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Demographic and clinical characteristics of central nervous system congenital anomalies: A one year review of cases managed at UDUTH Sokoto

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Abstract

Background: The spectrum of CNS congenital anomalies varies from one geographical location to another across the globe. This essentially can be solitary or multiple and can be cranial, spinal, combination of both cranial and spinal or others. The understanding of their distribution in each location could determine local workforce and facility needs and as well guide in the areas of sub specialization.

Objective: The aim of this study is to document the pattern of CNS congenital anomalies cases that had surgical intervention, at the Usmanu Danfodiyo University Teaching Hospital, Sokoto.

Methods: A cross-sectional, retrospective hospital based study from January, 2021 to December, 2021 of all patients with CNS congenital anomalies who had surgical intervention at the Neurosurgery Department, Usmanu Danfodiyo University Teaching Hospital, Sokoto, Nigeria.

Results: A total of 49 cases presented to our facility and had surgical interventions for central nervous system congenital anomaly. Of those 49 cases with CNS Congenital anomalies, there were 25 (51.0%) males and 24(48.98%) females, with a male-to-female ratio of 1.04: 1. The median age was 185 days, with age range of 2– 364days. Overall, the most commonly diagnosed neurosurgical CNS congenital anomalies in order of decreasing frequency were Congenital hydrocephalus 20 (40.82%), Myelomeningocele 14 (28.57%), Encephalocele 10 (20.41%), combination of Hydrocephalus associated with myelomeningocele 8(16.33%), Meningocele 3(6.12%), Lipomeningocele 1(2.00%), Dandy walker malformation 1(2.00%) , and Dandy variant1(2.00%) .

Conclusions: NTDs (Myelomeningocele, Meningocele and lipomeningocele) were the most common CNS anomalies (55.10%). However, congenital hydrocephalus, Myelomeningocele, Encephalocele, Hydrocephalus + MM, and Meningocele are common in our environment in decreasing frequency. Though, Lipomeningocele, Dandy walker malformation and dandy variant are relatively rare. Understanding of this distribution may guide service expansion and workforce needs, inspire sub specialization as well as direct research and government policy in this community.

Keywords: Central nervous system; Congenital anomalies; Congenital hydrocephalus; Encephalocele; Myelomeningocele

1. Introduction

In the main, 2 – 3% of births are associated with major congenital anomalies diagnosed around the birth period.[1] In comparison with other congenital malformations, central nervous system (CNS) congenital anomalies are unrecognized

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and underreported causes of childhood morbidity and mortality in Nigeria especially if intervention is delayed and most of Africa Congenital malformations of the central nervous system (CNS) are diverse, and complex in origin and development.[1–4] those that require neurosurgical intervention remain a major focus in neurosurgical centers.

The Prenatal detection of CNS malformations has substantially improved in the last decade. In developing countries, congenital anomalies of the CNS are sometimes not properly diagnosed and underreported (the conditions are attributed to forces of nature, which are beyond human intervention); and when recognized there is delay in presentation for surgical intervention[1,5]. Binitie in 1992 recorded 80 cases of CNS malformations in 2 years, although, study done 23years ago in Nigeria reported 111 cases. The overall incidence of CNS malformations is about 1 in 100 live birth.[1,6,7] most documented incidences and the pattern on this subject matter vary in different geographical locations. The degree to which the documented variations are attributable to differences in genetic predisposition, environmental factors or diagnostic facilities remains uncertain.[6–8] the population, of a tropical developing country like Nigeria, is a mixture of different groups where consanguinity is practically allowed especially in the Northern part for most tribal groups.

Studies on the incidence and pattern of different types of congenital abnormalities can provide valuable information for planning health care services, including preventive programmme. The aim of this study is to establish and document the frequency and pattern of CNS Congenital anomalies in our region (north western Nigeria) so that future preventive programmes can be planned. These findings may direct policies regarding facility expansions and upgrade as well as staff capacity development as they relate to Neurosurgical service delivery.

2. Materials and Methods

This is a cross-sectional, retrospective assessment of all new patients seen from January, 2021 to December, 2021 the inclusion criteria were all cases of CNS Congenital anomalies who had surgical intervention. Patients with CNS Congenital anomalies are seen at the Neurosurgery Outpatient Clinic, Emergency Department as well as inpatient referrals from SCBU or ICH of Usmanu Danfodiyo University Teaching Hospital, Sokoto, Nigeria.

The relevant information obtained from the patient's case notes included patient's age at presentation, gender, clinical (diagnosis and location), therapeutic data and diagnostic categorization. The data collection was done using a semi structured questionnaire and Microsoft Excel, and then exported toSPSS, version 25.0 (IBM Corp, Armonk, NY) for the analysis. The results are presented in simple frequency tables and percentages.

3. Results

A total of 115 new patients were seen over the study period. Only 49 (42.6%) cases were CNS congenital anomaly out of which 25 (51.0%) males and 24(48.98%) females, with a male-to-female ratio of 25:24. The median age was 185 days, with age range of 2– 364days. Majority of the patients, 28(57.14) were between the ages of 85 to 168days, whereas only 1 (2.00%) patient is at 364 days representing the least age group. There was bimodal peak age of presentation at 29 to 84 days and 85 to 168 days representing 17(34.69%) and 19(38.78) of patients respectively. Overall, the most commonly diagnosed Neurosurgical CNS Congenital anomalies in the North western Nigeria among new patients with Central Nervous System Congenital anomaly in order of decreasing frequency were Congenital hydrocephalus 20 (40.82%), Encephalocele 10(20.41%), Myelomeningocele with associated hydrocephalus) 8(16.33%), Myelomeningocele 5(10.20%), Meningocele 3(6.12%), Lipomeningocele 1(2.00%), Dandy walker malformation, 1(2.00%), Dandy variant1 (2.00%) and others 0%. More than half of the patients had hydrocephalus. This was associated with MM in 8(16.33%) necessitating cerebrospinal fluid shunting in 57% of cases. The most common clinical presentation of the spinal congenital malformation is lumbosacral 12(70.59%), while the Cervical, Thoracic 2 (11.76%) each and Sacral 1(5.88%) making it the least common site of the congenital spinal lesion. There were 32(65.30%) cranial, 9(18.37%) spinal, and combined cranial and spinal malformations in 8 patients (16.33%). Generally, most patients tend to present late (Table I). Only Sixteen percent of the patients presented within the neonatal period. Treatment was dependent on the pathology. These included ventriculoperitoneal shunts for hydrocephalus, excision and repair of MM, Lipomyelomeningocoele, Meningocele and Encephalocele. Cystoperitonial shunt for Dandy walker malformation and shunt placement for Dandy variants.

Table 1 Demographic profile

Variables	Frequency (%)
SEX	
Male	25(51.0%)
Female	24(48.98%)
AGE (DAYS)	
≤ 28	8(16.33%)
≥29	41(83.67%)
Total	49(100%)

Table 2 Distribution of CNS Congenital Anomalies

Clinical presentation	(%) Frequency
Cranial	32(65.30%)
Spinal	9(18.37%)
Cranial + Spinal	8(16.33%)
Total	49(100%)

Table 3 Spectrum of CNS Congenital Anomaly in regional centre for Neurosurgery

Congenital hydrocephalus	**20(40.82%)**
Encephalocele	10(20.41%)
Myelomeningocele + hydrocephalus	8(16.33%)
Myelomeningocele	5(10.20%)
Meningocele	3(6.12%)
Lipomeningocele Dandy Walker Malformation	1(2.00%) 1(2.00%)
Dandy Variants	1(2.00%)
Total	**49(100%)**

Table 4 Distribution base on site of spinal pathology

Site of pathology	Frequency (%)
Cervical	2(11.76%)
Thoracic	2(11.76%)
Lumbo sacral	12(70.59%)
Sacral	1(5.88%)
Total	17(100%)

4. Discussion

Sokoto is an ancient city in north-western Nigeria, a sub-Saharan country of Africa. It lies in the Rima Basin area between latitude 12º15' 29" North of the equator and longitude 13º 58'22" East of the Greenwich meridian, and it is situated at elevation 296 meters above the sea level.[9,10]

Global surveys have shown that the frequency of congenital anomalies varies greatly from 0ne geographical location to another. CNS malformations linked to essential cause of morbidity and mortality during the childhood period. Women, who previously have had a child with a NTD, have 2% risk of recurrence.[1,11] prenatal diagnoses of CNS anomalies are possible by measuring maternal serum concentration of α -fetoprotein and neuroimaging examination of the foetus. α -Fetoprotein, is the principal plasma protein of the foetus, is present in amniotic fluid. Its concentration in the amniotic fluid is increased when plasma proteins exude through a skin defect.[11]

Prenatal imaging is on the other hand non-invasive. In our region, delayed and or late presentations to tertiary institutions or medical care are exceptional and usual occurrences in developing nations with Nigeria not out of the norm. Despite the policy of substantial subsidization of the cost of treatment of children in our centre, patients still present late (84% after their neonatal period one month of age). This further supports the findings of Catibusic et al and Komolafe et al. in their respective study. It was made clear that the reasons for late presentations were lack of awareness, poverty and in some the expectation of demise of the baby.[11,12] NTDs is the most frequent CNS congenital anomalies; they result from failure of the neural tube to close spontaneously between the 3rd and 4th week of embryonic development.

The neural tube anomalies unlike many other structural malformations of interest are identifiable at birth. There is a global variation in the incidence rates of NTD as well as differences in individual regions. Several CNS anomalies including NTD and hydrocephalus are reported to be declining in incidence over the past three decades in the Western world.[13,14] NTDs (Myelomeningocele, Meningocele and lipomeningocele) were the most common CNS anomalies (55.10%) in our study. This is similar to what obtained in these series.[1,2,5,13,14]

Congenital hydrocephalus presents a special problem because it may be postnatal in origin and delay presentation. In our study we recorded a very high congenital hydrocephalus 20 (40.82%) The high frequency is further supported with what obtained in Komolafe et al [12] , Dandy – Walker malformation –2.00% and Dandy variants are relatively rare in most series . In our study, the frequency of myelomeningocele and encephalocele exist in the ratio of 1.3: 1. Earlier studies recorded ratio of myelomeningocele to encephalocele to be 3:1 in Nigeria and 8:1 in South Africa.[15,16]Prevention is certainly the best form of therapy. Primary prevention of CNS malformations is limited; with the exception of NTDs. Periconceptional folic acid supplementation and/or food fortification with folic acid have reduced significantly both the first occurrence and recurrence of NTDs in the off spring.

5. Conclusion

Late presentations of CNS congenital anomalies are common in our environment. NTDs were significantly more common. Health education about congenital malformations during ante-natal care, early neurological evaluation, definitive neuroimaging and timely intervention should be strongly considered.

Congenital hydrocephalus, Myelomeningocele, Encephalocele, Hydrocephalus with associated MM, and Meningocele are common in our environment. However, Lipomeningocele, Dandy walker malformation and dandy variant are relatively rare.

Understanding of this distribution may guide in the service expansion and workforce needs, inspire sub- specialization as well as direct research and government policy in this community.

Nongovernmental organizations wanting to have impact will be appropriately directed.

Compliance with Ethical Standards

Disclosure of conflict of interest

The authors declare no conflict of interest.

Statement of ethical approval

Ethical approval was sought and obtained from the Hospital's ethical committee.

Statement of informed consent

This was a retrospective study; no informed consent was obtained from patients.

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