

The Spectrum of Imaging Appearances of Müllerian Duct Anomalies: Focus on MR Imaging

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Abstract: *This study was designed with an aim to determine diagnostic efficiency of MRI in classification of sub-types of mullerian duct anomalies. I have studied 30 cases of diverse sub types of mullerian duct anomalies and the corresponding MRI findings. In this retrospective study, patients (n = 30) with clinical suspicion and diagnosed (ultrasound / HSG) of mullerian duct anomalies were referred to department of radiology and examined with MRI. Axial, sagittal, coronal T2 and axial T1 -weighted images were acquired. MRI based diagnosis was made and patients were grouped according to the ASRM classification system. MRI has emerged as the universally accepted standard in the imaging evaluation of Mullerian duct anomalies. Accuracies of up to 100% in the evaluation of Mullerian duct anomalies have been reported. MRI provides excellent delineation of both internal and external uterine anatomy. Most common anomaly in total study sample is hypo-plastic uterus (30%). MRI revealed seven patients with mullerian agenesis (23%), nine patients were diagnosed with hypoplastic uterus (30%), two patients with didelphys uterus (7%), three patients with bicornuate uterus (10%) and eight patients with septate uterus (26%) and one patient with arcuate uterus (4%).*

Keywords: Müllerian duct anomalies, Pelvic MRI, Uterine abnormalities, ASRM classification, Reproductive tract anomalies

1. Introduction

- Mullerian duct anomalies (MDAs) are a diverse group of rare congenital conditions resulting from abnormal development or non-fusion of the Mullerian ducts during fetal development, which give rise to the female reproductive tract. [1]
- Concomitant urinary malformations, particularly renal agenesis, are often present in these conditions. [1]
- Fetal intrauterine growth retardation, fetal malposition, multiple spontaneous abortions in the first trimester, infertility, premature labor and retained placenta are all known to be more common in patients with MDA. [2]
- The role of imaging is to detect and classify these Mullerian duct anomalies so that appropriate treatment is undertaken.
- The assessment of Mullerian duct anomalies has gained attention in recent years due to MR imaging.
- The ability of magnetic resonance imaging to precisely describe and illustrate the anatomical relationship of the female reproductive organs to surrounding structures has made it a potent imaging technique that complements and, in many situations, competes with alternate imaging modalities.

Embryology:

Embryologically, at six weeks of gestation, two pairs of mesonephric (wolffian) and paramesonephric (mullerian or female genital) ducts are present. Since the female foetus lacks the mullerian inhibitory factor, the mullerian ducts will expand in both directions while the mesonephric ducts will simultaneously regress.[3]

The mesothelial, mesenchymal, and primordial germ cells give rise to the ovaries, which are distinguishable by the tenth week of gestation. The gubernaculum, which later forms the utero-ovarian ligament and round ligament, guides the ovaries as they descend caudally from the urogenital ridge next to the

developing kidneys to the pelvis during Müllerian duct fusion.[4]

In patients with müllerian hypoplasia, the function of the ovaries is usually normal, which reflects their separate embryologic origins. The ovaries may be located superior to their expected location owing to failure of descent.[5]

Classification:

- The classification of the American Fertility Society (American Society for Reproductive Medicine; ASRM) is the most widely accepted classification of mullerian duct anomalies. It is a revision of the classification system first created by Buttram and Gibbons in 1979.[6]
- The classification is based on the extent of failure of normal development and thus groups anomalies with similar clinical manifestation and treatment together with prognosis for pregnancy outcome
 - Class I: mullerian agenesis or hypoplasia
 - Class II: unicornuate uterus
 - Class III: uterus didelphys
 - Class IV: bicornuate uterus
 - Class V: septate uterus
 - Class VI: arcuate uterus
 - Class VII: diethylstilbestrol (DES)-related anomalies

2. Materials & Methods

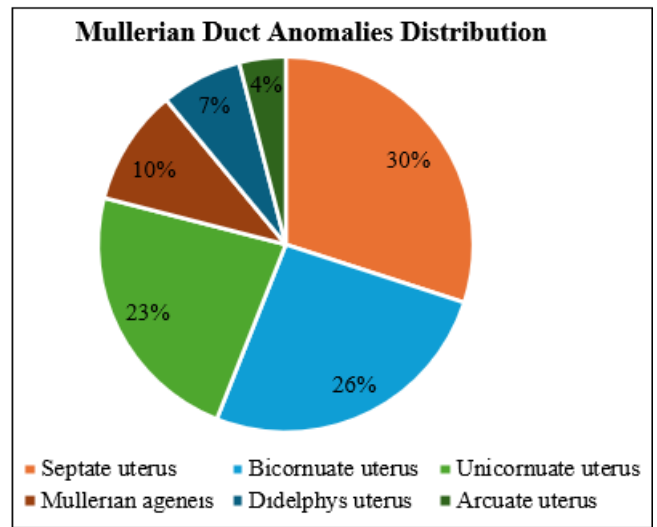
- In this retrospective study, patients (n = 30) with clinical suspicion and diagnosed (ultrasound / HSG) of mullerian duct anomalies were referred to department of radiology and examined with MRI.
- The mean age recorded was 22 years (age range 11–54years).
- They had variable clinical indications including amenorrhea, infertility, cyclic abdominal pain, and one case of post-partum bleeding.

- MRI was performed on a GE health care 1.5T MRI machine. Informed consent was obtained from all patients.
- No contrast material was used. T1- and T2 weighted images were acquired in Sagittal, trans axial and coronal planes.
- Additional coronal T2-weighted images with a large field of view were done for some patients to visualize the kidneys.
- Detailed imaging findings were recorded for each patient. Differentiation between septate and bicornuate uterus was done as follows: 1) If an external fundal indentation of < 1 cm on the coronal uterine plane (This is determined by drawing a line between the tops of both uterine horns and measuring the distance perpendicularly from this line to the lowest point of the indentation). 2) If the internal fundal indentation towards the endometrium is > 1 cm (measured by drawing a line between the two tubal ostia and measuring the distance from this line to the top of the myometrial or fibrous septum towards the endometrial cavity). 3) The leading edge of the septum is at an angle (indentation angle) of $< 90^\circ$. On the other hand, to diagnose a bicornuate/didelphys abnormality, the external fundal contour indentation to be by > 1 cm. [7] Other ways of calculation are available in the literature, which are particularly useful in equivocal cases, they are 4) If the intercornual distance is 4 cm seen more commonly in a bicornuate uterus. 5) if the intercornual angle is 105° , more suggestive of a bicornuate uterus. [8] 6) The septate uterus was further stratified into partial septate uterus, if the septum does not reach the external cervical os and complete septate uterus if it reaches the external os resulting in two separate cervical canals. [9,10]
- The resulting anomalies were then stratified according to the most widely used classification system of ASRM mullerian anomalies classification 2021.

- Associated anomalies particularly of the urinary system, special findings regarding the ovaries.

3. Results

- Variable congenital mullerian anomalies were found at different frequencies in a total study group of 30 patients.
- Most common anomaly in total study sample is septate uterus (30%). MRI revealed nine patients with septate uterus (30%), eight patients with bicornuate uterus (26%), seven patients with Unicornuate uterus (23%), three patients with mullerian agenesis (10%), two patients with didelphys uterus (7%), one patient with arcuate uterus (4%).



Class I: Müllerian Agenesis or Hypoplasia

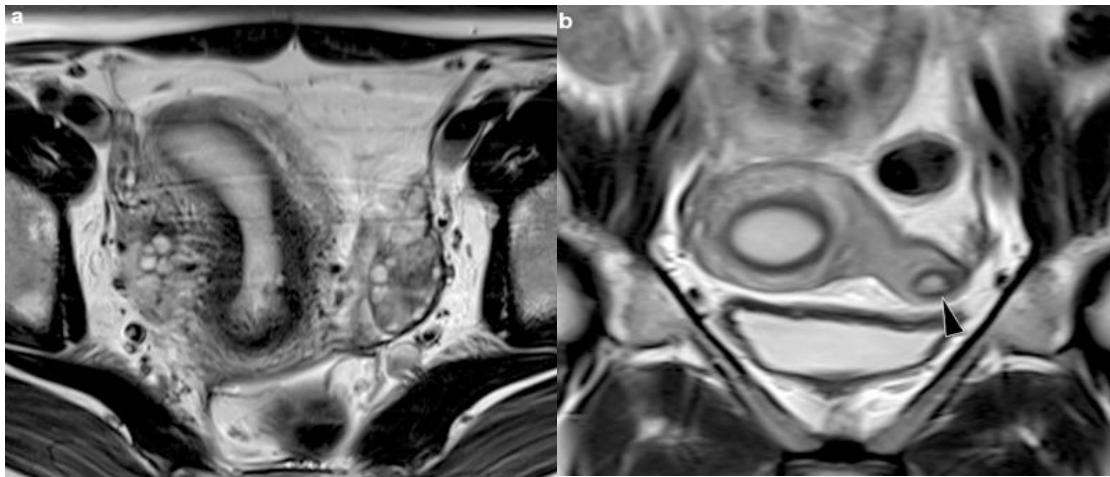


A 21-year-old female with primary amenorrhea.

- Sagittal T2-weighted image shows a triangular soft tissue (white arrow) above the bladder dome, which is continuous with the atrophic vagina.
- Coronal T2-weighted image shows a fibrous band-like structure (black arrows) connecting with bilateral remnants (black arrowheads)

- Axial T2-weighted image demonstrates bilateral müllerian remnants (white arrowheads) adjacent to bilateral normal ovaries (not depicted in this slice) in the pelvic cavity.

Class II: Unicornuate Uterus

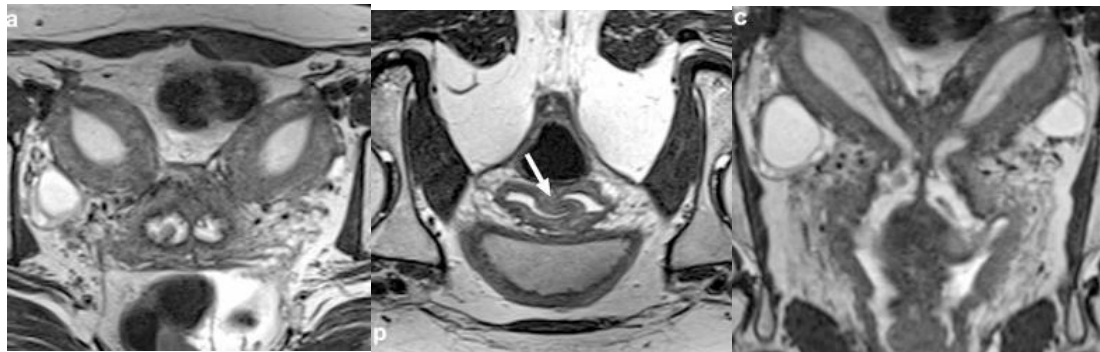


Unicornuate uterus with an obstructed non-communicating rudimentary horn (class IIb) in a 35-year-old female who was referred to our hospital for evaluation of infertility.

- a) Axial T2-weighted image shows the classic banana shape appearance of a right Unicornuate uterus with normal zonal anatomy.

- b) The left rudimentary horn has a small endometrial cavity (black arrowheads) and normal myometrial zonal anatomy on coronal T2-weighted images.

Class III: Uterus Didelphys



Uterus didelphys (Class-III) with vaginal septum in a 33-year-old female who underwent MRI examination for pre-pregnancy checkup

- a) Axial T2 weighted image shows complete duplication of uterine horns, with partial degree of fusion of adjacent cervixes
b) Axial T2 weighted image shows middle vagina with a longitudinal septum (white arrow)

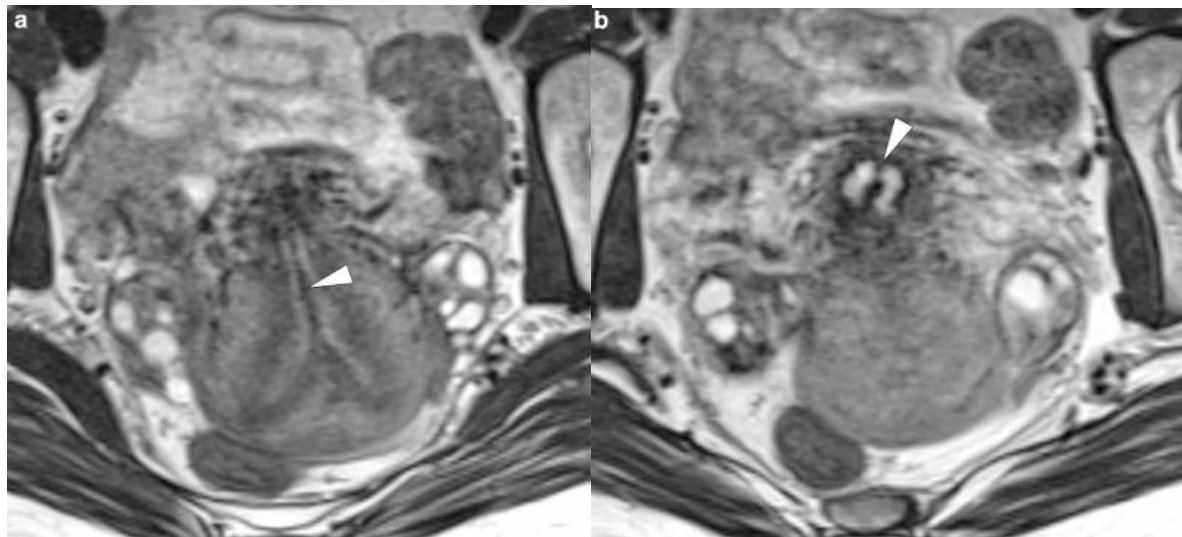
- c) Curved MPR of T2-weighted image shows duplication of the uterine horns, cervix, and proximal vagina without obstruction

Class IV: Bicornuate Uterus



Bicornuate uterus (class IV) in a 20-year-old female with right transverse vagina septum

- Coronal T2-weighted image shows bicornuate uterus with deep fundal cleft (>1 cm, black arrowhead).
- Axial T2-weighted image shows a transverse vagina septum in the right side of the upper vagina (white arrow)



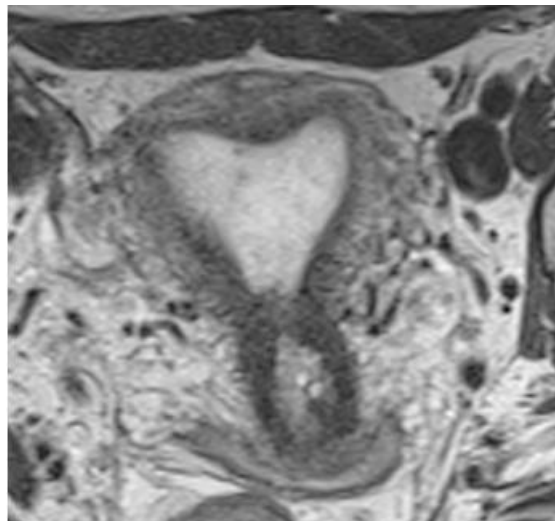
and adjacent multilocular cystic lesions (black arrowheads).

Class V: Septate Uterus

Complete septate uterus (class V) in a 33-year-old female with primary infertility. A, B Axial T2-weighted images show a backward uterus with normal external uterine contour and a low intensity fibrous septum (white arrowhead) originating from the muscular septum and extending into the cervical os.

DES causes a wide variety of disorders including uterine hypoplasia and a T-shaped uterine cavity (most common), cervical stenosis and vaginal adenosis, which increase the risk of vaginal clear cell carcinoma

Class VI: Arcuate Uterus:



Arcuate uterus (class VI) in a 38-year-old female with hypermenorrhea and chronic anemia.

Axial T2-weighted image shows a smooth, broad-based, shallow endometrial cavity with a normal convex external contour of the uterine fundus. Low intensity fibrous septum is absent.

Class VII: Diethylstilbestrol (DES)-Related Anomalies

Up to 15% of female offspring from women exposed to DES had uterine malformations and association with vaginal clear cell carcinoma.

4. Discussion

- MRI has emerged as the universally accepted standard in the imaging evaluation of Mullerian duct anomalies. Accuracies of up to 100% in the evaluation of Mullerian duct anomalies have been reported. MRI provides excellent delineation of both internal and external uterine anatomy. [10]
- T2 weighted images provide excellent detail regarding uterine zonal anatomy and are considered the mainstay of pelvic imaging.
- Not only MRI was able to correctly characterize the external uterine contour, but also it revealed the tissue composition of the septum muscular or fibrous septum. This particular finding is of great clinical importance as tissue composition significantly impacts on the surgical approach: hysteroscopic metroplasty with resection of the septum suffices in septate uterus, whereas a well-vascularized myometrial septum in bicornuate uterus requires a transabdominal approach in order to prevent life threatening bleeding. These results are in concordance with Carrington et al,[11] and Mintz et al,[12] who could show that MRI is an excellent tool for depiction of septum tissue composition.
- MRI does accurate characterization of the different subtypes of Mullerian duct anomalies could be often impossible using other diagnostic methods. Physical examination is of very limited value in diagnosis of MDAs.[13]
- HSG has been the first line of defense in diagnosis of anomalies of the female reproductive system for many years, but its diagnostic value is severely impaired by its lack to display the external uterine contour (which is

particularly important for differentiation of septate and bicornuate uterus) [13]

5. Conclusion

MRI is quite helpful in the differentiation of uterine anomalies. With its ability to provide information about the external and internal uterine anatomy and tissue composition, it represents a powerful tool in diagnosis and classification of patients with MDAs, and thereby creates a strong basis for choosing on further treatment options.

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