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Imaging Evaluation of Fetal Megacystis: How Can Magnetic Resonance Imaging Help?

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Evaluation of the kidneys, bladder, and amniotic fluid volume forms part of any standard obstetrical ultrasound. When a fetal genitourinary anomaly is suspected, a more detailed evaluation is necessary. This detailed imaging can be challenging in the setting of decreased or absent amniotic fluid or large maternal body habitus, and in complex malformations. In these situations, magnetic resonance imaging can help to better define the fetal anatomy and provide a more confident and specific prenatal diagnosis.
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Introduction

Megacystis is the term used to describe an enlarged bladder at any gestational age. During the second and third trimesters, this diagnosis is based on an overall subjective assessment and visualization of a persistently enlarged bladder. However, in the first trimester, specific size criteria have been defined.[1,2] The fetal bladder is first seen at approximately 10-12 weeks and its diameter should be no more than 6-8 mm. If the bladder is not seen by 15 weeks or if it is persistently enlarged (especially beyond 15 mm), concern for an underlying genitourinary malformation should be raised and further detailed evaluation during the second trimester is warranted.[3] This assessment typically includes a detailed ultrasound (US) of the kidneys, bladder, external genitalia, and amniotic fluid volume. The US may be complemented with fetal magnetic resonance imaging (MRI), which improves the evaluation of the kidneys with better detection of renal ectopia and cystic dysplasia. It also helps detect underlying anorectal malformations and allows better prediction of lung hypoplasia through calculation of fetal lung volumes in the third trimester, if needed.[4,5]

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The objective of this article is to describe the imaging evaluation of fetal megacystis and discuss how a combined approach with US and fetal MRI can be beneficial.

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Imaging Checklist for Fetal Megacystis

A systematic and organized approach to any imaging analysis enables collection of all the essential imaging clues and identification of the most likely diagnosis or at least the main differential considerations. The most important elements to be assessed in the presence of fetal megacystis are discussed.

Amniotic Fluid Volume

US can quantify the amniotic fluid volume. A frequently used method is the evaluation of the *single deepest vertical pocket* free of fetal parts and umbilical cord. In normal conditions it measures between 2 and 8 cm.[6] There is no standardized method for quantification of amniotic fluid volume with fetal MRI, but it allows a subjective overall assessment.

After the first trimester, if there is persistent renal dysfunction or lower urinary tract obstruction, the amniotic fluid volume is expected to decrease. Preserved amniotic fluid volume can be seen in less severe forms of lower urinary tract obstruction, in functional megacystis (where no anatomical obstruction is present), and in cases of obstructive megacystis with a combined proximal gastrointestinal obstruction (Fig. 1).[2] Alternatively, development of polyhydramnios in the third trimester has been described with megacystis-microcolon-intestinal hypoperistalsis syndrome, due to associated bowel dysmotility.[7,8]

Bladder

US evaluation of the bladder in the setting of massive megacystis, tortuous hydroureters, or extrinsic masses can be

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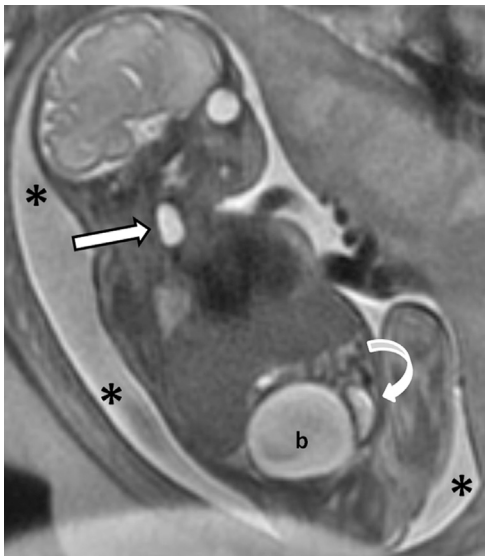

Figure 1 A 29-week female fetus with esophageal atresia and bladder outlet obstruction in the setting of cloacal malformation and VACTERL association. Fetal MRI T2-weighted coronal oblique image shows fluid dilatation of the cervical esophagus (arrow), known as the *pouch sign*, which is consistent with esophageal atresia; this finding was noted only once during the entire examination and was undetected with US. The stomach was small but the amniotic fluid volume (asterisk) was normal due to an associated bladder outlet obstruction. The bladder is enlarged (b) and there is hydrocolpos (curved arrow). Radial ray deficiency (not shown) and congenital heart defect were also present.

challenging owing to anatomical distortion and limitations in differentiating adjacent bowel. Fetal MRI allows a better understanding of the fetal anatomy, the level of obstruction, and associated findings.

The bladder is adjacent to the anterior abdominal wall and bordered on each side by the umbilical arteries (Fig. 2).[2] If a pelvic or abdominopelvic cystic structure keeps this anatomical relationship with the umbilical arteries and the abdominal wall, it truly represents the bladder. Otherwise, the cystic lesion could represent a different structure, such as hydrocolpos (Fig. 3), enteric duplication cyst, bowel dilatation, meconium pseudocyst, lymphatic malformation, ovarian cyst, anterior sacral meningocele, or a type IV sacrococcygeal teratoma.[9] The bladder configuration and content can provide diagnostic clues. Long-standing obstructive bladders may eventually develop wall thickening and trabeculation. In the presence of a dilated bladder and posterior urethra (known as the keyhole sign), posterior urethral obstruction should be considered (Fig. 4). Fetal MRI can help define more subtle or intermittent posterior urethral dilatation, as the examination allows visualization of the same anatomical area over a longer period of time (Fig. 5). Elongation of the bladder dome toward the abdominal cord insertion supports a patent urachus, potentially seen as a decompression pathway in the setting of severe bladder outlet obstruction (Fig. 6).[10] Enlarged bladders with lobulated contours or with large diverticula have been reported in cloacal malformations, and in some cases they may have associated layering debris, which represents meconium and supports an abnormal fistulous connection with the colon (Fig. 7).[11-13]

Evaluation of the Large Bowel for Potential Underlying Anorectal Malformations or Microcolon

Assessment of the bowel provides important clues in the context of genitourinary anomalies. Compared with US, fetal MRI is advantageous because it is able to easily track the course of the entire colon and rectum once it is distended with meconium. In normal circumstances, meconium displays a dark T2 and bright T1-weighted signal, starts accumulating in the distal rectum at 20-21 weeks, and fills the entire colon after 25 weeks.[14-16] Subsequently, the content and caliber of the large bowel increase homogeneously during the third trimester.[17] On a sagittal view, the rectum should be seen

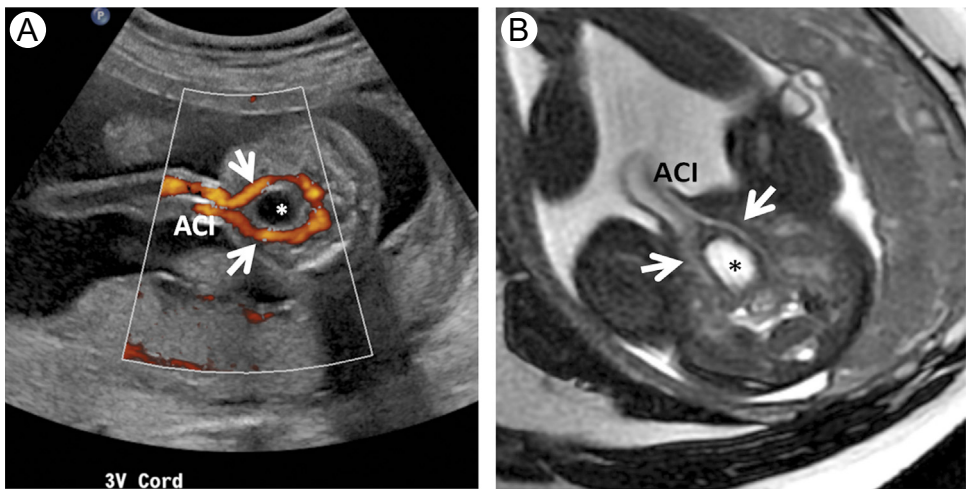

Figure 2 Normal fetal bladder with a 3-vessel cord. Axial color-Doppler US (A) and fetal MRI, balanced steady-state free-precession (B) images. The umbilical arteries (arrows) run from the abdominal cord insertion (ACI) parallel to the lateral walls of the bladder (asterisk) to join the iliac arteries. (Color version of figure is available online.)