

Lost in Transition – Long-Term Sequelae in Cloacal Exstrophy

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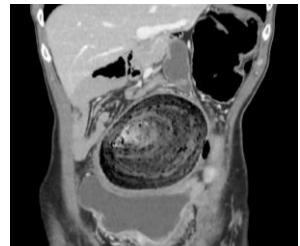
Background

Cloacal exstrophy represents one of the most complex congenital malformations of the lower urinary, genital, and gastrointestinal tracts. Advances in neonatal reconstruction have improved survival, yet many patients face lifelong functional and psychosocial challenges. Structured transition to adult care remains insufficiently established.

Case Summary

We report on a now 34-year-old woman with congenital cloacal exstrophy who has been under continuous care at our pediatric surgical center since early childhood. Following primary bladder closure and pull-through procedure, she underwent bladder augmentation with an ileal pouch and creation of a continent Mitrofanoff stoma, followed by vaginal reconstruction during adolescence. Subsequent procedures included vulvoplasty, labial reconstruction, and repeated anal sphincter dilations for recurrent stenosis.

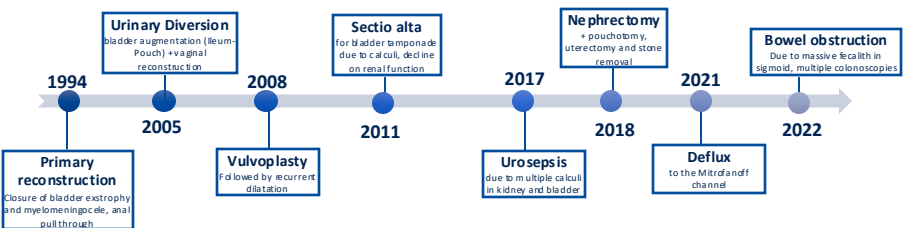
Long-Term Follow-Up



In 2011, the patient presented with hematuria and bladder tamponade due to multiple calculi. Renal imaging showed a decline of function on the left side (15%) as well as a grade II–III vesicoureteral reflux (VUR) on the left side. During open surgery (secto alta), multiple stones were removed from the pouch; however, due to adhesions and limited renal function, the left ureter and kidney were left in situ.

In December 2017, urosepsis occurred secondary to grade IV hydronephrosis and abscess-forming pyelonephritis of the left kidney, again due to multiple stones and neobladder. Consequently open radical nephrectomy with ureterectomy, pouchotomy, and stone removal was performed. Urodynamics showed stable pressures with VUR into the solitary kidney. In 2021, a Deflux injection was applied to the stomal channel, achieving stable continence and normal renal function.

From 2022, progressive bowel-related complications emerged. CT showed a large fecalith in the sigmoid colon with upstream fecal impaction to the left colonic flexure, requiring multiple colonoscopies for gradual removal. Examination also revealed severe anal stenosis. Surgical anal dilation was deferred due to adequate self-dilation management since childhood and an unclear sphincter status. Despite interventions, recurrent severe fecal impaction has frequently recurred since.



Conclusion

A structured, interdisciplinary transition pathway from pediatric to adult medicine is essential to ensure continuity of care for individuals with rare congenital malformations such as cloacal exstrophy. Close collaboration among pediatric and adult specialists — including surgeons, urologists, gynecologists, and psychologists — is vital to maintaining function, preventing complications, and supporting long-term well-being.

Takeaway Message

Long-term success in cloacal exstrophy extends beyond primary reconstruction. It relies on coordinated, lifelong multidisciplinary management and a structured transition into adult healthcare systems to safeguard both medical outcomes and quality of life.