

## Hidden Gut Issues

### Rethinking Food Choices and Meals for the Person with Prader-Willi Syndrome

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Researchers are discovering important problems in the gastrointestinal systems in persons with Prader-Willi syndrome (PWS). We have always known that persons with PWS of all ages can have problems with swallowing, choking, stomach issues and chronic constipation. Until recently, there had only been a few research publications to help us understand why this happens so frequently.

Recent research studies have documented low production of saliva (6), high risk of choking (7), high prevalence of constipation in adults with PWS (1), slow stomach emptying (gastroparesis) and gastric rupture (2, 3, 4). Dr. Roxann Gross and her team at The Children's Institute of Pittsburgh published important information in 2016 - the first research into swallowing, which revealed a high incidence of serious swallowing abnormalities in persons with PWS. None of the 30 subjects were aware of food still in their throats after "swallowing" - which is a risk for choking and pneumonia.

We know that persons with PWS have low muscle tone. **Poor muscle tone can affect the entire gastrointestinal system – mouth, esophagus, stomach and intestines.** With this new knowledge, we need to rethink food choices, timing and food preparation practices for the person with PWS. Any person with PWS who is already experiencing problems with their gastrointestinal system should be promptly evaluated by their healthcare professional and, if needed, referred to a gastroenterologist (physician who specializes in the digestive system). Even if your loved one with PWS is not currently experiencing symptoms such as choking or difficulty swallowing, we recommend caution and a few simple adjustments.

### What can be done to prevent or manage digestive problems for the person with PWS?

- Smaller quantities
- More moisture
- Softer consistency
- Less fiber

**Gastroparesis** is a condition where the movement and digestion of food in the stomach is slowed or stopped. Contractions in the stomach help crush the food and move it on to the small intestine, where further digestion and absorption of nutrients takes place. A person with gastroparesis may have food remain in their stomach for a very long period of time, which can support abnormal bacterial growth and the forming of a mass (called a bezoar) which can cause blockage or obstruction. Changing the diet to include foods that are easier to digest, plus making meals smaller in size are recommended as the first treatment for slow stomach emptying.

Foods high in fat and fiber require more work and time for the stomach to digest, and should be avoided. In most cases, fluids can pass through the stomach at a normal rate – even when gastroparesis is present. **Smaller-sized meals, foods that are semi-liquid or liquid and low fat, put less stress on the stomach and help the stomach to empty faster.** A soft, lower fiber diet may be safer, which is a change from the high fiber recommendation for constipation, which can be treated other ways.

Low production of saliva decreases the amount of moisture and lubrication needed for smooth passage of food from the mouth to the stomach. This issue, along with poor muscle tone, contributes to problems with food lodging in the throat. Staying hydrated between meals, along with good dental health, can help to minimize the problems of low production of saliva. Adding more moisture to food may help to prevent choking.

In the past, a low-calorie, high-fiber diet was the mainstay in management for both the child and adult with PWS. A low-calorie diet should continue, of course, but fiber should be reduced when slow gastric emptying is diagnosed or suspected. Since nutritional recommendations will vary based on age and other health issues of the person with PWS, it is important to receive specific dietary advice from your dietician or from PWSA (USA) nutritional publications. These new recommendations can be adapted for any individual plan such as low carb, lactose free, or vegetarian.

#### **More suggestions:**

- 1. Small frequent meals: Avoid three large meals per day; recommend three small meals and two snacks**
- 2. Liquids during and between meals: Encourage water or fluids between bites of food and after the meal**
- 3. Avoid: RAW vegetables and fruits, nuts and salads**
- 4. DO PROVIDE: Well cooked vegetables (softened) and/or mashed; fruits in softer form – applesauce, canned fruits in natural juice, and cooked cereal, smoothies, soups**

6. Probiotic foods: Daily (yogurt, kefir, sauerkraut) to restore and maintain healthy bacteria in the stomach
7. Ground or cut up meats: Turkey, beef, chicken
8. Eggs: Scrambled, fried or baked as quiches
9. Schedule the last meal as early as possible to allow time for food to be digested before bedtime
10. Stay upright after eating; schedule a walk or other activity to improve digestion
11. Work with your dietician to follow calorie, carbohydrate and other nutrient goals

**References:**

Kuhlman L, Joensson IM, Froekjaer JB, Krogh K, Farholt S: **A descriptive study of colorectal function in adults with Prader-Willi syndrome: high prevalence of constipation.** *Gastroenterology* 2014 **14**:63.

Arenz T, Schwarzer A, Pfluger T, Koletzko S, Schmidt H: **Delayed gastric emptying in adults with Prader-Willi syndrome.** *J Pediatric Endocrinol Metab* 2010, **23**(9):867-871.

Hoybe C, Barkeling B, Nashlund E, Thoren M, Hellstrom PM: **Eating behavior and gastric emptying in patients with Prader-Willi syndrome.** *Ann Nutr Metab* 2007, **51**(3):264-269.

Stevenson DA, Heinemann J, Angulo M, Butler MG, Loker J, Rupe N, Kendell P, Cassidy SB, Scheimann A: **Gastric rupture and necrosis in Prader-Willi syndrome.** *J Pediatr Gastroenterol Nutr* 2007, **45**(2):272-274.

Gross RD, Gisser R, Cherpes G, Hartman K, Maheshwary R. 2016. **Subclinical dysphagia in persons with Prader-Willi syndrome.** *Am J Med Genet Part A* 9999A:1-11.

Saeves, R, Reseland, J, Kvam, R, Sandvil, L, Nordgarden, H: **Saliva in Prader-Willi syndrome: quantitative and qualitative characteristics.** *Archives of Oral Biology* 2012, **57**(10): 1335-1341.

Stevenson, D, Heinemann, J, Angulo, M, Butler, M, Rupe, N, Kendell, P, Clerucuzio, L, Scheimann, A: **Deaths due to choking in Prader-Willi syndrome.** *American Journal of Medical Genetics* 2007 Mar 1: 143(5): 484-487.

Sample Planning for Meals/Snacks – 1000 Calories – in a person who does not have diabetes. These samples are not specific to any certain diet or nutritional choices you may make. (Check calories on individual products you purchase)			
Breakfast Goal: 225	chopped hard-boiled egg (70) 1 slice lite toast (45) ½TBSP margarine (25) 3 oz. DanActive Probiotic drink (70) 1 cup flavored water / coffee	1 cup unsweetened cereal (100) =½cup unsweetened plain almond milk (15) 1 lite cheese stick (60) ½cup lite applesauce (50) 1 cup flavored water / coffee	½cup lite Activia yogurt (60) ½lite English muffin (50) 1 TBSP lite cream cheese (30) 1 cup V8 juice (50) 1 cup flavored water / coffee
AM Snack	1 lite string cheese (60) 3 crackers (60)	1 cup lite Activia Greek yogurt (80)	1 tortilla (80) with 1 TBSP lite cream cheese (30)

Goal: 125	1 cup flavored water	½cup lgte fruit (45) 1 cup flavored water	1 cup flavored water
Lunch Goal: 225	WRAP - 1 tortilla (80), 3 oz turkey lunch meat (50), 1 TBSP lite mayo (20) ½cup cooked green beans (20) ½cup lite Activia yogurt (60) 1 cup flavored water	egg salad sandwich using lite mayo & lite bread (70+30+90) 1 cup cooked broccoli (30) 1 dill pickle (0) 1 cup flavored water	2 slices ham lunch meat (30) w/ 1 slice lite cheddar cheese (50) on 1 bagel thin (100) w/ 1 tsp mustard (0) 1 ½cup cooked cauliflower (50) ½cup SF Jell-O (0) 1 cup flavored water
PM Snack Goal: 125	½sandwich – 1 slice lite bread (45), 2 slices ham (30) ½cup lite fruit cup (50) 1 cup flavored water	½cup lite Activia yogurt (60) ½cup lite fruit cup (50) 1 cup flavored water	1 lite cheese stick (60) - wrap w/ham lunch meat (15) ½cup lite fruit cup (50) 1 cup ice tea
Supper Goal: 300	1 cup chili soup - ground turkey, crushed tomatoes, chopped celery, chili seasoning (200) 1 cup unsweet plain almond milk (30) 1 cup cooked cauliflower (30) ½cup lite pears (50) (warm & season with cinnamon)	tacos – 1 tortilla (80) topped with 2 ounces ground beef (80) w/ taco seasoning (10) top with ¼ cup salsa (25), ¼ cup lite shredded cheese (70), small amount shredded lettuce 1 cup unsweet plain almond milk (30) ½cup SF Jell-O (0)	grilled tuna melt sandwich – 2 slices light bread (90) w/ ¼ cup tuna (50) mixed with 1 TBSP lite mayo (30) 1 cup vegetable soup (90) 1 cup unsweet plain almond milk (30)

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