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(CASE REPORT)

Hematocolpos- variations: Case report

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Abstract

Hemato/hydrocolpos is an obstruction of the menstrual flow due to an anomaly of the genital tract, with imperforate hymen being the most common one, which is found in neonatal, infant, and adolescent girls. Diagnosis is often missed or delayed due to its rare incidence and nonspecific symptoms (usually misdiagnosed as constipation. In case of late diagnosis/ misdiagnosis and/or delay in treatment, late complications such as tubal adhesion, pelvic endometriosis, and infertility may develop. This article aimed to review the diagnosis and treatment of these urogenital anomalies by describing embryology, clinical presentation, imaging findings, surgical management, and postoperative outcomes.

Keywords: Hematocolpos; Hemoperitoneum; Imperforated Vaginal Hymen; Septum Vaginalis.

1. Introduction

Hemato/hydrocolpos is a medical condition in which menstrual blood or secretory fluid accumulates in the vagina due to vaginal obstruction. Hemato/hydrocolpos are caused by congenital urogenital anomalies or acquired vaginal occlusion due to infection, trauma, or sexual abuse [1]. There are mainly four congenital causes: imperforate hymen, distal vaginal agenesis, complete transverse vaginal septum, and obstructed hemivagina and ipsilateral renal anomaly (OHVIRA), the schemas of which are shown in Fig. 1 below:

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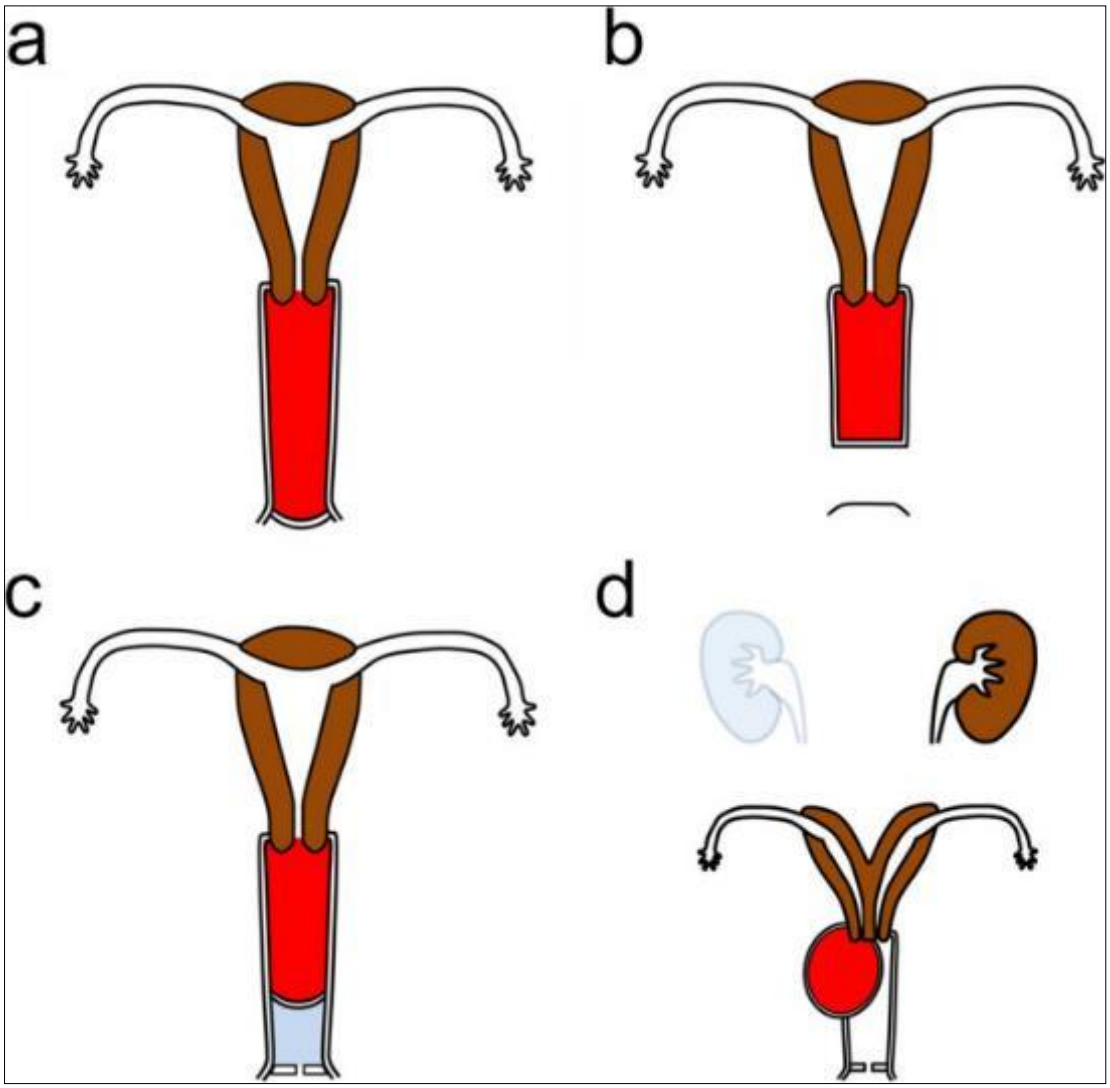


Figure 1 Coronal views of congenital urogenital anomalies causing hematocolpos. The accumulated blood in the vagina is colored in red.

a. Imperforate hymen.; b. Distal vaginal agenesis; c. Complete transverse vaginal septum; d. OHVIRA

1.1. Embryology of vaginal development

The embryonic origin of the vagina has been a historically hotly debated issue with several different contributions and origins described. Current molecular studies show the whole vagina epithelium is derived from the paramesonephric (Müllerian) duct with bone morphogenic protein 4 (BMP4) reshaping the intermediate mesoderm derived Müllerian duct into the vaginal primordium. Transgenic studies in mice also identified the developmental origin of vaginal epithelium derived solely from Müllerian duct epithelium. Vaginal development is also under negative control of androgens.

After fertilization of the egg, its division also begins and at 3rd or 4th day the molecule is formed. The molecule at 7th day transforms into a blastocyst which is distinguished in trophoblast and cavity of the blastocyst. On the inner surface of the wall of the blastocyst, a compact accumulation of cells is formed, the embryoblast, from which the embryo will develop. The amniotic cavity appears between the cells of the embryoblast, while on its inner surface the yolk sac develops. The amniotic cavity is lined internally by a layer of endothelial cells, which constitutes the amnion. Between these two cavities (the yolk sac and the amnion), the three primary germinal skins of the embryo are finally separated, from which various tissues of the body of the fetus will arise in the first 20 days. The three layers are the ectoderm to the amniotic cavity, mesoderm and endoderm or inner germinal skin. The combination of trophoblast and primary mesoderm forms the chorion. The two cavities moving towards the middle of the blastocyst are connected with its trophoblast with a stalk of mesoderm, the ventral stalk, from which the umbilical cord develops. During the third week

of fetal life from the middle vegetative skin the prokidneys and Wolff ducts are formed which are the primordial ureters and which end in the amara (primordial intestine).

In the 4th week, the prokidney is subducted and in its place develops the mesonephric, which is covered by the epithelium of the visceral cavity. This epithelium during the fifth week shows a tape-like papilla that extends from the 6th thoracic to the 2nd sacral somatotomy and is called the urogenital band. This band at its cephalic end is folded, while at the lower part it multiplies to form the primordial gonadal tumor, where primordial germ cells migrate from the wall of the cyst which are of extraembryonic origin. Thus, the undifferentiated gonads (gonads) are created. The gonads consist of three types of cells: 1. cells of the visceral epithelium, 2. Cells of the mesenchyme and 3. primary germ cells. With the migration of the primary germ cells, the gender differentiation of gonads into testes or ovaries begins, determined chromosomally from the moment of conception. When differentiated into female, the epithelium of the visceral cavity (germ epithelium) forms the genital trabeculae, between which the primary cells are installed, which in the case of the ovary are the oocytes. Thus, the ovary consists of: the medulla (zona vasculosa), which originates from the mesenchyme, and the cortex, which originates from the epithelial cells of the visceral cavity. These surround the oocytes forming the primordial follicles.

At the same time, Muller duct is formed, outside of Wolf duct, from diving of the epithelium of the visceral cavity. This new duct ends up in urogenital sinus, originating from the amara, forming there the crest of Müller. The fallopian tubes are formed from the head parts of Muller pores, while from the two caudal parts, which join together, the metrocoelic tube is formed, from which finally, with the disappearance of the septa in the middle line, the uterus and the upper part of the vagina are formed. Partially or totally remaining of the diaphragm results in the formation of congenital defects, which affect to a different extent the reproductive capacity of the woman. Overtime, the amara with a mesenchymal septum is divided into anterior and posterior quarters. The Wolf ducts export anteriorly and intestine posteriorly. Then the anterior part of the amara divides into two other cavities, from which will create the bladder and urogenital sinus.

The urogenital sinus joins the ducts of Muller and creates the genitalia tubercle, genital folds and genital tumors. From the genital tubercle the clitoris is formed, the post and its bridle, as well as the labia minora. The labia majora will arise from the genital tumors. Finally, the anus will be formed from a petal of the amara, while the perineum will be formed by two appendages from the mesenchyme between the urogenital and anus fold.

The uterovaginal development is shown in the figure below. After the caudal tip of the fused Müllerian ducts reaches the urogenital sinus, sinus-derived endodermal cells proliferate and form a solid sinovaginal bulb. The sinovaginal bulb proliferates and forms a vaginal plate. Proliferation continues at the cranial end of the plate and increases the distance between the developing uterus and urogenital sinus, and the core of the sinovaginal bulb degenerates and forms a cavity at 17–18 week's gestation. By the 5th month of pregnancy, the vaginal plate is completely canalized. The upper and lower parts of the vagina are considered to be derived from the Müllerian ducts and sinovaginal bulbs, respectively.

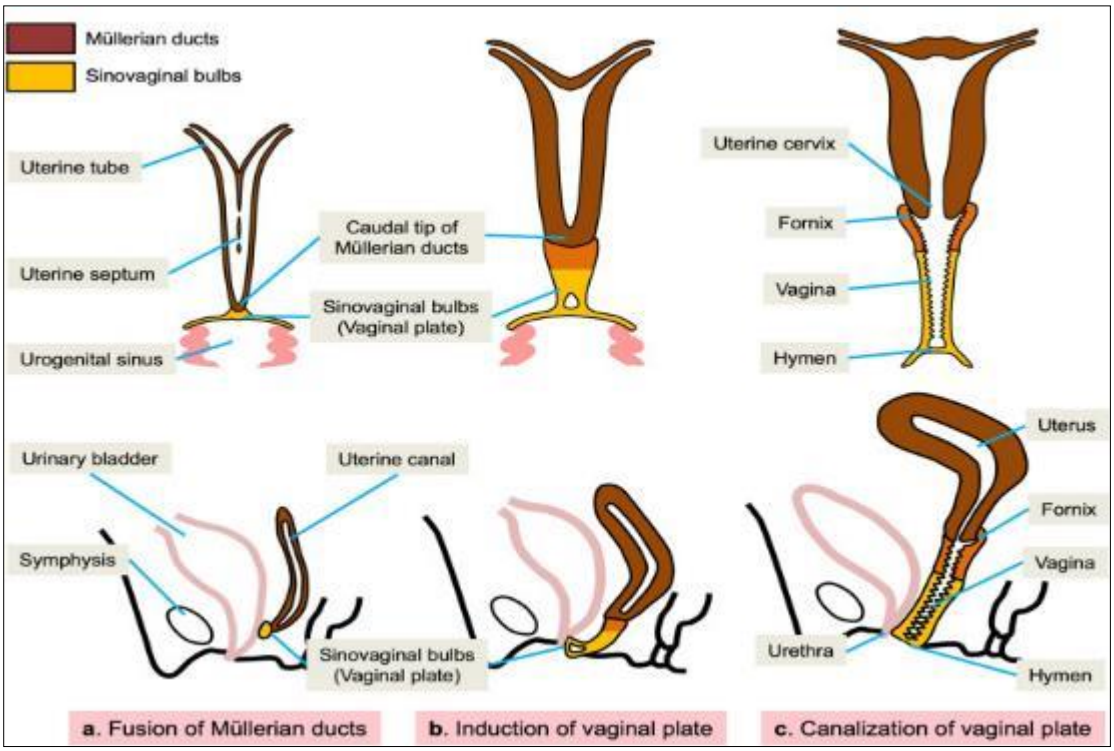


Figure 2 Schematic illustrations show a.Fusion of Mullerian Ducts b.Induction of vaginal plate c.Canalization of vaginal plate

a. After the caudal tip of the fused Müllerian ducts reaches the urogenital sinus, a sinovaginal bulb grows out of the sinus.

b.The sinovaginal bulb proliferates and forms a solid vaginal plate. Proliferation continues at the cranial end of the plate.

c. By the 5th month, the vaginal plate is entirely canalized and forms the vagina. The hymen is a thin mucosal tissue between the urogenital sinus and the sinovaginal bulb. A small opening is generally developed in the hymen during perinatal life.

2. Congenital anomalies

2.1. Imperforate hymen

In imperforate hymen, the vaginal orifice is occluded by a hymen without an opening. The prevalence of imperforate hymen is estimated at one in 1000 to one in 2000 females [5,21]. Most cases are thought to occur sporadically, despite the fact that rare cases of familial imperforate hymen have been reported [11,22].

In female neonates with an imperforate hymen, hematoma/hydrocolpos caused by maternal estrogen are incidentally discovered [23,24]. However, most of them with imperforate hymen present with symptoms such as: primary amenorrhea, cyclical pelvic pain or urinary retention secondary to hematocolpos or hematometrocolpos at puberty [8,19,25,26].

Hematocolpos due to imperforate hymen can be diagnosed easily by perineal inspection, which reveals bulging, bluish hymen without a vaginal opening [19,27,28]. However, this condition can be easily missed or misdiagnosed if a thorough recording of history is not taken or perineal examination is not performed. Abdominal ultrasonography can reveal hemato/hydrocolpos and MRI can depict the whole vagina distended with hematoma and the bulging hymen protruding from the introitus. MRI is a valuable imaging tool for assessing the extent of hematocolpos, hematometra and/or hematosalpinx, the thickness of the imperforate hymen, and related complications such as infection and endometriosis. The schematic illustration of the imperforate hymen is shown in Figure 3:

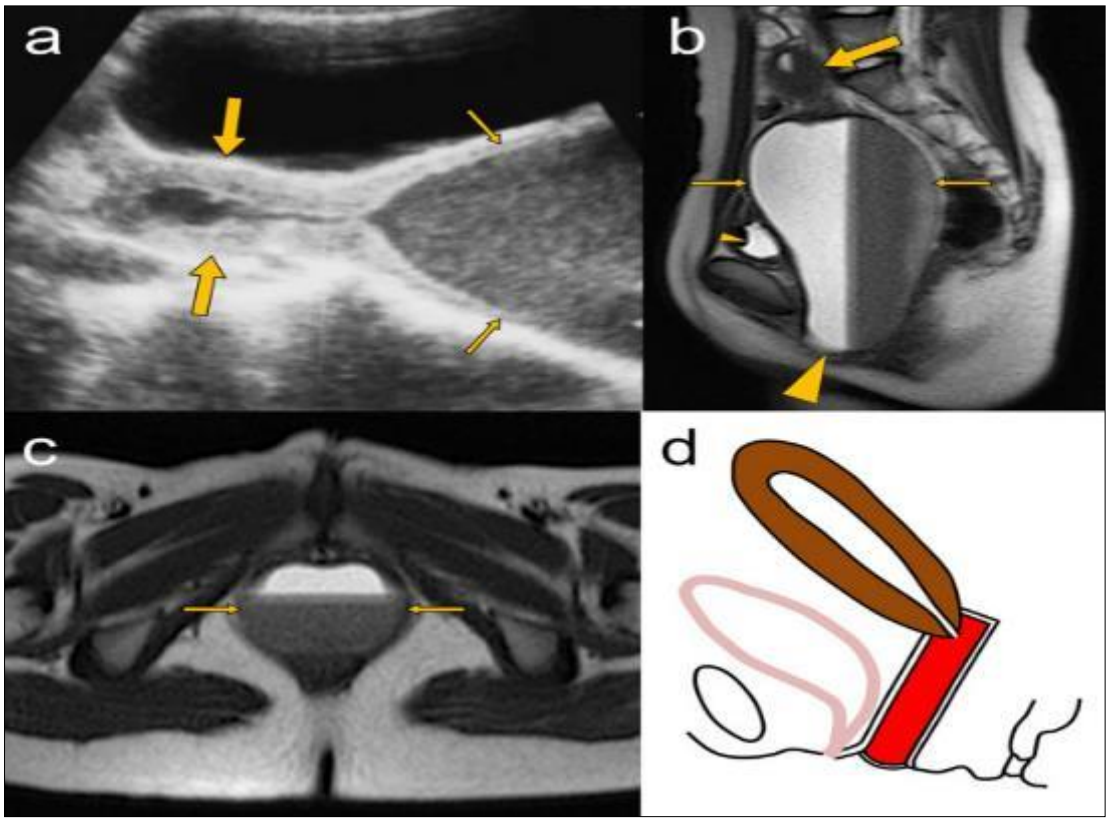


Figure 3 Schematic illustration of the imperforate hymen

Imperforate hymen is typically treated with hymenotomy (surgical incision of the hymen) or hymenectomy (surgical removal of the hymen). Although there are no differences in results between the two surgical techniques, hymenectomy was shown to be associated with higher problems than hymenotomy. Due to the significance of the initial intercourse hemorrhage, hymen-preserving operations such as simple vertical incision and annular hymenotomy are occasionally performed. Although problems such as re-closure, vaginal adenosis, or vaginal adhesion are recorded in only 6.6% of postsurgical patients [11] , close monitoring is required to guarantee no recurrence or sequelae. Long-term outcomes of appropriate surgery for imperforate hymen are good.

Prepubertal diagnosis of imperforate hymen has several advantages, including appropriate scheduling of surgery and prevention of severe complications associated with delayed treatments, such as tubal adhesions, pelvic endometriosis, and infertility. Given that the diagnosis of imperforate hymen is easily made by inspection of female genitalia, pediatricians should incorporate genital inspection of prepubertal females into standard clinical practice [11,21,27].

2.2. Distal vaginal agenesis

According to reports, vaginal agenesis affects one out of every 4000 to 10,000 females [32]. Vaginal agenesis is categorized into three types: complete, proximal, and distal [33], with the latter accounting for approximately 5% of the overall number. Distal vaginal agenesis occurs when the urogenital sinus fails to form the bottom region of the vagina or the vaginal plate fails to canalize partially. The American Society for Reproductive Medicine (ASRM) classifies distal vaginal agenesis as a class I A abnormality [34]. The majority of patients with distal vaginal agenesis experience a variety of symptoms as a result of menstrual blood retention during adolescence. Distal vaginal agenesis, as well as imperforate hymen, can be detected during genital examination; ideally, this condition should be identified during the newborn and prepubertal periods to decide the best timing for surgery and avoid difficulties associated with delayed treatment. [6]

Perineal examination indicates the absence of a hymen and vaginal opening, but with a slight concave indentation [6,9,35]. In individuals with hematocolpos, transabdominal ultrasonography can disclose the inflated upper section of the vagina with moving internal echoes (Figs.4a), whereas transperineal ultrasonography can show the length of the atretic vaginal segment. Although a CT scan can also reveal distal vaginal agenesis (Fig. 4b), an MRI can better identify the atretic vaginal segment (Figs.4c,d), allowing for a differential diagnosis from the more common hematocolpos caused by an imperforate hymen. MRI is required for the morphological assessment and surgical planning of distal

vaginal agenesis (adjusting the length of the graft) [9,35,37]. Figure(4e) provides a schematic representation of distal vaginal agenesis.

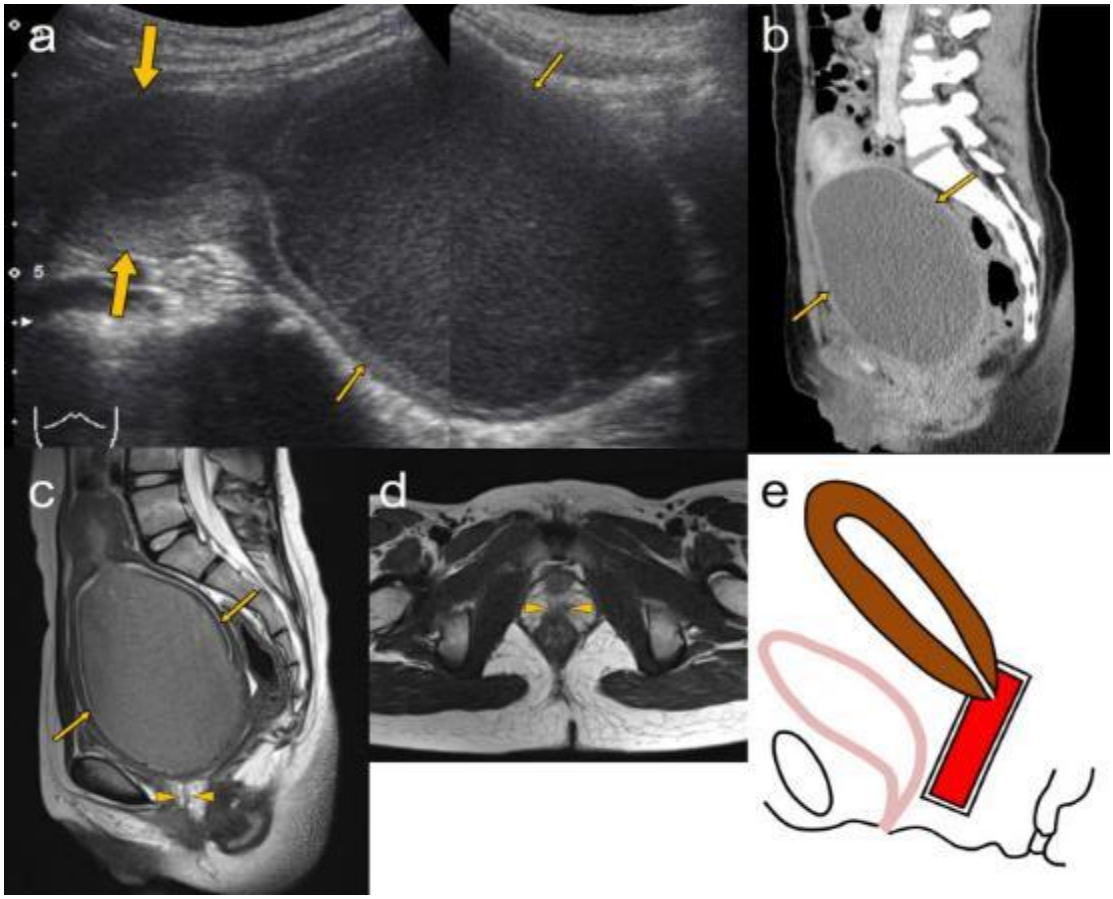


Figure 4 Schematic representation of distal vaginal agenesis.

Local pull-through vaginoplasty with direct anastomosis of the upper vaginal mucosa to the introitus via a perineal incision is used in surgical treatment when the distance between the perineal surface and the caudal side of the enlarged vagina is 2 cm or less. When a major portion of the vagina is missing, vaginoplasty using skin or bowel grafts can be performed. Fibrin glue promotes graft stability. The timing of surgery is controversial. If the diagnosis is made during the neonatal or prepubertal era, curative surgery should be delayed until hematocolpos appears during menarche. This is due to more dilated upper vaginal segments, making it easier to establish the appropriate surgical incision path and decreasing the graft length.

The most prevalent late consequence following a vaginoplasty is postoperative vaginal stricture, which has a negative impact on the patient's sexual life and pregnancy. Vaginal prosthesis, inflated silicone stents, and estrogen ointment are used to prevent vaginal stricture following surgery. MRI is a valuable imaging technique to exclude postoperative problems such as hydrometrocolpos caused by vaginal stenosis or shortening. (Fig.5)

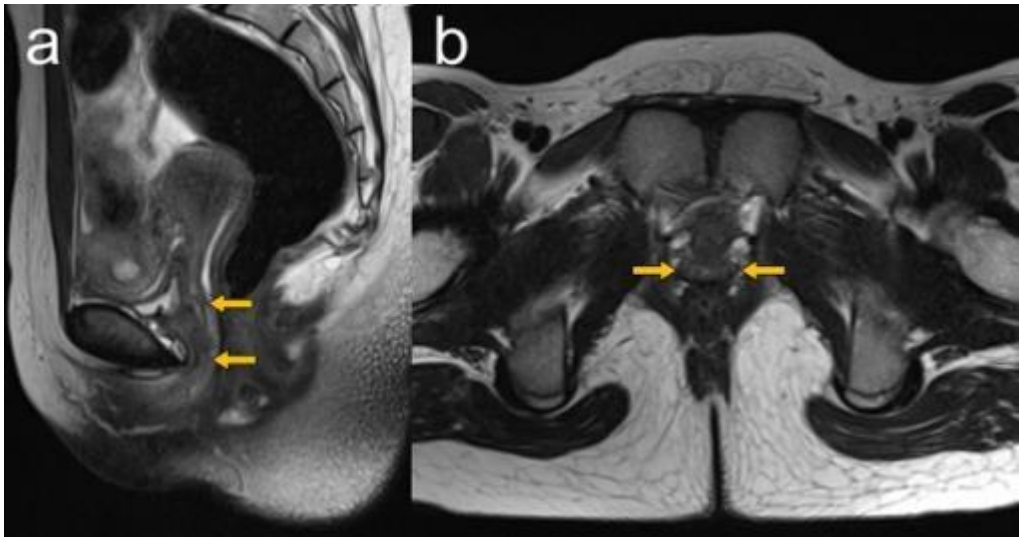


Figure 5 MRI is a valuable imaging technique to exclude postoperative problems such as hydrometrocolpos caused by vaginal stenosis or shortening.

2.3. Transverse vaginal septum

A partial vaginal septum is uncommon, while a complete septum is relatively rare. According to reports, transverse vaginal septum affects one in every 30,000 to 84,000 females [41]. The transverse septum can be seen at any level of the vagina; however, it is most commonly found in the upper vagina (46%), followed by the mid vagina (35%), and the lower vagina (14%) [42]. A transverse vaginal septum, a class IA anomaly according to the ASRM classification, is thought to emerge when tissue resorption fails between the sinovaginal bulb and the caudal point of the united Müllerian ducts [43]. This septum is a membrane of fibrous connective tissue that contains vascular and muscle components [41].

Thicker septa are more typically observed near the uterine cervix. During genital examination, no abnormalities are found in patients with upper or middle vaginal septum. A transverse vaginal septum is typically identified in persons who report with primary amenorrhea and cyclical pelvic pain owing to hematocolpos. Although early detection and correction of the transverse vaginal septum can improve outcomes, it is difficult to make a clear diagnosis before menarche [13].

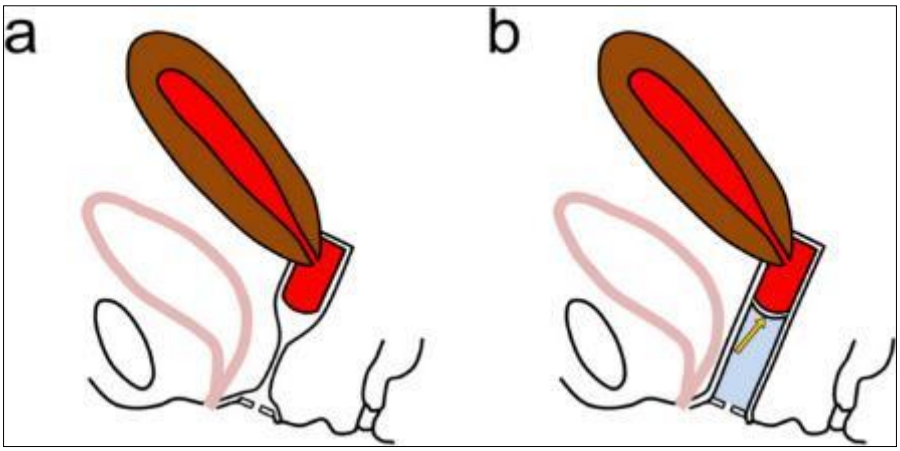


Figure 6 This technique has been shown to be effective for determining the level and thickness of the vaginal septum.

In patients with hemato/hydrocolpos, abdominal ultrasonography might detect the enlarged upper region of the vagina. In patients with a transverse vaginal septum, MRI can show the swollen upper and collapsed lower parts of the vagina, allowing the differential diagnosis to be made between hematocolpos caused by an imperforate hymen and vaginal agenesis [13]. However, standard MRI makes it difficult to discern between the vaginal septum and the collapsed typical lower vaginal wall. Infusing an adequate volume of ultrasound jelly through the vaginal introitus can extend the bottom

section of the vagina, resulting in better MRI delineation of the vagina septum. This technique has been shown to be effective for determining the level and thickness of the vaginal septum [10,35,43]. The schematic illustration is provided in Figure 6:

Before performing an end-to-end anastomosis of the upper and lower vaginas, all vaginal septa should be eliminated. To reduce the possibility of postoperative stenosis, a double-crossed Z-plasty with eight vaginal mucosal flaps is routinely performed. When end-to-end anastomosis is difficult due to thick vaginal septa, a biological graft is used to reconstruct a normal-sized vagina. To keep the vagina open after surgery, a vaginal mold or silicone stent is frequently used. The most prevalent consequence is post-operative vaginal stenosis. However, successful surgery can result in patients having a satisfying sex life.

2.4. Obstructed hemivagina and ipsilateral renal anomaly (OHVIRA)

Hydrocolpos due to congenital urogenital anomalies could be discovered during the prenatal and postnatal periods owing to the collection of secretions in the obstructed vagina under the influence of maternal hormones [47]. However, most patients are detected at puberty with several symptoms due to hematocolpos or hematometrocolpos. Clinical symptoms are often nonspecific and include suprapelvic tender mass, cyclical lower abdominal pain, constipation, vomiting, and urinary retention. Thus, such patients could be misdiagnosed as acute appendicitis, ovarian torsion, or urinary infection. Gold standard method for detecting hemato/hydrocolpos is MRI with its high-resolution and soft tissue contrast which provides useful information for the differential diagnosis of the obstructive causes. Ultrasonography, computed tomography (CT) can also be used for these patients.

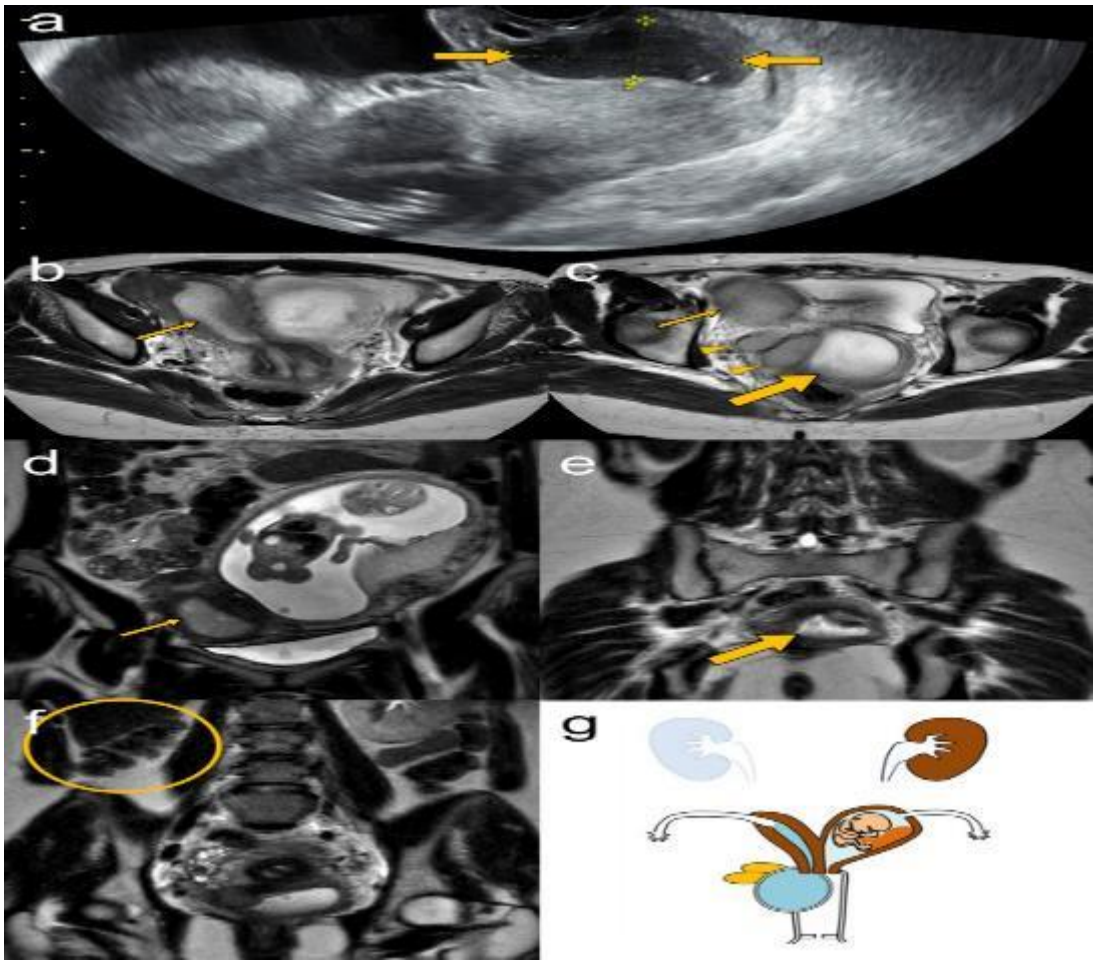


Figure 7 Congenital vaginal obstruction.

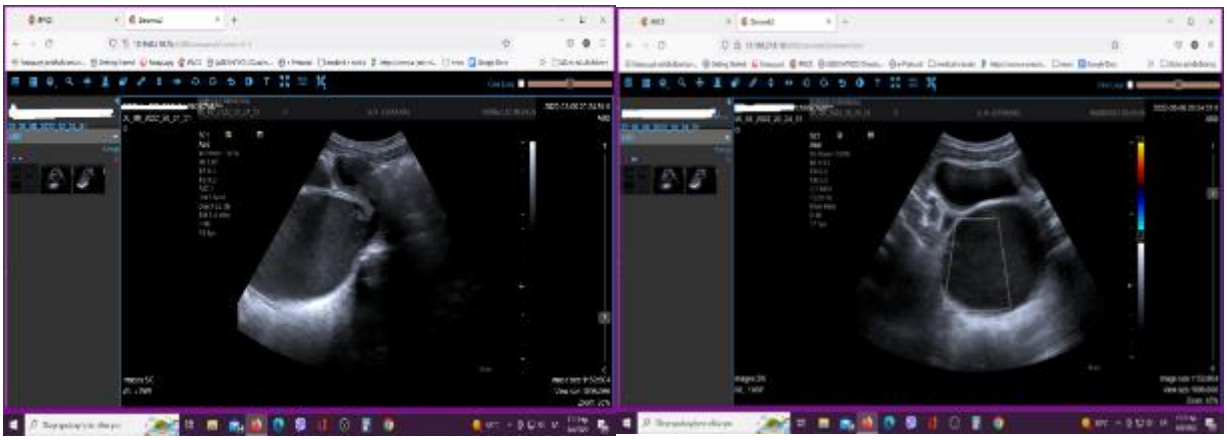
Surgical treatment is necessary for patients with hemato/hydrocolpos caused by congenital vaginal obstruction. Drainage of accumulated blood/ secretions in the vagina is preferable, but risks such as retrograde infections caused by the long-term transvaginal drainage should be considered. The uterus should not be squeezed for drainage as tubal adhesions and peritoneal endometriosis due to endometrial cell dissemination may lead to infertility. Below, we review

the embryology of the vaginal development along with the clinical and imaging findings, treatment options and possible outcome of the previously described congenital causes of hemato/hydrocolpos which is key to an early diagnosis and effective treatment.

Resection of the vaginal septum to remove obstruction is the preferred treatment, and complete resection is thought to result in good results and fertility [51]. Postoperative problems are uncommon; nonetheless, vaginal adenosis, vaginal stenosis, and re-closure of the vaginal septum have been observed [53,55]. Hemihysterectomy for a blocked uterine horn is not suggested due to the probability of fertility in a previously obstructed uterus. Women with uterus didelphys have a high pregnancy rate of 80%; nevertheless, higher rates of early delivery (22%), miscarriage (74%), and cesarean section (more than 80%) are seen. Early detection and surgical treatment can reduce problems and protect future fertility. The aberrant differentiation of the Wolffian and Müllerian ducts can result in renal defects, the most common of which is renal agenesis. Unilateral renal agenesis occurs in 1 out of every 1100 women, and more than 30% of those affected have ipsilateral Müllerian abnormalities [47,58]. If unilateral renal agenesis or other renal abnormalities are observed in newborn, baby, and prepubertal females, OHVIRA and other Müllerian defects commonly accompanied by the syndrome should be investigated [47,54,57].

3. Case presentation

A 13-year-old girl came to the Emergency Department of General Hospital of Lefkada due to a reported 24-hour urinary retention. From the ultrasound examination, an intra-abdominal as well as endometrial collection of hypochoic fluid, possibly blood, is observed. The bladder is full of urine. Catheterization of the bladder was performed with immediate remission of symptoms. This was followed by another ultrasound examination without differentiation of the image, with an image of fluid collection inside the abdomen, uterus and vagina. The possible diagnosis of atresia of the hymen and hematovaginal - hematouterine was made. This was followed by an overview of the external genital organs, where the diagnosis was made of the presence of a transverse septum in the lower part of the vagina and not atresia of the hymen due to the non-stretching of the hymen and the increased thickness of the septum. Due to the lack of specialized staff for such operations, she was referred to a 3rd degree Hospital with a Pediatric Gynecology department. The same diagnosis was made by the Alexandra Hospital where she was referred by the Children's hospital. The mother, a British citizen, of her own free will, immediately went to England where the operation was performed.



Figures 8 and 9. Fluid collection inside the abdomen.

4. Conclusion

In this review, we have described the embryological findings, imaging findings, and treatment options for congenital urogenital anomalies causing hematoma/hydraulic, namely, intact hymen, superior vaginal agenesis, complete transverse vaginal septum, and OHVIRA. Early identification and treatment reduce the risk of pelvic endometriosis. As a result, radiologists should be conversant with these imaging characteristics, and other clinicians, particularly pediatricians, gynecologists, and urologists, should keep these genitourinary abnormalities in mind while treating newborns, babies, and preadolescent women.

Compliance with ethical standards

Disclosure of conflict of interest

No one of the Authors have conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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