CASE REPORT

Double Cervix and Vagina with Septate Uterus: An Uncommon Müllerian Malformation

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Precis

A 20 year old nulligravida presents with an unusual müllerian anomaly which is elucidated by MRI and MR-based three dimensional reconstruction, and supports an alternative müllerian development theory.

Abstract

**Background:** The case of an unusual müllerian anomaly is described. **Case:** A 20-year-old nulligravid female was found to have a septate uterus with double...
cervix and a vagina with partial longitudinal vaginal septum. The anomaly presented symptomatically, was suggested by clinical examination, and confirmed by Magnetic Resonance Imaging (MRI) and intra-operative visualization. Visualization of the anomaly was enhanced by MR-based three dimensional (3D) reconstruction techniques. Description of double cervix and vagina in the setting of an anatomically normal or simple septate uterus is uncommon in the literature. **Conclusion:** This anomaly is inconsistent with the generally accepted understanding of müllerian development but fits with an alternative hypothesis. MRI and MR-based 3D reconstruction was useful in aiding visualization of the anomaly.

**Key words:** three dimensional reconstruction, müllerian anomalies, didelphys, septate uterus, magnetic resonance imaging

**Introduction**

Müllerian anomalies represent a vast array of structural abnormalities resulting from the improper development and fusion of the embryological müllerian ducts. The precise incidence of these abnormalities is unclear but is generally considered to occur in less than 5% of women. Use of improved imaging modalities, such as vaginal probe ultrasound and magnetic resonance imaging, has helped clinicians to better identify and characterize such anomalies.

A number of different schemes for classifying müllerian anomalies exist (1,3). This report of complete uterine septum with cervical duplication and longitudinal vaginal septum seems to be at odds with the common classification system (1,3), and may support an alternative hypothesis for müllerian derivation (4).

Conventional MRI was used to help classify the anomaly, and 3D reconstruction techniques were applied to aid visualization of the actual anatomy.

**Case Report**

A 20-year-old Colombian nulligravida presented with complaints of pelvic pain and dyspareunia. She reported menarche at age 14, with dysmenorrhea. She denied difficulty with tampon use except for leakage of menstrual flow despite vaginal placement of the tampon.

Gynecological examination revealed a phenotypic female, Tanner stage V, with normal external genitalia, a longitudinal vaginal septum in the upper half of the
vagina and two normal appearing cervices. The uterine contour was smooth and anteflexed without evidence of masses. For further evaluation, the patient underwent MRI studies of the pelvis. Conventional supine MRI studies were performed as follows: T2-weighted axial source images were obtained using a 1.5T magnet (General Electric Medical Systems, Milwaukee, WI) and a torso phased array coil wrapped around the pelvis. The following imaging parameters were employed: TR=4200msec, TE(eff) =108msec, 128 phase encodes, 24cm field of view, 3mm slice thickness, no gap. This study was interpreted by the coauthor (JRF), a senior radiologist who is experienced with müllerian anomalies. It revealed the longitudinal vaginal septum, dual cervices and a single uterus with two endometrial cavities separated by a complete septum. The MR image of a septate uterus with 2 cavities is shown in Fig 1. The left ovary was slightly elongated (4cm) but uniform in appearance and the right ovary was normal. The kidneys were noted to be in orthotopic position. A 3D reconstruction of the MRI dataset was then created (Fig 2).

The patient was taken to the OR for laparoscopic evaluation of pelvic pain and resection of the vaginal septum under general anesthesia. The patient had not yet attempted pregnancy and desired therapy for her dyspareunia and pelvic pain. Accordingly, she declined resection of the septum, preferring instead to await symptoms (i.e., infertility or recurrent pregnancy loss) before agreeing to resection of the uterine septum. Laparoscopy revealed normally placed fallopian tubes, two ureters in normal position and no evidence of endometriosis or adhesions to explain her pelvic pain. The uterus was noted to have a slight concavity centrally at the fundus. Pressure applied with a blunt probe suggested dense myometrium beneath this concavity (Fig 3). Both ovaries were mildly elongated and ovoid in appearance but otherwise normal. Vaginal exam confirmed the presence of a longitudinal vaginal septum in the upper half of the vaginal cavity. After resection of this fairly dense septum, two normal appearing cervices were identified (Fig 4). The patient's postoperative course was uneventful.

Discussion

This unusual müllerian anomaly of complete uterine septum, dual cervices and longitudinal vaginal septum does not fit in the commonly used classification system of anomalies suggested by Buttram (1). Review of the literature reveals this to be an uncommonly reported anomaly (2). Three cases of cervical and vaginal duplication with a normal uterine cavity have also been described, although in one case, one cervical canal ended blindly and did not communicate with the uterine cavity. These anomalies do not fit into the existing classification systems and are inconsistent with the theory of linear
caudal to cephalad müllerian fusion as described by Crosby and Hill in 1962 (3). Specifically, this theory holds that uterine development results from the fusion of the müllerian ducts during the 11-13th weeks of embryonic life, beginning at the caudal-most aspect known as the müllerian tubercle and proceeding in a cranial direction. Septal resorption was thought to follow shortly thereafter, beginning at any point of fusion and moving in either or both directions. The case reported here cannot be explained by this theory, because a dual vagina/cervix complex suggests failure of caudal fusion while a septate uterus indicates normal cephalad fusion with failure of septal resorption.

The present case does in fact fit better into the alternative hypothesis suggested by Musset et al. (4) and advocated by McBean and Brumsted (2) when they described another similar anomaly. Musset proposed an alternative, three-stage process in which the medial aspects of the müllerian ducts begin to fuse in the middle and proceed in both the cephalad and caudal directions simultaneously. This is then followed by rapid cellular proliferation between the ducts, forming the uterine corpus and cervix, and septal resorption, all of which occur in both directions simultaneously. According to this theory, the dual vagina/cervix complex could be explained by failed fusion of the müllerian ducts in the caudal direction beginning at the uterine isthmus. The septate uterus and vagina could then be explained by complete failure of normal septal resorption following normal fusion. This anomaly is classified by Toaff (8) as a Type 1A uterine communication and further supports Musset's theory of embryologic development.

This case report is interesting because it demonstrates the value of MRI as an adjunctive imaging modality for characterization of müllerian anomalies. It also demonstrates the utility of MR-based 3D reconstruction to aid visualization of the complex anatomy of this anomaly. In the past, most studies employed hystero-salpingography (HSG), ultrasound and/or laparoscopy/laparotomy/hysteroscopy to characterize these anomalies. However, there are several recent reports of the effective use of MRI in the evaluation of uterine anomalies (5-7). Fedele et al. (5) and Carrington et al. (6) reported correct classification of müllerian anomalies 100% of the time when using MRI as an adjunct to surgery. Doyle (7) correctly diagnosed 23 of 24 uterine anomalies (96%) with MRI, compared to 85% (11/13) for trans-vaginal ultrasound and 6% (1/17) for HSG. MRI has also been utilized to correctly identify a septate uterus where ultrasound had suggested didelphys.

Fedele et al. (5) used several diagnostic criteria on MRI used for differentiating between bicornuate/didelphic and septate uteri. Maximal fundal indentation < 10mm and an angle between the medial margins of the hemicavities of < 60
degrees differentiated septate from bicornuate/didelphic uteri. Additionally, they found that the use of T2-weighted images provided better differentiation of the endometrial and myometrial tissue planes, further helping to delineate types of double uteri. Our MRI study demonstrated a maximal fundal indentation of 4 mm. The angle between the medial margins of the hemicavities was 29 degrees. Evidence of complete septation between the cavities is shown in Fig 5 (arrows), and is identical to the image presented by Fedele (9) of their complete septate uterus (Fedele fig 4a). Based on these criteria, diagnosis of complete septate uterus was made. Accordingly, the patient chose to forego the risks of an additional procedure, namely hysteroscopy, in favor of expectant management.

MRI offers several clear advantages over the other diagnostic modalities. It is noninvasive and so does not require hospitalization, anesthesia or surgery. It allows visualization even in difficult surgical candidates with co-morbid illnesses or significant adhesive disease. It also allows for easy evaluation of the renal collecting system. This case supports the position of other investigators that MRI evaluation should be considered as a viable testing modality in the evaluation of müllerian anomalies. The 3D reconstructed images facilitate intuitive understanding of the anatomic relationships, which allows clinicians and trainees to better appreciate the structural relationships that underlie this anomaly.

References:


**Figures:**

![Fig 1: Axial MR image demonstrating double uterine cavities (white arrows), and uterine septum (Black Arrow). The ovaries are the circular structures on either side of the uterine cavities.](image-url)

Fig 3: Laparoscopic view of uterus. Probe demonstrates contiguous uterine fundus.
Fig 4: View of double cervix after resection of longitudinal septum. Double cervixes are labeled A and B.

Fig 5: Axial MR image showing complete septum (arrow). This is identical to Fedele's case of complete septum (Obstet Gynecol 1989; 74(6): p846 Fig 4a).