HIRSCHSPRUNG'S DISEASE

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Long-Term Bowel Symptoms Following Corrective Surgery

- Obstructive (11-42%)
  - Constipation
- Overflow incontinence
- Abdominal Distention
- Vomiting
- Fecal Incontinence (2.5-80%)
- Enterocolitis (2%-39%)
  - May be associated with mortality (0-30%)

Once a Hirschsprung’s always a Hirschsprung’s

Outcomes

- N= 259, 29 Years
- Normal Bowel Function: 68.7%
- Enterocolitis: 16.6%
- Constipation: 21.7%
- Incontinence: 10.3%
- Subsequent Sphincterotomies: 17.7%

**Significant Impact**

- Yanchar, et al. 1999
  - Bowel function had significant effect on activities and social lives: 50% (5-15y)
  - 24-33% parents of children <15 years reported significant disruption of family life
- Significantly lower quality of life scores found in those with fecal soiling or incontinence (Bai, et al. 2002)
- Enterocolitis is potentially fatal

**Causes of Bowel Symptoms Following Corrective Surgery**

- **Obstruction**
  - Anatomic
    - Anal Stenosis (2.9-19%)
    - Stricture
  - Functional
    - Residual/Acquired aganglionosis (≤20%)
    - Neuroenteric Abnormalities (NID)
    - Internal Anal Sphincter Achalasia (≤75%)
    - Idiopathic
    - Fecal retention

- **Fecal Incontinence**
  - Abnormal anorectal function
  - Rapid Transit/Shortened Colon

- **Enterocolitis**
  - Bacterial (e.g. C. dif.), other
Tools to Help Evaluate Bowel Symptoms Following Corrective Surgery

- History/Physical Examination
- Barium Enema
- Rectal Biopsy
- Anorectal Manometry
- Colonic Manometry
- Other
- Colonoscopy
- Transit studies

Physical examination

- Abdomen
- Spine
- Rectal exam
  - Tone
  - Stricture
  - Anastomosis
    - Must know type of operation
Child with HD s/p CS

History and physical examination
C. difficile testing

Non-retentive Fecal Incontinence
Obstructive symptoms and/or Enterocolitis

Expand differential diagnosis
Begin fiber and anti-diarrheals

Anal Stenosis
Anal dilatation and surgical evaluation

Barium Enema
Stricture or Transition Zone
Surgical evaluation

Therapies

- Social/Behavioral
- Medications
- Decrease IAS pressure
- Surgical

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Therapies

- Social/Behavioral
- **Medications**
  - Manipulate transit/stool consistency
  - Antibiotics
- Decrease IAS pressure
- Surgical

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Therapies

- Social/Behavioral
- **Medications:**
  - *Manipulate transit*
    - Accelerate transit (laxatives/prokinetics)
    - Bypass anal obstruction (rectal irrigations; suppositories)
    - Slow transit (loperamide, lomotil)
  - *Antibiotics*
    - Flagyl, cipro, augmentin, lactobacillus
- Decrease IAS pressure
- Surgical
Therapies

- Social/Behavioral
- Medications
- Decrease IAS pressure
- Surgical

Background: Non-relaxing Internal Anal Sphincter

- Nonrelaxation leads to a functional obstruction and increased resistance
- Higher rectal pressures required for defecation
- Children prone to fecal retention

Therapies for Nonrelaxing IAS

- Therapies
  - Surgical
  - Medical
Current Therapies

- **Surgery**
  - Myectomy/Myotomy in Hirschsprung's Patients
    - 20/29 (68.9%) “Good” outcome
    - 9/29 (31%) “Poor” or “Fair” outcome
    - 6/29 (20.6%) Occasional soiling


- **BOTOX**
  - Potent bacterial neurotoxin acting on neuromuscular junction blocking release of acetylcholine from presynaptic cholinergic nerves
  - Weakens muscle in focal and transient fashion
  - Large experience
  - Medical myectomy

- **Current Therapies: Nonrelaxing IAS**
  - Botulinum Toxin
    - Hirschsprung’s Related (18 Children)
      - 4/18 (22%) without any improvement
      - 2/18 (11%) had improvement less than 1 month
      - 7/18 (39%) with improvement for 1 to 6 months
      - 5/18 (28%) with improvement for greater than 6 months
      - 10/18 required additional injections
      - 8/9 with improvement > 1 month had documented decreases in sphincter pressures
      - 3/5 without significant improvement had documented decreases in sphincter pressures

Problems with Botox

- General Anesthesia
- Repeated injections
- Cost
- Experience
- Doesn't Always Work
- Incontinence may Occur
New treatment options??

Therapies to address IASA: Glycerin Trinitrate

- Nitric oxide is the primary neurotransmitter responsible for smooth muscle relaxation
- Topical organic nitrate application to the IAS results in relaxation
- Experience with acute/chronic anal fissure
- Topical organic nitrates have been used in small case series to relieve obstructive symptoms in children with HD s/p corrective surgery (Tiryaki, T et al. 2005)

Anal Pressures after GTN
Baseline and Lowest Anal Pressures

NITRIC OXIDE
- 15 patients with obstructive symptoms
  - 0.1 mg/kg BID
  - 60% good clinical response

Therapies
- Social/Behavioral
- Medications
- Decrease IAS pressure
- Surgical
**Surgery**
- ACE
- Segmental resections
- Total colectomy
- Colostomy/ Ileostomy

Take in account high non-relaxing internal anal sphincter pressure

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**Managing Obstructive Symptoms and/or Enterocolitis**

OS
- Anal Stenosis → Dilatations
- Physical Examination
- Barium Enema
- Stricture → Surgical evaluation
- Aggressive Bowel Regimen → Response → NO → Yes

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**Managing Obstructive Symptoms and/or Enterocolitis**

- Flex Sigmoid
- Rectal Biopsy
- Anorectal Manometry
- BoTox
- Aganglionosis Anatomic → Surgical Revision
- No improvement → Rectal therapies Repeat Botox
- Improvement → BoTox prn/ Myectomy
Managing Obstructive Symptoms and/or Enterocolitis

- Caloric manometry
- Segmental resection
- Diversion/Revision

Boston Children’s Hospital

<table>
<thead>
<tr>
<th>Symptoms</th>
<th>Obstructive Enterocolitis</th>
<th>Non-retentive fecal incontinence</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>3340 (55%)</td>
<td>740 (11.7%)</td>
</tr>
<tr>
<td></td>
<td>1603 (25.3%)</td>
<td>602 (10%)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Sex</th>
<th>Male</th>
<th>Female</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>48</td>
<td>11</td>
</tr>
<tr>
<td></td>
<td>(81.7%)</td>
<td>(18.3%)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Initial aganglionosis</th>
<th>Short-segment</th>
<th>Long-segment</th>
<th>Total colonic</th>
</tr>
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<tbody>
<tr>
<td></td>
<td>4200 (72%)</td>
<td>540 (14%)</td>
<td>10 (16%)</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Surgery</th>
<th>Soave</th>
<th>Duhamel</th>
<th>Swenson</th>
<th>Unknown</th>
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<tbody>
<tr>
<td></td>
<td>3553 (63.3%)</td>
<td>1556 (28%)</td>
<td>760 (11.6%)</td>
<td>260 (5%)</td>
</tr>
</tbody>
</table>

| Age                        | 5.1 ± 0.6 years | Down’s Syndrome | 1501 (15%) | Follow-up |
|----------------------------|-----------------|-----------------|-------------|
|                            | 36.4 ± 4.4 months |

(JPGN 2011; 53: 75–79)
WORK UP

<table>
<thead>
<tr>
<th>Condition</th>
<th>Count</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Residual aganglionosis</td>
<td>7</td>
<td>12%</td>
</tr>
<tr>
<td>Colonic/anastomotic stricture</td>
<td>8</td>
<td>14%</td>
</tr>
<tr>
<td>Non-intractable constipation</td>
<td>10</td>
<td>17%</td>
</tr>
<tr>
<td>Non-relaxing IAS as main reason for symptoms</td>
<td>22</td>
<td>38%</td>
</tr>
<tr>
<td>Colonic dysmotility abnormal (6)</td>
<td>4</td>
<td>6.8%</td>
</tr>
<tr>
<td>Rapid transit</td>
<td>2</td>
<td>3.4%</td>
</tr>
<tr>
<td>Bacterial overgrowth</td>
<td>2</td>
<td>3.4%</td>
</tr>
<tr>
<td>Food allergy</td>
<td>2</td>
<td>3.4%</td>
</tr>
</tbody>
</table>

(Treatment JPN 2011; 53: 75–79)

Treatment

<table>
<thead>
<tr>
<th>Treatment</th>
<th>Count</th>
<th>Percentage</th>
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</thead>
<tbody>
<tr>
<td>Aggressive bowel regimen only</td>
<td>5</td>
<td>8%</td>
</tr>
<tr>
<td>Clostridium botulinum toxin</td>
<td>38</td>
<td>63%</td>
</tr>
<tr>
<td>Repeat pull-through</td>
<td>9</td>
<td>15%</td>
</tr>
<tr>
<td>Myectomy</td>
<td>5</td>
<td>8%</td>
</tr>
<tr>
<td>Anal dilatation</td>
<td>4</td>
<td>6%</td>
</tr>
<tr>
<td>Cecostomy tube placement</td>
<td>2</td>
<td>3%</td>
</tr>
<tr>
<td>Stricuroplasty and resection</td>
<td>2</td>
<td>3%</td>
</tr>
<tr>
<td>Diversion (ileostomy/colostomy)</td>
<td>2</td>
<td>3%</td>
</tr>
</tbody>
</table>

(Treatment JPN 2011; 53: 75–79)

TREATMENT

JPGN 2011; 53: 75–79
CONCLUSIONS
- HD patients continue to have symptoms after surgical correction
- Once a Hirschsprung’s, always a Hirschsprung’s
- Work up needs to be methodical
- There are new medical therapies available for obstructive symptoms