



Newborn male with features of covered cloacal exstrophy with prolapsing bladder plate at penoscrotal hypospadiac opening

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A Swords

# NICU Ambiguous Genitalia consult

HPI:

- Full term infant born via SVD
- Pregnancy notable for IUGR, oligohydramnios, and multiple ultrasounds completed to determine fetal sex
- Determined to be female fetus

Exam:

- imperforate anus
- ambiguous genitalia
- meconium and urine draining from mucosal surface within bifid scrotum/partially fused labia

# Physical Exam

## Non-GU

- Imperforate anus
- Low-lying umbilicus
- Rectus belly diastasis
- Pubic symphysis diastasis



# Physical Exam

## GU

- Bilaterally descended testicles
- Hypospadiac phallus
- Prolapse of mucosal surface at penoscrotal junction with visible orifices
- Production of stool and urine via mucosal opening
- Appendage-like cylindrical structures bilaterally



# Physical Exam

GU

- Mucosal prolapse reduced



# Diagnostic findings

## Labs

- Normal renal function
- Karyotype 46,XY

# Diagnostic findings

## Imaging

- Pelvic US: no female organs
- Pelvic XR: with wide pubic symphysis diastasis, sacral anomaly
- Abdominal US: mild R hydronephrosis
- Pelvic MRI: lack of sacrococcygeal elements

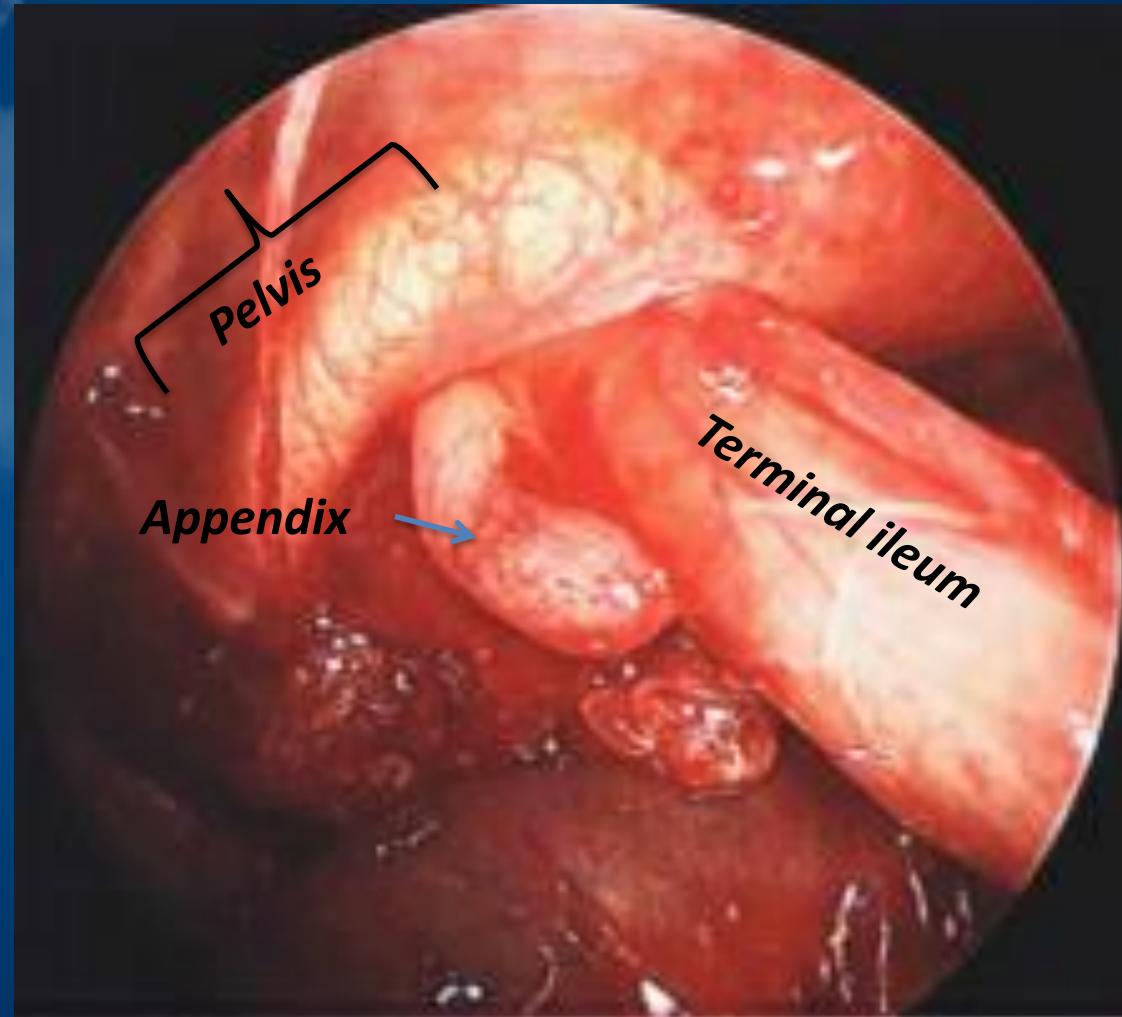
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# EUA, Laparoscopy

## Findings

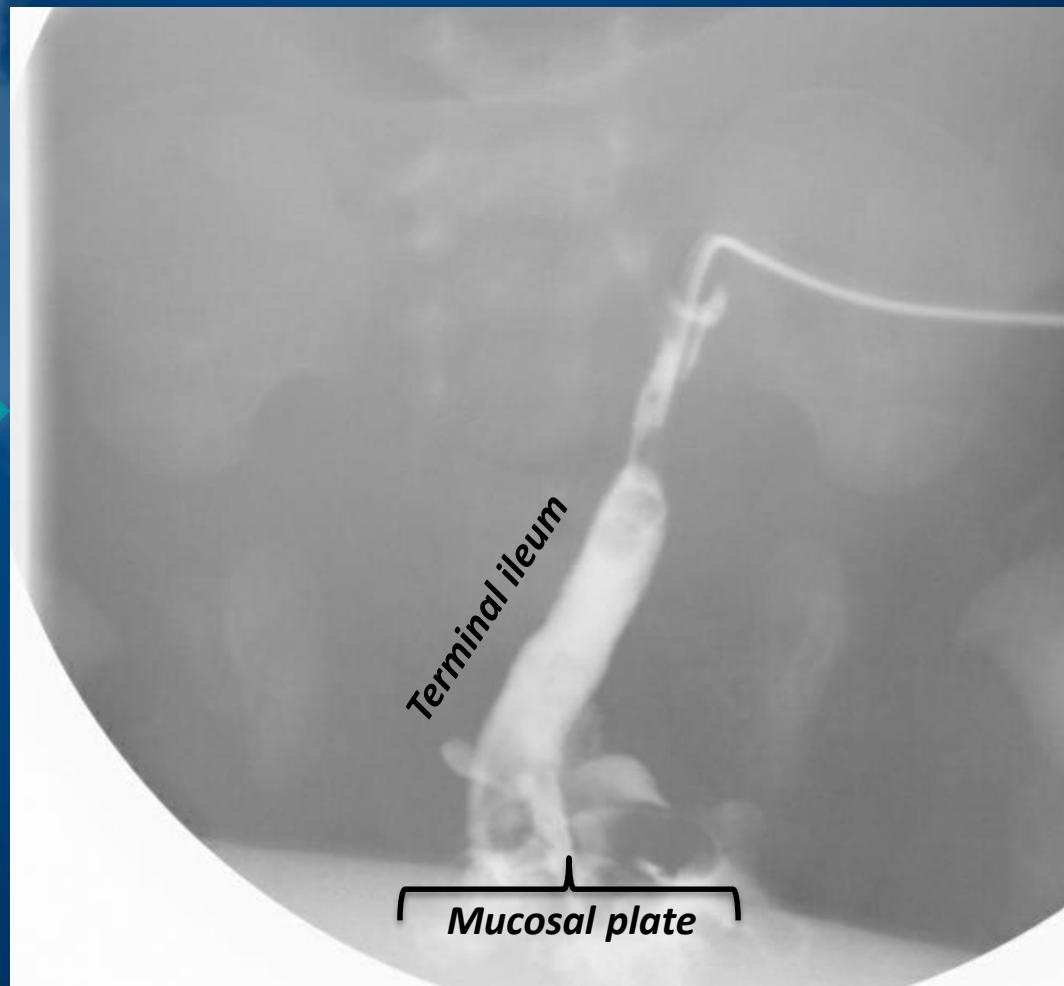
- Cystoscopy unable to distend mucosal space
- orifices identified on exam
- Only cecum
- colonic atresia
- Fecal diversion via end ileostomy with mucous fistula



# Fistulogram

## Findings

- No cecum visible



# First stage closure

## Findings

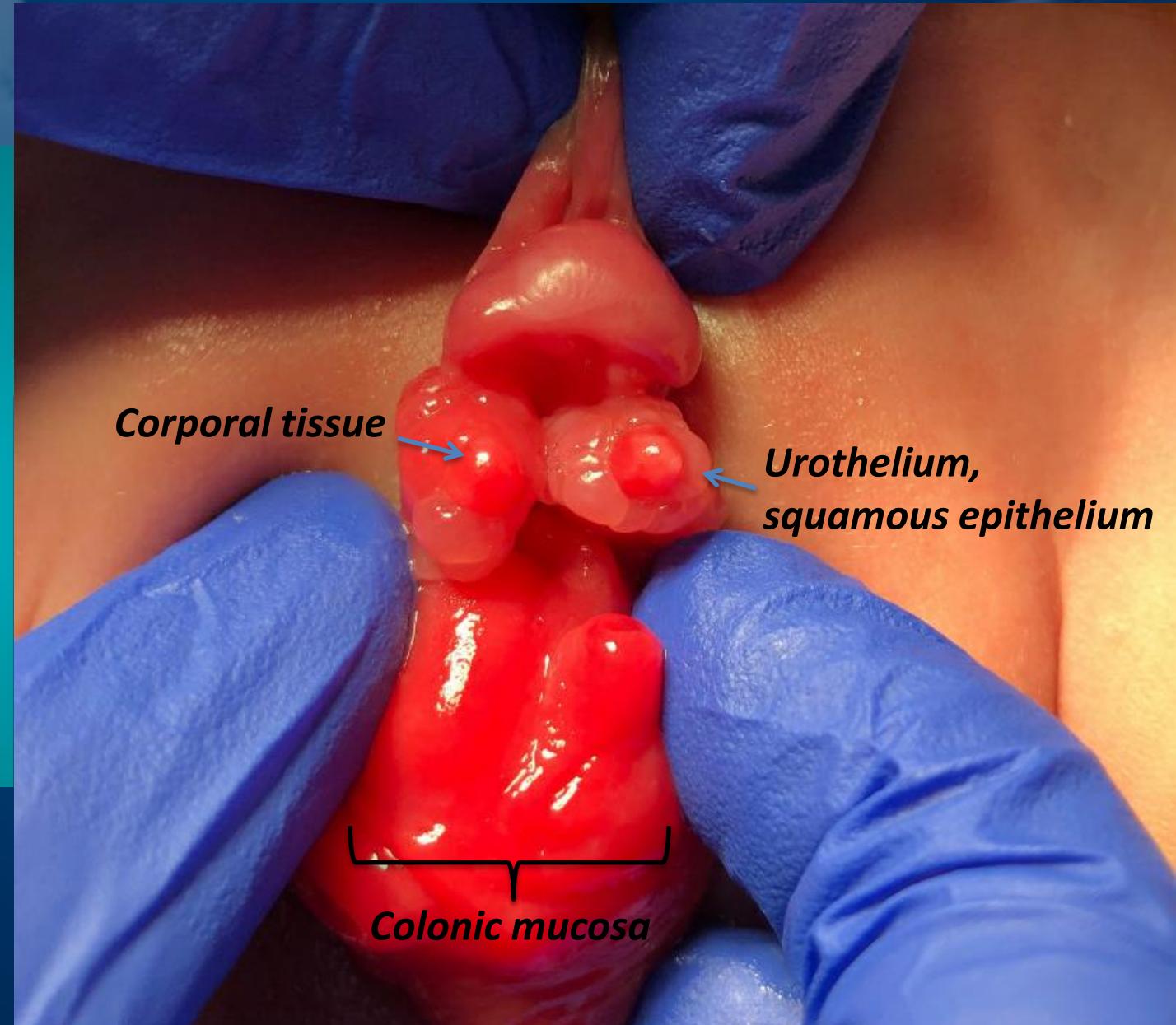
- Orifices identified



# First stage closure

## Diagnostic biopsies

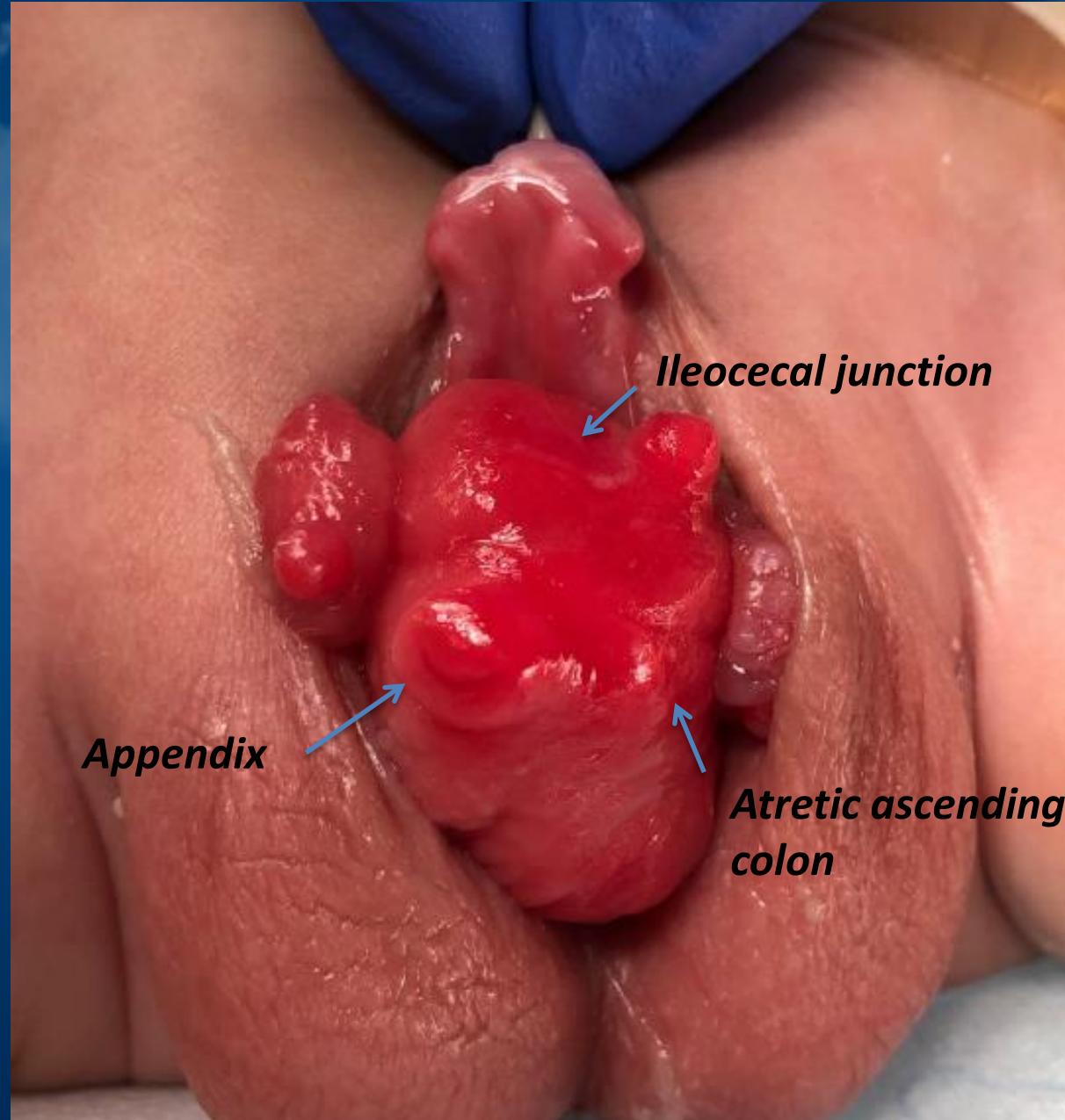
- Mucosal plate
- Bilateral cylindrical structures



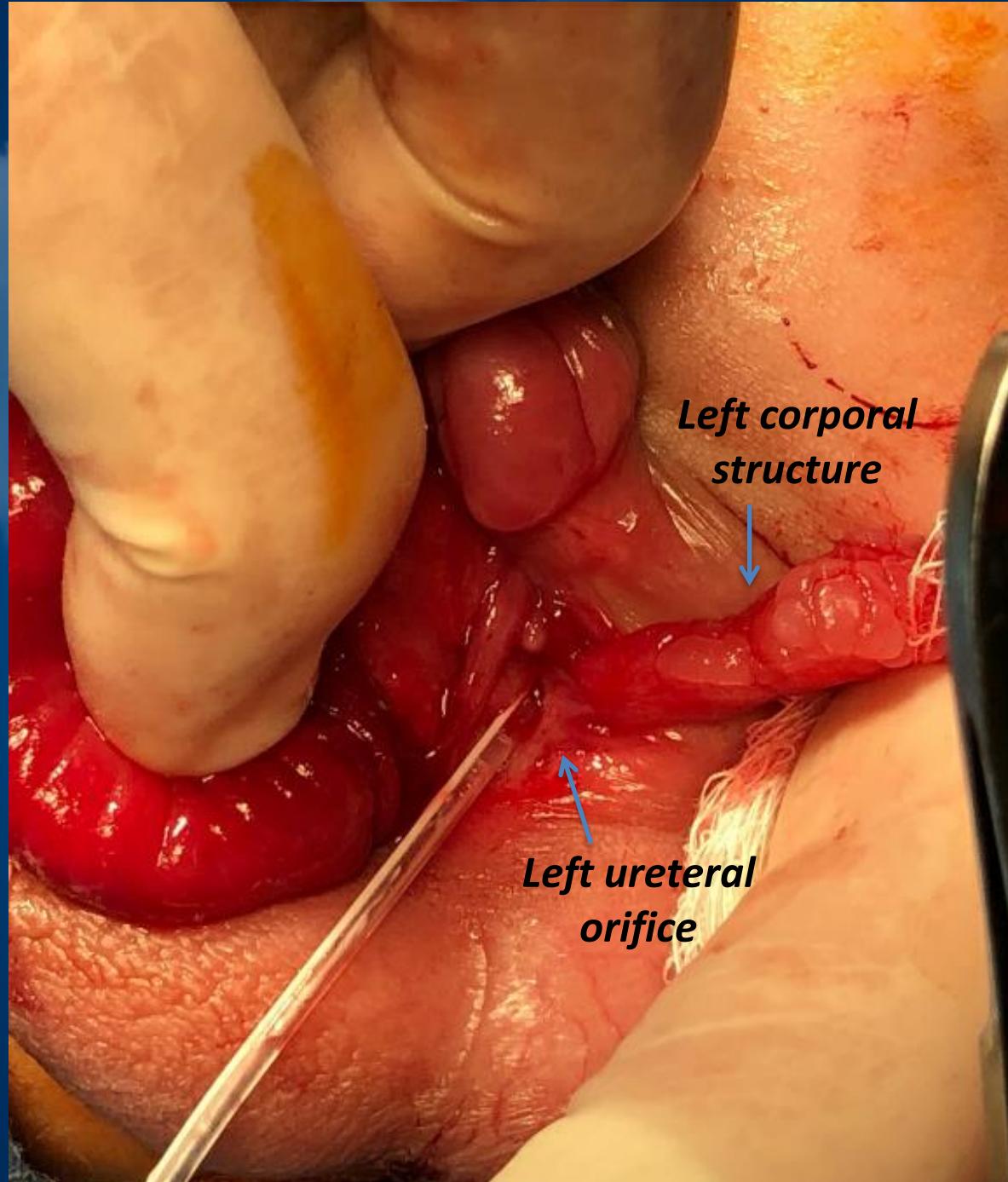
# First stage closure

## Findings

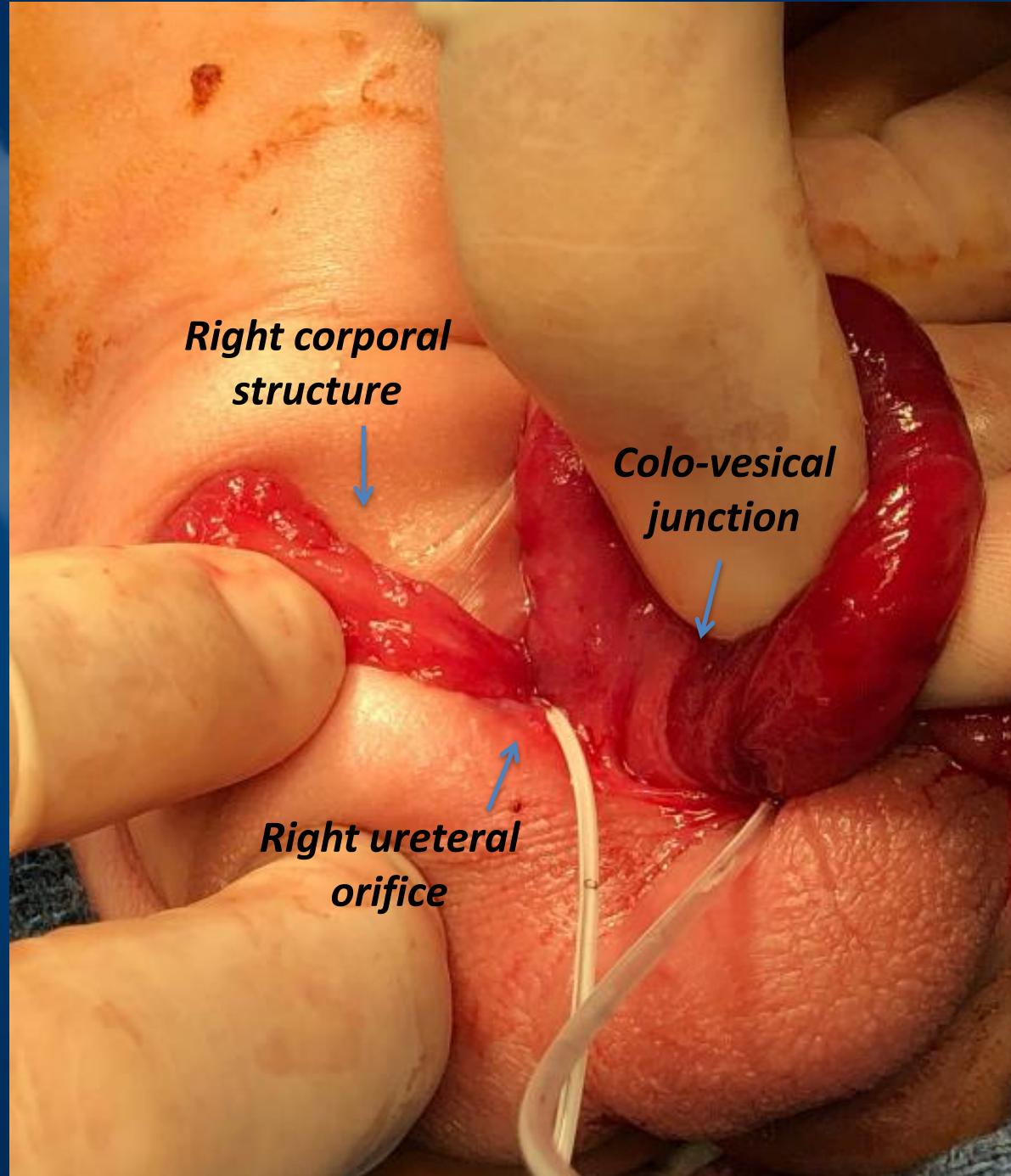
- Orifices identified



# Anatomic findings

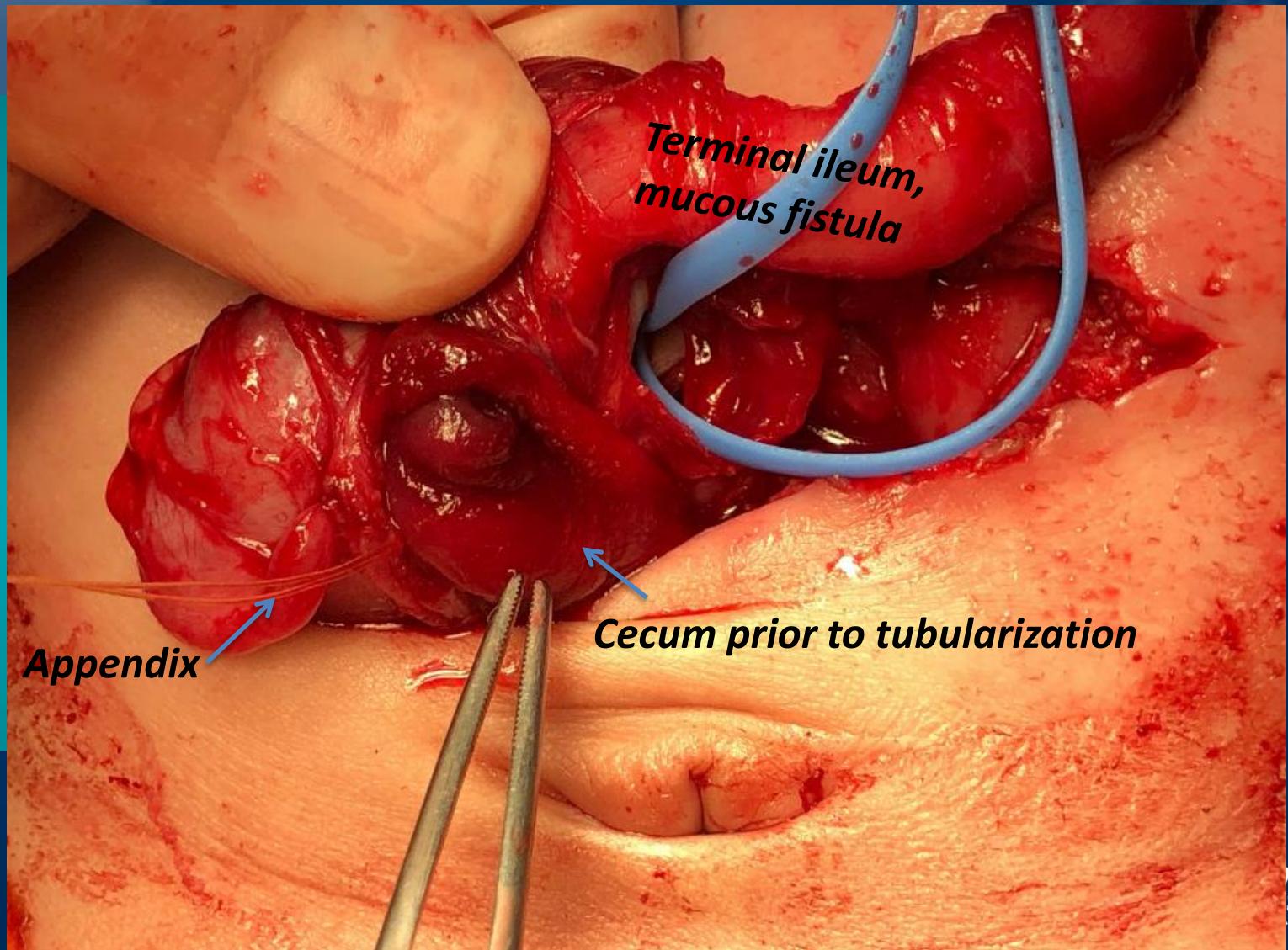


# Anatomic findings



# First stage closure

Findings



# First stage closure

Post-op

