

Diagnosis of midline anomalies of the fetal brain with the three-dimensional median view

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ABSTRACT

Objective To investigate the effectiveness of a simplified approach to the evaluation of the midline structures of the fetal brain using three-dimensional (3D) ultrasound.

Methods Sonographic examinations were performed in normal fetuses and in cases with anomalies involving the midline cerebral structures. Two-dimensional (2D) median planes were obtained by aligning the transducer with the anterior fontanelle and midline sutures by either transabdominal or transvaginal scans. Median planes were also reconstructed using 3D ultrasonography from volumes acquired from transabdominal axial planes of the fetal head (3D median planes), by either multiplanar analysis of static volumes or volume contrast imaging in the coronal plane (VCI-C). 2D and 3D median planes were compared qualitatively and quantitatively by measuring the corpus callosum and cerebellar vermis.

Results 2D median planes could be visualized in 54/56 normal fetuses. 3D median planes were obtained in all, usually more easily and rapidly. There was a good correlation between 2D and 3D images. Measurements of the corpus callosum and cerebellar vermis were highly correlated, with mean variations of 6% and 14%, respectively. The abnormal group included 13 fetuses (five with partial or complete agenesis of the corpus callosum, six with posterior fossa malformations, two with a combination of these two anomalies). In all cases the diagnosis could be made by both 2D and 3D views and was always confirmed by postnatal investigation. Although 2D median views were of better quality, 3D images were always adequate for diagnosis, both in normal and abnormal fetuses.

Conclusions 3D median planes are obtained more easily than 2D ones, and allow an accurate diagnosis of normal cerebral anatomy and anomalies. The 3D approach may be valuable particularly for rapid assessment of fetal cerebral anatomy in standard examinations. Copyright © 2006 ISUOG. Published by John Wiley & Sons, Ltd.

INTRODUCTION

Ultrasonography is commonly employed to evaluate fetal anatomy. It is widely accepted that the results of these examinations are variable, depending mainly upon the time dedicated to the scan and the expertise of the examiner. One of the major difficulties lies in obtaining views that are not easily accessible. At midgestation, most fetuses are in a horizontal lie, and transverse sections are usually easy to obtain. These scanning planes, however, do have many limitations. While examining the fetal head, one of the most important views is probably the so-called median plane, a sagittal section of the fetal head oriented along the midline, that provides unique information on intracranial structures such as the corpus callosum and the cerebellar vermis¹. Several authors have recently advocated the use of this scanning plane in the evaluation of fetal anatomy^{2–6}. Unfortunately this scanning plane is particularly difficult to obtain. Several approaches have been described, but they all require considerable ability, time and frequently a transvaginal examination.

Three-dimensional (3D) ultrasonography is now widely available, one of the advantages of which lies in the possibility of obtaining a volume and ‘slicing’ it along directions different from the ones used to acquire it⁷. The use of 3D sonography for obtaining median views of the

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fetal brain has been previously described⁸. The purpose of our study was to evaluate the feasibility of this approach in diagnosing normal and abnormal midline structures of the fetal brain.

PATIENTS AND METHODS

The study was performed in fetuses with both normal intracranial anatomy and brain anomalies. The normal group included low-risk fetuses in the second or third trimester of gestation that were consecutively examined and were found to have a normal sonogram. The abnormal group included fetuses with anomalies whose diagnosis requires the use of the median plane, namely abnormalities of the corpus callosum and Dandy–Walker complex.

Ultrasound examinations were performed with a Voluson 730 Expert (GE Healthcare, Milan, Italy). In each case, an attempt was made to obtain a median plane of the fetal brain with two-dimensional (2D) ultrasonography (2D median plane), aligning the transducer with the anterior fontanelle and the midline sutures by either

a transabdominal approach or, when this was not possible and the position of the fetus was favorable, by a transvaginal scan. A median plane was also obtained with 3D ultrasonography (3D median plane) as previously described⁸ by two different approaches: multiplanar analysis of a static volume (Figure 1) and four-dimensional volume contrast imaging in the coronal plane (VCI-C), a technology that allows the acquisition, in almost real time, of section planes at angles different from the one of the incident ultrasound beam (Figure 2). In each case, visualization of the relevant anatomic details of the median plane, the corpus callosum and cerebellar vermis in particular, was noted for both 2D and 3D images. Measurements of the corpus callosum and cerebellar supero-inferior diameter were also obtained as previously described^{4,8}. An attempt was made to visualize the main landmarks of the cerebellar vermis, namely the fastigium of the fourth ventricle and the two main fissures^{9–11}.

Abnormalities of the corpus callosum were diagnosed using criteria previously suggested. Partial agenesis of the corpus callosum was diagnosed when it

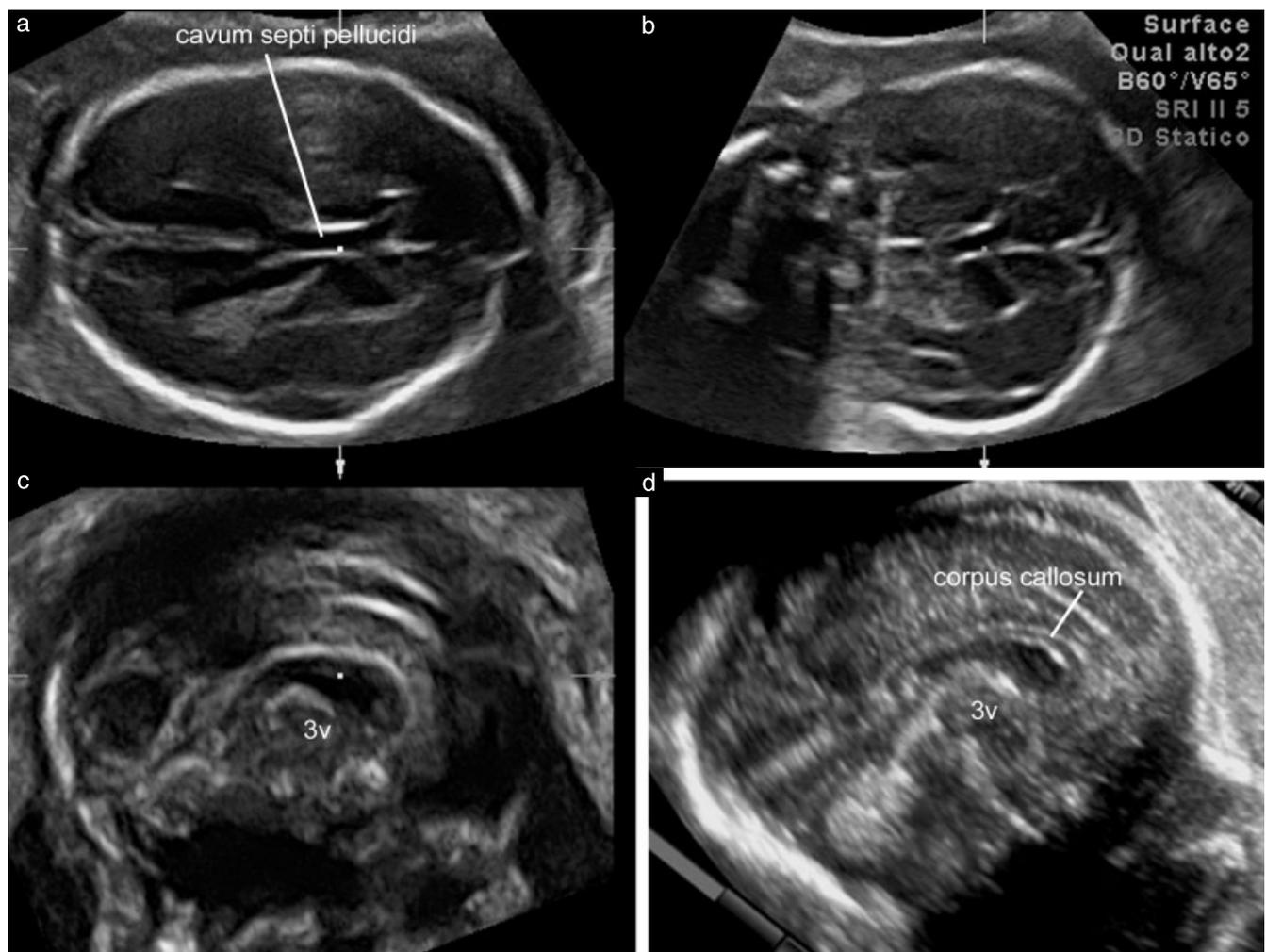


Figure 1 A comparison of three-dimensional (3D) and two-dimensional (2D) median views obtained in the same fetus. The 'start' image for the 3D examination corresponds to an axial section, parallel to the skull base and crossing the cavum septi pellucidi (a). The acquired volume is then displayed in multiplanar mode and the intersection of the planes aligned with the midline echo (a,b), to demonstrate in the sagittal plane a median view of the brain (c). The corresponding 2D image is demonstrated in (d). 3v, third ventricle.

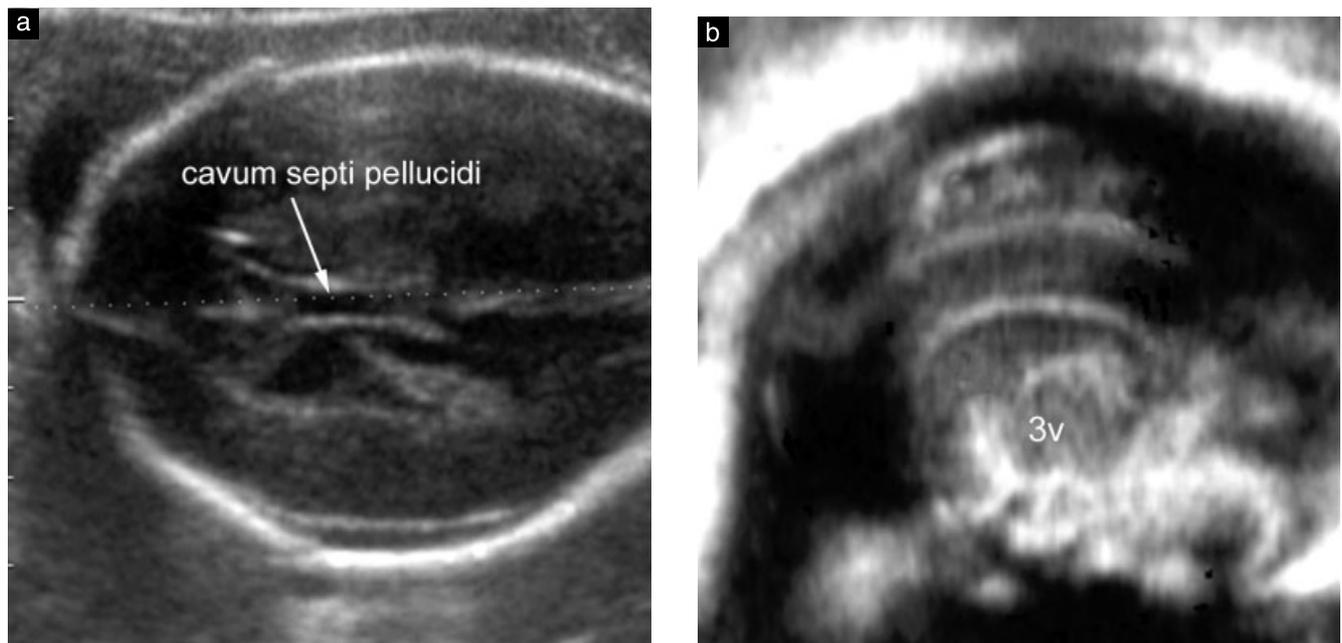


Figure 2 Three-dimensional (3D) median plane obtained with volume contrast imaging in the coronal plane (VCI-C) technology. The 'start' scan corresponds to that used for static 3D (a). The median plane is obtained by orienting the dotted line and is displayed simultaneously (b). 3v, third ventricle.

Table 1 Clinical data of fetuses with cerebral anomalies

<i>Cerebral anomaly (gestational age at diagnosis)</i>	<i>Outcome</i>
Complete agenesis of corpus callosum (21 weeks)	Termination of pregnancy
Complete agenesis of corpus callosum (22 weeks)	Termination of pregnancy
Complete agenesis of the corpus callosum with interhemispheric cyst (22 weeks)	Termination of pregnancy
Complete agenesis of corpus callosum (32 weeks)	Delivery at term. Normal neurologic examination at birth
Partial agenesis of the corpus callosum, agenesis of septum pellucidum (22 weeks)	Termination of pregnancy
Partial agenesis of the corpus callosum with hypoplasia of cerebellar vermis (22 weeks)	Delivery at term. Delayed neurologic development at 1 year of age
Partial agenesis of the corpus callosum with hypoplasia of cerebellar vermis (22 weeks)	Termination of pregnancy
Blake's pouch cyst (23 weeks)	Delivery at term, normal at 18-month follow-up
Blake's pouch cyst (22 weeks)	Delivery at 25 weeks, normal development at 6 months of age
Blake's pouch cyst (22 weeks)	Delivery at 34 weeks, dysmorphic syndrome to be defined
Blake's pouch cyst (27 weeks)	Delivery at 36 weeks, normal neurologic examination at birth
Megacisterna magna (30 weeks)	Delivery at term, normal development at 6 months
Megacisterna magna (25 weeks)	Delivery at term, normal neurologic examination at birth

appeared subjectively to be small and incomplete¹². Measurements were also taken and compared with published nomograms^{4,8}. The Dandy–Walker complex was categorized following the indications of Adamsbaum and co-workers⁹. Position and morphology of the cerebellar vermis were noted in each case. Rotation of the vermis was diagnosed when a fluid-filled space was seen dislocating the vermis from the brain stem. Hypoplasia was assessed subjectively, and the diagnosis was further supported when the main landmarks of the cerebellum (fastigium and two fissures) could not be identified^{9–11}. Measurements of the cerebellum were also obtained and compared with published nomograms. A detailed follow-up was obtained in all abnormal cases (Table 1).

RESULTS

Normal fetuses

Fifty-six cases with mean gestational age 24.3 ± 4.9 (range, 19–35) weeks were examined. The 2D median plane could be obtained in 54 (96%). Transabdominal sonography allowed adequate visualization in 43, while a transvaginal scan was required in the remaining 11 cases.

3D median planes obtained from multiplanar evaluation of static volumes and those obtained with VCI-C technology were similar (Figures 1 and 2) and both correlated well with 2D images. The 2D views were of superior quality, in that they always allowed identification of the corpus callosum as a thin sonolucent strip

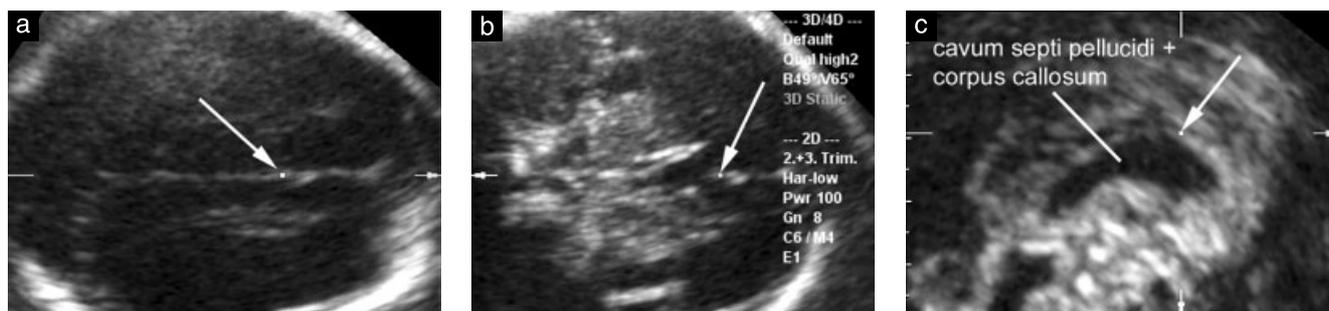


Figure 3 Multiplanar analysis of an ultrasound volume of the fetal brain obtained from the transverse plane, as in Figures 1 and 2. A comparison of the position of the dot (arrows) indicating the intersection of the three orthogonal planes identifies the superior echogenic border of the cavum septi pellucidi–corpus callosum complex as the lower edge of the midline echo.

with well defined echogenic contours overlying the cavum septi pellucidi. In 3D median planes the corpus callosum could not be clearly differentiated from the inferior cavum septi pellucidi (Figures 1 and 2). A single comma-shaped sonolucent structure was seen, outlined superiorly by an echogenic line that in multiplanar analysis could be identified as the lower extremity of the midline echo (Figure 3).

Measurements of the corpus callosum in 2D and 3D images correlated well (Figure 4). The mean difference was 1.6 mm or 6%, with an SD of 1 mm and 3%, respectively.

Visualization of the posterior fossa in 3D median planes was hampered by the acoustic shadowing of the petrous ridges of the base of the skull (Figure 5). Such shadow obscured the brain stem, frequently preventing the assessment of the normal relationship with the cerebellar vermis. The cerebellar vermis, outlined by the

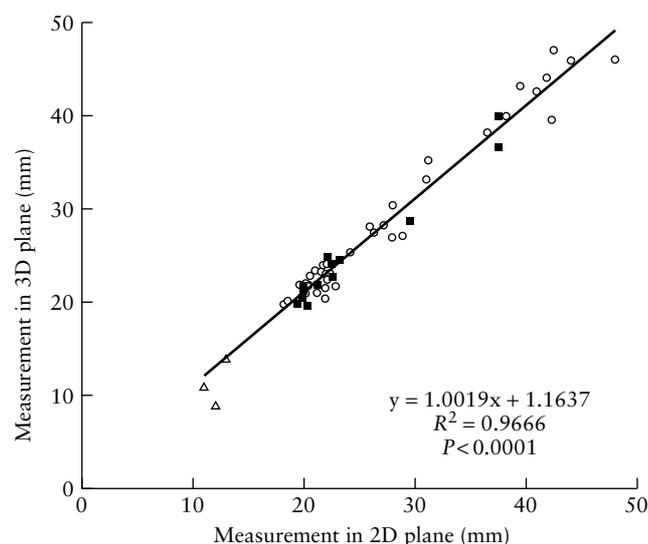


Figure 4 A comparison of measurements of the length of the corpus callosum in two-dimensional (2D) vs. three-dimensional (3D) median planes. Open circles indicate measurements in normal fetuses with static 3D sonography. Black squares indicate measurements with volume contrast imaging in the coronal plane (VCI-C). Triangles indicate three fetuses with partial agenesis of the corpus callosum, in which the 3D plane was always obtained from static volumes.

subarachnoid space, could be seen in all cases but the fourth ventricle and the two main fissures were clearly visualized in only 26/56 cases (46%), vs. 42/54 (78%) in 2D median planes. Measurements of the supero-inferior diameter of the cerebellar vermis obtained with 2D and 3D ultrasonography correlated well (Figure 6). The mean difference was 1.9 mm or 14%, with an SD of 1.5 mm or 15%, respectively.

Fetuses with brain malformations

The abnormal group included 13 fetuses ranging in gestational age from 21 to 32 weeks (Table 1). 3D median planes were comparable to 2D views and had diagnostic quality in all cases. Abnormalities of the corpus callosum were in general better demonstrated (Figures 7 and 8). A precise diagnosis of posterior fossa anomalies was also possible (Figures 9 and 10). At times, however, the relationship between the brain stem and the cerebellar vermis could not be clearly appreciated owing to shadowing artifacts. Furthermore, the integrity of the vermis had to be assessed mostly on a quantitative basis, as the main anatomic landmarks, the fastigium of the fourth ventricle and the main fissures, could not be demonstrated (Figure 11). Measurements of the corpus callosum in fetuses with partial agenesis and measurements of the cerebellum in cases with Dandy–Walker complex obtained from 2D and 3D images correlated well. The length of the corpus callosum in the three cases with partial agenesis was always well below the 5th centile⁴. In the two fetuses with Dandy–Walker complex and subjective diagnosis of vermian hypoplasia the supero-inferior cerebellar diameter was -2.1 and -1.5 SD from the mean. In the remaining cases, it fell within normal limits⁸.

The prenatal diagnoses were always confirmed postnatally by either autopsy or magnetic resonance imaging. Overall, four fetuses were terminated and the diagnosis was confirmed by anatomic dissection. One fetus with complete agenesis of the corpus callosum, three with Blake's pouch cyst, and two with megacisterna magna continued the pregnancy, were delivered at term and are alive and well at the time of writing. Although they seem to be developing normally, the follow-up is too short to

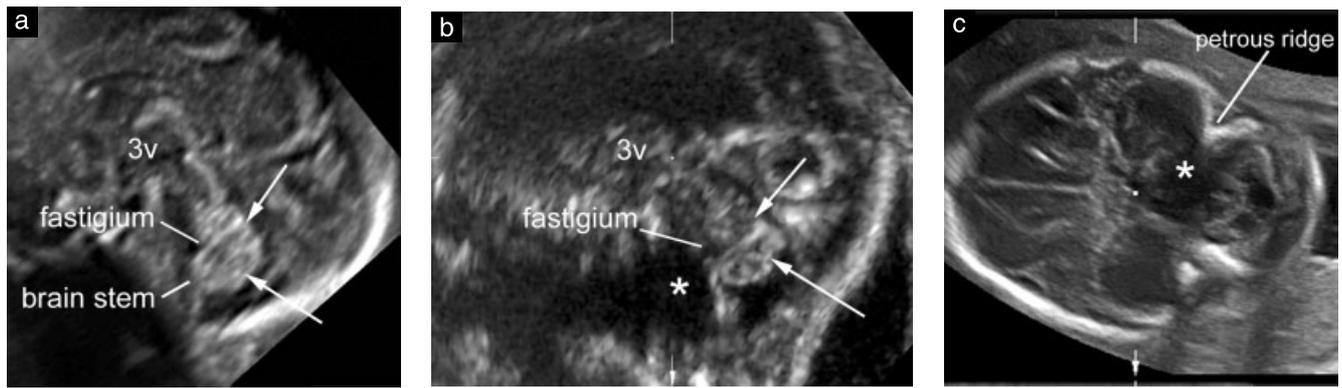


Figure 5 Views of the posterior fossa in two-dimensional (2D) (a) and three-dimensional (3D) median views (b). In the latter the cerebellar vermis is visible, while the brain stem is partly obscured by shadowing (*) from the petrous ridges of the temporal bone (c). In both (a) and (b) the three main landmarks of the cerebellar vermis, the fastigium and the two main fissures (arrows) are identified. 3v, third ventricle.

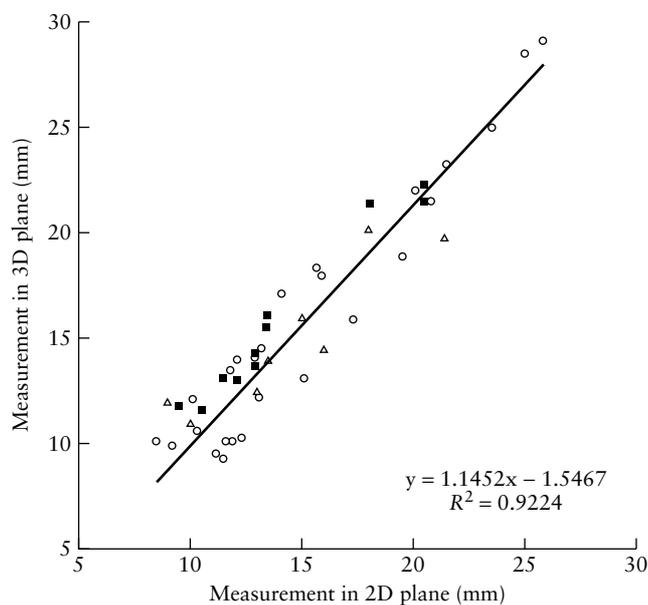


Figure 6 A comparison of measurements of the supero-inferior diameter of the cerebellar vermis in two-dimensional (2D) vs. three-dimensional (3D) median planes. Open circles indicate measurements in normal fetuses with static 3D sonography. Black squares indicate measurements with volume contrast imaging in the coronal plane (VCI-C). Triangles indicate eight fetuses with Dandy–Walker complex, in which the 3D plane was always obtained from static volumes.

allow a proper neurologic evaluation (Table 1). One fetus with Blake's pouch cyst is affected by a severe dysmorphic syndrome that is still awaiting a definitive diagnosis. One fetus with combined hypoplasia of the corpus callosum and cerebellar vermis is affected at 1 year of age by neurodevelopmental retardation that is likely to be severe.

DISCUSSION

Our study suggests that median views of the fetal brain reconstructed from sonographic volumes acquired originally from the transverse plane have diagnostic quality in the assessment of normal and abnormal

anatomy of the corpus callosum and cerebellum. Median views obtained directly with 2D ultrasonography are of superior quality; 3D sonography does not overcome the physical limitations of the technique. The thin sonolucent corpus callosum is not visualized when the ultrasound beam is oriented along the transverse plane of the fetal head. However our results indicate that it can be precisely inferred from the sonolucent complex it forms in association with the inferior cavum septi pellucidi. Acoustic shadowing from the base of the skull obscures the brain stem and at times does not allow precise assessment of the relationship between this and the cerebellar vermis. Furthermore, the landmarks of the cerebellar vermis are frequently not clearly demonstrated. Nevertheless, despite these limitations, all the cerebral anomalies that were encountered in our study were precisely diagnosed. It needs to be stressed that the categorization of abnormalities of the posterior fossa remains controversial, and the clinical implications of the different entities that are encountered are very uncertain. However, in our experience both 2D and 3D sonography proved correct in the definition of the anatomic findings.

To our knowledge this is the first time that a quantitative approach has been applied to the diagnosis of anomalies of the corpus callosum and of the cerebellar vermis. We have found that fetuses with partial agenesis of the corpus callosum had measurements well below the normal range. Fetuses with Dandy–Walker complex and megacisterna magna that were found postnatally to have a normally developed cerebellum had measurements of the vermis within normal limits. Of the two fetuses with hypoplastic vermis, one had a measurement below the normal limits and one was at the lower limit of normal. It must be stressed that the prenatal diagnosis of hypoplasia of the cerebellar vermis that correlates with abnormal neurodevelopment in postnatal studies is difficult and there is a lack of consensus on the optimal approach^{10,11}. Albeit limited, our data support the concept that biometry may be useful in supporting the qualitative examination for recognizing partial abnormalities of the corpus callosum and cerebellar vermis.

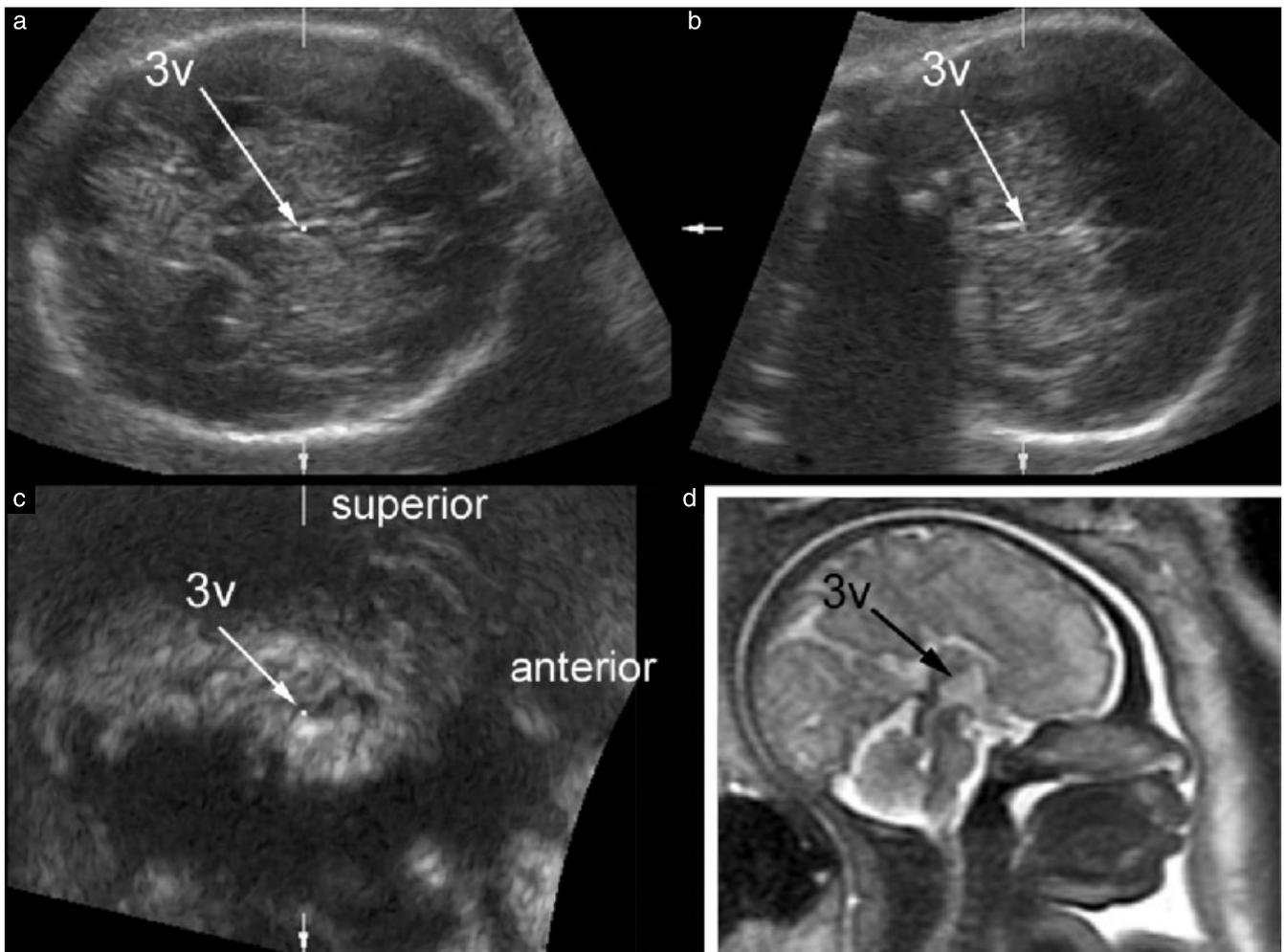


Figure 7 Complete agenesis of the corpus callosum in a third-trimester fetus examined with transabdominal three-dimensional (3D) ultrasound in multiplanar mode (a–c). The dot (arrow) corresponding to the intersection of the three orthogonal planes is positioned at the level of the third ventricle (3v), and the 3D median plane (c) demonstrates that above it the cavum septi pellucidi–corpus callosum complex is absent. A magnetic resonance image obtained in the same fetus confirms the diagnosis (d).



Figure 8 Partial agenesis of the corpus callosum. The small cavum septi pellucidi–corpus callosum is similarly demonstrated in the transvaginal two-dimensional (2D) median plane (a) and the transabdominal three-dimensional (3D) view (b). The same measurement of the hypoplastic corpus callosum (1.1 mm) is obtained in both the 2D and 3D images and correlates well with the autopsy specimen (c). 3v, third ventricle.

Multiplanar analysis of static volumes and VCI-C technology were equally effective in reconstructing median planes from transverse scans. VCI-C allows

elaboration of the median plane in almost real time, thus certainly minimizing movement artifacts. However, a static volume of medium quality is very rapidly obtained

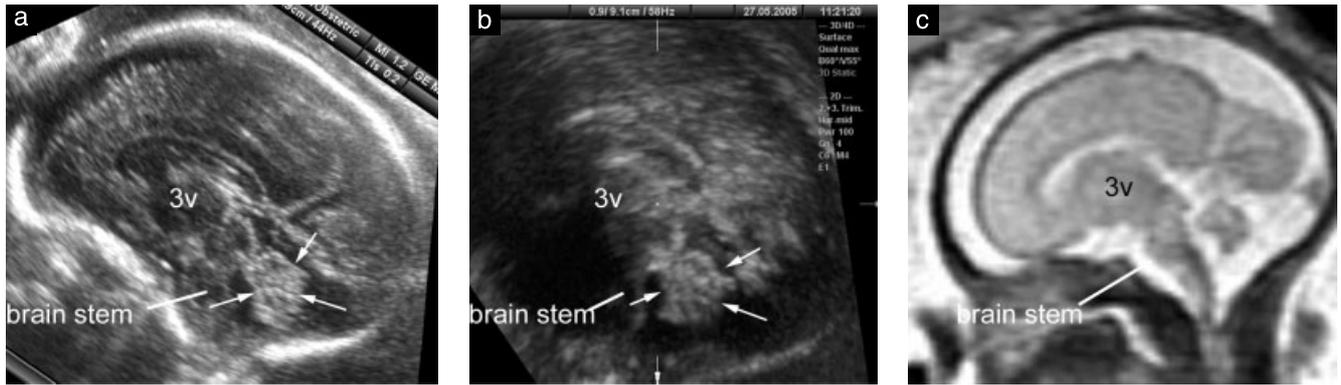


Figure 9 Fetus at 27 weeks' gestation with Blake's pouch cyst, the most frequent posterior fossa abnormality encountered in this study. The transabdominal two-dimensional (2D) (a) and three-dimensional (3D) (b) median planes demonstrate the superior rotation of a seemingly intact cerebellar vermis (the arrows point to the main anatomic landmarks, the fastigium of the fourth ventricle and main fissures), in the presence of a minimally dilated cisterna magna. A magnetic resonance image oriented along the same axis (c) is provided for comparison. 3v, third ventricle.

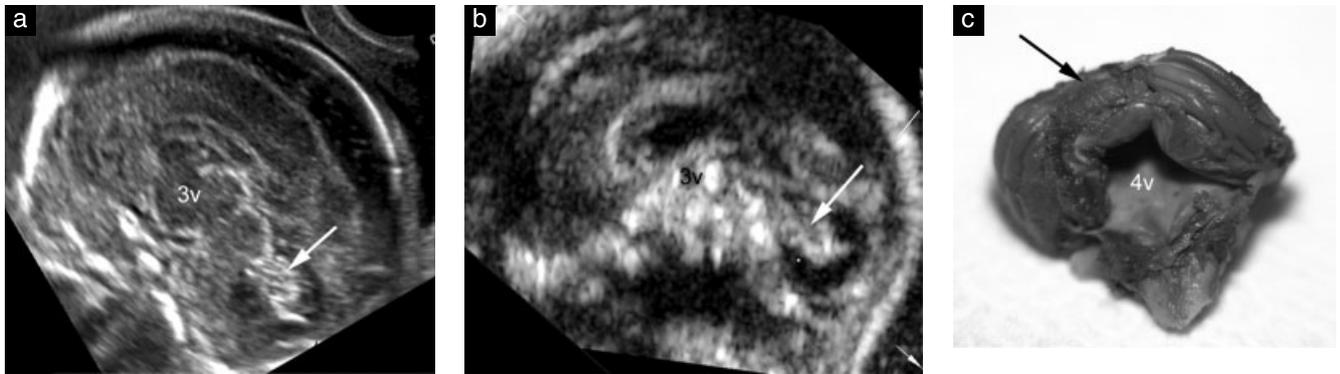


Figure 10 Enlargement of the cisterna magna with cerebellar rotation and hypoplasia of the vermis at 22 weeks' gestation. Both the two-dimensional (2D) (a) and the three-dimensional (3D) (b) median planes demonstrate a vermis (arrows) that appears small (the supero-inferior diameter is 10 mm, which is smaller than expected at 22 weeks' gestation). Failure to visualize the anatomic landmarks of the vermis adds to the index of suspicion. Autopsy (c) confirmed a prominent aditus to the fourth ventricle (4v) with hypoplasia of the vermis (arrow), which demonstrated absence of lobulation, and a generalized reduction in size of the entire cerebellum. 3v, third ventricle.

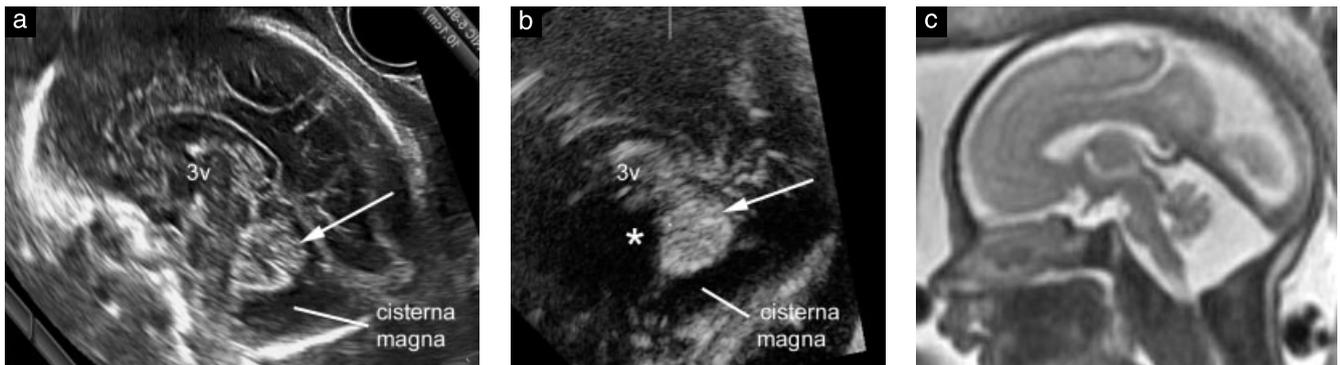


Figure 11 Megacisterna magna at 27 weeks' gestation; two-dimensional (a) and three-dimensional (3D) (b) images. Although the 3D median plane demonstrates an increased size of the cisterna magna and a seemingly intact vermis (arrow), shadowing obscures the brain stem (*), and the landmarks of the vermis are not clearly demonstrated. Magnetic resonance imaging confirms the diagnosis (c). 3v, third ventricle.

and has the advantage of allowing navigation in the three orthogonal planes, which may be useful at times, particularly when dealing with complex anatomy. A careful comparison of the sagittal and coronal planes is required to assess the integrity of the cerebellar vermis, and we have found that the simultaneous representation of the posterior fossa in the three

orthogonal planes is of particular value in this regard (Figure 12).

CONCLUSIONS

We believe that our results are valuable. Obtaining median views with standard 2D ultrasonography is difficult and

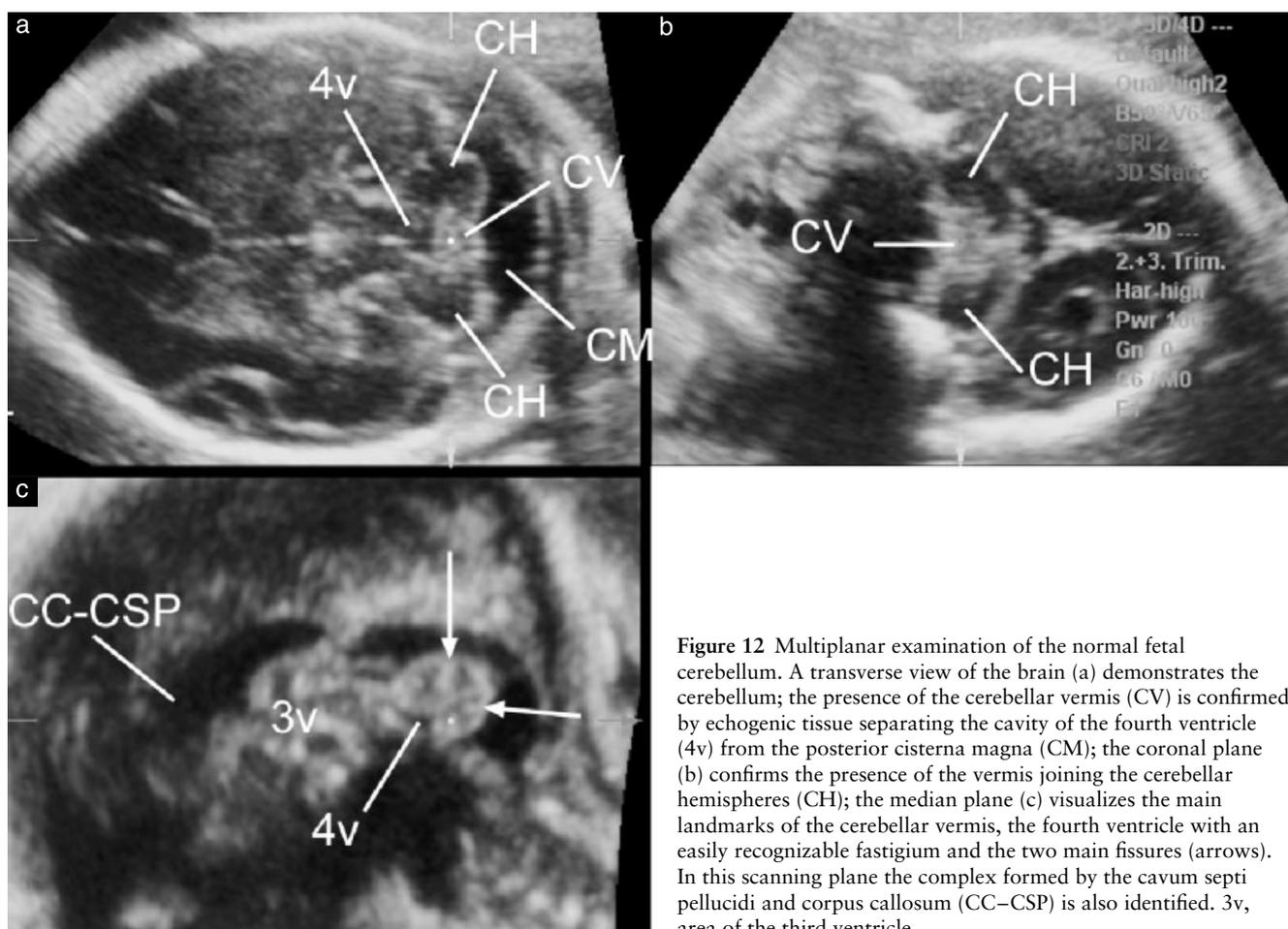


Figure 12 Multiplanar examination of the normal fetal cerebellum. A transverse view of the brain (a) demonstrates the cerebellum; the presence of the cerebellar vermis (CV) is confirmed by echogenic tissue separating the cavity of the fourth ventricle (4v) from the posterior cisterna magna (CM); the coronal plane (b) confirms the presence of the vermis joining the cerebellar hemispheres (CH); the median plane (c) visualizes the main landmarks of the cerebellar vermis, the fourth ventricle with an easily recognizable fastigium and the two main fissures (arrows). In this scanning plane the complex formed by the cavum septi pellucidi and corpus callosum (CC–CSP) is also identified. 3v, area of the third ventricle.

requires time. Conversely, the 3D approach is easily and rapidly performed. We expect that expert sonologists will continue to obtain median views with standard 2D ultrasonography as these allow sharper resolution of anatomic details, which may be critical, particularly when dealing with abnormal cases. One major limitation of 3D median planes is the impossibility of visualizing the brain stem, which plays an important role in assessing the severity of intracranial anomalies in general and posterior fossa anomalies in particular⁵. However, we suggest that the reconstructed median plane may have a role in the rapid assessment of normal anatomy in standard examinations, as well as in those abnormal cases in which a direct 2D scan cannot be obtained.

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