

Nutritional Strategies in the Management of Prader-Willi Syndrome, Gastroparesis and Constipation

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The following is a chart summarizing some of the nutritional strategies that can be utilized in planning and managing the diet and nutrition for a person with Prader-Willi Syndrome. It also provides some guidance for two common gastrointestinal problems often seen in these children and adults with PWS. It is not meant to replace the individual recommendations from your nutritionist or physician. There are many additional non-nutritional strategies that can help with preventing obesity and other health problems that are not included in this handout. This is an excerpt from a presentation that these two professionals performed at the PWSA (USA) conference – 2013.

Condition	Definition	Causes and Associated Findings	Nutritional Management Suggestions	Other
PRADER-WILLI SYNDROME	Two phases – both contribute to special nutritional challenges	Dysfunction in Hypothalamus: <ul style="list-style-type: none"> • Satiation problem • No structural abnormality in the brain has been found 		
“Failure to Thrive”	Characterized by poor feeding and poor weight gain	<ul style="list-style-type: none"> • Hormone deficiencies – Growth Hormone • Excessive sleepiness • Small jaw and teeth 	<ul style="list-style-type: none"> • Require special feeding techniques – feeding tubes, special nipples • Close monitoring of weight • Timed/ scheduled feedings • Referral to nutritionist 	<ul style="list-style-type: none"> • Important <u>NOT</u> to limit fats during this time. They are essential in brain development.
“Thriving too Well”	<ul style="list-style-type: none"> • Increased food pre-occupation and food seeking behavior – varying degrees of intensity • Weight gain and food security – lifelong struggle and challenge 	<ul style="list-style-type: none"> • Research suggests problem in peripheral satiety signaling. • Defective nutrient cycling • Body composition abnormality • Hormone deficiencies - GH 	<ul style="list-style-type: none"> • Low calorie – high nutrition diet • Recommend “Red, Yellow, Green Diet” • Avoid processed foods – corn dogs, chicken nuggets • Use fresh or frozen fruits and vegetables • Avoid “one pot meals” – number of foods served often important to person with PWS • Dining Out <ul style="list-style-type: none"> ○ Special – not routine ○ Research ahead ○ Nutritional information often found on website ○ Use care with coffee drinks 	KEY MANAGEMENT STRATEGIES: Food Security, Exercise & Diet/Nutrition

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GASTROPARESIS	<p>Slow movement of food through the stomach to the intestine.</p> <p>IMPORTANT to be under the care of a gastroenterologist (GI specialist)</p>	<ul style="list-style-type: none"> • Seeing greater number of persons with PWS being diagnosed with this condition. • Exact cause unknown. • Common complication of diabetes and some stomach surgeries. • Symptoms: <ul style="list-style-type: none"> ○ Reflux/regurgitation ○ Rumination ○ Nausea/vomiting ○ Pain in stomach area ○ Abdominal bloating/distension 	<ul style="list-style-type: none"> • <u>Small</u> frequent meals – 4-6 meals /day • Avoid foods high in fat • Chewing food well before swallowing. • Drinking fluids throughout meal • Low fiber diet • <u>High Fiber Foods to Avoid:</u> <ul style="list-style-type: none"> ○ Fruits – apples, berries, coconuts, figs, oranges & persimmons ○ Vegetables – Brussels’ sprouts, green beans, green peas, lettuce, potato peals, sauerkraut ○ Bran/whole grain cereals ○ Nuts /seeds /legumes/dried beans • <u>In severe cases</u> – semi-solid or liquid foods may be indicated. 	<p>Other recommendations:</p> <ul style="list-style-type: none"> • Sit upright for at least 1 hour after eating • Medications often tried – metoclopramide, erythromycin • Schedule last meal of day to early evening - allows time for food to pass through stomach. • Electrical stimulation – “stomach pacemaker”
CONSTIPATION	<ul style="list-style-type: none"> • Having a bowel movement fewer than 3 times per week. • Stools typically hard, dry, small and difficult to eliminate • Colon absorbs too much water and/or colon’s muscle contractions are slow or sluggish. <p>A person with PWS should be having at least 1-2 bowel movements daily.</p>	<p>Common Causes:</p> <ul style="list-style-type: none"> • Not enough fiber or water/fluids in diet • Lack of physical activity • Medications • Irritable bowel syndrome • Changes in life style – aging, travel • Over-use of laxatives • Ignoring urge to go • Specific conditions or diseases 	<p>Fiber:</p> <ul style="list-style-type: none"> • 14 grams of fiber per 1000 calories is recommended • Part of plant body cannot digest • Can cause constipation if not enough water/fluids in diet • Good sources: <ul style="list-style-type: none"> ○ Whole grains ○ Fruits/vegetables/legumes ○ Fiber One bars • Probiotics – bacteria that help maintain natural balance of micro-flora in intestines <ul style="list-style-type: none"> ○ Yogurt - Activa ○ Cottage cheese 	<p>Non-nutritional suggestions:</p> <ul style="list-style-type: none"> • Monitor frequency and consistency of BM’s • Exercise • Set aside time • Don’t ignore urge • Check with physician regarding use of laxatives
Gastroparesis AND Constipation	<p><u>Comments:</u></p> <ul style="list-style-type: none"> • Combination of problems often seen in persons with PWS. • Contradictory management often recommended (High vs. Low fiber) 		<p><u>Suggestions:</u></p> <ul style="list-style-type: none"> • Drink WARM fluids – especially in the AM before 1st meal – add lemon juice. • Use cooked vegetables and fruits – much less fiber than raw but still contains some fiber. • More liquids throughout the day - Smoothies as snack • Exercise 	

