Infantile hypertrophic pyloric stenosis

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history

1717: *Blair* described HPS (postmortem finding)
1888: *Hirschsprung* first complete description of HPS

1908: first surgical correction (splitting the hypertrophied pyloric muscle and closing the muscle transversely)

1912: *Ramstedt* suggested that closure of muscle was not necessary (current <u>standard operation</u> was established)

Incidence

Most common cause of GOO in infants
Prevalence of IHPS 1.5 to 4 per 1000 live births
More common in boys than girls (2:1 to 5:1)
It is generally agreed the IHPS is not congenital abnormality

Anatomy and histology

 Gross appearance of pylorus in IHPS: enlarged pale muscle
Hypertrophy and hyperplasia in circular layer
Hypertrophy in underlying mucosa

Etiology

No definitive cause

Evidence of genetic predisposition include :

- 1. Variability among race
- 2. clear male preponderance
- 3. increased risk in first-born infant
- 4. certain ABO blood type

Etiology(cont.)

- Environmental factors
- 1. method of feeding
- 2. seasonal variability
- 3. erythromycin exposure

Clinical features and DDx

Typical clinical findings :

- Nonbilious vomitting at 2 to 8 weeks of age (especially at 3 to 5 weeks)
- Blood in emesis as a result of gasteritis or esophagitis
- Indirect hyperbiliribinemia in 2% to 5% of infants (as high as 15 to 20 mg/dl
- Other medical cause of nonbilious vomitting ;

Gasteroenteritis , increased ICP , metabolic disorders

Other surgical cause of nonbilious vomitting ;

• antral webs, pyloric atresia, duplication cyst of the antropyloric region, and ectopic pancreatic tissue within the pyloric muscle

Diagnosis

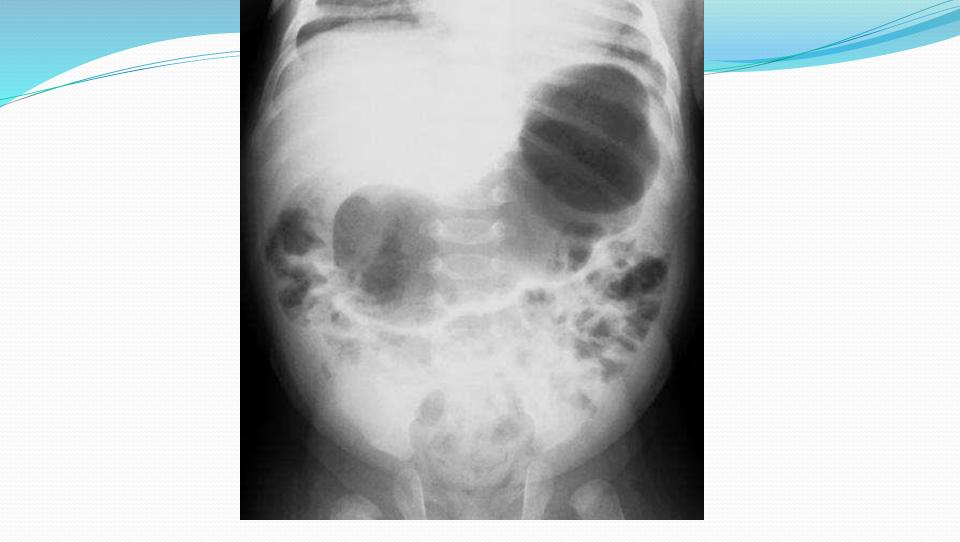
Cardinal features:

- Nonbilious projectile vomiting
- visible peristaltic waves in the left upper part of the abdomen.
- hypochloremic, hypokalemic metabolic alkalosis
- Definitive diagnosis can be made in 75% by careful P/E
- **Ultrasonography** is most common ,standard and choice diagnostic test for the diagnosis IHPS

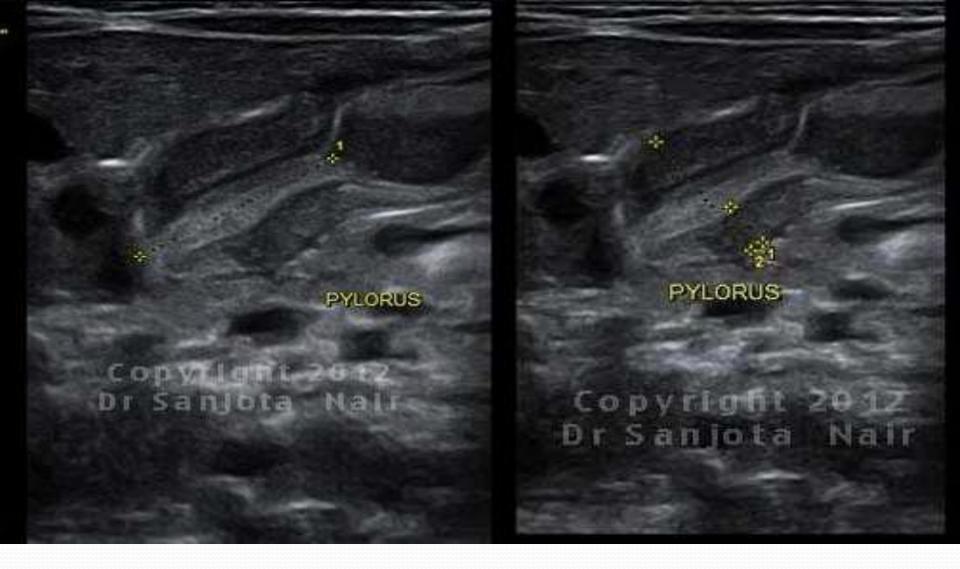
Findings in UGI :

- Elongated pyloric channel
- Indentation on the antral outline

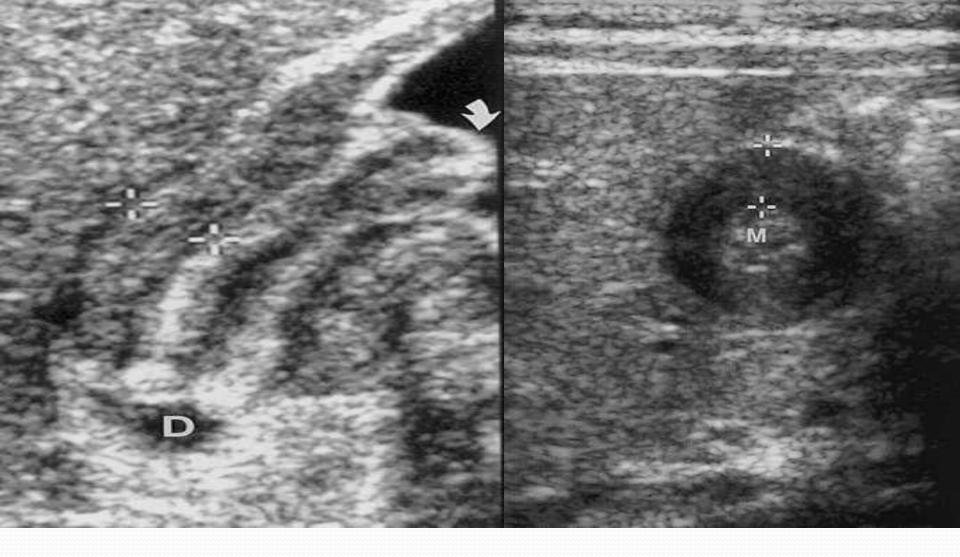
R/O of pyloric spasm is important



Supine radiograph in an infant with vomiting demonstrates the caterpillar sign of active gastric hyperperistalsis



In longitudinal plane the pyloric canal length is 18mm, The pyloric muscle thickness measuring 4.6 mm and the pyloric diameter measuring 14 mm



a. Sonograms in a patient with IHPS. (a) Longitudinal sonogram shows anterior thickened muscle (cursors). Double layer of crowded and redundant mucosa fills the channel and protrudes into fluid-filled antrum (arrow). D = fluid-filled duodenal cap. (b) Cross-sectional sonogram shows circumferential muscular thickening (cursors) surrounding the central channel and filled with mucosa (M).

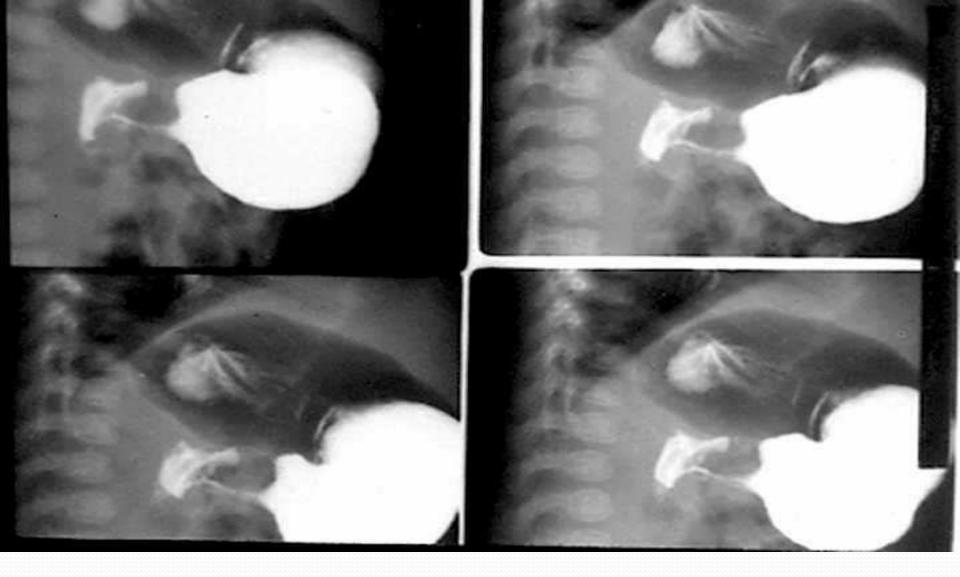
Mashroom sign Double/inple track sign / Teat sign

Diamond sign

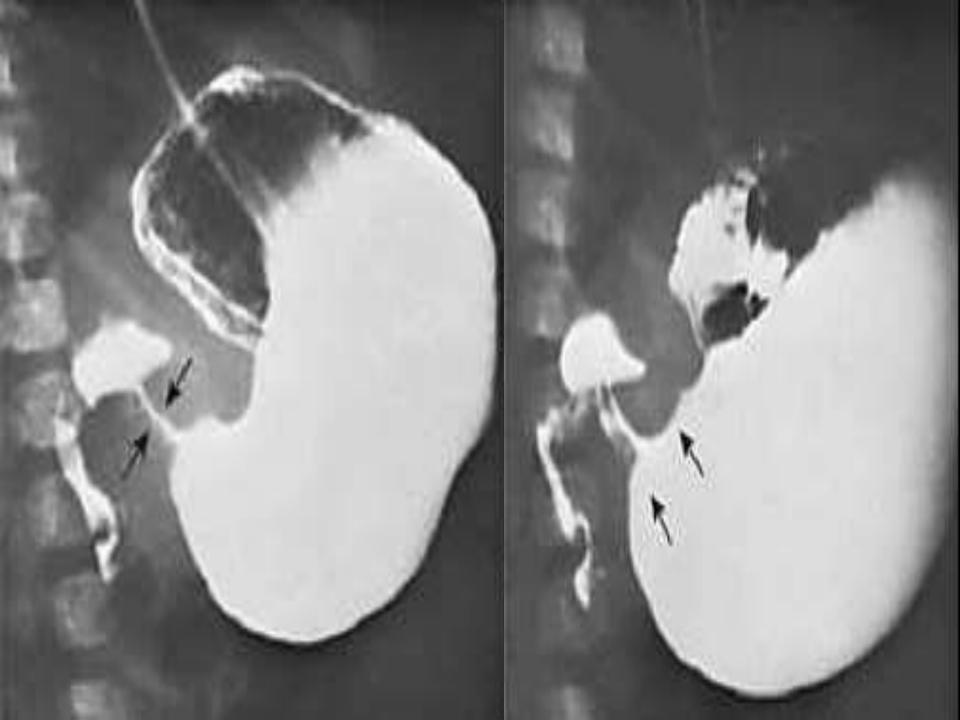
Beak sign

Caterpillar sign

Shoulder sign



INFANTILE HYPERTROPHIC PYLORIC STENOSIS



Treatment

Pre operative preparation:

- The length of preparation depend on the severity of the fluid and electrolytes abnormalities
- Three levels of severity primarily on the basis of the serum carbon dioxide : 1. slight (<25 meq/lit) 2.moderate (26 to 35 meq/lit) 3. severe (>35 meq/lit)
- Nasogastric tube should not be placed routinely
- Aggressive resuscitation should be avoided (possibly leading to seizure and ...)
- Ideal intravenous administration : 5% Dextrose in 0.45 N/S containing 20 meq/lit Kcl
- **Serum** K level should be carefully monitored.

- An initial rate for fluid resuscitation is 1.25 to 2 times the normal maintenance rate
- Normalizing the serum bicarbonate level usually lags behind normalization of fluid volume and serum K and Cl.
- **Hyperbilirubinemia resolves postoperatively.**

Operative procedure:

- It is important that fluid and electrolities must be corrected preoperatively including having a serum bicarbonate < 30 meq/Lit</p>
- Before the induction of anesthesia aspirate the stomach
- Choice operative procedure *Ramstedt Pyloromyotomy*

minimal laparatomy(open) technique

- Standard open approach is RUQ transverse incision
- Splitting of rectus muscle
- Alternative incision: supraumblical curve incision
- Pylorus can be stabilized by the index finger of the surgeon
- Serosa on the anterior wall of HPS is incised with a scalpel from proximal to hypertrophied muscle extending to pyloric vein
- Splitting the muscles with clamp or scalpel handle
- Most incomplete myotomies are a result of failure to extend it far enough proximally onto the antrum.
- □ **If perforation** occur the submucosa should be approximated with interrupted fine absorbable suture and a portion of omentum placed over this site.



Fredet- Ramstedt's Pyloromyotomy

Laparoscopic procedure

- ✓ the first description by Alain in 1991
- The infant is placed in supine at the end of table
- A 5 mm port in umbilicus another in LUQ and RUQ
- Grasper from LUQ for grasping of antrum
- And cautery from RUQ
- Splitting by pyloric spreader
- Leak test by insufflation of 60 ml air in NG tube



Laparoscopic Pyloromyotomy



Postoperative management

- Feeding can be started within 4 hours after surgical procedure
- If the Pt had hematemesis delay feeding 6 to 12 hours

Discharge 24 hr after feeding

Nonoperative management

Aspiration and malnutrition and prolonged hospital stay led to abandonment of this type of management

Complications

- **Vomitting**, frequent in the early postoperative period secondary to <u>gastroesophageal reflux</u>, <u>discoordination of</u> <u>gastric peristalsis</u>, or <u>gastric atony</u>
- Frequent vomiting persisting beyond 3 to 4 days may suggest an incomplete myotomy or an unsuspected perforation
- Contrast study may demonstrate a leak but is not
- helpful in evaluating the completeness of the myotomy
- It takes several weeks for the radiographic appearance of the pylorus to improve.
- Persistent and frequent vomiting 1 week beyond the pyloromyotomy may require reexploration.

