



pwsa AUSTRALIA

prader-willi syndrome association

Understanding Prader-Willi Syndrome – accessing the community.



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www.pws.org.au

Who is this guide for?

This guide is for service providers who assist someone with Prader-Willi syndrome (PWS) to access the community for daily living activities such as social access, leisure, recreation and sport. It will also help the understanding of any mainstream organisation which offers community activities, and has a PWS participant.

It can help to understand more about people with PWS and the impact they may have on you and on other people when you are supporting them.

What is Prader-Willi Syndrome?

PWS is a complex, multistage genetic disorder which affects multiple systems in the body. It significantly impacts behaviour, mental and physical health. People with PWS require cognitive, social, learning and health support throughout their lives. A person with PWS can live a healthy, fulfilling life when they have ongoing, consistent support from people who understand the intricacies of the syndrome.



Learn more about PWS
from the international
organisation

www.ipwso.org



What impact does a participant with PWS have on your role?

It's important to know that someone with PWS whose lifestyle is healthy, secure and purposeful is a joy to be with and has the potential to achieve great things!

The term 'duty of care' takes on new aspects when providing a safe and healthy environment for someone with PWS. Direct care service providers need to be diligent in a variety of ways and be aware of:

PWS traits	Impact and how you can help
Often present as very capable and willing to participate in activities	Abilities may be overestimated and more may be expected than they can deliver. As a result, inadequate support can be provided leading to a decline in the person's mood, health, safety and wellbeing.
Respond well to clear, simple instructions, consistent boundaries and positive feedback	Delivering instructions and feedback clearly and simply is likely to result in a cooperative, positive participant.
They often persevere on a subject or question, repeating it several times until directed to another topic or distracted by something else. This may present as telling great stories about themselves and their circumstances. They can expand stories, to the point of very plausible lies. Their anxiety and excitability may cause them to embellish reality.	Service providers need to be aware of the intellectual limitations. Otherwise the stories can lead to misunderstanding, disruption and possible failure. This behaviour can be an attempt to make a social connection but can put themselves and/or others at risk. They can't be relied on to recall, or accurately report events that have happened. Confirming reported "at risk" occurrences is essential, before acting.
Often have Asperger-like difficulties in social settings. They don't intend to be rude but are less aware of personal space and may speak loudly or butt in. They may persist on a topic of conversation when others have moved on to something else, or make dubious claims, causing listeners to avoid friendship.	Subtly modelling appropriate social interactions can help a PWS person learn how to act in a social situation.
The PWS brain takes longer to process what has been said to them, and to reply or act. Multi-faceted questions and multi-stepped tasks are confusing for people with PWS as they can 'get stuck' on the first step and appear to be stubborn or uncooperative. Generally have a poor short term memory.	Simple, direct questions or instructions can be more effective. They are capable of learning and applying rules and routines through consistent practice. The poor short term memory is often balanced by an excellent long term memory, especially for detail.
Generally not good at planning and organising. They might assure you they have packed everything needed for the day's outing, but still something critical may have been overlooked.	Find a subtle way to join the participant in 'double-checking' that they have everything. This is particularly important if there are several activities on one day.
Most people with PWS have a mild intellectual disability. They are 'concrete' thinkers and interpret literally what is said, due to reduced executive functions of the brain. It is not easy for them to understand abstract concepts, assess reality, make decisions, recognise consequences or switch tasks without much preparation.	They often need assistance to find a solution to a problem, or the correct way of doing something, as they are not good at problem solving.

PWS traits	Impact and how you can help
People with PWS have a heightened level of anxiety and excitability. Sudden changes in mood and temperament are common. They don't cope well with unexpected change, the thought of something new to their routine, or the expectation of accessing additional food or drink. Their social sensitivity is reduced so they can become loud and disruptive as a result of negative or positive responses to their feelings or perceptions.	Service providers need to be aware of these traits, particularly when organising events or celebrations. If emotions become heightened, the provider needs to have a strategy for reassuring the person.
They may demonstrate a poor concept of time. They can read a clock face but have little appreciation for the length of a day, a week or a month. Time in the day is generally related to meal times.	Service providers need to be very clear when needing time commitments because it may not be understood. Expressing how many times something occurs e.g. per week, is most effective if the relevant days are named. It may need to be written down.

Food security

People with PWS have a flaw in the part of their brain that determines hunger and satiety. This gives them a continuous urge to eat. It means they are constantly hungry and will seek food in the attempt to satisfy this hunger. Food, or the means of obtaining food, usually dominates their thoughts unless they are distracted or occupied with other activities. All people with PWS require a strictly limited energy intake to avoid ill health. Anyone dealing with someone with PWS needs to gently but consistently enforce this, unless agreed in advance with the caregivers.

It's important to be mindful of the significance of food and beverages in the presence of someone with PWS. Don't leave food visible or accessible to them as this will cause loss of concentration, increased anxiety and possible behavioural reactions. They may take food, irrespective of who the food belongs to, who has partially eaten the food or the quality of the food. Provide security around all food/beverage sources, including garbage bins, other people's food and money or credit cards. Someone with PWS doesn't see taking other people's money as 'stealing' but as an act of survival!

Impact on physical skills and ability

People with PWS have lowered muscle tone and muscle mass, reducing physical agility and speed. They may be unsteady on stairs, tire easily and often display 'daytime sleepiness'. They can be at greater risk of injury due to weaker muscles and less stable joints.

They have a very high pain threshold and rarely complain appropriately of real pain. However, they may exaggerate discomfort or express false pain in order to avoid something they don't like or as a means of obtaining more food. For example, if it is known that a trip to the hospital will result in more food than usual, they may express false pain to get that trip to the hospital. Reported pain must be taken seriously and investigated, to rule out underlying medical conditions.

Some people with PWS will pick open areas of their skin e.g. a mosquito bite, when stressed or bored. Unsightly, sores from constant picking can draw incorrect assumptions about abuse and mismanagement.

Determining how to dress for the weather is not easy for people with PWS due to impaired regulation of their body temperature. They can be at risk of hypothermia and hyperthermia so they need to be guided to dress appropriately and maintain good hydration. They can also present with a high fever despite the lack of infection or have a serious infection and no fever. Medical staff need to be aware of this as it can delay necessary treatment. It's important that a caregiver, house supervisor or ambulance is called if the person seems unwell.

How can you provide optimum service to someone with PWS?

Knowledge of PWS and the characteristics of the individual with PWS is vital. This, along with consistency of care, will maintain the safety and wellbeing of a person with PWS who attends community activities, whether specifically for the disabled or not.

COMMUNICATION

- Arrangements should be discussed with the person with PWS before they arrive. Welcome them with a calm and confident manner. Sit in a quiet place with minimal distractions to tell them about the place they have come to, the people they will meet and how things are done. Going through arrangements again will reassure them of consistency. Speak in simple short sentences; allow enough time for the person to think and respond. Do not give or ask for too much information at one time. Give them time to peruse the new environment and walk with them to show them around or introduce them to new people.
- Break any multi-faceted questions or explanations, and multi-stepped directions or tasks, into a series of single requests or statements. Remain positive and friendly but clear when giving instructions or directions. Identify a person of higher authority to whom the person with PWS can refer to for boundaries and confirmations, and to whom others can deflect to for decisions, in all their environments. Be specific in your questions or requests and prime them with the reason first – for example “The game is about to start. You need to put your boots on now.”
- Definite guidelines and boundaries help people with PWS to feel secure and not become overwhelmed or anxious. Explain rules and protocols simply and ask the person with PWS to repeat back to you what you have said. Regularly confirm rules and protocols and social mores (such as greetings). Although the words are understood the associated importance is often missed. Visual signs, pictures of a process or actions are understood more easily than words and can be effective to use as prompts for desired actions and references for confirmation.
- Include a parent, guardian or caregiver if requiring accurate or important information from, or to be given to, the person with PWS.
- Be aware of the person's physical capabilities. Short rests may be needed at regular intervals until the person is competent in the activity, if participating in sport or an unaccustomed physical activity. However, they may try to give up early, so it can help to motivate them with a ‘pep talk’.
- Try, at all times, to prevent the person with PWS being exposed to, or distracted by food or beverages that do not belong to them. Do not offer them food or beverages unless organised with caregivers. Most people with PWS will bring their own food/beverages with them, to maintain their strict diet routine, but are rarely able to resist the urge to eat or drink other food if accessible. Walk with them if they are passing areas of food preparation or food availability. Sit with them when a snack or meal break occurs to prevent food seeking. Try to provide companionship and distraction, not supervision.

BEHAVIOUR MANAGEMENT

- When possible, always prepare the person with PWS for change. If there is to be a change in the activity schedule, and it is known ahead of time, tell the person with PWS what change will occur several times before it happens. It helps to suggest that you “will need their help because...” before explaining about the change. This will appeal to their desire for responsibility and assist with their acceptance of the change. Ending the explanation with “will you be OK with this?” or “thanks so much for being OK about this” is a positive acknowledgement of their ability to be responsible or mature about change.
- If possible, try to offer the person with PWS choice, to give them a sense of control as well as to direct their thinking. Choice must be appropriate to the person's needs and ability and is best limited to two options.
For example: “would you like to sit on this chair or the bench, while you are waiting for your turn?”
- Check all stories, alleged food allowances or accusations with a parent or person responsible, before acting upon the information. If the information from the person with PWS requires action, confirm it with their responsible adult first. For example, they may tell you that they forgot to bring their lunch with them. Before providing food or money to buy food, check with their family or caregiver to ensure you won't be supplying a second lunch.
- Providers may need to facilitate appropriate social interactions for the participant with PWS. Modelling how to initiate, sustain and close a conversation or social encounter will help the participant learn and become more self-sufficient in social situations.
- If a problem arises, ask for their assistance in working out solutions to the problem. They do not like to be told what they are doing “is wrong and should be done this way”, but they will respond to “acquired agreement”.
For example: “Let's look at this together to work out what's not going well. What do you think?”

MEDICAL

- When a person with PWS sustains a fall or accident that may have caused injury, have the person thoroughly checked for such an injury. Do not rely on their self-reported pain or lack of pain. Be aware of their movement and behaviour after an incident. If they need to attend a medical service, have the Medical Alerts Booklet accompany them so the medical professional can be informed about PWS.
- If skin picking is happening remind the person with PWS that it's important to cover the area before leaving the house to prevent infection and resist further picking.
- Encourage people with PWS to respond to weather conditions as you would. If it's cold remind them to take or put a jacket on. If it's hot prompt them to take their jacket or jumper off.
- Diabetes is not uncommon in people with PWS. If the participant is diabetic, they may need reassurance that you are ready to assist if a diabetic event occurs. Some people are over-anxious about their diabetes and may ask for food. Conduct the skin prick test to provide objective information about what is needed (just in case it's an excuse to get food).
- Try to always communicate with a person with PWS in a positive calm manner. They respond poorly to raised voices, demands or threats of any kind. If you become anxious it heightens their level of anxiety and reduces their ability to act appropriately or cooperate. Aim to acquire their agreement to what is suggested.

Helpful Tips

People with PWS enjoy humour, being praised, and given (appropriate) responsibility!



When someone with PWS starts to become upset, keep calm and maintain a lowered but confident voice. It can help to have a pre-identified place for people to go to when they feel stressed or upset, to remove them from the immediate public area.



When accessing the community be sure to maintain companionship or an awareness of the person with PWS and direct them to the task at hand. One to one support is best when near places of food access, to ensure no unwarranted or inappropriate food access.



Deflect from an anxiety provoking subject or situation by highlighting something positive for the person or redirect to another task or topic.



Prevention is always the key to success!



If you are an organisation that offers mainstream activities to the general community, you can contact the NDIS about participation by a disabled person. To learn about a person with PWS, speak to their caregiver before they start your activities.

Further information

Video: Can't Stop Eating – UK documentary

<https://www.youtube.com/watch?v=dKCL2O0NfXg>

How you can help – for anyone interacting with a person who has PWS:

<http://pwc.org/wp-content/uploads/sites/18/2015/10/Brochure-Someone-You-Know-Helpful-Tips.pdf>

The International 'Standards of Care & Best Practice Guidelines for Prader-Willi Syndrome'

<http://www.ipwso.org/best-practice-guidelines-for-pws-care>

Confabulation in Prader-Willi Syndrome (IPWSO)

http://media.wix.com/ugd/a71d4c_a1faf165b293412d8620e5fe87450467.docx?dn=Confabulation%20in%20PWS.docx

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