SOS: Review of Common Neonatal GI Conditions

Jacqui Hoffman, DNP, ARNP, NNP-BC
NNP Track Coordinator, College of Nursing
University of Florida, Gainesville, FL
Neonatal Nurse Practitioner
Pediatricx 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 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 gastrochisis and the infant with an omphalocele;
- discuss clinical presentation, diagnostic evaluation, and management of the infant with a suspected abdominal obstruction.

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 Gastrochisis 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. Gastrochisis
   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


**SOS: REVIEW OF COMMON GI CONDITIONS**

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

---

**Embryology Review**

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

---

**Embryology Review**

- **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

---

**Embryology Review**

- **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 E.coli 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

---

Other finding 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 finding 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?

---

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

---

Other finding 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 finding 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 finding 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 finding 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 finding 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 finding 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 (NEC)

**Incidence**
- 1-3/1,000 live births
- Affects mainly premature infants, especially <1000 grams and/or < 28 weeks gestation
  - 2.2 – 8.3% of VLBW infants (<1500 gm) (Vermont Oxford, 2010), 8% ELBW infants (< 1000 gm)
- Varies between Medical Centers
- Occurs both sporadically & in clusters
- Age at presentation is inversely related to gestational age at birth (Gordon, Clark, Swanson & Spitzer, 2014)
- Incidence peaks at a postmenstrual age of about 29-31 weeks (Sharma & Hudak, 2013)

**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>Immature immune system</td>
</tr>
<tr>
<td>Feedings</td>
<td>Immature intestinal epithelial barrier</td>
</tr>
</tbody>
</table>

**Pathophysiology of NEC**

**Preterm/Immaturity** (Neu & Walker, 2011; Neu, 2014)
- Immature digestion, absorption, motility
- Immature immune system
- Immature intestinal epithelial barrier

**Microbiome**
- Dysbiosis (Torrazza et al, 2013)

**Inflammation**

**Role of Toll-like Receptor 4 (TLR4)**

Adapted from Sharma & Hudak, 2013

Choi, 2014
**Clinical Presentation**

- Varies from non-specific to a fulminant onset
- **Gastrointestinal symptoms**
  - Abdominal distention
  - Feeding intolerance
  - Emesis (may or may not be bilious)
  - Bloody stools
  - Abdominal wall erythema or bluish discoloration

**Clinical Presentation**

- Systemic instability mimicking sepsis
  - Increased apnea & bradycardia
  - Worsening of respiratory function
  - Lethargy
  - Poor perfusion, pallor, hypotension,
  - Temperature and/or glucose instability

**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/plug, Hirschsprung, Malrotation/volvulus
- Vascular accidents
  - Intestinal thromboembolic infarct
- Other
  - Intussusception, gastritis/gastric ulcer/perforation, swallowed
    maternal blood, pneumothorax → pneumoperitoneum

**Diagnostic Evaluation**

- History and physical findings
- Laboratory (poor sensitivity/specificity)
  - 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
Other Diagnostic Modalities

Abdominal Ultrasound
- Can identify even small volumes of gas
- Preferred for visualizing abdominal fluid and ascites
- Doppler study can be used to assess arterial perfusion to the bowel wall
- Portal venous gas can be detected easier vs X-ray

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
- Suspected NEC + 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
- Same as Stage I plus prominent abdominal distension +/- 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

Bell Staging Criteria for NEC (cont.)

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

- Surgery consult
- 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
- Supportive therapies based on clinical presentation (don’t forget pain management)

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
- Alternative to laparotomy – definitive treatment or temporizing measure
- 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
- Evidence of efficacy and safety
  - Human milk
  - Feeding protocols using careful advancement
- Measures that may be beneficial
  - Trophic feeds safer alternative to NPO
    - Each day NPO, increases risk by 8% (Kitsman et al, 2015)
  - Withholding feeds during transfusions or while on Indomethacin
  - Acidification of formula
    - Restrict use of H2-blockers
    - Pre/probiotics

### Morbidity and Mortality
- 20-30% overall mortality
  - Medical 21%, surgical (35-50%) (Hull et al, 2014)
- Complications (increase LOS/HC costs)
  - Intestinal strictures (affects up to 40%)
    - Bloody stools, FIT, feeding abnormalities, diarrhea
  - Feeding intolerance
  - Short bowel syndrome
  - Parenteral nutrition-induced cholestasis
  - Neurodevelopmental delay

### 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

Spontaneous Ileal Perforation (SIP)

- 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

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

- 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

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

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

**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 Preoperatively**
- 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

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

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

**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: 1-3/10,000 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

**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

**Omphalocele**

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

**Omphalocele**

- 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
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

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
GI Obstructions

- 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
- “String sign”
- “Double track sign”

**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 D Case Study (continued)**

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**

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 (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
  - 1:2,5000 live births
  - Females more commonly affected than males
  - High incidence of associated conditions; 30% infants associated with Trisomy 21

Moore & Persaud (2013). Figure 11-6

Moore & Persaud (2013). Figure 11-6
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

---

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

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

Jejunal and Ileal Atresia
- Clinical Presentation
- Surgical Management
- Prognosis
### 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

### 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)
- Fluid resuscitation
- Monitor for perforation
- If enemas are not effective in evacuating the meconium, surgery is indicated

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

Prognosis
- 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

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 Mag therapy, IDM, prematurity, hypothyroidism, hypotonia and sepsis

Clinical Presentation
- Failure to pass meconium
- Abdominal distention
- Visible loops of bowel
- Bilious emesis (late finding)
- DDx
  - Meconium ileus, Hirschsprung’s Disease

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

Management

- NPO, IVF
- Replogle to low-intermittent suction
- Digital rectal exam
- Contrast enema
- Further work-up?

Diagnostic Evaluation

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

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

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

Management

- NPO, IVF
- Replogle to low-intermittent suction
- Digital rectal exam
- Contrast enema
- Further work-up?
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

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

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)

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

- Small left colon syndrome
- Inguinal hernia
- Annular pancreas
- GI bleeding
- Malabsorption/maldigestion
- Gastroesophageal reflux
- Short bowel syndrome
- Biliary atresia
- Cleft lip/palate

### Questions?