

Rethinking Our Approach to Diet and Nutrition for the Person with Prader-Willi Syndrome

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Over the past several years, we have learned many new things in the area of gastrointestinal health for persons with Prader-Willi syndrome (PWS). For years, there have been many anecdotal reports of persons with PWS of all ages having problems with swallowing, choking, stomach issues and chronic constipation. Up until recently, there had only been a few research studies on these issues, specific to persons with PWS. Studies have now documented low production of saliva (6), a high risk of choking (7), a high prevalence of constipation in adults with PWS (1) and documentation of slow stomach/gastric emptying and gastric rupture (2, 3, 4). Dr. Roxann Gross, PhD. (5) reported at the PWSA (USA) 2015 conference that many persons with PWS in her study were found to have significant swallowing issues. Food was visualized on x-ray, lodged in throats, but persons with PWS were unaware of it. We know that persons with PWS have low muscle tone. **We are learning that this poor muscle tone affects their entire gastrointestinal system – mouth, esophagus, stomach and intestines.** All of these findings point to problems with feeding, nutrition and digestive health. With this new knowledge, we now need to rethink the approach to diet and nutrition for the person with PWS. It is important that any person with PWS who is experiencing problems with their gastrointestinal system, should be evaluated by a healthcare professional and, if needed, referred to a gastroenterologist (physician who specializes in diseases and problems with the gastrointestinal system). There can be many reasons for these types of problems.

So, what can be done to prevent and/or manage these problems using appropriate dietary measures for the person with PWS? Low production of saliva can impact the amount of moisture and lubrication needed for safe passage of food from the mouth to the stomach. This issue, along with poor muscle tone, may contribute to problems with food lodging in the throat and putting the individual at increased risk of choking. Another challenge arises in diet management for slow gastric emptying (gastroparesis) and constipation which contradict each other. According to the American College of Gastroenterology, a high fiber diet along with adequate fluids is recommended for prevention and treatment of constipation. In contrast, they recommend a low-fiber, low-fat diet for prevention and treatment of gastroparesis. So what diet approach should the person with PWS use when they may be at risk for both of these problems?

In the past, a low-calorie, high-fiber diet was the mainstay in management for both the child and adult with PWS. The “Red, Yellow, Green diet” was an approach that was utilized by many. However, the time has come to rethink what, when,

and how much we are feeding the child and adult with PWS. The low calorie diet must continue; however, fiber should be reduced. Since nutritional recommendations (calcium, protein, etc. needs) will vary based on **age** and **other health issues** of the person with PWS, it is important to receive specific dietary advice for a child/adult from a dietician or from PWSA (USA) nutritional publications. The examples shared in this article are **not specific** to any certain age group.

Adequate fluid intake along with good dental health can help to minimize problems with a low production of saliva. Gastroparesis is a condition where the movement and digestion of food in the stomach is slowed and in some cases, absent. Normally, contractions in the stomach help crush ingested food and move it in to the small intestine, where further digestion and absorption of nutrients takes place. It is common for a person with gastroparesis to have food remain in their stomach for a very long period of time. Retained food in the stomach can cause abnormal bacterial growth and form a mass (called a bezoar) which can cause blockage or obstruction. Diet is one of the primary treatment approaches for this problem. Foods high in fat and fiber require more work and more 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.** Persons with severe gastroparesis must have all food liquefied or pureed, with some requiring all nutrition be administered in a feeding tube. This is challenging for anyone; but is especially difficult for a person with PWS. A soft, lower fiber diet may be more appropriate.

Suggested Recommendations for a Soft, Lower Fiber diet for the person with PWS:

Maintain a low calorie diet.

1. Provide small, frequent meals. Avoid the three LARGE meals per day. Break up meals/snacks to three small meals and two snacks a day (six is only recommended if the person has diabetes). Reduce quantity of food being provided at one time.

2. Include more liquid or semi-liquid food items. Provide liquids during and between meals. Have person drink water or fluids between bites of food. (Helps moisten food and facilitate movement from mouth to the stomach; less work and time with food in stomach).

3. AVOID: RAW vegetables and fruits, nuts and salads. (YES, this is a change)

4. DO PROVIDE: vegetables that have been cooked (softened) and/or mashed, fruits in softer form – applesauce, fruits in natural juices, and cooked cereal.

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5. Soup - at least once a week (this has built-in softening and liquids)

6. Probiotic foods at least 1x daily (yogurt, kefir, sauerkraut). Helps restore and maintain healthy bacteria in the stomach.

7. Ground or cut up meat. (Ground turkey, chicken and/or beef)

8. Eggs – scrambled, fried or baked quiches

9. Schedule the last meal as early in the evening as possible.

(More time for food to be digested before going to bed)

10. Keep upright after eating. Schedule a walk or other movement. (Use gravity to help move food along)

11. HOW TO BEGIN AND PLAN: 1. Consult with dietician and determine specific calorie and nutritional needs for person with PWS. 2. Divide calories between three smaller meals and two snack times. (Suggest 6th meal time only if person with PWS has diabetes.) **THE FOLLOWING EXAMPLES HAVE BEEN PROVIDED. ■**

| Sample Planning for Meals/Snacks – 1000 Calories – in a person who does not have diabetes (Check calories on individual products you purchase) | | | |
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| Breakfast | <ul style="list-style-type: none"> o chopped hard-boiled egg (70) o 1 slice lite toast (45) o ½ TBSP margarine (25) | <ul style="list-style-type: none"> o 1 cup unsweetened cereal (100) o ½ cup unsweetened plain almond milk (15) o 1 lite cheese stick (60) o ½ cup lite applesauce (50) o 1 cup flavored water / coffee | <ul style="list-style-type: none"> o ½ cup lite Activia yogurt (60) o ½ lite English muffin (50) o 1 TBSP lite cream cheese (30) o 1 cup V8 juice (50) o 1 cup flavored water / coffee |
| Goal: 225 | <ul style="list-style-type: none"> o 3 oz. DanActive Probiotic drink (70) o 1 cup flavored water / coffee | | |
| AM Snack | <ul style="list-style-type: none"> • 1 lite string cheese (60) • 3 crackers (60) • 1 cup flavored water | <ul style="list-style-type: none"> • 1 cup lite Activia Greek yogurt (80) • ½ cup lite fruit (45) • 1 cup flavored water | <ul style="list-style-type: none"> • 1 tortilla (80) with 1 TBSP lite cream cheese (30) • 1 cup flavored water |
| Goal: 125 | | | |
| Lunch | <ul style="list-style-type: none"> o WRAP - 1 tortilla (80), 3 oz turkey lunch meat (50), 1 TBSP lite mayo (20) o ½ cup cooked green beans (20) o ½ cup lite Activia yogurt (60) o 1 cup flavored water | <ul style="list-style-type: none"> o egg salad sandwich using light mayo & lite bread (70+30+90) o 1 cup cooked broccoli (30) o 1 dill pickle (0) o 1 cup flavored water | <ul style="list-style-type: none"> o 2 slices ham lunch meat (30) w/ 1 slice lite cheddar cheese (50) on 1 bagel thin (100) w/ 1 tsp mustard (0) o 1 ½ cup cooked cauliflower (50) o ½ cup SF Jell-O (0) o 1 cup flavored water |
| Goal: 225 | | | |
| PM Snack | <ul style="list-style-type: none"> • ½ sandwich – 1 slice lite bread (45), 2 slices ham (30) • ½ cup lite fruit cup (50) • 1 cup flavored water | <ul style="list-style-type: none"> • ½ cup lite Activia yogurt (60) • ½ cup lite fruit cup (50) • 1 cup flavored water | <ul style="list-style-type: none"> • 1 lite cheese stick (60) - wrap w/ham lunch meat (15) • ½ cup lite fruit cup (50) • 1 cup ice tea |
| Goal: 125 | | | |
| Supper | <ul style="list-style-type: none"> o 1 cup chili soup - ground turkey, crushed tomatoes, chopped celery, chili seasoning (200) o 1 cup unsweet plain almond milk (30) o 1 cup cooked cauliflower (30) o ½ cup lite pears (50) (warm & season with cinnamon) | <ul style="list-style-type: none"> o 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 o 1 cup unsweet plain almond milk (30) o ½ cup SF Jell-O (0) | <ul style="list-style-type: none"> o grilled tuna melt sandwich – 2 slices light bread (90) w/ ¼ cup tuna (50) mixed with 1 TBSP lite mayo (30) o 1 cup vegetable soup (90) o 1 cup unsweet plain almond milk (30) |
| Goal: 300 | | | |

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