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REVIEW ARTICLE

Demystifying Posterior Cranial Fossa Lesions in the Fetus

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Abstract Posterior fossa lesions are rare. Introduction of high resolution ultrasound with 3D reconstruction and endovaginal evaluation of fetal brain through the fontanelle helps in better understanding of neuroanatomy. MRI is often used as an additional tool for evaluation in addition to ultrasound studies. Major components of posterior fossa such as cerebellar hemispheres, vermis, fourth ventricle, cisterna magna, and their normal variants and abnormalities can be demystified by ultrasound studies. Morphometry of these structures helps in the diagnoses of these abnormalities.

Keywords Fetal brain · Posterior fossa · Abnormalities · Ultrasound · Endovaginal MRI

Introduction

Posterior fossa abnormalities in the fetus include a wide variety of different entities ranging from normal variants to severe abnormalities [1]. It is essential to demystify these ultrasound features as it has direct bearing on the diagnosis, counseling, and management of these abnormalities.

Present ultrasound techniques make it possible to suspect posterior fossa abnormalities in the first trimester and confirm them at midgestation.

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This compilation was based on the prospective and retrospective evaluation by the author, of cases from 2011 to 2015.

Techniques involved in the evaluation of posterior fossa are followed as per ISUOG guidelines [2] authored by neurosonology task force which has recommended certain basic views and extended advanced evaluation of the fetal cranium.

The basic views of the cranium are transthalamic view (Fig. 1) for visualization of cavum septum pellucidum, transventricular view (Fig. 2) to see the size of the lateral ventricles at perito-occipital sulcus, and transcerebellar view (Fig. 3) to evaluate the posterior fossa.

Posterior fossa lesions are evaluated by certain specific views, namely suboccipito bregmatic view (Fig. 4), and the sagittal view (Fig. 5). These sections can be obtained transabdominally or transvaginally if the fetus is in cephalic presentation. Sagittal view of fetal cranium is an important component in the evaluation of posterior fossa and at times, is difficult to obtain by conventional 2D examination. In such circumstances, one can opt for 3D multiplanar reconstruction.

Embyologically, there are three cerebral vesicles, of which the fourth ventricle or the rhombencephalon is the largest. Rhombencephalon is initially covered by the small cerebellum, which gradually increases in size and engulfs the fourth ventricle. Important constituents of posterior fossa are vermis which connects both the cerebellar hemispheres that enclose fourth ventricle and its extension cisterna magna. Torcular herophili, which has the straight sinus is difficult to identify on ultrasound, approximately corresponds to tentorium. In the occipito-bregmatic view (Fig. 4) vermis is demonstrated as an echogenic rectangular structure and fourth ventricle is seen as a small fluid-filled structure in front of the vermis. In the sagittal section

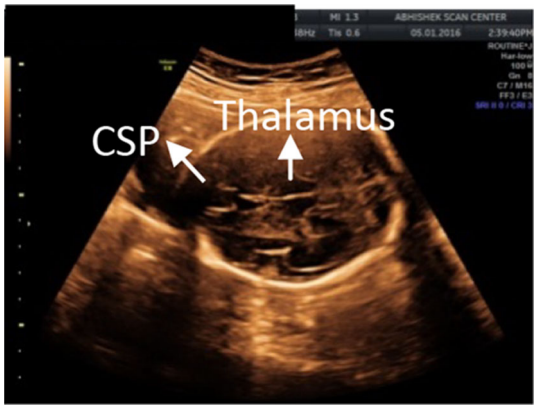

Fig. 1 Transthalamic section—midline falx, cavum septum pellucidum (CSP), and thalamus

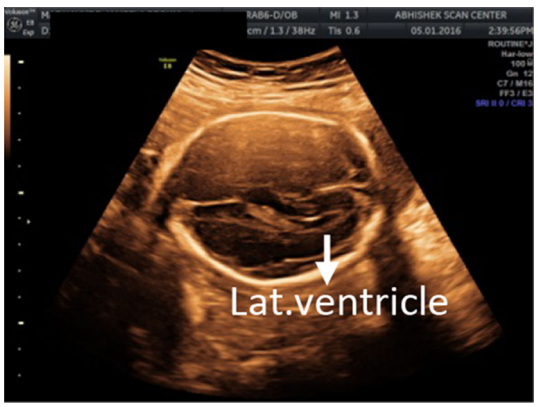

Fig. 2 Transventricular section—lateral ventricle posterior horn and choroid plexus

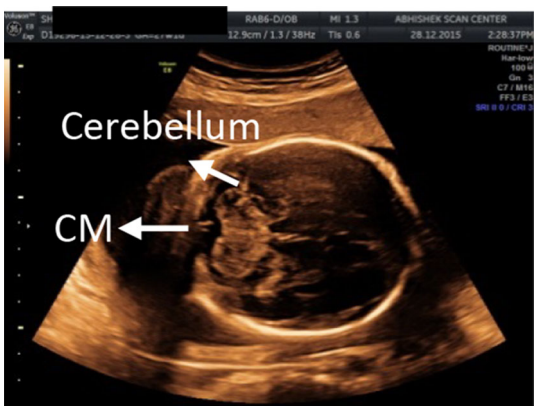

Fig. 3 Posterior fossa section—cerebellar hemispheres and cisterna magna (CM)

(Fig. 5), the vermis is seen as a triangular structure and fourth ventricle is identified as festigium. Sagittal section is useful in detailed evaluation of the vermian morphology and the biometry.

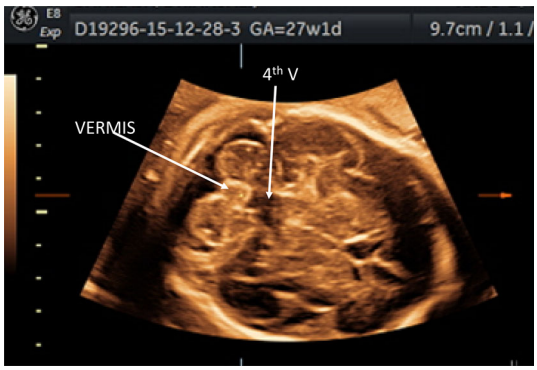

Fig. 4 Suboccipito bregmatic view—vermis and fourth ventricle

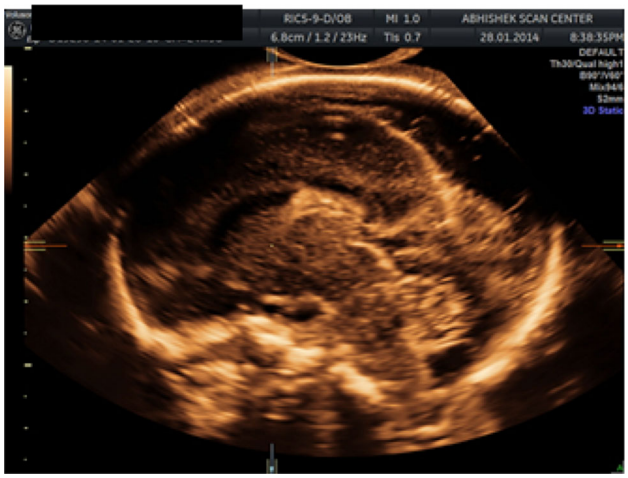

Fig. 5 Sagittal view—vermis, festigium, and tentorium

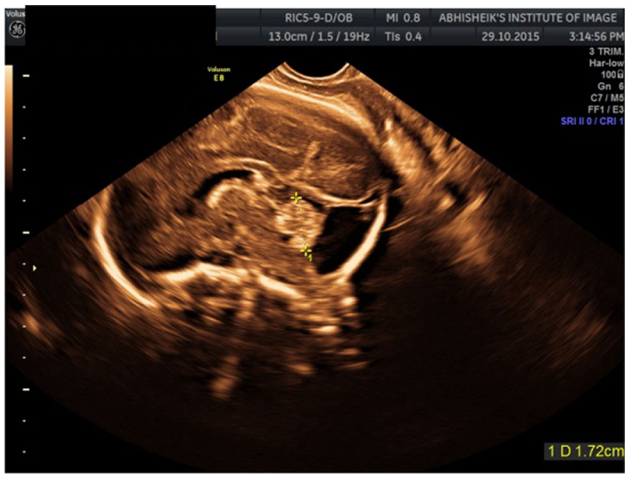

Fig. 6 Vermian morphometry—assessment of vermial morphology and biometry

Morphologically, vermis has 11 lobes but on ultrasound, three lobes are identified which are because of two fissures, namely the primary and secondary fissures (Fig. 6).

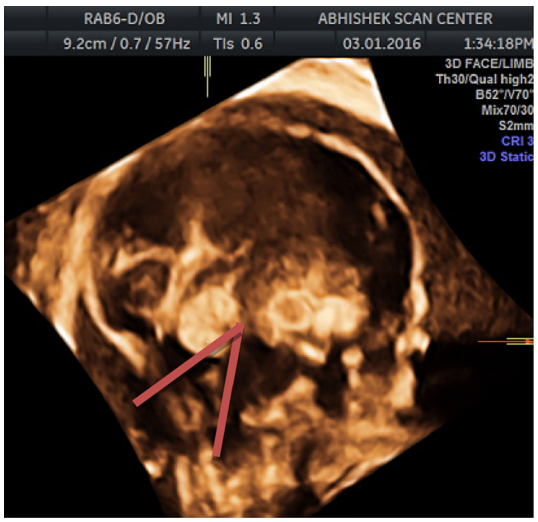

Fig. 7 Brainstem–vermis (BV) angle. Normal range of BV angle is <20°

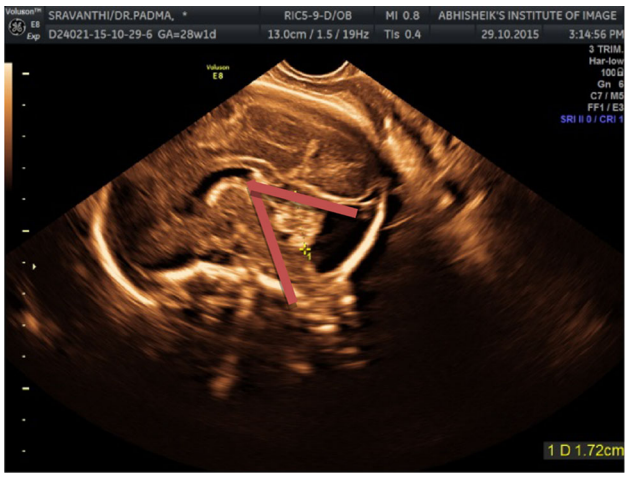

Fig. 8 Brainstem–tentorium (BT) angle. Normal range of BT angle is <45°

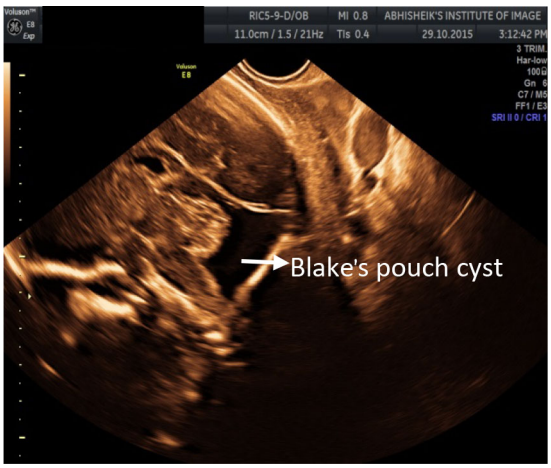

Fig. 9 Blake's pouch cyst—clear fluid and in continuation with fourth ventricle

Biometry of Vermis [3]

Superior–inferior diameter of the vermis is measured. This is dependent on gestational age ranging between 10 and 13 mm at 21–26 weeks of gestation.

Most of the posterior fossa lesions are associated with rotation of the vermis. This can be assessed subjectively or objectively. Objective assessment is done by calculating the brainstem–vermis angle (BV angle) [4] and by brainstem–tentorium angle (BT angle).

BV Angle

A line is drawn along the dorsal aspect of the brain stem and another line along the ventral aspect of cerebellar vermis (Fig. 7). Normal BV angle is <20°. In Blake's pouch cyst it is 25° and always <30°. In vermian hypoplasia, it is 30°–45°, and in Dandy–Walker malformation it is more than 45°.

BT Angle

A line is drawn along the tentorium and another along the dorsal aspect of brainstem (Fig. 8). Normal BT Angle is <45°, but in Blake's pouch cyst, the BT angle is 30°–50°, in vermian hypoplasia, it is 45°–65°, and in Dandy–Walker malformation, it is always more than 55°.

There are certain additional measurements such as tentorium—vermis angle, ratio between vermian diameter and biparietal diameter, which are rarely used.

The protocol (checklist) that is followed in evaluating posterior fossa lesions are:

1. Transcerebellar distance.
2. Depth of cisterna magna (2–10 mm).
3. Vermis—presence or absence, morphology, biometry, and rotation (BV and BT angles).
4. Size and shape of fourth ventricle.

Common Posterior Fossa Lesions

1. Blake's pouch cyst.
2. Megacisterna magna.

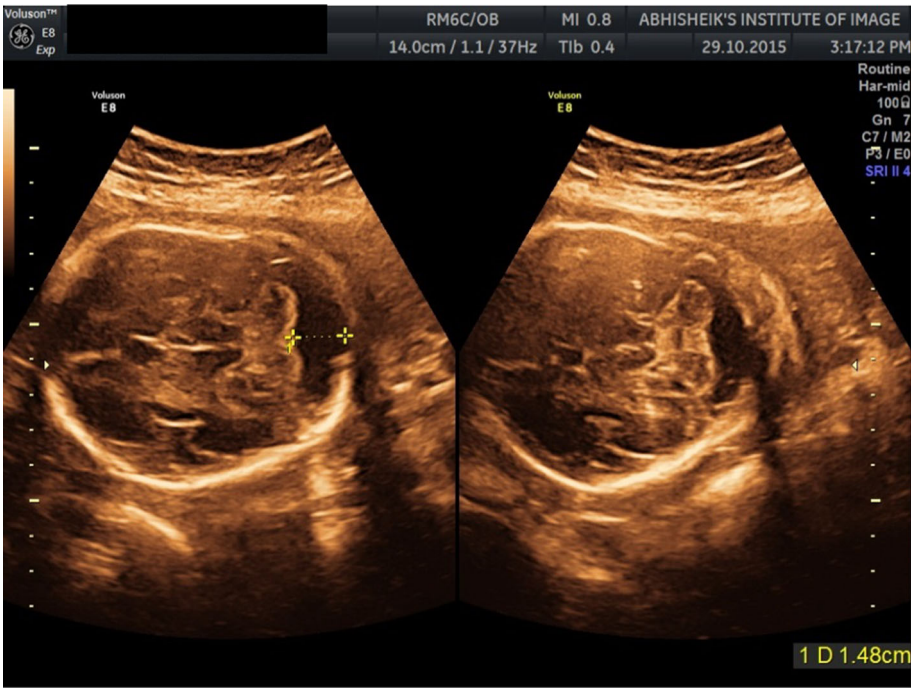

Fig. 10 Megacisterna magna—depth of cisterna magna >10 mm

3. Vermian hypoplasia.
4. Joubert syndrome (agenesis of vermis).
5. Dandy–Walker malformation.

Rare Posterior Fossa Lesions

Arachnoid Cyst, Cerebellar Hypoplasia, and Rhombencephalosynapsis

Blake's Pouch Cyst [5] (Fig. 9)

This has been described by an embryologist; hence, it is named after him. This is an evagination of area membranous inferior (AMI). Vermis develops from the area membranous superior (AMS). Blake's pouch is a finger-like projection which gets perforated by nine weeks of gestation and communicates with the cisterna magna. Blake's pouch cyst results due to persistence of finger-like projection with failure to perforate and communicate with cisterna magna. This is considered as a normal variant.

Sonographically, in sub-occipito bregmatic and sagittal view, vermis, fourth ventricle, and cisterna magna appear normal. The tentorium will be located at the normal level. BV angle is about 25°. It has to be differentiated from megacisterna magna. The fluid in the Blake's pouch cyst is clear and is in continuation with fourth ventricle, whereas the fluid in megacisterna magna is corpusculated and cisterna magna is more than 10 mm.

Megacisterna Magna [6] (Fig. 10)

Megacisterna magna is characterized when its depth is more than 10 mm with intact vermis. It may be associated with open fourth ventricle and clefting of cerebellum.

Vermian Hypoplasia [7] (Fig. 11a, b)

Axial view of the posterior fossa reveals the open fourth ventricle and normal cisterna magna. Torcular herophili or tentorium is seen at normal level but vermis appears half the size in sagittal view. Festigium and primary fissure cannot be identified. The BV angle will be between 30° and 45°.

Joubert Syndrome [8] (Fig. 12)

This condition is difficult to diagnose prenatally. This is associated with severe hypoplasia or agenesis of vermis. Extension of this syndrome is known as Joubert syndrome with related disorders (JSRD), which is characterized by polydactyly, encephalocele, and cystic kidneys, etc. Agenesis of cerebellar vermis results in close approximation of superior cerebellar peduncles resulting in molar tooth appearance. In the sagittal section, there will be bats wing or umbrella-type of fourth ventricle.

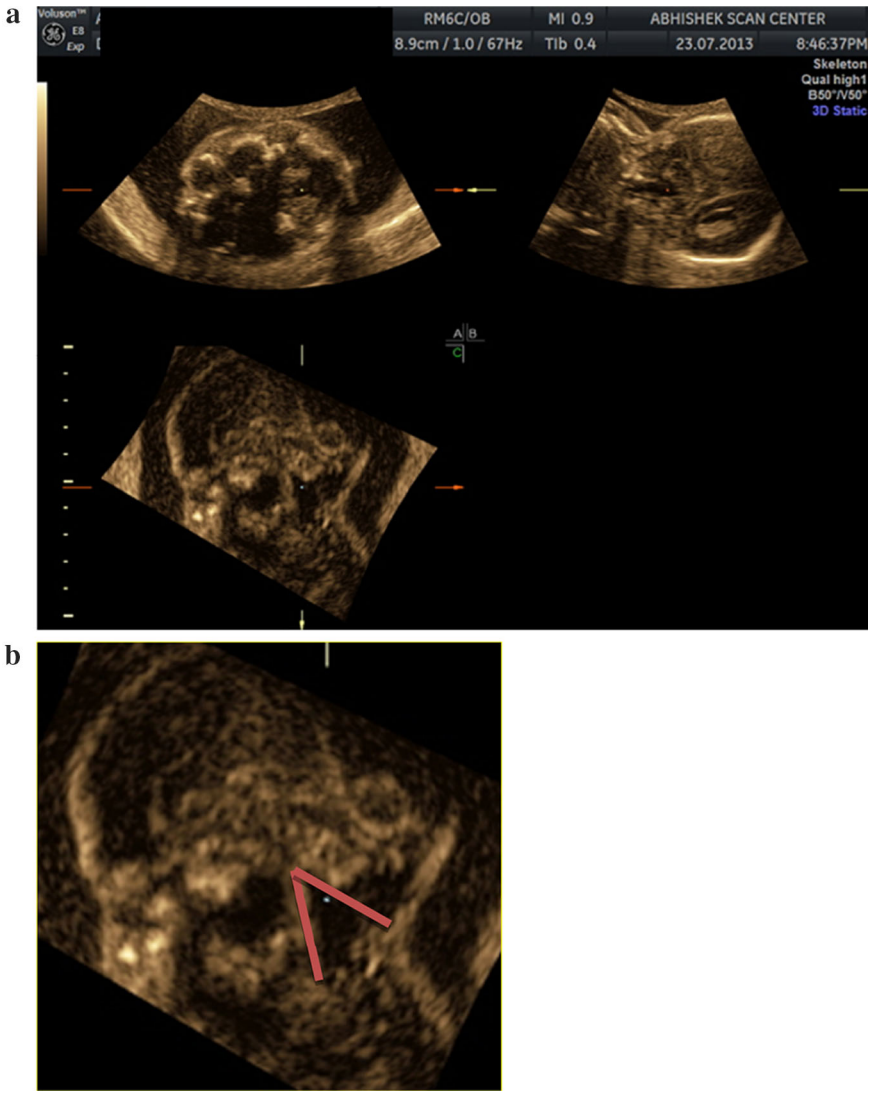

Fig. 11 **a** Vermian hypoplasia—inferior lobe of vermis small with marginal rotation and **b** vermian hypoplasia—BV angle of 30°–45°

Dandy–Walker malformation [9] (Fig. 13a–c)

It is often associated with ventriculomegaly, increased cisterna magna, and defects of cerebellar vermis. Prenatal ultrasound of the fetal cranium reveals in the axial plane, wide "V" shaped fourth ventricle. Sagittal section shows elevated tentorium, and hypoplastic rotated vermis (inferior lobe) with BV angle of more than 45°.

Uncommon Posterior Fossa Lesions

Arachnoid Cyst

It is a cystic lesion in the posterior fossa. This is eccentrically located displacing the cerebellum laterally. Other posterior fossa cystic lesion to be differentiated is Blake's pouch cyst.

Cerebellar Hypoplasia (Fig. 14)

This is an evolving abnormality. Early prenatal scan shows normal-sized cerebellum but later become hypoplastic (size less than half).

Rhombencephalosynapsis [10]

It is rare and is associated with congenital fusion of the cerebellar hemispheres and absence of the vermis.

First Trimester Evaluation of Posterior Fossa [11]

Anatomy of posterior fossa can be assessed in the 11–13.6 weeks transabdominal scan which is usually done for nuchal transculency assessment. The reference points to be considered are measurements of fourth ventricle (V), cisterna magna (CM), and the transcerebellar distance

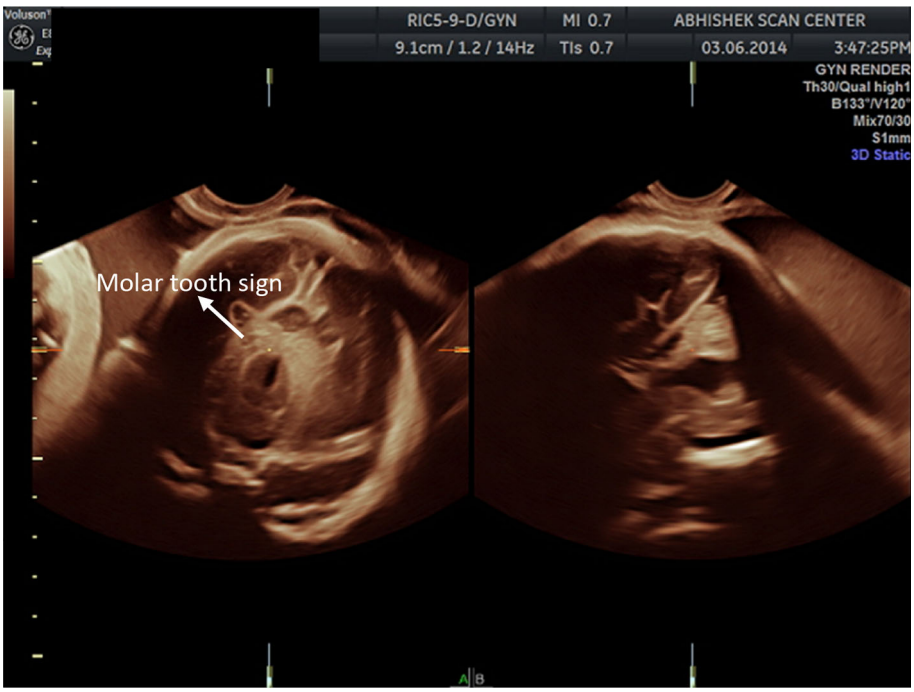

Fig. 12 Joubert syndrome—agenesis of cerebellar vermis (molar tooth sign)

(TCD). There is a linear correlation between CRL, fourth V, CM, and TCD [12] (Fig. 15a, b). Fourth ventricle cisterna magna complex can be evaluated by measuring intracranial translucency and brainstem to occipital bone diameter [13].

Prognosis

Fetal neurosonography enables to categorize and diagnose 90 % of posterior fossa lesions. Megacisterna magna and Blake's pouch cyst are common posterior fossa lesions and are rarely associated with other structural or chromosomal

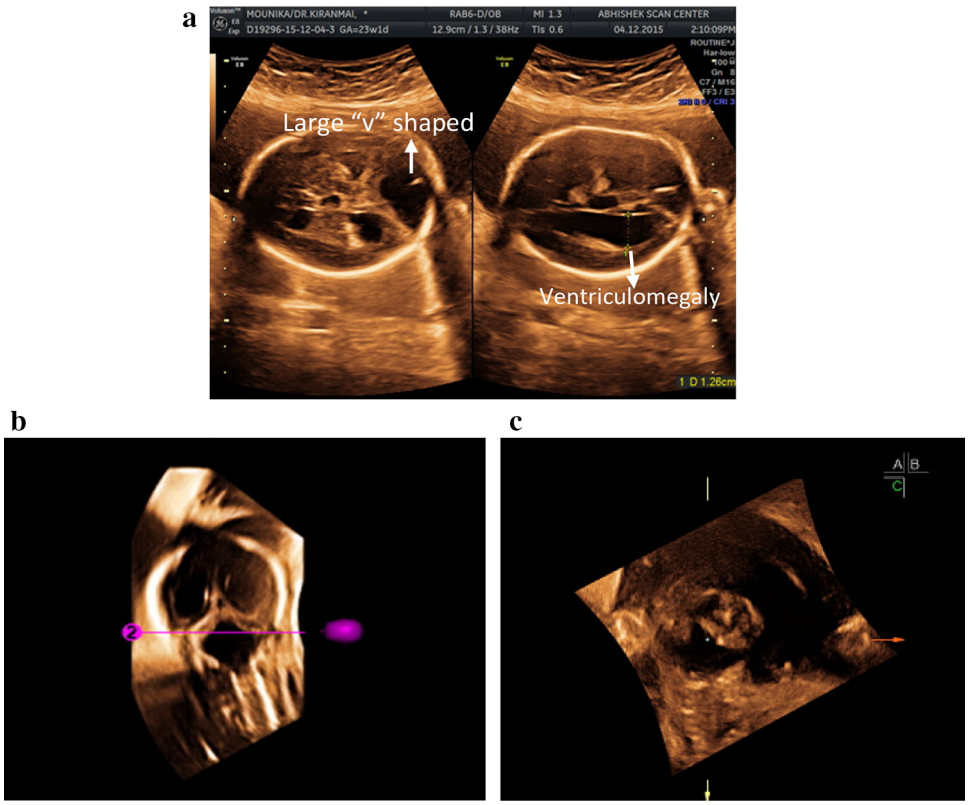

Fig. 13 **a** Dandy–Walker malformation and ventriculomegaly—large 'V' shaped cisterna magna, **b** coronal view—large cisterna magna and **c** reconstructed sagittal section—hypoplastic rotated vermis

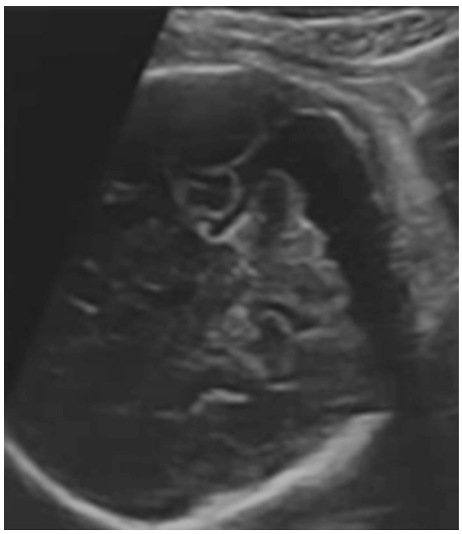

Fig. 14 Cerebellar hypoplasia—decreased intercerebellar distance with altered shape

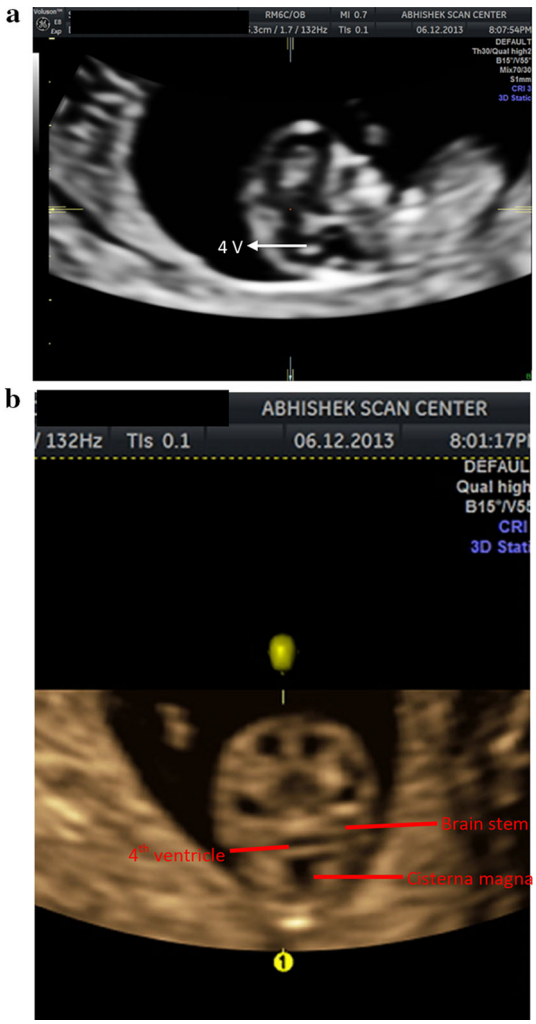

Fig. 15 **a** First trimester posterior fossa evaluation showing posterior fossa and cisterna magna. **b** Coronal view—brainstem, fourth ventricle, and cisterna magna

abnormalities. When they are isolated, there is a high possibility of spontaneous regression and normal developmental outcome. Dandy–Walker malformation and vermian hypoplasia have a guarded prognosis and are frequently associated with other abnormalities and neurologic impairment such as cognitive and psychomotor developmental delay [14].

Compliance with Ethical Standards

Conflict of interest None.

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