IMAGING IN DEVELOPMENTAL ANOMALIES OF GIT - II

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Derangement of embryological development can lead to malformations at any point along the gastrointestinal tract (GIT) from the oropharynx to the anorectum.

1] ANATOMICAL –

Attributed to embryological maldevelopment:

- Esophageal atresia with or without fistula
- antropyloric atresia
- antral diaphragm
- duodenal atresia and stenosis
- anorectal atresia.

- **Attributed to in utero catastrophic (ischemic) complication**:
- Jejuno-ileal atresia
- colonic atresia or stenosis
- complicated meconium ileus

- **2] FUNCTIONAL –**

- Meconium plug syndrome and its variants
- Megacyst

- **3] COMBINED – ANATOMICAL and FUNCTIONAL -**

- Hypertrophic pyloric stenosis
- Midgut volvulus (complicating midgut malrotation)
- Uncomplicated meconium ileus
- Colonic aganglionosis (Hirschsprung's disease)

IMAGING MODALITIES

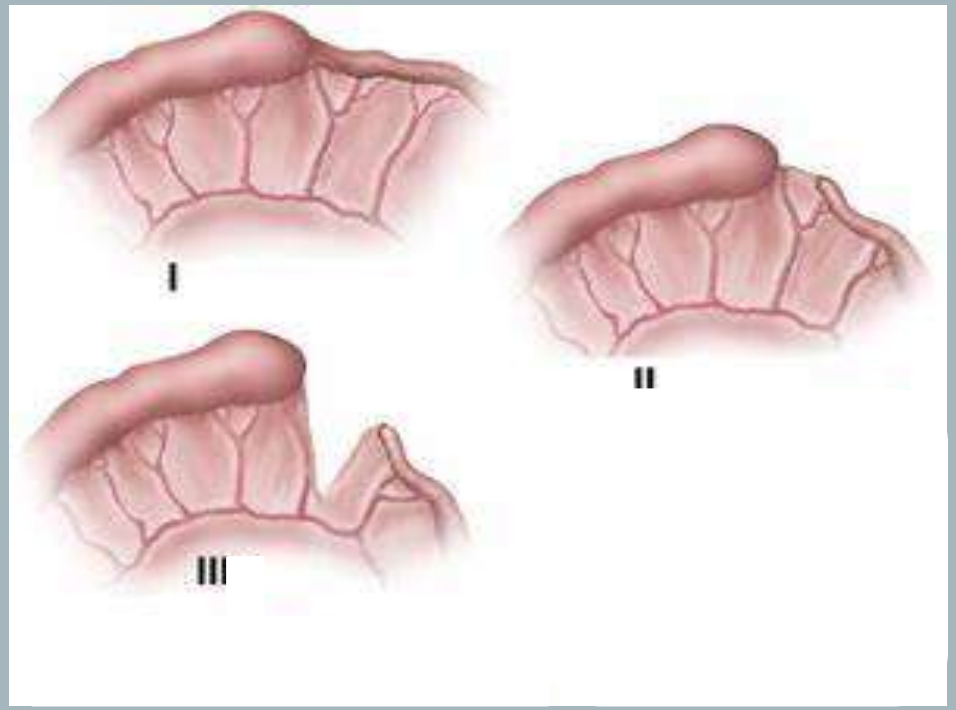
1) Plain radiograph
2) Contrast studies
3) Computed tomography
4) Magnetic resonance imaging

COLONIC OBSTRUCTION

- Anatomical or functional.
- Atresia of the colon, anorectal atresia, and aganglionosis or Hirschsprung's disease.
- Associated with VACTERL anomalies

COLONIC ATRESIA

- The classification system based on the anatomic appearance is same as with jejunoileal atresia.
- Type 1 : a diaphragmatic occlusion;
- Type 2 : a complete atresia with a blind, solid cord extending between the two ends of atretic segment;
- Type 3 : a complete atresia with complete separation and an associated V-shaped mesenteric defect



Contrast was introduced into the rectum via a catheter.
No passage of contrast proximal to the rectosigmoid junction despite multiple attempts – Suggestive of colonic atresia

HIRSCHSPRUNG'S DISEASE (AGANGLIONOSIS OF THE COLON)

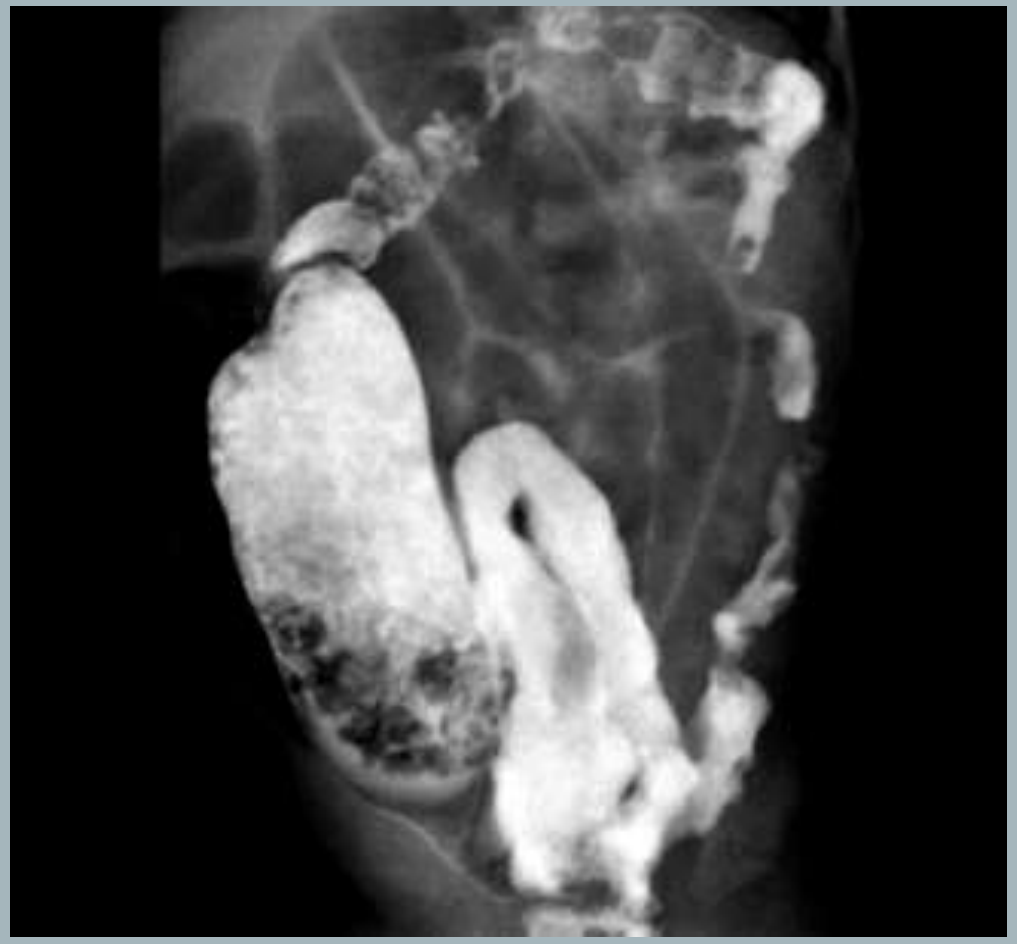
- Absence of normal ganglion cells in a segment of colon
- caused by abnormal neural crest cell migration, resulting in arrested distal migration of these cells.
- As the normal migration is continuous from proximal to distal, the part of the GI tract distal to the site of arrest is aganglionic.
- Always involves the anus and internal sphincter and extends proximally for a variable distance
- Hirschsprung's disease fail to pass meconium in the first 48 hours of life.

BARIUM ENEMA

- A dilated colon proximal to the distal and smaller aganglionic segment is the more typical finding.
- The transition zone generally is funnel-shaped and it is an important diagnostic feature.
- Irregular saw-toothed mucosal pattern due to disordered contractions in the aganglionic colon.
- The rectosigmoid index - compares the ratio of the rectal diameter to the sigmoid diameter - abnormal, if the sigmoid colon is more dilated than the rectum (R/S index <1).
- Delayed radiographs (24 hours) – Prolonged retention of barium (strong indicated)

Hirschsprung's disease:

Barium enema shows an abrupt transition from the narrow caliber rectosigmoid (aganglionic) to the larger caliber more proximal sigmoid colon



Barium enema

Colon of reduced caliber from rectum to hepatic flexure with markedly dilated colon (cecum and ascending colon) proximal to it—long segment aganglionosis

ANORECTAL ANOMALIES

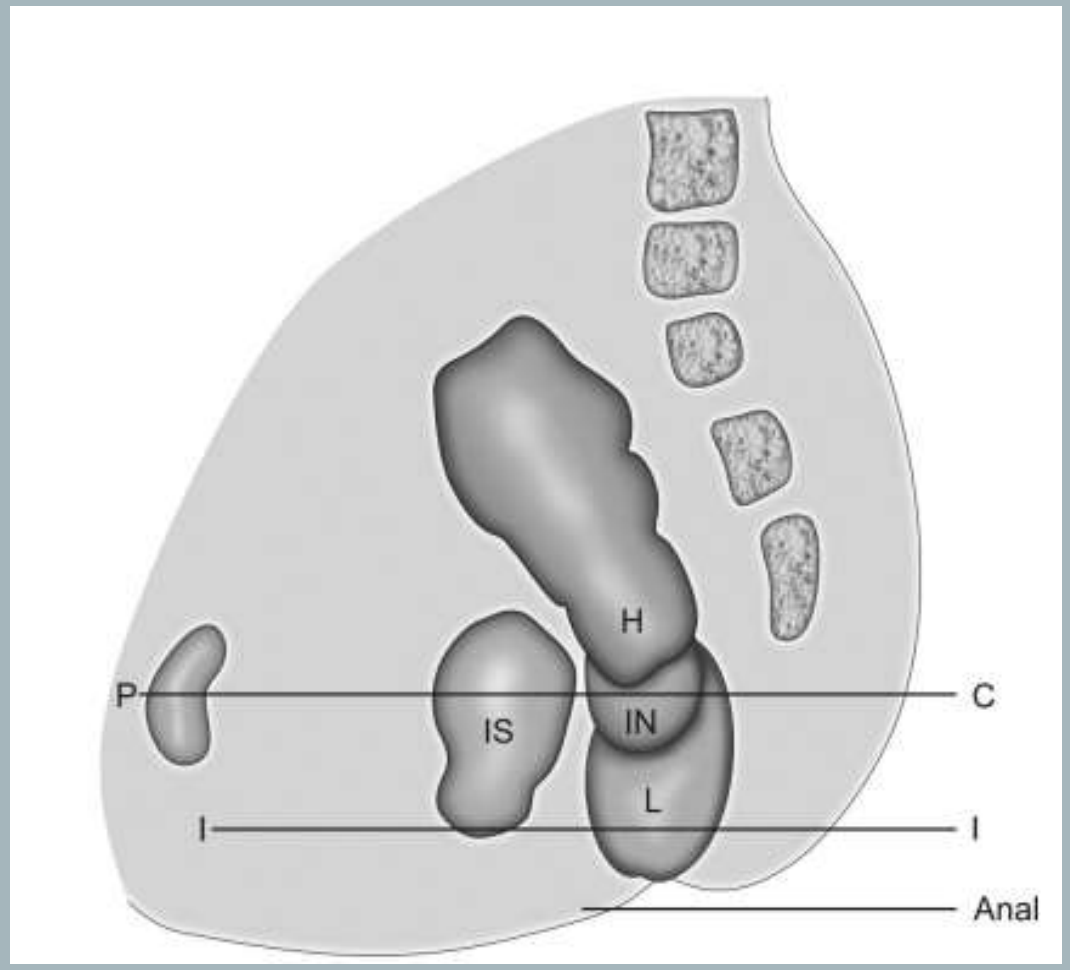
Anorectal anomalies:		
Level	*Male*	*Female*
High anomalies	1. *Anorectal agenesis:* a. Without fistula b. *With fistula:* i. Rectovesical ii. Rectoprostatic urethral (including H or N type) 2. Rectal atresia	1. *Anorectal agenesis:* a. Without fistula b. *With fistula:* i. Rectovesical ii. Cloacal anomalies iii. Rectovaginal (high) 2. Rectal atresia
Intermediate anomalies	1. *Anal agenesis:* a. Without fistula b. *With fistula:* i. Rectobulbar 2. Anorectal stenosis	1. *Anal agenesis:* a. Without fistula b. *With fistula:* i. Rectovaginal (low) ii. Rectovestibular 2. Anorectal stenosis
Low anomalies	1. *At normal anal site:* a. Covered anus, complete b. Anal stenosis 2. *At perineal site:* a. Anterior perineal anus b. Anocutaneous fistula	1. *At normal anal site:* a. Covered anus, complete b. Anal stenosis 2. *At perineal site:* a. Anterior perineal anus b. Anocutaneous fistula 3. *At vulvar site:* a. Anovestibular fistula b. Vulvar anus c. Anovulvar anus

ANORECTAL ATRESIA

IMAGING WORK-UP

- **1. Invertography**
- A newborn baby with ARA - held upside down for at least 3 minutes,
- Strict lateral view (in this upside down position) taken with the thighs of the baby flexed at hip, and beam accurately centered on the greater trochanter.
- Anal dimple and the natal cleft should be outlined by barium paste.
- **In a correctly taken radiograph:**
- (i) both the ischial bones would accurately superimpose
- (ii) the terminal blind bowel will be rounded and well-distended.

- **<u>Interpretation:</u>**
- Identify the relationship of the gas bubble in the blind pouch to the bony pelvis.
- (i) PC (pubococcygeal) line
- (ii) The I (ischial) line or I point
- (iii) The anal pit line
- A gas bubble situated above the PC line - "high" anomaly,
- One located between the PC and I line - "Intermediate" anomaly
- It extends below the I line - "low" anomaly



Line drawing showing high (H), intermediate (IN) and low (L) anomalies and their relationship to pubococcygeal (PC) and ischial (I) line

Low variety of anal atresia.

Coin piece is placed over the expected anus and the baby is turned upside down.
Gas filled large bowel loops noted.
Distance between the highest column of rectal gas shadow and metallic coin (skin surface) measures approx 1 cm

Rectum is located between the PC and I line -“Intermediate” anomaly

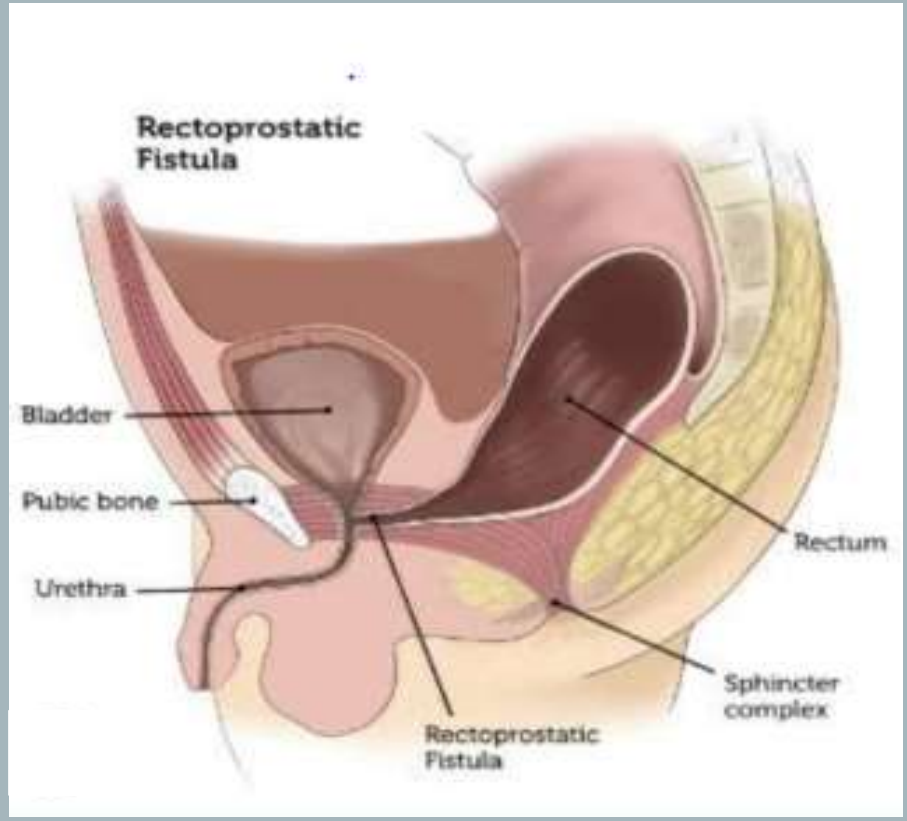
CONTRAST STUDIES

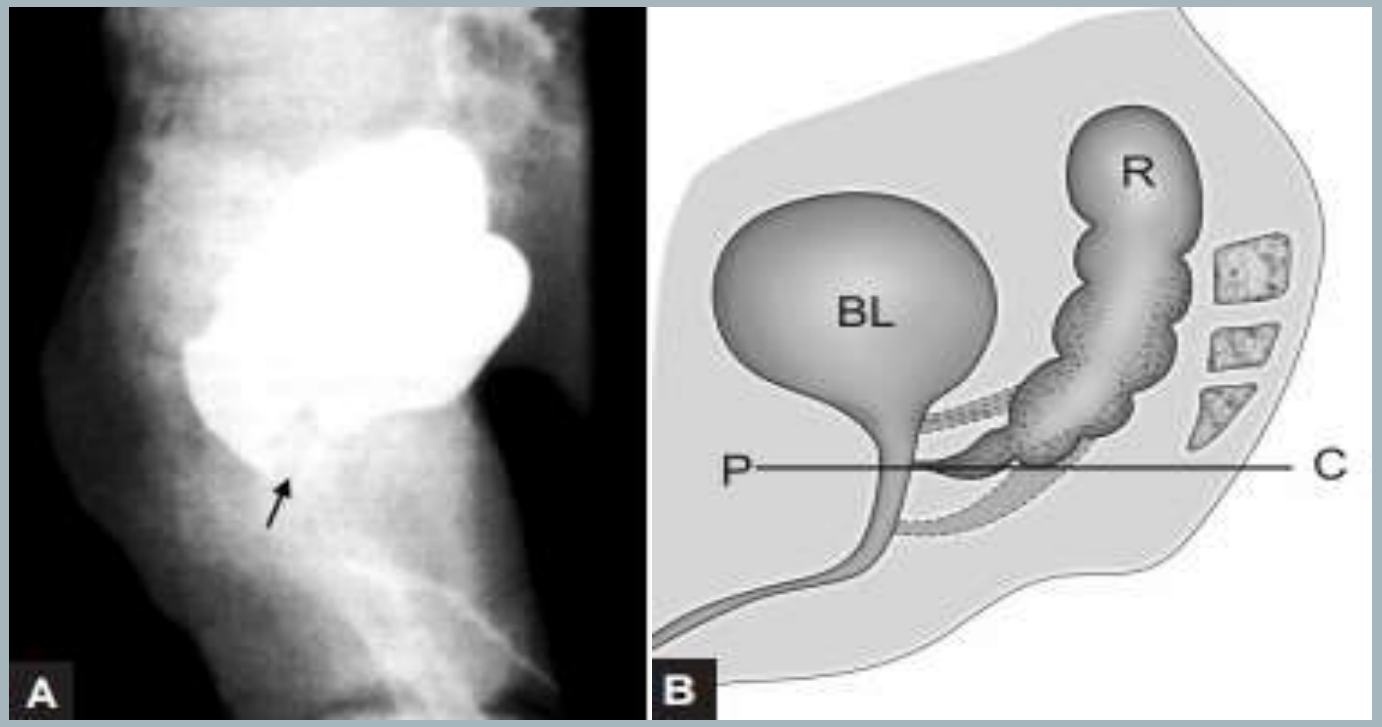
- Purpose of any contrast study :
- i. To confirm the level of blind bowel pouch
- ii. To document the presence (or absence) and level of the fistula between the GI (gastrointestinal) and the GU (genitourinary) system

ANORECTAL ANOMALIES IN MALES

High Anomalies

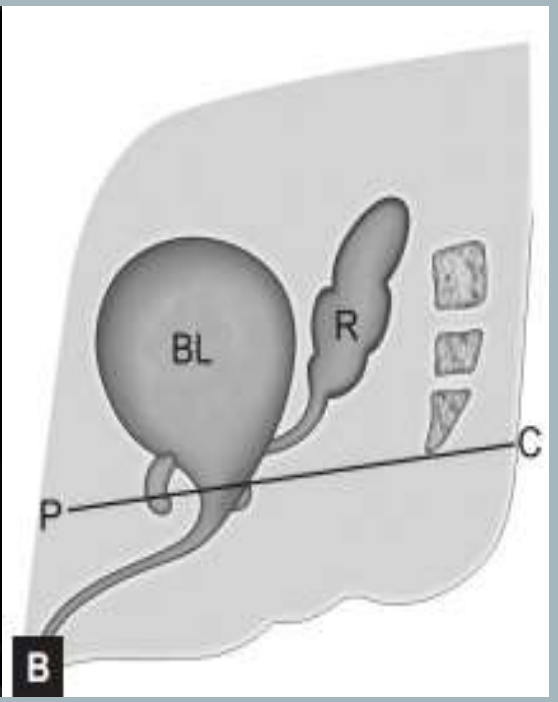
- The imperforate bowel or its fistulous connection lies above the puborectalis sling of the levator ani muscle.
- **1. Anorectal Agenesis without Fistula**
- Uncommon.
- Blind rectal pouch lying at or cranial to the PC line with no suggestion or demonstration of a fistula.
- **2. Anorectal Agenesis with Rectoprostatic Urethral Fistula**
- Most common type
- Most common site of fistulous opening is at the level of the verumontanum,





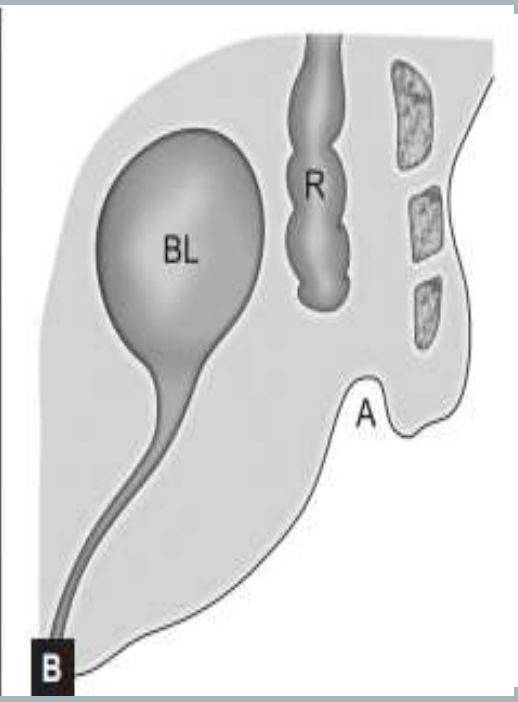
Rectoprostatic urethral fistula—schematic drawing (A) and a case (B). As indicated by dotted lines fistula may enter either more cranial or distal to verumontanum (arrow—fistula)

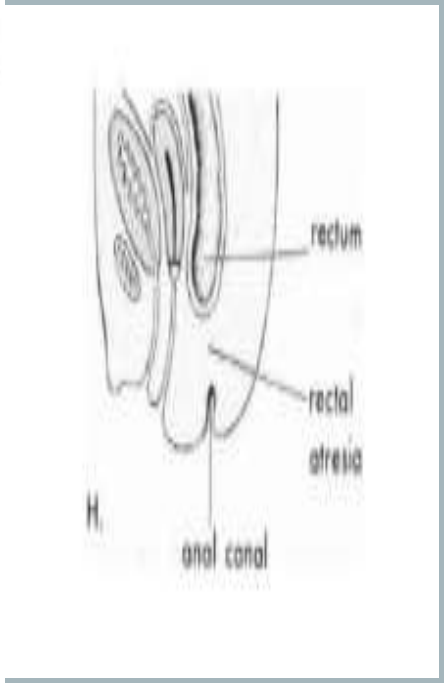
Rectovesical Fistula



The filling of urinary bladder immediately after opacification of the rectum

Rectal Atresia

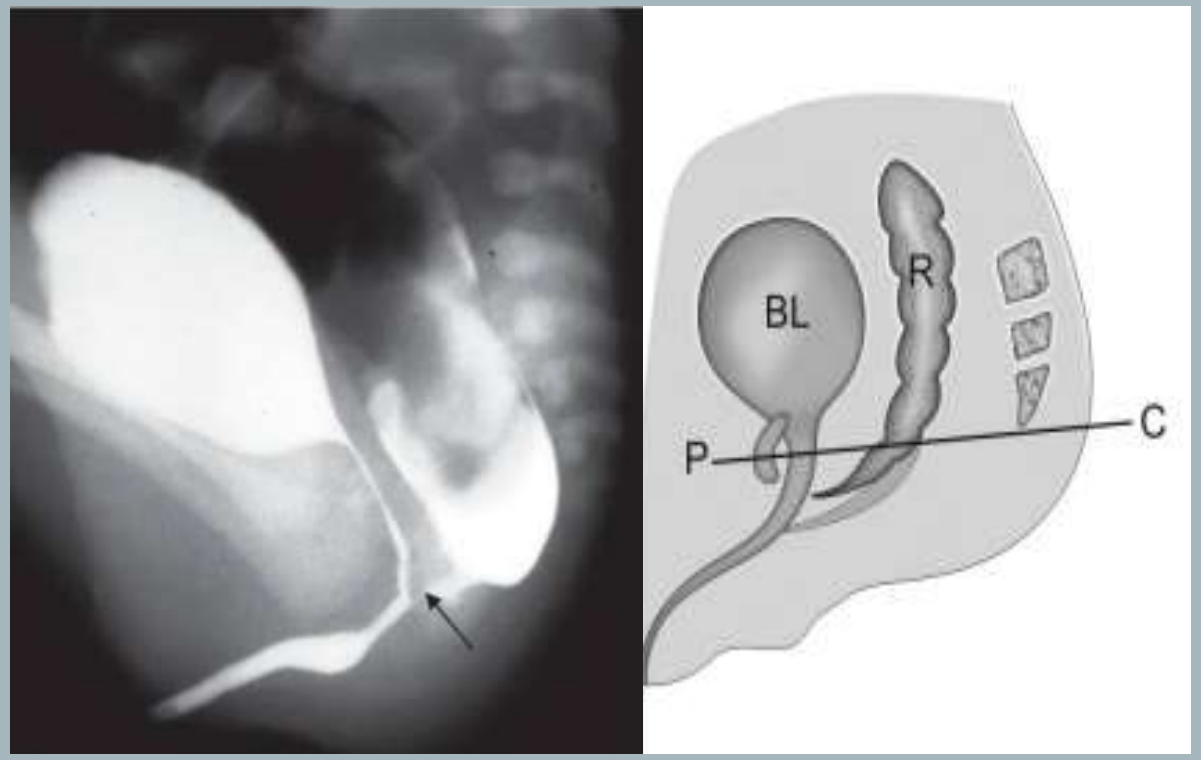




The terminal bowel ends blindly at varying levels.
Anus and the anal canal are normal and the bowel in between these two segments is deficient
Opacification done from both ends of the site of atresia (Distal cologram + anal canal opacification).
(arrows—atretic segment)

ANORECTAL ANOMALIES IN MALES

- **<u>Intermediate Anomalies</u>**
- 1. Anal Agenesis without Fistula
- 2. Anal Agenesis with Rectobulbar Urethral Fistula
- 3. Anorectal Stenosis



<u>Rectobulbar urethral fistula (Lateral Radiograph During High Pressure Distal Loopogram)</u>
Short, wide fistula entering directly into the posterior wall of bulb of urethra.
Long and narrow which enters into the under surface of penile urethra

Cystourethrogram study reveals rectal pouch which is in continuation with the urethra giving rise to rectourethral fistula
there is partial sacral agenesis is present

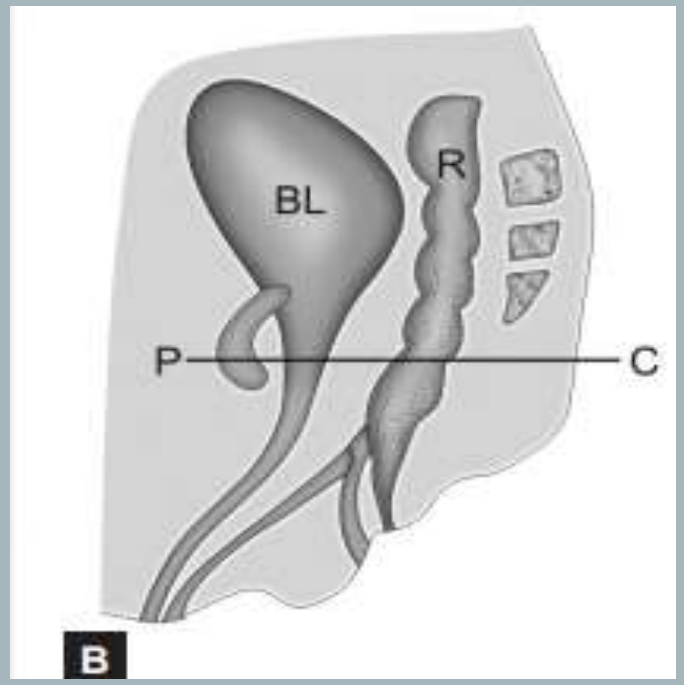
ANORECTAL ANOMALIES IN MALES

- **Low Anomalies**

Anal deformities”

1. Anocutaneous fistula (covered anus, incomplete)
2. Anterior perineal anus
3. Covered anal stenosis
4. Anal membrane stenosis
5. Covered anus, complete
6. Imperforate anal membrane

- **Anocutaneous Fistula**
- Anal canal is of normal caliber till the level of anal valves beyond which it is suddenly reduced to a tube of small caliber and runs forward covered by skin to open anywhere from the ventral surface of the penile shaft to an area adjacent to the anal site



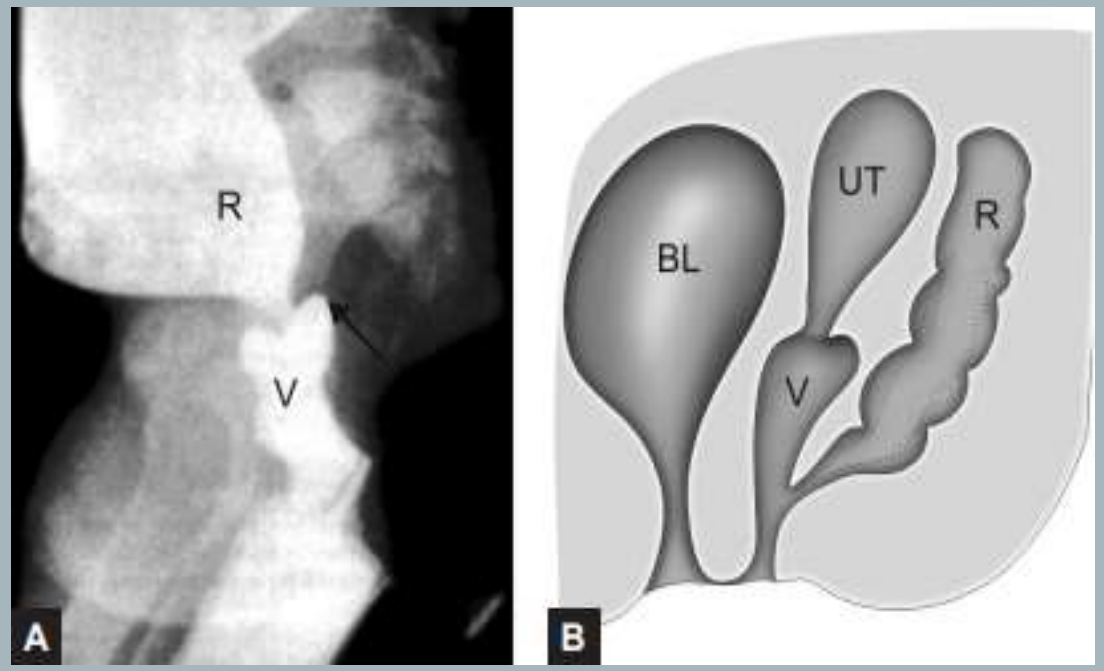
Anterior Perineal Anus

- Anus is normal in appearance but located more anteriorly than the normal site close to the posterior or membranous urethra.

ANORECTAL ANOMALIES IN FEMALES

High Anomalies

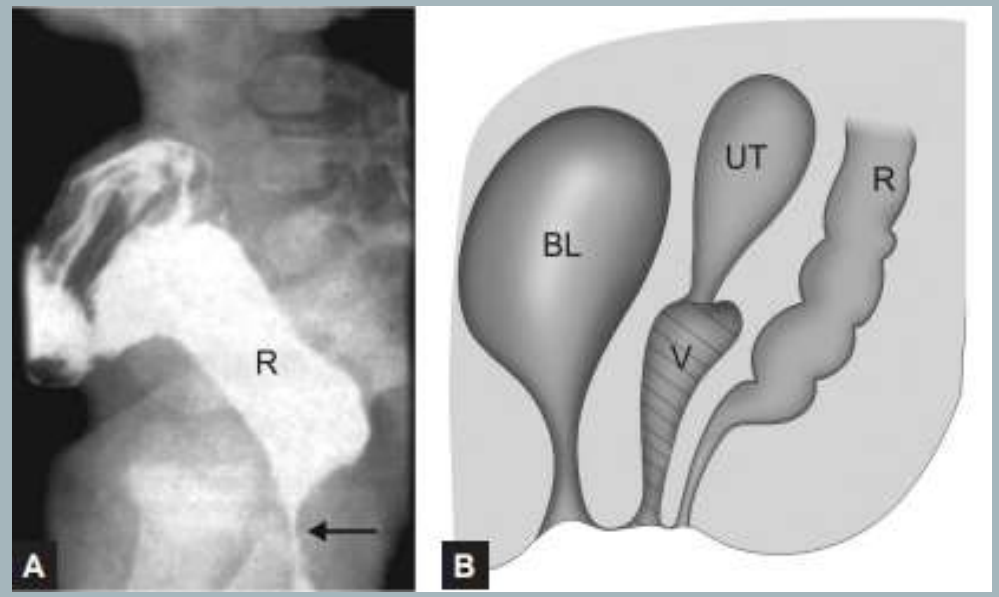
- **1. Anorectal Agenesis without Fistula**
- **2. Anorectal Agenesis with High Rectovaginal Fistula (Without a Urogenital Sinus Defect)Most common type**
- Vagina, urinary bladder and urethra are normal.
- Rectal fistula enters the posterior wall of the vagina.
- **3. Rectovesical Fistula**
- 4. **Rectal Atresia**



Rectovaginal fistula

ANORECTAL ANOMALIES IN FEMALES

- **Intermediate Anomalies**
- 1. **Anal Agenesis without Fistula**
- 2. **Rectovestibular Fistula**
  - From the anterior wall of the rectum arises a fistula which opens in the vestibule.
  - Three orifices in the vestibule Retrograde injection of contrast opacifies the fistula and the rectum
- 3. **Low Rectovaginal Fistula**
  - very rare.



Rectovestibular fistula—retrograde contrast injection. Note the long, narrow fistulous track (arrow)

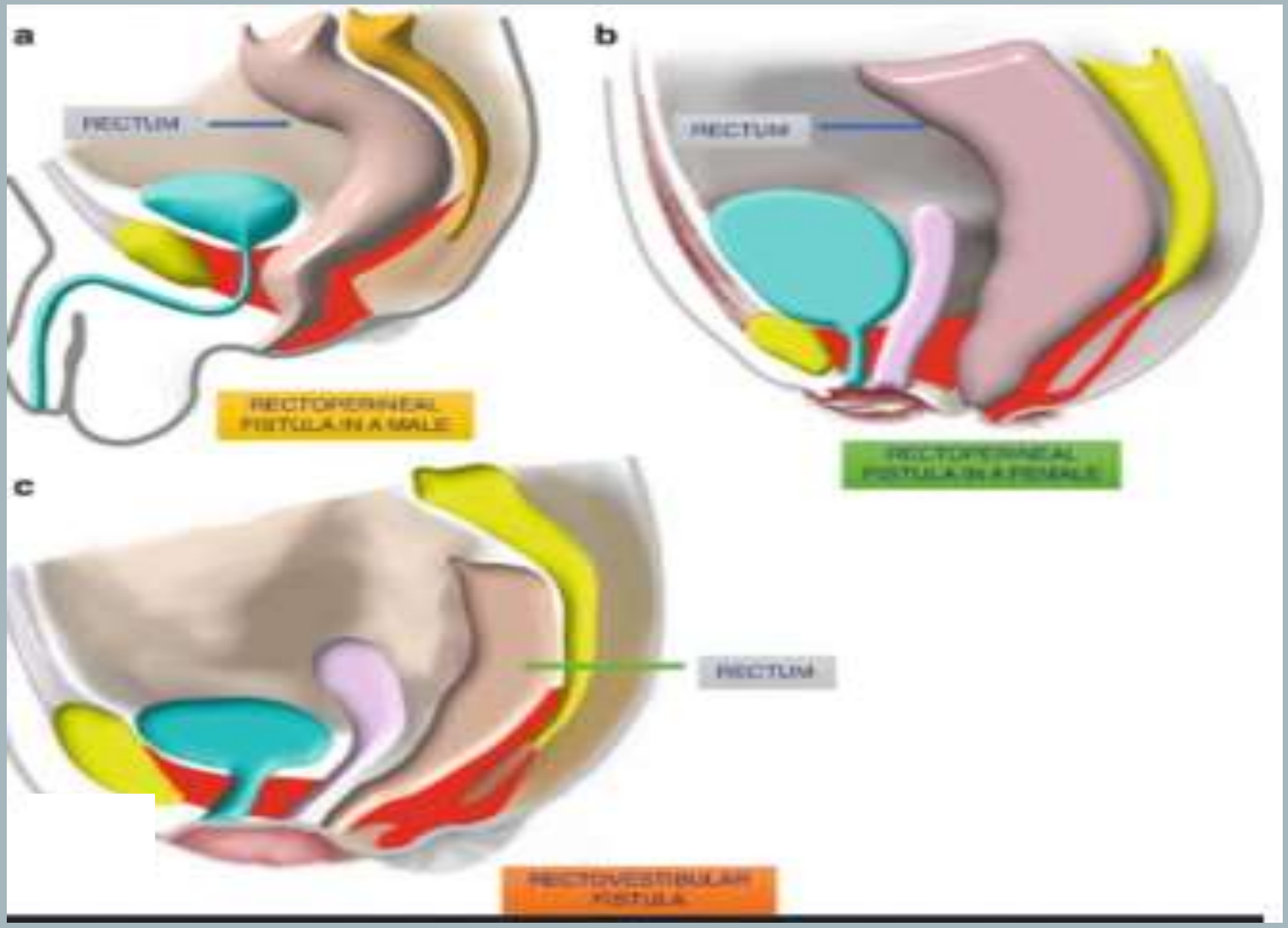
ANORECTAL ANOMALIES IN FEMALES

- **<u>Low Anomalies</u>**
- "anal deformities".
- 1. **Anovestibular Fistula** –
  - it opens in the vestibule.
  - length is very short (less than 1 cm)

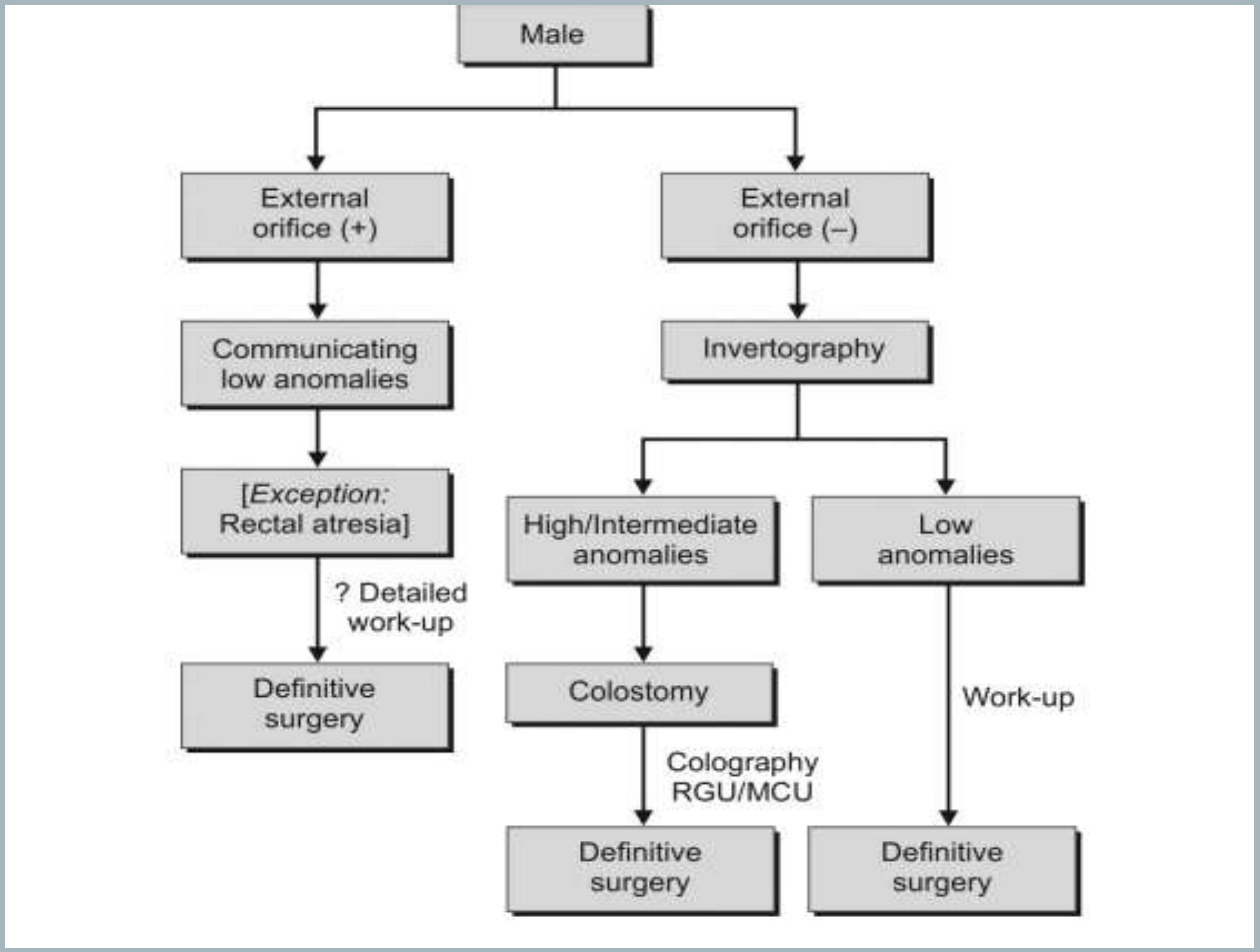
  2. **Anocutaneous Fistula/Anterior Perineal Anus**
- 3. **Anal Stenosis**

Associated anomalies with anorectal anomalies includes:

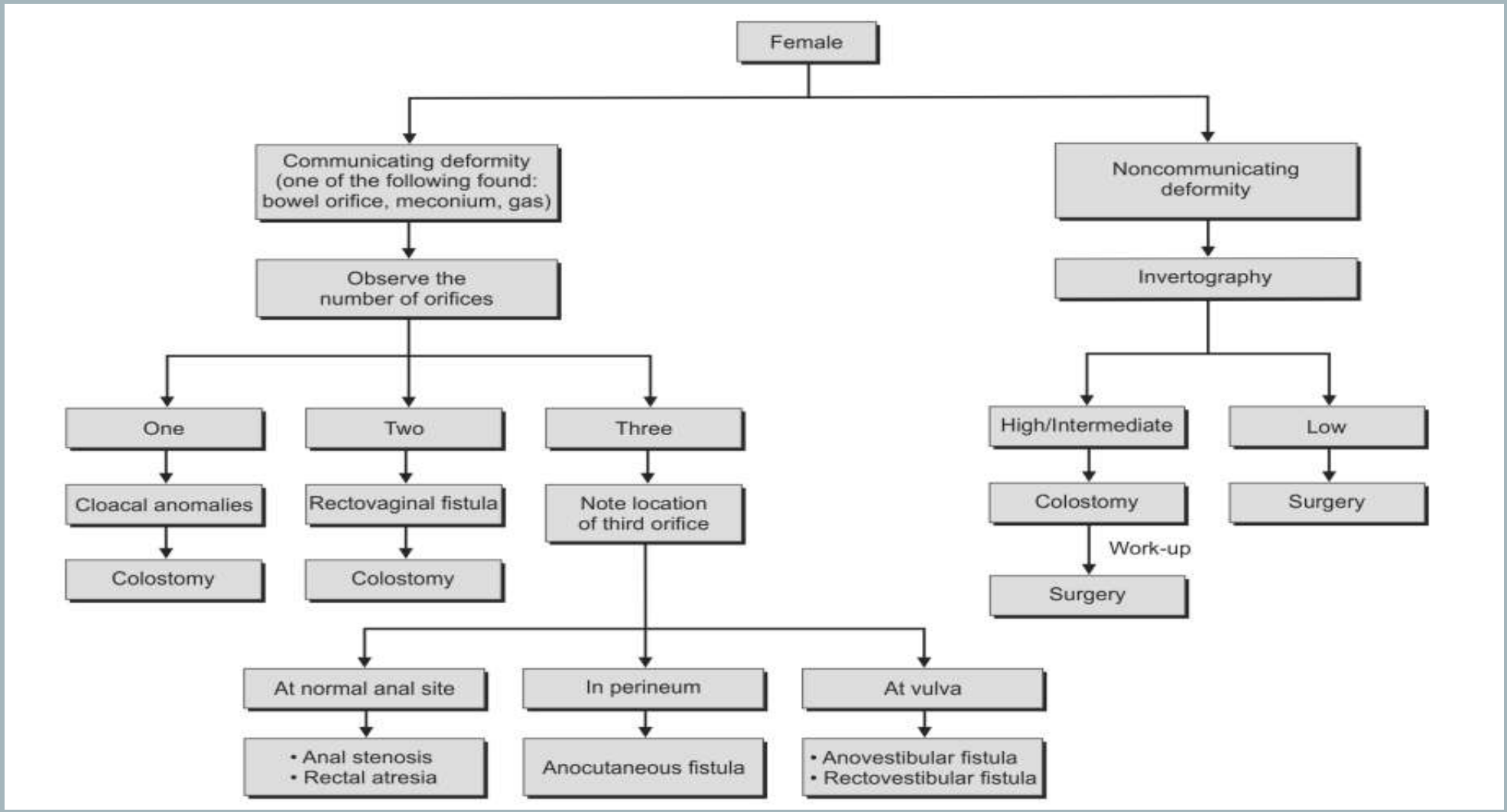
Esophageal atresia
Gross spinal abnormalities (Partial agenesis of sacrum)


a
RECTUM
RECTOPERINEAL FISTULA IN A MALE
b
RECTUM
c
RECTUM

DIAGNOSTIC ALGORITHM IN A MALE ANORECTAL ANOMALIES (ARA)



DIAGNOSTIC ALGORITHM IN A FEMALE ANORECTAL ANOMALIES (ARA)



THANK YOU