NGN NCLEX 2026 · VISUAL STUDY LIBRARY · PEDIATRIC GI

# Hirschsprung *Disease*

*Congenital aganglionic megacolon — the newborn who won't pass meconium*

PEDIATRIC GI | CONGENITAL | SURGICAL | NCJMM ALIGNED

⚑ SOURCE TRANSPARENCY

This topic is not present in the uploaded personal study notes. Content is synthesized from *Saunders Comprehensive Review for the NCLEX-RN Examination (2026 ed.)*, *ATI Nursing Care of Children*, *Lippincott Illustrated Reviews: Pediatrics*, and NASPGHAN clinical guidance on Hirschsprung disease and Hirschsprung-associated enterocolitis.

## 01 DEFINITION · OVERVIEW — What it is & why it matters

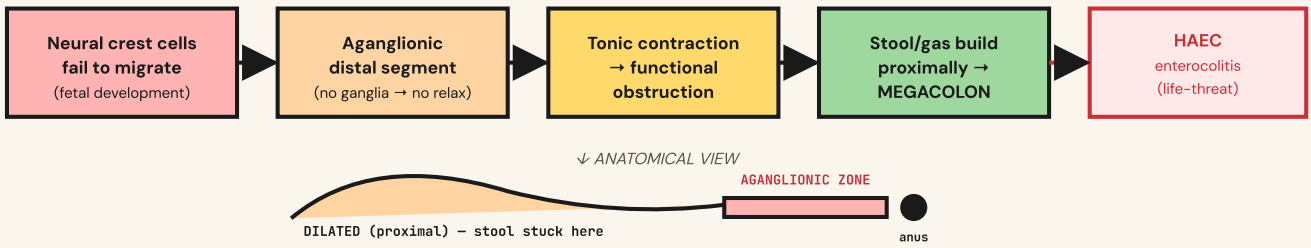
- **Hirschsprung Disease** is a **congenital absence of parasympathetic ganglion cells** in segments of the distal colon — most often the rectosigmoid region.
- Without ganglia, the affected segment **cannot relax or peristalse** → stool builds up proximally → the bowel above dilates → **functional intestinal obstruction** (megacolon).
- Most common cause of **lower bowel obstruction in newborns**. Higher incidence in males and in children with Down syndrome.
- Definitive treatment is **surgical resection** of the aganglionic segment (pull-through procedure).

## 02

PATHOPHYSIOLOGY

# From neural crest failure to megacolon

- During fetal development, **neural crest cells fail to migrate** all the way to the distal bowel.
- The distal segment is left **aganglionic** (no parasympathetic ganglia → no peristalsis).
- That segment stays **tonically contracted** — stool cannot pass through.
- Stool and gas accumulate **proximal** to the defect → the healthy bowel above dilates → **megacolon**.
- Stagnant stool + bacterial overgrowth → risk of **Hirschsprung-Associated Enterocolitis (HAEC)**.




## 03

SIGNS & SYMPTOMS

# By age of presentation

### 👶 Newborn (0–6 weeks)

- CARDINAL Failure to pass meconium within **24–48 hours** of birth
- Bilious (green) vomiting
- Abdominal distention
- Refusal to feed / poor feeding
- Reluctance to ingest fluids

### 👧 Older Infant / Child

- **Chronic constipation since birth**
- Ribbon-like, foul-smelling stools
- Abdominal distention with visible peristalsis
- Palpable fecal mass
- Failure to thrive / poor weight gain
- Anemia from malabsorption

**RED FLAG — HIRSCHSPRUNG-ASSOCIATED ENTEROCOLITIS (HAEC)**

Most dangerous, potentially fatal complication. Notify provider **immediately** if:

- **Explosive, foul, watery diarrhea** (sudden — in a child who normally can't stool!)
- Fever
- Rapidly worsening abdominal distention
- Lethargy, poor perfusion, signs of shock (tachycardia, hypotension)
- Bilious vomiting

PRIORITY NURSING INTERVENTIONS

# 04 ABC framework + Pre-op & Post-op

### A · Airway

- Monitor respiratory effort — severe abdominal distention can **splint the diaphragm**
- Elevate HOB to ease breathing

### B · Breathing

- Assess rate, depth, retractions, $SpO_2$
- Watch for tachypnea (early sepsis cue in HAEC)

### C · Circulation

- **Strict I&O, daily weights**
- Monitor for dehydration / shock (HAEC)
- IV fluid resuscitation as ordered

### 🔧 Pre-Op Priorities

- **NPO** status
- **NG tube** to low intermittent suction (decompression)
- IV fluids + electrolyte correction
- **Bowel prep with ISOTONIC saline enemas — NEVER tap water** (risk of water intoxication in children)
- Pre-op antibiotics (oral & systemic) to ↓ bowel flora
- **Measure abdominal girth daily** at the umbilicus (mark with pen)
- Vital signs every 4 hours; monitor temp closely
- Watch for HAEC at all times

### 🔧 Post-Op Priorities

- **NPO until bowel sounds return**; advance diet slowly
- **NO rectal temps, NO suppositories, NO digital exams** — protect suture line
- Position: **side-lying with knees flexed**; avoid stress on perineal wound
- Maintain NG decompression until peristalsis returns
- Stoma/colostomy care if temporary ostomy created
- Perineal/incisional skin care — keep dry & clean
- Pain management; monitor for infection
- Continue HAEC vigilance even *after* surgery

DIAGNOSTICS

# 05 Confirming the diagnosis

### Definitive

- GOLD STANDARD **Rectal suction biopsy** — shows *absence of ganglion cells* in the submucosa
- Often paired with histochemical staining (acetylcholinesterase activity)

### Supportive Studies

- **Contrast (barium) enema** — shows a "transition zone" between narrow aganglionic and dilated proximal bowel
- **Anorectal manometry** — absent rectoanal inhibitory reflex
- **Abdominal X-ray** — distended bowel loops, absence of rectal gas

06

PHARMACOLOGY

# Supportive drug therapy

| DRUG / CLASS | PURPOSE | KEY SIDE EFFECTS | NURSING CONSIDERATIONS |
|---|---|---|---|
| **IV broad-spectrum antibiotics** (e.g., ampicillin + gentamicin, piperacillin-tazobactam) | Treat / prevent HAEC; pre-op prophylaxis | Nephrotoxicity (gent), allergic rxn, C. diff overgrowth | Monitor renal function & peak/trough; watch IV site; assess for diarrhea |
| **Oral metronidazole** | HAEC treatment (anaerobic coverage) | Metallic taste, N/V, dark urine | Give with food if oral; monitor for neuro side effects (long term) |
| **Isotonic saline enema solutions** | Bowel prep / decompression / HAEC irrigation | Electrolyte shifts if used incorrectly | **Never use plain tap water** in children → water intoxication / hyponatremia |
| **Stool softeners** (e.g., docusate, PEG 3350) | Post-op & long-term — keep stool soft, prevent straining on anastomosis | Loose stools, cramping | Educate family on dose titration; ensure hydration |
| **IV crystalloids** (0.9% NS, LR) | Fluid resuscitation; correct dehydration/shock from HAEC | Fluid overload if excessive | Strict I&O, lung sounds, daily weights; monitor electrolytes |
| **Analgesics** (acetaminophen ± opioid post-op) | Pain control post-pull-through | Opioids → constipation, respiratory depression | Use FLACC / age-appropriate scale; opioids may delay return of peristalsis — minimize when possible |

07

NCLEX TEST TRAPS ⚠

# Wrong-thinking vs Right-thinking

| ✗ WRONG | ✓ RIGHT |
| --- | --- |
| "The baby will pass meconium late — just wait and observe." | **No meconium in 24–48 hrs is a RED FLAG.** Notify provider and prepare for diagnostic work-up — this is the cardinal sign of Hirschsprung. |
| Giving a **tap water enema** for bowel prep in a 4-week-old. | Use **isotonic (0.9%) saline** enemas. Children absorb free water rapidly → water intoxication, hyponatremia, seizures. |
| Taking a **rectal temperature** on the post-op pull-through child. | Use **axillary or tympanic** route. Rectal thermometers, suppositories, and digital exams can disrupt the suture line. |
| Confusing Hirschsprung with **intussusception** — "currant-jelly stools." | **Hirschsprung** = ribbon-like stools + no-meconium history.<br>**Intussusception** = sudden colicky pain + currant-jelly (blood + mucus) stools + sausage-shaped mass. |
| Confusing Hirschsprung with **pyloric stenosis**. | **Pyloric stenosis** = projectile *non-bilious* vomiting in a 2–8 week-old with an olive-shaped mass.<br>**Hirschsprung** = *bilious* vomiting + delayed meconium + distention. |
| Treating sudden **explosive diarrhea** post-op as a "good sign — bowel is working!" | That is **HAEC until proven otherwise** — life-threatening. Notify HCP, NPO, IV fluids, antibiotics, NG decompression, monitor for shock. |

08

PATIENT & FAMILY EDUCATION

# Discharge teaching priorities

## 🏠 Home Monitoring

- **Warning signs of HAEC** (teach BEFORE discharge): fever, foul/explosive diarrhea, abdominal swelling, lethargy, vomiting → **call provider or go to ER immediately**
- Monitor stool pattern, frequency, consistency
- Daily weights for first weeks if instructed
- Report any rectal bleeding, persistent vomiting, refusal to feed

## 💩 Bowel & Skin Care

- Keep **perianal skin clean and dry** — frequent diaper changes after pull-through
- Apply barrier cream as recommended
- If temporary **colostomy**: demonstrate stoma care, pouch changes, signs of complications (color change, prolapse, retraction, skin breakdown)
- Encourage adequate fluid & fiber intake (age-appropriate)

## 🥛 Nutrition & Growth

- Continue breast/formula feeding as tolerated post-op
- Watch growth — child may need extra calories to "catch up"
- Stool softeners may be needed long-term
- Avoid constipation — straining stresses the anastomosis

## 📅 Follow-up

- Keep ALL surgical follow-up appointments
- Some children need **anal dilations** at home — teach technique & frequency
- Long-term: some children have lingering constipation or fecal incontinence — bowel-training programs help
- Genetic counseling if family history (link to RET gene; ↑ risk with Down syndrome)

09

MEMORY BOOSTERS

# Mnemonics & pattern recognition

## H · I · R · S · C · H

- **H** — **H**as no ganglion cells
- **I** — **I**nability to pass meconium (within 24–48 hrs)
- **R** — **R**ibbon-like, foul-smelling stools
- **S** — **S**tretched bowel above defect (megacolon)
- **C** — **C**hronic constipation since birth
- **H** — **H**igh risk for enterocolitis (HAEC)

### 💡 Quick Associations

- **"No meconium by 48?"** → think Hirschsprung first
- **"Ribbon stool"** → narrow stool squeezed through stuck segment
- **"Explosive diarrhea in a constipated child"** → HAEC alarm
- **"Down syndrome + bowel symptoms"** → suspect Hirschsprung (3–10% co-occurrence)

### 🎯 Pattern Recognition: 3 N's of Hirschsprung

- **N**o meconium passage
- **N**o relaxation (aganglionic segment stays tight)
- **N**o rectal anything post-op (no temps, no suppositories, no digital exams)

10

NCJMM SIX-STEP CLINICAL JUDGMENT

# NGN Case Scenario

UNFOLDING CASE

*A* 2-DAY-OLD FULL-TERM NEWBORN *is admitted from the nursery. Mom reports the infant has* NOT PASSED MECONIUM SINCE BIRTH *. The baby has had* TWO EPISODES OF GREEN-TINGED EMESIS *, refuses to breastfeed, and has a* PROGRESSIVELY DISTENDED, FIRM ABDOMEN *. Vital signs: T 37.1°C, HR 168, RR 58, $SpO_2$ 96%. Abdomen tense; bowel sounds hypoactive.*

**1 — RECOGNIZE CUES**

No meconium passage in >48 hrs · bilious emesis · abdominal distention · refusal to feed · tachycardia · tachypnea · hypoactive bowel sounds.

**2 — ANALYZE CUES**

The combination of **delayed meconium + bilious vomiting + distention** in a newborn points to **distal bowel obstruction**. Bilious emesis indicates obstruction below the ampulla of Vater. Tachycardia + tachypnea are early signs of dehydration and respiratory compromise from abdominal distention splinting the diaphragm.

**3 — PRIORITIZE HYPOTHESES**

**#1 Hirschsprung disease** (most likely given classic presentation). Differential: intestinal atresia, meconium ileus (cystic fibrosis), malrotation/volvulus, imperforate anus. Rule out volvulus urgently — can become a surgical emergency.

**4 — GENERATE SOLUTIONS**

NPO · insert NG tube to low intermittent suction · establish IV access & start maintenance fluids · obtain abdominal X-ray and contrast enema · prepare for **rectal suction biopsy** · pre-op antibiotics · daily abdominal girth · continuous monitoring for HAEC and shock.

**5 — TAKE ACTION**

Place infant NPO and insert NG to suction · start IV NS bolus 10–20 mL/kg per protocol · obtain blood work (CBC, electrolytes, blood culture if febrile) · measure and mark abdominal girth at umbilicus · monitor VS q1h initially · prepare parents emotionally for biopsy and possible surgery · administer ordered IV antibiotics. **Use ONLY isotonic saline for any ordered enema — NEVER tap water.**

**6 — EVALUATE OUTCOMES**

**Expected:** abdominal girth decreases · NG drainage returns gastric contents · vital signs normalize · hydration restored (good urine output, moist mucous membranes) · biopsy confirms aganglionosis · NO signs of HAEC (no fever, no explosive diarrhea, no worsening distention) · family verbalizes understanding of plan of care.

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