

Neonatal hydromucometrocolpos: prenatal diagnosis with MRI

Hidromucometrocolpos neonatal: diagnóstico prenatal por RM

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Summary
The diagnosis of fetal malformations remains a challenge, despite advances in obstetric gynecology and fetal ultrasound. Hydromucometrocolpos is defined as a vaginal and uterine dilation caused by an accumulation of mucinous secretions usually due to an obstruction or malformation of the genital tract. We present a case of hydromucometrocolpos diagnosed by MRI in a 34-week fetus who presented an abdominal cystic mass in prenatal ultrasound. Upon birth, an imperforated hymen was detected with posterior drainage of the hidromucometrocolpos.

Resumen
El diagnóstico de las malformaciones fetales continúa siendo un reto, a pesar de los avances en obstetricia y ecografía fetal. El hidromucometrocolpos se define como una dilatación de la vagina y la cavidad uterina ocasionada por un cúmulo de secreciones mucosas debido habitualmente a una obstrucción del tracto genital o a una malformación. Se presenta un caso de hidromucometrocolpos diagnosticado por resonancia magnética en un feto de 34 semanas en el que se encontró una masa quística abdominal en la ecografía prenatal abdominal. Tras el parto se detectó en la exploración física un himen imperforado, con posterior drenaje del hidromucometrocolpos y reparación del himen.

Case report

Pregnancy of 34 weeks with a female fetus. Prenatal ultrasound showed dilatation of the vagina and uterine cavity of the fetus with abundant homogeneous content inside (Figure 1). Ultrasound examination showed no uterine, cervical or vaginal septa. In view of these findings, a fetal MRI was requested, which confirmed urethral and vaginal dilatation, reaching the vulvar introitus, associated with abundant intraluminal content in the uterus and vagina. This content presented a low signal in T1 sequences and a slight hyperintensity in T2 sequences, although with less signal than the content of the urinary bladder, findings that were suggestive of mucous content (Figure 2).

No ovarian masses, signs of compression of the urinary tract or malformations in the rectum, urinary bladder or urethra were observed. No fistulous tracts were identified with the bladder or rectum. In view of these findings, a diagnosis of imperforate hymen was proposed, which was confirmed and treated after delivery.

Discussion

Congenital malformations of the uterus occur in 0.5% of the population and vaginal malformations in 0.025% (1). Hydromucometrocolpos is an extremely rare condition in the fetus and neonate and consists of dilatation of the vagina and uterine cavity by an accumulation of mucous secretions or urine inside. Two types are defined: secretory (hydromucometrocolpos) and urinary (urometrocolpos) (2). The first is usually secondary to obstructive pathology of the lower third of the vagina, uterus or cervix that leads to an accumulation of intrauterine secretions produced by the estrogenic action of maternal hormones. Its main complication is ureterohydronephrosis due to its compressive effect on the distal third of the ureters, followed by ascites (1-3).

On the other hand, urometrocolpos is related to malformations of the urogenital sphere such as urogenital sinus or cloaca (2).

Imperforate hymen affects approximately 0.1 % of girls born at term and is the main cause of hydromucometrocolpos (1, 2), but is rarely associated with it during the fetal stage. This is because the accumulation of intrauterine secretions in the fetus is secondary to increased maternal estrogen levels which are usually kept low during the perinatal stage. Therefore, it usually debuts in adolescence, when hormone levels rise again, causing a characteristic clinical picture of cyclic abdominal pain and amenorrhea. Other less frequent causes of obstructive hydromucometrocolpos are vaginal stenosis and septum, vaginal atresia or hypoplasia, and cervical stenosis (2, 3).

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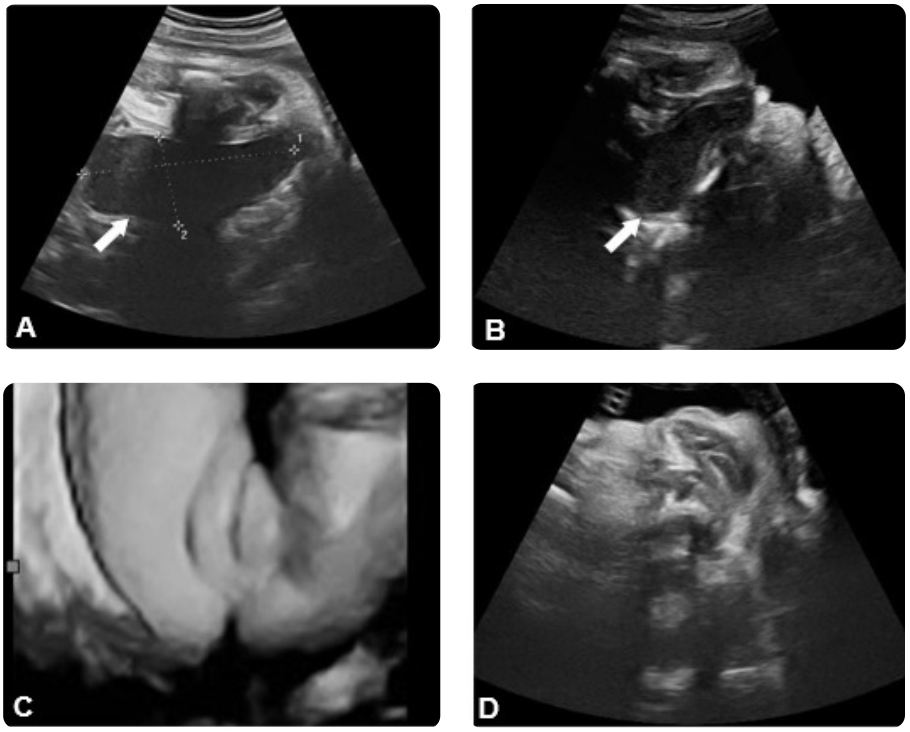


Figure 1. First case. Gestational age: 34 weeks. Hydromucometrocolpos due to imperforate hymen. a) Sagittal plane ultrasound. b) Coronal plane ultrasound. Dilatation of the vagina and uterine cavity with contents inside (white arrows) without presence of uterine, cervical or vaginal septa. c and d) 3D surface reconstruction of the vulva.

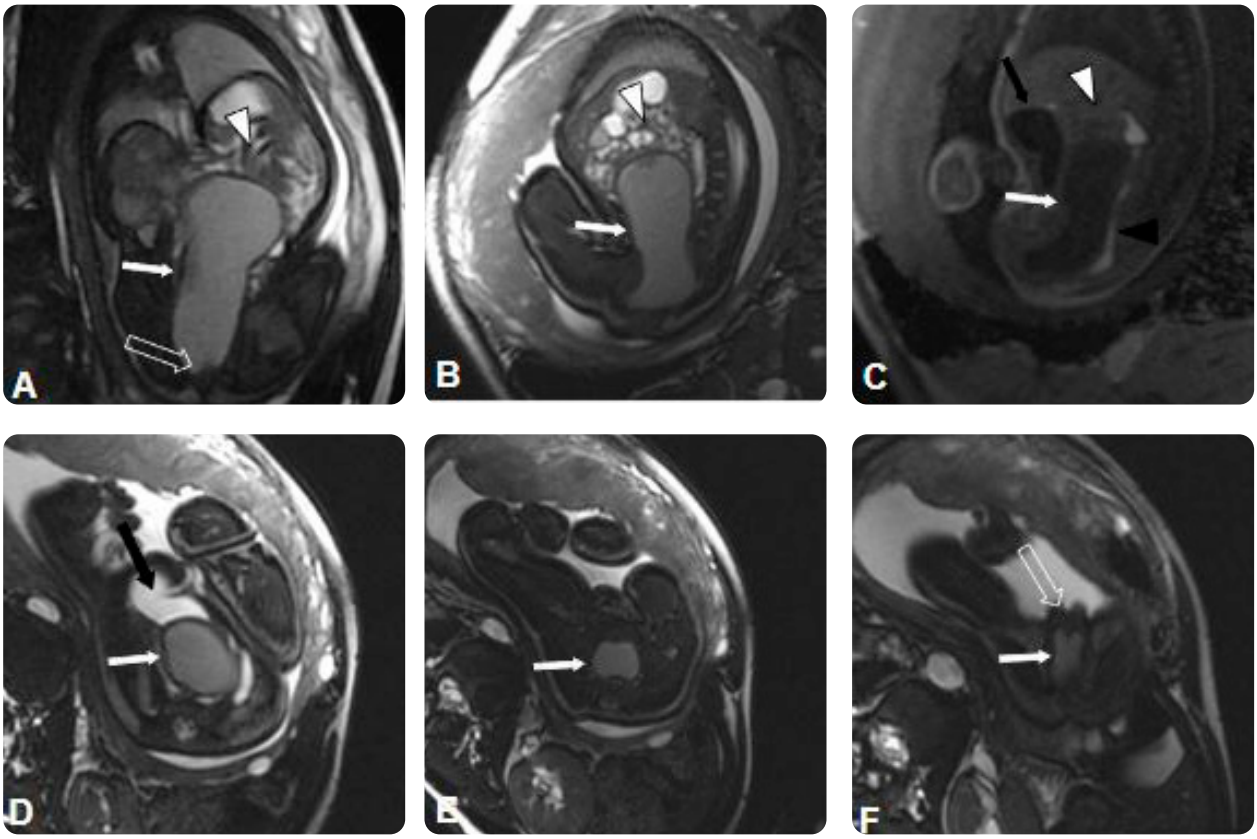


Figure 2. First case. EG: 34.3 s. Hydromucometrocolpos due to imperforate hymen. Fetal MRI. a) Coronal FIESTA. b) Sagittal FIESTA. c) Sagittal 3D T1-gradient echo. d-f) Axial FIESTA. Dilatation of the vagina and uterine cavity with hypointense content in its interior in T1 and slightly hyperintense in T2 suggesting mucosal content (white arrows). The dilatation reaches the vulvar introitus (hollow white arrow) without the presence of uterine, cervical or vaginal septa. It displaces in a superior and right lateral direction the bladder (black arrow) and the small bowel loops (white arrowheads) and compresses the rectosigmoid and descending colon (black arrowhead).

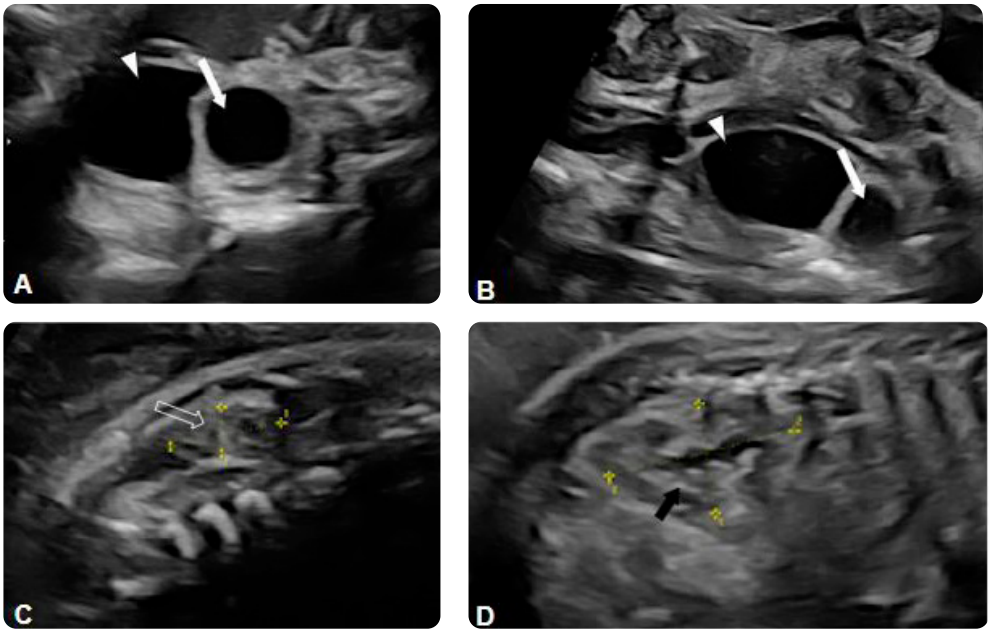


Figure 3. Second case. Gestational age: 27 weeks and 5 days. a-d) Fetal ultrasound. Hydromucometrocolpos (white arrowheads) displacing the bladder in anterior and superior direction (white arrows). The left kidney of normal dimensions (black arrow) and right kidney of small size (hollow white arrow) in relation to atrophy or hypoplasia.

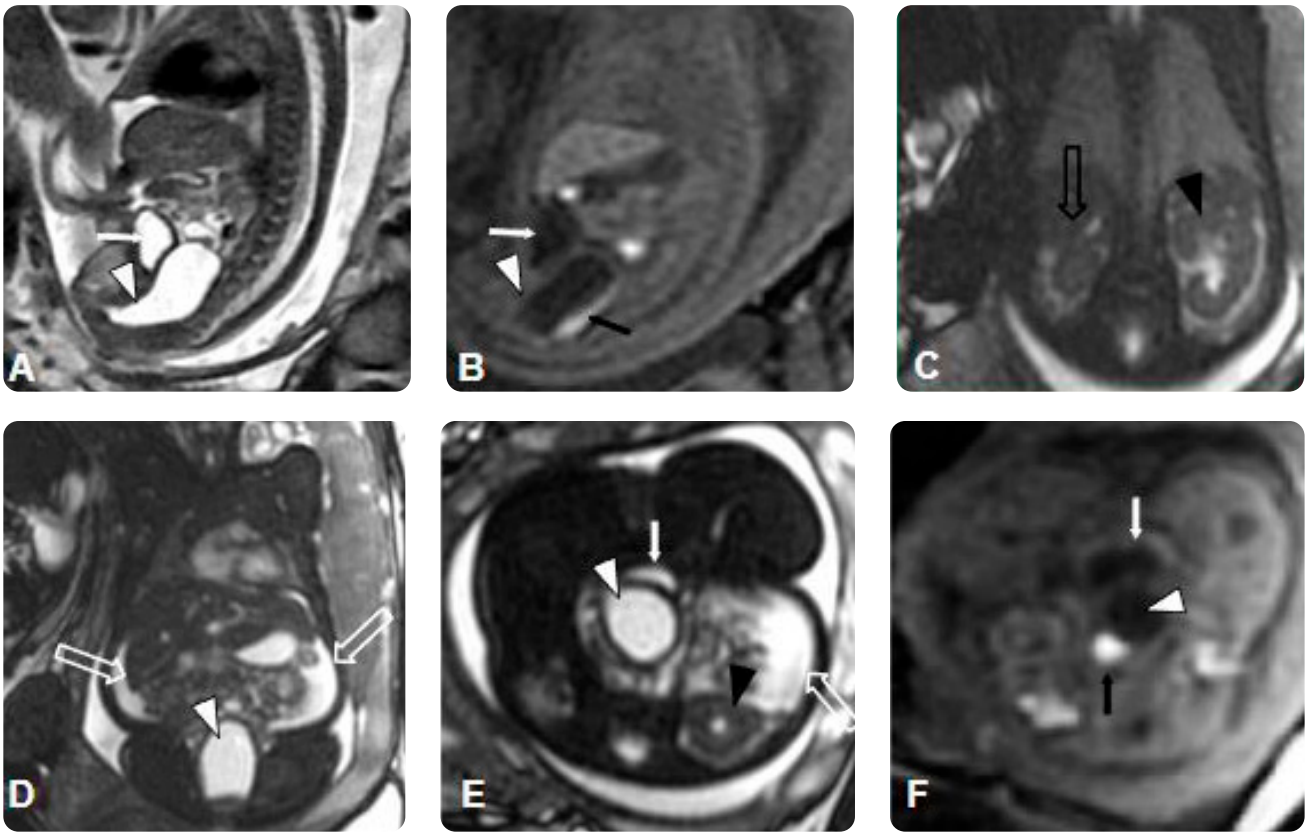


Figure 4. Second case. Gestational age: 28 weeks and 1 day. MRI. a) Sagittal SS FSE T2. b) 3D Sagittal LAVA. c) Coronal SS FSE T2. d) Coronal FIESTA. e) Axial FIESTA. f) 3D axial LAVA. Hydromucometrocolpos (white arrowheads) that displaces the bladder in anterior and superior direction (white arrows). The signal of both structures is similar, low signal in T1 and high signal in T2 suggesting communication of the content and suspicion of urogenital sinus (cases of isolated hydromucometrocolpos show low signal with respect to the bladder in T2 sequences). It is accompanied by moderate ascites of probable urinary origin. There is no dilatation of the rectum (black arrows), no communication between the rectum and the uterus and no enteroliths are visualized. The left kidney is of normal size and is accompanied by mild pyelocaliceal ectasia and dilatation of the left ureter along its entire course. The right kidney is small in size in relation to atrophy or hypoplasia (hollow black arrow). It is accompanied by moderate ascites of probable urinary origin (hollow white arrows).

Most cases of hydromucometrocolpos are sporadic, although this entity has been associated with various malformations, mainly of the urinary tract (ureteral duplication, urethral membrane and multicystic or hypoplastic kidneys). Likewise, its association with McKusick-Kaufman, Bardet-Biedl, Langer-Giedion and Pallister-Hall syndromes, among others, has also been described (2).

Prenatal diagnosis can be made by ultrasound, in which the uterus is visualized as a large cystic mass in the pelvis with abundant contents inside, usually hypoechogenic; although, occasionally with echoes inside in case of superinfection or bleeding (3).

The differential diagnosis includes (4): ovarian cysts or masses, gastrointestinal duplication cysts and a wide spectrum of bladder or rectal malformations. An attempt has been made to look for some specific sign of hydromucometrocolpos that allows its distinction from other pathologies that may appear in the prenatal period, mainly ovarian cysts. Sometimes two cystic images of different sizes (the uterus and the vagina) can be visualized, communicated by a small tract corresponding to the cervix, which together form the so-called "fjord sign" (5).

In this context, the development of fetal MRI has been a great advance in this field, since it allows obtaining high resolution images in several orthogonal planes, does not emit ionizing radiation and in most cases does not require maternal sedation (6, 7).

The first step in the study of this type of cystic lesions in the fetal pelvis is the exclusion of a cloacal malformation, due to the implications and therapeutic difficulties that this entails (4). The rectum is located anterior to the sacrum in the sagittal plane and can be visualized as a hyperintense column in T1 sequences, due to the increase of its meconium content after 22 weeks of gestation. An abnormally elevated location of the distal rectum with a hyperintense dilated colon on T2 sequences suggests urinary contents; a hyperintense content on T1 sequences in the uterus, vagina or bladder indicates the presence of meconium (4).

Once a rectal malformation has been excluded, obstructive genital malformations with mucus accumulation or genitourinary malformations with urine accumulation in the uterine/vaginal cavity should be ruled out.

Some studies have postulated that the comparison of signal intensity between bladder and uterine contents can help to differentiate both entities (4). Mucous secretions in the context of hydromucometrocolpos give the uterus a hypointense appearance in T2 sequences with respect to the bladder. In cases of urogenital sinus malformation and hydromucometrocolpos, the contents of the vagina and uterus present a marked hyperintensity in T2 sequences, similar to the bladder contents (4).

A second case of fetal hydromucometrocolpos was studied in this center due to the finding of ureterohydronephrosis in obstetric ultrasound (Figure 3). Although the fetal MRI subsequently performed showed frank T2 hyperintensity in both the uterus and bladder (Figure 4), at birth she presented an imperforate hymen without evidence of urological malformations.

Both pathologies can be accompanied by bladder obstruction, hydroureteronephrosis and urinary ascites.

Treatment of hydromucometrocolpos consists of drainage of the accumulated fluid, resolution of the obstruction of the genital tract by reestablishing communication between the vagina and vulva, and repair of concomitant malformations (8). Delay in treatment is associated with complications such as renal damage due to the compressive effect on the urinary tract, development of endometriosis, pyocolpos and uterine perforation (2).

Conclusion

Hydromucometrocolpos is an infrequent finding during the gestational stage and its most frequent cause is imperforate hymen. Fetal MRI can play a fundamental role as a complement to ultrasound in the prenatal stage.

References

1. Tang Ploog LE, Barrón López AR, Ayque Rosas FF. Hidrometrocolpos neonatal: a propósito de un caso de difícil diagnóstico. Rev. peru. ginecol. obstet. [Internet]. 2021 Oct [citado: 2022 dic 19];67(4):00019. Disponible en: http://www.scielo.org.pe/scielo.phpscript=sci_arttext&pid=S2304-51322021000400019&Ing=es
2. Calvo García MA. Complex urogenital and anorrectal malformations. En BM Kline-Fath, DL Bulas, R Bahado-Singh, Fundamental and advanced fetal imaging ultrasound and MRI, 2º Ed. Philadelphia: Wolters Kluwer; 2021. p. 833-846.
3. Madero G, Echeverry LM, Jiménez YL. Diagnóstico neonatal de hidrometrocolpos. Pediatria. [Internet]. 1 de julio de 2015 [citado: 2022 dic 19];48(3):75-9. Disponible en: https://revistapediatria.org/rp/article/view/38.
4. Capito C, Belarbi N, Paye Jaouen A, et al. Prenatal pelvic MRI: additional clues for assessment of urogenital obstructive anomalies. J Pediatr Urol. 2014;10:162-6.
5. Schumacher R, Brzezinska R. Hydrometrocolpos: The Fjord Sign. Ultraschall in der Medizin-European J Ultrasound. 2007;28(05):518-20. doi:10.1055/s-2007-963211
6. Millischer AE, Grevent D, Rousseau V, et al. Fetal MRI compared with ultrasound for the diagnosis of obstructive genital malformations. Prenat Diagn. 2017;37:1138-45.
7. Picone O, Laperelle J, Sonigo P, et al. Fetal magnetic resonance imaging in the antenatal diagnosis and management of hydrocolpos. Ultrasound Obstet Gynecol. 2007;30:105-9.
8. Bischoff A, Levitt MA, Breech L, Louden E, Peña A. Hydrocolpos in cloacal malformations. Pediatr Surg. 2010;45:1241-5.

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