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Actual epidemiological profile and management of central nervous system (CNS) malformations in a neurosurgery department in sub-Saharan Africa

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Introduction: Central Nervous System (CNS) malformations usually leave the family in disarray and doctors' staff powerless. In developed countries, the rigorous application of preventive measures has contributed to a significant reduction incidence of these malformations. In African societies, mystical-religious considerations and low socio-economic level contribute to the increased occurrence of these malformations. The purpose of this work was to evaluate the prevalence and management of CNS malformations in our neurosurgical department.

Methods: This was a retrospective study conducted in the neurosurgery department of Yalgado Ouedraogo Teaching Hospital, in Ouagadougou, from 2016 to 2019. All cases of CNS malformations managed in the department with a workable medical record were reviewed.

Results: Five hundred and twenty-four cases were selected. The prevalence of malformations was 7.05%. The mean age of patients was 1.48 years (1 day and 76 years). Thirty-one (5.9%) cases of CNS malformations were diagnosed antenatally. One hundred and twenty-four (23.6%) ultrasounds (including 51 antenatal), 418 CT scans (79.7%) and 9 MRIs (1.7%) were performed. Hydrocephalus [(n = 219 (41.7%)] and spina bifida [(n = 213 (40.6%)] were the most common pathologies. Surgical treatment was indicated in 435 cases (83%) of which 179 (41.1%) could be operated. After a follow-up of 15.8 months, there were 46.3% sequelae identified.

Conclusion: CNS malformations are common in our context. They are not always prone to be treated surgically and are at the origin of many neurological sequelae and lifelong disability. Hence there is an urgent need to focus on improving preventive measures in Sub-Saharan Africa.

Keywords: Malformations, CNS, spina bifida, hydrocephalus, encephalocele, AFTN

INTRODUCTION

The term "congenital CNS malformation" defines any abnormality in the shape or structure of the central nervous system, present at birth, due to abnormalities of the CNS development, occurring at any time in intrauterine life, regardless of its etiology. Anatomically, a distinction is made between neurulation abnormalities (anterior and/or posterior neurospora closure anomaly) and post-neurulation abnormalities (neurax and/or its container anomalies). These CNS malformations are rarely isolated [1].

The birth of a malformed child most often leaves the family in disarray, on the one hand, and the caregiver powerless on the other. In some circumstances, these are serious malformations that are sometimes incompatible with life or require complex treatment with disappointing results [2].

In developed countries, advances in antenatal diagnosis (amniocentesis, ultrasound, fetal MRI), genetic counseling and the rigorous application of preventive measures have contributed to a significant decrease in the incidence of these malformations [3]. In African societies, mystical-religious considerations, and low socio-economic level contribute to an increase in the occurrence of CNS congenital malformations in the population [4].

The purpose of this work was to evaluate the prevalence and management of CNS congenital malformations in our neurosurgical department.

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METHODS

This was a cross-sectional and descriptive study with retrospective collection of data in the neurosurgery department of Yalgado Ouedraogo University Hospital Center in Ouagadougou, from January 1st, 2016 to December 31st, 2019.

Study population consisted of records of patients admitted for outpatient evaluation or hospitalization during this period. All cases of CNS congenital malformation evaluated in our department with available medical records, i.e., containing at least the socio-demographic and clinical information of the patient, were included in the study.

RESULTS

Socio-demographic data

We recorded 8,098 admissions in the department, including 571 cases (7.0%) of CNS malformations, with 47 non-usable files. Five hundred and twenty-four (524) medical records were selected for this study. Figure 1 shows the flow diagram of CNS malformations and their sites.

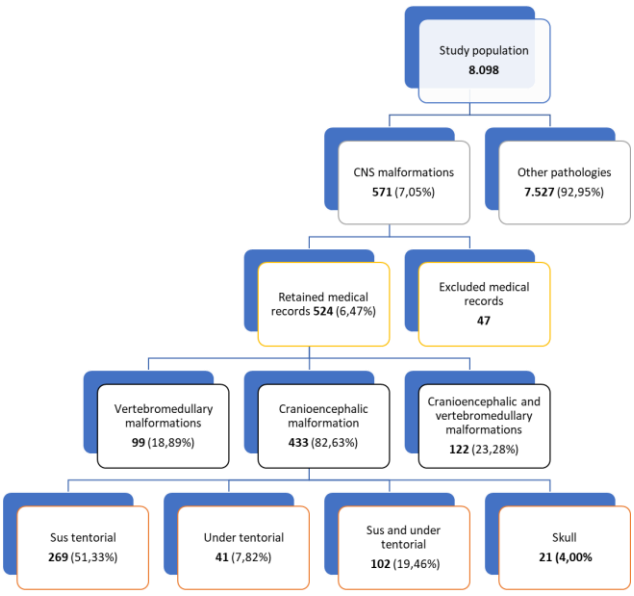

Figure 1- Flow diagram of CNS malformations and their sites

The mean age of the patients was 1.4 years with extremes of 1 day and 76 years. There were 317 infants (60.5%) among the patients. Four hundred and ninety-nine (499) cases (95.2%) of patients were less than 5 years of age. The distribution of patients by age group is shown in Table 1.

Table 1- Distribution of patients by age group

Age group	n	%
Newborn: 0 to 28 days	152	29,01
Infant: 28 days to 2 years	317	60,50
Early childhood: 2 to 6 years	32	6,11
Second childhood: 6 to 12 years old	9	1,72
Teenager: 12 to 18 years old	5	0,95
Adult: 18 to 65 years old	8	1,52
Senior: over 65	1	0,19
Total	**524**	**100**

There were 259 male patients (49.4%) and 264 female patients (50.4%). The sex was not reported in 01 patients (0.2%).

Two hundred and sixty-seven (267) cases (51%) resided in urban areas and 201 cases (38.3%) in rural areas. For 56 cases (10.6%), the provenance was not provided on the medical record.

Diagnostic Data

Thirty-one (31) cases of CNS malformations were diagnosed antenatally on average at the 33rd gestational week (range 3rd – 18th week). The mean time to admission was 8.3 months, ranging from 34 weeks (antenatal diagnosis) to 23 years of age.

The reason for consultation was spine congenital swelling (219 cases or 41.8%), macrocrania (200 cases or 38.2%), psychomotor retardation (126 cases or 24%), cephalic congenital swelling (63 cases or 12%), polymalformative syndrome (cephalic and spinal malformation, in 55 cases 10,5%), comitiality (29 cases or 5.5%), microcrania (15 cases or 2.9%), intracranial hypertension syndrome (13 cases or 2.5%) and torticollis (1 case or 0.2%). Macrocrania was associated with spinal swelling in 43 cases (8.2%).

On physical examination, among the 63 cases of congenital cephalic swelling, the swelling was localized in occipital in 43 cases (68.3%), naso-ethmoidal in 8 cases (12.7%), frontal in 8 cases. (12.7%), naso orbital in 3 cases (4.8%) and parietal in 1 case (1.7%). Regarding the 219 cases of the spine congenital swelling, there were 215 cases (98.2%) of spina bifida (including 198 cases localized in lumbar or lumbosacral, 14 cases in dorsolumbar and 3 cases in cervical) and 4 cases (1, 8%) of scoliosis (including 2 cases in lumbar, 1 case in lumbosacral and 1 case cervical). Among the 215 spina bifida, there were 172 cases (80%) of myelomeningocele and 43 cases (20%) of meningocele. In the 172 cases of myelomeningocele, paraplegia and sphincter disorders were noted in 121 cases (70.3%) and paraplegia/paraparesis without sphincter disorders in 51 cases (29.7%) The malformations or deformations associated with these various malformations of the CNS were clubfoot in 52 (9.9%), cleft lip and palate in 2 cases (0.4%); Crouzon

syndrome, Trisomy of chromosome 21, bent knee and genu valgus in 1 case (0.2%) each. Figure 2 presents some malformations in our series.

Figure 2- Photos of some malformations. (a) macrocrânie (hydrocephalus); (b) exencephaly + microcrania; (c) Crouzon disease; (d) spina bifida (myelomeningocele) ;(e) occipital encephalocele; (f) sincipial encephalocele (ethmoid naso)

One hundred and twenty-four (124 or 23.7%) ultrasounds (including 51 antenatal and 73 postnatal), 418 (79.8%) CT and 9 (1.7%) MRI (including 7 after CT) were performed. Among the 73 postnatal ultrasounds, there were 54 cases (74%) of transfontanellar ultrasounds, 11 cases (15.1%) of cardiac ultrasounds, 5 cases (6.8%) of abdominopelvic ultrasounds and 3 cases (4.1%) of the swelling ultrasound. In 53 cases (72.6%) postnatal ultrasound was not sufficient for diagnosis of the CNS malformation and was completed by CT. In the 20 other cases, it concluded to the CNS malformation diagnosis. In 20 cases (39.2%) out of 51, antenatal ultrasound had not concluded that there was a CNS malformation.

Therapeutic and Evolutive Data

There was a surgical indication in 435 cases (83%) of which 179 (41.1%) could be operated on and 256 (58.8%) were not operated. A surgical indication was not retained in 89 cases (17%). Patients with associated malformations outside CNS were referred to specialty departments indicated after CNS malformation surgery in neurosurgery. Physiotherapy was prescribed in all our patients with neurological limb deficit (73 cases).

Of the 179 patients who were able to undergo surgery, the average time between surgical indication and surgery was 31.2 days with extremes of 1 day and 8 years. Two hundred and twenty-eight (228) pathologies were operated including 115 hydrocephalus, 80 spina bifidas, 27 encephalocele, 4 compressive arachnoid cysts and 2 Arnold Chiari malformations. Of the 256 cases (58.9%) with a surgical indication who could not be operated; 231 cases (90.2%) lost follow-up. There were 22 cases (craniostenosis, polymalformative syndrome) in which it was more of a surgical abstention for insufficiency of the technical platform and 3 cases died before surgery. The summary of the diagnoses and therapeutic attitudes is shown in Table 2.

The average length of hospital stay for operated patients was 4.3 days, ranging from 1 to 25 days. At the end of this period, evolution was favorable in 45 cases (25.1%), stationary in 119 cases (66.4%), unfavorable in 14 cases (7.8%). There was 1 death (0.5%). In the long term, i.e., after an average postoperative duration evolution of 15.8 months, evolution was favorable without sequelae in 85 cases (47.5%), with slightly disabling sequelae in 75 cases (41.9%); with very disabling sequelae in 8 cases (4.5%). There were 9 lost to follow-up (5%) and 2 deaths (1.2%).

DISCUSSION

Prevalence and socio-demographic aspects

The annual prevalence of CNS malformations in our study (6.4%) was superimposed on that of other authors (5%) [5]. This prevalence noted in our series was much lower than some noted in the literature: 13.5% [4]; 31.8% [6]. These variations could be linked, on the one hand, to the level of the technical platform for the diagnosis of these malformations and, on the other hand, by the selection criteria of these studies. The mean age of diagnosis of CNS malformations was 17.6 months with extremes of 0 and 10 years [7]. This age is significantly higher than ours (8.21 months). The sex ratio also varies between studies. It was 0.86 [8] and 0.85 [4] for some authors. Others noted a sex ratio of 1.1 [7] and 1.54 [6]. Sex had no effect on the malformation [7]. Half of the patients in our study were from urban areas (51%). This is usually the temporary residence of patients who have come from rural areas for treatment in urban areas.

Diagnostic Aspects

Spine congenital swelling was the most noted reason for admission in our series (41.8%). This observation was also made by other authors with 68.1% of cases [1]. The association of macrocrania and spine congenital swelling in our series accounted for 8.2% of the reasons for admission. Other authors [1] noted it in 13.88%. This combination should suggest hydrocephalus associated with spina bifida. Polymalformative syndrome in our series accounted for 10.5% of the reasons for patient admission. It represented 29.6% admission reasons [9].

Antenatal ultrasound was performed in 80% [10] and 97% [7]. It was very little performed in our study (9.7%). This could be explained by the low accessibility (geographical and financial) of this examination to our populations. In addition, ultrasound is not a mandatory examination for monitoring pregnancy, according to the protocol in force in our country [11]. Antenatal ultrasounds performed could not lead to the diagnosis of CNS malformation in 39.2% of the cases in our

Table 1- Distribution of patients by age group

Diagnoses	Cases with a Surgical Indication		Cases without surgical indication	Total	
	Operated (n)	Not operated (n)	(n)	(n)	%
Spina bifida	37	38	0	75	14,31
Spina bifida + Hydrocephalus	25	49	0	74	14,12
Hydrocephalus	64	2	0	66	12,59
Cephalocele	21	33	0	54	10,3
Dandy Walker malformation	0	26	0	26	4,96
Spina bifida + Hydrocephalus + Clubfoot	18	7	0	25	4,77
Hydrocephalus + Dandy Walker malformation	2	23	0	25	4,77
Craniostenosis	0	20	0	20	3,81
Intracranial arachnoid cyst	4	0	15	19	3,62
Spina bifida+ Hydrocephalus +Arnold Chiari malformation+ Clubfoot	0	17	0	17	3,24
Microcephaly	0	0	16	16	3,05
Schizencéphaly	0	0	16	16	3,05
Spina bifida + Clubfoot	0	16		16	3,05
Hydranencephaly	0	0	15	15	2,86
Arnold Chiari malformation	2	10		12	2,29
Corpus callosum agenesis	0	0	11	11	2,09
Hydrocephalus + Cephalocele	6	3	0	9	1,71
Spina bifida+ Hydranencephaly	0	3	0	3	0,57
Semi lobar holoprosencephaly	0	0	2	2	0,38
Arachnoid cyst +Microcephaly	0	0	2	2	0,38
Lissencephaly+ Microcephaly	0	0	2	2	0,38
Schizencéphaly+ corpus callosum agenesis	0	0	2	2	0,38
Hemivertebra of L1	0	1	0	1	0,19
Hemivertebra of L1+ clubfoot	0	1	0	1	0,19
Caudal regression syndrome + knees flessum + club feet	0	0	1	1	0,19
Intraspinal arachnoid cyst	0	1	0	1	0,19
C1anterior arch agenesis with subluxation C1-C2+ Vertebral block C6-C7	0	0	1	1	0,19
Lissencephaly + craniostenosis	0	0	1	1	0,19
Spina bifida+ hydrocephalus+ Clubfoot+ Horseshoe kidney	0	1		1	0,19
Corpus callosum agenesis + cleft lip	0	0	1	1	0,19
Crouzon syndrome+ Semi lobar holoprosencephaly	0	0	1	1	0,19
Microcephaly+ Down Syndrome (trisomy 21)	0	0	1	1	0,19
Spina bifida + Genu valgum	0	1		1	0,19
Microcephaly+ nose bone malformation	0	0	1	1	0,19
Hydrocephalus+ Persistence of the peritoneovaginal canal	0	1	0	1	0,19
Spina bifida+ Hydrocephalus + Arnold Chiari malformation + Incomplete fusion 8th-9th ribs	0	1	0	1	0,19
Spina bifida+ Clubfoot+ Persistence of the peritoneopavaginal canal	0	1	0	1	0,19
Spina bifida+ Hydrocephalus+ Clubfoot+ left renal ptosis	0	1	0	1	0,19
Schizencephaly + Mega large cistern	0	0	1	1	0,19
Total	**179**	**256**	**89**	**524**	**100**

series. Elsewhere this diagnosis was not made in 25% [7] and 51% [12]. This would be related to the fact that some malformations express themselves little or not until birth, but also, ultrasound is an examination that depends on the operator and the performance of the ultrasound. The sensitivity, specificity, positive predictive value, and negative predictive value of ultrasound in the diagnosis of fetal CNS malformation in early pregnancy were 85.7%, 100%, 100% and 99.9%, respectively. [13]. Fetal MRI is more accurate and allows a better characterization of lesions but because of the acceptable performance of ultrasound and the poor geographical and financial accessibility of MRI, MRI is indicated in case of limitation of ultrasound. [10, 14]. In our context, it would be desirable to make ultrasound more accessible and mandatory for pregnancy monitoring. Postnatally, CT was performed in 79.8% in our study. Elsewhere it was MRI that had been performed in 100% of cases [5] while this rate was 1.7% in our study. This could be explained by the very low financial and geographical accessibility of patients in our study to these medical imaging techniques.

Neural tube defects were the most common in the literature [1,4,6,7,15]. The diagnoses were myelomeningocele (79.6%), encephalocele (12.5%) and Dandy Walker's malformation (9.1%) [1]. It was spina bifida in 80% and encephalocele in 19% [4]. We made the same observation in our series, probably related to the fact that neural tube defects and hydrocephalus are more symptomatic causing symptoms that worry and lead patients to consult most often, since they are hospital series for the most part.

Treatment of CNS malformations is most often surgical and depends on the type of malformation [7]. In our study a surgical indication was posed in 83%. Of the cases in our series where there was an operative indication, 41.1% had been operated. The reasons that could explain the fact that patients have not been operated despite a surgical indication were among others the insufficiency of the technical platform for a global management of certain patients (polymalformative syndrome); the large number of lost to follow-up (231 cases) which could be explained on the one hand, by the financial difficulties of patients to meet the expenses generated by this surgery and, on the other hand, by the long waiting times for surgery. Indeed, the average waiting time for surgery in our study was 31.2 days. This long average waiting time for surgery could be explained by the small number of nursing staff and the inadequacy of the technical platform (equipment and premises). The death rate pending surgery was 18% [6]. In our series we believe that most patients lost to follow-up among patients in whom there was a surgical indication died.

If the management of CNS malformation is not without great difficulty, that of associated malformations in our context pose even more problems. Thus, in no case of surgical management, the associated malformation was done in multi-teams at the same operative time as the management of the CNS malformation as is desirable in some cases. All CNS malformations were operated isolation and then the patient was referred to another specialty department for the management of the associated malformation.

The immediate postoperative evolution was favorable or stationary in 91.5% in our study. Elsewhere, it was in 84% [16]. While this immediate development seems generally satisfactory in our study, the medium- and long-term evolution was less so. There were 2 cases (0.4%) of deaths in our study. These 2 cases of death were patients with hydrocephalus including 01 cases associated with myelomeningocele. It is therefore a very non-lethal condition when treated in time. On the other hand, when the long-term follow-up is well carried out, disabilities and serious difficulties of social integration are to be feared, even in patients without surgical indication [17,18].

CONCLUSION

CNS malformations are common in our context. Diagnosis of these lesions and neurosurgical treatment are mostly possible. The main CNS malformations diagnosed were spina bifida, hydrocephalus and encephalocele.

A surgical indication was posed in almost all cases, but surgery could only be performed in less than half of these cases. The functional prognosis and long-term follow-up of patients still need to be improved through multidisciplinary care with the involvement of political decision-makers to improve the technical platform.

Given the large number of CNS malformations and the difficulties in managing them, emphasis should be placed on prevention.

DISCLOSURES

Ethical approval

This study was performed in line with the principles of the Declaration of Helsinki. Approval was granted by the local Ethics Committee.

Consent to participate

The patients gave consent to use their information and images for research purposes. *Consent for publication*

Conflict of interest

The authors declare no conflicts of interest with respect to the content, authorship, and/or publication of this article.

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CONTRIBUTIONS

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-Abdoulaye Adamou Babana: Conceptualization, Data curation, Formal analysis, Investigation, Methodology, Validation, Visualization, Writing – original draft, Writing – review & editing.
-Yakouba Haro: Conceptualization, Data curation, Methodology, Validation, Writing – review & editing.
-Abdoulaye Sanou: Conceptualization, Methodology, Validation, Writing – review & editing.
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-Julie Marie Adeline Wendlamita Kyelem Kafando: Conceptualisation, méthodologie, validation, rédaction – révision et édition.
-Abel Kabre: Conceptualisation, méthodologie, supervision, validation, rédaction – révision et édition.

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