# **Comprehensive Colorectal Surgical Care in the Pediatric Population**

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#### **Comprehensive Colorectal Surgical Care in the Pediatric Population**

<u>Requirements for Successful Completion</u> (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



COMPREHENSIVE COLORECTAL CENTER





#### **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.

## Uterus Bladder Urethra Rectum Vagina Common channel

CLOACA

COMPREHENSIVE COLORECTAL CENTER





# **Cloacal Exstrophy/OEIS**



COMPREHENSIVE COLORECTAL CENTER





# **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:

LOVE WILL.

- 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
- <u>Symptoms</u>:
  - 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**

Sacral Ratio = Less than 0.4

			POINTS	
ARM TYPE	Perineal Fistula	1		
	Rectal Stenosis	1		
	Rectal Atresia	1		
	Rectovestibular Fistula	1		
	Rectobulbar Fistula	1		POIN
	Imperforate Anus without Fistula	1		
	Cloaca < 3 cm Common Channel	2		
	Rectoprostatic Fistula	2		
	Rectovaginal Fistula	2		
	Rectobladderneck Fistula	3		3-4 = 600
	Cloaca > 3 cm Common Channel	3		Potential
	Cloacal exstrophy	3		Continen
				2
SPINE	Normal termination of the Conus (L1-L2)	1		5-6 = Fair Potential Continen
	Normal filum appearance	1		
	Abnormally low termination of the Conus (below L3)	2	r	
	Abnormal fatty thickening of filum	2		
	Myelomeningocele	3		
			/	Potential
SACRUM	Sacral Ratio = Greater than 0.7	1		Continen
	Sacral Ratio = Between 0.4 and 0.69	2	/	continen
	Hemisacrum	2	V	
	Sacral Hemivertebrae	2	1	
	Presacral Mass	2		
-				

3





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# Laxative Management

#### Senna

- 1-2mg/kg
- Varied bowel movement result time
- "Pusher"
- MiraLAX
  - Not used as frequently
  - "Musher"



Updated 2/9/17





#### **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

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# Appendicostomy (MACE) and Cecostomy

• Allows patient to perform enemas via antegrade route

Appendicostomy Colon rotated Appendix attached to belly button







## **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

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#### MACE

#### 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







## Childrensmercy.org/colorectal

#### **Colorectal Center**

Nationally ranked by U.S. News & World Report

Refer a Patient

🕗 Meet the Team

Email

Contact Us (816) 234-3151





## **Questions?**

#### Thank you!

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







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