

Information for Residential Care and Supported Living Staff

Prader-Willi Syndrome (PWS) is a rare lifelong **genetic** condition which is present from birth. Caused by an abnormality on chromosome 15, it affects the functioning of the hypothalamus. Main characteristics of the syndrome are:

- Hyperphagia (excessive appetite, overeating) – beginning around 2 -4 years
- Hypotonia (low muscle tone) – from birth
- Hypogonadism (immature sexual development)– from birth
- Short stature, compared with other family members
- Immature emotional and social development
- Learning disabilities ranging from severe to borderline. A minority of people with PWS do not have a measured learning disability but will not function at the level of their measured IQ
- Challenging behaviour, ranging from mild to severe (regardless of whether or not the person has a learning disability). A significant number of individuals with PWS also have a diagnosis of Autistic Spectrum Disorder or Attention Deficit Disorder.

Hyperphagia (excessive appetite)

There is currently no cure or appropriate medication to treat the excessive eating which is one of the major characteristics of the syndrome. The severity of this feature varies between individuals and may vary across time in any single individual. Low muscle tone, and an abnormal body composition in which there is more fat mass than lean muscle (even in individuals who are not overweight), means that a child or adult with PWS needs fewer calories than someone of the same age to maintain a healthy weight. Excess calorie intake can quickly result in significant obesity.

Currently the only way to prevent life-threatening obesity in PWS is to control access to food and to keep the person on a lower calorie diet – not easy when the person is hungry most of the time. Deaths at a very early age from obesity-related complications are unfortunately not uncommon.

Immature sexual development

Sex hormone treatment may be given to teenagers and adults of both sexes to enable them to become more sexually developed, but fertility is extremely rare. Worldwide there are only four reported cases of women with PWS having a child (none in the UK) and none of a man with PWS fathering a child.

Behaviour management

Challenging behaviour is common. Typical behaviours are:

- Temper outbursts (tantrums, rages), sometimes including aggressive verbal or physical assaults
- Food seeking and/or stealing
- Stubbornness and resistance to change, argumentativeness
- Perseveration (asking the same question, or reverting to the same subject over and over again)
- Compulsive behaviour and insistence on routine
- Attention-seeking and manipulative behaviour
- Skin-picking

- Lying and blame-shifting

Mental health problems may also emerge, and these seem to be particularly prevalent in those who have the **maternal disomy type** of PWS.

Adults with PWS

Some adults with PWS appear more able and articulate than is actually the case, and will tell outsiders that they know all about diet and are able control their own food intake – this is almost universally not the case. There are particular risks attached to this, and one is to underestimate the amount of support an individual needs, particularly where they are in a **supported living** situation.

Mental capacity around food and finances can be an issue, particularly where the person is generally high- functioning. Sometimes a Mental Capacity Assessment may be the only way to limit the person's access to food. Very rarely this has gone to the Court of Protection. Unfortunately, the case law for this is not yet in the public domain, but in those cases which are known to the PWSA, it has been deemed that the person does not have **capacity** around food.

Employment and day time occupations

It is extremely rare for an adult with PWS to be in full time paid employment, even when they have the intellectual capacity to do a particular job. The tendencies to food-seeking and stealing, sleepiness, and rage outbursts may all affect their overall abilities. However, part-time paid work or voluntary work is quite possible, and helps the person's self-esteem by providing them with a way of contributing to their community.

Best Practice Guide for employers

Healthcare and risk factors

Unusual features and potential risk factors in PWS

Health care

Information for GPs

Information for A&E and hospital staff

Personal qualities in staff which resonate best with those with PWS

- Fair but firm, and consistent in approach
- Not easily swayed or manipulated
- Positive attitude and tone of voice
- Calm in all situations
- Patient
- Able to build up a relationship of trust
- Able to communicate in clear and simple ways
- Reliable

Organisations providing care for people with PWS

Several organisations specialise in providing residential care and supported living for PWS, with particular emphasis on limiting access to food, calorie-controlled diets, and management of challenging behaviour. The PWSA UK maintains a list of these providers – **contact us** to obtain a copy of the list.

The PWSA UK publication *Beyond the Veneer: A Guide to the Essential Features of Residential Care and Supported Living for Adults with Prader-Willi Syndrome* is available to purchase and provides a checklist of things to consider when providing care for someone with PWS.

Enquiries and training

PWSA UK has a wide range of information and knowledgeable staff who can help with enquiries. **Training** is also available in-house for residential care and supported living staff and the Association also delivers the nationally accredited AIM Award – Understanding the Needs of Individuals with PWS.

Residential Care and Supported Living Forum

This Forum, organised by PWSA UK, runs three times a year (usually March, July and November in Derby) and provides an opportunity for senior managers to get together to share concerns and good practice in caring for people with PWS. Occasional single topic days are also held. To be included on the emailing list for this forum, contact jwaters@pwsa.co.uk

The International PWS Organisation has a Professional Provider and Caregiver Board and produces best practice guidelines for caregivers.