—Case Reports—

Suspected Low-Pressure Hydrocephalus Caused by Spinal Drainage after Subarachnoid Hemorrhage

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Hydrocephalus induced by low cerebrospinal fluid (CSF) pressure is extremely rare and sporadically reported. Subarachnoid hemorrhage, head trauma, and spinal drainage were reported to be causative factors for surgical treatment. A 33-year-old man with subarachnoid hemorrhage caused by right vertebral artery aneurysm rupture developed headache. Trapping surgery was performed, and a spinal drain was inserted from L4/5 for subarachnoid hemorrhage washout. On postoperative day 3, subdural fluid accumulation had increased at the posterior fossa craniotomy site and the cerebellar sulci had narrowed; the ventricles were slightly enlarged. The patient reported headache during head elevation. Low-pressure hydrocephalus (LPH) was suspected. After the spinal drain was removed, headache resolved, and cerebral ventriculomegaly disappeared. The subsequent clinical course was good. The patient was discharged 3 weeks after surgery. LPH is a rare disease caused by various factors and is treated by correcting liquorrhea or overdrainage, when present. Otherwise, drainage at negative CSF pressure is necessary. The symptoms and image findings for LPH are similar to those for intracranial hypertension and normal-pressure hydrocephalus. This report describes a suspected case of LPH caused by spinal drainage after subarachnoid hemorrhage and reviews the literature.

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Key words: low-pressure hydrocephalus, low intracranial pressure hydrocephalus, negative-pressure hydrocephalus, subarachnoid hemorrhage, spinal drain

Introduction

Low-pressure hydrocephalus (LPH) is an extremely rare, sporadically reported disease first described by Pang et al. in 1994. Although its developmental mechanism is unclear, subarachnoid hemorrhage, head trauma, posterior fossa surgery, lumbar puncture, and spinal drainage are putative causative factors for surgical treatment.

Some symptoms and image findings for LPH are similar to those for intracranial hypertension and normal-pressure hydrocephalus[1]. Symptoms include disturbance of consciousness, headache, vomiting, and cranial nerve palsy. Image findings are cerebral ventriculomegaly and periventricular edema (similar to intracranial hypertension). We treated a patient at Nippon Medical School Hospital with LPH due to spinal drainage after subarachnoid hemorrhage and report our findings.

Case Description

A 33-year-old man with subarachnoid hemorrhage (World Federation of Neurosurgical Societies classification, grade II) caused by right vertebral artery fusiform aneurysm rupture developed headache (**Fig. 1A, B, C**). The dissecting aneurysm lesion was located at a lower position, almost at the height of the foramen magnum and proximal to the posterior inferior cerebellar artery. Fine branches were observed from the periphery of the aneurysm (**Fig. 1C, D**). Consequently, a craniotomy for trapping was performed. The surgical approach involved a suboccipital craniotomy with an atlas laminectomy, and the lesion was trapped with cerebral aneurysm clips (**Fig. 1E**).

A spinal drain was inserted from L4/5 before surgery. Cerebrospinal fluid (CSF) was drained after surgery, for

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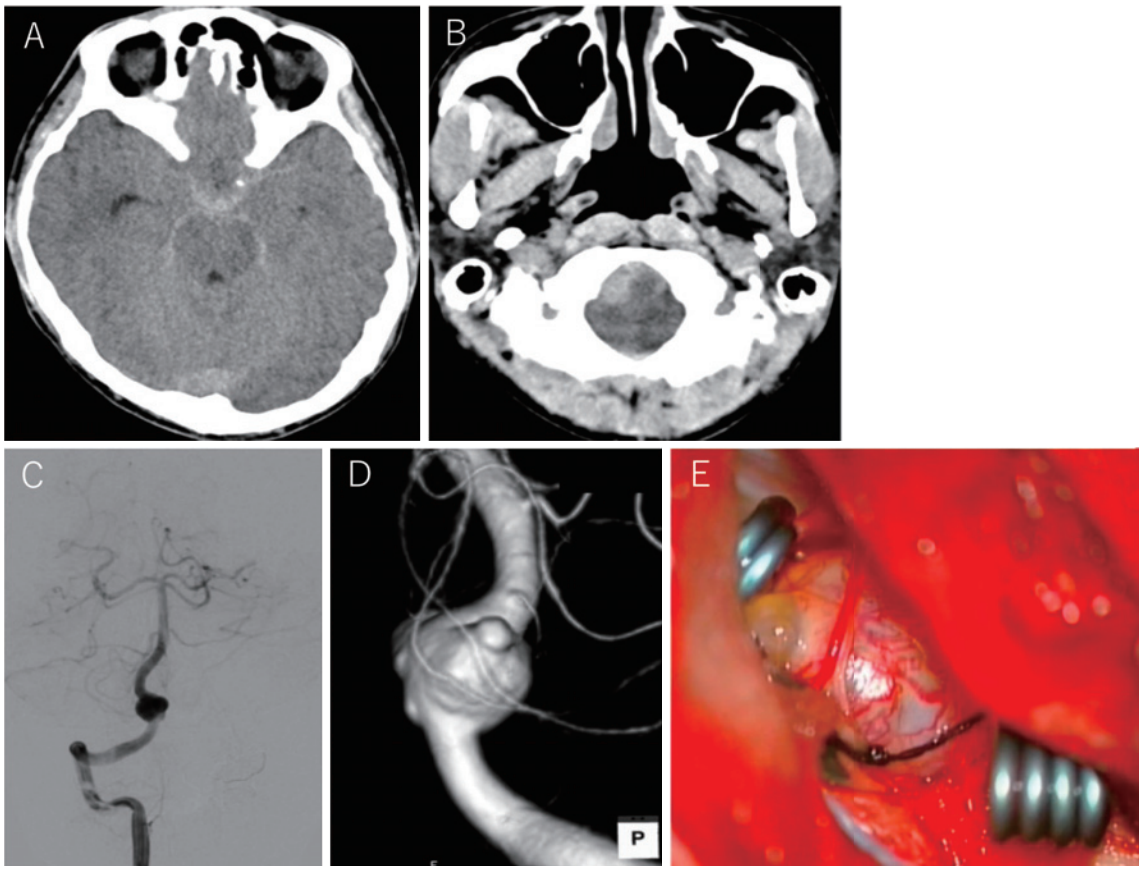

Fig. 1 Preoperative images and intraoperative findings.
(A) (B) Brain computed tomography (CT) images from a 33-year-old man with subarachnoid hemorrhage caused by right vertebral artery fusiform cerebral aneurysm rupture.
(C) (D) Digital subtraction angiography (DSA) and 3D-DSA images. The lesion was located proximal to the posterior inferior cerebellar artery.
(E) A photograph of trapping surgery.

washout of subarachnoid hemorrhage. On postoperative day (POD) 3, cerebral ventriculomegaly appeared, the subdural cavity at the posterior fossa craniotomy site was opened, and fluid accumulation was observed; cerebellar sulci were narrow and tight (**Fig. 2**). In addition, the patient reported headache during head elevation on the same day. Headache induced by head elevation, subdural fluid accumulation, and obscure sulci under spinal drainage were consistent with a diagnosis of low-pressure headache due to spinal drainage (**Fig. 2**). The spinal drain was therefore removed, which promptly relieved headache during head elevation. Furthermore, images showed that subdural fluid accumulation, obscure sulci, and cerebral ventriculomegaly improved (**Fig. 2**).

The spinal drainage volumes were 46 and 55 mL on POD 1 and 2, respectively (**Fig. 3**). On POD 3, headache during head elevation was noted, and images showed cerebral ventriculomegaly and subdural fluid accumulation. Moreover, 71 mL of fluid had already been drained within half a day. The drain height was adjusted according to the drainage volume per hour, and the target daily drainage volume was 100-120 mL. However, drainage volume was small, and the drain was lowered to −7 cm by using the external acoustic pore as the standard. Consequently, the cerebral ventricles were found to be enlarged despite the low CSF pressure. After drain removal, headache was promptly relieved, and CT findings improved (**Fig. 3**). Thus, LPH was thus diagnosed. The CSF data were cell 68/μL and protein 275 mg/dL when the spinal drain was inserted and cell 435/μL and protein 113 mg/dL when the drain was removed. The subsequent clinical course was good. Cerebral vasospasm and hydrocephalus did not recur. The patient was discharged 3 weeks after surgery.

Discussion

LPH is expressed as negative-pressure hydrocephalus (NegPH) when CSF pressure is negative. LPH and

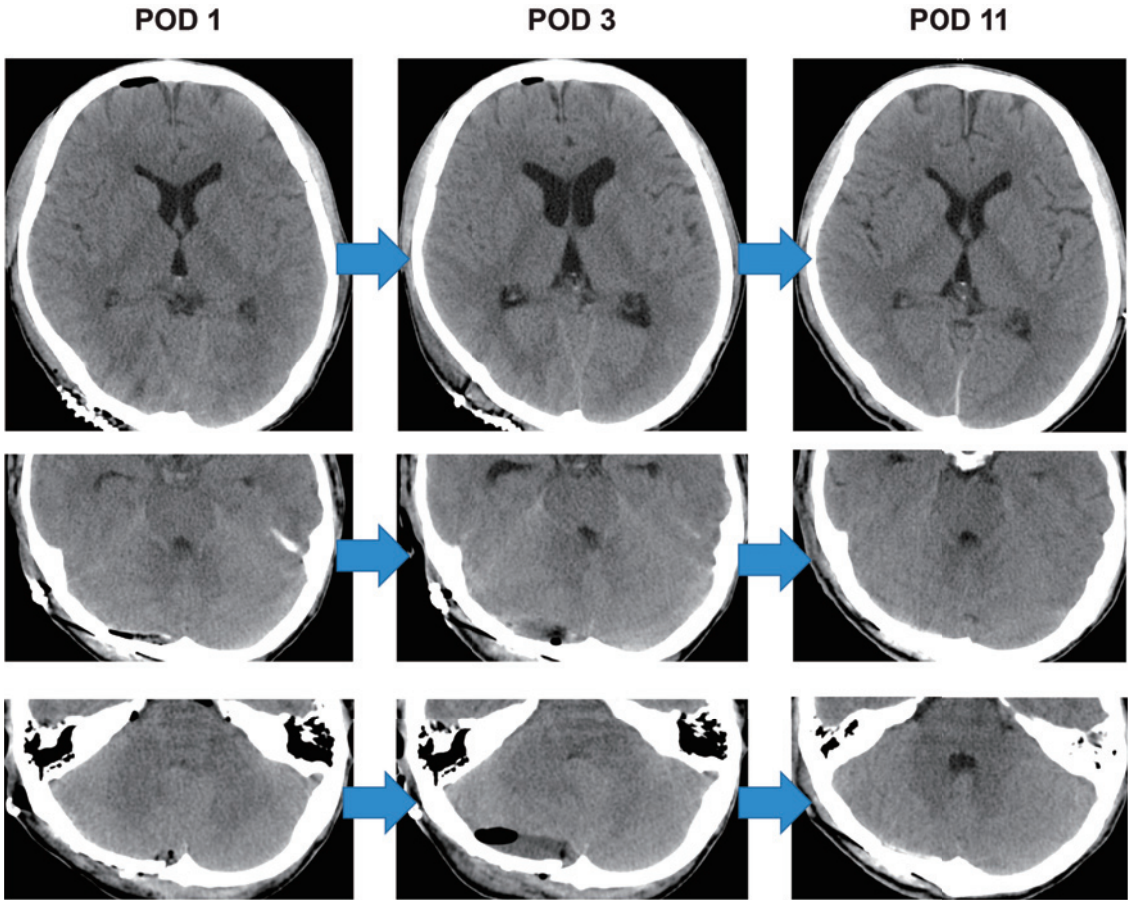

Fig. 2 Course of brain CT imaging.
On POD 3, CT images showed cerebral ventriculomegaly, opening of the subdural cavity at the posterior fossa craniotomy site, and fluid accumulation; the cerebellar sulci were narrow. After spinal drain removal (POD 11), subdural fluid accumulation, obscure sulci, and cerebral ventriculomegaly improved.

NegPH are clinically distinguished from other types of hydrocephalus. In addition, both types have rare characteristics that seem contradictory at a glance (i.e., cerebral ventriculomegaly in a state of low intracranial pressure). Symptoms include headache and disturbance of consciousness induced by low CSF pressure, dementia caused by cerebral ventriculomegaly, gait disturbance, and urinary incontinence. Many patients who underwent CSF pressure measurement by lumbar puncture and intracranial pressure monitoring had values of <80 mm H_2O. Although various putative causative factors and developmental mechanisms have been investigated, no definitive cause has been reported[2].

The developmental mechanism of the present case was examined (**Fig. 4**). CSF malabsorption caused by low CSF pressure resulting from excessive spinal drainage and compensatory intracranial hyperemia due to low CSF pressure may have occurred. Consequently, CSF accumulation and impaired CSF buffering capacity, leading to cerebral ventriculomegaly and hydrocephalus, may have resulted. Furthermore, increased brain compliance due to subarachnoid hemorrhage and surgery may also have impaired CSF buffering capacity, thereby contributing to cerebral ventriculomegaly. Furthermore, CSF buffering capacity may have been impaired by stenosis of the CSF tract due to cerebral ptosis resulting from low CSF pressure[3]. Cutler et al.[4] reported that CSF is not absorbed when intracranial pressure is <68 mm H_2O, which suggests that CSF malabsorption was noted in this case because of low CSF pressure, as the drain was lowered to −7 cm by using the external acoustic pore as the standard. According to the revised Monro-Kellie hypothesis, cerebral parenchyma + vascular bed + CSF cavity = constant[5]. Vascular bed and venous pressure increase in cases of low CSF pressure, resulting in CSF malabsorption. In addition, stasis is also a potential cause of hydrocephalus, apart from malabsorption. Deformity and distortion of the CSF tract due to cerebral ptosis (an elliptical deformity of the pons, dropping of the splenium of the corpus callosum, and narrowing of the prepontine

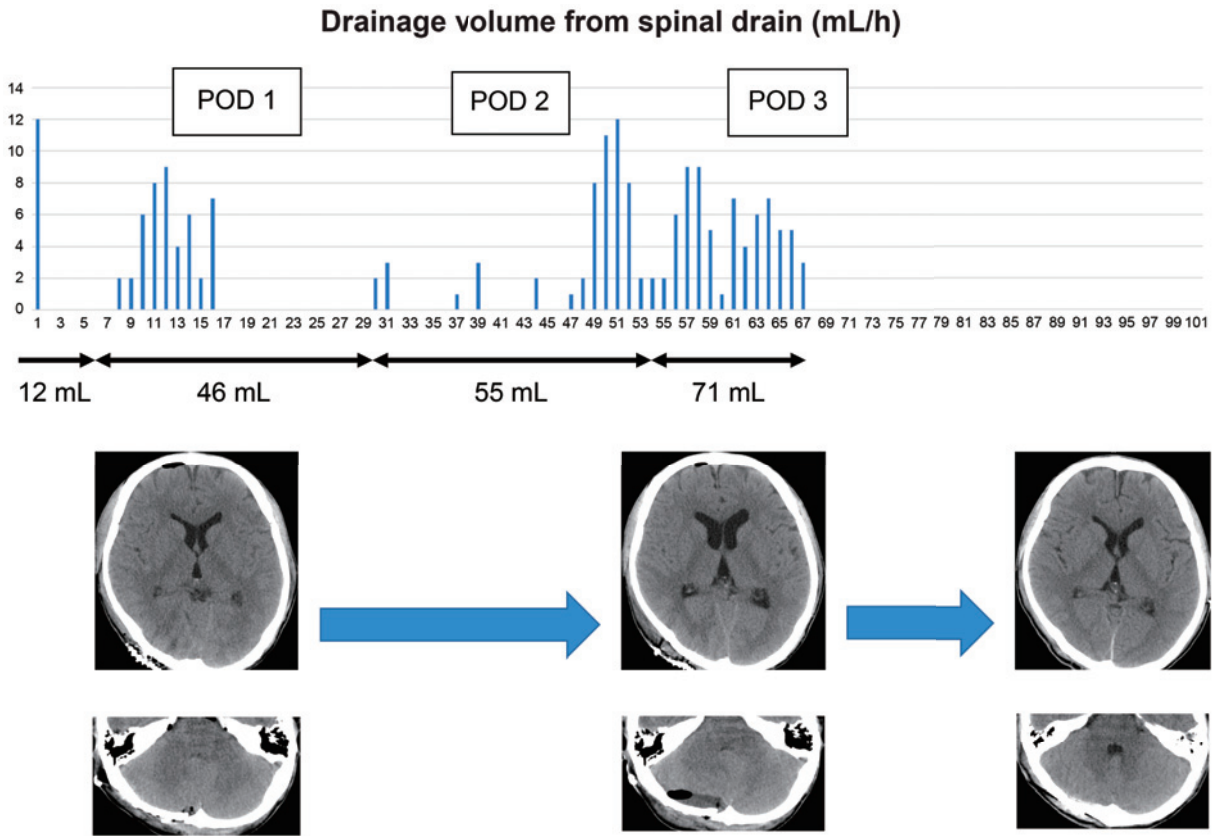

Fig. 3 Hourly drainage volume from the spinal drain and time course of CT findings.
Drainage volumes from the spinal drain were 46 and 55 mL on POD 1 and 2, respectively. Moreover, 71 mL of fluid was already drained in half a day on POD 3, when headache during head elevation, cerebral ventriculomegaly, and subdural fluid accumulation were noted. On POD 3, the spinal drain was removed, and CT findings improved.

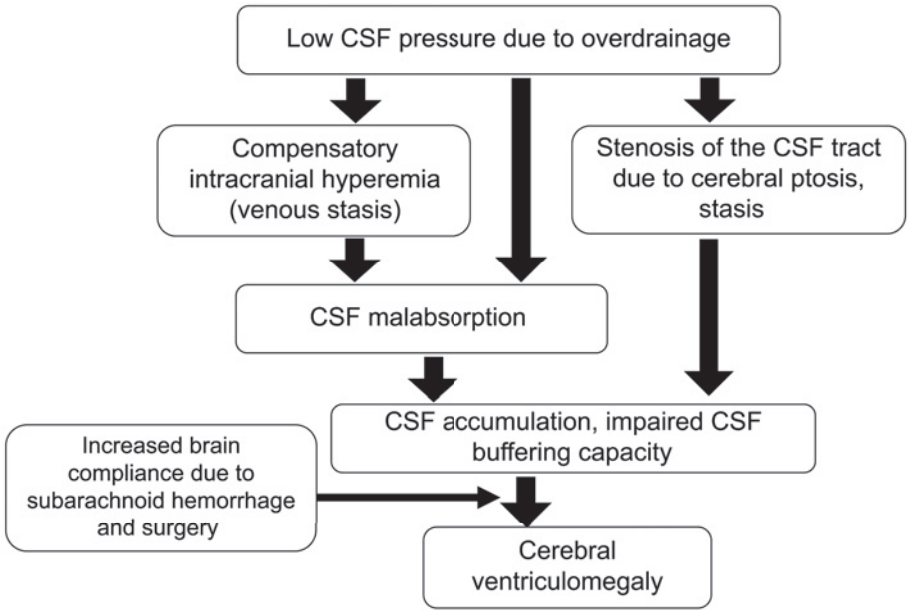

Fig. 4 Schema of the mechanism of onset.
Cerebrospinal fluid malabsorption due to low CSF pressure and compensatory intracranial hyperemia due to low CSF pressure caused CSF accumulation and impaired CSF buffering capacity, leading to cerebral ventriculomegaly and hydrocephalus. In addition, increased brain compliance due to subarachnoid hemorrhage and surgery impaired CSF buffering capacity. Furthermore, CSF buffering capacity was impaired by stenosis of the CSF tract due to cerebral ptosis caused by low CSF pressure.

Table 1 Summary of previously reported LPH cases (n = 19)

Author, year of publication	Age/ Gender	Underlying disease	Affected surgical treatment	Treatment	GOS at discharge
Cheng, 2017	23/M	Acute subdural hematoma	Craniotomy for hematoma evacuation	Negative-pressure CSF drainage, head elevation	5
	41/F	Subarachnoid hemorrhage	Craniotomy clipping	Negative-pressure CSF drainage, head elevation	5
	42/M	Brain contusion	Craniotomy	Negative-pressure CSF drainage, head elevation	3
	61/F	Hypertensive cerebral hemorrhage	Craniotomy for hematoma evacuation	Negative-pressure CSF drainage, head elevation	4
	62/M	Brain contusion	Craniotomy	Negative-pressure CSF drainage, head elevation	5
	8/F	Astrocytoma	Tumorectomy via craniotomy	Negative-pressure CSF drainage, head elevation	5
	39/M	Brain contusion	Craniotomy	Negative-pressure CSF drainage, head elevation	5
Pandey, 2017	26/M	Epidermoid tumor (posterior fossa)	Endoscopic transnasal transclival tumorectomy	Neck wrapping, CSF drainage 200 mL/day	5
Hunn, 2014	27/M	Pineal tumor	Craniotomy for extirpation	Negative-pressure CSF drainage, 120–360 mL/day	N/A
	51/M	Cavernous hemangioma	Craniotomy for extirpation	Negative-pressure CSF drainage, 120–360 mL/day	N/A
	65/F	Metastatic brain tumor	Craniotomy for extirpation	Negative-pressure CSF drainage, 120–360 mL/day	3
	12/M	Severe head trauma	Craniotomy	Negative-pressure CSF drainage, 120–360 mL/day	N/A
	36/M	Severe head trauma	Craniotomy	Negative-pressure CSF drainage, 120–360 mL/day	N/A
	54/F	Subarachnoid hemorrhage	Craniotomy clipping	Negative-pressure CSF drainage, 120–360 mL/day	N/A
	70/F	Unruptured cerebral aneurysm (MCA)	Craniotomy clipping	Negative-pressure CSF drainage, 120–360 mL/day	1
	63/M	Subarachnoid hemorrhage	Craniotomy clipping	Negative-pressure CSF drainage, 120–360 mL/day	4
Filippidis, 2011	61/M	Subarachnoid hemorrhage	Craniotomy clipping	Negative-pressure CSF drainage, neck wrapping	5
	67/M	Cavernous vascular malformation, dural arteriovenous Fistula	Embolectomy, γ knife, resection, decompressive craniectomy for postoperative cerebellar infarction	Liquorrhea repair, third ventriculostomy	3
	56/F	Dissecting aneurysm	Craniotomy clipping	CSF drainage, liquorrhea repair	5

cistern) attributable to low CSF pressure impaired CSF outflow[6].

Previously reported cases (n = 19) of LPH are shown in **Table 1**. All patients developed LPH after surgical treatment for cranial disease or trauma. One patient had undergone repair for postoperative liquorrhea, which was believed to be attributable to overdrainage due to spinal drainage. Thus, headache induced by low CSF pressure and image findings improved promptly after drain removal.

The cause of LPH in usually unclear, despite low CSF pressure, liquorrhea, and overdrainage. However, LPH may result from intracranial disease or craniotomy. In their pathophysiologic analysis, Cheng et al.[7] and Hunn et al.[8] reported that extracellular fluid deficiency changed the viscoelastic substance coefficient of cerebral parenchyma, leading to LPH. Treatment targeting hydrocephalus improvement by draining CSF by applying pressure lower than the intracranial pressure or negative pressure with ventricular drainage was reported for patients with no obvious liquorrhea. In addition, CSF drainage was continued while intracranial pressure was increased by lowering the head position, and venous pressure was increased by neck wrapping or reducing blood sodium concentration through fluid replacement with a hypotonic solution, in patients who developed negative intracranial pressures[9].

Appropriate treatment for LPH is sometimes delayed because symptoms and image findings are similar to those of intracranial hypertension and normal-pressure

hydrocephalus. Several LPH cases were difficult to diagnose and treat, and multiple shunt reconstructions were performed based on a diagnosis of shunt failure, because no improvement was noted despite partial reduction of shunt pressure with ventriculoperitoneal shunting. If cerebral ventriculomegaly does not improve even after shunt surgery for hydrocephalus, liquorrhea and the possibility of LPH should be considered. Moreover, the shorter the interval from onset to the start of treatment, the better the post-treatment prognosis and the more likely it is for ventricles to revert to their original size. With appropriate treatment, the prognosis is good. Thus, it is important to understand the pathology of LPH.

Conclusion

We treated a patient with LPH caused by spinal drainage after subarachnoid hemorrhage surgery. LPH should be considered when cerebral ventriculomegaly is present, even if liquorrhea and overdrainage are suspected. Furthermore, the symptoms and image findings are similar to those of intracranial hypertension and normal-pressure hydrocephalus, even though the condition might have resulted from intracranial disease or craniotomy. Moreover, LPH has a good prognosis if diagnosed early and treated appropriately. Thus, the possibility of low CSF pressure must be considered when treating hydrocephalus. Basic research will be helpful in clarifying CSF physiology.

Conflict of Interest: The authors declare no conflicts of interest.

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