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Commentary on management of müllerian tract anomalies associated with cloacal malformation and cloacal exstrophy

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ABSTRACT

We present a case of urethral prolapse in prepuberal female that presented with vulvar bleeding. A full medical history from both the patient and her parents in addition to a physical examination by the pediatric urology team were done and revealed incarcerated urethral prolapse. Treatments range from medical management to immediate surgical repair; in our case surgical repair was successfully performed. This case report discusses the uncommon manifestations of urethral prolapse in young girls with no previously known risk factors for urethral prolapse.

Key Words: Urethral prolapse, Incarcerated prolapse

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Description

Cloacal malformations and cloacal exstrophy are overall rare congenital malformations but involve the gastrointestinal and genitourinary tract and as such, require input from multiple specialties including pediatric surgery, pediatric urology, and pediatric gynecology. Cloacal malformations are found exclusively in female patients and are the most complex variant of anorectal malformations. In these patients, there is a single perineal orifice with a common channel of variable length and a confluence of the rectum, urinary tract, and reproductive tract. Cloacal exstrophy consists of a midline defect with two exposed and separated hemi-bladders attached to an exstrophied cecal plate, and often adjacent intussuscepted and eviscerated distal small bowel, as well as an omphalocele.

Most patients with a cloacal malformation and 46 XX patients with cloacal exstrophy have associated Müllerian tract anomalies. The incidence of Müllerian tract anomalies in patients with cloacal malformations is estimated at 53%-67%, most commonly duplication of the uterus and/or vagina. Although series are small given the overall rarity, 46 XX patients with cloacal exstrophy also have a high incidence of associated Müllerian tract anomalies with 43% having vaginal and uterine duplication in one of the largest series [1-3]. Historically, many patients with cloacal anomalies were managed by pediatric surgeons and underwent vaginoplasty or neo-vaginal construction at the time of their Posterior Sagittal Anoplasty (PSARP) [4,5]. However, even at the time of surgery, Müll erian anatomy is not always able to be adequately defined. It is the authors opinion, and one that is being discussed broadly within the colorectal and pelvic reconstruction community, that Müllerian reconstruction should be deferred with plan for close longitudinal follow-up,

ultimately with collaborative reconstruction alongside adolescent gynecology and potentially pediatric urology if needed, as patients get older and can participate in the decision-making [3].

As such, this puts these patients may present with Müllerian Outflow Tract Obstruction (OTO). Even patients who had a vaginoplasty in early childhood are still at risk for OTO due to the high rate of stricture. Patients with OTO tend to present in adolescence with cyclical abdominal pain and primary amenorrhea [6]. Given the high incidence of Müllerian tract anomalies in patients with cloacal malformations and the more rare patients with 46 XX with cloacal exstrophy, providers caring for these patients should anticipate OTO prior to presentation of symptoms as patients approach puberty [7].

As such, our institutional approach is to begin discussions surrounding OTO with patients and their caregivers early [8]. Müllerian anatomy should be part of the initial discussion with caregivers prior to any initial reconstruction even if there is no plan for reconstruction of gynecologic structures at that time. This should be reinforced at subsequent clinic visits with discussion of what to expect over time, especially as the child approaches puberty. At the time of thelarche, particularly if reconstruction has not yet taken place or if there is a known stricture, anatomy should be reviewed with consideration for repeat imaging to evaluate for the possibility of OTO. Pelvic Ultrasound (US) should be the initial imaging study and for patients at risk of OTO, repeat US should be obtained every 6 months to evaluate for occult OTO [8]. If ultrasound findings are unclear, pelvic MRI can provide further anatomical details. If OTO is suspected, the first step is menstrual suppression with hormone therapy [8].

Following menstrual suppression, there should be discussion with patients and their caregivers regarding next steps. Shared decision making is critical in this patient population and considerations should be made for pain management, desire and/or ability for future fertility, and patient readiness. For this reason, our institutional practice is to transition care of these patients to our Differences of Sexual Differentiation (DSD) clinic as they approach puberty [8]. Although cloaca and cloacal exstrophy do not have DSD, the clinic organizes access to all relevant providers from pediatric surgery, pediatric urology, endocrinology, psychology, and adolescent gynecology all of whom can lend expertise to the care of these patients. Options for surgical reconstruction are varied but the goal should be to relieve the outflow tract obstruction, and allow for future fertility and sexual activity, if that is in line with the patient's desires.

Conclusion

In conclusion, management of Müllerian tract anomalies in patientswithcloacalmalformationsand 46 XX patients with cloacal exstrophy is complex and requires input from multiple sub-specialties. Our recommendations are to delay gynecologic reconstruction at the time of initial intestinal reconstruction to allow for better delineation of Müllerian structures. This also allows for shared decision making with patients as their anatomy and resultant issues allow. As such, there should be a high index of suspicion for OTO as these children approach adolescence with close non-invasive monitoring with abdominal and pelvic ultrasounds after breast development begins. Also, the importance of on-going discussions about options and goals for reconstruction cannot be overstated.

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