

## Hirschsprung Disease Clinical Guideline

### Inclusion Criteria

Patients presenting with:

- Failure to pass meconium in first 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

Available resources:

- Hirschsprung Disease Patient and Family Education Sheet

Applicable Consultations:

- GI
- Pediatric General, Thoracic & Trauma Surgery

### Clinical Evaluation:

- Differential Diagnoses to consider in evaluation: meconium plug syndrome, small left colon syndrome (in diabetic mothers), distal small bowel/colonic atresia, and meconium ileus with cystic fibrosis
- Imaging:
  - AXR with generalized gaseous distention of the bowel, sometimes absence of rectal gas
  - Contrast enema. Area of bowel caliber change reflective of transition zone.
    - Do NOT irrigate prior to contrast enema
- Suction rectal biopsy: gold standard for diagnosis to establish aganglionosis performed at bedside
- While awaiting biopsy results, begin or continue irrigations to evacuate stool and relieve obstruction (irrigations are held for 12-24 hours after biopsy is performed to avoid trauma)
  - 20 ml/kg of NS divided in 3 aliquots every 6-8 hrs as ordered
- Results showing no ganglion cells and presence of nerve hypertrophy confirms Hirschsprung Disease.
  - Surgical recommendations:
    - Term baby with good transition zone visible→ consider primary pull through procedure
    - Term baby with no identifiable transition zone→ consider leveling biopsies and possible ostomy
    - Premature or Small baby→ consider scheduled irrigations and await growth prior to surgical intervention

**Hirschsprung Associated Enterocolitis:** greatest cause of *morbidity and mortality* in children with Hirschsprung's disease.

- Can occur *in both preoperative and postoperative for many years* after surgery
- Signs and symptoms: Fever, abdominal distention, bloating, vomiting, severe constipation, foul smelling and/or explosive stools.
  - Symptoms can mimic presenting symptoms of Hirschsprung disease
- Treatment: cefoxitin

### Preoperative:

- Discussion of PICC placement for TPN
- Rectal irrigations as ordered
- Nasogastric tube (salem sump) to low intermittent wall suction for gastric decompression
- Antibiotics with signs of enterocolitis

### Intraoperative:

- Intraoperative progressive biopsies performed to determine level at which ganglion cells are found (transition zone) and bowel resection is performed to remove the aganglionic segment
- Transanal endorecto pull through or leveling colostomy procedure performed

### Postoperative:

- NPO with TPN via PICC
- Salem sump to low intermittent wall suction
- Routine pain management medications
- Antibiotics: 48 hours cefoxitin
- Strict adherence to “nothing per rectum” orders. No rectal medications, suppositories, temperatures, stimulation due to risk of anastomotic disruption
- Often Foley catheter is left in place for 24-48 hours
- If ostomy created, 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 skin care should be started immediately postop. Utilize topical A&D ointment or product with zinc oxide. Air drying of the skin is recommended.
- Bacitracin to rectal incision for 1 week postoperative

### Discharge teaching:

- Routine postoperative dilatations will be started by surgeon in outpatient clinic about 2 weeks post-op. Parents will be taught how to perform daily or twice daily dilatations.
- Routine ostomy management care if ostomy present. Involve SWOT team.
- Routine rectal irrigation maybe required postoperative
- Extensive education on signs/symptoms of Hirschsprung Associated Enterocolitis
- Education on how to perform rectal irrigation
- Rectal irrigation video link for medical caregivers and families:

<https://cchmcstream.cchmc.org/MediasiteEX/Play/545154a603a844e8988ef74cd5b4c1c11d>

- Home equipment needed for rectal irrigations with case management support:
  - red rubber or foley catheters
  - 60 ml catheter tip syringe
  - normal saline
  - lubricant jelly
- Pediatric Surgery follow up appointment in 2-3 weeks

Approved by Evidence-  
Based Medicine Committee  
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Reassess the appropriateness of Care Guidelines as condition changes and 24 hrs after admission. This guideline is a tool to aid clinical decision making. It is not a standard of care. The physician should deviate from the guideline when clinical judgment so indicates.

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