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

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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
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

Corporal tissue

Urothelium, squamous epithelium

Colonic mucosa
Findings

- Orifices identified

First stage closure

Ileocecal junction

Appendix

Atretic ascending colon
Anatomic findings

- Left corporal structure
- Left ureteral orifice
Anatomic findings

- Right corporal structure
- Colo-vesical junction
- Right ureteral orifice
First stage closure

Findings

- Terminal ileum, mucous fistula
- Cecum prior to tubularization
- Appendix
First stage closure

Post-op