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Case Series

Preterm Delivery in a Nullipara with Undiagnosed Uterine Didelphys: A Case Report and Review of the Literature

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ABSTRACT

Uterine didelphys is a rare congenital abnormality which results from complete failure of fusion of the Mullerian duct during embryological phase of life. In pregnancy, uterine didelphys is commonly associated with adverse foetal outcome. We report a 25-year-old nullipara with previous history of miscarriages who presented at 33 weeks' gestation with complaints of preterm contraction with associated urinary symptoms. She was managed for urinary tract infection in pregnancy and symptoms resolved. Obstetric scan showed normal findings with no abnormality of the uterus or adjacent structures. She subsequently had Caesarean section for nullipara breech in labour with delivery of a live baby and an incidental intra-operative finding of uterine didelphys. Mother and baby were discharged home healthy on the 4th post-operative day and postpartum period was unremarkable. The rare incidence of this finding and the good perinatal outcome are the peculiarity of this report.

Keywords: Preterm, Urinary tract Infection, Uterine Anomaly, Uterine Didelphys

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INTRODUCTION

The uterus is formed during embryogenesis by fusion of the two paramesonephric ducts (Mullerian ducts) into a single uterine body.[1] The development, fusion, canalization or reabsorption of the Mullerian ducts normally occurs between 6 and 22 weeks in utero.[1,2] Abnormal embryological development of the Mullerian duct results in Mullerian duct anomalies [MDAs] which are congenital defects of the female genital system, some of which are septate uterus, bicornuate uterus, arcuate uterus and uterine didelphys.[1–3]

Uterine didelphys results from failure of fusion of the Mullerian duct which can be complete or partial, giving rise to various anatomical descriptions; with double uterine cavity, double cervix and double vagina or with double uterine cavity, double cervix and a single vagina or with double uterine cavity, single cervix and single vagina .[1,3] These may be associated with renal and skeletal anomalies. [1,4,5,6] The Genetic syndrome associated with this anomaly is known as Herlyn-Werner-Wunderlich (HWW) syndrome, also known as obstructed hemivagina and ipsilateral renal anomaly

(OHVIRA), and rarely have associated cardiac defects.[1,4,5,6]

The American Society of Reproductive Medicine (ASRM) 2021 classification of uterine didelphys is as shown in Figure 1.[7]

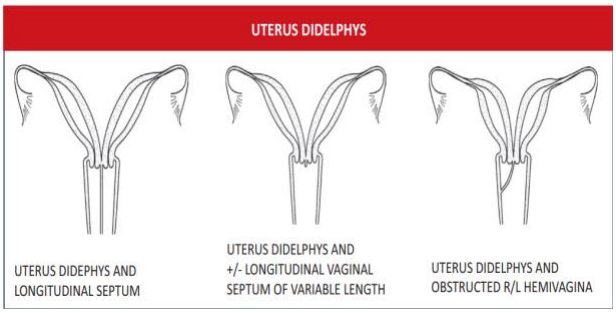

Figure 1 (photo credit ASRM): American Society of Reproductive Medicine in 2021 classification of uterine didelphys.[7]

Uterus didelphys is a relatively rare type of congenital malformation of the female genital tract.[2] The incidence globally ranges from 1% to 10% of women population.[2] It is challenging to know the exact occurrence of this anomaly, as it may go undetected when there are no medical and reproductive complications. The commonest uterine anomaly is septate uterus with a mean incidence of ~35% followed by bicornuate uterus (~25%) and arcuate uterus (~20%).[8,9]

Uterine didelphys is asymptomatic in most women.[8,9] However, it may be associated with dyspareunia, dysmenorrhea and infertility.[2,8–10] The degree of the dysmenorrhea and dyspareunia are dependent on the varying degree of longitudinal vaginal septum.[11] The obstetric complications include recurrent pregnancy loss, premature delivery, malpresentation, intrauterine growth restriction (IUGR), placental abruption, cervical insufficiency, spontaneous abortion, PROM, premature labour, Caesarean delivery due to breech presentation, and decreased live births.[8,9,12,13]

In this case report, we discuss a rare case of undiagnosed didelphys uterus in pregnancy delivered at our facility.

CASE REPORT

A 25-year-old Nigerian tertiary student who was referred from a primary health facility to the University of Benin Teaching Hospital (UBTH) as a case of preterm contractions for expert care. She was 33 weeks pregnant. She had presented at the referring health facility with complaint of abdominal pains of 3 days which was intermittent, sharp, non-radiating, worse on both flanks and transiently relieved by analgesics.

There was associated urinary frequency, nocturia, dysuria and low-grade fever, but no history of abnormal vaginal discharge, liquour drainage or bleeding per vaginam. There was history of spotting per vaginam (threatened miscarriage) in the first trimester for which she was managed with bed rest and haematinics. Symptoms resolved and she got better. She was not a known hypertensive, diabetic or patient with sickle cell disease, and had no known systemic illness. She had 2 previous spontaneous first trimester miscarriages which were complete with no complication.

She presented at the referring facility where she had registered her pregnancy at 16 weeks gestation with the aforementioned complaints. Following examination, a diagnosis of preterm contraction was made and she was referred to UBTH for further evaluation and access to neonatal care in anticipation of possible preterm delivery.

Her booking laboratory investigations were adjudged normal. She had 2 ultrasound scans done at 8 weeks, 20 weeks gestation and both reported an essentially normal pregnancy. She continued her routine medications as prescribed; tetanus immunization and haematinics.

At presentation in UBTH, she was a young woman in painful distress, not pale, febrile with temperature of 37.8 degrees Celsius, anicteric, acyanosed, not dehydrated with no pedal oedema. Her respiratory rate was 22 cycles per minute, not dyspnoeic and she was not in any respiratory distress. Her pulse rate was 84bpm, blood pressure was 120/70mmHg and heart sounds were S1 S2 only. The abdomen was enlarged, about 32 weeks size, no abdominal tenderness but there was left renal angle tenderness. Pelvic examination was essentially normal. Her full blood count showed elevated white blood cell count (leucocytosis) 14,200/ul, haemoglobin was 9.6g/dl, the haematocrit was 30%, and platelet count was 152,000/ul. Blood film for malaria parasite was negative. The urinalysis was positive for leukocytes and nitrites. The urine Microscopy showed numerous pus cells and the urine culture yielded growth of Escherichia coli which was sensitive to amoxicillin/ clavulanate. Results of other investigations performed were essentially normal. She was given analgesic, intravenous co-amoxiclav 625mg 8hourly for 72hours, intramuscular dexamethasone 12mg every 12 hours for 24hours, and placed on bed rest. Abdominal pain and fever resolved after 48 hours.

On the 6th day on admission, she had a repeat abdominal pain but no fever. Examination showed presence of uterine contractions of at least 2 in 10 minutes, of moderate intensity, lasting 35 seconds. A pelvic examination revealed 2cm dilated cervix, no liquour drainage and no bleeding. Urgent ultrasound scan done revealed a single viable breech foetus. A diagnosis of a nullipara breech in labour was made. She was counselled on the findings and the need for delivery by Caesarean section to which she consented. Packed Cell

Volume (PCV) was 33%. Two units of whole blood were Grouped and Crossed Matched for her.

She had a Lower Segment Caesarean section. Intra-operative findings (Figures II, III, & IV**)** were two distinct uteruses with separate uterine cavities both connecting to a common cervix down into the vaginal canal. A live female 2.2Kg neonate with Apgar Scores 8 (1 min) - 9 (5min), 0.5 kg placenta and membranes were delivered from the left uterus through the lower segment transverse incision. The right uterine cavity showed decidualization.

The incision made across the 2 uteruses was repaired in 2 layers, and rectus sheath and anterior abdominal closure was done as routine. Estimated blood loss was 350ml. She continued her postoperative antibiotics and haematinics. Her post-operative recovery was uneventful, her PCV was 31% on the 2nd day post operation. Baby at delivery was reviewed by the paediatrician who was in attendance and certified fit to be nursed with the mother when fully recovered. Baby tolerated oral intake and remained well with the mother. She was discharged with her baby 5 days post operation after counselling on need for neonatal immunization.

She and her baby were seen 6 weeks postpartum at the postnatal clinic. There was no complaint, mother and baby were healthy. The baby was being breast fed and weighed 5Kg. She was counselled about the incidental operative findings and possible challenges as seen in previous pregnancies, and the need for correction in the future. She was also counselled on various contraception methods (except for intrauterine device) and was discharged from the post natal clinic.

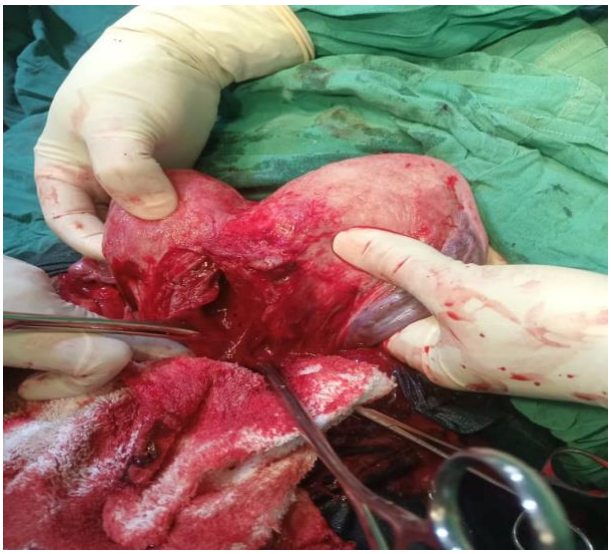

Figure II: Intraoperative view showing uterine didelphys following Caesarean section (foetus was located in the left uterine cavity)

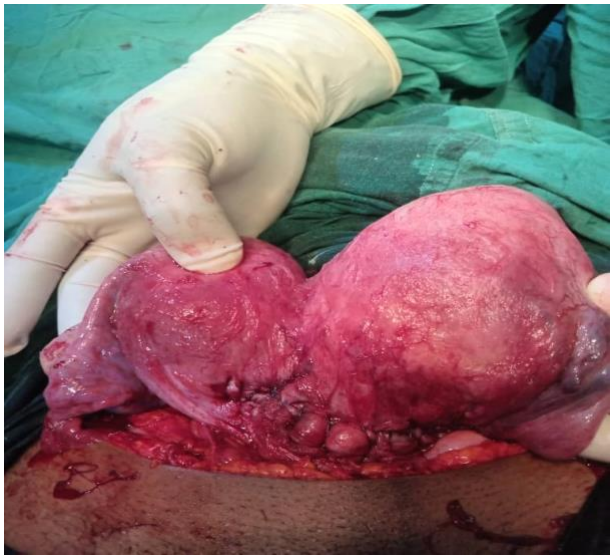

Figure III: Intraoperative view showing uterine didelphys after repairing the uteruses

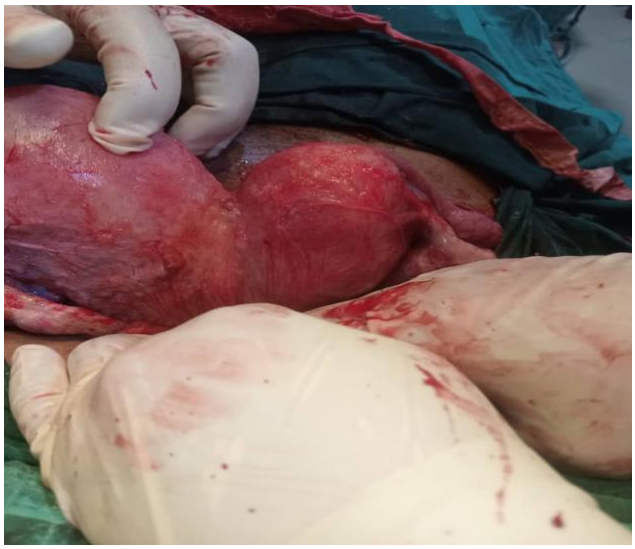

Figure IV: Intraoperative view showing uterine didelphys (Posterior view)

DISCUSSION

Uterine didelphys is a rare congenital malformation of the Mullerian duct.[2] It is characterized by double uterine cavity, cervix and in some cases the vagina.[1,3] Each uterus has one fallopian tube and ovary.[2,13] Uterine didelphys is associated with poor obstetric outcomes and remains a challenge to the obstetrician, especially when it is undiagnosed before onset of labour. The complications noted in this patient were premature delivery, preterm labour, malpresentation with Caesarean delivery of a breech presenting foetus. Other

possible complications are dysmenorrhea, dyspareunia, intrauterine foetal growth restriction, recurrent pregnancy losses, and premature rupture of membranes.

Pre-pregnancy, an excellent non-invasive investigation for the diagnosis is a 3-D transvaginal sonography with vaginal examination usually with the findings of any of the following: a longitudinal vaginal septum and two vaginal openings with two cervices, two cervices with a vaginal opening, or one cervix and a vaginal opening.[13] Others methods of investigation include sonohysterography, hysterosalpingography, hysterolaparoscopy and pelvic magnetic resonant imaging.[13] However, none of these pre-pregnancy diagnostic investigations were done for this patient.
Detection of uterine anomalies in early pregnancy is very important because its association with foetal anomaly will help inform on the extent of counselling as regards termination of pregnancies in congenital anomalies not compatible with life or help to improve pregnancy outcome where decision to continue is made in cases of compatibility with life.

In the case reported, the pre-operative diagnosis of uterine didelphys was not made in spite of the three-ultrasound done at various stages of pregnancy. The delay in diagnosis and missed diagnosis at Ultrasonography also common to other African and developing countries but less common in developed countries due to sophisticated tool for ultrasonography and better training. Pathan et al in India reported a case of uterine anomaly which was undiagnosed during antenatal period; this anomaly was only noted intraoperative at Caesarean Section.[6] Similar cases were reported by Okafor in Enugu Nigeria, Gudu et al in Ethiopia and Ojurongbe et al in Ogbomoso, Nigeria.[12–14] In contrast to this, the report by Golawki et al in Poland showed early diagnosis which helped in better planning for delivery and prognosis.[15] The reported reason for missed diagnosis may be the level of skill, experience, and index of suspicion in view of the history of recurrent miscarriages and preterm labour of the sonographer. This is likely due to poor training of sonographers in uterine anomaly scanning in Nigeria, other contributory factors may be the huge financial implications of acquiring such skills from international providers in Europe and America. This statement is without prejudice to the fact that it is a rare condition and may not have been experienced by the sonographer. The diagnosis in this case was made at Caesarean section.

Studies have shown increased rate of premature deliveries in women with uterine didelphys.[16,17] This is so with our case, that had preterm contractions at 33 weeks and was delivered at 35 weeks by Caesarean section for breech in labour. In a study by Heinonen, most pregnancies (76%) were located in the right uterus compare to the left.[15] However; in our case, the pregnancy was located in the left uterus.

There has been a case of adverse foetal outcome as reported by Okafor et al in Enugu Nigeria where they had intrauterine fetal death from failed induction of labour in a woman with undiagnosed uterine didelphys which necessitated an emergency caesarean delivery.[13] This may be due to lack of availability of modern diagnostic technique which may have caused the delay in establishing diagnosis, and thus delayed prompt interventions that could have averted the adverse foetal outcome.

The definitive management of uterus didelphys is surgical correction (Strassmann's metroplasty) which is indicated only for symptomatic patients with severe dyspareunia and recurrent pregnancy losses.[8,12] Metroplasty was not done in our case because she was asymptomatic and we decided not to complicate her surgery in the scenario of an incidental finding but would rather plan for corrective surgery in a non-pregnant state.

CONCLUSION

Uterine didelphys is a rare congenital anomaly often undiagnosed in the antenatal period because it may be asymptomatic, and when symptoms are found, they are often non-specific. Reports of poor obstetric outcomes have been associated with the condition. The successful obstetric management of this undiagnosed case until at Caesarean section does not preclude the need for astute approaches in the evaluation and care of pregnancy of this rare entity, to mitigate adverse outcomes. Our recommendation to physicians, sonologist and other skilled birth personnel is to consider rare conditions such as uterine anomalies during evaluation of patients presenting with preterm contractions with a background history of recurrent miscarriages.

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