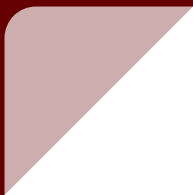


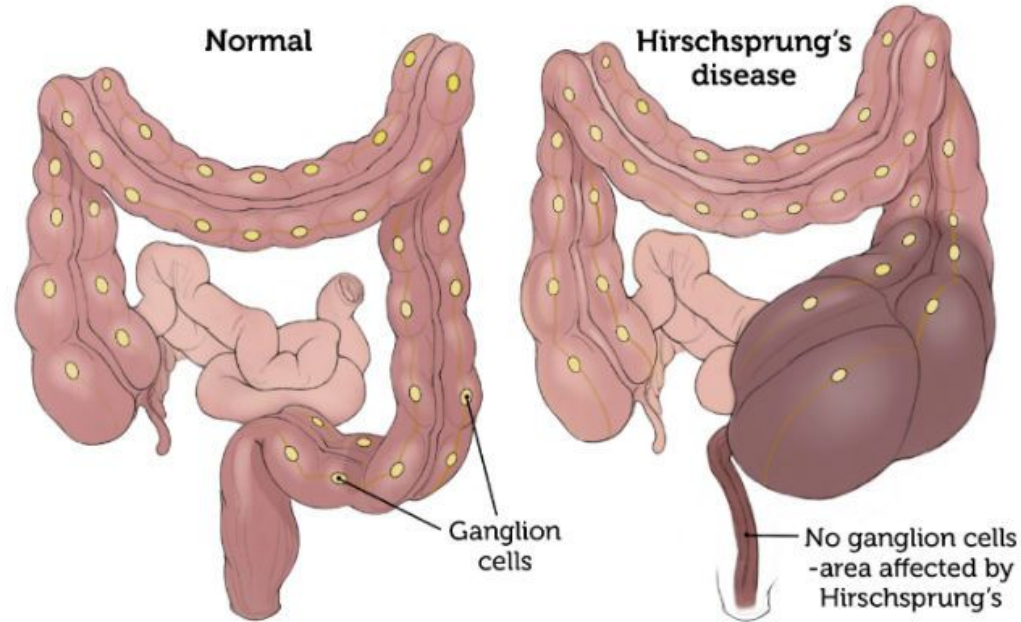
Hirschsprung's Disease

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What is Hirschsprung's Disease?

- Congenital disease that affects large intestines, most commonly the **distal colon and rectum**
- Unknown cause but associated with genetic mutations
- Improper development of the **ganglion (nerve) cells** in the colon
- No ganglion cells leads to inability to relax the segment, delaying progression of waste from digestion
- Colon swells with backed up stool causing **blockage of bowel** → constipation, partial or total obstruction



Risk Factors

- Parent or sibling with Hirschsprung's, especially mothers
- Males – **3-4x more common in boys** than girls
- Other Conditions – Down syndrome, Chagas disease, MEN II

Symptoms & Clinical Presentation

- Newborns
 - **Meconium ileus: failure to pass a bowel movement within first 24-48 hours of life**
 - Intestinal obstruction
 - Bilious (green/brown) vomiting
 - Abdominal distention (swollen belly)
- Children
 - Chronic constipation
 - Abdominal distention (swollen belly)
 - Passing small watery stools
 - Failure to thrive (delayed growth)
- Complication → **Enterocolitis**
 - Occurs in 30-40% of individuals affected by Hirschsprung's
 - Vomiting, diarrhea, signs of toxic megacolon → pain, fever, bloody diarrhea, rapid heart rate
 - Life-threatening serious intestinal infection → needs immediate treatment with antibiotics and bowel washouts



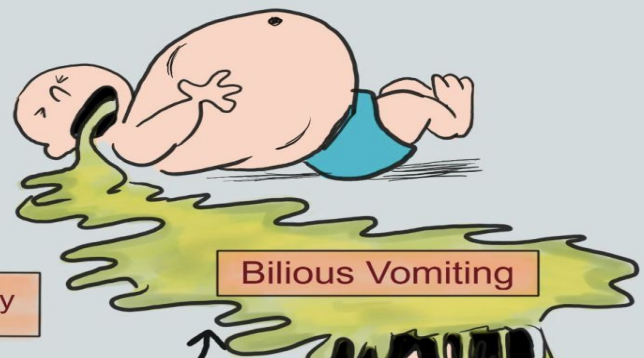
My belly Hurts...

Thank god Feeding you isn't a big problem....

But you are Odd with huge Abdomen...!!



No feeding Abnormality



Bilious Vomiting

Severe Abdominal Distention

Hirschsprung Disease Clinical Features

Creative-Med-Doses

Failure to pass Meconium



I wish something had come out of your Aboral End...!!
Enough vomiting...

Hope you have all things in ...
Right place...



You drank milk.. you vomited bile....

But you haven't Pooped
What's wrong with your Defecation system...

Any baby who hasn't passed *Meconium* within 24 hours of birth should be investigated for *Hirschsprung Disease*
It may save life...

Diagnosis

- **Rectal suction biopsy – definitive diagnosis**
 - Absence of ganglion cells in the sample tissue under microscopy
- Abdominal X-Ray – shows intestinal obstruction
- Contrast enema – transition zone between normal and affected bowel
- Anorectal manometry - balloons and pressure sensors to measure function of rectal nerves
 - Increased anal sphincter pressure and lack of relaxation of the internal sphincter with balloon rectal distension



Abdominal X-Ray with multiple loops of dilated small bowel



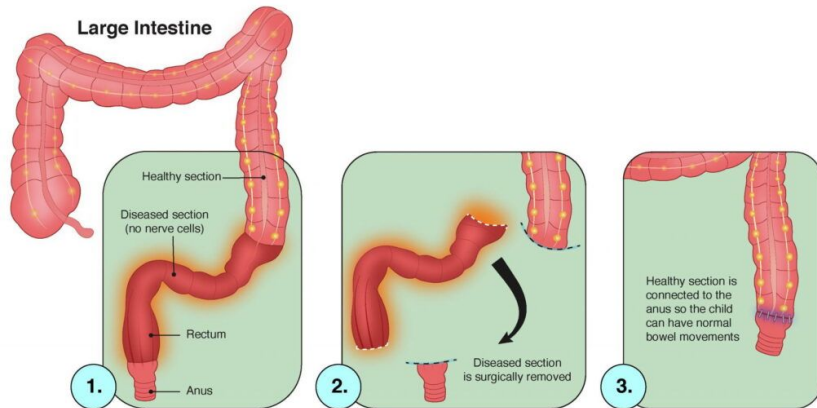
Contrast Enema with visible Transition Zone

Management → Surgical resection of the affected bowel segment

Pull Through Surgery

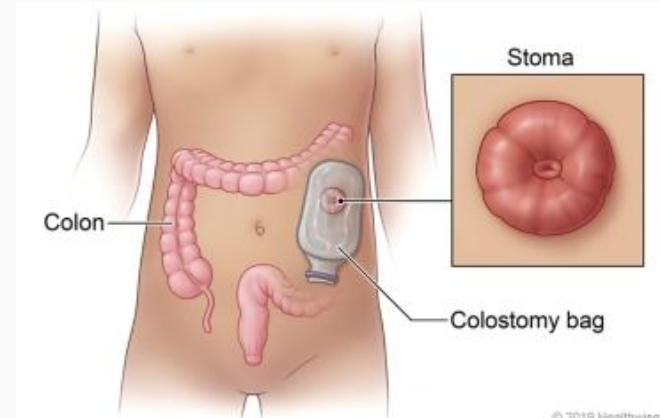
Diseased section is removed and healthy normal section is pulled through the colon from the inside and attached to the anus

Pull-Through Procedure



Ostomy Surgery


1. Diseased section removed and healthy portion connected to an opening created in the abdomen. Stool leaves through the opening into a bag attached to the end of the intestine that protrudes through the hole. Allows time for the lower part of the colon to heal.
2. Stoma is closed and healthy portion of intestines is attached to the rectum or anus



Awareness



SEPTEMBER



Hirschsprung's
Disease Awareness

MONTH

We may be RARE
but we've got ROAR

Ostomy gastrostomy major surgery leaks
bleeding catheters
bile medication
pain hospital
washouts short gut
perforated biopsy
worry complications
surgeon injections
Tubes ??

"BUT YOU DON'T
LOOK SICK..."

do it for the kids
SPREAD THE HIRSCHSPRUNG'S WORD



- 1 out of 5000 live births
- Accounts for 20% of neonatal bowel obstructions

WE WEAR BURGUNDY FOR



HIRSCHSPRUNG'S DISEASE AWARENESS

Resources

- Pance Prep Pearls - Dwayne A. Williams
- [https://www.mayoclinic.org/diseases-conditions/hirschsprungs-disease/symptoms-causes/syc-20351556#:~:text=Hirschsprung's%20\(HIRSH%2Dsproongz\)%20disease,muscles%20of%20the%20baby's%20colon.](https://www.mayoclinic.org/diseases-conditions/hirschsprungs-disease/symptoms-causes/syc-20351556#:~:text=Hirschsprung's%20(HIRSH%2Dsproongz)%20disease,muscles%20of%20the%20baby's%20colon.)
- <https://www.childrenshospital.org/conditions/hirschsprungs-disease>
- <https://www.mountsinai.org/health-library/diseases-conditions/toxic-megacolon#:~:text=Toxic%20megacolon%20is%20characterized%20by,that%20requires%20immediate%20medical%20treatment.>
- <https://www.rch.org.au/uploadedFiles/Main/Content/cprs/CPRS%20Booklet%20-%20Hirschsprung%20Asociated%20Enterocolitis.pdf>
- <https://rarediseases.org/rare-diseases/hirschsprungs-disease/>
- <https://www.health.state.mn.us/diseases/cy/hirschsprung.html#:~:text=Hirschsprung%20disease%20occurs%20in%201,rate%20of%2020%2D25%25.>