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Pyloric stenosis: evolution from pylorospasm?

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Abstract. Over a 10-year period, we have performed pyloromyotomy on 260 infants with hypertrophic pyloric stenosis (HPS), 10 of whom had a history suggestive of pyloric stenosis but initially had neither the physical nor radiological findings to confirm the diagnosis. All 10 demonstrated pylorospasm on upper gastrointestinal series (UGIS), were treated medically without improvement, and subsequently developed classic HPS confirmed by repeat UGIS. Age at diagnosis ranged from 3 to 16 weeks (mean 8 weeks). Vomiting was progressively more projectile and severe from the onset until diagnosis and operation, with a duration of 5–50 days (mean 24 days). In 9 of the 10 patients a second UGIS demonstrated the diagnostic signs of HPS in 8 and suggested an antral web in the 9th. The interval between the two UGIS ranged from 2 to 46 days (mean 13 days). The 10th patient had a palpable hypertrophic pyloric muscle 9 days after the first UGIS and was operated upon without a follow-up UGIS. All 10 patients had classic HPS at operation. We conclude that although most infants with pylorospasm on UGIS improve with medical management, a small but significant number go on to develop HPS. Awareness of this variant of pyloric stenosis and appropriate follow-up UGIS will help to avoid undue delay in correctly diagnosing infants with persistent non-bilious vomiting.

Key words: Pyloric stenosis – Pylorospasm – Evolution

Introduction

Hypertrophic pyloric stenosis (HPS) is the most common surgical disorder causing vomiting in infancy. Eighty-five to 90% of patients with a history suggestive of HPS can be diagnosed by palpation of the hypertrophied pyloric

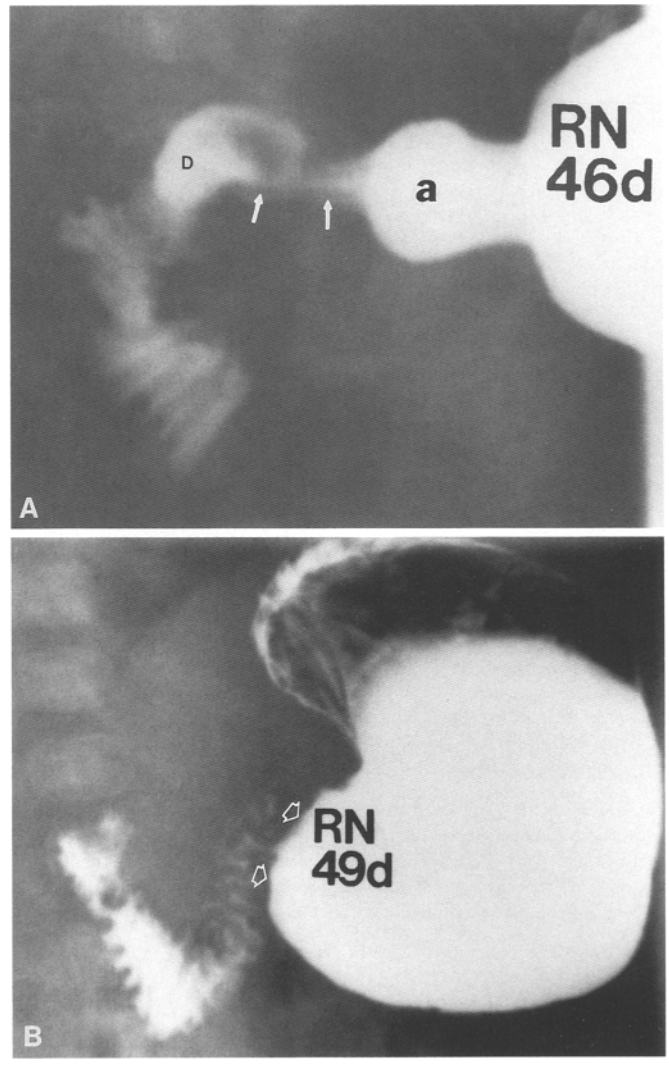

Fig. 1. Patient RN. **A** At 46 days of age: narrowed and elongated pylorus (*arrows*) and contracted antrum (*a*), which opened with prompt passage of barium (*D* = duodenum): diagnosis pylorospasm. **B** At 49 days of age: persistently narrowed and elongated pyloric channel with prominent shoulder sign (*arrows*) and minimal passage of barium: diagnosis pyloric stenosis

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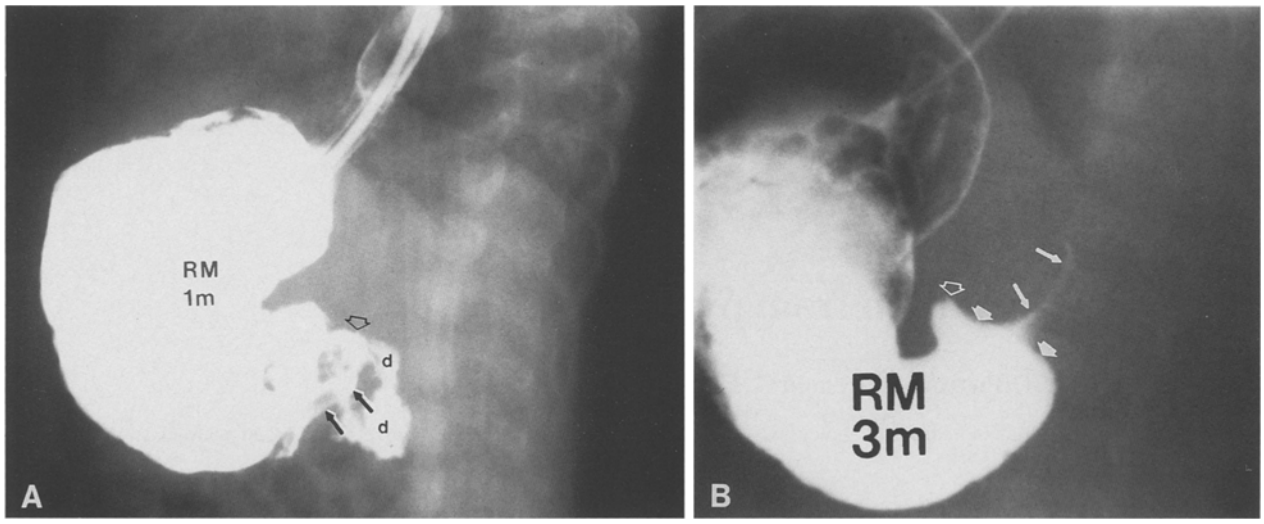

Fig. 2. Patient RM. **A** At 1 month of age: antral-pyloric narrowing (*arrows*) with intermittent good passage of barium into the duodenum (*open arrowhead* = duodenal bulb; *d* = duodenal sweep): diagnosis pylorospasm. **B** At 3 months of age: elongated (here incompletely filled), narrow pylorus (*arrow*) with prominent shoulder (*closed arrowheads*) and tit (*open arrowhead*) signs; poor gastric emptying: diagnosis pyloric stenosis

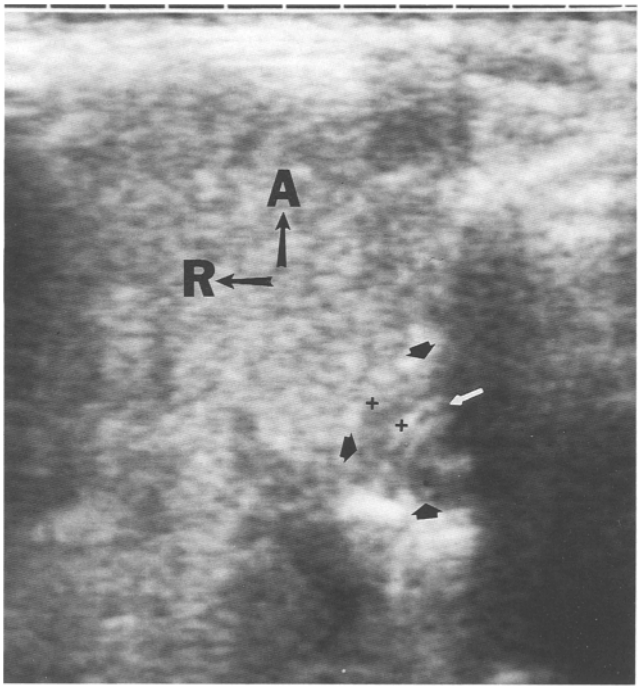

Fig. 3. Patient JP – 1 month of age, vomiting since birth. UGIS normal; ultrasound showed transient pyloric thickening (*arrowheads*; one wall {between +'s} = 4 mm) without significant elongation. Diagnosis pylorospasm. Vomiting resolved over 4 weeks. *White arrow* marks the pyloric lumen. Arrows point anteriorly (*A*) and rightward (*R*) for orientation

muscle. The remaining cases can usually be diagnosed by observing the classic radiological findings on upper gastrointestinal series (UGIS) [11]. More recently, the diagnosis has been quite accurately made by ultrasonography [2, 8]. Since 1977, however, in spite of the ease of preoperative diagnosis, we have cared for 10 infants with a history suggestive of HPS but with neither a palpable pyloric "olive" nor the characteristic changes of HPS on UGIS. Rather, they evidenced a form of pylorospasm that later evolved into classic HPS.

Clinical presentation and methods

Between September 1977 and September 1987, 260 pyloromyotomies were performed at C. S. Mott Children's Hospital, Ann Arbor. Of this group, 10 infants (3.8%) had a history typical of pyloric stenosis but on initial examination had neither the physical nor radiological findings of HPS. On UGIS all 10 demonstrated either pyloric narrowing without much hold-up of barium or pylorospasm with intermittent hold-up. They were treated medically for varying lengths of time with no improvement, and subsequent UGIS revealed classic HPS. All 10 infants then underwent pyloromyotomy.

Results

There were 7 males and 3 females ranging in age at diagnosis from 3 to 16 weeks (mean 8 weeks). Vomiting was progressively more severe and projectile from the onset until diagnosis and operation, with a duration of 5–50 days (mean 24 days). In 9 of the patients, a second UGIS demonstrated the diagnostic signs of HPS in 8 (Fig. 1 and 2) and persistent narrowing with only moderate hold-up suggestive of an antral web in the 9th. The interval between the two UGIS ranged from 2 to 46 days (mean 13 days). The 10th patient had a readily palpable hypertrophic pyloric muscle 9 days after the first contrast study and was operated upon shortly thereafter without a follow-up UGIS. All 10 patients had classic HPS at operation and recovered uneventfully from surgery.

Discussion

The precise etiology of HPS is unknown, and it is still uncertain whether this is an acquired or a congenital lesion.

Most infants develop symptoms 2 to 8 weeks after birth, and cases have first presented as late as 8 months of age [6, 13]. A genetic predisposition may play a role in some cases in that several instances of familial involvement have been reported [1, 3], and pyloric stenosis has occurred in twins and triplets [9, 14]. However, there are no convincing data to suggest that, overall, genetics or familial tendencies are overriding etiological factors.

One theory holds that the ganglion cells of the pyloric myenteric plexus are abnormal in either number or function [5, 15], however, this is more likely the result of degeneration secondary to overactivity and hypertrophy of the pyloric muscle than to intrinsic factors [19]. Furthermore, a recent electron microscopic study of myenteric plexus cells in infants with HPS concluded that they were normal [10]. There is very little evidence, therefore, to implicate abnormalities in the structure or number of ganglion cells as causative factors in HPS.

More recent evidence is accumulating that suggests that overactivity or prolonged spasm of the pyloric muscle may be a common pathway by which infants develop HPS. This may be caused by increased sensitivity of the pyloric ganglion cells to the gastrointestinal hormones secretin and cholecystokinin, which are known to cause both pyloric muscle spasm and hypertrophy [16]. The secretion of these hormones, in turn, is stimulated by gastric acid secreted by the parietal cells of the stomach, and many infants with HPS have been documented to have increased parietal cell mass [16]. Secretion of excess gastrin and vagal overstimulation have also been implicated in the etiology of HPS, both having been shown to induce pyloric muscle spasm and subsequent hypertrophy [4, 11, 12, 18]. Exogenous causes of HPS have also been documented, as in the case of five newborns who developed vomiting with pylorospasm following the administration of erythromycin estolate, which over a period of days developed into HPS [17].

Prolonged pyloric muscle spasm leading to muscular hypertrophy appears to be a common denominator in all of the proposed mechanisms and described causes of HPS, both intrinsic and extrinsic. Further support for this being the predominant problem is found in the successful medical treatment of HPS, which involves placing the pyloric muscle at rest with gastric decompression and anticholinergics [23].

If, when evaluating an infant with the classic symptoms of HPS, an abdominal mass (pyloric "olive") cannot be palpated, then some form of diagnostic imaging is indicated. During most of the 10 years encompassed by this review, we used the UGIS in this situation. Although we have recently been using ultrasound as the first diagnostic study when physical examination is equivocal or nonrevealing (Fig. 3) [2, 8], we continue to use the UGIS if ultrasound fails to reveal a classic hypertrophic pyloric olive. The UGIS not only demonstrates the antral, pyloric, and duodenal anatomy, but also permits evaluation of esophageal motility and the gastroesophageal (GE) junction for the presence of GE reflux and other causes of vomiting. This is particularly important when the radiologic appearance of the pyloric region is normal, in that it frequently enables the correct diagnosis, such as esophageal dysmotility or GE reflux, to be firmly established.

Pyloric stenosis is most often demonstrated radiographically by the classic well-known findings of an elongated pylorus producing a string sign and the hypertrophied pyloric muscle impression on the distal antrum creating the shoulder sign, the pyloric tit, and the beak sign [20]. Less often seen, but equally important to accurate diagnosis, are radiographic demonstrations of typical or incomplete muscle hypertrophy such as the double-track sign [7], the lesser curve mass, the spiculated antrum, the funnel antrum, and the pyloric niche [21]. In addition, there are a number of patients who have pyloric stenosis with no roentgenographically identifiable hypertrophied muscle mass but definite short-segment pyloric narrowing and obstruction [22]. These infants generally have a small muscle mass at the time of operation that was not readily palpable when the patient was awake. Most of these patients respond to pyloromyotomy, but in a few cases so little muscle hypertrophy is present that pyloroplasty may be the preferred procedure [22]. The etiology of short-segment pyloric stenosis with minimal muscle hypertrophy is unknown, but it may represent an early stage of classic HPS that, for as yet unknown reasons, smolders along without progressing to the usual degree of muscle hypertrophy.

Having taken into account all of the above possibilities, we made the diagnosis of "pylorospasm" in the 10 cases reported, which when followed clinically did not respond to medication or the passage of time, but developed into classic pyloric stenosis as confirmed by a subsequent diagnostic UGIS. As physicians and pediatricians consider pyloric stenosis earlier in infants presenting with non-bilious vomiting, we may see more cases of HPS in evolution from pylorospasm. The best clinical approach in these patients, based on the experience gained from this series of 10 infants, is to follow the patients until the vomiting resolves or until a definite diagnosis of HPS can be made. Definite HPS should be documented either by physical examination or by repeat diagnostic imaging (ultrasound or UGIS) so that adequate pyloric muscle hypertrophy is present to allow the surgeon to perform a standard pyloromyotomy. Otherwise, if the operation is done too early in the course of the disease, a more complicated operation such as a pyloroplasty may be required to alleviate the infant's gastric obstruction.

Our experience adds more evidence to the theory that pylorospasm is an important early finding in the development of HPS. Occasionally infants presenting early may have negative ultrasound or UGIS, and if symptoms persist a repeat study is indicated. Awareness of this early clinical picture in the course of HPS will help avoid undue delay in correctly diagnosing infants with persistent non-bilious vomiting.

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