

## Infantile Hypertrophic Pyloric Stenosis (IHPS) — Causes and Symptoms

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**Pyloric stenosis, also known as infantile hypertrophic pyloric stenosis (IHPS), is a condition that is characterized by pyloric muscle hypertrophy and hyperplasia, which leads to gastric outlet obstruction. Infants usually present in their third week of life with repeated, projectile vomiting, that is often associated with an abdominal olive-like palpable mass. The clinical picture is usually enough to make the diagnosis, but ultrasonography can help confirm the diagnosis of pyloric stenosis in most of the patients. Surgical correction with pyloromyotomy is the treatment of choice.**



### Definition of Pyloric Stenosis

Infantile hypertrophic pyloric stenosis (IHPS) is a condition **caused by a hypertrophy and hyperplasia of the muscularis propria of the pylorus in infants**. The pylorus defines a muscle that opens and closes to allow food to pass through the stomach into the intestine. This hypertrophy/hyperplasia results eventually in **gastric outlet obstruction**.

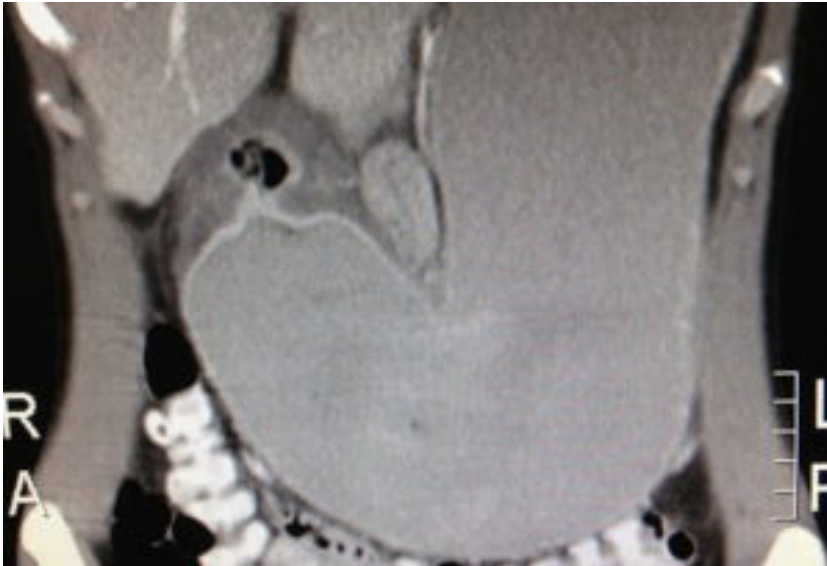


Image: "Pyloric stenosis." by Herecomesdoc - Own work. License: [CC BY-SA 3.0](https://creativecommons.org/licenses/by-sa/3.0/)

## Epidemiology of Pyloric Stenosis

Pediatric pyloric stenosis is a relatively common disorder in the United States with an estimated incidence of up to **4 per 1,000** live births. Fortunately, the condition is not associated with significant mortality.

For poorly understood reasons, pyloric stenosis seems to be slightly more common among whites compared to other ethnic groups. Additionally, pyloric stenosis is **4 times more common in males**, and 30 to 40% more common in first-born males.

Most of the patients are diagnosed at the age of 3 weeks. By 18 weeks, almost all patients with pyloric stenosis would have already presented to the health care system.

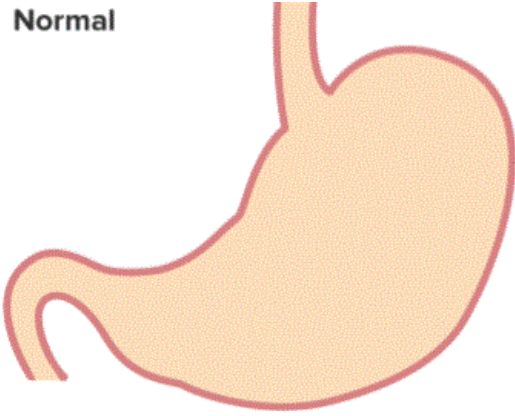
## Etiology of Pyloric Stenosis

The etiology of IHPS seems to be **both genetic and environmental**. For instance, exposure to **bottle-feeding** and **macrolide antibiotics** has been reported to increase the risk. Additionally, infants who have **abnormal mesenteric plexus innervation** or **deficiency of nitric oxide synthase** are at risk of developing IHPS.

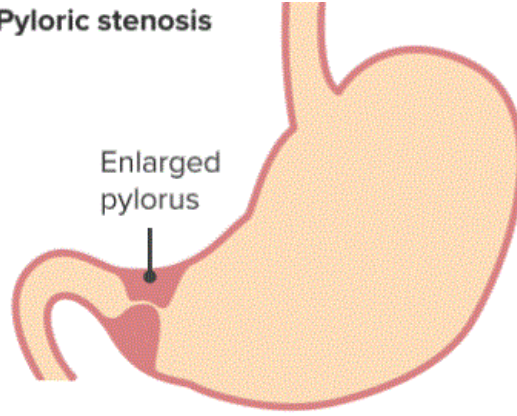
Unclear etiology but an association with:

- Maternal smoking
- Bottle feeding
- Genetic component
- Use of erythromycin/azithromycin

Normal



Pyloric stenosis



"Hypertrophic Pyloric Stenosis" Image created by Lecturio

## Pathophysiology of Pyloric Stenosis

Hypertrophy and hyperplasia of two layers of the pylorus "circular and longitudinal" lead to the development of symptoms. This eventually leads to **obstruction of the gastric outlet** causing **abdominal distension** and if left untreated **gastric dilatation**.

Signs and symptoms of Pediatric Pyloric Stenosis:

- Nonbilious projectile vomiting after 3 weeks of age
- The "happy vomiter"
- May have palpable olive in right upper quadrant or see peristaltic wave



[Image](#): "Pyloric stenosis, due to old callousulcer, showing gastrectasis and large number of peristaltic waves." by Internet Archive Book Images – <https://www.flickr.com/photos/internetarchivebookimages/14570579290/> Source book page: <https://archive.org/stream/americanquarterl04amer/americanquarterl04amer#page/n212/mode/1up>. License: [Flickr Commons](#)

## Causes of pyloric stenosis

The causes of pyloric stenosis are unknown, but environmental and genetic factors might play a role. IHPS usually is not present at birth and probably develops afterward. Several mechanisms and hypotheses have been suggested to explain why hypertrophy/hyperplasia of the pylorus occur in the first place:

- Patients with **family history** of IHPS are at risk of having neurons that are deficient in the enzyme nitric oxide synthase.
- Exposure in early age between 2 to 6 weeks to antibiotics like macrolides and erythromycin increases the risk of pyloric stenosis.
- Low serum lipids also can cause pyloric stenosis
- Persisting duodenal acidity is another associated factor.

**Nitric oxide** plays a vital role as a **neurotransmitter in the gastrointestinal tract**, where it relaxes sphincters by causing smooth muscle relaxation. Patients with nitric oxide synthase deficiency have an increased tone of the lower esophageal sphincter causing **achalasia**, increased tone of the pylorus causing pyloric stenosis and impaired gastric motility which can lead to **gastroparesis**.

There is no single gene that can be directly linked to pediatric pyloric stenosis but familial aggregation is usually evident in most of the cases.

## Clinical Presentation of Pyloric Stenosis

Since the level of the obstruction is at the gastric outlet, patients typically show **non-bilious vomiting** which can be projectile. After vomiting, infants are hungry again. Despite increased feeding, the infant will show **poor weight gain** due to repeated vomiting almost after each feeding.

This presentation is usually evident at 3 weeks of age. If an adequate fluid replacement is not attempted, the infant can develop **dehydration** and lethargy.

Physical examination can reveal an **olive-like, non-tender mass in the upper right abdomen**. Additionally, when gastric obstruction is almost-complete, gastric peristaltic waves can be visible before vomiting. Fortunately, the diagnosis nowadays can be achieved early enough, and these classical symptoms are no longer common.

## Diagnostic Work-up for Pyloric Stenosis

Due to repeated vomiting, patients with IHPS can become dehydrated. **Serum electrolytes, blood-urea-nitrogen, creatinine** and **blood pH** should be checked.

Patients with repeated vomiting due to IHPS lose large amounts of acid. This eventually leads to **metabolic alkalosis**. Loss of fluid also leads to contraction of the extracellular fluid, so the kidneys compensate by activation of the renin-angiotensin system activation. Increased angiotensin II and aldosterone leads to increased potassium secretion (hypokalemia), and further hydrogen secretion and HCO<sub>3</sub><sup>-</sup> reabsorption (more alkalosis). This metabolic process is called contraction alkalosis.

When dehydration is more severe, patients can develop **hyponatremia**, which can have deleterious effects on the brain. Even though these laboratory findings are helpful in the diagnosis of IHPS, they are becoming less common today due to the advances in imaging studies.

Patients with repeated vomiting after each meal, who are between 3 to 12 weeks of age, and have an olive-like mass on abdominal examination do not need any further studies, and the diagnosis of IHPS can be made clinically. When the clinical presentation is less typical, i.e. repeated vomiting without an olive-like mass, **ultrasonography** is the best imaging modality to aid in the diagnosis.

On ultrasonography, **increased pyloric muscles' thickness** is usually the main finding in pyloric stenosis. A pyloric muscle thickness of  $> 4$  mm is diagnostic of pyloric stenosis.



[Image](#): "Endoscopic image of Mallory-Weiss tear." by Samir.  
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Upper gastrointestinal barium studies should be reserved for patients with non-conclusive ultrasonography, and is not routinely used in the evaluation of IHPS. The "double track sign" is defined as the appearance of **two thin barium lines between the hypertrophic layers of the pylorus** and can be found in both pyloric stenosis and pylorospasm.

**Upper gastrointestinal endoscopy** is a last resort in the diagnostic work-up of pyloric stenosis. It can help exclude possible complications such as **Mallory-Weiss tears** due to repeated vomiting and confirm the diagnosis of pyloric stenosis by direct visualization of the gastric antrum and the pylorus.

When USG and UGI studies fail to diagnose clinical signs, endoscopy becomes the last option.

## Treatment of Pyloric Stenosis

It is important to evaluate infants for signs of **dehydration** and to assess the degree of fluid loss. Infants who are in shock due to severe fluid loss should undergo **fluid replacement therapy** according to the pediatric advanced life support guideline.

Accordingly, **correction of electrolytes imbalances, restoration of the normal acid-base balance** and **replacement of fluid** is essential at the emergency level.

The first step in the emergency management of an infant with IHPS is to **infuse a bolus of 20 mL/kg of a crystalloid solution** to correct dehydration.

Infants who are not dehydrated should receive 2 times their maintenance fluid replacement, which should contain either a **5% dextrose or a 0.33% sodium chloride solution** depending on whether the patient is hyponatremic or hypernatremic at

presentation to treat metabolic alkalosis. Patients with pyloric stenosis also develop hypokalemia and they should receive up to **4 mEq KCL per 100 ml fluid**.

Once the infant is hemodynamically stable, the treatment for pyloric stenosis is **corrective surgery**. The currently recommended procedure is a **Ramstedt pyloromyotomy** in which the pylorus and antrum are excised while the muscle layer is left intact. This procedure can also be done **laparoscopically**.

Laparoscopic pyloromyotomy has been found to be safe and as effective as the open approach but with lower morbidity, smaller skin incisions, and faster recovery.

Infants who are poor surgical candidates might benefit from **medical treatment**. Medical treatment consists of **nasogastric administration of atropine** which is believed to result in relaxation of the pylorus muscles and eventually relieve the obstruction after 21 days of daily administration. Atropine administration was evaluated so far in a single study that included only 12 patients and generalization or recommendations cannot be made yet until larger, well-controlled clinical trials can be made.

## Prognosis of Pyloric Stenosis

In patients with delayed or missed diagnosis, there is a risk of dehydration leading to shock. The prognosis for such cases with early diagnosis is good with laparoscopic pyloromyotomy.

## Patients frequently ask

### Is pyloric stenosis in adults possible?

Pyloric stenosis is possible in adults but mostly as a result of other conditions such as ulcers, cancer or abdominal surgery. At times it can be idiopathic although a very rare occurrence but it could occur in middle-aged men.

## References

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[Pediatric Pyloric Stenosis](#) via medscape.com

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