Published in final edited form as:

Semin Pediatr Surg. 2020 December ; 29(6): 150987. doi:10.1016/j.sempedsurg.2020.150987.

Long-term urologic and gynecologic follow-up and the importance of collaboration for patients with anorectal malformations

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Abstract

Anorectal malformations (ARM) are well recognized to be associated with anomalies in other organ systems. The introduction of screening protocols has increased the diagnosis of these anomalies and greater collaboration with other specialties has influenced the treatment and follow-up of patients with ARMs. Much of the medical literature regarding the treatment of anorectal malformations has focused on technical details of operations and early post-surgical outcomes. Recently, an increase in published data regarding the long-term sequelae of an ARM diagnosis has resulted in an emphasis extended follow up in this population. Patient support groups have highlighted complex issues in ARM patients persist into adulthood have advocated for improved transitional care. This article describes the benefits of long-term follow-up and identifies key issues in ARM patients with respect to urologic and gynecologic health. A collaborative model of care is outlined and suggested timings of screening for potential problems is described.

Urology

Reports of associated urological anomalies in ARM patients widely vary with reports from 18% to 85%.^{1,2} The range can be attributed to the differences in screening practices and completeness of screening. Most series with active screening protocols report a prevalence of around 50% across all ARM types.^{3,4} Even in developed countries there is often a lack of standardized screening which can result in an underestimation of the prevalence.⁵ It is critical to have urologic involvement in the care of patients with ARM as chronic kidney disease is the highest risk of decrease in life expectancy with end stage renal disease rates reported as high as 6.4% in complex ARM subtypes.⁶ Having a urologist's involvement in the collaborative center caring for these patients afford focused attention on bladder and kidney management in the effort to minimize potential long-term problems.

Case 1

A 12 yr old girl who had a previously repaired short common channel cloaca comes to your multi-disciplinary clinic for the first time. She was followed up by the pediatric colorectal surgeon and has gained fecal continence on a stimulant laxative regimen. Until now she has been dry for urine however in the last 6 months has had 2 urinary tract infections and developed urinary incontinence. She has a history of not passing urine more than twice a day and drinks less than 500mls/day. As the urologist you review her post-natal work-up and see that as a baby she had a normal renal tract ultrasound. No spinal imaging had been obtained. Today's renal tract ultrasound shows a thick-walled bladder with bilateral hydronephrosis. Her urinary creatinine is 2mg/dl. You organize urodynamics which reveal a high pressure, non compliant bladder.

There are 4 main factors that influence long term urological health:

1. The presence of intrinsic kidney or bladder anomalies at birth
2. The presence of sacral dysgenesis
3. The effect of the operative intervention to correct the ARM
4. The effect of spinal cord tethering causing bladder dysfunction which may change over time.

A normal renal and urinary tract assessment at birth does not preclude the development of potentially serious long-term complications which may remain clinically silent until there is severe chronic kidney disease.

A suggested screening protocol consists of:

Diagnostic work-up in neonatal period

- Renal tract ultrasound shortly after birth. The degree of hydronephrosis is likely to be underestimated if performed before 48 hours of age.
- Sacral X-rays
- VCUG on selected patient basis (distal ureteric dilatation on ultrasound, hydronephrosis)
- Spinal ultrasound at birth and MRI spine if there are abnormalities found on ultrasound.

Monitoring at outpatient visits

- Take a urological history at outpatient visits including whether the child has had urinary tract infections and the development or loss of urinary continence
- Annual renal tract ultrasound (More frequent if hydronephrosis or scarring)
- Annual renal blood work including cystatin C and eGFR
- Screening questions about symptoms of tethered cord

- Consider urodynamics if there are infections, worsening hydronephrosis, renal scarring or poor renal growth on renal ultrasound or in presence of symptoms of tethered cord

Renal failure

Preservation of renal function is arguably the most important role of the urologists caring for children with ARM. The type of malformation significantly affects the risk of developing end stage renal failure. In a series of 20 children with ARM who underwent kidney transplants, 13 had a cloaca and 2 had rectobladder neck fistulas.⁷ In this series over half had presented with renal failure at birth and interestingly the authors could not identify missed opportunities to prevent renal deterioration. The study found that just under 1% of ARM patients in their care underwent a kidney transplant. However, this may well underestimate the true lifetime risk of chronic kidney disease (CKD) because it looked at a database of patients treated in a pediatric center with relatively short follow-up. In a longitudinal study of cloaca patients up to 50% were found to have CKD and 11% progressed to end stage renal failure.⁶ In a resource limited setting then patients with 'higher' type malformations (cloacas in female patients and rectobladder neck fistulas in males) should be prioritized for urology input. Research has also demonstrated that abnormal sacral development is correlated with the presence of urological anomalies and those patients with lower sacral ratios are at higher risk of significant problems.⁴

The presence of a solitary kidney also increases the risk of progressing to renal failure. Das et al reported that 18% of their patients with a solitary kidney developed end stage renal failure at a median of 70 months of age.⁸

Hecht et al correlated the presence of ectopic ureters with an increased risk of chronic kidney disease needing renal transplant.⁹ However many of these patients had cloaca or bladder neck fistulas making it difficult to quantify what effect the ectopic ureter in itself had and how much was due to the original malformation and other associated anomalies. In patients with unilateral ectopic ureter 59% had decreased renal function in the ipsilateral kidney however worryingly they also showed that up to 29% also had dysfunction in the contralateral kidney.

Case 2

A 6 yr old boy with a previously repaired rectobladder neck fistula is incontinent for urine and feces and his parents are anxious for a solution now that he is in kindergarten. The pediatric colorectal surgeon has trialed a rectal enema regimen during a bowel management week which worked initially but now the boy is refusing to allow his parents to perform the enemas rectally. She has recommended a Malone appendicostomy. You organize urodynamics which shows a large capacity, flaccid, low pressure bladder which does not empty effectively. The boy has had several urinary tract infections over the past year. You think that a Mitrofanoff would enable him to gain continence and reduce the number of urinary tract infections. You discuss the case with your colorectal colleague and decide on a joint procedure possibly entailing splitting the appendix. At operation the appendix is short

and your colorectal colleague graciously allows you to use it for the Mitrofanoff whilst she creates a tubularized cecal flap for antegrade colonic enemas.

This case emphasizes that while tissue sharing is an important principle, splitting the appendix that is shorter than 7 cm may result in a mitrofanoff that is too short to tunnel into the bladder adequately or may result in difficulty getting the channel to reach the skin. If the appendix is too short, it should preferentially be used for the mitrofanoff and neo-malone or cecostomy tube can be placed for antegrade enemas.

Bladder Dynamics

Pre-operative urodynamic studies (UDS) in ARM patients are usually normal.^{10,11} Versteegh et al reported pre and post-operative UDS in a select group of their ARM patients and found that 37 of 51 (71%) patients had normal preoperative studies. However, they had only studied a small proportion of their patients (51 out of 341) and had investigated patients with more complex malformations which suggests that an even higher percentage of ARM patients will have normal bladder dynamics prior to operative repair.

Boemers et al found that 46 of 52 ARM patients with a normal sacrum had completely normal UDS, 6 patients had some detrusor instability however they stated that this finding is common in many young children and therefore classified 98% of studies as normal.¹² The presence of a sacral anomaly significantly influenced bladder function; 21 out of 38 (55%) patients had abnormal UDS. The type of sacral anomaly was important-those only missing S5 segment tended to have normal UDS however all patients who had absent S5 segment and at least half of S4 segment missing had lower urinary tract dysfunction.

Warne et al evaluated pre-operative UDS in cloaca patients as compared to other ARM patients and found that 9 out of 10 had abnormal UDS, namely detrusor overactivity.¹³ In this series only 1 of the 10 patients had a normal spinal cord and sacrum.

The effect of operative repair on bladder function seems to depend on the type of malformation and how much dissection is required close to pelvic nerve plexuses lying within the endopelvic fascia on the anterior surface of the rectum. Boemers et al were able to demonstrate urodynamic changes consistent with autonomic denervation of the bladder in 3 out of 32 patients.¹⁰ These were boys with rectourethral fistulas, 2 of whom had required a combined transabdominal and PSARP approach. Warne et al also showed a deterioration in bladder function in 5 out of 10 cloaca patients post operatively. The pattern changed from detrusor overactivity pre-operatively to a flaccid, atonic bladder post-operatively suggesting lower motor neurone damage. These were all patients with a long common channel (>3cm) who had been repaired with a Total Urogenital Mobilization (TUM) approach.

A more recent study by Versteegh et al did not find that TUM for cloacal reconstruction significantly changed pre and post-operative UDS for most patients.¹¹ They commented that in their patients they had not needed to divide all of the suspensory ligaments of the bladder and had maintained the bladder neck above the pelvic floor. There is accumulating evidence that TUM in long common channel cloacas or short common channel cloacas with a short

urethral take off may result in urinary incontinence and is likely not a good surgical option for primary repair as a result.¹⁴

Versteegh et al also highlighted that pre-operative UDS does not necessarily correlate with those patients who develop poor urological outcomes.¹¹ 50% of their patients with impaired urological function had normal pre-operative UDS and therefore they concluded that routine UDS pre-operatively was not indicated for all patients. This highlights that ongoing urological follow-up is crucial to make sure that deterioration in bladder function is appropriately diagnosed and managed.

The presence of a tethered cord in patients with rectobladder neck fistulas had been shown to be associated with very poor outcomes for bladder and bowel control.¹⁵ Bjorsum-Meyer et al were also able to show that the presence of spinal defects impacted on ICIQ voiding subscale in a wide range of ARMs in adult male and female patients however this did not significantly change the ICIQ incontinence or filling dynamics.¹⁶ Totonelli et al showed that untethering surgery for ARM patients helped neuromotor symptoms however it did not significantly improve bowel or bladder symptoms.¹⁷ It is not clear whether there is a role for prophylactic spinal cord untethering in ARM patients to try and minimize any effect on urological function. The active involvement of neurosurgery in a collaborative center looking after ARM patients is very helpful to ensure that opportunities for intervention are not missed.

Urinary Continence

Achievement of urinary continence is strongly related to the underlying anorectal malformation. Strenstrom et al found no significant difference in urinary continence between patients with rectoperineal fistulas and control patients however they did find an increased risk of urinary tract infections and requirement for anti-cholinergic medications.¹⁸ This contrasts with more severe malformations; in a series of 35 rectobladder neck fistulas assessed for urinary continence over the age of 4 years only 15% were continent, a further 43% were able to gain continence with Clean Intermittent Catheterization (CIC). Similar results are found in cloaca patients.¹⁹

Many complex ARMs require further urologic reconstructive surgery to achieve continence. Unlike patients with spina bifida, ARM patients are usually sensate and therefore introducing CIC in the older patients can be difficult. Boemers et al recommended that those at high risk of needing CIC would benefit from commencing it as neonates as they felt that children would then become accustomed to it.²⁰ For those that are unable to perform CIC per urethra then creation of a continent catheterizable channel (Mitrofanoff or Monti) is necessary. Often the appendix is used to create this channel and collaboration between the colorectal surgeon and urologist is vital to ensure sure that the appendix has not previously been used for creation of a Malone appendicostomy. With good pre-operative planning it is possible to split the appendix and perform simultaneous bowel and bladder reconstruction.^{21,22} These authors stress that the appendix should not be used for a Malone until the urologist involved can be confident that a catheterizable channel will not be necessary. As described previously the most common post-operative UDS finding

in cloaca patients is a floppy, atonic bladder and therefore bladder reconstruction may not be indicated.¹³ However, in patients with high pressure, neuropathic bladders then bladder augmentation can be crucial to preserve renal function. Again, with good collaboration, joint reconstructive operations can be performed and utilizing a pouch colon for coloplasty has been described.^{23,24}

For those children who have urinary incontinence due to detrusor overactivity, sacral nerve stimulation appears to be a promising therapy.²⁵ Sulkowski et al found that over 90% of patients were able to stop anti-cholinergic medications after placement of a sacral nerve stimulator.

Case 3

A 15 yrs old boy with a previously repaired rectoprostatic fistula attends for his routine follow-up appointment. The family travel from out of state because they like the multidisciplinary set-up in your center. For the last 6 months the boy has had 2 presentations with left sided testicular pain. At the first presentation he underwent a scrotal exploration which found an inflamed epididymis but no evidence of torsion. You take a full history and he denies being sexually active. He manages his bladder with intermittent catheterization per urethra and is dry between catheterizations. He is on long term anticholinergics for detrusor instability on prior urodynamics but admits that he hasn't been taking them regularly and has had occasional episodes of incontinence between cath. You organize a cystourethroscopy and urodynamics. At cystourethroscopy you note that the verumontanum sits above the bladder neck. Urodynamics also show that the intravesical pressure is high during periods of detrusor overactivity. After discussion with the boy and his family you decide to optimize his compliance with anticholinergics to see if reducing his episodes of high pressure will improve his episodes. He returns after 6 months of good compliance with anticholinergics and has not had any episodes of epididymo-orchitis. You advise him that compliance with his medication is likely the reason for this improvement but if symptoms return, an option would be to create a Mitrofanoff catheterizable channel as this may reduce episodes.

Testicular problems

Congenital urological problems which can impact on fertility are undescended testes and recurrent epididymo-orchitis. In a series of 136 males ARM patients Cortes et al found that nearly 20% had undescended testes and therefore part of the neonatal evaluation should include careful examination of the genitalia.²⁶ Trovalusci et al found an incidence of 33% of UDT in a series of 30 adult patients and half of these had bilateral UDT.²⁷ The mean testicular volume of these adult ARM patients was also significantly lower than controls. The AUA recommends orchiopexy between 6 to 18 months of age to try and preserve testicular function and reduce risk of testicular malignancy.²⁸ In patients with several congenital anomalies treatment for UDT may be delayed or overlooked. However, especially in bilateral UDT this may have a significant impact on fertility. Orchiopexy can be performed at the same time as other surgical procedures in an effort to minimize the number of general anesthetics a child receives.

Recurrent epididymo-orchitis is reported in up to 20% of male ARM patients and can lead to unnecessary scrotal explorations to rule out torsion.^{29,30} It is more common in rectobladder neck and rectoprostatic fistulas and associated with the presence of neurogenic bladder and urinary tract infections. In around a third of cases dividing the connection between the bowel and urinary tract will result in resolution.²⁹ However ectopic insertion of the ejaculatory ducts has been reported and in recurrent cases vasectomy can be considered as a treatment of last resort. Recently, there has been some reported success with endoscopic injection of the ejaculatory ducts at the verumontanum.^{31,32} Prevention of recurrence is important to try and minimize testicular damage.

Male Sexual Function and Fertility

Surveys of adult patients with ARMs have found that they tend to have a later sexual debut than the general population.^{27,33,34} The presence of persistent fecal or urinary incontinence into adulthood is known to affect their body self-image and psychosexual health.^{33,35} The development of the PSARP approach has led to a reduced incidence of urological trauma from operative repair and the incidence of problems with erectile dysfunction appear to be low.^{27,36}

Trovalusci et al reported sexual outcomes in 30 adult male ARM patients and found 1 patient with erectile dysfunction.²⁷ This series included a significant proportion with rectobladder neck fistulas (26.5%) and may not be representative of all ARM patients. A much more frequent finding was retrograde ejaculation, diagnosed by urinalysis, which affected 26.5% of patients. Of concern, they also reported a high rate of azoospermia/oligospermia affecting 13 of 28 (46%) patients. These issues are unlikely to be detected or diagnosed in a pediatric setting and therefore urological follow-up into adulthood is important. For patients who have retrograde ejaculation it may still be possible to father biological children with assisted reproductive technologies such as intracytoplasmic sperm injection. Improved long-term research is necessary in this field to better understand the impact on fertility.

Post-Surgical Urethral Complications

The innovation of the PSARP approach allowed identification of rectourethral fistulas from within the rectal lumen and led to a reduction of urethral injuries.³⁶ However, it is still possible to have urethral injuries or strictures following operative repair.³⁷ The involvement of urology in the multidisciplinary team allows for optimal management of these problems.

There is a delicate balance between not injuring the urethra by dissecting the fistula too close to the urethra and by leaving too much fistula behind. A posterior urethral diverticulum or remnant of the original fistula (ROOF) is a well reported post-operative complication and can be associated with urinary tract infections, post void dribbling, passing mucus per urethra and stone formation within the diverticulum.^{38,39} Cystourethroscopy and MRI pelvis may both be required to confirm the diagnosis and it is prudent to involve urological colleagues to excise the problematic ROOF.

The Role of a Urologist in a Collaborative Center

Urologists are important to ensure that renal and bladder function are appropriately investigated and managed during childhood. Collaboration ensures that joint surgical procedures can be performed when needed and the number of anesthetics can be minimized in complex patients.

Urologists are well placed to address sexual concerns as patients enter adulthood and can diagnose infertility issues which may not be recognized by pediatric surgeons.

Gynecology

Around 60% of girls born with cloaca have anomalies in their gynecological systems and this has been well recognized for many years.⁴⁰ Over recent decades research has highlighted that gynecological anomalies also occur frequently in rectoperineal and rectovestibular fistulas.⁴¹ However, in many centers where pediatric surgeons are working alone to look after girls with ARM, gynecological malformations may go undiagnosed until puberty. Even when Mullerian abnormalities are suspected, some may be difficult to diagnose with certainty prior to pubertal development as these structures remain small when unexposed to estrogen.

Case 4

A young baby with cloaca comes for an examination under anesthesia. As the gynecologist you are part of the surgical team and perform a vaginoscopy. This demonstrates a longitudinal vaginal septum, 2 cervixes and the rectal fistula entering high on the posterior vaginal wall. You are able to counsel the girl's parents that she has a uterus didelphus and recommend excising the septum as part of the cloacal reconstruction.

Neonatal care

The addition of a pediatric gynecologist during the initial evaluation, surgery or examination under anesthetic (EUA) allows for early assessment of gynecologic anatomy which will allow accurate counselling of the family as well as determine the type and timing of subsequent gynecologic interventions. Many parents appreciate frank and open discussions about their daughter's future sexual and reproductive health even while their child is still very young. The presence of a gynecologist in the team looking after girls with anorectal malformations raises the expectation that gynecological issues will be addressed and that concerns can be talked about freely.

Ultrasound evaluation of the internal Mullerian structures in the neonatal period can be a useful adjunct to surgical evaluation as estrogen production during the "mini-puberty" enlarges any uterine structures. Proactive initial surgical assessment at the time of EUA including vaginoscopy, if clinically indicated, helps to further elucidate anatomy including the vagina, number of cervixes, and presence or absence of a vaginal septum. Documentation of a normal vaginal introitus, vagina, and cervix allows an encouraging

discussion with the parents that their daughter will likely have a normal uterus and menstrual egress as well as the ability to engage in penetrative sexual intercourse.

The exam may reveal anomalies which may be missed if not evaluated by someone experienced in gynecology and there are case reports where a PSARP has been performed for a rectoperineal/rectovestibular fistula and vaginal agenesis has not been noticed by the operating surgeon.⁴² Failure to recognize a Mullerian anomaly such as vaginal agenesis at the time of primary operation may prevent the possibility of early repair of a menstrual outflow obstruction. The lack of early diagnosis of an outflow tract obstruction in patients with a uterus will result in pain and increased risk of endometriosis at the time of menarche which can have significant impact on fertility and quality of life. Furthermore, even in individuals in which the anomaly is not associated with an obstruction (such as absent uterus and vagina), their discovery later in life may further exacerbate the patient's negative self-perception, sense of loss and fear about other possible findings in the future.

In more complex cloacal malformations pre-operative vaginostomy assessing the gynecologic anatomy is crucial to successful pre-operative planning.⁴³ For example, a long common channel length is associated with a higher likelihood of requiring a vaginal replacement.⁴⁴ Alternatively, identifying a longitudinal vaginal septum and two cervixes during preoperative vaginostomy provides insight into the anticipated uterine anatomy as well as the opportunity to remove the septum at the time of the primary reconstruction.

Case 5

A 15 yrs old girl comes to your collaborative center for the first time for evaluation due to ongoing problems with fecal soiling. Her family are not clear what her initial diagnosis was. As the gynecologist you hold a consultation with her to discuss her menstrual and sexual health. She admits that she has not yet started her periods but that she has been suffering with cyclical, severe lower abdominal pain for the past year. She consents to an examination in the office with a chaperone. On inspection she only a small pit visible where the introitus would be expected to lie confirming distal vaginal agenesis.

You organize a pelvic and perineal ultrasound. She has demonstrable hematocolpos and hematemetria in what appears to an anatomically normal uterus. Both ovaries are seen and contain small cysts. The proximal vagina is around 2cm from the perineum. You explain the findings to the young woman and her family and offer the choice of surgery in the near future with the possibility of needing post operative vaginal dilations or suggest that if the young woman does not feel ready for this then you can give her hormonal suppression. The young woman opts for surgery and you are able to mobilize the vagina to reach the perineum without the use of skins flaps or mucosal grafts. She performs vaginal dilations at home and 6 months later has established regular periods and uses tampons.

Puberty

Puberty marks the time when the functionality of Mullerian structures can be definitively ascertained. A proactive approach including anticipatory guidance, pelvic ultrasound, and monitoring for painful menses to detect menstrual obstruction is recommended, especially in

girls where Mullerian anatomy was not documented or investigated at the time of anorectal repair or if there was any uncertainty in the congenital or acquired anatomy.

Puberty should occur at the usual time but can be delayed if there have been problems with nutrition and a girl is underweight. It is useful to discuss the physical changes which herald the onset of puberty with the patient and her parents. Rapid linear growth followed by thelarche, which is the onset of breast development, marks the beginning of puberty and typically occurs between the ages of 8 and 13 years. Girls require evaluation if there is no thelarche by age 13 year, no menarche by age 15 years, or if menses do not commence within 3 years of thelarche. This should be explained to the girl and her family and they should be alerted to the 'red flag' signs of amenorrhea, cyclical abdominal pain, and/or painful menses.

RED FLAG SIGNS

1. Primary amenorrhea has many potential etiologies, but in patients with ARM two important considerations include:
  - a. There is a lack of functioning endometrial tissue or uterine hypoplasia.
  - b. There is outflow tract obstruction – this may be congenital vaginal agenesis or a post-surgical problem such as vaginal stenosis. Cyclical abdominal pain associated with amenorrhea should prompt further investigations to look for outflow tract obstruction. An outflow tract obstruction can occur at any point in the reproductive tract.
2. Patients who are menstruating and experience severe dysmenorrhea also need to be evaluated for outflow tract obstruction. Patients who are menstruating but obstructed may have a duplicated Mullerian system with unilateral obstruction of a structure with functional endometrial tissue. This may either be:
  - a. Congenital e.g. obstructed horn in a bicornuate uterus
  - b. Post-surgical obstruction if not all Mullerian structures have been connected to the outflow tract. An example would be a cloaca patient with uterus didelphus and a neovagina where only one uterus has been connected to the neovagina.

Obtaining a screening pelvic ultrasound in asymptomatic patients six months after the start of thelarche allows for confirmation of the upper Mullerian anatomy. Screening ultrasounds may not be necessary for all rectoperineal or rectovestibular fistulas, if there has been a thorough gynecological assessment at the time of definitive repair which was normal. However, in centers with complex referral bases up to 20% of rectovestibular and 5% of rectoperineal fistulas have associated gynecological anomalies.⁴⁵ Even with a systematic approach to defining Mullerian anatomy at time of definitive repair nearly 40% of cloaca patients should be monitored for development of Mullerian obstruction at puberty.⁴⁶ Therefore, all patients who experience amenorrhea (with or without cyclic abdominal pain) or painful periods should have an diagnostic ultrasound to evaluate upper Mullerian anatomy. A subsequent pelvic MRI may be required to further characterize or

confirm abnormal ultrasound findings. Late diagnosis of obstructed menstruation is known to increase the risk of endometriosis, pelvic inflammatory disease and pyocolpos and should be avoided if possible.^{41,47} Furthermore, it is not unusual that patients who have undergone extensive surgeries in childhood develop ovarian and inclusion cysts with the advent of ovulation. An ultrasound may identify these and allow for prevention of enlargement through the use of hormonal suppression.

Obstructed Mullerian structures detected after puberty, are likely to require hormonal suppression in order to allow time for further discussion with the patient and her family to plan operative intervention. Vaginal surgeries to relieve obstructed menstruation require knowledge of the anatomy with special considerations to the presence and patency of the cervix. Repair of a vaginal septum may require post-operative vaginal dilation, in which case successful treatment is reliant on active involvement of the young woman. Alternatively, in cases where there is absent cervix or obstructed uterine horn, the best course of action may be removal of the Mullerian structures. Given the irreversible nature of such a procedure, it is important that the adolescent and family are allowed time to make a deliberate decision. Thus, hormonal suppression will provide symptomatic relief and prevent progression of endometriosis while allowing the adolescent to mature and shared decision making.

Menarche is also the time to start discussions with young women about assessment of the vaginal introitus and whether it is adequate to allow for tampon use and penetrative sexual intercourse. In older teenagers, an office visit with clinical examination will provide an opportunity to answer questions about anatomy, evaluate for pain and vaginismus, and access and educate about pelvic relaxation prior to vaginal dilator use if needed. It is not uncommon to identify longitudinal vaginal septums at this time. They do not necessarily obstruct menstrual egress but may interfere with tampon use, penetrative intercourse, or vaginal delivery. Resection of the septum can be performed as an outpatient procedure.

When vaginal stenosis is found the treatment depends on the location of the narrowing. In women whose reconstructive surgery has involved mobilization of their native vagina strictures can occur at the introitus and may respond to the serial dilation with vaginal dilators and pelvic floor physical therapy. If this is unsuccessful, then an introitoplasty may be required. Women who have had bowel neovaginas created as part of their reconstruction can suffer from strictures at the introitus as well as at the anastomosis between the bowel and native vagina. The internal stricture may require dilation under general anesthesia or further surgery to excise the stricture that might entail Z-plasties or buccal mucosa grafts.^{48,49} Patients who have had vaginal replacements have a high risk of requiring further surgical intervention. Couchman et al found that 57% of cloaca patients had at least 1 further operation to facilitate menstrual egress or sexual intercourse.⁴⁷

Sexual Functioning

Long term studies of sexual functioning in women with repaired anorectal malformations have found a high rate of sexual dysfunction in up to 50% of these individuals regardless of the type of anorectal malformation.⁵⁰ ARM patients tend to have a later sexual debut and

there is evidence that ongoing problems with urinary and fecal incontinence impact on their sexual health and body image.^{33,35,51}

This highlights the importance of collaboration between the entire team looking after ARM patients as fears around incontinence during sexual intercourse can have practical solutions. For example, in older patients who usually have a laxative regimen but are concerned about possible leakage or soiling during sexual intercourse, the option of performing an enema beforehand can be introduced by the colorectal surgeon. However, this may not be suggested if the woman feels uncomfortable discussing her sexual health with a pediatric or colorectal surgeon. van den Hondel surveyed adult ARM patients and found that many patients stated that sexual concerns had not been adequately addressed by medical practitioners.⁵⁰ This may reflect the fact that most ARM patients are managed solely by pediatric surgeons who may find it difficult to conceive that the baby with cloaca that they have looked after for many years is now a grown woman with sexual concerns. A recent survey of European pediatric surgeons found that half of the them did not feel confident discussing sexuality with their ARM patients and wished for more support and training in sexual health medicine.⁵²

The other crucial aspect is to ensure that the team can offer psychological support for patients regarding their anatomy, body image, and sexual function if struggling with anxiety or fear around expressing sexuality. Patients with dyspareunia or vaginismus can be helped with pelvic physiotherapy. Patients may not volunteer concerns unless specifically asked and may find it difficult to do so in front of their parents. When gynecologists are involved in a collaborative team, they optimize care through seeing patients over time, establishing rapport and offering private, confidential appointments without parents. We have found that at a first clinic appointment many patients will not feel comfortable discussing their sexual health however on subsequent appointments they may return with questions that lead to fruitful discussions and shared decision regarding further management.

Sexual Health and Contraception

Gynecologists and primary care physicians advise about safe sexual practices and the importance of the HPV vaccination which may not be addressed by pediatric surgeons. Women who do not have a functioning uterus may not realize that they will not require cervical cytology screening (Pap smear) but remain at risk of sexually transmitted infections and should be counselled accordingly.⁵³

Contraceptive options and advice will depend on the patient's overall health and uterine anatomy and a gynecologist who is comfortable with such complex cases, can consider all these factors in order to provide the best family planning for each patient. For example, although Intrauterine devices (IUD), a long acting reversible contraceptive, may provide some of the best pregnancy protection, they are contraindicated in patients with Mullerian anomalies. In patients with didelphic uteri, providers have reported placing IUD's in both horns as pregnancies have been reported when an IUD was placed in only one horn of a bicornuate uterus.⁵⁴ For those women with didelphic uteri there is very little evidence to guide the use of IUDs and it may be that other contraceptive options, including hormonal

contraceptives, are more appropriate. Alternatively, comorbidities associated with ARM, such as hypertension secondary to renal disease and some congenital heart abnormalities, may preclude the use of estrogen containing hormonal contraceptives.⁵⁵

Case 6

A 25 yrs old woman with a history of a previously repaired cloaca presents 8 weeks pregnant with her first pregnancy to your antenatal clinic. Unfortunately you do not have her previous medical records however she is not aware that she required a bowel vaginoplasty as part of her cloacal reconstruction. Her ultrasound scan shows a singleton pregnancy in what appears to be a bicornuate uterus. She is fully continent of feces and would like to opt for a Caesarean delivery to avoid the risk of a vaginal tear that might impact on her anal sphincter. She knows that she has a single kidney and she had a VSD repaired as a baby but neither she or her family know whether she had any screening for spinal anomalies as a child. In view of her VACTERL association you organize a spinal MRI to help determine whether she is suitable to have epidural anesthesia. This is reported as normal. She presents in labor at 35 weeks' gestation and is able to have a Caesarean section with epidural anesthesia and gives birth to healthy baby boy.

Pregnancy

There is a lack of long-term fertility outcomes in women with anorectal malformations and evidence is restricted to small case series.⁵⁶ There is no data about intent for conception, spontaneous conception rates, length of time until conception, or need for infertility treatment in women with ARM. A systematic review of fertility in ARM patients found 9 studies that reported the childbirth rate which was defined as the number of patients with children of their own divided by the total number of patients.⁵⁷ The mean age of the patients in these studies varied between 23 and 35 years. The overall childbirth rate was 27% (range 0-57%) in 429 patients however it included both male and female patients. There were only 2 studies that compared childbirth rate in ARM patients with healthy controls, both were from the same center and published in the 1990s.^{58,59} These two studies suggest the complexity of the ARM may impact childbirth rates as patients with rectoperineal and rectovestibular fistula had similar rates as healthy controls (39%) but patients with cloacas had a much lower rate (14%).

Women who have Mullerian agenesis associated with ARM almost universally have normal ovaries and therefore have the possibility of having biological children through surrogacy.⁶⁰ There are also increasing numbers of successful uterine transplant in patients with Mayer-Rokitansky-Kuster-Hauser syndrome resulting in live births but these have not been performed to date in women with ARMs.⁶¹

There are numerous considerations in women with ARM who become pregnant related to uterine anatomy, chronic UTIs and renal disease, VACTERL associated spine and cardiac conditions, and risks of both vaginal and surgical delivery. Women with cloacal malformations have a 50% risk of chronic kidney disease which may be unmasked by the higher metabolic demands of pregnancy and increased risk of urinary tract infections.^{6,62} Congenital heart disease increases the complexity of the antenatal care

requiring multidisciplinary care including cardiology and experts in high risk pregnancy. Older patients may not have had systematic screening for VACTERL association and may benefit from a spine MRI during pregnancy and an antenatal consultation with anesthesiology to discuss whether they are candidates for regional anesthesia for pain management or delivery. When a low lying conus has been diagnosed on MRI then successful epidural anesthesia can be performed using ultrasound guidance.⁶³

Mode of delivery should be individually tailored to the woman's original malformation, previous surgery and fecal continence status. In women who have had bowel vaginoplasties a planned Caesarean section is imperative. It is important to note that these patients may have significant intraabdominal scarring and altered anatomy which may require help from an experienced surgeon. In women who have had lower urinary tract reconstruction, such as bladder augmentation or creation of a catheterizable channel (Mitrofanoff), consideration should be made about the availability of urological help during pregnancy and delivery.⁶⁴

The delivery planning for women with native vaginas is more nuanced. Previous surgery may increase the likelihood of vaginal tears and disruption of the perineal body in scarred tissue planes. Ultrasonographic studies of pelvic musculature in women with previously repaired ARM have shown that all women had at least one muscular defect.⁶⁵ If the woman has full fecal continence then any risk of developing incontinence may be unacceptable to her and she should be offered a Caesarean section. However, for women who are reliant on an enema regimen for fecal continence it can be argued that a vaginal delivery will not change their continence status. There are several reports of successful vaginal deliveries even in the context of previous cloacal repair.^{56,66}

Concerns Specific to Bowel Vaginoplasties

Bleeding outside of menstruation in bowel vaginoplasties should be thoroughly investigated. Sometimes trauma from sexual intercourse can cause mucosal irritation or even small lacerations. These may resolve with abstaining from sexual intercourse however persistent bleeding should prompt vaginoscopy and a thorough examination of the entire mucosal surface.

Diversion colitis is not uncommon-a recent endoscopic study of 34 patients with sigmoid colon neovaginas found 65% had histological evidence of inflammation.⁶⁷ Complaints of excess mucus production and malodor correlated with the degree of inflammation. The inflammation can be treated with sodium butyrate vaginal enemas or 5-aminosalicylic acid pessaries.⁴⁹

There are also reports of inflammatory bowel disease developing in neovaginas which can be difficult to manage and may require vaginectomy.^{68,69}

Worryingly, there are case reports of malignancy in the long term follow-up of bowel neovaginas so any suspicious mucosal lesions should be biopsied.⁷⁰⁻⁷²

Long Term Gynecologic Health and Pelvic Organ Prolapse

As women with ARM age, longer term gynecological outcomes merit investigation. Currently, there is no data available concerning life-long risk of pelvic organ prolapse and timing of menopause in women with ARM. While the ovaries are typically not impacted in women with ARM, extensive pelvic surgery could potentially impact blood supply and accelerate menopause. Early menopause is associated with significant morbidity in women including cardiovascular disease and adverse impact on bone health. Given lack of menses in some patients born with ARM, early menopause may not be noted. There is no guidance regarding routine blood screening for menopause but symptoms such as hot flashes should be evaluated.

While there are anecdotal reports of bowel neovagina prolapse even relatively soon after primary reconstruction, evidence is lacking regarding the life-long risk of pelvic organ prolapse. Extensive pelvic surgery, neurologic abnormalities, and chronic constipation could all theoretically increase risk. There is uncertainty regarding the effectiveness as well as best time to initiate pelvic floor physical therapy to prevent or treat prolapse in women with ARM.

The Role of the Gynecologist in a Collaborative Center

Gynecologists offer a unique perspective to management of girls with anorectal malformations. Benefits to patients include earlier recognition of gynecological anomalies and potentially a reduction in the number of operative procedures.⁷³ When involved early they are able to counsel and educate parents about likely fertility outcomes or options for their daughter and manage expectations for further treatment or surgery. Furthermore, as the patient transitions to adult care, the gynecologist can help to address issues of sexuality, sexual function, family planning and other routine gynecologic care.

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