

Management of Idiopathic Chronic Hydrocephalus of the Adult in Guinea: A Prospective Study in 16 Patients

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Abstract

Object: Incidence of Idiopathic chronic hydrocephalus of the adult (ICHA) is increasing in Guinea due to the aging of the population. The goal of this study was to describe its epidemiology, clinical presentation, and surgical outcome in a low-resource medical environment. Method: Sixteen patients operated for a probable ICHA were prospectively studied according to a uniform protocol from June 2019 to December 2020. On computerized tomography, all operated patients had a clinical triad of gait disturbance, incontinence, dementia associated with ventriculomegaly, and transependymal edema. In addition, all patients underwent a single lumbar tap, improved clinically, and were subsequently offered a shunt consisting of a simple catheter in 37.5% and a non-adjustable high-pressure valve in 62.5%. Result: The mean age was 68.31 ± 10.4 (range 49 - 89). The sex ratio H/F was 1.67/1. Clinically, the most frequent comorbidity was a combination of hypertension and diabetes in 56.5% of cases. The mean time to diagnosis was 8.31 ± 2.75 months (range 3 - 14). The immediate postoperative mortality was 12.5% from a status epilepticus and pulmonary sepsis. In addition, we observed 2 cases of chronic subdural hematoma (12.5%) and 2 cases of shunt dysfunction (12.5%) in the first three months. The functional outcome was good in 23% of patients at one month (N = 13), 50% at three months (N = 10), and 87.5% at six months (N = 8). Conclusion: In carefully selected cases, the surgical outcome of ICHA in Guinea is comparable to high-income countries. Efforts need to be put in helping patients get covered with universal insurance and generally promote Neurosurgery in the country to improve the quality of care.

Keywords

Idiopathic Normal Pressure Hydrocephalus, Comorbidities, Ventriculoperitoneal Shunt, Lumbar Puncture

1. Introduction

Idiopathic Chronic Hydrocephalus of the Adult (ICHA) is a condition defined by an increase in the volume of brain ventricles resulting in a clinical syndrome characterized by gait disturbance, urinary incontinence, and cognitive decline and not caused by another central nervous system injury (infectious, vascular, or traumatic) [1]. Therefore, the initial observation of a normal Cerebrospinal fluid (CSF) pressure at lumbar puncture is no longer a criterion for affirming the diagnosis, which has helped move away from terminological discussions with normal pressure hydrocephalus in the French medical literature [2].

Appropriately identified, ICHA is a potentially reversible cause of dementia through permanent shunt placement. However, the clinical presentation being variable, the diagnosis and the management are perceived as a real challenge in Guinea, a low-income country with minimal medical resources, albeit an increasingly frequent cause of referral in neurosurgery.

This study aims to assess the extent of this disease in our practice and to describe the conditions, results, and problems of its management in Guinea.

2. Patients and Methods

This is a prospective study of patients operated on probable iCHA according to the international study guidelines for diagnosing and managing idiopathic Normal Pressure Hydrocephalus [3] from June 2019 to December 2020. It was carried out at the CHU of Donka, Guinea, a country of 13 million population in which Neurosurgery is in its earlier phase of development. The whole economic burden of medico-surgical care is entirely on each patient.

Patients with a history of stroke, neurotrauma, or brain tumor were excluded. All the patients underwent a brain scan and a depletive lumbar puncture. They were kept in hospital for 12 hours to evaluate the motor and executive functions by the up-and-go test in patients who could stand. This test asks the patient to stand up, walk 3 meters before returning to his starting position in less than 10 seconds [4]. Patients were then sent home, and families were asked to record any global change in cognitive status and motor disorders. In case of any clinical improvement, the selected patients were subsequently admitted for ventriculoperitoneal shunt either with a single catheter ligated distally with lateral fenestrations or with non-adjustable high-pressure valves. After the surgery, each patient was asked to lie flat for two days, then authorized to stand progressively as tolerated. Patients were seen at 1-month intervals for the first three months for clinical signs of hypodrainage and chronic subdural hematoma treated when appropriate by temporary distal catheter ligation and hematoma evacuation.

The parameters collected for the study were age, gender, time from admission to first symptoms, presence of comorbidities, clinical data before and after depletive lumbar puncture, Evans's index, and presence of transependymal edema in the scanner, shunt device used, immediate postoperative complications and during follow-up. The overall clinical course after the procedure was evaluated at 1, 3 and 6 months postoperative according to the classification of Bret and Chazal [5]. Thus, the functional status was classified as good in case of a return to the previous state or autonomy for the acts of daily life (defined as the primary success variable); average results with a real but incomplete benefit that does not allow for independent living; and poor results for cases of stability or aggravation. Death represents the final grade of this classification.

Qualitative variables were expressed in terms of numbers and percentages, and quantitative data in terms of averages with their respective standard deviation sometimes presented in tabular form. The Excel software for Windows Office 2018 has been used.

3. Results

After applying our inclusion and exclusion criteria, 16 files were selected for analysis, *i.e.*, 4% of patients operated in the department of neurosurgery during the study period. Extreme ages ranged from 49 - 89 years with an average of 68.31 ± 10.4 years and a peak frequency in the 61 - 70 age group (**Table 1**). The predominance was male with a sex ratio of H/F of 1.67/1. Clinically, 15 out of 16 patients had at least one comorbidity, the most frequent of which was the combination of high blood pressure and diabetes in 56.5% of cases (**Table 2**).

Age (years)	Number	Percentage
40 - 50	1	6.25
51 - 60	2	12.50
61 - 70	6	37.50
71 - 80	5	31.25
81 - 90	2	12.50
Total	16	100.00

Table 2. Patients distribution according to comorbidities.

Comorbidity	Number	Percentage
Hypertension (HTN)	2	12.5
Diabetes (DM)	1	6.25
HTN + DM	9	56.5
Benign prostatic Hyperplasia (BPH)	4	25.00
Lumbar Canal Stenosis (LCS)	1	6.25

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The mean time to diagnosis of our patients was 8.31 ± 2.75 months with extremes of 3 - 14 months. Adam Hakim's symptomatologic triad was complete in all patients. Two patients (12.5%) were bedridden with an impossible sitting position; 5 patients (31.2%) could sit with armrests, and nine patients (56.2%) had apraxia for walking. All patients experienced significant psychomotor retardation with apathy and temporospatial disorientation, and 4 of these patients developed akinetic mutism (25%). Thirteen patients (81.25%) had urinary incontinence associated with fecal incontinence in 3 of them. Five patients (31.5%) had a urinary urgency. The non-contrast head CT showed ventricular dilation and transependymal edema in all our patients with an average Evans index of 0.36 \pm 0.047 and extremes of 0.28 - 0.43. The depletive lumbar puncture improved motor execution on the up-and-go test in 10 cases (62.5%) and a subjective functional improvement perceived by the family at home in all cases. Six patients (37.5%) underwent surgery using a single catheter without a valve, and ten patients received a high-pressure valve (62.5%).

The postoperative course was marked in the first three weeks by 1 case of status epilepticus leading to death and one case of pulmonary sepsis. We observed 1 case of subdural hematoma at one month, who refused to be treated and subsequently lost to follow up; and a second one at three months, treated with temporary catheter ligation and burr-hole drainage of the hematoma. The other postoperative complications diagnosed in the follow-up were two cases of valve dysfunction; the first at one month refused the treatment, and was transferred abroad by the family. The second one at three months benefited from a shunt revision with a non-adjustable medium pressure valve. The functional success rate (good result) was 23% at 1 month (n = 13), 50% at 3 months (N = 10), and 87.5% at 6 months (N = 8) (Figure 1).

4. Discussion

ICHA is a good representative model of the long-time dogma that neurosurgery is too expansive in low resource settings and is perceived this way by hospital managers, doctors of other specialties, patients, and the public in Guinea. Some





evidence suggests that surgical treatment of hydrocephalus is cost-effective in most high-income countries [6] [7].

Still, in a setting where patients and families are responsible for the medical and pharmaceutical bills, the standard unpredictability of shunt surgical results and complications makes it challenging to integrate neurosurgical diseases like ICHA in the national public health network.

The aging population and the epidemiological transition are causing an exponential increase in the incidence of ICHA and should lead us to pay particular attention to it.

Life expectancy in the Republic of Guinea has increased from 35 years in 1965 to 61 years in 2019 [8]. This aging population has led to an increase in the number of neurological visits for dementia syndromes associated with significant motor disorders and urinary incontinence transiently reversed by a depletive lumbar puncture, hence the suspicion of ICHA. The average age of 67.5 years and the peak incidence in the 6th decade found in our study are slightly younger than the literature [9]. Prevalence was estimated to be 1.30%, and its incidence was 0.12% in a literature review conducted in 2015 [9]. In Africa, the prevalence was mainly determined in neurosurgery services. It averaged 2.5 cases per year in Dakar between 1997 and 2003 [10]. Fourcases were reported by Magadji et al. in 2015 in Yaoundé, Cameroun [11]. In our study, 16 cases were collected over 18 months. These figures, under-estimated mainly, are difficult to compare because of their temporality (the Dakar study dates back more than 15 years) and the significant variability in the diagnostic criteria and methodologies used. They do, however, reflect the growing interest in this condition in the continent's neurosurgery services.

Although the mechanism of occurrence of ICHA is still poorly understood, the most described physiopathological models are vascular: ischemia of the white peri-ependymal substance; reduction of venous compliance; redistribution of LCS pulsatile streams [12]. These assumptions are reinforced by the high frequency of vascular comorbidities in patients with probable ICHA: 56% of our patients had hypertension/diabetes, in line with the literature [13].

Gait disturbances are known to appear early in the course of the disease [14]. For a long time, they were the unique symptom in one of our patients, initially diagnosed with lumbar canal stenosis and operated on without improvement. They have subsequently evolved in 43.7% of our patients into an overall motor regression as described in 2005 by Chazal "in the reverse direction of the early childhood motor milestone chronology" [15]. Cognitive impairment subsequently began as a personality change, mood disorder, and then evolved into advanced psychomotor retardation in all cases and akinetic mutism in 37.5% of cases.

The nature of this cognitive profile and its evolution appearing clearly after motor disorders have been described as favoring the suspicion of ICHA over Alzheimer's Disease [16]. At the most severe stages of cognitive decline, as is the

case for our patients at diagnosis, this clinical distinction is more difficult to achieve [16]. Urinary disorders have evolved concomitantly with gait disorders in 31.2% of cases and are well described in the literature [17]. Nocturnal urinary urgency has been a source of misdiagnosis in three patients who underwent surgery for benign prostatic hypertrophy before being referred to neurosurgery for incontinence and cognitive impairment.

The clinical originality of our series compared to the literature lies in the completeness of Adam Hakim's triad in all our patients (this was one of the inclusion criteria we chose) and the severity of the clinical signs at the time of diagnosis. It reflects the long delay in diagnosis that we have found and the lack of knowledge of this pathology in the national care pathways.

Coupling the search for periventricular lucency and the measure of the Evans index on non-contrast head CT scan allows for quantification of ventricular dilatation, differentiating it from simple cortical-subcortical atrophy and also help to eliminate other apparent causes of secondary hydrocephalus [18], in the setting of the severe clinical profile of patients in our series. Unfortunately, brain MRIs are not affordable, and the contemporary sophistication in the volumetric and dynamic sequences for the diagnosis of ICHA [19] are not available and not necessary in Guinea, in our opinion.

Depletive lumbar puncture aims to show a transient clinical improvement (mainly on motor disorders) to be measured in the first 24 hours to reduce the number of false negatives [20], and whose positive predictive value would be 90% - 100% improvement after shunting. There are many tests to assess this improvement in the literature [21], and none were applied to our context, given the severity of the disorders. The only test used was an untimed version of the up-and-go test and the overall subjective impression of close family caregivers. All patients experienced a marked improvement in motor and cognitive disorders after depletive lumbar puncture. The added benefit of leaving this subjective assessment to the family was that they would adhere to the surgical indication in a context where neurosurgery is still an unknown discipline in the general population.

The curative treatment is surgical and is based on the internal derivation of the LCS to a site of extracranial resorption. In sub-Saharan Africa and our study, a ventriculoperitoneal derivation is the most used technique [10] [11], as in most Western studies [22]. It is the type of valve used that differs according to the socio-economic level of the study population. Adjustable valves make it possible to compensate for the complications due to initial low or high pressure [23]. However, the flow-regulating valves, non-externally adjustable valves, may have their place in a social context in which the postoperative follow-up of the patient is uncertain or even impossible [24]. In extreme conditions like ours, a single shunt catheter was used without a valve in 37.5 percent of cases. We used non-adjustable Chhabra valves in all others, mainly tested on pediatric series [25]. According to Warf and Coll, they have a very accessible cost and would not show any significant difference in terms of results and complications compared with the standard adjustable valves [25]. Despite the difference in materials used, the rates of complications of our study are comparable to those of the literature [26] and relate mainly to postoperative infection (0.06% of the cases in our study), chronic subdural hematomas (12.5% of the cases) and valve dysfunctions (12.5% of the cases). The overall functional result we assessed at one month, three months, and six months also appear to be in line with that of the literature [27], with a more significant improvement between 3 and 5 months [28].

These comparisons must take into account many limitations of our research: Our patients were not followed beyond six months, and only the patients with good and average results showed up at the 6-month follow up evaluation; one patient with a subdural hematoma and another with a valve dysfunction refused the treatment offered to them, perceiving the occurrence of complications as related to poor quality of care; We didn't precisely measure the opening pressure of the CSF during the depletive lumbar puncture; no brain scan was performed during follow-up, except for cases of complications confirmed by clinical examination; among the clinical signs, only motor symptoms were objectively measurable and improved correlating with the satisfaction of close caregivers, who were more reluctant to complete micturition schedules and generally discuss details of urinary continence and cognitive impairment of patients during follow-up.

5. Conclusion

A ventriculoperitoneal shunt is a safe and effective treatment of probable ICHA in selected cases. The functional outcome and complications rates appear to be equivalent in low-income countries compared to high-income countries. But the perception of the quality of neurosurgical care in the general public is altered by the fact that neurosurgery is still new in Guinea, and patients don't have the financial resources to afford more appropriate materials and further treatment of shunt complications. The lack of universal health coverage in Guinea is a major obstacle in the development of Neurosurgery in the country.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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