



pwsa VICTORIA

prader-willi syndrome association

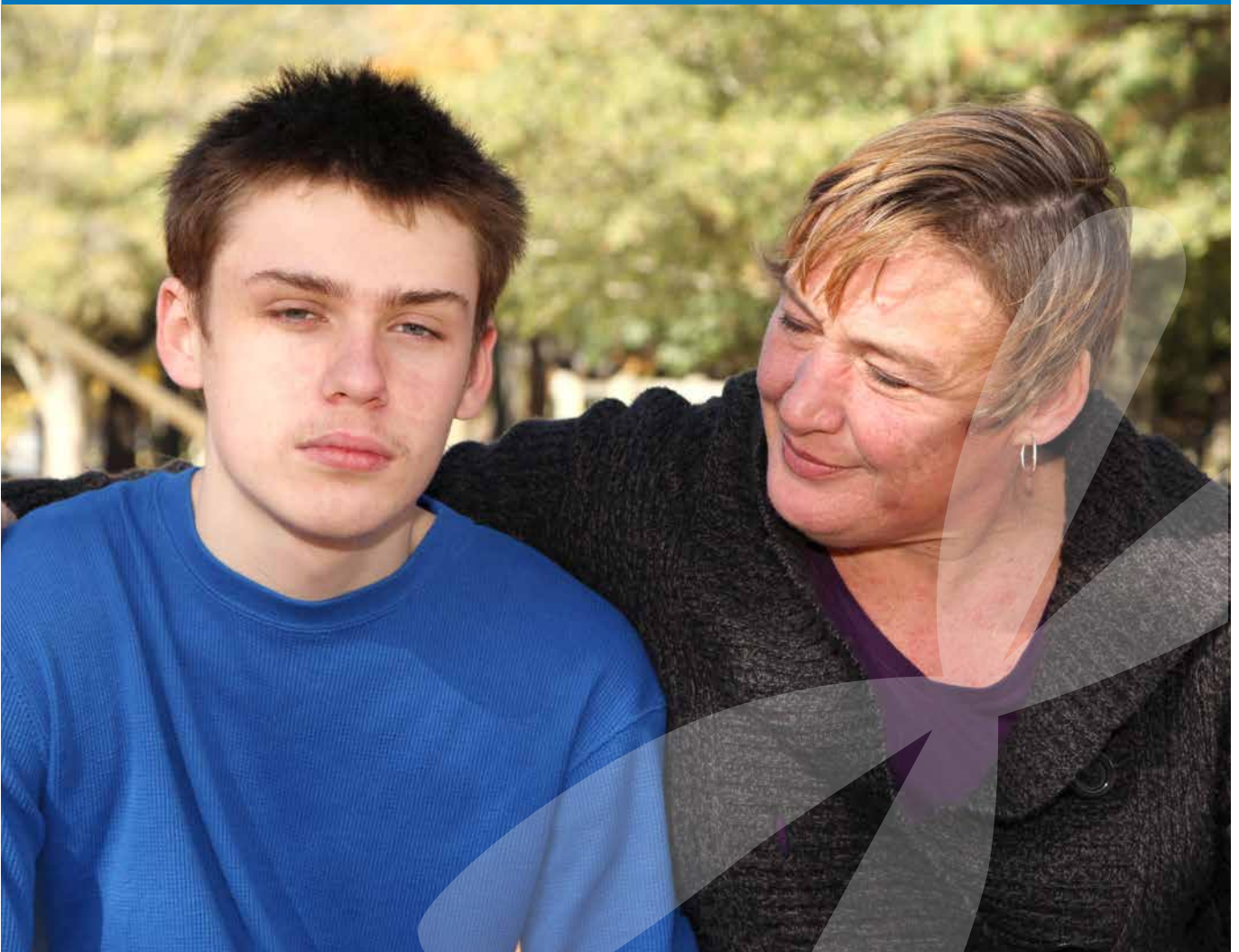
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IMPORTANT INFORMATION FOR PARAMEDICS

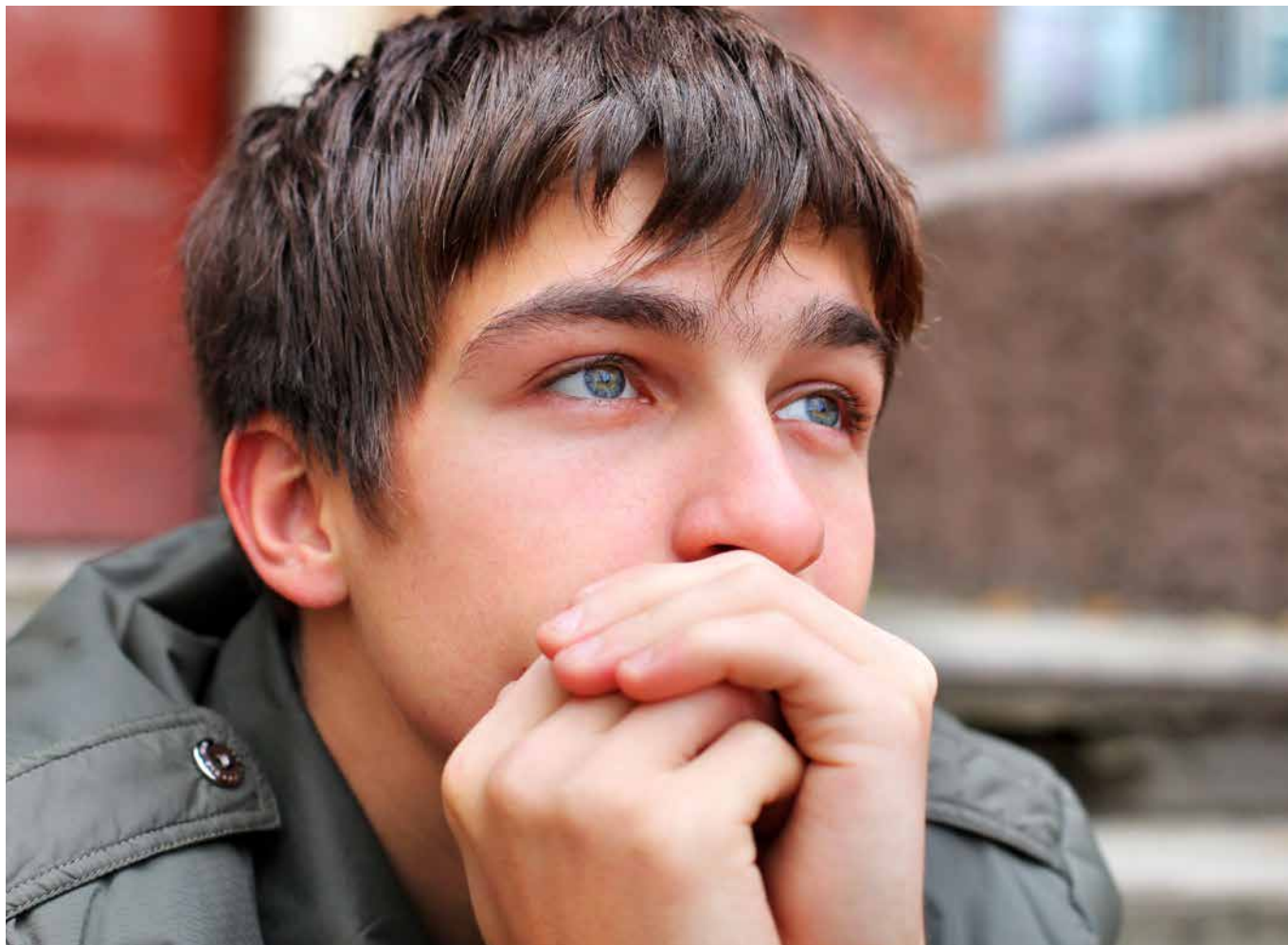
Dealing with a person who has Prader-Willi Syndrome



The purpose of this guide is to give the Paramedics information about the person who has Prader-Willi Syndrome (PWS).

The guide explains key characteristics of the disability, as they may be relevant when called to a medical incident.

This guide also gives information about how to support a person with this disability in times of high anxiety, when the person may be distressed, confused or obstructive.



PWS is a rare disability. Most Paramedics would not have heard of PWS. PWS has complex medical, behavioural, and emotional characteristics. This information will help you to better understand a situation when dealing with a person who has PWS.

PWS fundamentally, severely impairs a person's ability to self-regulate critical functions such as appetite, emotion, and temper response. It can create cognitive impairments as well. Therefore, your role as a professional is critical in helping a person with PWS manage themselves in a crisis, with minimal harm to self and others.¹

People with PWS typically have a mild intellectual disability, and even though some can speak well, they don't necessarily understand you. Some have a mental illness as well. They may have called the ambulance for a reason that is misunderstood.

Our goal is to help you understand the cause of PWS behaviours and put them into context. Then you can offer the support or medical assistance they need when the ambulance is called. Some understanding of PWS will increase the chance of a positive outcome and decrease the chances of the incident escalating.

1. PWSAUSA, To Law Enforcement Personnel Regarding Adults with PWS.

What approach works best?

PWS is a spectrum disability so each person who has PWS will present differently, with different degrees of cognition and impairment.

Even though there will be a wide range of individual abilities and personalities in the PWS population, these tips will help with how best to respond to how a person with PWS typically thinks and reacts.

- ▶ **Identify who you are and why you are there.** Please remember a person with PWS needs your help to explain clearly what is happening and why you are involved.
- ▶ **Check to see if there is a responsible adult available, with good knowledge of PWS, when you speak with the person with PWS, whenever possible.**
- ▶ **Keep calm and be consistent when communicating with the person with PWS,** particularly if the person is upset or agitated. This will help to de-escalate the situation. **Loud voices will only escalate the situation** as will threats and accusations. Negative approaches typically are not effective for people with PWS and can significantly worsen the situation.

They respond well to quiet, calm, confident speech and a calm manner is more likely to encourage their cooperation rather than lead to an escalation of oppositional attitude and non-compliance.²

- ▶ **Avoid arguments** - you will never win an argument with someone with PWS.
- ▶ **Use short sentences in easy-to-understand language.**
Most people with PWS have poor auditory processing and memory skills. **Just because you have given an instruction do not assume that message has been understood – always CHECK.**

Repeat or restate instructions to make sure they understand what you have said. Repeat or restate the instruction if you are unsure.

- ▶ **They respond well to limited, controlled choices. ie. 'either/or' choices.**
- ▶ In some circumstances, the **use of distraction and humour** is most effective.
- ▶ **Do not make promises you cannot or will not keep** – if what is promised is not delivered, the chances of a re-escalation of behaviours increases exponentially.
- ▶ **Avoid offering food or drinks** because it sets an expectation that more will be provided.
- ▶ **Do not offer them food as a means to placate or calm the person with PWS.**
- ▶ Unless necessary for the safety of the person or those around them **avoid physical contact or using restraints of any kind.** People with PWS will often react negatively if they feel backed into a corner and this can result in physical and verbal aggression and other destructive behaviours.
- ▶ **Physical restraint may cause breathing problems and injury without complaint, as people with PWS have a high pain threshold and often, poor circulatory and respiratory health.³**

2. Holland A Loughnan G, Law Enforcement Matters - Regarding People with Prader-Willi Syndrome www.ipwso.org/law-enforcement

3. Holland A Loughnan G, Law Enforcement Matters - Regarding People with Prader-Willi Syndrome www.ipwso.org/law-enforcement

Safety first always

- ▶ **Individuals with PWS are unable to accurately monitor their own health and wellbeing.**
- ▶ **Note that uncharacteristic or rapid changes to behaviours are often the first indicator of being unwell — In the absence of vomiting, high pain tolerance & absence of high-temperature readings, changes to behaviour and high fatigue levels may be the only warnings of pending illness.**
- ▶ **Seek the advice of the parent/support worker if possible or seek medical assistance immediately.**⁴
- ▶ **Do not try to talk them down or expect them to respond logically** — As a rule, in moments of PWS temper, reasoning does not work.
- ▶ **Once the person with PWS has obtained food do not take it off them. Best practice advice is to never take food away. It will only tend to escalate behaviours.**

Seek more information from support person

- ▶ **If a caregiver or parent is present, before directly addressing the person with PWS, ask the caregiver/parent if they have suggestions for the most effective way for you to approach the person.**
- ▶ **Seek confirmation of the parent/support worker if possible, about their mental health history.**

Hospital admission

Tips to facilitate successful handover to hospital staff
if the person with PWS requires hospital admission:



- ▶ **Refer to PWS Medical Alert Booklet to inform the Emergency Department staff of support that is required (see QR code for details).**
- ▶ **If possible, organise with carers or support staff to bring items to assist with stress management to the hospital i.e. noise-cancelling headphones, iPad, or soft toys.**

REFERENCES

PWSAUSA, To: Law Enforcement Personnel Regarding Adults with PWS, The Gathered View (ISSN 1077-9965), published bi-monthly by the Prader-Willi Syndrome Association (USA). Prader-Willi Syndrome Association (USA), 8588 Potter Park Drive, Suite 500, Sarasota Re-printed from, Florida 34238 800- 926-4797

PWS Medical Alerts Booklet www.pws.org.au

Holland A Loughnan G, Law Enforcement Matters - Regarding People with Prader-Willi Syndrome www.ipwso.org/law-enforcement

Foundation for Prader-Willi Research: Serious Mental Health Problems in PWS, www.fpwr.org/serious-mental-health-problems-in-prader-willi-syndrome

4. PWS Medical Alert Booklet www.pws.org.au

PWS SNAPSHOT

Prader-Willi Syndrome (PWS) is a complex genetic disorder.⁵

- Most people with PWS have a mild or moderate **intellectual disability** with associated learning difficulties, behavioural and **mental health** problems.
- There may also be some **physical, sensory, and receptive/expressive language impairment**.
- People with PWS **lack satiety**. They are always hungry. There is often **abnormal food-seeking behaviour**.
- People with PWS have difficulty managing **anxiety levels** and are very **stress-sensitive**.
- **Confabulation (storytelling and fantasizing) and impulsivity** are common PWS behaviours.
- **Uncooperative behaviour may quickly escalate to major temper outbursts** if not managed appropriately. **Seek the advice of parent/carer** regarding the behaviour and medical management and strategies likely to achieve maximum cooperation.
- Most people with PWS have a **high pain threshold** and **poor temperature regulation**.
- Most people with PWS are hypotonic with **poor muscle tone and coordination**.

Things to be aware of:

- High risk of **complications of obesity** including high blood pressure and diabetes.
- High risk of **obstructive sleep apnoea** and other sleep disorders.
- Risk of **undetected infections, injuries, and other medical conditions** due to a **high threshold of pain and poor body temperature regulation**.
- Presentation of **abdominal pain** can suggest a very serious underlying condition such as a **stomach tear and ruptures, severe constipation, and bowel obstruction**.
- **Vomiting rarely occurs** in those with PWS. As emetics are ineffective, hospital care should be sought in cases of **suspected poisoning**.
- May be **habitual skin pickers** and prone to skin pick wound sites. They also tend to **bruise readily**.
- May have an **adverse reaction** to standard doses of anaesthetic and to standard doses of **medication** especially those that may cause sedation.
- Are susceptible to excessive fluid intake and are at risk from **water intoxication following the use of medication with antidiuretic effects**.

FOR MORE INFORMATION PLEASE CONTACT:

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