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Review

Diagnostic imaging of posterior fossa anomalies in the fetus

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SUMMARY

Ultrasound and magnetic resonance imaging are the two imaging modalities used in the assessment of the fetus. Ultrasound is the primary imaging modality, whereas magnetic resonance is used in cases of diagnostic uncertainty. Both techniques have advantages and disadvantages and therefore they are complementary. Standard axial ultrasound views of the posterior fossa are used for routine scanning for fetal anomalies, with additional orthogonal views directly and indirectly obtainable using three-dimensional ultrasound techniques. Magnetic resonance imaging allows not only direct orthogonal imaging planes, but also tissue characterization, for example to search for blood breakdown products. We review the nomenclature of several posterior fossa anomalies using standardized criteria, and we review cerebellar abnormalities based on an etiologic classification.

1. Introduction

Evaluation of the posterior fossa is an essential part of routine fetal sonography. Standard measurements of the cisterna magna (between 2 and 11 mm) and ventricular atrium (<10 mm) confer a very high negative predictive value ($P < 0.005\%$) for abnormal central nervous system and spinal cord development [1]. On routine axial sonography, several of the pathologies appear similar but further assessment using direct orthogonal views of the posterior fossa are possible and further characterization is possible using three-dimensional ultrasound and magnetic resonance imaging (MRI).

2. Embryology

The cerebellum develops from the rhombic lips at the cranial end of the roof of the rhombencephalic vesicle and the vermis and cerebellum grow exophytically, inferiorly and laterally to cover it. This process is usually complete by around 18 weeks' gestation [2].

The vermis does not completely cover the roof of the fourth ventricle, and the lower part of the roof extends beneath the vermis into the overlying meninx primitiva as an evagination known as Blake's pouch [3]. Blake's pouch fenestrates to a variable degree down to the obex (the inferior recess of the fourth ventricle) leading to a communication with the subarachnoid space of the surrounding cisterna magna (Fig. 1).

3. The Dandy–Walker continuum: review of nomenclature

The Dandy–Walker continuum comprises classic Dandy–Walker malformation, Dandy–Walker variant (also known as inferior vermian hypoplasia), Blake's pouch cyst and mega cisterna magna [4,5]. A standardized definition of this spectrum of pathologies is shown in Table 1, though this is still controversial.

Current theories suggest that the spectrums of findings are due to a global developmental defect affecting the roof of the rhombencephalon leading to variable degrees of vermian hypoplasia and/or failure of fenestration of Blake's pouch (Fig. 2).

3.1. Classic Dandy–Walker malformation and Dandy–Walker variant

Classic Dandy–Walker malformation comprises vermian hypoplasia, dilatation of Blake's pouch and enlargement of the posterior fossa. Hydrocephalus is a frequent complication but not included in the definition, and is not usually present at birth. Dandy–Walker

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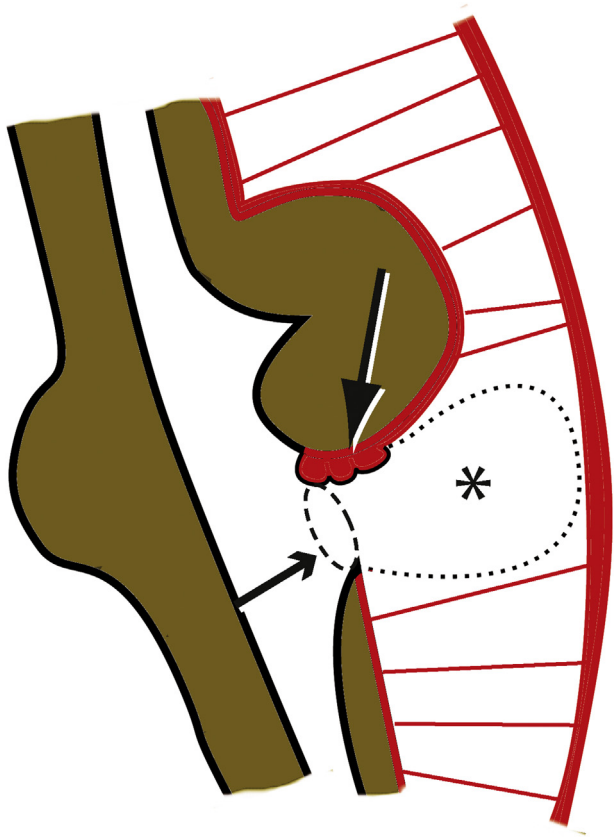

Fig. 1. Blake's pouch (*) is an evagination of the posterior membranous area of the roof of the fourth ventricle into the developing subarachnoid space. Once the pouch fenestrates, the neck of the pouch (short arrow) is known as the foramen of Magendie. The normal choroid plexus (long arrow) is attached to the superior margin of the pouch. (Adapted from Robinson and Goldstein [17] with permission.)

variant is used to describe those without an enlarged posterior fossa.

3.2. Inferior vermian hypoplasia

The term "inferior vermian hypoplasia" has been in use as an alternative to the nomenclature "Dandy–Walker variant" and has arisen out of the preconception that the vermis develops from superior to inferior, and that partial agenesis therefore involves its inferior part. This entire concept has recently been challenged [6], and therefore the preferred nomenclature in the Dandy–Walker continuum is simply "vermian hypoplasia".

3.3. Isolated inferior vermian hypoplasia

The term "isolated inferior vermian hypoplasia" has been used to describe the subset in which there are no known underlying or associated abnormalities. Though this group generally has a better outcome [7–10], it may be impossible to determine prenatally whether vermian hypoplasia is isolated since associated genetic and/or chromosomal abnormalities may be undetectable [11].

3.4. Blake's pouch cyst

Blake's pouch cyst is thought to result from inadequate fenestration of both Blake's pouch and the foramina of Luschka, leading to inadequate cerebrospinal fluid egress, enlargement of the pouch and failure of descent/rotation of the vermis towards the brainstem [12–14].

There is difficulty in distinguishing a Blake's pouch cyst from vermian hypoplasia because both have an enlarged Blake's pouch and a rotated vermis, but with differing appearance of the vermis [15]. The position of the choroid plexus is also thought to be important in that if it is displaced from its normal position on the inferior surface of the vermis, then it indicates that the anterior membranous area formed abnormally [16], and thus further evaluation of the vermis is warranted (Fig. 3).

3.5. Mega cisterna magna

Mega cisterna magna is thought to result from a defect of the posterior membranous area during embryogenesis. The vermis is normal but with mildly deficient fenestration of Blake's pouch insufficient to cause elevation of the vermis [17]. In this sense, mega cisterna magna may be considered a "mega Blake's pouch". By prenatal imaging the definition of mega cisterna magna is an enlargement of the cisterna magna to >10 mm, which often regresses in utero [18].

4. Assessment and classification

A wide variety of etiologies may lead to the similar imaging phenotypes previously described. The key determinants of outcome are the degree of hypoplasia in the cerebellum/vermis and brainstem, and the presence of associated abnormalities.

4.1. Anatomical assessment by imaging

4.1.1. Landmarks

By 17.5 weeks of gestation the major vermian landmarks should be present as determined by in-vitro studies, including the fastigial point and the primary fissure. The culmen and declive should be visible immediately above and below the primary fissure respectively (Fig. 4).

4.1.2. Growth

Normal biometric parameters for the vermis and cerebellum, pons and brainstem have been described both ultrasonographically and by MRI [19–23].

4.1.3. Lobulation

Assessment of vermian lobulation is critical because vermian and regional cerebellar morphology have been demonstrated to correlate with prognosis [24,25]. The vermian fissures become visible in the following specific sequence: pre-pyramidal at 21 weeks, pre-culmenate at 21–22 weeks, secondary fissure at 24 weeks, and all the fissures visible by 27 weeks [26]. Therefore, prior to 27 weeks of gestation, it may be difficult to determine whether vermian lobulation is appropriate for gestational age (Fig. 4).

4.1.4. Angulation

As the cerebellum and vermis develop, the angle formed between their attachment at the rhombic lips and the brainstem gradually decreases. This developmental appearance is often referred to as "closure" of the fourth ventricle. The normal fetal tegmento-vermian angle is usually <18°. A significantly elevated tegmento-vermian angle (>40–45°) is typically associated with the classic Dandy–Walker malformation [27] (Fig. 5).