Prenatal diagnosis and outcome of fetal posterior fossa

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Learning objectives

Posterior fossa malformations are best classified in terms of their embryogenesis from the rhomb encephalon. The diverse manifestations of the dysgenesis can be related to the stages at which development of the cerebellum became deranged. The cerebellar malformations can also be classified by their effect on the fourth ventricle and cisterna magna, and by whether any cystic spaces represent expansion of the rhomb encephalic vesicle or secondary atrophy of the parenchyma. The size of the posterior fossa depends largely on the size of the rhomb encephalic vesicle at the time that the mesenchyme condenses into the bony-dural walls of the posterior fossa, so identification of the size of the posterior fossa often proves to be more helpful in differentiating among these malformations than the simple presence, or absence, of a cyst.

This study aims to:

- Illustrate the normal posterior fossa anatomy in prenatal ultrasound and MRI.
- Define the advantages of fetal MRI to explore the fetal posterior fossa.
- Review and describe the spectrum of the various posterior fossa malformations diagnosed in antenatal.
Background

The primary prenatal imaging technique of fetal brain is ultrasound. It is able to explore perfectly the cerebral parenchyma. The study of posterior fossa has always been based upon the appearance on both axial and mid-sagittal views of the fetal head. The mid-sagittal plane allows a complete view of the posterior fossa, vermian and tentorium. Sometimes, ultrasound is limited in the exploration of posterior fossa because of the position of fetus and low amniotic fluid.

Fetal MRI offers several advantages over prenatal US. It has higher contrast resolution and can be easily performed using ultrafast T2-W sequences.

Because the fetal brain is a dynamic structure, it is important to familiarize with the normal appearance of the fetal brain at different gestational ages in order to be better able to identify and characterize abnormalities with fetal MRI.

Normative measurements of the pons on fetal MRI have been published and the pons should be measured in cases of suspected cerebellar or supratentorial abnormalities.

The brainstem can also be examined for focal or diffuse morphologic or signal abnormalities.

The dorsal pons and medulla normally appear hypointense on T2-W images and hyperintense on T1-weighted images relative to the ventral brainstem as early as 23-25 gestational weeks. The dorsal midbrain appears hypointense on T2 and hyperintense on T1 later in gestation, by about 31-32 weeks.

Diffusion-weighed imaging can be used to evaluate the brainstem, as there is a normal decline in mean diffusivity in the pons with increasing gestational age.
Findings and procedure details

Posterior fossa malformations are among the most common brain anomalies identified by current fetal imaging techniques. Despite the rapid progress in fetal imaging, the prenatal diagnosis of posterior fossa dysgenesis remains challenging due to both false-positive and false-negative diagnoses. As a result, a variety of classification schemes have been proposed for posterior fossa anomalies.

Advances in MRI during fetal and early postnatal life continue to provide important insights into normal and abnormal development of the cerebellum and brainstem. It is helpful in evaluating abnormalities of the posterior fossa. It allows direct visualization of the cerebellar hemispheres, vermis, and brainstem in three orthogonal planes and thus allows better assessment of their morphology. Normative measurements of the cerebellum, vermis, and brainstem on fetal MRI are available for different gestational ages.

Because many posterior fossa abnormalities are associated with supratentorial abnormalities, fetal MRI is also used to evaluate the supratentorial brain when an infratentorial abnormality is identified.

Fetal MRI is performed on 1.5-tesla MR scanner using a multi-channel phased array coil to allow increased coverage of the fetal head and increased signal-to-noise ratio. The mother lies supine during the course of the exam (typically 45-60 min). The mother is made as comfortable as possible during the MR exam in order to minimize fetal motion. It is primarily performed using ultrafast T2-W sequences known as single-shot rapid acquisition with refocused echoes.

Posterior fossa abnormalities evaluated by fetal MRI include Dandy-Walker continuum, cerebellar hypoplasia, mega cisterna magna and Chiari II malformation.

**Dandy Walker:**

The Dandy-Walker malformation was first reported by Dandy and Blackfan (1914) who described marked dilatation of the fourth ventricle and anterior displacement of the vermis, which they attributed to primary atresia of the cerebellar foramina.

It occurs in 1 per 25,000-30,000 births. There is a slight female predilection. Dandy-Walker malformation accounts for 2%- 4% of hydrocephalus and 14% of cystic posterior fossa malformations.

The typical Dandy-Walker malformation is characterized by a triad of 1) complete or partial agenesis of the vermis, 2) cystic dilatation of the fourth ventricle, and 3) an
enlarged posterior fossa with upward displacement of the lateral sinuses, tentorium, and torcular (fig 1,2)

**Cerebellar hypoplasia:**

Global cerebellar hypoplasia may result from several causes. It has been reported in:

- Chromosomal abnormalities (trisomies 13 and 18)
- Metabolic disorders (Zellweger or Smith-Lemli-Opitz syndrome)
- Several genetic syndromes (CHARGE syndrome or velocardiofacial syndrome)
- Migrational disorders

Unilateral cerebellar hypoplasia is more common than bilateral hypoplasia. Unilateral hypoplasia of the cerebellum is associated with hypoplasia of the ipsilateral dentate nucleus and the contralateral inferior olivary nuclei and pontine nuclei. Bilateral cerebellar hypoplasia is associated with hypoplasia of these nuclei bilaterally and readily appreciable diminution in the brain stem, particularly the pons and medulla (fig3)

**Mega cisterna magna:**

The term mega cisterna magna refers to a cystic malformation of the posterior fossa characterized by an intact vermis, an enlarged cisterna magna, and an enlarged bony/dural posterior fossa. The size of the mega cisterna magna is variable. The fourth ventricle communicates with the subarachnoid space. The brain stem and the cerebellum are usually normal. This condition accounts for 54% of the cystic posterior fossa malformations.

**Arachnoid cyst:**

It occurs in 10% in cases the posterior fossa. It is due to a duplication of the aracnoid membrane producing fluid-filled cysts. Arachnoid cysts do not communicate with the fourth ventricle or the subarachnoid space. There is no reported recurrence risk.

MRI reveals a well-circumscribed extraaxial fluid collection or cyst that is isointense relative to CSF with all sequences (fig 4)

**Black pouch cyst:**
Blake pouch cyst is caused by lack of fenestration of the Blake pouch, resulting in absence of communication between the fourth ventricle and the subarachnoid space and leading to tetra ventricular hydrocephalus. The cerebellum has a normal size and shape. Blake pouch cyst occurs sporadically, and no recurrence risk has been reported.

MRI shows the presence of a cyst in a retrocerebellar or infraretrocerebellar location, which is essentially a diverticulum of the consequently enlarged fourth ventricle (fig 5).

The presence of this cyst is responsible for the displacement of the choroid plexus inferior to the vermis along the antero superior aspect of the cyst. The consistent presence of hydrocephalus allows the differentiation of Blake pouch cyst from mega cisterna magna. Mild mass effect may result in indentation of the inferior vermis or of the caudal and medial aspects of the cerebellar hemispheres. The posterior fossa is typically normal in size. Supratentorial morphologic abnormalities other than hydrocephalus are usually absent.

**Chiari II:

The feature of the Chiari II malformation that have been most useful are the infratentorial findings, these include a small posterior fossa with effacement of the cisterna magna and deformation of the cerebellum called banana sign (fig 6).

Many supratentorial abnormalities have also been described. Included in these are callosal dysgenesis, a small third ventricle, enlarged interthalamic adhesions, a beaked tectum, polymicrogyria, heterotopias, skull deformities (flattened frontal bones called "lemon sign"), colpocephaly, and other cause of ventriculomegaly. (fig 7)
Fig. 1: Sagittal and axial T2W SS SFE MRI showing an enlarged posterior fossa with vermis agenesis and elevation of troncular in 22 gestational week fetus. Note the ventriculomegaly.

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Fig. 2: Axial and mid-sagittal fetal brain ultrasound showing a cystic dilatation of the 4th ventricle with vermian agenesis and troncular ascension.

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Fig. 3: Sagittal and coronal T2W SSFSE MRI showing bilateral cerebellar hypoplasia in 24 gestational week fetus.

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**Fig. 4:** Sagittal T2W SSFSE MRI shows a well-circumscribed extraaxial fluid cyst iso intense to CSF.

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**Fig. 5:** Sagittal and axial T2W SSFSE showing a normal size posterior fossa with an retrocerebellar cyst communicating with the 4th ventricule.

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Fig. 6: Axial plane of fetal brain ultrasound showing an anterior curving of the cerebellum "banana sign" due to small posterior fossa (A). Deformity of frontal bones which lose their convex contour and appear flattened "lemon sign"(B).

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Fig. 7: Axial and sagittal T2W SSFSE MRI confirming the ultrasound data with a small posterior fossa, ptosis of the cerebellar tonsils through the magnum orifice and deformity of frontal bones into "lemon sign". Note the supratentorial ventriculomegaly.

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Conclusion

Prenatal neurosonography and MRI are similarly accurate in the categorization of posterior fossa malformations.

Blake's pouch cyst and megacisterna magna are risk factors for associated anomalies but when isolated have an excellent prognosis, with a high probability of intrauterine resolution and normal intellectual development in almost all cases. Conversely, Dandy-Walker malformation and vermian hypoplasia, even when they appear isolated antenatally, are associated with an abnormal outcome in half of cases.

The early diagnosis may help to determine the prognosis of the pregnancy and indicate on time a therapeutic abortion.
References


