Standardized Toolbox of Education for Pediatric Surgery

APSA Committee of Education
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Malrotation
Malrotation

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History

- 1mo infant presents to ED after several episodes of bilious emesis and fussiness at home
• What other points of the history do you want to know?

• Duration of symptoms?
• Color of emesis? Post-prandial?
• How much/what has the baby being fed?
• Is the baby making urine, having bowel movements? Has there been any melena or hematochezia?
• Is the baby “lethargic”?
• Does the parent think the baby’s abdomen appears “bloated”, distended, or discolored?
• Pertinent PMH? (Any common associated anomalies such as Trisomy 21, cardiac defects, imperforate anus, h/o intestinal atresia at birth)
• Any medications?
• Relevant Family Hx?
• Relevant Social Hx?
Differential Diagnosis

• Malrotation
• Intestinal stenosis/web (atresia if newborn)
• Large bowel obstruction (Hirschsprung’s disease, imperforate anus, colonic atresia, meconium ileus)
• Intussusception
• Necrotizing enterocolitis or neonatal sepsis
• Gastroenteritis
• Reflux/overfeeding
Physical Exam

• **What specifically would you look for?**
  
  • **Vital Signs:** Is there evidence of shock (hypotension, tachycardia)?
  
  • **Appearance:** Is the capillary refill delayed (>3sec), is the baby lethargic?
  
  • **Relevant Exam findings:** Abdominal distension, peritonitis, abdominal wall erythema, distended stomach but otherwise scaphoid abdomen?
Studies (Labs, Imaging)

• **What labs needed?**
  – Electrolytes
  – CBC with differential
  – Type and Cross
  – Consider coagulation studies and VBG/ABG if patient is in shock

• **What Imaging Needed?**
  – UGI contrast study – MOST IMPORTANT
  – AXR
  – LGI
  – Abdominal US
  – CT scan
Study Results
Study Results
Study Results
Study Results
Case Discussion

• Malrotation
  – 1/6000 live birth incidence, but autopsy results suggest much higher incidence (possibly 1% of population)
  – 90% present within first year of life (75% within 1 month of life)
  – Disease of abnormal rotation or non-rotation of intestines
  – Presentation varies but most severe type is with midgut volvulus which is due to twisting of the abnormally fixated small bowel around a narrow-based mesentery and SMA
Case Discussion

Normal intestinal rotation:
(A) 6 weeks GA, nonrotation
(B) 8 weeks GA, incomplete
(C) 9 weeks GA, incomplete
(D) 11 weeks GA, incomplete
(E) 12 weeks GA, complete

Reference:
Malrotation and Midgut Volvulus

Normal

Malrotation

Red dotted lines represent the path of SMA and its branches.

Black circle demonstrates the clockwise torsion of the midgut on the straightened SMA.
Interval steps before / instead of surgery

- Instances when *emergent/urgent* Ladd’s procedure can be deferred:
  - Asymptomatic or incidentally found rotational anomaly
  - Complex cardiac disease (i.e. heterotaxy) with asymptomatic malrotation
  - Older patients with chronic symptoms but no volvulus

- Each of these may still warrant elective Ladd’s procedure – risk of future volvulus as high as 20%
Operation
Operation

- **Open Ladd’s Procedure**
  - RUQ transverse incision or midline laparotomy
  - Eviscerate intestinal contents
  - Detorse counter-clockwise if volvulus is present
    - Resect grossly necrotic bowel, consider second look laparotomy if questionable viability
  - Release Ladd’s cecal bands
  - Broaden the small intestine mesentery
  - Incidental appendectomy
  - Place small bowel on right and colon on left
Operation

Reference:
Operation
Operation

- **Laparoscopic Ladd’s Procedure**
  - 3-4 port sites (umbilical, R abdomen, L abdomen, +/- epigastric for liver retractor)
  - Controversial to perform in setting of acute volvulus but has been described
  - All steps of Ladd’s procedure except evisceration are performed. Lack of adhesion formation is main criticism but thus far not validated to make a significant difference in the literature
Complications

• **Peri-operative:**
  – Prolonged ileus
  – Wound infection
  – Intussusception
  – Early postoperative small bowel obstruction

• **Long Term**
  – Short bowel syndrome
  – Adhesive small bowel obstruction
  – Rarely, recurrent volvulus
Post-operative Management

• Prolonged antibiotics not necessary
• Await resolution of ileus (longer if bowel resection required or in older patients with chronic symptoms)
  – +/- NG tube decompression
• TPN and durable central line if short gut syndrome postoperatively
Questions

• The diagnosis of bilious emesis in a newborn is which of the following until proven otherwise?
  • (a) Pyloric stenosis
  • (b) Malrotation with or without midgut volvulus
  • (c) Duodenal atresia
  • (d) Hirschsprung’s Disease
Questions

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  - (a) Pyloric stenosis
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  - (d) Hirschsprung's Disease
Questions

• The most important diagnostic test to confirm malrotation is:
  • (a) Abdominal XR (supine and decubitus)
  • (b) Barium enema to evaluate position of cecum
  • (c) UGI contrast study
  • (d) Abdominal ultrasound
Questions

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  - (a) Abdominal XR (supine and decubitus)
  - (b) Barium enema to evaluate position of cecum
  - (c) UGI contrast study
  - (d) Abdominal ultrasound
Questions

• What are the key operative steps (in correct order) described in a Ladd’s procedure to correct malrotation?

• (a) Evisceration, detorse small bowel clockwise, appendectomy, release Ladd’s bands, widen mesentery

• (b) Evisceration, detorse small bowel counter-clockwise, appendectomy, release Ladd’s bands, widen mesentery

• (c) Evisceration, detorse small bowel counter-clockwise, release Ladd’s bands, widen mesentery, appendectomy

• (d) Detorse small bowel clockwise, eviscerate, release Ladd’s bands, widen mesentery, appendectomy
Questions

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  • (c) Evisceration, detorse small bowel counter-clockwise, release Ladd’s bands, widen mesentery, appendectomy
  • (d) Detorse small bowel clockwise, eviscerate, release Ladd’s bands, widen mesentery, appendectomy
Final Discussion/Review

• **Malrotation**
  – 75% present in 1\textsuperscript{st} month of life, 90% within 1\textsuperscript{st} year of life
  – Bilious emesis in newborn is malrotation until proven otherwise
  – UGI is diagnostic test of choice
  – Treatment is a Ladd’s Procedure:
    • Detorse counterclockwise, divide Ladd’s bands, widen mesentery, incidental appendectomy, place small bowel on right, large bowel on left
  – Evaluate for possible associated anomalies:
    • Intestinal atresias, cardiac anomalies, Trisomy 21, Imperforate anus, Meckel’s diverticulum
The preceding educational materials were made available through the American Pediatric Surgical Association.

In order to improve our educational materials we welcome your comments/suggestions:

www.eapsa.org