

## Healthcare in PWS 2–5 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

PWS children are usually more mobile at this age, partly due to natural improvement in their low muscle tone (hypotonia) and/or growth hormone treatment. With improved mobility and increased appetite, food seeking is characteristically noticed in PWS children as opposed to poor feeding in infancy – although it may not always have shown itself to be an issue yet for some children.

Sometimes children with PWS may also eat inappropriate items, such as modelling clay etc. 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 (See **Vomiting**). If he does eat something he should not have, seek medical advice.



#### NOTE

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

### Squint (strabismus)

A squint, where one or both eyes point in different directions, is common in babies and children with PWS, and is most likely due to poor muscle tone.

Corrective treatment is by patching or a simple surgical operation.

Your paediatrician or eye specialist will advise you on timing and best form of treatment.

See also “Vision and Care of the Eyes” <https://www.pwsa.co.uk/assets/files/eyes-birth-13.pdf>



## Orthopaedic problems

### 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.

### Hip dysplasia

Babies and young children should also have hip checks from an early age to look for symptoms of hip dysplasia. If treated early, there should be no long term problems.

## Vaccinations

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



**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](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.

### 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. Hence, do not try to feed your child faster than he is able to cope with.

## Undescended testes in boys

Undescended testes are quite common in boys with PWS. Your paediatrician will advise you on the timing for the best course of treatment, which often involves a simple operation to bring down one or both testes.



## 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.

## Vomiting/abdominal pain

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 a sign that little is wrong with your child.



### 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.

Also, they may not cry and alert you if they are injured or experiencing pain.

Thus, all known injuries must be assessed by a GP or paediatrician to exclude any serious problems. In time, you will be able to recognise subtle signs when your child is out of sorts or unwell.

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

Thank you to Dr Shankar Kanumakala from the PWS Clinic, Royal Alexandra Hospital, Brighton, and to Dr Nicola Bridges from the PWS Clinic, Chelsea & Westminster Hospital, for their help in checking this document.

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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.

## 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.
- **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 or infants)** 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](http://www.orpha.net/data/patho/Pro/en/Prader_Willi_EN.pdf)