Aswan Africa Obstetrics & Gynecology Journal, Volume 1/ No. 2/ May 2025 (68-85) online ISSN 3009-6480

Original Article

Updates in The Management of Obstructive Müllerian Anomalies

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Abstract

Keyword:

Obstructive Mullerian anomalies; Embryological disturbance; Female genital system.

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Abstract:

Obstructive Müllerian anomalies (OMAs) are among the most challenging conditions encountered by obstetricians and gynecologists. Embryological disturbances of the normal developmental process of female genital system can lead to a wide group of abnormalities, some of which are easy to treat whilst others are so sophisticated and so demanding in either in diagnosis or their surgical managements. Recurrent cyclic pains or progressive dysmenorrhea are the common theme of the clinical presentation of OMAs. Such characteristic symptoms represent important diagnostic clues for early diagnosis of OMAs. Early diagnosis of OMAs and prompt surgical correction is of a great value in obtaining the dramatic relive of painful symptoms in addition to avoiding the consequences of long-standing retrograde menstruation that represents a real threat to the future natural fertility of those young females. This article highlights briefly the updates of both instant and interval surgical managements of OMAs. For more details the reader can resort to the relevant references where the details of individual surgical approaches are adequately described.

Introduction

Developmental anomalies of the Müllerian duct system are among the most intriguing disorders encountered by obstetricians and gynecologists. The Müllerian duct serve as the embryonic foundation of the female reproductive system, differentiating to form the fallopian tubes, uterus, cervix, and upper vagina. Disruptions in this process can lead to a wide array of malformations, ranging from complete absence of the uterus or vagina to complete duplication of these structures. More frequently only uterine cavity defects are encountered[1] .

Müllerian anomalies frequently coexist with defects in the renal and axial skeletal systems, often becoming apparent during initial assessments for related conditions[2] .

Most of these anomalies occur in individuals with functional ovaries and normally developed external genitalia, typically becoming evident only after puberty. During childhood, normal external development and age-appropriate signs may mask internal reproductive abnormalities. After puberty, affected young women commonly present with menstrual irregularities, while delayed recognition can lead to infertility or pregnancy complications[3]. The prevalence of Müllerian duct anomalies varies widely, with estimates ranging from 0.16% to 10%. Among women experiencing recurrent pregnancy loss and undergoing hysterosalpingography (HSG), prevalence rates are around 8-10%. In contrast, women undergoing elective hysteroscopy (who better represent the general population) show a prevalence of approximately 2-3%. However, a Danish cohort of 622 women aged 20 to 74, assessed by saline contrast sonohysterography, reported a prevalence near 9.8%, particularly elevated among nulliparous women and those with oligomenorrhea[4].

Grimbizis et al. noted prevalence rates of 4.3% in the general and fertile populations, 3.5% among infertile women, and about 13% in women with recurrent pregnancy losses. Byrne et al. found a prevalence of 0.4% (4 per 1000 women) in a prospective study of 2,065 women undergoing sonographic exams for non-obstetric reasons. Nonetheless, ultrasonography may miss certain types of Müllerian defects, implying these figures may underestimate true prevalence[5].

Surgical Management of Obstructive Müllerian Duct Anomalies

Imperforate Vagina:

Imperforate vagina (IV), often caused by an imperforate hymen (IH), usually presents as hematometrocolpos. These cases are sometimes initially mistaken for constipation, ovarian masses, or urinary retention with bladder distension. Failure to examine the external genitalia in young girls can lead to improper diagnosis and management [6] .

A recent review of 35 studies covering 61 patients showed that premenarchal girls presenting with recurrent cyclic pain, constipation, tenesmus, back pain, urinary difficulties, lower abdominal pain, abdominal swelling, and absence of menstruation should be suspected of IV until proven otherwise. Physical examination may reveal a tender, pale, or bluish obstructing hymenal membrane, and ultrasound is a valuable diagnostic tool[7] .

The treatment goal is timely hymenotomy to drain accumulated menstrual blood, followed by hymenectomy to prevent recurrence. Follow-up is important to monitor for recurrences. However, hymenectomy may be controversial in conservative cultures due to sociocultural values. As a result, virginity-preserving procedures that maintain an annular hymenal opening are appealing alternatives. A circular hymenotomy performed with a 10 mm laparoscopy trocar under general anesthesia successfully preserved the hymen in 36 cases without complications. Follow-up

confirmed the patency of the annular hymen in all patients. This minimally invasive, safe, and effective technique should be considered a feasible alternative to traditional hymenotomy as it allows restoration of normal hymenal anatomy without[8].

Microperforated Hymen (MH):

Microperforated hymen is a rare partial obstruction of the hymenal membrane that often remains unnoticed until puberty, when it begins to affect the individual's quality of life. Unlike imperforate hymen, MH may allow some menstrual flow but can cause complications such as infections because retained secretions in the vaginal canal are exposed to the external environment, creating a pathway for pathogens. A typical presentation includes difficulty with vaginal intercourse[9].

Some patients experience regular menstruation, though bleeding may be scanty, prolonged, or pass through one side of the vagina. Physical examination and imaging help identify the micro perforation. Treatment by hymenotomy under general anesthesia is effective and generally results in satisfactory outcomes, enabling normal reproductive function post-procedure [10].

This anomaly is often overlooked, which may lead to underreporting of its true prevalence. Delayed symptom onset and psychological discomfort can postpone medical consultation and diagnosis. With thorough physical examination, MH can be identified and treated early, ideally before marriage, preventing complications including urethral dilation secondary to coital activitys[11].

Transverse Vaginal Septum (TVS):

Transverse vaginal septum (TVS) is classified based on its location from the vaginal opening—low (<3 cm), mid (3-6 cm), and high (>6 cm)—as well as by thickness (thin: <1 cm, thick: ≥1 cm) and whether it is perforated or complete. Complete TVS commonly presents with primary amenorrhea and cyclical lower abdominal pain,

whereas microperforated septa may give symptoms like microperforated hymen or may be asymptomatic[12] .

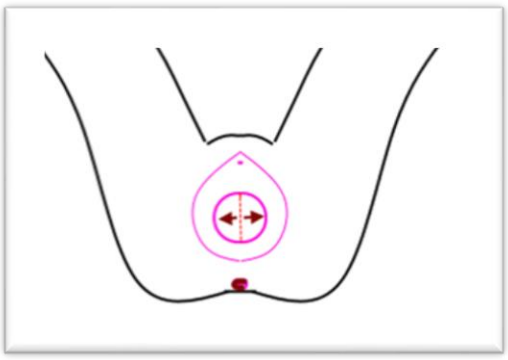
Surgical Management of TVS:

Simple Septum Excision:

Traditional management involves excision of the septum; the procedure is especially suitable for thin septa. This technique entails incision and evacuation of retained blood, followed by full excision of the septum and circumferential suturing. Use of vaginal estrogen tampons postoperatively has been reported to reduce the risk of vaginal stenosis[13].

Simple Vaginal Flap Technique:

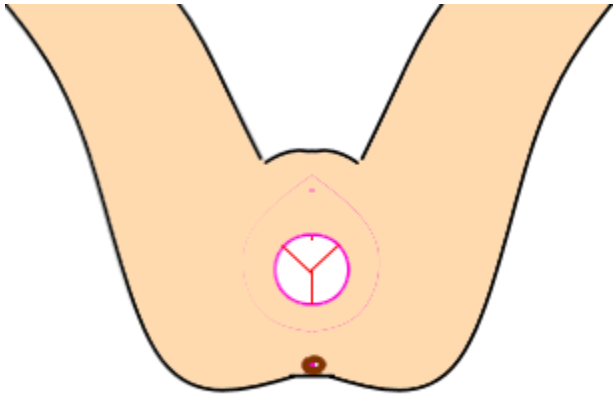
This alternative involves a vertical incision of the distal mucosa of the vaginal septum, submucosal dissection to create two lateral flaps, excision of the septum, and suturing of the flaps to the proximal vaginal mucosa to cover raw surfaces[14].



Interdigitating "Y" Flap:

This technique offers better preservation of vaginal length and less scarring. The septum, usually thick and of dual embryonic origin with distinct distal squamous and

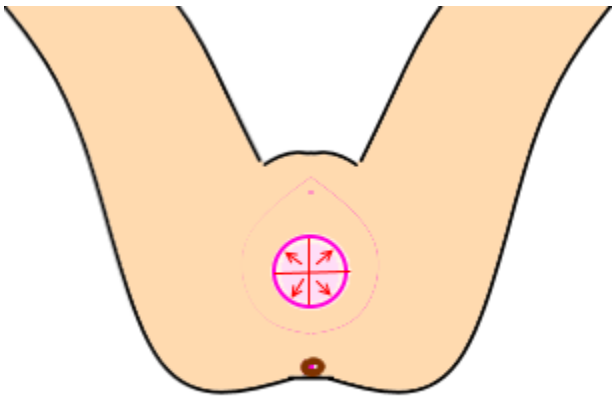
proximal glandular linings, is divided into distal and proximal flaps. The “Y” flap is particularly suitable for low and mid-vaginal septa due to better exposure and careful dissection[12].



the Y flap technique

Double-Cross-Plasty Technique:

Similar to the “Y” flap but involves creating four distal and four proximal flaps instead of three each. Both techniques help preserve vaginal length and reduce stenosis by distributing wound tension evenly. However, the “Y” flap is preferred because it involves fewer flaps of larger size, enhancing blood supply and technical ease[12].



Double-Cross-Plasty Technique

Vaginoscopic Resection with Hymen Conservation:

For patients desiring to preserve hymenal integrity due to cultural reasons, vaginoscopic resection performed under laparoscopic guidance with insertion of a Foley catheter helps maintain hymenal preservation. This method is effective in selected cases with relatively thin septa and results in good menstrual flow and no septal closure after 12 months [15].

Vaginal Atresia:

Vaginal atresia is a rare congenital condition characterized by partial or complete absence of the vagina while the uterus remains functional. Its incidence ranges from 1 in 4,000 to 1 in 10,000 females[16].

The uterus and cervix may appear normal or exhibit fusion or resorption defects, while the vaginal segment is replaced by fibrous tissue. Patients typically present at puberty with primary amenorrhea, cyclical pain, and a pelvic mass from hematocolpos. Untreated cases risk retrograde menstruation leading to endometriosis and pelvic adhesions[17].

Surgical treatment aims to relieve symptoms and drain hematocolpos. However, stenosis and strictures frequently develop, especially with longer obstruction segments. Historically, hysterectomy was used to alleviate symptoms and prevent complications such as pelvic inflammatory disease, but advances in surgical techniques now allow symptom relief with fertility preservation[18].

Surgical Approaches for Vaginal Atresia:

Modified McIndoe's Procedure:

After diagnosing segmental vaginal atresia, vaginoplasty involves creating a neovagina with an artificial skin graft (Terudermis®). A reverse T-incision on the lower uterine body enables canalization between the uterus and neovagina. A balloon catheter maintains patency, and subsequent cervical dilatations prevent reobstruction. At six years follow-up, normal menstruation was reported without recurrent hematometra[19].

Laparoscopic Vaginoplasty with Peritoneal Flaps and Cervicoplasty:

Two peritoneal flaps are harvested laparoscopically from the bladder and sacroiliac ligament areas. Following blunt dissection of the rectovesical and vesicocervical spaces, these flaps are transposed to line the neovaginal cavity. Cervicoplasty is performed to maintain the cervical canal, with Foley catheter and vaginal mold placement. Regular follow-up ensures no inflammation or obstruction, preserving menstrual flow[20] .

Combined Laparoscopic and Vaginal Cervicovaginal Reconstruction:

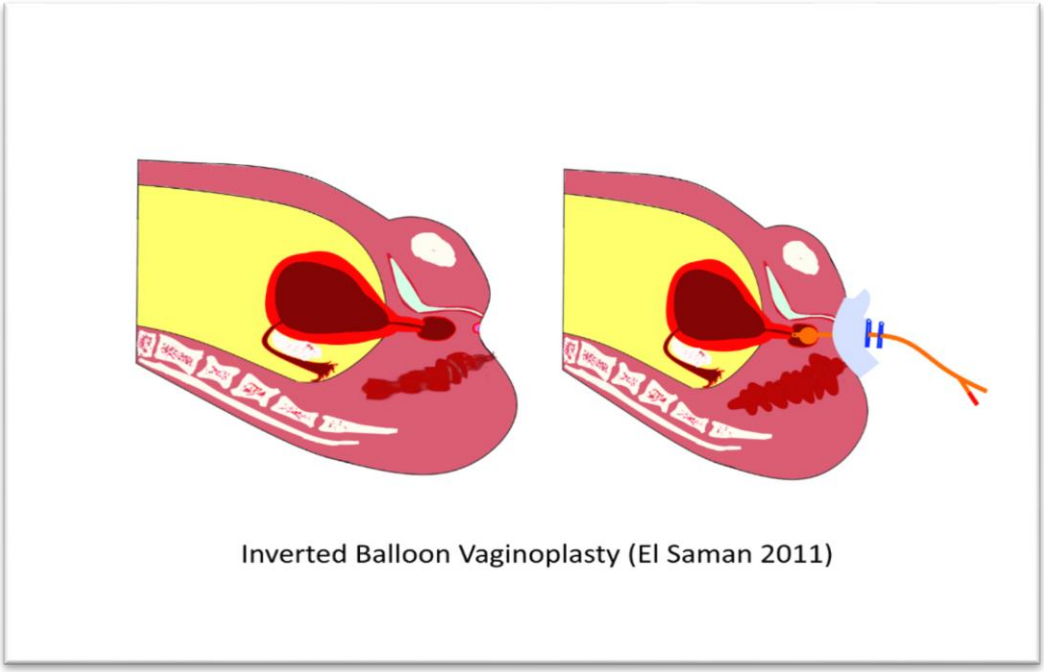
Using split-thickness skin grafts, this technique reconstructs cervicovaginal aplasia with good outcomes in menstruation resumption and no stenosis reported over several months follow-up[21].

However, most of the aforementioned techniques are demanding a sufficient skills, special instrumentation, and excessive dissection with subsequent scaring. The problem with scar formation is dual and affection both cosmetic and functional outcomes. In addition, scar contractures might result in recurrences. Therefore, there

was a real need for innovative approaches that can avoid the shortcomings of the available procedures[22-23]

Mechanical Tissue Expansion (Inverted Balloon Vaginoplasty):

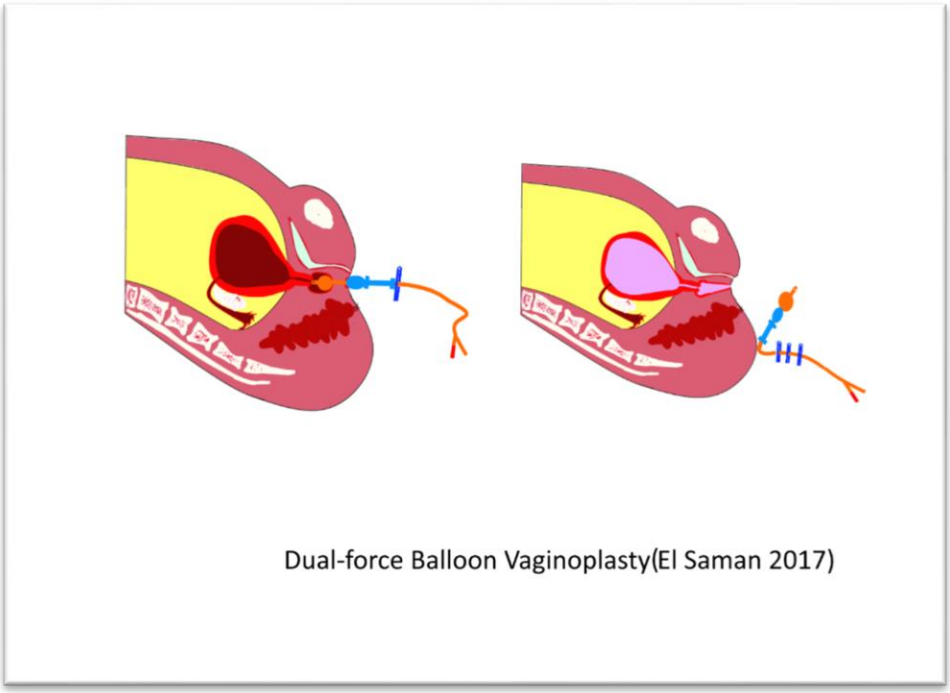
This was an innovative Egyptian method introduced by El Saman et. al, in the year 2011. Inverted balloon vaginoplasty (IBV) is only feasible in cases with a distended upper vaginal pouch. This pouch was pulled down via balloon catheter inserted under laparoscopic guidance. The supporting platform for pulling down was placed over the perineum in the form of bulky multilayered dressing. The outcomes of IBV were so satisfactory and provided a cosmetically appealing outcomes in a minimally invasive manner and giving a naturally covered neovaginal segment



Inverted Balloon Vaginoplasty (El Saman 2011)

Mechanical Tissue Expansion (Dual-force Balloon Vaginoplasty:

The dual-force technique uses simultaneous traction (pulling down) and pushing to avoid bulky dressings. The balloon catheter was inserted under laparoscopic or ultra sonic monitoring and an acrylic olive was positioned on the vaginal dimple. The acrylic olive was pushed up expanding the vaginal dimple this pushing of course was accompanied with simultaneous pulling on the upper vaginal pouch. This technique required minimal dissection and resulted in neovaginas lined with native epithelium, effective menstruation, and successful long-term marital relations with successful pregnancy in some cases[22].



Dual-force Balloon Vaginoplasty(El Saman 2017)

Isolated Cervical Aplasia or Obstruction:

Cervical atresia may be congenital or acquired, with acquired cases common after surgical procedures or conditions like radiotherapy or infections. Congenital cervical aplasia typically presents in adolescents with recurrent abdominal pain, necessitating multidisciplinary evaluation[23].

Treatment Options for Cervicovaginal Atresia:

Hysterectomy:

Reserved for postmenopausal women or those not desiring fertility, and in selected adolescent cases with severe disabilities or complex uterine anomalies[24].

Genitoscopic Ultrasound-Guided Cervix Fenestration and Balloon Dilatation:

Using vaginoscopy under anesthesia and ultrasound guidance, a needle is advanced through the atretic cervix into the uterine cavity for drainage. Balloon dilatation is performed repeatedly to maintain patency, with successful menstruation and symptom relief after multiple sessions[25].

Uterovestibular Anastomosis (UVA) via Vaginal Approach:

Minimally invasive vaginal surgery creates an anastomosis in congenital cervicovaginal agenesis cases, preserving fertility with favorable long-term anatomical and functional outcomes[27].

Laparoscopic-Assisted Uterovaginal/Vestibular Reconstruction:

Combining laparoscopy and vaginal techniques, a tunnel is created for uterine opening and hymenal incision anastomosis. Postoperative mucus production, absence of infection, and gradual neovaginal development occur without molds or pessaries[28].

Robotic-Assisted Laparoscopic Cervicouterine Anastomosis:

The cervix is mobilized and dilated laparoscopically, followed by incision and catheter placement to maintain patency. Closure is performed with absorbable sutures. Menstruation resumes with pain relief, offering a fertility-preserving alternative to hysterectomy[29].

Laparoscopic Decompression:

Used for symptomatic relief where specialized teams are unavailable, with subsequent referral for definitive treatment[30].

Abdominal Uterocervical Anastomosis:

In cases of uterine isthmus agenesis, abdominal anastomosis restores normal menstruation and relieves pain, maintaining patency on imaging[31]

Laparoscopic Cervico-Isthmic Anastomosis:

For traumatic cervix-uterine disjunction, laparoscopic end-to-end anastomosis with stenting prevents adhesions and stenosis, allowing symptom resolution[32].

Endoscopically Monitored Canalization:

Laparoscopic canalization with vaginoscopic or tactile monitoring treats isolated cervical atresia, ensuring bladder and urethral integrity. Follow-up shows regular menstruation, though further refinement is needed for cases with vaginal aplasia[33].

Microsurgical End-to-End Cervico-Cervical Anastomosis:

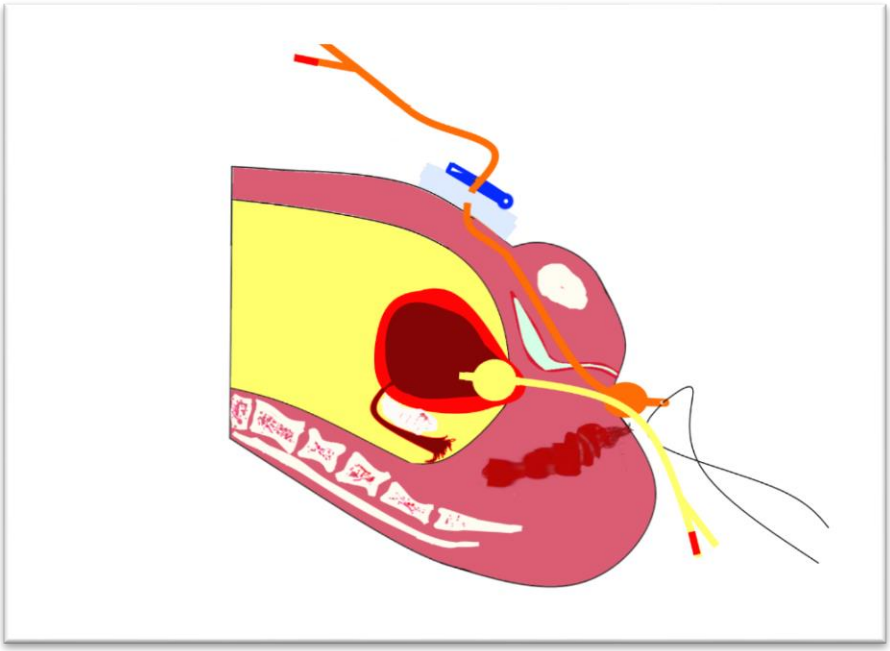
Rare congenital cervical fragmentation can be treated with microsurgical anastomosis, using Foley catheter stenting, leading to restored menstruation[34].

Trans-Myometrium Hysteroscopy-Guided Canalization:

A novel procedure uses laparoscopic-guided trans fundal hysteroscopy for cervical canal development in obstructive cervicovaginal agenesis, combined with balloon vaginoplasty if needed. Successful drainage and canalization are achieved, with regular menstrual cycles reported[35].

Combined retropubic balloon vaginoplasty and laparoscopic canalization:

Retropubic balloon vaginoplasty and laparoscopic canalization (RBV-LC) was a novel combination of procedures that offers a minimally invasive handling for cervicovaginal aplasia. The RBV-LC procedure was accomplished efficaciously in 4 females complaining recurrent cyclic pains as consequence of combined cervicovaginal Cystoscopy was accomplished to confirm urethral and urinary bladder integrity. RBV-LC was a practicable, effective, less invasive method for surgical treatment of cervicovaginal aplasia[36].



Disclosure of interests: The authors declare no relevant financial, personal, political, intellectual or religious interests.

Funding: No funding is relevant to this work.

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