

Dietary management for adults with PWS



Introduction

Limiting food intake to maintain the best level of health can be a great challenge for both adults with PWS and their parents and carers. However, it must be borne in mind that the consequences of severe obesity can place many limits on a person's quality of life, and their health will almost certainly suffer.

Effects of obesity

Besides life-threatening health issues, obesity can affect other areas of a person's well-being. Some of the most common outcomes of severe obesity in PWS are:

Illnesses

Diabetes (usually non-insulin dependent, but sometimes insulin is needed to control the condition), heart problems, breathing problems, sleep apnoea (cessation of breathing for short periods whilst asleep), oedema and lymphoedema (water retention), which can lead to cellulitis, a serious skin infection which can cause blood-poisoning.

Limitations on lifestyle

The ability to move about, already limited by poor muscle tone, becomes very difficult. Problems are experienced in using public transport, getting into cars, cinema seats, using the toilet, etc. Ordinary wheelchairs will not take the weight of a very obese person. Sleeping during the day becomes more frequent. Clothes become increasingly difficult to find.

Psychological and behavioural effects

Self-esteem may become very low and depression may result. Sleep apnoea causes breaks in the sleeping pattern, resulting in irritability, tiredness and increased behavioural problems during the day.

Currently, the only effective long-term prevention or treatment of obesity in PWS is a low-calorie diet, coupled with regular exercise.

**Low calorie =
low energy**

Although we commonly speak of low calorie foods, dietitians and other medical experts use the more correct term "low energy". Energy is measured in kilocalories (kcal) or kilojoules (kJ) - both are often featured on packaging, although kJ is becoming increasingly more common.

1kcal = 4.184 kJ



**PLEASE
NOTE!**
This

information does
not replace
guidance from
any health
professionals in
individual cases.

Eat and be happy?



Most parents and carers would agree that their son or daughter's quality of life and happiness does not begin and end with food, and that support should be offered to help them realise this. However, there may be some parents and carers who feel that their son or daughter, or the person they care for, will only be happy with unlimited access to food, or who, having tried everything within their power, just feel like giving up.

The welfare of your son or daughter should be at the centre of your decision-making and there are perhaps a few considerations you should think about before taking the step of withdrawing any sort of control.

- Does your son or daughter really want to become so overweight that they cannot move, and so overweight that they will die within a few years? Have you discussed this with them?
- What are the opinions of other family members, care staff and social workers?
- Is coping with the physical needs of an extremely overweight person, who will become less and less able to perform any actions for themselves, including personal hygiene, any easier than trying to support and help them with their diet and behaviour?
- Should a review of the person's capacity to make decisions about food under the **Mental Capacity Act** be considered? See <https://www.pwsa.co.uk/assets/files/Mental-capacity.pdf> or contact the PWSA office for a copy of this article.

Only you and your son or daughter can decide, but perhaps you need to think of the implications first, and to consider implementing some of the alternatives given below, before you give up entirely.

Supporting your son or daughter to maintain a healthy weight



If a person has Prader-Willi syndrome, keeping weight to a reasonable level is doubly difficult. They have the same nutritional requirements as everyone else, but because of poor muscle tone (and usually short stature if they have not received growth hormone treatment), people with PWS require fewer calories than most to maintain stable weight levels - yet they almost always feel hungry.

Most parents and professionals would agree that it helps the person adjust to the problems imposed by the syndrome if the person has been used to living on a restricted calorie intake since childhood. However, increasing levels of independence in adulthood can sometimes undo much of the good work done in childhood. Also, if adults have been diagnosed in their teens or later, they may need a great deal of support to adjust to a lower calorie intake if they have not previously been used to this.

Not everyone with the syndrome has the same problems around food. However, most have food as one of their main interests in life - so much so, that some people seem to be continually asking when the next meal is due, what it is, what everyone else is having, and so on.

Some would never steal food from shops or other people; some unfortunately will. A few are capable of preparing their own meals with little or no supervision; for others, time spent in the kitchen is time to eat as much as they possibly can, and even to secrete food on their person for later.

Continued ...

Ensuring a good nutritional balance

If you are in doubt about how to ensure that your son or daughter is getting all the nutrients they need, or are unsure about how many calories they require, you should get a referral from your GP or consultant to a registered dietitian.

The PWSA UK booklet, "Healthy Eating with Prader-Willi Syndrome," gives more information about the dietary and nutritional needs of people with PWS. <https://www.pwsa.co.uk/about-pws/publications/>

Supporting your son or daughter to maintain a healthy weight (continued)



Some people will eat items such as pet food, frozen food, garden produce (including potentially poisonous berries) and food from garbage bins, whilst others will not.

So far, no-one has been able to account for the reasons for these differences in eating behaviour, eg whether it is due to different levels of damage to the hypothalamus, or to different social, cultural and family influences. What is certain is that all will require some level of support.

The issue of self-control becomes all-important for adults with PWS. If control over food at home is impossible for the person with PWS, control in many other environments, such as shopping and going out independently, is also very limited.

This lack of personal control in so many areas of life may be one of the contributory factors to behavioural problems. It is therefore important to give back some control to your son or daughter in other ways, for instance, in agreeing a contract about food and diet, or consulting them about menus for the days or weeks ahead.



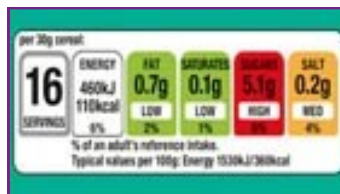
Some people become very upset by the word "diet", so you may need to use another term such as "healthy" or "sensible" eating. Responsibility and control can be provided in other ways,

such as asking your son or daughter to do their own laundry and keeping their room tidy.

Other ideas for supporting the dietary needs of people with PWS are given in the following pages, and range from almost total segregation from food to greater degrees of self-control.

Looking at food labels

Look at food labels carefully. Remember that "low fat" and "low sugar" do not always mean low energy (low calorie). Make sure you read "per portion", "per pack", "per 100g" etc correctly. For example, the label on a product may say 70kcal per 100g, but if there are 250g in the whole pot, the entire contents would amount to 175kcal.





Environmental controls

Environmental control means the removal of accessibility to food and thus temptation. If food is not available, your son or daughter is less likely to seek it. This is sometime referred to as **food security**. Environmental controls can take several forms, depending on level of need, and they will be needed for those people with PWS whose self-control around food is minimal. Here are some suggestions:

Locks on kitchen, fridge and pantry doors

These can be ordinary locks and keys, magnetic locks, padlocks, or more sophisticated electronic locks. Keep tools stored away safely - these may be used to force locks. You could consider giving back control to your son or daughter by providing them with their own locked table-top fridge (with their own code), stocked with small amounts of, for example, diet drinks or fruit). Even more control can be given by suggesting that your son or daughter buys the fridge and lock with their own money.

Do not leave food lying around

Do not put sauce bottles, bread, etc on the table before the meal commences. Clear everything from the table immediately after a meal and put leftovers straight into the rubbish bin or into the fridge for later (both of which may need to be locked). Though expensive, it may be worth buying a waste disposal unit to deter those who forage through rubbish bins. Make sure that family members and carers do not leave sweets, crisps etc in coat pockets or unlocked drawers.



If everyone in the family agrees, there should be some cupboards or rooms in the house which belong to each family member, and no one else should be allowed to go into them without their permission. This has to apply to your son or daughter with PWS as well, who should have his own private space.

Agree that food shops and restaurants are out of bounds if unsupervised

This is a difficult contract to make unless your son or daughter is never allowed out on their own. For those who do go out independently, the risk is always there. A sudden increase in weight may indicate unscheduled visits to food shops, but it may also mean that they have been obtaining food by some other means, such as stealing from shops, foraging in bins, or asking strangers for food or money. You can minimise some of the risk by alerting local shopkeepers and neighbours, but you may also need to agree with your son or daughter that going out independently is a privilege which they can have as long as they keep to the contract, and which will be withdrawn if they do not. Regular weighing is the best way of monitoring whether they are getting outside access to food. However, it is important not to make your son or daughter feel as if they are failing if the contract is not kept - emphasise what they *can* do whilst suggesting that next time someone goes with them.

Take complete control of access to food

If your son or daughter is very uncooperative about food, will not agree to any form of contract, and is severely overweight, you may have to consider taking complete control yourself. This is the sensible option in the face of overwhelming difficulties, but you should be aware that, as they are an adult, you do not have an automatic legal right to control their food intake. In most cases, common sense will prevail, but you may have to consider action under the Mental Capacity Act (see page 2), especially if your son or daughter is living away from home. You will need to involve social services if you do this.

Preparing, cooking and serving meals

There are many ways in which you can help your son or daughter reduce calories by adapting your methods of preparing, cooking and serving meals. These should not impinge to any great effect on the family's daily lifestyle. Here are some of the most commonly used:



- For snacks, provide fresh fruit or vegetables instead of biscuits or cakes. Buy smaller sizes of fruit such as apples and oranges. If a biscuit is a must, make it a plain biscuit and include the calories in the day's overall intake.
- Grill food, or use a microwave oven, wherever possible. Do not fry food.
- Use small thin slices of bread instead of thicker ones. Use small crackers and crispbreads and use a small amount of low-fat spread.



- Look for alternatives to desserts made with flour, sugar and fats. Try unusual fruits, low calorie jellies, sugar-free yoghurts or fromage frais.
- Use a smaller plate .
- Give larger portions of "free" vegetables and salads, but less of higher calorie foods. Make sure there are sufficient protein foods.
- You can avoid family arguments about one person having more than another by serving separate meals on trays in front of the television.
- Add water and/or extra vegetables to your son or daughter's helping of soups or stews to make it look larger. Avoid thick soups, cream soups and thickened stews.
- To prolong a meal, serve salad as a special first course, or even before dessert as in France.
- Cook correct amounts of food for each meal and ensure there are no extras or leftovers for second helpings.
- Encourage your son or daughter to have plenty of fluids eg low calorie squashes and water, unless this is restricted due to medical reasons.
- If your family is used to having second helpings, ensure your son or daughter has less in the first helping so that they can have a second helping along with everyone else.

Dietary management for adults with PWS

Eating out and parties

Eating out is one of the pleasures of life for many people. There is no reason why your son or daughter should be left out, but some advance planning and agreement may be required. These are a few of the options you could consider:

- **Some pubs and restaurants now offer smaller portion sizes. It is worth looking out for those.**
- **Agree that only certain foods, or only a small portion of any celebratory cake or dessert, can be eaten.**
- **Agree that if there are no "diet" drinks on the menu, the option will be mineral water.**
- **Agree with your son or daughter that they have only two of the courses on offer. Ask them to identify the healthier options on the menu, and praise them when they get it right.**



Social and educational controls

There are some kinds of eating behaviour which your son or daughter can learn to see as socially acceptable, or which will gain them favour with other people. The social environment can also be altered to be more supportive.

Educationally, most people with the syndrome benefit from knowing about the effect PWS has on them, and the need to limit their food intake for the sake of their health. People can therefore be educated about what is a reasonable portion of food to have on their plate, not to eat whilst they are talking, and to wait until others are served before commencing the meal. Reward or praise your son or daughter when they do this correctly.

Others can be supportive by not eating in front of your son or daughter, joining in with the "healthy eating" programme, and remembering not to leave leftovers lying around.

Many people can also learn to recognise foods which are high in calories and those which are lower in calories, and your son or daughter may enjoy shopping with you for the healthier options. Some people will be able to read food labels for themselves. It may be advisable to ask your son or daughter to help you make the shopping list before you set out - this will minimise arguments in the shop. Always praise the good choices, and suggest alternatives when an inappropriate choice is made.

Make sure that YOUR facts are correct before passing on your knowledge - it is very difficult for a person with the syndrome to "unlearn" something which they have been wrongly taught.

Taking more self control

A few people with PWS appear to benefit from having greater personal control over what they eat, especially in terms of improved behaviour. However, this is not a step to be taken lightly, as it can quickly get out of hand, and in any case, the person is unlikely to remain slim if allowed totally unsupervised access to food. Some other factors you may wish to consider before taking such a step are:



- Have an agreed trial period and take gradual steps eg begin by simply preparing vegetables.
- You have to be able to set a good example. If anyone in the rest of the family has difficulty controlling their appetite or the amount of food they eat, your son or daughter will find it very difficult to realise they have to control their eating habits when others do not.
- Your son or daughter's ability to understand or recognise high and low calorie foods is very important. Those with severe learning difficulties are unlikely to be able to prepare whole meals independently, but they may be able to carry out some of the other steps.
- Consider what you are going to do if it fails. One suggestion is to agree a contract with the person that if their weight goes beyond an agreed level, then you go back a step or two to something they are more able to manage. Write down the contract or draw pictures to represent it, and pin it in a prominent place in the kitchen or bathroom. The contract must involve weighing at least weekly as there may be an insidious decline in personal control which otherwise goes unnoticed.

Drinks and alcohol

There are now many diet drinks on the market, which are generally considered to be safe as long as they are not consumed in large quantities. Cola, while OK in moderation, has a decaying effect on teeth, which are often very vulnerable to decay in PWS. Therefore, if possible, a variety of other drinks should be offered.



Use skimmed milk for your son or daughter's tea and coffee, and sweetener rather than sugar. Some people may enjoy herb, fruit or lemon teas. If possible, use different types of sweetener in different drinks, so as to avoid a very high concentration of any one type of sweetener in the total intake. It is worth trying to cut down on sweeteners over time as too much sweetener could be harmful. A gradual reduction will usually go unnoticed.

Anecdotal evidence from parents suggests that alcohol has very similar effects on people with PWS as it does on the rest of the population, although a person with PWS may become intoxicated more quickly than most people, and having drunk smaller quantities of alcohol.

Going to the pub can be a very helpful socialising experience for adults with PWS, but if possible someone should go with them who understands their dietary needs. Alcoholic drinks have a high calorie content, but there are now some drinks on the market which are lower in calories.

If a visit to the pub is planned, try to make up for the extra calories by going without a dessert at mealtime. Low calorie lemonade, tonic water, etc can be used as mixers to keep overall alcohol and calorie levels lower.

Helping your son or daughter to lose weight

DATE	POUNDS	KILOGRAMS	DAILY CALORIES	PHYSICAL ACTIVITY

It may be necessary to support your son or daughter to lose several kilograms in weight. This will involve a decrease in daily calorie intake with, if possible, an increase in the level of physical activity.

You may need to get advice about calorie levels and calorie counting from your GP or dietitian. If they have little or no knowledge about PWS, send them appropriate leaflets on PWS in advance of the appointment.

Keep to realistic expectations: 0.5-1 kg (2-4lbs) per month is quite adequate.

Continued ...

Dietary management for adults with PWS

Exercise

Regular exercise is very important for people with PWS and whenever possible it should be incorporated into their daily programme. It is important for several reasons:

- It helps burn up energy (calories)
- It will improve muscle tone
- It improves circulation
- It distracts from eating
- It improves alertness

Exercise can be taken in regular sessions with the aid of computer games such as Wii Fit, exercise DVDs, exercise bikes and step machines (while watching favourite TV programmes or listening to music); by going to classes, or even by hiring a personal trainer using your son or daughter's Disability Living Allowance or PIP - this is especially good for those who thrive on one-to-one attention.

It can also be taken in less formal ways: housework, gardening, cycling, walking, riding, dancing, delivering newspapers, etc.



Helping your son or daughter to lose weight (continued)

Some people find a sponsored slim a very attractive way to lose weight, whilst others may get some benefit from going to a slimming club, or similar (check with the organisers first before going ahead).

It obviously helps if someone else in the family or a friend is also attempting to lose weight and can sympathise. Some may find working towards fitting into a fashionable outfit is a sufficient target, whilst others may need the extra support of colourful charts, and tokens towards rewards such as DVDs or outings.

Another idea is to start a "video diary" using a smart-phone or camcorder to take a few minutes of video each week, so that the person can see their weight loss from week to week, and talk about their difficulties and successes in losing weight. A successful video diary would be a very valuable resource for the PWSA UK and extremely useful for encouraging others. Alternatively, take a series of photographs, perhaps with the tape measure showing their measurements.

Whatever method of support is chosen, regular weighing is essential to ensure that it is working. If it is not, and unwanted weight is being gained, then the method needs to be re-examined to see what is going wrong, and perhaps additional support and/or different methods will be required.

Everyone who is involved in the daily life of your son or daughter should be made aware of the exact level of support which is being offered and adhere to it. Many people with the syndrome will quickly take advantage of loopholes in the system, or of newcomers, to obtain extra food. This includes family, friends, neighbours, local shops, staff at schools and colleges, day centres, respite centres, etc.

Praise your son or daughter whenever they meet or even exceed expectations.

Weight management is usually more likely to succeed if your son or daughter is involved in the discussions regarding the level of support which is being offered.

Other weight loss diets and treatments

"Miracle" diets

Generally speaking, commercial "miracle diets" may work in the short term, but their effects do not last. Some diets are dangerously short of essential nutrients - consult a registered dietitian or your GP before embarking on any of the latest fad diets.

Appetite suppressants

To date conventional appetite suppressants have had mixed results on people with PWS, often having unwanted side effects on the nervous system, and in a few cases triggering episodes of mental illness. They do not always succeed in reducing appetite and they are rarely effective in the long term. Effects in normal people only last about 6 weeks.

Surgery

Gastric by-pass operations can be performed, but usually as a last resort. There is a risk for very obese people under anaesthetic, and the side-effects of gastric by-pass, such as continual diarrhoea and unwanted folds of skin, can be very unpleasant. Wiring of the jaws is generally considered inadvisable.



Continued ...

Other weight loss diets and treatments (continued)

Hospitalisation

Hospitalisation may be the only alternative in cases of severe obesity.



A person may need to be admitted to hospital to reduce weight under supervision (see page 2 about the Mental Capacity Act). This will only work if the person does not gain access to food (eg from other patients) and nursing staff are aware of the person's cleverness in obtaining food from unusual sources.

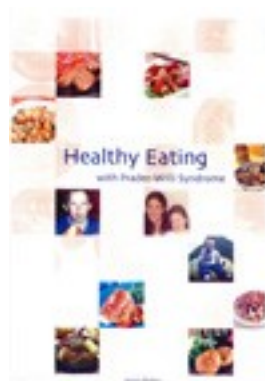
You may find it helpful to give hospitals a copy of **Information for Hospital and Emergency Department staff** <https://www.pwsa.co.uk/home/emergency-medical-information>

Cosmetic surgery to reduce skin folds after weight loss

Although this is a possibility, it is important to think through the implications of this process. Consideration should be given as to whether the person with PWS is likely to regain weight - this could have a very adverse effect on the scar tissue resulting from cosmetic surgery. Equally, would skin-picking at scars be problematic, possibly causing severe infections? Generally speaking, this is not a procedure that would be recommended for people with PWS and only carried out in exceptional circumstances.

Further information

Some of this article is taken from **Healthy Eating with Prader-Willi Syndrome** <https://www.pwsa.co.uk/about-pws/publications/> The book has sections on calorie counting, portion sizes, nutrition, meal planning and some sample menus and recipes.



Red Yellow Green system for Weight Management

– developed in Canada especially for people with PWS. You can read a presentation on this at

www.bcpwsa.com/aboutpws/nutritional_care.pdf

The book can be ordered from the Ontario PWS Association at www.opwsa.com

PWSA USA – also has various leaflets and books on dietary management. See www.pwsausa.org for more details.

Dietary management for adults with PWS

Helping your son or daughter understand about the need for a healthy weight

You may find it helpful to show your son or daughter the booklet **“Our Way of Life”**

There is also a workbook available, aimed mainly at children and younger people, which may be helpful for those with limited understanding. It is called **“Let’s Talk about PWS”**

<https://www.pwsa.co.uk/about-pws/publications/>

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