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Case Report

An atypical Müllerian duct anomaly: Duplicated cervices and vaginas with a single uterine cavity ☆

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ABSTRACT

Müllerian duct anomalies include a wide variety of developmental abnormalities involving the female reproductive system, many of which are not adequately represented by the current classification system used in the United States. Diagnosis can be made with imaging, but initial evaluation first requires a thorough physical exam. A 19-year-old female received a pelvic MRI for evaluation of a Müllerian duct anomaly following an abnormal pelvic exam. Imaging demonstrated a single uterine cavity which divides into 2 distinct cervices and vaginas. The patient received a hysteroscopic resection of her vaginal septum. This type of anomaly is extremely rare and associated clinical outcomes of potential infertility or complications with vaginal delivery are uncertain. Use of a more comprehensive classification system for Müllerian duct anomalies may assist with identification and research of such rare subtypes.

Introduction

Müllerian duct anomalies (MDAs) are anatomical variants of the female reproductive system due to congenital errors in Müllerian duct fusion and development. From weeks 8 to 18 of embryogenesis, the Müllerian ducts undergo a complex multistep process of elongation, fusion, and septal resorption [1]. A failure in any portion of this development can produce a wide variety of uterine anomalies, from uterine septations to complete duplication of the uterus and vagina. The incidence of uterine anomalies is estimated at 5.5% of the general female population with an increasing prevalence of up to 24.5% in populations experiencing miscarriage and infertility [2].

Patients with MDA may be asymptomatic or may present with primary amenorrhea, infertility, dysmenorrhea, or dyspareunia. MDAs are associated with an increased risk of adverse pregnancy outcomes which vary depending on the type of the anomaly. Pregnancies in patients with a known MDA are given special attention due to this increased risk. Treatments range from conservative management to a variety of surgical procedures.

Diagnosis of a distal MDA is initially made by pelvic examination revealing a double vagina or double cervix.

☆ Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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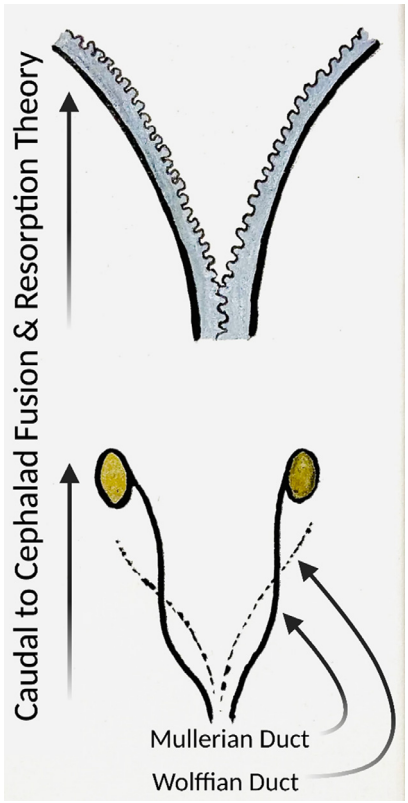


Fig. 1 – (Created with BioRender.com).

Laparoscopy has been the traditional gold standard for diagnosis of more proximal Müllerian anomalies. Other techniques have been used to support or make the diagnosis of a MDA, such as transvaginal ultrasonography, sonohysterography, magnetic resonance imaging (MRI), or hysterosalpingography. MRI can be particularly helpful due to its high accuracy and noninvasive nature. Three-dimensional ultrasound combined with pelvic examination has been shown to produce noninferior results to MRI and provides an alternative method of diagnosis [3].

The American Fertility Society (AFS) classification of Müllerian anomalies (published in 1988) has long been the most widely used classification system and was based on a system introduced by Buttram and Gibbons in 1979. This system described specific uterine subtypes such as the arcuate, septate, bicornuate, and unicornuate uterus as well as uterus didelphys, and uterine agenesis [4,5]. This initial classification system was founded on the principle that Müllerian fusion proceeds in a caudal to cranial direction (Fig. 1). This system was recently updated and expanded by the American Society for Reproductive Medicine (ASRM) in 2021 [6] (Fig. 2) and remains the dominant classification system used in the United States. However, several other international classification systems have been developed which are not based on the premise of caudal to cranial Müllerian fusion and which allow for more accurate characterization of certain unusual anomalies. Preeminent among these is the European Society of Human Reproduction and Embryology (ESHRE) and the European Society for Gynaecological Endoscopy (ESGE) [7] classification system, which divides classification separately by each individual embryologic subtype, namely the uterus, vagina, and cervix. In this case report, we discuss a rare type of MDA which is not classifiable by the AFS/ASRM system (Fig. 3), but which can be classified by the ESHRE/ESGE system.

Case report

A 19-year-old female with no pertinent past medical history underwent a routine pelvic examination which revealed duplication of the vagina and cervix. MRI of the pelvis was performed before and after administration of 12 mL of IV Dotarem contrast.

MRI showed a morphologically normal retroverted uterus with a normal endometrium. There was complete duplication of the cervix with suspicion for a longitudinal vaginal septum or vaginal duplication (Fig. 4). The patient was recalled for further imaging, at which time she self-administered ultrasound gel into her vagina, confirming the presence of a thick septum separating 2 hemi-vaginas (Fig. 5). The ovaries were normal in size, containing multiple follicles. No free fluid or other relevant findings were noted.

The patient received hysteroscopic resection of the vaginal septum using electrocautery as well as an endometrial biopsy due to abnormal appearing endometrial tissue on hysteroscopy. Anatomic findings on MRI were confirmed during the procedure. Pathology of the vaginal septum and endometrium demonstrated benign squamous mucosa.

Discussion

A single uterus with duplicated cervices and vaginas is an exceedingly rare MDA of uncertain clinical significance with the literature limited to a handful of case reports [8]. This anomaly is not classifiable according to the AFS/ASRM classification system [5,6] despite recent updates to the system to include numerous other types of anomalies. This is likely due to the fact that a duplicated cervix and vagina with a single uterine cavity opposes the assumed fundamental principle of caudal to cranial Müllerian fusion. The anomaly discussed in this case report, however, can be accurately described using the U1cC2V1 notation of the ESHRE/ESGE system [7], which divides categories by their individual embryologic origin.

Our patient was asymptomatic at the time of presentation, making initial diagnosis by pelvic examination crucial for accurate diagnosis and treatment. Possible presenting symptoms such as amenorrhea or dyspareunia are vague and elicit a broad differential, requiring careful physical examination for further evaluation. Undiagnosed and/or untreated similar uterine anomalies are associated with an increased risk of obstetric complications [9,10]. MRI proved helpful in this case in supporting the physical exam findings and demonstrating the complete nature of the vaginal septation. Administration of intravaginal gel aided in the demonstration of the vaginal septum. MRI was also useful in excluding other anomalies which

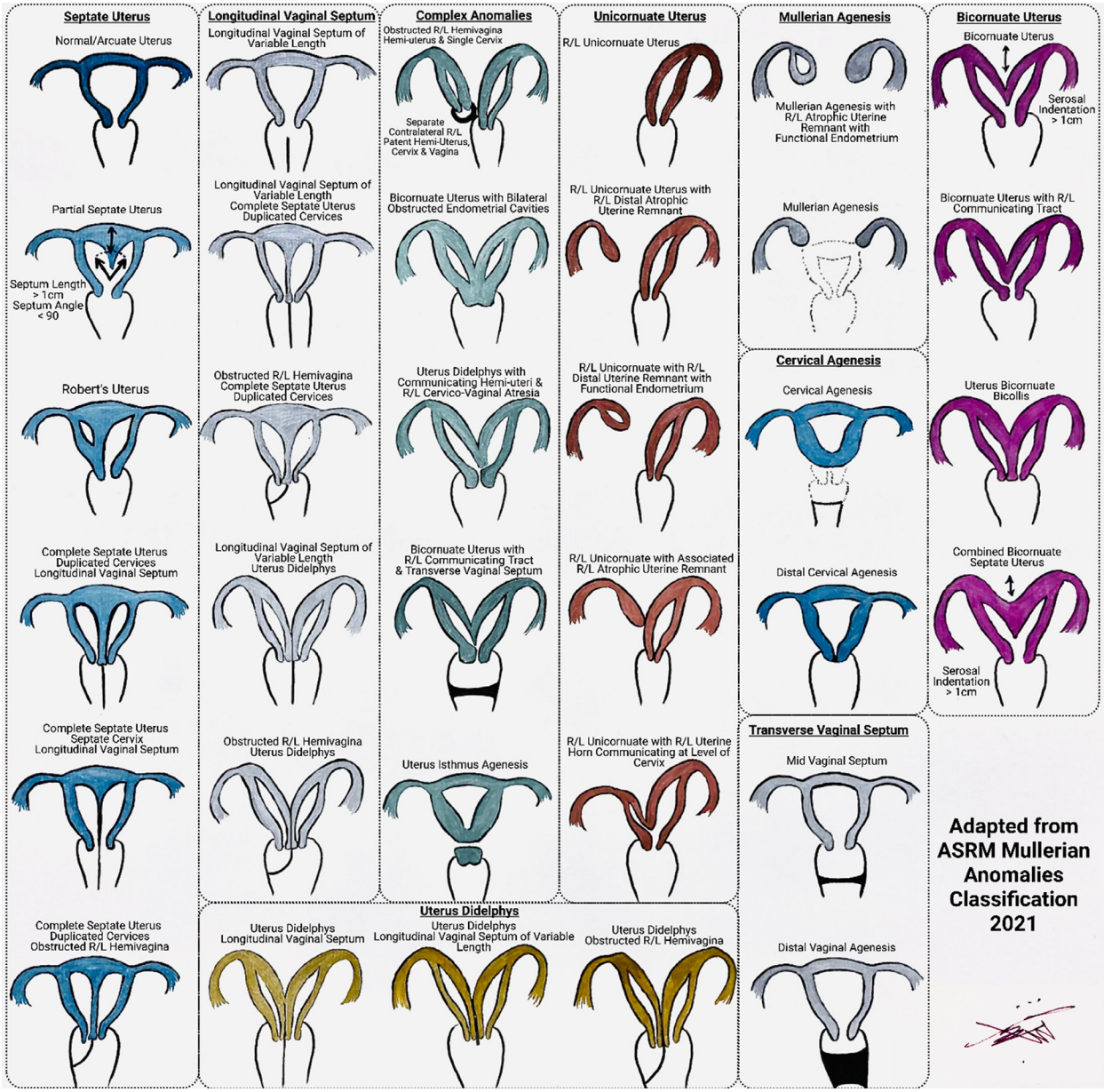
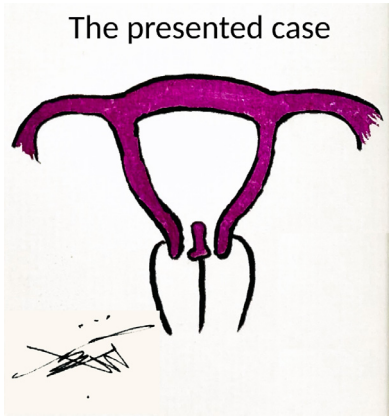


Fig. 2 – (Created with BioRender.com).

cannot be assessed on physical exam, such as a didelphys or septate uterus.

It is unclear whether the patient's MDA will affect fertility or future pregnancies due to lack of evidence on this subtype of Müllerian anomaly. Some studies suggest that the degree of uterine septation is the factor which most correlates with adverse pregnancy outcomes; however, our patient did not have a uterine septation. There is limited research on outcomes of isolated resection of a vaginal septum, but studies of combined uterine and vaginal septum resection have reported improvements in dyspareunia and dysmenorrhea [11]. Regarding duplicated cervices, however, there is no consensus on the management of this anomaly, and treatments range from simple observation to hysteroscopic metroplasty, with limited evidence for the efficacy of the latter [12]. Our patient's surgeons decided not to address the cervical duplication during the vaginal septoplasty due to the otherwise normal appearance of the uterus. More research is needed on this abnormality to determine the best and most appropriate management for this anomaly.

In conclusion, MDAs are a variety of congenital defects involving the female reproductive system which can present with a broad array of symptoms and degrees of severity. MRI, in conjunction with physical exam, is exceptionally useful in clarifying complex MDAs. Due to the significant variety of anomalies, patients require an individualized approach to diagnosis and management. Further research is needed to identify appropriate treatment for these highly variable anomalies. An updated classification system such as the one established by ESHRE/ESGE may improve research and clinical outcomes.



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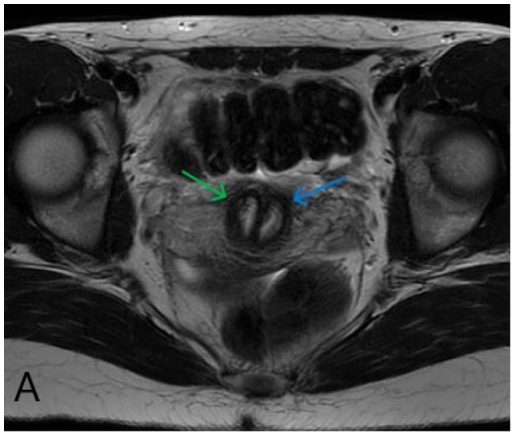


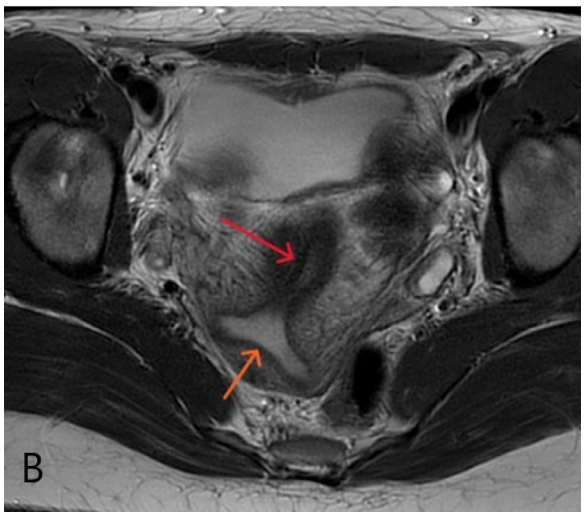
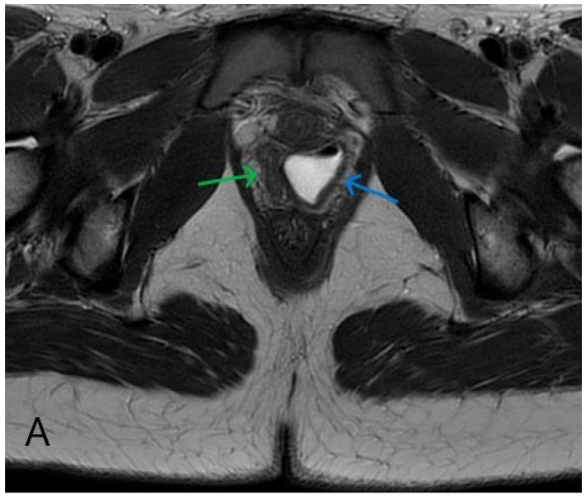


Fig. 4 – (A) T2 sequence axial image of the pelvis demonstrating a duplicated cervix. The green arrow is pointing to the right cervix and the blue arrow is pointing to the left cervix. (B) T2 axial image at a more superior cross-section demonstrating a single uterine cavity (orange arrow) and a longitudinal cervical septum (red arrow).



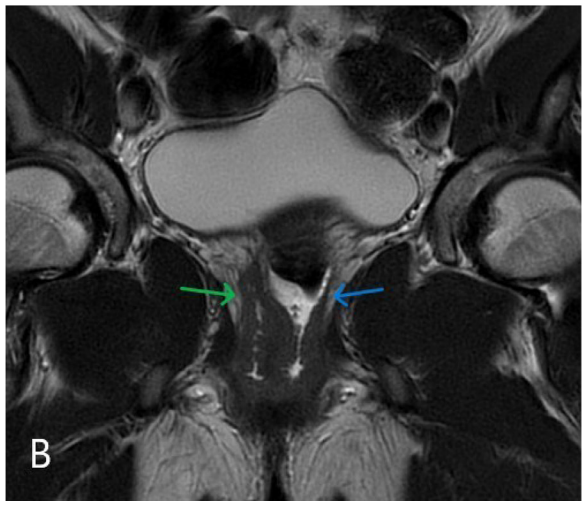


Fig. 5 – (A) Axial and (B) coronal T2 images at the level of the duplicated vaginas following administration of intravaginal gel. The gel remains within the left vagina (blue arrow), confirming complete duplication. The right vagina (green arrow) is collapsed.

Patient consent

Written, informed consent was obtained by the patient to publish this case.

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