Hypertrophic Pyloric Stenosis

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PRACTICE GAP

Providers should know the presentation of infants with hypertrophic pyloric stenosis (HPS) and obtain an ultrasonographic examination when circumstances suggest the diagnosis. Delay in diagnosis is not uncommon. This is avoidable by maintaining a low threshold to rule out HPS when managing infants with nonbilious vomiting.

OBJECTIVES After completing this article, readers should be able to:

1. Understand the common presenting signs and symptoms of infants with hypertrophic pyloric stenosis.
2. Recognize that a low threshold for ultrasonography may be warranted in infants with nonbilious vomiting.
3. Be aware that although pyloric stenosis is surgically correctable, it is imperative that the child is resuscitated appropriately and all serum electrolyte abnormalities are corrected before surgical intervention.
4. Understand that laparoscopic pyloromyotomy is safe and effective in treating pyloric stenosis and has outstanding long-term results.

ABSTRACT

Hypertrophic pyloric stenosis is a common condition seen in the first 1 to 3 months after birth. Patients typically present with nonbilious projectile emesis after feeds that may result in hypokalemic, hypochloremic metabolic alkalosis. Although inability to tolerate feeds is frequently seen with self-limited conditions such as reflux, a low threshold to obtain an ultrasonographic image is important to prevent a delay in diagnosis. Although operative intervention is the treatment, it is imperative that patients are hydrated and serum electrolyte concentrations normalized before the induction of anesthesia. Laparoscopic pyloromyotomy is safe and effective. Postoperative emesis is normal, and reassurance to parents is appropriate. There is no significant long-term physiologic impairment from pyloric stenosis after successful surgical intervention.

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INTRODUCTION

Hypertrophic pyloric stenosis (HPS) is, as the name denotes, due to hypertrophy of the muscularis of the pylorus, which results in stenosis. This means that the infant experiences partial gastric outlet obstruction, with predictable vomiting of nonbilious gastric contents. It is, notably, not congenital and does not appear in the first days after birth.

Nonbilious spitting up in the early stages of life is common and usually self-limited. Clinicians realize that bilious vomiting represents a potential surgical emergency and are perhaps inclined to worry less when vomiting is not bilious. The obstruction from HPS is partial and progressive. These factors contribute to a frequent delay in diagnosis of pyloric stenosis, which can result in dehydration, weight loss, and metabolic derangement. Compared with most healthy infants with nonbilious spit-up, patients with HPS will not improve without operative intervention.

Patients with HPS will not be helped by the standard interventions for children who are spitting up gastric contents and assumed to have reflux. As sequential nonoperative steps fail (formula changes, thickened feeds, antireflux medications), the therapeutic alliance between the family and the clinician is potentially strained. Delay in diagnosis can exhaust the emotional resources of the parents as the child develops progressive caloric depletion. When the correct diagnosis is finally made and an operation is required, the need for an operation, although curative, can cause significant fright.

This review addresses HPS, focusing on the impact of this diagnosis not only on the child but also on the family. This discussion is, in part, intended to limit the lag in diagnosis and offer information that if communicated to parents will reduce anxiety and maintain the therapeutic alliance with the primary care provider.

PRESENTATION

HPS is seen in approximately 0.2% to 0.4% of children. It is 6 times more likely to be seen in boys, frequently first-born males. (1) The age at presentation is variable. Although the median age at presentation is 6 weeks, it can be seen as early as 2 weeks and as late as 3 months. The classic presenting symptom is projectile, nonbilious emesis after feeds. With time, this can result in hypokalemic, hypochloremic metabolic alkalosis, poor weight gain, and dehydration. If left untreated, this can lead to severe dehydration and significant electrolyte derangements, resulting in lethargy. A recent report described a 5% rate of preoperative respiratory symptoms associated with worsened metabolic alkalosis. (2) Blood-tinged emesis is not uncommon when the infant has partial gastric outlet obstruction due to gastritis or esophagitis. Pyloric stenosis is occasionally associated with an elevated indirect serum bilirubin level. This is secondary to inadequate activity of glucuronyl transferase that is thought to be secondary to inadequate nutrition. Therefore, infants may present with jaundice (due to elevated indirect hyperbilirubinemia not due to cholestasis) that typically resolves after surgical intervention. (3)

Because the obstruction of the pylorus is partial, hydration can be maintained for some time. Thin feeds may pass through the pylorus, especially when parents are encouraged to give smaller, more frequent amounts to an upright infant. Although profound dehydration may not ensue, depletion of serum electrolytes can become severe when vomiting persists, but hydration is maintained with solutions much lower in sodium and chloride concentrations than gastric juice. This can cause false reassurance to both parents and physicians, ultimately contributing to a delay in diagnosis and a severe electrolyte derangement. A high level of suspicion is appropriate when an infant in this age group experiences nonbilious vomiting. There are many possible causes of infantile vomiting, representing multiple nosologic categories, and the potential investigations are numerous. (4) Therefore, it is sensible to exclude or confirm the relatively common HPS early in the diagnostic process.

Various risk factors for the development of HPS have been described, including prematurity, cesarean delivery, maternal smoking, young maternal age, and macrolide use. (5)(6)(7) The genetic pattern of pyloric stenosis is variable, but the classic form is sex influenced and multifactorial. Data have shown increased rate ratios for monozygotic and dizygotic twins, demonstrating a strong familial aggregation with a heritability of approximately 87%; however, it does not follow classic mendelian inheritance patterns. (8) Specific genome-wide significant loci have been reported to be associated with HPS, and research continues regarding this topic. (9)(10)(11)

DIAGNOSIS

Until the early 1980s the only way to make the diagnosis of HPS was to palpate the thickened pylorus (“feel the olive”) or perform an upper gastrointestinal (UGI) series. The former required training and time because the abdominal rectus muscles are thick, the pylorus is not fixed in the abdomen, and the hungry infant cries and
squirms. The UGI series includes exposure to ionizing radiation, possible hypothermia, and possible aspiration of contrast and gastric contents. The evolution of ultrasonography, a sensitive and selective way to make the diagnosis, has made the process both safe and relatively easy. Ultrasonography is considered positive for pyloric stenosis when the muscle thickness is greater than 3 mm and the length of the channel is 15 mm or more (Fig 1). (12)

If ultrasonography for HPS is positive, the road to cure without long-term morbidity is clear and straightforward. If it is negative, then the diagnosis of HPS is unlikely. However, it is important to know that with a normal ultrasonographic image, the diagnosis is not completely eliminated. Ultrasonography of HPS in its early stages may not meet the diagnostic criteria but will do so in the ensuing days. In cases in which the clinical picture remains suggestive, repeating the study is rewarding.

TREATMENT

The cure for pyloric stenosis is pyloromyotomy. This procedure is not an emergency. The metabolic derangement and dehydration should be corrected without delay. This is achieved with intravenous fluid resuscitation with both boluses and continuous infusion, nothing by mouth, and close monitoring of urine output. Intravenous boluses of normal saline are given until urine output is established, after which potassium is added to the administered maintenance fluid. The use of nasogastric decompression is no longer felt to be necessary. (13) The serum electrolyte concentrations are monitored to ensure that all derangements are normalized before proceeding with operative intervention. Some conditions causing partial intestinal obstruction carry the risk of intestinal ischemia, such as midgut volvulus and adhesive small-bowel obstruction. This is not the case with HPS. Clinicians should take as much time as is needed to restore homeostasis before the operation, thereby allowing a safe anesthetic. Anesthesiologists typically expect a bicarbonate level of 30 mEq/dL or lower ($<30$ mmol/L) and a serum chloride level of at least 100 mEq/dL ($\geq100$ mmol/L). (14) Without satisfactory improvements in the metabolic alkalosis, infants may experience decreased respiratory drive after general anesthesia and require prolonged intubation.

In the 19th century, patients with HPS faced a race against death. Some got better without operative intervention, but many died with depletion and malnutrition. The operative cure for HPS, the pyloromyotomy (longitudinal cutting of the muscularis without entry into the lumen) with no suturing, no bypass, no plastic restructuring of the narrowed channel, is a triumph in surgical history due to a quirk in biology. It is not surprising that splitting the hypertrophic muscle, allowing the narrowed pyloric channel to gape open, relieves the obstruction. It is perhaps curious that although the myotomy heals with the 2 cut gaping edges of muscle reapproximated, the stenosis does not recur. Before the discovery of the pyloromyotomy in the first decades of the 20th century, largely attributed to Pierre Fredet in France and to Conrad Ramstedt in Germany, more complicated operative steps were tried as desperate measures in severely depleted infants. Attempts at pyloric bypass (as with gastrojejunostomy) and plastic reconstruction of the thickened pylorus (pyloroplasty) were usually unsuccessful. Pyloromyotomy remains current now, more than a century since its discovery. Although the open technique, either with a right upper quadrant incision or a periumbilical incision, can be used, the laparoscopic approach has become a standard approach. A variety of data demonstrate the benefits and safety profile of this technique. (15)(16)(17) Figure 2 shows an image from a laparoscopic pyloromyotomy. New techniques such as endoscopic myotomy have been described, but their use...
is not at all widespread, and larger-scale studies are necessary to better evaluate the safety profile and long-term success. (18)

HPS demands a skilled and experienced pediatric anesthesiologist. Although the patient will be making an adequate amount of urine and the serum electrolyte concentrations will be in the normal ranges, gastric outlet obstruction persists at the time anesthesia is induced. This puts the infant at risk for aspirating acid-rich gastric contents. Being able to safely induce anesthesia in this setting, which demands rapidly establishing a secure airway in the small patient, is an essential life-sustaining skill for the pediatric anesthesiologist.

The use of atropine sulfate has been described for the treatment of HPS. Although this has not become standard practice, its use and potential success have been described. Atropine is an anticholinergic agent that induces smooth muscle relaxation, thereby potentially leading to relaxation of the hypertrophic pylorus. A recent systematic review and meta-analysis examining the use of atropine for HPS concluded that it is less effective than pyloromyotomy, and its use should be limited to poor candidates for surgery. (19) Its role after incomplete myotomy has also been described, but further data are needed. (20)

COMPLICATIONS
Pyloromyotomy is a safe procedure and is tolerated very well by infants, providing satisfaction for families as they often notice an immediate change in their infant’s ability to tolerate feeds. Significant complications, in addition to bleeding and infection, include unrecognized mucosal perforation and incomplete myotomy. Both rare events require reoperation. Although postoperative persistence of the obstruction is a well-known complication of pyloromyotomy, late recurrence of pyloric stenosis is not expected.

POSTOPERATIVE CARE
Postoperative anxiety can be limited if parents understand before the operation that spit-up and vomit, which is sometimes blood tinged, commonly occur for a while after the operation. Presumably, edema at the surgical site and some degree of gastroparesis from the chronic obstruction perhaps complicated by esophagitis and/or gastritis contribute to a few days of imperfect function. A variety of feeding methods have been trialed in an attempt to reduce postoperative emesis; however, recent data have demonstrated the benefits of ad libitum feeds, including reduction of length of stay and time to full feeds. (21) Although this innocuous emesis may be concerning for an incomplete myotomy, further evaluation is discouraged until at least 1 week after surgery. An ultrasonographic image or a UGI series in the initial days after surgery will show pyloric stenosis and confuse the clinical picture. Discharge to home is often on postoperative day 1 or 2, but this is not always possible. Parents are well served if they know that the hospital stay may take several days more for some patients.

Discharge from the hospital is appropriate when the patient can tolerate adequate feeds without vomiting and is clinically hydrated without receiving intravenous fluid. Parents should know that if, after discharge, the child vomits more than a minimal amount or does not appear well to them, they need to contact the clinical team. That concern should be shared because when patients are discharged as early as postoperative day 1, a perforation at the myotomy site causing peritonitis could present after leaving the hospital. Some practitioners have maintained all patients after pyloromyotomy on medication for reflux in the postoperative period, but no robust evidence supports that practice.

OUTCOMES
Efforts to identify long-term morbidity from HPS have generally found none of significance. The expected outcome is a memory for the parents and barely noticeable scars for the patients.

Figure 2. Laparoscopic image of hypertrophic pylorus after pyloromyotomy.
Summary

- On the basis of consensus, pyloric stenosis is a common entity in pediatric patients, commonly seen at approximately 6 weeks of age and more often in males than in females.

- On the basis of consensus, pyloric stenosis commonly presents with nonbilious, projectile emesis after feeds. A delay in diagnosis occurs when this is attributed to reflux or other etiologies, and formula changes and antireflux measures are offered.

- On the basis of consensus, delay in diagnosis not only causes further dehydration and electrolyte derangements in infants but also leads to increased parental anxiety and frustrations.

- On the basis of consensus and research (evidence quality A and B), ultrasonography is the imaging modality of choice for diagnosis, with a high accuracy.

- On the basis of consensus and research (evidence quality A and B), laparoscopic pyloromyotomy is safe and successful and is the treatment of choice after adequate hydration and improvement of electrolyte abnormalities.

- On the basis of consensus and research (evidence quality A and B), postoperative ad libitum feeding is appropriate and safe.

- On the basis of consensus, there are no long-term effects of pyloric stenosis after successful pyloromyotomy.

References for this article can be found at http://pedsinreview.aappublications.org/content/42/No. 10/539.
1. An infant is admitted to the hospital after radiologic confirmation of pyloric stenosis. On admission, a peripheral intravenous catheter is placed and a basic metabolic panel is obtained. Which of the following is the electrolyte derangement combination most likely expected to be seen in this patient?
   - A. Hyperkalemia, hyperchloremia, metabolic acidosis.
   - B. Hyperkalemia, hyperchloremia, metabolic alkalosis.
   - C. Hyperkalemia, hypochloremia, metabolic alkalosis.
   - D. Hypokalemia, hyperchloremia, metabolic acidosis.
   - E. Hypokalemia, hypochloremia, metabolic alkalosis.

2. A 6-week-old boy presents to your emergency department with 4 days of nonbilious, nonbloody emesis. His parents state that he has been eager to take his bottle every 2 to 3 hours until today, when he became sleepy and has not been waking to feed. On physical examination he is pale, sleeping in his parents’ arms, and responds appropriately during the examination. His anterior fontanelle is sunken. He is tachycardic, with the remainder of his vital signs within normal limits. Which of the following is the most appropriate diagnostic study to order on this patient at this time?
   - A. Abdominal computed tomography.
   - B. Abdominal ultrasonography.
   - C. Head computed tomography.
   - D. pH probe.
   - E. Upper gastrointestinal series.

3. An infant is evaluated in the emergency department for vomiting and dehydration. He is found to have pyloric stenosis on imaging studies, and a basic metabolic panel reveals several electrolyte derangements. Fluid resuscitation for this infant should begin with which of the following fluid regimens?
   - A. Intravenous dextrose 5% ½ normal saline with potassium.
   - B. Intravenous lactated Ringer bolus.
   - C. Intravenous normal saline bolus.
   - D. Nasogastric challenge with an oral rehydration solution.
   - E. Oral challenge with an oral rehydration solution.

4. An infant is being prepared for pyloromyotomy after fluid resuscitation and treatment of electrolyte derangements. While discussing the procedure with the family for informed consent, potential complications are reviewed. Which of the following is a common complication of a pyloromyotomy?
   - A. Infection of the surgical site.
   - B. Persistence of the pyloric obstruction.
   - C. Recurrence of pyloric stenosis.
   - D. Recurrent laryngeal nerve damage.
   - E. Unrecognized mucosal perforation.

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5. An infant who underwent pyloromyotomy a day earlier is being monitored closely in the hospital. He has not had any vomiting overnight and appears hungry. Vital signs are within normal limits, and physical examination findings are normal. Which of the following is the best feeding plan for this infant?

A. Ad libitum formula/human milk.
B. Clear fluids for 48 hours.
C. Continuous nasogastric feeds.
D. Limit the volume of each feed to less than 1 to 2 oz every 3 to 4 hours.
E. Nothing by mouth for 48 hours.