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Case Report

A Rare Case of a Twin Pregnancy in a 27 Year Old Patient with Uterus Didelphys

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Abstract

Uterus didelphys, or double uterus, is a congenital malformation of the female reproductive tract. It is estimated that between 0.5 - 6% of women may have a uterine anomaly. It occurs when the two Mullerian ducts fail to fuse, thus producing duplication of the reproductive structures. This is a case of a twin pregnancy in a 27-year-old patient with uterus didelphys. With obstructed hemivagina and ipsilateral renal agenesis. This is rare entity and referred to as Herlyn-Werner-Wunderlich syndrome (HWW). The patient was already diagnosed with uterus didelphys. Ultrasound imaging revealed fetus B had findings suspicious for ventriculomegaly with a lateral ventricle of 26.8 mm and oligohydramnios. Fetus A showed no anatomical abnormalities. This was a challenging caesarean section given the patients congenital uterine abnormality, further complicated by an almost fully dilated cervix in the first uterus. A transverse incision was made in the first uterus; a male infant was delivered cephalically with a moderate amount of clear liquor. The second uterus was incised; a female infant was delivered via forceps. Both placentas were delivered via controlled cord traction and each uterus was sutured in two layers. She was given prophylaxis for postpartum haemorrhage and Haematuria which resolved post-operatively prompted renal USS and CT urogram, which exhibited a right sided renal agenesis. Postpartum recovery was uneventful.

Keywords: *Twin Pregnancy; Uterus Didelphys; Congenital Malformation; Female Reproductive Tract*

Background

Uterus didelphys, or double uterus, is a congenital malformation of the female reproductive tract. It is estimated that between 0.5 - 6% of women may have a uterine anomaly. Uterus didelphys accounts for 8% of these. About one in 25,000 women with uterus didelphys gets pregnant with twins, one to each uterus [1]. The underlying aetiology of congenital müllerian defects is not well understood. Most of the defects are likely to be related to polygenic and multifactorial causes. It occurs when the two müllerian ducts fail to fuse, thus producing duplication of the reproductive structures. Generally, the duplication is limited to the uterus and cervix (uterine didelphys and bicollis [two cervices]), although duplication of the vulva, bladder, urethra, vagina, and anus may also occur. 15 - 20% of women with didelphic uterus also have unilateral anomalies, such as ipsilateral renal agenesis; the anomalies are on the right in 65% of cases. Obstetrical complications reported to occur more commonly with uterine anomalies include increased risks of miscarriage, prematurity, intrauterine growth restriction, antepartum and postpartum bleeding, cervical incompetence, abnormal fetal presentation, pregnancy-associated hypertension, and caesarean delivery [2]. Uterus didelphys has a term delivery rate of approximately 45%.

Purpose of the Study

We present a case of a twin pregnancy in a 27 year old patient with uterus didelphys. The report will discuss the epidemiology and complications of the condition and present an interesting case encountered in our unit.

Case Report

Case history

A 29 year old P2+1 with a known diagnosis of uterus didelphys presented for early pregnancy assessment at 8 weeks gestation. Obstetric history 1 previous spontaneous vaginal delivery at term. 1 previous caesarean section for breech presentation. 1 missed miscarriage at 6 weeks. 1st trimester ultrasound assessment revealed a viable singleton pregnancy in each uterus, each with a separate cervical canal.

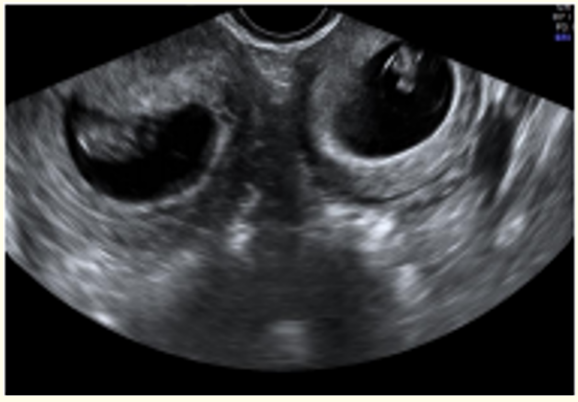

Image 1: *8 week scan showing two viable singleton pregnancies in each uterus.*

The patient was booked for consultant led antenatal care with regular ultrasound imaging every 4 weeks. She was assessed regularly throughout her pregnancy and had a relatively uncomplicated antenatal course. An elective LSCS was planned between 37 - 38 weeks 2nd trimester 20 weeks anomaly can: two well grown fetuses, no identifiable anomalies. Regular antenatal care: no concerns.

3rd trimester 35+4 weeks gestation: Ultrasound imaging revealed fetus B had findings suspicious for ventriculomegaly with a lateral ventricle of 26.8 mm (Image 2). Fetus A showed no anatomical abnormalities. A referral to fetal medicine was made .35+6: presented with spontaneous rupture of membranes and regular contractions. She was reviewed by the team on call, cervix was 6 cm dilated, other cervix not felt and a decision for an emergency caesarean section was made.

Outcome

Emergency caesarean section this was a challenging caesarean section given the patients congenital uterine abnormality, further complicated by an almost fully dilated cervix in the first uterus. A lower abdominal incision was made with routine dissection of the abdominal layers; both lower segments were clearly visualised. An adjoining septum was noted between the lower part of the uterus. A transverse incision was made in the first uterus; a male infant was delivered cephalically with a moderate amount of clear liquor. The second uterus was incised; a female infant was delivered via forceps. Of note, no liquor drained. Both placentas were delivered via con-

trolled cord traction and each uterus was sutured in two layers. Estimated blood loss was 2L. Post operative course haematuria which resolved postoperatively prompted renal USS and CT urogram, which exhibited a right sided renal agenesis. This had not been previously diagnosed. 15 - 20% of women with didelphic uterus have unilateral anomalies, such as ipsilateral renal agenesis; the anomalies are predominantly on the right side. Both infants were initially transferred to the neonatal unit for assessment. The female twin required oxygen and remained in the unit for observations. The male infant, with suspected ventriculomegaly, was assessed with cranial ultrasound which demonstrated no abnormalities. The patient was discharged home with her newborns on day 4 postnatal.

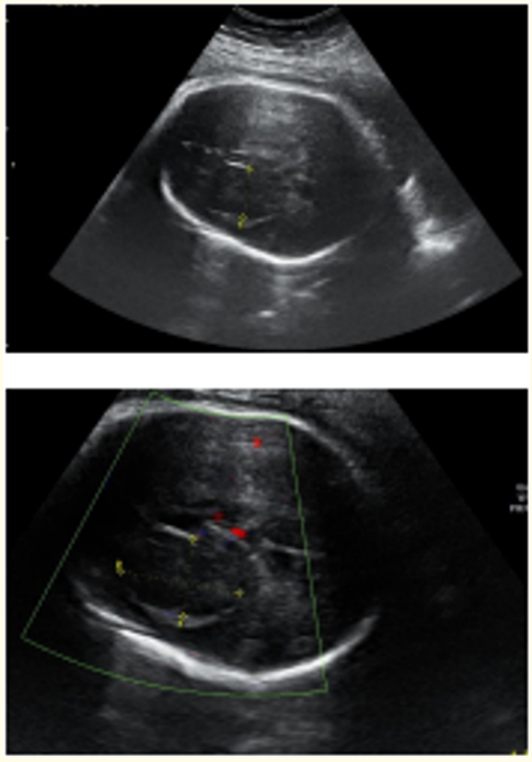

Image 2: *Fetus B showing an enlarged lateral ventricle.*

Discussion

Uterus didelphys is a rare congenital anomaly of the female reproductive tract and with twin pregnancy makes it one of the rare presentations. There are two uteri with two cervices, each has attached fallopian tube. A longitudinal vaginal septum may be associated in 75% of the cases. Other associated anomalies include vulva, urethra, bladder or anus [5]. Mullerian duct anomaly have been linked to fertility problems however it's not supported by strong evidence.

MRI is the best diagnostic method to identify these structural anomalies [4]. Most uterine anomalies are diagnosed by ultrasound and further details outlined by MRI. Uterus didelphys with two uterus attached appears heart shaped with two separate endometrial cavities inside [9]. Each uterus has endometrium, myometrium and serosal surfaces. Both uterus and cervices are fused together. Vagina may have longitudinal or transverse septum or obstructed hemivagina.

Most women with a didelphys uterus are asymptomatic, dyspareunia or dysmenorrhea may be the be the first presenting symptom.

Uterine abnormalities increase the risk of miscarriage due to implantation failure. Approximately 10% to 15% of recurrent miscarriage are associated with some type of uterine abnormality [7] resection of uterine septum to create single endometrial cavity is the surgical treatment option with varying of success rates [8].

A didelphys uterus may occur as a part of a syndrome called, Herlyn-Werner-Wunderlich (HWW) syndrome, it involves the triad of didelphys uterus, obstructed hemivagina, and ipsilateral renal agenesis [6].

Most women with related malformations need to be monitored closely throughout their pregnancy.

Accurate diagnosis is essential to determine the most effective management during childbirth. In our case at time of presentation only one uterus started labour with other cervix closed.

Women with uterine abnormalities have high risk of cesarean section which approaches 80% in case of mullerian duct anomalies [3].

Cesarean section appears safer for women with uterus didelphys, however safer vaginal delivery is also documented in some cases. An extensive analysis of more complex twin cases with uterus didelphys is needed to establish safer delivery choices especially in complex scenarios where two uterus may start labour separately. Twins in two separate uterus may labour spaced apart many hours and upto 8 weeks as mentioned in few case reports [10]. Anatomically, two uterus in this anomaly appear similar in structure but may function as two separate units [11].

Conclusion

Congenital Müllerian anomalies are challenging case scenarios for obstetrician-gynecologists. It may be associated with fertility issues or pregnancy failures. Women need meticulous care and monitoring throughout pregnancy and labour and sonographic surveillance of twin gestation for growth and cervical assessment.

Didelphys uterus is not an indication for cesarean delivery, however this is a customized decision based on maternal or fetal co morbidities or presence of thick vaginal septum. Ultrasound and MRI should be used to investigate renal anomalies and to rule out Herlyn-Werner-Wunderlich (HWW) syndrome.

Bibliography

1. P K Heinonen. "Uterus didelphys: a report of 26 cases". *European Journal of Obstetric Gynaecological Reproduction* 17.5 (1984): 345-350.

2. NE Nykiforuk. "Uterus Didelphys". *Canadian Medical Association Journal* 38.2 (1938): 175.

3. Pankaja S., *et al*. "Successful Pregnancy with Uterus Didelphys". *Journal of Andrology and Gynecology* 4 (2016): 3.

4. Fukunaga T., *et al*. "The spectrum of imaging appearances of müllerian duct anomalies: Focus on MR imaging". *Japanese Journal of Radiology* 35.12 (2017): 697-706.

5. Laufer MR and De Cherney AH. "Clinical Manifestations and Diagnosis of Congenital Anomalies of the Uterus". UpToDate Waltham, MA, USA: Wolters Kluwer South Holland, The Netherlands (2019).

6. S Suthar., *et al*. "Rupture uterus in pregnancy with didelphys uterus: a rare case report". *Journal of South Asian Federation of Obstetrics and Gynecology* 3.3 (2011): 149-150.

7. Lee RM and Silver RM. "Recurrent pregnancy loss: summary and clinical recommendations". *Seminars in Reproductive Medicine* 18.4 (2000): 433-440.

8. Propst AM and Hill JA. "Anatomic factors associated with recurrent pregnancy loss". *Seminars in Reproductive Medicine* 18.4 (2000): 341-350.

9. Salem S and Wilson SR. "Gynecologic ultrasound". In Rummack CM, Wilson SR, Charboneau JW (eds): Diagnostic Ultrasound. 3rd edition. St. Louis, MO, Mosby (2005): 534-537.

10. Brown O., *et al*. "A Twin pregnancy in a uterus didelphys". *Journal of Obstetrics and Gynaecology* 19.1 (1999): 82-83.

11. Makoto N., *et al*. "Twin pregnancy in each half of a uterus didelphys with a delivery interval of 66 days". *BJOG: An International Journal of Obstetrics and Gynaecology* 110.3 (2003): 331-332.

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