

GI/Surgical Conference

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Pediatric Surgery

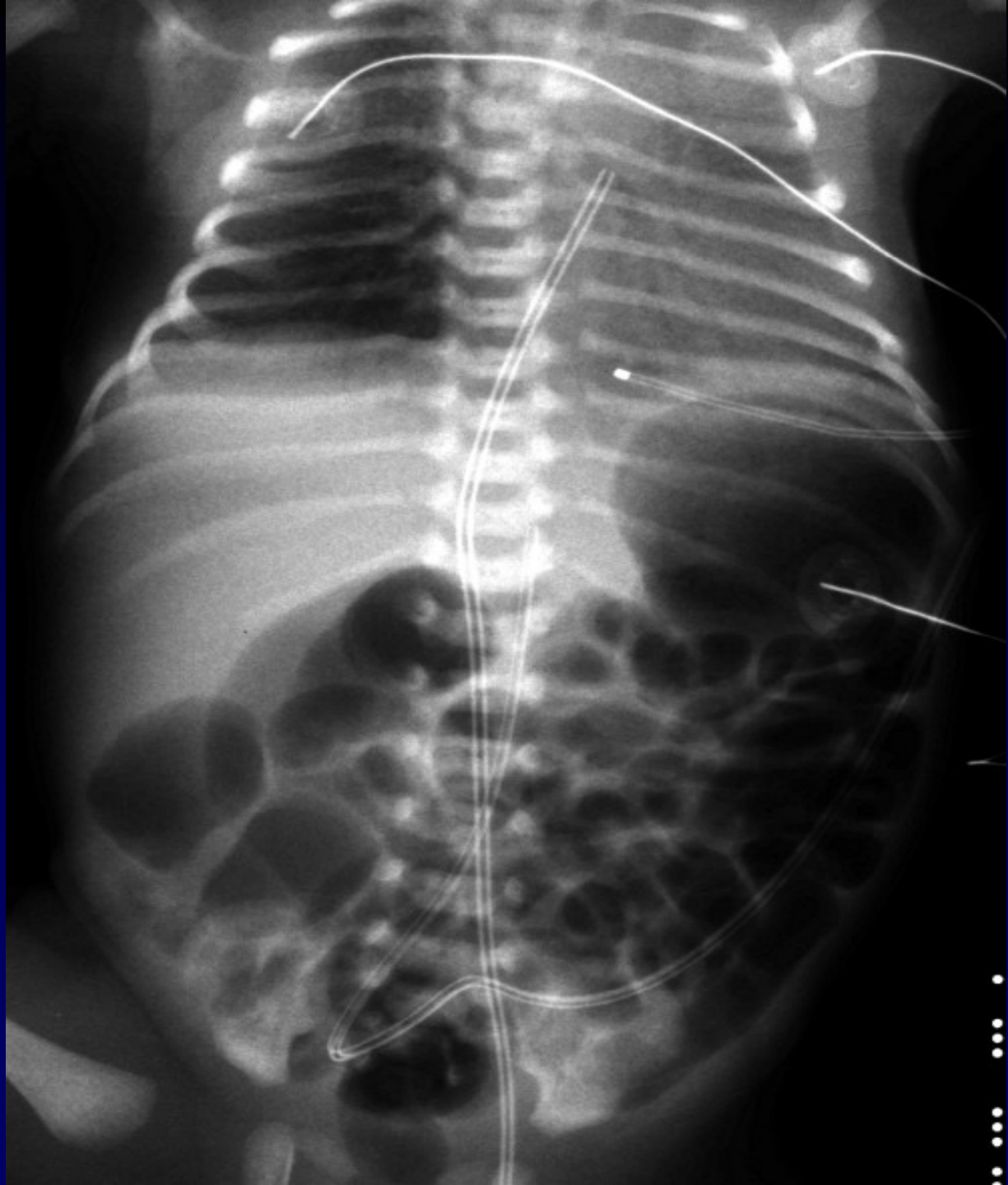
April 5, 2001

Case Presentation

- 32week premie with history of breech presentation and placental abruption
- Birth weight of 3lbs 10 oz
- On day of life number 3 baby boy became tachycardic, lethargic, and febrile
- Diagnosed with neonatal sepsis; treated in ISCU with IV Ampicillin/ Gentamicin
- Baby kept NPO with OGT & TPN

Case Presentation

- Examination revealed normal bowel sounds, a moderately distended, soft abdomen
- OGT output was around 40cc/day
- Leukocytosis resolved over the next week, but abdominal distension remained
- During 3rd week of life baby boy was clinically stable, but still with only rare smear of meconium stool



Case Presentation

- Small bowel obstruction was diagnosed and a surgical consultation was placed
- A barium enema was obtained...

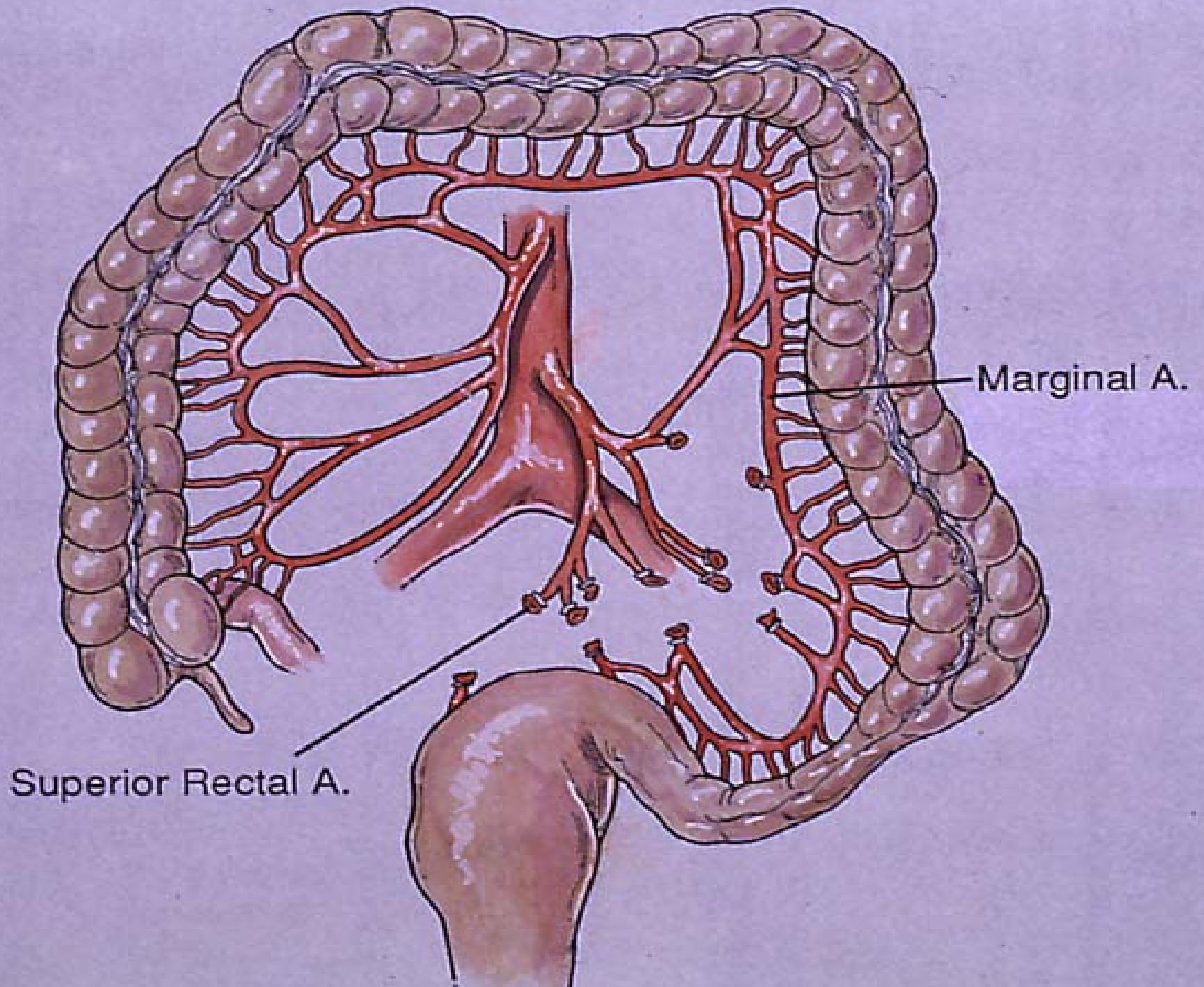




UTMB-GALVESTON

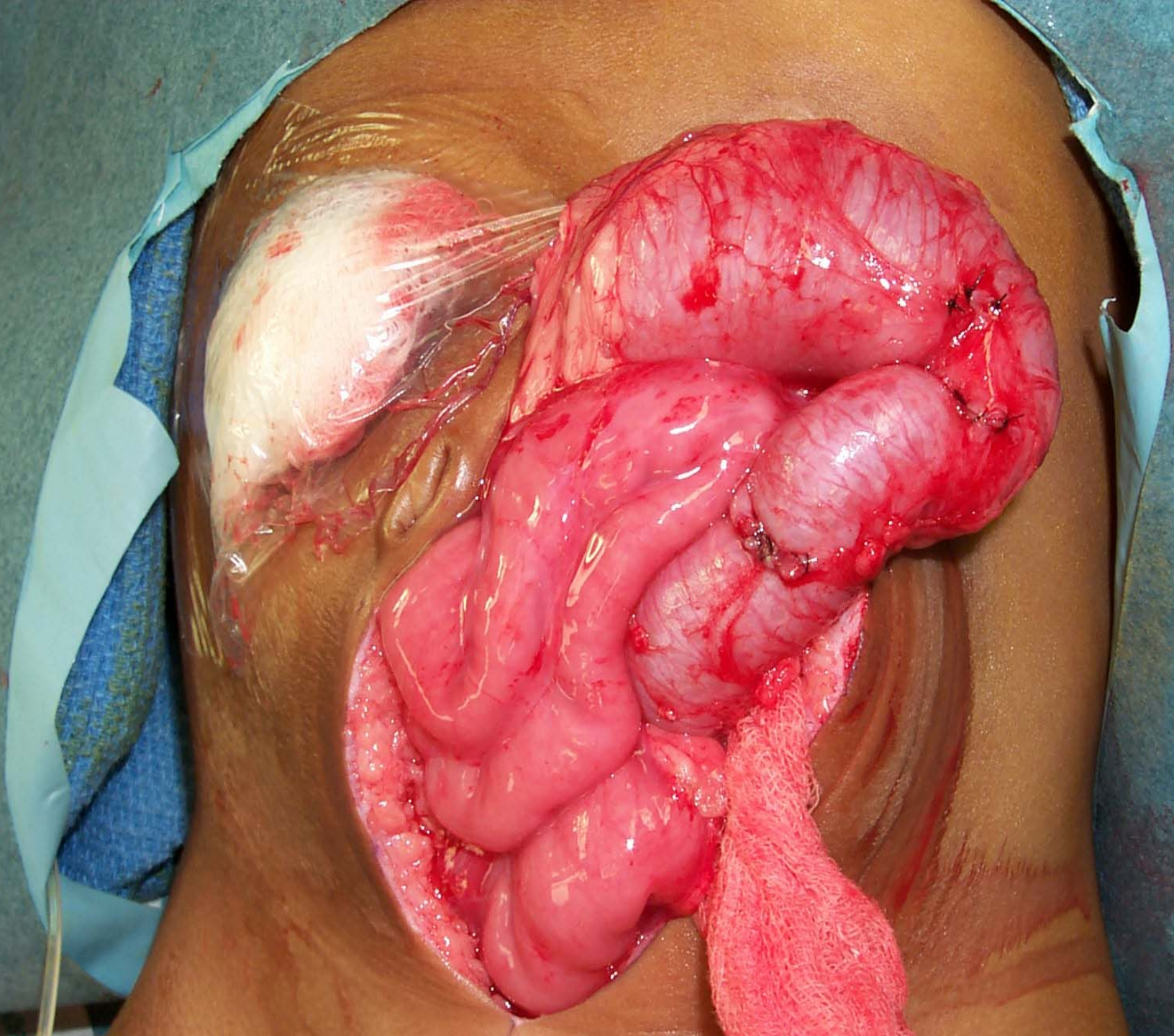
Hirschsprung's Disease

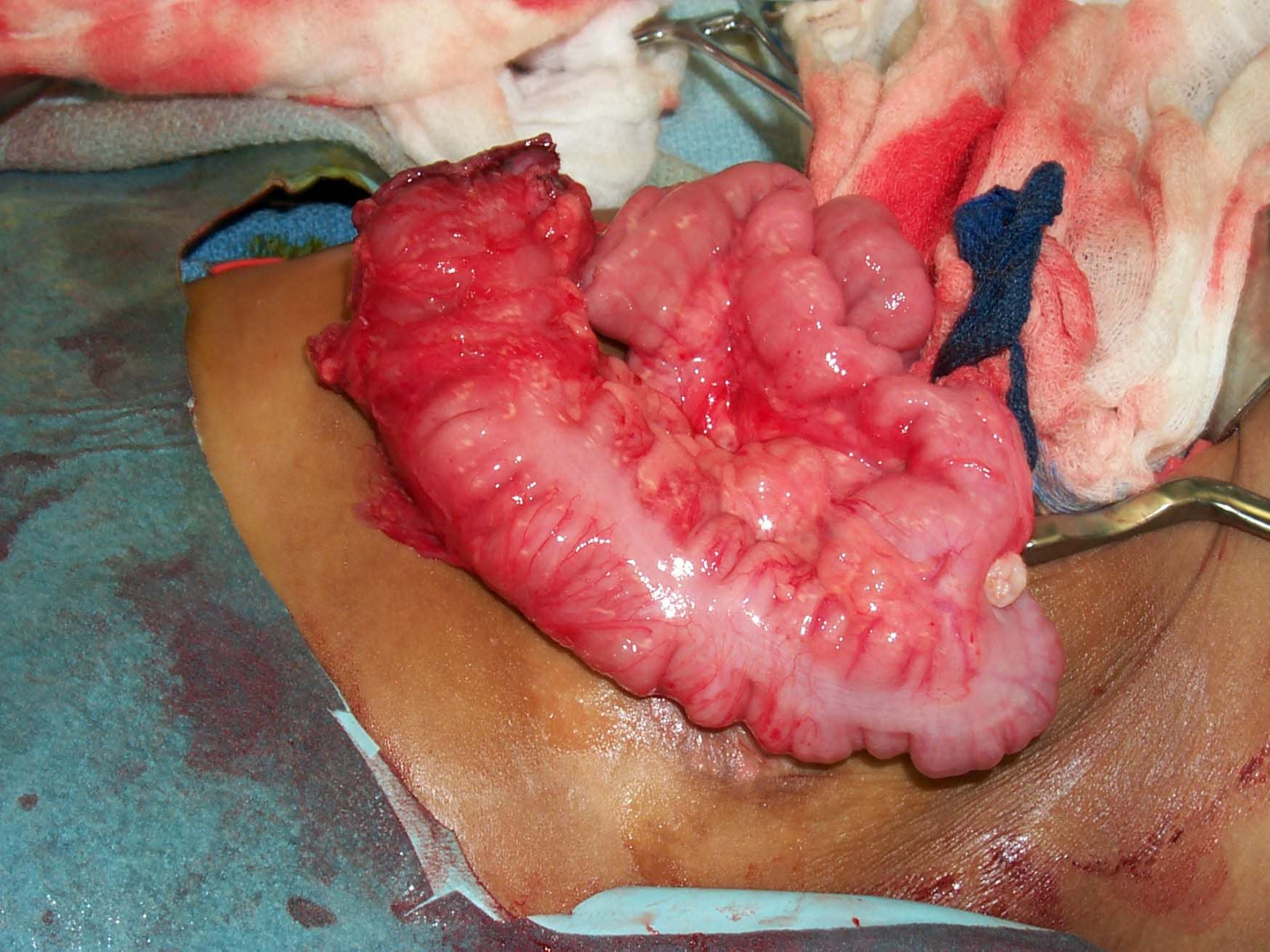
- Suction rectal biopsy confirmed the diagnosis and the baby was taken to the operating room for a temporizing procedure
- A colostomy was performed with the plan for a definitive operation -a colo-anal pull through procedure to be performed at 1 year
- the child now presents for this operation...

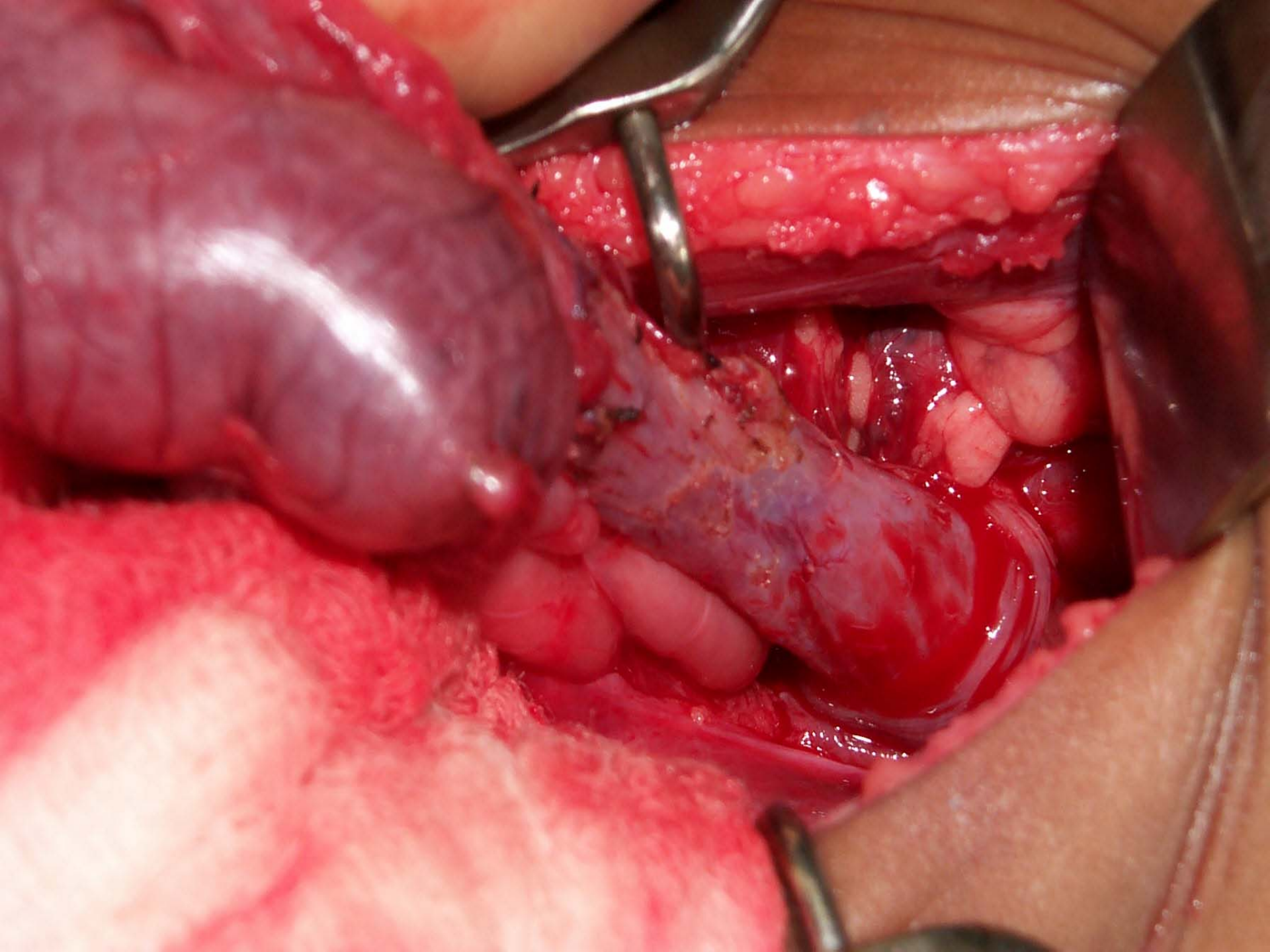


The Operation

(A Soave Colo-anal Pullthrough)











Hirschsprung's Disease

Steven W Williams, MD

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March 23, 2001

Hirschsprung's Disease

(Aganglionic Megacolon)

- *Definition:*
Absence of submucosal and myenteric ganglion cells in the distal alimentary tract which results in decreased motility in the affected segment.

History (Cont'd)

- 1691: Fredrick Ruysch 1st reported megacolon in autopsy of 5y/o girl dying of unremitting constipation
- 1886: Harald Hirschsprung classic description of megacolon in two infants
- 1901: Tittel noted absence of ganglia in bowel wall of 15mo with constipation
- 1940: Tiffen & colleagues proposed that proximally dilated segment result of distal pseudoobstruction

History (Cont'd)

- 1948: Orvar Swenson & Alexander Bill describe pathophysiology- aganglionic segment fails to relax during peristalsis
- 1949: Dr Swenson publishes 1st series of patients undergoing definitive operation
- 1956: Duhamel procedure devised to minimize complications of anterior rectal dissection

History (Cont'd)

- 1964: Soave modifies original endorectal pull through procedure, first described by Raviche and Sabiston, to be used in infants
- Present: Soave procedure utilizing laparoscopic technique is used most commonly for surgical treatment of Hirschsprung's disease at infancy

Background

- Incidence estimated at 1/6000 live births
- 80% of affected are male
- 8% have a family history
- Subsequent sibling risk at 5% (12% if first sibling has total gut involvement)
- Risk of affected child of pt is 2% if short segment or 30% if long segment

Associated Anomalies

- Multiple Endocrine Neoplasia IIA
- 8% of patients have Trisomy 21
- Various cardiac, CNS, GU, and GI tract anomalies are commonly found
- Can influence overall morbidity and mortality

Proposed Etiology

Embryology:

1. Precursor neuroblasts do not migrate to the affected segment
2. Neuroblasts are present but do not mature
3. Cells develop, but then degrade secondary to microenvironmental effects

Genetics:

1. Causative gene linked to chromosome 10
2. Associated with RET proto-oncogene (codes for a tyrosine kinase receptor molecule)

Diagnosis

Clinical Presentations:

- Usually diagnosed in the 1st year of life (mean age of 10 mos)
- Neonatal Signs:
 - abdominal distension (65%)
 - emesis (58%)
 - failure to pass meconium in first 24hrs (58%)
- 2nd only to NEC as the common cause of neonatal bowel obstruction





Diagnosis (Cont'd)

Radiographic findings:

- Plain films reveal dilated loops of bowel extending to the functional obstruction
- Barium enema usually able (80%) to see transition zone (no balloon, no prior rectal, delayed 24hr film)

Diagnosis (Cont'd)

Manometric findings:

- Increased resting tone of anal sphincter
- Sphincter does not relax with increased rectal ampullary pressure (90% diagnostic)
- Rarely used in clinical settings because of difficulty with data interpretation

Differential Diagnosis

Mechanical Obstructions:

- meconium ileus
- distal ileal or colonic atresias (stenosis)
- imperforate anus

Functional Obstructions:

- - prematurity
- small left colon syndrome
- meconium plug syndrome
- sepsis; electrolyte imbalance;hypothyroidism

Rectal Biopsy

- Necessary for definitive diagnosis
- Full thickness rectal biopsy from 1.5cm proximal to dentate line
- Suction rectal biopsy:
 - the most common method used in infants
 - does not require general anesthesia; however, only assess meissner plexuses
 - frozen section not recommended

Complications

- Neonatal bowel obstruction with risks for perforation
- Enterocolitis:
 - Clinical presentations in 6-20%
 - Significant mortality 30%
 - Risk factors: Down's syndrome, delay in diagnosis of Hirschsprung's > 1 week age
 - Signs: abdominal distension, diarrhea, fever, hematochezia, sepsis, explosive foul-smelling diarrhea after rectal exam

Enterocolitis

Treatment:

- Must recognize presenting clinical symptoms early to avoid complications
- Early decompression with rectal tube
- Operative decompression may be necessary
- IV antibiotics, fluids
- Vancomycin po as 1/3 of cases shown to be *Clostridium difficile* positive

Extent of Disease

- Always involves anal sphincter and extends proximally
- 75% rectum and sigmoid (short segment)
- 11% descending colon (long segment)
- 4% splenic flexure
- 2% transverse colon
- 8% total colon & proximal (total colonic)
- Longer segment involved in girls

Ultrashort Segment Hirschsprung's

- Aganglionosis extends only few centimeters proximal to dentate line
- Average age of diagnosis 6yo
- No radiographic transition zone
- Absent anorectal reflex by manometry
- Open, full thickness biopsy required
- Anorectal Myectomy

Treatment

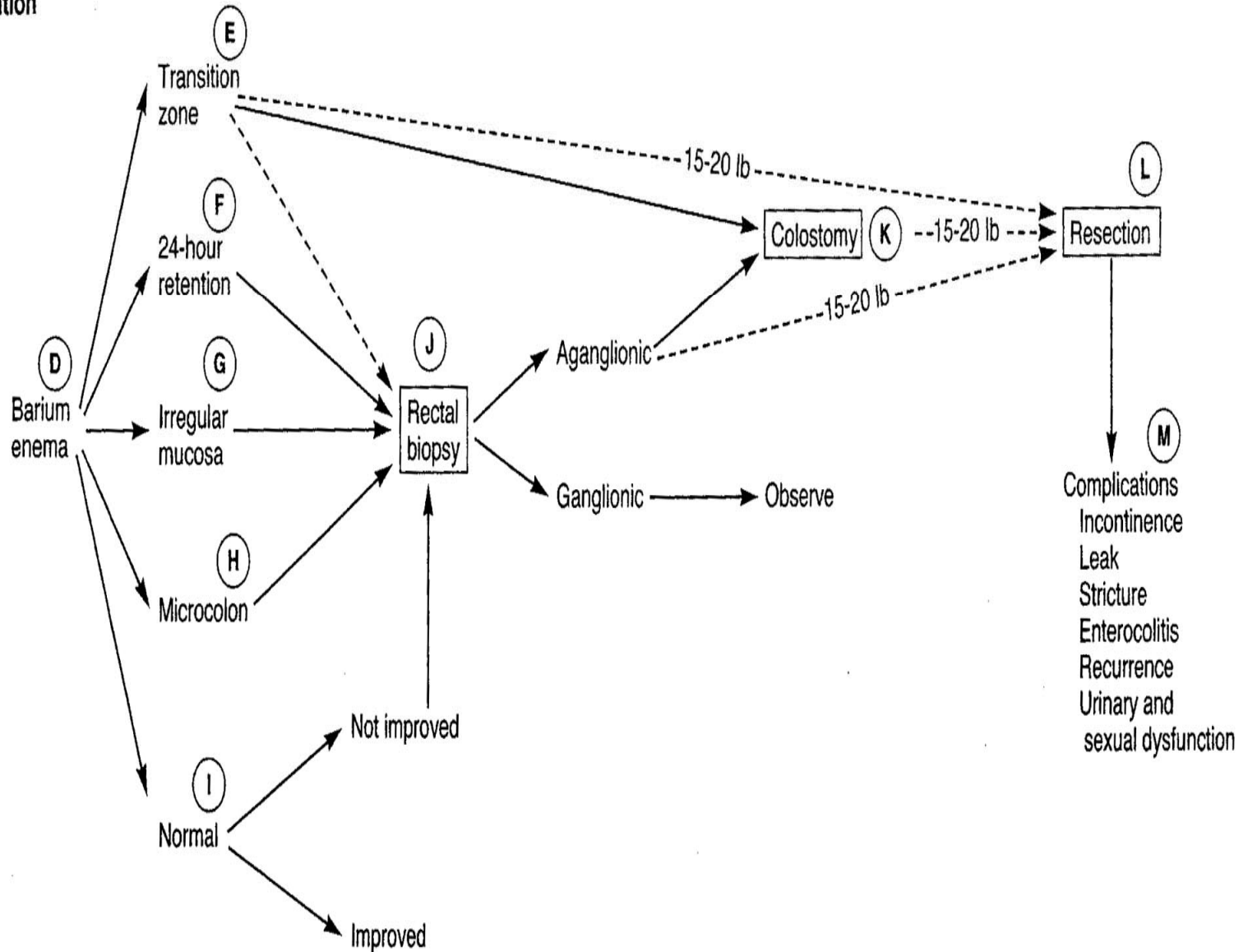
- Surgical treatment of Hirschsprungs involves removal or bypass of the affected segment
- Standard has been diverting colostomy and definitive operation when child > 10kg
- “leveling colostomy” -serial biopsies done to determine correct level
- Rectal irrigation regimen- requires bulk reducing diet & responsible caregivers

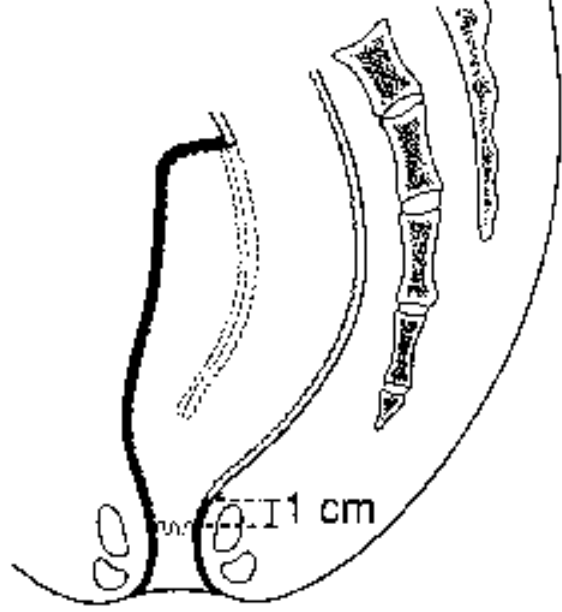
History and physical examination

- Child or infant (A)
- Delayed meconium
- Abdominal distention
- Poor feeding
- Vomiting, diarrhea

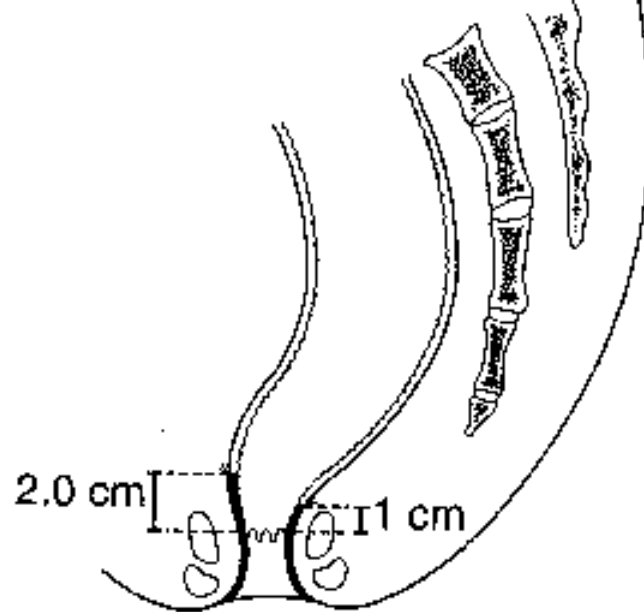
HIRSCHSPRUNG'S DISEASE*

- Lab (C)
- Abdominal x-ray

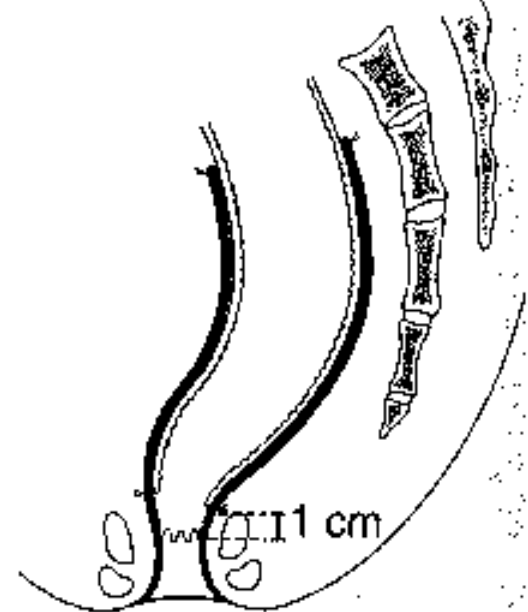




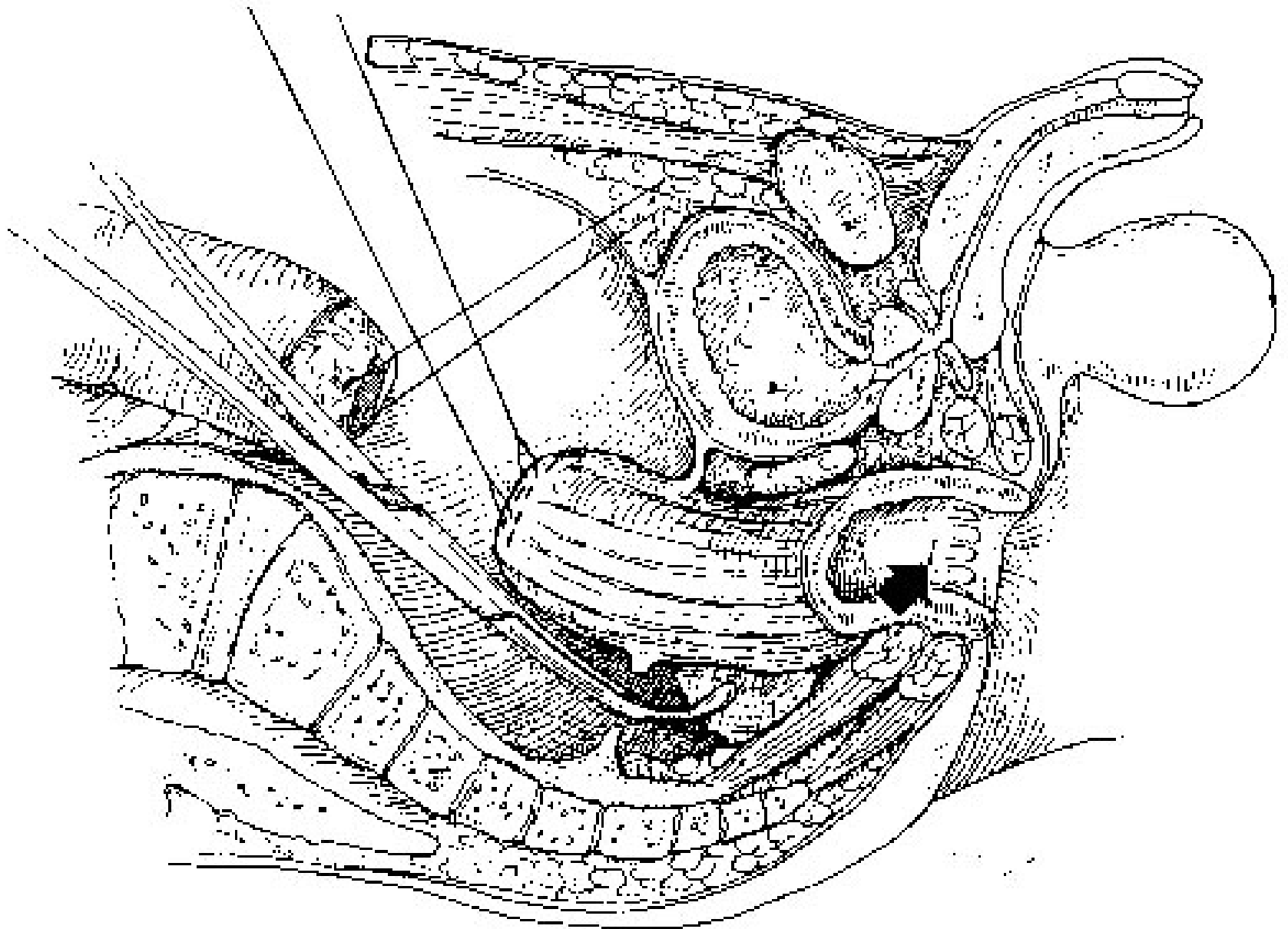
Duhamel

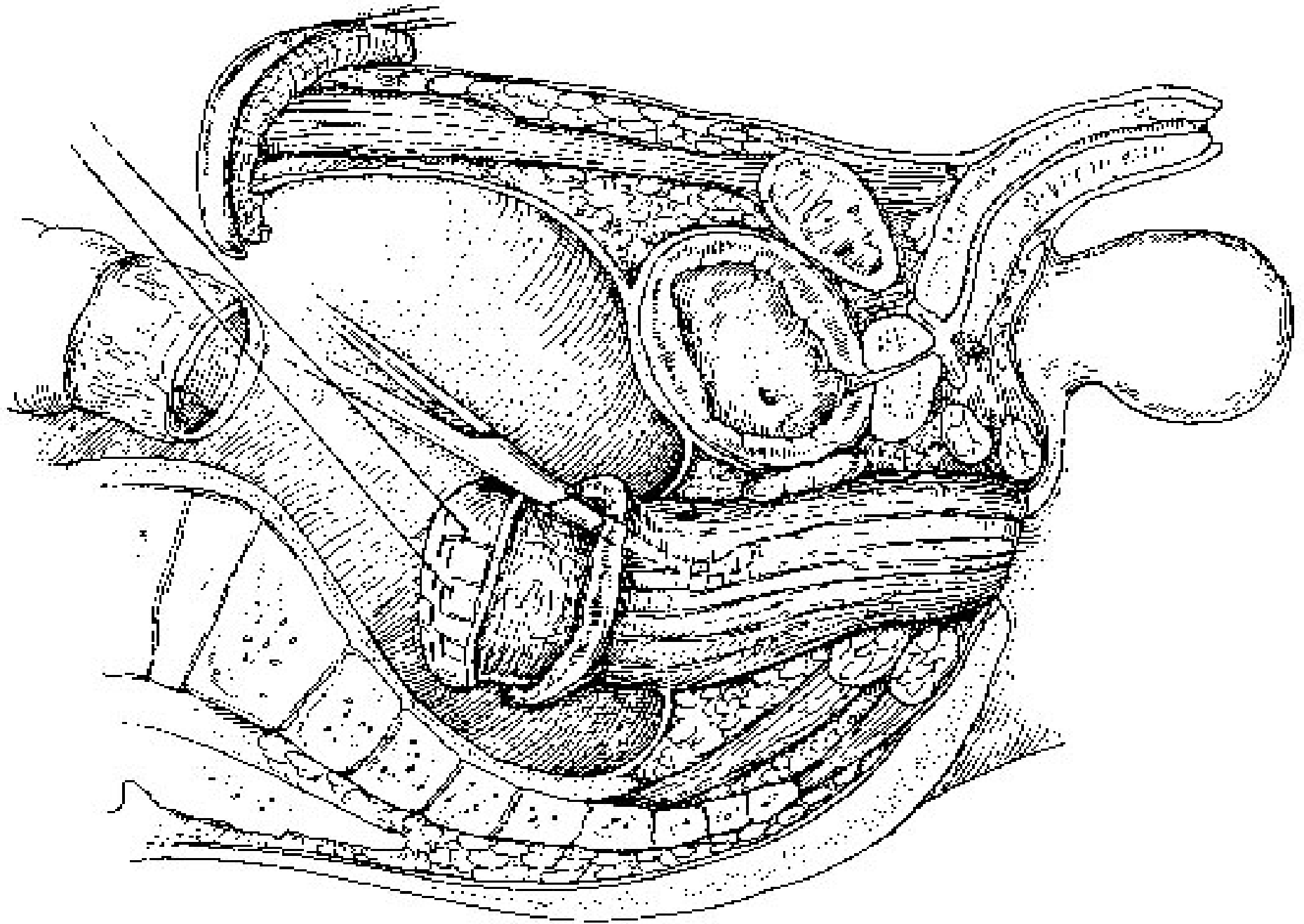


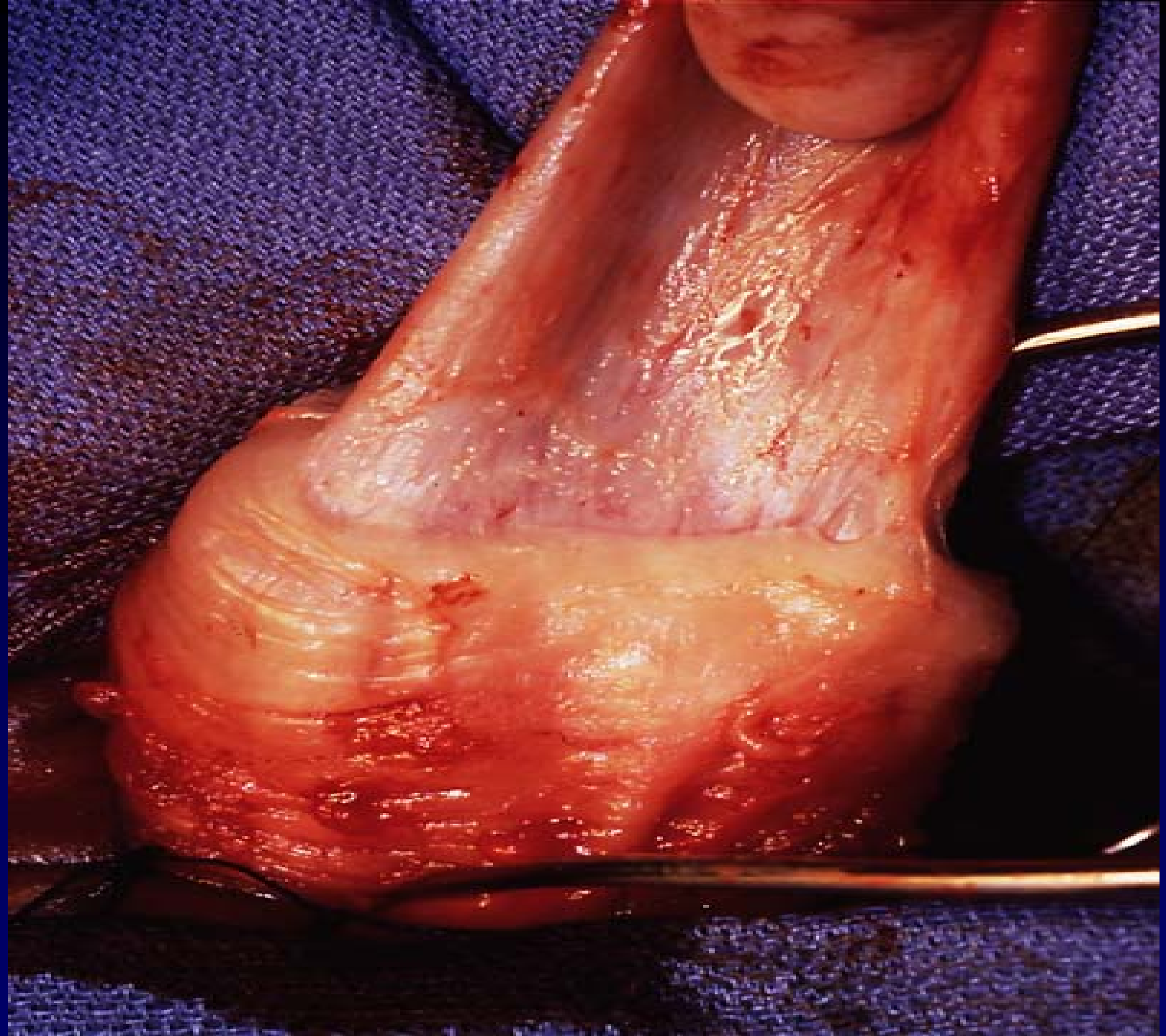
Swenson



Soave (ERPT)

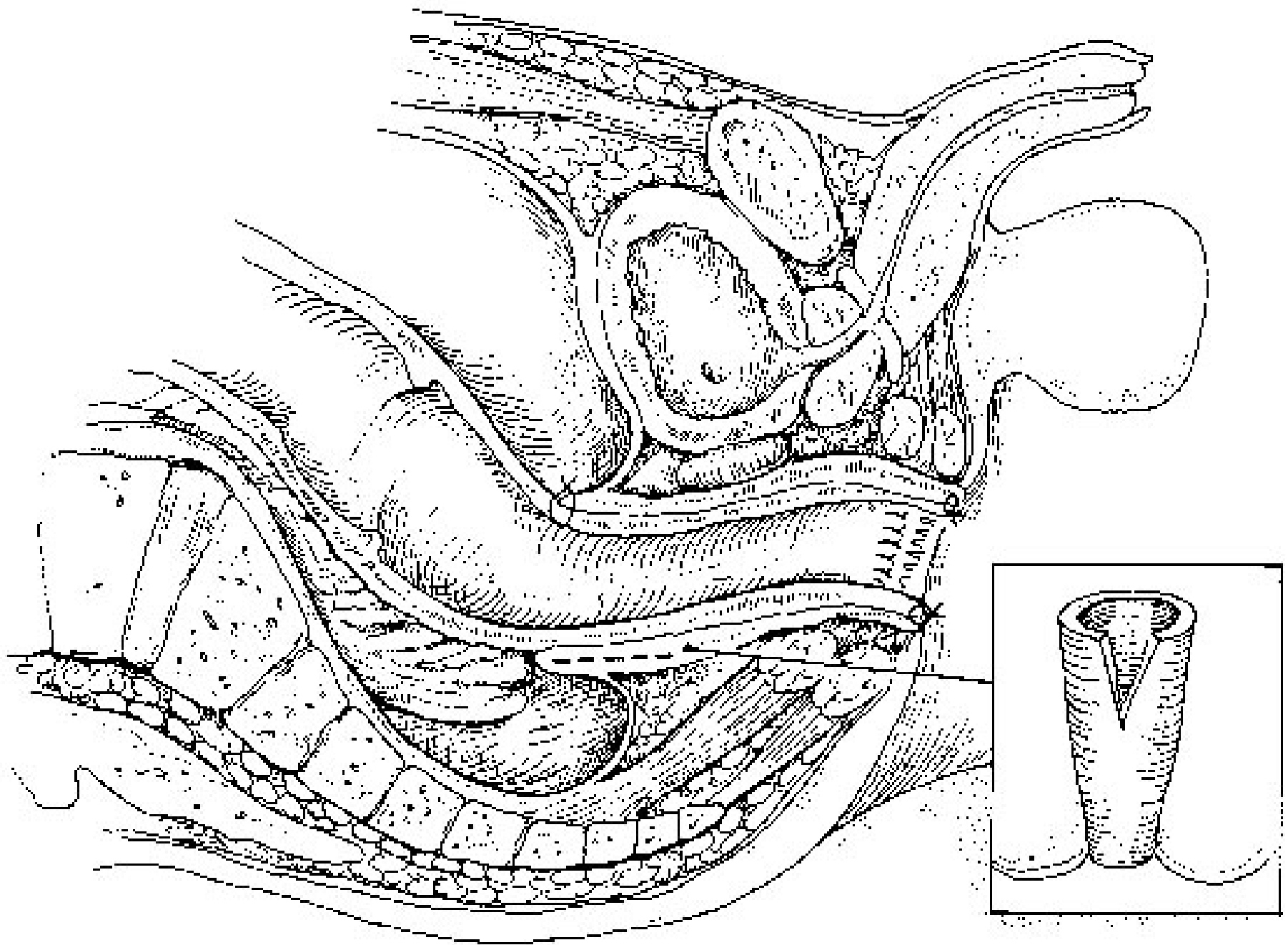


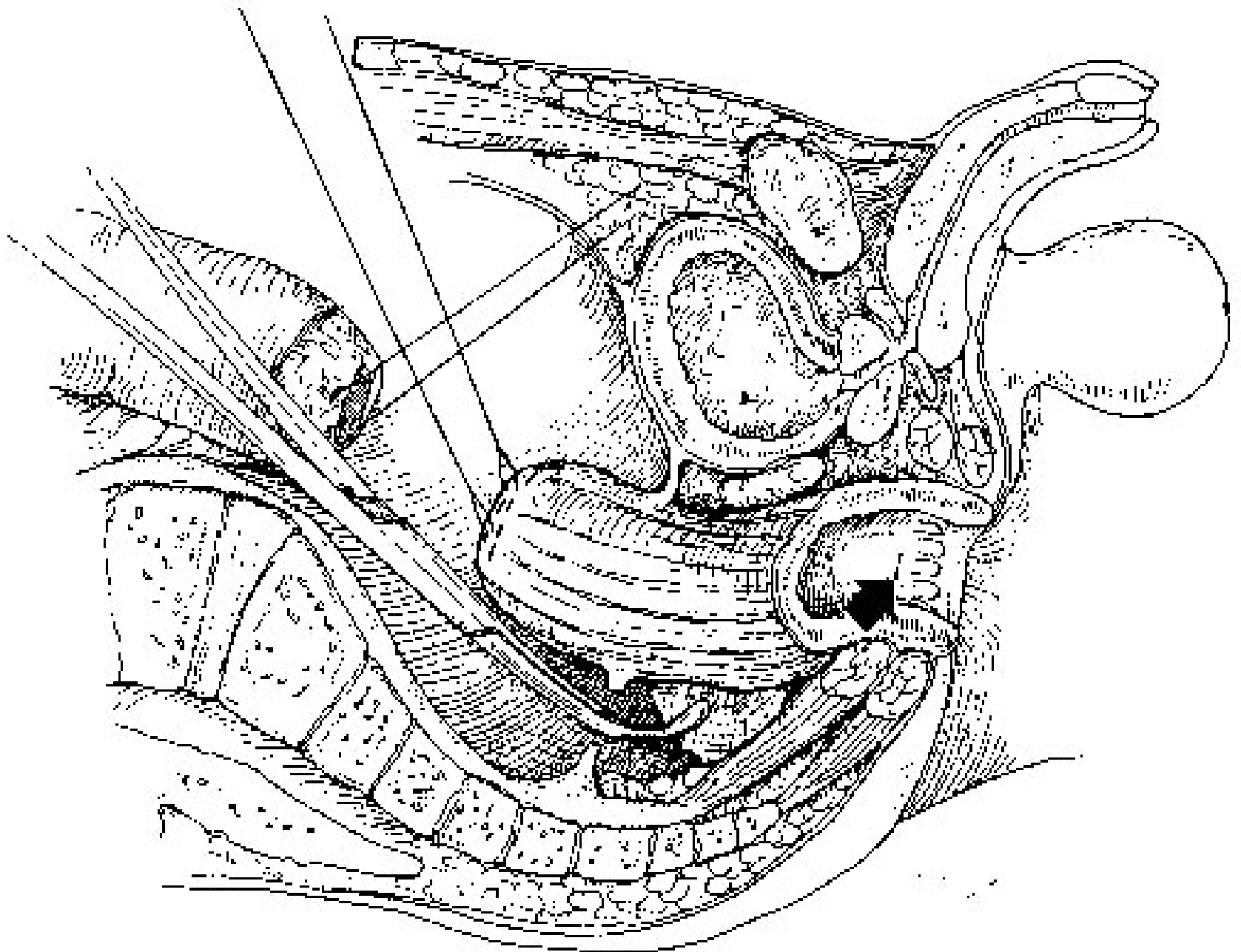


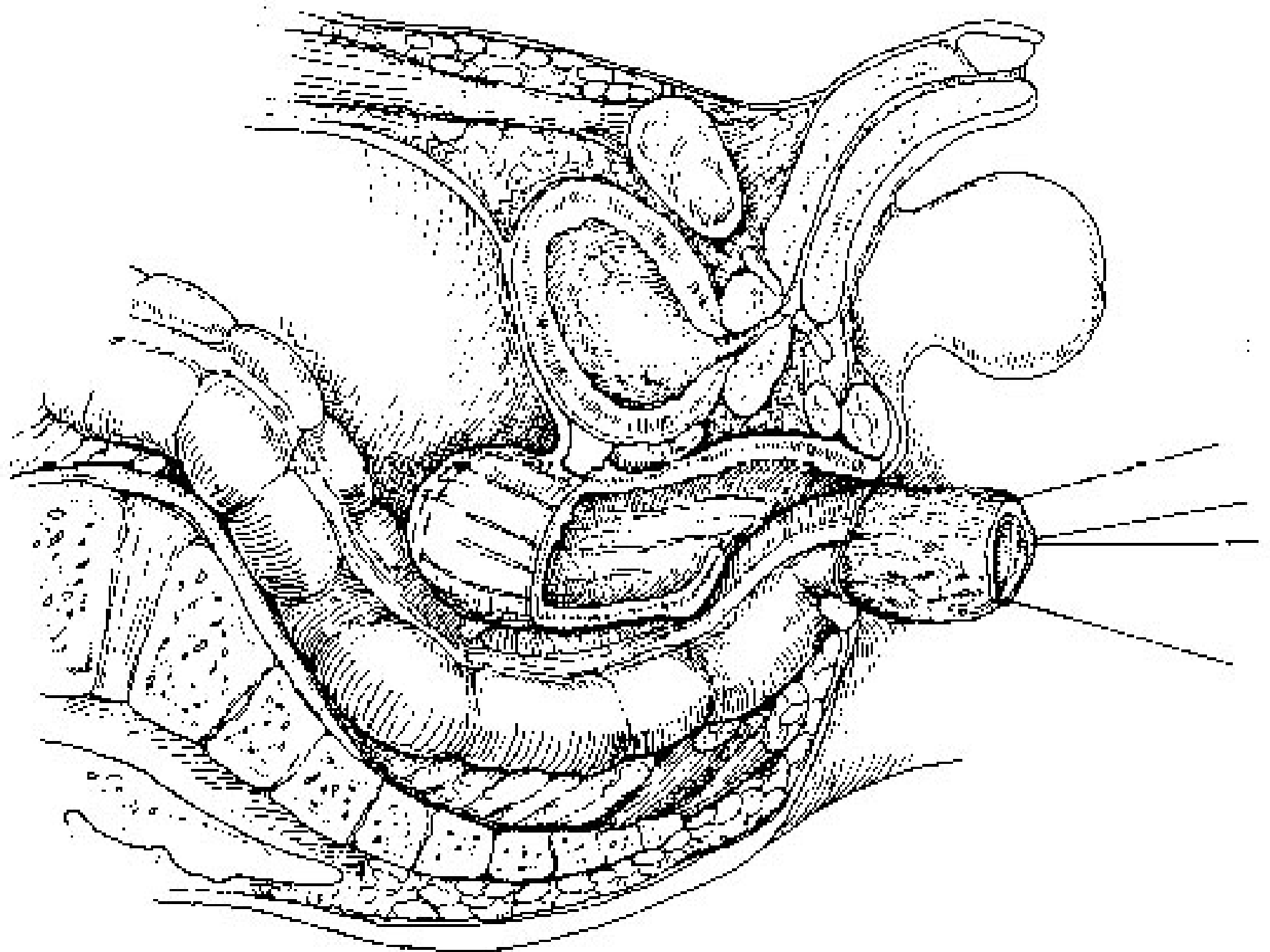


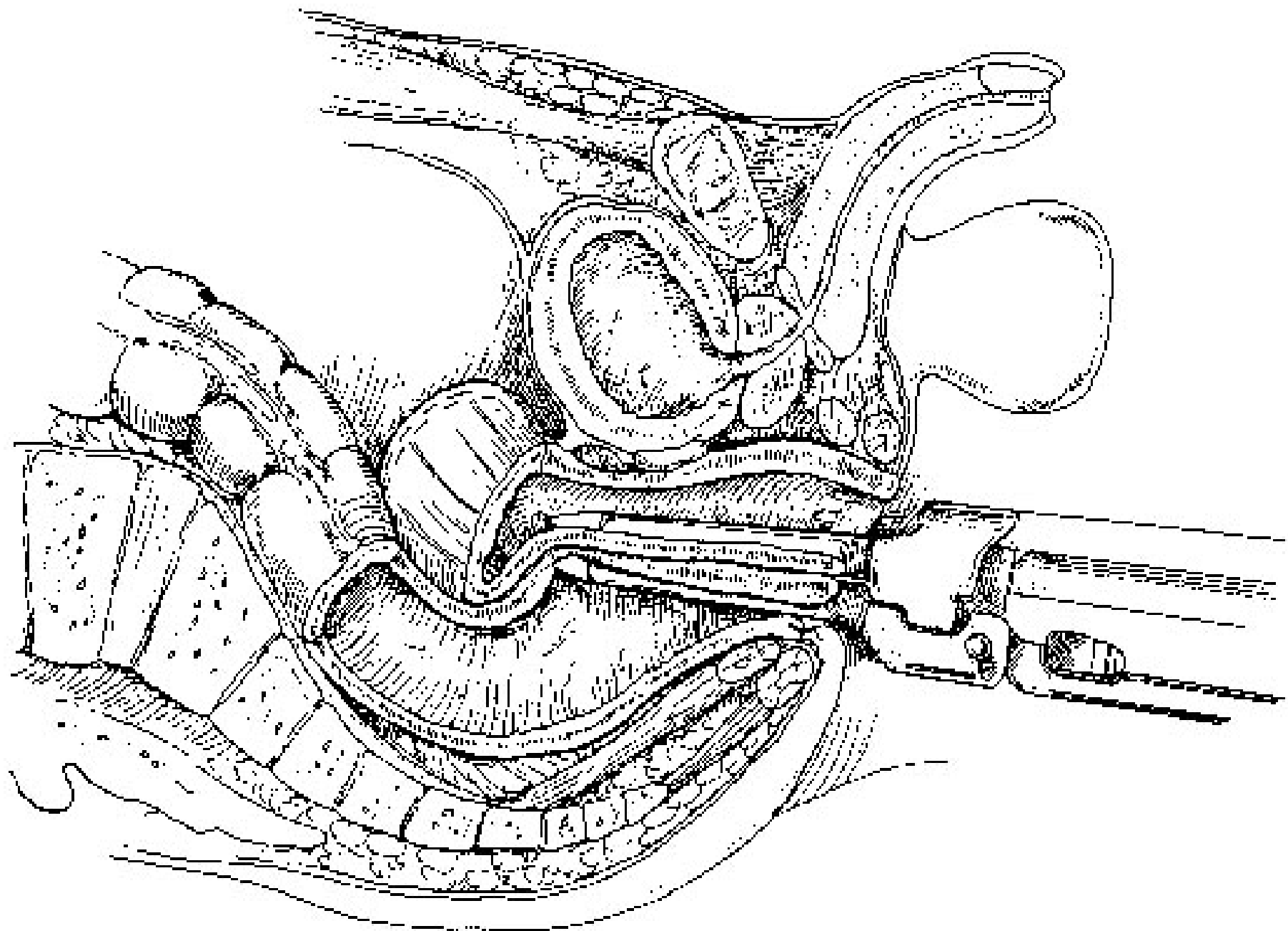


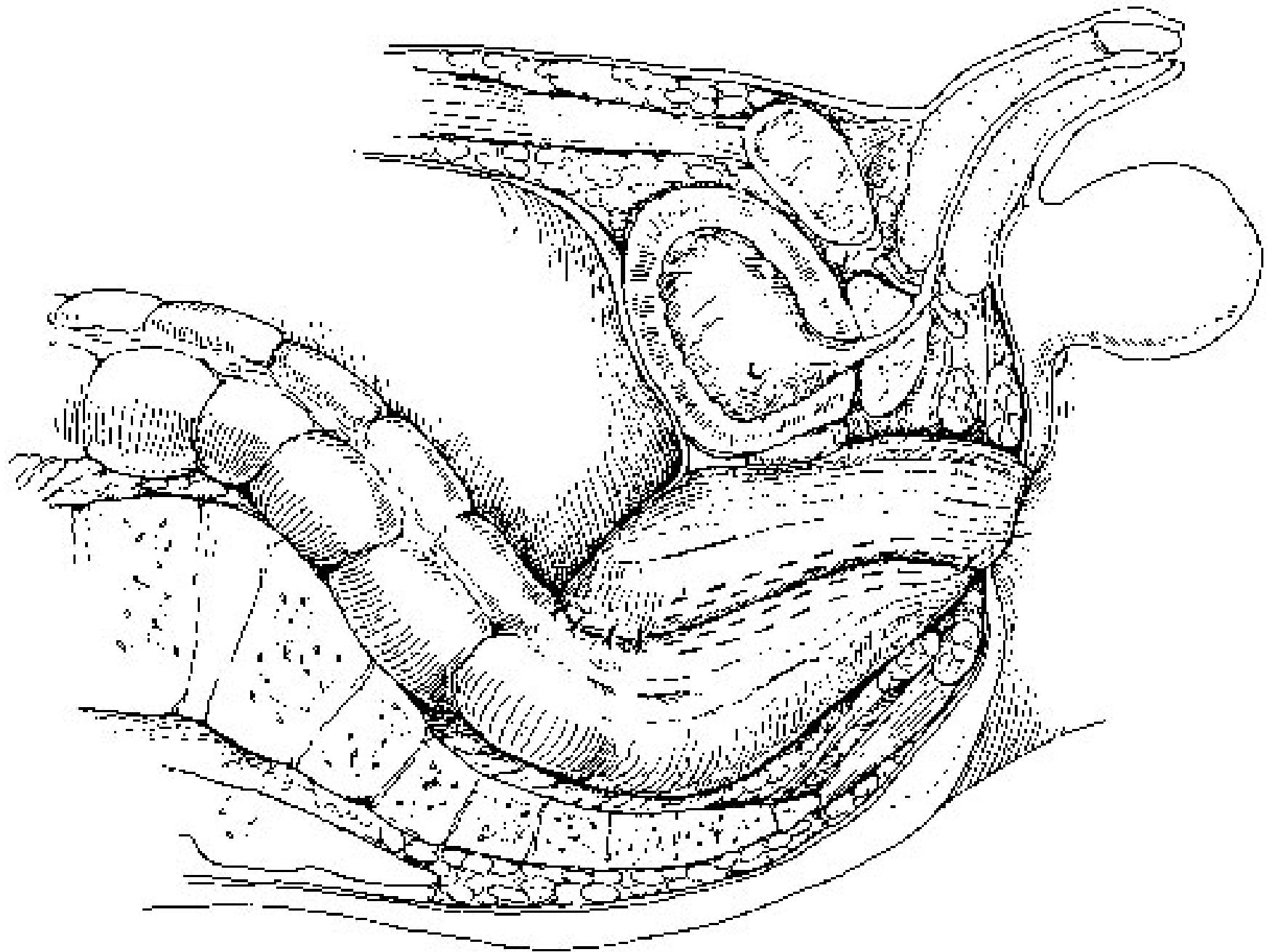
- aganglionic colon resection
- resect entire transition zone
- frozen section confirmation of ganglion cells at anastomotic site
- posterior myotomy











Definitive Operation

- No prospective studies compare 3 options
- **Swenson**: complete excision of aganglionic segment, but possible increase in postop enterocolitis & anal stricture rate
- **Duhamel**: technically easier & best redo operation, but increased constipation
- **Soave**: less anterior nerve injury, but higher diarrhea and incontinence

Postoperative Management

- Loperamide used as adjunct to decrease motility with cases of frequent loose stools
- Metronidazole used to alter flora
- Serial dilations for anal strictures
- Chronic constipation- barium enema may define anatomy, dilations, manometry, biopsy, redo pull through rare cases