Hypertrophic Pyloric Stenosis (HPS)

Patient and family information, brought to you by the Education Committee of APSA

Overview - "What is it?"

Hypertrophic pyloric stenosis (HPS) is a narrowing of the pylorus muscle. The pylorus is a sphincter muscle along the end of the stomach that controls the passage of food into the small intestine. (See Figure 1)

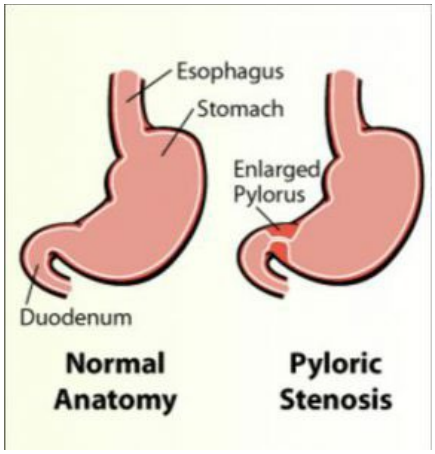

Figure 1: *Difference between normal pylorus and hypertrophic pyloric stenosis.*
(Image credit: surgery.ucsf.edu)

- When the sphincter becomes hypertrophied (thicker than normal), the opening of the pylorus becomes too narrow for food and/or liquids to pass through.
- There is literature to suggest that babies are not born with thickened pylorus muscle, but this condition develops over the first couple weeks of a baby's life.
- Hypertrophic pyloric stenosis is a condition occurring most commonly in first-born, Caucasian male infants, and occurs in approximately 1 in 300 to 900 live births.
- There is no proven cause of HPS. It occurs more frequently in males compared to females with a ratio of 2:1 to 5:1. It is believed to happen more often in the same family; however, most infants have no family history of HPS.

Other conditions that mimic pyloric stenosis: gastroesophageal reflux (GERD), gastroenteritis, increased intracranial pressure, food allergy and duodenal or pyloric atresia.

Signs and Symptoms - "What symptoms will my child have?"

HPS is usually diagnosed between two to eight weeks of life.

Nonbilious vomiting (yellow/clear without green) is the most common symptom associated with HPS.

Projectile vomiting (forceful vomiting shooting across the room) often occurs. The vomited fluid consists of formula/breastmilk and lacks green color. The vomiting may start with occurrence after some feeds and non-projectile initially, but typically progresses to occur after every feed. This may lead to delay of the diagnosis. Your infant may continue to act hungry after vomiting due to food not leaving the stomach. If vomiting continues over days, the baby will become dehydrated, resulting in fewer wet diapers and may not have tears when crying.

Blood levels of electrolytes (minerals such as sodium, bicarbonate and potassium) can be abnormal because of the adjustments and mechanisms the body undergoes to help with the dehydration.

Diagnosis - "What tests are done to find out what my child has?"

Physical Exam: Physicians will feel for the pylorus on abdominal exam, and a thickened pylorus feels like an "olive". This is typically difficult to appreciate while patients are awake unless baby is very dehydrated. Patients need to be relaxed, calm, and cooperative.

Labs: Blood work will be gathered for evaluation. Babies with HPS typically have abnormal labs due to vomiting.

Ultrasound: The first radiologic study performed and is standard for diagnosis. Ultrasound uses sound waves without radiation to create images of the pylorus. In HPS, the pyloric channel is longer and the pyloric wall thicker than normal. After measuring the pylorus, radiologists usually give babies Pedialyte and watch the liquid go through the pylorus. If the liquid does not go through the pylorus from the stomach into the duodenum, this is HPS.

- Ultrasound is operator dependent. Hospitals or centers inexperienced with care of children may not obtain adequate or accurate ultrasound images for diagnosis.

Upper gastrointestinal (UGI) contrast study: If the ultrasound is not diagnostic, or if the baby has already had surgery for pyloric stenosis and has return of vomiting symptoms, this study will be performed. The baby drinks a liquid and a special X-ray machine is used to watch the

liquid in the stomach. In HPS, only a small amount of contrast gets out of the stomach into the duodenum.

Treatment - "What will be done to make my child better?"

Surgery: The treatment for HPS is a surgery called a "pyloromyotomy". The goal of the surgery is to make a cut in the thickened pylorus muscle and open the passage between the stomach and the small intestine (see Figure 2). The surgery may be done laparoscopically or open. Surgery requires general anesthesia.

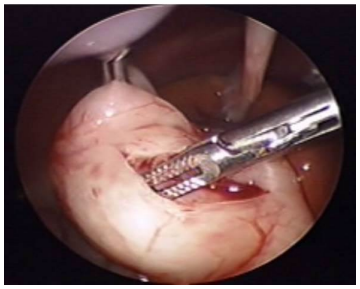

Figure 2: *Image of the surgeon widening the cut in the pylorus.*
(Image credit: *pedsurglibrary.com*)

- *Laparoscopic pyloromyotomy***:** Several small cuts (incisions) are made in the abdomen. Through one of the cuts, a video camera is placed. The surgery itself is done using small instruments placed through the other incisions. Sometimes the surgery cannot be performed laparoscopically.
- *Open pyloromyotomy*: A cut is made in the right upper part of the belly or around the belly button. The pyloromyotomy involves making a cut through the thickened pyloric muscle while leaving the inside layer of the stomach intact. This will allow food to pass through the pylorus and into the small intestine.

Risks/Benefits:

Risks:

- Risks of anesthesia and aspiration of stomach contents with placement of breathing tube, bleeding, infection and unintended damage to other structures in the body such as the duodenum or other small intestine.
- The inside lining of the stomach may also be cut during the pyloromyotomy which would cause leaking of stomach contents into the abdomen. This opening will be repaired and it may take longer to start feeding the baby in order to allow the area to seal.
- Inadequate or incomplete incision of the pylorus muscle. This would result in continued obstructive type symptoms such as vomiting. This is rare and a second surgery may be required to correct this.

Benefits: The stomach will be able to empty food into the small intestine, and the baby will be able to eat without vomiting.

Preoperative preparation: Before surgery can be performed, the infant will be admitted to the hospital and given IV fluids to improve the dehydration and correct abnormalities of the electrolyte levels in the blood.

- Correcting these problems are important to help baby tolerate surgery better and decrease risk of anesthesia.

Postoperative care:

- The baby will be allowed a few hours to recover from anesthesia.
- He or she will be allowed to drink at the discretion of the surgeon. Sometimes surgeons will start oral feeds at a smaller amount than before the surgery to see if your baby tolerates them at first. If baby tolerates, he or she will be given the normal amount of feedings. Some surgeons will allow the baby to try the full amount of feeds after the operating room. Sometimes children will vomit after surgery due to swelling; this can be normal. Infants will stay in the hospital until they are able to tolerate enough feeds without vomiting to stay hydrated.
- For pain, the baby is given acetaminophen (Tylenol®).
- If all goes well, your infant will be discharged within one day of surgery but depends on how he or she is tolerating feedings in order to stay hydrated.

Home Care - "What do I need to do once my child goes home?"

Diet: Your surgeon will discuss the feeding plan after surgery. Vomiting after some feeds may still occur. As long as baby is tolerating most feeds and staying hydrated (have adequate number of weight diapers), it is ok.

Activity: Your child can return to normal activity.

Wound care: Surgical incisions should be kept clean and dry for three days after surgery. Afterwards, the wounds may be washed with soap and water but not submerged in water for seven days after surgery. Most of the time, the stitches used in children are absorbable and do not require removal.

What to call the doctor for: Redness or drainage of the wound(s), fevers >101F, vomiting often, swelling of the belly, return of vomiting problems.

Follow-up care: You will follow up with the pediatric surgeon to make sure your infant is eating well, gaining weight and healing incisions.

Long Term Outcomes - “Are there future conditions to worry about?”

Long-term outcomes are excellent with a very low rate of complications. If your child has lots of vomiting after surgery, then your surgeon will likely do imaging studies to observe how the pylorus muscle looks after surgery.

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