Chapter 25
Surgery for Anomalies of the Müllerian Ducts

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DEFINITIONS

Hematometra—The distention of the uterus with blood or menstrual fluid.

Hematometrocolpos—The distention of the uterus and vagina with blood or menstrual fluid; because the vaginal wall is more distensible, the vagina will preferentially fill before the uterus.

Hydrocolpos—The distention of the vagina with fluid; often seen in infants with complex reproductive anomalies.

Metroplasty—Uterine reconstructive procedure.

Uterine anlagen—An underdeveloped uterine structure that is a remnant of a single embryologic müllerian duct.

Maldevelopment of the müllerian ducts occurs in a variety of forms, and each anomaly is distinctive. Nevertheless, some generalizations can be made. Classifications of vaginal anomalies based on certain anatomic findings are useful in organizing the type of malformation, but there usually are exceptions to each rule. Thus, what appears, after a preliminary diagnostic evaluation, to be an apparently isolated vaginal malformation may be found later to be associated with a uterine or renal anomaly. A comprehensive preoperative evaluation of patients with suspected malformations of the müllerian ducts is essential, but a clear understanding of the particular anomaly may not be established until the time of surgical correction. Reproductive surgeons must therefore be equally skilled in both uterine and vaginal reconstructions.

The patient with a uterovaginal anomaly often relies entirely on her physician to clarify the reproductive consequences associated with her diagnosis. The physician can help to allay her anxieties by making a prompt evaluation and giving a full and accurate description of the reproductive implications or the obstetric consequences of her particular uterovaginal anomaly.

CLASSIFICATION OF UTEROVAGINAL ANOMALIES

Classifications of uterovaginal anomalies originally were organized on the basis of clinical findings. Our improved understanding of the embryologic development of most uterovaginal anomalies has enabled categorization on this basis. The 1988 American Fertility Society (AFS) classification of müllerian anomalies (Table 25.1) offers an alternative based on the degree of failure of normal uterine development. Anomalies are grouped according to similarities of clinical manifestations, treatment, and prognosis for fetal salvage. The AFS classification system is weighted primarily toward disorders of lateral fusion and does not include associated vaginal anomalies, although the scheme does allow the user to describe anomalies involving the vagina, tubes, and urinary tract as associated malformations.

No classification of müllerian maldevelopment can focus entirely on the uterus; the vagina is often involved, and sometimes the tubes are involved as well. This discussion follows a suggested modification of the AFS classification of uterovaginal anomalies (Table 25.2) that comprises four groups based on embryologic considerations.

TABLE 25.1 American Fertility Society Classification of Müllerian Anomalies[a]

Class I. Segmental, müllerian agenesis-hypoplasia	
	A. Vaginal
	B. Cervical
	C. Fundal
	D. Tubal
	E. Combined anomalies
Class II. Unicornuate	
	A. Communicating
	B. Noncommunicating
	C. No cavity
	D. No horn
Class III. Didelphys	
Class IV. Bicornuate	
	A. Complete (division down to internal os)
	B. Partial
Class V. Septate	
	A. Complete (septum to internal os)

B. Partial

Class VI. Arcuate

Class VII. Diethylstilbestrol related

[a]This classification allows the user to indicate the malformation type and provides additional findings to describe associated variations involving the vagina, cervix, tubes (right, left), and kidneys (right, left).
Adapted from the American Fertility Society. The American Fertility Society classifications of adnexal adhesions, distal tubal occlusion, tubal occlusion secondary to tubal ligation, tubal pregnancies, müllerian anomalies and intrauterine adhesions. *Fertil Steril* 1988;49:944, with permission. Copyright © 1988, Elsevier.

TABLE 25.2 American Fertility Society Classification of Uterovaginal Anomalies

Class I. Dysgenesis of the müllerian ducts

Class II. Disorders of vertical fusion of the müllerian ducts

- A. Transverse vaginal septum
  - 1. Obstructed
  - 2. Unobstructed
- B. Cervical agenesis or dysgenesis

Class III. Disorders of lateral fusion of the müllerian ducts

- A. Asymmetric-obstructed disorder of uterus or vagina usually associated with ipsilateral renal agenesis
  - 1. Unicornuate uterus with a noncommunicating rudimentary anlage or horn
  - 2. Unilateral obstruction of a cavity of a double uterus
  - 3. Unilateral vaginal obstruction associated with double uterus
- B. Symmetric-unobstructed
  - 1. Didelphic uterus
    - a. Complete longitudinal vaginal septum
    - b. Partial longitudinal vaginal septum
    - c. No longitudinal vaginal septum
  - 2. Septate uterus
    - a. Complete
      - 1) Complete longitudinal vaginal septum
      - 2) Partial longitudinal vaginal septum
      - 3) No longitudinal vaginal septum
    - b. Partial
      - 1) Complete longitudinal vaginal septum
      - 2) Partial longitudinal vaginal septum
      - 3) No longitudinal vaginal septum
  - 3. Bicornuate uterus
    - a. Complete
      - 1) Complete longitudinal vaginal septum

2) Partial longitudinal vaginal septum

3) No longitudinal vaginal septum

b. Partial

1) Complete longitudinal vaginal septum

2) Partial longitudinal vaginal septum

3) No longitudinal vaginal septum

4. T-shaped uterine cavity (diethylstilbestrol related)

5. Unicornuate uterus

a. With a rudimentary horn

1) With endometrial cavity

a) Communicating

b) Noncommunicating

2) Without endometrial cavity

b. Without a rudimentary horn

Class IV. Unusual configurations of vertical-lateral fusion defects

Modified from the American Fertility Society. The American Fertility Society classifications of adnexal adhesions, distal tubal occlusion, tubal occlusion secondary to tubal ligation, tubal pregnancies, müllerian anomalies and intrauterine adhesions. *Fertil Steril* 1988;49:944.

Class I: Dysgenesis of the Müllerian Ducts

Dysgenesis of the müllerian ducts, which includes agenesis of the uterus and vagina (the Mayer-Rokitansky-Küster-Hauser [MRKH] syndrome), is an impairment of the reproductive system characterized by no reproductive potential other than that achieved by in vitro fertilization in a host uterus.

Class II: Disorders of Vertical Fusion of the Müllerian Ducts

Disorders of vertical fusion can be considered to represent faults in the junction between the downgrowing müllerian ducts (müllerian tubercle) and the upgrowing derivative of the urogenital sinus. Typically, these disorders are characterized by an atretic portion of the vagina that can be quite thick, extending through more than half the distance of the vagina, or it can be quite thin and limited to a small obstructing membrane.

Regardless of the length of the septum, a disorder of vertical fusion should be regarded as a transverse vaginal septum and classified as either obstructed or unobstructed. The socalled partial vaginal agenesis with uterus and cervix present is probably a misnomer for a large segment of atretic vagina. Cervical agenesis or dysgenesis is also included in the group of disorders of vertical fusion.

Class III: Disorders of Lateral Fusion of the Müllerian Ducts

Disorders of lateral fusion of the two müllerian ducts can be symmetric-unobstructed, as with the double vagina, or asymmetric-obstructed, as with unilateral vaginal obstruction. Obstructions associated with disorders of lateral fusion are particularly noteworthy in that they are observed clinically only as unilateral obstructions that almost invariably are associated with absence of the ipsilateral kidney. Bilateral obstruction is thought to be associated with bilateral kidney agenesis and subsequent nonviability of the developing embryo.

The three varieties of asymmetric obstruction with ipsilateral renal agenesis are:

1. Unicornuate uterus with a noncommunicating horn that contains menstruating endometrium
2. Unilateral obstruction of a cavity of a double uterus
3. Unilateral vaginal obstruction

The five groups of symmetric-unobstructed disorders of lateral fusion are:

1. The didelphic uterus
2. The septate uterus
3. The bicornuate uterus
4. The T-shaped uterine cavity, which may be hypoplastic and irregular and which is associated with diethylstilbestrol (DES) exposure in utero
5. The unicornuate uterus with or without a rudimentary horn

The first three groups are types of double uteri; differentiation between a septate uterus (second group) and a bicornuate uterus (third group) requires visualization of the fundus. The septum within the septate uterus is complete or partial. When the septum is complete, that inevitably involves the cervical region with a longitudinal vaginal septum that can extend to the introitus or partially down the vagina. The bicornuate uterus also can have a partial or almost complete separation of the uterine cavities. The term *arcuate uterus* is used primarily by radiologists to refer to a slight septum in the uterine fundus that forms no clear separation of the uterine cavities. This type of uterus is usually included in the category of partial septate uterus.

The unicornuate uterus may have an attached horn with a cavity that communicates with the unicornuate uterus, or there may be no uterine horn or a uterine horn with no cavity.

Some debate has focused on whether the unicornuate uterus with a communicating horn can represent a hypoplastic side of a bicornuate uterus.

Class IV: Unusual Configurations of Vertical-Lateral Fusion Defects

This final category includes combinations of uterovaginal anomalies and other disorders. Unusual uterovaginal configurations have been described that do not fit a particular category, and vertical and lateral fusion disorders can coexist.

Unusual configurations of vertical-lateral fusion defects can be seen with abnormalities of the lower urinary tract. Singh and coworkers have described a patient who was noted to have a persistent hymen and a longitudinal vaginal septum with a didelphic uterus. The patient was noted also to have a double urethra and bladder and left renal agenesis.

Obstructive lesions require immediate attention to relieve retrograde flow of trapped mucus and menstrual blood and increasing pressure on surrounding organs and structures. When no obstruction is present, attention may not be required immediately, but it will always be required eventually to establish or improve reproductive or coital function.

EMBRYOLOGY

The reproductive organs in the female (and in the male) consist of external genitalia, gonads, and an internal duct system between the two. These three components originate embryologically from different primordia and in close association with the urinary system and hindgut. Thus, the developmental history is complex (**Figs. 25.1** and **25.2**). Even in the 3.5- to 4-mm embryo, it is possible to recognize the bilateral thickenings of the coelomic epithelium known as the gonadal ridges medial to the mesonephros (primitive kidney) in the dorsum of the coelomic cavity. At approximately the 6th week of gestation, in the 17- to 20-mm embryo, the gonad can be distinguished as either a testis or an ovary.

In the female, the labia minora and majora develop from the labioscrotal folds, which are ectodermal in origin. The phallic portion of the urogenital sinus gives rise to the urethra. The müllerian (paramesonephric) duct system is stimulated to develop preferentially over the wolffian (mesonephric) duct system, which regresses in early female fetal life. The cranial parts of the wolffian ducts can persist as the epoöphoron of the ovarian hilum; the caudal parts can persist as Gartner ducts. The müllerian ducts persist and attain complete development to form the fallopian tubes, the uterine corpus and cervix, and a portion of the vagina.

Origin of the Müllerian Ducts

Approximately 37 days after fertilization, the müllerian ducts first appear lateral to each wolffian duct as invaginations of the dorsal coelomic epithelium. The site of origin of the invaginations remains open and ultimately forms the fimbriated ends of the fallopian tubes. At their point of origin, each of the müllerian ducts forms a solid bud. Each bud penetrates the mesenchyme lateral and parallel to each wolffian duct. As the solid buds elongate, a lumen appears in the cranial part, beginning at each coelomic opening. The lumina extend gradually to the caudal growing tips of the ducts.

Eventually, the caudal end of each müllerian duct crosses the ventral aspect of the wolffian duct. The paired müllerian ducts continue to grow in a medial and caudal direction until they eventually meet in the midline and become fused together in the urogenital septum. A septum between the two müllerian ducts gradually disappears, leaving a single uterovaginal canal lined with cuboidal epithelium. Failure of reabsorption of this septum can result in a septate uterus. The most cranial parts of the müllerian ducts remain separate and form the fallopian tubes. The caudal segments of the müllerian ducts fuse to form the uterus and part of the vagina. The cranial point of fusion is the site of the future fundus of the uterus. Variations in this site of fusion can result in an arcuate or bicornuate uterus. Complete failure of fusion can result in a didelphic uterus.

Isolated case reports continue to challenge established embryologic mechanisms of müllerian development. Dunn and Hantes reported a case of a double cervix and vagina with a blind cervical pouch challenging the theory of unidirectional fusion. Engmann and colleagues reported a unicornuate uterus with normal external uterine morphology, with bilateral fallopian tubes; however, only the right fallopian tube communicated with the uterine cavity. The patient suffered from pain because of the obstruction egress of the stimulated endometrial tissue. The patient was treated with removal of the obstructed cavity. The authors propose that this may represent failure of canalization of one of the müllerian ducts. Additional reports are necessary to fully evaluate potential variations in embryologic development.

Development of the Vagina

The vagina is formed from the lower end of the uterovaginal canal, which developed from the müllerian ducts and the urogenital sinus (**Fig. 25.2**). The point of contact between the two is the müllerian tubercle. A solid vaginal cord results from proliferation of the cells at the caudal tip of the fused müllerian ducts. The cord gradually elongates to meet the bilateral endodermal evaginations (sinovaginal bulbs) from the posterior aspect of the urogenital sinus below. These sinovaginal bulbs extend cranially to fuse with the caudal end of the vaginal cord, forming the vaginal plate. Subsequent canalization of the vaginal cord occurs, followed by epithelialization with cells derived mostly from endoderm of the urogenital sinus. Recent proposals hold that only the upper one third of the vagina is formed from the müllerian ducts and that the lower vagina develops from the vaginal plate of the urogenital sinus. Recent studies also suggest that the vaginal canal is actually open and connected to a patent uterus and tubes, even in early embryonic life, and that the vagina does not form and later become canalized from an epithelial cord of squamous cells growing upward from the urogenital sinus. Most investigators now suggest that the vagina develops under the influence of the müllerian ducts and estrogenic stimulation. There is general agreement that the vagina is a composite formed partly from the müllerian ducts and partly from the urogenital sinus.

At approximately the 20th week, the cervix takes form as a result of condensation of stromal cells at a specific site around the fused müllerian ducts. The mesenchyme surrounding the müllerian ducts becomes condensed early in embryonic development and eventually forms the musculature of the female genital tract. The hymen is the embryologic septum between the sinovaginal bulbs above and the urogenital sinus proper below. It is lined by an internal layer of vaginal epithelium and an external layer of epithelium derived from the urogenital sinus (both of endodermal origin), with mesoderm between the two. It is not derived from the müllerian ducts.

Anomalies in Organogenesis of the Vagina

Anomalies in the organogenesis of the vagina are easily understood. If there is failure in the development of the müllerian

ducts at any time between their origin from the coelomic epithelium at 5 weeks of embryonic age and their fusion with the urogenital sinus at 8 weeks, the sinovaginal bulbs will fail to proliferate from the urogenital sinus, and the uterus and vagina will fail to develop. Congenital absence of the uterus and the vagina, known as the MRKH syndrome, is the most common clinical example of this anomaly.

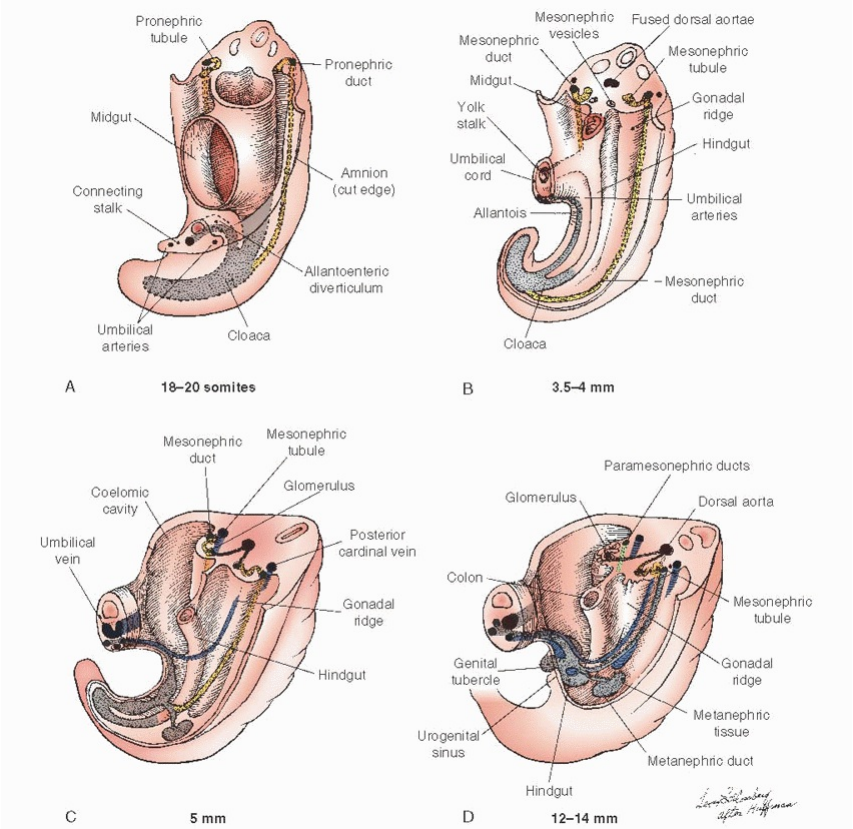


FIGURE 25.1 Diagrammatic representation of the development of the female reproductive organs and structures in early embryogenesis. **A:** At the 18- to 20-somite stage (4th week), the gonadal ridges have not yet begun to form. **B:** In the 3.5- to 4-mm embryo (5th week), the gonadal ridges can be recognized as thickenings of the coelomic cavity just medial to the mesonephric tubules. (Gonadal differentiation into either testis or ovary does not occur until the 6th week of development.) The allantoenteric diverticulum is joined caudally to the dilated cloaca. **C and D:** The genital tubercle and labial folds form in the region just anterior to the cloaca. The cloaca later divides into the ventral urogenital sinus and the dorsal rectum. The development of the urinary system closely parallels that of the reproductive system. The nonfunctioning pronephric tubules shown in **(A)** develop to form the mesonephric ducts shown in **(B)** and **(C).** The permanent kidneys eventually develop from the metanephric tissue, and the urinary collecting system develops from the metanephric ducts. The paramesonephric (müllerian) ducts are apparent by the 12- to 14-mm stage **(D).** (Their subsequent development is illustrated in **Figure 25.2.**)

Transverse Vaginal Septum

A transverse vaginal septum can develop at any location in the vagina but is more common in the upper vagina at the point of junction between the vaginal plate and the caudal end of the fused müllerian ducts. This defect presumably is caused by failure of absorption of the tissue that separates the two or by

failure of complete fusion of the two embryologic components of the vagina. A large segment of vagina can be atretic. In past reviews, this has been termed *partial vaginal agenesis with a uterus present.* Elucidation of the cause of a high transverse vaginal septum is more difficult. A local abnormality of the vaginal mesoderm or failure of canalization of the epithelial vaginal plate can provide the answer, but why the abnormality should occur at this particular site is not evident. The proportion of the vagina originating from the urogenital sinus can at times be considerably more than one fifth, and a high transverse vaginal septum thus may represent the junction of an abnormally long urogenital sinus contribution and a short müllerian portion.

Alternatively, the high transverse septum could be the sequela of a local infection of the septum at the end of the vagina. Septa in other areas of the vagina are unexplained by this theory, which has not gained widespread acceptance.

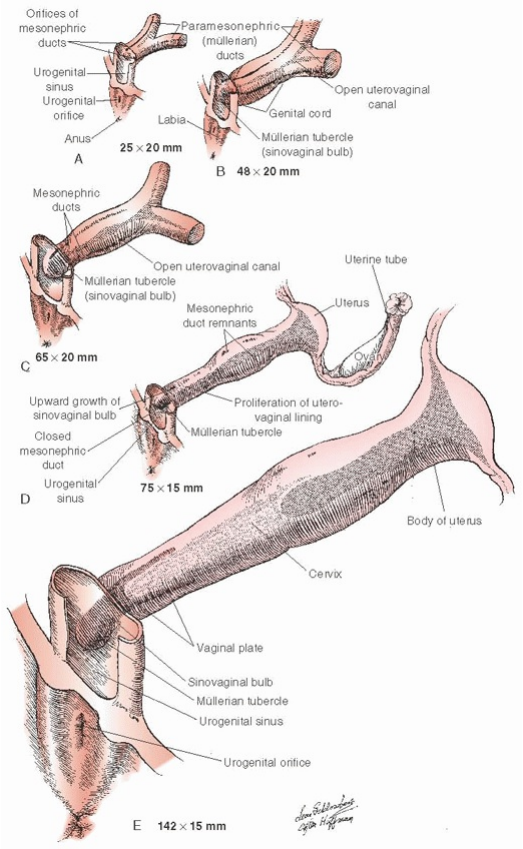


FIGURE 25.2 Further development of the paramesonephric (müllerian) ducts and the urogenital sinus. **A:** Early development of the paramesonephric ducts. The cranial ends of the paramesonephric ducts develop first. These ends remain open to form the fimbriated ends of the fallopian tubes. The paramesonephric ducts grow caudally and cross the mesonephric ducts ventrally. **B:** Eventually, they fuse together to form the uterovaginal canal. **C:** Further caudal development brings this structure into contact with the wall of the urogenital sinus, producing the müllerian tubercle. The caudal ends of the fused paramesonephric ducts form the uterine corpus and cervix. Together with the urogenital sinus, they also form the vagina. The cranial point of fusion of the paramesonephric ducts marks the location of the future uterine fundus. The fallopian tubes form from the unfused cranial parts of the paramesonephric (müllerian) ducts. The proliferation of the lining of the uterovaginal canal above the upward growth of the sinovaginal bulb from below **(D)** forms the vaginal plate **(E),** which later becomes canalized to leave an open vaginal canal. Thus, the vagina is of composite origin. The mesonephric ducts in the female degenerate but can persist into adult life as Gartner ducts.

Disorders of Ineffective Suppression of Müllerian Ducts

When abnormal gonadal development is caused by ineffective suppression of the müllerian ducts, ambiguous external genitalia frequently are accompanied by a small rudimentary uterus or a partially developed vagina. Additionally, when there is a genetic loss of cytoplasmic receptor proteins within androgenic target cells, such as occurs in the androgen insensitivity syndrome (formerly called testicular feminization syndrome), the vagina is incompletely developed because the existing male gonads suppress the development of the müllerian ducts. Because these genetically XY patients have phenotypic female genital anatomy without a completely formed vagina, it is important that a vagina be nonsurgically (dilatation) or surgically created to ensure a satisfactory sexual experience.

Congenital anorectal malformations (imperforate anus with rectoperineal or rectovestibular fistula or more complex anomalies like cloaca or cloacal exstrophy) have been reported to occur with reproductive anomalies. These anomalies can be associated with maldevelopment of the müllerian and mesonephric duct derivatives.

Müllerian Duct Abnormalities

Abnormalities in the formation or fusion of the müllerian ducts can result in a variety of anomalies of the uterus and vagina: single, multiple, combined, or separate. Just as the entirely separate origin of the ovaries from the gonadal ridges accounts for the infrequent association of uterovaginal anomalies with ovarian anomalies (see this chapter), so do the close developmental relationships of the müllerian and wolffian ducts explain the frequency with which anomalies of the female genital system and urinary tract are associated. Failure of development of a müllerian duct is likewise associated with failure of development of a ureteric bud from the caudal end of the wolffian duct. Thus, the entire kidney can be absent on the side ipsilateral to the agenesis of a müllerian duct.

Depending on the timing of the teratogenic influence, renal units can be absent, fused, or in unusual locations in the pelvis. Ureters can be duplicated or can open in unusual places, such as the vagina or uterus. Jones and Rock have noted that failure of lateral fusion of the müllerian ducts with unilateral obstruction is associated consistently with absence of the kidney on the side with obstruction. Bilateral obstruction has not been observed clinically, presumably because it would be associated with bilateral renal agenesis, a condition that would not allow the embryo to develop. According to Thompson and Lynn, 40% of female patients with congenital absence of the kidney are found to have associated genital anomalies.

Much investigation has been undertaken to determine a genetic relationship in the development of disorders of the müllerian ducts. Familial aggregates of the most common disorders of the müllerian differentiation are best explained on the basis of polygenic or multifactorial inheritance. No information exists on the number and chromosomal location of responsible genes. Single mutant genes are responsible for the McKusick-Kaufman syndrome and the hand-foot-genital syndrome. Hand-foot-genital syndrome is a rare, dominantly inherited condition that affects both the distal limbs and the genitourinary tract. A nonsense mutation of the HOXA13 gene has been identified in several families. HOX gene mutations have been reported in several families with multiple müllerian abnormalities. Genital malformations may also be associated with heterozygous DNA sequence variations of the HOXA10, HOXA11, and HOXA13 genes. To date, involvement of the Y chromosome in the pathogenesis of müllerian anomalies has not been considered. The 2004 report by Plevraki and colleagues suggests the possible role of testis-specific protein 1-Y gene in patients with uni- or bilateral gonadal agenesis and uterovaginal dysgenesis. Timmreck and colleagues narrowed the genetic considerations by noting that in an evaluation of 40 women with developmental abnormalities of the uterus and vagina and 12 normal controls, no mutations of the WNT7A gene—a gene associated with murine Müllerian duct development—were found. Reproductive abnormalities involving the uterus and vagina may also be associated with other more complex malformation syndromes in which the molecular basis of many of the syndromes remains unknown.

CONGENITAL ABSENCE OF THE MÜLLERIAN DUCTS

The disorders of müllerian agenesis include congenital absence of the vagina and uterus. Often referred to in the literature simply as congenital absence of the vagina (vaginal

agenesis), this condition is more accurately labeled aplasia (or dysplasia) of the müllerian ducts because the lower vagina generally is normal, but the middle and upper two thirds are missing. Despite the absence of the uterus, rudimentary uterine primordia are found that are comparable to each other in size and appearance. Tubes and ovaries in patients with congenital absence of the müllerian ducts generally are normal. The syndrome, usually referred to as the MRKH syndrome, is associated with a heterogeneous group of disorders that have a variety of genetic, endocrine, and metabolic manifestations and associated anomalies of other body systems.

Characteristics of Women with Müllerian Agenesis

- Congenital absence of the uterus and vagina (small rudimentary uterine bulbs are usually present with rudimentary fallopian tubes)
- Normal ovarian function, including ovulation
- Sex of rearing: female
- Phenotypic sex: female (normal development of breasts, body proportions, hair distribution, and external genitalia)
- Genetic sex: female (46,XX karyotype)
- Frequent association of other congenital anomalies (skeletal, urologic, and especially renal)

Partial agenesis of the vagina with the uterus present and a transverse vaginal septum both are categorized as disorders of vertical fusion. These two disorders have a low incidence of associated urinary tract anomalies, another circumstance that sets them apart from the MRKH syndrome.

Realdus Columbus first described congenital absence of the vagina in 1559. In 1829, Mayer described congenital absence of the vagina as one of the abnormalities found in stillborn infants with multiple birth defects. Rokitansky in 1838 and Küster in 1910 described an entity in which the vagina was absent, a small bipartite uterus was present, the ovaries were normal, and anomalies of other organ systems (renal and skeletal) were frequently observed. Hauser and associates emphasized the spectrum of associated anomalies. Pinsky suggested that congenital absence of the vagina is part of a symptom

complex and not a true syndrome. Over the years, the disorder has come to be known as the MRKH syndrome, the Rokitansky-Küster-Hauser syndrome, or simply the Rokitansky syndrome (Fig. 25.3). Counseller found that the condition occurred once in 4,000 female admissions to the Mayo Clinic. Evans estimated that vaginal agenesis occurred once in 10,588 female births in Michigan from 1953 to 1957.

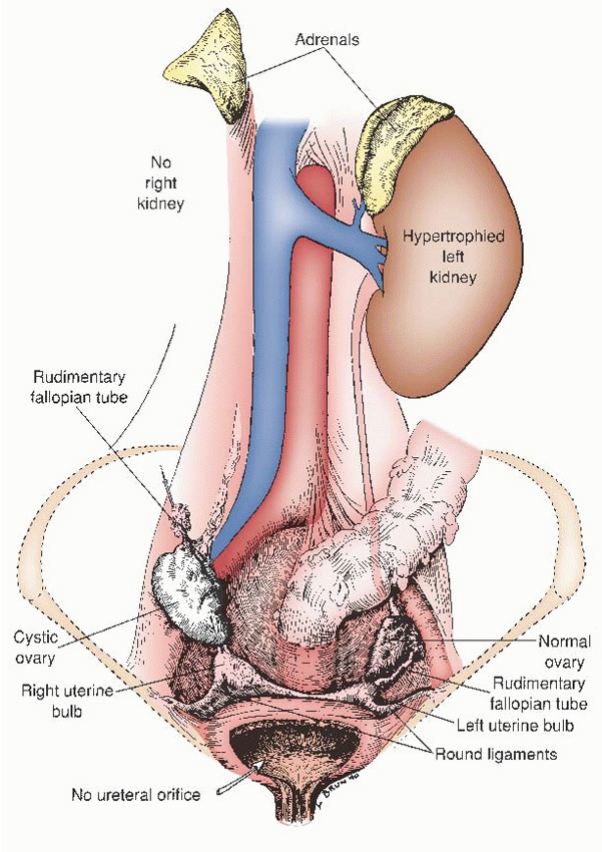


FIGURE 25.3 Typical findings in a patient with MRKH syndrome. Note the absence of the right kidney and right ureteral orifice. The uterus is represented by bilateral rudimentary uterine bulbs joined by a band behind the bladder. The ovaries appear normal although there is malposition of the right ovary.

Individuals with an absent vagina and the classic MRKH syndrome usually are first seen by a gynecologist at age 14 to 15 years, when the absence of menses causes concern. Such young women have a normal complement of chromosomes (46,XX) and usually have normal ovaries and secondary sex characteristics, including external genitalia. Menstruation does not appear at the usual age because the uterus is absent, but ovulation occurs regularly. There are some exceptions to the rule of normal ovaries. For example, polycystic ovaries and gonadal dysgenesis have been reported in patients with congenital absence of the vagina. Plevraki and colleagues reinforced the importance of consideration of such conditions, as one of the six women with MRKH evaluated over a 12-month period demonstrated hypergonadotropic hypogonadism that was due to the bilateral absence of gonadal tissue. Additionally, nested polymerase chain reaction demonstrated the presence of testisspecific protein 1-Y-linked (TSPY) gene in two women.

Etiologic Factors

An exclusively genetic etiology cannot be ascribed to vaginal agenesis because almost all patients have a normal karyotype (46,XX) and because the discordance of vaginal agenesis in three sets of monozygotic twins has been reported. The occurrence of complete vaginal agenesis in sisters with a 46,XX karyotype suggests an autosomal mode of inheritance for these patients. Shokeir investigated the families of 13 unrelated females with aplasia of müllerian duct derivatives. Similarly affected females were found in 10 families. Usually, there was an affected female paternal relative, suggesting female-limited autosomal dominant inheritance of a mutant gene transmitted by male relatives.

Other investigators point to the variety of associated anomalies as support for the etiologic concept of variable expression of a genetic defect possibly precipitated by teratogenic exposure between the 37th and the 41st gestational day, the time during which the vagina is formed. Knab has suggested five possible etiologic factors of the MRKH syndrome:

1. Inappropriate production of müllerian regressive factor in the female embryonic gonad

2. Regional absence or deficiency of estrogen receptors limited to the lower müllerian duct

3. Arrest of müllerian duct development by a teratogenic agent
4. Mesenchymal inductive defect
5. Sporadic gene mutation

Knab believes that the teratogenic and the mutant gene etiologies are the most probable.

Anomalies Associated with Müllerian Agenesis

Many patients with müllerian agenesis have associated anomalies of the upper müllerian duct system together with associated anomalies of other organ systems. By gentle rectal examination, the physician can feel an absence of the midline müllerian structure that should represent the uterus. The physician instead feels a smooth band (possibly a remnant of the uterosacral ligaments) that extends from one side of the pelvis to the other. In MRKH syndrome, the uterus is represented by bilateral rudimentary uterine bulbs that vary in size, are not usually palpable, are connected to small fallopian tubes, and are located on the lateral pelvic side wall adjacent to normal ovaries. Depending on their size, these rudimentary uterine bulbs may or may not contain a cavity lined by endometrial tissue (Fig. 25.4). If present, the endometrial tissue can appear immature or, rarely, can show evidence of cyclic response to ovarian hormones. The endometrial cavity does not communicate often with the peritoneal cavity because the tube may not be patent at the point of junction between the tube and the rudimentary uterine bulb. In rare instances, however, active endometrium can exist within the uterine anlagen and the endometrial cavity, enabling communication with the peritoneal cavity through patent fallopian tubes. Reports have described several patients with functioning endometrial tissue in one or both rudimentary uterine bulbs (Fig. 25.4B). The patient can develop hematometra because of cyclic accumulation of trapped blood. Cyclic abdominal pain is relieved by excision of the active uterine anlagen. A patient with MRKH syndrome was reported who had a 4-cm endometrioma removed from the left ovary by laparotomy at the time of operation to create a vagina. Myomas have been reported to form in the muscular wall of inactive uterine anlagen, and dysmenorrhea has been attributed to their presence. A small myoma has been found, in addition to the tube and ovary, in the inguinal canal and in the inguinal hernia sac. Due to the rarity of the condition, interaction with the radiologist about the patient's history is important to make an accurate diagnosis.

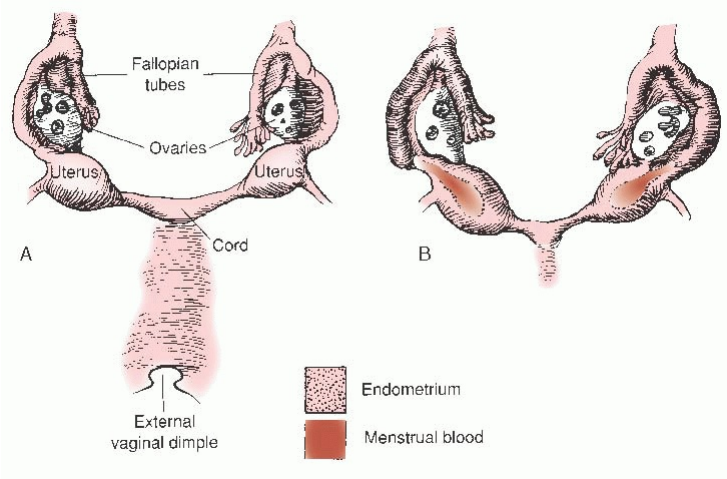


FIGURE 25.4 Patients with congenital absence of the vagina can show variation in the development of the upper müllerian ducts. **A:** Bilateral rudimentary uterine bulbs without endometrium. **B:** Bilateral rudimentary uterine bulbs containing a cavity lined with functioning endometrial tissue. Crosssectional view shows presence of menstrual blood.

Chakravarty and colleagues and Singh and Devi have demonstrated that the rudimentary bulbs have the potential for function. These authors used these rudimentary uterine bulbs to reconstruct a midline uterus. The reconstructed uterus was then connected to a newly constructed vagina. A surprising number of patients who have undergone this procedure have experienced cyclic menstruation, although recurrent stenosis and obstruction of the rudimentary horns are the most common results of such efforts. The authors of this chapter have had no experience with this technique and question its usefulness. In the majority of cases, these rudimentary uterine bulbs usually are insignificant structures that cause no problems.

Associated Urologic and Renal Anomalies

Fore and associates reported that 47% of patients in whom evaluation of the urinary tract was performed had associated urologic anomalies. In other studies, approximately one third of patients with complete vaginal agenesis were found to have significant urinary anomalies, including unilateral renal agenesis, unilateral or bilateral pelvic kidney, horseshoe kidney, hydronephrosis, hydroureter, and a variety of patterns of ureteral duplication. A significant number of patients with partial vaginal agenesis also have associated urinary tract anomalies.

Associated Skeletal and Other Anomalies

Associated skeletal anomalies have been recognized since congenital absence of the vagina was first described. In a review of 574 reported cases, Griffin and associates found a 12% incidence of skeletal abnormalities. Most of these abnormalities involve the spine (wedge vertebrae, fusions, rudimentary vertebral bodies, and supernumerary vertebrae), but the limbs and ribs also can be involved. Other anomalies include syndactyly, absence of a digit, congenital heart disease, and inguinal hernias, although the latter are more often present in patients with androgen insensitivity syndrome than in patients with MRKH syndrome (Fig. 25.3). Consideration of cardiac anomalies is also important, as Pittock and colleagues reported a substantial incidence of cardiac defects (16%) when reviewing a group of 25 patients with MRKH at the Mayo Clinic.

The recognized association of absent vagina with imperforate anus and rectovestibular fistula will likely be diagnosed in infants and treated by the pediatric surgeon at the time of the rectal pull through (most commonly with sigmoid neovaginoplasty). However, the condition may not be noted or treatment may be deferred to young adulthood. Knowledge of this association can be important at the time of treatment as creation of an adequate neovagina with dilation may be impaired by scarring at the previous fistula site in the vestibule. Assessment of vaginal adequacy as a young adult is important as the neovagina created in childhood may need revision or augmentation.

Treatment for Disorders of Müllerian Agenesis

Pretreatment Considerations

If functioning endometrial tissue is present with the anlagen, then symptoms from cryptomenorrhea will begin shortly after female secondary sex characteristics develop. Prompt removal of the active uterine bulbs affords complete relief of symptoms.

Occasionally, older patients with the classic MRKH syndrome consult a gynecologist because of difficult or painful intercourse. The indication for operation in these patients is obvious. Of all patients, they are the most satisfied with the operative results.

Most commonly, patients aged 14 to 16 years are seen by a gynecologist because of primary amenorrhea. An examination may not have been performed by a previous physician because the patient was “too young,” but various hormonal medications may have been given with the hope that menstruation would begin. An inaccurate examination may have led to the mistaken diagnosis of imperforate hymen. Futile attempts to incise the hymen may have resulted in scarring of the apex of the vaginal dimple before a correct diagnosis of congenital absence of the vagina was finally made. In the past, it was customary to advise delaying surgery to create a vagina for these young patients until just before their marriage. More recently, it has become usual to perform the procedure when patients are 17 to 20 years old and are emotionally mature and intellectually ready to manage the potential postoperative requirements for care, including manipulation of the vaginal form or the use of dilators.

Psychological Concerns

Insufficient attention has been given to the psychological aspects of this problem. The patient with congenital absence of the vagina cannot be made into a whole person simply by creating a perineal pouch for intercourse. Establishment of sexual function is only one concern and may be the easiest problem to correct. Evans reported that 15% of his patients have real psychiatric difficulty. He and David and associates suggest that psychiatric help should be initiated before the treatment to create a neovagina. Weijenborg and ter Kuile described the effect of a group program on women with Rokitansky syndrome. The authors held group sessions conducted by a gynecologist, a female social worker, and a woman with Rokitansky syndrome. Seventeen patients participated. Three women had elected not to create a vagina, six women created a vagina by dilatation or sexual activity, and eight women had undergone a vaginoplasty. Indices of psychological distress were measured before the program, at initiation of the program, and at the last group session. The results demonstrated that women with Rokitansky syndrome felt less anxious, less depressed, and less sensitive to interpersonal contact after participation in the semistructured program. These data support the value of group interaction in patients with Rokitansky syndrome.

Learning about this anomaly, especially at a young age, is a shock and is accompanied by diminished self-esteem. Such patients can be encouraged by having their gynecologist offer appropriate psychological and medical therapy with an experienced multidisciplinary team. The gynecologist can also point out that the patient will functionally be like other young women who have had a hysterectomy because of serious pelvic disease and who have satisfied their desire to be a parent through adoption or gestational surrogacy. When receiving this diagnosis, patients and families are looking for reassurance about the future. Liao and colleagues reported on women with Rokitansky (treated with dilation, no treatment, or surgery) who completed four questionnaires assessing health-related quality of life, emotional distress, and sexual function and underwent a vaginal examination. Of the 87 eligible participants, 56 (64%) took part in the study. Thirty-six women had used dilators in the past, and 7 (who were sexually active) had undergone vaginal surgery (laparoscopic Vecchietti 4, McIndoe 1, bowel 1, skin flap vaginoplasty 1). The range of time from surgery to participation in the study was 5 to 16 years. Twelve patients had no intervention to create a neovagina. The participants reported overall better physical health and poorer overall mental health compared with normative data. Anxiety levels were higher, especially for women who had treatment to create a neovagina (dilation or surgical treatment). Vaginal length had a positive correlation with overall sexual satisfaction but was not related to overall quality of life. Kimberley and colleagues in Australia reported on quality of life and sexual experience of patients with vaginal atresia or agenesis. Seventy patients were identified, but nine were excluded who were younger than 17 years, newly diagnosed, or developmentally delayed. Thirty-four women responded with 28 actual participants in the study. Of the 20 patients who completed the sexual satisfaction questionnaire, only four were treated surgically (Sheare procedure 3, McIndoe 1) and the remaining 16 created a neovagina with dilation. The quality-of-life outcomes as measured by the World Health Organization Quality of Life (WHOQOL-BREF) questionnaire showed overall quality of life to be comparable to that of the average Australian population. In addition to quality-oflife evaluation, psychosocial assessment is important as well. There was a strong correlation between the quality-of-life scores and sexual satisfaction, highlighting the importance of psychological and psychosocial supports in the management of young women affected by the condition. Time since diagnosis had a positive influence on overall sexual satisfaction, with 92% of women who received a diagnosis more than 5 years ago demonstrating satisfactory sexual function scores. Callens and her colleagues in the Belgian-Dutch Study Group on Disorder of Sex Development (DSD) attempted to evaluate the psychosexual and anatomical outcomes of patients after dilation or surgery to create a neovagina; however, only 7 of the 35 participants were noted to have a diagnosis of MRKH (all treated with skin graft neovaginoplasty). Of those patients, only two were examined to correlate the anatomic findings with the psychosexual responses. All of the patients completed at least some part of the Female Sexual Function Index (FSFI), a validated measure in the vaginal reconstruction population, with the mean score of 25.0, which falls below the 26.55 cutoff, implying that they are at risk for developing sexual dysfunction. The authors acknowledged that their population was not randomized for treatment, was recruited exclusively from a clinical sample, and may have been biased by including patients who created a sufficient vagina by coitus alone with those that followed a dilation program. Regardless, their results support that any treatment for vaginal hypoplasia may be of limited utility without psychological expertise to address other aspects of self-perception. More work in the

area of overall quality of life, well-being, and emotional/sexual wellness is needed in this population.

When counseling patients, gestational surrogacy should definitely be included in the discussion. Until recently, the literature had provided only sparse evidence regarding the use of this modality in this population. Beski and colleagues confirmed the use of gestational surrogacy in a small population. The treatment cycles resulted in six clinical pregnancies (42.9% pregnancy rate per embryo transfer and 54.5% per oocyte retrieval) and three live births (21.4% per embryo transfer, 27.3% per retrieval, and 50% per patient). Several authors have reported on the genetic offspring of patients with vaginal agenesis. Petrozza and colleagues reported a retrospective study in 1997, describing a large number of treatment cycles for patients with Rokitansky syndrome. The authors attempted to determine an inheritance pattern of the syndrome through a questionnaire sent to all centers performing surrogacy treatment in the United States. A total of 162 in vitro fertilization/surrogacy treatment cycles were reviewed for 58 patients with congenital agenesis of the uterus and vagina. The treatment resulted in 34 live births (17 girls, 17 boys). One child had a nonspecific middle ear defect and hearing loss. The authors concluded that congenital absence of the uterus and vagina was not commonly inherited in a dominant fashion. These findings suggest that inheritance of this disorder in children of affected mothers is likely via a polygenic mechanism. In this population, none of the 17 female infants born to affected mothers exhibited Rokitansky syndrome. Ovulation induction is similar to other patients; however, oocyte retrieval may be more challenging. Typically, oocytes are retrieved transvaginally; yet, after creation of a neovagina, the ovaries may not be as easily accessible. In addition, ovaries may be ectopically located, higher in the abdomen. Barton and colleagues reported success with a transabdominal approach without decrease in safety or efficacy. Five of the sixty-nine women who underwent abdominal follicular aspiration were noted to have congenital reproductive anomalies.

Patient Cooperation

Regardless of which operative technique is chosen, the patient must cooperate if the operation is to be successful. In many of the available vaginal reconstruction techniques, dilation or use of a vaginal mold will be necessary. For example, when a McIndoe operation is performed, patients must understand the need to wear a form continuously for several months and intermittently for several years until the vagina is no longer subject to constriction and until regular intercourse is taking place. No surgery should be performed until preoperative evaluation determines that the patient understands her essential role in its success. This is especially important when the patient is a younger teenager. The single most important factor in determining the success of vaginoplasty is the psychosocial adjustment of the patient to her congenital vaginal anomaly.

Laboratory and Diagnostic Testing

A complete chromosomal analysis should be considered in all patients. If there is a suspicion of ovarian dysgenesis, androgen insensitivity syndrome, or some aberration of the classic MRKH syndrome, then a consideration of additional SRY analysis should be entertained to assess the possible presence of any Y chromosome. A contrast study such as a magnetic resonance (MR) urogram or an intravenous pyelogram should be done preoperatively. This also provides an adequate survey for anomalies of the spine. If a pelvic mass is present, then additional special studies may be indicated to differentiate between hematometra, hematocolpos, endometrial and other ovarian cysts, and pelvic kidney. The MR urogram may facilitate the evaluation of the reproductive, urologic, and skeletal systems with one radiographic study.

Evaluation of Cyclic Pain

Some patients without a pelvic mass report cyclic pain. This pain can be ovulatory or possibly a result of dysmenorrhea originating in well-developed rudimentary uterine bulbs. The physician can differentiate between the two by asking the patient to keep a pain diary and reviewing the diagnostic imaging with an experienced reconstructive gynecologic surgeon or radiologist to ensure that an obstructed anlagen is not present. Occasionally, there is a question about whether a patient has congenital absence of the vagina or an imperforate hymen with cryptomenorrhea. The diagnosis is clarified before operative intervention by using radiographic imaging. Pelvic ultrasonography can often detect hematocolpos or an

obstructed uterine anlagen distended with menstrual blood. Magnetic resonance imaging (MRI) can differentiate the two diagnoses, if necessary.

METHODS OF CREATING A VAGINA

There is no unanimity of opinion regarding the correct approach to the problem of vaginal agenesis (Table 25.3). With the development of the Ingram method for vaginal dilatation, fewer patients require surgical vaginoplasty. The American Congress of Obstetricians and Gynecologists has supported nonsurgical creation of a neovagina as first-line therapy since 2006. Increased utilization of additional surgical techniques has broadened the discussion of surgical treatment of vaginal agenesis. The role of tissue expanders in vaginoplasty has been reviewed by Patil and Hixon. Labial expansion with an expander having a capacity of 250 mL provides a flap

10 cm long and 8 cm wide with a 4-cm projection. Thus, wellvascularized flaps can be available to provide an outlet for stenosis-free vaginoplasty. This approach has been suggested to maximize the success of surgical vaginoplasty. A review of the methods devised for the formation of a vagina follows. The editors of this book have found the modified McIndoe technique to give the most consistently satisfactory results.

TABLE 25.3 Classification of Methods to Form a New Vagina

Nonsurgical (intermittent pressure on the perineum)
Active dilatation
Passive dilatation
Surgical
Without the use of abdominal contents
Without cavity dissection
Vulvovaginoplasty
Constant pressure (Vecchietti)
No attempt to line cavity (now unacceptable)
Lining cavity with grafts
Split-thickness skin grafts (McIndoe operation)
Dermis grafts
Amnion homografts
Lining cavity with flaps
Musculocutaneous flaps
Fasciocutaneous flaps
Subcutaneous pedicled skin flaps
Labial skin flaps (can be created with tissue expander)
Penoplasty (transsexualism)
With use of abdominal contents (cavity lining with)
Peritoneum
Free intestinal graft
Pedicled intestine

Nonsurgical Methods

In 1938, Frank described a method of creating an artificial vagina without operation. In 1940, he reported remarkably satisfactory results in eight patients treated with this method. His follow-up study showed that a vagina formed in this manner remained permanent in depth and caliber, even in patients who neglected dilatation for more than 1 year. It has been emphasized that the pelvic floor itself is embryologically deficient in some patients. Indeed, the ease with which some patients are able to create a vagina with intercourse alone or with other intermittent pressure techniques can be explained on this basis. Five patients were reported to have developed enteroceles, one after coitus alone, three after a Williams vulvovaginoplasty, and three after a McIndoe operation. This complication can develop when the vaginal mucosa is brought in close proximity to the pelvic peritoneum, but a relative embryologic weakness or an absence of endopelvic fascia can also contribute to this complication. Rock, Reeves, and associates at the Johns Hopkins Hospital reported that an initial trial of vaginal dilatation was successful in 9 of 21 patients.

Prompted by the rewarding results of Broadbent and Woolf, Ingram has described a passive dilatation technique of creating a new vagina. Instructing his patients in the insertion of dilators (Fig. 25.5) specially designed for use with a bicycle seat stool, Ingram was able to produce satisfactory vaginal depth and coital function in 10 of 12 cases of vaginal

agenesis and 32 of 40 cases of various types of stenosis.

FIGURE 25.5 Vaginal dilators for use in Ingram passive dilatation technique to create a new vagina. The set from Faulkner Plastics consists of 19 dilators of increasing length and width.

The Ingram technique for passive dilatation has several advantages. The patient is not required to press the dilator against the vaginal pouch. A series of graduated Lucite dilators slowly and evenly dilate the neovaginal space. The patient should be carefully instructed in the use of dilators, as recommended by Ingram, beginning with the smallest dilator. The patient is shown and instructed with the use of a mirror how to place a dilator against the introital dimple. The dilator may be held in place with a supportive undergarment and regular clothing worn over this.

The patient is shown how to sit on a racing-type bicycle seat that is placed on a stool 24 inches above the floor. She is instructed to sit leaning slightly forward with the dilator in place for at least 2 hours/day at intervals of 15 to 30 minutes. Follow-up is usually at monthly intervals, and the patient can expect to graduate to the next size larger dilator approximately every month. An attempt at sexual intercourse may be suggested after the use of the largest dilator for 1 or 2 months. Continued dilatation is recommended if intercourse is infrequent. In our experience, functional success rates are outstanding. Roberts, Haber, and Rock previously reported the largest series of vaginal agenesis patients who used the Ingram method of dilatation to create a neovagina. The records of 51 patients with müllerian agenesis were reviewed: 37 patients attempted vaginal dilatation, and 14 young women underwent a surgical intervention. Functional success was defined as satisfactorily achieving intercourse or accepting the largest dilator without discomfort in the clinic visit. All patients were followed up for at least 2 years and for an average of 9.25 years. Functional success was achieved in 91.9% of those who attempted dilatation (**Table 25.4**). Thus, passive dilatation should be suggested as an initial therapy for vaginal creation. If dilatation is unsuccessful, operative vaginoplasty is indicated.

Edmonds and colleagues reported on their experience with managing 360 patients with MRKH from 1998 to 2010. Two hundred forty-five patients requested vaginal dilation for treatment. The mean age of dilating patients was 18.6 years (16 to 22 years). Success was defined as achieving sexual satisfaction and functional vaginal length (6 cm) and was achieved in 232 of 245 patients (94.7%). The mean time to complete therapy was 5.5 months (2 to 19 months). Despite having large number of patients in the study, the applicability outside of the United Kingdom could be questioned. The UK program consists of an intensive program in which patients have an identified clinical nurse specialist for teaching and support, one or two specific providers, and an inpatient admission (an average of 3 days) to teach dilation technique. Also, sexual satisfaction was not measured in a structured way for all patients; a subset of 60 patients were assessed using a sexual function questionnaire with the answers compared to a normal population. In the United States,

Gargollo and colleagues described their 12-year experience treating patients with vaginal agenesis. Rokitansky syndrome was the primary diagnosis in 64 of the 69 patients included in the retrospective review. The mean age at start of vaginal dilation was 17.5 years (14 to 35 years), and mean follow-up time was 19 months (0 to 100). Progressive perineal dilation was the treatment of choice. The total success rate was 88% achieving functional success (ability to achieve satisfactory vaginal intercourse, vaginal acceptance of the largest dilator, or vaginal length of at least 7 cm), during a mean of 18.7 months of therapy. Their work not only described an outpatient regimen that could be applicable in the United States but also highlighted the integral role that the multidisciplinary team plays in patient success. They also reinforced that dilating can be successful even in patients with a small vaginal dimple; thus, validating an attempt would be reasonable in almost all patients with vaginal agenesis.

TABLE 25.4 Outcomes of Patients with Vaginal Agenesis Who Attempted Dilatation

PATIENTS	TOTALS	PERCENT
Successful dilatation	34/37	91.9%[a]
Failed dilatation	3/37	8.1%

[a] $P < 0.001$.

Modified from Roberts CP, Haber MJ, Rock JA. Vaginal creation for müllerian agenesis. *Am J Obstet Gynecol* 2001;185:1349, with permission. Copyright © 2001, Elsevier.

Surgical Methods

During the past three decades, experience has proved the Abbe-Wharton-McIndoe procedure (more popularly called the McIndoe operation) for dealing with complete absence of a vagina to be generally superior to others in most cases. In special circumstances, alternative methods of creation of a neovagina may be indicated.

Historical Development of Surgical Procedures

In 1907, Baldwin used a double loop of ileum to line a space dissected between the rectum and bladder, leaving the mesentery connected to the bowel. The continuity of the intestinal tract was reestablished by an end-to-end anastomosis. He reported that the new vagina was absolutely normal in every way. In 1910, Popaw constructed a vagina using a portion of the rectum that was moved anteriorly. This operation was modified by Schubert in 1911. The rectum was severed above the anal sphincter and moved anteriorly to serve

as the vagina. The sigmoid was sutured to the anus to reestablish the continuity of the intestinal tract. Both operations had soberingly high morbidity and mortality rates, and their popularity declined. Today, segments of sigmoid are used most often to create a vaginal pouch or extend vaginal length in patients who have lost vaginal function as a result of extensive surgery or irradiation for pelvic malignancy. Some patients who are treated for multiple genitourinary or gastrointestinal abnormalities may be treated with a bowel vaginoplasty during a combined procedure.

Less formidable procedures involving dissection of a space between the bladder and rectum and lining of this space with flaps of skin from the labia or inner thighs also were tried. Marked scarring resulted, and hair usually grew in the vagina. Extensive plastic procedures to construct a vagina are no longer necessary or desirable and have been discarded in favor of safer procedures unless there is the problem of maintaining a vaginal canal after an extensive exenterative operation for pelvic malignancy. In this case, the physician may want to consider using the gracilis myocutaneous flap technique described by McCraw and associates in 1976.

The Abbe-Wharton-McIndoe Operation

This operation for creating a new vagina began with simple surgical attempts to create a space between the bladder and the rectum. These early attempts were often made in patients with cryptomenorrhea. However, such a space usually would constrict because the surgeon would fail to recognize the importance of prolonged continuous dilatation until the constrictive phase of healing was complete.

At the Johns Hopkins Hospital in 1938, Wharton combined an adequate dissection of the vaginal space with continuous dilatation by a balsa form that was covered with a thin rubber sheath and was left in the space. He did not use a split-thickness skin graft. Instead, he based his operation on the principle that the vaginal epithelium has remarkable powers of proliferation and in a relatively short time will cover the raw surface. Recalling that a similar process occurs in the fetus when the epithelium of the sinovaginal bulbs and the urogenital sinus form the vaginal canal, Wharton merely applied this same principle in the adult. This simple procedure is entirely satisfactory as long as the space is kept dilated long enough to allow the epithelium to grow in. Occasionally, however, even after several years, the vault of the vagina remains without epithelial covering. Coital bleeding and leukorrhea result from the persistent granulation tissue, and there is a tendency for vaginas constructed by this method to be constricted by scarring in the upper portion. In Counseller's 1948 report from the Mayo Clinic of 100 operations to construct a new vagina, 14 were performed by the Wharton method, with excellent results in all 14 patients. It was stated that the disadvantages of persistent granulation tissue with bleeding and leukorrhea were of no consequence. This has not been the experience of the editors of this book.

When inlay skin grafts were first used to construct a new vagina, the results were poor because the necessity for dilatation of the new vagina again was not recognized. Severe contraction, uncontrolled by continuous or intermittent dilatation, almost invariably spoiled the results. Although Abbe and others preceded him by many years in using a skin-covered prosthesis in neovaginal construction, it was Sir Archibald McIndoe, at the Queen Victoria Hospital in England, who popularized the method and gave it substantial clinical trial. He emphasized the three important principles used today in successful operations for vaginal agenesis:

1. Dissection of an adequate space between the rectum and the bladder
2. Inlay split-thickness skin grafting
3. The cardinal principle of continuous and prolonged dilatation during the contractile phase of healing

Other tissues such as amnion and peritoneum have been used to line the new vaginal space, but they have not had substantial success. However, Tancer and associates reported good results with human amnion. Karjalainen and associates stated that a more physiologic result was achieved with an amnion graft than with a skin graft. Nevertheless, concerns about the transmission of human immunodeficiency virus with human amnion now limit this option.

Technique of Abbe-Wharton-McIndoe Operation

TAKING THE GRAFT

After a careful pelvic examination is performed under anesthesia to verify previous findings, the patient is positioned for taking a skin graft from the buttocks. For cosmetic reasons, the graft should not be taken from the thigh or hip unless for some reason it cannot be obtained from the buttocks. Patients may be asked to sunbathe in a bathing suit before coming to the hospital so that its outline can be seen; an attempt should be made to take the graft from both buttocks within these borders. The quality of the graft determines to a great extent the success of the operation. We have found the Padgett electrodermatome to be the most satisfactory instrument for taking the graft. With relatively little experience and practice, the gynecologic surgeon can successfully cut a graft of controlled width and thickness (**Fig. 25.6**). The instrument is set and checked for taking a graft approximately 0.018 inch thick and 8 to 9 cm wide. The total graft length should be 16 to 20 cm. If the entire graft cannot be taken from one buttock, then a graft 8 to 10 cm long is needed from each buttock.

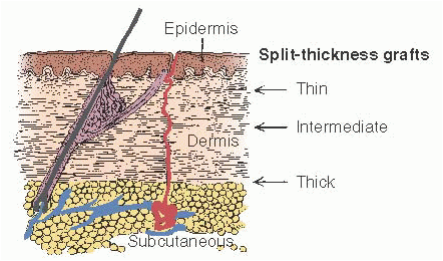


FIGURE 25.6 Section of split-thickness skin grafts. Grafts should be uniform in thickness. The Padgett electrodermatome is set to take a graft approximately 0.018 inch thick. A graft that is slightly thick is better than a thin graft.

The skin of the donor site is prepared with an antiseptic solution (povidone-iodine), which is then thoroughly washed away. The skin is then lubricated with mineral oil as assistants steady and stretch the skin tight. Considerable pressure should be applied uniformly across the dermatome blade. The thickness of the graft must have minimal variation. A graft that is a little too thick is better than one that is a little too thin. There should be no breaks in the continuity of the graft. The graft is placed between two layers of moist gauze, and the donor sites are dressed. The donor site is soaked with a dilute solution of epinephrine for hemostasis, and a sterile dressing is applied. A pressure dressing is then placed over the site; this dressing can be removed on the seventh postoperative day. The sterile dressing dries in place over the donor site and ultimately will fall off by itself. Moistened areas on the dressing can be dried with cool air. If there is separation and evidence of some superficial infection, then merbromin can be applied to these areas.

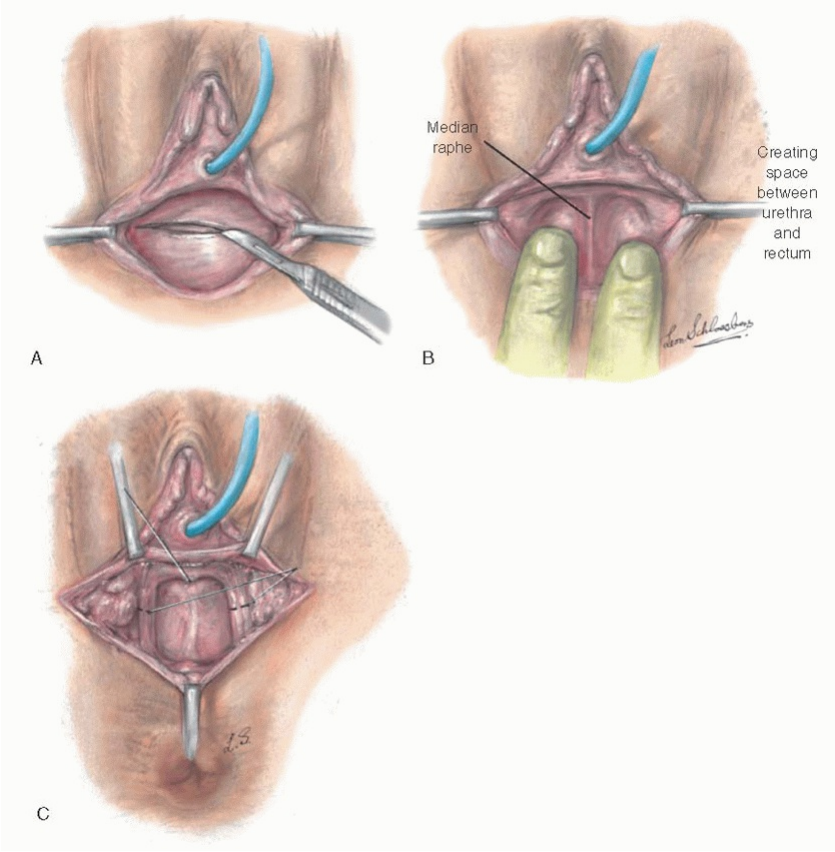


FIGURE 25.7 The McIndoe procedure. **A:** A transverse incision is made in the apex of the vaginal dimple. **B:** A channel can usually be dissected on each side of the median raphe. The median raphe is then divided. Careful dissection prevents injury to the bladder and rectum. **C:** A space between the urethra and bladder anteriorly and the rectum posteriorly is dissected until the undersurface of the peritoneum is reached. Incision of the medial margin of the puborectalis muscles will enlarge the vagina laterally.

CREATING THE NEOVAGINAL SPACE

The patient is placed in the lithotomy position, and a transverse incision is made through the mucosa of the vaginal vestibule (Fig. 25.7A). The space between the urethra and bladder anteriorly and the rectum posteriorly is dissected until the undersurface of the peritoneum is reached.

This step may be safer with a catheter in the urethra and sometimes a finger in the rectum to guide the dissection in the proper plane. After incising the mucosa of the vaginal vestibule transversely, the physician often is able to create a channel on each side of a median raphe (Fig. 25.7B), starting with blunt dissection and then dilating each channel with Hegar dilators or with finger dissection. In some instances, it may be necessary to develop the neovaginal space by dissecting laterally and bringing the fingers toward the midline. The median raphe is then divided, thus joining the two channels. This maneuver is helpful in dissecting an adequate space without causing injury to surrounding structures.

To avoid subsequent narrowing of the vagina at the level of the urogenital diaphragm, it may be helpful to incise the margin of the puborectalis muscles bilaterally along the midportion of the medial margin (Fig. 25.7C). Although useful in all circumstances, incision of the puborectalis muscle is more important in cases of androgen insensitivity syndrome with android pelvis, in which the levator muscles are more taut against the pelvic diaphragm, than in cases of gynecoid pelvis. Incision of the puborectalis muscle causes no difficulty with fecal incontinence, significantly improves the ease with which the vaginal form can be inserted into the canal in the postoperative period, and has eliminated the problem of contracture of the upper vagina caused by a poorly applied form. The dissection should be carried as high as possible without entering the peritoneal cavity and without cleaning away all tissue beneath the peritoneum. A split-thickness skin graft will not take well when applied against a base of thin peritoneum. All bleeders should be ligated by clamping and tying them with very fine sutures. It is essential that the vaginal cavity be dry to prevent bleeding beneath the graft. Bleeding causes the graft to separate from its bed, resulting in the inevitable failure of the graft to implant in that area and in local graft necrosis.

PREPARING THE VAGINAL FORM

Early skin grafts were formed over balsa, which has the advantages of being an inexpensive, easily available, light wood that can be sterilized without difficulty. It also can be whittled easily in the operating room to a proper shape to fit the new vaginal space. However, uneven pressure from the form can cause a skin graft to slough in places, and pressure spots are associated with an increased risk of fistula formation. The Counseller-Flor modification of the McIndoe technique (Fig. 25.8) uses, instead of the rigid balsa form, a foam rubber mold shaped for the vaginal cavity from a foam rubber block and covered with a condom. The foam rubber is gas sterilized in blocks measuring approximately 10 × 10 × 20 cm. The block is shaped with scissors to approximately twice the desired size, compressed into a condom, and placed into the neovagina (Fig. 25.8A-C). The form is left in place for 20 to 30 seconds with the condom open to allow the foam rubber to expand and conform to the neovaginal space (Fig. 25.8D). The condom is then closed, and the form is withdrawn. The external end is tied with 2-0 silk, and an additional condom is placed over the form and tied securely (Fig. 25.8E, F).

SEWING THE GRAFT OVER THE VAGINAL FORM

The skin graft is then placed over the form and its undersurface exteriorized and sewn over the form with interrupted vertical mattress 5-0 nonreactive sutures (Fig. 25.8G, H). Where the graft is approximated, the undersurfaces of the sutured edges are also exteriorized.

The graft should not be “meshed” to make it stretch farther, and the edges of the graft should be approximated meticulously around the form without gaps. Granulation tissue develops at any place where the form is not covered with skin. Contraction usually occurs where granulation tissue forms. After the form has been placed in the neovaginal space, the edges of the graft are sutured to the skin edge with 5-0 nonreactive absorbable sutures, with sufficient space left between sutures for drainage to occur. The physician must be careful not to have the form so large that it causes undue pressure on the urethra or rectum. A balsa form should have a groove to accommodate the urethra. With a foam rubber form, this is unnecessary. A suprapubic silicone catheter is placed in the bladder for drainage. If the labia are of sufficient length, then the form can be held in place by suturing the labia together with two or three nonreactive sutures.

REPLACING WITH A NEW FORM

After 7 to 10 days, the form is removed and the vaginal cavity is irrigated with warm saline solution and inspected. This is usually performed with mild sedation and without an

anesthetic. The cavity should be inspected carefully to determine whether the graft has taken satisfactorily in all areas of the new vagina. Any undue pressure by the form should be noted and corrected. It is especially important that there not be too much pressure superiorly against the peritoneum of the cul-de-sac. Such a constant upward pressure could result in weakness with subsequent enterocele formation. The new vaginal cavity must be inspected frequently to detect and to prevent pressure necrosis of the skin graft.

The patient is given instructions on daily removal and reinsertion of the form and is taught how to administer a lowpressure douche of clear warm water. She is advised to remove the form at the time of urination and defecation, but otherwise to wear it continuously for 6 weeks. A neoprene form, which is much easier to remove and keep clean than a foam rubber form, is substituted for the original form in 6 weeks. A new form is molded with a sterile sheath cover (condom) to fit the size of the vaginal canal. The patient is instructed to use the form during the night for the following 12 months. If there has been no change in the caliber of the vagina by that time, then it is unlikely to occur later, and insertion of the form at night can be done intermittently until coitus is a frequent occurrence. However, if there is the slightest difficulty in inserting the form, then the patient should be advised to use the form continuously again. Most patients are able to maintain the form in place simply by wearing tighter underclothes and a perineal pad. Douches are advisable during residual vaginal healing and discharge.

RESULTS AND COMPLICATIONS

Results with the McIndoe operation have improved over the years. Recently, reported percentages of satisfactory results have ranged from 80% to 100%. The serious complications formerly associated with the McIndoe operation have been significantly reduced by improvements in technique and greater experience. Serious complications do still occur, however, including a 4% postoperative fistula rate (urethrovaginal, vesicovaginal, and rectovaginal), postoperative infection, and intraoperative and postoperative hemorrhage. Failure of graft take is also still reported as an occasional complication. Failure of graft take often leads to the development of granulation tissue, which might require reoperation, curettage of the granulation tissue down to a healthy base, and even regrafting. Minor granulation can be treated with silver nitrate application. The functional result is more important than the anatomic result in evaluating the success of this operation. Although a vagina of only 4 cm is adequate for some couples, in most instances, a vagina smaller than 4 cm causes major problems.

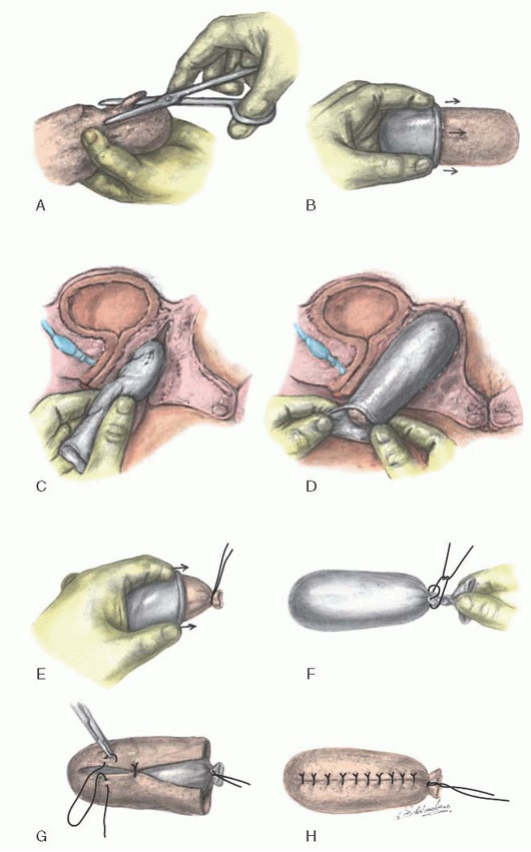


FIGURE 25.8 Counseller-Flor modification of the McIndoe technique. **A:** A form is cut from a foam rubber block. **B:** A condom is placed over the form. **C:** The form is compressed and placed into the vagina. **D:** Air is allowed to expand the foam rubber, which accommodates to the neovaginal space. The condom is closed and the form removed. A second condom is placed over the form **(E)** and tied securely **(F). G:** The graft is then sewn over the form with interrupted 5-0 nonreactive sutures. **H:** The undersurfaces of the sutured edges of the graft are exteriorized. The vaginal form is ready for insertion into the neovagina.

The postoperative results have improved significantly since the balsa vaginal form was replaced by the foam rubber form. Between 1950 and 1989, the McIndoe operation was performed on 94 patients at the Johns Hopkins Hospital. During these 39 years, 83% of the 94 patients had a 100% take of the graft; in only three cases was there a significant area over which the graft failed.

Urethrovaginal fistula has become even more infrequent since the introduction of the suprapubic catheter and the foam rubber form. The catheter is removed when the patient is voiding well and has no residual urine. In general, the patient is able to void without difficulty within the first few days of the procedure. Prophylactic broad-spectrum antibiotics started within 12 hours of surgery and continued for 7 days are of definite value in reducing the incidence of graft failures from infection in the operative site.

Because of the excellent results obtained after a modified McIndoe vaginoplasty, this operation is recommended as the procedure of choice for women unable or unwilling to obtain a neovagina with dilatation methods. Women with a flat perineum with no dimple or pouch have no alternative other than the McIndoe vaginoplasty to obtain a neovagina for comfortable sexual relations.

Desirability of the modified McIndoe procedure may be increased by the use of alternative graft harvest sites to conceal possible aesthetic concerns of the buttock site, as proposed by

Höckel and colleagues. The authors proposed the use of split skin harvesting from the scalp because thin (0.25-mm) split skin grafts do not seem to hamper hair growth at the donor site nor lead to hair growth at the recipient site. Because alopecia has been reported as a complication associated with technical errors, more experience is necessary before advocating the scalp as a potential graft harvest site.

It is important that a McIndoe operation be performed correctly the first time. If the vagina becomes constricted because of granulation tissue formation, injury to adjacent structures,

or failure to use the form properly, then subsequent attempts to create a satisfactory vagina are more difficult. The first operation has the best chance of success.

Ozek, like many other surgeons, modified the McIndoe procedure by describing an X-type perineal incision and the use of a perforated vaginal mold during the postoperative period. He postulated that this incision minimized stricture at the vaginal introitus and provided greater ease of dissection of the vaginal cavity. He reinforced that the overall procedure is simple with a generally uneventful postoperative course. Complications included infection, failure of skin graft take, stress urinary incontinence, partial graft loss, and vaginal stricture. All were treated satisfactorily except the patient with stress urinary incontinence.

Despite any minor modifications of the McIndoe vaginoplasty, the essential components of dissection of an adequate space, split-thickness skin grafting, and continuous dilatation during the contractile phase of healing remain unchanged. Recent reviews continue to support the safety and efficacy of the procedure. Hojsgaard and Villadsen reported 26 patients who underwent vaginoplasty, 18 of whom had Rokitansky syndrome. All patients were recorded as having a satisfactory result with complete graft take, adequate vaginal dimensions, and no strictures or fistulas giving symptoms. Complete take was achieved in 33% of patients within a week postoperatively, and after one further grafting procedure, an additional 38% had complete take. The intraoperative and early postoperative complications were perforation of the rectum in one patient (3.8%) and postoperative bleeding in three patients (11.5%). The late complications were vaginal stricture in three patients (11.5%), urethrovaginal fistula in two patients (7.7%), and rectovaginal fistula in one patient (3.8%). Alessandrescu and colleagues described the surgical management of 201 cases of Rokitansky syndrome. The surgeon substituted a modified transverse perineal incision and a perforated, rigid plastic mold. Intraoperative and postoperative complications consisted of two rectal perforations (1%), eight graft infections (4%), and 11 infections of graft site origin (5.5%). Sexual satisfaction was investigated with both objective and subjective criteria. Among the 201 cases, 83.6% had anatomic results evaluated as “good,” 10% as “satisfactory,” and 6.5% as “unsatisfactory.” More than 71% of patients rated their sexual life as “good” or “satisfactory” and reported that they had been able to experience orgasms related to vaginal intercourse. Twenty-three percent reported the ability to have sexual intercourse but had no ability to achieve orgasm, and only 5% expressed dissatisfaction with their sexual performance. Strickland and colleagues reported on the coital satisfaction, perception of vaginal competence, and impact on lifestyle of adult women undergoing vaginoplasty as adolescents. Ten of twenty-two women responded to a questionnaire at a median of 18 years (range 5 to 13 years) following surgical intervention with a McIndoe vaginoplasty. All of the women had sexual experience, and 80% were sexually active at the time of evaluation. The most frequent difficulty reported was vaginal dryness and lack of lubrication with sexual intercourse. Ninety percent of the subjects expressed satisfaction that sexual ability was acceptable. This experience also supports the role of the McIndoe vaginoplasty in providing young women with vaginal agenesis long-term coital ability and minimal disabilities.

DEVELOPMENT OF COMPLICATIONS

Several case reports exist of malignant disease developing in a vagina created by various techniques; these reports were reviewed by Gallup and colleagues, as well as others. The authors reported a patient who was initially treated for intraepithelial malignancy by total vaginectomy combined with a split-thickness skin graft vaginoplasty to reconstruct a functional vagina. The authors noted a lesion in her vaginal apex 7 years later. These findings suggest that epithelium transplanted to the vagina can assume the oncogenic potential of the lower reproductive tract. The evidence supports a risk of neoplastic change in both skin and bowel grafts, with a reported average interval from vaginoplasty to diagnosis on the order of 19 years or greater. Epithelium transplanted to the vagina will assume the oncogenic potential of the lower reproductive tract. With less than 25 cases of primary carcinoma of the neovagina reported, it is presently unclear if transplantation of bowel or other graft tissue alone changes the malignancy risk. Consideration of a plan for surveillance and counseling regarding transmission risk for virally related dysplastic change in the lower genital tract is also extremely important for any patients undergoing neovaginoplasty. Neovaginal vault prolapse has also been reported in patients managed both medically and surgically. Without apical or lateral suspension by endopelvic fascial attachments, subsequent prolapse may develop with any type of neovagina. Prolapse in neovaginal segments created by dilation, McIndoe technique, skin flaps, and bowel segments is reported. Abdominal suspension via sacrocolpopexy, abdominal suspension to the Cooper ligament, vaginal resection of redundant prolapsed bowel neovagina, and sacrospinous ligament suspension have all been described as treatment. Kondo and colleagues report a recurrence rate after surgical treatment of 25%.

The Williams Vulvovaginoplasty

Construction of a perineal bridge to help contain the vaginal mold was a routine part of the operation described by McIndoe, but it was not adopted subsequently by others. However, Williams described a similar vulvovaginoplasty procedure in 1964 and advised that it could be used to create a vaginal canal (**Fig. 25.9**). In 1976, he reported that the procedure was unsuccessful in only 1 of 52 patients. Feroze and coworkers reported that the anatomic results were good in 22 of 26 patients. According to these authors, the advantages of the Williams operation are its technical simplicity, its absence of serious local complications even when performed as a repeat procedure, the ease of postoperative care, the absence of postoperative pain, the speed of recovery, the possible elimination of dilators and consequent applicability to patients who do not intend to have regular intercourse in the near future, and the higher success rates of primary and repeat procedures. The technique is not applicable to patients with poorly developed labia. It does result in an unusual angle of the vaginal canal, which is reported to straighten to a more normal direction with intercourse. If a very high perineum is created, urine can momentarily collect in the pouch after urination, giving the impression of postvoid incontinence. Failure of the suture line to heal by primary intention results in a large area of granulation tissue and most likely an unsatisfactory result.

Williams believes that if the urethral meatus is patulous, a vulvovaginoplasty should not be performed because the urethra might be stretched further by coitus. He suggests that varying deficiencies in muscular and fascial tissue can explain

why some patients with uterovaginal agenesis are able to develop a satisfactory vaginal canal with simple intermittent pressure with coitus, whereas others are prone to develop enteroceles.

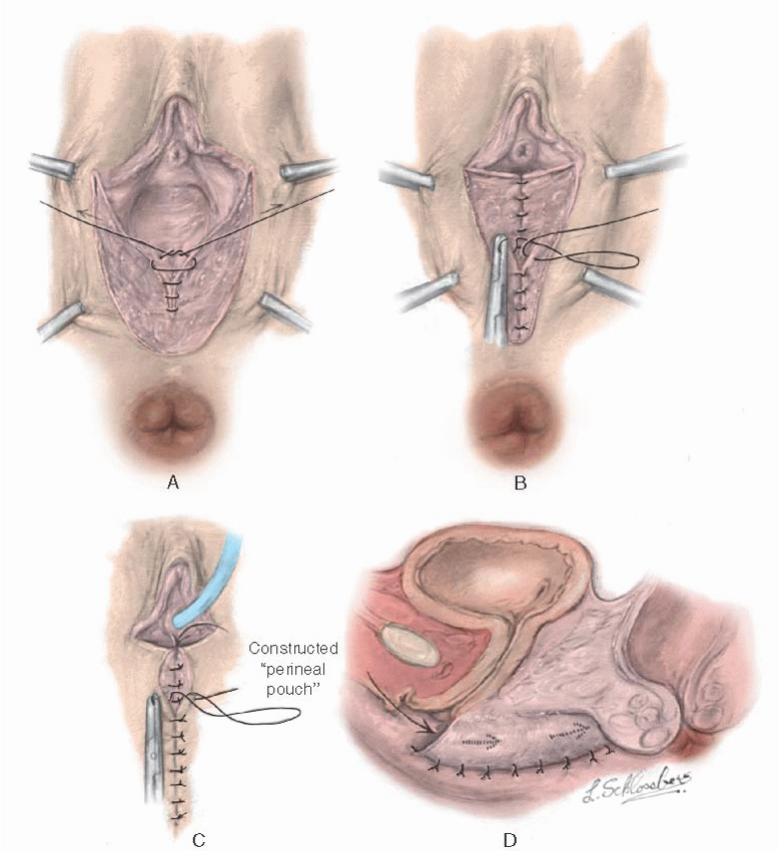


FIGURE 25.9 The Williams vulvovaginoplasty. **A-C:** No. 3-0 polyglycolic acid sutures can be used throughout to close both inner and outer skin margins and the tissue between. **D:** The entrance to the pouch should not cover the external urethral meatus.

The Williams vulvovaginoplasty is a useful operation and should certainly be considered the operation of choice for patients needing a follow-up to an unsatisfactory primary operation to create a neovagina or a supplement to a small vagina resulting from extensive surgery or radiation therapy, when options are limited. It remains an option for the rare patient with a solitary kidney low in the pelvis who does not have room for dissection of an adequate vaginal space.

Alternative Techniques

Schätz and colleagues reported three patients who underwent the George modification of the Wharton-Sheares neovaginoplasty technique. Wharton combined an adequate dissection of the vaginal space with continuous dilatation by a balsa form that was covered with a thin rubber sheath and was left in the space. His operation was based on the principle that the vaginal epithelium will proliferate and, in a relatively short time, will cover the raw surface. This modification of an already simple procedure may provide an alternative in areas where multidisciplinary, reconstructive pelvic surgery teams are not available. Because the procedure includes the mere dilatation of the bilateral müllerian ducts and lining of the superior aspect with peritoneum, it eliminates some of the more complex aspects of the other surgical options. However, with a mean follow-up period reported as only 12 months (range of 2 to 23 months), reported long-term complications associated with the Wharton method (persistent granulation tissue, coital bleeding, leukorrhea, and scarring of the upper portion) cannot be fully evaluated. Longer follow-up and a larger patient experience are both necessary to consider this procedure as a primary option for patients with MRKH. Walch and colleagues from Austria reported the outcome of 10 patients with MRKH treated with the George modification of the Wharton-Sheares approach. Mean follow-up from surgery was 33.5 ± 22.4 months (range 3 to 77 months). Data collected on follow-up included cytologic smears of the neovagina, swabs, culture, and hybrid capture for human papillomavirus (HPV). The mean vaginal length was 8.3 cm (range 7 to 10 cm). Of the 8 patients from whom a biopsy was able to be obtained, all showed nonkeratinizing stratified squamous epithelium. The sexual function scores obtained by the Rosen FSFI questionnaire were comparable to previously published control patients. The authors propose that an important drawback of this technique may be the lifelong routine need for the use of vaginal dilators (Table 25.5).

TABLE 25.5 Comparison of Female Sexual Function Index Values for Different Types of Neovaginoplasty

	WHARTONSHEARESGEORGE SURGERY (*n* = 7)	SIGMOID VAGINOPLASTY (*n* = 11)	DAVYDOV LAPAROSCOPIC VAGINOPLASTY (*n* = 25)	LAPAROSCOPIC DAVYDOV TECHNIQUE (*n* = 40)	LAPAROSCOPIC DAVYDOV TECHNIQUE (*n* = 6)	LAPAROSCOPIC MODIFIED VECCHIETTI TECHNIQUE (*n* = 27)	LAPAROSC VECCHIETT TECHNIQUI 40)
Desire	4.3 ± 1.3	4.7 ± 0.9	4.4 ± 0.9	4.3 ± 0.7	3.9 ± 1.2	4.4 ± 0.8	4.2
Arousability	5.4 ± 0.5	4.9 ± 0.6	4.4 ± 1.1	4.7 ± 0.8	4.0 ± 1.1	4.8 ± 0.8	4.6
Lubrication	5.1 ± 1.5	5.0 ± 0.9	4.5 ± 1.4	5.1 ± 0.6	4.4 ± 1.0	5.0 ± 0.8	4.5
Orgasm	4.9 ± 1.0	5.3 ± 0.8	4.1 ± 1.3	5.0 ± 0.6	3.3 ± 1.6	4.6 ± 1.0	4.4
Satisfaction	5.1 ± 1.5	4.7 ± 1.6	4.6 ± 1.1	4.8 ± 1.5	4.0 ± 1.8	5.4 ± 0.6	5.2
Pain	5.1 ± 1.1	3.5 ± 2.4	4.4 ± 1.4	4.7 ± 1.0	1.9 ± 1.9	5.0 ± 0.9	5.2

Total score	29.9 ± 4.3	28.0 ± 5.0	26.5 ± 5.6	31.8 ± 0.8	21.4 ± 5.3	29.0 ± 3.2	30.2
Reference		Communal P H et al.	Giannes A et al.	Bianchi S et al.	Allen LM et al.	Fedele L et al.	Bianchi S et

Note: Values are given as means ± SD. Patients without sexual activity within the past month were not included in this FSFI evaluation. Total FSFI score ranges from 0 to 36. Modified from Walch. Long-term outcomes after Wharton-Sheares-George surgery. *Fertil Steril* 2011;96:492, with permission. Copyright © 2011, Elsevier.

Since the initial report of the use of oxidized cellulose material instead of a skin graft to line the neovaginal space, more than 30 cases have been reported using several different commercial products. No operative or postoperative complications have been reported and epithelialization was noted between 1 and 12 months after the procedure. Limited long-term data are available regarding this technique. Dornelas and colleagues in Brazil recently reported on 11 patients treated with this modality, eight of which had MRKH. The mean follow-up for the entire group was 14 months (6 to 24 months). Patients who were sexually active at the 6-month assessment were administered the FSFI, translated into Portuguese, with all the results in the "very good" range. The authors reinforced that if sexual intercourse is suspended for prolonged periods, vaginal dilation may be required to preserve vaginal length. The proposed advantages include the shorter operative times and easily available material that does not require additional surgical expertise. Several authors, including Adamyan, Soong et al., and Templeman and colleagues, have described the laparoscopic use of the peritoneum to create a neovagina in patients with vaginal agenesis. Adamyan and Soong et al. reported a group of 45 patients without significant postoperative complications. The most common postoperative problem involved the formation of granulation tissue at the vaginal vault. Templeman and colleagues described the laparoscopic mobilization of peritoneum for the creation of a neovagina in only one patient. The peritoneum was grasped through a perineal dissection and sutured to the introitus. A purse-string closure was placed at the apex. Stenting of the neovagina was continued for 3 months postoperatively followed by rigid dilator use. At 9-month follow-up evaluation, an 8- by 2-cm vagina was described, with squamous epithelialization present. Both groups describe the technique as safe and efficient, producing a neovagina with apical granulation tissue as the only complication. Bianchi and colleagues reported on eighty patients with Rokitansky syndrome. Forty patients underwent a Davydov procedure. The team highlighted that the Davydov procedure is a better choice in patients with female hypospadias but may have more risk in patients with a pelvic kidney or patients with previous pelvic surgery and adhesions. They also argued that the Davydov approach immediately achieves good functional results and there is less need for vaginal dilation than other methods to maintain the end result. They found a mean vaginal length of 7.25 + 2.1 cm at discharge and 8.5 + 1.6 cm at 12 months postprocedure. At 12 months postprocedure, the FSFI was completed by all the patients in the study with globally optimal results. No important complications occurred, although one patient experienced introital stenosis, which did not require surgery. However, they also again reinforced the risk of significant complications including rectal and bladder perforation. Lastly, they commented on potential limitation of the Davydov in the situation of postoperative failure. They suggest that this procedure yields poor chance of surgical correction due to the difficulty of recanalization of the urethrovesicorectal space. Giannesi and colleagues reported an assessment of sexuality via a self-report questionnaire after the laparoscopic Davydov procedure. The FSFI was also administered to both 28 women who underwent a laparoscopic Davydov procedure and 28 agematched controls. The anatomical result was judged to be satisfactory (>6 cm) in 26 of 28 patients with a mean vaginal length of 7.2 ± 1.5 cm. No statistical difference was found between the subjects and the controls in all six domains of the FSFI; however, the authors note that 6 of the 25 (24%) operative subjects who completed the entire FSFI had a poor FSFI result. Of note, the areas of greatest concern were in the areas of lubrication and pain. Liao and colleagues reported on 31 patients with MRKH who underwent surgery with a laparoscopic Davydov procedure. Seven patients were lost to follow up, so only 24 patients completed the surgery and requested follow-up. The FSFI was administered to evaluate function of patients who became sexually active and compared them with 50 randomly selected, age-matched healthy women. There was no significant difference in the frequency of sexual intercourse between the control group and the 20 cases who were sexually active. There was no statistical difference in scores of all six domains of the FSFI between the cases and the control subjects. The data from both groups suggest positive functional results.

The Vecchietti operation was first described in 1965 by Giuseppe Vecchietti. He subsequently reported his cumulative 14-year experience in 1979 and 1980. Veronikus and colleagues reviewed the use of the technique and described a laparoscopic modification that uses cystoscopy to confirm bladder integrity. The Vecchietti procedure is a surgical technique for the treatment of vaginal agenesis that constructs a dilatation-type neovagina in 7 to 9 days. The procedure uses specialized equipment including a traction device, a ligature carrier, and an acrylic-shaped olive. The process is in two steps, with essential operative and postoperative components. The operative phase involves positioning the olive at the perineum and the traction sutures extraperitoneally. Classically performed through a Pfannenstiel incision, the ligature carrier introduces the suture into a newly dissected vesicorectal space. The olive is threaded with suture at the perineum, and the suture is reintroduced at the abdomen. The suture is then guided lateral to the rectus muscles bilaterally in a subperitoneal fashion and advanced along the sidewall. The traction device, which provides constant traction on the olive, is positioned on the abdomen. During the postoperative invagination phase, the neovagina is created by applying constant traction to the olive. The process reportedly occurs at a rate of 1.0 to 1.5 cm per day, developing a 10- to 12-cm vagina in 7 to 9 days. Prior to being discharged, patients are instructed on how to use a vaginal obturator on an outpatient basis. Borruto reported on Vecchietti's personal series of vaginal agenesis patients, comprising 522 consecutive patients. The surgical complications included one bladder and one rectal puncture with the ligature carrier and three cases of vaginal vault bleeding. At 100% follow-up at 1 month, dyspareunia was initially reported to be 12%, but resolved in all cases by 3 months. There were no reported failures of the neovaginal construction with 1- and 2-year follow-up of 70% and 30%, respectively.

Modifications of the Vecchietti approach include the use of laparoscopy and elimination of the dissection of the vesicorectal space. The first description of a laparoscopic modification was published by Gauwerky and colleagues. The vesicorectal space was dissected laparoscopically. The threads of the olive device were positioned using a probe introduced into the abdomen through the perineum. Six small abdominal incisions were used for laparoscopic instruments and the traction springs of the specialized device. In 1995, Laffarque and others described a laparoscopic intervention, creating a neovagina in three patients without dissection of the vesicorectal space. At completion of the procedure, cystoscopy was used to confirm bladder integrity. Some experts believe the theoretical risk of bladder or rectal perforation without the dissection of the vesicorectal space is unacceptably high. Brucker and colleagues in Tübingen, Germany, reported on their experience with the Vecchietti procedure in 101 patients with 93 treated for vaginal agenesis. They stratified their patients to three groups: Group 1 with conventional instruments and tunneling in the vesicorectal space, Group 2 included conventional instruments and no dissection in the vesicorectal space, and Group 3 combined both

specialized instruments and no vesicorectal dissection. Group 3 had the longest mean vaginal length immediately postprocedure (9.6 cm compared to 8.9 cm in Group 1 and 7.8 in Group 2) and shortest traction time (4.8 days compared to 11.7 days in Group 1 and 7.5 days in Group 2). Mean followup for Group 2 was 37.7 months compared to 15.5 months in Group 3. Without tunneling and the new specialized instruments, the mean operative time was decreased from 113.0 to 47.5 minutes with a significant reduction in complication rate for bladder lesions and no bowel lesions. Bianchi and colleagues also described their experience with 80 patients who underwent laparoscopic creation of neovagina. Forty of these patients underwent a laparoscopic Vecchietti procedure. Mean operating room time was 30 ± 9.6 minutes. Their group did not perform a Vecchietti procedure for patients with female hypospadias due to risks to the urethra with introital pressure. Preferentially, they performed a laparoscopic Vecchietti for patients with a pelvic kidney, in lieu of other laparoscopic options. They noted that the Vecchietti is easier as a laparoscopic procedure since the perineal approach is not necessary with the absence of dissection in the vesicorectal space. When comparing the outcomes to the laparoscopic Davydov procedure, they reported initially longer vaginal length with the Davydov approach (6.3 ± 0.7 cm for Vecchietti, 7.25 ± 2.1 cm for Davydov), which persists at 12 months; however, both remain within the clinically normal range (7.5 ± 1.1 cm for Vecchietti, 8.5 ± 1.6 cm for Davydov). Additionally, there were no important differences in sexual quality of life, as demonstrated by FSFI scores. Fedele and colleagues modified the approach to use a combined laparoscopic-ultrasonographic technique. The ultrasound assists in identifying the space of connective tissue between the bladder and rectum. The operating time for this modified procedure was only 40 minutes. After 10 days, the patient engaged in sexual intercourse. One-month evaluation confirmed a 12-cm vaginal length. Long-term follow-up outcomes are not available. This technique may even be considered in patients who have been treated for imperforate anus with vaginal agenesis who were not treated in childhood. Sexuality after the laparoscopic Vecchietti procedure requires more data to fully assess. Fedele and colleagues provided the initial report, describing 50 of 52 cases (96%) with a vaginal length greater than 7 cm. All patients succeeded in having vaginal intercourse, 49 (82.6%) had a stable sexual relationship, and 49 (94.2%) were globally satisfied with their sexual life. Since then, the FSFI was introduced and validated as a tool to assess the sexual function in patients undergoing reconstruction. In 2008, Fedele reviewed the entire population, which at that time included 110 patients, including the 27 patients who completed the FSFI at 12 months after surgery. No significant difference was found between the patients and the controls

in the domains regarding desire, arousal, and satisfaction. The scores for the domains on lubrication, orgasm, and comfort, as well as in the total score, resulted in a statistically significant difference, with slightly lower scores for patients with Rokitansky. The lowest scores were in patients who had low scores for desire and arousal, with the authors suggesting that this may actually reflect a more pervasive psychological aspect affecting the total score.

Urologic surgeons involved in urogenital reconstruction are familiar with the role of buccal mucosa in the reconstruction of the urethra and mucosally lined genital surfaces. More recently, gynecologic surgeons have reported the use of buccal mucosal grafts in surgical creation of a complete neovagina. Reports have described using buccal mucosa that was fenestrated prior to grafting to increase the size of the graft. Patients were initially hospitalized with the graft in place and then discharged to continue mold use at home for several weeks. Authors reported vaginal lengths greater than 8 cm; however, limited patient numbers and abbreviated follow-up have been reported. Perhaps, the most advantageous role for buccal neovaginal grafting may be in augmentation vaginoplasty for patients who have had prior suboptimal attempts at vaginal reconstruction or those with more complex reconstructive needs, such as DSD conditions or cloacal anomalies. Benefits of this procedure include the tissue similarities compared to native vagina, an easily accessible source with excellent healing and no visible scar. More data are necessary to weigh the possible disadvantages of numbness of the lip/cheek and difficulty opening the mouth due to scarring or contracture at the graft donation site.

Bowel vaginoplasty is a well-known alternative for creation of a neovagina. The Ruge procedure and others are characterized by the formation of a neovagina using sigmoid colon grafts. Advocates propose that scar formation and vaginal stenosis occur less often than with other procedures; however, the disadvantage is the necessity of an abdominal laparotomy. Ota and colleagues reported a laparoscopic-assisted Ruge procedure. Mesenteric dissection and sigmoid resection were performed laparoscopically. A 3.5-cm incision was used for appropriate bowel suturing. The segment of sigmoid colon was mobilized and brought to the introitus. The serosal layer of the pediculate end was stabilized to pelvic peritoneum. The patient remained hospitalized for 14 days. The benefit of this modification is certainly the accomplishment of a difficult surgical procedure endoscopically. Other advantages include the functional, ample vaginal length (12 cm) without postoperative dilatation. Disadvantages include the extended postoperative hospitalization period and the small number of patients evaluated.

Reported surgical outcomes by authors such as Hensle and colleagues and Communal and associates support the role of sigmoid neovaginoplasty, especially in patients with MRKH. Until recently, long-term data regarding sexual function had not been available. Parsons and colleagues retrospectively reviewed 28 cases at a mean of 6.2 years after sigmoid vaginoplasty. Seventynine percent of patients were reportedly “very satisfied” with sexual function, and 21% were “comfortable” with the outcome. Communal and colleagues administered the FSFI to 11 patients after creation of a sigmoid neovagina. The mean score of the 8 women who were attempting intercourse was reported to be equivalent to that of “normal” patients without vaginal agenesis. Carrard et al. reported on 59 patients with MRKH, 11 treated with the Frank method and 48 who underwent sigmoid vaginoplasty. The mean time after surgery was 6 years (10 months to 17.8 years). Forty patients (68%) answered the questionnaire, 35/48 (73%) who had surgery and 5/11 (45%) who were treated with dilation. The mean total FSFI score was 28 in the operated group: 21 patients of the 30 currently sexually active respondents (70%) had a score above the cutoff for sexual dysfunction. Women with MRKH syndrome treated with sigmoid vaginoplasty could be considered “normal” in terms of desire, arousability, lubrication, orgasm, and global sexual satisfaction. However, discomfort scores were higher in these patients. The only significant between-group difference was in terms of vaginal discharge discomfort. In comparison, in the group treated by the Frank method, the mean total FSFI score was 30 and each domain score was similar to “normal” women's score. Three patients (75%) had a score above the 26.55 cutoff. Hensle and colleagues provided additional information by reporting the sexual function of 57 patients who underwent creation of a bowel neovagina. Forty-two of the patients were reported to have MRKH. Eight patients underwent ileovaginoplasty; two patients were then subsequently treated with a colonic neovagina. The follow-up period varied from 18 months to 24 years with a mean of 8.8 years. Outcomes

were evaluated both by a retrospective chart review and the female sexual dysfunction questionnaire described as an Institutional Review Board-approved, validated instrument. Of the 36 patients who responded, 31 were sexually active. Seventy-eight percent of the entire group of patients, including additional diagnoses precipitating neovaginoplasty, reported sexual desire, 33% sexual arousal, 33% sexual confidence, and 78% sexual satisfaction. Also, 56% reported frequent orgasms, 22% occasional orgasms, and 22% no orgasms. Thirty-two patients (89%) reported adequate lubrication for sexual activity; 34 patients used home douching, and 20 required pads for mucus production. Djordjevic and colleagues reported on the use of the rectosigmoid in neovaginal replacement. They reported on 86 patients, of whom 54 had vaginal agenesis. The mean follow-up was 47 months (range 8 to 114 months). The mean vaginal length was 13 cm in patients with vaginal agenesis. Mucous production was reported in 19/54 (35%) in the first 6 months; however, after 6 months, only 2 (3.7%) patients noted the complaint. Introital stenosis and vaginal prolapse were each only reported in 2 cases (3.7%). Sexual outcome was evaluated with the FSFI with sexual function described as satisfactory in 46/54 patients (85.18%).

One of the long-term risks with colonic neovaginal replacement is the possible involvement of the neovagina in systemic inflammatory bowel disease. Only a handful of cases have been reported; however, involvement of the neovagina has been reported as significant, requiring reoperation and repeat vaginal replacement. Endoscopic surveillance with biopsy should be considered essential in patients with symptoms of unexplained discharge and pain.

Makinoda and colleagues reported a nongrafting method of vaginal creation. This group reported 18 women who underwent a two-step protocol. The initial step used noninvasive dilatation using a vaginal mold based on the technique of Frank. The second step was a surgical procedure via a perineal approach. The apex of the dilated vaginal space was incised, and further dissection between the bladder and rectum was carried to the peritoneal cavity. After peritoneal perforation, the uterine structures, when present, were pulled down and sutured to the newly created vaginal space. A firm vaginal mold was inserted and recommended for use for 6 months postoperatively. The authors propose a benefit of avoidance of grafting. The time course for success with the initial dilatation step may be unacceptably long in many patients (mean = 10.90 ± 9.8 months). No significant surgical complications were reported despite the theoretical risk of ureteral contortion and kinking when pulling the rudimentary uterine structures inferiorly. During the follow-up period, shrinkage of the vaginal length and diameter was noted in some patients who had been noncompliant with the mold or without coitus. The authors noted a minimal vaginal length of 5 cm in a patient who was noncompliant with the mold and not sexually active. The authors dispute the necessity of any lining of the neovaginal space. In their experience, significant narrowing or contraction of the margins did not occur. They maintain that the vaginal space is maintained by suturing the muscular buds of the uterus to the pressure-created neovaginal space.

A spatial W-plasty technique using a full-thickness unilateral groin graft has been described in a limited number of patients by Chen and others. The authors advocate an earlier intervention with the premise that a full-thickness graft may grow as the patient grows. This is recommended to eliminate psychological issues in patients who would be treated during the late teens, when sexual identity may be forming.

An exciting innovation is the use of autologous in vitro grown vaginal tissue. Panici and colleagues in Rome reported a two-step procedure. The first step involved acquiring a fullthickness biopsy of the vestibule. The tissue was processed and incubated. The time interval between the biopsy and the fully differentiated mucosal tissue for grafting was 2 weeks. When the tissue was ready for grafting, the vaginal canal was prepared in the Abbe-McIndoe technique and gauze was placed with the cell stratum facing the newly created vaginal canal walls. Surgical time was reported as 18 minutes for the second step, and estimated blood loss was less than 100 mL. The gauze was kept in place with a mold for 5 days; the mold was then removed, and the vagina was irrigated. Estimated success was 90%. Although this technique is in its infancy, the benefit of having the neovagina lined with physiologic vaginal tissue is significant both for function and psychological implications. Challenges include the logistics of the two-step procedure and the need for capabilities for tissue culture. Additional experience with this type of orthotopic vaginal grafting may lead to groundbreaking changes in the treatment of this condition.

Acquired Vaginal Insufficiency

Unusual types of infection and atrophy can rarely cause closure of part of the vagina, but acquired vaginal inadequacy most often is the result of treatment of various gynecologic malignancies with surgery or radiation, or a combination of both. Restoration and maintenance of vaginal function are important elements of the treatment plan for such malignancies, especially when the patient is young and otherwise healthy. The unique surgical challenges associated with creation of a neovagina in patients treated with radiation therapy require the experience and expertise of surgeons familiar with this population. The techniques of vaginal reconstruction in gynecologic oncology have been reviewed by Magrina and Masterson, by Pratt, and by McCraw and associates.

DISORDERS OF VERTICAL FUSION

The problems associated with vertical fusion include transverse vaginal septum with or without obstruction. Although imperforate hymen is a vertical fusion problem, the hymen is not a derivative of the müllerian ducts; therefore, this condition is discussed elsewhere (see Chapter 24).

Transverse Vaginal Septum

No reliable epidemiologic data exist regarding the incidence of transverse vaginal septum. Reported incidences vary from 1 in 2,100 to 1 in 72,000. It is probably less common than congenital absence of the vagina and uterus. It has been diagnosed in newborns, infants, and older adolescent girls. Its etiology is unknown, although McKusick has suggested that some and perhaps most cases are the result of a female sex-limited autosomal recessive transmission. There is a developmental defect in vaginal embryogenesis that leads to an incomplete fusion between the müllerian duct component and the urogenital sinus component of the vagina. The incomplete vertical fusion results in a transverse vaginal septum (AFS class IIA) that varies in thickness and can be located at almost any level in the vagina (**Fig. 25.10**). Lodi has reported that 46% occur in the upper vagina, 40% in the midvagina, and 14% in the lower vagina. Rock, Zacur, and associates have noted septa in the upper, middle, and lower thirds of the vagina in 46%, 35%, and 19% of patients, respectively. In general, the thicker septum is noted to be more common closer to the uterine cervix. In contrast to congenital absence of the müllerian ducts, the transverse vaginal septum is associated with few urologic or other anomalies. Imperforate anus and bicornuate uterus can be found, as

reported by Mandell and colleagues. The lower surface of the transverse septum is always covered by squamous epithelium. The upper surface can be covered by glandular epithelium, which is likely to be transformed into squamous epithelium by a metaplastic process after correction of the obstruction.

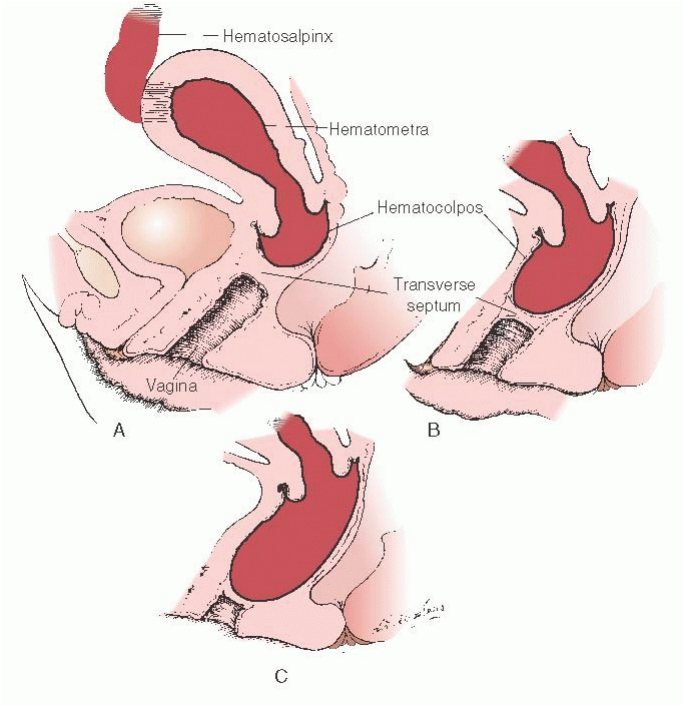


FIGURE 25.10 Positions of septum responsible for complete vaginal obstruction. High **(A),** mid- **(B),** and low **(C)** transverse vaginal septa. Note the position of the hematocolpos. Lower vaginal septa allow more blood to accumulate in the upper vagina. The vaginal mass shown in **(C)** is more accessible through rectovaginal examination.

In neonates and young infants, imperforate transverse vaginal septum with obstruction can lead to serious and life-threatening problems caused by the compression of surrounding organs by fluid that has collected above the septum. The fluid undoubtedly comes from endocervical glands and müllerian glandular epithelium in the upper vagina that have been stimulated by the placental transfer of maternal estrogen. Continued fluid collection in infants, even after the 1st year, has been reported; thus, the possibility of a fistula between the upper vagina and the urinary tract should be considered. The distended upper vagina creates a large pelvic and lower abdominal mass that can displace the bladder anteriorly, displace the ureters laterally with hydroureters and hydronephrosis, compress the rectum with associated obstipation and even intestinal obstruction, and limit diaphragmatic excursion to indirectly compress the vena cava and produce cardiorespiratory failure. Fatalities have been reported. The hydrocolpos develops along the axis of the upper vagina and therefore may not necessarily cause the outlet or perineum to bulge when there is compression of the mass from above. After careful preoperative radiologic and endoscopic investigations of the infant, the septum should be removed through a perineal approach. Bilateral Schuchardt incisions may be required to ensure that the septum has been removed. Because of the subsequent tendency for vaginal stenosis and reaccumulation of the fluid in the upper vagina, follow-up studies to assess the recurrence of urinary obstruction are important. Vaginal reconstruction may be required in later years to allow satisfactory menstruation and coitus.

A hematocolpos may not develop until puberty. Symptoms include cyclic lower abdominal pain, no visible menstrual discharge, and gradual development of a central lower abdominal and pelvic mass. Sometimes, a small tract opens in the septum, some menstrual blood escapes periodically, and symptoms are variable. A septum large enough to allow pregnancy to occur can still cause dystocia during labor. Cyclic hematuria may be present if a communication between the bladder and upper vagina exists. The pelvic organs of a woman with a transverse vaginal septum are shown in **Figure 25.11.** The woman developed severe cyclic pain at the time of onset of menstruation, but there was no external bleeding until menstrual blood finally began to flow through the small sinus. Pelvic examination per rectum revealed a cervix and a normal-sized corpus. The ovaries were palpable but adherent, probably because of organized blood from hematosalpinx and hematoperitoneum. Remarkably, the woman had little dysmenorrhea after beginning to menstruate externally. Coitus was fairly satisfactory before surgical correction, but the shortness of the vagina was something of a handicap. The obstructing membrane was excised, and an anastomosis of the upper and lower vagina was performed.

The findings of 26 patients with complete transverse vaginal septum reported from the Johns Hopkins Hospital by Rock, Zacur, and colleagues have shown that associated congenital anomalies include urinary tract anomalies, coarctation of the aorta, atrial septal defect, and malformations of the lumbar spine. Vaginal patency and coital function were successfully established in all patients, and 7 of 19 patients

attempting pregnancy eventually had children. The incidence of endometriosis and spontaneous abortion was high. A lower pregnancy rate and more extensive endometriosis were present when the transverse septum was located high in the vagina, suggesting that retrograde flow through the uterus and fallopian tubes occurs earlier in these patients. More extensive dissection between the bladder and rectum was required to identify the upper vagina when the septum was thick and high. Exploratory laparotomy was necessary in five patients to guide a probe through the uterine fundus and cervix and to assist in locating a high hematocolpos.

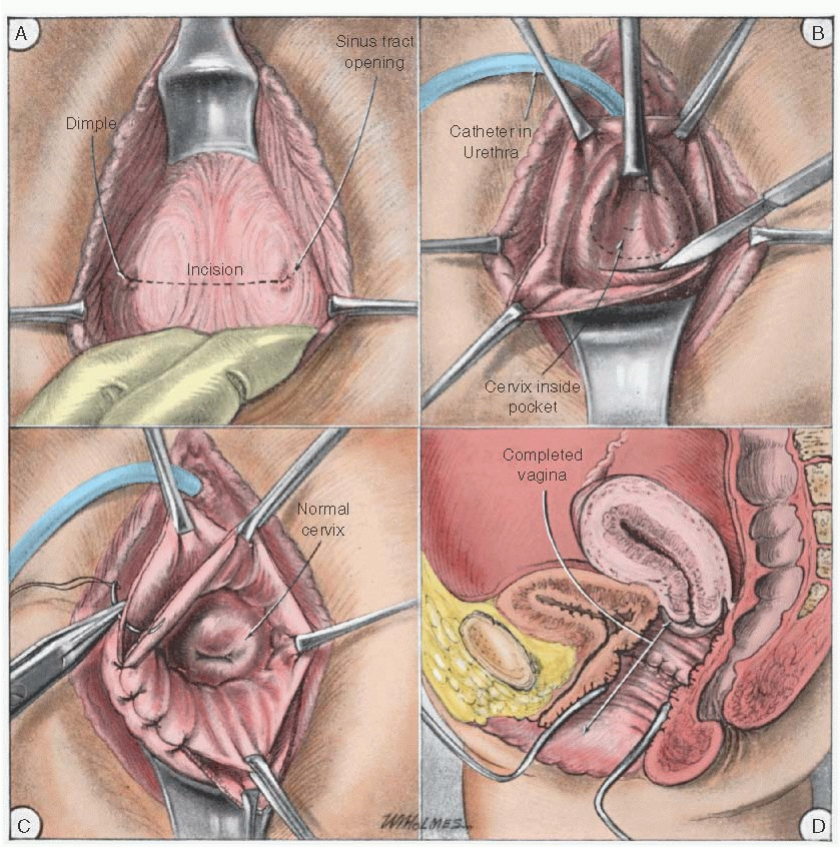


FIGURE 25.11 Surgical correction of transverse vaginal septum. **A:** The upper end of a short vagina. The small sinus tract opening, through which the patient menstruated, is shown. The line of incision is drawn through the mucous membrane between the vaginal dimple and the sinus. **B:** Areolar tissue is dissected through to the pocket of mucosa that covered the cervix. The mucosa is incised. **C:** An anastomosis is made between the lower vagina and the upper vagina. **D:** Completed vagina. It is slightly shorter than normal but of normal caliber.

Surgical Technique for a Transverse Vaginal Septum

A transverse incision is made through the vault of the short vagina (Fig. 25.11A). A probe is introduced through the septum after a portion of the barrier has been separated by sharp and blunt dissection. The physician usually finds some areolar tissue in dissecting the space between the vagina and the rectum. Palpation of a urethral catheter anteriorly and insertion of a doublegloved finger along the anterior wall of the rectum posteriorly provide the proper surgical guidelines so that the bladder and rectum can be avoided during this blind procedure. After the dissection is continued for a short distance, the cervix can usually be palpated, and continuity can be established with the upper segment of the vagina (Fig. 25.11B, C). The lateral margins of the excised septum are extended widely by sharp knife dissection to avoid postoperative stricture formation. The edges of the upper and the lower vaginal mucosa are undermined and mobilized enough to permit anastomosis with the use of interrupted delayed absorbable sutures (Fig. 25.11C). Figure 25.11D shows the completed anastomosis with a vagina that is of normal caliber but has a length slightly shorter than average. A soft foam rubber vaginal form covered with a sterile latex sheath can be placed in the vagina and removed in 10 days for evaluation of the healing process. The form can be worn for 4 to 6 weeks until complete healing has occurred. After this, coitus is permitted. If the patient is not sexually active, then vaginal dilatation may be necessary to maintain established patency. Alternatively, a silicone elastomer (Silastic) vaginal form can be inserted at night until the constrictive phase of healing is complete. A combination of a perineal (transvaginal) and

laparoscopic approach may also be useful. Laparoscopy can provide assistance with mobilization of the upper vagina, using dissection both anteriorly at the bladder and in the posterior cul-de-sac. This technique can decrease tension on the anastomosis of upper and lower vagina and minimize the risk of stricture.

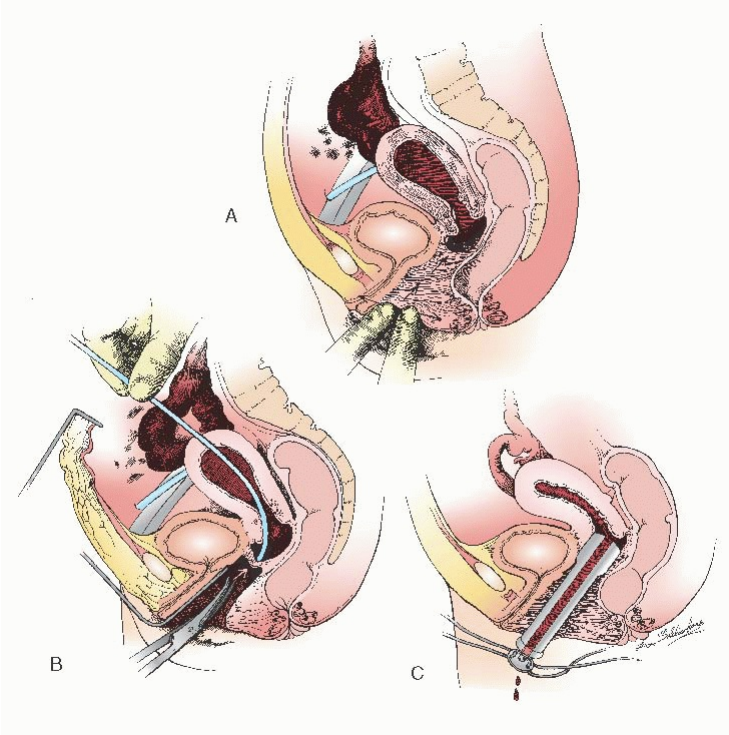


FIGURE 25.12 Correction of an atretic vagina. **A:** A large portion of atretic vagina is palpated with two fingers. Once the vaginal space is developed, it may be necessary to open the abdomen via laparotomy and pass a probe through to the uterine fundus **(B)** to tent out the septum, which may then be safely excised. **C:** An acrylic resin (Lucite) form is then placed into the vagina and secured with rubber straps.

High Transverse Vaginal Septum

If the length of the obstructing transverse vaginal septum is such that reanastomosis of the upper and lower vagina is impossible, as is the case with a high transverse vaginal septum, in which a significant portion of the vagina is atretic, then a space is created between the rectum and bladder to permit identification of the obstructed vagina (Fig. 25.12). The mass that has resulted from accumulated menstrual blood must be distinguished from the bladder anteriorly and the rectum posteriorly, a process that is facilitated by the mass itself. When differentiation is impossible, however, exploratory laparotomy can be performed. During this procedure, a probe is passed through the fundus of the uterus to tent out the vaginal septum and enable the surgeon to excise it from below and resect it safely.

In most surgical procedures to remove the high transverse vaginal septum, the obstructing membrane can be readily identified (Fig. 25.13), after which the operator can probe the mass with an aspirating needle to identify old menstrual blood. The upper vagina is then opened and the septum excised. Because the distance between the septum and the upper vagina is too great to permit an anastomosis, an indwelling acrylic resin (Lucite) form, consisting of a bulbous end and a channel through which menstrual blood can drain, is placed into the vagina and anchored with a retaining harness. The bulbous end of the form, in most instances, is retained in the upper vagina and should be left in place for 4 to 6 months while epithelialization is accomplished. After its removal, vaginal dilatation should be practiced on a daily basis for 2 to 4 months to prevent contracture of the space. It is essential to the success of the operation that the new space not become constricted; to avoid constriction, the form must be worn for many months during the constrictive phase of healing. As an alternative to the Lucite form, the physician can consider using a split-thickness graft to bridge the gap. The graft is usually sutured in situ in the vagina rather than sutured to a form. An ingenious but rather complicated Z-plasty method of bridging the gap has been described by Garcia and by Musset. A simpler flap method was described by Brenner and associates.

A transverse vaginal septum diagnosed after the onset of puberty presents numerous problems. Often, a large segment of the vagina is absent, making anastomosis of the upper and lower segments difficult. Furthermore, postoperative vaginal dilatation is necessary to prevent stenosis at the anastomosis site. Poor compliance with dilatation in a poorly motivated pubertal patient is always a concern. However, rarely is the surgeon able to delay vaginoplasty until the patient is more mature because of increasingly severe cyclic abdominal pain caused by the hematocolpos. Thus, a difficult vaginoplasty can have less than optimal results.

Hurst and Rock have described an alternative approach to maximize surgical resection and anastomosis in women with a high transverse vaginal septum. Aspiration of the hematocolpos under ultrasound guidance was necessary to relieve the acute pain and delay surgery. Continuous oral contraceptives were used to delay recurrence of hematocolpos. Most important, vaginal dilatation was used to lengthen the lower vaginal segment to facilitate resection and reanastomosis (Fig. 25.14). In all three patients, the approach was successful.

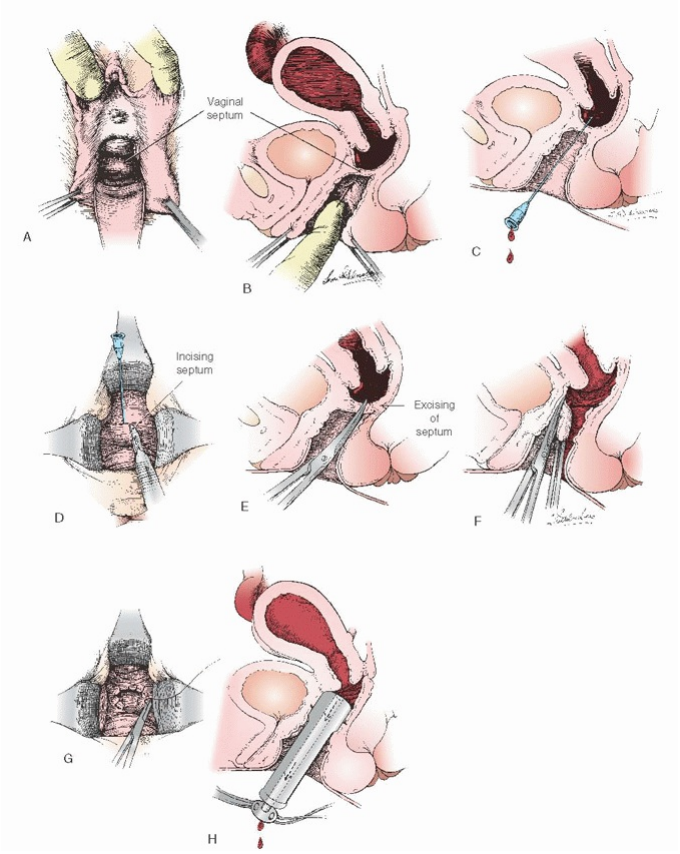


FIGURE 25.13 A high transverse vaginal septum. **A:** The neovaginal space is dissected, revealing a high obstructing vaginal membrane. **B:** This can be palpated with the middle finger. **C:** A needle is then placed into the mass. **D:** The incision is made with a sharp knife, and considerable bleeding can occur. **E:** The septum is excised. **F:** The septum is removed. **G:** After the septum is removed, the wall of the septum is oversewn with interrupted sutures of 2-0 chromic catgut. **H:** Because the distance between the septum and the upper vagina is too great to allow anastomosis, an acrylic resin (Lucite) form is placed in the vagina so that epithelialization can occur over the form while vaginal patency is maintained. The form, in place, is fitted with a plastic retainer. Rubber straps can be placed through the retainer and attached to a waist belt to allow constant upper pressure so that the form is retained in the upper vagina. Modification of this method includes a small adapter to allow drainage through the acrylic resin (Lucite) form, preventing the accumulation of old blood and mucus in the upper vagina.

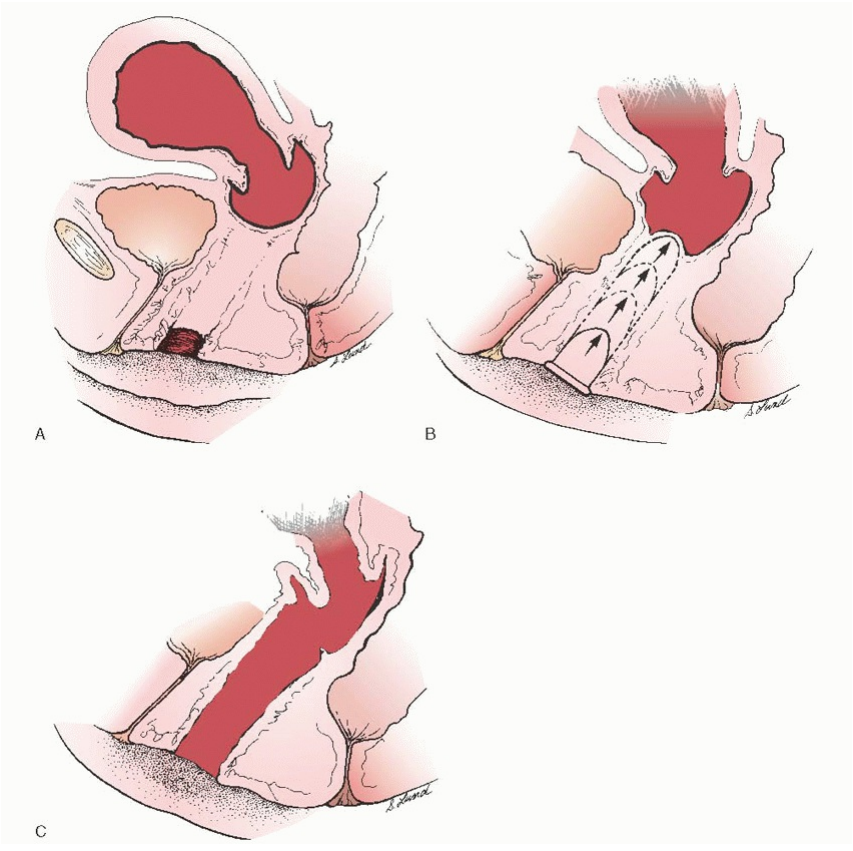


FIGURE 25.14 A: High transverse vaginal septum demonstrating a small hematocolpos and hematometra. Upper to lower vaginal anastomosis at this stage can result in stenosis at the anastomosed site. **B:** Vaginal depth is increased with passive dilatation using progressively larger dilators. **C:** A primary upper to lower vaginal anastomosis can be performed easily after dilatation.

Congenital Absence or Dysgenesis of the Cervix

Agenesis or atresia of the cervix (AFS class IIB) is a relatively infrequent müllerian anomaly. When this anomaly does occur, it is often in association with absence of a portion or all of the vagina. In many cases of cervical agenesis or atresia, retention of menstrual blood initiates symptoms of cyclic lower abdominal pain without menstrual flow, causing the patient to seek gynecologic evaluation and care. In past times, diagnosis was suspected on the basis of a history and physical findings but was not proved until the time of surgery. Today, diagnosis of cervical agenesis or atresia is still usually difficult before operation, but the possibility of making a correct diagnosis before surgery does exist, with the help of modern diagnostic tools. Early diagnosis offers significant advantages in patient care, the most important of which is effective presurgical planning and preparation.

Diagnosis of Cervical Dysgenesis

Patients with congenital absence of the cervix present a diagnostic challenge. Patients with cervical aplasia with a functioning midline uterine corpus have aplasia of the lower two thirds of the vagina with an upper vaginal pouch. Similarly, some patients have a considerable atretic segment of vagina and an upper vaginal pouch with a properly developed uterine cervix and corpus above. Differentiation of these two müllerian anomalies is essential. Ultrasonography may be helpful. Valdes and associates have reported the use of preoperative ultrasonography in the evaluation of two patients with atresia of the vagina and cervix. Magnetic resonance imaging has been found to be

helpful in confirming this diagnosis, as reported by Markham and associates. The lower uterine segment and cervical tissue can be carefully examined (**Fig. 25.15**). With cervical dysgenesis, there is no vaginal dilatation with the accumulation of blood, as seen with a high transverse vaginal septum. Both ultrasonography and MRI are most helpful when they are correlated with the findings of a careful pelvic examination under anesthesia.

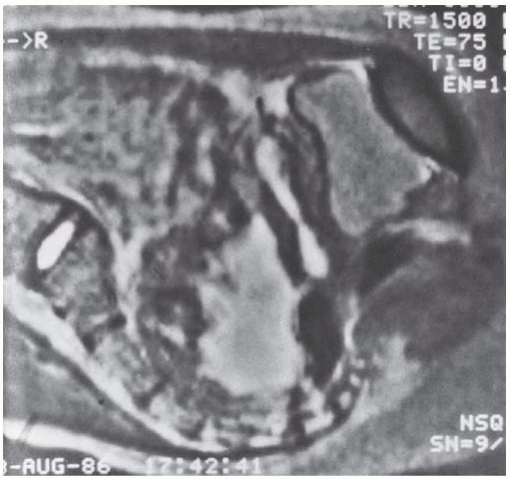


FIGURE 25.15 Magnetic resonance T1-weighted image showing atretic segment of distal cervix. The tip of an atretic cervix is shown. No vagina is noted.

Anatomic Variations of Congenital Cervical Anomalies

Two basic categories of cervical anomalies have been observed in several configurations. Patients exhibiting the first type, cervical aplasia, lack a uterine cervix (**Fig. 25.16A**), and the lower uterine segment narrows to terminate in a peritoneal sleeve at a point well above the normal communication with the vaginal apex. Omurtag and colleagues' report of uterine torsion and acute pain in a 13-year-old with hematometra reminds the reconstructive gynecologic surgeon of the consideration of uterine torsion in the absence of a well-formed cervix. The uterus can be susceptible to rotation on its horizontal axis, particularly with the mass effect of hematometra. The second type, cervical dysgenesis, can be described in four subtypes:

1. Cervical body consisting of a fibrous band of variable length and diameter (endocervical glands may be noted on pathologic examination) (**Fig. 25.16B**)
2. Intact cervical body with obstruction of the cervical os (the cervix is usually well formed, but a portion of the endocervical lumen is obliterated) (**Fig. 25.16C**)
3. Stricture of the midportion of the cervix (which is hypoplastic with a bulbous tip and no identifiable cervical lumen) (**Fig. 25.16D**)
4. Fragmentation of the cervix (with portions that can be palpated below the fundus and that are not connected to the lower uterine segment) (**Fig. 25.16E**)

Associated anomalies of the urinary tract are rare, but they do occur. Variable portions of the vagina can be atretic. Cervical obstruction is most often associated with a vagina of normal length.

Treatment

When both the vagina and cervix are absent and a functioning uterine corpus is present, it is difficult to obtain a satisfactory fistulous tract through which menstruation can occur. Many methods have been tried, most of them involving creation of a passage through the dense fibrous tissue between the uterine cavity and the vagina and placement of a stent to keep the tract open. Occasional successes in maintaining an open passageway and normal cyclic menstruation have been reported, but endocervical glands do not develop, and there is no way to compensate for the absence of the cervical mucus, which plays an important role in sperm transport. Even though cyclic ovulatory periods can be achieved in a few patients, pregnancy is unlikely. Eventually, the uterovaginal tract closes from constriction by fibrous tissue. Endometriosis can develop along the tract. Endometriosis also can develop in ovaries and other pelvic sites because of retrograde menstruation. Recurrent and severe pelvic infection is a common problem and may require total hysterectomy and removal of both ovaries. As in vitro fertilization procedures began to offer the possibility for a host uterus to carry a pregnancy to term, procedures to establish a fistulous tract were abandoned. Nevertheless, Cukier and associates in 1986 reported treating a patient with congenital absence of the cervix by construction of a splint that extended into the neocervical canal such that a split-thickness skin graft could actually be placed within the endocervical canal. This patient has continued to menstruate without difficulty, although pregnancy has not been accomplished.

Many authors have recommended hysterectomy as an initial procedure for a patient with a functioning uterine corpus and congenital absence of the cervix and vagina. A hysterectomy eliminates much needless suffering from associated problems such as cryptomenorrhea, sepsis, endometriosis, and multiple operations. If the hysterectomy is performed soon enough, before the problems become great, it may be possible to conserve the ovaries and their useful functions. The reconstructive surgeon should be prepared to perform a vaginoplasty if hysterectomy is performed, particularly if there has been a vaginal dissection. If the neovaginal space is allowed to close and scar, then future operations to develop an adequate neovagina are associated with increased risks of graft failure and fistula formation.

Despite the overall poor results from reconstruction for congenital absence of both the cervix and the vagina, clinical experience suggests that cannulization procedures can be worthwhile in a few carefully selected cases with adequate stroma to allow a cervicovaginal anastomosis. If a long segment of cervix is fibrous cord, a cervical grafting technique may be required. If a fragmented cervix is noted, then hysterectomy is usually warranted. The few patients who have achieved a pregnancy after cervical reconstruction have had a well-formed cervical body. Anecdotal reports of resection of the dysgenetic cervix and reconstruction with uterovaginal anastomosis may be promising. The isthmus of the uterus is anastomosed directly to the vagina as in patients with cervical malignancy who undergo trachelectomy. Successful pregnancies have

been reported. More experience with this type of reconstruction is necessary to fully understand the unique challenges that may occur in this population compared to patients who experience disease in an otherwise normally developed cervix.

Anecdotal case reports occasionally appear in the literature confirming the necessity of palpable cervical tissue. Letterie described the development of a cervicovaginal tract in an adolescent patient with a core of cervical tissue. The tract remained patent for menstrual flow for 2 years; however, pregnancy had not yet been attempted. Data published by the

senior author regarding the long-term follow-up of 21 patients with abnormal cervical development support the success of cannulization in only selected patients with sufficient rudimentary cervical tissue. All of the patients with fragmentation of the cervix (*n* = 4) eventually underwent hysterectomy. Only those patients with a well-formed cervical body, with at least a palpable cord or only distal obstruction, achieved successful surgical outcomes (4/7 patients). Only one patient with distal obstruction treated with cannulization using a full-thickness skin graft achieved pregnancy.

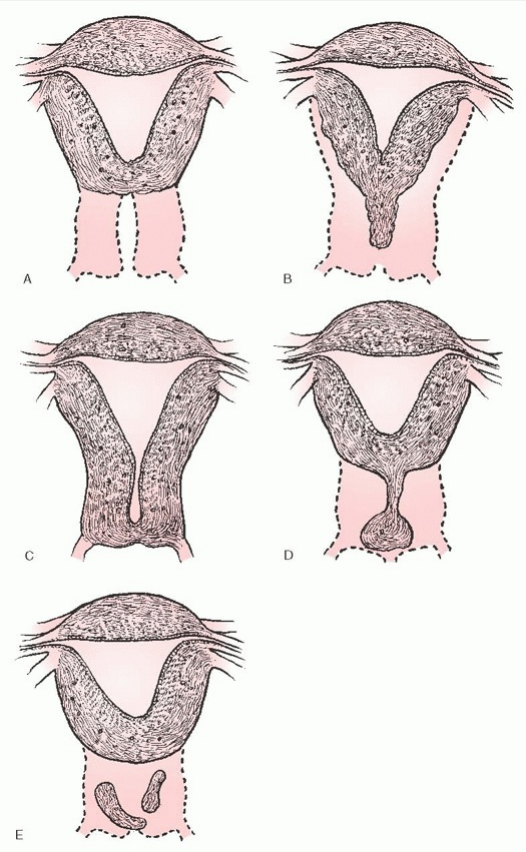


FIGURE 25.16 Congenital cervical anomalies. **A:** The fundus of the uterus is noted without a cervix. **B:** The cervical body consists of a fibrous band of variable length and diameter that can contain endocervical glands. **C:** The cervical body is intact with obstruction of the cervical os. Variable portions of the cervical lumen are obliterated. **D:** Stricture of the midportion of the cervix, which is hypoplastic with a bulbous tip. No cervical lumen is identified. **E:** Cervical fragmentation in which portions of the cervix are noted with no connection to the uterine body. Hypoplasia of the uterine cavity can be associated with cervical cord fragmentation.

Others have reported the anastomosis of a well-developed distal vagina to a substantial midline uterine body. Creighton and colleagues described the use of the laparoscope for this procedure; however, Deffarges and associates examined the outcomes of 18 patients after open uterovaginal anastomosis. Twenty-two percent of patients required additional surgery. Eighty-three percent of the cases were associated with upper genital tract complications. Six spontaneous pregnancies also were reported in four of the patients. Despite the small potential for pregnancy, it is imperative to realize the potential for complications, including sepsis and even death as a result of ascending infection. Fedele and colleagues reported successful management of cervical atresia in twelve patients with vaginal aplasia with laparoscopy. All women experienced regular menstrual cycles. The mean vaginal length at 6-month follow-up was 6 cm. At the time of publication, none of the patients had attempted pregnancy. Kriplani and colleagues described 14 consecutive patients with congenital absence of the uterine cervix (**Table 25.6**). Laparoscopic-assisted uterovaginal anastomosis with placement of a silicone stent was performed. McIndoe vaginoplasty was performed concomitantly in patients with vaginal agenesis. Cervical dysgenesis was present in 5 (35.7%) and cervical agenesis in 9 (64.2%). Mean follow-up after surgery was 3.8 + 1.2 years. Postoperatively, all but one patient experienced regular menses and relief of cyclic pain. One patient underwent hysterectomy because of genital infection and restenosis. The authors reported that five patients were sexually active and reported it to be satisfactory. Pregnancy occurred in 3/5 patients. Reoperation and subsequent hysterectomy have been reported in as much as half of patients undergoing cervical reconstruction as the primary procedure. Kriplani attributes the use of the silicone stent in the neocervix until the resolution of inflammation as one of the reasons the incidence of restenosis was decreased in their population. Only one patient (7.4%) required hysterectomy for recurrent infection and stenosis.

TABLE 25.6 Published Reports of Laparoscopic-Assisted Management of Cervical Agenesis and Vaginal Aplasia

SOURCE, YEAR	NO. OF PATIENTS	AGE, YEAR	LAPAROSCOPICASSISTED PROCEDURE	CERVICAL REMNANT	VAGINA	VAGINAL LENGTH AFTER SURGERY, MEAN, CM	SEXUALLY ACTIVE/PREGNANT
Lee et al., 1999	1	12	Reconstruction of cervix and vagina using skin graft	Absent	Absent	7	NA
Creighton et al., 2006	1	16	Uterovaginal anastomosis	Absent	Proximaplasia	NA	1/0
Fedele et	12	12-17	Uterovestibular anastomosis	Present (*n*	NA	6 ± 1.6	6/NA

al., 2008				= 2)			
Raudrant et al., 2008	1	13	Uterovaginal anastomosis	Absent	Absent	Small	NA
El Saman, 2009	4	14/18	Laparoscopic canalization and combined retropubic balloon vaginoplasty	Absent	Absent	9-11	NA
Daraï et al., 2009	1	16	Uterovaginal anastomosis	Absent	Absent	6	NA
El Saman, 2010	5	13-16	Endoscopically monitored canalization	Absent	Absent in 2	NA	2/0
Nguyen et al., 2011	1	21	Laparoscopically assisted reconstruction of PTFE graft lining neocervix	Absent	Present	NA	1/0

NA, data not available or not applicable; PTFE, polytetrafluoroethylene. Modified from Kriplani A, Kachhawa G, Awasthi D, et al. Laparoscopic-assisted uterovaginal anastomosis in congenital atresia of uterine cervix: follow-up study. *J Minim Invasive Gynecol* 2012;19:477, with permission.

The surgical approach for the rare opportunity for cervical cannulation remains a challenge. Several additional factors may also influence the surgical outcome: the size of the created channel, the duration of stenting of the channel, the presence of rudimentary endocervical glands in the region of the created channel, the presence of a native vagina adjacent to the created channel, or the number of menses allowed to flow through the stented channel. Because the surgical approach for cervical cannulation is based only on several case reports, limited

data regarding safety and efficacy are available to share with patients and families. A frank discussion with both the patient and her family regarding the potential risks and morbidity is imperative. Even reportedly successful attempts have involved multiple surgical interventions, such as a reported cervicoplasty with bladder mucosa described by Bugamann and colleagues; the 12-year-old patient had to undergo at least one previously failed attempt at reconstruction before the reported successful procedure. Very young adolescents may be subjected to multiple surgical procedures without good evidence of success. These more aggressive, fertility-sparing procedures may be best suited for patients who have had good menstrual suppression and are more mature at the time of the discussion of risks and benefits and decision for surgery.

DISORDERS OF LATERAL FUSION

Failures of lateral fusion of the two müllerian ducts cause vaginal anomalies that are grouped as obstructed or unobstructed.

The Unobstructed Double Uterus (Bicornuate, Septate, or Didelphic Uterus)

Complete failure of medial fusion of the two müllerian ducts can result in complete duplication of the vagina, cervix, and uterus. Partial failure of fusion can result in a single vagina with a single or duplicate cervix and complete or partial duplication of the uterine corpus. A failure of absorption of the uterine septum between the two fused müllerian ducts causes the septum to persist inside the uterus to a variable extent while the external appearance remains that of a single uterus. The septum can be so complete that it divides both the uterine cavity and endocervical canal into two equal or unequal components. More often, incomplete disappearance of the septum leaves only the upper uterine cavities divided. Each of these and a variety of other forms of double uteri have their own individual features of clinical significance. When no obstruction is present, surgical reconstruction is performed primarily because of difficulties with reproduction.

Some aspects of lateral fusion disorders remain controversial because information is still inaccurate or incomplete. Many reports are based on small samples of selected patients, patients who have been diagnosed as having one anomaly or another based on incomplete data, and patients who have received unification operations without preliminary studies to rule out other causes of reproductive difficulty. A comparison of results from one series to the next is difficult because authors have used different classifications based on a variety of embryologic, anatomic, physiologic, functional, and radiologic considerations. Unknown numbers of uterine anomalies may have escaped detection because reproductive performance is generally acceptable and gynecologic difficulties do not necessarily occur.

The müllerian ducts undergo multiple steps in development, including caudal, medial growth followed by fusion and later resorption of the remaining septum. Apoptosis has been proposed as a mechanism by which the septum regresses. Bcl-2, a protein involved in regulating apoptosis, was found to be absent from the septa of several uteri. The absence of this critical protein may play a pivotal role in the persistence of the septum and lateral fusion disorders.

Historical Development of Surgical Procedures

Ruge, in 1882, first reported excision of a uterine septum in a woman who had suffered two pregnancy losses. The woman subsequently carried a pregnancy to term. Paul Strassmann of Berlin and later Erwin Strassmann, his son, were strong advocates of uterine unification operations. The studies of Jones and Jones have contributed greatly to modern understanding of the management of uterine anomalies. Their studies began with a report in 1953 of a series that was started in 1936. Updates have been published from time to time. Wheeless, Rock, Andrews, and others have joined in these reports.

Diagnosis of Uterine Anomalies

If a uterine anomaly is associated with obstruction of menstrual flow, then it causes symptoms that will come to the attention of the gynecologist shortly after menarche. Unobstructed uterine anomalies are diagnosed later in a variety of circumstances. Young girls may notice difficulty in using tampons or later difficulty with coitus if a longitudinal vaginal septum is present. This can lead to the diagnosis of an associated uterine anomaly. A patient with an anomalous upper urinary tract on intravenous pyelogram may be found to have a uterine anomaly on gynecologic evaluation. A uterine anomaly is occasionally found when a patient reports dysmenorrhea or menorrhagia or when a dilatation and curettage (D&C) is performed. A palpable mass may be a uterine anomaly but should be confirmed as such by ultrasonography, MRI, hysterography, or laparoscopy. Woelfer and colleagues described the use of three-dimensional ultrasonography in screening for congenital uterine anomalies. During an investigation of the correlation of uterine anomalies with obstetric complications, the authors assessed the potential value of three-dimensional ultrasound for screening. More than 100 women with uterine anomalies were identified. Seventytwo arcuate uteri, twenty nine septate, and five bicornuate uteri were described. The authors emphasized how the threedimensional ultrasound may overcome the limitations of conventional two-dimensional ultrasonography in providing a coronal view of the uterus, thus differentiating between arcuate, bicornuate, and subseptate uteri. This technique remains investigational. Semmens has pointed out that the diagnosis of a uterine anomaly can also be made from astute observation of an abnormal uterine contour during pregnancy, either in the antepartum period or at the time of abdominal or vaginal delivery. The abnormal contour is caused by a combination of fetal malpresentation and an anomalous uterus. An anomalous uterus can also be diagnosed when a pregnancy occurs despite the presence of an intrauterine contraceptive device. Persistent postmenopausal bleeding despite recent D&C can lead to a diagnosis of an anomalous uterus. Sometimes the diagnosis is made as an incidental finding at laparotomy. Historically, most uterine

anomalies were diagnosed after hysterosalpingography to evaluate infertility or reproductive loss, usually from repeated spontaneous abortion, but increasingly, uterine anomalies are detected on MRI performed for other indications.

Uterine Anomalies and Reproductive History

Although some uterine anomalies can cause infertility, most patients with uterine anomalies are able to conceive without difficulty. There is no question that uterine anomalies can be associated with perfectly normal reproductive performance. Overall, however, the incidences of spontaneous abortion, premature birth, fetal loss, malpresentation, and cesarean section are clearly increased when a uterine anomaly is present. It is impossible to predict which patients with uterine anomalies will have these problems.

Etiology of Reproductive Failure

The etiology of reproductive failure in patients with uterine anomalies remains unclear. Mahgoub believes that the presence of a uterine septum can lead to spontaneous pregnancy loss because of diminished intrauterine space for fetal growth or because of implantation of the placenta on a poorly vascularized septum. Mizuno and associates have attached importance to the inadequacy of vascularization of the uterine septum. Associated cervical incompetence, luteal phase insufficiency, and distortion of the uterine milieu have all been implicated in the etiology of increased reproductive loss. However, it is as yet unexplained why some patients with a uterine anomaly have normal reproductive function, whereas others abort early in pregnancy. Interestingly, it has been reported that the chance for a liveborn child increases with each pregnancy loss. It is unknown whether this apparent "conditioning" of the uterus is due to better vascularization, better myometrial stretching and accommodation, or some other factor.

A medical history of three or more episodes of spontaneous abortion or premature labor merits evaluation of the uterine cavity to determine whether structural abnormalities of the uterus are present. An abnormality is found in approximately 10% of such cases. Among chronic early second-trimester aborters, the incidence may be higher. The etiology of spontaneous abortion is complex, and a complete workup should be done even when an anomalous uterus has been found. A careful history should include a detailed discussion of each previous pregnancy loss and inquiry into DES exposure or other drug or chemical toxicity, specific medical illnesses, and exposure to contagious diseases. A family history should emphasize reproductive failures among family members of both the patient and her partner. Specific medical diseases such as thyroid disease, diabetes mellitus, renal disease, and systemic lupus erythematosus should be ruled out. The possibility of infection by such agents as *Neisseria gonorrhoeae, Chlamydia, Mycoplasma, Toxoplasma,* and *Listeria* should be considered. Chromosome analyses should be done. Abnormalities in aborted tissue are found in more than 50% of spontaneous abortions, and abnormalities appear in up to one fourth of couples with a history of habitual abortion. Identifying such couples makes it possible to offer genetic counseling for subsequent pregnancies. Uterine leiomyomas, especially lower uterine segment and submucous leiomyomas, can cause spontaneous abortion. Basal body temperature charts, serum progesterone determinations, and endometrial biopsies timed in the luteal phase help determine the presence of luteal phase deficiency. The cervix should be studied for incompetence.

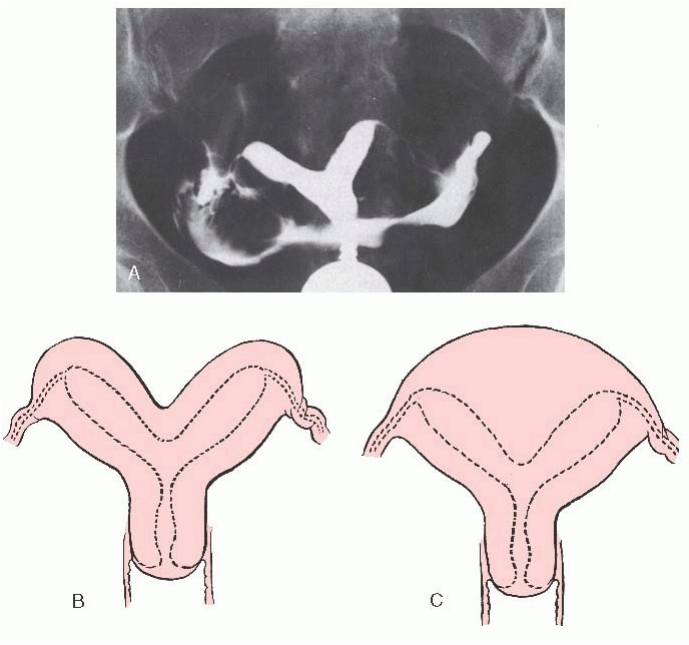


FIGURE 25.17 A: A hysterogram of a double uterus. A bicornuate uterus **(B)** and a septate uterus **(C)** are types of double uteri. Visualization of the fundus is required to determine the type of uterus.

Couples with multiple etiologies for reproductive loss should have all other problems corrected before metroplasty is considered. Indeed, correcting other factors first may correct the problem of reproductive loss without metroplasty. In 1977, Rock and Jones reported on seven patients who had anomalous uterine development and extrauterine factors in the etiology of their reproductive loss. These patients had already had 16 pregnancies, 5 (29%) of which resulted in a liveborn child. After therapy to correct the extrauterine factor, the success rate increased to 71%. Stoot and Mastboom reported an impressive increase in reproductive performance among uterine anomaly patients by simple improvement of abnormal carbohydrate metabolism.

Hysterographic Studies

Proper technique during the performance of hysterosalpingography to diagnose uterine anomalies is important. The hysterogram must be taken at right angles to the axis of the uterus for a true assessment of the deformity to be made. The study is best done under fluoroscopy. A septate uterus cannot be distinguished from a bicornuate uterus by hysterogram alone (**Fig. 25.17**). The external uterine configuration also cannot usually be determined by pelvic examination alone, but some idea of the configuration can be obtained by ultrasonography.

McDonough and Tho have suggested the use of double-contour pelvic pneumoperitoneum-hysterographic studies for precise identification of müllerian malformations. Of course, laparoscopy is even more certain. If the uterine corpus has not been previously visualized, the physician must be prepared to correct either anomaly (i.e., obstructed or unobstructed), depending on the findings at surgery.

Additional Testing

A complete investigation should also include an assessment of tubal patency and an MRI urogram or an intravenous pyelogram. A variety of upper urinary tract anomalies are seen, including absence of one kidney, horseshoe kidney, pelvic kidney, duplication of the collecting system, and ectopically located ureteral orifices. The lower urinary tract (bladder and urethra) is much less often anomalous.

The Double Uterus and Obstetric Outcome

The percentage of full-term pregnancies with various types of double uteri in an unselected series of women who have not been operated on is unknown. For all types combined, it is probably approximately 25%. In patients selected for operation, it probably increases from approximately 5% to 10% to approximately 80% to 90%. Because patients with uterine anomalies who have relatively normal obstetric histories cannot be identified, there is confusion in the literature about which anomalies are more often associated with obstetric difficulties and which are relatively benign in their effect. Special diagnostic procedures to detect uterine anomalies are not usually performed before reproductive performance is tested. A didelphic uterus is the exception. This anomaly can be diagnosed easily on routine pelvic examination by identification of two complete cervices and perhaps also a longitudinal vaginal septum. A study by Heinonen in Finland of 182 women with uterine anomalies indicated that pregnancies in the septate uterus had a better fetal survival rate (86%) than they did in the complete bicornuate uterus (50%) or in the unicornuate uterus (40%). These findings differ from prevailing opinions that the septate uterus is associated with the highest reproductive loss, as proposed by Jones and Jones. A 2011 report by Woefler and associates supports Jones and Jones's opinions by noting that women with a septate uterus had a significantly higher proportion of first-trimester loss than did women with a normal uterus.

In 1968, Capraro and colleagues reported on 85 patients with uterine anomalies seen between 1962 and 1966. One uterine anomaly was seen for every 645 admissions (0.145%). Metroplasty was considered necessary in only 14 (16%) of these 85 cases. According to Jones and Jones, only one third of patients with a double uterus have important reproductive problems. In most instances, the presence of a double uterus is not in itself an indication for metroplasty.

In 1980, Jewelewicz and coworkers estimated the spontaneous abortion rate to be 33.8% in women with a bicornuate uterus, 22.2% in those with a septate uterus, and 34.6% in those with a unicornuate uterus. More recently, Ludmir and associates reported that high-risk obstetric intervention did not significantly increase the fetal survival rate for uncorrected uterine anomalies. Capraro and associates found a preoperative fetal salvage rate of 33.3% for the septate uterus, 10% for the bicornuate uterus, and 0% for the didelphic uterus. Postoperatively, the fetal salvage rate was 100% for the bicornuate uterus, 80% for the septate uterus, and 66% for the didelphic uterus. The report gives the improved salvage figures, compared with several previous studies, after abdominal metroplasty.

Ravasia and colleagues described the incidence of uterine rupture in a cohort of women with müllerian duct anomalies who attempted vaginal birth after cesarean delivery (VBAC). Of the 1,813 patients who attempted VBAC between 1992 and 1997, only 25 patients with known müllerian duct anomalies attempted a trial of labor. This included 14 patients with a bicornuate uterus, five with a septate uterus, four with a unicornuate uterus, and two with uterine didelphys. Uterine rupture was diagnosed in two patients with müllerian anomalies. The authors proposed several mechanisms for the higher incidence of uterine rupture in this population: abnormal development of the lower uterine segment, previous scar similar to a vertical or classic incision, and the possibility of abnormal traction on the uterine scar during labor.

The Didelphic Uterus

A didelphic uterus with two hemicorpora is easily diagnosed because all patients have two hemicervices visible on speculum examination and most, if not all, have a longitudinal sagittal vaginal septum. In the series reported by Heinonen and associates, all 21 patients with a didelphic uterus had a vaginal septum. Conversely, a patient with a longitudinal vaginal septum usually has a didelphic uterus. The indication for uterine unification is related to the role of this anomaly as an etiologic factor in reproductive loss. Of all the uterine anomalies (except arcuate uterus), the didelphic uterus is associated with the best possibility of a successful pregnancy. However, there is still some increase in perinatal mortality, premature birth, breech presentation, and cesarean section for delivery. Heinonen and associates reported a fetal survival rate of 64% without metroplasty. Musich and Behrman stated that the didelphic uterus offers the best chance for a successful pregnancy (57%) and should not be considered an appropriate indication for metroplasty. However, W. S. Jones considered the didelphic uterus to give the worst obstetric outcome. In the opinion of the editors of this book, a unification operation for a didelphic uterus is not often indicated, and the results may be disappointing, especially when an attempt is made to unify the cervix. Not only is this procedure technically difficult in a complete didelphic anomaly, but it can also result in cervical incompetence or cervical stenosis.

The Septate Uterus

Most patients who are evaluated for repeated pregnancy loss and who are found to have a uterine anomaly have a septate uterus. A few have other anomalies, mostly the bicornuate uterus. Proctor and Haney's review of women with recurrent first-trimester pregnancy loss reinforces the role of the septate uterus in repeated pregnancy loss. Of 35 women reviewed with a divided uterine cavity on hysterosalpingogram, all women were found through diagnostic hysteroscopy and laparoscopy to have a septate uterus. In our experience, fetal survival rates are higher after septate uterus repair than after other repairs. In 1977, Rock and Jones reported on 43 patients with septate uteri selected for Jones metroplasty at the Johns Hopkins Hospital. Of these 43 patients, 95% became pregnant postoperatively, 73% carried to term, and 77% delivered a liveborn child. Similarly, hysteroscopic metroplasty for the septate uterus provides a substantial improvement in obstetric outcome. Data obtained from retrospective studies suggest that hysteroscopic metroplasty is associated with favorable outcomes, with a pregnancy rate of approximately 80% and a miscarriage rate of only approximately 15%. Recent prospective observational studies reported similar findings. Pabuccu and Gomel reported the reproductive outcome of 61 patients; however, the patients had unexplained infertility, and nearly 15% were also treated with cervical cerclage. Litta and colleagues also reported an 83.3%

term delivery rate in their population of women with a septate uterus who underwent hysteroscopic metroplasty. Patton and colleagues described 16 women with a complete uterine septum. The preoperative pregnancy loss rate was 81%. Eleven of the fourteen septa treated with a hysteroscopic approach were successfully removed. The remaining three unsuccessful hysteroscopic procedures and two additional patients were treated using the Tompkins metroplasty. Postoperatively, 9 women conceived after hysteroscopic surgery, and term live births occurred in 9 of 12 (75%) conceptions. The most controversial area remains the resection of the cervical portion of the septum. Parsanezhad and colleagues reported a multicenter, randomized controlled trial regarding the management of the cervical septum. Surgical issues, complications, and pregnancy outcomes were compared. Operating times, distending media deficits, and perioperative complications were all substantially better in the cervical septum resection group. The reproductive outcomes were similar; however, the cesarean section rate was higher in the group in which the cervical septum was spared. The histologic features of the septum in this abnormal uterus have been described. Dabirashrafi and colleagues noted less connective tissue in uterine septa. Poor decidualization and placentation were suggested as a cause.

Finally, the AFS class Va uterus (a double cervix and uterine cavity with a single fundus) can result from a rotation abnormality during the descent of the müllerian ducts. Among the reported cases of the septate uterus, the incidence of the complete septum involving the cervix varies from 4% to 29%. If the dextrorotating müllerian ducts overrotate, the senior author theorizes that the septum fails to absorb after fusion of the ducts (J. A. Rock, *personal observations,* 1991). In virtually every patient with a complete septate uterus, the left cervix is higher than the right. In one patient, one cervix has been noted above the other (**Fig. 25.18**). This rotation abnormality may be a factor associated with lack of absorption of the uterine septum in these patients.

Uterine Anomalies and Menstrual Difficulties

Dysmenorrhea and abnormal and heavy menstrual bleeding have been reported to occur more frequently with any form of double uterus and to be relieved after unification operations. Capraro and associates reported several cases in which dysmenorrhea was cured by metroplasty. Erwin Strassmann also believed that all cases of dysmenorrhea and menorrhagia associated with uterine anomalies were relieved by unification of the two uterine cavities. Generally, however, dysmenorrhea and menorrhagia are inappropriate indications for uterine unification, and the operation should not be performed solely for these reasons. Grynberg and colleagues reported on women who experienced infertility, pregnancy losses, dyspareunia, or dysmenorrhea. In their series, 22 patients underwent hysteroscopic resection of complete uterine septum and resection of longitudinal vaginal septum. The data suggested that hysteroscopic incision of the septum did not improve reproductive outcomes (miscarriage rate increased from 25% to 43%, cervical cerclage rate was not significantly changed). In their series, the advantage of surgery was limited because the miscarriage and the preterm delivery rates were not improved after metroplasty. This publication reminds reconstructive gynecologists that careful consideration of indications for surgery is imperative before surgery. Until more data are available, systematic surgical intervention for young women diagnosed with complete uterine septum without evidence of pregnancy loss should be discouraged.

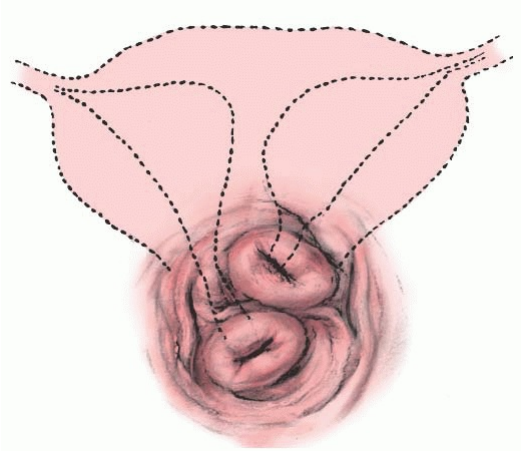


FIGURE 25.18 A double uterus with two cervices and a single fundus (class V). Note that the left cervix is positioned over the right cervix. This rotation abnormality may be a factor associated with a lack of absorption of the uterine septum.

Uterine Anomalies and Infertility

Opinions differ considerably in terms of whether infertility is a proper indication for metroplasty. Erwin Strassmann stated that primary infertility could be cured in 60% of patients with uterine anomaly if all other causes of infertility were excluded. Strassmann reported eight metroplasties for primary sterility that yielded nine pregnancies and seven liveborn children, although the number of patients who conceived was not given. Similar reports of small numbers of patients can be found throughout the literature. Heinonen and Pystynen indicated that uterine anomalies are rarely the reason for infertility. Nonuterine causes of infertility must be ruled out before metroplasty, as a last resort, is considered.

Certainly, a full infertility investigation to rule out other causes should be completed before the anomalous uterus is blamed. Even when no other cause for infertility is found, if the uterus is septate or bicornuate, then there may not be any proper indication for metroplasty. This question of when to perform metroplasty simply has not yet been answered. The decision is difficult and becomes even more difficult when the opportunity for metroplasty presents itself because a septate or bicornuate uterus requires laparotomy for some other reason, such as endometriosis or tubal occlusion.

Surgical Technique for Uterine Unification

Historically, the septate uterus has been unified with either the Jones or the Tompkins procedure. Clinical reports by Chervenak and Neuwirth; by Daly, Walters, and colleagues; by DeCherney and associates; and Israel and March have favorably compared hysteroscopic (resectoscopic incision of a uterine septum) with the more traditional transabdominal approach. Term pregnancy rates after these procedures have approached 80% to 85%. Several attempts may be necessary to incise a wide septum, although the septum usually can be incised completely at the first operation.

Transcervical Lysis of the Uterine Septum

Abdominal metroplasty for transfundal incision or for excision of the septum associated with the septate uterus generally has been abandoned. With hysteroscopic scissors, the procedure can be tedious, especially with a large, broad septum. Although

the hysteroscope and scissors are still used for cutting the septum, the resectoscope has been found to be comparable. The optics are excellent, and the septum can be electrosurgically incised with little difficulty. Laser-assisted procedures have also been described.

Before transcervical lysis of a uterine septum, a gonadotropin-releasing hormone agonist may be given for 2 months to reduce the amount of endometrium that can obscure the surgeon's view during the procedure. Many authors do not consider routine preoperative preparation of the endometrium essential and may only use medications in procedures involving exceptionally wide septa or complete septa that involve the lower one third of the uterine cavity or the cervical canal. If medical preparation is not used, surgical intervention should be scheduled during the early proliferative phase of the cycle to avoid bleeding and impaired visualization from a vascular endometrium associated with the secretory phase. Transcervical lysis is usually performed in conjunction with laparoscopy under general endotracheal anesthesia. The uterine cavity is distended with dextran 70 (Hyskon) by way of the resectoscope, which is inserted into the cervix. The septum is then electrosurgically incised by advancing the cutting loupe, using the trigger mechanism of the resectoscope. The uterine septum is incised until the tubal ostia are visualized and there is no appreciable evidence of the septum. The procedure is performed under simultaneous laparoscopy to limit the risks of uterine perforation. The laparoscopic light can be turned off so that the light from the hysteroscope can be clearly visualized through the fundus. Most patients can be discharged within 4 hours of the procedure. There is no role for placement of a postoperative intrauterine device. The benefit of routine procedure-related antibiotic therapy has not been well supported with evidence; however, it is recommended to administer antibiotics before the procedure and to continue for 5 days after surgery to limit the risks of infection. If excessive bleeding occurs after the procedure, a Foley catheter should be placed in the uterine cavity for tamponade and removed in 4 to 6 hours. Hormonal therapy is the most commonly used postoperative treatment regimen. The aim of the treatment is the promotion of rapid epithelialization. Dabirashrafi and colleagues reported that estrogen therapy did not appear to demonstrate a benefit. Further evidence is necessary before dismissing the current trend of postoperative estrogen therapy.

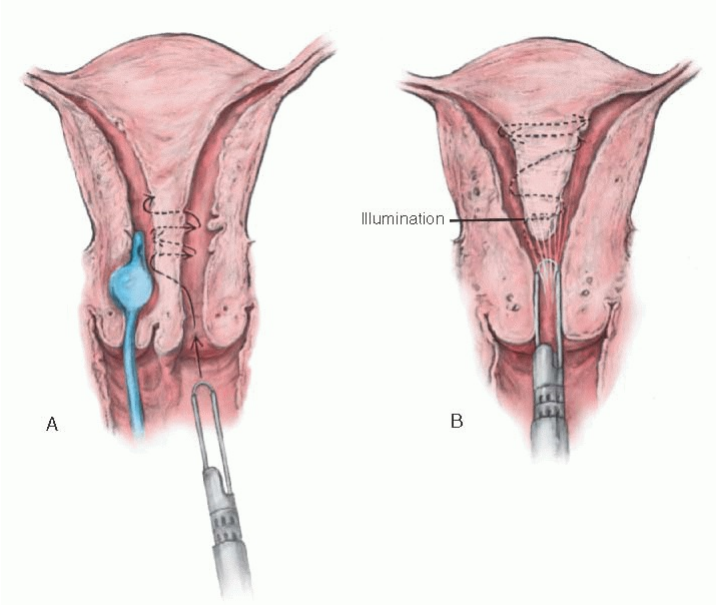


FIGURE 25.19 Resectoscopic metroplasty. **A:** A Foley catheter is placed in one cavity of a complete septate uterus (American Fertility Society class Va uterus). The resectoscope is inserted in the opposite cavity, and the septum is incised until the Foley is visualized. The septum can be easily incised with the resectoscope until both internal os are visible. **B:** A septate uterus with a single cervix. The septum can be incised with the straight loupe of the resectoscope.

Transcervical lysis also can be performed to repair a complete septate uterus (i.e., a single fundus with two cavities and two cervices). In this instance, a no. 8 Foley catheter is inserted into one cervix and indigo carmine is injected into the cavity. The other cavity is distended with dextran 70 (Hyskon) by way of the resectoscope. The septum is electrosurgically incised at a point above the internal cervical os until the Foley catheter is visualized. The septum is then incised in a superior direction until the tubal ostium is visualized and there is no appreciable septum (Fig. 25.19).

After transcervical lysis of a uterine septum, a 2-month delay before attempting pregnancy is suggested to allow complete resorption of the septum. Delivery may be vaginal. The Jones procedure is used to repair a septate uterus when a particularly broad septum cannot be easily incised with the resectoscope. The Strassmann procedure is used for unification of a bicornuate uterus. The safety and efficacy of hysteroscopic resection of the uterine septum in patients with a class Va septate uterus has been demonstrated by the senior author. Historically, case reports, such as that of Hundley and colleagues, were the only source of information about this interesting variant; however, one of the largest populations of patients with a complete septum was reported in 1999 by Rock, Roberts, and Hesla. The patients underwent hysteroscopic metroplasty with preservation of the cervical portion of the septum. With the exception of one case of pulmonary edema, no significant intraoperative or postoperative complications were reported. Postoperative hysteroscopy revealed only minor fundal septal remnants without clinical significance.

The Modified Jones Metroplasty

In the modified Jones unification operation (Fig. 25.20), the abdomen is generally opened through a transverse incision. If only the unification operation is planned, then a Pfannenstiel incision is permissible. The pelvic viscera are inspected. The septate uterus may demonstrate a median raphe across the fundus, but it is surprising how often the corpus looks normal. To facilitate manipulation, a traction suture of heavy silk is placed through the top of the septum. This suture is removed from the site when the septum is excised.

No attempt is made to stain the uterine cavity with methylene blue. Normal unstained endometrial tissue can be easily differentiated from the myometrium.

There are essentially two methods to control bleeding during this procedure. In the first, a tourniquet is applied at the junction of the lower uterine segment and cervix by inserting a 0.5-inch Penrose drain through an avascular space in the broad ligaments just lateral to the uterine vessels on each side. The tourniquet is placed around the lower uterine segment and is tied anterior to the uterus. Because the uterine corpus receives a significant blood supply through the ovarian arteries, tourniquets should also be tied around the infundibulopelvic ligaments on each side, using the same hole in the broad ligament. All tourniquets must be tied tightly enough to occlude both the arterial supply to and the venous drainage from the uterus. If only the venous drainage is occluded, then the corpus becomes engorged and congested, and bleeding is increased. If the arterial supply is occluded, then the uterus blanches, and the bleeding is minimal. A sterile Doptone can be used to establish disappearance of uterine artery pulsations. Hypotensive anesthetic techniques used in conjunction with the tourniquets allow a uterine unification operation to be accomplished with negligible blood loss.

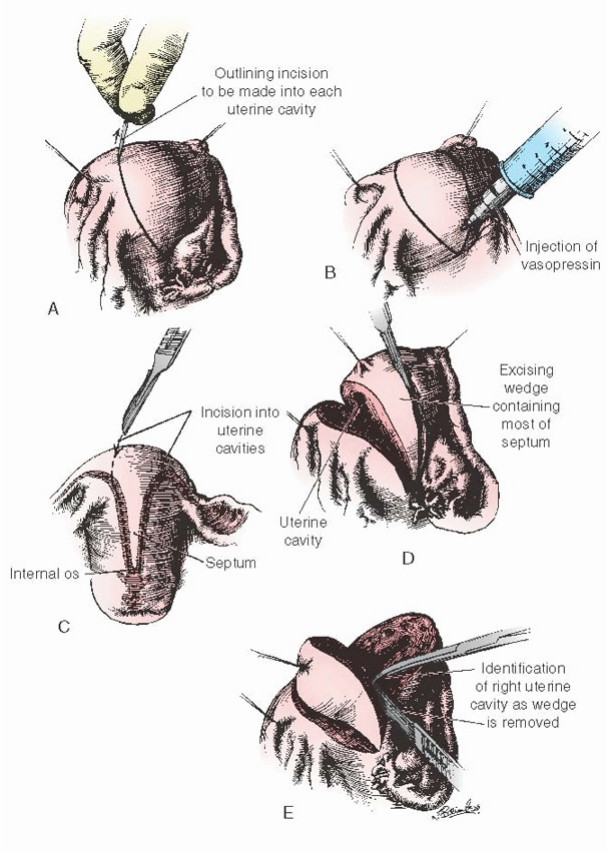


FIGURE 25.20 The modified Jones metroplasty. See the text for a full description of the various steps in the operative repair of a septate uterus by excision of a wedge.

The alternative method for hemostasis uses up to 20 units of vasopressin that is diluted in 20 mL of saline and injected into the anterior and posterior walls of the uterus before the incision is made.

The uterine septum should be surgically excised as a wedge (**Fig. 25.20D**). The incisions begin at the fundus of the uterus. The approach to the endometrial cavity should be handled

carefully so that it is not transected (**Fig. 25.20E**). The original incisions at the top of the fundus are usually within 1 cm—and sometimes even less—of the insertion of the fallopian tubes. If the incision is directed toward the apex of the wedge, however, there seems to be little danger of transecting the tube across its interstitial transit in the myometrium.

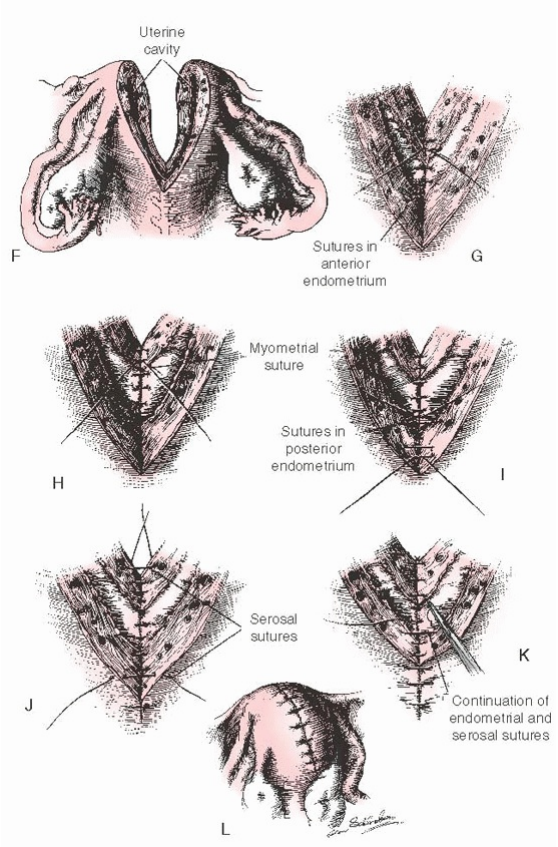


FIGURE 25.20 (*Continued*)

After the wedge has been removed, the uterus is closed in three layers with interrupted stitches; 2-0 nonreactive suture on an atraumatic tapered needle is convenient. Two sizes of

needles are needed: a half-inch needle for the inner and intermediate layers and a large needle (three fourths half round) for the outer muscular layer. The inner layer of stitches must include approximately one third of the thickness of the myometrium, because the endometrium alone is too delicate to hold a suture and will be cut through. The inner sutures should be placed through the endometrium and the myometrium in such a way that the knot is tied within the endometrial cavity (**Fig. 25.20G, H**). While the suture is being tied, the two lateral halves of the uterus should be pressed together both manually and with the guy sutures to relieve tension on the suture line and to reduce the possibility of cutting through. These sutures are placed alternately, first anterior and then posterior. After the first few stitches are placed and before the first layer is completed, the second layer can be started to reduce tension.

As the operation proceeds, the third layer of stitches is begun in the serosa both anteriorly and posteriorly (**Fig. 25.20I-K**). Finer, nonreactive suture material can be used to approximate the serosal edges of the uterus more precisely to prevent adhesion formation to the suture line (**Fig. 25.20K, L**). By the conclusion of the operation, the uterus appears near normal in configuration. The striking feature is usually the proximity of the insertions of the fallopian tubes. Special care must be exercised not to obstruct the interstitial portions of the fallopian tubes while placing the fundal myometrial and serosal sutures.

The final size of the uterine cavity seems to be relatively unimportant to reproductive capability; uterine symmetry appears to be a more important factor. Often, the constructed cavity is quite small compared with the normal uterus.

Whether the surgeon removes the septum with the Jones procedure or lyses the septum transcervically, postoperative hysterogram films often show small dog-ears that are leftover tags from the original bifid condition of the uterus. Such dog-ears do not seem to interfere with function, although a postoperative roentgenogram cannot be considered normal in the sense that it does not have the appearance of a normal endometrial cavity after such an operation. If a double cervix is present, the physician should not attempt to unify the cervix because an incompetent cervical os will result. To allow the uterine incision the best possible opportunity to heal, a delay of 4 to 6 months in attempting pregnancy is advised after abdominal metroplasty.

The Jones Metroplasty versus the Tompkins Procedure

The technique of modified Jones metroplasty is a compromise between the classic Jones metroplasty and the Tompkins metroplasty. In the Jones operation, the entire septum is removed. In the Tompkins operation, a single median incision divides the uterine corpus and septum in half. The incision is carried inferiorly until the endometrial cavity is reached. Each lateral septal half is then incised to within 1 cm of the tubes. No septal tissue is removed. The myometrium is reapproximated, taking care not to place sutures too close to the interstitial portion of the tubes. Proponents of the Tompkins technique suggest that it is simpler than the classic Jones procedure, that it conserves all myometrial tissue and leaves the uterotubal junction in a more normal and lateral position, and that it provides better results than the Jones metroplasty. Good results with the Tompkins technique have been reported by McShane and colleagues.

The Wedge Metroplasty versus Transcervical Lysis

There are obvious advantages to a transcervical incision of a uterine septum for patients with a septate uterus. Morbidity is decreased after the procedure, and delivery can be vaginal. Term pregnancy rates are comparable with those after abdominal metroplasty for repeated pregnancy wastage.

Most of the septa associated with a septate uterus can be cut through the cervix by way of the hysteroscope or the resectoscope. Nevertheless, cases of broad uterine septum can benefit from the wedge metroplasty, and reconstructive surgeons should be knowledgeable in its performance.

The Strassmann Metroplasty

The Strassmann procedure is not easily adapted to the septate uterus, but it is the procedure of choice for unification of the two endometrial cavities of an externally divided uterus, both bicornuate and didelphic (**Fig. 25.21**). A bicornuate uterus cannot be repaired through transcervical lysis because perforation will result. When there has been failure of fusion of the two müllerian ducts, inspection of the pelvic cavity often reveals a broad peritoneal band that lies in the middle between the two lateral hemicorpora. This rectovesical ligament is attached anteriorly to the bladder, folds over and is attached between the uterine cornua, continues posteriorly in the cul-de-sac, and ends with its attachment to the anterior wall of the sigmoid and rectum. It is not invariably present, but when it is, its potential significance in the etiology of the anomaly, possibly by preventing the two müllerian ducts from joining, must be considered. This rectovesical ligament must be removed before a unification procedure can be performed (**Fig. 25.21A**).

For hemostasis, tourniquets are used in a manner similar to that described for the modified Jones procedure. The two uterine cornua are incised on their median sides in their longitudinal axes, deeply enough to expose the uterine cavities (**Fig. 25.21B**). Superiorly, the incision must not be too close to the interstitial portion of the fallopian tubes. Inferiorly, the incision is carried far enough to join the two sides into a single endocervical canal. If it appears that a deeper incision will compromise the competence of the cervix, then a double cervical canal can be left. If the cervix is already duplex, then it should not be joined. As the incision in the myometrium releases the internal stresses in the walls of the hemicorpora, each one everts and is perfectly positioned for apposition, almost as if the original intention in embryologic development is finally to be realized. The suture technique for joining the two sides (**Fig. 25.21C-E**) is exactly the same as for the modified Jones procedure. The suture line in the uterine corpus should be observed for several minutes to determine the adequacy of hemostasis. Occasionally, it is necessary to place one or two extra sutures to control bleeding.

A uterine suspension can be performed as necessary. However, in the event of pregnancy, the shortened round ligaments can produce symptoms from an enlarging uterus. Presacral neurectomy in association with uterine unification should be considered only in patients with severe midline dysmenorrhea.

The cervix should be dilated to ensure proper drainage from the uterine cavity. This can be accomplished transvaginally after the abdominal procedure or from above by inserting a dilator through the cervical canal into the vagina to be removed later.

The operative technique should always be consistent with the goal of maintaining or enhancing fertility and possibly achieving a successful pregnancy. Tissue surfaces should be kept moist throughout the procedure, and instruments should be selected and used in such a way that tissue damage is minimized. Abdominal packs should be placed in plastic bags to avoid adhesions, or no-lint laparotomy pads can be used. Talc should be carefully washed from gloves, and meticulous aseptic technique should be used. The appendix should not be removed. Lactated Ringer solution containing heparin and corticosteroid can be used for peritoneal lavage throughout the procedure.

Cervical Incompetence Associated with a Double Uterus

When a patient with an anomalous uterus, with or without unification, becomes pregnant, she must be watched closely for evidence of cervical incompetence, especially if a history of previous reproductive loss suggests cervical incompetence. Heinonen and associates improved fetal survival rate from 57% to 92% by cervical cerclage. Cerclage was used mostly in patients with a partial bicornuate uterus. In these patients, the fetal salvage rate was improved from 53% before cerclage to 100% afterward. Prematurity also was decreased, from 53% to 3%. The authors stress that cervical incompetence, not the uterine anomaly, is the proper indication for cerclage in these patients. However, the frequency with which these problems are found together suggests the importance of doing a careful evaluation for both problems. Some reproductive losses from a uterine anomaly might be prevented by cerclage of an incompetent cervix during metroplasty. However, routine cerclage at the time of metroplasty is not recommended.

Attempts to unify a double cervix or a septate cervix also 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 cesarean section if it appears that the cervix will cause dystocia.

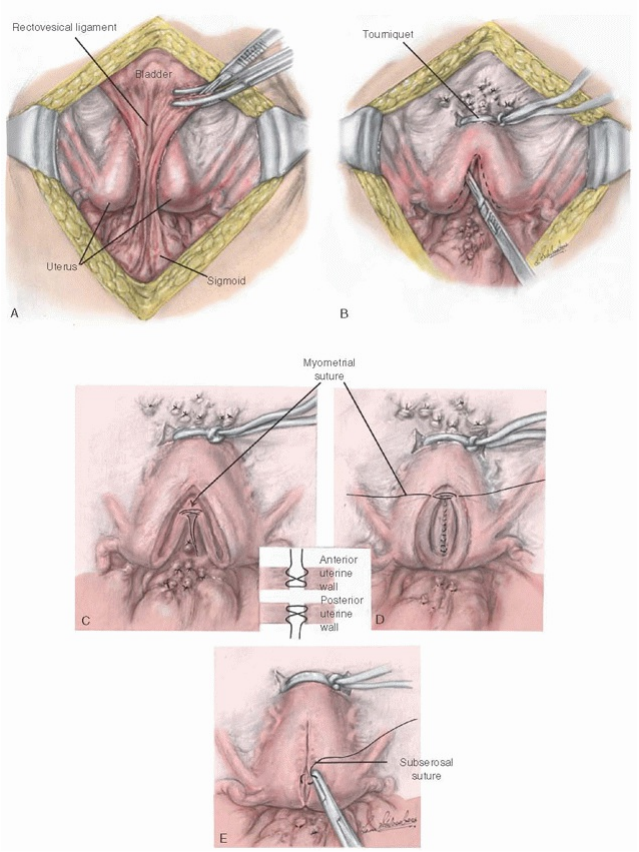


FIGURE 25.21 The Strassmann metroplasty with modification. **A:** If a rectovesical ligament is found, it should be removed. **B:** An incision is made on the medial side of each hemicorpus and carried deep enough to enter the uterine cavity. The edges of the myometrium will evert to face the opposite side. **C and D:** The myometrium is approximated by the use of interrupted vertical figure-of-eight 3-0 polyglycolic acid sutures. One should avoid placing sutures too close to the interstitial portion of the fallopian tubes. **E:** A continuous 5-0 polyglycolic acid subserosal suture is used as a final layer. Tourniquets are removed, and defects in the broad ligament are closed.

Mode of Delivery after Metroplasty

The scar formed in the myometrium after unification is as strong as, if not stronger than, the scar formed after cesarean section. The biologic conditions under which healing occurs are entirely different in these two situations. Endomyometritis is a common complication after cesarean section but is not a complication of uterine unification. Of 71 known pregnancies in Strassmann's collected series reported in 1952, 61 were delivered vaginally. There were no cases of uterine rupture during pregnancy or delivery. Lolis and colleagues reported the reproductive outcome of 22 women who underwent the Strassmann metroplasty for a bicornuate uterus; 88% achieved pregnancies that ended with the delivery of a viable infant. All were delivered by cesarean section without evidence of scar rupture. Despite evidence that the uterine scar heals securely after unification operations, our policy is to recommend delivery by elective cesarean section in all patients who have undergone abdominal metroplasty. Patients can deliver vaginally after a metroplasty by hysteroscope or resectoscope.

Diethylstilbestrol-Related Uterine Anomalies

Exposure of the female fetus to DES can cause significant anomalous development of the uterus, as reported by Kaufman and associates and by Haney and colleagues. The T-shaped uterus is the variant most commonly seen. It is associated with an increased rate of spontaneous abortions, preterm deliveries, and ectopic pregnancies.

Nagel and Malo determined the feasibility of correcting the uterine malformations seen in DES-exposed women by incising constriction rings and septa. Their goal was to incise the irregular uterine walls until the cavity assumed a smooth, straight line from the lower uterine cavity to the uterine tubal ostium. Their results suggested that metroplasty can decrease pregnancy loss but does not enhance fertility. The editors of this book suggest that the rare patient can benefit from a uterine reconstructive procedure, but that most will not. Surgeons may never develop a large series to document efficacy of surgical outcomes because patients with this anomaly are close to aging out of the natural reproductive years (last dispensed in 1971 in the United States). DES-exposed patients must be monitored closely for evidence of dilatation and effacement of the cervix early in pregnancy. Cervical cerclage may be indicated in some patients.

Unicornuate Uterus

A unicornuate uterus can be present alone or with a rudimentary horn or bulb on the opposite side. In a series reported by Heinonen and associates, 11 of 13 patients with a unicornuate uterus had a rudimentary horn, and two did not. The rudimentary anlagen (uterine muscle bundle or bulb) can communicate directly with the unicornuate uterus. In some instances, there is no cavity within the anlagen, or there is no rudimentary horn. Most rudimentary horns are noncommunicating (90% according to O'Leary and O'Leary). The two sides may be connected by a fibromuscular band, or there may be no connection and no communication between the two uterine cavities. Fedele and associates have found sonography useful in determining the presence of not only a rudimentary horn but also a cavity within.

Associated Anomalies

Urinary tract anomalies are often associated with a unicornuate uterus. On the side opposite the unicornuate uterus, there may be a horseshoe or a pelvic kidney, or the kidney may be hypoplastic or absent. This is especially true if there is associated müllerian duct obstruction. When all müllerian duct derivatives and the kidney are absent on one side, this implies failure of development of the entire urogenital ridge, including the genital ridge where the ovary forms. In addition, the ovary may be malpositioned (Fig. 25.22). Rock, Parmley, and associates reported a unilateral ovary located above the pelvic brim in four cases of uterine anomalies. The orifice of the müllerian duct develops at approximately the level of the fourth thoracic vertebra (T4) in the embryo. The tip subsequently migrates along the course of the müllerian duct into the pelvis. The orifice of the duct or the fimbriated end of the tube comes to lie in the pelvis as a result of differential growth of the fetus. The subsequent differential growth is retarded so that the portion of the urogenital ridge that gives rise to both the gonad and tube does not displace into the pelvis. Malpositions of the ovary and tube are the result.

Reproductive Performance

According to Heinonen and associates, the unicornuate uterus carries the poorest fetal survival rate (40%) of all uterine anomalies. In 1957, Jones reported similar findings. The

abnormal shape, the insufficient muscular mass of the uterus, and the reduced uterine volume and inability to expand may explain the poor obstetric outcome.

Moutos and colleagues compared the reproductive performance of the unicornuate uterus with that of the didelphic uterus. Twenty of the 29 women with a unicornuate uterus produced a total of 40 pregnancies, whereas 13 women with a didelphic uterus produced a total of 28 pregnancies. The percentages of pregnancies resulting in preterm delivery, term delivery, and living children were similar in both groups. The authors concluded that reproductive performance of the unicornuate uterus was not different from that of the didelphic uterus, that it is uncommon for either malformation to be a primary cause of infertility, and that there is insufficient information to support recommendation of placement of a cervical cerclage in the absence of cervical incompetence. Reichman, Laufer, and Robinson reviewed 20 published reviews on patients with a unicornuate uterus. They examined 290 total women who were reported in the literature; 175 patients conceived with 468 pregnancies reported. They reported 2.7% ectopic pregnancy, 24.3% first-trimester loss, 9.7% second-trimester abortion, 20.1% preterm delivery, and 10.5% intrauterine fetal demise. They reported a 49.9% livebirth rate. The authors also suggested that the current data available about the benefit of cervical cerclage (nonrandomized and lacking control subjects) seem inadequate to support a role for cervical incompetence for recurrent losses in patients with a unicornuate uterus.

Because most cases of unicornuate uterus have a noncommunicating rudimentary uterine horn on the opposite side, there is danger of pregnancy in the rudimentary horn from transperitoneal migration of sperm or ovum from the opposite side. According to Holden and Hart, approximately 350 cases of pregnancy in a rudimentary horn have been reported since the original case report by Mauriceau in 1669. O'Leary and O'Leary found the corpus luteum on the side contralateral to the rudimentary horn containing a pregnancy in 8% of cases. Signs and symptoms of an ectopic pregnancy develop with eventual rupture of the horn if the pregnancy is not detected early. Rupture through the wall of the vascular rudimentary horn is associated with sudden and severe intraperitoneal hemorrhage and shock. Death can occur in a few minutes. It is surprising that the current mortality rate has decreased to 5%.

Very little, if anything, can be done to improve the reproductive performance of patients with a unicornuate uterus. The physician should observe closely for signs and symptoms of preterm labor. Cervical incompetence is likely less common than previously suggested, yet if present, cerclage should be

considered as indicated. Andrews and Jones have suggested that removal of the rudimentary uterine horn may improve the chances of a successful pregnancy, eliminating the risk of ectopic pregnancy in the rudimentary horn as well as the risk of possible retrograde menstruation and the development of endometriosis. Typically, the procedure is a straightforward laparoscopic procedure with the potential for substantial benefit. Cases of asymmetric development of the unicornuate uterus with an opposing rudimentary uterine horn are not amenable to unification.

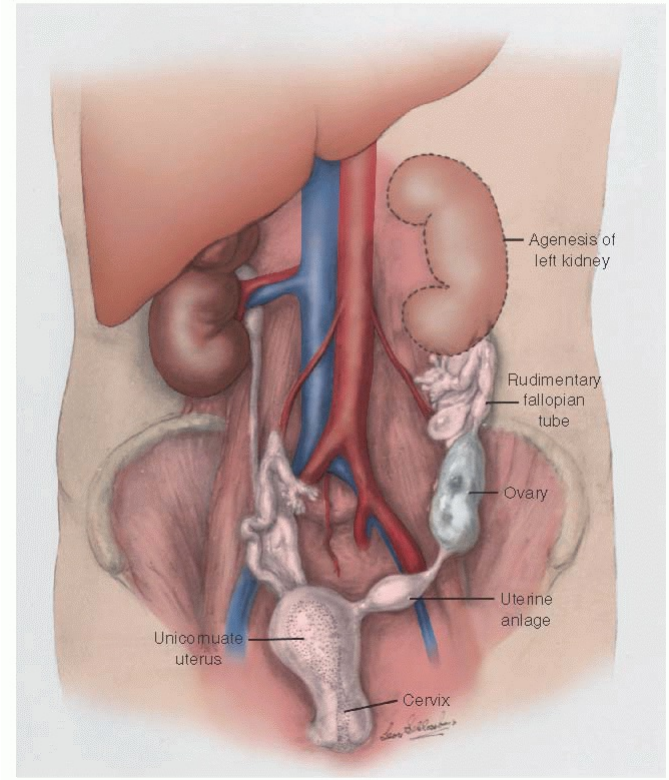


FIGURE 25.22 A unicornuate uterus associated with ovarian malposition on the left. Note that the ovary and the tube are slightly above the pelvic brim. In this instance, the ovary measured 6 inches long.

Longitudinal Vaginal Septum

Failure of fusion of the lower müllerian ducts that form the vagina can result in a vagina with a longitudinal septum. The septum can be partial or complete. Young patients have difficulty using tampons. In cases of didelphic uterus with a longitudinal vaginal septum, one uterine hemicorpus is usually better developed than the other. If intercourse consistently occurs on the vaginal side connected to the uterine hemicorpus that is less well developed, then infertility or repeated pregnancy loss could result. For these reasons, the septum should be removed (when the patient is not pregnant) unless there is a contraindication. This can usually be accomplished easily with reasonable precautions against injury to the urethra, bladder, and rectum.

Haddad and colleagues reported their experience over a 24-year period with management of the longitudinal vaginal septum. The retrospective review of 202 patient charts described a complete septum (extending from cervix to introitus) in 45.6% of patients, high partial in 36.1%, and a medium or low partial, involving only the distal vagina, in 18.3%. Uterine malformations were noted in 87.8% of cases. The frequency of uterine malformations was 99.4% in cases of complete or partial high septum and 30.3% in cases of partial medium or low septum. The most common malformation was class Va complete septate uterus in 59.5% of malformations, followed by class III uterus didelphys (24.3%), and class Vb partial septate uterus (15%). Section or resection was performed in 201 cases. Bladder injury in one patient was the only reported complication. As highlighted by the high prevalence of associated uterine malformations in this review, management should always include an assessment of uterine anatomy.

Asymmetric Obstruction of the Uterus or Vagina

Unicornuate Uterus and Noncommunicating Uterine Anlagen Containing Functional Endometrium

If one müllerian duct develops normally while the opposite müllerian duct fails to develop or develops incompletely, then a relatively normal unicornuate uterus is found on one side

and the cervix, musculature, uterine cavity, endometrium, fallopian tube, blood supply, and ligamentous attachments are absent or hypoplastic to a varying degree on the other side.

Obstruction to menstruation can also occur to varying degrees on the improperly developed side. For example, if a rudimentary uterine horn does not communicate externally but does have an endometrium-lined uterine cavity, then clear symptoms of obstructed menstruation may begin soon after menarche, and severe dysmenorrhea will be present. Cryptomenorrhea can be overlooked as the diagnosis because there is cyclic menstruation from the opposite side. It is important to make the diagnosis as soon as possible, because if the lumen of the tube communicates with the endometrial cavity of the rudimentary uterus, then retrograde menstruation and pelvic endometriosis will develop, and reproductive potential can be damaged. During the operation illustrated in **Figure 25.23,** which was performed to remove an obstructed rudimentary uterine horn, the fallopian tube was obstructed, and retrograde menstruation was impossible. Occasionally, the fallopian tube connected

to the rudimentary uterine horn may not be patent because of incomplete development. Multiple case reports regarding the laparoscopic resection of obstructed uterine anlagen have supported the use of multiple techniques (stapling, bipolar or monopolar cautery, and the harmonic scalpel). Fedele and colleagues reported a series of 10 patients who have done well; however, the follow-up was only reported out to 6 months postprocedure. The authors strongly recommend the removal of the associated fallopian tube to minimize the risk of an ectopic pregnancy.

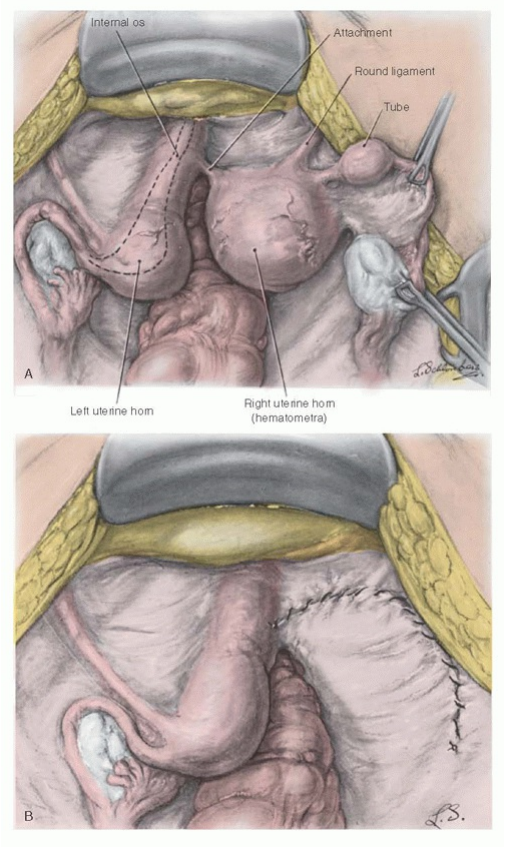


FIGURE 25.23 A: A noncommunicating rudimentary horn with functional endometrium that contains menstrual blood under pressure. Note the congenital abnormality of the fallopian tube, which prevented retrograde menstruation. **B:** The same patient after excision of the rudimentary horn.

Unilateral Obstruction of a Cavity of a Double Uterus

Another example of a rare obstructed lateral fusion problem is the complete septum between two uterine cavities illustrated in **Figure 25.24.** One cavity communicated with a cervix, and the other did not. This could represent an example of unilateral failure of cervical development. The patient reported incapacitating dysmenorrhea that appeared shortly after the menarche and lasted 5 days. A tense, cystic mass was palpable in the right half of the pelvis. The operation, described originally by Jones in the second edition of this book, consisted of making an incision through the anterior wall of the cystic right portion of the uterus. It was found to contain old menstrual blood. The entire septum was excised, and the uterus was reconstructed by anastomosis of the two cavities. A continuous lockstitch was reinforced by interrupted myometrial sutures, and the plastic reconstruction of the uterus was completed by a third layer of interrupted sutures uniting myometrium and serosa. Steinkampf and colleagues reported a similar case in a 17-yearold girl with progressive pelvic pain. The authors described an accessory noncommunicating uterine cavity, which they treated by excision at laparotomy.

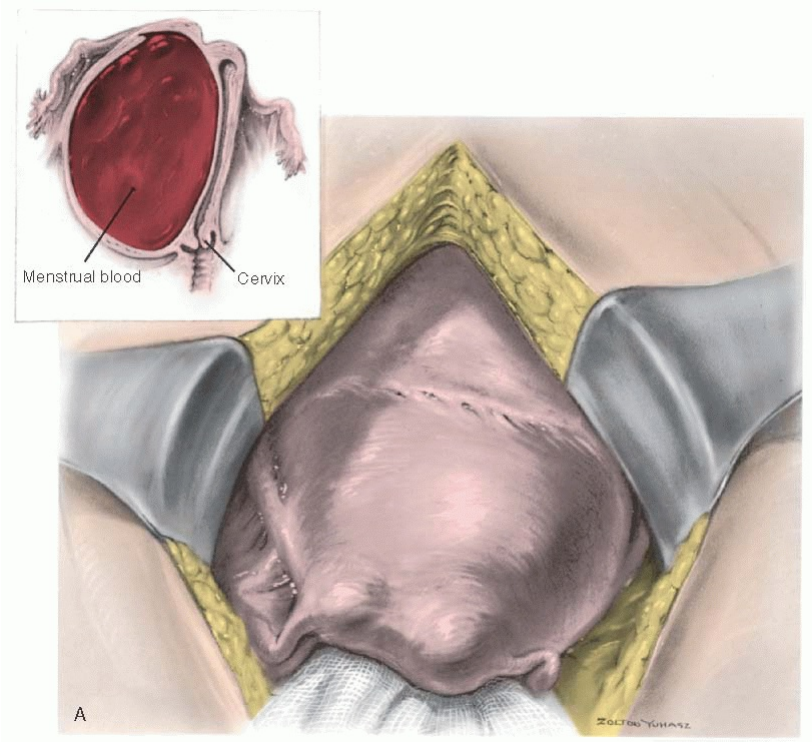
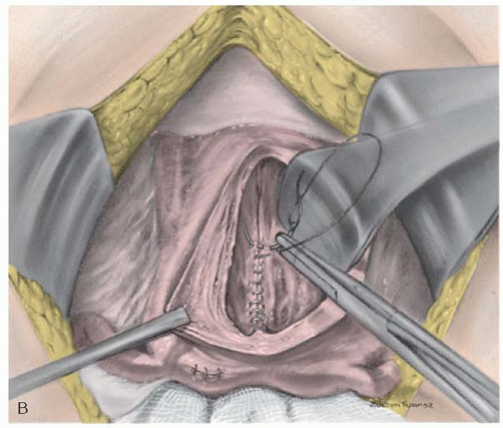


FIGURE 25.24 A: A double uterus seen at operation. Hematometra in the right uterine cavity (**inset**), which does not communicate with the other cavity or the cervical canal.

Sanders and colleagues described several cases in which the role of interventional radiology was crucial in the management of obstructive anomalies. The report described the drainage of a noncommunicating right uterine cavity distended with blood in a unicornuate uterus in a 14-year-old patient. Adequate access was established by using ultrasound-guided needle aspiration followed by a hysteroscopic excision. The assistance of interventional radiologic procedures, including percutaneous drainage and dilatation of small maldeveloped areas, may allow access to areas otherwise inaccessible by conventional mechanisms and assist in preserving reproductive function.

Double Uterus with Obstructed Hemivagina and Ipsilateral Renal Agenesis

The unique clinical syndrome consisting of a double uterus, obstruction of the vagina (unilateral, partial, or complete), and ipsilateral renal agenesis is rare. The renal agenesis (mesonephric involution) on the side of the obstructed vagina associated with a double uterus and double cervix is suggestive of an embryologic arrest at 8 weeks of pregnancy that simultaneously affects the müllerian and metanephric ducts. The exact cause is unknown.



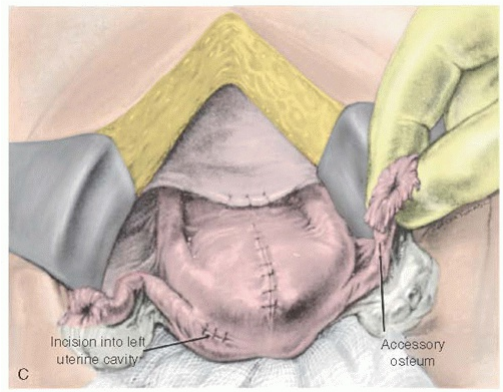


FIGURE 25.24 (*Continued*) **B:** The septum of the double uterus has been excised, and anastomosis is performed to unite the two cavities. **C:** Anastomosis is completed. The small incision in the left uterine cavity was made before the septum was removed for the purpose of orientation.

Diagnostic Groups

Clinical symptoms vary depending on the uterovaginal relations in individual cases, but the syndrome can be described generally in three groups. Group 1 patients have complete unilateral vaginal obstruction without uterine communication, resulting in a paravaginal mass and symptoms of severe dysmenorrhea and lower abdominal pain. Menses are regular. Group 2 patients have an incomplete unilateral vaginal obstruction without uterine communication. The presenting symptoms are lower abdominal pain, severe dysmenorrhea, excessive foul mucopurulent discharge, and, in some instances, intermenstrual bleeding. Group 3 patients have complete vaginal obstruction with a laterally communicating double uterus. They have a paravaginal mass, lower abdominal pain, and dysmenorrhea. Menses are regular. A 10-year review of patients with this anomaly was published by Phupong and colleagues. Most patients presented with dysmenorrhea (73%) or a pelvic or paravaginal mass (71%). The right uterus and vagina were affected in 63.5% of patients.

Because menses in patients with this syndrome are rarely irregular, the possibility of this syndrome as a diagnosis can easily be overlooked. A careful pelvic examination is necessary to make the correct diagnosis. Magnetic resonance imaging can identify the obstructed vagina, double uterus, and absence of a kidney on the side of the obstruction (Fig. 25.25), but it may not be helpful if there is incomplete vaginal obstruction or a uterine communication. With the onset of more universal prenatal imaging, the diagnosis of unilateral renal agenesis in a female fetus should prompt the obstetric team to interact with pediatric providers about the possible association with müllerian anomalies, particularly obstructing hemivagina with double uterus. The obstructed vagina may even prolapse through the introitus in newborns with mucocolpos of the obstructed vagina. A postdelivery pelvic ultrasound, combined with renal/bladder

sonogram, can provide data on the uterovaginal anatomy as well as rule out hydroureter and hydronephrosis.

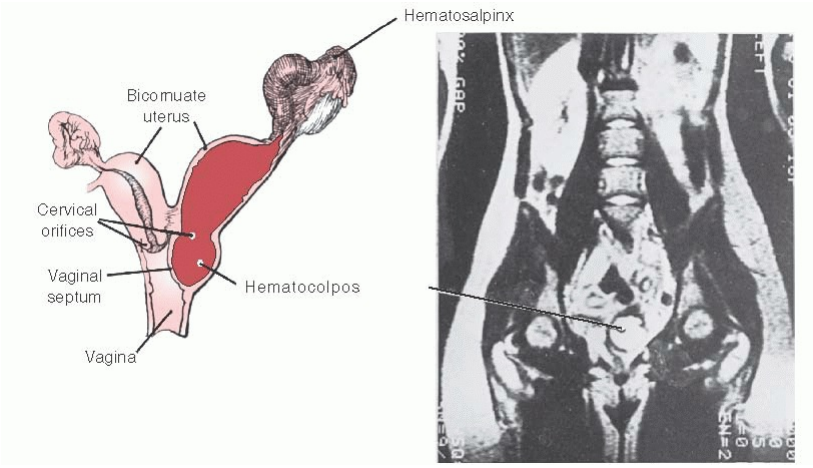


FIGURE 25.25 A double uterus with unilateral complete vaginal obstruction and ipsilateral renal agenesis. Magnetic resonance imaging reveals the left hematocolpos, both uteri, and absence of the left kidney on the side of the vaginal obstruction.

Complete unilateral vaginal obstruction (Group 1) can go unrecognized for a number of years after the onset of menses unless the obstructed vagina is significantly smaller causing hematometra to develop more quickly. The vagina is quite distensible and can accommodate a large amount of accumulated blood in the obstructed side. There is sufficient absorption of menstrual blood between periods so that each subsequent flow can add to the increments of accumulated blood without pain. Nevertheless, once retrograde menstruation occurs, endometriosis invariably is the result.

Surgical Treatment

Careful excision of the vaginal septum is the treatment of choice for a unilateral vaginal obstruction. Prophylactic antibiotics should be administered before surgery. After opening the vaginal pouch, the surgeon should use suction and lavage to remove the pooled blood and mucus. Phupong and colleagues' review also confirms the successful use of this primary therapy in 84.3% of patients. Haddad and colleagues reported a similar experience in a report describing patient management over a 27-year period. Excision of the vaginal septum was successful in 88% of patients, with complete excision in one procedure in 92% of those patients. In cases of pyocolpos or hematocolpos, distention and stretching of the septal tissue may increase the risk of inadequate resection and possible postoperative stenosis; the authors found the use of a twostep graduated resection advantageous to ensure adequate resection. A limited resection (3 cm) was performed to allow adequate drainage, followed by a return to the operating room in approximately 1 month to remove any remaining septum.

Because the obstructing septum is usually thick, removal can be difficult. Cooper and Merritt proposed the novel use of a tracheobronchial stent to maintain patency after excision of the obstructing vaginal septum. The stent allows maintenance of patency while epithelialization of the vaginal walls occurs. This technique can be particularly useful in high obstructing septa or in very young patients when access for reapproximation of vaginal mucosa is virtually impossible. In most cases, clamps should be used to isolate a generous vaginal pedicle while the suture is being tied in place to prevent slippage of tissue. Such pedicles generally retract during healing, and formation of a vaginal stenosis is avoided. In most instances, surgery is restricted to excision of the septum. A handheld harmonic scalpel can be advantageous for negotiating an appropriate pedicle and providing excellent hemostasis. Abdominal exploration is usually unnecessary; however, in cases with a high vaginal obstruction, an abdominal approach may provide the best surgical access to unify the vaginas. Uterine reconstruction is not indicated for cases of lateral communication of the uterine horns. Some authors have reported the use of hemihysterectomy in patients with a high, thick-walled obstruction, massive ovarian involvement, endometriosis, or adenomyosis; however, this is generally not recommended in young patients.

Reproductive Performance

Reproductive performance for patients with this disorder is usually consistent with that of patients with a double uterus unless the delay in diagnosis and resection of the obstructing septum has been sufficient to destroy tubal function or to cause the development of endometriosis. Haddad and colleagues' review was notable for a predominance of pregnancies (80%) in the contralateral endometrial cavity.

UNUSUAL CONFIGURATIONS OF VERTICAL-LATERAL FUSION DEFECTS

Unusual configurations of both vertical and lateral fusion defects may occur simultaneously. Figure 25.26 depicts the radiographic evaluation (MRI) of a young woman in whom cryptomenorrhea developed above a transverse vaginal septum. The MRI study depicting the hematocolpos also suggests the longitudinal vaginal septum. Incision, drainage, and resection of the transverse vaginal septum allowed appropriate evaluation of the more proximal müllerian anatomy. The artist's depiction in Figure 25.26A demonstrates the unusual constellation of a uterus didelphys with an intrauterine communication of the cavities and a longitudinal vaginal septum. This type of atypical combination occurs frequently enough to emphasize the importance of proper delineation of individual anatomy preoperatively for proper surgical preparation.

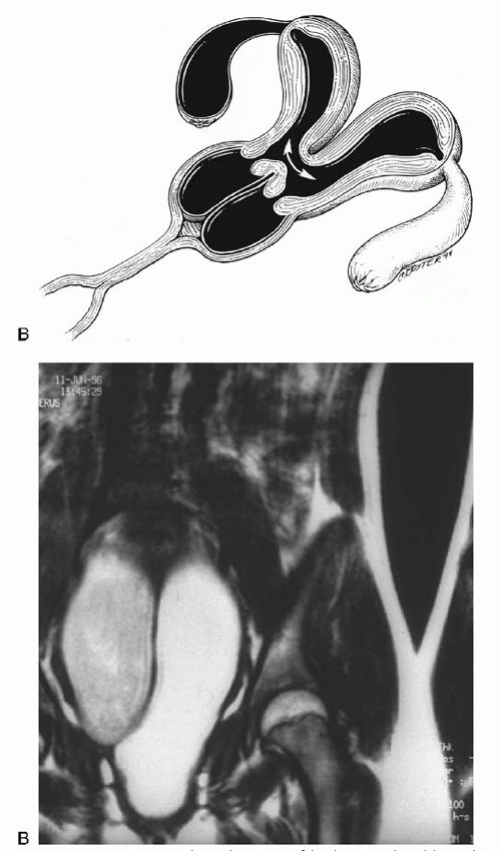


FIGURE 25.26 An unusual combination of both vertical and lateral fusion defects. **A:** A uterus didelphys with an intrauterine communication and a longitudinal vaginal septum are present proximal to a transverse vaginal septum. **B:** The magnetic resonance image demonstrates the presence of both septa.

Müllerian duct anomalies can occur in association with a variety of other problems. For example, Stanton reported that in a series of 70 patients with bladder exstrophy, 30 (43%) had reproductive tract abnormalities. He suggested that the true figures were actually higher. Müllerian abnormalities included absence of the vagina; septate vagina; unicornuate, bicornuate, and didelphic uterus; and absent uterus. Fewer müllerian anomalies are seen with epispadias. Jones investigated anomalies of the external genitalia and vagina in 30 patients with bladder exstrophy seen at the Johns Hopkins Hospital and suggested operative techniques for correction of these anomalies. Techniques for the management of other gynecologic and obstetric problems (especially uterine prolapse) also have been discussed by Weed and McKee and by Blakeley and Mills. A number of other rare combinations of congenital malformations of the vagina and perineum have been found in association with uterine anomalies. Their surgical correction, especially in children, is reported by Hendren and Donahoe and by others. Several authors have considered the uterovaginal anomalies that occur in association with multiple other gastrointestinal and genitourinary abnormalities. Goh and colleagues described an infant girl with complete duplication of the bladder, urethra, uterus, and vagina associated with a urogenital sinus and an anterior ectopic anus. Gastol and colleagues and Magalhaes and associates also described children with complete duplication of the bladder, urethra, vagina, and uterus. These complex anomalies include significantly more defects than lateral fusion concerns in the müllerian ducts. These cases emphasize the variable anatomy in this rare group of anomalies and that a substantial effort should be placed on defining anatomy before surgical exploration and management. Sheldon and colleagues reviewed 13 consecutive cases of vaginal reconstruction in pediatric patients with multisystem anomalies. The review emphasized several important principles involved in the surgical management: (a) all anticipated perineal reconstruction should be performed in a single stage, (b) urethral catheterization has an important role, (c) urinary reconstruction is often intimately involved in the vaginal reconstruction, (d) avoidance of overlapping suture lines is essential for optimal healing, (e) maximum growth potential of the neovagina should be considered, and (f) meticulous follow-up of proper routine dilatation of the neovagina should be expected. Coordinated reconstruction of all organ systems is especially important in these complex cases.

Müllerian duct anomalies are seen with the McKusick-Kaufman syndrome, an autosomal recessive disorder. Other clinical findings reported with this syndrome include hydrometrocolpos, postaxial polydactyly, syndactyly, congenital heart disease, intravaginal displacement of the urethral meatus, and anorectal anomalies. In 1982, Jabs and colleagues added an unusual case to the few cases previously reported in the literature.

Müllerian duct anomalies may also affect the development of the fallopian tube. Although extremely rare, episodes of unilateral or bilateral absence of the fallopian tube have been reported. Of the less than 10 cases in the literature, Eustace reported two of the described cases. He hypothesized that compromise of the local blood supply to the caudal aspect of the müllerian duct was a more likely cause than a fusion disorder. This situation could affect fallopian tube development to a variable extent with even some effect on ovarian development.

BEST SURGICAL PRACTICES

- Vaginal dilatation should be the first line of treatment for creation of a neovagina in patients with müllerian agenesis.
- The gynecologic literature supports the historical role of the McIndoe vaginoplasty as a safe and effective procedure for surgical creation of a neovagina, when necessary. The surgical tenants of the McIndoe procedure are the basis for innovative techniques in surgical creation of the neovagina.
- Appropriate preoperative evaluation of reproductive and pelvic anatomy remains a critical step before a patient is taken to the operating room for treatment of any müllerian anomaly.
- In patients with müllerian agenesis, removal of uterine anlagen that contain endometrium and have the potential to cause obstruction and subsequent pelvic pain should be strongly considered.
- Hysteroscopic metroplasty is a successful, minimally invasive technique for removal of a uterine septum.
- Resection of a uterine septum has been shown to improve pregnancy success in patients with a history of recurrent pregnancy loss.
- Strassmann metroplasty should be considered in select women with a bicornuate uterus who have experienced recurrent pregnancy loss or preterm delivery.

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