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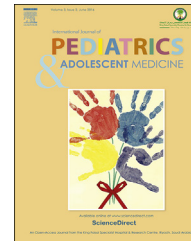


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CASE REPORT

Low complete transverse vaginal septum, vesico-ureteric reflux and low anorectal malformation: Case report and review of literature



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KEYWORDS

Transverse vaginal septum;
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Abstract The complete, imperforate transverse vaginal septum is one of the rare anomalies of the female reproductive tract. This anomaly is a disorder of vertical fusion of the Müllerian ducts and can present with or without obstruction. It has been classified as Type IIA as per the American Fertility Society (AFS) classification. Its thickness and site varies in the vaginal canal; the upper and middle third septa are common. The diagnosis can be made in newborns, infants, and adolescent girls. It is found associated with urological anomalies, anorectal malformation (ARM), and bicornuate uterus. We present a case of a 13-year-old girl with hematometrocolpos due to an imperforate complete low vaginal septum, left-sided vesico-ureteric reflux (VUR), and ectopic anus (low ARM). Copyright © 2016, King Faisal Specialist Hospital & Research Centre (General Organization), Saudi Arabia. Production and hosting by Elsevier B.V. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

1. Introduction

Müllerian anomalies are congenital defects of the female reproductive tract resulting from failure in the development of the Müllerian ducts and their associated structures. The incidence of these anomalies has been estimated to be

1 in 70,000 females [1]. The cause of the anomalies is not known, but it is currently believed to be multifactorial. The patient might present in the neonatal period, during adolescence, or early adulthood, and the anomaly might affect the reproductive capacity. Transverse vaginal septum is a rare anomaly, and a low septum is even rarer. When clinically suspected, investigations leading to the diagnosis include imaging methods such as hysterosalpingography, ultrasonography (USG), magnetic resonance imaging (MRI), and cystoscopy. The management depends on the age at presentation, the site, and the thickness of the septum. Regular follow-up and strict adherence to vaginal dilatation are pre-requisites for good long-term results.

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2. Case summary

A 13-year-old pre-menarchal girl presented with history of episodic, colicky abdominal pain since 3 months of age. She had a history of anoplasty performed for ectopic anus in the neonatal period. There was a large, non-tender cystic lump arising from the pelvis. On rectal examination, a cystic bulge could be felt anteriorly up to the anal verge. The external genitalia were normal. There were three openings in the perineum with the anus placed slightly anteriorly (Fig. 1). The Tanner staging for both the external genitalia and the breasts was Stage II.

Abdominal ultrasound suggested hematometrocolpos with fullness of the left pelvi-calyceal system. Micturating cystourethrography (MCU) showed grade 5 vesico-ureteric reflux (VUR) on the left side and a large, distended bladder. Perineal ultrasound revealed a low transverse vaginal septum of 8–10-mm thickness. An MRI of the pelvis showed hematometrocolpos and a low transverse vaginal septum. The distance between the lower end of the vagina and the introitus was 15 mm (Fig. 2). During examination under general anesthesia, the vaginal pit was shallow, but there was no bulging at the outlet. Vaginoscopy was attempted, but it was not possible as there was a complete low septum.

Intraoperatively, stay sutures were placed on the septum, and the septum was marked off (Fig. 3). It was aspirated and opened by a cruciate incision. The margins were then sutured (Fig. 4). Postoperatively, the vaginal form was kept.

A postoperative ultrasound showed no evidence of hematometrocolpos. The patient is awaiting management of ARM & VUR.

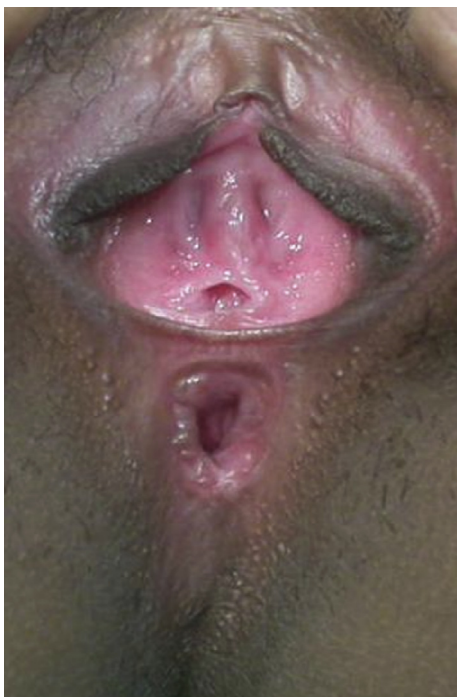


Figure 1 Local examination showing three openings in the perineum with anus placed little anteriorly. No evidence of bulging membrane (as seen in imperforate hymen).

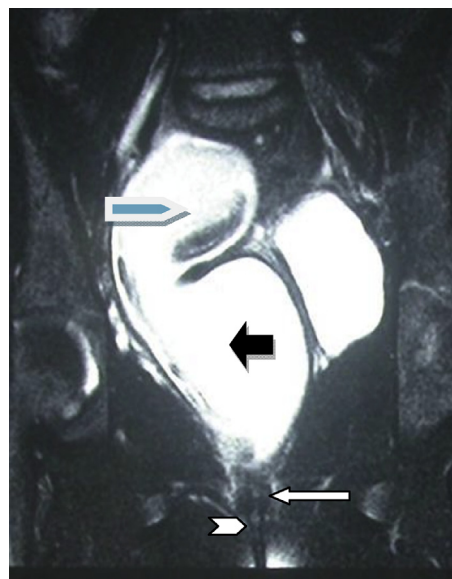


Figure 2 MRI of the pelvis showing hematometrocolpos and a low transverse vaginal septum (white arrow). The uterus (blue arrow) distended upper vagina (black arrow) and the collapsed lower vagina (white arrow head) are seen. The distance between lower end of vagina and the introitus was 15 mm.

3. Discussion

The female genital system develops from the paired Müllerian ducts and in close association with the urinary system and hindgut [1]. The paired Müllerian ducts develop from the coelomic epithelium lateral to the mesonephric ducts and cross medially to fuse in the midline [2]. The most cranial parts of the Müllerian ducts, which remain separate from the fallopian tubes, and the caudal segments proceed caudally to join the urogenital sinus, where they produce an elevation called the Müllerian tubercle [1,2].

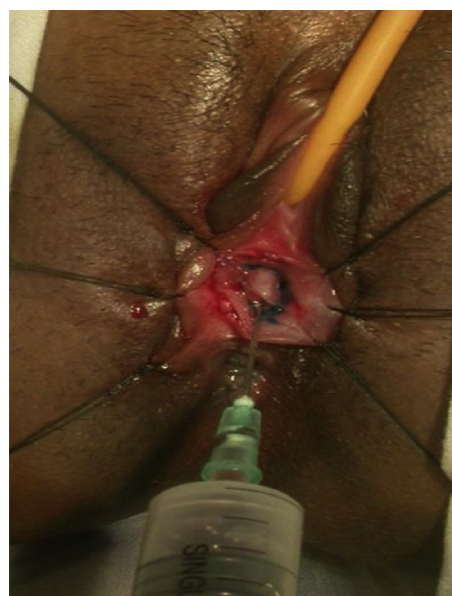


Figure 3 Intra-operative image showing the stay sutures and the cruciate incision.

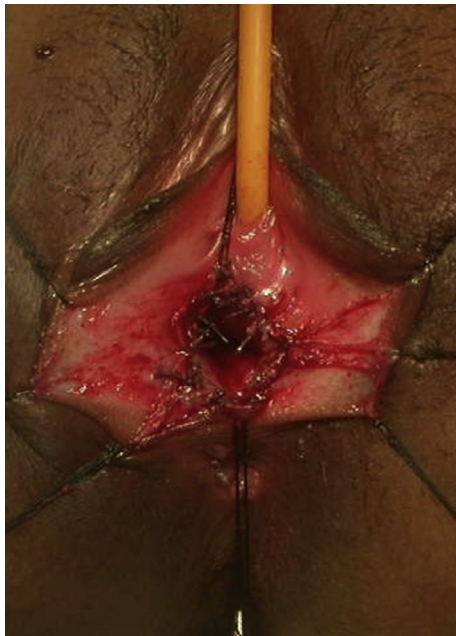


Figure 4 Intra-operative image showing completed suturing.

These fuse and form the uterus, cervix, and the proximal two-thirds of the vagina [2]. The sinovaginal bulbs are paired caudal ectodermal outgrowths that proliferate to form a cord of tissue that develops into the distal vaginal plate. This plate is later canalized in a caudal to cranial direction to form the distal vagina [2].

The close proximity of the mesonephric and paramesonephric ductal systems helps to explain the common association of paramesonephric abnormalities and ipsilateral renal anomalies. Even mesodermally derived organs as cranial as the cervical vertebra and the tracheo-esophageal anlagen can be affected in association with congenital abnormalities of the mesonephric and paramesonephric ductal systems, as is observed in the vertebral defects, anal atresia, tracheo-esophageal fistula with esophageal atresia, radial and renal dysplasia (VATER) and Müllerian duct aplasia, renal aplasia, and cervicothoracic somite dysplasia (MURCS) associations [2].

The anomalies of the female reproductive system can be grouped into four main categories as per the American Fertility Society (AFS) classification [3]:

- 1) Those resulting from either hypoplasia or agenesis,
- 2) Those caused by vertical fusion, which can be obstructive or non-obstructive (canalization abnormalities resulting from abnormal contact of the müllerian structures with the urogenital sinus),
- 3) Those resulting from lateral fusion (duplication), and
- 4) Unusual configurations and combinations of defects.

Delaunay first described the transverse vaginal septum in 1877 [4]. It is believed to arise from a failure in fusion or canalization (or both) of the urogenital sinus and Müllerian ducts. The cause is unknown although some cases might be the result of a female sex-limited autosomal recessive transmission [1] or exposure to certain agents in utero [5]. The septum is composed of fibrous connective tissue and

vascular muscular elements. Although the lower surface is always covered with squamous epithelium, the upper surface can be covered by either vaginal squamous epithelium or columnar epithelium, with or without metaplasia [1].

The estimated incidence is 1 in 70,000 females [2]. A complete transverse vaginal septum can be located at various levels in the vagina, but there is a higher frequency in the middle and upper third of the vagina. In one large series by Lodi in 1951, the distribution was 46% in the upper vagina, 40% in the middle vagina, and 14% in the lower vagina [6]. The septa are usually less than 1 cm thick and frequently have a small central or eccentric perforation [2], giving an impression of a vaginal vault without a cervix [3]. The septa are frequently accompanied by urinary tract abnormalities, such as unilateral renal agenesis, ectopia, or fusion [7], as well as musculoskeletal, gastrointestinal, and cardiac defects [1]. The most common genital anomalies associated are uterus bicollis with an obstructing partial vaginal septum and the unicornuate uterus with a rudimentary horn [7]. It has also been associated with bilateral tubal atresia [1]. Incomplete treatment of Transvaginal ultrasound (TVS) allows menstrual flow to escape periodically. The diagnosis can be made on transvaginal sonography, computed tomography, and magnetic resonance imaging [3].

The clinical presentation depends on whether it is complete or partial. With complete septum, the menstrual blood accumulates in the genital tract resulting in hematocolpos and hematometra. Such patients usually present at adolescence with cyclic lower abdominal pain, and occasionally, a lower abdominal mass (hematometra) can be palpable. Hematocolpos might not develop until puberty, but complete obstruction might cause serious compression of the surrounding structures in neonates and infants. A high transverse vaginal septum might cause cyclic hematuria if communicating with the urinary system. An incomplete septum allows partial egress of menstrual blood, and such patients complain of dysmenorrhea, hypomenorrhea, dyspareunia, foul-smelling vaginal discharge [2,4], and dystochia. It might also be asymptomatic and appear as an incidental finding during routine gynecologic examination [4]. However, unlike an imperforate hymen, examination of the genitalia reveals no evidence of bulging at the introitus.

Transperineal, transvaginal, and vesical hydrosonegography have been used in defining both the vaginal structure and pathological conditions; however, the fine details as to whether the septum is complete or incomplete, or the number and dimensions of vaginal compartments cannot be determined [1]. A transvaginal approach, coupled with catheter instillation of saline solution, provides a more accurate assessment [1]. MRI can help determine whether a cervix is present so that a high septum can be differentiated from congenital absence of the cervix [2].

The treatment of a TVS is individualized, and the goal is to relieve cyclic abdominal pain and prevent development of endometriosis to preserve fertility [5,8,9]. Vaginal dilators are the preferred non-surgical choice for patients with small septa [5]. Dilatation techniques can be used in lieu of surgery, before surgery in order to improve outcomes, and after surgery to prevent strictures, scarring, or stenosis of surgical site [2]. A low transverse vaginal septum is treated by a transverse incision over the vault of the short vagina followed by anastomosis after identifying the

cervix [5]. An indwelling stent or a soft foam rubber vaginal form is used in the immediate postoperative period followed by vaginal dilatations. Coital function is usually normal after surgery, and pregnancy rates are 40–50% for septum in the lower or middle third of the vagina.

If the patient's pain from hematocolpos is manageable, surgery can be delayed with suppression of endometrial activity by a GnRH agonist or continuous oral contraceptives [2]. This treatment might allow time for dilation of the lower vaginal segment, potentially improving the ease of surgical repair [2], especially in cases of a high septum.

Small vaginal septa (<1 cm in thickness) can be treated by excision with a simple end-to-end anastomosis of the vaginal epithelium or a Z-plasty [5]. Larger septa (>1 cm) might require pre-operative vaginal dilatation followed by a longitudinal Z-plasty technique to reduce stenosis and contraction of scarring at the site [5]. The Olbert balloon catheter technique has also been described to maximize the vaginal mucosa available for anastomosis and to avoid postoperative narrowing of the vagina [5].

Vaginal stenosis at the site of resection is the most common complication [2]. Postoperative vaginal dilation or a vaginal mold might help in decreasing the scarring and stenosis of the surgical site [5]. Postoperative vaginal dilation is critical to the success of the procedure, which is usually affected by the apprehension of the adolescents. Such apprehensive patients might be better served with a long-term mold or stent [5]. Daily guidance with patience is necessary to help these adolescent girls gain confidence for vaginal dilatation [5].

Adverse outcomes after surgery, such as dyspareunia, menstrual irregularities, fertility issues, and preterm labor, are common. Recognition and intervention at younger ages by draining the collected blood and possibly preventing endometriosis is necessary to preserve fertility [5,8,9]. Rock et al reported that patients were less likely to conceive after surgical correction, and even if they did so, there was a 50% chance of spontaneous abortion [9]. Therefore, these patients and their family members should be educated about these potential long-term complications and about regular follow-up while attempting to conceive.

4. Conclusions

Müllerian anomalies are a diverse group of developmental disorders involving the internal female reproductive tract.

Establishing an accurate diagnosis is hence essential for planning treatment and management strategies.

Conflict of interest

None to be declared by the authors.

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None to be declared by the authors.

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