Treatment of Parenchymatous Degeneration of the Brain by Ventriculo-Atrial Shunting of the Cerebrospinal Fluid

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Dementia of the elderly has, until now, been considered a hopeless condition. The socio-economic factors of the institutional care of these patients in an increasingly elderly population make this a problem worthy of intensive study.

Recently, some cases of dementia with psychomotor retardation and ataxia associated with "normal cerebrospinal fluid pressure" and enlarged ventricular systems have been described in which ventriculo-atrial shunting of the cerebrospinal fluid has led to dramatic improvement in mental function. In these rare patients, there was no evidence of complicating intrinsic cerebral disease. The clinical picture of "symptomatic occult hydrocephalus with normal cerebrospinal fluid pressure" was attributed to the hydrocephalus.1,2 Patients with senile dementia or other degenerative brain disease often present a similar clinical picture and also have an enlarged ventricular system which is attributed to wasting of brain tissue, the so-called hydrocephalus ex-vacuo. We have employed ventriculo-atrial shunting in five patients with hydrocephalus ex-vacuo and parenchymatous degenerative disease of the brain in an attempt to alter the relentless progression of their disease.

To document the changes brought about by the operation, motion pictures, psychometric tests, and pneumoencephalograms were carried out before the ventriculo-atrial shunt and 8 weeks later. To make the postoperative assessment of mental function as objective as possible, the tests were conducted by psychologists not involved in the decision regarding the desirability of surgery in these patients. We are reporting the clinical histories and responses to ventriculo-atrial shunting in two of our patients, and summarizing the results in all five patients (Table 1).

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Case Histories

Case 1. A 73-year-old accountant had experienced 7 years of mental deterioration, slowness, incontinence, unsteady gait, and memory loss before admission to the hospital after a subarachnoid hemorrhage in January, 1966. A ventriculo-atrial shunt was carried out in March, 1966, which resulted in a remarkable restoration of mental and motor function and a return of continence.

First admission. The patient was first examined at the Cincinnati Veterans Administration Hospital in January, 1964. During the previous 5 years, slow progressive mental deterioration had been noted by his wife. Two months before admission, the patient had an episode of confusion and lethargy. A subarachnoid hemorrhage was suspected but a lumbar puncture was not performed at another hospital. General examination was normal except for a blood pressure of 180/110. He was not oriented in time or place but could answer simple questions. He could not subtract 7 from 20 and could not recall an address after 3 minutes. The neurological examination was otherwise normal, except for a broad-based, slow, shuffling gait, and bilateral Babinski responses. A lumbar puncture showed an opening pressure of 120 mm of water and clear CSF. Bilateral common carotid angiograms were normal. In February, 1964, psychometric tests showed a halting circumstantial speech; he would start a sentence, stop in the middle, and then continue on an entirely different subject. He had difficulty in understanding what was required of him and tests had to be abandoned because he could not perform any given task. No assessment of mental function or I.Q. was possible. He was discharged in February, 1964.

Second admission. The patient was readmitted on January 28, 1966. His memory had deteriorated further, and he had frequent episodes of fecal and urinary incontinence.
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**TABLE 1**

*Summary of clinical features, pneumoencephalography, and results of ventriculo-atrial shunting of CSF in patients with parenchymatous degeneration of the brain*

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Age &amp; Sex</th>
<th>Clinical Diagnosis</th>
<th>Clinical Manifestations</th>
<th>Duration</th>
<th>Pneumoencephalography</th>
<th>Results of Operation</th>
<th>Follow-Up</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>79 M</td>
<td>Alzheimer's disease</td>
<td>Dementia, loss of memory, ataxia, incontinence</td>
<td>7 yrs</td>
<td>Dilatation of ventricles</td>
<td>No change</td>
<td>Marked improvement in mental function, gait, continence</td>
</tr>
<tr>
<td>2</td>
<td>54 M</td>
<td>Alzheimer's disease, Peripheral Neuropathy</td>
<td>Dementia, loss of memory, ataxia</td>
<td>7 yrs</td>
<td>Dilatation of ventricles, cortical atrophy</td>
<td>Slight reduction in ventricular size</td>
<td>Marked improvement in mental function and gait</td>
</tr>
<tr>
<td>3</td>
<td>56 M</td>
<td>Alzheimer's disease</td>
<td>Dementia, mute, unable to swallow, pressure sores, incontinent, bedridden, contractures</td>
<td>8 yrs</td>
<td>Dilatation of ventricles, cortical atrophy</td>
<td>No change</td>
<td>Marked improvement in memory, return of limited vocabulary, eats unaided, sits in chair, healing of pressure sores</td>
</tr>
<tr>
<td>4</td>
<td>72 M</td>
<td>Creutzfeldt-Jakob Disease</td>
<td>Dementia, incontinence, few words left, bedridden, pressure sores, myoclonic jerks</td>
<td>8 mos</td>
<td>Dilatation of ventricles, cortical atrophy</td>
<td>Postmortem study; slight decrease in ventricular size</td>
<td>Improvement in swallowing, return of some sentences, disappearance of myoclonus</td>
</tr>
<tr>
<td>5</td>
<td>55 M</td>
<td>Huntington's Chorea</td>
<td>Dementia, severe chorea; institutionalized</td>
<td>6 yrs</td>
<td>Absence of head of caudate nucleus</td>
<td>No change</td>
<td>No change</td>
</tr>
</tbody>
</table>

* Autopsy findings: Carcinoma of lung, multiple pulmonary abscesses, Creutzfeldt-Jakob disease.

Eight days before admission, he had become listless and complained of headache, and his gait had become worse. When admitted, the blood pressure was 140/80 in the right arm. He did not know the date, season, or where he was. There was marked perseveration, apraxia, finger agnosia, right-left confusion, and he would frequently fall asleep. Motor power was good in the arms and legs, but the patient could not perform any purposeful movements. Deep tendon reflexes were normal and bilateral Babinski responses were present. A lumbar puncture on the day of admission showed xanthochromic fluid, opening pressure at 260 mm of water, 1,300 red blood cells, and no white blood cells per high-power field. Gait was tested 4 weeks after admission; at this time he could not walk or stand unsupported. A pneumoencephalogram showed symmetrical dilation of ventricles (Figs. 1, 2). Psychometric tests on February 25, 1966, showed bizarre responses to questions and given tasks; again, no I.Q. estimate was obtainable.

**Ventriculo-atrial shunt.** On March 18, a ventriculo-atrial shunt with a medium pressure Spitz-Holter valve (opening pressure 30-40 mm CSF) was performed. Eight weeks after the shunt, the patient was able to walk unaided and had become continent. Psychometric testing on May 24, 1969, showed him cooperative and able to concentrate for long periods of time; the Wechsler I.Q. was 111. A repeat pneumoencephalogram showed no change in the ventricular system. He is now completely recovered.

The occurrence of a subarachnoid hemorrhage 5 years after the onset of symptoms may have contributed to the accelerated decline in mental function. There is, however, no doubt that the primary disorder was parenchymatous degeneration of the brain.

**Case 2.** A 54-year-old man had a 7-year history of loss of memory, ataxia, and difficulty in walking, and had had to use a cane for 1½ years. He was forced to retire from his job as manager of a gambling casino because of his difficulty with numbers.

**First admission.** The patient was first admitted to our hospital in November, 1964. The general examination was normal.
logical examination showed he was oriented in time and place, but could not repeat a sentence containing two names and a number. His gait was wide-based and shuffling, and he could not turn without help. His motor power and tone were normal; a left Babinski response was present. The radioactive brain scan was normal, but pneumoencephalograms showed symmetrical dilation of the lateral ventricles, enlargement of the cisternae magna and pontis, and dilated sulci over the convexity of the hemispheres. He was thought to have degenerative brain disease of undetermined etiology.

Second admission. In October, 1965, after a fall 2 days earlier, the patient was admitted to the hospital. His disability had progressed so that he could not walk or solve simple problems. Blood pressure was 140/90. The cranial nerves were intact. Strength, when tested recumbent, was normal. There were bilateral Babinski responses; pain, touch, pinprick, and joint position were impaired in the feet up to about mid-calf and were normal throughout the rest of the body. Lumbar-puncture opening pressure was 130 mm of water; the cerebrospinal fluid protein was 55 mg%. A repeat pneumoencephalo-

Fig. 1. Case 1. Preoperative (left) and postoperative (right) pneumoencephalograms show no change in ventricular size after ventriculo-atrial shunting.

Fig. 2. Case 1. Preoperative (left) and postoperative (right) pneumoencephalograms show no change in depth of cerebral mantle.
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FIG. 3. Case 2. Preoperative (left) and postoperative (right) pneumoencephalograms show a slight reduction in the size of the lateral ventricles. The cerebral mantle has increased from 2.5 to 3 cm in depth.

gram (Fig. 3 left) was similar to the one performed a year earlier. Psychometric tests showed that he had difficulty in reproducing simple patterns and dots or figures. A formal assessment of intelligence quotient was very difficult.

A ventriculo-atrial shunt was performed on November 23, 1965. Eight weeks later, the patient was able to walk without a cane, and showed considerable improvement in mental alertness. Pneumoencephalograms showed a slight reduction in the size of the lateral ventricles (Fig. 3 right). Four months later, he no longer needed to be cared for by his children, with whom he had been living for the past 3 years, and returned to his club, coping well with his personal needs and able to walk without assistance.

Discussion

Improvement after ventriculo-atrial shunting was evident in all patients with markedly enlarged ventricular systems and cortical atrophy, but one patient with a small ventricular system and parenchymatous degeneration of the brain (Case 5) was not helped by the operation (see Table 1). The most striking change was in alertness, motor performances, and intellectual capacity. Some patients who required institutional care have been discharged to their previous environment after the operation (Cases 1 and 2).

In patients with “symptomatic normal pressure hydrocephalus,” disturbance of CSF circulation has been blamed for the dementia; clinical improvement after ventriculo-atrial shunting occurs concurrently with a decrease in ventricular size. Since there was no intrinsic disease of the brain, the symptomatology was attributed to the enlarged ventricles.

Contrary to the findings in patients with normal-pressure symptomatic hydrocephalus, our patients with hydrocephalus ex-vacuo showed clinical improvement without significant change in the size of the ventricular system. This supports the view that brain substance had been lost and that the enlarged ventricles themselves did not con-
tribute to the clinical picture. Since the patient with Huntington’s chorea showed no improvement after the shunting, it appears that lowering of the CSF pressure alone in the presence of a normal-sized ventricular system has no effect on parenchymatous degenerative brain disease. Since the amount of CSF secreted remains constant over a wide range of pressures, it is unlikely that a change in amount was responsible for the clinical improvement. The lowering of the CSF pressure by the operation and unchanged ventricular size would be reflected in a diminution of the force exerted by the CSF on the ventricular walls (force = pressure X area).2

By exclusion, then, we postulate that in patients with hydrocephalus ex-vacuo a lowering of the total force applied by the CSF to the brain might have led to the clinical improvement. This, in turn, could result in an increase in blood supply to the brain, which might account for some return of function in the remaining neurons. From the results reported here, it would seem that the best results from ventriculo-atrial shunting in patients with parenchymatous degenerative brain disease can be expected only in cases of marked dilatation of the ventricular system. Ventriculo-atrial shunting may not be a cure, but a return of continence, ability to walk, and some capacity to manage affairs, however short-lived, is worthwhile, particularly if achieved by a relatively simple surgical procedure.

Summary

We have discussed how ventriculo-atrial shunting has been associated with a spectacular improvement in mental and motor functions of some patients with parenchymatous degenerative diseases of the brain and hydrocephalus ex-vacuo. We have postulated that the procedure lowers the “force” exerted on the brain by the cerebrospinal fluid and thus improves the blood supply to the remaining neurons. We have presented the clinical histories of two patients in detail and summarized the results in three others.

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References