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Embryology

Elixir Embryology 102 (2017) 44498-44501

ISSN: 2229-712X

# "COMMON VAGINAL ANOMALIES" EMBRYOLOGICAL BASIS AND ITS CLINICAL IMPORTANCE

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**ARTICLE INFO**

**Article history:**

Received: 9 December 2016;

Received in revised form:

13 January 2017;

Accepted: 26 January 2017;

**Keywords**

Paramesonephric ducts,
Mesonephros,
Hematometrocolposatresis,
Mullerian,
Didelphys.

**ABSTRACT**

The embryologic growth of the vagina is still uncertain. The urogenital sinus and the mesonephric (wolffian) and paramesonephric ducts possibly all play a part in the development of the vagina. Duplication, agenesis, mesonephric duct remnants, and hymen abnormalities are amongst the more common congenital anomalies of the vagina. Due to close developmental correlation between the genital and the urinary tracts, relations of anomalies in both systems are common. Mullerian malformations are normally associated with abnormalities of the renal and axial skeletal systems, and they are frequently the first encountered when patients are originally inspected for related conditions.



## Introduction

Congenital abnormalities of the female genital tract are defined as deviations from normal anatomy resulting from embryonic maldevelopment of the Mullerian or paramesonephric ducts. Congenital malformations of the female genital tract may be the result of a clear disorder in one stage of embryonic development, or more than one stage of normal development[1]. Vaginal abnormalities include vaginal agenesis, vaginal atresia, Mullerian aplasia, transverse vaginal septa and its associated anomalies[2]. Vaginal agenesis is a condition that progresses before birth, in which the muscular canal (vagina) to your uterus fails to develop completely. When this happens, other problems also may appear. For example, you may have a small uterus or, more commonly, no uterus at all.Vaginal atresia is a condition in which the vagina is abnormally closed or absent[1,2]. The main causes can either be complete vaginal hypoplasia, or a vaginal obstruction, frequently caused by an imperforate hymen or, unusually, a transverse vaginal septum. Vaginal atresia is a congenital defect causing in uterovaginal outflow tract obstruction[2,3]. It occurs when the caudal portion of the vagina, contributed by the urogenital sinus, fails to form. This caudal portion of the vagina is replaced with fibrous tissue. Mullerian aplasia is a disorder that affects the reproductive system in females[4]. It is caused by abnormal development of the Mullerian ducts, which are arrangements in the embryo that develop into the uterus, fallopian tubes, cervix, and the upper part of the vagina. Individuals with Mullerian aplasia typically have an underdeveloped or absent uterus and may also have anomalies of other reproductive organs[5]. Women with this condition have normal female external genitalia, and they have pubic hair and develop breast normally at puberty, and they do not begin menstruation by age 16 (primary amenorrhea) and will never have a menstrual period[4,5]. Affected women are unable to have children (infertile).

Women with Mullerian aplasia have higher-than-normal levels of male sex hormones called androgens in their blood (hyperandrogenism), which can cause acne and excessive growth of facial hair (facial hirsutism). Kidney abnormalities may also be present in some affected individuals[5,6]. A transverse vaginal septum is a condition in which a wall of tissue running horizontally through the vagina. It is a congenital disorder, present at birth. In associated anomalies the urethra can open into the vaginal wall or the vagina into a persistent urogenital sinus. Associated rectal abnormalities include vaginorectal fistula, vulvovaginal anus, recto-sigmoidal fistula[6].

## Incidence

The incidence of vaginal anomalies is slightly higher at 3-6%. In general, women with regular abortions have an incidence of 5-10%, with the highest incidence of Mullerian defects occurring in patients having third-trimester miscarriages[7]. The most commonly reported vaginal abnormalities are transverse septum, vaginal atresia, vaginal agenesis and mullerian aplasia. The exact distribution depends on the study and on the geographic location. The prevalence of mullerian duct anomalies also differs , with reports ranging from 0.16-10%[7,8].

## Ontogenesis of the normal fate of Mullerian duct

In females the paramesonephric (Mullerian) ducts arise from the mesoderm lateral to the mesonephric ducts in the seventh week as pivotal invaginations of the coelomic epithelium on the upper pole of each mesonephros[9]. The paired paramesonephric and mesonephric ducts represent the indifferent stage of the foetal internal genital canal systems. The paramesonephric ducts are the precursors of the uterus, fallopian tubes, cervix and upper vagina. The paramesonephric ducts grow caudally, coursing lateral to the urogenital ridges. In the eighth week the paired paramesonephric ducts lie medial to the mesonephric ducts[10]. The paramesonephric ducts fuse to form a confluence.

This process is referred to as Mullerian organogenesis and this represents the initial stage in the development of the upper two-thirds of the vagina, the cervix, uterus and both fallopian tubes[10,11]. The cranial end of the fused ducts gives origin to the future uterus which contains mesoderm which forms the uterine endometrium and myometrium. The unfused cranial ends of the paramesonephric duct, a funnel shaped configuration and remain open to the future peritoneal cavity as the fimbriae portions of the fallopian tubes. The caudal end of the fused ducts will form the upper two-thirds of the vagina[11].

During seventh and ninth weeks when the lower segments of the paramesonephric ducts fuses, the lateral fusion of paramesonephric ducts occurs[12]. At this stage a midline septum is present in the uterine cavity, this usually regresses at around 20 weeks but can persist. Vertical fusion occurs in the eighth week when the lower most fused paramesonephric ducts fuse with the ascending endoderm of the sinovaginal bulb[12,13]. The lower third of the vagina is formed as the sinovaginal node (bulb) canalizes. The sinovaginal node insets into the urogenital sinus at Muller's tubercle. The hymen, a membrane separating the vagina from the urogenital sinus develops and is normally perforated by birth[13].

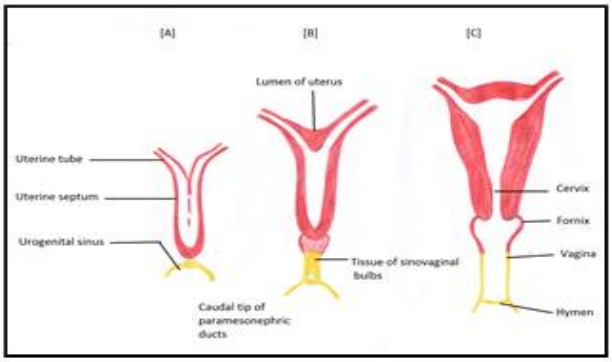


**Fig1. The schematic diagram shows the normal development of vagina [A] regression of uterine septum. [B] Canalization of sinovaginal bulb. [C] Normal vagina.**

**Ontogenesis of the vaginal anomalies**

During normal developmental stages when interruptions occur, mullerian defects occur and are frequently grouped according to the failed developmental mechanism that gives rise to a given malformation [14]. This includes agenesis or hypoplasia, lateral fusion defects, vertical fusion defects, transverse septum. Actually, many Mullerian duct anomalies often exist. The most common Mullerian duct defects involve the vagina and the uterus; these anomalies are the most commonly corrected by surgically [15]. Vaginal agenesis is categorized by an absence or hypoplasia of the uterus, proximal vagina, and, in some cases, the fallopian tubes [14, 15].

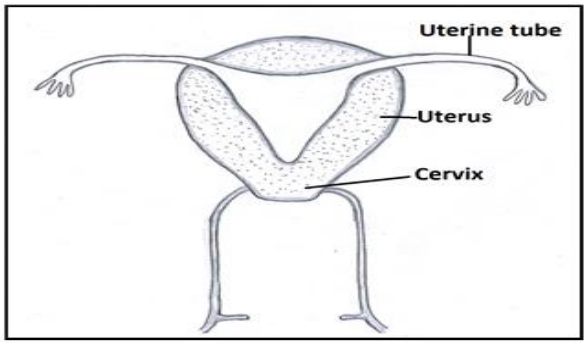


**Fig 2. The schematic diagram shows the absence of vagina (Vaginal atresia).**

Vaginal agenesis is the result of developmental failure of the sinovaginal bulbs. Without the sinovaginal bulbs, the vaginal plate cannot form. In this condition uterus is absent because the UVP induces differentiation of the sinovaginal bulbs [15]. Incomplete vaginal agenesis with a normal upper genital tract is uncommon and must be distinguished from vaginal atresia. Vaginal atresia is due to an interruption in UGS development and is usually allied with normal Mullerian-derived structures [16].

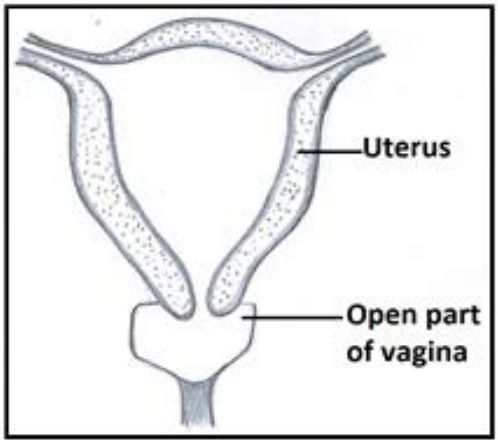


**Fig 3. In this schematic diagram shows the open part of vagina indicates the closed vagina (vaginal agenesis).**

Lateral fusion defects are divided into symmetric and asymmetric groups and are further divided into obstructive or non-obstructive groups [16]. These defects occur by means of arrests in different stages of Mullerian duct development. In general, the arrested stages include partial fusion of the caudal Mullerian ducts, failed septum resorption, and imperfect development of all or part of a duct [17]. Asymmetric, disruptive lateral fusion defects of the Mullerian system are frequently related with unilateral mesonephric agenesis and evident as renal agenesis, ureter agenesis, or both. Müllerian aplasia can be partial or complete [16, 17]. Partial Mullerian aplasia is characterized by a normal uterus and small vaginal pouch distal to the cervix. Complete mullerian aplasia is the most common and it is characterized by congenital absence of the vagina and the uterus. The fallopian tubes are normal, and the ovaries have normal endocrine and oocyte function [18]. Horizontal vaginal septa are included in this classification because they occur as consequence of defective lateral fusion; however, other horizontal septa arise from mesodermal hyper proliferation or persistence of epithelium during canalization.

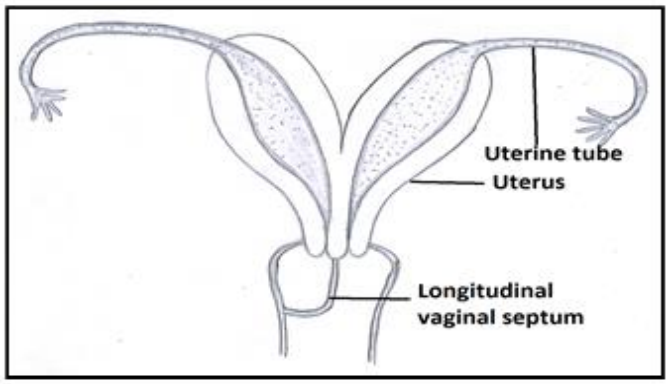


**Fig 4. The schematic diagram shows the development of longitudinal vaginal septum.**

Isolated horizontal vaginal septa are usually clinically benign, though obstacle can occur when septa are associated with other Mullerian anomalies, and surgical excision may be

required [19, 20]. Disorders of vertical (transverse) fusion result from abnormal canalization of the vaginal plate and, in some cases, the UVP and the sinovaginal bulbs fails to fuse [20]. These disorders can result in the formation of a transverse vaginal septum (TVS), an imperforate hymen, and, vaginal atresia. The TVS is subdivided according to whether the defect is complete or partial [21]. TVS can be supplemented by urinary tract anomalies. A partial septum may occur in females exposed to DES. Imperforate hymen and vaginal atresia result from structural defects involving derivatives of the UGS [22].

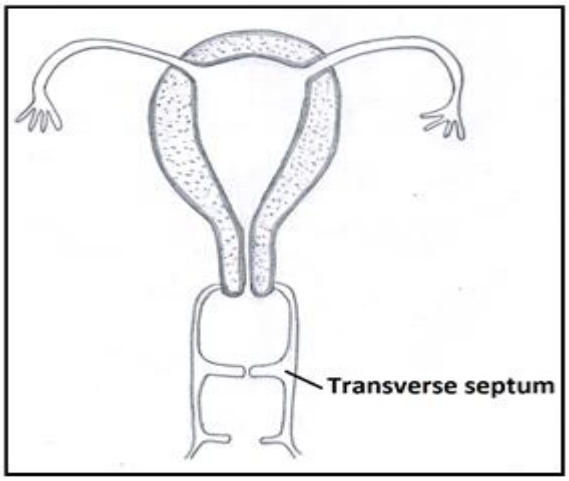


**Fig 5. The schematic diagram shows the development of transverse septum.**

**Discussion**

As the paramesonephric (Mullerian) ducts grow caudal, they reach the urogenital sinus by about the ninth week and fuse with it to form an elevation known as the Mullerian tubercle, with the openings of the paramesonephric ducts on either side of it. A ribbon of epithelium replaces the uterovaginal canal and is the precursor of the vagina [23]. Between 16th and 20th weeks the vagina is formed by the development of lacunas; complete canalization later occurs to form the vaginal lumen .The principal congenital anomalies of the vagina include, longitudinal septum, Transverse septum, vaginal agenesis, and vaginal atresia and Mesonephric remnants [24]. The longitudinal septum's produces the "double vagina" with the appearance of a double-barreled shotgun. Such septum may be complete or incomplete [25]. They are frequently associated with uterus didelphys, and all arrangements have been encountered, including the presence of a normal uterus and cervix [26]. Due to retention of mucus or blood, one cervix can be blocked off. Because the external genitalia typically appear normal, such septum is frequently not diagnosed unless painful coitus or labor dystocia occurs [26, 27]. Simple incision with suitable ligation of bleeding points is required. Fistula formation or vaginal stenosis may result from incautious or extensive surgery. Longitudinal septum occurs almost twice as frequently as transverse septum. Transverse septums vary from complete occlusion to a mild constriction [28]. And there is a small opening allowing blood to drain. There is no external sign of the blockage in the bulging membrane associated with imperforate hymen. In nearly all patients, the internal organs are normal and pregnancy is frequent. Transverse

Septum represents failure of complete canalization of the vaginal epithelial mass [28]. The stenosis is caused by a narrowing of fibromuscular bond. If menstruation and coitus occur without trouble, the condition may not be spotted until pelvic examination reveals its existence .

The symptoms depend on the presence of suitable uterine drainage [29, 30]. A mass may develop in the lower abdomen, due to hematometrocolposatresis in case of complete vaginal atresia. Pelvic abscesses may develop in such entrapped blood. Dystocia sometimes caused by vaginal atresia and make cesarean section the safest method of delivery [31]. Depending upon the degree of stenosis and the firmness of the constricting band, treatment is given. No treatment may be required, or two or three longitudinal incisions may suit. Attempts at complete excision of an annular segment of vaginal wall may result in postoperative scarring or fistula formation [32, 33]. For amenorrhea agenesis of the vagina may be detected on pelvic examination. Absence of a vagina is always associated with absence of a hymen. Agenesis of the vagina is usually misdiagnosed as imperforate hymen. In vaginal agenesis there is an absent or rudimentary uterus; ovarian agenesis may also be present [33]. Nearly all patients without a vagina have associated anomalies of the urinary tract, including renal agenesis, ectopic pelvic kidney, and ureteral anomalies. Lower vaginal atresia is when the lower portion of vagina fails to develop appropriately [34, 35]. The lower portion of the vagina is replaced by fibrous tissue, causing blockage. These are the consequence of failure of development of the epithelial vaginal mass or from failure of development of the urogenital sinus [36]. A congenital imperforated hymen is a result of failure of completion of the canalization or cavitations of the epithelial plug that fills the vagina. The condition is usually exposed at puberty, when the patient develops a lower abdominal pain, abdominal mass and bulging vaginal mass full of mucus or blood [35, 36]. A related condition is a microperforate hymen, in which no hymeneal opening can be found but a tiny opening is present which allows passage of menstrual blood [37].

**Conclusion**

Vaginal disorder one of the Müllerian anomalies are a morphologically different group of developmental disorders that involve the internal female reproductive tract. An accurate diagnosis is essential for planning treatment and management schemes. And it is diagnosed by the physical examination and ultrasonography. The surgical approach for correction of Mullerian duct anomalies is specific to the type of malformation and may vary in a specific group [38].

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