

1 What is Prader-Willi Syndrome (PWS)?

PWS is a complex genetic disorder that affects both males and females from birth and throughout their lives. Main characteristics are: low muscle tone with consequent motor developmental delays, a mild to moderate learning difficulty, incomplete sexual development, and emotional and social immaturity, which can lead to challenging behaviours. During childhood, an overwhelming and insatiable chronic appetite usually develops which, without rigorous food management and exercise regimes, leads to food seeking, stealing and life threatening obesity. PWS occurs randomly in about 1:22,000 births. It is estimated there are about 2,000 living with PWS in the UK.

2 Other Characteristics

Many people with PWS also exhibit characteristic facial & other physical features: almond-shaped eyes, a narrow forehead (measured across), a downturned mouth with a triangular-shaped upper lip, small hands & feet. They also have poor large muscle strength, often coupled with poor coordination and balance. Muscle tone can be improved with appropriate therapy and exercise. Small muscle strength is usually better. Some physical features can be improved with growth hormone treatment. Most people with PWS have borderline or moderate learning disabilities. A minority have severe learning difficulties.

3 Life as an Adult

As adults, people with PWS have varying abilities in attaining independence, although all will need some form of support or monitoring to help with controlling their food intake, and thus their weight. Many will have the intellectual and physical ability to work, but are usually ill-equipped on an emotional and social level to deal with the stresses and demands of the ordinary workplace.

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However, they can make a positive contribution to society in many ways and may be involved in voluntary work, craft work, or have a part-time job. Many people live with their families, but an increasing number are living in residential homes, or being supported to live in the community. In the past, life expectancy was short because of health problems associated with massive obesity but nowadays life expectancy is increasing because of better dietary management and better understanding of the problems associated with PWS. More information about PWS



6 PWS & Mental Capacity: Intervention

In most cases early intervention and preventative actions can result in positive outcomes in both dietary & behaviour management. Consideration should be given to:

- Positively engaging the person from the beginning of your involvement
- Obtaining informed consent (where possible)
- Carry out a mental capacity assessment where appropriate

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PWS and Mental Capacity

An important consideration is whether the person has mental capacity to understand the risks associated with actions/lack of actions, e.g. risks of overeating & obesity versus benefits of following dietary management plans, or learning about staying safe, and having the associated executive capacity/functioning to be able to carry out the appropriate actions. Remember mental capacity is decision-specific, not a blanket term of 'has' or 'has not' mental capacity.

7 Raising a Safeguarding Concern

Responding to the needs of a person with PWS depends upon the level of risk/harm posed to the person and/or others, and whether the person is able to protect themselves and determine their own action.

Workers and volunteers should refer to the threshold guidance to determine whether a safeguarding concern should be raised.

More information about PWS is available at: [the PWSA UK website](#) and the [NHS website's PWS page](#).

