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7-2019

### Hirschsprung-Associated Enterocolitis

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#### Recommended Citation

Shann, Erin, "Hirschsprung-Associated Enterocolitis" (2019). *Nursing Student Class Projects (Formerly MSN)*. 364.

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# Hirschsprung-Associated Enterocolitis

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## Introduction

Hirschsprung disease (HD) occurs when there is an abnormal development in the enteric nervous system leading to ganglion cells being absent in the colon (Avansino & Levitt, 2017). One in 5,000 live births are affected by HD and is often associated with other factors such as trisomy 21 and a genetic mutation specifically in the Ret proto-oncogene (Avansino & Levitt, 2017). HD presents as severe abdominal distention, failure to pass a meconium stool within the first 48 hours of life, difficulty feeding, bilious vomiting in the newborn period, and as prolonged severe constipation in the older child (Avansino & Levitt, 2017).

Undiagnosed HD can lead to a life-threatening condition called enterocolitis. Enterocolitis presentation varies per individual with the most common symptoms including abdominal distention, foul-smelling explosive stools, loud rumbling noises coming from the abdomen, and fever (Langer et al., 2017). Hirschsprung-associated enterocolitis (HAEC) is challenging to diagnose due to non-specific symptoms and is underreported (Gosain et al., 2017). Proper preventative measures and treatment for those presenting with symptoms of enterocolitis are needed to decrease the risk of developing severe HAEC.

## Presentation of Case

A 2-year-old male with history of HD status-post primary pull-through. He has a history of one episode of enterocolitis pre-pull-through and five episodes post-pull-through. He has received two rectal Botox injections on two separate occasions, multiple rounds of antibiotics, and as needed rectal irrigations with 200 milliliters (mL) of 0.9% sodium chloride with a 16 French (Fr) red rubber catheter. He continues to experience sporadic fevers, lethargy, abdominal distention, and loud rumbling noises from his abdomen.

## Incidence & Presentation of HD

- 1 in 5,000 live births.
- Often associated with Trisomy 21 and the Ret proto-oncogene genetic mutation.
- Severe abdominal distention.
- Failure to pass meconium stool within 48 hours of life.
- Difficulty feeding.
- Bilious vomiting in the newborn period.
- Prolonged severe constipation in the older child.

(Avansino & Levitt, 2017).

## Signs & Symptoms of HAEC

- Undiagnosed HD can lead to a life-threatening condition called enterocolitis.
- Presentation varies per individual.
- Abdominal distention.
- Foul-smelling explosive stools.
- Loud rumbling noises coming from abdomen.
- Fever.

(Langer et al., 2017).

## Pathophysiology

- First line of defense in a healthy intestinal tract from adhesion and infections is a mucosal barrier (Nakamura et al., 2018).
- Gel-forming mucins, Goblet cells, which is the major component of mucus, block the direct attachment of bacteria to the epithelial layer (Nakamura et al., 2018).
- Mucin 2 (MUC2) is expressed in humans and is the predominant mucin (Nakamura et al., 2018).
- Trefoil factor 3 (TFF3) is critical for wound healing. TFF3 synergizes with mucin and enhances the protective barrier of the mucus layer (Nakamura et al., 2018).
- SAM pointed domain-containing ETS transcription factor (SPDEF) “drives terminal differentiation and maturation of secretory progenitors into goblet cells” (Nakamura et al., 2018, p.121).
- Krueppel-like factor 4 (KLF4) is a “goblet cell-specific differentiation factor in the colon. It controls goblet cell differentiation and activates mucin synthesis” (Nakamura et al., 2018, p.121).
- Critical for health to have proper functioning of the intestinal barrier between the host and microbe (Nakamura et al., 2018).

## Significance of Pathophysiology

- Cause of HAEC is unknown (Gosain et al., 2017).
- Several hypotheses for the cause of HAEC include “dysbiosis of the intestinal microbiome, impaired mucosal barrier function, altered innate immune responses, and bacterial translocation” (Gosain et al., 2017, p. 518).
- Goblet cell dysfunction may result in intestinal barrier dysfunction contributing to HAEC (Nakamura et al., 2018).

## Diagnosis of HAEC

- Challenging to diagnosis due to non-specific symptoms and has been underreported (Gosain et al., 2017).
- HAEC has been reported in 6% to 60% prior to pull-through surgery and 25% to 37% after surgery (Gosain et al., 2017).

Grade	Description	Clinical History	Physical Examination	Radiographic Findings
I	Possible HAEC	<input type="checkbox"/> Anorexia <input type="checkbox"/> Diarrhea	<input type="checkbox"/> Mild abdominal distention	<input type="checkbox"/> Normal <input type="checkbox"/> Mild ileus gas pattern
II	Definite HAEC	<input type="checkbox"/> History of past episode of HAEC <input type="checkbox"/> Explosive diarrhea <input type="checkbox"/> Fevers <input type="checkbox"/> Lethargy	<input type="checkbox"/> Fever <input type="checkbox"/> Tachycardia <input type="checkbox"/> Abdominal distention <input type="checkbox"/> Abdominal tenderness <input type="checkbox"/> Explosive gas/stool on DRE	<input type="checkbox"/> Ileus gas pattern <input type="checkbox"/> Air/fluid levels <input type="checkbox"/> Dilated loops of bowel <input type="checkbox"/> Recto-sigmoid cutoff
III	Severe HAEC	<input type="checkbox"/> Obstipation <input type="checkbox"/> Obtunded	<input type="checkbox"/> Decreased peripheral perfusion <input type="checkbox"/> Hypotension <input type="checkbox"/> Altered mentation <input type="checkbox"/> Marked abdominal distention <input type="checkbox"/> Peritonitis	<input type="checkbox"/> Pneumatosis <input type="checkbox"/> Pneumoperitoneum

Table 1. Guideline for the diagnosis of HAEC with grading I (possible HAEC) to III (severe HAEC). (Table from Fig.1 Gosain et al, 2017).

## Treatment of HAEC

- Based on the severity of symptoms (Gosain et al., 2017).
- Proper technique of rectal irrigations one to three times per day (Avansino & Levitt, 2017).
- Intravenous fluids (Avansino & Levitt, 2017).
- Broad-spectrum antibiotics (Avansino & Levitt, 2017).
- Further evaluation if corrective surgery is needed to remove retained HD (Langer et al., 2017).
- Rectal Botox (Langer et al., 2017).
- Probiotics “play a protective role in maintaining intestinal mucosal integrity through a number of different interactions, including alterations in mucosal cytokine expressions, competing with intestinal pathogens for mucosal receptors, thereby increasing transepithelial resistance” (Nakamura, Lim, & Puri, 2017).

Grade	Disposition	Diet	Antibiotics	Irrigations	Surgery
I	Outpatient	<input type="checkbox"/> Oral hydration	<input type="checkbox"/> PO metronidazole	<input type="checkbox"/> Consider rectal irrigations	
II	Outpatient or Inpatient	<input type="checkbox"/> Clear liquids or NPO <input type="checkbox"/> IVF hydration	<input type="checkbox"/> Metronidazole (PO or IV) <input type="checkbox"/> Consider broad spectrum coverage [ampicillin (IV) and gentamicin (IV) or piperillin/tazobactam (IV)]	<input type="checkbox"/> Rectal irrigations	
III	Inpatient, possible ICU	<input type="checkbox"/> NPO <input type="checkbox"/> IVF hydration	<input type="checkbox"/> Metronidazole (IV) <input type="checkbox"/> Broad spectrum coverage [ampicillin (IV) and gentamicin (IV) or piperillin/tazobactam (IV)]	<input type="checkbox"/> Rectal irrigations	<input type="checkbox"/> Proximal diversion for failure to improve with non-operative management <input type="checkbox"/> Exploration for pneumoperitoneum

Table 2. Guideline for management of HAEC based on guidelines for the diagnosis of HAEC (Table 1). (Table from Fig.2 Gosain et al, 2017).

## Proper Technique of Rectal Irrigations

- Large bore Foley catheter (20-24 Fr).
- Up to 1 to 2 liters of 0.9% sodium chloride.
- Use 20mL of sodium chloride at a time with a 60mL cath tip syringe.
- Irrigate child until output is clear.
- Child should not retain more than 20mL per kilogram (kg).

(Avansino & Levitt, 2017).

## Implications for Nursing Care

- Education to families on proper rectal irrigation technique (Avansino & Levitt, 2017).
- Make sure the family has the appropriate supplies for rectal irrigations (Avansino & Levitt, 2017).
- Knowledge and education on recognizing the signs and symptoms of HAEC (Gosain et al., 2017).
- Education on importance to follow the recommendations given by the healthcare provider.
- Collaboration with healthcare providers for the best overall care for the patient.

## Conclusion

HAEC continues to lead to life-threatening conditions for individuals with HD. HAEC continues to be underreported and undertreated. Proper preventative measures and treatment for those presenting with symptoms of enterocolitis are needed to decrease the risk of developing severe HAEC. Further research is needed on the underlying cause and pathophysiology of HAEC.

## Topic Selection

The author works for the Center for Colorectal and Pelvic Reconstruction (CCPR) at Nationwide Children's Hospital. She takes care of patients with HD and has recognized the importance of proper education on the signs and symptoms of HAEC. The author also recognizes the importance for all healthcare providers to provide proper treatment plans based on the patients symptoms, and the importance for all healthcare providers to understand the proper technique for rectal irrigations.

## References



## Additional Sources



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