



Gynecologic concerns in patients with cloacal anomaly



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ABSTRACT

Children with anorectal malformations (ARM) constitute a significant group within a pediatric surgery practice. It is important with female cases of anorectal malformations to consider the association of gynecologic anomalies, especially at the time of the definitive repair. However, it is critical to consider the association of such gynecologic anomalies when caring for patients with a cloacal anomaly. If not recognized, an opportunity to diagnose and treat such anomalies may be missed with the possibility of negative implications for future reproductive capacity. With the knowledge of the associated anomalies and long-term sequelae, surgeons can provide better care for girls and important counseling for parents. Knowledge of reproductive related issues in females with cloaca allows the pediatric surgeon an opportunity both, to provide optimal surgical management in infancy, childhood, and into young adulthood and to collaborate medically and surgically with an experienced gynecologist in patients with more complex anatomic variations. Appropriate counseling for patients and families about potential reproductive concerns that may develop many years after the definitive surgical repair allows preparation and planning to preserve future fertility.

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Gynecologic concerns in patients with anorectal malformations

Children with anorectal malformations (ARM) constitute a significant group within a pediatric surgery practice. Some anorectal malformations, such as an imperforate anus with a rectovestibular fistula, may seem straightforward and managed with a posterior sagittal anorectoplasty (PSARP); however, the association of gynecologic anomalies in all ARM has been firmly established.^{1,2} However, it is critical to consider the association of such gynecologic anomalies when caring for patients with a cloacal anomaly. If not recognized, an opportunity to diagnose and treat such anomalies may be missed at the definitive surgery with the possibility of negative implications for future sexual performance and reproductive capacity. With the knowledge of the associated anomalies and long term sequelae, surgeons can provide better care for girls and important counseling for parents and to collaborate with an experienced gynecologist in patients with more complex anatomic variations.

Evaluate for associated anomalies

Many pediatric surgeons anecdotally report that they customarily repair most ARM, other than a cloaca, without evaluating for

the presence of a gynecologic anomaly. However, most pediatric surgeons are aware of the strong association of gynecologic anomalies with a cloacal anomaly, including 53–67% of female patients with uterovaginal anomalies.^{3–5} It is essential to understand the type of reproductive anomaly and expected effects both on short and long term outcomes. Non-obstructive uterovaginal anomalies will not produce acute concerns at puberty, thus management may be deferred. Since possible menstrual obstruction at puberty may lead to catastrophic consequences in patients with complex cloaca, obstructive anomalies should be identified and addressed early.

In cloacal anomalies, it is necessary to separate both the urinary and colorectal systems from the reproductive tract; however, it is also essential in such cases to appreciate the uterovaginal anatomy to prevent adverse outcomes at puberty and menarche. Previous literature has reported a 36–41% rate of menstrual obstruction at puberty.^{6,7} If not diagnosed early, not only can this complication produce significant pain, often requiring urgent surgical intervention, but can also lead to infertility. In less complex cloacal anomalies (shorter length common channel and those without associated renal anomalies), a vaginal septum can be effectively treated during the initial repair of the rectum. This can be accomplished with one anesthetic exposure, with optimal surgical exposure, and without the possible psychological concerns associated with such a procedure in adolescence.

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Ways to evaluate reproductive anatomy

Vaginoscopy should be performed on all girls before or during the definitive repair of any complex ARM. This can be performed under coincident anesthesia at the time of the PSARP, with preparation to undertake vaginal septum resection if diagnosed. However, in more complex malformations such as cloaca, an exam under anesthesia with both cystoscopy and vaginoscopy can often be beneficial for surgical planning. If girls or young women were treated in infancy, without a complete gynecologic evaluation, endoscopy can be combined with any other indicated surgical procedures during childhood. The goal is to have as much information as possible regarding reproductive anatomy so that a definitive plan can be in place for implementation on or around the onset of menarche. Accurate knowledge of the reproductive anatomy allows both parents and providers to adequately prepare well ahead of anticipated pubertal changes and arrange necessary surveillance or planned interval procedures.

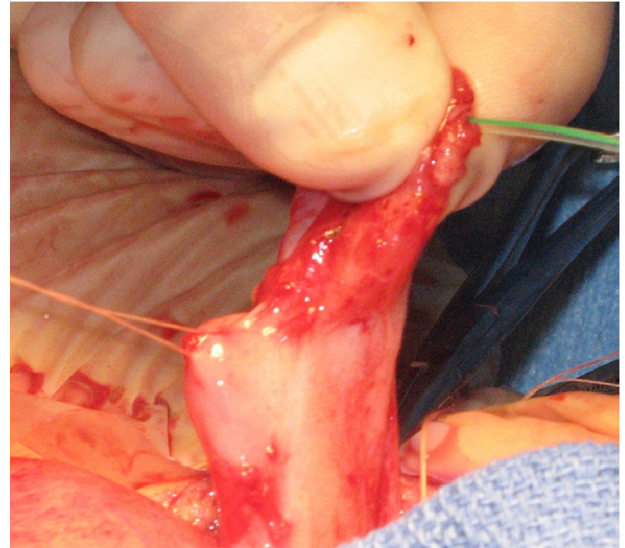
Endoscopy of the common channel allows evaluation of the vaginal anatomy; documentation of vaginal duplication with two hemivaginas and a septum, documentation of the septal length, and total vaginal length. The cervix or cervixes can also be visualized. This allows documentation of the cervical appearance, including cervical development, the position of the cervix/cervixes in the vagina(s), and the presence or absence of mucus at the ectocervix (Figure 1). This also provides assessment of adequacy of native vaginal tissue for reconstruction and possible need to plan for possible neovaginal graft.

Assessment of the internal reproductive anatomy should be performed whenever an intra-abdominal procedure is performed. Since patients with cloaca undergo diversion with creation of a colostomy before the definitive surgery, that is the earliest surgical opportunity to assess/document internal reproductive anatomy. However, if entry into the abdomen was not indicated at the time of definitive repair, evaluation of the reproductive anatomy can then be performed at the time of colostomy closure. If an assessment was not performed at either the definitive repair or the colostomy closure, this assessment can be performed later, perhaps with the combination of laparoscopy at the time of creation of an appendicostomy as indicated for bowel management. In complex cloaca patients, the timing of urinary reconstruction is typically long before pubertal onset and may also represent another opportunity for assessment. It is advantageous to perform an assessment of the internal anatomy before the onset of menses



Fig. 1. The ectocervix is seen in the midline of the apex of the vaginal with mucus noted suggesting patency of the cervical region of the uterus.

A



B

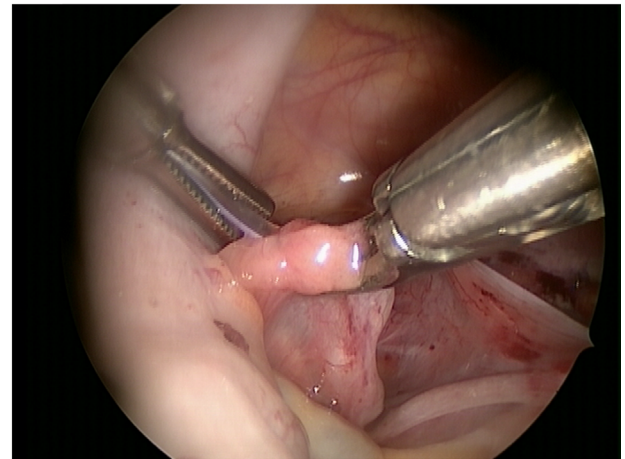


Fig. 2. (A) A #3 pediatric feeding tube is gently placed into the distal aspect of the fallopian tube to instill saline when testing patency of the Müllerian system. (B) The pediatric feeding tube may also be gently placed into the distal fallopian tube laparoscopically to evaluate patency of the Müllerian system.

to assess the possible risk for obstruction to menstrual flow. During either a laparoscopy or laparotomy, the uterus/uteri should be identified. The insertion of the Fallopian tubes into the uterine body should be documented, in addition to the communication of the Fallopian tubes with the ovaries.

Patency of the Müllerian system should also be confirmed. Pediatric feeding tubes can be used to cannulate the distal aspect of each fallopian tube; gentle compression of the fimbriae around the tube allows the antegrade instillation of saline through the Fallopian tube, uterus, cervix, and vagina of the Müllerian system bilaterally [Figure 2(A)]. This assessment of patency can provide reassurance regarding the future outflow of menstrual products. Although almost all females with cloaca have normal ovaries, this should also be confirmed. Assessment of the Müllerian system can also be performed during laparoscopic procedures. Antegrade instillation of saline may be performed using laparoscopic instrumentation with the same pediatric feeding tube [Figure 2(B)]. Alternatively, each cervix can be cannulated with a ureteral catheter during vaginoscopy; dye is then instilled in a retrograde fashion while the fallopian tubes are observed with the laparoscope for spillage from each fallopian tube, thus confirming retrograde patency.

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