REVIEW ARTICLE

Three-dimensional Ultrasonographic Evaluation of the Fetal Posterior Fossa

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Abstract

Posterior fossa malformations can be considered as very frequent brain anomalies. Irrespective of the rapid development of fetal imaging, the frequency of both false-negative and false-positive diagnoses of posterior fossa anomalies did not change, making the diagnosis difficult. Fetal posterior fossa abnormalities may be broadly divided into hindbrain malformations, including diseases with cerebellar or vermian agenesis, aplasia or hypoplasia, and cystic posterior fossa anomalies, and cranial vault malformations or Chiari malformations. Categorization of posterior fossa anomalies is still controversial and there is no uniform approach. Multidimensional mode in fetal neurosonography by 3D probes has several advantages over standard 2D probes. Unlimited offline analysis by using three orthogonal planes of the fetal brain, tomographic ultrasound imaging (TUI), and volume contrast imaging (VCI) may be obtained by a single 3D acquired data. These neuroimaging modes allow obtaining more precise information on fetal posterior fossa and the results are comparable to those obtained with fetal magnetic resonance imaging (MRI). Three-dimensional ultrasonography is an invaluable instrument for differential diagnosis of posterior fossa anomalies. Transabdominal or transvaginal 3D ultrasonography allows thorough assessment of the complex anatomic structures of the posterior fossa and improves diagnostic accuracy and diagnostic confidence having a positive clinical effect in most of the cases.

Keywords: Congenital malformations, Fetal posterior fossa, Three-dimensional ultrasonography.

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Introduction

Posterior fossa malformations can be considered as very frequent brain anomalies. Irrespective of the rapid development of fetal imaging, the frequency of both false-negative and false-positive diagnoses of posterior fossa anomalies did not change, making the diagnosis difficult.[1–3] Fetal posterior fossa abnormalities may be broadly divided into hindbrain malformations, including diseases with cerebellar or vermian agenesis, aplasia or hypoplasia, and cystic posterior fossa anomalies, and cranial vault malformations or Chiari malformations.

Evaluation of the fetal posterior fossa is an essential part of routine fetal ultrasonography. Thanks to advances of neuroimaging in the last 20 years increased the frequency of the evaluation of the posterior fossa anomalies. However, fetal posterior fossa contains complex anatomic structures, and therefore, conventional two-dimensional ultrasonography may sometimes have limited value in detecting anomalies especially in the second trimester of gestation. And when an abnormality is found by standard two-dimensional ultrasonography, many pathologies can be suspected: from normal findings to severe anomalies that may look alike or even identical. Nevertheless, categorization of posterior fossa anomalies is still controversial and there is no uniform approach.[4–6] Differential diagnosis of these pathologies may require additional sonographic planes such as sagittal and coronal planes and acquiring those may be rather difficult in certain cases. Fetal MRI and 3D or 4D ultrasonography allow complete assessment of the complicated anatomic structures of the posterior fossa increasing diagnostic possibilities in most of the cases.[7] Brainstem, cerebellum, fourth ventricle (4V), and cisterna magna are better assessed with these techniques. Having said that, fetal MRI is available only in specialized centers and may not be accepted by the family due to the complexity of the procedure. Another novel option is 3D or 4D evaluation of the fetal brain. Ultrasonographic machines with 3D or 4D capabilities are readily available in most of the perinatology or Ob and Gyn centers, and families are more familiar and feel safe with those machines though fetal MRI is superior to ultrasonography in cases with oligohidramnios, maternal obesity, and fetal skull ossification.

By transabdominal ultrasonography, fetal brain anatomy may be visualized fairly in axial sections. However, fetal cranial bone ossification, thick maternal abdominal wall, and anterior placenta may render transabdominal ultrasonography difficult and ineffective. This obstacle may be overcome by high-resolution transvaginal ultrasonography and high-quality images may be obtained from acoustic window through anterior fontanelle and sagittal sutures. Although it is critically valuable in case of the vertex presentation, acquiring essential sections may be difficult especially for those who are not acquainted with transvaginal fetal neurosonography. Multidimensional mode in fetal neurosonography by 3D transvaginal probes has several advantages over standard 2D transvaginal probes. Unlimited offline analysis by using three orthogonal planes of the fetal brain, TUI, and VCI may be obtained by a single 3D acquired data. These neuroimaging modes allow obtaining more precise information

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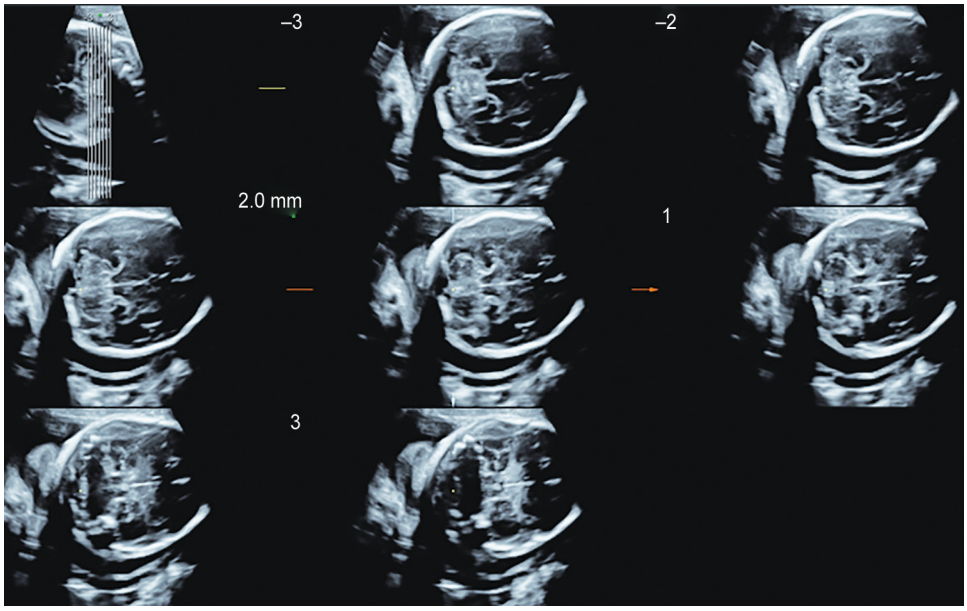

Fig. 1: A 3D volume data set of axial acquisitions of fetal posterior fossa displayed in tomographic mode

on fetal posterior fossa and the results are comparable to those obtained with fetal MRI. Paladini et al. showed that the overall accuracy rates of expert neurosonography and fetal MRI were similar (91.3% vs 94.4%), respectively, and fetal MRI was more useful after 24 weeks of gestation.[8]

Axial view is used for the assessment of cerebellar anatomy including depiction of the normal shape of the vermis in the middle and normal cerebellar hemispheres on both sides. Visualization of inferior part of the vermis separating 4V from the cistern is also important (Fig. 1). When the fetus is in the breech presentation, the cerebellar vermis can be depicted transabdominally through the anterior fontanelle similarly to the depiction of the corpus callosum (Figs 2 and 3).

In order to diagnose anomalies of the vermis, it should be visualized together with neighboring structures in the axial planes in midsagittal view, enabling objective assessment of the vermian size and the shape (Figs 2 to 5).

In this paper, we would like to summarize the main fetal posterior fossa abnormalities and emphasize the importance and significance of 3D ultrasonography in differential diagnosis of those abnormalities.

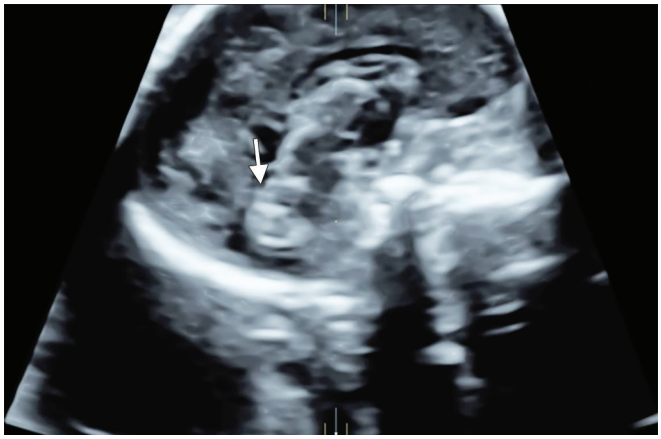

Fig. 2: Vermis (arrow) and posterior fossa can be reconstructed after a 3D volume acquisition. In this reconstruction, the corpus callosum can also be seen in this midline section

Vermian–Cerebellar Hypoplasia

All forms of incomplete development of the vermis and cerebellum defined as vermian–cerebellar hypoplasia can be detected by ultrasound.[9] If there is no fluid collection which may happen in vermian hypoplasia, then the diagnosis is difficult and puzzling.[10] Paladini et al. showed that 3D ultrasound may be helpful in the diagnosis of the significant hypoplasia of the cerebellar vermis, showing excellent correlation with MRI in terms of morphometric measurements of the vermis,[11] which makes 3D ultrasound an appealing choice.

Dandy–Walker Malformation

Dandy-Walker malformation (DWM) is a cerebellar abnormality characterized by the dysgenesis of cerebellar vermis and cystic dilatation of the 4V. The best diagnostic clue for DWM is a large fetal posterior fossa with big cerebrospinal fluid (CSF) cyst and the 4V appears contiguous with the posterior fossa cyst. The tentorium and torcular herophili are elevated. Hydrocephaly, aqueductal stenosis, dysgenesis of the corpus callosum, brainstem dysplasias, migration anomalies, schizencephaly, lipomas, cephaloceles, and lumbosacral meningocele may be associated with DWM. Differential diagnosis of DWM may be difficult and contains Blake's pouch cyst (BPC) and vermian agenesis/hypoplasia. Diagnosis of DWM by using only axial transabdominal ultrasonography may result in erroneous diagnosis.[12] For a proper diagnosis, assessment of the vermis and torcular herophili is essential by a median view.[5,6,13] Three-dimensional ultrasonography may be invaluable for those cases due to the ability to control the planes using the orthogonal planes as reference.[14,15] Furthermore, VCI may assist visualization of subtle anatomical details[14,15] (Fig. 6).

Blake's Pouch Cyst

Blake's pouch is a finger-like outpouch of the 4V and is an embryological remnant. Enlargement of this remnant may lead to

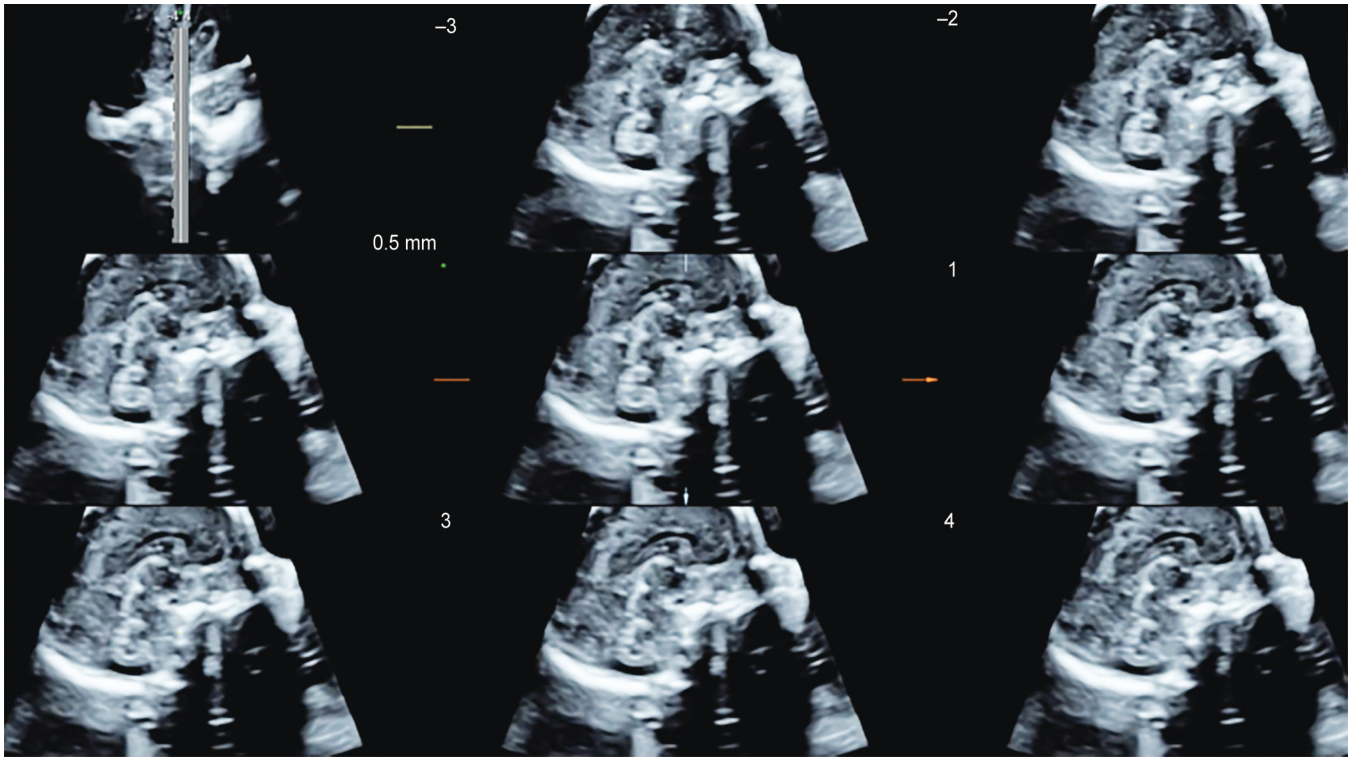

Fig. 3: Sagittal and parasagittal sectional planes after a transabdominal 3D acquisition through the fontanel with a rendering in tomographic mode. The focus is on the midline structures, which are well-recognized as the corpus callosum and vermis

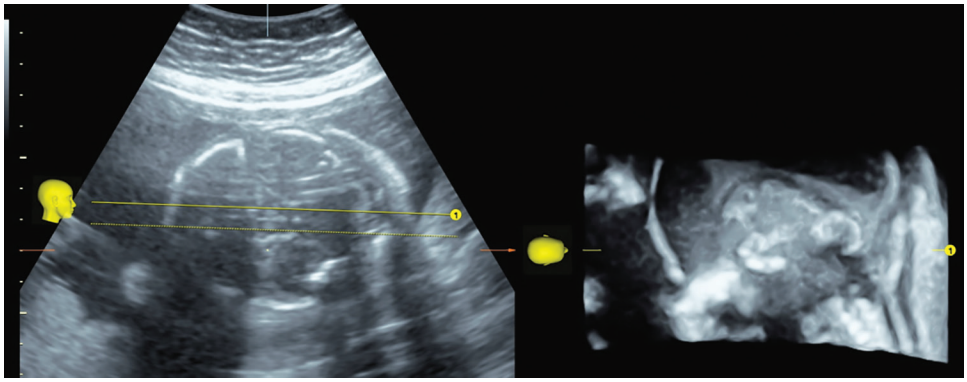

Fig. 4: Omniview with volume contrast imaging for the demonstration of vermis and brainstem

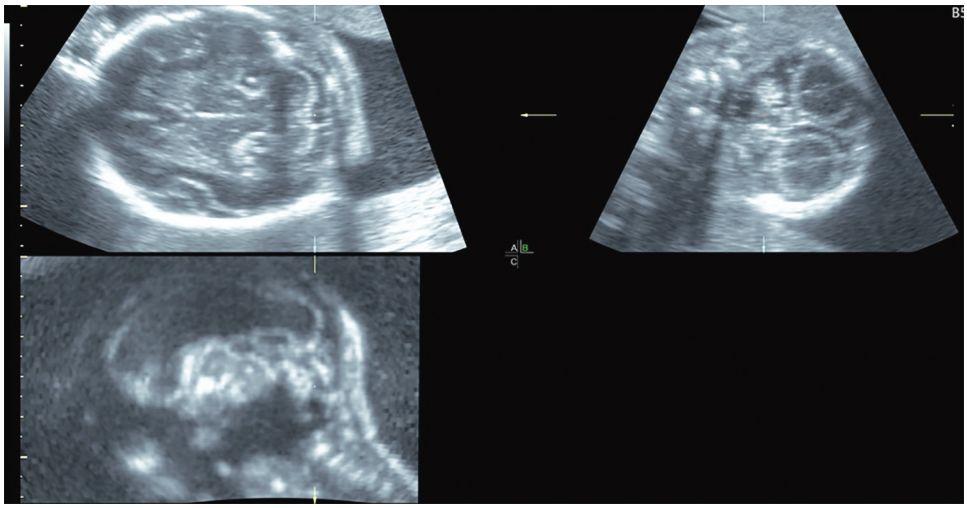

Fig. 5: Posterior fossa with multiplanar imaging

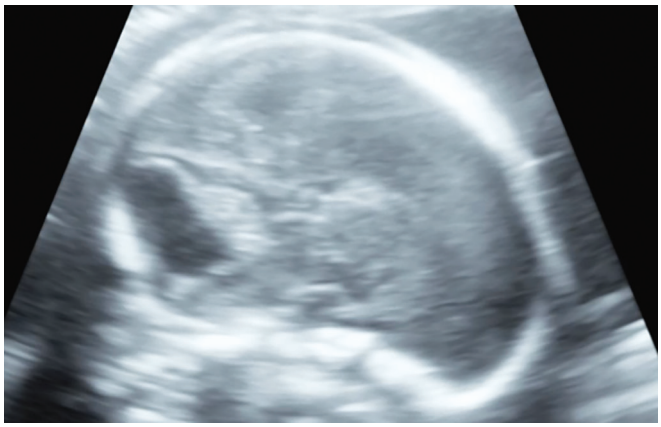

Fig. 6: Detailed sonographic demonstration of Dandy–Walker malformation with volume contrast imaging mode

a slight superior displacement of the vermis and is called BPC. As this gives an impression of 4V contiguous with the posterior fossa cyst, it is an important entity in the differential diagnosis of DWM. In BPC, the cerebellar vermis, cisterna magna, and torcular herophili are in normal position. Assessment of these characteristic features is possible with 3D ultrasonography. Paladini et al. found that the upper wall of the cyst may be visible in 11/19 of the cases.[16]

Megacisterna Magna

Widening of the cisterna magna called megacisterna magna while cerebellum looks normal including the vermis and both hemispheres. It is usually an isolated finding with good prognosis if associated anomalies are not present.[17] Nevertheless, it is an important entity because it should be differentiated from DWM, BPC, and vermian agenesis/hypoplasia. The key plane is the sagittal one, and by 3D ultrasonography, essential median and paramedian planes may be obtained easily.

Spinal Dysraphism and Chiari II Malformation

Spinal dysraphism refers to a vertebral open defect in which the spinal contents protrude through the bony content. In Chiari II malformation, open spinal dysraphism results in herniation of the cerebellar vermis and brainstem through the foramen magnum.[18] The diagnosis of Chiari II malformations may be achieved by scanning the spine in three planes (axial, sagittal, and coronal) by 2D ultrasound. However, due to maternal body properties and fetal position, not all planes may be accessible in all cases. Using 3D ultrasonography, the spine may be displayed along three orthogonal scanning planes, and therefore, 3D ultrasonography presents obvious advantages.

As a conclusion, different fetal posterior fossa abnormalities may have similar appearances and differential diagnosis is crucial as they have distinct prognosis. Three-dimensional ultrasonography is an invaluable instrument for differential diagnosis of these entities. Transabdominal or transvaginal 3D ultrasonography either transabdominal or transvaginal is used to assess the complex anatomic structures within the posterior fossa, and by detailed evaluation, it improves diagnostic accuracy and diagnostic confidence having a positive clinical impact in most of the cases.

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