Children born with anorectal malformations (ARMs) face a variety of medical considerations throughout their lifetimes. At birth, these challenges are difficult to prioritize, predict, and overcome. As individuals with ARM grow older, they should collaborate with their medical team to consider their reproductive and sexual health. Current literature indicates that a patient’s knowledge and status of their own reproductive and sexual health greatly impacts their overall health.

The internal reproductive system in females develops from embryonic structures called Müllerian ducts (one duct system on each side of the body). These paired ducts migrate to fuse and form one reproductive tract. The reproductive tract usually has a fallopian tube on each side of the body connected to a single uterus (with one cervix) and a single vagina. In some cases, medical professionals may refer to these tissues as Müllerian structures, especially if the development remains incomplete. During fetal development, the ovaries develop from a different source of tissue than the other reproductive organs and are therefore typically normal in patients with ARM. The degree to which the Müllerian structures are impacted contributes to the reproductive considerations in females with ARM.

Assessment of the female reproductive structures ideally should occur as early in life as possible. This assessment may be combined with other procedures requiring anesthesia such as cystoscopy or the definitive anal repair. Additional opportunities to evaluate Müllerian structures could occur at follow-up procedures such as the surgical creation of continent or catheterizable channels. If surgical assessment is not an option, then imaging, such as a pelvic ultrasound or magnetic resonance imaging (MRI), may be helpful. The Müllerian structures can be visualized more clearly on imaging if estrogen is present in the patient. Therefore, ideally any imaging assessment should be done between the ages of 0-6 months when estrogen is present due to maternal exposure, or at the onset of puberty (usually marked by the start of breast development). Growth and maturity of the patient will also help to determine the shape, size, and quality of the uterus and cervix.

At the time of any planned surgical evaluation, it is beneficial to use the opportunity to determine the development and configuration of the Müllerian structures. Variations in development can occur depending on the type and severity of the ARM. Individuals may have a didelphys uterus (two half-uteri), a bicornuate uterus (a single heart-shaped uterus), a unicorneate uterus (half uterus) or a normally shaped uterus. Within these scenarios, the cervix and/or the vagina can also be single or duplicated.

In some circumstances, one of the Müllerian structures may be under-developed and/or not connected to the larger, more developed uterus. In this case, it is generally recommended to remove the under-developed structure to prevent menstrual obstruction and pain in the future. Leaving the other normally-developed well-functioning side for future menstruation and pregnancy is a safe option.

With the results of these assessments in mind, the impact of reproductive health in young adult female patients with ARM is
centered on four components:

- Sexuality and sexual function/health
- Becoming pregnant
- Maintaining pregnancy
- Delivery outcome of pregnancy

**Sexuality and Sexual Function**

An individual’s state of continence for urine and/or stool has been linked to their psychosocial and psychosexual development. These issues can affect a young woman’s self-perception and hinder interactions with her peers. Recent literature highlights an association of impaired sexual function with low self-esteem, body dissatisfaction, anxiety, and depression, as well as impaired social and emotional function. Patients with ARMs may experience these symptoms or conditions and thus may benefit from additional psychological support during adolescence and into adulthood.

Sexual function in terms of the ability to achieve orgasm/pleasurable intercourse also has an impact on sexual health. The clitoris is usually unaffected; therefore, satisfying sexual stimulation and orgasm should be achievable. It is important for all women, regardless of their anatomy, to understand their anatomy and learn what is pleasurable to them. Women with an ARM typically are well-versed with their anatomy and can sometimes be at an advantage (compared to other women) in their ability to interact with their partner about their anatomy and guide them about what feels pleasurable.

**Becoming Pregnant**

Becoming pregnant is typically not a significant concern in females with ARM since the ovaries, responsible for housing and releasing eggs, are typically normal. However, the decision to pursue pregnancy should be carefully considered and planned. A dialogue with the primary care provider, obstetric consultant, and the individual’s significant other can help in deciding if carrying a pregnancy is recommended or possible. Other components should be considered before planning a pregnancy, such as an evaluation of kidney health, the care and function of continent channels such as a Malone or Mitrofanoff, and potential risk of complications due to other complex reconstructive surgeries, such as bladder augmentation. Until medical clearance for pregnancy can be obtained, contraceptive use is encouraged for females with ARM.

In rare cases, patients who have had numerous abdominal surgeries may develop abdominal or pelvic scarring (adhesions). The scarring may involve the fallopian tubes which can prevent natural conception. The patency of the tubes can be confirmed with straightforward testing ordered by an obstetrician/gynecologist, if indicated. Some papers have described a possible higher risk for miscarriage or inability for proper implantation of a pregnancy in these cases.

**Maintaining a Pregnancy**

Maintaining a pregnancy until a reassuring gestational age is the highest priority. An important focus is around the quality and shape of the individual’s uterus. In patients with uterine abnormalities, the size and pliability may be decreased in comparison to a more fully developed uterus. This may contribute to a higher risk of preterm contractions, labor and delivery. Considerations to review with the medical team also include the risk of an unusually placed placenta or the possibility of a baby in a “breech” position (feet or buttocks first instead of head first).

In a recent review from our center, the risk of early delivery was confirmed. The review identified 40 pregnancies in 25 women with ARMs. More than half of the patients delivered early, although far enough in
the pregnancy for the preterm baby to generally do well (>32 weeks gestational age). The majority delivered by cesarean section (64%). More research is needed in this area to determine best practice for delivery for women with a history of a repaired ARM.

Delivery
The route of delivery should also be discussed by the patient and the medical team. The decision to deliver vaginally or by Cesarean section (also known as C-section) is influenced by other medical factors and previous surgical treatment. Many factors are reviewed when deciding the safest mode of delivery such as prior types of surgery, the size/integrity of the perineal body, and the site of catheterizable channels.

Previous surgeries in both the abdomen and the perineal body need to be considered. Multiple surgeries in the abdomen predispose women to the development of adhesions and scar tissue, making subsequent operations more difficult. The obstetric and surgical team should work together in preparation for delivery by reviewing previous surgeries, current anatomy, and impact on mode of delivery. Previous skin incisions can often be utilized by the obstetricians/surgeons during possible C-sections to minimize additional visible scars. If a C-section is chosen by the patient and health care team, it may be helpful to involve surgeons from other disciplines during surgery to protect reconstructed anatomy such as a Malone, bladder augmentation, or a Mitrofanoff.

The perineal body can be more fragile due to previous surgeries such as anoplasty. A smaller or scarred perineal body causes an increased risk for a tear to occur during a vaginal delivery. Such a tear (even when repaired) may increase the risk of fecal incontinence if involving the rectum. Protecting continence is always of utmost importance whether at the Malone/Mitrofanoff or the urethra/anus.

New Innovations
In rare situations, other conditions that affect the ovaries occur in conjunction with an ARM, such as primary ovarian insufficiency, at-risk inheritable conditions such as BRCA (breast cancer predisposition) positive status, or other congenital anomalies. In these situations, fertility preservation procedures may be considered. Fertility preservation is defined as the process of saving an individual’s eggs, sperm, embryos, or reproductive tissue for future biological children. The standard methods available to females consist of embryo or oocyte (egg) freezing. Ovarian tissue cryopreservation (OTC) is an investigational option primarily for younger patients. OTC is a surgical procedure performed to remove part or the entire ovary to freeze (cryopreserve) for possible future use for childbearing. OTC for patients with ARMs remains rare and is only used in unique situations.

Uterine transplantation is currently under investigational protocol for individuals with other congenital conditions such as solitary uterovaginal agenesis. It is not currently offered for patients who have undergone complex pelvic reconstruction such as patients with a history of cloaca, but with further research, it may be a possibility in the future.

Alternatives
Families are formed through various avenues. Gestational surrogacy, also known as gestational carriers or surrogates, is an option for women who may be unable to safely carry a pregnancy. This is a process where the embryo is created via in vitro fertilization (IVF), using the eggs and sperm of the intended parents or donors, and then transferred into the surrogate’s uterus to carry the pregnancy. Therefore, the child is not biologically related to the gestational carrier, but is biologically related to the person whose eggs were used.
Adoption is another way for individuals to have children. Foster parenting and blended families are additional ways to form a family. Alternatively, some women choose not to raise or carry children.

**Key Sources:**


Cincinnati Children’s Medical Center Hospital Comprehensive Fertility Care & Preservation Program: [https://www.cincinnatichildrens.org/service/f/fertility-preservation](https://www.cincinnatichildrens.org/service/f/fertility-preservation)


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