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Original Article

Cure of Cephalocele in Niger: A Report of 33 Cases Over a Three-Year Period in Niamey

Le Traitement Chirurgical de la Céphalocèle au Niger : Bilan d'une Série de 33 Cas sur une Période de Trois Ans à Niamey

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Abbreviations
CSF: cerebrospinal fluid
CT: Computed Tomography
M/F: male/female
NTDs: Neural Tube Defects

ABSTRACT

Introduction. Neural tube defects (NTDs) are a public health burden in countries with limited resources, particularly in Niger. In this work, we focused on cephalocele which is an entity of NTDs. The aim was to describe epidemiological, therapeutical, and outcome of cephalocele at our neurosurgical center. **Methods**. This was a prospective study recruiting 33 patients suffering from encephalocele treated at the neurosurgical center in the National Hospital of Niamey. It was conducted over 03 years with an average follow-up of 6 months. ANOVA test was applied to appreciate good prognostic factors (CI 95%). **Results**. More cases of cephaloceles were reported in 2022. The sex ratio was M: F 0,73 and most of the mothers' age ranged from 20 to 30 years old (48%), with a consanguinity rate of 74%. 98% of cephaloceles were located on the skull vault. All patients benefited from a brain CT scan, depicting that 57% had meningoencephalocele. 28 patients were operated on with 18% post-surgical complications. 89% of our patients had a good evolution. The prognosis was associated with the aspect of skin lining (p=0.02), general status on admission (p=0.01), and malformation contents (p=0.03). **Conclusion**. The post-surgical prognosis of cephaloceles remains a challenge because of the burden on society. Generally, sensitization remains a necessary means of preventing NTDs. However, once it occurs, early consultation is important. Other prognostic factors mentioned in the literature were not significant in this series, showing the necessity of a study with a larger sample.

RÉSUMÉ

Introduction. Les anomalies du tube neural (ATN) constituent un fardeau pour la santé publique dans les pays aux ressources limitées, en particulier au Niger. Dans ce travail, nous nous sommes concentrés sur la céphalocèle qui est une entité des ATN. L'objectif était de décrire l'épidémiologie, la thérapeutique et les résultats de la céphalocèle dans notre centre neurochirurgical. **Méthodes**. Il s'agit d'une étude prospective recrutant 33 patients atteints d'encéphalocèle traités au centre neurochirurgical de l'hôpital national de Niamey. Elle s'est déroulée sur 03 ans avec un suivi moyen de 6 mois. Le test ANOVA a été appliqué pour apprécier les facteurs de bon pronostic (IC 95%). **Résultats**. Plus de cas de céphalocèle ont été rapportés en 2022. Le sex-ratio était M : F 0,73 et l'âge de la plupart des mères se situait entre 20 et 30 ans (48%), avec un taux de consanguinité de 74%. 98% des céphalocèles étaient situées sur la voûte du crâne. Tous les patients ont bénéficié d'une tomodensitométrie cérébrale, montrant que 57% d'entre eux avaient une méningo-encéphalocèle. 28 patients ont été opérés avec 18% de complications post-opératoires. 89% de nos patients ont eu une bonne évolution. Le pronostic était associé à l'aspect du revêtement cutané (p=0,02), à l'état général à l'admission (p=0,01), et au contenu de la malformation (p=0,03). **Conclusion**. Le pronostic post-chirurgical des céphalocèles reste un défi en raison du fardeau qu'il représente pour la société. D'une manière générale, la sensibilisation reste un moyen nécessaire de prévenir les ATN. Cependant, une fois qu'elle survient, il est important de consulter rapidement. D'autres facteurs pronostiques mentionnés dans la littérature n'étaient pas significatifs dans cette série, ce qui montre la nécessité d'une étude avec un échantillon plus important.

INTRODUCTION

Neural tube defects (NTDs) are congenital and originate during the central nervous system formation.
The incidence is estimated at 300,000 new cases per year with a prevalence of 18.8 for 10,000 live births. These data fluctuate according to the part of the world concerned [1]. Many parameters have been identified as being related to the occurrence of NTDs in sub-Saharan Africa. Be it preventable or non-preventable factors, once the defect occurs the prognosis is at stake, it might be functional or life-threatening. The epidemic notion mentioned in 1971 is therefore widely accepted. The NTDs represent a high public health burden, socially and economically weighing on families and governments. The latter is essentially due to the after-effects presented by the survivors [1–3]
Its multifactorial character permitted it to implement some prevention axes, like food fortification with folates, which

contributed to considerably reducing the incidence [3, 4]. But there is still a lot to do with non-preventable factors like genetics and environmental ones [4].
Compared to spina bifida, cephaloceles are less frequent and in Niger is represented with a ratio of 5.6/1 [5]. Apart from the scarcity of research on cephaloceles in Africa, the prevalence of cephaloceles is 0.02%[3]. As a distinct presentation of a neural tube defect, cephalocele represents a challenge during the phase of treatment according to his anatomic characteristics, requiring mastery to avoid complications related to the surgery procedures [3]
This study aimed to give an update of the epidemiology and therapeutic aspects of cephaloceles, to appreciate the prognosis.

MATERIALS AND METHODS

It was a descriptive prospective study covering 36 months (from January 1st, 2021, to December 30th, 2023), including all infants admitted for cephaloceles in the neurosurgery department of the National Hospital of Niamey. That length of the study period allowed us to have a sufficient sample that would permit the execution of statistical tests to give value to our research.
Patients admitted for other NTDs those presenting with a post-traumatic mass or tumoral mass were not included in this study.
A data collection form helped us to gather information from the infant's or newborn's parents, from the medical file and the consultation and operative room records.
Data were curated and analyzed using SPSS 16.0 and an ANOVA test was applied with a 95% confidence interval ($p<0,05$), allowing us to check the association between some parameters and the prognosis (Tables 1 and 2).
This study was approved by both ethics committees of the faculty of health sciences and the National Hospital of Niamey. Oral consent was required from the parents of the infants before anonymous data collection.
The principal handicap of this study was the follow-up of the patients especially because of the lack of telephone contact from some patients' carers.

RESULTS

Epidemiological data

During the study period, the hospital incidence of cephaloceles was 3.63% and 2022 had more admissions, with 14 cases within the year (Table 1). The sex ratio M: F was 0.73. 87.88% of cases were brought to consultation during the neonate's period. The patients' mothers are relatively young, with 48.48% aged between 20 and 30 years. There is a particularity to notice that mothers are more literate than fathers (16% of women against 6% of men). Despite the diagnosis of the cephalocele during prenatal care (87.87% between 1st and 6th prenatal visit), 94% of women were put to birth through normal delivery, among which 45% were septic deliveries. Consanguinity remains highly represented in our setting (73%).

Table 1. Overview of the socio-epidemiologic parameters of cephaloceles.

Parameters	N (%)	p-value[a]
Hospital Incidence	**33/909 (3.63)**	-
2021	12/333 (3.60)	
2022	**14/297 (4.71)**	
2023	7/279 (2.50)	
Annual admission		-
2021	12 (36.36)	
2022	**14 (42.42)**	
2023	7 (21.22)	
Sex ratio	0.3	-
M	14 (42)	
F	**19 (58)**	
Infants' age		0,447
0-29 jours	**29 (87.88)**	
1-24 mois	3 (9.09)	
2-4 ans	1 (3.03)	
Mother's age		
< 20 YOb	5 (15.15)	
20-30 YO	**16 (48.48)**	
31-41 YO	4 (12.12)	
> 41 YO	8 (24.4)	
Illiteracy		-
Men	**31 (94)**	
Women	27 (84)	
Pregnancy		-
Primigeste	7 (21.21)	
Paucigeste	16 (48.48)	
Multigeste	**5 (15.15)**	
Great multigeste	**5 (15.15)**	
Parity		-
Primipara	6 (18.18)	
Paucipara	17 (51.51)	
Multipara	**5 (15.15)**	
Great multipara	**5 (15.15)**	
Prenatal visits		-
0	2 (6.6)	
1-3	14 (42.42)	
4-6	15 (45.45)	
7-9	2 (6.6)	
Consanguinity	**24 (73)**	-
Delivery mode		-
Normal delivery	31 (94)	
Cesarian section	2 (6)	
Septic deliveries	**15 (45)**	0.243

[a] p-value evaluating parameters with correlation with the patients' evolvement; [b] YO = years old

Clinical data, management, and outcome (Table 2)

The major location (Figure 2) of the tumefaction is the cranial vault (98%) and 57.14% of them are meningoencephaloceles (Table 2). We noticed that associated malformations were present in 4 infants. All the patients benefited from a brain CT scan before surgery, with or without a spine CT scan.

Table 2. Overview of clinical, therapeutic, and evolutive aspects

Clinical aspects	N (%)	p-value[a]
Tumefaction		0.43
Opened	4 (12)	
Closed	**29 (88)**	
Skin lining		**0.028**
Healthy skin	**24 (73)**	
Poorly epidermized	9 (27)	
General status		**0.013**
Altered	2 (6)	
Preserved	**31 (94)**	
Malformations' location		0.55
Skull vault	**32 (98)**	
Skull base	1 (2)	
Associated malformation		-
Yes	**4 (12.12)**	
No	29 (87.88)	
Cephaloceles' contents		**0.024**
Meningoencephalocele	**16 (57.14)**	
Encephalocele	4 (14.28)	
Meningocele	6 (21.42)	
Unprecised	4 (14.28)	
Therapeutic		-
operated	28 (85)	
Non operated	5 (15)	
Immediate complications		-
Yes	**5 (18)**	
No	23 (82)	

[a] p-value showing the correlation between the parameters and the patient's clinical evolution; [b] post-surgical follow-up over 7 days.

85% were operated upon, following classical steps (Figures 3 and 4). The cure of the cephalocele was associated in some cases with the cure of other malformations (Figure 1).

Complications are considerable (18%) and are essentially made up of post-surgical infection, losing stitches, CSF leaks, hydrocephaly, and on a further note, death (10.71%).

DISCUSSION

Socio-epidemiology

Cephalocele is an entity of neural tube defects common in Niger [5–7]. The socio-economic context makes it an epidemic. Nowadays the global incidence in the general population is difficult to evaluate. That is why a strategy is being put in place to perform a study at a sub-regional level to appreciate the actual societal burden [1].

At the hospital level, cephalocele represents 3.33% of admitted patients in our neurosurgery department. Hospital incidence fluctuates according to the geographic region. At Zinder, in Niger, it represents a mean of 14.66 cases per year [7], slightly above Niamey (11 cases per year). This incidence in Niger is relatively higher than the one in developed countries because of preventive measures applied there; notably genetic counseling, therapeutic abortion, and folate food fortification. [1, 4, 6, 8]

The female predominance observed (sex ratio 0.73) is still maintained compared to previous studies performed in Niger [6] and other studies in the literature [9–11]. However, some authors are reporting an equilibrium between males and females [5] or a male predominance [7, 12]. This doesn't allow us to establish gender determinism.

The patients in Niger consult relatively earlier [6, 7], even though the average age during the consultation remains higher compared with the one in Maghreb and Western countries [10], despite the care policy for infants aged between 0 and 5 years put in place in Niger. This discrepancy is certainly favored by universal health coverage and health insurance policy which are performing well in those countries. We highlight that we did not have patients above 5 years. Some authors in the literature have registered children, teenagers, and adults [2, 9, 11, 13–15].

Consanguinity, multiparity, young aged or older aged mothers, identified as predisposing factors in the literature [4, 5, 7, 11] were found, but without correlation with the occurrence of cephaloceles in our study. Prenatal consultation which is a keynote for the screening of the malformation [16] depends on the literacy level of the mother ($p = 0.03$). This brings up the importance of community sensitization, particularly to women of childbearing age, on sexuality and the importance of prenatal follow-up.

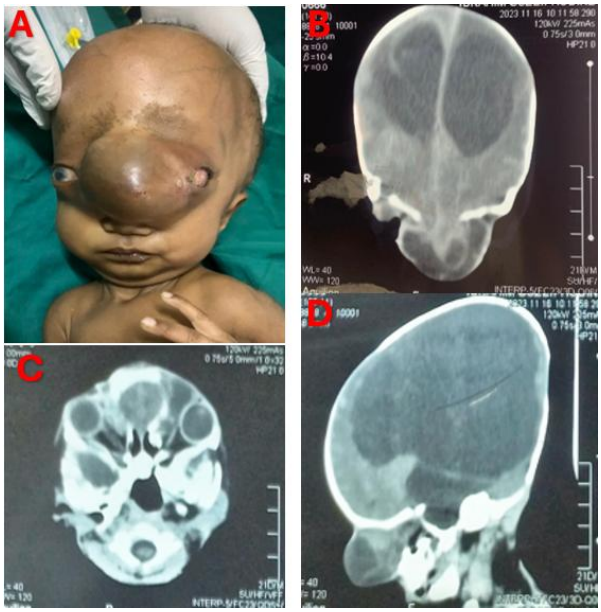

Figure 1. Clinical presentation and brain CT scan of an anterior cephalocele. (A) naso-orbital tumefaction with hypertelorism; (B) axial view of brain CT scan showing herniation of brain tissue through frontal bones; (C) axial view of the cranial base showing a tissue process through ethmoidal bone and in both orbital cavities; (D) left para sagittal reconstruction through the orbital cavity showing an intra-orbital process with mixed content and ventricular dilation.

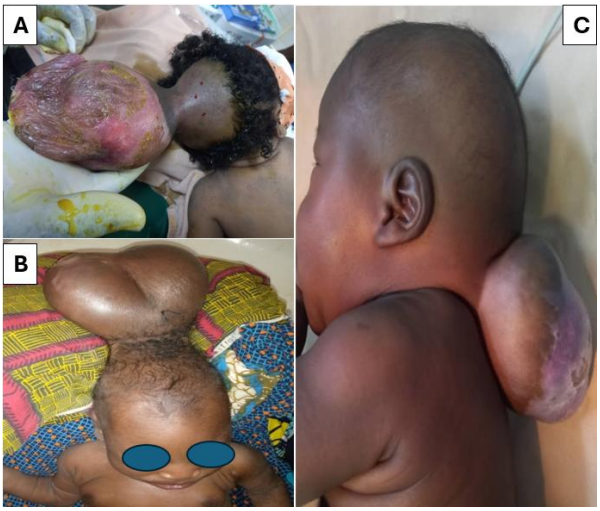

Figure 2. Some clinical presentations of cephaloceles. (A) parietal cephalocele with poor skin lining, skin break, and CSF leak indicating an urgent repair. (B) parietal cephalocele with a good skin lining. (C) occipital cephalocele partly covered with a good skin lining.

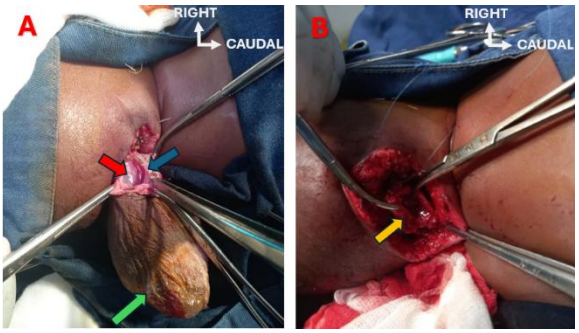

Figure 3. Cure of occipital cephalocele. (A) Resection of the sack: skin lining poorly epidermized (green arrow), the opening of the dura mater (blue arrow), and exploration of the mass content comprising meninges and CSF (red arrow); (B) after resection of the exceeding tissue, closure of the dura mater edges (orange arrow) with non-absorbable suture (polypropylene 4/0).

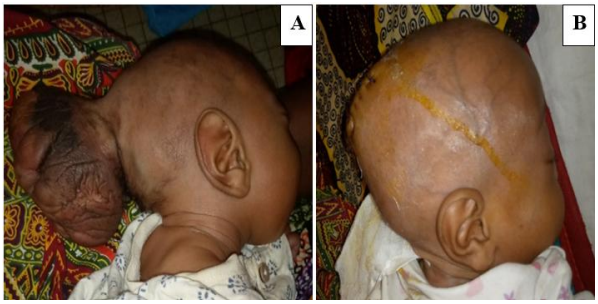

Figure 4*. Surgical repair of a parietal cephalocele. (A) Before surgery; (B) After surgery.*

Clinical presentation

94% of our patients admitted with a good general health status are due to early consultation because of the care policy for children between 0 and 5 years which is free of charge.

98% of malformations are located on the cranial vault, 87.5% being occipital. The latter represents the preferred location in the literature [2, 5, 7, 10–12] and might be associated with a poor prognosis. In this series, the correlation is not significant between the location and immediate post-surgical prognosis. The skin lining aspect and or CSF leak in cephalocele is a decisive criterion for urgent surgical care of the malformation. *Yucetas et Uçler* had 44% of patients with skin fissures with CSF leak [10], 11.38% for *Sani et al* at Zinder [7], and 4.3% for *Juan F. Martinez-Lage et al* [11].

The brain CT scan performed for all our patients had the advantage of showing more details on the bone defect location and is also paramount for the surgery planification and bone defect repair. It also helps to an extent, to appreciate the content of the malformation. The MRI, which is more precise to appreciate the content, was out of reach because of the unavailability of an anesthesiologist to sedate infants during the procedure.

Based on the brain CT scan information, meningoencephalocele was the most represented anatomo-clinic entity (57.14%). In this series, there is a significant correlation between the nature of the mass content and the global evolution of the patient ($p=0.024$).

Surgical care

The average hospitalization duration before the surgery was 13 days, this is closer to *Sani et al* at Zinder who found 12 days [7]. The surgical care of the malformation located at the vault is a classic procedure. For our lone case of skull base cephalocele, we proceeded with a conventional surgery through a bi-coronal classical approach in the absence of a multidisciplinary team like *S. Bakhti et al* in Morocco [9]. Classic surgery care of sincipital cephaloceles gradually gives way to endoscopy [14]. The care of cephaloceles in our series was done with associated malformations (hydrocephalus, spina bifida) during the same surgical period. Thus, we had a ventriculoperitoneal shunt (VP shunt) or cure of spina bifida associated with the repair of the cephalocele. All our patients benefited after surgery of analgesic therapy and antibiotic coverage due to our bacteriologic environment. Some authors also performed concomitant surgeries during the cure of the cephalocele, especially the VP shunt.[12]

Post-surgical evolution

The evolution after surgery in our series is not free from complications. We had one case for each complication encountered namely hydrocephalus, loose stitches, CSF leak, suppurative wound, and death. For the factors influencing the prognosis, it differ according to studies *Juan et al* found no correlation between mortality and the size nor the content of the mass [11]. According to *Kabre et al,* the prognosis is linked with the occurrence of post-surgical complications like hydrocephalus or wound infection and also with the presence of another malformation [17]. *Sanoussi et al,* in 2009, found that the purity of cephalocele, the size of the mass, the association with hydrocephalus, and the age less than 15 days were strongly correlated with the mortality [6]. In this series, the general health status of the infant at admission, the skin lining of the mass, and the content of the mass are statistically significant for the prognosis. (Table 2).

CONCLUSION

Neural tube defects, amongst which cephaloceles, remain a great challenge in our context; looking at the numerous predisposing factors encountered. The care of cephalocele on the technical note is no longer an issue but the prognosis is highly correlated to the age of the infant on admission. Some efforts are still necessary as far as prevention is concerned, with a view to eradicating this malformation whose consequences are heavy for the patient, the family, and society, because of the risk of sequelae and neurologic handicaps depending on the case presentation. Complications being related to clinical presentation are also shown to be related to the skin lining and the content of the mass. This will allow us to carry out some studies to define the insights into those relations found on the complication's venue.

DECLARATIONS

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Assoumane Issa Ibrahim: Conceptualization, Methodology, Writing – review & editing, Validation
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Kelani Aminath: Supervision
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Conflicts of Interest
The authors declare no conflicts of interest.

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