



Evaluation and Management of a Patient with Hirschsprung Disease

Description of the Activity	<p>Hirschsprung disease (HD) is a condition that every pediatric surgeon will encounter during their career. Pediatric surgeons may encounter these patients during the newborn period or later in life for those not diagnosed in early infancy. The surgeon must be able to diagnose and treat these patients in both the inpatient and outpatient settings and on an acute and elective basis. Given the lifelong implications of HD, the pediatric surgeon will ensure proper transition of care as the patient becomes an adult.</p>
Functions	<ul style="list-style-type: none">❖ Nonoperative/Preoperative<ul style="list-style-type: none">➤ Perform a targeted history and physical, including an inquiry into the timing of the first passage of meconium.➤ Recognize the signs and symptoms of Hirschsprung-associated enterocolitis (HAEC) and its management.➤ Identify initial radiographic studies used to aid in the diagnosis, including interpretation and limitations of the studies.➤ Determine whether urgent surgical intervention is warranted, including indications for stoma creation.➤ Discuss the role of rectal biopsy in the diagnosis, including biopsy techniques and the advantages and disadvantages of each.➤ Formulate an operative approach based on the patient's age, weight, and clinical presentation.➤ Recognize when additional workup for comorbidities is needed (trisomy 21, congenital central hypoventilation syndrome, hereditary HD).➤ Obtain informed consent, describing the indications, risks, benefits, alternatives, and potential complications of the planned operation, including nuances relevant to the patient's individual condition and comorbidities, and ensure familial understanding. Document the informed consent discussion in the medical record.➤ Recognize the signs, symptoms, and severity of HAEC.➤ Demonstrate understanding of the differential diagnosis of neonatal distal bowel obstruction.➤ Devise an operative plan, and communicate it to the operative team (anesthesia, nursing, techs, assistants), including patient position, anesthesia needs, special instrumentation, and postoperative planning.❖ Intraoperative<ul style="list-style-type: none">➤ Manage the perioperative environment, including temperature regulation in the neonate and sites for peripheral intravenous lines, as well as room setup, equipment needed, procedural time-out, specimen processing (and communication with pathology), wound classification, and the importance of debriefing.➤ Recognize the correct patient positioning based on the age of the patient and the surgical intervention being performed.➤ Administer preoperative antibiotics.➤ Collaborate with the perioperative health care team to ensure an environment that promotes the optimal care of the patient.➤ Recognize the histologic findings of HD and additional stains used to improve diagnosis.➤ Ensure a circular segment of the pull-through segment is reviewed by the pathologist intraoperatively to ensure circumferential ganglion cells.➤ Pull-through:<ul style="list-style-type: none">▪ Recognize the common approaches to abdominal dissection based on the level of the transition zone (open, laparoscopic, transanal).



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- Identify the crucial steps of the operation: sequential biopsies to identify the transition zone, skeletonization of the aganglionic bowel, and mobilization of the proximal ganglionic bowel to allow a tension-free delivery to the anus.
- Recognize the recommended minimum distance from the biopsy site to the pull-through segment to avoid a transition zone pull-through.
- Laparoscopic Soave:
 - Identify the common positioning of trocars.
 - Recognize the need to mobilize the colon while preserving the marginal artery.
 - Determine adequate length by grasping the bowel 10 to 20 cm proximal to the transition zone and directing it to the pelvis.
 - Recognize the location of the mucosal incision in relation to the dentate line.
 - Identify the plane between the submucosa and the muscularis, and recognize the need to continue in this plane until the intra-abdominal dissection is met.
 - Recognize the need to divide the muscular wall, pull the colon through the muscular cuff, divide the cuff posteriorly, and divide the colon.
 - Complete the anastomosis by placing traction sutures to ensure proper orientation, and then additional sutures to create a tight anastomosis.
- Swenson:
 - Perform circumferential mobilization of the rectum to the pelvic floor or as low as can be safely performed.
 - Recognize the need to stay close to the rectal wall, identify and avoid the ureters, and in boys, protect the vas deferens.
 - Perform transanal dissection by incising full-thickness through the rectal wall circumferentially, 1 cm above the dentate line. Recognize the continued need to stay close to the rectal wall.
 - Recognize complications associated with each operative technique (Swenson vs Soave vs Duhamel).
- Leveling ostomy:
 - Access the abdominal cavity using a laparoscopic or open approach.
 - Identify the target of the seromuscular biopsy.
 - Determine the location of the ostomy based on the findings of the frozen section.
 - Consider a 4-quadrant biopsy at the point chosen for the stoma to avoid a transition zone stoma.
 - Recognize long-segment HD with serial biopsies and the need for proximal diversion.
- ❖ Postoperative
 - Communicate the postoperative plan of care to the family and other involved health care team members.
 - Verbalize the importance of avoiding instrumentation/manipulation of the rectum to minimize the risk of anastomotic disruption.
 - Recognize the need for short-term follow-up after discharge (within 2-3 weeks) to determine the need for routine rectal dilations or continued rectal irrigations.
 - Identify the signs, symptoms, and treatment of HAEC, and recognize its occurrence before or after definitive surgical intervention.
 - Recognize special populations at increased risk for HAEC (patients with trisomy 21 and long-segment disease).
 - Recognize the need for transfer to a higher-level facility for patients with long-segment HD.



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Scope

- ❖ In scope
- ❖ Diagnoses
 - HAEC
 - Normal-segment HD
- ❖ Procedures
 - Botox injection
 - Laparoscopic-assisted /open pull-through
 - Leveling colostomy/ileostomy
 - Rectal biopsy (punch or open)
 - Rectal irrigation/decompression
 - Soave/Swenson with or without Duhamel
- ❖ Special populations:
 - Hereditary populations
 - Long-segment
 - Trisomy 21
- ❖ Out of scope
 - Diagnoses/procedures
 - Anorectal malformation (see specific EPA)
 - Idiopathic constipation



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<p style="text-align: center;">1</p> <p>Framework:</p> <p>The learner demonstrates understanding of information and has basic skills</p> <p>What a new pediatric surgery fellow should know</p> <p>Entrustment:</p> <p>The attending will show and tell or the learner acts as first assistant.</p>	<ul style="list-style-type: none"> • With active guidance, performs a relevant H&P including inquiry of passage of first meconium or stooling history • With active assistance, performs a suction rectal biopsy in a patient with suspected HD and interprets pathologic findings • With active guidance, identifies a patient in need of rectal irrigations, including one presenting with HAEC, and provides general parental education on their performance • With active guidance, interprets findings of initial studies such as a contrast enema to aid in the diagnosis of HD (e.g., abnormal rectosigmoid ratio) • With active assistance, communicates the basic facts of HD to the family in a respectful and culturally sensitive manner 	<ul style="list-style-type: none"> • With active guidance, participates in preparation and positioning of the patient and demonstrates understanding of variances depending on the HD surgical intervention to be performed • With active assistance, performs leveling biopsies and serves as first assistant with the pull-through procedure (open or lap-assisted) • With active assistance, recognizes HD transition zone pathophysiology and the implications of a transition zone pull-through • With active assistance, articulates surgical options and general principles such as identification of the transition zone, preservation of the blood supply of the proximal intestine, and the importance of tension-free repair and avoiding injury to the dentate line • With active assistance, performs an open rectal biopsy when a suction rectal biopsy is nondiagnostic or not possible and verbalizes complications of the procedure • With active assistance, discusses intraop biopsy/specimen findings with the pathologist 	<ul style="list-style-type: none"> • With active assistance, communicates the basic steps of the postop plan to the family and other health care team members, including immediate postop needs and the need for short-term follow-up • With active assistance, recognizes the short- and long-term complications of a pull-through procedure such as HAEC, anastomotic stricture, cuff abscess, soiling/incontinence, and obstructive symptoms • With active assistance, identifies the rationale for long-term multidisciplinary management



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		<ul style="list-style-type: none"> With active guidance, recognizes a patient with long- segment HD while performing a leveling biopsies, and identifies the need for proximal diversion. With active guidance, considers HD in a patient undergoing diversion after presenting with an acute abdomen (i.e., cecal or appendiceal perforation) and performs the appropriate workup (i.e., rectal biopsy). 	
<p style="text-align: center; font-weight: bold;">2</p> <p>Framework:</p> <p>The learner demonstrates understanding of the steps of the operation but requires direction through principles and does not know the nuances of a basic case</p> <p>Entrustment:</p> <p>The learner can use the tools but may not know exactly what, where, or how to do it.</p> <p>The attending gives active help throughout the case to maintain forward progression or may need to</p>	<ul style="list-style-type: none"> With indirect supervision, performs a relevant H&P, including inquiry of passage of first meconium or stooling history in an uncomplicated patient with HD With direct supervision, performs a suction rectal biopsy in a patient with suspected HD and interprets pathologic findings With direct supervision, identifies a patient in need of rectal irrigations, including one presenting with HAEC, and provides parental education on their performance With indirect supervision, interprets findings of initial studies such as a contrast enema to aid in the diagnosis of HD (e.g., abnormal rectosigmoid ratio) With direct supervision, thoroughly explains the pathophysiology of HD to the 	<ul style="list-style-type: none"> With direct supervision, prepares and positions the patient depending on the HD surgical intervention to be performed With direct supervision, performs the pull-through procedure (open or lap-assisted) With direct supervision, recognizes HD transition zone pathophysiology and implications of a transition zone pull-through With direct supervision, articulates surgical options and general principles such as identification of the transition zone, preservation of the blood supply of the proximal intestine, and the importance of tension-free repair and avoiding injury to the dentate line 	<ul style="list-style-type: none"> With direct supervision, communicates the postop plan to the family and other health care team members, including immediate postop needs and the need for short-term follow-up With direct supervision, recognizes the short- and long-term complications of a pull-through procedure such as HAEC, anastomotic stricture, cuff abscess, soiling/incontinence, and obstructive symptoms and delineates a basic treatment plan for short-term complications With direct supervision, identifies the rationale for long-term multidisciplinary management



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<p>take over the case at a certain point</p>	<p>family in a respectful and culturally sensitive manner and obtains informed consent</p>	<ul style="list-style-type: none"> • With direct supervision, performs an open rectal biopsy when a suction rectal biopsy is nondiagnostic or not possible and verbalizes complications of the procedure • With direct supervision, discusses intraop biopsy/specimen findings with the pathologist • With direct supervision, recognizes a patient with long-segment HD while performing a leveling biopsies, and identifies the need for proximal diversion. • With direct supervision, considers HD in a patient undergoing diversion after presenting with an acute abdomen (i.e., cecal or appendiceal perforation) and performs the appropriate workup (i.e., rectal biopsy). 	
<p>3</p> <p><u>Framework:</u></p> <p>The learner has a good understanding of surgical options and techniques but does not recognize abnormalities and does not understand the nuances of a complicated case</p> <p><u>Entrustment:</u></p>	<ul style="list-style-type: none"> • With indirect supervision, performs a targeted H&P and identifies the diagnostic workup and surgical management for a complex patient with HD presenting with an acute abdomen, trisomy 21, ELBW, or a delayed presentation • With indirect supervision, performs a suction rectal biopsy in a patient with suspected HD and interprets pathologic findings 	<ul style="list-style-type: none"> • With indirect supervision, prepares and positions the patient depending on the HD surgical intervention to be performed • With indirect supervision, performs the pull-through procedure (open or lap-assisted) • With indirect supervision, recognizes HD transition zone pathophysiology and implications of a transition zone pull-through 	<ul style="list-style-type: none"> • With indirect supervision, communicates a comprehensive postop plan to the family of a complex patient and other health care team members, including short- and long-term complications/goals of care • With indirect supervision, recognizes the short- and long-term complications of a pull-through procedure such as HAEC, anastomotic stricture, cuff abscess, soiling/incontinence, and



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<p>The learner can perform the operation/task independently in the uncomplicated patient</p> <p>or</p> <p>The attending provides passive/indirect supervision/suggestions in the complicated patient but still allows the learner to perform the operation/task themselves</p>	<ul style="list-style-type: none"> With indirect supervision, identifies a patient in need of rectal irrigations, including one presenting with HAEC, promptly institutes management, and provides comprehensive parental education on their performance With indirect supervision, identifies uncommon fluoroscopic imaging findings that might suggest total colonic disease (e.g., microcolon, normal-appearing colon) With indirect supervision and in a respectful and culturally sensitive manner, discusses the proposed operative plan with the family and uses shared decision-making to ensure a personalized plan of care that aligns with the family's values, goals, and preferences 	<ul style="list-style-type: none"> With indirect supervision, articulates surgical options and general principles such as identification of the transition zone, preservation of the blood supply of the proximal intestine, and the importance of tension-free repair and avoiding injury to the dentate line With indirect supervision, performs an open rectal biopsy when a suction rectal biopsy is nondiagnostic or not possible and verbalizes complications of the procedure With indirect supervision, discusses intraop biopsy/specimen findings with the pathologist With indirect supervision, recognizes a patient with long-segment HD while performing a leveling biopsy and identifies the need for proximal diversion With indirect supervision, considers HD in a patient undergoing diversion after presenting with an acute abdomen (cecal or appendiceal perforation) and performs the appropriate workup (rectal biopsy) 	<p>obstructive symptoms and delineates a comprehensive treatment plan for short-term complications</p> <ul style="list-style-type: none"> With indirect supervision, identifies the rationale for long-term multidisciplinary management and occasionally participates in this management (participation in clinic, discussion of follow-up care with the primary surgical attending)
<p>4</p> <p>Framework:</p>	<ul style="list-style-type: none"> Independently performs a targeted H&P and promptly identifies the diagnostic workup and surgical management for a complex patient with HD presenting with 	<ul style="list-style-type: none"> Independently prepares and positions the patient depending on the HD surgical intervention to be performed 	<ul style="list-style-type: none"> Independently communicates a comprehensive postop plan to the family of a complex patient and other health care team members, including



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<p>The learner has a strong and in-depth understanding of surgical options and techniques</p> <p><u>Entrustment:</u></p> <p>Can perform the operation/task independently in complicated cases</p> <p>or</p> <p>The attending may need to provide indirect supervision or suggestions in the context of extremely rare or severely complicated cases</p>	<p>an acute abdomen, trisomy 21, ELBW, or a delayed presentation</p> <ul style="list-style-type: none">Independently performs a suction rectal biopsy in a patient with suspected HD and interprets and articulates pathologic findingsIndependently identifies a patient in need of rectal irrigations, including one presenting with HAEC, promptly institutes management, and provides comprehensive parental education on their performanceIndependently identifies uncommon fluoroscopic imaging findings that might suggest total colonic disease (e.g., microcolon, normal-appearing colon)Independently and in a respectful and culturally sensitive manner, discusses the proposed operative plan with the family and uses shared decision-making to ensure a personalized plan of care that aligns with the family's values, goals, and preferences	<ul style="list-style-type: none">Independently performs the pull-through procedure (open or lap-assisted)Independently recognizes HD transition zone pathophysiology and implications of a transition zone pull-throughIndependently articulates surgical options and general principles such as identification of the transition zone, preservation of the blood supply of the proximal intestine, and the importance of tension-free repair and avoiding injury to the dentate lineIndependently performs an open rectal biopsy when a suction rectal biopsy is nondiagnostic or not possible and verbalizes complications of the procedureIndependently communicates with the pathologist before the procedure, intraoperatively, and postoperatively regarding specimen findingsIndependently, recognizes a patient with long- segment HD while performing a leveling biopsies, and identifies the need for proximal diversion.Independently considers HD in a patient undergoing diversion after presenting with an acute abdomen (i.e., cecal or	<p>short- and long-term complications/goals of care</p> <ul style="list-style-type: none">Independently recognizes the short- and long-term complications of a pull-through procedure such as HAEC, anastomotic stricture, cuff abscess, soiling/incontinence, and obstructive symptoms as well as unique concerns in a patient with total colonic aganglionosis and delineates a comprehensive treatment plan for short- and long-term complicationsIndependently identifies the rationale for long-term multidisciplinary management and frequently participates in this management (participation in clinic, discussion of follow-up care with the primary surgical attending)



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		appendiceal perforation) and performs the appropriate workup (i.e., rectal biopsy).	