

IMPORTANT PRADER-WILLI SYNDROME CONSIDERATIONS

PWS is a complex neurodevelopmental disorder affecting multiple body systems. Knowledge may be limited due to rarity - experienced specialist advice can be helpful. Useful resource: **RareCAP.org** (RD Clinical Activity Protocol)

1.

PWS affects the endocrine system and causes dysregulation of many hormones. These include growth hormone, oestrogen/testosterone, thyroxine (sometimes), cortisol, insulin and other metabolic / appetite-regulating hormones, and vasopressin.

 - Contact and consult the endocrinologist on call, or patient’s endocrinologist.
2.

Gastro-intestinal issues are common in PWS, including constipation, impaction, gastroparesis, obstruction, and infection. Risk of perforation, rupture & necrosis, especially after a food binge. Suspect a binge, even if denied. Evaluate any abdominal distension or pain, appetite loss, or vomiting (less likely / poor emetic reflex.)

 - Use the IPWSO evaluation chart for GI symptoms and/or a suspected food binge. (Inside Medical Alerts booklet or online: www.pws.org.nz/wp-content/uploads/2018/05/GI-algorithm-chart-ipwso-pwsausa.pdf)
3.

Mental illness presents atypically in PWS. Psychosis is believed to be more common, especially in the mUPD genetic subtype. Symptoms can present as cycloid psychosis with acute onset. Mood disorders are also more prevalent, although highs and lows likely to present atypically with true mania rarely seen. However, delirium is also thought to be common in PWS and difficult to distinguish from mental illness.

 - Consider delirium first during evaluation to exclude a medical or medication-related cause.
 - Be aware of atypical responses to medications, especially those with sedative effects. Lower doses are usually required. Side effects more likely. Hyperglycaemia risk. Water intoxication risks if antidiuretic.
4.

Unusual reactions to anaesthesia are possible – prolonged and exaggerated responses. A patient with PWS may also have health issues that alter the course of anaesthesia. Monitor closely during and post anaesthesia.

 - Considerations include: hypotonia, thick saliva/airway management, temp instability, assume food in stomach even if denied, obesity issues, apnoea, scoliosis, difficult IV access, possible GHD and CAI.
5.

Other important health information for consideration:

 - High pain threshold** – may mask serious illness, infection or injury. May be unable to localise pain.
 - Temperature instability** – Consider normal body temp may be low and/or may not be raised when unwell.
Body temp may also be higher than expected. Hypothermia / hyperthermia risk.
 - Dysphagia & choking risk** – Poor oral/motor skills, emetic & gag reflexes. Hypotonia, reduced/thick saliva.
 - Increased risks for** - glucose intolerance, ketoacidosis, sepsis, pulmonary embolism, oedema, seizure.

EMERGENCY ADMISSION FOR PATIENTS WITH PRADER-WILLI SYNDROME



PERSONAL INFORMATION

My name is

My date of birth is

My ethnicity is

My address is

I live

☐

with family / whānau

☐

in a home supported by a community support service

I make my own health choices

☐

Yes

☐

No

☐

with support

The main person who supports me with healthcare

Their contact details

(Family / WGuardian / SP)

Other important people to contact when I get healthcare:

Please circle (SP = support person)

Name

Ph

(Family / WGuardian / SP)

Name

Ph

(Family / WGuardian / SP)

My NHI number

My endocrinologist is

My GP is

GP Practice

Other specialists I see (i.e. paediatrician, orthopaedist, respiratory physician, psychiatrist, physiotherapist)

Name

Specialism

Name

Specialism

Name

Specialism

COMMUNICATING WITH ME - PWS causes auditory processing and working memory difficulties.

☐

Please tell me one thing at a time, give me a few extra seconds to process it, and check I understand.

☐

Please give me time to think before I answer.

☐

I understand better when people speak clearly and use clear language.

Here are some other ways I like to communicate or ways you can support me:

MY SPECIAL DIET – PWS means that dietary intake usually needs to be reduced by 20-40% and it is recommended to follow a reduced carbohydrate diet and avoid sweet foods. PWS also causes hyperphagia which means that someone with PWS may be preoccupied with thoughts of food, overriding all other thoughts or concerns. Individuals living with PWS may go to extreme lengths to seek or obtain food that will often surprise you. Providing ‘food security’ helps - reassurance about upcoming meals (without false promises) and reducing stress by removing visible food and opportunities to access food.

If there is anything else you need to know about my special diet, or any allergies, here is some information:

MY LIKES AND DISLIKES – PWS is associated with anxiety, stress sensitivity, and emotional dysregulation. Here are some tips about what might make me anxious or upset in a hospital, and what I like that helps me.

MEDICAL INFORMATION

OTHER DIAGNOSES? I live with PWS and other diagnoses listed below (e.g., ASD, ADHD, gastroparesis, T2DM):

MEDICAL HISTORY – I have or have had the following health or mental health issues that you should be aware of:

MY MEDICINES

Medication	Dose	How often

Date updated _____