



# Nutrition Guidelines: Toddlers through Adolescence with Prader-Willi Syndrome

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### **Dedication:**

This booklet is dedicated to all the individuals with Prader-Willi syndrome (PWS) and their families. The authors' hope is this information provides parents and caregivers with basic nutrition knowledge they need to create the healthiest environment possible for their children with PWS. For more information on PWS, and/or state and local contacts, please visit [www.pwsausa.org](http://www.pwsausa.org) or contact the Prader-Willi Syndrome Association | USA directly at (941) 312-0400.

## Introduction

Prader-Willi syndrome (PWS) is a genetic disorder affecting about 1 in 12,000-15,000 individuals (both sexes, all races). It has many characteristics, the most challenging being the constant obsession with food, also referred to as hyperphagia. Most individuals with PWS feel an insatiable drive to obtain food and eat most of the time. While there is not yet any prescribed medical treatment for the hyperphagia symptom, there have been successes with weight control through calorie-controlled diet, exercise, behavior management, and structured living situations. It is important to note that there is generally a spectrum among individuals with these characteristics from mild to severe.

In 2012, Miller, et.al described PWS in multiple stages:

- Phase 0: Decreased fetal movements in utero and lower birth rate
- Phase 1a: Hypotonia with difficulty feeding and may have failure to thrive (0-6 months; median age at completion 9 months)
- Phase 1b: No difficulty feeding and growing appropriately on growth curve (5-15 months; median age of onset 9 months)
- Phase 2a: Weight increasing without an increase in food or calories (20-31 months; median age of onset 2 years)
- Phase 2b: Weight increasing with an increase in food seeking (3-5 years; median age of onset 4 ½ years)
- Phase 3: Hyperphagic, food-seeking, lack of satiety (5 years-adulthood; median age of onset 8 years).
- Phase 4: Appetite for some adults is no longer insatiable and is able to feel full (adulthood)

## Obesity Prevention

The prevention of obesity is one of the most important goals for the family of a child with Prader-Willi syndrome. Your child with PWS is at increased risk for obesity because the child will become hyperphagic (excessive hunger and rarely feeling full) coupled with the fact that he or she only needs about two-thirds of the calories needed by most people.

Preventing your child from becoming overweight or obese is necessary to reduce the risk of breathing problems, heart disease, hypertension, fatty liver disease and diabetes that can develop early in childhood. This can be done with a nutrient dense, calorie-controlled diet and consistent environmental controls that will help establish a healthy eating pattern for life. Obesity prevention requires a balance. The challenge is to help your child feel satisfied without giving him or her too many calories, yet provide all of the vitamins and minerals needed to grow well.

Some parents can become so fearful about their child becoming obese that they may overly restrict calories and certain food groups and then the child fails to grow. In certain situations, if weight and length have plateaued for a period of time, the degree of food restriction may need to be loosened. Parents or caregivers should monitor growth closely and speak to their physician and qualified registered dietitian for guidance on this issue.

## Weight & Height Measurements

Frequent tracking of height and weight measurements are essential for monitoring your child's growth and planning an appropriate diet. Keep your own records at home. Your doctor will likely plot your child's growth measurements on a growth chart. There are growth charts specific to individuals with Prader-Willi syndrome, and the Centers for Disease Control (CDC) has growth charts for all children. The CDC charts are able to calculate a child's body mass index (BMI) to determine the degree of overweight or obesity beginning at age two years and older.

It is ideal to always weigh your child on the same scale, at the same time of the day, and in the same type of clothing (e.g. in pajamas, first thing in the morning). Measuring height measurements (marked on a wall taken with the child standing up straight) should be taken every three months. If your child is gaining weight, but is not growing taller, he or she may be getting too many calories. Check closely what he or she is eating at meals. If meals, snack and physical activity has not changed, check to see where the extra food may be coming from (e.g., refrigerator, cabinets, family members, friends, school). Ask your doctor or registered dietitian for an appropriate goal for your child's weight.

## Calorie Needs

Children need calories to maintain a normal rate of growth and calorie needs change as the child grows and moves through the phases of PWS. Your child's needs may vary and in early life, a child with PWS may need a high calorie diet to grow. Once a calorie level is estimated, a child's growth should be monitored regularly to be sure that the level is accomplishing the right goal (i.e., weight maintenance or weight loss). This may require frequent weight and length/height check visits with the pediatrician to make sure your child is plotted on the growth charts according to the trends. Children on growth hormone will have significant improvement in linear growth, decrease of fat mass, and increase of lean mass. Your medical team will use your child's current intake and current weight/height or BMI to determine if extra calories are needed.

To maintain weight, children need about 20-28 calories per inch of height OR 10-11 calories per centimeter every day. A slightly overweight child's weight goal may be to maintain weight so that he or she can grow into that weight. To lose weight, children need about 18 calories per inch of height each day OR 8-9 calories per centimeter. An appropriate and safe goal for weight loss would be about two pounds per month.

## Exercise

A good exercise plan is essential and important to start as early as possible. The entire family should participate, as children might need extra encouragement. Not only does this benefit the child with PWS, but it also keeps the entire family healthier. Daily walks, bike riding, swimming, gymnastics, martial arts like karate and Tae Kwon Do are examples of good physical activities. There are bound to be some hikes in your area of the country. Visit [www.localhikes.com](http://www.localhikes.com) to find some.

There are also video games that involve strenuous activity like X Box games and Wii Fit.

Exercise that is regularly scheduled helps burn extra calories and prevent obesity. Strenuous physical activity and/or competitive sports should be undertaken only with the approval of your child's doctor.

## Nutrition Through The Years

**Birth and Infancy:** Many children with Prader-Willi syndrome have early feeding difficulties. At birth they appear hypotonic (floppy) and have a poor suck/swallow reflex making sucking and swallowing breast milk or formula difficult. Many children may need a nasal-gastric feeding tube or have a gastric tube placed while they gain strength to suck and swallow on their own. Breast milk is preferred, but if breast milk is not available, there are infant formulas that provide the optimal calories, protein and fat children need to grow. It's important to work closely with your physicians, dietitian and therapists to help the child gain the strength to eat by mouth and have the feeding tube, if placed, removed as soon as possible. In addition, these medical professionals can also help you with the introduction of solid foods (table foods) to your child.

**Toddlers/Preschool Age:** After those early months of life, children usually begin to develop better feeding skills and are able to eat and grow appropriately without a feeding tube. Establishing good nutrition and feeding schedules is essential. Parents must pay attention to what they feed and how they feed their children. During this time, children may start Phase 2a, which means that weight will increase without an increase in calories. At this time, paying closer attention to calories and an intensive review by a registered dietitian may become crucial. If calories are reduced, a dietitian needs to make certain all nutrients are still met.

**School Age Children & Teenagers:** As children grow, they naturally want to be more social. This shift in their social situation will most certainly allow for more access to food. It's essential to make certain all school administrators, teachers, friends, family and coaches are aware of the diagnosis and that the child cannot have unlimited access to food. Also, physical activity is even more crucial to balance calorie intake. Scheduling at least an hour of activity 6-7 days per week is helpful to control weight.

**Adults:** As teenagers transition into adulthood, new challenges occur in regards to weight management and food security. Many adults participate in day programming, which promotes socialization and a level of independence but also poses an increased threat to food security. Strict monitoring of caloric intake should continue through adulthood with caloric requirements stabilizing and/or decreasing with age. Physical activity remains crucial to support ongoing weight maintenance and/or loss as well as bone health.

## What to Feed Your Child

Having a child with Prader-Willi syndrome requires parents to develop a good working knowledge of nutrition. Variety is important for optimal nutrition. The nutritional goal is to offer very nutrient dense foods that are satisfying. Examples are vegetables, fruits, lean meat, chicken, turkey, fish, pork, dairy, eggs, nuts, beans, and seeds. Steer clear from foods in packages with too much sugar, unhealthy fats or salt. Reading labels is essential and ingredients lists that are full of unrecognizable ingredients are not ideal in a daily diet.

Amount Per Serving	
<b>Calories</b> 100	Calories from Fat 30
%Daily Value*	
<b>Total Fat</b> 3.5g	<b>5%</b>
Saturated Fat 0g	<b>1%</b>
Trans Fat 0g	
<b>Cholesterol</b> 0mg	<b>0%</b>
<b>Sodium</b> 290mg	<b>12%</b>
<b>Total Carbohydrate</b> 18g	<b>6%</b>
Dietary Fiber 6g	<b>25%</b>
Sugars 2g	
<b>Protein</b> 4g	

### Nutrition 101...

- **Calories** are made up of carbohydrates, proteins and fats. There are 4 calories per gram of carbohydrates, 4 calories per gram of protein and 9 calories per gram of fat. Study food labels closely and review these websites for more details and information about general nutrition:

- [www.kidshealth.org](http://www.kidshealth.org)
- [http://kidshealth.org/kid/stay\\_healthy/food/calorie.html](http://kidshealth.org/kid/stay_healthy/food/calorie.html)
- [www.cdc.gov/nutrition](http://www.cdc.gov/nutrition)
- <http://www.cdc.gov/nutrition/everyone/basics/>

- **Carbohydrates:** This group is important for energy in the body, but there are some that are more nutritional than others that you should choose more often. The healthiest carbohydrates are fresh fruits and vegetables, whole-wheat products, brown rice, beans, peas, and lentils. Milk and yogurt are also sources of carbohydrates. Unhealthier carbohydrates are white bread, white pasta, white rice, muffins, donuts, cookies, brownies, and candy. Read food labels closely and it's ideal to look for more than 3 grams of fiber per serving for healthier carbohydrates.

- **Protein:** Protein is important for building, maintaining and replacing the tissues in the body. Muscles, organs and the immune system are made up of proteins and so it is important to have good sources of protein in the diet. Protein comes from meat, chicken, turkey, fish, eggs, dairy, beans, nuts and seeds. Each meal and snack should have a good source of protein to make for a balanced diet.

- **Fats:** Science and medicine have revised the recommendations as they relate to fat as part of a healthy diet. This recommendation also translates to people with Prader-Willi syndrome. For many years fat was considered unhealthy and low fat diets were recommended. New research has broken fats down into different components that require further explanation:

- o **Monounsaturated/Unsaturated fats:** These fats have a beneficial effect on heart health when eaten in moderation and used to replace saturated fat and trans fat in a diet. Omega-3, a type of monounsaturated fat, can lower triglyceride levels and increase HDL cholesterol (the good cholesterol). Several other studies also suggest that these fatty acids may help lower blood pressure.

- Examples: Fatty fish like salmon, mackerel, herring, lake trout, sardines, and albacore tuna are high in monounsaturated fats and the very good omega-3 fatty acids. The American Heart Association recommends eating these fatty fish at least two times a week. Eating a variety of these fish will help minimize any potential effects due to environmental pollutants. Omega-3 fats can also be found in flax seeds, walnuts, sardines, salmon, beef and soybeans. Canola oil, soybean oil, and olive oil are also good sources. While all of these foods are healthy, calories still count, so offer these foods in proper portions.

- o **Saturated Fats:** These fats are found naturally occurring in some foods that provide important nutrients for a healthy diet. So the key here is to offer these foods in moderation and focus more on fats that are unsaturated as listed above.

- Examples: fatty beef, lamb, pork, poultry with skin, lard, eggs, cream, butter, cheese, and other dairy products from whole/reduced fat.

- o **Trans Fatty Acids (Trans Fats):** Trans fat needs to be reduced or eliminated whenever possible. These fats can raise blood cholesterol levels and contribute to heart disease. They provide very few nutrients and are high in calories.

- Examples: These include fried foods like doughnuts, and baked goods including cakes, pie crusts, biscuits, frozen pizza, cookies, crackers, and stick margarines and other spreads.

- For the infant and very young child it is important to not overly restrict fat intake because it is still needed in the diet for adequate brain development and growth. When looking at calories, 30% of calories should come from fat.

## Exchange System

The exchange system is a good plan because it is balanced, flexible, easy to follow and easy to modify as your child grows. Foods are divided into six groups based on their calories and nutrients. They are: starch/bread, vegetables, fruit, meat, dairy products, and fat. A daily food plan can be developed with the help of a registered dietitian that is based on your child's nutrient needs and food preferences. The following table, "Daily food Plan Using the Exchange system," shows the number of food choices allowed from each group for a variety of calorie levels. Serving sizes are described in the "Food Exchange Lists" at the end of this booklet.

	Calories Per Day				
	600	800	1000	1200	1400
Exchange Groups	<b>Number of Servings Per Day</b>				
Starch/Bread	2	3	3	3	3
Vegetables	7	7	7	7	7
Fruit	3	4	4	4	4
Meat	2	2	5	7	8
Dairy	1.5	2	2	2	2
Fat	1	1	1	1	1

\*Fat should be used sparingly.

Standard portion sizes are too large for younger children. Foods in the correct portion size are selected each day according to the meal pattern. The foods that most children like from the exchange group are incorporated in the Food Exchange Lists at the end of this booklet.

When using the exchange system, it is still important to read food labels and compare them with the exchange list for portion size and calories. If the calories are significantly different, choose a different product or adjust the serving size.

## Calorie Counting

Families may prefer to count calories rather than follow the exchange system. When counting calories, careful meal planning is necessary so that your child's diet includes all of the food groups. Careful reading of the nutrition information on the food labels is essential. Similar products may vary in calorie content. For example, one slice of diet bread has about 40 calories compared to regular bread, which has about 80 calories per slice. Be sure to serve the portion size specified in the label. Even if families opt to not count calories, it's important to have a general sense of foods that are low and high in calories when planning meals and snacks. Learning to read food labels is important and reading about nutrition is necessary.

## Vitamin/ Mineral Supplements

Because the calorie level of the diet is low, it is challenging to offer all the vitamins and minerals the children need for growth on less than 1000-1200 calories per day. A complete multivitamin supplement is often recommended. One key when choosing your multivitamin is looking for the front label to include the term "complete."



One mineral that is difficult to get in adequate amounts is calcium. Calcium is important because it helps your child grow strong bones. Your child's calcium needs will change with age. One 8-oz. glass of milk has 300mg of calcium. If your child is allergic to milk, soy milk, unsweetened almond, hemp or other nondairy fortified milk can be a great option. If your child consumes too little or no dairy products, they will need a calcium supplement. For some children, limiting milk to 2 servings per day and adding a supplement at the end of the day can help save some of the calories for other foods. A registered dietitian can review your child's diet to see if they are getting the proper amounts of all vitamins and minerals.

**Hydration**

One of the challenges with children with Prader-Willi syndrome is their hydration status. Put simply, they often do not like to drink water so it's important to devise strategies to increase water intake. Adding the juice of oranges, lemons, or limes can help. You can also soak strawberries or apples and cinnamon overnight in water to make water more palatable to drink. Cucumbers can also flavor water. Juice and soda are not recommended because they provide excess calories that can cause weight gain.

**Notes:**

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## How to Feed Your Child

Structuring meals and snacks is essential. As a child moves from milk to more table food, setting a structure for eating provides support and predictability for your child. Posting menus and rules about mealtime behavior is also a good strategy to set expectations. But as parents, you must do it and follow through. Limit access to unsupervised buffet situations where food is readily available and easy to take.

## Special Events

Treats and other high-calorie foods are a part of life. These foods, in very small amounts, may be planned into your child's diet. When your child has a treat, adjust his or her total calories by subtracting the calories from the daily total.

When incorporating treats or high-calorie foods, they must be planned ahead of time and included on your menus. Food is not to be used as a reward for good behavior. When given spontaneously, treats can be viewed by your child as a reward. Unplanned treats can also cause increased anxiety surrounding food.

Special events and holidays are important for all families. Try to make food less important during holidays. Stress decorating, gift giving, and family togetherness.

Determine what types of food will be served at your event. If limited options are going to be available, serve small meals at other points of the day and allow the individual to have a small portion of a few foods at the event. Depending on the event, packing a meal may be a good alternative.

## Food Access

Along with strict diet control, your child's access to food must be controlled. Food-seeking, gorging and hoarding are all typical behaviors of children with PWS. Many families lock kitchen cupboards, refrigerators and other food storage areas. Families should have a fairly strict schedule for meals and snack times. Most children with PWS appear to like a set routine. This may help to decrease food seeking and hoarding. Serve food directly on the plates. After meals, put the food away immediately and restrict access to the kitchen.

Instruct all family members and all caregivers (e.g., extended family, teachers, neighbors, etc.) on the importance of preventing access to food. One way is not to give in to begging and temper tantrums for food. Undesirable behavior over food needs to be dealt with calmly, firmly and with consistency. Extra food is dangerous to a child with PWS. When a child's weight continues to increase and he or she is following a strict diet at home, the problem may be access to food away from home.

It is important to recognize that the degree of food issues in PWS can be a significant source of stress to the family and home environment. If the family works as a team, is consistent in approach and structure of food issues, and sibling needs are also recognized, the stress can be managed as well. Families are encouraged to seek counseling, if stresses around managing diet and food access become overwhelming.

**Notes:**

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Prevention or control of obesity is one of the most challenging tasks for the parents of a child with PWS. It requires constant work and good communication between all who are involved in the child’s care and it can be done. This is a summary of what needs to be done:

- Monitor growth closely and keep in close contact with physicians
- Balance of carbohydrate, protein and fat
- Read food labels closely
- Structure meals/snack times
- Fresh food is always best
- No juice, soda or other calorie containing drinks (unless milk is worked into diet)
- Intensive, frequent physical activity as a family
- Be an advocate for your child with PWS. Educate all parties involved in taking care of your child so they have an understanding of PWS and can keep the child safe and healthy

## Food Charts

<b>Starch/Bread</b> (80 calories per serving) Aim for whole grains when possible.	<b>Serving Size</b>
<b>Bread, Cereal &amp; Pasta</b> Cooked Cereals Ready-to-eat unsweetened cereal Pasta (enriched or whole grain, cooked) Rice (white or brown), cooked Quinoa, cooked Couscous Granola Bagel English Muffin Hot Dog or Hamburger Bun Light Hot Dog or Hamburger Bun Pita (6" across) Tortilla (6" across) Bread (white or wheat) Light/Diet Bread (white or wheat)	½ cup ¾ cup (1 oz) ½ cup ½ cup 1/3 cup 1/3 cup ¼ cup ½ bagel ½ muffin ½ bun 1 1 1 1 slice 2 slices
<b>Dried Peas, Beans, Lentils</b> Beans (cooked) Lentils (cooked)	½ cup 1/3 cup
<b>Starchy Vegetables</b> Corn or peas Corn on the cob (6" long) Baked Potato (small) Mashed Potatoes Winter Squash (acorn or butternut) Yams or Sweet Potatoes	½ cup 1 1 (3 oz) ½ cup ¾ cup ½ cup
<b>Crackers/Snacks</b> Animal Crackers (plain) Graham Crackers (2 1/2" square) Popcorn (no fat added) Pretzels (sticks) Saltine-type crackers	8 3 3 cups 40 6
<b>Starchy Foods Prepared with Fat</b> Pancake (4" across) Waffle (4 1/2" across) Taco Shell (6" across) French Toast	1 1 1 1

<b>Dairy Products</b> (90 calories per serving) Children under 2 years of age should be given whole milk.	<b>Serving Size</b>
Skim milk Soy milk (unsweetened) Rice milk (unsweetened) Almond milk Hemp milk (unsweetened) Non-fat yogurt (check label for calories)	8 oz 8 oz 8 oz 8 oz 8 oz 4-6 oz

<b>Fat</b> (45 calories, 5 grams of fat per serving)	<b>Serving Size</b>
Margarine/oil/mayonnaise Diet Margarine/diet mayonnaise Salad Dressing Reduced Calorie Salad Dressing Peanuts Seeds, Pine Nuts, Sunflower (without shells) Bacon Cream Cheese	1 teaspoon 2 teaspoons 1 tablespoon 2 tablespoons 20 small or 10 large 1 tablespoon 1 slice 1 tablespoon

<b>Fruit</b> (60 calories per serving)	<b>Serving Size</b>
Fresh, Medium Fruit Berries or Melon Canned Fruit (in juice) Dried Fruit Raisins	1 1 cup ½ cup ¼ cup 2 tablespoons

Meat and Meat Substitutes	Serving Size
<p><b>Lean Meat</b> (55 calories, 3 grams of fat per serving)</p> <p>Cooked poultry, without skin Cooked lean meat Cooked fish Tuna (in water) Non-fat cottage cheese Diet Cheese (less than 55 calories/oz) 95% Fat-Free luncheon meats</p>	<p>1 oz 1 oz 1 oz 1 oz 1 oz ¼ cup 1 oz 1 oz</p>
<p><b>Medium-Fat Meats and Substitutes</b> (75 calories, 5 grams of fat per serving)</p> <p>Most beef products Most pork products Skim or part-skim milk cheese:     Ricotta     Mozzarella     Diet Cheese (56-80 calories/oz) Egg Tofu</p>	<p>1 oz 1 oz ¼ cup 1 oz 1 oz 1 4 oz</p>
<p><b>High-Fat Meat and Substitutes</b> (110 calories, 8 grams of fat per serving)</p> <p>Pork spareribs and sausage All fried fish products All regular cheese Luncheon meat- bologna, salami Turkey or Chicken hotdog Peanut Butter</p>	<p>1 oz 1 oz 1 oz 1 oz 1 1 tablespoon</p>



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