# Practical Portions for Prader-Willi Syndrome







## **Practical Portions for Prader-Willi Syndrome**

Prader-Willi Syndrome (PWS) is a rare genetic condition causing an overwhelming and uncontrollable drive to eat. If obesity develops, this can impair quality of life and can be life-limiting.

People with PWS need about 30% fewer calories than their peers. This means that managing portion sizes is crucial for quality of life.



For more information, visit Prader-Willi Syndrome Association UK: **www.pwsa.co.uk** 

This portion size resource provides practical guidance on how to regulate food intake, helping people with PWS to lead healthier lives.

Portion size management is important for individuals with PWS for the following reasons:

- Hyperphagia: People with PWS experience an overwhelming drive to eat (hyperphagia). This intense hunger can lead to excessive food consumption, which can lead to rapid weight gain. Effective portion control can help mitigate this challenge by limiting the amount of food consumed.
- 2. Hypotonia: PWS is characterised by low muscle tone (hypotonia). This affects physical activity and metabolism. Appropriate portion sizes can help ensure that people with PWS receive adequate nutrition without causing excessive weight gain that could overburden their muscles.
- 3. Health risks: Hyperphagia makes it very difficult for people with PWS to make their own choices about food. Without proper dietary management, people with PWS can become obese, leading to serious health issues, for example diabetes, cardiovascular problems, and joint strain. Portion control can help maintain a healthier weight and reduces the risk of associated complications.
- 4. Quality of Life: Managing portion sizes can allow people with PWS to enjoy a better quality of life. It helps them maintain a healthy weight, take part in physical activities, and avoid obesity-related limitations.

## About this booklet

#### Who is it for?

The information in this booklet aims to support anyone with Prader-Willi Syndrome (PWS) or who cares for a child, young person or adult with PWS.

#### What will it help you to do?

It will help you and your family to understand and choose a balanced diet for good health now and in the future. It can also help you to explain and show appropriate portion sizes to others.

#### Why is a healthy diet important?

A healthy balanced diet provides the variety and quantity of food and drink to support growth and maintain health throughout life. Getting into the habit of choosing healthy foods and appropriate portion sizes now helps limit the risk of health problems later, for example heart disease and type 2 diabetes. The illustration on pages 4-5 shows the food groups that form a healthy balanced diet for people with PWS.

#### What is an appropriate portion size?

The photos and information on the following pages provide guidance based on estimates of what people with PWS need at different ages. We recommend that you work with a dietitian to select the right portion sizes.

The exact number of calories a person with PWS needs is hard to define and can be very individual. However, it is estimated that people with PWS need about 30% fewer calories than people without PWS.

The right size of portion (and therefore the right number of calories) will depend on many factors including age, weight gain pattern and activity levels. Adjustments will be needed as these factors change with time. For example, in adolescence, during the growth spurt, boys may need to have more calories (approximately 250 calories more a day).

Regular reviews of weight are essential to guide intakes and optimise health in people with PWS. Height and weight should be measured at hospital appointments. Some people and families also find it helpful to assess weight regularly at home.

#### Helpful hints for measuring appropriate portion sizes

In this booklet, each photo shows a portion with the name, description, weight and handy measure. These photos are examples of foods from each food group. To have a balanced diet, people with PWS will need to eat a wider variety than this. If there is something they like to eat which is not shown, we recommend that you discuss this with a dietitian or health professional.

## A healthy diet with PWS





Water, lower fat milk, sugar-free drinks including tea and coffee all count.



#### **Cutlery and crockery**

The cutlery and crockery shown here are the size of those used in the photographs. You can use similar household utensils to help measure your own food portions.

This circle is the same size as the plate used in the photos (22cm/8½ inches diameter). Serving food on a small plate like this, instead of a large plate, can make portions look bigger.

The bowl used in the photos is 17cm (61/2 inches) diameter.

All tablespoons are heaped unless otherwise stated.





Practical Portions for Prader-Willi Syndrome 7

## How much food should a person with PWS eat every day?

The table below shows the recommended number of portions from each food group a person with PWS needs every day. Depending on individual needs, your healthcare professional may suggest something different.

Food Group	Number of portions each day
Vegetables	At least 3
Fruit	2
Potatoes, bread, rice, pasta and other starchy carbohydrates	6-8
Beans, pulses, fish, eggs, meat and other proteins. Include at least 1 portion of oily fish such as salmon, herring or mackerel every week.	2-3
Dairy and alternatives	3
Oils and spreads	3
Foods and drinks high in fat, salt or sugar	Not recommended

The photos on the following pages provide examples of foods, and typical portion sizes for different ages. Use these examples as a practical guide to help you provide balanced and appropriately sized meals and snacks. Always try to offer a variety of food from each group to provide all the nutrients needed for growth and health.

People with PWS need fewer calories but have the same vitamin and mineral requirements as people without PWS. **Therefore, taking an over the counter, age-appropriate vitamin and mineral supplement (including iron, zinc, vitamin A and D) is recommended.** Vitamin D deficiency is common in the UK. Speak to your GP about additional vitamin D supplementation.

This booklet includes commonly eaten foods. Some of these foods are best avoided as they are unhealthy. However, if they are included, only have them very occasionally. They have been marked with a red  $\bigcirc$ .

## **Cooking methods**

Try to choose healthier cooking methods. For example, steaming, baking, grilling, boiling and microwaving are all better than frying.



\*\*This [resource] would help me so I can provide myself with a balanced diet without putting myself at risk.

**Adult with PWS** 









We have included wholegrain/brown versions of pasta, bread and rice as these are recommended.







16 NDR-UK



Fruit portions for 2-3 years old are approximately 30g. For all other age groups, fruit portions are approximately 55g. Vegetable portions for 2-3 years old are 40g. For all other age groups, vegetable portions are 80g as recommended in the Eatwell Guide.

vitamins and minerals. Compared to fruit, they generally provide fewer calories.









## Dairy and alternatives (3 portions per day)



## Dairy and alternatives (3 portions per day)



#### Dairy and alternatives (3 portions per day)

19-64 years

14-18 years

11-13 years

7-10 years

4-6 years

2-3 years

Age range



\*If choosing dairy alternatives, always go for alternatives that are fortified with calcium. Rice milk is not suitable for under 5's

Cheddar

cheese (

like either

piece.

cheese, Cream

## Oils and spreads (3 portions per day)

19-64 years	1½ teaspoon (7g)	1 teaspoon (3g)	1½ teaspoons (12g)	3 teaspoons (30g)
14-18 years	1½ teaspoon (7g)	1 teaspoon (3g)	2 teaspoons (16g)	3½ teaspoons (35g)
11-13 years	1½ teaspoon (7g)	1 teaspoon (3g)	1½ teaspoons (12g)	3 teaspoons (30g)
7-10 years	1 teaspoon (5g)	1 teaspoon (3g)	1 teaspoon (8g)	2½ teaspoons (25g)
4-6 years	1 teaspoon (5g)	1 teaspoon (3g)	1 teaspoon (8g)	1½ teaspoons (15g)
2-3 years	1 teaspoon (5g)	1 teaspoon (3g)	1 teaspoon (8g)	1½ teaspoons (15g)
Age range	Reduced-fat spread 🔴	<b>Oil O</b> (olive, rapeseed, sunflower, vegetable)	Mayonnaise, reduced-fat	Tomato ketchup 🔴

## **Snacks**

Depending on how much a person with PWS eats at mealtimes, small portions of healthy snacks can help them meet their daily energy and nutritional needs.

Some people with PWS find it helpful to have a healthy snack routine in place. Others may find it easier to have no snacks.

Fruit such as strawberries, raspberries and melon are good snack options. They are lower in calories than bananas and grapes. Dried fruit is not recommended as it is high in sugar.

Try to avoid sweets and snacks that are high in fat and/or sugar as they have no nutritional value and provide lots of calories.

## Fluid

- Children aged 2-3 years should aim to drink 6-7 glasses of fluid a day. From 4 years onwards, children and adults should aim to drink 6-10 glasses of fluid a day. A glass is about 200ml.
- Tap water is the best way to quench thirst. Many parents/carers find it difficult to get a person with PWS to drink water. Try adding a slice of lemon, lime or cucumber for flavour, offer ice cubes, or choose a sugar-free drink.
- Skimmed milk is a good option, unless a dietitian has advised differently. It contains calories so check the portion guidance on page 22.
- Energy, caffeinated and alcoholic drinks are not recommended. They often contain high amounts of calories and provide no nutritional benefit.
- A sugar-free drink can be an alternative to a snack.

## **Plated meals**

Meals often include a combination of foods from different food groups. The following example shows how you can make up a balanced meal to ensure a person with PWS gets all the nutrients they need to grow, develop and maintain health.



Below is an example meal plan for a 12-year-old girl with PWS. It is an example of how you can include the recommended number and size of portions from each food group in a day.

		Monday	
Meal/ snack	Food	Food group	Number of portions
Breakfast	1½ wheat biscuits	Potatoes, bread, rice, pasta and other starchy carbohydrates	1
	210ml skimmed milk	Dairy and alternatives	1
	1½ plums	Fruit	1
Snack	3 breadsticks	Potatoes, bread, rice, pasta and other starchy carbohydrates	1
	2 tbsp. hummus	Beans, pulses, fish, eggs, meat and other proteins	1
	2 slices wholegrain bread	Potatoes, bread, rice, pasta and other starchy carbohydrates	2
	1½ tsp. reduced-fat spread	Oils and spreads	1
Lunch	3 slices roast beef	Beans, pulses, fish, eggs, meat and other proteins	1
	5cm cucumber and 6 cherry tomatoes	Vegetables	1
	²/ <sub>3</sub> banana	Fruit	1
Snack	<sup>2</sup> / <sub>3</sub> small matchbox sized piece of cheese	Dairy and alternatives	1
	<sup>3</sup> / <sub>4</sub> wrap	Potatoes, bread, rice, pasta and other starchy carbohydrates	1
Dinner	½ salmon fillet, oven cooked	Beans, pulses, fish, eggs, meat and other proteins	1
	2 egg-sized potatoes	Potatoes, bread, rice, pasta and other starchy carbohydrates	1
	3 heaped tbsp. carrots	Vegetables	1
	3 heaped tbsp. broccoli	Vegetables	1
	1 tsp. olive oil	Oils and spreads	1
	1 fromage frais (85g)	Dairy and alternatives	1

## Food security and food seeking tips

Food security refers to creating a safe food environment for people with PWS. This should ensure that they do not feel stressed: they are not tempted to take food they don't need, but feel reassured that food will be available soon i.e. with the next meal. Removing the physical temptation to take food also removes the thought of food. This can be reassuring and help people to focus on and enjoy other things.

Meals should follow a regular schedule to ensure predictable mealtimes with a consistent order of when they happen. It can be helpful if these are not only defined by time e.g. "we will have lunch once we have finished this walk".



<sup>66</sup>This will be...a great resource to share with the rest of the family as they never understand quantities that are suitable for our daughter.<sup>39</sup>

Parent of child with PWS.

Many parents/carers can provide food security by providing food routines and limiting access to food to help manage hyperphagia (increased hunger).

These include:

- Locks on kitchen doors and/or cupboards. Locks are not just a physical security but psychological as well. Removing opportunities to access food can take away anxiety around this.
- Routines in place for meal and snack times. Knowing when they are going to eat can be reassuring and help to reduce stress for those with PWS. A visual timetable/plan can help too.
- Meal/menu planning in advance.
- Dish up meals at the worktop rather than on the table.
- Remove any leftover food from sight.

Food seeking can be difficult to manage. However, reducing opportunities to find food can help, not just at home but in school too. PWSA UK can provide advice and guidance to schools.

Here are some tips to help:

- Ensure your child's teacher has a good understanding of the condition and is aware of the role and responsibility they have.
- Keep lunch boxes and snacks out of sight.
- Do not use food such as rice, pasta or flour dough in play.
- Ensure supervision during lunch breaks and social times.
- Encourage other children not to swap or share food.
- Do not use food as a reward.
- Keep pet foods such as seeds and cereals out of sight.
- Ask other parents not to bring sweets and cakes to celebrate birthdays.
- Consider not having unhealthy foods at home.

## A message for people with PWS

This resource aims to help those who support a person with PWS understand what an appropriate amount of food is to be healthy.

If you are reading this for yourself and there is something that you do not understand, ask for help from your family or healthcare professional.



## **Useful websites**

Prader-Willi Syndrome Association UK (PWSA UK): **www.pwsa.co.uk** 

International Prader-Willi Syndrome Organisation (IPWSO): **www.ipwso.org** 

Notes

Developed in collaboration with NDR-UK and PWSA UK. Authors:

Chris Smith, RD, Senior Paediatric Dietitian and Honorary Senior
Clinical Lecturer, University Hospitals Sussex NHS Foundation Trust
Dr Evelien Gevers, MD PhD FRCPCH, Consultant and Honorary Reader
in Paediatric Endocrinology and Diabetes, Barts Health NHS Trust – Royal
London Hospital, and Centre for Endocrinology – Queen Mary University
of London
Jennifer Sanderson, Paediatric Endocrine Clinical Nurse Specialist, NHS
Greater Glasgow and Clyde
Lisa Singh, RD, Specialist Paediatric Dietitian, NHS Greater Glasgow and Clyde
Jackie Lodge, Chief Executive, PWSA UK
Peri Wallach, Therapy Assistant, NHS Greater Glasgow and Clyde
Ruth Consterdine, Deputy Manager, PWSA UK
Jenni Henderson, Development Officer, NDR-UK

Ν	0	t	e	S
	_	_	_	_

This information has been produced for Nutrition and Diet Resources UK (NDR-UK) by Registered Dietitians and other relevant health professionals. It is protected under copyright and may not be modified or reproduced without prior written permission from NDR-UK. At the time of publication the information contained within the leaflet was, to the best of our knowledge, correct and up-to-date. Always consult a suitably qualified dietitian and/or your GP on health problems. NDR-UK cannot be held responsible for how clients/patients interpret and use the information within this resource nor is it responsible for the content of external sites or links.





© NDR-UK Ref 9540. First published: 2024 To re-order visit **www.ndr-uk.org**  Part funded by the Scottish Government.

NDR-UK - the UK's practitioner-led nutrition and diet resource provider. Registered charity number SC041043, company number SC364677.