IDIOPATHIC NORMAL PRESSURE HYDROCEPHALUS' OUTCOME AFTER CEREBROSPINAL FLUID SHUNTING

Leonidas Grigorakos[1], Lavrentios Roussos[2], Pavlos Myrianthefs[3], Ioannis Alamanos[4], Christos Kelesis[5], Chryssoula Lemonidou[6], Nikolaos Sakelaridis[7]

Abstract: **The aim of this study was to evaluate the clinical outcome of patients diagnosed with Idiopathic Normal Pressure Hydrocephalus (INPH) after cerebrospinal fluid (CSF) shunting. Thirty patients diagnosed with INPH were treated with CSF shunting. The patients were evaluated preoperatively and 6 months postoperatively, in terms of their clinical outcome of gait, cognitive function and urinary incontinence. Sixteen patients (53%) showed an average improvement of their clinical symptoms and 6 months after shunting were able to function independently. Ten patients (34%) were able to return to their every day functioning. In four patients (16%) there was no clinical improvement. Our data suggest that patients diagnosed with INPH and subjected to CSF shunting had a significant clinical improvement in the 6-month postoperative follow-up. Positive outcome on patients with INPH after CSF shunting is highly correlated with immediate and accurate diagnosis based on the presence of the classic clinical "Adam-triad", preoperative cerebrospinal fluid pressure monitoring and drainage response.**

Keywords: Cerebrospinal fluid shunting, Idiopathic Normal Pressure Hydrocephalus, Outcome

INTRODUCTION

Idiopathic Normal Pressure Hydrocephalus (INPH), primarily a condition of the elderly, is defined as a hypokinetic-rigid gait disorder with incontinence and cognitive decline-constituting the classical "Adam-triad" - and normal cerebrospinal fluid (CSF) pressure on lumbar puncture [1]. It is considered to be a disorder of CSF circulation, as analysis of CSF pressure recordings and infusion studies demonstrate an increased resistance to CSF absorption and an increased frequency of pathological vasogenic waves [2]. Radiological features include dilated ventricles and the presence of periventicular white matter lesions, without extensive atrophy [3]. The mainstay of therapy for INPH is CSF diversion via a ventriculoperitoneal or ventriculoatrial shunt. The success rate of shunt surgery and reported short-term and long-term effects of shunting vary considerably between studies and range from no statistically significant improvement to improvement rates of 70% [4, 5].

INPH remains a diagnostic and therapeutic dilemma for the clinician, since patients very often present with dementia and there is a great difficulty in distinguishing them from other neurodegenerative conditions. Its importance as one of the most common reversible causes of dementia, by means of CSF shunting, justifies the efforts to unravel this complex problem.

In this prospective study we present our experience gained from patients with INPH, evaluating their clinical performance pre and postoperatively.

STUDY MATERIAL AND METHODS

We describe thirty patients diagnosed with INPH and operated in the Department of Neurosurgery at Trauma Hospital (KAT, Athens) during the last 6 years. After operation patients were closely monitored for two days in the high dependency unit (HDU). All patients were preoperatively examined and re-evaluated at 6 months postoperatively. Diagnostic criteria for INPH included the presence of typical gait disturbance, organic mental symptoms and urinary incontinence and enlarged ventricles on computed tomography (CT). The diagnosis also required an insidious onset and no evidence of antecedent causes of secondary hydrocephalus were revealed. Eight patients were admitted to the hospital for continuous CSF pressure monitoring and in twelve patients we performed controlled diagnostic CSF drainage. Twenty-eight patients subsequently received ventriculoperitoneal shunt using medium or high-pressure valves; one patient was submitted to ventriculoatrial and one to lumboperitoneal shunt. The postoperative follow-up consisted of clinical evaluation. Cognitive function improvement was evaluated from either the patient's or family's prospective. Urinary function improvement was defined as a decrease in incidence of urinary frequency, urgency or incontinence. Gait improvement was documented by change in detailed clinical evaluation and documentation of dependence on assistive devices. The overall day to day functioning of the patient was also evaluated. Symptoms were classified as "good improvement" if the patient could return to his every day functioning and was able to work, "average improvement" if the patient was able to function independently at home but still had some neurological deficit and "bad outcome" if the patient and the family members could not detect any change in the overall functioning in the following six months. In the group of "bad outcome" we also included those patients who had initial postoperative improvement in any of their symptoms but deteriorated later.

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RESULTS

Demographics and clinical baseline assessment prior to shunting are shown in Table I.

Table I. Clinical features and data of the patients (N=30)

		Number	%
1.	Age (mean years)	64	
2.	Symptoms at baseline		
	Gait disturbance	30	100%
	Mental disorder		
	-memory decline	30	100%
	-cognitive dysfunction	8	27%
	-psychological disturbances	9	30%
	-personality disorder	11	36%
	-mood disorder	2	7%
	Urinary incontinence	20	67%
3.	Symptom duration at time of shunting		
	3 months	8	27%
	6 months	12	40%
	1 year	2	7%
	2 years	4	13%
	3 years	4	13%

All thirty patients were over 60 years old, with a mean age of 64 years. All patients by the time of their presentation had memory difficulties, 8 presented with cognitive dysfunction, 9 had psychological disturbances, 11 personality disorder and 2 presented with mood disorder. All the patients had gait and balance disability, and almost half of them needed assistance in order to walk. Twenty patients had urinary incontinence, at the time of presentation. The overall duration of the symptoms before the shunting operation varied from 6 months to 3 years. In the 6-month postoperative follow-up, ten patients were evaluated as having sufficient improvement, sixteen patients presented average improvement and four patients were evaluated as having bad outcome. The criteria for clinical outcome assessment are described in detail in the section Methods (see above). The follow-up results are shown in Table II. Overall, a favorable outcome after shunt surgery was seen in 26/30 (87%) of our INPH patients.

Table II. Six-month outcome after shunting (N=30).

Outcome	*Number (%)*
Good improvement	10 (34%)
Average improvement	16 (53%)
Bad outcome	4 (13%)

DISCUSSION

INPH was initially described as a treatable form of dementia. Recent estimates of the incidence of this condition are in the region of 6% of patients with dementia. The symptoms of INPH can vary among individuals and may be confused with those of patients with multi-infarct dementia, dementia of the Alzheimer type, or even Parkinson's disease. The pathophysiological mechanism of the disease is correlated with the CSF dynamic disturbance, the expansion of the ventricular system and the resultant blood flow decrement and metabolic disturbance of the subcortical structures [6, 7]. There is some evidence to suggest that the cerebral vasculature may have a role in the pathogenesis of INPH and some studies have shown that a global cerebral blood flow reduction, more severe in the frontal lobes, has been observed in patients with INPH [8,9]. Consequently, neurological functions like movement but also more complex functions (e.g. psychomotor speed, attention and concentration, memory and learning ability) may be impaired, explaining the presence of gait disturbance, cognitive deterioration and incontinence seen in most hydrocephalic patients [10].

Shunting INPH patients is controversial because of the difficulty in distinguishing it from other neurodegenerative conditions. Thus, accurate diagnosis of INPH is the key to successful treatment. Clinical, radiological (CT, MRI) and invasive diagnostic procedures (diagnostic CSF removal, continuous intracranial pressure monitoring and hydrodynamic study methods) have been suggested as helpful diagnostic tools for pre-selection of possible shunt candidates [11,12]. Functional neuroimaging techniques (CT perfusion, fMRI, brain SPECT, MRI spectroscopy) are more attractive for predicting outcome in patients with INPH due to their noninvasive nature. There are studies showing significant improvement in motor performance after CSF removal, being accompanied by bilateral increased activation in the supplementary motor area [13-15]. However, the diagnostic reliability, prognostic value and complications of such diagnostic concepts are still a matter of debate and the question still arises as to whether the benefits outweigh the risks in shunting INPH patients. To date, there is no standard for outcome assessment of shunt treatment and the literature available on this topic has been marked by various definitions of clinical improvement and varying postoperative follow-up protocols and periods. Traditionally, gait has been the motor dysfunction most studied regarding improvement after CSF shunting [3]. Variable improvement rates reported are not only because of different criteria for selection of patients but also because of different post-operative assessment procedures and follow-up intervals. Among various studies, overall success rate of shunt surgery ranges from 30 to 96% [4,16], and reported long-term effects also vary considerably from 26 to

50% [17, 18]. A long-term study following 25 patients for 5 years, showed that 47% felt their gait to be better, 29% felt their urinary symptoms to be better and 38% felt their memory to be better than in the preoperative state [19]. In our retrospective study of 30 INPH patients, we found significant improvement in clinical symptoms after 6 months of shunt treatment. The overall beneficial outcome after shunting was 87%. We believe that there could be two reasons explaining the apparently high rate of success in our study. A great percentage of patients had the classic triad of symptoms during pre-operative clinical assessment, so there was a relatively diagnostic certainty of INPH and also, the short-term follow-up probably could not detect any shunt malfunction, which is a frequent cause of long-term poor outcome [20].

CONCLUSION

In conclusion, we suggest that CSF shunting in INPH patients is beneficial for their short-term positive clinical outcome, quality of life and day-to-day functioning. Positive outcome is also highly correlated with immediate and accurate diagnosis.

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AUTHOR'S PROFILE

1. Dr. Leonidas Grigorakos is an Associate Professor of Medicine (ICU) at the National and Kapodistrian University of Athens (Faculty of Nursing), and Director of the Intensive Care Unit at Trauma Hospital of Athens, KAT, Kifissia, Athens, Greece.
2. Dr. Lavrentios Roussos is Director of the Neurosurgery Department at Trauma Hospital of Athens, KAT, Kifissia, Athens, Greece.
3. Dr. Pavlos Myrianthefs is Professor of Medicine (ICU) at National and Kapodistrian University of Athens (Faculty of Nursing).
4. Dr. Ioannis Alamanos is Director of the Intensive Care Unit at Trauma Hospital of Athens, KAT, Kifissia, Athens, Greece.
5. Dr. Christos Kelesis is Neurosurgerist at the General Hospital Athens "Korgialenio-Benakeio", Hellenic Red Cross, Greece.
6. Prof. Cryssoula Lemonidou is Head of the Faculty of Nursing at the National and Kapodistrian University of Athens. She teaches fundamentals of and nursing deontology, research methods and special issues in nursing.
7. Dr. Nikolaos Sakellaridis is Director of the Neurosuyrgery Department at Tzaneio General Hospital of Pireus, Greece.