Anorectal malformation:

common variants

Samuel Negash M.D.

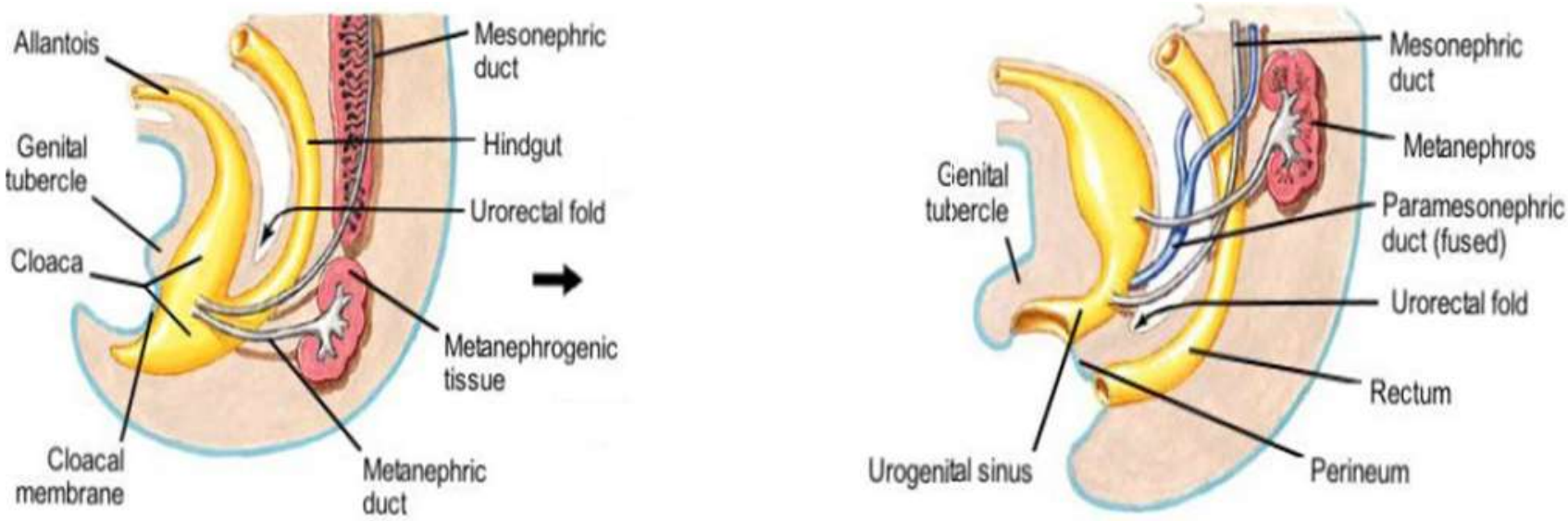
Pediatric Surgery Resident

Addis Ababa University

Outline

1. Basics
2. Newborn Management
3. Definitive Repair
4. Follow-up

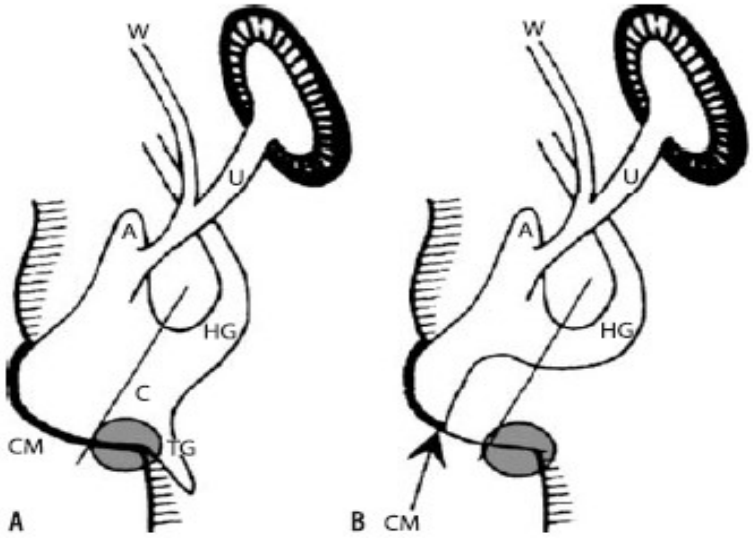
Embryology

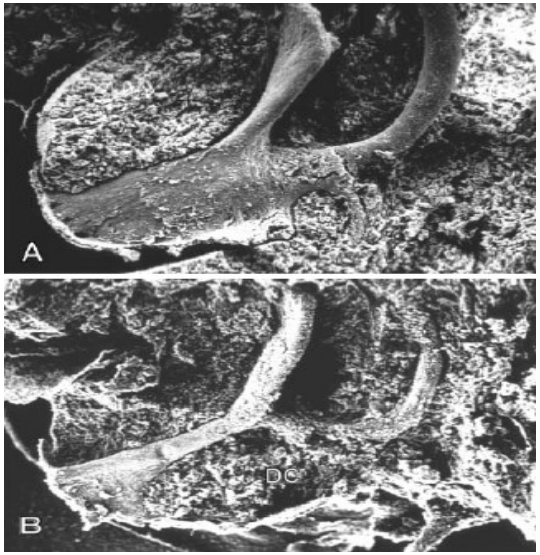


- Theories of ARM based on theories of normal development: failure of lateral fusion of urorectal septum or fusion of septum with cloacal membrane has not been seen in animal studies.

ጥቁር አንበሳ
TIKUR ANBESSA

Embryology





- Studies in Animals: Examination of mammalian embryos (pigs/mice with congenital ARM and SD mice, Adriamycin rat) has found **deficiency in dorsal part of cloacal membrane and adjacent dorsal cloaca**. Cloacal membrane is too short (doesn't extend to the region of the tail groove) and dorsal cloaca is missing. Possible cause could be abnormal infiltration of mesenchyme in dorsal part.

ጥቁር አንበሳ
TIKUR ANBESSA

Epidemiology

- 1 in 4,000 to 5,000 newborns
- Slightly more common in males
- Risk of having a 2nd child with ARM is 1%

Classification

Major clinical groups		Rare/Regional variants
Male	**Female**	
Perineal fistula	Perineal fistula	Congenital pouch colon
	Vestibular fistula	Rectal atresia
Urethral fistula • bulbar • prostatic	Cloaca • short channel • medium channel • long channel	Recto-vaginal fistula
Vesical fistula		H type fistula
Without fistula	Without fistula	Others

- Krikenberg (2005): terms low, intermediate, and high imperforate anus are not used anymore. Historically cloaca considered rare while rectovaginal was commonly reported but now the reverse is true.

ጥቁር አንበሳ
TIKUR ANBESSA

Associated Anomalies

- VACTRL association – at least 3/6
  - **Urologic** – 50% (renal agenesis, VUR)
  - **Cardiac** – 30% (ASD, VSD, TOF)
    - 10% require intervention
  - **Vertebral**
    - Fusion
    - segmentation defect - hemi-v., butterfly v., wedge v.)
    - Dysplasia
    - Number - absent/supernumerary
  - **Esophageal atresia** - 8%
  - **Limb**
- Spinal – 25%
  - tethering- low conus, lipoma, syringomyelia
  - caudal regression- short spinal cord;
  - Currarino syndrome – presacral mass
- Gynecologic
- Chromosomal

ጥቁር አንበሳ
TIKUR ANBESSA

2. Newborn Management

ጥቁር አንበሳ
TIKUR ANBESSA

1st 24 hours of life

Exclude associated anomalies

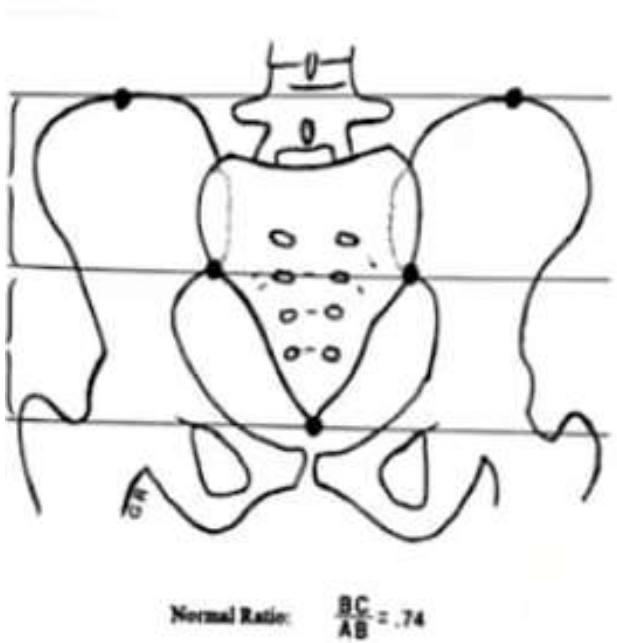
- NG tube
- X-ray - Babygram
- Ultrasound – abdomen and spine
- Echo – can be delayed until after colostomy unless cyanotic

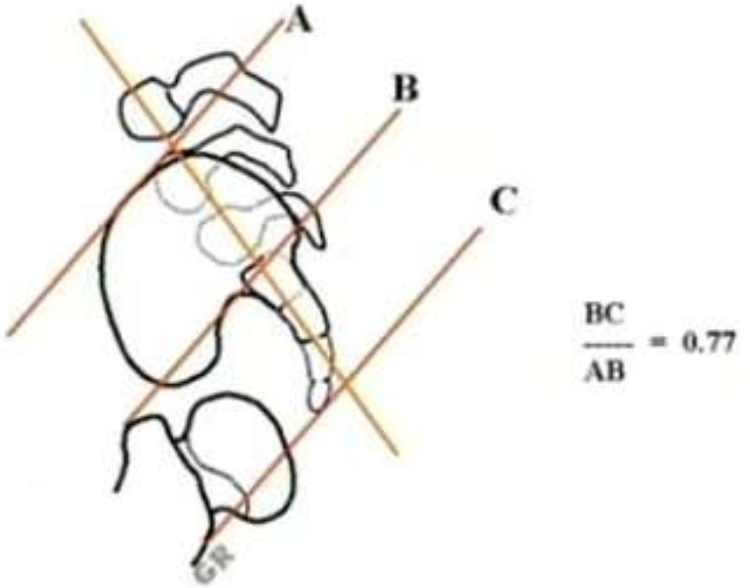
Determine type of ARM

- Physical exam (supine, relaxed, legs bent upwards, spread perineal body, Lift labia)
  - Shape of buttocks gluteal groove
  - Anal dimple (presumed muscle complex)
  - Signs of perineal fistula
  - Number of openings in vestibule

ጥቁር አንበሳ
TIKUR ANBESSA

Sacral ratio

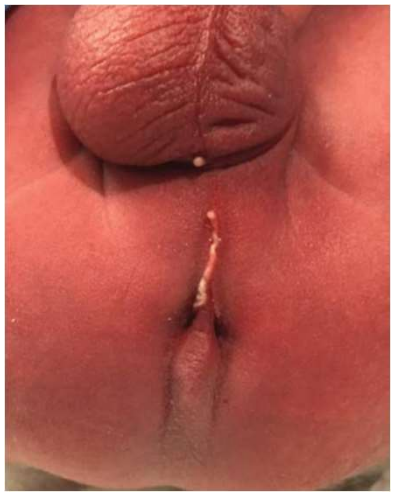
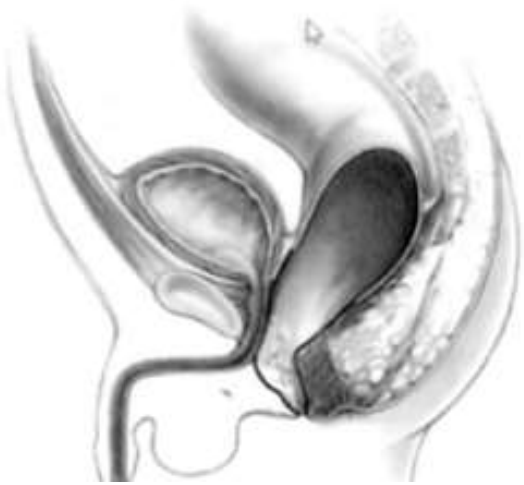
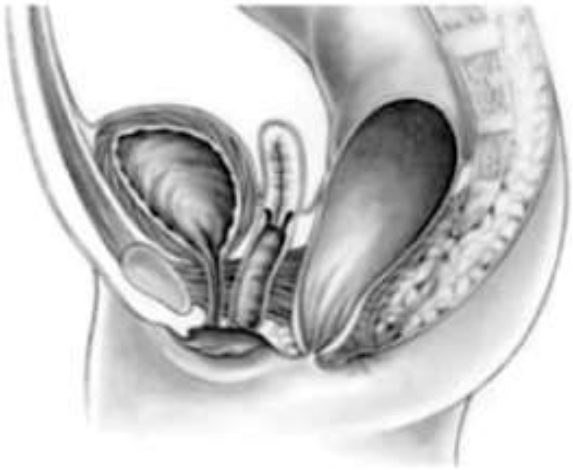
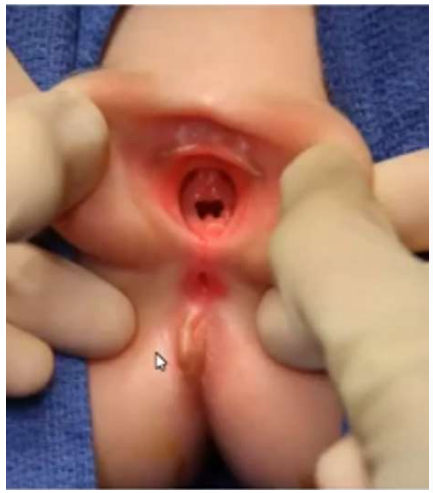




- Sacral X-ray can be used to objectively asses the sacrum (hypoplasia/caudal regression). **AP view shows sacral defects** while **lateral view is best to calculate sacral ratio**. Draw line at top of iliac crest, sacroiliac joint and last sacral bone visible. 0.4-0.7 have intermediate prognosis. Those above and below have good and poor prognosis.

ጥቁር አንበሳ
TIKUR ANBESSA

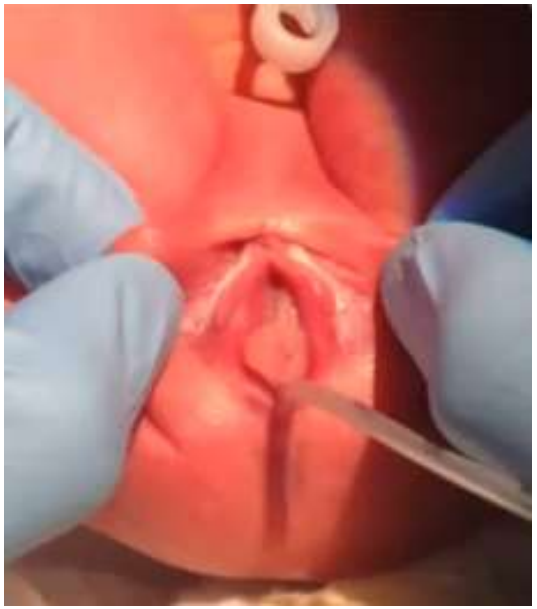
Perineal Fistula

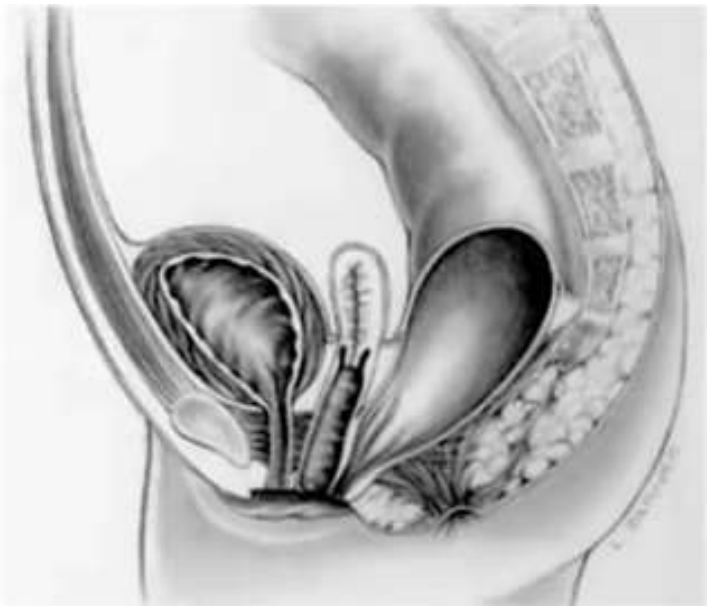





- The most benign form of ARM. Rectum forms fistula anterior to anal dimple. Almost all are continent but suffer from severe constipation. Perineal fistula may be present male patients without obvious perineal opening as a Bucket handle defect or subepithelial tract. In females there might be difficult to differentiate from anterior anus (short perineal body but anus in center of muscle complex) which requires examination under anesthesia

ጥቁር አንበሳ
TIKUR ANBESSA

Vestibular fistula

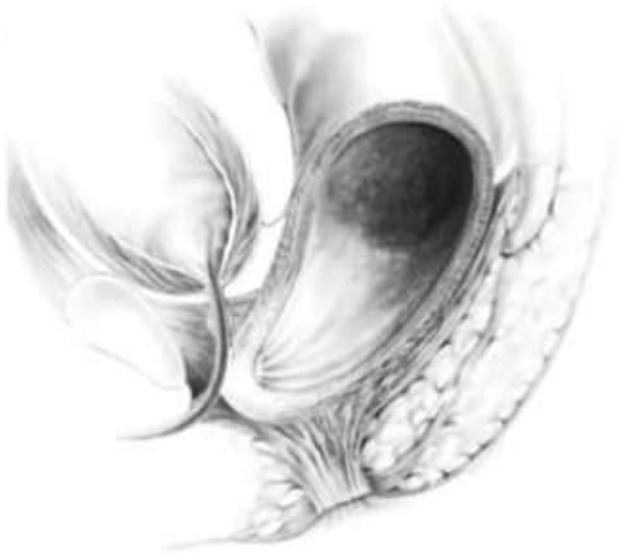




- The most common ARM in females. The fistula is just posterior to the vagina and shares a common wall. Sometimes opening may not be visible but 8F NGT will pass behind the prolapsed/edematous hymen. Around 95% are continent but suffer from severe constipation.

ጥቁር አንበሳ
TIKUR ANBESSA

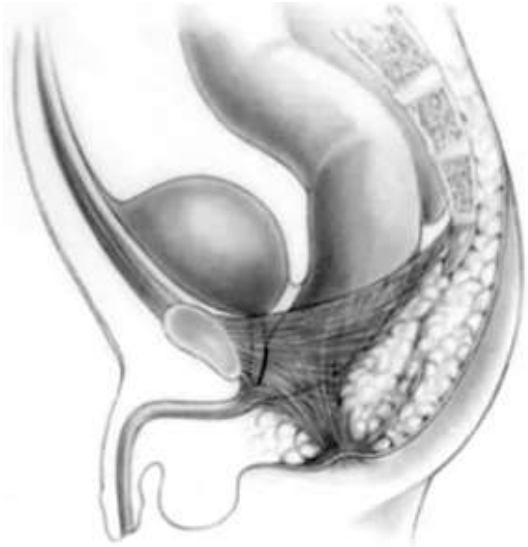
Without fistula

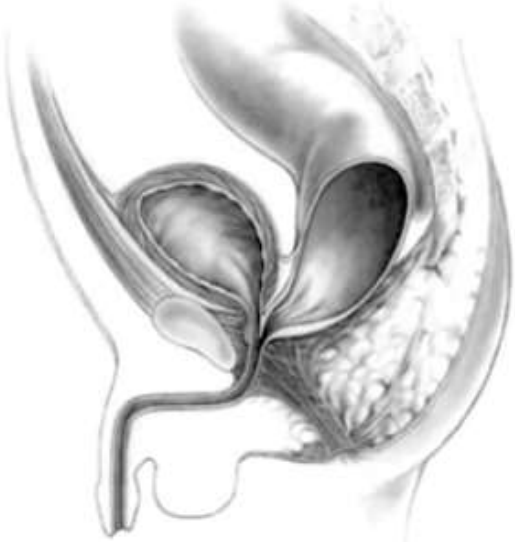


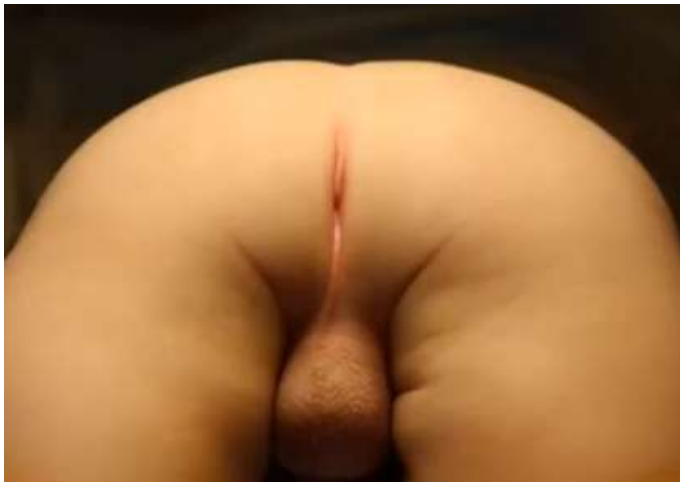
- Relatively rare form of ARM (5%). Commonly associated with Down's syndrome (50% of ARM without fistula have DS. >90% of ARM with DS have no fistula). Perineal exam usually reveals well developed perineum. Even though there is no fistula, the rectum may be closely adherent to the urethra. Even though they have DS, it doesn't interfere with their good prognosis for continence.

ጥቁር አንበሳ
TIKUR ANBESSA

Urethral fistula



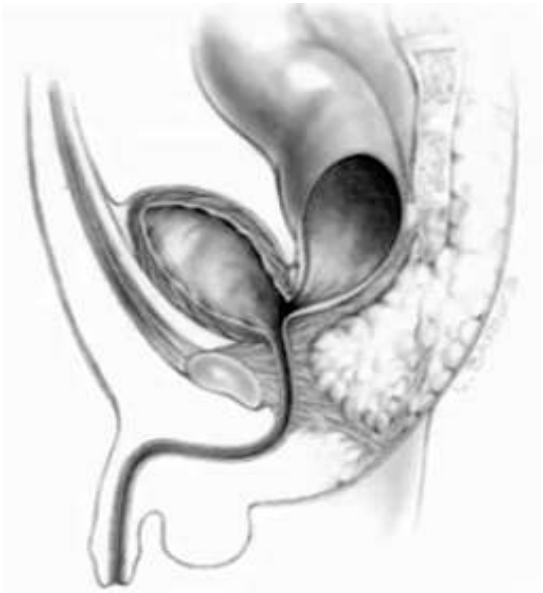


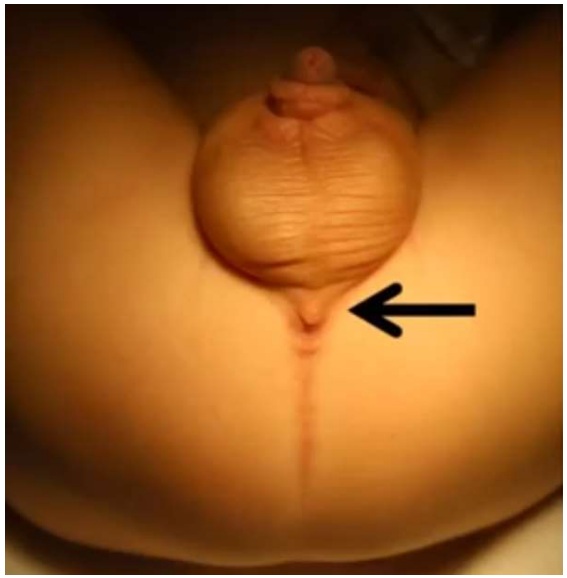


- Urethral bulbar fistula is the most common ARM in males and perineum has good development. Urethral prostatic fistula has a poorly developed perineum (figure) and only 65% achieve continence. Both have common wall with the urethra that requires meticulous dissection

ጥቁር አንበሳ
TIKUR ANBESSA

Bladder neck fistula

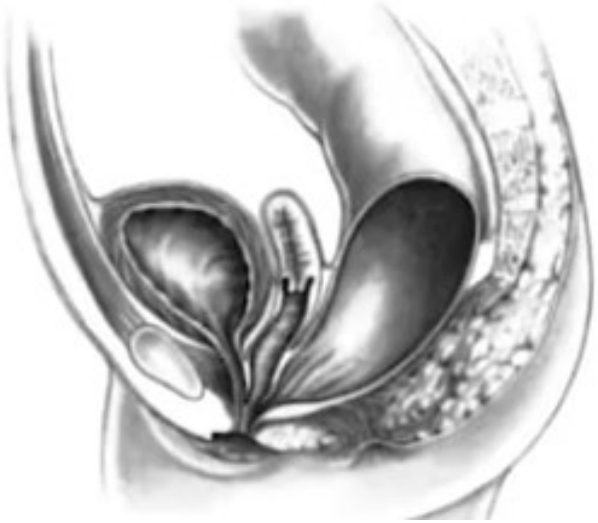


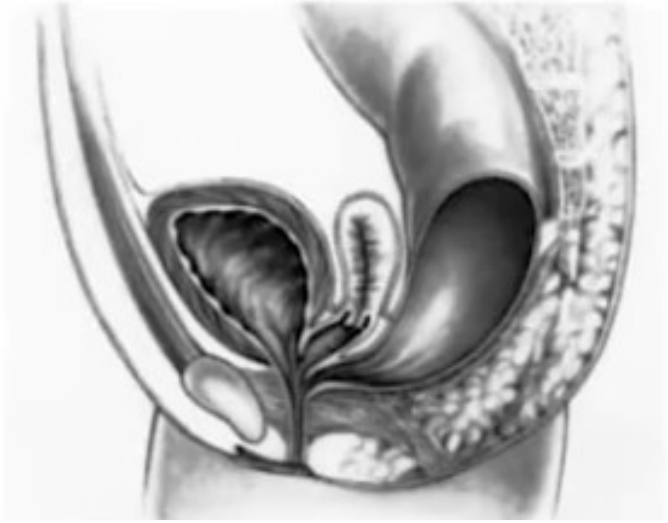


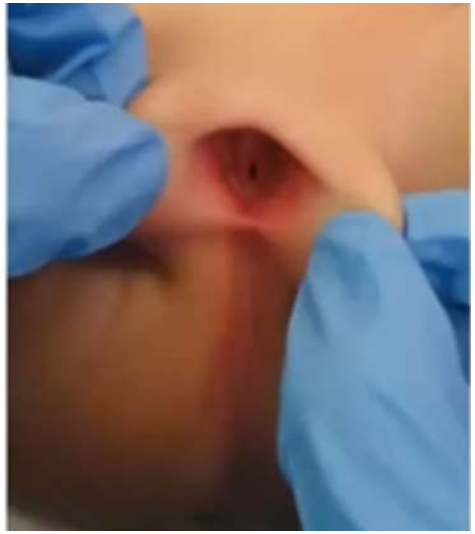
- Highest malformation in males (10%). Rectum enters urinary tract just at the peritoneal reflection, . Perineum is flat with center of the sphincter close to the base of the scrotum. Requires abdominal approach to separate fistula. Prognosis for bowel control is poor (~15%)

ጥቁር አንበሳ
TIKUR ANBESSA

Cloaca







- Highest malformation in females. Short common channel has good prognosis (~70%). Long common channel has poor prognosis with most also requiring CIC.

ጥቁር አንበሳ
TIKUR ANBESSA

20 -24 hours after birth

- Repeat physical exam (distal colonic pressure will delineate a fistula)
- cross-fire film (In cases without visible fistula)

Colostomy

- **Cloaca**
- **High rectum on x ray** (no fistula,) (prostatic/vesical fistula)

Primary Repair

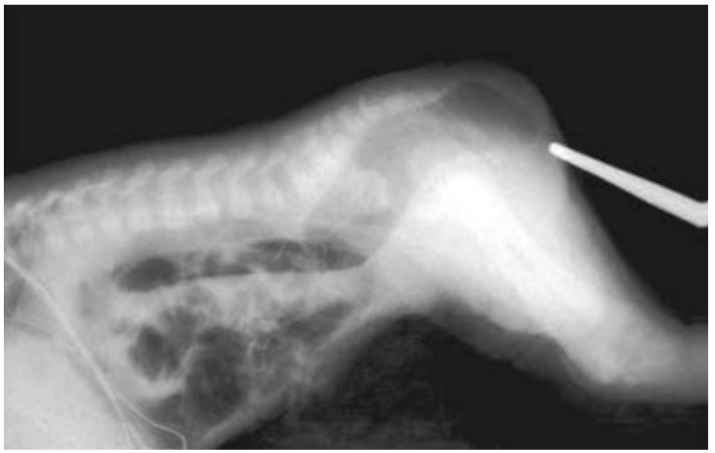
- **Perineal fistula**
- *? Vestibular fistula*
- *Low rectum on x ray* (**no fistula**,) (? bulbar fistula)

Dilatation (delayed repair at 2-3mo + colostomy)

- **Perineal / *Vestibular* fistula**
- ill from associated anomalies
- premature
- surgeon not comfortable with primary repair

ጥቁር አንበሳ
TIKUR ANBESSA

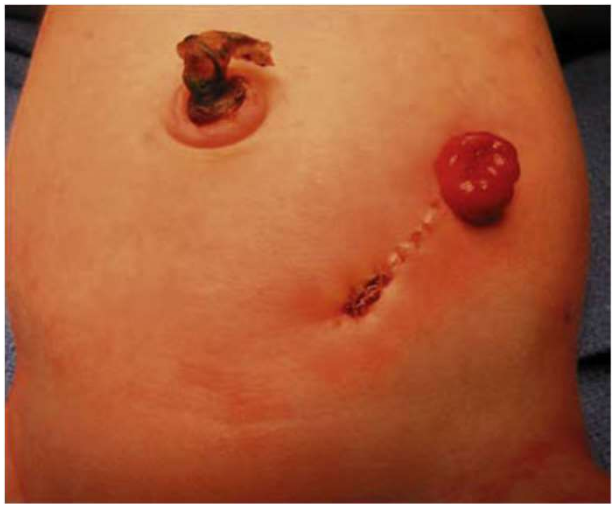
Cross-fire film

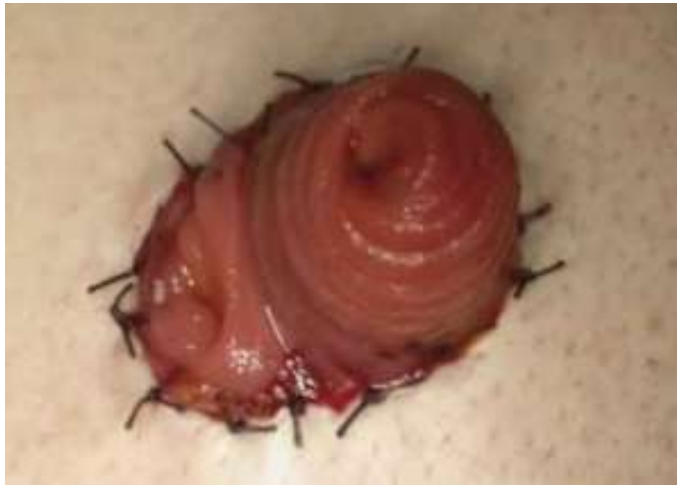


- Historically invertogram was used but currently Prone cross table lateral X-ray should be done if there is no apparent fistula after 24 hours. Roll placed beneath the hips of the infant to elevate the buttocks and allow air to migrate superiorly to the end of the rectum. Low rectum (amenable to primary repair) is below coccyx (in patient with normal sacrum) or close to perineal skin (within 1-2cm)

ጥቁር አንበሳ
TIKUR ANBESSA

Colostomy





- Proximal sigmoid colostomy: Descending sigmoid junction is preferred site to avoid prolapse, avoid excess urine absorption, avoid difficulty during distal colostogram and have adequate length for pullthrough. Stomas should be fully diverting to avoid megarectum and UTI. This can be done via **divided** stoma (mucus fistula with far enough skin bridge to apply stoma bag). It can be done laparoscopically b/n 24-48hr of life (prior to massive abdominal distention) . A **diverting loop** stoma is also acceptable if done with Turnbull technique (distal end simply sutured to skin and proximal end everted like ileostomy).

ጥቁር አንበሳ
TIKUR ANBESSA

Decision making in major co-morbidities

- **ARM + cardiac lesion:** most common is VSD (rarely impact patient undergoing colostomy)
- **ARM + EA/TEF:** passage of air through fistula expedite rectal distention (earlier colostomy)
- **ARM + DA:** colon may not dilate b/c limited meconium (care on selecting bowel for stoma)
  - Marking colon during the transverse laparotomy incision for duodeuno-duodenostomy will help
- **ARM + EA/TEF and DA**: Top down approach (TEF repair >> duodenoduodenostomy >> colostomy)
  - If patient is unstable, duodenal atresia surgery might be done later but a gastrostomy is mandatory
- **ARM + long gap EA**: colonic replacement not preferred (gastric tube/pullup is first line).
  - Co-existing duodenal atresia is beneficial b/c excess stomach volume available for gastric tube

ጥቁር አንበሳ
TIKUR ANBESSA

3. Definitive Repair

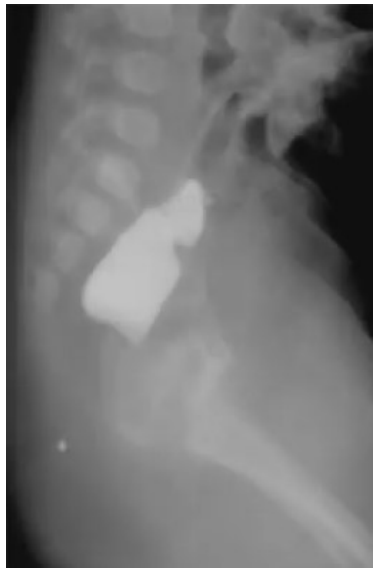
Timing of surgery

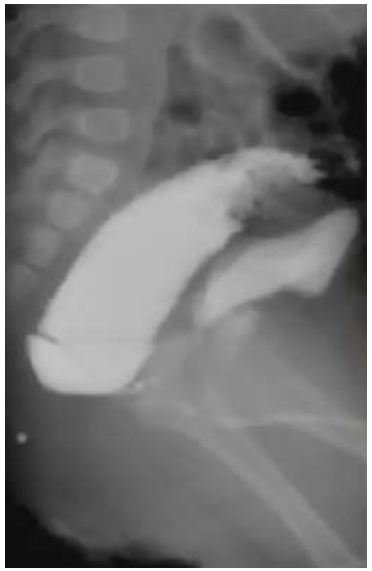
Early in life (2-3 months)

- Less time with stoma
- Less discrepancy b/n proximal & distal bowel during colostomy closure
- Easier anal dilatation
- Potential to improve acquired local sensation

ጥቁር አንበሳ
TIKUR ANBESSA

Colostogram

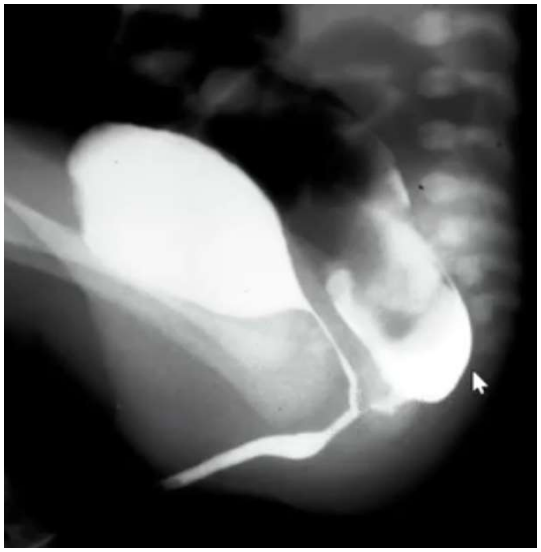
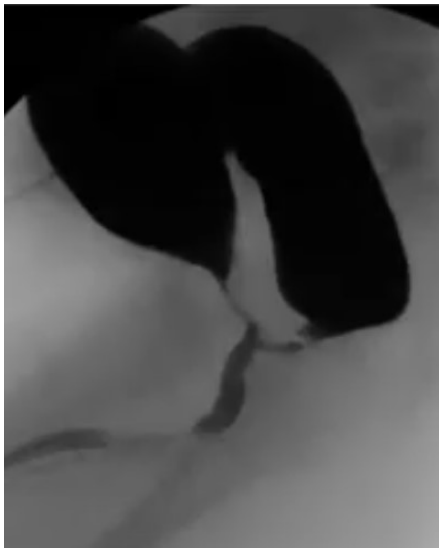
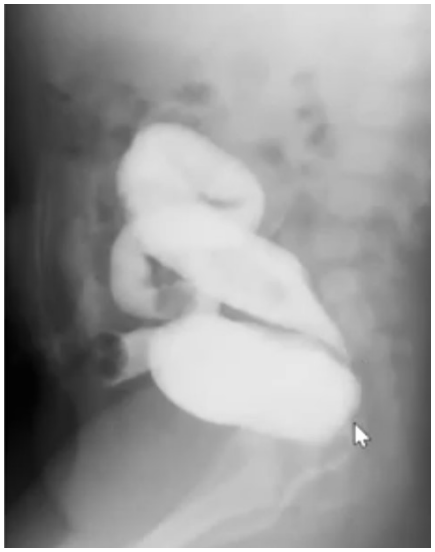




- High pressure distal colostogram technique: in lateral position and marker on anal dimple, insert foley catheter through distal stoma and inflate. Inject contrast with high pressure. If rectum stops flat, it indicates the pressure is not adequate to overcome muscle at pubococcygeal line. With more pressure sphincter is overcome and bladder is filled with contrast. Additional film is needed with child voiding to visualize the urethra. If fistula is below sphincter (bulbar) the distal urethra may be filled with contrast preferentially (difficult to fill bladder).

ጥቁር አንበሳ
TIKUR ANBESSA

Colostogram

- Road map for surgery (determine approach): Elbow can be used to symbolize urinary tract fistulas. Fistula opening below curve is bulbar fistula (rectum bulges through incision when levator is divided). Fistula above curve is prostatic (rectum found higher –close to coccyx during PSARP). Fistula higher up is bladder neck, which is usually above pubo-coccygeal line (requires abdominal approach). A high prostatic fistula may also be approached laparoscopically if distal rectum is narrow. Amount of bowel length available for the pullthrough is also evaluated on the colostogram.

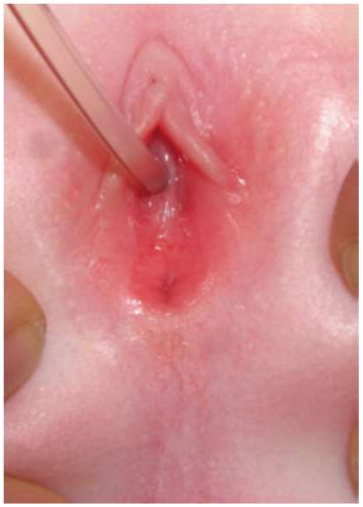
ጥቁር አንበሳ
TIKUR ANBESSA

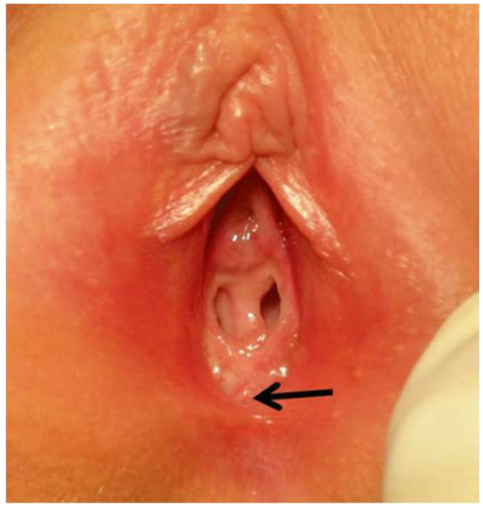
Bowel preparation

- Literature regarding bowel prep in children is lacking.
- There are single center studies which have shown no benefit of bowel preparation.
- However, mucosa-skin anastomosis is higher risk and needs mechanical preparation
  - **Children without colostomy (**primary PSARP**)**
  - **Children with short distal stoma limb** (proximal colon needed for reconstruction)
  - Avoid impaction/hard stools post-op as this may cause stress on the anastomosis.

ጥቁር አንበሳ
TIKUR ANBESSA

Vaginoscopy / Cystoscopy

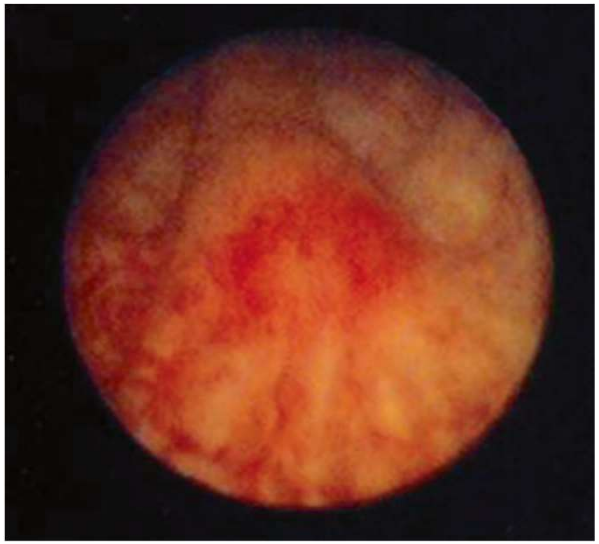




- Routine vaginoscopy should be performed during definitive repair because **17% of ARM with RVF** have associated utero-vaginal anomalies. Absent vagina may be treated by pullthrough if proximal is dilated or need replacement (8% also have absent mullerian). Vaginal septum is always associated with hemicervix/uterus. It should be resected at same surgery if diagnosed. If diagnosis is missed it may have significant gynecologic and obstetric implications.

ጥቁር አንበሳ
TIKUR ANBESSA

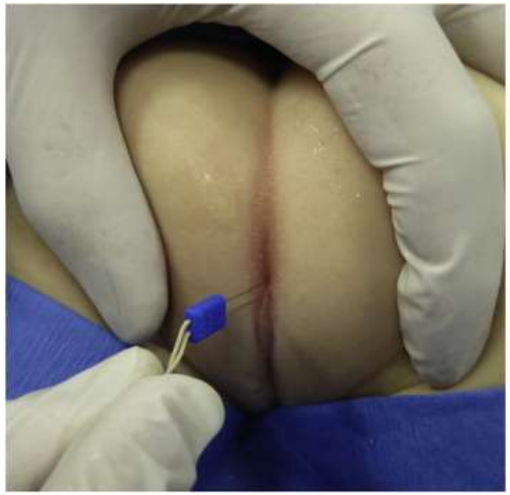
Vaginoscopy / Cystoscopy



- Routine Cystoscopy in boys has shown an anomaly previously not known- ectopic veromontanum (45% in bladder neck fistulas). It may be located on bladder neck or around trigone. These patients ejaculate into the bladder and require artificial assistance for fertility (retrive sperm from urine).

ጥቁር አንበሳ
TIKUR ANBESSA

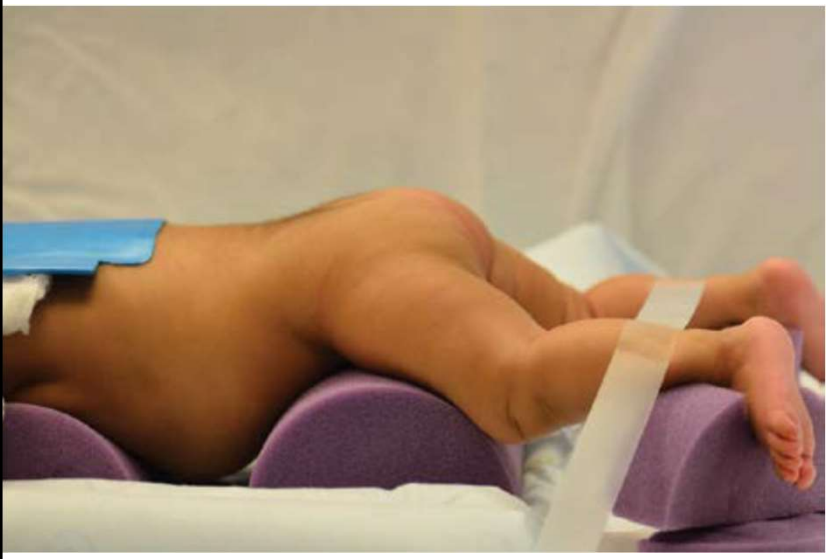
Electrical stimulation

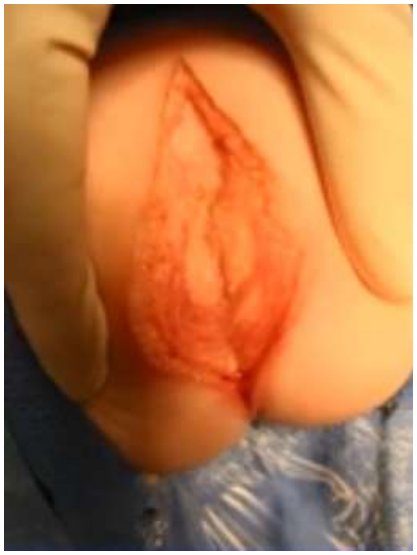


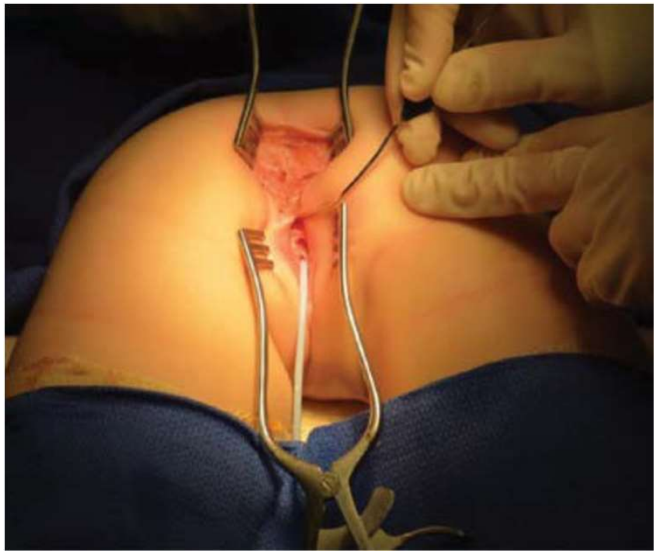
- **Very important to identify muscle complex.** To obtain the best contraction no muscle relaxant should be given and a continuous current should be used. The tip of the stimulator is placed on perineal skin and moved along the midline until no contractions are visible. the best contraction is expected to be where the anal dimple is visible and/or where the skin has a different color, generally pinker and darker than the rest of the intergluteal fold. The muscle complex of an infant is usually 2–3 cm long and there may be right and left asymmetry. The sphincter should be marked prior to starting the operation.

ጥቁር አንበሳ
TIKUR ANBESSA

PSARP



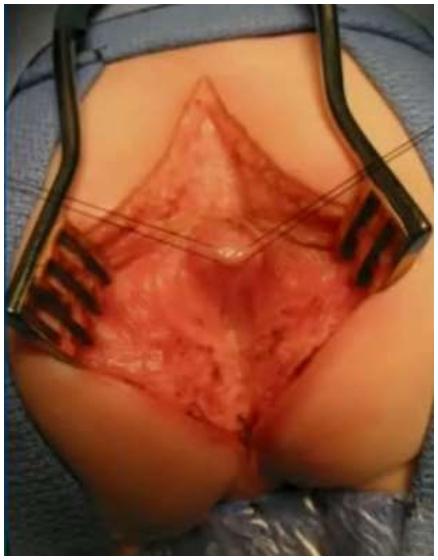
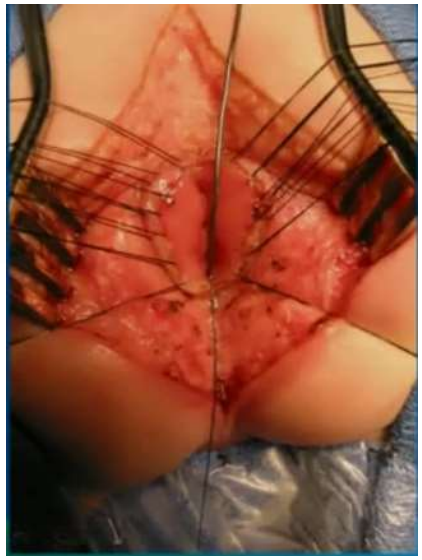
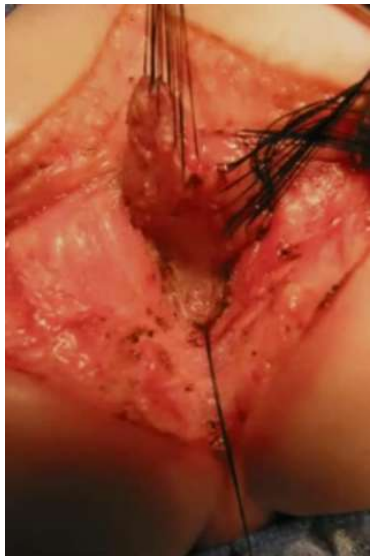
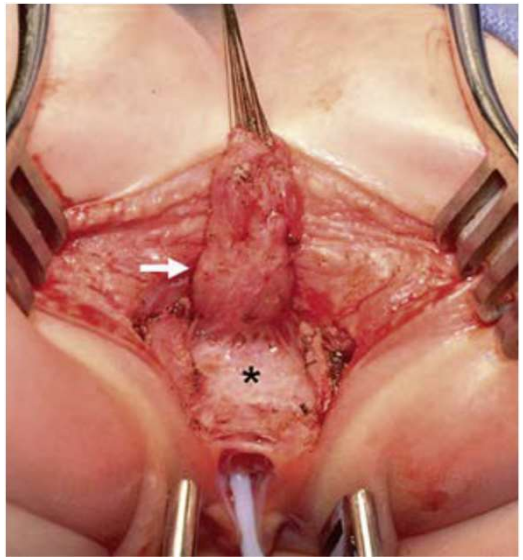




- **In prone position** with an indwelling urinary catheter placed (may require cystoscopy/guidewire in males) before turning the patient and a roller pad underneath the pelvis. A midline incision is made from coccyx to fistula or to base of scrotum *(perineal fistula requires minimal incision ~2cm).* Weitlander retractors or disposable hooks are used for symmetrical exposure. Incision is continued splitting parasagittal fibers, muscle complex & levators in midline. *(levators uslally not opened in RPF & RVF)*

ጥቁር አንበሳ
TIKUR ANBESSA

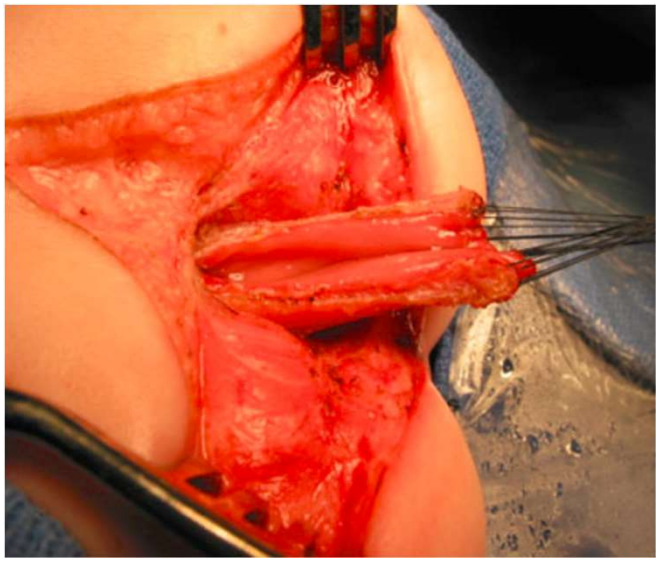
PSARP

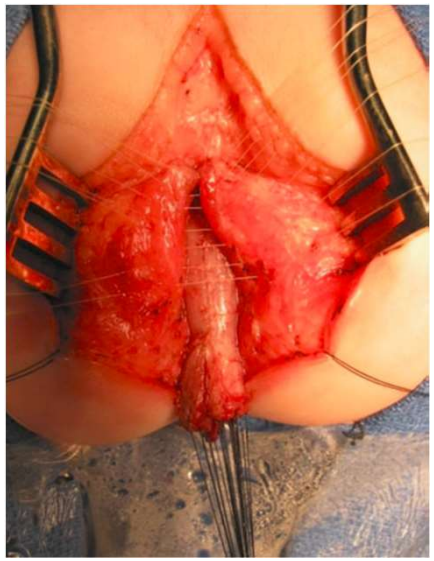





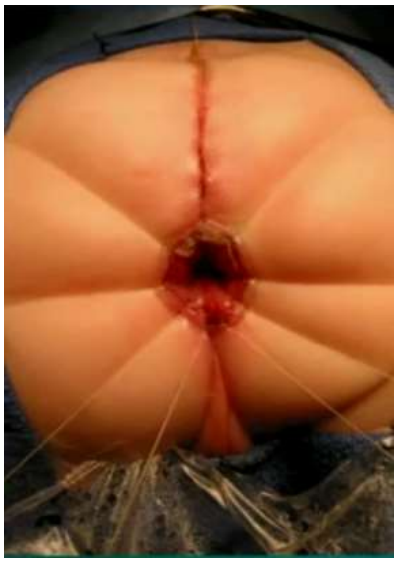
- Rectum is identified below coccyx and held with stay stich. Rectum is opened distally adding stay sutures until fistula to urinary tract is exposed. Then place stay sutures in midline above fistula and divide. Continue dissection in submucosal plane to separate the common wall between rectum and genitourinary tract (important in RVF to avoid tension).Full separation realized when find areolar plane in between. Then ligate the fistula.

ጥቁር አንበሳ
TIKUR ANBESSA

PSARP



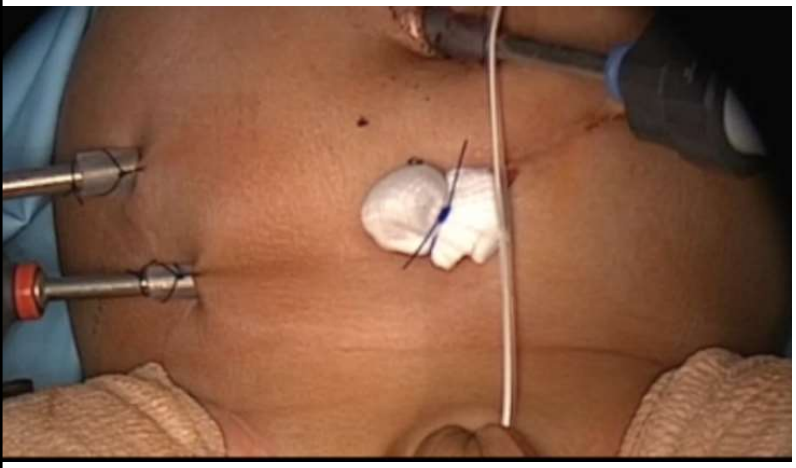


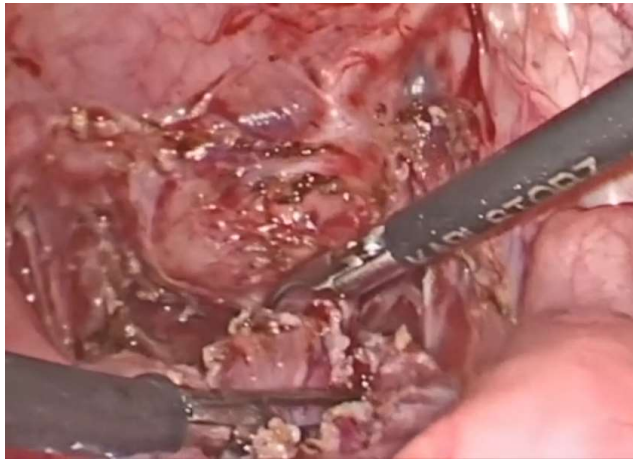


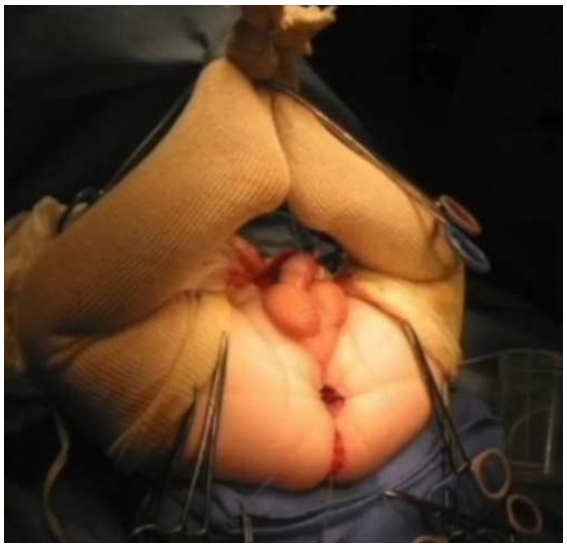
- White fascia over rectum is opened to mobilize rectum close to its wall without injuring it. Divide and burn extrinsic vessels and bands while applying traction until enough length is gained (up to peritoneal reflection). Anterior rectal wall is repaired and Approximate muscle complex taking bite of posterior rectal wall (rectopexy). Rarely rectum might require tapering bu removingpart of posterior wall and suturing. Neo anus fashioned in center of muscle complex with absorbable stiches, taking full thickness bite of skin and rectum, placed under slight tension. Initial stiches are placed in the four cardinal points then 3 stiches added per quadrant (total of 16 stiches).

ጥቁር አንበሳ
TIKUR ANBESSA

Laparoscopy assisted PSARP



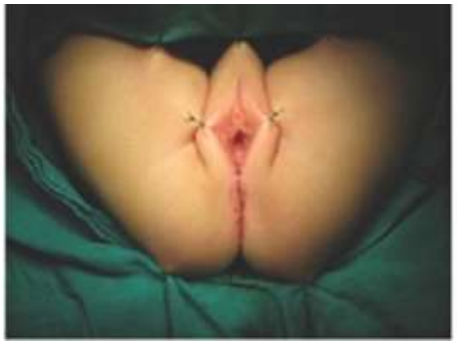
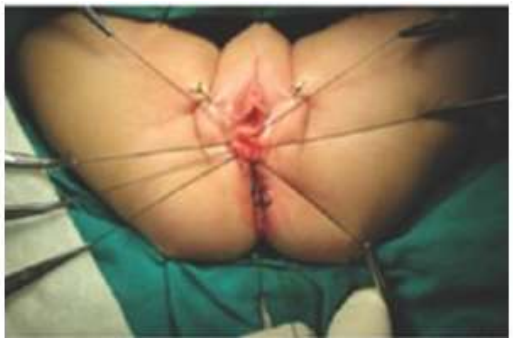
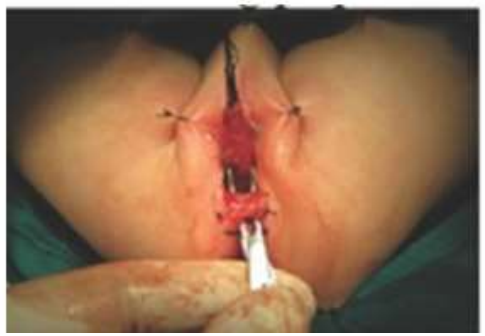
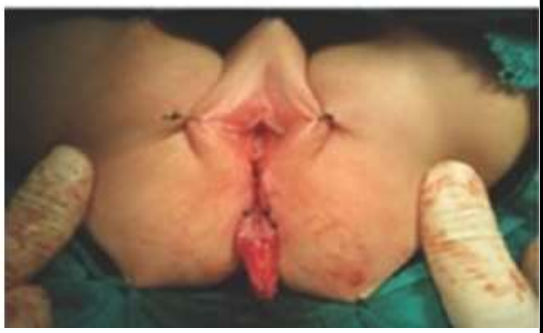




- **LAARP**: In supine position laparoscopy ports inserted. Bladder fixed to anterior abdominal wall for exposure. Distal rectum dissected close to its wall until caliber of rectum becomes thin as the dissector. Fistula is either transfixed or ligated with endoloop (clips not recommended). Patient leg is placed up, PSARP incision made and hegar dilator used to deepen dissection to reach pouch of douglas. Rectum is pulled thorough bluntly created tract. Anoplasty then done in supine position.

ጥቁር አንበሳ
TIKUR ANBESSA

ASARP

- Various modifications have been made to PSARP for RVF to preserve perineal skin bridge and levator muscle (Anal transposition, NSARP, TFARP).
- ASARP modification shows spreading of vestibule, mobilizing fistula and transposing rectum in center of muscle complex under direct vision.

ጥቁር አንበሳ
TIKUR ANBESSA

4. Follow up

Postop care

- **Antibiotic**
  - IV broad spectrum (prophylactic) – can be continued 24 hr postop
  - Ointment (optional) – applied to sutures on perineum for 7 days
- **Feeding**
  - Protective colostomy – feed after recovery from anesthesia (same day)
  - Primary PSARP for perineal fistula in neonates – feed after 48-72 hr
  - Primary PSARP for RVF or for RPF in older infants – feed after 5-7 days
- **Catheter** - If catheter falls off it should not be re-inserted
  - rectovestibular or perineal fistula - 24 hours
  - rectourethral fistula – 5-7 days
  - Rectovesical fistula, additional urethral repair – 10 - 14d
- **Urologic evaluation** – ultrasound at 3 months postop, other tests as needed

ጥቁር አንበሳ
TIKUR ANBESSA

Dilatation

Size of dilator according to age	
Age	**Hegar size (mm)**
1-4 mo	12
4-8 mo	13
8 -12 mo	14
1-3yr	15
3-12 yr	16
>12 yr	17

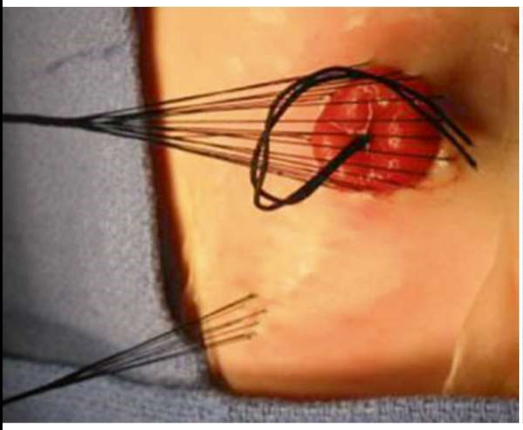
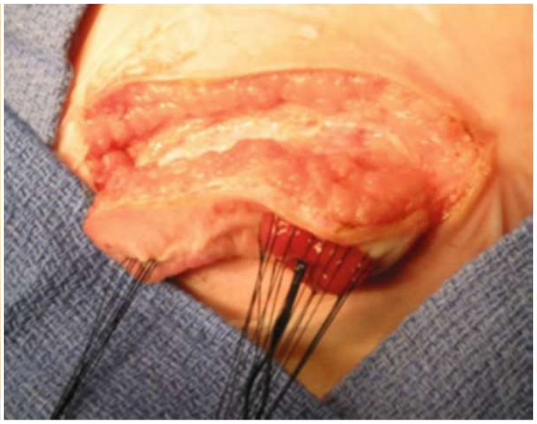
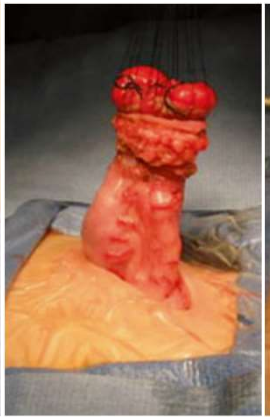
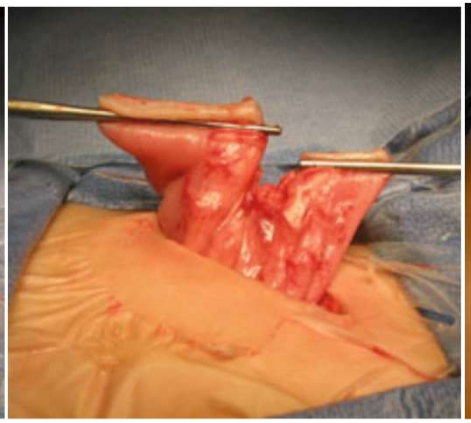
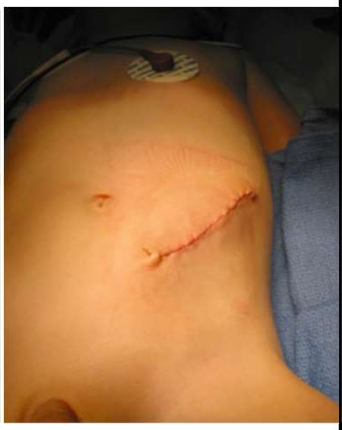
The size of the anal orifice in the neonate

= 1.3 + (BW in kg × 3)

- Dilatation is started 2 weeks with Hegars or other available tools (candles). Initiate dilatation bi-daily with smallest size anus can admit and increase size weekly until target is reached. Once adequate size is reached colostomy can be closed but daily calibrations are recommend for a couple more months. Should not be painful and bleed after first few days.

ጥቁር አንበሳ
TIKUR ANBESSA

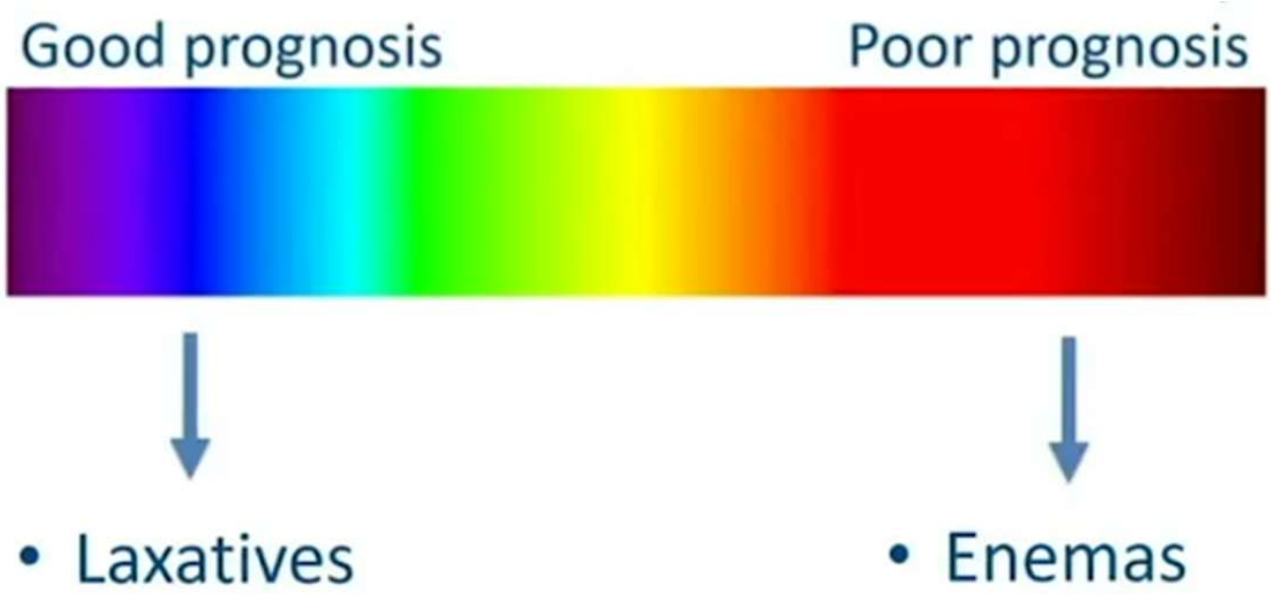
Colostomy closure

- Multiple silk sutures are placed at the mucocutaneous junction of both stomas. Traction is applied and wedge incision made to separate stoma from abdominal wall. Stomas are resected to use a fresh portion of bowel to perform anastomosis.

ጥቁር አንበሳ
TIKUR ANBESSA

Outcome



- Bowel movement becomes more regular at about 3–6 months after colostomy closure. The diet is tailored to ensure 1-3 bowel movements each day. A patient is likely to be toilet trained if he/she has 1-3 bowel movements per day, is clean in between bowel movements, and feels the urge to push. Patients with good prognosis suffer from constipation. Poor prognosis patients suffer from incontinence. At least 25% have true incontinence and require bowel management.

ጥቁር አንበሳ
TIKUR ANBESSA

Check points

- *Neonatal age – rule out associated anomalies, plan the future*
- **Early age** (colostomy closure, weaning) - vigilance to diagnose constipation
- **Toilet training age** (4yr) – asses continence, start bowel management for some kids
- Beginning school – avoid accidents, psycho-social evaluation
- Vacation trials – Laxative for those with borderline bowel control to stop enema
- Older, independent – antegrade enema procedure for those requiring enema
- Puberty, later life – check on fertility, sexual and reproductive function

**ideal follow-up - once a year to confirm bowel management continues to work*

ጥቁር አንበሳ
TIKUR ANBESSA

Thank You!

ጥቁር አንበሳ
TIKUR ANBESSA