

HIRSCHSPRUNG DISEASE

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Variant of
Hirschsprung

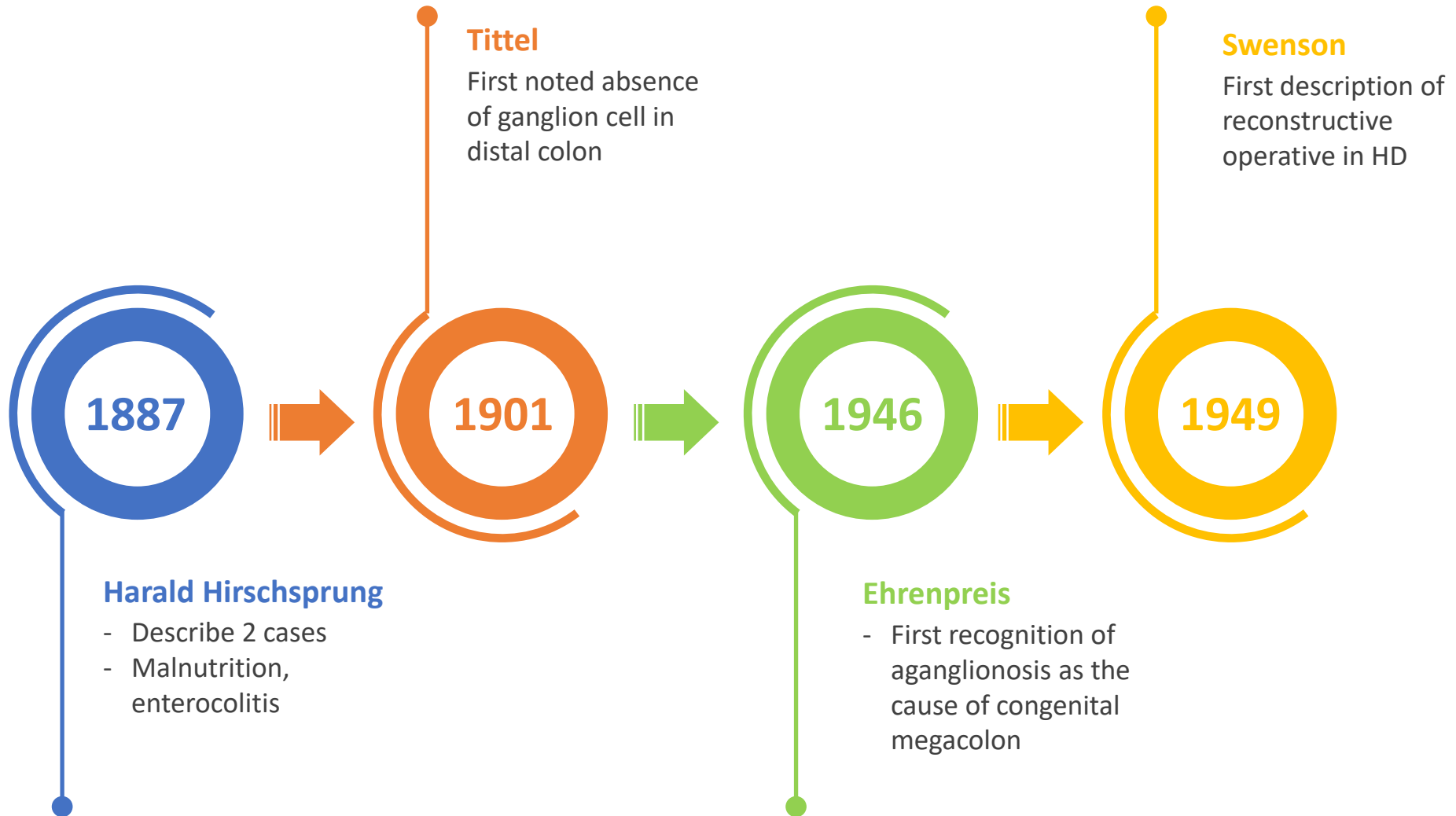


INTRODUCTION & HISTORY

INTRODUCTION

- Also known as “Congenital megacolon”
- Absence of ganglion cell in myenteric and submucosal plexus

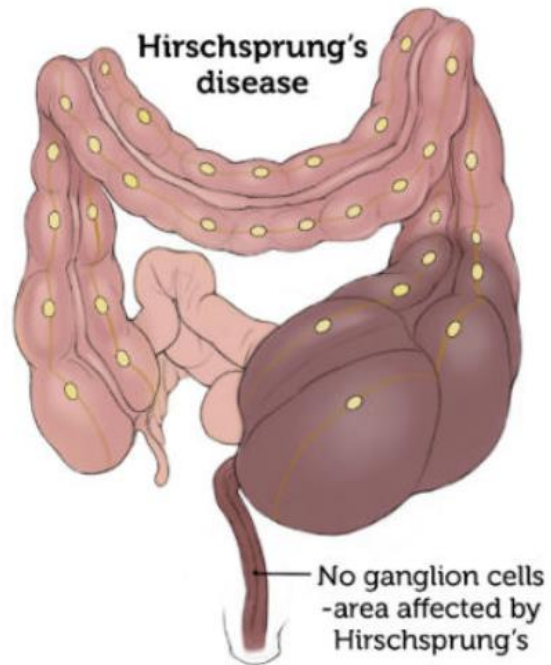
HISTORY



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EPIDEMIOLOGY

INCIDENCE



- 1 in 5000 live birth
- 80% seen TZ in rectum or rectosigmoid
- 5-10% have total colonic aganglionosis

ASSOCIATED SYNDROME

Box 34.1 Congenital Anomalies and Conditions Commonly Associated With Hirschsprung Disease

- Down syndrome (trisomy 21)
- Neurocristopathy syndromes
 - Waardenberg–Shah syndrome
 - Yemenite deaf-blind-hypopigmentation
 - Piebaldism
 - Other hypopigmentation syndromes
- Goldberg–Shprintzen syndrome
- Smith–Lemli–Opitz syndrome
- Multiple endocrine neoplasia 2
- Congenital central hypoventilation syndrome (Ondine’s curse)
- Isolated congenital anomalies
 - Congenital heart disease
 - Malrotation
 - Urinary tract anomalies
 - Central nervous system anomalies
 - Other



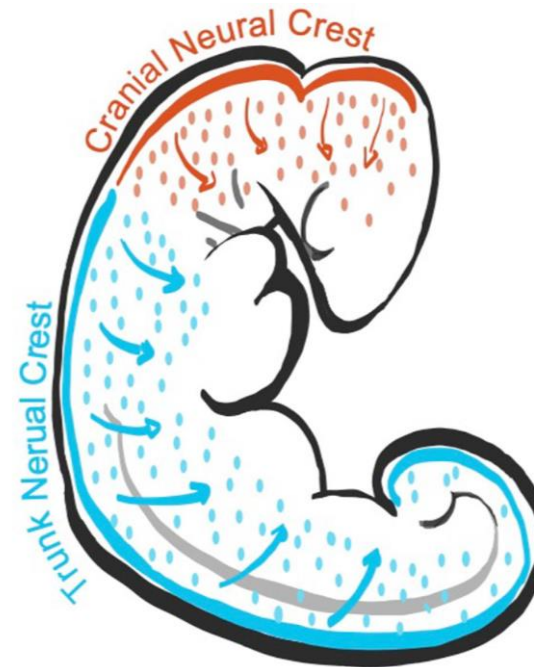
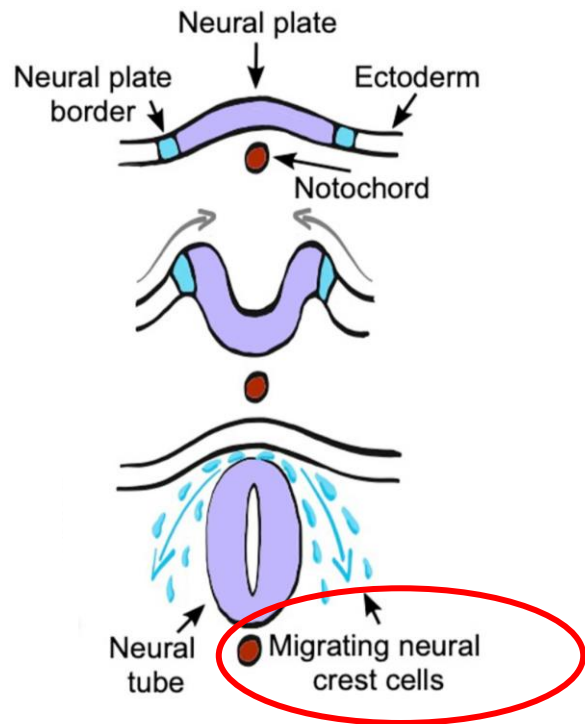
ETIOLOGY

ETIOLOGY

Neural crest

13wks postconception

ganglion cell



Proximal

Distal

THEORY

1. Neural crest cell never reach the distal intestine
 - Due to early maturation or differentiation into ganglion cell
2. Neural crest cell reach their destination but fail to survive or differentiate
 - Due to an inhospitable microenvironment

GENETIC BASIS OF DISEASE

RET proto-oncogene

- Encodes tyrosine kinase receptor
- Found in patients with familial and long-segment involvement
- Related with Neurturin, Glial cell line-derived neurotrophic factor(GDNF)
- Mechanism : Assoc with early neuronal cell death

Other

- Endothelin family genes esp endothelin-3 endothelin-B receptor
- SOX-10 gene
- ZFHX1B, Phox2B, HedgehogNotch complex

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CLINICAL PRESENTATION & DIAGNOSIS

CLINICAL PRESENTATION

Prenatal

- Rare
- Mostly U/S find fetal intestinal obstruction in total colonic disease

Neonatal period

**Mostly present at this time

- Abdominal distension
- Bilious vomiting
- Feeding intolerance
- Delayed passage meconium beyond 24 hr (90%)

CLINICAL PRESENTATION

Later childhood

- Chronic constipation
- Failure to thrive
- Gross abdominal distention
- Dependence on enema without significant encopresis

CLINICAL PRESENTATION



- 10 % of HD present with Hirschsprung-associated enterocolitis (HAEC)
 - Fever
 - Abdominal distension
 - Diarrhea
 - Toxic megacolon
 - Perforation
- May confuse due to HD mostly present with constipation

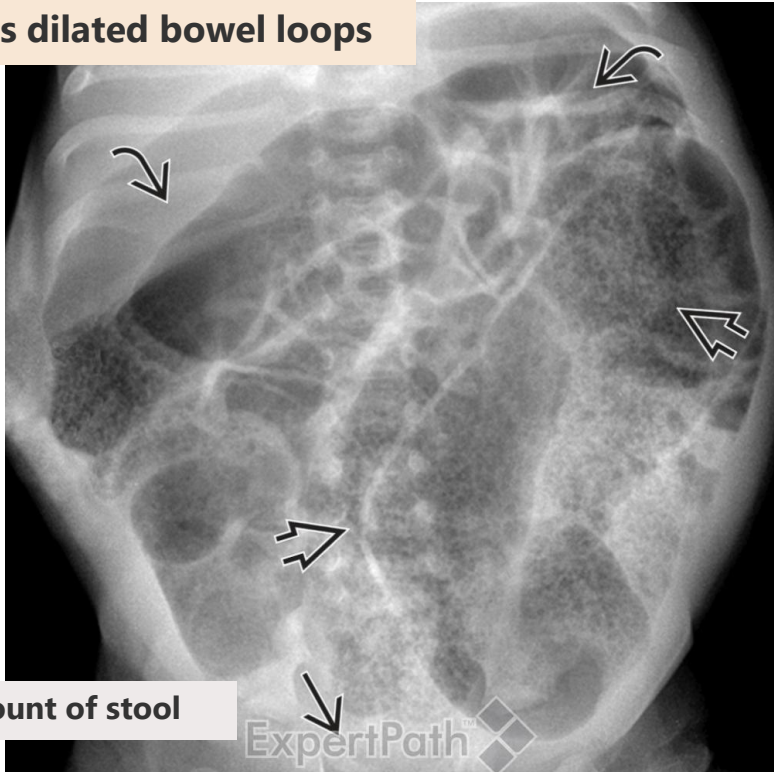
DIFFERENTIAL DIAGNOSIS

- Intestinal atresia
- Meconium ileus
- Meconium plug syndrome

INVESTIGATION

Plain radiography

Numerous dilated bowel loops

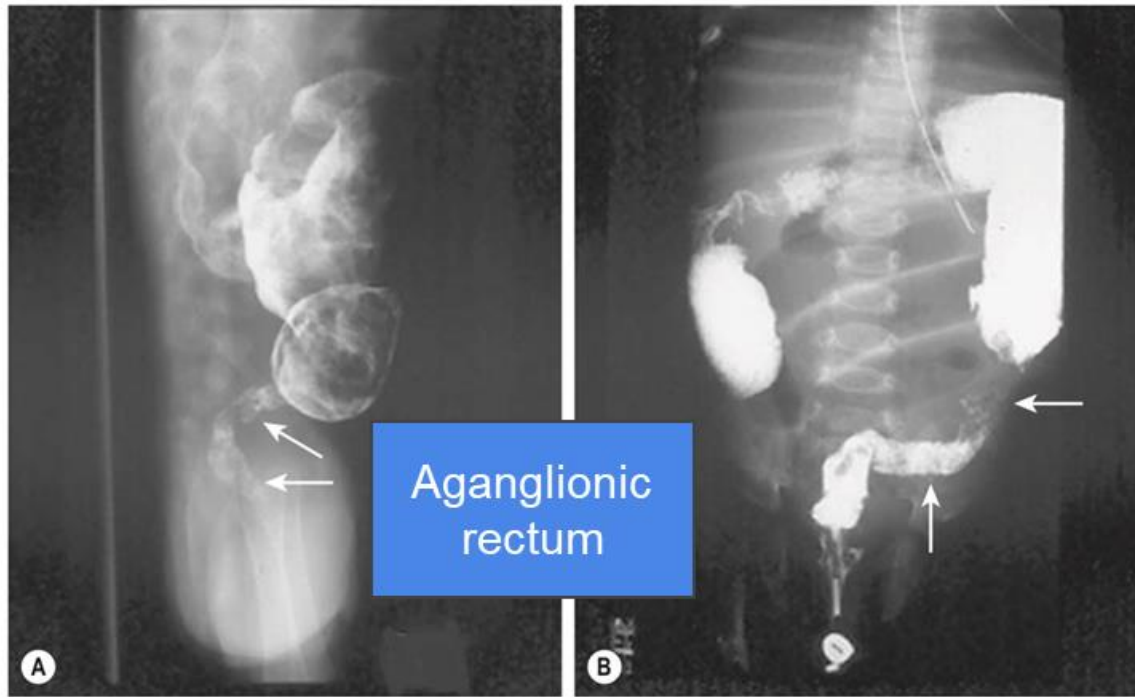


large amount of stool

- Dilated bowel loop throughout abdomen

INVESTIGATION

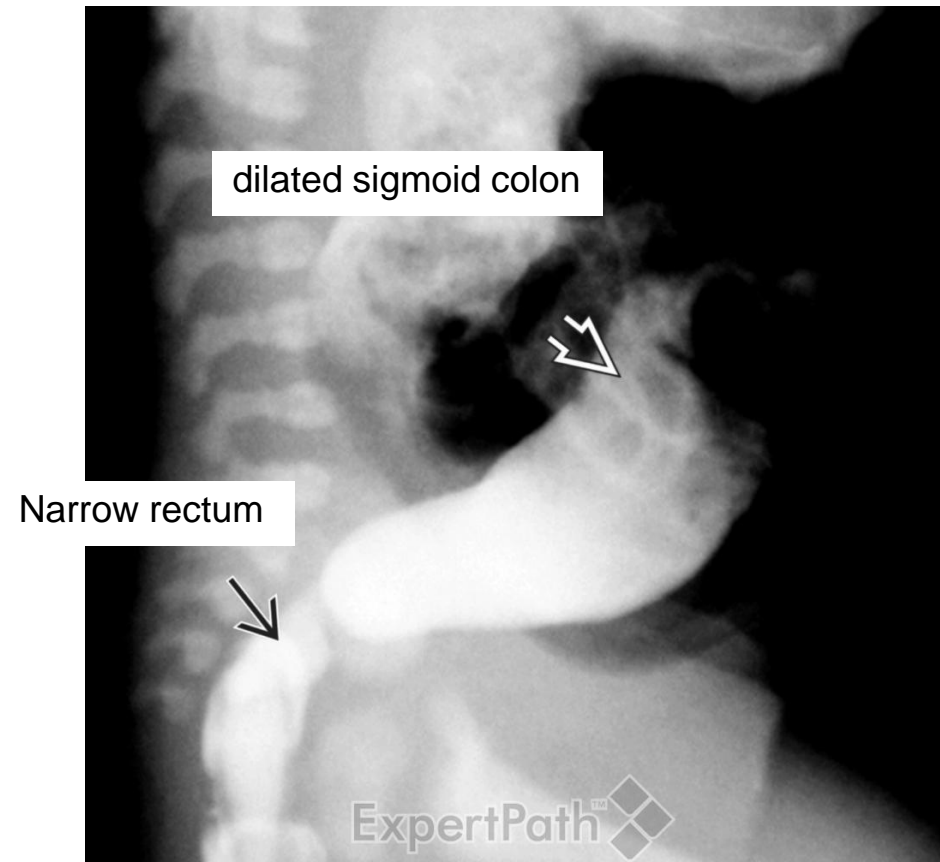
Water-soluble contrast enema



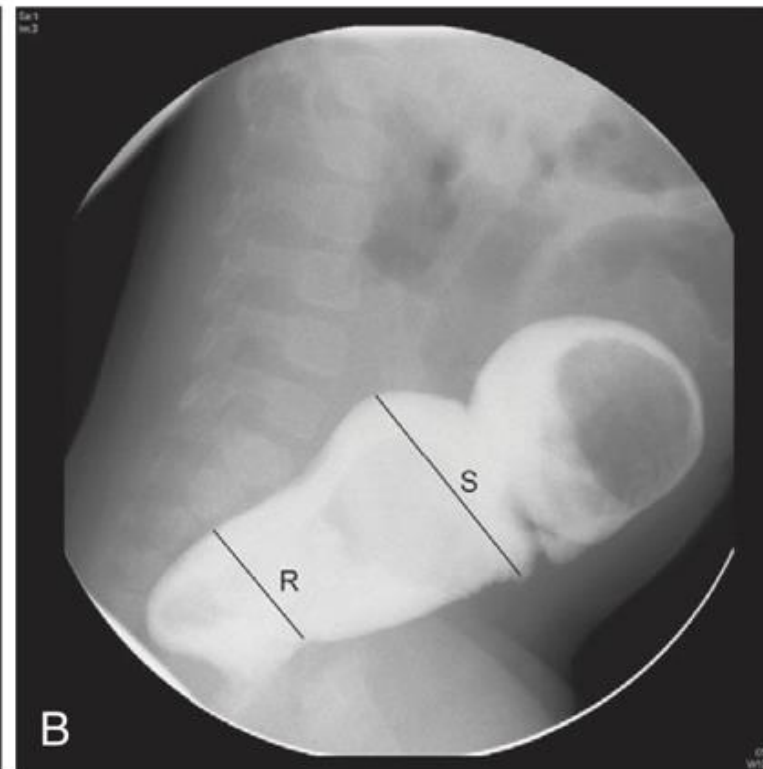
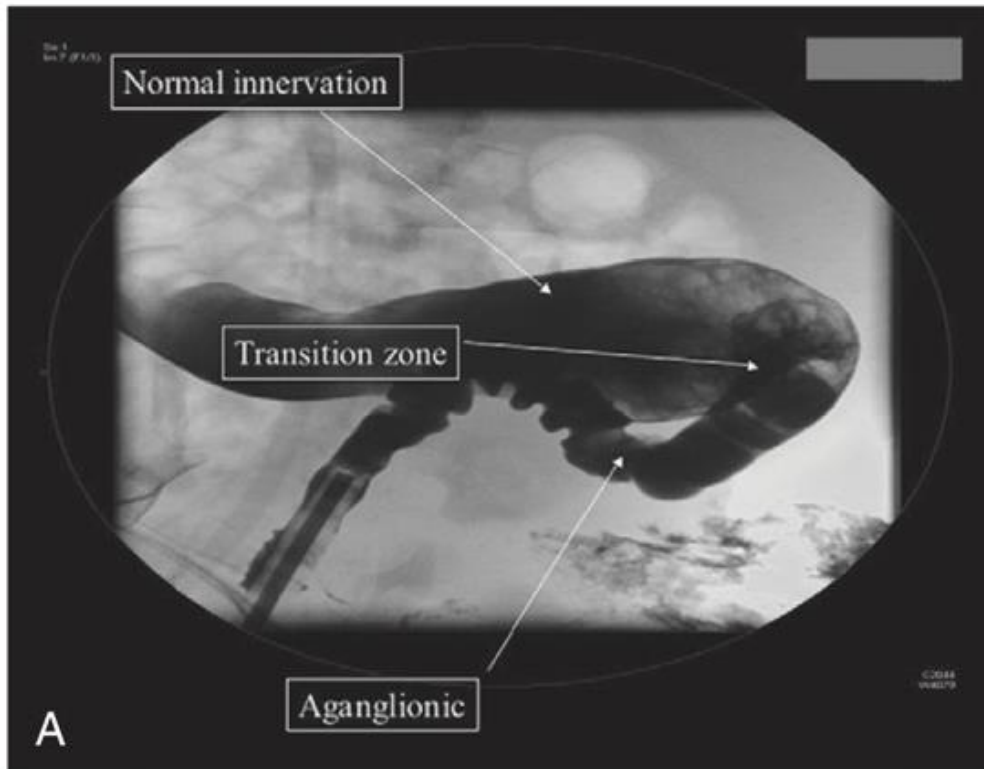
- Transitional zone between normal and aganglionic bowel
- Best seen in lateral view
- 10% may not be seen

- Water enema can be definitive treatment in other conditions
 - Meconium ileus
 - Meconium plug syndrome

INVESTIGATION : WATER SOLUBLE ENEMA



INVESTIGATION : WATER SOLUBLE ENEMA



$$R : S < 1$$

INVESTIGATION : WATER SOLUBLE ENEMA



FIGURE 101-7 Contrast enema in a child with total colonic Hirschsprung disease. There is no transition zone in the colon, and the colon is foreshortened with a "question-mark" configuration.

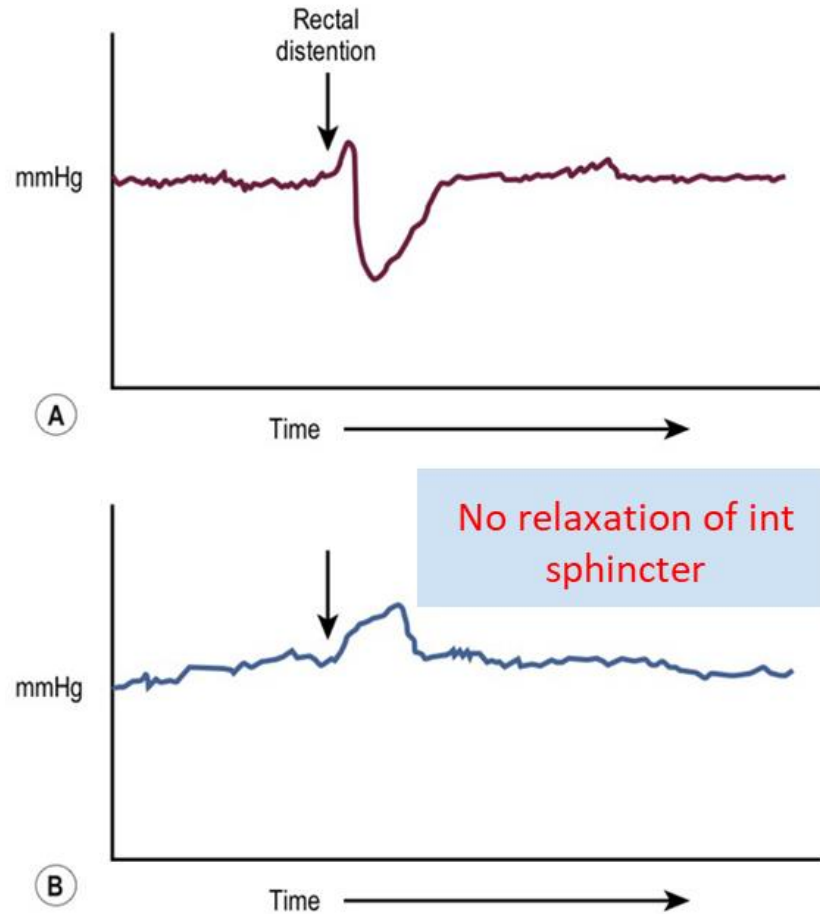
INVESTIGATION



Fig. 34.2 Retention of contrast is seen on this postevacuation film, which was obtained 24 hours after the contrast enema.

- Obtain plain radiography 24 hr later
 - Retention of contrast very suggest HD
- False negative
 - Massive rectal distension
 - Very short aganglionic segment

INVESTIGATION

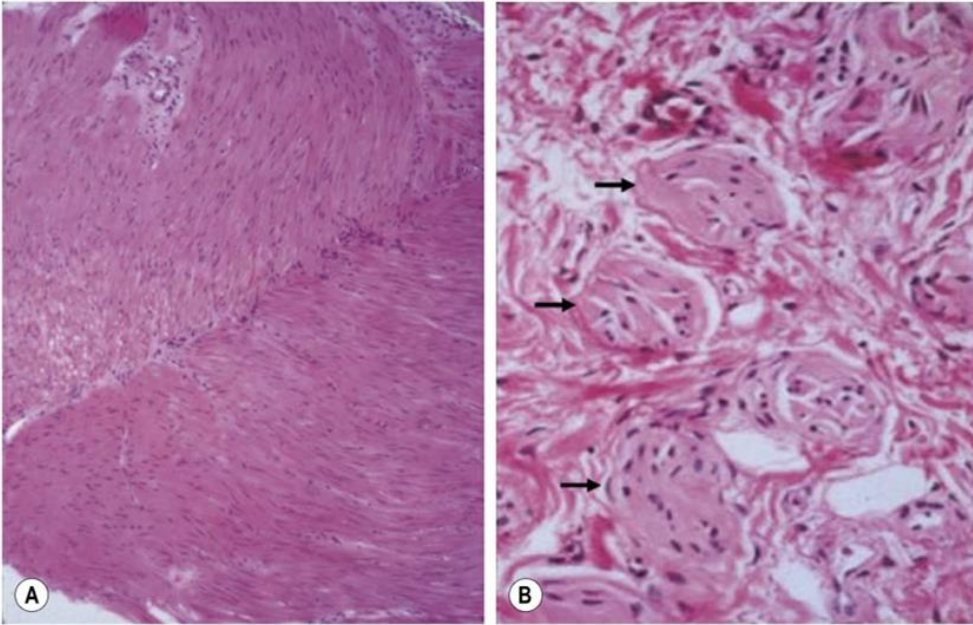


Anorectal manometry

- Presence of recto-anal inhibitory reflex(RAIR)
- Reflex relaxation of int-anal sphincter response to rectal distension
- Useful in older child with constipation
- False positive
 - Contraction of external anal sphincter
 - Artifact from moving or crying

Normal RAIR -> exclude HD

RECTAL BIOPSY



Absence of ganglion cell in submucosal and myenteric plexus

Hyperthrophic nerve trunk

- **Gold standard** diagnostic technique
 - Suction rectal biopsy
 - Less reliable in older children (4 year)
 - sampling error
 - Immaturity of ganglion cell
 - > 1-1.5 cm above dentate line
 - Full thickness biopsy

HISTOLOGY

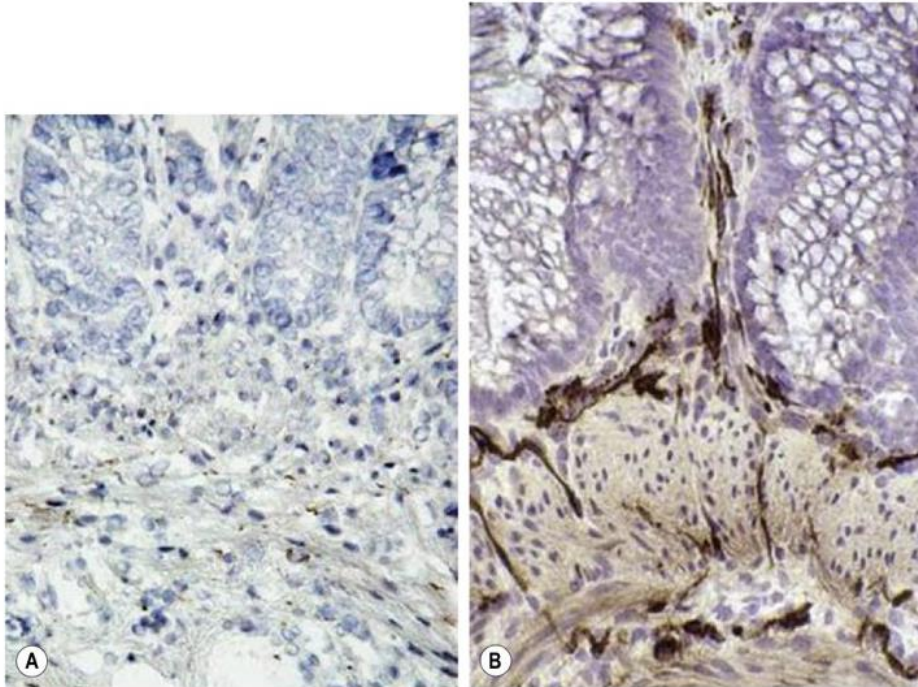


Fig. 34.5 Cholinesterase staining in (A) normal colon and (B) colon affected by Hirschsprung disease.

- Hematoxylin and eosin stain for **acetylcholinesterase**
- Characteristic pattern in submucosa and mucosa of HD

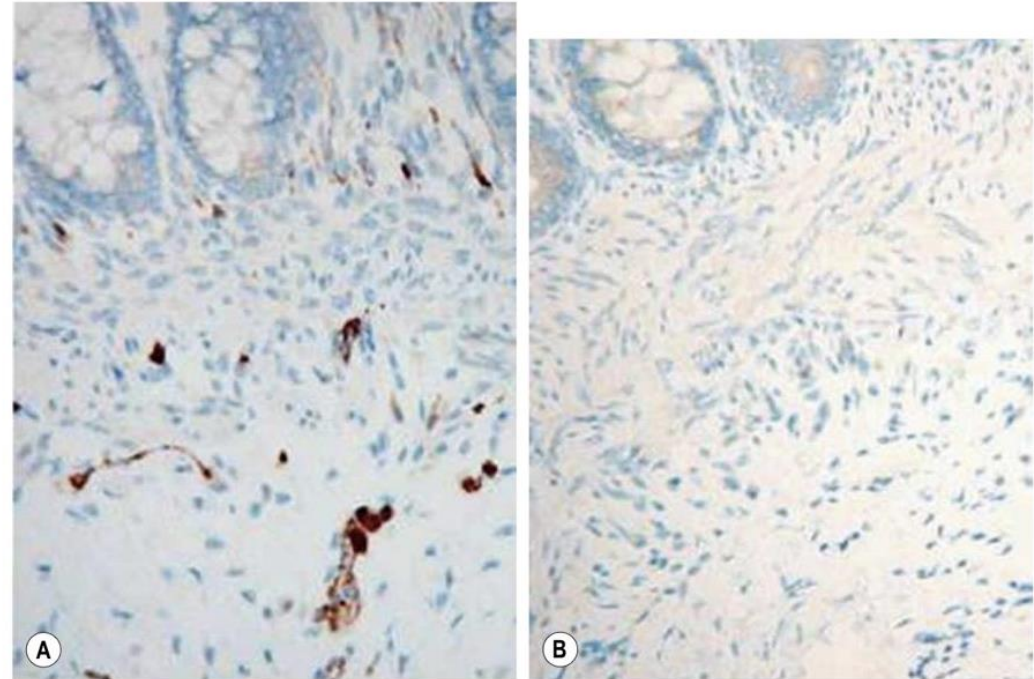
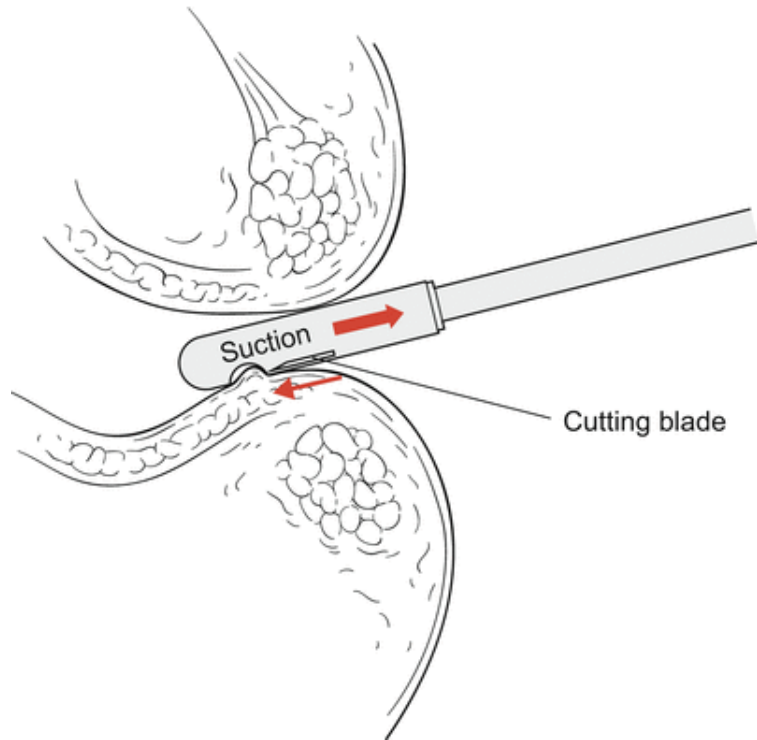


Fig. 34.6 Calretinin staining is seen in (A) normal colon but not in colon (B) affected by Hirschsprung disease.

- Calretinin – always absent in HD

ADEQUATE TISSUE BIOPSY



- Normal paucity of ganglion cell at 0.5 -1.0 cm above dentate line
- Biopsy should be taken $> 1.0-1.5$ cm
- Biopsy too proximal may miss short aganglion segment

ALGORITHM FOR DIAGNOSIS HIRSCHSPRUNG DISEASE

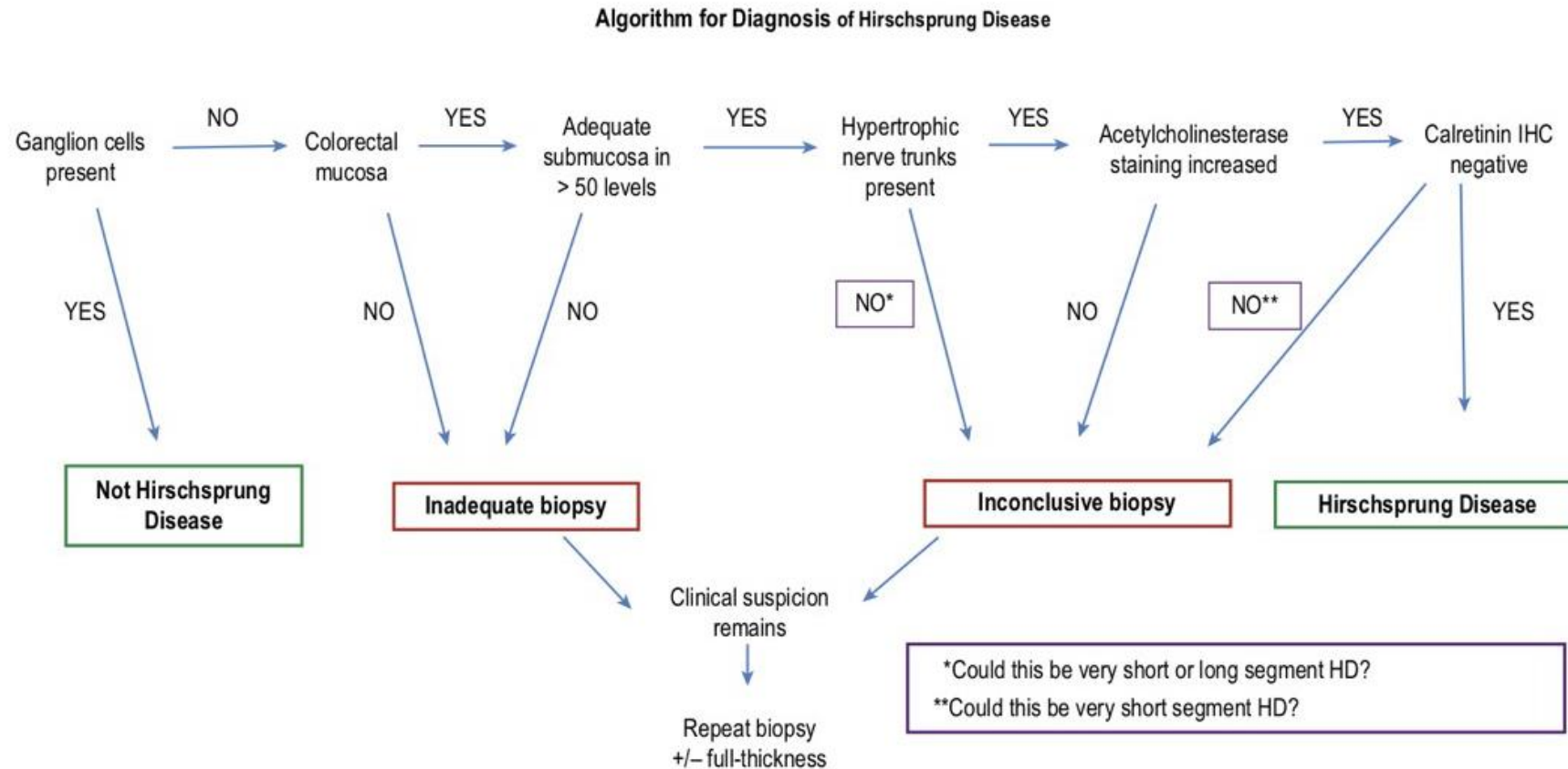


Fig. 34.7 Algorithm for the pathologic diagnosis of Hirschsprung disease from a rectal biopsy. (Courtesy Dr. Gino Somers and Dr. Glenn Taylor, Department of Pathology, Hospital for Sick Children, Toronto.)



MANAGEMENT

PREOPERATIVE PREPARATION

Initial management

- Resuscitated with intravenous fluid
- Broad-spectrum antibiotics
- Nasogastric drainage
- Rectal decompression using rectal stimulation or irrigation
- Investigate associate abnormalities before surgery

Semi-electively operation

- Discharge
- Feeding with breast milk or formula
- Combination with rectal irrigation
- Advice HAEC

PREOPERATIVE PREPARATION

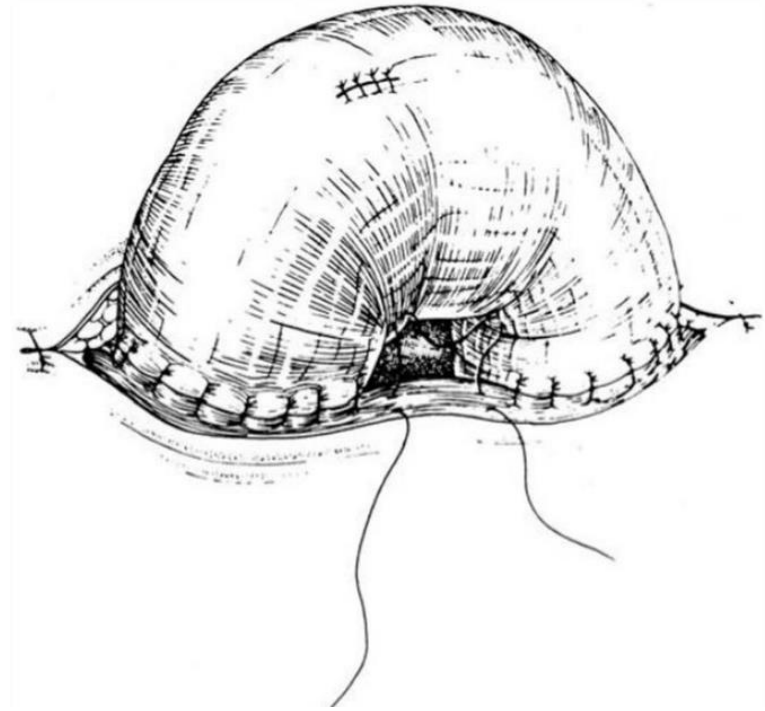
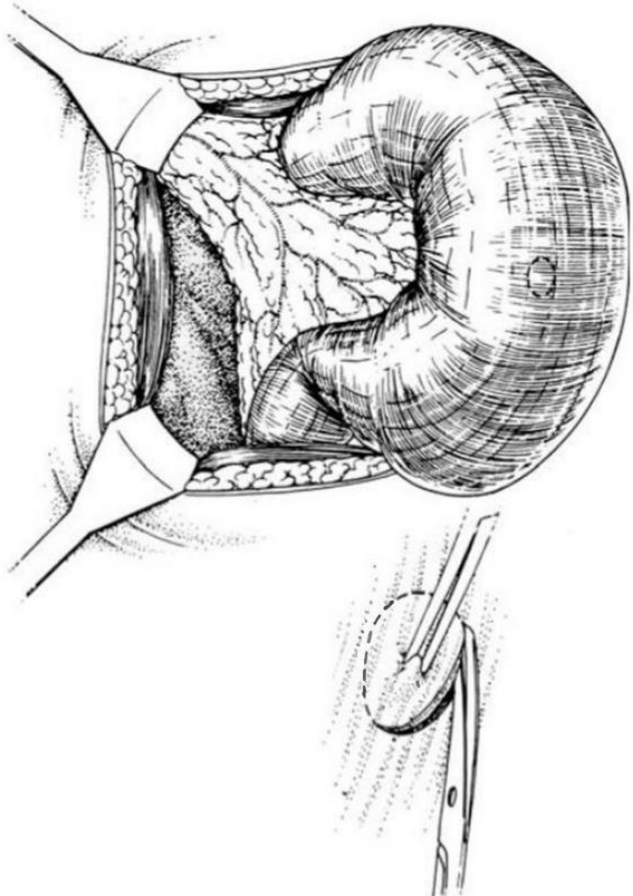
- Older child with extremely dilate colon
 - Pull-through should be delayed until diameter of colon has decreased sufficiently
 - Weeks to months may be need
 - Colostomy sometime gain benefit



INDICATION FOR COLOSTOMY

- Severe enterocolitis
- Perforation
- Malnutrition
- Massively dilated proximal bowel
- Not possible to reliably identify TZ on frozen section

COLOSTOMY



SURGICAL MANAGEMENT

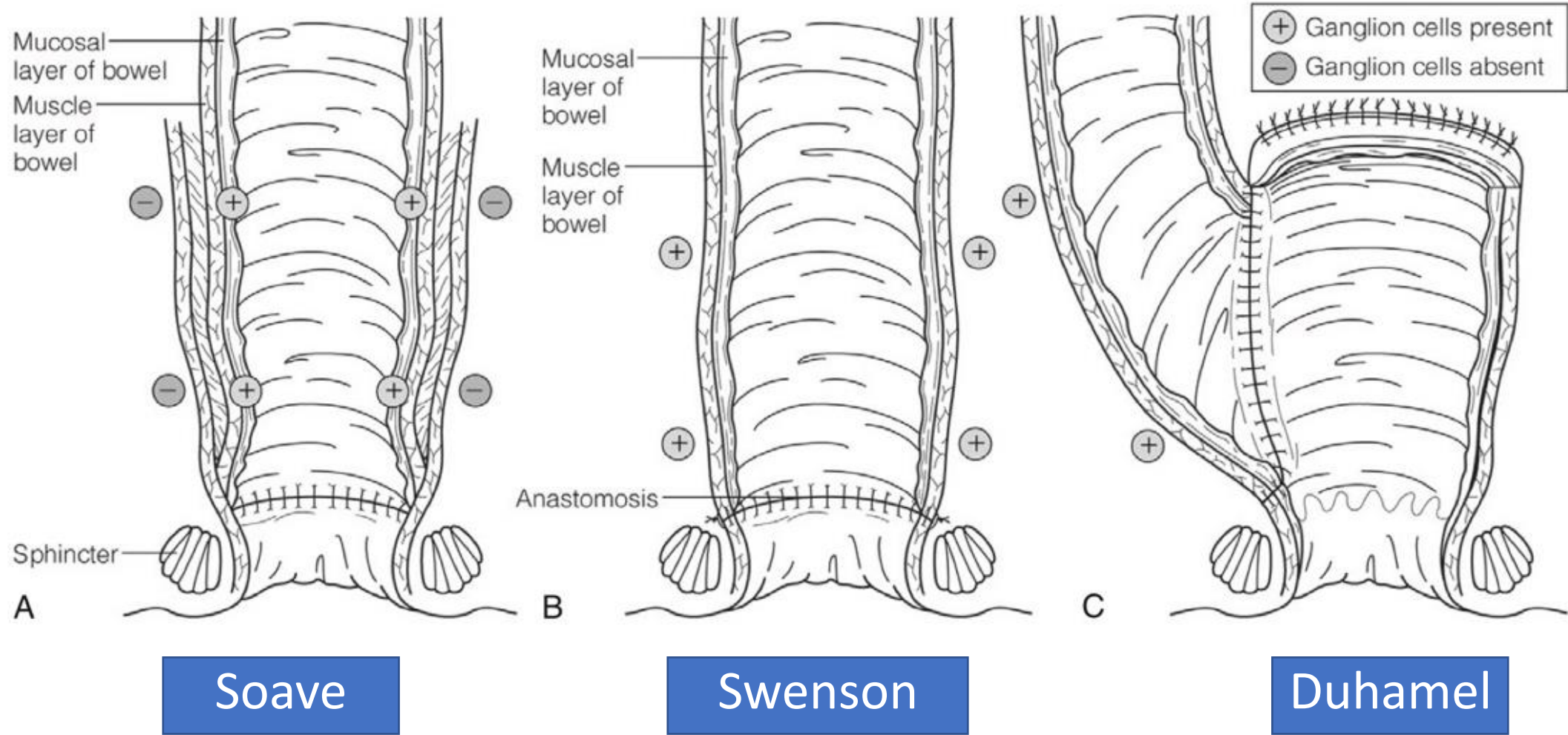
- Aim

- Remove aganglionic bowel and reconstruct intestinal tract bring normal bowel to anus
- Preserve normal sphincter function

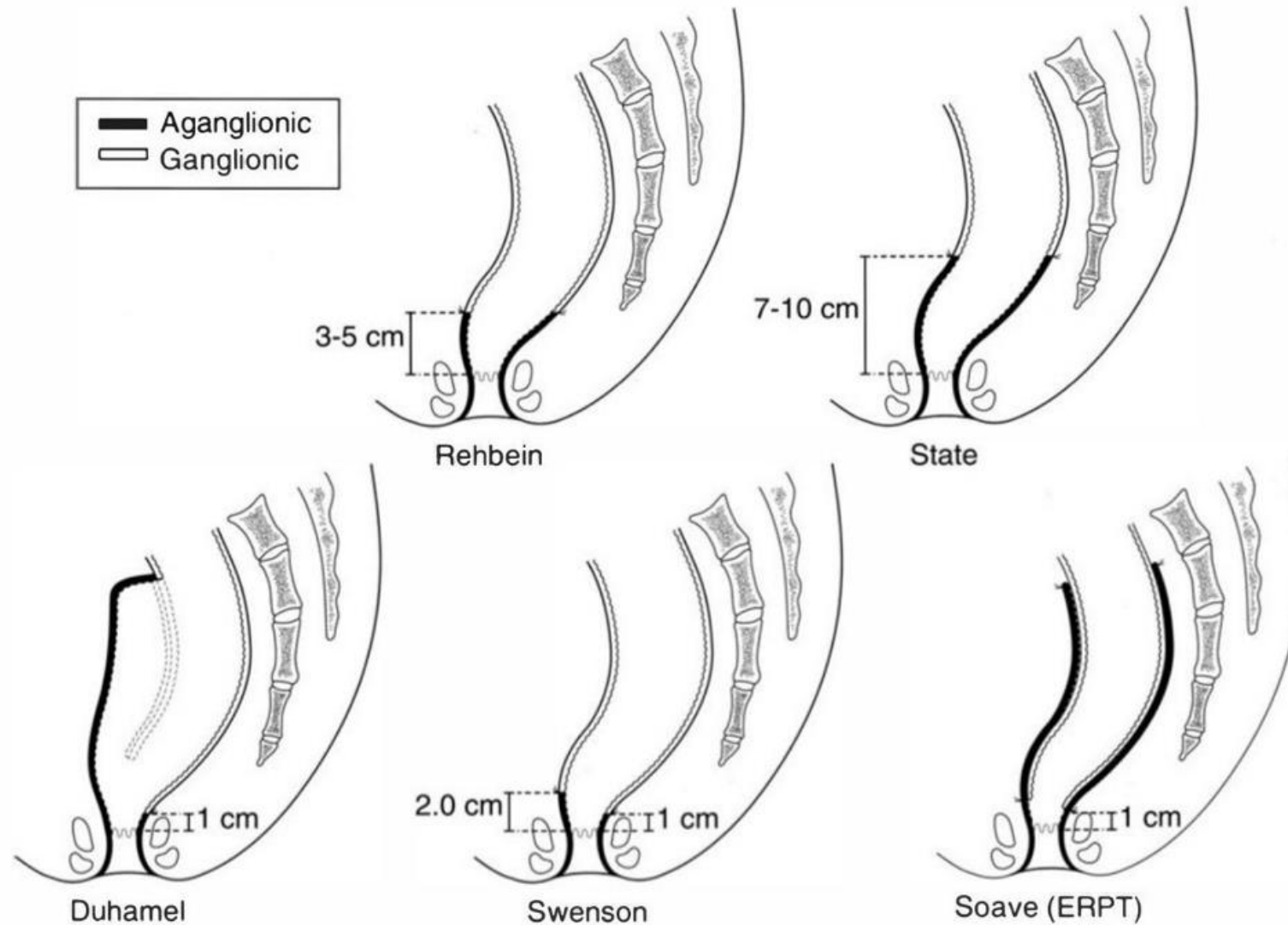
- Procedure

- Swenson
- Duhamel
- Soave

OPERATIVE PROCEDURE



OPERATIVE PROCEDURE



SWENSON'S PROCEDURE

- Initially develop one stage procedure
- High incidence of stricture, leak led to preliminary colostomy with definitive pull through 3-12 months later
- Nowadays one-stage is safe, avoid morbidity and stoma, more cost effective

SWENSON PROCEDURE

- Remove entire aganglionic colon + end to end anastomosis above anal sphincter
- Important to keep dissection in the correct plane along rectal wall to avoid injury to deep pelvic nerve, vessel

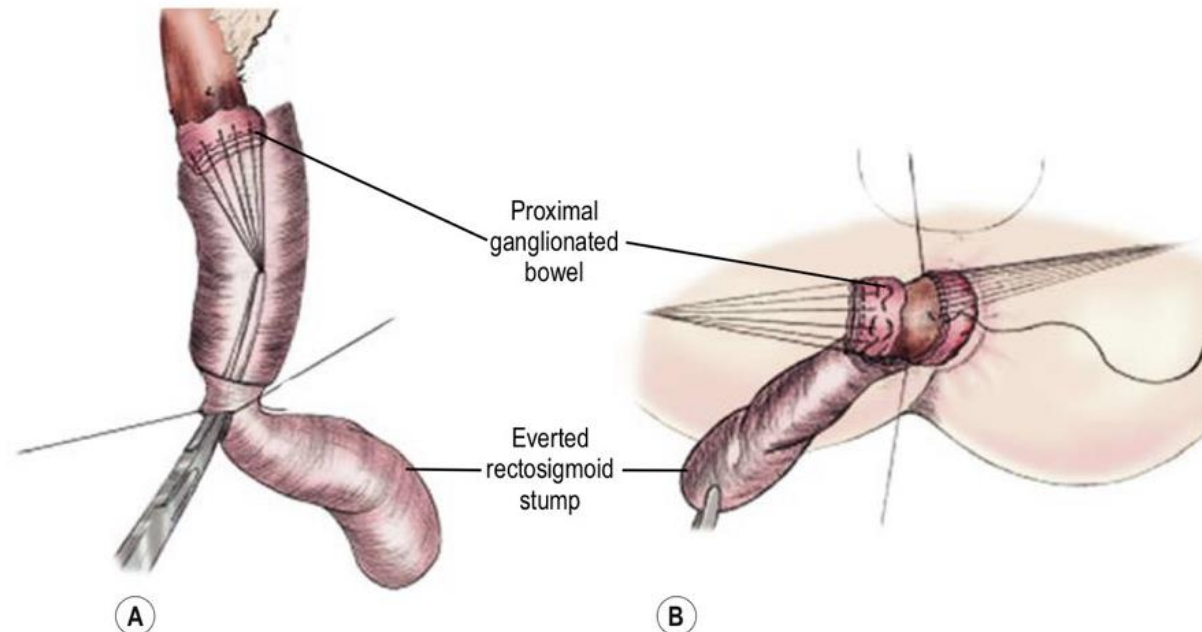


Fig. 34.8 The principles of the Swenson pull-through procedure are seen in these drawings. **(A)** The proximal ganglionated bowel is grasped through an incision in the prolapsed rectosigmoid stump. **(B)** The ganglionated bowel is then sewn to the anus.

SWENSON PROCEDURE

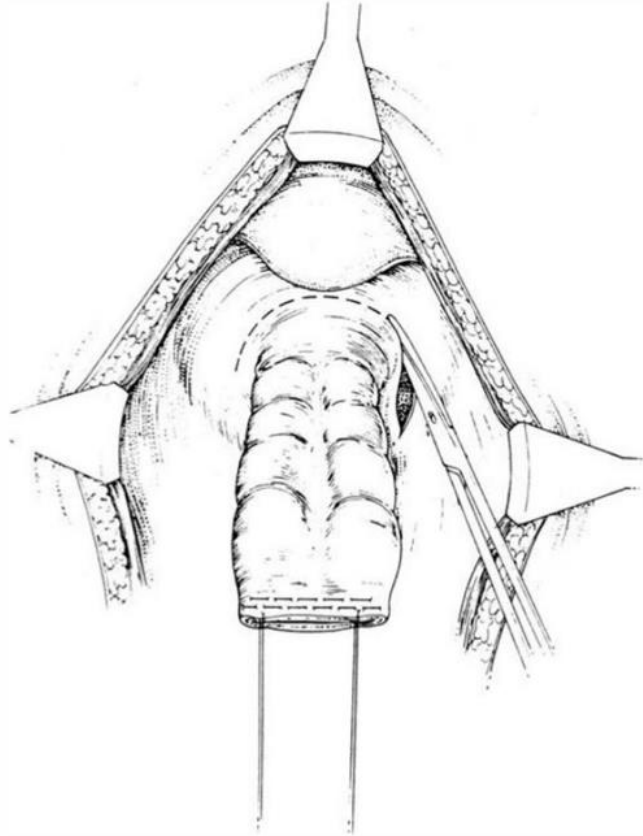


FIGURE 44-28 The peritoneal reflection is incised anteriorly.

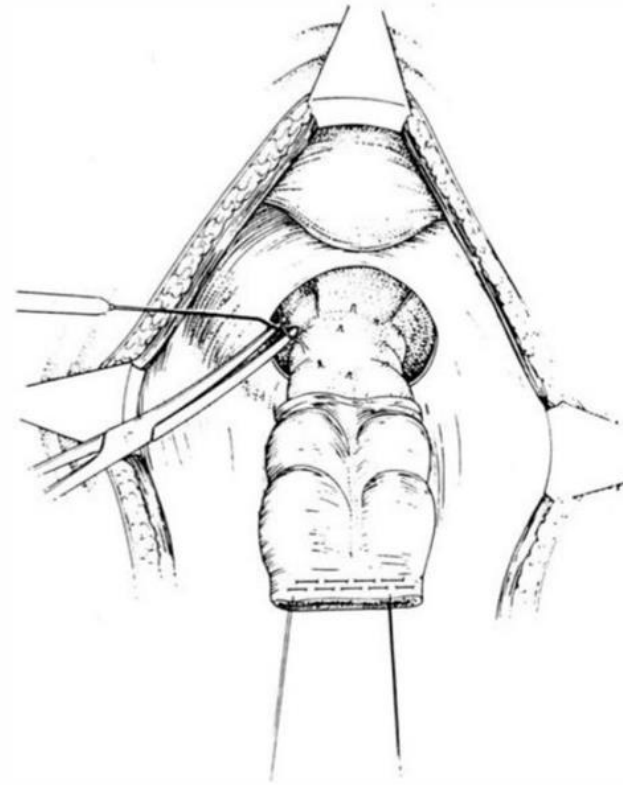


FIGURE 44-29 Dissection continues along the outer rectal wall, taking care to proceed less aggressively anteriorly, to within 2 cm of the verge.

SWENSON PROCEDURE

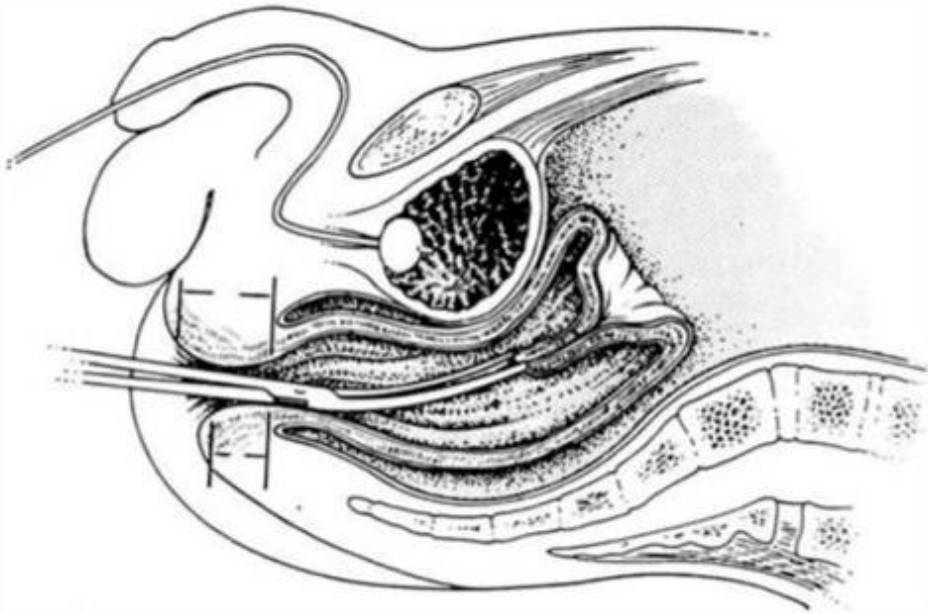


FIGURE 44-30 The full-thickness mobilized rectum is everted out of the anal canal.

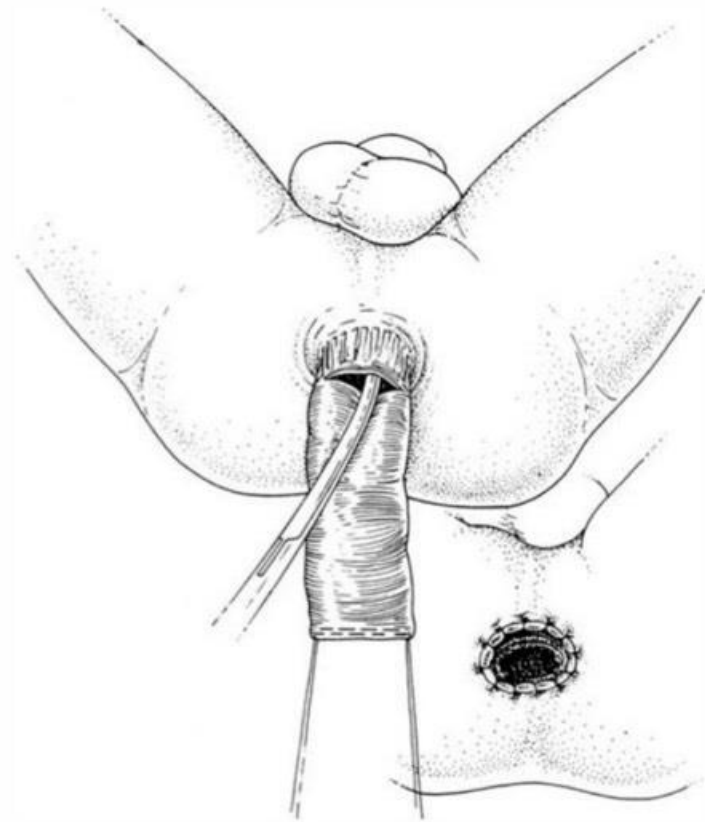
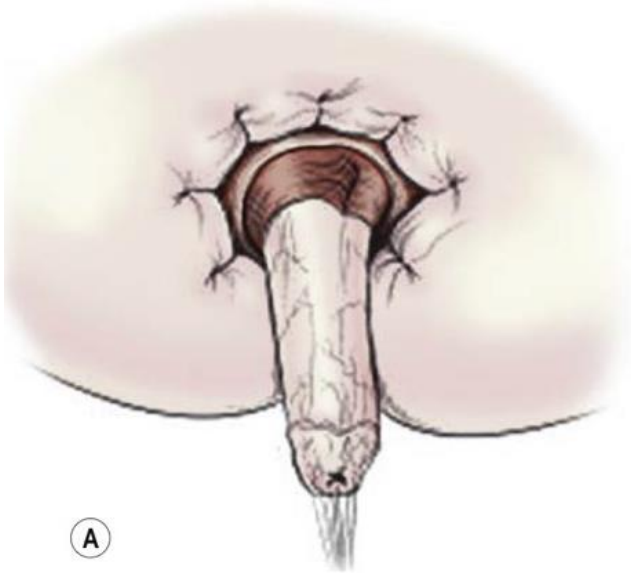


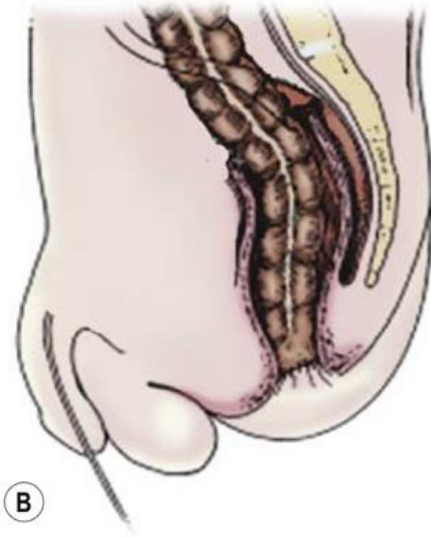
FIGURE 44-31 The anterior half of the everted bowel is incised, the ganglionated proximal colon is advanced through the pelvic space, and the extrapelvic end-to-end anastomosis is completed with interrupted sutures.

SOAVE PROCEDURE

- Designed to **avoid the risk** of injury to important pelvic structure
- Submucosal endorectal dissection and positioning the pull-through bowel within aganglionic muscular "cuff"



A



B

Long term constipation due to incomplete excision of aganglionic rectum

SOAVE PROCEDURE

- Typically perform in newborn period (1-2 weeks of life)

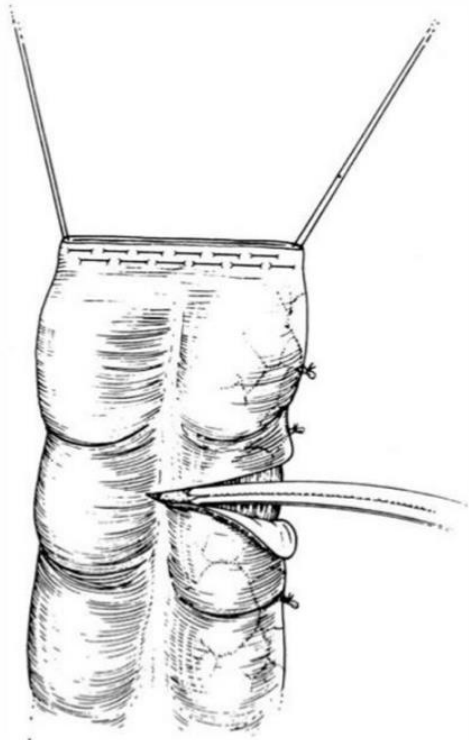


FIGURE 44-21 Developing the plane between the submucosal and muscular layers in a circumferential fashion.

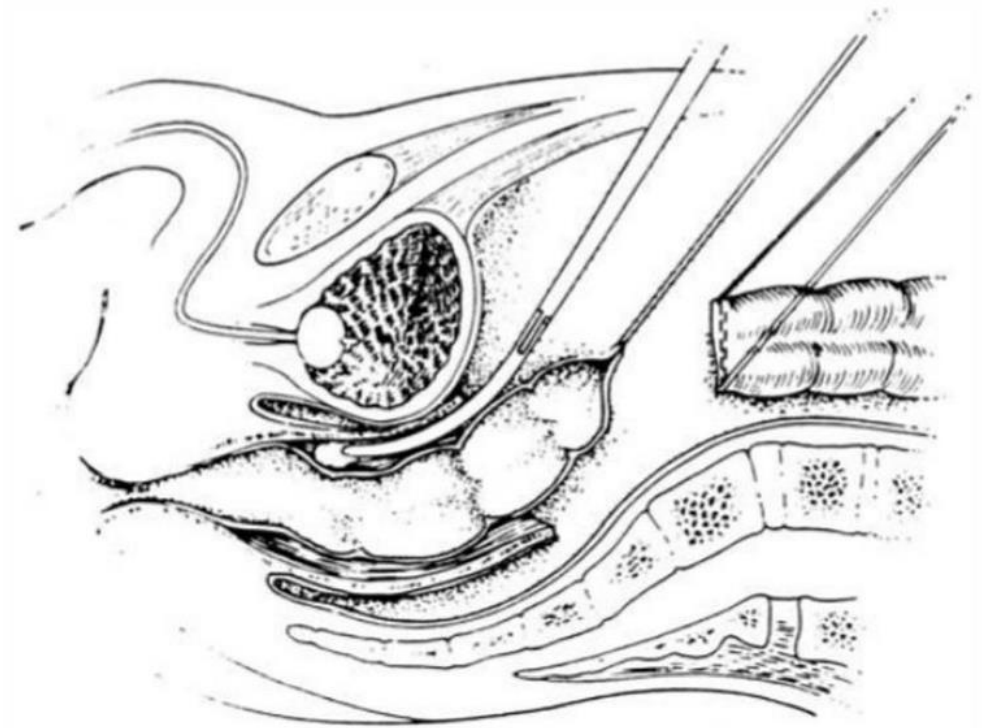


FIGURE 44-22 The endorectal dissection is carried distally to within 1 to 1.5 cm from the anal opening.

SOAVE PROCEDURE

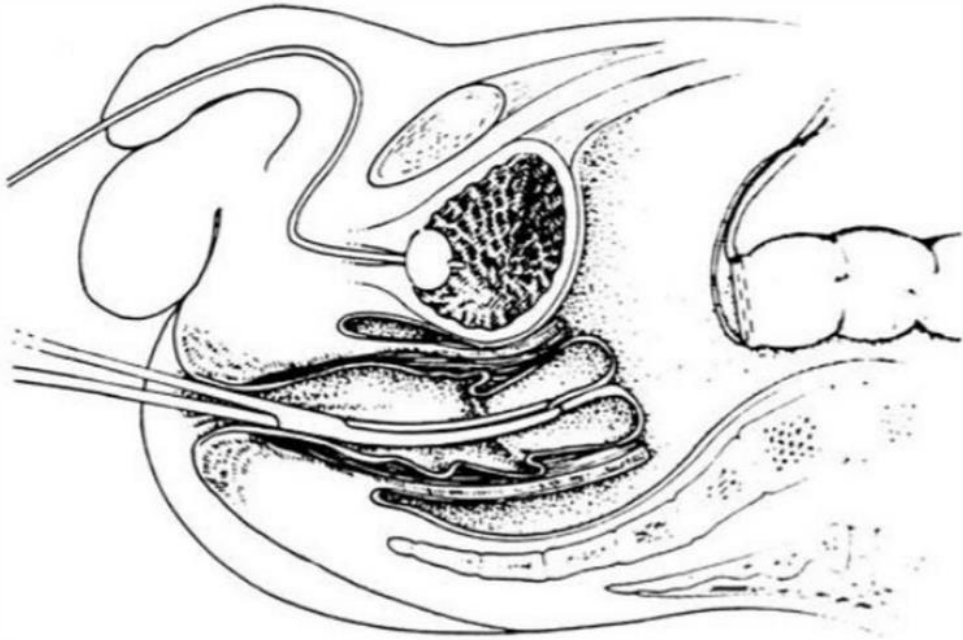


FIGURE 44-23 The mucosal tube is grasped with a transanal inserted clamp and everted out of the rectal canal.

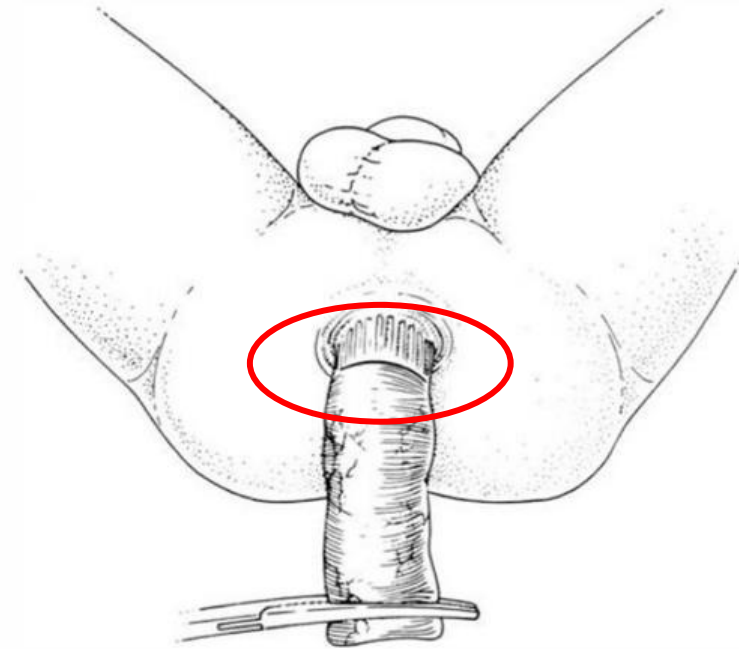


FIGURE 44-24 With traction on the mucosal tube, incision is made 0.5 to 0.7 cm above the dentate line anteriorly.

SOAVE PROCEDURE

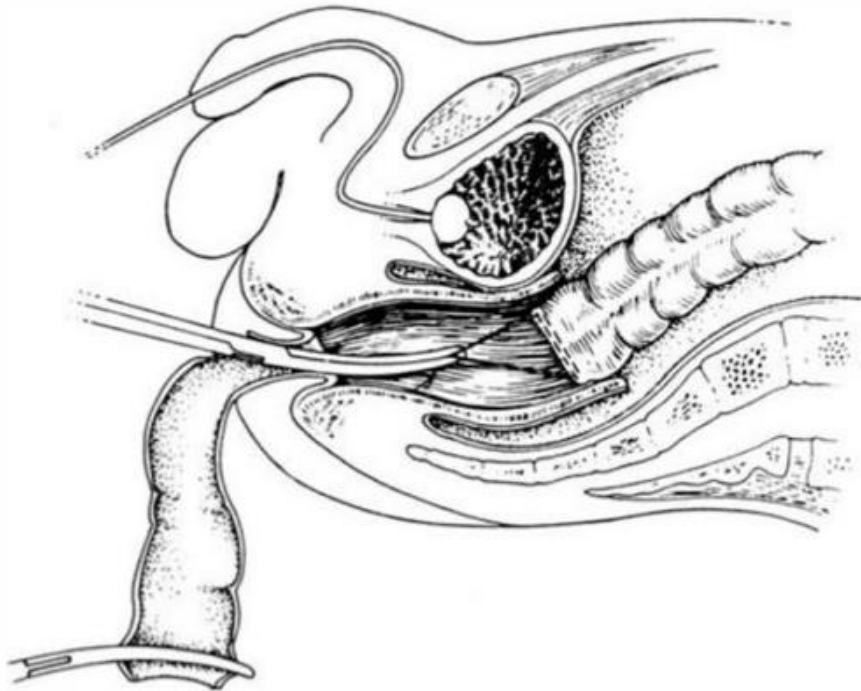


FIGURE 44-25 The proximal ganglionated bowel is pulled under direct vision through the residual muscular sleeve.

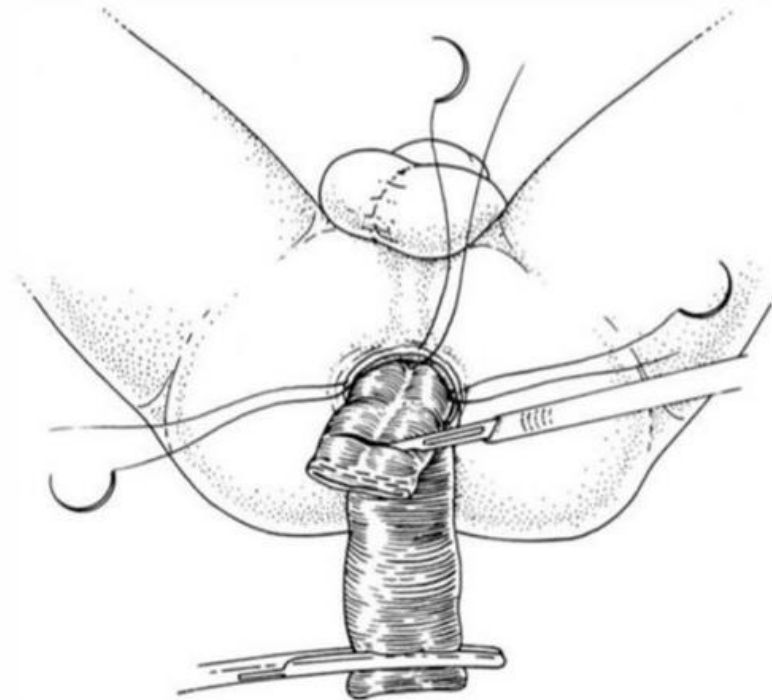
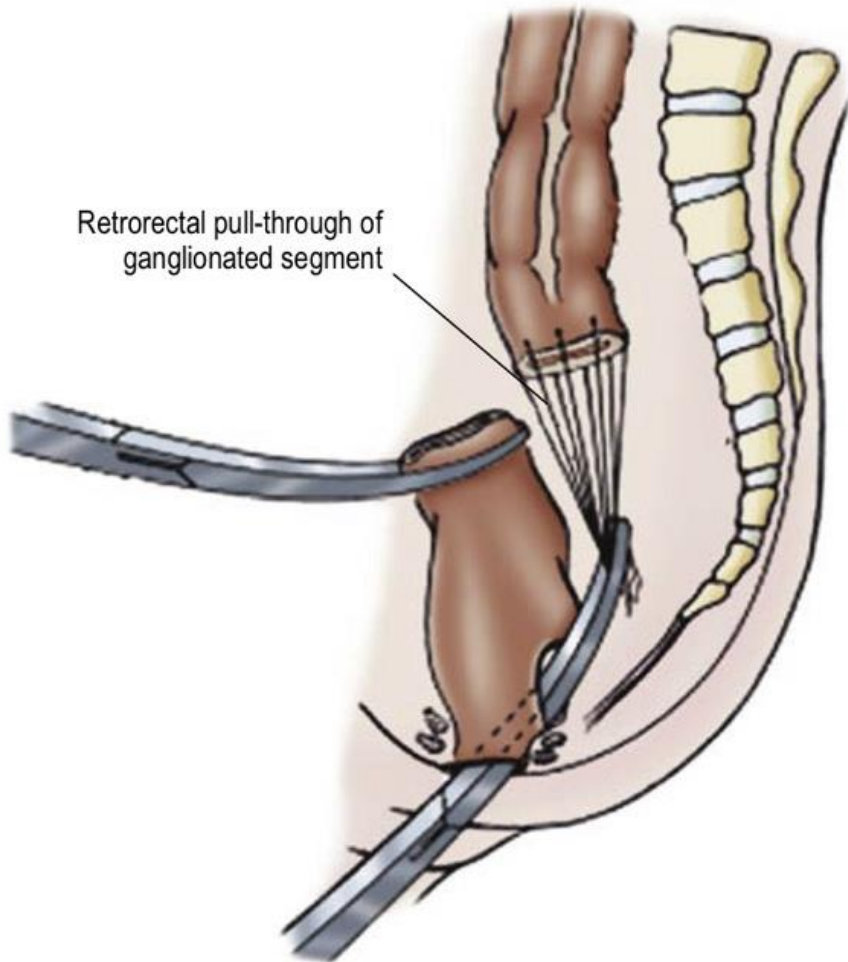


FIGURE 44-26 The anterior wall anastomosis is completed between the distal anterior rectal wall just above the dentate line to the pulled-through rectum.

DUHAMEL PROCEDURE



1. Bring normal colon down through bloodless plane between rectum and sacrum and
2. Joining two walls with a linear stapler to create a new lumen that is aganglionic anteriorly and normally innervated posteriorly

DUHAMEL PROCEDURE

- Advantages
 - Easier and safer
 - Less pelvic dissection
 - Large anastomosis -> ↓ risk of stricture

DUHAMEL PROCEDURE

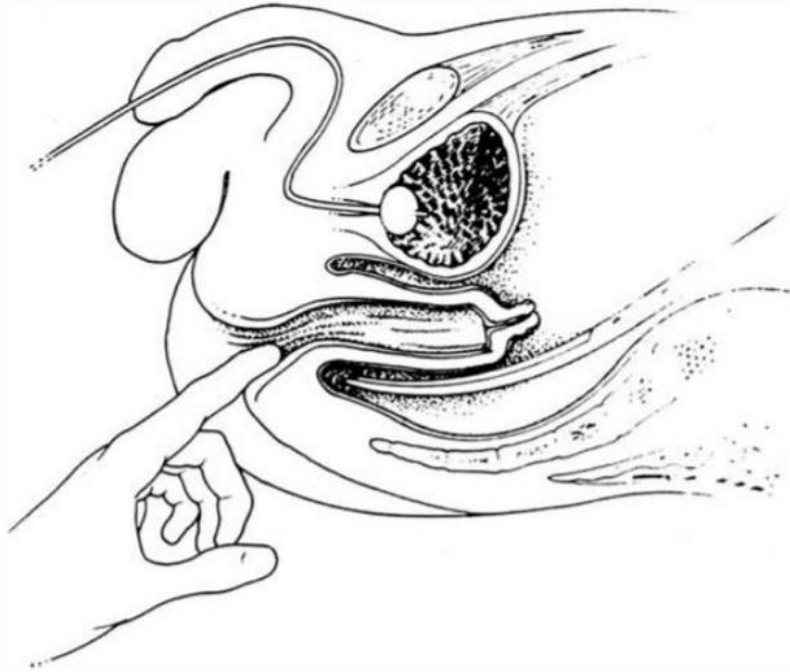


FIGURE 44-8 Development of the retrorectal space for the Duhamel pull-through.

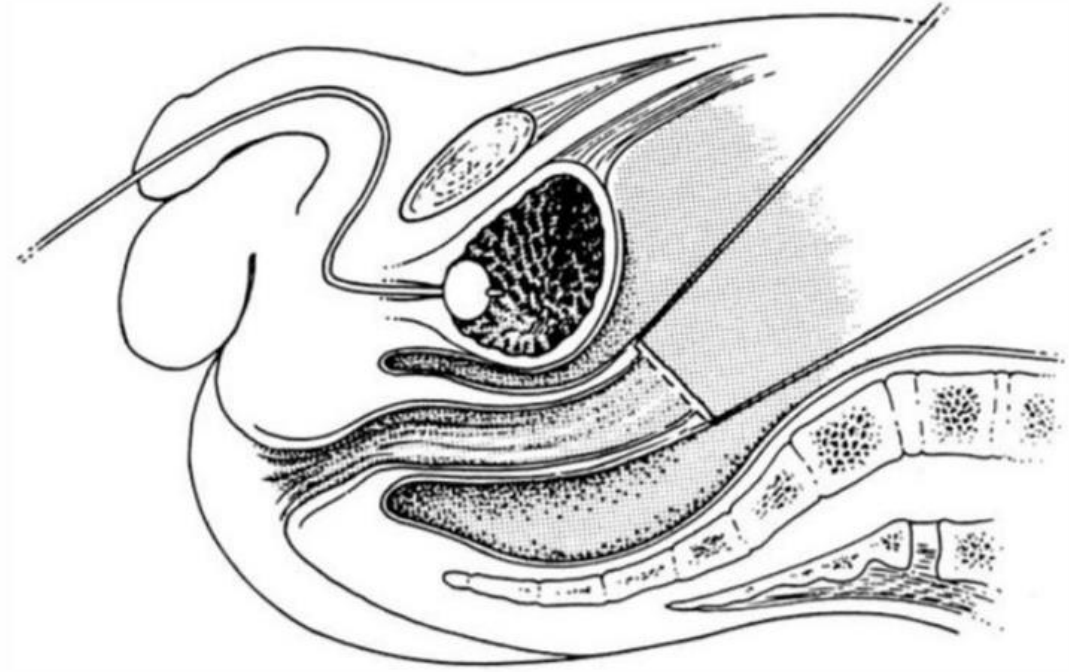


FIGURE 44-9 Traction sutures suspend the distal rectum in preparation for the retrorectal pull-through.

DUHAMEL PROCEDURE

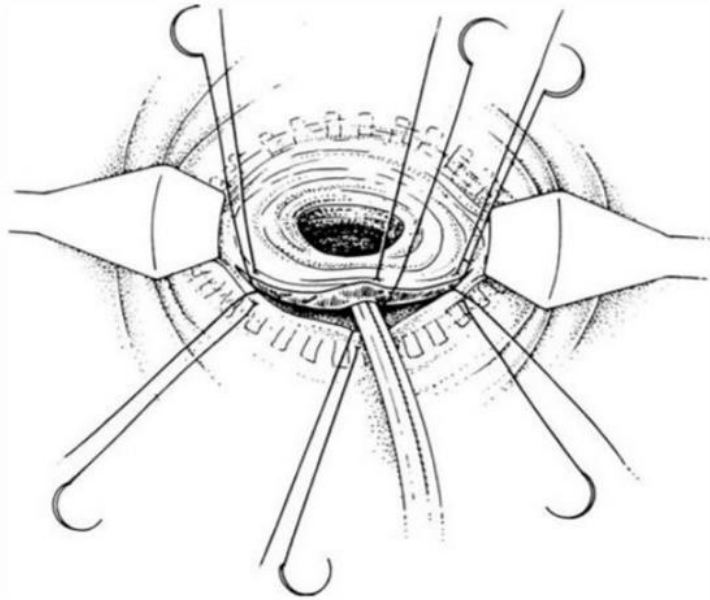


FIGURE 44-10 A ring-clamp is inserted into the retrorectal space from below.

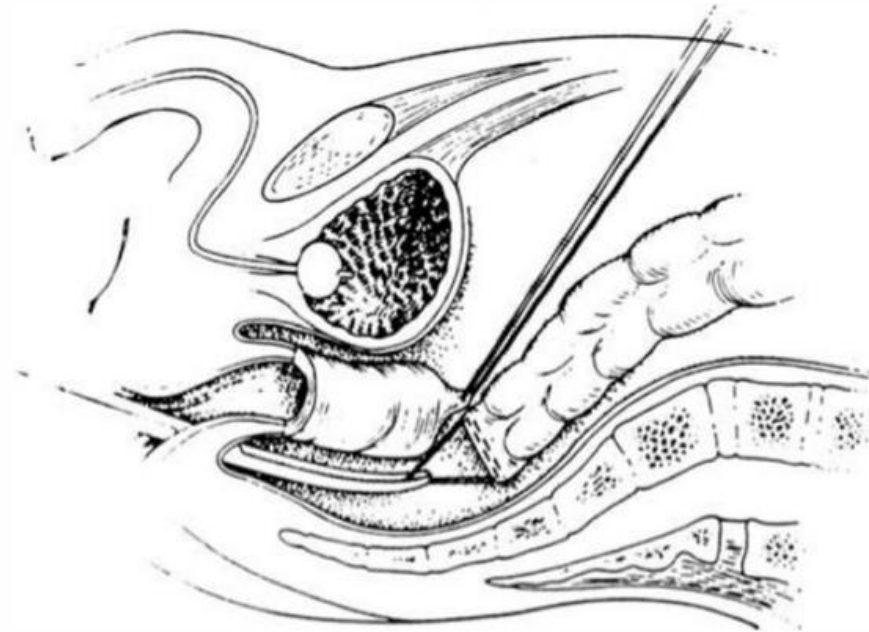


FIGURE 44-11 The ganglionated bowel is gently pulled through the retrorectal space and angulation/twisting is prevented.

DUHAMEL PROCEDURE

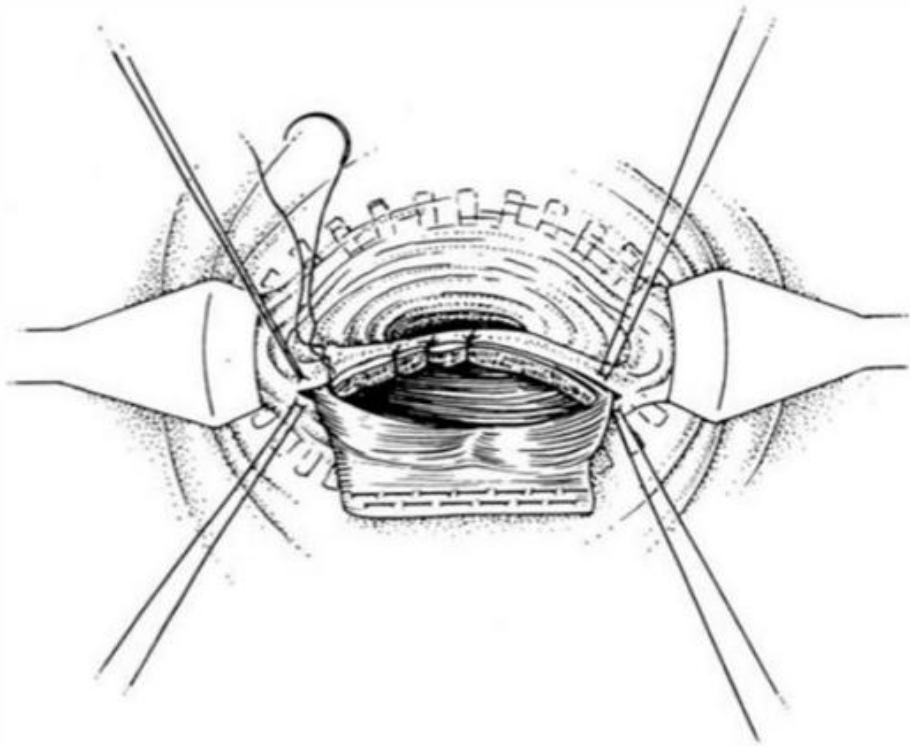


FIGURE 44-14 A single-layer anastomosis is made from the posterior wall of the "old" rectum to the anterior wall of the pulled through rectum.

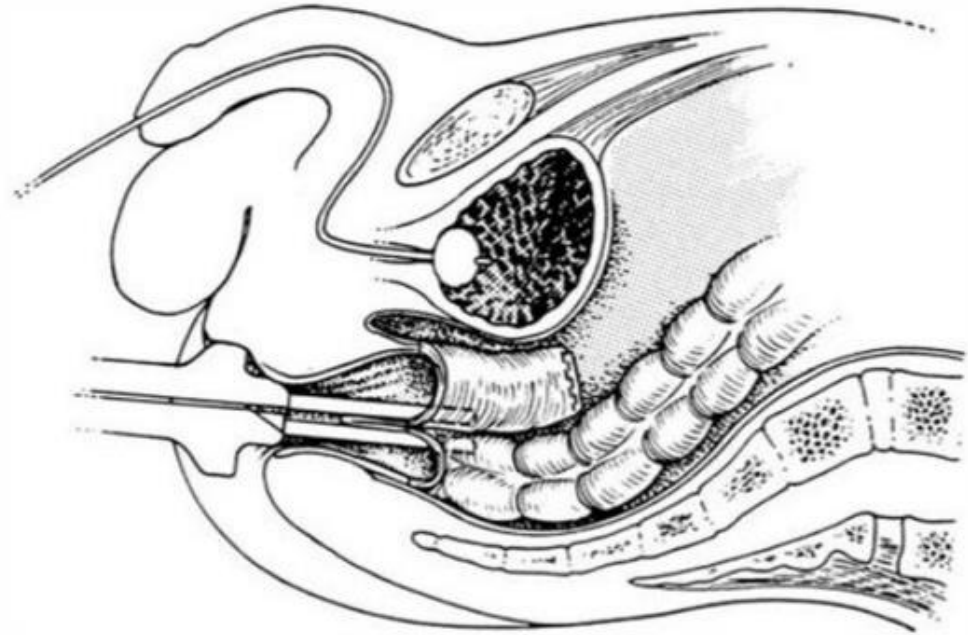
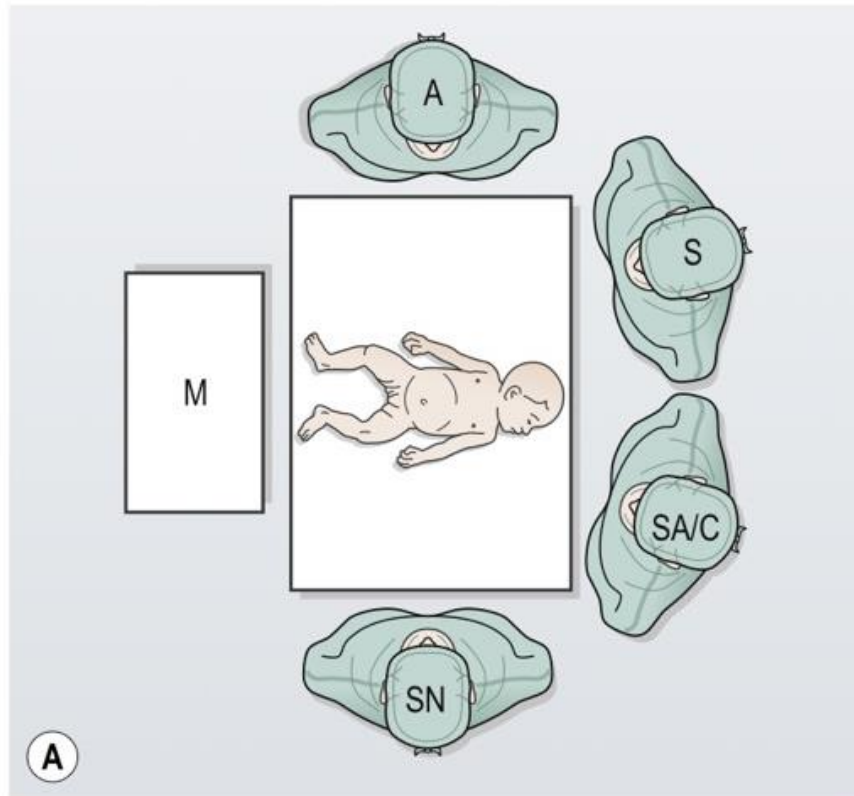


FIGURE 44-16 A GIA stapler is placed transanally, one arm in the rectum, and one arm in the pulled-through rectum.

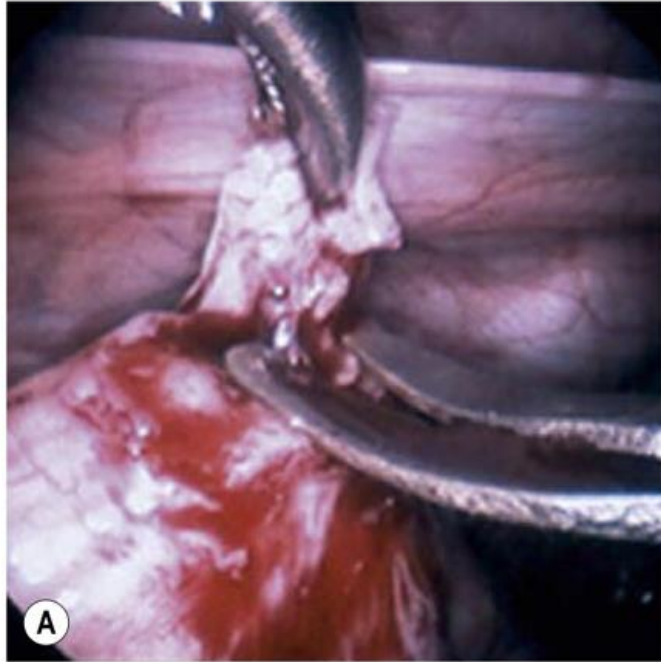
LAPAROSCOPIC PULL-THROUGH

- First described in 1995 by Georgeson
- Step
 1. Laparoscopic biopsy -> identify TZ
 2. Laparoscopic mobilization of rectum below peritoneal reflection
 3. Short mucosal dissection through a perineal approach
 4. Rectum prolapsed through anus and performed anastomosis from below

LAPAROSCOPIC PULL-THROUGH



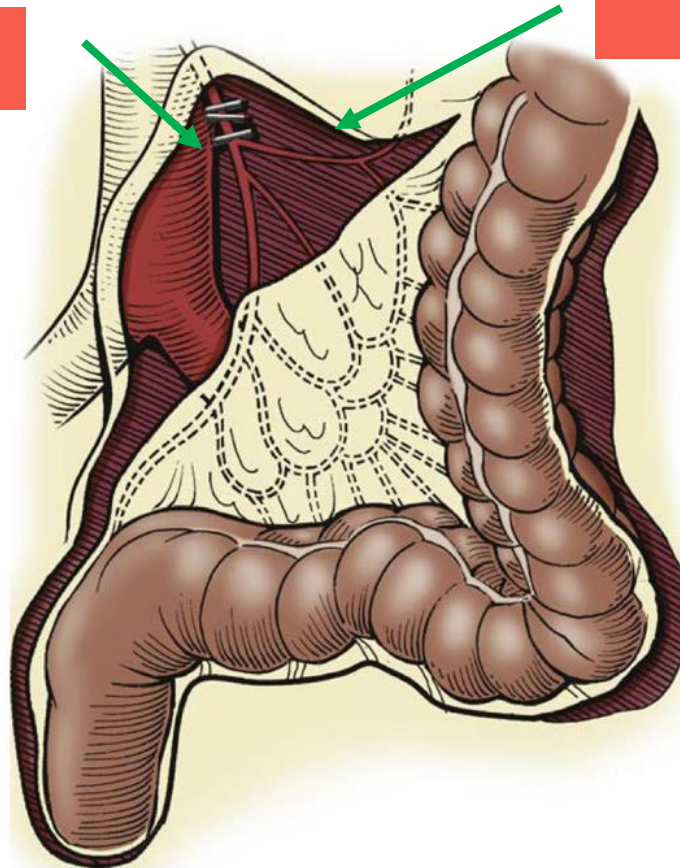
LAPAROSCOPIC PULL-THROUGH



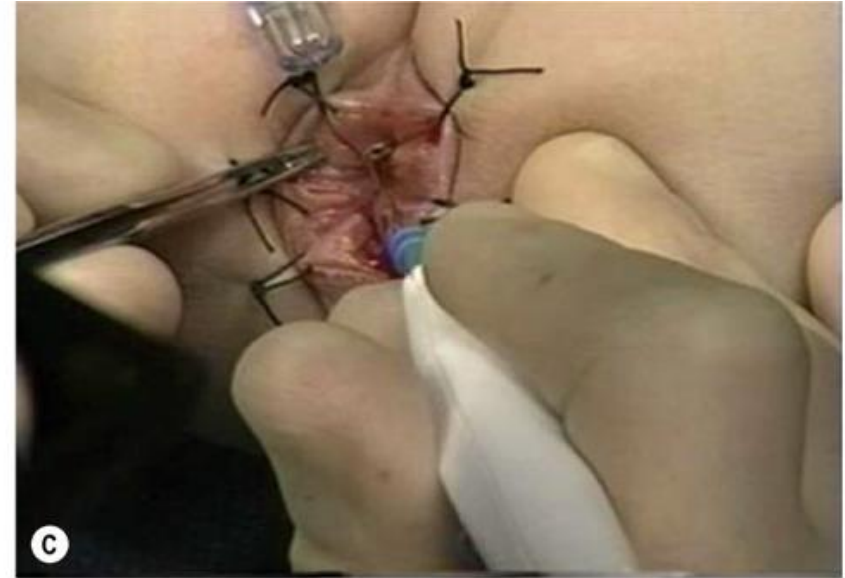
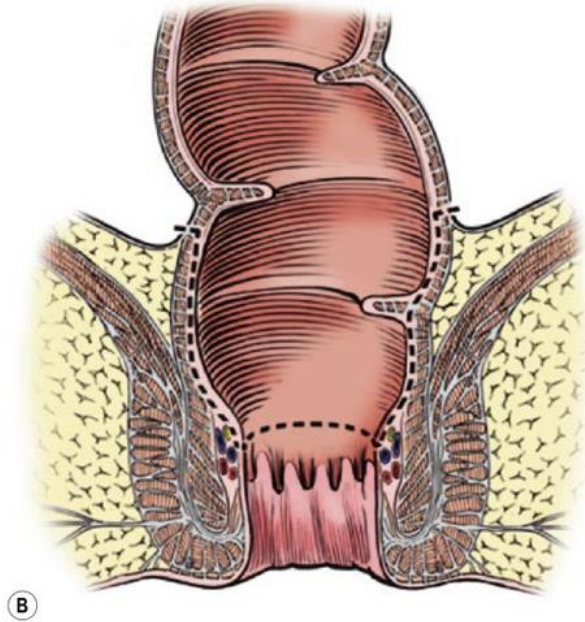
LAPAROSCOPIC PULL-THROUGH

Inferior mesenteric artery

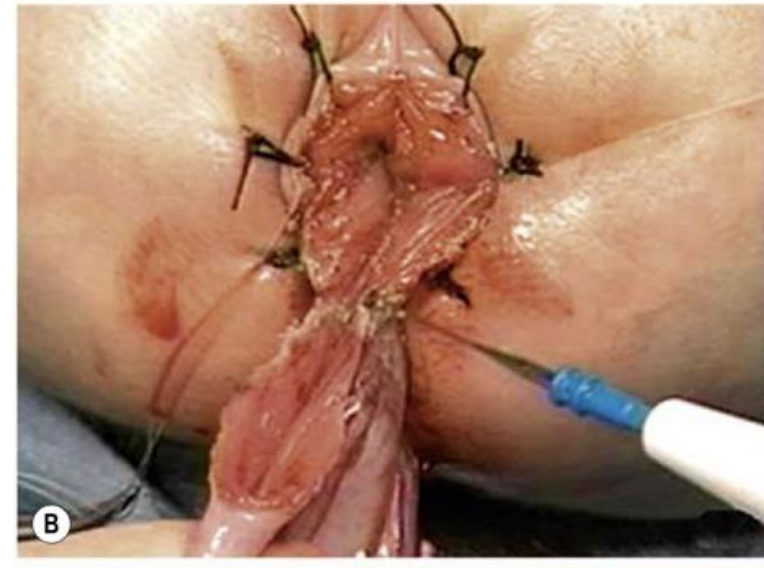
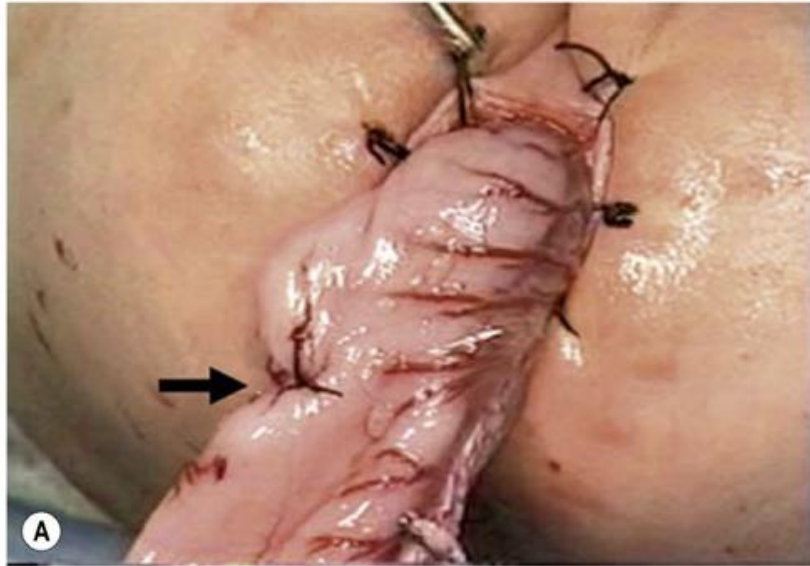
Left colic artery



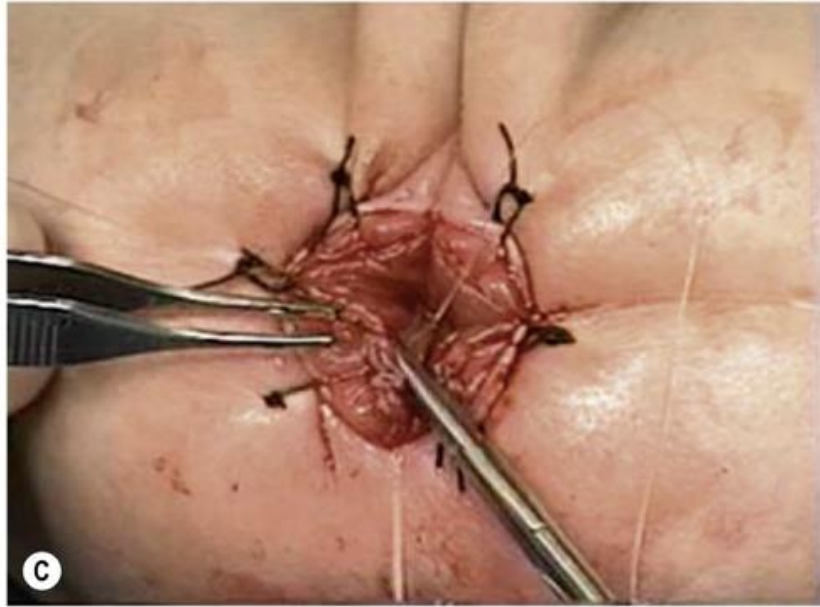
LAPAROSCOPIC PULL-THROUGH



LAPAROSCOPIC PULL-THROUGH



LAPAROSCOPIC PULL-THROUGH



TRANSANAL (PERINEAL) PULL-THROUGH

- Position – prone or lithotomy
- Mucosal incision 0.5 – 1.0 cm above dentate line
- Submucosal dissection varies according to surgeon
- When transitional zone is reached the anastomosis is performed from below

TRANSANAL (PERINEAL) PULL-THROUGH

- Rectal muscle is incised circumferentially
- Dissect continued on rectal wall divided vessel that enter rectum
- Entire rectum and part of sigmoid can be delivered to anus
- When TZ reached, anastomosis is performed from below
- In case proximal TZ
 - Laparoscopy or umbilical incision is needed to mobilized left colon and/or splenic flexure

TRANSANAL (PERINEAL) PULL-THROUGH



TRANSANAL (PERINEAL) PULL-THROUGH

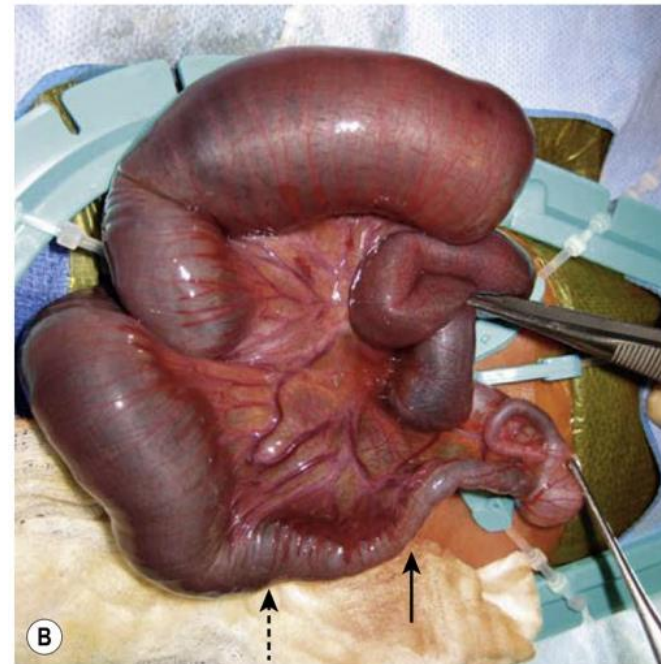
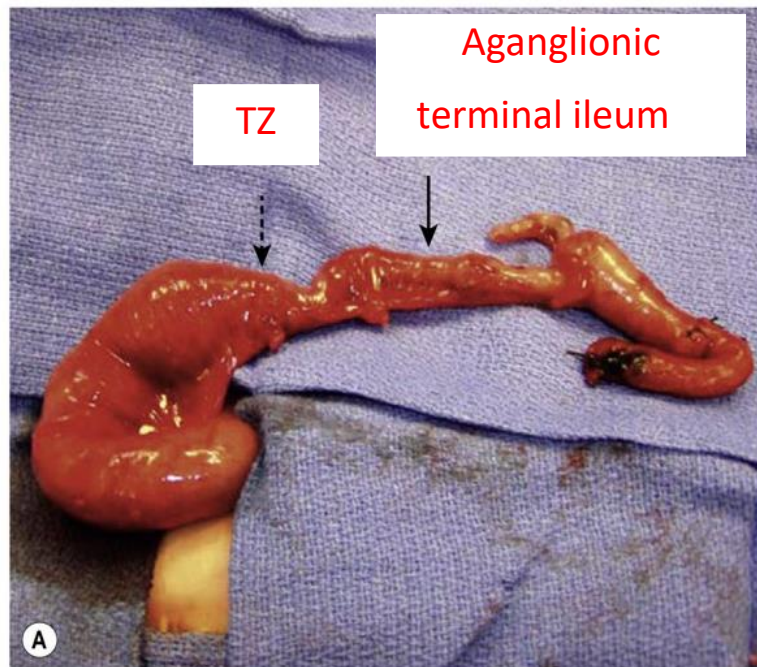


SHORT SEGMENT HIRSCHSPRUNG

- Some advocated nonoperative long-term management
 - Enema
 - Laxative
- Some suggest simple myectomy may be adequate
- Do not provide good quality of life
- Most surgeon still recommended pull-through procedure

LONG SEGMENT AGANGLIOSIS

- Usually defined as transition zone is proximal to **mid-transverse colon**
- Most common is total colonic aganglionosis which may also include distal ileum



LONG SEGMENT AGANGLIOSIS

- Presentation
 - Distal small bowel obstruction (may not present until after weaning from breast milk)
- Investigation
 - Contrast enema - narrow " question mark" colon
- Biopsy
 - No ganglion cell, most have no hypertrophic nerve



LONG SEGMENT AGANGLIOSIS

- Management
 - Sequential colonic biopsies via laparotomy or laparoscopy or umbilical incision
 - Cecum is preferable site for initial biopsy
 - Once diagnosis
 - Determine the extent of aganglionosis based on frozen section
 - **Create stoma** at most distal point that has normally innervated bowel
 - Gastrostomy considered for continuous feeding

OPERATIVE TECHNIQUES FOR RECONSTRUCTION

1. Straight pull-through
 - Bring normal innervated ileum to just above anal sphincter
 - Swenson
 - Duhamel
 - Soave
2. Colon patch
3. Ileal J-pouch construction

OPERATIVE TECHNIQUES

- Colon patch
 - Side to side anastomosis between normal and aganglionic colon
 - Small bowel for motility and colon as reservoir

Left colon (Martin)

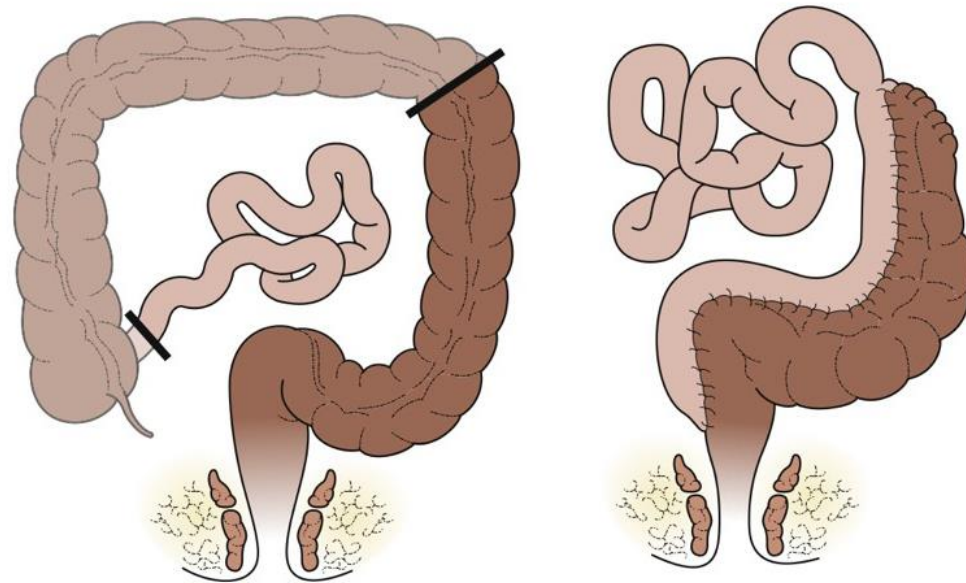
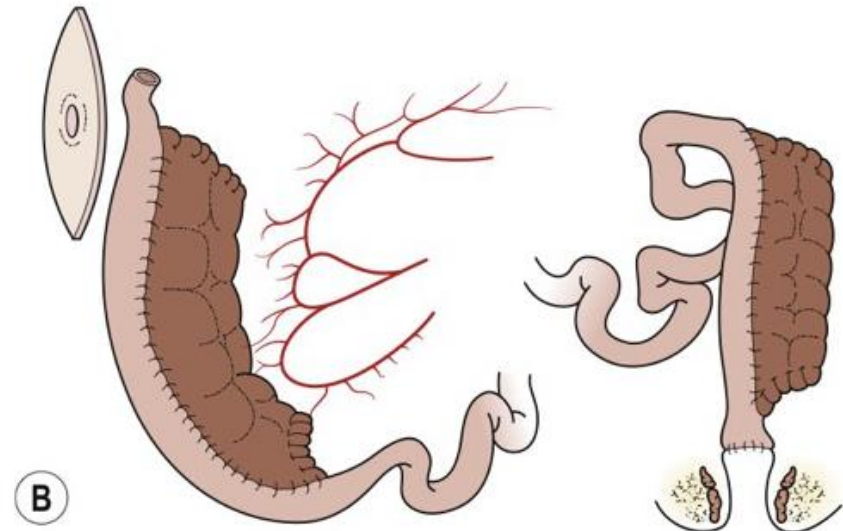
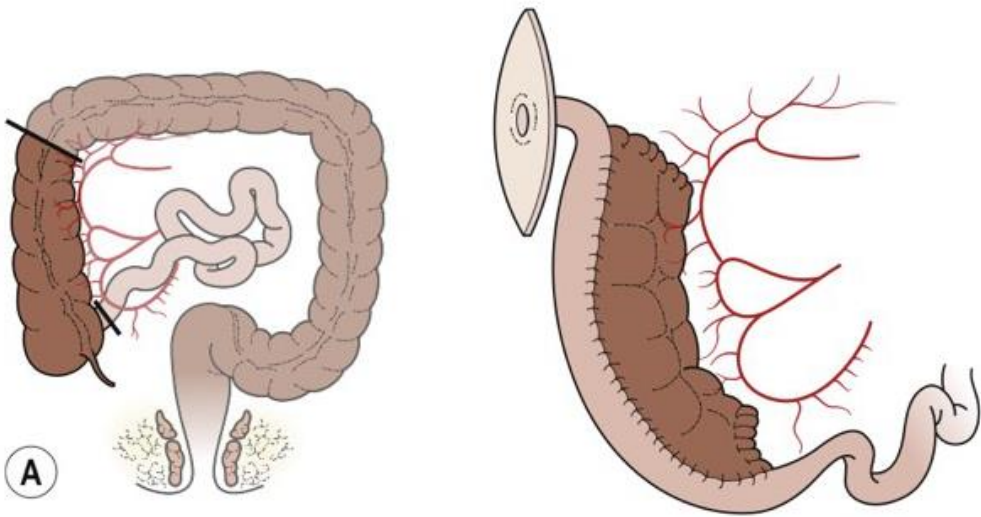


Fig. 34.19 This diagram depicts the Martin procedure for total colon aganglionosis.

OPERATIVE TECHNIQUES

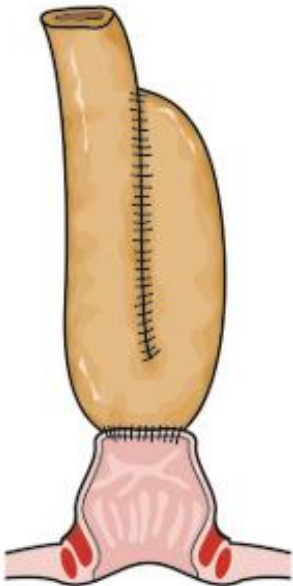
Right colon (Kimura)



COLON PATCH

- Colon patch
 - In theory -> decreased stool output due to better water absorption
 - In reality -> aganglionic colon tends to dilate -> enterocolitis

J pouch



- Straight pull-through or J-pouch tends to decrease stool frequency

NEAR TOTAL INTESTINAL AGANGLIOSIS

- In entire intestinal tract agangliosis usually leave 10-40 cm of innervated jejunum
- Not enough functional bowel to support enteral nutrition -> intestinal failure
- For patient develop significant dilatation of normal innervated bowel ,tapering, imbrication or bowel lengthening procedure such as Bianchi or serial transverse enteroplasty (STEP) may be helpful

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POSTOPERATIVE MANAGEMENT & COMPLICATION

POSTOPERATIVE MANAGEMENT

- Patient undergo laparoscopic or transanal pull-through can be fed immediately discharge in 24-48 hr
- Dilation
 - Anastomosis calibrated with appropriately sized dilator or finger 1-2 week after procedure
 - Daily vs weekly for 4-6 weeks
- Protect buttocks with barrier cream to prevent skin breakdown
- Educate family sign of enterocolitis

POSTOPERATIVE COMPLICATION

- Wound infection
- Intra-abdominal bleeding
- Anastomotic complication
 - Leak
 - Stricture
- Intestinal perforation
- Bowel obstruction



LONGTERM OUTCOME

LONG-TERM OUTCOME

- Ongoing obstructive symptoms
- Soiling
- Enterocolitis

1. OBSTRUCTIVE SYMPTOMS

- Symptoms

- Abdominal distension
- Vomiting
- Bloating
- Ongoing severe constipation

Box 34.2 Causes of Obstructive Symptoms After Operation for Hirschsprung Disease

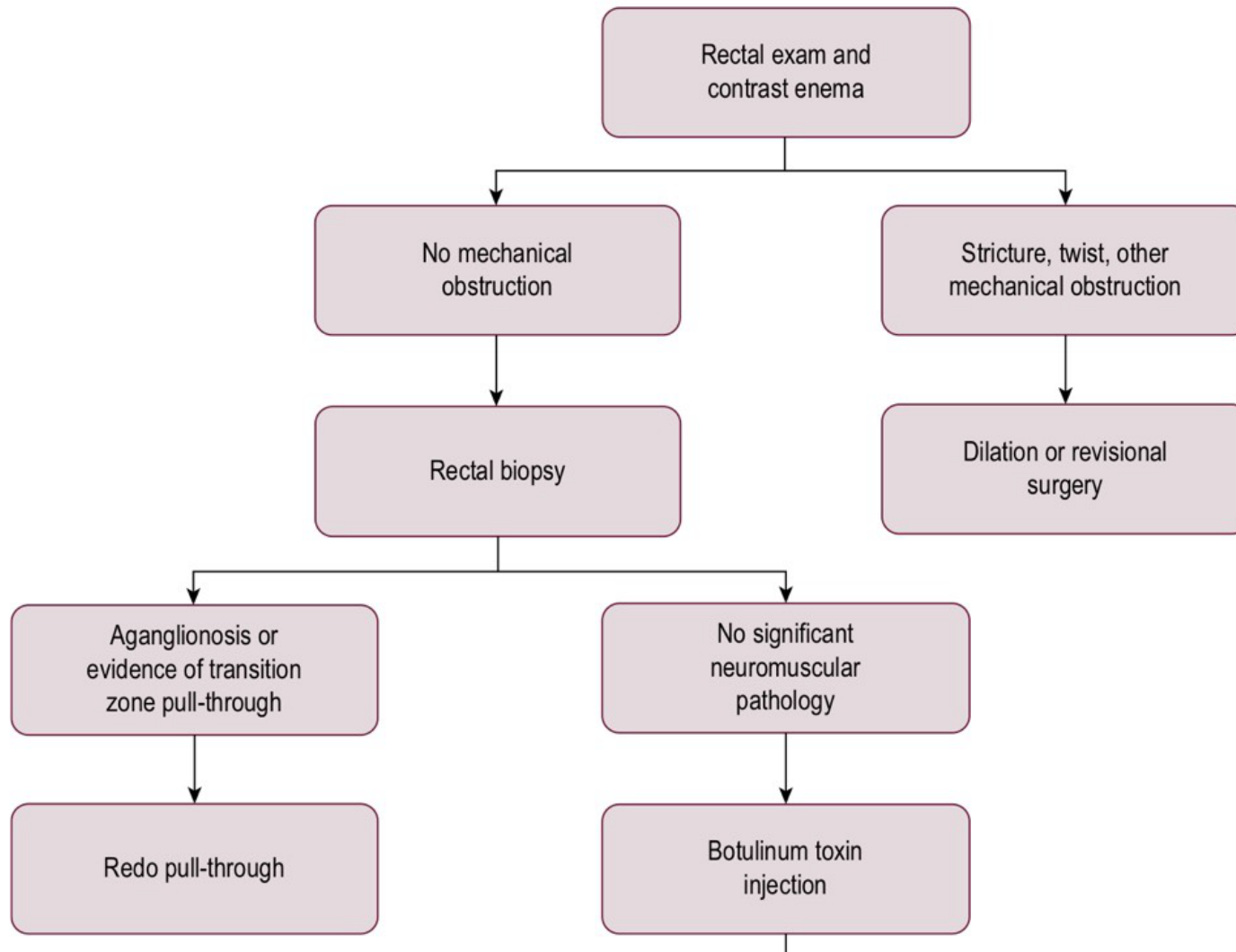
Mechanical obstruction

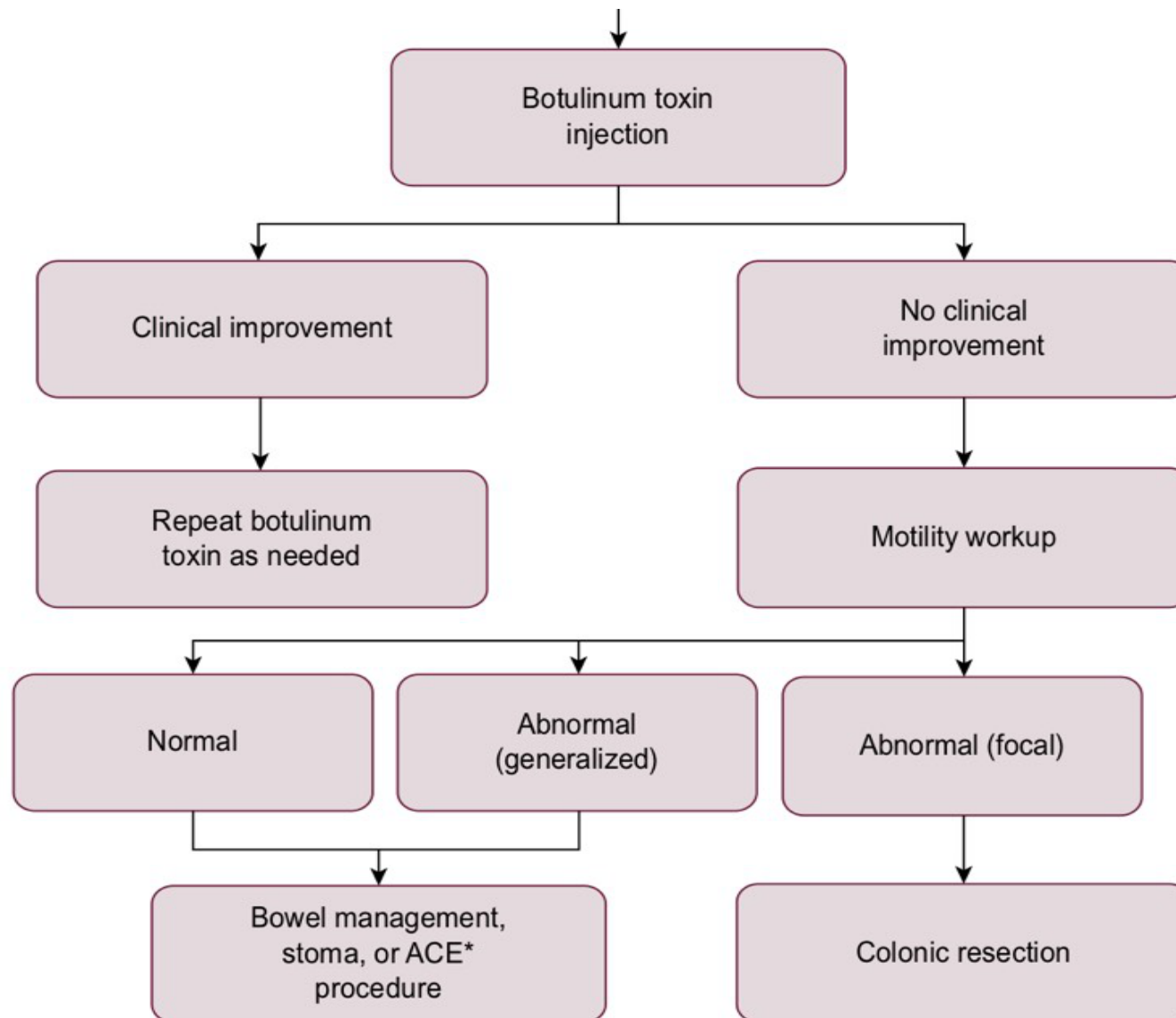
Recurrent or residual aganglionosis

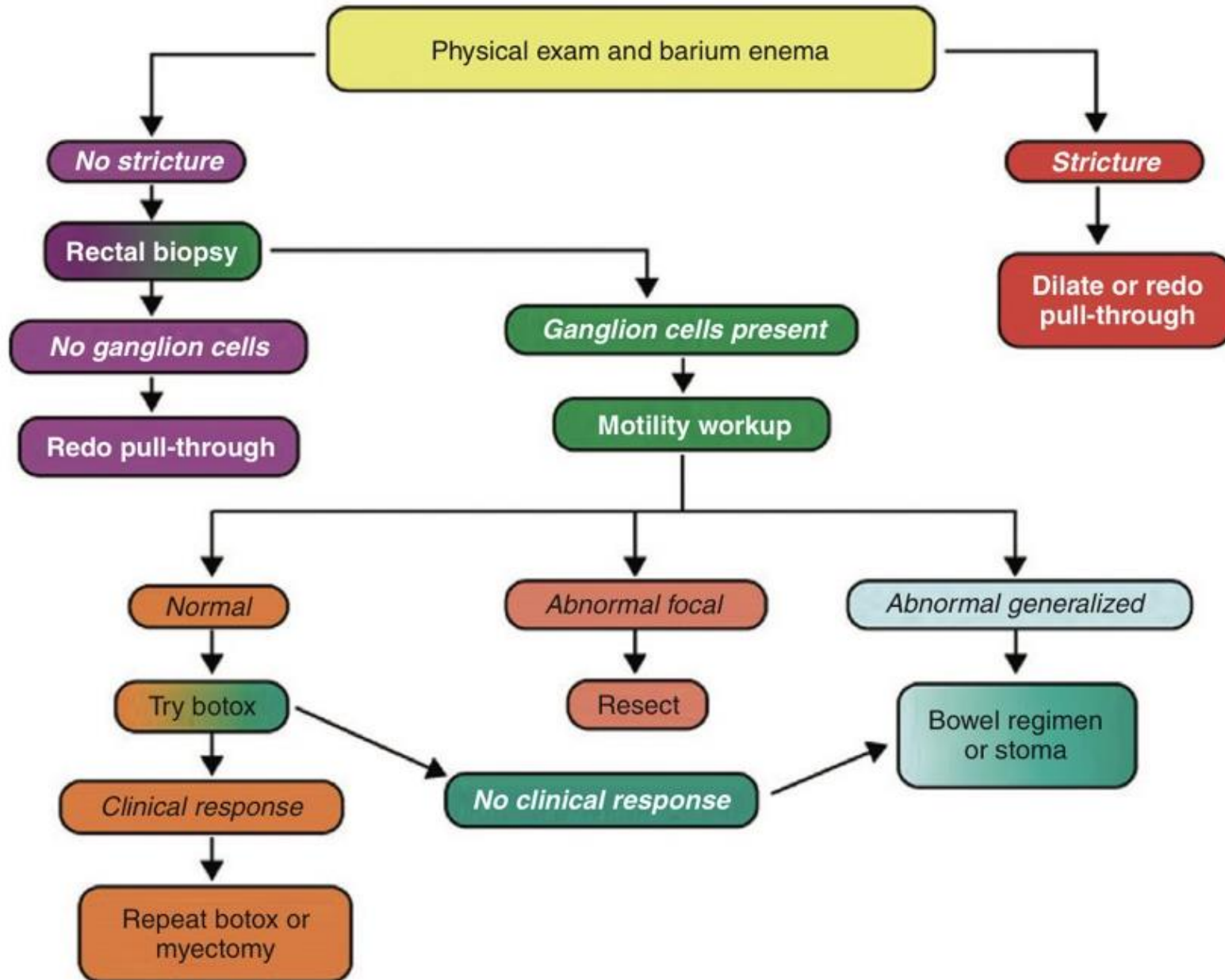
Motility disorder involving the ganglionated bowel

Internal anal sphincter achalasia

Functional megacolon (stool-holding behavior)







1.1 MECHANICAL OBSTRUCTION

- Most common is stricture
- Occur more in Swenson and Soave
- Duhamel may have retain "spur" of anterior aganglionic bowel

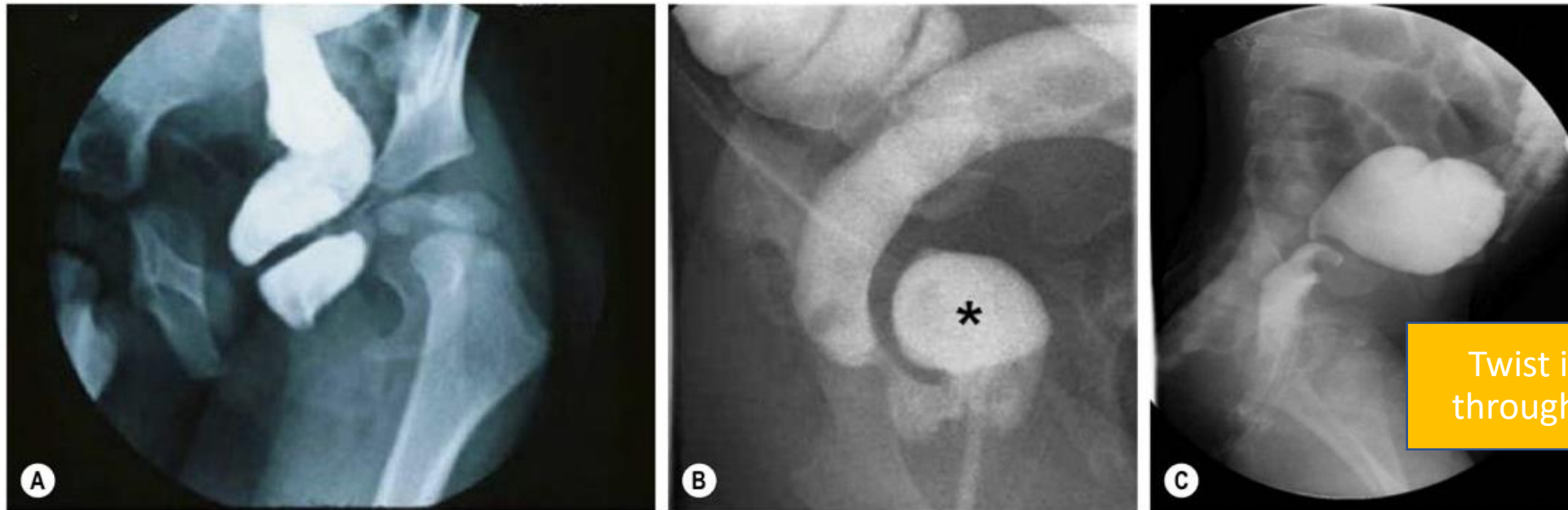


Fig. 34.22 Causes of mechanical obstruction after a pull-through are shown. **(A)** Stricture following a Soave procedure. **(B)** Anterior aganglionic "spur" (asterisk) following a Duhamel procedure. **(C)** Twisted transanal pull-through.

MECHANICAL OBSTRUCTION

- Diagnosis

- Digital rectal exam
- Barium enema

- Management

- Repeat dilatation by finger, dilator, balloon
- Recalcitrant stricture use antegrade dilatation with Tucker dilator + intralesional steroid or topical mitomycin C
- Revision of pull-through

1.2 PERSISTENT OR ACQUIRED ANGANGLIONOSIS OR TRANSITION ZONE PULL-THROUGH

- May due to error in histologic analysis, transition zone pull through, loss of ganglion cell
- Diagnosis
 - Biopsy above colo-anal anastomosis looking for the presence and number of ganglion cells
- Management
 - Repeat pull-through

1.3 MOTILITY DISORDER

- Often have abnormality motility through intestinal tract
- Diagnosis
 - Radiologic shape study, radionuclide colon transit study , colonic manometry , laparoscopic biopsy
- Management
 - Resect abnormal part
 - Diffuse treat with bowel management : antegrade enema , prokinetic agent

1.4 INTERNAL SPHINCTER ACHALASIA

- Obstructive symptoms caused by lack of normal RAIR
- Diagnosis
 - Clinical response to intrasphincteric botulinum toxin
- Management
 - Internal sphincterotomy or myectomy
 - Can spontaneous resolve : prefer chemical sphincterotomy with intrasphincteric botulinum toxin

1.5 FUNCTIONAL MEGACOLON

- Result of **stool holding behavior**
- More common in HD because of their predisposition to constipation
- Management
 - Bowel management regimen : laxative + behavior modification strategies
 - Antegrade enema or proximal stoma

2. FECAL SOILING



Fig. 34.23 Loss of the dentate line is seen in this child who had a transanal pull-through with an anastomosis that was done too low.

- Three broad causes for soiling after pull-through
 1. Abnormal sphincter function
 2. Abnormal sensation
 - Inability to feel rectal distension
 - Loss of transitional epithelium
- Management
 - Surgery
 - Constipate diet, stimulant laxative, rectal or antegrade enema

2. FECAL SOILING

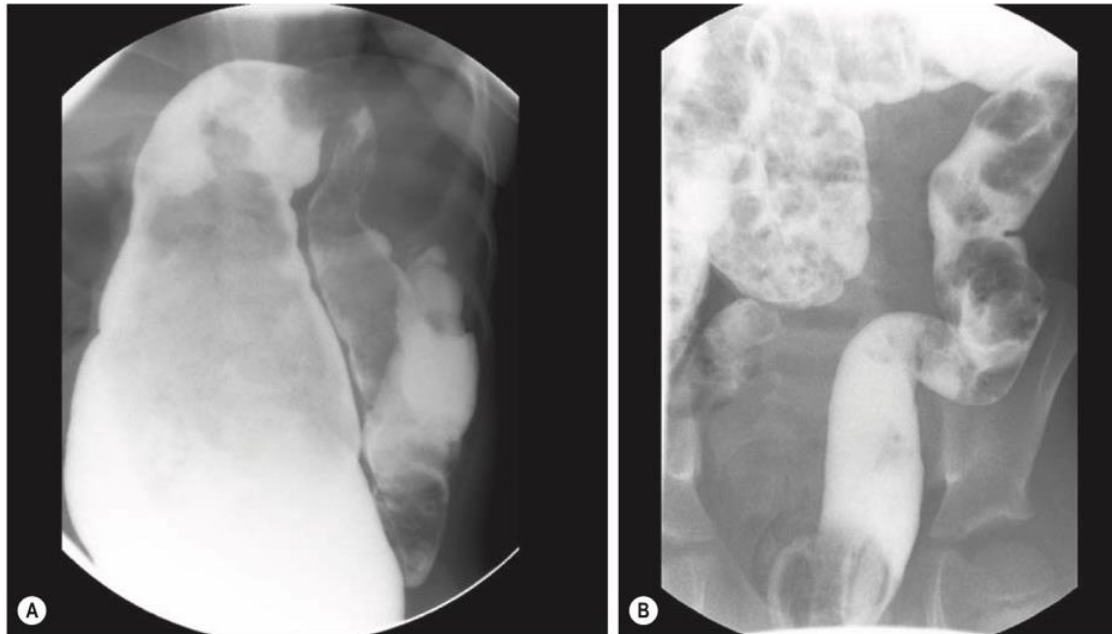


Fig. 34.24 The difference between obstipation and hyperperistalsis forms of pseudo-incontinence can be demonstrated on contrast enema. **(A)** With obstipation, the rectum is extremely dilated and the sigmoid colon may be very redundant. **(B)** With hyperperistalsis, the caliber of the sigmoid and rectum is more normal.

3. Pseudo-incontinence

Associated with severe constipation

- Massively distended rectum and overflow of liquid stool

Associated with hyperperistalsis of the pulled-through bowel

- Inability of anal sphincter to achieve control despite normal sphincter function

FECAL SOILING

- Management

- Severe constipation : laxative
- Sphincter sensation inadequate
 - stimulant laxative such as senna or bisacodyl
 - Inadequate achieving rectal emptying : require retrograde or antegeade enemas
- Stool-holding behavior with normal sphincter worsen behavioral problem by rectal enema

FECAL SOILING

- Management

- No constipation with hyperperistalsis, abnormal sphincter function or sensation
 - Constipating diet + loperamide
- Slow transit constipation or stool holding behavior
 - high-fiber diet and passive laxative therapy

3. ENTEROCOLITIS

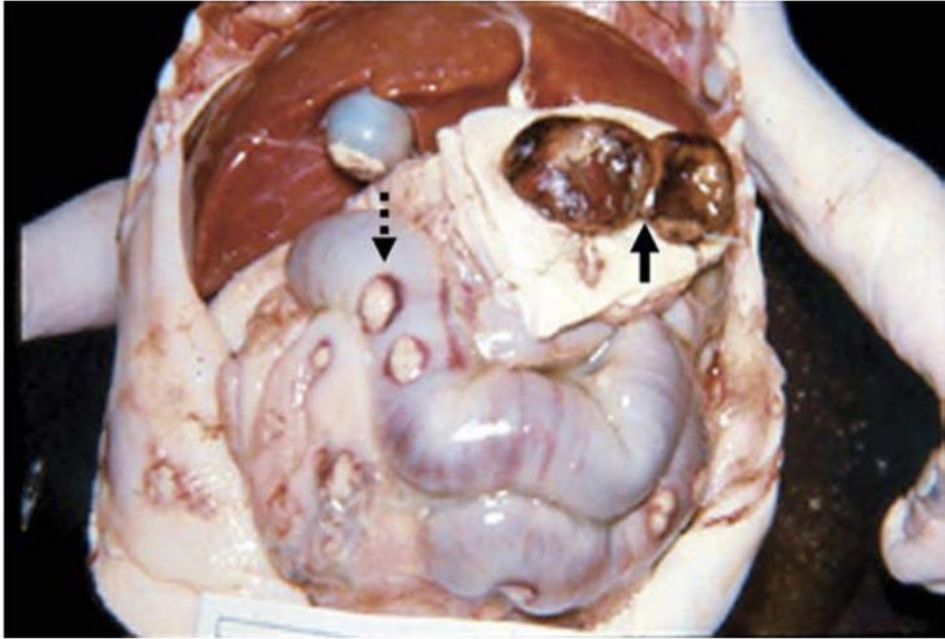
Table 34.1 Hirschsprung-Associated Enterocolitis (HAEC) Score

	Score
CRITERIA HISTORY	
Diarrhea with explosive stool	2
Diarrhea with foul-smelling stool	2
Diarrhea with bloody stool	1
Previous history of enterocolitis	1
PHYSICAL EXAMINATION	
Explosive discharge of gas and stool on rectal exam	2
Distended abdomen	2
Decreased peripheral perfusion	1
Lethargy	1
Fever	1
RADIOLOGY	
Multiple air–fluid levels	1
Dilated loops of bowel	1
Sawtooth appearance with irregular mucosal lining	1
Cut-off sign in rectosigmoid with absence of distal air	1
Pneumatosis	1
LABORATORY	
Leukocytosis	1
Shift to left	1
TOTAL	20

- Etiology
 - unknown
 - Stasis -> bacterial overgrowth such as Clostridium difficile or rotavirus
- Risk factor
 - Younger children
 - Longer segment disease
 - Trisomy 21
- Clinical
 - Fever, abdominal distension, diarrhea, leukocytosis , intestinal edema on an abdominal film
- Management
 - Nasogastric drainage, IV fluid, broad-spectrum antibiotic , decompression of rectum

A score of 10 or higher was associated with a positive diagnosis of HAEC by an international panel of experts.¹⁰⁸

ENTEROCOLITIS



- Prevention
 - Routine irrigation
 - Long-term metronidazole or probiotic agent
- Most resolve after first 5 years

HIRSCHSPRUNG-ASSOCIATED INFLAMMATORY BOWEL DISEASE

- Small group develop condition resembling Crohn disease
- Chronic inflammation, fistula formation
- Risk factor
 - Long-segment, trisomy 21
- Anti-inflammatory medication are ineffective

The background of the slide features a repeating pattern of medical icons in a light gray color. The icons include a clipboard with a checklist, a pill capsule, a circular pill with a diagonal line, and a plus sign. These icons are arranged in a grid-like pattern across the central white band of the slide. The text 'VARIANT OF HIRSCHSPRUNG' is centered in this band.

VARIANT OF HIRSCHSPRUNG

VARIANT HIRSCHSPRUNG DISEASE

- Intestinal neuronal dysplasia
- Hypoglanlionosis
- Internal sphincter achalasia
- "Ultra-short segment" HD
- Desmosis coli

1. INTESTINAL NEURONAL DYSPLASIA

- **Type A**
 - Absent sympathetic innervation of the myenteric and submucosal plexus
 - Hyperplasia of myenteric plexus
- **Type B**
 - Dysplasia of submucous plexus, thickened nerve fibers and giant ganglia
 - ↑ acetylcholinesterase staining, ectopic ganglion cell in lamina propria

2. HYPOGANGLIONOSIS

- Rare form of dysganglionosis
- Sparse and small ganglia, usually in distal bowel
- Abnormalities in acetylcholinesterase distribution
- Management
 - Resect abnormal colon and perform pull-through procedure

Immature ganglion (seen in preterm) -> self limit

3. INTERNAL SPHINCTER ACHALASIA

- Normal ganglion cell but lack RAIR
- Clinical
 - Obstructive symptoms
 - Severe constipation
- Diagnosis
 - Anorectal manometry
 - Rectal biopsy
- Management
 - Bowel management regimen
 - Anal sphincter myectomy
 - Botulinum toxin, nitroglycerine paste, topical nifedipine

4. ULTRASHORT SEGMENT HIRSCHSPRUNG DISEASE

- Aganglionic segment less than 3-4 cm (1-2 cm)
- Hypertrophic nerves, abnormal cholinesterase staining may be absent
- Management
 - Simple anal sphincter myectomy VS Excision of aganglionic segment and pull-through reconstruction

5. DESMOSIS COLI

- Chronic constipation associated with total or focal lack of connective tissue net in the circular and longitudinal muscles

REFERENCES

