Incidental Finding of a Müllerian Malformation on Two-dimensional Ultrasound: What Now?

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Section 2 – Answer

Case

A 25-year-old woman, nulligravida, with no relevant medical or surgical history, chronic medication, gynecologic symptoms, or history of infertility, underwent controlled ovarian stimulation for voluntary oocyte donation.

During vaginal ultrasound (US)-guided oocyte retrieval, the presence of a longitudinal vaginal septum reaching the introitus was noticed, separating the vagina into two parts, with two symmetrical small uterine cervixes [Figure 1].

On vaginal US, two cervical canals [Figure 2] and two uterine cavities [Figures 3 and 4] were noticed. At the level of the miduterus, there was a median discontinuation of the uterine wall, separating the uterine body in two [Figure 5-7].

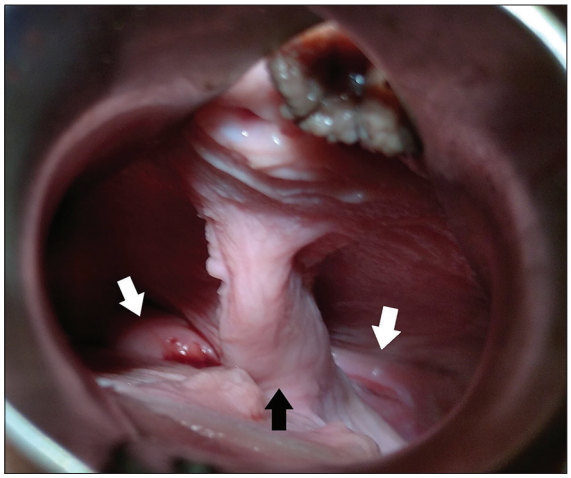

Figure 1: Complete longitudinal nonobstructing vaginal septum (black arrow) and two cervixes (white arrows)

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Interpretation

Congenital malformations of the female urogenital tract, with a prevalence of 2%–10%, are the result of an impaired development of the Müllerian ducts and/or urogenital sinus. The fallopian tubes, the uterus, and the upper part of the vagina have their embryological origin in the Müllerian ducts, while the urogenital sinus originates the most distal part of the vagina. Any disruption in any part of this process may lead to an impaired genitourinary development.

There are two main worldwide accepted classifications that of the American Fertility Society (AFS) and that of the European Society of Human Reproduction and Embryology (ESHRE) and the European Society of Gynecologic Endoscopy (ESGE). The former is simpler and based on the embryologic development, dividing the anomalies into seven categories – agenesis/hypoplasia, unicornuate, didelphys, bicornuate, septate, arcuate, and diethylstilbestrol-related abnormality.[1] The European system consists of a more functional classification,

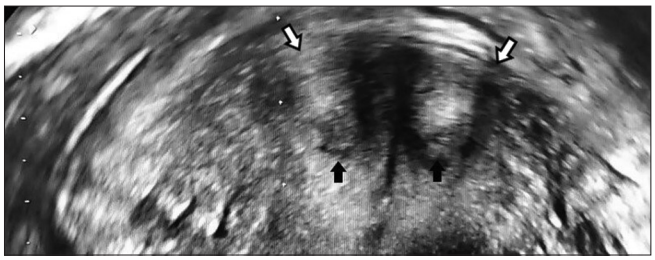

Figure 2: Two-dimensional vaginal ultrasound, axial section at the level of the uterine cervix, showing two cervical canals (black arrows) with distinct outlines (open arrow pointing uterine serosa)

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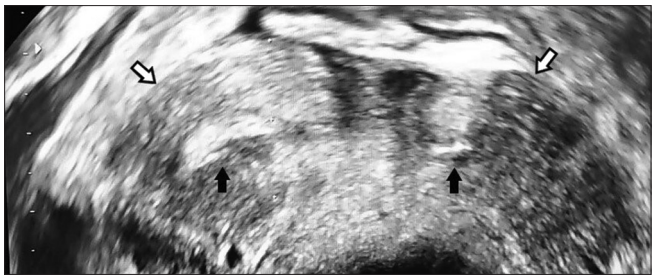

Figure 3: Two-dimensional vaginal ultrasound, axial section at the level of the uterine isthmus, showing two uterine cavities (black arrows) but a single uterine body (open arrow pointing uterine serosa)

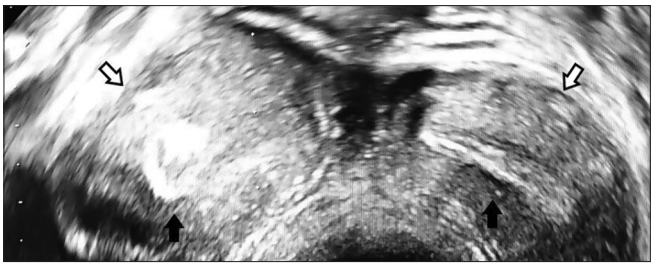

Figure 4: Two-dimensional vaginal ultrasound, axial section at the level of the uterine body, showing two uterine cavities (black arrows) but a single uterine body (open arrow pointing uterine serosa)

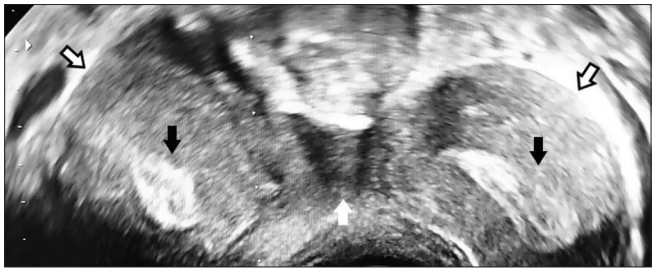

Figure 5: Two-dimensional vaginal ultrasound, axial section at the level of the uterine fundus, showing two uterine cavities (black arrows), and the beginning of the discontinuation of the midline wall (open arrows pointing the serosa, white arrow pointing the median part of the uterine fundus)

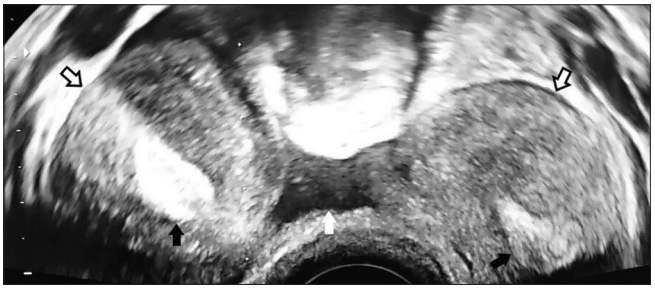

Figure 6: Two-dimensional vaginal ultrasound, axial section at the level of the uterine horns, showing two uterine cavities (black arrows), and two separated uterine horns (open arrows pointing the serosa, white arrow pointing free fluid lying on the uterine fundus, and interposed between the two uterine horns)

dividing the anomalies into subclasses for each affected organ – uterine body (U), the cervix (C), and the vagina (V).[2,3] There seems to be no genetic cause for these disorders and most of the patients are asymptomatic, although some may present dyspareunia, dysmenorrhea, obstruction of the reproductive tract, infertility, or poor obstetric outcomes. Thus, most frequently, these abnormalities are found incidentally during gynecological examination or US.[4]

Both pelvic US and Magnetic resonance Imaging (MRI) are considered the gold standard for diagnosing uterine congenital disorders. To carry out a complete evaluation of any uterine malformation, it is not only important to focus on the endometrial cavity but also to search for defects of the uterine outline. T-shaped cavities, infantile uterus, and complete or partial septate uterus (ESHRE U1 and U2) consist of isolated defects of the uterine cavity. On the other hand, bicorporal uteri (U3) present a median separation of the uterine body.[5]

US is especially useful in disorders affecting the uterine cavity. Sequential axial two-dimensional US (2D-US) sections may differentiate between two uterine cavities (AFS III: uterus didelphys; ESHRE U3: Bicorporal uterus) and a single cavity with a septum (AFS V/ESHRE U2: Septate uterus). Furthermore, the same procedure may give a sense of the septum's extent in relation to the length of the endometrial cavity if the septum reaches the internal cervical orifice, then it is called a complete septum (ESHRE U2B). Three-dimensional US (3D-US) is of great value to describe cavity defects such as septa or T-shaped uterus, because a full reconstruction of the cavity is performed, thus enabling the visualization of the whole cavity in a single image.[5]

US may have a limited role to evaluate the outline of the uterus, as subtle indentations of the uterine fundus may pass unnoticed. Again, serial axial 2D-US sections may give a sense of the extent of fundal indentations. Theoretically, 3D-US would be useful to define the uterine fundus shape, but most of the times it is hard to get a section visualizing the whole uterine wall, especially if the two horns are distant from each either, as in this case (caused by interposed liquid), limiting the use of this technique.[6] The diagnosis of cervical and especially vaginal defects may not be possible exclusively using US, but this may easily be completed by gynecological examination.

MRI is the best tool to diagnose uterine malformations. Based on MRI, it is not only possible to evaluate uterine cavity but also to accurately define the outline of the uterus and disorders of the cervix and the vagina, if present.[7]

Alternatively, to radiologic exams, combining hysteroscopy with laparoscopy may also diagnose Müllerian malformations.

These abnormalities alone do not require treatment unless they have any clinical impact. Patients with vaginal septa may require surgical correction if it impairs sexual intercourse or obstructs the genital tract.[8] Furthermore, patients with infertility or history of obstetric complications due to cavity defects, such as septum or T-shaped uterus, may require metroplasty.[9] However, in the presence of a uterine outline defect, such as bicorporal uterus, surgical correction may not be

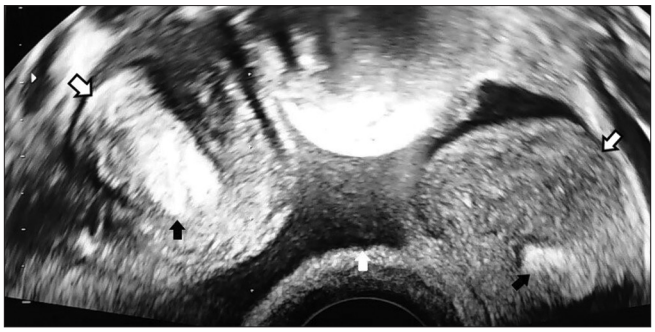

Figure 7: Two-dimensional vaginal (ultrasound), axial section at the level of the upper limit of the uterine horns, showing two uterine cavities (black arrows) and two uterine horns (open arrows pointing the serosa, white arrow pointing free fluid lying on the uterine fundus, and interposed between the two uterine horns)

feasible as it may compromise the integrity of the uterine wall and thus reduce the possibility of reconstructing a functional organ. Furthermore, in cases of uterine hypoplasia, not only the uterine cavity but also the whole uterus is small in all its dimensions, so surgery is of little interest and hormonal therapy (pseudogestational treatments) may be useful.

This case describes the diagnosis of a congenital Müllerian malformation based solely on physical examination and 2D US, given that this occurred in an oocyte donation context, and thus more complex techniques were not feasible.

At physical examination, there was a complete longitudinal nonobstructing thick vaginal septum-V1 category. There was one cervix in each side of the vaginal septum, both symmetrical and of slightly reduced size-double normal cervix-C2 category.

On US, sequential axial sections toward the uterine fundus revealed two uterine cavities, progressively further away from each other. This was enhanced by the presence of a thickened endometrium as a result of ovarian hyperstimulation for oocyte donation. At the miduterus level, there was a midline discontinuation of the uterine wall, separating the uterine body into two parts. This was better perceived because of the presence of free fluid lying on the uterine serosa, as a consequence of the ovarian stimulation and puncture. These features confirmed the presence of two endometrial cavities and an important fundal uterine indentation partially separating the uterine body in two, giving the appearance of a heart-shaped uterus. This corresponds to a bicorporal uterus or U3 according to the ESHRE/ESGE classification. Nevertheless, it is difficult to define the extent of the uterine indentation and the length of the uterine septum based solely on 2D US. When clinically relevant, a 3D scan (US or MRI) may be used to define a subclass within the U3 malformations.

According to the ESHRE/ESGE classification of the Müllerian malformations, this patient's genital tract may be classified as U3C2V1 – bicorporal uterus, double normal cervix, and complete longitudinal nonobstructing vaginal septum, – or Class III (didelphys) according to the AFS classification.

This case highlights the importance of the 2D US as a dynamic radiologic exam in the diagnosis of Müllerian malformations when more accurate exams such as 3D US or MRI are not available.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal her identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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