Roundtable Presentation
Hirschsprung’s Disease

Disclosure Information

There were no financial interests or relationships or conflicts of interest to disclose for any of the Hirschsprung’s Disease roundtable moderators.

Hirschsprung’s Disease:
Anatomy and Physiology
Jenny Kreiss, RN, MN, PNP-GC
Seattle Children’s Hospital
Seattle, WA
Objectives:  
Anatomy and Physiology

- 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’s Disease and disorders of abnormal motility

Hirschsprung’s Disease

- Hirschsprung’s: 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

Making the diagnosis: 
Anatomy and physiology

Contrast enema demonstrates transition zone 
Absence of ganglion cells in myenteric plexus
Mechanics of intestinal motility

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.

Hirschsprung’s Disease: Preoperative care

Monica Holder, BSN, RN, CPN
Cincinnati Children’s Hospital
Cincinnati, OH

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

Enterocolitis

Prevention and Treatment
• Irrigations!
• Irrigations!
• Irrigations!
• Flagyl

Hirschsprung’s Disease:
Intraoperative Care
Steadman McPeters, DNP, CPNP-AC, RNFA
University of Alabama at Birmingham, School of Nursing
Birmingham, AL
Objectives: Intraoperative Care

- The attendee will have been given the opportunity to understand a brief description of the operation
- The attendee will have been given the opportunity to understand the care provided intraoperatively to the HD patient
- The attendee will have been given the opportunity to understand how to receive and give handoff to other members of the health care team

Description of the Operation

- Definitive Tx of HD – surgical resection of aganglionic bowel & anastomosis of the ganglionic bowel to the distal rectum, directly above the pectinate line
- Traditional approach: 2-or 3-stage repair in which a colostomy is primarily preformed for decompression of the colon
- Common surgical approach: Primary repair with 1-stage technique
- Experienced clinical pathologist
- Earlier diagnosis
- Swenson Procedure
- Duhamel Procedure
- Soave Endorectal Pull-Through
- Advantages to endoscopic/laparoscopic surgery

Intraoperative Care

- Pediatric Nurse Practitioner Role (CPNP)
- Registered Nurse First Assistant Role (RNFA)
- Preparing for the case
- Interaction with the patient & family
- When to communicate with the family
Interdisciplinary Team

- Receiving hand-off pre-surgery
- Giving hand-off post-surgery
- OR RN
- PACU RN
- NICU MD/CRNP/RN

Hirschsprung’s Disease: Inpatient Postoperative Management
Jennifer Quilty, DNP, ARNP
Children’s Surgical Associates
Orlando, FL

Inpatient postoperative management

- Primary pull-through or Ostomy creation initially have the same postoperative management in the hospital
  - IVF and antibiotics
  - NPO with OG in place until return of bowel function per rectum or via ostomy
    - Slowly advance feeds when ready (usually 24-48 hours)
  - Prep diaper area for stool and impending diaper rash if pull-through
  - Keep dressing dry and intact
Inpatient postoperative management

• Teachings- planning for discharge: Ostomy
  • Parent teaching with WCON and set up for outpatient appointment
  • Supplies for discharge
  • Monitor incision site
  • Parent teaching for signs of enterocolitis
  • Follow up with surgery team appointment

Inpatient postoperative management

• Teaching-planning for discharge- Soave
  • Diaper area care teaching with supplies
  • Teaching for signs of enterocolitis
  • Monitor incision site (if pt has an incision)
  • Follow up appointment with surgery after discharge

Hirschsprung’s Disease:
Postoperative outpatient clinic & long term care
Teri Coha, RN, MSN, APN, CWOCN
Ann & Robert H. Lurie Children’s Hospital of Chicago
Objectives: Outpatient and Long Term Care

- Participants will understand the initial postoperative needs of the patient: ostomy care or prevention/care of contact dermatitis.
- Participants will understand that HD is not “cured” after the operation & that children with HD will need ongoing follow-up.

Outpatient postoperative care

- Irritant contact dermatitis
  - Cleansers
  - Skin prep
  - Barrier pastes
- Ostomy care
  - Ostomy company programs and education materials
  - Clothing companies
  - Odor eliminator products

Outpatient postoperative care

Ongoing Care

- Hirschsprung’s associated enterocolitis (HAEC)
  - can occur before or after pull through
  - decreases to ~5% by age 5-6 or 2-3 years after pull though if child is diagnosed later in life
- Other
  - C-diff
  - Constipation or multiple loose stools
  - Stricture
  - Failure to thrive
  - Pelvic nerve injury
### Outpatient postoperative care

**Bowel management**

- should not toilet train prior to 3-4 years of age
- address continence prior to entering school to avoid the negative impact of fecal incontinence at school.
- children will avoid social activities, sports, gym
- children labeled as “holders” or “defiant”
- diagnosis of ADD or ADHD

Goal: social continence using enemas, laxatives, stool softeners, pectin, imodium, lomotil

APN run bowel management clinics/visits/phone calls