Ins and Outs of Gastrointestinal Disorders: An Update

Kathleen M. Cox, RN, MS, PNP
Pediatric Gastroenterology
Lucile Packard Children’s Hospital at Stanford University

Gastrointestinal Problems in the Developmentally Disabled Population

- Chronic gastrointestinal problems affect majority of children with CP and neurodevelopmental disabilities
- These conditions include:
  - Gastroesophageal reflux 32%
  - Abdominal Pain and Gastritis 32%
  - Constipation 74%
  - Dysphagia 60%
  - Malnutrition 33%


<table>
<thead>
<tr>
<th>MEDICAL DIAGNOSES IN CHILDREN WITH DEVELOPMENTAL DELAY</th>
</tr>
</thead>
<tbody>
<tr>
<td>AGE (YRS)</td>
</tr>
<tr>
<td>-----------</td>
</tr>
<tr>
<td>2</td>
</tr>
<tr>
<td>2</td>
</tr>
<tr>
<td>2</td>
</tr>
<tr>
<td>3</td>
</tr>
<tr>
<td>5</td>
</tr>
<tr>
<td>8</td>
</tr>
<tr>
<td>9</td>
</tr>
<tr>
<td>GT = Gastrostomy tube</td>
</tr>
</tbody>
</table>

MEDICAL DIAGNOSES IN CHILDREN WITH DEVELOPMENTAL DELAY

<table>
<thead>
<tr>
<th>MEDICAL DIAGNOSES IN CHILDREN WITH DEVELOPMENTAL DELAY</th>
</tr>
</thead>
<tbody>
<tr>
<td>AGE (YRS)</td>
</tr>
<tr>
<td>-----------</td>
</tr>
<tr>
<td>14</td>
</tr>
<tr>
<td>14</td>
</tr>
<tr>
<td>15</td>
</tr>
<tr>
<td>16</td>
</tr>
<tr>
<td>16</td>
</tr>
<tr>
<td>18</td>
</tr>
<tr>
<td>18</td>
</tr>
</tbody>
</table>

*SIB - self-injurious behavior
NEW MEDICAL DIAGNOSES IN SEVERELY MR WITH SIB*

<table>
<thead>
<tr>
<th>Age (Yrs)</th>
<th>Diagnosis</th>
<th>Rx, Response</th>
</tr>
</thead>
<tbody>
<tr>
<td>3</td>
<td>lactose intol.</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>dysphagia, otitis, constip</td>
<td>Improved</td>
</tr>
<tr>
<td>8</td>
<td>dysphagia, otitis</td>
<td>Improved</td>
</tr>
<tr>
<td>8</td>
<td>esophagitis, gastritis</td>
<td>Improved</td>
</tr>
<tr>
<td>9</td>
<td>constipation</td>
<td>Improved</td>
</tr>
<tr>
<td>9</td>
<td>bur in nose</td>
<td>Improved</td>
</tr>
<tr>
<td>35</td>
<td>duodenal ulcer</td>
<td>Improved</td>
</tr>
</tbody>
</table>

*SIB - self-injurious behavior

SYMPTOMS WHICH OFTEN HAVE A MEDICAL CONDITION AS A CAUSE

- Symptoms:
  - trouble swallowing
  - vomiting
  - posturing
  - constipation or diarrhea
  - crying/grimacing
  - self-injurious behavior
  - weight loss

Screening Studies

Laboratory Studies
- CBC with differential
- ESR and/or C Reactive Protein
- Chemistry Panel with Albumin, AST, ALT, GGT, Amylase
- Stool: Hemoccult, WBC, pH, Reducing Substance, Fecal Fat, Giardia, H. Pylori Antigen

Imaging Studies
- KUB

PREVALENCE OF GER IN 110 PATIENTS WITH SEVERE MR

- 55.3% had significant GER.
- 64.7% with GER had esophagitis.
- GER correlated with cerebral palsy and use of anticonvulsant therapy.

Case 1: Gastroesophageal reflux

- 5 yr F with severe MR
- Behavior: aggressive, head banging, kicking, hair pulling, whining, arching, feeding resistance
- W/U:
  - screening labs - nl
  - KUB neg
  - UGI: hiatal hernia
- Rx:
  - omeprazole
  - metaclopramide
- Response:
  - marked decrease in self-injurious behavior and improved appetite and feeding behavior

Case 2: Gastroesophageal Reflux

- 3 year old M with Autism, diarrhea, feeding aversion, hoarse tone to voice, disrupted sleep pattern
- W/U:
  - Stools studies + for Giardia
  - ENT Referral
  - Multiple vocal cord polyps and significant erythema of upper airway
- Rx for Giardia with diarrhea resolved
- Upper Endoscopy
  - severe esophagitis
- Nissen Fundoplication
  - resolution of hoarse tone to voice, improved feeding behaviors and longer sleep intervals

Symptoms Suggestive of Gastroesophageal Reflux Disease

- c/o Abdominal pain
  - supine position
  - postprandial:
    - citrus, carbonated, spicy foods, caffeine, chocolate, peppermint
- Respiratory Sx
  - pneumonia, bronchitis
  - chronic lung disease
  - apnea, asthma
  - upper airway congestion
  - coughing

- Esophagitis
  - heartburn, dysphagia, gagging
  - GI bleeding/anemia

- Regurgitation & vomiting

- Other
  - dental erosion
  - horse tone to voice
  - malodorous breath
  - c/o sore throat, globus sensation

Assessment of Developmentally Disabled Individual

- Observation of behaviors
- Identification of any associations: behaviors, eating behavior, position, medications, surgeries, illness
- Associated physical symptoms
- Alleviating and aggravating factors
### Gastroesophageal Reflux Evaluation

- Upper GI series
- Esophagram
- Modified Barium Swallow
- Upper endoscopy
- 24 hour ph probe
- Tc99m Technesium Gastric Emptying Study

### GI X-ray Studies to Evaluate GER Symptoms, Swallowing Difficulty or Vomiting

- Liquid & solid barium meal swallowing study
- Esophagram
- UGI series
- Small bowel follow-through

Purpose:
Evaluation for oropharyngeal coordination, mucosal abn. (esophagitis, stricture) anatomical abn. (vascular ring, malrotation, hiatal hernia).

### Upper Endoscopy in Children With Gastroesophageal Reflux

- Upper endoscopy
- Esophageal biopsies
- Gastric biopsies

Purpose:
Evaluates for esophagitis, allergies, anatomical abnormalities (hiatal hernia) and gastritis.

### 24 hr. Esophageal pH Study

- 24 hr. esophageal pH study

Purpose:
Quantitates severity of GE reflux and correlates GER with symptoms (cough, pain, SIB).
**Tc$^{99m}$ Solid & Liquid Meal Gastric Emptying Study**

- **Purpose:** Evaluates for delayed gastric emptying, quantitates GE reflux, & detects aspiration.
- **Quantitate GE reflux in esophagus over 1 hr.**
- **4-12 hr. follow-up scan of lung to detect microaspiration.**

**Summary of Standard Treatments for GER**

- **Dietary**
- **Feeding techniques**
- **Gastric acid inhibitors:** Antacids, H$_2$ blockers, proton pump inhib.
- **Prokinetic agents:** metaclopramide, erythromycin
- **Nasogastric, nasojejunal, gastrojejunal tube feeding**
- **Surgery - fundoplication**
- **Endoscopic therapies**

**Helicobacter Pylori - Gastritis**

- **Cause of abdominal pain, gastritis, ulcer disease and associated with gastric carcinoma**
- **Transmitted via oral – oral route**
- **Incidence of H. Pylori is higher in children and adults in residential populations than in the general population* **
- **Chronic gastritis may be due other causes:** medication induced postviral allergy


**Diagnosis of Helicobacter Pylori**

- **Diagnostic Studies**
  - Upper gastrointestinal endoscopy with biopsy for histologic identification -gold standard
  - Urea breath testing
  - Stool antigen
  - Serologic IgG Immunoassay
**Treatment of *Helicobacter Pylori* Infection**

- **Triple therapy**
  - 1 to 2 weeks of 2 of the following antibiotics: amoxicillin, tetracyline (not to be used under 12yrs.), metronidazole or clarithromycin
  - Plus: Bismuth, an H2 blocker or a proton pump inhibitor

**Case 3: *H. Pylori* Infection**

- **8 yr F with severe MR**
- **Behavior:** arm biting, head banging, hitting, kicking
- **W/U:**
  - Endoscopy - *H. pylori* with ulcers
  - Tc⁹⁹m gastric emptying - delayed
- **Rx:** PeptoBismol, metranidoazole, amoxacillin, omeprazole, cisapride
- **Response:** SIB reduced from 28% to <1%


**Prevalence of Constipation in People with Severe Intellectual Disability***

- **69%** of 215 with severe MR had constipation (<3 BMs/wk or use of laxatives >3 times/wk).
- Significant correlation with:
  - Non-ambulatory
  - Use of anticonvulsants, benzodiazepines, Baclophen, H₂ receptor antagonist or PPI
  - Food refusal
  - I.Q. < 35


**Definition: Constipation & Encopresis**

- **Constipation -** Stools characterized as hard, large, infrequent, painful, and/or difficult to pass.
- **Normal frequency and consistency of stooling varies considerably from 3/day to 3/week.**
- **Encopresis -** Fecal incontinence. May be voluntary or involuntary.
**Constipation in the Developmentally Disabled**

- **Contributing factors**
  - Inadequate dietary fiber and fluid
  - Physical inactivity
  - Defective intestinal innervation
  - Gastointestinal dysmotility
  - Poor muscle tone/coordination
  - Hyper or hypotonia
  - Lack of erect posture
  - Lack of urge to defecate

**Case 4: Constipation**

- 9 yr M with severe MR
- Behavior: hand biting, hitting face, scratching chest, head banging, decreased communication skills
- Rx: clean out: enemas, suppositories oral laxatives on a daily basis
- Response: “decreased SIB & improved appetite & communication”
- W/U: UGI – severe constipation

**Case 5: Constipation**

- 2 1/2 yr. old female with cognitive delay, nonverbal, gastrostomy dependent for nutrition, blind, nonambulatory, diabetes insipidus. Constipation since birth refractory to rx.
- Rectal exam: decrease sphincter tone, absent anal wink
- W/U
  - Anorectal manometry
  - Low sphincter pressure, lack of response to rectal distention.
  - MRI: arachnoid cyst, syringomyelia T5-T7
- Neurosurgical repair
  - Resolution of constipation, increased mobility

**Common Associated Presenting Complaints of Constipation**

- Stool incontinence
- Abdominal pain
- Abdominal distention
- Diarrhea
- Rectal bleeding
- Rectal pain
- Failure to thrive, loss of appetite
- Vomiting
- Irritability
- Lethargy

Functional fecal retention

Physical examination
- abdominal exam, palpation of the lumbosacral spine, DTR’s of lower extremities, anal wink, rectal exam
- Abdominal x-rays
- Lumbosacral films
- Anorectal manometry and EMG
- Barium enema
- Sigmoidoscopy and rectal biopsies
- MRI of the spine

Diagnostic Evaluation of Constipation & Encopresis in Children

Abdominal x-rays

Treatment of Fecal Retention
- High fiber & high fluid diet
- Clean out: Fleets enemas, Biscodyl suppositories, Mg citrate, Miralax or Golytely.
- Meds: stool softers: mineral oil, MOM, sorbitol, Miralax/Glycolax or lactulose stimulants: Biscodyl tablets or Senna.
- Fiber: Metamucil, Citrucel, Benefiber
- Reinforcement: regular toileting, rewards
High Fiber & High Fluid Diet for Constipation

<table>
<thead>
<tr>
<th>AGE</th>
<th>FIBER</th>
<th>WEIGHT</th>
<th>FLUID</th>
</tr>
</thead>
<tbody>
<tr>
<td>1-3 yrs.</td>
<td>5-7 gms.</td>
<td>0-10 kg.</td>
<td>100 cc/kg</td>
</tr>
<tr>
<td>4-6 yrs.</td>
<td>10 gms.</td>
<td>10-20 kg.</td>
<td>75-90 cc/kg</td>
</tr>
<tr>
<td>7-10 yrs.</td>
<td>15 gms.</td>
<td>20-30 kg.</td>
<td>60-70 cc/kg</td>
</tr>
<tr>
<td>11-14 yrs.</td>
<td>20 gms.</td>
<td>&gt;30 kg</td>
<td>40-50 cc/kg</td>
</tr>
<tr>
<td>15-18 yrs.</td>
<td>20-25 gms.</td>
<td>1 kg = 2.2 lbs.</td>
<td></td>
</tr>
</tbody>
</table>

Treatment for symptoms refractory to medications and dietary management

- Behavioral Psychological consultation
- Behaviorist consultation
- Dyssynergia Biofeedback
- Persistent megacolon or neurogenic colon Partial colectomy, appendicostomy, cecostomy

Feeding Difficulties - Dysphagia

- Feeding difficulties are common in individuals with developmental disabilities.
- Oral-motor dysfunction is present majority (90%) of children with cerebral palsy *
- 20-30% of hemiplegic and diplegic children with CP are underweight for age *
- Chronic pulmonary aspiration affects 41% of children with neurodevelopmental disabilities **

**Del Guidice: Brain & Development 21: 307-311, 1999

Spectrum of Feeding Issues

- Oral/pharyngeal motor dysfunction
- Oral sensitivity
- Feeding Aversion
- Insufficient dietary intake
- Insufficient fluid intake
- Increased nutrient losses
- Alterations in Energy requirements
- Congenital Abnormalities
Feeding Evaluation

- **Feeding Team**
  - Dietician
  - Speech language pathologist/ Occupational therapist
  - Practitioner
- **Routine lab:** CBC with Diff, Stool guiac, Chem Panel, Serum Zinc, Iron Studies, Chest X-ray
- **Modified Barium Swallow**
- **Indirect Calorimetry**

Treatment

- **Treatment**
  - Individualized based on feeding ability, lifestyle of the individual and medical condition
  - May be a combination of enteral and oral feeding
  - Age appropriate nutrients with specialized diets only as needed
  - Should include Speech Language Therapy/ Occupation Therapy to optimize oral feeding potential
  - Frequent reassessment

Treatment of Insufficient Caloric Intake

- **Treatment**
  - Regular diet with increased caloric concentration &/or modification in consistency
    - Increased fats, vegetable oil, olive oil
  - High caloric formulas and supplements
    - Duocal, Polycose, Scandishake
  - Feeding techniques
    - consideration of enteral feeding tubes
  - Medications
    - Periactin (Cyproheptadine)
    - H2Blockers, Proton Pump Inhibitors
    - Prokinetics

- **Goals:**
  - To improve the quality of life of the individual
  - To reduce risk of illness secondary to malnutrition and medical complications of underlying conditions
  - To improve nutritional status