



Health & Wellbeing



Caring for those with Prader-Willi Syndrome.

This information sheet has been designed to provide information for those caring for people with PWS on a day-to-day basis. Each PWS person is an individual and may have some or all of the below mentioned characteristics.

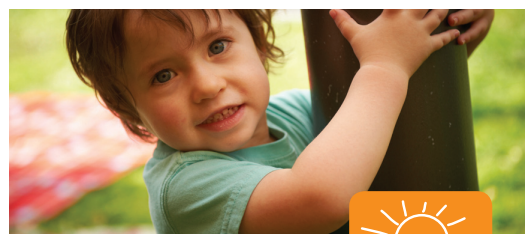
In the day to day medical care of people with PWS the specific areas of concern which require regular monitoring include:

- Weight and body mass index
- Blood pressure
- Sleep Apnoea: even when obesity isn't a problem
- Scoliosis
- Osteoporosis: due to lack of sex hormone
- Diabetes: type 2
- Oedema: water retention
- Skin infections
- Constipation
- Hypothyroidism

PWS presents a number of challenging characteristics that impact on the health and wellbeing of a person with PWS. All personnel involved in the medical care and management of people with PWS should be aware of these characteristics.

“People with PWS are generally as healthy as the general population, and may also be prone to the same illnesses and diseases. However there are some unusual features of PWS which require special attention and may not always be evident to those who have little experience of the syndrome. “

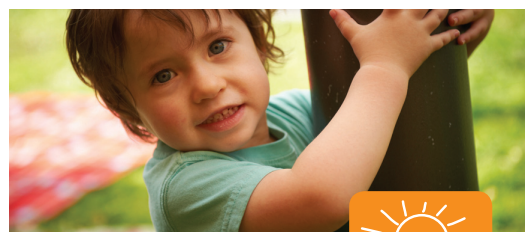
“The most obvious way of preventing illness or death is to keep weight in reasonable limits. Currently this can only be achieved by restricting food intake, supplementing with exercise and close supervision.”¹



Health & Wellbeing (cont'd)

Medical alerts that could be life threatening:

Features	Description
Anaesthesia, medication reactions	May have unusual reactions to STANDARD doses of medications & anaesthetics. Use caution with medications that cause sedation.
Water intoxication	This relates to the use of certain medications with antidiuretics, as well as from excess fluid intake alone.
High pain threshold	May mask presence of infection or injury. This fact should be recorded on their medical file.
Bone fractures	Due to the high pain threshold, it is not unusual for a person with PWS to have an undetected broken bone.
Respiratory concerns	Increased risk for respiratory difficulties eg. Hypotonia, weak chest muscles & sleep apnoea possible complications.
Lack of vomiting	Emetics may be ineffective, repeated doses may cause toxicity. Presence of vomiting may signal a life threatening illness.
Risk of choking	There is a risk of choking arising from people with PWS trying to eat food quickly, either because they habitually do this, or because they are trying to disguise the fact that they have taken food to eat which they should not have done.
Severe gastric illness	General feeling of unwellness rather than localized pain. Abdominal distension or bloating, pain and/or vomiting may be signs of life threatening gastric inflammation more common in PWS than in general population.
Body temperature abnormalities	Fever may be absent despite serious infection. Hypothermia may occur during minor illness or during an anaesthetic.
Skin lesions & bruising	Many individuals with PWS bruise easily but, because of the high pain threshold, are often unable to say how they came by the bruise.
Hyperphagia	Excessive appetite.
Surgical & Orthopaedic concerns	Surgeons need to be aware of narrow airway, underdevelopment of the larynx & trachea, hypotonia, oedema & scoliosis.
Diabetes	Children with PWS have low insulin levels, but they don't need higher levels.
Cortisol hormone insufficiency	There has been a suggestion that some people with PWS may be at increased risk of stress hormone (cortisol) deficiency due to hypothalamic problems but this has not been found in all studies and probably occurs only rarely.



Health & Wellbeing (cont'd)

In addition to these more serious conditions mentioned above, the following list provides additional health concerns, that if not managed and monitored can escalate into more serious health issues.

Features	Description
Respiratory problems	Common in infants and assistance with clearing secretions may be needed. Insufficient fluid intake, due to feeding difficulties, can occur.
Bowel problems	Difficult to detect due to decreased pain threshold. Lack of vomiting, already obese... eg, Stomach already distended.
Rectal picking	May lead to increased infection.
Dental problems	Soft tooth enamel, thick sticky saliva and poor oral hygiene.
Strabismus	Crossed eyes.
Delayed gross motor development	
Language / speech disorders	

In addition to careful monitoring health wellbeing, carers and professionals involved in the care of a person PWS, need to be aware that the major causes of illness and death can be quite sudden and unexpected.

Common causes include:

- Obesity (heart problems and respiratory problems)
- Pneumonia (ensure that the person is vaccinated)
- Gastric necrosis (due to binge eating)
- Choking
- Septicaemia
- Complication from diabetes
- Food poisoning

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