

CLINICAL CASE - TEST YOURSELF

Neuro/Head and Neck Radiology

Routine brain US in a full-term neonate

Ioanna Tritou¹, Georgios Niotakis¹, Maria Raissaki²¹Department of Radiology, Venizeleion General Hospital, Heraklion, Crete, Greece²Department of Radiology, University Hospital of Heraklion, University of Crete, Medical School, Crete, Greece

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PART A

Brain ultrasonography (US) was performed in a full-term male neonate (**Fig. 1**). He was born to a mother with gestational diabetes after an otherwise uncomplicated pregnancy and delivery, without any clinical abnormalities and with normal head circumference

(50-85% percentile) and body weight (85% percentile). A follow up brain US (**Fig. 2**) and magnetic resonance imaging (MRI) (**Fig. 3**) were performed 1 month later, while his most recent MRI was at 5 years of age (**Figs 4, 5**).

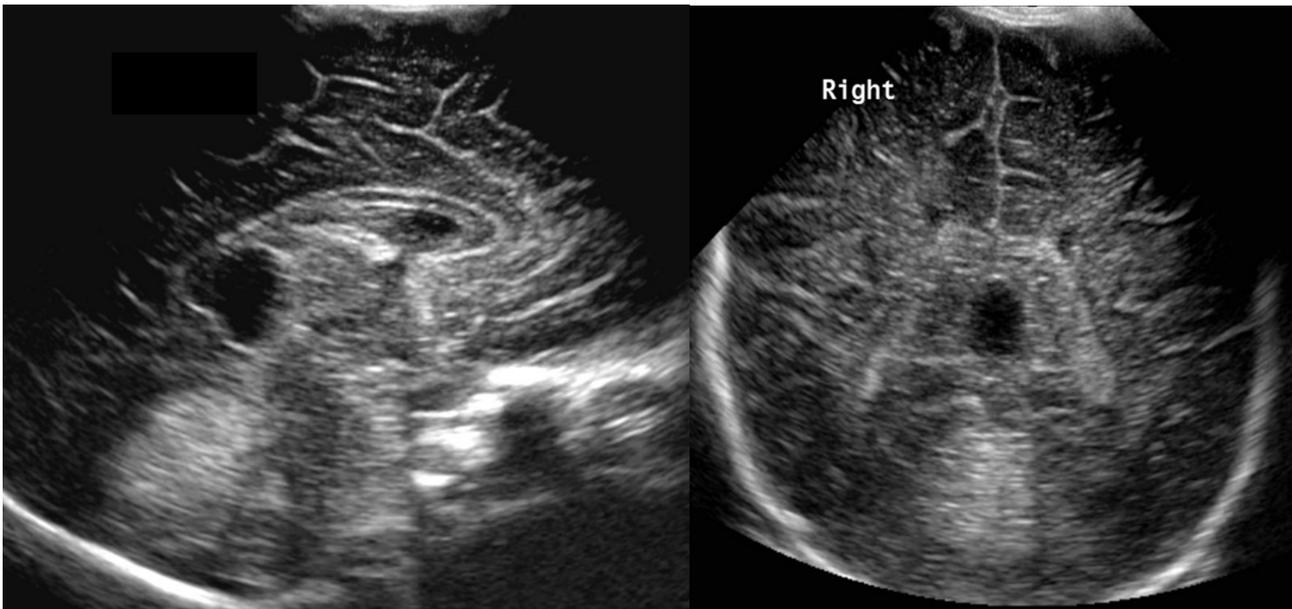


Fig. 1. Brain US, mid-sagittal scan (left) and coronal scan (right), at day 7 of life.



CORRESPONDING
AUTHOR,
GUARANTOR

Maria Raissaki, MD, PhD,
Associate Professor in Paediatric Radiology, Department of Radiology, University
Hospital of Heraklion, University of Crete, Medical School, Crete, Greece,
32 Sofokleous Str, 71306, Heraklion, Crete, Greece, Email: mraissaki@yahoo.gr

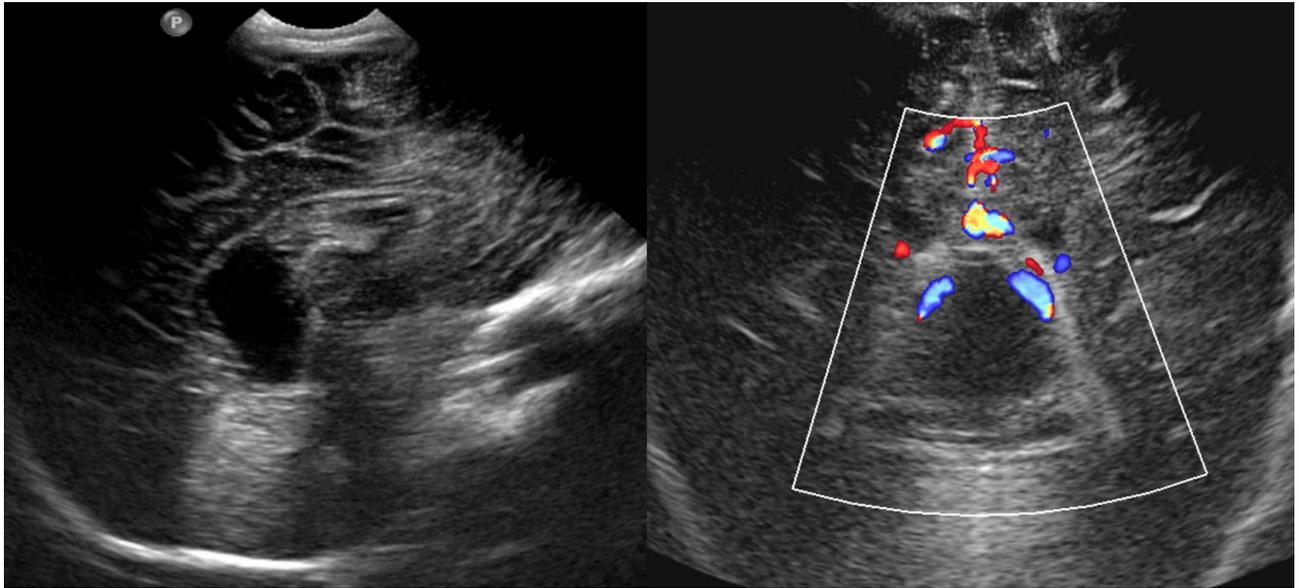


Fig. 2. Brain US, mid-sagittal scan (left) and coronal scan with colour Doppler (right) at 1.5 months of age.

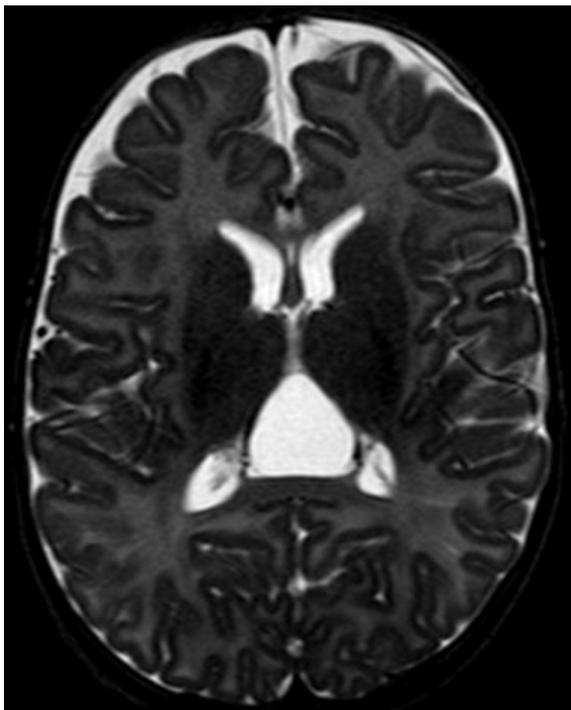


Fig. 3. Brain MRI at 1.5 month of age axial T2W image.

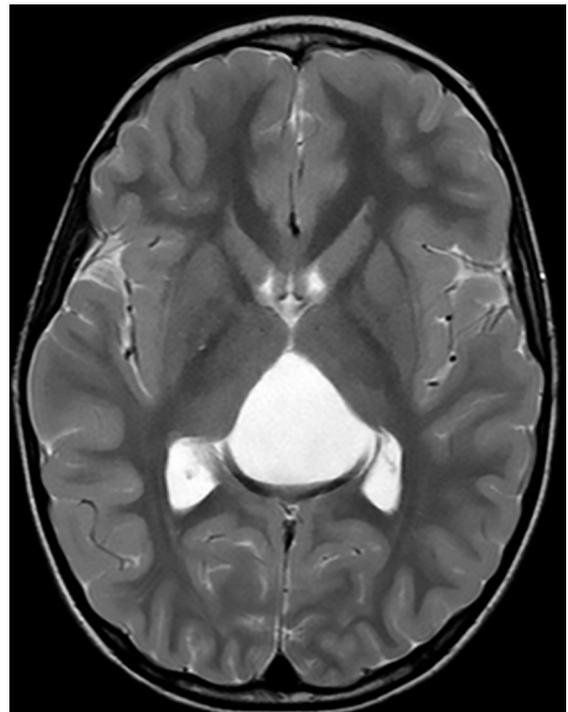


Fig. 4. MRI at 5 years of age, axial T2W image.

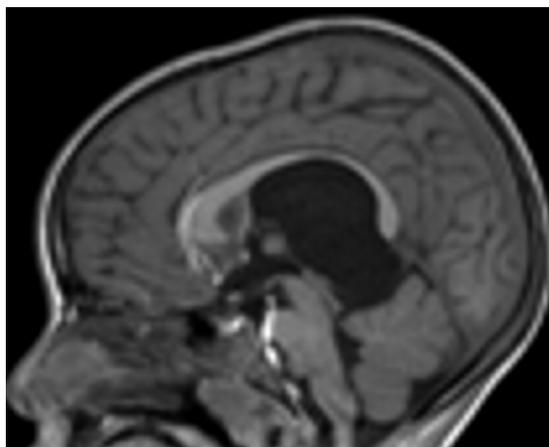


Fig. 5. MRI at 5 years of age, sagittal T1W image.

PART B

Diagnosis: Progressively enlarging cavum velum interpositum cyst

US revealed the presence of a cyst at the anatomic region of the velum interpositum (**Fig. 1**), measuring 1.2 cm in diameter and consistent with the diagnosis of a cavum velum interpositum (CVI) cyst.

A follow-up US performed one month later showed an increase in size of the CVI cyst, measuring 3 cm in diameter, causing lateral displacement of the internal cerebral veins (**Fig. 2**).

Complementary brain MRI disclosed the CVI cyst, which exhibited fluid signal in all sequences (**Fig. 3**), increased diffusivity and no contrast enhancement (not shown). The CVI cyst measured 2 cm (axial) x 1.9 cm (antero-posterior) x 3.3 cm (cranio-caudal diameter). As the child did not have any major neurologic deficits or clinical and radiological signs of hydrocephalus or any midline structure shift, it was decided to be followed-up annually. Subsequent MRI exams showed progressive enlargement of the cyst that reached the size of 4.3 cm (axial) x 4.6 cm (antero-posterior) x 5.4

cm (cranio-caudal diameter) at 3 years of age. Following consultation, surgery was temporarily discouraged because there were no headaches and annual follow-up continued. The latest MRI at 5 years of age showed size stabilisation during the last 1.5 year with mild compression of adjacent structures (**Figs. 4, 5**).

The velum interpositum is a midline potential sub-arachnoid space containing cerebrospinal fluid (CSF), entirely surrounded by pia matter, gradually regressing between the seventh month of foetal life and the second year of age [1, 2]. It represents a part of the normal brain development of the cerebral midline structures. Thus, it may be evident prenatally and usually disappears after birth in full term babies. It is located superior to the pineal gland, below/medially to the fornices and above the internal cerebral veins. Its boundaries include the hippocampal commissure and corpus callosum superiorly, the tela choroidea inferiorly and the crus of the fornix on each side supero-laterally. It lies between the posterior halves of the two lateral ventricles and may extend anteriorly

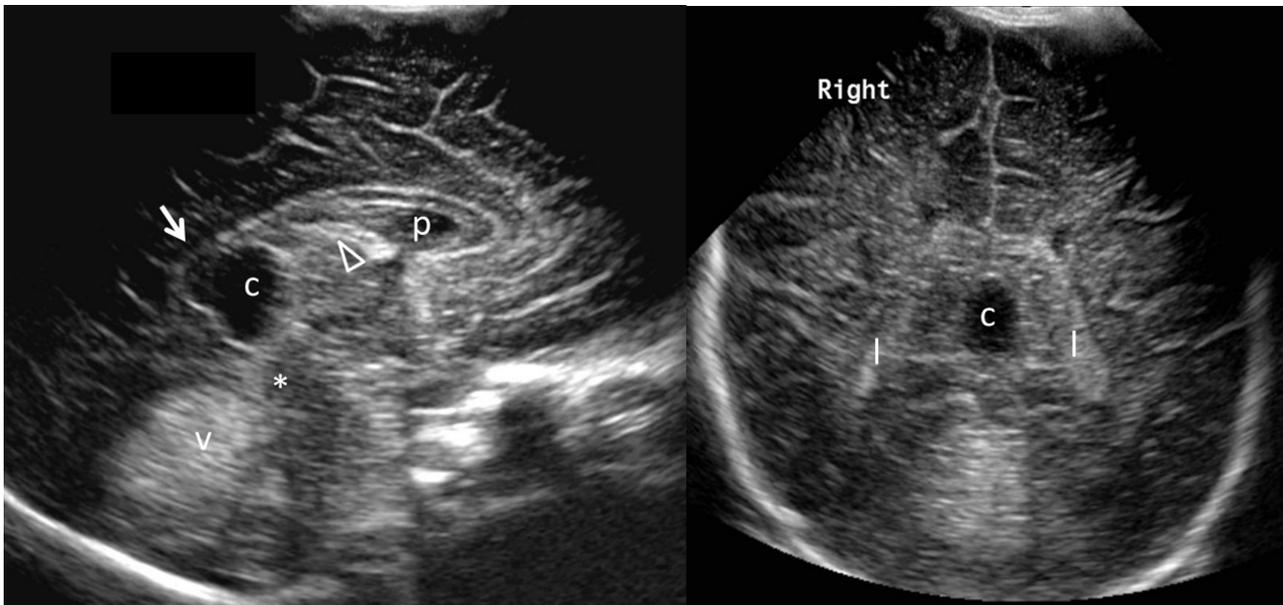


Fig. 1. Brain US, mid-sagittal scan (left) and coronal scan (right) at day 7 of life. A cystic structure representing the CVI (c) is seen below and anterior to the splenium of the corpus callosum (arrow). The CVI is separated from the cavum septum pellucidum (p) by the column of the fornix (arrowhead) and is away from the quadrigeminal plate (*) and the vermis (v). l: choroid plexi of the lateral ventricles.

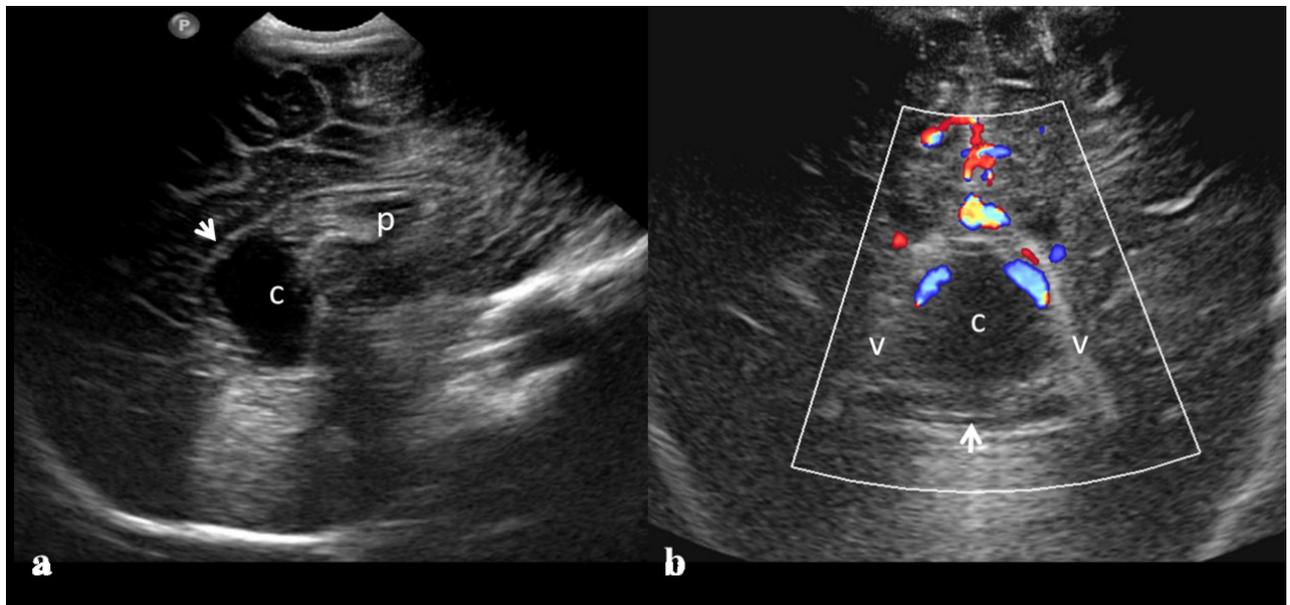


Fig. 2. Brain US, mid-sagittal scan (left) and coronal scan with colour Doppler (right) at 1.5 months of age. The CVI cyst (c) has enlarged and remains below and anterior to the splenium of the corpus callosum (arrow) and between the lateral ventricles, which appear normal in size (v: choroid plexi). The internal cerebral veins (in blue) are laterally displaced. p: cavum septum pellucidum.

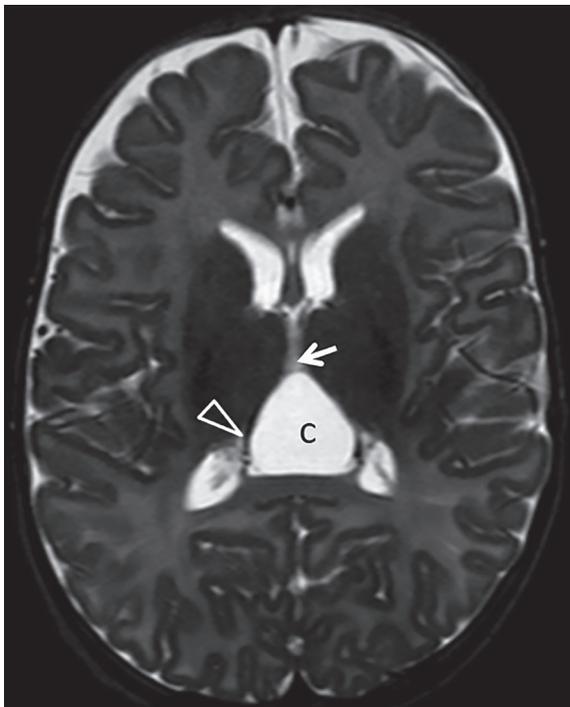


Fig. 3. Brain MRI at 1.5 month of age, T2-weighted sequence, axial scan. The CVI cyst (c) is demonstrated as a midline, triangular-shaped, well-demarcated hyperintense lesion, with its base in contact with the splenium and its apex at the floor of the third ventricle (arrow). The internal cerebral veins are seen as flow voids (arrowhead), been displaced inferiorly and laterally.

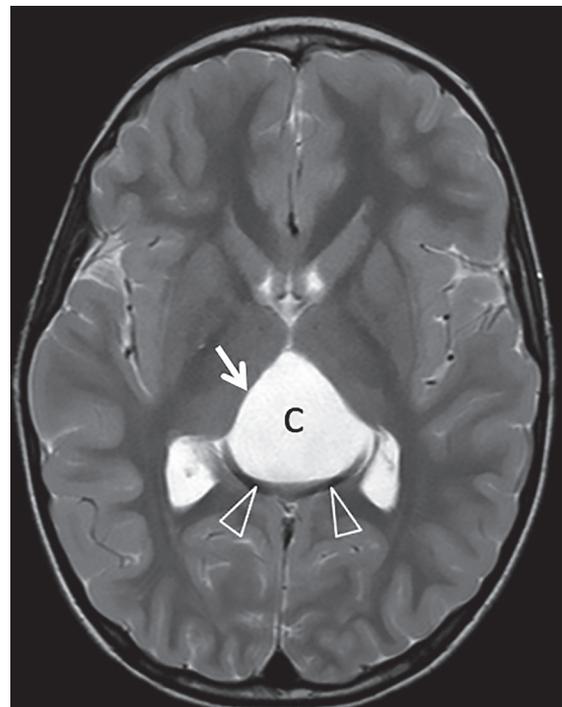


Fig. 4. MRI at 5 years of age, axial T2W image. The CVI cyst (c) has enlarged causing mild compression (arrow) on adjacent thalami. Note inferior and lateral displacement of the internal cerebral veins (flow voids, arrowheads) which distinguishes CVI from a quadrigeminal plate cyst and a pineal cyst. There is no ventriculomegaly while white matter signal intensity is unremarkable.

to the roof of the third ventricle as far as the interventricular foramina. Caudally, it opens into the cisterna vena magna Galeni between the two layers of the tela choroidea [1-3].

The enlargement of this potential space may be explained by a ball-valve-type of connection between the CVI cistern and its open posterior part to the cisterna venae magna Galeni. Therefore, its course is unpredictable. If this happens, it is called CVI [1]. In case it reaches 10 mm in diameter, and/or causes mass effect it is called a CVI cyst and it is arbitrarily considered an arachnoid cyst by some authors, although this is controversial [4, 5]. CVI cysts of a moderate size have been described [6, 7]. A large cyst with documented progressive enlargement as in our case is rare.

The clinical significance and natural history of CVI cysts are unclear. CVI cysts, when found antenatally by US, usually remain stable in size, may be accompanied by ventriculomegaly and usually disappear soon after birth or between 6 months and 1 year of age. CVI cysts have also been detected in children older than 2 years old and in adults and are thought to represent an incidental persistent primitive structure without associated additional midline anomalies [3, 5-9]. However, psychotic disorders, psychomotor retardation, seizures, autism, headaches and memory disturbances have also been reported in patients with CVI cysts [3, 4, 8, 10]. Because CVI cyst course is unpredictable, follow-up is recommended to exclude increase in size and hydrocephalus as a potential complication [3, 4, 6].

CVI appears as a triangular or rounded (in the case of the cyst)-shaped cystic space between the posterior parts of the bodies of the lateral ventricles with no or mild mass effect against neighbouring structures. Its apex lies just behind the foramen of Monro. On sagittal images, it is described as an "inverted helmet" with the convex part looking down and bordered by the internal cerebral veins and its upper part usually flat, taking the shape of the inferior part of the splenium [9]. However, CVI may extend anteriorly with concomitant splaying and anterior displacement of

the fornices, lateral and downwards displacement of inferior cerebral veins and potential hydrocephalus [9]. Its caudal extension may cause compression of the quadrigeminal cistern, the brainstem and the cerebellum [3-10].

The differential diagnosis of a CVI cyst should include other midline cysts based on the location, the effect on fornices/internal cerebral veins and ancillary findings. An enlarged third ventricle due to hydrocephalus coexists with lateral ventricle enlargement. An aneurysm of the vein of Galen is easily excluded due to specific location and colour Doppler appearance. A cavum vergae cyst coexists with a cavum septum pellucidum, lies above the internal cerebral veins and is seen separately from them while a CVI cyst encloses internal cerebral veins at its lower and lateral borders [4, 7]. A quadrigeminal plate arachnoid cyst and a pineal cyst, in contrast to a CVI cyst, lift the inferior cerebral veins upwards because the arachnoid membrane in the quadrigeminal plate and the pineal gland are located below them. An epidermoid cyst exhibits restricted diffusion [7-10].

Treatment is controversial. Follow-up is advised when clinical symptoms are lacking or considered not serious and no hydrocephalus or midline shift exist [5, 7, 9]. Endoscopic fenestration of the cyst has been applied as the surgical method of choice in adults, especially in those with hydrocephalus, headaches and other symptoms attributed to compression by the CVI cyst [4-6, 10]. In our patient, management was the result of long discussions with the neurosurgeons, who considered that the cyst, albeit enlarging in the past, was currently stable. Since the boy had not been experiencing significant or deteriorating disabilities, a conservative management with close clinical and imaging follow-up was jointly decided. The patient has been closely monitored since and further neurosurgical consultation will be sought if headaches or additional neurological symptoms occur. **R**

Conflict of interest

The authors declared no conflicts of interest.

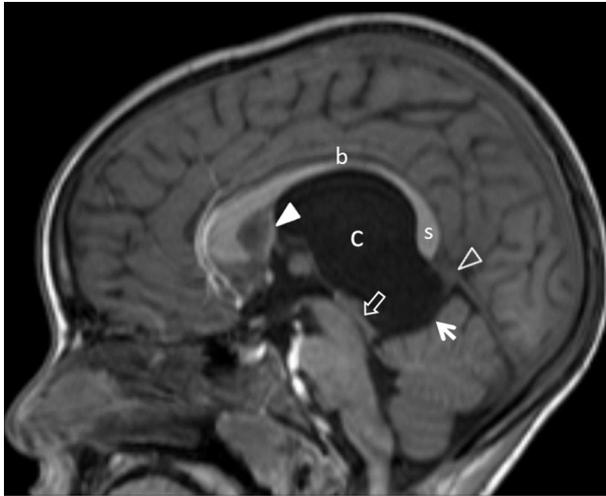


Fig. 5. MRI at 5 years of age, sagittal T1W image. The thin-walled CVI cyst (c) causes anterior displacement of the fornix (white arrowhead) and mild elevation and compression of the corpus callosum body (b) and splenium (s). Note downwards extension with compression-distortion of the tectum (open arrow), vermis (white arrow) and posterior displacement of pineal gland (open arrowhead).



KEY WORDS

Cavum Vellum Interpositum; Cyst; Symptomatic; Ultrasonography; MRI

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