Beneficial Effect of Siphoning in Treatment of Adult Hydrocephalus

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**Objective:** To increase awareness about the treatment of adult patients with shunt-nonresponsive hydrocephalus—a state characterized by marked ventriculomegaly, low intracranial pressure, and a patent cerebrospinal fluid diversionary shunt.

**Design:** Retrospective analysis of hospital and outpatient records.

**Patients:** Four patients with symptomatic ventriculomegaly and patent ventriculoperitoneal shunts treated with a protocol of progressive ventricular hypotension induced by external cerebrospinal fluid drainage.

**Results:** Severe clinical manifestations exhibited by the patients, including parkinsonian features, Parinaud syndrome, and extensor posturing, completely reversed once a normalization of ventricular size was achieved. External ventricular drainage pressures as low as −30 cm H2O were required to reduce ventricular size. All patients finally received a shunt incorporating a standard medium differential pressure valve with no antisiphon device.

**Conclusions:** Shunt siphoning may be an essential mechanism by which cerebrospinal fluid shunting is effective in many patients with adult hydrocephalus. Cerebrospinal fluid shunts that contain an antisiphon device are ineffective in these patients, despite the attainment of “physiologic” intracranial pressures. Based on reported experimental and clinical evidence, it seems that the cause of this condition may be related to abnormally high intracranial compliance.

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ADULT hydrocephalus differs from that of childhood in that the former is usually not associated with persistently elevated intracranial pressure (ICP). A diagnosis of hydrocephalus in children is seldom questioned because the findings of ventriculomegaly in association with elevated ICP are pathognomonic. Conversely, ventriculomegaly in adults can be confounded by degenerative changes of the brain parenchyma. In many patients with adult hydrocephalus—particularly so-called normal pressure hydrocephalus—the diagnosis is considered “confirmed” only if improvement occurs after a cerebrospinal fluid (CSF) shunting procedure. This is partly because of a lack of definitive clinical or neuroradiographic criteria to predict which patients with presumed normal pressure hydrocephalus will respond to a shunt. A risk of this treatment-dependent diagnostic approach is that patients who do not improve after shunting may be labeled as “nonresponders” and, therefore, considered to be misdiagnosed. This diagnostic uncertainty is partly caused by our incomplete understanding of the pathophysiological features of adult hydrocephalus and the mechanisms by which shunting procedures work. Whereas intermittent elevations in ICP together with a sustained transmantle pressure are likely to be responsible for ventricular enlargement, clinical deterioration has not been temporally correlated with ICP abnormalities. Instead, the distorting effects of ventriculomegaly may be the primary mechanism of neural dysfunction.

The obscure relationship of intraventricular pressure to the pathogenesis of adult hydrocephalus has direct consequences regarding treatment. Studies comparing the efficacy of various opening pressures of differential pressure valves focus on a treatment rationale for preventing ICP elevations. For unclear reasons, these same studies disregarded the siphoning phenomenon that invariably accompanies this type of shunt. Such bias likely arises from the fact that, in the literature, CSF siphoning is primarily associated with overdrainage complications. This inconsistent presentation of shunt-ICP physiological features reveals a limited understanding of the mechanisms by which shunting is effective for the treatment of adult hydrocephalus. The understanding of shunt-ICP physiological features is highly pertinent to the subgroup of patients who do not respond to a standard shunt procedure. If one believes...
PATIENTS AND METHODS

PATIENTS

From February 1, 1994, to January 31, 1997, 4 adult patients were referred to the University of California at Los Angeles Adult Hydrocephalus Program for persistent neurologic deterioration secondary to hydrocephalus. Clinical presentations are summarized in Table 1. Within 5 years of referral to the program, all patients were diagnosed as having hydrocephalus and were successfully treated previously. In general, each patient enjoyed a symptom-free period after the initial shunt procedures but then had progressive neurologic decline that did not respond to shunt revisions. Although the presentations were unique, neither headache nor nausea was a complaint. Three patients had parkinsonian features; all were practically bedridden because of severe gait apraxias, increased motor tone, or decreased responsiveness. Two of 4 patients were assessed by at least 1 neurologic consultant as being irreversibly brain damaged. This assessment had presumably been largely based on the finding that the measured ICP was not elevated. Computed tomographic (CT) scans revealed marked ventriculomegaly clearly out of proportion to cerebral atrophy.

TREATMENT PROTOCOL

After determination of a patent VP shunt and a low normal ICP, each patient was treated using a modified protocol of Pang and Altschuler. First, a long-tunneled ventriculostomy was performed. The existing shunt was either removed or disconnected. In patient 4, the pressure-volume index was measured in the operating room. Aside from a 24-hour perioperative dose of antibiotic medications, no further antibiotic drugs were given. An external ventricular drain was connected to a collection system that had an integral 1-way valve to prevent air reflux (Cordis Co, Miami, Fla). All patients were cared for in the intensive care unit while the ventriculostomy was in place. The initial drainage level was set at 0 cm, referenced to the external auditory meatus.

A limited CT scan (3 planes, 1 cm thickness, demonstrating the third and lateral ventricles above the level of the corneas) was obtained on alternate days. The external ventricular drainage level was lowered 2 to 3 cm/d until a reduction in ventricular size occurred. At this maximum negative drainage level, ventricular size normalized (judged by results of previous neuroimaging studies and the degree of atrophy present) and the patient underwent placement of a VP shunt with a medium differential pressure valve. After surgery, the patient was observed on the ward for 2 to 3 days, then discharged from the hospital.

That prevention of intermittent ICP elevations is the therapeutic goal, then a combination of a patent shunt and a near-zero ICP is presumably adequate. Conversely, if reduction in ventricular size is the goal, reversal of the transmantine pressure gradient must be achieved. Because such a reversal may require significantly negative intraventricular pressures, this opens the possibility that a shunt may be therapeutically incompetent, despite being patent. We report our experience with 4 adult patients whose common finding was that siphoning was essential to effectively treat their hydrocephalic condition. All had marked ventriculomegaly, patent ventriculoperitoneal (VP) shunts, and a near-zero baseline ICP. These patients were considered “nonresponders” by consulting physicians, but, after aggressive treatment, all made full recoveries. The study of their clinical presentations and response to the treatment protocol used adds new insight to our understanding of the treatment of the enigmatic disorder of adult hydrocephalus.

RESULTS

Table 2 summarizes the treatment courses of our patients. In patient 4, the calculated pressure-volume index was 108. Three patients required a significantly negative external drainage level before ventricular size began to decrease (−19 to −30 cm H2O). Aside from small daily variations in the results of neurologic examinations, significant improvements in the levels of consciousness, vocalization, parkinsonian findings, gait, and motor tone were not noticed until ventricular size showed definite reduction on CT (Figure 1 and Figure 2). This improvement in neurologic function typically occurred over a 24-hour period. A dramatic improvement of the Parinaud syndrome occurred in all patients, although complete resolution was delayed up to 3 weeks after hospital discharge. Three patients were discharged directly home, whereas patient 4 was transferred to a rehabilitation facility for 2 weeks.

Outcome was excellent in all 4 patients (follow-up, 28-46 months). Patients 1, 2, and 3 returned to independent lives, the younger patients regaining their previous full-time employment. Patient 4’s moderate short-term memory deficit remained after the original aneurysmal subarachnoid hemorrhage. Patients 1 and 4 required subsequent shunt revisions for mechanical obstructions at 9 and 28 months, respectively. Each did well with short hospital stays.

COMPLICATIONS

Complications are summarized in Table 2. Introduction of intraventricular air produced a significant setback in 2 patients. Patient 2 inadvertently pulled on the external ventricular drain tubing, breaking the ventriculostomy at the point that it was sutured to the scalp. A moderate amount of intraventricular air was introduced. A new ventricular catheter was immediately placed. The slight enlargement in ventricular size resulted in approximately 7 extra days in the treatment protocol (an extended intensive care unit stay). The introduction of a large amount of intraventricular air occurred in patient 4 when a 3-way stopcock cracked while the patient was being drained at ~30 cm H2O. The patient, whose mental status had significantly improved, immediately lost consciousness and had a generalized seizure. After pneumoencephalographic maneuvers to localize most of the air in the frontal horns, she underwent fluoroscopic-guided bifrontal ventriculostomy for removal of the air. This required starting the external ventricular drain protocol again, resulting in a 14-day extension of the intensive care unit stay.
A small, unilateral subdural effusion formed during treatment in patient 2, as recognized by the CT imaging protocol, which prompted the placement of a subdural catheter connected distal to the valve at the time of VP shunt placement. This subdural effusion immediately disappeared after surgery.

Our experience with 4 patients suggests that reduction in ventricular size by induced ventricular hypotension was a necessary and effective treatment for their condition. This cause-and-effect relationship suggests that the effectiveness of standard VP shunts in some patients with adult hydrocephalus is derived from the siphoning effect of CSF drainage. This is not the first article to describe shunt-nonresponsive clinical states. McCullough and Wells26 and McCullough27 reported experience with 4 patients whose conditions deteriorated after the addition of an antisiphon device (ASD) to a previously existing VP shunt. Neurologic recovery and reduction in ventricular size occurred after removal of the ASD. In 1994, Pang and Altschuler24 described a disorder they termed the low-pressure hydrocephalic state. Their series consisted of 12 patients, between 4 and 72 years old, who had progressive neurologic deterioration, ventriculomegaly, and normal ICP. Abnormally high intracranial compliance was considered to be the primary problem. Unlike McCullough’s series, none of these patients had ASDs. Using an external ventricular drainage setup, they reconstituted intracranial compliance, reduced ventricular size, and reversed neurologic symptoms. Mean ± SD maximum negative pressure required was -5.7 ± 3.6 mm Hg (range, -1.5 to -14.7 mm Hg). All patients eventually received a low or medium differential pressure valve shunt.

The previous initial studies describing shunt-nonresponsive patients enabled us to recognize our first

### Table 1. Clinical Presentations*

<table>
<thead>
<tr>
<th>Patient No./ Age, y/Sex</th>
<th>Clinical History and Symptoms</th>
<th>Intervening Events Immediately Before Referral</th>
<th>Clinical Condition at the Time of Referral</th>
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<tbody>
<tr>
<td>1/21/M</td>
<td>Posttraumatic hydrocephalus, undergone VP shunt at age 16 y complicated by meningitis 3 y later; he had multiple shunt revisions, each with an MDPV; presented 7/96 with a 1-mo history of intermittent upward gaze paresis and dizziness; CT scan showed normal-sized ventricles</td>
<td>Parenchymal ICP monitoring revealed wide variation in pressures, consistent with intermittent shunt function; the shunt was revised to a Level 1 Delta valve; he deteriorated over 24 h to lethargy, with massive enlargement of his ventricles; immediate shunt revision revealed no evidence of mechanical obstruction; because of lack of improvement, the following day an external ventriculostomy was placed; initial supine ICP was 6 cm H2O</td>
<td>Lethargic, localizing to pain; Parinaud syndrome (upward gaze palsy, poor light and near convergence, convergence retraction nystagmus), mild 6th CN paresis on the right</td>
</tr>
<tr>
<td>2/35/M</td>
<td>Idiopathic aqueductal stenosis diagnosed at age 32 y presenting with progressive headache; underwent a VP shunt (MDPV); on 4/96 he presented with intermittent headaches and double vision; CT scan showed small ventricles</td>
<td>Slit ventricle syndrome was suspected, and an additional MDPV was added to the system; this resulted in massive enlargement of the ventricles, loss of consciousness, and extensor posturing; the shunt was revised to a Level 1 Delta valve; because of no improvement, the shunt was reexplored the following day, finding a patent system and 0 cm H2O ICP</td>
<td>Alert, expressionless face; hypophoncic; Parinaud syndrome, 6th CN palsies; severe hypertonia with frequent extensor posturing; resting tremor; systemic hypertension, diaphoretic</td>
</tr>
<tr>
<td>3/61/F</td>
<td>Seen at age 60 y with progressive gait ataxia and incontinence, suspected idiopathic NPH; underwent a VP shunt (MDPV) with resolution of symptoms; on 11/95 seen with lethargy and increased ventricular size</td>
<td>A peritoneal shunt obstruction was diagnosed and the shunt was revised to include a Level 1 Delta valve; she continued to be lethargic after surgery with large ventricles, therefore a ventriculostomy was placed; the supine ICP was 4 mm Hg</td>
<td>Lethargic with Parinaud syndrome; lower extremities hyperreflexic, marked gait apraxia</td>
</tr>
<tr>
<td>4/66/F</td>
<td>Hydrocephalus developed after aneurysmal subarachnoid hemorrhage on 2/94; a VP shunt (MDPV) was complicated by infection; after antibiotic drug treatment, replaced shunt was complicated by bilateral subdural hygromas necessitating a subdural-peritoneal shunt; neurologic status began to deteriorate 10/94, with decreasing cognition and increasing ventricular size</td>
<td>A shunt malfunction was suspected, and the shunt was revised to include a Level 1 Delta valve; she continued to deteriorate, with decreased responsiveness and increasing ventricular size; a shunt tap revealed a supine ICP of 2 cm H2O; this procedure was complicated by the introduction of a moderate amount of intraventricular air</td>
<td>Alert, with marked psychomotor delay and single-word verbal responses; hypophoncic, expressionless face; severe gait ataxia; cogwheeling rigidity and resting tremor</td>
</tr>
</tbody>
</table>

*VP indicates ventriculoperitoneal; MDPV, medium differential pressure valve; CT, computed tomography; ICP, intracranial pressure; CN, cranial nerve; and NPH, normal pressure hydrocephalus.
†Manufactured by Medtronic/PSMedical, Goleta, Calif.
patient with this disorder. However, these studies did not address the known hydrostatic effects of shunts on ICP in relation to the treatment protocols suggested. Despite multiple studies and articles17–19,21,28,29 demonstrating the principles of shunt pressure dynamics, emphasis has been placed on choosing an “appropriate” valve closing pressure.12,15,24 The effect of shunt siphoning has been ignored, concentrating instead on reducing elevated ICP. Several studies17–20 documented that, in patients harboring intraventricular air, intraventricular hypotension is encountered in every case once the patient is placed in an upright position. Because the resultant pressure gradient is large compared with valve opening pressures, significant negative ventricular pressures are encountered, even with the use of high-pressure valves.

The reason intracranial hypotension must be induced to effectively treat patients with adult hydrocephalus appears to arise from altered biomechanical tissue properties known to occur with this disorder. Tans and Poortvliet30 demonstrated that the rate and completeness of ventricular size reduction after shunting correlated well with the degree of elevation of the preoperative pressure-volume index. The extraordinarily high pressure-volume index measured in 1 of our patients supports the findings of Pang and Altschuler,24 who demonstrated high intracranial compliance in many patients with the low-pressure hydrocephalic state. These clinical findings agree with experimental hydrocephalus models31,32 demonstrating that progressive ventricular enlargement is associated with an increase in intracranial compliance. As suggested by Pang and Altschuler,24 the failure of standard CSF diversion shunts in these patients occurs because the hypercompliant intracranial compartment effectively absorbs a broad range of intraventricular pressures.

The physics behind this state are probably best explained by applying the Laplace Law, as proposed by Hakim et al.33 A sufficiently negative transmural pressure is apparently necessary to overcome the wall tensions maintaining the enlarged ventricular cavity. In our patients, like those reported by McCullough,27 the inclusion of an ASD effectively prevented ventricular hypotension. Future studies measuring the transmural, ventricular, and subarachnoid pressures simultaneously, as previously done by Hof and Barber,4 during subatmospheric drainage, may further our understanding of the mechanisms involved.

Because all differential pressure valve shunts allow siphoning, it could be argued that the intracranial hypotension that accompanies them should be sufficient for the treatment of patients with high intracranial compliance states. Based on our experience and that of Pang and Altschuler,24 we speculate that treatment failure in these patients by standard shunting methods can occur for at least 2 reasons. The common feature in these patients is that a sufficiently negative ventricular pressure (or negative transmural pressure) is not reached or is not present for an adequate duration to produce a reduction in ventricular size. As demonstrated by patients 2 and 4, sustained negative pressures lower than −28 cm H2O were necessary to achieve a reduction in ventricular size. Such pressures are uncommonly achieved in patients with a VP shunt17–19,28 and are unlikely to be sustained unless the patient is able to maintain a completely upright position for prolonged periods. Our patients were practically bedridden and, therefore, would not have likely achieved sufficiently negative pressures. Even excluding the valve entirely, as suggested by McComb,34 would likely have been ineffective for the patients described in this study.

Based on known postural changes of ICP after placement of a differential pressure valve VP shunt,35 it is clear

### Table 2. Treatment Results and Complications

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Initial BFD, cm (Evans Ratio)</th>
<th>Final BFD, cm</th>
<th>Maximum Negative EVD Level, cm</th>
<th>ICU Stay, d</th>
<th>Treatment Results†</th>
<th>Complications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>4.8 (0.35)</td>
<td>3.1</td>
<td>−19</td>
<td>21</td>
<td>Alert, oriented; moderate upward gaze palsy; normal motor examination results</td>
<td>Required a subsequent shunt revision 9 mo after treatment for ventricular catheter obstruction</td>
</tr>
<tr>
<td>2</td>
<td>5.0 (0.39)</td>
<td>4.3</td>
<td>−28</td>
<td>28</td>
<td>Alert, oriented; mild upward gaze palsy; bilateral 6th cranial nerve paresis; no tremor, normal tone; mildly ataxic gait, walking without assistance</td>
<td>Intraventricular air was introduced after the patient inadvertently pulled out the ventriculostomy catheter on ICU day 13; growth of a small subdural effusion required a subdural catheter connected to the VP shunt</td>
</tr>
<tr>
<td>3</td>
<td>5.0 (0.39)</td>
<td>3.2</td>
<td>−5</td>
<td>9</td>
<td>Alert, disoriented; improved facial animation; mild upward gaze palsy</td>
<td>None</td>
</tr>
<tr>
<td>4</td>
<td>5.4 (0.48)</td>
<td>4.3</td>
<td>−30</td>
<td>54</td>
<td>Alert, oriented; poor short-term memory; conversant, normal voice; improved facial animation; no tremor or rigidity; walking with walker</td>
<td>A large amount of intraventricular air was introduced secondary to a break in the 3-way stopcock in EVD on ICU day 26, resulting in obtundation and seizures; an operative procedure was required to remove the air; approximately 2 y after treatment, a conversion to a VA shunt was required secondary to peritoneal adhesions</td>
</tr>
</tbody>
</table>

†Results of neurologic examination before shunt and after external cerebrospinal fluid drainage.

* BFD indicates bitemporal distance (maximum distance of lateral aspects of frontal horns of the lateral ventricles); Evans ratio, the BFD divided by the maximum biparietal distance of the inner skull table; EVD, external ventricular drainage; ICU, intensive care unit; VP, ventriculoperitoneal; and VA, ventriculoatrial.
that a sustained intracranial hypotension state cannot be maintained after discharge from the hospital. It is then reasonable to question why a “normalized” ventricular size state continued during delayed follow-up. We hypothesize that this is caused by a change in ventricular compliance that accompanies the reduction in ventricular size. Results of experimental studies\(^2\) indicate that intracranial compliance positively correlates with ventricular size. Therefore, as ventricular size normalizes, the pressure-volume relationship predicts that greater intraventricular pressures are required to result in an increased ventricular size. A significant increase in pressure, however, would not occur because CSF would be diverted through the shunt, regardless of what position the patient was in. This reduced compliance effect, combined with continued intermittent intracranial hypotension associated with the upright position, likely accounts for the sustained “normal” ventricular size seen in our patients. Assuming that this explanation is correct, it is less important which valve opening pressure is chosen once ventricular size is normalized. We did not see the necessity of slowly increasing the external ventricular drainage level to determine the most “appropriate” shunt valve, as described by Pang and Altschuler.\(^2\)

In this and previous series,\(^2\) the knowledge that each patient had responded favorably to shunting prompted an aggressive treatment plan, with the assumption that symptomatic deterioration was reversible. Without this history of previous shunt responsiveness, it is less clear which patients should be treated aggressively. The rationale behind, and the response to, the treatment method used in this study...
suggests that some patients are at the far end of the intracranial compliance spectrum, and I was characterized by poor responsiveness to standard shunting procedures.

There is no consensus regarding the most appropriate valve to use for the treatment of adult hydrocephalus. One dilemma is that use of the low differential pressure valve, considered the most effective valve, likely predisposes the patient to the highest risk of an overdrainage-related complication. Efforts to reduce this have focused on reproducing “normal” ICP dynamics, typically by adding an ASD, siphon-control device, or differential flow-limiting device. The efficacy of these devices in preventing complications or treating normal pressure hydrocephalus has yet to be demonstrated.

Because of the relatively high frequency and serious morbidity associated with overdrainage complications, we do not suggest that a low differential pressure valve is the optimal first choice based on its siphoning characteristics. A more logical and safe approach may be afforded by programmable valves. Adult hydrocephalus patients could be started with the valve set in the high-pressure position, then slowly adjusted downward based on clinical response. In patients who do not respond at the lowest pressure setting, a treatment protocol similar to that described herein might be considered. However, each patient in this series had responded to CSF shunting previously, and therefore it is not known whether this treatment protocol is applicable to patients with newly diagnosed normal pressure hydrocephalus. Furthermore, progressive subatmospheric ventricular draining protocol is not without risk and typically entails a prolonged intensive care unit stay. This is emotionally taxing for the patient and carries a significant health care expense. The nursing staff must be familiar with the concept of siphoning, and the utmost attention must be given to preventing a ventriculostomy infection. Further studies will be required to assess the utility of this management strategy. Nevertheless, the experience gained with the treatment of these 4 patients significantly adds to our understanding of how and why CSF shunting works in adult hydrocephalus.

In conclusion, it cannot be assumed that a patent shunt and low normal ICP are diagnostic of a failure to respond to CSF shunting. In patients in whom there is a high clinical suspicion that they are symptomatic because of ventriculomegaly, a treatment plan aimed at reducing ventricular size by achieving sufficiently negative intraventricular pressure should be initiated.

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REFERENCES