

**Prader-Willi Syndrome Association Response to the  
Consultation on the Personal Independence Payment: Assessment thresholds and Consultation**

Prader-Willi Syndrome is a complex, rare genetic syndrome which affects about 1:29,000 live births.

Typically people with Prader-Willi Syndrome (PWS) can exhibit:

- weak muscle tone, sometimes coupled with poor co-ordination and balance
- excessive uncontrollable appetite and a decreased calorific requirement which without controlling factors leads to obesity and a reduction in life expectancy
- central nervous system and endocrine dysfunction causing varying degrees of learning disability, typically an IQ of 60 - 70
- somnolence
- immature hypothalamus causing poor emotional and social development, which can cause extremely challenging behaviour
- immature development of sexual organs
- short stature, unless treated with growth hormone during childhood

These characteristics impact on the ability of people with PWS's ability to find and maintain paid employment. Although some individuals may have the physical and intellectual ability to work, they are often ill equipped on both an emotional and social level to function within the workplace.

Healthcare professionals will, on average only see one case of PWS during their working life and so may not have any knowledge of the syndrome. Even if they have cared for someone with PWS, its rarity means that their experience could be atypical. Rare syndromes such as PWS will create a need for the assessors to be suitably trained before assessing someone with the syndrome.

In the case of PWS, a person's disability in many cases is not at all obvious and they may well present at first sight as perfectly capable of looking after themselves. Their ability to understand what is said to them is often very easily overestimated.

**Activity 1 – Preparing food and drink**

Some people with PWS would present at an interview as able to prepare and cook a simple meal unaided and would also both claim that they could, and prefer to do so. However, the excessive and uncontrollable appetite of those with PWS makes it inappropriate for most to have access to food unsupervised. Their appetite and obsession with food is such that, if unsupervised, their lives become endangered through obesity, stomach and bowel complications. These are life threatening and need to be judged as requiring supervision 'to prevent a potentially dangerous incident occurring'.

### **Activity 3 – Managing therapy or monitoring a health condition**

The examples of possible therapeutic activities include exercise regimes, which people with PWS need to both improve muscle tone and to help combat obesity, but for someone with PWS managing a low calorie diet is also a 'long term home therapeutic activity . . . without which their health is likely to deteriorate'. The obesity that people with PWS not on a low calorie diet develop is life threatening and only a combination of reducing calorific intake together with an exercise regime will combat it. The fat cells of someone with PWS also have a higher calorific value than normal, making losing weight harder. The poor muscle tone associated with PWS, together with obesity also make exercise more difficult, and more difficult to manage.

### **Activity 8 – Engaging socially**

People with PWS may present at interview as able to engage socially as this will be an anticipated, planned, controlled meeting. However, people with PWS have an immature hypothalamus giving adults the level of control over their emotions usually associated with that of a young child. Challenging behaviour and uncontrolled temper tantrums are both common features that may not be apparent at an interview.

### **Activity 7 - Communicating**

Some people with PWS may seem at interview to be articulate but in reality they are likely to have difficulties with sequential processing, so struggle understanding complex verbal information. It would be essential for an assessor to take this into account when assessing someone with PWS.

### **Activity 9 – Making financial decisions**

This activity considers the ability of an individual to make financial decisions, but does not take into account those for whom it would not be in their best interests to take their own financial decisions. Some people with PWS may be quite capable of calculating the costs of goods and change required, of calculating personal budgets, but given the power to do so, will spend everything on food, so putting their health and welfare at risk. There needs to be a descriptor for those who '**should** not manage financial decisions unaided'

For the Attention of Susan Passmore CEO PWSA(UK)

Dear Susan,

We have not met but may I add my message of welcome to you, to PWSA(UK).

I am writing in response to the request for consultation over PIP.

I feel it is important that those of us responding to this consultation should do it via the Association to make our voice as loud as possible.

My point is this. It seems to me that one very big question is being begged. How and by whom are the assessors to be trained as to what the needs of a person with PWS are. As we know there are hundreds of conditions, many very rare and many giving rise to unique needs and different degrees of disability. How can an assessor be aware of all of them and be qualified to give an opinion as to the qualification or otherwise of a person to receive a benefit on all of them. In the case of PWS, a persons disability in many cases is not at all obvious and they may well present at first sight as perfectly capable of looking after themselves. Their ability to understand what is said to them is often very easily overestimated. May I urge you to make use of our team of medically skilled friends to put this point to the DWP.

I am sure I am stating the obvious and that you are already right up with it, but I just wanted to add my voice.

With best wishes

Julien Courtauld

(I was for many years a Trustee of the Association)