CdLS Foundation

GI Challenges in CdLS

Carol Potter, MD
Nationwide Children’s Hospital
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Objectives

• Malrotation
• Bowel Obstruction
• Constipation
• Pancreatitis
Malrotation

- Abnormal attachment of the intestine to the back abdominal wall
- Allows twisting of the small intestine around the artery and bowel
- Present in 5% of patients with CdLS
MALROTATION AND MIDGUT VOLVULUS

Ladd’s Bands

Midgut Volvulus
Chronic Volvulus

- Twisting of the bowel enough to cause partial obstruction and vomiting
- Intestine is still viable
- May wax and wane
- Should be immediately fixed
Acute Volvulus

• Complete obstruction of the bowel with ischemia
• Medical Emergency
• Pain, vomiting, very ill
• May result in loss of small intestine
Diagnosis of Malrotation

- Barium studies show malposition of the first part of the small intestine
- Some benign variants may mimic malrotation on UGI
- Most vomiting in CdLS is not going to be malrotation
- You have malrotation or you don’t—-it doesn’t develop
Therapy for Malrotation

- Varies by country
- If associated with vomiting and volvulus on UGI
  - Urgent surgery
- If no vomiting and no volvulus
  - Some elect to wait
- If acutely ill
  - Surgical emergency with out UGI
Questions?
Bowel Obstruction

- Congenital Malformations
- Post surgical complications
- Tube related obstruction
- Constipation
Congenital GI Malformations

- More common in CdLS
- May present with vomiting and feeding intolerance
- Radiology often helpful
- High Index of Suspicion
Pyloric Stenosis
Pyloric stenosis

- Hypertrophy of muscle
- In very young babies
- Relentless vomiting
- Diagnosis with US
- Treated with surgery
Duodenal Atresia

• Presents at birth
• May be associated with pancreas abnormalities
• Requires surgical intervention
Diaphragmatic Hernia

- Bowel is in the chest
- Small ones may not be obvious at birth
Cecal Volvulus
Post Surgical Complications

• Adhesions
  • Scar from surgery
  • May block bowel
  • The more you fix it the worse it is

• Hernia

• Infections

• Perforations

• Tube Complications
Adhesions
Hernia
Tube Migration
Questions?
Constipation

- Very common in CdLS
- They may be having a bowel movement everyday
- They have an underlying dysmotility
- Chronic fecal burden leads to stretching and loss of muscle tone
Normal

Faecal impaction with overflow diarrhoea
Sigmoid Volvulus

- Chronic constipation stretches the colon
- The stool filled colon falls over on itself and twists off the blood supply
- Insidious presentation with abdominal pain, and distension that then becomes fulminant
- Diagnosis with CT scan
- Treated with a scope or surgery
- May reoccur
Sigmoid Volvulus
# Bristol Stool Chart

<table>
<thead>
<tr>
<th>TYPE</th>
<th>Description</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Separate hard lumps, like nuts - difficult to pass, indicates constipation</td>
</tr>
<tr>
<td>2</td>
<td>Sausage-shaped but lumpy, indicates constipation</td>
</tr>
<tr>
<td>3</td>
<td>Sausage-shaped with surface cracks, ideal stools as easier to pass</td>
</tr>
<tr>
<td>4</td>
<td>Sausage shaped, smooth and soft, ideal stools as easier to pass</td>
</tr>
<tr>
<td>5</td>
<td>Soft blobs with clear-cut edges, may indicate diarrhoea and urgency</td>
</tr>
<tr>
<td>6</td>
<td>Fluffy pieces with ragged edges, a mushy stool, may indicate diarrhoea and urgency</td>
</tr>
<tr>
<td>7</td>
<td>Watery, no solids, entirely liquid, may indicate diarrhoea and urgency</td>
</tr>
</tbody>
</table>
Constipation

- Many ways to treat it
- Don’t under treat it
Constipation Therapy

• Pulling more water into the stool
  • Miralax
  • Milk of Magnesia
  • Fiber

• Drinking more water won’t help
  • Colon just absorbs it
Stimulants

- May be needed due to dysmotility
- Senna
  - Liquid, pills, and chews
- Bisacodyl
  - Tablets and suppositories
Fermentation/Gas Production

- Lactulose
- Juice
Questions?
Pancreatitis

- Inflammation of the pancreas

- Causes include
  - Infection
  - Trauma
  - Drug
  - Malformation

- Symptoms
  - Pain and Vomiting
  - Can be very sick

- Diagnosis
  - US
  - Lab work
Pancreatitis - Treatment

- Vigorous hydration (IV)
- Supportive
- Nutrition
- Watch for complications
Surgical Complication

- Adhesions
- Hernias
- May be hard to diagnose
- Fixing them may cause more problems than it solves
Questions?
Thank you for attending!

We hope to see on the next webinar.

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