

Female Hypospadias and Congenital Uterine Müllerian Anomaly with Proved Fertility: A Rare Case Report

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ABSTRACT

Congenital anomalies of the female genital tract result from müllerian duct anomalies and/or abnormalities of the urogenital sinus or cloaca. Due to the close developmental relationship between the genital and the urinary tracts, association of anomalies in both systems are common. This article reviews the appearance of developmental anomalies of the female urinary (hypospadias) and genital tracts and points out complication associated with bicornuate uterus. A 40-year-old primigravid woman conceived a pregnancy after 10 years of primary infertility. The prenatal course was uncomplicated. At 38 weeks' gestation, she was successfully delivered by elective low transverse cesarean section.

Keywords: Bicornuate uterus, Hypospadias, Infertility.

INTRODUCTION

Abnormal fusion of the müllerian ducts during embryological life results in a variety of congenital uterine malformations; the etiology of such abnormalities remains unknown. Müllerian duct anomalies occur in 0.1 to 3% of women, bicornuate uteri are thought to represent approximately 10% of müllerian duct anomalies; 15 to 25% of women with such congenital uterine anomalies have problems with fertility and reproduction, such as miscarriage, premature labor, premature rupture of the membranes or malpresentation.¹ Having a pregnancy in a patient with a bicornuate uterus is rare, especially, if it is associated with female hypospadias. In a female, external meatus is normally located between the clitoris and the vagina. Female hypospadias is a developmental anomaly in the female in which the urethra opens into the vagina. In females, hypospadias is much less common than in males. It appears about once in every 500,000 live female births.

CASE REPORT

A 40-year-old woman was undergoing investigation after 10 years of primary infertility when she conceived spontaneously. Prior to conception, hysterosalpingography showed the presence of a uterine malformation but could not determine whether the malformation was a completely bicornuate uterus or a uterus with a complete septum. Investigations had not been completed before the patient conceived. In the first ultrasound examination, a pregnancy of single viable fetus was diagnosed. Subsequent 3D ultrasound examinations demonstrated single viable fetus in left uterine horn in bicornuate uterus, subsequently confirmed at cesarean section (Fig. 1).

During pregnancy, the patient was monitored with ultrasound examinations every 2 to 3 weeks to assess fetal growth and to measure cervical length to assess the risk of premature delivery. Fetus grew appropriately for their gestational age, with no apparent difference in estimated fetal weight. The cervical length was consistently > 30 mm. Combined screening for chromosomal defects in the first trimester was also performed, showing a low risk of abnormality for fetus (estimated risk 1:1410). The prenatal course of this pregnancy, therefore, had none of the complications reported.



Fig. 1: Bicornuate uterus with right horn (normal) and pregnancy in left horn (post LSCS)

At 38 weeks' gestation, elective cesarean section was performed. During cesarean section, it was first diagnosed that patient was having hypospadias because there was no urethral opening between clitoris and vaginal introitus. Urethral opening was on anterior vaginal wall (Fig. 2). A single viable fetus in cephalic presentation was delivered successfully by low transverse cesarean section in left horn (Fig. 3). Birth weight was 3 kg, and Apgar score at 1 and 5 minutes were 8/9 and 10/10 respectively. To stimulate uterine contraction after delivery, intravenous carbetocin was given after manual removal of placenta. There were no complications during the surgery or

postpartum, and the patient was discharged five days after delivery with her healthy baby.

DISCUSSION

The müllerian ducts (or paramesonephric ducts) are paired ducts of mesodermal origin in the embryo. They run laterally down the side of the urogenital ridge and terminate at the müllerian eminence in the primitive urogenital sinus. In the female, they will develop to form the fallopian tubes, uterus, and the upper portion of the vagina; in the male, they regress.

A bicornuate uterus was thought to be associated with infertility² but nowadays it is not associated with recurrent pregnancy loss.³ This case also had no history of recurrent pregnancy loss. It is possible to diagnose a bicornuate uterus using gynecologic sonography, specifically sonohysterography and MRI.⁴ However, as there is no indication to do operative procedures on asymptomatic women, the presence of a bicornuate uterus may not be detected until during pregnancy or delivery. Women with congenital uterine malformation usually have higher incidence of infertility and complications with poorer obstetric outcome during pregnancy and delivery.² In a male, the external opening of the urinary tract (external meatus) is normally located at the tip of the penis. In a female, it is normally located between the clitoris and the vagina. Female hypospadias is an anomaly of the female urogenital apparatus, a defect of posterior wall of urethra and anterior vaginal wall, in which external urethral orifice opens into the cavity of the vagina. Hypospadias may be inherited, although it is mostly an isolated finding. Maternal age, number of previous pregnancies, deliveries, stillbirths, abortions, preeclampsia history, twinning, number of cesarean sections not complicated with female hypospadias.

We understood that this case was a very unusual and potentially high-risk pregnancy because association with hypospadias and potential complications included infertility, miscarriage, premature labor, premature rupture of the membranes, malpresentation, and intrauterine growth restriction can be associated with such uterine anomaly. Additionally, our patient was a 40-year-old primigravida with a previous history of infertility, and this was therefore a premium pregnancy.

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Fig. 2: Female hypospadias (catheter lying in vagina)



Fig. 3: Repair of scar in left uterine horn after lower segment cesarean section