

Adapted from the PWS Admissions Policy of Royal Prince Alfred Hospital, Sydney, Aust.

**Introductory notes to accompany
on presentation at the hospital emergency department, or for any admission.**

Known medical conditions:

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Known allergies:

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GP:

Parents:

Public Guardian:

SUPPORT ORGANISATION CONTACT:

Prader-Willi Syndrome Australia

T: 1800 797287 1800 pwsaus | W: www.pws.org.au | E: info@pws.asn.au

Prader-Willi Syndrome Inpatient Management

Prader-Willi syndrome (PWS) is a complex multistage genetic disorder that results in a malfunctioning of the hypothalamus.

Most people with PWS have a mild or moderate intellectual disability with specific cognitive problems. They are also socially and emotionally developmentally delayed. They are hypotonic, shorter in stature, have low growth hormone and low gonadotrophin levels. They have a specific phenotype and have **no satiety**.

They are therefore always hungry, suffer from hyperphagia and readily gain weight.

It's important to note that **vomiting rarely occurs** in those with PWS.

People with PWS may search for food from any source – other people's lockers, meal trays, kitchens, garbage bins, cafeterias, tea lady's etc. They may even steal money to purchase food.

Due to their **cognitive problems** - poor sequential processing, concrete thinking and resistance to change, they may present as stubborn, demanding, generally uncooperative and at times aggressive. **Problem behaviours can be minimized by:**

- 1) Preparing the PWS patient for new things, or new people who will be doing things to them;
- 2) Giving them time to fully understand what is happening (or going to happen to them) or what they are being told;

- 3) Keeping instructions, the same from person to person who may be dealing with them (even slight or insignificant differences can cause a reaction). If something is done one way the first time they will expect it to be done the same way next time, unless it has been fully explained to them that it will be different or done by someone else – *before* the event.
- 4) Being clear, concise and consistent in *all* interactions with the PWS patient

People with PWS have difficulty managing **anxiety** levels and are very **stress sensitive**. Often increased anxiety levels can cause self-injurious behaviours to escalate (eg skin picking).

People with PWS have a high pain threshold but often exhibit a child-like fear of pain – such as for injections. If this is the case with the patient, the suggestion of a pain relieving gel may help. KY gel may even do the job!

Uncooperative behaviour may escalate into major temper tantrums if they are not managed appropriately. This can be distressing for all involved and may take some time to diffuse. They respond well to quiet, calm, confident speech that endears their cooperation rather than opposes their apprehension or oppositional attitude. They respond well to limited, controlled choices. They respond well to distraction and humour.

Many people with PWS may also have **mental health issues**. Seek the advice of parents/carers regarding previous medical management and strategies for gaining maximum patient cooperation.

On presentation to the Emergency Department or Admissions:

- It is suggested that **people with PWS be given as much supervision as possible while admitted to hospital**. This may require a “special” nurse being appointed to their care and/or where possible a single room.
- They **require low fat meals and no more than 8kcal per cm of height per day**. Please do not offer a standard menu unless there is an appropriate support person present. This will avoid false expectations about food choices, increased anxiety and a significant possibility of arguments or a breakdown in behaviour.
- People with PWS are **prone to confabulation** (story telling). Therefore, a responsible **carer/parent is required to be present** for:
 - an accurate patient history
 - the explanation of care plans, making future appointments,
 - any important information for the client

The PWS Medical Alerts booklet should be presented.

If not, please go to this website www.pws.org.au for important information about complications that are unique to Prader-Willi Syndrome.

Refer to the following documents for additional information on PWS:

PWS - A Primer for Clinicians' <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3217845/>

'PWS – A Primer for Psychiatrists'

[http://www.pittsburghpartnership.com/handouts/Pittsburgh%20Partnership%20Psychiatrists%20Primer%20for%20Care%20of%20PWS%20\(2\).pdf](http://www.pittsburghpartnership.com/handouts/Pittsburgh%20Partnership%20Psychiatrists%20Primer%20for%20Care%20of%20PWS%20(2).pdf)

<http://www.ipwso.org/for-medical-professionals>