnshire, PE25 2BS
Skipton General Hospital	Keighley Road, Skipton, North Yorkshire, BD23 2RJ
Solihull Hospital	Lode Lane, Solihull, West Midlands, B91 2JL
South Molton Hospital	Widgery Drive, South Molton, Devon, EX36 4DP
South Tyneside District Hospital	Harton Lane, South Shields, Tyne and Wear, NE34 0PL

Southampton General Hospital	Tremona Road, Southampton, Hampshire, SO16 6YD
Southend Hospital	Prittlewell Chase, Westcliff-on-Sea, Essex, SS0 0RY
Southport and Formby District General Hospital	Town Lane, Southport, Merseyside, PR8 6PN
St Albans City Hospital	Waverley Road, St. Albans, Hertfordshire, AL3 5PN
St Ann's Hospital	St. Ann's Road, London, Greater London, N15 3TH
St Bartholomew's Hospital	West Smithfield, London, Greater London, EC1A 7BE
St George's Hospital	Blackshaw Road, London, Greater London, SW17 0QT
St Helens Hospital	Marshalls Cross Road, St. Helens, Merseyside, WA9 3DA
St James's University Hospital	Beckett Street, Leeds, West Yorkshire, LS9 7TF
St Peter's Hospital	Guildford Road, Chertsey, Surrey, KT16 0PZ
St Peter's Hospital	Spital Road, Maldon, Essex, CM9 6EG
St Thomas' Hospital	Westminster Bridge Road, London, Greater London, SE1 7EH
St. Margaret's Hospital	The Plain, Epping, Essex, CM16 6TN
Stafford Hospital	Weston Road, Stafford, Staffordshire, ST16 3SA
Stamford and Rutland Hospital	Ryhall Road, Stamford, Lincolnshire, PE9 1UA
Stepping Hill Hospital	Poplar Grove, Stockport, Cheshire, SK2 7JE
Stoke Mandeville Hospital	Mandeville Road, Aylesbury, Buckinghamshire, HP21 8AL

Stratford Hospital	Arden Street, Stratford-upon-Avon, Warwickshire, CV37 6NX
Stretford Memorial Hospital	226 Seymour Grove, Stretford, Manchester, Greater Manchester, M16 0DU
Stroud General Hospital	Trinity Road, Stroud, Gloucestershire, GL5 2HY
Sunderland Eye Infirmary	Queen Alexandra Road, Sunderland, Tyne and Wear, SR2 9HP
Sussex Eye Hospital	Eastern Road, Brighton, East Sussex, BN2 5BF
Sutton Hospital	Cotswold Road, Sutton, Surrey, SM2 5NF
Tameside General Hospital	Fountain St, Ashton Under Lyne, Lancashire, OL6 9RW
Tewkesbury General Hospital	Barton Road, Tewkesbury, Gloucestershire, GL20 5QN
The County Hospital	The County Hospital, Union Walk, Hereford, Herefordshire, HR1 2ER
The Great Western Hospital	Marlborough Road, Marlborough Road, Swindon, Wiltshire, SN3 6BB
The Ipswich Hospital	Heath Road, Ipswich, Suffolk, IP4 5PD
The James Cook University Hospital	Marton Road, Middlesbrough, Cleveland, TS4 3BW
The Johnson Community Hospital	Spalding Road, Pinchbeck, Spalding, Lincolnshire, PE11 3PB
The Princess Royal Hospital	Apley Castle, Grainger Drive, Telford, Shropshire, TF1 6TF
The Queen Elizabeth Hospital	Gayton Road, King's Lynn, Norfolk, PE30 4ET
The Royal Liverpool University Hospital	Prescot Street, Liverpool, Merseyside, L7 8XP
The Royal London Hospital	Whitechapel, London, Greater London, E1 1BB

The Royal Marsden Hospital (London)	Fulham Road, London, Greater London, SW3 6JJ
The Royal Victoria Infirmary	Queen Victoria Road, Newcastle Upon Tyne, Tyne and Wear, NE1 4LP
The Whittington Hospital	Magdala Avenue, London, Greater London, N19 5NF
Tiverton and District Hospital	Kennedy Way, Tiverton, Devon, EX16 6NT
Torbay Hospital	Newton Road, Torquay, Devon, TQ2 7AA
Torrington Hospital	Calf Street, Torrington, Devon, EX38 7BJ
Trowbridge Hospital	Adcroft Street, Trowbridge, Wiltshire, BA14 8PH
University College Hospital	235 Euston Road, London, Greater London, NW1 2BU
University Hospital (Coventry)	Clifford Bridge Road, Coventry, West Midlands, CV2 2DX
University Hospital Of North Durham	North Road, Durham, County Durham, DH1 5TW
University Hospital Of North Staffs	Hartshill Road, Stoke-on-Trent, Staffordshire, ST4 7PA
University Hospital Of North Tees	Hardwick Road, Stockton-on-Tees, Cleveland, TS19 8PE
Upney Lane Centre	132 Upney Lane, Barking, Essex, IG11 9LX
Victoria Hospital (Deal)	London Road, Deal, Kent, CT14 9UA
Victoria Hospital (Sidmouth)	Sidmouth, Devon, EX10 8EW
Victoria Infirmary (Northwich)	Winnington Hill, Northwich, Cheshire, CW8 1AW
Walton Hospital	Rice Lane, Liverpool, Merseyside, L9 1AE
Warrington Hospital	Lovely Lane, Warrington, Cheshire, WA5 1QG

Warwick Hospital	Lakin Road, Warwick, Warwickshire, CV34 5BW
Watford General Hospital	60 Vicarage Road, Watford, Hertfordshire, WD18 0HB
Watford General Hospital	Vicarage Road, Watford, Hertfordshire, WD18 0HB
West Berkshire Community Hospital	London Road, Benham Hill, Thatcham, Berkshire, RG18 3AS
West Cornwall Hospital (Penzance)	St Clare Street, Penzance, Cornwall, TR18 2PF
West Cumberland Hospital	Homewood, Hensingham, Whitehaven, Cumbria, CA28 8JG
West Suffolk Hospital	Hardwick Lane, Bury St Edmunds, Suffolk, IP33 2QZ
Westmorland General Hospital	Burton Rd, Kendal, Cumbria, LA9 7RG
Weston General Hospital	Grange Road, Uphill, Weston-Super-Mare, BS23 4TQ
Wharfedale Hospital	Newall Carr Road, Otley, West Yorkshire, LS21 2LY
Whipps Cross University Hospital	Whipps Cross Road, London, Greater London, E11 1NR
Whitby Community Hospital	Springhill, Whitby, North Yorkshire, YO21 1EE
Whitstable and Tankerton Hospital	174-176 Northwood Road, Whitstable, Kent, CT5 2HN
Whitworth Hospital	330 Bakewell Road, Darley Dale, Matlock, Derbyshire, DE4 2JD
William Harvey Hospital (Ashford)	Kennington Road, Willesborough, Ashford, Kent, TN24 0LZ
Worcestershire Royal Hospital	Charles Hastings Way, Worcester, Worcestershire, WR5 1DD

Workington Community Hospital	Park Lane, Workington, Cumbria, CA14 2RW
Wycombe Hospital	Queen Alexandra Road, High Wycombe, Buckinghamshire, HP11 2TT
Yeatman Hospital	Yeatman Hospital, Hospital Lane, Sherborne, Dorset, DT9 3JU
Yeovil District Hospital	Higher Kingston, Yeovil, Somerset, BA21 4AT
York Hospital	Wigginton Road, York, North Yorkshire, YO31 8HE

## Scotland



<b>Hospital Name</b>	<b>Hospital Address</b>
Abbey Carrick Glen Hospital	Dalmellington Road, Ayr, KA6 6PG
Aberdeen City Hospital	Urquhart Road, Aberdeen, AB24 5AU
Aberdeen Royal Infirmary	Foresterhill, Aberdeen, AB25 2ZN
The Ayr Hospital	Dalmellington Road, Ayr, KA6 6DX
Balfour Hospital	New Scapa Road, Kirkwall, Orkney, KW15 1BH
Borders General Hospital	Melrose, TD6 9BS
City Health Clinic	9 Earl Grey Street, Edinburgh, EH3 9BN
Crosshouse Hospital	Crosshouse, Kilmarnock, KA2 0BE
Dr Gray's Hospital	Pluscarden Road, Elgin, IV30 1SN
Dumfries and Galloway Royal Infirmary	Bankend Road, Dumfries, DG1 4AP
Dalrymple Hospital	Stranraer, DG9 7DQ
Falkirk and District Royal Infirmary	Major's Loan, Falkirk, FK1 5QE
Gartnavel General Hospital	1053 Great Western Road, Glasgow, G12 0YN
Golden Jubilee National Hospital	Beardmore Street, Clydebank, Glasgow, G81 4HX
Glasgow Royal Infirmary	84 Castle Street, Glasgow, G4 0SF
Hairmyres Hospital	Eaglesham Road, East Kilbride, G75 8RG
Hay Lodge Hospital	Neidpath Road, Peebles, EH45 8JG
Inverclyde Royal Hospital	Larkfield Road, Greenock, PA16 0XN
Jubilee Hospital	Bleachfield Street, Huntly, AB54 8EX
Monklands Hospital	Monkscourt Avenue, Airdrie, ML6 0JS

Montrose Royal Infirmary	Montrose, DD10 8AJ
The New Victoria Hospital	Grange Road, Glasgow, G42 9LF
Ninewells Hospital	Dundee, DD1 9SY
Peterhead Community Hospital	21 Links Terrace, Peterhead, AB42 2XB
Princess Alexandra Eye Pavilion	Chalmers Street, Edinburgh, EH3 9HA
Perth Royal Infirmary	Perth, PH1 1NX
Queen Margaret Hospital	Whitefield Road, Dunfermline, KY12 0SU
Raigmore Hospital	Old Perth Road, Inverness, IV2 3UJ
Royal Aberdeen Children's Hospital	Westburn Road, Foresterhill, Aberdeen, AB25 2ZG
Royal Hospital for Sick Children	Yorkhill, Glasgow, G3 8SJ
Royal Alexandra Hospital	Corsebar Road, Paisley, PA2 9PN
Royal Hospital for Sick Children	9 Sciennes Road, Edinburgh, EH9 1LF
Stirling Royal Infirmary	Livilands, Stirling, FK8 2AU
Southern General Hospital	Glasgow, G51 4TF
Spire Murrayfield Hospital	122 Corstophine Road, Edinburgh, EH12 6UD
St John's Hospital at Howden	Howden, Livingston, EH54 6PP
Stobhill ACH Hospital	133 Balornock Road, Glasgow, G21 3UW
Turner Memorial Hospital	Turner Street, Keith, AB55 5DJ
Uist and Barra Hospital	Balivanich, Benbecula, HS7 5LA
Victoria Hospital	Hayfield Road, Kirkcaldy, KY2 5AH
Western Isles Hospital	Macaulay Road, Stornoway, Isle of Lewis, HS1 2AF
Western Infirmary	Dumbarton Road, Glasgow, G11 6NT

Wishaw General Hospital	50 Netherton Street, Wishaw, ML2 0DP
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**Wales**

<b>Hospital Name</b>	<b>Hospital Address</b>
Bridgend Clinic, Princes of Wales Hospital	Coity Road, Bridgend, CF31 1RQ
Bro Ddyfi Community Hospital	Machynlleth, SY20 8AD
Bronglais General Hospital	Aberystwyth, SY23 1ER
Bryn Beryl Hospital	Pwllheli, LL53 6TT
Colwyn Bay Community Hospital	Hesketh Road, Colwyn Bay, LL29 8AY
Dolgellau & Barmouth District Hospital	Dolgellau, LL40 1NP
Ffestiniog Memorial Hospital	Blaenau, Ffestiniog, LL41 3DW
Glan Clwyd Hospital	Bodelwyddan, Rhyl, LL18 5UJ
H M Stanley Hospital	St Asaph, LL17 0RS
Llandrindod Wells Hospital	Temple Street, Llandrindon, LD1 5HF
Llandudno General Hospital	Llandudno, LL30 1LB
Longfield Medical Consulting Rooms	196 Stow Hill, Newport, Gwent, NP20 4HB
MDA Clinic	65 Cardiff Road, Llandaff, Cardiff, CF5 2AA
Neath Port Talbot Hospital	Baglan Way, Port Talbot, SA12 7BX
Nevill Hall Hospital	Brecon Road, Abergavenny, NP7 7EG
Prince Charles General Hospital	Merthyr Tydfil, CF47 9DT
Prince Philip Hospital	Bryngynmawr, Dafen, Llanelli, SA14 8QF
Royal Glamorgan Hospital	Ynysmaerdy, Llantrisant, CF72 8XR
Royal Gwent Hospital	Cardiff Road, Newport, Gwent, NP20 2UB
Sancta Maria Hospital	Ffynone Road, Swansea, SA1 6DF
Singleton Hospital	Sketty Lane, Swansea, SA2 8QA

South Pembrokeshire Hospital	Fort Road, Pembroke Dock, SA72 6SX
Spire Cardiff Hospital	Croescadarn Road, Pentwyn, Cardiff, CF23 8XL
Spire Yale Hospital	Wrexham Technology Park, Croesnewydd Road, Wrexham, LL13 7YP
St Asaph Consulting Rooms, Yale Hospital	St Asaph rooms, 88 Bowen Court, St Asaph, LL17 0JE
St Joseph's Hospital	Harding Avenue, Malpas, Newport, Gwent, NP20 6ZE
St Tysilio Medical Centre	Llanfairpwll, Anglesey, LL61 5YR
Tenby Cottage Hospital	Church Park, Tenby, SA70 7EE
Twywn Hospital	Bryn Hyfryd Road, Twywn, LL36 9HH
University Hospital of Wales	Heath Park, Cardiff, CF14 4XW
Vale Healthcare Cardiff By Clinic	Dunleavy Drive, Cardiff Bay, Cardiff, CF11 0SN
Victoria War Memorial Hospital	Salop Road, Welshpool, SY21 7DU
West Wales General Hospital	Glangwili, Carmarthen, SA31 2AF
Withybush General Hospital	Fishguard Road, Haverfordwest, SA61 2PZ
Wrexham Maelor Hospital	Croesnewydd Road, Wrexham, LL13 7TD
Ysbyty Gwynedd	Penrhosgarnedd, Bangor, Gwynedd, LL57 2PW
Ysbyty Penrhos Stanley	Holyhead, LL65 2PR
Ystrad Mynach Hospital	Caerphilly Road, Ystrad Mynach, Hengoed, CF82 7XU
Ystradgynlais Community Hospital	Glanrhyd Road, Ystradgynlais, Swansea, SA9 1AE

**Registration of visual impairment  
Form CVI**

Children with severe visual impairment will be seen by a consultant ophthalmologist who will issue a Certificate of Visual Impairment (CVI).

This formally certifies the child as either:

- Severely sight impaired (formerly “blind”)
- Sight impaired (formerly “partially sighted”)

The form is sent to Social Services, who will register the child, arrange for an assessment of needs and provide information about the services and benefits available.

### **Severely sight impaired**

The definition of severely sight impaired is “so blind as to be able to perform any work for which eyesight is essential.”

This is equivalent to 100% disablement for the DLA deaf/ blind deeming provision.

A person should be certified as severely sight impaired if they have:

- Visual acuity less than 3/60 or 1/18 Snellen.
- Visual acuity between 3/60 and 6/60 with a very restricted visual field.

### **Sight impaired**

The definition of sight impaired is “substantially and permanently handicapped by defective vision caused by congenital (present at birth) defect or illness or injury.”

A person should be certified as sight impaired if they have:

- Visual acuity between 3/60 and 6/60 with a full visual field
- Visual acuity between 6/24 and 6/60, with a moderately restricted visual field or aphakia (absent lens) or opacities (clouding) blocking vision in the eye itself.
- Visual acuity 6/18 or better with a gross defect of visual fields (of both eyes, such as hemianopia) or marked contraction of the visual field (i.e. in retinitis pigmentosa or glaucoma).

Infants and young children who have congenital ocular abnormalities leading to visual defects should be certified as sight impaired unless they are obviously severely sight impaired.

Children should be certified as severely sight impaired or sight impaired according to the corrected binocular vision.

## **Form RVI**

Staff in the hospital eye service may issue form RVI (Referral of Vision Impaired patient) to refer a patient (with their consent) for a social care assessment. This should be done as soon as social needs become apparent,

but where certification is not currently appropriate or cannot be carried out, for example if they are not being seen by a consultant.

### **LVL leaflet**

People with visual impairment can also self refer themselves to social services using leaflet LVL.

## **What you need to know about Wilms tumours**

### **What is Wilms tumours?**

Wilms tumour is a type of kidney cancer that affects young children.....

- [Wilms tumours](#)

### **What are the effects and signs?**

The most common feature of Wilms tumour is an abdominal mass or abdominal swelling.....

- [Effects of Wilms tumours](#)

### **How is it assessed?**

Investigations are likely to include Blood tests, Urine tests, Imaging studies .....

- [Assessment of Wilms tumours](#)

### **How is it treated and managed?**

Most children over the age of 6 months will begin their treatment with 4 weeks of pre-operative chemotherapy.....

- [Treatment of Wilms tumours](#)

### **What evidence is available?**

The best source of information will be the hospital based specialist nurse or the consultant oncologist .....

- [Evidence](#)

### **How long will the needs last?**

- [Prognosis and duration of the award](#)



## **What is a Wilms tumour?**

Wilms tumour is a type of kidney cancer that affects young children. The tumour is an abnormal proliferation of a type of cell called a metanephric blastema cell. These cells are embryological remnants leftover from kidney development in the womb. Tumours grow within the kidney itself, a tumour confined to the kidney may be termed 'intrarenal' in the medical evidence. 40% of tumours extend from the kidney into the renal vein and a smaller number extend into the ureter and sometimes the bladder.

It is the fifth most common cancer in childhood. The mean age at diagnosis of children affected is 3.5 years. In 6% both kidneys are affected by Wilms tumour.

10% of children who develop Wilms tumour have a recognisable congenital abnormality, the most common are listed below. Such children are routinely screened for Wilms tumour through early childhood:

- WAGR syndrome (Wilms Aniridia Genitourinary anomalies mental Retardation) – 50% of children with WAGR develop Wilms tumour and 5% of children with Wilms tumour have this syndrome – aniridia means the iris or coloured part of the eye is absent
- Denys-Drash Syndrome – rare, Wilms tumour develops in most cases. Other features are abnormal genitals (e.g. Intersex) mesangial renal sclerosis – abnormal kidney changes that cause nephrotic syndrome and renal failure
- Horseshoe kidney – single abnormally shaped kidney
- Hypospadias – abnormally placed urethral opening
- Cryptorchism – testis not present in scrotum
- Beckwith Wiedemann Syndrome – 7.5% develop Wilms tumour. Other features include abdominal hernia of varying severity, macroglossia (large tongue), enlarged internal organs and hemihypertrophy (overgrowth of one half of the body leading to asymmetry)

5 year survival after treatment of Wilms tumour has improved dramatically in the UK from 29% in the 1960s to 80% in the most recent figures.

### **Incidence/prevalence**

Around 80 children develop Wilms tumour in the UK each year. The vast majority will be under 5 years old at diagnosis. Overall survival is very good

- at least 90%. The majority of children have good prognosis disease and expected survival approaches 100% in this group.

**What are the effects and signs?**

The most common feature of Wilms tumour is an abdominal mass or abdominal swelling noticed by the parents or the child. It is usually painless. Occasionally bleeding within the tumour may lead to loin pain. More rarely

the tumour may cause high blood pressure, high temperatures, blood in the urine (haematuria) and even more rarely anaemia – low blood count.

### **Indicators of severe functional restriction**

These indicators of severe functional restriction identify the children who are most likely to undergo prolonged treatment:

- The child has bilateral Wilms tumour (stage 5)
  - The tumour has spread to other parts of the body such as the lung (stage 4)
  - The child is having radiotherapy as part of their treatment
  - The child is in the 'high risk group'
  - Recurrent disease - Wilms tumour has returned after treatment and further treatment is planned/ongoing
- 
- [Care and mobility considerations](#)

### **How is it assessed?**

#### **Blood tests**

- Full blood count (FBC)
- Clotting studies (acquired von Willebrand disease affects up to 8% of children with Wilms tumour)
- Liver function
- Urea and electrolytes (kidney function)
- Calcium

#### **Urine tests**

- Microscopy and culture

#### **Imaging studies**

These are very important and include:

- Ultrasound scan –may include 'Doppler study' a scan of the renal vein and inferior vena cava to see if there is tumour or

tumour related clot in the main vein in the body or the renal vein that drains in to it from the kidney

- Computed Tomography (CT) scan
- Magnetic Resonance Imaging (MRI) scan

CT and MRI are used to confirm that the tumour is within the kidney and not for example a neuroblastoma in the adjacent adrenal gland. These scans assess the extent of the spread of the tumour prior to surgery.

### **Tumour biopsy**

Tumour biopsy is usually required to diagnose and stage Wilms tumour. This is often carried out even when the tumour cannot be completely removed. In the UK biopsy is done prior to chemotherapy.

### **Histology**

This is the appearance of the cells from the tumour biopsy under the microscope. The appearance of the cells and other markers indicate how likely the tumour is to recur after treatment. A tumour with features associated with a low risk of recurrence is called 'low risk'. In the medical evidence children may be referred to as 'low risk' or in the 'low risk group'. These terms refer to the histology of the tumour they had and along with the spread of the tumour (the stage) is the most important determinant of how much treatment is required. Tumours may have characteristics that show a higher risk

of recurrence after treatment, these are the 'intermediate risk' tumours and those with that show they are most likely to recur are called 'high risk'.

## **Staging of Wilms tumour**

### Stage 1

35% of children will be stage 1:

- Tumour is confined to the kidney
- The tumour has not broken through the layer around the kidney called the renal capsule

### Stage 2

15% of children:

- the tumour extends beyond the kidney or into blood vessels but is completely removed at surgery

### Stage 3

35% of children are in this group because one or more of the following is found during the operation to remove their tumour:

- the tumour is not completely removed at surgery
- lymph nodes in the abdominal cavity contain tumour cells
- tumour ruptures spilling tumour cells before or during surgery
- tumour cells are found growing away from the tumour on the peritoneal lining of the abdominal cavity (peritoneal deposits or seedlings)

### Stage 4

15% of children:

- Tumour cells have spread via the bloodstream to the lung, liver, bones or the brain and can be seen on CT or MRI scanning

### Stage 5

5% of children:

- Wilms tumour has developed in both kidneys 'bilateral' disease

## **How is it treated and managed?**

### **Overview**

Treatment of children with Wilms tumour will be at regional paediatric cancer centres that may be some distance from the child's home. Type and duration of treatment will be based on two factors:

- stage of tumour
- histology – low, intermediate or high risk groups

Most children over the age of 6 months will begin their treatment with 4 weeks of pre-operative chemotherapy followed by surgery. Most children will have some post operative chemotherapy treatment as well. Wilms tumour is rare in children under 6 months - they usually have surgery as the first stage of their treatment.

The likely treatments are listed using staging information but this will not be enough information to work out the duration of treatment in some cases. In these cases, the total duration of treatment needed will not become clear until after the child has had surgery to remove their tumour. Care needs relate directly to type and duration of treatment and recovery time.

### **Stage 1 low and intermediate risk groups**

Children will undergo 4 weeks chemotherapy before surgery and 0 or 4 weeks chemotherapy after surgery. Duration of stay in hospital after surgery is one week. Typical chemotherapy agents used include:

- Vincristine
- Actinomycin D
- 

### **Stage 1 with high risk disease, stages 2 and 3**

These children undergo preoperative chemotherapy for 4 weeks followed by surgery. Following surgery they are likely to remain in hospital for a week. Adjuvant chemotherapy begins after surgery and lasts for varying periods up to 34 weeks depending on the risk group of the tumour. The following drugs are likely to be used:

- Vincristine
- Actinomycin D
- Doxorubicin

### **Stage 4**

This group begins treatment with 6 weeks of chemotherapy prior to surgery. They will then undergo radical surgery to remove the kidney and surrounding tissue, recovering in hospital for a week or more afterwards. Further

treatment may include radiotherapy to the kidney area or abdomen and/or radiotherapy to the lungs. Chemotherapy follows usually for up to 34 weeks depending on the features of the tumour and includes:

- Vincristine
- Actinomycin D
- Doxorubicin

And it may also include:

- Carboplatin
- Etoposide
- Cyclophosphamide

Children in this group may undergo a treatment protocol that lasts up to a year.

### **Stage 5**

This group of children have a Wilms tumour in each kidney. Radical surgery to remove both kidneys would put the child on kidney dialysis. Treatment plans aim to avoid this by shrinking tumours with chemotherapy prior to surgery. In most cases one tumour is smaller than the other and part of one or both kidneys can be preserved. This group of children begin treatment with biopsy of their tumours followed by 6 weeks of chemotherapy and rescanning to plan surgery. Using this approach, some of one or both kidneys can be preserved. Children will have further chemotherapy or radiotherapy depending on the size and extent of the tumour in each kidney. Adjuvant chemotherapy may extend for up to 34 weeks.

### **Recurrence of Wilms Tumour**

Wilms tumour may recur in the abdomen or other areas of the body such as the lungs. Survival following treatment for recurrence is variable, being good in some groups and poor in others. Treatment is likely to be tailored to the site of the recurrence and take account of treatment already given for the primary tumour. For example, if no radiotherapy has been given, radiotherapy may be used. Most children will undergo a combination of radiotherapy,

surgery where possible and chemotherapy. Some children may undergo high dose chemotherapy and stem cell transplant.

- [Complications and side effects of treatment](#)
- [Care and mobility considerations](#)

**What evidence is available?**



<b>Sources of Evidence</b>	<b>Information Provided</b>	<b>Limitation of Evidence</b>
Claimant/carer	First source of information	May not be objective, does not have specialist knowledge
GPFR	The GP should be able to confirm the diagnosis and provide contact details for the paediatric oncologist managing the child's Wilms tumour.	All children will be managed at their local paediatric oncology centre. The GP will be able to confirm the diagnosis but may not be able to give details of current or planned treatment or prognosis of disabling effects.

<p>Hospital FR</p>	<p>The best source of information will be the hospital based specialist nurse or the consultant oncologist. They will be able to provide details of stage of the Wilms tumour and the current and planned treatment and confirm disabling effects. They are the best source of information on duration of planned treatment and prognosis of disabling effects.</p> <p>The most important information to check will be:</p> <p>Confirm diagnosis.</p> <p>Confirm treatment is being received – e.g. some type of chemotherapy – planned duration of chemotherapy.</p> <p>Confirm severe disability due to specific side effects of therapy if claimed e.g. rare disabling side effects of chemotherapy. Always ask for prognosis of side effects.</p>	<p>Details of planned treatment may change depending on response to treatment or the development of side effects, information provided may become quickly out of date.</p>
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**How long will the needs last?**

Full recovery is expected in the majority of children, even those who develop recurrent disease. Enduring side effects of treatment may persist after treatment or occur later in life, these are listed in the treatment section.

Care needs related to treatment and its side effects are expected, the expected duration of planned treatment is key. This information is always available after surgical treatment is completed.

If needs are identified first awards are recommended for one year.

#### Recurrence of Wilms Tumour

Age at date of claim	Award Period
1 - 15	Award for 3 years

Wilms tumour may recur within 12 months of original treatment or years later. In all cases of tumour recurrence 3 year awards are recommended.

Care needs will continue indefinitely in children with further recurrence and terminal illness.

Note: The majority of children will be well on renewal with no care needs.

#### **Special Rules**

Consideration of entitlement under the Special Rules provisions is a separate consideration.

You must take all information into account when considering the duration of care and mobility needs. You must base the duration of care and mobility needs on the particular circumstances of the child.

#### **Care and mobility considerations**

##### **Care**

All children undergoing treatment for Wilms tumour will at least undergo radical surgery and chemotherapy and develop care needs during their chemotherapy treatment. Duration of treatment will be 'key' to needs assessment.

Older children undergoing treatment may have more care needs than younger children. Children aged 5 and above are likely to be substantially self caring – any help with feeding, toileting and mobility in this age group is likely to be related to their condition rather than immaturity. Children may be immunosuppressed and unable to attend school on a regular basis, requiring care and protection from infection at home. Preschool children who normally attend nursery may need to be cared for in a safe environment rather

than nursery to reduce risk of infection. Some children may still attend but reduce their hours.

Children of all ages are likely to require extra emotional support and practical help from their parents related to both treatment and the disease. In relation to treatment, parents will need to spend time doing the following for any child undergoing chemotherapy treatment:

- Supporting their child through painful or distressing treatment
- Ensuring any oral drug treatments are taken as prescribed, despite side effects
- Care of central line, if used (see [Equipment used \(access\) for giving chemotherapy](#))
- Monitoring their child for the side effects of treatment - this includes monitoring for signs of:
  - infection
  - easy bruising
  - bleeding, and
  - anaemia
- Protecting their child from infection during periods of immunosuppression and monitoring for signs of infection
- Encouraging their child to eat during periods of stomatitis or mucositis (sore dry mouth) and providing mouth care
- Providing an appropriate diet for the child when immunosuppressed, some children will require enteral feeding (tube feeding), some children develop food fads and food aversion, some children require dietary supplements
- Emotionally supporting their child through their illness e.g. dealing with hair loss/time away from school/ being different to peers
- Episodes of severe fatigue may endure for many months related to chemotherapy treatment and anaemia. Younger children will require help with all aspects of self care and dressing because of their age. Older children may also require such help due to severe fatigue

Some children will require additional care for:

- Peripheral neuropathy related to chemotherapy – numbness or tingling may make using the hands difficult due to numbness.

This is rare. May affect personal care, toileting ability, feeding and dressing. May affect walking

- A small number of children will develop end stage renal failure and may need kidney dialysis in addition to their anti-cancer treatment. Kidney transplant is common after Wilms tumour treatment and is usually delayed for 1 to 2 years after successful treatment
- Terminal illness
- 

## **Mobility**

Severe fatigue related to chemotherapy treatment and anaemia may affect walking for periods during treatment but this is not a continual effect over 6 months or more.

A small number of children may experience side effects of chemotherapy that can affect walking and these include numbness in the feet affecting balance (risk of falls) and hand foot syndrome. Full recovery is expected in these cases. The best source of evidence on mobility problems and ability to recover is the treating oncologist or specialist nurse.

## **Complications and side effects of treatment**

### **Immediate side effects of surgery**

The immediate effects include:

- bleeding
- damage to blood vessels, the spleen or the intestines during surgery, and
- small bowel obstruction

These complications are likely to lead to a substantial increase in the duration of the hospital stay following surgery but unlikely to lead to enduring care or mobility needs.

### **Short term side effects of chemotherapy**

The effects of chemotherapy and the care needs that arise from ensuring the child has their chemotherapy treatment and copes with the side effects are the most likely reason for care needs to develop. Duration of care needs related to chemotherapy will relate to duration of treatment, which may extend from several weeks to almost 1 year. The immediate side effects of

chemotherapy occur during treatment and can be expected to resolve within weeks of treatment finishing. These include:

- Nausea and vomiting
- Diarrhoea
- Hair loss
- Increased risk of infection
- Bruising and bleeding
- Extreme tiredness
- Mucositis
- Reduced nutritional intake
- Rarely peripheral neuropathy and difficulty walking

### [Long term disabling effects of chemotherapy treatment](#)

#### **Long term complications of Wilms tumour treatment**

##### Renal failure and dialysis

End stage renal failure occurs when the kidneys can no longer deal with waste products produced by the body and dialysis is necessary for ongoing survival. Only 1% of children with Wilms tumour develop end stage renal failure and of these 70% had bilateral tumours. Renal failure may occur at the time of treatment because of the removal of kidney tissue or later on due to damage to remaining kidney tissue by radiotherapy. In 99% of children treated for Wilms tumour the remaining kidney compensates for the kidney that was removed.

##### Heart Failure

Chemotherapy drugs, particularly doxorubicin, damage heart muscle cells. 25% of children receiving doxorubicin will have some evidence of heart muscle damage on testing. The overall incidence of heart failure after treatment for Wilms tumour is 1.7%. This rate rises to 5.4% in children who also received radiotherapy to the chest area.

##### Lung function

Lung function be affected by radiotherapy to the lungs and may cause breathlessness on exertion. Only a small number of children will have

radiotherapy to the lungs so this is a rare problem following treatment for Wilms tumour.

#### Hepatic Veno-occlusive disease

Liver damage caused by radiotherapy and chemotherapy drugs, particularly Actinomycin D, may cause jaundice, ascites, enlarged liver and weight gain. Around 8% are affected and recovery is expected.

#### Hormonal insufficiency and infertility

Radiotherapy may affect the testes or more rarely the ovaries, puberty may not progress normally and sex hormone treatment may be required.

#### Second cancers

The risk of these is increased as with all childhood cancers and in Wilms tumour is 1.6% at 15 years follow up.

[Problems in adults who had cancer treatment as children](#)

#### **Equipment Used (Access) for giving Chemotherapy**

Name	Access	Area of the body
Hickman Line	Venous	This type of line is tunnelled under the skin of the chest wall and inserted into the large veins in the neck. The end of the line is outside the skin about half way down the front of the chest.
PICC (peripherally inserted central catheter)	Venous	This is a long line inserted into a vein in the arm and threaded up to the large veins in the neck. The end of the line is outside the skin near the crook of the elbow.
Portacath/implantable port/port	Venous	This is also inserted into the neck veins but the end of the line is under the skin, usually of the chest wall. A small lump will be palpable under the skin and this is the chamber into which, drugs can be injected.
Peritoneal port	peritoneal	These catheters are usually inserted surgically and have ports underneath the skin of the abdomen.
Omayya reservoir	Spinal fluid	The reservoir is a chamber placed under the skin of the scalp, this communicates via a tube with the spinal fluid. This avoids repeated spinal tap procedures.

### **Problems in adults who had cancer treatment as children**

People in this category have much longer to develop the long term or enduring side effects of chemo and radiotherapy. The oldest members of this group will have had their treatment in the 1970s. What will happen to them as they age is unknown. Some childhood survivors have already developed significant enduring problems because of their treatment, either during



treatment or some years later. The number of adults 'at risk' in this category is set to rise.

Cancer therapy, in particular chemotherapy made great progress in the 1980s and 1990s and for the first time high rates of cure were achieved in some of the common childhood malignancies such as leukaemia and lymphoma. Over time treatment has been modified to become as effective as possible with as few side effects as possible. Significant long term side effects of treatment given in the past are increasingly being recognised. These side effects generally occur because of changes in normal tissue caused by the treatment, these changes take many years to cause symptoms or become apparent. The medical profession is still in the early days of recognising and researching these disorders.

Over time more members of this group can expect to either develop these problems or have them recognised. A breakdown of problems is provided by treatment. Effects tend to be greater when treatment of cancer began at a young age (under 3) and when large doses of chemotherapy and

radiotherapy were necessary. Common cancers in children include leukaemia, lymphoma, brain tumours, bone and soft tissue sarcomas.

Type of cancer treatment	Disabling effect
Cranial irradiation esp. if combined with intrathecal chemotherapy	Neurocognitive defects – reduced IQ, attention deficit, poorer motor/verbal skills, may be severe enough for a Statement of Education Needs (SEN), deafness, epilepsy.
Cranial irradiation, effects are worse if this was combined with abdominal radiation	Hormonal effects including growth impairment in childhood, hypothyroidism, increased risk of infertility, early menopause.
Cranial irradiation	Obesity and its disabling effects.
Chemotherapy or radiotherapy	Increased risk of infertility.
Chemotherapy esp. anthracycline doxorubicin	Heart problems including heart failure, myocardial infarction, arrhythmias and sudden death at young age.
Radiotherapy to chest	Lung problems – breathlessness.
Steroids, methotrexate, inactivity due to illness	Osteopaenia/Osteoporosis.
Chemotherapy or radiotherapy	Second cancers especially brain tumour.
Radiotherapy to abdomen (bladder/bowel/liver)	Chronic diarrhoea, malabsorption, bladder problems, kidney problems including rarely kidney failure.

### Long term disabling effects of chemotherapy treatment

Peripheral neuropathy (neuritis, plexopathy) - is a toxic effect on nerves, which prevents nerves from working properly.

- Motor nerves – these control movement of muscles, damage may lead to clumsiness or in severe cases paralysis of muscles supplied by the affected nerves. Some recovery may

occur once treatment is stopped but the changes are usually slow to improve or permanent

- Sensory nerves – these nerves enable the sensation of touch, damage to them results in numb areas or areas of pins and needles. In some cases pain fibres are affected and this can lead to pain syndromes

Abnormal sensations can make simple activities like making a cup of tea, fastening a button or walking difficult, impossible or painful and distressing.

- Breathlessness - has many causes including lung problems and heart failure, which may have been caused by the

chemotherapy or radiotherapy. Lung damage or heart problems caused by certain drugs may be irreversible or progressive

- Leukaemia - is more common in children who have had chemotherapy or radiotherapy treatment
- Infertility – chemotherapy and radiotherapy can cause infertility

These side effects are rare.

## **Children's Sources of Evidence**

A - G
<a href="#">Audiological Reports</a>
<a href="#">Bayley Scales of Infant Development</a>
<a href="#">The Common Assessment Framework (CAF)</a>
<a href="#">Educational Psychology Service</a>
<a href="#">General Practitioner (GP) Report</a>
H
<a href="#">Health Care Professional Reports</a>
<a href="#">Health Care Professional (HCP) Examination Reports</a>
<a href="#">Hospital Report</a>
I
<a href="#">Individual Education Plan</a>
<a href="#">IQ Percentiles &amp; the Wechsler Scale</a>
K
<a href="#">Kaufman Assessment Battery for Children</a>
M
<a href="#">Medical Services Doctor</a>
N
<a href="#">National Curriculum Key Stages</a>
<a href="#">Note in Lieu</a>
O
<a href="#">Occupational Therapist</a>
P
<a href="#">Pastoral Support Programme</a>

<a href="#">Physiotherapist</a>
<a href="#">Portage Report</a>
<a href="#">Portage Visitor</a>
S
<a href="#">School Action and School Action plus</a>
<a href="#">Social Worker</a>
<a href="#">Special Educational Needs</a>
<a href="#">Specialist Nurse</a>
<a href="#">Stanford-Binet Intelligence Scale</a>
T
<a href="#">Transition (14+) Plan / Review</a>
W
<a href="#">Wechsler Pre-school and Primary Scale of Intelligence</a>

## **Audiological Reports**

An Audiological report (audiogram) is a technical assessment to establish the extent of a customer's deafness.

An audiogram demonstrates the level of hearing loss at different frequencies and shows whether the hearing loss is due to a conductive,

sensorineural or mixed cause. The level of disability in the CCM guidance is based on audiograms carried out without hearing aids being worn by the customer.

An audiological report can be used as further evidence when a customer claims the DLA higher rate mobility component because of a combination of hearing loss and blindness (deaf / blind deeming provisions).

Decision Makers can request an Audiological report but only when the customer has not had an Audiogram in the last six months.

### **The Bayley Scales of Infant Development**

The Bayley Scales of Infant Development (BSID) have been used extensively worldwide to assess the development of infants. The test is given on an individual basis and takes 45-60 minutes to complete. It is administered by examiners specifically trained in BSID test procedures. The examiner presents a series of test materials to the child and observes the child's responses and behaviours. The test contains items designed to identify young children at risk for developmental delay. BSID evaluates individuals along three scales:

- **Mental scale:** This part of the evaluation, which yields a score called the mental development index, evaluates several types of abilities: sensory/perceptual acuities, discriminations, and response; acquisition of object constancy; memory learning and problem solving; vocalisation and beginning of verbal communication; basis of abstract thinking; habituation; mental mapping; complex language; and mathematical concept formation
- **Motor scale:** This part of the BSID assesses the degree of body control, large muscle coordination, finer manipulative skills of the hands and fingers, dynamic movement, postural imitation, and the ability to recognise objects by sense of touch (stereognosis)
- **Behaviour rating scale:** This scale provides information that can be used to supplement information gained from the mental and motor scales. This 30 item scale rates the child's relevant

behaviours and measures attention/arousal, orientation/engagement, emotional regulation and motor quality

The BSID are known to have high reliability and validity. The mental and motor scales have high correlation coefficients (.83 and .77 respectively) for test-retest reliability.

## **The Common Assessment Framework (CAF) Form**

### **What is the CAF form?**

The CAF is a shared assessment and planning framework for use across all children's services and all local areas in England. It aims to help early identification of children and young people's additional needs and promote co-ordinated service provision to meet them. The Common Assessment Framework is not in use in other parts of the United Kingdom.

### **What does the CAF form consist of?**

- A pre-assessment checklist to help decide who would benefit from a common assessment.
- A process to enable experts in the children and young people's workforce to undertake a common assessment and then act on the result.
- A standard form to record the assessment
- A delivery plan and review form

### **Who is the CAF for?**

The CAF is aimed at children and young people with additional needs who have needs that are not being met by their current service provision.

### **Who will use the CAF form?**

Staff working in health, education, early years and child care.

See - [Common Assessment Framework form](#)

## **The Educational Psychology Service**

### **What is an Educational Psychologist?**

Educational Psychologists (EPs) provide guidance and support to schools on a range of issues including Special Educational Needs (SEN). They work



with parents, carers and teachers to identify and support the SEN of individual children at home and at school.

All EPs have qualifications in Psychology and Educational Psychology and are not usually medically qualified. They may also be qualified teachers with teaching experience.

### **What does their work include?**

EPs help children from Nursery school to age 19. They assess how best to help by observing and working with children at home and at school. The EP works closely with parents and school staff to plan a programme of support.

Usually the Special Educational Needs Coordinator (SENCO) at school will contact an EP after deciding the child might benefit from their help. The school then contacts parents for permission to provide the specialist support.

Parents / carers, the school and the EP review the plan to make sure the child's needs are being met.

### **General Practitioner (GP) Reports**

GPs are the first point of contact for people with a health condition and provide ongoing care to their patients. They are responsible for diagnosing and treating illnesses and may refer the patient to hospital for specialised care, in which case a report is usually sent back to the GP. GPs will have long term knowledge of the patient and hold the patient's records including hospital correspondence. GPs generally are not trained and do not assess a patient's function and are therefore not usually in a position to give information about their patients' ability to perform daily living activities.

It is reasonable to expect a GP's report to contain factual information such as diagnosis, history of the condition, clinical findings, results of special tests, medication and treatment plan.

The GP may have very limited information on people who have had stable, long-term disabilities, often since childhood, for example children and adults with learning disabilities. Claimants with mental health problems or learning disabilities may be mainly under the care of community psychiatric services or other specialist teams and it is often more appropriate to make an initial request for a factual report from another agency or professional.

### **Health Care Professional (HCP) Examination Reports**

A HCP is a practitioner involved with patient care. This would include doctors, nurses, occupational therapists and physiotherapists. They will have received training in disability assessment medicine. In addition they are

instructed in the assessment of care and mobility needs relating to the entitlement to DLA/AA/PIP.

A HCP examination report will provide details of the diagnosis (where known), brief history of the condition, treatment, a record of clinical examination, severity and likely disabling effects of the condition on day-to-day living.

The HCP examination report is able to provide a critical appraisal of whether a person's claimed care and mobility needs are reasonable in the light of the disabling condition(s).

A HCP examination report can be particularly helpful in situations where the existing medical evidence (usually factual reports) appear to be contradictory, or where factual reports do not appear to cover areas relevant to entitlement.

When the Case Manager decides to request a HCP examination report, it is worth considering the exact nature of the information required and to formulate a submission with some specific questions for the HCP. It may also be useful to enclose copies of factual reports, especially if existing evidence is contradictory.

### **Health Care Professional**

The term "health care professional" means an individual who has had specialised training to provide health care items or services, treatment, assistance with activities of daily living or medications to patients.

### **Hospital Reports**

National Health Service hospitals and trusts are required by the NHS Executive to provide factual reports incidental to the claimant's treatment when requested by the Benefits Agency. The hospitals or their employees are not paid on an individual basis for the reports. It is the responsibility of the hospital or trust management to ensure that the information is supplied.

Although patients are under the care of a designated consultant, this doctor does not necessarily have to complete the report. An alternative member of

the medical team, another consultant or other healthcare professional may carry out the task.

The hospital report will contain information obtained from the medical records and the professional's knowledge of the patient. It will consist of detail relating to diagnosis, special tests, clinical findings, medication etc.

A hospital doctor will not have the background knowledge to answer specific questions relating to the criteria for benefit entitlement.

Hospitals are likely to be the most appropriate source of information when the medical diagnosis is uncommon or treatment is very specialised.

### **Individual Education Plan**

The child's teacher is responsible for working with the child on a day to day basis, but may decide to write down the actions of help in an IEP. The IEP

will include details of the conditions giving rise to the child's needs and could also include:

- what special or additional help is being given
- who will provide the help and how often
- what help the child could be given at home
- the child's targets
- how and when progress will be checked

Other names for an IEP, which may also be used, are:

- Individual Behaviour Plans (IBP)
- Individual Behaviour Management Plans (IBMP)
- Behaviour Support Plan (BSP), and
- Behaviour Target Report (BTR)

Formal reviews of the plan should be held at least twice a year.

<b>Name</b>		<b>Stage</b>	
<b>Area/s of concern</b>		<b>Yr group IEP No</b>	
<b>Class Teacher</b>		<b>Start date</b>	
<b>Supported by</b>		<b>Review date</b>	
<b>Proposed support</b>		<b>Support began</b>	
Targets to be achieved			
Achievement criteria			
Possible Resources and Techniques			
Possible strategies to use in class			
Ideas for support teacher / assistant			
Parents / carers need to			
Student needs to			

### **IQ percentiles and the Wechsler Intelligence Scale for Children (WISC)**

The Wechsler Intelligence Scale for Children (WISC) is an intelligence test for children between age 6-16 that can be completed without reading or writing. Tests include Vocabulary – straightforward questions over meaning of words, Matrix reasoning – children are shown an array of pictures with

one missing square and select the picture that fits from 5 options and Letter / Number sequencing. The WISC generates an IQ score.

The IQ percentile table below illustrates the IQ score and the corresponding percentile rank. A percentile rank shows the percentage of people that scored above and below a certain score. For example, the 50<sup>th</sup> percentile means 49 out of 100 per cent of the people who took the test scored higher scores and 49 got lower scores. The 5<sup>th</sup> percentile means 75 out of 100 scored higher and 25 scored lower.

The IQ scale only goes as low as 45. A percentile score of less than 1 equates to an IQ score of 65.

<b>IQ Score</b>	<b>Percentile rank</b>
100 – average IQ	50th
95	37th
90	25th
85	16th
80	9th
75	5th
70	2nd
65	1st
60	0.38
55	0.13
50	0.043

The WISC is used both as an intelligence test and a clinical tool. It can be used to show discrepancies between a child's intelligence and their

performance at school and it is this discrepancy that Educational Psychologists look for when using this test.

### **Kaufman Assessment Battery for Children**

The Kaufman Assessment Battery for Children (KABC) is a standardised test that assesses intelligence and achievement in children aged 2½ years to 12½ years.

#### **Purpose**

The KABC was developed to evaluate preschoolers, minority groups, and children with learning disabilities. It is used to provide educational planning and placement, neurological assessment, and research. The assessment is to be administered in a school or clinical setting and is intended for use with English speaking, bilingual, or non-verbal children.

#### **Precautions**

The KABC is especially useful in providing information about nonverbal intellectual abilities. However, it has been criticised for not focusing on measures of verbal intelligence in the Mental Processing Composite score, which measures intelligence. Additionally, the separation of intelligence and achievement scores has been questioned by researchers who claim the two terms are misleading. For example, many subtests in the achievement composite are in fact measures of intelligence rather than achievement (knowledge acquired through school and/or home environment). The KABC

should be used with caution as the primary instrument for identifying the intellectual abilities of children.

Administration and interpretation of results (as with all psychometric testing) requires a competent examiner who is trained in psychology and individual intellectual assessment - preferably a psychologist.

### **Description**

Administration of the KABC takes between 35 and 85 minutes. The older the child, the longer the test generally takes to administer. It is comprised of four global test scores that include:

- sequential processing scales
- simultaneous processing scales
- achievement scales
- mental processing composite

There is an additional non-verbal scale that allows applicable subtests to be administered through gestures to hearing impaired, speech/language impaired, or children who do not speak English.

The test consists of 16 subtests -10 mental processing subtests and six achievement subtests. Not all subtests are administered to each age group, and only three subtests are administered to all age groups. Children ages two years, six months are given seven subtests, and the number of subtests given increase with the child's age. For any one child, a maximum of 13 subtests are administered. Children from age 7 years to 12½ years are given 13 subtests.

The sequential processing scale primarily measures short-term memory and consists of subtests that measure problem-solving skills where the emphasis is on following a sequence or order. The child solves tasks by arranging items in serial or sequential order including reproducing hand taps on a table, and recalling numbers that were presented. It also contains a subtest



that measures a child's ability to recall objects in correct order as presented by the examiner.

The simultaneous processing scale examines problem-solving skills that involve several processes at once. The seven subtests comprising this scale are:

- facial recognition
- identification of objects or scenes in a partially completed picture
- reproduction of a presented design by using rubber triangles
- selecting a picture that completes or is similar to another picture
- memory for location of pictures presented on a page, and
- arrangement of pictures in meaningful order

The achievement scales measure achievement and focus on applied skills and facts that were learned through the school or home environment. The subtests are:

- expressive vocabulary
- ability to name fictional characters, famous persons, and well-known places
- mathematical skills
- ability to solve riddles
- reading and decoding skills; and
- reading and comprehension skills

The sequential and simultaneous processing scales are combined to comprise the mental processing composite. This composite measures intelligence on the KABC and concentrates on the child's ability to solve unfamiliar problems simultaneously and sequentially. The simultaneous processing scales have a greater impact on the mental processing composite score than do the sequential processing scales. The mental processing composite score is considered the global estimate of a child's level of intellectual functioning.

## **Results**

The global scales on the KABC each have a mean or average score of 100 and a standard deviation of 15. For this test, as with most measures of intelligence, a score of 100 is in the normal or average range. The standard

deviation indicates how far above or below the norm a child's score is. For example, a score of 85 is one standard deviation below the norm score of 100.

Test scores provide an estimate of the level at which a child is functioning based on a combination of many different subtests or measures of skills. A trained psychologist is needed to evaluate and interpret the results, determine strengths and weaknesses, and make overall recommendations based on the findings and behavioural observations.

### **Medical Services Doctors**

The doctors employed by Medical Services are particularly skilled and knowledgeable in the field of medical disability analysis. Most carry out regular examinations for benefit including the Personal Capability Assessment, Severe Disablement Allowance, Industrial Injury Disablement Benefit and DLA/AA. They also train contracted doctors to carry out the above types of examination and monitor the reports of these doctors, providing further training if needed.

Advice is given to Decision Makers on request in all the benefit strands.

Medical Service doctors also liaise with general practitioners, hospital doctors and other external bodies with regard to individual claims or other benefit matters.

Their experience in all strands of the medical benefits enhances their expertise in interpreting the effects of functional impairment across a wide range of disability.

Medical Services doctors can advise Decision Makers in a number of ways:

- They can identify who is likely to be the best person to approach for a medical report, whether a doctor or other healthcare professional and provide advice on the basis of the reply
- They can explain and interpret medical terminology in all types of medical reports. This can include the nature of diagnoses,

the use of medication, the meaning of clinical examination findings and the likely disabling effects of any given condition

- They will be able to interpret clinical information, analyse it and describe the extent of care or mobility restrictions likely to arise from a specific medical or disabling condition
- They can identify inconsistencies or contradictions in the evidence and advise if further evidence is likely to be useful
- They can give advice regarding response to treatment and prognosis

## **National Curriculum Key Stages**

### **England and Wales**

Age	Stage	Year	Assessment
3-4	Early years foundation stage		
4-5		Reception	
5-6	Key Stage 1	Yr 1	
6-7		Yr 2	<p>Teacher assessment in English, Maths &amp; Science</p> <p>Expected to work at levels 1-3 in key stage 1.</p> <p>A child age 7 is expected to achieve level 2</p>
7-8	Key Stage 2	Yr 3	
8-9		Yr 4	
9-10		Yr 5	
10-11		Yr 6	<p>National Tests &amp; Teacher assessments in English, Maths &amp; Science.</p> <p>Expected to work at levels 2-5 in key stage 2.</p> <p>A child age 11 is expected to achieve level 4.</p>
<p>* A child working at level 1 or 2 at age 11 is not formally tested – given teacher assessment only.</p>			
11-12	Key Stage 3	Yr 7	
12-13		Yr 8	

13-14		Yr 9	Teacher assessment in English, Maths & Science and other foundation subjects.  Expected to work at levels 3-7 in key stage 3.  A child age 14 is expected to achieve level 5/6.
14-15	Key Stage 4	Yr 10	Some children take GCSE's.
15-16		Yr 11	Most children take GCSE's or other national qualifications.
<p>* Attainment levels for all subjects range between 1- 7 (1 being the lowest). High performing children may be awarded level 8 or exceptional performance.</p> <p>If children are working way above or below the expected level, the school must provide extra support.</p>			

### Scotland – Age 5-14 Curriculum

Refers to pupils in Primary school (years P1-P7) and years 1 & 2 of Secondary education (years S1-S2).

Level A should be attained in the course of P1-P3 by almost all pupils.

Level B should be attainable by some pupils in P3 or even earlier, but certainly by most in P4.

Level C should be attainable in the course of P4-P6 by most pupils.

Level D should be attainable by some pupils in P5-P6 or even earlier, but certainly by most in P7.

Level E should be attainable by some pupils in P7/S1, but certainly by most in S2.

The Scottish Curriculum is currently going through a national review. Curriculum for Excellence is the Scottish curriculum review programme which aims to produce a streamlined educational experience for pupils from 3-18 years old.

### **P Scales**

The P scales are differentiated performance criteria and should be used with pupils from 5-16 years of age. . They are relevant to pupils with learning difficulties and outline attainment for pupils working below level 1 of the national curriculum.

There are P scales for each subject in the national curriculum and for religious education. The P scales use 8 performance levels to illustrate the learning that leads to national curriculum level 1.

- Levels P1 to P3 show the earliest levels of general attainment with subject-focused examples.
- Levels P4 to P8 show subject related attainment.

The levels in the P scales operate independently of chronological age. Teachers should not expect pupils to reach any given level at a particular age or to progress through the levels at a pre determined rate. However, the

P scale descriptors are designed to identify the kinds of progress a pupil may make over a year or key stage.

### **Note in Lieu**

A note in lieu is issued if, after a statutory assessment, the LEA decides that the needs can be met with the support being provided by the child's school.

A Note in Lieu describes -:

- the child's special educational needs
- gives the reasons why a Statement has not been written
- provides suggestions for the school about how to meet the child's needs
- describes any non-educational needs the child has and the support which might be helpful

Copies of all the advice and reports received as part of the statutory assessment will be sent along with the Note in Lieu to parents / carers.

The Note in Lieu will be used by the school to help them provide the most appropriate support for the child. The note contains advice which has been based on the assessment reports, but it is not a legally binding document.

Explanation – in lieu means 'instead of' - in this case instead of a SEN.

### **Occupational Therapists (OTs)**

The prime function of OTs is to maximise the independence of the patients through assessing their needs and prompting the restoration of the maximum use of function. As specialists in the provision of disability equipment they offer practical advice on adapting the environment to the needs of the individual.

OTs will be found working in many areas of health care; learning disability, mental illness, geriatrics, physical disability, paediatrics, day care and

rehabilitation centres. They work in a variety of settings including hospitals, day centres, claimant's own homes and residential accommodation.

Their work focuses on enabling people to perform every day activities such as washing, dressing, cooking and shopping. They help people to readjust to independent living after discharge from hospital.

Where OTs work with people with mental health disorders or learning disabilities, they prepare these patients to return to as full a life as possible in the community. This may involve psychology or behavioural programmes including group work, for example, desensitisation programmes for agoraphobia or social skills training for people with long-term mental disorders.

### **Reports from Occupational Therapists**

Reports from occupational therapists can provide very useful information since they link disability to need.

Sometimes OTs can prove difficult to identify as a source of information, especially if the patient is only seen on one or two occasions before discharge or in their own home.

If an elderly person is assessed at home a copy of the report may be available in his or her general practice records.

Claimants themselves may also have a copy of their assessment report.

### **Pastoral Support Programme**

The Pastoral Support Programme (PSP) is a school based and co-ordinated intervention to help individual pupils to improve their social, emotional and behaviour skills. The PSP will act as a preventative measure for those pupils at risk of being excluded.

If the pupil has another plan then ideally the PSP should be integrated. Rather than set up a separate PSP for pupils with an IEP (Individual Education Plan), schools should ensure IEP's for pupils at serious risk of being



excluded reflect appropriate approaches, strategies and support to meet their additional needs.

A PSP should be set up for a pupil:

- Who has several fixed exclusions that may be leading to a permanent exclusion
- Who is identified as being at risk of failure at school through the feeling of being alienated from other people
- Where the situation is complex and a range of agencies are needed to support children and young people

The PSP should:

- use information gathered from a range of sources including, the child, parents and carers, school and other relevant professionals
- set out specific and realistic targets, and how they will be measured, agreed by all involved, including the child, and broken down into manageable chunks
- identify the input and support from the school, parents and carers, the child will receive to help them reach the agreed targets
- identify the input and support from all other relevant professionals and agencies the child will receive to help them reach the agreed targets
- identify the recognition and rewards the child will receive when they demonstrate efforts to meet the agreed targets
- identify the consequences if the child does not demonstrate efforts to meet the agreed targets including any sanctions that may be applied
- identify how long the PSP will last and when it will be reviewed. Any PSP should be given appropriate time to ensure the pupil has opportunity to demonstrate efforts to improve. A PSP should have a time limit, for example, 16 working weeks.

During this time progress should be regularly monitored and adjustments made to the PSP as necessary

<b>Name of Pupil</b>		<b>Date of Birth</b>	
<b>Address</b>		<b>Tel No</b>	
<b>Ethnicity</b>		<b>Post Code</b>	
<b>Purpose of Pastoral Support programme</b> <ol style="list-style-type: none"> <li>To prevent permanent exclusion and/or improve attendance.</li> <li>To help.....better manage his/her behaviour/attendance and to identify precise and realistic behaviour outcomes for him/her to work towards.</li> <li>To improve achievements</li> </ol>			
<b>Specific areas of concern</b>			
1.			
2.			
3.			
<b>Pupil's views</b>		<b>Carer's views</b>	
<b>Other involved agency views (e.g. Governors, Discipline Committee, Social Worker etc)</b>			
<b>Agency</b>			
1.			
2.			
3.			
<b>Support Plan</b>			
<b>Task</b>	<b>Target Date</b>	<b>By Whom</b>	

1.			
2.			
3.			
Rewards ( if applicable )			
Contingency Plan ( Intervention if circumstances demand )			
1.			
2.			
Date/Time Review meeting approx. 6 weeks		Venue	

## Physiotherapists

A physiotherapist has expertise in preventing and treating diseases/disabilities, particularly in relation to the neuromuscular, musculoskeletal, cardiovascular and respiratory systems.

Physiotherapists have a major contribution to make in restoring independence after a traumatic incident such as a stroke or head injury, after major surgery such as amputation, joint replacement or organ removal.

They also provide treatment for muscle, ligament, or tendon injuries, spinal conditions and rehabilitation after fractures.

When a physiotherapist treats a person for a minor illness/injury e.g. ankle injury or mechanical low back pain, the course of treatment is relatively short-lived involving two to six sessions over a similar number of weeks. Treatment after a major surgery e.g. joint replacement or amputation will be more prolonged but will be finite in length.

A few long-term disabilities may require continuing treatment on a regular basis, but this is not frequently the case. In these situations the patient may be able to carry out the exercises themselves or their carer is taught to do the procedure e.g. chest physiotherapy in cystic fibrosis. Many children with physical disabilities find their needs change as they grow. It is therefore

common for them to remain under the care of a physiotherapist, with regular reviews, until their growth is largely complete.

Review of posture, pattern of walking and mobility in general is very much part of the role, together with pain-reducing and muscle-strengthening techniques.

Physiotherapists assess patients for and can prescribe appropriate aids. These include walking sticks, crutches, frames and wheelchairs. Other equipment might include TENS machine for pain relief, incontinence aids, splints, neck collars and callipers.

### **Reports from Physiotherapists**

A report from a physiotherapist could be expected to contain a brief medical history, diagnosis of the disabling condition and details of clinical examination and treatment plan.

Information about limb function and ability to walk with appropriate appliances will often be available.

In milder conditions information will relate to a specific episode of treatment relating to the affected area of the body, and the report is unlikely to cover the disabling effects of other generalised conditions.

### **Portage**

Portage is a home-visiting educational service for pre-school children with special or additional needs and their families.

Portage assesses the needs of young children with delayed development and then, in partnership with parents, builds on the abilities the child already has to stimulate the development of new skills, by breaking them down into quickly achievable steps.

Portage originated in the late 60's in a town called Portage in Wisconsin, USA. It was set up as a home based early intervention model for families who had pre-school children who were either delayed in their development

or had been diagnosed with a medical condition which were known to affect development.

How does it work?

Initially the child's development is observed by a Portage visitor and recorded in the following areas -:

- Social skills
- Self help
- Motor skills
- Cognitive skills
  
- Language skills
- Infant stimulation

Looking at the skills a child has acquired, plus emerging skills he/she is beginning to demonstrate, the portage visitor and parents decide on teaching objectives. Each skill is broken down into small steps via a series of linked activities to stimulate the child's development. Portage programmes are specifically designed to meet the needs of the individual child and to enable him/her to learn as effectively as other children. Portage emphasises the positive and builds on what children can already do.

### **More Information about a Portage Visitor**

A portage home visitor (or portage worker), provides a home-visiting service for pre-school children with developmental or learning difficulties, physical disabilities or other special needs. They help parents to encourage their

children's development by suggesting activities and daily routines to make learning fun.

Their work includes:

- observing the child and talking to the parents to identify the skills the child already has
- deciding with the parents which skills are most important for the child's future learning
- suggesting a programme of activities for the parents and child to practise together
- breaking down tasks that are difficult or take a long time into small steps
- providing an activity chart or notebook for parents to record their child's progress
- visiting each week to check on progress and agree on new goals
- writing progress reports and working with the parents to develop long-term goals

They work closely with other professionals, such as health visitors, social workers, physiotherapists and speech therapists.

A Portage Visitor must have experience of working with children under the age of five, and an understanding of child development. They would also usually need one of the following:

- a relevant professional qualification such as teaching, social work or nursing
- an early years qualification such as NVQ Level 3 in Early Years Care and Education.

### **School Action/School Action Plus**

School and early education settings place great importance on identifying Special Educational Needs (SEN) so they can help the child as early as possible. Children learn in different ways and can have different levels or

kinds of SEN. So if the child has SEN, the school will increasingly, step by step, bring in specialist expertise to help with the difficulties they may have.

The school must inform parents /carers if they start giving extra or different help to the child because of their SEN. The basic level of extra help is known as School Action and could be -:

- A different way of teaching things
- Some extra help from an adult
- Using particular equipment like a computer or specialist desk.

The equivalent in Scotland is called Additional Support Needs (ASN). The principle difference being not confined to children whose need for special attention arises from disability or learning difficulties but from a range of factors e.g. being in hospital, death of a close family member.

The child's teacher is responsible for working with the child on a day to day basis, but may decide to write down the actions of help in an Individual Education Plan. The IEP could include:

- what special or additional help is being given
- who will provide the help and how often
- what help the child could be given at home
- the child's targets
- how and when progress will be checked

Sometimes the school will not write an IEP but will record how they are meeting the child's needs in a different way, perhaps as part of the lesson plans.

If the child does not make enough progress under School Action, the teacher or SEN coordinator (SENCO) will ask for advice from other people outside the school. This could include a specialist teacher or a speech and language therapist. This kind of extra help is called School Action Plus.

The key test for taking School Action, moving to School Action Plus, or considering whether a statutory assessment is necessary is whether the child is making adequate progress. Essentially, what is considered to be adequate



progress for a particular child is a matter for the teacher's professional judgement

## **Social Workers**

The title social worker is currently used to describe a variety of people employed in the public, private and voluntary sectors. There are great differences in the level of professional training and the type of work undertaken.

The biggest employers of professionally qualified social workers are local authority Social Services Departments. The social workers may be based in fieldwork offices or in hospital. Some specialise in areas of work such as learning disabilities, mental health, physical disabilities, childcare and hearing and sight impairment.

Social workers carry out assessments on disabled people to determine the level of care needed at home or the requirement for day care or residential services. These assessments will include information about the client's ability to do day-to-day tasks and to perform personal care. The social workers may work closely with occupational therapists that are employed on the social work team by the same local authority.

Residential social workers and day care workers are employed in residential settings and day care centres. A care co-ordinator may have specific responsibility for a particular client. Many are very experienced in their field of care but are not professionally qualified. They usually have detailed first hand knowledge of a client's disability and day-to-day functioning.

## **Reports from Social Workers**

In those clients with disabilities who have undergone a formal assessment to determine the level of help which they need in their own homes or other residential settings the social worker can provide useful knowledge in determining benefit entitlement.

Some of the information will relate to activities such as the ability to shop or clean the home, which are not relevant to entitlement. In those with more severe disabilities evidence pertaining to personal care may be available.

Residential social workers will be able to give detailed information about care, including at night, similar to that provided by other relatives or carers living in the home.

## **Special Educational Needs (SEN)**

School & early education settings place great importance on identifying SEN so they can help the child as early as possible. Children learn in different ways & can have different levels or kinds of SEN. So if a child has SEN,

the school will increasingly, step by step, bring in specialist expertise to help with the difficulties they may have.

The school must inform parents /carers if they start giving extra or different help to the child because of their SEN. The basic level of extra help is known as School Action & could be:

- A different way of teaching things
- Some extra help from an adult
- Using particular equipment like a computer or specialist desk

The equivalent in Scotland is called Additional Support Needs (ASN). The principle difference being not confined to children whose need for special attention arises from disability or learning difficulties but from a range of factors e.g. being in hospital, death of a close family member.

### **Individual Education Plans (IEP)**

The child's teacher is responsible for working with the child on a day to day basis, but may decide to write down the actions of help in an IEP. The IEP could include:

- what special or additional help is being given
- who will provide the help & how often
- what help the child could be given at home
- the child's targets
- how & when progress will be checked

Sometimes the school will not write an IEP but will record how they are meeting the child's needs in a different way, perhaps as part of the lesson plans.

If the child does not make enough progress under School Action, the teacher or SEN coordinator (SENCO) will ask for advice from other people

outside the school. This could include a specialist teacher or a speech & language therapist. This kind of extra help is called School Action Plus.

The equivalent in Scotland is called an Individualised Educational Programme. There is no difference to the English IEP.

[Click here for more information about IEP's](#)

**Statement of Special Educational Needs (SSEN) is in the process of being replaced by Education, Health and Care Plan (EHCP).**

EHCPs began to be introduced across England and Wales during 2014. They replace the current system of Individual Healthcare Plans (IHPs) and Statements of SEN. Processing of the EHCPs has been simplified in order to confirm the child's SEN more efficiently, with data viewed by only the necessary parties. It formulates the spectrum of the child's needs, rather than focussing upon the educational limitations.

In forming an EHCP, each child must be assessed individually, even if similar cases have previously arisen. This is because every child will have different needs and react differently to the condition they are suffering, so every EHCP should be unique and heavily focused on the individual personality and needs of that child. The initial draft of an EHCP is formed amongst the school, child's parents, and school nurse. At the end of the process the

local authority (LA) has to make the decision as to whether or not to issue an EHCP.

If the LA refuses to issue an EHCP, the family has a right to appeal to the Special Educational Needs and Disabilities Tribunal.

The EHCP should detail:

Child's special educational needs and special educational provision

Healthcare provision assessed as reasonably required

Social care provision made under the Chronically Sick and Disabled Persons Act (1970), and any other social care provision assessed as reasonably required.

The LA has the legal duty to secure the educational provision specified in the EHCP, i.e., to ensure that the provision is delivered.

When SEN cannot be met by resources generally available to local schools, the assessment will identify the child's needs and any special help that is required and will be produced in the following format:

Part 1 – Child's details (e.g. name, address, parent / carer) and a list of the advice the authority received as part of the assessment

Part 2 – Gives details of the child's special educational needs

Part 3 – Describes all the special help to be given for the child's needs

Part 4 – Gives the type & name of the school the child should go to & how any arrangements will be made out of school hours or off school premises

Part 5 – Describes any non-educational needs the child has

Part 6 – Describes how the child will get help to meet any non-educational needs.

A child's statement of Special Educational Needs (SEN) should include mandatory information which Local Authorities (LAs) must request & consider when producing a statement of SEN. In addition to the actual statement, which will set out emotional & behavioural problems which may impact on learning, as well as other conditions & health problems, parents will also have the reports considered to assist the LA to produce the statement. This will always include a medical report, a report from Nursery/School & a report from the Educational Psychologist.

Children who have learning support needs may not necessarily have a statement of SEN or EHCP. This is because the threshold for consideration,

and the speed of determination, of a statement/EHCP is high & is variable from LA to LA.

Under the system of SSEN, those children whose condition might impact on attendance, but not their learning support, would not have one e.g. a child being treated for cancer may miss a whole year of school but they would not have a statement because the health problems do not impact on learning ability, only attendance. One of the aims of EHCP is to be more holistic, and bring such children into the compass of a system that supports the spectrum of their needs, not simply the educational ones.

Many children who have ADHD/ADD & other significant conditions may not have a statement because the condition does not impact sufficiently on their learning abilities. They would not usually have a statement unless they also had other conditions that impacted on their education such as dyslexia.

If a child does have a statement/EHCP, this is a reliable indicator that the child does require a high level of additional support because they have a significant condition. However, the absence of a statement/EHCP does not mean that the child does not have a significant learning impairment.

The equivalent in Scotland is called Co-ordinated Support Plan (CSP). There is no difference to the English statement of SEN.

### **Note in Lieu**

If, after carrying out an assessment, the LEA decides not to draft a statement, they may issue a note in lieu. It may resemble a SEN but it has no legal force & does not necessarily provide any additional funding to meet the child's needs.

[Click here for more information about Note in Lieu](#)

### **Transition Plan**

This is for children moving from Secondary school to further or higher education. It outlines what the child wants to achieve in the next few years & what support they will need to live as independently as possible. It covers every aspect of their life, including education, employment, housing, health, transport & leisure activities. Most plans are drawn up in year 9.

The equivalent in Scotland is called Transition Planning / Future Needs Assessment. There is no difference to the English TP.

### **Transition (14+) Plan/Review**

This is for children moving from Secondary school to further or higher education. It outlines what the child wants to achieve in the next few years and

what support they will need to live as independently as possible. It covers every aspect of their life, including education, employment, housing, health, transport and leisure activities. Most plans are drawn up in year 9.

Pupil Ref no..... Pupil's name..... Date of birth.....

Transition needs	Action	Agencies responsible
Academic		
Career / Future		
Health / Therapy		
Personal / Family / Social		

General Action		
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### **Specialist Nurses**

Specialist nurses play a key role in the management of patient care. Working closely with doctors and other members of the multidisciplinary team, they educate and support patients, relatives and carers.

The specialist nurse has in depth knowledge of the physical, psychological and social effects of a specific condition.

Most specialist nurses are hospital based and have access to patient's medical records.

Some will visit patients in their own homes following discharge from hospital.

Some hospitals employ nurse specialists for:

- Blood disorders
- Gastro-enterological
- Gastro-intestinal
- Cardiac
- Respiratory
- Care of the Elderly
- Diabetes
- HIV/AIDS
- Incontinence
- Learning disability
- Oncology
- Neurology



- Pain Management
- Paediatrics
- Psychiatry
- Renal Medicine
- Skin Disease

### **Community Psychiatric Nurses (CPNS)**

CPNs are specialist nurses with the treatment and care of people with mental health disorders. They work both in hospitals with inpatients and with outpatients in the community where they see patients in clinics or visit in their homes.

CPNs work closely with general practitioners, psychiatrists, social services and voluntary groups. Some specialise in drug and alcohol dependency.

People with more severe mental health problems may be on supervised discharge from hospital and the community psychiatric nurse acts as the care co-ordinator.

CPNs administer drug treatment by regular injection, monitor the effects of treatment, and carry out psychotherapeutic techniques such as behaviour therapy and counsel patients and their carers.

### **Community Matrons**

Community matrons are highly experienced, senior nurses who work closely with patients (mainly those with a serious long term condition or complex

range of conditions) in a community setting to directly provide, plan and organise their care.

For further information click on the following link:

[NHS Careers/Matron](#)

### **Reports from Specialist Nurses**

Reports from the specialist nurse can provide details of the diagnosis, brief medical history, clinical examination findings, medication, treatment plan and response, variability of the condition, disabling effects and prognosis.

In people who have more than one medical condition, the specialist nurse may not be able to provide detailed information outside their own area of expertise or to be able to comment on multiple disabilities.

### **The Stanford-Binet Intelligence Scale**

The Stanford-Binet Intelligence Scale is a standardised test that measures intelligence and cognitive abilities in children and adults, from age two through to mature adulthood.

#### **Purpose**

The Stanford-Binet Intelligence Scale was originally developed to help place children in appropriate educational settings. It can help determine the level of intellectual and cognitive functioning in preschoolers, children, adolescents and adults and assist in the diagnosis of a learning disability, developmental delay, mental retardation, or giftedness. It is used to provide educational planning and placement, neuropsychological assessment and research. The Stanford-Binet Intelligence Scale is generally administered in a school or clinical setting.

#### **Precautions**

The Stanford-Binet Intelligence Scale is considered to be one of the best and most widely used intelligence tests available. It is especially useful in providing intellectual assessment in young children, adolescents, and young adults. The test has been criticised for not being comparable for all age ranges. This is because different age ranges are administered different subtests. Additionally, for very young preschoolers, it is not uncommon to receive a score of zero due to test difficulty or the child's unwillingness to co-

operate. Consequently, it is difficult to discriminate abilities in this age group among the lower scorers.

Administration and interpretation of results of the Stanford-Binet Intelligence Scale requires a competent examiner who is trained in psychology and individual intellectual assessment, preferably a psychologist.

## **Description**

Administration of the Stanford-Binet Intelligence Scale typically takes between 45 to 90 minutes, but can take as long as 2 hours 30 minutes. The older the child and the more subtests administered, the longer the test generally takes to complete. The Stanford-Binet Intelligence Scale is comprised of four cognitive area scores which together determine the composite score and factor scores. These area scores include:

- Verbal Reasoning,
- Abstract/Visual Reasoning,
- Quantitative Reasoning, and
- Short-Term Memory

### **Abstract and visual reasoning are analysed in Stanford-Binet intelligence tests**

Score is considered to be the best estimate of "g" or "general reasoning ability" and is the sum of all of subtest scores. General reasoning ability or "g" is considered to represent a person's ability to solve novel problems.

The composite score is a global estimate of a person's intellectual functioning.

The test consists of 15 subtests, which are grouped into the four area scores. Not all subtests are administered to each age group; but six subtests are administered to all age levels.

These subtests are:

- Vocabulary
- Comprehension
- Pattern Analysis
- Quantitative
- Bead Memory, and
- Memory for Sentences

The number of tests administered and general test difficulty is adjusted based on the test taker's age and performance on the sub-test that measures word knowledge. The subtest measuring word knowledge is given to all test takers and is the first subtest administered.

The following is a review of the specific cognitive abilities that the four area scores measure:

- The Verbal Reasoning area score measures verbal knowledge and understanding obtained from the school and home learning environment and reflects the ability to apply verbal skills to new situations. Examples of subtests comprising this factor measure skills which include: word knowledge, social judgment and awareness, ability to isolate the inappropriate feature in visual material and social intelligence, and the ability to differentiate essential from non-essential detail
- The Abstract/Visual Reasoning area score examines the ability to interpret and perform mathematic operations, the ability to visualize patterns, visual/motor skills, and problem-solving skills through the use of reasoning. An example of a subtest which determines the Abstract/Visual Reasoning score is a timed test that involves tasks such as completing a basic puzzle and replicating black and white cube designs
- The Quantitative Reasoning area score measures: numerical reasoning, concentration, and knowledge and application of numerical concepts. The Quantitative Reasoning area is

combined with the Abstract/Visual Reasoning area score to create an Abstract/Visual Reasoning Factor Score

- The Short-Term Memory score measures concentration skills, short-term memory, and sequencing skills. Subtests comprising this area score measure visual short-term memory and auditory short term memory involving both sentences and number sequences. In one subtest that measures visual short-term memory, the participant is presented with pictures of a bead design, and asked to replicate it from memory

## **Results**

The numbers of correct responses on the given subtests are converted to a Standard Age Score or SAS score which is based on the chronological age of the test subject. This score is similar to an I.Q. score. Based on these norms, the Area Scores and Test Composite on the Stanford-Binet Intelligence Scale each have a mean or average score of 100 and a standard deviation of 16. For this test, as with most measures of intelligence, a score of 100 is in the normal or average range. The standard deviation indicates how above or below the norm a child's score is. For example, a score of 84 is one standard deviation below the norm score of 100. Based on the number of correct responses on a given subtest, an age-equivalent is available to help interpret the person's level of functioning.

Test scores provide an estimate of the level at which a child is functioning based on a combination of many different subtests or measures of skills. A trained psychologist is needed to evaluate and interpret the results, determine strengths and weaknesses, and make overall recommendations based on the findings and observed behavioural observations.

## **The Wechsler Pre-school and Primary Scale of Intelligence**

The Wechsler Pre-school and Primary Scale of Intelligence (WPPSI) is used to assess the intelligence of children aged 2:6 years through 7:3 years. It provides composite scores that represent intellectual functioning in specified cognitive areas e.g. Verbal Intelligence Quotient (VIQ) and Performance Intelligence Quotient (PIQ) as well as providing a composite score

that represents an overall Intelligence Quotient (IQ). Processing Speed is measured by two supplemental tasks.

The scores obtained represent estimates of a child's true scores. That is, they reflect a child's abilities combined with some degree of measurement error. The Confidence Interval gives the range of probable scores:

- Composite Scores are Standardised Scores - have an average of 100 and a standard deviation of 15. The average range is between 85 and 115
- The Confidence Interval represents the band of scores where a child's true score is likely to be 90% or 95% of the time

**Performance on subtests can be reported in a number of ways, these include:**

Percentile – the number of children of a similar age out of 100 that a child would have performed as well as or better than. 50 is the average score.

Scaled Scores - lie between 1 and 19, with the average range between 7 and 13 (standard deviation of 3). These represent a child's performance relative to same-age peers.

Age Equivalent – the age at which 50% of children will achieve at this level.

Not all tasks on the WPPSI need to be completed on every occasion. However, a sufficient number of subtests need to be completed to report upon

an area of ability. The selection of tasks will vary according to each specific situation. The tasks that make up the WPPSI-III are detailed below:

### **Verbal Tasks**

<b>Task</b>	<b>Ability Measured</b>	<b>Description of Task</b>
Information	Crystallised intelligence, long-term memory, and the ability to retain and retrieve knowledge from school and the environment	The child responds to a question either by pointing or verbally. Where no verbal response is required, the child responds by choosing a picture from four response choices.
Vocabulary	Word knowledge, verbal concept formation, fund of knowledge, learning ability, long-term memory and degree of language development	Children are either shown a picture or told a word. For picture items the child names the pictures. For the verbal items the child gives a definition of the word read out loud to them.
Word Reasoning	Verbal Comprehension, analogic and general reasoning ability, the ability to integrate and synthesize different types of information, verbal abstraction, domain knowledge and the ability to generate alternative concepts	The child is asked to identify the common concept being described in a series of increasingly specific clues
Comprehension	Verbal Reasoning, and conceptualisation, the ability to evaluate and utilise past experience, verbal comprehension and expressions and the ability to demonstrate practical information	The child answers questions based on his or her understanding of general principles and social situations.



Similarities	Verbal reasoning and concept formation	The child is read an incomplete sentence containing two concepts that share a common characteristic. The child is asked to complete the sentence by providing a response that reflects the shared characteristic.
Receptive Vocabulary	Comprehension of verbal directives, auditory and visual discrimination, auditory memory, auditory processing and the integration of visual perception and auditory input.	The child looks at a group of four pictures and points to the one the examiner reads out loud to them.
Picture Naming	Expressive language ability, word retrieval from long-term memory and association of visual stimuli with language	The child names a picture that is displayed.

## Performance Tasks

<b>Task</b>	<b>Ability Measured</b>	<b>Description of Task</b>
Block Design	Analyse and synthesise abstract visual stimuli	The child views a constructed model or a picture and uses one or two colour blocks to recreate the design within a specified time limit.
Matrix Reasoning	Fluid Intelligence	The child looks at an incomplete matrix and selects the missing piece from 4 or 5 response options.
Picture Concepts	Abstract and categorical reasoning	The child is presented with two or three rows of pictures and chooses one picture from each row to form a group with a common characteristic.
Picture Completion	Visual perception and organization, concentration and visual recognition of essential details of objects	The child views a picture and points to or names the important part missing from the picture
Object Assembly	Visual-perceptual organization, integration and synthesis of part-whole relationships, nonverbal reasoning and trial-and-error learning	The child is presented with a standardised configuration of puzzle pieces and allowed 90 seconds to fit the pieces together to form a meaningful whole.

## **Processing Speed**

Tasks	Ability Measured	Description of Task
Coding	Processing speed, short-term memory, learning ability, visual perception, visual-motor coordination, visual scanning ability, cognitive flexibility, attention and motivation	The child copies symbols that are paired with simple geometric shapes. Using a key, the child draws each symbol in its corresponding shape.
Symbol Search	Processing speed, short-term visual memory, visual-motor coordination, cognitive flexibility, visual discrimination and concentration	The child scans a search group and indicates whether a target symbol appears in the search group by marking the appropriate symbol with a pencil.