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Research Article

Chronic Hydrocephalus in Adults (ACH): Experience of the Neurosurgery Team at the Yalgado Ouédraogo University Hospital (CHU-YO)

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Abstract

Introduction: Adult Chronic hydrocephalus(ACH) was first described in 1965 as normal pressure hydrocephalus (NPH) by Adams and Hakim [1].

Its initial definition was based on the clinical triad (Adams and Hakim triad) associating cognitive, gait and sphincter disorders in association with dilatation of the ventricles and normal CSF pressure on lumbar puncture (LP).

The aim of this study is to lay the foundations of the epidemiology of ACH in Burkina Faso and to describe the difficulties associated with its management from January 2015 to December 2021.

Patients and Method: This was a cross-sectional, descriptive, retrospective study over a 7-year period, including all adult patients managed for chronic hydrocephalus and with usable clinical records.

Results: Thirty-four cases of chronic adult hydrocephalus were included. The mean age was 51.8 years. The sex ratio was 3.85.

The clinical signs were gait disorders in 32 cases (94.12%), intellectual disorders in 26 cases (76.47%), and sphincter disorders in 25 cases (73.53%).

Brain CT scan showed tetraventricular dilatation in 24 patients (70.6%). Preoperative lumbar puncture was performed in 9 cases (26.47%). Thirty (30) cases (88.24%) underwent a ventriculoperitoneal shunt and only one case (2.94%) underwent a ventriculocisternostomy.

After an mean follow-up of 7 months, the results of the treatment were favorable, with improvement in 32 cases (94.1%).

Conclusion: Chronic hydrocephalus in adults is still an uncommon condition in our context. Treatment significantly improves quality of life. Mass screening could be used to recruit a large number of patients for treatment.

Keywords: Adams Hakim Triad; Chronic Hydrocephalus; Adults; Shunting

Abbreviations

ACH: Chronic Hydrocephalus in Adults; LP: Lumbar Puncture; CT Scan: Computerized Tomography Scans; NPH: Normal Pressure Hydrocephalus; CSF: Cerebrospinal Fluid

Introduction

Hydrocephalus is a distension of the ventricular cavities due to an accumulation of cerebrospinal fluid (CSF) responsible for intraventricular hyperpressure. Chronic adult hydrocephalus was first described in 1965 as normal pressure hydrocephalus (NPH) by Adams and Hakim [1].

Its initial definition was based on the clinical triad of cognitive, gait and sphincter disorders associated with dilatation of the ventricles and normal CSF pressure on lumbar puncture (LP). Nowadays, chronic hydrocephalus in adults (ACH) refers to a clinico-radiological entity combining a clinical triad of gait disorders, neuro-cognitive deterioration (dementia syndrome) and urinary incontinence with ventricular ectasia on imaging.

Considering the conditions of the diagnosis based mainly on the clinical triad and ventricular ectasia in most neurosurgical teams without study of intraventricular pressures and the observation of a tendency for these pressures to rise in patients with NPH, the terminology was revised with the proposal of the term chronic hydrocephalus without taking into account CSF pressures or age [2]. Since then, chronic hydrocephalus has been the most appropriate term for this condition [2,3]. Two forms of chronic hydrocephalus can be described: idiopathic and secondary.

This condition is particularly relevant in developing countries, where geriatric medicine is underdeveloped.

The aim of this study is to lay the foundations of the epidemiology of ACH in Burkina Faso and to describe the difficulties associated with its management.

Patients and Methods

This was a cross-sectional, descriptive, retrospective study conducted from 2015 to 2021 in the neurosurgery department of the Yalgado Ouédraogo University Hospital. It included all patients admitted and managed for signs suggestive of the Adam and Hakim clinical triad with ventricular ectasia on imaging. Two cases were excluded for lack of usable records.

Results

Epidemiology

In 7 years, 34 patients were treated. These patients comprised 27 men (79.41%) and 7 women, with a sex ratio of 3.85.

The mean age was 51.8, with extremes of 24 and 81.

Contributing factors were found in 22 cases (64.71%), including a history of cranioencephalic trauma and bacterial meningitis. We were therefore able to identify two clinical forms: idiopathic chronic hydrocephalus (12 cases; 35.29%) and secondary chronic hydrocephalus (22 cases; 64.71%).

Diagnosis

The mean consultation time was 08 months, with extremes of 1 month and 36 months. Gait disorders, dementia and urinary incontinence were noted in 32 patients (94.12%), 26 cases (76.47%) and 25 cases (73.53%) respectively. Table 1 summarises the different symptomatic presentations of the patients.

	Number	Percentage (%)
Mono-symptomatic	6	17,65
Walking disorders Dementia syndrome	5 1	14,71 2,94
Bi-symptomatic Gait disorder + dementia syndrome Walking disorders + urinary incontinence Dementia + urinary incontinence	6 3 2 1	17,65 8,83 5,88 2,94
Complete triad (difficulty walking + dementia + urinary incontinence)	22	64,70
Total	34	100

Table 1: Distribution of patients by reason for consultation.

Cranioencephalic CT scans were performed in all patients in our series and revealed 22 cases (64.71%) of tetraventricular hydrocephalus and 12 cases (35.29%) of triventricular hydrocephalus. Figure 1 illustrates the hydrocephalus of one of the patients in our series.

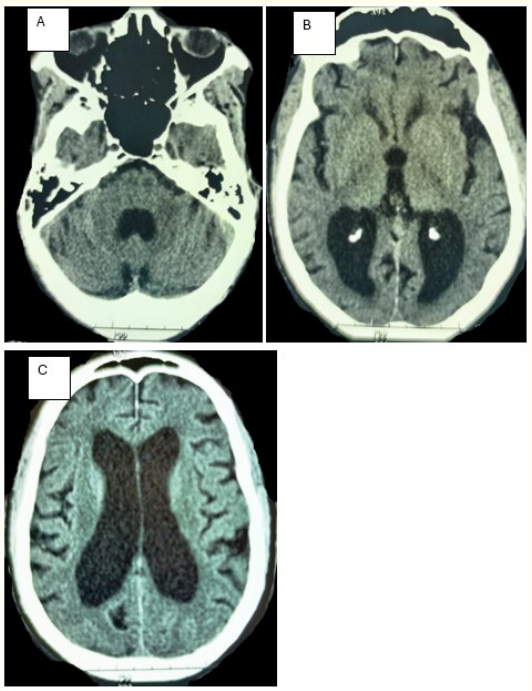

Figure 1: Cranioencephalic CT scan of a patient admitted with Adams and Hakim triad. These axial sections show tetraventricular hydrocephalus with dilatation of the fourth ventricle (A), dilatation of the third ventricle (B), dilatation of the lateral ventricles (C), cortical atrophy and periventricular hypodensity of transependimal resorption.

Therapeutic tests

A series of 2 to 3 subtractive lumbar punctures per patient was performed in 9 patients (26.47%). The volume of CSF evacuated varied between 30 and 50 milliliters. CSF pressure was not measured in any of our patients. This test was conclusive in all cases, with a transient improvement in symptoms.

Treatment

The average waiting time was 36.3 days, with extremes of 7 days and 180 days. Surgery was proposed in all cases, but only in 31 cases, i.e. 91.18% of cases, after varying periods of hesitation about accepting surgical treatment. In the other cases (3 cases; 8.82%), because of non-acceptance of surgery, medical treatment (acetazolamide) of short duration (2 weeks) was proposed.

Surgical treatment included ventriculoperitoneal shunting in 30 cases (88.24%) and ventriculocisternostomy in 1 case (2.94%). A medium-pressure valve was used in all cases.

Evolution

In our series, the average post-operative follow-up time was 7 months, with extremes of 2 months and 18 months.

In the short term, complications such as chronic subdural haematoma due to hyperdrainage were observed in 2 patients (6.45%) and led to secondary revision surgery to replace the medium-pressure valve with a high-pressure valve after evacuation of the haematoma.

After an average postoperative follow-up of 7 months, we noted clinical improvement in 32 cases (94.1%), including a return to the previous state with complete recovery in 24 patients (70.6%) and incomplete recovery in 8 patients (23.5%), and clinical deterioration in 2 cases (5.9%).

Discussion

Adult chronic hydrocephalus was first described in 1965 by Adams and Hakim [1] under the term normal pressure hydrocephalus (NPH) with a precise definition based on a clinical triad that combined cognitive disorders, gait disorders and sphincter disorders associated with dilatation of the ventricles and normal cerebrospinal fluid (CSF) pressure on lumbar puncture (LP). Although both tetraventricular and triventricular forms are described in the first publication, the most commonly described type of chronic hydrocephalus remains tetraventricular.

Pathophysiology

The mechanisms by which hydrocephalus occurs can be summed up as an imbalance between CSF secretion, flow and resorption. There are three pathophysiological modes that can lead to excessive accumulation of CSF in the ventricular system and result in hydrocephalus: hypersecretion, obstruction of CSF flow (obstructive hydrocephalus) and resorption deficit [4]. The most common pathophysiological mechanism in chronic tetraventricular hydrocephalus is resorption deficit. Chronic triventricular hydrocephalus may be related to degenerative stenosis of the midbrain aqueduct [5] or untreated acute hydrocephalus, initially stabilised and secondarily decompensated in the chronic form with the characteristic clinical triad [6].

Several theories have been put forward to explain the different elements of the clinical triad.

Progressive ventricular dilatation and pulsations of the cortex against the internal table of the calvarium could generate radial shearing forces on the cerebral parenchyma, thus contributing to the onset of dementia [6-8].

Stretching and compression of the fibers of the corticospinal tract in the corona radiata near the lateral ventricles due to interstitial edema linked to the increase in intracranial pressure, and poor perfusion of the periventricular white matter and the prefrontal regions are all disturbances of the motor innervation of the limbs which may be the cause of walking disorders [8,9].

At an early stage, the periventricular sacral fibers of the corticospinal tract are stretched, leading to a loss of voluntary (supraspinal) control of bladder contractions [10]. At a more advanced stage of the disease, dementia may contribute to incontinence [11].

Epidemiology aspects

Chronic hydrocephalus is a rare condition that affects 0.1-0.5% of patients over the age of 60 [12] but its frequency may be underestimated due to diagnostic uncertainties [13]. The idiopathic form is less common than the secondary form. The idiopathic form occurs in people over 60 years of age [6,12,14] but the secondary form occurs at any age. No significant gender predominance has been demonstrated [8,14] but some authors have reported a male predominance in 66.7% to 83.3% of cases [6,15] mainly related to risk factors in the secondary form. In our series, a male predominance of 79.41% was found, which could be explained by the greater frequency of risk factors in men.

Diagnosis aspects

The clinical diagnosis of chronic hydrocephalus is very difficult because of the neurodegenerative diseases that can present with the same symptoms [6,14]. The clinical triad of Adams and Hakim is described in 50% to 70% of cases [6,12,14,16]. In our series, the complete triad was present in 64.70% of cases. Problems with balance and walking, which are symmetrical unlike idiopathic Parkinson's disease, are usually the first signs of the disease and the most severe [12,17]. In our study, 94.12% of our patients presented with gait disorders, and this high frequency may be linked to the long delay in diagnosis.

Cognitive disorders are polymorphous and very difficult to diagnose precisely, requiring neuropsychological tests. These signs usually appear late in the final phase of the disease. At the time of diagnosis, intellectual disorders are present in 66% to 83% of cases [6,13,16]. Extreme cases of genuine psychiatric symptoms may occur [7]. In the present series, cognitive disorders were observed in 76.47% of cases, but this frequency may be underestimated due to the non-use of neuropsychological tests. Urinary disorders vary from simple bladder hyperactivity, which is more frequent and gives rise to urgenturia, to urinary incontinence per se, often associated with fecal incontinence, occurring late in the course of dementia [18-20]. In our case, the urinary disorder observed was sphincter incontinence, reflecting the long delay in diagnosis.

Cerebral imaging shows ventricular ectasia, which supports the diagnosis, specifies the type of ventricular ectasia (triventricular or tetraventricular) and contributes to the differential diagnosis by showing other pathologies likely to explain the patient's symptomatology [17,21].

Although CT and MRI are both effective in demonstrating ventricular ectasia, MRI remains the reference examination for signs of the Adams and Hakim [14]. MRI must combine morphological sequences (3D sequences with high T2 weighting) and flow-sensitive imaging (velocimetric sequences). In developing countries, brain scans are the most widely used due to their greater accessibility [15,22]. In our series, all patients underwent CT scan only.

Therapeutic tests aspects

The diagnostic difficulties of chronic hydrocephalus and the great variability in response to treatment of ventricular ectasia encourage practitioners to carry out therapeutic tests before proposing definitive treatment [5,17,23]. A distinction is made between clinical therapeutic tests, including lumbar puncture and external lumbar drainage over 72 hours, and manometric tests, including the perfusion test and intracranial pressure measurements. These positive therapeutic tests have a high positive predictive value for a satisfactory clinical response to surgical treatment. In our case, only the lumbar puncture therapeutic test was used in 26.47% of cases and was positive in all cases, thus contributing to acceptance of surgery. Given the high probability of association of other degenerative pathologies with chronic

hydrocephalus, it is recommended that associated pathologies be treated before considering either diagnostic and therapeutic tests or surgical management proper, especially since symptomatic resolution could motivate a diagnostic and therapeutic revision [17].

Therapeutic aspects

In the past, treatment modalities included lumbar punctures (a series of 2 to 3 successive punctures) [14,24], drug treatment with acetazolamide [25] and surgical treatment, but nowadays treatment is mainly surgical with ventriculoperitoneal shunting and ventriculocisternostomy. Ventriculoperitoneal shunting is the standard treatment because the typical form of chronic hydrocephalus is tetraventricular, with impaired resorption of CSF [9,14,21]. To date, there is no evidence that any particular type of valve is superior, but it is recognised that adjustable-pressure valves offer a greater possibility of adjusting drainage flow according to the patient's clinical response after surgery, and with a lower risk of complications [17,26,27].

Repeated lumbar punctures are characterised by a low success rate that remains transient, with an increased risk of infectious complications, and are therefore increasingly excluded from the list of therapeutic means [24].

Evolution aspects

Chronic hydrocephalus in adults shares the same postoperative complications as acute hydrocephalus, with a few minor differences [14,17,28]. The complications are infectious, functional (hyperdrainage with or without chronic subdural hematoma [9,29,30]. In our series, all the complications observed (6.45%) were functional complications, essentially related to the fixed-pressure shunt valves. In general, the degree of improvement was greater for gait disorders, cognitive disorders and urinary dysfunction [30-32]. The rate of postoperative clinical improvement varies from 69% to 100% for gait disorders, depending on the series [29,30,33] but urinary and cognitive disorders improve shortly after shunting [14].

Overall, when the indication for surgery is well established, clinical improvement is found in 70 to 90% of patients [9,16,21]. In our series, clinical improvement was noted in 94.1% of patients.

Conclusion

This study shows that adult chronic hydrocephalus is an uncommon condition in Burkina Faso. The diagnostic process is complex, especially in our context where geriatric medicine is underdeveloped. Early treatment can significantly improve quality of life. Mass screening could enable a large number of patients to be recruited for treatment and a better study of the particularities of this pathology in our country.

Conflicts of Interest

We declare no conflicts of interest in the development of this document.

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