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Case Report

Spontaneous Ventriculostomy: Report of Three Cases Revealed by Flow-Sensitive Phase-Contrast Cine MR Imaging

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Summary: Spontaneous ventriculostomy is a rare condition that occurs with the spontaneous rupture of a ventricle, resulting in a communication between the ventricular system and the subarachnoid space. Three cases of spontaneous ventriculostomy through the floor of the third ventricle that occurred in cases of chronic obstructive hydrocephalus are presented. The communication was identified via flow-sensitive phase-contrast cine MR imaging. Spontaneous ventriculostomy is probably a result of a rupture of the normally thin membrane that forms the floor of the third ventricle and, with long-standing obstructive hydrocephalus, creates an internal drainage pathway that spontaneously compensates for the hydrocephalus.

Obstructive hydrocephalus can cause rupture of the walls of the ventricles, generally at the level of the medial wall of the trigone or at the pineal recesses of the third ventricle. This rupture results in direct communication of the ventricular system with the subarachnoid space; a condition called spontaneous ventriculostomy (SV) (1, 2). Only one previous report has described an SV through the floor of the third ventricle into the suprasellar and prepontine cisterns diagnosed by means of ventriculography (3). This rare communication occurs with rupture of the thinnest part of the floor of the third ventricle, an area located between the median eminence of the tuber cinereum and the mamillary bodies (1). In this paper, we report three cases of SV that were discovered by MR imaging. This rare condition, which results in an internal drainage pathway that probably compensates the hydrocephalus, should be kept in mind in the preoperative assessment of obstructive hydrocephalus when endoscopic fenestration of the floor of the third ventricle is considered as an alternative to extracranial shunt.

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Case Reports

Technique

We collected three cases of SV through the floor of the third ventricle from our case records at the Vall d'Hebron University Hospitals. All three patients initially underwent cranial CT, which showed ventricular enlargement. The patients then were studied with MR imaging consisting of sagittal, axial, and coronal T1-weighted (450–550/14–15 [TR/TE]) images, and axial proton density–weighted and T2-weighted conventional spin-echo (2500/20–80) images. In one case (case 1), coronal and sagittal T2-weighted fast spin-echo images (5000/90) also were obtained.

As part of our routine workup to determine treatment options for adult patients having hydrocephalus, who are potential candidates for ventricular shunting, we performed a phase-contrast sequence to examine CSF pulsations. These sequences were performed with retrospective cardiac-gating, midsagittal, two-dimensional fast imaging with steady-state precession (2D FISP) using the following parameters: 70/13 (TR/TE); flip angle, 15°; section thickness, 4 mm; matrix, 192×256; field of view, 250 mm; and measurement time, 7 minutes 24 seconds. Sensitivity was set to produce a phase change of 180° for a velocity of 100 mm/s in the superior–inferior direction. The 16 flow-encoded images obtained per cardiac cycle were evaluated in a closed-loop cine format to assess qualitatively the dynamics of the CSF pulsation in the posterior fossa and craniocervical junction. The gray scale of the flow-encoded images is proportional to velocity of CSF along the craniocaudal axis. By convention, negative velocities (represented in black) are craniocaudal (systolic), and positive velocities (represented in white) are caudocranial (diastolic).

Case 1

A 17-year-old woman reported continuous mild headache over 1 year. The neurologic examination was normal. Cranial CT study revealed a supratentorial hydrocephalus and cranial MR imaging, which included sagittal, coronal, and transverse T1- and T2-weighted images (Figs 1A–B), showed an intrinsic, nonenhancing, tectal mass that had produced aqueductal stenosis. Axial proton density–weighted images did not show periventricular edema, indicative of acute obstructive hydrocephalus (Fig 1C). The flow-sensitive phase-contrast cine MR imaging confirmed the obstructed CSF flow through the stenotic aqueduct. Additionally, MR imaging enabled identification of an abnormal CSF wave through a gap located in the floor of the third ventricle between the tuber cinereum and the mammillary bodies that connected the ventricular system with the suprasellar and prepontine cisterns (Fig 1D). A retrospective analysis of the conventional T1- and T2-weighted images showed loss of integrity of the floor of the third ventricle between the tuber cinereum and the mammillary bodies. However, there were no indications of CSF flow across the floor of the third ventricle, even on the coronal and sagittal T2-weight-

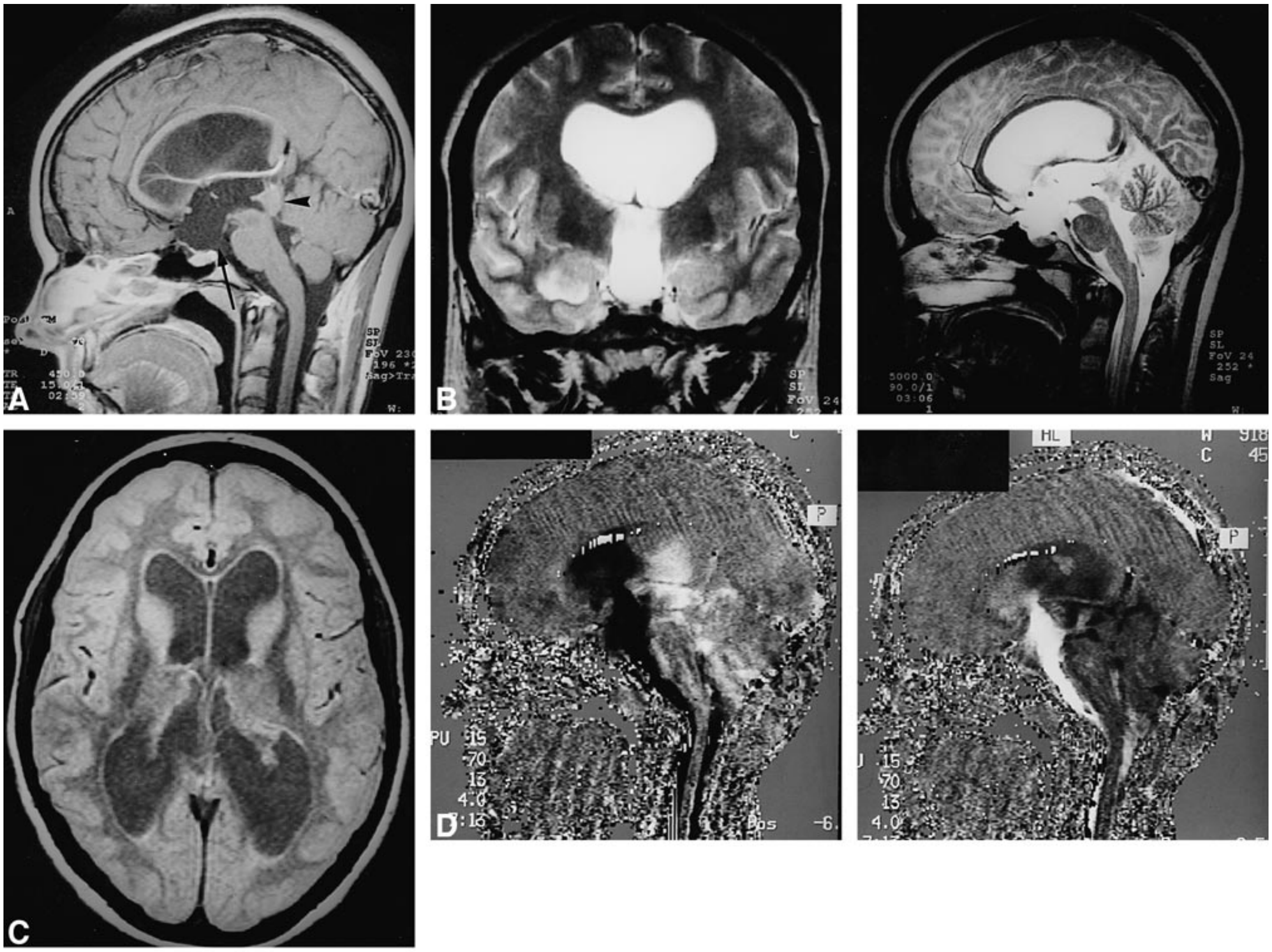

FIG 1. Aqueductal stenosis due to presumed benign tectal glioma.

Sagittal midline contrast-enhanced T1-weighted image [(SE 450/15/2)(TR/TE/excitations)] shows nonenhancing bulbous enlargement of tectum (*arrowhead*), obliterating aqueduct and producing severe hydrocephalus. A small gap in floor of third ventricle in front of mamillary bodies can be identified (*arrow*).

B. Coronal (*left*) and sagittal (*right*) T2-weighted images (FSE 5000/90/2) show no flow-void across floor of third ventricle.

C. Proton-density-weighted axial image (SE 2200/20/1) at level of lateral ventricles shows marked hydrocephalus but no periventricular edema, indicating acute obstructive hydrocephalus.

D. Sagittal midline phase-contrast MR imaging [(2D-FISP 70/13/15°) (TR/TE/flip angle)]. Images represent a point in midsystole (*left*) and middiastole (*right*). Pulsatile CSF flow is seen within anterior of third ventricle and pontine cistern. CSF flow is rapid in anterior and slow in posterior of third ventricle, and absent through aqueduct.

E. Follow-up sagittal midline T1-weighted image (SE 450/15/2) performed after 4 years shows no changes in ventricular size and tectal mass.

ed images (Fig 1B). The patient underwent continuous intracranial pressure monitoring using a fiberoptic extradural device. Mean pressure was less than 12 mm Hg and no abnormal waves were detected, establishing a final diagnosis of arrested hydrocephalus. According to these findings, and in the absence of clinical evidence of hydrocephalus, the patient was not considered to be a candidate for shunt operation. A final diagnosis of benign tectal glioma was established after MR imaging showed no changes during a 4-year follow-up (Fig 1E).

Case 2

A 28-year-old woman was admitted to the neurosurgical department to evaluate possible shunt treatment for hydrocephalus. Seven years earlier, she had suffered a closed head injury with a Glasgow coma scale of 13 at admission. An emergency cranial CT scan showed severe hydrocephalus, but the patient was not considered a candidate for ventricular shunting or any other radiological test. The patient complained of intermittent, mild headache over the 7 years, therefore a new evaluation of the hydrocephalus was considered. Cranial MR imaging performed during the admission we describe in this report revealed obstructive hydrocephalus due to aqueductal stenosis (Fig 2A). The size of the ventricular system had not changed from the initial CT scan performed 7 years previously, and no periventricular edema indicative of acute obstructive hydrocephalus was seen. The flow-sensitive phase-contrast cine MR imaging confirmed the obstructed CSF flow through the stenotic aqueduct, and identified an abnormal CSF wave through the floor of the third ventricle behind the tuber cinereum and

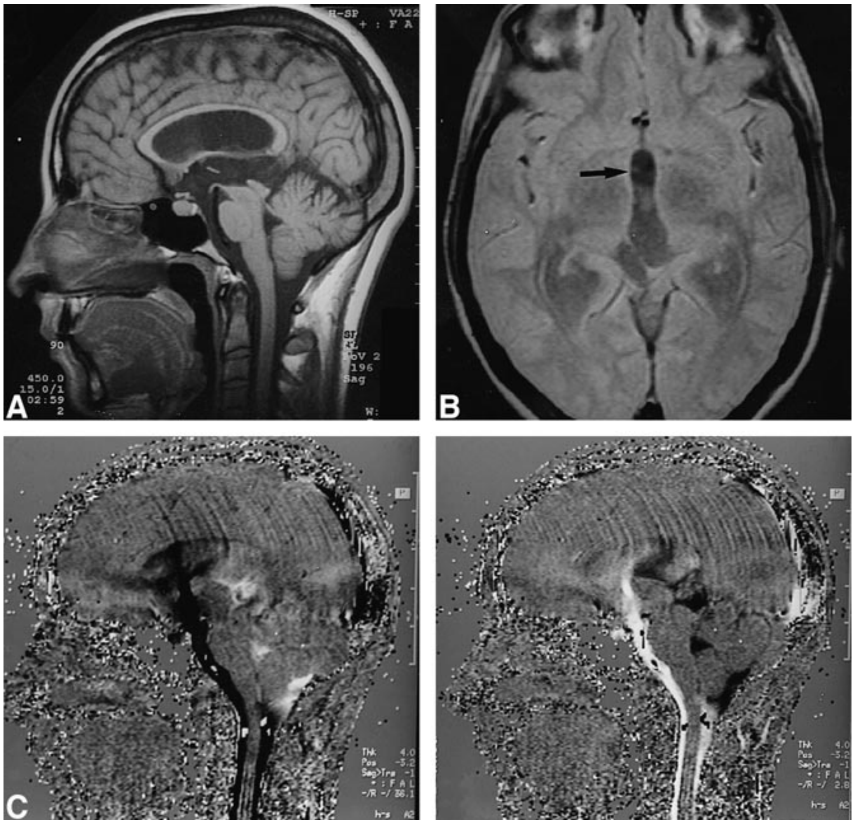

FIG 2. Aqueductal stenosis.

A. Sagittal midline T1-weighted image (SE 450/15/2) reveals moderate ventricular dilatation with diffuse narrowing of aqueduct.

B. Proton-density-weighted axial image (SE 2200/20/1) at level of third ventricle shows abnormal flow-void at anterior recesses of third ventricle (*arrow*).

C. Sagittal midline phase-contrast MR imaging (2D-FISP 70/13/15°). A midsystolic image (*left*) shows caudal flow (*black signal*) at level of floor of third ventricle and prepontine cistern, while in middiastolic image (*right*) flow is reversed (*white signal*), indicating bidirectional flow through floor of third ventricle.

in front of the mammillary bodies (Fig 2B), which connected the ventricular system with the suprasellar and prepontine cisterns. Continuous intracranial pressure monitoring demonstrated a mean pressure less than 12 mm Hg and an absence of abnormal waves, establishing a final diagnosis of arrested hydrocephalus. According to these findings, and in the absence of clinical evidence of hydrocephalus, the patient was not considered to be candidate for shunt operation.

Case 3

A 67-year-old woman was admitted after a minor closed head injury. An emergency CT scan showed no hemorrhagic lesions, but a severe hydrocephalus was detected and subsequently studied by means of MR imaging. This exam showed a supratentorial obstructive hydrocephalus due to aqueductal stenosis without periventricular edema (Figs 3A–B). Flow-sensitive phase-contrast cine MR imaging confirmed the obstructed CSF flow through the stenotic aqueduct and identified an abnormal CSF wave through a gap in the floor of the third ventricle that connected the ventricular system with the subarachnoid spaces (Fig 3C). As the patient was completely symptom-free, intracranial pressure monitoring was not considered. The patient remained asymptomatic after a clinical follow-up of 18 months.

Discussion

Spontaneous ventriculostomy has been described mainly at postmortem examination or with ventriculography, but only rarely with pneumoencephalography (2, 5), and never, to our knowledge, with MR imaging. Previously reported cases were noted to have occurred secondary to obstructive hydrocephalus caused by neoplastic disease or by benign stenosis of the aqueduct, and occasionally congenital stenosis of the foramen of Monro. SV probably occurs more frequently than the literature suggests, and the absence of reported cases in the last two decades simply reflects the routine clinical use of CT, rather than ventriculography and pneumoencephalography, in the diagnosis of ventricular system diseases. The use of MR imaging in our three cases gave the diagnosis of SV, although the condition was not initially suspected using conventional spin-echo MR imaging, even on sagittal and coronal T2-weighted images as performed in case 1 (Fig 1B). With these sequences, it is possible to identify a CSF flow void through the floor of the third ventricle in patients who have a functioning percutaneous ventriculostomy. However, the absence of this finding does not necessarily preclude communication between the third ventricle and the subarachnoid space; false-negative results are a possibility (6). With flow-sensitive phase-contrast cine MR imaging, we were able to identify the obstructed CSF wave across the aqueduct and an abnormal pulsatile CSF wave through a small opening in the floor of the third ventricle, which permitted free communication between the supratentorial ventricular system and the subarachnoid space. The opening consisted of a gap in the thinnest and weakest segment of the floor of the third ventricle located between the median eminence of the tuber cinereum and the mammillary bodies, surrounded by the fornix and mamillothalamic tract posterolaterally and the hypothalamic nuclei laterally (1, 7). This segment of the floor of the third

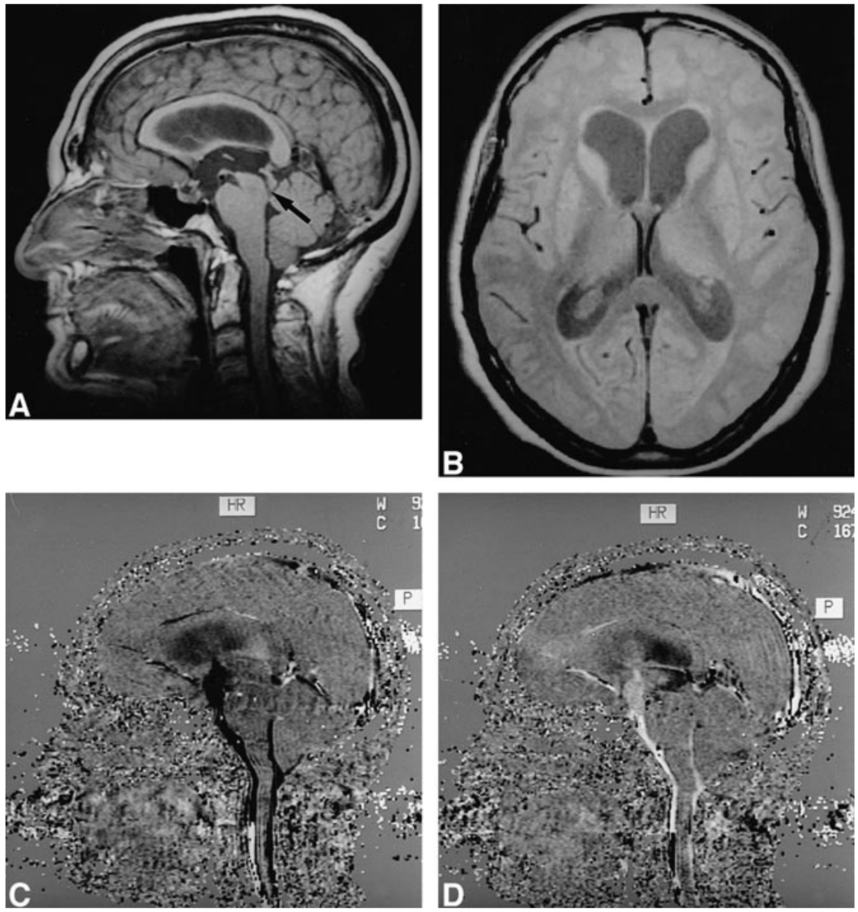

FIG 3. Aqueductal stenosis.

A. Midline sagittal T1-weighted image (SE 550/14/2) in patient with moderate hydrocephalus shows distal aqueductal stenosis (*arrow*).

B. Proton-density axial image (SE 2200/20/1) at level of lateral ventricles shows moderate hydrocephalus but no periventricular edema, indicating acute obstructive hydrocephalus.

C–D. Sagittal midline phase-contrast MR imaging (2D-FISP 70/13/15°). Images represent a point in mid-systole (*C*) and middiastole (*D*). Craniocaudal pulsatile flow is seen through floor of third ventricle.

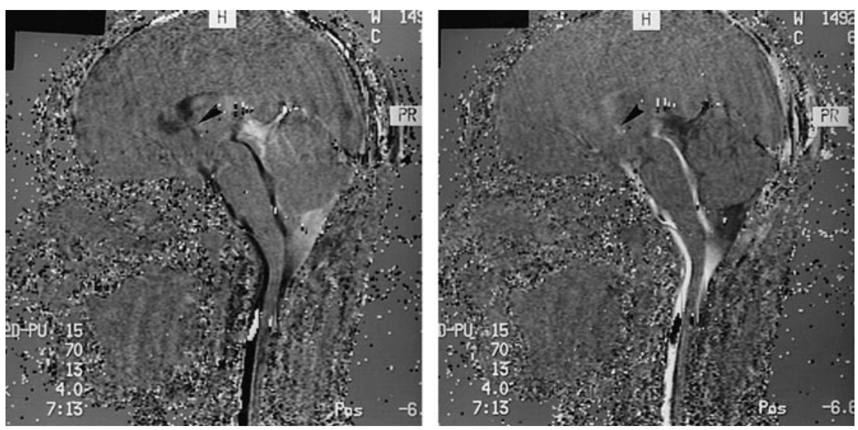

FIG 4. Normal CSF flow cine-MR study. Sagittal midline phase-contrast MR imaging (2D-FISP 70/13/15°). Midsystolic image (*left*) shows caudal flow within foramen of Monro (*arrow*), aqueduct, and fourth ventricle. No CSF flow is seen in anterior of third ventricle. In middiastolic image (*right*), flow is reversed indicating bidirectional flow through ventricular system and subarachnoid spaces. By convention, caudal flow (systolic) is represented in black and cranial flow (diastolic) is represented in white.

ventricle is the area selected by neurosurgeons in which to perform endoscopic ventriculostomy in obstructive hydrocephalus (8). We were able to identify the disruption of the floor of the third ventricle with the standard coronal and sagittal T1-weighted sequences only in retrospect, whereas it was easily identified with flow-sensitive phase-contrast MR imaging. This technique noninvasively depicts the CSF flow in the ventricular system and subarachnoid spaces. The flow is a result of cardiac pulsations transmitted to intracranial arteries and capillaries, causing a specific series of CSF and brain parenchyma displacements. In normal conditions, CSF shows an alternating upward (diastolic) and downward (systolic) motion in each cardiac cycle that can be recognized and measured in the ventricular system, cisterns, and subarachnoid spaces (9) (Figs 4A–B). In healthy persons, antegrade flow of CSF is initiated at the foramen of Monro at 8% of the cardiac cycle. The wave of CSF flow continues through the aqueduct into the proximal fourth ventricle. At the time CSF is flowing in an antegrade direction through the aqueduct, it is also flowing in an antegrade direction through the pontine cistern and upper cervical subarachnoid space. Onset of retrograde flow is initiated at 42%

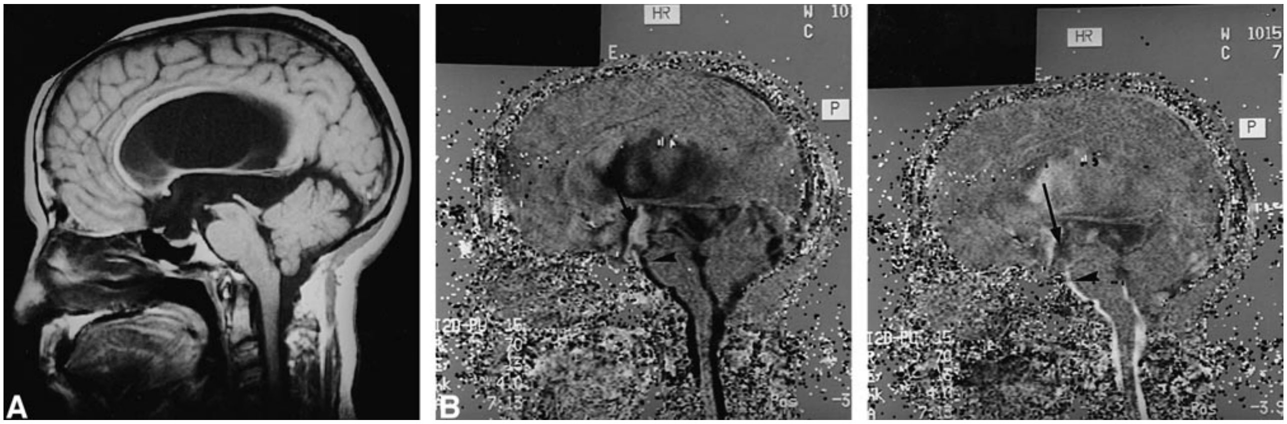

FIG 5. Aqueductal stenosis without spontaneous ventriculostomy in patient with active obstructive hydrocephalus that required ventricular shunting.

A. Sagittal midline T1-weighted image (SE 450/15/2) reveals severe ventricular dilatation with distal narrowing of aqueduct.

B. Sagittal midline phase-contrast MR imaging (2D-FISP 70/13/15°). Images represent a point in midsystole (*left*) and middiastole (*right*). Craniocaudal pulsatile flow is not seen through aqueduct. Note asynchronic CSF flow between anteroinferior part of third ventricle (*arrow*) and prepontine subarachnoid spaces (*arrowhead*).

of the cardiac cycle, first in the posterior cervical subarachnoid space and lower fourth ventricle, followed by retrograde flow starting in the aqueduct at 54% of the cardiac cycle, and then in the anterior cervical subarachnoid space (9). In healthy subjects, ventricular CSF systole and diastole are constantly slightly out of phase with CSF systole and diastole in the subarachnoid space (10). Flow-sensitive phase-contrast MR imaging has been used to demonstrate CSF motion in healthy individuals and in patients with communicating or obstructive hydrocephalus, and it permits the reassessment of our understanding of human physiology (9, 11) and allows for a better method of selecting those patients with chronic communicating hydrocephalus who would benefit most from shunt implantation (9), as well as for the functional analysis of percutaneous endoscopic third ventriculostomy patency (11). A previous report of flow-sensitive phase-contrast MR imaging in aqueductal stenosis demonstrated absence of aqueductal flow, but no apparent CSF flow, across the floor of the third ventricle connecting the ventricular system and the subarachnoid spaces (10). We obtained the same findings in other patients with aqueductal stenosis (Fig 5).

Although the final decision to shunt is made on the basis of the patient's symptoms and the information obtained by continuous intracranial pressure monitoring (4, 12), flow-sensitive phase-contrast MR imaging could be useful for initial selection of those patients with hydrocephalus who might require an additional, more invasive test to arrive at a definitive treatment decision.

The three cases reported are of adults diagnosed with obstructive hydrocephalus due to aqueductal stenosis, without significant related symptomatology, and with normal intracranial pressure monitoring, who were given the final diagnosis of arrested hydrocephalus not requiring ventricular shunting (13). The higher incidence of this rare condition in adults suggests an acquired rupture of the thinnest segment of the floor of the third ventricle as a result of long-standing CSF pulsations against it due to chronic obstructive hydrocephalus. In this condition, the floor of the third ventricle is thinned out to a transparent membrane that pulsates with each heart beat, as has been demonstrated in endoscopic examination of the ventricular system (8). This spontaneous communication has not been described in acute obstructive hydrocephalus, probably because the normal elasticity of the ventricular walls prevents their rupture when there is a rapid increase of intraventricular pressure.

SV through the floor of the third ventricle can be easily identified with the use of flow-sensitive phase-contrast MR imaging, which shows the same findings as those described for normal functioning percutaneous endoscopic third ventriculostomies (6, 12). Identification of this rare condition adds to our understanding of human pathophysiology and should be kept in mind in the preoperative assessment of obstructive hydrocephalus when endoscopic fenestration of the floor of the third ventricle is considered as an alternative to extracranial shunts in noncommunicating hydrocephalus.

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