Case 14563

Eurorad

The clinical relevance of anatomical variants!

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Section: Genital (female) imaging
Area of Interest: Genital / Reproductive system female
Procedure: Diagnostic procedure
Imaging Technique: MR
Imaging Technique: Ultrasound
Special Focus: Congenital Case Type: Clinical Cases
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Patient: 13 years, female

Clinical History:

A 13-year-old girl was admitted with pelvic pain. She referred previous episodes of less severe dysmenorrhoea. Past medical history was not significant. Physical examination and lab tests were unremarkable, except for mild tenderness on middle lower abdomen. Menarche occurred at 11 years of age and she was in the secretory phase.
Imaging Findings:

Ultrasonography showed two completely separate uterine horns, each with its own endometrial cavity, without communication between the two endometrial cavities.
MR images confirmed the presence of a large mass occupying the lower pelvis with signal characteristics of blood products - high signal intensity on T1-WI and low signal intensity on T2-WI.
Two structures with a muscular wall were identified superiorly to the mass, but just the right structure was in direct communication with the mass. These structures represent the two uterine horns, with the right uterus communicating with a massively dilated blood-filled right hemivagina.
MR imaging demonstrated widely divergent uterine horns, with a midline fundal cleft greater than 1 cm in the external contour of the uterus, two separate cervices, and a unilateral hemivaginal septum that causes the obstruction.
On the coronal T2-WI, the left kidney is in its normal location and appears unremarkable, while the right kidney cannot be identified.
Discussion:

Müllerian duct anomalies (MDA) are developmental malformations that resuIt from disturbance of normal growth of the müllerian ducts, which originate in the uterus, fallopian tubes, cervix, and proximal two-thirds of the vagina.

MDA can be classified according to the three main stages of the embryologic female genital tract development: ductal development, ductal fusion, and septal reabsorption.

Uterus didelphys results from complete failure of müllerian duct lateral fusion that normally occurs between the 6th and 11th weeks of gestation. Uterus didelphis constitutes approximately 5% of MDA. [1, 2]

In this abnormality two distinct uteri and cervix can be identified, without communication between the two

endometrial cavities. In each uterus, the endometrial-to-myometrial width and ratio are preserved, and demonstrate normal zonal anatomy. Varying degrees of duplicated proximal vagina are present in a majority of these patients because a complete or partial longitudinal vaginal septum is associated in 75% of cases. In some of these patients defects in vertical fusion producing a transverse vaginal septum can also occur, with subsequent ipsilateral obstruction. [3]

Clinical presentation of uterus didelphys is associated to the presence of obstruction. In the absence of vaginal obstruction, uterus didelphys is usually asymptomatic. With unilateral vaginal obstruction it may become symptomatic at menarche and manifest as dysmenorrhea that overlaps with regular menses from the contralateral patent side, so these patients do not have amenorrhoea. This obstruction results in haematocolpos/haematometrocolpos and can cause retrograde menstrual flow, which can be the origin of endometriosis, infections, and pelvic adhesions. [4]

Diagnosis is clinically important because of the high associated miscarriage rate, and an unfavourable obstetric outcome.

Because of the close embryologic development of the urinary and genital systems renal anomalies occur more frequently among MDA patients, and renal agenesis is the most common associated anomaly.
Renal agenesis has a stronger correlation with obstructed type MDA and especially with obstructed uterus didelphys, located on the same side as the obstruction. [5]

MR imaging is the preferred imaging method for MDA evaluation because it can identify the type of uterine anomaly, the level of possible vaginal obstruction, and the presence or absence of an ipsilateral kidney. MR also provides reliable delineation of the external uterine contour and clear delineation of internal and external uterine anatomy in multiple planes. [6]

Management depends on the presence or absence of an obstruction and/or symptoms. Vaginal septoplasty is the preferred approach in the setting of haematocolpos or dyspareunia and laparoscopic resection of any haematosalpynx can also be performed. [4]
Differential Diagnosis List: Uterus didelphys with obstructing hemivaginal septum and ipsilateral renal agenesis, Bicornuate uterus, Unicornuate uterus with rudimentary contralateral horn

Final Diagnosis: Uterus didelphys with obstructing hemivaginal septum and ipsilateral renal agenesis

References:

Robbins JB1, Parry JP, Guite KM, Hanson ME, Chow LC, Kliewer MA, Sadowski EA. (2012) MRI of Pregnancy-Related Issues: Müllerian Duct Anomalies. AJR Am J Roentgenol 198(2):302-10 (PMID: 22268172)
Grimbizis GF1, Gordts S, Di Spiezio Sardo A, Brucker S, De Angelis C, Gergolet M, Li TC, Tanos V, Brölmann H, Gianaroli L, Campo R. (2013) The ESHRE/ESGE consensus on the classification of female genital tract congenital anomalies. Hum Reprod 28(8):2032-44 (PMID: 23771171)
Junqueira BL1, Allen LM, Spitzer RF, Lucco KL, Babyn PS, Doria AS. (2009) Müllerian duct anomalies and mimics in children and adolescents: correlative intraoperative assessment with clinical imaging. Radiographics 29(4):1085-103 (PMID: 19605658)
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Epelman M1, Dinan D, Gee MS, Servaes S, Lee EY, Darge K. (2013) Müllerian duct and related anomalies in children and adolescents. Magn Reson Imaging Clin N Am 21(4):773-89 (PMID: 24183525)
Behr SC1, Courtier JL, Qayyum A. (2012) Imaging of Müllerian Duct Anomalies. Radiographics 32(6):E233-50

(PMID: 23065173)

Figure 1

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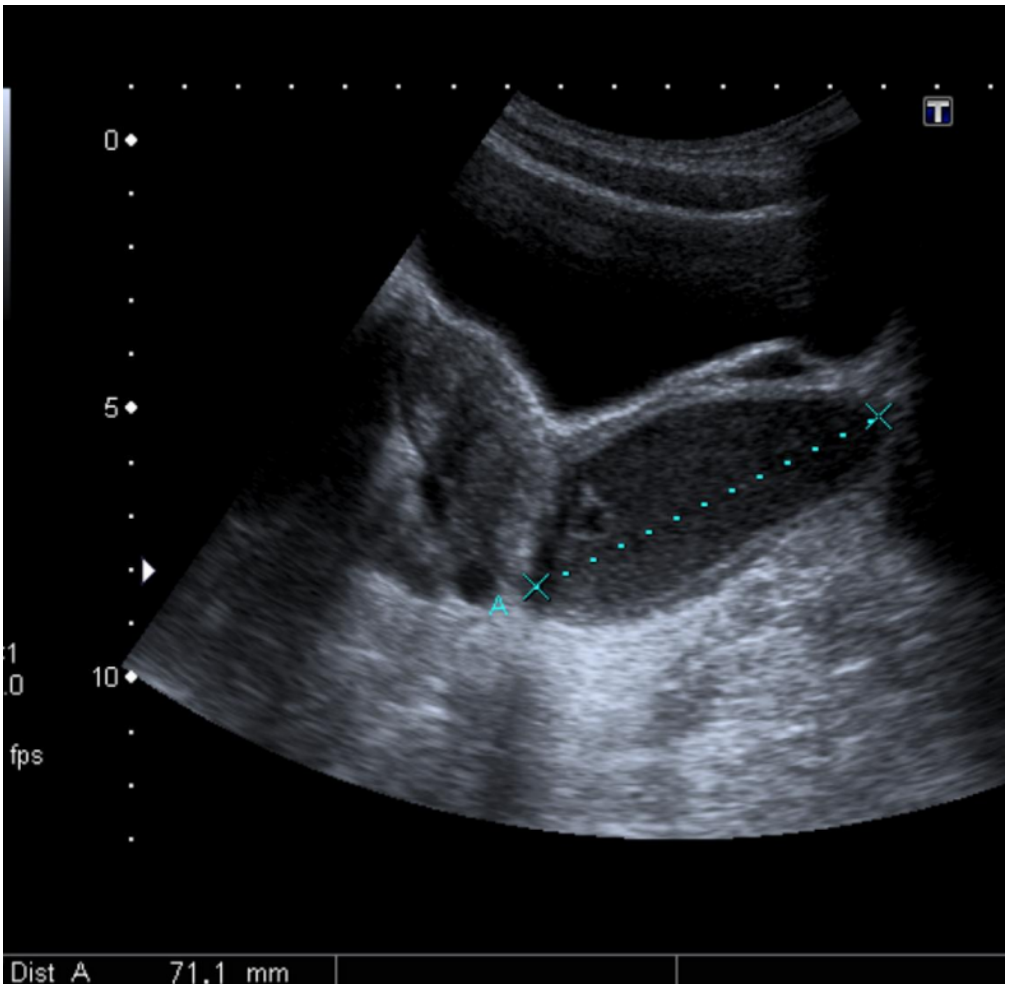

Description: Sagittal transabdominal US image shows an elliptical echogenic mass, which represents the vaginal cavity markedly distended with blood products (haematocolpos). The uterus extends from the superior aspect of the mass. **Origin:** RDM, Department of Radiology, CHEDV, Portugal.

Figure 2

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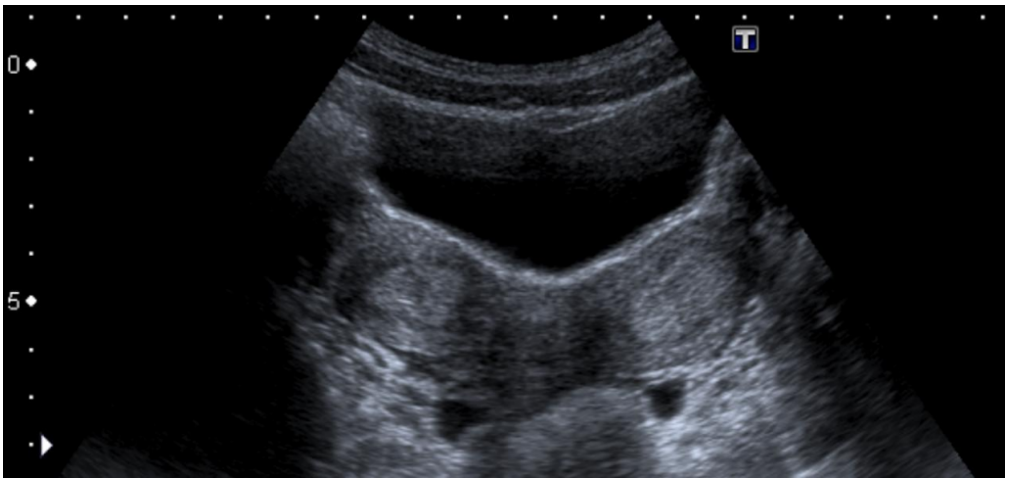

Description: Transverse transabdominal US image shows uterus didelphys, with two uterine horns and two separated endometrial cavities. **Origin:** RDM, Department of Radiology, CHEDV, Portugal.

Figure 3

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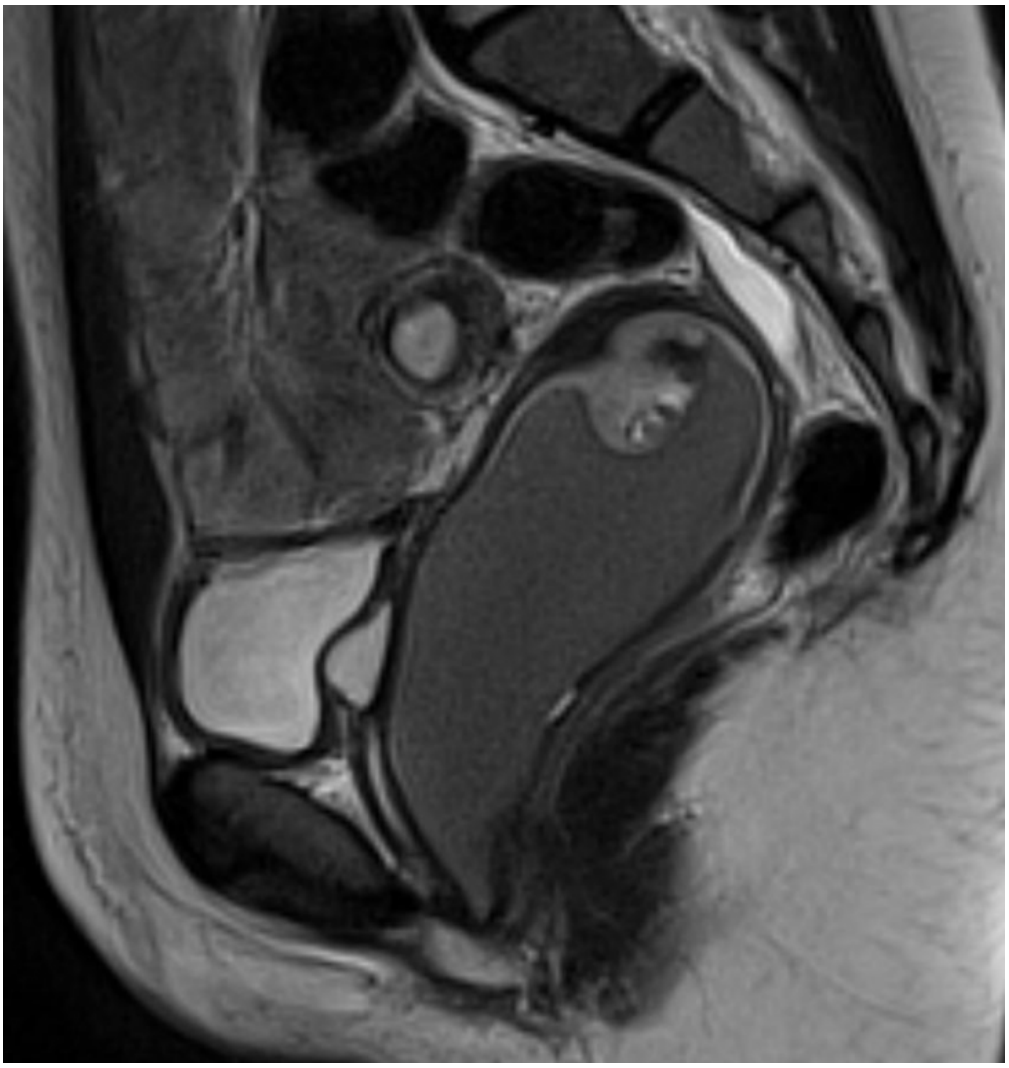

Description: MR image shows a dilated hemivagina whose content has signal characteristics of blood, forming a blind-ending pouch secondary to obstruction. **Origin:** RDM, Department of Radiology, CHEDV, Portugal.

Figure 4

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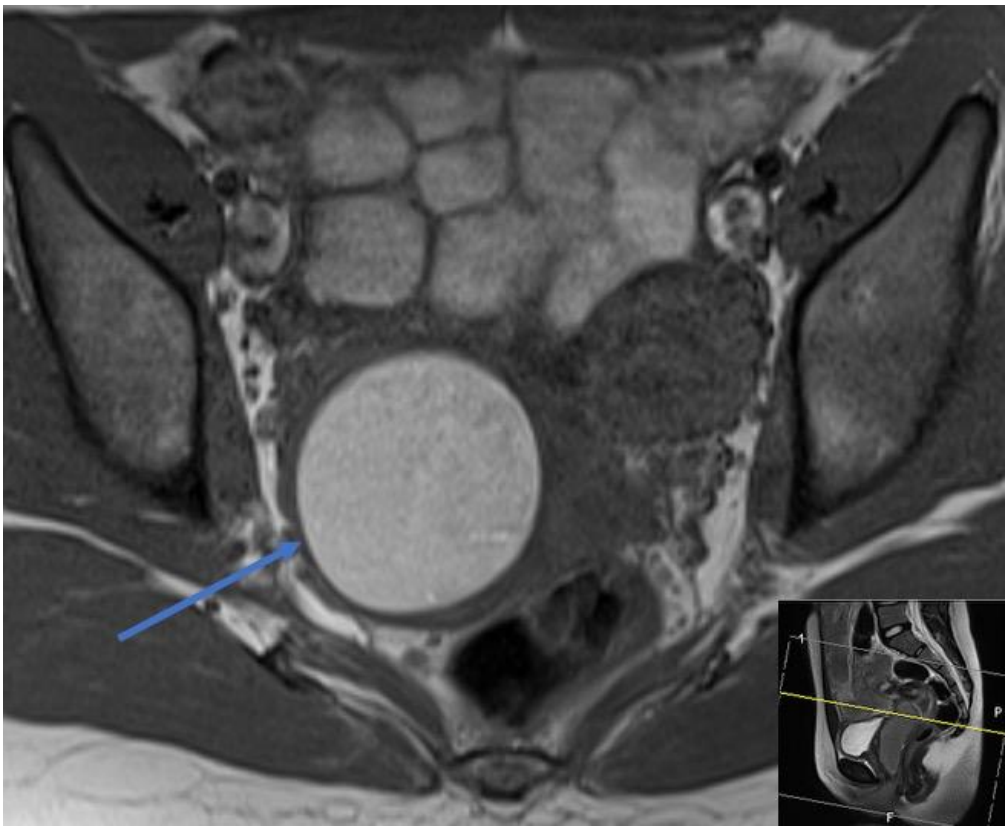

Description: A large haematocolpos centrally (arrow), a finding that corresponds to the obstructed right hemivagina. Mild dilatation of the right endometrial cavity and a nondistended left endometrial cavity are also seen. **Origin:** RDM, Department of Radiology, CHEDV, Portugal.

Figure 5

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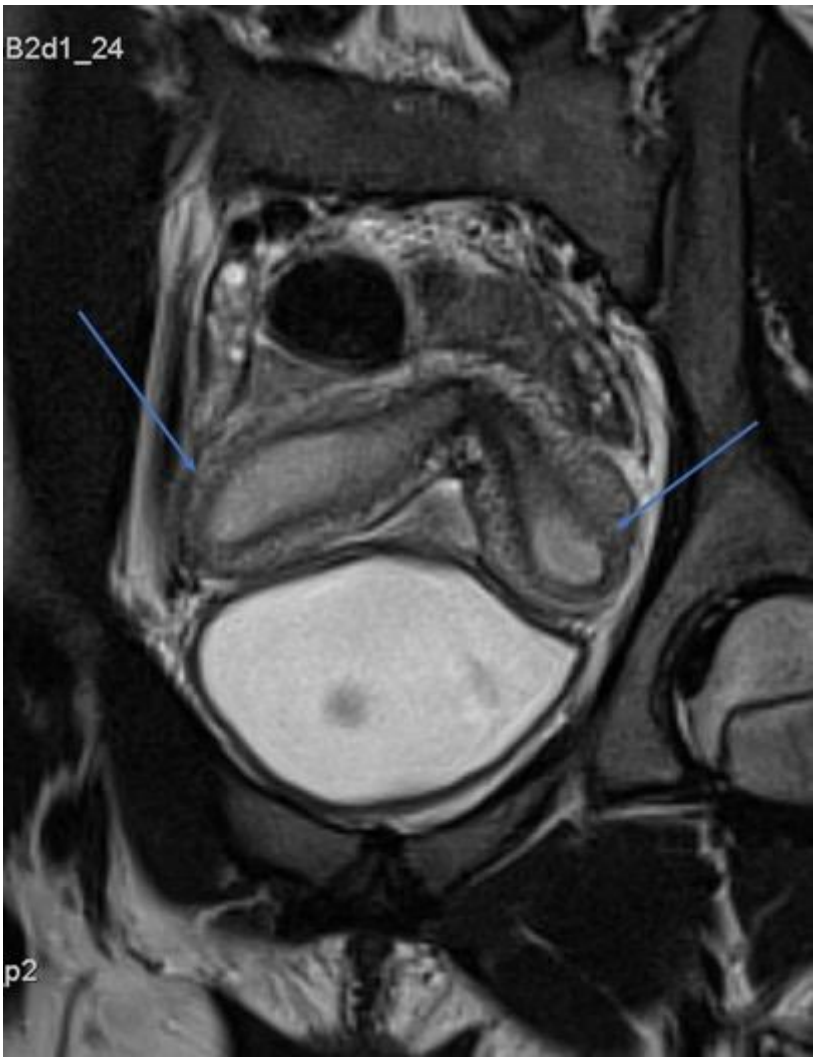

Description: Coronal T2-weighted image of a uterus didelphys, obtained in plane with the uterus, shows two widely divergent uterine horns (arrows) separated by a deep fundal cleft. **Origin:** RDM, Department of Radiology, CHEDV, Portugal.

Figure 6

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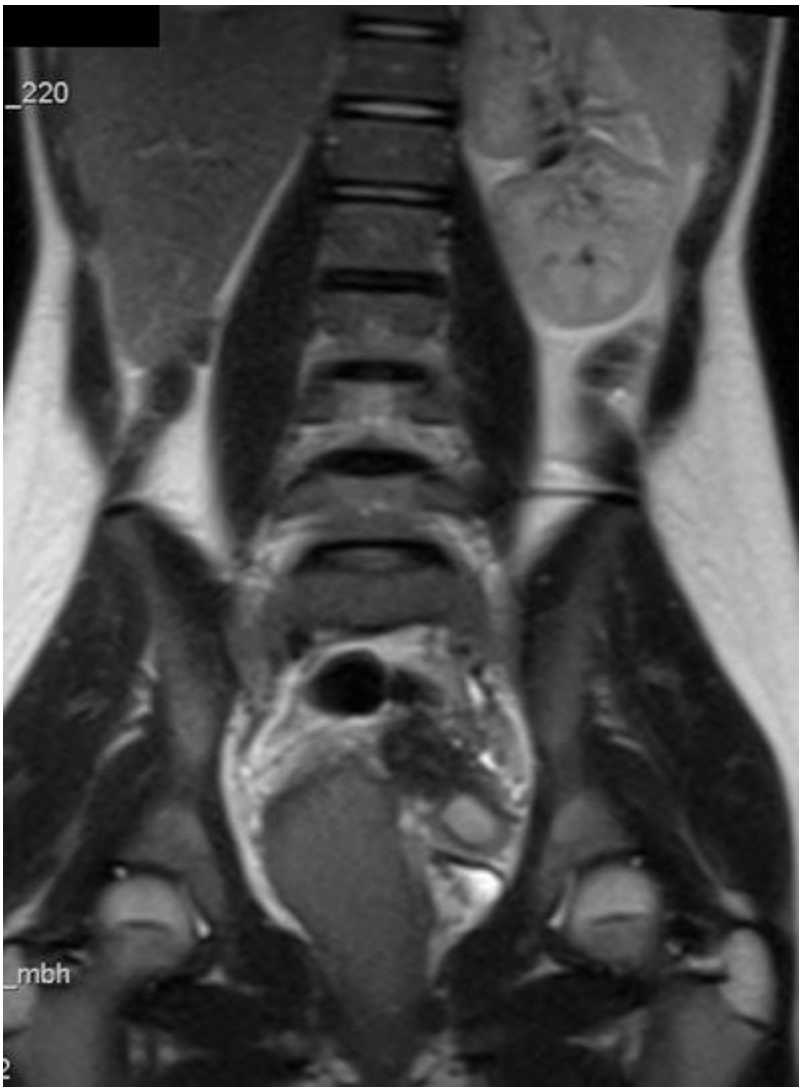

Description: Coronal fast spin-echo T2-WI shows a solitary left kidney, confirming renal agenesis with bowel loops in the right renal fossa, which is ipsilateral to the obstructed hemivagina. **Origin:** RDM, Department of Radiology, CHEDV, Portugal.

Figure 7

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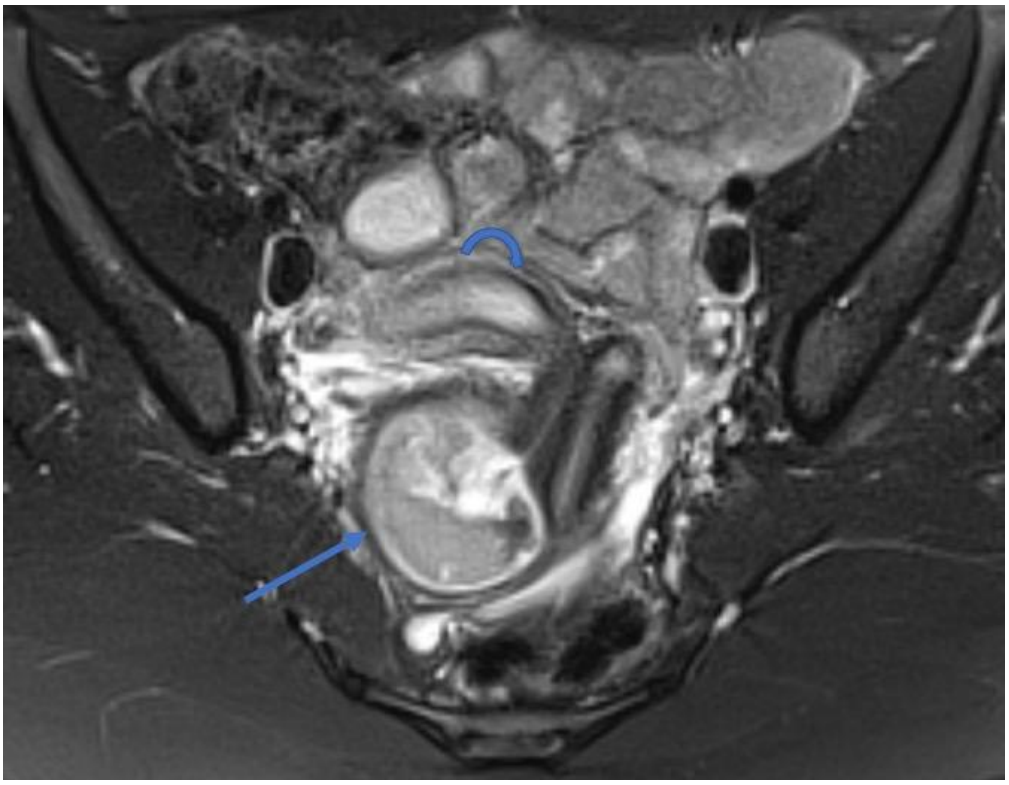

Description: Transverse oblique T2-WI obtained with fat suppression demonstrate communication of the right uterine horn (curved arrow) with the dilated left hemivagina (arrow); the appearance of the left uterine horn is normal. **Origin:** RDM, Department of Radiology, CHEDV, Portugal.

Figure 8

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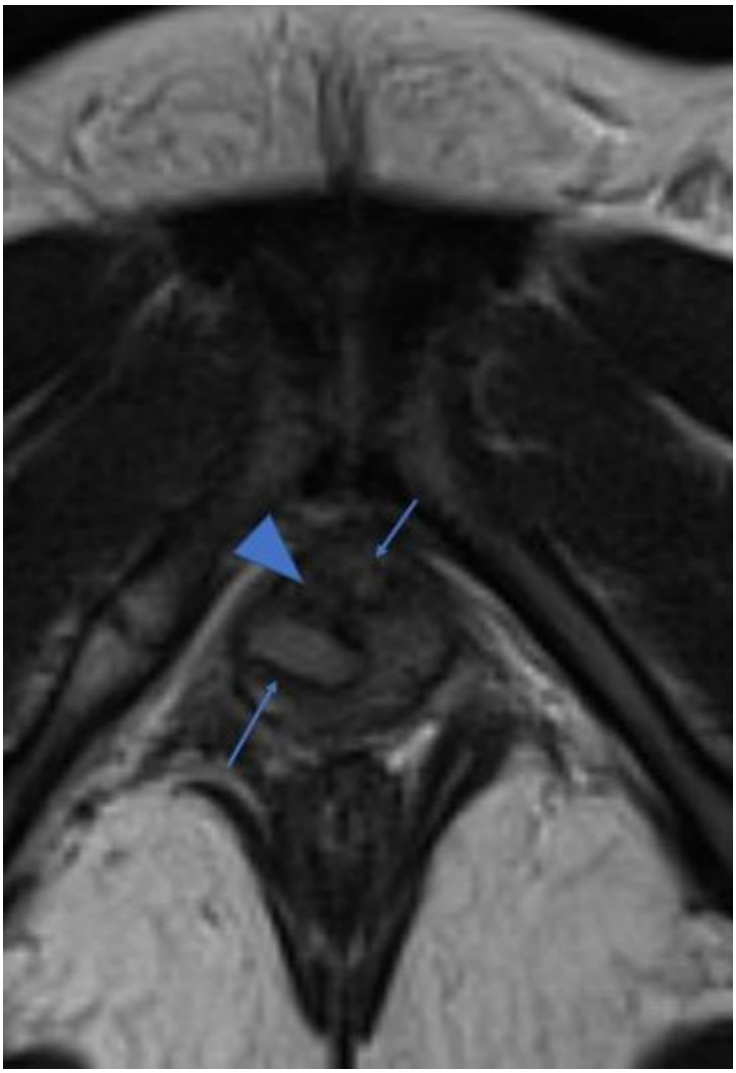

Description: The distal third of the two hemivaginas (arrows) separated by a band of fibrous tissue (arrowhead); the obstructed right hemivagina is already dilated, containing heterogeneous debris, which indicates distal obstruction. **Origin:** RDM, Department of Radiology, CHEDV, Portugal.