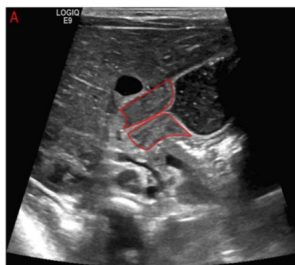
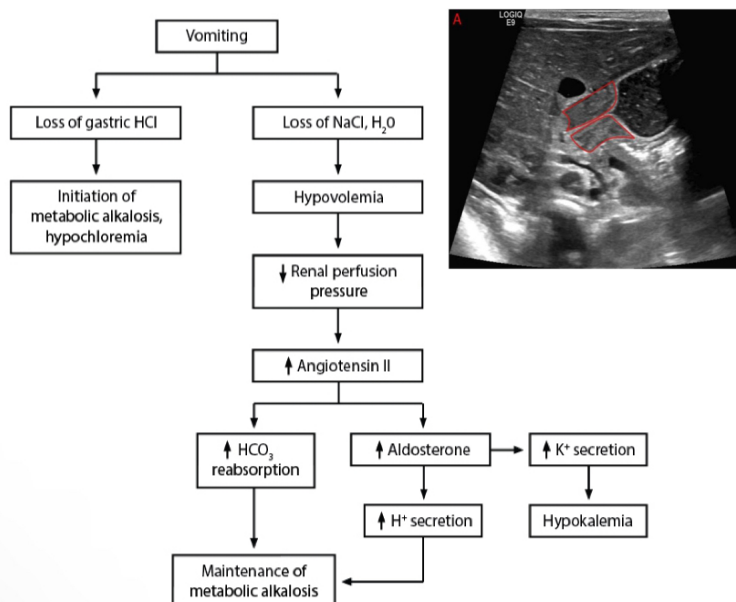


IV. Intestinal Anatomic Obstructions That Result in Vomiting

A. Hypertrophic pyloric stenosis

1. **Definition.** Thickening of the circular pylorus muscle results in gastric outlet obstruction with projectile vomiting.
2. **Epidemiology**
 - a. Incidence is 2–3.5 in 1000 live births.
 - b. **Caucasians and first-born male children are affected more commonly.** The male-to-female ratio is 4:1.
3. **Etiology**
 - a. The **etiology is unknown.** Abnormal muscle innervation, breastfeeding, early administration of oral erythromycin, and decreased production of nitric oxide synthase have all been implicated.
 - b. Associated conditions include duodenal atresia, tracheoesophageal fistula, trisomy 18, and Cornelia de Lange syndrome.
4. **Clinical features**
 - a. *Vomiting of nonbilious*, milky fluid starts during the second or third week of life.
 - b. Vomiting becomes more forceful, is often described as **projectile**, and typically occurs immediately after feeding.
 - c. Affected infants may be **irritable but hungry**.
 - d. Jaundice occurs in 5% of patients, and it is associated with low levels of the enzyme glucuronyl transferase.
 - e. *Dehydration and electrolyte abnormalities from vomiting may occur.*
5. **Evaluation**
 - a. **Physical examination**
 1. The hypertrophied pylorus muscle may be palpable just above and to the right of the umbilicus. It is often referred to as the **“olive.”**
 2. Abdominal peristaltic waves may be visible after feeding.
 - b. Prolonged vomiting of gastric contents classically results in **hypochloremic, hypokalemic, metabolic alkalosis.**
 - c. **Diagnostic studies**
 1. **Ultrasound** is used to measure the pylorus muscle length and thickness and is the **diagnostic method of choice.**
 2. **UGI study** may demonstrate an elongated, narrow pyloric channel (**“string sign”**).
6. **Management**
 - a. Electrolyte abnormalities and dehydration should be corrected before surgery.
 - b. Surgical correction is with a **partial pyloromyotomy**, in which the circular pylorus muscle fibers are transected.

Laboratory derangements in pyloric stenosis



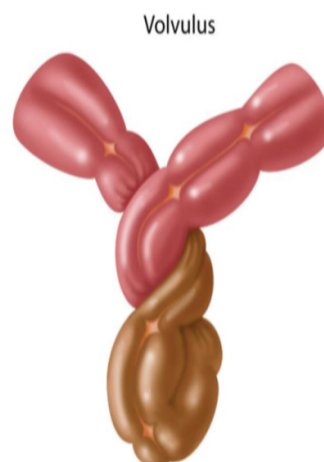
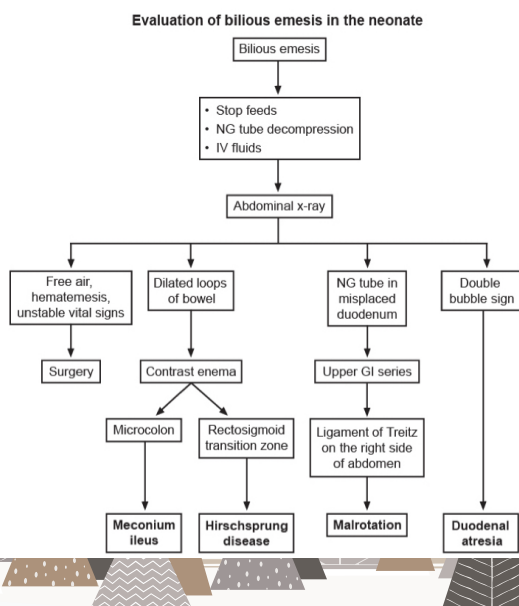
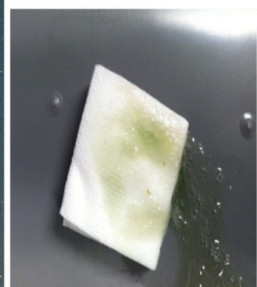
Infantile hypertrophic pyloric stenosis

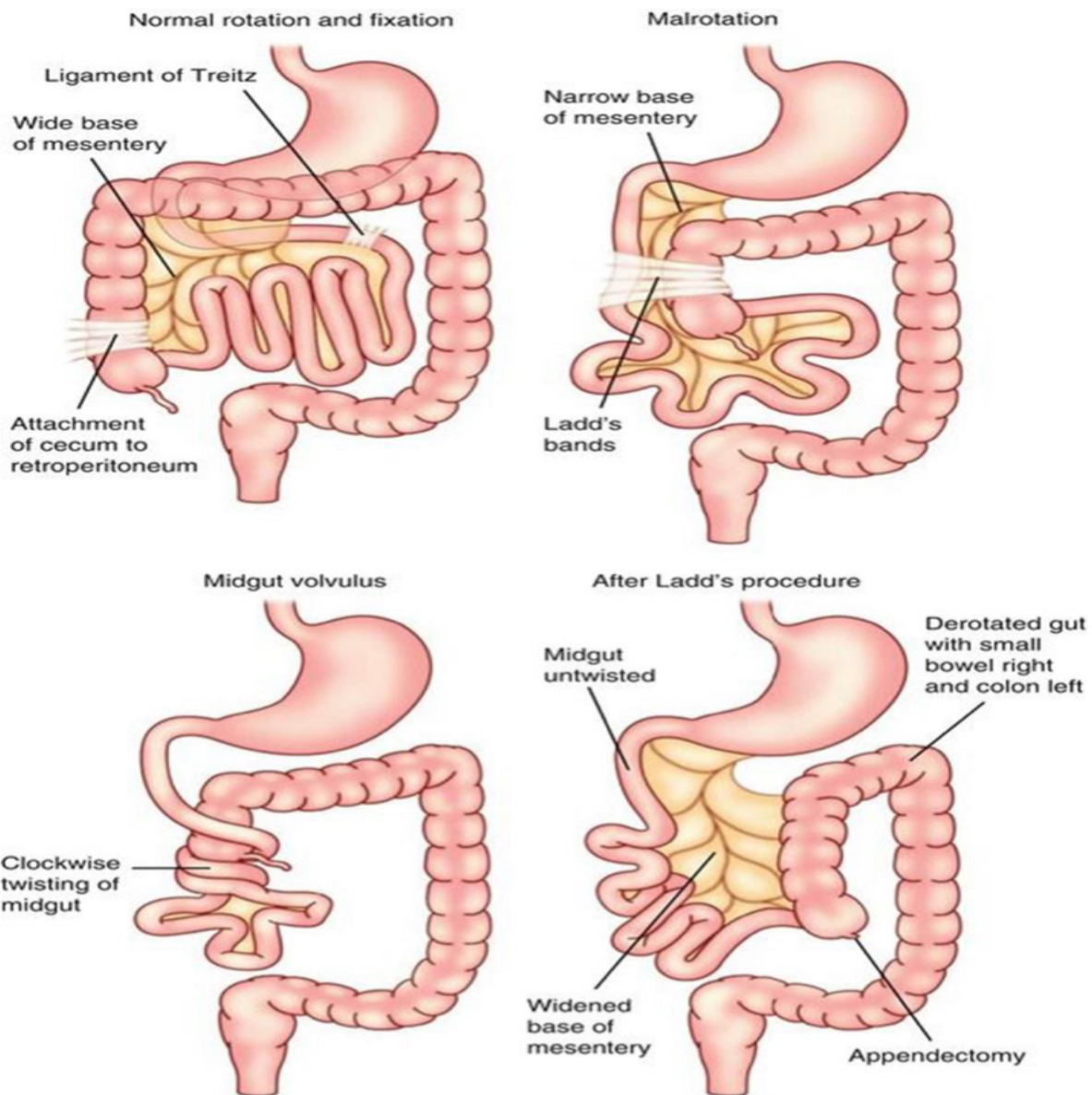
Risk factors	• First-born boy • Erythromycin • Bottle feeding
Presentation	• Projectile nonbilious emesis • Poor weight gain • Dehydration • Olive-shaped abdominal mass
Diagnostic studies	• Hypochloremic metabolic alkalosis • Thickened pylorus on abdominal ultrasound
Treatment	• Intravenous rehydration • Pyloromyotomy

muscle fibers are transected.

B. Malrotation and midgut volvulus

1. **Definition.** Malrotation is an anatomic abnormality of intestinal rotation that allows the **midgut to twist around the superior mesenteric vessels (i.e., volvulus)**. This may obstruct and infarct the bowel.
2. **Epidemiology**
 - a. Incidence of malrotation is 1 in 500 live births, with a **2:1 male predominance**.
 - b. Malrotation is common in patients with heterotaxy, a condition in which the internal organs are abnormally arranged in the chest and abdomen, and may be associated with small bowel atresia, Hirschsprung disease, and intussusception.
3. **Pathogenesis.** The intestines normally return to the abdomen through the umbilical cord during the 10th week of gestation and undergo counterclockwise rotation about the axis of the superior mesenteric artery. The intestines are then fixed to the abdominal wall. Malrotation occurs when normal bowel rotation is interrupted.
 - a. Lack of fixation of the small bowel results in **peritoneal bands (Ladd bands)** that can compress the duodenum, causing mechanical obstruction.
 - b. The narrow pedicle, which suspends the small bowel and the base of the superior mesenteric vessels, can easily twist, leading to ischemia and midgut volvulus.
4. **Clinical features**
 - a. **Bilious vomiting and sudden onset of abdominal pain in an otherwise healthy infant** is the classic presentation. Older children may have intermittent, crampy abdominal pain and vomiting.
 - b. Anorexia, distension, and blood-tinged stools are common.
 - c. Physical examination may initially be normal with peritonitis occurring later. Shock and cardiovascular collapse may occur as bowel ischemia progresses.
5. **Evaluation**
 - a. **Abdominal radiographs** may show gastric or proximal intestinal distension and obstruction, often with little or no distal bowel gas.
 - b. **Upper intestinal contrast imaging** (UGI study with small bowel follow-through) is the **diagnostic tool of choice**, and demonstrates the abnormal position of the ligament of Treitz to the right of midline, partial or complete duodenal obstruction, and the jejunum to the right of midline.
 - c. **Lower intestinal contrast studies** may reveal the cecum in the left abdomen or right upper quadrant.
6. **Management**
 - a. Volvulus is a **surgical emergency** requiring immediate exploration, untwisting of the gut, resection of any nonviable segments of intestine, and fixation of the gut to prevent recurrence.
 - b. **Fluid resuscitation**, nasogastric suctioning, and broad-spectrum parenteral antibiotics should be administered.
 - c. TPN may be required if a large segment of the bowel is resected.





C. Atresias

1. Duodenal atresia and stenosis

- Definition.** Duodenal atresia is a congenital obstruction of the duodenum caused by failure of the lumen to recanalize at 8–10 weeks' gestation.
- Epidemiology**
 - Intestinal atresia is the **most common cause of obstruction in the neonatal period**.
 - Incidence is approximately 1 in 10,000 live births, with a **male predominance**.
 - One-fourth of cases occur in patients with **Down syndrome**.
- Clinical features**
 - Diagnosis may be suspected if prenatal ultrasound demonstrates gastric dilation with **polyhydramnios**.
 - At birth, physical examination of patients with duodenal atresia may reveal a **scaphoid abdomen with epigastric distension**. Infants have feeding intolerance and vomiting.
 - Duodenal stenosis may present with emesis, weight loss, and FTT.
 - Other congenital defects (e.g., malrotation, esophageal atresia, congenital heart disease, renal anomalies) may also be present.
- Evaluation**
 - Abdominal radiography** shows air in the stomach and proximal duodenum,

creating a “double bubble” sign in infants with atresia.

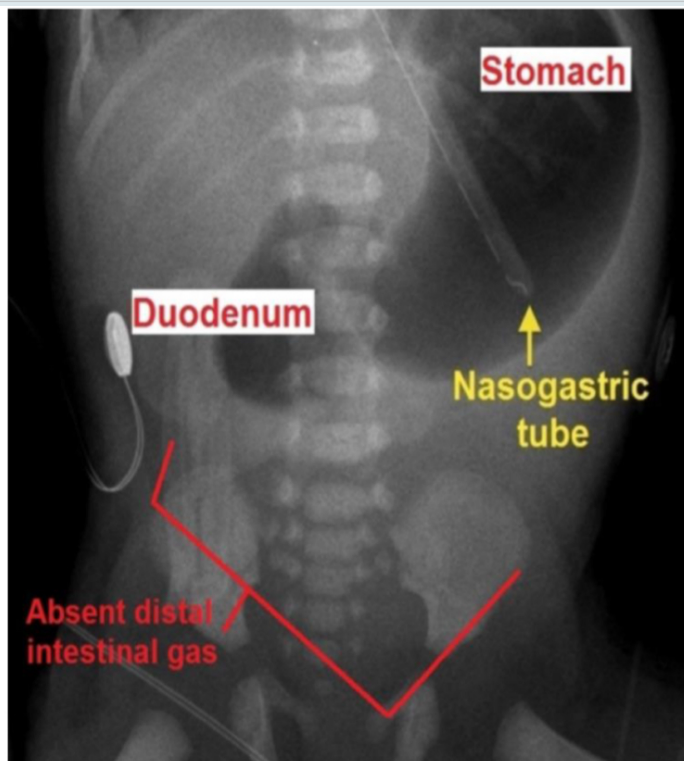
2. UGI study is very effective at diagnosing atresia and stenosis.

e. **Management**

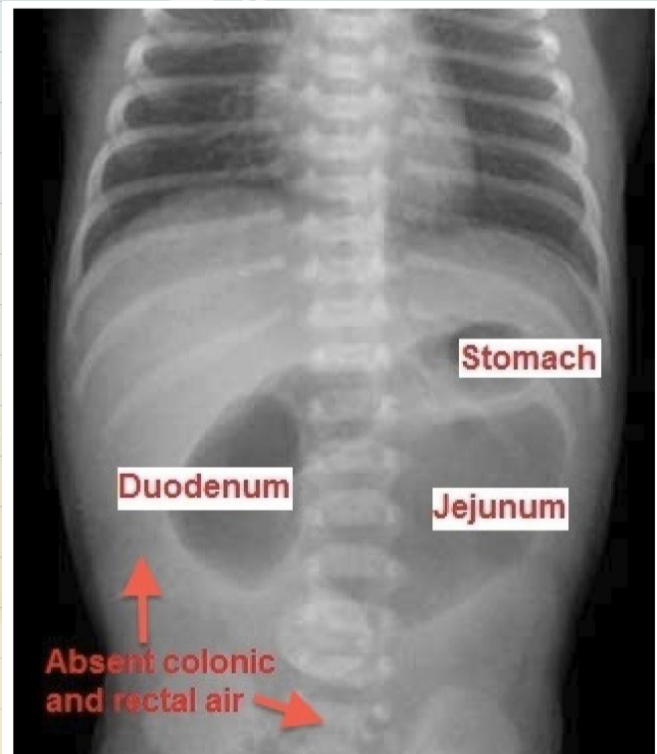
1. Nasogastric decompression, hydration, and correction of electrolyte abnormalities (typically hypochloremic metabolic alkalosis) are necessary.
2. Atresia is surgically corrected by duodenoduodenostomy.
3. Exploration for malrotation and other luminal obstructions is also performed at the time of surgical correction.

2. **Jejunioileal atresia**

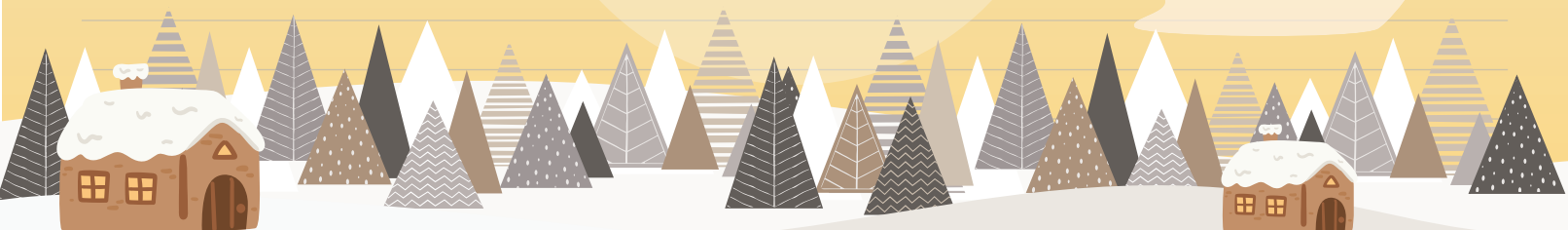
- a. **Definition.** This congenital obstruction of the jejunum or ileum is caused by a mesenteric vascular accident during fetal life.
- b. **Epidemiology.** The incidence is approximately 1 in 3000 live births. It is not usually associated with other anomalies.
- c. **Clinical features.** Bilious emesis and abdominal distension occur within the first few days of life.
- d. **Evaluation.** Abdominal **radiographs** reveal *air–fluid levels*, and **contrast studies** reveal the atresia.
- e. **Management.** Patients should undergo nasogastric suctioning and fluid resuscitation before surgical resection and anastomosis of the atretic segment of the small bowel.



Duodenal Atresia

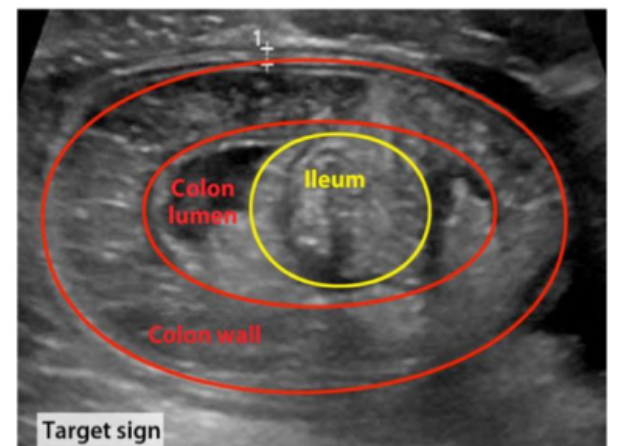
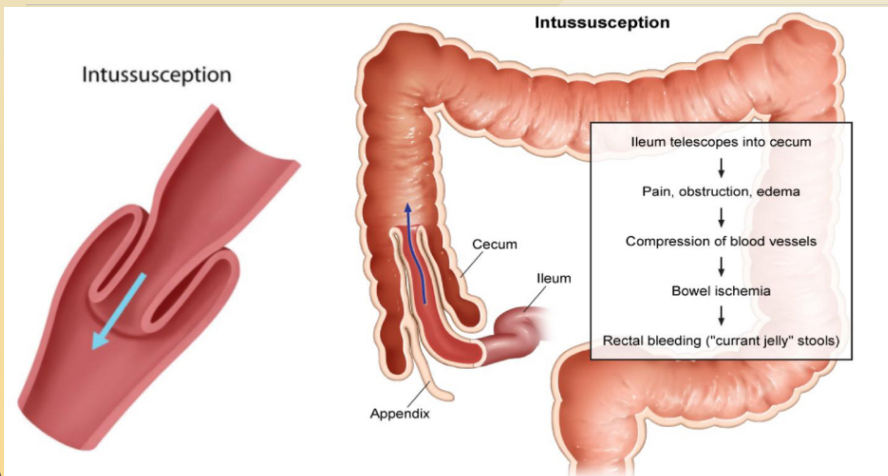


Jejunal atresia



D. Intussusception

1. **Definition.** Intussusception is the *telescoping or invagination of a more proximal portion of the intestine into a more distal portion*.
2. **Epidemiology.** The incidence is 1.5–4 in 1000 live births, with a slight **male predominance**. Peak incidence occurs at **5–9 months of age**. **Intussusception is the most common cause of bowel obstruction after the neonatal period in infants younger than 2 years.**
3. **Pathogenesis and etiology**
 - a. Intussusception may occur at a variety of sites within the intestine, but **ileocolic intussusception is the most common location**.
 - b. Etiology is generally unknown, but a **lead point** such as Meckel diverticulum, polyp, intestinal duplication, Peyer patch, or lymphoma may act to draw the proximal intestine inward. However, a lead point is identified in only 5% of cases and is **more common in older children**.
 - c. Intussusception causes bowel wall edema and hemorrhage, and may lead to bowel ischemia and infarction.
4. **Clinical features**
 - a. Previously healthy infants or children may present with **sudden onset of crampy or colicky abdominal pain**. The pain often occurs in intervals followed by periods of calm. Infants may cry and draw their legs toward the chest.
 - b. Vomiting and **lethargy** are common.
 - c. Stools may be normal or have a bloody, **“currant jelly”** appearance because of intestinal ischemia and mucosal sloughing.
 - d. Occasionally, a **sausage-shaped mass** may be palpated in the abdominal right upper quadrant, representing the intussusception.
5. **Evaluation**
 - a. Initial management should include fluid resuscitation.
 - b. Radiographs are of limited usefulness but may reveal dilated loops of the bowel, or pneumoperitoneum if perforation has occurred.
 - c. *Abdominal ultrasound* is the most accurate diagnostic tool in the hands of an experienced ultrasonographer. **Contrast enema** (barium or Gastrografin) can also be used to both establish the diagnosis and to reduce the intussusception. Contrast enema may show the classic **“coil spring”** sign representing the intussusception.
6. **Management**
 - a. **Air or contrast enemas**, often under ultrasound guidance, successfully reduce the intussusception in 80–90% of cases.
 - b. If the contrast enema fails to reduce the intussusception, or if the child has signs of peritonitis or pneumoperitoneum, **operative reduction** is indicated.
 - c. The **risk of recurrence** is 5% after contrast reduction and 1% after surgical repair.

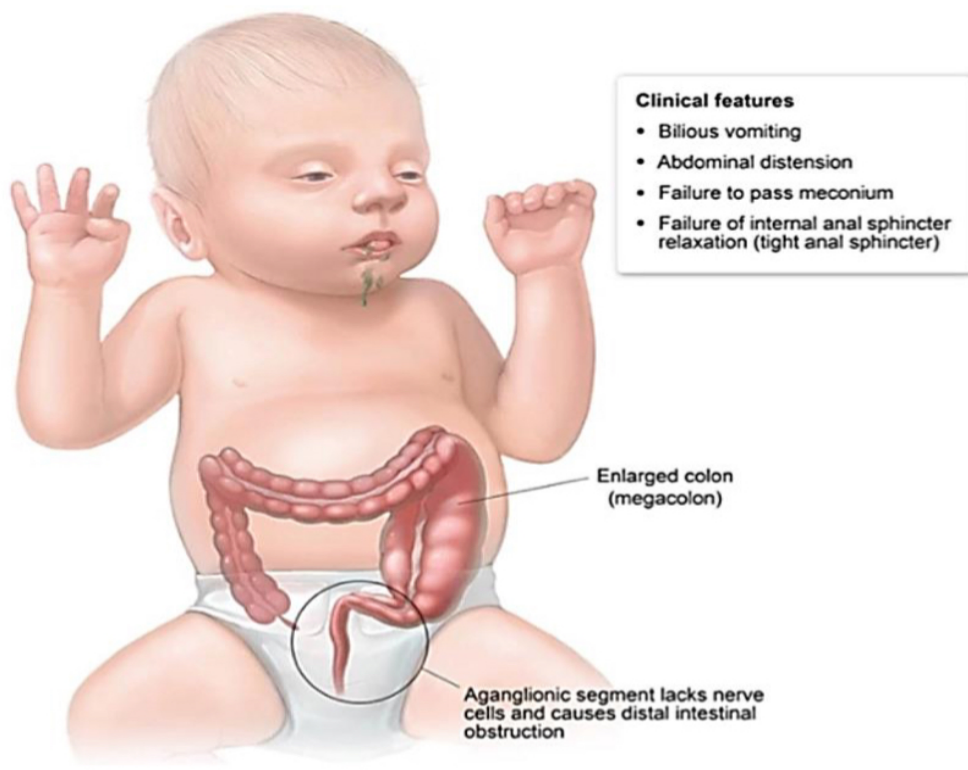


Intussusception

Risk factors	• Recent viral illness or rotavirus vaccination • Pathological lead point ◦ Congenital malformation of the intestines (eg, Meckel diverticulum) ◦ Henoch-Schönlein purpura ◦ Celiac disease ◦ Intestinal tumor ◦ Polyps
Clinical presentation	• Sudden, intermittent abdominal pain • "Currant jelly" stools • Sausage-shaped abdominal mass • Lethargy or altered mental status
Diagnosis	• "Target sign" on ultrasound
Treatment	• Air or saline enema • Surgery for removal of lead point

Hirschsprung Disease

- Hirschsprung disease (congenital aganglionic megacolon) is a **congenital lack of innervation of the distal bowel by the Auerbach plexus**.
- The disorder results from failed development of the enteric nervous system of a variable portion of the distal gut and **most commonly involves the rectosigmoid**. The affected colonic segment **cannot relax and therefore is chronically contracted**.
- Neural crest cells start migrating to the intestinal wall very early during embryonic development. They give rise to ganglion cells of the submucosal (Meissner) and myenteric (Auerbach) plexi of the bowel wall.
- **The arrest of migration of neural crest cells causes Hirschsprung disease, in which a segment of colon is deprived of ganglion cells. Since neural crest cells migrate caudally, the rectum is always involved in Hirschsprung disease.**
- There is **frequent association with Down syndrome, and it is more common in boys than in girls** (approximately 4:1).

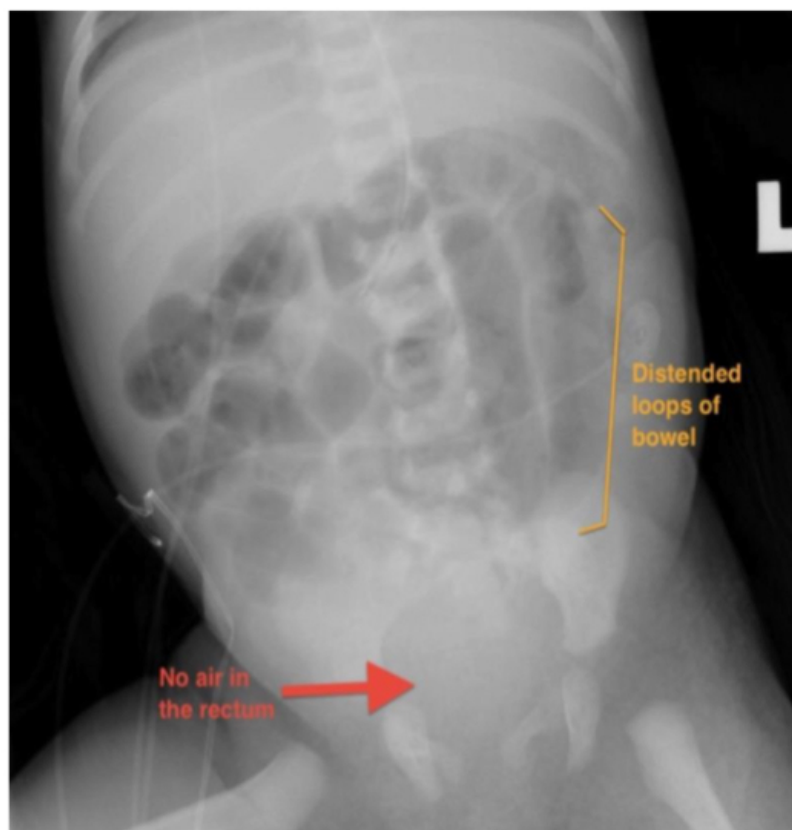


▪ Presentation:

- Of unaffected infants, 90% pass first meconium within 24 hours, **whereas children with Hirschsprung do not pass meconium for over 48 hours or fail to pass meconium at all.**
- Extreme constipation is followed by **large bowel obstruction** (poor feeding, abdominal distension, absent air in the rectum).
- Rectal examination can produce an explosive expulsion of gas and stool ("**squirt sign**") from temporary relief from the obstruction.

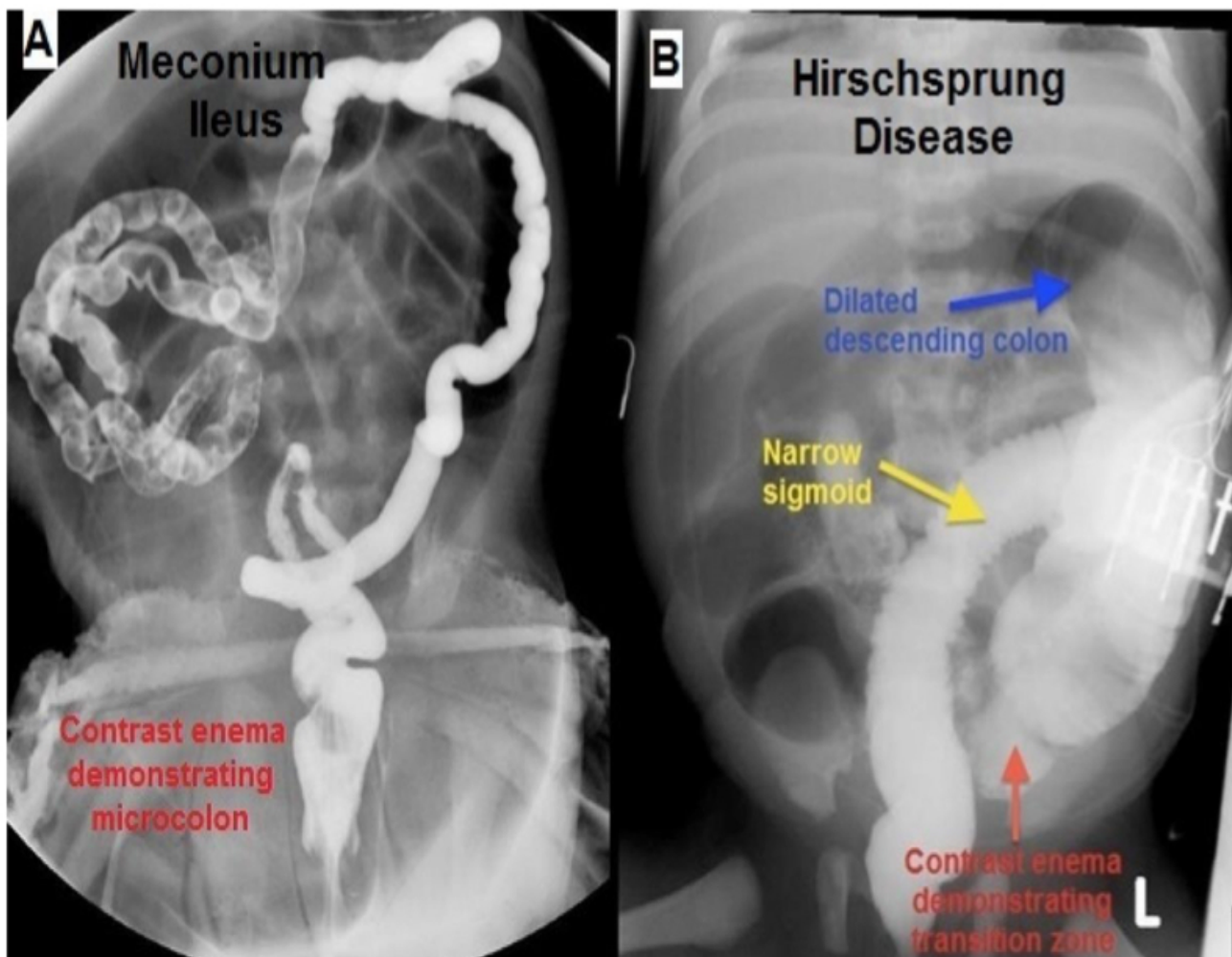
▪ Diagnostic Tests/Treatment:

- **Plain x-rays show distended bowel loops with a lack of air in the rectum.**
- **If there is no evidence of perforation (free air under the diaphragm), contrast enema can potentially delineate the level of obstruction.** A transition zone may be seen between the narrowed aganglionic segment and the normally innervated, dilated colon (megacolon).
- **Manometry will show high pressures in the anal sphincter.**
- **The mainstay of diagnosis is a full thickness biopsy** that reveals a lack of ganglionic cells in the submucosa.
- A 3-stage surgery procedure is curative.



❖ N.B:

- **Meconium ileus and Hirschsprung disease (congenital aganglionic megacolon) should be considered in any neonate with delayed passage of meconium** as 99% of healthy, full-term infants pass stool within 48 hours of birth.
- These 2 conditions have overlapping clinical features but **can usually be differentiated by the level of intestinal obstruction and meconium consistency.**
- Meconium ileus is virtually diagnostic for cystic fibrosis (CF). Although only 20% of patients with CF develop meconium ileus **almost all newborns with meconium ileus have CF.**
- A mutation in the CF transmembrane conductance regulator gene results in abnormal chloride and sodium transport and thick, viscous secretions in multiple organs. **Thick, inspissated meconium is difficult to propel, resulting in obstruction at the level of the ileum and a narrow, underdeveloped colon (microcolon) compared to more typical rectosigmoid obstruction seen in infants with Hirschsprung disease.**
- Administration of hyperosmolar enema (Gastrografin) **can potentially break up the inspissated meconium and dissolve the obstruction.** Surgery is required if therapeutic enema is unsuccessful.
- In contrast to Hirschsprung disease, rectal examination would not relieve any meconium due to its inspissated consistency. Meconium ileus is not associated with Down syndrome.



Differentiating features of Hirschsprung disease and meconium ileus		
	Hirschsprung disease	Meconium ileus
Associated disorder	Down syndrome	Cystic fibrosis
Typical level of obstruction	Rectosigmoid	Ileum
Meconium consistency	Normal	Inspissated

OTHER CONGENITAL DISORDERS

GASTROSCHISIS:

- abdominal wall defect not involving the umbilicus, through which intestinal contents have herniated.
- the bowel is not covered by peritoneum or amniotic membrane.
- As a result, prolonged contact with the amniotic fluid typically causes a thick, exudative covering (a "peel") on the exposed bowel.
- **Gastroschisis is not associated with extraintestinal anomalies**, but segments of intestinal atresia are common.
- After surgical reduction of the defect, return of normal bowel function may be slow and requires prolonged parenteral nutrition for infants with long atretic segments (short bowel syndrome) and infants with a thick peel.



OMPHALOCELE

- abdominal wall defect at the umbilicus caused by failure of the intestine to return to the abdomen during fetal life.
- The bowel remains within the umbilical cord and is covered by peritoneum and amniotic membranes.
- This defect is associated with other congenital anomalies, especially:
 1. cardiac defects
 2. **Beckwith-Wiedemann syndrome** (somatic overgrowth, hyperinsulinemic hypoglycemia, risk for Wilms tumor)
 3. intestinal complications.
- Treatment is surgical closure, which sometimes must be performed in stages to fit the bowel into a congenitally small abdominal cavity.



DUPLICATION CYSTS

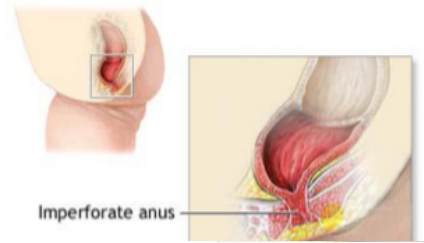
- Abnormal fluid-filled structures lined with intestinal or gastric mucosa.
- They are found within the mesentery and lie adjacent to normal bowel.
- These cysts do not communicate with the adjacent bowel lumen.
- some cysts may cause symptoms in early infancy or grow slowly for years, eventually causing problems secondary to:
 1. perforation
 2. bowel obstruction
 3. intussusception
 4. volvulus.
- Diagnosed by ultrasound, CT scan, or contrast studies.
- Treatment is surgical excision.



ANORECTAL MALFORMATIONS

- Including imperforate anus and its variants, are embryologic defects recognized at birth by the absence of a normal anal opening.
- Evaluation of these infants should include observation for emergence of meconium from the urethra or fistulas on the perineum.
- A urinary catheter should be placed if urinary distention is present.
- In low lesions, a fistulous opening that drains meconium is present on the perineum.
- Low lesions commonly are associated with fistulization between the bowel and bladder, vagina, or urethra.

- Lateral x-rays show the defect level and show gas in the bladder caused by a fistula.
- These children are treated initially by colostomy to divert the fecal flow, with subsequent anogenital reconstruction.
- The internal sphincter muscle is functionally absent in high lesions, and continence after repair is difficult to achieve.
- **All** children with imperforate anus require MRI of the lumbosacral spinal cord because of high incidence of **tethered spinal cord**.
- Urologic dysfunction is common and should be evaluated appropriately.



What happens when you have a tethered spinal cord?

As a result, the spinal cord can't move freely within the spinal canal. This can cause the spinal cord to stretch out as the spine grows, leading to possible nerve damage, pain and other symptoms. In most cases, the condition becomes worse over time.

