Infantile hypertrophic pyloric stenosis

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INTRODUCTION

Infantile hypertrophic pyloric stenosis (IHPS) is a disorder of young infants caused by hypertrophy of the pylorus, which can progress to near-complete obstruction of the gastric outlet, leading to forceful vomiting.

The clinical manifestations, diagnosis and treatment of IHPS is discussed below. The differential diagnosis of vomiting in infants and other related content is discussed separately. (See "Approach to the infant or child with nausea and vomiting" and "Anesthesia for pyloromyotomy in infants".)

EPIDEMIOLOGY

IHPS occurs in approximately 2 to 3.5 per 1000 live births, although rates and trends vary markedly from region to region [1-7]. It is more common in males than females (4:1 to 6:1) [1,6,8-12] and in infants born preterm as compared with those born at term [13-15]. Approximately 30 to 40 percent of cases occur in first-born children (approximately 1.5-fold increased risk) [8,9,13,16], and it is less common in infants of older mothers [17]. Symptoms usually begin between three and five weeks of age and very rarely occur after 12 weeks of age [18,19].

ETIOLOGY

The etiology of IHPS is unclear but probably is multifactorial, involving genetic predisposition and environmental factors. Neonatal hypergastrinemia and gastric hyperacidity may play a role [10]. Prematurity (<37 weeks gestation) may be a risk factor [14]; one study reported an incidence of 2.99/1000 in preterm infants compared with 2.25/1000 in term infants [20].

Environmental factors — Maternal smoking during pregnancy increases the risk for IHPS by 1.5 to 2-fold [13,14,17,21]. Several studies have suggested that bottle feeding rather than breastfeeding increases the risk for IHPS [15,17,22-25]. In particular, a large population-based study from Denmark found that bottle feeding during the first four months of life increased the risk for IHPS more than fourfold (hazard ratio 4.62, 95% CI 2.78-7.65), and a similar increase in risk was seen for infants that were both bottle- and breastfed [24]. Most of the included subjects used formula for bottle feeds rather than expressed breast milk, so the study did not delineate whether the increased risk is related to the formula or to the mechanism of bottle feeding. Similarly, a large study from Washington state also found that bottle feeding increased the risk for IHPS after adjustment for sex and maternal smoking (odds ratio [OR] 2.31, 95% CI 1.81-2.95) [17].

Genetic factors — Familial aggregation of pyloric stenosis was examined in a large population registry from Denmark [6]. Significant familial aggregation was noted, with a nearly 200-fold higher rate among monozygotic twins and a 20-fold increase among dizygotic twins or siblings as compared with individuals with no affected relatives. The heritability estimate from this study was 87 percent. The maternal and paternal contributions to heritability were similar, suggesting that the intrauterine environment was not an important causal factor.

Several genetic loci that confer a predisposition to IHPS have been identified. A genome-wide association study identified a susceptibility locus that contains the *APOA1* gene (apolipoprotein A1) cluster [26]. This finding provides a possible mechanistic explanation for observed associations between low plasma cholesterol at birth and risk of IHPS. The study also confirmed previous findings of two other susceptibility loci (near *MBNL1* and *NKX2-5*), which are small contributors to overall risk [26,27].

Macrolide antibiotics — Both erythromycin and azithromycin are associated with increased risk of IHPS, particularly when administered to infants younger than two weeks of age [12,28-33]. The risk of IHPS with clarithromycin is not known [34]. (See "Pertussis infection in infants and children: Treatment and prevention", section on 'Choice of regimen'.)

In a retrospective cohort of more than one million infants younger than 90 days, the overall rate of IHPS was 2.29 per 1000 [12]. The risk of IHPS varied with the age of exposure to azithromycin or erythromycin as follows:

- Azithromycin prescribed at age 0 to 14 days – Adjusted OR (aOR) 8.3, 95% CI 2.6-26.0
- Azithromycin prescribed at age 15 to 42 days – aOR 3, 95% CI 1.2-7.2
- Erythromycin prescribed at age 0 to 14 days – aOR 13.3, 95% CI 6.8-15.9
- Erythromycin prescribed at age 15 to 42 days – aOR 4.1, 95% CI 1.7-9.9

A possible association between the use of macrolide antibiotics by women during late pregnancy or while breastfeeding and the development of IHPS in their offspring has been suggested [32,35-37] but is not supported by a meta-analysis [33].

Cases of pyloric stenosis after the use of oral macrolide antibiotics should be reported to the US Food and Drug Administration safety information and adverse event reporting program (MedWatch).

CLINICAL MANIFESTATIONS

Classic presentation — The classic presentation of IHPS is the three- to six-week-old infant who develops immediate postprandial vomiting that is nonbilious and forceful (often described as "projectile" vomiting). The infant then demands to be refed soon afterwards (a "hungry vomiter"). These characteristics were seen in a series of 132 infants who were diagnosed with IHPS in the 1970s, in which 91 percent presented with projectile vomiting after feedings [8]. The majority (83 percent) were boys, and 31 percent were firstborn. In another review of infants diagnosed between 1957 and 1969, the mean age at which the infants began vomiting was 22 days [38]. IHPS rarely occurs after 12 weeks of age [18].

Patients were classically described as being emaciated and dehydrated, with a palpable "olive-like" mass at the lateral edge of the rectus abdominis muscle in the right upper quadrant of the abdomen (see 'Palpation of the "olive"' below). The frequency of palpation of the "olive" typically was quite high (up to 92 percent) in a report from 1975 [8] but has been noted to be less common in subsequent reports [39].

Laboratory evaluation classically showed a hypochloremic, metabolic alkalosis due to the loss of large amounts of gastric hydrochloric acid, the severity of which depended upon the duration of symptoms prior to initial evaluation [40]. Similarly,

hypokalemia is common in infants who have been vomiting for longer than three weeks but typically is not seen in those with a more recent onset of symptoms [41].

Earlier presentation — The typical presentation has changed over time, probably because infants are diagnosed earlier than in the past. Vomiting is still the most common symptom, but infants tend to be better nourished and generally present without significant electrolyte imbalances [42]. This difference was illustrated in a study comparing the presentation of a total of 283 infants diagnosed in the decades prior to 1975, 1985, and 1995 [43]. The mean age at presentation was significantly younger in the more recent decades (mean age 5.4 weeks in 1975 versus 3.4 weeks in 1995). Electrolyte abnormalities were absent in most infants (88 percent) in all groups. A minority of infants present before two weeks of age, and these infants are more likely to have a positive family history of IHPS [44]. These infants tend to have more subtle ultrasonographic abnormalities compared with those presenting after two weeks of age.

The earlier diagnosis of IHPS might be explained by advances in diagnostic imaging or to an increased awareness of the disorder among clinicians. One study found that clinicians in the modern era were less likely to be able to palpate the pyloric "olive" (from 87 percent to 49 percent in the two different time periods) [39]. This finding is probably due to ready availability and reliance on ultrasound to make the diagnosis immediately upon suspicion, so that clinicians in the modern era have less practice at palpating the pyloric "olive" [45]. In addition, infants are now more likely to be evaluated earlier in the course of the disease, when they are well nourished, which makes palpation of "olive" more difficult.

Atypical presentation — In addition to the presenting symptoms described above, the diagnosis of IHPS should be considered in young infants with repeated nonbilious vomiting, hypochloremic alkalosis, and/or rapid clinical improvement after rehydration, even in the absence of projectile vomiting or a palpable "olive" [46,47]. In a few infants with IHPS (<2 percent), the vomiting may be bilious [48].

In infants with medical or surgical problems, the presentation may be atypical or the symptoms initially attributed to other etiologies. In infants with congenital anomalies that affect swallowing (eg, central nervous system anomalies, cleft lip and palate), vomiting may not be projectile [46]. In infants with gastrointestinal anomalies, postoperative vomiting may initially be attributed to adhesions or obstruction at an anastomotic site [46].

In premature infants with IHPS, vomiting may be less forceful, voracious appetite and exaggerated gastric peristalsis may be lacking, and ultrasonographic criteria for diagnosis may not apply [46,47,49,50]. Premature infants may also present at an older chronologic age than term infants [20,51]. One study found a mean age at presentation of 7.6 weeks in infants born prior to 34 weeks gestation versus 5.4 weeks in term infants [51]. In hospitalized premature infants, non-projectile vomiting, weight loss, and lethargy may initially be attributed to sepsis; in this case, negative cultures, rapid improvement with intravenous fluids, and metabolic alkalosis (rather than acidosis) in such infants should prompt consideration of IHPS [46].

Clinical associations — Numerous clinical associations have been described with IHPS.

- Hyperbilirubinemia is one of the most frequent clinical associations of IHPS (occurring in 14 percent of cases in one series [52]). The combination of IHPS and hyperbilirubinemia is known as the icteropyloric syndrome. Unconjugated hyperbilirubinemia is more common than conjugated hyperbilirubinemia; it is often related to the poor hydration and nutritional status of the presenting infant and tends to resolve soon after surgical intervention [53]. In some of these cases, the unconjugated hyperbilirubinemia is an early manifestation of Gilbert syndrome, a benign condition that is caused by mutations in the *UGT1A1* gene (bilirubin uridine diphosphate-glucuronosyltransferase) [52,54]. If the hyperbilirubinemia is conjugated, other etiologies should be considered, including liver disease or sepsis. (See "Unconjugated hyperbilirubinemia in the newborn: Pathogenesis and etiology" and "Gilbert syndrome and unconjugated hyperbilirubinemia due to bilirubin overproduction" and 'Differential diagnosis' below.)

- Midgut malrotation has been described in 0.1 to 5 percent of infants with IHPS, which is a higher percentage than in the general population, but it is unclear whether this is due to higher diagnostic surveillance [55-58]. A familial syndrome with malrotation, congenital short bowel, and IHPS also has been reported but is rare [59-61]. (See "Intestinal malrotation in children".)

- Small case series have described an association between IHPS and an absent or hypoplastic mandibular frenulum, as well as joint hypermobility [62,63]. These minor abnormalities are not functionally significant but may provide clues to the pathogenesis of the disorder, if confirmed in other series.

Other clinical diagnoses occasionally described in patients with IHPS include eosinophilic gastroenteritis [64] and gastric ulcer [65], which may cause pyloric narrowing. Reported associations include hiatal hernia [66], diaphragmatic hernia [58,67],

congenital heart disease [55,58,68], esophageal atresia/tracheoesophageal fistula [55,58], cleft lip and palate [58], obstructive defects of the urinary tract [55], propionic acidemia [69], congenital nephrotic syndrome [70], and congenital hypothyroidism [71]. At least one case of congenital pyloric stenosis has been diagnosed in an infant with an intrauterine history of polyhydramnios [72]. A clinical picture of IHPS was described in a one-month-old infant with familial hyperlipidemia who had an intense hyperechogenicity of the thickened pyloric muscle on ultrasound and fatty infiltration of the pyloric muscle layer at surgical exploration [73]. The symptoms resolved after implementation of a fat-restricted diet.

EVALUATION

Patients with suspected IHPS should be evaluated with a focused physical examination and laboratory testing.

History — Vomiting in IHPS is typically forceful, nonbilious, and tends to occur immediately after feeding. The force and timing can help to distinguish IHPS from physiologic gastroesophageal reflux, in which most episodes of vomiting are not forceful and may occur 10 minutes or more after the meal. A history of bilious vomiting does not exclude IHPS but should raise concern about more distal intestinal obstruction, such as malrotation with volvulus or Hirschsprung disease.

Other elements of the history include:

- Appetite – Classically, infants with IHPS have a strong appetite
- Urine output – Low urine output (four or fewer wet diapers daily) suggests dehydration
- Stool – Gross or occult blood in the stool is not typical for IHPS and suggests the possibility of cow's milk protein-induced proctocolitis or infectious colitis
- Diet – Exposure to cow's milk or soy proteins, through formula or breast milk, is the typical trigger for a dietary protein intolerance
- Medications – Administration of macrolide antibiotics to the mother or infant
- Exposures to caretakers with diarrheal disease
- Family history of pyloric stenosis (see 'Genetic factors' above)

Palpation of the "olive" — The hypertrophied pylorus is palpable in 50 to 90 percent of infants with IHPS. When present, it is virtually pathognomonic of the disease (99 percent specificity in one study [74]), and many surgeons will proceed directly to the operating room based on this finding, without further testing. However, identification of a palpable pylorus (called the "olive") has fallen from 73 to 27 percent over time, with clinicians in the current era making the diagnosis primarily by pyloric ultrasonography [75].

The olive is felt as a firm mass at the lateral edge of the rectus abdominis muscle in the right upper quadrant of the abdomen, approximately the size and shape of an olive. To palpate the "olive," the abdomen should be examined when the infant is quiet to avoid interference from tensed abdominal muscles; providing a pacifier dipped in a sucrose solution may be helpful to quiet the infant during the examination. Ideally, the examination should be performed immediately after emesis because the mass is less likely to be obscured by a distended antrum. As an alternative, the gastric contents can be emptied with a nasogastric tube, which helps to decompress the distended stomach and enhances the palpability of the pyloric mass. Peristaltic waves may be seen progressing across the child's upper abdomen from left to right just before emesis.

The abdomen should also be evaluated for distension and bowel sounds. Abdominal distension or high-pitched bowel sounds suggest intestinal obstruction rather than IHPS, especially if associated with bilious vomiting. Such infants should be evaluated promptly with abdominal radiographs. A stool should be visually inspected for mucus and blood and tested for occult blood. Rectal bleeding raises the possibility of cow's milk protein intolerance in a healthy-appearing infant or of intussusception in an unwell infant. (See 'Differential diagnosis' below.)

Other physical findings — The infant should also be assessed for the following physical findings to evaluate for complications and help exclude disorders that can cause symptoms similar to IHPS:

- Weight and length – To assess nutritional status, especially if the infant has had prolonged vomiting. When serial growth points are available, plotting them on a growth chart helps to establish whether there has been chronic or acute growth failure.
- Mucous membranes and skin turgor – To assess hydration.

- Skin and sclerae – To assess for jaundice, which occurs in patients with IHPS and icteropyloric syndrome but also raises the possibility of underlying liver disease. The presence of eczema raises suspicion for a food allergy. (See 'Clinical associations' above and 'Differential diagnosis' below.)

- Genitalia – To assess for genital ambiguity. Congenital adrenal hyperplasia (CAH) can present in the neonatal period with adrenal crisis. In female infants, CAH is also associated with virilization of the genitalia. (See 'Differential diagnosis' below.)

Laboratory testing — In most infants with suspected IHPS, and especially in those who are ill-appearing, basic laboratory testing is appropriate to assess for dehydration and electrolyte depletion. At a minimum, the evaluation should include an electrolyte panel and complete blood count (CBC).

- Electrolytes – Patients with recent onset of symptomatic IHPS usually have normal laboratory results. Those with prolonged symptoms tend to have low serum chloride and potassium and elevated bicarbonate (a **hypochloremic alkalosis**). Either hypernatremia or hyponatremia may be present. One study showed that serum pH >7.45, chloride <98, and base excess >+3 gave a positive predictive value of 88 percent in diagnosing IHPS in infants presenting with vomiting [76].

  By contrast, an infant with a **hyperkalemic acidosis** is unlikely to have IHPS and should be urgently evaluated for other causes, including adrenal crisis (eg, CAH). (See 'Differential diagnosis' below.)

- Blood urea nitrogen (BUN) and creatinine – To help assess for dehydration and renal insufficiency (eg, prerenal azotemia due to dehydration).

- CBC – The CBC should be normal in infants with uncomplicated IHPS. Abnormal results do not exclude IHPS but should raise suspicion of another cause of vomiting (eg, infection).

- Bilirubin – Infants with jaundice should be evaluated for total and conjugated bilirubin, aspartate aminotransferase (AST), alanine aminotransferase (ALT), and alkaline phosphatase or gamma-glutamyl transpeptidase (GGTP). Unconjugated hyperbilirubinemia is consistent with IHPS (see 'Clinical associations' above). If there are elevations in conjugated bilirubin, ALT, or AST, the infant should be evaluated for underlying liver disease. (See 'Differential diagnosis' below.)

Infants who are severely dehydrated or ill-appearing may warrant further evaluation to rule out other causes including sepsis, intestinal obstruction, or a primary metabolic disorder. (See "Clinical features, evaluation, and diagnosis of sepsis in term and late preterm infants".)

Imaging — When the "olive" and/or peristaltic waves cannot be detected, the diagnosis of IHPS can be confirmed by imaging. In general, ultrasonography is the procedure of choice for infants with typical clinical features of pyloric stenosis [11,74,77]. A fluoroscopic upper gastrointestinal (UGI) series is preferred if the infant has bilious vomiting or other features suggesting more distal obstruction, as outlined in the radiography Appropriateness Criteria tables for infants with vomiting (variants 2 and 4) [78].

Ultrasonography — Ultrasound is the procedure of choice for infants presenting with typical symptoms of IHPS (new onset of nonbilious vomiting in infants up to three months of age), if a sonographer with experience in detecting pyloric stenosis is available. In experienced hands, the sensitivity and specificity of ultrasonography for IHPS are above 95 percent, but the accuracy is operator dependent [79-81]. If the ultrasound is negative or equivocal but the patient remains symptomatic, the study should be repeated a few days to a week later.

IHPS is characterized by a classic "target" sign on transverse view. The measurements most commonly used are pyloric muscle thickness (PMT), pyloric muscle length (PML), and pyloric diameter (PD) (image 1). Criteria defining the upper limits of normal range from PMT 3 to 4 mm, PML 15 to 19 mm, and PD 10 to 14 mm [82-84]. Measurements within or above these ranges support the diagnosis of IHPS. These measurements may not be applicable in premature infants [47,49]. Normal values vary with the size of the infant, so results should be interpreted with caution in particularly small or young infants [50,85,86]. Each of these measurements has been touted as the most reliable of the three [87-90]. However, all three measurements typically are used together in practice.

Other less frequently proposed ultrasonographic criteria for the diagnosis of IHPS have included pyloric volume (PV) and pyloric ratio (PR). The PV has been defined as equal to 0.25 pi × PD(2) × PML; infants with IHPS were found to have a significantly higher value than were those without IHPS [91]. The PR is described as PMT/PD and is significantly higher in infants with IHPS, with a sensitivity and specificity of 96 and 94 percent, respectively [92].

Ultrasonographic diagnosis of IHPS has potential pitfalls. The ultrasonographic findings of pylorospasm may, at least transiently, mimic those of IHPS [93]. The ultrasonographer also must be aware of potential false-positive readings caused by a fluid-filled duodenal bulb and gastric antrum and false-negative interpretation resulting from poor visualization caused by overlying bowel gas [94]. A posterior sonographic view may be helpful in the latter situation [95].

Fluoroscopic upper gastrointestinal series — A fluoroscopic UGI series is generally used only if the physical examination and ultrasonography are nondiagnostic or if there are signs or symptoms suggesting more distal obstruction, such as bilious vomiting or dilated small bowel on conventional radiography. The classic signs on the UGI study are an elongated pyloric canal (the "string" sign) (image 2), two thin tracks of barium along the pyloric canal created by compressed pyloric mucosa (the "double-track" sign), a tapered point ending the pyloric canal (the "beak" sign), and a prepyloric bulge of barium (the "shoulder" sign) [96-98]. The main disadvantage of a UGI is radiation exposure.

Upper endoscopy — Upper endoscopy usually is reserved for patients in whom other imaging modalities are inconclusive (or if warranted to evaluate for another gastrointestinal disorder because of symptoms or signs that are atypical for IHPS). In infants with IHPS, the mucosa of the antrum and pylorus appear thickened [99]. Endoscopy is useful for obtaining tissue sample if eosinophilic gastritis is suspected, as a mimicker of IHPS.

DIAGNOSIS

The diagnosis of IHPS is suspected when an infant presents with suggestive features, as described above. The diagnosis can be confirmed by palpation of an "olive" or by the presence of typical findings on imaging studies. In many centers, ultrasonography is performed to confirm the diagnosis of IHPS prior to surgery, even if an "olive" is palpated. (See 'Evaluation' above.)

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of vomiting in early infancy includes the following disorders (table 1). In most cases, these are easily distinguished from IHPS by the history, physical examination, and/or initial laboratory tests. (See "Approach to the infant or child with nausea and vomiting", section on 'Neonates and young infants'.)

- Gastroesophageal reflux – Physiologic reflux in newborns and infants is common and is characterized by effortless regurgitation in an otherwise healthy infant (a "happy spitter"). The regurgitation may seem forceful at times. Physiologic reflux is not associated with electrolyte abnormalities, tends to be chronic rather than progressive, and rarely causes weight loss. (See "Gastroesophageal reflux in infants".)

- Cow's milk protein intolerance – Intolerance to cow's milk or soy protein typically presents with colitis (blood-tinged stools) but also may affect the small bowel (enteropathy), presenting with recurrent vomiting. The presence of blood-tinged stools in infants who are fed cow's milk or soy protein (through formula or through the maternal diet, if breastfed) raises this possibility. (See "Food protein-induced allergic proctocolitis of infancy".)

- Adrenal crisis – Newborn infants with adrenal insufficiency may present with an adrenal crisis, which is manifested by vomiting and dehydration. This is a life-threatening condition and should be evaluated and treated urgently. Key features of adrenal crisis are disproportionate hypotension and **hyper**kalemic **acid**osis (rather than the hypokalemic alkalosis typically seen in IHPS). The most common cause of adrenal insufficiency in infants is congenital adrenal hyperplasia (CAH). Females with CAH tend to have virilized or ambiguous genitalia; males usually have no obvious genital abnormalities. (See "Clinical manifestations and diagnosis of adrenal insufficiency in children", section on 'Adrenal crisis'.)

- Intestinal obstruction – Causes of intestinal obstruction in early infancy include malrotation (with or without volvulus), Hirschsprung disease, and intussusception. Intestinal obstruction should be considered in infants with bilious vomiting and abdominal distension, especially if high-pitched bowel sounds or bloody stools are present. If intestinal obstruction is suspected, the specific diagnosis is often suggested by the patient's history and then confirmed with appropriate radiologic imaging, including a plain radiograph. (See "Approach to the infant or child with nausea and vomiting", section on 'Intestinal obstruction'.)

- Liver disease – Liver disease in infants may present with symptoms resembling IHPS, including vomiting, poor weight gain, and jaundice. Infants with biliary atresia also may have acholic (very pale-colored) stools. Infants with conjugated hyperbilirubinemia should be further evaluated for liver disease, including biliary atresia, biliary cysts, and metabolic disorders. (See "Unconjugated hyperbilirubinemia in the newborn: Pathogenesis and etiology" and "Gilbert syndrome and unconjugated hyperbilirubinemia due to bilirubin overproduction" and "Causes of cholestasis in neonates and young infants".)

TREATMENT

Definitive management of IHPS is surgical pyloromyotomy. The timing of surgery depends upon the clinical status of the infant. If the child is well hydrated with normal electrolytes, and if surgeons with expertise in the procedure are available, surgery may take place on the day of diagnosis [100]. Surgery should be delayed in the setting of dehydration and/or electrolyte derangements [101].

Fluid and electrolyte management — Infants presenting with normal electrolyte values and no dehydration, as is the case with more than 60 percent of patients, should receive maintenance intravenous fluids such as 5% dextrose with one-half normal saline (0.45% NaCl) and 10 to 20 mEq KCl per L. Infants with moderate or severe dehydration require more intensive management by administering fluids with higher NaCl concentrations (typically normal saline [0.9% NaCl]), given as an initial fluid bolus and/or higher rates of administration (1.5 to 2 times maintenance) until the calculated fluid deficit is repleted. In severely dehydrated infants, renal function should be assessed prior to adding potassium to the intravenous fluids. (See "Treatment of hypovolemia (dehydration) in children".)

An expert panel recommended that laboratory values be normalized to at least the following thresholds prior to pyloromyotomy: pH ≤7.45, base excess ≤3.5, bicarbonate <26 mmol/L, sodium ≥132 mmol/L, potassium ≥3.5 mmol/L, chloride ≥100 mmol/L, and glucose ≥4.0 mmol/L (72 mg/dL) [102]. If alkalosis is not corrected prior to surgery, it has been associated with an increased risk of postoperative apnea [103,104]. Patients with severe hypokalemia (serum potassium <2.5 mEq/L) or hyponatremia (serum sodium <120 mEq/L) have increased risks and should be managed particularly carefully, with expert consultation if needed. (See "Hypokalemia in children" and "Hyponatremia in children: Evaluation and management".)

Pyloromyotomy

- **Technique** – The classical operation for IHPS is Ramstedt pyloromyotomy, which involves a longitudinal incision of the hypertrophic pylorus with blunt dissection to the level of the submucosa; it relieves the constriction and allows normal passage of stomach contents into the duodenum. Laparoscopic pyloromyotomy (picture 1) is a minimally invasive version of the Ramstedt procedure that has been associated with a lower incidence of postoperative emesis and a shorter

hospital stay but occasionally results in incomplete pyloromyotomy or mucosal perforation [105-108]. A transumbilical approach also may be used but has longer operating time [109].

Open and laparoscopic pyloromyotomy were compared in a prospective trial in 200 infants with ultrasonographically confirmed IHPS, who were randomly assigned to open or laparoscopic pyloromyotomy [105]. There were no differences between groups in operating time, time to full feeding, or length of stay. However, infants in the laparoscopic group had fewer episodes of emesis (1.9 versus 2.6) and received fewer doses of analgesia (1.6 versus 2.2) than those in the open group. A similar randomized study also reported more rapid return to enteral feeding and shorter hospital stay among infants treated laparoscopically, although in 3 to 5 percent of laparoscopically performed cases, the pyloromyotomy was incomplete [106]. The study was performed at six centers with extensive experience in laparoscopic techniques.

- **Resuming feeds postoperatively** – Feeding can be resumed in most infants within a few hours after surgery. Modest regurgitation occurs in as many as 80 percent of infants after pyloromyotomy [8] and should not delay feedings. Vigorous postoperative vomiting is infrequent. In a meta-analysis, infants offered ad lib feedings four hours after operation tolerated full feedings sooner and had a shorter hospital stay compared with infants receiving an incremental feeding schedule, despite having more emesis episodes [110]. Radiologic evaluation should be performed if vomiting persists beyond five days postoperatively [111], with the understanding that interpretation of the study may be difficult because of postoperative swelling.

- **Postoperative monitoring** – Infants with IHPS are at risk for apnea postoperatively because of their young age and effects of anesthesia; we suggest monitoring for apnea for at least 24 hours postoperatively. Those with underlying alkalosis may be at increased risk based on limited and inferential data [104]. As mentioned above, this is one reason why alkalosis and other electrolyte abnormalities should be corrected prior to surgery.

- **Complications** – The rate of serious complications from pyloromyotomy is low. Mucosal perforations occur in fewer than 1 percent of patients and are readily recognized intraoperatively [112]. The pyloromyotomy is incomplete in approximately 1 percent of patients treated with a laparoscopic approach by an experienced surgeon and is even rarer in those undergoing the open procedure.

- **Follow-up** – Infants should be monitored through surgical recovery and return to normal feeds. Thereafter, they need only routine pediatric care, including monitoring of growth, unless new symptoms develop. Pyloromyotomy is curative in the vast majority of infants [112]. Gastroesophageal reflux is common, as it is in other healthy infants, and does not warrant concern unless it is very severe or accompanied by other symptoms (see "Gastroesophageal reflux in infants"). Infants with other persistent or recurrent symptoms warrant further evaluation, including for disorders other than pyloric stenosis. If abdominal ultrasonography is performed postoperatively, caution should be exercised in interpretation because the thickened muscle and enlarged diameter persist to eight months and one year, respectively [113].

Balloon dilation — Endoscopically guided balloon dilation for IHPS has been described [114]. However, because balloon dilatation does not reliably disrupt the seromuscular ring of the pylorus [115], attempts at this technique are best reserved for patients in whom general anesthesia would pose a significant risk or in whom a surgical approach to the pylorus is not possible.

Conservative management — Conservative management of infants with IHPS using anticholinergics has also been described. This approach typically involves a trial of continuous nasoduodenal feedings, generally lasting several months, until the obstructive process becomes less significant as the infant gains weight [116]. Oral and intravenous atropine sulfate (which relaxes the pyloric musculature) also have been described in small case series [117-121]. Small nonrandomized studies and a meta-analysis suggest that oral treatment with atropine sulfate is effective in approximately 75 percent of patients [121-123]. Because data on conservative management are very limited, and because surgery is both safe and effective, we suggest conservative management only for infants in whom a surgical approach is either not advisable or not feasible.

INFORMATION FOR PATIENTS

UpToDate offers two types of patient education materials, "The Basics" and "Beyond the Basics." The Basics patient education pieces are written in plain language, at the 5th to 6th grade reading level, and they answer the four or five key questions a patient might have about a given condition. These articles are best for patients who want a general overview and who prefer short, easy-to-read materials. Beyond the Basics patient education pieces are longer, more sophisticated, and more detailed. These articles are written at the 10th to 12th grade reading level and are best for patients who want in-depth information and are comfortable with some medical jargon.

Here are the patient education articles that are relevant to this topic. We encourage you to print or e-mail these topics to your patients. (You can also locate patient education articles on a variety of subjects by searching on "patient info" and the keyword(s) of interest.)

- Basics topics (see "Patient education: Pyloric stenosis in babies (The Basics)")

SUMMARY AND RECOMMENDATIONS

Infantile hypertrophic pyloric stenosis (IHPS) is characterized by hypertrophy of the pylorus, eventually progressing to near-complete obstruction of the gastric outlet.

- The etiology of IHPS is obscure but probably is multifactorial, involving genetic predisposition and environmental factors. In particular, IHPS has been associated with the administration of macrolide antibiotics to infants during the first few weeks of life and perhaps to their mothers during last gestation or lactation. (See 'Etiology' above.)

- Symptoms usually begin between three and five weeks of age and very rarely occur after 12 weeks of age. Typical presenting symptoms are nonbilious vomiting that is forceful and occurs immediately after feeding, while the infant remains hungry. A firm, "olive-like" mass may be palpable in the right upper quadrant of the abdomen, and there may be signs of dehydration. In a few infants, the vomiting may be bilious. In the past, infants were often malnourished at presentation. In premature infants with IHPS, the vomiting may be less forceful and they may not display hunger. (See 'Clinical manifestations' above.)

- In most infants with suspected IHPS, and especially in those who are ill-appearing, basic laboratory testing is appropriate to assess for dehydration and electrolyte depletion. At a minimum, the evaluation should include an electrolyte panel and complete blood count (CBC). Infants with IHPS may have normal results or may have low serum chloride and potassium and elevated bicarbonate (a hypochloremic alkalosis). (See 'Laboratory testing' above.)

- The diagnosis of IHPS is usually established by abdominal ultrasound, on which IHPS is characterized by increased pyloric muscle thickness (PMT) length and diameter (image 1). The accuracy of ultrasound for the diagnosis of IHPS is high if the sonographer is experienced in diagnosing this disorder. A fluoroscopic upper gastrointestinal (UGI) series may be used

if the physical examination and ultrasonography are nondiagnostic or if there are signs or symptoms suggesting more distal obstruction, such as bilious vomiting or dilated small bowel on conventional radiography (image 2). (See 'Imaging' above.)

- The differential diagnosis of IHPS includes physiologic gastroesophageal reflux and cow's milk protein intolerance, which are relatively benign. More severe disorders that may present with vomiting in the first two months of life include adrenal crisis, intestinal obstruction (eg, due to malrotation, Hirschsprung disease, or intussusception), and liver disease (eg, biliary atresia or metabolic defects). These disorders should be considered in infants with atypical presenting features, including hypotension, vomiting that is bilious, abdominal distension, or jaundice with conjugated hyperbilirubinemia (table 1). (See 'Differential diagnosis' above.)

- IHPS is typically treated with surgical pyloromyotomy. If the child is well hydrated with normal electrolytes, and if surgeons with expertise in the procedure are available, surgery may take place on the day of diagnosis. Surgery should be delayed in the setting of dehydration and/or electrolyte derangements until these abnormalities are corrected with appropriate fluid and electrolyte therapy. Mild regurgitation after pyloromyotomy is common and should not delay the initiation of feeding. (See 'Pyloromyotomy' above.)

REFERENCES

1. To T, Wajja A, Wales PW, Langer JC. Population demographic indicators associated with incidence of pyloric stenosis. Arch Pediatr Adolesc Med 2005; 159:520.
2. Habbick BF, To T. Incidence of infantile hypertrophic pyloric stenosis in Saskatchewan, 1970-85. CMAJ 1989; 140:395.
3. Hedbäck G, Abrahamsson K, Husberg B, et al. The epidemiology of infantile hypertrophic pyloric stenosis in Sweden 1987-96. Arch Dis Child 2001; 85:379.
4. Sule ST, Stone DH, Gilmour H. The epidemiology of infantile hypertrophic pyloric stenosis in Greater Glasgow area, 1980-96. Paediatr Perinat Epidemiol 2001; 15:379.

5. Sommerfield T, Chalmers J, Youngson G, et al. The changing epidemiology of infantile hypertrophic pyloric stenosis in Scotland. Arch Dis Child 2008; 93:1007.
6. Krogh C, Fischer TK, Skotte L, et al. Familial aggregation and heritability of pyloric stenosis. JAMA 2010; 303:2393.
7. Kapoor R, Kancherla V, Cao Y, et al. Prevalence and descriptive epidemiology of infantile hypertrophic pyloric stenosis in the United States: A multistate, population-based retrospective study, 1999-2010. Birth Defects Res 2019; 111:159.
8. Gibbs MK, Van Herrden JA, Lynn HB. Congenital hypertrophic pyloric stenosis. Surgical experience. Mayo Clin Proc 1975; 50:312.
9. Chen EA, Luks FI, Gilchrist BF, et al. Pyloric stenosis in the age of ultrasonography: fading skills, better patients? J Pediatr Surg 1996; 31:829.
10. Rogers IM. The enigma of pyloric stenosis. Some thoughts on the aetiology. Acta Paediatr 1997; 86:6.
11. Godbole P, Sprigg A, Dickson JA, Lin PC. Ultrasound compared with clinical examination in infantile hypertrophic pyloric stenosis. Arch Dis Child 1996; 75:335.
12. Eberly MD, Eide MB, Thompson JL, Nylund CM. Azithromycin in early infancy and pyloric stenosis. Pediatrics 2015; 135:483.
13. Krogh C, Gørtz S, Wohlfahrt J, et al. Pre- and perinatal risk factors for pyloric stenosis and their influence on the male predominance. Am J Epidemiol 2012; 176:24.
14. Svenningsson A, Svensson T, Akre O, Nordenskjöld A. Maternal and pregnancy characteristics and risk of infantile hypertrophic pyloric stenosis. J Pediatr Surg 2014; 49:1226.
15. Zhu J, Zhu T, Lin Z, et al. Perinatal risk factors for infantile hypertrophic pyloric stenosis: A meta-analysis. J Pediatr Surg 2017; 52:1389.
16. Mandell GA, Wolfson PJ, Adkins ES, et al. Cost-effective imaging approach to the nonbilious vomiting infant. Pediatrics 1999; 103:1198.
17. McAteer JP, Ledbetter DJ, Goldin AB. Role of bottle feeding in the etiology of hypertrophic pyloric stenosis. JAMA Pediatr 2013; 167:1143.
18. Spicer RD. Infantile hypertrophic pyloric stenosis: a review. Br J Surg 1982; 69:128.

19. Aboagye J, Goldstein SD, Salazar JH, et al. Age at presentation of common pediatric surgical conditions: Reexamining dogma. J Pediatr Surg 2014; 49:995.
20. Stark CM, Rogers PL, Eberly MD, Nylund CM. Association of prematurity with the development of infantile hypertrophic pyloric stenosis. Pediatr Res 2015; 78:218.
21. Sørensen HT, Nørgård B, Pedersen L, et al. Maternal smoking and risk of hypertrophic infantile pyloric stenosis: 10 year population based cohort study. BMJ 2002; 325:1011.
22. Pisacane A, de Luca U, Criscuolo L, et al. Breast feeding and hypertrophic pyloric stenosis: population based case-control study. BMJ 1996; 312:745.
23. Habbick BF, Khanna C, To T. Infantile hypertrophic pyloric stenosis: a study of feeding practices and other possible causes. CMAJ 1989; 140:401.
24. Krogh C, Biggar RJ, Fischer TK, et al. Bottle-feeding and the Risk of Pyloric Stenosis. Pediatrics 2012; 130:e943.
25. Wayne C, Hung JH, Chan E, et al. Formula-feeding and hypertrophic pyloric stenosis: is there an association? A case-control study. J Pediatr Surg 2016; 51:779.
26. Feenstra B, Geller F, Carstensen L, et al. Plasma lipids, genetic variants near APOA1, and the risk of infantile hypertrophic pyloric stenosis. JAMA 2013; 310:714.
27. Feenstra B, Geller F, Krogh C, et al. Common variants near MBNL1 and NKX2-5 are associated with infantile hypertrophic pyloric stenosis. Nat Genet 2012; 44:334.
28. Friedman DS, Curtis CR, Schauer SL, et al. Surveillance for transmission and antibiotic adverse events among neonates and adults exposed to a healthcare worker with pertussis. Infect Control Hosp Epidemiol 2004; 25:967.
29. Honein MA, Paulozzi LJ, Himelright IM, et al. Infantile hypertrophic pyloric stenosis after pertussis prophylaxis with erythromcyin: a case review and cohort study. Lancet 1999; 354:2101.
30. Centers for Disease Control and Prevention (CDC). Hypertrophic pyloric stenosis in infants following pertussis prophylaxis with erythromycin--Knoxville, Tennessee, 1999. MMWR Morb Mortal Wkly Rep 1999; 48:1117.
31. Morrison W. Infantile hypertrophic pyloric stenosis in infants treated with azithromycin. Pediatr Infect Dis J 2007; 26:186.

32. Lund M, Pasternak B, Davidsen RB, et al. Use of macrolides in mother and child and risk of infantile hypertrophic pyloric stenosis: nationwide cohort study. BMJ 2014; 348:g1908.
33. Abdellatif M, Ghozy S, Kamel MG, et al. Association between exposure to macrolides and the development of infantile hypertrophic pyloric stenosis: a systematic review and meta-analysis. Eur J Pediatr 2019; 178:301.
34. Hauben M, Amsden GW. The association of erythromycin and infantile hypertrophic pyloric stenosis: causal or coincidental? Drug Saf 2002; 25:929.
35. Mahon BE, Rosenman MB, Kleiman MB. Maternal and infant use of erythromycin and other macrolide antibiotics as risk factors for infantile hypertrophic pyloric stenosis. J Pediatr 2001; 139:380.
36. Cooper WO, Ray WA, Griffin MR. Prenatal prescription of macrolide antibiotics and infantile hypertrophic pyloric stenosis. Obstet Gynecol 2002; 100:101.
37. Sørensen HT, Skriver MV, Pedersen L, et al. Risk of infantile hypertrophic pyloric stenosis after maternal postnatal use of macrolides. Scand J Infect Dis 2003; 35:104.
38. Dodge JA. Infantile hypertrophic pyloric stenosis in Belfast, 1957-1969. Arch Dis Child 1975; 50:171.
39. Macdessi J, Oates RK. Clinical diagnosis of pyloric stenosis: a declining art. BMJ 1993; 306:553.
40. Touloukian RJ, Higgins E. The spectrum of serum electrolytes in hypertrophic pyloric stenosis. J Pediatr Surg 1983; 18:394.
41. Nmadu PT. Alterations in serum electrolytes in congenital hypertrophic pyloric stenosis: a study in Nigerian children. Ann Trop Paediatr 1992; 12:169.
42. Taylor ND, Cass DT, Holland AJ. Infantile hypertrophic pyloric stenosis: has anything changed? J Paediatr Child Health 2013; 49:33.
43. Papadakis K, Chen EA, Luks FI, et al. The changing presentation of pyloric stenosis. Am J Emerg Med 1999; 17:67.
44. Demian M, Nguyen S, Emil S. Early pyloric stenosis: a case control study. Pediatr Surg Int 2009; 25:1053.
45. Shaoul R, Enav B, Steiner Z, et al. Clinical presentation of pyloric stenosis: the change is in our hands. Isr Med Assoc J 2004; 6:134.
46. Weinstein MM, Seibert JJ, Erenberg A. Six atypical presentations of congenital hypertrophic pyloric stenosis. Clin Pediatr (Phila) 1979; 18:120.

47. Janik JS, Wayne ER, Janik JP. Pyloric stenosis in premature infants. Arch Pediatr Adolesc Med 1996; 150:223.
48. Piroutek MJ, Brown L, Thorp AW. Bilious vomiting does not rule out infantile hypertrophic pyloric stenosis. Clin Pediatr (Phila) 2012; 51:214.
49. Cosman BC, Sudekum AE, Oakes DD, de Vries PA. Pyloric stenosis in a premature infant. J Pediatr Surg 1992; 27:1534.
50. Bisset RA, Gupta SC. Hypertrophic pyloric stenosis, ultrasonic appearances in a small baby. Pediatr Radiol 1988; 18:405.
51. Costanzo CM, Vinocur C, Berman L. Prematurity Affects Age of Presentation of Pyloric Stenosis. Clin Pediatr (Phila) 2017; 56:127.
52. Hua L, Shi D, Bishop PR, et al. The role of UGT1A1*28 mutation in jaundiced infants with hypertrophic pyloric stenosis. Pediatr Res 2005; 58:881.
53. Stringer MD, Brereton RJ. Current management of infantile hypertrophic pyloric stenosis. Br J Hosp Med 1990; 43:266.
54. Trioche P, Chalas J, Francoual J, et al. Jaundice with hypertrophic pyloric stenosis as an early manifestation of Gilbert syndrome. Arch Dis Child 1999; 81:301.
55. Applegate MS, Druschel CM. The epidemiology of infantile hypertrophic pyloric stenosis in New York State, 1983 to 1990. Arch Pediatr Adolesc Med 1995; 149:1123.
56. Zeidan B, Wyatt J, Mackersie A, Brereton RJ. Recent results of treatment of infantile hypertrophic pyloric stenosis. Arch Dis Child 1988; 63:1060.
57. Croitoru D, Neilson I, Guttman FM. Pyloric stenosis associated with malrotation. J Pediatr Surg 1991; 26:1276.
58. Schärli A, Sieber WK, Kiesewetter WB. Hypertrophic pyloric stenosis at the Children's Hospital of Pittsburgh from 1912 to 1967. A critical review of current problems and complications. J Pediatr Surg 1969; 4:108.
59. Brereton RJ. Midgut malrotation associated with hypertrophic pyloric stenosis had concomitant congenital short bowel. J Pediatr Surg 1992; 27:796.
60. Levin MD, Troian VV. [Combined malrotation of the intestines with pyloric stenosis in a 6-day-old infant]. Klin Khir 1984; :56.
61. Sansaricq C, Chen WJ, Manka M, et al. Familial congenital short small bowel with associated defects. A long-term survival. Clin Pediatr (Phila) 1984; 23:453.

62. De Felice C, Di Maggio G, Zagordo L, et al. Hypoplastic or absent mandibular frenulum: a new predictive sign of infantile hypertrophic pyloric stenosis. J Pediatr 2000; 136:408.

63. De Felice C, Di Maggio G, Toti P, et al. Infantile hypertrophic pyloric stenosis and asymptomatic joint hypermobility. J Pediatr 2001; 138:596.

64. Blankenberg FG, Parker BR, Sibley E, Kerner JA. Evolving asymmetric hypertrophic pyloric stenosis associated with histologic evidence of eosinophilic gastroenteritis. Pediatr Radiol 1995; 25:310.

65. Kelsey D, Stayman JW Jr, McLaughlin ED, Mebane W. Massive bleeding in a newborn infant from a gastric ulcer associated with hypertrophic pyloric stenosis. Surgery 1968; 64:979.

66. Iijima T, Okamatsu T, Matsumura M, Yatsuzuka M. Hypertrophic pyloric stenosis associated with hiatal hernia. J Pediatr Surg 1996; 31:277.

67. al-Salem AH, Grant C, Khwaja S. Infantile hypertrophic pyloric stenosis and congenital diaphragmatic hernia. J Pediatr Surg 1990; 25:607.

68. Feenstra B, Gørtz S, Lund M, et al. Co-occurrence of infantile hypertrophic pyloric stenosis and congenital heart defects: a nationwide cohort study. Pediatr Res 2019; 85:955.

69. Lehnert W, Junker A, Wehinger H, et al. [Propionic acidemia associated with hypertrophic pyloric stenosis and bouts of severe hyperglycemia (author's transl)]. Monatsschr Kinderheilkd 1980; 128:720.

70. Grahame-Smith HN, Ward PS, Jones RD. Finnish type congenital nephrotic syndrome in twins: presentation with pyloric stenosis. J R Soc Med 1988; 81:358.

71. Hirano T, Jogamoto M, Tamura K. Association of congenital hypothyroidism with congenital hypertrophic pyloric stenosis. Acta Paediatr Jpn 1988; 30:371.

72. Houben CH, Kiely EM. Congenital hypertrophic pyloric stenosis with associated polyhydramnios in a premature infant. Eur J Pediatr Surg 1997; 7:184.

73. Veyrac C, Couture A, Bongrand AF, et al. Atypical pyloric stenosis in an infant with familial hyperlipidemia. Pediatr Radiol 1996; 26:402.

74. White MC, Langer JC, Don S, DeBaun MR. Sensitivity and cost minimization analysis of radiology versus olive palpation for the diagnosis of hypertrophic pyloric stenosis. J Pediatr Surg 1998; 33:913.

75. Bakal U, Sarac M, Aydin M, et al. Recent changes in the features of hypertrophic pyloric stenosis. Pediatr Int 2016; 58:369.

76. Oakley EA, Barnett PL. Is acid base determination an accurate predictor of pyloric stenosis? J Paediatr Child Health 2000; 36:587.

77. Breaux CW Jr, Georgeson KE, Royal SA, Curnow AJ. Changing patterns in the diagnosis of hypertrophic pyloric stenosis. Pediatrics 1988; 81:213.

78. American College of Radiology appropriateness criteria for vomiting in infants up to 3 months of age. Available at: https://acsearch.acr.org/docs/69445/Narrative/ (Accessed on August 31, 2018).

79. Sivitz AB, Tejani C, Cohen SG. Evaluation of hypertrophic pyloric stenosis by pediatric emergency physician sonography. Acad Emerg Med 2013; 20:646.

80. Niedzielski J, Kobielski A, Sokal J, Krakós M. Accuracy of sonographic criteria in the decision for surgical treatment in infantile hypertrophic pyloric stenosis. Arch Med Sci 2011; 7:508.

81. Hernanz-Schulman M. Pyloric stenosis: role of imaging. Pediatr Radiol 2009; 39 Suppl 2:S134.

82. Kofoed PE, Høst A, Elle B, Larsen C. Hypertrophic pyloric stenosis: determination of muscle dimensions by ultrasound. Br J Radiol 1988; 61:19.

83. Rohrschneider WK, Mittnacht H, Darge K, Tröger J. Pyloric muscle in asymptomatic infants: sonographic evaluation and discrimination from idiopathic hypertrophic pyloric stenosis. Pediatr Radiol 1998; 28:429.

84. Hallam D, Hansen B, Bødker B, et al. Pyloric size in normal infants and in infants suspected of having hypertrophic pyloric stenosis. Acta Radiol 1995; 36:261.

85. Said M, Shaul DB, Fujimoto M, et al. Ultrasound measurements in hypertrophic pyloric stenosis: don't let the numbers fool you. Perm J 2012; 16:25.

86. Iqbal CW, Rivard DC, Mortellaro VE, et al. Evaluation of ultrasonographic parameters in the diagnosis of pyloric stenosis relative to patient age and size. J Pediatr Surg 2012; 47:1542.

87. Blumhagen JD, Maclin L, Krauter D, et al. Sonographic diagnosis of hypertrophic pyloric stenosis. AJR Am J Roentgenol 1988; 150:1367.

88. Kovalivker M, Erez I, Shneider N, et al. The value of ultrasound in the diagnosis of congenital hypertrophic pyloric stenosis. Clin Pediatr (Phila) 1993; 32:281.

89. Stunden RJ, LeQuesne GW, Little KE. The improved ultrasound diagnosis of hypertrophic pyloric stenosis. Pediatr Radiol 1986; 16:200.

90. Tunell WP, Wilson DA. Pyloric stenosis: diagnosis by real time sonography, the pyloric muscle length method. J Pediatr Surg 1984; 19:795.

91. Westra SJ, de Groot CJ, Smits NJ, Staalman CR. Hypertrophic pyloric stenosis: use of the pyloric volume measurement in early US diagnosis. Radiology 1989; 172:615.

92. Lowe LH, Banks WJ, Shyr Y. Pyloric ratio: efficacy in the diagnosis of hypertrophic pyloric stenosis. J Ultrasound Med 1999; 18:773.

93. Cohen HL, Zinn HL, Haller JO, et al. Ultrasonography of pylorospasm: findings may simulate hypertrophic pyloric stenosis. J Ultrasound Med 1998; 17:705.

94. Khamapirad T, Athey PA. Ultrasound diagnosis of hypertrophic pyloric stenosis. J Pediatr 1983; 102:23.

95. Sargent SK, Foote SL, Mooney DP, Shorter NA. The posterior approach to pyloric sonography. Pediatr Radiol 2000; 30:256.

96. Meuwissen T, Sloof JP. Roentgenologic diagnosis of congenital hypertrophic pyloric stenosis. Acta Pediatr 1932; 14:19.

97. Haran PJ Jr, Darling DB, Sciammas F. The value of the double-track sign as a differentiating factor between pylorospasm and hypertrophic pyloric stenosis in infants. Radiology 1966; 86:723.

98. SHOPFNER CE, KALMON EH Jr, COIN CG. THE DIAGNOSIS OF HYPERTROPHIC PYLORIC STENOSIS. Am J Roentgenol Radium Ther Nucl Med 1964; 91:796.

99. Liacouras CA, Cook-Sather SD, Schreiner MS, Bellah RD. Endoscopic findings in hypertrophic pyloric stenosis: appearance in classic and evolving disease. Gastrointest Endosc 1997; 45:371.

100. Poon TS, Zhang AL, Cartmill T, Cass DT. Changing patterns of diagnosis and treatment of infantile hypertrophic pyloric stenosis: a clinical audit of 303 patients. J Pediatr Surg 1996; 31:1611.

101. Nakayama DK, Taylor LA. Hypertrophic pyloric stenosis in infancy. An analysis of treatment outcome. N C Med J 1998; 59:310.
102. van den Bunder FAIM, Hall NJ, van Heurn LWE, Derikx JPM. A Delphi Analysis to Reach Consensus on Preoperative Care in Infants with Hypertrophic Pyloric Stenosis. Eur J Pediatr Surg 2020; 30:497.
103. Steven IM, Allen TH, Sweeney DB. Congenital hypertrophic pyloric stenosis: the anaesthetist's view. Anaesth Intensive Care 1973; 1:544.
104. Tigges CR, Bigham MT. Hypertrophic pyloric stenosis: it can take your breath away. Air Med J 2012; 31:45.
105. St Peter SD, Holcomb GW 3rd, Calkins CM, et al. Open versus laparoscopic pyloromyotomy for pyloric stenosis: a prospective, randomized trial. Ann Surg 2006; 244:363.
106. Hall NJ, Pacilli M, Eaton S, et al. Recovery after open versus laparoscopic pyloromyotomy for pyloric stenosis: a double-blind multicentre randomised controlled trial. Lancet 2009; 373:390.
107. Siddiqui S, Heidel RE, Angel CA, Kennedy AP Jr. Pyloromyotomy: randomized control trial of laparoscopic vs open technique. J Pediatr Surg 2012; 47:93.
108. Staerkle RF, Lunger F, Fink L, et al. Open versus laparoscopic pyloromyotomy for pyloric stenosis. Cochrane Database Syst Rev 2021; 3:CD012827.
109. Kim SS, Lau ST, Lee SL, et al. Pyloromyotomy: a comparison of laparoscopic, circumumbilical, and right upper quadrant operative techniques. J Am Coll Surg 2005; 201:66.
110. Sullivan KJ, Chan E, Vincent J, et al. Feeding Post-Pyloromyotomy: A Meta-analysis. Pediatrics 2016; 137.
111. Hulka F, Harrison MW, Campbell TJ, Campbell JR. Complications of pyloromyotomy for infantile hypertrophic pyloric stenosis. Am J Surg 1997; 173:450.
112. Hall NJ, Eaton S, Seims A, et al. Risk of incomplete pyloromyotomy and mucosal perforation in open and laparoscopic pyloromyotomy. J Pediatr Surg 2014; 49:1083.
113. Muramori K, Nagasaki A, Kawanami T. Ultrasonographic serial measurements of the morphologic resolution of the pylorus after Ramstedt pyloromyotomy for infantile hypertrophic pyloric stenosis. J Ultrasound Med 2007; 26:1681.

114. Ogawa Y, Higashimoto Y, Nishijima E, et al. Successful endoscopic balloon dilatation for hypertrophic pyloric stenosis. J Pediatr Surg 1996; 31:1712.

115. Hayashi AH, Giacomantonio JM, Lau HY, Gillis DA. Balloon catheter dilatation for hypertrophic pyloric stenosis. J Pediatr Surg 1990; 25:1119.

116. Yamashiro Y, Mayama H, Yamamoto K, et al. Conservative management of infantile pyloric stenosis by nasoduodenal feeding. Eur J Pediatr 1981; 136:187.

117. Nagita A, Yamaguchi J, Amemoto K, et al. Management and ultrasonographic appearance of infantile hypertrophic pyloric stenosis with intravenous atropine sulfate. J Pediatr Gastroenterol Nutr 1996; 23:172.

118. Theobald I, Rohrschneider WK, Meissner PE, et al. [Hypertrophic pyloric stenosis: sonographic monitoring of conservative therapy with intravenous atropine sulfate]. Ultraschall Med 2000; 21:170.

119. Singh UK, Kumar R, Prasad R. Oral atropine sulfate for infantile hypertrophic pyloric stenosis. Indian Pediatr 2005; 42:473.

120. Kawahara H, Takama Y, Yoshida H, et al. Medical treatment of infantile hypertrophic pyloric stenosis: should we always slice the "olive"? J Pediatr Surg 2005; 40:1848.

121. Lukac M, Antunovic SS, Vujovic D, et al. Is abandonment of nonoperative management of hypertrophic pyloric stenosis warranted? Eur J Pediatr Surg 2013; 23:80.

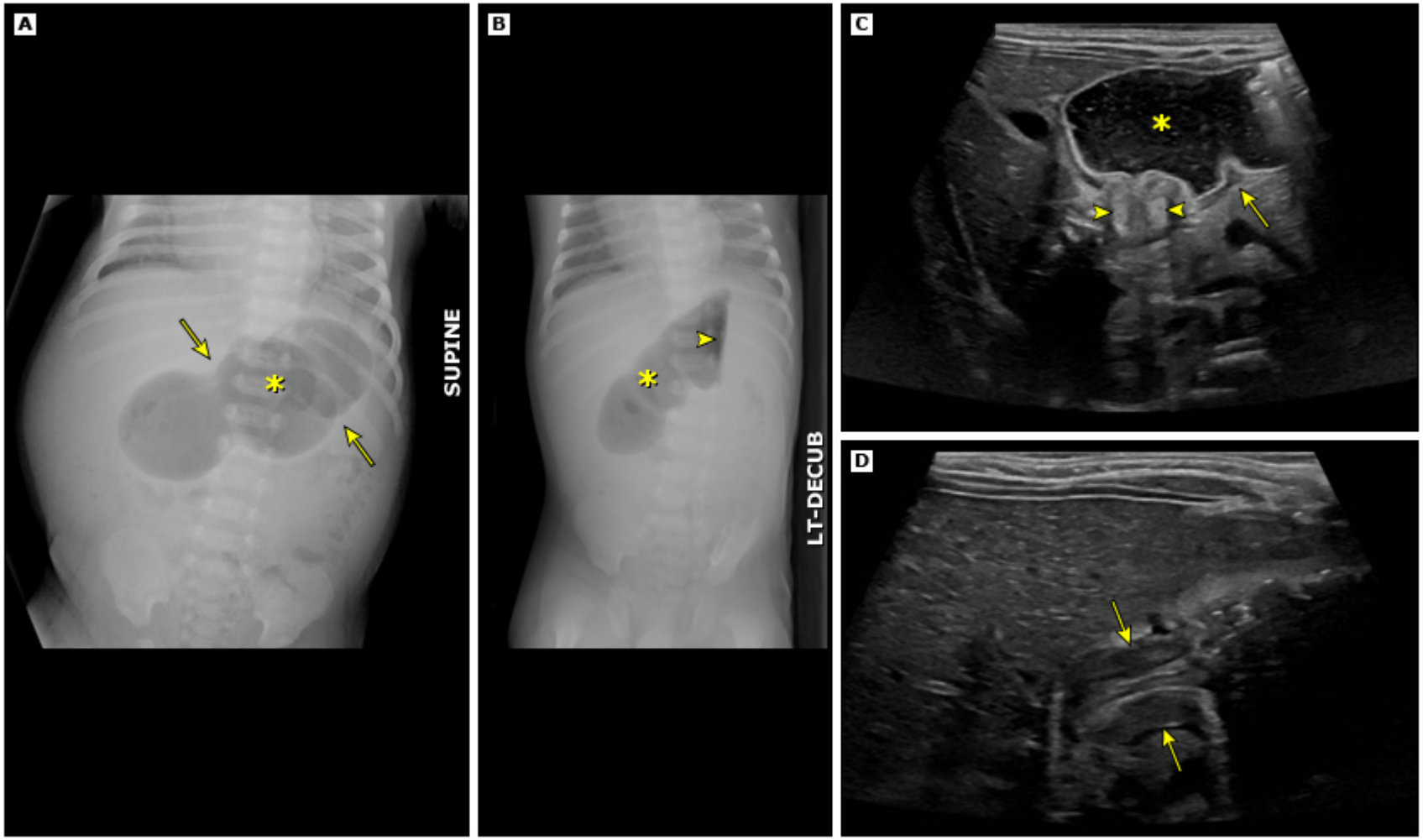
122. Mercer AE, Phillips R. Question 2: can a conservative approach to the treatment of hypertrophic pyloric stenosis with atropine be considered a real alternative to surgical pyloromyotomy? Arch Dis Child 2013; 98:474.

123. Wu SF, Lin HY, Huang FK, et al. Efficacy of Medical Treatment for Infantile Hypertrophic Pyloric Stenosis: A Meta-analysis. Pediatr Neonatol 2016; 57:515.

Topic 5890 Version 38.0

GRAPHICS

Pyloric stenosis on radiograph and ultrasound



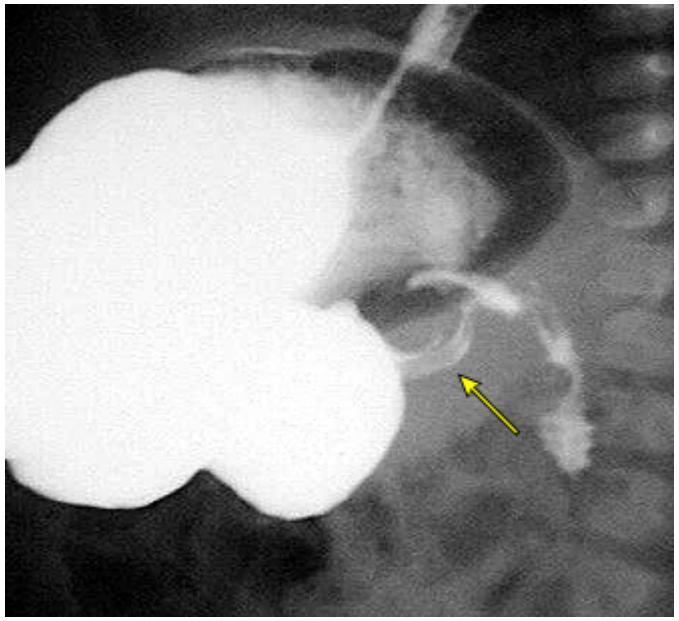
This 4-week-old male infant presented with projectile nonbilious emesis and failure to thrive. Panel A is a supine radiograph and Panel B a lateral decubitus radiograph of the abdomen, revealing a dilated stomach with a single air fluid level (arrowhead) and peristaltic waves (arrows), which constitute the "caterpillar sign" of pyloric stenosis. Panel C is a sonographic image of the distal stomach and pylorus and shows a dilated stomach (asterisk), with peristaltic waves (arrow) and pyloric wall thickening

(arrowheads). Panel D is a sonographic image of the epigastrium and shows wall thickening (arrows) and lengthening. The pyloric muscle thickness measures 5 mm and pyloric muscle length measures 18 mm, consistent with pyloric stenosis.

Courtesy of Joseph Farnam, MD.

Graphic 96412 Version 2.0

Pyloric stenosis



A fluoroscopic upper gastrointestinal study of pyloric stenosis. Note the characteristic "apple core" or "string" sign caused by the narrowed lumen of the pylorus (arrow).

Courtesy of Mary L Brandt, MD.

Graphic 69939 Version 5.0

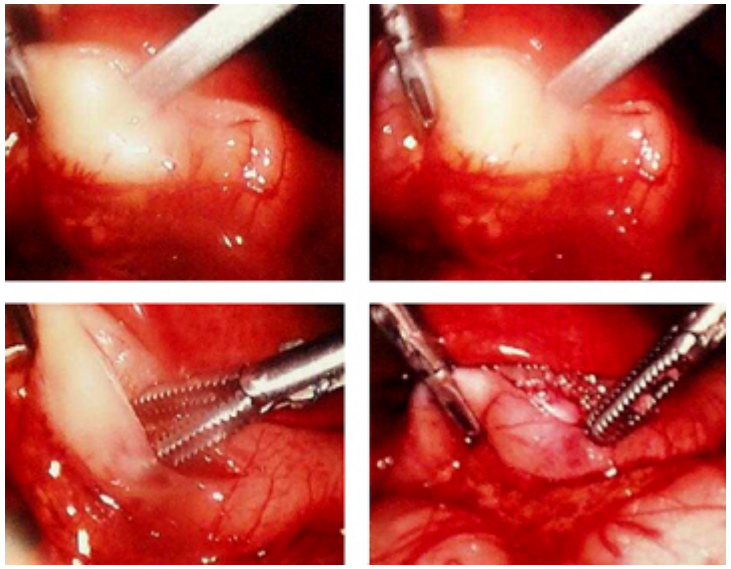
Important causes of vomiting in early infancy

Physiologic reflux or GERD
Pyloric stenosis
Necrotizing enterocolitis
Obstruction
Malrotation with midgut volvulus
Hirschsprung disease
Intussusception
Congenital atresias, stenoses or webs
Gastroenteritis
Metabolic disorders
Feeding intolerance (may be associated with cardiac, pulmonary, renal, or neuromotor disorders)
Adrenal crisis
Dietary protein intolerance (eg, milk protein enteritis)
Urinary tract infection
Toxic ingestion
Increased intracranial pressure (eg, subdural hematoma from child abuse, or hydrocephalus)
Hepatobiliary disease

GERD: gastroesophageal reflux disease.

Graphic 94332 Version 1.0

Laparoscopic pyloromyotomy



Intraoperative photographs of a laparascopic pyloromyotomy for an infant with hypertrophic pyloric stenosis. The pyloric muscle is opened using an arthroscopy blade and then spread open with a grasping forcep.

Courtesy of Mary L Brandt, MD.

Graphic 74120 Version 2.0

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Conflict of interest policy

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