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Uterus didelphys - a successful fetal outcome

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Abstract

Incomplete fusion of the Mullerian or Paramesonephric ducts results in the most common types of uterine malformations: uterus didelphys, uterus bicornis bicollis, uterus bicornis unicollis, uterus subseptae, uterus arcuatus, uterus unicornis . Uterus bocornis bicollis, is characterized by double or single vagina, double cervix and two single- horned uterus which show partial fusing of their muscular walls with duplication running right down to the uterine orifice. These malformations are rare but known to be associated with infertility, spontaneous miscarriages, intrauterine growth restriction, preterm deliveries, preterm perlabour rupture of membranes, breech presentation and increased rate of caesarean delivery. However, normal reproductive performance has been seen in association with them.

Keywords: Uterus Didelphys, MDAs, HSG

Introduction

Communicating uteri are a distinct class of uterine malformations characterized by the presence of a communicating tract between two otherwise separate utero cervical cavities. This malformation was first identified by Musset in 1967. Then musset et all and leo toaff proposed a comprehensive classification of communicating uteri. Ten different groups of communicating, uteri have been classified. Five of them are variant or subgroups. This morphological classification is based on the constant presence of isthmenian communication on the degree of separation of mulleins duct and their partial Artesia. These malformations are of accidental finding clinically because they do not affect patients clinical condition.

Uterus didelphys

Double uterus with an incidence of 1:2000 pregnancies is a type of mullerian duct anamoly. Fusion of the mullerian ducts normally occurs between the 6^{th} to 11^{th} weeks of gestation to form the uterus, fallopian tubes, cervix, and proximal two-thirds of the vagina. Any disruption of mullerian duct development during embryogenesis can result in a broad complex spectrum of congenital abnormalities termed mullerian duct anomalies (MDAs). The overies and distal third of the vagina originate from the primitive yolk sac and sinovaginal bud, respectively. Therefore, MDAs are not associated with anomalies of the external genitalia or ovarian development.

Diagnosis of MDAs is clinically important because of the high associated risk of infertility, endometriosis, and miscarriage, such that an estimated 15% of women who experience recurrent miscarriages are reported to have MDAs. MDAs are also commonly associated with renal anomalies, with a reported prevalence of 30%–50%, including renal agenesis (most commonly unilateral agenesis), ectopia, hypoplasia, fusion, malrotation, and duplication (1,3-5). Other congenital anomalies commonly associated with MDAs include those of the vertebral bodies (29%), such as wedged or fused vertebral bodies and spina bifida (22%–23%), cardiac anomalies (14.5%), and syndromes such as Klippel-Feil syndrome (7%).

Buttram and Gibbons proposed an MDA classification in 1979, which was subsequently modified by the American Society for Reproductive Medicine in 1988 (formerly the American Fertility Society). Accurate MDA recognition and classification are critical because treatment varies by the anomaly subtype. Of particular importance is correct identification of a septate uterus, since the septum may be composed predominantly of fibrous tissue; recurrent miscarriage in these patients is attributed to implantation of the embryo onto a poorly vascularized septum. Even with today's state-of-the-art imaging techniques, classification of MDAs may be challenging; when a specific designation cannot be made, it is best to describe the anatomy rather than to force the MDA into a category.

Imaging plays an essential role in MDA diagnosis and treatment planning. Currently, magnetic resonance (MR) imaging is the preferred means of evaluation. However, selection of the initial imaging modality is often dictated by the presenting clinical scenario (eg, primary amenorrhea, pelvic pain, or infertility). Hysterosalpingography (HSG) is routinely used in an initial evaluation of infertility; it allows assessment of the uterine cavity and fallopian tube patency but does not provide any information about the external uterine contour.

In younger patients or acute cases, ultrasonography (USG) is the preferred method because it is readily available, inexpensive, and rapid and does not use ionizing radiation. Field-of-view restrictions with USG, patient body habitus, and artifact from bowel gas may result in a request for further imaging with MR imaging. With the advent of three-dimensional (3D) techniques, USG may have the future potential to match the capabilities of MR imaging. Currently, however, MR imaging remains the preferred MDA imaging method, as it exquisitely details both the uterine cavity and external contours and has shown excellent agreement with clinical MDA subtype diagnosis.

To simplify the embryologic process, we adopted the three-stage approach used by Robbins et al : ductal development, ductal fusion, and septal reabsorption.

During the first 6 weeks of development, the male fetus and female fetus are indistinguishable, with both demonstrating paired mesonephric (wolffian or male genital) ducts and paramesonephric (müllerian or female genital) ducts. The presence of a Y chromosome is associated with production of müllerian-inhibiting factor. Therefore, after 6 weeks gestation, the absence of müllerian-inhibiting factor in the female fetus promotes bidirectional growth of the paired müllerian ducts along the lateral aspect of the gonads in conjunction with simultaneous regression of the mesonephric ducts. Interruption of müllerian duct development during this time gives rise to aplasia or hypoplasia of the vagina, cervix, or uterus.

Müllerian duct growth is accompanied by midline migration and fusion of these paired ducts to form the uterovaginal primordium. Interruption of the mullerian

duct *fusion* process gives rise to bicornuate uterus and didelphys MDA subtypes.

Between 9 and 12 weeks gestation, the fused müllerian ducts undergo a process of reabsorption of the intervening uterovaginal septum. Interruption of müllerian duct development during this *reabsorption* phase gives rise to septate or arcuate MDA subtypes. The reabsorption process is thought to occur in both cranial and caudal directions. The bidirectional reabsorption model is more congruent (than the previously suggested unidirectional model) with some forms of MDA such as isolated vaginal septum.

Although interruption in this phase of development is used to explain differences in MDA subtypes, both incomplete müllerian duct fusion and partial reabsorption of the uterovaginal septum may be difficult to differentiate. A common imaging and clinical challenge is the ability to distinguish a bicornuate uterus from a septate uterus. The sepate uterus carries a high risk of miscarriage and may be managed with resection of the septum. Absence of a cleft in the external uterine fundal contour with a duplicated endometrial cavity is the key feature used to diagnose a septate uterus rather than a bicornuate uterus.

Prevalence

The reported prevalence of MDA varies widely in the literature, ranging from 1%–5% in the general population to 13%–25% among women with recurrent pregnancy loss. This wide range of reported prevalence may be due to a variety of reasons including, but not limited to, lack of a universal classification system.

In a recent analysis of 94 observational studies, Chan et al reported an MDA prevalence of 5.5% in the general population, 8% in infertile women, 13.3% in women with a history of miscarriage, and 24.5% among women who have experienced miscarriage and infertility. They found that arcuate uterus was most common in the general population, affecting 3.9% of women, followed by bicornuate uterus (0.4%). Among women who experienced challenges conceiving (eg, infertility or miscarriage), septate uterus was a frequent finding, affecting 15.4% of women.

US and MR imaging play important roles in the diagnosis and evaluation of suspected MDA. HSG is typically indicated in the initial stages of an infertility work-up.

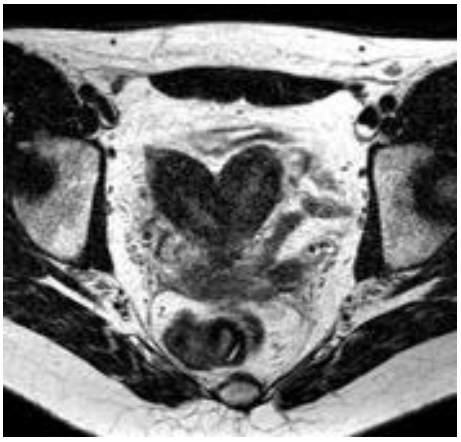

Fig. 1

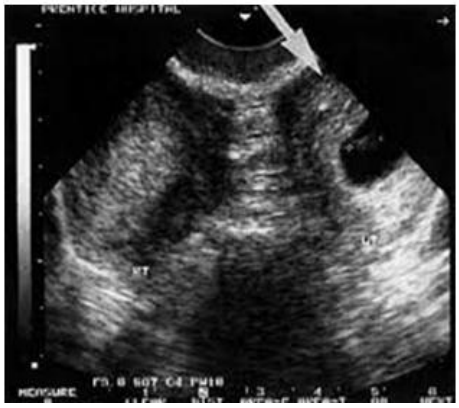

Fig. 2

While the presence of a divided rather than triangular uterine cavity at HSG may suggest the presence of an MDA, it is not possible to differentiate between subtypes. MR imaging and US provide greater anatomic detail; both of these imaging methods provide information on the external uterine contour, which is an important diagnostic feature of MDAs. Furthermore,

both MR imaging and US may be used to assess for oncomitant renal anomalies; renal anomalies occur at a higher rate among MDA patients.

An HSG examination is performed with fluoroscopy; a catheter is placed into the cervical canal, and a balloon is inflated to prevent contrast agent leakage. Water-soluble contrast material is then slowly introduced into the uterine cavity, with select fluoroscopic spot images obtained to evaluate uterine configuration, uterine filling defects, and fallopian tube patency (Fig 3). HSG allows evaluation of only the component of the uterine cavity that communicates with the cervix; since the anatomic information is limited without the ability to evaluate the external contours of the uterine fundus, HSG has little clinical utility in MDA evaluation.

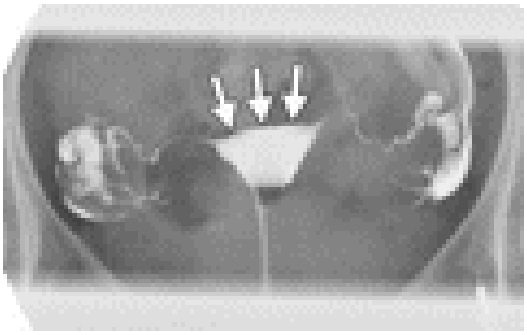

Fig. 3: Frontal HSG image shows a normal fundal contour of the endometrial cavity (arrows).

Case Study: A 21 year old 2nd gravida presented to OBG Dept of J.L.N. Medical College Ajmer with amenorrhoea of 71/2 Month . She had a first trimester abortion in first pregnancy .She had no complaints,but an early ultra sound done at 7week of gestational age detected a bicornuate uterus with pregnancy in the right horn. Per speculum examination showed two cervix. At 30 weeks of gestation she was admitted with complaints of pain abdomen. Tocolytics given and patient steroided for lung maturity. USG done revealed decreased liquor and asymmetrical IUGR of the foetus. Doppler showes early fetoplacental insufficiency. Patient treated with amino infusion. Antepartum foetal monitoring done with modified BPP and Doppler. Tocolytics stopped 48hrs after giving corticosteroids. At 30 weeks of gestation the patient presented with prelabour premature rupture of membranes. In view of non-reassuring foetal heart rate, an emergency lower segment caeserian section was done with the following intra operative findings. A bicornuate uterus with pregnancy in the right horn seen. The left horn in enlarged to around 14 weeks size and tubes on either cornu are healthly. A live male child in right occipitio posterior position weighing. 1.7 kg was delivered. Placenta is located anteriorly in the upper segment. By exteriorizing the uterus, findings are confirmed and communication between the two cavities is noted. She had uneventful post operative period and was discharged on 8th post operative day.

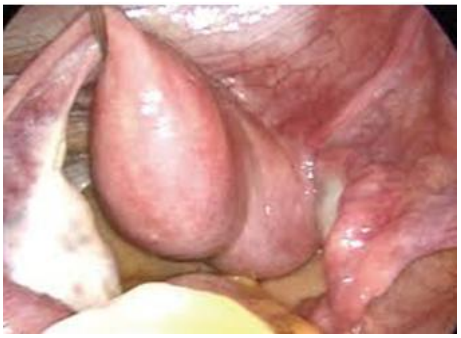

Fig. 4

Fig. 5

Discussion

The incidence of congenital uterine malformation is estimated to be 3-5% in general population. Bicornuate uterus is a congenital uterine anomaly that results from defective lateral fusion of the paramesonephric ducts at about the tenth week of intrauterine life around the fundus.Commnucating uteri account for 1-2% of malformation. Communicating uteri are often occasional findings during HSG usually performed for infertility or suspected malformation. The occurrence of a pregnancy in bicornis bicollis uterus gas been reported only sporadically in the literature. The detection of congenital uterine anomalies will increase because of heightened physician awareness and improved diagnostic modalities. In this case the Congenital uterovaginal anomalies can have adverse effects on pregnancy outcome. Offspring of the mothers with bicornuate uterus had a four times higher risk congenital defects than infants born to woman with normal uteri.

Conclusion

Early diagnosis and proper antenatal care is required to successfully manage a pregnancy with bicornate uterus. Patient with mullerian duct anomalies are known to have a higher incidence of infertility, repeated fist, second trimester spontaneous abortions intrauterine growth retardation, fetal malpositions, preterm labor, prelabour preterm rupture of mambreans & retained placenta. Reduced liquor may increase the difficulties while delivering the baby at cesarean section. Anticipation and preparedness to deal with these known complications will ensure positive outcome for the mother and baby.

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