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Original

Very Low-pressure Hydrocephalus : A New Clinical Entity and Issues of Treatment

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Abstract : Secondary normal pressure hydrocephalus (NPH) frequently occurs after severe head injury and cerebrovascular disease. This condition is usually treated by surgically implanting a cerebrospinal fluid (CSF) shunt with a pressure-setting valve or programmable valve. However, some patients do not respond to the shunt operation. Among these non-responders, we found 7 patients whose pressure-setting shunts were mechanically patent, but were not functioning due to very low intracranial pressure (ICP). In these 7 cases, continuous ICP monitoring indicated low pressure with occasional negative pressure, and the patients' consciousness improved during negative-pressure CSF drainage. We performed shunt revisions with zero setting on-off valves, which raised the mean functional independence measure (FIM) scores from 26 to 62. Four patients in a persistent vegetative state (PVS) regained their ability to communicate and recovered to the level of severely disabled (SD). We propose very low-pressure hydrocephalus (VLPH) as a new clinical entity, and describe the process of diagnosis and treatment.

Key words : normal pressure hydrocephalus, intracranial pressure, CSF shunt, persistant vegetative state, low pressure, secondary hydrocephalus

Introduction

Secondary normal pressure hydrocephalus (NPH) is frequently associated with severe head injury and cerebrovascular disease[3, 19]. This condition is currently treated by surgically implanting a cerebrospinal fluid (CSF) shunt with a pressure-setting valve or programmable valve[2, 19, 20]. A CSF shunt may help to improve various symptoms, including conscious disturbances, inactive states, and dementia, although its efficacy can only be predicted by a properly performed tap test or infusion test[7]. CSF shunts still fail to improve symptoms in a significant number of patients with NPH, even when the tap or infusion test has proven to be effective[14, 19]. Recently, we encountered 7 cases of secondary NPH in which CSF shunts with programmable valves failed due to the patients' very low intracranial

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Table 1. Case presentation

Patient	Age / sex	Primary disease	Treatment	Duration	GOS
1	85 / F	SAH	Clipping	4	PVS
2	66 / M	ASDH	Removal	7	SD
3	43 / M	SAH	IVS	5	PVS
4	73 / M	ICH	Conservative	28	SD
5	47 / M	SAH	Clipping	2	PVS
6	44 / M	SAH	Clipping	2	PVS
7	54 / F	SAH	IVS	12	SD

GOS : Glasgow outcome scale, PVS : persistent vegetative state, SD : severely disabled, duration : Months from admission to placement of the zero-shunt, SAH : subarachnoid hemorrhage, ASDH : acute subdural hematoma, ICH : intracerebral hematoma, removal : removal of hematoma, IVS : intravascular surgery

pressure (ICP). We successfully treated these patients using CSF shunts with zero-pressure valves. Four out of the seven patients, who were in a persistent vegetative state (PVS) preoperatively, recovered some communication abilities with the CSF shunt. We therefore propose the new clinical concept of low-pressure hydrocephalus (VLPH), and report on the diagnosis and treatment for this condition.

Materials and Methods

We examined 7 patients for this study comprising 1 patient with acute subdural hematoma, one with intracerebral hemorrhage, and 5 with subarachnoid hemorrhages (SAH). All of the patients presented with chronic secondary NPH that had been treated with a CSF shunt with a programmable valve (Codman Hakim adjustable valve, pressure setting 30–200 mm H_2O), from October 2000 to May 2005. Table 1 lists the patient characteristics.

In the acute stage of the patients' primary disease, CSF drainage was performed and their neurological conditions improved. Patients then received a CSF shunt with a programmable valve ; however, their clinical conditions did not improve as expected. Even after adjusting the valve pressure to the minimum of 30 mm H_2O, the patients' clinical conditions continued to worsen, with all Evans' ratios[18] remaining unchanged on CT scans. We first eliminated shunt malfunction as a cause of failure, then initiated continuous ICP monitoring in which the level of the external auditory meatus in the flat supine position was determined to be zero pressure. ICP monitoring revealed consistently low ICP occasionally falling below zero pressure for all cases. Therapeutic external CSF drainage with negative pressure was thus performed for several days, resulting in improved neurological conditions in all patients. CSF shunts with zero-pressure valves (On-off flashing reservoir, Heyer-Schulte NL850-0150) were then fitted in all patients (zero-shunt).

The time from the onset of primary disease to the beginning of a neurologically stable state ranged from 2 to 12 months. The patients' functional independence measure (FIM) scores[8] at that time were between 18 and 52. Four patients were diagnosed as being in

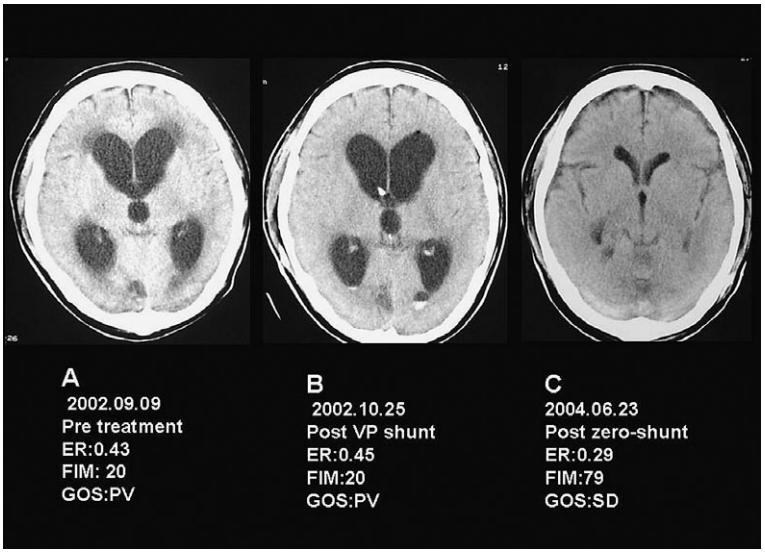

Fig. 1. Changes in ventricular size observed from CT scan in case 3. Evans ratio (ER) is 0.43 in CT obtained before CSF shunt placement (A). ER is 0.45 in CT obtained after placement of the CSF shunt with programmable valve (B). ER is 0.29 in CT obtained 8 months after zero-shunt placement (C).

Table 2. Results of zero-shunt (pre-post zero shunt)

Patient	FW	GOS (pre-post)	FIM score (pre-post)	ER (pre-post)
1	24	PVS-SD	18–36	0.42–0.37
2	10	SD-MD	30–63	0.48–0.39
3	9	PVS-SD	20–79	0.43–0.29
4	29	SD-MD	52–95	0.40–0.37
5	3	PVS-SD	18–43	0.31–0.24
6	4	PVS-SD	18–29	0.41–0.25
7	4	SD-MD	28–89	0.36–0.28
Mean			26–62**	0.40–0.33**

FW: follow-up duration (month), GOS: Glasgow outcome scale. FIM: (minimum 18, maximum 126), ER: Evans ratio. ** $P < 0.01$

a PVS and three patients were deemed severely disabled (SD). The duration of follow-up was 4 to 24 months after placement of the zero-shunt. The patients' neurological conditions were evaluated based on their FIM score and the Glasgow outcome scale (GOS)[6]. Ventricle size was evaluated by the Evans ratio on CT images.

Results

The patients' Evans ratio changed significantly from a mean of 0.40 prior to placement of the zero-shunt to a mean of 0.33 after the shunt placement ($P < 0.01$ paired t-test; Fig. 1, Table 2). FIM scores were also significantly improved in all patients from a mean of

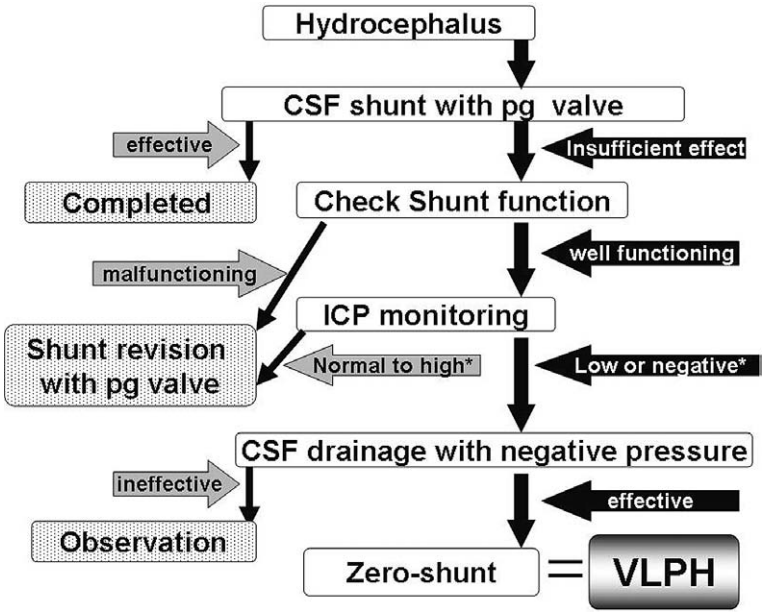

Fig. 2. Flow diagram for diagnosis and treatment of very low-pressure hydrocephalus (VLPH). Pg valve : programmable valve, CSF : cerebrospinal fluid, ICP : intracranial pressure, * pressure

26.3 to a mean of 62.0 after the zero-shunt was positioned ($P < 0.01$ paired t-test ; Table 2). After placement of the zero-shunt, all of the patients were able to communicate with their families and the medical staff, two patients were able to eat independently, and four became only partially dependent on help for eating. At the last follow-up date, four patients who were evaluated as being in a PVS preoperatively, improved to an SD state (from FIM 18.5 to 46.8) after the zero-shunt placement (Table 2). The patients experienced no complications including subdural hematoma or infection after placement of the zero-shunts. However, two patients needed anti-siphon devices implanted several months after placement of the zero-shunt due to the onset of headaches while in a sitting position.

Discussion

Since Adams *et al*[1] reported three cases of NPH in 1965, the CSF shunt became generally accepted as the most effective treatment of NPH. However, failure rates remained high after placement of such shunts (10–30% of patients with NPH), despite a thorough preoperative examination[7, 11, 14, 20]. Several authors recently reported NPH patients with failed CSF shunts presenting with low ICP[4, 10, 12, 13], a condition they called low-pressure hydrocephalus (LPH). These patients were successfully treated with negative-pressure CSF drainage or CSF shunts with low- or zero-pressure valves. Our study also examined a group of patients with low or negative ICP identified by ICP monitoring.

Historically, the term LPH has been used interchangeably with NPH[16, 17] due to the

association of NPH with relatively low ICP compared to high-pressure hydrocephalus. Even now with the concept of NPH widely known, some authors still use the term "low pressure hydrocephalus" when referring to NPH[9]. Therefore, the term LPH is not suitable for the condition that we have discussed in this paper. We propose the term "very low-pressure hydrocephalus" (VLPH) to distinguish it more strongly from NPH, and so that patients with VLPH could be diagnosed by a careful investigation. Fig. 2 shows a flow diagram used to distinguish VLPH patients from those with secondary hydrocephalus from a non-functioning routine CSF shunt. Based on the diagram, we could select eligible patients for whom the zero-shunt operation should be performed.

The causative mechanisms of VLPH remain unknown. Patients in this study included those with secondary hydrocephalus due to severe head injury or cerebrovascular disease. In the acute stage they showed high-pressure hydrocephalus, which manifested after several months with very low ICP. CSF production may be downregulated due to chronically increased ICP[15] or viscoelastic alterations in the brain due to prolonged over-stretching[5, 13].

A literature search revealed only 35 patients matching our described concept of VLPH, including the present 7 cases[4, 10, 12, 13]. There may be considerably more patients with VLPH who are being improperly treated after receiving CSF shunts with pressure-setting valves. It is worthy to note that four of our patients who were in a PVS preoperatively recovered enough function postoperatively to communicate and eat.

Conclusions

We propose that patients with failed CSF shunts should be closely examined for signs of VLPH. Placement of a zero-shunt may provide a chance for these patients to recover neurological function, and may help to solve a major medico-social problem by improving the quality of life in patients with severe brain disease.

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