

A CASE OF UTERINE DIDELPHIS WITH CERVICAL ATRESIA: A CASE REPORT

Eskinder Kebede, MD¹, Yitbarek Fantahun, MD, MPH¹, Hamelmal Abate, MD¹

ABSTRACT

Congenital uterine malformations are deviations from normal anatomy resulting due to defective fusion of Mullerian ducts or the paramesonephric ducts in the developing embryo.

The prevalence of female genital tract anomalies is 4%-7% in general population and up to 8%-10% in women who have recurrent pregnancy loss.¹ The type and degree of anatomical distortion has associated health implications that may include, cyclical abdominal pain, reproductive failure, obstructed menses and inability to engage in sexual intercourse.²

Uterus didelphys (double uterus) is a developmental abnormality that results from the failure of fusion of the Mullerian ducts leading to separate uterine cavities and 2 cervixes 1. It is a rare anomaly accounting for 5% of all uterine anomalies.³ Cervical agenesis is a rare Mullerian anomaly with an incidence of 1 in 80,000 females.⁴ It represents 3% of all uterine anomalies. It is rarely associated with a functioning uterus (4.8%). Cervical agenesis is often associated with vaginal atresia (less than 50%). It is important to classify these anomalies for easy diagnosis and plan appropriate preoperative treatment.

KEYWORDS: Case report, Cervical atresia, Mullerian anomaly, Uterine Didelphis.

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¹ Department of Obstetrics and Gynecology, School of Medicine, Addis Ababa University, Addis Ababa, Ethiopia

BACKGROUND

Mullerian duct anomalies (MDAs) are congenital defects of the female genital system that arise from embryological mal development of the Mullerian ducts. These abnormalities result from either failure of development, fusion, canalization, or reabsorption, which normally occurs between 6 and 22 weeks in utero. As estimated by most resources, the incidence of these abnormalities ranges between 0.5 to 5.0% in the general population.⁵

Most women with a didelphys uterus are asymptomatic, but some present with dyspareunia or dysmenorrhea in the presence of a varying degree of longitudinal vaginal septum. Rarely, hematocolpos, hematometocolpos, and renal anomalies are reported in association with didelphys uterus. Despite some of these complications, there are many cases of women with a didelphys uterus that did not exhibit any reproductive or gestational challenges. When classifying these anomalies solely based on abnormal development, four major types are apparent. The most recent and widely used classification systems for the different types of Mullerian duct abnormalities were created by Buttram Jr. and Gibbons (1979) and the American Fertility Society (1988)⁶

Cervical agenesis is a rare Mullerian anomaly with an incidence of 1 in 80,000 females. It represents 3% of all uterine anomalies. It is rarely associated with a functioning uterus (4.8%).⁽⁴⁾ MRI is the investigation of choice for evaluation of Mullerian duct anomaly due to its high accuracy and detailed delineation of uterovaginal anatomy. MRI has a reported accuracy of up to 100% in the evaluation of mullerian duct anomalies.⁷

CASE PRESENTATION

A 20 years-old Ethiopian female who is the first child for her family, has a younger sister who is 16 years old, has cyclic menses since 13. She has no family history of similar illness. She was referred to the outpatient gynecology clinic of Addis Ababa University Hospital in March 2016 with a

history of primary amenorrhea and severe lower abdominal pain and vomiting occurring at regular intervals over a period of 5 years. The medical and surgical history was normal. Physical examination revealed normal breast development and other sexual characters. On a genital examination a normal eschuteon was observed and the hymen was intact, with a normal perforations. The vagina was 9 cm in length as measured by trans hymenal catheter. The abdominal pelvic ultrasound showed duplicated uterine fundus, Didelphys uterus. The right uterus measures 6.9 cm in length and has 1cm fluid collection and the left uterus measures 7.4 cm in length, with minimal hematometra. Magnetic resonance imaging (MRI) revealed a double uterus with widely separated horns, and a rudimentary right horn and cervical atresia. Both kidneys were normal sized and located in their usual place.



Figure 1. Ultrasonographic image of the separated uterine horns. Notice the minimal endometrial collection in both cavities.

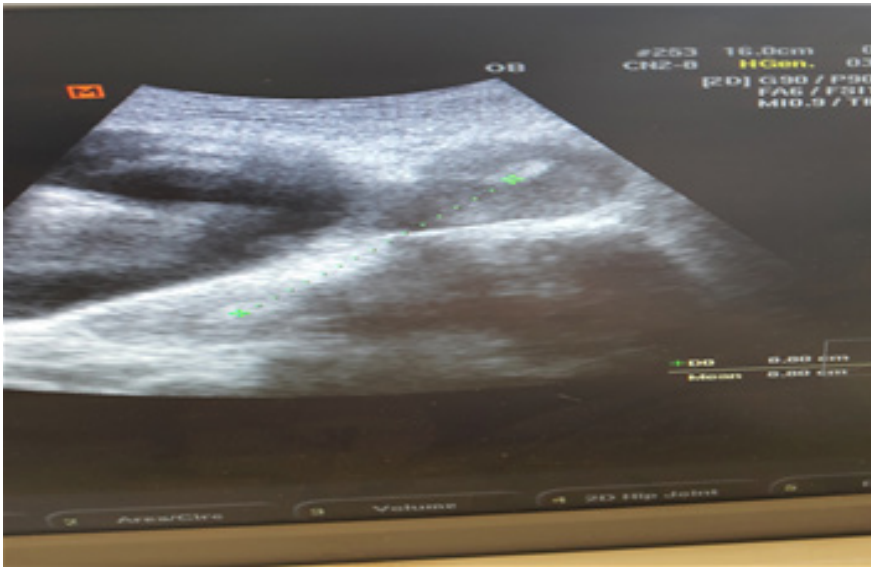


Figure 2. Ultrasonographic image showing the widely separated uterine horns.

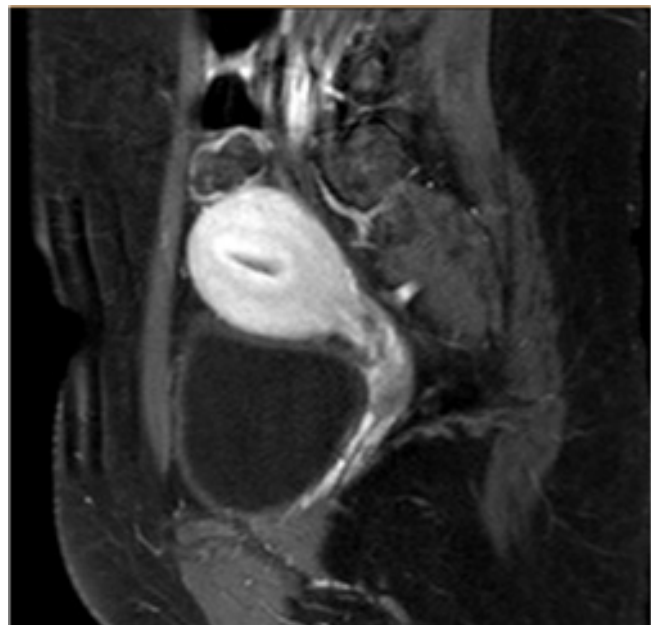
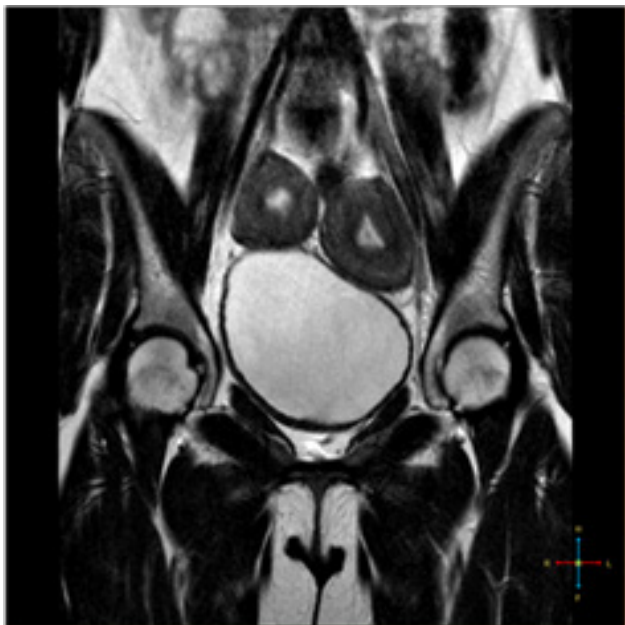
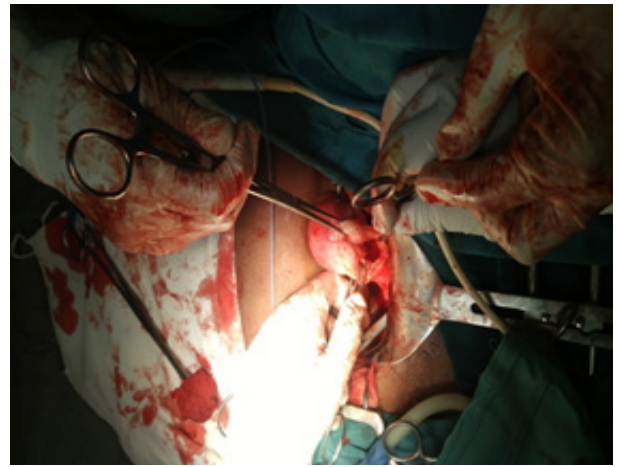


Figure 3. Magnetic resonance imaging showing a uterine didelphys with a rudimentary right horn. Notice that the right hemi-uterus leads to an atretic cord like cervix and vagina

After written informed consent was taken, the patient was taken to the operating theatre and the abdominal cavity was entered via midline infra-umbilical incision. Intraoperative there was double uterus with their tubes and ovaries which were healthy looking bilaterally (Fig 2).

Surgical therapy included resection of rudimentary horn and a trial of creation of a patent cervix by

making a midline vertical cervical and upper vaginal incision with a subsequent trial of canalization using a metallic probe but was difficult to place a stent because of sever atresia and cord like long cervix without canal. Finally hysterectomy was done.



DISCUSSION

This case report, we discuss a rare case of didelphys uterus and cervical atresia in a woman with a history of primary amenorrhea.

A didelphys uterus is characterized by complete failure of the Mullerian ducts to fuse leading to separate uterine cavities and two cervixes. A longitudinal vaginal septum may also present in most cases. Initial suspicion of the condition followed by the diagnosis usually begins with a routine speculum exam where visualization of anatomical abnormalities warrants further investigation. Further, because the Mullerian ducts develop often in association with Wolffian ducts, abnormalities of the kidneys may be found in conjunction with uterine abnormalities.

Uterine cervix agenesis is an extremely rare congenital anomaly and it occurs in 1 in 80,000 to 100,000 births (Sugunuma et al 2002). Patients affected by this rare, “non communicating”, abnormality have a functional uterus, but due to lack of cervix they get primary amenorrhea with cyclic pelvic pain, due to hematometra⁸.

Congenital agenesis or dysgenesis of the uterine cervix is a class IB in the American Fertility Society classification system (1998). The presence of a normal uterine corpus is a challenge for the clinician because a successful surgical repair could restore normal menses and potentially preserve the patient’s fertility. However, current opinion in the literature considers complete agenesis to be the most

difficult anatomic form of cervico-uterine anomaly to correct. In this form, a total hysterectomy is recommended, because of the high incidence of complications or failure when attempting surgical correction.⁹

A didelphys uterus remains a very rare Mullerian duct anomaly in comparison to other anomalies described in the Buttram and Gibbons classification. Most women with a didelphys uterus are asymptomatic, but may present with dyspareunia or dysmenorrhea in the presence of a thick, sometimes obstructing, vaginal septum. This obstructing vaginal septum can lead to hematocolpos/hematometocolpos and thus present as chronic abdominal pain as well. Rarely, genital neoplasms and endometriosis are reported in association with cases of didelphys uterus.

The fertility of women with untreated didelphys uterus has been shown by some sources to be better than those with other Mullerian duct abnormalities but still less than women with normal uterine anatomy. There is also an increased risk of spontaneous abortion, fetal growth retardation, and prematurity with an estimated 45% (or lower) chance of carrying a pregnancy to term in comparison to a normal uterus, which is similar to that of a unicornuate uterus. This indicates poor reproductive performance, but still not as poor as a septate or bicornuate uterus which are more common amongst the MDAs.

A didelphys uterus has been shown in many case reports to occur as a part of a syndrome, more specifically called, Herlyn-Werner-Wunderlich (HWW) syndrome, also known as obstructed hemivagina and ipsilateral renal anomaly (OHVIRA). It is a very rare congenital anomaly of the urogenital tract involving Mullerian ducts and Wolffian structures, and it is characterized by the triad of didelphys uterus, obstructed hemivagina, and ipsilateral renal agenesis. This condition can cause hematometrocolpos or hematocolpos on the side of the obstructed hemivagina which produces a mass effect with subsequent lower abdominal pain. Most cases present after menarche as intense lower abdominal pain and/or a protruding mass over the vaginal introitus. A preliminary pelvic Ultrasound is done followed by an MRI to confirm the diagnosis. The uterine cervix provides a conduit for menstrual flow, a barrier to infection from vaginal microflora, maintenance of an intrauterine pregnancy, and mucus for sperm transfer. Atresia (dysgenesis) of the cervix may result from local segmental atrophy. Buttram has suggested a classification for "muillerian"agenesis or hypoplasia. Cervical anomalies are designated class IB. There is a consensus in the international literature that hysterectomy is the procedure of choice in a patient with cervical agenesis. Alternatively, when cervical dysgenesis is noted, reconstruction of the cervix may be warranted. The goals of reconstructive surgery for cervical malformations are to provide a conduit for menstruation to relieve pain and preserve reproductive potential. The goals are usually achieved when there is substance to the cervix. Pregnancy has been documented after cervical reconstruction when cervical stroma is substantial. Rock et al(1995) clearly defined the different anatomic findings that they had encountered in their study.⁹ They classified anatomic forms of their 21 cases into 4 categories: cervical agenesis , cervical fragmented , cervical cord and cervical obstruction . Patients with cervical dysgenesis may have one of three anatomical variations.

They performed reconstructive surgery in the clearly defined categories, yielded differing prognoses their⁴ attempts at surgical correction of cervical agenesis and fragmented dysgenetic groups, invariably failed requiring reoperative and hysterectomy within 6 months. Their 5 attempts at surgical correction of cervical cord and obstruction dysgenetic groups, lead to a⁴ success with one pregnancy. Patients with atresia or cervical fragmentation are not usually candidates for canalization. Patients with either cervical obstruction or a fibrous cord may reasonably be considered for reconstruction.

CONCLUSION

In our case as both uterus were suspended high in pelvis. Trials of canalization using a metallic probe was difficult to place a stent because of sever atresia and cord like long cervix without canal. With no cervix and no connection between the suspended uterus and vagina, there was no possibility of creating a utero-vaginal anastomosis, hence hysterectomy was done

DECLARATION:

Ethical approval and consent to participate:

Informed written consent was obtained from the patient for treatment.

Consent to publish: Consent was taken for the publication of the case and the accompanying images.

Availability of data and materials: NA

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CORRESPONDING AUTHOR

Yitbarek Fantahun, MD, MPH

Department of Obstetrics and Gynecology, School
of Medicine, Addis Ababa University, Addis Ababa,
Ethiopia

Email: yitbarekobgyn@gmail.com/

Yitbarek.fantahun@aau.edu.et

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