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Chronic ("normal pressure") hydrocephalus in childhood and adolescence

A review of 16 cases and reappraisal of the syndrome

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Abstract "Normal pressure" hydrocephalus (NPH) is generally considered to be a disorder of the adult and geriatric population. Only a few reports have described the possible occurrence of this condition in children. A series of 16 patients aged less than 20 years forms the basis of the present report. Among these 16 patients, 11 had a clearly identified etiologic factor and 7 had had a shunt previously implanted. The majority of patients exhibited at least two elements of the adult's triad of psychomotor retardation (14 cases) and/or psychotic-like symptoms (4 cases), gait anomalies (8 cases), and sphincter disturbances (3 cases). Six patients had their intracranial pressure (ICP) monitored. ICP values were estimated to be within the normal limits for age. All the 16 patients underwent shunting or shunt revision. Surgical results were as follows (mean follow-up 20±17.2 months): a good response to shunting was obtained in 12 cases ("cured": 5, improved: 7), while the other 4 patients failed to improve. It seems likely that associated parenchymal disorders have played a major role in therapeutic failures. In children showing ventricular dilation on computed tomographic (CT) analysis and a clinical picture of subtle psychomotor deterioration, it may be difficult to distinguish an active disorder of the CSF dynamics from "arrested hydrocephalus." Since intracranial manometry cannot be undertaken as a routine procedure, less invasive methods such as cerebrospinal fluid (CSF) tap test, psychometric, or urodynamic tests deserve special attention as reliable predictors of outcome after shunting. Because most patients undergo shunting without prior assessment of their CSF pressure, the term "chronic hydrocephalus" is proposed as an alternative designation to "NPH," since there is little argument for maintaining an instrumentally based definition of the syndrome.

Key words Hydrocephalus · Adult · Pediatrics · Cerebrospinal fluid · Intracranial pressure · Psychometry · Shunt · Shunt obstruction

Introduction

In a recent report on 243 adult patients with a so-called normal pressure hydrocephalus (NPH), it was proposed by the authors of the present paper to substitute for this widely adopted but questionable term "normal pressure hydrocephalus" the more realistic designation "chronic hydrocephalus of the adult," because today the great majority of patients undergo a shunting procedure without prior assessment of their intracranial pressure (ICP) [6]. Since its first description by Adams and Hakim in 1965 [1], this syndrome has generally been regarded as specific to the adult and geriatric population. Although there is no reason why

Table 1 Summary of pediatric cases in which shunting (or shunt revision) was carried out for "normal pressure hydrocephalus" (*AS* aqueduct stenosis, *gait* anomalies of gait, *IV hemorrhage* intraventricular hemorrhage, *PMR* psychomotor retardation, *sphincter* sphincter disturbances, *ST glioma* supratentorial glioma)

Patient no.	Age (years)	Sex	Etiology	Previous shunt, duration (months)	Signs and symptoms	Result after shunt follow-up (months)
1	4	F	AS	Yes, 50	PMR, gait, macrocephaly	Cured, 15
2	4	M	?	Yes, 30	PMR	Improved, 6
3	7	M	AS	No	PMR, seizures	Cured, 55
4	7	F	AS	No	PMR, gait and sphincter, macrocephaly	Improved,18
5	10	M	Multiple abcesses	Yes, 10	PMR, macrocephaly	Unimproved, 14
6	11	F	?	No	PMR, gait	Improved, 43
7	12	M	Neonatal IV hemorrhage	Yes, 40	PMR, macrocephaly	Unimproved, 18
8	14	F	Meningo-myelocele	No	PMR, macrocephaly	Unimproved, 60
9	15	M	Neonatal IV hemorrhage	No	PMR, "psychotic-like," macrocephaly	Unimproved, 8
10	15	M	ST glioma radiotherapy	No	PMR	Improved, 9
11	15	F	?	No	"Psychotic-like," gait	Cured, 6
12	16	F	ST glioma radiotherapy	Yes, 145	PMR, "psychotic like," sphincter	Improved, 9
13	16	F	AS	No	PMR, gait	Improved, 16
14	17	F	?	No	PMR, gait, macrocephaly	Improved, 24
15	19	F	Meningio-matosis	Yes, 20	"Psychotic-like," gait and sphincter	Cured, 18
16	19	F	?	Yes, 210	PMR, gait	Cured, 5

it should not occur at any age, few papers have dealt with the pediatric form of "NPH." In the present study, a series of 16 consecutive patients aged less than 20 years is analyzed.

Clinical material and results

The cases of 16 pediatric patients who underwent shunt placement for "NPH" during the past 5 years (1990–1994) were retrospectively reviewed. Each patient's clinical history and computed tomographic (CT) data fulfilled the following criteria:

- Existence of long-standing impairment of the neuro- or psychological status with subtle deterioration of intellectual performance and slowing down of psychomotor development
- Absence of overt signs of increased ICP
- Presence of seemingly stabilized ventricular dilation on serial CT documentation
- Suspicion of an active disorder of the cerebrospinal fluid (CSF) dynamics, prompting shunting or revision of a previously placed shunt

Etiological and clinical data

Our 16 cases are summarized in Table 1. The age of patients at shunting (or shunt revision) ranged from 4 to 19 years (mean 12.5± 5 years). In the 5 patients over 15 years of age, the onset of symptoms had taken place in the pediatric period. In 11 patients a causative factor was present (intracerebral hemorrhage at birth: 2 patients; intracranial supratentorial neoplasms at a late stage: 3 patients (with radiotherapy delivered in 2); meningomyelocele: 1 patient; aqueduct stenosis: 4 patients; bacterial meningitis with multiple brain abcesses: 1 patient). In the remaining 5 patients, no etiologic factor was identified. Seven children had had a previous shunt implanted for an average duration of 72 months (±75.5 months). The other 9 patients were "shunt-free" when their symptoms were ascribed to active hydrocephalus and shunting was considered.

Poor intellectual performance and/or psychomotor retardation were present in all patients but two. A "psychotic-like" presentation, unresponsive to chemotherapy, was noted in four patients. Anomalies of gait comparable to those reported in the corresponding adult population were present in eight patients and recent sphincter disorders in three. Macrocephaly was present in seven patients.

CT data

Ventricular size was graded on axial CT images using the bifrontal index (BFi), which was obtained by dividing the maximum ventric-

ular span at the level of the frontal horns by the width of the intracranial content at the same level (normal BFi <0.30). The mean BFi value at time of diagnosis was 0.63 (±0.18). Except in two patients who had shown a recent increase in their BFi values, ventricular size had remained stable through several months or years, during which "arrested hydrocephalus" had been diagnosed.

Instrumental data

Further procedures aiming at identifying an active CSF disorder were undertaken in 10 of the 16 patients of the series:

- Intracranial manometry and related procedures [mean ICP, presence of A and B waves, CSF infusion test with measurement of the resistance to outflow (*Ro*)]: six patients. All six patients were estimated to have normal mean ICP levels for their age. A and B waves were present in two together with *Ro* values above 12 mmHg/ml min.
- CSF tap test by lumbar puncture: two patients.
- Urodynamic evaluation of bladder function: one patient.
- Psychometric tests (using Mini Mental Test and the Wechsler Child Intelligence Scale): five patients.

In the other six patients, the decision for shunting (or shunt revision) was undertaken on the basis of clinical and CT data only, without further investigative procedures.

Operative results

All 16 patients of the series were operated on. In the seven previously shunted patients, a mechanical cause of shunt malfunction was disclosed and corrected at surgery. In the other nine patients, a primary shunt was placed (ventriculoatrial in two, ventriculoperitoneal in seven). There was no operative morbidity or mortality. Results were as follows (mean follow-up: 20±17.2 months): four patients remained unchanged after surgery; seven responded partially to operation with improved academic performance and better responsiveness and awareness; five were regarded as cured relative to their condition at presentation.

Discussion

Like adult hydrocephalus, it now seems certain that in many cases infantile hydrocephalus may present as a chronic disorder, either primarily or in patients with a shunt that is malfunctioning. It has been suggested that progressive "NPH" may be more common among children than adults if it is looked for [16]. To date, however, the pediatric form of the syndrome has received less attention than its adult counterpart [5, 10, 14, 17, 18, 24, 26, 27]. There are at least three explanations for this: (1) Like adults, the majority of hydrocephalic children undergo a shunting operation on the basis of their clinical and CT profile only, without prior assessment of their CSF pressure. (2) Even nowadays, what actually constitutes "normal" CSF pressure in childhood remains under debate. Most authors agree in that there is a relationship between mean ICP and the children's age [9, 23, 28]. ICP has been measured at subatmospheric levels in the neonate, increasing to a 20–70-mmH_2O range during infancy, and ultimately reaching its adult level by the age of 8. (3) It is likely that a proportion of children who are regarded as having "arrested hydrocephalus," on the argument that they show neither overt signs of increased ICP nor any change in ventricular size on repeat CT, actually have an active, although slowly progressive, disorder of their CSF dynamics.

From the clinical standpoint, most of these children show deterioration of a longstanding neurological deficit which had seemed stable or subtle worsening of a previously known psychomotor retardation. These may be difficult for both families and physicians to distinguish from the patients' baseline functions. Deterioration of the intellectual abilities may not be obvious to the parents, even if teachers sometimes refer to it because of failure at school. "Psychotic-like" disorders were also present in four patients, in two of whom they were a presenting symptom. All these four patients were over 15 years of age. Their parents had noticed changes in their behavior and personality. Despite the fact that they had no history of psychiatric disease and their CT scans showed marked ventricular dilation, their condition was assumed to be purely psychotic and they were given chemotherapy, which proved unsuccessful. They all improved after a shunt was at last inserted. This kind of "psychotic" presentation has been also described in adult "NPH" patients, including the reversibility of the behavioral symptoms by CSF shunt [7, 8, 22].

It is worth noting that the other elements of the Hakim and Adams triad may also be present in this pediatric population. Eight patients in our series exhibited gait anomalies of various types, either as long-standing retardation of walking which had recently worsened or in a pattern similar to the adult form with unsteady gait and short steps with frequent falls. Anomalies of micturition in hydrocephalic children may possibly have been overlooked or ascribed to enuresis. Three of our patients had bladder incontinence with no previous history of sphincter disturbances. In one, urodynamic assessment showed that his bladder capacity was initially less than 100 ml, and it increased to over 200 ml after 30 ml CSF was removed via a lumbar puncture. This tends to prove that in some hydrocephalic children, loss of supraspinal control of detrusor inhibition may be involved, as in the adult [2, 6].

As mentioned in previous studies [10, 12], shunted patients with shunt malfunction may not uncommonly show subtle deterioration of psychointellectual performance without overt signs of intracranial hypertension, as occurred in the seven previously shunted patients in our series. It should be emphasized that in a significant proportion of shunted children, progressive worsening of intellectual performance or lack of improvement after shunting together with persistent ventricular dilation on CT may be misdiagnosed as "shunt-independent arrested hydrocephalus," the fact being overlooked that the hydrocephalus is still active and shunt revision may be of benefit.

ICP recordings and CSF infusion tests have yielded information as to the manometric profile of pediatric "NPH" patients that is grossly similar to that available for the corresponding adult population [10–12, 25]. Patients showing A and B waves on a "normal" ICP baseline are likely to improve after shunt placement. *Ro* levels above 12 mmHg/ml min also seem to have a favorable predictive significance, as in the adult [6, 19, 20]. Although helpful in identifying patients who are likely to improve after shunt insertion (or revision), hydrodynamic tests cannot be recommended as a routine procedure in all institutions, since they remain invasive and their clinical reliability depends to a large extent upon the investigators' experience.

In a few cases, a definite decision to shunt cannot be taken on the basis of the patient's clinical and CT data. In these circumstances, it is suggested that minimally invasive procedures be employed in these young candidates for shunting. The CSF tap test [6, 29] and urodynamic tests [2, 6] deserve special attention here because they have proved reliable predictors of the outcome after shunting.

Psychometric tests are also a valuable tool as they can reveal progressive deterioration of intellectual performance in patients with apparently stable hydrocephalus, thus prompting a decision to shunt [15, 16, 27]. Although there are differences in results among the various tests employed; it is generally agreed that there is a discrepancy between the performance and the verbal intellectual quotient of these patients [14]. Alterations in fine psychomotor functions are responsible for the clumsiness of these children and are most likely to improve after shunting [27].

Surgical results are consistent with those obtained elsewhere in the literature. Twelve of the 16 patients in the present series improved after placement or adequate revision of a CSF shunt, while 4 did not improve. As shown by both their clinical history and their CT images, it was clear that three of these four "failed" patients suffered from associated parenchymal disorders which played a major role in their poor outcome. Two of them had had an intracerebral hemorrhage in the neonatal period and the third had suffered multiple brain abcesses from bacterial meningitis. Failures to improve after shunting in pediatric patients with "NPH" have to be seen as parallel to the adult cases in which failure of shunt treatment was related to additional parenchymal disorders, either degenerative or ischemic in origin [3, 4, 6, 21]. These disorders have been widely recognized as predictors of a poor prognosis, possibly because they promote increased ventricular compliance as a consequence of alterations in the viscoelastic properties of the parenchyma [5, 12, 13]. Whatever the age of the patients, any noninvasive method designed to identify such parenchymal lesions would appear a major advance in the correct selection of patients to be shunted.

Conclusion

In children showing progressive deterioration of their neurological and/or psychological status together with ventricular dilation on CT, an active process of hydrocephalus should be considered. Although difficult to differentiate in some instances from "arrested hydrocephalus," this condition must be clinically identified since a shunting procedure may be expected to benefit the majority of patients, at least the same proportion as among adults.

Manometric studies have confirmed that the majority of these patients have low or "normal" CSF pressure in apparent contrast to their CT images, which show ventricular dilation. Despite this fact, it seems worth taking this opportunity to question the continued use of the term "normal pressure hydrocephalus" in this population of patients, in whom ICP is rarely assessed in current clinical practice. Since the decision to shunt is most commonly taken on the basis of clinical and CT data only, we propose substituting the more realistic label "chronic hydrocephalus" for all patients of this group, regardless of their age, instead, since there is little argument today for defending an instrumentally based definition of the syndrome.

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