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REVIEW

Pyloric stenosis: role of imaging

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Introduction

Pyloric stenosis is an acquired condition, which typically develops between 2–12 weeks of postnatal life. The infants present with nonbilious vomiting, and presentation overlaps with other causes of vomiting, particularly reflux. If unrecognized or untreated, the condition leads to worsening and protracted vomiting, typically described as "projectile", with supervening dehydration, hypochloremic alkalosis secondary to loss of electrolytes, paradoxical aciduria in an effort to conserve sodium, and, if untreated, eventually culminates in emaciation and death. Although unrecognized and often fatal for centuries, in the twentieth century great strides were made in the recognition and surgical treatment of pyloric stenosis, a condition which has now become routinely familiar to all pediatric radiologists, pediatric surgeons and pediatricians, with uniformly excellent outcome.

The epidemiology of pyloric stenosis is dependent on racial and geographic extraction, with a likely polygenic influence. Among white populations of northern European descent, the incidence of pyloric stenosis is approximately 2–5/1,000 live births. This incidence decreases by 20–30% among Caucasians in India, and even further among Black and Asian populations (0.7/1,000 live births) [1]. The proband concordance between monozygotic twins is 0.25–0.44, and that between dizygotic twins falls to 0.05–0.10 [2]. There is a greater than five-fold increase in incidence among first-degree relatives [1]. Regardless of underlying cause, the phenotype is most often present in males. Affected mothers have a likelihood of pyloric stenosis in 20% of their sons and 7% of their daughters, while affected fathers have a likelihood of pyloric stenosis in 5% of their sons, and 2.5% of their daughters, with a boy:girl ratio at presentation cited between 2.5–5.5:1 [2].

Pathophysiology

The cause and pathophysiology of pyloric stenosis remain elusive, despite the fact that the condition is common, and despite, or perhaps because of, the success of modern surgical management. Current investigations have focused on studies of the hypertrophied musculature, and multiple abnormalities have been identified. These include innervation abnormalities, e.g., deficiency in interstitial cells of Cajal [3, 4], in the quantity of nerve terminals [5] and of markers for nerve-supporting cells [6], in peptide-containing fibers [7, 8], and in nitric oxide synthase activity [9] and messenger RNA production [10]. The muscle itself has been found to have increased expression of insulin-like growth factor-I messenger RNA [11], and increased platelet-derived and insulin-like

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growth factors [12]. It is therefore hypothesized that there is failure of muscle relaxation, with increased synthesis of growth factors, leading to muscle hyperplasia, hypertrophy, and obstruction. These findings, although impressive, fail to explain the development of the condition after the postnatal inception of feedings, fail to take into account the hyperemia and marked hypertrophy of the mucosa filling and obstructing the antropyloric channel [13, 14], and the fact that, as early as 4 months after surgical release of obstruction, assay results for nerve growth factor, interstitial cells of Cajal, and nitric oxide synthase activity have returned to normal [15].

Another hypothesis [16, 17] suggests that infants who develop pyloric stenosis are born with an increased parietal cell mass, which leads to increased secretion of gastrin. Gastrin has a trophic effect on gastric mucosa, stimulating DNA, RNA and protein synthesis, increasing the number of parietal cells, and is a major promoter of acid secretion [18]. Infants in the first 2 months of life have been found to have increased serum gastrin levels, with poor or no feedback mechanism and response to feedings [19]. Thus, infants born with a high parietal cell mass are hypothesized to have higher acid production, antral contractions, and mucosal hypertrophy which initiates a cycle of repeated pyloric contraction and delayed gastric emptying, likely mediated by the hyperemic changes seen on Doppler [14], culminating in the findings of mucosal and muscular hypertrophy and gastric outlet obstruction.

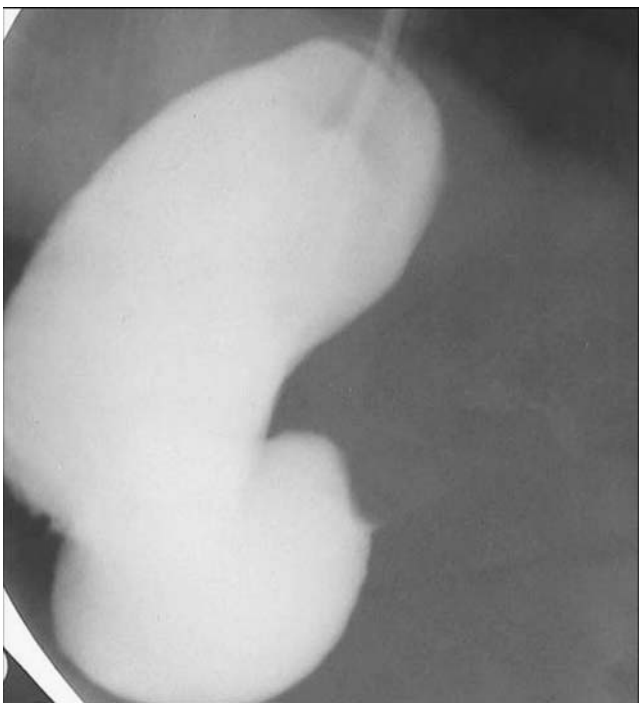

Fig. 1 Spot image from an upper GI, outlining gastric peristalsis propelling contrast through the narrowed antropyloric canal into the base of the duodenal cap. Note orogastric tube initially used to empty the stomach prior to introduction of contrast material. Mean sensitivity and specificity of this examination is quoted as approximately 95% and 98.5%, respectively [20, 28, 32]

Overall cost to society

The costs of diagnosis and treatment of pyloric stenosis vary with the decision tree for diagnosis, the type of surgery performed, the skill of the physicians involved, and the rate of complications.

In a retrospective study of 234 infants, 150 with pyloric stenosis [20], the total charges for those with pyloric stenosis were $2,454.00, with a potential savings of $100.00 per patient, if diagnostic imaging was only performed after abdominal palpation, and if the sensitivity of this examination were at least 38%, assuming no further imaging is done if olive is not palpated.

A retrospective study of 780 patients [21] found that the mean hospital charges for patients treated by general surgeons were $5,121.00, whereas those for patients treated by pediatric surgeons were $4,496.00, largely due to a difference in the frequency of complications.

In a prospective study of 116 infants in the UK [22], clinical examination had a sensitivity of 72% and specificity of 97%; US had sensitivity of 97% and specificity of 100%. Use of US initiated a change in management in the clinically false negative group at a cost of 3% of total, and obviated surgery in the clinically false positive patient, at a savings of £1,602.

A hypothetical decision tree study in which the upper GI was assigned sensitivity and specificity of 100%, and US 90% and 100% respectively, concluded that the upper GI is the most cost-effective initial study in the evaluation of the vomiting infant [23], using mean charges derived from 24 institutions. Not surprisingly, as the two sensitivities became equal, the cost ratio of US/upper GI became one. Assuming a median incidence of pyloric stenosis of 0.30, and that all patients with negative US would go on to upper GI, the authors found that an US followed by upper GI strategy would only be cost-effective if the cost of US were 0.66 of that for upper GI. If imaging were done after a negative abdominal palpation, and the incidence of patients with pyloric stenosis decreased to 0.02–0.18, the cost of US would need to be less than 0.50 for the US strategy to be cost-effective. Some of the assumptions in this study have been challenged [24], particularly the pre-test probability, the assumptions of the sensitivity and specificity of the US and upper GI examinations, and that every patient with negative US would necessarily undergo upper GI.

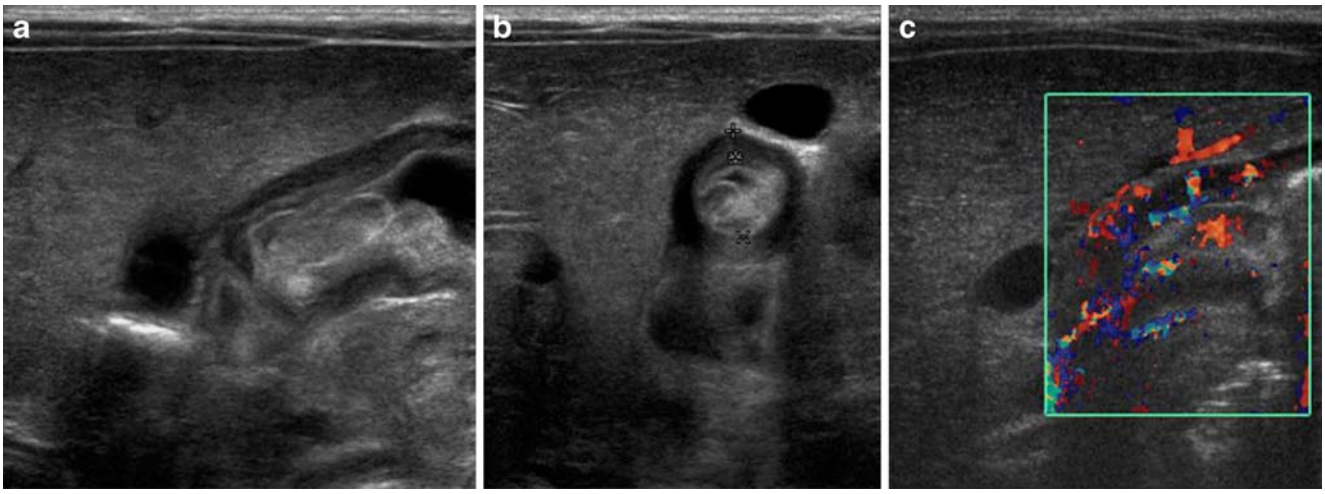

Fig. 2 Longitudinal (**a**) transverse (**b**) and Doppler (**c**) US images through the abnormal pylorus, outlining the increased vascularity to the thickened muscle measuring approximately 3.6 mm, and to the thickened mucosa, which fills and obstructs the 10.7 mm wide antropyloric channel. This degree of mucosal thickening is uncommon. Mean sensitivity and specificity of this examination, in experienced hands, is quoted as 99.3% and 99.8%, respectively [20, 22, 29, 30, 34–36]

Diagnostic examinations

The ideal diagnostic examination should be accurate, noninvasive, and rapid. There is controversy regarding most efficient diagnosis, with some authors advocating endoscopy and even MRI as the most accurate diagnostic method [25, 26]. In this discussion, we will consider three common and valid methods of evaluating patients with clinical concern for pyloric stenosis: abdominal palpation, upper GI and US (Table 1).

Clinical examination

Abdominal palpation is the first line evaluation of patients suspected of pyloric stenosis. However, it is probably the most operator-dependent of the three available choices. Successful palpation of the enlarged pylorus requires an experienced examiner, a significant time commitment of 10–20 min, a calm infant, and may need to be repeated or may necessitate evacuation of a distended stomach via NG tube [27], further increasing the time necessary to calm the infant and achieve a successful examination. In a retrospective study of 212 patients seen between 1974 and 1977, abdominal palpation by surgeons was successful in 99%, whereas in 187 patients seen between 1988 and 1991, it was successful in 79% of patients. Among non-surgeons, the success rate was 47% and 33% respectively [28]. In another retrospective study of 234 patients and five surgeons, the sensitivity ranged between 31% and 100%, with a mean sensitivity and specificity of palpation calculated at 74% and 99% respectively [20]. In yet another study of 200 infants with vomiting, the sensitivity and specificity of abdominal palpation was 85% [29]. A further study of 323 patients found a sensitivity of 72% and a specificity of 91% for abdominal palpation [30]. Although prior to the advent of sonography for this diagnosis, some authors have suggested sedation in order to increase the sensitivity of abdominal palpation, which increased from 70 to 100% in a small series of ten patients reported in 1976 [31].

Therefore, abdominal palpation can be accurate, it is most often noninvasive, and can be time-consuming.

Upper GI

Upper GI is the first imaging examination applied to the diagnosis of pyloric stenosis, and although it requires skill and experience, it is the least operator-dependent of the three diagnostic examinations discussed. The diagnosis is dependent upon passage of contrast material through the narrowed and obstructed channel; therefore the study necessitates further filling of the stomach with contrast, or first emptying the stomach via orogastric tube, and using fluoroscopy until some of the contrast is propelled from the stomach through the abnormal antropyloric channel (Fig. 1).

When performed by an experienced radiologist, the upper GI is accurate in the diagnosis or exclusion of pyloric stenosis, but few studies today specifically address its sensitivity and specificity. In a series of 46 patients without a palpable olive reported in 1967 [32], upper GI had a sensitivity of 96% and a specificity of 100%. In 7% of patients, there was high grade obstruction, with no passage of contrast through the channel after 30 min of fluoroscopic examination. In another study, the results of upper GI done on patients with pyloric stenosis during two time periods: 1974–1977 and 1988–1991 were reviewed.

During the first time period, the sensitivity of upper GI done at the outside hospital was 89.1%, similar to 90.6% in the second time period; the sensitivity of upper GI at the referral center was 96% in the early period, again not much different from 95% in the later period, but likely reflecting additional expertise at the referral center [28].

Therefore, upper GI is accurate, it is somewhat invasive because of the potential for vomiting and aspiration, potentially protracted fluoroscopy time and radiation exposure, and can at times be time-consuming for the same reason. It is an appropriate examination in cases in which there is continued clinical suspicion after negative abdominal palpation, if expertise in the US examination is low with a consequently low sensitivity and specificity.

Ultrasound

The US examination, similar to abdominal palpation, requires a skilled and experienced examiner. However, unlike the clinical examination, US requires neither a calm infant nor an empty stomach, and unlike the upper GI, is not dependent upon passage of gastric contents through the abnormal channel for diagnosis, as both the lumen and the surrounding musculature are directly visualized (Fig. 2); the child with a distended stomach does not need to drink, and there is no radiation exposure.

The seminal report of US in the diagnosis of five patients with pyloric stenosis examined with the static B-scanner, was published in the New England Journal in 1977 [33], and used the pyloric diameter (mean 2.3 cm, range 1.8–2.8 cm) as the diagnostic criterion. The advent of real-time ultrasound equipment and visualization of increasing detail led to muscle thickness and channel length being recognized as the increasingly important criteria for diagnosis. Analysis of the ensuing literature must be viewed within the context of evolution of the technique in unison with evolution of the equipment and of our ability to visualize increasing details of the antropyloric junction.

In a 1986 prospective study of 200 infants with vomiting, the authors, utilizing mechanical sector transducers operating at 7.5 mHz, found a mean muscle thickness of 3.4 mm, range 3–5 mm, and a mean pyloric length of 22.3 mm, range 18–28, recognizing the importance of real-time evaluation, lack of opening of the abnormal channel, and changes in the normal channel secondary to antropyloric peristaltic activity. They were able to discriminate between normal patients and patients with pyloric stenosis with a sensitivity and specificity of 100%, with no false positives or false negatives [29].

In a subsequent 1988 study including 323 examinations utilizing 5–7.5 mHZ transducers investigators found a sensitivity of 98.2% for US, and specificity of 100%, despite classifying an initially "suspicious" case diagnosed as pyloric stenosis 4 days later, as a false negative. These authors found a mean muscle thickness of 4.8 mm, range 3.5–6 and a mean pyloric length of 17.8 mm, range 11–25 [30].

In a retrospective 1991 study of 145 infants, investigators found that muscle thickness of 3 mm or greater was diagnostic of pyloric stenosis. Muscle thickness was <1.5 mm in 98% of normal patients, and <2 mm in 100%; "equivocal" thickness between 2 and 3 mm was found in six patients, two of whom went on to develop pyloric stenosis [34]. When appropriate referral for surgical therapy is taken as the end-point of the examination, the sensitivity and specificity of US were 100% and 99%, respectively.

A further 1994 study of 152 consecutive patients with non-palpable olives scanned with linear transducers operating at 7.5 mHz, and using real-time information of a non-opening channel and muscle thickness of 3 mm or greater, found a sensitivity and specificity of 100%, with the end-point being appropriate surgical referral [35].

Therefore, US in experienced hands, is accurate, non-invasive, and rapid in establishing the diagnosis of pyloric stenosis.

Repeat or follow-up examinations

Pyloric stenosis is a condition that develops after birth, in a population in which vomiting is prevalent and often secondary to reflux; therefore, depending upon pediatrician or parental anxiety, patients may present for imaging during the evolution of pyloric stenosis. The rate at which pyloric stenosis evolves is not known, and data are sparse on whether pylorospasm is always a self-resolving condition, or whether it is one of the initial steps in the development of pyloric stenosis in some patients.

Table 1 Performance characteristics of diagnostic examinations

	Sensitivity	Specificity
Palpation by surgeon	31–99% (mean 72.5%) [20, 22, 28–31]	85–99% (mean 93.3%) [20, 22, 29, 30]
By non-surgical clinician	26–47% (mean 36.7%) [28, 30]	
Ultrasound (in experienced hands)	97–100% (mean 99.3) [20, 22, 29, 30, 34–36]	99–100% (mean 99.8) [20, 22, 29, 30, 34–36]
Upper GI	90–100% (mean 95%) [20, 28, 32]	99.5% [20]

In a retrospective evaluation of 145 patients, six (4%) of the infants had equivocal findings, with muscle thickness greater than 2 mm, but less than 3. Pyloric stenosis had developed on follow-up examinations in two of these, with muscle thickness of 4 mm 2 weeks later. Pylorospasm resolved in two, and a diagnosis of milk allergy and eosinophilic gastroenteritis was made in the remainder [34].

In a prospective study of the vascularity of the pyloric muscle and mucosa in 75 patients, 41 of whom had pyloric stenosis, one patient, first imaged at 2 weeks of age because of high clinical concern due to familial history, was found to develop pyloric stenosis on follow-up examination, by 6 weeks of age [14].

Based on available data, in the small minority of patients with equivocal findings in the appropriate age group, if symptoms do not resolve, it is important to do a follow-up examination to assess for the interim development of pyloric stenosis. Performing an upper GI at the time of the initial examination would not resolve the issue, and may result in inappropriate surgical referral [35].

Conclusions

During the nineteenth and twentieth centuries, pyloric stenosis has evolved from an unknown condition with poor prognosis, to one that is well recognized with uniformly good outcome. However, diagnostic controversy centers on the relative sensitivity and specificity of the various techniques, and relative monetary cost. The data to date suggest that examiner skill is an important component of these statistics, and should be integral in decision-making. Abdominal palpation should be the first examination when pyloric stenosis is suspected, and if unsuccessful, should be followed by US if performed by experienced medical personnel. Upper GI can be done if accurate sonography is not available, or if reflux is the primary consideration.

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