

CHAPTER 115

VAGINAL AND UTERINE DUPLICATIONS

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Introduction

Congenital anomalies of the vagina, cervix, and uterus arise from errors in embryogenesis and are characterised by diversity in anatomic features, clinical presentation, and reproductive performance.¹ Uterine anomalies are associated with both normal and adverse reproductive outcomes.

Demographics

The true prevalence of vaginal and uterine duplications is unknown because the anomaly may not manifest until the reproductive years of the individual.² Although a prevalence of 3.2% in the healthy fertile population has been quoted in one study, another study quoted an incidence of 0.1–0.5%.^{3,4} Rackow and associates reported an incidence of 3–4% in fertile and infertile women, 5–10% in women with early recurrent pregnancy loss (RPL), and up to 25% in women with late first and second trimester pregnancy loss or preterm delivery.^{3,4} Uterine and vaginal duplications can present at birth, in childhood, in adolescence, or in adulthood.^{5,6,7}

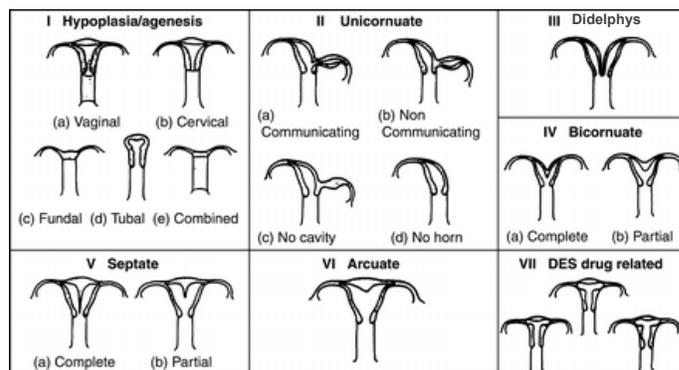
Aetiology/Pathophysiology

The aetiology of uterine anomalies is embryological in nature. Both male and female embryos initially have two pairs of genital ducts, the mesonephric and paramesonephric ducts. The paramesonephric duct arises as a longitudinal invagination of the coelomic epithelium of the anterolateral surface of the urogenital ridge in a 5- to 7-week embryo. Cranially, the duct opens in the coelomic cavity with a funnel-like structure; caudally, it runs first lateral to the mesonephric duct, but then crosses it ventrally to grow in a caudomedial direction. In the midline,

it comes in close contact with the paramesonephric duct from the opposite side.^{3,4} The two ducts are initially separated by a septum but later fuse to form the uterine canal. The caudal tips of the combined ducts project into the posterior wall of the urogenital sinus.

With the descent of the ovary, the upper two-thirds of the fused paramesonephric ducts develops into the uterine tube, and the lower third develops into the uterine canal. The fused walls of the paramesonephric ducts break down to form the cavity of the uterus. Defects in the fusion

Figure 115.1: Diagrammatic representation of the AFS classification of Müllerian duct anomalies.



Source: The American Fertility Society Classification of adnexal adhesions, distal tubal occlusion, tubal occlusion secondary to tubal ligation, tubal pregnancies, Müllerian anomalies and intrauterine adhesions. Fertility and Sterility, 1988; 49(6):944–955. Reproduced with kind permission of Elsevier®.

Table 115.1: The AFS classification.

Classification	Clinical Finding	Description
I	Segmental or complete agenesis or hypoplasia	Agenesis and hypoplasia may involve the vagina, cervix, fundus, tubes, or any combination of these structures. Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome is the most common example in this category.
II	Unicornuate uterus with or without a rudimentary horn	When an associated horn is present, this class is subdivided into communicating (continuity with the main uterine cavity is evident) and noncommunicating (no continuity with the main uterine cavity). The noncommunicating type is further subdivided on the basis of whether an endometrial cavity is present in the rudimentary horn. These malformations have previously been classified under asymmetric lateral fusion defects. The clinical significance of this classification is that they are invariably accompanied by ipsilateral renal and ureter agenesis.
III	Didelphys uterus	Complete or partial duplication of the vagina, cervix, and uterus characterises this anomaly.
IV	Complete or partial bicornuate uterus	Complete bicornuate uterus is characterised by a uterine septum that extends from the fundus to the cervical os. The partial bicornuate uterus demonstrates a septum, which is located at the fundus. In both variants, the vagina and cervix each have a single chamber.
V	Complete or partial septate uterus	A complete or partial midline septum is present within a single uterus.
VI	Arcuate uterus	A small septate indentation is present at the fundus.
VII	Diethylstilbestrol (DES)-related abnormalities	A T-shaped uterine cavity with or without dilated horns is evident.

Source: The American Fertility Society Classification of adnexal adhesions, distal tubal occlusion, tubal occlusion secondary to tubal ligation, tubal pregnancies, Müllerian anomalies and intrauterine adhesions. Fertility and Sterility, 1988; 49(6):944–955. Reproduced with kind permission of Elsevier®.

of the two paramesonephric ducts and failure of the breakdown of the fused middle walls gives rise to congenital anomalies of the uterus.^{3,4,8} The upper third of the vagina is derived from the uterine canal, and the lower two-thirds is of urogenital sinus origin. Shortly after the solid tips of the paramesonephric ducts reach the urogenital sinus, two solid evaginations grow out from the pelvic part of the sinus. These evaginations, the sinovaginal bulbs, proliferate strongly and form a solid plate. The proliferation continues at the cranial end of the plate, thus increasing the distance between the uterus and the urogenital sinus. By the 5th gestational month, the vaginal outgrowth is entirely canalised. The wing-like expansions of the vagina are of paramesonephric origin.⁸ Duplication and atresia of the uterine canal, or lack of fusion of the paramesonephric ducts in a localised area or throughout the length of the ducts, may explain all the different types of duplication of the uterus. In its extreme form, the uterus is entirely double (uterus didelphys).⁸

Classification

In 1979, Buttram and Gibbons⁹ proposed a classification arranged into six subgroups. This classification was revised in 1988 by the American Fertility Society (AFS) (Figure 115.1, Table 115.1).

Clinical Presentation

Complete duplication of the urogenital system in girls is a very rare congenital anomaly. It is characterised by widely variable modes of presentation, depending on the anomaly. Some children present early in the newborn period with abnormalities of the external genitalia, anorectal malformation, or urinary tract obstruction.³

Other groups remain asymptomatic for many years, and the anomalies are found during evaluation for infertility or as incidental findings in otherwise healthy young women.⁷ They are also most likely to have a higher rate of spontaneous abortion (about 32.2%), preterm birth rate (28.3%), RPL, foetal abnormalities, presentations, and obstructed labours.⁴

Patients with uterine duplication may also present with menorrhagia because of the increased surface area of the two endometrial cavities, dysmenorrhoea, and pelvic pain. Associated anomalies of the urinary system may be coexistent. Other findings include obstructed labour and antepartum and postpartum hemorrhage.

Physical examination may reveal the presence of two vaginal orifices.⁷

Investigation

Several radiologic techniques are useful for evaluating anomalies of the female reproductive tract. Modalities employed include hysterosalpingography, ultrasonography (US), sonohysterography, computed tomography (CT) scan, and magnetic resonance imaging (MRI).¹⁰

Hysterosalpingography

Hysterosalpingography, commonly used to assess the patency of the fallopian tubes, can provide further information about the contour of the endometrial cavity and the presence of any complex communication in the setting of a Müllerian anomaly^{3,4} (Figure 115.2).

Pelvic Ultrasound Scan

Definitive diagnosis of a uterine abnormality requires assessment of the external uterine contour. Transabdominal, transvaginal, or transperineal US effectively evaluates the internal and external uterine contour; detects a pelvic mass, haematometra, or haematocolpos; confirms the presence of ovaries; and assesses the kidneys. Transrectal US has been reported to help in defining the pelvic anatomy, which can be especially useful in young patients.¹¹ Timing the study to the secretory phase of the menstrual cycle provides better visualisation of the endometrium and thus the internal uterine contour⁴ (Figure 115.3).

Sonohysterography

Sonohysterography can further delineate the intracavitary space and internal and external uterine contours. Three-dimensional (3D) US (Figure 115.4) is a highly accurate imaging modality that provides

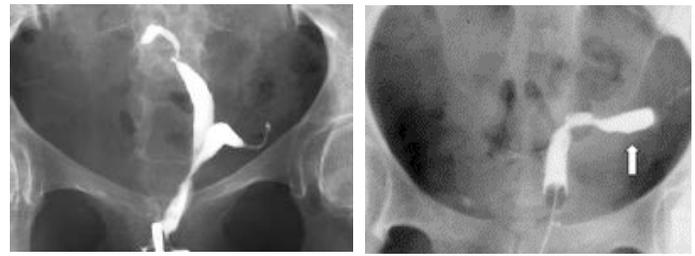


Figure 115.2: Hysterosalpingography of bipartite (left) and unicornuate (right) uteri.

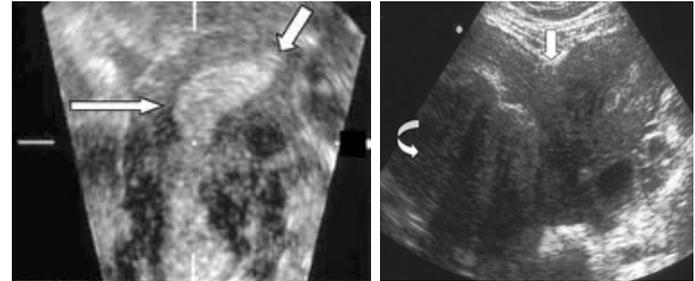


Figure 115.3: Sonographic appearance of Müllerian duct anomaly.

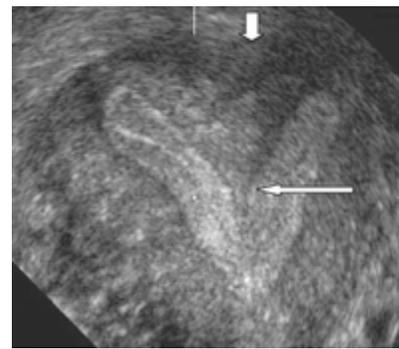


Figure 115.4: 3D sonographic view of a bicornuate uterus.

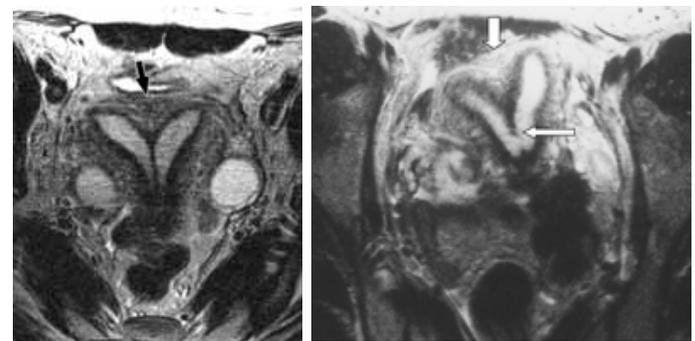


Figure 115.5: Magnetic resonance imaging appearance of bicornuate uterus.

thorough views of the pelvic anatomy and detailed visualisation of the uterus. This is a reliable method of evaluating Müllerian anomalies.¹⁰

A combination of sonohysterography and laparoscopy will sufficiently diagnose a uterine anomaly. The urinary system also needs to be evaluated by using US scan and an intravenous urography because of the coexistence of uterine anomalies and urinary abnormalities.¹⁰

Magnetic Resonance Imaging

MRI is considered the gold standard technique for diagnosing Müllerian anomalies; it is both sensitive and specific. This modality provides excellent delineation of internal and external uterine contours. MRI (Figure 115.5) can distinguish a myometrial versus a fibrous uterine

division and thus distinguish among bicornuate, didelphic, and septate uterus as well as determine the extent of a uterine or vaginal septum. MRI can also identify a rudimentary uterine horn and determine whether functional endometrium is present. However, in low-resource settings such as those found in the African continent, MRI may not be available, or if available may be unaffordable to some patients.^{4,5,11}

Management

The initial management of a patient with uterine and urinary duplication is to investigate and determine the degree of the anomaly and associated anomalies of the urinary system. A serum electrolytes and urea and blood cross match are needed.^{7,11} The management of uterine and urinary duplication in the younger group when associated with anorectal anomalies is done at the time of the repair of the anorectal anomaly.⁷

Management of these conditions typically relates to three primary issues: menstrual problems, fertility problems, and sexual function problems. The presence and severity of these areas, along with the type of anomaly present, will help to dictate the appropriate management. It should be stated that the mere presence of an abnormality does not necessitate treatment unless the patient is symptomatic as a result of it.⁴

Menstrual disturbances typically relate to outflow anomalies most commonly represented by a transverse or blocking septum, but also are seen with an absence of the vagina or cervical anomalies. In these circumstances, menstrual blood builds up behind the blockage and can result in pain. Additionally, it has been found that patients with blocked outflow are at increased risk of developing endometriosis—likely secondary to increased retrograde menses. The major concern in treating these patients is to create a passageway through which the menses can flow.

As mentioned previously, infertility is probably the most common presenting complaint in affected patients. This may be the result of problems relating to fertilisation (due to blockage of the sperm's path), implantation, or pregnancy maintenance. The type of abnormality will guide the approach to treatment. Many assisted reproductive techniques are now available.

Sexual function can be affected in a couple of ways. The most obvious is in the situation of a complete absence of the vagina. In this circumstance, normal intercourse would be impossible and creation of a neovagina may be appropriate. In the case of both a transverse and longitudinal septum, a physical barrier may make intercourse difficult, painful, or even impossible, as the calibre and length of the vagina may be altered.⁴

After consideration of the above, a decision can be made by both the patient and physician as to how to proceed. In cases of minor abnormalities, such as a uterine partial septum or even a complete septum, hysteroscopic resection can be an appropriate choice, particularly if fertility is an issue. The advantages of hysteroscopy include the shorter duration of surgery, smaller blood loss, lower costs, reduced morbidity, and shorter hospital stay compared with abdominal surgery.^{1,3} In select women with RPL or preterm delivery, uterine reconstruction with Strassman metroplasty achieves unification of two endometrial cavities in a divided uterus and is associated with a live birth rate >80%.⁴

Attempts to unify a double cervix or a septate cervix are not recommended because of the possibility of causing cervical incompetence. However, a double or septate cervix can adversely affect the outcome of delivery if vaginal delivery is attempted, and delivery should be by caesarean section if it appears that the cervix will cause dystocia.¹²

Vaginal septum, if the patient is symptomatic, can usually be treated with a simple resection if small.⁴

In cases of absence of the vagina, surgical and nonsurgical methods can be used to create a neovagina.¹³ An appropriate choice in this circumstance depends on the presence or absence of a uterus. If the uterus is present, then creation of a neovagina, along with a communication to the cervix, would be important if menstrual abnormalities exist. If these do not exist, or if there is no uterus, then nonsurgical methods should be employed initially.¹³ The nonsurgical approach entails the use of subsequently larger vaginal dilators to stretch the area where the vagina is to be created.

Multiple surgical procedures have been described. The McIndoe surgical procedure for vaginal agenesis is the most well known. A space is dissected between the rectum and the bladder, and a split-thickness skin graft from the buttocks is used to form the vagina; a specially crafted dilator at the time of the procedure creates continuous dilatation of the vagina while the graft heals. Other procedures include Williams vulvovaginoplasty, musculocutaneous flaps, and free intestinal grafts, which are easier and with better results due to the absence of stenosis and need for lubrication.¹² The disadvantage of the intestinal conduits can be excess mucus production and the magnitude of surgery. The decision of which approach to take is dictated by the patient's characteristics and needs.

Given the major advances in infertility treatments, it is possible to perform an *in vitro* fertilisation (IVF) cycle through the myometrial wall. Therefore, a direct connection of the uterine cavity to the vagina through the cervix may not be an issue when considering fertility problems. When fertility is not an issue and the patient is suffering from menstrual problems, hysterectomy can be a consideration.¹⁴

Laparoscopy should be used to excise obstructed, rudimentary uterine horns and adjacent tubes in patients with a unicornuate uterus. It should also be used for hysterectomy in cases of cervical agenesis and in neovaginoplasty procedures in cases of vaginal agenesis. In many women, the malformation results in obstructed and retrograde menstruation, thereby facilitating the development of endometriosis. During laparoscopy, this diagnosis may be confirmed and the endometrial foci may be resected.¹

Postoperative Complication

The complication of pelvic infection can be prevented by administering broad spectrum antibiotics intra- and postoperatively. Haemorrhage may occur if the sutures are not properly applied; therefore, meticulous application of sutures intraoperatively is very important.¹²

Prognosis and Outcome

A good prognosis with improvement in reproductive performance subsequently has been documented.^{15,16} The pregnancy outcome has improved following unification procedures. Other causes of infertility have to be excluded by investigation, however, before embarking on this procedure.¹⁶

Ethical Issues

In Africa, genital surgery is a taboo for some patients and their families. Delayed presentation is very common, and refusal of treatment is common in some communities. One of the major challenges is convincing the patients and their families to perform vaginal dilatations; that is why the choice of surgery depends mainly on social conditions.

Evidence-Based Research

Table 115.2 presents a randomised study of two different techniques in surgical excision of a uterine septum.

Table 115.2: Evidence-based research.

Title	Small-diameter hysteroscopy with Versapoint versus resectoscopy with a unipolar knife for the treatment of septate uterus: a prospective randomized study
Authors	Colacurci N, De Franciscis P, Mollo A, Litta P, Perino A, Cobellis A
Institution	Department of Gynecology, Obstetrics and Reproductive Sciences, Second University of Naples, Naples, Italy
Reference	J Minim Invasive Gynecology 2007; 14(5):622–627
Problem	Role of two different techniques of metroplasties were analysed in surgical excision of a uterine septum.
Intervention	Small diameter hysteroscope with Versapoint™, resectoscope with unipolar knife.
Comparison/control (quality of evidence)	Two groups of patients. In group A (n = 80), a 26F hysteroscope with unipolar knife was used for excision of the uterine septum. In group B (n = 80), a 5-mm hysteroscope with Versapoint device was used. The study was conducted in 2001–2005.

Outcome/ effect	Operative time and fluid absorption were significantly lower in group A (23.4±5.7 vs 16.9±4.7). The complication rate was significantly lower in group B than in group A. No difference in any of the reproductive parameters was observed between the two groups: pregnancy and delivery rates were 70% and 81.6%, respectively, in group A versus 76.9% and 84%, respectively, in group B. Nine women (18.4%) from group A and 8 women (16%) from group B experienced spontaneous abortions.
Historical significance/ comments	This report randomly compared two hysteroscopic techniques in performing metroplasties for a large group of women having uterine septa. The study used a variety of parameters to compare the two groups: operative parameters were operative time, fluid absorption, complications, and need for second intervention; reproductive outcome parameters were pregnancy, abortion, term and preterm delivery, modality of delivery, and cervical cerclage. Most patients (54/82) delivered by caesarean section without differences according to the hysteroscopic technique used for metroplasty (65% in group A vs 67.7% in group B) or to the gestational age (65.1% of term and 68.7% of preterm deliveries). Small-diameter hysteroscopy with bipolar electrode for the incision of the uterine septum is as effective as resectoscopy with a unipolar electrode regarding reproductive outcome and is associated with a shorter operating time and a lower complication rate.

Key Summary Points

- Müllerian anomalies are congenital defects of the female reproductive tract resulting from failure in the development of the Müllerian ducts and their associated structures. Their cause has yet to be fully clarified; it is currently believed to be multifactorial.
- The embryological development of the female reproductive system is closely related to the development of the urinary system, and anomalies in both systems may occur in up to 25% of these patients. Other associated malformations may affect the gastrointestinal tract (12%) or musculoskeletal system (10–12%).
- Müllerian anomalies are frequently asymptomatic and are often missed in routine gynecological examinations. Nevertheless, a history of pelvic pain following menarche, dysmenorrhea, and an increase in abdominal volume are complaints suggestive of uterine anomalies. In addition, primary amenorrhea and changes to menstrual flows may be present.
- Due to the complexity of presentations, diagnosing Müllerian malformations requires the use of more than one imaging method in 62% of the cases.
- The treatment for Müllerian anomalies varies according to the specific type of malformation found in each patient.
- Müllerian anomalies consist of a wide range of defects that may vary from patient to patient. Therefore, their management must also be individual, taking anatomical and clinical characteristics into consideration, as well as the patient's preference.

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