Pediatric Surgery

LIMU

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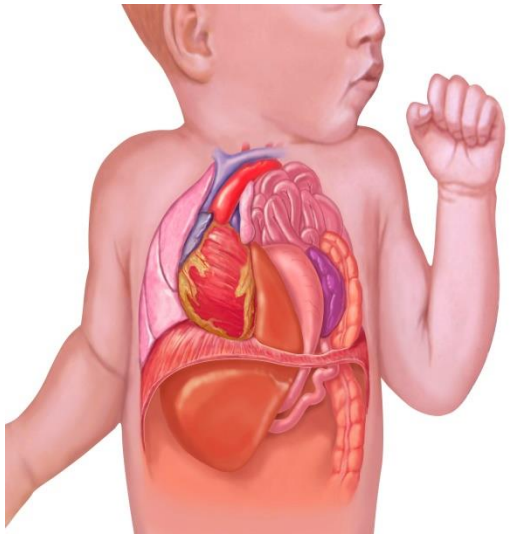
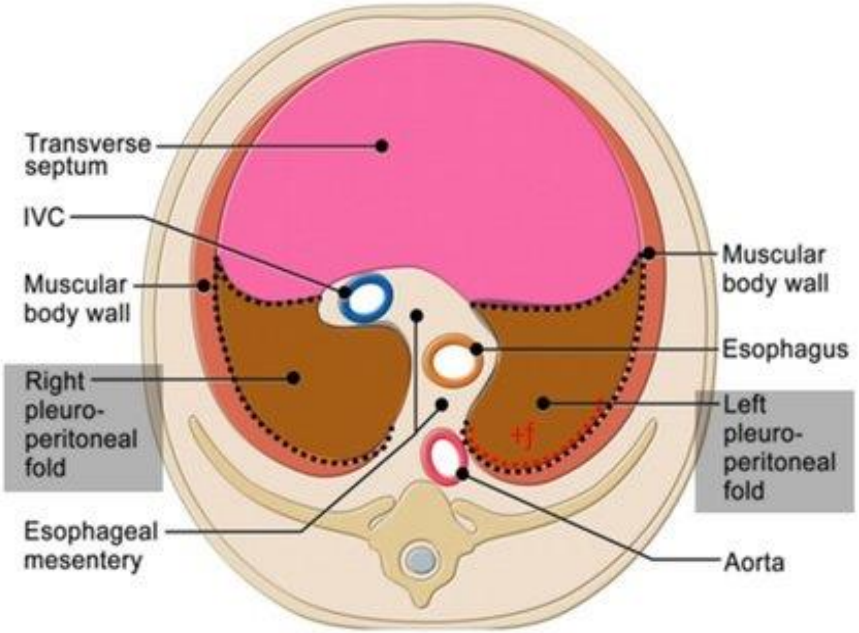
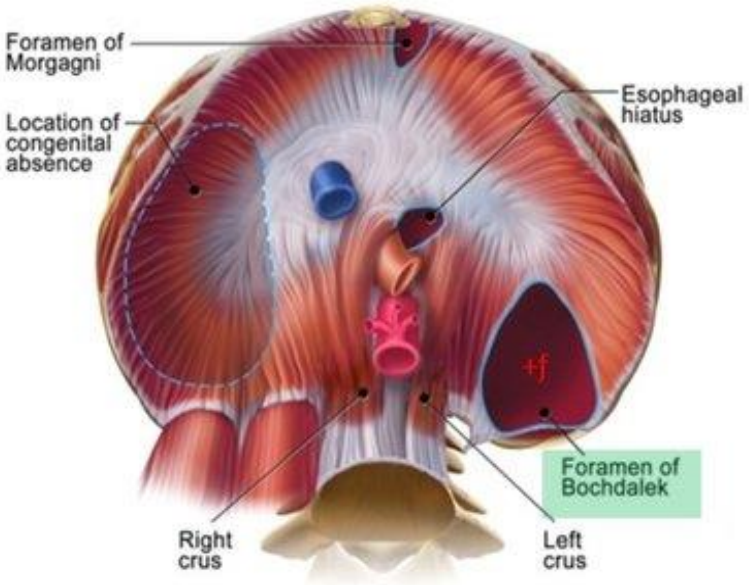
Congenital Posterolateral Diaphragmatic Hernia (CDH)

- ☺One of most severe conditions of neonate
- ☺Defect in diaphragm during early fetal development
- ☺ left side most commonly affected
- ☺content of the hernia:
  - small bowel
  - colon
  - spleen
  - stomach
  - liver, kidney, tail of pancreatic



The Bochdalek hernia accounts for up to 90% of the hernias seen, with 80–90% occurring on the left side.
The Morgagni hernia accounts for 2–6% of CDH.
Occasionally bilateral (<5%).


Transverse septum
IVC
Muscular body wall
Right pleuro-peritoneal fold
Esophageal mesentery
Muscular body wall
Esophagus
Left pleuro-peritoneal fold
Aorta


Foramen of Morgagni
Location of congenital absence
Esophageal hiatus
Foramen of Bochdalek
Right crus
Left crus

【pathophysiology】

1、Hypoplasia of the lung

characterized by a reduction in pulmonary mass and the number of bronchial divisions, respiratory bronchioles, and alveoli.

2、Pulmonary hypertension

Abdominal viscera into the thoracic cavity → compression of the lung, PaO_2↓$PaCO_2$↑→ acidosis, hypoxemia（PH<7.30）→pulmonary vessels spasm →vessel resistance↑, right to left shunting through patent ductus arteries and foramen ovale↑→aggravate acidosis and hypoxemia in the body circulation (fetal circulation syndrome)

diaphragmatic hernia

Clinical manifestations：

1，Severe respiratory distress，cyanosis, vomit

2，Breath sounds: diminished on the side of hernia

3，Heart sounds: deviated to the contralateral chest

4，Scaphoid abdomen

Associated anomalies – in up to 30% of cases,:

CNS lesions,

Esophageal atresia,

Omphalocele, and

Cardiovascular lesions.

It is also recognized in :

trisomies 21, 13, and 18 and Turner syndromes.

【 diagnosis 】

Prenatal diagnosis

ultrasound: abdominal organ visible in the fetal chest diagnose 50% of cases.

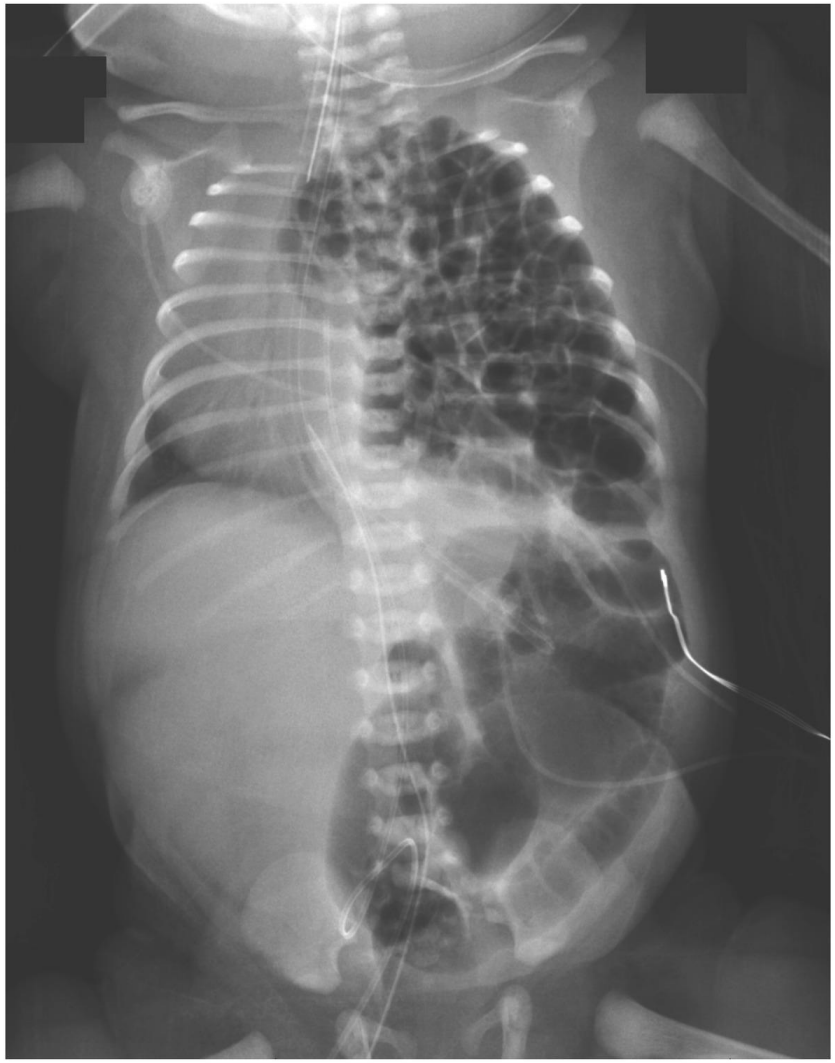
polyhydramnios, chest mass, mediastinal shift, and gastric bubble or a liver in the thoracic cavity.

High-speed fetal MRI

diagnosis after birth

X-ray film:

- Typical air-filled stomach and bowels in the chest, which continues into the abdominal cavity.
- Diaphram can not be seen at the affected side.
- Absence or scarcity of intestine in the abdominal cavity



Treatment

- **Before delivery: cortisone could induce the maturation of pulmonary tissue**
- **Preoperative preparation:**

（1）mechanical ventilation with pure oxygen

（2）nasogastric tube to decompress stomach and intestine

（3）semi-supine and inclined to the ipsilateral side， keep warm

（4） i.v. fuild, correction of acidosis

(5）surgical repair

Congenital Esophageal Atresia Tracheoesophageal Fistula

- ***Incidence: 1/3000***
- ***associated anomalies common***
- ***Impediment of recanalization and interruption of septation of trachea and esophagus***

VACTERL syndrome

Vertebral

Anorectal

Cardiac

Tracheal

Esophageal

Renal

radial, (**Limb**)

Cardiac and vertebral anomalies are seen in **32%** and **24%**, respectively.

CHARGE syndrome

Coloboma of the eye ***C***entral nervous system anomalies

Heart defects

Atresia of the choanae

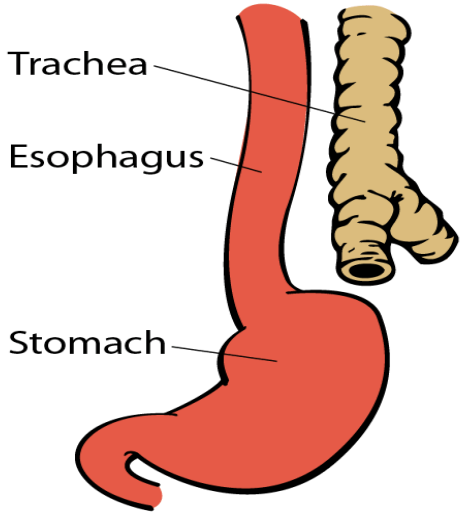
Retardation of growth and/or development

Genital and/or urinary defects [hypogonadism]

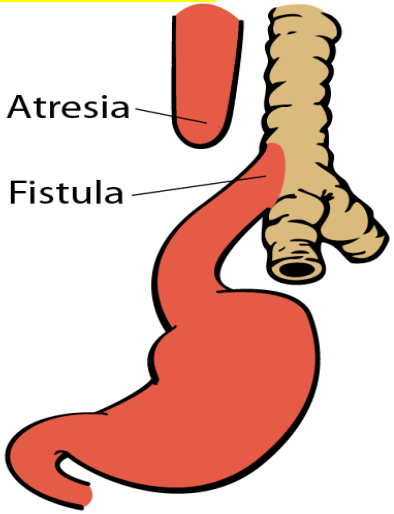
Ear anomalies and/or deafness

CHD and anophthalmia–esophageal–genital syndrome.

%85 A

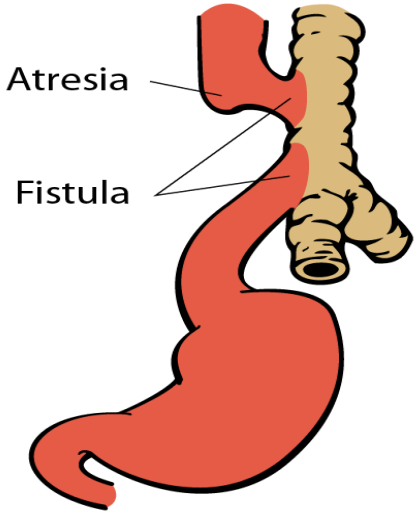


Normal Anatomy

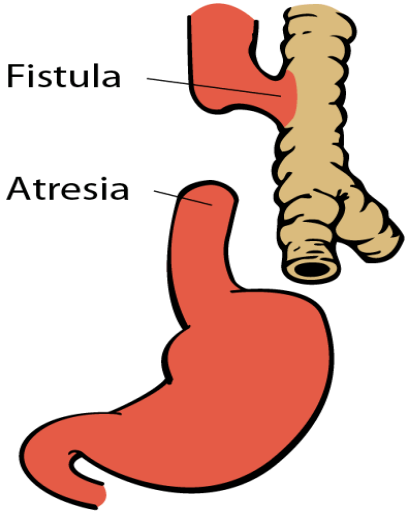


Atresia with distal Fistula

1.4% - E

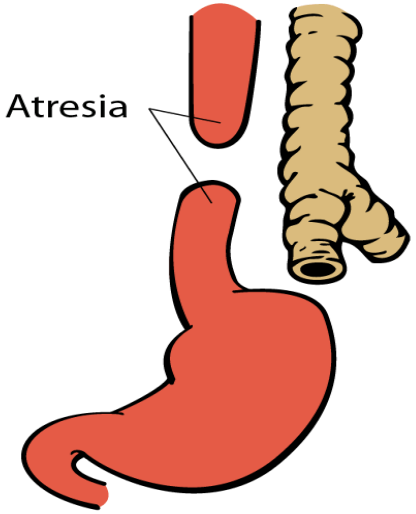


Atresia with double Fistula



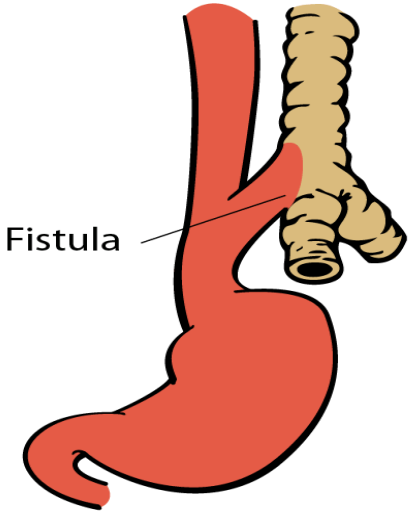
Atresia with proximal Fistula

0.6%- D



Atresia

% 8 - B



Fistula

C - %4

【clinical findings】

1、**drooling saliva, unable to swallow**

2、**cough and choke and may become cyanotic after feeding**

3、**chemical and aspiration pneumonia**

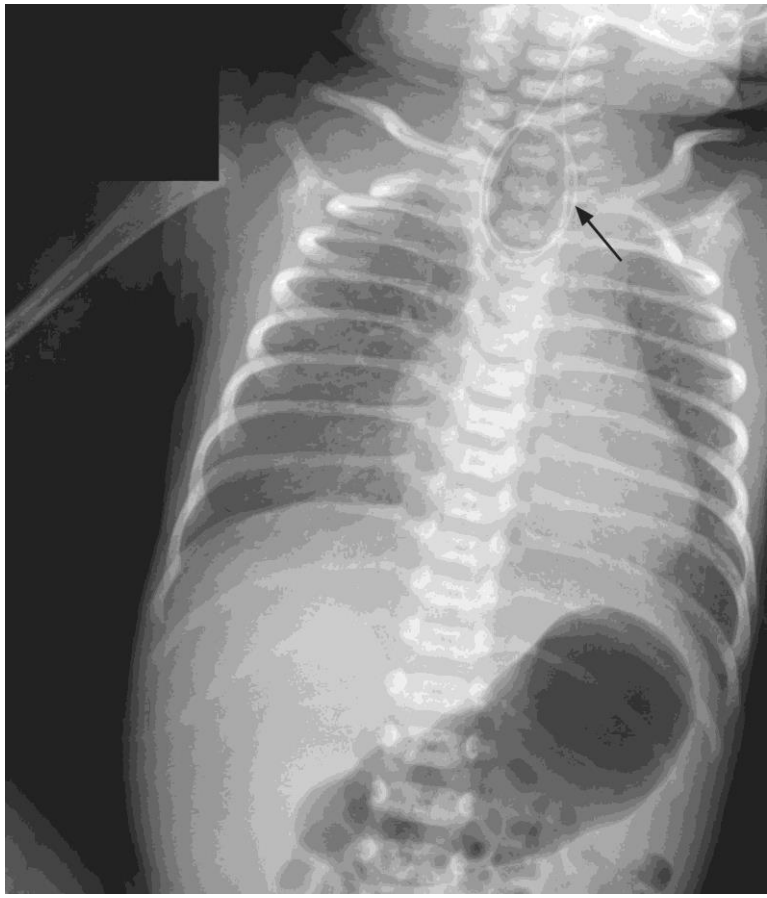
4、**abdominal distention or scaphoid abdomen**

【 diagnosis 】

1、**prenatal diagnosis**

2、 **nasogastric tube can not reach stomach.**

3、 **X-ray film show the coiling of the tube in the upper mediastinum**



Preoperative preparation

- supine and elevated to 30～40°
- Catheter was put at the blind end of the esophagus for continuous drainage
- oxygen inhalation, incubator
- i.v. Fluid.
- surgical repair
- Prognosis: 98%~100% survival rate..

Prone positioning minimizes movement of gastric secretions into a distal fistula, and esophageal suctioning minimizes aspiration from a blind pouch.

Surgical ligation of the TEF and primary end-to-end anastomosis of the esophagus via right-sided thoracotomy constitute the current standard surgical approach.

If the gap between the atretic ends of the esophagus is >3 to 4 cm (>3 vertebral bodies), primary repair cannot be done; options include using gastric, jejunal, or colonic segments interposed as a neoesophagus.

Careful search must be undertaken for the common associated cardiac and other anomalies.

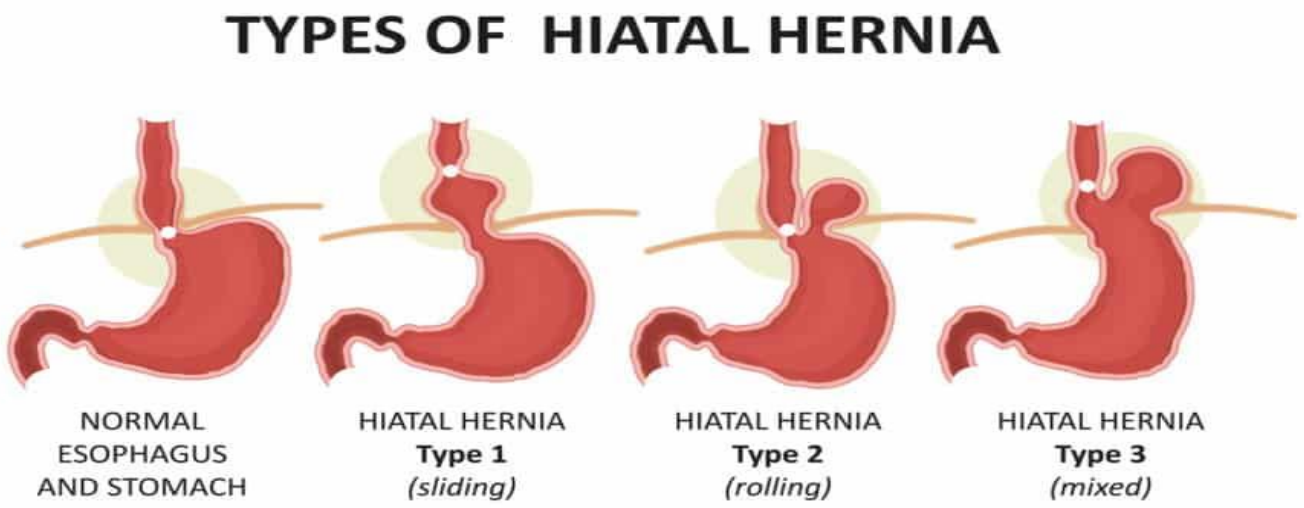
Hiatus hernia

Type 1 common sliding hernia : in which the gastroesophageal junction slides into the thorax.Often associated with GER.

Type 2 paraesophageal : in which a portion of the stomach (usually the fundus) is insinuated next to the esophagus inside the gastroesophageal junction in the hiatus .

Type 3 A combination of sliding and paraesophageal types (type 3)

Diagnosis usually made by an upper gastrointestinal series and upper endoscopy.



Hypertrophic Pyloric Stenosis

【pathophsiology】

1、olive shaped mass：length 2～3.5cm，thickness 0.4～0.6cm，pale in color with consistency of cartilage

2、Muscular hypertrophy of all the layers of the pylorus , most significant in the circular layer, causing the stenosis

Hypertrophic Pyloric Stenosis

【symptoms】

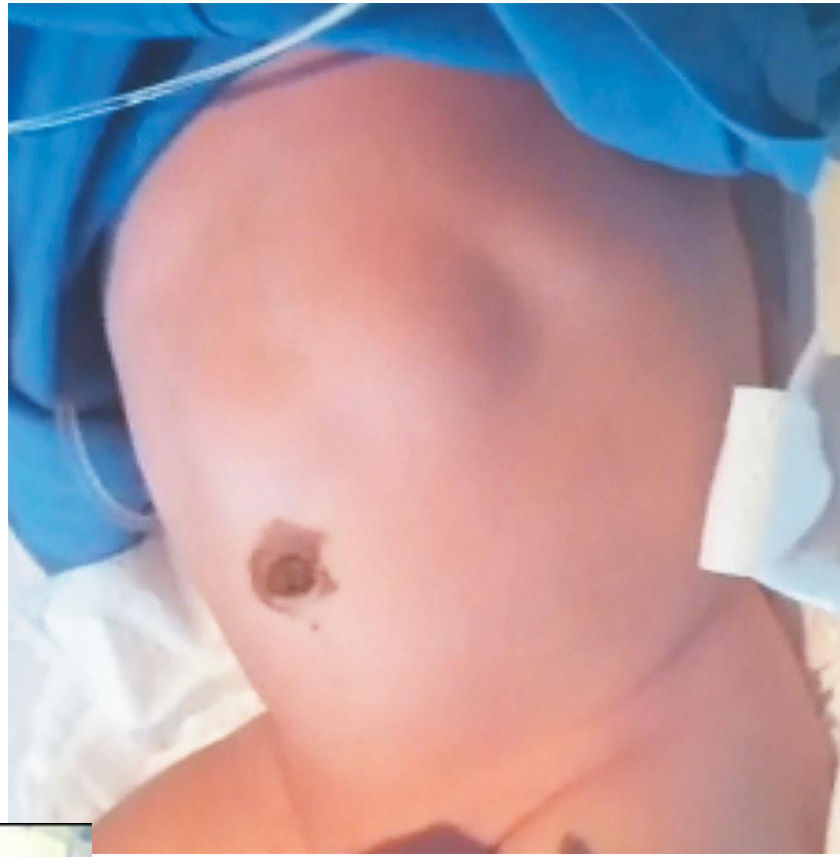
1、 projectile vomiting: onset:2-3 weeks after birth and progressive with time; vomitus: non-bilious milk and milk curds

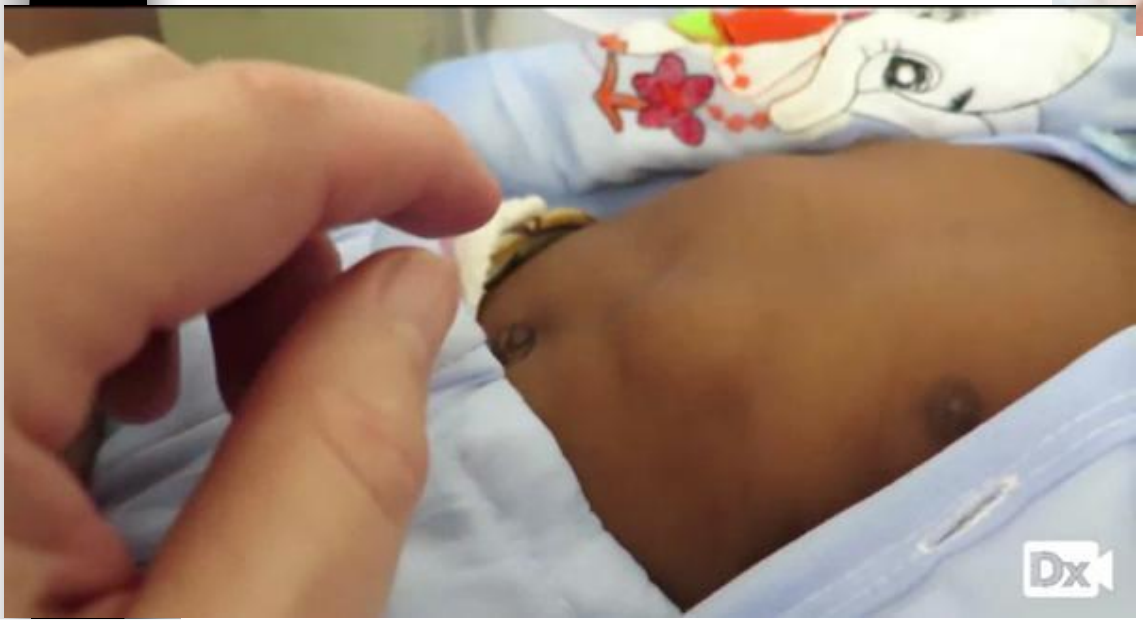
2、jaundice ： deficiency in liver enzyme and compression of the biliary tract

3、 overall condition: dehydration, weight lose, hypo-chloride metabolic alkalosis, oliguria

4、abdominal examination: distention of epigastrium, visible gastric waves, presence of a palpable pyloric tumor (unique physical sign)

Visible gastric waves in P.Stenosis



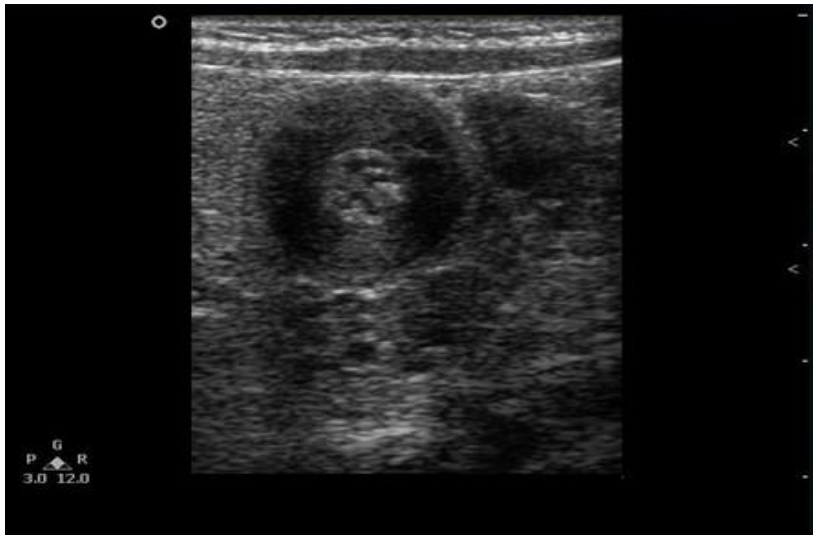


【Diagnosis】

1、typical vomiting and mass in the epigastrium

2、ultrsound: muscular thickness≥0.4cm,
SD=thickness×2/diameter≥50% (Target sign)

3、GI for cases with difficulty in diagnosis:
①distention of the stomach
②strong gastric waves
③elongated and narrow pyloric channel
④delay in stomach emptying



Treatment

- Correction of dehydration.
- Correction of acid base imbalance.
- Correction of the electrolyte imbalance.

(Hypokalemic hypochloremic metabolic alkalosis)

Surgical Correction

Ramstedt pyloromyotomy

Intestinal Atresia and Stenosis

【Clinical findings】

1、vomiting

onset: from first time of feeding
to a few days after birth
vomitus: bilious or feculent

2、abdominal distention

high: confined to epigastrium
low: full abdomen distention

3、failure to pass meconium:

normally meconium was passed within the first 24hrs of life and cleared in 2-3 days.

4、General condition

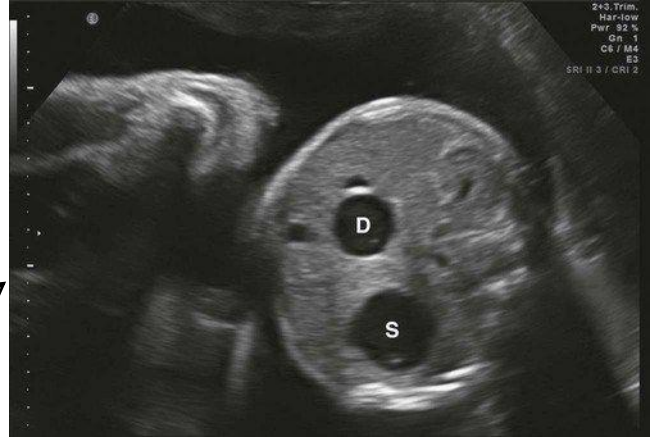
In Duodenal atresia

- 50% of infants are premature.
- Concomitant congenital anomalies are common:

✓ congenital heart disease (30%)
✓ malrotation (20–30%)
✓ annular pancreas (30%)
✓ renal anomalies (5–15%)
✓ esophageal atresia with or without tracheoesophageal fistula (5–10%)
✓ skeletal malformations (5%)
✓ anorectal anomalies (5%).

trisomy 21 is identified in up to one-third of patients.

【 Diagnosis 】



1、**prenatal ultrasound** → sonographic double-bubble

2、**Clinical findings:**

bilious vomiting 24-48hrs after birth

abdominal distention

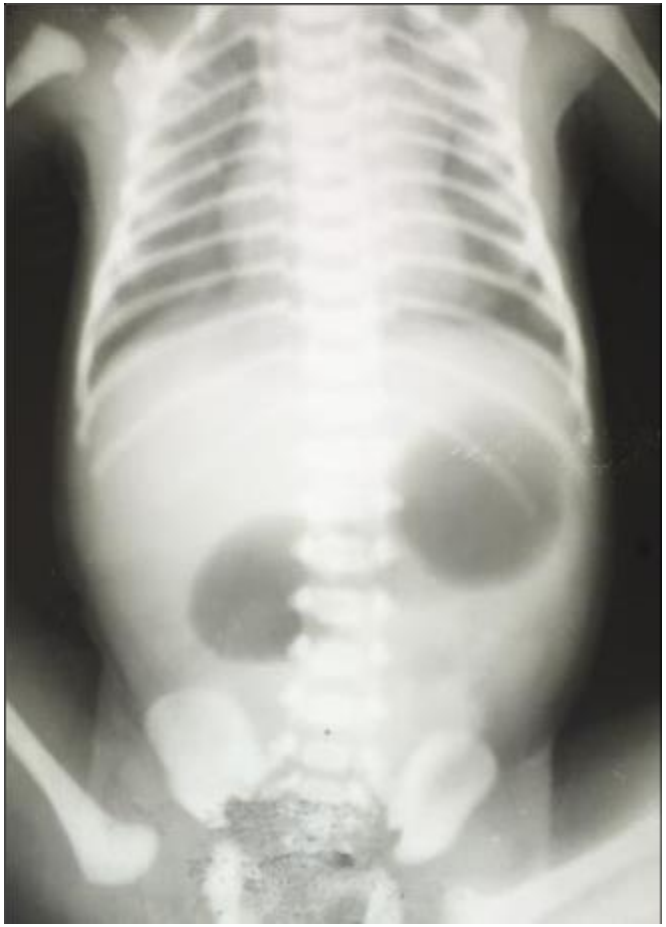
failure to pass meconium

3、**X-ray：**

duodenal atresia——Double bubble sign

jejunal atresia——triple bubble sign

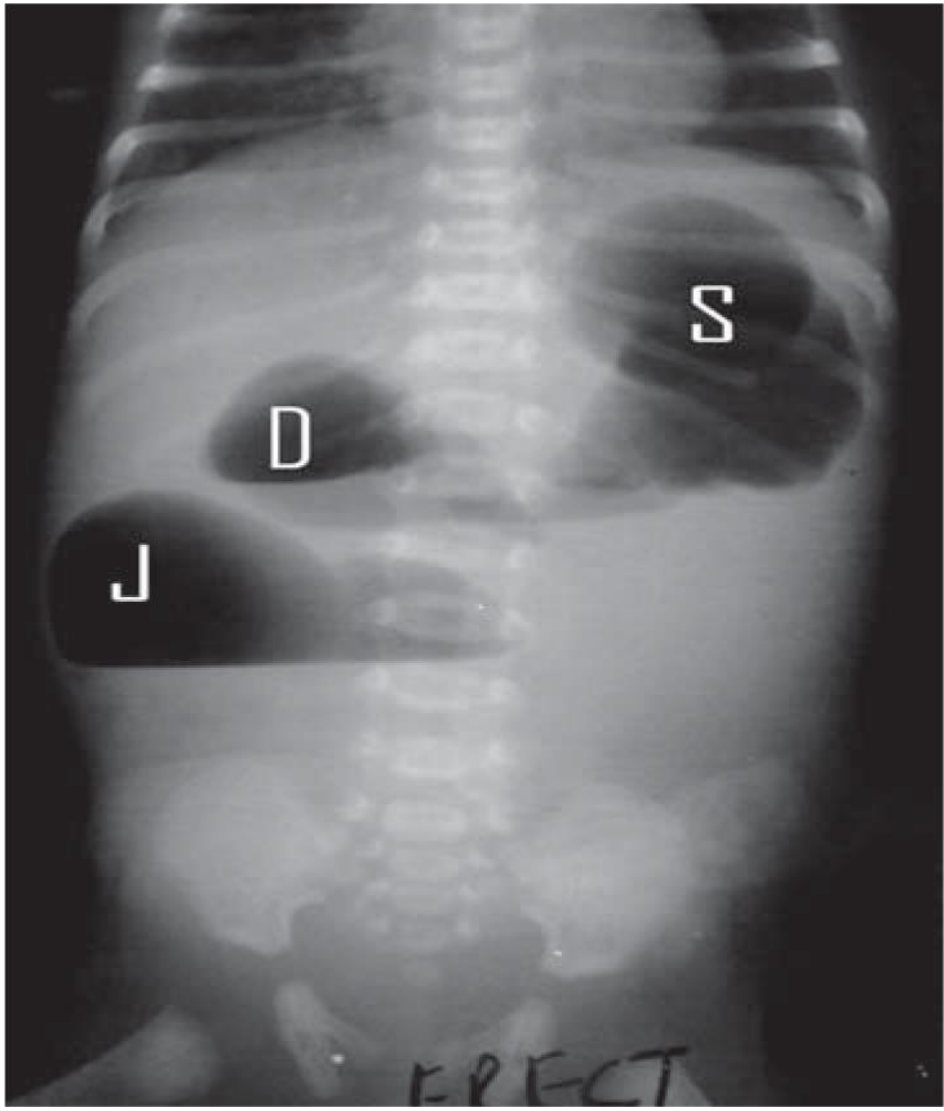
low intestinal atresia——multiple air-fluid level

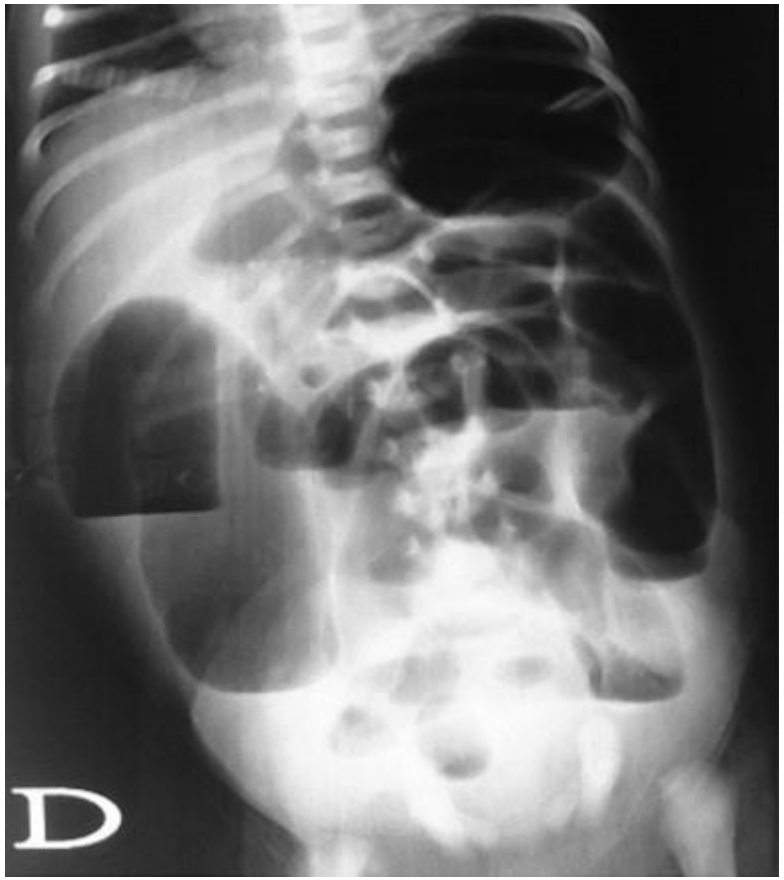


SUPINE ABDOMINAL RADIOGRAPH OF A NEWBORN PRESENTING WITH BILLOUS VOMITING SHOWS A DISTENDED STOMACH AND DUODENAL AIR BUBBLES "DOUBLE BUBBLE" SIGN. THERE IS NO GAS DISTAL TO THE DUODENAL BUBBLE.

The **triple bubble sign:**

The classic radiographic appearance observed in jejunal atresia .





Low intestinal atresia

【 Treatment 】

The only option is surgery：

intestinal septum excision

Intestine resection and anastomosis

Congenital Malrotation of Intestine

【Definition】

Malrotation is the term used to define the group of congenital anomalies resulting from aberrant intestinal rotation and fixation

【Embryology】

Week 6~8: Herniation of midgut into the umbilical cord with a 180 degree of counterclockwise rotation along the axis of superior mesenteric artery

Week 10: Return to the abdominal cavity with a final 90 degree of rotation to complete the 270-degree counterclockwise rotation

【 Pathology 】

Nonrotation and Incomplete rotation: abnormal positioning of the proximal small bowel and the cecum

Duodenum compressed by abnormal peritoneal band(Ladd's band): high incomplete extrinsic obstruction

Midgut volvulus: torsion of the narrow mesenteric pedicle produces an acute closed-loop intestinal obstruction and vascular insufficiency.

Proximal jejunum fused to the ascending colon by anomalous peritoneal attachments

Malrotation

- **Pathology：**

Compression of duodenum

Kinked and foreshortened proximal jejunum by peritoneal band

Midgut volvulus

【Clinical manifestations】

Emesis： **bilious, intermittent， occur at 3-5 days after birth or asymptomatic**

Abdominal distention: **confined in epigastrium, diffuse to the full abdomen in bowel necrosis**

Stool: **normal meconium, bloody stool suggests volvulus and necrosis**

Newborn: **normal meconium， intermittent vomiting after 3-5 days of birth， no abdominal distention, hard stool**

Children and infant： **asymptomatic since birth, intermittent onset or sudden onset of volvulus**

Clinical manifestations

- **Symptoms of volvulus: bloody vomitus and stool, abdominal tenderness**

【X-ray film】

1、Plain X-ray film: double-bubble sign

2、barium enema: cecum in the upper or left abdomen

3、GI： incomplete duodenal obstruction; ligament of Treitz not to the left of the midline; abnormal position of the proximal jejunal loops to the right of the midline

Treatment

Principles：

Asymptomatic malrotation

- most recommend surgical treatment
- some believe operation only necessary in young children

High intestinal obstruction

- operated on promptly, but not necessarily emergently

Volulus with sign of bowel necrosis

- immediate operation

Treatment

Ladd's operation

- All volvulus is clockwise so the small bowel must be rotated in a counterclockwise fashion
- Expose duodenum by division of the Ladd's bands
- Dissection additional peritoneal bands to convert the mesenteric pedicle to a wide plane
- Alignment of small bowel to the right and colon to the left of the abdominal cavity

Hirschsprung's Disease

Anatomy

1、**distended segment: proxiaml colon enlarged with muscular hypertrophy**

2、**stenosis segment：distal colon spasm**

3、**transitional segment：between distal and proximal segments**

Histology

1、lack of ganglion cell in the neural plexus of the affected segment of intestine

2、hypertrophied nerve trunk stain positive for acetylcholinesterase

3、Disarray of adrenergic fibers

【Pathophysiology】

Arrest of cranial to caudal migration of neuroblasts derived from neural crest precursors along the intestinal tract with vagal nerve fiber at 6-12 weeks of gestation, which results in aganglionosis of the distal bowel.

1、spasm of affected segment
no normal peristalsis

2、internal sphincter spasm
no normal defecation reflex

3、proximal bowel distended with histologic evidence of muscular hypertrophy

【clinical findings】

Neonate：

1、emesis： bilious or feculent

2、abdominal distention

3、delayed passage of meconium

4、rectal examination： tightness of internal sphincter,rectal emptiness， withdraw brings out meconium and gas

5、after bowel irrigation, temporary subsiding of the symptoms

Children and infant：

1、History of neonate constipation

2、Malnutrition , anemia

3、Chronic abdominal distention

【Diagnosis】

Barium enema

Demonstration of a spasmodic distal intestinal segment with dilated proximal bowel

Failure to evacuate barium from colon within 24hours

simplicity of the method

accuracy in neonate 80%

not suitable for short segment type

Anorectal Manometry

- **Aid diagnosis through identification of the rectoanal inhibitory reflex which is absent in the vast majority of children with Hirschsprung's disease**
- **Drawbacks: false-positive in older children due to masking of the relaxation response by contraction of the external sphincter**

Rectal biopsy (Definitive diagnosis)

- Suction biopsy
  - Biopsy taken at 1-2cm above the dentate line
  - looking for the presence or absence of ganglion cells and hypertrophied nerve trunks
  - simplicity, accuracy, absence of complications
  - False-negative (age, mucosal edema, tissue quality, experience)

Histologic staining of mucosa

Increased AChE content in the nerve fibers of the lamina propria and muscularis mucosae

Full-thickness Rectal Biopsy

Complexity and complications, possible effect on future definitive surgery

【Complications】

1、**Enterocolitis**

Most frequently encountered and life-threatening:

from *constipation to diarrhea*

peritonitis and sepsis

fever and abdominal distention

digital exam：massive amount of odorant stool

2、**Perforation：**

Cecum peforation

Bowel necrosis and perforation

【Treatment】

1、**Colon irrigations:**

isotonic fluid

one to two times a day

＃Do not use tape water

2、Colostomy：

3、Surgical options

Swenson

Duhamal

Rehbein

Soave

Neonatal surgery

Laparoscope's

Transanal pullthrough

Intussusception

the most common cause of intestinal obstruction in children under 2 years of age.

1 portion of proximal intestine (intussusceptum) telescopes into a more distal portion (intussuscipiens).

Once this prolapse has occurred, lymphatic and venous congestion develops, resulting in edema, strangulation, ischemia, and ultimately necrosis.

Additionally, the lumen of the intussuscepted portion of the bowel collapses, causing intestinal obstruction.

Intussusception is fatal if spontaneous reduction does not occur and it is left untreated; therefore, prompt diagnosis and treatment are critical for successful management.

Intussusception

PATHOGENESIS AND EPIDEMIOLOGY

uncommon prior to 3 months of age, and children ages 3 months to 3 years are most commonly affected. It peaks between 5 and 7 months old.

incidence : 75 cases per 100,000 in the first year of life.

Boys : girls 2: 1.

idiopathic : 90% of cases.

ileocolic .

Mechanism: extra mucosal lead point such as Peyer's patch hypertrophy or mesenteric lymphadenitis.

Secondery:

Viral gastroenteritis (most commonly adenovirus), Henoch–Schönlein purpura, intestinal lymphoid hyperplasia , Meckel diverticulum, intestinal polyps, intestinal duplication, hemangioma, tumors, and ectopic pancreas.. !!!! **recurrent intussusceptions.**

An association between intussusception and the tetravalent live attenuated rotavirus vaccine was identified in 1999.

classic triad

intermittent, crampy abdominal pain.

palpable abdominal mass.

“currant jelly” stools.

Abdominal pain :

sudden onset manifest as sudden crying, with flexed hips and knees.

normal between these episodes of pain. Lethargy and pallor eventually develop.

Over time, signs of obstruction develop, including bilious emesis and abdominal distention.

a sausage-like mass is felt on palpation.

“currant jelly stools”

Due to mucosal ischemia It is bloody sloughing mucosa. It is a **late** sign .

Ultimately, perforation occur focal or diffuse peritonitis .

Investigations

Abdominal radiograph.

limited capacity to confirm the diagnosis.

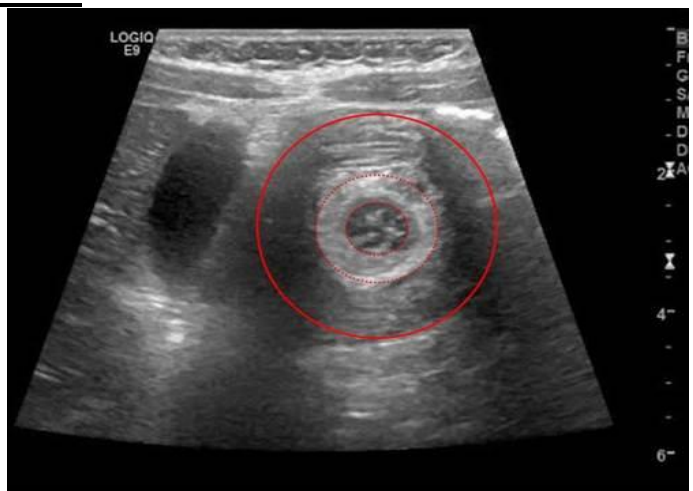
findings suggestive of an intussusception

proximal bowel dilation , distal air outlining the intussusceptum, or pneumoperitoneum, , may be evident in 74% of cases.

Abdominal ultrasound

diagnostic accuracy of approximately 85%. **"target" sign.**

A target sign may also be visible on **CT scan.**



treatment

IV fluids and exclude pneumopertitonium.

Radiographic enema reduction using water soluble contrast or air and guided by USS or Fluroscopy.

Bowel perforation (< 1%),

Recurrence Following successful reduction in 8% to 15% of children within 6 months with one-third occurring within the first 24 hours.

Operative intervention

- failed enema reduction.
- perforation, peritonitis, or hemodynamic instability, or if a pathologic lead point is identified. Example : Meckle's diverticulum.

Types of surgery

- Manual reduction.
- Bowel resection

bowel ischemia, pathologic lead point, or inability to manually reduce the prolapsed segment.

Inguinal hernia

- **Direct 1 %**

results from muscle weakness in the floor of the inguinal canal (internal oblique and transversus abdominis muscles).

- **indirect.** In 99 %.

  secondary to a patent processus vaginalis.

The processus vaginalis accompanies the testis on its descent from the retroperitoneum into the scrotum, and normally obliterates by term.

In girls, the processus vaginalis accompanies the round ligament into the labia majora.

Meckel diverticulum

Most common congenital anomaly of the GI tract occur in 2–3% of all infants. .

Caused by the incomplete obliteration of the omphalomesenteric duct during the 7th wk of gestation.

Partial or complete failure of involution of the omphalomesenteric duct results in various residual structures. And Meckel diverticulum is the most common of these structures .

A typical Meckel diverticulum is a 3-6 cm outpouching of the ileum along the antimesenteric border 50-75 cm (approximately 2 feet) from the ileocecal valve

Classic presentation： rule of 2s

- found in approximately **2%** of the general population.
- usually located **2 feet** proximal to the ileocecal valve.
- approximately **2 inches** in length.
- can contain **2 types** of ectopic tissue (pancreatic or gastric).
- generally present before the **age of 2 yr.**
- found **twice** as commonly in females.

Meckel diverticula are lined by an ectopic mucosa,which is most commonly of gastric origin, but it can also be pancreatic, jejunal, or a combination of these tissues.

The acid-secreting mucosa causes intermittent painless rectal bleeding by ulceration of the adjacent normal ileal mucosa.

Anemia. Current jelly colored stool. Melena.

Intestinal obstruction : the diverticulum may acts as the lead point of an intussusception.

Diverticulitis inflammed Meckel manifests like acute appendicitis.

Diagnosis

The most sensitive study is a Meckel radionuclide scan, which is performed after intravenous infusion of technetium-99m pertechnetate. The mucus-secreting cells of the ectopic gastric mucosa take up pertechnetate, permitting visualization of the Meckel diverticulum.

Routine radiological study and barium study rarely fill the diverticulum.

Treatment

Symptomatic Meckel diverticulum is surgical excision.

A diverticulectomy can be performed safely as either a laparoscopic or open procedure.

Acute appendicitis

- peak incidence : second decade.
- Perforation rates have remained around 40% .
- Appendiceal ultrasound is highly sensitive for diagnosis.
- <50% of cases in children have the classic presentation.

Progressive Abd pain – Tenderness – Rebound tenderness - Gaurding - dysuria

- Children early in the disease can appear well and demonstrate mild symptoms, minimal findings on physical examination, and normal laboratory studies.
- Perforation and advanced peritonitis : severe illness with bowel obstruction, renal failure, and septic shock.

Treatment : hydration – Appendectomy

Thank you !