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Congenital Central Nervous System Abnormalities in Bauchi, Nigeria: Our Experience in 4 Years

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Abstract

Introduction: Congenital central nervous system abnormalities are among the leading cause of infant morbidity and mortality in developing countries. These abnormalities impair functions and cosmetic challenges among the survivors. We aimed to describe the pattern of the congenital CNS abnormalities in our environment.

Materials and Methods: We carried out a retrospective study on children who presented and managed for congenital CNS abnormalities from October 2018 to September 2022. Clinical data of the patients were retrieved from their record files, admission record and operation notes. The data were analyzed and results expressed.

Results: Two-hundred and thirteen children with congenital CNS abnormalities were reviewed within the study period. Congenital hydrocephalus (48%) was the most common abnormalities seen followed spinal bifida (27%). Forty-eight percent of the children presented after two month of life. Peri-conception folic acid intake/supplementation and febrile illness occurrence during first trimester of the pregnancy were reported as 15% and 29% of the mothers respectively.

Conclusion: congenital hydrocephalus and spinal bifida are the most common congenital CNS abnormalities in our practice. Many of the mothers of the affected children did not have peri-conception folic acid intake or supplementation. Prevention, early detection during ante ante-natal care and early presentation at birth of the cases are needed efforts in our society.

Keywords: congenital, central nervous system, abnormalities, management

Introduction

Congenital abnormalities are structural or functional anomalies which are present at birth or spoon after birth. Most of the congenital abnormalities impair functions, and cosmetic challenges. The impact of congenital abnormalities are enormous on the child affected, the parents and the health care systems. The low and middle income countries like ours are severely affected in all ramifications [1]. Congenital abnormalities of central nervous system are complex and vary both in origin and development, with potential for life long morbidity when treatment is delayed. The classification of congenital abnormalities can be grouped into neural tube defects, disorders of structural specifications, disorders of brain growth and size, disorders of skull growth and shape [2]. This classifications have been made possible from the advancement in medical practice with the use of genetics and molecular biology [3]. Associated

congenital abnormalities have been shown sometimes to occur simultaneously with central nervous system congenital abnormality [4].

The overall incidence of central nervous system abnormality is about 1 in 100 births, higher frequency has been reported in spontaneous abortion, which shows that this abnormality has higher intrauterine fatality [5]. Many reports have suggested that the incidence and pattern of congenital central nervous system abnormalities vary across different geographical locations [6, 7].

Sixty percent of the causes of congenital abnormalities in human beings are unknown while in about 25% of the cases of congenital abnormalities, the aetiology is multifactorial, which depicts interplay between environmental and genetic risk factors [8]. Folic acid deficiency increased the risk of neural tube defects and it has been shown that folate confers about 50-70% protective effect on women who consumed adequate among of it against neural tube defects [9]. Exposure during pregnancy to drugs such as thalidomide and phenytoin, alcohol, cigarette smoking, certain environmental chemicals and radiation have all been implicated in the causation of congenital malformations [10, 11]. The occurrence of congenital malformations has also been associated with advanced maternal and paternal age, parental consanguinity, increasing birth order and low birth weight [11, 12].

The study on the pattern of spread of central nervous system congenital abnormalities will help in proffering solutions; prevention the occurrence, access to health care services, prompt diagnosis and alleviating the morbidity with surgical interventions. The aim of this study was to present the account of various types of central nervous system congenital malformation in our hospital over 48 months.

Materials and Methods

The study was a cross-sectional retrospective study of all children with central nervous system malformation that presented to neurosurgery unit, department of surgery, Abubakar Tafawa Balewa University Teaching Hospital, Bauchi over 48 months (October 2018 - September 2022). The data were obtained from the patients' case file and recorded into a semi-structured proforma. All patients were examined and diagnosis were made from combination of clinical presentation and cranial ultrasound/computer tomography scan/magnetic resonance image.

The data collected include; age, age at presentation, gender, preconception folic acid usage, antenatal services, febrile illness during pregnancy family history of similar abnormality, type of malformations and associated congenital abnormality. Analysis of the data was done using SPSS version 22.0 (Statistical Package for Social Sciences version 22 statistical software Chicago, IL, USA).

Results

Two-hundred and thirteen children with congenital central nervous system abnormality during the study period. About half of these patients presented within 1 month of birth and presentation after one year of age accounted for about 17.8%. Male children constituted 61% with male to female ratio of 1:1.5. Children with congenital CNS abnormality were delivered by mothers whose age was less than 25years in about 55% while fathers above 45 years of age were responsible for 84% of the children with congenital CNS abnormality (Table 1).

Passive smoking was encountered among 59% of the mothers of the children with congenital CNS abnormality and only 15% of them had preconception folic acid ingestion. Alxohol intake among these mothers was insignificant only seen in 7% and antenatal care was accessed by only 36%. 29% of the women experienced febrile illness during pregnancy and most of the affected children (68%) were below number 4 in birth order (Table 2).

The most common congenital CNS abnormality was hydrocephalus accounted for 48% of the cases. Spinal bifida is the second most common abnormality (27%), however, spinal bifidal associated with hydrocephalus made up 16% of the patients. Other congenital CNS abnormalities in order of frequency were encephalocele (8%) and 2% for Dandy-Walker syndrome (Table 3).

Characteristics	*Frequency (%)*
Age at presentation	
< 1month	112(52.6)
1- 12 months	63 (29.6)
> 12months	38(17.8)
Gender	
Male	129 (61)
Female	84(39)
Paternal age	
< 35years	60(28)
35 – 45years	69 (32.5)
> 45years	84 (39.5)
Maternal age	
<25yeasr	116 (55)
25 -35years	52 (24)
>35years	45 (21)

Table 1: Demography of the patients and the parents.

Characteristics	*Frequency (%)*
Passive smoking	
Yes	126 (59)
No	87(41)
Preconception folic acid	
Yes	32 (15)
No	181 (85)
Alcohol intake	
Yes	15 (7)
No	198 (93)
Antenatal visit	
Yes	76 (36)
No	137 (64)
Febrile illness	
Yes	61 (29)
No	152 (71)
Birth order	
<4	145 (68)
≥4	68 (32)

Table 2: Maternal factors associated with congenital CNS abnormality.

Spinal bifida has some associated congenital abnormalities which include talipes (26%). Polydactyl (5%) and omphalocele accounted for 3.5%. encephalocele was associated with hydrocephalus in about 7.8% while polydactyl associated encephalocele was 6% (Table 4).

Congenital CNS abnormality	*Frequency (%)*
Spinal bifida	58 (27)
Hydrocephalus	102 (48)
Spinal bifida & Hydrocephalus	33 (16)
Encephalocele	16 (8)
Dandy-Walker Syndrome	4 (2)

Tables 3: Distribution of Congenital CNS abnormality.

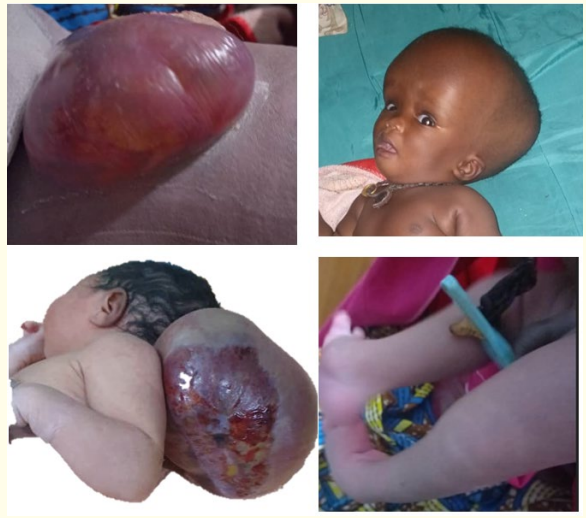

Figure 1: Various forms of congenital CNS and associated abnormalities

Congenital abnormalities	*Frequency (%)*
Spinal bifida Talipes Polydactyl Omphalocele	 15 (26) 3 (5) 2 (3.5)
Hydrocephalus Encephalocele	 8 (7.8)
Spinal bifida & Hydrocephalus Talipes	 16 (28)
Encephalocele Polydactyl	 1 (6)
Dandy-Walker Syndrome Talipes	 1 (25)

Table 4: Associated congenital abnormalities with congenital CNS abnormalities.

Discussion

Congenital CNS anomalies are among the major causes of childhood morbidity and mortality in many countries around the world. Surveys all over the world have shown that the frequency of birth defects varies greatly from region to region and from country to country. A male preponderance was observed in our study population, there have been similar reports on this [13-15] whereas there has been report of equal sex distribution as well [16]. The reason for this sex variation cannot be explained and this could be by chance. Late presentation of the children to the hospital are normal phenomenon in developing countries including our setting. About 48% of our study population presented after one month of birth and even some presented after I year of birth. This similar finding was reported by Komolafe et al [17].

Consanguinity in marriage was observed in about 35% of our study population. Consanguineous marriages have been shown to increase the occurrence of congenital anomalies as some recessive genes becomes expressed [18, 19]. Ignorance, poverty and expectation that the baby would die have been reported as some of the factors causing late presentation. Maternal age is an important risk factors in the occurrence of congenital anomaly. 45% of the mothers were aged 25 years and above. Existence of direct relationship between maternal age and incidence of congenital anomaly have been reported by some studies [20, 21]. Regards smoking, none of the mother smoked but 59% 0f them were passive smokers.

Folic acid deficiency has increasing risk of neural tube defect anomaly. With so many observational and interventional studies have shown 50-70% protective effect on women who took adequate amount of folic acid before conception against neural tube defect development [22, 23]. In our study, most of the women (80%) had no preconception folic acid and this could explain the high incidence of neural tube defect. Only 7% of the mothers admitted alcohol consumption prior to or during pregnancy. Alcohol intake, most especially chronic alcohol usage has been implicated in causing congenital abnormality, though the safest level of alcohol during pregnancy is not well known [24].

Febrile illness was seen in 29% of the mothers in this study, though occurrence of febrile illness has been implicated in congenital conditions particularly cardiovascular system anomalies, whether it has effect on the occurrence of congenital CNS anomaly cannot be explained in our study.

Hydrocephalus is the leading congenital CNS abnormality in the current study, this is similar to study in Ibadan, Nigeria few decades ago [25]. The send most prevalent conegenital CNS abnormality in our study is neural tune defect, this finding was in contrast to some studies in the recent times in our environment where neural tube defect (myelomeningocele) has become the leading anomaly [13-15]. There is worldwide variation regards the incidence rates of NTD in individual countries and regions. Several congenital CNS abnormalities including neural tune defect and hydrocephalus are reported to be declining in incidence over the past three decades in the Western world [7, 26].

The wide spectrum presentation of these abnormalities in this study has some associated congenital abnormalities, some within the CNS, while some are from other systems. Myelomeningocele and encephalocele ratio in the current study is 3.6:1. This is not different from some studies in our environment [27, 28]. Though a higher ratio of 8:1 has also been reported by De Clerk et al [29].

Conclusion

Congenital hydrocephalus was observed to be most common presented congenital CNS abnormality in our practice. Late presentations of these cases are still encountered in our environment. Pre and peri-conception folic acid supplementation and/or food fortification as a primary prevention will be of help. Advocacy and health education on the congenital CNS abnormality will reduce incidence, improve early presentation and encourage timely surgical intervention.

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