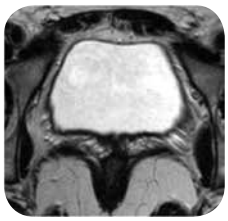

Müllerian Duct Anomalies: Diagnostic Evaluation with Magnetic Resonance Imaging

Anomalías en los ductos Müllerianos: Evaluación diagnóstica por resonancia magnética

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Summary

The Müllerian ducts are two embryological structures that undergo three main processes during their formation: development, ductal fusion, and septal resorption. Müllerian duct anomalies (MDAs) include a large spectrum of malformations that involve the uterus, fallopian tubes, cervix, and the upper two-thirds of the vagina. The severity and morphological characteristics are given by the specific embryological moment in which the disorder occurs. Women with these anomalies present symptoms such as primary amenorrhea, infertility, and obstetric complications. Magnetic resonance imaging (MRI) has become a very useful tool for evaluation, given that it is a noninvasive technique that does not use ionizing radiation, while allowing multiplanar evaluation for a proper assessment of the external uterine contour, which makes this method a useful technique for accurate diagnosis and surgical planning.

Resumen

Los ductos Müllerianos son dos estructuras embrionarias que en su desarrollo pasan por tres procesos fundamentales: desarrollo, fusión ductal y reabsorción septal. Las anomalías de los ductos Müllerianos (ADM) incluyen un gran espectro de malformaciones que comprometen el útero, trompas de Falopio, cuello uterino y los dos tercios superiores de la vagina. La gravedad de las ADM y sus características morfológicas están dadas por el momento embriológico específico en el que aparece el trastorno. Las mujeres con estas anomalías consultan principalmente por amenorrea primaria, infertilidad y complicaciones obstétricas. La resonancia magnética (RM) se ha convertido en una herramienta muy útil para su adecuada evaluación por tratarse de una técnica no invasiva, que no utiliza radiación ionizante y permite una evaluación multiplanar con adecuada valoración del contorno uterino externo, lo que la constituye en una técnica necesaria para el diagnóstico preciso, así como para la planeación quirúrgica de las pacientes.

Introduction

The Müllerian ducts are two embrionary structures that fuse and reabsorb during the Gestation to develop the uterus, fallopian tubes, the cervix and the upper two thirds of the vagina (1). The merger of the Müllerian ducts normally occurs between the sixth and eleventh weeks of gestation (2). Anomalies of the Müllerian ducts (MDA) have a prevalence ranging from 1% to 5% of the General population and between 13% and 25% in patients with a history of recurrent abortions (3-5). Any alteration in the normal development of the ducts during embryogenesis results in a broad spectrum of congenital abnormalities. The diagnosis of MDA is of great value due to its association with infertility, endometriosis, major risk of miscarriages (described in 15% of patients) and its association with abnormalities in renal development (6-9).

Embriology

Robbins and colleagues described three stages In the embryogenesis of the Müllerian ducts:

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Ductal development: Occurs after the sixth week due to the absence of Müllerian inhibitory factor. The interruption in the development of the Müllerian ducts in this period leads to aplasia or hypoplasia of the vagina, cervix and uterus.

Ductal fusion: In the development of the Müllerian ducts there is medial migration and fusion of the pair of ducts to form a uterovaginal primordium. The interruption of this leads to bicornuate and didelphic uterus.

Septal reabsorption: Between the ninth and twelfth week of gestation, the uterovaginal septum is reabsorbed. The absence of this reabsorption results in a septate or arcuate uterus. This reabsorption is carried out in a bidirectional (cranial and caudal) direction (10).

Diagnostic approximation through imaging

Hysterosalpingography is used to evaluate the permeability of the fallopian tubes and is the first test that is practiced in patients who consult for infertility (11-13). When the suspicion is specific for an MDA, ultrasound and MRI are the initial examinations, especially MRI, since it allows a greater anatomical detail of the internal uterine cavity and of its external contours.

The protocols usually used by MRI include axial images enhanced in T1 and T2 and coronal oblique T2 images, which are very important to evaluate the external contour of the uterine fundus. At this moment, with the appearance of T2-weighted three-dimensional sequences, cuts of a sub-millimeter thickness can be achieved and from these perform multiplanar reconstructions, which reduces the acquisition time of images (1).

MR protocol

The MR protocol includes: turbo spin echo images (TSE potentiated in T2) in the 3 planes (oblique, axial and sagittal coronal), oriented with the axes of the uterus (figure 1). The oblique coronal plane Is important in the diagnosis of MDA as it allows the external contour of the uterine fundus. They can be used as T2-weighted three-dimensional (3D) sequences with millimeter cut thickness for multiplanar reconstructions, and thus reduce the acquisition time of the image. As well, T1-weighted axial images with fat saturation of the entire pelvis are included to identify blood content, for example, in the study of hematocolpos or to identify foci of endometriosis As an associated disease in patients with infertility problems. It is important to include an image in the enhanced coronal plane In T2 (HASTE) with a large field of view (FOV), including Kidneys, since MDA are associated with renal abnormalities (agenesis, renal ectopy) because they share the same embryological origin.

The administration of contrast medium, gadolinium compounds (Gd) and diffusion sequences are performed within our pelvic protocol to evidence additional disease identified incidentally.

Classification

Although there is no universally accepted classification for malformations of the Müllerian ducts, the proposal by Buttram and Gibbons (14) in 1979, and subsequently revised in 1988 by the American Society of Reproductive Medicine (15) is the most accepted. It divides it into 7 subtypes:

» Class I: hypoplasia or uterine agenesis
» Class II: unicorn uterus
» Class III: uterus didelphys
» Class IV: bicornuate uterus
» Class V: septate uterus
» Class VI: arcuate uterus
» Class VII: diethylstilbestrol (DES) related abnormalities

Agenesis or hypoplasia (I)

An early alteration in the development of the Müllerian ducts results in the most extreme form of malformations: the Mayer-Rokitansky-Küster-Hauser syndrome (16) (Figure 2). This syndrome consists of agenesis or hypoplasia of the proximal two thirds of the vagina, cervix and uterus. Ovaries and external genitalia are normal in these patients who, generally, manifest primary amenorrhea at puberty. MRI is the exam of choice for the evaluation of these anomalies. The most important treatment In these patients is the reconstruction of a neovagina.

Unicorn uterus (II)

It is the result of the normal development of a single Müllerian duct with complete or near complete failure in the formation of the contralateral duct. There are 4 subtypes of this anomaly (figure 3).

» Absence of rudimentary horn
» Rudimentary horn without uterine cavity
» Rudimentary horn with uterine cavity communicating to the normal side
» Rudimentary horn with non communicating uterine cavity

Identifying endometrial tissue in the rudimentary non-communicating horn is of great use, since it is associated with pelvic pain and endometriosis due to retrograde menstrual flow (17,18). Other possible associated risks with this anomaly are higher possibilities of abortion, ectopic pregnancy, preterm delivery and uterine rupture (19).

In patients with this type of anomaly one can be observe, in an associated manner, renal malformations by 40%, with renal agenesis being the most frequent associated renal anomaly.

Uterus didelphys (III)

This malformation constitutes 5% of MDA and occurs due to the absolute absence in the merger of the Müllerian ducts, showing a duplication of the uterine horns, cervix and, in 75% of the patients, proximal vagina (20). The vaginal septum may lead to the develop-

ment of hematometrocolpos, obstructing the exit of the endometrial cavity, while the nonobstructive didelphic uterus is usually asymptomatic and is usually diagnosed in an incidental manner. In case there is obstruction of a hemivagina, it can manifest as cyclic pelvic pain, associated with a mass in the hemivagina on physical examination.

In the MRI, one can observer a complete separation of both uterine cavities, without communication between them, with a slit to the uterine fund greater than 1 cm, as well as two uterine necks and a longitudinal vaginal septum (Figure 4). The slit toward the uterine fundus greater than 1 cm has been described as a finding with a sensitivity and specificity of 100% to differentiate the anomalies in the fusion from the abnormalities in the reabsorption of the septum (21).

The surgical management of these patients depends on whether or not there is obstruction of a hemivagina, in which case the vaginal septum is removed. If not, it does not require surgery.

Bicornuate uterus (IV)

This MDA is due to incomplete fusion of the proximal part of the Müllerian ducts, with two uterine cavities in the fundic portion that attaches to the uterine isthmus. In this anomaly, the uterine fundus has a deep indentation greater than 1 cm, the intercornual distance is greater than 4 cm and the intercornual angle is greater than 105º (Figure 5). It may be associated with duplication of the cervix and a vaginal septum.

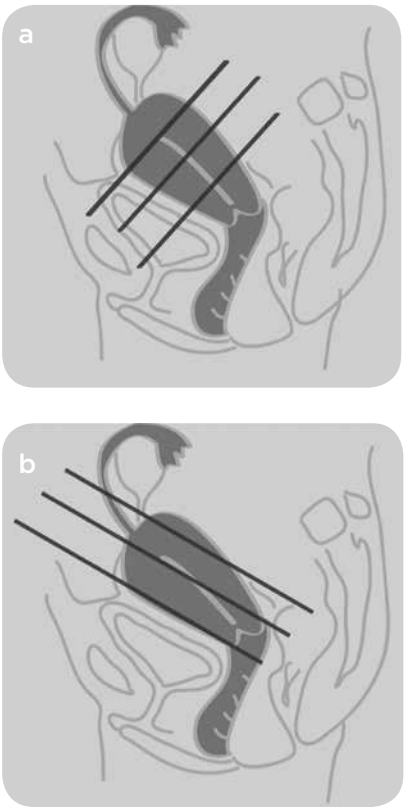

Figure 1. Acquisition of MR images. a) Oblique coronal plane to the major axis of the uterus and b) axial or perpendicular to the long uterine axis.

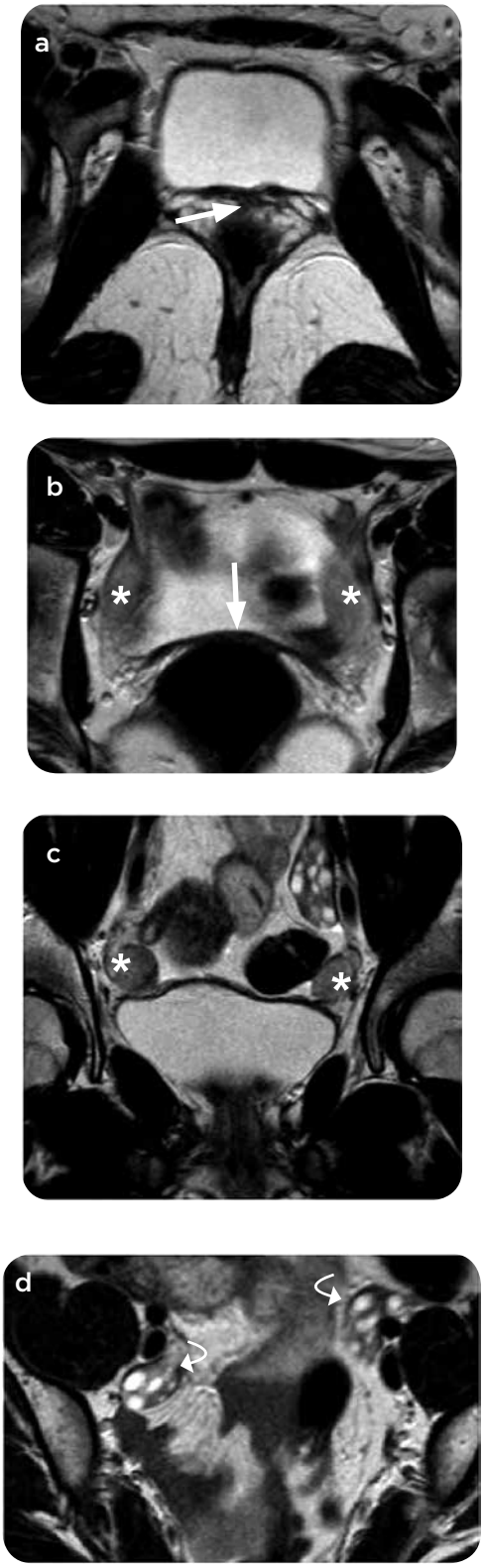

Figure 2. Mayer-Rokitansky-Küster-Hauser syndrome. a, b and d). Axial TSE MR enhanced in T2; c) T2 enhanced coronal TSE: absence of the uterus, cervix and vagina (arrows), uterine embryonic remnants on both sides of the pelvis are observed as ovoid images with intermediate solid signal (asterisks in b and c). Ovaries with normal characteristics on both sides of the pelvis (curved arrows).

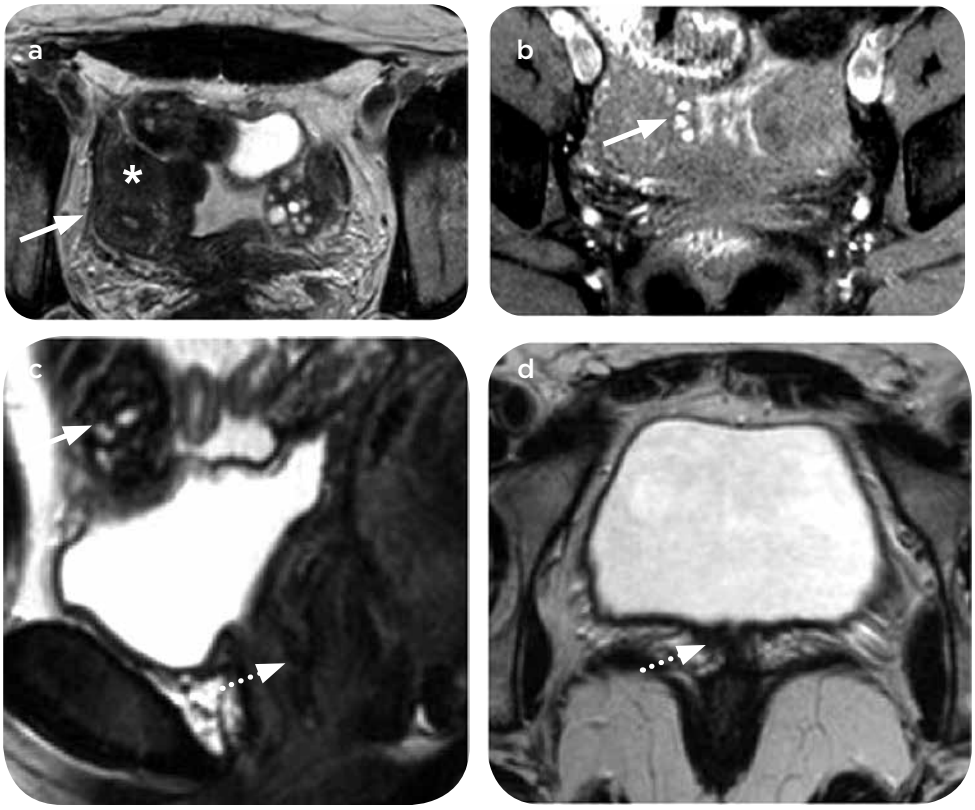

Figure 3. Unicorn horn uterus with rudimentary horn and communicating endometrial cavity. a and d) T2 axial TSE; b) Axial T1 with fat saturation; c) T2 sagittal TSE. Right unicorn uterus of oblong morphology (arrow in a) with focus of intramural adenomyosis towards the left slope of the fundus (asterisk) that is evidenced as high signal punctiform focci, in the myometrial thickness (arrow in b and c). The uterus is not identified in the midline between the bladder and rectum (dotted arrows in c and d).

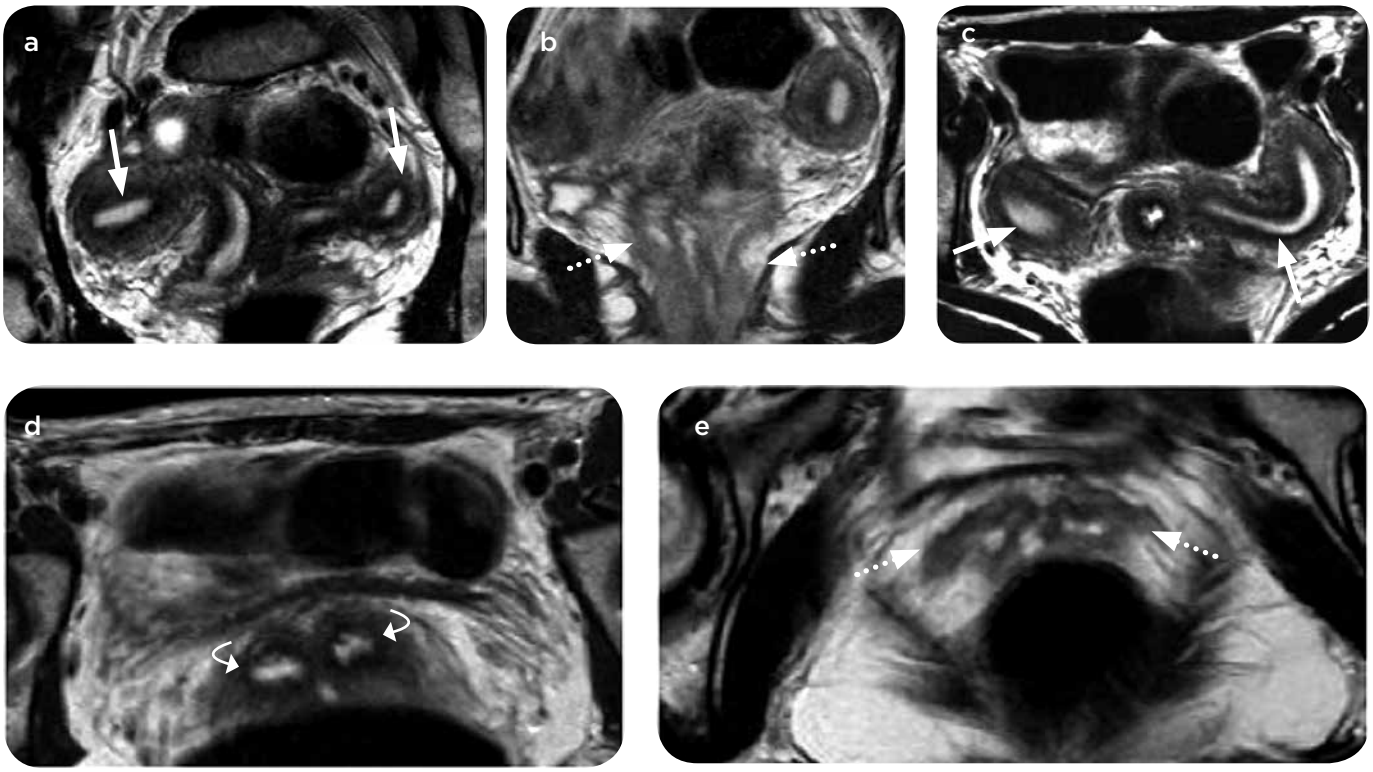

Figure 4. Uterus didelphys. a and c) Oblique coronal MRI TSE enhanced in T2. b, d and e) Axial TSE enhanced in T2. Two completely separated and divergent uterine cavities, with independent endometrial cavity (arrows in a and c). Two uterine necks (curved arrows in d) and two vaginas (dotted arrows on b and e).

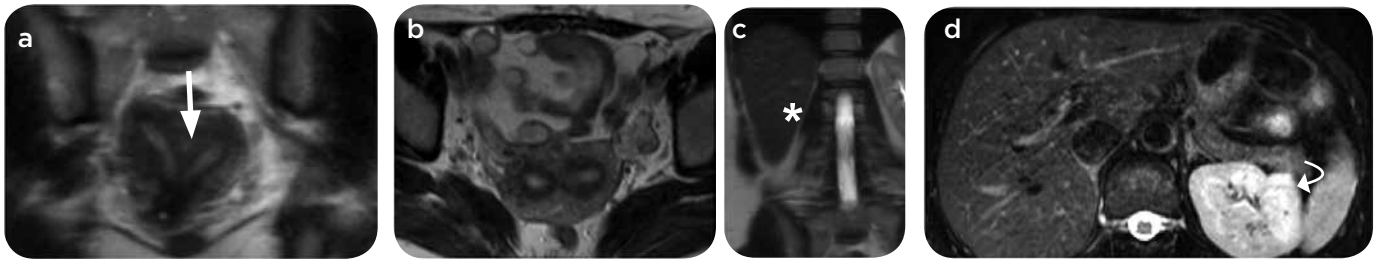

Figure 5. Bicornuate uterus. a and b) Coronal and axial T2 TSE. The uterus is seen with two uterine horns communicating to the lower slope, with a 1-cm slit towards the uterine fundus (arrow in a) and intercornual separation of 4 cm; c) coronal T2 HASTE and d) axial TSE T2 with fat saturation: Absence of the right kidney (asterisk) with left kidney of normal morphological characteristics (curved arrow).

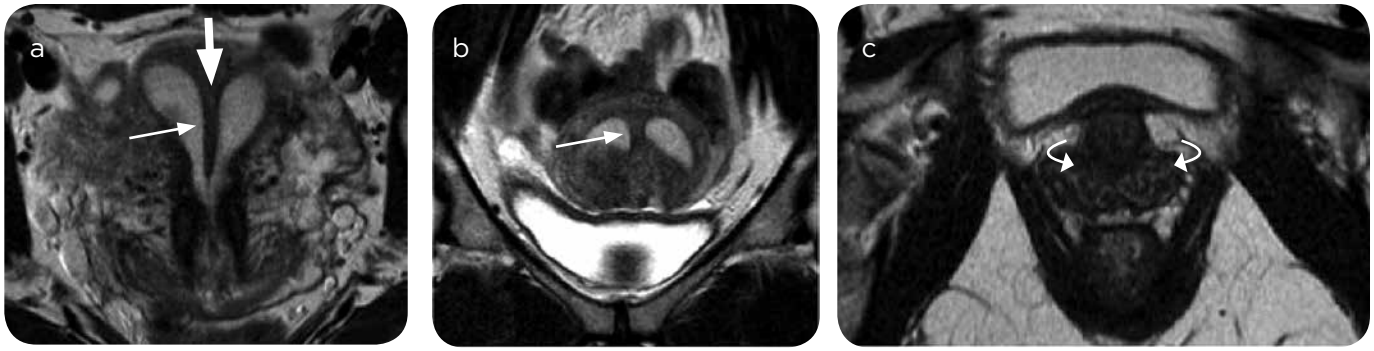

Figure 6. Complete septate uterus. a) Coronal T2-weighted TSE MR, oblique to the major axis of the uterus; b) coronal TSE enhanced in T2; c) axial TSE enhanced in T2. External uterine contour of the convex fundus (thick arrow in a), with intercornual distance less than 4 cm. Complete septum extending from the fundus to the internal cervical orifice (arrow in a and b). Vaginal partition forming two vaginas (curved arrows in c).

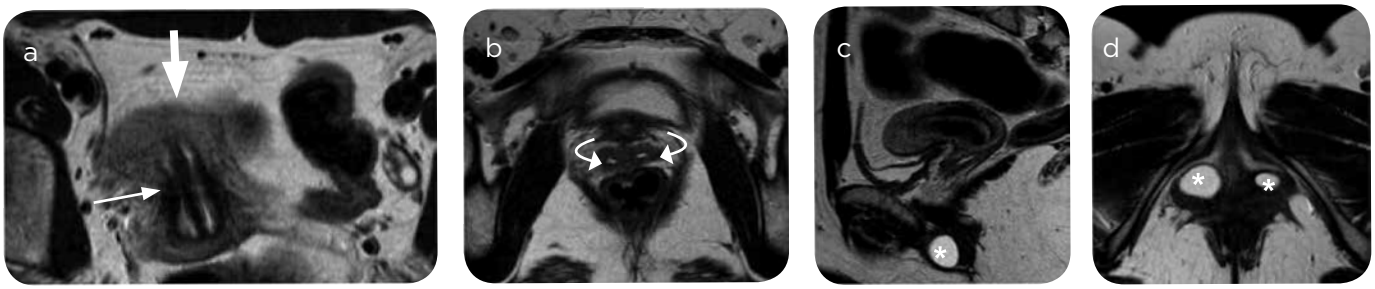

Figure 7. Complete septate uterus with septate vagina and two Bartolino cysts. a) TSE T2 coronal MRI oblique to the longitudinal uterine axis; b and d) Axial TSE T2 MRI; c) sagittal TSE T2. External contour of the convex fundus (arrow). Complete septum reaching the internal cervical orifice (thick arrow). Two vaginas are seen separated by vaginal septum (curved arrows). Bilateral cysts of Bartolino in both slopes of the vaginal introitus (asterisks).

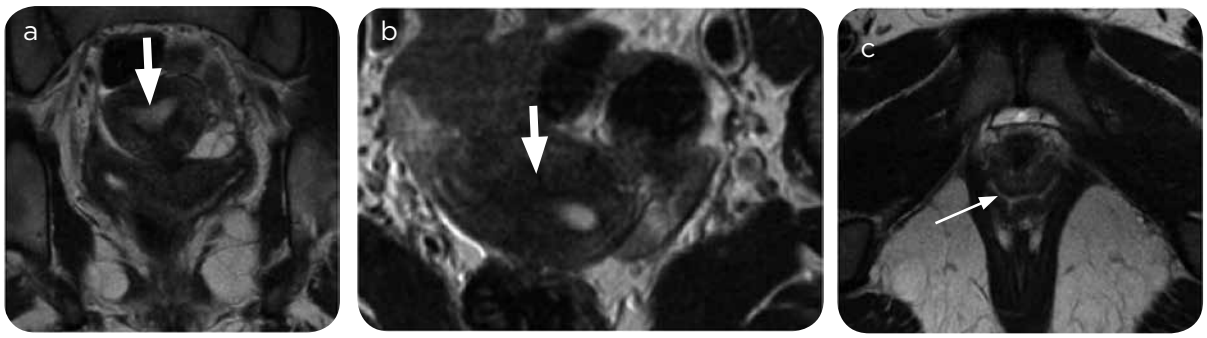

Figure 8. Arcuate uterus. a) TSE T2 Coronal MRI oblique to the longitudinal axis of the uterus; b and c), Axial TSE T2. Outline of the external fundus with imprint of the myometrium on the uterine cavity over the fundus (arrow in a and b). Single vagina is observed (arrow in c).

Septate uterus (V)

It is the most frequent anomaly, with an incidence of 55% of MDA. It is due to a failure in total or partial resorption of the uterovaginal septum (22-24). The septate uterus may be complete or incomplete depending on whether or not the internal cervical opening is reached. This type of malformation presents the highest incidence of recurrent abortions, so its diagnosis is important (25). The cleft of the uterine fundus should be less than 1 cm, and its contour convex or straight (figures 6 and 7), with intercornual distance less than 4 cm and intercornual angle less than 75º. Unlike bicornuate uterus, septate requires surgical treatment with hysteroscopic resection of the septum, to improve the obstetric outcomes.

Arcuate uterus (VI)

The malformation of the Müllerian ducts is milder, in that there is an almost total resorption of the uterovaginal septum, which, through images, is seen as a small fundus indentation (figure 8). In the evaluation, the uterine indentation is formed by myometrium and the external contour of the uterus is normal. This anomaly is usually asymptomatic and has no impact on reproductive life.

Anomalies associated to diethylsilbestrol (DES) (VII)

DES is a non-steroidal estrogen that was used around 1950 for some obstetric conditions, such as abortions and preclampsya. Later, it was discontinued in 1971 because it was found that in daughters of the patients exposed to this drug there was a higher prevalence of clear cell carcinoma and T-shaped infant hypoplastic uterus (26, 27). This condition is best evaluated by hysterosalpingography, in which an irregular endometrium is seen, with a decreased size upper uterine segment and a characteristic "T" shape. With MRI it is observed as a hypoplastic uterus with T-shaped endometrium and stenosing bands, which give the endometrium an irregular appearance (28).

Conclusion

The malformations of the Müllerian ducts are infrequent alterations, but with important clinical implications such as infertility, obstetric complications, endometriosis, among others. For its specific diagnosis the MRI has become the main imaging technique. It is fundamental to know the normal embryological process of the Mullerian ducts during the formation of the different structures to understand the cause of its morphological findings and, in this way, make a more precise diagnosis.

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