Review Article

Obstetrical Outcomes in Adult Patients Born with Complex Anorectal Malformations and Cloacal Anomalies: A Literature Review

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A B S T R A C T

Patients born with complex anorectal malformations often have associated Müllerian anomalies, which might affect fertility and obstetrical outcomes. Other vertebral-anorectal-tracheoesophageal-renal-limb associations, such as renal or cardiac anomalies, could also affect pregnancy intention, fertility rates, and recommendations about mode of delivery or obstetrical outcomes. Associated conditions present at birth, like hydrocolpos, could also potentially affect fertility. Depending on the complexity of the anomaly, primary reconstruction might include vaginoplasty, vaginal interposition, perineal body reconstruction, and extensive pelvic dissection. After the initial reconstruction, patients might have multiple additional surgeries for stoma reversal, bladder augmentation, and creation of conduits, all with potential for pelvic adhesions. Pregnancy intention, fertility rates, mode of delivery, and obstetrical outcomes data are limited in this patient population, making it challenging to counsel patients and their families. We sought to evaluate all available literature in an attempt to better counsel families. A PubMed literature search was undertaken to review this topic. Search terms of "cloaca," "anorectal malformation," "pregnancy," "cloacal exstrophy," "vaginal delivery," and "cesarean section" were used and citation lists from all identified articles were checked to ensure that all possible articles were included in the review. We also outline comorbidities from the fetal period to adulthood that might affect reproductive health. Of the articles on anorectal and cloacal anomalies, 13 reports were identified that covered obstetrical outcomes. They were in patients with previous anorectal malformation, cloaca, and cloacal exstrophy repair. Twenty-four pregnancies were reported in 16 patients. Two ectopic pregnancies, 5 spontaneous miscarriages, 1 triplet pregnancy, and 16 singleton pregnancies were reported with a total of 19 live births. Regarding the method of conception, 15/18 pregnancies occurred spontaneously and 3/18 were via assisted reproductive technology with in vitro fertilization. There were 19 live births, of which at least 8 were preterm. Müllerian anatomy was reported in 8 of 13 articles. Only 2 patients underwent vaginal delivery (1 patient with repaired cloaca malformation had an operative vaginal delivery and 1 patient with repaired imperforate anus with rectovaginal fistula had a normal spontaneous vaginal delivery). The remaining patients all underwent a cesarean section. There were no reported cases of maternal mortality, and maternal morbidity was limited to recurrent urinary tract infections and worsening chronic kidney disease. There is a paucity of information regarding obstetrical outcomes in adult anorectal and cloaca patients. However, patients with previous cloacal repairs have achieved pregnancy spontaneously, as well as with in vitro fertilization. Patients with repaired cloacal malformations are at increased risk of preterm birth and cesarean delivery. Most patients with cloacal anomalies have an associated Müllerian anomaly and therefore have an increased risk of preterm labor. From our review we conclude that contraception should be offered to patients not desiring pregnancy, and cesarean section is likely the preferred mode of delivery. On the basis of this review, we recommend proactive data collection of all such patients to document outcomes and collaboration among providers and between centers devoted to this complex patient population.
Key Words: anorectal malformations, mullerian anomalies, pregnancy, delivery mode, complex pelvic malformations

Introduction

Anorectal malformations (ARMs) encompass a wide range of abnormalities with varying degrees of severity. In women, ARMs include rectoperineal fistula, imperforate anus with rectovestibular or rectovaginal fistula, and cloacal malformations.[1] Cloaca is the most complex ARM in women and is often associated with other anomalies included in a vertebral-anorectal-tracheoesophageal-renal-limb association, which includes vertebral anomalies, ARMs, cardiac anomalies, trachea–esophageal fistula, renal anomalies, and limb defects.[2] In cloacal malformations, there is a confluence of vagina, urethra, and rectum into a common channel, which is identified on the perineum as a single orifice. Urinary, genital, and intestinal openings end together in a single common channel in the perineum, requiring a complex reconstruction to separate and reconstruct the 3 different systems.[3]

In addition to the ARM and potential associated anomalies, many aspects of the clinical course can potentially affect fertility and obstetrical outcomes. These patients require a colostomy at birth. In addition to a colostomy if a hydrocolpos is present, patients with cloacal malformation might require either transperineal or transabdominal drainage to prevent ureteral obstruction. When other life-threatening anomalies have been repaired and the baby is

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growing adequately, primary reconstruction is performed (usually at 6-12 months of age) to restore normal anatomy.[4] Depending on the complexity of the original malformation, it might also involve a posterior sagittal approach only or might additionally require laparotomy.

Müllerian anatomy is variable, but most patients with cloacal malformations have an associated Müllerian anomaly, most commonly uterine didelphys. Depending on the original anatomy, patients might need to undergo vaginal interposition with bowel at the time of their primary reconstruction. This primary repair typically involves perineal body reconstruction and longitudinal vaginal septum, if present, is resected.

Patients with cloacal malformations might undergo additional procedures that might affect their fertility, obstetrical outcomes, or recommendations regarding mode of delivery, such as colorectal procedures including colostomy takedown, bowel resection and reanastomosis, and creation of a Malone appendicostomy. Additional urologic procedures include bladder augmentation, reimplantation of ureters, and creation of a Mitrofanoff appendicovesicostomy.

Traditionally, pediatric surgeons were the primary providers for these girls throughout the newborn period until puberty, after which patients were transitioned to adult providers. Unfortunately, there is a paucity of long-term data in this patient population on adolescent or adult outcomes, particularly regarding sexual functioning, pregnancy intention, fertility rates, mode of delivery, and obstetrical outcomes. Today, a coordinated effort starting at the prenatal stage and spanning the patient's entire life cycle is recommended as this patient population matures into adolescence and adulthood; evidence-based counseling is challenging and best practice recommendations regarding reproductive health, pregnancy, and delivery outcomes are limited.[5] The purpose of this literature review was to determine which obstetrical outcomes have been reported for patients with history of complex ARMs.

Methods

A literature search was performed using the search terms “cloaca,” “anorectal malformation,” “pregnancy,” “cloacal exstrophy,” “vaginal delivery,” and “cesarean section” in PubMed. In addition, citation lists from all identified articles were reviewed to ensure that all possible publications were included in the review. We included any reference reporting a pregnancy.

Results

Our literature review identified 13 articles (of 863 total articles related to general ARM and cloaca postoperative functional outcomes) reporting outcomes related to pregnancy and delivery in patients with cloaca, cloacal exstrophy, or ARMs (Table 1). These 13 articles were a combination of case reports and retrospective reviews; no prospective studies were identified.

Twenty-six pregnancies were reported in 17 patients. Two patients were reported to have a cloacal exstrophy, 2 with imperforate anus with rectovaginal fistula, and 13 with cloacal anomalies. Cloacal anomalies are typically divided into complex or “high” anomalies defined by a common channel longer than 3 cm, and “low” anomalies where the common channel measures less than 3 cm. Unfortunately, none of the articles regarding obstetrical outcomes differentiated between the type of cloacal anomalies, length of the common channel, length of the urethra, or type of reconstruction the patient underwent (complete urogenital separation vs total urogenital mobilization). The maternal age was reported in 11/14 articles with a mean age of 29 years.[6,9–11,14,15,19–31]

Mode of conception was reported in 18 patients. Of these, 15/18 pregnancies occurred spontaneously and 3/18 via assisted reproductive technology with in vitro fertilization. Two of the spontaneous conceptions was the result of anal intercourse with a nonrepaired rectovaginal fistula.

Two ectopic pregnancies, 5 spontaneous miscarriages, 1 triplet pregnancy, and 16 singleton pregnancies were reported with a total of 19 live births. Gestational age was noted in 12 of the pregnancies that resulted in a live birth. Most of these pregnancies (8/12) were delivered prematurely, with the average gestational age of 32.5 (range, 28-36) weeks. These premature deliveries were most commonly the result of spontaneous premature rupture of membranes (PPROM) and not medically indicated deliveries. Only 4 pregnancies (4/12) delivered at term. Mode of delivery was most commonly cesarean section, with 1 normal spontaneous vaginal delivery (rectovaginal fistula) and 1 operative vaginal delivery (repaired cloacal malformation).

There were no reports of maternal mortality. Maternal morbidities most commonly reported were urinary tract infections and worsening chronic kidney disease.

Four patients were noted to have adhesive disease that required modification of the standard surgical technique to avoid injury to the bladder and/or bowel at the time of the cesarean section. One patient was unable to undergo digital cervical examinations because of vaginal stenosis.

Discussion

In this review, we describe all pregnancy information available in the published literature regarding outcomes in patients with a complex ARM, including persistent cloacal malformation and imperforate anus with rectovaginal fistula. We found 13 articles, with varying and somewhat limited information regarding the type of original malformation, surgical repair, associated anomalies, pregnancy intention, length to conception, pregnancy complications, and maternal and fetal outcomes. This literature most likely reflects only a portion of this patient population who have achieved pregnancy, with or without successful conception or live birth. With such limited information, counseling patients and their families is challenging.

It is encouraging, however, to share with patients and their families that patients with cloacal malformations and imperforate anus with Müllerian anomalies have safely undergone pregnancy and given birth to live-born babies. There was no reported maternal mortality, and reported maternal morbidity included recurrent urinary tract infections and worsening of chronic kidney disease. Patients

and their families should understand that most patients in the literature gave birth prematurely and by cesarean section. It is clear that all patients with complex ARMs would benefit from preconceptual counseling. Impairment of renal function, associated cardiac anomalies, vertebral abnormalities, or tethered cord could all affect obstetrical care and surgical planning. Pelvic adhesions, bladder augmentation, and placement of Mitrofanoff and/or Malone conduits could affect type of skin incision and hysterotomy.

Besides the limitations of retrospective studies, the reports often lacked information about the original type of malformation, type of repair, and additional surgeries, as well as associated anomalies. Often the original surgeon performing the initial reconstruction might not necessarily provide reproductive-related health care or follow the patient long term. Over the course of their lives, this patient group has often seen many physicians and their medical records are often fragmented. Ten patients were described as having repaired cloacal anomalies, 2 with imperforate anus and rectovaginal fistula, and 1 with repaired cloacal exstrophy. None of the articles distinguished between the type or complexity of the cloacal malformation and their repair, which could affect fertility rates and obstetrical outcomes. It is possible that poorer outcomes would occur in patients with a more complex anomaly. Therefore, it is difficult to draw any conclusions from these limited data. In our center we are committed to proactively collecting key data items so these can be used for future reproductive health counseling and obstetrical decision-making.[5]

To better understand the potential complications these women might experience during pregnancy and delivery, we outline comorbidities from the fetal period to adulthood that might affect reproductive health.[4] This serves to outline potential real and/or theoretical concerns to be addressed with patients and their families regarding reproductive potential and pregnancy outcomes.

Fetal Ascites/Hydrocolpos

In utero, fetuses with cloacal malformations often develop outflow obstruction because of the confluence of the urinary tract into a small common channel. Hydrocolpos develops when urine flows into the vagina.[32] Very rarely, because of the outflow obstruction, urine refluxes through the fallopian tubes in the abdominal cavity and leads to fetal urinary ascites.[33] After birth, the hydrocolpos typically needs to be drained to avoid urinary outlet and ureteral obstruction.[34] Currently, there is no consensus on the best approach to the initial management of hydrocolpos. Intermittent transperineal catheterization of the common channel and/or transabdominal vaginostomy have both been proposed.[35] Although the main concern is relief of the obstructed urinary tract and prevention of renal damage, complications of an undrained hydrocolpos such as peritonitis and pyocolpos could theoretically also affect the long-term fertility by causing inflammation of the fallopian tubes and vaginal tissue.[32] At present, no long-term data are available about the effect of urinary fetal ascites or hydrocolpos on future fertility. We are unable to comment regarding the presence of hydrocolpos and type of drainage on future fertility.

Müllerian Anomalies

More than half of patients born with a cloacal malformation have an associated Müllerian anomaly, most commonly uterine didelphys, but other anomalies including unicornuate uterus and uterine agenesis have been reported as well.[36] Patients with Müllerian anomalies have an increased risk for adverse obstetrical outcomes, primarily preterm delivery, PPROM, small for gestational age newborns, and increased rates of cesarean delivery because of fetal malpresentation.[37] However, most of the current obstetrical outcomes data in the literature are on the basis of women with isolated Müllerian anomalies,[37,38] and it is unclear how a Müllerian anomaly in the setting of a cloacal malformation with previous hydrocolpos and pelvic and/or genital reconstruction might further increase the risk of infertility or poor obstetrical outcomes. The patients reported in the literature showed, as expected, a high rate of Müllerian anomalies, with 5 patients with uterine didelphys, 2 with unicornuate uterus, and 1 with bicornuate uterus. Not surprisingly, there was a significant proportion of patients who experienced PPROM, preterm delivery, and cesarean delivery. Because it is unclear how the presence of a Müllerian anomaly in the setting of a cloacal malformation with or without previous hydrocolpos and pelvic and/or genital reconstruction affects fertility and/or obstetrical outcomes, patients and their families for now should be counseled similarly on the basis of known information about isolated Müllerian anomalies. Furthermore, the risk of cervical incompetence is not reported in cloacal patients but might be relevant.[7]

Pelvic Adhesions

Most patients with cloacal malformations and other types of complex ARMs undergo at least 3 laparotomies; initially to perform a colostomy in the neonatal period with potential hydrocolpos drainage, secondarily during primary reconstruction, and last during colostomy reversal. In the most severe cases, children might also undergo subsequent abdominal or pelvic procedures for additional urinary reconstruction, such as bladder augmentation as we found in 1 patient in this series.[12,17] In a long-term follow-up of patients with cloacal malformations, Warne et al reported a 36% rate of reoperation in patients with a previous cloacal repair.[39] These procedures could lead to intra-abdominal and pelvic adhesions. Reoperation is more difficult, often takes longer, and is associated with a higher complication rate secondary to pelvic adhesions.[40] As mentioned earlier, adhesions might also be present because of previous in utero or neonatal hydrocolpos and ascites. Adhesions have been associated with dyspareunia, infertility, and bowel obstruction.[40]

Patients and their families should be counseled that adhesive disease might increase the complexity of a cesarean section. In this patient population, 4 patients were noted to have adhesive disease that required modification of the standard surgical technique to avoid injury to bladder and/or bowel.[11,17,19,40,41] This modification of a standard surgical cesarean section has also been described in other complex pelvic anomalies such as bladder exstrophy.[41] In 1

Table 1
Reported Obstetrical Outcomes in Women with Complex Anorectal Malformations

Reference	Patient n	Age, years	Time to conception	OB history	Müllerian anatomy	Associated anomalies	Type of reconstruction	Vaginal status
1. Salvi et al[6]	1	33	Unknown	G6P0050	Didelphic	Renal cross fused, duplex, system	PSARVUP BN reconstruction; abdominoplasty	Vaginal dilation for stricture twice
2. Shrim et al[7]	1	37	>4 Years	G1P0	Didelphic	Renal agenesis, previous pyosalpinx, persistent bilateral hydrosalpinx	PSARVUP twice	Vaginoplasty at 33 years
3. Greenberg et al[8]	1	27	>2 Years	G1P0	Didelphic	N/R	PSARVUP; multiple laparotomies; BO	Introitoplasty; perineal body reconstruction; native vagina pull-through
4. Greenberg et al[9]	1	29	2 Years between first pregnancy and second pregnancy	G2P2	Didelphic	N/R	PSARVUP; multiple laparotomies; BO	Introitoplasty; perineal body reconstruction; native vagina- through
5. Sato et al[10]	1	24	N/R	G1P0	Unicornuate	Renal agenesis	PSARVUP; resection of atretic right Müllerian structure	Sigmoid vaginoplasty
6. Resnik et al[11]	1	28	N/R	G1P0	N/R	N/R	PSARVUP; ureteral reimplant	Multiple urethral and vaginal dilations
7. Waters[12]	1	28 and 29	6 Months	G1P0 and G1P1	N/R	N/R	Rectum and vagina ended together; common channel urethra	Rectovaginal fistula not repaired
8. Maruotti et al[13]	1	37	N/R	G1P0	Bicornuate	CKD	PSARVUP; bladder augmentation	N/R
9. Hendren[14]	4	N/R	N/R	N/R	N/R	N/R	N/R	N/R
10. Couchman et al[15]	1	N/R	N/R	N/R	N/R	N/R	N/R	N/R
11. Gezer et al[16]	1	21	N/R	G1P0	Didelphic; no left ovary and tube; only present the right horn	Bladder exstrophy, ARM	Bladder augment, PSARVUP	Single vaginal opening
12. Ljubić et al[17]	1	19	N/R	G1P0		ASD repair; no previous perineal repair	None	Imperforate anus with rectovaginal fistula
13. Hamai et al[18]	1	26	N/R	G1P0	Unicornuate	Previous nephrectomy	Imperforate anus with rectovaginal fistula; neorectum used for coitus	Rectovaginal fistula not repaired

Obstetrical history: G (gravida) P (para).
abx, antibiotics; ARM, anorectal malformation; ASD, atrial septal defect; BN, bladder neck reconstruction; BO, bowel obstruction; CKD, chronic kidney disease; FAVD, forceps-assisted vaginal delivery; IVF, in vitro fertilization; N/A, not applicable; N/R, not reported; OB, obstetrical; PL, preterm labor; PRM, premature rupture of membranes; PSARVUP, posterior sagital anorecto-vagino-uretroplasty; UTI, urinary tract infection; VAVD, vacuum-assisted vaginal delivery; VUR, vesicoureteral reflux.

case, a transvesical cesarean section was performed because of multiple adhesions and abnormal position of the bladder.[11] Surgical planning must address not only potential adhesive disease, but also altered anatomy and the presence of a Malone appendicostomy and Mitrofanoff appendicovesicostomy.

Perineal Body Scarring/Vaginal Introital Stenosis

We previously reported a high incidence of reoperation in 28 girls (2 with cloacal malformation) with previously repaired ARMs.[19] The most common indications for reoperation were an abnormal perineal body and introital

Vaginal intercourse	Spontaneous	Implantation	Number of fetus	Pregnancy complications	Vaginal delivery	Episiotomy	Cesarean section	Follow-up
Painful	Yes	Right uterine horn	1	Abdominal pain; cervical exam not possible; recurrent UTI PL	N/A	N/A	28 Weeks Pfannestiel (no adhesions)	N/R
N/R	IVF; no patent fallopian tubes	Left uterine horn	1	Recurrent UTI	N/A	N/A	37 Weeks	3 days N/R complication
Yes	Yes	Left uterine horn	1	PL	34 Weeks VAVD FAVD (Tucker-McLean); sulcus vaginal laceration	No	N/A	6 Weeks; well healed
Yes	IVF; no patent fallopian tubes	Left uterine horn	Triplets	PL	N/A	N/A	30 Weeks; transverse hysterotomy	N/R
Yes	Yes	Right fallopian tube	1	Ectopic pregnancy treated surgically	N/A	N/A	N/A	4 Days; well healed
No, but spontaneous conception	Yes	N/R	1	PRM; 3 weeks' bed rest	No	N/A	34 Weeks; transvesical (multiple adhesions)	6 Months; well healed
No introitus; transrectal coitus	Yes, twice	N/R	1 and 1	P0: none P1: none	No	N/A	40 Weeks; supravesical extraperitoneal twice	7 Years; good functional outcome
N/R	Yes	N/R	1	Evolution CKD; liver disease; UTI	No	N/A	33 Weeks; longitudinal hysterotomy	1 Month; improved renal function
Yes	Yes	N/R	N/R	N/R	No	N/A	Yes, 4 patients	N/R
N/R	IVF	N/R	1	Complex preterm delivery	No	N/R	N/R	N/R
Yes	Yes	Yes	1	UTI grade 2 VUR requiring abx twice; fetal distress; preterm birth	No	N/A	36 Weeks; fundal uterine incision/ supraumbilical midline incision	N/R
No anal opening; normal introitus	Yes	N/R	1	N/R	Yes	N/R	38 Weeks; vaginal delivery with no complications	N/R
No introitus, transrectal coitus	Yes	Rudimentary left horn	N/A	Ectopic implantation	N/A	N/A	N/A	N/A

stenosis. In reproductive-aged women, an abnormal or reconstructed perineal body might lead to an increased rate of third- and fourth-degree perineal lacerations during vaginal delivery, pelvic floor dysfunction, and dyspareunia.[19] Particularly in patients with adequate fecal continence, avoiding obstetrical anal sphincter injuries is paramount. An abnormal perineal body might affect recommended mode of delivery (favoring cesarean delivery) and obstetrical outcomes (increased risk of third- and fourth-degree lacerations) in patients with complex anorectal cloacal malformations.

In the current review 5/13 patients reported an abnormal vaginal introitus in adulthood after their primary cloacal repair in childhood, which led to additional surgical procedures (introitoplasty/vaginoplasties).[6,7,9,11] We also identified 1 patient who was unable to undergo digital cervical examinations because of vaginal stenosis. Couchman et al recently investigated the long-term outcomes of early

vaginoplasty performed as part of the initial cloacal repair and reported that 56% of patients had required 1 or 2 further vaginal reconstructions because of inability to have intercourse or reported dyspareunia.[15] Patients and their families should be counseled that vaginal and/or introital stenosis can affect ability for vaginal penetrative intercourse, ability to perform routine obstetrical care, as well as recommendations regarding mode of delivery.

Vaginal Interposition

Depending on the complexity of the cloacal malformation, a vaginal interposition might be needed at the time of primary reconstruction or during subsequent reconstructive procedures.[20] The literature notes that this is necessary in 22%-26% of cases.[4,15,21] There are many options available for vaginal substitution, each offering unique advantages and disadvantages. Surgeon preference and experience clearly influences the choice of neovagina[22] but all vaginal interpositions have the potential to affect vaginal penetrative sexual intercourse and recommendations regarding mode of delivery.[1] Although the timing for vaginal reconstruction remains controversial, most pediatric surgeons create a bowel neovagina at the time of the primary reconstruction early in life.[23] There are some reports on assessing sexual health in women with intestinal vaginal interpositions; it is reassuring that most report a low rate of complications, with most reporting normal vaginal intercourse, presence of orgasm, and adequate lubrication. However, 18% of patients reported dyspareunia.[24–27]

In this review we identified 1 case of pregnancy in a patient with a bowel neovagina that resulted in an ectopic pregnancy.[10] Patients and their families can be counseled that penetrative vaginal intercourse and conception is reported with bowel neovagina. It is important to note that most articles involving vaginal interpositions are on patients with Mayer-Rokitansky-Küster-Hauser syndrome, not cloacal malformations. Cloaca patients undergo more complex reconstructive procedures, so it is not possible to fully extrapolate the long-term sexual data from Mayer-Rokitansky-Küster-Hauser syndrome patients to a cloaca cohort.

Mode of Delivery

Counseling on the mode of delivery is complex and involves a multidisciplinary approach, with attention paid to the patient's surgical history, anatomy, degree of fecal and urinary continence, current pregnancy comorbidities, and reproductive life plans. Women with a previous cloacal repair, vaginal interposition, or urinary reconstruction are typically counseled to avoid vaginal delivery.[1,14,28,29] Although we acknowledge the paucity of evidence-based data, we agree with this recommendation on the basis of unique anatomic features in this population after cloacal reconstruction. All cloaca patients have undergone some type of repair of the vagina and perineal body, which likely places them at an increased risk of damage to the vagina and/or perineal body as a result of vaginal delivery, because previously repaired/scarred tissue does not stretch as well as unrepaired tissue. They theoretically have an increased risk of obstetrical anal injury, and higher-grade perineal lacerations might result, potentially resulting in impaired fecal continence. Furthermore, a reconstructed urethra, bladder, and/or bladder neck might suffer functionally from significant trauma caused by vaginal delivery.[30] Although a reconstructed vagina could be fully functional for menstrual egress and coitus, the presence of stenosis scarring might make it difficult to perform transvaginal sonography or a digital cervical examination. Limited access to the cervix (or cervices) makes assessment difficult in patients with threatened preterm labor.

Although we do believe that cesarean section is safer in the cloaca malformation population, it is not without risks. The high rate of vaginal interposition (which depends on a single vascular pedicle, usually a branch of the left colic artery), reconstructed urethra and bladder, continent stomas such as Malone appendicostomy or Mitrofanoff appendicovesicostomy, and the presence of pelvic adhesions, make cesarean section difficult in these patients.[31] Previous surgical reconstructions and distorted pelvic anatomy increase the risk of damage to surrounding structures; particularly if mobilization of the uterus is necessary or preferred during closure of the hysterotomy, or if the closure of the hysterotomy incision is not done in situ, but rather after lifting the uterus through the abdominal incision, a common practice of many obstetricians.[31] If cesarean section is the chosen mode of delivery, whether emergent or elective, a surgical team that includes someone familiar with the anatomy might reduce the risk of injury. It is of utmost importance that a reconstructive surgeon familiar with the anatomy be present to reduce the risk of injuring the bladder or vascular pedicles. This is most important if the patient has had previous bladder augmentation, a continence stoma, or a vaginal replacement. Patients living far from a maternity unit with suitable resources might consider a planned elective cesarean delivery to avoid the potential for emergent cesarean section elsewhere, in an effort to minimize such injuries.[28] Encouraging patients to obtain operative reports that document their previous abdominal and pelvic procedures is important.

A successful operative vaginal delivery has been reported in 2 patients. The first one had a previous cloacal malformation repair, although specific details about the type of the cloacal malformation, the length of the common channel, or how the vagina was reconstructed were not included in the report. The patient was noted to have a vaginal laceration that healed by 6 weeks postpartum, but there is no additional obstetric information provided.[8] The second patient had an unrepaired congenital rectovaginal fistula and underwent a successful normal spontaneous vaginal delivery without complication.[17]

Renal Impairment

Patients with persistent cloaca are at high risk for chronic kidney disease and upper renal tract deterioration.[42] In some series nearly 80% of patients with cloacal malformations had long-term renal impairment, and 5% have experienced end stage renal failure.[43] It is well known that there are renal risks during pregnancy after genitourinary reconstruction. Alterations in renal function associated with

urinary tract reconstruction and pregnancy have been observed in some series, because these patients are more likely to have underlying hydronephrosis or renal insufficiency.[44] Patients with previous pelvic surgery and good renal function or only mild impairment might have no notable change in function, although they might still be at greater risk for an intervention with nephrostomy tube or stent placement for obstruction during pregnancy.[45] The highest risk period for obstruction appears to be between 20 and 28 weeks of gestation, and patients with underlying renal impairment might require ultrasonographic surveillance every 2 weeks during that time.[46] In this series 2 patients delivered via cesarean section at 33 and 34 weeks gestation secondary to worsening renal function in the setting of chronic kidney disease that risked maternal health.[13,16] Although not reported in the literature, patients with chronic kidney disease might also be at increased risk of gestational hypertension and preeclampsia.

Cardiac Anomalies

The observed prevalence of congenital heart anomalies among patients with ARM is much higher (9%-37%) than that observed in the general population (approximately 1%).[47] Congenital heart disease is present in 17% of patients with cloaca, similar to the rate observed in the other types of ARMs.[48] The most frequent defects are ventricular septal defects, atrial septal defects, restrictive patent ductus arteriosus, and persistent foramen ovale. A small percentage of patients might present with more complex cardiopathies such as tetralogy of Fallot or primary pulmonary hypertension.[48]

For women with cardiac anomalies, individualized preconception counseling and pregnancy planning should be a vital component of their medical management, particularly if the heart defect is present in a patient with cloaca. Women with simple congenital heart defects generally tolerate pregnancy well. However, women with complex congenital defects, with or without surgical repair and/or residual defects, should be managed in tertiary care centers with a multidisciplinary team of physicians experienced in adult congenital heart disease and high-risk obstetrics who collaboratively participate in pregnancy planning, management, and care through childbirth and postpartum.[49] In our review, 1 patient had a repaired atrial septal defect and underwent an uncomplicated normal spontaneous vaginal delivery at term.[17]

Contraception

All menstruating patients, including those with ARMs, should undergo counseling about risk of unintended pregnancy and offered safe and effective contraception. Contraceptive counseling must consider Müllerian anatomy as well as renal and cardiac comorbidities. Sexual practices should also be discussed because conception was reported after anal rather than vaginal penetrative intercourse. Patients might not recognize the risk of pregnancy and potential need for contraception despite the lack of penetrative vaginal intercourse. In this review, there were 2 reported ectopic pregnancies, which remains a life-threatening condition. Pelvic adhesive disease and Müllerian anomalies are both risk factors for ectopic pregnancy.[10,18] Surgical treatment of ectopic pregnancy might be difficult because of the complexity of a surgical intervention and medical treatment with methotrexate is contraindicated if there is chronic kidney disease.

Anesthesia Concerns

Tethered cord syndrome (TCS) is a diverse clinical entity and is caused by excessive tension on the spinal cord.[50] In patients with ARM and cloaca, the incidence of TCS is estimated between 15% and 30% of cases, which might underestimate the true incidence because some of these patients are asymptomatic and are underdiagnosed.[51–53] In cases of cloacal exstrophy, this prevalence increases to 90%-100%.[54,55]

Spinal anesthesia is a safe, routinely performed procedure in vaginal and cesarean section deliveries. It is recommended that the dural puncture be made at or below the L3-L4 intervertebral space because the conus medullaris ends at the inferior margin of L1 in most adults and extends to L2 in a few persons.[56,57] In patients with TCS, the conus medullaris often terminates at a lower level, often extending to L5-S1. In this condition, dural puncture should be avoided because of the possibility of injury of the spinal cord.[50] In preparation for potential spinal anesthesia in patients with ARMs/cloacal malformations with an unknown spinal status, a spinal magnetic resonance imaging examination should be performed to evaluate for a spinal malformation, which might alter the delivery mode of anesthesia.[58] No anesthesia information was included in this reported patient group. None of the articles in this series mentioned the patient's spinal status.

Conclusions

There are multiple potential factors that can affect fertility, pregnancy course, and mode of delivery in patients with cloaca specifically and ARMs generally. These comorbidities should be taken into account when counseling families and patients. It is crucial to facilitate good communication between pediatric surgeons and obstetricians at the time of transitioning care, as well as to report their pregnancy outcomes with detail paid to the type of cloaca, type of reconstruction, and associated anomalies to better understand obstetric outcomes in this high-risk population. Because of the complex anatomy and underlying medical comorbidities in cloaca patients, we advocate that they are managed by high-risk obstetricians if possible in a tertiary hospital, with a multidisciplinary collaborating team.

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