Case Report

H-type fistula ARM

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30 wks GA, Prenatal US scan

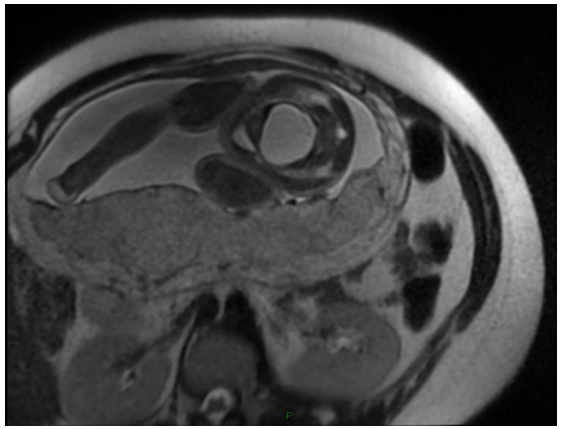
- anechogeic area posterior to the bladder
  54 x 40 x 45 mm
  extending down to the perineum with visible orifice
  suspected for hydrometrocolpos

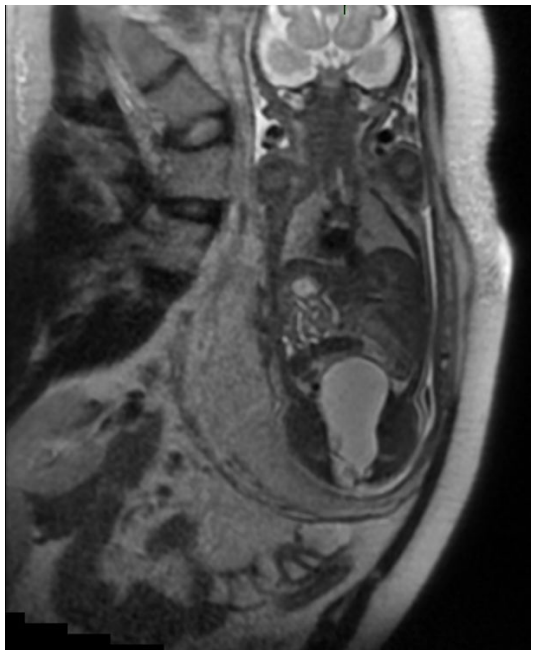
- normal kidneys

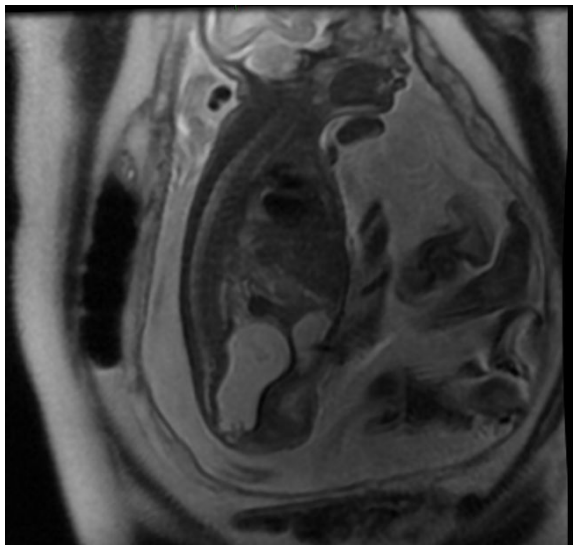
- single umbilical artery

30 wks GA, Fetal MRI

- perisacral mass
- 62 x 36 x 30 mm
- suspected for complex urogenital malformation with hydrometrocolpos





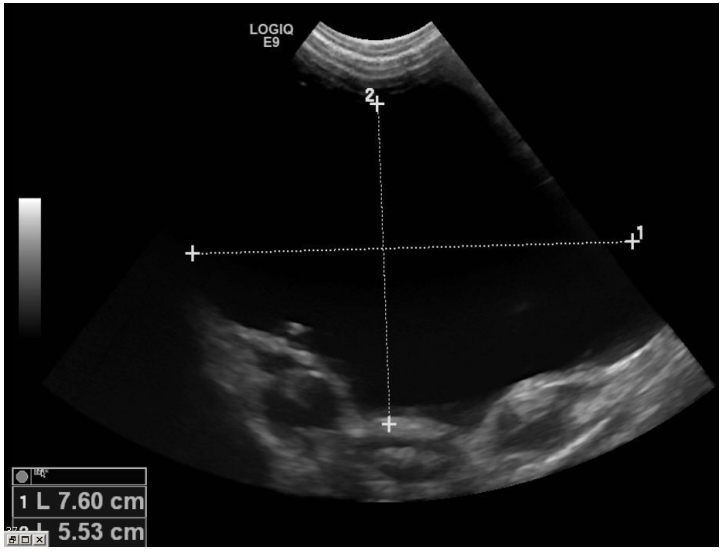


At birth (39 wks GA), O/E

- Normal APGAR score
- Normal perineum with three normally located orifices
- No anomalies detected other than
  - single umbilical artery
  - palpable abdominal mass
- NPO, NPT, Urinary catheter, normal looking patent rectum
- Meconium observed normally from D1 of life

D1-D3 of life, US scan

- Fluid filled abdomino-pelvic presacral mass (occupying inferior 2/3 of abdomen)
- 9 cm x 7.6 cm x 5.5 cm
  not comunicating with bladder or uterus
- 5-10 mm far from the perineal skin plane
- bowel loops compressed
- rectum not identifiable
- normal kidneys with no hydroureteronephrosis



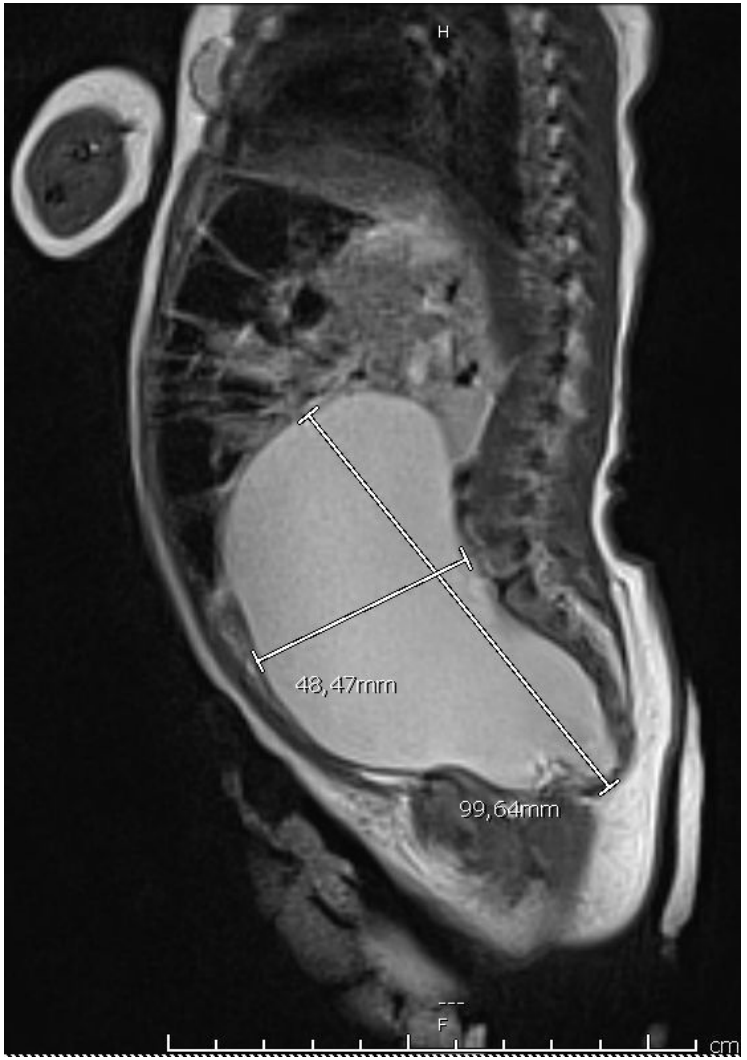
What do you think it is?

Differential Diagnosis:

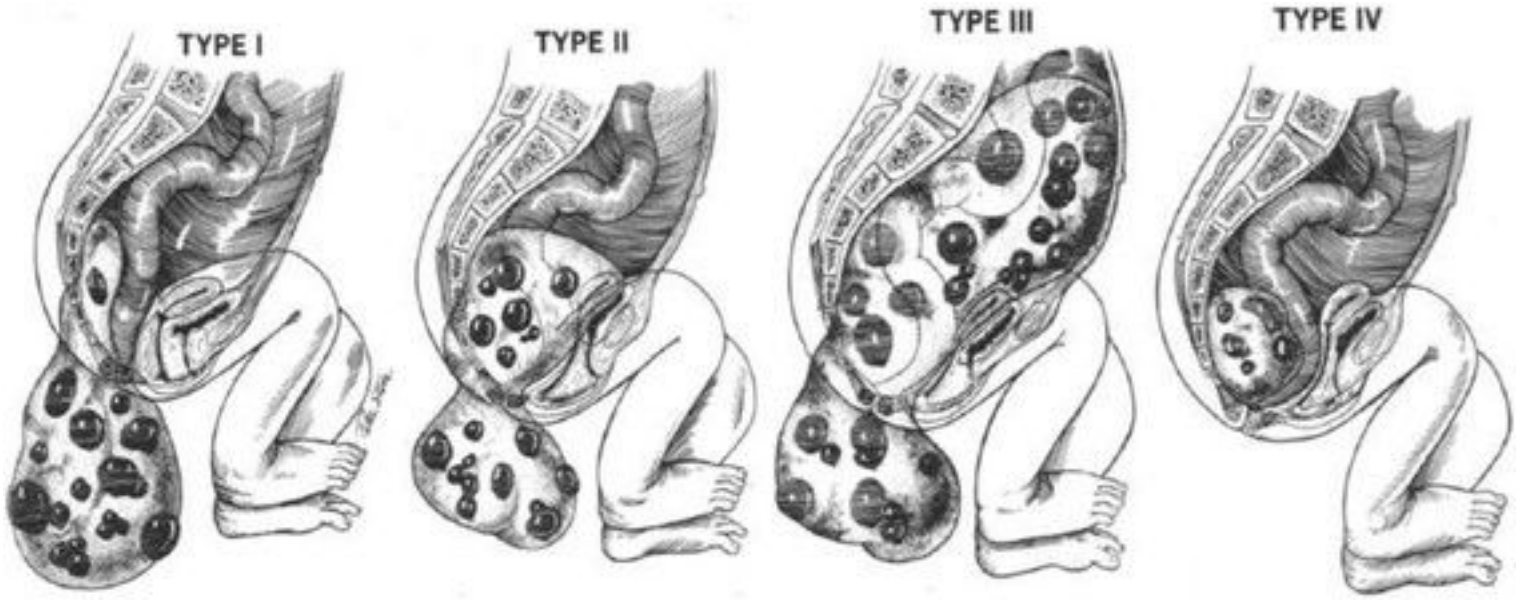
1. Hydrocolpos
2. Mesenteric cyst
3. Ovarian cyst
4. Cystic teratoma

D5 of life, MRI

- Fluid filled abdomino-pelvic mass
- 99 x 48 x 80 mm
- with multiple septa in the pelvic portion
- not clearly separable from the sacrococcigeal plane
- Suspected cystic sacrococcigeal teratoma



Altman classification



D7 of life, AXR

- Paucity of gas in the inferior abdomen
- Bowel loops not particularly dilated

D25 of life, Surgery

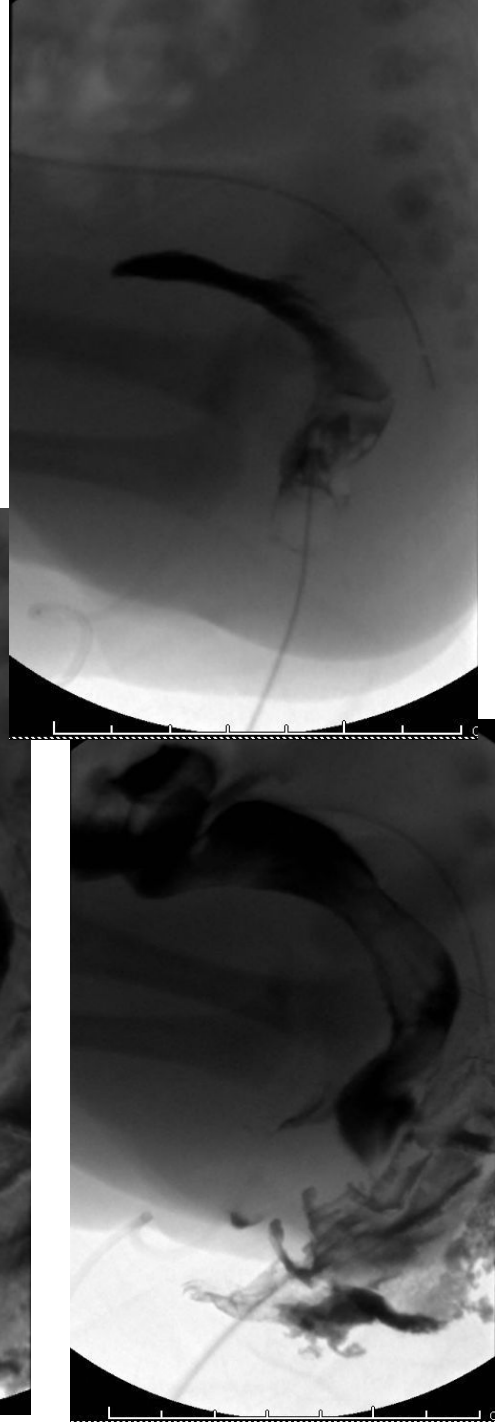
- Xifo-pubic laparotomy and mass excision
  - cystic retroperitoneal presacral mass extending down between pre-sacral plane and rectum
  - accidental opening of posterium rectal wall (repaired in double layer interrupted sutures)

D3 post-op

- Passage of stool from both vagina and rectum

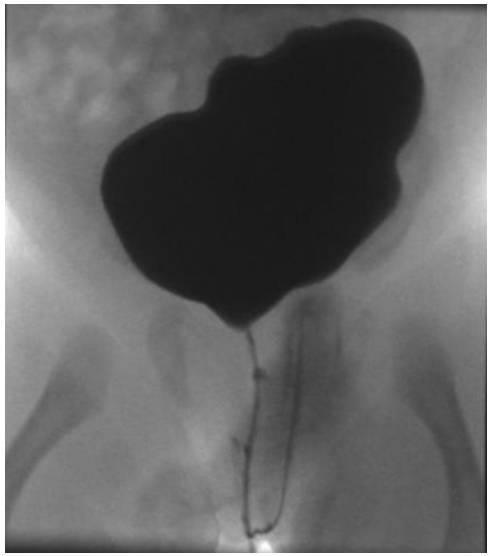
D6 post-op, Barium enema

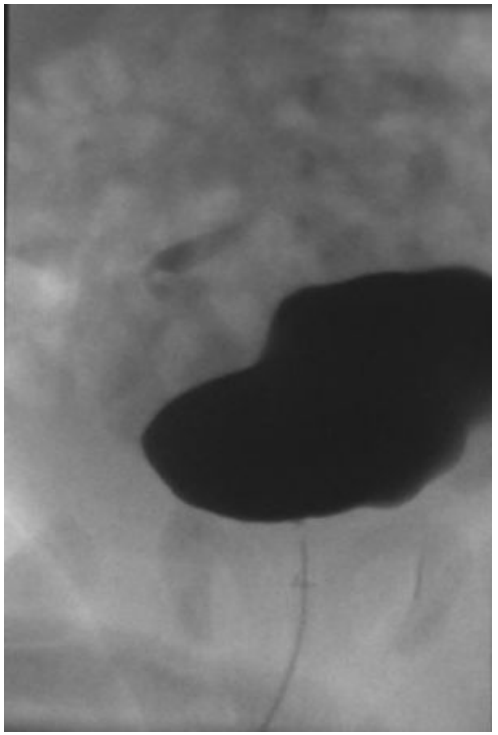
- Recto-vaginal fistula
- Iatrogenic??
  - but anterior rectal wall was untouched at surgery
- Congenital??
  - initially asymptomatic because of rectal compression
  - now only visible with deflated balloon



D13 post-op, MCUG

- Normal
- Mild R VUR



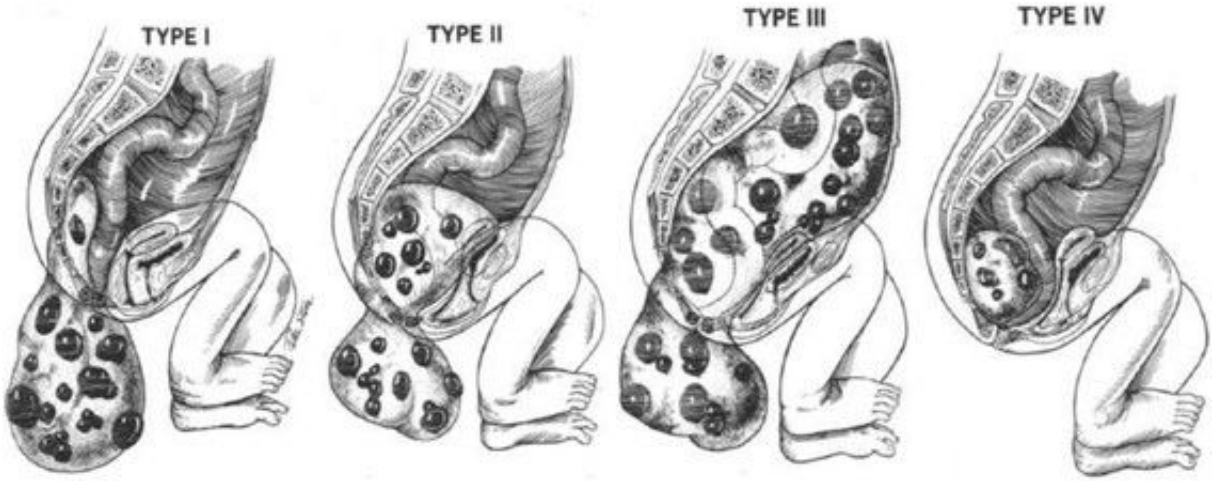


D25 post-op (D50 life), Surgery

- Sigmoid colostomy (distal sigmoid closed and left in pelvis)

- Post-op normal course
  - D3 stoma working
  - D4 feeds started

oncology



- Hystopathology:
  - Mature cystic teratoma, (Gonzales-Crussi grade 0)
- Elevated pre-op Alpha fetoprotein values falling post-op
- Favourable histology but requiring coccyx removal
  - Scheduled at the time of recto-vaginal fistula repair

H -TYPE FISTULA

Table 2 Standards for diagnosis international classification (Krickenbeck)

Major clinical groups	Rare/regional variants
Perineal (cutaneous) fistula	Pouch colon
Rectourethral fistula	Rectal atresia/stenosis
Prostatic	Rectovaginal fistula
Bulbar	H fistula
Rectovesical fistula	Others
Vestibular fistula	
Cloaca	
No fistula	
Anal stenosis	

- A rare anorectal malformation

Pediatr Surg Int (2017) 33:3–13
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REVIEW ARTICLE

Diversities of H-type anorectal malformation: a systematic review on a rare variant of the Krickenbeck classification

Shilpa Sharma[1] · Devendra K. Gupta[1]

- More common in female (2.5-6 times)
- More common in Asia (up to 16%)
- Anus can be stenosed and/or ectopic (perineal fistula)

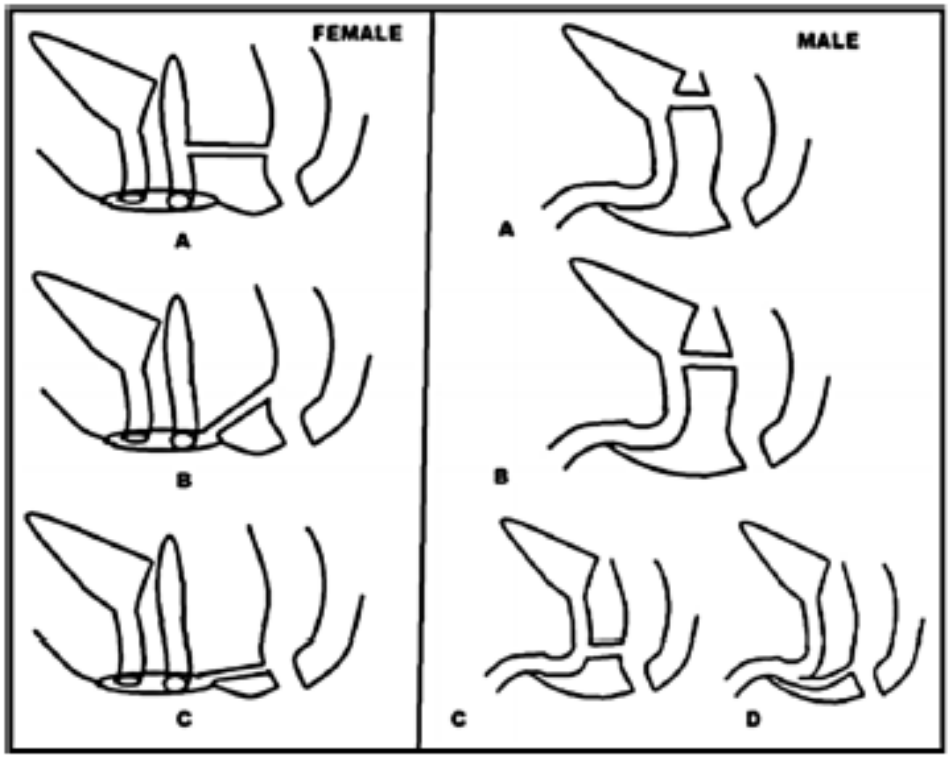

Fig. 1 H-type anorectal malformation may be described as *A* recto-vaginal fistula, *B* rectovestibular and *C* anovestibular in females, and *A* rectovesical fistula, *B* rectobladder neck, *C* rectoprostatic urethral, and *D* rectobulbar urethral in Males

H-type Fistula

- Presentation
  - passage of stool from the vestibule (+/-abscess) in girls
  - passage of urine from the rectum in boys (urethral ipoplasia stenosis)
- Confirmation
  - Contrast study
  - Vaginoscopy – urethroscopy
- Associated anomalies may be present in up to 20–60%
  - VACTERL
  - presacral mass
- Often associated with anorectal stenosis (37%)

Journal of Pediatric Surgery, Vol 31, No 4 (April), 1996: pp 559-562

H-Type Anorectal Malformations: Incidence and Clinical Characteristics

By R.J. Rintala, L. Mildh, and H. Lindahl

Helsinki, Finland and Liverpool, England

Table 1. Associated Anomalies

	Males	Females
Vertebral/sacral anomalies	6	3
Renal anomalies	4	2
Vesicoureteric reflux	1	3
Hypospadias/urethral anomalies	3	0
Cardiac anomalies	3	1
Esophageal atresia	2	0
Malrotation	3	0
High rectal stenosis	1	1
Limb anomalies	3	0
Cleft palate	1	0
Meningocele	1	0
Presacral teratoma	0	1
Total no. of anomalies	28	11
Total no. of patients	6	6

Which surgical procedure ?

1. Perineal repair
2. Limited PSARP
3. ASARP
4. Vestibuloanal pull-through
5. Endorectal pull-through
6. Anterior wall of rectum pull-through
7. Transanal excision of the fistulous tract
8. Endorectal mucosal advancement flap

(overall recurrence rate of 5–60%)

Management of H-type rectovestibular and rectovaginal fistulas

Taiwo A. Lawal, Kaveer Chatoorgoon, Andrea Bischoff, Alberto Peña, Marc A. Levitt*

Colorectal Center for Children, Cincinnati Children's Hospital Medical Center, Cincinnati, OH 45229, USA

Journal of Pediatric Surgery (2011) **46**, 1226–1230

Fig. 2 The posterior sagittal approach was used in the first 5 cases. Silk sutures were placed at the rectal end of the fistula to provide traction.

The essential technical point for repair is to get the healthy anterior rectal wall to cover the area of fistula on the posterior vagina

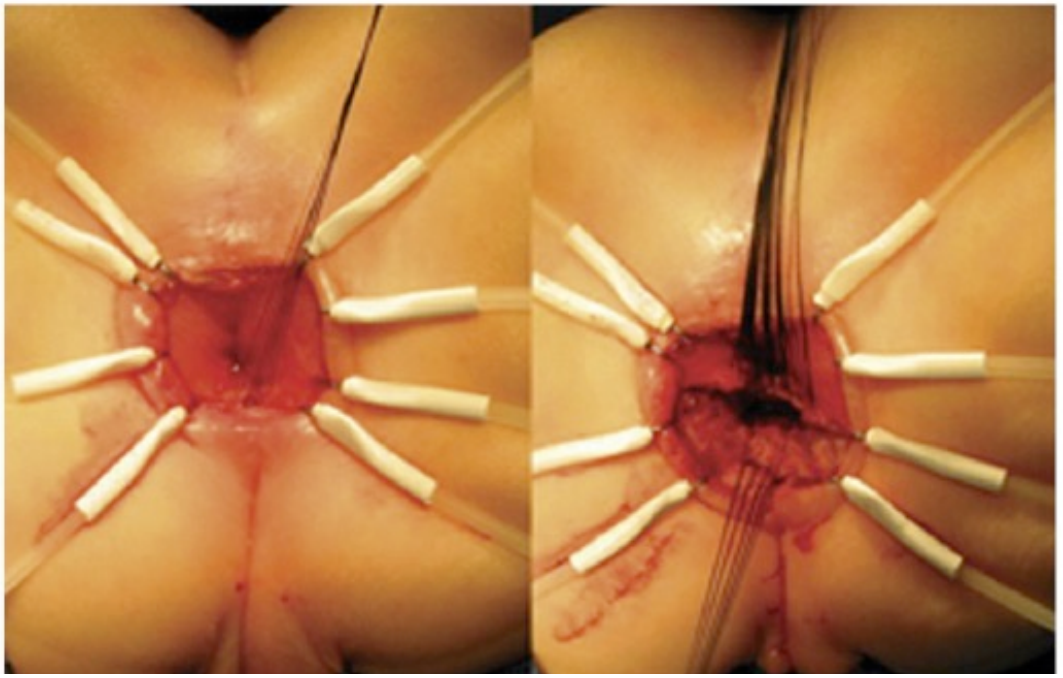

Fig. 3 The transanal approach in prone position for the mobilization of the anterior rectal wall (180°), ligation of the fistula, and repair. The anterior wall of the rectum is mobilized and pulled distally to cover the area of the fistula with healthy rectal wall.

Thank you