Uterine didelphys with dicavitary twin gestation: A case report

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ABSTRACT

Uterine didelphys is a rare type of congenital uterine anomaly resulting from incomplete fusion of the paramesonephric ducts during embryogenesis. We report the case of a 27-year-old multiparous woman who presented with ovulation-induced dicavitary dichorionic diamniotic twins in known uterine didelphys. At 29 + 5 weeks of gestation, the patient had preterm prelabour rupture of membranes followed by threatened pre-term labour in the right uterus only, which settled with tocolysis. The pregnancy continued for a further 9 days, at which time uterine tightenings returned and the right cervix was fully dilated, resulting in successful vaginal delivery of the right twin. As a rare phenomenon, there is sparse literature on the management of dicavitary twin gestation in uterine didelphys. This case report adds to the evidence for independent functioning of uteri and cervices in cases of uterine didelphys which may enable interval delivery delay in this cohort with known increased risk of preterm birth.

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1. Background

Congenital uterine anomalies arise from abnormal embryological development of the paramesonephric ducts (or Müllerian ducts) [1]. Various anomalies result from arrested development of the uterovaginal primordium during the 8th week, by incomplete or failed development of parts of one or both paramesonephric duct, incomplete fusion of the paramesonephric ducts or incomplete canalisation of the vaginal plate [1]. Prevalence of all types of female congenital reproductive tract malformations is estimated at 4–7% and they are mostly benign [2]. The type and degree of anatomical distortion has associated health implications that may include reproductive failure, obstructed menses and inability to engage in sexual intercourse [2]. In pregnancy, uterine anomalies have been associated with recurrent miscarriage, placental abruption, intrauterine growth restriction, malpresentation, preterm delivery and increased rate of caesarean section [2].

The most widely accepted classification system is that of the American Society of Reproductive Medicine, which describes Müllerian anomalies anatomically [3]. Uterine didelphys is a rare type of anomaly with estimated prevalence of 0.3%, caused by failure of fusion of the inferior parts of the paramesonephric ducts resulting in separate uterine cavities with two cervices and a double or single vagina [1,4]. Uterine didelphys with dicavitary twin gestation is exceedingly rare: the reported incidence is 1 in 1,000,000 [5].

2. Case Presentation

A 27-year-old gravida-3 para-1 abortus-1 woman at 9 weeks of gestation presented to the outpatient clinic to establish her antenatal care. She had pre-pregnancy hysteroscopic results that demonstrated uterine didelphys, with two separate uteri each communicating with separate cervical canals such that no communication existed between the uterine cavities. She had a history of resection of a longitudinal vaginal septum. A normal renal tract had been documented. Her obstetric history was significant for a term vaginal singleton delivery 4 years prior, complicated by retained placenta and massive post-partum haemorrhage (PPH) (estimated blood loss of 4000 mL requiring blood transfusion, intensive care admission and prolonged length of stay), and subsequent pregnancy followed by an early spontaneous miscarriage.

Her background history included polycystic ovarian syndrome on regular metformin but she was otherwise fit and well with no previous abdominal surgeries, normal body mass index (BMI), non-smoker and no allergies.

The current pregnancy was an ovulation induced dichorionic diamniotic twin pregnancy with one fetus in each uterine cavity confirmed on early pregnancy dating ultrasound. Combined first-trimester screening returned low risk for fetal chromosome anomalies. Ultrasound at 16 weeks showed two cervical canals with a thin septation between them that were long and closed (3.1 cm and 3.2 cm). Subsequent regular ultrasound scans at 18, 22, 26 and 28 weeks demonstrated normal fetal morphology, normal and concordant fetal growth, normal placental blood flow and amniotic fluid levels. She presented to our birth unit with clinical history of premature

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prelabour rupture of membranes (PPROM) at 29 + 5 weeks of gestation, which was confirmed on vaginal speculum examination and oligohydramnios was seen on bedside ultrasound for twin 1 on the maternal right. Cephalic presentation was confirmed for both twins. A few hours later the patient developed irregular uterine tightenings in the right uterus only. She was otherwise well, with no clinical signs or biochemical evidence of infection. She was managed as an inpatient for PPROM initially with intravenous antibiotics, nifedipine tocolysis, betamethasone intramuscular steroid injections for fetal lung maturation and intravenous magnesium sulfate infusion for fetal neuroprotection. Uterine tightenings settled after 24 h and she was changed to oral antibiotics (erythromycin) and magnesium sulfate infusion was ceased. She was managed as an inpatient.

Nine days later, at 31 weeks of gestation, there was return of her uterine tightenings in both uteri that became more regular and painful. Vaginal examination revealed a fully dilated right cervix with fetal vertex at maternal ischial spines. Two large-bore intravenous cannulas were secured and she was transferred to operating theatre urgently for imminent vaginal delivery with known high risk of PPH and possible need for caesarean section for delivery of twin 2 in the left uterine cavity. Shortly thereafter, twin 1 on maternal right was spontaneously delivered vaginally in good condition, APGAR scores of 9 and 9 and weighed 1535 g. No oxytocics were given in attempt to prolong pregnancy for the second twin. Attempted physiological 3rd stage with gentle cord traction was unsuccessful; however, there was no excess bleeding. Pelvic ultrasound demonstrated twin 2 in left uterus and left cervix long and 1 cm open. Decision was made to leave placenta in situ in the right uterus and manage expectantly.

Approximately 24 h later, contractions in both uteri returned but were mild and irregular. On examination the left cervix was 5 cm dilated but with high fetal head. Fetal tachycardia seen on cardiotocography (CTG) combined with elevated inflammatory parameters (leucocytosis of 14.1 × 10^9/L with 79% neutrophilia, reference range 3.9–11.1 × 10^9/L and rise in C-reactive protein [CRP] 56 mg/L, reference range <3 mg/L) and in the context of known prolonged rupture of membranes and placenta in situ, chorioamnionitis was suspected. A decision was made for urgent caesarean section delivery of the twin in the left uterus. In lithotomy position a Pfannanstiel incision with lower uterine segment transverse hysterotomy of the left uterus was performed and the second twin was delivered cephalic with forceps in good condition, APGAR scores of 7 and 9 and weighed 1700 g. The first placenta in the right uterus was delivered complete vaginally. Intraoperatively there was PPH of 1000 mL secondary to uterine atony which necessitated blood transfusion for symptomatic anaemia (haemoglobin 75 g/L, reference range 115–165 g/L). She was continued on antibiotics until inflammatory parameters were down trending and her clinical condition improved. She was well and discharged from hospital 6 days after caesarean section.

3. Discussion

This case describes a rare presentation of twin pregnancy occupying separate uterine cavities in a woman with known uterine didelphys and illustrates the challenges involved in antenatal management and delivery planning.

A wide range of congenital uterine anomalies exist on a spectrum from very mild, such as arcuate uterus, to very significant aberrations, such as uterine didelphys, resulting from complete failure of paramesonephric duct fusion during embryogenesis [6]. Due to low prevalence, reproductive outcomes for women with uterine anomalies are often studied as a group rather than stratified by classification. The literature suggests that women with uterine anomalies are at increased risk of adverse pregnancy outcomes such as preterm birth, caesarean delivery due to fetal malpresentation and fetal growth restriction [7,8]. A systematic review by Chan et al. identified that canalisation defects, such as septate uteri, were associated with reduced conception rate and increased rate of first-trimester miscarriage when compared with unification defects such as arcuate, bicornuate and didelphys uterus [9].

However, data is mostly from singleton pregnancies and outcomes for twin gestations are mostly limited to case reports. Since twin pregnancies are already considered a high-risk cohort for most adverse pregnancy outcomes, it is not possible to discern from the literature whether adverse outcomes are related to uterine anomaly or twin pregnancy itself [6]. A small retrospective cohort study of twin pregnancies by Fox et al. concluded that patients with uterine anomalies had significantly worse pregnancy outcomes including rescue cerclage, preterm birth and lower birth weight but not fetal growth restriction [6]. However, these study findings are limited given very small sample size of only 17 patients with uterine anomaly, including only 1 patient with uterine didelphys.

Mode of delivery for women with congenital uterine anomalies should be discussed antenatally with consideration of previous pregnancy mode of delivery, multiple pregnancy, fetal presentation, other pregnancy related complications and/or pre-existing medical conditions and patient preference. The reported increased risk of caesarean section delivery is related to increased incidence of fetal malpresentation and fetal distress secondary to labour dystocia, rather than didelphys itself being an indication for caesarean section [10] and successful vaginal births have been previously reported in cases of uterine didelphys twin gestation [5,11]. A rare scenario described by Post et al. of uterine didelphys with dicovitary twin gestation required caesarean section delivery due to incomplete left cervical canal, therefore incompatible with vaginal delivery of the corresponding twin [10].

Given that the patient previously had a term vaginal singleton delivery, cephalic presentation of both twins and patient preference, it was considered suitable to aim for vaginal delivery for this pregnancy. However, the mode of delivery for the aftercoming twin was uncertain as the effect of the labour on the cervix of the non-presenting twin could not be predicted. As seen in this case and similarly previously reported case by Maki et al. it is possible for dicovitary twin gestations to have independently functioning uteri and cervixes [12]. In this case, the right-sided uterine gestation had PPROM, threatened preterm labour (TPL) with cervical dilatation while the left uterus remained apparently quiescent for several days even through vaginal delivery of the first twin. Delayed-interval delivery may be possible for cases of dicovitary twin uterine didelphys, in part owing to independent functioning uteri. This was seen in a case reported by Nohara et al. with a delivery delay of 10 weeks after emergency caesarean section for fetal distress at extreme prematurity, enabling the second twin to continue to 35 weeks of gestation [13], and again in a case of reported by Jan et al. with a 23-day interval delivery delay successful vaginal delivery of the second twin after PPROM and vaginal delivery of the first twin [14]. Furthermore, given this patient now has a uterine scar of the left uterus, it is appropriate to discuss implications of this for future pregnancies. Successful trial of labour after caesarean (TOLAC) with subsequent pregnancy in the contralateral uterine cavity (without scar) has been reported [15]. It is even possible to consider TOLAC, as seen in a case described by Wong, of successful vaginal birth after caesarean section with uterine didelphys with subsequent singleton gestation in the same uterine cavity [16].

4. Conclusion

Given the paucity of data on management and pregnancy outcomes, delivery planning for twin pregnancy with uterine anomaly should be considered high risk and individualised to each patient to optimise outcomes.

This case report adds to the literature for management of a very rare obstetric condition.

Contributors

Alison Laura King was the main author, conducted the literature review and drafted the manuscript.
Sarah Pixton was involved in the case, and reviewed and revised the manuscript.

Valeria Lanzarone was involved in the case, and reviewed and revised the manuscript.

All authors were responsible for the conceptualisation of the case report, saw and approved the final version of the paper and take full responsibility for the work.

Conflict of Interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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Patient Consent

Obtained.

Provenance and Peer Review

This case report was peer reviewed.

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