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Committee on Adolescent Health Care

The North American Society for Pediatric and Adolescent Gynecology endorses this document. This Committee Opinion was developed by the American College of Obstetricians and Gynecologists' Committee on Adolescent Health Care in collaboration with committee member Anne-Marie E. Amies Oelschlager, MD.

Müllerian Agenesis: Diagnosis, Management, and Treatment

ABSTRACT: Müllerian agenesis, also referred to as müllerian aplasia, Mayer-Rokitansky-Küster-Hauser syndrome, or vaginal agenesis, has an incidence of 1 per 4,500–5,000 females. Müllerian agenesis is caused by embryologic underdevelopment of the müllerian duct, with resultant agenesis or atresia of the vagina, uterus, or both. Patients with müllerian agenesis usually are identified when they are evaluated for primary amenorrhea with otherwise typical growth and pubertal development. The most important steps in the effective management of müllerian agenesis are correct diagnosis of the underlying condition, evaluation for associated congenital anomalies, and psychosocial counseling in addition to treatment or intervention to address the functional effects of genital anomalies. The psychologic effect of the diagnosis of müllerian agenesis should not be underestimated. All patients with müllerian agenesis should be offered counseling and encouraged to connect with peer support groups. Future options for having children should be addressed with patients: options include adoption and gestational surrogacy. Assisted reproductive techniques with use of a gestational carrier (surrogate) have been shown to be successful for women with müllerian agenesis. Nonsurgical vaginal elongation by dilation should be the first-line approach. When well-counseled and emotionally prepared, almost all patients (90–96%) will be able to achieve anatomic and functional success by primary vaginal dilation. In cases in which surgical intervention is required, referrals to centers with expertise in this area should be considered because few surgeons have extensive experience in construction of the neovagina and surgery by a trained surgeon offers the best opportunity for a successful result.

Recommendations and Conclusions

The American College of Obstetricians and Gynecologists makes the following recommendations and conclusions:

- Patients with müllerian agenesis usually are identified when they are evaluated for primary amenorrhea with otherwise typical growth and pubertal development.
- Rudimentary müllerian structures are found in 90% of patients with müllerian agenesis by magnetic resonance imaging. On ultrasonography, these rudimentary müllerian structures are difficult to interpret and may be particularly misleading before puberty.
- Evaluation for associated congenital anomalies is essential because up to 53% of patients with müllerian agenesis have concomitant congenital malformations, especially of the urinary tract and skeleton.
- All patients with müllerian agenesis should be offered counseling and encouraged to connect with peer support groups.
- Future options for having children should be addressed with patients.
- Primary vaginal elongation by dilation is the appropriate first-line approach in most patients because it is safer, patient-controlled, and more cost effective than surgery.
- Because primary vaginal dilation is successful for more than 90–96% of patients, surgery should be reserved for the rare patient who is unsuccessful with primary dilator therapy or who prefers surgery

after a thorough informed consent discussion with her gynecologic care provider and her respective parent(s) or guardian(s).

- Regardless of the surgical technique chosen, referrals to centers with expertise should be offered. The surgeon must be experienced with the procedure because the initial procedure is more likely to succeed than follow-up procedures.
- Although vulvar and vaginal intraepithelial neoplasia are possible, routine cytology testing is not regularly recommended because of the lack of a cervix.
- Sexually active women with müllerian agenesis should be aware that they are at risk of sexually transmitted infections and, thus, condoms should be used for intercourse. Patients should be appropriately screened for sexually transmitted infections according to the guidelines for women without müllerian agenesis.
- Patients should be given a written medical summary of their condition, including a summary of concomitant malformations. This information may be useful if the patient requires urgent medical care or emergency surgery by a health care provider unfamiliar with müllerian agenesis.

Background

Müllerian agenesis, also referred to as müllerian aplasia, Mayer–Rokitansky–Küster–Hauser syndrome, or vaginal agenesis, has an incidence of 1 per 4,500–5,000 females (1). Müllerian agenesis is caused by embryologic underdevelopment of the müllerian duct, with resultant agenesis or atresia of the vagina, uterus, or both. The vaginal canal is markedly shortened and may appear as a dimple below the urethra. A single midline uterine remnant may be present or uterine horns (with or without an endometrial cavity) may exist. The ovaries, given their separate embryologic source, are typically normal in structure and function, though they may be found in atypical locations.

Differential Diagnosis

Patients with müllerian agenesis usually are identified when they are evaluated for primary amenorrhea with otherwise typical growth and pubertal development. Müllerian agenesis is one of the most common causes of primary amenorrhea in patients with typical thelarche and adrenarche. On physical examination, patients with müllerian agenesis have normal height, breast development, body hair, and external genitalia. The vagina is present and may appear as a small flush dimple, or longer, without a cervix at the vaginal apex. The differential diagnosis of a patient presenting with primary amenorrhea and a shortened lower vagina includes obstructing vaginal or uterine anomalies, including imperforate hymen, transverse vaginal septum, or cervical atresia.

Additionally, 46,XY differences (or disorders) of sex development may present with primary amenorrhea and absent uterus, including androgen insensitivity syndrome.

The initial evaluation of an adolescent patient with primary amenorrhea includes a physical examination to assess for signs of appropriate or delayed puberty. In patients with müllerian agenesis, thelarche and adrenarche will be appropriate for age, but genitourinary examination will reveal a distal vagina that may be shortened without a palpable cervix. Imaging will not identify a typical midline uterus. Other diagnoses that may be confused with müllerian agenesis include vaginal or uterine obstructions or 46,XY differences of sex development.

An evaluation to exclude obstructive anomalies, including imperforate hymen, distal vaginal atresia, transverse vaginal septum, and cervical atresia, should be performed. On physical examination, the imperforate hymen with hematocolpos will appear as a blueish-colored bulging membrane without the typical hymenal fringe. Distal vaginal atresia may appear as a pink vaginal dimple and may be bulging without a hymenal fringe. The patient with a low transverse vaginal septum usually will have a normal hymen with more proximal obstruction of the vaginal canal. A rectal examination often will identify a bulging of the proximal vagina. In adolescents or women with cervical atresia, the vagina typically is shortened or may appear as a dimple. Although external examination may be similar to müllerian agenesis, imaging studies are indicated to delineate internal anatomic structures. Postpubertal pelvic imaging usually will reveal hematocolpos, or hematometra, or both. In addition to presenting with primary amenorrhea, all these conditions may occur with symptoms of cyclic or persistent abdominal or pelvic pain and a pelvic mass due to the obstructed outflow tract.

One of the most common conditions that may be confused with 46,XX müllerian agenesis is androgen insensitivity syndrome. Both conditions often present with primary amenorrhea, shortened vagina, and absent cervix. In patients with androgen insensitivity, the gonads are testes, which produce normal androgens. Patients with androgen insensitivity have typical breast development because of the peripheral aromatization of testosterone to estrogen. The lack of functional androgen receptors results in decreased or absent pubic and axillary hair. The vagina is typically shortened and the uterus or cervix does not develop because of in utero production of müllerian-inhibiting substance by the testes. Although the incidence of androgen insensitivity syndrome in females is 1:20,000, it may be as high as 1.1% in female infants with inguinal hernias (2, 3). The diagnosis of androgen insensitivity syndrome is made by assessing serum testosterone levels, which will be in the typical male range, followed by karyotype analysis, which is 46,XY. This is an X-linked recessive condition and androgen receptor sequencing can be performed for

confirmation. Although the testes are intraabdominal in most women with complete androgen insensitivity syndrome, they are not dysgenetic and, therefore, the risk of gonadoblastoma in adolescence and young adulthood is estimated to be 2% (4). The estrogen produced from the peripheral aromatization of testosterone produced by the testes will result in spontaneous progression through puberty. Therefore, prophylactic gonadectomy should be delayed until after puberty when the patient truly understands the risks and benefits and can choose to have the procedure, and is prepared to take ongoing hormone replacement therapy to maintain bone mass and decrease the risk of low-bone-mineral density. The risk of gonadoblastoma in nonpalpable gonads is higher in patients with the clinical diagnoses of partial androgen insensitivity and testosterone synthesis disorders compared with patients with complete androgen insensitivity syndrome. There is a lower threshold for gonadal biopsy with orchiopexy or gonadectomy (5). Given the controversy and complexity of decision making surrounding gonadectomy for patients with differences of sex development, referrals to centers with expertise should be offered.

If the physical examination of the patient with primary amenorrhea demonstrates delayed puberty, a serum follicle stimulating hormone level (FSH) and karyotype should be performed. The most common genetic etiology of pubertal delay and primary amenorrhea is Turner syndrome with a 45,X karyotype and an elevated FSH. Additional testing for the presence of Y chromatin (mosaicism) should be considered. The patient with Turner syndrome usually will have short stature, a typical length vagina, cervix and uterus present, and delayed puberty due to hypogonadism.

Another etiology of pubertal delay with typical external female genitalia and absent cervix is *CYP17A1* deficiency. This is a rare autosomal recessive heterogeneous form of congenital adrenal hyperplasia with an incidence of 1:50,000 to 1:100,000 that may be confused with müllerian agenesis. Individuals will have impaired sex steroid and cortisol synthesis and overproduction of aldosterone with resultant hypertension and hypokalemia. An individual with a 46,XX karyotype will have a uterus and vagina, but an individual with a 46,XY karyotype may have phenotypically female external genitalia, a shortened vagina, no uterus, and intraabdominal testes. The diagnosis can be confirmed by testing of the *CYP17A1* gene and careful interpretation of adrenal steroids, including elevated serum deoxycorticosterone and corticosterone levels and low cortisol, androgens, and estrogen levels (6, 7).

Evaluation of the Patient With Müllerian Agenesis

Initial evaluation of the patient without a uterus may include the following laboratory tests: testosterone

level, FSH level, and karyotype. Initial radiologic evaluation includes transabdominal, translabial, or transrectal two-dimensional or three-dimensional ultrasonography to assess for the presence of a midline uterus. Rudimentary müllerian structures are found in 90% of patients with müllerian agenesis by magnetic resonance imaging (MRI). Additionally, MRI can assess for the presence of endometrial activity within the müllerian structures (8).

If active endometrium is present, the patient may experience cyclic or chronic abdominal pain. On ultrasonography, these rudimentary müllerian structures are difficult to interpret and may be particularly misleading before puberty (9). The MRI should be ordered with specific instructions to assess for müllerian remnants and the results should be interpreted by a radiologist with experience in evaluating müllerian tract structures (8). The MRI typically can be done without contrast, but this decision can be left to the discretion of the radiologist.

Although laparoscopy is not necessary to diagnose müllerian agenesis, it may be useful in the evaluation and management of patients who report pelvic pain. Patients may experience pain from ovulation or endometriosis, which may improve with hormonal suppression. Patients also may develop endometriosis from retrograde menstruation from obstructed uterine horns. When obstructed uterine horns with the presence of active endometrium without an associated cervix and upper vagina are identified, then laparoscopic removal of the unilateral or bilateral obstructed uterine structures should be performed (10). In most cases, surgical excision of the uterine horn results in improvement of the endometriosis (11).

Evaluation for associated congenital anomalies is essential because up to 53% of patients with müllerian agenesis have concomitant congenital malformations, especially of the urinary tract and skeleton (12). Multiple studies have confirmed the prevalence of renal anomalies in patients with müllerian agenesis to be 27–29%; therefore, ultrasound evaluation of the kidneys is warranted for all patients (13, 14). Skeletal anomalies (eg, scoliosis, vertebral arch disturbances, hypoplasia of the wrist) have been reported in approximately 8–32% of patients; therefore, spine radiography (X-ray) may reveal a skeletal anomaly even in asymptomatic patients (12–14). There is an increased, but small, rate of hearing impairment in patients with müllerian agenesis (12). A variety of uterine anomalies, including müllerian agenesis, can be seen with VATER/VACTERL association (vertebral anomalies, anorectal malformations, cardiovascular anomalies, tracheoesophageal fistula, esophageal atresia, renal anomalies, limb defects) (15).

Karyotype evaluation of patients with müllerian agenesis will be 46,XX in most individuals. Given the heterogeneity of müllerian agenesis, it is not surprising that there have been several karyotype rearrangement abnormalities reported, including duplications and deletions,

as well as individual gene mutations such as the *WNT4* and *WNT9* genes (1). A consultation with a geneticist with experience with müllerian agenesis may be helpful for additional genetic testing.

Psychosocial Counseling and Support

All patients with müllerian agenesis should be offered counseling and encouraged to connect with peer support groups. The psychologic effect of the diagnosis of müllerian agenesis should not be underestimated. Many patients experience anxiety and depression, question their female identity, and grieve their infertility. These patients struggle with how to share their conditions with family members, peers, and romantic partners (16, 17). The best predictor of good emotional outcome after diagnosis is a good relationship between the patient and her parent(s) or guardian(s) and the ability to share feelings with family and friends (18). Contact with a support group of young women with the same diagnosis may be especially helpful (10). In addition, parents and guardians also may benefit from counseling to address their feelings and to allow them to support their child better. See For More Information for resources.

Fertility

Future options for having children should be addressed with patients, including adoption and gestational surrogacy. Assisted reproductive techniques with use of a gestational carrier (surrogate) have been shown to be successful for women with müllerian agenesis. Female offspring of women who achieved pregnancy by assisted reproductive technology usually have normal reproductive tracts, although familial aggregates of müllerian agenesis have been reported (19–21). Uterine transplantation has resulted in live births, but given limited data, this procedure currently is considered experimental (22, 23) and is not widely available. Understanding future fertility options allows adolescents and young women to understand their potential for becoming parents, which may help them cope with the diagnosis and its implications.

Management of Patients With Müllerian Agensis

Management of patients with müllerian agenesis includes psychosocial counseling as well as treatment of the anatomic anomalies. Options include vaginal elongation and the surgical creation of a neovagina.

Vaginal Elongation

Primary vaginal elongation by dilation is the appropriate first-line approach in most patients because it is safer, patient controlled, and more cost effective than surgery (24, 25). When well-counseled and emotionally prepared, almost all patients (90–96%) will be able to achieve anatomic and functional success by primary vaginal dilation (26, 27). Although it is a successful approach, many

obstetrician–gynecologists do not receive training in primary vaginal dilation and may not feel equipped to counsel and coach their patients adequately (28). Additional training for the obstetrician–gynecologist or referral to a health care provider with experience guiding patients through primary dilation therapy (eg, an experienced pelvic floor physical therapist) may be warranted.

Assessing Patient Readiness

Nonsurgical or surgical vaginal elongation should wait until the patient is emotionally mature and expresses the desire to proceed with therapy. There are multiple risks of failure of dilation (eg, poor motivation, unstable relationships, interpersonal conflict, parental misunderstanding of diagnosis, sociocultural factors, and mental health issues), most of which are not anatomic and may predict poor adherence to postoperative dilation. Cognitive issues that affect adherence to dilation may include the following: limited comprehension of the diagnosis and anatomy, young age, underlying learning disability, and inadequate knowledge of the dilation process. Logistical barriers to successful dilation include lack of privacy and limited ability to travel to clinic for close follow-up. In a study of adolescent girls and women in whom müllerian agenesis was diagnosed, respondents reported lack of motivation, uncertainty that dilation would be successful, and the perception of dilation as a negative experience as barriers to use (29). Finally, anatomic considerations include discomfort and pain, scar from prior procedures, the absence of dimple, and the presence of multiple congenital anomalies (30). The patient should be encouraged to wait to start dilation until she feels emotionally and physically ready to begin the process.

Technique

Dilation should take place in a supportive setting with close monitoring that is tailored to the individual adolescent or woman. Initially, the patient should have a thorough examination with a mirror so that she can identify her clitoris, urethra, and distal vagina. She should be able to understand and demonstrate the appropriate location and angle to place the dilator. She should be instructed to place progressive dilators on the distal vaginal apex for 10–30 minutes one to three times per day (30, 31). There are many dilator options available and the patient may want to try different dilators or vibrators to determine which ones are the most comfortable to use. Online support groups may provide links to purchase dilators online. Strategies for privacy should be discussed. Ideally patients may be seen weekly or biweekly for close follow-up to monitor progress, to manage adverse effects (including pain and bleeding), and to provide encouragement. Involvement of an experienced pelvic floor physical therapist also may be beneficial (32). Notably, there is no set length that must be achieved before penetrative intercourse is permitted; indeed, elongation by vaginal intercourse alone can be successful (33, 34).

Troubleshooting

Common adverse effects reported with dilation include urinary symptoms, bleeding, and pain. If these are experienced, the patient should be evaluated if possible to assess for vaginal abrasion, urinary dysfunction, and urinary tract infection (35). The most commonly recommended treatments for bleeding are to increase use of lubricant, switch to a wider or softer dilator, and rest the pelvis until the bleeding has ceased. Treatments for pain include switching to a wider or softer dilator and increasing use of lubricants. The patient also should be assessed for dysfunctional voiding and vaginismus.

Defining Success and Failure

Patients who have previously attempted primary dilation may have been told or may assume that they “failed” dilation; however, close questioning often reveals that the patients may have not had an adequate understanding of the process and may not have been appropriately coached (29). A dilated vagina may not appear on examination as a typical vagina; however, appearance does not dictate function. Although some studies define success anatomically by a length of 6 cm or longer (31), the best definition of success is a vagina that is functional for comfortable sexual activity, as reported by the patient. There is no starting length associated with functional success, and, therefore, even patients with a flush vaginal dimple should be encouraged to dilate as first-line therapy. Based on expert opinion, patients who successfully use dilation therapy may require continuation of dilation on an intermittent basis if they are not regularly engaging in vaginal intercourse (30, 31). Patients who have stopped dilating should be reassured that they will not cause themselves harm, but they may need to resume dilation before sexual activity in the future. The patient should be empowered to determine when she is ready to start dilation and encouraged to proceed with dilation at her own rate.

Surgical Creation of a Neovagina

Surgical creation of a vagina requires ongoing postoperative dilation or vaginal intercourse to maintain adequate vaginal length and diameter; therefore, it is not a method to avoid vaginal dilator therapy. Because primary vaginal dilation is successful for more than 90–96% of patients, surgery should be reserved for the rare patient who is unsuccessful with primary dilator therapy (26, 27) or who prefers surgery after a thorough informed consent discussion with her gynecologic care provider and her respective parent(s) or guardian(s). Unlike primary vaginal dilation therapy, failing to adhere to postsurgery dilation can have deleterious effects.

The primary aim of surgery is the creation of a vaginal canal to allow penetrative intercourse. The timing of the surgery depends on the patient and the type of procedure planned. Surgical procedures often are performed in late adolescence or young adulthood when the patient

is mature enough to agree to the procedure and to be able to adhere to postoperative dilation.

Several surgical techniques may be used to create a neovagina. Regardless of the surgical technique chosen, referrals to centers with expertise should be offered. The surgeon must be experienced with the procedure because the initial procedure is more likely to succeed than follow-up procedures. Patients should be thoroughly counseled about surgical pain and the need for very close postoperative care. Compared with primary vaginal dilation, vaginoplasty complications are much more common and include bladder or rectal perforation, graft necrosis, hair-bearing vaginal skin, fistulae, diversion colitis, inflammatory bowel disease, and adenocarcinoma (31). At present, there is no consensus in the literature regarding the best option for surgical technique to afford the best functional outcome and sexual satisfaction (36).

Historically, the most common surgical procedure used to create a neovagina has been the modified Abbe-McIndoe operation. This procedure involves the dissection of a space between the rectum and bladder, placement of a stent covered with a split-thickness skin graft into the space, and the diligent use of vaginal dilation postoperatively. Other procedures for the creation of the neovagina are the Vecchietti procedure and other laparoscopic modifications of operations previously performed by laparotomy (37). The laparoscopic Vecchietti procedure is a modification of the open technique in which a neovagina is created using an external traction device that is affixed temporarily to the abdominal wall (38). Another procedure, the Davydov procedure, was developed as a three-stage operation that requires dissection of the rectovesicular space with abdominal mobilization of a segment of the peritoneum and subsequent attachment of the peritoneum to the introitus (39–42). Other vaginoplasty graft options include bowel, buccal mucosa, amnion, and various other allografts. Postoperative dilation is essential to prevent significant neovaginal stenosis and contracture; therefore, these techniques are not recommended if the patient objects to dilation. Dilators must intermittently be used until the patient engages in regular and frequent sexual intercourse.

General Gynecologic Care

Health care providers should be aware that some routine gynecology questions, including the date of last menstrual period, are unnecessary and may make patients have less confidence in the health care team. The patient should be asked about any vaginal discharge, bleeding, pelvic pain, or dyspareunia. Pelvic examinations should be performed if there are concerns about complications, vaginal stricture, or stenosis. If a patient is symptomatic, vaginal speculum examination and inspection should be performed to check for possible malignancy, colitis, ulceration, or other problems. Although vulvar and vaginal intraepithelial neoplasia are possible, routine cytology

testing is not regularly recommended because of the lack of a cervix. However, if an abnormal lesion is identified, biopsy is warranted. Although the vagina may not appear typical postprimary dilation or surgery, appearance does not dictate function.

Sexually active women with müllerian agenesis should be aware that they are at risk of sexually transmitted infections and, thus, condoms should be used for intercourse. Patients should be appropriately screened for sexually transmitted infections according to the guidelines for women without müllerian agenesis (43). Human papillomavirus vaccination of girls and young women is recommended because it may decrease the risk of vulvar and vaginal neoplasia and genital warts (44, 45). Finally, patients should be given a written medical summary of their condition, including a summary of concomitant malformations. This information may be useful if the patient requires urgent medical care or emergency surgery by a health care provider unfamiliar with müllerian agenesis.

Conclusion

The most important steps in the effective management of müllerian agenesis are correct diagnosis of the underlying condition, evaluation for associated congenital anomalies, and psychosocial counseling in addition to treatment or intervention to address the functional effects of genital anomalies. Because of the sensitivity of MRI imaging, laparoscopy is seldom required to make the diagnosis, but may be appropriate in a patient presenting with pelvic pain. Nonsurgical vaginal elongation by dilation should be the first-line approach. In cases in which surgical intervention is required, referrals to centers that have health care providers with expertise in this area should be considered because few surgeons have extensive experience in construction of the neovagina and surgery by a trained surgeon offers the best opportunity for a successful result.

For More Information

The American College of Obstetricians and Gynecologists has identified additional resources on topics related to this document that may be helpful for ob-gyns, other health care providers, and patients. You may view these resources at: www.acog.org/More-Info/MullerianAgenesis.

These resources are for information only and are not meant to be comprehensive. Referral to these resources does not imply the American College of Obstetricians and Gynecologists' endorsement of the organization, the organization's website, or the content of the resource. The resources may change without notice.

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