Jejunoileal Atresia

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Case Presentation

- Full term male.
- C-section due to breech presentation.
- Apgar 8/9.
- Birth weight- 3.2 kg
- Small omphalocele. Umbilical cord with 3 vessels identified.
- Arthrogryposis multiplex congenita

- Mother is 19 y/o. No PMH.
- Uncomplicated pregnancy.
- Fetal U/S x 4 were negative
Case Presentation

- Hospital Day (HD) #1
  - NPO/ IVF’s
  - Labs WNL.
  - Pediatric Surgery consulted.
- HD#2-5
  - Abdominal distention.
  - Bilious vomiting.
  - Not passed meconium.
  - NPO, OGT placed, TPN.
  - Ampicillin and Gentamicin.
  - Echo- negative.
  - Karyotype XY
Case Presentation

- Contrast Enema
  - Microcolon that ends in LLQ.
  - Terminal ileum not visualized.
Contrast Enema
Exploratory Laparotomy

- RUQ transverse incision.
- Dilated distal ileum.
  - Enterotomy and evacuation of meconium.
- Omphalocele with incarcerated loop of ileum.
- Atresia of terminal ileum, absence of cecum and ascending colon.
- Ladd’s procedure for Malrotation of intestines.
Exploratory Laparotomy

- Resection of dilated distal small bowel.
- Colon assessed for patency.
- End-to-end two layered anastomosis between ileum and transverse colon with 4-0 PDS suture.
- Mesenteric defect closed with 4-0 PDS suture.
- Repair of abdominal wall defect.
Post-op Course

- POD#1
  - Extubated
  - On TPN
- POD#5
  - +BM
- POD#6
  - OGT removed
- POD#7
  - Enteral feedings started
- POD#20
  - TPN discontinued
- POD#28
  - Discharged
Jejunoileal Atresia
Intestinal Atresia

Duodenal

Jejunoileal
Intestinal Atresia

**Duodenal**
- Failure of vacuolization of the duodenum from its solid cord stage.¹
- Associated with
  - Down Syndrome
  - Prematurity
  - Polyhydramnios
  - Malrotation
  - Annular pancreas
  - Biliary atresia
  - Cardiac, renal, esophageal, anorectal anomalies.

**Jejunoileal**
- Intrauterine mesenteric vascular accident.
  - Louw and Barnard (1955)²
  - Vascular insult late in gestation.
    - Volvulus, intussusception, internal hernia, abdominal wall defects.
  - Cystic fibrosis - 10% cases.¹

Jejunoileal Atresia

- Incidence - 1/1,000 to 1/5,000 births.
- Equal male to female ratio.  
- Mortality
  - 90% before 1950
  - 1-10% currently
    - Advances in surgical technique, neonatal intensive care, TPN

Jejunoileal Atresia Type I

- Membranous atresia or web.
- Bowel length is normal.
- Intact mesentery.
Jejunoileal Atresia Type II

- Blind ends of bowel connected by fibrous cord.
- Bowel length usually normal.
- Intact mesentery.
Jejunoileal Atresia Type III A

- Blind bowel loop endings.
- Total bowel length may be affected.
- Mesenteric defect of variable size.
Jejunoileal Atresia Type III B

- Apple peel or Christmas tree
- Occluded SMA
- Proximal bowel
  - Dilated
  - Ends near ligament of Treitz.
- Distal bowel
  - Wrapped around a marginal vessel from ileocolic or right colic artery.
- Significant loss of mesentery and bowel length.
- Malrotation
Jejunoileal Atresia Type IV

- Multiple atresias.
- May be any combination of types.
- Up to 20% of patients have multiple atresias.
Presentation

- Prenatal
  - Ultrasound
    - Signs of small bowel obstruction
      - Dilated loops of bowel.
      - Hydramnios.
      - Not specific for jejunoileal atresia.
    - More reliable for duodenal atresias.
Presentation

- Abdominal distention
  - More prominent with distal ileal atresias.
  - Bowel perforation
    - Abdominal wall erythema
    - Increased tenderness
    - Hemodynamic instability
- Bilious emesis.
- Passage meconium does not rule out atresia.
- Incomplete obstructions may presents later in infancy or childhood.
Differential Diagnosis

- Meconium ileus
- Hirschprung’s disease
- Malrotation
- Volvulus
- Intestinal duplication
- Internal herniation
Imaging

- Abdominal Radiograph
  - Dilated loops of bowel
    - Depend on level of atresia
  - Absence of distal gas.
  - Calcifications
    - Meconium peritonitis consistent with bowel perforation.
Imaging

- Contrast enema.
  - Assess patency of colon
    - Rule-out multiple atresias.
    - Rule-out other causes of bowel obstruction.
  - Assess size of colon
    - Distal atresias may result in microcolon.
Treatment

• Pre-op
  • Maintain IV hydration
  • Electrolyte replacement
  • Orogastric decompression
  • Broad-spectrum antibiotics
Surgical Exploration

- Transverse upper abdominal incision.
- Small bowel is eviscerated and inspected.
  - Site of atresia is identified.
  - Coexisting anomalies are identified.
- Distal limb is cannulated and perfused with warm saline to confirm patency.
- Restoration of gastrointestinal continuity.
Surgical Exploration

- Restoration of gastrointestinal continuity.
  - Goal is to preserve bowel function and length.
  - Proximal loop is dilated
    - Stasis and functional obstruction if anastomosed to distal loop.
    - Size mismatch may lead to technical difficulty in creating the anastomosis.
Surgical Exploration

- Normal bowel length.
  - Dilated proximal loop is resected.
  - End-to-end anastomosis.
  - Closure of mesenteric defect.
- Inadequate bowel length.
  - Tapering jejunoplasty on antimesenteric border.\(^5\)

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Surgical Exploration

- Multiple atresias
  - Normal bowel length and atresias grouped together.
    - Single resection and anastomosis.
- Inadequate bowel length
  - Multiple resections and anastomosis.
Postoperative Care

- Gastric decompression and parenteral nutrition until bowel function returns.
- Overall survival > 90%
- Operative mortality < 1% 4
- Type III B atresias
  - Higher risk of proximal limb necrosis, short gut and malabsorption.

A proposed classification system for familial intestinal atresia and its relevance to understanding of the etiology of jejunoileal atresia

Nicholas A. Shorter, Anthony Georges, Agnes Perenyi, Eugene Garrow
Division of Pediatric Surgery, Department of Surgery, SUNY-Downstate Medical Center, Brooklyn, NY, USA.

- Familial cases of intestinal atresia have been described.
- Classification system for types of familial intestinal atresia.
  - Class 1- Pyloric atresia - Autosomal recessive
  - Class 2- Duodenal atresia- Autosomal recessive
  - Class 3-Hereditary multiple atresia - Autosomal recessive
  - Class 4- Apple peel atresia - Autosomal recessive
  - Class 5- Colonic atresia – X-linked recessive
Etiology of jejunoileal atresia

- Normal embryologic development
  - Intestinal Tube and Blood Vessels
- Embryologic development disrupted by
  - Genetic Mechanisms
- Congenital Malformation of Blood Vessels
- Volvulus, Intussusception, Abdominal Wall Defect
- In-utero Vascular Accident
- In-utero Vascular Accident
- Bowel Necrosis and Absorption
- Intestinal Atresia
Abdominal Wall Defects

Omphalocele

- Abdominal contents
  - protrude through umbilical ring
  - covered with peritoneal membrane

- Associated anomalies
  - Trisomies 13, 18, 21
  - Cardiac
  - Genitourinary

- Treatment
  - Pre-op – keep sac moist
  - Primary closure of fascia
  - Silastic silo and delayed fascial closure
Abdominal Wall Defects

Gastroschisis

- Defect on the right side of umbilical ring.
- Abdominal contents not covered.
- Associated anomalies are infrequent
  - Intestinal Atresia (15% cases)
- Treatment
  - Cover bowel with saline-soaked gauzes
  - Primary closure of fascia
  - Silastic silo and delayed fascial closure
References