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Results of the Management of Paediatric Hydrocephalus at Teaching Hospital of Conakry-Guinea: Series of 167 Cases

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Abstract

Objective: The main goal of this study was to evaluate the long-term results of the management of paediatric hydrocephalus in the neurosurgery department of teaching hospital of Conakry. **Methods:** This was a retrospective multicentre study of descriptive and analytical type covering the period from January 1, 2012 to October 31, 2019. We collected 167 cases of paediatric hydrocephalus treated during our study period. **Results:** an annual average frequency of 21 cases/year. The mean age of the patients was 12,0158 ± 23,0889 months. The male sex was the most represented (sex ratio = 1.16). Macrocrania was the main reason for consultation (95.81%) and acquired aetiologies represented 56.84% of cases. The most performed surgical technique was the VP shunt (55.09%) and the timing of surgery was 13.38 ± 31.88 days. The shunt disfunction rate was 32.93% and the average rate of revision of the shunt was 1.25 revisions/patient. During our study we recorded 43.84% of death. **Conclusion:** The results of the management of paediatric hydrocephalus depend on the time to perform the surgery. Short and long-term follow-up is of paramount importance in the progression of the child treated for hydrocephalus. The transition period from paediatric age to adulthood would make it possible to better appreciate the impact of hydrocephalus on the psychomotor development of the child and his socio-professional integration.

Keywords

Results, Paediatric Hydrocephalus, Management, VP Shunt, ETV

1. Introduction

Paediatric hydrocephalus is a complex pathology most often diagnosed in child-

hood and requires long-term monitoring. It is a disorder of CSF hydrodynamics leading to dilatation of the ventricular system associated with higher intracranial pressure. Its multiple and diverse aetiologies are mostly congenital. Frequent in infants with serious consequences on the developing brain, it requires immediate diagnosis and surgical intervention. The aim of this study was to evaluate the long-term results of the management of paediatric hydrocephalus in the neurosurgery departments of the Conakry University Hospital and, above all, to identify and analyse the factors that could influence the vital, cognitive and functional prognosis of our patients.

2. Materials and Methods

We conducted a retrospective, multicentre, descriptive, and analytical study, the protocol of which was approved by the Medical Ethics Committee of the Neurosurgery Department of the Conakry University Hospital. This study involved a series of patients aged between 0 and 15 years; admitted and treated for paediatric hydrocephalus between January, 1st, 2012 and October, 31st, 2019 in the neurosurgery departments of the Conakry University Hospital. Only complete medical records of patients treated with post-operative follow-up were included in this study. Any incomplete records and records of patients who did not undergo surgical treatment were excluded.

Patient Selection: For each department, records were compiled using the local archiving system (manual and/or computerized, as appropriate). Furthermore, the neurosurgery department at Donka National Hospital underwent a relocation due to renovations to the hospital premises from 2015 to the present. This relocation likely prevented us from exhaustively collecting all medical records of patients treated for paediatric hydrocephalus in our departments during the study period.

Data: For each patient, a pre-established patient information sheet was created. We visited each centre to access the patients' medical records, consult the existing database (if available), and, if possible, analyse imaging when available. The medical record review allowed us to collect the following parameters: age, sex, place of origin, medical history, consanguinity, maternal age, clinical presentation, admission timing (from onset of symptoms to consultation), imaging results, indications, timing of surgery (diagnosis to surgery), surgical technique, complications, deaths, and survival time (from discharge to last consultation). We know that when children are young, the assessment of developmental progression is done in a global way, still little differentiated since the differentiation of functions is progressive. The development of the child being progressive, we used a scale which allowed us to assess the evolution of the child under several aspects (motor skills, communication, cognitive and affective processes, etc.). This was the Denver developmental screening test II (DDST II) this tool scoring which allowed the assessment of the prognosis to screen children from birth to age 6 years. It was considered unfavourable, when all the points obtained. After 6 weeks of follow-up, the Denver score was < 90% of the expected Denver score for a child of the same age not

treated for hydrocephalus.

Statistical analysis was performed using SPSS version 21 software. It covered the entire series as well as specifically non-communicating hydrocephalus. A descriptive analysis was performed (mean ± standard deviations and/or medians for quantitative variables and percentages for qualitative variables). For univariate analysis, the Chi-square test was used to compare two qualitative variables. The alpha risk was set at 5% ($p < 0.05$) for all statistical analyses. For the analysis of overall survival with or without recurrence, it was performed by the Kaplan-Meier method and the Logrank (Mantel-Cox) test. The median survival (plus or minus standard deviation) was evaluated. A multivariate analysis was also performed using a descending stepwise Cox model to search for prognostic factors. The factors studied were: Age (less than 02 years), Type of hydrocephalus (tri-ventricular), Presence of another associated malformation, Age of the mother, Presence of convulsive seizures, Presence of consciousness disorders, Time to treatment, Derivation technique, Number of surgical revisions, Occurrence of associated complications, and Mortality.

3. Results

During the study, we collected 317 consecutive cases, of which 150 patients were excluded, including 15 unoperated, 92 empty files, 15 files not found, and 28 incompletes. Our study sample was 167 patients.

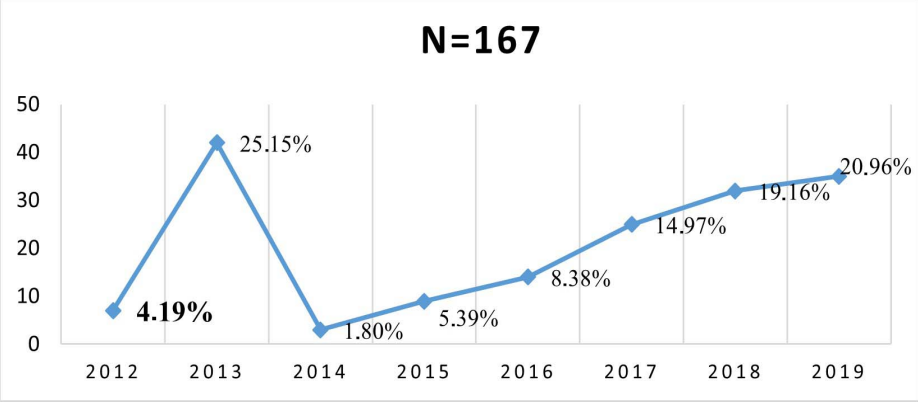


Figure 1. Illustrates the annual rate of our patients.

The annual rate frequency was 21 cases/year (**Figure 1**).

The average age was 12.0158 months ± 23.0889; the median age was 4 months with extremes of 1 day and 15 years (**Figure 2**).

A slight male predominance with a sex ratio of 1.17. Only 5/167 (2.99%) of our patients consulted within 72 hours of the onset of the first symptoms, with the timing of consultation of 2.8484 ± 3.8036 months (**Table 1**).

The clinical presentation was dominated by progressive macrocrania, which was found in 95.81% of patients, followed by bulging fontanelle (92.81%), axial hypotonia (92.22%), dilated veins (85.03%), and sunset gaze (74.25%) (**Figure 3**).

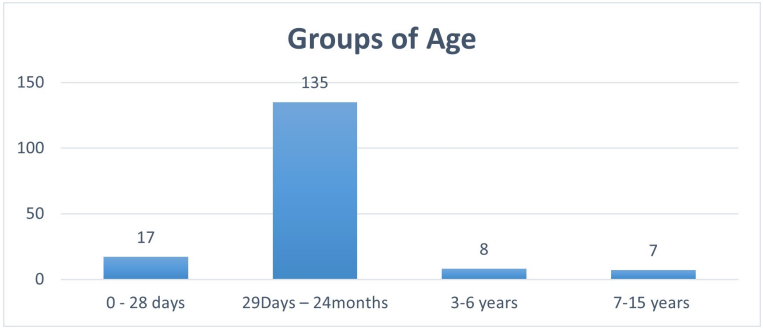


Figure 2. Groups of age repartition.

Table 1. Descriptive analysis of Socio-demographic characteristics of our patients.

Features	Frequencies	Percentages
Sex		
Male	90	53.89%
Female	77	46.11%
Inbreeding		
yes	22	13.17%
No	145	86.83%
Pregnancy monitored		
yes	18	10.78%
No	149	89.78%
Premature birth		
yes	4	2.40%
No	163	97.60%

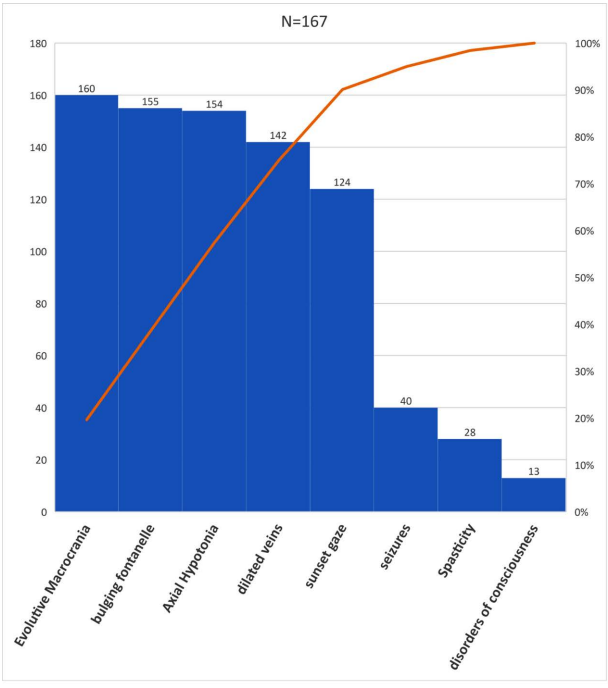


Figure 3. Clinical presentation of our patients.

Table 2. Descriptive analysis of Typology and aetiologies.

Parameters	Workforce	Percentages
anatomical type		
communicative	70	47%
Non-communicating	97	53%
Aetiologies		
Congenital without myelomeningocele	57	34.11%
Congenital with myelomeningocele	23	13.78%
Post infectious	72	43.11%
Post haemorrhagic	7	4.2%
Tumoral	8	4.8%

Congenital aetiologies. We treated 128/167 patients within more than 72 hours, for a mean of 13.3832 ± 31.8874 days, with a median of 7 days and a range of 1 day to 3 months (Table 2).

Table 3. Descriptive analysis of surgical techniques and timing of surgery.

	Workforce	Percentages
Support technique		
VP Shunt	92	55.09%
ETV	56	33.53%
ETV + VP Shunt	11	6.59%
EVD	8	4.79%
Response time		
≤24 hrs	07	4.19%
48 hrs - 72 hrs	32	19.16%
>72 hrs	128	76.65%

The most performed surgical technique was the VP shunt (55.09%) and the timing of surgery was 13.38 ± 31.88 days (Table 3).

The most common germs were Enterobacter cloacae in 2cases and Meningococcus with the same rate (Figure 4).

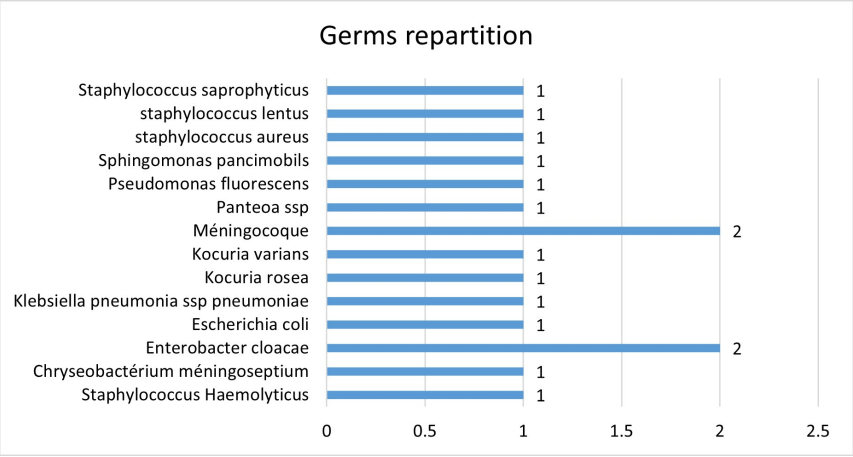
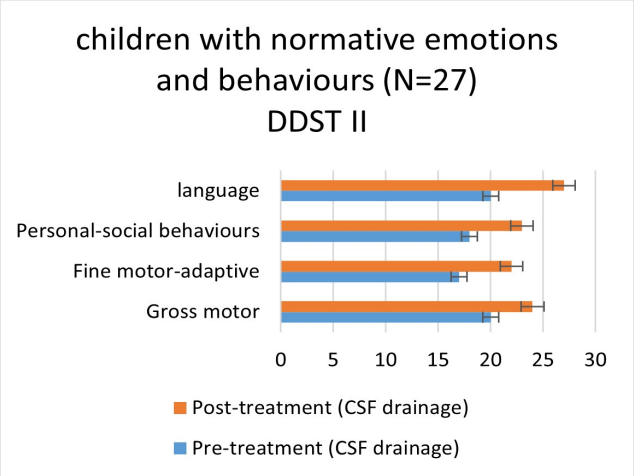


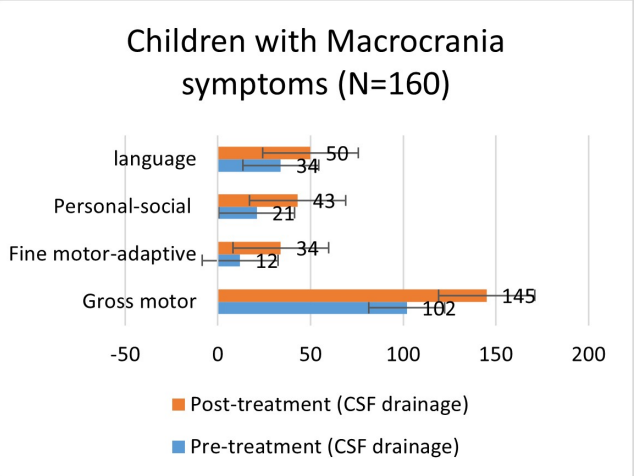
Figure 4. Infectious germs repartition.

Table 4. Descriptive analysis of post-operative complications of our patients.

Complications	Number	Percentage
Mechanical		**39 (23.34%)**
Obstruction	24	
Migration	03	
Hyper drainage/slit ventricle syndrome	09	
Rupture	04	
Infectious		**14 (08.38%)**
Shunt Infections	8	
Site Infections	14	
Haemorrhagic		**2 (01.20%)**
Haemorrhages	02	
Abdominal Issues		**4 (02.40%)**
Perforations	02	
Peritoneal pseudo-cyst	01	
Intestinal occlusion	01	
Others complications		**17 (10.18%)**
CSF leak	05	
Cognitive	04	
Post-operative seizures	02	
hormonal's disorders	01	
Hygroma	04	
Mesencephalic aqueduct Syndrome	02	
Inefficacity of ETV opening holes	09	
Subgaleal Collection	02	
Malposition	08	



(A)



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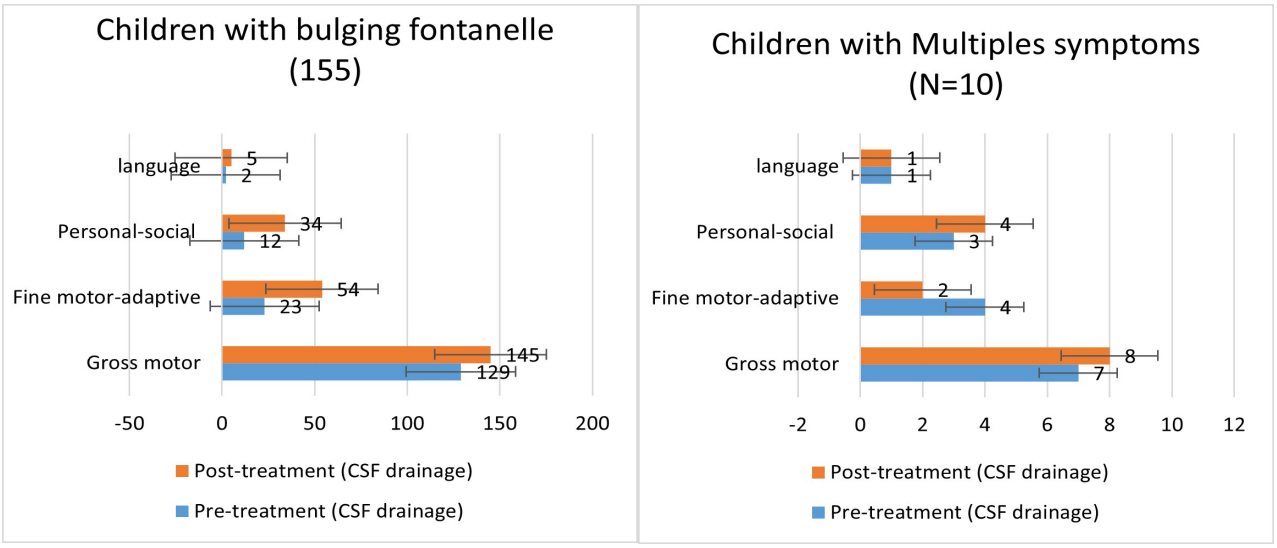

(C) (D)

Figure 5. Differential Diagrams of response of treatment showing changes in Denver developmental screening test II (DDST II) subscale scores with the subgroups' symptoms with paired samples CSF drainage pre and post treatment. significant changes observed for all children underwent CSF drainage and best improvement was found to children with normative emotions and behaviors (Denver Scores improved).

The majority of our patients were infants whose Denver development screening test was used to assess their summary of subscale scores (Figure 5). The univariate analysis of the series allowed us to have a statistical difference in survival according to the delivery voice. We found a significant difference with a median survival of 4.347 months ($p \leq 0.005$) (Figure 6).

Aqueduct stenosis was an etiology found in 42/167 with death as a censored variable, the statistical difference for survival according to etiology (aqueduct stenosis) was significant with a median survival of 3.996 months ($p \leq 0.005$) (Figure 7).

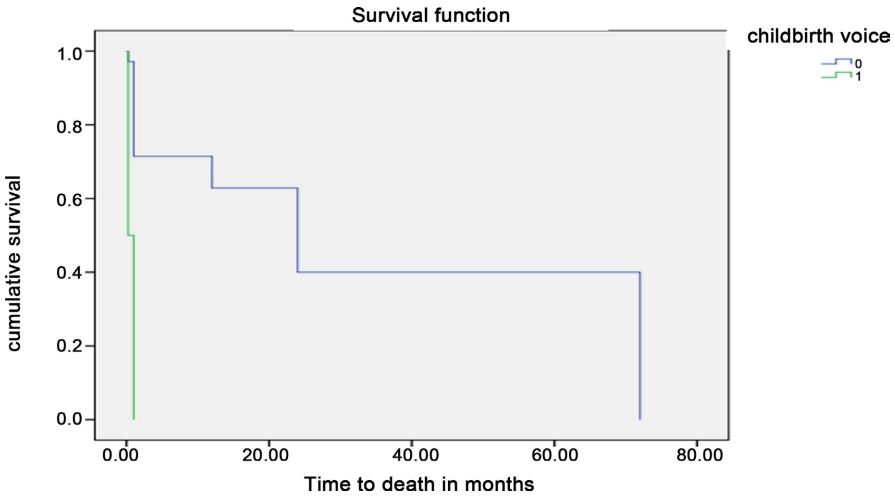

Figure 6. Survival function according to the route of delivery. $X^2 = 8.500$ (ddl = 1) P-value = 0.004.

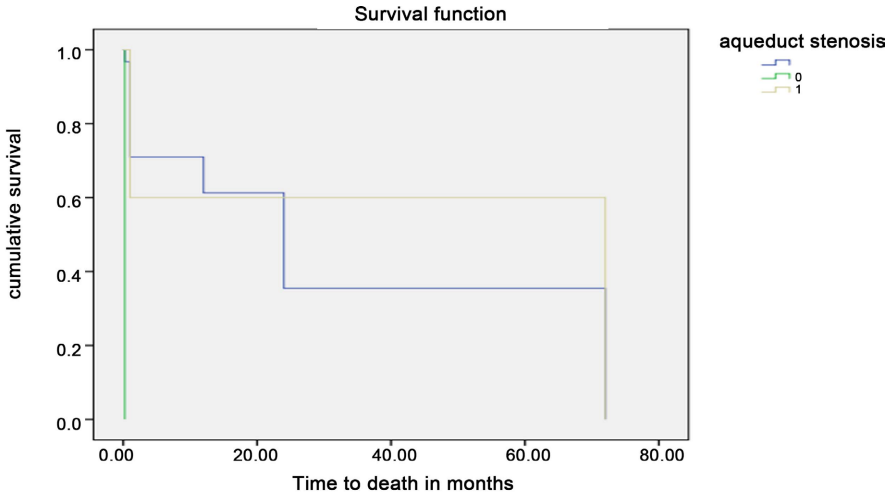

Figure 7. Survival function as a function of aetiology (aqueduct stenosis). $X^2 = 18.007$ (ddl = 2) P-value = 0.025.

The mechanical complications were the main cause of the shunt dysfunction found in 39 (23.34%) cases (**Figures 8-10**) and infectious complications represented in 08.38% specifically germs we find in **Figure 4**. Other rare complications were found including sylvian aqueduct syndrome and global rostral midbrain dysfunction in 2 cases (01.20%) cases and abdominal issues also the haemorrhages. Abdominal complications intestinal occlusion, and Extrusion of Ventriculoperitoneal Shunt Catheter (**Figure 11(A)** and **Figure 11(B)**) significant changes were observed for all children who underwent CSF drainage (**Figure 5**). During our study we recorded 43.84% of deaths.

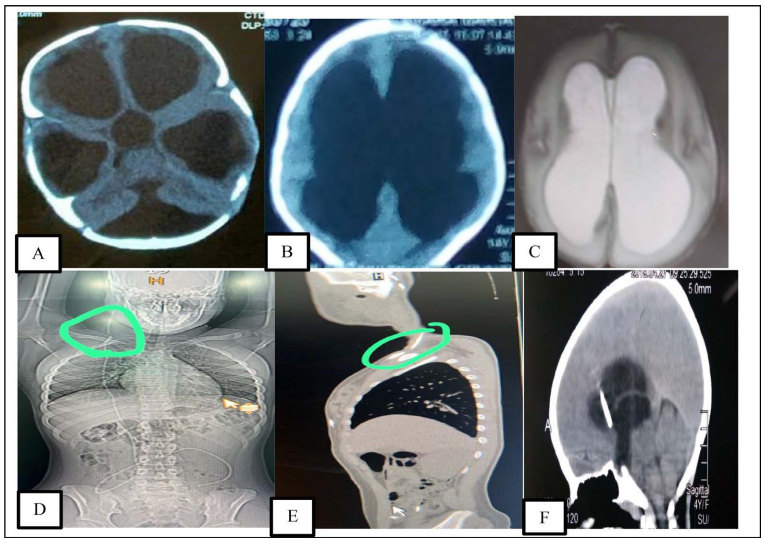

Figure 8. (A) Cranial CT show congenital hydrocephalus with Dandy walker malformation (B) Cranial CT scan ventricle dilatation (C) Brain MRI illustrated predominant occipital ventricle horn dilatation (D) Chest and Abdominal X-Ray illustrate rupture of VP shunt abdominal catheter at cervical level (E) CT scan ill-starred the same rupture (F) Sagittal CT scan reconstruction show the intracranial drain.

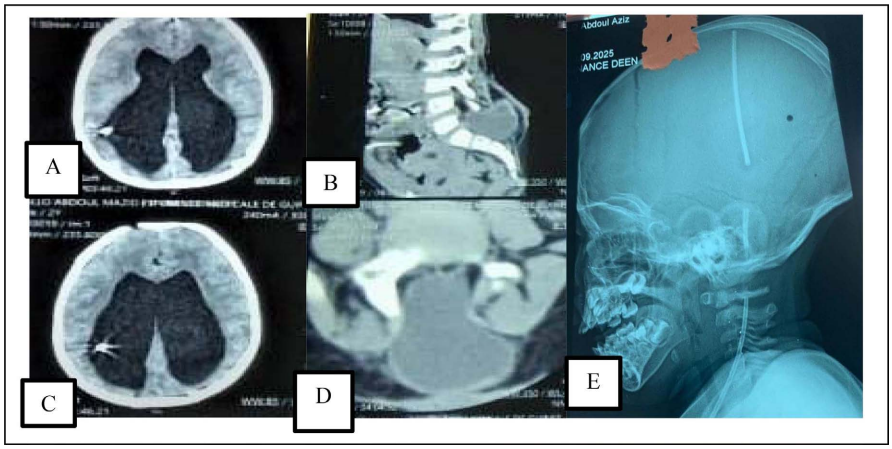

Figure 9. 6 years-old male child diagnosed congenital hydrocephalus with myelomeningocele L4 who underwent VP Shunt at early 3 days of life (A) and (C) predominant dilatation occipital horn ventricle, (B) sagittal reconstruction and (D) axial CT of lumbar CT scan illustrate a myelomeningocele.

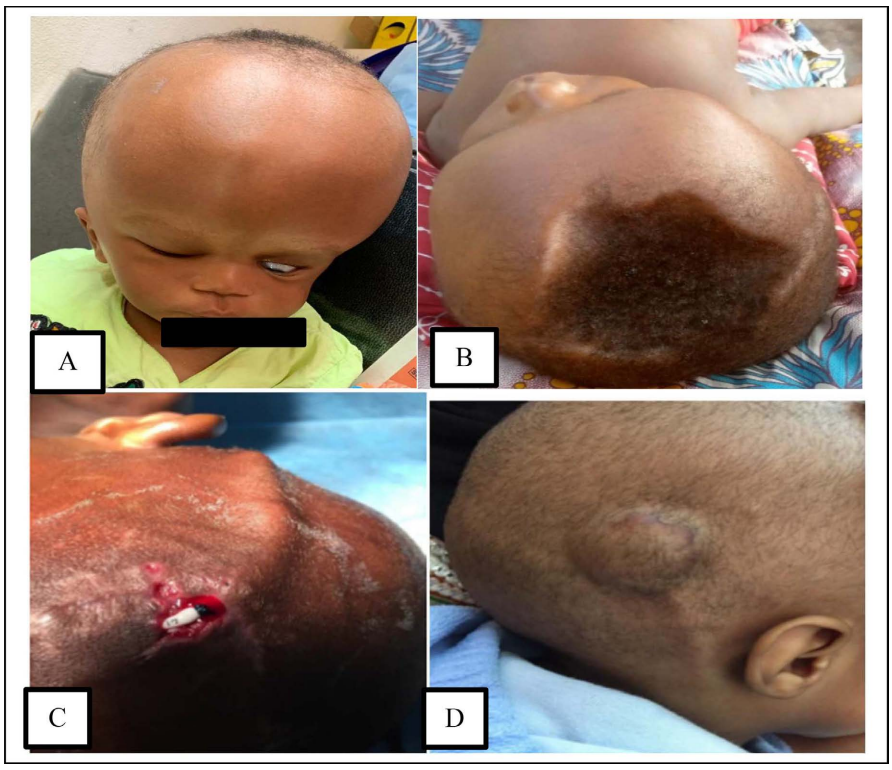

Figure 10. (A) Palpebral ptosis (B) anterior fontanelle depression with child surfing congenital hydrocephalus aqueduct stenosis who underwent right VP Shunt for treatment link to mechanical complication presented hyper drainage, (C) cranial Scalp ulceration (D) VP shunt Dysfunction.

Limitations of the study

Paediatric hydrocephalus is very common. In the scientific literature, studies reported on small series, often mixing different types of hydrocephalus and were homogeneous at all ages. Therefore, the analysis of prognostic factors in children

remains difficult. Our work therefore allows us to study the evolution of a large number of paediatric hydrocephalus cases, although it is retrospective, therefore faced the inability to carry out an exhaustive census of patients and a large number of patients were lost to follow-up.

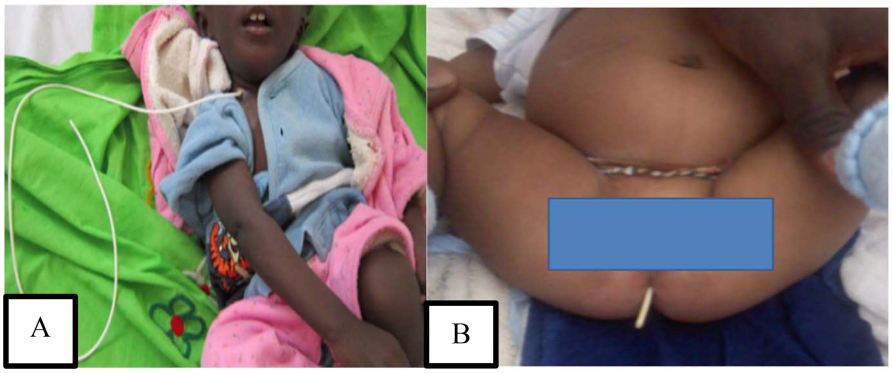

Figure 11. VP Shunt complications (A) exteriorization of abdominal drain (B) anal extrusion drain.

4. Discussion

The incidence of paediatric hydrocephalus is very difficult to establish, however, it is reported by Junior T. *et al.* in a study carried out in 2018 in Congo an average frequency of 7.7 cases/year [1], while in the USA, Sherman C. *et al.* [2] found in 1981 an incidence of congenital hydrocephalus of 66 cases per 100,000 live births with an association with myelomeningocele in 28 cases (16/100,000). The average age being 12.0158 months, infants were the most affected 135/167 cases (80.84%) with a median age of 4 months and extremes ranging from 1 day to 10 years. Philip M. *et al.* [3] Pierre A.B. *et al.* [4] reported an average age of 6.5 months respectively in Kenya in 2010 and in Italy in 2017. Our results can be justified by the delay in diagnosis highlighted by an average consultation time of 2.8484 months ± 3.8036 which could be explained on the one hand by the lack of awareness of the urgent nature of the pathology by the healthcare staff and on the other hand by the mystical connotation most often evoked in the African context. Fifty-three-point eighty-nine percent (53.89%) of the patients in our series were male, giving a sex ratio of 1.16. In 2016, a previous study conducted in Guinea by SOUARE I. *et al.* [5] found a sex ratio of 1.01, while a study conducted by Anne H. P. *et al.* [6] in Norway in 2015 also highlighted a male predominance with a sex ratio of 1.17. Our results correspond to those in the literature.

The clinical presentation of paediatric hydrocephalus varies according to age. The symptoms differ based on age. It is marked mainly in newborns and infants by progressive macrocrania which, according to the WHO, is defined as a head circumference greater than two standard deviations, *i.e.*, the 98th percentile; in young children, the intracranial hypertension syndrome is similar to that of adults. In 2012 in Togo, ADJENOU V. *et al.* [7] reported the presence of macrocrania in 40% of cases while Sergio F. S. *et al.* [8] in a study carried out in Mozam-

bique in 2014 found the presence of macrocrania in all patients in their series (100%). In Sweden, a study carried out by Persson E. K. *et al.* [9] in 2006 reported macrocrania in 46% of patients in their series. Our results are similar to data from the literature.

The diagnosis of paediatric hydrocephalus can sometimes be identified early with prenatal screening by ultrasound or MRI during pregnancy, others can be diagnosed after birth. A physician may suspect based on a child's rapidly expanding circumference's head neuroimaging MRI or CT of Brain can provides detailed views of the brain an d can confirm the diagnosis [10].

The aetiologies of paediatric hydrocephalus are diverse, multiple and mostly congenital. They are due to prenatal, perinatal and postnatal events. In our study, they were found in 84 cases/167 or 48.5% with an association with myelomeningocele in 14.37% of cases. In the scientific literature, the respective rates of 43.03%, 64% and 60% were reported by Junior T. *et al.* [1], Persson E. K. *et al.* [9] and Nalin G. *et al.* [10] respectively in Spain in 2005, in Sweden in 2006 and in the USA in 2007. Our results are lower than the data in the literature and could be explained by the absence of maternal-fetal monitoring (89.22% of pregnancies not monitored), the frequency of underdiagnosed forms (post-haemorrhagic forms frequent in premature babies due in 11% of cases to intracerebral haemorrhage [9], post-infectious forms due to germ infections such as treponema pallidum [11]; Lymphocytic Choriomeningitis Virus [12] neglected or poorly treated during pregnancy).

The management of paediatric hydrocephalus is urgent. First of all, the treatment aims to reduce the pressure on the brain. Multiples surgical technics provided this goal, CSF shunt a common surgical procedure that involves implanting a thin, flexible tube (shunt devise) to drain the excess CSF from the ventricles system to another part of body where it can be absorbed. The endoscopic Third Ventriculostomy (ETV) another surgical option where allowed created a small opening in the floor of a third ventricles to allow CSF to flow to posterior fossa cisterns. In our series, 128 patients/167 were treated within a period of more than 72 hours, *i.e.* an average of 13.3832 days ± 31.8874 with a median of 7 days and extremes of 1 day and 3 months. In Mali in 2012, the average time to treatment according to BA Momar *et al.* [13] was 3 months compared to a time of 138.5 days reported by Junior T. *et al.* [1] in Congo in 2018. Our results could be justified by the delay in parental agreement for surgery and the frequency of post-infectious cases.

Ventriculoperitoneal shunting was the most commonly used technique (55.09%) in our series. In Kenya and Turkey, it was performed at a rate of 93% and 97.1% according to Ali H. T. *et al.* [14] and Esther G. *et al.* [15] respectively in 2010 and 2019. According to Kouzo H. *et al.* [16] it was used in 91.9% of cases in a study carried out in Japan in 2007. Our results are consistent with the literature. The outcome depends on the early treatment and the aetiologies involved. In our study, it was favourable in 97.28% of cases. This rate was 82.4% in Congo in 2018 [1].

In our study, the average follow-up of patients was 10.2350 months ± 8.6417. This period was 36 months in Mozambique in 2014 [8] compared to 26 months found in the series of Schyamal B. *et al.* [17] in the USA in 2016. Our result could be explained by the loss of patient contact over time. Ventriculoperitoneal shunting (VP Shunt) being the most performed technique, its complications are mechanical and infectious. In our series, mechanical complications were the most frequent (36.53%). This rate was 20% in Kenya in 2010 [15], 80% in the USA [10] and 55.4% in Japan in 2007 [16]. During this study, valve obstruction was the main complication (32.93% of cases). In a study conducted in Kenya by Philip M. *et al.* [3] in 2010, valve obstruction was reported in 53.8% of cases. Our results could be explained by the proportion of lost to follow-up recorded in our series.

Endoscopic Third Ventriculostomy (ETV) is a minimally invasive surgical procedure to treat paediatric hydrocephalus by creating a opening hole in the third ventricle floor to bypass blockage in the CSF flow diversion [18]. It's particularly effective for obstructive hydrocephalus especially in children over six months of age or walking children [19]. The potential benefits of ETV such as lower infectious rate and better long-term quality of life compared to others technics CSF flow shunting. The success depends on patient selection and can be affected by factors like inflammations, infections and the specific aetiologies of hydrocephalus [20]-[22]. Others procedures external ventricle drainage (EVD) or subgaleal ventricle drainage (SGVD) the both allowed the temporary CSF drainage [23].

The postoperative complications of paediatric hydrocephalus are critical and many of them are rely to the shunt system dysfunction but among the mechanical complications related to shunting equipment, obstruction appears to be the most frequent, others complications like abdominal complications intestinal occlusion, Extrusion of Ventriculoperitoneal Shunt Catheter was observed in scientific literature and some very rare extrusion of peritoneal part of ventriculoperitoneal shunt complication was peroral extrusion [24]. The very rare complications like sylvian aqueduct syndrome and global rostral midbrain dysfunction associated with shunt malfunction often was described by some authors [25]-[27] and epilepsy [28]. This rate was 53.8% in Kenya in 2010 [3]. However, our results (**Table 4**) are lower than those of Anne H. *et al.* [6] in Norway in 2015 who reported an average of 3.3 revisions/patient.

The cognitive and behavioural outcomes for paediatric hydrocephalus are variable and may include motors dysfunction, personal social changing, intellectual efficiency and the language disability but outcome depended often on some factors like age of disease, aetiology, early management and the post-operative complications [29]. But some children may achieve normative emotions and behaviour or near-normative intelligence and can lead healthily live after treatment [24]. In our study the significant changes observed for all children who underwent CSF drainage and the best improvement was found to children with normative emotions and behaviours (Denver Scores improved). After all, some children can have difficulties with learning, concentration and social skill [30]. The particular-

ity of paediatric hydrocephalus lies in these multiple aetiologies but also and above all in its impact on the psychomotor development of the child. It is marked by physical handicap, the main motor sequela of hydrocephalus in children, with a variable frequency. Indeed, it was found in 20% of cases by Anne H. *et al.* [6] in Norway, in 39.3% of cases by Sérgio F. S. *et al.* [8]. The delay in diagnosis (average consultation time: 3 months), was the main cause of complications and poor prognosis. This chronicity compromises both the functional, sensory and intellectual prognosis. Thus, Susann A. *et al.* [31] found 83% of visual disorders in Sweden in 2006. As for the intellectual and cognitive prognosis, Persson E. K. *et al.* [9] reported 34% of intellectual delay in Sweden in 2006. Vichon v. *et al.* [32] mentioned a cognitive delay of 48% in France in 2012.

The mortality rate of hydrocephalus is relatively variable from one population to another. Mortality in paediatric hydrocephalus varies according to the series in the scientific literature; it is estimated at 18.1% in France in 2012 [20] and at 7.4% in Turkey in 2019 [14]. This could be explained in our context on the one hand by the delay in treatment marked by the large number of poor results found after a treatment delay of >72 hours without statistical correlation ($p > 0.05$).

5. Conclusion

Our study shows that age at diagnosis (<2 years), delay in treatment, and valve revisions (>3) are independent prognostic factors. Cerebrospinal fluid (CSF) drainage appears to be the only indicated treatment for hydrocephalus, particularly in infants < 2 years of age. It should be performed urgently and will improve prognosis when performed early. CSF appears to be an effective therapeutic weapon for non-communicating (obstructive) hydrocephalus. Its use in communicating hydrocephalus appears more controversial, and our results show a negative effect on overall child survival. The prognostic value of antenatal diagnosis of congenital aetiologies encourages fundamental research for early intrauterine therapeutic management. Studies are needed on the significance of certain factors, including the derivation technique and the mother's age.

Conflicts of Interest

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

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