Learning objectives

- Describe the anatomy of the antropyloroduodenal region

- Explain how to perform the ultrasound and contrast studies examinations

- Describe and illustrate the imaging findings in the Hypertrophic Pyloric Stenosis in ultrasound and in contrast examinations

- Describe the major pitfalls in the ultrasound diagnosis of the Hypertrophic Pyloric Stenosis

- Review the main differential diagnosis of this pathology

Background

- Hypertrophic Pyloric Stenosis (HPS) was described for the first time in 1717, by Blair(1);

- HPS implies muscle hypertrophy and hyperplasia primarily involving the circular layer and hypertrophy of the underlying mucosa, resulting in partial or complete obstruction of the pyloric channel (1);

- The Infantile form of HPS is the most frequent cause of gastric outlet obstruction in infants(1);

- The prevalence of HPS is higher in caucasian infants than in African-American and Asian children(1);

- The prevalence of the Infantile form of HPS rounds 1.5 to 4.0 per 1000 live(1);

- HPS is more common in boys than girls (2:1 to 5:1)(1);

- The clinical presentation of HPS is based on the onset of nonbilius projectile vomiting(1);
- The peak of initial presentation occurs at 4th week of life(2);

- 95% of cases of HPS appear between the 3rd and 12th weeks of life(2);

- Unusually the diagnosis is made in patients less than 10 days old(2);

- The aetiology of HPS is not understood, but both genetic and environmental factors seem to play a role in the pathophysiology(1);

- The diagnosis of HPS can be made when an olive-shaped mass is palpated in the epigastrium, representing the thickened pylorus(2);

- The US examination is the first line method for the diagnostic of HPS(2);

- The sensitivity and specificity of US rounds the 100%(2);

- HPS is not a surgical emergency (2).

**Findings and procedure details**

- **The anatomy of the normal antropyloroduodenal region**

- The stomach is divided in two parts, by the *incisura angularis*: the body, on left, and the pyloric portion, on the right(3);

- The pyloric region is divided in two parts, by the *sulcus intermedius*: the pyloric vestibule on the left, and the pyloric antrum or pyloric canal on the right(3);

- The pylorus is situated at the outlet of the stomach - regulating gastric emptying - and is structure continuous with the longitudinal smooth muscle layer between the antrum and duodenum(4);

- The pyloric antrum has 2.5 cm length and ends at the pylori orifice(3);
During normal gastric emptying, the pylorus opens, allowing passage of luminal content to distend the duodenum and duodenal motor activity is stimulated(4);

• **The anatomy of the antropyloroduodenal region in patients with HPS**

- The pyloric ring is transformed into a channel with variable length (1.5-2.0 cm), which separates the normally distensible portion of the antrum from the duodenal cap(3);

- The pyloric canal has a thickened muscle: diameter of the canal lumen is equal or greater than 6 mm (3);

- The obstructed lumen is filled compressed and redundant mucosa creating an obstructed passage to the gastric contents (3).

• **Ultrasound (US) Evaluation - Technique**

- Use a high-resolution linear-array transducer (5-12 MHz) (2,5);

- First position of the linear-array transducer: transverse oblique plane parallel to the right lower costal margin (2);

  *Use the liver as an acoustic window, in order to eliminate shadowing caused by gas distal to the stomach (2)*;

- Second position of the linear-array transducer: relative cephalad positioning of the probe with slight downward angulation (2);

  *The gallbladder is the landmark, once it is just lateral to the pylorus (2,5).*

- Third position of the linear-array transducer: right posterior oblique positioning of the infant, to allow gastric liquid to distend the antropyloric region OR right anterior oblique position to allow liquid to gravitate to the antropyloric region, when the stomach is largely gas filled(2);
The stomach should be filled with liquid to allow improved visualization of the pylorus (2,5);

- The radiologist should search for the normal triangular duodenal bulb, distal to the gastric antrum (2).

NORMAL PYLORUS

- The liquid passes quickly from the antrum into the duodenal bulb (2,5);

- Bulls-eye pattern (in cross section view) (5):
  - the outer anechoic rim - represents the normal circular muscle;
  - the inner echogenic layer - represents the mucosa and submucosa;
  - the inner most anechoic center - represents fluid in the pyloric canal.

- In the longitudinal plane the radiologist can demonstrate the same anatomic features (Figure 1) (5).
**Fig. 1**: Ultrasound image showing the outer anechoic rim (which represents the normal circular muscle) (red arrow), the inner echogenic layer (which represents the mucosa and submucosa) (green arrow) and the inner most anechoic center (which represents fluid in the pyloric canal) (yellow arrow).

**References**: - Viseu/PT

- **ABNORMAL PYLOROS - HPS**

- The key for establishing the diagnosis is the persistent abnormal thickening of the pyloric muscle (2,5);

- The stomach can remain distended several hours after the last feeding, because of the gastric outlet obstruction (2);

- The "cervix" sign: appearance of HPS on long-axis US images, resembling the uterine cervix (2) (Figure 2);
Fig. 2: The ultrasound imaging showing the "cervix" sign.

References: - Viseu/PT

- The "donut" lesion: represents the sonolucent thickened muscle, located medial to the gallbladder and anterior to the kidney (Figure 3) (5);
Fig. 3: The ultrasound imaging showing the "donut" lesion.

References: - Viseu/PT

- On longitudinal section it is possible to show the continuity between the anechoic rim of thick muscle and the thin muscle of the gastric antrum (5);

The morphology of the channel and real-time observations are more important than numeric value (2).

- The standard measurements are performed on long-axis view:

1. measures are taken at the superficial and deep borders of a single layer of the hypoechoic pyloric muscle - the threshold value for a diagnosis of HPS is generally greater than 3-4 mm (2);
2. abnormal length measure ranges between 14 - 20 mm. This measure is useful for the performing the pyloromyotomy and in cases in which muscle thickness is borderline(2) (Figure 4);
Fig. 4: The ultrasound imaging showing the increase of the longitudinal length measure (16.9 mm) and an increase in the thickness of the pyloric muscular wall (11.9 mm).

References: - Viseu/PT

PITFALLS IN THE ULTRASONIC RECOGNITION OF NORMAL PYLORUS

-The muscle hypertrophy needs to present in both the cross sectional and longitudinal planes, to avoid diagnostic errors(5);

-The evaluation of the muscle thickness must be made truly perpendicular to the axis of the pylorus or aligned with its axis, with cross sectional sonograms or with longitudinal sonograms, respectively. When the sections are not properly oriented may suggest muscle thickening, leading to an erroneous diagnostic (5);
- During the examination the fluid should be seen passing through the canal, once the normal collapsed pylorus can produce a false appearance of muscle thickening-"pseudothickening" (5);

- Gastric overdistention condicionates the appearance of the most common pitfall, once this situation leads to false-negative result, because it displaces the antrum and pylorus posteriorly. To avoid this situations the transducer should be placed more laterally toward the right flank to better visualization of the displaced pylorus (2);

- The Upper gastrointestinal Examination (UGI)

- When is necessary to realize UGI in a suspect of HPS ?

- This examination should be realized in a patient with persistent unexplained emesis, in order to search for a hiatal hernia, an antral web, or a duodenal obstruction (2);

- How to realize UGI?

- The patient first drinks the barium meal. Insertion of a nasogastric tube is not necessary (3);

- The patient should be positioned in the right anterior oblique position, to facilitate gastric emptying (3);

- Intermittent fluoroscopic observations should be done (3).

- What to expect in a normal UGI?

- The intermittent fluoroscopic should be done, since peristalsis may transiently simulate an elongated and abnormal canal (3);

- A thin pyloric ring can be the imagiologic manifestation of the bridge between the prepyloric antrum and the dudenal cap (3);

- The prepyloric antrum is widely distensible between normal peristaltic waves (3).
- What alterations to expect in the UGI, in patients with HPS?

- In HPS the pyloric channel appears "elongated", because there is a failure in the relaxation of the pre-pyloric antrum(3);

- The *string sign* - appears when the channel is outlined by a string of contrast material (3);

- The *double-track sign* - appears when several linear tracts of contrast material are separated by the intervening mucosa. (Figure 5)(3);

- The *tit sign* - appears when the luminal barium is transiently trapped between the peristaltic wave and the muscle (3) (Figure 6);

- The HPS diagnosis can be inferred when the pyloric opening time is delayed longer than five minutes. The opening time is found to be between fifteen and sixty minutes, in the majority of the HPS cases(6).

Fig. 5: Double-track sign - Contrast studies showing redundant mucosa separates barium into two columns in pyloric channel.

*References:* - Viseu/PT
**Fig. 6**: Tit sign - contrast study showing an outpouching created on lesser curve by distorted muscle in hypertrophic pyloric stenosis.

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**Table 1**: Generic comparison between UGI and Ultrasound in the diagnosis of HPS (3).

*References:* - Viseu/PT

**LIST OF THE DIFFERENTIAL DIAGNOSIS:**

- Gastroenteritis(1);
- Food allergy(1);
- Gastroesophageal reflux(1);
- Pylorospasm(1);
- Antral web(1);
- Pyloric duplication(1);
- Ectopic pancreatic tissue in the wall of the pylorus (1);
- Hiatal hernia(3);
Preampullary duodenal stenosis(3).

Treatment

- The Ramstedt Pyloromyotomy is the operative procedure of choice is (1,3), whether performed via abdominal incision or at laparoscopy. This procedure leaves the intact mucosa bulging through the incision by the division of the hypertrophied muscle(3). This procedure is associated with remarkably low morbidity (1).

- Nowadays, HPS is not a life-threatening disease and has a mortality rate that is nearly 0% (3).

Images for this section:
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Conclusion

This review of the anatomy, diagnostic methods, imaging findings and main differential diagnosis of hypertrophic pyloric stenosis provides greater confidence of the radiologist in making this diagnose.

Personal information


References