Management of Acute Obstructive Uterovaginal Anomalies

ABSTRACT: Obstructive uterovaginal anomalies may present after puberty with amenorrhea, dysmenorrhea, pelvic pain, recurrent vaginal discharge, or infertility. The evaluation of a patient with a suspected obstructive reproductive anomaly should include a detailed medical history, physical examination, and imaging. The genital examination is critical to differentiate a patient with an imperforate hymen from a patient with labial adhesions, urogenital sinus, transverse vaginal septum, or distal vaginal atresia. Pelvic ultrasonography is the initial imaging method recommended for a patient with cyclic pain and amenorrhea or a patient with persistent dysmenorrhea. It is important to note that diagnosis of a uterine or vaginal anomaly by imaging before puberty can be challenging and misleading because of the small size of the prepubertal uterus and the lack of endometrial stimulation and menstrual distention of the vagina. Consultation with a radiologist experienced with imaging of uterovaginal anomalies may be helpful to determine the most accurate diagnosis. In general, obstructive vaginal and uterine anomalies are not surgical emergencies, and the complexities of these conditions are best managed by gynecologic care providers familiar with the surgical management of these conditions. Given the high risk of stenosis and complications associated with transverse vaginal septum, distal vaginal atresia, and cervical atresia, referral to a center with expertise in the management of these anomalies is paramount. The best long-term outcome is achieved with a complete evaluation, clear understanding of the anomaly, mobilization of appropriate surgical resources, sufficient preoperative counseling, and planned surgical intervention.

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

- In general, obstructive vaginal and uterine anomalies are not surgical emergencies, and the complexities of these conditions are best managed by gynecologic care providers familiar with the surgical management of these conditions.
- Obstructive uterovaginal anomalies may present after puberty with amenorrhea, dysmenorrhea, pelvic pain, recurrent vaginal discharge, or infertility.
- Müllerian obstructions are more common in patients with renal, vertebral, anorectal, cardiac, tracheoesophageal, and limb anomalies. Females at high risk of obstruction should be monitored closely for early signs and symptoms of obstruction during pubertal development and initial menses.
- If the diagnosis is not certain or there is a concern for a distal vaginal atresia, transverse vaginal septum, cervical atresia, or an obstructed uterine horn, magnetic resonance imaging (MRI) is recommended.
- Simple incision and drainage of hematocolpos should not be performed because it is associated with a high rate of ascending infection and sepsis and may complicate definitive surgery.
- Given that adolescents may not be sufficiently mature or prepared to adhere to a postoperative dilation schedule, obstetrician–gynecologists and
other gynecologic care providers should consider menstrual suppression and delay of surgical intervention until the patient is prepared to perform postoperative vaginal dilation.

- The best long-term outcome is achieved with a complete evaluation, clear understanding of the anomaly, mobilization of appropriate surgical resources, sufficient preoperative counseling, and planned surgical intervention.

**Background**

Obstetrician–gynecologists should be familiar with uterovaginal anomalies, which may have an incidence as high as 7% in females (1). Obstructive uterovaginal anomalies may present after puberty with amenorrhea, dysmenorrhea, pelvic pain, recurrent vaginal discharge, or infertility. The evaluation of a patient with a suspected obstructive reproductive anomaly should include a detailed medical history, physical examination, and imaging. Accurate diagnosis is critical to manage the obstruction properly. The best long-term outcome is achieved with a complete evaluation, clear understanding of the anomaly, mobilization of appropriate surgical resources, sufficient preoperative counseling, and planned surgical intervention.

**Embryology**

During embryogenesis, the müllerian ducts fuse and develop into the fallopian tubes, uterus, cervix, and upper vagina. Distally, the urogenital sinus separates into the urethra and distal vaginal. Any portion of the development or canalization process may be incomplete, which results in müllerian anomalies. Obstetrician–gynecologists should be aware of obstructive conditions that result from a failure of the müllerian ducts to fuse. Incomplete vertical fusion of the caudal müllerian duct with the sinovaginal bulbs leads to cervical atresia, transverse vaginal septa, or distal vaginal atresia. Incomplete resorption of uterine or vaginal septa leads to rudimentary and obstructed uterine horns or obstructive hemivagina and ipsilateral renal anomaly (also referred to as OHVIRA or Herlyn-Werner-Wunderlich syndrome). Patients with a history of a solitary or multicystic dysplastic kidney have a risk of an obstructed uterine horn or an ipsilateral obstructing vaginal septum with a patent contralateral vagina (2). Most patients with obstructive hemivagina and ipsilateral renal anomaly have a didelphic, bicornuate, or septate uterus. Obstructive uterine horns may be found in a hernia, laterally in the pelvis, or connected to a patent uterus. A rudimentary horn is most commonly encountered when there is vaginal agenesis or in the presence of a renal anomaly (Fig. 1, Fig. 2). Müllerian obstructions are more common in patients with renal, vertebral, anorectal, cardiac, tracheoesophageal, and limb anomalies. Females at high risk of obstruction should be monitored closely for early signs and symptoms of obstruction during pubertal development and initial menses (3). See Figure 3 for the most commonly used classification system for uterine anomalies.

**Symptoms**

The most common symptom associated with obstruction is pain. In a patient who presents with pain, the presence of menstruation does not rule out obstruction. Pain may occur in a cyclic manner or may be continuous. However, in patients who are amenorrheic, a pattern may be more challenging to identify. Other associated symptoms include urinary frequency,
dysuria, hematuria, and urinary retention. Case series report urinary retention in up to 46% of patients with hematometrocolpos caused by an imperforate hymen (4). Because of the rectal pressure associated with low vaginal obstructions, patients may report feeling the need to defecate but may be unable to do so. Patients who have a transverse vaginal septum with spontaneous perforation may report sudden onset of heavy menstrual bleeding. If there has been a chronic microperforation with ascending infection, symptoms may include chronic ongoing vaginal discharge or pelvic inflammatory disease. Patients with longitudinal vaginal or hymenal septa may report difficulty placing or removing tampons. Patients also may report bleeding through tampons and dyspareunia. Additional risks include septal tearing with coitus and obstructed labor.

**Physical Examination**

Examination should begin with an evaluation of sexual maturity (Tanner staging) to assess the degree of pubertal development. An abdominal examination may reveal tenderness or an abdominal mass due to an enlarged obstructed uterus. The genital examination is critical to differentiate a patient with an imperforate hymen from a patient with labial adhesions, urogenital sinus, transverse vaginal septum, or distal vaginal atresia. Downward labial traction can open and separate the labia majora and minora sufficiently to visualize the distal introitus. An imperforate hymen with hematocolpos (blood in the vagina) will reveal a dark-colored or bluish-tinged bulge without hymenal fringe identified (Fig. 4). Distal vaginal atresia will appear as pink mucosa without discoloration (Fig. 5). If the patient can tolerate a digital examination, a shortened vagina on examination without a palpable cervix may be consistent with a transverse septum, cervical atresia, or vaginal agenesis. There may or may not be a bulging proximal vagina if there has been a spontaneous perforation. For the patient with obstructive hemivagina and ipsilateral renal anomaly, there may be a bulge along the patent vagina wall that deviates the patent vagina away from the obstructed side (Fig. 6). Frequently, the cervix on the patent vaginal side is pulled medially toward the septum and may be difficult to palpate. Often, a patient cannot tolerate a speculum examination. If the patient can tolerate a speculum examination, using a smaller, shortened, Huffman speculum or a nasal speculum may allow visualization of a vaginal septal microperforation, which may appear as a small dot or opening with either dark blood or purulent discharge noticeable (Fig. 7). For adolescents, a digital rectal examination is often less painful than a vaginal examination and may be particularly helpful to confirm the presence of the cervix or assess the distance from the hematocolpos to the perineum with vaginal or hymenal obstructions.

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**Figure 3.** The classification system of Müllerian duct anomalies used by the American Fertility Society. (Reprinted from Chandler TM, Machan LS, Cooperberg PL, Harris AC, Chang SD. Müllerian duct anomalies: from diagnosis to intervention. Br J Radiol 2009;82:1034–42.)
Imaging

Pelvic ultrasonography is the initial imaging method recommended for a patient with cyclic pain and amenorrhea or a patient with persistent dysmenorrhea (Fig. 8). A preimaging discussion with a radiologist is helpful. In adolescents, transabdominal pelvic ultrasonography is recommended before proceeding with transvaginal ultrasonography, particularly for those who have not used tampons or had sexual intercourse. Often hematocolpos may be identified posterior to the bladder. Obstructed uterine horns may be difficult to identify by pelvic ultrasonography because they may be located laterally and much higher in the pelvis (5). If the diagnosis is not certain or there is a concern for a distal vaginal atresia, transverse vaginal septum, cervical atresia, or an obstructed uterine horn, MRI is recommended. Magnetic resonance imaging is especially useful to identify the presence and location of the uterine corpus, endometrial cavity, cervix, and proximal vagina (6). The use of vaginal gel or a vitamin E capsule placed in the distal vagina may assist in determining the distance between the distal and proximal vagina, which will help with surgical planning for a transverse vaginal septum resection.

It is important to note that diagnosis of a uterine or vaginal anomaly by imaging before puberty can be challenging and misleading because of the small size of the prepubertal uterus, the lack of endometrial stimulation, and the lack of menstrual distention of the vagina (7). Consultation with a radiologist experienced with imaging of uterovaginal anomalies may be helpful to determine the most accurate diagnosis.

In patients with identified müllerian anomalies, renal imaging should be obtained (8, 9). If the urinary bladder and ureters cannot be distinguished clearly from hydrocolpos, a magnetic resonance urogram or ureteral stenting can be considered (10).
General Principles of Treatment

In general, obstructive vaginal and uterine anomalies are not surgical emergencies, and the complexities of these conditions are best managed by gynecologic care providers familiar with the surgical management of these conditions. Given the high risk of stenosis and complications with transverse vaginal septum, distal vaginal atresia, and cervical atresia, referral to a center with expertise in management of these anomalies is paramount. In fact, hematocolpos creates vaginal distention, increasing the size of the proximal vaginal tissue that can be mobilized to bridge the distance between the proximal and distal vagina. Therefore, delaying surgical intervention may allow further distention of the distal vagina or thinning of the septum, which may decrease the risk of stenosis.

There are multiple strategies to allow time for referral to an experienced gynecologic surgeon for definitive surgery. Simple incision and drainage of hematocolpos should not be performed because it is associated with a high rate of ascending infection and sepsis and may complicate definitive surgery. Indications to expedite surgery include urinary retention, severe pain, and ascending infection. If a patient presents with acute urinary retention, an indwelling urinary catheter may be placed. Stool softeners and pain medication also should be prescribed. If a patient presents with acute pyometrocolpos, broad spectrum antibiotics should be initiated (11) and surgical management should be expedited.

Oral medroxyprogesterone acetate, 20 mg by mouth three times per day, may be given to suppress menses acutely (12). Continuation of complete menstrual suppression can improve patient pain and provide time for referral to a surgeon with expertise. Relief of pain will not be immediate but will improve with time, typically in 7–14 days. Options for menstrual suppression include continuous combination oral contraceptive pills, progestin-only pills or injections, or gonadotropin-releasing hormone agonists (Table 1) (1). Given that adolescents may not be sufficiently mature or prepared to adhere to a postoperative dilation schedule, obstetrician–gynecologists and other gynecologic care providers should consider menstrual suppression and delay of surgical intervention until the patient is prepared to perform postoperative vaginal dilation. Menstrual suppression allows time for an adolescent to mature, understand the diagnosis and dilation process, and actively participate in shared decision making.

Patient and Family Counseling

Many patients and families will have limited understanding of the anomalies of the reproductive tract and will benefit from education regarding embryology and the physiology of menstruation. Patients should be informed that the best long-term outcome is achieved when a complete evaluation to accurately diagnose the obstructive anomaly has been performed, and the patient has been completely evaluated, has been counseled by a surgeon with expertise in managing acute obstruction, and is mentally ready. For procedures that may require vaginal dilation, patients should be educated about the process of dilation and be mature enough and prepared to dilate postoperatively (1, 13). Patients with distal vaginal atresia or transverse vaginal septum can be encouraged to dilate preoperatively to thin the distance between the proximal and distal vagina to decrease the need for graft (14). Patients with these anomalies should be counseled that postoperative dilation may be necessary to prevent stenosis and reoperation.

Surgical Management

After a comprehensive evaluation and stabilization, surgical management of the obstruction may be appropriate. Surgical management is dependent on the type of obstructive vaginal or uterine anomaly. If the obstetrician–gynecologist does not have experience with distal vaginal atresia, high or thick transverse vaginal septa, and cervical atresia, referral to a specialty care center or to a surgeon with expertise in these conditions is warranted. Given the high risk of infection with an obstruction, the use of prophylactic antibiotics is recommended (15).

Imperforate Hymen

Before puberty, the imperforate hymen without hematocolpos appears similar to distal vaginal atresia; therefore, surgical intervention should be delayed until the obstetrician–gynecologist can confirm that a uterus and vagina are present during thelarche. See ACOG...
Distal Vaginal Atresia

Distal vaginal atresia is notable for the presence of the upper vagina, cervix, and uterus without evidence of hymenal tissue at the introitus and is associated with anorectal and urologic anomalies. The first-line approach for management of distal vaginal atresia involves a pull-through vaginoplasty. A pull-through vaginoplasty involves making an introital incision and dissecting between the urethra, bladder, and rectum until the proximal obstructed vagina is reached. The proximal vagina is then mobilized and anastomosed to the introitus. If the proximal vagina is 3 cm or greater from the introitus, the risk of stenosis is high, and an interposition graft may be necessary (17). For high atresia, a combined abdominal...
and vaginal approach using laparoscopy or laparotomy may be considered (18).

**Transverse Vaginal Septum**

The locations of transverse vaginal septa include the upper one third of the vagina (46%), middle one third of the vagina (40%), and lower one third of the vagina (14%) (19). Associated anomalies include imperforate anus, bicornuate uterus, coarctation of the aorta, atrial septal defect, and malformations of the lumbar spine. As in patients with a distal vaginal atresia, the risk of vaginal stenosis is high; therefore, surgical intervention is best performed when the patient is committed to postoperative dilation.

Surgical management includes completely excising the intervening septal tissue. Because the risk of stenosis and injury to the bladder and rectum is high, the surgeon should proceed with caution. Surgeons who are inexperienced with the procedure should consider referring patients to a center with surgeons experienced in excising the septal tissue. The proximal vagina may be mobilized with care to avoid the bladder, rectum, and anastomosed to the distal vagina. A Z-plasty technique may increase the length of the vaginal flaps and decrease the risk of narrowing the vagina (20). For long transverse septa, an interposition graft also may be considered. Postoperative stenting may be required for longer vaginal septa and with interposition grafting (21).

**Obstructed Hemivagina and Ipsilateral Renal Anomaly**

During renal imaging before puberty, patients with obstructive hemivagina and ipsilateral renal anomaly present with a lateral bulging cystic mass at the introitus or an incidental hydrocolpos posterior to the bladder (Fig. 6). Patients who present postmenarche often report severe dysmenorrhea. A vaginoscopy may be performed to assess for a microscopic perforation between the two vaginal canals. Care should be taken to resect as much of the septal tissue as possible with extreme care to avoid both of the cervices, bladder, and rectum. Often the obstructed cervix is dilated and may be pulled toward the medial septal tissue (2, 22). It is important to note that there may be an ectopic ureter in the obstructed vagina, and if patients have urinary drainage after excision of the septum, the dysplastic kidney may need to be excised (23). This condition usually is associated with uterine didelphys, a bicornuate bicornis uterus, or a complete septate uterus.

**Longitudinal Vaginal Septum**

Longitudinal vaginal septum is commonly associated with uterine didelphys, bicornuate uterus, or complete septate uterus. The excision should be performed with extreme care to avoid the bowel, bladder, and urethra, which can be tented toward the septum. Additionally, care should be taken to avoid injuring the cervix or cervices. The vaginal septum typically is clamped and transected, and the mucosa is approximated between the two sides after the septal tissue has been excised.

**Cervical Atresia**

Surgical management of cervical atresia is controversial because of the high risk of complications, stenosis, and ascending infection associated with reconstructing the cervix and vaginal anastomoses. Although there have been reports of menstrual suppression with pregnancies achieved by in vitro fertilization, as well as pregnancies after interposition graft anastomoses, there also have been reports of deaths associated with sepsis and very high rates of reoperation (24). Given the challenging nature of this diagnosis and high risk of complications associated with surgical intervention, patients with suspected cervical atresia should be referred to an experienced surgeon for definitive diagnosis, counseling, and management. If a patient undergoes a hysterectomy for cervical atresia, vaginal elongation for vaginal sexual activity typically can be accomplished by vaginal dilation alone without further surgical treatment. Dilator therapy should be deferred until the patient is ready (25).

**Rudimentary Horn**

A rudimentary horn may be identified at the time of diagnosis of a prepubertal herniorrhaphy, but typically it is resected after puberty (26). Because it is difficult to assess for the presence of a cervix and small upper vagina before puberty, incidentally identified prepubertal horns should not be resected unless there is no potential for fertility. If MRI indicates no endometrium in a uterine horn, resection typically is not necessary, and observation is reasonable. If there is an active endometrium, laparoscopic resection of the uterine remnants to decrease pain, retrograde menstruation, and endometriosis is warranted (25). Menstrual suppression can be considered to decrease pain if a patient prefers to defer operative intervention. Given the high rate of urologic anomalies associated with uterine anomalies, avoiding aberrant ureters during surgery is important (Fig. 6) (27, 28).

**Postoperative Care**

Patients at high risk of vaginal stenosis, including those who have had pull-through vaginoplasty, resection of a thick transverse vaginal septum, or those with a vaginal graft, typically will require postoperative hospitalization on bedrest with an indwelling vaginal stent. A vaginal stent may be fashioned from foam, silicone, or packed gauze covered with a sterile condom or glove. It may be left in place from 3 to 7 days depending on the extent of grafting. While the stent is in place, an indwelling urinary catheter, progestin hormonal suppression, antibiotics, and deep vein thrombosis prophylaxis should be prescribed. Once the stent has been removed, the patient may be discharged with close

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postoperative follow-up for monitoring of healing and dilation, ideally every 1–2 weeks until the patient is comfortable with the dilation process and pain is improved. Dilator therapy should not be initiated until the graft or suture line has healed enough to permit dilation. Gynecologists should continue to monitor patients closely to address patient concerns and encourage adherence with dilation (29).

**Long-Term Follow-up**

All patients with a history of obstruction or müllerian anomalies are at high risk of ongoing endometriosis. Although the initial treatment of the obstruction may alleviate pain, patients should be monitored closely and counseled about menstrual suppression options for ongoing dysmenorrhea (30). All patients with a history of an obstructing septum or distal atresia have a risk of stenosis and may require ongoing dilation or additional surgical intervention (29). Although fertility may not be affected after early intervention for obstructing vaginal lesions, because endometriosis is associated with infertility and uterine anomalies are associated with adverse obstetric outcomes, patients should receive long-term follow-up care to address any reproductive issues proactively (31).

**For More Information**

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

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

**References**


