

## A Parkinsonism as a Component of Sylvian Aqueduct Syndrome : Effect of Floating Cranioplasty and Distal Catheter Elongation

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The sylvian aqueduct syndrome is a global rostral midbrain dysfunction induced by a transtentorial pressure gradient through the aquaeductus. Several months after ventriculoperitoneal shunt, a patient with hydrocephalus began experiencing a constellation of midbrain dysfunction symptoms, including bradykinesia, medial longitudinal fasciculus syndrome, third nerve palsy, and mutism. These were indicative of cerebral aqueduct syndrome. In addition, the patient showed posture-dependent underdrainage or overdrainage. All symptoms were resolved after distal catheter elongation and floating cranioplasty. We present a case of reversible parkinsonism, which developed in a patient with shunted hydrocephalus and aqueductal stenosis, and discuss the diagnosis and treatment of the sylvian aqueduct syndrome. We also review the literature to address problems of drainage and potential treatment modalities.

**KEY WORDS :** Sylvian aqueduct syndrome · Drainage failure · Floating cranioplasty · Distal catheter elongation · Parkinsonism.

### Introduction

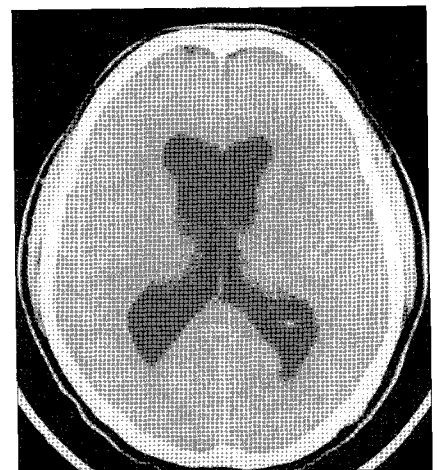
According to the definition by Barrer and colleagues<sup>2)</sup>, the sylvian aqueduct syndrome is a global rostral midbrain dysfunction induced by a transtentorial pressure gradient through the aquaeductus. When observed during shunt malfunction, it may be associated with a more complex clinical picture. The sylvian aqueduct syndrome is progressive and may be life-threatening in the absence of appropriate treatment<sup>11)</sup>. Currently, the underlying pathophysiological mechanism is not fully understood.

We report a case of parkinsonism as a component of the sylvian aqueduct syndrome, which was resolved without the need for antiparkinsonian drugs after distal catheter elongation with floating cranioplasty<sup>10,13)</sup>. Neurosurgeons generally focus on the structures around the perimesencephalic subarachnoid space and not on the aquaeductus. In this case, we focused on the vulnerable aquaeductus for the transtentorial pressure gradient.

### Case Report

#### History

A 65-year-old man sustained minor head trauma in a traffic accident. Computed tomography(CT) revealed triventriculomegaly without symptoms of hydrocephalus (Fig. 1). In the absence of symptoms, the patient received no treatment for ventriculomegaly.



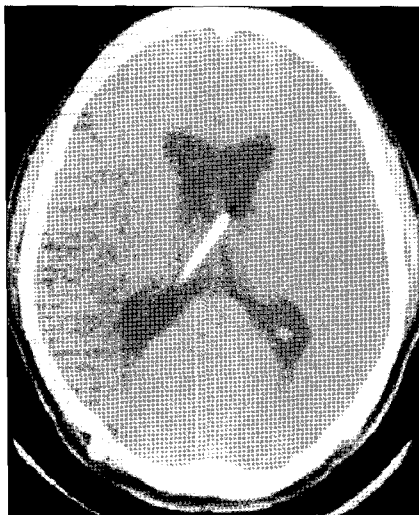
**Fig. 1.** Brain computed tomography after traffic accident showing ventriculomegaly without hydrocephalus symptom. Bifrontal index=40.

• Received : September 1, 2005 • Accepted : December 8, 2005  
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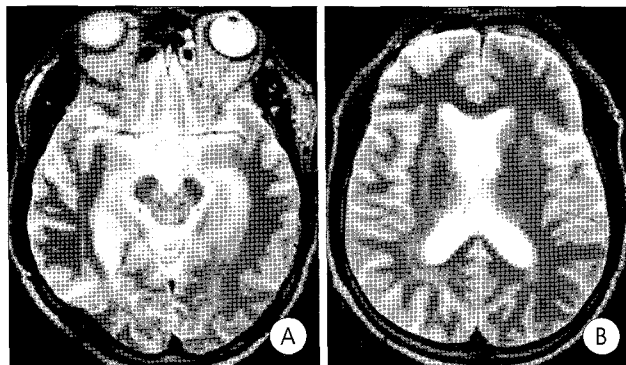
**Table 1.** Changes of symptom, ventricle size, ventricular pressure by time table

	Symptom	Pcsf	BFI	Diagnosis	Pset
PRO 20 months	Concussion	–	40	Ventriculomegaly	–
PRO 35 days	Ataxia, dysarthria	190	42	Hydrocephalus	–
Operation	Ventriculoperitoneal shunt	190	–	–	160
PO 7 days	Symptom improved	160	38	–	140
PO 8 months	Diplopia, headache	–	30	Ischemic CVD	–
PO 9 months	Parkinsonism, ptosis	–	12	–	–
PO 10 months	Parkinsonism, hypersomnia, upward gaze limitation	–	–	Parkinsonism	–
PO 1 year	Akinetic mutism	100	40	–	90
PO 1 year 4 days	Hypokinesia	90	11	Slit ventricle	150
PO 1 year 5 days	Hypokinesia	150	30	–	–
PO 13 months (ROP)	Distal catheter elongation and floating cranioplasty	–	11	Sylvian aqueduct syndrome	170
ROP 10 months	No neurological problem	–	–	Slit ventricle	–

Pcsf : lumbar cisternal or valve pressure, BFI : bifrontal index, PVL : periventricular lucency, SDF : subdural fluid or hematoma collection, Pset : setting pressure, PRO : preoperative, OP : operative, PO : postoperative, ROP : reoperative, CVD : cerebrovascular disease



**Fig. 2.** Postoperative six-day brain computed tomography showing decreased size of ventricles under setting pressure of 160mm cerebrospinal fluid.



**Fig. 3.** Postoperative eight-month brain magnetic resonance imaging under setting pressure of 140mm cerebrospinal fluid showing no significant abnormal findings in parenchyma (A) or ventricles (B), with symptom of headache and diplopia. Diagnosed with ischemic pontine cerebrovascular disease.

Over the next two years, the patient experienced progressive symptoms of dizziness and gait and speech difficulties. Neurological examination showed ataxia and dysarthria and slightly increased deep tendon reflexes. A cranial CT scan revealed slightly increased ventricle size with sulcal effacement. The lu-

mbar cisternal pressure was 190mm CSF. Routine laboratory examinations were within normal limits. There was no definite history of cerebrovascular accident or hypertension. The diagnostic and therapeutic courses were summarized in Table 1.

### Operation

A ventriculoperitoneal (VP) shunt with a Codman Hakim programmable valve (Codman & Shurtleff, Inc., Raynham, MA) was performed two years after the patient's traffic accident. The ventricle was approached via the right occipital route. The initial valve pressure was

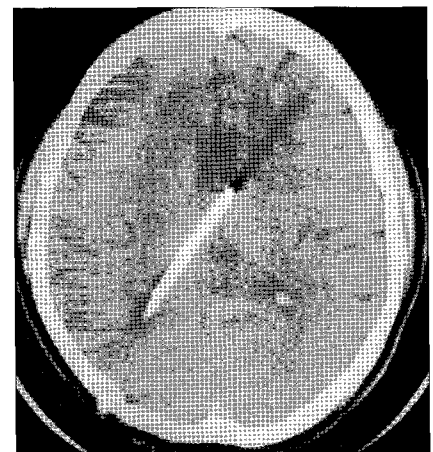
set at 160mm CSF and gradually adjusted down to 140mm CSF at postoperative day six (Fig. 2).

### Postoperative course

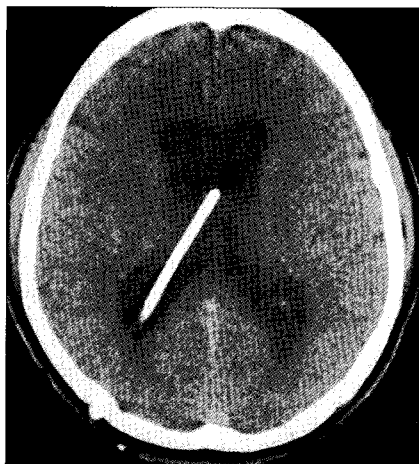
The patient was in good health until diplopia and headache developed eight months after shunting. Magnetic resonance imaging and angiography showed no definite abnormalities in the size or contour of the ventricles, in the brain parenchyma, or in the cerebral vasculature. Clinically, the patient was diagnosed with ischemic type pontine cerebrovascular disease (Fig. 3). After conservative treatment, his symptoms were relieved. However, at nine months postoperatively, bradykinesia and ptosis developed. He was diagnosed with parkinsonian syndrome, with overdrainage of the shunt system at 100mm CSF on CT imaging (Fig. 4). At one year, he experienced confusion, and a CT scan revealed underdrainage at 100mm CSF. Thereafter, the valve pressure was adjusted according to the size of the ventricle with concomitant partial effects. We realized it as a posture-dependent drainage problem for the first time (Fig. 5, 6).

### Reoperation

Thirteen months after shunting, the patient was di-



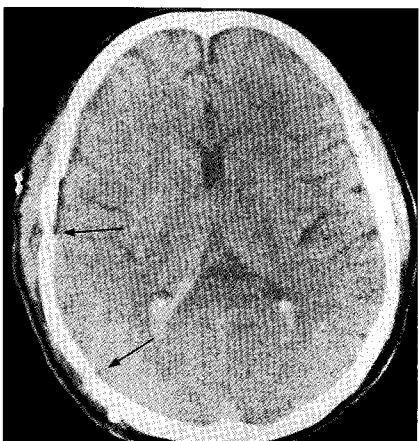
**Fig. 4.** Postoperative nine-month brain computed tomography under the setting pressure of 100mm cerebrospinal fluid showing slit ventricle. Parkinsonism was diagnosed with ptosis and bradykinesia.



**Fig. 5.** Brain computed tomography showing underdrainage under the valve pressure of 100mm cerebrospinal fluid after 20 minutes lying down. Bifrontal index=40.

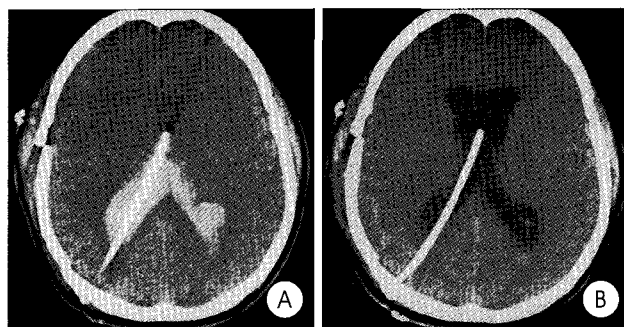


**Fig. 6.** Brain computed tomography after 20 minutes standing up under the valve pressure of 90mm cerebrospinal fluid showing slit ventricle with mutistic bradykinesia. Erroneously diagnosed with slit ventricle syndrome.

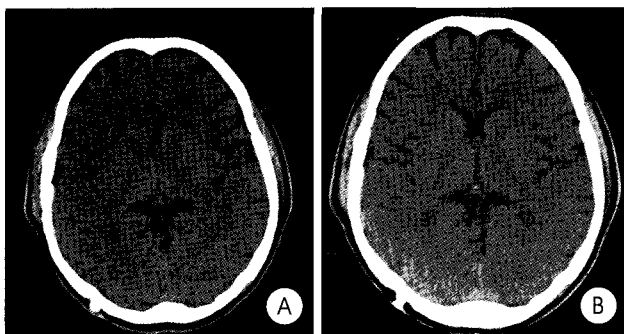


**Fig. 7.** Immediate post-reoperative brain computed tomography showing slit ventricle. The floating bone is also visible (arrow).

agnosed with Sylvian aqueduct syndrome with a drainage problem. Accordingly, we decided to perform a shunt revision, involving distal catheter elongation for overdrainage and floating cranioplasty, replacing the craniectomized bone without fixation, for increasing the compliance (Fig. 7). Postoperatively, an immediate subtle effect was followed by a semicomatose mentality at two days, and a CT scan revealed whole ventricular hemorrhage. At ten days, a CT scan showed scanty intraventricular hemorrhage with slightly increased lateral ventricle size (bifrontal index = 38)(Fig. 8). At two months postoperatively, a CT scan revealed slight underdrainage or overdrainage of the shunt system relative to the patient's body posture. The patient was advised to avoid sudden movements from a lying position to an upright position. Over the next ten months, all of the patient's



**Fig. 8.** Brain computed tomography on two days after operation showing whole intra ventricular hemorrhage (A), and on ten days after operation showing almost resolved hemorrhage with ventricular dilatation (B).



**Fig. 9.** Brain computed tomography of ten months after reoperation under the valve pressure of 100mm cerebrospinal fluid after upright for two hours (A) and then after lying down for two hours (B), showing slit ventricle. Note slightly smaller lateral ventricle after upright than after lying down.

symptoms slowly resolved. The patient was discharged and returned to his active life, without ataxia, headache, parkinsonian syndrome, subtle midbrain dysfunction, or need for any medication. MRI and CT scanning revealed slit lateral ventricle syndrome, enlarged subarachnoid spaces, and no periventricular lucency. However, the patient showed minimal posture-dependent ventricle changes (Fig. 9).

## Discussion

As one of the symptoms of hydrocephalus, the cerebral aqueduct syndrome is characterized by midbrain dysfunction syndrome, which involves parkinsonian syndrome, third nerve palsy such as ptosis, diplopia, and akinetic mutism. The cerebral aqueduct syndrome is responsible for significant anatomical distortion of the structures located at the level of the tentorial hiatus, which is well tolerated because of the slow progression of the aqueductal obstruction.

Suzuki and colleagues<sup>19)</sup> reported the resolution of eyelid dysfunction by cerebrospinal fluid diversion and suggested that chronic hydrocephalus was involved in the development of ptosis after minor head injury. Our patient presented with the symptom of hydrocephalus two years after a minor traffic accident.

A sudden change in cerebrospinal fluid pressure at the time of injury may have induced functional impairment at the level of the vulnerable periaqueductal structures, which barely withstood the longstanding ventriculomegaly, resulting in the clinical features observed in our patient.

### Pathophysiological theories

Before the identification of the anatomical structure responsible for vertical gaze, some authors proposed that cystic dilation of the suprapineal recess with herniation in the quadrigeminal cistern and dilation of the upper portion of the aqueduct rostral to the obstruction could distort and compress the tectal plate in which the center for vertical gaze was thought to be located<sup>6,9,14,16-18</sup>.

However, most of these theories have been revised since the identification of the anatomical structure responsible for upward gaze, which is located in the periaqueductal gray matter ventral to the aqueduct in the dorsal interstitial nucleus of the medial longitudinal fasciculus<sup>3</sup>.

In communicating hydrocephalus, the CSF hydrostatic pressure is equally distributed throughout all the compartments of the central nervous system<sup>15</sup>. There is no development of abnormal stress within the brain parenchyma. Thus, CSF pressure may reach high levels, producing only symptoms of intracranial hypertension without any other focal neurological signs.

During ICP measurement in patients with aqueductal stenosis, Cinalli and colleagues<sup>9</sup> reported very interesting results. Simultaneous recordings of supratentorial and infratentorial pressures before shunt revision revealed a higher supratentorial pressure in all cases, with a transtentorial pressure gradient ranging from 4 to 10mm Hg (mean, 6mm Hg). The general patterns of the recordings were deeply altered, and, during sleep, the pattern of paradoxical sleep was lost. Pressure waves were sometimes observed in a plateau configuration. They found no correlation between ICP levels and the severity of the clinical condition. They also found diminished compartmental pressure difference after shunting or third ventriculostomy.

### Therapeutic management

Endoscopic third ventriculostomy (ETV) may be an ideal solution for drainage problems associated with aqueductal stenosis because it converts the bicompartamental brain to a unicompartamental brain between supra- and infratentorial compartments. However, according to Tisell and colleagues<sup>20</sup>, the long-term effectiveness of ETV for adult patients with noncommunicating hydrocephalus was sufficient in only 50% of the cases. These cases demonstrate that VP shunt may be a suitable therapeutic option for cases of hydrocephalus and aqueductal stenosis.

Czosnyka and colleagues<sup>5</sup> reported that the rate of compli-

cations related to overdrainage may be reduced by distal catheter elongation, which increases the resistance of these valves by 100% to 200%. In the present study, we performed distal catheter elongation for an overdrainage problem with adequate success. This should be the preferred therapeutic procedure for preventing the siphon effect.

Allan and Chaseling<sup>11</sup> reported two cases of diagnosed slit-ventricle syndrome and elevated ICP. Each patient underwent ICP monitoring before and after subtemporal decompression. They presented the dramatic changes in the ICP measurements along with findings from one-year follow-up examinations. Heisey and colleagues<sup>8</sup> proposed not only that the total cranial compliance depends on the mobility of the intracranial fluid volumes of blood and cerebrospinal fluid when there is an increase in intracranial volume (ICV) but also that it varies as a function of cranial compliance attributable to the movement of the cranial bones at their sutures. Floating cranioplasty, in our terminology, is a good substitute for subtemporal craniectomy, which is associated with problems of disfigurement<sup>7,12</sup>.

Initially, our patient presented with difficulties in gait and speech, typical symptoms of hydrocephalus. His past medical history was suggestive of aqueductal stenosis without symptoms. VP shunt with immediate symptom relief was followed by several cycles of overdrainage or underdrainage during subsequent months, which was misdiagnosed as ischemic pontine cerebrovascular disease or parkinsonian syndrome. We did not recognize the sylvian aqueduct syndrome with complicated shunt problems until thirteen months after VP shunt. To address this drainage failure, we performed distal catheter elongation to correct overdrainage and floating cranioplasty to increase the compliance. The underdrainage problem was assumed to be controlled by lowering the valve pressure. Although the shunt revision was complicated by intraventricular hemorrhage, the patient was able to return to a normal life, without headache, midbrain dysfunction, parkinsonism, or the need for antiparkinsonian medication.

We report the case of a patient with a constellation of midbrain dysfunction symptoms, including parkinsonian syndrome, indicative of the sylvian aqueduct syndrome. Long standing, unrecognized, posture-dependent drainage failure associated with midbrain dysfunction was finally controlled by distal catheter elongation and floating cranioplasty.

### Conclusion

In the treatment of patients with hydrocephalus, achieving a balance between intracranial pressure and volume is critical. To prevent the sylvian aqueduct syndrome, neurosurgeons should concentrate on symptoms of midbrain dysfunction pre- and post-shunt therapy. Midbrain dysfunction in the sylvian

aqueductal syndrome, including parkinsonism, maybe reversible by third ventriculostomy, distal catheter elongation, subtemporal decompression, or floating cranioplasty.

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## Commentary

With a literature review, the authors presented a complicated case of sylvian aqueductal syndrome developed in a patient with aqueductal stenosis after ventriculo-peritoneal shunt. The patient suffered from dorsal midbrain dysfunction symptoms such as third nerve palsy, MLF syndrome, mutism, and bradykinesia during the follow-up period after VP shunt, which were caused by viciously-cycled shunt malfunction and overdrainage. They recommended the elongation of the distal catheter of VP shunt device for the prevention of over-drainage and what they called, "floating cranioplasty" for increasing the compliance.

I think this paper is very interesting and informative in the management of complicated case of hydrocephalus associated with aqueductal stenosis especially in old ages. The complex syndrome composed of upward gaze palsy, retraction-convergence nystagmus, lid retraction, sun-set eye sign, and abnormalities on convergence, known as "Parinaud's syndrome" has been also called as various names of Koerber-Salus\_Elschnig, dorsal midbrain syndrome, pretectal syndrome. sylvian aqueduct syndrome, When the shunt malfunction was affected by aqueductal stenosis in old ages, it is well known that this syndrome can be associated with more complex features such as Parkinsonian syndrome, akinetic mutism, memory disturbances, staring gaze, pyramidal signs, and coma. This clinical entity was defined by Barrer, et al. as "global rostral midbrain dysfunction". The pressure difference between supratentorial compartment and infratentorial compartment might be the main cause of the "global rostral midbrain dysfunction". The best way to manage the "global rostral midbrain dysfunction may be endoscopic third ventriculostomy to equalize transtentorial pressure difference. For the patient in this paper I would like to recommend a endoscopic third ventriculostomy or a medium-pressure shunt device with an anti-siphon as an alternative for the patient before choosing elongation of distal catheter with "floating cranioplasty" procedure.

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