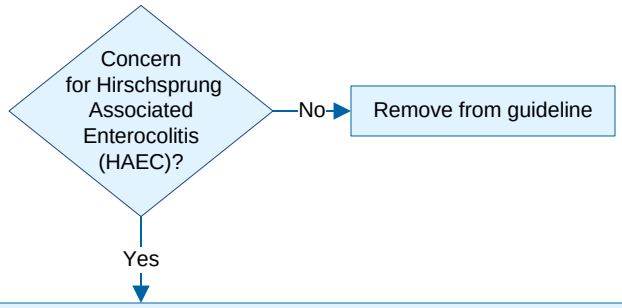
Hirschsprung Disease Care Guideline

Emergency Department / Initial Workup and Diagnosis

Inclusion Criteria:

Patients presenting with:

- Failure to pass meconium within 24-48 hours of life after *confirmation of a patent anus*
- Abdominal distention, non-bilious or bilious emesis, and constipation
- Any patient with suspected or confirmed Hirschsprung Disease
- Bilious emesis ONLY if malrotation with volvulus ruled-out



Differential Diagnoses to Consider

- Meconium plug syndrome
- Small left colon syndrome (seen with diabetic mothers)
- Distal small bowel/colonic atresia
- Meconium ileus with cystic fibrosis

Assessment and Immediate Treatment

- Assess for the presentation of the above inclusion criteria and if initial concern for enterocolitis:
  - If enterocolitis is strongly suspected or the patient appears critically ill or septic:
    - Initiate rectal irrigation immediately using a minimum of 20 mL/kg of normal saline (NS).
    - Continue irrigation until the effluent runs clear.
    - Refer to Patient Care Policy F963v1 - Retrograde and Antegrade Colonic Irrigation (#Rectal Irrigation, #MACE, #Malone, #Cecostomy, #Appendicostomy).
  - Start Metronidazole 10 mg/kg IV q8h.
- Consult surgery.

Imaging

- AXR with generalized gaseous distention of the bowel, sometimes absence of rectal gas.
- Contrast enema – used to identify bowel caliber changes indicative of a transition zone.
  - If the patient is clinical stable, do not perform rectal irrigation prior to the contrast enema.
  - If the patient appears critically ill or septic, initiate rectal irrigation immediately, following the recommendations in “Assessment and Immediate Treatment”.
  - Schedule the contrast enema once the patient is clinically stable.

Further Workup

- **Anorectal Manometry**
  - The recto-anal inhibitory reflex (RAIR) is an involuntary anal reflex that follow distention of the rectum.
  - In Hirschsprung’s patients, the RAIR is absent during manometry testing.
  - An absent RAIR indicates a need for rectal biopsy.
- **Rectal Biopsy**
  - The gold standard for diagnosis to establish aganglionosis performed at bedside, unless > 1yo.
    - < 1yo: Suction rectal biopsy, performed at bedside, no anesthesia required, do not need to be NPO.
    - > 1yo: Full thickness rectal biopsy, which is performed in the OR and requires general anesthesia.
    - While awaiting biopsy results, begin or continue irrigations (irrigations are held for 12-24 hours after biopsy is performed, to avoid trauma).
      - ➢ Minimum volume 20 mL/kg NS divided in 3 aliquots, as ordered.
  - Results showing no ganglion cells and presence of nerve hypertrophy, with calretinin staining confirms Hirschsprung Disease.

Discharge Teaching

- ALL parents should be taught signs and symptoms of enterocolitis *(see chart on page 3)* and how to do rectal irrigations.
- Educate parent to call surgery office and start irrigations for no stool for 24hrs, lots of foul-smelling flatus/stool or abdominal distention.
- Rectal irrigation video link for medical care givers and families:
  - https://cchmcstream.cchmc.org/MediasiteEX/Play/545154a603a844e8988ef74cd5b4c1c11d
- DME need for rectal irrigations with case management support:
  - Red rubber or foley catheters
  - 60mL catheter tip syringe
  - Normal saline
  - Lubricant jelly
- Pediatric Surgery follow-up appointment in 2-3 weeks.

Approved Evidence Based Medicine Committee
Original: 9/18/2019
Review/Revised: 9/18/2024

Hirschsprung Disease Care Guideline with Enhanced Recovery After Surgery (ERAS) Principles

Surgical Care

Preoperative Considerations

- Integrated ERAS principles:
  - Educate patient and family on operative procedure, post-operative care, including appropriate expectation of post-operative care and discharge criteria. Patient and family education materials are distributed to the family.
- Feeds as tolerated if abdominal exam is stable and patient is stooling.
- Discussion of PICC placement for TPN if NPO / not tolerating feeds.
- Orogastric/Nasogastric tube (Salem sump) to low-intermittent wall suction for gastric decompression for ongoing emesis.
- Rectal irrigations as ordered.
- Begin teaching parents rectal irrigations.
- Metronidazole 10 mg/kg q8hrs if with signs of enterocolitis.

Intraoperative Considerations

- Term baby with good transition zone visible on contrast enema, or with enterocolitis that is controlled with irrigations ⇨ consider primary pull-through procedure.
- Term baby with no identifiable transition zone ⇨ consider progressive biopsies and possible ostomy.
- Intraoperative progressive biopsies performed to determine level at which ganglion cells are found (transition zone) and bowel resection is performed to removed the aganglionic segment.
- Premature or small baby ⇨ consider scheduled irrigations and await growth prior to surgical intervention, possible discharge home prior to operation.
- If planning for ostomy:
  - Consider applying stool to perianal area for x1 hour/day for 1-2 months prior to pull-through surgery to acclimate skin to stool and prevent perineal breakdown postoperatively.
  - Teach family how to perform rectal irrigations of the diverted colon, to be performed once a week.
- Laparoscopic-assisted transanal endorectal pull-through can be with or without leveling colostomy procedure performed.

Postoperative Care

- NPO until return of bowel function, consider IV nutrition for longer than 5 days NPO.
- Surgeon's preference to insert salem sump and set to low-intermittent wall suction.
- Routine pain management medication.
- Cefoxitin 30mg/kg IV q8h x 2 doses, Dosing Guidelines: neonates, postoperative prophylaxis.
- S/p laparoscopic-assisted transanal endorectal pull-through strict adherence to "nothing per rectum" order. No rectal irrigations, medication, suppositories, temperatures, or stimulation due to risk of anastomotic disruption.
- Often Foley catheter is left in place for 24-48 hours.
- If ostomy is created, consult Skin, Wound, Ostomy Team (SWOT) and assess stoma for tissue perfusion, evidence of prolapse or retraction, and stool output.
- Expect frequent loose stools, making anorectal skin care extremely important. Prophylactic perianal care should be started immediately postoperatively.
- SWOT consult as needed.
- Referral to Colorectal Clinic.

Discharge Teaching

- ALL parents should be taught signs and symptoms of enterocolitis *(see chart on page 3)* and how to do rectal irrigations.
- Educate parent to call surgery office and start irrigations for no stool for 24hrs, copious amounts of foul-smelling flatus/stool or abdominal distention.
- Routine ostomy management care if ostomy present. Involve SWOT team.
- Routine rectal irrigation may be required routinely postoperatively.
- Rectal irrigation video link for medical care givers and families:
  - https://cchmcstream.cchmc.org/MediasiteEX/Play/545154a603a844e8988ef74cd5b4c1c11d
- Coordinate with case management to arrange Durable Medical Equipment (DME) for rectal irrigations:
  - Red rubber or foley catheters
  - 60mL catheter tip syringe
  - Normal saline
  - Lubricant jelly
    - Ensure ostomy supplies are included, if necessary.
- Pediatric Surgery follow-up appointment in 2-3 weeks.
  - Sizing of the anastomosis, and possible dilations may be started by the discretion of the surgeon at this time.
- Follow-up outpatient appointment with SWOT, if needed.

Hirschsprung Disease Care Guideline

Treatment for Patients with Known Diagnosis

Hirschsprung Associated Enterocolitis (HAEC): greatest cause of ***morbidity and mortality*** in children with Hirschsprung Disease.

- Can occur in both preoperative and postoperative for many year after surgery. Parents should call Colorectal Clinic or Surgeon On-Call once symptoms start ***immediately***. Decision to treat inpatient or outpatient is made at the discretion of the provider.

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

From “Guidelines for the diagnosis and management of Hirschsprung-associated enterocolitis”

- **Criteria to start at-home treatment:**
  - Grade 1 (possible HAEC) – outpatient management can typically be used.
    - Rectal irrigations and oral hydration, possibly PO metronidazole.
  - Grade II (definite HAEC) – may be inpatient or outpatient.
    - Follow-Up: 24-48 hours by phone.
      - If improvement: continue with irrigations x 7 days total, call at end of treatment stop if symptoms resolved.
      - If NO improvement: evaluate in person (clinic same-day or next day. ED immediately if worsening and after hours.
- **Criteria to be evaluated inpatient:**
  - **Workup:**

Study/Exam	Moderate	Severe
2 View ABD XR	• Ileus gas pattern	• Air fluid levels • Dilated bowel loops • Pneumatosis • Recto-sigmoid cut off
CBC w/ diff	• Leukocytosis	• Elevated neutrophil count • Bandemia
Rectal Exam	• Explosive stool	• Explosive stool

  - **Treatment:**
    - Rectal irrigations: should be started ***immediately.***
    - IV fluid hydration.
    - Metronidazole 10 mg/kg IV q8h.
      - May start with IV until tolerating PO.
      - If stool toxin positive for *Clostridioides difficile* (*C.diff*), change antibiotic to Vancomycin PO.
    - Bowel rest.
    - Close monitoring.
  - **Discharge Criteria:**
    - Adequate PO intake.
    - Tolerating PO antibiotics.
    - Symptom improvement.
  - **Discharge Treatment:**
    - Continue rectal irrigations (BID x 10 days total, then as needed).
    - Complete 7-day antibiotic course.
  - **Follow-Up outpatient**

Hirschsprung Disease Care Guideline
References

Das, K., & Mohanty, S. (2017). Hirschsprung disease - current diagnosis and management. *Indian Journal of Pediatrics, 84*(8), 618-623. https://doi.org/10.1007/s12098-017-2371-8 (Level V)

Freedman-Weiss, M. R., Chiu, A. S., Caty, M. G., & Solomon, D. G. (2019). Delay in operation for Hirschsprung disease is associated with decreased length of stay: a 5-Year NSQIP-Peds analysis. *Journal of Perinatology: Official Journal of the California Perinatal Association, 39*(8), 1105-1110. https://doi.org/10.1038/s41372-019-0405-y (Level III)

Gosain, A., Frykman, P. K., Cowles, R. A., Horton, J., Levitt, M., Rothstein, D. H., . . . Goldstein, A. M. (2017). Guidelines for the diagnosis and management of Hirschsprung-associated enterocolitis. *Pediatric Surgery International, 33*(5), 517-521. https://doi.org/10.1007/s00383-017-4065-8 (Level V)

Langer, J. C., Rollins, M. D., Levitt, M., Gosain, A., de la Torre, L., Kapur, R. P., . . . Goldstein, A. M. (2017). Guidelines for the management of postoperative obstructive symptoms in children with Hirschsprung disease. *Pediatric Surgery International, 33*(5), 523-526. https://doi.org/10.1007/s00383-017-4066-7 (Level V)

Soh, H. J., Nataraja, R. M., & Pacilli, M. (2018). Prevention and management of recurrent postoperative Hirschsprung's disease obstructive symptoms and enterocolitis: Systematic review and meta-analysis. *Journal of Pediatric Surgery, 53*(12), 2423-2429. https://doi.org/10.1016/j.jpedsurg.2018.08.024 (Level I)

Vilanova-Sanchez, A., & Levitt, M. A. (Eds.). (2020). *Pediatric Colorectal and Pelvic Reconstructive Surgery* (1st ed.). Boca Raton: CRC Press. https://doi.org/10.1201/9780429027789