Complementary role of MRI in the study of fetal genitourinary system.

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Learning objectives

- To examine the ultrasound (US) and fetal magnetic resonance (MR) anatomy of the genitourinary system.
- To evaluate the utilities of the main techniques of imaging based on US and fetal MR.
- To describe the congenital anomalies in both techniques.
- To introduce our experience in US and fetal MR of the genitourinary system.

Background

The genitourinary pathology is relatively common, accounting for 30% - 50% of the overall structural anomalies at birth stage. This pathology may just require from simple monitoring in mild cases up to intrauterine derivation or immediately post birth surgery in severe cases.

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Given that fetal urine is the main source of amniotic fluid, the pathology at this level have a great influence in the fetal pulmonary maduration. The genitourinary anomalies include: urinary tract dilatations (the most frequent), development variants, renal cystic diseases and malignant pathology. These pathologies may be isolated entities or associated to systemic syndromes. Consequently, if an anomaly is detected, we should carry out a further evaluation in the remaining systems.

Findings and procedure details

ULTRASOUND OF THE FETAL GENITOURINARY SYSTEM

US is the initial image technique in the screening and it is a useful tool in the monitoring of genitourinary pathologies. Therefore most of these anomalies may be detected by this technique. At our institution, the first ultrasound screening is conducted by the obstetrics and gynecology unit. Only in case of a suspicious pathology, the patient may be transferred to an ultrasound specific center. This first US evaluation is carried out at 20-22 week of gestation, trying to answer 7 clue questions:

1) Are there two kidneys?
2) Do both kidneys have a normal size and echogenicity?

- Unilateral renal enlargement: multicystic dysplastic kidney, mesoblastic nephroma
- Increase of renal echogenicity: autosomal recessive polycystic renal disease, cystic renal dysplasia

3) Are there macroscopic renal cysts?

- Multicystic dysplastic kidney, cystic renal dysplasia, autosomal recessive polycystic renal disease.

4) Are the ureters visible?

- The normal ureters are not displayed. If they are dilated we have to regard distal obstruction, vesicoureteric reflux and primary megaureter.

5) Is the bladder visible?

- If the bladder is absent it usually is due to a failure in the urine production
- There are bladder anomalies of morphology or localization in certain systemic syndromes.

6) Does the bladder have a normal size?

- Marked distention of the bladder (megacystis): posterior urethral valves, urethral atresia, Prune - Belly syndrome.

7) Do adrenals glands have a normal morphology and position?

- The normal adrenal look is an "ice cream sandwich" appearance, with a hypoechoic cortex and a hyperecogenic medulla. Normal morphology is often an inverted-V or inverted-Y appearance.
- Flattened: renal agenesis or ectopia.
- Discoid morphology: adrenal enlargement in congenital adrenal hyperplasia (we have to look for the presence of female fetus virilization)
- Unilateral adrenal mass: neuroblastoma, adrenal hemorrhage, extralobar sequestration.

8) Are there normal genital?

**FETAL MR OF THE GENITOURINARY SYSTEM**

When ultrasound is not enough to evaluate the genitourinary system or MR can provide us valuable information that in other way would require the use of ionizing radiation, it is at that point when we use this technique. Next, the circumstances to evaluate the genitourinary system with fetal MR are described as follow:

**Fetal MR indications.**

- Anhydramnios, oligohydramnios.
- Maternal indication: obesity, suspected uterine rupture, placental anomalies, leiomyoma of the uterus, ovarian tumor or abscesses and/or pelvimetry measurements.
- To evaluate the fetal pulmonary maduration.
- When ultrasound may be unable to provide a definitive diagnosis

**Preparation.**

- Informed written consent from each women.
- To provide recent US.
- To identify the gestational week: fetal MR is done during the second or third trimester.
- Supine position during the second trimester and lateral position in advanced gestational stages.
- Optional sedation with the possibility to use alprazolam.
- Gadolinium is not administered.

**Technical considerations.**

- **Morphologic evaluation.**

Ultrafast T2-weighted sequences: they are essentials in the genitourinary system evaluation. At our institution we employ the next sequences:

- First-choice: Single shot fast spin echo.
- Others: turbo or fast spin echo T2 WI without mother breathe synchronization with 3D or MIP reconstructions and FIESTA T2 sequences.
Normal anatomy:

§ Renal parenchyma: Intermediate intensity signal, lower than the fluid and higher than the hepatic parenchyma (fig. 1).

§ Ureters and urethra: they only are visible if they are dilated.

§ Bladder: it is filled and emptied during the exploration. We have to avoid confusing the urinary jets with pathology (fig. 2).

§ Adrenal glands: low intensity signal, similar to liver in T2 weighted images.

 o T1 weighted sequences (SPGR sequences): they are useful to evaluate gastrointestinal system (hyperintense meconium fills the colon from 24 week) (fig. 3) and to differentiate the adrenal hemorrhage, which present high intensity signal, from the neuroblastoma, which has intermediate intensity signal.

- Functional evaluation.

-Echo-planar diffusion weighted images:

§ They are very sensitive detecting renal parenchyma, which usually shows high intensity signal and low signal on the ADC maps (fig. 4).

§ Preliminary results of recent reports point out to their usefulness in the renal function evaluation. The comparison with maternal kidneys may help us.

 o -Indirect signs of renal function:
   • Amniotic fluid volume
   • Pulmonary maduration degree
   • Bladder repletion degree

MAIN PATHOLOGICAL ENTITIES OF THE GENITOURINARY SYSTEM.

URINARY TRACT DILATATION

The urinary tract dilatation constitutes the highest percentage within the genitourinary anomalies (fig. 5). When we detect a dilated collecting system the most straightforward etiological considerations are:

- -Obstructive pathologies:
  • Ureteropelvic junction anomalies (35%)
- Ureterovesical junction anomalies (10%)
- Ureteroceles
- Bladder outlet obstructions: posterior urethral valves are the main entity.

- Vesicoureteric reflux.

Duplicated collecting system is another frequent entity that we should consider.

The first exploration must be always the ultrasound, the MR study being reserved in cases where the US is inconclusive or equivocal. (fig. 6).

An anteroposterior pelvis diameter greater than 3mm is considered pathological. There are several degrees depending on the measures and gestational age:

- Second trimester: mild = 4 - 7 mm, moderate = 8 - 10 mm and severe >10 mm.
- Third trimester: mild = 1 - 10 mm, moderate = 10 - 15 mm and severe >15 mm.

We should evaluate the possible existence of dysplasia, displayed as an increased echogenicity or a decrease intensity signal in diffusion weighted images.

- **Ureteropelvic junction obstruction**: hydronephrosis that ends abruptly at ureteropelvic junction, with a normal sized ureter and bladder.
- **Posterior urethral valves**: typically, the bladder (megacystis) and the posterior urethra should be distended, they provide us a "keyhole image" with a variable degree of ureterohydronephrosis and oligohydramnios.
- **Duplicated collecting system**: they follow the Weigert-Meyert law:
  - Upper pole ureter: it has an ectopic insertion and it is dilated due to obstruction, with and associated ureterocele in many cases (fig. 8)
  - Lower pole ureter: orthotopic insertion, with possible vesicoureteric reflux associated.

**RENEAL CYSTIC DISEASE.**

Firstly it is important to difference it from hydronephrosis:

- Peripheral localization: cysts.
- Central localization: collecting system dilatation.

**Obstructive cystic renal dysplasia**:

It is associate to an obstructive uropathy in which any obstructive cause leads to the renal cystic formation (fig. 9). The obstructive cystic renal dysplasia may present unilaterally or
bilaterally (resulting in oligohydramnios) or segmentary, in case of duplicated collecting system.

It is characterized by:

- Different sized renal cysts, primarily peripheral ("string of beads").
- Reniform normal morphology conservation.
- Echogenic parenchyma between the cyst, caused by dysplasia and microcysts.
- Differential diagnosis:
  - Multicystic dysplastic kidney: no ureter or bladder dilatation.
  - Hydronephrosis: dilated calyces connects with pelvis.

**Multicystic dysplastic kidney**

It is characterized by the replacement of renal parenchyma by cysts, most often is a unilateral condition (80%). It requires monitoring because it is usually accompanied by vesicoureteric reflux and contralateral ureteropelvic junction obstruction.

Main imaging features:

- Paraspinal mass occupying the flank with different sizes cysts, without connections between them, peripheral and central.
- Renal enlargement with loss of its shape, unable to identify renal parenchyma. (fig. 10, 11 y 12)

**Recessive polycystic renal disease**

It is a genetic disorder that affects the distal renal tubules (bilateral and symmetric cystic renal disease) and bile ducts (hepatic fibrosis).

It is characterized by:

- Medulla exclusive involvement.
- Bilateral involvement with marked renal enlargement and hyperechogenic parenchyma with loss of corticomedullary differentiation and intact cortex (hypoechoic / hypointense ring)

**NORMAL DEVELOPMENT VARIANTS**
They obey to a renal tissue absence, ectopic localization and fusion and ascent abnormalities.

Differential diagnosis:

- When we detect an empty flank, we have to evaluate the contralateral kidney looking for a probably compensatory hypertrophy.
  - Hypertrophy is present: renal agenesis is suspected (fig. 13).
  - No hypertrophy: it may be a pelvic kidney (fig. 14).

- When there is an abnormal renal morphology, we must suspect a horseshoe kidney. In this variant exists parenchymal or fibrous connection between both kidneys (isthmus), located previous to aorta, more often connecting the lower poles.

- In case of an empty flank and an atypical enlarged contralateral kidney, we must think about a cross fused renal ectopia. To study this kind of anomalies is preferable use fetal MR, especially diffusion sequences.

**MESOBLASTIC NEPHROMA**

If we found a retroperitoneal mass:

- Renal origin: mesoblastic nephroma is the first abnormality to consider.
- Adrenal origin: neuroblastoma, adrenal hemorrhage, extralobar sequestration.

Mesoblastic nephroma is the most common solid kidney neoplasm in fetuses and neonates. It is a benign entity with an excellent prognosis.

It is characterized by:

- Large sized renal mass, solid, well circumscribed and hipervascular. It associates polyhydramnios frequently (70%).
- Iso or hyperechogenic relative to normal renal parenchyma with a vascular hypoehogenic ring in Doppler ("ring sign").
- MR is used to confirm location and relationships. Mesoblastic nephroma presents as homogeneous mass with a moderate intensity signal on T2 weighted images.

The image is not useful to differentiate from Wilms tumor. Age plays an important role, because the Wilms tumor is exceptional in fetus.

**NEUROBLASTOMA**
It is the most common malignant neoplasm in neonatal age. It originates from adrenal medulla (90%) or from the sympathetic neural tissue.

Warning signs:

- Unidentifiable adrenal.
- Inferiorly displaced kidney.

Its main features are:

- Composition:
  - Complex cysts with septation (50%).
  - Homogeneous solid, echogenic and moderate intensity signal on T2 WI.
  - Mixed

- Metastases: most often in liver, we must suspect them by the existence of hepatomegaly or hydrops fetalis.

- In Doppler-colour it shows diffuse vascularization.

- Clinical: variable. Some cases may experience spontaneous regression, up to 40% (fig 15) or differentiate into benign ganglioneuromas.

- Differential diagnosis:
  - Extralobar sequestration: the most identifying feature is the existence of a single irrigation artery arising from aorta. In these cases adrenal gland is recognizable.
  - Adrenal hemorrhage: it is hyperintense on T1 WI, regresses and shows no flow.
  - Duplicated collecting system: dysplastic and dilated upper renal pole can resemble an adrenal cystic injury.
  - Retroperitoneal teratoma: calcifications helps in their identification.
  - Mesoblastic nephroma.

**ADRENAL HAEMORRHAGE.**

It is displayed as a clearly defined lesion with hyperintense signal on T1 and T2 weighted sequences.

**ADRENAL CYST.**
We must suspect this entity when we detect a well-defined mass that shows hyperintense on T2 WI and hypointense on T1 WI (unlike the adrenal hemorrhage) in the theoretical location of the adrenal gland; no clearly identify some or all of it. (Fig 16 y 17).

**FETAL BLADDER ANOMALIES.**

**Absent bladder.**

It may be obey to an inadequate filling due to a renal disease or severe obstruction, or to an altered bladder structure (bladder or cloacal exstrophy).

**Megacystis.**

When we detect this finding we must first look for an obstruction in the bladder outlet. There are other entities associated to megacystis, like Prune - Belly syndrome (fig. 18), megacystis - microcolon syndrome and cloacal malformations.

**OUR EXPERIENCE**

Our hospital is reference for fetal medicine in eastern Andalucía. Since 2006 we have collected a total of 413 fetal MR. The 82% were brain studies and the 18% of the rest of the body, being the most frequent suspected pathologies diaphragmatic hernias and gastrointestinal abnormalities. Genitourinary system MR account for 4%, that were a total of 18 cases performed in third trimester (fig. 19).

Below, the disease distribution is reflected. N = 18 cases.

- 6 bilateral urinary tract dilatation (fig. 5 y 6)
- 5 multicystic dysplastic kidney cases (fig. 10, 11 and 12)
- 1 duplicated collecting system with multicystic dysplastic kidney in upper moiety and ureterocele (fig. 8)
- 1 left renal agenesis (fig. 13)
- 2 neuroblastomas (fig. 16 - 18)
- 2 adrenal cysts (fig. 19 and 20)

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**Images for this section:**
**Fig. 1:** Normal renal parenchyma in a healthy fetus. Axial (A) and coronal (B) T2-weighted SSFSE sequences. Normal kidneys show intermediate intensity signal (arrows).

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Fig. 2: Normal bladder in a healthy fetus. Coronal (A) and (B) sagittal T2-weighted SSFSE sequences. The normal bladder filled by urine show T2 hyperintensity (white arrows). On coronal T2-weighted sequence (A) also appears others two high signal structures corresponding to the stomach (open white arrow) and gallbladder (black arrow).

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Fig. 3: Normal gastrointestinal tract in a healthy fetus. Coronal (A and B) T1-weighted SPGR sequences demonstrate normal colon entirely filled by high signal intensity meconium (white arrows), in this case, it also can be visualized within distal small intestine (black arrow).

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**Fig. 4**: Axial echoplanar diffusion-weighted sequences showing bright signal in normal renal parenchyma (arrows).

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Fig. 5: Bilateral hydronephrosis. Coronal (A) and sagital (B) T2-weighted SSFSE sequences and coronal diffusion-weighted sequences (C). The kidneys are enlarged with a dilated collecting system (*) and ureters (white arrows). There is also thinning of renal parenchyma (A and B). Both kidneys show restricted diffusion suggesting normal renal function (C).

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**Fig. 6:** Fetal US shows a duplex collecting system with dilatation of the renal pelvis and calices in both poles of the fetal kidney. Fetal ultrasound was enough to reach a certain diagnosis.

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Fig. 7: Fetal US demonstrate urethral atresia secondary to posterior urethral valves. The bladder and posterior urethra are dilated showing the typical keyhole sign.

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**Fig. 8:** Segmental multicystic dysplastic in the upper moiety of the right kidney with a duplex collecting system. Axial (A), sagittal (B) and coronal (C) T2-weighted SSFSE sequences in a fetus of GW 31 + 0. The upper moiety of the right kidney is dilated (white arrows), the lower moiety has a normal parenchyma (open white arrow) and intravesical ureterocele (black arrow).

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**Fig. 9:** Obstructive cystic dysplasia of left kidney. Coronal (A) and axial (B) T2-weighted SSFSE sequences (arrows).

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**Fig. 10:** Multicystic dysplastic right kidney. Sagital (A) and coronal (B) T2-weighted SSFSE sequences and coronal T1-weighted SPGR sequences (C). Multiple cysts not communicating with each other (white arrows) and no evidence of recognizable right renal parenchyma. The colon is filled by meconium without associated gastrointestinal anomalies (open white arrow).

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Fig. 11: Ultrasound equivalence in the same patient of figure 8. Multiple internal cysts of varying sizes, not communicating with each other.

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Fig. 12: Multicystic dysplastic left kidney. Axial (A) and coronal (B) T2-weighted SSFSE sequences. Left renal parenchyma can be visualized enlarged and completely replaced by cysts of varying sizes (arrows).

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**Fig. 13:** Left renal agenesis. Left sagittal (A), right sagittal (B) and axial (C) T2-weighted SSFSE sequences. Left empty renal pouch.

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**Fig. 14:** Multicystic dysplastic left pelvic kidney. Coronal (A), axial (B) and sagittal (C) T2-weighted SSFSE sequences. No renal parenchyma is identified in the renal fossa. A multicystic formation is observed in the pelvis, in contact with the posterior wall of the bladder (*) corresponding to multicystic dysplastic kidney (white arrows).

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**Fig. 15:** Left neuroblastoma. Coronal T2-weighted SSFSE sequence (A), coronal T1-weighted SPGR sequence (B) and fetal US performed at 2 years (C). A septate cystic lesion is identified in the theoretical location of the left adrenal gland (arrows). This lesion regressed progressively until disappearing completely as fetal US showed at 2 years.

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**Fig. 16:** Left adrenal cyst. Coronal (A), axial (B) T2-weighted FIESTA sequences and coronal (C) T1-weighted SPGR sequence in a fetus of GW 28 + 0. A retrogastric cystic lesion with benign characteristics is observed (*). Normal right adrenal gland and part of the left adrenal gland is identified (white arrows). The stomach is displaced and it has normal characteristics (S).
Fig. 17: Ultrasound equivalence showing the adrenal cyst in the same patient of figure 19 (arrow).
**Fig. 18:** Prune belly syndrome. Fetal US showing a megacystis thin-walled with anterior abdominal wall underdevelopment.

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**Fig. 19:** Graph showing the percentage distribution of fetal MRI studies performed in our center since 2006.
Conclusion

1. Multidisciplinary collaboration is essential for maximum diagnostic yield in the congenital pathology of the genitourinary tract.
2. Ultrasound is the primary imaging modality in the management of the genitourinary abnormalities.
3. Fetal MR should be considered as a useful complementary tool in the evaluation of these anomalies during the third trimester of pregnancy. Its role in the assessment of fetal adrenal and genitourinary system is not fully defined and there are few publications, most of them with small cases series.
4. SS-FSE T2-weighted sequences are essential to evaluate the morphology of the genitourinary system, as they provide an accurate visualization of the renal pelvis, dilated urinary tract and cystic renal lesions.
5. Fat-suppressed SPGR T1-weighted sequences allow the assessment of the gastrointestinal tract and differentiate between tumors and haemorrhages.
6. Echoplanar diffusion-weighted images are excellent in detecting renal parenchyma and are also promising since they allow the evaluation of renal function and not only morphology.

Personal information

References


