The syndrome of normal-pressure hydrocephalus

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A series of 40 patients with the syndrome of normal-pressure hydrocephalus is presented. Diagnosis was based on clinical and computerized tomography (CT) scan criteria and was followed by the insertion of a ventriculoperitoneal shunt in every instance. All patients responded favorably to treatment (four of them had a fair outcome), and this response was maintained. There was one postoperative death, but no other serious complications. This study demonstrates that it is possible to diagnose the syndrome of normal-pressure hydrocephalus on the basis of clinical and CT scan criteria without any other invasive investigations. Such patients should respond favorably to an adequately functioning shunting system.

KEY WORDS • normal-pressure hydrocephalus • computerized tomography • ventriculoperitoneal shunt • hydrocephalus

The term "normal-pressure hydrocephalus" (NPH) is used to describe a clinical syndrome mainly comprising gait disturbance, dementia, and urinary incontinence, and is associated with dilatation of the ventricular system of the brain and normal cerebrospinal fluid (CSF) pressure at lumbar puncture. Since the initial reports from the Massachusetts General Hospital, numerous series of patients apparently suffering from this syndrome have been described in the literature. In these studies, attention has mainly concentrated on establishing diagnostic criteria and prognostic factors that would reliably predict a favorable outcome following a ventricular shunting procedure.

In this report, we present the results of treatment of 40 patients with the syndrome of NPH. The questions we have attempted to answer include: Are there any criteria on the basis of which we can select with certainty at least some of the patients with the syndrome? Are there any patients with the syndrome who do not respond favorably to a shunting procedure? and Why do they not respond?

Clinical Material and Methods

The study concerns 40 patients in whom the diagnosis of NPH was based on clinical and computerized tomography (CT) criteria. All the patients were hospitalized in either the Neurosurgical or the Neurological Departments of the Army General Hospital or the Army Veterans Administration Hospital between January, 1979, and August, 1983. For treatment and inclusion in the study, patients must have had a clinical history and symptomatology suggestive of the syndrome and the demonstration of a dilated unobstructed ventricular system on the CT scan, together with some additional features which excluded dilatation ex vacuo of the ventricular system. Lumbar puncture with measurement of the CSF pressure was not considered necessary and was therefore performed according to the needs of each patient. Apart from routine investigations, no other tests were performed before treatment was undertaken.

In this series of patients we did not consider separating patients with a known cause of their communicating hydrocephalus from those with an unknown etiology (idiopathic). The reasons for this are discussed below.

Patient Population

Almost half of the patients were selected from a pool of patients with otherwise unexplained gait disturbance and dementia. The remainder were mostly neurosurgical patients under care or being followed for other intracranial disorders. There were 24 patients with communicating hydrocephalus of known etiology, and 16 in whom no cause for the hydrocephalus could be identified from either the investigations or the patient's medical history, based on information given mainly by the relatives (Table 1).

Gait disturbance was present in every patient and ranged from a mild deficit in balance to complete inability to walk or even stand. Excluding the four
patients with subarachnoid hemorrhage (SAH) who were bedridden at diagnosis, in every instance the onset of the disability was insidious and required some time to become apparent to the patient or his relatives. This period of time was shorter in the group with known etiology (several weeks to 1 year, with one exception) than in the idiopathic group (1 to 4 years). In four patients in the idiopathic group, inability to walk was the only presenting symptom, save for a mild memory deficit considered “normal” for the patient’s age.

Frank dementia (akinetiC mutism in three cases) was the predominant feature in eight patients. Impairment of memory, mainly short-term, slowing of thought, and lack of attention or initiative were the most common forms of disturbance in the remaining 24 cases. In four patients the dementia antedated the disturbance of gait by several months to years; in fact it dominated the clinical picture. One of these patients had suffered severe ischemic damage to the hypothalamus and the medial aspects of the frontal lobes following rupture of a giant anterior communicating artery aneurysm, another had a severe closed head injury, residual aphasia, hemiparesis, and personality changes, and two patients were in the idiopathic group. These patients are discussed separately, as they form the group of “fair” responders to a shunting procedure.

Urinary incontinence was present in almost half the patients (19 out of 40), and in every instance it followed disturbance of gait and mentation. Most often it took the form of lack of concern, but in a few patients it appeared as urgency of micturition.

Other symptoms or signs, such as headache, dizziness, visual disturbances, or pyramidal or extrapyramidal signs, were less frequent and are not considered in this study. In three patients (two with SAH and one with a history of head injury), the diagnosis of the syndrome was suspected when their preexisting neurological deficits began to deteriorate. Interestingly, their CT scans demonstrated, in addition to ventricular dilatation, the presence of areas of low density roughly corresponding to the underlying ischemic white matter which disappeared following treatment. Finally, failure to make a satisfactory recovery following SAH or surgery for intracranial tumor (meningioma or pituitary adenoma) led to the correct diagnosis in four other patients.

Computerized Tomography Findings

Most of the CT scans were obtained on an Ohio Nuclear “Delta” head scanner and displayed on a 160 × 160 matrix. In the eight most recent cases, a Pfizer AS and EO450 body scanner was also used.* Four CT scan criteria were used in this study, based on our previous experience and the experience of others. These criteria were the presence on the CT scan of: 1) dilatation of the unobstructed ventricular system; 2) obliteration of the cerebral sulci; 3) areas of periventricular low density; and 4) “rounding” of the frontal horns of the lateral ventricles. We considered these characteristics as indicating that the coexisting ventricular enlargement was not an ex vacuo variant. Subsequently, only patients satisfying at least one of the first two criteria, in addition to the ventricular dilatation, were offered treatment and included in the study.

Dilatation of the unobstructed ventricular system was present in every patient and was considered a prerequisite for treatment and inclusion in the study. Only patients with more than moderate ventricular enlargement were selected, in order to overcome arguments regarding the actual size of the ventricles. Thus, only patients in whom the ratio of the width of the lateral ventricles at the level of the foramina of Monro to the transverse inner diameter of the skull at the same level was above 1.5 were included.

Obliteration of the cerebral sulci (Figs. 1, 2, and 3), as seen in the uppermost CT cuts, was present in 31 cases (Table 2); thus, no patients were excluded from the study on the basis of the absence of this criterion.

* Delta scanner 25 manufactured by Ohio Nuclear, Inc., 2910 Aurora Road, Solon, Ohio. Pfizer AS and EO450 body scanner manufactured by Pfizer Medical Systems, Inc., 9052 Annapolis Road, Columbia, Maryland.

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**TABLE 1**

*Clinical features in 40 patients with the NPH syndrome*

<table>
<thead>
<tr>
<th>Etiology</th>
<th>No. of Cases</th>
<th>Age Range</th>
<th>Duration of Symptoms (range)</th>
<th>Deterioration†</th>
<th>Mutism</th>
<th>Gait Disturbance</th>
<th>Dementia</th>
<th>Urinary Incontinence</th>
</tr>
</thead>
<tbody>
<tr>
<td>SAH</td>
<td>10</td>
<td>20–63</td>
<td>3 wks–10 mos</td>
<td>4</td>
<td>2</td>
<td>6‡</td>
<td>8</td>
<td>7</td>
</tr>
<tr>
<td>craniotomy</td>
<td>6</td>
<td>42–64</td>
<td>3 wks–2 yrs</td>
<td>2</td>
<td>0</td>
<td>6</td>
<td>5</td>
<td>2</td>
</tr>
<tr>
<td>CNS infection</td>
<td>3</td>
<td>15–58</td>
<td>2 mos–1 yr</td>
<td>0</td>
<td>0</td>
<td>3</td>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>head injury</td>
<td>5</td>
<td>58–75</td>
<td>6 mos–10 yrs</td>
<td>1</td>
<td>1</td>
<td>4‡</td>
<td>4</td>
<td>3</td>
</tr>
<tr>
<td>unknown (idiopathic)</td>
<td>16</td>
<td>52–80</td>
<td>1–4 yrs</td>
<td>0</td>
<td>0</td>
<td>16</td>
<td>12</td>
<td>6</td>
</tr>
<tr>
<td>total cases</td>
<td>40</td>
<td></td>
<td></td>
<td>7</td>
<td>3</td>
<td>3‡</td>
<td>32</td>
<td>19</td>
</tr>
</tbody>
</table>

* NPH = normal-pressure hydrocephalus; SAH = subarachnoid hemorrhage; CNS = central nervous system.
† Deterioration of preexisting deficits or failure to make a good recovery.
‡ The remaining patients were bedridden.
Normal-pressure hydrocephalus

Fig. 1. The chief computerized tomography characteristics in a 70-year-old woman with normal-pressure hydrocephalus syndrome. a and b: Marked ventricular dilatation and “rounding” of the frontal horns of the lateral ventricles are seen. c: Periventricular areas of low density are indicated by arrows. d: The cerebral sulci are obliterated (arrows).

Fig. 2. Obliteration of the cerebral sulci (arrows) in a 72-year-old man with ventricular enlargement. Note the considerable dilatation of the subarachnoid spaces at the lower levels (including the Sylvian fissures).

Fig. 3. Left Pair: Obliteration of the cerebral sulci in a 58-year-old man with the normal-pressure hydrocephalus syndrome (small arrow). Note the widening of the Sylvian fissures (large arrow). Right Pair: Reappearance of the cerebral sulci 1 year following treatment (small arrows). Note the diminution in the size of the Sylvian fissures (large arrow).

The width of the cerebral sulci and its predictive value regarding response to treatment have been discussed previously. In our study, obliteration of the sulci was sometimes observed in combination with dilatation of the subarachnoid spaces at lower levels, including the Sylvian fissures (Figs. 2 and 3). This finding was considered to indicate obstruction of the subarachnoid space of the convexity (Table 2). In a few cases, which were followed postoperatively with CT scanning, reappearance of the cerebral sulci was documented (Fig. 3), and the validity of this criterion was reinforced.
**TABLE 2**  
**Computerized tomography characteristics in 40 patients with the NPH syndrome***

<table>
<thead>
<tr>
<th>Etiology</th>
<th>No. of Cases</th>
<th>&gt; Moderate Ventricular Enlargement</th>
<th>Obliteration of Cerebral Sulci</th>
<th>Periventricular Areas of Low Density</th>
<th>&quot;Rounding&quot; of Frontal Horns</th>
<th>Cortical Atrophy</th>
</tr>
</thead>
<tbody>
<tr>
<td>SAH</td>
<td>10</td>
<td>10</td>
<td>8 (2)†</td>
<td>8</td>
<td>7</td>
<td>1</td>
</tr>
<tr>
<td>craniotomy</td>
<td>6</td>
<td>6</td>
<td>4 (1)†</td>
<td>5</td>
<td>5</td>
<td>1</td>
</tr>
<tr>
<td>CNS infection</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>3</td>
<td>-</td>
</tr>
<tr>
<td>head injury</td>
<td>5</td>
<td>5</td>
<td>4 (2)†</td>
<td>3</td>
<td>4</td>
<td>2</td>
</tr>
<tr>
<td>unknown (idiopathic)</td>
<td>16</td>
<td>16</td>
<td>12 (4)‡</td>
<td>8</td>
<td>9</td>
<td>4</td>
</tr>
<tr>
<td>total cases</td>
<td>40</td>
<td>40</td>
<td>31 (9)‡</td>
<td>27</td>
<td>28</td>
<td>8</td>
</tr>
</tbody>
</table>

* NPH = normal-pressure hydrocephalus; SAH = subarachnoid hemorrhage; CNS = central nervous system.
† Numbers in parentheses indicate cases with dilatation of the subarachnoid spaces at lower levels (Sylvian fissures included).

In one SAH patient, in one head-injured patient, and in two in the idiopathic group, cerebral sulci were clearly visible in the uppermost cuts, but in the remaining cases no firm judgment could be made for technical reasons (including movement artifacts or no available uppermost CT cuts). The combination of the clinical picture and the presence of areas of periventricular low density in these last patients was, however, considered sufficient for inclusion of the patients in the study (Fig. 4). This feature was taken to indicate transependymal movement and absorption of CSF, which is known to take place in communicating hydrocephalus. Granholm has shown that periventricular areas of low density are present in cases of hydrocephalus and disappear following shunting. This was subsequently confirmed by others. Overall, 27 patients exhibited periventricular low-density areas on CT scanning, and this was taken as an absolute indication for treatment in every patient (Figs. 1 and 4). Interestingly, this feature was seen in four of our patients with clearly visible cortical sulci (Fig. 4), suggesting that obliteration of the subarachnoid space in these areas is not necessarily present in every patient with this syndrome, and that it is at least partially dependent on the actual size of the cerebral gyri.

Finally, "rounding" of the frontal horns of the lateral ventricles (Fig. 1) was demonstrated in 28 patients and was considered as equivalent to the pneumoencephalographic (PEG) finding that the greater impact of ventricular dilatation is on the anterior portions of the ventricular system. This feature did not serve as an absolute indication for treatment in this series.

An interesting group of eight patients, half of whom were in the idiopathic category, showed CT features suggestive of cortical atrophy. This as a rule was better seen on CT cuts lower than the uppermost ones; for example, cuts through the upper part of the lateral ventricular bodies. No attempt was made to distinguish these appearances etiologically, as this was beyond the scope of this study. It is emphasized, however, that in every patient of this group the combination of clinical and additional CT scan features such as those mentioned above suggested that the coexistent ventricular dilatation was at least partially due to obstruction of

![Fig. 4. Areas of periventricular low density (arrows) in a 75-year-old patient with ventricular dilatation 6 months following head injury. The cerebral sulci are clearly visible.](image-url)
Normal-pressure hydrocephalus

the CSF bulk flow, and required shunting. This was shown to be the case in every patient of this group, even in the three patients who showed only a fair response to treatment.

Management

Following diagnosis and routine preoperative investigations (chest radiograph, electrocardiogram, blood biochemical analysis, blood count and hematocrit, and urinalysis), all the patients were given prophylactic flucloxacinil (Floxapec) in a dose of 500 mg three times daily for 1 day pre- and 2 days postoperatively. Surgery was performed under general anesthesia. A ventriculoperitoneal shunting system was installed in every patient, incorporating a medium-pressure Holter valve and a standard Rickham reservoir.† The standard surgical technique was used, with special care taken that no part of the device came in contact with the patient’s skin. Operative time ranged from 50 to 70 minutes. Patients were encouraged to leave their bed in the 2nd postoperative day and were usually discharged home between the 8th and the 10th day. Follow-up examination was performed at 3 and 6 months, at 1 year, and thereafter every year.

Results

Response to treatment was graded according to the criteria proposed by Black,6 which are as follows:

Excellent: resumed pre-illness activity without deficit
Good: resumed pre-illness activity with moderate deficit
Fair: improved but no return to previous work
Poor: no change or worse.

Table 3 summarizes the results of treatment. Thirty patients achieved an excellent result. Of the remaining 10, six were judged as good responders in that they obtained complete resolution of gait disturbance but not the memory deficit. They were nevertheless able to return to their previous activities. Four patients showed only a fair response to treatment. These patients deserve special mention. The first patient was a 23-year-old man who suffered two SAH’s within 1 month from rupture of a giant anterior communicating aneurysm. He was referred with a history of gait disturbance of 10 months’ duration and a longer history of short-term memory deficit. He was also incontinent of urine. A CT scan showed ventricular dilatation with periventricular lucencies and visible cerebral sulci (Fig. 4). A few days following surgery he began to converse and take a few steps with support, but 6 months after surgery he remains dysphasic with right hemiparesis, and at times he is incontinent of urine. The third patient was a 72-year-old man, who presented with a 3-year history of gait disturbance leading to inability to walk or stand and frank dementia of 2 years’ duration. He was also incontinent of urine. A CT scan demonstrated some cortical atrophy, ventricular enlargement, and definite obliteration of the cerebral sulci, together with dilatation of the subarachnoid spaces at lower levels (Fig. 2). One year after surgery, this patient walks normally; he is continent of urine but continues to have severe memory deficit.

The last patient in this group, a 73-year-old man, was referred with a history of gait disturbance of 10 months’ duration and a longer history of short-term memory deficit and mild dementia. His medical history included coronary artery bypass surgery for myocardial ischemia and two mild ischemic strokes of the right cerebral hemisphere. A CT scan showed ischemic lesions and cortical atrophy in both hemispheres and ventricular dilatation combined with obliteration of the cerebral sulci. Soon after surgery he was able to walk almost normally (and he remains able to do so); however, his dementia remains unchanged 6 months postoperatively.

There were four additional patients (two with SAH and two in the idiopathic group) who showed an initial satisfactory response to surgery only to return to their preoperative level in 1 to 2 days. In every instance, palpation of the valve showed that it emptied and filled

† Medium-pressure Holter valve and Rickham reservoir, supplied by Codman and Shurtleff, Inc., Randolph, Massachusetts.
properly, and the patients' postoperative CT scans showed no change in the size of the ventricles and no additional features. Pumping of the valve was then started three times daily (30 pumpings each time) as inadequate function of the system was suspected. This produced a definite improvement in the patients' condition within 48 hours. Improvement was dramatic in two cases. Eventually, three of these patients achieved an excellent and one a good response to treatment. Pumping, performed by instructed relatives, was successfully discontinued in two patients in 3 months, and is continuing in the rest (6 months and 1 year later, respectively).

There was one postoperative death in this series. This was in an 80-year-old man with a 4-year history of gait disturbance leading to inability to stand or walk; he had no other symptoms. He responded dramatically to the shunting procedure, and was able to walk on the 3rd postoperative day. He was discharged home on the 9th postoperative day, but died from bronchopneumonia in another hospital 5 weeks later.

Obstruction of the proximal end of the shunt system was diagnosed in one patient and the ventricular catheter was revised. Four patients developed postoperative urinary tract infections or bronchopneumonia, and were treated successfully with the appropriate antibiotics. There were no cases of shunt infection or postoperative subdural hematomas.

**Discussion**

Eighteen years since its initial description,2,27 the syndrome of normal-pressure hydrocephalus continues to present questions, mainly regarding reliable diagnostic criteria and prognostic factors associated with a shunting procedure.6,22,36,53,56 There is little disagreement about the clinical symptoms that constitute the syndrome. Disturbance of gait, dementia, and urinary incontinence are regarded as the predominant signs.1,2,16,27 The demonstration of a dilated ventricular system on neuroradiological studies is the cardinal feature, but it is now well appreciated that the CSF pressure is not normal in these patients, at least not continuously.9,11,22,28,48,54,55 The demonstration of a definite and at times dramatic improvement of a patient's symptoms following a shunting procedure1,2,27,46 has stimulated tremendous interest in this treatable cause of dementia.10

The initial enthusiasm51 has gradually subsided, leaving behind a voluminous amount of information. Numerous studies dealing with the syndrome and proposing criteria for selection of patients appropriate for treatment have appeared in the literature. These include PEG features,23,24,36,37,56,58,64 iodine-131-labeled human serum albumin (RIHSA) scanning patterns following intrathecal administration,12,29,37,50,58 lumbar45,48 or ventriculolumbar48 subarachnoid infusion tests, and evaluation of the patients' mental function before and after withdrawal of small amounts of CSF.17,55 Measurement of cerebral blood flow,39 CT features,6,11,22,26,36 and patterns of continuously measured and recorded intracranial pressure (ICP)10,11,22,28,48,54,55 have been proposed as predictors of a response to treatment. It is generally agreed that continuous ICP monitoring offers one of the most reliable means for selection of patients likely to respond to treatment.48 Since the pioneering work of Symon, et al.,25 many other investigators have demonstrated that the ICP in these patients is not normal, thus refuting the title of the syndrome. They also showed that the presence in the record of the so-called "B-waves" is of important prognostic significance.9,11,28,48 More recently, Børgesen, et al.8 found that measurement of the conductance to outflow of the CSF is more useful in predicting the outcome to treatment than measurement of the ICP.

Obstruction of the bulk flow of the CSF in the subarachnoid space distal to the outlet foramina of the fourth ventricle has been proposed as the initial defect in this syndrome. This eventually leads to ventricular dilatation, which is responsible for the clinical picture.7,23,27,38 Several pathoanatomical reports have substantiated this proposition, demonstrating leptomeningeal fibrosis mainly in the basal cisterns or the subarachnoid space of the convexities of the hemispheres with or without changes of the arachnoid granulations in patients with known etiology (SAH, central nervous system infection, head trauma, intracranial surgery) or unknown (idiopathic) cause of their hydrocephalus.12,13,29,49,50,57 Additional findings include ventricular ependymal disruption, periventricular white matter edema, subependymal glial reaction, and periventricular loss of myelin staining,13 features that are considered to be caused by disturbance of the CSF flow and ventricular enlargement. As transependymal and possibly transchoroidal absorption of CSF is enhanced,40,42 the initial intracranial hypertensionsubsides but the ventricles continue to enlarge, leading to destruction of periventricular nerve fibers.61 How ventricular enlargement continues in the presence of a fall in CSF pressure is difficult to explain. Hakim and Adams37 offered the proposition that, since the total force on the ventricular wall is related to both area and pressure, if the area of the ventricles is increased, a smaller pressure is required to maintain their dilated state. This theory has since been taken further by other authors.15,26,62 It is also interesting to note that experimental obliteration of the subarachnoid space leads to ipsilateral ventricular dilatation,25 which lends support to the proposition that obliteration of the subarachnoid space of the convexities contributes to an increase in cerebral mantle pressure between the ventricles and the subdural space.25,31

It is well known that some patients are found at postmortem examination to have changes due to both Alzheimer's disease and leptomeningeal fibrosis, causing obstruction of the subarachnoid space.13 Postmortem studies of patients with the NPH syndrome showing extensive hypertensive cerebrovascular disease with
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multiple small infarcts but without gross leptomeningeal changes should be viewed with caution; well-organized subdural membranes were present in two of the three reported cases and in the third the diagnosis of the syndrome was not certain.\textsuperscript{14,35}

In clinical practice, when a patient presents with gait disturbance and memory deficit (with or without urinary incontinence), a decision must be made whether a radiologically demonstrated ventricular dilatation is clinically significant and therefore requires shunting, or whether it is only the result of wasting of cerebral substance (dilatation \textit{ex vacuo}). The existing body of evidence does not offer reliable criteria on which to base with certainty the distinction between these entities.\textsuperscript{6,7,48} The basic question remains: Do any cases of NPH syndrome not respond to treatment? And if, yes, Why do they not respond? Almost all investigative efforts have been concentrated in "identifying the winners;" that is, the patients who will respond favorably to a shunting procedure.\textsuperscript{10} As a result, conclusions have been based on the response to treatment of groups of patients with ventricular dilatation fulfilling various criteria. The validity of these criteria was subsequently evaluated in retrospect according to the observed response of the patient to the shunting procedure. It is obvious that in this process two variables are at work. First, the clinical material may not be pure. It is inevitable that some patients with dilatation \textit{ex vacuo} and/or some with both the syndrome and additional cerebral pathology (such as Alzheimer's disease) escape recognition and are included in the studies. These cases should not have been included in the evaluation process because if they had been recognized at the beginning they would not have been treated by shunting. The second factor involved the effectiveness of treatment. In most of the studies, this was judged on the supposition that a properly selected and adequately functioning shunt system had been installed. It is surprising how few studies have addressed the question of evaluation of shunt function, particularly in cases of idiopathic NPH that failed to respond to treatment.\textsuperscript{4,36,56,65}

Separation of the group of patients with NPH of unknown etiology (idiopathic group) by many authors\textsuperscript{6,22,26,32,52} from those with known cause, as if different pathological mechanisms are involved in the two groups, has not in our opinion contributed to the clarification of the issues. If there are different mechanisms involved, then we must accept that we are dealing with different entities. In this respect, it is of interest to note the results of a recent study which showed similar patterns of ICP recordings in two groups of hydrocephalic patients, one with known etiology and another of the idiopathic type.\textsuperscript{11} It could, of course, be claimed that patients of the first group, as a rule, respond favorably to treatment,\textsuperscript{56} and this is not so in the idiopathic group. It is, however, also true that the diagnosis of the syndrome is usually certain from the medical history in the first group and, when response to treatment is not satisfactory, a vigorous search for the possible factors follows. This usually identifies an inadequately functioning shunt, which is subsequently revised.

In our study we attempted to investigate the possibility of using relatively simple criteria as a basis for diagnosing the NPH syndrome, avoiding invasive techniques. The clinical criteria used are in accordance with those reported previously.\textsuperscript{6,16,22,24,36,47,53,56} Gait disturbance was present in every patient (that is, no patient without this symptom was considered for treatment). As a rule, gait disturbance heralded the other symptoms,\textsuperscript{16,56} and improved in every patient following treatment. Dementia was the dominant symptom in eight patients and was present in 32. Following treatment it definitely improved in 22 patients, was slightly ameliorated in six, and remained unchanged in four (all cases with underlying additional cerebral pathology). Urinary incontinence, when present, resolved following treatment. The selection of CT scan criteria was based on published evidence,\textsuperscript{6,11,22,26,36,44,60} and on the belief that the CT scan appearance would represent existing pathological changes, as reported by previous investigators.\textsuperscript{12,13,25,29,49,50,57}

The criteria used in this study can be criticized as strict. However, considering the fact that all the patients diagnosed as suffering from the NPH syndrome on the basis of these criteria responded favorably to shunting, we conclude that they are at least valid in selecting with certainty patients who do have the syndrome. Our suggested indications for treatment are given, and the opportunity to study the response to them in a homogeneous group is offered.

With regard to treatment, we used a medium-pressure valve system in all patients in an attempt to avoid the development of subdural hematomas, which are more common when low-pressure systems are used.\textsuperscript{23} We also used the same system to standardize treatment throughout the series. The finding that four of the patients eventually required pumping of their valves in order to achieve and maintain a satisfactory result implies that in these cases a low-pressure or high-flow system was probably required. It also suggests that in these patients every effort should be undertaken to assure adequate function of the shunting system before failure of the treatment is accepted.

Four patients showed a "fair" response to treatment in that they only achieved improvement of gait disturbance. We believe that these patients represent examples of cases where the NPH syndrome is superimposed on preexisting cerebral pathology (ischemia, trauma, possible Alzheimer's disease, and multi-infarct dementia, respectively).\textsuperscript{11,19,36,56} Surgical treatment is only expected to alleviate symptoms caused by the NPH syndrome without influencing the underlying pathology, and in our opinion this is what occurred in these patients. Thus, the second conclusion that emerges from this study is that all patients with the NPH syndrome respond or should respond to the placement of an adequately functioning shunting device.
This study suggests that it is possible to base the diagnosis of the syndrome of NPH on clinical and CT criteria. Favorable response to treatment in patients so selected can be obtained without unacceptably high mortality or morbidity rates.

Addendum

Since submission of this manuscript, a relevant report has appeared (Salibi NA, Lourie GL, Lourie H: A variant of normal-pressure hydrocephalus simulating Pick’s disease on computerized tomography. Report of two cases. J Neurosurg 59:902–904, 1983). In this paper, dilatation of the Sylvian fissures, mimicking atrophy of Pick’s disease, was shown to be a feature in two cases of the NPH syndrome, both of which responded favorably to shunting.

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