**Background**

[Pyloric stenosis](http://emedicine.medscape.com/article/929829-overview), also known as infantile hypertrophic pyloric stenosis (IHPS), is the most common cause of intestinal obstruction in infancy. IHPS occurs secondary to hypertrophy and hyperplasia of the muscular layers of the pylorus, causing a functional gastric outlet obstruction.

In 1717, Blair first reported autopsy findings of pyloric stenosis. Although the description of the signs and symptoms of infantile hypertrophic pyloric stenosis can be found in the 17th century, the clinical picture and pathology were not accurately described until 1887 by the Danish pediatrician, Hirschsprung. Prior to 1912, early successful surgical procedures included gastroenterostomy, pyloroplasty, and forcible dilatation via gastrostomy. In 1912, Ramstedt observed an uneventful recovery in a patient following pyloroplasty, where sutures used in reapproximating the seromuscular layer had disrupted. Following this observation, he began leaving the split muscle layer unsutured in all subsequent repairs. The Ramstedt pyloromyotomy remains the standard procedure for pyloric stenosis today.

**Pathophysiology**

Marked hypertrophy and hyperplasia of the 2 (circular and longitudinal) muscular layers of the pylorus occurs, leading to narrowing of the gastric antrum. The pyloric canal becomes lengthened, and the whole pylorus becomes thickened. The mucosa is usually edematous and thickened. In advanced cases, the stomach becomes markedly dilated in response to near-complete obstruction.

The causes of infantile hypertrophic pyloric stenosis are multifactorial.[[1]](javascript:showrefcontent('refrenceslayer');) Both environmental factors and hereditary factors are believed to be contributory. Possible etiologic factors include deficiency of nitric oxide synthase containing neurons, abnormal myenteric plexus innervation, infantile hypergastrinemia, and exposure to macrolide antibiotics.

Nitric oxide has been demonstrated as a major inhibitory nonadrenergic, noncholinergic neurotransmitter in the GI tract, causing relaxation of smooth muscle of the myenteric plexus upon its release. Impairment of this neuronal nitric oxide synthase (nNOS) synthesis has been implicated in infantile hypertrophic pyloric stenosis, in addition to achalasia, diabetic gastroparesis, and [Hirschsprung disease](http://emedicine.medscape.com/article/178493-overview).

Rogers has suggested, that persisting duodenal hyperacidity, secondary due to a high parietal cell mass (PCM) and loss of gastrin control, produces pyloric stenosis from repeated pyloric contraction in response to hyperacidity.[[2]](javascript:showrefcontent('refrenceslayer');)

No specific pattern of inheritance exists. It is more common in first-born white males of northern European ancestry and more concordant in monozygotic than dizygotic twins. It also has predominance in children of affected parents (as many as 7%).

A nationwide study of nearly 2 million Danish children born between 1977 and 2008 shows strong evidence for familial aggregation and heritability of pyloric stenosis. Results of the study found a heritability rate of 87% in affected families, lending to the idea that familial aggregation may be explained by shared genes that affect responses to postnatal factors in causing pyloric stenosis.[[3]](javascript:showrefcontent('refrenceslayer');)

**Epidemiology**

**Frequency**

**United States**

The incidence of infantile hypertrophic pyloric stenosis is 2-4 per 1000 live births.

**Mortality/Morbidity**

Death from infantile hypertrophic pyloric stenosis is rare and unexpected. The reported mortality rate is very low and usually results from delays in diagnosis with eventual dehydration and shock.

**Race**

Infantile hypertrophic pyloric stenosis is more common in whites than Hispanics, blacks, or Asians. The incidence is 2.4 per 1000 live births in whites, 1.8 in Hispanics, 0.7 in blacks, and 0.6 in Asians. It is also less common amongst children of mixed race parents.

**Sex**

Infantile hypertrophic pyloric stenosis has a male-to-female predominance of 4:1, with 30% of patients with infantile hypertrophic pyloric stenosis being first-born males.

**Age**

The usual age of presentation is approximately 3 weeks of life (1-18 wk). Approximately 95% of infantile hypertrophic pyloric stenosis cases are diagnosed in those aged 3-12 weeks. Infantile hypertrophic pyloric stenosis is rare in premature infants. In addition, premature infants have a delayed diagnosis secondary to low birth weight and atypical presentation.

**History**

* Classically, the infant with pyloric stenosis has nonbilious vomiting or regurgitation, which may become projectile (in as many as 70% of cases), after which the infant is still hungry.[[4]](javascript:showrefcontent('refrenceslayer');)
* Emesis may be intermittent or occur after each feeding.
* The emesis may become brown or coffee color due to blood secondary to gastritis or a [Mallory-Weiss tear](http://emedicine.medscape.com/article/187134-overview) at the gastroesophageal junction.
* The infant begins to show signs of dehydration and malnutrition, such as poor weight gain, weight loss, marasmus, decreased urinary output, lethargy, and shock.
* The infant may develop jaundice, which is corrected upon correction of the disease.

**Physical**

* In as many as 60-80% of the infants with infantile hypertrophic pyloric stenosis (IHPS), a firm, nontender, and mobile hard pylorus that is 1-2 cm in diameter, described as an "olive," may be present in the right upper quadrant at the lateral edge of the rectus abdominus muscle. This is best palpated after the infant has vomited and when calm, or when the gastric contents have been removed via nasogastric tube.
* Clinicians may also observe gastric peristalsis just prior to emesis as the peristaltic waves try to overcome the obstruction.
* Signs of dehydration include depressed fontanelles, dry mucous membranes, decreased tearing, poor skin turgor, and lethargy.
* The classic signs of infantile hypertrophic pyloric stenosis are becoming less common. The mean age of presentation is getting significantly younger, and infants are not developing the physical signs or electrolyte abnormalities they were 20 years ago. Additionally, the availability of diagnostic imaging is allowing clinicians to make this diagnosis before other clinical manifestations appear.

**Causes**

* The etiology of infantile hypertrophic pyloric stenosis is unknown and is probably multifactorial.

**Prehospital Care**

* As with all pediatric resuscitations, prehospital care in patients with pyloric stenosis should be consistent with pediatric advanced life support (PALS) recommendations for infants who are dehydrated or in shock.
* Immediate treatment requires correction of fluid loss, electrolytes, and acid-base imbalance. Once intravenous access is obtained, the dehydrated infant should receive an initial bolus (20 mL/kg) of crystalloid fluid. The infant should remain nothing by mouth (NPO).

**Emergency Department Care**

* Infantile hypertrophic pyloric stenosis (IHPS) is a medical emergency.
* Immediate treatment requires correction of fluid loss, electrolytes, and acid-base imbalance. Once intravenous access is obtained, an initial fluid bolus (20 mL/kg) of crystalloids should be infused immediately if the infant is dehydrated.
* More than 60% of infants present to the ED with normal electrolyte values or are not in clinical shock. These infants should receive 1.5-2 times maintenance intravenous fluid: 5% dextrose in 0.25% or 0.33% sodium chloride with 2-4 mEq KCl per 100 mL replacement. The infant's fluid status should be continuously reassessed with special attention to acid-base status and urine output.
* The definitive treatment for infantile hypertrophic pyloric stenosis is corrective surgery.
* The Ramstedt pyloromyotomy is the procedure of choice, during which underlying antro-pyloric mass is split leaving the mucosal layer intact.
  + Traditionally, the pyloromyotomy was performed through a right upper quadrant transverse incision. Recent studies have compared the operative time, cost, and hospital stay associated with the traditional incision, a circumbilical incision (believed to have improved cosmesis), and a laparoscopic procedure. The laparoscopic pyloromyotomy has been found to be safe and effective, with shorter operative times and hospital stay.
  + A study from the United Kingdom observed less time to full feedings, less analgesia, less emesis, and faster discharge in the laparoscopic group compared with the traditional approach.[[6]](javascript:showrefcontent('refrenceslayer');)
  + A study from France showed that laparoscopic pyloromyotomy does not decrease the incidence of postoperative vomiting and may lead to a risk of inadequate pyloromyotomy.[[7]](javascript:showrefcontent('refrenceslayer');)
  + Pyloromyotomy performed in specialized centers in pediatric surgery and a general surgery teaching hospital had similar complication rates in a study from the Netherlands.[[8]](javascript:showrefcontent('refrenceslayer');)
  + Recently, various surgical approaches, such as the supraumbilical skin-fold incision and umbilical incision, have been used with easy access, and these approaches have better cosmetic results. Also, a study from Montreal showed superior cosmesis with the supraumbilical (SU) approach than with the right upper quadrant (RUQ) approach.[[9]](javascript:showrefcontent('refrenceslayer');)
* Nonsurgical treatment for infantile hypertrophic pyloric stenosis with atropine sulfate, both intravenous and oral, has shown encouraging results. In one study, infants were given 21 days of atropine via nasogastric tube and regression of pyloric hypertrophy was monitored sonographically. One patient needed intravenous atropine, as nasogastric tube feedings were not tolerated for the first 2 days, but the patient did well subsequently. In this study, all 12 patients were successfully treated nonsurgically without complication.
* Surgical correction is considered the standard of care for all patients with infantile hypertrophic pyloric stenosis; therefore, medical management should be reserved for patients who are poor surgical candidates or whose parents are opposed to surgery.

**Consultations**

* A surgeon comfortable with neonatal care should be consulted as soon as the diagnosis of infantile hypertrophic pyloric stenosis is entertained.

**Medication Summary**

* Surgical correction is considered the standard of care for infantile hypertrophic pyloric stenosis (IHPS).
* Limited data are available for nonsurgical treatment