# HIRSCHSPRUNG'S DISEASE

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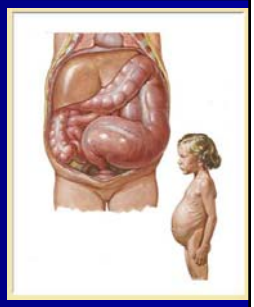

## Long-Term Bowel Symptoms Following Corrective Surgery

- Obstructive (11-42%)
  - Constipation

Once a Hirschsprung's always a Hirschsprung's

- Fecal incontinence (2.5-86%)
- Enterocolitis (2%-39%)
  - May be associated with mortality (0-30%)

## Outcomes

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

Menezes, M, et al. Long-term results of bowel function after treatment for Hirschsprung's disease: a 29 year review. *Pediatr Surg Int* 2006

## 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

## Causes of Bowel Symptoms Following Corrective Surgery

- 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

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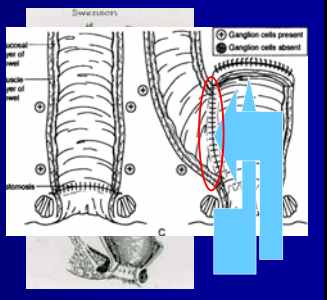
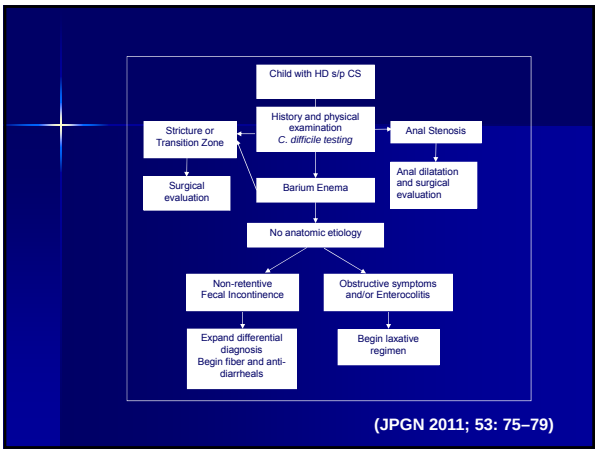
- Other
  - Colonoscopy
  - Transit studies

## Physical examination

- Abdomen
- Spine
- Rectal exam
  - Tone
  - Stricture
  - Anastomosis
    - Must know type of operation






(JPGN 2011; 53: 75–79)

## Therapies

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

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  - Manipulate transit/stool consistency
  - Antibiotics
- Decrease IAS pressure
- Surgical

## 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

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- Social/Behavioral
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## 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

**Wildhaber, BE et al. "Posterior Myotomy/Myectomy for Persistent Stooling Problems in Hirschsprung's Disease" Journal of Pediatric Surgery 39: 920-926 2004**

## 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

**Minkes, RK et al. "A Prospective Study of Botulinum Toxin for Internal Anal Sphincter Hypertonicity in Children with Hirschsprung's Disease" Journal of Pediatric Surgery 2000 35: 1733-1736.**

## BOTOX


Am J Gastro 2009;104:976

## BOTOX

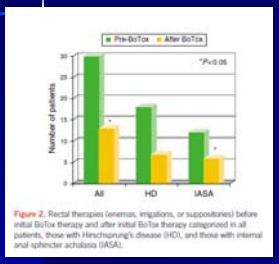



Figure 2. Rectal therapies (enemas, irrigations, or suppositories) before initial BoTox therapy and after initial BoTox therapy categorized in all patients, those with Hirschsprung's disease (HD), and those with internal anal sphincter achalasia (IASA).

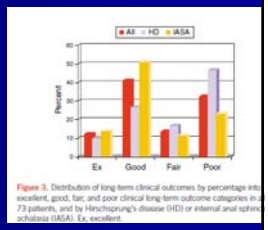



Figure 3. Distribution of long-term clinical outcomes by percentage into excellent, good, fair, and poor clinical long-term outcome categories in all 73 patients, and by Hirschsprung's disease (HD) or internal anal sphincter achalasia (IASA). Ex, excellent.

Am J Gastro 2009;104:976

## Problems with Botox

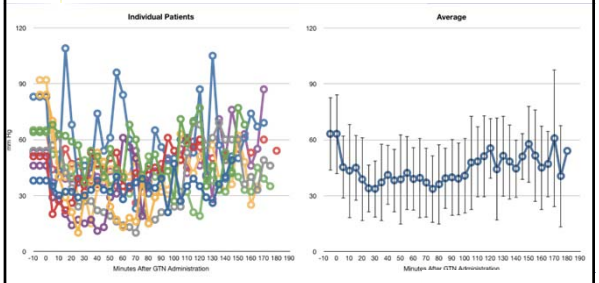
- General Anesthesia
- Repeated injections
- Cost
- Experience
- Doesn't Always Work
- Incontinence may Occur

## New treatment options??

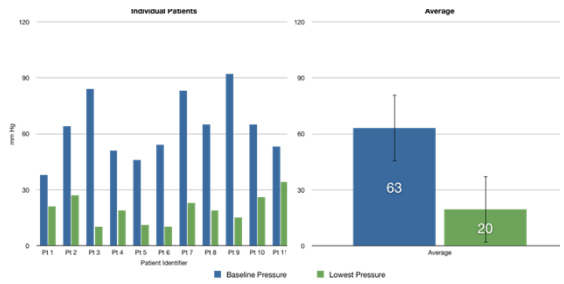

## Therapies to address IASA: Glycerin Trinitrate

- 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

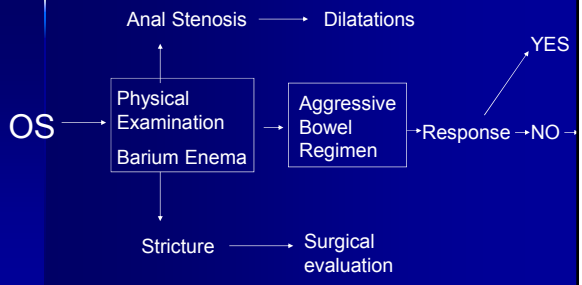
- Social/Behavioral
- Medications
- Decrease IAS pressure
- Surgical

## Surgery

- ACE
- Segmental resections
- Total colectomy
- Colostomy/ Ileostomy

Take in account high non-relaxing internal anal sphincter pressure

## Managing Obstructive Symptoms and/or Enterocolitis

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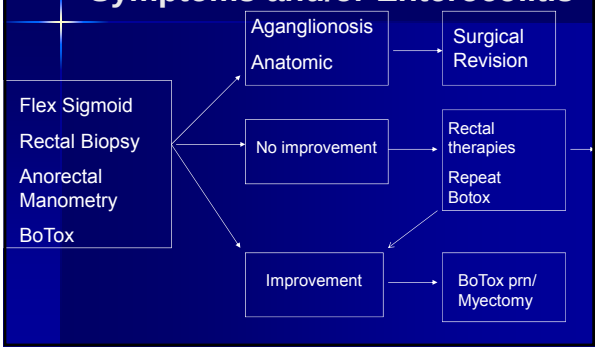

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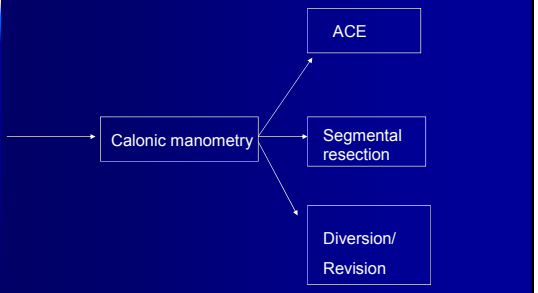


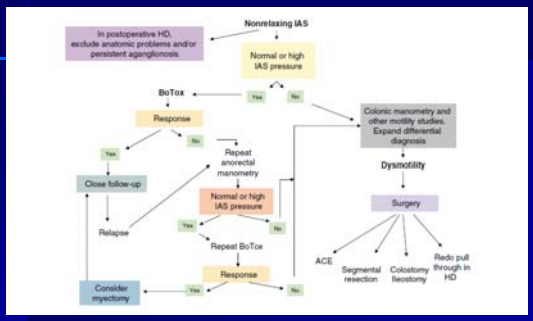


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## Boston Children's Hospital

| | | |
|---|---|---|
| Symptoms | Obstructive<br>Enterocolitis<br>Both<br>Non-retentive fecal incontinence | 33/60 (55%)<br>7/60 (11.7%)<br>14/60 (23.3%)<br>6/60 (10%) |
| Sex | Male<br>Female | 49 (81.7%)<br>11 (18.3%) |
| Initial aganglionosis | Short-segment<br>Long-segment<br>Total colonic | 42/60 (72%)<br>8/60 (14%)<br>10 (16%) |
| Surgery | Soave<br>Duhamel<br>Swenson<br>Unknown | 38/60 (63.3%)<br>12/60 (20%)<br>7/60 (11.6%)<br>3/60 (5%) |
| Age | | 5.1 ± 0.6 years |
| Down's Syndrome | | 7/60 (12%) |
| Follow-up | | 39.4 ± 4.4 months |

(JPGN 2011; 53: 75–79)

## WORK UP

| | |
|---|---|
| Residual aganglionosis | 7 (12%) |
| Colonic/anastomotic stricture | 8 (14%) |
| Non-intractable constipation | 10 (17%) |
| Non-relaxing IAS as main reason for symptoms | 22 (38%) |
| Colonic dysmotility (abnormal) (6) | 4 (6.8%) |
| Rapid transit | 2 (3.4%) |
| Bacterial overgrowth | 2 (3.4%) |
| Food allergy | 2 (3.4%) |

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## Treatment

| | |
|---|---|
| Aggressive bowel regimen only | 5 (8%) |
| *Clostridium botulinum* toxin | 38 (63%) |
| Repeat pull-through | 9 (15%) |
| Myectomy | 5 (8%) |
| Anal dilatation | 4 (6%) |
| Cecostomy tube placement | 2 (3%) |
| Stricturoplasty and ressection | 2 (3%) |
| Diversion (ileostomy/colostomy) | 2 (3%) |

## TREATMENT

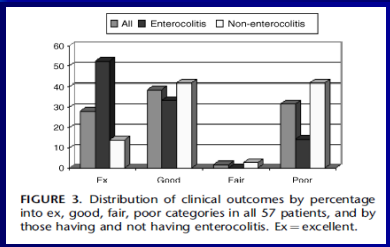


FIGURE 3. Distribution of clinical outcomes by percentage into ex, good, fair, poor categories in all 57 patients, and by those having and not having enterocolitis. Ex = excellent.

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## 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 symtpoms