

## Case Report

# Complete Uterine Septum with Cervical Duplication and Longitudinal Vaginal Septum: An Anomaly Supporting Alternative Embryological Development

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ABSTRACT

The diagnosis and management of Mullerian abnormalities have revolutionized with the advent of magnetic resonance imaging, three-dimensional ultrasound, and endoscopic techniques. All the earlier unclassified abnormalities can now be classified as per the new European Society of Human Reproduction and Embryology 2013 nomenclature. The presence of complete uterine septum with cervical duplication and longitudinal vaginal septum reinforces the alternative theory of Mullerian development. The presence of this complex anomaly is discussed in the light of embryonic development along with management issues.

**KEYWORDS:** Embryology, laparoscopy, Mullerian duct

## INTRODUCTION

Mullerian abnormalities are rare and have a varied incidence of 1%–6%.<sup>[1]</sup> The earlier diagnosis was based on clinical, sonographic images, and during laparoscopy or laparotomy. With the availability of good three-dimensional (3D) ultrasound machines, magnetic resonance imaging (MRI), laparoscopy, and hysteroscopy, the diagnosis of Mullerian abnormalities has revolutionized. This is a very rare case of the complete uterine septum with a double cervix with vaginal septum. As per the earlier used American Fertility Society (AFS) classification, this case is one of the examples which falls in the unclassified group. However, as per the European Society of Human Reproduction and Embryology (ESHRE) 2013, all such anomalies can be clearly classified [Table 1].<sup>[2]</sup> They also challenge the unidirectional development theory and support the bidirectional development theory. Only a few such cases have been reported so far. This case highlights that management guidelines are not clear and surgical management has to be tailor made as per the clinical requirements.

## CASE REPORT

A 32-year-old nulliparous married woman presents with complaints of chronic lower abdomen pain, dyspareunia, and infertility. Her general physical examination was normal with body mass index of 22 kg/m<sup>2</sup>. On speculum examination, a longitudinal vaginal septum was seen in the proximal portion of the vagina and the cervix was partially seen beyond the septum, at a high up position [Figure 1]. On vaginal examination, a single cervix was felt high up, slightly deviated to the right, and a normal anteverted uterus. An ultrasound showed a subseptate uterus with midline thick muscular septum and normal adnexa. MRI revealed septate uterus with septa extending till the cervix [Figures 2 and 3]. No anomaly was found in any other organ.

To complete the diagnosis, consent was taken for vaginal septum removal, followed by diagnostic hysterolaparoscopy. The couple was given the option of septoplasty in the same sitting or later in case of pregnancy complications. They chose to have it later.

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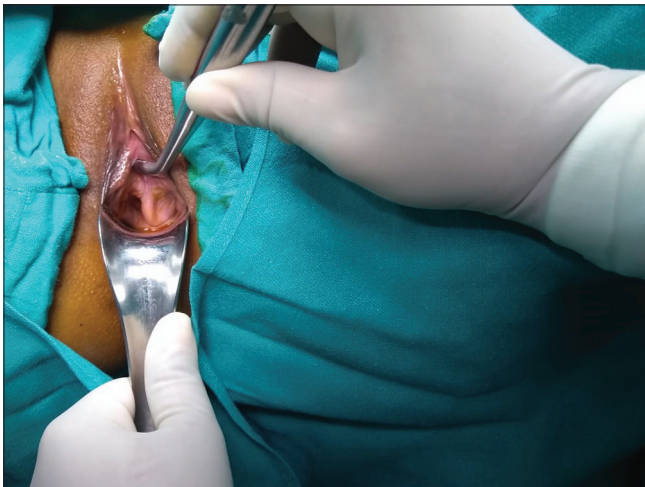
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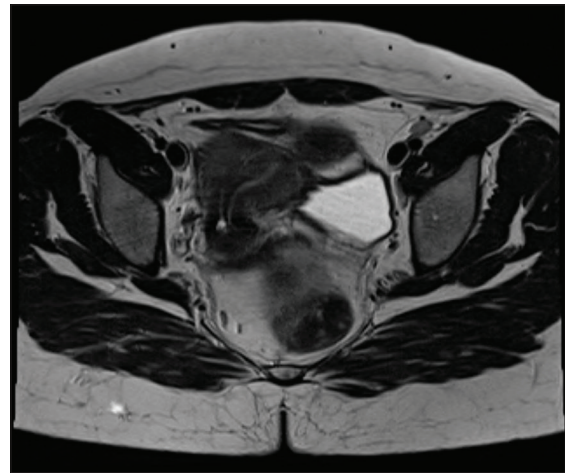
**Table 1: ESHRE classification of female genital tract anomalies**

U-Uterine anomaly		C- Cervical, V-Vaginal anomaly
Main Class	Sub Class	Coexistent class
U0 Normal uterus		C0-Normal cervix
U1 Dysmorphic Dysmorphic	a- T shaped b- Infantalis c- Others	C1-Septate uterus C2-Double normal cervix C3-Unilateral cervical aplasia C4- Cervical aplasia
U2 Septate Uterus	a- Partial b- Complete	V0-Normal vagina V1- Longitudinal nonobstructing vaginal septum V2- Longitudinal obstructing vaginal septum V3- Transverse vaginal septum with or without imperforate hymen
U3 Bicorniporeal	a-Partial b-Complete c-Bicorniporeal septate	V4- Vaginal aplasia
U4 Hemi-uterus	a- With rudimentary cavity (horn communicating or not) b- Without rudimentary cavity (horn without cavity or no horn)	
U5 Aplastic	a- With rudimentary cavity (bi or unilateral horn)	
U6 Unclassified malformations		

Final diagnosis: U C V

**Figure 1:** Speculum examination revealing septa in the vagina

Under anesthesia, the vaginal septum was held in between two allis forceps and divided in between the two with cautery. After the septum was removed, two dimples were seen high up in the vagina. They were double cervix confirmed by magnification with the hysteroscope. Two separate sounds could be inserted through the two cervixes, ruling out any cervical atresia [Figure 4]. The uterocervical length in both sides was 3.5 inches. Hysteroscope was introduced through both the cervixes. The septum was found to be complete till the level of the cervix delineating two separate cervical canals and two cavities. Cavities on both sides of the septum were found to be similar in size. The two ostia were visualized on the respective sides of the uterus. On introducing the laparoscope, the fundus was found to be smooth with a 2 cm fundal midline

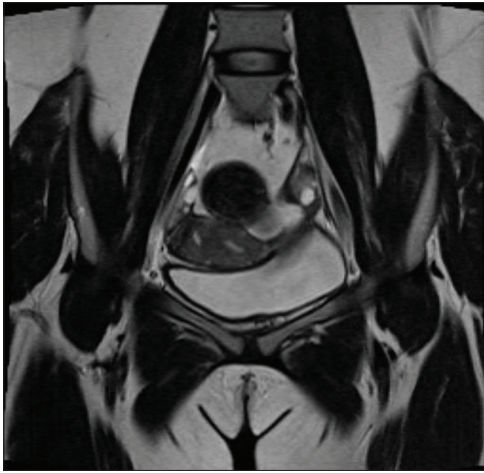
**Figure 2:** Magnetic resonance imaging (axial view) showing two uterine cavities extending till cervix with single fundus

fibroid and with normal tubes and ovaries [Figure 5]. Thus, a diagnosis of complete uterine septum with two cervixes and longitudinal vaginal septum was made. On chromopertubation spill was seen on both sides.

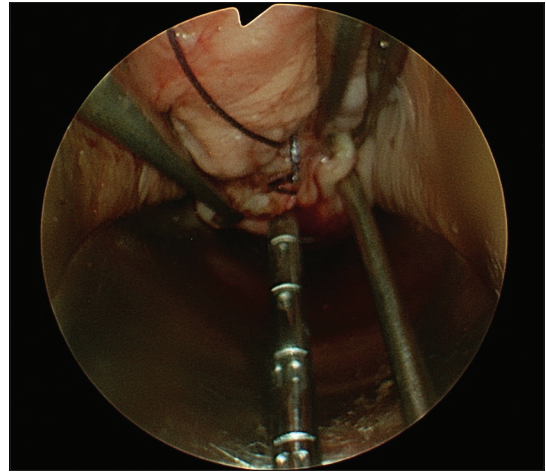
After 6 weeks of the vaginal septum resection, she reported an improvement in the dyspareunia and the heaviness in the lower abdomen [Figure 6]. She is being kept under follow-up to look for pregnancy complications and manage accordingly.

## DISCUSSION

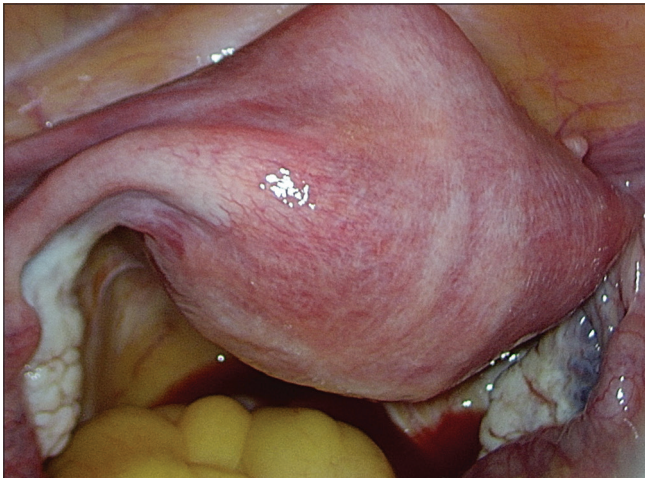
Embryologically, the uterus, cervix, and vagina are formed from the Mullerian ducts. There can be an absence of fusion or a full range of defects arising from anomalous fusion. These may occur as a result of genetic, environmental causes, and spontaneous developmental



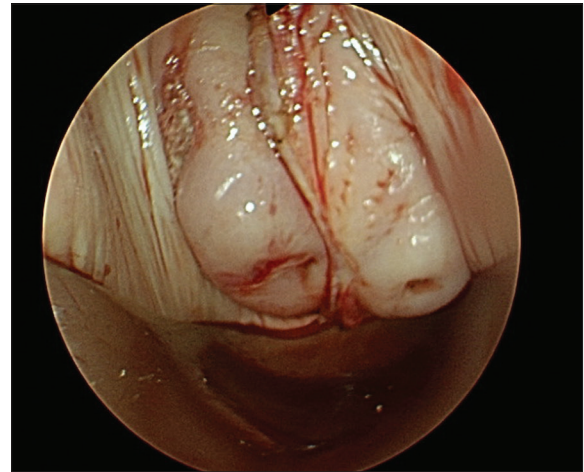
**Figure 3:** Magnetic resonance imaging (coronal view) showing two uterine cavities



**Figure 4:** Intraoperative view of double cervixes with sound in each uterine cavity



**Figure 5:** Laparoscopic view of the single fundus with small anterior fibroid



**Figure 6:** Six weeks postoperative view of both the cervixes

disabilities.<sup>[3]</sup> This is a very rare case which questions the embryological development of the female genital tract. If we assume that cervical agenesis occurs due to the failure of fusion of the Mullerian ducts on the two sides, then it is not possible to have duplication of the cervix with a smooth, single fundus. The development, therefore, is not unidirectional from caudal to cephalad direction. This supports an alternative embryological hypothesis that fusion and resorption start at the isthmus and proceed in the cranial and caudal directions at the same time. This was supported by a study by Chang *et al.* They studied the anomalies associated with vaginal septum and came across similar cases.<sup>[4]</sup>

A number of similar cases have come into literature in the last three decades.<sup>[1,4-6]</sup> All these support the alternative embryological hypothesis. While the AFS classification left many anomalies nonclassified, the ESHRE-2013 classification can incorporate this and many other anomalies.<sup>[2]</sup> As per ESHRE, it can be

denoted as U2bC1V2 making it easy to understand the complete anomaly. The complete diagnosis is possible only with a combination of MRI and diagnostic hysterolaparoscopy.<sup>[5]</sup> The septate uterus (U2) is associated with a number of obstetric complications such as second-trimester abortions, preterm labor pains, malpresentations, and infertility.<sup>[2]</sup> However, in this case, a trial for normal conception was planned. As the septum was complete, each cavity was uniformly smooth and in connection with a cervix. This might not be so bad for a spontaneous normal pregnancy. A surgery on the other hand would be highly challenging. There is also a high chance of cervical incompetence and cervical dystocia later.

The detailed classification also enables in planning the management of the patient depending on the primary complaints. In this case, as the primary complaint was a chronic pelvic discomfort and dyspareunia, the vaginal septum was removed and the complete uterine septum and its two cervixes were left as such. With uncovering

of the second cervix, subfertility may not be an issue. In case of pregnancy complications, a second stage surgery may be required.

In a similar case reported by Barbanti *et al.*, a primigravida woman with 8 weeks missed abortion was incidentally diagnosed with the same anomaly. She too had a resection of the vaginal septum after uterine evacuation for complaints of long-standing dyspareunia. Further procedure was planned in case of future pregnancy complications.<sup>[5]</sup> Ribeiro *et al.* reviewed those with a complete uterine septum, double cervix, and a vaginal septum. They found that most of them had dyspareunia and infertility.<sup>[6]</sup> In a study by Chang *et al.*, five nulligravida women underwent clinical, radiological, and surgical workup for complaints mainly pertaining to the vaginal septum and were found to have complete uterine septum with double cervix and a longitudinal vaginal septum.<sup>[4]</sup> Studies suggest that surgical interventional treatments should be targeted to relieve symptoms and preserve fertility.<sup>[7,8]</sup>

The need for surgical or conservative management is controversial. The resection of the vaginal septum is easy and should be done in all. To improve obstetric outcomes, hysteroscopic resection of the septum is recommended.<sup>[7,8]</sup> Regarding cervical unification, different studies have different recommendations. While some recommend it routinely, often under 3D transrectal sonography, others prefer to leave it to avoid problematic bleeding during surgery and cervical incompetence later on.<sup>[8]</sup>

## CONCLUSION

This case is one of the rare Mullerian anomalies, which is completely diagnosed with an MRI and diagnostic hysteroscopy and laparoscopy. The ESHRE classification lists a wider range of anomalies compared to the AFS system. However, in women with complete uterine septum and double cervix, the role of prophylactic metroplasty in patients with no previous miscarriages or pregnancy complications is still not clear. A long-term

follow-up of such patients will enable us to take a more informed decision in deciding the management.

## Declaration of patient consent

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

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Nil.

## Conflicts of interest

There are no conflicts of interest.

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