Proposal to Disability Support Services for PWS specific residential care homes

Background

About PWS – the challenges and risks

Prader-Willi syndrome is a rare genetic neurodevelopmental disorder. It typically causes intellectual disability with symptoms that include impacts on cognition, executive functioning, mood, emotional regulation, impulse control, appetite, and metabolism. PWS is also a complex medical condition. Due to the rare nature of the syndrome and its complexity, it is essential that those who support a person living with PWS have a good understanding of the syndrome, and that consistency is maintained in providing knowledgeable and experienced support.

Some of the greatest risks to people living with PWS are caused by hyperphagia. The control centres that regulate appetite and food reward signalling do not function correctly in PWS, resulting in increased feelings of hunger or starvation, a decreased sensation of satiety, and preoccupations or obsessions with food. This usually means that a person experiences a strong drive to obtain food at any opportunity and is at risk of consuming dangerous

amounts of food. Sometimes the food obtained may be unhygienic, taken from the floor or a bin, and sometimes the food may be unsafe to eat, i.e. raw meat, or meat containing bones. People living with PWS also tend to have voracious eating habits due to intense hunger and their enhanced enjoyment of food, and they will often eat any unauthorised food they obtain very quickly to consume it before being caught or seen. Hyperphagia causes several risks:

- Dangerous volumes of food intake may lead to life-threatening gastric inflammation, necrosis, or rupture. This risk is heightened by slow gastric motility, which is very common, meaning that it takes longer for food to pass through the digestive system – constipation and obstruction are common problems. Due to hypotonia (low muscle tone), a person with PWS is also less likely to vomit to clear any excessive intake. If a food binge has occurred and has caused serious gastric complications, these may also go unnoticed until severe due to a higher pain threshold often experienced in PWS

- Voracious eating habits put people with PWS at a high choking risk. Studies have shown that choking causes 6 – 8% of deaths in PWS (and unlike the typical population, these deaths have occurred mainly in adulthood). Risk of choking is also increased due to dysphagia, poor oral/motor coordination, poor gag reflex, and hypotonia.

It is essential that the recommended '**food security**' strategy is used for managing hyperphagia. 'Food security' is achieved by keeping food securely out of reach, sight and mind, whilst also providing security about what will be available and when. This not only keeps a person with PWS safe but also greatly improves behaviour because thoughts of food and food-related anxiety are reduced, allowing them to focus on other things. People living with PWS respond well to food routines and do not respond well to changes in expectation. Any unplanned extras provided can be very disruptive, both in the short and long term, as these extras may be thought about obsessively and expected again.

It is important to carefully manage the distribution and availability of food, but food management is complicated further by the dietary requirements of persons with PWS. Due to their altered body composition and metabolism, people living with PWS gain weight very easily and require a reduced calorific, low carbohydrate diet to maintain a healthy weight. They also have an increased risk for developing diabetes, and sweet foods should be limited. Regular exercise is also very important, although hypotonia, coordination difficulties and fatigue can make activities more challenging and create a reluctance to exercise. Some people with PWS develop an understanding of their dietary needs, and with support some people might be able to make good food choices, but understandably, they are still unlikely to cope well with seeing others eat food they cannot have, and strategies need to be used to manage this.

PWS is also characterised by a complex behaviour profile. In a paper published in 2021 by the International PWS Clinical Trials Consortium, **consensus definitions and descriptions** were produced for 6 key characterising features: anxiety, obsessive compulsive behaviour, rigidity, temper outbursts, social cognition deficits, and behaviours related to hyperphagia. These behaviours have a significant impact on the daily functioning and quality of life for the person with PWS and their families.

Challenging behaviour in PWS is additionally compounded by executive functioning deficits, in particular, difficulties with thought and activity switching. The impairment of executive functioning affects cognitive processes such as flexible thinking, problem solving, attention and impulse control.

It is important that all persons in support roles work together and are consistent in their approach to managing challenging behaviours. People with PWS have a preference for routine and predictability and are also extremely stress sensitive and prone to outbursts, which can at times result in destructive or aggressive behaviours that are detrimental to the person with PWS and others around them. It is essential to use proactive rather than reactive behaviour management strategies.

Successfully supporting people living with PWS requires a particular skill set. Ideally, a support person needs to be reliable and trustworthy, have good communication skills, the ability to remain calm in an escalating situation, the ability to respectfully and sensitively manage complexities and behavioural nuances of the syndrome, be attuned to manipulation and ploys that people with PWS might use, and be wary of risk, especially when supporting people with PWS in new environments, and have the ability to manage this.

The challenge when supporting adults living with PWS is balancing a rights-based service delivery approach with managing risk. Supporting people with disabilities usually focuses on promoting their independence, but in Prader-Willi syndrome, this is associated with increased access to food without appropriate support and careful management. Support providers need to be able to appropriately balance independence, empowerment, opportunity, dignity and rights, with restrictions of liberty and responsibility for health and safety.

Impact of PWS on families

It has been shown in several studies that Prader-Willi syndrome can be one of the most challenging syndromes for families to manage. For example, a **study published in 2018** found that the mean ZBI scores in 142 PWS caregivers in the USA were higher than those measured in caregivers for persons with dementia, Alzheimer's and traumatic brain injury, all conditions known to have high levels of behavioural challenges and to be difficult for caregivers. Results showed that caregivers experienced strikingly high caregiver burden, particularly caregivers of adolescents and young adults with PWS.

Another **study published in 2021** also demonstrated the high care burden of families living with PWS and highlighted the importance of having dedicated medical care for both somatic and neuropsychiatric symptoms.

A **study published this year** was the first to use a multi-country sample and comprised parents and siblings from 31 countries, including New Zealand. The study found high levels of anxiety, depression and stress in parents and siblings of people with PWS, with siblings exhibiting clinically relevant levels of PTSD symptoms.

Issues with access to residential care in New Zealand

In recent years, the wait times for residential care placements appears to have grown. Families are often told there are no places available and the wait for a placement may take a few years. This puts enormous strain upon families who are sometimes in desperate need of respite. However, at the same time, families are concerned for the health and safety of their son / daughter with PWS and need to know that a residential provider can deliver the specialised support needed and the necessary environmental restrictions. Unfortunately, the options for specialised care and support are very limited in New Zealand.

PWSA(NZ) is also aware there is often a reluctance amongst providers to accept clients with Prader-Willi syndrome due to the high risks perceived and a lack of ability or an unwillingness to provide appropriate restrictive practices. For adults living at home with their families, the reluctance of providers to modify their practice also creates barriers to accessing vocational services, and there are very few adult respite facilities in New Zealand that can cater for the needs of people with PWS.

We have observed that some people with PWS appear to be entering the residential care system at a younger age than in the past because their family has reached a crisis point; this is often through Oranga Tamariki, a mental health facility or the court system. We believe there are several factors contributing to this: more pupils with PWS being declined ORS funding from MOE, unsupported pupils leaving school earlier, less utilisation of special residential schools, lack of appropriate respite supports, lack of accessible support services, difficulties accessing mental health services, and a lack of psychiatric expertise in PWS. This route into the residential care system could be described as an 'ambulance at the bottom of the cliff' and can have devastating, lasting impacts on families and result in irreversible health consequences for the person with PWS when services are unprepared.

The status of residential care for people living with PWS in New Zealand

At present, there is only one PWS specific home in New Zealand, and this is provided by Spectrum Care Trust in Auckland. This home has been successfully established for many years and provides support for 4 female clients living with PWS. There has been little change in the occupancy over the years (one client moved with family to Australia) which is testament to the home being well managed and the residents being content. Weight management is good with no clients experiencing excessive weight gain or the typical health related issues associated with obesity in PWS.

Spectrum Care Trust have earned a reputation for being experienced providers of quality residential care for PWS and currently support 6 other clients with PWS in residential homes across the Auckland area. One of these clients is living with a flatmate who has a similar need for restraints around food, and weight management is good. Two others live together with 3 other housemates who have food seeking tendencies that need to be managed. Due to the knowledge base and practice models Spectrum Care have developed as an organisation, the other clients in mixed disability homes have reasonably good weight management in place.

Unfortunately, not all the stories we hear about residential care are so positive. It is quite common to hear of things going very wrong from the start and adults living with PWS having to abruptly move to a new home or provider. This can be due to a lack of preparation and staff training by the provider, a lack of food security leading to an escalation in behavioural challenges, and a lack of consistency in staffing and service provision.

It is essential that staff have a good understanding of the syndrome and are well equipped and trained to manage the challenging behaviours typical in PWS. When this doesn't happen, we tend to see arguments, power struggles and punitive management approaches, and it is the person with PWS who loses out or suffers as a result. When staff understand what the problems and stumbling blocks are likely to be, strategies and plans can be implemented that enable a smoother transition into residential care. Constant evaluation and planning are needed, but the ability to evolve practice is hindered by a rapid turnover of staff and the contrasting approaches that staff might be used to when supporting clients with other disabilities.

Sadly, we know of two cases in New Zealand where previously healthy young people with PWS have died suddenly in food related incidents due to a lack of understanding about the syndrome (ages 20 and 38), and other cases of early mortality due to complications of morbid obesity (ages 18, 19, 34, 36, 37, 40). We also know of several cases in New Zealand where adults living in residential care have faced criminal charges due to incidents relating to food or poor behaviour management. We believe the number of people with PWS who have been through the justice / RIDSAS systems is disproportionately high. Sometimes this has also resulted in people with PWS receiving very high needs support packages (i.e. 2:1 support) when it is quite possible this would not be necessary if they had previously been able to access higher quality support.

Unfortunately, many of the adults living with PWS that we meet through our work are either obese or morbidly obese and experiencing negative health consequences as a result. This is not helped by the limited availability of medical expertise for adults with PWS, and whilst we encourage residential providers to facilitate annual health checks, medical care for adults with PWS is often overseen by a GP rather than a specialist.

The PWSA(NZ) maintains an information website, has various printed publications freely available, and offers training courses for residential services and schools. However, we can only stress the importance of receiving PWS training, but we are unable to ensure that providers utilise this service. Some providers in New Zealand regularly engage with us for training whilst others do not.

Case example 1

J was a classic example of a person with PWS who did not meet the current criteria for ORS funding (2 applications declined), despite having specific learning disabilities, intellectual disability, challenging behaviours and being diagnosed with ASD as an adult. (PWS is often viewed as an 'umbrella diagnosis' and separate diagnoses are seldom evaluated, but it is parental view that J probably also had ADHD.) Increasingly challenging behaviour led to J having to leave school earlier than expected with no transition services in place. J's family struggled to find things for J to do with insufficient support and had to wait almost a year for their NASC to review J's support package after his change in circumstances. At his review, they requested an application be made for a residential placement and were told there was a wait of about 2 years and that someone would be in touch to do the assessment (the family never heard any more on this.) When they finally received an increase in J's support package, they experienced difficulties in utilising this IF package at first due to J's reluctance to try some things, refusal of some services to accept J, and the need to find suitable support workers.

After leaving school, J 's behaviours initially improved somewhat because the stress of that environment had been removed. However, around a year later, J started exhibiting a sudden increase in aggression. There had also been a couple of incidents of concern where J had mentioned hearing voices. J's family had tried to access mental health services several times during his adolescence and after an episode of hearing voices, but J had been discharged twice, and despite impulse control disorder being diagnosed, J was only ever prescribed an antidepressant /anxiety SSRI medication which appeared to be having no effect. J's family believe the sudden worsening in J's mental health was exacerbated by increased feelings of frustration at not being able to maintain employment, be more independent, and a lack of stimulation. At the age of 19 years, J's rapidly escalating behaviour culminated in a traumatic event which has had long lasting devastating consequences for J's family.

J had to be removed from the family home for the protection of his younger siblings and was taken to the emergency department for psychiatric assessment. The assessing clinician made a judgement that they could not

be certain J was experiencing psychosis, despite mental illness and psychosis presenting atypically in PWS, and J was therefore held on remand in police cells for a night whilst officers sought an emergency respite placement, then transferred to Rimutaka Prison the following day after it was determined there were no respite beds available in the entire Wellington Region. As J's family had no extended family or support contacts where J could be remanded, J had to spend 3 weeks in the prison ISU until they managed to get him transferred for further assessment to a secure youth forensic inpatient mental health facility. Although J was technically an adult, it was agreed that the youth services would be more appropriate for J.

J was remanded to this facility for almost 5 months which was an extremely stressful period for him. The prison-like environment and the uncertainty of what was going to happen to him saw his behaviour and mood drastically deteriorate, including a suicide attempt and A&E admission. J was weaned off his SSRI whilst multiple assessments took place, but none of the assessors had expertise in PWS and because no further episodes of psychosis were observed, it was determined that J be court ordered under the IDCCR Act to become a secure compulsory care recipient with a RIDSAS provider. Just before leaving the mental health facility, J was prescribed an antipsychotic medication.

J's family were very concerned about the proposed RIDSAS placement because although they were assured the kitchen would be secure, the provider had no experience of PWS, had not received any training, and the location was not close to their home or conducive to J being able to return to any of the activities he had previously enjoyed. However, they were informed there were no other options - J had to live with a RIDSAS provider, the service closer to his home did not have a bed, and the judge would rule according to the recommendation of the forensic services care coordinator. J's family supplied the RIDSAS provider with lots of publications / written resources about PWS and urged them to book PWS training. The compulsory care coordinator completed a needs assessment and care plan that contained little health information about PWS, no clinical advice was sought, and J's family were not shown these documents at the time.

The antipsychotic medication worked effectively, J was calm and there were no behavioural incidents during the 6 months J lived with the RIDSAS provider. However, J became withdrawn and stayed in his room. He was very unhappy in his new living environment away from his home, and largely because he was living with 5 other residents who were older men, some of whom he found particularly frightening, and he was probably exposed to bullying or threats. J also spent much time with nothing to do and the few activities offered could not be described as the rehabilitation that was intended by his placement there, with the exception of a Stepping Stones course that was started after several months and could have been attended from a different location.

J's family were very concerned about the lack of stimulation and J's low mood, but they were also extremely alarmed by J's rapidly increasing weight. J's diet was being very poorly managed and he gained 16kg in 12 weeks, and 20kg in total during the time he lived with this provider. After complaints, the provider did make some effort to address the dietary issues and weight management but by this time, J had developed diabetes. Tragically, J died in May 2023 at just 20 years old after developing ketoacidosis which was triggered by an abdominal infection. It became apparent from postmortem that the provider was not supervising what J was eating because a small abscess caused by a subsequently healed perforation of the small intestine was found, indicating that J had eaten something sharp. It is believed this could have happened up to a week prior and there would have been signs of J's worsening condition for at least 2 days before medical help was finally sought. J was in a life-threatening condition of severe DKA on admission to hospital and it is the view of the pathologist that J probably would have survived had he been admitted earlier.

Support records have shown there were clear signs of J being unwell that should have been acted upon by support staff, including J spending his last day there in his bed. None of the staff working at the home at this time appeared to be aware of any of the important health alerts associated with PWS and failed to act on any of the 'red flags' that are well known to anyone with an understanding of the syndrome. Staff at the home had not received any PWS training and it appears the information materials provided were not shared or used.

J's family are expecting nothing less than a gross negligence prosecution against the residential provider and the investigation is currently ongoing. However, J's story is not only an example of the need for improved access to more appropriate residential placements with knowledgeable, trained support staff, but also an example of the need for better support systems that could have prevented the series of events that led to J's situation and the tragic loss of a previously healthy boy who just over a year earlier had been excitedly expressing hopes and dreams for his future. J's family firmly believe if J had received better education, mental health and social supports, J's story would have been very different. Also, if J's story were examined from a cost only perspective, it would likely reveal that the costs involved in the provision of those earlier supports would have been less than those of incarceration, a very high-cost forensic inpatient stay, multiple assessments, judicial costs, 1:1 RIDSAS funding, and hospitalisations.

Case example 2

F. is a 40 year old female with PWS and added complications of morbid obesity, diabetes Type II, and IBS. Because of her obesity her balance is not good and, should she fall, she requires the services of an ambulance to get her back on her feet.

She is supported by a residential agency in the Wellington region which has shown a very proactive interest in understanding the syndrome and giving the best possible support. She is the only person in the rented house and has 24/7 care, of which 8 hours are double staffed. However, in the past she has had many interactions with the police and the court system because of her inability to contain her mood swings, which have become more violent since her teenage years. The failure of services to support F appropriately has caused much upheaval in her life and significant health decline.

In childhood F was able to attend Salisbury Residential School which showed a remarkable ability to manage her dietary needs, behaviour, education, and her mental wellness. She had 4 years at Salisbury and came home to a small-town school which was not particularly successful.

At the age of 17 she was able to enter residential care with one of NZ's largest residential service providers with 1:1 staffing, including overnight. However, her mood swings became elevated with dangerous consequences ending in court appearances for shoplifting, stealing, violent behaviour and breakages, and for attempting to do harm to a staff person. She was sentenced to one year at a RIDCAS service, which was an extreme lock-down environment with no food control, combatant behaviour control, and some risky male clients. This was entirely inappropriate, but her family was unable to cope, and the previous provider was reluctant to have her return to their services.

This was a harrowing time for F. and for her family. When the year ended, she moved back home very overweight, and her family had to set about finding new residential services. They found a placement in the Manawatu with a second female who also had PWS and a third client who had dietary needs. Although this was considered an

excellent result, it actually was not. The two clients with PWS were not compatible, although at times they managed to trick staff into situations allowing them freedom of the kitchen. Staff were not trained in the specific needs of PWS and after a year, F. was sent back to the RIDCAS provider where – as there were no available beds – she was sleeping on the floor.

Her family took her back for the second time and continued to seek good residential care. After a year at home, they found a placement willing to take her. With training from PWSA(NZ) on a regular basis, and specific requirements for dietary needs, the stage was set for success. However, people with PWS can be extremely good at sabotage. Meals were compromised for the sake of good behaviour, F. went back to begging, asking for money, asking neighbours for food, etc. Her behaviours escalated as her hunger increased. Police were being called on a regular basis, staff turnover was high. Extreme stress was felt by family, staff, and by F. herself.

Many attempts were made by her mother to have F. seen by a psychiatrist but she was merely put on a waiting list and the only time she saw a forensic psychiatrist was after one of her frequent court appearances. The forensic psychiatrist was unable to diagnose or prescribe appropriate treatments in the context of PWS. Finally, after one year of desperately seeking for her daughter to be seen by specialists, a team was put together comprising F's GP, mental health team members, a psychiatrist familiar with PWS, a consultant psychologist, her residential Team Leader, and her mother.

It was only through utter persistence that F. was able to meet with team members, for team members to agree on a forward plan *including* a new drug regime, and regular appointments with her GP to monitor progress, that improvement to her whole demeanour started to be noticed. It took just one week before staff were commenting on how equitable F had become, how there were no more fights with staff; her family noticed the difference, and F. herself stated she was far less anxious, far less angry, and starting to enjoy her life. Four months later the improvement is remarkable. She has accepted her new dietary regime and although weight loss is the absolute end goal, this is slow to happen. F's morbid obesity is of ongoing concern to all and everything is being done to help support her in this journey.

Case example 3

H was born in a small Marlborough town in the late 1980s into a very supportive family, extended family and community. Due to geographical isolation, she had no access nor was offered access to medical specialists needed to treat Prader-Willi syndrome. Her primary school years went fairly smoothly (with teacher aide support), but when H moved to intermediate / high school and her support hours were reduced at the same time as the challenging traits and characteristics of Prader-Willi syndrome started developing (as they normally do in PWS during adolescence), problems started to occur.

H attended Salisbury Residential School for two years which had good knowledge and successful history of working with PWS. These were probably H's happiest and most rewarding times. On returning to her local high school at 15, her ORS funding had been removed so there was little support - behavioural problems, food seeking/stealing, struggling to achieve were prominent. H and her family decided staying at school was not an option as they were unable to offer the support needed to keep her safe. This was a very difficult time for H and the family. There were no respite or residential options in the small town where they live.

After several years of trying to find a placement for H in the Marlborough area, she moved to a residential service in Nelson for three years with limited success. The first year was in a shared home with others but this proved too difficult. The staff were unable to manage the challenges and complexities of Prader-Willi syndrome and H was moved into her own home which was intended to have 24-hour support. However, the support was not consistent, H gained significant weight and was involved with shoplifting and social challenges in the community.

It was decided to try a different setting for H and she was moved to a home in Manawatu. H is an example of a person with PWS with high functioning / borderline intellectual disability which can cause even more challenges for providers and staff supporting her. At this house there were other residents with high levels of intellectual disability and in some cases limited speech and vocabulary. H was quite under-stimulated and bored, with a lot of time spent sitting around or joining staff outside on many cigarette breaks. Behaviour problems and meltdowns were increasing while H lived there. The service struggled to provide positive behaviour support and the family were very concerned about H's mental health, so they decided to move her nearer to home where they could better support her. Although she had managed to lose a lot of weight she had gained, she was now an addicted smoker.

Once again there were very limited options for any residential placement, so it was decided to trial an Individualised Funding / community support combination package whereby H lived in a shared flat with one other person with a disability. This IF package was very generous and enabled a large support team to be put around H. However, she managed to outsmart most people, and the result was shoplifting and stealing, problems with the police and various members in the community, rapid weight gain and health problems.

The family relationship had become very strained by this point with the family deciding they could no longer support H's lifestyle and choices and the effect this was having on her health and their health. They faced a challenging decision and encouraged H to choose another path, or they would have to step back from her life. Fortunately, H chose to move into a residential setting not far from home. In the past this had not been seen as a good option but due to her declining health and higher support needs, it needed to happen. This was a rural property with nine people in three dwellings, a family-like setting that H actually thrived in. She lived happily and reasonably successfully there for six years. There were a variety of challenges for the staff, but most of these were health related. Despite their best efforts, H's weight continued to increase and her health deteriorate, with several episodes of cellulitis and major infections. H had approximately 10 hospital admissions during the time she lived at this home and the recovery time seemed to take longer after each one.

When a new manager was appointed at the home, they decided rather quickly that H caused too many problems and needed to go; she did not want to go – and had nowhere to go. This resulted in a year of neglect and abuse for H that was extremely stressful and tragic for H and her family. Her health deteriorated even more, and H was very isolated. A formal complaint was made to Whaikaha, who eventually ensured H had funding to move elsewhere.

In February 2023 H moved into a flat near her family and her community with a small but wonderful support team around her, and most of her needs at this stage were health related. Due to the impact of neglect and abuse she had experienced, H had also lost the will to live and chose to give up trying to live with PWS. She died in July 2023 at the age of 34 weighing 160 kg, the tragic loss of a beautiful soul who should have had a much better life.

H's family believe that the constant changes were very detrimental for H. She, like many others with PWS who have had bad experiences and outcomes, have had to fit the system (that didn't work for them) and this caused so

many problems for H when her life was already challenged by being born with PWS. They believe this proves there is a need for PWS specific residential homes with consistent, qualified staff that can accommodate and support their needs whilst providing a good and healthy life.

H's family also believe H's health problems could have been avoided with early access to specialist treatment and care that is needed to keep a person with PWS well. They would dearly like to see adult multidisciplinary clinics established that H could have attended on a regular basis so that there was oversight and health management (mental and physical) for her, her family, and those trying to provide support.

Case example 4

L is 17 years old and currently living in a national forensic ID secure unit after being placed under the IDCCR Act. L's story is another account of lack of services and the services not being able to manage the level of care he requires. L grew up in a South Island city and has several other diagnoses alongside Prader-Willi syndrome which include intellectual disability, ADHD, anxiety and autism.

L was mainstream schooled and several attempts to apply for ORS were declined. He managed well until age 10 when school became more difficult and peer gaps widened. L's behaviour also became more challenging and demanding. L's family reached breaking point and were not aware or advised of any qualified support or respite services. They did receive funding for some respite care hours and L spent some time with his aunt, but this was not enough to provide the support and break the family needed. After a serious assault on his mother and a desperate cry to the police, L was taken into Oranga Tamariki (OT) care for the protection of his family from aggressive behaviours. Violent and inappropriate behaviours also led to L being stood down a few times from intermediate school. An application to Halswell Residential College was declined.

OT were unable to find appropriate in-home care for L, so he was taken to the local DHB's respite care facility where he lived for many months. OT were then able to contract a bespoke placement to a care agency and L moved to a house where he lived alone for a couple of years. During this time L was excluded from school in his first year at college (in a learning support unit) after some antisocial behaviours. This decision was contested by OT, L's family and PWSA because it was believed these behaviours were typical of PWS and could have been avoided, but L spent significant time unable to attend school. L was then accepted as a bespoke student to the only special school in the region, arranged with support from MOE because L did not have the required ORS funding. L was schooled off site in a church with one other part time pupil, but this was situated across the road from 2 food outlets on a State Highway which caused a huge amount of anxiety for L.

Staff at L's home were finding it very difficult to support L, the staff turnaround became unmanageable for the service provider to continue, and they gave OT 48 hours notice of the service terminating. An emergency temporary care provider not specialising in disability came to look after him, but in the month that he was supported by them he gained 10kg because they did not maintain his strict meal plan.

A further agency offered to support L but was only just setting their service up in L's home region, so they asked that he be taken out of region to one of their other homes while they did this (a city more than 5 hours away). This was a stressful time for L to be living away from his family and routines. L's aggression grew even more. He was damaging his home on a weekly basis, smashing through doors and windows to access food. He started to abscond from his residence and steal food. Police involvement was becoming more regular as the providers could

not restrain him or stop him leaving the property. After 8 months they transferred L back to his home city and after less than a year they also terminated their services as they were unable to recruit staff quickly enough. L had committed a number of offences - wilful damage of his home, assault and shoplifting (he was trespassed from a number of places.)

OT were scrambling again to find care providers and had to bring in agency staff from a provider in the North Island. This provider worked hard to upskill staff and engaged extensively with PWSA for training. Staff were experienced, had the best relationship with L and provided the consistency he required, but it was challenging for them as staff teams were travelling back and forth from the North Island (at great cost).

L's behaviour continued to escalate. He became miserable and suicidal but was declined mental health support on several occasions. L was running into traffic and managed an overdose of one of his medications. L was seen in ED numerous times and would sometimes require restraining in ED. This was all very stressful for him and his family. The family had been asking for years to have a full medication review as he was on many different medications which were not appearing to help. When L finally did get to see the mental health team, they felt his behaviours and suicide attempts were due to situational distress rather than mental health. The family have a current complaint with HDC following the lack of supportive services provided by the local hospital.

At this point, L had 2-3 support staff 24 hours a day. These staff teams were still travelling across from the North Island and were reaching burn out. OT continued to look across the country for other options for L but were not able to find anything suitable. The provider agency offered to build a service around L in the North Island and L was transferred out of region again, displacing him from his family for a second time but it was necessary to keep the support team resilient.

It took some time for extensive modifications to be made to the new house for L. Unfortunately, transition did not go well and L continued to abscond, stealing from the local supermarket and then committing a burglary at a neighbouring house. L had been held in police cells overnight on 2 occasions and spent 3 days in a youth justice facility, a very inappropriate place for a young person with a disability. He was then remanded in a youth mental health secure unit for a 30-day assessment. It is a result of that assessment that it was found to be appropriate for L to receive the current level of secure care. Since being in the secure unit with no means of leaving and no access to unauthorised food, there has been a significant improvement in L's behaviour and mood. He is now able to relax a bit more and is enjoying the on-site school. The medical team are doing the long-awaited medication review and medications are being rationalised and slowly removed.

For L's family it has provided a sense of relief to know he and the public are safe, but they are heartbroken that L has had to go through a criminal path to get the right support. It has been a long frustrating process to get L the care he requires. L will be 18 this year and there is much uncertainty about his future as he will be unable to stay where he is indefinitely, but with adolescence and early adulthood notorious for being the most challenging period in this very complex syndrome, it will be essential that the right steps are taken so that L transitions successfully through the next few years and can move forward without having to experience more negative setbacks.

Case example 5

T is a 22 year old young man living with PWS in rural Otago. T lives at home with his parents as there are currently no other options available. T's parents describe him as quite low functioning and similar to a 3-4 year old

intellectually which results in the need for 24/7 supervision at all times - sometimes 2 on 1 care. T also has a diagnosis of ADHD and has cyclical moods ranging from low mood/depression to mania/psychosis. He takes antipsychotic and mood stabiliser medication.

After three attempts, T was granted ORS funding at primary school. By the time T was at high school his behaviour had become increasingly inappropriate and aggressive, and after two further attempts T was granted Very High Needs ORS. During his time at school, T was stood down for aggressive behaviour which included hitting pupils and teachers. The common PWS traits of rectal picking and skin gouging were a major problem during the school years and were very difficult for staff to deal with. Running away from school was also a problem and there were several occasions when he managed to escape from school grounds and would be on the main roads before it was noticed he was missing. Food seeking was always an ongoing issue. T's challenging behaviour meant his school day became shorter and shorter until he was only attending for 2 hours per day. Eventually it was agreed that school was no longer suitable for T and he left before he turned 21.

T started at a local day centre for adults with intellectual disabilities, but this was short lived as staff were unable to manage T's aggressive and non-compliant behaviour. Unfortunately, a female support worker was injured with a black eye, and holes were kicked in walls and doors. After the first Covid lockdown, it was agreed that T would not return to the centre.

Consequently, T's mother gave up her job to stay at home full-time and provide care and support for T. Since T has been assessed as having very high needs, his family receive a reasonable amount of Individualised Funding for respite care. However, it is hard for them to find suitable support workers who can manage T and are familiar with PWS. When they have found caregivers, they have not been able to retain them. T's parents have relied heavily on T's two siblings for respite care, but they have now left home and no longer live close by.

T currently spends his days with his parents as there are no other options for him. He is a fully grown young man weighing 106kg and is sometimes too much for his mother to handle. Both parents are in their late fifties and wonder how much longer they will be able to manage T with such little respite care available. They would really welcome the availability of a residential facility not too far from home that can manage the specific food seeking and other challenging behaviours of PWS with appropriately trained staff. Having seen T's behaviour at its worst due to lack of skills and training when T attended the day centre and being fully aware of what can go wrong with inappropriate support, T's family are understandably very anxious about T's future.

Residential care placement considerations for supporting people living with PWS

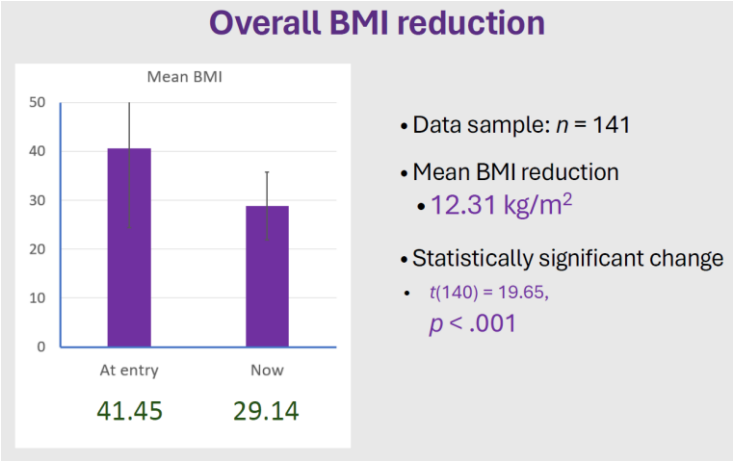
- Independent living is not known to have been successfully achieved and people with PWS usually require high levels of 24 hour support.
- Modifications or alterations are usually needed to the home to ensure a PWS safe environment.
- The location / neighbourhood of the home needs to be considered, for example a home that is too close to a food outlet is not advised. A calm area is ideal with connections to social environments.
- People with PWS need to live with restrictive practices, particularly regarding food accessibility, which will impact on other residents in the home.
- A person living with PWS will need an individualised dietary plan to maintain or lose weight.

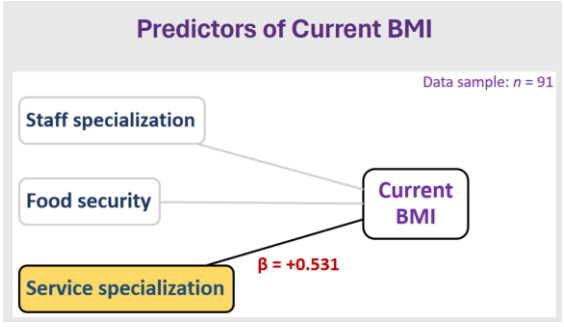
- Support is best provided by skilled, capable staff who receive ongoing professional development.
- PWS specific training is essential and must be made available to all new staff before commencing employment in the home.
- High turnover of staff is problematic when trying to provide the necessary consistency in routines and house rules, and use of casual staff is not recommended.
- Appropriate levels of support may differ from concepts of what constitutes appropriate levels for other residents with similar intellectual or behaviour disorders.
- Links need to be established with health and allied health teams, and sometimes specialist behaviour support services.
- Mixed disability settings can be challenging for people with PWS as the needs of all residents need to be considered.

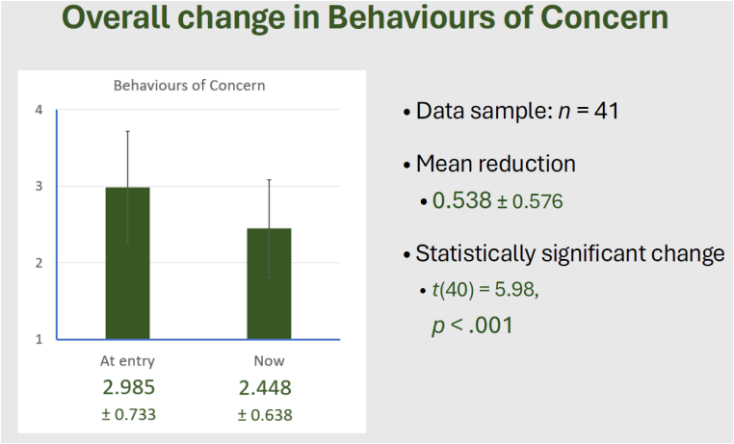
Improved outcomes of PWS specific residential care

The most suitable option for residential care and support will vary according to the needs of each person living with PWS, but it is the general view of many clinicians and experts that PWS-specific care represents best practice. This was confirmed by a paper published in Feb 2024 which examined the impact of residential care in PWS and concluded “full-time care services have a high likelihood of enhancing the lives of people with PWS within one year with long-lasting benefits, especially if those services are exclusive and specialised around the particular needs of PWS.”

The International PWS Organisation (IPWSO) collated information from service providers in 6 countries (Denmark, Germany, Ireland, Switzerland, UK, USA) to form an anonymised dataset for statistical analysis. Results showed significant reductions in weight, BMI and behaviours of concern after joining a full-time care service, with people living in PWS-exclusive services showing the greatest improvements in weight, BMI, and BOC. The paper also states, “Reductions in BOC were associated with greater, rather than less, social contact, suggesting that these improvements were not achieved at the expense of broader freedoms, such as the opportunity to meet with families and friends.”







Hughes, B.M., Holland, A., Hödebeck-Stuntebeck, N. *et al.* Body weight, behaviours of concern, and social contact in adults and adolescents with Prader-Willi syndrome in full-time care services: Findings from pooled international archival data. *Orphanet J Rare Dis* **19**, 48 (2024). https://doi.org/10.1186/s13023-024-03035-x

Other advantages of PWS specific residential care

From a management perspective, it is easier to implement appropriate practice models, restrictive practices, and consistent behaviour management strategies in PWS specific homes because of the similar support needs of residents. The similarity in nutritional needs can also simplify the ordering and preparation of food. Residents are also likely to experience similar challenges with mobility and exercise capacity which makes it easier to plan beneficial exercise programmes and motivate residents to participate. The coordination of transport could also be less challenging because whilst residents would each be engaged in their own interests, social and vocational activities, there would likely be some shared activities and necessary health visits.

Recruitment and retention of support workers remains a significant challenge for residential care providers. This creates additional problems when providers support clients with a diagnosis or health condition that requires staff have a good level of understanding of the condition, and where the use of casual agency staff should be avoided if possible. In a home that provides PWS specific care, there would always be staff with PWS training and experience working at the home and it is therefore much easier to on-board and upskill new staff.

Last year, alongside a scholarship grant from IPWSO, we supported Bjørn Christensen, a Service Delivery Lead at Community Connections, to attend the International Professional Providers and Caregivers Conference in Berlin. His conference report included the following comments regarding staff retention:

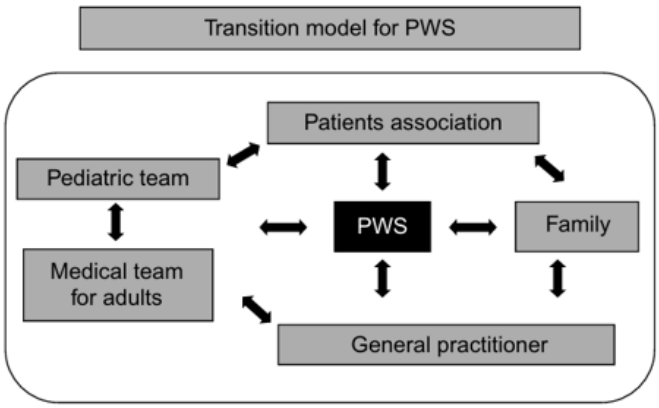
- “Acknowledging the specialisation that comes with work alongside someone with PWS means that we can then set a professional benchmark for how we want our staff to be trained in order to succeed. Success = Staff Retention!”
- “Succession planning is critical.”
- “Something that I really agreed on with this (*staff retention*) workshop was creating a realistic preview of what supporting someone with PWS looks like. If someone is given the right onboarding and has also been a good snapshot of what is needed to succeed, I think it is plausible that they are more likely to stay employed with us for a greater length of time.”

In addition to greater reductions in weight and BMI observed in services exclusive and specialised in PWS, we believe specialised residential services have the potential to achieve other health and wellness benefits. Residential services tend to use their own health advisors, but these advisors typically do not have extensive

knowledge of PWS and can miss subtle health changes that are more significant in PWS. A study found that many health problems in adults with PWS remain undiagnosed (**61%**). A service that supports a larger number of clients with PWS is more likely to develop expertise in the health advisor role. We would also hope that if multiple PWS patients were registered with a local GP, their experience and expertise would develop too. Individuals may also have improved access to allied health specialists and counsellors if they worked with household groups rather than individuals when appropriate. In a PWS specific home, residents would also have the opportunity to benefit from a mutually supportive environment because the residents would understand some of the needs and anxieties of their peers.

Unfortunately, we do not have ideal health services for adults living with PWS in Zealand at present. Paediatric care is usually multidisciplinary and often comprises an endocrinologist, developmental paediatrician, dietitian, physical therapist and sometimes others, but once patients transfer to adult services, multidisciplinary care ends. Ideally, an endocrinologist should be overseeing patient care, but some adults only see their GP. Scientific papers list comorbidities that should be evaluated in PWS, i.e. **Poitou C, Holland A., Höybye C, et al 2022** and **Crinò A. et al 2016** which focus on the transition period because it is during this pivotal period during and post transition that patients are most at risk of health deterioration (obesity, diabetes, psychiatric problems etc). The reduction in health services, loss of expertise and history (specialists who may have worked with a patient for many years) is very poorly timed with this period of high risk. It is also notoriously difficult for patients to access psychological and psychiatric support, which is often needed at this time, particularly from professionals with PWS expertise.

We would like to see New Zealand develop a multi-agency approach to supporting adults living with Prader-Willi syndrome – disability services working together with health services to form multi-disciplinary teams. We suggest facilitating this approach could be assisted by teams being able to oversee the care of patient groups in PWS specific residential care homes. Ideally, Health NZ would establish PWS clinics for adults similar to those held for paediatric patients in the main centres. Care would focus on endocrine, psychiatric, nutritional and social issues. In a retrospective study that compared parameters depending on transition services received, Poitou, Holland, Höybye et al showed that "*a coordinated care pathway with organized transition from a pediatric hospital to a Reference Center for adults with PWS, with multidisciplinary follow-up involving health and social care professionals, contributed to better metabolic and psychological health in adulthood.*" As the focus areas of transition are medical, social and environmental, we see a role for residential services in this process.



Allied health, residential and disability support services would fit within Crinò et al's transition model for PWS.

Poitou, Holland, Höybye et al and Crinò et al also both stress the importance of someone overseeing, coordinating and monitoring transitional care.

Potential cost-saving implications of PWS specific residential care

Whilst there would be an initial set up cost involved in either building purpose-built homes or modifying existing housing, we propose that providing PWS specific residential care could be more cost effective in the long term:

- Less homes requiring modification overall.
- Implementation of appropriate and consistent behaviour management strategies can lead to a reduced need for 1:1 support.
- Reduced impact on high-cost services such as the forensic and high needs framework.
- Reduced costs for the health system if obesity is avoided.
- Could work in collaboration with Health NZ to provide streamlined health services, i.e. clinic check-ups rather than multiple individual appointments, reduced number of house visits for health professionals, combined patient health advice, i.e. a dietitian working with a household rather than individuals separately.
- Streamlining of behavioural support services to people with PWS (can sometimes be offered to a household rather than individuals.)
- May reduce grocery costs if menu planning is for people who have similar dietary needs rather than catering for the dietary needs of people with mixed disabilities.
- May reduce transport costs if some travel is combined, i.e. clinic appointments.
- Service specialisation means 'in-house' training could be developed which may reduce time and cost of external training needs.

Examples of PWS specific residential care

There is significant variation in what PWS specific residential care looks like internationally, as well as variability in what might be offered by different providers within the same country. There is no single correct method of providing accommodation and support for people with PWS, considering individual needs must be taken into account and PWS specific care is still evolving with new ways of thinking, but service providers are generally guided by IPWSO's best practice guidelines.

IPWSO's research investigating the impact of full-time care services on people with PWS found that people living in homes with a larger number of residents had greater improvement in behaviours of concern, but this evidence was slightly skewed by a bimodal dataset which included a group that lived with almost 30 cohabitants, whereas most individuals lived with fewer than 10 cohabitants. The large group of cohabitants were most likely from **Latham Centers**, a campus-style accommodation and support provider in the USA which has an international reputation for providing first-rate support for people living with PWS. Latham has excellent expertise with a team of highly trained clinicians, special education teachers, direct support professionals, nursing teams, and support staff which provide comprehensive therapeutic services. Whilst families often look to providers like Latham wishing there was something similar available where they live, it is inevitable that facilities like this cannot exist in every area which means residents would likely have to move away from their families and existing connections.

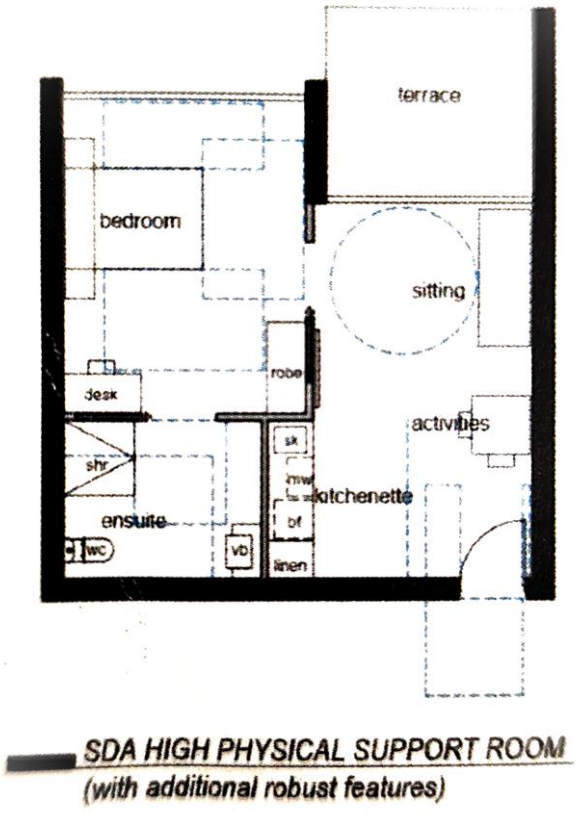
In New Zealand and Australia, smaller community based residential services are favoured that usually accommodate between 2 and 5 occupants. A similar approach is taken in other parts of the world with some variability – some houses in the UK and Europe accommodate slightly higher numbers of residents. In this proposal, we list some examples of PWS specialist providers with links below, but we will look at some Australian examples in more detail.

Like other disability supports in Australia, residential care is funded through the NDIS. Accommodation providers in Australia can become **SDA** registered (providers of specialist disability accommodation that meet SDA **design criteria**) and NDIS participants seeking specialist support may need to have SDA approval in their NDIS plan. Many SDA homes in Australia are provided through partnerships between established support providers and one of the **many SDA property managers** such as **AccessAccomm**. Companies like **NDIS Property Australia** work with **investors**, providers and support coordinators to assist stakeholders in creating solutions.

Interaction Disability Services established Australia's first PWS specific accommodation model in 1992 and now have multiple PWS specific homes in Sydney and the Central Coast. In partnership with AccessAccomm, Interaction have **developed purpose-built accommodation** for supporting people living with PWS. The home comprises two standalone purpose-built homes adjacent to each other, with three tenants residing in each. They have been designed to the 'Robust Level' of SDA Design Standard and include higher accessibility space, ensuites, and an office for 24/7 support. Potential house layout below:

Interaction explain their PWS specific homes allow for consistent practice, with essential medical and lifestyle supports. Healthy food options are based on the recommendations of the PWS clinic for adolescents and adults at Royal Prince Alfred Hospital, Sydney. Meals are large in size but low in calories with lots of fresh vegetables, fruit and salads, and 6 meals a day. Most residents participate in daily exercise programmes at a gym or on treadmills. Staff are highly trained, skilled and experienced. Interaction state that "PWS specific shared accommodation is a cost-effective option."

Multicap are another provider of PWS specific care and plan to open **Queensland's first purpose-built supported home** for people living with PWS to meet an identified unmet need. Multicap have explored contemporary support models for PWS to provide a supported independent living home that provides a service model based on best practice. The design and layout of the home has been informed by extensive research and consultation with international experts, parents, and caregivers. The home will accommodate 5 people, with overnight support staff, in villa style accommodation where each resident has their own living space with views of the natural environment. Each villa will have security features, independent environmental controls, an ensuite bathroom, and a kitchenette allowing residents to engage with food preparation as part of their personal development if appropriate. The property will also have a secure communal kitchen, and considerations of design have even included how groceries will be brought into the home. Outside there will be walking tracks and exercise equipment in the garden.



SDA HIGH PHYSICAL SUPPORT ROOM
(with additional robust features)

Another example of a PWS specific home in Australia is this property in Victoria, supported by **Scope**.

Other international examples of PWS specific residential care

AME Community Services, USA

11 homes in Minnesota, each accommodating 3-4 residents. Most common staffing level is 1:2. The level of food security needed in each home can vary with residents living in the home most appropriate for them. Several therapeutic and behavioural services available.

www.pwsausa.org/minnesota-prader-willi-group-home and www.amecommunity.com/who-we-serve

Prader-Willi Homes of Oconomowoc, USA

11 homes nestled into the community in and around Oconomowoc. 40 years service history. Clients from across the USA. The large number of homes allows PWHO to have a large and diverse staff team, including allied health.

www.pwho.com/homes/

Abilities Midwest, USA

4 homes in quiet residential neighbourhoods within inviting, small-town communities in Wisconsin.

www.abilitiesmidwest.com/program-results

Resilience Healthcare, Ireland

2 residential homes with another opening soon, and 1 PWS respite facility. Support team includes allied health. Individual rooms / apartments with ensuite facilities, and general areas such as an activities room and gym.

www.resilience.ie/prader-willi-syndrome

Prader-Willi specific homes in Denmark

Denmark has a similar population to New Zealand and has 10 group homes which accommodate 70-80 adults living with PWS. (There are also some other houses where 2-3 people with PWS live with others where the primary issue is behaviour.) Not all homes have apartments, some have rooms and shared bathrooms, but the final goal is a private apartment for each person. Dietitian input, once/week physio, visits to PWS Center (medical).

Grankoglen - www.sua.rm.dk/tilbud/grankoglen

Spruce - new architect designed home with 8 apartments for 2 groups of 4, with common areas and gym (see slides 47-61 for images): https://download2.eurordis.org/documents/pdf/sss/6-HA-PWDK-Susanne-Blichfeldt.pdf

Consensus, UK

12 homes in different parts of the UK, 40 years experience – supported over 200 people with PWS in last 10 yrs. Have developed 'PWS Academy', an interactive digital academy for accessible staff training. Some facilities are large scale, i.e. Gretton House with 17 rooms + 3 s/c apartments in a rural location, whereas others are smaller.

www.consensussupport.com/services/gretton-house

Some positive stories: www.consensussupport.com/the-new-gretton-pws-breakfast-bar and www.consensussupport.com/keiths-journey-at-clare-house

Dolphin Homes, UK

Abbey House accommodates 8 residents with PWS, all rooms with ensuite bathrooms.

www.dolphinhomes.co.uk/abbey-house/

Priory Group, UK

5 homes in different parts of the UK (3 around Hastings) housing 3, 5, 6, 8, and 9 occupants. Most rooms ensuite.

www.priorygroup.com/residential-services/prader-willi-syndrome-support

Voyage Care, UK

4 or more homes in different parts of the UK each housing 6-7 occupants, such as Redbank and Esmer.

www.voyagecare.com/service/redbank-house and www.voyagecare.com/service/esmer-house

Prader-Willi specific homes in Germany

Germany has approximately 16 residential homes tailored to the care and support of people with Prader-Willi syndrome. We have not been able to source individual websites, but well-known experts in PWS, Dr. Norbert Hödebeck-Stuntebeck and Dr. Hubert Soyer, are involved with these homes and could provide further information.

www.prader-willi.de/pws/adults/?lang=en

The demand for PWS specific residential care

Statistics and parental interest

PWSA(NZ) has 103 adults (18 years+) listed in our database as currently living with PWS. It is likely that there are more adults living with PWS that we are not aware of, but we cannot access this information in the absence of a national patient registry.

In 2022 we surveyed families whose son or daughter was living at home and asked what their preferred options would be for residential care, with the aim of helping us connect families who might be looking for the same things. There was a slight preference for PWS specific homes, but most families know that these are only currently available in Auckland. However, there were two overarching themes - that the home must cater for their child's PWS needs, and a consistent requirement that the home is located within the same region as where the family live. There were a couple of other comments to do with the developmental level of housemates and the need for male support staff.

From our work as a DIAS provider and working with families, we know that parents of children with PWS often need to become experts in their child's diagnosis and work incredibly hard to keep their child healthy. When transition planning begins, parents often ask us what the options are for residential care and express concerns about expertise. The transition process to residential care requires a significant leap of faith for parents and this is one reason why several adults living with PWS remain living at home whilst this may not be in their best interests or the best interests of their family, but these families do not feel there is an alternative suitable option.

Interest from providers in developing PWS specific care

Comments from IPWSO PPCB Conference report by Bjørn Christensen (Community Connections):

"The overwhelming majority of what I heard made me realise that we are not alone in some of the challenges that we face at Community Connections. It seems that the number one thing that we need as an organisation is a broader connection to the PWS community, here in New Zealand, and also abroad."

"Going forward, I think we need to establish a network of professionals who are providing care to people with PWS around the country. I know that we have the PWS association, but Community Connections is a perfect example of a NZ organisation that is currently siloing information, rather than seeking and sharing it."

"Too often we are stretched beyond capacity which leaves us at a disadvantage with supporting our person with PWS. This means that we are not able to give them the kind service/support that they deserve. I am thinking more and more that we need to think about employing a specialist team for their service. We have a large funding contract for the person with PWS we support. It is my opinion that we are flogging a dead horse with the way that we are using it. Could we reduce staffing down to 1:1 but at the same time; a) get a nutritionist b) a nurse to administer insulin c) increased behavioural support? The end goal here being that we are moving towards a specialist PWS home. I am not seeing much of a difference whether we have 2:1 or 1:1. It is about specialisation and competence. I have thought for a little while now that the person with PWS's current living arrangements are

not working for them - could we look at making a much more specialised PWS service that first becomes established with the person with PWS with the end goal being to have other clients with PWS move in too?"

"I would very much like to visit the group homes that we have up in Auckland. I heard a lot of very positive and interesting things about group homes in Europe and the USA. I would be interested to see what sort of model is happening in the services in Auckland."

PWSA(NZ) has also worked very closely with Complex Staffing Solutions who have also expressed interest in developing specialism in PWS specific care.

Conclusion

We would like to conclude by asserting we understand we cannot adopt a 'one size fits all' approach and we need to have flexibility in living options. We also understand that delivery of residential disability supports should be guided by EGL principles, particularly 'self-determination', 'person-centred,' and 'ordinary life outcomes'. However, with few appropriate support options available for adults living with PWS, we currently have limited choice. This is an area of unmet need that needs to be addressed.

There are lots of considerations for families when looking for a residential placement, such as location of the home, compatibility of housemates, the amount of restrictions in place (which should always be the lowest amount necessary), the service offered, and of course, the general feeling of the home, but families should not have to worry if specific needs related to PWS will be understood and managed appropriately.

We would like to see New Zealand develop its PWS expertise, ideally through multi-agency collaboration, but most importantly, we would like to see people with Prader-Willi syndrome being set up for success. All too often this does not happen and when things start to go wrong and inevitable failures occur, recovery can be too difficult. We need to be proactive in ensuring these failures are avoided.