Comprehensive Colorectal Surgical Care in the Pediatric Population

Wendy Lewis, APRN II, FNP-C
& Christine Warner, APRN II, CPNP-PC
Comprehensive Colorectal Surgical Care in the Pediatric Population

Requirements for Successful Completion (to receive the 1.0 contact hour(s) for this activity):
The participant will:
• Sign-in on electronic roster
• Complete an electronic evaluation

Provider Approval Statement:

Children’s Mercy Kansas is approved with distinction as a provider of nursing continuing professional development by the Midwest Multistate Division, an accredited approver by the American Nurses Credentialing Center’s Commission on Accreditation.

Conflict of Interest:
No conflicts of interest have been identified for the planners or presenters of this activity.
Objectives

• Become familiar with the Comprehensive Colorectal Center
• Understand the difference between Anorectal Malformation, Hirschsprung Disease, Cloaca, and Cloacal Exstrophy
• Differentiate between initial management/images associated with each of the diagnoses
• Recognize the different types of bowel management courses the colorectal team utilizes in these patient populations
What is the Colorectal Center?

Children’s Mercy is home to one of the eight Comprehensive Colorectal Centers in the nation.

We take a team-based approach to care for children with colorectal complex colorectal and pelvic conditions.
Pediatric Colorectal Care
Comprehensive Colorectal Center: Multidisciplinary Team

Colorectal Surgery
• Dr. Rebecca Rentea
• Wendy Lewis, APRN
• Christine Warner, APRN

Urology
• Dr. Alonso Carrasco
• Dr. John Gatti
• Mary Langston, APRN

Psychology
• Dr. Christina Low-Kapalu

Gynecology
• Dr. Anne-Marie Priebe
• Dr. Julie Strickland
• Dr. Ashli Lawson

Gastroenterology
• Dr. John Rosen
• Dr. Gracielle Bahia

Colorectal Nursing Staff
• Mariama Faty, RN
• Maddie Jermain, RN

Program Coordinator
• Margaret Martin-McLain, RN
Diagnoses managed by the Colorectal Center

- Anorectal Malformation
- Cloaca and Cloacal Exstrophy
- Hirschsprung Disease
- Functional Constipation
Why is a center important?

• Our center performs 400-500 surgical cases per year

• Of those 200-250 are complex

• In comparison the average pediatric surgeon performs 1-2 of these types of complex cases per year

• Most pediatric surgeons will never operate on a cloaca in their career
Anorectal Malformation

Described based upon:
- Size
- Location
- Type of Fistula
Anorectal Malformation Screening

VACTERL Work-up
V - Vertebral Anomalies
A - Anorectal Malformation
C - Cardiac Anomalies
T/E - Tracheoesophageal fistula/ Esophageal Atresia
R - Renal Anomalies
L - Limbs Anomalies

• Other areas to assess:
  • Sacrum
  • Gynecological
Sacrum & Spine

Strongly associated with bladder and bowel continence
Posterior Sagittal Anorectoplasty

- cut into the correct muscle complex
Case Study:

- 5-year-old female
- ARM- rectovestibular with tethered cord
- Family motivated to attempt laxative treatment
- Laxative treatment resulted in daily accidents and streaking despite high dose- 75 mg Senna and fiber daily
- Ultimately transitioned to high volume enemas (300mL NS+9mL Castille) with success
- At an older age may consider an appendicostomy or another trial of laxatives
Cloaca

A type of anorectal malformation that occurs when the rectum, vagina, and urinary tract form a single channel rather than 3 openings.
Cloacal Exstrophy/OEIS
Case Study

• 2 year old female
• History of cloacal exstrophy
• Has approximately 12 inches of colon, stooling into ostomy
• Voiding out of a vesicostomy
• Currently: working towards reconstruction to occur around age 4, completing nightly enemas into the ostomy with no stool in the bag all day as a result of this daily enema
Hirschsprung Disease

- Aganglionic portion of colon
- Various presentations

*Not always diagnosed at birth*
Hirschsprung Presentation

• Newborn:
  • Failure to pass meconium within first 48 hours of birth
  • Abdominal distension
  • Enterocolitis
  • Failure to thrive (<5%)
  • Explosive, foul smelling stools

• Older children:
  • Unmanageable chronic constipation
  • Delayed growth
Hirschsprung Diagnosis
Hirschsprung Management

- Rectal Irrigations during infancy
- Some patients are diverted in NICU/infancy
- Pull-through procedure at 8-12 weeks of life
  - Total colonic children are pulled through at 6-12 months
- Anal sphincter Botox injection
- Lifelong bowel management
Hirschsprung Associated Enterocolitis

- Enterocolitis- inflammation of the lining of the mucosa
- May occur before and after surgery- Life threatening!
- High occurrence in patients with history of HD
- Causes: stasis of stool, bacterial or viral infection
- Symptoms:
  - Abdominal pain, Fever, Diarrhea, Vomiting
Rectal Irrigations
MEDICAL ALERT

This patient has Hirschsprung Disease and is at risk of Hirschsprung’s Enterocolitis

Symptoms include: lethargy, fever, vomiting, diarrhea, and abdominal distention

Immediate treatment: Stop oral feeds. Give IV antibiotics (Cipro/Flagyl), IV fluids, NG tube, rectal irrigation (washout). Do not give enema.

Date:

This child is a patient of Dr. Rentea and Children’s Mercy. Please contact the on-call Pediatric surgeon at 1 (800) GO-MERCY immediately upon arrival to your facility.
Functional Constipation

Referred to Colorectal Surgery Specialty after failed management of constipation through other specialties.

Diagnoses

• Motility disorders
• Neurogenic Bowel
• Sacral deformities, sacral agenesis, caudal regression
Bowel Management

Goals:
• Be clean and continent for stool
• Have a predictable stooling pattern

Strategies:
• Utilize a variety of treatment options
• Treatment variations may change throughout child’s lifetime
Bowel Management Considerations

Oral Laxatives vs. Enemas/Flushes

- Anorectal Malformation
  - Fistula/defect level
  - Spine and sacrum status
- Cloaca
  - Channel Length
  - Spine and sacrum status
- Hirschsprung Disease
  - Transition zone
- Constipation
  - Rectal Prolapse
  - Motility Disorders
## Continence Potential

### ARM TYPE

<table>
<thead>
<tr>
<th>Condition</th>
<th>Points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Perineal Fistula</td>
<td>1</td>
</tr>
<tr>
<td>Rectal Stenosis</td>
<td>1</td>
</tr>
<tr>
<td>Rectal Atresia</td>
<td>1</td>
</tr>
<tr>
<td>Rectovestibular Fistula</td>
<td>1</td>
</tr>
<tr>
<td>Rectobulbar Fistula</td>
<td>1</td>
</tr>
<tr>
<td>Imperforate Anus without Fistula</td>
<td>1</td>
</tr>
<tr>
<td>Cloaca &lt; 3 cm Common Channel</td>
<td>2</td>
</tr>
<tr>
<td>Rectoprostatic Fistula</td>
<td>2</td>
</tr>
<tr>
<td>Rectovaginal Fistula</td>
<td>2</td>
</tr>
<tr>
<td>Rectobladderneck Fistula</td>
<td>3</td>
</tr>
<tr>
<td>Cloaca &gt; 3 cm Common Channel</td>
<td>3</td>
</tr>
<tr>
<td>Cloacal exstrophy</td>
<td>3</td>
</tr>
</tbody>
</table>

### SPINE

<table>
<thead>
<tr>
<th>Condition</th>
<th>Points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal termination of the Conus (L1-L2)</td>
<td>1</td>
</tr>
<tr>
<td>Normal filum appearance</td>
<td>1</td>
</tr>
<tr>
<td>Abnormally low termination of the Conus (below L3)</td>
<td>2</td>
</tr>
<tr>
<td>Abnormal fatty thickening of filum</td>
<td>2</td>
</tr>
<tr>
<td>Myelomeningocele</td>
<td>3</td>
</tr>
</tbody>
</table>

### SACRUM

<table>
<thead>
<tr>
<th>Condition</th>
<th>Points</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sacral Ratio = Greater than 0.7</td>
<td>1</td>
</tr>
<tr>
<td>Sacral Ratio = Between 0.4 and 0.69</td>
<td>2</td>
</tr>
<tr>
<td>Hemisacrum</td>
<td>2</td>
</tr>
<tr>
<td>Sacral Hemivertebrae</td>
<td>2</td>
</tr>
<tr>
<td>Presacral Mass</td>
<td>2</td>
</tr>
<tr>
<td>Sacral Ratio = Less than 0.4</td>
<td>3</td>
</tr>
</tbody>
</table>

### TOTAL POINTS

- **3-4 = Good** Potential for Continence
- **5-6 = Fair** Potential for Continence
- **7-9 = Poor** Potential for Continence

---

LOVE WILL.
Laxative Management

• **Senna**
  • 1-2mg/kg
  • Varied bowel movement result time
  • "Pusher"

• **MiraLAX**
  • Not used as frequently
  • "Musher"
High Volume Retention Rectal Enemas

- Large volume enemas
  - 200-500mL saline or water
- Additives to stimulate colon
  - Bisacodyl
  - Glycerin
  - Castile
- Evacuate stool from descending/rectosigmoid colon
- Results in a complete bowel movement within 45-60 minutes
- Clean for stool until next enema
- Predictable timing
Appendicostomy (MACE) and Cecostomy

- Allows patient to perform enemas via antegrade route
One Centers Experience

- The first year of the mini-ace over 20% of placements had troubleshooting complications
- Implemented a teaching process and post operative process
- 9 months after implementation saw a 5% decrease in postoperative call back
MACE

*Golden Ticket*

- Less drainage
- Less infection risk
- Less risk for stenosis
- No long term need for tube
- May “split” Appendix to use for MACE/Mitrofanoff
Case Study

- 17 year old female
- Functional Constipation/IBD
- Lifelong medical management for chronic constipation
- Desired more predictable stooling pattern and independence
- MACE created
- Now flushes with 500mL NS+30mL Glycerin with daily bowel movements
Cecostomy

- Tube placed into cecum
- Chait tube or Mini ace tube
- 1st change done in IR
- Main complaints:
  - leaking from cecostomy
  - granulation tissue
Bowel Management Boot Camp

• One-week program
• Nurse practitioners, surgeon, gastroenterologist, psychologist
• Two visits, beginning and end
• Daily abdominal x-rays
• Daily communication
Case Study

- 17 year-old female
- Cloaca
- Had been formally managed on enemas and had an appendicostomy placed
- Goal: wanted to take medication and not do a flush
- Transitioned to oral laxatives successfully during boot camp
- Able to successfully complete in cheerleading and wear underwear
Questions?

Thank you!

Contact us at: 
welewis@cmh.edu 
cnwarner@cmh.edu
References


• Wood RJ, Levitt MA. Anorectal Malformations. Clinics in colon and rectal surgery 2018;31(2):61-70