SOS: Review of Common Neonatal GI Conditions

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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

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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
Embryology Review

- **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

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 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 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.

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?
Necrotizing Enterocolitis

**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
- Most commonly involves terminal ileum and proximal colon

**Pathophysiology**
- Precise pathogenesis remains unknown
- Multifactorial
  - Prematurity
  - Formula feeding
  - Intestinal ischemia
  - Abnormal bacterial colonization

**NEC**

- Intestinal Ischemia
- Abnormal bacterial Colonization
- Formula Feeding
- Prematurity (mucosal immaturity)
Clinical Findings

- 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/atresia, imperforate anus, meconium ileus/ptag, Hirschsprung's, Malrotation/volvulus
- Vascular accidents
  - Intestinal thromboembolic infarct
- Other
  - Intussusception, gastritis/gastric ulcer/performation, swallowed maternal blood, pneumothorax → pneumoperitoneum

Diagnostic Evaluation

- History and physical findings
- Laboratory
  - Leukopenia
  - Neutropenia
  - Thrombocytopenia
  - Coagulation disturbances (DIC)
  - Lyte abnormalities (hypo-natremia and/or kalemia)
  - Metabolic acidosis
  - Glucose instability
Diagnostic Evaluation

- Radiographic findings
  - Ileus
  - Pneumatosis intestinalis
  - 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

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

- Stage IIIA – Advanced NEC
  - Resp & metabolic acidosis, require 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 - pneumoperitoneum on X-ray

Medical Management

- Bowel rest (NPO), hyperalimentation
- GI decompression – replogle (sump tube)
- 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

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

Outcomes

- Complications
  - Intestinal strictures
    - Bloody stools, FTT, feeding abnormalities, and diarrhea
  - Prolonged hospitalization
  - Feeding intolerance
  - Short bowel syndrome
  - Parenteral nutrition-induced cholestasis
  - Neurodevelopmental delay
- 30-50% Overall Mortality
Spontaneous Ileal Perforation (SIP)

- Cause unknown
- Occurs more frequently in VLBW and ELBW
- Risk Factors
  - More likely to have received postnatal steroids, have a PDA treated with indocin, and to have received vasopressor support use
  - Some studies show association with chorioamnionitis
- Most commonly perforation occurs in terminal ileum

Pathophysiology

- 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 week of life
  - May have few symptoms
  - Lack of infectious symptoms
  - Pneumoperitoneum on x-ray

Spontaneous Ileal Perforation (SIP)

- Treatment
  - NPO
  - Replogle to low, intermittent suction
  - Surgery consult
  - Antibiotics
  - Supportive care
- Prognosis
  - Decreased mortality and neurodevelopmental impairment compared to infants with NEC
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?

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)

- 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
  - Infraumbilical or intraumbilical 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

- Diagnosis
  - Prenatal ultrasound
  - Elevated maternal serum α-fetoprotein
  - Increased incidence of oligohydramnios unless intestinal atresia (then polyhydramnios), fetal growth restriction and meconium-stained fluid
  - Physical exam at birth

- Clinical Presentation
  - Herniated bowel that may be edematous or even matted protruding through an abdominal wall defect located lateral to an intact umbilical cord (usually to the right)
  - Occasionally liver herniated
  - No peritoneal sac
  - Usually isolated defect without other non-GI anomalies
- Differential diagnosis (DDx)
  - Ruptured omphalocele, cloacal extrophy
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 to low, intermittent suction

- 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
**Gastroschisis**

- **Surgical Management**
  - Staged closure using prosthetic silo
  - Gradual manual reduction 1-2 times per day over 5-10 days

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

**Gastroschisis**

- **Complications**
  - Sepsis
  - Prolonged ileus
  - Complications from prolonged TPN
  - Strictures
  - Scar revision
  - 10% will have intestinal atresia
  - Mortality < 10%

**Omphalocele**

- **Abdominal wall defect with herniation of abdominal contents into the umbilical cord**
- **Etiology**
  - Incomplete return of bowel into abdomen or incomplete closure of anterior abdominal wall
- **Incidence:** 1/3,000 to 1/10,000 live births
  - 3:1 male-to-female predominance; more common in older maternal age
  - All will have malrotation but other GI anomalies rare
  - 50-70% will have associated anomalies; Beckwith-Wiedemann syndrome should be considered
Omphalocele

- **Diagnosis**
  - Prenatal ultrasound
  - Elevated maternal serum α-fetoprotein
  - Amnioentesis to rule out chromosomal abnormalities
  - Fetal echocardiogram recommended
  - Physical exam at birth

- **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
  - Giant omphalocele (>5cm)
  - Look for dysmorphic features and other anomalies

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

- **Management preoperatively**
  - Delivery
    - Facility with surgical services
    - C-Section recommended if large defect containing liver
    - Avoid bag/mask ventilation
  - Use latex-free products
  - Bowel bag or sterile, saline soaked dressing
  - NPO – replogle to low, intermittent suction

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**Classical omphalocele**

**Ruptured omphalocele**
Omphalocele

- 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

- 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

- Complications
  - Mortality rate variable depending on associated anomalies and size of defect
  - Gastroesophageal reflux is common
  - Decreased GI motility not as common compared to gastroschisis
  - 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
  - Biliary - 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
- Incidence: 1-4/1,000 live births
  - First born more often affected
  - 5:1 Male-to-female predominance
  - Appears to be a familial risk
  - More common among Caucasian infants
  - Associated conditions uncommon
### Hypertrophic Pyloric Stenosis

**Clinical Presentation**
- Nonbilious vomiting, usually around 3-6 wk of age, that becomes projectile with time
- Signs and symptoms of dehydration, electrolyte abnormalities, and poor weight gain

**DDx**
- GER, sepsis, small bowel obstruction

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**Diagnosis**
- History and Physical exam
- Abdominal ultrasound
  - Preferred diagnostic test
  - “String sign” or “Apple core”
- Upper GI contrast study

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**Management preoperatively**
- Baseline laboratory studies
- Correct electrolyte and acid-base imbalances
- Fluid resuscitation may be necessary
- Replogle to low, intermittent suction

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**Surgical Management**
- Pyloromyotomy
- Laparotomy
- Laparoscopy
Hypertrophic Pyloric Stenosis

- Management Postoperatively
  - 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?

- Complications
  - 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 (Cont.)

- 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 (Cont.)

- Feedings were resumed and the infant’s discharge was cancelled. The infant remained in the nursery overnight for further observation. The infant continued to breast feed well overnight. The following afternoon just before the pedi came in to do evening discharge rounds, the abdomen was noted to be significantly distended. An OGT was placed and approximately 20ml bilious aspirate was noted. The pedi requested a neonatology consult.

Baby D Case Study (Cont.)

- 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 (Cont.)

- 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 (Cont.)

- 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 – 10th week of gestation
  - Usually occurs in 2nd part of duodenum
- Incidence
  - 1/6,000 to 1/10,000 births
  - High incidence of associated conditions
    - Trisomy 21 (~30%)
    - CHD (30%)

Clinical Presentation

- Vomiting, usually bilious, in first 24hr of life
- Abdominal distention, generally confined to the upper abdomen
- Can have meconium passage
- 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!
Duodenal Atresia

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

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

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

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

Duodenal Atresia

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

Jejunal-Ileal Atresia

- Failure of the lumen of the bowel to form properly

- Etiology
  - Mesenteric vascular insult with subsequent necrosis and reabsorption of the affected segment(s)

- Incidence
  - 1/1,000 live births
  - Usually presents as an isolated defect
  - Found more frequently in the distal ileum, followed by the proximal jejunum -> distal jejunum -> proximal ileum
Jejunal-Ileal Atresia

**Clinical Presentation**
- Bilious emesis
- Progressive abdominal distention (greater with more distal atresias)
- May initially pass meconium, then none

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

**Diagnosis**
- 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
  - The lower the obstruction, the greater the amount of air
  - “Triple-bubble” — proximal jejunal atresia
- Contrast enema
Jejunal-Ileal Atresia

- Management preoperatively
  - 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

Jejunal-Ileal Atresia

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

- Complications
  - 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 terminal ileum due to meconium
- Etiology
  - Unknown; due to hyposecretion of pancreatic enzymes or abnormal hyperviscous secretions from the mucous glands of the small intestine
- Incidence
  - Majority of cases (90%) are associated with cystic fibrosis

Clinical presentation

- Usually presents within 24-48 hr
- Abdominal distention with thickened, palpable bowel loops
- Bilious emesis
- Failure to pass meconium
- Bowel perforation with peritonitis may occur (will have tenderness and/or erythema)

DDx

- Meconium plug, small bowel atresia, Hirschsprung’s Disease

Diagnosis

- 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
Meconium Ileus

**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
Meconium Ileus

- Management postoperatively
  - 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

- Complications
  - Post-operatively
    - Volvulus, perforation with peritonitis, infection
  - Long-term
    - If CF diagnosed, will need careful f/u
    - M&M due to complications of CF

Meconium Plug Syndrome

- Mechanical obstruction of the distal colon/rectum due to thick, inspissated meconium
- Etiology
  - Unknown
- Incidence
  - 1:100 newborns
  - Associated with pre-eclampsia with magnesium therapy, IDM, prematurity, hypothyroidism, hypotonia and sepsis

- Clinical presentation
  - Failure to pass meconium in 1st 24-48 hours
  - Abdominal distention
  - Visible loops of bowel
  - Biliary emesis (late finding)

- DDx
  - Meconium ileus, Hirschsprung Disease, ileal atresia, septic ileus
Meconium Plug Syndrome

- Diagnosis
  - History and Physical exam
  - Abdominal X-ray
    - Dilated loops of bowel and few air fluid levels
  - Water-soluble contrast enema
    - Diagnostic and therapeutic

http://www.medcyclopaedia.com/library/topics/volume_vii/m/meconium_plug_syndrome.aspx

Meconium Plug Syndrome

- Management
  - NPO, IVF
  - Replogle to low-intermittent suction
  - Contrast enema
  - Further work-up?

- Outcome
  - Excellent outcome after relief of obstruction

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
  - Can be isolated finding or associated with other malformations such as abdominal wall defects, intestinal atresia, imperforate anus, cardiac anomalies, congenital diaphragmatic hernia, and Beckwith-Wiedemann syndrome
Malrotation/Volvulus

- 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
Malrotation/Volvulus

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

MALRODUCTION/VOLVULUS

• Surgical Management
  o Laparotomy (Ladd’s procedure)
  o Appendectomy commonly done

Malrotation/Volvulus

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

Malrotation/Volvulus

• Complications
  o Prognosis is excellent if uncomplicated
  o Mortality/morbidity influenced by amount of intestine involved, presence of shock/sepsis, prematurity, and other associated conditions
  o 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 GI anomalies not common; increased risk of hearing loss, ocular neuropathies, and decreased peripheral nerve function

Hirschsprung’s Disease

- Clinical presentation
  - Failure to pass meconium
  - Bilious vomiting and abdominal distention
  - H/O constipation or abnormal stooling
  - Enterocolitis

- DDx
  - Malrotation/volvulus, meconium ileus or plug, distal ileal atresia, colonic atresia

Hirschsprung’s Disease

- Diagnosis
  - Clinical presentation
    - 50% diagnosed by 1st month of life
  - Abdominal X-ray
    - Diffuse dilated loops of bowel along with decreased or absent air in the rectum
  - Barium enema
    - Transition zone may not be seen in newborn
    - Retained barium >24 hr after study in full-term infants
  - Anal manometry
  - Rectal biopsy
    - Definitive diagnosis
Hirschsprung’s Disease

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

Hirschsprung’s Disease

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

Hirschsprung’s Disease

- Management postoperatively
  - Replogle to low, intermittent suction
  - NPO, IVF until enteral nutrition started
  - Pain management
  - Routine ostomy care
  - Rectal irrigation
  - Rectal dilations
  - Consider genetic counseling
Hirschsprung’s Disease

- Complications
  - Dysmotility of the colon
    - Stooling abnormalities (15%) – 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

Anorectal Malformations

- 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)
Anorectal Malformations

- 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


Types

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

- High
  - Located above the sphincter muscle
  - Usually has a fistula
  - 10% will have tethered cord

Anorectal Malformations

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

Anorectal Malformations

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

- 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

- Management postoperatively
  - Varies 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
<table>
<thead>
<tr>
<th>Anorectal Malformations</th>
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<tr>
<td><strong>Complications</strong></td>
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<tr>
<td>o Urinary incontinence</td>
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<tr>
<td>o Fecal incontinence</td>
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<td>o Postoperative colostomy complications</td>
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