Congenital Hypertrophic Pyloric Stenosis

Case Studies [1] | December 03, 2013
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Case history: A 4-week-old male neonate was brought with the complaints of vomiting after feeds for the past two weeks. It was projectile, nonbilious vomiting, progressively increasing in severity. On examination, no lump (‘olive’) was palpable. Transabdominal ultrasound was performed.

Image 1: Axial oblique ultrasound shows thickened, hypoechoic muscular wall (5.4 mm) and elongated pyloric canal (17.2 mm approximately).

Diagnosis: Congenital hypertrophic pyloric stenosis. Pyloric canal length was greater than 16 mm, and single wall thickness greater than 3 mm.
Image 2: Ultrasound shows the fluid filled, distended stomach, due to delayed gastric emptying.

Image 3: The distended stomach displaces the hypertrophied pyloric canal posteriorly, which resembles the uterine cervix (cervix sign). To the experienced radiologist and surgeon, the hypertrophied pylorus often looks obvious at first glance: “the hot dog in a bun” appearance.

Discussion: Hypertrophic pyloric stenosis is idiopathic thickening of pyloric muscle in infancy which creates progressive gastric outlet obstruction. It is typically seen in 2- to 12-week-old infants with worsening projectile vomiting. Dehydration, hypochloremic alkalosis, and jaundice may develop. Boys outnumber girls, 4:1. On physical examination, 10 percent to 40 percent of patients have either hyperperistaltic waves or a palpable pyloric “olive.” The latter finding is rarely felt except by experienced observers, wherein imaging may not be indicated. Otherwise, ultrasound is the procedure of choice. The cut-off values for muscle thickening, and elongated pyloric channel vary by author. Commonly accepted threshold values for HPS
- Single wall thickness of pylorus > 3 mm
- Pyloric channel length > 16 mm
- Pyloric diameter > 15 mm

On cross-section, the hypoechoic ring of hypertrophied pyloric muscle around the central echogenic mucosa, gives the appearance of bull’s eye/ dough-nut (target sign). Echogenic mucosal lining also tends to hypertrophy, and becomes redundant. Gastric hyperperistalsis and obliterated pyloric lumen are seen on dynamic exam. When duodenal bulb is easily identified, distended with fluid, diagnosis of HPS is unlikely.

Color Doppler shows increased flow in both the muscle and mucosa of infants with HPS. Barium studies may be used if the history is atypical, the emesis is bilious, and to evaluate for other causes of vomiting.

Image 4: Sonographic image resembling few of the UGI study findings. White arrow: Teat sign, Yellow arrow: Beak sign; Blue arrow: Shoulder sign.

The characteristic findings on UGI study are:
- Pyloric wall thickness > 10 mm
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• Elongation and narrowing of pyloric canal (2-4 cm in length)
• "Double / triple track sign" - Crowding of mucosal folds in pyloric channel
• "String sign" - Passing of small barium streak through pyloric channel
• Twining recess = "diamond sign" - Transient triangular tentlike cleft/niche in midportion of pyloric canal with apex pointing inferiorly secondary to mucosal bulging between two separated hypertrophied muscle bundles on the greater curvature side within pyloric channel
• "Pyloric teat" - Outpouching along lesser curvature due to disruption of antral peristalsis
• "Antral beaking" - Mass impression upon antrum with streak of barium pointing toward pyloric channel
• Kirklin sign = "mushroom sign" - Indentation of base of bulb (in 50%)
• Gastric distension with fluid
• Active gastric hyperperistalsis - "Caterpillar sign" (hyperperistaltic waves)

DIFFERENTIAL DIAGNOSIS
Pylorospasm (muscle thickness between 1.5-2 mm), can best be regarded as delayed gastric emptying. After a variable period (days to weeks), the symptoms of pylorospasm disappear. Whether or not pylorospasm represents a forme fruste of pyloric stenosis is not certain because adrenogenital syndrome, dehydration, and sepsis have also been implicated as causes. Finally, gastroenteritis, pyloric channel (stress) ulcer, and congenital abnormalities such as antral web and gastric duplication may cause symptoms similar to those of HPS. However, USG is usually successful in excluding HPS when one of these other conditions is present.

REFERENCES
3. Pediatric radiology, the requisites, 3rd edition.
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