

# Prader-Willi Syndrome – Eating

INFOSHEET for Family members and Care providers of Adults with PWS

## Summary



Living with Prader-Willi Syndrome (PWS) means that controlling food intake is difficult. This compulsion to eat is life-threatening. Morbid obesity results from overeating and can lead to a range of health complications and even death. Compulsive eating and obsession with food usually begins early in life. Sometimes raw, frozen, discarded or spoiled food and other inedible or toxic substances may be eaten necessitating supervision in general.



This compulsion cannot be regulated by medication. The person cannot help it, the compulsion to eat is hard-wired. People with PWS experience higher rates of mental illness like depression, obsessive compulsive disorder, and psychosis. Signs of these mental illnesses should be taken seriously.



Because people with PWS have low muscle tone and inactivity, they need an especially balanced low-calorie diet with vitamin and mineral supplementation and exercise.



Regular weighing and diet review are recommended.



The best meal/food plan is one that can be applied routinely and consistently. Food restriction may require locking the kitchen and food storage areas.



If weight is controlled, life expectancy and quality may be greatly improved and the person can enjoy good health. People with PWS can live long, healthy, happy lives!



## *A Meaningful Life*

We all want to live meaningful and active lives. So do people with PWS! Consider these questions:

- Is s/he occupied in meaningful activities/work during the week?
- Is s/he living with people they want to live with?
- Is s/he contributing to their local community?

Having a meaningful and active life will mean the person is happier and that will reduce 'behaviours' associated with frustration and loneliness. It also means having less time to think of food!



## *Routine*

Routine is necessary for people with PWS. They want to know what is going to happen next, who is going to be on duty, what's on the menu for dinner, who is coming to visit, and so on.

Keeping to the routine is not always possible, so explaining changes in advance is vital.



## *Dietary Management*

It is important that food management starts early.

It is important that you recognise that food-seeking will be a problem which may result in morbid obesity, and that you take steps to control this before it becomes a life-threatening situation.

Because of the chemical dysfunction, there is no on/off mechanism that tells the brain, "I've eaten enough". What happens instead is that the brain keeps telling the stomach, "you're starving, you need food", and the drive to find food overrides everything else.

The vast majority of people with PWS show excessive eating behaviours; some of these include stealing food, stealing money in order to buy food, taking food from others, breaking locks on cupboards, and so on.

Unfortunately it is also very easy for people with PWS to gain weight. Managing weight gain becomes critical.

Management means locks on pantries, fridges, food cupboards - not straight away, but when food-seeking becomes apparent.



## Counting the Calories

Calories are what the body uses as its source of fuel and energy. Energy from food is calculated in calories to give a quantity we can measure. For example, a boiled egg has 80 calories.

Most foods are labelled with the energy level per 100g; sometimes it is in calories and sometimes in kilojoules or kJs. A rough guide to convert calories to kilojoules is to multiply by 4.

1 Calorie  $\approx$  4 kJ

The amount of calories a person needs depends on their age and physique. For an overweight adult (not on growth hormone therapy), a diet of 1,000 calories per day is recommended. That breaks down into around 300 calories per meal (3 a day) and 100 calories left over for drinks and snacks.



## Exercise and Calories

It is much harder for a person to exercise if they are overweight, so try to involve the person in planning so that they can choose things to do that they enjoy.

Even slow exercise requires energy which means the burning of calories. Burning calories is the key to managing weight. Some examples are:

- Sitting for one hour uses 100 calories.
- Doing housework for one hour uses 180 calories.
- Gardening for one hour uses 220 calories.
- Brisk walking for one hour uses 330 calories.
- Jogging for one hour uses up to 750 calories.

Try to encourage exercise at all times, even hidden exercise, such as hanging out the clothes, window shopping, parking further from the shops or house, and climbing the stairs.

There are all sorts of exercises designed to strengthen muscles. Start an organised and fun exercise programme as soon as possible.



### *Further Reading*

Healthy Eating with Prader-Willi Syndrome. By J Waters, 2007. PWSA UK

Prader-Willi Syndrome: practical ideas to help with weight control. By M Gellatly. 2001. PWSA UK

Cookbook: Low fat, low sugar recipes for the Prader-Willi Syndrome diet. By D Unterberger. 2003 PWSA USA

Red-Yellow-Green Weight Control System booklet. By K Balko. 2006. PWSA Ontario USA <http://www.pwsausa.org/publications/index.htm>

DVD: Food, Behavior and Beyond - Practical Management for the Child or Adult with PWS. By LM Gourash and JL Forster. 2005

These books and DVD are generally sold by the PWS Associations in the respective countries or could be borrowed from the PWS Association in your state (Vic, NSW and QLD)

Australia <http://www.pws.org.au/>

New Zealand <http://www.pws.org.nz/publications/>

UK <http://www.pwsa.co.uk/index.php/what-is-pws/186-publications>

USA <http://www.pwsausa.org/publications/index.htm>

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