Case Reports in Women's Health 22 (2019) e00118

Contents lists available at ScienceDirect

Case Reports in Women's Health

journal homepage: www.elsevier.com/locate/crwh

Twin gestation in a uterus didelphys with only one functional cervix: A case report

Rebecca J. Post *, Claire L. Templeman, Richard M. Benoit

Department of Obstetrics & Gynecology, Kaiser Permanente Los Angeles Medical Center, Los Angeles, CA, United States of America

ARTICLE INFO

Article history:
Received 5 April 2019
Received in revised form 21 April 2019
Accepted 25 April 2019

Keywords:
Twin gestation
Uterus didelphys
Hypoplastic cervix
Mullerian duct anomalies
Cervical anomalies

ABSTRACT

Introduction: Twin gestation in a uterus didelphys with one fetus in each uterine cavity is rare and presents unique challenges in antepartum and intrapartum care.
Case Presentation: A 35-year-old woman with a uterus didelphys became pregnant with twins, with one fetus in each uterus, after intrauterine insemination of a single visible cervix. Multiplanar ultrasonography showed the presence of one complete cervix and a second hypoplastic cervix; it was unclear whether she could deliver both twins vaginally. Her pregnancy was complicated by fetal growth restriction of twin B. At 38 weeks, the patient underwent scheduled cesarean section and delivered two viable twins.
Conclusion: Determining the precise anatomy of Mullerian duct anomalies, including the cervix and vagina, is important for obstetrical management.

1. Introduction

Failure of the Mullerian ducts to fuse during embryonic development can result in a wide spectrum of female reproductive tract malformations, ranging from a minor variation in uterine shape to complete absence or duplication of the uterus, cervix, and vagina. Mullerian duct anomalies have been associated with increased risk of infertility, pregnancy loss, and adverse pregnancy outcomes depending on the extent of abnormality [1]. The American Society for Reproductive Medicine (ASRM) created the most widely known classification system for Mullerian duct anomalies in 1988; however, it is almost entirely limited to describing uterine abnormalities. Thus, the European Society of Human Reproduction and Embryology and the European Society for Gynaecological Endoscopy (ESHRE/ESGE) proposed a new classification in 2013, which accounts for variations of cervical and vaginal anatomy [2]. Identifying the exact nature of a patient's anomaly is crucial to understanding the potential impact on reproductive health.

The uterus didelphys is one of the least common anomalies, estimated to occur in 1 in 1000 women, and is classically defined as the presence of two uteri, two cervixes, with or without a longitudinal vaginal septum, though other variations exist [1,3]. Twin gestation with a fetus occupying each uterus has been reported in a small number of case studies. Simultaneous pregnancies may pose some unique obstetrical issues when two complete uteri can contract independently and can have different rates of cervical dilation [4]. Management decisions such as determining feasibility of vaginal delivery or the surgical approach to cesarean delivery is unclear and left to the discretion of the obstetrician.

This case report discusses a patient with an exceedingly rare situation – a twin pregnancy occurring in a uterus didelphys, with one twin in each uterine cavity. However, this case is further unique in that only one cervix was visible on pelvic exam. Whether both twins could deliver vaginally or how a pregnancy occurred in the left uterus without an identifiable cervix was initially unclear. The issues surrounding the evaluation of Mullerian duct anomalies in pregnancy and their implications during labor and delivery are discussed.

2. Case Presentation

A 35-year-old woman was found to have a Mullerian duct anomaly during investigation of her primary infertility. Magnetic resonance imaging showed she had a uterus didelphys with two uteri and two cervixes, as well as an absent left kidney. However only one cervix was identified on pelvic exam. Hysterosalpingogram (HSG) was performed by injecting dye into the single cervix, resulting in filling of the right uterine cavity and spillage from the right fallopian tube (see Fig. 1). With an uncertain diagnosis of her Mullerian duct anomaly, the patient underwent several cycles of controlled ovarian hyperstimulation and intrauterine insemination (IUI) through the single identifiable cervix. The patient achieved pregnancy after the fourth IUI cycle using a regimen of 5 days of clomiphene (at 100 mg/day) followed by 9 days of human menopausal gonadotropin (8 days of 2 ampules and 1 day of 3

* Corresponding author at: Kaiser Permanente Los Angeles Medical Center, Department of Obstetrics & Gynecology, 4900 Sunset Blvd, 5th Floor, Los Angeles, CA 90027, United States of America.
E-mail addresses: rebecca.j.post@kp.org (R.J. Post), claire.l.templeman@kp.org (C.L. Templeman), richard.m.benoit@kp.org (R.M. Benoit).

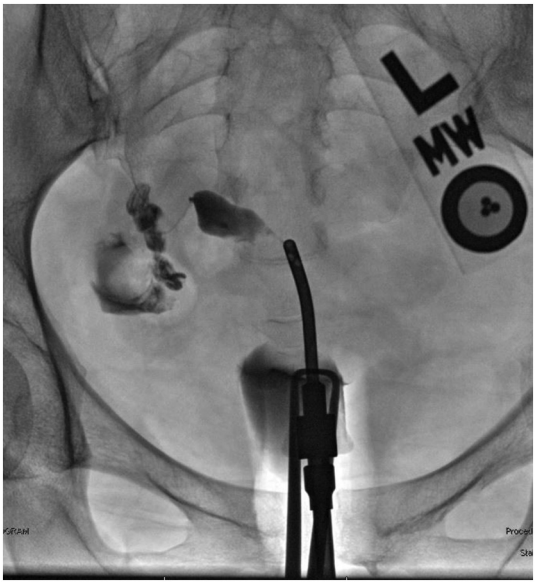

Fig. 1. Hysterosalpingogram showing filling of right uterus and spillage from the right fallopian tube.

ampules) while monitoring the number and develop of ovarian follicles using serial transvaginal ultrasound scans. Once 1–2 potential mature follicles were identified in the right ovary, ovulation was trigged with human chorionic gonadotropin injection, and IUI was subsequently performed. Ultrasound at 8 weeks of gestation revealed the presence of dicavitary twins, with one fetus situated in each uterus.

The patient was followed by the perinatology team and was monitored throughout the pregnancy with serial growth ultrasound scans and non-stress testing due to her unusual uterine anatomy. The right twin developed growth restriction at 37 weeks, with an estimated fetal weight at the fifth percentile for gestational age but with normal fetal Doppler velocimetry. Both twins remained in cephalic presentation at term (see Fig. 2). Delivery was recommended around 38–39 weeks, given twin gestation with fetal growth restriction. Several discussions were had with the patient regarding route of delivery since it was unclear whether the left uterus had a cervix due to the discrepancy between her prior imaging and pelvic exam. Detailed three-dimensional multiplanar ultrasound was performed and confirmed the presence of two cervixes; however, the left cervical canal appeared to be incomplete without any communication to the vagina (see Fig. 3a–f). A small communication was identified between the right and left cervical canals, as shown in Fig. 3f. Thus, it was uncertain whether the left twin would be able to be delivered vaginally. The patient elected to proceed with cesarean delivery.

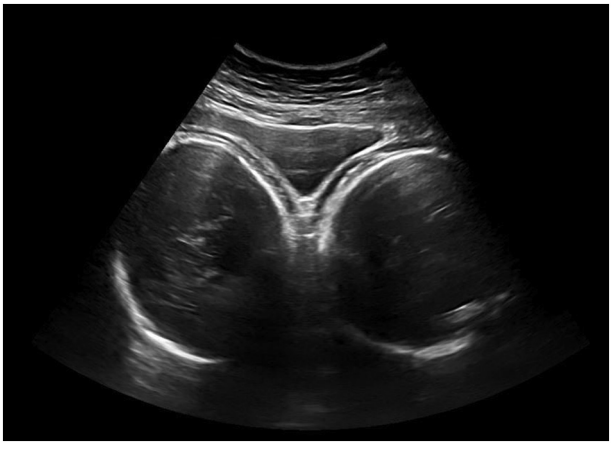

Fig. 2. A two-dimensional ultrasound image showing twin gestation with one fetus in each uterus at 32 weeks, both twins in cephalic presentation.

At 38 and 6/7 weeks of gestation, the patient underwent primary cesarean section with a low midline vertical skin incision to maximize exposure. The left uterus was found to be more anterior, and so the left twin was delivered first (twin A), followed by the twin in the right uterus (twin B) via bilateral low transverse uterine incisions. An intraoperative exam revealed a complete uterus didelphys (see Fig. 4). The right uterus was confirmed to have a complete cervical canal while the cervix of the left uterus was found to end in a blind pouch. Thus, the patient's uterine anomaly was confirmed to be ESHRE/ESGE Class U3b-C3-V0. Estimated blood loss was 1.6 L, mostly attributed to bleeding from the bilateral hysterotomies, but she did not require blood transfusion. Twin A weighed 3195 g with Apgar scores of 8 and 9, and twin B weighed 2705 g (sixth percentile for gestational age) with Apgars of 8 and 9. The postoperative course was uncomplicated, and both twins were discharged with their mother two days after delivery.

3. Discussion

This case involved a rare scenario of twin gestation in a didelphys uterus with each fetus occupying a separate uterine cavity and illustrates the challenges of diagnosing and managing pregnancies with uterine anomalies. Mullerian duct anomalies include a variety of anatomical defects, from a minor groove in the uterine fundus to complete duplication of the female genital tract. Yet, most of the literature examining pregnancy outcomes with uterine anomalies group all of the different classes together and compare them to pregnancies with a normal uterus. Few studies have investigated the clinical significance of each specific Mullerian duct anatomic defect due to their low prevalence [5]. One of the largest studies examining patients with uterus didelphys specifically was a retrospective cohort study following 49 patients over an average of 9.1 years; 94% of these women had at least one pregnancy (all singleton gestations). The complications reported were spontaneous abortion (21%), pregnancy-induced hypertension (13%), preterm delivery (24%), breech presentation (51%), fetal growth restriction (11%), and cesarean delivery (84% due to malpresentation or labor dystocia) [3].

Multifetal gestation in a uterus didelphys has been reported in only a small number of case studies. Most of these women were delivered by cesarean section either electively or due to anatomical obstruction or fetal distress; however, successful spontaneous vaginal deliveries of both twins have also been reported [6,7]. Thus, the presence of a uterus didelphys is not an indication itself for cesarean delivery but carries a higher likelihood of cesarean delivery for malpresentation or for labor dystocia reasons. Dicavitary twin gestations are further complicated since the two uteri and two cervixes can function independently. Maki et al. reported a case of twins in a double uterus and demonstrated independent contractions in the two uteri for approximately 90% of labor on tocometry [4]. Nohara et al. described a patient with uterus didelphys and dicavitary twins, complicated by preterm rupture of membranes in the left uterus with subsequent contractions. Due to fetal distress, the left twin was delivered by emergent cesarean section at 25 weeks gestation. The right uterus remained quiescent until 35 weeks and delivered the second twin via spontaneous vaginal delivery [8].

The diagnosis of Mullerian duct anomalies is not always easily discernible and may require utilizing multiple imaging modalities or more invasive procedures such as hysterosalpingography, hysteroscopy, laparoscopy, or laparotomy. Furthermore, uterine anomalies have been reported that do not belong to any of the traditional ASRM classifications, such as a uterus didelphys with one cervix. More recently, three-dimensional mutliplanar ultrasonography has been utilized as a noninvasive method to determine uterine anatomy with accurate results [1]. Multiplanar ultrasound proved useful for our patient case, as it not only confirmed the presence of two cervixes, but

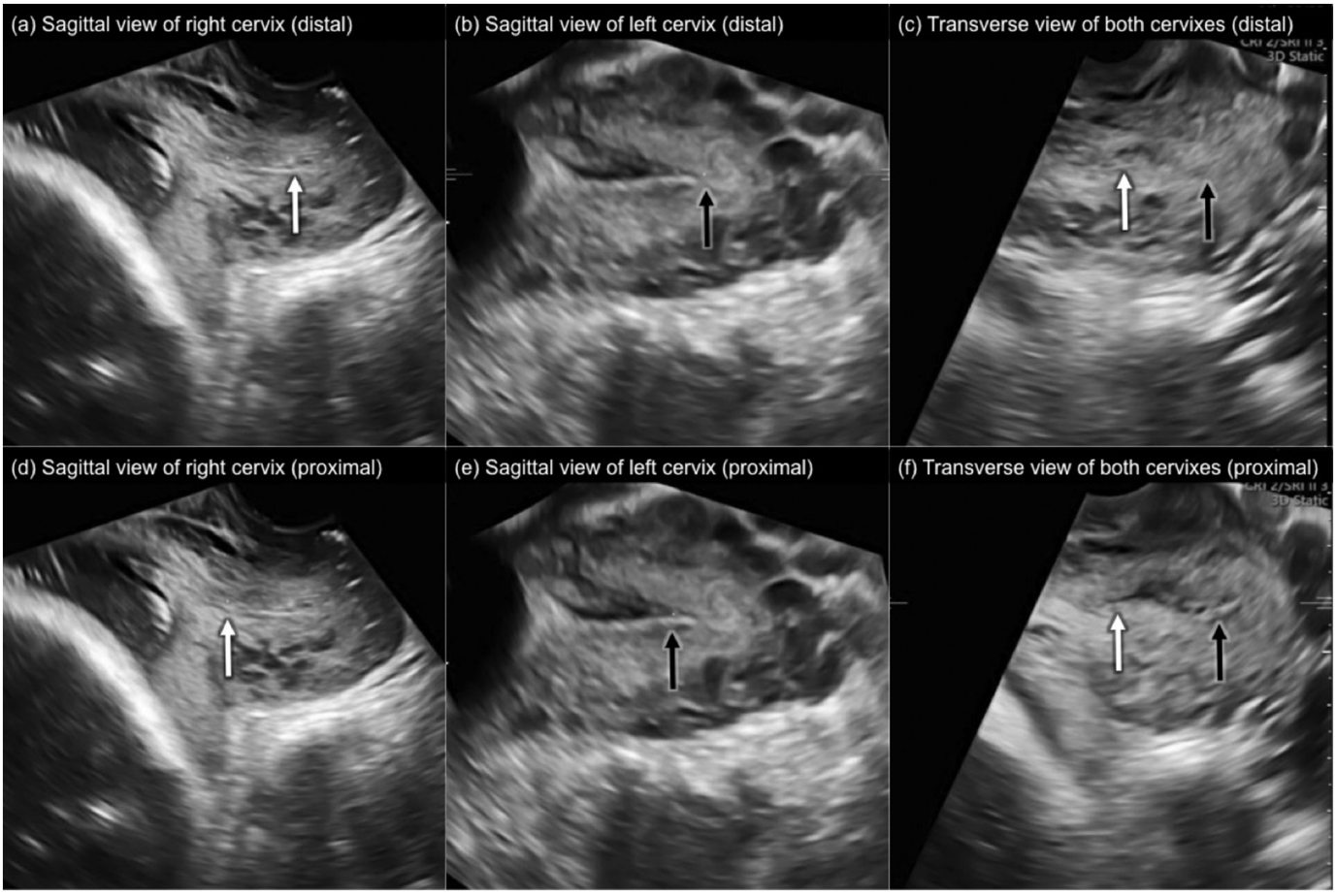

Fig. 3. Multiplanar images obtained using three-dimensional ultrasound of the two cervixes. Fig. a–b–c show the simultaneous sagittal and transverse views of the distal portions of the cervixes: the right uterine cervix (as shown by white arrows) has a complete cervical canal, while the left uterine cervix (as shown by the black arrows) has an incomplete cervical canal without an apparent communication to the vagina. Figures d–e–f show the simultaneous sagittal and transverse views of the proximal portions of the cervixes: the right uterine cervix (white arrows) and left uterine cervix (black arrows), with a small communication between their cervical canals (Fig. 3f).

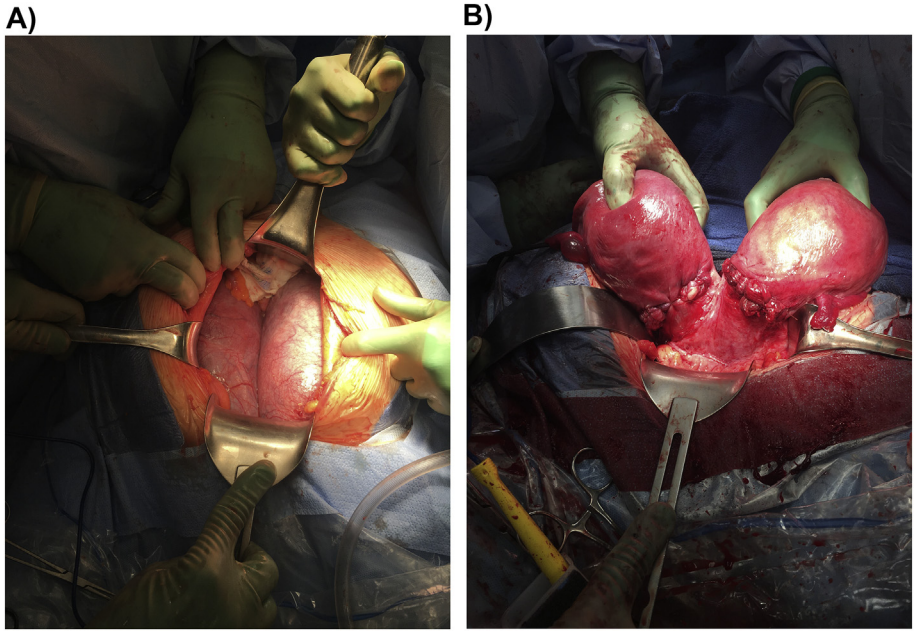

Fig. 4. Intraoperative photos of uterus didelphys during cesarean section: (a) upon abdominal entry, and (b) after repair of the bilateral hysterotomies.

also revealed the second cervix to be incomplete with no apparent communication to the vagina. Based on this information, cesarean delivery was recommended since it was unlikely the left twin could be delivered vaginally with a hypoplastic cervix.

How the patient became pregnant in both uteri when intrauterine insemination was only performed through the right cervix was initially a mystery. Given only the right ovary had 1–2 potentially mature follicles seen on transvaginal ultrasound, the patient most likely ovulated twice from the right ovary, with one oocyte entering the right fallopian tube, and the other oocyte entering the left fallopian tube. But this does not explain how semen was able to enter the left uterus. The HSG suggested there was no communication between the uteri, as injection of the dye through the right cervix resulted in filling of the right uterus only. However, multiplanar ultrasound identified a communication between the right and left cervical canals, which could provide a route for the semen injected into the right uterus to reach the left uterus. If the HSG catheter was placed above the level of this cervical connection, it is possible it could have been missed on HSG imaging. The presence of the communication between the right and left cervical canals would also explain how this patient did not develop hematometra during menses.

This patient case involved an unusual situation of two fetuses occupying two separate uterine cavities with only one functional cervix, demonstrating the diversity of Mullerian duct anomalies. Confirming the precise anatomy of the entire female genital tract, including the uterus, cervix, and vagina, is essential to predicting potential obstetrical outcomes. Multiple imaging modalities may be required for diagnosis when maternal anatomy is ambiguous. Three-dimensional multiplanar ultrasound was particularly useful in our case. Further studies are needed to investigate the clinical significance of each anatomical variation.

Contributors

Rebecca J. Post managed the patient, conducted background research, and was involved in drafting and revising the manuscript.

Claire L. Templeman managed the patient and was involved in revising the manuscript.

Richard M. Benoit contributed to the conception of the study and was involved in revising the manuscript.

Conflict of Interest

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

Funding

The authors received no financial support for the research, authorship, or publication of this article.

Patient Consent

Obtained.

Provenance and Peer Review

This case report was peer reviewed.

Acknowledgements

We would like to thank our colleagues Amy Dhesi, MD, Marsha Baker, MD, and Aline Ketefian, MD, from the department of Reproductive Endocrinology & Infertility at Kaiser Permanente Los Angeles Medical Center, for providing their insight into this patient's infertility workup and treatment.

References

[1] B. Bhagavath, E. Greiner, K.M. Griffiths, T. Winter, S. Alur-Gupta, C. Richardson, et al., Uterine malformations: an update of diagnosis, management, and outcomes, Obstet. Gynecol. Surv. 72 (6) (2017) 377–392.
[2] G.F. Grimbizis, S. Gordts, A. Di Spiezio Sardo, S. Brucker, C. De Angelis, M. Gergolet, et al., The ESHRE/ESGE consensus on the classification of female genital tract congenital anomalies, Hum. Reprod. 28 (8) (2013) 2032–2044.
[3] P.K. Heinonen, Clinical implications of the didelphic uterus: long-term follow-up of 49 cases, Eur. J. Obstet. Gynecol. Reprod. Biol. 91 (2) (2000) 183–190.
[4] Y. Maki, S. Furukawa, H. Sameshima, T. Ikenoue, Independent uterine contractions in simultaneous twin pregnancy in each horn of the uterus didelphys, J. Obstet. Gynaecol. Res. 40 (3) (2014) 836–839.
[5] N.S. Fox, A.S. Roman, E.M. Stern, R.S. Gerber, Rebarber A. Saltzman, Twin pregnancy in patients with a uterine anomaly, J. Matern. Fetal Neonatal Med. 27 (4) (2014) 360–364.
[6] A. Tyagi, B. Minocha, S. Prateek, Delayed delivery of second twin in uterus didelphys, Int. J. Gynaecol. Obstet. 73 (3) (2001) 259–260.
[7] H.N. Al Yaqoubi, N. Fatema, Successful vaginal delivery of naturally conceived dicavitary twin in didelphys uterus: a rare reported case, Case Rep. Obstet. Gynecol. (2017), 7279548.
[8] M. Nohara, M. Nakayama, H. Masamoto, K. Nakazato, K. Sakumoto, K. Kanazawa, Twin pregnancy in each half of a uterus didelphys with a delivery interval of 66 days, BJOG 110 (3) (2003) 331–332.