

## Prader-Willi Syndrome (PWS)

Prader-Willi Syndrome (PWS) is a genetic disorder first identified by three Swiss doctors in 1956, Drs Prader, Labhart and Willi. Its features include food obsession, shortness of stature and poor muscle tone. It affects boys and girls alike, and occurs in families from all backgrounds. It is estimated that one in every 10,000 to 12,000 births will have the disorder. It is the most common known genetic cause of life-threatening obesity in children.

Research indicates that PWS may be the result of four different genetic abnormalities on chromosome 15. In approximately two-thirds of cases there is a deletion on chromosome 15 coming from the father. In about 30% of cases both copies of the chromosome are inherited from the mother, instead of one from her and one from the father. If you have one child with PWS it does not necessarily follow that you are any more likely than the population at large to have another, but genetic counselling is advised. Genetic testing is by DNA (blood) sample of both partners and the affected child.

### Early indications

PWS, being a genetic disorder, is there from birth. In the very early months parents may notice that their baby has difficulty sucking, is very quiet and sleepy compared to other babies, and appears to be very 'floppy'. Later, from about six months to a year, their baby will become more alert and awake as the tendency to sleep recedes, but the early feeding problems will be replaced from the age of two onwards by an almost insatiable desire to eat, although the intensity of this feature will vary considerably between individuals.

### Physical characteristics

These may include short height, very small hands and feet, narrow face, small mouth, almond-shaped eyes and underdeveloped sexual organs. It has also been noted that up to half of Caucasian children with PWS will be fair-haired and have blue or grey eyes, regardless of their parents' colouring.

### Developmental delays

Motor and language skills will be affected, resulting in late walking and talking plus problems with articulation.

### Behaviourally

In their early years the children are friendly, loving and placid, but behavioural difficulties usually set in and become more severe as the children grow and they have to cope with their insatiable desire for food. Stealing and hiding food may become common activities as the obsession grows, and many outbursts of temper and rage can be put down to these 'needs' being necessarily moderated by their parents or carers. Others may relate to any change in their accepted daily routine which will upset them. Anxiety and worry may result in compulsive skin-picking and other similar self-injurious behaviours.

### Other

Possibly high pain threshold, sleep disorders, breathing difficulties, curvature of the spine and physical inactivity. Psychiatric problems are not uncommon in adolescents and adults.

### In general

Comprehension is generally better than verbal skills. Moderate learning difficulties across the board are common, but severe learning difficulties are rare. Many individuals will have specific difficulties with maths, writing, short-term memory and auditory processing.

Children with PWS have good visual organisation (parents have observed a higher than average ability to do jigsaw puzzles), good reading skills and a wider than average vocabulary. When they are older they may be over-friendly, chatter a lot, be immature compared with their peers and be socially isolated.

**Remember**, not all babies and children with PWS will display the same characteristics, or to the same degree, as another child with the disorder. Control of their diet is the major problem with this disorder and must be seen as a prime goal if the young person with PWS is not to become obese with subsequent life-threatening or damaging implications. This is especially true when a young person with PWS is allowed more independence and thus more access to food. Unfortunately, appetite suppressants are not generally successful and sometimes have side-effects.

The majority of children with Prader-Willi Syndrome will become adults with Prader-Willi Syndrome, and as long as their general health stays good and their tendency to obesity can be controlled, their life expectancy will be much the same as the general population.

### Hints

- As with many disorders, a multi-disciplinary approach is advised and should include input from physio-, occupational and speech and language therapists
- Ask your GP for referral to a dietician / nutritionist for a controlled diet and eating plan
- Encourage a daily exercise routine from a very early age: seek help from your local sports centre
- It is not a good idea to use food as a reward with these children! Try a rewarding trip to the swimming pool, gym, walk on the beach or in the countryside.

### Teachers should be aware that their student with PWS may have problems with

- coordination
- handwriting
- maths
- abstract concepts
- attention span, especially if required to listen for 'long' periods
- socialising skills
- controlling their hunger
- controlling their temper, particularly if they want to eat and food is withheld
- worries and anxieties involving all the above which may result in minor self-injury

### Other Useful Contacts

**PWSA (UK)** 125a London Road, Derby DE1 2QQ. Offers support and advice to parents, professionals and individuals with PWS. (Mon to Fri 9.30-3.30/Ans)

**Web:** [www.pwsa.co.uk](http://www.pwsa.co.uk)

**Tel:** 01332 365676

**Email:** [admin@pwsa.co.uk](mailto:admin@pwsa.co.uk)

**Gretton Homes** Gretton House, 3 High St, Gretton, Northants NN17 3DE. Gretton Homes are a group of 8 residential holistically run care homes supporting almost 70 people with PWS of 18+ years.

**Web:** [www.grettonhomes.co.uk](http://www.grettonhomes.co.uk)

**Tel:** 01536 770325

**Email:** [info@grettonhomes.com](mailto:info@grettonhomes.com)

**Putney House & Putney Cottages** 47 Scarborough Road, Bridlington YO16 7PE. Specialist residential homes for adults with PWS offering a variety of support and opportunities.

**Tel:** 01262 674818

**Tel:** 01262 674818

**Web:** [www.themtg.co.uk](http://www.themtg.co.uk)

**Email:** [ken.davenport@themtg.co.uk](mailto:ken.davenport@themtg.co.uk)

### Publications

Contact the **Prader-Willi Syndrome Association (UK)** for their publications list. Their book **Babies & Children with PWS: A Handbook for Parents & Carers** is free (one copy) to parents and carers who join the Association, £5.00 (Including P&P) thereafter, or to others.

**Prader-Willi Syndrome: A Practical Guide.** Author Jackie Waters. 1999. Published by David Fulton Publishers Ltd. ISBN1-85346-614-X. Around £20.00 + p&p. Available from PWSA (UK) or visit [www.amazon.co.uk](http://www.amazon.co.uk).

### Internet sites

[www.ipwso.org](http://www.ipwso.org) the website of the International Prader Willi Syndrome Organisation

[www.pwsausa.org/](http://www.pwsausa.org/) is the Prader-Willi Syndrome Association (USA) site

Note: The OAASIS Information Sheets use 'he'/'his'/'him' rather than the cumbersome 'he / she'/'his / her'/'him / her'. No sexism is intended. The sheets are checked annually; please ensure you have the current version.

### OAASIS publications

- SEN Information Sheets
- First Guide to ...
- Cards explaining 9 learning disabilities
- Legal advice Information Sheets
- SEN 'How to' guides

All OAASIS publications can be downloaded from [www.oaasis.co.uk](http://www.oaasis.co.uk)