2 Surgical Techniques for Vaginal Agenesis With and Without a Functioning Uterus

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Müllerian Anomalies

Background and Terminology

Anomalies of the female urogenital tract vary in structure, etiology, location, and presentation. Müllerian anomalies are defects caused by malformations or dysfunction in Müllerian duct development that occurs during female embryogenesis. Anomalies may have complete underdevelopment of the Müllerian duct system with agenesis or atresia of the vagina, uterus, and/or fallopian tubes. These disorders are referred to as Müllerian agenesis, Müllerian aplasia, Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome, and vaginal agenesis [1]. The incidence of Müllerian or vaginal agenesis is 1 per 4500–5000 females [1], whereas the exact prevalence of all Müllerian anomalies is unknown. Defects in the female reproductive tract development are estimated in 7% of healthy reproductive-aged women [2]. Many abnormalities of the female reproductive tract are likely undiagnosed, but may be seen more frequently in women with miscarriages or infertility [3].

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The presentation of a Müllerian anomaly depends on the stage of embryogenesis dysfunction that occurs, the location and structure of the defect, and the presence or absence of obstruction. Obstruction refers to outflow blockage with backup of fluids, especially mucus or menstrual blood, when a uterus or uterine structure with a functioning endometrium is present. This occurs in patients with transverse vaginal septum, distal vaginal atresia, imperforate hymen, or obstructed uterine horn(s). Obstruction does not occur in patients with Müllerian agenesis or MRKH as they lack a uterus unless incomplete Müllerian duct development results in an isolated uterine horn. This is seen in patients with OHVIRA (Obstructed Hemivagina, Ipsilateral Renal Anomaly), or Herlyn-Werner-Wunderlich syndrome, who have complete duplication of the Müllerian duct system comprising of a uterine didelphys and two vaginas. In these patients, one vagina is not patent causing outflow obstruction on that uterine side and a renal anomaly is found on the ipsilateral side.

Terminology regarding Müllerian anomalies is often confusing as multiple names for each diagnosis may be used interchangeably. There is clinical distinction between patients with vaginal agenesis (Müllerian agenesis) who lack a developed uterus, and those with an obstructive Müllerian anomaly such as distal vaginal atresia, or vaginal agenesis involving a normal uterus. Multiple classification systems exist to attempt to

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categorize the wide variety of these reproductive tract defects and are described in the following sections. The focus of this chapter will be concerning Müllerian anomalies involving abnormal vaginal development. There is distinction between clinical recommendations for patients with Müllerian or vaginal agenesis who lack a uterus, and those with obstructive Müllerian anomalies or vaginal agenesis and a functioning uterus (distal vaginal atresia). Each type of Müllerian anomaly is unique, and patient management should always be individualized.

Embryology

Development of the female reproductive structures begins at 6–7 weeks of gestation and is guided by the presence or absence of the SRY (sex-determining region Y) gene [4]. At approximately 37 days of fertilization, the Müllerian ducts appear lateral to the Wolffian duct system. In the absence of the SRY gene, the Müllerian structures fuse in the midline, and canalize until 14 weeks to caudally join the sinovaginal bulb, or vaginal plate. The Müllerian ducts ultimately develop into the fallopian tube, uterine cavity, cervix, and upper one-third of the vagina. Distal to the vaginal plate, the urogenital sinus, which is derived from the fetal cloaca, fuses with the Müllerian structures and canalizes to form the lower two-thirds of the vagina and the hymen [2, 4]. Defects in female reproductive tract development can occur at any point in fetal development. Failed vertical fusion of the Müllerian duct with the sinovaginal bulb may result in cervical atresia, a transverse vaginal septum, or distal vaginal agenesis. Underdevelopment and/or incomplete canalization of the upper Müllerian duct structures can cause structural uterine anomalies such as a uterine septa, a bicornuate uterus, or a longitudinal vaginal septum.

Classification Systems

There are several different classification systems for Müllerian anomalies. The most widely used system in the United States is proposed by ASRM (The American Society for Reproductive Medicine) and is based on the uterine structure (Fig. 2.1). In Europe, the most widely used classification terminology, also based on uterine shape, is from The European Society of Human Reproduction & The European Society for Gynaecological Endoscopy (ESHRE-ESGE) [5]. Another classification system that may be clinically useful, called the Acién Classification of Genital Tract Anomalies, is based on the type of embryologic dysfunction that occurs during different phases of female urogenital development [5]. The VCUAM (vagina cervix uterus adnex-associated malformation) Classification was developed to simplify the grouping while remaining precise [6]. There are downsides to all of the classification systems available, and most are difficult to apply in routine gynecologic practice. These limitations may include the lack of genetic, syndromic, or ovarian considerations. In addition, most of the existing classification systems may exclude those with hybrid or very rare anomalies. Disorders of the female urogenital tract are diverse and difficult to characterize in a precise way; they exist on a spectrum of structural and developmental issues that occur in embryogenesis. ASRM has published a more comprehensive classification system, ASRM MAC 2021, which describes anomalies involving the uterus, cervix and vagina for clinical application. The focus of this chapter will be regarding ASRM Class 1 Müllerian anomalies from the 1988 classification system and the mullerian agenesis category from the 2021 tool.

Vaginal Agenesis With and Without a Functional Uterus

Differential Diagnosis

A patient presenting with primary amenorrhea and evidence of vaginal or Müllerian agenesis should be evaluated for other reproductive tract anomalies that appear clinically similar and can be misdiagnosed. The differential diagnosis includes imperforate hymen, transverse vaginal septum, androgen insensitivity syndrome, and Swyer's syndrome. In patients with MRKH

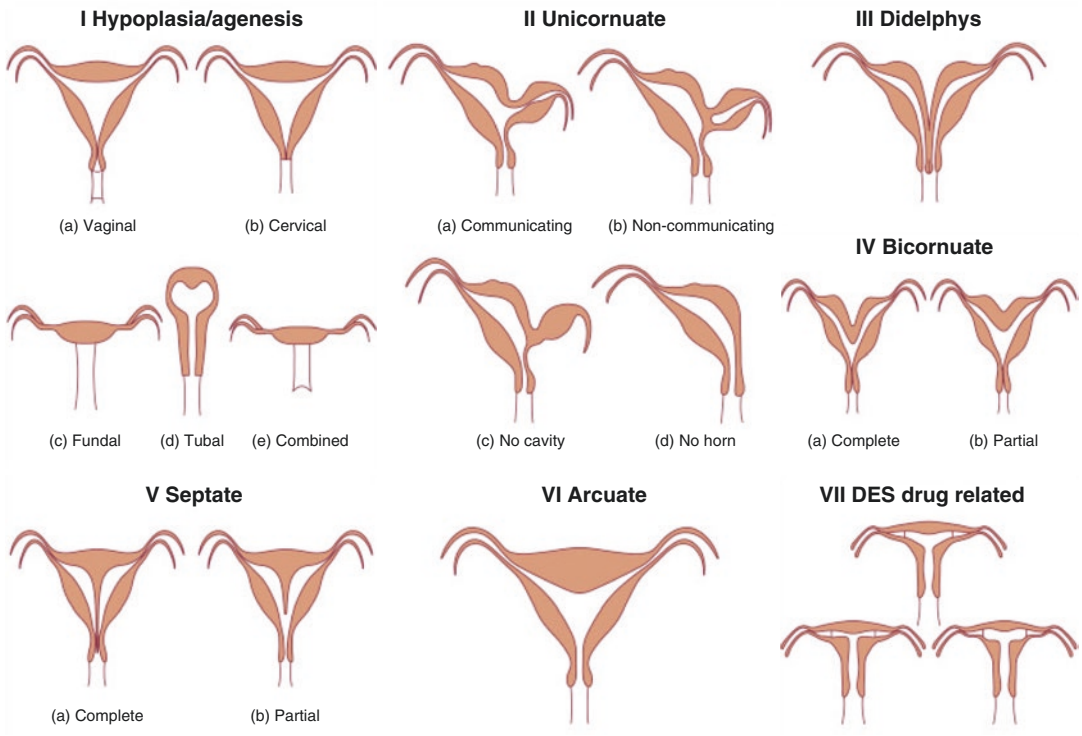

Fig. 2.1 Classification of Müllerian anomalies from The American Fertility Society. (Reprinted from Rackow et al. [67]. Copyright (2017), Elsevier publishing)

(Müllerian agenesis) and partial androgen insensitivity syndrome, physical exam will show normal secondary sexual characteristics, such as full breast development, pubic/axillary hair, and absent vagina. Patient's with Swyer's syndrome present with delayed puberty, absent breast tissue, and a normal functional vagina and uterus. An imperforate hymen and transverse septum may also present with obstruction, primary amenorrhea, and worsening cyclic abdominal pain similar to vaginal agenesis with a functioning uterus (distal vaginal atresia). Prior to puberty, a young girl with labial adhesions seen on physical exam can be mistaken for vaginal or Müllerian agenesis.

Evaluation and Diagnosis

Patients with any type of Müllerian anomaly present at variable points in life depending on the severity of the defect and presence or absence of outflow obstruction. Identifying Müllerian anomalies on a routine pediatric exam is uncommon and diagnosis is often delayed. Neonates with an imperforate hymen or obstructive Müllerian anomaly, such as a transverse vaginal septum or distal vaginal atresia, may present with a protruding vaginal mass caused by obstructed mucus (mucocolpos). These reproductive tract anomalies are ideally diagnosed prior to puberty in order to prevent the anticipated blockage, back-up of menstrual products, and pelvic pain by educating the patient and parents and in some cases surgical correction. Unfortunately, most are diagnosed after puberty with symptoms of an acute outflow obstruction. These patients present with absent menses, abdominal discomfort and/or a pelvic mass caused by painful build-up of menstrual blood in the upper genital tract (hematocolpos). For patients with vaginal agenesis, the diagnosis is often missed until late adolescence as patients are asymptomatic and have normal growth and development.

A thorough physical exam in the office is obligatory for any patient diagnosed with a

Müllerian anomaly. This exam should include an evaluation of the external genitalia and assessing the patient for secondary sex characteristics of breast development and pubic and axillary hair growth. Performing an exam under anesthesia may be necessary if a patient does not tolerate an exam in the office, and imaging is inconclusive. In this operative setting, complete visualization of the anatomy may include vaginoscopy and/or diagnostic laparoscopy especially for complex anomalies and if a vagina with incomplete Müllerian development is present. Müllerian anomalies are frequently associated with renal, vertebral, anorectal, cardiac, tracheoesophageal and limb anomalies, and assessment of defects involving these organ systems during a patient's evaluation should be considered [7].

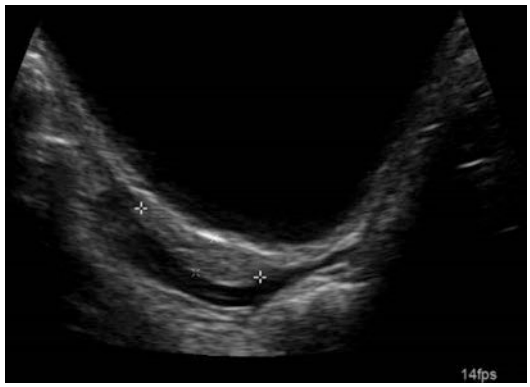

Fig. 2.2 Pelvic ultrasound of a Müllerian anomaly. Example of a pelvic ultrasound showing a hypoplastic uterine structure in Müllerian anomaly work-up of a patient with an absent vagina

Imaging is helpful with both obstructive and nonobstructive Müllerian anomalies; a pelvic ultrasound should be performed early in the evaluation. A pelvic ultrasound assesses uterine structures, identifies masses or hematocolpos, and evaluates the adnexa (Figs. 2.2 and 2.3a, b). Ultrasound is beneficial as the initial imaging modality of choice as it is minimally invasive, low cost, and readily available. However, a normal ultrasound does not rule out the presence of a genital tract anomaly. Magnetic resonance imaging (MRI) is more useful in visualizing urogenital anatomy by looking more closely at soft tissue structures in close proximity (Fig. 2.4). An MRI study can be especially useful in girls with Müllerian agenesis and coexisting urogenital or colorectal anomalies. However, this should not be the initial diagnostic imaging of choice given its high cost, frequent challenges obtaining the study on young children, and overall good reliability of images obtained with ultrasound instead.

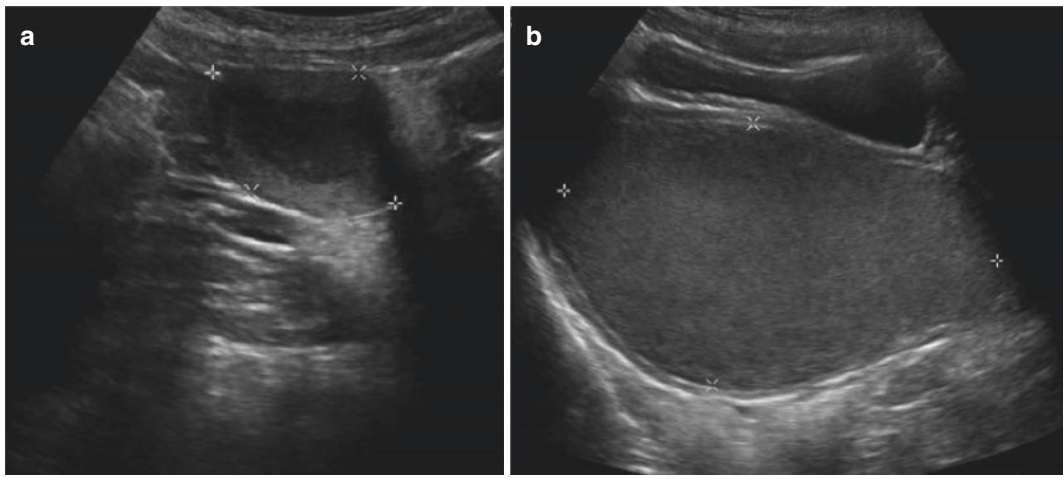

Fig. 2.3 (**a**, **b**) Pelvic ultrasound of an obstructive Müllerian anomaly. Diagnostic ultrasound in a patient with primary amenorrhea and cyclic pelvic pain showing hematometra (**a**) and large distended hematocolpos (**b**). There are normally developed upper reproductive tract structures, including a uterus and ovaries, but an absent distal vagina (distal vaginal atresia)

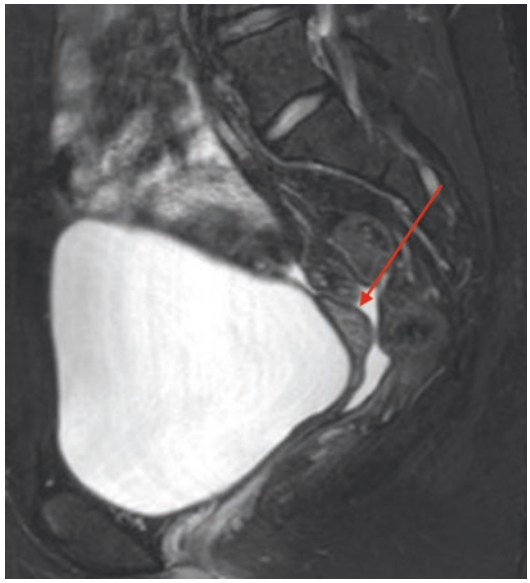

Fig. 2.4 Pelvic MRI of a Müllerian anomaly. T2-weighted magnetic resonance imaging (MRI) of the patient from Fig. 2.2. The hypoplastic uterine structure (red arrow) posterior to the distended bladder does not show an endometrial stripe and there is no vaginal structure

In order to provide appropriate clinical counseling, the complete evaluation (including history, physical, and imaging) should precede disclosure of the final diagnosis. Receiving the diagnosis of a Müllerian anomaly, especially vaginal agenesis, can be emotional with psychosocial implications for patients and families, especially regarding vaginal dilation or vaginal surgery at a young age. Concerns over future reproductive capabilities and sexual functioning are common and deserve adequate counseling throughout care. There should be multiple visits with a multidisciplinary team, including pediatric gynecology or urology, psychology, and reproductive endocrinology, to carefully review the patient's anatomy, management options, and address any emotional or psychologic problems in the process.

Reproductive Considerations

Women with Müllerian tract defects containing a functioning uterine structure can be reassured of favorable reproductive outcomes and fertility. In obstructive anomalies, patients can have normal sexual activity and child-bearing capabilities with surgical correction of the anatomic blockage. In patients with distal vaginal atresia, there is a normal upper vagina, cervix, and uterus. These patients and those with vaginal agenesis involving a cervix and functioning uterus have reproductive success following surgical correction by pull-through vaginoplasty or a similar procedure. These surgical procedures are described in the following sections.

Almost all patients with Müllerian anomalies have normal functioning ovaries as the development of the gonads is separate in embryogenesis. For patients with vaginal agenesis involving uterine atresia, artificial reproductive technology (ART) by oocyte harvesting and in-vitro fertilization can produce biologic offspring with the use of gestational surrogacy [8, 9]. Patients with disorders that lack functioning ovaries, such as Turner syndrome, Gonadal dysgenesis, or androgen insensitivity syndrome, may present similar to those with Müllerian anomalies or Müllerian agenesis; however, counseling and management in these patients are different and will not be discussed here.

In women who cannot carry a pregnancy and are opposed to gestational surrogacy, information should be provided regarding available reproductive options such as adoption or uterine transplantation. The first live birth in a patient with MRKH syndrome after uterine transplantation was reported in 2014 in Gothenburg, Sweden. Since then, there has been rising clinical interest and several US centers have conducted uterine transplantation with successful results [10, 11]. In a survey of women diagnosed with Mayer-Rokitansky-Küster-Hauser (MRKH) syndrome by Chmel et al., 62% expressed interest in uterine transplantation at the time of Vecchietti neovagina creation after being counseled on the risks, benefits, and lack of long-term data on the experimental procedure [12]. There is some debate regarding the ethics with living donors and the recipient patient's risk-to-benefit of elective uterine transplantation as it is a complex, lengthy procedure, and requires postoperative

immunosuppression. Reported data on adverse outcomes after transplantation is limited, but few cases of surgical complications include (but are not limited to): graft-vs-host disease, thromboembolisms, hemorrhage, pelvic infections/abscess, graft ischemia, emergency hysterectomy (transplant removal), fistula, cystitis, cuff dehiscence, and vaginal stenosis. However, most (more than half) of uterine transplant procedures performed internationally did not have any reported surgical or medical complications in the postoperative period. This innovative procedure, although complex, will likely continue to increase in success as a surgical option for select patients desiring fertility and pregnancy who have uterine absence.

Vaginal Agenesis with a Functioning Uterus (Distal Vaginal Atresia)

Presentation and Management

Unless diagnosed earlier in childhood, variants of vaginal agenesis with a functioning uterus, also referred to as distal vaginal atresia, present with pain and outflow obstruction at menarche. These anomalies, as well as transverse vaginal septum or imperforate hymen, are considered obstructive Müllerian anomalies and are treated similarly. Management of complex obstructive Müllerian anomalies, such as noncommunicating uterine horns or OHVIRA syndrome, is not discussed here.

Vaginal agenesis with a functioning uterus, imperforate hymen, or transverse vaginal septum will ultimately require surgery to allow for normal female reproductive function and spontaneous passage of menstrual blood. If one of these obstructive anomalies is identified before menarche, surgical management can be performed at an appropriate time to prevent obstruction at puberty. If not diagnosed before spontaneous menses begin, these patients will present with acute obstructive symptoms including abdominal pain, amenorrhea, and a pelvic mass or vaginal bulge caused by a hematocolpos (Fig. 2.5).

Fig. 2.5 Acute obstruction with bulging hematocolpos. Vaginal bulge from obstructed menses (hematocolpos) in an adolescent with an obstructive Müllerian anomaly. Differential diagnosis includes vaginal agenesis with a functioning uterus (distal vaginal atresia), transverse vaginal septum, and imperforate hymen

In the setting of an acute obstruction at the time of diagnosis, management options include either menstrual suppression to delay surgery or immediate surgical decompression. Delay in surgical management can be considered if pain is well-controlled, the patient is uninfected, and is able to void without issues. This strategy is preferred for young girls who may not be appropriately mature for surgery and possible postoperative vaginal dilator therapy if required. Pursuing immediate surgical management and decompression of an outflow obstruction is often necessary if the patient has uncontrolled pain, voiding difficulty, or evidence of infected hematocolpos.

The surgical procedure performed on patients with vaginal agenesis and a functioning uterus (distal vaginal atresia) is referred to as the pull-through vaginoplasty. Prior to surgery, the diagnosis of a suspected obstructive Müllerian anomaly should be confirmed, and the patient's urogenital anatomy assessed by an external genital exam and imaging. If able, the distance between the leading edge of the upper vagina and

the top of the vaginal dimple or lower vagina should be estimated preoperatively. In surgical planning, if there is a large distance between the proximal and distal vaginal ends, vaginal dilation, or use of an interposition graft may be necessary. The guidelines for management for these larger vaginal agenesis or septal defects are described below.

In performing the vaginal pull-through procedure, the visible apex of the lower vagina, or vaginal dimple is incised transversely and sharp dissection is carried toward the upper vagina that may be bulging from acute obstruction. A large hematocolpos can aid in this procedure by pushing the leading edge of upper vagina closer to the introitus and decreasing the thickness of the tissue being incised. Once old menstrual blood products, if present, have been adequately drained at entry into the upper vagina, the initial incision is extended laterally. For adequate diameter of the vaginal opening and to prevent stricture formation, additional small cuts are made at the superolateral and inferolateral aspects of both ends of the transverse incision (at the 1 o'clock, 5 o'clock, 7 o'clock, and 11 o'clock positions respectively). The upper vaginal mucosa should be tagged or grasped as soon as possible in the procedure and ultimately sutured circumferentially to the level of the lower vagina or introitus (Fig. 2.6).

Surgical management of transverse vaginal septum or an imperforate hymen is similar to that of distal vaginal atresia. Excision of a transverse vaginal septum involves surgical removal of the intervening septal tissue and anastomosis of the proximal and distal vagina. This is sometimes accomplished with a Z-plasty technique, involving creation of vaginal flaps sutured superiorly and inferiorly along the vaginal canal in order to increase vaginal length and minimize the risk of stenosis [1].

Larger transverse septal or vaginal atretic defects are those with more than 3–4 cm between the leading edges of the upper and lower vagina. These anomalies are managed differently as they have higher risk for stricture or stenosis. To decrease the risk in these patients, preoperative dilation of the lower vagina to approximate the upper and lower vagina and/or the use of a mucosal skin bridge (such as a buccal graft) at surgery may

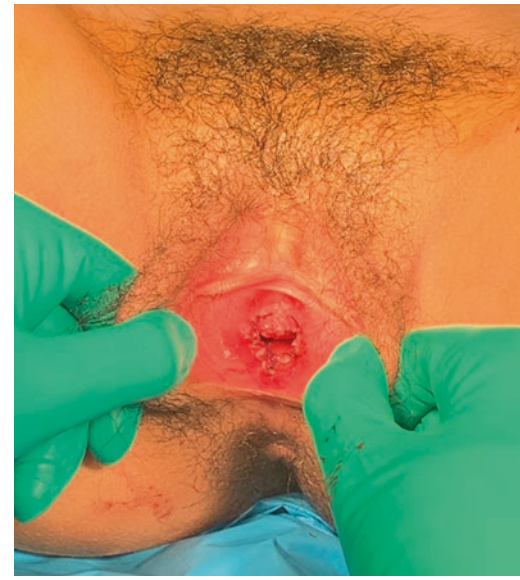

Fig. 2.6 Surgical correction of distal vaginal atresia by pull-through vaginoplasty. Vaginal canal after the pull-through vaginoplasty technique described in this chapter

be useful [7, 13, 14]. Postoperative use of soft or hard dilators to prevent strictures is often recommended after pull-through vaginoplasty, but has not been shown to benefit patients with defects measuring less than 3 cm. In those that vaginal dilator use after surgery is recommended, patients should perform vaginal dilation daily using hard dilators for 10- to 30-minute intervals. Alternatively, soft flexible vaginal dilators can be inserted and remain in place 24 hours-a-day for a week at-a-time with removal only to void. Patients with grafts placed surgically should not initiate dilator therapy until a follow-up exam confirms adequate graft healing and update though will likely need a vaginal mold to help prevent stenosis [1].

Risks and complications of surgical management of distal vaginal atresia, imperforate hymen, transverse vaginal septum, or vaginal agenesis with a functioning uterus include infection, vaginal scar tissue formation, vaginal stenosis or stricture, dyspareunia and need for repeat surgery [15]. Repeat surgeries to manage recurrent obstructions from postoperative vaginal stenosis have increased morbidity. Each subsequent surgery is more challenging with increasing risk of recurrent stenosis and treatment failure.

Müllerian Agenesis (Vaginal Agenesis Without a Functioning Uterus)

A patient with Müllerian or vaginal agenesis without a uterus should be counseled on the management options including primary vaginal dilation, surgical creation of a vagina (neovagina), or no treatment. The purpose of pursuing neovagina creation is for patient psychological wellness and sexual satisfaction. Primary vaginal dilation is considered the mainstay of treatment, but multiple surgical procedures are available to create a vagina if this fails. Referral to a tertiary center with a qualified multidisciplinary team should be considered, especially for girls with complex urogenital, cloacal, or colorectal anomalies, as they may have more favorable outcomes for the management of vaginal agenesis [15].

Nonsurgical Management (Primary Vaginal Dilation)

Vaginal elongation by using dilators is considered the first-line treatment of MRKH, vaginal agenesis, or Müllerian agenesis [16]. Important to the success of dilation is beginning when the patient is ready and committed to spending the time needed to perform dilation. Advantages of nonsurgical elongation of the vagina include the ability for the patient to proceed at her speed, no need for anesthesia, no hospitalization or scarring, and less risk of vaginal stenosis, pain, and expense compared with surgical management [17]. Disadvantages include longer time until successful creation of a neovagina and the need for patient privacy. Possible risks of using dilators to create or elongate the vagina include discomfort during dilation, bleeding from abrasions, and inadvertent dilation of the urethra instead of the vagina. It is necessary to have frequent follow-up visits with a healthcare provider comfortable with counseling and instruction of vaginal dilator use.

Graduated hard dilators come in a wide variety of sizes (Fig. 2.7). A knowledgeable medical professional should be able to select appropriate starting vaginal dilator size and the rate of advancing the diameter used during the dilation process. The usual process in selecting size is to start with the smallest size that is comfortable for the patient but provides appropriate soft tissue pressure and then advance up. Teaching adolescent and adult women how to use the dilators requires educating the patient about their own

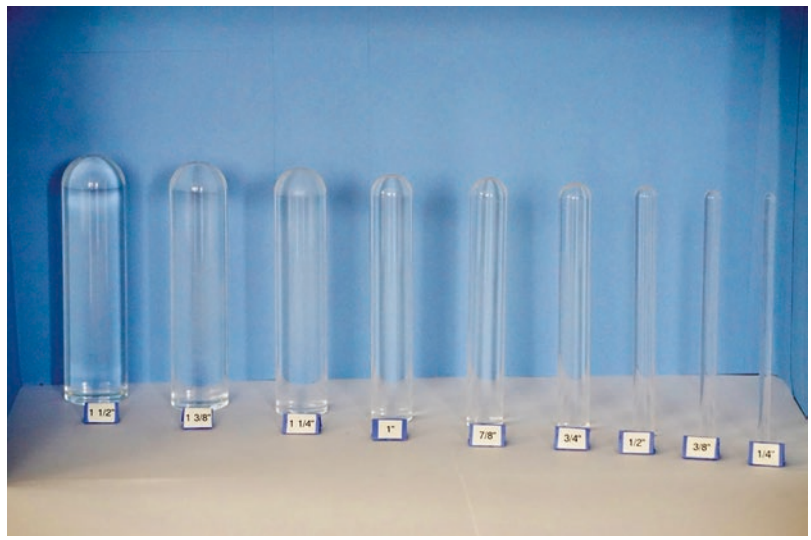

Fig. 2.7 Graduated hard vaginal dilators. Graduated plexiglass vaginal dilators ranging from ¼ inch to 1½ inch. If desired and to assist with dilation using a stationary seat, the length of the dilator may be modified by a carpenter or professional who is comfortable with cutting that specific material qualified company

external anatomy. The use of a mirror and frequent office visits are helpful in the success of patient's vaginal dilator therapy. Historically, vaginal dilation used hard dilators cut down to slightly extend beyond the patient's introitus when placed in the vaginal space and was held in place with spandex underwear [18, 19]. Patients were then instructed to sit on a bicycle seat attached to a firm board in order to apply pressure on the dilator at the perineum in order to perform dilation. Now, patients often find it more comfortable to lie down with hips flexed, retract the labia with their nondominant hand, and hold the lubricated dilator using the dominant hand at the perineum pointing downward toward the sacrum to create pressure on the dilator (Fig. 2.8). Ideally the patient uses the dilator for 10–30 minutes one to three times a day and gradually advances to a larger sized vaginal dilator when appropriate. It is recommended patients empty their bladder before and after dilation sessions. Patients who need to stop or interrupt dilation therapy during the graduated dilator process may need to use a smaller size dilator than previously used when they start-up again.

One study of patients with vaginal agenesis reported 80–92% success in creating a functional vagina using graduated hard dilators [20]. Adequate vaginal length, typically 6–7 cm, is attained once the patient can comfortably have vaginal intercourse. Vaginal dilation can also be achieved by penetrative sexual activity exclusively, or in combination with hard dilators, which has similar success rates [21]. Attention should be given to each patient's self-esteem and mood during the treatment process. Often, patients feel depressed over the need to use dilators, and are especially distressed if this treatment fails.

Surgical Management

Multiple surgical techniques to create a functional vagina (neovagina) in patients with vaginal agenesis have been performed since the early twentieth century. These techniques are secondary to vaginal dilation in management, and are often reserved for those who have failed or are unable to perform dilation [22]. The advantage of surgery over primary dilation is more rapid creation of a functional vagina. No singular technique has been found to be superior in terms of outcomes and patient satisfaction [3]. Each surgery has unique requirements, complications, and outcomes (Table 2.1). Regardless of type of surgery, complication rates are significantly reduced if performed prepuberty (14%) as opposed to postpuberty (58%) [23].

All surgical neovagina methods require close follow-up with a minimum of annual exams for evaluation of sexually transmitted infections, vaginal strictures, and evaluation of rare malignancies that may be associated with certain neovagina epitheliums [1, 24]. Almost all procedures require the use of vaginal dilators or molds after

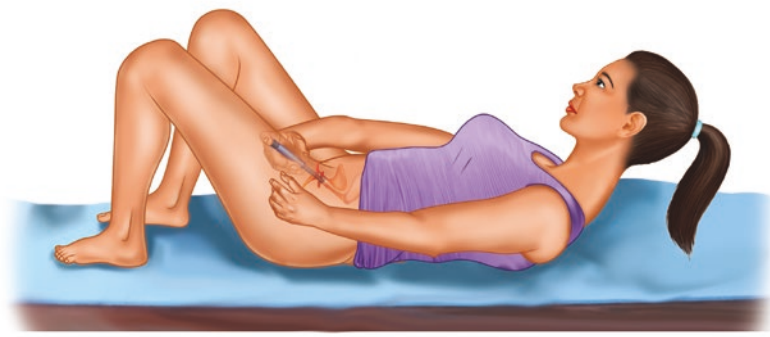

Fig. 2.8 Preferred patient positioning for vaginal dilation. The preferred patient position for vaginal dilation described in the text

Table 2.1 Summary of options for vaginal creation (neovagina) in patients with vaginal agenesis

Procedure	Description	Advantages & outcomes	Disadvantages & complications
Nonsurgical options			
Primary Vaginal Dilation	Patient performs vaginal dilation using graduated hard dilators or coitus *Considered first-line option*	Success in 80–92% Vaginal length 6.7–8.7 cm Avoidance of surgery and hospitalization Ability to perform at patient's preferred speed Decreased risk of stricture, stenosis, or dyspareunia Low-cost	Complication 5–35% Laborious and time consuming to patients Longer time to end result Emotional distress Need for routine privacy Discomfort during dilation Bleeding caused by abrasions Inadvertent urethral dilation Failure of therapy and requiring surgery
Surgical neovagina techniques			
Abbe-McIndoe Vaginoplasty, 1938 Or Modified McIndoe Procedures	Perineal dissection with split-thickness skin grafting performed in two surgeries Modified McIndoe techniques use of an alternative (mucosal-like) material grafted into the vaginal space Example: Human amnion Autologous buccal mucosa Surgical adhesive barrier Artificial created dermis	Success in 85% Vaginal length 7.4 cm	Complications 19–65% Prolonged hospitalization Pain Secondary surgery for vaginal mold removal Required postoperative dilator use Wound infection Neovagina stricture or stenosis Neovaginal fistula Graft failure Scarring at graft site Complications requiring reoperation
Davydov Procedure, 1974	Autologous pelvic peritoneum is surgically connected to the external vaginal opening	Success in 68–87% Vaginal length 7.8 cm High sexual satisfaction scores	Complications 14% Required postoperative dilator use Surrounding organ injury during procedure Neovaginal fistula Pelvic adhesions Granulation tissue formation Vaginal stricture, vaginismus Complications requiring reoperation
Vecchietti Procedure, 1965	Active perineal dilation by gradual increased tension of a surgically placed device over 1 week followed by a second surgery to remove the apparatus	Success in 80–98% Vaginal length 9.5 cm High sexual satisfaction scores	Complications 11–13% Prolonged hospitalization Secondary surgery for device removal Pain Required postoperative dilator use Urinary tract infection Granulation tissue formation Postoperative fever Vaginal stricture or stenosis Complications requiring reoperation

Table 2.1 (continued)

Procedure	Description	Advantages & outcomes	Disadvantages & complications
Intestinal Vaginoplasty, 1892	Surgical transposition of a closed loop segment of large or small bowel	Success in 73–83% Vaginal length 10 cm Option for surgery before puberty No requirement for postoperative dilation	Complications 7–79% Introital stenosis and subsequent need for dilation Trauma or discomfort with intercourse Excessive malodorous vaginal discharge Mucosal prolapse Small bowel obstruction Fistula formation Complications requiring reoperation
Vulvovaginal Pouch or Williams Procedure, 1964	Full-thickness skin graft and external labial suturing to create a vertical perineal pouch for intercourse	Success in 95% Vaginal length of 10–12 cm Minimally invasive reversible procedure High sexual satisfaction Low complication rates No requirement for postoperative dilation unless vagina is short	Complications unknown to 4% Immediate postoperative hospitalization Bleeding with intercourse Wound infection Hematoma Irritation from graft with hair-bearing skin Disfiguring scars Change in anatomic axis for intercourse Awkward angle of vaginal intercourse Scarring at graft sites Complications requiring reoperation

Complied from data in McIndoe et al. [25], Buss et al. [52], Herlin et al. [38], Højsgaard et al. [53], Klingele et al. [54], Davydov et al. [36], Willemsen et al. [55], Allen et al. [56], Giannesi et al. [57], Borruto et al. [41], Borruto et al. [42], Brucker et al. [43], Rall et al. [58], Baldwin et al. [46], Karateke et al. [47], Carrard et al. [48], Communal et al. [59], Hensle et al. [60], Nowier et al. [61], Burgu et al. [23], Parsons et al. [62], Williams et al. [49], Creatsas et al. [51], and Creatsas et al. [63]

surgery for an extended period of time. With vaginal molds, the device can be created from flexible foam material covered by a condom that the patient wears continuously with removal only to urinate or defecate. Alternatives include intermittent vaginal dilation with hard dilators and/or eventually regular intercourse. The appropriate length of time for postoperative vaginal dilation or mold placements is individualized to the type of surgery, risk of stenosis, patient goals for vaginal length, and if the patient plans to use coitus to maintain patency. Careful consideration should be taken in deciding candidates for a surgical neovagina procedure. The risks of surgery and the postoperative requirements, including dilation and close follow-up, should be disclosed to the patient before surgery. As maturity to perform these tasks is needed, the provider may consider delaying surgical neovagina surgery until the patient is more ready at an older age.

Techniques for Surgical Vaginal Construction (Neovagina)

Active Dissection of the Perineal Space

The Abbe-McIndoe or McIndoe Vaginoplasty was first described in 1938 [25]. This technique uses a split-thickness graft (0.018–0.022 in) from the buttock to line a surgically created neovaginal space in a two-step surgical technique for girls with vaginal agenesis [25, 26]. Similar to a pull-through procedure described earlier in this chapter, the top of the distal vagina, or vaginal dimple,

is incised transversely and midline dissection is carried inward at the sub-urethral level. Once adequate space is obtained, a sterile mold is created using expandable foam material within a condom, covered with affixed split-thickness skin graft, and placed in the surgically created vaginal space. The edges are then sewn to the introitus, and labia are sewn closed to keep the mold in place. The patient remains in the hospital for a week, on bedrest, with a foley catheter and placed on a stool softening bowel regimen before returning to the operating room for mold removal and graft assessment. After surgery, the patient must follow a meticulous regimen using vaginal dilators to maintain vaginal patency and prevent stricture or stenosis while the graft continues to heal. Initially, the patient keeps a mold or flexible dilator in place continuously for minimum of a month; removing only to urinate and defecate. This is followed by insertion of a flexible or hard dilator several times a day for 3–6 months. Eventually, the dilator is used only nightly until the patient can maintain vaginal patency with intercourse alone [26].

Modifications of the McIndoe technique with split-thickness skin grafts are frequently performed and have variable functional outcomes. These modified techniques involve alternative graft materials which are placed on the vaginal mold similar to the initial McIndoe procedure. Tissues that have been successfully used in modified McIndoe procedures include autologous buccal mucosal grafts [27, 28], human amnion [29–31], and artificial adhesion barriers with *Interceed* [32, 33]. Similar to the split-thickness skin graft, these different mucosa-like materials are affixed to the temporary vaginal mold. Postoperative use of vaginal dilators is essential for preventing the vaginal strictures commonly experienced with the classic McIndoe procedure. More recently, the use of an artificially-created dermis by medical recombinant fibroblast growth factor mucosa has been described [34]. Lastly, full-thickness skin grafts obtained by harvesting myocutaneous rectus abdominis skin, gracilis or pudendal fasciocutaneous flaps, or skin taken from the lower abdominal wall by pfannensteil-like excision are reported [35]. These skin grafts can be associated with large disfiguring scars [35]. Full-thickness graft techniques are considered a last surgical option for creation of a neovagina, and are difficult to graft into the surgically created vaginal space [35].

Autologous Pelvic Peritoneum

The Davydov method for surgical vagina creation was initially described in 1974, and has reported outcomes similar to the McIndoe techniques [36–38]. The initial portion of the procedure is identical to the perineal approach of the McIndoe procedure, but pelvic dissection is slightly deeper and directed toward the abdominal peritoneum of the Pouch of Douglas. Then, via laparoscopy, the peritoneum is advanced by "push-down" approach from the pelvis and brought to the level of the introitus [13, 37, 39, 40]. The abdominal portion of the peritoneum is then closed in a purse-string fashion [13, 37, 39, 40]. Complications of this approach can include vaginal stricture or stenosis, as well as potential bladder or ureteral injury, which may in time may lead to vesicovaginal fistula formation [37, 40].

Active Perineal Dilation

The Vecchietti-technique, performed since approximately 1965, utilizes a method of active tension on the perineum for creation of a neovagina, and is the preferred technique used at European centers [41, 42]. A plexiglass olive, or modified dilator, is attached to the vaginal introitus with permanent sutures and secured through the perineum. The sutures are run through the lower abdominal wall to a metal device that allows for increasing tension on the perineum by tightening the sutures externally to stretch the blind vaginal pouch until sufficient vaginal length is achieved [26, 41–44]. This surgery requires a week of hospitalization with parenteral anesthesia for pain, as well as a second short-interval surgery for removal of the tension device and olive when dilation is complete. After removal of the device, the patient is instructed to perform continued vaginal dilation with hard dilators to maintain this newly created space and length [13, 26, 41–44]. A more recent strategy to the Vecchietti is a laparoscopic-assisted balloon vag-

inoplasty that was introduced in 2007 [45]. The active portion of dilation at the perineum is instead accomplished by feeding a retropubic foley catheter to the introitus and slowly increasing tension on the opposite end of the foley [45].

Intestinal Vaginoplasty

A method often preferred by pediatric surgeons, bowel loop or sigmoid vaginoplasty, is one of the oldest procedures for surgical creation of a vagina. As opposed to the other neovagina methods, bowel vaginoplasty for creation of a vagina can be performed in infancy or childhood [13, 15]. First described in 1892, the procedure was made popular in the United States in the early 1900s, and both small and large bowels have been used [46]. Steps of the procedure include open or laparoscopic mobilization of the bowel to the introitus with end-to-end re-anastomosis to create a blind vaginal pouch [15, 46–48]. Blood supply of the utilized bowel segment is maintained during the procedure.

Labial Skin Flap or Vulvovaginal Pouch

The Williams vulvovaginoplasty is an alternative surgical option to consider in certain patients as the surgery is reversible, minimally invasive, and does not require entry into the pelvis. Good candidates for creation of an artificial vagina by the Williams technique may include those who have failed dilator therapy, are unable to perform vaginal dilation, individuals with extensive urogenital malformations, such as a cloaca, or for patients who are unsuccessful in creating a functional vagina with one of the other described surgeries [13, 15, 49, 50]. This technique may be the preferred option for patients with severe pelvic scarring from prior procedures or pelvic radiation [13, 15, 49, 50]. The Williams vaginoplasty involves the creation of an exterior "kangaroo pouch" horizontal to the perineum. The pouch-space is created by suturing full-thickness skin flaps from the labia in a "U-shaped" configuration [15, 49–51]. Patients must be counseled on the different axis required for intercourse after this procedure, and the need for dilator use or regular intercourse to prevent adhesion formation and maintain the space as a functional vagina.

Complications and Outcomes of Surgical Neovagina Techniques

In studies of surgical techniques for vaginal construction in patients with Müllerian agenesis, no single procedure is superior and each carries unique disadvantages, side effects, or complications (Table 2.1). Although surgical management may produce more rapid results for these patients, surgery is still considered secondary to vaginal dilation in the management of vaginal agenesis due to their surgical risks and potentially morbid complications [22].

Active Dissection of the Perineal Space

Long-term surgical outcomes for the McIndoe procedure have been well-studied and functional success was reported in 85% of girls who underwent this procedure in a study by Buss et al. [52]. Reported complications of this procedure come in a wide range of severity and rates range from 19% to 65% [38, 53]. Disadvantages of this technique include the need for two surgeries, hospitalization, prolonged postoperative dilator, and unique surgical risks including graft failure. Surgical complications include vaginal stenosis, disfiguring scar at donor graft site, wound infection, and fistula formation [52, 54]. The McIndoe techniques involving alternative material grafted into the vaginal space have similar reported outcomes and there is inconclusive evidence demonstrating advantage over the classic split-thickness Abbe-McIndoe procedure.

Autologous Pelvic Peritoneum

In a retrospective cohort study by Willemsen et al., 160 women with vaginal agenesis who underwent neovagina procedure by the Davydov technique were studied for long-term outcomes [55]. Women who underwent the Davydov procedure had a 68–87% success in creating a functional vagina (defined as length greater than 5 cm) with a mean vaginal length of 7.8 cm; the results did not change significantly if the patient performed vaginal dilation prior to surgery [55]. Sexual satisfaction scores after this procedure are shown to be similar for scores in sexual arousal, lubrication, orgasm, and comfort in studies com-

paring these patients to a random female control population [55, 56]. The rate of adverse outcomes associated with the Davydov peritoneal neovagina is reported at 14% in one study [57]. Complications associated with this procedure include rectal or bladder injury at the time of the procedure, recto-neovaginal fistula, pelvic adhesions, granulation tissue formation, stricture, and vaginismus [55, 56].

Active Perineal Dilation

The laparoscopically assisted neovagina creation by Vecchietti active perineal dilation results in a successful creation of a functional vagina in over 98% [42, 58]. The reported mean length of 9.5 centimeters with sexual satisfaction scores that are similar to female age-matched controls in several studies [43, 58]. Similar to the McIndoe procedure, the Vecchietti method requires postoperative hospitalization and two surgeries 1 week apart. There must be aggressive pain control and close monitoring during the active portion of vaginal dilation. Typical complications of the Vecchietti procedure are postoperative fever, urinary tract infection, granulation tissue formation, vaginal stricture, and rarely, urethral necrosis [43, 58]. Rates of adverse outcomes of either laparoscopic or open approach Vecchietti are 11–13% [41].

Intestinal Vaginoplasty

Bowel vaginoplasty has been performed for over a century with multiple long-term studies of outcomes, complications, and modifications of the procedure. Most studies of this surgery report a 73–83% patient satisfaction with final vaginal length (mean of 10 cm) and good sexual function [48, 59–61]. Advantages of bowel vaginoplasty include the lack of multiple procedures or required postoperative vaginal dilation.

Multiple complications and complaints after bowel vaginoplasty have been reported and the exact rate of all postoperative issues is difficult to know. The biggest disadvantages include the common complaint of copious foul smelling mucus discharge requiring daily pad use or douching, trauma with intercourse (especially if small bowel is used), and an abnormal vaginal length later in life if the surgery is performed in childhood [13, 15, 23, 47, 48, 62]. The occurrence rates of significant complications range from 7% to 79% and include introital stenosis requiring dilation (especially if blood supply is compromised), mucosal prolapse, small bowel obstruction, and fistula formation [23, 47, 62].

Labial Skin Flap or Vulvovaginal Pouch

The Williams vulvovaginoplasty is the most simple, noninvasive neovagina surgical technique and is the only reversible option currently available. First described by Williams 1964, the Williams procedure and other reported surgical modifications demonstrate good results in sexual function and patient satisfaction with final vaginal length [49, 51]. Creatsas et al. found that of 178 patients with MRKH, approximately 95% were successful in obtaining a vaginal length of 10–12 cm and 94% of patients reported satisfactory quality of sexual life after surgery [51]. Data on complication rates is limited as this procedure is rarely performed. In reports by Creatsas et al., specific issues such as wound complications, hematoma formation, or need for dilation each occurred in about 4%. Those requiring dilation (4.5%) had a 7–9 cm neovagina and most reported good sexual function after dilator therapy [51, 63]. Patient complaints after Williams procedure are often of postoperative bleeding, need for initial hospitalization, scarring at the graft site or vulva, different vaginal axis with intercourse, and irritating hair growth within the vagina [51, 63].

Criteria of "success" vary between surgeries and studies. Most cited sources define surgical success by a patient's satisfaction with the neovagina, subsequent sexual function, and/or a final vaginal length of more than 5–6 cm.

"Vaginal length" in centimeters is reported in the referenced studies as mean, range, or an average.

Conclusion

Management of patients with vaginal agenesis, with or without a functional uterus, is complex and clinical recommendations depend on age at diagnosis and the clinical presentation. Disclosing

the diagnosis of a reproductive tract anomaly can be distressing especially with anomalies that affect future fertility or in patients that require vaginal dilation or surgery. Frequent counseling visits with the patient and family by a provider familiar with the management of these complex disorders are required. When appropriate, girls and women without a developed uterus should be provided with information on available advanced fertility options and their alternatives if childbearing is desired. Referral to a specialty or tertiary center may be necessary as developmental defects involving other organ systems are frequently encountered in girls with genital tract anomalies and a multidisciplinary approach is preferred.

Patients with vaginal agenesis and their families can be reassured of the favorable outcomes in most by vaginal dilation only. If dilation is not an option or is unsuccessful for these patients, many surgical techniques exist for the creation of a neovagina with high success rates and good sexual satisfaction. If a provider is comfortable recommending and performing one of these procedures, the postoperative requirements and unique surgical complications should be carefully reviewed before surgery.

Preventative health care and screening recommendations for women and girls with Müllerian anomalies should not be overlooked. Genital tract anomalies are not contraindications to the human papillomavirus (HPV) vaccine [64]. This vaccine is administered in late childhood and is recommended for all patients regardless of their genital anatomy as it can help decrease virus transmission to sexual partners and prevent HPV-related oropharyngeal or genital tract malignancies [64]. Patients with Müllerian agenesis or cervical atresia do not fall under the USPTF (US Preventative Services Task Force) criteria for routine cervical cancer screening regardless of sexual activity or HPV vaccination status [65]. Those with an identifiable cervix should undergo routine cervical cancer screening starting at age 21 according to current guidelines [65]. Patients with duplicated Müllerian systems require cytology samples of each cervix at each screening. Safe sex counseling and annual screening for sexually transmitted infections, especially Gonorrhea and Chlamydia, are recommended for all sexually active women younger than 25 years, or older if additional risk factors are present [66]. Lastly, preventative health visits that include an annual pelvic and breast exam by a healthcare provider are recommended for all women regardless of their anatomy.

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