Pediatric Case Reports

Repair of Iatrogenic Urethral and Bladder Neck Injury Due to Missed Diagnosis of Mayer-Rokitansky-Küster-Hauser Syndrome

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Mayer-Rokitansky-Küster-Hauser syndrome is a congenital malformation disorder resulting in agenesis of the proximal vagina, absence of cervix, and variable development of the uterus. This report describes the repair of a complex iatrogenic urethrovaginal injury due to a missed diagnosis of Mayer-Rokitansky-Küster-Hauser. Our treatment utilized a primary urethroplasty through a transvaginal approach with bladder neck reconstruction and a Martius flap for secondary coverage. Urinary continence was restored postoperatively. UROLOGY 134: 213–216, 2019. © 2019 Elsevier Inc.

CASE PRESENTATION
A 15-year-old female was referred to our center with a 4 month history of total urinary incontinence (UI) following a partial hymenectomy for primary amenorrhea from a presumed imperforate hymen. A magnetic resonance study of the pelvis performed posthymenectomy showed absence of a normal uterus and proximal vagina with normal ovaries and bladder (Fig. 1). The patient’s past medical, antenatal, developmental, and family history were unremarkable.

Examination under anesthesia demonstrated Tanner V external genitalia. Flexible cystoscopy identified a completely incised urethra at the 6-o’clock position, extending through the ventral portion of the external urethral sphincter into and past the BN (Fig. 2). The ureteric orifices were uninjured. The bladder could be filled to a normal capacity. Total UI was demonstrated on removal of the cystoscope.

Treatment options were reviewed with the patient and her family. The patient’s goal was to regain urinary continence. Surgical options discussed included: (1) BN reconstruction with a pubovaginal sling and Mitrofanoff appendicovesicostomy, (2) creation of vaginal channel using distal urethra, closure of BN, and Mitrofanoff appendicovesicostomy, and (3) reapproximation of the incised distal urethra and BN from a transvaginal approach. The last option was chosen as this represented the most likely intervention that would allow the patient to regain continence without catheterization. She declined a vaginoplasty as part of the reconstruction.

SURGICAL PROCEDURE
The patient underwent a transvaginal BN reconstruction and urethroplasty with Martius flap and suprapubic cystotomy. She was prepped and draped in dorsal lithotomy position. A 16-French Foley catheter was placed to demonstrate the margins of the incised urethra (Fig. 3a). The urethral edges and BN were dissected laterally, which allowed for tension-free reapproximation of the urethral edges. The lateral edges of the vaginal cuff were dissected posteriorly.

The BN and urethral edges were reapproximated in 2 layers. The primary suture line was imbricated into the lumen with a secondary layer starting at the BN and extending into the distal urethra with 4-0 and 5-0 absorbable suture, respectively (Fig. 3b). Rigid cystoscopy verified that ureteric orifices were uninjured. The bladder demonstrated adequate capacity without leakage. A 12-French suprapubic catheter was placed.

A Martius flap from the left labia majora was harvested to cover the repair. The Martius flap was transposed between the urethral repair and the vagina.

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margins were approximated with running 4-0 nonabsorbable sutures (Fig. 3c). A Penrose drain was placed in the left labia majora and the incision was closed using 5-0 absorbable sutures. A 12-French Foley catheter was inserted into the bladder and secured.

Vaginal packing was removed on postoperative day 3. The drain and urethral catheters were removed postoperative day 7. She was discharged from hospital with the suprapubic catheter on postoperative day seven and kept on prophylactic cephalaxin and oxybutynin three times daily until outpatient follow-up.

FUNCTIONAL OUTCOMES
There were no postoperative complications. Pain was well controlled on acetaminophen. A bladder capacity of 200 mL with no evidence of leak was demonstrated on cystogram. With the suprapublic catheter plugged, the patient was able to hold contrast without UI and emptied her bladder with a good urinary stream. The suprapubic catheter was then removed at this appointment. She was instructed to continue pelvic floor exercises and to void every 2-3 hours to cycle her bladder.

At 6-month follow-up the patient was content with the results of surgery. She had no urinary tract infections and lower urinary tract symptoms. She had experienced minimal stress UI with strenuous exercise.

DISCUSSION
Female urethral and BN injuries are rare, most commonly caused by iatrogenic complications of pelvic surgery. They

Figure 1. MRI Pelvis. (a) Axial T2WI MR shows a cord-like fibrous structure (→) at the expected location of the lower vagina, (b) Sagittal T2WI MR shows a small triangular structure (black arrow). No discernible cervix is seen. The vagina is replaced by a fine cord-like structure (white arrow).

Figure 2. Cystoscopic view of transected urethra and bladder neck on ventral aspect. No fistula seen at the time of cystoscopy. Normal ureteric orifices were identified at the time of cystoscopy. (Color version available online.)
can also be caused by obstetrical trauma, sexual trauma with foreign objects, radiation, pelvic cancers, and long-term erosion secondary to indwelling catheters. These injuries are even more uncommon in pediatric patients. In this case report, we describe the first reported iatrogenic urethral and BN injury during a presumed hymenectomy that occurred as a result of a missed diagnosis of MRKH.

Female urethral injuries present with varying symptoms depending on the extent and location of injury. Smaller injuries distal to the urethral sphincter may present with postvoid vaginal urine leakage. More extensive injuries involving the urethral sphincter present with varying degrees of incontinence. Other symptoms of severe urethral injury include intermittent positional incontinence, irritated perineal skin, and repeated vaginal fungal infections. Due to the functional and anatomic variability between types of urethral injury, current guidelines do not recommend a standardized treatment strategy for management of urethral injury.

Conservative management, including indwelling urinary catheterization and anticholinergic medication, are used for small injuries that present early. If unsuccessful, surgery is often necessary to restore continence. Most surgical repairs for urethral injury are performed via a transvaginal approach. Primary repair is performed for smaller fistulas. Graft tissue is used when primary repair is not possible due to anatomic disturbance, fibrosis of the surrounding tissue, or insufficient fascia for a second layer closure.

In this case, a Martius flap was used as it can be used to minimize the risk of fistulization for repairs distal and proximal to the external sphincter. Simple vaginal flaps are used for small fistulas distal to the sphincter, however, our patient possessed relative contraindications to this method due to her young age and anatomic features secondary to MRKH (small and under developed vaginal wall). Gracilis muscle flaps can also be used, but require additional surgical incisions and incur the risk of partial flap necrosis due to poor vascular supply, although there are benefits for larger, more complex fistulas. Buccal mucosa grafts have been used in urethral injury repairs, and do not require dissection of the urethra, minimizing risk of bleeding and damage to nervous structures. However, this approach also required additional surgical sites.

CONCLUSION

MRKH is a rare congenital malformation characterized by agenesis of the distal vagina and cervix with partial uterine development. This case report is the first to describe iatrogenic urethral injury incurred following a hymenectomy completed on a patient with a missed diagnosis of MRKH with successful subsequent transvaginal BN reconstruction and urethroplasty with Martius flap. This repair restored total urinary continence and should be considered in similar cases.

References


