Hydrocephalus in children less than 1 year of age in northern Mozambique

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Received: 22 May 14  Accepted: 22 September 14  Published: 08 December 14

Abstract

Background: In developed countries, the incidence of neonatal hydrocephalus ranges from 3 to 5 cases per 1000 live births, but little is known about the frequency of hydrocephalus in Africa. In Mozambique, there is no primary information related to this disorder, but using the above data, the expected incidence of neonatal hydrocephalus would range from 2900 to 4800 cases per year.

Methods: This study is based on 122 children younger than 1 year with neonatal hydrocephalus, followed up between January 2010 and December 2012, their origin and treatment, and aims to evaluate difficulties with diagnosis, treatment, and follow‑up in northern Mozambique.

Results: Identified cases were mainly less than 6 months old (77%), with severe macrocephaly and the classic stigmata of this condition. A high rate of follow‑up loss (44.3%) was detected, particularly among children from more distant locations. Our findings contrast with the expected 1000-1700 cases that would occur in the area during the study period, being considerably lower.

Conclusions: Hydrocephalus is a serious problem in sub-Saharan Africa, whose effects can be minimized by a better organization of the health system in hydrocephalus prevention, referral, and follow‑up. New management alternatives to provide treatment to more children with this disorder and reduction of the follow‑up difficulties caused due to geographical reasons for the children undergoing treatment are essential.

Key Words: Hydrocephalus, infant, mozambique, nampula, pediatric

INTRODUCTION

Since the middle of the last century, hydrocephalus has been treated using valve‑regulated ventricular derivations, which continue to be widely used.[15] The poor cognitive and motor development in children, the loss of the cognitive function, and visual loss can complicate untreated hydrocephalus and persist after treatment,[15,38] with the consequent impairment interfering in their development in an important way.
In developed countries, the incidence of congenital hydrocephalus has been estimated at 0.5 cases per 1000 live births and the incidence of neonatal hydrocephalus has been estimated at 5-5 cases per 1000 live births, with a male predominance. The incidence distribution is bimodal with most hydrocephalus cases occurring among children. It is one of the most common developmental disorders, surpassing Down’s syndrome or congenital deafness. It is, therefore, the most often-treated neurosurgical condition in pediatric practice.

Although hydrocephalus is reported to be more common in developing countries, its prevalence is yet to be determined. The current incidence of hydrocephalus in sub-Saharan Africa is unknown. In this region, it is estimated that less than 10% of cases are annually treated using the ventriculoperitoneal shunt (VPS) systems. It is known that many children with hydrocephalus in sub-Saharan Africa are not taken to health facilities for treatment due to poverty, erroneous cultural interpretations about hydrocephalus in children, and various other reasons. Most children who suffer from this medical condition unfortunately do not even have the chance to be treated.

The crude birth rate in Mozambique was 41.6/1000 in 2011, with a projected population of 23,049,621 inhabitants, which gives an estimate of 958,864 births/year. Using data from developed countries to estimate incidence, 480 new cases of congenital hydrocephalus and from 2900 and up to 4800 new cases of neonatal hydrocephalus are predicted each year in Mozambique.

Comparing the developing countries (as is the case of Mozambique) with developed countries, an incidence of congenital hydrocephalus greater than or equivalent to that in developed countries would be expected, but non-congenital (acquired) etiologies of hydrocephalus in children are expected to be different—the post-hemorrhagic cause associated with prematurity being the most common in Western countries and post-infectious hydrocephalus (PIHC) being more common in developing countries.

It is believed that the increased prevalence in Africa is due to an increase in birth rate and, thus, an increase in occurrence of congenital abnormalities, as well as increased rates of neonatal infections.

Low socioeconomic status is a risk factor for all non-genetic (acquired) defects, including hydrocephalus. Non-genetic structural defects can occur for several reasons. Hydrocephalus is strongly associated with spina bifida, and this defect is caused by a deficiency of folates/folic acid in the maternal diet. Multivitamin supplements during pregnancy have singly been shown to reduce the risk of hydrocephalus.

Low maternal age has been associated with a higher risk of hydrocephalus, probably because younger mothers do not undergo prenatal screening. Maternal age at first birth is lower in developing countries than in developed countries. In Mozambique, most primigravidae are in the age group ranging between 20 and 24 years.

In north Mozambique, prenatal care provided by health personnel (doctors, nurses, midwives or auxiliary or traditional birth attendants) reaches 90-96% of pregnant women; however, assisted deliveries and post-delivery/neonatal care by skilled health personnel remain low throughout the country (54%), ranging from 35.6% in Cabo Delgado province to 60.3% in Niassa province (Nampula province – 55.3%) in the northern region. These percentages are directly related to the low level of education of pregnant women.

The Nampula’s Central Hospital (HCN), a tertiary hospital, is the reference facility for the country’s northern region, having an estimated population of almost 8,000,000 inhabitants (more than one-third of the country’s population), with a crude birth rate of 41.6/1000 inhabitants (2011).

In this study, data were collected regarding the baseline clinical presentation and the initial approach to all cases of hydrocephalus (122) in patients under the age of 1 year treated at Nampula’s Central Hospital (HCN), from 1 January 2010 to 31 December 2011 and followed up until 31 December 2012.

**MATERIALS AND METHODS**

Between 1 January 2010 and 31 December 2011, 122 pediatric cases of hydrocephalus, under 1 year of age, were treated surgically at Nampula’s Central Hospital (HCN), from the provinces of Nampula, Cabo Delgado, and Niassa. of a total of 152 cases of operated hydrocephalus (570 neurosurgical procedures were performed during the same period).

The follow-up period was until 31 December 2012.

We included all cases of hydrocephalus patients younger than a year of age, submitted to surgery, who were referred to the Nampula’s Central Hospital (HCN) and came from the catchment area of the hospital, i.e. the provinces of Nampula, Cabo Delgado, and Niassa. The diagnosis was clinically based (signs of macrocephaly and intracranial hypertension) and confirmed by cranioencephalic computed tomography (CT). A referral network exists from primary health centers, to rural, provincial, and finally central-level hospitals, and some patients have to travel nearly 700 km to have access to the care of a neurosurgeon.

The provisional diagnosis is made clinically by the healthcare professional who observes the patient (at the
health center or rural hospital) and the child is sent to the provincial referral hospital. After evaluation at the provincial referral hospital, the child is remitted for evaluation by a neurosurgeon who, with the current clinical and CT information, establishes the diagnosis and defines the treatment plan.

During the data collection period, only one neurosurgeon was active at the Nampula’s Central Hospital (HCN).

Data were collected by the above-mentioned neurosurgeon, assisted by a graduate student of medicine, by completing a specific protocol for this purpose – after the parents or guardians of the children signed an informed consent.

Six HIV-infected and malnourished infants were admitted with hydrocephalus, but excluded from this study, as they presented gravely ill and were not able to be stabilized for anesthesia and surgical intervention. All six of these patients died.

For the cephalic perimeter assessment, performed by the same neurosurgeon, standard WHO head circumference by age Z score tables were used.[49]

The Alberta Infant Motor Scales (AIMS) tool was used for the evaluation of psychomotor development, and the results were grouped or summarized into four groups (appropriate, slight mental retardation, moderate mental retardation, and severe mental retardation).

The data were entered and analyzed with the software program IBM SPSS Statistics, version 20.0.

RESULTS

We calculated the expected number of cases corresponding to the 2-year period of this study. To do this, the official projections for the population by province, the crude birth rate, and the incidence rate of neonatal hydrocephalus in the developed world were used, since at present there is no data on the incidence of hydrocephalus in sub-Saharan Africa.[45] With this methodology, Mozambique is expected to have between 2900-4800 new cases of pediatric hydrocephalus each year, with 1000-1700 (in a conservative perspective) of those coming from the Northern region of the country.

For northern Mozambique, the expected incidence of neonatal hydrocephalus cases between 2010 and 2011 is shown in Table 1, and in summary only about 15% of the expected cases were referred. However, there was no statistically significant difference between the expected cases and the cases diagnosed/referred (P = 0.200; Chi square), when different provinces were compared with each other.

Among the cases referred to the Nampula’s Central Hospital (HCN), 8% came from the city of Nampula, 53% from Nampula province (average distance 350 km), 29% from the province of Cabo Delgado (405 km from the province capital – Pemba), and 10% from Niassa province (695 km from the province capital – Lichinga).

There was no gender predominance (sex ratio 1:1) and 77% were younger than 6 months. Distribution by age is shown in Figure 1.

Macrocephaly was present in all children (reason for referral), with 77% infants having above the 95th percentile of cephalic perimeter, 16.4% between 95th and 90th percentile, and 6.6% between 90th and 75th percentile. The anterior fontanelle was taut without bulging in 56.7% of patients, bulging and slightly depressible in 28.3%, bulging and not depressible in 6.7%, and without bulging and depressible in 8.3%. Sunset eye sign were present in 47.5%.

Psychomotor development was adequate in 45.9%, 39.3% presented a slight retardation, 12.3% with a moderate retardation and 2.5% had a severe retardation.

All cases underwent surgical treatment, which consisted in setting in place a VPS system. The National Health System in Mozambique currently offers the Chhabra Surgiwear® valve system; however, in 8% of cases, the non-valved Harare type system had to be used because of rupture of the stock.

The duration of postoperative hospitalization for cases without immediate surgical complications was, on average, 8 days, and for those cases with any post-operative complication was 26.4 days (minimum 13 days, maximum 68 days).

| Table 1: Expected incidence and referred cases of neonatal hydrocephalus during a period of 2 years in northern Mozambique (2010-2011) |
|---------------------------------------------------------------|
| **Est.* population** | **Est.* births** | **EI** | **RC (n)** | **RC (%)** |
| population** | (2010+2011) | (2 years) | (n) | (%) |
| Mozambique | 23,049,621 | 1,796,086 | 5,388-8,980 | 122 | 11.0-18.3 |
| Northern provinces | 7,977,134 | 652,549 | 1,957-3,263 | 71 | 3.7-6.2 |
| Nampula | 4,647,841 | 381,790 | 1,145-1,909 | 39 | 5.3-8.8 |
| Cabo delgado | 1,797,335 | 148,159 | 444-741 | 12 | 2.0-3.3 |
| Niassa | 1,531,958 | 122,600 | 368-613 | 12 | 2.0-3.3 |

*Estimated, **Expected incidence, ***Referred cases (n), ****Percentage of referred cases (%) in relation to expected incidence
Global complication rate was 5.7% – three cases of obstruction of the ventricular system, one migration of the ventricular catheter, one migration of the abdominal catheter, and one peritonitis.

During the study period (36 months), the follow-up loss was 44.3% (54 patients): 1 patient in the city of Nampula (representing 10% of the patients coming from this location), 20 patients in Nampula province (representing about 30% of the patients coming from this location), and more than 70% patients lost to follow-up from the provinces of Cabo Delgado and Niassa (35 patients).

At follow-up (56%), between 13 and 34 months (average 22.4 months), all patients had a functioning system.

**DISCUSSION**

The natural history of hydrocephalus leads to severe neurological deficits associated with major morbidity, causing sufferers of this condition to be totally dependent in their daily lives and in need of closer medical follow-up, which leads to a large social and economic impact on families.[9] Appropriate and timely treatment can reduce this impact.[40]

Hydrocephalus has been known since 1811 when Cooke reported a case.[8] Since then, definition, etiology, symptoms, classification, and treatment options are debated more often, and there are (on Pubmed database) more than 27,000 articles related to the issue. In Africa, the first article related to hydrocephalus was published by Clifford in 1963.[7] and 20 years later and important case series on hydrocephalus in Africa was published by Peacock and Currer.[31] Less than 1% of articles about hydrocephalus, indexed to Pubmed are related to Africa. The care of hydrocephalus in sub-Saharan Africa is hampered by economic constraints (poverty), difficulties for patients and families in securing transportation means and access to adequate care, and cultural mistakes/misunderstandings related to hydrocephalus.[23,45]

There is no epidemiological data on hydrocephalus in Mozambique. Cerebrospinal fluid (CSF) shunt systems, as well as neurosurgeons able to set them in place, are not available for most children and, when available, most people are not able to acquire a VPS system. This paper is the basis for a more exhaustive study in future and seeks to contribute to the improvement of care of hydrocephalus in Mozambique.

The diagnosis of hydrocephalus is clearly underestimated in northern Mozambique, probably because personnel not trained to diagnose hydrocephalus perform deliveries and neonatal care. Moreover, neonatal care only reaches half of the pregnant women in northern Mozambique (50.4%),[26,27] which further implies a reduced capacity of diagnosis of neonatal hydrocephalus (50%). As shown in Table 1, only 20% of expected cases are referred for neurosurgeon evaluation.

Patients referred to the Nampula’s Central Hospital (HCN) arrive there with evolved forms and severe hydrocephalus stigmata – more than 90% with a cranial perimeter above the 90th percentile and taut fontanelle, 15% with a significantly retarded psychomotor development, and more than 50% with some degree of impairment. These evolved forms can be avoided by early diagnosis and timely treatment.

There is, therefore, a crucial need to train health personnel from the most basic care units for pregnant women and newborn to the care units for the general population in order to ensure early diagnosis and timely referral.

Most infants presented obstructive hydrocephalus (98.4%). This diagnosis was confirmed only with the use of cranioencephalic CT, since there is no magnetic resonance image (MRI) apparatus at Nampula’s Central Hospital (HCN). The main causes are presented in Figure 2.
In Africa, post-infectious etiology varies from 7% to 60%, and is correlated to the level of health of the considered population. The ratio of congenital to “post-meningitic” hydrocephalus in Zimbabwe (a neighbor country) was reported to be 2:1. PIHC represents one-third of this series and has therefore been included in reported literature.

Although the methods for the study of hydrocephalus are MRI and CT, the transfontanellar ultrasound plays an important role in the diagnosis and characterization of brain lesions in the newborn (e.g., hemorrhage, ventricular dilatation, leukomalacia, malformations, etc.). For its safety and ease of implementation and for the possibility of serial exams to assess the evolution of brain lesions, transfontanellar ultrasound is considered a method of choice in the evaluation of the newborn at risk, being even, in most cases, the only method needed. In Mozambique, this resource is not routinely used and it should be considered, since it is painless, relative easy to perform, more readily available than MRI and CT, and provides valuable clinical information. Capacitating Mozambican health care workers to perform this type of ultrasound examination is a priority.

Even with higher rates of major complications, VPS remains the primary treatment option in the orientation of hydrocephalus. The complication rates range between 1% and 60% by 2 years after surgery, including failures of the systems and infections. Until recently, VPS systems used in Mozambique were of the simple type, consisting of a silicone tube, reported in Zimbabwe and Malawi series. Those were rudimentary systems without a valve regulation and with inherent higher risks and complications. Currently, the Chhabra Surgiwear systems, manufactured in India, are in use, and costing the Mozambican Health System about $100 USD/unit. It is noteworthy that 89% of the complications occurred in Harare shunts and 11% in Chhabra. All cases underwent further surgery to remove the dysfunctional system and for installing a Chhabra Surgiwear valve system. The ideal VPS system does not exist, and the availability of each system has been a major limiting factor, rather than the surgeon’s preference. Chhabra Surgiwear VPS systems (now routinely used in Mozambique) have similar complication rates to systems used in the Western countries, without statistically significant differences in complications, as reported in a study in Uganda.

Operations took place in most of the cases within an acceptable time frame, given the logistic conditions. Surgeries that took place between 1 and 2 weeks after evaluation by a neurosurgeon were due mostly to lack of surgical time. In cases presenting with co-morbidities including respiratory and gastrointestinal infections, and malaria, surgery was delayed for 2 weeks. However, the diagnosis was late because most of the children presented severe stigmata of the clinical condition, which could be minimized by early diagnosis and referral.

In this series, only seven cases of complications related to the surgical procedure were reported (5.7%). The follow-up loss is considerable and it certainly affects this value, making the complication rate inconsiderable. Of course, a new healthcare orientation on hydrocephalus in Mozambique is of utmost importance to overcome this local difficulty. In the series of sub-Saharan Africa, the described complications vary between 7% and 69% and are related to mechanical causes (obstruction and VPS system migration) (11-54%) and infection (7-69%). Even when the VPS placement is possible, it is more dangerous in the African context than in developed countries because of the infectious complications and shunt’s malfunctioning. Infectious complications of VPS implantation can be reduced by implementing protocols that train staff in infection control measures related to the preoperative, operative, and postoperative period, as reported by Choux et al., being fundamental to adapt this knowledge to local reality. The follow-up losses are proportional to the distance of patients’ residences from the Nampula’s Central Hospital (HCN). The economic difficulties related to extreme poverty, poor road infrastructure, and long distances are challenging constraints for the appropriate follow-up of these children.

A large number of children may alternatively benefit from endoscopic third ventriculostomy (ETV), which is a treatment option, without heterologous material, with a long-term lower rate of associated complications and with a follow-up that we believe can be less frequent, thus reducing the socioeconomic impact on the families of these patients.

The average cost of this equipment is around $50,000 USD, with investment recovered after about 500 procedures, without including shorter periods of hospitalization, reduced need for hospital visits for follow-up, fewer complications, and its strong socioeconomic impact on families, as reported on ETV procedures.

**CONCLUSIONS**

Hydrocephalus in sub-Saharan Africa is a serious health problem. Although more investigation and publishing is increasingly being carried out, there is still little data on its incidence, prevalence, and causes.

To improve the quality of care of patients with hydrocephalus in a country with a social, economic, and cultural context, such as Mozambique, is a challenge that goes far beyond the aspect of neurosurgical treatment.
In northern Mozambique, like in the rest of the country, and in sub-Saharan Africa, there is an urgent need for strategies to address a medical condition that has devastating consequences when left untreated.

In addition to appropriate assistance during pregnancy and delivery, early diagnosis and appropriate orientation are essential.

Measures including folic acid supplementation for the prevention of neural tube defects, improved follow-up of pregnant women, improved public educations, improvement and capacity development in health centers, particularly in peripheral areas, can help improve early diagnosis, retention in care, and patient follow-up.

Due to the simplicity and lower price of the exam (compared with MRI and CT), the transfontanellar ultrasound should be considered for diagnosis in Mozambique.

There is a need to implement viable therapeutic options, such as ETV, which have lower costs for the health system in the short and medium term, giving the best result for the patient and having lower socioeconomic impact on the families of patients.

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