

Diagnostic imaging of hypertrophic pyloric stenosis (HPS)

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Background. Imaging of the abdomen in children with suspected hypertrophic pyloric stenosis has been traditionally performed by plain film radiography and upper gastrointestinal contrast studies. In many clinical situations, this approach has been modified or replaced by ultrasound examination. The authors aimed to analyse the value of diagnostic algorithm in children with hypertrophic pyloric stenosis confirmed at surgery in our hospital.

Patients and methods. The authors made a five year retrospective review of hospital records of all children operated on for HPS in Clinical Hospital Centre Zagreb - Rebro and found out that 14 boys, between 2 (17 days) and 10 weeks of life (75 days) underwent surgery due to HPS.

Results. Specific radiographic signs were: string sign, double track sign, elongation and narrowing of pyloric canal, mushroom sign, gastric distension with fluid and beak sign. Ultrasound was performed in 9 patients, one of them was false negative (sonographer admitted that he had no experience), the rest were positive.

Conclusions. If the physical examination is negative or equivocal, sonography by an experienced sonographer must be performed. If the ultrasound finding is negative, than the infant should undergo to barium upper gastrointestinal studies (UGI). If HPS isn't a primary diagnostic question, it's better to perform UGI first in order to make a correct diagnosis.

Key words: pyloric stenosis - radiography - surgery; hyperthrophy; child

Introduction

Hypertrophic pyloric stenosis (HPS) is actually idiopathic hypertrophy and hyperplasia of the circular muscle fibers of the pylorus with proximal extension into the gastric antrum.¹ The cause of HPS remains unknown. HPS is inherited as a dominant polygenic trait. Some authors reported even familial occurrence of HPS in twins.² This, however, was rather an acquired than congenital condition.¹ Others presented an example of "secondary" HPS in a patient with

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prostaglandin - induced foveolar hyperplasia of antrum.³

Recently, it has assumed that, in some cases, HPS is caused by *Helicobacter pylori*.⁴ The latest research supports the hypothesis of a selective immaturity of the enteric glia in the muscular layers of infantile HPS.⁵

Hypertrophic pyloric stenosis is the most common acquired obstruction of the young infant. It is more common in boys than girls, by a 5:1 ratio and develops usually between the second and eighth weeks of life.

The clinical features of bile-free progressive projectile vomiting, visible gastric peristaltic waves, and an olive shaped palpable abdominal mass in the right upper quadrant are frequently diagnostic. Depending on how long symptoms have been present, little patients may present with dehydration and hypokalemic alkalosis, irritability, weight loss, and failure to thrive.

Plain film radiography has no role in the diagnosis of pyloric stenosis. Massive gastric distension (>7 cm diameter) is seen commonly in other conditions and is not at all specific. If the child vomits before the filming, the gastric distension may be relieved.

For *barium upper gastrointestinal studies* (UGI), we must empty gastric contents via nasogastric tube before and after the study due to a high incidence of reflux in these patients. Positive fluoroscopic and radiographic signs include elongated pyloric canal (string sign), antral beaking, pyloric teat, flattening of the prepyloric area of the lesser curvature (shoulder sign), and usually active gastric hyperperistalsis (caterpillar sign). Sometimes a double or triple column of barium is present as two or three parallel lines (double/triple track sign) caused by the crowding of mucosal folds in the pyloric canal. The base of the bulb can be indented by thickened shoulder of pyloric muscle (mushroom sign). Delayed gastric emptying is the least reliable indicator of HPS and can be seen with pylorospasm, gastric hypotonia, sepsis and ileus.

The *ultrasound (US) examination* is performed with the patient in the supine, and later, in the right lateral decubitus position. Overlying bowel gas or gastric distension may occasionally hinder the sonographic diagnosis of HPS. To resolve this problem, a novel approach for obtaining posterior views of the pylorus was reported.⁶

Ultrasound examination of the pyloric region includes both transverse and longitudinal images of the pylorus. The most common measurement used is *pyloric muscle thickness* obtained with transverse scanning of the pylorus. The muscle is usually hypoechoic, but it can have a nonuniform pattern.⁷ The muscle appeared to be more echogenic in its near and far fields and less echogenic on its sides due to anisotropic effect which is related to the orientation of the ultrasound beam with respect to the circular fibers of the pyloric muscle. The *transverse pyloric diameter*, including the lumen and both walls of the pylorus, is less frequently measured. The *pyloric canal length* (echogenic) may be measured, and is shorter than the surrounding *pyloric muscle length* (hypoechoic structures). Several different pyloric muscle indices also have been used to detect HPS.⁸⁻¹⁰

There has been disagreement as to the exact measurements to be used for pyloric stenosis. Authors have published different numbers for these different measurements.¹¹⁻¹³

Dähnert¹ suggested, that pyloric muscle thickness ≥ 3 mm, transverse pyloric diameter ≥ 13 mm with pyloric canal closed and pyloric canal length ≥ 17 mm are diagnostic of HPS.

Other sonographic signs are: "target sign" (hypoechoic ring of the hypertrophied pyloric muscle around echogenic mucosa centrally on cross-section), "cervix sign" (indentation of the muscle mass on the fluid-filled antrum on longitudinal section), "antral nipple sign"¹⁴ (redundant pyloric canal mucosa protruding into the gastric antrum), exaggerated retrograde peristaltic waves and

delayed gastric emptying of fluid into the duodenum.

The authors aimed to analyse the value of diagnostic algorithm in children with hypertrophic pyloric stenosis confirmed at surgery in Clinical Hospital Rebro.

Patients and method

This is a retrospective review of hospital records of all children operated on for pyloric stenosis in Clinical Hospital Centre Zagreb - Rebro from 1st January 1995 to 31st December 1999. Fourteen infants underwent surgery due to hypertrophic pyloric stenosis during the period in question. They were all boys, between 2 (17 days) and 10 weeks of life (75 days).

UGI study was performed with 5-10 ml of diluted barium on Siemens Sireskop 3. Sonographic examination was performed in the standard supine and right lateral decubitus position, using a GE Logiq 400 scanner and 5.0-MHz convex traducer and 6.6-MHz linear traducer.

Results

Clinical findings in our patients are presented in Table 1.

In all infants, the radiological diagnosis was made on the basis of upper gastrointestinal series (Table 2).

Specific radiographic signs were: string sign, double track sign, elongation and narrowing of pyloric canal, mushroom sign, gas-

Table 1. Clinical symptoms and laboratory data

Clinical symptoms and laboratory data	No.
Bile-free projectile vomiting	14
Olive shaped muscular mass	2
Dehydration and hypokalemic alkalosis	9
Weight loss, failure to thrive	3

Table 2. Methods of imaging

No.	UGI	US
14	14 (100%)	9 (64%)

UGI= barium upper gastrointestinal studies; US= ultrasound examination

tric distension with fluid and beak sign. Ultrasound was performed in 9 patients, one of them was false negative (the sonographer admitted that he has no experience), the rest were positive. Ultrasound signs and measurements were: target sign, transverse pyloric diameter, pyloric muscle wall thickness and pyloric canal length. All measurements were consistent with the diagnosis of HPS.

Discussion

Nowadays, the reliance on diagnostic imaging has been increasing.¹⁵ During palpation performed by paediatrician or surgeon the infant must be calm; this is time consuming, and may even be impossible, if the stomach is distended. Some authors stated that the technique of palpating a pyloric mass became "a declining art".¹⁶ Many publications on this subject stressed that the diagnosis often can be made by physical examination and that imaging procedures don't need to be routinely performed.¹⁷⁻²⁰ Only children with a negative or equivocal physical examination should go to ultrasonography. Currently, ultrasonography has replaced the upper gastrointestinal (UGI) examination as the method of choice for establishing the diagnosis.²¹⁻²³ US is more economical, there is no exposure to ionising radiation such as in UGI studies, and allows to follow up the patients, but it demands a highly experienced sonographer. It has also been reported that over-reliance on ultrasound scans only lead to negative explorations.²⁴

There are also other opinions: that the UGI is less expensive than the US as the first strategy in the evaluation of the infant with sus-

pected HPS.^{25, 26} An advantage of the UGI is that it has slightly higher sensitivity for pyloric stenosis than does US scan. UGI also provides definitive information in the evaluation of the vomiting infant regarding other potential diagnoses such as gastroesophageal reflux, malrotation and intestinal obstruction. If the clinical findings are doubtful, it is justified to perform UGI because of concomitant pathology. One of the papers presented the cases of pyloric stenosis associated with malrotation.²⁷

In a recent publication, reporting of the attempts to develop a cost- and time-effective algorithm for differentiating HPS from other medical causes of emesis in infants, it is recommended that the child is given nothing by mouth for 3 to 4 hours before gastric aspiration. The aspirated volume ≥ 5 ml implicated gastric outlet obstruction and ultrasonography was performed. If this examination was positive for HPS, the child was referred for surgery. If US was negative, upper gastrointestinal series were performed. The aspirated stomach contents volume < 5 ml suggested another medical cause of emesis; therefore UGI was performed.^{28, 29}

In our hospital UGI was performed always on surgeon's request, even clinical and US findings were positive. Surgeon's trust and confidence in UGI versus US is changing very. They sometimes neglect the ionising radiation during UGI studies. On the other hand, because US is very operator -dependent imaging modality, false positive and false negative results can compromise this method. This is no wonder because HPS is rare pathology. An additional problem is that, in large centres like our hospital, we have no department of paediatric radiology.

Differential diagnosis of HPS after the imaging includes infantile pylorospasm in which the muscle thickness is between 1.5 and 3 mm. In this condition, antral narrowing is of variable calibre, gastric emptying is delayed, the pylorus is elongated, antral peristalsis is

functioning. Muscle thickness or pyloric length measurements may overlap those accepted as positive for HPS. Image or measurement variability is an important clue for diagnosing pylorospasm.³⁰ Milk allergy and eosinophilic gastroenteritis can also mimic the clinical symptoms and US appearance of idiopathic HPS.³¹ Eosinophilic gastro-enteritis is characterised by hypertrophy of the hypoechoic muscular layer and also thickening of the mucosal and submucosal layers of the pylorus. It is also helpful to search for thickening of the antral wall. The differential diagnosis for possible HPS encompasses several other gastrointestinal tract abnormalities, including gastroesophageal reflux, duodenal obstruction, and pyloric membrane, or webs. After the imaging we didn't have any differential diagnostic difficulties.

Treatment is surgical (pyloromyotomy). We follow up the operated children with US. Recently, some attempts have been made in the treatment with atropine sulfate; all infants were followed by sonography to observe the anatomical changes (shortening of the pyloric canal, followed by thinning of the muscular layer).^{32, 33} We have no experience in such treatment. Our review of the literature suggests that this kind of treatment hasn't found general clinical acceptance.

The infant with symptoms that clearly suggests pyloric stenosis must be examined by an experienced physician prior to imaging. If the physical examination is negative or equivocal, sonography by an experienced sonographer must be performed.

If the US is negative, than the infant should go to UGI.

If HPS isn't primary diagnostic question, it's better to perform UGI first to establish the correct diagnosis.

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