

Healthcare in PWS 5–10 years



Introduction

Children with PWS are generally as healthy as other children, but are also prone to the same childhood 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 or when the unusual features are subtle or mild and hence more difficult to identify.

These features are included in this article, along with some health issues which are quite common in children with PWS. Please note that the issues vary considerably between individuals with PWS and not all children with PWS will necessarily have them all. As time goes by, you will be able to identify which of those issues may apply to your child.

Food seeking

By this age most children are quite mobile, can run, jump and climb and go up and downstairs, partly due to natural improvement in their hypotonia (poor muscle tone) and/or growth hormone treatment.

With improved mobility and increased appetite, food seeking may become more evident.

Many children can be taught what is edible and what is not; but unless you can be absolutely sure, it is best not to leave your child unsupervised, especially as he is less likely to be sick after eating something unsuitable or inappropriate. If he does eat something he should not have, seek medical advice appropriately.



NOTE

For ease of reading, the text refers to “he, him, his”, but the details are equally relevant to girls, unless otherwise stated.

Vomiting

Individuals with PWS do not commonly exhibit a vomiting reflex. Many children with PWS are rarely sick, even when unwell.

If your child has a fever, or is not responding as they usually do, you should seek your doctor's help, to check them out.

Lack of vomiting cannot be taken as sign that little is wrong with your child.

See also the section on TEMPERATURE

Continued ...



Food seeking (continued)

The degree to which children of this age will seek food does vary considerably between individuals. Some are happy to be part of a dietary regime where they only eat what is put in front of them, others will ask for and seek food whenever an opportunity arises.

Children with PWS may be quite indiscriminate in what they eat, e.g. poisonous berries, out-of-date food, frozen food, food from waste bins or off the ground. There are reports of people with PWS eating large quantities of items like salt or shampoo, though this is not thought to be very common.

The lack of vomiting reflex (see page 1) may mask or hide the fact that the person has ingested inappropriate items. A dramatic weight increase within a day - especially if coupled with reports of abdominal distress (and occasional vomiting) - may be a sign that your child is severely ill. Similarly, symptoms such as loss of appetite, diarrhoea or significant fluid retention are also reasons for concern.

Vaccinations

Unless your GP or paediatrician advises otherwise, your child should receive ALL the usual childhood vaccinations against disease, including flu- there are unlikely to be any side effects from these, other than those usually experienced.



Scoliosis

Children with PWS have a high risk of developing scoliosis (curvature of the spine to one side). Hence your child's spine or back should be examined regularly (or at least once a year as a minimum) to identify if he has scoliosis and to institute any intervention if needed.

Scoliosis develops in children with PWS for a number of reasons. Children do have poor muscle tone and poor muscle power; this often leads to poor posture and development of scoliosis. Often associated obesity or increased weight can make the scoliosis worse.

Parents worry that treatment with growth hormone can precipitate or worsen the scoliosis. However, growth hormone is generally safe and does not cause an increased incidence of scoliosis. However, in those who already have scoliosis, it may worsen and hence should be monitored more closely. If a particular child has severe scoliosis, then growth hormone therapy is contra-indicated.

Conservative measures like good posture, good care of the back and physiotherapy are all helpful and useful. Generally no other treatment is required for mild scoliosis. However for moderate to severe scoliosis, interventions are needed (such as a spinal brace). In very severe and complex cases of scoliosis (and additional kyphosis or forward bending of the spine) surgical intervention may be needed.

Skin lesions

A common feature of PWS is skin-picking, which may be first noticed at this age. The article on **Behaviour Management** <https://www.pwsa.co.uk/assets/files/Behaviour-children.pdf> gives you more information about how to manage this. It is extremely important to monitor any wounds for infection and obtain appropriate treatment from your GP if this occurs.



Information for hospitals and GPs

You will find further information to give to hospitals and GPs included in this pack or on our website at <https://www.pwsa.co.uk/information-for-professionals/health>

Respiratory problems/Obstructive sleep apnoea

Children with PWS may have respiratory problems and are prone to chest infections. Sleep apnoea (where the child momentarily stops breathing while asleep) is sometimes seen in children with PWS; this is exacerbated when the child also has increased weight gain or obesity. When sleep apnoea is suspected, please alert your doctor because this will require further specialist investigations and treatment as appropriate.



Bruising

Many children with PWS can bruise easily; additionally they may not cry out at the time of injury or accident, due to high pain threshold. Hence the presence of a bruise should alert you to actively look for any other signs of an injury to your child.

Temperature

A child with PWS may have below-normal temperatures at times and may not have a high temperature even when seriously ill. Even slight temperature elevations should be considered as a warning sign and to observe your child closely for any other clues. If in doubt, ask your doctor to examine him. It is also a good idea to ask your doctor to make a note of his temperature when he is healthy so that you have a base line guide for variations.



Risk of choking

Choking happens when food or fluids gets into the airway, rather than going into the stomach. This can happen in any child, but lack of vomiting reflex makes it more difficult and tricky to identify in a child with PWS. Encourage your child to eat slowly and chew food properly before swallowing.



PWS Specialist Clinics

In a few areas of England and Scotland, hospitals run specialist clinics for children with PWS.

Specialist clinics currently run at:

- Chelsea & Westminster Hospital, London
- Royal Alexandra Hospital, Brighton
- Birmingham Children's Hospital
- Royal Hospital for Sick Children, Glasgow
- Royal Stoke University Hospital, Stoke on Trent

These clinics allow you to see several specialists in one day and are staffed by professionals with a special interest in PWS.

PWSA UK representatives attend the clinics, when possible, to offer non-medical support.

Health care in PWS

High pain threshold

Individuals with PWS frequently have decreased sensitivity to pain and thus there is a potential danger of under-estimating the problem.

Thus, all known injuries must be assessed by a GP or paediatrician to exclude any serious problems. If your child shows symptoms that are out of the ordinary, it is also a good idea to have him checked over.

Following a significant fall or other injury, your child should be closely monitored for a change in posture, walking or movement of limbs.

Observe for deformities, swelling or bruising as these may indicate an undetected broken bone or fracture.

Thank you

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Obesity-related problems

Obstructive sleep apnoea and breathing difficulties (respiratory failure) are the most common problems for the child who is significantly overweight. Good and pro-active dietary management can help prevent obesity and obesity related problems at this age. However, it is extremely important to have regular check-ups to ensure weight is within the normal range; also this will help to put measures in place if weight starts to creep up.

Low cortisol levels

Cortisol is an important stress hormone which the body produces all the time, but increases the production to help deal with illness, infections or stress. There have been several studies looking at adrenal function in individuals with PWS. A study from Holland raised the possibility that deficiency of cortisol was much more common than expected in PWS; however later studies have not reproduced this result.

Individuals with PWS are probably more likely than the rest of the population to have cortisol deficiency, and the current guideline suggests that individuals are not tested routinely, but a test should be done if there is any concern. Symptoms of low cortisol include tiredness, low blood pressure, episodes of low glucose or fainting.

Anaesthesia

There is nothing inherent in PWS which gives cause for concern with the administration of anaesthesia. However, all individual health problems related to PWS should be taken into account. These include:

- **Obesity** (may be associated with obstructive sleep apnoea, high blood pressure, poor breathing pattern (hypo-ventilation) which is further exacerbated by hypotonia or poor muscle tone etc.).
- **High pain threshold** (see previous page).
- **Temperature instability** - you should inform the anaesthetist about your child's usual temperature range.
- **Food seeking behaviour** is common in children with PWS; though generally not in infants. However, the anaesthetist or health professional should assume that your child has eaten and has food in his stomach, unless you can verify otherwise.
- **Low muscle tone (especially in younger children)** may cause difficulties in their ability to cough and clear secretions from their airway. In addition, thick saliva may further complicate airway management.
- **Excessive post-operative drowsiness** may be present in some children.

Further information about anaesthesia

You can download information about anaesthesia and PWS to hand to your child's anaesthetist at:

www.orpha.net/data/patho/Pro/en/Prader_Willi_EN.pdf