

PROFILE AND MANAGEMENT OF CENTRAL NERVOUS SYSTEM
MALFORMATIONS IN A TERTIARY HOSPITAL

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ABSTRACT

Background: Central Nervous System (CNS) malformations rank among the commonest malformations. These may be identified in utero or noticed immediately after birth. Most studies showed Myelomeningocele, a Neural Tube Defect (NTD) to be the commonest. Severe defects have a significant impact on long term disability. While some are preventable, most are surgically managed. **Objective:** To study the profile and management of CNS malformation in a tertiary hospital. **Method:** we undertook a 3-year retrospective study of the profile and surgical management, including the postoperative complications among Neonates admitted into the Special Care Baby Unit (SCBU). Data on place of delivery, Gender, Gestational age, Mode of delivery, Prenatal Diagnosis (Ultrasound Scan), types of malformations, types of surgeries, and postoperative complications were retrieved. **Result:** We found 71 patients with CNS anomalies (31.8% of all congenital anomalies), in delivery 10 (14.1%), out delivery 46(64.8%). Male=43, females=28. Term gestation 66 (93%), preterm 3(4.2%), post-term 2(2.8%). Delivered by Vagina (SVD) in 65(91.6%), Caesarean section 4(5.6%), and undocumented in 4(5.6%). Prenatal diagnosis in only 3(4.2%). Myelomeningocele was 35(49.3%), meningocele was 9(12.7%), Hydrocephalus was 18(25.3%), anencephaly was 1(1.4%), occipital Encephalocele was 6(8.5%), Sincipital Encephalocele was 2(2.8%). Operated on 53(75%), LAMA/lost to follow was 13(18%), preoperative death was 5(7%). Excision and closure of Neural Tube Defect were 34(64.2%), VP Shunt was 13(24.5%), Excision and closure of occipital Encephalocele were 5(9.4%) craniofacial excision and repair of Sincipital Encephalocele was 1(1.9%). CSF leak in 4(28.6%), wound infection in 3(21.4%), shunt infection and obstruction in 2(14.3%) each. meningitis, hypertrophied scar, and pseudo meningocele in 1(7.1%) each. **Conclusion:** The commonest CNS anomaly is a Myelomeningocele a preventable condition, Sincipital Encephalocele is not common in our environment. Concomitant treatment of hydrocephalus averts post excision CSF Leak.

Key words: CNS Malformation, Complications, Encephalocele, Myelomeningocele, Surgeries.

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INTRODUCTION

Congenital Malformation is structural defects identified in utero or noticed immediately after birth, characterized by either a single or multiple defects. It has a worldwide prevalence of 2-3%,¹ is associated with high prenatal mortality (one in three babies), and significant post-natal defects.² Worthy of note is that 94% of severe malformations

occur in low- and middle-income countries and the impact of this on long-term disability, which may be significant on individuals, families, healthcare systems, the economy, and societies cannot be overemphasized.² Central nervous system (CNS) congenital anomalies are one of the most common congenital abnormalities.³

CNS anomalies include those of neural tube formation (Neural Tube Defects), regionalization (holoprosencephaly), cortical development (microcephaly), posterior fossa structures (aplasia or hypoplasia of the cerebellar hemispheres, Dandy-Walker malformation), and combined (agenesis of the corpus callosum). Several studies had indicated that Neural Tube Defects (NTDs) constitute a majority of these anomalies.^{5,7}

The reported prevalence of congenital Central Nervous System (CNS) malformations was 8.8% in Tanzania.⁸ In the middle belt of Nigeria, an incidence rate of 7/1000 live new-borns had been reported.⁷ Ambe et al¹⁰ found the prevalence of CNS anomalies in a neonatal unit in Maiduguri to be 3.4%. Incidence and the pattern of congenital CNS anomalies tend to vary depending on the geographical locations.^{11,12} However, there is no known study on the prevalence, pattern, and surgical outcome of CNS anomalies at the Federal Medical Centre Yola (FMCY), a tertiary health facility in North-Eastern Nigeria.

MATERIALS AND METHOD

This is a retrospective study of cases of CNS anomaly admitted to Federal Medical Centre, Yola (FMCY), Adamawa State, North-eastern Nigeria over a three year period (January 2017 to December 2019). Yola is located on latitude 9.21 and longitude 12.48 and it is situated at an elevation of 162 meters above sea level. The hospital has a 363-bed capacity. It serves as a referral centre not only for Adamawa state but the nearby state of Taraba, parts of Borno, and Gombe states including the neighbouring part of Cameroon republic. The Hospital SCBU admits neonates only, with a capacity for 25 patients and equipped with 2 neonatal ventilators. Data on patient's Characteristics, system(s) involved/type, and the surgical intervention offered and complications were retrieved from clinical notes of all patients with congenital anomalies admitted

into the Special Care Baby Unit (SCBU), with emphasis on CNS anomalies using a proforma design for the study. The data was presented in tables and a pie chart.

RESULTS

A total of 2,294 neonates were admitted into the SCBU during the study period. Two Hundred and Twenty-Three (9.7%) of this had various congenital anomalies. Of this number, Seventy-one (31.8%) had a Central Nervous System (CNS) anomaly. CNS involvement is second to the Gastrointestinal system only. The characteristic feature of the patients with CNS anomalies is as shown in table 1. Only 10 patients of those with CNS involvement were delivered in our centre - FMCY (14.1%), while most (61) were referred following delivery elsewhere (64.8%). There were 43 males and 28 females with Male to Female ratio of 1.5:1.

The main mode of delivery was Vaginal (91.6%); conducted at term (93.0%). prenatal Ultrasound Scan (USS) diagnoses were available in only 3 patients (4.2%).

Table 1: characteristics of patients with CNS Anomalies (n=71)

PARAMETERS	NUMBER	PERCENTAGE
Place of Delivery:		
in FMCY	10	14.1%
Out of FMCY(Referred)	46	64.8%
Undocumented	15	21.1%
Gender:		
Male	43	60.6%
Female	28	39.4%
Gestational age:		
Preterm	3	4.2%
Term	66	93.0%
Post-term	2	2.8%
Mode of delivery:		
Caesarean Section	2	2.8%
Vaginal Delivery	65	91.6%
Undocumented	4	5.6%
Prenatal Diagnosis (by USS):		
Yes	3	4.2%
No	68	95.8%

Neural Tube Defects (Myelomeningocele + Meningocele) was found in 54 (76.1%) of the CNS malformations followed by 18 cases of Hydrocephalus (10 Chiari Malformation, 6 Aqueductal stenoses, and 2 Dandy-Walker malformations), Encephalocele (6 occipital and 2 Sincipital) and Anencephaly (1) respectively as in the table below.

Table 2: Types of CNS Malformations (n=71)

Type of Malformation	Number(n=71)	Percentage
Myelomeningocele (MM)	2	2.8 %
= Two (2) level	33	46.5%
= One (1) level	9	12.7%
Meningocele		
Hydrocephalus:	10	14.0%
= Chiari II Malformation	6	8.5%
= Post NTD repair	2	2.8%
= Post Infective	1	1.4%
Anencephaly		
Encephalocele:	6	8.5%
= Occipital	2	2.8%
= Sincipital		

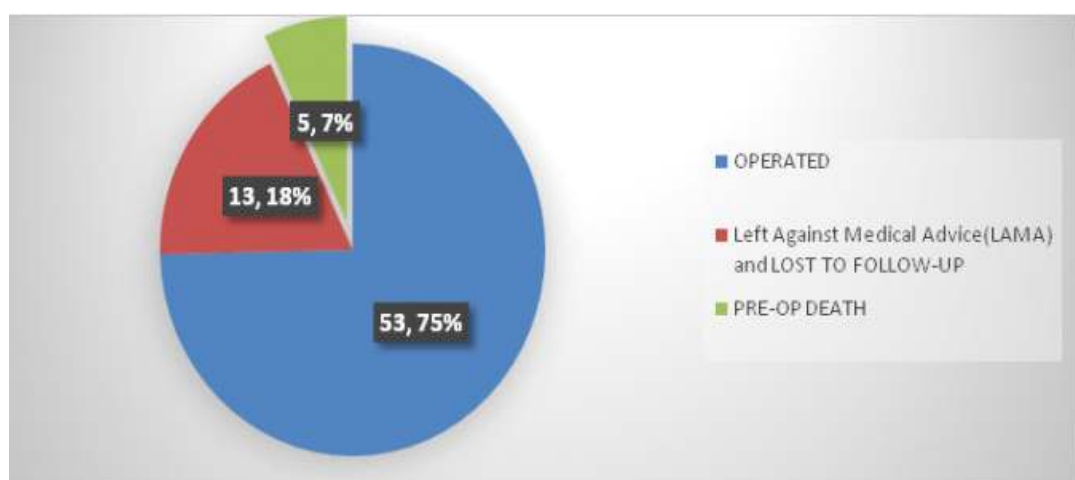


Figure 1: Pie Chart Showing management outcome(n=71)

Table 3: Showing Surgeries (n=53)

CLINICAL DIAGNOSIS	NUMBER	PERCENT	SURGERY
Myelomeningocele	26	49.1%	Excision and Closure
Meningocele	8	15.1%	Excision and Closure
Occipital Encephalocele	5	9.4%	Excision and Closure
Sincipital Encephalocele	1	1.9%	Craniofacial excision and closure
Hydrocephalus	13	24.5%	Ventriculoperitoneal shunt + 1 Cyst - peritoneal shunt

Table 4: Post-operative complications (n=14, 26.5%)

OPERATION	COMPLICATIONS	NUMBER	PERCENTAGE
Myelomeningocele excision	Cerebrospinal fluid Leak	4	28.6%
	Pseudo meningocele	1	7.1%
	Wound infection	3	21.4%
	Meningitis	1	7.1%
Meningocele excision	Hypertrophic scar	1	7.1%
V-P Shunt	Shunt obstruction	2	14.3%
	Shunt infection	2	14.3%

DISCUSSIONS

Two hundred and twenty-three(223) congenital anomalies were identified during the study period, Central Nervous System was involved in 71 (31.8%). this is less than the findings in Makurdi (38.4%)¹³ and cross river state (36.4%)¹ in Nigeria. While other studies in Maiduguri, North-Eastern Nigeria by Ambe JP et al,¹¹ in Jos,¹⁵ and Bida¹⁶ in Nigeria revealed far less than our findings, despite the similarity in climatic conditions with Maiduguri. Jos and Bida are known to be major cultivators of Fruit and vegetable, a rich source of folic acid, known to prevent NTD.

We found CNS anomalies to be slightly higher in males than females, with male to female ratio of 1.4:1. This is similar to the finding in Bida¹⁶ (1.4:1) and Jos¹⁵(1:1), but a reverse finding was reported in Sokoto¹⁷(1:1.4) and Lagos¹⁸(1:1.1)

Only 10(14.1%) Patients were delivered in our Hospital, while the majority were referred to our facility 46 (64.8%).An undocumented place of delivery was found in 15(21.1%) of the patients. Despite the few numbers delivered in our hospital, this study finding connotes a better uptake of obstetric service utilization than the study in jos¹⁵ which revealed that all of the patients were delivered outside and subsequently referred to their Hospital (100%).

In our study, prenatal Ultrasound Scan diagnoses were available in only 3 patients (4.2%). Ultrasound detected congenital CNS anomaly may account for up to 24.5% (Lagos).¹⁸ Another finding detected 27.6%.¹⁷ No foetal Magnetic Resonant Imaging (MRI) done. The prenatally diagnosed

malformation may allow the parents to decide in terminating, intrauterine repair, or elective delivery by Caesarean section and early surgical intervention.

The commonest CNS anomaly we found was Neural Tube Defect (NTD) in 54(76.1%). Most studies have revealed NTD as the commonest CNS anomaly. Our finding is similar to the findings in Sokoto,¹⁷Enugu,²⁰ and Jos Nigeria.¹⁶ However, this is about double the finding from Iraq,²¹ Kano²² (38.6%), and about ten times the report from Anambra²³ which may be explained by either public awareness or some local diets rich in folic acid.

Myelomeningocele was the commonest NTD that we found 45(63.4%).This is similar to the finding by Binitie O.P in Jos.²³ However, it is higher than the Lagos finding (57.4%) but less than the finding in Ilorin (73.8%).²⁵

Fifty-three patients (75%)had surgical interventions. our surgical interventions are far more than the findings of Emine Aydin et al²⁶ in Turkey who operated on only 29.6% of the 108 CNS Anomalies of their series and 38% of 61 in Ibadan Nigeria.²⁷NTD Surgery (Myelomeningocele and Meningocele) were the commonest surgical operation 34 (64.2%), out of which Myelomeningocele contributed 26(49.1%). A study in Ilorin,²⁵ Nigeria reveals 74% of surgery for myelomeningocele and meningocele. Thirteen (24.5%) had shunts for Hydrocephalus (ventriculoperitoneal) including 1 who had cyst – Peritoneal shunt for Dandy-Walker Malformation (DWM). Endoscopic Third Ventriculostomy (ETV)

was not available in our facility. Six patients with Encephalocele had surgeries of excision and closure in 5 occipital types, but a combined craniofacial excision in 1 Sincipital type.

Thirteen (18%) of the patient Left Against Medical Advice (LAMA) and lost to follow up. Globally Leaving Against Medical Advice has been reported to range between 1 – 25%.²⁸ Our finding is nearly similar to a study conducted in Gombe, North-Eastern Nigeria by Jalo et al²⁹ whose finding shows that 15.0% of patients with congenital anomaly signed and left against medical advice. Factors behind this may be related to cultural practices. Low average household incomes are identified as one of the reasons³⁰ among others. A paediatric surgery study in India by Pandey, et al³¹ identified the causes of LAMA in preoperative patients to include financial constraints, poor prognosis explained to the patient's attendants, the poor general condition of the patient, dissatisfaction with the available resources.

Postoperative complications were observed in 14 patients (26.5%). This is about 5 times more than the findings from a study in Pakistan.³¹ Our Post NTD surgery complications were CSF leak 4(28.6%), wound infection 3(21.4%), meningitis 1(7.1%), pseudo meningocele 1(7.1%), and a hypertrophied scar in a case of excised meningocele 1(7.1%). All the observed CSF Leaks were from the patients that were at risk, who subsequently developed hydrocephalus after excision of myelomeningocele. Odebode²⁵ in Ilorin found CSF leak, wound sepsis, and wound dehiscence involving 2(4.5%) patients each out of the 45 he operated. Noman MA,³² in

Pakistan also observed the following complications of CSF leaks (8.1%), sepsis in (8.1%), and 11.2% of patients developed postoperative meningitis. Post VP Shunt complications that we encountered are shunt obstructions 2(14.3%) and shunt infections 2(14.3%). An Indian study³³ shows a shunt infection rate ranging from 0 to 31.4%, with a mean of 10 to 15%. Our shunt infection rate is within a reported rate of 8 – 15% from a review by m. Paff et al.³ Some studies have shown shunt infection incidence of 5%-15%.^{35? 36? 37} A study by Merkle³⁸ et al. shows the cumulative rate of infection at 6.1%. Despite the variability in the rate of shunt infection, children are at higher risk than adults.³

CONCLUSIONS

Our study found that CNS involvement is the second most common congenital anomaly, with Myelomeningocele a subtype of NTD as the commonest anomaly. Most of the anomalies were not identified before delivery as the prenatal diagnosis was poor. Non-concomitant treatment of evolving hydrocephalus may result in Cerebrospinal Fluid (CSF) leak following excision and closure of myelomeningocele.

We are suggesting raising awareness at the community level on the need to prevent NTD by the use of a periconceptional folic acid supplement.

LIMITATIONS: Because our study is a retrospective, therefore we are suggesting a prospective study for more information

CONFLICT OF INTEREST: None.

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