Ultrasound Obstet Gynecol 2006; **27**: 490–493
Published online in Wiley InterScience (www.interscience.wiley.com). **DOI:** 10.1002/uog.2768

Rotation of the vermis as a cause of enlarged cisterna magna on prenatal imaging

Y. ZALEL*, Y. GILBOA*, L. GABIS†, L. BEN-SIRA‡, C. HOFFMAN§, Y. WIENER* and R. ACHIRON*

*Departments of *Obstetrics & Gynecology, †Pediatric Neurology and ‡Pediatric Radiology, The Chaim-Sheba Medical Center, Tel-Hashomer and §Pediatric Radiology, Sorsky Medical Center (Affiliated to the Sackler School of Medicine, Tel-Aviv University), Ramat-Gan, Israel*

KEYWORDS: cerebellar vermis; cisterna magna; prenatal sonography; rotation; sagittal plane

ABSTRACT

Objectives *Dandy–Walker complex is a continuum of developmental anomalies of the posterior fossa which includes vermian rotation. However, vermian rotation alone may be benign. The aim of this study was to describe our experience with sagittal-plane prenatal ultrasound in the diagnosis of rotation of the vermis in cases of suspected enlarged cisterna magna on routine antenatal imaging, and to describe the follow-up of these patients.*

Methods *Seven women, who were referred to our ultrasound unit for evaluation of an enlarged fetal cisterna magna and suspected agenesis of the vermis on axial-plane imaging, underwent further multiplanar studies of the posterior fossa and measurements of the vermis.*

Results *The mean maternal age was 27 (range, 20–33) years and the mean gestational age at diagnosis was 19.5 (range, 18–31) weeks. The standard axial plane image showed a 'direct communication' between the cisterna magna and the fourth ventricle. In the mid-sagittal plane, the vermis was clearly delineated, with posterosuperior rotation. Vermis size was within normal limits for gestational age in all cases. Findings were confirmed by prenatal magnetic resonance imaging (MRI) in two cases and postnatal MRI and/or sonography in five. During a mean follow-up of 4.5 (range, 1–7.5) years, all children developed normally, with no neurological complications.*

Conclusion *The finding of an enlarged cisterna magna on standard- (axial-)plane ultrasound should be evaluated further in the sagittal plane to determine whether the cause is rotation of a normal vermis. This may spare patients unnecessary tests, anxiety and, in some cases, pregnancy termination.*

INTRODUCTION

Dandy–Walker malformation is a rare congenital syndrome. Originally it was characterized as a combination of ventriculomegaly, large cisterna magna, and defective cerebellar vermis. Different definitions are used, including dysgenesis of the cerebellar vermis, cystic dilation of the fourth ventricle, and a range of abnormalities of the posterior fossa, with upward displacement of the tentorium[1,2]. It is distinguishable from megacisterna magna, defined as a cisterna magna > 10 mm in diameter, and Dandy–Walker variant, which consists of vermian agenesis and cystic dilation of the fourth ventricle without enlargement of the posterior fossa[3].

Dandy–Walker malformation has an estimated prevalence of 1 : 30 000 births. It manifests clinically in the first year of postnatal life as hydrocephalus, accounting for 4–12% of all cases of hydrocephalus in infants. The mortality rate is 24% and about 40–70% of survivors have subnormal intelligence. The clinical significance of megacisterna magna and Dandy–Walker variant is unknown[4]; they are apparently associated with neurological compromise, but no clear-cut prognostic data exist[3]. Hence, the specific diagnosis is of utmost importance.

In 1989, Barkovich *et al.*[5], using postnatal magnetic resonance imaging (MRI), reported that Dandy–Walker malformation, Dandy–Walker variant and megacisterna magna actually represent a continuum of developmental anomalies of the posterior fossa. In 12 patients, they noted an enlarged cisterna magna with no intervening vermis

Correspondence to: Prof. Y. Zalel, Obstetrics & Gynecologic Dept., Sheba Medical Center, Tel-Hashomer (Affiliated to the Sackler School of Medicine, Tel-Aviv University), Ramat-Gan, 52621, Israel (e-mail: zalel_y@netvision.net.il, zalel_y@a2z.net.il)

Accepted: 8 February 2006

linking it to the fourth ventricle. On sagittal imaging, however, the vermis appeared severely rotated superiorly and slightly hypoplastic, leading them to suggest, for the first time, that vermian rotation, a benign condition, could cause apparent enlargement of the posterior fossa. Klein *et al*.[6] also described rotation of the vermis using MRI in a series of cases of Dandy–Walker malformation. In all cases, the vermis was either partially agenetic or severely malformed and the volume of the posterior fossa was enlarged, with ascent of the cerebellar tentorium and torcular. Guibaud[7] also used MRI to describe horizontalization of the vermis in cases of retrocerebellar cystic formation. Vermian rotation, however, has never been confirmed by ultrasound during the prenatal period.

The aim of this study was to describe the use of ultrasound in the sagittal plane to detect rotation of the vermis in fetuses with suspected enlargement of the cisterna magna.

METHODS

Seven women were referred to the ultrasound unit of a major tertiary center, between the years 1995 and 2004, for evaluation of an enlarged fetal cisterna magna and possible vermian agenesis detected on routine antenatal ultrasound examination. No other brain or extra-central nervous system malformations were noted.

In order to visualize the posterior fossa and the vermis, we performed sonography in the transverse (axial), sagittal and coronal planes. The vermis was evaluated in the sagittal plane for position and structure, and its height and width were measured, as described previously[8,9]. The size of the cisterna magna, and the appearance of the fourth ventricle and brain stem were also evaluated. Fetal karyotyping was performed. All women were offered consultations with a geneticist and a pediatric neurologist.

The structure of the vermis was further evaluated prenatally by MRI in two cases and postnatally with MRI in one case, based upon the neurologist's decision, and ultrasound in four cases. All children were examined by a pediatric neurologist at birth, and were followed up in the pediatric neurology clinic and/or by a pediatrician.

RESULTS

The mean age of the women was 27 (range, 20–33) years and the mean gestational age at diagnosis was 19.5 (range, 18–31) weeks. In the axial plane, there appeared to be a direct communication between the cisterna magna and the fourth ventricle (Figure 1). The cisterna magna measured 10–15 (mean, 11) mm in diameter, indicating possible vermian agenesis. In the sagittal plane, however, the normal structure of the vermis could be very well delineated (Figure 2), and measurements were appropriate for gestational age[8,9]. Amniocentesis revealed a normal karyotype in all cases.

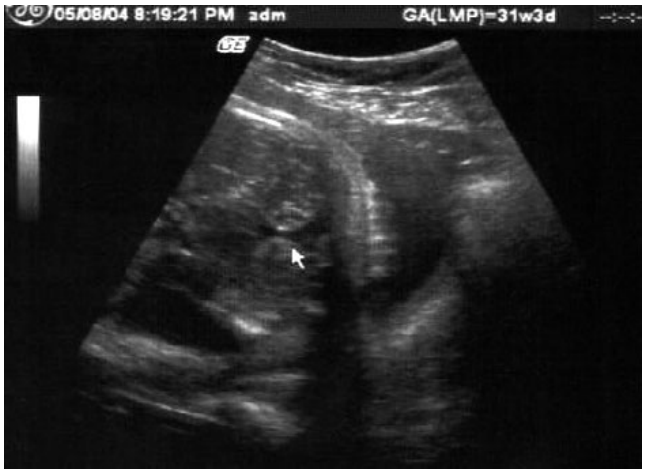

Figure 1 Axial ultrasound image showing the posterior fossa at 31 weeks' gestation. Note the apparent enlarged cisterna magna and direct communication between the cisterna magna and fourth ventricle (arrow).

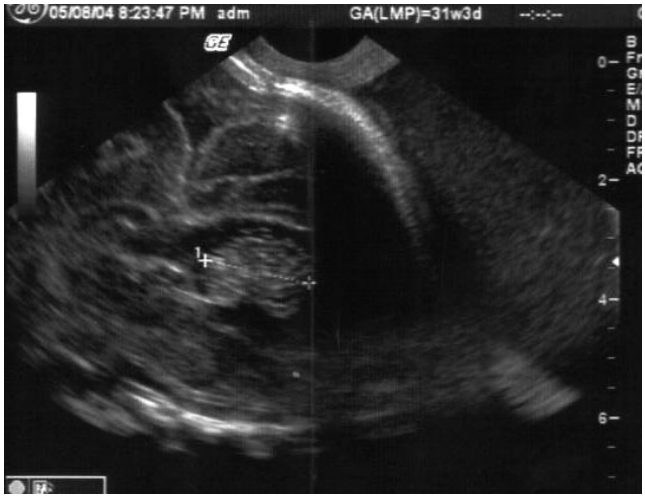

Figure 2 Oblique sagittal section of the same patient as in Figure 1, showing the intact and rotated normal vermis.

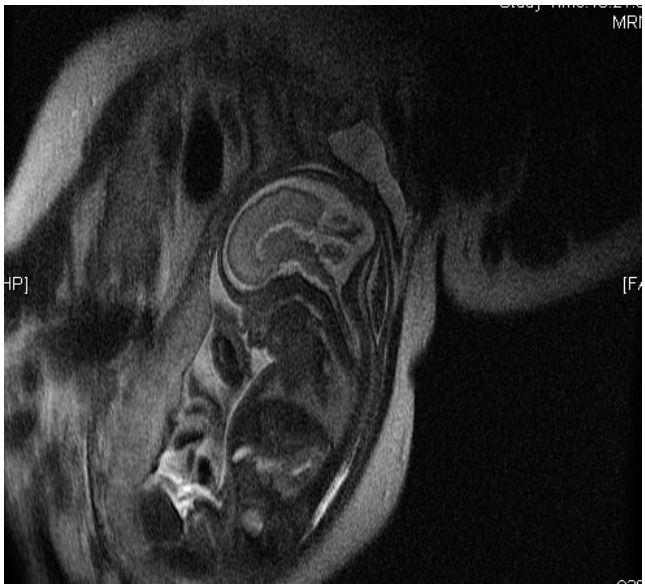

Figure 3 Magnetic resonance image of a fetus at 28 weeks' gestation demonstrating rotation of an apparently normal vermis.

Table 1 Background and clinical data of seven cases of fetal vermian rotation

Gestational age (weeks) at diagnosis	*Gender*	*Diameter of CM (mm)*	*Additional findings*	*Additional work-up (genetic/radiological)*	*Length of follow-up (years)**
31	Female	12	Asymmetry of ventricles	Amniocentesis, neonatal sonography	1
20	Female	10	Intrauterine growth restriction	Amniocentesis, MRI	5
18	Male	11	None	Amniocentesis, neonatal MRI	7.5
22	Male	15	None	Amniocentesis, neonatal sonography	7
24	Male	11	Heterogeneous choroid plexus	Amniocentesis, MRI	1
18	Female	11	None	Amniocentesis, neonatal sonography	7
18	Female	10	None	Amniocentesis, neonatal sonography	3

*None of the infants had abnormal neurological findings on follow-up. CM, cisterna magna; MRI, magnetic resonance imaging.

The prenatal MRI (Figure 3) and postnatal MRI and ultrasound examinations confirmed the prenatal sonographic findings (Table 1). During a mean follow-up of 4.5 (range, 1–7.5) years, all children achieved the normal developmental milestones, with no neurological complications.

DISCUSSION

Imaging of the posterior fossa has become an integral part of screening for fetal anomalies, and the sonographic evaluation of the cerebellum, vermis and cisterna magna has proven to be of value in the detection of several malformations.

Vermian agenesis is commonly found in Dandy–Walker malformation and Dandy–Walker variant, as well as in association with other malformations and syndromes[10,11]. It may be complete or partial (i.e. inferior)[12,13]. Embryogenesis of the cerebellar vermis begins rostrally in the midline during the 9th gestational week. The fusion process continues caudally to form the remainder of the cerebellar hemispheres (neocerebellum)[14,15]. Only by 16–18 weeks' gestation is the fourth ventricle completely covered by the cerebellar hemispheres and vermis, and by 18 weeks the communication between the fourth ventricle and the cisterna magna is covered as well. Therefore, the diagnosis of vermian agenesis (especially partial agenesis) cannot be made prior to 18 weeks' gestation[16,17]. Even after this period, sagittal-plane sonography cannot detect subtle defects in the cerebellum and/or vermis. Pilu *et al.*[18] reported that, in their experience, the vast majority of fetuses with apparently isolated enlargement of the cisterna magna or a small vermian defect were found on clinical and neuroimaging evaluation at birth to be entirely normal. They also suggested that prenatally it may be impossible to diagnose correctly in all cases a defect in the cerebellar vermis[19].

At present, during standard antenatal sonography, measurements of the cisterna magna are made in the axial plane (Figure 1). This plane does not, however, have the capability of clearly assessing the integrity of the vermis and can therefore lead to over- or underestimation of any defect. Prompted by the study of Barkovich *et al.*[5], wherein the excellent resolution in the sagittal plane afforded by MRI led to a revision of the classification of Dandy–Walker complex, we assumed that serial sonographic coronal and sagittal sections of the posterior fossa would more accurately visualize the vermis. This technique could avoid the false appearance of an enlarged cisterna magna on angled semi-coronal imaging[20]. We found that the vermis could be well delineated in the sagittal plane (Figure 2), and its longitudinal diameter (as well as its transverse diameter and the vermis–pons ratio) could be measured and compared with the results of our former studies[8,9].

The normality of the vermis and cisterna magna were confirmed by pre- or postnatal ultrasound or MRI in all cases (Figure 3). All seven women who were referred for possible vermian agenesis were ready to terminate the pregnancy on the basis of the diagnosis. However, after our evaluation, all continued with the pregnancy, and at a follow-up of 1–7.5 years, the children were found on clinical and neuroradiological evaluation to be developing normally.

These findings confirm the conclusion of Barkovich *et al.*[5] that rotation of the cerebellar vermis alone is a benign entity and could cause apparent enlargement of the cisterna magna on axial imaging scans. Therefore, we suggest that all cases of enlarged cisterna magna or suspected vermian agenesis on standard antenatal ultrasound be re-evaluated in the sagittal plane. We are aware that although sagittal ultrasound slices are undoubtedly necessary to explore adequately anomalies of the posterior fossa, the presence of an apparently normal vermis on a sagittal sonographic plane is far from sufficient to assess the normality of the posterior fossa or to rule out pathology associated with a poor prognosis. However, since we have created nomograms of the vermis and cerebellum, as well as nomograms of the fetal pons[8,9] all these measurements can be taken into account when determining the normality of the vermis in the presence of enlarged cisterna magna.

We recommend the addition of MRI to confirm sonographic data, especially to determine the volume of the posterior fossa. MRI can be used to determine the global volume of the posterior fossa, the position of the tentorium cerebelli, and in the biometric and morphological analysis of the cerebellum, vermis, brainstem and fourth ventricle. It is especially valuable in

the late stages of gestation or when the fetal position is unfavorable. It should be emphasized, however, that MRI has its limitations, including difficulty in imaging the vermis due to partial voluming of (i.e. shadowing by) the adjacent cerebellar hemispheres, parental anxiety, its costs, and the difficulties caused by fetal movement. Early MRI (prior to 25 weeks' gestation) adds little new information compared with a well-conducted detailed ultrasound scan. In fact, in the second trimester of pregnancy, midline sagittal imaging is sometimes easier to perform with ultrasound than it is with MRI[7,21,22].

The very high proportion of disagreement between prenatal sonographic diagnoses of Dandy–Walker malformation or variant and autopsy findings[23] further emphasizes the need for accurate sonographic demonstration of the cerebellar vermis. Our method may avoid a false diagnosis of vermian agenesis, sparing patients unnecessary tests and anxiety, and, in some cases, unnecessary termination of pregnancy. However, it should be emphasized that, as suggested by Guibaud[7] and Adamsbaum *et al.*[22], the term 'Dandy–Walker variant' is confusing and should be avoided. Instead, a differential diagnosis should be made between partial agenesis and rotation of the vermis.

Visualization of enlargement of the cisterna magna is challenging. It requires meticulous sonographic examination and possibly MRI imaging, especially in the sagittal plane. We recommend establishment of a universal nomenclature and, following this, determination of the prognosis of each anomaly. This study adds, in our opinion, a further prenatal sonographic entity that can help in the evaluation of abnormalities of the posterior fossa. Further studies and multicenter cooperation are needed to delineate this observation in particular and the posterior fossa in general.

ACKNOWLEDGMENT

We thank Gloria Ginzach and Hanni Penn for their editorial and secretarial assistance.

REFERENCES

1. Dandy WE. The diagnosis and treatment of hydrocephalus due to the occlusion of the foramina of Magendie and Luschka. *Surg Gynecol Obstet* 1921; **32**: 112–128.
2. Taggart JK, Walker AE. Congenital atresia of the foramens of Luschka and Magendie. *Arch Neurol Psychiatr* 1942; **48**: 583–595.
3. Raybaud C. Cystic malformations of the posterior fossa abnormalities associated with development of the roof of the fourth ventricle and adjoint meningeal structures. *J Neuroradiol* 1982; **9**: 103–133.
4. Ecker JL, Shipp TD, Bromley B, Benacerraf B. The sonographic diagnosis of Dandy-Walker and Dandy-Walker variant: associated findings and outcomes. *Prenat Diagn* 2000; **20**: 328–332.
5. Barkovich AJ, Bent KO, Norman D, Edwards MS. Revised classification of the posterior fossa and cyst-like malformations based on the results of multiplanar MR imaging. *Am J Neuroradiol* 1989; **10**: 977–988.
6. Klein O, Pierre-Kahn A, Boddaert N, Parisot D, Brunelle F. Dandy-Walker malformation: prenatal diagnosis and prognosis. *Childs Nerv Syst* 2003; **19**: 484–489.
7. Guibaud L. Practical approach to prenatal posterior fossa abnormalities using MRI. *Pediatr Radiol* 2004; **34**: 700–711.
8. Zalel Y, Seidman D, Brandt N, Lipitz S, Achiron R. The development of the fetal vermis: an in-utero sonographic evaluation. *Ultrasound Obstet Gynecol* 2002; **19**: 136–139.
9. Achiron R, Kivilevitch Z, Lipitz S, Gamzu R, Almog B, Zalel Y. The development of the human fetal pons: In utero ultrasonographic study. *Ultrasound Obstet Gynecol* 2004; **24**: 506–510.
10. Altman NR, Naidich TP, Braffman BH. Posterior fossa malformations. *Am J Neuroradiol* 1992; **13**: 691–724.
11. Nyberg DA, Cyr DR, Mack LA, Fitzsimmons J, Hickok D, Mahony BS. The Dandy-Walker malformation: prenatal sonographic diagnosis and its clinical significance. *J Ultrasound Med* 1988; **7**: 65–71.
12. Blazer S, Berant M, Sujuv PO, Zimmer E, Bronshtein M. Prenatal sonographic diagnosis of vermal agenesis. *Prenat Diagn* 1997; **17**: 907–911.
13. Chang MC, Russell SA, Callen PW, Filly RA, Goldstein RB. Sonographic detection of inferior vermian agenesis in Dandy-Walker malformations: prognostic implications. *Radiology* 1994; **193**: 765–770.
14. Kollias SS, Ball WS, Jr, Prenger EC. Cystic malformations of the posterior fossa: differential diagnosis clarified through embryologic analysis. *Radiographics* 1993; **13**: 1211–1231.
15. Lemire RJ LJ, Leech RW. *Normal and Abnormal Development of the Human Nervous System*. Harper & Row: Hagerstown, MD, 1975; 144–163.
16. Bromley B, Nadel AS, Pauker S, Estroff JA, Benacerraf BR. Closure of the cerebellar vermis: evaluation with second trimester US. *Radiology* 1994; **193**: 761–763.
17. Babcook CJ, Chong BW, Salamat MS, Ellis WG, Goldstein RB. Sonographic anatomy of the developing cerebellum: normal anatomy can resemble pathology. *AJR Am J Roentgenol* 1996; **166**: 427–433.
18. Pilu G, Perolo A, Falco P, Visentin A, Gabrielli S, Bovicelli L. Ultrasound of the central nervous system. *Curr Opin Obstet Gynecol* 2000; **12**: 93–103.
19. Pilu G, Visentin A, Valeri B. The Dandy-Walker complex and fetal sonography. *Ultrasound Obstet Gynecol* 2000; **16**: 115–117.
20. Laing FC, Frates MC, Brown DL, Benson CB, Di Salvo DN, Doubilet PM. *Radiology* 1994; **192**: 247–251.
21. Raybaud C, Levrier O, Brunel H, Girard N, farnarier P. MR imaging of brain malformations. *Childs Nerv Syst* 2003; **19**: 455–470.
22. Adamsbaum C, Moutard ML, Andre C, Merzoug V, Ferey S, Quere MP, Lewin F, Fallet-Bianco C. MRI of the fetal posterior fossa. *Pediatr Radiol* 2005; **35**: 124–140.
23. Carroll SG, Porter H, Abdel-Fattah S, Kyle PM, Soothill PW. Correlation of prenatal ultrasound diagnosis and pathologic findings in fetal brain abnormalities. *Ultrasound Obstet Gynecol* 2000; **16**: 149–153.

