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Three Cases of Congenital Anomalies U2bC2V1 with different Reproductive Outcomes: Is it Cause of Infertility?

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ABSTRACT

Background: Congenital uterine anomalies (CUAs) consist structural disorders of the female genital tract due to abnormal fusion or resorption of the Müllerian ducts. The prevalence of CUAs is 5.5% in the general population, 8.0% in infertile women and 13.3% in women with a history of miscarriage.

Results: We present three cases of women with complex genital tract anomalies (U2bC2V1), including cervical duplication and vaginal septum with different reproductive outcomes.

Conclusion: A complete uterine septum with double functional cervix may have a wide spectrum of reproductive outcomes. We advised nulliparous women to complete their evaluation by performing hysteroscopy. It provides reliable information for the anatomical status of the cervix, tubal ostia and, especially, uterine cavity. In cases of infertility or recurrent pregnancy losses, the dilemma is whether to proceed with the resection of the uterine septum in combination with resection of vaginal one.

Key Messages: A complete uterine septum with double functional cervix may have a wide spectrum of reproductive outcomes; The association of uterine anomalies with the woman's fertility status remains controversial; Patients with septate uterus and history of infertility or poor pregnancy outcome should complete their investigation and proceed with resection of septum.

INTRODUCTION

Congenital Uterine Anomalies (CUAs) comprise of structural disorders of the female genital tract that arise because of abnormal fusion or resorption of the Müllerian ducts.

The prevalence of CUAs is estimated to be at 5.5% in the general population, whilst up to 8.0% in infertile women, 13.3% in women with a history of miscarriage and 24.5% in those who were infertile that have suffered a miscarriage [1]. The past few years a combination of Three-dimensional (3D) ultrasound, Magnetic Resonance imaging (MRI), laparoscopy and hysteroscopy had contributed significantly to the classification these anomalies. In contemporary medicine, the majority of cases can be diagnosed in a less invasive way because of the increased use of MRI and 3D ultrasound [2].

CUAs are mostly asymptomatic and incidentally diagnosed during the annual check-up or investigation of recurrent pregnancy loss or infertility. The reproductive outcomes in women with such anomalies range widely: from normal to severe adverse outcomes, including preterm birth or miscarriage [1,3].

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In this work we present three cases of women upon patient consent with complex genital tract anomalies classified as per the ESHRE consensus (U2bC2V1), including complete uterine septum, cervical duplication and vaginal septum respectively, with different reproductive outcomes [4].

CASE PRESENTATION

1st Case

A 51-year-old G2P1 (vaginal delivery) presenting with severe worsening menorrhagia attributed to multiple leiomyomas, was referred to our clinic. Pelvic examination revealed an enlarged uterus, small vertical vaginal septum in the upper third of vagina and two external cervical orifices. Complete septate uterus and cervical duplication were found during 2D (Two Dimensional) Transvaginal Ultrasound (TVUS) and confirmed by MRI. Since she had already undergone three myomectomies and after excluding of endometrial pathology by conducting endometrial biopsy, hysterectomy was recommended. Those findings were also confirmed intraoperatively. The histopathology report mentioned the presence of two intracervical canals, 3.5 cm each, and two external cervical orifices **(Figure 1)**.

2nd Case

A 25-year-old G0P0 woman presenting with dyspareunia attended our clinic for consultation. The gynecological examination revealed a longitudinal non-obstructing vaginal septum with a well-formed cervix in the left and a hypoplastic one in the right side. During 2D ultrasound complete septate uterus was revealed and patient was referred for pelvic MRI to confirm this finding. MRI findings enabled classification of the identified anomaly as U2bC2V1. The patient underwent resection of the vaginal septum and diagnostic hysteroscopy from both cervical orifices. Laparoscopy was performed simultaneously due to a persistent ovarian simple cyst, measuring 6 cm in diameter (which was resected) and confirmed normal contour of uterus **(Figure 2)**.

3rd Case

A 38-year-old G0P0 infertile woman presented to our clinic for fertility counseling and dyspareunia management. The patient was evaluated by pelvic examination, TVUS and hormonal blood test assessment for ovarian reserve markers. Gynecological examination revealed a vertical vaginal septum on the left side of vagina with a well-formed cervix in the right side. The left part of the vagina was too small to examine the left cervix. TVUS revealed 2 normal cervixes and 2 endometrial cavities completely separated by septum. The external contour of the uterus, according to 2D ultrasound and MRI, was found to be normal. Hysterosalpingography was performed only from the right side and revealed well-formed right side of the uterine cavity and patent right fallopian tube. After TVUS and blood test assessment, the patient was diagnosed with poor ovarian reserve. The AFC (Antral Follicle Count) was 4 and her AMH (Anti-Mullerian Hormone) was 0.9 ng/ml. Partners' sperm count was found to be within normal range. Therefore, the patient's infertility was attributed mostly to poor ovarian reserve and the possibility of *In Vitro* Fertilisation (IVF) was discussed. The woman was proceeded with diagnostic hysteroscopy both of cavities and resection only of the vaginal septum (**Figure 3**).

DISCUSSION

Septate uterus is believed to evolve due to the failed fusion or resorption of the Müllerian ducts, during the period of embryologic development [5]. According to the new ESHRE

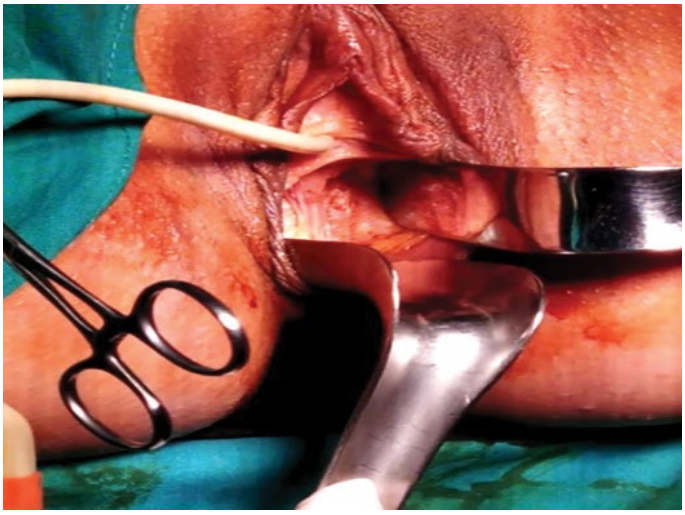

Figure 1: Supporting image from case 1 as discussed in text.

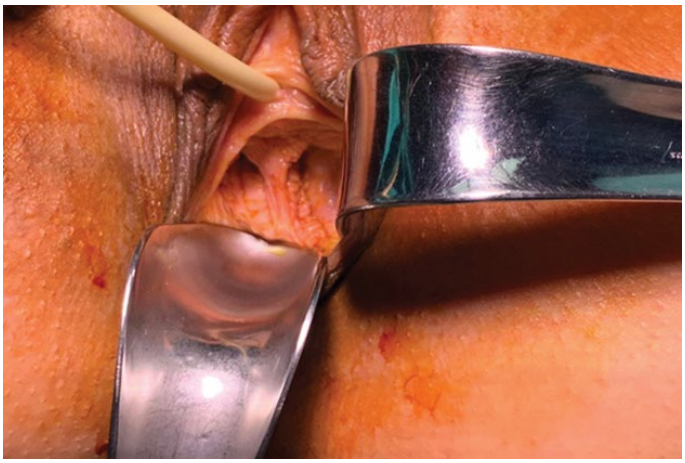

Figure 2: Supporting image from case 2 as discussed in text.

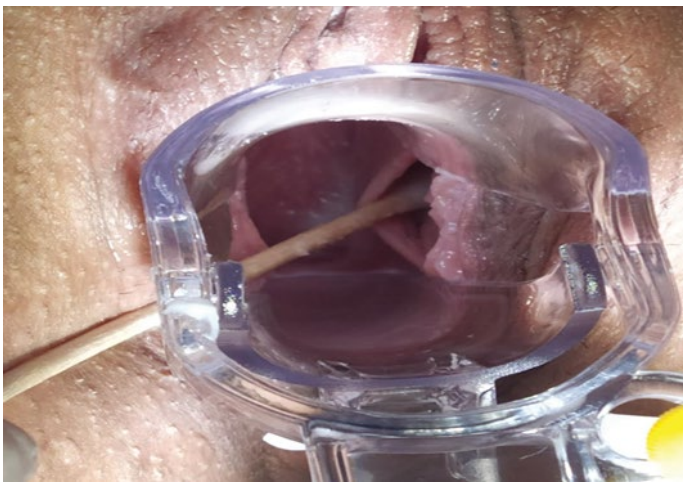

Figure 3: Supporting image from case 2 as discussed in text.

(European Society of Human Reproduction and Embryology)/ ESGE (European Society for Gynaecological Endoscopy) classification system of female genital tract congenital anomalies, septate uterus involves all those cases with a normal external contour and an internal indentation extending >50% of the uterine wall thickness [4].

During the last few years, cases of congenital anomalies combining cervical duplication with a uterus of normal shape have been reported. The different types of CUAs have been associated with varying degrees of suboptimal reproductive outcomes. Women with canalization defects seem to have the poorest reproductive performance, including reduced conception rates, which more often than not are linked to implantation failure [6-9]. The present study describes three cases of women with duplicated cervices and septate uteruses. A complete uterine septum with double "normal" cervix may have a wide spectrum of reproductive outcomes. We advised nulliparous women to complete their evaluation by performing hysteroscopy. This provides reliable information of the anatomical status of the vagina, cervix, uterine cavity and tubal Ostia. There is no doubt that women with dyspareunia should undergo resection of vaginal septum. In cases of infertility or recurrent pregnancy loss, the dilemma remains as to whether or not to proceed with the resection of the uterine septum in combination with resection of the vaginal section.

Several studies have evaluated the association between congenital anomalies and reproductive outcomes with controversial conclusions since they are associated with both acceptable as well as adverse outcomes [10]. Canalization defects, such as septate uterus, seem to be associated with poor obstetrical outcomes including reduced conception rates, increased risk of miscarriages and preterm births. A systematic review of 3805 women with both canalization and unification defects reported decreased pregnancy rates in the canalization defect group and significant higher risk for preterm birth, miscarriage and fetal malpresentation. Additionally, women with septate uterus appear to have poorer outcomes when compared with those with subseptate uterus [9]. Naeh A, et al. [11] compared the course of 167 pregnancies of women with CUAs to the general population. Overall higher rates of major adverse outcomes were reported in the CUAs group but safe conclusions regarding the CUAs subtype could not be drawn. Although the exact mechanism that causes infertility in women with septate uterus remains obscure, implantation alterations seem present the most plausible explanation [12]. Conversely, as infertility is a multifactorial condition, it is hard to be exclusively attributed to septate uteri [5]. These findings were confirmed by the first case of 51-year-old woman, who had no history of infertility and had one normal delivery.

Septate uterus is the most frequent congenital anomaly accounting for 35% of all uterine anomalies. Despite that, there is still no consensus regarding the proper management [13]. The most frequent intervention in these women includes hysteroscopic septum resection. Various observational studies report that the restoration of the anatomy of the uterus improves live birth rates [14-18]. Bendifallah S, et al. [15] have analyzed pregnancy rates amongst 128 women with primary infertility or recurrent miscarriage and septate uterus. After metroplasty, via hysteroscopy, 60.9% of women became pregnant while the birth rate in this group was reported at 53.1% and the miscarriage rate significantly decreased [15]. However, the outcome seems to be good even without an intervention. Rikken JF, et al. [19] reports the lack of evidence to support the surgical intervention and the need for relevant RCTs. From nine comparative studies regarding miscarriages and live birth rates with and without metroplasty, three supported better outcomes by surgery while six found expectant management safer and equally effective. Furthermore, the possibility of intrauterine synechiae after hysteroscopic resection of uterine septum should also be considered. Recently an international-multicenter-open-label -randomized controlled trial compared reproductive outcomes in women with septate uterus using either expectant management either septum resection in 80 patients was published. According to their findings live birth rates, miscarriages and preterm birth rates were not improved by the septum resection. More specifically, live birth rate occurred in 31% of the metroplasty group and 35% of the expectant group [20].

In our institution, we suggest all patients with complete uterine septum to perform diagnostic hysteroscopy. We used to adopt the expectant management in women without history of infertility if at least one endometrial cavity was found to be “normal” during diagnostic hysteroscopy. In infertile women, after excluding all possible causes of infertility (male factor, poor ovarian reserve etc.), the possibility of resection of uterine septum should be considered.

CONCLUSION

Congenital anomalies are not as rare as they may previously thought to be. The association of these anomalies with the woman’s fertility status remains controversial issue as their impact may vary from asymptomatic presentation to severe reproductive adverse outcomes. Symptomatology spectra and literature controversies makes the appropriate management of these patients challenging. The available research remains limited and consequently insufficient to allow for any safe conclusions to be drawn. However, here we suggest that patients with septate uterus and history of infertility or poor pregnancy outcome to complete their investigation and proceed with resection of septum.

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