Abdominal and gastrointestinal tract congenital anomalies. A fetal MR study

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Purpose

- To show the normal anatomy of the fetal abdomen and digestive tract by magnetic resonance imaging (MRI).
- To describe the findings of the fetal abdominal and gastrointestinal tract anomalies.
- To evaluate the usefulness of MRI in the diagnosis and determine if it provides additional data to the ultrasound findings.
Methods and Materials

Introduction

Prenatal diagnosis consists of acquiring genetic, anatomic, biochemical, and physiological information to determine the likelihood of fetal alterations that might have repercussions during gestation and/or after birth. It is essential to provide families with this information and genetic counseling to enable decisions regarding the therapeutic alternatives for any anomalies detected before birth.

Until recently, ultrasound (US) examination was the only non-invasive method for the evaluation of fetal anatomy. However, with the development of ultrafast magnetic resonance (MR) sequences this technique is proving to be an invaluable adjunct to US, providing additional data that can have an important impact on the final diagnosis.

Congenital abdominal anomalies can occur in many fetal organs (the kidneys, ureters, bladder, genitals, gastrointestinal tract, liver, spleen, mesentery, and/or peritoneal cavity). The accurate determination of the location and morphological characteristics of an anomaly is essential for diagnosis. MR's ability to discriminate among tissues makes it a great help in the diagnosis of these pathologies.

As some of these anomalies can be difficult to detect clinically at birth, prenatal diagnosis is important to ensure early intervention and minimize complications.

Our objective is to illustrate the MR findings for anomalies of the abdomen and gastrointestinal tract that do not involve the genitourinary system.

Methods and materials

Our gynecology and obstetrics service is a referral center for a population of approximately 400,000 inhabitants and receives patients with suspected or confirmed fetal anomalies from other centers. Approximately 3,500 second-trimester US examinations are performed here annually. Our prenatal diagnostic committee, made up of obstetricians, neonatologists, pediatricians, geneticists, biochemists, psychologists, pediatric surgeons, pathologists, and radiologists, has been in operation since the early 1990s. This committee analyzes the anomalies detected or suspected and proposes the most appropriate methods for further study in each case. Approximately 1% of pregnancies are studied with MR.

Since 1997, 704 pregnant women carrying a total of 733 fetuses have undergone MR at our center. In 138 fetuses an abdominal or gastrointestinal tract anomaly was confirmed with clinical or radiologic evidence after birth or with the results of the autopsy in cases where legal interruption of pregnancy was performed.
Since current legislation in our environment allows the interruption of pregnancy for therapeutic purposes until the 22th week of pregnancy, most MR studies have been performed between the 20\textsuperscript{th} and 22\textsuperscript{nd} weeks of gestation.

**MR technique**

No special preparations were necessary for any of the patients or fetuses. All MR studies were performed using a 1.0 or 1.5 Tesla. A multiple-element phased-array body coil was used to obtain T2-weighted images (WI) using the SS-HF-RARE or HASTE sequences; occasionally, single volumetric images were obtained using SS-RARE sequences. T1-WI were obtained using ultrafast gradient-echo sequences. The average time employed for each examination was approximately 20 minutes.

**Normal anatomy of the abdomen and gastrointestinal tract**

**Amniotic fluid**

Given that some gastrointestinal tract anomalies associate polyhydramnios, assessment of amniotic fluid volume provides useful information. MRI can produce volumetric images with SS-RARE sequences where a subjective assessment of the amniotic fluid volume can be made (Fig. 1), as well as general information of the fetal morphology is obtained.

**Gastrointestinal tract (1)**

**Oropharyngeal cavity**

The oropharyngeal cavity is usually well-depicted on MR. The axial plane is best for evaluating the upper maxillary. The palate, tongue, and pharynx are best visualized in the sagittal plane, although all three spatial planes need to be studied for thorough evaluation of this region (Fig. 2).

**Gastrointestinal tract**

The esophagus is not normally visible unless it is dilated or image acquisition happens to coincide with fetal swallowing.

Visualization of the diaphragm is excellent in both the coronal and sagittal planes.

The stomach should be seen from early on as a hyperintense cavity in T2-WI; the jejunum and the ileum are identified on T2-WI as hyperintense serpiginous structures distributed throughout the abdomen but mainly occupying the left hemiabdomen; the colon is hypointense on T2-WI and hyperintense on T1-WI due to the presence of meconium. These structures need to be studied in the axial, coronal, and sagittal planes of the fetus. The rectal ampulla is best evaluated in the sagittal plane using T1 sequences (Fig. 3).
Liver, spleen, pancreas, peritoneal cavity and abdominal wall

The liver is well-depicted in all planes. It is fairly hypointense in T2-WI and of intermediate signal intensity on T1-WI. The fetal liver is located in the right side of the abdomen and it tends to be somewhat larger in proportion to body size than the liver after birth. The gallbladder is well visualized and it is hyperintense on T2-WI. The spleen is more difficult to evaluate; its signal intensity in T2 sequences is somewhat higher than that of the liver. The pancreas is not usually seen, possibly due to its small size. The peritoneal cavity is a virtual cavity and is not visualized unless there is ascites. The abdominal wall is easily recognized in all spatial planes on T2-WI. It is also easy to recognize the umbilical cord and its point of insertion (Fig. 4).

Anomalies of the abdomen and gastrointestinal tract

We have considered that facial and oropharyngeal cavity anomalies are part of the digestive tract as they play an important role in swallowing so are included under this heading.

Oropharyngeal cavity

Cleft lip and cleft palate

Alteration of the fusion of the fronto-nasal and maxillary processes and the primary and secondary palatine processes. Cleft lip and cleft palate can occur separately or together. The etiology of the two anomalies are distinct (the lips fuse at 7-8 weeks’ gestation and the palate fuse at 12 weeks).

The overall incidence is 0.15 % of all births, which represents 7.5 % of all fetal anomalies. Predisposing factors include rubella, medication, alcohol, tobacco, and drug abuse. These anomalies are sometimes difficult to diagnose and this is especially true of cleft palate.

Cleft lip and cleft palate are often associated to other anomalies: chromosomopathies, particularly 13 and 18, syndromic presentations, CNS anomalies, skeletal anomalies, and cardiovascular anomalies.

If one parent has this anomaly, the rate of recurrence is 3-5 %. If, moreover, a sibling has this anomaly, the rate of recurrence is 14-17%. If it is part of an autosomal dominant syndrome the recurrence is 50% (Fig. 5, Fig. 6, Fig. 7 and Fig. 8) (2, 3).

RM  Sequences  Projections
Defect in lip and/or palate  T2  All (*)
Small stomach (**)
Polyhydramnios (***)

(*) It is absolutely essential to evaluate the three standard projections to reach the correct diagnosis. Assessment can be very difficult

(**), (***) Not always present

Micrognathia

Micrognathia is present in numerous syndromes (Treacher Collins syndrome, Goldenhar syndrome, Roberts syndrome, Pierr-Robin syndrome, and others) and chromosomopathies (Trisomy 10, 18, 9). The alteration in the anatomy of the mandible can impair fetal deglution.

The prognosis depends on the syndrome or chromosomopathy and associated anomalies (Fig. 9).

<table>
<thead>
<tr>
<th>RM Sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Shortening of the mandible</td>
<td>T2 Midline sagittal</td>
</tr>
<tr>
<td>Small stomach (*)</td>
<td></td>
</tr>
<tr>
<td>Polyhydramnios (**)</td>
<td></td>
</tr>
</tbody>
</table>

(**), (**) Not always present

Macroglossia

This is a rare entity. In Beckwith-Wiedemann syndrome, the tongue tends to be greatly enlarged. We examined a fetus with macroglossia but were unable to detect this anomaly. Trisomy 21 is usually associated to variable degrees of macroglossia. Fetal swallowing can be impaired and there may be polyhydramnios.

The size of the tongue can be difficult to evaluate with MR; the tongue is hypointense in T2-weighted sequences.

Tumors and masses

These are uncommon, the most common being:

- **Teratoma (Epignathus).** It has an incidence of 1 in 35,000 live births. Approximately 6% are associated to other anomalies: facial clefts, bronchial cysts, hypertelorism, and heart defects.
• **Epulis (Giant-cell granuloma).** An uncommon benign pedunculated tumor, arising anteriorly from the maxillary alveolar ridge. There are cases published about a triple X female fetus (Fig. 10) (4).

• **Hemangioma - cystic lymphangioma.** The most common tumors in infants and most are present at birth. The most common location for lymphangioma is in the neck and they usually extend toward the thorax (Fig. 11).

• **Goiter.** Enlargement of the thyroid can result from either hypo- or hyperthyroidism. The incidence of congenital hypothyroidism is 1 in 3,700 live births. Altered thyroid function in fetuses with maternal Graves disease is 2-12%.

These masses can impair fetal deglution and cause polyhydramnios. After birth they can impair swallowing and breathing.

<table>
<thead>
<tr>
<th>RM</th>
<th>Sequence</th>
<th>Projections (*)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Teratoma</td>
<td>Complex mass</td>
<td>T2</td>
</tr>
<tr>
<td></td>
<td>Polyhydramnios</td>
<td></td>
</tr>
<tr>
<td>Épulis</td>
<td>Hypointense mass</td>
<td>T2</td>
</tr>
<tr>
<td></td>
<td>Polyhydramnios</td>
<td></td>
</tr>
<tr>
<td>Hemang. / lynphang.</td>
<td>Cystic mass</td>
<td>T2</td>
</tr>
<tr>
<td></td>
<td>Polyhydramnios</td>
<td></td>
</tr>
<tr>
<td>Goiter</td>
<td>Hypointense mass</td>
<td>T2 and T1</td>
</tr>
<tr>
<td></td>
<td>at T2 and hyperintense at T1</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Poly / Oligohydramnios</td>
<td></td>
</tr>
</tbody>
</table>

(*) All spatial projections are necessary to achieve a correct diagnosis. We point the projection that we think gives more information in each case.

**Proximal gastrointestinal disorders**

**Esophageal atresia**

This results from incomplete division of the foregut into the ventral respiratory portion and the dorsal digestive portion by the tracheoesophageal septum. It has an incidence of 2 to 12 cases in 10,000 live births.

Atresia with distal tracheoesophageal fistula is the most common presentation (85-90% of cases) (Fig. 12). The remaining entities are atresia without fistula (10%) (Fig. 13),
tracheoesophageal fistula in H (fistula without atresia), atresia with proximal fistula and atresia with double fistula proximal and distal.

Up to 50-70 % of cases are associated to other anomalies (gastrointestinal 28%, cardiopathy 24%, genitourinary 13%, musculoskeletal 11%, CNS 7% and others). 3-4% of cases are associated to chromosomopathies, especially Down's syndrome.

There is no increased risk of recurrence (7,8).

<table>
<thead>
<tr>
<th>RM</th>
<th>Sequence</th>
<th>Projection</th>
</tr>
</thead>
<tbody>
<tr>
<td>Small stomach (*)</td>
<td>T2</td>
<td>Midline sagittal</td>
</tr>
<tr>
<td>Polyhydramnios</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Proximal esophageal pouch (**)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

(*) The small stomach can be a normal finding or be due to other causes (deglution disorders, facial defects, tumors, CNS lesions, oligohydramnios...)

(**) Not always present

Hiatal hernia

Very rare in the newborn. May be associated with Marfan syndrome, asplenia and polysplenia. We must make the differential diagnosis with the diaphragmatic hernia, esophageal duplication cyst and bronchogenic cyst. Recurrence is very low.

With MRI you can see the distal esophagus and the stomach herniated dilated as hyperintense structures on T2-weighted sequences.

We have not studied any fetus with this anomaly.

Duodenal obstruction

Obstruction of the duodenum can be caused by atresia (Fig. 14), stenosis, intraluminary diaphragm, annular pancreas, intestinal malrotation with Ladd's band (Fig. 15), or midgut volvulus.

The estimated incidence of these anomalies is 1 / 5,000 pregnancies. Vascular impairment during gut development can cause duodenal stenosis and atresia. Duodenal atresia and annular pancreas are associated in most cases.

Approximately 50% of duodenal anomalies are associated to other anomalies (gastrointestinal, skeletal, cardiovascular, genitourinary, and chromosomal).
The associated gastrointestinal anomalies can include atresia at any other point of the digestive tract, biliary atresia, pancreatic ductal atresia. Trisomy 21 is present in 30-40% of cases of duodenal atresia.

The differential diagnosis includes choledochal cyst, hepatic cyst, duplication cyst (9,10).

<table>
<thead>
<tr>
<th>RM</th>
<th>Sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Enlarged stomach (*)</td>
<td>T2</td>
<td>All</td>
</tr>
</tbody>
</table>

Dilatation of the duodenum

Polyhydramnios

(*) The stomach may be normal or small if duodenal obstruction is associated to esophageal atresia.

Small bowel abnormalities

Atresia/ stenosis

Atresia of the small intestine affects approximately 1 in every 3,000-5,000 live births.

Vascular impairment during development is the most likely cause of this anomaly. Atresia most often affects the distal ileum (36%) or the proximal jejunum (31%) (Fig. 16), and is multiple in approximately 6% of cases.

While associated extraintestinal anomalies are rare, other intestinal anomalies (other atresias, malrotation, volvulus, duplication, gastroschisis, meconium ileus) are not. Meconium ileus can be a complication of atresia.

A large proportion of cases are affected by cystic fibrosis. It is normally very difficult to differentiate between this entity and meconium ileus, volvulus, and Hirschsprung's disease. It can sometimes simulate abdominal cysts.

<table>
<thead>
<tr>
<th>RM</th>
<th>Sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dilatation of loops proximal to the obstruction</td>
<td>T2 and T1 (**)</td>
<td>All</td>
</tr>
<tr>
<td>Microcolon</td>
<td>T2 and T1 (**)</td>
<td>All</td>
</tr>
<tr>
<td>Polyhydramnios (*)</td>
<td>T2 and T1 (**)</td>
<td>All</td>
</tr>
</tbody>
</table>

(*) Likelihood increases when atresia is more proximal.
In T2-weighted sequences the contents of the intestine proximal to the atresia is less intense than usual. The signal intensity is increased in T1-weighted sequences, which can help to differentiate these intestinal loops from dilated ureters.

Meconium ileus

Meconium ileus is the result of functional obstruction of the distal ileum by abnormally thick meconium. It is the earliest sign of cystic fibrosis; almost all newborns with meconium ileus have cystic fibrosis and 10-15% of all patients with cystic fibrosis have meconium ileus.

Cystic fibrosis is the most common autosomal recessive disease and is found in 1 of every 3,000 newborns. Meconium ileus is often (50%) associated with gastrointestinal complications such as volvulus, jejunoileal atresia, intestinal perforation, and meconium peritonitis.

The differential diagnosis should include jejunoileal atresia, volvulus, and Hirschprung's disease. The risk of recurrence when a sibling has cystic fibrosis is 25%. We have not encountered this anomaly.

<table>
<thead>
<tr>
<th>RM Sequences Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dilatation of small bowel loops T2 and T1 All</td>
</tr>
</tbody>
</table>

Altered signal intensity (*)

Microcolon

Ascites (**) 

Polyhydramnios +/-

(*) Owing to the high protein content, increased mucus, and decreased water in the meconium in this entity, signal intensity on T2-WI is lower than usual and higher than usual on T1-WI.

(**) Whenever there is intestinal perforation as a complication

Meconium peritonitis

Caused by intestinal perforation during gestation. Meconium peritonitis causes chemical peritonitis and the formation of fibrous tissue masses that calcify (Fig. 17).

It occurs in approximately 1 of every 2,000 pregnancies.
In most cases (65%), it is the consequence of meconium ileus, intestinal atresia, or volvulus (13,14,15).

| RM Sequences Projections |
|--------------------------|------------------|
| Dilatation of bowel loops T2 All |

Ascites

Peritoneal calicifications (*)

(∗) Difficult to see, if seen they are hypointense in T2-weighted sequences.

Hyperechoic bowel // "Altered bowel signal intensity"

Hyperechoic bowel is an echographic term that does not refer to a pathologic entity; rather it expresses a subjective impression of the behavior of bowel contents.

It is a controversial term as potential associations are with pathologic entities such as aneuploidy, intestinal obstruction, congenital infection, fetal death, cystic fibrosis, and intrauterine growth retardation.

It is seen in approximately 0.5% of second-trimester ultrasound examinations, and it may be caused by alterations in the components of meconium (similar to that described above for meconium ileus), blood swallowed by the fetus, intestinal wall edema, or ischemia.

MR can evaluate this condition, possibly also subjectively, showing lower signal intensity for the bowel in T2-weighted sequences and higher signal intensity in T1-weighted sequences (Fig. 18) (16).

Large bowel abnormalities

Large bowel atresia and anorectal atresia

Large bowel atresia is uncommon (less than 10% of all intestinal atresias). Vascular impairment seems to be the most probable etiology. Imaging findings for this entity are not well-defined, although dilated intestinal loops above the atresia and altered signal intensity of bowel contents may be seen in both T1 and T2-weighted MR sequences. We have not encountered this anomaly.

Anal atresia is more common; its incidence is estimated at 1 in 5,000 live births.

In up to 70% of cases other anomalies (especially of the genitourinary tract) are associated. Other associated anomalies include skeletal (especially vertebral),
gastrointestinal (especially tracheoesophageal atresia), cardiovascular, CNS, and chromosomopathies (especially trisomies 21 and 28).

It should be differentiated from other causes of intestinal dilatation, large bowel atresia, and Hirschsprung’s disease.

We studied two fetuses with anorectal atresia using MR but their gestational age (14 and 16 weeks) did not allow us to reach the diagnosis.

<table>
<thead>
<tr>
<th>RM Sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Dilatation of the colon</td>
<td>T1 (**)</td>
</tr>
</tbody>
</table>

Amniotic fluid normal (*)

(*) As long as it is not associated to a high bowel atresia (polyhydramnios) or to bilateral kidney malformation (oligohydramnios).

(**) T1-weighted sequences depict the colon best: in these sequences meconium is hyperintense, which makes it possible to differentiate it from other dilated structures (especially ureters)

Hirschsprung’s disease

The estimated incidence is 1 in 10,000-20,000 live births. The rectum is always affected, while the portion of the colon affected varies. Small bowel affection is rare (8% of cases). Prenatal diagnosis is difficult and the differential diagnosis is with other causes of intestinal obstruction. Association with Down’s syndrome has been described.

We suppose that MR would enable us to see dilatation of the segments of the colon proximal to the aganglionic region and altered signal intensity of the meconium.

Hirschsprung’s disease is usually sporadic; however, 4% to 6% of cases are familial. We have not encountered this anomaly.

Other intestinal anomalies

Heterotaxy syndromes

The terminology around this syndrome has been confusing; it includes terms such as situs solitus, situs ambiguous, situs inversus (Fig. 19), asplenia/polysplenia syndrome (Fig. 20), and cardiiosplenic syndrome.

These terms describe the abnormal arrangement of the organs in the thorax and abdomen. Findings must be evaluated on a case-by-case basis to allow patients to be classified into a particular subtype of this anomaly. The overall incidence is 1 of
every 10,000 adults. There is a very high prevalence of congenital heart defects in this population (18,19)

Abdominal anomalies are very common and form part of this syndrome (polysplenia, asplenia, central or left-sided liver, small stomach, central or right-sided stomach, esophageal atresia, duodenal atresia, biliary atresia, intestinal malrotation, anomalous morphology of the bronchi, as well as genitourinary, skeletal, and CNS anomalies). It is difficult to establish the recurrence rate as many factors are involved in the etiology of heterotaxy syndrome. Heart defects, so often associated to this syndrome, cannot usually be diagnosed with MR.

<table>
<thead>
<tr>
<th>RM</th>
<th>Sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Abnormal position of organs in the tórax and abdomen (*)</td>
<td>T1 / T2</td>
<td>All</td>
</tr>
</tbody>
</table>

(*) The position of the stomach is often the clearest, most easily evaluated sign.

**Abdominal masses** (20,21)

**Enteric duplication cyst**

Duplications of the digestive tube are uncommon congenital abnormalities found anywhere along the alimentary tract from the tongue to the anus.

Intestinal duplication is located on the mesenteric side of the gut and does not usually communicate with intestinal lumen. The most common location is the distal portion of the ileum (Fig. 21), followed by the distal portion of the esophagus and the stomach. Although these lesions rarely cause intestinal occlusion in utero, they can cause intestinal occlusion and/or abdominal pain after birth, due to volvulus, invagination, or bleeding of the cyst. Sometimes other anomalies such as bronchopulmonary sequestration (Fig. 22) or vertebral anomalies are associated, especially in esophageal duplications.

They should be differentiated from other abdominal cysts (ovarian cyst, mesenteric cyst, choledochal cyst) and dilatation of bowel loops or the urinary system (22).

<table>
<thead>
<tr>
<th>RM</th>
<th>Sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Generally rounded mass</td>
<td>T2 / T1 (*)</td>
<td>All</td>
</tr>
</tbody>
</table>

(*) Hyperintense structure on T2-WI and hypointense on T1-WI. The combined use of these sequences can help to differentiate this entity from dilated bowel loops.

**Mesenteric cyst**
It is difficult to determine their incidence because many of these cysts are asymptomatic. They are considered to be lymphatic anomalies. They are often single, unilocular lesions (Fig. 23), although they can be multilocular. They are usually located in the mid-abdomen within the mesentery, omentum, or retroperitoneum and sometimes are mobile and change position.

Mesenteric cysts are not usually associated to other anomalies.

As in the case of duplication cysts, they need to be differentiated from other cystic anomalies of the abdomen.

<table>
<thead>
<tr>
<th>RM</th>
<th>Sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Uni or polylobulated mass</td>
<td>T2 (*)</td>
<td>All</td>
</tr>
</tbody>
</table>

(*) They are usually hyperintense on T2-WI unless complicated with hemorrhage.

**Ovarian cyst**

This is a common cause of abdominal cysts in females during the prenatal period; it occurs in approximately 1 in 2500 pregnancies. It usually presents during the third trimester (Fig. 24).

Excessive stimulation of the fetal ovaries by placental and maternal hormones seems to play a role in their etiology. Most are unilateral, although they can be bilateral. In fetuses they can be simple cysts or they can complicate with torsion and bleeding. They sometimes disappear in utero, though the majority disappear after birth.

They need to be differentiated from other abdominal cysts, dilation of bowel loops, and dilations of the ureters. Ovarian cysts are not normally associated to other fetal anomalies.

<table>
<thead>
<tr>
<th>RM</th>
<th>Sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hyperintense, rounded structure in the female fetal abdomen (*)</td>
<td>T2</td>
<td>All</td>
</tr>
</tbody>
</table>

(*) In the case of simple cysts. When complicated by torsion and/or bleeding, they can be have septa and contents with different signal intensity.

**Hepatic and splenic masses**
These lesions are very rare in the prenatal period. Cysts are the most common. They may appear within liver parenchyma or "hang" from its lower edge. They can be single or multiple. Multiple lesions usually form part of autosomal recessive polycystic kidney disease. Single lesions usually correspond to a ciliated foregut cyst or epidermoid cyst. Masses in the spleen are almost always epidermoid cysts.

They should be differentiated from duplication cysts, mesenteric cysts, urachal cysts, choledochal cysts, and ovarian cysts.

Single cysts are not usually associated to other anomalies; however, we examined a fetus with a hepatic cyst and severe renal and cardiac anomalies (Fig. 25).

<table>
<thead>
<tr>
<th>RM sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Uni or polylobulated mass</td>
<td>T2 (*)</td>
</tr>
</tbody>
</table>

(*) In T2-weighted sequences they are hyperintense unless they have bled, in which case they have different signal intensities in their interior.

Other masses are: hepatosplenomegaly (caused by fetal hydrops, congenital infection, Beckwith-Wiedemann syndrome or Zellweger syndrome), hemangioma, hemangioendothelioma (Fig. 26), hamartoma and metastasis.

Other cysts and masses

**Neuroblastoma** (Fig. 27) is the most common solid neoplasm in infants. It affects 1 of every 7,000-10,000 live births. It usually arises in the adrenal gland. It is almost always detected in the third trimester of gestation. **MR** can show solid, cystic, or more complex lesions. These tumors can metastasize, especially to the fetal liver and the placenta. They can disappear before birth.

**Infradiaphragmatic extralobar pulmonary sequestration** (Fig. 28). 8-10% of cases of extralobar pulmonary sequestration are found in the abdomen. They are often associated to type 2 congenital cystic adenomatoid malformation. More than 90% are located on the left side. On **MR** they are seen as well-defined masses between the diaphragm and left kidney. Unlike neuroblastoma, they may appear in the second trimester.

Other cysts and masses, such as suprarenal hemorrhage (MR can be an excellent technique for its diagnosis) and teratoma can affect the fetus.

**Ascites**

Fetal ascites can be an isolated finding or it can be associated to hydrops fetalis. Isolated ascites is normally secondary to an intraabdominal problem rather than a systemic
anomaly and it is often associated to fetal malformations. Genitourinary problems, usually from obstructive uropathy, are the most common cause. Other causes of isolated ascites are gastrointestinal in origin, usually meconium peritonitis, hepatic anomalies, heart defects (Fig. 29), infections, metabolic disorders, and it is sometimes idiopathic (Fig. 30).

Hydrops fetalis is defined as an excess of total body water and this term is employed when fluid is detected inside at least two body cavities or in a single cavity when anasarca is present. Hydrops fetalis can have a wide variety of causes, which can be divided into two major groups: immunologic, related to incompatibility between fetal and maternal red blood cells; and non-immunologic. At present the majority of cases are non-immunologic and the prevalence is approximately 1 in 3,000 live births.

<table>
<thead>
<tr>
<th>RM</th>
<th>Sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Free fluid in the peritoneal cavity (*)</td>
<td>T2</td>
<td>All (**)</td>
</tr>
</tbody>
</table>

(*) If part of hydrops fetalis, there will be free fluid in other fetal cavities.

(**) When part of hydrops fetalis, it is necessary to find the cause of the hydrops fetalis.

**Ventral wall malformations**

**Gastroschisis** (24)

This is the herniation of the abdominal viscera of the fetus into the amniotic cavity, secondary to a small defect affecting all of the layers of the abdominal wall (Fig. 31). It affects approximately 1 in 3,000 live births.

There is a strong association with maternal age, being more common in young mothers, and also with drug abuse. Associated anomalies are infrequent. Intestinal atresia or stenosis can be found in 7-30% of cases.

The risk of recurrence has been estimated at 3.5%, although it is generally sporadic.

<table>
<thead>
<tr>
<th>RM</th>
<th>Sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bowel loops outside the abdominal cavity</td>
<td>T2, volumetric SS-RARE</td>
<td>Axial, sagittal</td>
</tr>
</tbody>
</table>

| Normal position of insertion of the umbilical cord (*) | | |

--

Page 16 of 62
These findings differentiate gastroschisis from omphalocele.

**Omphalocele**

This is a central defect that affects the navel; the hernia is wrapped in a membrane composed of two layers: the peritoneum and the amnion (Fig. 32). The incidence is approximately 1 in 4,000 live births.

Other associated anomalies are present in approximately 54% of cases, the most common being heart defects, followed by CNS anomalies. Neural tube defects and genitourinary and gastrointestinal malformations. It is associated to trisomies 18 and 13. Beckwith-Wiedeman syndrome is often associated to omphalocele (25).

<table>
<thead>
<tr>
<th>RM</th>
<th>Sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Bowel loops and other abdominal viscera outside the abdominal cavity</td>
<td>T2, volumetric SS-RARE (***)</td>
<td>Axial, sagittal</td>
</tr>
<tr>
<td>Cord insertion in the omphalocele (*)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Peritoneal lining (**)</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

(*) These findings differentiate it from gastroschisis.

(**) This sequence is important to visualize the point of insertion of the umbilical cord.

**Cleft sternum, ectopia cordis, and pentalogy of Cantrell**

These are defects at the level of the sternum. The pentalogy of Cantrell is based on the association of omphalocele, anterior diaphragmatic hernia, sternal cleft, ectopia cordis, and intracardiac defects. The incidence is unknown. It is usually associated with heart defects, cleft lip with or without cleft palate, encephaloceles, exencephalia, and sirenomyelia. It is also associated with trisomies 13 and 18, and Turner's syndrome.

**MR** allows us to see the heart outside the thorax; in the pentalogy of Cantrell pericardiac effusion, pleural effusion, and omphalocele can be seen. We have not encountered these anomalies (26,27).
Bladder exstrophy

The anterior wall of the bladder is absent and the posterior wall is exposed to the exterior (Fig. 33).

Its occurrence is estimated at 1 in 30,000 births, with a male predominance of 2.3:1. In 10% it occurs in same-sex twins.

Certain anomalies are frequently associated, such as epispadias, a defect in the descent of the testes, and bilateral inguinal hernias in males; while cleft clitoris is common in females.

This diagnosis should be suspected in cases where the bladder is absent in the context of normal amounts of amniotic fluid. The presence of a soft-tissue mass on the surface of the lower anterior wall of the abdomen, corresponding to the exstrophied bladder, may also be detected.

<table>
<thead>
<tr>
<th>RM</th>
<th>Sequences</th>
<th>Projections</th>
</tr>
</thead>
<tbody>
<tr>
<td>Absence of the urinary bladder (*)</td>
<td>T2</td>
<td>All</td>
</tr>
</tbody>
</table>

Normal kidneys

Normal amount of amniotic fluid

Irregularities in the anterior wall (**) 

(*) This is the most important finding, together with visualization of normal kidneys and normal amounts of amniotic fluid

(**) Difficult to visualize

Other anomalies of the anterior wall

- **Urachal cyst** is a total or partial failure of the urachus to regress that gives rise to a communication between the bladder and the interior wall of the abdomen. This anomaly can involve a completely permeable urachus, a urachal diverticulum, a urachal sinus, or a urachal cyst. Depending on the type of anomaly, **MR** will depict an anomaly in the entire bladder or a cystic lesion that is hyperintense on T2-weighted images, corresponding to an urachal diverticulum or cyst. There are often other associated anomalies.

- **Inguinal hernia** (Fig. 34) is a rare cause of a scrotal mass. It is often an isolated anomaly and is more common in males than females. It
results from a defect in the closure of the processus vaginalis; situations in which intraabdominal pressure is increased (ascites, meconium peritonitis, intestinal obstruction) can lead to this anomaly. It is necessary to differentiate it from other scrotal masses such as hydrocele, testicular tumor, and torsion of the testes. MR can detect the prominence of the scrotal sac and, depending on the contents of the hernia, structures of distinct characteristics inside.

- **Prune belly syndrome** (Fig. 35) is usually defined as atrophy of the musculature of the abdominal wall. We believe that this terminology is confusing and that Prune belly syndrome is the consequence of other anomalies (almost always of the low urinary tract) rather than an entity in itself. The marked distension of the bladder and ureters produce distension of the abdomen and atrophy of the abdominal wall. MR depicts the anomalies of the urinary tract and the marked distension of the abdomen.
Images for this section:

**Fig. 1:** Fetal anatomy: SS-RARE volumetric single image to assess quantitatively the amount of amniotic fluid and an overview of the fetus

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**Fig. 2:** Fetal anatomy: fetal oropharyngeal cavity. The palate is well depicted in the sagittal and coronal planes (red arrows), the upper jaw is identified in the axial plane (white arrows). The tongue is hypointense and well depicted especially in the sagittal plane (red asterisk). The nose and lips are identified in the coronal plane (black arrows).

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Fig. 3: Fetal Anatomy: A: Midline sagittal image showing the mediastinal esophagus (red arrow), visible only during swallowing, and the trachea (white arrow) B: diaphragm (white arrows), stomach (red arrows) and small intestine (asterisks) C: colon (red arrow), hyperintense structure due to meconium in T1-weighted sequences and hypointense on T2.

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Fig. 4: Fetal anatomy. Liver (red arrows), gallbladder (red asterisk), spleen (white arrow), bladder and male external genitalia (black arrows)

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**Fig. 5:** Cleft lip: fetus at 23 weeks’ gestation. Images are shown in the 3 spatial planes of the oropharyngeal cavity in which it is observed the integrity of the maxilla in sagittal and axial plane (white arrows) and the anomaly of the upper lip (red arrows).

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**Fig. 6:** Unilateral cleft lip and cleft palate: fetus at 21 weeks’ gestation. In the axial plane it is well depicted a large defect in fusion of the maxilla (red arrow). In the coronal plane, can be identified the communication of the oral cavity with the left nostril (white arrows).

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Fig. 7: Bilateral cleft lip and cleft palate: fetus at 22 weeks' gestation. Although difficult, it can be seen on coronal images the slits in the upper lip (black arrows) and bilateral communication of the oral cavity with the nasal cavity (red arrows). In the sagittal plane cannot be identified the maxilla (normal image in the box below left for another fetus without anomaly). The axial image shows the upper jaw which is interrupted in the midline (white arrows).

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Fig. 8: Midline cleft lip and cleft palate: fetus at 21 weeks’ gestation. Midline cleft lip and upper jaw (red arrows). This fetus associated a semilobar holoprosencephaly and chromosomal 13q partial deletion.

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**Fig. 9:** Micrognathia: fetus at 21 weeks’ gestation. It is depicted a small jaw (black arrows). At birth, the diagnosis was confirmed: Goldehan syndrome. The patient also associated preauricular tags and hemivertebrae, anomalies not depicted with MRI.

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Fig. 10: Epulis: fetus at 21 weeks’ gestation. A hypointense mass protruding from the mouth and upper jaw is clearly depicted (red arrows). The tongue and palate are normal. There is amniotic fluid in the mouth and pharynx and stomach (asterisk) is normal. Yet there seems to be some difficulty with swallowing because there polyhydramnios. After birth underwent surgery with satisfactory evolution.

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**Fig. 11:** Cervicothoracic cystic lymphangioma: fetus at 32 weeks’ gestation. A cervicothoracic multicystic heterogeneous mass (red arrows) is clearly identified. We could not identify the trachea and esophagus and assumed that was compressed because the stomach was small (asterisk) and there was polyhydramnios. The fetus died within minutes after birth due to respiratory problems.

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Fig. 12: Esophageal atresia with distal tracheoesophageal fistula, right renal ectopia: fetus at 33 weeks’ gestation. It is observed upper esophageal dilation partially compressing the trachea, the distal esophagus is also displayed (red arrows). There is polyhydramnios (asterisk) and the stomach is little filling (white arrow). The CT scan after birth shows esophageal atresia and distal tracheoesophageal fistula (black arrow). Fetal MRI was performed for suspected renal ectopia in prenatal ultrasound. MRI detected renal ectopia, pelvic kidney (yellow arrow) and esophageal anomaly.

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Fig. 13: Esophageal atresia without tracheoesophageal fistula and duodenal stenosis: fetus at 26 weeks' gestation. The stomach is small (red arrow) and there is polyhydramnios. There is dilatation of proximal esophagus (red arrowhead). The Rx after birth revealed the absence of abdominal air and proximal esophageal pouch, confirming the diagnosis of esophageal atresia without tracheoesophageal fistula. It was decided to postpone the surgery and it was performed a feeding gastrostomy, through which barium study was conducted and there was discovered a duodenal stenosis (green arrows), an anomaly that had not been diagnosed on fetal ultrasound or MRI. The retrospective assessment of fetal resonance imaging showed this abnormality (white arrows).

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**Fig. 14:** Duodenal atresia: fetus at 20 weeks' gestation. It is observed gastric and duodenal dilatation (red arrows). There polyhydramnios. The fetus died at 28 weeks' gestation possibly by placental insufficiency. The autopsy showed atresia of the fourth portion of duodenum.

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Fig. 15: Duodenal stenosis Ladd's band, intestinal malrotation: fetus at 32 weeks' gestation. Significant dilation was observed until its fourth duodenal portion. The stomach was distended discreetly. The volume of amniotic fluid is normal. The large intestine is to the left of the abdomen (white arrows). Radiographs after birth and surgery confirmed this anomaly. There was a Meckel diverticulum (black arrow), an anomaly that had gone unnoticed in the ultrasound and MRI.

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**Fig. 16:** Jejunal atresia: fetus at 25 weeks' gestation. It was observed dilatation of jejunal loops, which are hypointense on T2-weighted and hyperintense on T1 (red arrows). The urinary bladder is marked with a red asterisk. The rectal bulb is just as empty as there is virtually microcolon (green arrows). X-rays after birth showed dilatation of jejunal loops and microcolon.

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Fig. 17: Meconium peritonitis: fetus at 26 weeks’ gestation. It is observed bowel dilatation, ascites (red arrows) and moderate polyhydramnios. In the axial plane image shows a possible peritoneal calcification in the left abdomen (green arrow). The asterisk indicates the umbilical artery in the abdominal portion (also hypointense and not to be confused with intraabdominal calcifications). The CT scan after birth shows peritoneal calcifications and meconium pseudocyst (white arrows).

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Fig. 18: Altered bowel signal intensity (meconium peritonitis): fetus at 21 weeks' gestation. It is noted decreased signal intensity of bowel loops (red arrows). The ultrasound showed hyperechoic bowel loops. The child was born premature. Radiographs at birth showed typical abdominal calcifications of meconium peritonitis (white arrows).

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**Fig. 19:** Heterotaxy syndrome, situs inversus: fetus at 23 weeks’ gestation. The heart is in the correct position (black arrow) but the stomach is on the right side of the abdomen (red arrow). The ultrasonography and barium study confirmed this abnormality at birth, spleen (white asterisk), stomach (white arrows) and liver (green asterisk).

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Fig. 20: Heterotaxy syndrome, polysplenia syndrome: fetus at 20 weeks' gestation. The liver occupies a central position in the abdomen (red arrows). The stomach (asterisk) is also located in the center of the abdomen. The fetal autopsy showed polysplenia, anomaly that could not be demonstrated with MRI. This fetus also had cardiopathy. It is shown the anatomical specimen of these anomalies.

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Fig. 21: Intestinal duplication: fetus at 24 weeks’ gestation. It is identified an structure hyperintense on T2-weighted sequences and hypointense on T1 (red arrows) above the bladder (red asterisks). Radiograph at birth show intestinal obstruction. The surgery showed intestinal duplication bowel and midgut volvulus as a cause of intestinal obstruction.

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Fig. 22: Extrapulmonary sequestration and gastric duplication: fetus at 22 weeks' gestation. Fetal ultrasound detected a cyst behind the stomach. Fetal MRI in addition to viewing the abdominal injury for a gastric duplication (red asterisk) could detect a lesion at the base of the left hemithorax (white arrows) corresponding to extrapulmonary sequestration and had gone unnoticed in the study by ultrasound. CT studies after birth confirmed the presence of these anomalies.

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Fig. 23: Mesenteric cyst (not confirmed): fetus at 20 weeks’ gestation. Fetal MRI demonstrates a hyperintense rounded lesion in the right abdomen (red arrow). The x-ray, ultrasound and CT scan after birth showed the same lesion (red arrows). Further studies with ultrasound and CT showed spontaneous disappearance. The diagnosis of mesenteric cyst is the most probable.

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Fig. 24: Ovarian cyst: fetus at 36 weeks' gestation. Fetal MRI shows a hyperintense lesion in the left abdomen (red arrows). Ultrasound at birth and CT at four months of age showed that the lesion has calcifications inside. The ultrasound at 15 months was only able to visualize the left ovary (white arrow), the right ovary and the lesion had disappeared. The diagnosis of ovarian cyst is the most probable.

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**Fig. 25:** Liver cyst: fetus at 17 weeks' gestation. This fetus with agenesis of one kidney and contralateral dysplasia (white arrow) also had a cyst. Hyperintense lesion is seen apparently in connection with the liver (red arrows) with fluid-fluid level inside. The liver cyst can be seen in the autopsy specimen.

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**Fig. 26:** Hepatic hemangioendothelioma: fetus at 37 weeks' gestation. Fetal MRI study showed a left abdomen mass (white arrows) of uncertain nature located above and in front of the left kidney, below the liver and behind the stomach. Surgery showed that this mass was "hanging" from the bottom edge of the left hepatic lobe and corresponded to a hemangioendothelioma.

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Fig. 27: Neuroblastoma fetus at 32 weeks' gestation. Fetal images show a right adrenal mass moderately hyperintense on T2 and hypointense on T1-weighted sequences. A CT scan done after birth can be seen that the mass has grown and metaiodobenzylguanidine scintigraphy confirmed that this mass corresponds to a right adrenal neuroblastoma (arrows).

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**Fig. 28:** Infradiaphragmatic extralobar pulmonary sequestration (not confirmed): fetus at 23 weeks' gestation. Fetal MRI shows a heterogeneous mass (red arrows) in the left abdomen, between the diaphragm, stomach (white arrows) and above the kidney. All radiological tests identify it after birth, the adrenal gland is normal (black arrow in ultrasonound) and the study was negative with metaiodobenzylguanidine. Could not be find abnormal vascular supply, although we believe that it could correspond to an infradiaphragmatic extralobar pulmonary sequestratio. The analytical data were normal and has not undergone surgery. The child is 7 years old is currently asymptomatic and in periodic inspections the lesion has disappeared.

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**Fig. 29:** Ascites: fetus at 25 weeks’ gestation. There is ascites (red arrows) and significant cardiomegaly (black arrow). The diagnosis of endocardial fibroelastosis was made at necropsy which showed ventricular dilatation and mostly-white endocardium of the interventricular septum. Microscopically, endocardial thickening is observed due to collagen and elastic fibers.

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Fig. 30: Fetal ascites and hydrops: fetus at 26 weeks' gestation. Significant ascites (asterisk) as well as soft tissue edema (red arrows) and pleural effusion (black arrows) can be identified. The fetus died in utero at 33 weeks of gestation, the cause of fetal hydrops was unknown.

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Fig. 31: Gastrochisis: fetus at 22 weeks' gestation. It is observed in both the sagittal and axial images that the bowel loops are outside the abdominal cavity (red arrows), are not covered by peritoneal coverings and the umbilical cord is inserted in the correct position (black arrows).

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Fig. 32: Omphalocele: fetus at 21 weeks’ gestation. The fetal bowel loops can be seen outside the abdominal cavity (red arrows) and that are covered by peritoneal membranes (white arrows). The umbilical cord is inserted into the omphalocele itself (black arrows). The SS-RARE volume imaging may be helpful to identify the umbilical cord.

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**Fig. 33:** Bladder extrophy: fetus at 32 weeks’ gestation. Both kidneys were of normal morphology (white arrows) but the bladder could not be identified (asterisk) and there were irregularities in the lower abdomen anterior wall (red arrow). The volume of amniotic fluid is normal. Courtesy of Dr. Fernando Mas, ERESA. Valencia

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Fig. 34: Inguinal hernia: fetus at 30 weeks’ gestation. The scrotal distention (red arrows) and testes (white arrows) can be depicted. The inguinal canal seems very "open" (asterisk). After birth she underwent surgery. The hernia contained omentum.

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**Fig. 35:** Prune belly syndrome: fetus at 17 weeks’ gestation. Large bladder expansion is observed that produces abdominal distention and causes thinning of the abdominal wall muscles (red arrows). Hydrocephalus is also seen (white arrow). This fetus had a Down syndrome and also associated an esophageal atresia and annular pancreas that could not be diagnosed with MR or ultrasound.

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Results

Our results

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<table>
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<tr>
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<tbody>
<tr>
<td>Total MR studies</td>
<td>704</td>
</tr>
<tr>
<td>Total fetuses</td>
<td>733</td>
</tr>
<tr>
<td>Total cases digestive anomalies</td>
<td>138</td>
</tr>
<tr>
<td>• MR detected</td>
<td>117 (84.7%)</td>
</tr>
<tr>
<td>• US detected</td>
<td>106 (76.8%)</td>
</tr>
<tr>
<td>• MR and US detected</td>
<td>122 (88.4%)</td>
</tr>
<tr>
<td>MR contributed and added data</td>
<td>36 (26.08%)</td>
</tr>
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Conclusion

Conclusions

• Abdominal congenital abnormalities are frequent and varied. MR can help in their prenatal diagnosis.

• In our study, MRI detected more anomalies than the US, although there are exceptions that are best identified by the US.

• Some anomalies are better diagnosed with US, and MR cannot detect all fetal anomalies but when both techniques are used together, the final number of prenatal abdominal anomalies detected increases.

• In our study MR added data in 26.08% of cases.

• We propose the use of MR whenever a fetal abdominal or gastrointestinal anomaly is suspected.
References

Bibliography


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Images for this section:

**Fig. 36:** UDIAT logo

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**Fig. 37:** Hospital Universitari de Sant Joan Logo

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