Intestinal Obstruction in Newborns

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Morbidity and Mortality Conference
Case Presentation

- SG - 10 day old baby girl, born at 30 weeks gestation
- Associated with an in utero twin fetal demise
- Apgar score 2, 7 (1, 5 min); Birth weight 1.3 kg
- PMH: patent PDA
- Increasing abdominal distention and obstipation for first 4 days of life
Physical Exam

- VS: T 97.8 F  BP 68/34  HR 164  RR 16  O₂ sat 91%
- CV: RRR, S1S2 normal
- Pulm: CTA bilaterally; intubated
- Abdomen: very distended with visible bowel loops, soft, nontender
- Rectal: unable to go beyond 1.5 cm

Labs:
- CBC: 3.1/12/36/261
- BMP: 138/3.8  108/24 8.4/0.7 <158
- PT/INR/PTT: 13/1.33/29.8
Radiologic Studies

- **AXR**: dilated small bowel loops with no air in pelvis
- **Barium enema**: microcolon with only left colon visualized
- **Upper GI**: no progression of contrast beyond the stomach consistent with marked ileus
Intraoperatively

- 6/9: Exploratory laparotomy
  - Reduction of jejunal volvulus
  - Intestinal resection of atretic segments
  - Tapering enteroplasty of the dilated reduced bowel with primary anastomosis
Meconium
Hospital course

- Abdomen remained nondistended and soft
- No bowel movement for 3 ½ weeks except for mucus plugs
- Upper GI series: dilated proximal loops of bowel
- 7/5: Exploratory laparotomy, adhesiolysis, small bowel resection, enterorrhaphy (from staple line) and intestinal decompression with removal of meconium plugs
Hospital Course

- Post operatively no bowel movement for 2 weeks
- 7/19: exploratory laparotomy, adhesiolysis, intestinal decompression through enterotomy, jejunal resection of previously volvulated segment
- Meconium passed on POD #3.
- Extubated currently with normal bowel function.
Intestinal Obstruction in the Newborn

- Think of...
  - Atresia
  - Malrotation with volvulus
  - Meconium ileus
  - Hirschsprung’s disease
Differential Diagnosis in neonate with abdominal distention and obstipation

- Meconium Ileus
- Distal jejuno-ileal atresia
- Hirschsprung’s Disease
Intestinal Atresia

- Congenital obstruction caused by complete occlusion of intestinal lumen
- Mesenteric vascular accidents in utero
- Multifactorial
- Incidence:
  - Jejunoileal: 1 in 330 (US) - 1 in 1500
- Increased in maternal use of pseudoephedrine and ergotamine + caffeine
Clinical Presentation

 Symptoms:
 - Bilious emesis - proximal
 - Abdominal distention - distal
 - Maternal polyhydramnios (24%)
 - Failure to pass meconium

 Prenatal Ultrasound
 - More detectable in duodenal atresia
 - Multiple distended loops of bowel with vigorous peristalsis
 - Echogenic bowel
 - <1/3 cases recognized
Radiographic findings

- High Jejunal Atresia
- Few air fluid levels
Ileal Atresia
Intestinal Atresia

Classifications

- **Type I:** mucosal atresia with intact bowel wall and mesentery
- **Type 2:** 2 atretic blind ends joined by fibrous cord + intact mesentery

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Intestinal Atresia

Classifications

Type 3:
- Type 3A: two ends of atresia separated by V shaped mesenteric defect
- Type 3B: “Christmas tree” deformity
  - Bowel distal to the atresia receives its blood supply in a retrograde fashion from ileocolic or right colic artery

Type 4: multiple atresias
- “String of sausage” or “string of beads”

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Type 3 Intestinal Atresia
Operative Interventions

- In proximal jejunal atresia - resection at ligament of Treitz followed by end – to- oblique anastamosis
- If limited length - tapering enteroplasty
  - Retention of dilated blind proximal segment - functional obstruction
  - Smooth muscle hypertrophy and enlargement of bowel diameter
  - Ineffective peristalsis
Attempting to preserve bowel length
Morbidity and Mortality

- Most common cause of early death: infection related to pneumonia, peritonitis or sepsis

- Postoperative complications:
  - Functional intestinal obstruction at anastomosis
  - Anastamotic leak (15%)

- De Lorimier and associates concluded that resection improved survival in jejunal atresia from 39% to 66%.
  - Little effect on overall survival in ileal atresia
Hirschsprung’s Disease

- Incidence: 1 in 5000 live births
- M:F ratio 4:1
- Among families of children with HD incidence ↑ to 6%
- Associated with trisomy 21 (4.5-16%) atresias
Any child with constipation dating back to newborn period
  90% diagnosed as newborns

History of delayed passage of meconium \texttt{within 48 hours of life}

Other si/sx:
  Abdominal distention absent at birth and tight anus
  Poor feeding
  Emesis
Radiographic Studies

- AXR- distended loops of intestine with paucity of air in rectum

- Contrast enema (76-92% accuracy)
  - Narrow spastic distal intestinal segment with a dilated proximal segment
  - **the point of caliber change is the key radiographic finding**
  - Most commonly the transition point at rectosigmoid
Hirschsprung’s Disease

Constipation
Anorectal Manometry

- Measures absence of a relaxation reflex after a distending bolus is created in the rectal lumen
- Elevated resting anal sphincter pressure
Rectal Biopsy

- GOLD STANDARD for diagnosis of Hirschsprung’s disease
- Diagnostic accuracy 99.7%
- Alternate method: full thickness posterior rectal wall biopsy
Cause of Hirschsprung’s Disease

- Sporadic occurrence accounts for 80-90% of cases
- RET gene mutations in 35% of sporadic cases and 49% of familial cases
- 5-10% show mutations in other genes
Embryology

3 phases in development of enteric nervous system:

- **Induction phase**
- **Neural crest cell migration**
- **Differentiation of the neural crest cell precursors**

- 5 wks gestation- NC cells in esophagus
- 7 weeks- midgut
- 12 weeks- distal colon
Theories of embryologic defect:

- “Failure of migration”
- “Hostile environment”
In infancy the intestine may appear fairly normal.

As infant ages- proximal ganglionic bowel hypertrophies.

Rectosigmoid – 80%

Histology:
- Absence of ganglion cells in the distal intestine
Endorectal Pull-Through

- Single stage operation; Initially described by Soave in 1964
- LLQ incision
- At least 5 cm proximal to the first area of normal ganglion cells
- Endorectal dissection approximately 2 cm below peritoneal reflection by incising the seromuscular layer
  - Carried down 0.5cm of the dentate line in newborns
Endorectal Pull through -2

- Evert mucosal-submucosal tube and incise on anterior half
- Normal ganglionic intestine brought down to this point
- Anastamosis with absorbable sutures
Different surgical techniques

Laparoscopic

Transanal
Complications

- **Early Post-Pull-Through**
  - Intestinal obstruction (8-13%)
  - Early anorectal stenosis (10-20%)

- **Late**
  - Incontinence (3-8%)
  - Constipation (6-30%)
Meconium Plug Syndrome

- Confirmed by contrast enema radiograph to find “plugs” in sigmoid or descending colon
- Spontaneously pass after withdrawal of the enema catheter
- Pathogenesis may relate to bowel hypomotility
- Associated with: prematurity, hypotonia, hypermagnesemia, sepsis, hypothyroidism, and Hirschsprung’s disease in 5%
- Sweat test and rectal biopsy
Meconium Ileus

- 1905- Landsteiner

- Seen in 20-35% patients with **cystic fibrosis**
  - Mutation in CF transmembrane regulator (CFTR) gene

- Patients with meconium ileus represent a distinct phenotype
  - Earlier presentation and worse pulmonary function

- Survival 95-100% with aggressive management, nutrition and close monitoring of pulmonary function
Intestinal obstruction secondary to intraluminal accumulation of inspissated and dessicated meconium

EARLIEST clinical manifestation of CF- 20.8%

CFTR- chromosome 7, band q31 (F508 mutation)
  Cyclic adenosine monophosphate-induced chloride channel

Reduced clearance of secretions
  Respiratory, GI, biliary, pancreatic and reproductive systems
Pathogenesis of Meconium Ileus

- Intestinal glandular disease plays a dominant role
  - Pancreatic disease plays a secondary role

- Meconium in CF patients is twice as high in concentrations of sodium, potassium and magnesium; \( \uparrow \) protein nitrogen

- Degradation enzymes

- Protein + mucopolysaccharides \( \rightarrow \) highly viscid rubbery meconium

- Complications: volvulus of heavy loop with perforation, peritonitis, atresia
  - microcolon
Meconium Ileus

- **Uncomplicated**
  - Presents immediately at birth
  - Abdominal distention, bilious emesis, failure to stool

- **Complicated**
  - Presents in utero or postnatally
  - Bowel obstruction with evidence of perforation and/or necrosis
Clinical Features

- Family history CF 10-33%
- Polyhydramnios
- Physical Exam:
  - Abdominal distention
  - Visible peristaltic waves
  - Palpable doughy bowel loops
  - Putty sign
Radiologic Studies

- **In utero:**
  - echogenic bowel in 3rd trimester;
  - distended bowel

- **Abdominal XR:**
  - Great disparity in size of bowel loops
  - NO or few air fluid levels
  - “soap bubble” or “ground glass” in meconium peritonitis

- **Contrast Enema**-
  - microcolon
Lab studies

- **Labs:**
  - Sweat test- 100mg
  - Sweat Cl >60 mEq/L

- **Stool Studies**
  - Meconium albumin>80mg/g
  - Stool trypsin >80 mg/g
Meconium Ileus
Non-operative Management

- **Gastrograffin enema**
  - Hyperosmolar, water-soluble, radioopaque solution
  - 0.1% polysorbate 80 (solubilizing agent) and 37% organically bound iodine
  - Transient osmotic diarrhea and a putative osmotic diuresis

- **Advantages**: reduction in pulmonary morbidity, decreased hospital length of stay

- **Disadvantages**: delay, intestinal injury, hypovolemia
Operative Management

- Enterotomies with irrigation
  - Warmed saline, 50% diatrizoate solution, H₂O₂, Mucomyst
  - Meconium milked distally into the colon or enterotomy
  - Enterostomy + T-tube
    - Continued irrigation until POD #14 where catheter removed

- Mikulicz double-barreled enterostomy

- Santulli – proximal chimney enterostomy
Bishop-Koop procedure

- Limit intraoperative bowel trauma in neonatal period
- Resect disparately enlarged ileal loop filled with meconium
- Create an approximately sized end of prox to side of distal ileum
- Access to the insertion of a catheter into the distal bowel containing the meconium pellets
- Permit eventual enterostomy closure by bedside ligation of “chimney” stoma
Various Techniques

End-to-end anastomosis

Mikulicz resection

Proximal

Bishop-Koop resection

Santulli resection

Fig. 69-2 Operative options for the surgical treatment of meconium ileus.
Postoperative Management

- Instillation of 2%-4% acetylcysteine delivered through a nasogastric tube to solubilize the residual meconium
- When gut patency verified – elemental formula
- Supplemental pancreatic enzymes also begun with formula
- Short term post operative antibiotics
- Total parenteral nutrition
Survival rates approaching 100%

No overall significant differences in outcome was present with regard to patient gender, complication of meconium ileus or type of operation performed.
Hypothesis: the role of contrast enema in simple meconium ileus is not as effective as previously reported (30-50% reduction)

- 37 cases of simple MI over a 12 year period (16M:21F)
  - 22/37 patients had CF (89%)
- 8/37 cases (22%) had successful relief of obstruction
  - 3/8 required one attempt only
Demographics and Results

No differences between groups in demographic variables

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<thead>
<tr>
<th>Table 1</th>
<th>Demographics of simple MI patients</th>
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<tbody>
<tr>
<td></td>
<td>Contemporary group</td>
</tr>
<tr>
<td>Gestational age (wk)</td>
<td>36 ± 3.8</td>
</tr>
<tr>
<td>Birth weight (kg)</td>
<td>3 ± 0.9</td>
</tr>
<tr>
<td>Male (n)</td>
<td>10</td>
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<tr>
<td>Age at diagnosis (d)</td>
<td>4.7 ± 6</td>
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Significant decrease in successful resolution of obstruction by enema

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<tr>
<th>Table 2</th>
<th>Results of contrast enema in simple MI patients</th>
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<tr>
<td></td>
<td>Contemporary group (n = 18)</td>
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<tr>
<td>Successful reduction</td>
<td>1 (5.5%)</td>
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<tr>
<td>No. of attempts</td>
<td>1.4 ± 0.7</td>
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Discussion

- Low perforation rate with a high failure rate of enema.

- Enema substrate used in this study (cysto-conray II- 400 mOsm/kg water) less osmotically active than Gastrografin (1940 mOsm/kg water).

- Small sample size- need a prospective multicenter evaluation.
References


- **Schwartz’s Principles of Surgery.** Chapter 39: PEDIATRIC SURGERY

- **Dalla V, Grosfeld JL, West KWI. Intestinal atresia and stenosis: a 25 year experience with 277 cases. Arch Surg. 1998 May; 133 (5) 490-6.**

- **Current Diagnosis and treatment, Surgery. 13th edition. Pediatric Surgery**
Normal Rotation and Fixation of Duodenojejunal Loop
Normal Rotation and Fixation of Cecocolic loop

0°

90°

ao.

S.M.A.
Normal Rotation and Fixation of Cecocolic loop
4th week of fetal life the embryo is at 5 mm stage
- Intestinal tract is a straight tube with slight anterior bulge in central portion

8th week
- Duodenojejunal rotates during extracoelomic phase to 180 degrees

10th week
- Intestines return to abdomen
Malrotation

- All abnormalities of intestinal position and attachment
  - **Atypical malrotation** - ligament of Treitz is to the left of the midline
  - **Nonrotation**
  - **Incomplete rotation**
Acute Midgut Volvulus

- Narrow pedicle formed by the base of the mesentery in malrotation predisposes midgut to clockwise twisting from duodenum to transverse colon

- Various causes:
  - Unusual movement of torso
  - Abnormal intestinal peristalsis
  - Segmental bowel distention
Midgut Volvulus

**Acute Midgut Volvulus**
- First month of life
- Bilious emesis
- As vascular compromise progresses → intraluminal bleeding
- Crampy abdominal pain
- Complete obstruction → distention, hypovolemia, shock

**Chronic Midgut Volvulus**
- Children > 2 years
- Lymphatic and venous obstruction with enlargement of mesenteric LN
- Sx:
  - chronic vomiting,
  - intermittent colicky abdominal pain
  - diarrhea
  - hematemesis
Duodenal Obstruction

- Ladd’s bands extending across the 3rd portion of duodenum
- Most common in neonates
- Forceful bilious emesis
- “double bubble sign”
- **Upper GI contrast study**
Radiologic Diagnosis

- Contrast radiography

Normal

MIDGUT VOLVULUS
“Z sign”- incomplete rotation and broad peritoneal bands extending across and fixing the involved small intestine without an accompanying volvulus.

- Midgut Volvulus
- Z sign
- Colon Malposition
(1) Evisceration of the bowel and inspection of mesenteric root

(2) Counterclockwise derotation of midgut volvulus

(3) Lysis of Ladd’s peritoneal bands with straightening of the duodenum along the right abdominal gutter

(4) Appendectomy

(5) Placement of the cecum in the left lower quadrant
Ladd Procedure

- Supraumbilical right transverse incision
- 2 constant anatomic points: Pylorus and splenic flexure
At 12-24 hours recovery of questionable bowel or demarcation is obvious

3 principles should be considered:
- Preserving minimum length of intestine required for survival is of highest priority
- Avoid anastomoses between end of intestine of questionable viability
- Resection of entire midgut will necessitate lifelong parenteral nutrition and small intestinal transplantation
Postoperative Management

- In uncomplicated duodenal obstruction- peristalsis in 1-5d

- Complications:
  - Postoperative intussusception (3.1%)
    - Abdominal distention and bilious emesis on POD # 5-8
  - Recurrent volvulus (0.5-1.25%)
  - Bowel obstruction (4%)
  - Death (associated with peritonitis)