

Research Letter

A mullerian anomaly “without classification”: Septate uterus with double cervix and longitudinal vaginal septum

Nilufer Yigit Celik, Baris Mulayim*

Department of Obstetrics and Gynecology, Alanya Medical and Research Center, Baskent University, Alanya, Antalya, Turkey

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The incidence of congenital uterine anomalies in the general population is estimated to be 0.001–10% [1]. The most commonly seen anomalies are septate and arcuate uteri, which represent 75% of all mullerian anomalies. The majority of mullerian duct anomalies are considered to be sporadic or multifactorial in nature; however, polygenic and genetic patterns of inheritance have been described [2,3].

There are two theories to explain the development of mullerian duct anomalies, the classical unidirectional regression theory, and an alternative bidirectional theory in which it is hypothesized that the process proceeds simultaneously in both the cranial and the caudal directions. Anomalies such as a complete septum with a duplicated cervix or an isolated vertical upper vaginal septum could only be explained by the second theory [4,5]. We report the case of a woman with a complete uterine septum, and a double cervix with a longitudinal vaginal septum, and search the available literature for similar cases.

A 28-year-old nulligravida woman was admitted to our hospital due to being unable to conceive. She had been married for 2 years and had never used any contraceptive methods. She had regular menses but had dysmenorrhea and sometimes also dyspareunia. A longitudinal vaginal septum from the midportion of the vagina to the cervix, and the presence of two separate cervixes were identified following a speculum examination. Transvaginal ultrasound showed a normal uterine contour, then hysterosalpingography was performed. When a contrast media was injected into the right cervix, the uterine cavity was filled, however, the fallopian tubes were not. When the contrast media was then injected into the left cervix and uterine cavity, the left fallopian tube and pelvic cavity filled with contrast media, but the right fallopian tube was not filled with contrast media. Following

this, magnetic resonance imaging revealed a single uterus with two endometrial cavities and two cervixes associated with the longitudinal vaginal septum (Fig. 1A). No additional genitourinary abnormalities were established. Her Day 3 follicle-stimulating and estradiol values, and her husband's semen analysis were also normal.

The patient underwent both vaginal and endoscopic surgery. The vaginal septum was excised using scissors. Hysteroscopy revealed a complete uterine septum, which was resected. Following hysteroscopy, laparoscopy was performed, which confirmed that there was a normal uterine fundus without indentation, and normal ovaries and fallopian tubes (Fig. 1B). Methylene blue was injected through both cervixes sequentially, during laparoscopy. When the methylene blue was injected through the right cervix first, it filled the left fallopian tube and pelvic cavity, but not the right fallopian tube. However, when methylene blue was injected through the left cervix it filled the left fallopian tube and pelvic cavity, but not contralaterally. Following uterine septum resection, one uterine cavity was established, making it unnecessary to inject methylene blue into both left and right cervixes. After surgery, timed intercourse was recommended due to normal ovarian function, semen analysis and tubal patency and she has been trying to conceive expectantly for 7 months.

This unique mullerian anomaly—consisting of a septate uterus with cervical duplication and longitudinal vaginal septum—was first described by McBean and Brumsted [6]. This anomaly differs from the classical embryological definition of mullerian anomalies. Traditionally it is believed that the uterus and vagina are formed embryologically by three processes, lateral fusion, vertical fusion, and resorption. Abnormalities of the uterus and vagina are believed to be caused by disruption of one of the above processes, resulting in failure of either fusion or canalization. Failure for the mullerian ducts to fuse can result in uterus didelphys (double uterus) or septate uterus. The incidence of uterus didelphys in fertile women has been calculated to be about 0.16% [7]. Septate uteri account for 35% of structural uterine anomalies

* Corresponding author. Baskent Universitesi, Alanya Hastanesi, Kadın Hastalıkları ve Doğum Bölümü, Alanya, Antalya, Turkey.

E-mail address: brsmlym@yahoo.com (B. Mulayim).

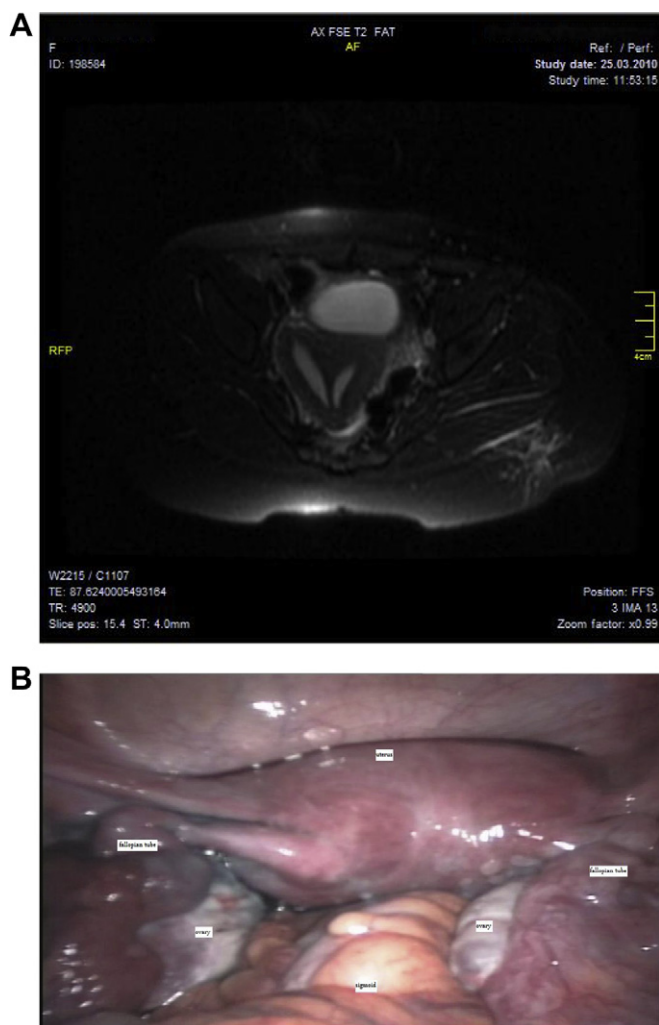


Fig. 1. (A) Magnetic resonance imaging revealing a single uterus with two endometrial cavities. (B) Laparoscopy showing a normal uterine fundus without indentation, and normal ovaries and fallopian tubes.

and is the most common, with a prevalence of 1–2% in the general population. The true incidence of triad, septate uterus with cervical duplication and a longitudinal vaginal septum, is not known [8,9].

There have been cases of septate uterus with double cervix and vaginal septum or normal uterus with double cervix and vaginal septum reported in the literature. However, there is no uniform theory to explain all these different mullerian anomalies. In 1962, Crosby and Hill described the theory of linear caudal to cephalad mullerian fusion [4]. According to their theory, during the 11th to 13th weeks of embryologic life, mullerian ducts fuse from the caudal to cephalad points. Septal resorption in either direction immediately follows the fusion. However, this theory

did not hold the cases of double cervix and vaginal septum that was the result of failed caudal fusion, and the cases of septate uterus and double cervix and vaginal septum that was the result of failed caudal fusion and failure of septal resorption with normal cephalad fusion [10].

Another hypothesis was described by Musset et al in 1967 [5]. This hypothesis proposed that mullerian duct fusion begins at isthmic level, then proceeds bidirectionally. This hypothesis is consistent with the cases of double cervix and vagina, and normal uterus (failed caudal fusion of the mullerian duct, beginning at the uterine isthmus, and normal septal resorption after normal fusion in the cranial direction).

In the literature, another spectacular case was reported by Moawad et al in 2009. This case demonstrated the coexistence of two rare gynecologic anomalies in a 15-year-old girl: uterus didelphys and longitudinal vaginal septum coincident with an obstructive transverse vaginal septum [7].

The literature includes a wide spectrum of mullerian anomalies and patients present with a variety of symptoms. Some cases have been misdiagnosed and maybe some have never been diagnosed.

In conclusion, especially for unique mullerian anomalies, known incidence does not reflect the true incidence. In addition, the proposed embryological theories do not explain the development of all these peculiar anomalies. This case is presented in an attempt to alert all gynecologists of the need to establish the true incidence of this anomaly.

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