

Reproductive performance of women with müllerian anomalies

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Purpose of review

This review discusses current diagnostic techniques for müllerian anomalies, reproductive outcome data, and management options in reproductive-age women.

Recent findings

Multiple retrospective studies have investigated reproductive outcomes with müllerian anomalies, but few current prospective studies exist. Uterine anomalies are associated with normal and adverse reproductive outcomes such as recurrent pregnancy loss and preterm delivery, but not infertility. Furthermore, unicornuate, didelphic, bicornuate, septate, arcuate, and diethylstilbestrol-exposed uteri have their own reproductive implications and associated abnormalities. Common presentations of müllerian anomalies and current diagnostic techniques are reviewed. Surgical intervention for müllerian anomalies is indicated in women with pelvic pain, endometriosis, obstructive anomalies, recurrent pregnancy loss, and preterm delivery. Although surgery for most uterine anomalies is a major intervention, the uterine septum is preferentially managed with a hysteroscopic procedure. Several recent studies and review articles discuss management of the septate uterus in asymptomatic women, infertile women, and women with a history of poor reproductive outcomes. Current assessment of reproductive outcomes with uterine anomalies and management techniques is warranted.

Summary

Müllerian anomalies, especially uterine anomalies, are associated with both normal and adverse reproductive outcomes, and management in infertile women remains controversial.

Keywords

congenital uterine anomalies, müllerian anomalies, recurrent pregnancy loss, septate uterus

Abbreviations

DES diethylstilbestrol
HSG hysterosalpingogram
RPL recurrent pregnancy loss

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Introduction

Congenital anomalies of the female reproductive tract may involve the uterus, cervix, fallopian tubes, or vagina. Depending on the specific defect, a woman's obstetric and gynecologic health may be adversely affected. Uterine anomalies are the most common of the müllerian anomalies, but the true incidence is not known since many women are asymptomatic, and sensitive imaging modalities have only recently become available [1,2]. Uterine anomalies are associated with both normal and adverse reproductive outcomes; they occur in approximately 3–4% of fertile and infertile women, 5–10% of women with recurrent early pregnancy loss, and up to 25% of women with late first or second-trimester pregnancy loss or preterm delivery [3–6]. Overall, uterine anomalies are associated with difficulty maintaining a pregnancy, and not an impaired ability to conceive [1,4]. The proper management of infertile women with uterine anomalies is controversial.

Embryology of the female reproductive tract

Normal development of the female reproductive tract requires a complex series of events: müllerian duct elongation, fusion, canalization, and septal resorption; failure of any part of this process can result in a congenital anomaly. Müllerian development occurs in close association with development of the urinary tract; thus, anomalies of the kidney and ureter are commonly identified in females with müllerian anomalies. Gonadal development occurs as a separate process, beginning by 7 weeks of gestation; therefore, women with müllerian anomalies usually have normal ovaries and ovarian hormone production.

The paired müllerian (paramesonephric) ducts are identifiable by week 6 of development, and arise from coelomic epithelium along the lateral walls of the urogenital ridge. These solid tissue structures elongate caudally, cross the wolffian (mesonephric) ducts medially, and fuse in the midline to form the primitive uterovaginal canal. By week 10, the caudal end of the fused müllerian ducts connects with the urogenital sinus. Next internal

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canalization of the müllerian ducts occurs, resulting in two channels divided by a septum. The septum is subsequently resorbed in the caudal to cephalad direction; this is completed by week 20. The fused caudal portion of the müllerian ducts becomes the uterus and upper vagina, and the unfused cephalad portion becomes the fallopian tubes.

The lower vagina has a different embryologic origin. Upon contact between the müllerian ducts and the urogenital sinus, sinovaginal bulbs originate and proliferate toward the caudal end of the uterovaginal canal, forming a solid vaginal plate. The lumen of the lower vagina is formed by degeneration of cells at the center of the vaginal plate; this process occurs in a caudal to cephalad direction and is complete by week 20. The hymenal membrane separates the vaginal lumen from the urogenital sinus. The central epithelial cells of the hymenal membrane usually degenerate prior to birth, and the hymen persists as a thin fold of mucus membrane at the introitus.

Classification of müllerian anomalies

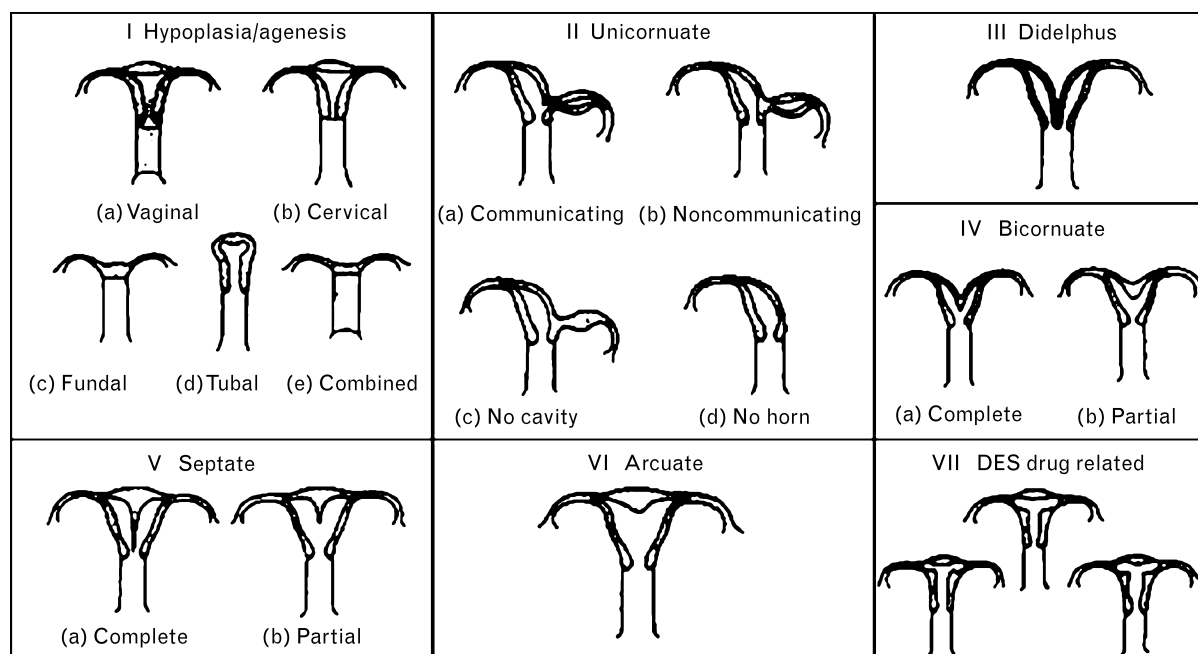
Congenital anomalies of the female reproductive tract are typically classified into three main categories: agenesis and hypoplasia, lateral fusion defects, and vertical fusion defects. A fourth group is composed of women exposed to diethylstilbestrol (DES) *in utero*. Agenesis and hypoplasia can occur with any or multiple müllerian structures. Lateral fusion defects occur due to failure of migration

of one müllerian duct, fusion of the müllerian ducts, or absorption of the intervening septum between the ducts. This is the most common category of müllerian defect, and can result in symmetric or asymmetric and nonobstructed or obstructed structures [7]. Depending on the population studied and the imaging modalities used, the most common uterine malformation is the arcuate, septate, or bicornuate uterus [1,3–5]. Vertical fusion defects result from abnormal fusion of the müllerian ducts with the urogenital sinus, or problems with vaginal canalization. These lesions can cause menstrual flow obstruction.

The etiology of congenital anomalies of the female reproductive tract is poorly understood. Karyotypes are normal (46XX) in 92% of women with müllerian anomalies, and abnormal in 7.7% of these women [8]. The majority of these developmental abnormalities are infrequent and sporadic, and are thus attributed to polygenic and multifactorial causes [2].

There is no universally accepted standard classification for congenital anomalies of the female reproductive tract. In 1988 the American Fertility Society (renamed the American Society for Reproductive Medicine) created a classification system that focuses on the major categories of uterine anomalies (Fig. 1) [9]. Documentation of associated anomalies of the vagina, cervix, fallopian tubes, and urinary system should be included.

Figure 1 American Society for Reproductive Medicine classification system for müllerian anomalies



DES, diethylstilbestrol. Reproduced from [9].

Clinical presentation of müllerian anomalies

Although many women with müllerian anomalies are asymptomatic, several gynecologic signs and symptoms are associated with specific anomalies. Women may present with cyclic or noncyclic pelvic pain and dysmenorrhea suggestive of an obstructive anomaly, retrograde menstruation, and endometriosis [1]. Endometriosis is a common finding in women with obstructive and non-obstructive müllerian anomalies, and may be a cause of infertility [10^{••},11]. Primary amenorrhea with pelvic, vaginal, or back pain or a pelvic mass is concerning for a transverse vaginal septum or an imperforate hymen. Müllerian agenesis (congenital absence of the uterus and vagina) presents with amenorrhea. Abnormal uterine bleeding may occur with a septate uterus, a partial or microperforate obstruction, or a longitudinal septum when only one tampon is used (two are required).

Of all müllerian anomalies, those involving the uterus are most commonly implicated in poor obstetric outcomes. Uterine anomalies are associated with diminished cavity size, insufficient musculature, impaired ability to distend, abnormal myometrial and cervical function, inadequate vascularity, and abnormal endometrial development [2,4,12–20]. These abnormalities of space, vascular supply, and associated local defects contribute to increased rates of recurrent pregnancy loss (RPL; 21–33%), preterm delivery, and malpresentation associated with uterine anomalies [1,3,5,13,16,21–23]. Intrauterine growth restriction is similarly attributed to abnormal vascularization and a smaller uterine cavity [19]. An increased cesarean delivery rate is associated with higher rates of malpresentation and vaginal anomalies such as a longitudinal vaginal septum. Furthermore, pregnancy may occur in an obstructed or rudimentary uterine horn; these conceptions rarely last until term, and 89% rupture [24]. Other associated obstetrical complications include cervical incompetence, pregnancy-induced hypertension (due to renal abnormalities), and antepartum and postpartum bleeding [1,4,5].

Evaluation of müllerian anomalies

Several radiologic techniques are useful for evaluating congenital anomalies of the female reproductive tract. Modalities employed include hysterosalpingogram (HSG), ultrasonography, sonohysterography, and MRI. Each imaging technique has inherent strengths and limitations; therefore a combination of several techniques may best evaluate a particular abnormality.

HSG is commonly used to assess the patency of fallopian tubes, and can provide further information about the contour of the endometrial cavity and the presence of any complex communications in the setting of a müllerian anomaly [25,26]. Definitive diagnosis of a uterine

abnormality requires assessment of the external uterine contour, and this is poorly defined by HSG [26]. The HSG correctly diagnoses 55% of septate and bicornuate uteri, and the addition of ultrasonography improves this result to 90% [27].

Transabdominal, transvaginal, or transperineal ultrasonography effectively evaluates the internal and external uterine contour, detects a pelvic mass, hematometra or hematocolpos, confirms the presence of ovaries, and assesses the kidneys. Timing the study to the secretory phase of the menstrual cycle provides better visualization of the endometrium and thus the internal uterine contour [28]. Sonohysterography can further delineate the intracavitary space, and internal and external uterine contours [29]. When available, three-dimensional ultrasonography is a highly accurate imaging modality that provides thorough views of the pelvic anatomy and detailed visualization of the uterus [5,30]; this is a reliable method for evaluating müllerian anomalies [26,30].

MRI is considered the gold standard technique for diagnosing müllerian anomalies, and is both sensitive and specific [25]. This modality provides excellent delineation of internal and external uterine contours [26]; MRI can distinguish a myometrial versus a fibrous uterine division and thus distinguish between bicornuate, didelphic, and septate uteri, and can determine the extent of a uterine or vaginal septum. Furthermore, MRI can identify a rudimentary uterine horn and determine if functional endometrium is present. The ability of MRI and three-dimensional ultrasonography to evaluate müllerian anomalies has not been directly compared.

Anomalies of the urinary tract are frequently seen with müllerian anomalies, thus appropriate urinary tract imaging is necessary. Upper urinary tract abnormalities include renal anomalies (20–30%) such as a horseshoe or pelvic kidney or renal agenesis, duplication of the collecting system, and ectopic ureters [1]. In the setting of an obstructive müllerian defect such as a noncommunicating uterine horn or an obstructed hemivagina, ipsilateral renal agenesis is commonly seen. Over 50% of the time, renal agenesis is predictive of an obstructive ipsilateral müllerian anomaly [24]. Recommended imaging techniques for the urinary tract are an intravenous pyelogram, renal ultrasound, or a computed tomography (CT) scan.

Indications for surgical intervention

Due to the availability of sophisticated imaging techniques, diagnostic surgical procedures such as an exam under anesthesia, vaginoscopy, hysteroscopy, and laparoscopy are rarely necessary to diagnose müllerian anomalies. Historically, laparoscopy and hysteroscopy were the gold standard for evaluating the contour of

Table 1 Reproductive outcomes in women with congenital uterine anomalies

Uterine anomaly	Number of studies	Number of patients	Number of pregnancies	Abortion rate	Preterm birth rate	Term delivery rate	Live birth rate
Unicornuate	11	151	260	36.5	16.2	44.6	54.2
Didelphys	8	114	152	32.2	28.3	36.2	55.9
Bicornuate	4	261	627	36	23	40.6	55.2
Septate	4	198	499	44.3	22.4	33.1	50.1
Arcuate	3	102	241	25.7	7.5	62.7	66

Rates are averaged and presented as a percentage. Data from Grimbizis *et al.* [4].

the uterine fundus and assigning the proper classification to a uterine anomaly [31].

Surgical correction of uterine anomalies in asymptomatic women or women with primary infertility is controversial. In general, uterine anomalies do not prevent conception or implantation, and these women can have normal reproductive outcomes. The mean prevalence of müllerian anomalies in infertile women is 3.4% (range 1.0–26.2%) [4], similar to the prevalence in the fertile population, which suggests that these abnormalities have little effect on fecundity [10^{••}]. In comparison, the prevalence of uterine anomalies in women with RPL is substantially higher at 12.6% [4]. When women with uterine anomalies undergo IVF, they have similar clinical pregnancy rates compared with women with normal uteri, but experience higher rates of miscarriage and preterm delivery [32].

Currently, surgery for müllerian anomalies is indicated for women with pelvic pain, endometriosis, obstructive anomalies, and poor obstetric outcomes such as RPL and preterm delivery. Prior to performing surgery, it is important to exclude extrauterine factors which may cause pregnancy loss [1,2,33[•]]. The goals of surgery include treatment of pelvic pain, restoration of pelvic anatomy and uterine architecture, and preservation of fertility. Inherent developmental abnormalities, however, such as abnormal myometrium or altered vascularization, may permanently impair uterine function [4,34[•]].

Unicornuate uterus

During embryogenesis, the failure of one müllerian duct to develop and elongate results in a unicornuate uterus. This asymmetric lateral fusion defect usually results in a functional uterus with a normal cervix and fallopian tube, and varying configurations of abnormal müllerian development on the contralateral side: agenesis, or a rudimentary uterine horn (74%) [35]. This rudimentary horn may be noncommunicating (70–90%) or communicating with the unicornuate uterus, and may have no endometrial cavity or some functional endometrium [24]. Although rudimentary horns are commonly asymptomatic, an obstructed horn with active endometrium can result in cyclic or chronic pelvic pain, endometriosis, or a horn gestation [24]. Furthermore, this anomaly is associated

with a high incidence of renal abnormalities (40%), usually ipsilateral to the anomalous side [1,13].

The unicornuate uterus is associated with higher rates of endometriosis, premature labor and delivery, and malpresentation [13]. Women with unicornuate uteri have impaired pregnancy outcomes; a compilation of studies reveals a spontaneous abortion rate of 36.5%, a preterm delivery rate of 16.2%, a term delivery rate of 44.6%, and a live birth rate of 54.2% [4] (Table 1). Surgical reconstructive procedures have not been shown to improve pregnancy outcomes [36]. Although prophylactic cervical cerclage has been recommended to improve pregnancy outcomes [37–39], women with müllerian anomalies should be managed expectantly with adherence to standard indications for cerclage placement [2,7[•],34[•]]. Additionally, removal of a functional rudimentary horn is recommended as treatment for pelvic pain and endometriosis, and to prevent conception in an obstructed horn [24,34[•]].

Uterine didelphys

The failure of fusion of the two müllerian ducts results in duplication of müllerian structures; a didelphic uterus has two uteri, two endometrial cavities, and two cervixes. A longitudinal vaginal septum is present in 75% of cases [40]. An obstructed hemivagina can occur with uterine didelphys, and this constellation of findings is associated with ipsilateral renal agenesis [41[•],42^{••},43[•]].

This uterine anomaly is associated with modest reproductive outcomes: a pooled spontaneous abortion rate of 32.2%, a preterm birth rate of 28.3%, a term delivery rate of 36.2%, and a live birth rate of 55.9% [4] (Table 1). In select women with RPL or preterm delivery, uterine reconstruction with the Strassman metroplasty should be considered [2,17]. The Strassman metroplasty achieves unification of two endometrial cavities in a divided uterus (bicornuate or didelphys), and is associated with a live birth rate greater than 80% [17]. Several experts believe, however, that existing data do not support repair of a didelphic uterus to improve pregnancy outcome [1,34[•],44]. In contrast, incision of the longitudinal vaginal septum is indicated for an obstructed hemivagina with hematocolpos, dyspareunia, or difficulty with tampon placement.

Bicornuate uterus

The bicornuate uterus occurs due to incomplete fusion of the two müllerian ducts at the level of the fundus, resulting in a single cervix and two endometrial cavities. The degree of separation between the two endometrial cavities can be variable, extending as far as the internal cervical os depending on the length of incomplete müllerian duct fusion. The external uterine contour has an indented fundus, arbitrarily defined as more than 1 cm, and the vagina is generally normal [26,45,46].

This anomaly is associated with obstetrical complications including pregnancy loss, preterm labor, and malpresentation. Grimbizis *et al.* [4] identified an overall spontaneous abortion rate of 36%, a preterm birth rate of 23%, a term delivery rate of 40.6%, and a live birth rate of 55.2% (Table 1). Furthermore, the incidence of preterm delivery varies with the degree of cavity separation in partial (29%) and complete (66%) bicornuate uteri [38].

In the setting of RPL or preterm delivery and a bicornuate uterus, all other etiologies of pregnancy loss must be excluded prior to considering a uterine reunification procedure. The Strassman metroplasty should be reserved for select women based on poor reproductive outcomes [2,17]. Furthermore, the bicornuate uterus is associated with a high incidence of cervical incompetence (38%) [14]. Although studies have identified improvements in fetal survival rates and decreased preterm delivery rates with a cervical cerclage [21,38,47], expectant management and appropriate adherence to standard indications for cerclage placement are warranted.

Septate uterus

A defect in resorption of the midline septum between the two müllerian ducts results in a fibromuscular uterine septum. The degree of septation is variable; a complete septum extends from the uterine fundus through the cervix, and a partial septum demonstrates resorption of a portion of the caudal aspect of the septum. Despite the endometrial cavity abnormality, the external uterine contour appears normal. A longitudinal vaginal septum is found most frequently with a septate uterus [48]. Endometriosis has been identified in up to 30% of fertile and infertile women with septate uteri [10^{••},49[•]].

The septate uterus is considered the most common of the uterine anomalies, occurring in approximately 1% of the fertile population [10^{••}], and is associated with the poorest reproductive outcomes [1,2,5,12,26,31,50]. A compilation of studies of partial and complete septate uteri identified a pregnancy loss rate of 44.3%, a preterm delivery rate of 22.4%, a term delivery rate of 33.1%, and a live birth rate of 50.1% [4] (Table 1). Raga *et al.* [5] further differentiated the timing of pregnancy loss into early (25.5% before 13 weeks) and late (6.2% between 14 and 22 weeks)

periods; a higher rate of reproductive failure is seen in the first trimester [10^{••}]. In comparison, a recent study [42^{••}] of reproductive outcomes with complete septate uteri identifies a spontaneous abortion rate of 27%, a preterm delivery rate of 12%, a term delivery rate of 61%, and a live birth rate of 72%. Compared with the rest of the uterus, septal tissue has been shown to have decreased vascular supply and abnormal overlying endometrium [45,51,52]; these findings help to explain the etiology of pregnancy loss with the septate uterus.

Hysteroscopic metroplasty has been demonstrated to significantly improve the live birth and miscarriage rates to approximately 80 and 15%, respectively [4,31], and is recommended when the uterine septum is implicated in RPL, second trimester loss, malpresentation, or preterm delivery [1]. The hysteroscopic approach is preferred due to its safety, simplicity, and excellent posttreatment results [31]. Concomitant laparoscopy enables evaluation of the pelvis and external uterine contour, and guides the extent of septum resection. Traditionally the cervical portion of a complete septum is left intact due to the risk of cervical incompetence [53,54], but a recent small randomized study [55[•]] demonstrated that resection of the cervical septum is associated with a less complicated surgical procedure and equivalent reproductive outcomes. Postoperative formation of intrauterine synechiae is rare, and routine use of an intrauterine balloon catheter, estradiol supplementation, or antibiotics have not been shown to be necessary [56–58]. A follow-up examination should be performed 1–2 months after the procedure; ultrasonography, HSG, and hysteroscopy are reasonable approaches [31].

Prophylactic hysteroscopic metroplasty in infertile women or women without a history of adverse reproductive outcomes is a controversial procedure since many women with a septate uterus can have reasonable pregnancy outcomes, and there is no established causal relationship between a septate uterus and infertility [1,4,6,10^{••},21,42^{••},59,60]. After hysteroscopic metroplasty in women with unexplained infertility, a modest improvement in pregnancy and live birth rates is demonstrated in nonrandomized trials [60]; these rates are significantly higher after metroplasty in women with RPL, which highlights the difference in fertility between these two populations [31,60,61]. Prophylactic metroplasty, however, may prevent miscarriage or other obstetrical complications, and is recommended in order to optimize pregnancy outcomes in women with prolonged infertility, in women older than 35 years, and in women planning to pursue assisted reproductive technologies [1,5,12,62,63].

Arcuate uterus

The arcuate uterus has been variably classified as a normal, bicornuate, or septate uterus. Anatomic hallmarks include

a slight midline septum with a broad fundus, sometimes with minimal fundal cavity indentation. Of all uterine anomalies, obstetrical complications are least common in anomalies, obstetrical complications are least common in women with an arcuate uterus [1,26]. Women with an arcuate uterus have an overall term delivery rate of 62.7% and a live birth rate of 66% [4] (Table 1), but one study [5] identified rates as high as 78% and 83%, respectively. Compared with women with a normal uterus, women with an arcuate uterus have a higher proportion of second trimester losses and preterm labor [50]. Reconstructive procedures on an arcuate uterus, however, do not improve pregnancy outcomes [36].

Diethylstilbestrol exposure

DES is a synthetic estrogen that was used between the 1940s and 1971 for the treatment of RPL, premature delivery, and other complications of pregnancy [64]. If a woman was exposed to DES *in utero*, uterine abnormalities are common. In one population of women with DES exposure *in utero*, 69% had uterine abnormalities on HSG [65]. The T-shaped uterine cavity was the most common abnormality (70%); other anomalies include a hypoplastic uterus, midfundal constriction rings, intrauterine filling defects, and endometrial cavity adhesions. DES exposure is also associated with cervical abnormalities such as cervical hypoplasia, hoods, collars, and pseudopolyps, and vaginal abnormalities such as adenosis, vaginal ridges, and transverse septa. The association between in-utero DES exposure and vaginal clear cell adenocarcinoma was identified in 1971, at which time DES use in pregnancy was prohibited [66]. Therefore, in-utero DES exposure will continue to affect women of reproductive age for another 10–15 years [2,67].

Uterine anomalies due to in-utero DES exposure are associated with an increased risk of adverse reproductive outcomes. Compared with controls, these women experience twice the risk of spontaneous abortion, and nine times the risk of ectopic pregnancy [67]. Due to uterine and cervical anomalies, pregnant women with a history of DES exposure are at risk of cervical incompetence; options include expectant management, bedrest, and prophylactic or emergent cerclage placement. Studies have shown a term delivery rate greater than 70% in DES-exposed women without and with cerclage (both prophylactic and emergent procedures) [23,67,68]. Cerclage placement may benefit DES-exposed women, thus appropriate candidates should be considered based on standard indications for cerclage placement.

Cervical anomalies

Most cervical abnormalities accompany vaginal or uterine anomalies; they are rarely isolated, so an MRI is necessary to define the anatomy [1]. Cervical anomalies include agenesis, atresia, abnormal length or width, obstruction,

and hypertrophy [1]. Cervical atresia is a rare anomaly; these women can present with primary amenorrhea and cyclic pelvic pain due to hematometra and retrograde menstruation. Successful pregnancies have occurred after utero-vaginal anastomosis for cervical atresia [69]. Surgical correction of obstructive cervical anomalies, however, rarely results in a patent passage and is associated with a high risk of ascending infection; a hysterectomy is often necessary [70,71]. The ovaries should be preserved, hence pregnancy can be achieved with IVF and a gestational carrier.

Vaginal anomalies

Vaginal anomalies include a transverse vaginal septum (a vertical fusion defect), a longitudinal septum, and an imperforate hymen. Although the transverse septum and imperforate hymen are not associated with other müllerian anomalies [72], the longitudinal vaginal septum often occurs with uterine anomalies such as a septate or didelphic uterus. Regardless, the vaginal anomalies themselves should not interfere with reproductive outcomes.

Each vaginal anomaly requires careful assessment of pelvic anatomy to make the correct diagnosis, and surgical repair is necessary in the presence of an obstructive anomaly. A transverse vaginal septum results from failure of fusion between the müllerian ducts and the urogenital sinus or abnormal vaginal canalization, and requires excision of the septum and vaginal anastomosis. A longitudinal vaginal septum may cause dyspareunia, difficulty with tampon placement, or obstructed labor, and should be excised if symptomatic or if the woman desires restoration of a normal vaginal canal. An imperforate hymen occurs due to incomplete degeneration of the central portion of the hymen, and requires excision of the excess hymenal tissue.

Vaginal agenesis

Vaginal agenesis is an uncommon condition, and most frequently occurs as congenital absence of the vagina with variable uterine development (Mayer-Rokitansky-Kuster-Hauser syndrome). This developmental anomaly occurs due to agenesis or hypoplasia of müllerian duct development. The incidence of this abnormality is one in 5000 female births [73,74]. These women have a 46XX karyotype, and normal ovaries, ovarian function, female external genitalia, and secondary sexual characteristics, but experience primary amenorrhea [75••]. This diagnosis must be differentiated from vaginal agenesis, androgen insensitivity, low-lying transverse vaginal septum, and imperforate hymen. Müllerian agenesis is also associated with extragenital anomalies: urologic (25–50%) and skeletal (10–15%) abnormalities, cardiac defects, auditory deficits, digital anomalies, and cleft palates [75••,76,77•,78•,79].

Although vaginal agenesis is usually accompanied by uterine agenesis, 7–10% of women have a normal but obstructed uterus, rudimentary uterus, or uterine horns with functional endometrium [80,81]. Functional endometrial tissue in an obstructed uterine remnant can cause pelvic pain from hematometra, hematosalpinx, and endometriosis. Imaging, usually with MRI, is essential to identify the structures present. If a normal uterus, cervix, and upper vagina is present with lower vaginal agenesis, surgical correction of the vaginal anomaly is warranted.

Women with müllerian agenesis can achieve pregnancy with their own oocytes through IVF and the use of a gestational carrier. These women respond to gonadotropin stimulation similarly to women with normal pelvic anatomy [82]. Only one congenital anomaly has been identified in the offspring of 34 women with müllerian agenesis [74]. To enable sexual intercourse, a neovagina can be created with vaginal dilators or surgery; several successful approaches are available [75**].

Conclusion

Although müllerian anomalies are relatively uncommon, they can significantly impact reproductive outcomes. A high index of suspicion is warranted in adolescents and reproductive-age women with pelvic pain, dysmenorrhea, abnormal bleeding, RPL, second trimester pregnancy loss, or preterm delivery. Detailed pelvic imaging with modalities such as HSG, ultrasonography, and MRI is necessary, as well as evaluation of the urinary tract as indicated. When a müllerian anomaly is identified, the woman should be counseled about reproductive prognosis, pregnancy outcomes, and evidence-based management. Many of these women can have normal reproductive outcomes, but intervention is recommended in the event of poor obstetric outcomes. Moreover, due to the ease and low morbidity associated with hysteroscopic metroplasty for a septate uterus and demonstrated improvement in obstetric outcomes, this procedure should be considered in infertile women with a septate uterus prior to advanced fertility treatment. The diagnosis and classification of müllerian anomalies are facilitated by the availability of excellent imaging techniques; hence, current data on reproductive outcomes with uterine anomalies should be assessed.

References and recommended reading

Papers of particular interest, published within the annual period of review, have been highlighted as:

- of special interest
- of outstanding interest

Additional references related to this topic can also be found in the Current World Literature section in this issue (p. 292).

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