Prader-Willi
SYNDROME ASSOCIATION | USA
SAVING AND TRANSFORMING LIVES

Gastroparesis, Gastric Dilatation, with Gastric Necrosis and PWS

What Do We Know?

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PWSA | USA receives calls from around the country about people with PWS who are experiencing gastrointestinal problems. More persons with PWS are being diagnosed with a problem called GASTROPARESIS (a slow emptying of the stomach). This condition occurs when there is a delay or slowing in the contraction of the stomach muscle. Because of this delay, stomach contents build up and distention can occur. The stomach is a muscle that contracts very much like our heart muscle. Unlike the heart, instead of pushing blood, the stomach pushes food out of its cavity into our intestine for further digestion. It is not surprising to learn that persons with PWS who already have low muscle tone may have poor muscle tone in the internal muscles of their body.

Feeling full is our body's mechanism for regulating the amount of food that the stomach can accommodate. When a person eats, the stomach stretches. It may become "over stretched" or distended when one overeats. We know that persons with PWS do not have the normal mechanism of registering fullness as they eat. They are at a very high risk of over distending their stomachs.

It is believed that if persons with PWS over distend their stomach, it can stretch to the point that it cuts off its blood supply causing necrosis (death of the stomach tissue). This can then result in rupturing of the stomach with highly acidic gastric juices leaking into the chest and abdominal cavity. This is a life threatening situation. It must be quickly diagnosed and treated. Over the past few years, we have seen more persons binge eat and develop this condition – especially during the holiday season. The medical terms used to describe this is GASTRIC DILATATION WITH GASTRIC NECROSIS and STOMACH PERFORATION. Unfortunately, most of these cases have resulted in death and been diagnosed in a postmortem examination. Persons with PWS may be at higher risk for gastroparesis. Many parents and caregivers report that some children and/or adults with PWS experience rumination (the regurgitation of undigested food from the stomach back up to the mouth). It was believed to be a behavioral issue. Is this or could this problem caused by gastroparesis? We do not know. We may need to change its management approach.

Some of the risk factors seen in both conditions are summarized below.

Risk factors for Gastroparesis	**Risk Factors Seen in Persons with PWS**
• Diabetes – most common cause	• Diabetes – many persons with PWS have diabetes.
• Adrenal and thyroid gland problems	• Many persons with PWS have been found to have low functioning of their thyroid gland.
• Certain drugs weaken the stomach – many antide-pressants and heart medications	• Many persons with PWS take antidepressant medications as part of behavior management and some may be taking heart medica-tions.
• Neurologic or brain disorders such as Parkinson's, stroke and brain injury	• We continue to learn the effects of PWS on brain functioning.

It has been found that people with PWS who have suffered from gastric dilatation with gastric necrosis, have had a recent binge episode. It is also more common in those whose weight is under control. The usual symptoms seen in gastroparesis include abdominal distention or bloating, abdominal pain, heartburn, vomiting and regurgitation of stomach fluid into the mouth. These symptoms can be very difficult to detect in persons with PWS. If vomiting occurs or there are any signs of acute abdominal illness, a person with PWS should be evaluated by a health care professional immediately.

If a person with PWS is experiencing gastrointestinal symptoms, he/she may be referred to a gastroenterologist (a doctor who specializes in disorders of the stomach and intestine). Optimally, gastroparesis is diagnosed through a gastric or stomach emptying test. Food that has been "marked" is given to the patient. A scanner then tracks the time it takes for food to leave the stomach. Another test that may be performed is an electrogastrogram (EGG). This is a test similar to the EKG of the heart. The EGG measures electrical waves that normally sweep over the stomach. In gastroparesis, these electrical waves are slower than normal.

If caught early, gastroparesis can be treated. Any underlying medical condition should be treated. Diet and nutrition is adjusted. Since fats delay stomach emptying, foods high in fats are avoided. Because high fiber food stays in the stomach for a long time; they may be restricted. Liquids leave the stomach faster so they normally are not limited. Eating frequent small feedings 4-6 times a day may be helpful. In many cases, medications may be used to stimulate the stomach to contract and empty more normally. It is important to follow the recommendations of your health care professional and/or dietician.