

Information for Primary School Staff



Introduction

Prader-Willi syndrome (PWS) is a complex developmental disability that results from a defect on chromosome 15. It causes a malfunction in the area of the brain called the hypothalamus, which controls aspects such as appetite, temperature and emotion.

PWS can be characterized by:

- Hypotonia (low muscle tone)
- Hypogonadism (underdeveloped sex organs)
- Hyperphagia (uncontrollable hunger)
- Cognitive impairment
- Challenging behaviours

Health professionals that may be involved with this child include:

- Dietician
- Endocrinologist
- Geneticist
- Occupational Therapist
- Ophthalmologist
- Orthopedic nurse or surgeon
- Orthoptist
- Orthotist
- Paediatrician
- Physiotherapist
- Speech therapist

(You may need to seek advice from the above in addition to reading this leaflet)



YouTube information video

You can view a 35 minute YouTube video which gives more information about children with PWS in a school setting at <https://www.youtube.com/watch?v=3F415xDp1A&feature=youtu.be>

A child with Prader-Willi syndrome is joining your school

This pack contains information which is appropriate to the age of this child and will help you to manage their needs, maintain a safe environment and facilitate learning.

But do remember- if you have any queries, we are here to help. We are only a phone call or email away.



What's special about this child?

Please remember that PWS is just a part of your pupil. They will have similarities and differences, as all other children do. Many people with PWS have specific and valuable attributes and skills. Many have excellent reading and writing skills as well as fine motor skills and are particularly clever with jigsaw puzzles and fine handiwork. They often have excellent long term memory and visual processing skills.

What to expect from this child and how you can best support them

Learning Disabilities and challenges

Many children with PWS experience some level of learning disability. The level of which varies from one child to the next. Please do be aware that even if the child presents as able, there may be underlying issues with regard to comprehension and cognitive ability. It is common for children with PWS to face challenges in expressive language use and short term memory.

What can you do to help?

Ensure instructions are repeated and ask the child to repeat instructions back to you. Check understanding and include 'refreshers'/ revisit previous work regularly.

Dietary needs and food seeking behaviours

Food seeking behaviours are common in people with PWS and the pupil is likely to have lots of anxiety surrounding food. These types of behaviour are a result of the damage to the hypothalamus, meaning the child is unable to experience the sensation of fullness. It is very unlikely that a child with PWS would be able to make informed or safe decisions around food, particularly when left unattended.

What can you do to help?

It is important that all staff are aware and familiar with the child's dietary needs. It can sometimes be helpful to make other pupils aware of this too, providing you have parental permission. Anxiety can be reduced if food is kept out of sight and some form of food 'security' is in place. This might include keeping food storage areas locked, clearing empty plates away immediately after eating or ensuring lunch boxes are not left unattended. It is advised that you discuss any food within the classroom with the pupil's parents beforehand and that any food consumption for special occasions is arranged with them in advance. This can allow parents to 'borrow' calories from their evening meal. The pupil will need reassurance of his/her food routine which should include fixed times.



Emotional challenges

Some children with PWS have difficulty in controlling and understanding their emotions. You may find that emotional outbursts occur when the child feels anxious, frustrated, confused or that the situation is out of their control.

What can you do to help?

Ensure staff work together to reduce anxiety and to create positive and safe working environments. Reassure the child where possible and focus on achievements and goals.

Continued ...

What to expect from this child and how you can best support them (continued)

Anxiety

Most children with PWS suffer from some form of anxiety. Anxiety levels can increase when the child is faced with changes to their routine or when they are unable to predict the near future. At times, you may be able to see the child's anxiety growing with behaviours such as skin picking or fidgeting.

What can you do to help?

Ensure lines of communication between other staff and parents/carers are open in order to provide consistency and reassurance.

Be clear about rules and routines and try not to deviate from these. If a change occurs that is unavoidable, inform the child as early as possible and be prepared to confirm these changes as often as needed.

Perseveration or repeated questioning

Repeated questioning is a common feature of PWS, which usually occurs when the child requires reassurance or if they have been unable to process the information given. On occasions, the child may use repeated questioning to gain 1:1 attention and verbal responses.

What can you do to help?

Agree to answer the question just 3 times. On the 3rd occasion, ask them to repeat your answer back and confirm they have understood your response. If the child persists, try reducing verbal responses by shaking/nodding your head, or using visual reminders which they can refer to (eg. if the question is "what time is lunch?" try drawing a clock face on a paper plate to indicate your answer).

Time management

Some children with PWS struggle to move on from one activity to another, and you may find they are reluctant to move on until the task is completed. Most children with PWS are able to focus more effectively on several smaller tasks, as oppose to one large, on-going project.

What can you do to help?

Provide short activities and be clear on time frames or when you expect it to be completed. Ensure a gentle reminder is given as to how much time is remaining.

If the child insists they cannot move on until the task is complete, try issuing an "extension card", enabling them to borrow time which they can cash in later to complete the task.

Continued ...

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Quick tips for behaviour management

- Use positive language
- Keep verbal responses to a minimum during behaviour incidents
- Use humour where possible
- Appear disinterested in negative behaviour
- Be consistent with your responses as a whole staff team
- Use a structured discipline procedure
- Work collaboratively with parents/carers

What to expect from this child and how you can best support them (continued)



Day time sleepiness

Note that children with PWS can often suffer from disturbed sleeping patterns. It is likely that this child will grow tired throughout the day and may require frequent rest breaks.

What can you do to help?

Ensure you have a quiet space available where the child can go to rest. It may be that the child requires a short nap (approx. 10 mins), and this may be required more than once a day.

Rewards

Positive behaviour should always be rewarded as children with PWS respond well to praise. They are also likely to enjoy responsibility and being told they've "done a good job". Doing this can boost the child's self-esteem and confidence.

What can you do to help?

Most importantly, food should **never** be used as a reward for a child with PWS. It can be useful to record achievements and praise on a graph or star chart. You can make this available for the child to refer to and the visual representation will make this accessible to them. When rewarding the whole class with food items, ensure you have consulted parents beforehand. They may provide a low calorie alternative or make arrangements for the calories to be borrowed from their evening meal.



We have a useful LEAPS Framework available on request, which includes further guidance for staff. Contact us for a copy

Supporting this child

Many children with PWS will have an Education, Health and Care plan. In addition to the support outlined in this, we would always recommend some form of support during social times and when around food to avoid high levels of anxiety or food seeking behaviours.

Children with PWS respond well to 1:1 support, and often this will be a requirement of their EHC plan. If you do already have support in place, do bear in mind that it can be very easy for people with PWS to become attached to this person, particularly if they are together for the entire school day.

You may notice some obsessive behaviours surrounding this person. Whilst the consistency and reassurance of a familiar face can be beneficial, it is important to avoid the pupil taking 'ownership' of their support worker or TA. This can be easily overcome by the TA working with various pupils in the class-room, showing that he/she is not there solely for the pupil with PWS.

It is common for children with PWS to be fond of adult company and the child may need some support in socialising with peers. Encouraging them to take part in lunchtime clubs or activities is beneficial in aiding their social development.

You may find that the child will still require the support of a TA during these times, and it can often be useful to provide a quiet, 'safe' place if they become overwhelmed.

Supporting this child (continued)

Sometimes a Positive Support programme can help develop social skills and relationship skills. You may notice the pupil struggles with conversational skills such as turn taking, waiting for others to finish speaking or listening to others. In cases such as these, it can be useful to play turn taking games such as board games or role playing games.

It is important to listen carefully to the parent(s) of the PWS child as they will understand their child's individual needs and idiosyncracies better than anyone else. It may appropriate to have more regular meetings with the parents than with other children in your class. A diary recording notable events in the day may also benefit both parent and teacher.

Inclusion

Including a young child with PWS is relatively easy, given that they are sociable and affection in nature. As with all children, you should check that activities are appropriate to their ability and check that instructions have been understood. You may need to repeat Instructions or use signs to demonstrate.

It can sometimes be useful to educate the other children on the needs of this child, with the consent of his/her parents. This may mean using social stories or persona dolls. In doing this, you can reduce the chances of other children sharing food and can create a nurturing, safe environment.

An increasing number of children are receiving dual diagnosis of PWS and **autism**. If the child is also on the autistic apectrum, they are likely to need extra support in social situations and in play.

Special Medical concerns

There can be health complications in PWS so it is important to be aware of warning signs that something is wrong. **If this child vomits or complains of stomach pains it is imperative that this is taken seriously.** Children with PWS are unlikely to vomit at all and doing so **may** be a sign that something is wrong.

Due to their **high pain threshold**, any complaints of pain or discomfort should be taken seriously. Most children with PWS will only complain of this if the pain is extreme, and even then the person may only describe it as an ache or discomfort. It is important that staff are aware that bruising can occur easily, even as a result of a slight knock or bump.

Children with PWS also have **poor body thermostats** so this child may need extra support regarding appropriate dress and regulating temperature.

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Medicines

It is likely that the child will take medication, usually in the form of a growth hormone (GH) injection. GH has positive effects in terms of muscle tone and height and is to be taken on a daily basis.

It is more than likely that this will be administered in the home, but if you are asked to keep medication on site please seek advice from the child's consultant or visit <https://www.gov.uk/government/publications/supporting-pupils-at-school-with-medical-conditions--3>

Special Medical concerns (continued)

Some children with PWS will **skin pick**, which is usually a sign of anxiety. Skin picking requires careful monitoring to avoid infection and sores.

Sticky saliva is also common in children with PWS, and drinking plenty of fluids can help with the discomfort (a crusting around the mouth) as well as the difficulties caused by this when swallowing food.

Scoliosis is a common problem in PWS which causes a sideways curve to the spine. This can be very mild, requiring little or no treatment or, when more severe, can require bracing or surgery. If the child is wearing a brace, it is important that the back is kept as straight as possible at all times.

PWSA UK

Suite 4.4 , Litchurch
Plaza

Litchurch Lane
Derby DE24 8AA

T: 01332 365676

E: admin@pwsa.co.uk

W: www.pwsa.co.uk

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