Cystic masses and pseudomasses in the fetal pelvis: a differential diagnosis based on fetal MRI and US findings.

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Learning objectives

The purpose of this pictorial essay is to develop a practical approach to the interpretation of pelvic cystic masses observed in utero, based on a retrospective review of the prenatal imaging studies performed in our institution, with emphasis on fetal MRI exams. Practical teaching points to determine the anatomic origin of the lesion and to limit the possible differential diagnosis will be described.

Background

Fetal anomalies are always a stressing condition for parents and physicians. The ability to diagnose anomalies in utero has continuously improved in the last two decades as a result of the generalization of prenatal ultrasound (US) screening imaging exams. Although most fetal pelvic masses are detected at US, prenatal MRI is increasingly being used in inconclusive or complex cases, as it may bring additional information in terms of the anatomic origin, extension and tissue characterization of the detected pathology. Thanks to the technical developments in prenatal imaging techniques, radiologists are now able to better clarify the nature of a pelvic mass in utero in a significant percentage of cases.

Pelvic masses account for 10% of all fetal masses detected on prenatal screening US. They often have a cystic appearance at imaging. The diagnosis of a "pelvic cyst" is wide, as it may correspond to a tumoral pathology, a congenital anomaly or be the result of an anomalous accumulation of liquid in a pelvic whole organ, mainly the urine bladder in males and the vagina and/or uterus in females. Therefore, the differential diagnosis of pelvic cystic lesions and pseudo-lesions changes according to the fetus’s sex. The most often detected pathologies appear in Table 1.

<table>
<thead>
<tr>
<th>URO-GENITAL</th>
<th>MALES</th>
<th>BOTH</th>
<th>FEMALES</th>
</tr>
</thead>
<tbody>
<tr>
<td>URO-GENITAL</td>
<td>LUTO(^{(1)})</td>
<td>Urachus anomalies</td>
<td>Ovarian cysts</td>
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<tr>
<td></td>
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<td></td>
<td>Hymen imperforatus</td>
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<td></td>
<td></td>
<td></td>
<td>Uterine anomalies</td>
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<tr>
<td></td>
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<td></td>
<td>Cloacal dysgenesis</td>
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<tr>
<td>DIGESTIVE</td>
<td></td>
<td>Rectal duplication cyst</td>
<td>MMIH syndrome(^{(2)})</td>
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<tr>
<td>TUMORAL</td>
<td></td>
<td>Sacrococcygeal teratoma</td>
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When a cystic appearing mass is detected in utero, the following issues should be addressed: is it a congenital lesion or an anomalous distended whole pelvic organ?; is it really pathologic?; which is its anatomic origin and the most possible diagnosis?; which are the repercussions over the adjacent fetal organs? and the implications for pregnancy, including fetal prognosis?; when and how should follow up occur?

Findings and procedure details

All fetal MRI performed in our Institution between 2007-2013 were reviewed. Patients with pelvic cystic lesions or anomalous liquid distension of pelvic whole organs were selected. For this pictorial review, we have evaluated the prenatal US and MRI images and their suggested diagnosis and compared then with the final diagnosis, obtained from autopsies, pathologic exams and/or postnatal imaging studies. All MRI studies were performed in a 1,5 Tesla scanner. The usual fetal MR protocol is described in Table 2 and include T1-VIBE; T2-TRUE FISP and T2-HASTE sequences obtained in all three spatial fetal planes.

<table>
<thead>
<tr>
<th>Slice thickness (mm)</th>
<th>TR (msec)</th>
<th>TE (msec)</th>
<th>Angle</th>
<th>FOV</th>
<th>Matrix</th>
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</thead>
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<td>88</td>
<td>135</td>
<td>350X350</td>
</tr>
<tr>
<td>T2-TRUEFISP</td>
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<td>3,22</td>
<td>70</td>
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<tr>
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<td>1,6</td>
<td>12</td>
<td>255x340</td>
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</tbody>
</table>

Table 2.- Usual protocol (sequences parameters) for fetal imaging.
Once a cystic appearing lesion is detected at prenatal US exam, determination of the sex of the fetus is the first step to establish a possible differential diagnosis (Table 1). If the US is incomplete, technically deficient or in case of large, complex lesions, fetal MRI would be performed to improve the information about the lesion and to orientate the therapy during pregnancy and in the immediate postnatal period.

In the mid-line sagittal plane in male fetuses, the pelvis can be divided in two compartments: the anterior one, containing the genito-urinary organs and limited dorsally by the posterior wall of the urine bladder and the posterior pelvis, containing the distal bowel, the presacral space and the sacrum. Figure 1 shows the anatomic schema and the division of the pelvis for males.

For female fetuses, the pelvis can be divided in three regions: the anterior one, containing the urine bladder and once more, limited by the posterior wall of the bladder; the middle pelvis, with the genital organs (vagina and uterus) and the posterior one, limited ventrally by the anterior wall of the rectal ampulla and containing the rectum, the presacral space and the sacrum. Figure 2 shows the anatomic schema with the division of the pelvis for females.

T1- and T2-weighted images obtained in the midline sagittal plane are extremely helpful to identify the anatomic origin of a pelvic cyst or pseudocyst. This plane allows an easy identification of the fetal bladder, filled with urine, that appears homogeneously hyperintense on T2- and hypointense on T1-weighted MR images. The bladder should be visible in all fetuses after the 12th GW.(1). The normal urine voiding time in utero is every 55-155 minutes, but in normal fetuses, the fetal bladder is never completely empty (2), even after voiding, which allows its identification. The distal rectum contains meconium and should also be visible. It is observed after the 20th GW as a tubular structure, hyperintense on T1-weighted images (Fig 3). In case of pelvic masses, the direction of the displacement of the bladder and the rectum helps to recognize the location of the pathology and limits its differential diagnosis (Fig. 4).

The anatomic location of the most important cystic images in the fetal pelvis is described in Table 3 for males and in Table 4 for females. Ovarian cysts and mesenteric cysts may be found not only in the pelvis, but also in the fetal abdomen, whereas lymphangiomas do not respect the anatomical planes.

**Table 3.**

<table>
<thead>
<tr>
<th>Males</th>
<th>Anterior pelvic space</th>
<th>Posterior pelvic space</th>
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</thead>
<tbody>
<tr>
<td></td>
<td>LUTO</td>
<td>Rectal duplication cyst</td>
</tr>
</tbody>
</table>
Urachus anomalies | Sacrococcygean teratoma (SCT)  
Anterior meningocele

Table 4.

<table>
<thead>
<tr>
<th>Females</th>
<th>Anterior pelvic space</th>
<th>Middle pelvic space</th>
<th>Posterior pelvic space</th>
<th>pelvic space</th>
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<tr>
<td></td>
<td>LUTO</td>
<td>Cloacal dysgenesis</td>
<td>Rectal duplication cyst</td>
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<tr>
<td></td>
<td>Urachus anomalies</td>
<td>Hydrometrocolpos</td>
<td>SCT</td>
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<tr>
<td></td>
<td></td>
<td>Uterine anomalies</td>
<td>Ant. Meningocele</td>
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</tbody>
</table>

**Ovarian cysts** are the most frequently accounted pelvic cysts in female fetuses. Ovarian cysts are easily identifiable at US and they rarely require additional prenatal imaging. Most of them are simple, benign, functional cysts resulting from excessive stimulation of the fetal ovaries by the placental and maternal hormones (3). US detection occurs mostly between the 23th and the 35th GW. Ovarian cyst should be considered as the first possible diagnosis in a female fetus with a cystic structure in the pelvis or lower abdomen and with normal urinary and gastrointestinal tracts. Their sonographic appearance depends on the size of the cyst and the presence or not of complications. Simple cysts are anechoic and may be uni- or multilocular, as they can occasionally have internal septations (Fig 5). Large cysts have an increased risk of torsion and hemorrhage and may present internal echos, fluid-fluid levels and thickened septa (Fig.6-7). The natural course of ovarian cysts is spontaneous, slow regression post natally.

Isolated congenital **hydrocolpos or hydrometrocolpos** is very uncommon. It arises from the accumulation of secretions from the reproductive glands and the impairment in its evacuation, caused mostly by an imperforated hymen, a midline vaginal septum or a vaginal atresia (4-6),(Fig 8). Its sonographic appearance is a well-defined, cystic pelvic mass in the middle pelvic space of a female fetus. The distended vagina, filled with liquid or with debris, shows a pear-inversed form, with a recognizable uterus on top, that may or not be distended (5). Diagnosis occurs mostly during the third trimester. A full bladder helps to differentiate the fluid filled genitalia from the urinary system. Hydrocolpos is often associated with hydronephrosis, because of the posterior compression of the bladder. Occasionally, ascites may be observed.

**Cloaca** is an extremely rare malformation seen in female fetuses with an incidence of 1 in 50000 studies (7). In cloacal malformation there is a single common perineal
opening for the genital, urinary and gastrointestinal tracts (Fig.9). It is thought that the anomaly results from the failure of the uroseptal septum to join the cloacal membrane between the 4-6thw. of embryonic development. Cloaca can lead to bladder obstruction, hydrometrocolpos and/or colonic dilatation. Nevertheless, the final diagnosis is first postnatal in most of the cases, with identification of a single perineal orifice (8). These children require a complete postnatal evaluation, as cloaca are often associated with multisystemic anomalies. Genital duplication is often observed, as in our case (Fig.10).

**MMIH syndrome** is a rare condition characterized by a massively dilated urinary bladder, incomplete intestinal rotation, microcolon, and small bowel dilatation (8,9). It occurs predominantly in girls and its exact cause is unknown. The typical prenatal findings include megacystis and hydronephrosis with absence of polyhydramnios. Documentation of the fetal sex is important because this entity occurs almost exclusively in females, thus helping to differentiate it from other forms of LUTO.

**LUTO**, (acronym of fetal Lower Urinary Tract Obstruction) is characterized by a wide variety of pathologies causing obstruction of the lower urinary tract such as post-urethral valves (PUV’s), urethral stenosis and urethral atresia. PUV accounts from almost half of LUTO cases and its incidence is 1 per 8000 to 1 per 25000 births in males (8). The exact etiology is unknown but it is probably due to an abnormal insertion of the mesonephric ducts to the fetal cloaca. Early LUTO is associated with a high fetal morbidity and mortality, consequence of a severe hydronephrosis, with secondary cystic renal dysplasia, resulting in abnormal renal function. Moreover, renal compromise can lead to severe oligohydramnios, often associated with secondary pulmonary hypoplasia. Imaging findings suggesting the diagnosis of LUTO include a large, thick-walled bladder and a dilated proximal urethra (classically known as the keyhole sign). As referred, oligohydramnios, renal cystic changes and occasionally hypoplastic lungs may be observed (Fig.11-12). Postnatal treatment of PUV consists in immediate post natal catheter placement for bladder drainage with secondary surgical cystoscopic valve ablation.

The **urachus**, or median umbilical ligament, is a midline tubular structure that extends upward from the anterior dome of the bladder toward the umbilicus. It usually involutes before birth. Failure of urachal obliteration can give raise to various abnormalities such as patent urachus (48%), urachal cyst (31%), urachal sinus (16%) and vesicourachal diverticulum (5%) (10). In case of patent urachus a urine leakage from the umbilicus is noted in the neonatal period. Sonographically the patent urachus is observed as a longitudinal, tubular structure connecting the bladder with the umbilicus (Fig.12). Urachus cysts are seen in 2 per 100.000 births (11). Their imaging finding is that of an anechoic anterior midline cystic structure, lying between the bladder and the umbilicus, either contiguous or separated from the bladder. In utero, the differential diagnosis between
urachus anomalies, bladder extrophy or omphalocele is sometimes difficult to make. The treatment is postnatal surgical excision in case of symptoms.

**Enteric duplication cysts** are rare lesions, with an incidence of about 1 per 10,000 births. They may arise from any point of the GI tract, with the ileum being the most often location. Although rectal duplication cysts are extremely rare, they should be considered in the differential diagnosis of cystic pelvic lesions in utero. They probably result from the failure of the intestinal lumen recanalization during embryogenesis. Sonographically they appear as an elongated, tubular or spherical unilocular cyst, close to and in contact to the bowel. Typically, they present a thick, multilayered wall. Duplication cysts may communicate or not with the intestinal lumen (12). In MRI the rectum can be identified after the 20th WG as a high T1 and low T2 signal structure, filled with meconium (Fig.13-14). Detection of peristalsis and the typical thickness of the muscular wall can be a distinguishing feature from other cystic lesions. Obstruction, intussusception and volvulus are frequent complications and therefore, the treatment is mostly the post natal surgical ablation. (13).

**Sacrococcygeal teratomas (SCT)** are the only cystic tumoral lesion in the fetal pelvis. Teratoms are the most common fetal tumors, with a reported incidence of one every 35000 to 40000 births and the pelvis is one of the most frequent locations. Because of the acoustic shadowing by fetal pelvic bones US cannot always define the most cephalad extent of the tumor. Following the **Altman/AAPSS classification**, the SCT are classified in 4 types, according with their anatomical location: type I (47% of cases, best prognosis), primarily external; type II (34%), dumbbell shaped with equal external and internal portions; type III (9%), primarily internal within abdomen and pelvis; type IV (10%, worse prognosis), entirely internal with no visible external component, (Fig.17).

They are typically heterogeneous at US imaging, with liquid and solid components. Moreover, they may present different echogenic patterns secondary to tumor necrosis, cystic degeneration, internal hemorrhage and/or calcification (Fig.15-16), (14). The prognosis depends mainly on their size, classification, anatomic extension and content of the tumor.

**Mesenteric cysts and lymphangiomas** are lymphatic cysts in origine. Although they can appear similar macroscopically, lymphangiomas are more frequently larger in size, multilocular, with fluid-fluid levels. Lymphangiomas do not respect the anatomical planes and can rapidly increase in size in case of infection or hemorrhage. Mesenteric cysts are smaller and usually located in the mid abdomen. They are mobile and their position may change at different US examinations (Fig.18), (1).
**Anterior sacral meningoceles** are characterized by the anterior herniation of sacral meninges secondary to focal erosion or associated to hypoplastic sacral and/or coccygeal vertebral segments. They are often observed as part of the Currarino triad (anorectal anomalies, caudal regression syndrome and presacral mass) or in conditions with prominent dural ectasia (NF1, Marfan, homocystinuria). The best diagnostic clue is an homogeneous presacral cystic mass which communicates with the thecal sac through an anterior sacral defect.

**Meconium peritonitis** from bowel perforation in utero can occasionally lead to a meconium pseudocyst. Features like dilated bowel, associated with peritoneal or bowel calcifications and ascites may help in the diagnosis (1).

Images for this section:
Fig. 1
Fig. 2

anterior pelvic space  middle pelvic space  posterior pelvic space
**Fig. 3:** Normal fetal pelvis in T2 sagittal plane in a male (a), and female (b) fetus, with a hypointense rectum (R) and hyperintense bladder (B). T1 sagittal plane (c) of a female pelvis with hyperintense rectum and hypointense bladder anteriorly.

**Fig. 4:** -(a) The rectum (R) which contains the hyperintense meconium, is displaced anteriorly by a voluminous, hypointense mass (SCT) in this T1-weighted MR image. (b) Observe the well-defined, hypointense mass (rectal duplication cyst) located posterior- and inferiorly to the rectum in this T1-weighted image (c). The distended, fluid-filled vagina (V), in this case of imperforated hymen, displaces the rectum posteriorly in this T1-weighted sagittal image (black arrows).
Fig. 5: Simple ovarian cysts. Examples of US images (a-c) of non-complicated, unilateral ovarian cysts in three different patients. Ovarian cysts are the most often detected pelvic and abdominal cysts in female fetuses and rarely require additional imaging studies. Internal, fine septations are usually observed.
Fig. 6: Bilateral, non-complicated ovarian cysts. Occasionally, ovarian cysts are bilateral as in this patient with voluminous, non-complicated cysts (a: bilateral; b: right ovary; c: left ovary)
**Fig. 7:** Hemorrhagic ovarian cyst. Axial- (a), sagittal- (b) and 3D (c) US images of an unilateral hemorrhagic ovarian cyst in a female fetus (24th w.) The internal septations are thicker and the cyst is heterogeneous

**Fig. 8:** Hymen imperforatus with secondary hydrocolpos. Axial (a) and sagittal T2-HASTE (b) MR images (35th w.) shows a retrovesical located, homogeneous hyperintense structure, that corresponds to the fluid-filled, distended vagina (V). Note the ventral displacement of the bladder (B) and the bilateral hydronephrosis (a); (B: Bladder) .(c) Sagittal T1-VIBE image shows the posteriorly displaced and compressed rectum (black arrow), hyperintense because of the meconium-filling.(d) Postnatal clinic exam reveals the protrusion of the non perforated hymen. No other anomalies were found
**Fig. 9:** Cloacal anomaly is characterized by a unique external perineal opening with a short (A) or long (B) common canal for the genital, urinary, and digestive systems.

**Fig. 10:** Cloacal anomaly with genital duplication. (a) Coronal T2-HASTE MR image (35th w.) shows the two symmetric pelvic cystic structures, which correspond to fluid-filled
hemivaginas in this female fetus with genital duplication (V). Parasagittal T2-HASTE (b) and T1-VIBE MR images (c) show one of the distended hemivaginas (V) as well as the fluid-filled hemiuteri (U). The bladder is anteriorly displaced (arrow) and the rectum is not identifiable in its normal position. Note the associated oligohydramnios.

Fig. 11: LUTO (Posterior urethral valves). Coronal and axial US images (12th w.) in this male fetus show a distended urine bladder (B) and evidence the dilated proximal urethra in a classic «keyhole» appearance (a, black arrow). Bilateral hydronephrosis and ureteral dilatation is also observed (b, yellow marks). Sagittal- (c) and 3D US reconstruction images (d) 2 w. later show increasing distension of the bladder and cranial displacement of the diaphragma.
**Fig. 12:** LUTO (Posterior urethral valves), patent urachus with giant urachus- and umbilical cyst. Coronal (a and b) and sagittal (c) T2-HASTE MR images show the distended urin bladder (B) with thick, irregular wall. Note the dilatation and the « keyhole » appearance of the proximal urethra (b and c, white block arrow) and the presence of urine ascites in this male fetus. The bladder dome is opened in a patent urachus (black arrow). The ureters are extremely dilated (U) and there is bilateral hydronephrosis with reduced size of the kidneys (black small arrows, b). The clinical imaging shows a giant urachus and umbilical cord cyst.

**Fig. 13:** Rectal duplication cyst. Axial US (a) and T2-HASTE (b) MR images (--th w.) at the level of the ischio-rectal fossae show the well defined cystic lesion (star), homogeneously anechogenic at US and fluid isointense on T2-WI, with thick cyst wall in this male fetus. Sagittal T2-HASTE MR image (c) evidences that the lesion is separated from the coxis (c). Sagittal T1-VIBE (d) MR image shows that the cyst is located posteriorly to the rectum (R), which is filled with protein rich, hyperintense meconium. (B: Bladder)
**Fig. 14:** Rectal duplication cyst. (a) Axial and (b) sagittal T2-HASTE MR images performed at day 4th shows the pelvic cyst, located posteriorly to- and exercising a mass effect over the rectum. The cyst presents a well defined, thick wall, characteristic for bowell duplication. Images confirm that the lesion is separated from the coxis. (c) Lateral view of contrast enema at day 4th reveals the mass effect of the cyst at the posterior wall of the rectum (black arrow). Diagnosis was confirmed at pathology.

**Fig. 15:** Parasagittal T2-TRUF1 (a) and T1-VIBE (b) and axial T2-TRUF1 MR images (c) (36th w.) show the voluminous, heterogeneous tumor and reveals its anatomic relationship with the coxis. The intrapelvic tumor (star), mostly cystic, displaces the bladder (B) cranially and the rectum ventrally (white arrows, c). The postnatal babygram shows the huge mass. Observe the enlarged distance between the ischial bones in this female baby.
**Fig. 16:** Sagittal (a) and axial (b) fetal US (-- th w.) images and coronal T2-HASTE MR image (c) show the presacral tumor (T), with solid (ST) and cystic tumor (CT) components. The tumor causes urethral obstruction with megabladder secondary bilateral hydronephrosis. Note the pyelocalicial dilatation of the right and left kidneys (RK, LK) as well as the clearly identifiable urin enlarged bladder neck (white arrow, c).

**Fig. 17:** source pedsradiology.com
Fig. 18: Axial (a) and sagittal (b) US images (- w.) show an uniloculated, anechogenic, well defined cyst (c) in the left hemiabdomen, located anterior to the kidney (K) and caudal to the spleen (S). Mesenteric cysts show a fine cyst wall, opposed to duplication cyst, which typically present a thick wall (L: Liver)
Conclusion

Detection of fetal pelvic cystic lesions at US screening exams is common. An accurate knowledge of the developing foetal anatomy is crucial to limit the differential diagnosis and to evaluate the risks. Because of its multiplanar capacity and its high tissue characterization, prenatal MRI may help to identify the exact anatomic location of the lesion and to establish the most possible diagnosis in a significant number of cases. This additional information may help the parents and the medical team to take the most appropriate therapeutic decisions during pregnancy and/or in the immediate postnatal treatment.

Personal information

References

1. Deborah Levine, Atlas of fetal MRI

