Letters to the editor

Poor Prognosis/Good Outcome in Patients with NPH


Abstract

Object. The authors set out to describe the outcome in a subgroup of patients with normal-pressure hydrocephalus (NPH) in whom prognostic factors were poor. This subgroup of patients who had received shunts was selected according to strict criteria.

Methods. From a cohort of 56 patients with NPH in whom shunts were placed, the authors selected a subgroup with four of the factors traditionally considered to indicate poor prognosis: idiopathic type, cortical atrophy, longstanding symptoms, and presence of dementia in addition to old age. Twelve patients met the inclusion criteria.

Results. After receiving shunts, 92% of the patients showed clinical improvement on the NPH scale: gait improved in 100% of patients, sphincter control in 90%, and dementia in 33%. Improvement was significant for gait and sphincter control, general NPH score, and most daily life activity scales. No significant differences regarding clinical, cognitive, or functional changes following surgery were found in comparison with the rest of patients (the good prognosis subgroup).

Conclusions. The clinical condition of patients with NPH who present with traditionally accepted markers of poor prognosis can improve after surgery, especially as regards gait and sphincter control. The authors assert that the presence of these markers should not be considered to be an absolute criterion for ruling out shunt surgery in cases of NPH syndrome.

This letter concerns the authors’ second factor indicative of poor prognosis, cortical atrophy and its delineation. The computerized tomography (CT) scans presented in Fig. 3 of their paper were obtained in a patient who was presumed to have dilated cortical sulci (taken to indicate cortical atrophy) and who, against the odds, responded favorably to a shunt placement. Yet these scans demonstrate widening of the sylvian fissures and the cerebral sulci at the midlevel of the cerebral convexities, in addition to that of the periventricular lucencies. No uppermost CT scan cuts are shown. In my study, I described the presence of obliteration of the cerebral sulci in the uppermost CT scan cuts and dilation of the subarachnoid spaces at lower levels, including the sylvian fissures, in patients with NPH. The finding was considered to indicate obstruction of the subarachnoid spaces of the convexity of the hemispheres with subsequent dilation of all the spaces below the level of obstruction. In Fig. 3 of my paper I presented CT scans obtained in a patient with NPH that showed dilation of the sylvian fissures in addition to marked ventricular enlargement. He responded well to treatment, and both the ventricles and the sylvian fissures diminished in size postoperatively. This pattern was also documented in the two patients described by Salibi, et al., and is also shown in the postoperative scans of the patient presented in Fig. 3 by Poca and colleagues. Figure 2 of their paper refers to a patient with NPH. The legend reads “despite enlarged sylvian fissures and cortical sulci, the patient demonstrated intracranial hypertension on the ICP [intracranial pressure] tracing, with 51% of high-amplitude B waves.” I presume that the authors use this case as an example of a patient with NPH and cortical atrophy who despite the latter has intracranial hypertension. If this assumption is correct, then I believe that their statement is misguided and misleading. Unless the authors can demonstrate widening of the cerebral sulci in the uppermost CT scan cuts, the patient should not be considered to have cortical atrophy. Widening of the subarachnoid spaces at the midlevel of the hemispheres and that of the sylvian fissures most probably follows obstruction of the subarachnoid spaces high in the convexity of the hemispheres. The pathological substrate of this obstruction is unknown, but possibly it is attributable to compression and obliteration of the spaces by the expanding ventricles.

This feature should not be considered a bad prognostic criterion in patients with the syndrome. It certainly should not be confused with cortical atrophy.

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References


Response: We thank Dr. Vassilouthis for his letter. We are aware of his studies in the field of NPH and appreciate his interest in and comments on our work. His letter provides us with the opportunity to broaden discussion of the still-controversial topic of the real significance of increased cortical sulci size in patients with NPH and to explain the actual ICP values that these patients may have in further detail.

Dr. Vassilouthis’ comments focus on the patient whose CT scan is shown in Fig. 3 of our article. As Dr. Vassilouthis correctly points out, this scan shows a widening of the sylvian fissures and the cerebral sulci at the midlevel of the cerebral convexities in addition to that of periventricular lucencies. For the sake of space and clarity, only two CT scan slices were included; the figure was chosen not to discuss whether or not the cortical sulci were dilated but to show how a patient with moderate ventricular dilation only and widened cortical sulci can have NPH and improve after shunt insertion. The uppermost slices are shown in Fig. 1 of this letter. We believe that after seeing this figure, Dr. Vassilouthis will agree that the cortical sulci are not obliterated in them and therefore that our point is clinically relevant and not “misleading and misleading.”

In all patients with suspected NPH, we analyze the sulci size at different levels, as well as sylvian fissure size. In our study, we used cortical sulci size as inclusion criteria, not
sylvian fissure size; however, in all 12 patients, the size of all these structures increased simultaneously. We believe that this is evident in Fig. 2 of our article, which shows extreme enlargement of some cortical sulci. Cortical sulci enlargement are also evident in one of the slices of the presurgical CT scan shown in Fig. 3 (upper right) of our article, and is even more obvious in Fig. 1 of this letter, which shows the patient’s complete set of magnetic resonance imaging slices. Both figures provide evidence that the patients included in our study had cortical atrophy.

Figure 2 of our paper was chosen to emphasize that patients with what is still known as NPH can have intracranial hypertension even when cortical atrophy is evident. This phenomenon has been mentioned by several authors and we described it extensively in 1991 in a paper published in Acta Neurochirurgica (Wien) in which the following comment was added by the editor, “The authors decided—against suggestions of our reviewers—to include patients into their study who had higher than normal ICP values and therefore cannot be considered to suffer from NPH. The argument of the authors has been, that they presented only patients with typical clinical syndromes. With some reluctance we finally have accepted the paper as it stands. But we still are convinced that it would have been better to include only patients into this interesting study who really belong to the Normal Pressure Hydrocephalus group.”

After this paper was published, and after the increased use of continuous ICP monitoring in this syndrome, this phenomenon was also reported independently by other authors. Evidence is accumulating that in some patients with NPH syndrome, mean ICP is not always normal, even when these patients have no symptoms of intracranial hypertension but show the classical symptoms of the NPH syndrome. In some of these patients, continuous ICP monitoring shows either isolated episodes of intracranial hypertension (usually during rapid eye movement sleep) or continuously elevated mean ICP. Although the term “normal-pressure hydrocephalus” as first described by Hakim continues to be used, the alternative term “adult chronic hydrocephalus” (which does not indicate ICP value or any physiopathological theory) is being increasingly adopted and better defines this complex and frequently encountered syndrome.

The association of marked sylvian fissure enlargement and obliterated cortical sulci in some patients is well known. Data from recent studies have revealed that enlarged sylvian fissures and basal cisterns and some focally dilated sulci are compatible with a favorable response to shunt insertion in patients with NPH. The role of sylvian fissures as reservoirs of cerebrospinal fluid (CSF) analogous to those of the ventricular system has been repeatedly reported, and these patients’ condition has even been incorrectly diagnosed as

Fig. 1. Complete images corresponding to the presurgical T2-weighted magnetic resonance images shown in Fig. 3 of our original paper. The figure shows enlargement of all cortical sulci, including those in the uppermost slices.