PRADER-WILLI SYNDROME ASSOCIATION NEW ZEALAND
ADVOCACY · EDUCATION · SUPPORT

3rd March 2023

Review of Adult Decision-Making Capacity Law
Submission by the Prader-Willi Syndrome Association NZ

The PWSA is pleased that decision-making law is being reviewed and hope that it will become more reflective of both changing attitudes toward disability and an improved understanding of decision-making ability and limitations. The PWSA agrees with the guiding principles of the review.

The focus of our submission is how changes might specifically impact adults living with Prader-Willi syndrome. We do not know of any adults with PWS in New Zealand who manage to live completely independently, and they generally live in supported living accommodation supported by a service provider, or at home supported by their family. By nature of the syndrome, adults with PWS have affected decision-making and either have informal supported decision-making arrangements in place or have welfare guardianship court orders in place.

The preliminary issues paper states that court orders allowing someone to make decisions for a person can be put in place if a person is assessed as not having decision-making capacity. However, decision-making capacity is far more complex in PWS – there are some areas specific to PWS where a person does not have that capacity, but other areas in which capacity may not be affected.

The specific area of concern is decisions related to food and food environments. This concern also extends to uncontrolled access to money which can be used to obtain food. Hyperphagia is a key symptom of PWS for which there are no treatments. A person living with PWS needs a carb and calorie restricted diet in order to maintain a healthy weight because their altered body composition affects their metabolism, meaning weight gain can happen rapidly. Their low muscle tone (hypotonia) also makes excess weight very difficult to lose. It is a particularly cruel condition because coupled with the need for a restricted diet, they also have an altered appetite in which satiety is affected. Affected hormones and brain chemistry create increased feelings of hunger and altered food reward signalling which can drive them to seek food with great intensity. This can be extremely dangerous and lead to life-threatening situations. It can also lead to death if a person has uncontrolled access to food and experiences life-threatening gastric inflammation, stomach rupture or necrosis. Sadly, we know of a confirmed case in New Zealand where this has happened to an adult who was living in residential care.

The definition given to assess whether a person has decision-making capacity is “do they understand the nature and consequences of their decisions and can communicate the decision made?” In Prader-Willi syndrome, many adults would understand the nature and consequences of decisions regarding food. They are often aware of their dietary needs, the reasons for them and have grown up following dietary plans and having controlled access to food. Many people with PWS have the best of intentions

to have self-control over their food intake and can be very convincing that they will be able to make the right choices around food. However, hyperphagia is a biologically driven behaviour and despite understanding the nature of potential consequences of a decision regarding food, they cannot reliably make safe decisions in this area.

What we often see in New Zealand is adults with PWS leaving home and moving into a supported living situation and then gaining weight quite rapidly. This can be because the providers do not fully understand the controls and safeguards necessary in this syndrome, because there was insufficient preparation or there are inappropriate safety systems in place. We are also seeing adults with PWS finding it harder to obtain places in supported living accommodation, partly due to current staffing limitations, but also because service providers can be reluctant to accept a client with PWS either because they do not want to (food security may not work with other clients in the house), or they are unable to (cannot have a locked kitchen or the risks are deemed too high for the support they can offer). Unfortunately, there are very few PWS specific living options in New Zealand and Enabling Good Lives is not set up nationwide as yet, which may have the potential to allow more flexible living arrangements.

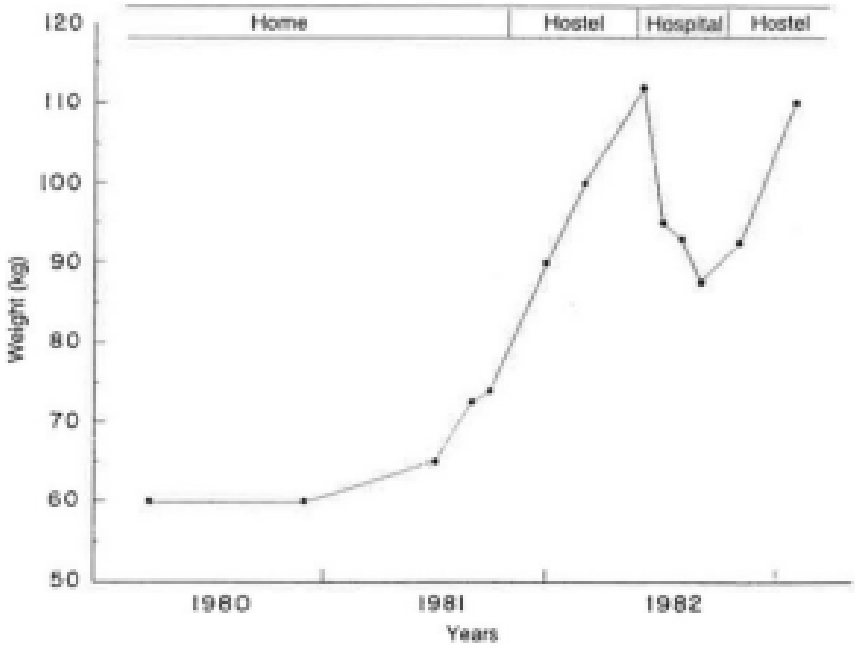

Figure 2 The weight chart of a person with Prader–Willi syndrome showing the large weight increase which occurred when access to food was unsupervised in a group home for people with learning disabilities.

The above diagram shows the first piece of research undertaken by Professor Tony Holland (University of Cambridge) in the 1980s, which shows the increase in weight experienced by a young woman with PWS after she left home and went to live in a hostel, followed by a spell in hospital, and then back to the hostel again.

What we also see in New Zealand is that there are frequent breakdowns in initial placements so that adults with PWS may move from one service provider to another. We know that inconsistency in support leads to weight fluctuations, physical and mental health deterioration, and challenging behaviours accompanied by increased risk, leading to placement breakdowns.

The Transition from Paediatric to Adult Care in Individuals with Prader-Willi Syndrome – C Poitou, A Holland, C Höybye, L C G de Graaff , S Bottius, B Otterlei and M Tauber, pub. 2022

"The transition of individuals with PWS from adolescence to adult life is challenging because of multiple comorbidities and complex disabilities. Individuals and caregivers face psychological, medical and social issues. This period of profound changes is thus prone to disruptions, and the main risks being the worsening of the medical situation and loss to follow-up of the individuals. Medical care may be poorly adapted to the needs of individuals because of a lack of knowledge concerning the syndrome and also lack of the necessary specific skills."

" This vulnerable period needs to work on the environment and extended support. Coordination between the caregivers and social actors is essential to improve the care pathways and daily life of individuals, which are very strongly intertwined. Actors involved in these pathways have a crucial role to help the families."

What we would like to see is good decision-making arrangements in place from the start that do not set a person up for failure.

We believe there needs to be a balance between the priorities of a person's right to make their own choices and managing the risks associated with PWS. We believe that to keep a person with PWS safe from harm, it is necessary to have some restrictions on decision-making and how / where a person lives. There is vast scientifically backed evidence to support that it is necessary for an adult to act on behalf of an adult with PWS in decisions regarding food and food environments in order that risks can be managed.

That said, decision-making arrangements need to involve the person with PWS and a team of people working together to ensure accountability. If a decision-making arrangement is not open and collaborative, things may not go well and the person with affected decision-making is being set up to fail.

We would like to see holistic decision-making arrangements that involve a team of people who have a good understanding of the person with PWS. These arrangements should include family members who will not only know and understand their child well but have often become 'experts' in PWS and the specific health needs of their child's rare disorder. Good communication and access to information is essential amongst the team. Regular team meetings are ideal and a clear, accessible complaint system with mediation available. The person with PWS needs to have choice in who the team members might be by having a say in who works well with them.

We would also like to see adaptable decision-making arrangements because all people with PWS are different and have differing levels of learning disability, impulse control and behavioural challenge, requiring different levels of support. As decision-making capacity can vary from person to person, therefore the level of restricted practice also needs to vary. If choices are overly restricted, this can create further problems, but if any necessary controls are not in place, this can result in harm to the person with PWS or those around them. Safeguards need to be in place for the affected person and the whole support team.

The suggestion of templates in the preliminary paper could be considered for PWS. They could not only help a person with PWS clarify what decisions they can make on their own and which decisions they need help with, but a template could also be helpful in the setting up of decision-making arrangements, i.e. one that is specific to the risks associated with PWS and protection from harm.

Appendix:

1. We attach a copy of an article co-authored by Professor Tony Holland, written for PWSA UK. Prof Tony Holland is a Professor of Psychiatry at the University of Cambridge and Head of the Cambridge Intellectual & Developmental Disabilities Research Group. He is also the President of the International Prader-Willi Syndrome Organisation. His work has included research to inform the UK Mental Capacity Act 2005.

   The article is titled "**The Mental Capacity Act: Supporting People with PWS**" and was revised in 2022.
   (also available here:
   https://irp.cdn-website.com/1b38aac2/files/uploaded/PWSA%20MCA%20guidance%20October%202022.pdf

2. We also recommend the following video presentation by Prof Tony Holland on the subject of 'making sense of mental capacity':

   The Changing Faces of PWS - PWSA UK's virtual conference, 7th November 2021.
   https://www.youtube.com/watch?v=Efxe5gf8GoI

3. **Supporting adults who have Prader-Willi syndrome: caregivers' perspectives on the ethical and practical dilemmas** : a thesis presented in partial fulfilment of the requirements for the degree of Doctor of Philosophy in Psychology at Massey University, Wellington, New Zealand.

   A thesis by Dr John Ford - please ask if you are unable to source this. We can seek permission to forward a copy.

4. The following review may also be of interest:

   A scoping review of case law relating to support and treatment for people with Prader-Willi Syndrome - Anna Murray , Isla Kuhn , Gautam Gulati , Elizabeth Fistein, pub 2021.

   Aims: 1) To review case-law from English-speaking common law jurisdictions concerning support arrangements for people with PWS with a view to identifying issues that have required the intervention of the courts. 2) To identify principles on which to base clinical guidelines relating to the issues identified, ensuring that such guidelines are consistent with ethical and human rights imperatives.
   (https://pubmed.ncbi.nlm.nih.gov/34481216/)

We can provide further supporting documents. Please ask us if you require any further studies / reviews / reports, or further information about PWS.