Is normal pressure hydrocephalus a valid concept in 2002? A reappraisal in five questions and proposal for a new designation of the syndrome as “chronic hydrocephalus”

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The authors question the current validity of the traditional concept of normal pressure hydrocephalus (NPH) as it was described by Adams and Hakim in 1965. The classic features of the disease are addressed. It is concluded that most of the historical statements made three decades ago need to be revised. Especially, the term “normal pressure” hydrocephalus probably does not match the actual manometric profile of patients with NPH. Similarly, the terms “curable” and “reversible” dementia are inadequate to designate the mental alterations of NPH. That NPH is non-specific to the adult population is also stressed, since it may be not uncommonly encountered in paediatrics, especially in an implanted shunt malfunction. The term “chronic hydrocephalus” without reference to cerebrospinal fluid pressure and to the age of the patient is proposed instead of NPH, which seems out of step with current knowledge of the pressure profile and with the diagnosis and decision making context in patients with so called NPH.

Normal pressure hydrocephalus (NPH) was described in 1965 by Hakim and Adams as a new syndrome in three patients showing a characteristic clinical triad of altered mentation, gait difficulties, and sphincter disturbances together with ventricular dilatation in air studies and normal pressure of the cerebrospinal fluid (CSF) at lumbar puncture. This apparently paradoxical condition has prompted a considerable number of studies aimed at explaining the pathophysiological background of the disease and at identifying the best candidates for CSF shunting surgery.

At the beginning of the 21st century, many controversies remain about the nosographic concept of the disease, its pathophysiology, diagnostic tests, and the most reliable predictors of a good response after shunting. In 1990, we published a comprehensive report on this topic and we proposed that the new designation “chronic hydrocephalus” be used instead of the NPH, which seemed inadequate. In 2002, it is interesting to question the current validity of Adams and Hakim’s historical concept by addressing the five following issues:

1. Was NPH actually a newly discovered disease when reported in 1965 by Adams and Hakim?
2. Is CSF pressure really normal in patients with NPH?
3. Is NPH really a cause of curable dementia?
4. Is NPH specific to the adult and elderly population?
5. Is CSF shunt the only treatment modality?

1. WAS NPH ACTUALLY A NEWLY DISCOVERED CONDITION WHEN REPORTED BY ADAMS AND HAKIM IN 1965?

The answer to this somewhat provocative question is negative. Several papers published before 1965 clearly addressed this problem in patients showing the apparent paradox of progressive symptomatic hydrocephalus without overt signs of intracranial hypertension. Especially, a pioneering paper by Riddoch provided as early as 1936 a description of the syndrome that was closely similar to that proposed by Adams and Hakim three decades later: “The clinical picture was one of slowly progressive dementia. All this indicates that the main explanation of the absence of headache, vomiting and papilledema lies in the very gradual obstruction to the CSF circulation. The absence of increased pressure in the ventricles and on lumbar puncture supports this view.” Strikingly similar descriptions are also available in other early contributions, such as those by McHugh and by Messert and Baker, with a special mention of relief of symptoms after shunting in the latter.

2. IS CSF PRESSURE REALLY NORMAL IN PATIENTS WITH NPH?

This question is of utmost importance because it addresses the pathophysiological background of the syndrome. It should be remembered that the historical labelling “normal pressure” hydrocephalus was based on the finding that all three reported patients showed low CSF pressures at lumbar puncture. It is now widely recognised that a single...
limited in time, CSF pressure measurement by lumbar puncture yields a poor estimation of the real intracranial pressure (ICP) profile of patients with NPH.

Moreover, there is an abundant literature based on ICP monitoring that acknowledges that CSF pressure may actually not be normal in this population. In this view, the presence of numerous B waves on prolonged CSF pressure recording indicates a general trend to episodic high ICP levels, which was interpreted by some authors as a favourable predictor of response after shunting. Although the predictive value of CSF infusion tests remains debatable, measurements of resistance to outflow and of the pressure-volume index showed that, in the case of a sudden increase of ICP, compensatory mechanisms were altered and easily overwhelmed in patients with NPH. Additionally, there is clinical evidence from daily practice that makes questionable the current use of the term NPH: only a few specialised institutions have gained expertise in routine CSF infusion tests. In most neurosurgical centres, the decision to use shunting in such patients is undertaken on the basis of their clinical and computed tomography (CT) or magnetic resonance imaging (MRI) presentation without assessment of their CSF pressure.

The importance of the clinical and CT parameters in decision making has been clearly confirmed in the above mentioned report based on 243 cases and in other recent contributions. Specifically, it has been shown that good results after shunting can reasonably be expected in patients showing a ventricular dilatation with a frontal horn ratio exceeding 0.50 on CT studies along with one or more of the following criteria:

- Presence of a clearly identified etiology;
- Predominant gait difficulties with mild or absent cognitive impairment;
- Substantial improvement after CSF withdrawal (CSF tap test);
- Normal sized or occluded sylvian fissures and cortical sulci on CT or MRI;
- Absent or moderate white matter lesions on MRI.

The presence at the first evaluation of such favourable parameters justifies by itself the decision for shunting and makes the use of ancillary tests unnecessary, including the CSF dynamic tests, which are invasive and rely to a large extent on the operator's expertise. In patients lacking favourable criteria, a 3–5 day external lumbar drainage has been reported to be a reliable predictor of outcome after shunting. The predictive value of CSF hydrodynamics assessed by T2 weighted images and cine phase contrast MRI in patients with NPH is being evaluated.

Difficulties in the nosographic designation of the syndrome are acknowledged by the number of alternative terms that have been proposed, which cannot be listed here. Since the early recognition of the syndrome, it has been suggested that the term NPH might not be the most appropriate for the condition. Because the traditional terminology matches neither the usual conditions of decision making nor the real ICP profile of patients with so called NPH, we proposed in 1990 to designate this syndrome “chronic hydrocephalus”.  

3. IS NPH REALLY A CAUSE OF CURABLE DEMENTIA?

Unfortunate terminologies such as “hydrocephalic dementia” or “curable dementia” have been used extensively to designate the mental disorders of NPH. Such confusing terms have generated the widespread myth that some forms of “true” dementia can be cured by shunt surgery, this view being apparently supported by some biopsy or necropsy findings showing that degenerative or ischaemic brain lesions may be associated with ventricular dilatation. This erroneous belief abusively led to extensive application of shunt operations in demented patients, with uniformly devastating results; this may be an explanation for the disappointment among the neurosurgical community during the first few years that followed Adams and Hakim's publication. Still, nowadays, neurosurgeons are occasionally requested to use shunting in such patients based on the argument that “they have nothing to lose”.

Although a specific psychometric profile is difficult to define in NPH, psychometric studies have widely documented the fact that patients with NPH did not fit the criteria of degenerative (Alzheimer-type) or arteriosclerotic dementia. Although psychometric tests may not differentiate NPH from other types of subcortical mental alterations, they have proved helpful in distinguishing subcortical from cortical cognitive deficits. Patients with NPH exhibit subcortical-type mental deficits including forgetfulness, decreased attention, inertia, and mental slowness with a pattern of memory impairment that differs from that of Alzheimer's disease and other cortical encephalopathies. The differential diagnosis with Alzheimer-type dementia is further supported by the fact that patients with NPH do not exhibit the “aphasia-apraxia-agnosia syndrome”, which belongs to the picture of cortical dementia.

True dementia, as defined in the Diagnostic and statistical manual of mental disorders, third edition, revised, does not belong to the clinical triad and “demented patient” should definitely be excluded from clinical descriptions of NPH. When massive intellectual loss is present in the clinical presentation, other diagnoses should be considered and shunt surgery should be refused even in cases where CT studies illustrate some degree of ventricular dilatation, which is also a common finding in demented patients. In such cases, the presence of significant hippocampal atrophy on MRI studies has been reported to be an important neuroimaging criterion for the diagnosis of Alzheimer's disease.

4. IS NPH SPECIFIC TO THE ADULT AND ELDERLY POPULATION?

In other words, is there a paediatric counterpart to adult NPH? This question raises the problem of the normal ICP level in children, which remains a debatable matter. It is generally agreed that there is a relation between ICP and the age of children, with ICP values progressively increasing from subatmospheric levels in the neonate to a range of 20–70 mm H2O during infancy and ultimately to its adult levels by the age of eight years.

Several studies reported in the literature have shown that a condition comparable with NPH may not be uncommon in childhood. From a clinical standpoint, a paediatric form of the triad may be characterised as follows:

- Subtle mental deterioration manifested by poor or decreasing scholar performances;
- Anomalies of gait manifested either by walking retardation in infants or by repeated falling spells in older children;
- Anomalies of micturition with delayed bladder control, whose relation to hydrocephalus is frequently overlooked or ascribed to enuresis. Urodynamic evaluation of bladder dysfunction in hydrocephalic children has shown that a mechanism grossly similar to that found in adult patients was involved, namely a failure of the supraspinal detrusor inhibitory centres.

Paediatricians should be aware of the possible occurrence of slowly progressive symptomatic hydrocephalus in children. Especially, patients with shunt malfunction may show subtle deterioration of their psychointellectual performances without overt signs of intracranial hypertension. Such a condition...
may be misdiagnosed as a “shunt independent arrested hydrocephalus”, overlooking the fact that hydrocephalus is still active and that shunt revision may be beneficial.  

5. IS CSF DIVERSION THE ONLY TREATMENT MODALITY?

Despite several attempts to introduce non-shunt methods of treatment, alternatives to shunting remain marginal or even speculative. In most instances of suspected NPH, the crucial problem is whether to shunt or not to shunt. Ventriculoperitoneal shunts have become standard treatment although ventriculotrial shunts are still a second line option in patients with a history of multiple laporotomies or with defective peritoneal CSF resorption. Although proposed as an attractive and safer modality, a lumboperitoneal shunt has never gained widespread use in patients with NPH, probably because its application is strictly limited to proven communicating forms of hydrocephalus and because of the high rate of tonsillar herniation reported in patients with such a shunt implanted.  

Whatever the type of shunt, the major drawback of all shunting methods is the high rate of shunt related complications resulting in the highest morbidity in neurosurgical practice. Recent improvements in shunt technologies are aimed at reducing the incidence of technical failures and of siphon effect related complications of shunts. Technical refinements include antisiphon devices, flow controlled valves, and valves with adjustable pressure. The cost effectiveness of such devices compared with conventional hydrostatic valves is still under debate. 

Besides the vast majority of patients with NPH, in whom shunt surgery is the only possible treatment, in a limited number of cases other modalities may be considered. Curing a causative factor is seldom possible in secondary NPH but should be proposed in the few cases where CSF flow is compromised by intracranial tumours or malformations, without previous shunt implantation. Medical treatment of NPH includes acetazolamide and repeated lumbar punctures, which have occasionally yielded prolonged clinical improvement. Although justified in patients with high surgical risks, conservative management usually results in mild and transient relief of symptoms and shunt surgery is ultimately required. Except for rare cases of aqueductal stenosis related NPH, there has been little enthusiasm for third ventriculostomy in patients with NPH, mainly because hydrocephalus has been regarded in most instances as resulting from a communicating CSF circulation disorder. A recent contribution suggests that third ventriculostomy may be an attractive alternative to shunting, even in cases where the aqueduct is patent on imaging studies, in reducing the tissue stress to periventricular areas by reducing the transaqueductal pressure gradient.  

However, third ventriculostomy cannot be recommended as a first line treatment and its efficiency in patients with NPH requires further validation.  

CONCLUSION

Some of the statements made by Adams and Hakim 35 years ago remain valid today. This is true for the clinical presentation since a majority of patients at diagnosis fit the criteria of the triad, with the notable reservation that the term “true” dementia is not attributable to its psychiatric component. It is also true that some carefully selected patients respond to shunting, which remains the standard today. However, an unacceptably high rate of shunt related complications justifies all attempts at promoting alternative methods of treatment. On the other hand, it has become obvious that the other elements of the historical concept of NPH need to be revised. NPH should not be regarded as an age related disease specific to the adult and elderly population. A similar condition may be encountered during childhood, with a clinical presentation that does not differ basically from that of the adult and may be erroneously ascribed to “arrested hydrocephalus”, which is a distinct condition. More important, the term NPH is questionable because it matches neither the real conditions of the current diagnosis—which is established in most institutions on the basis of the clinical and CT presentation only, without assessment of the ICP—nor the actual CSF manometric profile of such patients. This is acknowledged by the results of dynamic tests that showed a general trend to increased, albeit compensated, pressure levels or at least an inability of compensatory mechanisms to dampen a sudden increase of ICP in patients with NPH.  

For these several reasons, we propose a more relevant nosographic designation for the NPH syndrome by renaming it “chronic hydrocephalus” without reference to age and CSF pressure. Chronic hydrocephalus is an active process that should be replaced in the sequence of events that result from failure of the circulatory resorptive mechanisms of the CSF. It may be regarded as an intermediate state of balance between uncompensated hypertensive hydrocephalus and asymptomatic hydrocephalus (in which compensatory systems are fully effective). As proved by follow up monitoring of shunting in patients, insertion of a shunt in those suffering from chronic hydrocephalus often provides a clinical cure without changes in their ventricular size. Shunting may therefore be regarded as an additional compensatory system allowing chronic hydrocephalus to turn in to asymptomatic hydrocephalus, which may be the only condition that really deserves the label NPH.

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References


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