Vaginal Outlet Obstruction - A Review of Cases
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Abstract
The paper focuses on anatomical varieties of developmental anomalies of female vagina’s outlet, emphasizing the role of their early recognition in preventing retrograde menstruation by surgical treatment at the beginning of puberty.

Three different types of vaginal outlet obstructions are described: imperforate hymen - from the group of hymen misdevelopment, transversal vaginal septum - from the group of failed transverse Mullerian fusion, and vaginal atresia - from the Mullerian aplasia group of disorders. Diagnoses were made in different age of life, from newborn to the teenage years. Clinical and ultrasound examinations dominated in establishing the diagnosis. Computed tomography and magnetic resonance imaging were included in search for associated congenital abnormalities. The time period of the analysis was 2003-2012.

The total number of vaginal outlet obstructions was 19. Imperforate hymen was diagnosed in 15 patients - 4 neonates, 1 toddler, 2 pre-pubertal girls without any oestrogen stimulation and 8 pubertal females may be helpful in early diagnosis in preventing morbidity and mortality.

Prenatal ultrasound and careful genital examination of the newborn females may be helpful in early diagnostics of very distal vaginal obstructions like imperforate hymen and distal vaginal septum. Vaginal atresia is difficult to notice before the puberty and the menstruation cycle. A search for associated genital anomalies is necessary. In pubertal girls who haven’t got a menarche and suffer the recurrent lower abdominal pain, the pelvic ultrasound is recommended to identify the cause(s).

Keywords
Vagina; Developmental abnormalities; Atresia; Hydrocolpos; Hematocolpos

Abbreviations
IH: Imperforate Hymen; MRI: Magnetic Resonance Imaging; CT: Computed Tomography; TVS: Transverse Vaginal Septum; VA: Vaginal Atresia

Introduction
The vaginal outlet mechanism consists of undisturbed fluid passage from vagina through hymenal opening. The genital tract outflow is important for secretion and menstrual effluxion and as a pathway in reproductive function. Congenital outflow obstruction may occur at different levels and with different clinical presentations [1].

Embryological development of vagina results from lower portion paramesonephric ducts fusion and regression forming the uterovaginal primordium (gives rise to the uterus and superior part of vagina). Contact of the uterovaginal primordium with urogenital sinus induces formation of paired outgrowths named sinovaginal bulbs. The sinovaginal bulbs fuse to form the vaginal plate. The cells of the fused bulbs undergo apoptosis to form the lumen of the vagina. Until late fetal life the lumen of the vagina is separated from the cavity of the urogenital sinus by a membrane – the hymen [2,3].

Misdevelopment can result in any of the following three:
• imperforate hymen (failure of epithelial degeneration),
• low, mid, or high transverse septum of the vagina (incomplete unit), or in
• atresia of the vagina resulting in persisting of a portion of solid cells cord.

Recently interest has focused on expression and function of the mammalian HOX genes as a possible etiology of these genital developmental abnormalities [4].

Despite different origin of some parts of the vagina and hymen, their obstructed forms are clinically manifested as hydro (metro) colpos in the neonatal period and hemato (metro) colpos at the beginning of puberty.

In this report on our ten-year experience the importance is illustrated of early diagnosis in preventing morbidity and mortality.

Variation in Hymenal Development
Regular development of ring like hymenal structure provides normal vaginal outflow. Until late fetal life, the lumen of the vagina is separated from the urogenital sinus by invagination of the posterior wall forming a hymenal membrane. The hymen usually ruptures during the perinatal period and remains as a thin fold of mucous membrane circumferentially at the entrance of the vagina [2].

There is a spectrum of variations in hymenal configuration, like different type of fenestration, septa, etc. with more or less clinical significances. Imperforate hymen (IH), as the result of failed hymen rupture is the most important in this misdevelopment spectrum.

Imperforate Hymen
IH is an external congenital genital anomaly when nonperforated membrane of the hymenal tissue covers the entrance into vagina. The membrane composed of vaginal epithelium and epithelium of the urogenital sinus is interposed by mesoderm [5]. Imperforation of
hymenal tissue may also be present in variations as micro perforate, septate, stenotic or cribriform hymen.

The maternal hormones affect the reproductive tract of the female infant both in utero and during the early neonatal period, increasing the secretions of the uterine and cervical glands. The possible explanation is that the target tissue may be more sensitive to or exposed to higher levels of maternal estrogen [6]. By the age of one month vaginal secretion becomes poor and alkaline, the epithelium is thin, the individual cells small and devoid of glycogen, and the basal layer comparatively inactive [7].

If maternal estrogenic stimulation and vaginal obstruction coexist hydro (metro) colpos will develop. The retained fluid in hydrometrocolpos consists of vaginal and uterine secretions and may be acidic and serous or mucoid, containing large numbers of desquamated epithelial cells, leukocytes, and erythrocytes [8].

In the setting of imperforate hymen, efflux of these secretions is not possible, and the newborn may present with a whitish bulging mass filling the introitus (hydrocolpos or mucocolpos). If hydrocolpos is small or not detected at birth, secretions resorb up to the end of first months of life, and the condition may be undetected until puberty. Also, if there is a low level of maternal hormones, there is no extra genital secretion and the obstruction will usually remain unnoticed until puberty.

Hydrocolpos is the mass usually consisting of fluid-filled vagina but the uterus is displaced upwards. Why the uterus is not always dilated is not known.

The clinical presentation in neonates is a translucent bulging membrane-hymen between labia as a result of distension of the vagina and uterus due to accumulation of excessive mucous secretion. Soon after delivery or a few days later a lower abdominal mass filling the introitus (hydrocolpos or mucocolpos). If hydrocolpos is not possible, and the newborn may present with a whitish bulging mass filling the introitus (hydrocolpos or mucocolpos). If hydrocolpos is small or not detected at birth, secretions resorb up to the end of first months of life, and the condition may be undetected until puberty. Also, if there is a low level of maternal hormones, there is no extra genital secretion and the obstruction will usually remain unnoticed until puberty.

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The hydro (metro) colpos may compress the adjacent bladder and ureters, bowel, and pelvic veins, causing problems of urinary retention, constipation, and edema or cyanosis of the lower extremities [1,9]. Secondary infection of the vaginal fluid, usually with colonic organisms, is not uncommon. Other abnormalities are published as syndromic forms of hydrometrocolpos, associated with genetic syndromes as McKusick-Kaufman syndrome or Bardet-Biedl syndrome [10-12].

At puberty, the accumulation of menstrual blood is clinically present as hemato (metro) colpos. The accumulated secretion evert the membrane producing a bluish-colored bulging mass at the introitus (mid and high transverse vaginal septa or vaginal atresia will not present with this characteristic interlabial mass) [6]. Clinical signs at the beginning of puberty may be as follows: recurrent pelvic, lower abdominal or back pain or lower abdominal midline mass [13-15]. Severe long-standing obstruction may cause urinary retention, constipation and hydrolephrosis [16]. The pressure of the vagina on the ureters crossing the pelvic brim results in hydromecephalus and hydrouretur.

Compression of the vena cava and the iliac vessels causes cyanosis, edema, and ecchymosis of the perineum, lower extremities, and abdominal wall. The rectum is less commonly involved, but constipation may complicate hydro (metro) colpos.

Rarely the reflux of endometrial tissue through the fallopian tubes (hematosalpinx) may result in secondary endometriosis [17].

Diagnosis of hydro (metro) colpos is possible by fetal ultrasound from 25th gestational week [18-20]. Hydrocolpos should be considered when an abdominopelvic cystic mass is diagnosed in a female fetus. Also, it is necessary to prenatally detect whether there are associated anomalies [21].

In cases when pelvic mass is difficult to distinguish from other abnormalities – inappropriate fetal position, if mother is obese, or oligohydramnios is present, an ultrafast MRI is suggested [22-24]. MRI shows the exact location and extension of the cystic mass, bladder or cloacal abnormalities if they exist [25]. The differential diagnosis of the pelvic mass includes saccroccygeal teratoma and anterior meningocele.

In neonates, clinical finding and pelvic ultrasound showing a dilated vagina are sufficient for diagnosis. Abdominal ultrasonography reveals a large, sonolucent midline mass displacing the bladder forward and the rectum posteriorly [9]. Associated anomalies have to be excluded. In pubertal girls MRI is preferable than CT in visualisation of other congenital abnormalities [26].

Differential diagnosis includes vaginal atresia or agenesis, transverse vaginal septum, vaginal cyst, hymenal cyst or paraurethral cyst, cloacal malformation, etc. It is important to distinguish an imperforate hymen from transverse vaginal septum because of possible influence on fertility.

Treatment of imperforate hymen can be performed in infancy, childhood, or in adolescence. Under general anesthesia hymen is incised in newborns, and after that the fluid is drained. The best solution in adolescent girls is an elliptical excision of the membrane close to the hymenal ring, with running absorbable suture along the incised edges to prevent recurrence. Slow evacuation of the retained material is suggested and a Foley catheter placed in vagina can be used for drainage. A local anaesthetic should be applied: lidocain sol injected or lidocain gel topically. Estrogen topic use is also a good idea.

Puncture and drainage of hematocolpos without a definitive surgical procedure is not recommended. The fluid is viscous and may not be adequately drain, which increases the risk of an ascending infection. Simple incision of the hymen may be associated with postoperative stenosis. Spontaneous rupture of the IH with hematocolpometra is described and hymenotomy may not be needed if adequate opening for menstrual discharge is warranted [27]. After decompression repeated ultrasonography is necessary over the next few months for confirmation of normal vaginal outflow.

A complication like acute abdomen due to bilateral hematosalpinx and unilateral rupture of the hematosalpinx has been published [28].

Over the last ten years (2003-2012) 15 females were admitted at the Pediatric Surgery Hospital in Novi Sad and treated for IH.

Four of them were neonates:

- Three were full-term neonates from unremarkable pregnancies. Primary health care paediatricians had noticed the enlarged abdomen (Figure 1A) a few days after delivery,
the diagnosis was confirmed by ultrasound and the operative treatment was successful. Mild hydronephrosis in two neonates regressed after treatment. Also vascular pressure on pelvis and distal extremities (Figure 1B) disappeared immediately after decompression. No other anomalies were detected. Two of them were diagnosed by fetal ultrasonography during the last week of pregnancy.

- One premature infant in the 30th week of gestation was born and admitted to Intensive Therapy unit for duodenal atresia. Physical examination revealed small hymenal protrusion (Figure 2). After duodenal surgery hymenotomy was performed, but parents were informed about the local setting and the necessity of observation up to the pubertal age. This has been the youngest female with hymenal incision and we shall continue with regular follow-ups.

One was the infant aged two:

- A two year female suffered recurrent urinary infection. She had a complete adhesion of the labia minora and after adhesiolysis it was expected that uroinfections would stop. Micturition urethrocystography performed for repeated infection diagnosed bilateral vesicoureteral reflux. During examination for endoscopic treatment of reflux, IH was noticed and an elliptical incision was performed during the same procedure for reflux.

Two were the prepubertal girls:

- A girl of 9 went to a urology consultant because at her birth her parents were informed that she had IH though without a complication. An incision was proposed before the age of puberty. External genitalia were normally developed but hymen consisted of translucent imperforate membrane. Two years later she started to menstruate with regular cycles.

- An amenorrheic girl of 10 had an increased leucorrhoea, and in primary care the vaginal outflow obstruction was noticed.

There were no signs of dilated vagina, external genital examination revealed membrane instead of typical hymenal appearance. During intervention a pin-point opening in the cephalad part of the hymen was noticed. We defined it as an incomplete hymenal obstruction. An elliptical excision was performed. Ten months later menarche appeared followed by irregular cycles.

Eight were the pubertal amenorrheic girls:

- They were sent to consultation for extreme constipation, urinary retention, abdominal tumor, nodi haemorrhoidales, and four of them for recurrent abdomino-pelvic pain. Genital examinations revealed the IH and hematocolpos; ultrasound confirmed the diagnosis and all girls underwent elliptical hymenal incision with running sutures successfully. Regular follow-ups and ultrasound confirmed a physiologic cycle of menstruation. No other anomalies were noticed.

**Congenital Anomalies of Fusion in Vagina Development**

The sinovaginal bulbs complete their growth by the 16th gestational week, forming the primitive vaginal plate. Canalisation of this plate is completed with forming of vagina by the 5th month [29]. Failure of canalisation might be present with a spectrum of anomalies that rarely cause symptoms during infancy but disturb the time of menstruation.

According to the American Fertility Society (AFS) classification, Mullerian duct abnormalities [30,31] manifest as a wide spectrum caused by arrest at different stages of development (lateral, horizontal or longitudinal fusion disorders), or as a result of abnormal canalisation of vaginal plate or failure of fusion (disorders of vertical or transverse fusion) [32,33].

**Transverse Vaginal Septa**

Transverse vaginal septa (TVS) may occur at any level above the hymen. It is also caused by the failure of vaginal plate canalisation. As a result high, mid or low diaphragm-like formations are present obstructing the outflow. Patients are primary amenorrheic.

Clinical presentation is rarely as hydro (metro) colpos (usually presented with low TVS) but most often as hematoo (metro) colpos. Symptoms of recurrent abdominal or pelvic pain are in the patients’ history data. Abdominal ultrasound and external genitalia examination lead to a diagnosis. A bulging membrane protruding through the vulva is not always an imperforate hymen. In some cases a transverse septum higher in the vagina is seen to protrude through a normally perforated hymen, membrane bulging through a normal hymen. It is possible that some patients with a protruding membrane are incorrectly diagnosed with IH.

When the obstruction is due to a very low vaginal septum, the accumulated secretion averts the membrane producing a bulging mass at the introitus. Mid and high transverse vaginal septa or vaginal atresia will not present with this interlabia mass [6,9]. Hydro (metro) colpos from a low membrane is usually not associated with other congenital anomalies unless the obstructed vagina is one-half of a duplicate system [34]. This is how hydro (metro) colpos differs from vaginal atresia or a mid or high transverse septum, which is almost always associated with severe and often multiple congenital anomalies of the genitourinary and gastrointestinal tract [6].
The diagnosis is usually made by physical and ultrasound examination. Surgery for a transverse vaginal septum depends on location and thickness. Excision of the septum and maintained vaginal patency are the main tasks. Sometimes there is indication for vaginoplasty. In cases of strictures repeated dilatation is required.

A follow-up examination 4-6 weeks later is recommended to ensure that the scar tissue has not caused a recurrence of obstruction.

Our experience includes three patients with low TVS:

- Neonatal period - A two-day-old female newborn with enormous abdominal distension, respiratory insufficiency due to bilateral massive pneumonia and hydromineral imbalance was admitted to the hospital. Prenatal US were not performed. A firm smooth mass occupied the lower and medium abdomen up to the costal margin. External genitalia were normal. Ultrasound examination revealed large fluid filled cystic mass extending from pelvis occupying the whole abdomen, moving the intestine cranially. At the top of the cyst a uterine like formation was noticed. Urinary tract was normal. Vaginoscopy was performed through normal hymenal aperture, and 5 mm upward a blind septum was noticed bulging into vagina. An incision and insertion of small polietilen F4 catheter was adequate to drain 600cc of odorous mucoid fluid. General condition deteriorated with life-threatening septic presentation and the neonate died five days after the operation.

- Pubertal age - Two girls of thirteen had a history of recurrent low abdomen pain lasting a few months. Ultrasound revealed a large vagina filled with menstrual blood. On examination of external genitalia a bulging but not translucent membrane obstructed the entrance of the vagina (Figure 3A). Rectal examination revealed a pelvic mass. Associated anomalies were not detected. Under general anaesthesia an elliptical excision was done through the septum formation. It was few millimetres thick and not resembled the hymen. Slowly, 300 ml from one vagina and 350 ml from the other one were evacuated. Running sutures were used in trying to prevent recurrence or stenosis. Microscopic analysis of tissue specimen revealed the possibility of a low transverse septum (Figure 3B). Both girls are now under the outpatient care of juvenile gynecologists.

Vaginal Atresia

Vagina originates from two embryonic structures: the upper part from Mullerian duct system and the lower third from the urogenital sinus. Canalisation of the vaginal canal is complete by the 20th week.

Misdevelopment presented as failure of fusion or canalisation of these two systems in vertical plane may be clinically present with a spectrum of Mullerian duct anomalies. Vaginal atresia (VA) is one of them. Missing portion of the vagina is replaced with fibrous tissue [17]. According to the American Society for Reproductive Medicine 1998. Classification, vaginal atresia is categorized as Type I [35]. Some authors believe that variants of VA, formerly called partial vaginal agenesis, are more correctly classified as variants of a transverse vaginal septum [17].

In VA the appearance of the external genitalia can be normal or vulva slightly retracted upwards. This is why this abnormality cannot be detected without a careful examination of the genitalia.

Clinical findings vary depending on the anatomy of the vaginal outlet and the changes in the upper vagina and uterus. The upper vagina becomes enormously distended when the girl starts to menstruate, usually producing a palpable abdominal mass arising from the pelvis. The Fallopian tubes can be normal, although they may be distended allowing escape of the fluid into the peritoneum. Other anomalies are occasionally seen with hydrometrocolpos. Some other combinations of structural anomalies may be present, such as Mayer-Rokitansky-Küster-Hauser syndrome, Bardet –Biedl syndrome, Frase syndrome, and Winter syndrome [36-39].

The diagnosis is usually made when symptoms of obstruction are obvious. Ultrasound, abdominal and endorectal [40], and other imaging studies confirm the physical examination. Urogram would demonstrate anterior and superior displacement of bladder and, possibly, hydrenephrosis and hydroureter. As Mullerian agenesis can be associated with other anomalies, particularly those of kidneys and skeleton [41,42], further investigations are indicated.

Therapy is directed to relieve the obstruction of the vaginal outlet and provide normal sexual life and reproductive function. Regarding the pathological problem, cultural and religious considerations, it is necessary to inform the parents about the risk of defloration. Vaginal reconstruction is required, sometimes by an abdomino-perineal approach, but stenosis and fistula formation could complicate postoperative period.

We have had only one case of VA:

- Pubertal age - A twelve-year-old amenorrheic girl was admitted as an emergency case for the symptoms and signs of acute abdomen. History taking revealed that a healthy and sport oriented girl started to suffer a lower abdominal pain six month earlier, followed by parestetic feelings on the both legs. Progressing symptoms and the lower back intensive pain after abdominal ultrasound turned attention to a large cystic mass arising from the pelvis which was suspected to be uterus didelphys in the form of hematometra (Figure 4A), and also a solitary kidney on the right (Figure 4B). CT scan confirmed the diagnosis. External genitalia appearance was normal (Figure 4C). Emergency laparoscopy revealed two large masses of hematodidelphys uterus and bilateral hematosalpinx with perforation on the left. Transabdominal drainage was performed. When general condition improved transvaginal desobstruction and vaginoplasty were performed. Treatment involved dissection through the atretic segment with pull-through anastomosis of both proximal and
distal parts. Regular dilatation was performed over the next few months, preventing the stenosis.

Conclusion

The genital tract’s expulsion function is important especially from the beginning of puberty and menstruation. The outflow obstruction may occur at different levels with variations in clinical presentation. It is also noticed that Mullerian abnormalities may be a component of a multiple malformation syndrome.

In many patients they remain undetected until puberty, when patients reach menstrual cycle and experience the retrograde menstruation. Most patients have the history of amenorrhea, recurrent lower abdominal cyclical pain, or lower abdominal midline mass. Severe long-standing obstruction may cause urinary retention, hydronephrosis and constipation. Recurrent urinary infection is also a common clinical condition in differential diagnostics of vaginal outlet obstruction.

Examination of the newborn and inspection of genitalia are still essential. Suspicion of absence of vaginal outlet can arise during this simple evaluation which may indicate further investigation. Ultrasound is essential in diagnostics but in some cases CT scan or MRI is necessary prior to surgery. Surgery of vaginal outlet obstruction depends on local anatomy. Simple incision / excision or vaginal reconstruction can be performed. Postoperative follow-up is necessary until the normal menstrual cycle is established.

References