

A Comprehensive Overview of GI Issues in Prader-Willi Syndrome

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The prevalence of GI issues in persons with PWS of all ages is quite high. In addition to early feeding difficulties, reflux and aspiration symptoms are frequent problems in infancy. Problems with digestion affect approximately 35% of adults; constipation and diarrhea problems occur in 20-35%. Gastroesophageal reflux disease, GERD, is common.

Dr. Scheimann outlined suggestions to treat GERD in infants: use thickened feedings, avoid overfeeding, use more prone positioning, and eliminate all exposure to tobacco smoke. For older children and adults, she recommended: avoid lying down after eating a meal, elevate the head of the bed, lose weight, avoid all tobacco, and avoid foods and medications that may cause reflux. Fundoplication is a surgical option when lifestyle changes and medications aren't enough.

Oral problems are common, including small mouths causing teeth crowding and enamel erosion. Salivary flow is generally far less than normal [dry mouth products such as Biotene can be helpful]. Factors predisposing someone to choking, a serious and not uncommon occurrence in persons with PWS, include hyperphagia (high drive for food), thick saliva, weak pharyngeal muscles, and reflux.

Use of the "pace and chase technique for liquid consumption with meals is helpful in preventing the symptoms of esophageal dysphagia and choking. Dr. Scheimann advises all care providers to learn the Heimlich maneuver, treat reflux and gastritis symptoms, encourage chewing during meals, and of course, always supervise persons with PWS.

Risk factors for developing gallstones, also not uncommon, include obesity, low fiber/high fat diet, and diabetes mellitus. 70-80% of normal adults in one study had no biliary symptoms when their gallstones were detected - the majority of healthy adults did not require treatment for gallstones unless symptoms arose such as right sided abdominal pain or pain after meals.

Constipation and encopresis (involuntary fecal soiling) are common problems. Factors that add to constipating conditions include developmental factors (cognition, genetics,

fluid intake, etc.) and alter anatomy (low muscle tone, malrotation, etc.). Rectal ulcers can occur when there is chronic constipation.

General guidelines to treat constipation in infancy include the careful use of glycerin suppositories or softening agents such as Karo syrup and the increase of fiber intake when solids are introduced.

She cautioned against using enemas, suppositories and finger dilations unless recommended by the physician. For the school age child and adult, her suggestions included a cooked fiber-rich diet plus water and the continuous use of medications, such as Miralax with appropriate amounts of fluid daily rather than intermittent dosing.

Gastric motility (the rate at which the stomach empties) and an impaired vomit reflex (controlled by the central nervous system) contribute to serious stomach expansion and stomach rupture problems that can cause death. Warning signs and immediate hospitalization or ER evaluation for potential gastric rupture or gastric necrosis included a binge eating episode followed by abdominal discomfort, recent history of gastritis or ulcer.

While a variety of bariatric surgery techniques have been attempted in persons with PWS, the long-term results have been very poor. Research continues to explore viable treatment options, but bariatric surgery is not currently one of them.

Updated and reviewed by Dr. Scheimann on July 30, 2019 s.medical.giissuesscheinmann.2019

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