PWSA UK
PRADER-WILLI SYNDROME ASSOCIATION UK

# Information for Nursery 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 30 minute YouTube video which gives more information about children with PWS in a nursery setting at https://www.youtube.com/watch?v=wEpfpl6O9xo**

**A child with Prader-Willi syndrome is joining your nursery**

**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?

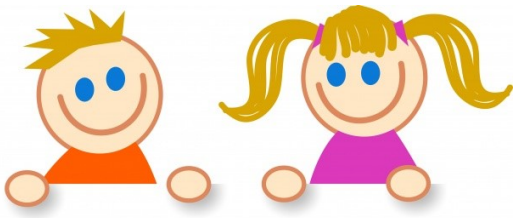
Most children with Prader-Willi Syndrome have an affectionate and friendly disposition. They respond well to humour and are often able to make others laugh. Although they are likely to have some level of learning disability, children with PWS are often skilled when it comes to creative work and puzzles. Although the child may have limited language, they are likely to be socially aware and enjoy the company of other children and adults.

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

### Developmental Milestones

Developing the skill of walking is usually later in children with PWS than we would expect to see in children without the syndrome. It varies from child to child, but usually takes place between 18 months to 3 years of age. The average age for sitting unaided is 12 months, but again this can vary from one child to the next.

Although not all children with PWS experience difficulty with speech and language, delayed speech development is common. Vocabulary may be minimal and the child may stick to certain topics, saying or asking the same thing over and over. The child may receive speech therapy, although it can also be useful to develop strategies to help the child express their needs. This could be in the form of signing or with the use of pictures, symbols or actions.



Toilet training can be a long and difficult task for children with PWS, and it is important that you do not assume that the child is able to toilet independently. If the child is no longer in nappies, it is likely they will still require support with personal care and hygiene.

Although some babies with PWS are tube fed due to their low muscle tone, children later develop a heightened interest in food between the ages of 2 and 7 years. This may manifest in food seeking behaviours, taking food without consent or a preoccupation with food. Many children with PWS enjoy talking about food and need reassurance regarding meals and times.

### How can you best support a child with weak muscle tone?

Hypotonia, or weak muscle tone, is present in all children with PWS. It is important that activities which focus on gross motor skills are used as this can help with core stability and muscle tone.

It is useful if you are able to incorporate physical activities into your daily routines in order to manage weight and increase muscle tone. This could be a gentle "wake up, shake up" session or just general play. Be aware that the child may require special equipment such as a standing frame or specialist seating.

## Information to be gained from Parents/carers

Before the child joins the nursery, it is important that you gain information from parents/carers about the individual needs to this child. This may include:

- How the child sits and stands (any specialist equipment?
- Does the child have input from a physiotherapist?
- Does the child crawl or walk?
- Is the child able to grasp objects?
- Does the child experience difficulty with speech, vision or hearing?
- Information about diet, as recommended by a consultant
- Which professionals does this child see? (be sure to gain contact details)
- Does the child tolerate touch, messiness on their hands, noise etc?

We would also advise that you ask parents/carers to describe how *they* feel PWS affects their child. This will give them an opportunity to voice their concerns and help to paint a picture of how this child functions on a day-to-day basis.

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

**Contact us for a copy**

## Adapting routines and environments

You may have some routines in place at your nursery that would be unsuitable for a child with PWS, such as rolling snacks or self service lunches.

A child with PWS should not have unlimited access to food and will find the availability of this food distracting and at times distressing. We would always suggest that food is only available at set times and is easily portioned.

Ideally, snacks should be in the form of fruit or vegetables or other low calorie snacks. If you are unsure about portion sizes or food items, always seek advice from parents. Where possible, the child should be supervised around food to prevent food seeking and promote positive eating habits. In young children with PWS, a verbal reminder is often enough to discourage negative behaviours.

A child with PWS is likely to grow tired throughout the day and may require rest breaks or naps. It is a good idea to ensure you have a quite space available for this.

**Information for Nursery staff**

## Developmental milestones

**Please note– these will vary from child to child, but generally speaking we would expect to see:**

**General alertness and responsiveness—6 months**

**Sitting unaided– 12 months**

**Walking—24 months**

**Speech—3 years**

**Heightened interest in food– 2 to 7 years**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## 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.

## 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

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.

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.