Infantile hypertrophic pyloric stenosis

Synonyms: congenital hypertrophic pyloric stenosis, hypertrophic pyloric stenosis

Pathogenesis

This condition is caused by diffuse hypertrophy and hyperplasia of the smooth muscle of the antrum of the stomach and pylorus. It usually occurs in infants aged 2-8 weeks. The pyloric muscle hypertrophy results in narrowing of the pyloric canal, which can then become easily obstructed [1] .

Genetic studies have identified susceptibility loci for infantile hypertrophic pyloric stenosis (IHPS) and molecular studies have concluded that smooth muscle cells are not properly innervated in this condition [2] .

Epidemiology [3]

- The incidence of pyloric stenosis is 2 to 5 in 1,000 live births per year.
- It is more common in males, with a male:female ratio of 4:1.
- There is a familial link, with a polygenic hereditary pattern.
- Pyloric stenosis is more common in the white population. It is less commonly seen in Indian, Asian and Black populations.
- HPS occurs very rarely in adults and must then be differentiated from gastric cancer [4] .

Presentation

- Typical presentation is onset of vomiting at 2-8 weeks of age (late presentation up to 6 months can occur but is very rare) [5] [6] :
    - Vomiting: non-bilious, often but not always projectile and usually 30-60 minutes after a feed, with the baby remaining hungry.
    - Vomiting increases in frequency over several days.
    - Vomiting also increases in intensity until it becomes projectile.
    - Slight haematemesis may occur.
- Persistent hunger, weight loss, dehydration, lethargy, and infrequent or absent bowel movements may be seen.
- Stomach wall peristalsis may be visible.
- An enlarged pylorus, classically described as an 'olive', may be palpated in the right upper quadrant or epigastrium of the abdomen:
    - The 'olive' is best palpated at the start of a feed but is often missed [7] .
    - With the infant supine and the examiner on the child's left side, gently palpate the liver edge near the xiphoid process.
    - Then displace the liver superiorly; downward palpation should reveal the pyloric olive just on, or to the right of, the midline.
    - It should be possible to roll the pylorus beneath the examining finger.

Differential diagnosis

- Feeding problem or milk intolerance.
- Gastro-oesophageal reflux.
- Gastroenteritis.
- Duodenal atresia, oesophageal atresia or other bowel obstruction in the newborn.

- Intestinal malrotation/acute midgut volvulus.

Investigations

- Serum electrolytes (for correction of imbalances before surgical repair); there is often metabolic alkalosis with severe potassium depletion. However, biochemical disturbances are now much less common with earlier diagnosis[7] .
- Ultrasound is reliable and easily performed and has replaced barium studies as the main investigation[8] . There is a normal variation of pylorus muscle measurements with age and gestation but ultrasound has a very high sensitivity and specificity[9] .

Management

- Pre-operative management is directed at correcting the fluid deficiency and electrolyte imbalance.
- Ramstedt's pyloromyotomy is easily performed and is associated with minimal complications.
- Laparoscopic pyloromyotomy is also performed and is an effective alternative where suitable facilities are available. Time to achieve full enteral feeding has been found to be significantly shorter in those treated laparoscopically vs those having open pyloromyotomy[10] .

Complications

- Vomiting can lead to dehydration, weight loss, and severe electrolyte disturbance (hypokalaemic and hypochloraemic metabolic alkalosis).
- Operative complications include mucosal perforation, continued postoperative bleeding (very rare), and persistent vomiting due to incomplete pyloromyotomy (rare).
- Foveolar cell hyperplasia (FCH) has been reported as a rare cause of persistent gastric outlet obstruction in patients with IHPS[11] . An extended pyloromyotomy is required to manage this.

Prognosis[3]

- Prognosis is excellent unless diagnosis is delayed and prolonged severe dehydration occurs.
- Mortality is rare after pyloromyotomy.

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