Adult idiopathic communicating hydrocephalus with and without shunting

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SUMMARY The outcome in 37 adult patients with idiopathic communicating hydrocephalus treated by ventriculostriatal shunting is presented. Only 33% showed definite improvement, and no diagnostic procedures accurately predicted the outcome of surgery. These were compared with a “control” group of 12 patients who were not shunted; 50% of these were stable for up to 36 months. These findings, and the high frequency of serious complications (35%), suggest caution in recommending a shunt procedure.

Ventriculostriatal (VA) shunting for diversion of cerebrospinal fluid in adults has been used extensively since Adams et al. (1965) described an occult form of communicating hydrocephalus in adults as a potentially treatable cause of dementia. Since experience with the evaluation, definition, and treatment of this condition has not been uniform, we thought it useful to review our experience. All adult patients who underwent ventriculostriatal shunting between January 1967 and June 1974 for suspected idiopathic communicating hydrocephalus have been reviewed.

The following hypotheses were tested.
1. Normal gait predicts lack of improvement after ventriculostriatal shunting.
2. Prominent gait disorder predicts improvement.
3. Gait disorder early in the course of the illness predicts improvement.
4. Pneumoencephalographic findings “typical” of idiopathic communicating hydrocephalus predict improvement.
5. Cisternographic findings “typical” of idiopathic communicating hydrocephalus predict improvement.
6. “Typical” findings of idiopathic communicating hydrocephalus on both pneumoencephalogram and cisternogram have greater predictive value than either alone.
7. The combination of prominent gait disorder and “typical” radiographic findings of ICH is highly predictive of improvement after surgery.
8. The absence of prominent gait disorder and the lack of “typical” radiographic findings of ICH are highly predictive of a poor success rate after shunting.

In addition, we reviewed the courses of patients seen from 1972 to 1976 inclusive who met clinical and radiological criteria for idiopathic communicating hydrocephalus but who were not shunted. Although not matched with the treated patients, these were considered “control” subjects to evaluate the natural history of unoperated idiopathic communicating hydrocephalus. This matter deserves consideration while reflecting on the success and complications of ventriculostriatal shunting for this condition.

Patients and methods

Patients in this series, whether shunted or not, were considered at the time of evaluation to have idiopathic communicating hydrocephalus. While the results of diagnostic studies varied, all patients showed ventricular enlargement, and most had some cisternographic evidence for obstruction to CSF reabsorption. In the years 1967–1974 there was already conflicting opinion regarding criteria for diagnosis of idiopathic communicating hydrocephalus and debate on the indications for ventriculostriatal shunting. For the potential benefit to
dementia, surgery was recommended to patients who had some evidence of idiopathic communicating hydrocephalus by pneumoencephalography or cisternography, but not necessarily by both (Salmon and Armitage, 1968; Shenkin et al., 1973).

Two groups of patients were studied. The first ("surgical") group was composed of 40 consecutive patients who underwent ventriculostial shunting for idiopathic communicating hydrocephalus between January 1967 and June 1974 at the Washington University Medical Center. Sufficient data could not be traced for three patients. The remaining 37 were unselected patients who met the following criteria. All were above 50 years of age, had mild, moderate, or severe dementia, some radiological evidence of communicating hydrocephalus, and no known pre-existing cause for communicating hydrocephalus—for example, severe head trauma, meningitis, or subarachnoid haemorrhage. All but seven had a mild, moderate, or severe gait disorder. At the time of surgery these patients were considered to have idiopathic communicating hydrocephalus, and although a few were suspected of having a concomitant cerebral degenerative process or vascular disease, the hydrocephalus was considered to be of primary importance. These patients were treated surgically at or close to the time the diagnosis of hydrocephalus was made.

The second ("control") group was composed of 12 patients evaluated at this institution between 1972 and 1976, who were found to have idiopathic communicating hydrocephalus and who did not undergo ventriculostial shunting immediately. These patients fulfilled similar diagnostic criteria. For various reasons these patients were not shunted for many months after diagnosis (three patients) or, never underwent surgery (nine patients). Shunting was strongly considered or recommended in each case although many of these patients had other problems as well. None, however, had a pre-existing factor causing meningeal irritation.

INVESTIGATIONS
All patients were first examined to exclude other systemic disease, and while a few showed minor abnormalities of cardiac, renal, pulmonary, or hepatic function, none had significant failure of these organ systems. While other neurological tests, such as radionuclide brain imaging, electroencephalography or cerebrospinal fluid infusion tests (Trotter et al., 1974) were performed in many patients, the studies of principal interest were those neuroradiological procedures which directly evaluated the ventricular system or CSF dynamics (pneumoencephalography, computed cranial tomography, and radionuclide cisternography).

Pneumoencephalography was performed on all patients involved in this study until computed axial tomography (CAT) became available in 1974. For the purposes of this study the pneumoencephalograms were reviewed retrospectively without knowledge of the clinical outcomes. The studies were classified as either compatible with idiopathic communicating hydrocephalus or not ("other"). The pneumoencephalographic classification was defined as (1) lateral ventricles moderately or markedly enlarged, (2) no evidence of prominent cortical atrophy, and (3) no mass lesion obstructing the CSF pathways. The pneumoencephalograms labelled "other" depicted such findings as moderately or even markedly enlarged ventricles but also substantial cortical or central atrophy.

After its introduction in 1974, CAT quickly became the procedure of choice in the investigation of adult patients with suspected communicating hydrocephalus and dementia. Computerised tomography scans were performed on an EMI head unit with a vertical slice thickness of 13 mm. Most patients were studied both before and after the administration of iodinated contrast agent. Essentially the same criteria for idiopathic communicating hydrocephalus were used to interpret CAT images as had been used for pneumoencephalography (Gado et al., 1976).

Radionuclide cisternography was performed with either $^{131}$-human serum albumin or $^{111}$In-DTPA which was adopted after 1972 because of its superior physical properties for imaging and lower radiation exposure for the patient. The following criteria were used at the time of retrospective review without knowledge of clinical outcome. Ventricular reflux with retention for 48–72 hours and a definite or probable block of CSF flow in the subarachnoid space were required to confirm a diagnosis of idiopathic communicating hydrocephalus. Subarachnoid block was considered to be present if there was failure of normal ascent of the radiopharmaceutical or absence of accumulation in the parasagittal region on sequential late images obtained at least 24 hours apart. A pattern of transient ventricular reflux or delayed but complete ascent of the radiopharmaceutical to the parasagittal region or both was interpreted as probable cerebral atrophy and listed as "other." This latter classification includes: (a) cisternograms with ventricular retention of less than 24 hours and with definite subarachnoid block, (b) those with ventricular retention for more than 24 but less than 48 hours and with prob-
able subarachnoid block, (c) those with ventricular retention for more than 48 hours but without subarachnoid block, and (d) those with incomplete data.

Surgical Procedures

Ventriculoatrial shunting was performed by members of the neurosurgical group at Washington University. Initially, low pressure Hakim valves were used. Subsequently medium and high pressure Hakim valves were used as well as the multipurpose antisiphon valve (MPASV) (Heyer-Schulte Inc., Goleta, California). A few Holter valves of low and high pressure were also used. The change from the use of valves opening at lower pressure (<8 cm H2O) to those opening at higher pressures (11–14 cm H2O) resulted from the concern that the lower intracranial pressures might have been the cause of some of the complications observed.

Later Evaluation

The patients were followed up by the neurosurgery staff and by one of us (CPH) who established contact with the patients in 1974 and thereafter. At the time of the first follow-up, a minimum of nine months had elapsed after either the shunt procedure or, in the case of the “control” subjects, the evaluation suggesting idiopathic communicating hydrocephalus.

The following criteria were employed at the time of follow-up. For each of the individuals in whom a ventriculoatrial shunt was placed the course after surgery was categorised as considerably improved either in gait or mental function or both, questionably improved, stable, or worse. For those who were considerably improved it was clear both to the family and physician that the patient was walking much better and/or had improved memory and other intellectual functions. In the case of those with questionable improvement there was a subjective impression on the part of the family that the patient was better while no objective improvement was evident in the neurological evaluation. These patients were, therefore, grouped with those whose downhill course before surgery appeared to stabilise afterwards for a period. For those patients who were described as stable or questionably improved, it was possible to establish whether this stability had lasted for six months, 12 months, or longer. Those with a period of possible stability less than six months before further deterioration were considered together with the deteriorating group for purposes of shunt evaluation. For the control patients, the follow-up established whether they were improved in gait or intellectual function or both, stable, or worse.

Statistical comparisons between groups were made by the binomial test or the Mann-Whitney rank test (Snedecor and Cochran, 1967).

Results

The findings in the surgical group of 37 patients are summarised in Table 1. There were 26 men and 11 women. The mean age at shunting was 66 years (range 56–81 years). The mean duration of symptoms before operation was 24 months and the mean length of follow-up was 31 months.

All patients presented with dementia; 30 had a gait disorder; and all were treated at the conclusion of the initial evaluation. All were followed until death or for at least 18 months after surgery so that the long-term effects of the shunting procedure could be assessed. This group experienced a large number of complications which will be discussed below. Because of these complications, the effect of the shunt on the course of the disease was obscured in 10 patients. Consequently a subgroup of 27 surgical patients was selected from the original 37 in whom the follow-up period was sufficiently long (more than six months except for one patient, case 6, whose steady deterioration was clear before death 45 days after surgery) and free from complications to allow adequate evaluation of the shunting procedure.

Outcome in 27 Surgical Patients

Overall in this group, nine patients improved (33%), seven were stable (26%), and 11 became worse (41%). If the group is divided according to the clinical presentation, there are the following results. Of six patients who had no gait disorder, none improved, three remained stable (50%), and three worsened (50%). Of 13 patients who had slight gait disorder, five improved (39%), two were stable (15%), and six worsened (46%). Of eight patients who had a moderate or severe gait disorder, four improved (50%), two remained stable (25%), and two worsened (25%). Of 15 patients whose gait disorder developed before or at the same time as the dementia, seven (46%) improved, four (27%) were stable, and four (27%) worsened.

We can also subdivide the surgical group according to radiographic criteria. There were 25 who had pneumoencephalograms classified as idiopathic communicating hydrocephalus. Of these, nine improved (36%), seven were stable (28%), and nine worsened (36%). Of 16 patients who had cisternograms labelled as idiopathic communicating hydrocephalus, six (38%) improved, four stabilised (25%), and six worsened.
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Gait/dementia—0=no abnormality, 1=mild abnormality, 2=moderate or severe abnormality.
NA=Not available.
*=Effect of shunt could not be evaluated.
N=Necropsy.
B=Biopsy.
PEG=Pneumoencephalography.
ICH=Idiopathic communicating hydrocephalus.
Table 2  "Control" patients

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<th>Patient</th>
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Patient 42 was shunted nine months later. Only gait improved.
Patient 43 was shunted seven months later. Only obtundation improved.
Patient 47 was shunted 12 months later. Only gait improved questionably.
*Initial "dementia" apparently the result of depression.
(38%). Of the eight patients whose cisternograms were classified "other," two (25%) improved, two were stable (25%), and four worsened (50%). There were 16 surgical patients who had both pneumoencephalography and cisternography consistent with idiopathic communicating hydrocephalus: six of these improved (38%), four were stable (25%), and six worsened (38%).

There were nine patients who had a prominent gait disorder and full radiological criteria (pneumoencephalography and cisternography) for idiopathic communicating hydrocephalus. In this group five improved (56%), two stabilised (22%), and two worsened (22%).

Because of the small numbers involved the only statistically significant figure in these comparisons of subsets within the surgical group was the rate of improvement in those patients who had both a prominent gait disorder and the radiological criteria for the condition as opposed to those whose clinical and radiological evaluations did not meet such criteria fully (P = 0.03). Further, it appears that patients whose gait disorder is at least as prominent as the dementia have a better chance of improving after ventriculoatrial shunting than those who do not, and that the pneumoencephalographic and cisternographic criteria for idiopathic communicating hydrocephalus, and particularly results of the two radiographic studies together, suggest a more favourable outcome.

Clinical and radiographic data for the 12 control patients are presented in Table 2. For these the mean age at initial evaluation was 70 years, the mean duration of symptoms before initial evaluation was 21 months, and the mean length of follow-up was 24 months. There were six men and six women in this group, followed for seven to 64 months.

These 12 patients were more nearly homogeneous with respect to the results of cisternography, pneumoencephalography and CAT. All presented with both dementia and gait disorder with the exception of one patient (case 49) who had only a questionable dementia at the initial assessment that later reversed with treatment for depression. Another patient with a progressive gait disorder (case 41) had only a mild dementia at initial testing that was not confirmed later.

These patients were not shunted at the initial evaluation for several reasons. Seven were not treated because the attending neurologist was reluctant to advise ventriculoatrial shunting. Four patients (or their families) refused treatment. One had other medical problems judged sufficient to contraindicate surgery. Three patients (cases 42, 43 and 47) were shunted after periods of observation of seven, nine, and 12 months respectively, and all showed mild improvement.

Table 3 gives a comparison of the age, sex distribution, duration of symptoms, and length of follow-up in these three groups of patients: surgical patients whose course could be assessed, surgical patients in whom complications rendered shunt assessment impossible, and control patients. The groups, though small, were all comparable with respect to these features.

Table 4 shows the outcomes in the 27 surgical patients whose course could be assessed and in the 12 control patients who were not treated. Both shunted and control patients could be divided into three groups—those who clearly improved, those who remained stable for at least six months or more after surgery (or after the initial evaluation), and those who continued to worsen. When the numbers of those improved were compared with a combination of those stable or worse, there was a significant difference between surgical and control patients (P < 0.01).

While the varied course after a shunt procedure has been demonstrated before, this comparison emphasises the equally varied course that patients can experience without a shunt. It should be noted that all of the controls who became progressively worse after the initial evaluation had a deteriorating gait while in only three did intellectual function change significantly. Only one of these situations was a progressive dementia.
Adult idiopathic communicating hydrocephalus with and without shunting

patients (case 43) deteriorated rapidly; the others became worse only very slowly. Six of the 12 control patients never deteriorated in the follow-up period of 12–36 months, and one actually improved.

The serious complications in the group of patients who underwent ventriculoatrial shunting are shown in Table 5. In addition to the usually recognised complications (subdural haematoma, intracerebral haemorrhage soon after placement of the shunt, shunt malfunction, and seizures), non-haemorrhagic stroke was also encountered. Seven cerebral infarctions occurred within seven weeks of surgery. Two immediately followed the procedure, and three were in the hemisphere opposite the shunt. They are included here because of the likelihood that this is an excessive incidence of infarction in a group of 37 patients over a seven week period, perhaps related to altered intracranial dynamics. Thus, there were 19 major complications in 16 patients (43% of the total group). In 13 patients (35%), the complications were serious (subdural haematomas, intracerebral haemorrhages, and non-haemorrhagic strokes), and resulted in death or permanent neurological deficit. Subdural haematomas occurred both in patients with high (3/13) and those with low or medium pressure valves (3/22). None of these complications was seen in the control patients.

It should be noted, parenthetically, that between June 1974 and March 1978 an additional 21 patients with idiopathic communicating hydrocephalus were shunted at this institution, 19 with the MPASV. While it is not possible at this time to assess the long-term results in this group, there were none of the serious complications listed above in the immediate period after surgery.

Discussion

This study suffers from problems common to retrospective investigations. Clinical data were sometimes incompletely described and sometimes not available for review. Ancillary diagnostic data were often lacking (lumbar CSF pressures and CSF infusion tests, for example). Similar flaws are common to the vast majority, if not all, of the studies of sizeable numbers of patients evaluated for possible shunting. We believe, nonetheless, that our experience is worth discussing both in relation to the data available and as a framework for the design of future prospective investigations. We recognise the difficulty of applying any statistical methods to such a small and selected population but we have offered one such approach in addition to the data.

There have been numerous clinical studies of communicating hydrocephalus in adults with dementia since the original recognition and successful treatment of this problem by Adams et al. (1965). After its introduction, radionuclide cisternography was immediately applied to the evaluation of this condition (Bannister et al., 1967), and the results of this test along with those of pneumoencephalography have been regarded as important criteria for shunting. Excellent summaries have appeared recently (Fisher, 1976; Katzman, 1977).

Table 6 contains a summary of the results of studies published between 1965 and 1977 concerning shunt therapy of patients over 40 years of age with idiopathic communicating hydrocephalus. In this tabulation, patients with previous major head trauma, subarachnoid haemorrhage, or meningeal infection have been excluded. Some series were excluded when there was not a clear statement on this point. Apart from the initial report by Adams et al. (1965) we included only those series with five or more patients.

In all, 307 patients with idiopathic communicating hydrocephalus could be identified who were treated with ventriculoatrial or, in a few cases,
with ventriculoperitoneal shunting; 144 of these patients (47%) appeared to have improved by more than a minimal amount. Given the selectivity involved, this result probably represents an exaggerated rate of favourable response in this setting. Complication rates similar to that in the present study have been noted in other recent studies (Wood et al., 1974; Laws and Mokri, 1976; Greenberg et al., 1977). The pattern of improvement varies from an apparent complete remission of symptoms to significant improvement for a few months followed by further deterioration. Only a few patients have been documented to have had a complete remission of symptoms lasting for years.

Many authors accept the proposition that certain pneumoencephalographic and cisternographic criteria can predict whether or not the patient will respond to a shunt procedure, an attitude best exemplified by Messert and Wannamaker (1974). These criteria include enlargement of the lateral and third ventricles (possibly the fourth also), dilatation of the temporal horns, and a corpus callosal angle of less than 120° (Greitz and Grepe, 1971). Criteria by radionuclide cisternography include penetration of the tracer isotope into the lateral ventricles where it remains for several days; little or no activity is observed progressing to the convexity.

Salmon (Salmon and Armitage, 1968; Salmon, 1972) was the first to cast serious doubt on the usefulness of these criteria, pointing out that in his series they did not seem reliably predictive of improvement after shunting. He said that the thickness of the brain mantle measured at the frontal horns was the most reliable indicator of success. The thinner the brain substance (less than 40 mm), the better the chance for success. This latter criterion, however, has not been found to be useful by several subsequent investigators (Stein and Langfitt, 1974; Greenberg et al., 1977). These more recent studies also cast doubt on the reliability of any criterion beyond large ventricles in predicting success.

Our own results suggest that neither the absence of atrophy shown by pneumoencephalogram, nor ventricular retention of isotope for 48 hours, nor an apparent convexity block to CSF flow, can predict the ultimate outcome accurately. Although these criteria remain compelling to many neurologists in that they relate logically to the suspected pathophysiology of idiopathic communicating hydrocephalus, it is understood that correction of CSF flow abnormalities may not necessarily alleviate abnormal neuronal metabolism or pathological anatomy, whether these changes are caused by the hydrocephalus itself or by some other process.

Some authors (Messert and Wannamaker, 1974) have held that parenchymal disease of the brain produced only hydrocephalus ex vacuo, not amenable to treatment with ventricular shunting. Others (Salmon, 1972; Greenberg et al., 1977), however, have reported that certain patients treated with a shunt may improve, even though they appear to have cerebral atrophy of various causes. Both Alzheimer's disease (Coblentz et al., 1973; Sohn et al., 1973) and cerebrovascular disease (Earnest et al., 1974; Koto et al., 1977) have been associated with communicating hydrocephalus but one expects that any improvement from ventriculoatrial shunting in these disorders would be temporary and might benefit the gait problem more than the dementia. Katzman (1977)
suggests that patients with Alzheimer’s disease and idiopathic communicating hydrocephalus should not be shunted whereas those with hypertensive cerebrovascular disease and idiopathic communicating hydrocephalus can be considered for surgery. There are not enough data on patients with proven Alzheimer’s disease or cerebrovascular disease and idiopathic communicating hydrocephalus to be dogmatic, but our experience is consistent with Katzman’s. Note that our patients, cases 9 and 24, with proven cerebrovascular disease improved with shunting whereas the patients with Alzheimer’s disease (cases 13 and 18) continued to deteriorate.

Of course, the clinical problem is the difficulty of predicting the presence of Alzheimer’s disease, cerebrovascular disease, or normal brain parenchyma in the setting of radiological criteria for idiopathic communicating hydrocephalus. Earlier and more prominent gait disturbance would favour hydrocephalus (with or without cerebrovascular disease) over Alzheimer’s disease. However, there have been reports of improvement from shunting when mental deterioration was prominent and gait little affected (Greenberg et al., 1977). One cannot dismiss entirely the possibility of benefit when dementia is the dominant clinical sign. While some have suggested that brain biopsy be used to exclude Alzheimer’s disease before shunting, we have been reluctant to advise it regularly.

A case can be made for the following position. If a patient with dementia has an early and prominent gait disorder and pneumoencephalographic (or CAT) and cisternographic criteria for idiopathic communicating hydrocephalus, there is a good chance (50% or greater) of improvement with ventriculotrial shunting. The lack of these individual criteria reduces the chances of a favourable outcome. The frequency of serious surgical complications encourages reluctance to advise shunting in the absence of supportive clinical and radiographic criteria. An occasional patient with similar clinical and radiographic findings may improve and others remain stable without shunting. Clinical judgment may well dictate a period of weeks or months of observation, especially when clinical or radiographic findings are not “typical.” Serial CAT scans or cisternography may be helpful.

We believe this problem will remain a difficult one for the clinician. Our experience indicates that the CSF infusion test gives results that can be predicted by cisternography (Trotter et al., 1974). Similarly our data on cerebral blood flow and its alteration by CSF withdrawal in demented patients (Grubb et al., 1977) do not allow us to predict accurately the results of shunting. Prolonged intracranial pressure recordings have been recommended by some for their predictive value (Chawla et al., 1974; Crockard et al., 1977; Hartmann and Alberti, 1977). Our experience with this technique in studying possible idiopathic communicating hydrocephalus is limited to one patient in this series (case 7). In addition to appropriate radiographic findings, high pressure waves were observed in this patient over two days of intraventricular recording. However, shunting improved only his gait but not his severe dementia.

It may well be too late (in view of clinical bias and risks of some of the procedures) to propose an approach to these complex issues. The ideal plan would be a prospective study of those patients with dementia and enlargement of the lateral ventricles. Careful clinical description would document details of mental and gait abnormalities. Is hydrocephalic dementia different from Alzheimer’s dementia? Simple quantitation of these clinical abnormalities and an estimate of the likelihood of multi-infarct dementia should be possible. One would like to have data on continuous lumbar and intracranial CSF pressure recordings, CSF production and absorption, and CSF infusion results. Information from brain biopsy would be useful but the risk may not be justified. Can one identify a subgroup of patients with Alzheimer’s disease or multi-infarct dementia whose course is adversely (and reversibly) affected by the complication of idiopathic communicating hydrocephalus?

Improvement in the instrumentation of shunt systems may allow investigations that will indicate which of their characteristics offer best control of intracranial pressure, hydrocephalus, and related clinical problems. Serial studies on patients after shunting might include (for ideal investigative purposes) quantitative clinical assessment, lumbar and intracranial pressure recordings, CAT and cisternographic data, documentation of shunt patency and functional characteristics. We doubt that serial observations on cerebral blood flow and cerebral metabolism would be illuminating but cannot prove it.

The dilemma of idiopathic communicating hydrocephalus remains with us. Only a rare patient benefits dramatically from present techniques. Most of these have gait disorder and dementia, but some may present in other ways. Since we do not have certain criteria for predicting the outcome of shunting and since the operation has major risks, there are still opportunities for clinical debate and much need for
careful prospective study.

While some support can be found for all eight of our original hypotheses, only the seventh has some statistical significance. This would suggest that at this time patients with presumed idiopathic communicating hydrocephalus who have an early and prominent gait disorder together with "typical" radiographic findings might be candidates for ventriculocisternal shunting. In other cases, a delay with careful clinical observation and, perhaps serial CAT scans, might be recommended before surgery is considered.

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