A Second Successful Pregnancy in a Hybrid Uterus – Coexistence of Bicornuate and Septate Uterus: A Case Report

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Abstract

Congenital malformations of the uterus are rare. Pregnancies in these uterine abnormalities are usually associated with poor reproductive outcomes such as recurrent pregnancy losses, preterm birth and intrauterine growth restriction. Presented here is the case of a 21-year old G2P1(1001), who was diagnosed intraoperatively with a septate bicornuate uterus during her second Cesarean delivery. She had to undergo abdominal delivery for both her pregnancies due to malpresentation, and delivered live healthy babies with no gross structural defects. Pathophysiology and classification of congenital uterine malformations are discussed in the case, as well as the diagnostics and management for such conditions.

Key words: bicornuate uterus, septate uterus, uterine anomalies

Introduction

Congenital anomalies of the female genital tract are deviations from normal anatomy due to errors in embryological development of the Müllerian or paramesonephric ducts.[1] These conditions are rare, estimated to be 3% - 5% of the general population.[2,3] There are numerous variations in mullerian anomalies published, but two of the most commonly documented are bicornuate uterus and septate uterus. The incidence rate of a hybrid uterus with both bicornuate and septate configuration is unknown, but is assumed to be very rare. A descriptive study[4] approximated its incidence to be at 4.8% among cases of double-chambered uterine anomalies. Depending on the type and the degree of anatomical distortion, mullerian anomalies are associated with reproductive problems,[1] and are usually associated with recurrent pregnancy loss, fetal growth restriction, malpresentation, and preterm birth.[5]

Reported here is the case of a 21-year old G2P1(1001), who had 2 successful pregnancies, G1, a term breech and G2, a preterm breech, despite having a hybrid uterus with coexisting septate and bicornuate types. Both pregnancies were delivered via Cesarean section due to malpresentation. This paper shall illustrate intraoperative findings of a bicornuate septate uterus, and classify this according to the ASRM and ESHRE/ESGE guidelines. Pathophysiology of congenital uterine malformations is likewise discussed, as well as the diagnostics and management for such conditions.

The Case

This is a case of a 21-year old G2P1 (1001), Filipino, who came in at the emergency room for preterm labor, and subsequently admitted for emergency repeat low transverse cesarean section for fetal malpresentation.

Starting first trimester, regular prenatal check-ups were done in a local health center, beginning one month age of gestation. Patient denied exposure to radiation nor viral exanthems and had no history of infections nor blood pressure elevation for the

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entirety of pregnancy. Patient regularly took prenatal multivitamins, folic acid, and ferrous sulfate. Routine prenatal laboratory workups all showed normal results. An ultrasound done at 32 weeks age of gestation, just five days prior to her admission, revealed a single live intrauterine pregnancy in breech presentation of about 32 weeks age of gestation, with good cardiac activity at 135 bpm and estimated fetal weight of 1970 grams, with posterior high lying placenta grade II, and normal amniotic fluid index.

The patient had no known comorbidities, and with no maintenance medications previously taken. She denies having any vices, nor illicit drug use. Patient underwent low transverse Cesarean section for fetal malpresentation in 2018 in another institution, and delivered a live, term baby boy with birthweight of 2,400 grams, with no fetomaternal complications, nor gross fetal abnormalities.

A few hours prior to admission, patient started to feel regular, moderate to strong uterine contractions. Vital signs were stable and the patient was conscious, coherent and not in respiratory distress. She had warm moist skin, anicteric sclerae, and pink palpebral conjunctivae. Cardiopulmonary, gastrointestinal and neurologic examinations were all normal. Patient had globular abdomen, with fundic height of 26 cms, and fetal heart rate of 150 bpm. Upon internal examination, the cervix was fully-dilated, with intact bag of water, and fetal breech, and feet palpable through the fully dilated cervix.

The patient was immediately admitted for emergency repeat low transverse Cesarean section (LTCS). Intraoperatively, there were noted dense adhesions between the anterior lower third of uterus and peritoneum with intact previous LTCS scar. A gravid uterus with well-formed lower uterine segment was noted. There was adequate amount of clear amniotic fluid. A preterm live baby girl in right sacrum transverse position was delivered; with APGAR score of 9,9, birth weight of 1.95kg, birth length of 45cm, 35 weeks by pediatric aging. There were no gross anatomical deformities noted in the baby. Placenta was implanted posteriorly with three-vessel cord. The uterus was well-contracted. Upon further examination, there were two cornua noted, with more than 1 cm serosal indentation noted in between the cornua. There was only one cervix, and one vagina noted on further exploration, supporting an intraoperative diagnosis of a bicornuate uterus. Furthermore, there was a finding of partial intrauterine septum, that extended from the fundus up to the level of the internal os, thereby dividing the uterine cavity into 2 equal halves (Figure 1). The rest of the pelvic organs were grossly normal. The septum was left intact, and the uterus was repaired by layers.

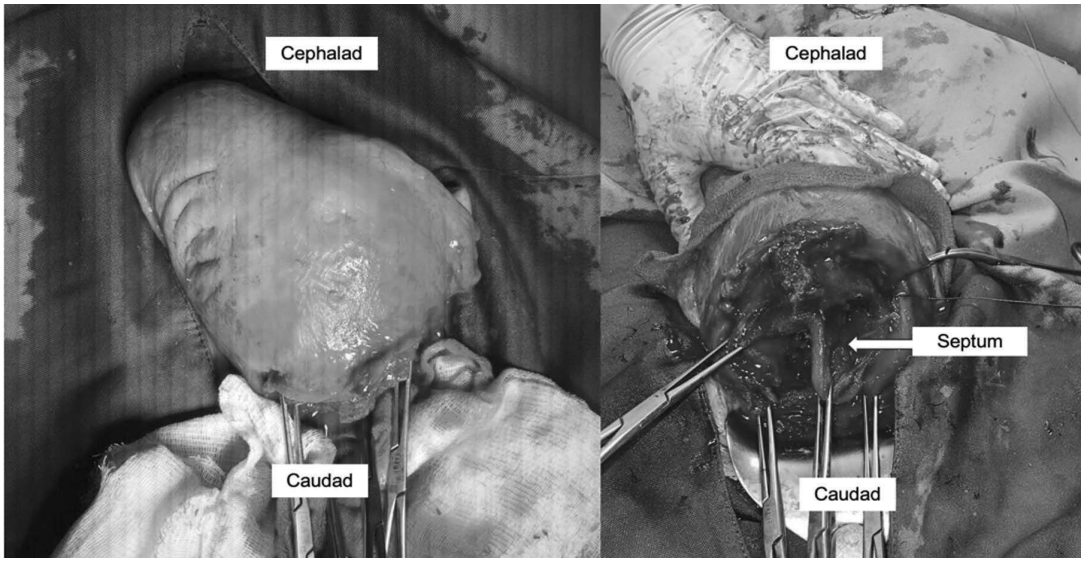

Figure 1. The exteriorized postpartum bicornuate uterus showing the cesarean incision post-delivery of the baby (left); Note the characteristic heart-shaped configuration, and the enlarged left cornu from which the baby was extracted. Note the intraoperative finding of partial uterine septum that extended from the fundus up to the level of the internal os, dividing the uterine cavity into 2 halves (right).

Postpartum, the patient had no subjective complaints, nor postpartum complications. She was sent home stable, on day 2 postpartum. Her baby, however, stayed at the neonatal intensive care unit for further observation due to prematurity, but was subsequently discharged stable, after a few days.

Discussion

Congenital malformations of the uterus stem from abnormalities in the combination, canalization, and resorption of the septum during the development of Mullerian ducts in the early fetal stages. Statistics show that in the general population, the incidence of uterine malformations is estimated to be between 3% and 5%, but prevalence is significantly higher among women with infertility and poor obstetric outcomes. Various published reports have described numerous variations in mullerian anomalies, but two of the most commonly reported are bicornuate uterus and septate uterus. The incidence rate of a hybrid uterus with both bicornuate and septate configuration is still unknown, but is assumed to be very rare. A descriptive study by El Saman et al4 approximated its incidence to be at 4.8% among cases of double-chambered uterine anomalies.

Classification System for Congenital Uterine Anomalies

Uterine anomalies are classified according to the American Fertility Society (AFS)/American Society for Reproductive Medicine (ASRM) and The European Society of Human Reproduction and Embryology (ESHRE)/ The European Society for Gynecological Endoscopy (ESGE) classifications systems. The latest ASRM classification system (Mullerian Anomalies Classification 2021 or MAC2021)[7], divides uterine malformations into nine categories: 1) Mullerian agenesis, 2) Cervical agenesis, 3) Unicornuate uterus, 4) Uterus didelphys, 5) Bicornuate uterus, 6) Septate uterus, 7) Longitudinal vaginal septum, 8) Transverse vaginal septum, and 9) Complex anomalies. Our index case is classified under "combined bicornuate septate uterus" (Figure 2). The ESHRE/ESGE classification is based on malformations of the uterine cervix and

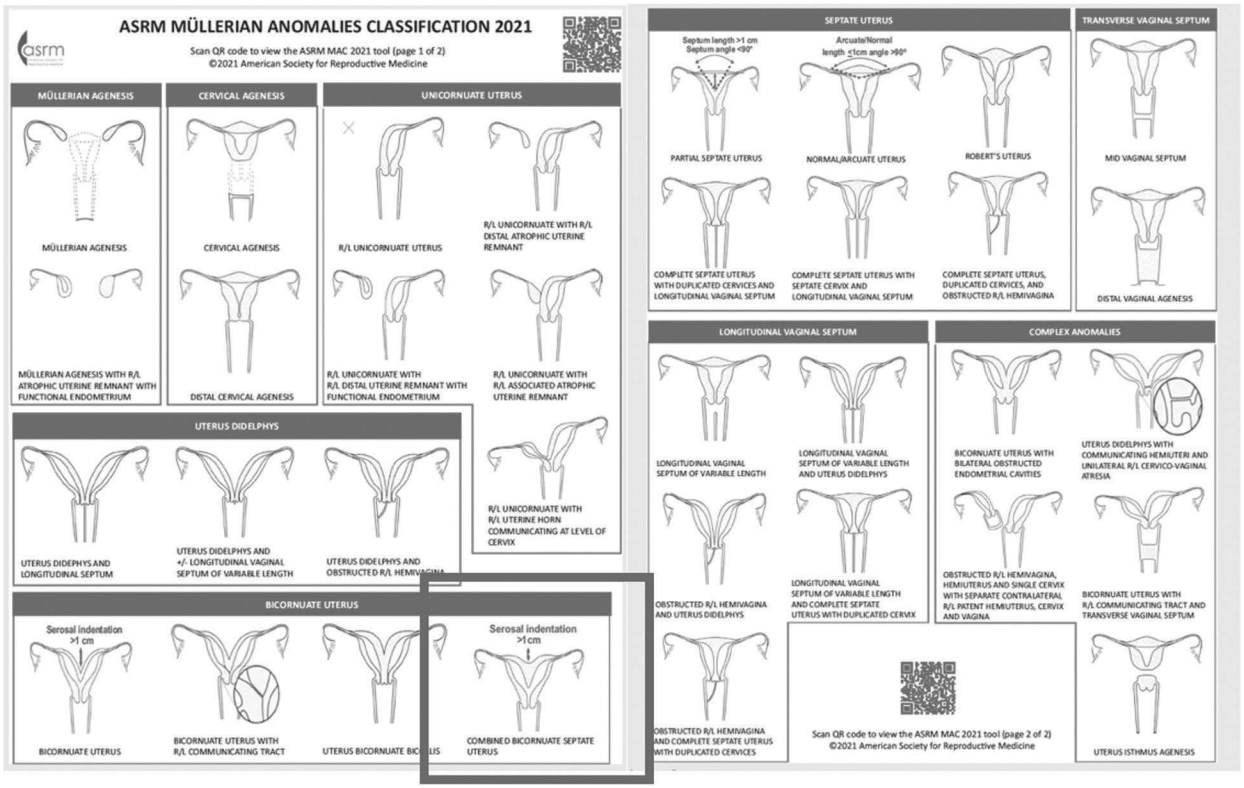

Figure 2. Classification system of uterine anomalies based on the new American Society for Reproductive Medicine Mullerian Anomalies Classification 2021 (MAC2021)[7]. Index case falls under "combined bicornuate septate uterus"

vagina and can even classify complex Mullerian anomalies.[8] The design of this classification system is based on the deviation of uterine anatomy derived from the same embryological origin and on the anatomical variations of the main classes expressing different degrees of uterine deformity with clinical significance respectively.[9] Using this classification, our index case falls under "class U3/C bicorporeal septate" (Figure 3).

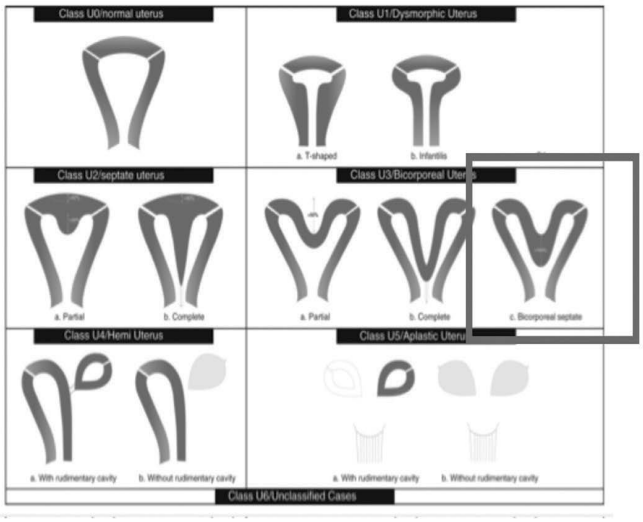

Figure 3. Classification of uterine anomalies based on the ESHRE/ESGE (2013).[1] Index patient falls under the category "class U3/C bicorporeal septate."

A bicornuate uterus (also known as the "heart-shaped" uterus), is a congenital uterine anomaly where the uterus has two separate horn-like structures, giving it a characteristic bicornuate appearance. This results from the incomplete fusion of the Mullerian ducts. It is often asymptomatic and may be incidentally detected only during routine evaluations. Bicornuate uterus can be associated with other congenital anomalies, such as renal abnormalities including agenesis of the kidney and ureter.

On the other hand, a septate uterus is the most commonly diagnosed congenital Müllerian anomaly.[10] This condition occurs when a band of tissue called a septum divides the uterine cavity partially or completely. A complete uterine septum extends from the fundus to the level of the external cervical os, while partial uterine septum extend from the uterine fundus to the cervical internal os.[11] The uterine septum is the congenital uterine anomaly most closely linked to recurrent pregnancy loss.[10]

Pathophysiology

The exact etiology behind Müllerian ducts anomalies is not fully understood. It is likely multifactorial, involving both genetic and environmental factors. Studies have suggested that genetic abnormalities or variations in gene expression could play a role in the development of uterine anomalies. Genetic research has identified several candidate genes (such as Pax, Lim1, Emx2, Wnt4, Wnt9b) that may be involved in Müllerian duct development, but further studies are needed to elucidate their exact roles and how variations in these genes may contribute to uterine anomalies. Additionally, environmental factors such as exposure to certain medications, toxins, or hormonal imbalances during critical periods of embryonic development may also influence Müllerian duct fusion and contribute to uterine anomalies.[12]

Around 7th to 9th week age of gestation, the Müllerian ducts fuse in the midline to form the uterovaginal canal. If there is impedance during the fusion of the Müllerian ducts (failure of complete and normal fusion), it can result in partial fusion and lead to anomalies such as a bicornuate uterus. Resorption of the septum comes later, at around the 9th to 13th week age of gestation, when the midline septum between the fused ducts disappears, and canalization of the uterovaginal canal occurs, forming a continuous cavity. However, if the resorption process is interrupted or incomplete, the septum may persist, leading to the formation of a septate uterus (failure of canalization).[13]

The coexistence of both bicornuate and septate varieties in a hybrid uterus supports the theory that lateral fusion and canalization may occur concurrently.[14]

Diagnosis

This patient came to this institution already heavily pregnant and in advanced labor, so a preoperative diagnosis of a mullerian anomaly will be quite difficult to establish. Ideally in nonpregnant women, several imaging modalities are available for diagnosing and managing congenital uterine

anomalies: 1) Hysterosalpingography (HSG), 2) 2D and 3D Ultrasound, and 3) Magnetic Resonance Imaging (MRI).

Most women with bicornuate and septate (or both) uterus usually are asymptomatic, and may be discovered only incidentally during routine pelvic imaging or diagnostic procedures. However, it is more likely to be identified after a first-trimester pregnancy loss, recurrent pregnancy loss, or other adverse obstetric outcomes such as preterm birth or breech presentation.

HSG is particularly useful for evaluating the general shape of the uterine cavity and assessing the patency of the fallopian tubes. In cases of suspected bicornuate uterus, HSG can help measure the intercornual angle (angle formed between the two uterine horns). A normal uterus typically has an intercornual angle of less than 105 degrees. If the angle is greater than 105 degrees and two separate uterine horns are visible, it suggests a bicornuate uterus.

Ultrasound is often the initial imaging modality used due to its non-invasive nature and lack of radiation exposure. A 3D ultrasound, in particular, is increasingly utilized for its higher precision and reliability in diagnosing mullerian anomalies. particularly in visualizing the uterus's shape and structure.[15] In fact, a study by Bermejo, et al[15] concluded that there is a high degree of concordance between 3D ultrasound and MRI in the diagnosis of uterine malformations.

When using ultrasound as a modality to diagnose mullerian anomalies, El Saman, et al[4] proposed that the presence of a "fundal depression" should not always be considered as a marker for "pure bicornuate variety", where there is no role for hysteroscopic interventions. He encouraged gynecologists to consider the presence of "fundal depression" to carefully evaluate the patient and look for possible "downward extension", such as a septum. He also advised gynecologists to consider the presence of a "fundal depression" as an indication to analyze the depth of fundal depression on laparoscopy, and the length of dividing septum through hysteroscopy.[4]

Lastly, MRI is considered the gold standard for diagnosing uterine anomalies due to its non-invasiveness, lack of radiation exposure, and ability to produce high-resolution images in multiple planes, thereby providing a comprehensive view of the anomaly.[16]

Management

The often controversial clinical questions in cases like this would be the following: (1) Should the uterine septum be removed during the cesarean section?, and (2) Should all women with bicornuate uterus and septum, regardless of reproductive outcomes and gravidity, undergo surgical management?

Pregnancies in women with uterine abnormalities are classified as "high-risk" due to their association with poor reproductive outcomes. The abnormal uterine shape can pose challenges for the developing fetus, potentially leading to complications during pregnancy and delivery, which may include increased risk of recurrent pregnancy loss, preterm birth, malpresentations, growth restriction, and fetal deformities. Therefore, careful monitoring and management are essential to mitigate these risks and optimize outcomes for both the mother and the baby.[17]

For this index case, the patient delivered two live babies (G1 live term, and G2 live preterm), both breech presentation, which necessitated 2 Cesarean deliveries. Both babies were healthy, with no deformities nor structural malformations. Although she had a preterm delivery for her second pregnancy, she did not have any history of miscarriage, which is quite remarkable for cases of hybrid uterus such as this. The inadequate room for rotation brought about by the coexistence of bicornuate and septate varieties can explain the fetal malpresentation.[18] Although the baby delivered did not have any gross anatomical deformities, fetal limb deformities are possible in cases such as this index patient, and this might result from prolonged pressure on the limbs due to a lack of space brought about by the septum and within the uterine horn where fetal development took place.[19]

A prospective study by Pang, et al[20] (n = 138) compared reproductive outcomes among women with septate uterus who underwent hysteroscopic resection versus those who did not. The authors concluded that while hysteroscopic septoplasty significantly improved pregnancy outcomes in women with a history of recurrent miscarriages, it did not show significant effect on reproductive outcomes among women with no history of poor pregnancy outcomes.

Given that the index patient does not have history of infertility, recurrent pregnancy losses, intrauterine growth restriction nor fetal limb deformities, the decision of the surgeons to leave the septum intact was acceptable. Had this been a case of recurrent preterm deliveries with unfavorable fetal outcomes, the authors would have leaned more towards recommending removal of the septum.

In contrast to the septate uterus, most patients with a bicornuate uterus have pregnancy outcomes close to those of the general population. Several reports have described successful term pregnancies, thereby rendering surgical correction unnecessary. A report by Moltot, et al[19] described a 28 year old G3P3 (2012) who, despite having bicornuate unicollis, was able to successfully deliver 2 live and grossly normal babies at term (G2 and G3). Namazi, et al[21], in their case report of a woman with bicornuate uterus, concluded that surgical correction of bicornuate uterus is only recommended in patients with a history of poor pregnancy outcome after other potential causes have been ruled out.

Osazuwa and Ejenobo[22] presented the antenatal management of a case of term twin gestation in a primigravid woman with bicornuate septate uterus, who delivered 2 live and healthy babies via Cesarean section. The patient described in their paper underwent prophylactic cerclage at 13 weeks age of gestation using the McDonald's technique, as an adjunctive procedure to minimize the risk of a miscarriage or preterm birth. Their patient was also given 2 courses of steroids at 28 and 32 weeks age of gestation.

Generally, surgical management of septate or bicornuate uterus is recommended for the following:[23] 1) patients with uterine septum and a history of: recurrent pregnancy loss after exclusion of other causes of recurrent pregnancy loss, or dysmenorrhea if medical therapy is not effective; 2) patients with bicornuate uterus and history of recurrent pregnancy loss, after exclusion of other causes of recurrent pregnancy loss.

Summary

In this case report, the authors explore the existing literature to gain insights into the characteristics, diagnosis, and treatment strategies for congenital malformations of the uterus. Overall, recognizing the potential implications of uterine malformations on reproductive health and implementing appropriate measures for diagnosis and management are essential steps in improving the care of patients with this conditions. An important aspect in management is the physician's and patient's shared decision whether or not surgical correction is needed. Patient education is significant as this can affect future pregnancies, since the adverse reproductive outcomes of such congenital anomalies dictates certain morbidity in the newborn.

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