SOS: Review of Common Neonatal GI Conditions

Jacqui Hoffman, DNP, ARNP, NNP-BC
NNP Track Coordinator; Clinical Assistant Professor
University of Florida, Gainesville, FL
Neonatal Nurse Practitioner
Pediatrix Medical Group, Tampa, FL

The speaker has signed a disclosure form and indicated she has no significant financial interest or relationship with the companies or the manufacturer(s) of any commercial product and/or service that will be discussed as part of this presentation.

Session Summary

This lecture provides a general overview of common neonatal GI problems to help the attendant prepare for certification exams.

Session Objectives

Upon completion of this presentation, the participant will be able to:

- identify the three parts of the primordial gut and the common structures/organs arising from each;
- contrast the difference between the infant with a gastroschisis and the infant with an omphalocele;
- discuss clinical presentation, diagnostic evaluation, and management of the infant with a suspected abdominal obstruction;
- describe the clinical presentation, diagnostic evaluation and management of the infant with necrotizing enterocolitis.

Test Questions

1. A previously well full-term infant presents with bilious vomiting. What is the first disease process that the infant should be evaluated for?
   - a. Pyloric stenosis
   - b. Sepsis
   - c. Malrotation with midgut volvulus

2. Omphalocele and Gastroschisis can best be differentiated by:
   - a. Assessing involvement of the umbilicus
   - b. Identifying the organs exposed by the defect
   - c. Noting the presence of a membranous covering

3. Which of the following gastrointestinal conditions is associated with a high incidence of associated malformation?
   - a. Gastroschisis
   - b. Omphalocele
   - c. Jejunoileal atresia
4. A term male neonate at 50 hours of age has abdominal distention and episodes of vomiting. No meconium has been passed since birth except during your physical when a rectal examination is done. An abdominal X-ray is non-specific; a contrast study depicts areas of dilatation and constriction in the sigmoid colon. The most likely diagnosis is:
   a. Meconium ileus
   b. Malrotation with volvulus
   c. Hirschsprung’s disease

5. Almost all infants pass meconium by:
   a. 12 hours of life
   b. 24 hours of life
   c. 48 hours of life

6. A 4 week old male infant presented with projectile vomiting of nonbilious emesis. The physical exam reveals a small “olive-shaped” mass in the abdomen. The most likely diagnosis is:
   a. Meconium plug
   b. Pyloric stenosis
   c. Necrotizing enterocolitis

References


**Session Outline**

See presentation handout on the following pages.
SOS: REVIEW OF COMMON GI CONDITIONS

Jacqui Hoffman, DNP, ARNP, NNP-BC
NNP Track Coordinator
University of Florida
NNP Pediatric Medical Group, Tampa

Embryology Review

- Primordial gut
  - Forms during the 4th week
  - Divided into 3 separate parts:
    - Foregut
    - Midgut
    - Hindgut

Foregut
- Oral cavity, pharynx, tongue, tonsils, salivary glands
- Upper and lower respiratory system
- Esophagus
  - Reaches final length by 7 wk gestation
- Stomach
- Duodenum
  - Develops from caudal part of foregut, cranial part of midgut & splanchnic mesenchyme
- Liver and biliary apparatus, gallbladder, pancreas, spleen
- Blood supply – celiac artery

Midgut
- Small intestine
- Ascending colon and large portion of transverse colon
- Cecum
- Appendix
- Blood supply – superior mesenteric artery
- Physiologic umbilical herniation during the 6th week with return of intestines to abdomen by week 10

Hindgut
- Distal third of the transverse colon
- Descending colon
- Sigmoid colon
- Rectum and upper part of anal canal
- Epithelium of the urinary bladder
- Urethra
- Blood supply – inferior mesenteric artery
Baby A Case Study

- Baby A is a former 1720 gm 28.5 EGA female. She was delivered by stat C-Section when mother presented in active preterm labor with rapidly advancing dilation and fetus noted to be in breech position. Maternal urine culture was positive for Ecoli that was Ampicillin resistant. Infant was intubated in the delivery room, given a dose of surfactant and extubated to nasal CPAP. Infant developed worsening respiratory distress and required intubation for the next 6 days. Infant was started on Caffeine after extubation for apnea of prematurity.

Baby A Case Study (continued)

- Other findings included a HR of 178 at rest, capillary refill time of 4-5 seconds, abdomen distended with visible loops of bowel, and absent bowel sounds.
- What diagnoses would you consider in this infant?
- What diagnostic work-up would be most appropriate at this time?
- What management options should be started immediately?

Other Areas to Review

- Function of various organs
- Concurrent development of nervous system
- Motility and factors that may affect this
- Developmental considerations
- GI hormones
- Absorption of various nutrients

Baby A Case Study (continued)

- Baby A received 48 hr antibiotic with final blood culture remaining negative. Trophic feeds of preterm formula were started on DOL 2 (MOB did not want to breast feed). The feeds were advanced to full feeds over the next 7 days. When feeds were at 100 ml/kg/day, the caloric content was increased to 24 cal/oz. On DOL 11 the infant was noted to have temperature instability, increased A&B events requiring vigorous stimulation and a 22ml bilious gastric residual.
Necrotizing Enterocolitis (NEC)

**Incidence**
- 1-3/1,000 live births
- 5.6-7.2% VLBW infants (< 1,500 gm) [Vermont Oxford Database]
- 8% ELBW infants (< 1,000 gm)
- Varies between Medical Centers
- Occurs both sporadically & in clusters
- Age at presentation is inversely related to gestational age at birth

**Pathogenesis**
- Precise pathogenesis remains unknown
- Multifactorial

<table>
<thead>
<tr>
<th>Preterm</th>
<th>Term</th>
</tr>
</thead>
<tbody>
<tr>
<td>Intestinal Immaturity</td>
<td>Hypoxia-ischemia</td>
</tr>
<tr>
<td>Abnormal Microbial Colonization</td>
<td></td>
</tr>
</tbody>
</table>

**Clinical Presentation**
- Gastrointestinal symptoms
  - Abdominal distention
  - Feeding intolerance
  - Emesis (may or may not be bilious)
  - Bloody stools
  - Abdominal wall erythema or bluish discoloration
- Systemic instability mimicking sepsis
  - Apnea & bradycardia, poor perfusion, lethargy

**Differential Diagnosis (DDx)**
- Mucosal inflammation
  - NEC, allergic colitis, gastritis/stress ulcer
- Infection
  - Systemic infection, infectious gastroenteritis, pseudomembranous colitis
- Congenital abnormalities and malformations
  - Intestinal stenosis/atroresia, imperforate anus, meconium ileus/plug, Hirschsprung's, Malrotation/volvulus
- Vascular accidents
  - Intestinal thromboembolic infarct
- Other
  - Intussusception, gastritis/gastric ulcer/perforation, swallowed maternal blood, pneumothorax → pneumatoperitoneum
Diagnostic Evaluation

- History and physical findings
- Laboratory
  - Neutropenia, left-shift of neutrophils
  - Thrombocytopenia
  - Coagulation disturbances
  - Hemolytic anemia
  - Metabolic acidosis
  - Glucose instability
  - Hyponatremia

Diagnostic Evaluation

- Radiographic findings
  - Ileus
  - Pneumatosis intestinalis (intramural air)
  - Dilated loops
  - Thickened bowel wall
  - Pneumoperitoneum
  - Portal venous gas
Bell Staging Criteria for NEC

- **Stage I – Suspected NEC**
  - Temperature instability, apnea & bradycardia, ↑ gastric residuals, mild abdominal distention, occult blood in stool, normal or mild ileus on X-ray

- **Stage II – Definite NEC**
  - Same as Stage I plus prominent abd distention +/- tenderness, absent bowel sounds, grossly bloody stools, ileus or dilated bowel loops with focal pneumatosis on X-ray

- **Stage IIA – Mild NEC**
  - Mild acidosis & thrombocytopenia, abd wall edema & tenderness +/- palpable mass, extensive pneumatosis +/- portal venous gas and early ascites on X-ray

- **Stage IIB – Moderate NEC**
  - Vital sign & laboratory evidence of deterioration, shock, evidence of perforation, and pneumoperitoneum on X-ray

- **Stage IIIA – Advanced NEC**
  - Resp & metabolic acidosis, mechanical ventilation, hypotension, oliguria, DIC, worsening wall edema & erythema with induration, prominent ascites with persistent bowel loop but no free air on X-ray

- **Stage IIIB – Advanced NEC**
  - Vital sign & laboratory evidence of deterioration, shock, evidence of perforation, and pneumoperitoneum on X-ray

Medical Management

- Bowel rest (NPO), hyperalimentation
- Gastrointestinal decompression
- Blood culture & broad-spectrum antibiotics
- Serial abdominal girth measurements
- Serial abdominal X-rays, CBC, coagulation studies, electrolytes & blood gases based on clinical condition
- Parenteral nutrition
- Supportive therapies based on clinical presentation
Surgical Management

- Absolute indications
  - Pneumoperitoneum
  - Clinical deterioration despite maximal medical treatment
  - Abdominal mass with persistent intestinal obstruction or sepsis
  - Development of intestinal stricture

Surgical Management

- Peritoneal drainage
- Exploratory laparotomy with resection of diseased bowel, enterostomy & stoma formation

Management Postoperatively

- NPO, IVF (central line)
- Replogle to low, intermittent suction
- Pain management
- Broad-spectrum antibiotics; ?Clindamycin
- Ostomy care

Prevention of NEC

- Prevent preterm delivery
- Measures shown to be effective in prevention
  - Human milk
  - Feeding protocols using careful advancement
- Questionable measures
  - Withholding feeds during transfusions or while on Indomethacin
  - Acidification of formula
  - IgG, IgA
  - Oral antibiotics
  - Pre- or pro-biotics

Morbidity and Mortality

- Complications
  - Intestinal strictures
    - Bloody stools, FTT, feeding abnormalities, and diarrhea
  - Prolonged hospitalization
  - Feeding intolerance
  - Short bowel syndrome
  - Parenteral nutrition-induced cholestasis
  - Neurodevelopmental delay
- 20-30% overall mortality

Spontaneous Ileal Perforation (SIP)

- Cause unknown
- Occurs more frequently in VLBW & ELBW
- Risk Factors
  - Postnatal steroid, Indocin, and vasopressors use
  - Some studies show association with chorioamnionitis
- Most commonly perforation occurs in terminal ileum
Pathogenesis

- Medications or other exposures lead to mucosal hyperplasia, submucosal thinning and smooth muscle necrosis
- These occurrences lead to bowel wall fragility and depletion of endothelial nitric oxide

Spontaneous Ileal Perforation (SIP)

- Clinical presentation
  - Sudden onset, typically in the first two weeks of life
  - May have few symptoms
  - Lack of infectious symptoms
  - Pneumoperitoneum on x-ray

- Treatment
  - NPO
  - Replogle to low, intermittent suction
  - Surgery consult
  - Antibiotics
  - Supportive care

- Morbidity and Mortality
  - Decreased mortality and neurodevelopmental impairment compared to infants with NEC

Abdominal Wall Defects

Umbilical Hernia

- Protrusion of tissue or viscera through the umbilical fascial ring

- Incidence
  - Unknown
  - Estimated to be 18% in white infants and as high as 42% in black infants
  - Increased incidence in preterm infants and low birth weight infants
  - Can be associated with certain syndromes and disease processes (Trisomy 21, congenital hypothyroidism, Beckwith-Wiedemann syndrome)

Spontaneous Ileal Perforation (SIP)

Baby B Case Study

- A 17 y/o G1P0 white female presented to the ER with complaint of abdominal pain. She was diagnosed with active labor and transferred immediately to L&D where she precipitously delivered in the bed. A 3200gm male infant, estimated to be 38 weeks was given Apgar scores of 9/9. Physical exam demonstrated an abdominal wall defect with exposed intestinal contents.

- What is your DDx? Is there other information you would like?
Umbilical Hernia

- Clinical Presentation
  - Protrusion of the umbilicus especially when crying or straining
  - Fascial defect is usually < 2 cm in diameter
  - Redundant umbilical skin

- DDx
  - Small omphalocele

Umbilical Hernia

- Diagnostic work-up
  - Diagnosed by physical exam

- Management
  - Majority spontaneously close if defect is small by 3 years of age
  - Surgery recommended if hernia persists after 4-5 years of age
    - Intraumbilical or infraumbilical incision
    - Hernia sac excised and fascial defect is sutured

Gastroschisis

- Abdominal wall defect with herniation of abdominal contents lateral to the umbilical cord
- Etiology unknown
  - Vascular accident during embryogenesis
- Incidence: 1/4,000 to 1/20,000 births
- Association with teen pregnancies and low socioeconomic status
- Malrotation is almost universal

Gastroschisis

- Clinical Presentation
  - Herniated bowel that may be edematous or even matted protruding through an abdominal wall defect located lateral to an intact umbilical cord
  - Occasionally liver herniated
  - No peritoneal sac
  - Usually isolated defect without other non-GI anomalies

- Differential diagnosis (DDx)
  - Ruptured omphalocele, cloacal extrophy

Gastroschisis

- Diagnosis
  - Prenatal ultrasound
  - Elevated maternal serum α-fetoprotein
  - Physical exam at birth
Gastroschisis

**Management Preoperatively**
- Delivery
- Facility where surgical services available
- Avoid bag/mask ventilation
- Use latex-free products
- Bowel bag or sterile, saline soaked dressing
- Right side-lying position
- NPO - replogle (sump tube) to low, intermittent suction

**Management**
- Aggressive fluid management
  - Increased total fluids
  - I&O, BP and perfusion monitoring
- Radiant warmer/isolette
- Thorough physical exam
- Broad-spectrum antibiotics
- Baseline laboratory studies

**Surgical Management**
- Primary closure - preferred
  - Dependent on size of defect and bowel edema
  - May experience:
    - Decreased cardiac output,
    - Respiratory compromise, and/or
    - Compromise of perfusion to kidneys, intestines and lower extremities
- Staged closure using prosthetic silo
  - Gradual manual reduction 1-2 times per day over 5-10 days

**Postoperatively**
- Monitor for increased intraabdominal pressure
- Sedation/pain management
- Mechanical ventilation
- Antibiotic therapy
- Fluid and feeding challenges
  - Central line for prolonged TPN

**Morbidity and Mortality**
- Sepsis
- Prolonged ileus
- Necrotizing enterocolitis (NEC)
- Complications from prolonged TPN
- 10% will have intestinal atresia
- Mortality < 5%
**Omphalocele**

**Clinical Presentation**
- Herniated single loop of bowel to bowel + other abdominal organs through the base of intact umbilical cord
- Abdominal cavity may be scaphoid in appearance
- Protective transparent sac, occasionally this may rupture
- Look for dysmorphic features & other anomalies

**DDx**
- Gastroschisis, umbilical hernia, patent urachus

**Diagnosis**
- Prenatal ultrasound
- Elevated maternal serum α-fetoprotein
- Amniocentesis to r/o chromosomal abnormalities
- Physical exam at birth

**Management preoperatively**
- Intravenous fluids
- Radiant warmer/isolette
- Thorough physical exam
  - Screening echocardiogram
  - Obtain chromosomes
  - Consider radiographic evaluation or other evaluation as deemed necessary
- Broad-spectrum antibiotics

**Etiology**
- Incomplete return of bowel into abdomen or incomplete closure of anterior abdominal wall

**Incidence:**
- 1-3/10,000 live births
  - 3:1 male-to-female predominance; more common in older maternal age
  - 50-70% will have associated anomalies; Beckwith-Wiedemann should be considered

**Abdominal wall defect with herniation of abdominal contents into the umbilical cord**

**Clinical Presentation**
- Herniated single loop of bowel to bowel + other abdominal organs through the base of intact umbilical cord
- Abdominal cavity may be scaphoid in appearance
- Protective transparent sac, occasionally this may rupture
- Look for dysmorphic features & other anomalies

**DDx**
- Gastroschisis, umbilical hernia, patent urachus

**Management preoperatively**
- Intravenous fluids
- Radiant warmer/isolette
- Thorough physical exam
  - Screening echocardiogram
  - Obtain chromosomes
  - Consider radiographic evaluation or other evaluation as deemed necessary
- Broad-spectrum antibiotics

**Abdominal wall defect with herniation of abdominal contents into the umbilical cord**

**Clinical Presentation**
- Herniated single loop of bowel to bowel + other abdominal organs through the base of intact umbilical cord
- Abdominal cavity may be scaphoid in appearance
- Protective transparent sac, occasionally this may rupture
- Look for dysmorphic features & other anomalies

**DDx**
- Gastroschisis, umbilical hernia, patent urachus

**Management preoperatively**
- Intravenous fluids
- Radiant warmer/isolette
- Thorough physical exam
  - Screening echocardiogram
  - Obtain chromosomes
  - Consider radiographic evaluation or other evaluation as deemed necessary
- Broad-spectrum antibiotics

**Abdominal wall defect with herniation of abdominal contents into the umbilical cord**

**Clinical Presentation**
- Herniated single loop of bowel to bowel + other abdominal organs through the base of intact umbilical cord
- Abdominal cavity may be scaphoid in appearance
- Protective transparent sac, occasionally this may rupture
- Look for dysmorphic features & other anomalies

**DDx**
- Gastroschisis, umbilical hernia, patent urachus

**Management preoperatively**
- Intravenous fluids
- Radiant warmer/isolette
- Thorough physical exam
  - Screening echocardiogram
  - Obtain chromosomes
  - Consider radiographic evaluation or other evaluation as deemed necessary
- Broad-spectrum antibiotics

**Abdominal wall defect with herniation of abdominal contents into the umbilical cord**

**Clinical Presentation**
- Herniated single loop of bowel to bowel + other abdominal organs through the base of intact umbilical cord
- Abdominal cavity may be scaphoid in appearance
- Protective transparent sac, occasionally this may rupture
- Look for dysmorphic features & other anomalies

**DDx**
- Gastroschisis, umbilical hernia, patent urachus

**Management preoperatively**
- Intravenous fluids
- Radiant warmer/isolette
- Thorough physical exam
  - Screening echocardiogram
  - Obtain chromosomes
  - Consider radiographic evaluation or other evaluation as deemed necessary
- Broad-spectrum antibiotics
Omphalocele

**Surgical Management**
- Primary closure
  - Dependent on size of defect and abdominal cavity size
  - Monitor for cardiac, respiratory, renal and even liver compromise
- Staged closure using prosthetic silo
  - Gradual manual reduction 1-2 times per day over 5-10 days

**Management postoperatively**
- Monitor for increased intraabdominal pressure
- Monitor LFTs
- Sedation/pain management
- Mechanical ventilation
- Antibiotic therapy
- Fluid and feeding challenges
  - Central line for maximum nutrition

**Morbidity and Mortality**
- Mortality rate variable depending on associated anomalies and size of defect
- Gastroesophageal reflux is common
- Bowel obstruction
- Ventral hernia

**GI Obstructions**

**Mechanical**
- Congenital intrinsic
  - Atresias, stenoses, meconium ileus, anorectal malformations, enteric duplications
- Congenital extrinsic
  - Volvulus, peritoneal bands, annular pancreas, cysts/tumors, incarcerated hernias
- Acquired
  - NEC, intussusception, peritoneal adhesions

**Functional**
- Intrinsic
  - Hirschsprung disease, meconium plug syndrome, ileus, peritonitis
- Extrinsic
  - Intestinal pseudo-obstruction syndrome
GI Obstruction Pearls

- **Polyhydramnios**
  - More common in proximal obstructions
- **Abdominal distention**
  - More common in distal obstructions (and TEF)
- **Emesis**
  - Bilious - more common when obstruction is distal to the ampulla of Vater
  - Early onset indicates high obstruction; late - low
- **Normal meconium patterns**
  - 94% pass meconium by 24 hr of age; 99.8% by 48 hr

GI Obstruction Generalizations

- **Management**
  - Replogle to low, intermittent suction
  - NPO, IVF
  - Abdominal x-ray and/or contrast study
  - Consult pediatric surgeon

Hypertrophic Pyloric Stenosis

- Hypertrophy of pylorus, resulting in stricture of the outlet from the stomach to the small intestine
- **Etiology**
  - Exact cause unknown; hereditary component
  - **Incidence:** 1-4/1,000 live births
    - First born more often affected
    - 4:1 Male-to-female predominance
    - More common among **Caucasian** infants
    - Associated conditions uncommon

Clinical Presentation

- Nonbilious vomiting, usually around 2-6 wk of age, that becomes projectile with time
- Signs and symptoms of dehydration, poor weight gain
- **DDx**
  - GER, sepsis, small bowel obstruction

Diagnostic Evaluation

- History and Physical exam
- Abdominal ultrasound
- Upper GI contrast study
Preoperative Management
- Baseline laboratory studies
- Correct electrolyte and acid-base imbalances
- Fluid resuscitation may be necessary
- Replogle to low, intermittent suction

Surgical Management
- Pyloromyotomy
  - Laparotomy
  - Laparoscopy

Postoperative Management
- Pain management
- Fluid and feeding challenges
  - Monitor serum electrolytes, I&O, weight
  - Feedings started 6-8hr post-op
  - Never place gastric tube post-op
- Genetics consult?

Prognosis
- Generally, complete recovery with no residual effects
- Surgery corrects stenosis and stenosis generally does not reoccur
- Persistent vomiting first few days post-op, resolves quickly

Baby C Case Study
Baby C is a 3675 gram male infant delivered by spontaneous vaginal delivery at 39.1 wk EGA. The mother had breast fed the infant twice with no reported issues. At the next feeding, the mother was back in the operating room postpartum hemorrhage, therefore, the infant was fed a small amount of term formula. The infant was noted to have approximately 20ml bilious emesis. An OGT is placed and while awaiting X-ray, additional bilious aspirate is noted.

Baby C Case Study (continued)
- What diagnoses would you consider in this infant?
- What diagnostic work-up would be most appropriate at this time?
- What management options should be started immediately?
Baby D Case Study

Baby D is a 3150 gram term female infant delivered by scheduled repeat C-Section. Mother's past medical and this pregnancy history were unremarkable. The infant has been breast feeding ad lib demand for the past 3 days with good voiding and stooling pattern. Just prior to discharge, the infant was noted to have a small bilious emesis. An abdominal X-ray was non-specific and a barium enema was read as the cecum somewhat high-riding otherwise normal; further correlation with infant's clinical condition recommended.

Baby D Case Study (continued)

What diagnoses would you consider in this infant?
What diagnostic work-up would be most appropriate at this time?
What management options should be started immediately?

Baby E Case Study

Baby E is a 3620 gram 36.5 wk EGA late-preterm male infant delivered by spontaneous vaginal delivery. Maternal GBS status was not on the chart; mother reported she thought it was negative. SROM x 19 hours with maternal temperature of 100.9 noted just prior to delivery. A screening CBC (nl) and blood culture were drawn on the infant per hospital policy. The infant was breast fed on demand every 1-3 hours with good latch scores.

Baby E Case Study (continued)

At 28 hours of age, the infant developed increasingly poorer latch scores. Over the next several hours, the infant was noted to have increasingly abdominal distention with the girth up 3 cm. After the last feeding at approximately 37 hours of age, the infant had a large bilious emesis. In reviewing the chart, vital signs have been stable, infant has been voiding, no stool has been documented since birth.
Baby E Case Study (continued)

What diagnoses would you consider in this infant?
What diagnostic work-up would be most appropriate at this time?
What management options should be started immediately?

Small Bowel Obstructions

Duodenal Atresia

- Congenital obstruction of the duodenum, usually distal to the ampulla of Vater
- Etiology
  - Unknown; thought to be from failure of recanalization of the duodenum during the 8th week of gestation
- Incidence
  - 1:2,500 live births
  - Females more commonly affected than males
  - High incidence of associated conditions; 30% infants associated with Trisomy 21

Clinical Presentation

- Vomiting, may be clear or bilious
- Abdominal distention
- If incomplete atresia or stenosis, may not present in the immediate NB period

- DDx
  - Midgut volvulus, malrotation, meconium ileus, meconium plug, Hirschsprung’s disease

Bilious emesis in the neonate is midgut volvulus until proven otherwise!
**Diagnostic Evaluation**

- Prenatal ultrasound
- History of polyhydramnios
- Abdominal X-ray
  - Classic double-bubble

**Duodenal Atresia**

- Classic double-bubble

**Preoperative Management**

- NPO, IVF
- Replogle to low, intermittent suction
- Thorough physical exam to detect associated anomalies
  - Screening echocardiogram

**Surgical Management**

- Duodenoduodenostomy - removal of atretic portion with reanastomosis of the ends of the bowel
- Most infants will have a gastrostomy tube placed

**Postoperative Management**

- Pain management
- Low, intermittent suction to G-Tube
- Fluid and feeding challenges
- Prophylactic antibiotic therapy
**Prognosis**

- Possible leaking at anastomosis site
- Prognosis is dependent on associated anomalies

**Jejunal and Ileal Atresia**

- Failure of the lumen of the bowel to form properly
- Etiology
  - Mesenteric vascular insult with subsequent necrosis and reabsorption of the affect segment(s)
- Incidence
  - 1/1,000 live births
  - Usually presents as an isolated defect

**Jejunal and Ileal Atresia**

Clinical Presentation

- Bilious emesis usually within the 1st 24 hours of life
- Progressive abdominal distention
- May initially pass meconium, then none
- DDx
  - Midgut volvulus, malrotation, meconium ileus, meconium plug, Hirschsprung’s disease

**Diagnostic Evaluation**

- Presence of symptoms
- ? History of polyhydramnios
- Prenatal ultrasound
- Abdominal X-ray
  - Gas or fluid-filled dilated loops of bowel with scant amounts of gas distal to the obstruction
  - “Triple-bubble” - proximal jejunal atresia
- Contrast enema

**Triple Bubble**

Preoperative Management
- Replogle to low, intermittent suction
- NPO, IVF
- Correct any electrolyte imbalances

Surgical Management
- Surgical procedure dependent on amount of intestine involved
  - End-to-end or end-to-oblique-side anastomosis
  - Externalization of the proximal and distal ends

Postoperative Management
- Pain management
- Replogle to low, intermittent suction
- Fluid and feeding challenges
- Prophylactic antibiotic therapy

Prognosis
- Ileus
- Peritonitis, if perforation occurred
- Short bowel syndrome (SBS)
- Strictures or adhesions
- Leak at anastomosis site
- Decreased survival in neonates with multiple atresias

Meconium Ileus
- Mechanical obstruction of the distal lumen due to meconium
- Etiology
  - Unknown; due to hyposecretion of pancreatic enzymes or abnl viscid secretions from the mucous glands of the sm. intestine
- Incidence
  - Majority of cases are associated with cystic fibrosis

Clinical Presentation
- Abdominal distention with thickened bowel loops often visible
- Bilious emesis
- Failure to pass meconium
- Bowel perforation with peritonitis (will have tenderness and/or erythema)
- DDx
  - Meconium plug, small bowel atresia, Hirschsprung’s Disease
Diagnostic Evaluation

- Prenatal ultrasound may show peritoneal calcifications
- History and Physical exam
- Abdominal X-ray
  - Dilate proximally, microcolon distally
  - “Soap-bubble” appearance
- Contrast Enema
  - Will demonstrate microcolon
- Gene probe studies

Management

- NPO, IVF
- Replogle to low-intermittent suction
- Pediatric surgery consult
- Broad-spectrum antibiotics
- Hyperosmotic enema (if no contraindication)
- If enemas are not effective in evacuating the meconium, surgery is indicated

Surgical Management

- If prenatal/postnatal perforation is present or if enemas were not effective in evacuating the meconium, surgery is indicated
- Laparotomy with end-to-end anastomosis or creation of stoma

Postoperative Management

- Pain management
- Replogle to low, intermittent suction
- Fluid and feeding challenges
  - Need pancreatic enzymes
- Antibiotic therapy
- Genetic counseling
  - DNA
  - Sweat chloride
- Pulmonary toilet
  - CPT, aerosols, humidity
<table>
<thead>
<tr>
<th><strong>Prognosis</strong></th>
<th><strong>Meconium Plug Syndrome</strong></th>
</tr>
</thead>
</table>
| - Post-operatively  
  - Volvulus, perforation with peritonitis, infection  
  - Long-term  
  - If CF diagnosed, will need careful f/u  
  - Morbidity and mortality due to complications of CF | - Mechanical obstruction of the distal colon/rectum due to thick, inspissated meconium  
- Etiology  
  - Unknown  
- Incidence  
  - 1:100 newborns  
  - Associated with pre-eclampsia with Mag therapy, IDM, prematurity, hypothyroidism, hypotonia and sepsis |

<table>
<thead>
<tr>
<th><strong>Clinical Presentation</strong></th>
<th><strong>Diagnostic Evaluation</strong></th>
</tr>
</thead>
</table>
| - Failure to pass meconium  
  - Abdominal distention  
  - Visible loops of bowel  
  - Bilious emesis (late finding)  
  - DDx  
  - Meconium ileus, Hirschsprung’s Disease | - Diagnosis  
  - History and Physical exam  
  - Abdominal X-ray  
  - Dilated loops of bowel and few air fluid levels  
  - Water-soluble contrast enema  
  - Diagnostic and therapeutic |

<table>
<thead>
<tr>
<th><strong>Management</strong></th>
<th></th>
</tr>
</thead>
</table>
| - NPO, IVF  
  - Replogle to low-intermittent suction  
  - ? digital rectal exam  
  - Contrast enema  
  - Further work-up? |
Malrotation/Volvulus

- Abnormal rotation and fixation of intestines during 7th to 12th week of gestation
- Etiology
  - Unknown; occurs when the intestines do not rotate and/or the mesentery does not fixate during embryologic development
- Incidence
  - 1:6,000 live births
  - Associated with abdominal wall defects, intestinal atresia, imperforate anus, cardiac anomalies, and Trisomy 21

Clinical Presentation

- Most cases present during 1st month
- Bilious vomiting
- May be abdominal distention, tenderness
- May have bloody emesis or stools
- Signs of shock and sepsis if necrosis
- DDx
  - Small bowel atresia, NEC

Diagnostic Evaluation

- Clinical presentation
- Abdominal X-ray
- Abdominal ultrasound
- Contrast Upper GI Study

Preoperative Management

- Considered a surgical emergency if symptomatic
  - Delay in diagnosis can result in loss of the entire midgut
- Replogle to low, intermittent suction
- NPO, IVF
- Broad-spectrum antibiotics

http://embryology.med.unsw.edu.au/Notes/gi02.htm
Surgical Management
- Laparotomy (Ladd’s procedure)
- Appendectomy commonly done

Post-operative Management
- Replogle to low, intermittent suction
- NPO, IVF until return of bowel function
- Pain management
- Feedings are introduced slowly with elemental formula commonly required

Prognosis
- Prognosis is excellent if uncomplicated
- Mortality/morbidity influenced by amount of intestine involved, presence of shock/sepsis, prematurity, and other associated conditions
- Short bowel syndrome (SBS) if large portion of bowel was removed

Hirschsprung’s Disease
(Congenital megacolon or aganglionic megacolon)
- Absence of ganglion cells in the colon
- Etiology
  - Caused by failure of migration of neural crest neuroblasts to the distal portion of the colon
  - Incidence: 1:5,000 births
  - Occurs predominantly in white males
  - 1/3 will have positive family history
  - Associated anomalies not common
  - Increased risk of hearing loss, ocular neuropathies, and decreased peripheral nerve function

Clinical Presentation
- Failure to pass meconium
- History of constipation
- Bilious vomiting and abdominal distention
- Enterocolitis

DDx
- Malrotation/volvulus, meconium ileus or plug, small bowel obstruction
Diagnostic Evaluation

- Clinical presentation
- Abdominal X-ray
  - Non-specific
- Barium enema
  - Transition zone
- Anal manometry
- Rectal biopsy
  - Definitive diagnosis

Management

- Medical management attempted initially
- Management preoperatively
  - Replogle to low, intermittent suction
  - NPO, IVF
  - Broad-spectrum antibiotics
  - Colonic irrigation

Surgical Management

- Staged repair
  - Colostomy with later pull-through procedure
- Complete pull-through repair
  - Laparoscopic surgery

Postoperative Management

- Replogle to low, intermittent suction
- NPO, IVF until enteral nutrition started
- Pain management
- Routine ostomy care
- ? rectal irrigation
- Rectal dilations
- Consider genetic counseling

Prognosis

- Dysmotility of the colon
  - Stooling abnormalities – constipation or incontinence
- Rectal stenosis
- Stricture formation
**Anorectal Malformations**

- Wide spectrum of abnormalities characterized by a stenotic or atretic anal canal
- Malformations include:
  - Persistent cloaca
  - Anal stenosis
  - Membranous anal atresia
  - Anal agenesis
  - Anorectal agenesis (imperforate anus)
  - Rectal atresia

**Etiology**

- Failure during embryonic development of differentiation of the urogenital sinus and cloaca

**Incidence**

- 1:4,000 to 1:5,000 births
- More common in males
- Associated with GU, vertebral, CV and esophageal atresia with TEF (think VATER/VACTERL)

**Clinical Presentation**

- Presenting signs and symptoms will vary depending on type of defect
- It takes 24 hours for a fistula to declare itself
- Physical exam may reflect absence or narrowing of anal opening, or a deep anal dimple
- Failure to pass meconium, meconium in the urine (males) or meconium in the vaginal outlet

---


Classifications

- **Low**
  - The rectum has descended below the sphincter muscle complex
  - Rarely a fistula

- **High**
  - Located above the sphincter muscle
  - Usually has a fistula

Diagnostic Evaluation

- Physical exam
  - Visual and digital exam
- X-ray of the sacrum
  - Wangensteen-Rice technique – inverted lateral radiograph
- Perineal and spinal ultrasonography
- MRI, ECHO
- Voiding cystourethrogram

Preoperative Management

- Replogle to low, intermittent suction
- NPO, IVF
- Broad spectrum antibiotics
- Serial urinalysis
- Observe closely for enterocolitis or possible perforation

Surgical Management

- Varies depending on type of defect
- ? colostomy needed
- ? urinary diversion needed
- Management differs between males & females
Surgical Management - Males

- Low lesion (perineal fistula)
  - Serial dilations or perineal anoplasty
- High lesions
  - Colostomy
  - Definitive repair at 3-12 mon of age (posterior sagittal anorectoplasty)

Surgical Management - Females

- Single perineal orifice
  - Colostomy, urinary diversion, drain hydrocolpos
- Perineal fistula
  - Serial dilations or perineal anoplasty
- Vestibular fistula
  - Colostomy or primary repair

Postoperative Management

- Varied depending on surgical procedure required
- IV antibiotics for 48-72 hours
- Antibiotic ointment 8-10 days
- NPO, IVF
- Replogle to low, intermittent suction
- Pain management
- Colostomy care
- Foley catheter for 8-14 days if rectourethral fistula
- Anal dilations after corrective surgery

Prognosis

- Urinary incontinence
- Fecal incontinence
- Constipation
- Postoperative colostomy complications

Other Miscellaneous Conditions to Review

- Inguinal hernia
- Testicular torsion
- Annular pancreas
- Intussusception
- GI bleeding
- Gastroesophageal reflux
- Short bowel syndrome
- Biliary atresia