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Significant Individual Support Needs – Supportive Practices and Modified Environments for people living with Prader-Willi Syndrome

Position Paper (Draft)

Prader-Willi Syndrome Australia, July 2020

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EXECUTIVE SUMMARY

This paper is written in support of the Prader-Willi Syndrome Australia (PWSA) statement that:

> *People with PWS find it difficult to sustain life in the absence of significant and ongoing specialized support practices. What constitutes 'ongoing specialized support' will vary from individual to individual. Each case has to be taken on its own merits, depending on the ability of the person to make specific decisions about their own welfare, especially eating.*
>
> *Practices that deliver food security enable a person with PWS to live a more ordinary life. Personal freedom, in line with United Nations Convention on the Rights of Persons with Disabilities, should be pursued at all times. However, practices which modify the support provision environment to enable 'due care and diligence' could potentially have an impact on 'personal choice and control.*

These statements and this position paper will be revisited and updated as part of an international Personal Choice and Control review being undertaken by the International Prader-Willi Syndrome Organization (IPWSO)[i], its 100+ member countries and associated support organisations. The review is aimed at drawing up World Best Practice Guidelines. Such guidelines will encourage least-restrictive practices, while recognizing that some elements of food security and ongoing support practices are required to sustain life, and enable people living with PWS to reach their full potential.

KEY FACTS

- Prader-Willi syndrome (PWS) is a complex multi-system, multi-stage genetic disorder which has only ever been seen to result in early death if appropriate supports are unavailable or withdrawn.
- Personal choice and control can be maximized when appropriate and informed supports are in place.
- Support when around food is essential to maintain life for a person with PWS, due to their genetically altered satiety regulation and changed brain structure and function.

[i] The International Prader-Willi Syndrome Organization (IPWSO), website www.ipwso.org

- Supports around food access must be delivered in a highly sophisticated manner; This is essential to avoid exacerbating other aspects of the PWS condition (such as anxiety-related behaviours of concern), whilst optimizing personal choice and control.

- 'Reasonable and necessary' supports enabled by adequate public/community funding[1] will be the most significant contributing factors to increased life-expectancy for the community of people with PWS.

To enable people living with PWS to live as close to an ordinary life as possible (optimum health and wellbeing), and experience maximized personal choice and control (personal independence), Prader-Willi Syndrome Australia (PWSA) recommends:

Recommendation 1: Specialized and ongoing training and intensive support provision for challenging behaviors must be implemented, to enable maximized personal choice and control

Recommendation 2: Each person should be assessed to gauge their functional impairments caused by over-eating and behaviours of concern, with suitable, least restrictive supports implemented to enable good health and wellbeing

Recommendation 3: A holistic and collaborative approach must be taken to support a person living with PWS

This multi-disciplinary approach is needed because food is available in most settings in life and managing only the domestic environment will not be enough support for the person with PWS. A holistic approach requires a 'lead' provider or nominee to be responsible. Much 'behind the scenes' supporting of daily life is needed to keep anxiety-producing occurrences to a minimum and address any adverse consequences of poor collaboration. Common examples are consistent practices to support dietary planning, or contingency arrangements to overcome daily challenges, like transport.

PURPOSE

This Paper describes the current position of PWSA in relation to significant supportive practices that promote personal choice and control, which may be deemed too restrictive by some jurisdictions.

The necessity for this Position Paper has been brought about by:

- The adoption of the United Nations Convention on the Rights of Persons with Disabilities (UN CRPD) by Australian legislators and by PWSA.
- The advent of the National Disability Insurance Scheme (NDIS).
- The subsequent and ongoing entry of many new service providers into the disability sector.

The need for a consistent and specialist approach to supporting persons with PWS has become possible under the NDIS. NDIS Plans will need to include adequate funding to ensure that service providers can deliver appropriate outcomes. The nature of this rare eating disorder and accompanying behaviours of concern, requires significant and highly skilled supportive practices.

This Paper will help service-providers and regulators to understand the continuum of significant supportive practices that are needed, to ensure that people with PWS can live a full and ordinary life, and exercise choice, control and their human rights to the greatest extent possible. Some people may deem the practices described as being restrictive, but they are in fact life-saving for a person with PWS.

SCOPE

This position paper is primarily designed to inform disability service providers supporting persons with PWS who are in residential specialist disability accommodation or receiving other services from disability providers in the community.

BACKGROUND

PWS is a complex multistage genetic disorder affecting multiple systems in the body. It significantly impacts on behaviour, mental and physical health. People with PWS require cognitive, social and learning 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 PWS.

PWS presents at birth and continues, with significant intensity, throughout life. PWS occurs equally in males and females and affects all races. Hyperphagia[ii], food-seeking and behaviours of concern are compounded by a typically low muscle to fat ratio and low energy requirements, resulting in rapid weight gain. "Consequentially, without careful monitoring and intervention around diet and activity, weight gain can be very rapid, leading to obesity, health complications and early death."[2] "Obesity and its complications are major causes of morbidity and mortality for people with PWS."[3] People with PWS are also at a much higher risk of developing psychiatric illnesses.

PWS is recognised as the most common genetic cause of life-threatening childhood obesity[4], a condition that invariably carries over to adulthood. People with PWS have a flaw in the part of the brain called the hypothalamus. This part of the brain is an important supervisory centre and hormone regulator. The hypothalamus, when functioning normally, registers feelings of hunger and satiety (fullness). For individuals with PWS this does not occur, and most individuals with PWS will never feel full. They have a continuous urge to eat. This part of the brain also controls metabolism of fats and carbohydrates.

In Australia, the general population can expect to live for an average 82 years, however, people with PWS have a much lower life expectancy, with the average life expectancy being less than 38 years of age. This is mainly due to the lack of knowledge about PWS, combined with maladaptive behaviours that cause fatal ill health or accidental death. The risk of early death can be significantly reduced when appropriate, reasonable and necessary supports, environmental modifications, and PWS-specific supportive practices are put in place, including access to food (food security). This is the current accepted international best practice for people with PWS.

Characteristics relevant to significant supportive practice

[ii] Hyperphagia (noun used in medicine): an abnormally great desire for food; excessive eating

- Most people with PWS have a mild or moderate intellectual disability with additional cognitive problems, including poor executive brain function.[5]
- There may also be some physical, sensory and receptive/expressive language impairment.
- They have malfunctioning satiety[iii]. They therefore present as always hungry, will constantly food-seek, suffer from hyperphagia and readily gain weight. They usually display genetically related abnormal food seeking behaviour. They have a reduced resting metabolism related to an altered body composition (muscle to fat ratio), leading to an abnormally lower rate of energy expenditure. Best practice support would include a low energy diet coupled with regular physical activity, necessitated by these physiological characteristics.
- People with PWS experience extreme limitations in their ability to self-regulate in relation to food consumption. Their resulting obesity is not their own choice. International research[6] on the brains of people with PWS has shown that their physiological response to food is different to the neuro-typical person.
- Although there may be variation in the intensity of the food-seeking drive, people with PWS have a preoccupation with food, along with the overriding desire to eat. Therefore, the energy intake of every person with PWS must be regulated by responsible support persons, or they will die prematurely from obesity related, or other, co-morbidities.
- People with PWS have chronic behaviour disturbance. There is "a typical neurobehavioral profile that includes altered intellectual functioning and centrally driven maladaptive behaviours".[7] People with PWS have difficulty managing anxiety levels and are very stress sensitive. Oppositional behaviour may quickly escalate to major temper outbursts, if not supported appropriately by persons with a good understanding of PWS.
- It cannot be assumed that once the capacity of a person with PWS has been built, the supports can be withdrawn. Successful independent living has not yet been demonstrated anywhere worldwide.
- People with PWS find it extremely difficult to extrapolate from one situation to another. Their ongoing, genetic impairment affects executive brain function, satiety and regulation of anxiety and emotions. The risk of early death from overeating or accidental death may never be totally eliminated and supports of some type must always be in place to manage food security (and money), and guard against their increased risk of accidental death.

CORE PRINCIPLES

In preparing this discussion paper, PWSA has identified the following core principles:

1. PWSA endorses the United Nations Convention on the Rights of Persons with Disabilities and support all individuals in their pursuit of maximised personal choice and control.

   Quinn[8] coined the "sword and shield" concept to describe the core role of the United Nations Convention on the Rights of Persons with Disabilities. The Convention offers a 'shield' to defend against intrusion (restrictions) on rights by the State or others, as well as an assertive 'weapon' to demand resources (supports) needed to exercise those rights.

[iii] Satiety: the feeling of having eaten sufficient food

PWSA believes that Quinn's analogy justifies the case for seeking 'reasonable and necessary' NDIS support funding to enable improved personal choice and control by the person with PWS, leading to a more 'ordinary life', when living within complex functional impairments.

In maximising personal freedom, we recognise the need to safeguard the competing rights of individuals to practice choice and control in an environment that is least restrictive.

2. We identify that an ethical "*tension exists between a genetic deterministic perspective and that of individual choice* "[9].

> *"If the person is found to lack capacity, the common-law principles of acting in the person's 'best interests' using the 'least restrictive alternatives' may be helpful. Allowing serious weight gain, in the absence of careful consideration of these issues is an abdication of responsibility*".[10]

3. We identify that PWS is a spectrum disorder and there are subtle differences in the genetic variation causing PWS. Not everyone affected by PWS will have all the characteristics. The degree to which they are affected may also vary. We recognize that there is no one solution to fit all individuals who have PWS:

> *" Adoption of the capacity approach implies that each case has to be taken on its own merits depending on the ability of the person to make specific decisions about eating. There may be some people who never have the capacity to make decisions about their eating while others may be deemed capable, especially if they have been educated about diet, and strategies about reducing weight such as exercise. In those lacking capacity, preventing access to food by locking the kitchen would be justified under the 'best interest' principle."* [11]

It must be recognized that the degree of capacity will span a spectrum, and this range of capacity to make decisions about their eating must be taken into consideration when making decision about the type of support required on individual merit.

In some instances, people with PWS may lack capacity. "Capacity refers to a functional concept, determined by the person's ability to understand, retain, and weigh up information relevant to the decision in order to arrive at a choice, and then to communicate that choice. Approaches to decision-making in adults who lack capacity include anticipatory decisions made through advance health care statements or decisions by proxy based on 'best interests' or 'substituted judgement".[12]

A paper from the UK by Holland addresses the difficulties in assessing capacity in PWS:

> *"It will often be that families and care professionals have very useful anecdotal evidence that begins to explore the difficulties that people with PWS face when applying these executive functions to decisions about food. You do not have to be certain that such deficits exist - rather your judgement about capacity is made on the balance of probabilities.*
> *Evidence is crucial. For instance, keeping weight records can show that a person is gaining weight even though they may say they are not. Keep very detailed records and clear evidence that a serious problem exists."*[13]

4. We identify that PWS is a complex multi-system, multi-stage genetic disorder.[14]

   Research has found that there may be up to 9 distinct phases over the lifetime of a person with PWS, each with different support needs. Therefore, it follows that the management strategies may need to change over time. Strategies used by the person(s) supporting the individual with PWS surrounding food security[15], and associated behaviours, will need to undergo regular review and assessment throughout the person's life. What was relevant food security and behaviour support strategies at 16 years old may not be best practice at 30.

5. Those who are supporting the person with PWS have a common law 'duty of care'[16] to manage the environment to optimize food security according to the needs of the individual.

   > *" The person with PWS will always need food security and behavioural support. The proper goal for someone with PWS is maximal functioning with supports, not independent functioning."*[17]

   Unquestionably food is a major source of stress for someone with PWS:

   > *"Families and professionals [support staff] often mistakenly believe that the patient cannot be happy unless he has as much food as he demands. Because efforts to limit food, if attempted without establishing food security cause increased stress and behaviour problems.*

   Successful Behaviour management of PWS means that uncertainty about food must be eliminated as much as possible."[18]

6. An essential element of successful behaviour management is the skill of the staff to deliver the dietician's diet plan and not defer to the manipulative elements of PWS behaviour which lead to an escalation of syndrome-driven behaviours. A total staff commitment to the consistent management of a dietary plan, as detailed by the dietitian, is critical to the success of individual wellbeing. The success of the implementation is directly related to the level of understanding of PWS and level of skill of the staff in providing support to avoid complex PWS behaviours. This in turn correlates directly with all staff having ongoing PWS training in best practice dietary management and behaviour support.

   The achievement of consistent dietary management and behaviour support allows the person with PWS to minimize anxiety /stress levels and to get on with their life in a productive, ordinary and meaningful way.

HOW CAN LIVING AN ORDINARY LIFE WITH PWS BE ACHIEVED?

Behaviour support of PWS in residential settings includes physical environment modification to optimize food security, according to the needs of the individual.

> *"Intervention* [in the form of support around minimising exposure to excess calories] *is justified on the grounds that it puts people with PWS on more of a level playing field with people who do not face the challenges that they have, as a result of genetic difference."*[19]

It is the policy of PWSA to promote residential practices that pursue maximised personal choice and control. We are also aware that these freedoms will look different for each individual, depending on their capacity for self-determination.

A healthy life in the case of people living with PWS will always involve elements of 'food security' and/or 'food control'.

We support the arguments of by Piers Gooding and Tony Holland in the context of the UN CRPD:

> *"Supported decision-making" does not appear in the CRPD but generally refers to efforts to boost the decision-making of persons with disabilities, offering them resources for making choices from 'good' options about how to live."*[20]

Professor Tony Holland (pers. com.) has said:

> *Support* [to food control/security] *improves capacity and therefore improves engagement. This is in line with what the UN CRPD wants for people with disability. I think we see this in people with PWS - those living in food unrestricted environments are not able to engage for many reasons and may also have limited capacity to understand the risks and to balance those risks in deciding what to do. With the 'support' of food security it can be very different - in that context their understanding may also improve to the extent that they have sufficient understanding of the risks [and] they are willing to consent to the restrictions that a food secure environment implies.*[21]

See Appendix 2 for a table of accepted techniques to support food security and/or control.

A consideration: How does food security differ from food control?

- **Food Security**: Pittsburgh Partnership defines food security for people with PWS as psychological security in relation to a clear plan or understanding for all food consumed including non-calorie food and beverages. There is variation across the syndrome cohort. However, most individuals must have no doubts (or concerns) about what they will be receiving at each meal/snack, and no expectations (or hopes) of obtaining food outside their food plan, which will result in no disappointment that can lead to behaviours of concern. In the case of people with PWS, food security therefore exists when individuals have certainty regarding what they would eat, when they would eat it, and how much they would eat. This reduces stress, anxiety and adverse behaviours, enabling a productive and healthy life. Ensuring food security can involve significant and ongoing individual support and capacity building.
- **Food Control**: Due to the changed brain development of people with PWS, which results in hyperphagia (overeating), as a result of an uncontrollable urge to eat, individuals are rarely able to resist consuming excess calories. If not managed, this consistent over consumption of energy results in obesity and related co-morbidities, eventually reducing life expectancy. In many cases, food control is needed, involving the establishment of physical mechanisms or barriers to prevent additional food access, such as locked kitchens

or pantries. This process is a restrictive practice and should be closely monitored to asses if any restrictions can be eased or removed over time.

See Appendices 1 and 2 for detailed strategies to support food security and food control.

CONCLUSION

Best practice PWS support involves a multifaceted and holistic approach that utilizes a wide range of positive behaviour support and support strategies, and organisation and communication strategies. It involves the input of professional advice and constant assessment and refining of programs and strategies.

All of this is driven by the needs and capacity of the person who has PWS. If the best professional advice from the Support Team deems that locks/physical barriers are required in certain cases, then this should be allowed as this is a proven method to deliver the best practice support for healthy calorie consumption and reducing behaviours that otherwise lead to social isolation.

Other considerations such as prudent planning of new housing builds/modifications is also important. These would factor in design features such as butler pantries and open plan kitchen, so staff can support the healthy eating plan implementation in a robust, PWS friendly environment that enables sufficient personal space.

These support practices around diet may be required in other settings, such as educational or day program environments, together with community access activities.

Food security and food control are a continuum of practice, implemented to a greater or lesser degree, depending on the individual. Restrictive practices should never be viewed as the beginning and end of food security or behaviour support practices. It is too simplistic to talk about management of food security for PWS in terms of just locks and barriers.

NDIS PLANNING

The NDIS offers great opportunities for developing individual capacity in the area of personal choice and control for people living with PWS. However, enabling maximised personal choice and control is complex due to the multi-system, multi-stage nature of the syndrome. The syndrome presents a collection of functional impairments.

A holistic approach to duty of care is required of providers, to support the collection of PWS impairments and enable a balance. A lack of balance leads to a disruption of the person's ordinary life. Therefore, the NDIS will need to provide considerable 1:1, or small group support funding to enable increased freedom and minimise other practices that may be deemed restrictive. The degree to which the individual can participate in self-determination should be assessed on a case by case basis, with the understanding that there will always be at least an element of support needed for healthy food choices.

Since independent living by people with PWS has not been successfully achieved, the planning process will need to address food security and/or food control arrangements in the supported residential setting.

Support and guidance from the care team/allied health/medical professionals to prepare the evidence of functional impairment(s) needs to be sought, documented and submitted during the planning process. The team's support will also be essential in the initial stages of transition to the residential house and after the move is complete. The support person(s) should have regular input and guidance including, but not limited to, the following:

- Dietician
- Endocrinologist
- Psychologist /Psychiatrist
- Respiratory Specialist
- Exercise physiologist
- Parents/Support workers

Their professional opinions should be stated in any NDIS planning documents, to provide clear and transparent guidance to the Support Coordinator about the behavioural practice and physical supports needed.

The planning request should also include "Investigating Housing Solution" funding. This will enable the Participant to work with a Specialist Support Coordinator to search for *appropriate* housing. It also covers completion of a housing plan and funding of various allied health assessments to determine the supports the person with PWS would need when they transition from the family home to the supported residential setting, or transfer from one supported setting to another.

ONGOING REVIEW OF PERSONAL CHOICE AND CONTROL BY PWSA AND OTHERS

PWSA is currently taking a lead role in a world-wide review of personal choice and control (restrictive practices) in relation to PWS. Participating in the review are:

- Individuals living with PWS and their families
- The International Prader-Willi Syndrome Organisation (IPWSO)
- Numerous national person and family Associations
- Major support providers that provide specialist disability accommodation (SDA) to over 600 people living with PWS
- Psychiatrists/psychologists, professors, clinicians and lawyers from various universities and institutions

The purpose of this review is to establish a set of more detailed world best practice principles that enable maximised personal choice and control, thereby minimising, and where possible, eliminating, restrictive practices. Australia has come a long way when one compares practices on an international basis, however, we recognise that more needs to be done to promote personal freedoms in line with the United Nations Convention on the Rights of Persons with Disabilities.

This review will take some time to complete, before international best practice principles can be progressed. In the interim, PWSA has undertaken its own independent review, and will continue to refine our position in relation to supportive practices that are designed to maximise personal choice and control and empower a healthy and full life for people living with PWS across Australia.

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SUGGESTED FURTHER READING

Prader-Willi Syndrome Australia; Providing *Prader-Willi Syndrome support in residential settings* (2016), PWSA website http://www.pws.org.au/

'*Standard of care & Best Practice Guidelines for Prader-Willi Syndrome*, International Prader-Willi Syndrome Organisation https://www.ipwso.org/best-practice-guidelines

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Description of different strategies to be used – a global perspective

Best practice should support a healthy, ordinary and fulfilling life. Supporting a person living with PWS currently demands of the support workers a comprehensive and sophisticated response. It is their responsibility to support people with PWS, while recognising and upholding the person's statutory and common law rights, including a right to receive 'due care and diligence' in the delivery of support by the team.

In order to deliver the standard of support required, the staff and support workers must have a sound understanding of PWS. They must have the confidence and fortitude to manage the multi-system, multi-stage spectrum of PWS behaviours. In addition to PWS specific training, the support worker must have a manageable workload. That enables them to consistently implement the organisational aspect of communication, behaviour support and food security measures that PWS support demands.

The provision of consistent food security is as complex as it is demanding. It requires a well-trained team of support staff, to provide the environment that ensures there is minimal opportunity for excess calorie consumption, and minimised behavioural outbursts that arise from inconsistency.

- Healthy diet: All individuals require education and training to understand the complexities of a healthy diet, including people with PWS and their support team. No exception. Unsupported access to inappropriate food will endanger the lives of people with PWS.
- All staff, including casual support workers, should participate in specific PWS training in the areas of dietary and positive behaviour support planning, effective communication strategies and health/wellbeing support strategies.
- Advanced planning of quality, low calorie meal choices, as recommended by a dietitian.
- Shopping for the house reflects the dietary advice received – this does not involve bulk buying of highly processed food. It does include thoughtful purchases, such as limits to pre-packaged processed foods, purchase of fresh fruit & vegetables in the quantities stipulated by dietitian. Consideration of portion sized when shopping can reduce the likelihood of over consumption.
- Storage of food reflects advice from the dietitian – allowed snacks, allocated daily and accessible (within the parameters of diabetic intake patterns, if applicable). In instances where the person is deemed to lack capacity in relation to making appropriate decisions about their eating, physical barriers (eg. Locks) in the kitchen area may be justified under the 'best interest' principle.
- Meals served by staff should reflect portion sizes as advised by dietician.
- Before leaving the home to eat out, careful planning assists in identifying which foods are compatible with their diet, and thus managing expectations about choice during the outing.
- Scheduled daily activities with the place and time of meals clearly identified helps the person with PWS to engage in a meaningful and purposeful daily program. They are then less likely to be focused solely on food.

- Consistent supervision at meal preparation, eating and after meal 'food clearing' to safeguards against consumption of food being taken from others, or accessed from food storage or scrap storage places. Subject to the individual's hyperphagia drive, all food will most likely require secure stored at all other times.
- Exercise should become an integral part of daily routine.
- Access to money may need to be supervised and receipts kept to support healthy living in line with their dietician's advice.

The support team needs to encourage and monitor food access arrangements at all times. The goal is to obtain the agreement of the person with PWS to the arrangements. A contract based on informed consent is sometimes appropriate. For example:

> "I (the person) do not like being overweight and want to return to a normal weight range/X kilo. You (support person,) can support me in managing my diet in a food secure environment, thereby assisting me in reducing the stressors associated with food in my life. I will try to abide by my diet and the agreed food security routine and measures, which are (list measures)".

Such an agreement must be consistently applied and the person with PWS must be supported emotionally and socially in all settings, to maintain their consent, in the face of many temptations.

The provision of appetising, well presented, quality food, in small serving portions, at regular meal and snack times, has been shown to assist emotional wellbeing for the person living with PWS.

Adherence to consistent food routines can reduce friction in most cases. What these food security measures look like, will vary from individual to individual and will need to be established, maintained and reviewed, with the support of the individual's Care Team. 'Food security' is a psychological state of mind. When food security is not achieved, other management strategies will have to be explored including the use of locks.

It is essential that this support is implemented with appropriate 'duty of care'.

The restriction of choice of foods (i.e. low fat, low calorie foods) as recommended by a dietitian, has many precedents in everyday practice in the community. Disability support houses/schools/day care centres regularly restrict food choices for people who have coeliac or peanuts/seafood or other allergies. Therefore, the restriction of food choices to people with PWS should not be a contentious issue. Many schools/residential support houses/day-care places enforce rules prohibiting the bringing of certain foods, based on medical/dietician's advice due to the fact someone in that house or school has a severe allergy to certain foods. Excess .food (callories) of any type is, over time, life-threatening to those with PWS.

The PWS low-calorie diet is comparable to the needs of others with specific requirements in everyday practice in the community. If this diet is not adhered to, obesity and related co-morbidities will risk the health and wellbeing of the person with PWS. This is similar to an anaphylaxis reaction risk for the person with the allergies; albeit one type is fast acting (anaphylaxis) and the other slow (morbid obesity).

While PWSA does not necessarily promote a specific method of enabling food security, numerous different practices and devices exist across the globe. Four alternative practices are outlined below.

It should be noted that some of these programs will necessitate considerable increased support provision and capacity building. Existing practices, if working successfully for the wellbeing of the person with PWS, should be maintained until new programs for food security are fully developed. Transition to a new support practice should only occur once residents have been consulted, capacity of the residents has been developed, the program has been fully researched and developed, support workers have been fully trained and the support organisation is satisfied that they have the resources to implement the new program in an effective and sustainable manner.

The primary aim of implementing an alternative food security practice, such as those cited below, would be to enable a least restrictive practice, while maintaining good health and wellbeing. Choosing the most appropriate option will rely heavily on individual capacity and organisational capacity.

Alternative food security practices include:

- **Pittsburgh Partnership**, Linda Gourash, MD; Janice Forster, MD[A], suggest a food security support strategy:
    - "No doubt" The person with PWS is able to relax and think less about food when they know the set times when meals arrive. Advanced planning provides them with expectations which will be reliably fulfilled.
    - "No hope" of getting anything different from what is planned. This requires more measures to be put in place to ensure they are not successful in acquiring food. Chances to obtain food are stressful so therefore should be eliminated. This can require additional supports to be put in place to ensure the person is supported in successfully maintaining their diet, rather than successfully acquiring additional food calories. Implementation of appropriate supports will depend on the individual's history and capability of unplanned food acquisition.
    - "No disappointment" No false expectations. What you promise you deliver. This will reduce the number and intensity of angry outbursts.

- **Prader-Willi Syndrome Institute Germany** (PWS-ID), Dr Norbert Hödebeck-Stuntebeck and Dr Hubert Soyer[B] promote a food security program that encourages individual food access training:
    - "We say 'Yes we can, yes we can, yes we can!'. We all have to deal with doubt, with hope and with disappointment because these are conditions of normal life. And we must prepare/help people with PWS to become competent for new and sudden changes, for visions and for situations of disappointments, so that they learn to develop coping strategies for such situations. If we do not give them the chance to come in contact with such situations, they have no chance to learn to manage them in a more and more proper way and become more and more independent from others.

[A] Founding Directors, Pittsburgh, Pennsylvania, USA www.pittsburghpartnership.com
[B] Directors, www.pws-id.de

Over the years, we saw that people with PWS are much more self-controlled than professionals or parents think [is possible]." Norbert Hödebeck-Stuntebeck

- **Arc of Alachua County,** Mark Swain and his associates[C] at Ark have implemented a system of food dollars based around calorie content. Individuals are supported to manage their own daily calorie intake by trading food dollars for food. This method enables increased personal choice and control within a set daily calorie target.

- **Grankoglen,** Carina Knudsen and Mai-Britt Rosengreen[D] in Denmark:
  - Danish experts believe that the person with PWS cannot be cured of their hyperphagia, so they should be relieved of the daily stress of worrying about food, thereby freeing them up for other, more fulfilling life pursuits.
  - Grankoglen delivers 'restaurant service' as their preferred method of food security. Professional cooks are employed to undertake menu planning, food preparation, food serving and food service to the table.

Note: Some support providers in Australia may find this Danish concept challenging in relation to personal choice and control, however, it is worth noting that people living with PWS in Denmark live more than 50% longer on average than those living in Australia

Summary

Best practice PWS support involves a multifaceted and holistic approach that utilizes a wide range of positive behaviour support and support strategies, and organisation and communication strategies. It involves the input of professional advice and constant assessment and refining of programs and strategies.

All of this is driven by the needs and capacity of the person who has PWS. If the best professional advice from the Support Team deems that locks/physical barriers are required in certain cases, then this should be allowed as this is a proven method to deliver the best practice support for healthy calorie consumption and reducing behaviours that otherwise lead to social isolation.

Other considerations such as prudent planning of new housing builds/modifications is also important. These would factor in design features such as butler pantries and open plan kitchen, so staff can support the healthy eating plan implementation in a robust, PWS friendly environment that enables sufficient personal space.

These support practices around diet may be required in other settings, such as educational or day program environments, together with community access activities.

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Food security and food control are a continuum of practice, implemented to a greater or lesser degree, depending on the individual. Restrictive practices should never be viewed as the beginning and end of food security or behaviour support practices. It is too simplistic to talk about management of food security for PWS in terms of just locks and barriers.

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Appendix 2

Food security management for PWS in mixed disability residential support houses

Tensions created by PWS food security/PWS daily exercise routine in mixed disability support houses	Suggested approach/solution	How can this be achieved	Extra resources required
PWS specific diet is difficult to implement in a house where other residents don't have specific dietary requirements	Given that support workers have a duty of care to provide all residents with a healthy well balance diet – a Healthy Living approach for all residents should be a long-term goal. A good PWS diet is a healthy living diet with smaller servings. It takes time and skill to change poor dietary habits, particularly for a person with PWS. But a skilled support worker with the support of the dietician can coordinate a transition to, and maintenance of a healthier diet	Specific training for support workers on how to present and prepare a well-balanced diet that is appetising. Healthy cooking methods should always be used in any SDA, i.e. fresh, without additives, managed calorie intake, minimal use of deep frying, few calorie rich sauces and condiments. Outings should not involve unhealthy food choices; a healthy range of options should pre-identified and built in by the dietitian.	Dietician should train all residents and support workers on site, about healthy cooking methods, food storage, serving sizes, including appropriate healthy snacks. Regular servings of fresh fruit and vegetables should be included in the diet. Support workers and residents may initially need the assistance of a dietitian to develop a file of dietitian approved recipes with attached shopping list for each recipe. Support workers must set aside time to refer to, and understand, an individual's meal plan and then adhere to it consistently, e.g. cut lunches healthy options

Tensions created by PWS food security/PWS daily exercise routine in mixed disability support houses	Suggested approach/solution	How can this be achieved	Extra resources required
		Smaller plates can be used to make servings look more generous. Involve the residents in the menu planning process, utilising either/or choices to enable healthy options. Develop a weekly menu planner in consultation with the residents, including prepared lunches & snacks Encourage residents to help prepare their meal on a roster, according to capabilities and with support	Support worker hours should be set aside for 1:1 food preparation, with no other residents in proximity - can lead to stress and outbursts for the person with PWS
Many house staff are too time poor to shop in accordance with the dietitian's plan, resulting in minimal time for a planned nutritious menu	Healthy menus should start at the point of purchase; that determines the quality of the resident's diet. The goal of the support worker should be to purchase fresh quality food in the quantities	Weekly menu and dietary requirements of residents should inform shopping. Only food on the list should be purchased. Where possible, purchase items in specific, small serving	Dietitian and House coordinator to work with the staff to fine tune the purchase of foods for the house in line with a healthy eating policy. Staff, support workers and residents to develop a clear understanding of

Tensions created by PWS food security/PWS daily exercise routine in mixed disability support houses	Suggested approach/solution	How can this be achieved	Extra resources required
Shopping is done in a rush with no preplanning; pre-prepared items are bought in bulk to save time. Processed 'treat' foods/snacks are bought in high rotation. This can result in quantities of less healthy food being stored at the house and potential food wastage.	required for the weekly menu planner	sizes – yogurts, snack packs in 45 gm boxes	food labelling, energy content and ingredient lists.
Food purchased in bulk can often be stored in pantry/cupboards that are readily accessed by all residents. Additional food could be consumed by	Minimizing the quantity of food purchased per shop - focusing on quality of the food and correct storage of foods will reduce the amount stored. If the food is presented in an attractive and appetizing way, the healthy option will become	Allocating the number of snacks per day that are recommended by the dietitian Have fruit readily available in quantities recommended by the dietitian	Dietitian & house supervisor audit how food is stored in the house All staff identifies and records any breeches of food security for the person with PWS and problem solve strategies to rectify. Staff informed about, and prevent further food 'leakage' by being

Tensions created by PWS food security/PWS daily exercise routine in mixed disability support houses	Suggested approach/solution	How can this be achieved	Extra resources required
people with PWS, with the practice continuing repeatedly without being noticed by support workers.	routine once the change to routine is bedded down. Change for anyone takes time, particularly if they have an intellectual disability. Realistic timelines and outcomes must be factored into any change to food security routines. In a case where the person is deemed not to have the capacity to make decisions about their food intake, stress can be alleviated, but not eliminated, if pantry and fridge are locked. Note: Other residents can experience disadvantage due to this practice.	Encourage the residents to eat at the table together rather than in their rooms If healthy options are available this increases the likelihood of the residents choosing to eat these healthy options, particularly if they are praised for doing so Introduction of locks and barriers should be a considered and measured approach. How they are implemented into day to day house routine must be carefully planned and monitored	more vigilant and diligent about practices
Residents of the home can experience disadvantage in enjoying food-related celebrations and	Every resident regardless of whether they have PWS or not has the right to socialise and participate in community events	Support the residents beforehand to rehearse how they can make healthy choices when they go out	Work with the Dietician to understand calorie banking or calorie trading Support workers then monitor and record the excess calories taken in

Tensions created by PWS food security/PWS daily exercise routine in mixed disability support houses	Suggested approach/solution	How can this be achieved	Extra resources required
eating out due to the dietary requirements of the person with PWS			at a celebration and calculate what has to be cut back over the next few days in exchange. They must inform staff on following shifts of the temporary arrangements, so it is reliably applied. Support staff apply a visual representation of the trading arrangements on the wall to help them understand what's being negotiated, and stick to the trade once commenced
When the person with PWS has nothing to do they actively seek food	The daily planner for a person with PWS should be fully allocated with engaging and purposeful tasks - limited downtime should be a primary goal of any support/daily activity plan	Household duties/activities should be included to minimize and to redirect away from food seeking - get the mail, set the table, gardening tasks	House coordinator to set up a list of tasks for each resident Interesting and engaging activities that involve community participation & don't involve food should be emphasized
The person with PWS keeps asking the other residents for food or takes food from other residents' bag or rooms	This is a code of conduct issue. If the House Code of Conduct states very clearly – no sharing of food & no entering other residents' bedrooms without	Support workers to have a range of management and support strategies to distract/redirect to avoid food seeking	Team meeting to communicate any issues and revise strategies. If behaviours continue, seek the advice of psychologist

Tensions created by PWS food security/PWS daily exercise routine in mixed disability support houses	Suggested approach/solution	How can this be achieved	Extra resources required
/kitchen and this causes tension within the house	permission, then this becomes an expectation that this behaviour is not accepted All residents of the house should be fully aware of the house Code of Conduct and what it means for them in day to day application.	All the support workers consistently respond to incidents with appropriate consequences that have been discussed prior to incidents Staff know and apply particular consequences relevant to the situation, without the person with PWS escalating to behaviours of concern	Highly trained and skilled staff to apply the supports or interventions. If the incidents escalate this may require a review of the living arrangements
The residents are not interested in any exercise, favouring a sedentary lifestyle such as watch TV and play on iPad	Support workers to model good exercise habits; residents to be given the opportunity to participate in a range of activities involving exercise both formally and informally Walking is encouraged rather than being driven everywhere to gradually increase their exercise pattern	Support workers to help the residents organise and plan events that include walking swimming, team sports	Exercise physiologists Local government sport participation officers can assist with the introduction of regular exercise into the house routine Person supported to participate in community access activity at local gym Skilled support workers can model habits for the person with PWS so

Tensions created by PWS food security/PWS daily exercise routine in mixed disability support houses	**Suggested approach/solution**	**How can this be achieved**	**Extra resources required**
	Membership of local gym should be encouraged and supported		that, for example, they regularly participate in a daily exercise routine before dinnertime. Staff should exercise *with* the person, modelling and appreciating the fitness activity
Residents can store and eat food in their rooms. The person with PWS can take food and consume more than dietitians' recommendation without being observed and over-eating goes un-noticed.	Discourage food being kept in rooms on a health & cleanliness rationale In some case this will be included in the house Code of Conduct, where it clearly states what can or can't be consumed in rooms, eg. drinks can be consumed in rooms but not foods	Support worker to encourage and support the residents to take pride in their room and keep it clean and free of food Support Staff to be aware of what they do and model the same respect for eating arrangements	Support worker must negotiate alternative storage arrangements if food is brought into the house by a resident Staff must educate residents about the arrangements
Person's weight is going up despite appropriate food access in the home. Note: Excess food can	Support workers to support individuals in monitoring weight by recording on a regular basis Key worker to analyse weight gain pattern to support the	Support worker to monitor client behaviour upon returning from community Evidence of additional food consumption should be	Staff must liaise with other providers (e.g. day program) to understand what food can be accessed there and negotiate new food security practices at the other location

Tensions created by PWS food security/PWS daily exercise routine in mixed disability support houses	Suggested approach/solution	How can this be achieved	Extra resources required
cause bowel blockages, stomach rupture and death	person in avoiding rapid weight increase with subsequent health and mobility deterioration, identifying potential food sources	noted and monitored, e.g., food scraps, packaging, food spills on clothes	Records can be maintained to monitors patterns. Additional supports should be initiated in community access locations if additional food is being accessed
Person with PWS wakes up at night, wanders to kitchen and takes food, or non-food items to eat (e.g. rubbish)	Introduction of the use of assistive technology and/or additional active night support or physical barriers such as locking the kitchen at night	Introduce assistive technology such as light motion sensors or motion sensor mats to alert staff of night- time wandering Note: Support may have to be upgraded to active night roster to achieve the benefits of this technology. If problem persists it may be necessary to lock the kitchen, fridge and pantry, at night.	Use of Assistive technology Roster active night support staff to support the person with PWS, to redirect them away from food seeking Team meeting to communicate any issues and revise strategies. If behaviours continue, seek the advice of psychologist and OT, as needed Highly trained staff to apply the interventions
Person with PWS requests that the kitchen cupboards be locked for their own	Support the person with PWS in their choice		Seek the advice of psychologist and OT

Tensions created by PWS food security/PWS daily exercise routine in mixed disability support houses	**Suggested approach/solution**	**How can this be achieved**	**Extra resources required**
safety and peace of mind Note: Other residents may not want this			This may require a review of living arrangements Seek OT and Specialist Support Coordinator reports
Support workers use of junk food or snacks as reward for compliant behaviour	Discontinue the practice of any food rewards Use intrinsic and more suitable extrinsic rewards	Ongoing training for staff in the importance of not using any food as management tool House coordinator support the support workers to use more suitable rewards	Seek dietitian advice Review staff training about appropriate positive behaviour support strategies Organise upskilling of staff and ensure better practices are applied