Hirschsprung Round Table

Hirschsprung Anatomy and Physiology
Jenny Kreiss, MN, PNPPC-BC

Objectives
• Participants will review embryology of ganglion cell neuron migration
• Participants will describe mechanics of intestinal peristalsis and motility
• Participants will describe pathologic evidence of abnormal ganglion cells as basis for diagnosis of Hirschsprung Disease and disorders of abnormal motility
• Participants will describe the history of Hirschsprung Disease over time
Hirschsprung Disease

- Hirschsprung: a congenital malformation defined as the absence of ganglion cells in the myenteric and submucosal plexuses of the terminal rectum +/- more proximal bowel.
- Functionally marked by intestinal obstruction caused by the inability of the gut to transmit a peristaltic wave along the aganglionic segment.
- Population incidence 1:5000 live births.
- Male to female ratio 4:1, particularly prevalent in short-segment.
- Embryonic vagal neural crest origin of enteric neural system, colonized in oral-to-anal migration. The entire length of intestine is colonized by week 7 in humans.
- Causal theories: multiple genes, hostile environment.

History

- Harold Hirschsprung presented his classic description of "congenital dilatation of the colon" to the pediatric congress in Berlin, 1886.
- Previous descriptions of children with megacolon date back to the 17th century.

Radiologic evidence

Peristalsis is an interaction of the muscles, nerve cells and tendinous connective tissue of the muscularis propria. Failure of any of these components results in motility disorder. **Aganglionosis results in inability of the smooth muscle to relax, creating a functional bowel obstruction.** Contrast enema demonstrates transition zone.
Pathologic evidence

An adequate suction rectal biopsy should be ≥3 mm in greatest dimension, and at least one-third of the biopsy should be submucosa. Hypertrophic nerves (≥40 μm) are typically present in the aganglionic submucosa in HD. A biopsy with ganglion cells contains sparse thin AChE-positive nerves in the muscularis mucosae and lamina propria. In contrast, coarse large AChE-positive nerves are present in a biopsy from a HD patient. Calretinin immunoreactivity exists in small nerves in the muscularis mucosae and lamina propria, when ganglion cells are present. No calretinin immunoreactivity is present in the mucosa or superficial submucosa of aganglionic bowel.

Adapted from Kapur, R.  Practical Pathology and Genetics of Hirschsprung’s Disease. Seminars in Pediatric Surgery, Volume 18, Number 4, November 2009.

Fun Fact: Nomenclature du jour

Current APSA convention: named procedures and diseases have no apostrophe’s:

- Hirshsprung (not Hirschsprung’s)
- Crohn (not Crohn’s)
- Ladd (not Ladd’s)
- Stamm
- Malone
- Duhamel

Preoperative Care

Monica Holder, BSN, RN, CPN
Patty Kern, BSN, RN CPN, CCRC
Newborn Suspicion of HD

- Newborn with delayed meconium elimination
  - Contrast Enema
  - Rectal Biopsy
  - Irrigation teaching for enterocolitis

Enterocolitis

Signs and Symptoms

- Fever
- Abdominal Distention
- Not Stooling
- Vomiting
- Explosive Diarrhea
- Foul Smelling Stool/Gas

Prevention and Treatment

- Irrigations!
  - Irrigations!
  - Irrigations!
- Flagyl

http://seraph.cchmc.org/MediasiteEx/Play/5451515dab9a6b4e30b8f8b74d9b4d9c1
Intraoperative Care
Sarah-Ross Tolin, MSN, RN, CNL, CPN
Sarah Wilhelm, BSN, RN

Surgical Interventions
- Indications for Surgery
  - Diagnosis
  - Infection
  - Treatment
- Surgical Interventions
  - Rectal biopsy
  - Diverting ostomies
  - Transanal repair

Positioning & Total Body Prep
- Surgical Positioning
- Total Body Preparation
### Biopsy & Specimen Management

- Fresh Specimen vs. Frozen Specimen
  - When
  - Where
  - Why

### Post Operative Care

Kimberly Cogley, MSN/MBA

### Skin Care

- Care of the perineum
  - Skin not used to stool
  - Barrier cream
  - Clean and dry
- Care of the ostomy
  - WOCN consult
  - Home supplies
  - Need for distal segment wash outs
- Incisions (dependent of surgical approach)
  - Remove dressing
  - Bathe once dressings removed
Nutrition

- Feeding
  - Return of bowel function
  - Clear liquids
  - Formula/Breastmilk

- Parental nutrition requirement
  - Current nutritional status
  - Slow return of bowel function
  - Dilation of colon – what extent?

Ready for Discharge

- Maintain skin
- Care of colostomy
- Nutritional needs met
- Understanding of potential postoperative complications
- Contact information for department

Most important thing is to give lots of love and care
Objectives

- Participants will be able to describe the needs of the patient with Hirschsprung’s disease (HD) that are addressed when they are seen in clinic postoperatively.

- Participants will understand that HD is not “cured” after the operation and that children with HD will need ongoing follow-up and may encounter several problems which may include: constipation, loose stools, incontinence, enterocolitis, bacterial overgrowth and stricture.

Immediate Postop Management

- Diaper dermatitis (most common)
- Dilations
- Enterocolitis (HAEC)/Pouchitis/C. difficile infections
- Irrigations
- Stricture
- Ostomy
- Growth & Development
- Diet

Long Term Follow Up

- Growth and development
- Bowel management
- School issues (504 plan/IEP)
- Coordination of care
- Education
Past, Present, & Future

**Past**

Mortality rates: higher (2.4% - 15.6%)

Why do you think that mortality rates were so high over this 40 year period?

**Present**

Mortality rate: 1.1%

Laparoscopic pull-through

Increased understanding that there are long term issues with HD patients postoperatively (fistulae, bowel atresia, Hirschsprung’s, stenosis,

- Bowel Management programs
- Hirschsprung’s consortium
- Medical management (e.g. Flagyl/Bactrim)
- Understanding of bacterial overgrowth

**Future**

Mortality rates: decreasing

Hirschsprung’s associated enterocolitis

- Early recognition and treatment
- Abdominal distention and vomiting are major presenting clinical features
- More advances in medical field, improved immune functions

HAEC: Hirschsprung’s associated enterocolitis

Microbiome, altered mucin levels, impaired immune functions

Discussion

What is your dilation protocol?

What modalities are you using for bowel management?

Do you notice any dietary issues in HD patients?

Do you teach families irrigations? If so, how are the irrigations performed in your institution?

What do you use for diaper rash prevention/treatment?

How do you identify HAEC in your institution?

Do you have a protocol for treatment?

References


