

HIRSCHSPRUNG'S DISEASE

(Congenital Megacolon)

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DEFINITION:

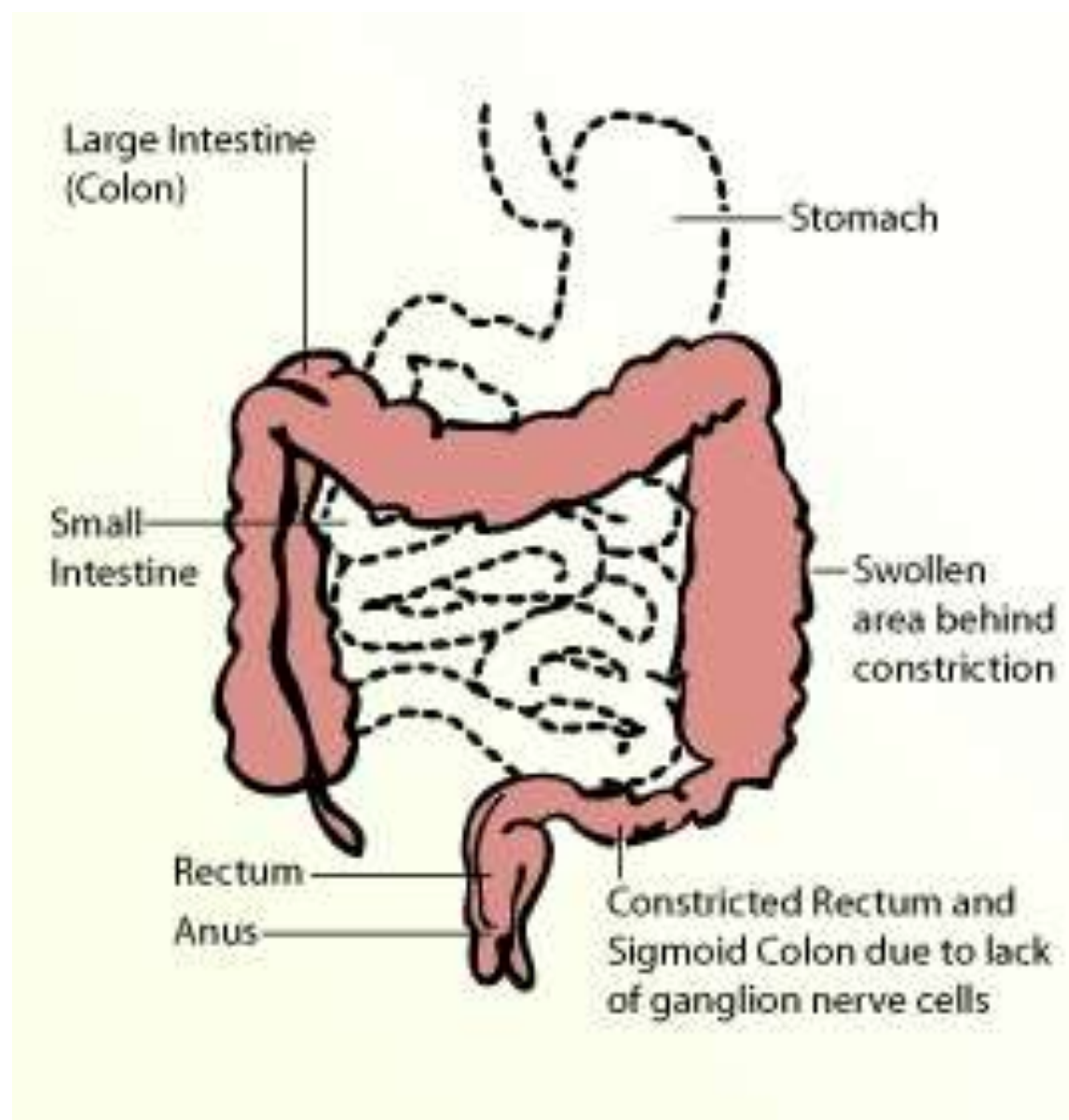
Obstruction caused by lack of propagation of peristaltic wave due to absence of ganglionic cells in myenteric and submucosal plexus which leads to spasm of aganglionic bowel i.e. failure to relax.

Types

- 1. Short segment (rectosigmoid) 75% have ganglionic cells down to the level of rectosigmoid and below that is aganglionic gut.
- 2. Long segment-15% have aganglionic cells up to descending colon or transverse colon.
- 3. Total colonic aganglionosis- 5-7% have involvement of descending colon and even distal small bowel.

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Embryogenesis

- Cellular and molecular abnormalities in the development of enteric nervous system along with incomplete migration of neural crest cells.
- Neural crest derived “neuroblasts” cells migrate in cranial to caudal direction into developing gut from 5th to 12th week of gestation. But in Hirschsprung’s disease, there is failure of migration of these cells and segment involved will depend at what stage arrest occurs.

Clinical Features

Incidence – 1 to 3 per 5000 live births.

Sex ratio- male to female 4:1

1. History of delayed passage of meconium i.e. failure to pass meconium within 48hrs of birth.
2. Constipation and intermittent diarrhea.
3. Distended abdomen and bilious vomitings in later stages.
4. Feeding intolerance with poor weight gain.
5. Acute presentation as enterocolitis that is passage of foul smelling stools with abdominal distension and fever. Patient can go into sepsis and shock and even death can occur. Patient needs an immediate treatment.
6. P/R-Collapsed rectum and patient passes paste like stools after withdrawal of finger.

Older children > 1yr of age

1. Signs and symptoms of constipation and enterocolitis
2. Protuberant abdomen with visible peristalsis.
3. Peripheral edema and hypoproteinemia
4. Extremity wasting

Diagnosis

- Plain X ray abdomen- Gaseous distension of the colon with few air fluid levels and absent rectal gas.
- Barium Enema study:
 1. Contrast study should be done on unprepared colon, under fluoroscopy, non balloon catheter should be used and give small amount of barium just to delineate the dilated colon and take a delayed 24H film.
 2. Findings- There is irregular colonic contraction in distal collapsed aganglionic colon with distension of proximal dilated colon and transition zone intact.
 3. Retention of barium on delayed 24 hr film is also suggestive of Hirschsprung's disease.

Plain X ray



Barium enema



- **Anorectal manometry**- used in older children
- **Normal reflex** - distension of the rectum (increased rectal pressure) leads to reduction in internal sphincteric pressure i.e. relaxation of the anorectum.
- But in Hirschsprung's disease this reflex is absent i.e. when rectum distends internal sphincter fails to relax.

Confirmatory Diagnosis

- Rectal biopsy –open full thickness
 - suction biopsy
- Full thickness biopsy is taken 1-2cm above the dentate line from posterior rectal wall and looked for ganglionic cells. In Hirschsprung's disease, there is absence of ganglionic cells along with hypertrophied nerve bundles.
- Suction biopsy done in small children and neonates and stained with acetylcholinesterase staining and if there is increased activity of acetylcholine esterase, this is also confirmatory of Hirschsprung's disease.

Management

- Basic principle is the resection of the aganglionic portion of the bowel and bringing the ganglionic bowel for coloanal anastomosis.
- Operative procedure is usually done as staged procedures.

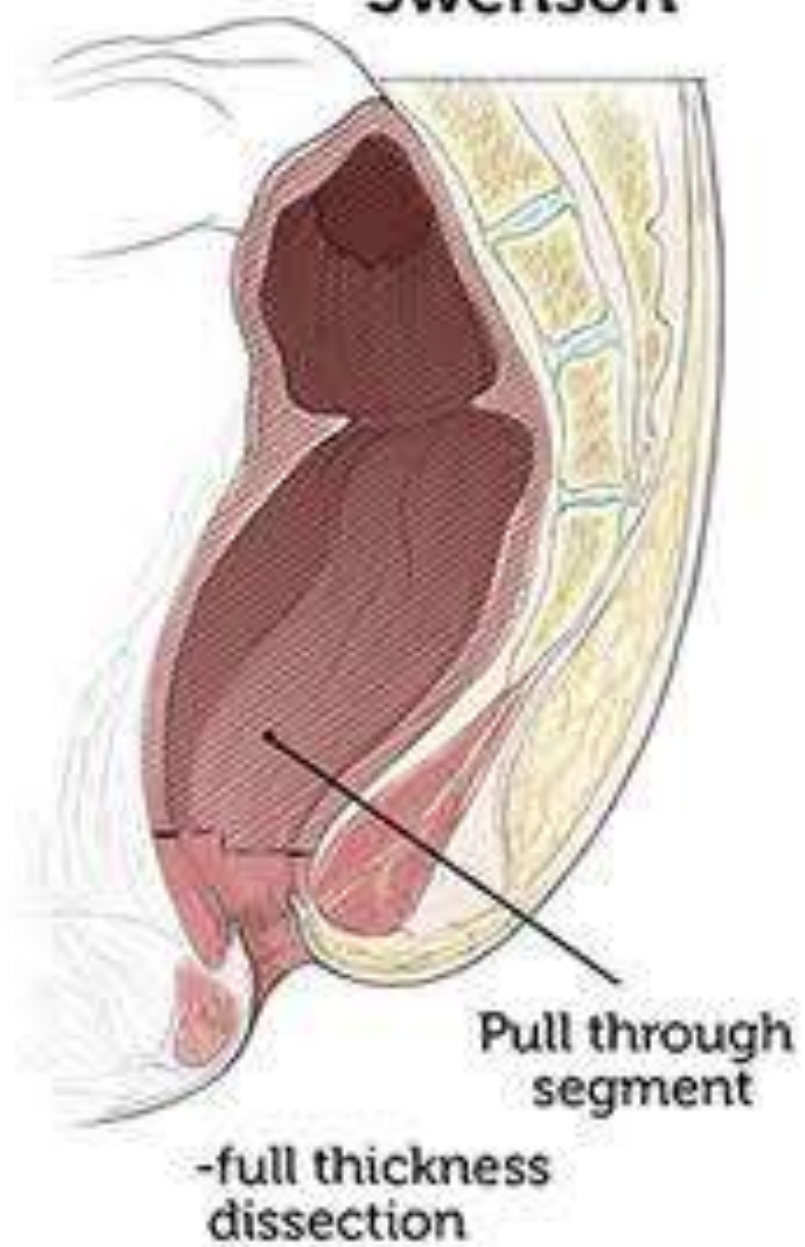
Two stage - 1st stage, diverting colostomy is done proximal to the transition zone with resection of the aganglionic bowel and in the second stage colostomy is pulled down for coloanal anastomosis.

Three stage- 1st diverting colostomy, 2nd resection of the aganglionic bowel with coloanal anastomosis and 3rd colostomy closure.

Various operations done are

- Swenson's Procedure
- Soave Boley Procedure
- Duhamel's Procedure

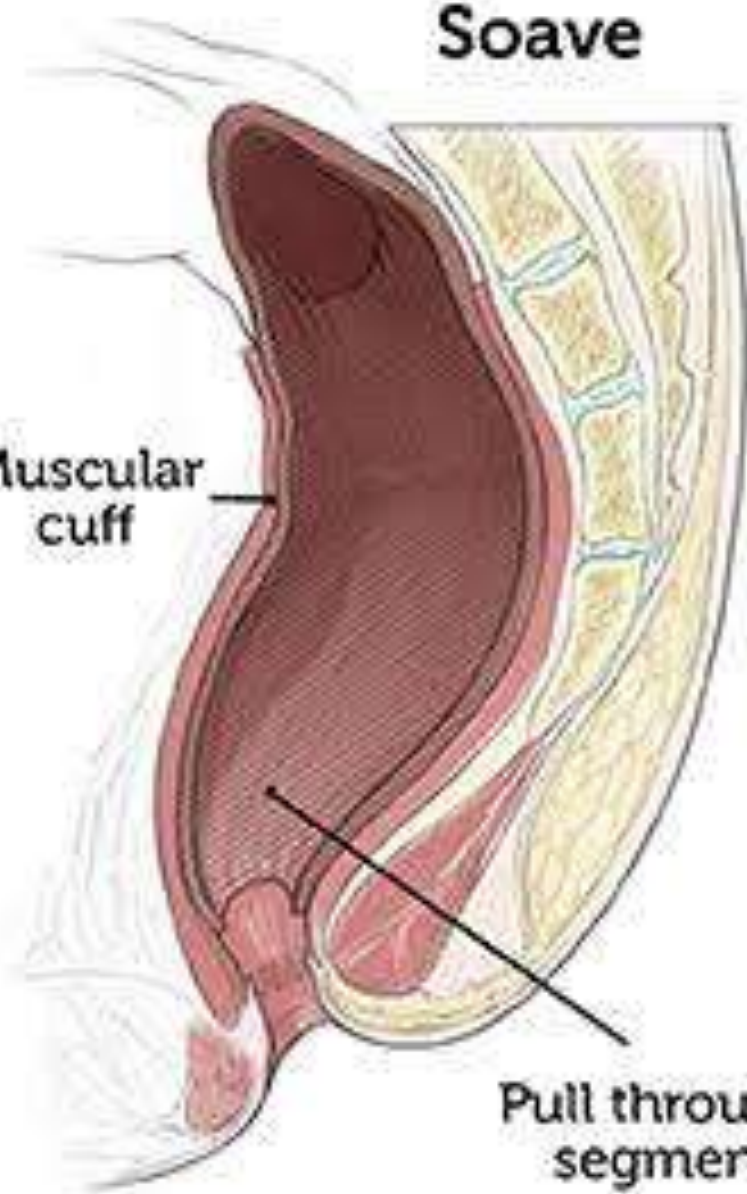
Swenson



Soave

Muscular
cuff

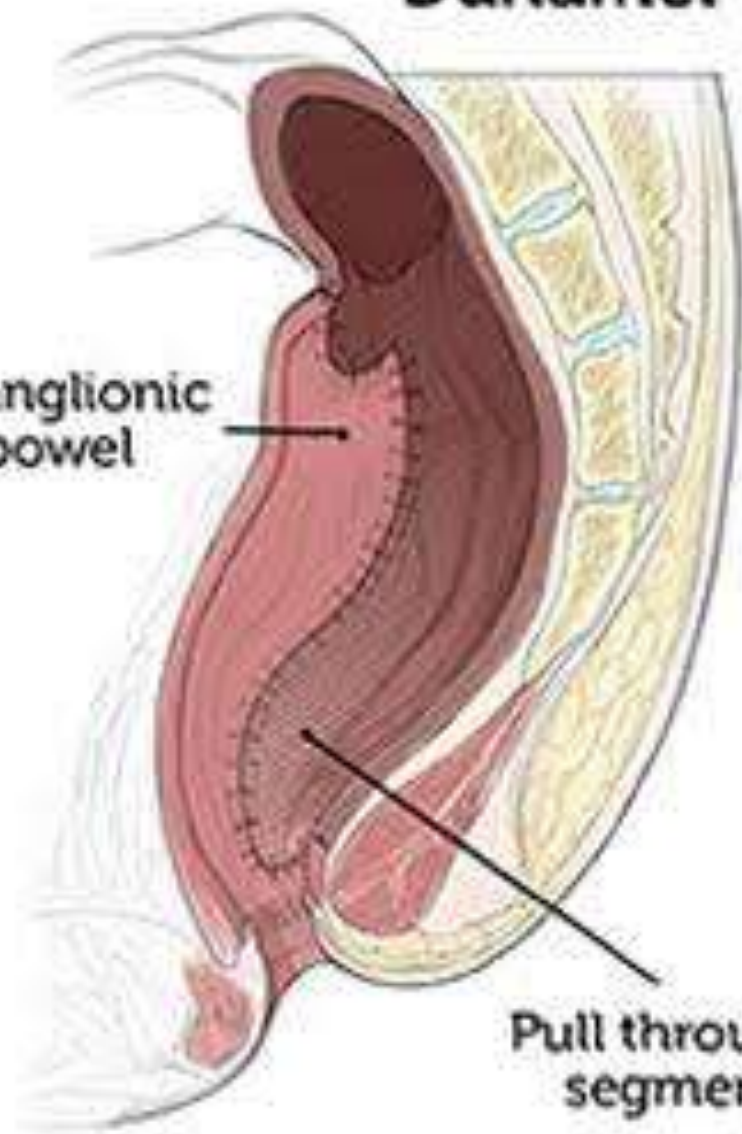
Pull through
segment



Duhamel

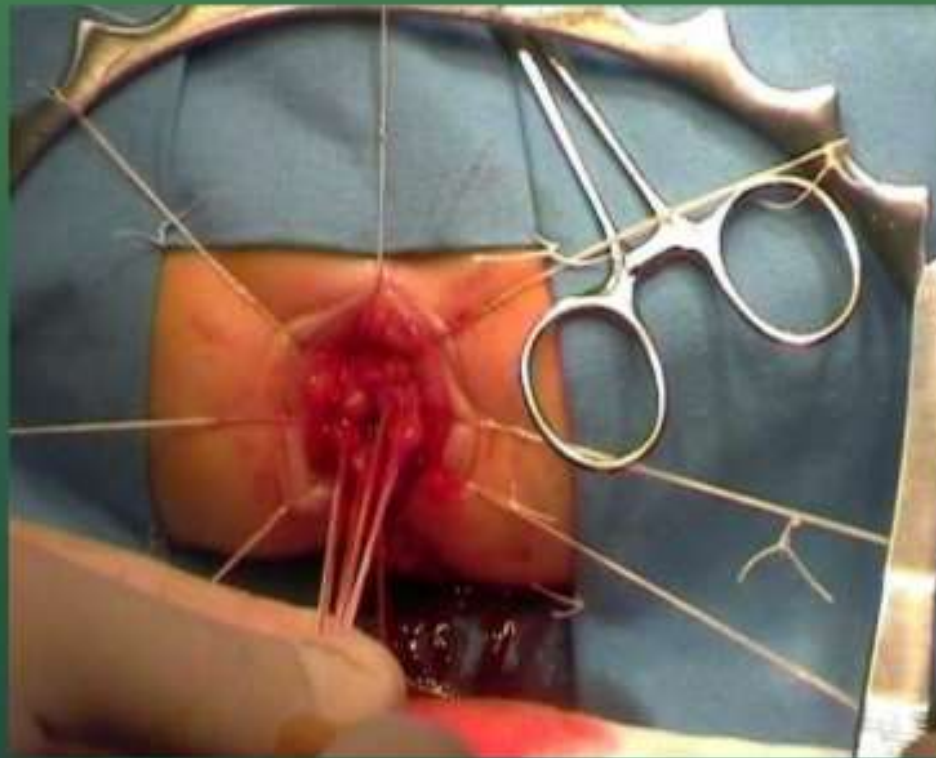
Aganglionic
bowel

Pull through
segment



- Single stage- Transanal endorectal pull through procedure is done these days in patients with minimal dilatation of the colon.
- Dissection and mucosectomy is done from below and then ganglionic bowel is pulled for coloanal anastomosis.

Transanal Endorectal Pull-Trough



THANKS