Portal Venous Gas and Gastric Pneumatosis in an infant with Pyloric Stenosis – a rare presentation of a common condition.

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Disclosures

- None.

Objectives

1. Review clinical and diagnostic imaging features of infantile hypertrophic pyloric stenosis (IHPS).

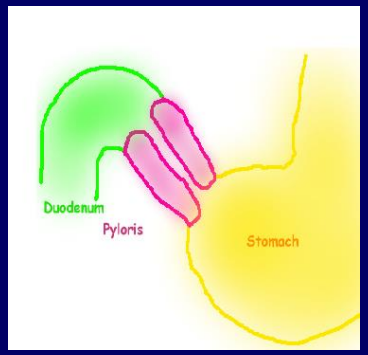
2. Discuss an atypical case of infantile pyloric stenosis that presented with portal venous gas and gastric pneumatosis and highlight the significance of recognising this condition.

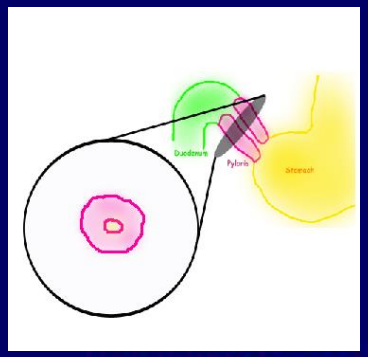
How rare is this association ?

- There have been very few reported cases of portal venous gas in IHPS (2,3,4) and gastric pneumatosis in IHPS (5,6,7).
- Only one previous case report (1) describes both portal venous gas and gastric pneumatosis in an infant with pyloric stenosis.
- We believe ours is the only other such case report !

Introduction:

- Infantile hypertrophic pyloric stenosis (IHPS) is a common condition affecting young infants, in which the antropyloric portion of the stomach becomes abnormally thickened and manifests as obstruction to gastric emptying.
- Not present at birth, but mechanical obstruction typically develops in the first 2-12 weeks of life.
- Most common condition requiring surgery in infants.
- Has a male predilection (M:F is 4:1).
- Etiology remains unknown.





Clinical Presentation:

- The infant presents with a recent onset of forceful nonbilious vomiting, typically described as “projectile.”
- Initially intermittent, the frequency of emesis increases with time.
- Vomitus may be stained with blood due to rupture of small capillaries in gastric mucosa.
- Dehydration and weight loss are often present. Occasionally, indirect hyperbilirubinemia may be seen.
- The infant may appear to be starving and crying inconsolably.

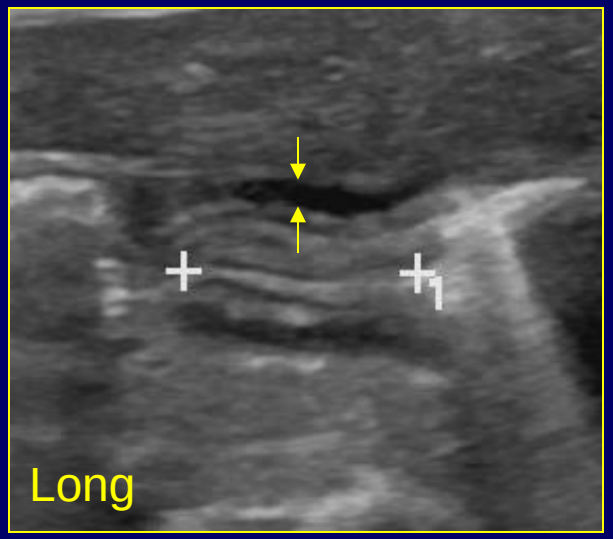
Clinical Exam:

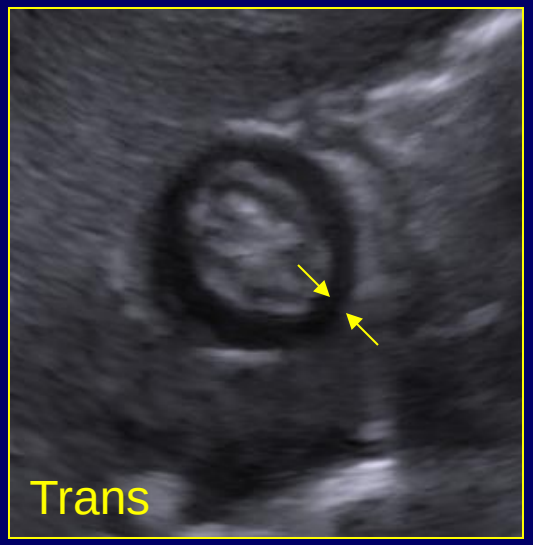
- On exam, distended stomach and vigorous active peristalsis may be visible through a thin abdominal wall.
- An experienced clinician may be able to palpate an olive-sized firm mass representing hypertrophied pylorus, but this could be challenging.
- Hypochloremic metabolic alkalosis is the characteristic biochemical abnormality because vomiting of gastric contents leads to depletion of sodium, potassium and hydrochloric acid.

Diagnostic Imaging in Pyloric Stenosis:

- Ultrasonography is the primary imaging modality that visualizes the pyloric muscle thickness and does not use ionizing radiation.
- Plain radiographs of the abdomen are often performed as baseline to rule out more emergent conditions such as bowel obstruction, midgut volvulus, pneumatosis intestinalis and free air.
- Fluoroscopic upper gastrointestinal study is used to exclude malrotation, but may be used in symptomatic infants with negative ultrasound.
- CT/ MRI is considered inappropriate.

Pyloric Ultrasound: Normal

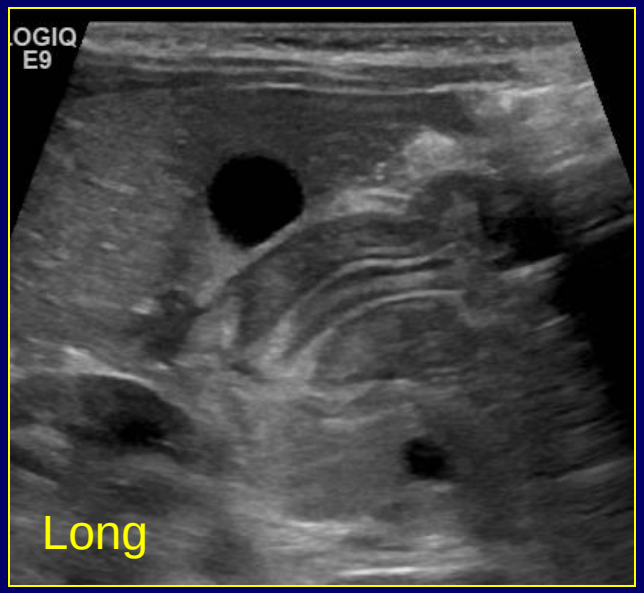


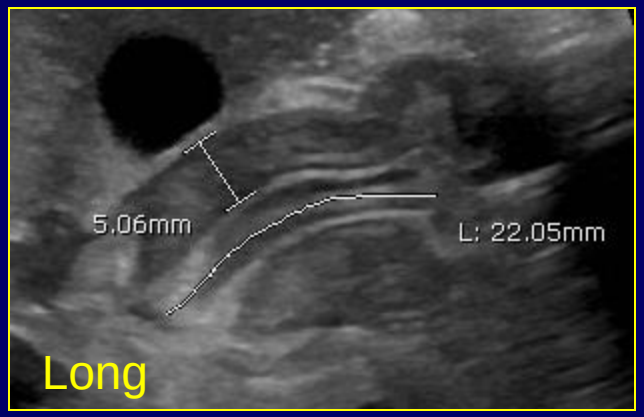


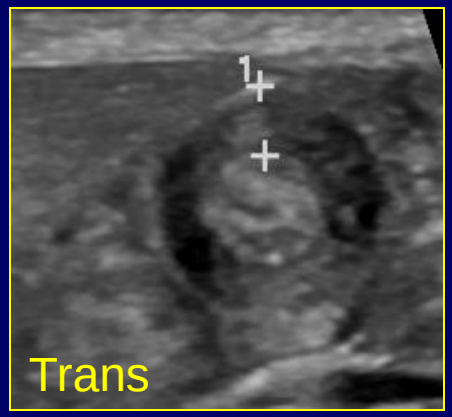
Normal pyloric muscle-
- Thin hypoechoic layer, (between arrows)
- Measures <2mm thick

Normal pyloric channel-
- 11 to 14 mm in length

Pyloric Ultrasound: Positive for IHPS



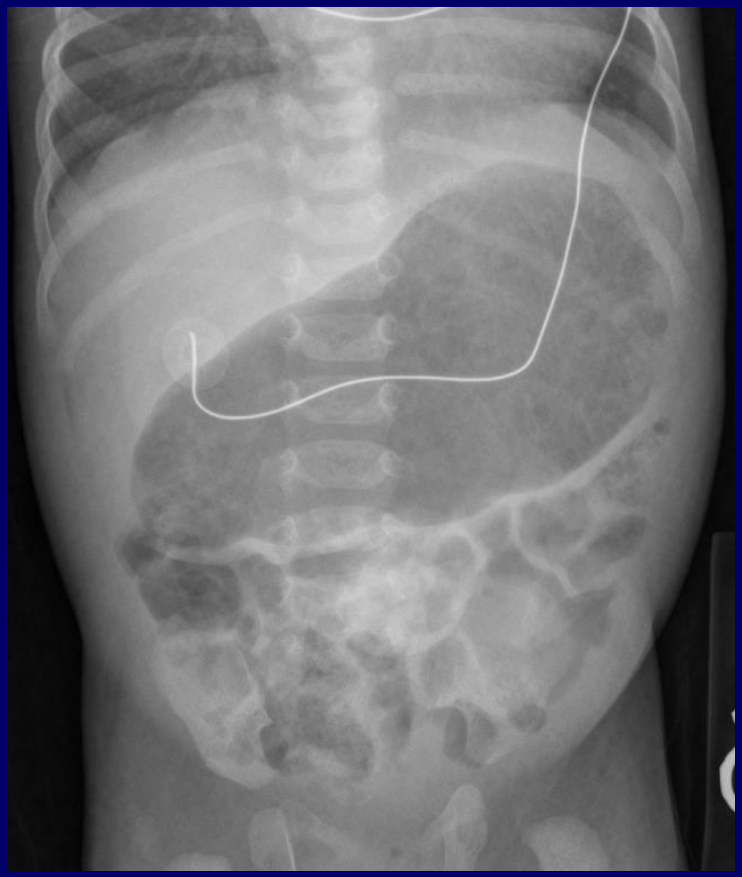




Sonographic criteria:

1. Pyloric muscle thickness >3mm
2. Pyloric canal length >17mm
3. Absence of passage of peristaltic wave through pylorus (observed real-time)

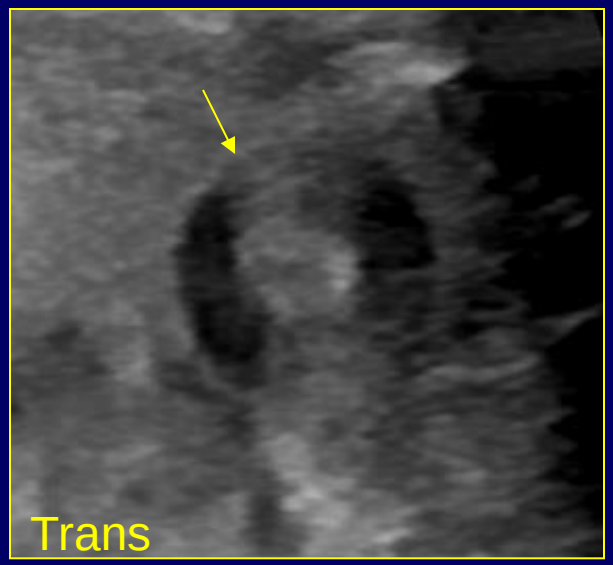
Radiographic signs of pyloric stenosis:



Plain Abdomen radiograph may show:

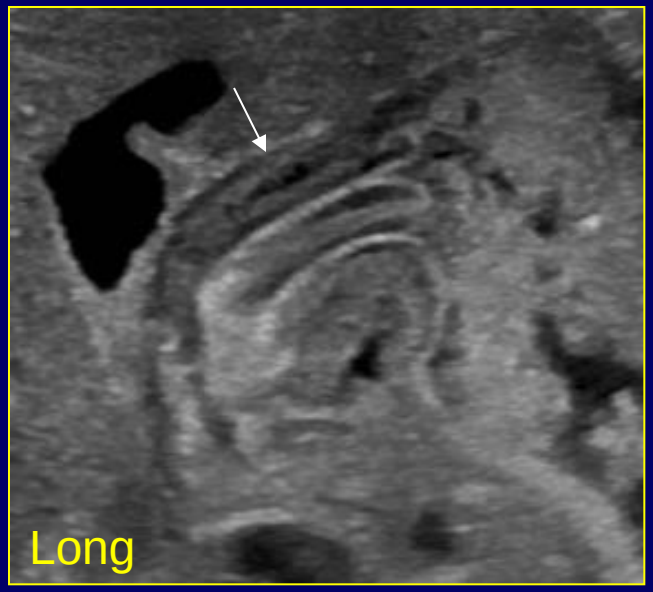
- Marked gastric distension
- Mottled frothy gastric contents
- Caterpillar stomach sign – indentation of gastric air shadow by obvious peristaltic waves
- Paucity of distal bowel gas

Sonographic signs of pyloric stenosis:



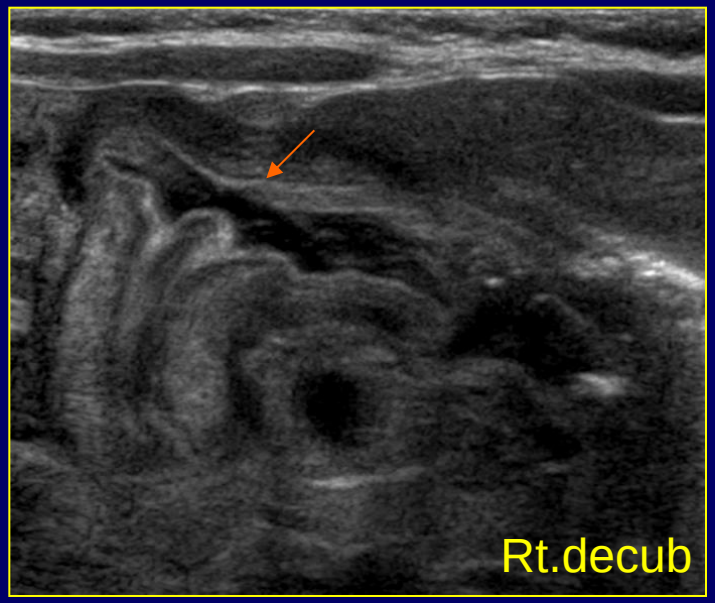
1. Target sign – Peripheral ring of hypertrophied hypoechoic muscle surrounding central echogenic mucosa, resembling a doughnut (yellow arrow)

Sonographic signs of pyloric stenosis:



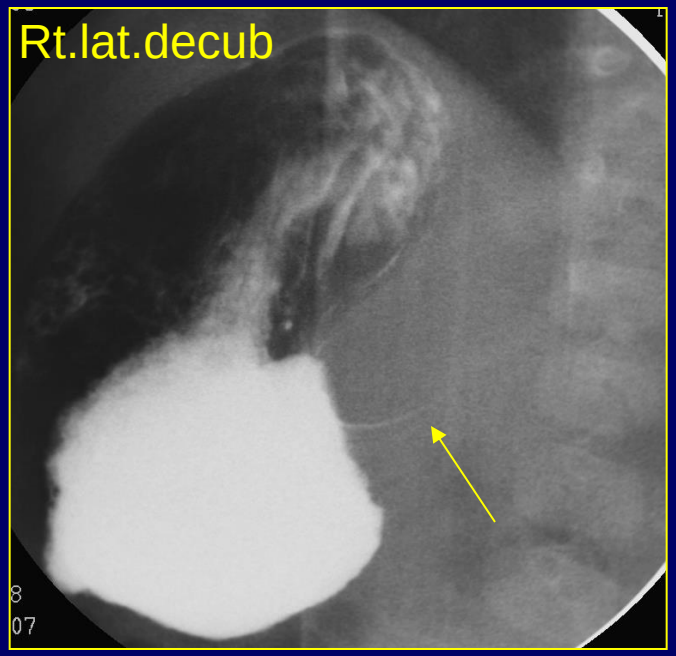
2. Cervix sign - Extension of hypertrophied pyloric muscle into the antrum and elongated pyloric channel form an image that resembles a cervix (white arrow)

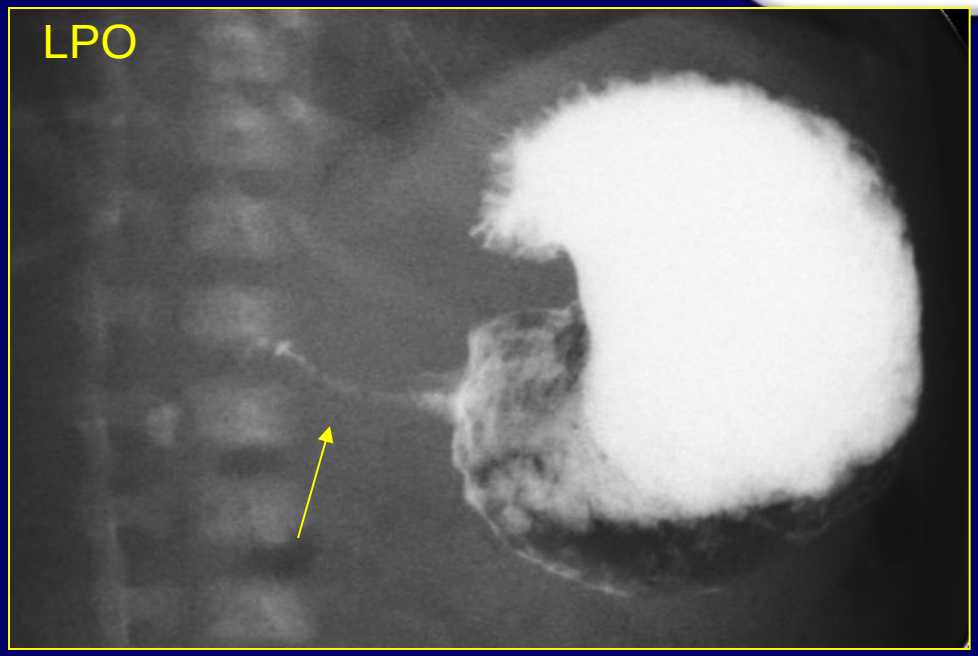
Sonographic signs of pyloric stenosis:



3. Antral nipple sign – redundant pyloric mucosa protruding into the gastric antrum (orange arrow)

Fluoroscopic signs of pyloric stenosis:

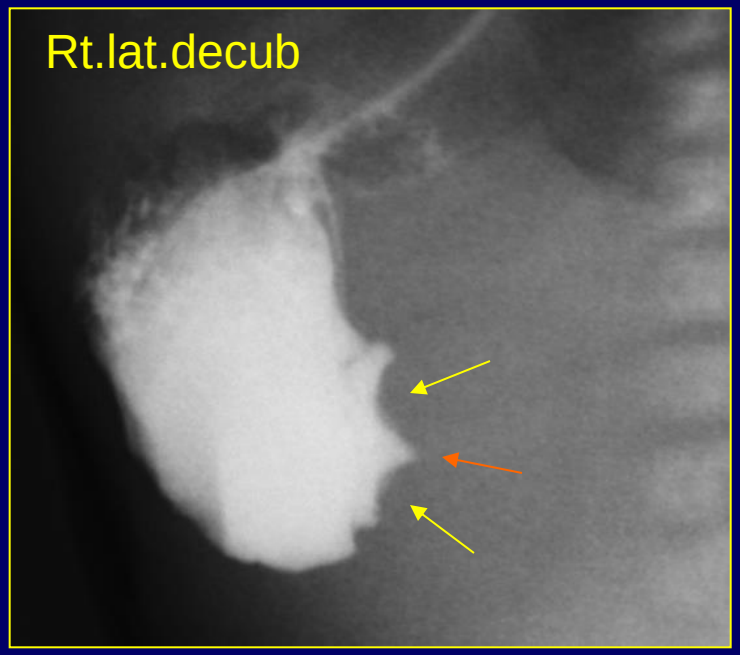




1. Delayed gastric emptying

2. String sign – Central streak of contrast lining narrowed lumen of elongated pylorus (yellow arrow)

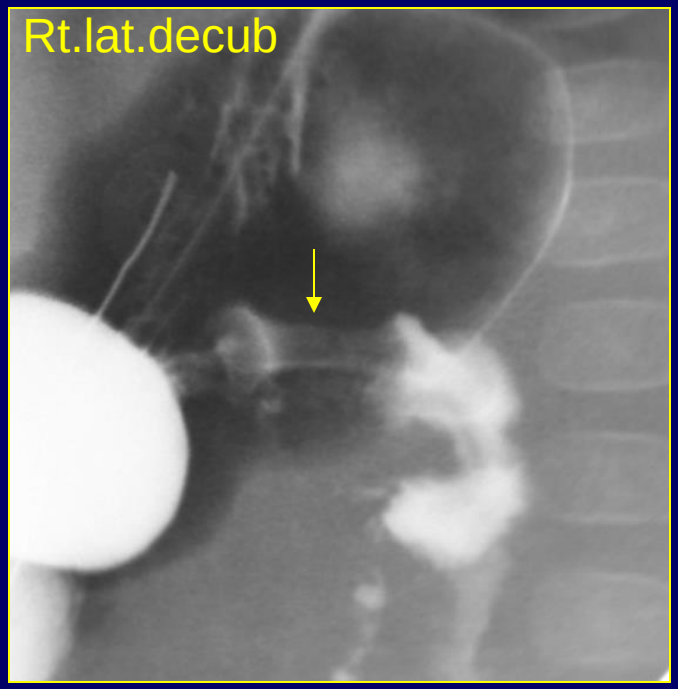
Fluoroscopic signs of pyloric stenosis:

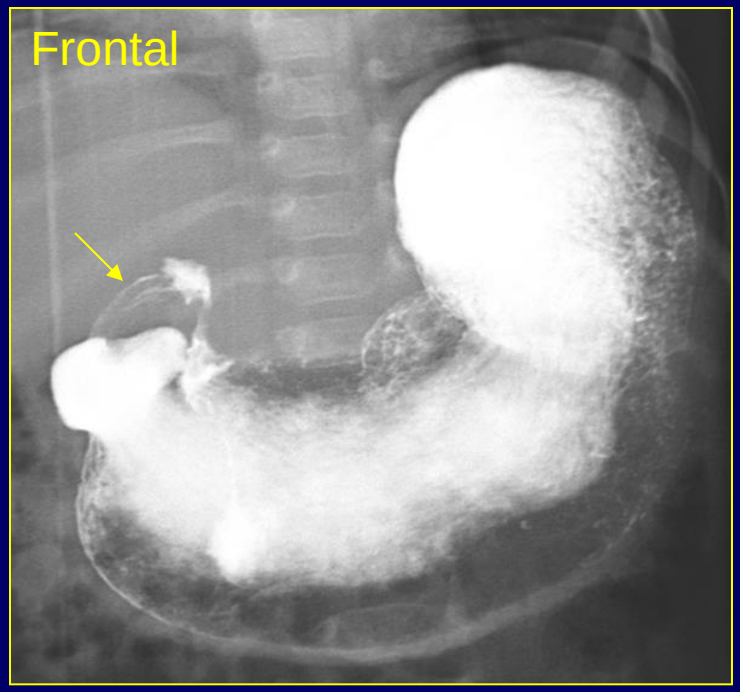


3. Beak sign – Contrast enters the proximal pyloric channel resembling a beak (orange arrow).

4. Shoulder sign – seen when the hypertrophied pylorus indents the contrast filled antrum (yellow arrows)

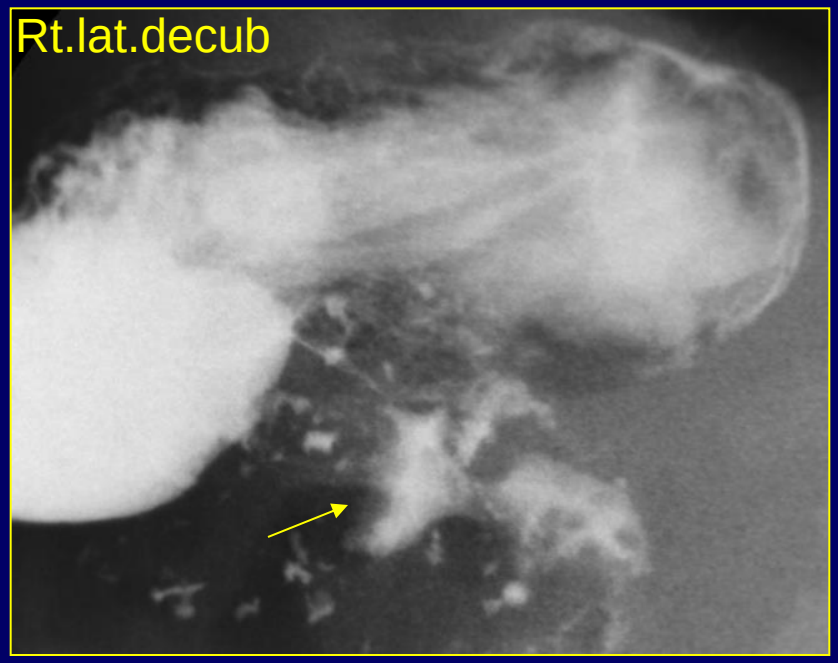
Fluoroscopic signs of pyloric stenosis:





5. Double track sign – Twin parallel streaks of contrast seen lining pyloric channel due to intervening redundant mucosa (yellow arrow)

Fluoroscopic signs of pyloric stenosis:



6. Mushroom sign – caused by mass effect from hypertrophied pylorus on base of duodenal bulb (arrow)

Here comes our interesting case……..

History: A 4 week-old previously healthy full term male infant presented with two week history of sweating with feeds, post-feed perioral cyanosis and frequent non-bilious vomiting.

Reason for ER visit : Apparent life-threatening event (ALTE) at home when he vomited after a feed and began turning purple with slow breathing. He responded to suctioning of the nose and mouth.

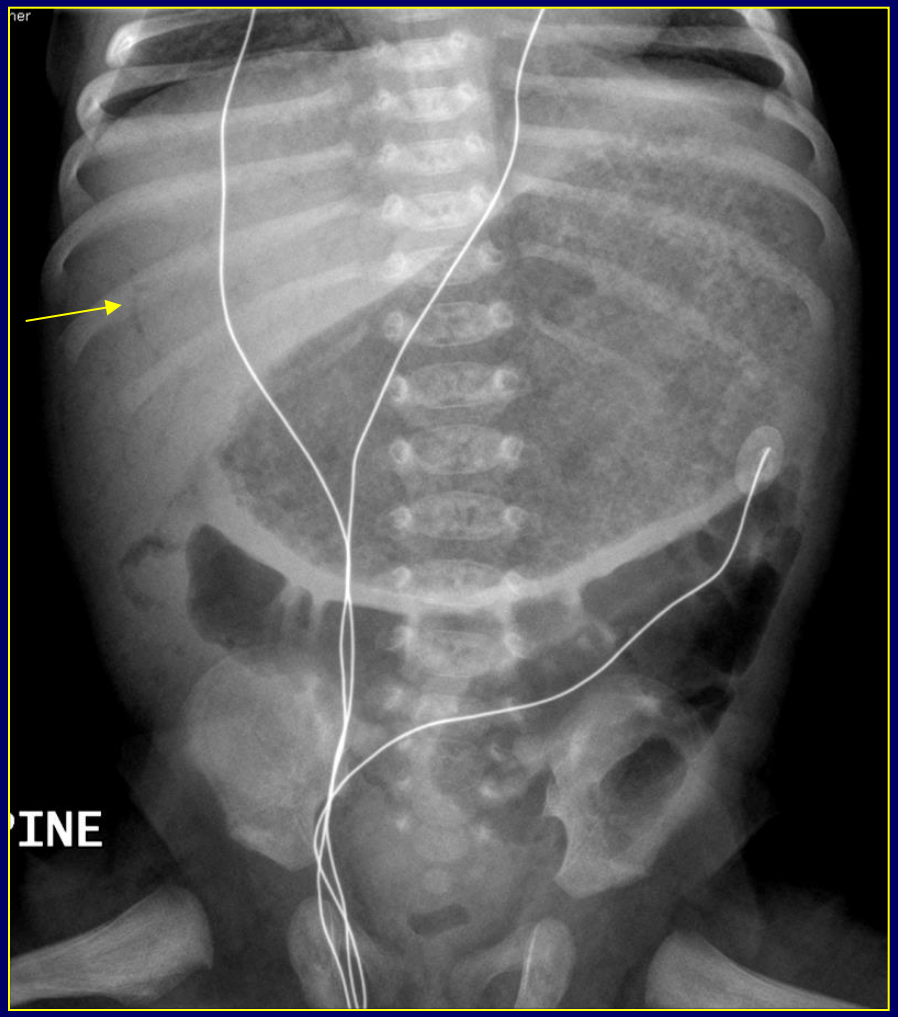
Other development history: Gaining weight appropriately, No fever or significant illness.

Physical Exam: Fussy, dehydrated and appeared very sick.

Clinical Differential consideration:

1. Severe gastroesophageal reflux
2. Cardiac etiology
3. Tracheoesophageal fistula
4. Pyloric stenosis (considered unlikely due to presence of cyanosis)

Plain radiograph of abdomen:



Frontal abdomen radiograph :

1. Massive gastric distension

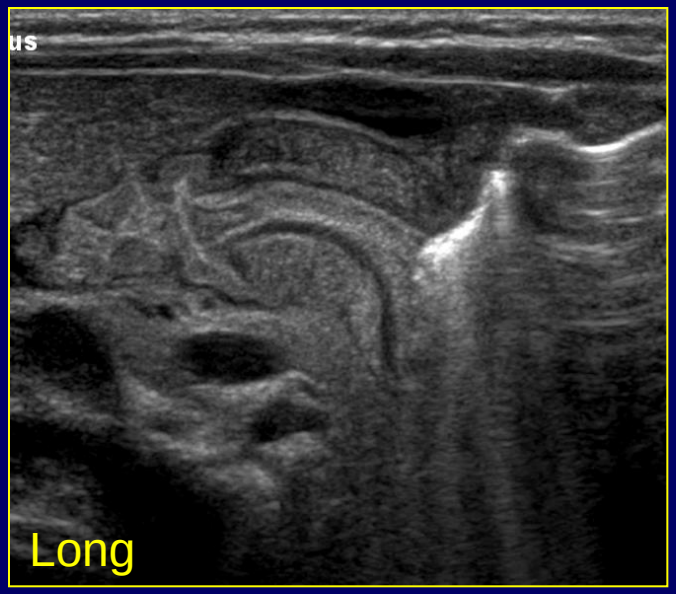
2. Mottled bubbly appearance of stomach concerning for gastric pneumatosis

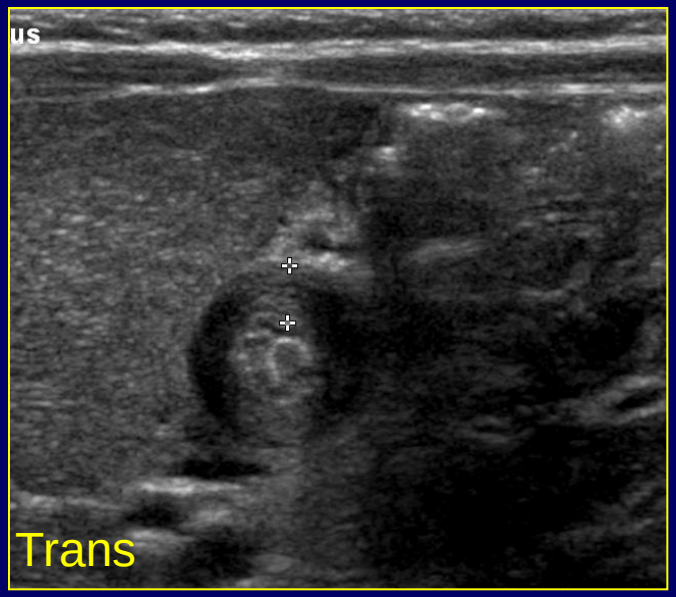
3. Linear branching lucencies (arrow) overlying liver consistent with portal venous gas

In the meantime:

Since gastric pneumatosis and portal venous gas are considered ominous in gastrointestinal disease, a complete abdominal sonogram was ordered to rule out underlying mesenteric ischemia/ embolic event.

Sonography of abdomen:



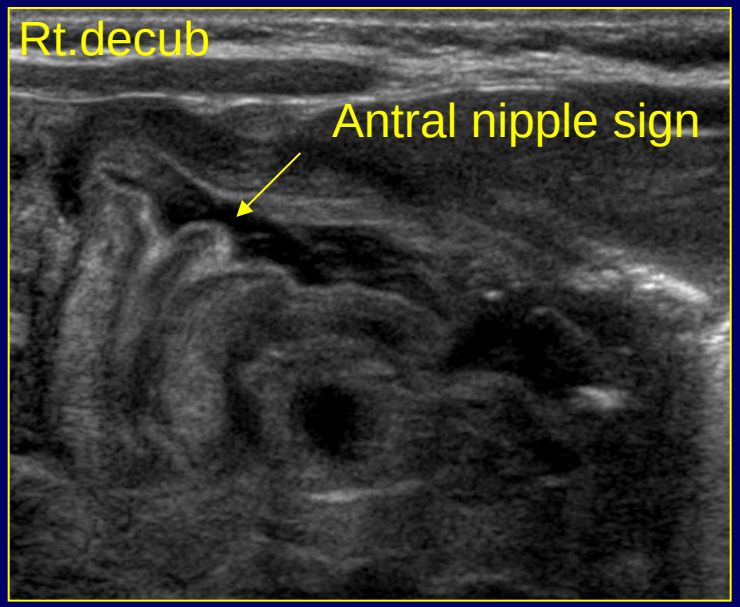


Pyloric muscle was thickened measuring 4 mm.

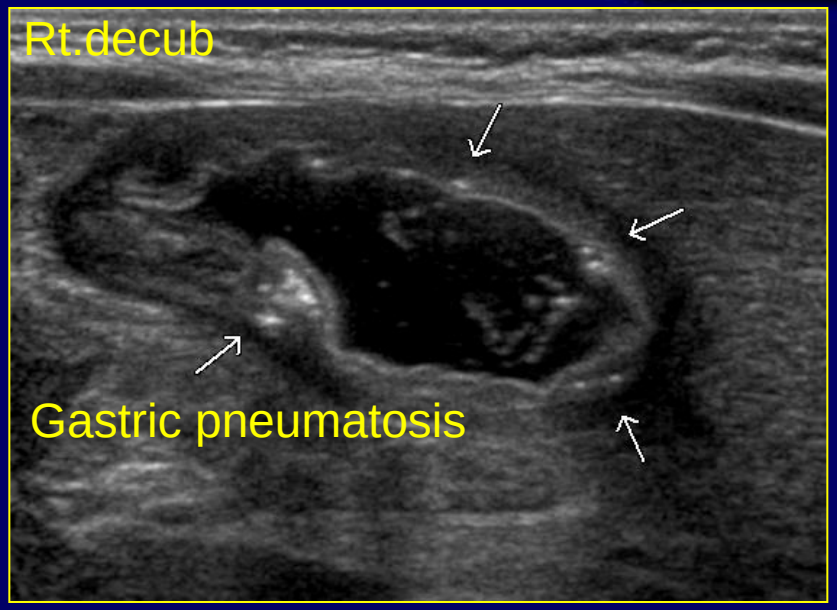
Pyloric channel was elongated measuring 20 mm.

Sonogram confirmed pyloric stenosis.

Sonography of abdomen:



Antral nipple sign of protruding mucosa seen in right decubitus position.



Gastric wall thickening with echogenic specks representing gastric pneumatosis were noted.

By the way……

- PV gas was not seen on sonogram, not unusual due to dynamic nature of movement of air in the wall of GI tract and portal venous system.

- Blood tests : Hypochloremic metabolic alkalosis, a characteristic biochemical disturbance in pyloric stenosis.

- Cardiac exam : Normal

Management:

Surgery : Laparoscopic pyloromyotomy, uneventful recovery.

Discussion:

Pyloric stenosis

↓

Gastric outlet obstruction

↓

Increased intraluminal pressure in stomach

↓

That forces gas through intact gastric mucosa into venules

↓

Then into veins, eventually draining into portal venous system

Discussion:

- Gastric pneumatosis and PV gas are considered ominous signs that raise possibility of bowel ischemia and necrotizing enterocolitis.
- However, a mechanical cause of gastric outlet obstruction such as pyloric stenosis may occasionally present with gastric pneumatosis and associated portal venous gas.

Summary:

It is important to recognise that infantile hypertrophic pyloric stenosis is a benign cause of gastric pneumatosis and portal venous gas for which pyloromyotomy is curative.

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