

Gastrointestinal Disorders

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Disclosures

The following speaker of this CME activity has no relevant financial relationships with commercial interests to disclose.

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Objectives

- Discuss normal embryologic development of the gastrointestinal system
- Describe common gastrointestinal disorders, clinical presentation, and management strategies
- Describe radiographic findings of common gastrointestinal disorders



Embryology

Folding of embryonic disc week 3
Trachea and esophagus have common origin
Incomplete partitioning of tracheoesophageal septum- stenosis or atresia
Digestive system develops from 3 structures

Foregut

Oral cavity, pharynx, esophagus, upper & lower respiratory system, stomach to second part of duodenum, liver, pancreas, spleen, and blood supply of celiac artery

Midgut

3rd part of duodenum to first 2/3 of large intestine and blood supply of superior mesenteric artery

Hindgut

Remaining large intestine through rectum, bladder, urethra, and blood supply of inferior mesenteric artery

GI Tract

Main Functions

Absorption and digestion of nutrients
Elimination of waste products
Maintenance of fluid and electrolyte balance
Protection of host from toxins and pathogens

Prenatal Assessment

Family History - genetic syndrome
Maternal History/Physical
Complications - polyhydramnios, oligohydramnios
Conditions
Medications
Fetal ultrasound - stomach present, bowels intact?



Abdominal Assessment

Normal

Round, soft, symmetrical, pink, active bowel sounds, liver 1-2 cm below right costal margin, spleen rarely palpable (left costal margin), kidneys 4-5cm

Muscular Development- diastasis recti

Umbilicus/Cord - pearly white, 3 vessels, detaches 10-14 days

Abnormal

Erythema, dusky, mottled, distended, organomegaly, ascites, scaphoid, asymmetrical, mass, visible bowel loops or peristalsis, hyper/hypoactive bowel sounds

Prune belly syndrome, omphalocele, gastroschisis, bladder exstrophy, hernias (umbilical, inguinal, femoral)

Omphalitis, patent urachus, granuloma

Omphalocele



Abdominal wall defect - failure intestines return from cord into abdominal cavity
Herniation abdominal contents into umbilical cord
May also contain liver and other organs

Incidence

2.5/10,000 live births
3:1 male:female predominance

Etiology

Multifactorial, environmental, chromosomal

Associated conditions

Prematurity(SGA), cardiac defects (50%), syndromes and anomalies (50-65%), neurologic/neural tube anomalies(40%), other midline defects, atresia and malrotation, GU anomalies

*** What other follow-up is important with these babies?

Gastroschisis



Abdominal wall defect
Herniation abdominal contents lateral to umbilical cord
No protective sac
Liver occasionally herniated

Incidence

4:10,000 births, mothers <20 years old, low socioeconomic status, exposure vasoconstrictors (cocaine, NSAIDS, decongestants)

Etiology

Unclear, theories include vascular accident or rupture of umbilical stalk

Associated Conditions

Prematurity, IUGR, malrotation, atresia and/or strictures, intestinal dysfunction

Omphalocele/Gastroschisis

Management

Deliver where pediatric surgical services available

Avoid bag/mask ventilation

Bowel decompression

Bowel bag/sterile saline soaked dressing

Right side-lying positioning

Aggressive fluid management

Antibiotics, long term IV access awaiting GI motility

Surgery

Primary vs. stage closure

Pain management, respiratory support, follow cardiac output, bowel perfusion, gradual restart enteral feedings

Complications

Sepsis, ileus, NEC, prolonged TPN, intestinal atresia (10%), mortality (<5%)

Genetics

Gastrointestinal Obstructions

Mechanical

Atresia, stenosis, meconium ileus, anorectal malformations, volvulus, annular pancreas, cysts tumors, incarcerated hernias, NEC, intussusception, adhesions

Functional

Hirschsprung's disease, meconium plug syndrome, ileus, peritonitis, pseudo-obstruction syndrome

Clinical Presentation

Polyhydramnios - proximal

Abdominal distention - distal and TEF

Bilious emesis - distal to ampulla of Vater

Treatment – repleg, LIS, NPO, IVFs, KUB and/or contrast study, consult pediatric surgery

*** **What is a normal stool pattern?**



Esophageal Atresia/Tracheoesophageal Fistula

Esophageal Atresia - interruption esophagus

Tracheoesophageal Fistula- trachea fails to differentiate from esophagus (4 weeks)

Etiology - genetic factors

Incidence - 1/1000-2500 births

Associated anomalies - 25-40% CHD, 25-30% Vertebral anomalies, 15% VACTERL association, LBW, CHARGE

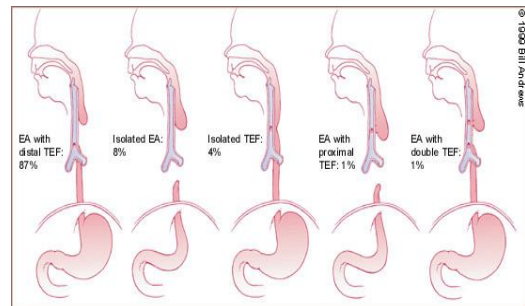
Diagnosis – U/S - small/absent stomach, polyhydramnios

Clinical Presentation

- copious secretions
- choking, coughing, respiratory distress
- emesis, distention
- cyanosis - onset of feeding
- inability to pass gastric tube
- recurrent pneumonia

Surgery – primary, staged (large gap esophageal segments), dilatations

Types of EA and TEF



Hypertrophic Pyloric Stenosis

Hypertrophy of pylorus, resulting in stricture of outlet from stomach to small intestine

Cause

Unknown, hereditary component (5%), PGEs

Incidence

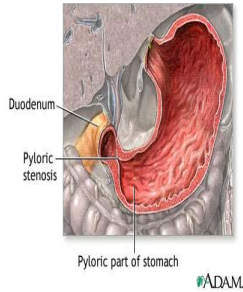
1-5/1,000 live births, more common in Caucasian, term, trisomy 18 & 21, boys>girls, smoking mother

Clinical Presentation

Non-bilious vomiting usually 2-6 weeks of age, becomes projectile, dehydration, poor weight gain

Diagnosis

History and PE - olive mass RUQ, Visible peristalsis sometimes
Abdominal ultrasound, UGI contrast study



Management Pyloric Stenosis

Correct dehydration - electrolyte, acid-base imbalances

Fluid resuscitation

Decompression

Surgical management

Pyloromyotomy (incision in muscle to release stricture)

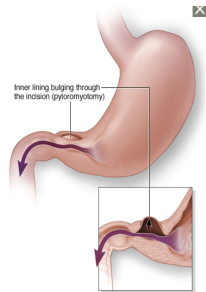
Pain management

Fluid and feeding challenges

Feeds started 6-8 hours post-op

Never place a OG/NG tube post-op

**May have vomiting first few days post-op, resolves quickly



Duodenal atresia/web/stenosis

Intestinal atresias - most common cause obstruction in newborn
Obstruction of duodenum, usually distal to **Ampulla of Vater**

Associated conditions: genetic (30% occurrence), cardiac (30%), malrotation (20%)

Which genetic disorder commonly associated?

Incidence: 1/7000 live births

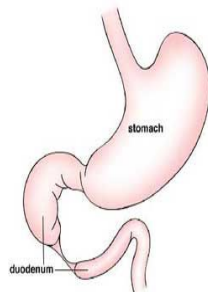
Classic **"double bubble"**

Abdominal distention, upper abdomen fullness, jaundice, bilious vomiting

Surgery - Duodenostomy - removal atretic portion with reanastomosis of bowel

May have gastrostomy tube placed

Restart feeds - 3-10 days, slow progression



Jejunal and Ileal Atresia

Etiology - Failure bowel lumen form properly - mesenteric vascular insult, necrosis, reabsorption affected segments

Associated conditions - malrotation, meconium peritonitis/ileus

Usually isolated defect

Incidence 1/1000 live births

5 Types I, II, IIIA (most common), IIIB (apple peel), IV (sausage links)

Clinical Presentation Bilious emesis- first 24-48 hours

Progressive abdominal distention

Initially pass meconium

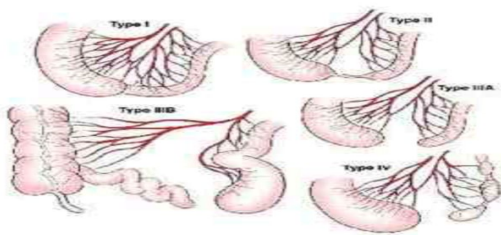
Jaundice

KUB "triple bubble", contrast study to confirm, microcolon

Treatment - surgery (resection/reanastomosis) or ileostomy/jejunostomy, bowel rest 7-14 days, TPN, gradual advancement feeds (elemental, breastmilk)

Complications - delayed gastric emptying, slow motility, strictures

Types of Jejunoileal Atresias



Four subtypes of jejunoileal atresia

Jejunal and Ileal



Malrotation with Volvulus

*** Billious emesis is midgut volvulus until proven otherwise!

Malrotation

Abnormal rotation/fixation intestines during 6th-10th week gestation, predisposes to obstruction

Volvulus

Blood flow compromised, bowel ischemic, dilates

Incidence: 1/5000 live births

Diagnosis PE, KUB, UGI (Whirlpool sign)

Abdominal distention

Hypotension and shock

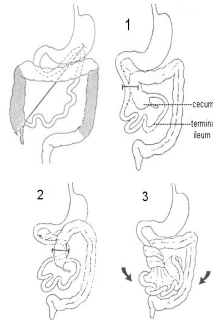
Surgical emergency

IVF, NPO, replete, antibiotics

Surgery - Laparotomy, Ladd's Procedure

Await return of bowel function

Complications - short bowel syndrome, strictures



Meconium Ileus

Obstruction of intestine - overly thick meconium

Deficiency digestive enzymes - from pancreas

Incidence: 1/2000 live births of Caucasian infants

3 Cardinal Signs intestinal obstruction

- generalized abdominal distention
- bilious vomiting
- failure to pass meconium within 24 hours

90% infants with meconium ileus have CF only 15% with CF have meconium ileus
KUB "soap-bubble" appearance, dilated proximal bowel, distal microcolon, scattered calcifications

BE: microcolon

Non-surgical - water soluble enemas (diagnostic and therapeutic)

Surgery - resection of compromised bowel, reanastomosis or stoma placement

CF Testing, follow Genetics, Pulmonary, GI

Meconium Plug Syndrome

Mechanical obstruction of distal colon/rectum due to thick meconium

Associated: maternal magnesium sulfate therapy, maternal diabetes, prematurity, hypothyroidism, hypotonia (CNS abnormality), sepsis

Incidence: 1/100 newborns

Presentation: failure pass meconium, abdominal distention, bowel loops (hyperactive sounds), bilious emesis (late finding)

Diagnosis: PE

KUB (diffuse dilated loops of bowel)

Water soluble enema (diagnostic and therapeutic)

75% expel plug spontaneously, avoiding surgery

Consider CF testing, consider work up Hirschsprung's Disease



Hirschsprung's Disease

Congenital absence of ganglion cells

Failure migration of neural crest neuroblasts to distal portion of colon
Normally, nerve cells signal muscles in colon to push stool toward anus
Stool moves through until reaches part lacking nerve cells

- Functional obstruction
- Distention
- Bilious vomiting
- Failure to pass meconium
- Constipation - lack nerve cells in part/all large intestine

Males predominate 4:1, 1/5000 live births
30% have family history - "congenital bowel disease"

Hirschsprung's Disease

Diagnosis

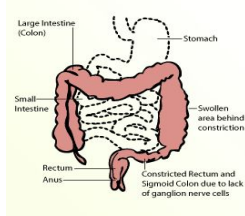
KUB- proximal bowel dilatation and absence air in rectum
Contrast studies - constricted rectum, dilated bowel above & transition zone
Rectal biopsy

Surgical Management

Colostomy with later pull-through procedure
Complete pull-through repair (laparoscopy)
Anal dilatations

Complications

Incontinence, constipation, strictures, rectal stenosis



Imperforate Anus

Anorectal Malformation
Connection to VACTERL complex

Etiology

Failure to differentiate urogenital sinus and cloaca
High type imperforate anus - more complicated
Low type imperforate anus - frequently includes fistula

Treatment

Colostomy initially
Surgery - closure of fistula, creation of anal opening, repositioning rectal pouch into anal opening, creating adequate nerve/muscle structures around rectum/anus to provide bowel control
Many have incontinence issues



Necrotizing Enterocolitis

Necrosis of mucosal/submucosal layers of GI tract
Pathogenesis unknown

Etiology - prematurity, formula feeding, intestinal ischemia, bacterial colonization

Presentation - abdominal distention, emesis, **bloody stools**, discolored abdomen, systemic instability (A's & B's, poor perfusion, lethargy), risk of perforation

KUB - ileus, **pneumatosis**, dilated loops, thick bowel wall, pneumoperitoneum, portal venous gas

Labs - neutropenia, thrombocytopenia, metabolic acidosis, glucose instability

Management - Bowel rest, gastric decompression, broad-spectrum antibiotics, serial KUBs (free-air), CBCs, electrolytes, blood gases/lactates

Surgery - pneumoperitoneum

Complications - strictures



Necrotizing Enterocolitis

Bell Staging Criteria

Stage I Suspected NEC

Temperature instability, apnea & bradycardia, mild abdominal distention, bloody stools, normal or mild ileus KUB

Stage II Mild NEC

Stage I + prominent abdominal distention, +/- tenderness, absent BS, grossly bloody stools, dilated bowel loops with focal pneumatosis on KUB

Stage IIB Moderate NEC

Mild acidosis, thrombocytopenia, abdominal wall edema and tenderness, extensive pneumatosis, +/- portal venous gas

Stage IIIA Advanced NEC

Respiratory/metabolic acidosis, mechanical ventilation, hypotension, oliguria, DIC, worsening wall edema, prominent ascites, no free-air

Stage IIIB Advanced NEC

Deterioration (VS and labs), shock, evidence of perforation, and pneumoperitoneum on KUB



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Short Bowel Syndrome

Chronic malabsorption and malnutrition due to bowel shortening

Etiology

50% from NEC, jejunal/ileal atresia, midgut volvulus, gastroschisis, omphalocele

Prognosis

Dependent on bowel length (15-20cm with ileocecal valve) or (30-45cm without ileocecal valve) and site of intestinal loss

Management

Stabilize fluid/electrolytes, TPN support, gradual increase feedings, supplemental vitamins (ADEK, B12 if ileum lost)(B12 if stomach lost)

Surgical management

Small bowel transplant (may include liver)



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Ostomy Care

Assessment - Ostomy output (goal <20 ml/kg/d)

Consistency - seedy/liquid

Weight gain/loss

Electrolyte imbalance

Stoma - beefy red, moist, measure length

Complications - prolapse, inverted stoma, bleeding stoma, infection (yeast), skin irritation

"Dumping" - may need elemental formulas, continuous feeds, decrease feedings, use supplementation TPN/ILs to grow

May need replace output - 0.9%NaCl/0.45%NaCl over 4-6 hours

Surgical reanastomosis - 6 weeks

Nursing Care - daily weights, weigh all output, monitor labs, skin care

Manage bacterial overgrowth - often occurs when ileocecal valve absent



Ascension

Biliary Atresia

Biliary Atresia

Blockage or complete lack of bile duct - drains bile from the liver

Bile builds up inside liver - damage and eventually liver failure

Symptoms

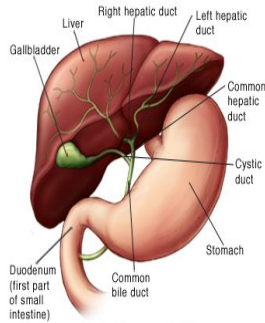
Jaundice, acholic stools, dark urine, portal hypertension

Diagnosis

HIDA scan, liver biopsy

Treatment

Surgery (Kasai procedure), most successful before 8 weeks of age, 80% require liver transplant



Ascension

Gastroesophageal Reflux (GER)

Retrograde movement of gastric contents into esophagus

Symptoms - vomiting, growth retardation, aspiration pneumonia, esophagitis

Risks - prematurity, high bowel obstruction

Treatment

Small frequent feeds/thickened feeds

Positions - held upright for 30 minutes after feeding

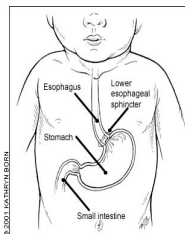
Supine when asleep per AAP guidelines

Resolves spontaneously by 12-18 months

Severe cases

Prokinetics, PPI/H2 antagonists

Nissen fundoplication (rare)



Ascension

GI Bleeding/Perforation

Anal fissure - most common cause GI bleeding- tear of mucocutaneous tissue, red blood streaks on stool

Other causes GI bleeding

Swallowed maternal blood

Allergic colitis - milk protein allergy

Indomethacin (treatment PDA) - intestinal vasoconstriction, platelet dysfunction

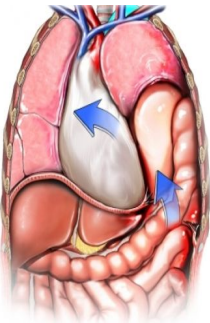
Maternal medications - placental transfer of ASA, phenobarbital - coagulation abnormalities)

Stress gastritis/ulcer- prematurity, mechanical ventilation, dexamethasone

Less common causes: bacterial/viral enteritis (Shigella, E. Coli), coagulopathies (DIC), NEC, Hirschsprungs, Meckel diverticulitis, liver disease

Treatment - Vitamin K prophylaxis, Apt test (tests mother vs baby), sepsis evaluation, liver and coagulation testing, surgical/hematology/GI consult, antibiotics, hypoallergenic formula, blood transfusion, endoscopy/biopsy

Congenital Diaphragmatic Hernia



Bochdalek Hernia

Most common kind of CDH - 95% cases
Hole in posterior lateral corner of diaphragm
Abdominal viscera moves into chest cavity
Majority (80-85%) occur left sided

Morgagni - <5% anterior

Presentation

Diminished breath sounds, shift heart tone, barrel chest, scaphoid abdomen

Complications

Pulmonary hypoplasia
Pulmonary vasoconstriction
Pulmonary hypertension

Practice

The infant you are caring for is suspected of having Hirschsprung's disease. What diagnostic procedures do you anticipate the infant to undergo prior to treatment of this disease?

- 1) Plain abdominal films and blood work including amylase and lipase
- 2) Rectal biopsy, plain AXR and barium enema
- 3) Rectal biopsy and MRI

Practice

Which of the following is the most common cause of intestinal obstruction in the newborn?

- 1) Intussusception
- 2) Meconium ileus
- 3) Intestinal atresia

Practice

A previously well full-term infant presents with bilious vomiting. What is the first disease process that the infant should be evaluated for?

- 1) Pyloric Stenosis
- 2) Sepsis
- 3) Malrotation with Midgut Volvulus

Practice

A full term baby with Down's syndrome has started vomiting. You notice his abdomen is becoming distended. Which of the following conditions may be the cause of this problem?

- 1) High meconium plug
- 2) Malrotation and volvulus
- 3) Duodenal atresia

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