Further Experience with the Syndrome of "Normal" Pressure Hydrocephalus*


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During the past 10 years, it has become possible to separate from the large number of adults with dementia a small group whose symptoms are related to normal pressure hydrocephalus. The typical early clinical picture is characterized by the gradual development over weeks to months of mild impairment of memory, physical and mental slowness, unsteadiness of gait, and urinary incontinence. With the patient lying on his side, the CSF pressure measured in the lumbar subarachnoid space is less than 180 mm of water. Pneumoencephalography (PEG) reveals marked ventricular enlargement and, in classic cases, a lack of filling of the subarachnoid space over the cerebral convexities. Following this procedure, there is often a marked increase in the symptoms and signs. Improvement follows a shunt operation. This report analyzes our surgical experience with a total of 28 cases, including five previously reported.1,2,9

Results

For purposes of analysis, we have divided the cases into three large groups (Table 1). Group 1 includes those cases of complete or partial cerebrospinal fluid (CSF) obstruction in which no definite etiology could be established as the cause of the enlarged ventricular system and neurological symptoms. In Group 2 are the cases of complete or partial CSF obstruction in which specific etiological factors were present; the clinical syndrome, at times, was superimposed on some degree of residual deficit due to the underlying disease process. Group 3 includes cases of atrophic (ex vacuo) hydrocephalus without obstruction of the CSF flow (Alzheimer's disease suspected).

Group 1 is subdivided into three categories. Group 1 A consists of patients with the typical clinical findings and radiographic features of the syndrome of normal pressure hydrocephalus and a good response to surgical treatment. All were females in the 6th or 7th decades. Group 1 B had the same clinical features but only partial obstruction of CSF flow, the PEG revealing some air in the Sylvian or frontal subarachnoid space but none over the parietal convexities. A fair-to-good response results from a shunting procedure. Group 1 C includes patients with a clinical picture suggesting normal pressure hydrocephalus but lacking some of the characteristics of the complete clinical syndrome. Partial obstruction of CSF flow was demonstrated, but little or no response to shunting occurred.

The histories of the cases in Group 2 revealed a definite etiological factor as the probable cause of the enlarged ventricular system and CSF obstruction. Group 2 A includes four cases related to subarachnoid hemorrhage; Group 2 B, four post-trauma cases; Group 2 C, one patient with a third ventricle tumor previously reported, and one with aqueduct stenosis, possibly due to tumor.

Group 3 contains five patients with dementia in whom the clinical diagnosis of Alzheimer's disease was made. These cases had marked ventricular enlargement, and, in addition, air outlined dilated cerebral subarachnoid spaces, with no evidence of obstruction of CSF flow. In this group of cases with atrophic hydrocephalus there was no response to treatment.

Except for two patients in whom a Torkildsen procedure was performed, all patients were treated with a ventriculovenous shunt.

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There was no shunt infection in any of these cases. Five revisions were required because of obstruction of the ventricular catheter.

We now use the Hakim valve, usually in the medium pressure range, with a closing pressure of 50 to 70 mm. It has been reported¹⁸ that this valve system provides a greater margin of safety than other valves in cases with variations of CSF volume or protein. A summary of our initial experience with this valve in the treatment of all forms of hydrocephalus has been reported.¹⁶

**Selected Case Reports**

**Group 1 A**

**Case 1.** This 63-year-old woman was well until October, 1958, when she had a peculiar episode of generalized weakness and fatigue. Examination was normal but she subsequently developed unsteadiness of gait and persistent fatigue, and complained of inability to think clearly, poor memory, and slowness of thought. Urinary incontinence developed.

Examination revealed slowness in mental responses, decrease in attention and concentration, and lack of spontaneity. She walked slowly on a normal base with short steps, and in walking tandem lurched slightly to one side or the other. The deep tendon reflexes were brisk in the legs, and at times the left plantar reflex was absent or suggestively extensor.

The lumbar CSF pressure was 174 mm. A PEG revealed enlarged ventricles (span greater than 70 mm) and no surface air. Following this procedure she became worse. Lumbar CSF pressure at 1 and 2 weeks after the procedure was 180 mm, but 3 weeks later was 300 mm. Four weeks later ventricular pressure was 210 mm.

A ventriculoalovenous shunt was placed. Thought process returned to a near normal level and walking improved. Approximately 6 weeks later, she fell, fracturing her femur, and subsequently had marked mental deterioration. The lumbar CSF pressure was 240 mm. On reopening the operative wound, it was found that the plastic tube between the valves had been broken, presumably by the fall. With replacement of this valve, the CSF pressure was reduced to a range of 115 to 130 mm, and mental function again returned to normal.

One month after the last operation specific tests using the Wechsler Adult Intelligence Scale and Wechsler Memory Scale were superior in both verbal and nonverbal areas, and general memory was very superior, a marked improvement over the preoperative tests. Further improvement was documented 3 years later. The patient remains well and in good health 9 years after operation.

**Case 2.** This 66-year-old woman developed unsteadiness of gait followed by mental deterioration at about the same time she was treated for carcinoma of the breast. Initial evaluation revealed her to be confused, with poor memory. A PEG showed ventricular enlargement (span 60 mm) with no air over the surface. Following this procedure mental