# Overview of Anorectal Malformation

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# Disclosures

- I have no disclosures.

# Objectives

- Demonstrate understanding of the spectrum of anorectal malformation.
- Demonstrate understanding of the severity of sequelae associated with each anorectal malformation.
- Describe the workup for a neonate with anorectal malformation.
- Identify the indication of primary versus secondary surgical repair for anorectal malformation.

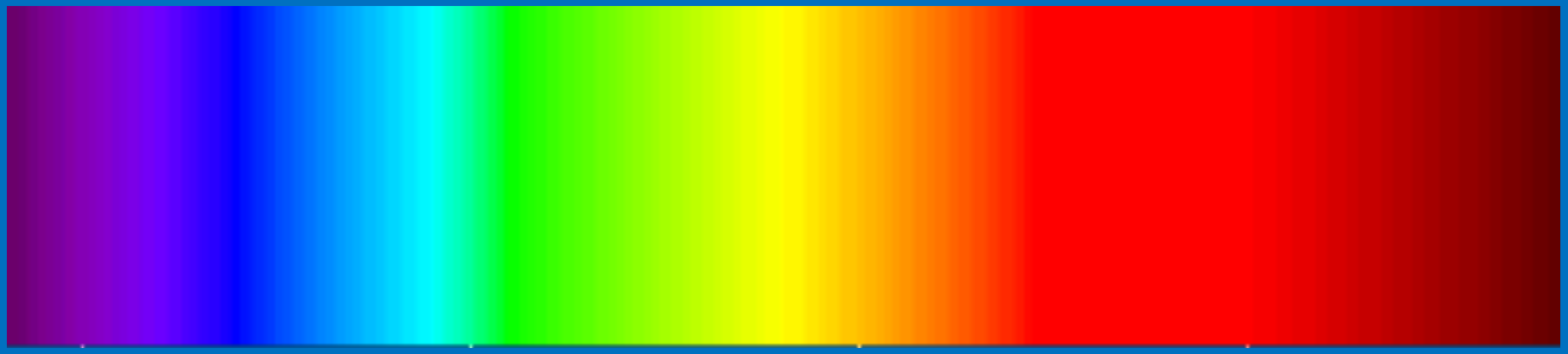
# Objectives

- Identify important aspects of long-term management of anorectal malformation.
- Summarize the rationale behind a multidisciplinary approach when caring for patients with anorectal malformation.

# Spectrum of Anorectal Malformations

Perineal Fistula

Cloacal Exstrophy



# Types of Anorectal Malformation

| Level of severity | Male | Female |
|---|---|---|
| Least severe/best prognosis | Perineal fistula | Perineal fistula |
| | Anorectal malformation without fistula | Anorectal malformation without fistula |
| | Recto-bulbar fistula | Recto-vestibular fistula |
| | Recto-prostatic fistula | Recto-vaginal fistula |
| | Recto-bladder neck fistula | Cloaca |
| Most severe/worst prognosis | Cloacal exstrophy | Cloacal exstrophy |
| | | |

# Identifying Anorectal Malformation

- Perform a thorough newborn exam
- Observe for meconium passage through a normally placed anus
- Listen to parents!

A newborn workup for a patient born with an anorectal malformation should include which of the following?

A. Echocardiogram
B. Babygram and NG tube
C. Renal ultrasound
D. Sacral radiographs
E. Spinal ultrasound
F. A, B, D, and E
G. All the above

# Newborn Screening for Patients with ARM

- Echocardiogram- 30% patients have cardiac anomalies
- NG tube and babygram- 8% have esophageal atresia and 3% duodenal atresia
- Renal ultrasounds- 50% have associated urologic conditions

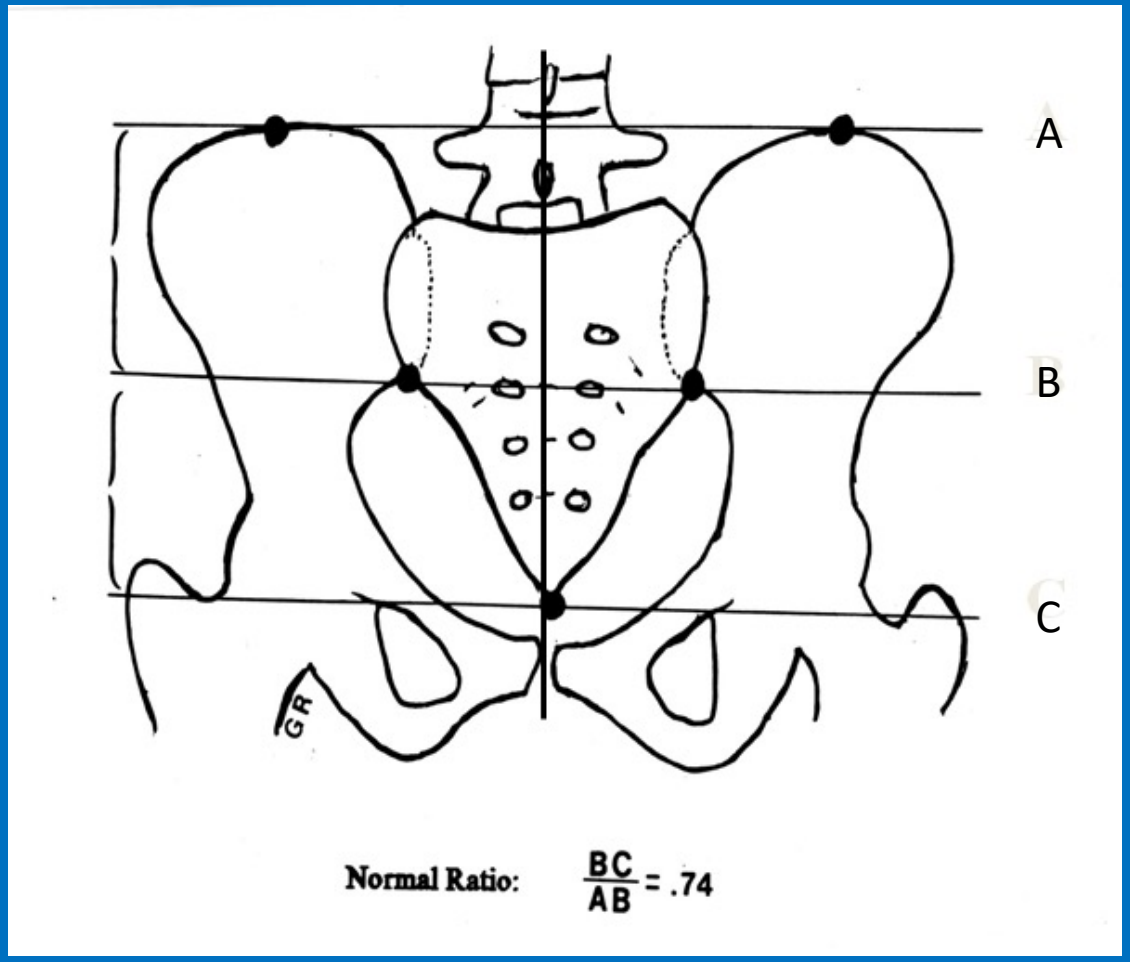
# Newborn Screening for Patients with ARM

- Spinal X-ray- AP and lateral – to rule out hemisacrum and calculate the sacral ratio
- Spinal ultrasound- to rule out tethered cord – present in 25%
  - Unless >3 months old, then get MRI

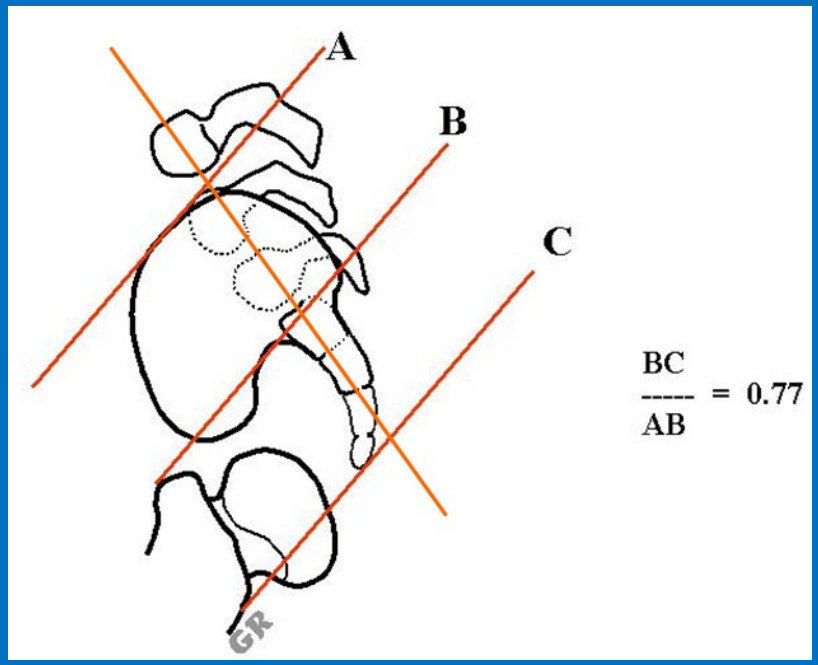
# Bowel Control Factors

- Tethered cord
- Qualities of the sacrum
- Presence of presacral mass
- Type of malformation

# Sacral Ratio AP Radiograph

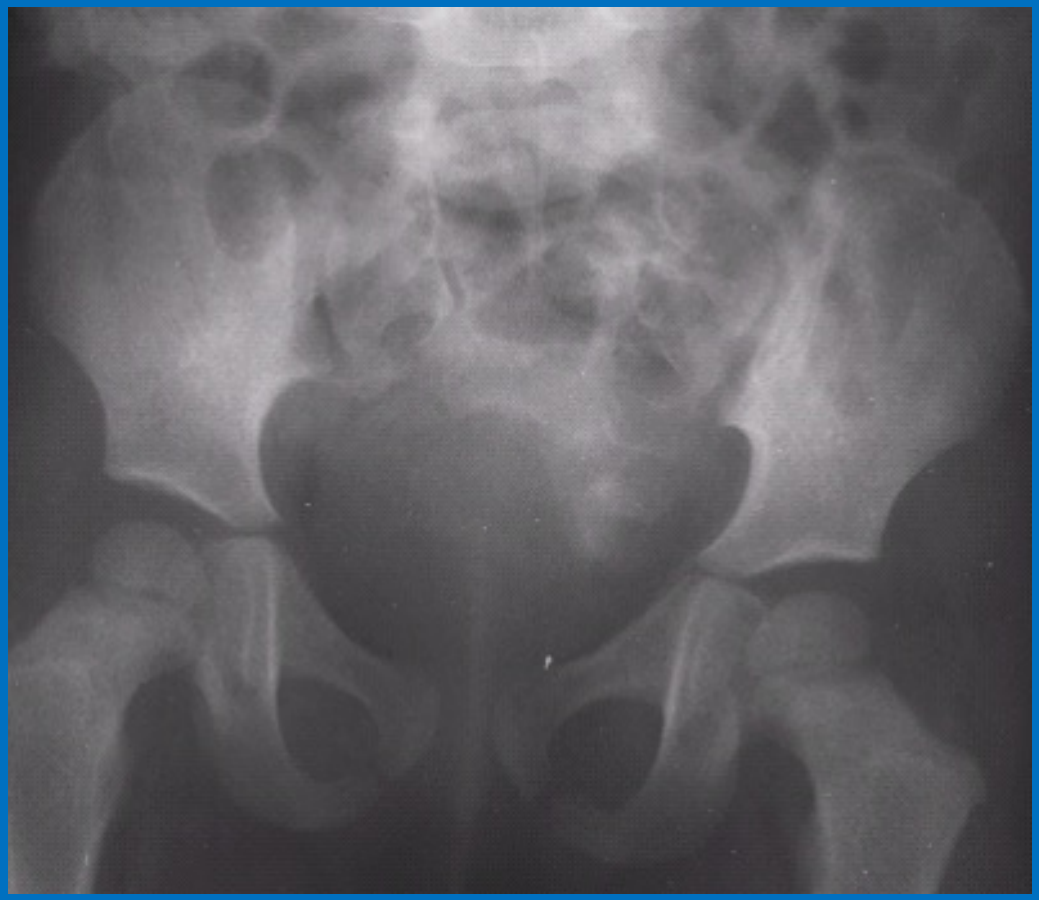


# Sacral Ratio Lateral Radiograph




- Sacral ratio of >0.7 or greater represents good prognosis for bowel control.
- Ratio 0.41- 0.69 are considered undetermined.
- Ratio of < or equal to 0.4 represents poor prognosis for bowel control.

# Based on this radiograph what further imaging would you recommend ?

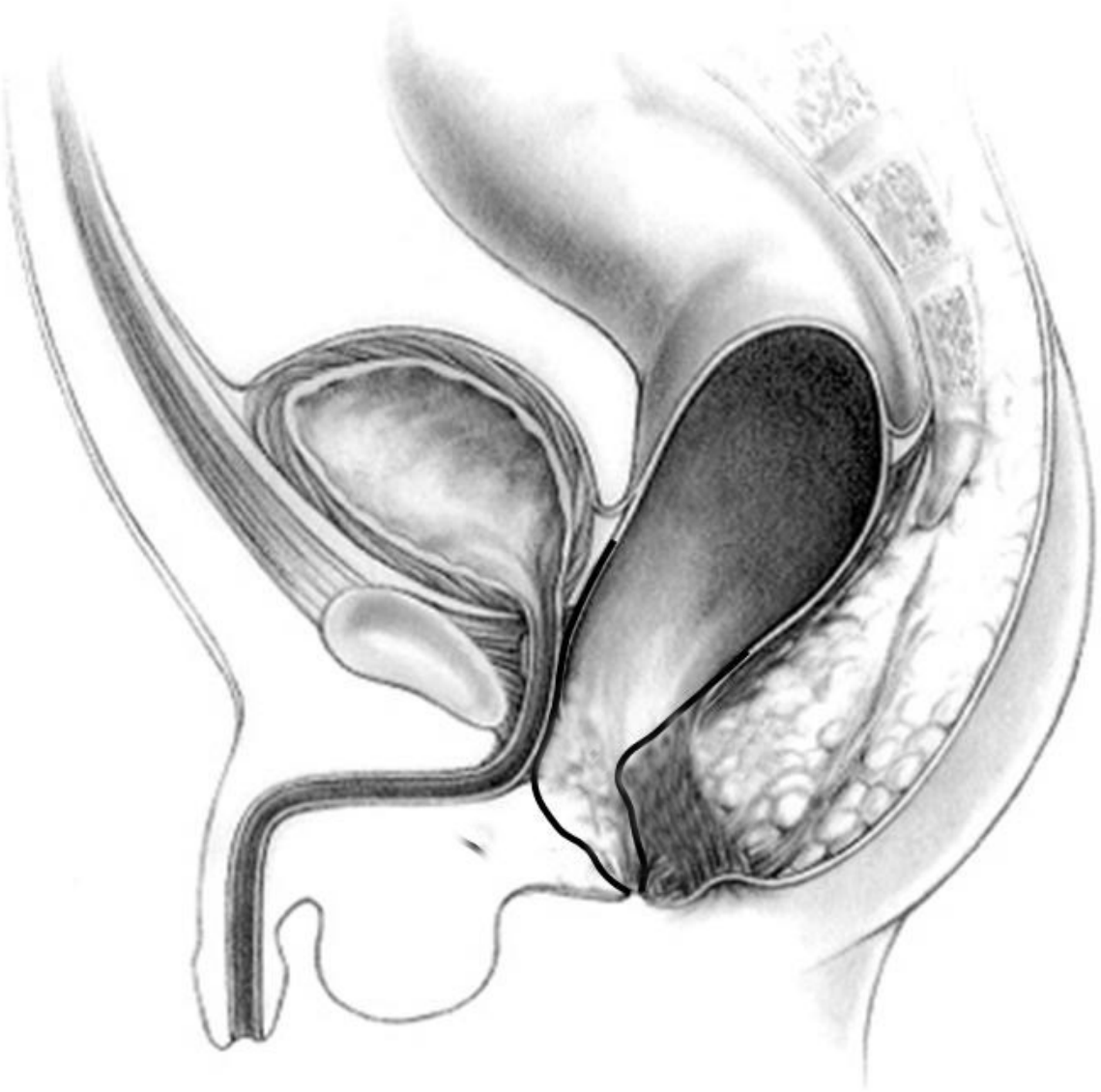


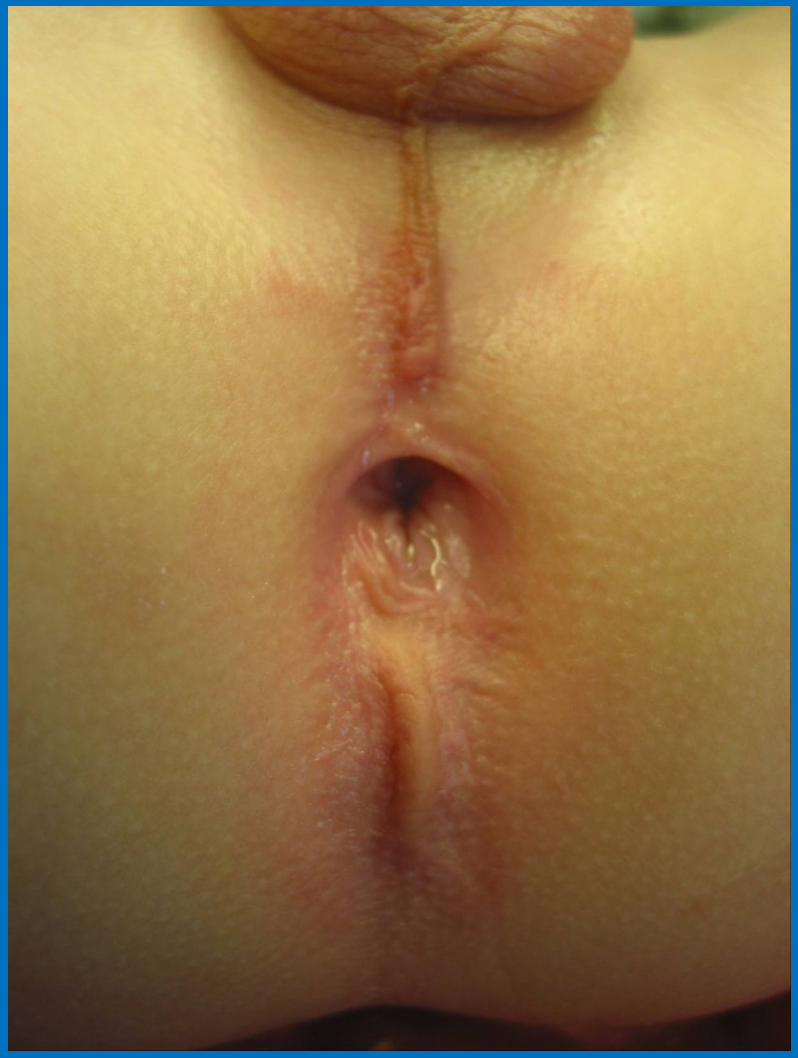
A. No further imaging this is a normal sacral radiograph
B. Ultrasound of the spine
C. CT scan of the abdomen
D. MRI pelvis
E. I don’t know

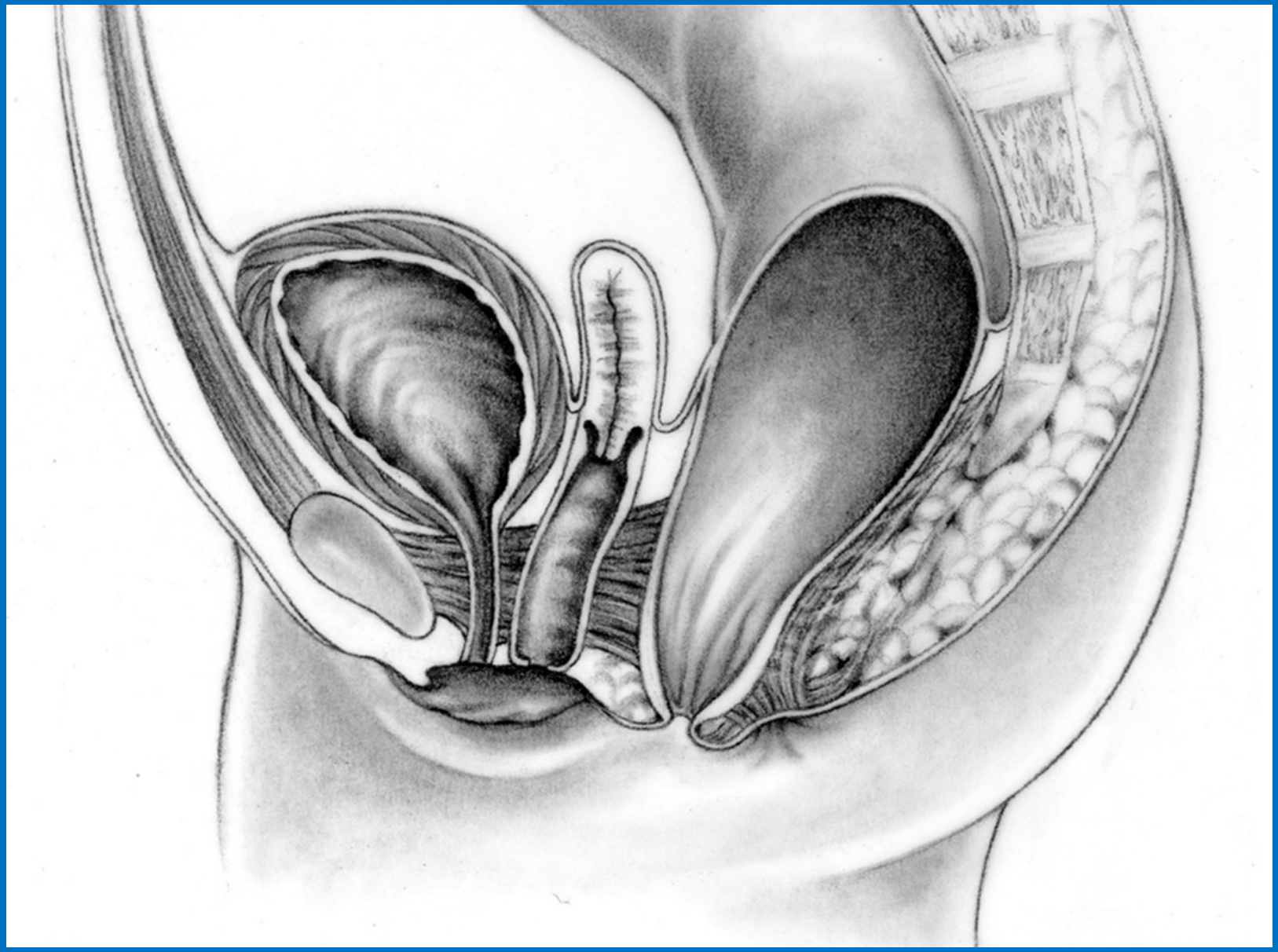
# Recto-perineal Fistula

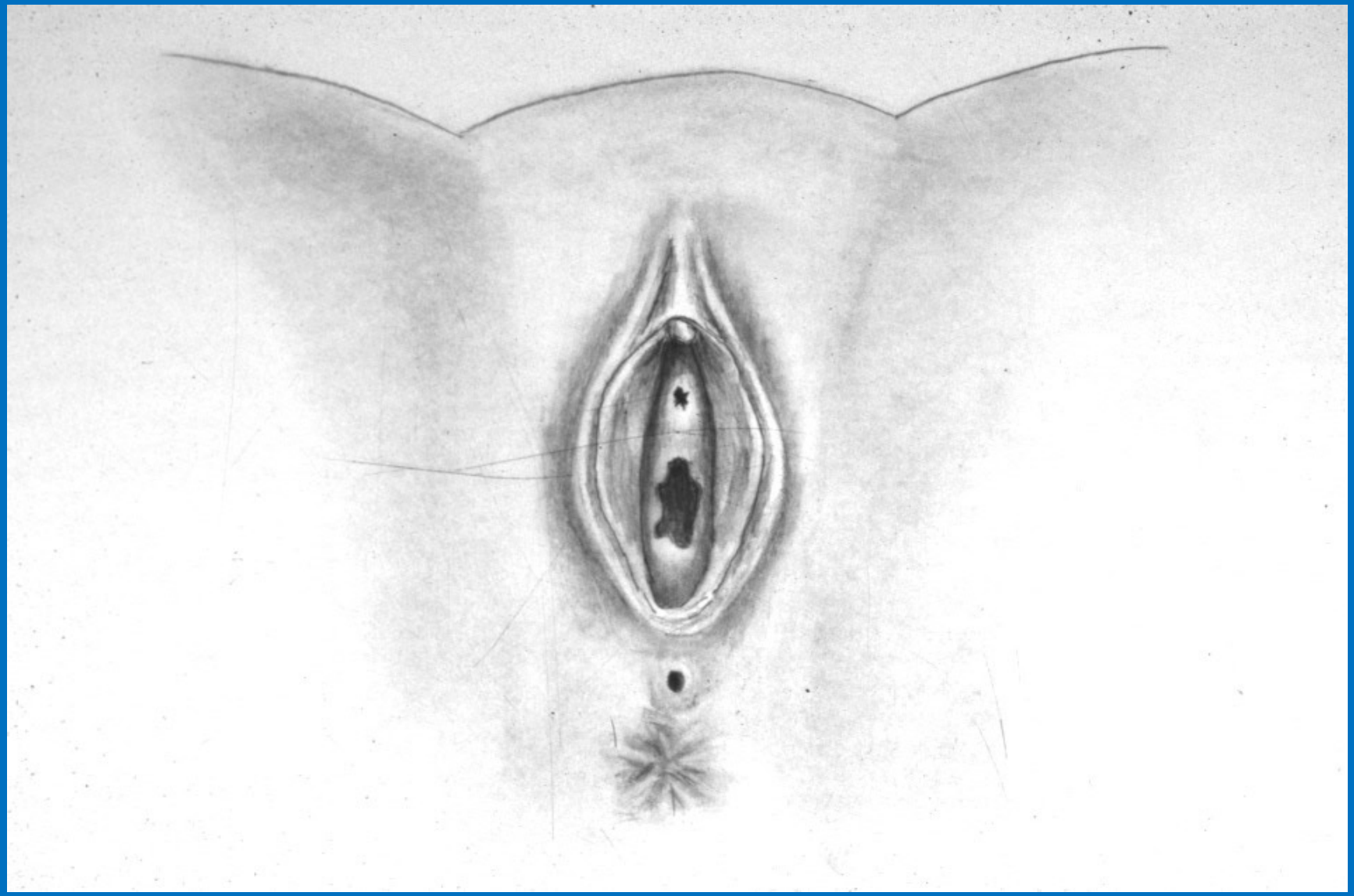
- Most benign type of anorectal malformation
- 100% chances of bowel control provided:
  - Good operation
  - No tethered cord
  - Good sacrum
  - No presacral mass

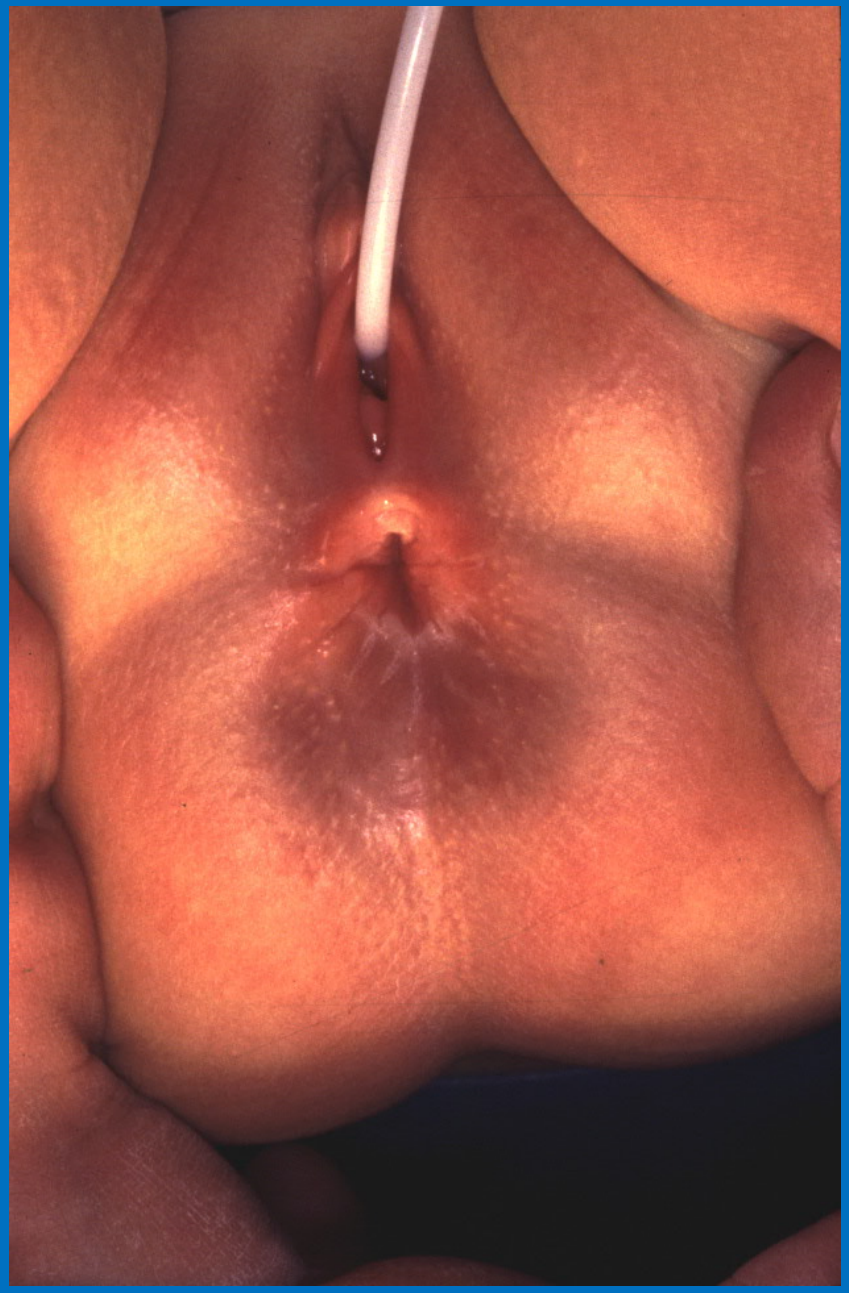
- Patients will suffer from SEVERE constipation







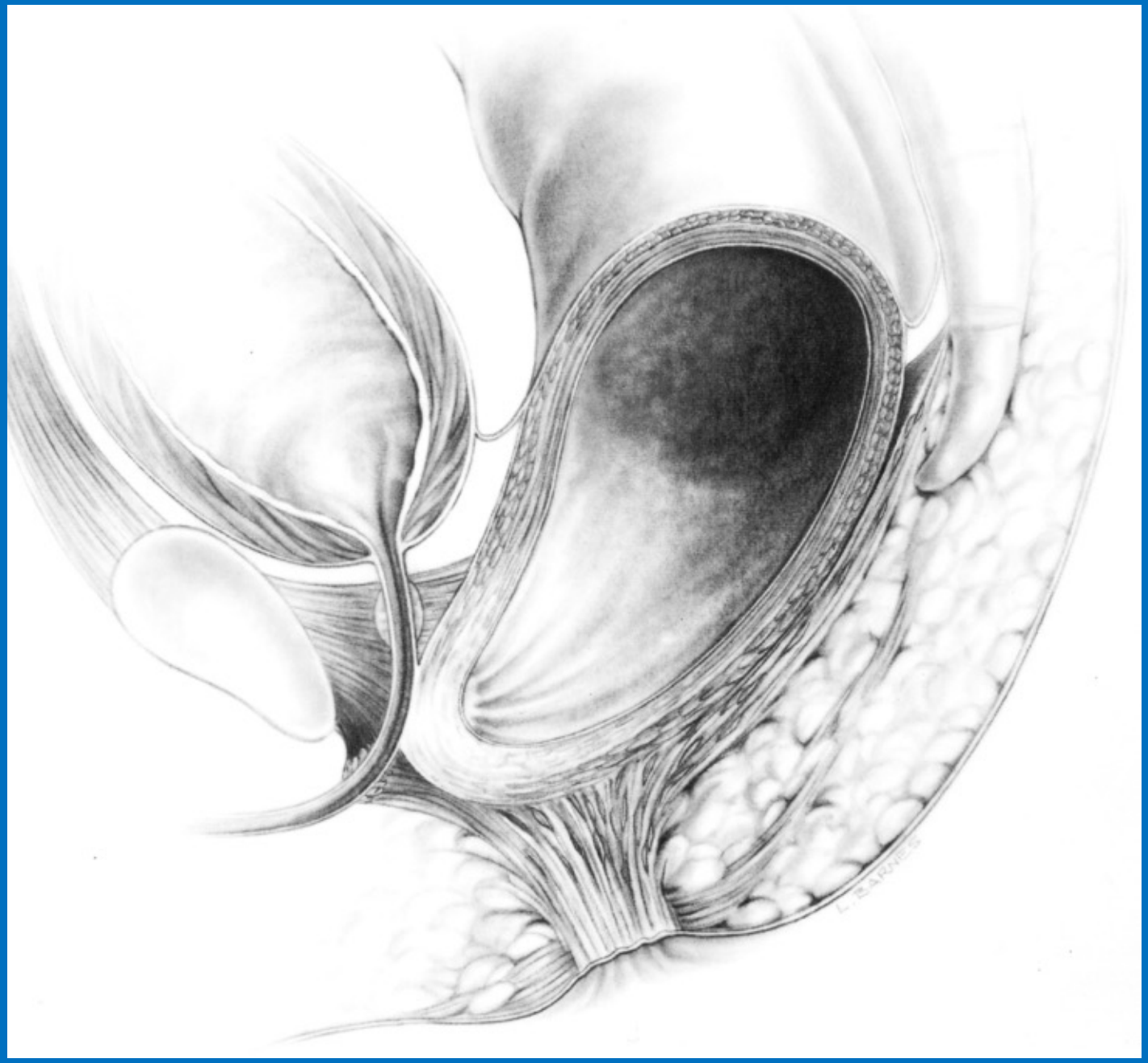




# No Fistula

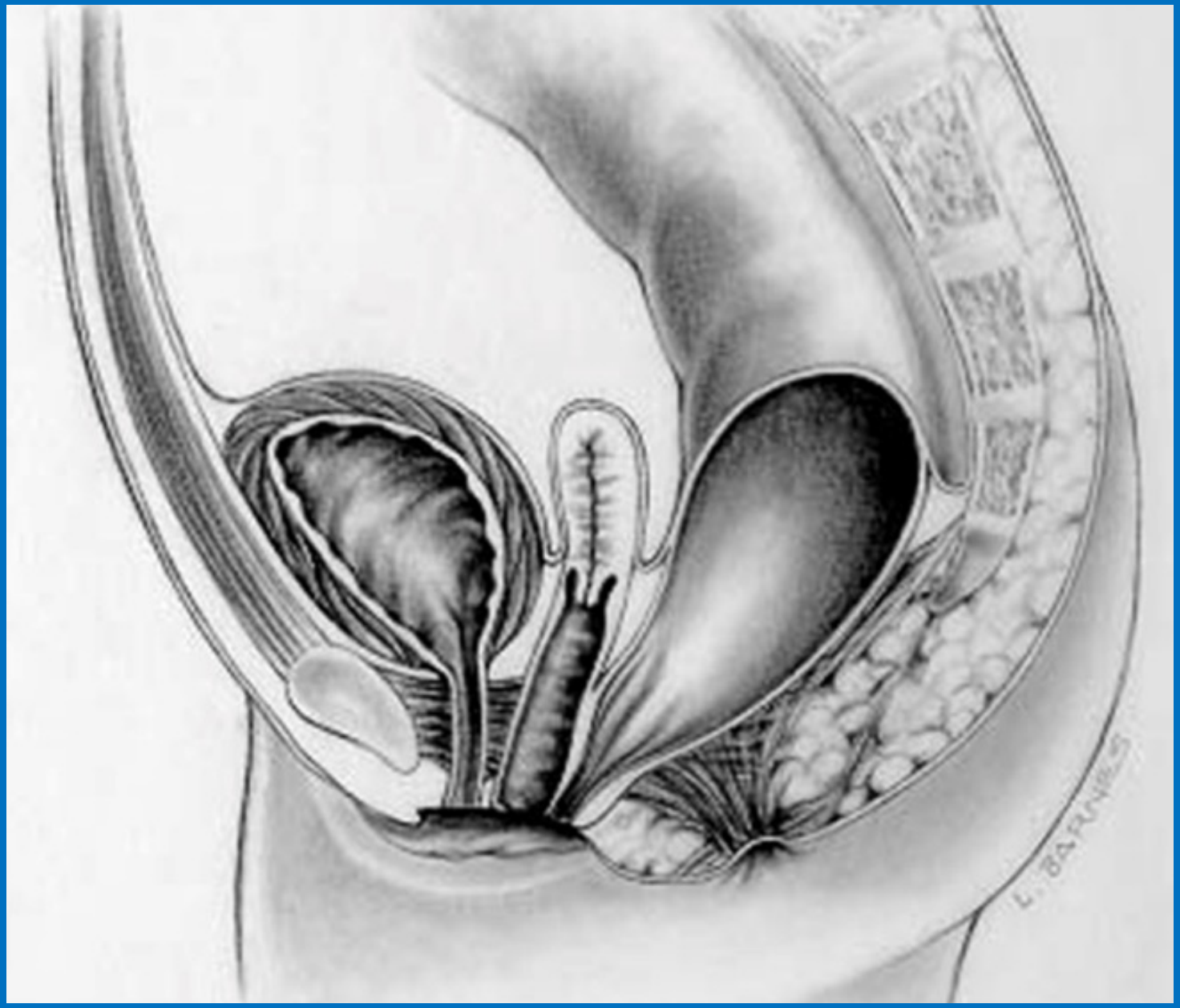
- 80% to 90% chances of bowel control provided:
  - Good operation
  - No tethered cord
  - Good sacrum

- Patients with trisomy 21 have higher incidence of anorectal malformation without fistula.
- Patients will suffer from SEVERE constipation.



# Recto-vestibular Fistula

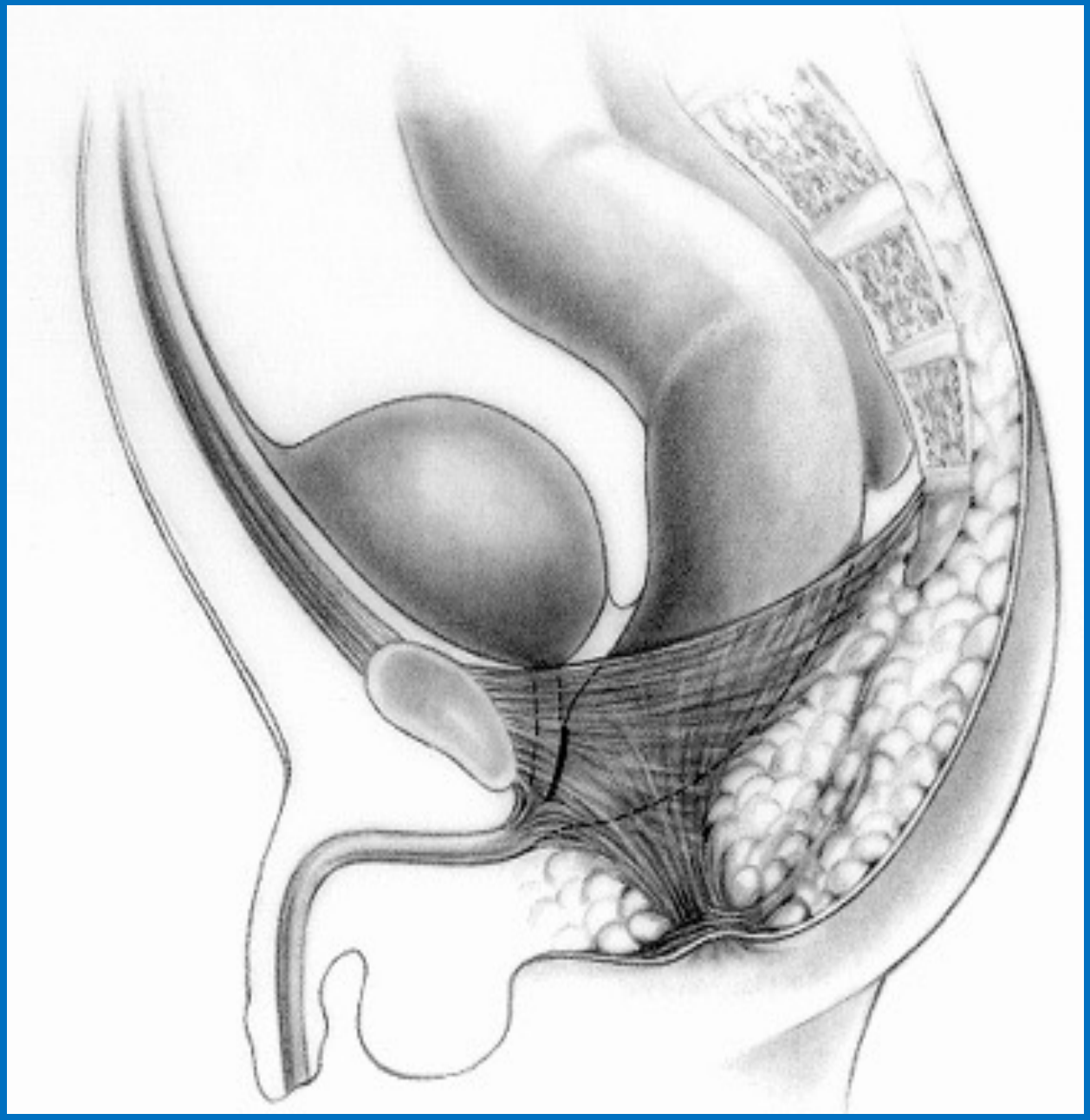
- Most common anorectal malformation in female patients.
- Good prognosis for bowel control (95%) provided:
  - Good operation
  - Normal sacrum
  - No tethered cord
- Patients will suffer from SEVERE constipation



# Recto-urethral Bulbar Fistula

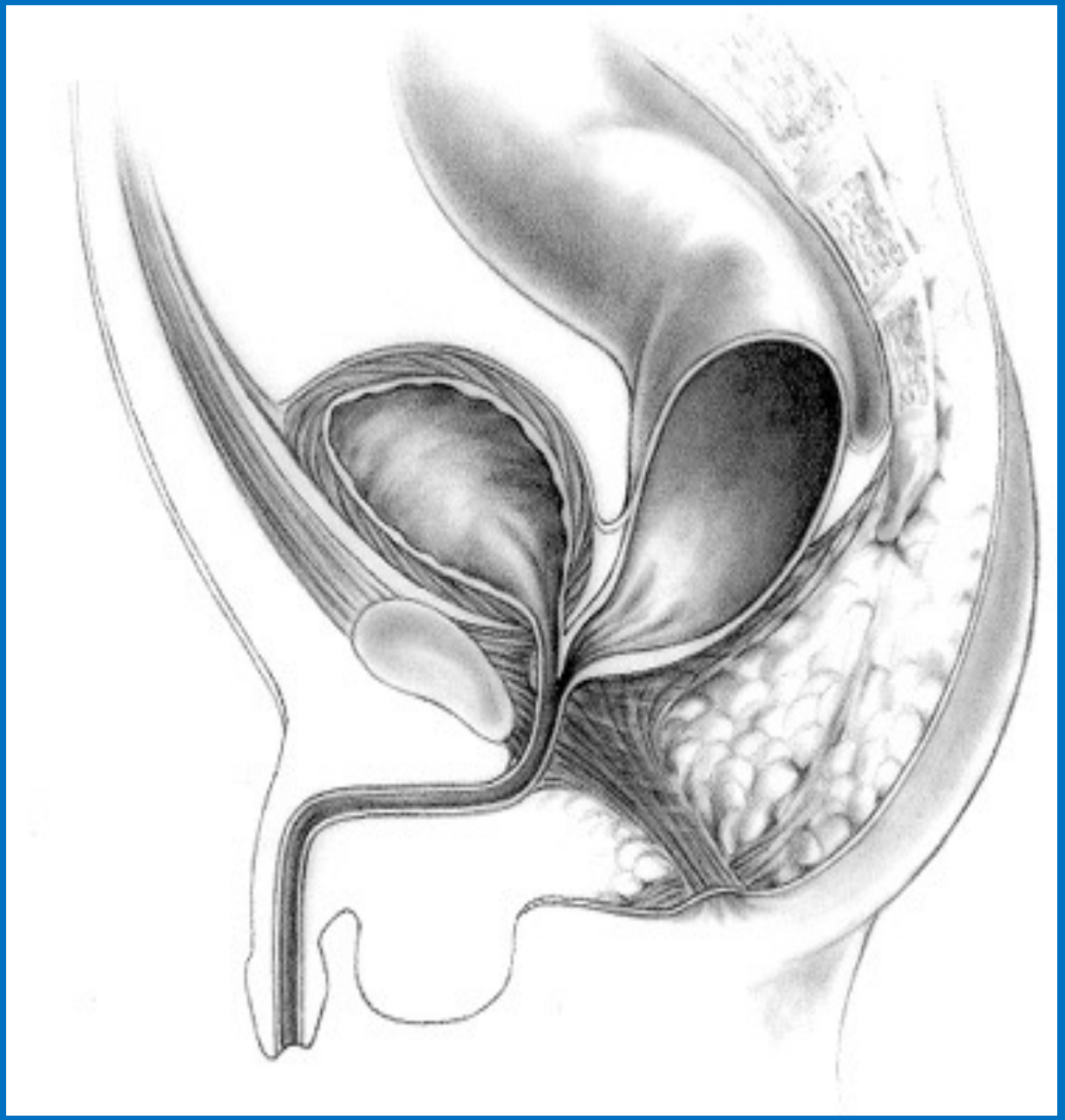
- 85% chances of bowel control provided:
  - Good operation
  - No tethered cord
  - Good sacrum

- Majority of patients will suffer from constipation.



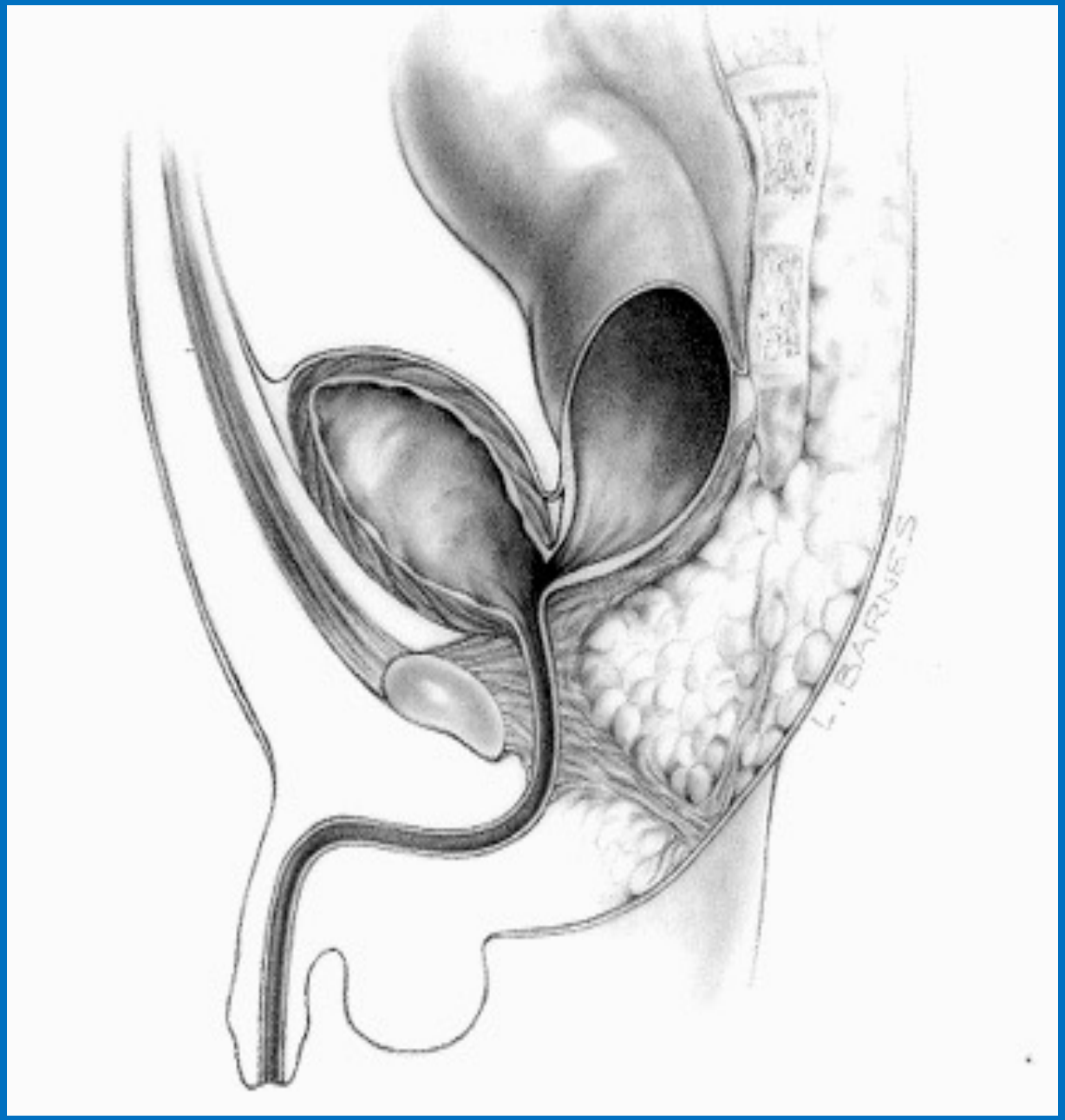
# Recto-urethral Prostatic Fistula

- 60% chances of bowel control.



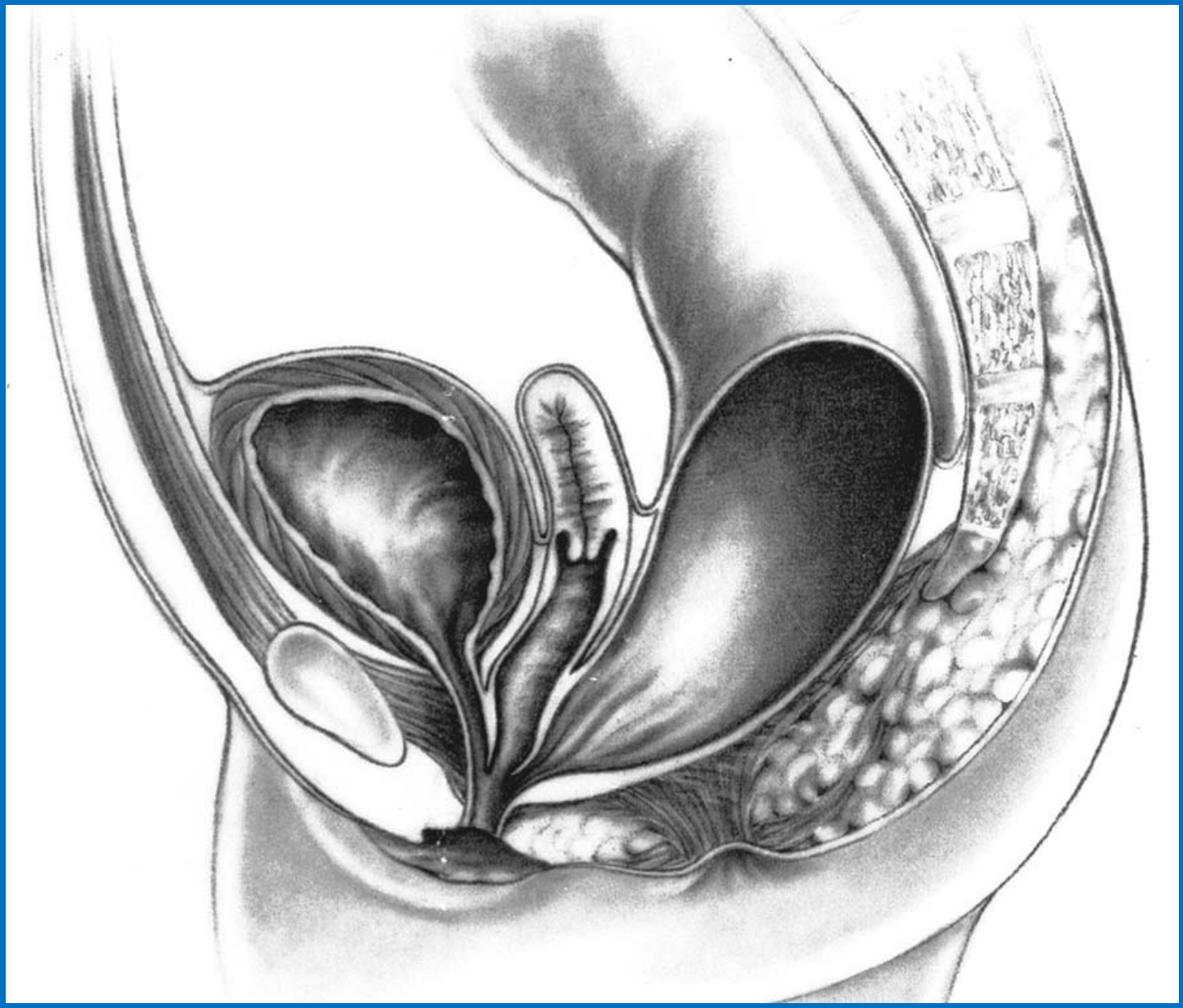
# Recto-bladderneck Fistula

- Poor prognosis for bowel control.
- Only 15% of patients will have bowel control.


L. BARNES

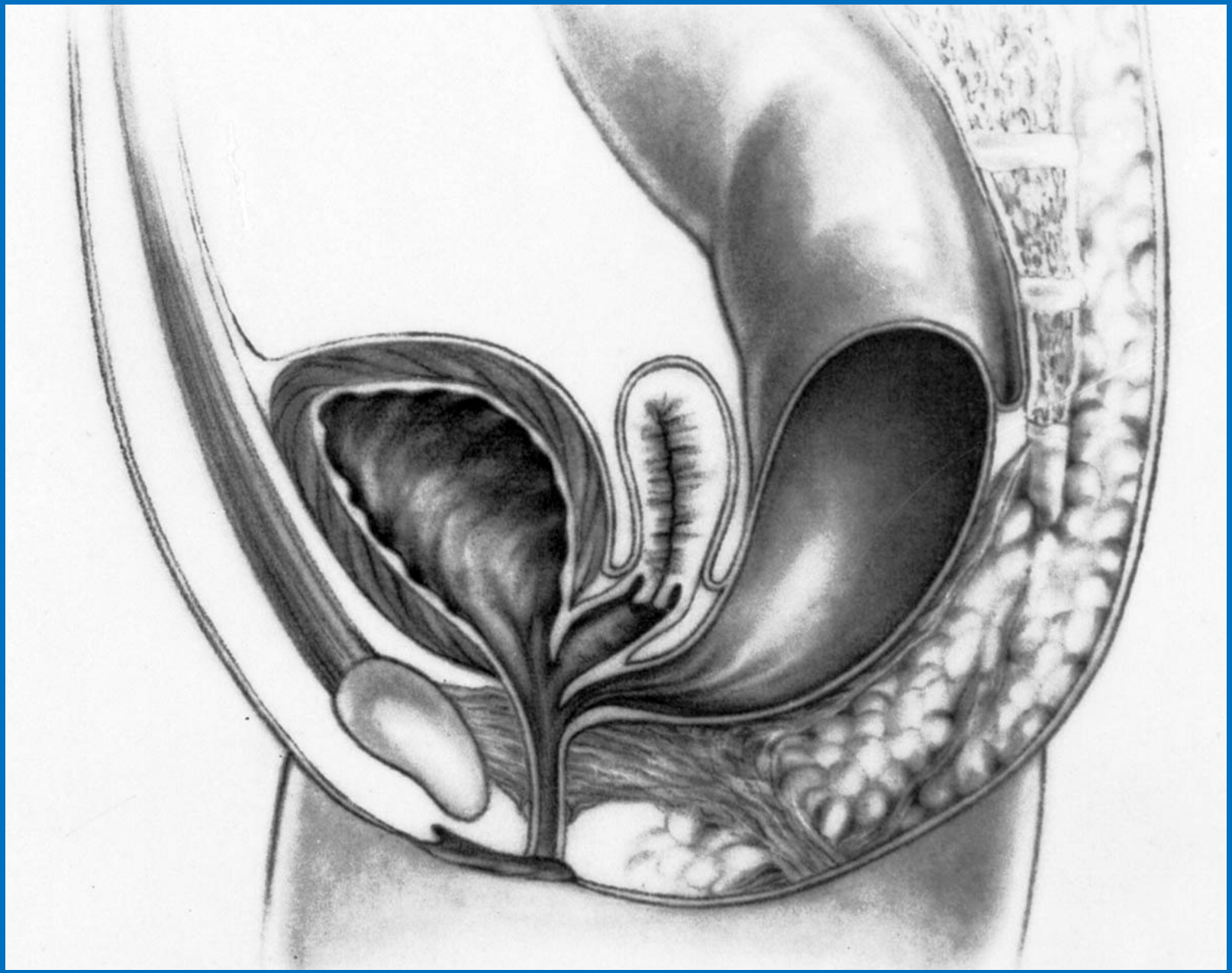
# Cloaca

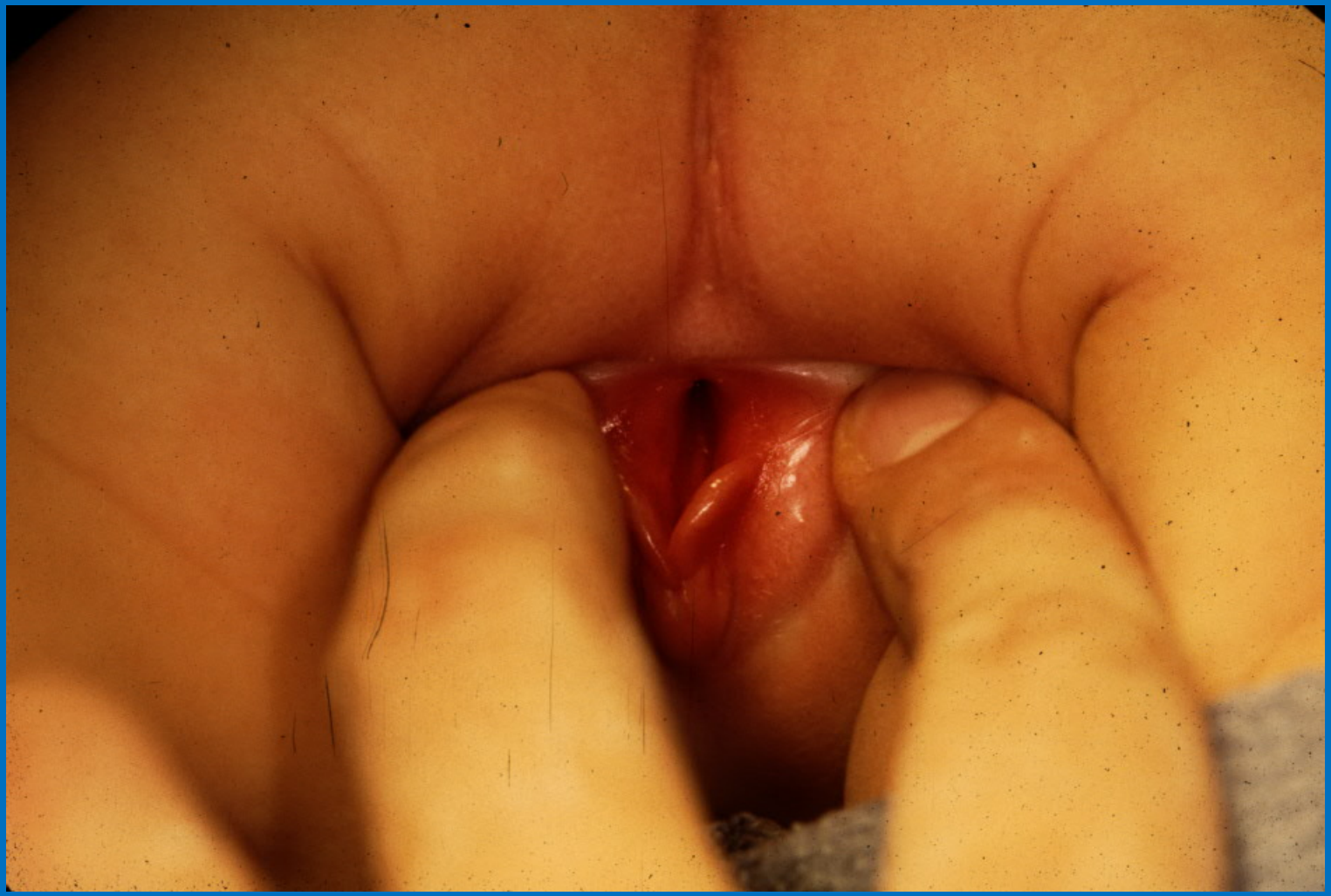
- Good prognosis for bowel control:
  - Normal sacrum
  - No tethered cord
  - Good surgical repair
  - Less than 3 cm common channel



# Cloaca

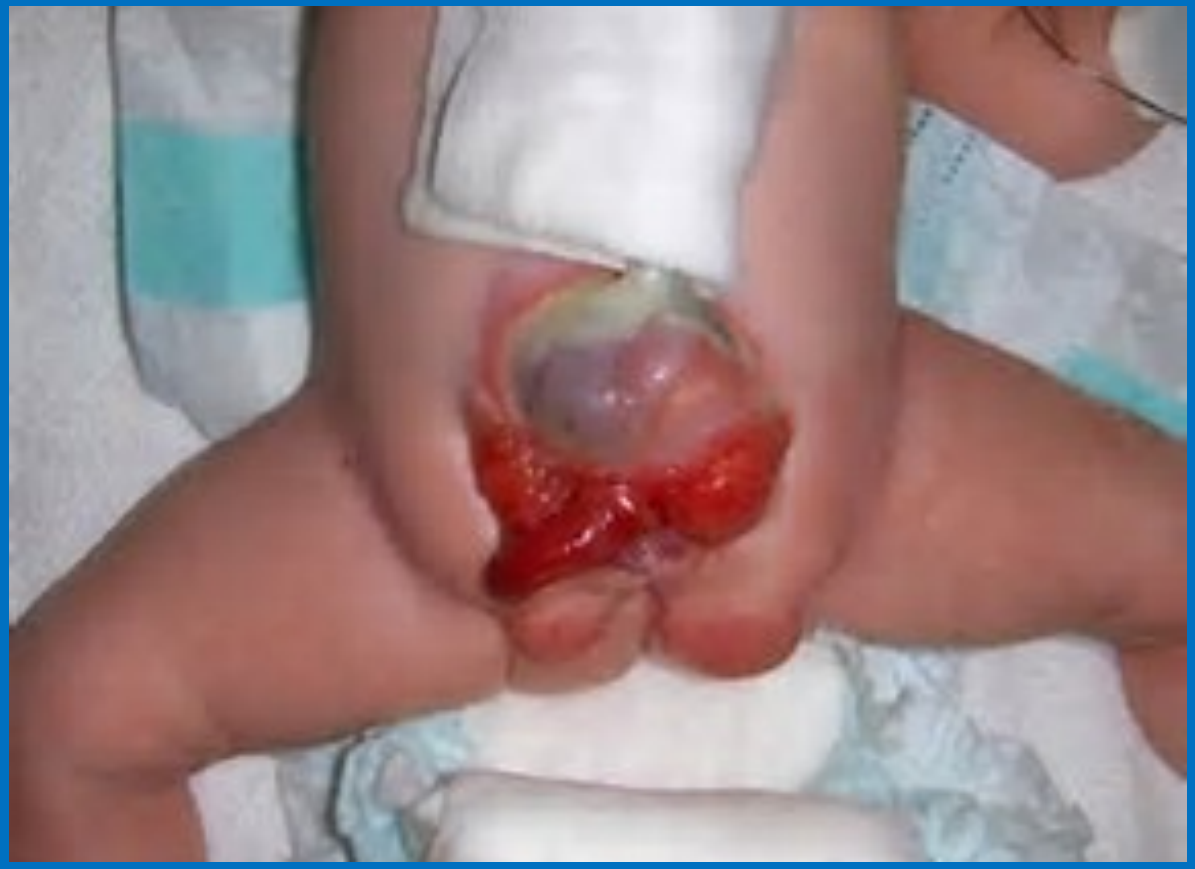
- Poor prognosis for bowel control:
  - Sacral ratio <0.4
  - Tethered cord
  - Reoperations
  - Long common channel



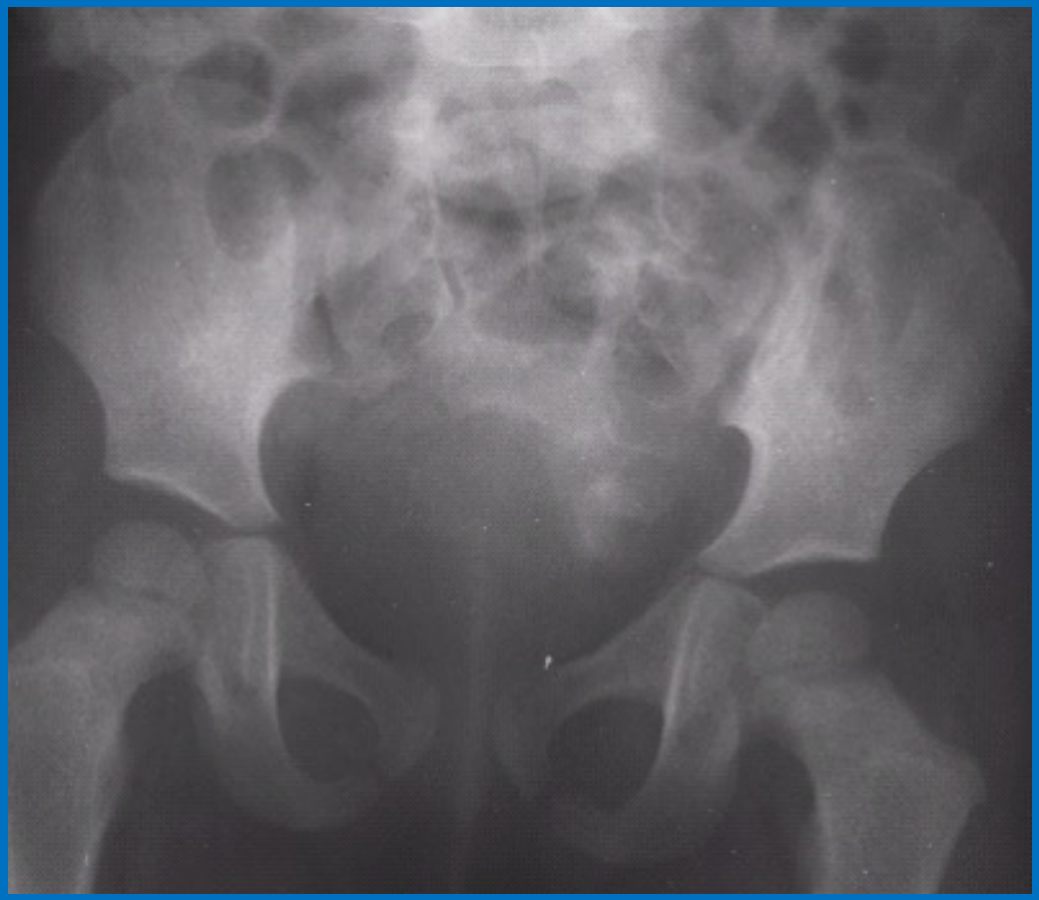


# Cloacal Exstrophy

- Abdominal wall defect with bowel exposed, anorectal malformation, and bladder and reproductive organs separated into two halves
- Often misidentified as “ambiguous genitalia”
- Different amount of colonic length
- Poor prognosis for continence



# What type of anorectal malformation is most common in patients presenting with a hemisacrum?



A. Cloaca
B. Vestibular fistula
C. Perineal fistula with presacral mass
D. Cloacal exstrophy
E. I don't know

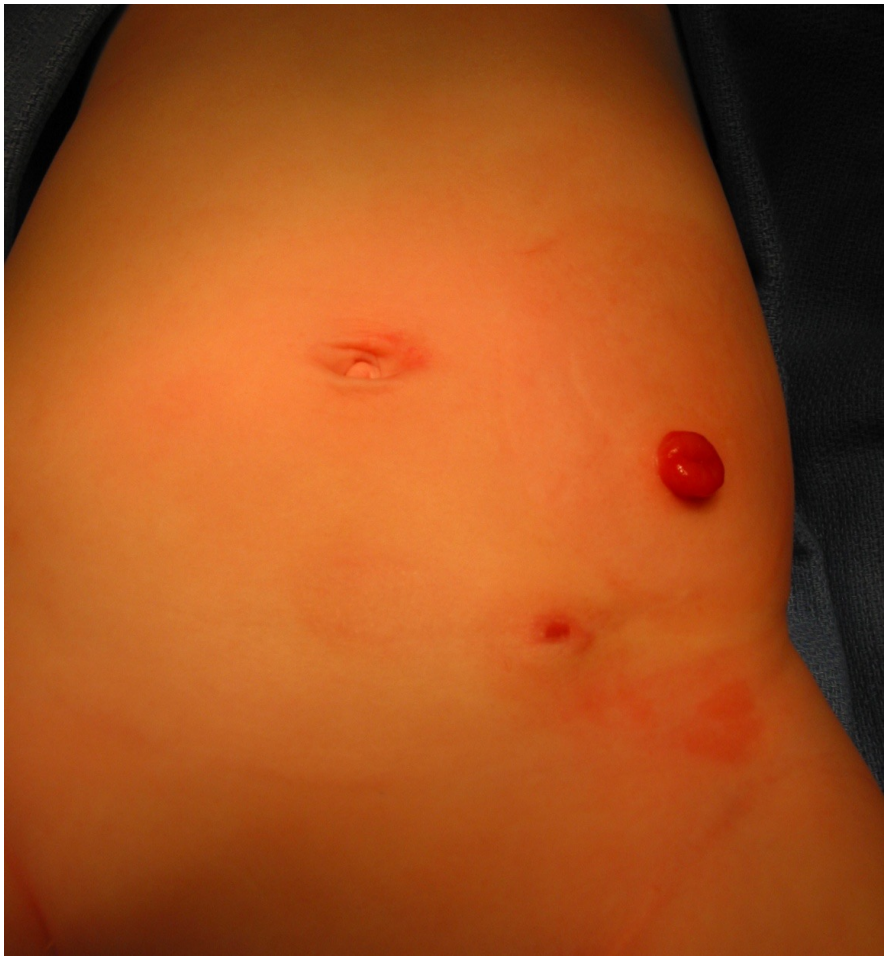
# Surgical Management of Anorectal Malformation

- Primary repair versus delayed repair with diverting colostomy
- Depends upon the complexity of the malformation and the ability to pass stool

# What is the preferred type of stoma for a patient with anorectal malformation requiring diversion?

A) Loop colostomy
B) Divided colostomy in the transverse colon
C) Divided colostomy in the descending colon
D) Ileostomy
E) I don't know

# Colostomy with Mucous Fistula

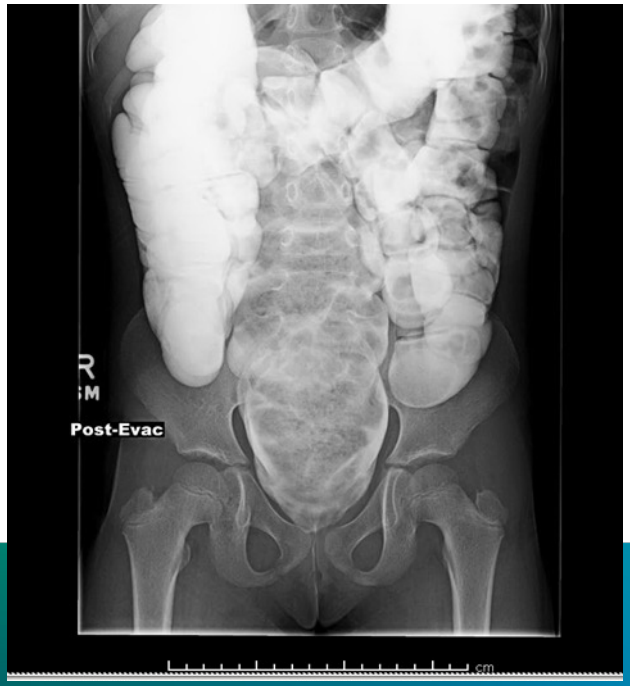


# Long-term Considerations: Infancy

- Constipation
- Close follow up is important
- Abdominal radiographs to monitor stool burden
- Pedialax or small doses of Miralax


# Infancy to Toddlerhood

- Stooling in a diaper is age-appropriate
- Continue to follow closely for constipation
- Aggressive management to prevent colonic dilation



# Preschool and Older

- Attempt toilet training with understanding that patient may not have bowel control
- If unable to toilet train by 4 y/o, refer for bowel management
- Psychosocial implications

# Adulthood

- Female patients who have had a vaginoplasty require vaginoscopy when nearing puberty.
- This allows to evaluation of the patency of the vagina prior to menstruation.
- Patients are followed throughout life into adulthood.

# CHCO Multidisciplinary Team

- Colorectal Surgery
- Urology
- Neurosurgery
- Gynecology
- Nurse Practitioners
- Nurses
- Social Work
- Psychology

# Conclusions

- Anorectal malformation is a spectrum of congenital problems with differing prognoses
- Requires a multidisciplinary approach and thorough newborn screening
- Long-term follow up is key

# Thank you!

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