Who is this guide for?

This guide is for NDIA Technical Advisory Team members who are providing advice during planning, or reviewing a plan for a person with Prader-Willi Syndrome (PWS). This could be an initial or subsequent plan, or managing an appeal.

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

The traits which affect someone with PWS are cognitive, behavioural, emotional and physical. There is a high risk of underestimating the difficulties experienced by someone with PWS. This can lead to inadequate service support and a decline in the participant’s health, safety and wellbeing.

If a plan for someone with PWS is to be successful it needs to allow for a holistic and coordinated approach, so that the intervention of one support does not undermine the efforts of another.

PWS is uncommon, affecting about 1 in 14,000 people in Australia. To assess a plan for someone with PWS effectively, educate yourself about the many facets of this disability prior to the assessment. Gaining a comprehensive understanding of PWS support needs will help ensure consistent plan design and decision making for these participants, across the NDIS.

While PWS is a ‘spectrum’ disorder, there are a set of common characteristics. People with PWS have chronic behaviour disturbance and can be argumentative, aggressive and destructive. During the teenage years and into adulthood, lying in a plausible way and exaggerating (confabulation) increases, as does manipulation. Due to cognitive impairment, they are impulsive and have poor executive brain function, often leading to very poor decision making. When they are in a state of anxiety, they can present a risk to themselves and others around them. Their level of support needed can vary from month to month. Therefore, their plans may be at high risk of needing more frequent revisions than average. Evidence about the complexity of PWS is in Appendix 1.

The support of a person with PWS will include many ‘hidden’ costs. The extra administrative costs are likely to revolve around challenging behaviour (eg refusing to attend a medical appointment, causing a repeat booking, getting involved with the police which staff have to negotiate, causing property damage for which staff have to make insurance claims and organize repairs, etc.). Restrictive practices related to locking up food sources must be managed diligently.1,2 Research from Anglicare3 provides case studies and highlights the hidden costs in supporting people with challenging behaviours, including those making food demands.

How will you provide optimum service to a person with PWS?

- Preparation is essential to understand how the idiosyncrasies of this syndrome make it so complex. PWS is not like other intellectual disabilities. See Appendix 1 for further information.
- Talk to the person’s family, guardian or carer privately, before the assessment is concluded.
- People with PWS can be talkative. Make sure that information supplied by the person with PWS, upon which decisions are being made, is corroborated by another party.
- If you are meeting the person with PWS, make sure you have read the PWSAA ‘NDIS planning for people with Prader-Willi Syndrome’ Guideline. This explains how to interact with the participant. They typically have auditory processing difficulties so a phone call is unlikely to provide reliable information.
- People with PWS are prone to premature mortality, so plans should include supported accommodation.

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Tips

Before meeting someone with PWS, read the guideline PWSAA ‘NDIS planning for people with Prader-Willi Syndrome’, which explains their communication style.

There are a number of Guidelines about how service providers can best support people with PWS in the Australian context. These will give further insight to the complexity of the condition and the support focus. The Guidelines are on the Prader-Willi Syndrome Association of Australia website, www.pws.org.au.

Check if there is an Administrator in place for the participant. NDIS payments should not be made directly to a person with PWS as they are at risk of buying and eating inappropriate amounts of food. A food binge episode leaves a person with PWS highly vulnerable to dying as a result of necrosis of the stomach.4, 5

Further information

The International ‘Standards of Care & Best Practice Guidelines for Prader-Willi Syndrome’
http://www.ipwso.org/#/best-practice-guidelines-for-pws-care/c1jq

Confabulation in Prader-Willi Syndrome (IPWSO)
http://media.wix.com/ugd/a71d4c_a11faf165b293412d8620e5fe87450467.docx?dn=Confabulation%20in%20PWS.docx

Video: Can’t Stop Eating – UK documentary https://www.youtube.com/watch?v=dKCL2Q0NfXg

Video: My Deadly Appetite: Prader-Willi Syndrome - US documentary https://www.youtube.com/watch?v=ItCFF1Nal8c

IPWSO – Latest Scientific Papers and Research list http://www.ipwso.org/#/news/c21kz


Appendix 1: Complexity of Prader-Willi Syndrome – a summary of indicative evidence from medical specialists

CHALLENGING BEHAVIOR – A SUMMARY OF INSIGHTS FROM RESEARCHERS AND SPECIALISTS

- There is “a typical neurobehavioral profile that includes altered intellectual functioning and centrally driven maladaptive behaviors” and a “commonality of hoarding, cognitive rigidity along with the need for sameness, temper outbursts and emotional lability, repetitive and perseverative behaviors”. Also found was “cognitive dysfunction, social and emotional immaturity” along with “manipulative and sometimes illegal behaviors designed to acquire food” and “increasing oppositional tendencies.”

- “Many individuals also have specific emotional and learning disabilities which can impact on behaviour… evident even where the person is functioning at a relatively high intellectual level.”

- “Individuals with PWS typically have intellectual disabilities (borderline to mild/moderate mental retardation) and exhibit a higher overall behavior disturbance compared to individuals with similar intellectual disability.”

- “People with PWS are more prone to temper outbursts than people without PWS at similar levels of development.”

- There is “…a striking autistic-like behavioral phenotype in the majority of the PWS individuals, particularly deficits in the quality of language and communication and of imagination and interests.”

- “…the most common error is to underestimate the complexity of the disorder…”

- “… staff supporting someone with Prader-Willi Syndrome will need to have a good understanding of the disability in order to keep the person safe and respond to their highly specialised needs.”

SPECIALISED ACCOMMODATION SUPPORT

- If food access is left unmanaged, people with PWS die younger, and have a higher mortality rate than other (matched) intellectually disabled people, due to obesity factors.

- In relation to the impact of specialised accommodation for people with PWS, research has found a significant and beneficial impact on resident health, with a reduction in weight and diabetes medication.

- “For adults with PWS, one successful living situation for behaviour and weight management is a group home specially designated for individuals with PWS. Affected individuals generally require a sheltered employment environment.”

HEALTH

- The typical health issues for a person with PWS (who may be non-compliant with treatment) have been highlighted by the International PWS Organisation:

  “Striving for food combined with a lower than normal calorie requirement and, frequently decreased activity related to the hypotonia, will result in rapid weight gain if not controlled externally, leading to obesity and eventually the consequences of obesity: respiratory difficulties, heart disease, diabetes, and other problems.”

- IPWSO reports other common issues including back and joint problems, oedema (fluid retention), dry skin (picking, sensitive), osteoporosis, stomach and bowel problems, sleep problems, unexpected reactions to some medications, temperature instability and sensitivity, increased risk of psychiatric behaviours, hormone deficiency and dental problems. The symptoms of these conditions can exacerbate the anxiety of a person with PWS, and the anxiety can exacerbate the physical manifestations.

The 2015 Australasian PWS covered many facets of the disorder. The conference papers index, links to session videos on the ‘PWSA Vic’ Youtube channel: https://www.youtube.com/channel/UCNoz8qMUCB3E_AjUDSlvvyKg

NB. Footnotes on following page
Assessing NDIS participants with Prader-Willi Syndrome


17. Description of PWS clinical features http://www.ipwso.org/#!what-is-prader-willi-syndrome/clxd