Sylvian aqueduct syndrome with slit ventricles in shunted hydrocephalus due to adult aqueduct stenosis

Report of 3 cases


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The authors report on 3 patients who developed sylvian aqueduct syndrome (SAS) in the context of shunt dysfunction and slit ventricles. All 3 patients had received shunts for adult onset hydrocephalus due to aqueduct stenosis and were stable for years before presenting with loss of upward gaze, convergence-retraction nystagmus, and slit ventricles, all due to shunt overdrainage. All 3 improved after either shunt revision or a third ventriculostomy procedure. Although it is well known that SAS can be caused by shunt blockage producing a transtentorial pressure gradient, these cases emphasize that an identical clinical pattern can occur with a reverse transtentorial pressure gradient and slit ventricles due to shunt overdrainage. The authors propose a simple management plan for patients with shunted hydrocephalus who develop SAS. (DOI: 10.3171/JNS/2008/109/11/0939)

Key Words • aqueduct stenosis • hydrocephalus • Parinaud syndrome • slit ventricles • sylvian aqueduct syndrome • third ventriculostomy

LESIONS of the dorsal midbrain, rostral to the superior colliculi, produce pathognomonic eye movement abnormalities, which include failure of upward gaze, convergence-retraction nystagmus, lid retraction, skew deviation, and pupillary light-near dissociation. Confusingly, some or all of this constellation of abnormal eye movements have been called, virtually synonymously, Parinaud syndrome, Koerber–Salus–Elschnig syndrome, pretectal syndrome, dorsal midbrain syndrome, paraaqueductal syndrome, and SAS, which we use in this paper. It has long been recognized that obstructive hydrocephalus, usually due to aqueduct stenosis, can present with SAS, which resolves with effective shunting. It is also recognized that in patients with shunted hydrocephalus the development of SAS usually indicates shunt malfunction with large ventricles and high pressure due to underdrainage. The underlying pathophysiological mechanism in these cases appears to be a pressure differential across the tectum, with supratenorial pressure higher than infratentorial pressure.

What has not yet been fully recognized is that in patients with shunted hydrocephalus, SAS can also occur with slit ventricles and low pressure due to shunt overdrainage. In this paper we report on 3 such patients; in all 3 cases the supratenorial pressure was measured and confirmed to be low or negative. In 2 patients the SAS resolved after an endoscopic third ventriculostomy procedure and in the third patient after insertion of an antisiphon device. We propose that in these patients the mechanism responsible for the development of SAS was also a pressure differential across the tectum, with supratenorial pressure lower, not higher, than infratentorial pressure. We outline an algorithm for the diagnosis and treatment of patients with shunted hydrocephalus who develop SAS.

Case Reports

Case 1

This 61-year-old man presented to one of the authors in 2005 with a 3-month history of multiple discrete episodes of confusion, unsteadiness, and diplopia. He vomited during one of these episodes but headache was not
a feature of his presentation. The symptoms were worse when he was in the upright position and better when he was recumbent. Neurological examination revealed a confused man, disoriented to time and place. He had bilateral lid retraction, upward gaze paralysis, and convergence-retraction nystagmus. His pupillary reflexes were normal and his optic discs were flat.

He had originally presented elsewhere in 2000 with intermittent symptoms of headache, ataxia, dizziness, and vomiting. An MR imaging study at the time revealed hydrocephalus due to aqueduct stenosis, and a VP shunt was inserted. The patient's symptoms resolved following the shunt insertion and subsequent CT scans revealed small ventricles. The patient had a history of childhood meningitis, during which the aqueduct stenosis may have been acquired.

After a few hours in the upright position, the patient underwent a brain CT scan that showed slit ventricles (Fig. 1A), but MR imaging (Fig. 1B) performed after 24 hours in the recumbent position showed enlarged ventricles without any other abnormality, specifically in the midbrain. Intracranial pressure monitored using an independent Rickham reservoir (Integra LifeSciences Corp.) was consistently between −2 and +5 mm Hg (−27 and +68 mm H2O) while the patient was recumbent. A CT ventriculogram confirmed aqueduct stenosis. His symptoms were believed to be due to shunt overdrainage. His non-functioning Delta valve (Medtronic) was replaced with a programmable Hakim valve (Codman) with an antisiphon device. His SAS completely resolved within 48 hours and a postoperative CT scan revealed normal-sized ventricles. Three years later his symptoms have not recurred.

Case 2

This 53-year-old man presented to one of the authors in 2003 with a 9-month history of horizontal diplopia, more marked when looking to the left. He had no headaches. He had originally presented elsewhere in 1989 with 3 weeks of headaches after a minor head injury during a baseball game; MR imaging showed hydrocephalus due to aqueduct stenosis. A VP shunt was inserted and was revised 1 year later when he presented with headaches due to a blocked shunt. He remained well for the next 10 years with serial imaging revealing slit ventricles.

In 1999 he was examined, at another institution, for loss of motivation and memory. At the time, he was noted to have absent upward gaze and “diplopia consistent with a left fourth nerve palsy.” The MR imaging study at the time again showed only slit ventricles. Depression was diagnosed and the eye movement abnormalities were attributed to the original head injury.

In 2003 we found not only loss of upward gaze but also convergence-retraction nystagmus, most obviously elicited by a downwardly directed optokinetic stimulus. His pupillary reflexes were normal and his optic discs were flat. Brain MR imaging (Fig. 2A) again showed slit ventricles, unchanged since 1999, and no other abnormality, specifically in the midbrain.

Intracranial pressure monitored using an independent Rickham reservoir revealed pressures of < 5 mm Hg (68 mm H2O) indicating that the shunt was overdraining. A CT ventriculogram confirmed the aqueduct stenosis. The shunt was revised with a Codman Medos Hakim programmable valve. Adjustment of the valve failed to normalize ventricular size; the ventricles would be either slit (Fig. 2B) or dilated (Fig. 2C) with only 10–20 mm H2O changes in valve opening pressure. The patient became unconscious when the ventricles were dilated and rapidly improved with adjustment of the valve.

To equalize the pressures between the supratentorial and infratentorial compartments it was decided that an en-
Endoscopic third ventriculostomy would be performed. The shunt was left in situ but was programmed to the highest setting (200 mm H₂O). Following the third ventriculostomy, the patient’s diplopia quickly resolved and his upward gaze improved. The ventricles were now midsized (Fig. 2D). Three years following the endoscopic procedure his SAS has resolved.

Case 3

This 24-year-old man presented to one of the authors in 1999 with persistent headache and diplopia. He had originally presented at another institution in 1998 with headaches and was found to have enlarged ventricles. Although only the lateral and third ventricles were enlarged, a dynamic MR imaging study was inconclusive concerning cerebrospinal fluid flow in the aqueduct. Intracranial pressure monitoring showed normal baseline pressures with peaks as high as 20 mm Hg (270 mm H₂O). Because the patient had had meningitis at 4 years of age, he was believed to have communicating hydrocephalus. A VP shunt was inserted, after which his headaches resolved. He presented 1 year later, again with headaches, and was found to have a disconnected shunt, which was revised, but his headaches were only partly relieved. Imaging studies at that time revealed small ventricles. Over the next 12 months his headaches became more frequent and he noted occasional diplopia.

On neurological examination in 1999 we found absent upward gaze above the horizontal meridian and convergence-retraction nystagmus. His pupillary reflexes were normal and his optic discs were flat. Magnetic resonance imaging showed unilateral ventricular collapse but no midbrain abnormality (Fig. 3A). Intracranial pressure monitored using an independent Rickham reservoir was consistently < 10 mm Hg (150 mm H₂O). A CT ventriculogram and dynamic MR imaging showed aqueduct stenosis (Fig. 3D). His symptoms were presumed to be due to shunt overdrainage, so the shunt was revised using a high-pressure valve with an antisiphon device. After 24 hours he developed more headache and became somnolent; MR imaging now showed ventriculomegaly (Fig. 3B) with ICPs of 5–10 mm Hg (68–135 mm H₂O). At this stage an endoscopic third ventriculostomy was performed. Over the next month his headaches and SAS completely resolved, and his MR imaging results showed normal midsized ventricles (Fig. 3C) with a patent third ventriculostomy (Fig. 3E).

Discussion

Sylvian aqueduct syndrome and even more extensive rostral midbrain dysfunction can occur in patients with shunt malfunction in hydrocephalus, specifically when the underlying cause is aqueduct stenosis. Cinalli and associates reported a retrospective analysis of 28 such patients. All had triventricular enlargement at the time they had SAS; 6 of the 28 had MR imaging and all showed anatomical deformation and focal hyperintensities in the midbrain. Supratentorial and infratentorial pressures were measured simultaneously in 6 patients, and supratentorial pressure was higher than infratentorial pressure; that is, there was
a transtentorial pressure gradient in all 6, although the solutrentorial pressure was actually normal in 3 patients. On this basis the investigators proposed that in patients with shunted hydrocephalus and SAS there is a transtentorial pressure differential, creating mechanical stress on the midbrain tectum, and they recommended a third ventriculolostomy procedure to equalize the pressures between the supratentorial and infratentorial compartments.

Although it is clear that a transtentorial pressure gradient can somehow cause midbrain dysfunction, specifically SAS, it might be that the gradient could be in either direction. Cinalli et al. noted that 4 of the patients deteriorated after shunt revision and supratentorial pressure was normal or low in all 4, and that in the 3 patients in whom the pressure recordings were repeated after shunt revision, the transtentorial pressure gradient had reversed in all 3. In a retrospective analysis of the symptoms of low ICP in 14 patients with shunted hydrocephalus, Foltz and Blanks reported that at least 1 patient had SAS, and Shallat and associates reported 2 patients with slit ventricles after shunt insertion who developed SAS.

Our 3 patients presented with adult onset hydrocephalus due to aqueduct stenosis and were treated with VP shunts, after which they remained well for 1–15 years before becoming symptomatic with SAS in the context of slit ventricles. Low or negative supratentorial ICP was confirmed in each patient using a Rickham reservoir. In each case a Rickham reservoir was used to measure the pressure and to avoid a further operation when inserting a fiberoptic monitor. In all cases the pressure trace was good and the presumption was made that the pressures were accurate. We propose that the SAS in our patients was also caused by a transtentorial pressure gradient, but with pressure in the supratentorial compartment lower rather than higher in the infratentorial compartment. We note that just as overdrainage from lumboperitoneal shunting can produce a pressure gradient and brain shift at the foramen magnum, it appears that overdrainage from VP shunting can produce a similar pressure gradient at the tentorial notch.

After confirming the “low” supratentorial pressure and aqueduct stenosis we revised the ventricular shunts, raising the opening pressure of the valve to increase supratentorial pressure and thereby lowering the pressure gradient across the tectum. The ventricular compliance, however, was such that the ventricles were either slit or grossly dilated in 2 of the patients (Cases 2 and 3), making incremental or empirical changes to shunt-valve opening pressure ineffective at resolving the clinical problem.

There is some evidence of differences in ventricular compliance between dogs with aqueduct stenosis and those without. In Case 3, the measured ICP was normal (5–10 mm Hg or 68–135 mm H2O) when the ventricles were dilated after increasing shunt-valve opening pressure, but the patient became unconscious due to low or negative pressure hydrocephalus. By using the Codman Medos Hakim programmable valve in Case 2 and a medium-high pressure Hakim valve in Case 3, it was possible to enlarge the ventricles to enable an endoscopic third ventriculostomy to be performed. The patient in Case 1 was the only one to respond to raising the opening pressure of the shunt valve. In this patient the ICP was gravity-dependent: the ventricles were slitlike and overdrained when the patient was upright, and enlarged and underdrained when he was recumbent. This patient was able to tolerate enlarged ventricles without symptoms, unlike the patients in Cases 2 and 3 who were very sensitive to ventricular enlargement.

How does a transtentorial pressure gradient cause dorsal midbrain dysfunction, specifically SAS? In patients with underdrainage and a high supratentorial pressure, there does appear to be anatomical deformation due to axial enlargement of the third ventricle and T2 signal change in the midbrain, but this was not true in our patients. Furthermore, whereas the nuclei and pathways controlling vertical eye movements, the pupils, and the eyelids are located in the dorsal midbrain, it is not obvious how dysfunction and even lesions of the area produce the specific functional pattern found in SAS. Although it is known that the rostral interstitial nucleus of the medial longitudinal fasciculus controls vertical saccadic eye movements and that the interstitial nucleus of Cajal is the vertical oculomotor integrator, its not easy on this basis to explain how dysfunction of these 2 structures could produce selective absence of all upward, not downward, eye movements and only above the horizontal meridian.

We therefore suggest a management plan for patients with shunted hydrocephalus who develop SAS, consisting of ICP monitoring to check supratentorial pressure followed by ventriculography to confirm the diagnosis of aqueduct obstruction. Both of these tests are important because slit ventricles with raised ICP and underdrainage (due to shunt obstruction) as well as low ICP with overdrainage can both cause SAS. Shunt revision should be undertaken with a programmable valve to increase opening valve pressure gradually. If the compliance of the ventricles allows a gradual increase in ventricular size with increasing valve resistance, then the pressure differential across the supratentorial and infratentorial compartments should equalize and might resolve the clinical problem. If the ventricular compliance is such that the ventricles are either slit or enlarged and causing symptoms of hydrocephalus, then the only treatment is a third ventriculostomy with the goal of shunt independence. For these reasons, a third ventriculostomy should be considered as a first-line treatment of aqueduct stenosis.

Disclaimer
The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper.

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Accepted December 7, 2007.

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