Cases in Pediatric Gastroenterology

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Disclosure

- I have no relevant financial relationships with the manufacturers(s) of any commercial products(s) and/or provider of commercial services discussed in this CME activity
- I do not intend to discuss an unapproved/investigative use of a commercial product/device in my presentation.

Objectives

- Learn about the organic causes of constipation
- Learn about NASPGHAN guideline for Cyclic Vomiting Syndrome.
- Be familiar with critical issues with magnets ingestions
- Learn about the management of gallstones
Case 1

- 3 Y.O old with hx of constipation since birth.
- Passes one small caliber stool q 3 days. No leaking
- Failed medical therapy
- BE is normal
- Full thickness rectal bx is normal

ROME III criteria

Functional constipation criteria

- Functional constipation (2 or more of following in child at least FOUR years of age with insufficient criteria for dx of IBS and these criteria fulfilled for at least 2 months before Dx)
- < 2 defecations in toilet per week
- At least 1 episode of fecal incontinence per week
- History of retentive posturing or excessive volitional stool retention
- History of painful or hard bowel movements
- Evidence of a large fecal mass in the rectum
- History of large diameter stools that may obstruct the toilet

Organic Causes of Constipation

- Newborn
  - Hirschsprung’s disease
  - Intestinal atresia/stenosis
  - Meconium plug syndrome
  - Small left colon syndrome
  - Meconium ileus
  - Imperforate anus
  - Intestinal volvulus
Hirschsprung’s Disease

- Congenital Aganglionic Megacolon
- Absence of intramural ganglion cells
- 1 in 5000 live births
- 4 males : 1 female
- 85% - Short segment disease.
- 15% - Entire colon (1 male : 1 female)


Clinical Presentation

- Failure to pass meconium within the first 48 hours of life
- Abdominal distention and feeding difficulties
- Bilious emesis from partial bowel obstruction
- Diarrhea: Hirschsprung’s disease–associated enterocolitis
- Colonic perforation: most frequently involving the cecum
- Refractory constipation
- Fecal soiling occasionally may occur

*Harrison M.W. Am J Surg 1986*

Diagnosis

- Barium enema
- Rectal suction biopsy
- Full‐thickness biopsy
- Ano‐rectal manometry
Recto-Anal Inhibitory Reflex

RAIR present with balloon inflation

Treatment

- **(B)** Duhamel-Martin operation: The aganglionic rectum is preserved, the normally ganglionated proximal bowel is brought down behind the native rectum, and the wall between the two is obliterated with a stapling device to create a single common lumen.

- **(C)** Soave-Boley operation: The mucosa and submucosa of the distal rectum is removed, preserving the outer muscle layer, and the proximal ganglionated bowel is delivered through the muscle cuff and sutured just above the dentate line.

Pediatric Gastrointestinal and Liver Disease, 4th Edition
By Robert Wyllie, MD and Jeffrey S. Hyams, MD
Internal Anal Sphincter Neurogenic Achalasia (IASNA)

- Earlier onset of symptoms than functional constipation.
- Less fecal soiling
- Less withholding behavior

- Treatment with Intraspincteric injections of botulinum toxin (dose 15-25U per quadrant)
- Variable response to botulinum injection
- Duration of response ranged from 1 week to 18 months.

Anal Achalasia

- Post-hirschsprung’s disease repair
  Effects of botulinum toxin injection on anal achalasia after pull-through operations for Hirschsprung’s disease: a 1-year follow-up study

- Milk allergy? reversible

  Food intolerance and chronic constipation: manometry and histology study
  Giuseppe Iacono1, Sebastiano Bonamini1, Catagino Scalici1,
  Enrica Margari2, Linda Di Prema2, Magnozzi Bonini3, Gianpaolo Di Gregorio4,
  Davide Nello2 and Antonio Carroccoli1

- Which of the following statements about Hirschsprung’s disease is FALSE?

  a. There are no ganglion cells seen in Auerbach’s plexus.
  b. There is an increased incidence of Down syndrome.
  c. It is more common in girls.
  d. It may be associated with enterocolitis.
  e. It may involve the small intestine
Case 2

- 10-year-old female with recurrent, stereotypic episodes of nausea and vomiting, separated by intervals of symptom-free periods.

- Her symptoms last for 2-3 days, 30-50 episodes per day

- Vomiting, usually begin early in the morning and associated with severe epigastric pain.

**Differential Dx**

**Vomiting**

- **Acute**
- Chronic recurrent
- Cyclic recurrent

- GI
- Infectious: GE, OM, Pharyngitis, Hepatitis, Sinusitis
- GU: Pyelo, UPJ obstruction
- Endocrine: DKA
- Neuro: Concussion, Subdural hematoma, Reye’s syndrome
- Other: Toxic ingestion, Food poisoning
### Differential Dx

#### Vomiting

- **Acute**
- **Chronic recurrent**
- **Cyclic recurrent**

**Chronic recurrent**

- Two-third of recurrent vomiting cohort
- Not acutely ill or dehydrated
- Frequent, >2 episodes/week
- No stereotype
- Daytime
- Ratio of GI to extra-GI causes 7:1
- UGI mucosal injury most common (esophagitis, gastritis)

**Cyclic recurrent**

- One-third of recurrent vomiting cohort
- Severe, dehydrated
- High intensity, >6 emesis/h at peak
- Infrequent, <2 episodes/week
- Stereotypic and early morning
- Ratio of extra-GI to GI causes 5:1
- CVS most common

**Infectious:**

- Sinusitis

**GI:**

- Malrotation with volvulus
- Acute hyponephrosis 2° to UPJ obstruction (Dietl's crisis)

**Endocrine:**

- DKA, Addison’s disease, MCAD deficiency, partial OTC deficiency, MELAS syndrome, Acute intermittent porphyria
- Neuro: Arnold-Chiari malformation, Migraine
Cyclic Vomiting Syndrome

- CVS has been described in all races and ethnicities
- Prevalence is not known. 2 population based studies done in pediatric age group; indicated a prevalence of 0.04% and 1.9% respectively
- Affect all age groups
- Affect girls more than boys (3:2)

Phases of CVS

The interepisodic phase "well phase"
- Symptoms free
- Weeks to months

The pre-emetic "prodromal phase"
- Nausea, palor, tachycardia, acoustic phene etc.
- Minutes to hours (median 1.5h)

The "emetic phase"
- Intense nausea and vomiting
- Hours to days (median 4h)

Recovery phase

CVS and Migraine

Profile of Symptoms in Cyclic Vomiting Syndrome, Abdominal Migraine, and Migraine Headaches

<table>
<thead>
<tr>
<th>Percentage of patients with symptom</th>
<th>CVS</th>
<th>Abdominal Migraine</th>
<th>Migraine Headache</th>
</tr>
</thead>
<tbody>
<tr>
<td>Nausea 90-95%</td>
<td>100%</td>
<td>60-64%</td>
<td>10-20%</td>
</tr>
<tr>
<td>Abdominal pain 85-91%</td>
<td>100%</td>
<td>60-64%</td>
<td>10-20%</td>
</tr>
<tr>
<td>Headache 50-59%</td>
<td>100%</td>
<td>60-64%</td>
<td>10-20%</td>
</tr>
<tr>
<td>Associated symptoms 87%</td>
<td>91%</td>
<td>30-60%</td>
<td></td>
</tr>
<tr>
<td>Lethargy 91%</td>
<td>91%</td>
<td>90-100%</td>
<td>20-60%</td>
</tr>
<tr>
<td>Palor 74%</td>
<td>97%</td>
<td>90-100%</td>
<td>20-60%</td>
</tr>
<tr>
<td>Tenderness 74%</td>
<td>91%</td>
<td>90-100%</td>
<td>20-60%</td>
</tr>
<tr>
<td>Nausea 72%</td>
<td>75-91%</td>
<td>40-100%</td>
<td></td>
</tr>
<tr>
<td>Photophobia 52%</td>
<td>1-42%</td>
<td>27-61%</td>
<td></td>
</tr>
</tbody>
</table>

Central Questions....

• What is the appropriate laboratory, radiographic and endoscopic evaluations?

• Does prophylactic treatment improve long-term outcomes?

• During an acute attack of CVS, does treatment improve outcomes – as compared to no treatment or alternative treatment options?

CVS: Diagnostic Criteria

• 5 attacks total or ≥ 3 over 6 months

• Episodic nausea and vomiting 1h-10d, ≥ 1 wk apart

• Stereotypical pattern w/in individuals

• Vomiting ≥ 4 X/h Return to baseline health between

• Return to baseline health for weeks to months

• No attributable to a disorder

• No attributable to a disorder

4 Red Flags

1. Bilious vomiting, abdominal tenderness and/or severe abdominal pain

2. Attacks precipitated by intercurrent illness, fasting, or high protein meal

3. Abnormalities on neurological examination

4. Progressively worsening episodes or conversion to a continuous or chronic pattern
Diagnostic algorithm (> 2 yrs old) guidelines

GI sx s Triggers Abn neuro
Abd U/ S, ALT, GGT, lipase
Metabolic - LA, NH3, AA, carnitine, Ur ketones, OA

CVS Criteria
Labs - glucose, lytes, BUN, Cr, UGI x-ray
No red flags
Prob CVS
POS: Treat. NEGATIVE
Treat empirically

Cyclic Vomiting Syndrome
ED or Hospital Protocol
- Dark, quiet room, VS q 4-6 h
- If dehydrated, bolus NS 20 mL/kg
- D10 0.45 NS+ KCl @ 1.5X maintenance
- IV ondansetron 0.3 mg/kg/dose q. 6 h
- IV lorazepam 0.05-1.0 mg/kg/dose q. 6 h
- If pain, ketorolac 1.0 mg/kg/dose ≤ 30 mg
- Admit if > 5% dry, no urine X12h Na+ < 130 mEq/L or AG > 18 mEq/L, or unable to stop vomiting

Prophylactic Medications

Clinical Report
North American Society for Pediatric Gastroenterology, Hepatology, and Nutrition Consensus Statement on the Diagnosis and Management of Cyclic Vomiting Syndrome

Prophylactic Medications

Titrate dose q. 1-4 wks X 2 CVS cycles
– ≤ 5 yr: Cyproheptadine 1st line (Pizotifen)
– > 5 yr: Amitriptyline 1st line
– 2nd line: Propranolol

Dosing – Cyproheptadine 0.25 – 0.5 mg/kg/d t.i.d. or q.hs
– Amitriptyline 1.0 mg/kg q.hs
– Propranolol 0.25 – 1.0 mg/kg/d divided t.i.d.
• You are evaluating a 5-year-old girl who has had four attacks of vomiting lasting 4 to 8 hours. What is the recommended initial diagnostic test to be performed before considering a trial of empiric therapy?

a. Brain MRI  
b. Upper gastrointestinal radiographic series  
c. Abdominal ultrasound  
d. Esophagogastroduodenoscopy  
e. CT scan of the abdomen

Case 3:

• A 13-year-old boy with abdominal pain, vomiting, dysphagia and reflux-like symptoms for 2 years.

• Pt had history of Nissen fundoplication at 5 months of age for persistent projectile vomiting.

• A trial of Antacid didn’t help.

• UGI series was normal

Case Presentation:

A cord-like foreign body (FB), 5 cm long in the distal esophagus with irritated, edematous and ulcerated esophageal mucosa. The FB was retrieved without complications and patient had complete resolution of symptoms on follow up.
Foreign Body Ingestion

- Childhood curiosity appear to be the major risk factors for accidental ingestion

- American Association of Poison Control-95,705 cases of foreign-body ingestions in 2011 in patients <20 years (74,725 ≤ 5 year old)

Clin Toxicol 2012; 50:911–1164

Foreign Body Ingestions

- The majority of FB will progress through the GI tract without problem (80-90%)

- The challenge for the clinician is to predict which objects will not pass, or pose risk of a serious complication that would warrant removal!


Foreign Body Ingestions

- High incidence of complications has been reported with:
  - Large objects (greater than 2 cm diameter or 5 cm long)
  - Sharp-ended FB
  - Batteries
  - Magnets
Foreign Body Ingestions - Coins

Quarter: 24 mm
Nickle: 20 mm
Dime: 17.8 mm
Penny: 19 mm

Small coins (15–20 mm) are less likely to get stuck in the esophagus than are larger coins (20–35 mm).

Magnet ingestion

- Increasingly reported in pediatric patients
- In 2012: 93 cases of magnet ingestion (age 1-13 years, at least 372 magnets ingested)
  - Attach to one another across bowel wall
  - Cause necrosis and perforation
  - Deaths recently reported


Magnet Ingestion

JPGN 2012; 54:828
Magnet Ingestion

• 2003-2006 - 20 cases of magnet ingestion and injury in children were reported 75% with bowel perforation.

• 2007 - The U.S Consumer Products Safety Commission (USCPSC) issued the first warning after the death of a 20-month-old-child.

MMWR 2006;55:1296–1300

Magnet Ingestion

• 2009 – USCPSC banned the sale of high-powered rare earth magnets to children younger than 14 years.

• July 2012 - USCPSC came to an agreement with most manufacturers regarding voluntary recall.

JPN 2012;55:239-42

Warning! SWALLOWED MAGNETS ARE DANGEROUS

Magnet ingestion can lead to bowel blockages, damage or even death.

• Take steps to protect your child from swallowing magnets.
• Ask that your child’s medicine be placed in a child proof container.
• Keep children under the age of 6 away from magnets.
• Make sure the magnets are in your child’s possession at all times.
• If children do not have access to magnets, immediately report this to your local or national USCPSC office.
Case 4

- 15-year-old girl presents to ED with a 12-hour history of acutely worsening abdominal pain

- Pain is epigastric/RUQ, constant, sharp, 7/10 pain, radiates to her chest and back.

- Pain is associated with nausea, vomiting x 4, and poor appetite.

### Case 4

<table>
<thead>
<tr>
<th>Blood work</th>
<th></th>
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</thead>
<tbody>
<tr>
<td>ALT</td>
<td>334 IU/L</td>
</tr>
<tr>
<td>AST</td>
<td>250 IU/L</td>
</tr>
<tr>
<td>Total Bilirubin</td>
<td>2.6 mg/dl</td>
</tr>
<tr>
<td>Direct Bilirubin</td>
<td>1.2 mg/dl</td>
</tr>
<tr>
<td>Amylase</td>
<td>580 IU/L</td>
</tr>
<tr>
<td>Lipase</td>
<td>2904 IU/L</td>
</tr>
</tbody>
</table>
Pediatric Gallbladder Disease

- Gallbladder disease in children is evolving and studies suggest an increasing frequency of gallbladder disease and resultant cholecystectomies in children.

- Prevalence in younger than 16 years: 0.15% in 1959, currently 1.9% to 4.0%.

(Pediatrics, 2000;31:411–417)

Pediatric Gallbladder Disease

- Cholelithiasis: Gallstones
- Acute cholecystitis: acute inflammation in GB
- Choledocholithiasis: CBD stones
- Acute cholangitis: bacterial infection of the biliary ducts

(Pediatrics, 2000;31:411–417)

Semin Liver Dis 1990;10:159–170

*JPGN 2006;42:66–70

JPGN, 2000;31:411–417

JPGN, 2000;31:411–417
GB disease: Imaging

• Abdominal US

• Abdominal CT scan

• HIDA scan

• MRCP, EUS

• ERCP


GB Disease: Imaging

• MRCP is indicated in patients with suspected choledocholithiasis, to confirm the presence or absence of common bile duct stones.

• Endoscopic ultrasound (EUS) can give a description of both gallstones and choledocholithiasis as well as pancreatic parenchymal and ductal abnormalities

Clin Imaging 2007; 31:93-101

GB Disease: ERCP

Pediatric Gastrointestinal and Liver Disease, 4th Edition
By Robert Wyllie, MD and Jeffrey S. Hyams, MD
• THANK YOU!