Anorectal malformations

Complications

and Redo Operations

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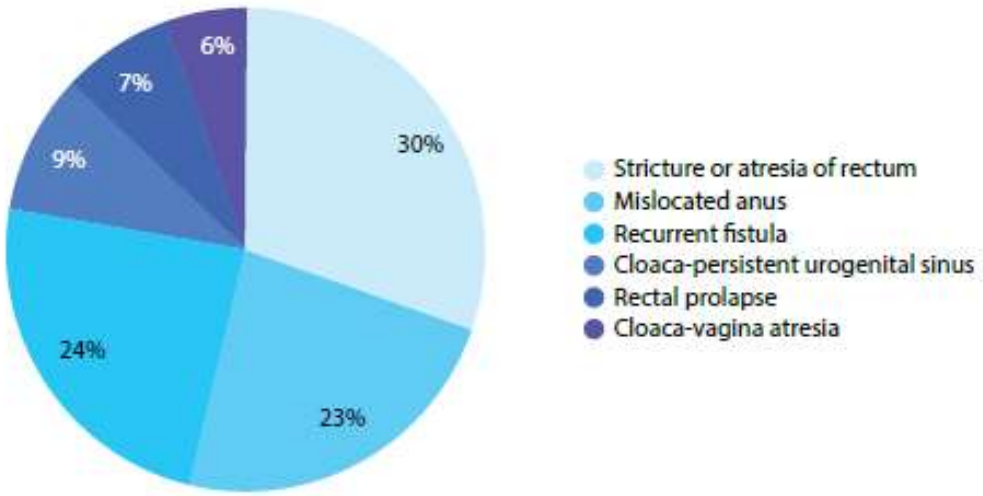
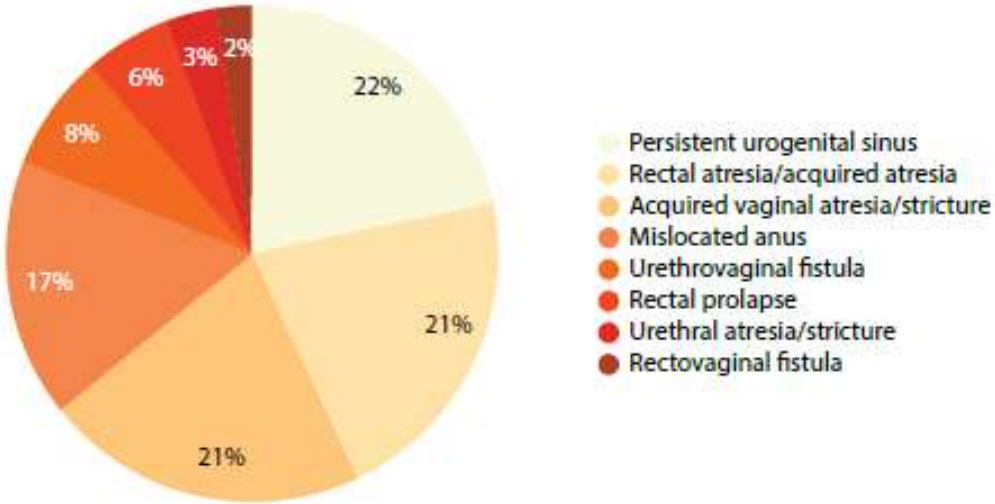
Introduction

- Despite advances in surgery for ARM, complications are common
  - Mainly due to underlying malformation
  - Significant portion are iatrogenic (preventable)
- Best result when proper surgery done during first procedure
- Reoperation may be considered for several reasons

Pena A et al. *J Ped Surg* 2007; 42: 318–325



Levitt MA et al. J Ped Surg 2011; 46: 1250–1255



Common indications for redo surgery in ARM are **rectal stricture, mislocated rectum and recurrent fistula**. Rectal stricture and mislocated rectum are also common indications in cloaca in addition to **persistent urogenital sinus, and vaginal stricture.**

OUTLINE

1. Functional problems
2. Failed Repair (Catastrophes)
3. Urogenital complications
4. Fistula complications
5. Rectal prolapse
6. Other

1 FUNCTIONAL PROBLEMS

- ✓ Constipation
- ✓ Incontinence

Constipation

- Most common complication
- More common in good prognosis ARM
- **Related to preop rectal dilatation** (inadequate irrigation of distal stoma, loop colostomy, transverse colostomy)
- distal colon dilates if inadequately managed (vicious cycle)
- Treat first with large volume enema until colon disimpacted. followed by daily laxatives.
- May require sigmoid resection if laxative requirement is high

Missed presacral mass

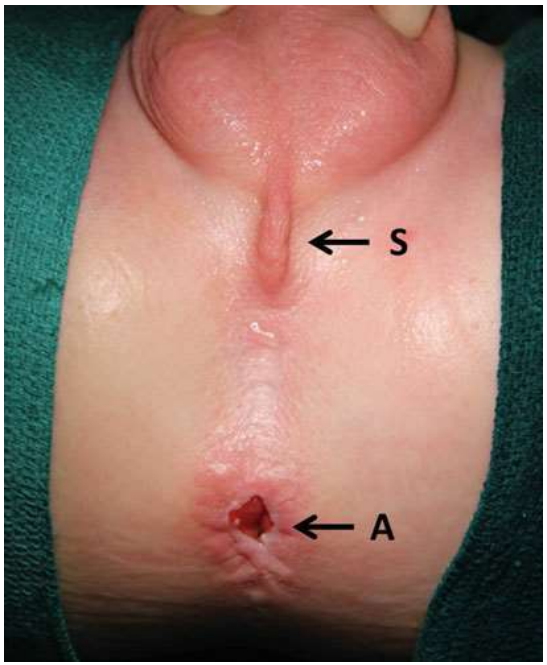
- Present with severe constipation with megacolon
- Can be teratoma/dermoid, lipoma, meningocele, or a combination of all of these.
- Associated with ARM with RPF and hemi-sacrum
- AP view of sacrum shows sacral defect
- Repair requires resection of mass and mobilization of normal rectum above the narrow area

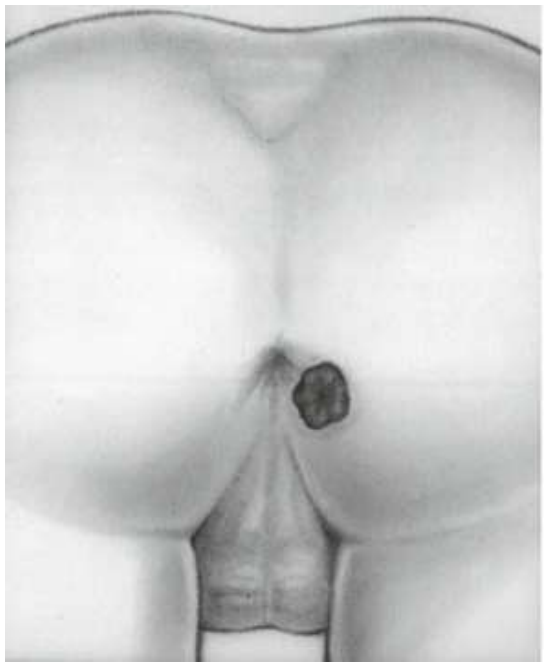
Incontinence

- Evaluate patients thoroughly b/c there may be multiple causes
- Borderline incontinence require **regulation of colonic motility**
  - Diarrheal type – meds to slow down colon & constipating diet
  - Constipating type – laxatives (after dismipacting enema)
- Mislocated rectum may require **surgery**
- Poor prognosis ARM requires **bowel management**

Mislocated rectum

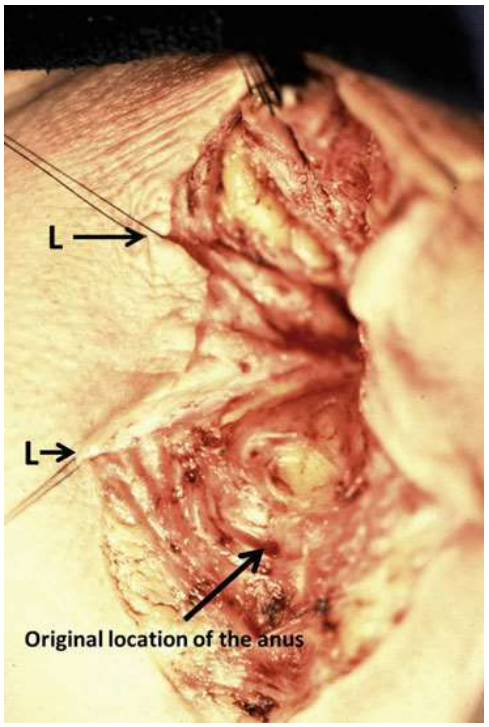
- Initially all patients with incontinence were re-operated with PSARP but only 30% had significant improvement
- Surgical indications were modified to only those with
  - Complete mislocation
  - Good prognosis ARM (RPF, RVF, bulbar fistula, no fistula)
  - Normal sacrum and no tethered cord'
- Be very clear to parents that we can't guarantee recovery (with new indication 2/3 improved but 85% still soil underwear)



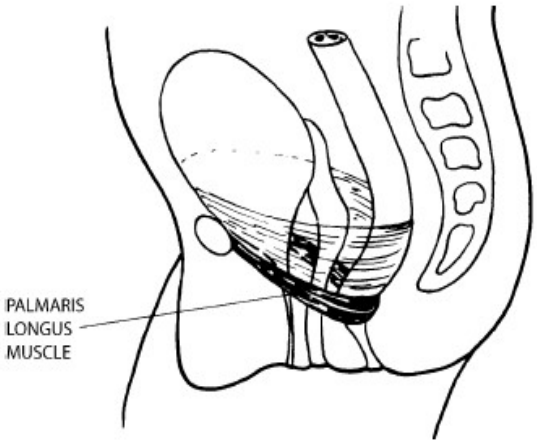


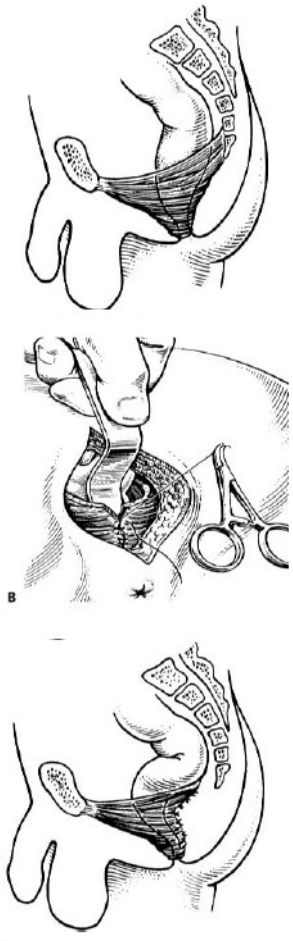
Mislocated rectum is commonly anterior followed by posterior. Lateral mislocation is rare. Mislocation can be total or partial.

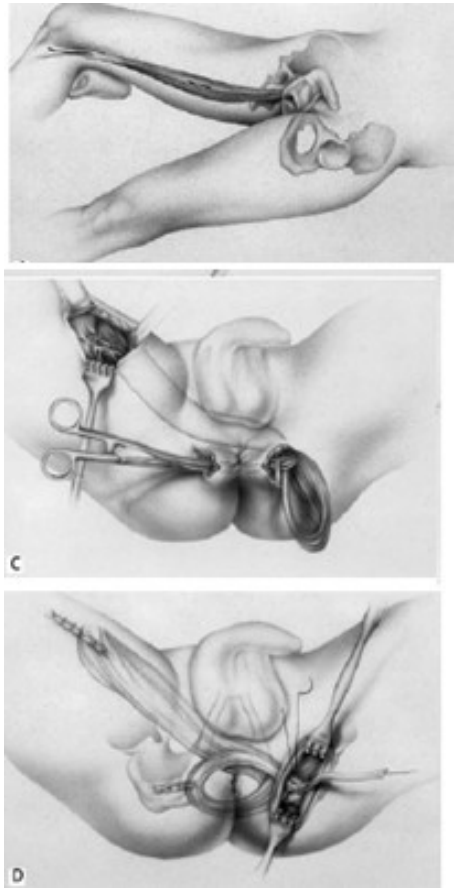
Inadequate perineal body and narrow anus not centered in muscle complex may occur if Inadequate mobilization of rectum from vagina in RVF



Reoperation for mislocated rectum via posterior sagittal incision rectum is mobilized, limits of sphincter are electrically determined and anoplasty done with anchoring to sphincter mechanism. Surgery is easier since most of these patients did not suffer from infections (anatomy clear), not necessary to separate the rectum from urogenital structures, no fistula, no problem of bowel length.







Other Reoperations to improve incontinence many procedures have been suggested with questionable results. "Missed puborectalis". Levatorplasties. "Gracilis sling" sphincter substitution). Artificial sphincters have also been used

Missed tethered cord

- Incontinence can be 2ry to tethered cord..
- Tethering may be found in 10-50% of ARM
- Diagnosis will be missed unless screened
- Decision for surgery (dethetering) is controversial
  - most recommend prophylactic surgery unless discovered late (>10yr)
  - Relationship of tethering to fecal/urinary symptoms in ARM is questionable (maldevelopment of sacral innervation)
  - Constipation is not an indication for unthetering

2

FAILED REPAIR (CATASTROPHES)

- ✓ Infection/abscess
- ✓ Dehiscence / retraction
- ✓ Rectal stricture

Wound Infection / abscess

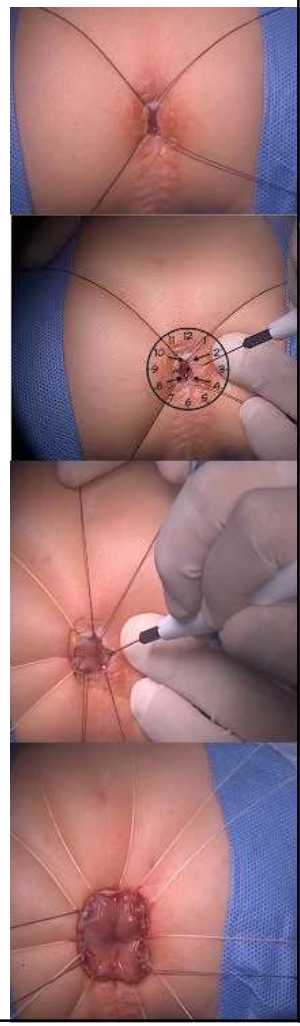
- **Usually affects skin and subcutaneous tissue only**
  - Fecal contamination (1ry surgery, loop colostomy)
  - May be re-sutured in the immediate postop period
  - may heal with 2ry intention without functional sequalae
- **May lead to dehiscence/retraction** >> incontinence, strictures and, recurrent fistulas.
  - Severe pelvic fibrosis “frozen pelvis” or “cement pelvis”
  - struggle to find the rectum and to mobilize it adequately

Dehiscence and retraction

- Combination of
  - **Excessive tension** (inadequate mobilization / colostomy created too distally)
  - **Ischemia**
  - Infection
  - *Mucosal only sutures (anoplasty should be full thickness).
- *May lead to severe stricture or disappearance of anal opening*
- Reoperation requires PSARP
  - rectum high in pelvis surrounded by fibrosis)

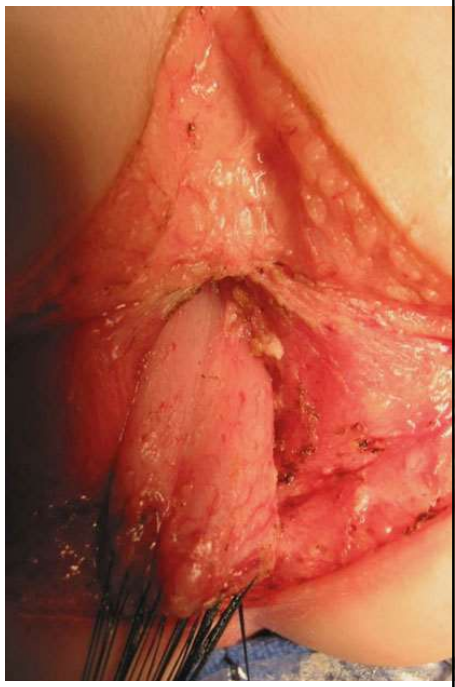
Stricture

- Short (<1cm) ring like strictures
  - Occur if parents **didn't follow dilatation protocol**.
  - Can be treated with Heineke-Mikulicz type of plasty
- Longer strictures
  - result of **ischemia** to distal rectum
  - Redo PSARP (rectum mobilized until fibrotic portion can be removed)



Preventing catastrophic comp

- **mobilization:** difficult if working outside the fascia
  - *The rectum is covered by a very characteristic white fascia that contains vessels to the rectum. Surgeon must dissect fascia off the rectum, remaining as close as possible to the rectal wall. Bands and the extrinsic rectal blood supply must be divided to gain rectal length.*
- **ischemia**: occurs if dissection is too close to rectum
  - *Intramural blood supply of the rectum is excellent; and the rectum can be dissected to gain significant length provided the wall is not injured.*



3 Urologenital problems

- ✓ Injuries
- ✓ Neurogenic bladder
- ✓ Urethral stricture
- ✓ Vaginal stricture

Urogenital injuries

- More common in high prostatic/bladder neck fistulas
- Not knowing anatomy (**Lack of distal colostogram**)
- Includes injuries to
  - Bladder neck, Urethra, or ectopic ureter
  - Vas deferens, Seminal vesicles, Prostate
  - nerve damage (neurogenic bladder & impotence)

Neurogenic bladder

- **Rare in male ARM**
  - Related to very abnormal sacrum and spine anomalies
  - Due to denervation of bladder & bladder neck during dissection of rectum (wrong plane) or not staying in the midline
- **Related to cloaca-** deficient emptying mechanism
  - very good bladder neck with floppy flaccid bladder (bladder smooth and large not typical "Christmas tree")- require CIC
  - Bladder neck affected in patients with very long common channels – require continent diversion

Urethral stricture

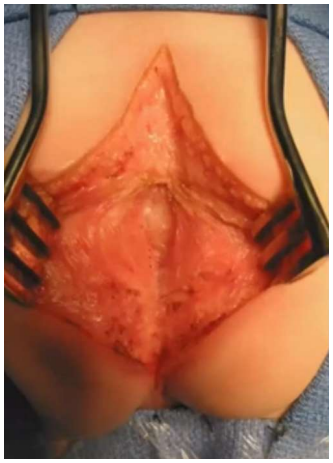
- **Cloaca-** ischemia of neourethra
- **Male** – accidental transection of urethra
    - Redo PSARP with rectum mobilization to expose urethra. Both urethral ends identified and anastomosed

Vaginal stricture

- Tension or Ischemia on mobilizing high vagina
- Excessive dissection during separation of vagina from urethra
- Same approach as for persistent urogenital sinus

Preventing Urologic problems

- **proper (high pressure) distal colostogram**
  - *flat distal rectum at level of pubococcygeal line, indicates low-pressure (inadequate) study. Distal rectum should be bulging under pressure before the surgeon can make this diagnosis before making diagnosis of no fistula*
- **Stay in the midline** (nerves go laterally)



4 FISTULA COMPLICATIONS

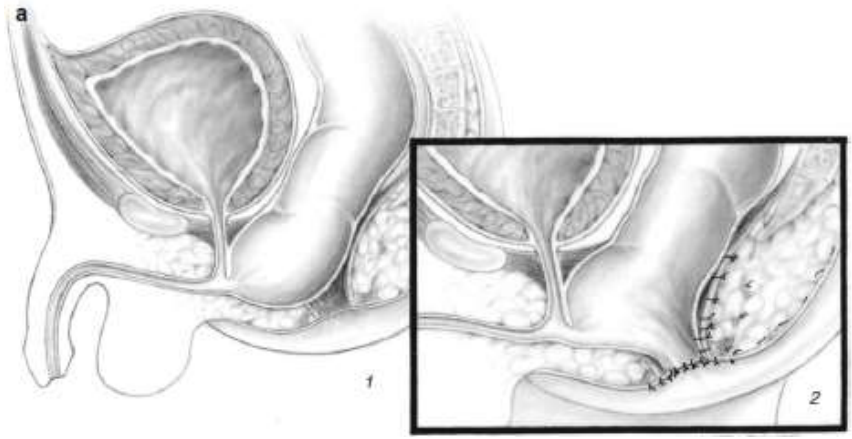
- ✓ Recurrent fistula
- ✓ Persistent fistula
- ✓ Acquired fistula
- ✓ Remnant fistula

Recurrent fistula

- Fistula repaired but recurred
- Occurs if **damaged rectal wall in front of sutured urethra (**not mobilized adequately)
- redo PSARP more difficult (enough rectum mobilized to be sure normal rectum in front of urethral suture)

Persistent fistula

- missed fistula
- Usually bulbar fistula with rectum bulging down mistaken for low ARM



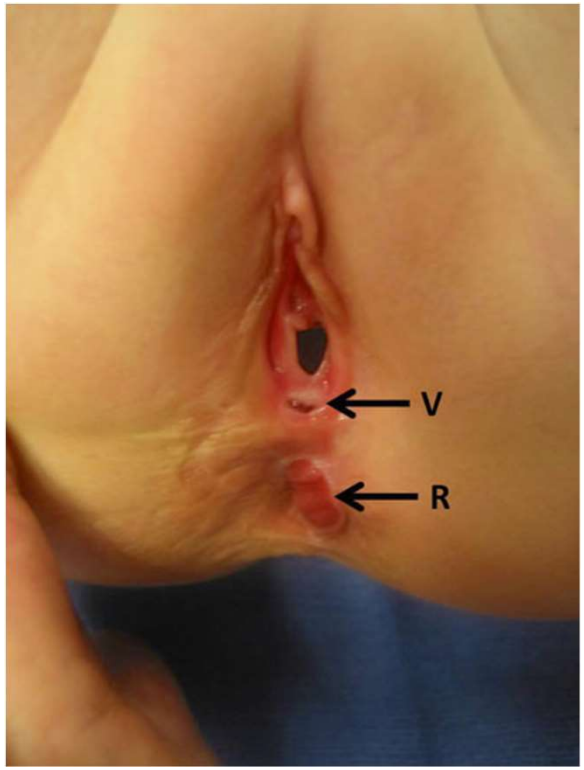
Acquired fistula

- New fistula in a patient with RPF
- undergoing Surgery without Foley catheter in urethra
- (inadvertent injury)- urethral transection
- Leakage of urine through anopasty
- Attempt to pass cystoscope and leave urethral catheter
- If fail, a suprapubic bladder catheter should be inserted and redo PSARP with urethral dilation or urethroplasty

Acquired fistula in females: **Recto-vaginal fistula** may occur in after failed repair of vestibular fistula . **Urethro-vaginal fistula** may occur In cloaca that required vaginal mobilization and separation, Prevent by 90°rotation of vagina and coverage of posterior urethra with ischiorectal fat pad

Remnant fistula

- Retained rectum left attached to posterior urethra
  - *** common in bulbar fistula approached by laparoscopy*
- Presents as urethral diverticula
- Pseudoincontinence, stones, UTI, orchiepididymitis, risk of malignant change, difficult to catheterize
- Can be fixed with PSARP (similar to primary case)

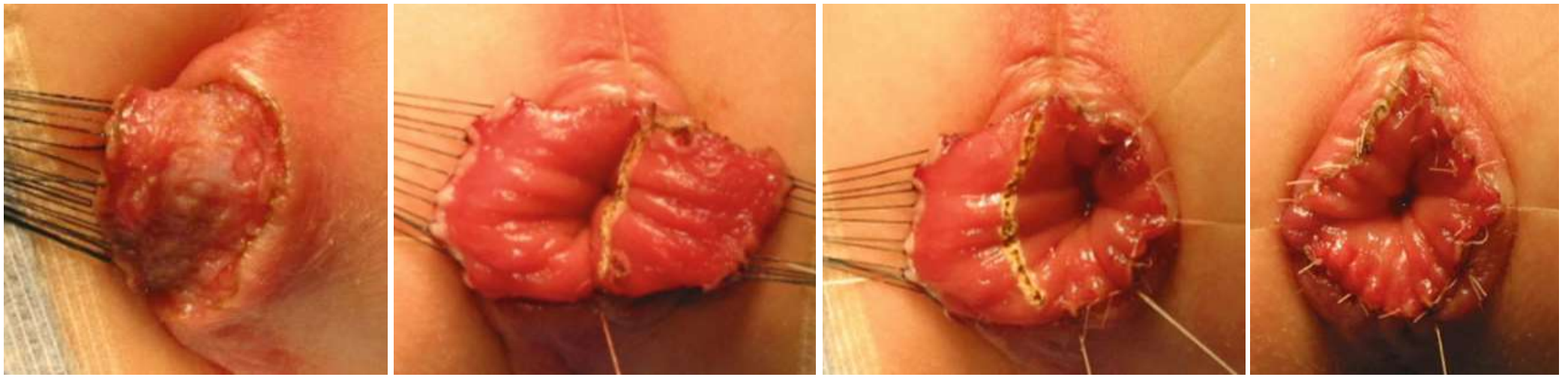


Remnant fistula In females pocket of original vestibular fistula may be left in a patient operated with the **erroneous diagnosis of vaginal fistula** where original distal rectum left behind and more proximal segment mobilized for anoplasty

5 RECTAL PROLAPSE

Prolapse

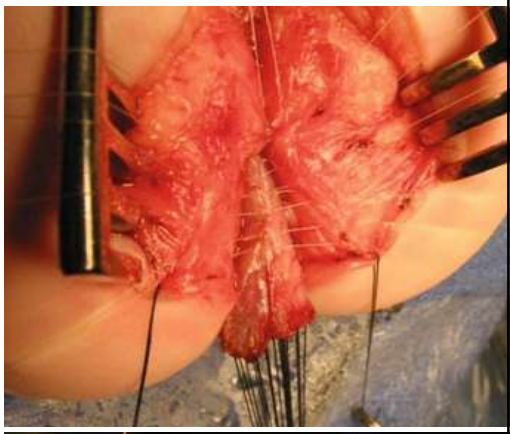
- Incidence 3%,
  - **Lack of fixation of rectum to sphincter**
  - **Absent/poor sphincter** mechanism. (high ARM)
  - **Exacerbated with straining** in case of constipation
- If significant can lead to ulceration, bleeding, mucus production and interfere with sensation.
- Repair requires bowel prep if done after colostomy closed

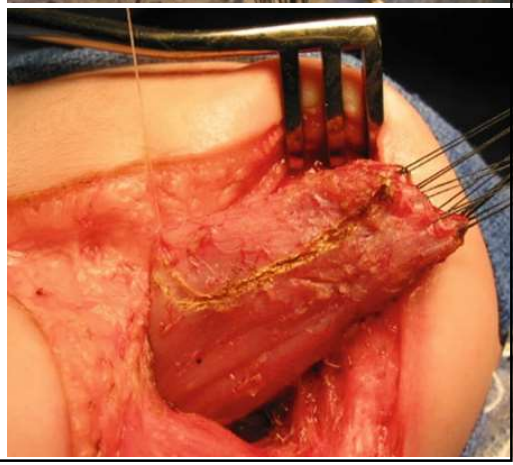


Reoperation for prolapse mucocutaneous junction incised circumferentially, mobilization of full thickness redundant rectum. Adequate length resected. Anoplasty done. There is risk of recurrence but never as severe as the original

Preventing prolapse

1. fixation of rectum to sphincter
2. anoplasty under slight tension so that after the sutures of the anoplasty are cut, the rectum retracts slightly with no mucosa being visible.
3. tapering a dilated rectum if necessary





6 Other

- ✓ Persistent Urogenital sinus
- ✓ Femoral nerve palsy

Cloaca--Persistent Urogenital sinus

- Cloaca patients mistaken for vaginal fistula and underwent only PSARP (only rectal component was repaired)
- Reoperation is approached posterior sagtally (rectum must be dissected and reflected out of the way) and urogenital sinus repaired as in other cloaca

Femoral Nerve palsy

- Transient palsy can be observed particularly in adolescents
- **excessive pressure in the groin** during PSARP
  (can be avoided with adequate cushioning)


THANK YOU!