Case Presentation

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History of Present Illness

- xx-year-old male presenting with a four-day history of worsening suprapubic abdominal pain.
- Reports nausea, and anorexia, but no vomiting, diarrhea, or constipation.
- Denies dysurea, and any prior hx of similar pain.
- Denies fever or chills at home.
- The patient was seen by PMD two days prior to the admission; sent home with Vicodin.
- Two episodes of bilious vomiting in the ER.
Past Medical History
- Denies
- No history of chronic gastrointestinal complains.

Past Surgical History
- None

Medications
- None

Allergies
- NKDA
Physical Examination

- Temp: 102.2F, BP: 137/75, HR: 100, RR 16
- Neuro – Awake and Alert
- Chest: Clear to auscultation bilaterally
- CVS: RRR, normal S1&S2, no murmurs
- Abdomen: + BS, soft, nondistended, with moderate suprapubic tenderness, no rebound, no guarding
- Rectal: no masses; guaiac negative
**Laboratory Results**

- CBC: WBC 11.3 (Neut-91%), H/H 13/38, Plat 270
- Chem7: Na 134, K 3.9, Cl 100, BUN 17, Cr 0.9, Glu 94
- LFT: TP 6.9, Alb 3.9, AST 18, ALT 18, AP 64, TB 1.0
- Coags: INR 1.3, PT 13, PTT 28
- UA: negative

**Imaging**

- AXR: non-obstructive bowel gas pattern
- CT: appendicitis, and nonrotation or incomplete rotation of the GI tract.
Operative Findings

- The patient was taken for exploratory laparotomy
- Gangrenous appendix was identified in the RLQ
- Ascending colon was fixed to the retroperitoneum at the midline.
- The duodenum formed a C-loop around the head of the pancreas but did not cross the midline.
- Thin bands attached transverse colon to the right lateral abdominal wall.
- Operative procedure: appendectomy, and Ladd’s procedure.
Intestinal Malrotation in Adults

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Historical Perspective

- 1700’s – first reported cases
- 1898 – Mall described normal development of fetal midgut
- 1911 – Waugh first correlated symptoms of malrotation to the embryology
- 1923 – Dott provided classic three stage description of rotation of the entire midgut.
- 1936 – Ladd first described surgical therapy
- 1968 – Delvin reported first cases of adult malrotation.
Embryology

- Fourth week of gestation
  - Primitive intestinal tract forms a loop with the SMA at its axis.
  - Duodenojejunal (DJ) loop lies above the SMA.
  - Cecocolic (CC) loop lies below the SMA.
Embryology

- **STAGE I**: (Weeks 5-10)
  - The DJ & CC loops rotate 90° counterclockwise
  - Brings the DJ loop to the right and CC loop to the left of SMA
Embryology

**STAGE II**: (Weeks 10-12)

- Bowel returns to the abdomen.
- The DJ loop completes 270° rotation.
- The DJ flexure is brought to the left of the SMA.
Embryology

- **STAGE II: (Weeks 10-12)**
  - CC loop completes $270^\circ$ rotation
  - Cecum is brought superior and anterior to the SMA.
Embryology

- **STAGE III:** (Weeks 12-term)
  - Cecum descends to the RLQ.
  - Fixation of intestine to the posterior body wall.
Natural History

- Incidence – unknown
  - Rotational anomalies: 1/200 to 1/500 live births
  - Symptomatic malrotation: 1/6000 live births.
- Among patients diagnoses with malrotation:
  - 46% present in the 1st week of life.
  - 68% diagnosed by 1mth of life.
  - 85% diagnosed by 1year of life.
- Remainder (15%) – clinically silent for variable duration.
- Many patients remain asymptomatic for life.
Types of Intestinal Malrotation

- Nonrotation
  - Bowel returns to coelom
  - Failure to rotate
  - Small bowel to the right
  - Colon to the left
Types of Intestinal Malrotation

- Reversed rotation
  - Rare anomaly
  - 90 clockwise rotation around SMA.
  - Cecum & transverse colon dorsal to SMA.
  - Duodenum anterior to SMA.
Types of Intestinal Malrotation

- Incomplete rotation or Malrotation
  - Premature arrest of normal rotation
  - Ladd’s bands
  - Absent ligament of Treitz

- Narrow mesenteric base
- Prone to clockwise twisting
Types of Intestinal Malrotation

- Fixation
Clinical Features

- Asymptomatic
- Chronic abdominal complaints
- Atypical symptoms from a common abdominal disease.
- Acute obstructive symptoms & signs of impending abdominal catastrophe.
Diagnosis

- Plain Abdominal Films
  - Neither sensitive nor specific
  - Suggestive findings
    - Right-sided jejunal markings
    - Absence of stool filled colon in RLQ
Diagnosis

- **UGI**
  - accuracy > 80%
  - Findings:
    - Right-sided DJ junction or proximal jejunal loops
    - Partial obstruction of the duodenum.
    - Midgut volvulus - “Corkscrew” appearance of proximal bowel
Diagnosis

- Barium Enema
  - Less sensitive and specific than UGI
  - Findings:
    - Cecum high in the RUQ
      - 20% have normal appearing cecum.
    - 6% of patients without malrotation have high riding cecum.
Diagnosis

- Ultrasound
  - Vertical relationship or left-right inversion of SMA & SMV.
  - “Whirlpool sign”
  - Radiologist dependent
Diagnosis

- CT Scan
  - Common findings:
    - Intestinal malposition.
    - Vertical relationship or left-right inversion of SMA & SMV.
    - Underdeveloped or absent uncinate process of pancreas.
Diagnosis

- **Internal hernia:**
  - Small bowel obstruction
  - Underlying malrotation

- **Midgut volvulus:**
  - Whirlpool sign
  - Duodenal obstruction
  - Congestion of mesenteric vasculature
  - Underlying malrotation
Management

- Acute abdominal catastrophe:
  - Rapid fluid resuscitation
  - Exploratory laparotomy

- Ladd’s procedure
- Resection of nonviable bowel
- May need 2nd look operation
Management

- Chronic abdominal complaints:
  - Diagnosis and management are commonly delayed.
  - May have incomplete resolution of symptoms.
  - In majority of patients surgery is appropriate.
  - Reversed rotation
    - rotate midgut 360 counterclockwise
  - Ladd’s procedure
Management

- Incidental finding on evaluation:
  - Based on primary diagnosis
    - If urgent operation needed, address malrotation if
      1. it does not add undue risk to the procedure
      2. patient can tolerate added operative time
    - If elective operation needed,
      Treatment can be incorporated into the operative plan

- Discovered at operation
  - Issue of consent
    - Treat immediate problem
    - Investigate malrotation electively
Management

- Asymptomatic malrotation
  - Most controversial
  - Three views:
    1. no intervention
      - Discussion with patient of nature and risks of condition
    2. operative intervention for everyone
      - “Any risk for midgut volvulus... warrants operative intervention”
    3. selective intervention
      - Based on presence of “high risk” anatomy.
      - i.e. significantly lower risk of volvulus in patients with DJ junction left of midline
Ladd’s Procedure

- Detorsion of midgut volvulus, reduction of internal hernia

- Division of peritoneal bands tethering duodenum, small bowel mesentery, cecum, and mesocolon

- Mobilization of duodenum after division of Ladd’s bands

- Appendectomy

- Placement of cecum in the left abdomen and duodenum inferiorly
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