



MANAGING DIET AND THE FOOD ENVIRONMENT

UNDERSTANDING THE NUTRITIONAL PHASES OF PWS

In the past, PWS was described as a 2 stage syndrome - failure to thrive followed by hyperphagia. It is now known that the changes in appetite and weight gain which occur in PWS are more gradual and complex.

<u>Phase</u>	<u>Ages</u>	<u>Characteristics</u>
0	prenatal-birth	Decreased fetal movements and lower birth rate than siblings
1a	0-9 months	Hypotonia with difficulty feeding and decreased appetite
1b	9-25 months	Improved feeding and appetite; growing appropriately
2a	2.1-4.5 years	Weight increasing without appetite increase or excess calories
2b	4.5 - 8 years	Increased appetite and interest in food, but can feel full
3	8 years - adulthood	Hyperphagic (abnormally increased appetite); rarely feels full
4	adulthood	Appetite is no longer insatiable (only very few adults)

Parents of children diagnosed in infancy have the opportunity to instil diet modifications and healthy eating habits well before the child's appetite or interest in food increases. As a result, when phase 3 begins it is often less severe in families who have implemented early intervention measures.

The age at which phase 3 begins varies, but at approximately 8 years, an intense preoccupation with food can develop accompanied by food related anxiety. A child may rarely feel truly full; they may eat more than they should if opportunity arises, seek or steal food, eat from unsavoury sources or in rare cases, eat inedible items. However, the intensity of hyperphagia symptoms varies in individuals and some families report that their child never takes food and they don't need to lock cupboards.

WHY DIETARY MANAGEMENT IS ESSENTIAL IN PWS

It is important to understand the **high risk of obesity in individuals with PWS due to the high fat to muscle ratio causing reduced energy expenditure**, and that this risk is increased by the difficulties faced in imposing a restrictive diet on a person who can be preoccupied with or obsessive about food and may be feeling **consistently hungry**. To maintain a healthy weight at phase 3, an individual with PWS usually requires **dietary restriction of approximately 50-70% of RDA** and food cupboards are typically locked to prevent food related anxiety and keep the person with PWS safe. An individual with PWS can rapidly gain weight over a short period of time if not supervised and it is very difficult to lose again due to **reduced exercise capacity**. It is essential to support the strategies families have in place, especially as their efforts can be additionally compounded by the behavioural challenges of PWS.

There is also an **elevated risk of diabetes in PWS so it is advised to avoid foods with a high glycaemic load** – foods high on the Glycaemic Index cause spikes in blood glucose and insulin. Food also poses a serious health risk if a person with PWS were to gain unsupervised access to a significant quantity of food. A person with PWS **could quickly consume a large quantity of food resulting in choking or a perforated stomach lining**. Unfortunately, those who do not understand the reasons for dietary management in PWS often see a slim, healthy person and cannot understand why there is a need to be strict about food and diet.

CURRENT DIETARY ADVICE FOR PWS

It is important to know that there is no single, correct way of feeding a person who has Prader-Willi syndrome – different approaches work for different individuals and families. To date, the underlying drive to seek food in PWS is not fully understood. Whilst several diets are recommended for weight management, very little is also known about the scientific benefit of these for individuals with PWS. Traditionally, parents are usually advised to provide a low calorie (typically low fat) diet which can be adjusted for age, growth and height by a dietitian. However, only one published study so far has examined macronutrient amounts (carbohydrate, fat and protein) for PWS:

Dr Jennifer Miller and her team at the University of Florida trialled a reduced energy intake diet with the following macronutrient distribution: 45% carbohydrate, 30% fat and 25% protein with at least 20g of fibre per day. They found that the children whose parents had adhered to the macronutrient distribution of the diet had lower bodyfat and weight compared to those who had only followed energy intake (calorie) recommendations (J Hum Nutr Diet. 2013 Feb;26(1):2-9. doi: 10.1111/j.1365-277X.2012.01275.x. Epub 2012 Oct 18.)

Dr Miller currently advises that a combination of a **reduced carbohydrate diet (45% or less) with 25-30% protein and increased exercise, results in improved body composition with a decreased need for calorie restriction.** She recommends choosing good quality carbohydrates and foods with a low Glycaemic Load.

More trials are currently underway. One is comparing a low carbohydrate versus a low fat diet in PWS with the hypothesis that the higher fat diet will decrease hyperphagia and increase satiety (feeling of fullness). Other trials are examining modified atkins and modified ketogenic diets for PWS. There is much interest in the effects of dietary interventions on weight, body composition, hyperphagia, energy and behaviour, but until the optimal composition of macronutrients for PWS has been scientifically determined and any long term risks considered, we can only endorse the reduced carbohydrate 'Miller Diet 2013' and the similar [Mediterranean Diet](#) trialed at Latham Centers USA and recommended by Melanie Silverman, renowned dietitian with expertise in PWS.

Please discuss dietary choices with a dietitian. Dietary advice will vary amongst dietitians - very few dietitians in New Zealand currently recommend low carb / high healthy fat diets, but most will support your choices. **A dietitian will help with recommending foods, calculating energy requirements and looking for any dietary deficiencies.**

It is important to ensure the PWS diet is a **balanced diet with sufficient micronutrient intake (vitamins and minerals)**. Therefore, it is recommended to omit or limit 'empty calories' (foods of low nutritional value) from a restrictive PWS diet, such as highly processed foods and treat foods. **Sufficient dietary fibre is also important to avoid problems with constipation, common in PWS.**

DIETARY MANAGEMENT STRATEGIES

Sweet foods, treats and rewards

Current medical advice for PWS is to follow a low carbohydrate diet and **avoid sweet foods**. It is known that continual blood sugar spikes increase diabetes risk, but they also do little for satiety. After eating sweet foods we are left feeling hungry again shortly afterwards. Foods containing artificial sweeteners are no better because our body doesn't receive the expected calories that our brain associates with the sweet taste and therefore craves more. Brain scan imaging in PWS has

also revealed that people with PWS already experience a higher 'reward' from food so this is exacerbated by consuming sweet foods which are known to be addictive, high reward foods. Some families decide to avoid sweet treats altogether and only use non-food treats. Others decide to make or buy only healthy foods as treats. If occasional food treats are allowed, it is important these are scheduled so not too many are given and anxiety is not raised by uncertainty about whether they will be received. Be prepared for the expectation that any treats given will be received again, and an excellent long term memory means they will not forget! **Ask the family for their rule about treats and support them in their plan. Do not provide any unplanned extras.** It is preferred if food is not used as a reward because the anticipation of the reward raises anxiety. Any changes in diet or food rules can also raise expectation that these will happen again - consistency minimises future battles over food.

Understanding food groups and food labels

Good carbs are low GI vegetables and low fructose fruits and bad carbs are starchy vegetables and refined carbs lacking in fibre. Foods high in protein are good satiety foods, i.e. chicken, fish, lean red meat, eggs, Greek yoghurt and nuts. Although oats are mainly carbohydrate, they contain more protein than other grains, plus a good dose of fibre. Beans are also mainly carbohydrate, but contain protein and fibre, ranking high on satiety scales. The fats to avoid are trans fats and processed oils, but foods containing healthy fats should be included in a PWS diet, i.e. fish, nuts, seeds, avocados, olive and coconut oil.

Check the carbohydrate and fat content of foods. Often foods marketed as 'low fat' have a higher carbohydrate content to make them taste better. The energy content (Kilojoules, KJ) may also be higher than the full fat version too. You should also check for low sugar content. Overall, it's best to avoid highly processed foods containing refined grains, hydrogenated oils and high fructose corn syrup. Highly processed foods and 'bad carbs' tend to just make a person with PWS feel bloated or lethargic, possibly due to poor carbohydrate metabolism.

Which drinks are best?

Thirst awareness is also controlled by the hypothalamus and often affected in PWS, therefore drinking water often needs to be encouraged. Drinks like water, naturally flavoured water, unsweetened tea and coffee are preferred to fruit juices and sugary drinks. Fruits can be high in fructose but when eaten as a whole fruit, the fibre content reduces the blood sugar spike - this doesn't happen with juice. Diet drinks are often not ideal because they usually contain artificial sweeteners and encourage a preference for sweet drinks.

The importance of exercise

Restricting diet is not the only way to maintain a healthy weight. Regular exercise is vitally important because in addition to maintaining energy balance (energy in=energy out), it also builds strength. **An improved body composition (increased muscle mass) and metabolism reduces the amount of dietary restriction required.** Individuals with PWS can be reluctant to exercise, so it's a good idea to build regular exercise into the routine early on.

The importance of consistency

Planned and scheduled meals and snacks reduce food related anxiety and the repetitive questioning that might occur if there was uncertainty about what will be eaten, when and how much. Individuals with PWS thrive on routine and prefer predictability, but there are times when flexibility is needed, so it's a good idea to have healthy snacks readily available so they can still be

reassured about food when a change of plan or 'eating on the go' is necessary. **Having clear food rules are also helpful, i.e. what is the rule for seconds or dessert?**

Ask all those who work with or care for the individual with PWS to be consistent with the routines and rules you have in place. Asking others to follow the principles of 'food security' (see below) are a good way to ensure consistent management of the food environment.

Planning responses and being prepared

Have a consistent approach for how you will deal with requests for food – they will keep asking if they think there is hope. Plan your response for an occasion when food is taken, whilst keeping in mind that they are unable to control the drive to eat. If they are encouraged to tell exactly what they have taken, this will help keep them safe in the event of an unknown amount of food being eaten.

It is also useful to **be prepared with some methods for making less seem more and avoiding fairness complaints**. Some tips from carers are:

- using smaller plates to decrease portion size whilst also filling the plate
- bulk filling a plate with some good foods like salad
- serving food out of sight to avoid comparisons
- offering foods which you know will take longer to eat so it seems like more
- not eating in front of the person with PWS when they can't.

Teaching the person with PWS about nutrition

Help the person with PWS learn about making good food choices. They will be able to understand the reason for their special diet and know that other people need special diets too. Until a cure is found for hyperphagia in PWS, it's likely that the food environment will need to remain controlled, but many older individuals with PWS try desperately hard to avoid temptation and make the right choices - some become quite the nutrition expert!

Celebrations and social events involving food

Considering the needs of people with PWS at events involving food is no different to catering for people with food allergies or intolerances. Most individuals and families will not mind PWS health needs being shared with others and the PWSA can help with providing information if needed.

Make a plan with the individual with PWS and their family for celebrations and events involving food. If forewarned that there will be shared food, they have the option to either eat a planned alternative that you take along or to make allowance by adapting dietary intake before or after (there will need to be a limit on how much is allowed and carers will need to know how much was eaten.) With help, the person with PWS may learn to make good food choices at social events. Eating out at restaurants, attending parties and social occasions need not be avoided if you plan ahead. **Rehearsing or role playing food situations so the person with PWS knows exactly what they are allowed can be effective.** Of course, it's more helpful if their needs have been considered - encourage others to avoid buffet style events and keep food out of sight.

The availability of food can be relentless with birthdays, shared lunches, fundraising bake sales and sausage sizzles etc. It would provide great relief to individuals with PWS and their families if **alternative food free celebrations** were sometimes considered. With a general population shift toward reducing sugar in our diet, **new ideas for food free and healthy fundraising are emerging.** A healthy eating policy on food in the care environment would be of benefit to all.

Food security

Food security is not just about keeping food out of reach – it is also about keeping food out of sight and mind. **Removing temptation to take food also removes the thought of food** which is preoccupying all thought. By taking that thought away, an individual is able to focus on other things. Many older children with PWS actually ask for food to be locked away because it relieves the pressure of resisting urges and they want to follow their diet.

Keeping food out of mind requires more than just storing it out of sight. Food security is achieved by ensuring there is **no uncertainty about what and when food will be available and there is no hope of any extras outside the schedule**. No doubt and no hope leads to no disappointments - this method is taught by Drs Forster and Gourash at The Pittsburgh Partnership who coined the phrase 'food security' for PWS.

NO DOUBT

There is a predictable routine in which food is scheduled. (Focus on the sequence rather than the time of meals.) The person with PWS is able to relax and think less about food. Predictability of the menu also reduces anxiety.



NO HOPE (or chance)

There will be no unplanned extras outside the schedule and no opportunities for gaining access to food. Chances to obtain food cause stress. Food acquisition capability varies and can fluctuate, so modify the measures that need to be taken.

NO DISAPPOINTMENT

The person only has expectations that will be reliably carried out. No other expectations have been raised so there is no disappointment when these are not realised or fulfilled.

Tips for achieving food security

- ✓ The individual is aware of the schedule and where eating food fits within the schedule.
- ✓ The individual has clear knowledge of the rules surrounding mealtimes.
- ✓ The individual knows which food is for which snack or meal if they have a packed lunch.
- ✓ The individual is forewarned and reassured if the schedule is disrupted for any reason.
- ✓ There are no suggestions or promises of extras, and no surprises.
- ✓ Peers, staff and involved community members know not to offer unplanned food to the individual with PWS.
- ✓ Treats are not used as incentives or unplanned rewards.
- ✓ Food is not used as a consequence, i.e. the withdrawing or delaying of food.
- ✓ Supervision is provided during travel and transitions if required and around food sources.
- ✓ Opportunities for access to food are eliminated. This may require an inaccessible or lockable area for food storage.
- ✓ The individual cannot gain unpermitted access to money or have an opportunity to buy unpermitted food.
- ✓ Plans are in place for any special occasions and celebrations involving food and the individual is aware of the plan in advance.
- ✓ The individual is supported with making appropriate food choices when this is allowed.
- ✓ There are no consequences if food is stolen – encourage the sharing of what was eaten.