Infantile Hypertrophic Stenosis of the Pylorus: Tips and tricks for Radiology Residents

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Learning objectives

To describe and illustrate the imaging characteristics of infantile hypertrophic stenosis of the pylorus, giving practical dynamic tips not found in textbooks to ensure a rapid and correct diagnosis.

Based on our own clinical experience and an up-to-date review of the literature, we describe and illustrate the anatomy of the antropyloroduodenal region, the normal appearance of the pylorus on imaging studies, the characteristics of hypertrophic stenosis, how to perform the examination, possible complications, and posttreatment changes on imaging studies.

After viewing this educational exhibit, radiologists will be familiar with the imaging characteristics of hypertrophic stenosis as well as with the complications and changes associated with its treatment.

The purpose of this exhibit is to help radiology residents reach a correct diagnosis quickly for the most common cause of nonbilious vomiting in newborns.
Background

Definition and Etiology

Infantile hypertrophic pyloric stenosis, the most common surgical condition producing emesis in infancy, was first described by Hirschsprung in 1888.

The pyloric muscle is hypertrophied and the pyloric channel becomes narrow and elongated, causing gastric outlet obstruction. It typically develops between 2 and 8 weeks after birth. The incidence is approximately 2 to 5 per 1000 live births. It is more common in boys: the ratio of boys to girls affected is 4.8:1.

The cause is unknown, although various genetic factors, environmental factors (e.g., sleeping in the prone position, erythromycin, and maternal smoking, among others), and hormonal factors are thought to play a role in the pathogenesis. It seems that various anomalies are present in the muscular layer, including anomalous distribution of nerve endings, peptide alterations, decreased synthesis of nitric oxide (due to downregulation of the nNOS gene), ultrastructural changes and decreased number of cells of Cajal, and increased production of insulin-like growth factor. Together, these factors are thought to result in failure of the mechanism of muscular relaxation, increasing the production of growth factors and giving rise to hypertrophy.

There is a familial predisposition. About 20% of the sons and 7% of daughters of female index cases develop the disease, whereas 5% of sons and 2% daughters of male index cases are affected.

Pylorus Anatomy

The pyloric portion is the most distal part of the stomach, which is connected to the duodenum through the pyloric sphincter or ring.

The incisura angularis divides the stomach into the gastric body (to the right of this notch) and the pyloric part (to the left). The pyloric part is further divided by the sulcus intermedius into two parts: the pyloric antrum (the most proximal part, which connects to the body of the stomach) and the pyloric canal (the most distal part, which connects to the duodenum). The pyloric antrum terminates in the pyloric orifice, in the pyloric sphincter.

The pyloric sphincter is a zone of intermittent high pressure. It contains a circular layer of smooth muscle that is capable of both tonic and fascicular contraction, enabling gastric
emptying. A definite separation between the pyloric sphincter and the gastric antrum or the head of the duodenum cannot be clearly identified. Unlike most intestinal sphincters, the pyloric sphincter maintains the same lumen (approximately 1 cm wide) most of the time.

**Fig. 1**: Normal anatomy of the antro-pyloric region

**References**: Radiodiagnóstico, UDIAT CD - Sabadell/ES
Practical Analysis

*Let's play a game! Imagine…*

- **On a day like any other day, we are in the US examination room when the emergency department consults us about a baby with suspected infantile hypertrophic stenosis of the pylorus.**

Do you know the characteristics of a child with suspected infantile hypertrophic stenosis of the pylorus?

Patients with suspected infantile hypertrophic stenosis of the pylorus are usually between two and eight weeks old, although this condition has been reported in babies as young as one week and as old as five months. These babies tend to lose weight despite a large appetite.

Typically, they develop nonbilious vomiting after meals, with blood-stained vomit due to gastritis, that increases to become "projectile". If diagnosis and treatment are delayed, hypertrophic stenosis of the pylorus can lead to dehydration, hypochloremic alkalosis, sodium and potassium deficiency, and paroxysmal aciduria.

Is it considered an immediate radiologic emergency?

No, it is considered a relative radiologic emergency. The first and most important thing is to stabilize the patient and correct any electrolyte disturbances. Surgery should be deferred until the infant is appropriately resuscitated.

What do you think is the best imaging technique to establish the diagnosis?

For more than 20 years, high resolution US has been the first choice for diagnosing infantile hypertrophic stenosis of the pylorus: it is noninvasive, fast, widely available, economical, does not require contrast agents, does not use ionizing
radiation, and has nearly 100% sensitivity and specificity. High resolution US enables the direct visualization of the muscle and of the pyloric canal.

Nowadays, barium upper gastrointestinal series are reserved for patients in whom the US findings are inconclusive and those in whom reflux is strongly suspected.

The clinical diagnosis hinges on palpation of the thickened pylorus or "olive". Abdominal palpation is accurate but not always successful, depending on factors such as the experience of the examiner, the presence of gastric distention, and a calm infant.

How should the US be done? Should the stomach be full or empty?

High-resolution linear probes (7-15 MHz) should be used. These probes are capable of identifying all the layers of the pylorus.

The stomach should not be emptied before the examination because this makes identification of the antropyloric area difficult.

If the stomach is filled with gas, placement of the patient in a right anterior oblique position permits fluid to gravitate to the antrum for adequate evaluation. On the other hand, if the stomach is markedly distended, the duodenal cap may be displaced caudally and medially, rendering the pylorus extremely difficult to access. In such cases, if the patient is slowly moved toward the supine and even the left posterior oblique position, the pylorus will be able to rise anteriorly for optimal examination.

Less experienced practitioners should try to locate the gallbladder first. The pylorus is usually located adjacent to the gallbladder (see axial and longitudinal diagrams).

Do you know the US appearance of a normal pylorus?

Although there is no general consensus about the normal measurements of the pyloric canal, the most accepted figures are:

- Thickness of the muscle $\leq 3$ mm, and it is hypoechoic
Anteroposterior pyloric diameter ≤ 14mm
Length of the pyloric canal ≤ 17 mm

A good way for radiologists to remember these parameters is the mnemonic: P < 3.1416 (3 mm muscle thickness, 14 mm AP diameter, and 16 or 17 mm long).

Fig. 2: Normal measures of the pyloric channel

References: Radiodiagnóstico, UDIAT CD - Sabadell/ES
Fig. 3: Longitudinal slice from an abdominal sonogram showing the characteristics of the normal pylorus (a) with correct relations to the sphincter; note the passage of gastric contents through the sphincter (b). Note the correlation with the upper gastrointestinal tract examination (UGI).

References: Radiodiagnóstico, UDIAT CD - Sabadell/ES

Do you know the characteristic findings of infantile hypertrophic stenosis of the pylorus on US?

In hypertrophic stenosis of the pylorus, thickening of the pyloric muscle leads to lengthening of the pyloric canal. On US, the hypertrophy of the pyloric muscle is seen as a heterogeneous hypoechoic ring with a hyperechoic center on transversal slices. The hypoechoic ring corresponds to the thickened muscle and the hyperechoic center corresponds to the redundant edematous surface of the mucosa.

In longitudinal slices, lengthening of the pyloric canal is seen as a characteristic image: the "double-track sign" (Fig. 9). Additionally, the pylorus bends and raises
the duodenal bulb, giving rise to the "umbrella sign". Both these signs correlate well with the images on barium upper gastrointestinal series. The presence of prolapsed pyloric cannel mucosa into the gastric antrum results in "the antral nipple sign".

Fig. 4: Axial diagram showing the anatomic relations of a hypertrophic pyloric stenosis.

References: Radiodiagnóstico, UDIAT CD - Sabadell/ES
Fig. 5: Axial view showing the characteristic sonographic findings for hypertrophic pyloric stenosis.

References: Radiodiagnóstico, UDIAT CD - Sabadell/ES
Fig. 6: Longitudinal diagram showing the anatomic relations of a hypertrophic pyloric stenosis.

References: Radiodiagnóstico, UDIAT CD - Sabadell/ES
Fig. 7: Longitudinal view showing the characteristic sonographic findings for hypertrophic pyloric stenosis.

References: Radiodiagnóstico, UDIAT CD - Sabadell/ES
Fig. 8: Correlation between findings at sonography and UGI for the "double-track sign" (Figs. a & b) and for the "umbrella sign" (Figs. c & d).

References: Radiodiagnóstico, UDIAT CD - Sabadell/ES
Fig. 9: Three-month-old patient with hypertrophic pyloric stenosis. Note the "nipple sign": the presence of pyloric canal mucosa prolapsing into the gastric antrum (arrow).

**References:** Radiodiagnóstico, UDIAT CD - Sabadell/ES

A degree of hyperechogenicity may be seen in the hypertrophied pyloric muscle.
**Fig. 10:** One-month-old patient with a one-week history of nonbilous vomiting. Longitudinal slice shows marked thickening of the muscular layer of the pylorus. Note the slight hyperechogenicity.

**References:** Radiodiagnóstico, UDIAT CD - Sabadell/ES

Infantile hypertrophic stenosis of the pylorus causes exaggerated peristaltic waves in the gastric antrum, a sign of the antrum struggling to overcome the pyloric stenosis.

In some cases, it is possible to see the emptying of gastric contents and air into the duodenum through a hypertrophied pylorus (*abdominal plain films can show proper intestinal pneumatization*).

It is important to observe the failure of the pyloric canal to relax in real time!
Can a child with normal findings on a US examination develop infantile hypertrophic stenosis of the pylorus?

Yes, the clinical syndrome always precedes the imaging findings; thus, when US rules out infantile hypertrophic stenosis of the pylorus, we should recommend clinical follow-up and repeating the US study within a few days.
Ten-day-old patient who presented with an episode of nonbilious vomiting. Sonogram shows a normal-appearing pylorus (Figs. a & b). Repeat sonogram 12 days later done for persistence of symptoms shows a clearly hypertrophic pylorus (Figs. c & d).

References: Radiodiagnóstico, UDIAT CD, Sabadell/ES

Great! We have successfully diagnosed infantile hypertrophic stenosis of the pylorus! Now we need to contact the surgical team so they can operate on the child.

Do you know what the treatment consists of?

The current treatment of choice is pyloroplasty (Fredet-Weber-Ramsted extramucosal pyloromyotomy). Before the operation, electrolyte disturbances must be corrected. Stabilizing the child can delay the intervention by 24 h to 48 h. Like all surgical techniques, complications can occur. Medical treatment with antispasmodics (atropine) was used until the mid-1960s and is currently be reappraised by various investigators.
• After the intervention, the child was discharged; however, one week later he presented at the emergency department with persistent vomiting after meals.

What diagnostic possibilities should you consider?

If done correctly, the surgical intervention solves the problem, and no cases of relapse have been reported. The characteristic US findings can persist for six to eight weeks after surgery, which is the time necessary for the hypertrophied pyloric muscle to return to its normal size and appearance. We should not consider these findings to represent failed surgery! In this case, the cause of our patient's vomiting was gastroenteritis.

Fig. 13: Sonogram in a patient who presented a new episode of vomiting 15 days after intervention for hypertrophic pyloric stenosis shows that the hypertrophy of the pylorus persists.

References: Radiodiagnóstico, UDIAT CD - Sabadell/ES
Conclusion

Take-home points

• In a baby who presents with vomiting between the ages of two and eight weeks, we should consider infantile hypertrophic stenosis of the pylorus the first diagnostic option.
• "#ylorus": mnemonic for the measurements of the normal pylorus.
• Normal US findings do not rule out infantile hypertrophic stenosis of the pylorus: if the symptoms persist or worsen (projectile vomiting), it is necessary to repeat the study.
• Not all cases of infantile hypertrophic stenosis of the pylorus result in total obstruction—in some cases gastric content can be observed to pass through the pyloric canal.
• After surgery, the US findings do not return to normal for six to eight weeks.
References


