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Case Report

Clinical Experience of Ventriculoperitoneal Shunt Dysfunction in a Patient with Hydrocephalus after Brain Injury: A Case Report

Shean-Jing Lin, Yun-Yee Chen, Mu-Jung Kao, Ching-Yang Lin, Sheng-Huang Hsiao

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We report a case of an adult patient with post-traumatic hydrocephalus who was treated using a ventriculoperitoneal (VP) shunt with a fixed pressure setting valve. Slit ventricle syndrome was found two months later and an antisiphon device (ASD) was added. A fixed pressure valve with ASD was found not to be optimal in terms of improving his neurological symptoms and a programmable valve system was therefore implanted with a pressure setting of 140 mmH₂O in the beginning. The valve then was gradually programmed to reduce down to 70 mmH₂O over two months. This case demonstrates that selection of a shunting device is difficult, and even after deliberate selection and use of an antisiphon device, slit ventricle syndrome could not be avoided. In addition, the patient in this case had increased episodes of seizure that were associated with acute intracranial hypertension; these were caused by obstruction. Good clinical monitoring of such patients is needed because of the intermittent nature of the high intracranial pressure symptoms. Notwithstanding this, minor complications involving shunt dysfunction may be neglected once the initial treatment has been completed in the neurosurgical ward and the patient has been transferred to a rehabilitation ward. It should be noted that this patient was very sensitive to over-drainage and siphoning was required in order to generate an effective transmantle pressure gradient. Thus, the highest valve setting available was used at the beginning of treatment and this was followed by a methodically lowering of the opening pressure based on the clinical response and computed tomography findings. Overall, the programmable valve shunt system would seem to be more effective when there is a need to deal with shunt dysfunction non-invasively. (Tw J Phys Med Rehabil 2009; 37(4): 251 - 259)

Key Words: hydrocephalus, programmable VP shunt, rehabilitation, slit-ventricle syndrome

INTRODUCTION

Hydrocephalus following head injury was first noted as early as 1914 by Dandy and Blackfan. Most of the reports to date support the hypothesis that traumatic brain injury (TBI) results in an alternation of the cerebrospinal fluid (CSF) flow and dynamics, which, in turn, leads to post-traumatic hydrocephalus. In 1956, ventricular shunts dramatically changed the outlook for hydrocephalus.

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Hydrocephalic adults, who have undergone a shunting procedure, have been reported to have complication and revision rates of between 35% and 80%. Complications associated with CSF shunting procedures may be categorized into the following three major groups: (1) complications related to the operative procedure itself, such as shunt infection; (2) mechanical problems of obstruction, fracture, and migration; and (3) complications occurring as a result of the flow characteristics of the shunt, such as overdrainage or underdrainage. Overdrainage is manifest as slit ventricle syndrome or subdural hygromas, and underdrainage is manifest by failure of the ventricles to change in size and persistent symptoms.

However, ventriculomegaly in adults can be confounded by degenerative changes within the brain parenchyma, and such a diagnosis is only considered as "confirmed" if improvement occurs after the shunting procedure. In addition, the dynamic changes in pressure that occur when a person is upright must be taken into consideration when choosing an optimal opening pressure valve. This diagnostic uncertainty may be the cause of the surprising high overall incidence of ventricular over-shunting.

This paper describes the use of a programmable valve shunt system for the management of slit ventricle syndrome. With the availability of this accurate programmable system, the need for a second operation to implant a different pressure valve was eliminated.

## CASE HISTORY

The 43-year-old male patient had a car accident on 12 March 2006. He was transported to hospital and his Glasgow coma scale (GCS) was E1M1V1 when he arrived at the emergency room of CGMH. The radiological impression after brain CT scans was one of acute mixed subdural and epidural hemorrhage over the right cerebral convexity and fractures of the right temporal and parietal bones. An emergency operation was performed immediately for removal of an intracerebral hematoma following craniectomy [Figure 1-A]. He was then transferred to our hospital on 10 April 2006. Aphasia was noted at that time and the GCS was E4M6V1. Therefore, ventriculoperitoneal (VP) shunting was performed using a Hakim medium-pressure valve that has a range from 80 to 130 mm H₂O. His GCS was measured as E4M6V3 during the following days and CT scans were undertaken on the 13 May 2006 [Figure 1-B], when over-drainage was suspected although he was doing quite well at that time. However, the coma scale dropped to E4M6V1 two months later and over-drainage syndrome was diagnosed using neuroimaging studies, where a slit-like configuration of the lateral ventricles was noted on 30 June 2006 [Figure 1-C]. Initially, the neurosurgeon replaced the medium-pressure valve with a high-pressure valve with a range from 120-170 mm H₂O, but no improvement was found postoperatively [Figure 1-D]. At this point, on 20 July 2006, a shunt ligation over the left medial clavicle head was performed. Seven days later, brain-bulging over the skull-defect site was noted; the shunt ligation was released and an antisiphon device (ASD) was introduced to the previous ligated VP shunt system. The GCS remained E4M6V1 after the ASD insertion. After cranioplasty on 07 August 2006, the patient’s GCS failed to improve and he was transferred to a rehabilitation unit [Figure 1-E]. Initially, when he was transferred, his GCS was E4M6V1, and left hemiparesis (power 3/5 in the upper and lower limbs) was still noted. The Modified Rankin scale of global disability was grade 5 and the Rancho Los Amigos was stage III after the rehabilitation program was completed. However, after discharge, he had episodes of seizure more and more frequently, although repeated measurements of his valproate level were within normal limits. At the time, the neurologist diagnosed the events as refractory seizures. After a grand mal seizure, due to a deteriorating neurological status in Dec 2007, he was readmitted to our neurosurgery ward. He underwent a revision of the VP shunt and a Codman Hakim programmable valve set at 140 mmH₂O was introduced on 19 Dec 2007. The valve was adjusted from 140 mmH₂O to 70 mmH₂O over the next two months and his GCS improved from E4M6V1 to E4V5M6 [Figure 1-F, 1-G]. The event chronology of this patient is presented in [Table 1] including the pressure and the GCS after each procedure. On being transferred to the rehabilitation unit again, he regained 4/5 power in his left limbs and his activity of daily life (ADL) improved from totally dependence to minimally supported. He became self-caring, required only a little assistance and reached a Modified Rankin scale of global disability of grade 3. The Rancho Los Amigos stage was VI after continuing rehabilitation therapy. Neuropsychological
testing showed subnormal verbal fluency, and verbal recall. Nonetheless, his verbal communication was appropriate to meeting basic needs. At a half year of follow-up, he had no further seizures with the pressure setting at 70 mmH2O. The nasogastric (NG) tube was removed during the second rehabilitation program and he started oral intake independently. He is able to walk for 50 meters without support and from this point onwards he has been in a stable condition.

Figure 1. (A) CT image of the patient 1 month after the patient sustained a severe TBI with hydrocephalus and periventricular lucency; (B) CT image of the patient after the VP shunt was performed; (C) Slitlike configuration of the lateral ventricles under VP shunt with medium-pressure setting valve; (D) mild degree of slitlike ventricle under VP shunt with high-pressure valve; (E) CT image of the patient after cranioplasty with high-pressure setting valve and ASD insertion; (F) CT scans after programmable VP shunt system performed with pressure setting at 140 mmH2O; (G) CT scans with programmable VP shunt system with pressure setting at 70 mmH2O.
Table 1. Event chronology happened to the patient after every procedure

<table>
<thead>
<tr>
<th>Date</th>
<th>Event</th>
<th>Valve setting</th>
</tr>
</thead>
<tbody>
<tr>
<td>10 April 2006</td>
<td>GCS: E4M6V1, Aphasia was noted</td>
<td>Conventional graded valve with median pressure: 80-130 mmHg</td>
</tr>
<tr>
<td>13 May 2006</td>
<td>GCS: E4M6V3, Overdrainage was suspected</td>
<td>Conventional graded valve with median pressure: 80-130 mmHg</td>
</tr>
<tr>
<td>30 June 2006</td>
<td>GCS changed from E4M6V3 to E4M6V1, Slit ventricle syndrome was noted.</td>
<td>Conventional graded valve with high pressure: 120-170 mmHg</td>
</tr>
<tr>
<td>20 July 2006</td>
<td>No improvement was found after shunt revision</td>
<td>Shunt ligation</td>
</tr>
<tr>
<td>27 July 2006</td>
<td>Brain-bulging over skull-defect site was noted</td>
<td>Shunt ligation released and ASD was introduced</td>
</tr>
<tr>
<td>07 Aug 2006</td>
<td>Transferred to rehabilitation ward, GCS: E4M6V1</td>
<td>High pressure valve with ASD</td>
</tr>
<tr>
<td>19 Dec 2007</td>
<td>Refractory seizure was noted and shunt revision was done</td>
<td>Codman Hakim programmable valve set as 140 mmHg</td>
</tr>
<tr>
<td>28 Jan 2008</td>
<td>Transferred to rehabilitation ward</td>
<td>Codman Hakim programmable valve set as 70 mmHg</td>
</tr>
</tbody>
</table>

**DISCUSSION**

The incidence of post-traumatic hydrocephalus has been reported to be as low as 0.7% or as high as 29%. There is no consensus regarding the most appropriate valve to use for the treatment of adult hydrocephalus. This is because the valve opening pressure is defined at the time of the shunt implantation and the surgeon has to anticipate the patient’s drainage needs and the contribution from the siphoning effect; it is based on these factors, the surgeon must choose an adequate opening pressure of the valve. In addition, the optimal drainage pressure for an individual patient may vary over time.

During the last 50 years, a variety of valve designs has evolved. These can be broadly classified into graded pressure, siphon control and flow-regulated valves. Initial studies found that graded pressure shunts are more effective in reducing the signs and symptoms of hydrocephalus. It has recently been emphasized that designs including flow control of the CSF are beneficial when meeting the needs of individual patients. CSF will vary depending on the size, position and activity of the person. Many factors must therefore be considered when selecting an appropriate shunt system.

For the management of post-subarachnoid-hemorrhage (post-SAH) and post-traumatic-brain-injury (post-TBI) hydrocephalus, it is important not to assume that an elevated intracranial pressure (ICP) is always present. If the pressure is normal preoperatively, the ICP may have to be lowered by the shunt to non-physiological negative values in order to reduce ventricular size. Some patients may require a siphon- induced negative ICP as low as -300 mmH2O in order to obtain neurological improvement as a result of a reduction in ventricular size. One dilemma is that use of a low differential pressure valve is likely to predispose the patient to a high risk of over-drainage-related complications. The major problems arising from overdrainage are subdural hematoma, premature closure of cranial sutures in infants, stenosis or occlusion of the aqueduct, slit ventricle syndrome and low ICP syndrome.

Slit ventricle syndrome (SVS) is one possible over-drainage-related complication characterized by (1) the slit-like configuration of the lateral ventricles as noted with imaging, and (2) intermittent headaches, unrelated to postures, which are frequently accompanied by nausea, vomiting, drowsiness, irritability and impaired mental activity. Incontinence, increases in focal neurological deficits already present, possible seizures, and lethargy
suggest a sudden increase in ICP. A previous study stated that slit-like ventricles are obvious on CT scans in approximately 5% of successfully shunted patients, and that one third of these have the signs and symptoms associated with SVS.\textsuperscript{[11]} An incidence of 5.3% for SVS in 302 shunted hydrocephalus patients has been reported in one study and, in another study, approximately one third of 106 patients with a VP shunt had SVS.\textsuperscript{[12,13]}

In this case, the patient had a VP shunt with a Hakim medium-pressure valve, which was introduced at that time when post-TBI hydrocephalus was first diagnosed. However, various shunt revisions were carried out due to the presence of SVS. After the high-pressure valve with ASD was used, repeated brain CT scans showed enlargement in ventricular size, but his neurological status remained unchanged. Larsson et al. reported that clinical effects and ventricular size are independent when the adjusted shunt opening pressure is within the 50-170 mmH\textsubscript{2}O range.\textsuperscript{[5]} Yoshioka et al showed that shunt operation may decrease the overall mortality rate, but it also increases the likelihood of severe disability or the development of a vegetative state.\textsuperscript{[14]} For these patients, a potential drawback of the use of ASD is that siphoning may be required in order to generate an effective transmantle pressure gradient. These patients are good candidates for a programmable valve without ASD because siphoning can usually be controlled with this adjustment.

The Codman Hakim programmable valve was developed by Hakim during the late 1980s. The pressure setting of the valve can be adjusted non-invasively by the use of a programmer that transmits a group of coded magnetic impulses; this allows 18 pressure settings, ranging from 30 to 200 mm H\textsubscript{2}O in 10-mm H\textsubscript{2}O steps to be set. It has been reported that several patients are sensitive to changes in opening pressure of as little as 10 mmH\textsubscript{2}O.\textsuperscript{[15,16]} It is possible to monitor the patient’s condition even after a very slight change in opening pressure and thus we were able to change the pressure gradually to reach an optimal level. Adjustments of the opening pressure are thought to help patients undergo intracranial conditioning. It is believed that over-drainage syndrome can usually be well managed merely by adjusting the opening pressure of the shunt valve. Kamano et al demonstrated that it was possible, using a VP shunt with a programmable valve, to treat over-drainage-related complications such as subdural hematoma by merely adjusting the valve to a higher setting.\textsuperscript{[17]} However, it is still difficult to dilate long-standing and extremely narrowed ventricles.\textsuperscript{[18]}

Several types of shunt revision with different valve pressures or ASD insertions were tried in our patient, but his cognition status did not get better and episodes of seizure occurred more and more frequently. Seizures arising from SVS in a patient with over-drainage disturb the patient’s routine daily rehabilitation exercises, and interfered with their functional outcome. Clinically, however, minor complications of shunt dysfunction may be neglected and remain untreated by staff once the initial treatment has been completed in the neurosurgical ward and the patient is transferred to a rehabilitation ward. However, such dysfunction is indeed an important disorder that may influence the rehabilitation outcome.\textsuperscript{[19]}

When a person with a shunt assumes an upright position, the valve will only close when the ICP is sufficient negative to offset the hydrostatic pressure difference caused by hydrostatic effect of the vertical length of tubing.\textsuperscript{[2]} This phenomenon is typically called siphoning, and the ASD was developed to combat this problem and prevent ventricular hypotension. However, the absorption of CSF depends on the pressure difference between ICP and CSF pressure in the superior sagittal sinus, and the difference in pressure between ICP and the sinus needs to be maintained even in a sitting position.\textsuperscript{[20]} The balance of the intraparenchymal venous pressure and the CSF pressure helps to maintain the brain’s parenchymal tissue free from distortions and leaves the ventricular size undisturbed. Based on Bergsneider et al’s experience, treatment failure involving a standard VP shunt with ASD can occur for two reasons. One is that a sufficient negative ventricular pressure is not reached; the other one is that there is a lack of the time needed to produce a reduction in ventricular size.\textsuperscript{[19]} This patient improved dramatically in cognition and ADL after the mismatching of the valve to the patient’s requirements has been corrected with a pressure setting of 70mmH\textsubscript{2}O. This showed that siphoning may be required in order to generate an effective transmantle pressure gradient in this patient. In addition, he also had SVS, which was associated with an increased rate of seizure due to an intermittently high ICP. Hirayama emphasized that an intermittently high ICP
may be due to temporary ventricular catheter occlusion.\textsuperscript{[21]} This patient underwent insertion of a VP shunt with a programmable valve set at 140 mmH\textsubscript{2}O initially, in order to resolve SVS in the beginning. This was then gradually lowered to an opening pressure of 70 mmH\textsubscript{2}O over a period of two months to provide enough negative ventricular pressure; in these circumstances, his clinical symptoms improved dramatically. At the half year follow-up, with a pressure setting of 70 mmH\textsubscript{2}O, he had no further seizures.

In addition, the presence of cavum septum pellucidum (CSP) was noted in this patient. This is the congenital abnormality in the general population and, in this cases, was pinpointed from the CT scans. When a cavity between the two leaflets of the septum is present, it is known as CSP. It also referred to as the “fifth ventricle” since it is filled with CSF; however, it is not visibly connected to the ventricle system in humans.\textsuperscript{[22]} In the course of normal development, it closes in 15\% of humans within one month after birth and there is 85\% closure within the first 6 months of life.\textsuperscript{[23]} Bogdanoff and Natter found the prevalence of CSP to be less than 1\% of the general adult population.\textsuperscript{[24]} Some authors have suggested that a small CSP is common and reflects normal anatomical variations. A large CSP with a length of more than 6 mm is more common among patients with schizophrenia.\textsuperscript{[25-27]} In our patient’s case, there was neither a history of schizophrenia, nor were there marked symptoms of schizophrenia found during admission. Although negative symptoms such as alogia, avolition and affective flattening were difficult to recognize due to the patient’s impaired cognition status, he had no disorganized speech, delusion, hallucination, or disorganized behavior over at least 6 months. Schunk has stated that increased pressure within the ventricles may cause secondary dilation of the septum pellucidum.\textsuperscript{[28]} CSP is known to be common and pathological in brain damaged boxers. Based on the above, we hypothesize that the presence of the CSP may be due to tearing of the septal wall, which caused spinal fluid to collect within the cavum and is without any relationship to his neurological symptoms.

| CONCLUSIONS |

Shunt dysfunction during hospitalization for rehabilitation appears to be present in approximately 13.2\% of patients with a VP shunt.\textsuperscript{[19]} Therefore, because shunt dysfunction interferes with the functional outcome during rehabilitation, we should pay more attention to the symptoms of shunt dysfunction. Shunt overdrainage may occur due to an improperly set shunt valve or due to siphoning of the CSF caused by gravity when the patient is upright. The decision to choose a given fixed pressure valves system is made purely based on the surgeon’s experience. Black et al have recommended using a medium-pressure-valve as an initial shunt in all patients with normal pressure hydrocephalus.\textsuperscript{[29]} According to the initial CT scans, which showed brain tissue bulged out of the craniotomy wound, the rounded appearance of the frontal horns and the enlargement of the temporal horns and ventricular size, a medium-pressure-valve was chosen to implantation in this case. The selection of valve was considered to be correct because the GCS improved in the following days. However, over-drainage occurred two months later due to the siphoning effect in the upright position and an ASD was inserted. However, this carried the risk of increased intracranial pressure, which, in this case, seemed to be associated with intermittent patient seizures. The seizures were controlled after sufficient negative intraventricular pressure was reached by slowly reducing the opening pressure by means of a programmable valve. The VP shunt with a Codman Hakim programmable valve, as has been found by other authors could be fine tuned by us and allowed titration of the opening pressure more precisely than is possible with a traditional pressure setting valve. This allows the brain parenchyma to adapt over a longer time period.\textsuperscript{[30]} The programmable valve system afforded a more safe approach to adjust the opening pressure as changes in the setting could be based on the clinical response. In addition, a VP shunt using a programmable valve system is convenient and valuable when treating shunt dysfunction in a rehabilitation ward non-invasively.

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引流管功能異常在水腦症病患之治療經驗：病例報告

林筱菁  陳允宜  高木榮  林慶揚  蕭勝煌

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本文報告一位 43 歲男性因車禍導致顱內出血併發水腦症，接受顱骨與血塊移除及植入引流管手術。術後兩個月發現有腦室細隙現象 (slit-ventricle syndrome)，於是將中壓式腦室腹膜引流管 (ventriculoperitoneal shunt with median pressure valve) 更改為高壓式腦室腹膜引流管 (ventriculoperitoneal shunt with high-pressure valve)。然而術後神經學症狀仍未改善，於是將抗虹吸裝置 (anti-siphon device) 置入並執行顱骨回填手術。病人一連串術後影像學上腦室細隙現象雖有改善，但因癲癇發作次數頻繁，且以抗癲癇藥物治療無效。病人因癲癇發作再次入院後，改以可調式腦室腹腔引流管 (programmable ventriculoperitoneal shunt)。一開始設定為 140 mm H₂O，之後根據其臨床神經學症狀，在兩個月內的時間內往下調整為 70 mmH₂O。癲癇在半年追蹤期內不再發生，病人臨床上從昏迷指數 (Glasgow coma scale) 由 11 分進步到 15 分，認知功能量表 (Rancho Los Amigos stage) 由 3 級進步到 5 級，modified Rankin scale 由 5 級進步為 3 級。腦室細隙現象不僅由影像學上診斷，有時仍會以頭痛、嗜睡、噁心、認知功能不良、失禁、癲癇頻繁的發作來表現，被認為是因爲短暫的引流管阻塞導致腦壓升高。因此必須先將引流管設為高壓，改善其腦室細隙現象，之後往下調整為低壓以便產生足夠的負壓來引流腦脊髓液，以達到腦脊髓液吸收與製造的平衡，避免腦組織產生扭力 (torsion)。病人於臨床神經學的症狀便會有重大的進步。由於放置引流管後腦室細隙現象發生率約 5%，而且會影響到病人復健的成果，所以在復健的過程中需了解引流管的術後併發症，而可調式腦室腹膜引流管也提供了一種非手術性的調整引流壓力的方法，以供醫師遇到似案例時的參考。（台灣復健醫誌 2009；37(4)：251 - 259）

關鍵詞：水腦症 (hydrocephalus)，可調式引流管 (programmable VP shunt)，復健 (rehabilitation)，腦室細隙症候群 (slit-ventricle syndrome)

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