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Ventriculomegaly and pericerebral CSF collection in the fetus: early stage of benign external hydrocephalus?

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Abstract *Background and purpose:* Mild ventricular dilatation or an asymmetric aspect of the atrium with prominent subrachnoid spaces is a challenging clinical condition in utero that requires prenatal MRI to rule out a destructive lesion or brain malformation. We report five cases that demonstrated benign external hydrocephalus postnatally, together with the prenatal MRI to define prenatal criteria of so-called benign external hydrocephalus. *Methods and results:* The prenatal MR images of five cases showing typical features of external hydrocephalus postnatally were reviewed. All cases showed in utero mild ventricular dilatation at the level of the atrium with enlargement of the subarachnoid spaces homo-, contra- or bilaterally, and predominantly in the parieto-occipital areas. The head circumference was normal in all cases with no family history of macrocephaly. MRI did not demonstrate either abnormal signal within brain parenchyma or loss of the normal layering of the developing brain. *Conclusion:* Posterior mild ventricular dilatation and prominent subarachnoid spaces in a posterior distribution can be considered an early stage of benign external hydrocephalus that is nicely illustrated by MRI.

Keywords Fetal MRI · Fetal ventriculomegaly · Benign hydrocephalus

Introduction

So-called benign external hydrocephalus (EH) is characterized postnatally by macrocephaly, enlarged ventricles (at the level of the frontal horns in a symmetric way), enlarged subarachnoid spaces from accumulation of cerebrospinal fluid (CSF) in the anterior parts of the hemispheres, and a square forehead [3]. The pathophysiology of benign enlargement of the ventricles includes prematurity (and intraventricular hemorrhage with the result of diminishing the ability of arachnoid granulations to absorb CSF), osseous origin (skull-base dysplasia), vascular origin (arteriovenous malformation), and metabolic disease such as mucopolysaccharidoses. However, in most cases the cause remains unknown and is considered to be idiopathic (or constitutional). In addition, ventricular dilatation is one of the most common clinical conditions [1] in cases referred for fetal brain magnetic resonance imaging (MRI) (38% in our experience), the most challenging condition being the evaluation of mild ventricular dilatation.

We report five documented cases of EH and point out that the criteria in the prenatal period are different from those in the postnatal period. The power of MRI to illustrate the development of the subarachnoid spaces is fascinating and also very effective in the managment of fetal ventricular dilatation.

Case reports

Five cases of mild ventricular dilatation with pericerebral accumulation of CSF depicted in utero are reported that postnatally proved to be idiopathic benign external hydrocephalus based on the clinical and/or neuroradiological features.

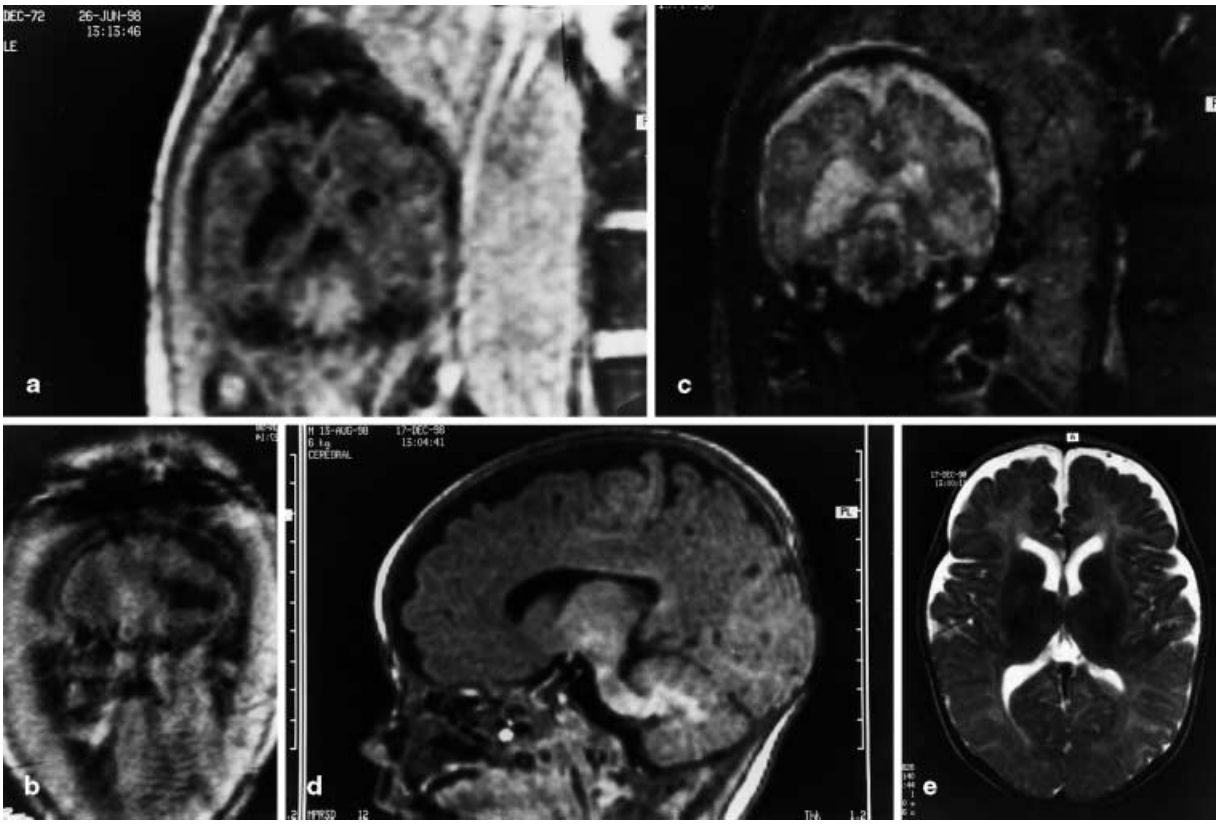


Fig. 1 Prenatal MRI in week 30: coronal (**a**) and sagittal (**b**) T1 WI, coronal T2 WI (**c**) show mild unilateral ventricular dilatation at the level of the atrium (**a, c**), associated with prominent subarachnoid spaces (**a–c**) bilaterally in the parieto-occipital areas. Postnatal MRI at 4 months of age: parasagittal T1WI (**d**) and axial T2WI (**e**) show typical features of benign EH as frontal accumulation of CSF as opposed to antenatal MRI

Case 1

B.V. was referred to fetal MRI following the depiction of mild enlargement of the posterior horns on US in week 30. Fetal brain MRI (T1- and T2-weighted images) performed in week 30 of gestational age (GA) demonstrated unilateral dilatation of the posterior horn (12 mm versus 6 mm of the contralateral side) associated with bilateral enlargement of the subarachnoid spaces (Fig. 1a–c). No abnormal signal changes were seen within the parenchyma. Because of breech presentation, the baby was delivered in week 39 through caesarian section with no context of birth asphyxia. Postnatal brain MRI performed at 4 months of age showed morphological changes of benign EH (Fig. 1d,e).

Case 2

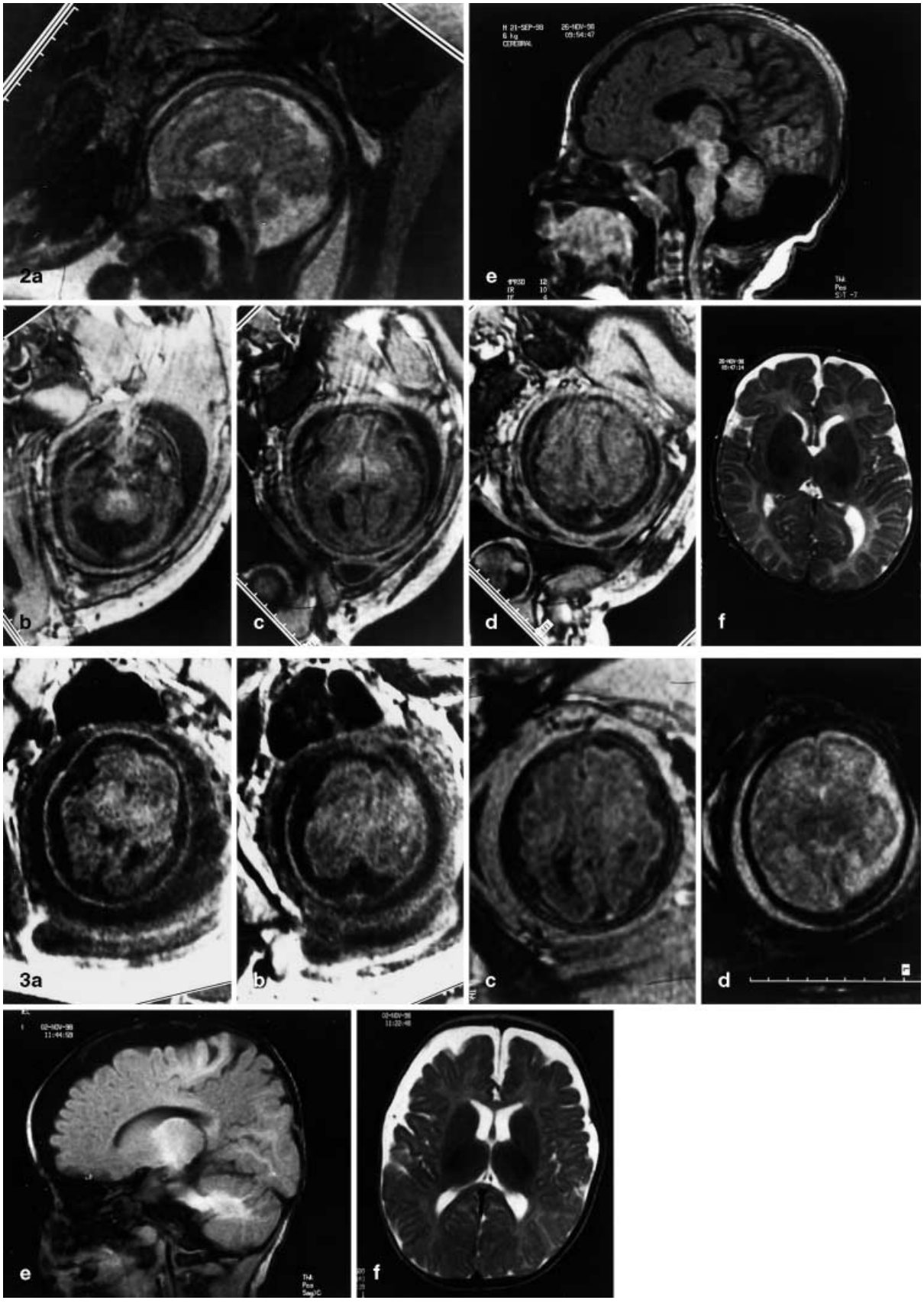
C.L. was referred at week 32 for suspected vermian agenesis. MRI (T1- and T2-weighted images) showed enlarged subarachnoid spaces within the posterior fossa, with no vermian agenesis, thus ruling out Dandy-Walker malformation, but with abnormal insertion of the cerebellum tentorium consistent with Blake's pouch abnormality (Fig. 2a,b). Unilateral mild ventricular dilatation of the posterior horn (11 mm versus 4 mm) was seen, which was associated with enlarged subarachnoid spaces homolaterally (Fig. 2a–c). Brain parenchyma dysplayed normal features. The head circumference was normal. The baby was born at term after an uneventful vaginal delivery. MRI performed at 2 months of age showed typical features of EH (Fig. 2f), associated with a posterior cerebellar cyst consistent with a Blake's pouch (Fig. 2e).

Case 3

M.E was referred for fetal brain MRI because of mild ventricular dilatation in a co-twin of a monochorionic biamniotic twin preg-

Fig. 2 Prenatal MRI in week 32: sagittal T2 WI (**a**), axial T1 WI (**b, c, d**) demonstrate Blake's pouch abnormality within the posterior fossa (**a, b**), associated with unilateral mild ventricular dilatation at the level of the posterior horn (**c**) and prominent subarachnoid spaces homolaterally in the parietooccipital area (**d, a**). Postnatal MRI at 2 months of age: sagittal T1 WI (**e**) and axial T2 WI (**f**) show the posterior cerebellar cyst consistent with a Blake's pouch, associated with the typical feature of EH, that is, frontal accumulation of CSF as prominent frontal horns and subarachnoid spaces in the frontal areas ▶

Fig. 3 Prenatal MRI in week 27: axial T1 WI (**a, b**) demonstrates unilateral mild ventricular dilatation at the level of the posterior horn (**a**) associated with prominent subarachnoid spaces bilaterally in the posterior areas (b) and contralateral enlargement of the subarachnoid spaces (**a**). Follow-up prenatal MRI in week 31: axial T1 WI (**c**) and T2 (**d**) WI show persistent mild unilateral ventricular dilatation (**c**) associated with contralateral subarachnoid spaces (**d**). Postnatal MRI at 5 months of age: parasagittal T1 WI (**e**) and axial T2 WI (**f**) show typical features of EH with frontal accumulation of CSF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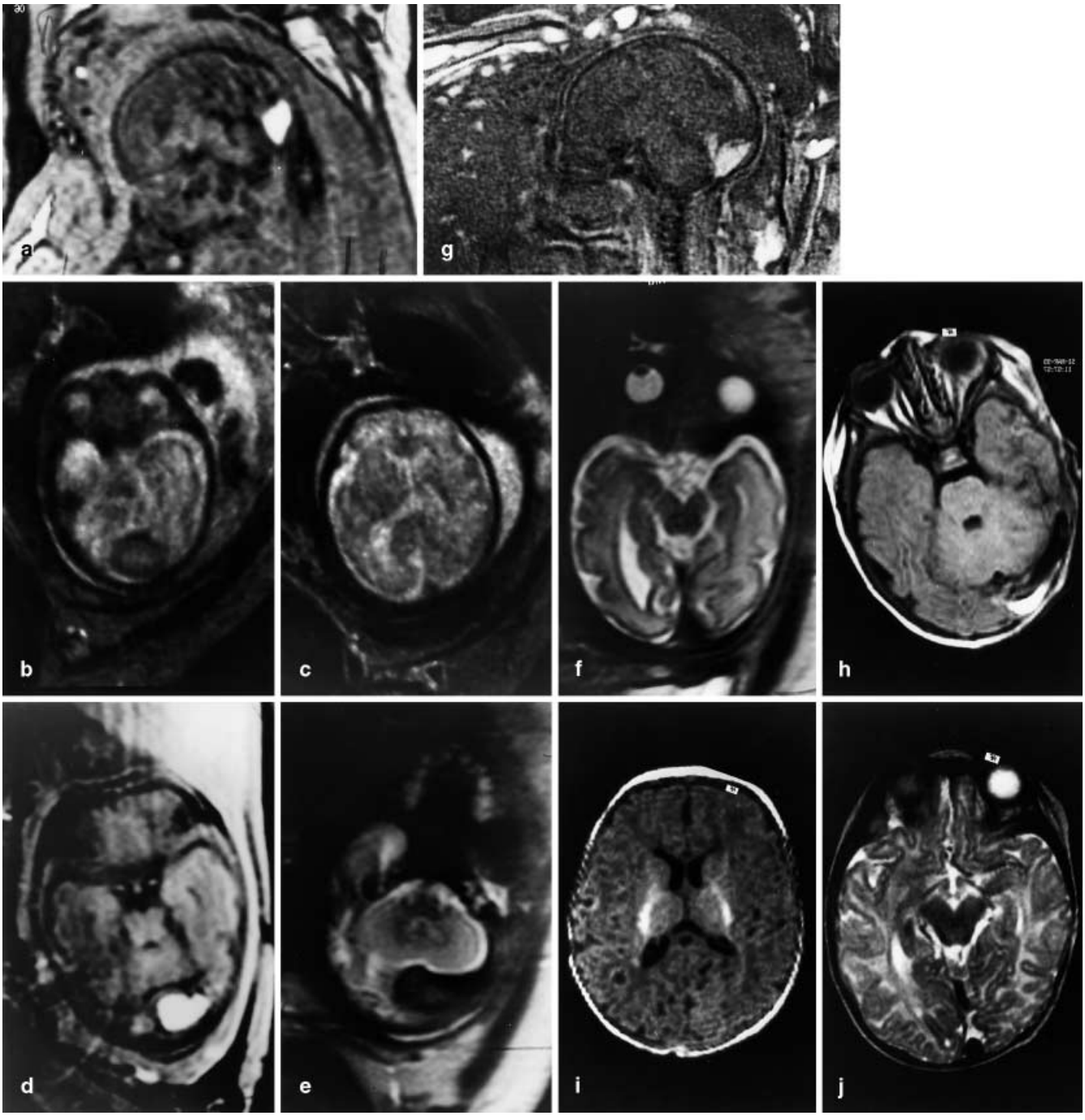

Fig. 4 Prenatal MRI in week 26: sagittal T1 WI (**a**) and axial T2 WI (**b, c**) show a lesion of the torcular as bright signal on T1 WI (**a**) and heterogeneous signal on T2 WI (**b**) consistent with thrombosis of the torcular, associated with mild unilateral ventricular dilatation at the level of the atrium (**c**). Follow-up MRI in week 31: axial T1 WI with fat saturation (**d**), axial HASTE images (**e, f**), and sagittal angiographic image (**g**) demonstrate thrombosis of the torcular and lateral sinus as bright signal on T1 WI (**d**) and low signal on haste image (**e**), associated with mild unilateral ventricular dilatation of the posterior horn (**f**). Note that angiographic image does not show abnormal vessels, ruling out a fistula (**g**). Postnatal MRI at 2 months of age: axial T1 WI with fat saturation (**h**), axial gradient echo T1 WI (**i**) and axial T2 WI (**j**) confirm the venous abnormality (**h, j**) associated with typical feature of EH (**i**)

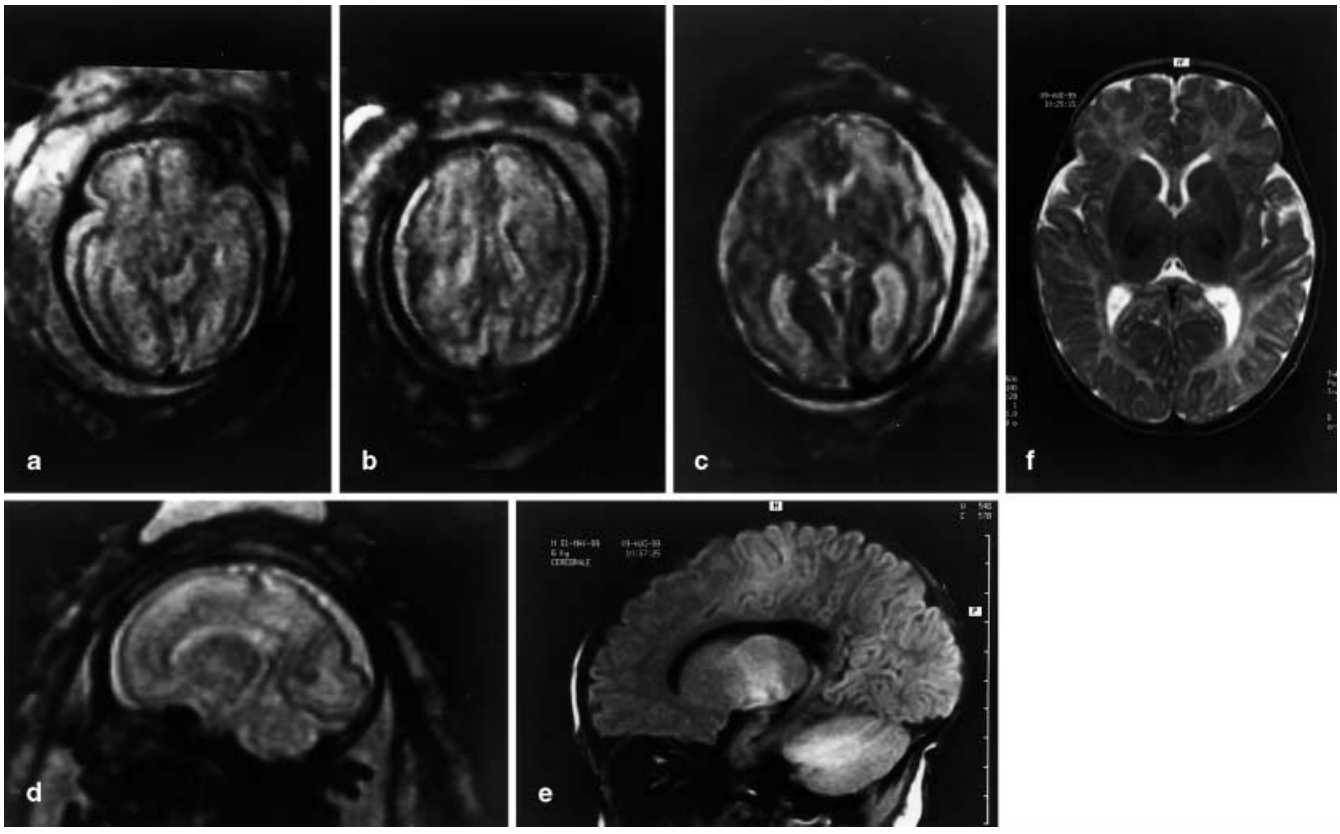

Fig. 5 Prenatal MRI in week 25: axial T2 WI (**a, b**) show unilateral mild ventricular dilatation of the posterior horn (**a**). The subarachnoid spaces are prominent bilaterally (**b**) – consistent with the gestational age. Follow-up prenatal MRI at week 29: axial (**c**) and parasagittal (**d**) T2 WI demonstrate a persistent mild ventricular dilatation at the level of the atrium (**c**) associated with prominent subarachnoid spaces in the parieto-occipital area (**d**). Postnatal MRI at 2 months and 15 days of age: parasagittal T1 WI (**e**) and axial T2 WI (**f**) show frontal accumulation of the CSF as prominent subarachnoid spaces in the frontal areas

nancy. MRI (T1- and T2-weighted images) performed at week 27 showed mild unilateral ventricular dilatation of the posterior horn (9 mm versus 4 mm) (Fig. 3a) associated with enlarged subarachnoid spaces bilaterally in the posterior areas and contralateral enlargement of the subarachnoid spaces overlying the cerebral hemisphere (Fig. 3a,b), with no loss of either signal contrast or cerebral mantle layering. A follow-up fetal brain MRI (T1- and T2-weighted images), performed in week 31, demonstrated unilateral ventricular dilatation posteriorly (12 mm versus 6 mm) (Fig. 3c) associated with contralateral enlargement of the subarachnoid spaces (Fig. 3d). No abnormality was depicted within the brain parenchyma. Delivery was uneventful at week 37. Post-natal brain MRI, performed at 5 months of age, demonstrated the typical morphological features of EH (Fig. 3e, f).

Case 4

D.B was referred for suspected arachnoid cyst of the posterior fossa in week 26. MRI (T1- and T2-weighted images) demonstrated a lesion within the torcular as bright signal intensity on T1-weighted images (WI) (Fig. 4a) and heterogeneous signal intensity on T2 WI (Fig. 4b). This lesion did notcommunicate with the subcutaneous fat, ruling out a midline developmental abnormality, such as a dermoid cyst or dermal fistula. The lesion was brighter than the subcutaneous fat on T1 WI, ruling out a lipoma. Unilateral mild ventricular dilatation of the posterior horn (11 mm versus 5 mm) was also seen (Fig. 4c). No signal abnormality was depicted either in the white matter, cortical ribbon, germinal matrix, or in the basal ganglia. A follow-up fetal brain MRI was performed in week 31, including T1 WI, Haste images, T1 WI with fat saturation, and inversion recovery images. This MRI confirmed spontaneous thrombosis of the torcular and lateral sinus (Fig. 4d,e) with no brain injury of hypoxoischemic origin. Unilateral mild ventricular dilatation was seen (Fig. 4f), associated with slightly enlarged subarachnoid spaces homolaterally. No vessel abnormality was seen on angiographic images (Fig. 4g). Post-natal brain MRI at 2 months of age did not show an arteriovenous fistula, but instead thrombosis of the torcular and of the lateral sinus (Fig. 4h, j) associated with typical features of EH (Fig. 4i). No coagulation disorder was found.

Case 5

S.V. was referred for mild ventricular dilatation. MRI in week 25 showed unilateral mild ventricular dilatation of the posterior horn (13 mm versus 8 mm) (Fig. 5a, b), with no signal abnormality of either cortical ribon, white matter or germinal matrix. The subarachnoid spaces appeared large, but this feature was consistent with the gestational age. A follow-up MRI was performed in week 29, showing mild ventricular dilatation posteriorly (12 mm versus 9 mm; Fig. 5c) with prominent subarachnoid spaces bilaterally in a posterior distribution (Fig. 5d). Postnatal MRI performed at 2 months and 15 days of age, following an uneventful delivery, showed typical features of EH (Fig. 5e,f).

In summary, this retrospective analysis of five cases shows that the prenatal criteria of benign external hydrocephalus are not similar to those in the postnatal period. Prenatal criteria include: (1) mild ventriculomegaly as asymmetric dilatation at the atria level; (2) enlargement of the subarachnoid spaces posteriorly at the parieto-occipital levels either bilaterally, homo- or contralaterally to the ventricular dilatation; (3) the head circumference is normal. The accumulation of CSF in utero dysplays a more posterior distribution, as opposed to the postnatal period with frontal accumulation of CSF. Note that there was no family history of enlarged head circumference in any of these cases.

Discussion

As already mentioned in the introduction, the pathophysiology of benign EH includes prematurity, osseous origin, vascular origin, and metabolic disease. All of the children in this study were born at term with no intrapartum history, ruling out intraventricular hemorrhage as a possible cause. Case 4 fit the criteria for a vascular origin. However, in the postnatal MRI no arteriovenous fistula or abnormal venous drainage was found . One could also say that case 2 fit the definition of an abnormal venous drainage origin because of the presence of a posterior fossa cyst. However, this case is not consistent with a Dandy-Walker malformation. The enlarged retrocerebellar CSF spaces do not lead to brainstem compression, abnormal venous drainage thus being unlikely. The five cases reported can be considered to be idiopathic EH since no etiology was found, including the family history.

The postnatal criteria of benign EH, as already mentioned, include macrocephaly, frontal accumulation of CSF as enlarged ventricles at the level of the frontal horns and enlarged subarachnoid spaces in the anterior parts of the cerebral hemispheres, and a square forehead. These criteria are not available in utero, where the accumulation of CSF displays a more posterior distribution, in an asymmetric way, as opposed to the postnatal period; the posterior distribution manifests as enlarged ventricles at the atria level and as enlarged subarachnoid spaces in the parieto-occipital area. These antenatal features probably reflect the development of the CSF pathway. Primitive subarachnoid space develops by cavity formation of the meninx primitiva in stage 14 [2]. Formation of the primitive subarachnoid space is known to spread from the ventral portion of the mes- and rhombencephalon caudally to the spinal cord and cranially to the prosencephalon. It also tends to spread from the ventral to the dorsal portion of the neural tube. The primitive subarachnoid space is well developed at the end of the embryonic period. The primitive subarachnoid space can be called subarachnoid space after separation of the arachnoid membrane from the primitive duramater in the fetal period and is fully developed by week 30 of fetal life [4]. Arachnoid granulations develop in patches as depressions in the wall of the venous sinuses (lacunae laterales, that is, at the level of the posterior third of the superior sagittal sinus, their formation proceeding in a frontal direction). Depressions are depicted during week 26. Typical arachnoid villi are seen in week 35 while arachnoid granulations are seen in week 39 [4]. From these histologic studies the formation of the subarachnoid spaces, through cavitation of the meninx primitiva with its consequent accumulation of CSF, spreads from the ventral to the dorsal portion of the neural tube, thus leading to accumulation of CSF in the posterior areas and in the posterior fossa. Arachnoid villi and arachnoid granulations are seen later in the posterior part of the superior sagittal sinus with the intercellular spaces in the matrix that could be filled with CSF, again probably responsible for posterior accumulation of CSF.

MRI is a powerful tool for illustrating the morphological changes of the developing brain [5–8]. Along with the improvement of MR techniques, more and more subtle cases of ventricular dilatation are being referred to MRI units. Ventricular dilatation may be of destructive origin, malformative origin, hydrocephalic origin, destructive and hydrocephaic origin, or malformation. A malformation is easy to depict because of the specific morphological features [9–11], the most difficult condition being the evaluation of histogenetic disorders. A destructive origin is most challenging, but the diagnosis can still be achieved through the ventricular morphology, the loss of cell layer visibility, loss of normal signal contrast, and loss of the maturation milestones [12]. Although more cases are needed with postnatal imaging correlations, we will now be able to suggest benign external hydrocephalus in utero based on the peculiar MRI features: asymmetric posterior ventricular dilatation associated with enlarged subarachnoid spaces in the parieto-occipital areas. However, we have to bear in mind the fact that postnatal imaging correlations by MRI or CT scan are very often difficult to obtain since these children usually do not show neurologic abnormality or developmental delay.

References

1. D'Ercole C, Girard N, Boubli L, Potier A, Chagnon C, Raybaud C, Blanc B (1993) Prenatal diagnosis of fetal cerebral abnormalities by ultrasonography and resonance imaging. Eur J Obstet Gynecol Reprod Biol 50:177–184
2. D'Ercole C, Girard N, Cravello L, Boubli L, Potier A, Raybaud C, Blanc B (1998) Prenatal diagnosis of fetal corpus callosum agenesis by ultrasonography and magnetic resonance imaging. Prenat Diagn 18:247–253
3. Filly RA, Goldstein RB, Callen PW (1991) Fetal ventricle: importance in routine obstetric sonography. Radiology 181:1–7

4. Girard NJ, Raybaud CA (1992) In vivo MRI of fetal brain cellular migration. J Comput Assist Tomogr 16:265–267
5. Girard N, Raybaud C (1998) Nuclear magnetic resonance brain imaging in the prenatal period (in French). Arch Pediatr 5 [Suppl 2]:171–174
6. Girard N, Raybaud C, D'Ercole C, Boubli L, Chau C, Cahen S, Potier A, Gamerre A (1993) In vivo MRI of the fetal brain. Neuroradiology 35:431–436
7. Girard N, Raybaud C, Poncet M (1995) In vivo MR study of brain maturation in normal fetuses. AJNR Am J Neuroradiol 16:407–413
8. Girard N, Raybaud C, Poncet M (1996) In vivo magnetic resonance imaging of the fetal brain. In: Guerrini R, Andermann F, Canapicchi R, Roger J, Zifkin BG, Pfanner P (eds) Dysplasia of cerebral cortex and epilepsy. Lippincott-Raven, Philadelphia, pp 101–103
9. Gomez DG, DiBenedetto AT, Pavese AM, Firpo A, Hershoan DB, Potts DG (1981) Development of arachnoid villi and granulations in man. Acta Anat 111: 247–258
10. Maytal J, Alvarez LA, Elkin CM, Shinnar S (1987) External hydrocephalus: radiologic spectrum and differentiation from cerebral atrophy. AJNR Am J Neuroradiol 8:271–278
11. Osaka K, Handa H, Matsumoto S, Yasuda M (1980) Development of the cerebrospinal fluid pathways in the normal and abnormal embryos. Child's Brain 6:26–38
12. Sonigo P, Elmaleh A, Fermont L, Delezoide AL, Mirlesse V, Brunelle F (1996) Prenatal MRI diagnosis of fetal cerebral tuberous sclerosis. Pediatr Radiol 26:1–4