Gastroparesis: The Newest Threat

by Lisa Graziano, M.A., PWCF Executive Director
Janalee Heinemann, M.S., PWSA (USA) Director of Medical Affairs
Ann Scheimann, M.D., MBA, Gastroenterologist and PWS Specialist

If you're a member of the Prader-Willi California Foundation and the national Prader-Willi Syndrome Association (USA) then you have read about or discussed at a meeting of some sort this thing called gastroparesis. There have been articles about the fact that it exists, alerts about it (*Medical Alert: Gastrointestinal Issues in Individuals with PWS*), and now a peek at the likely very high incidence rate. What we haven't yet received enough information about is how do we know if our child/adult may has it and what can we *do* about it. This is the focus of this article.

Please know that this article will likely be a difficult read for those of us who care for and love someone with PWS. Knowledge is power, however, and so if the information contained within this article helps inform care providers enough that they can keep someone with PWS safer, then the read is worth it.

In 1999 PWSA (USA)'s *The Gathered View* included an article about PWS specialist Rob Wharton's, M.D. discovery of what he termed Acute Idiopathic Gastric Dilation. What Dr. Wharton saw in his patient was that for some unknown (idiopathic) reason the stomach (gastric) was quickly (acute) pushed out (distended) and caused the stomach tissues to die. If not immediately treated with surgery this condition may lead to death.

Over the following years, particularly with closer examination by PWS and GI specialist Ann Scheimann, M.D., it has become clearer that a great number of other people with PWS have a stomach that empties too slowly. In fact, Dr. Scheimann now believes ***it is highly probable that a significant number of people with PWS have some degree of a slow emptying stomach***. The medical name of this disorder is gastroparesis: the muscles in the wall of the stomach work poorly and prevent the stomach from emptying properly. As a result, food stays in the stomach longer than it should. Over time, the volume of accumulated food in the stomach can cause the stomach to become full. Like a balloon that has too much air, the stomach of someone with PWS that contains too much food can respond in one of two ways: it will rupture or the food will push so hard against the stomach lining that it compresses and weakens the cells in the stomach. Both of these conditions cause massive internal infection and can quickly lead to death.

Other important factors to consider are that some medications such as narcotic pain relievers and anticholinergic medications can also cause the stomach to empty too slowly (as well as cause dry mouth symptoms). Abnormally high blood glucose (sugar) levels or undetected hypothyroidism can also slow stomach emptying therefore it is important to control blood glucose levels and screen periodically for hypothyroidism.

At the same time the stomach empties too slowly, the bowel intestinal tract also seems to empty too slowly. This means that digested food that the body turns into waste product and must eliminate from the body as feces/stool is not entirely eliminated leaving too much stool in the intestinal tract.

I've spoken with many parents and care providers who've believed that because their child or adult has a bowel movement every day that this means they don't have a slow emptying bowel. This is not necessarily true. Even with a regular daily bowel movement the intestinal tract may not empty appropriately. As the colon becomes more backed up with retained stool, the ability to evacuate stool is less effective. Over a long period of time, continuous, constant hard pushing has resulted in some people with PWS to experience rectal prolapse. The feeling of constant fullness and pressure on the anus or itching of the skin from irritation from bile acids present in the stool may contribute to reasons why some people with PWS insert their fingers into their anus or pick at it.

As the colon becomes more impacted with retained stool, emptying of the stomach commonly slows down. This means that gastric rupture or dilation are at dangerously elevated risks.

How to Detect Gastroparesis and Slow Emptying Bowel

How do we know if the individual with PWS we're caring for has gastroparesis or a slow emptying bowel? What are the signs? What are the symptoms? What do we look for? The answers are, unfortunately, that there probably aren't many easily recognizable signs or symptoms.

The symptoms of a slow emptying stomach are primarily nausea, vomiting, abdominal fullness after eating, and/or pain. But for persons with PWS who often have a blunted pain threshold and an absent vomit reflex, symptoms of gastroparesis or Acute Idiopathic Gastric Dilation can be extremely difficult to detect.

Because the abdominal core muscles are generally weaker in persons with PWS, the stomach can often appear to be more rounded. If food is not emptied quickly enough the stomach can look rounded (distended) and feel "too firm" to the touch. On the other hand, for those who are taking growth hormone medication and are therefore more lean, the stomach can already feel "firm" to the touch.

The most likely answer to how we treat the potential for gastropareisis and slow emptying bowel is to presume they exist and treat them as if they exist.

The definitive test to identify delayed stomach emptying is the Gastric Emptying Study which measures the amount of time it takes for food to empty from the stomach and enter the small intestine. The test is done in the nuclear medicine section of a hospital. The patient fasts overnight and eats a breakfast that contains a tiny amount of radioactive material. The patient then lies flat and still on an exam table under a large "arm" that measures the amount of food particles that evaporate from the stomach over a period of time, generally four hours is the appropriate amount of time for the emptying study, following a mixed meal of liquid and solids. There are no side effects from a gastric emptying study; the radioactive material is not absorbed into the body and is eliminated in the stool. The test can be difficult for kids under the age of 10 to complete because it is critical that the person lie perfectly still throughout the duration of the test. It is important to make certain that other factors such as constipation and/or thyroid disease are well controlled prior to completion of the test.

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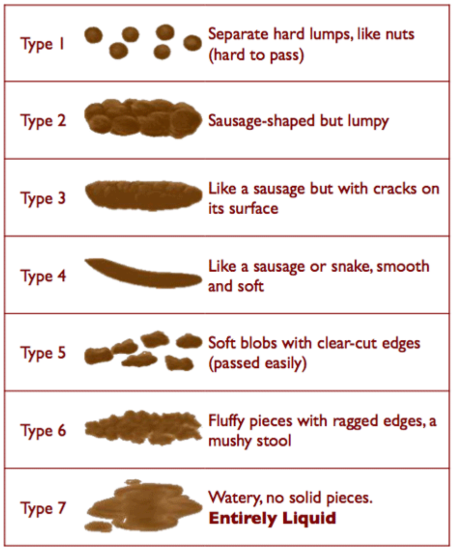
Treatment Strategies

1) As with all treatment of PWS symptoms, the first approach is to always provide Food Security: a) provide a healthy, low-calorie, low carbohydrate diet; b) serve meals and snacks at structured times/sequences of time throughout the day; and c) restrict all access to food.
2) Request from the PWCF or the PWSA (USA) information about GI issues in persons with PWS.
3) If there are GI concerns present, consider consultation with either a pediatric or adult gastroenterologist, dependent upon age. Provide the physician with your GI issues documents.
4) Discuss the pros and cons of a Gastric Emptying Study.
5) Discuss the use of medications such as metoclopramide (Reglan) and erythromycin to improve stomach emptying.
6) Discuss an assessment for stool buildup (e.g., palpation, x-ray). The Bristol Stool Chart can be used to screen/track progress of the management of constipation. Normal stools should be Bristol Class 4 (See Table below).
7) Discuss the use of over-the-counter medications such as Miralax to improve stool elimination and over-the-counter probiotics to help regulate the balance of helpful organisms (microflora) in the intestines
8) If there are challenging issues for your primary GI specialist physician, suggest the GI specialist contact a PWS GI specialist by contacting the PWCF or the PWSA (USA).

Bristol Stool Chart



We continue to learn more about the gastrointestinal and bowel emptying issues of Prader-Willi syndrome and as we do, we will inform you. Maintain your membership in the PWCF and the PWSA (USA) so that you stay as informed as possible about the management of gastroparesis, slow emptying bowel, and all other PWS-related medical issues.

For more information about Prader-Willi syndrome contact:

Prader-Willi California Foundation	310.372.5053 | 800.400.9994 in CA | www.pwcf.org
Prader-Willi Syndrome Association (USA)	941.312.0400 | 800.926-4797 | www.pwsausa.org