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Congenital Anomalies of the Central Nervous System in the Niger Delta: A Neurosurgical Perspective

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ABSTRACT

Congenital anomalies of the neuraxis are an important cause of morbidity and mortality in the perinatal period and during infancy; presentation is rare after childhood though some present for the first time in older children and adults. There is a dearth of data from our geographically distinct population referred to as the Niger Delta, which includes Nigeria's major oil producing areas. A retrospective study of 213 patients surgically managed on our service from June 2006 to May 2014. We studied demographic, clinical and radiologic data as well as outcomes. Data was analysed using SPSS 21.0. We surgically managed 213 patients representing 17% of all neurosurgical (and 53% of paediatric neurosurgical) operations in our centre during the 8-year period. Most patients, 70%, presented in infancy; only 8% presented in the neonatal period and 2.8% after 15 years. Patients with neural tube defects presented earlier. Overall male to female ratio was 1.2: 1; however, there are more males born with hydrocephalus spina bifida while the reverse is the case with the others in this study. Age range at time of operation was 18 days to 34 years. The other anomalies managed surgically were encephalocoeles, craniosynostosis, porencephalic cysts and Adeloye- Odeku disease. The majority, 37.5%, were born to parents who lived in Warri, Sapele and other parts of Delta State as well as Port-Harcourt and Bayelsa; 31%, Benin City, 17% Edo North, 7.5% from the East (Onitsha etc), 3.7% from the West (Akure, Ondo) and 3.7% from the Middle belt (Taraba, Lokoja). Fifty-five percent, 55.5%, were from very low income/ low socio-economic settings, 38% from lower middle class and 6.5% upper middle class. Mean maternal age was 30.6 years (range 18 to 41 years). Likelihood of congenital anomalies increased with both maternal age and the sum of maternal and paternal ages. All the mothers had antenatal care in various facilities, 75.5% had pre-natal ultrasonography which detected all cases of hydrocephalus, but only 25% of encephalocoeles and none of the others. None had exposure to irradiation or teratogenic medication during pregnancy. Family history of previous congenital anomaly was positive in one case of spina bifida and two siblings with encephalocoele both operated in our facility. Congenital anomalies of the CNS are common in paediatric neurosurgical practice. Most present in early infancy while an index of suspicion, meticulous clinical and radiological examination will detect about 2.5 to 5% presenting between the second and third decades. Further study may be necessary to determine other factors which might be involved in congenital anomalies in the sub region.

Keywords: Congenital Anomalies; Neuraxis; Morbidity and Mortality; Childhood; Niger Delta; Paediatric Neurosurgical Practice

INTRODUCTION

Congenital anomalies of the central nervous system (CNS) are among the most common obvious systemic congenital anomalies as well as the leading causes of morbidity, mortality and foetal loss [1 – 6]. They are also among the few systemic congenital anomalies compatible with life; they have a high prevalence in low and middle income countries where they may account for up 30% of neonatal deaths related to visible congenital anomalies [6 - 11] Though these anomalies are noticeable at birth, ignorance, poverty and cultural stigmatisation result in late presentation in these settings [1,10]. The normal development of the central nervous system (CNS) includes five stages: (a) induction during which the mesoderm induces the ectoderm to develop into neural tissue, (b) formation of the neural tube, (c)

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regionalisation and specification which require the interractions between genes and signal molecules to form forebrain, midbrain, hindbrain and spinal cord (as well as neural crest and glial cells, and sensory, motor neurons and interneurons) in the rostro-caudal and dorsoventral axes, (d) proliferation and migration to form the peripheral nervous system and population of the six layers of the cerebral cortex, and (e) connection and selection with associated axon overgrowth to establish synaptic connections failing which apoptosis occurs [12] We present, in this study, the epidemiological pattern of surgically managed congenital abnormalities treated at our neurosurgical unit which receives patients from several states in Nigeria's oil producing region.

PATIENTS AND METHODS

Two hundred and thirteen patients (213) in all had various operations for congenital anomalies of the neuraxis. All the patients were admitted into neurosurgical service from the emergency unit, neurosurgical out patient service and by transfer from paediatric and neonatal wards.They had detailed neurosurgical examination. Radiological evaluation included transfontanelle ultrasonography, computed tomography and /or magnetic resonance imaging. Plain radiographs were only used to assess patients after cerebrospinal fluid (CSF) shunts. Laboratory evaluation included full blood counts, electrolytes, urea and creatinine, blood sugar, CSF biochemistry and microbiology. Other investigations were echocardiography used to exclude congenital heart diseases in selected patients and neuroophthalmic tests to document vision in patients with intracranial anomalies. Urgent CSF aspiration by ventricular puncture through the anterior fontanelle at mid-pupillary line was carried out for patients with hydrocephalus who suffered raised intracranial pressure prior to surgery.Patients with scalp ulcers, ulcerated encephalocoeles, and myelomeningocoeles with exposed placode or other forms of sepsis were managed with broad spectrum antibiotics initially and/or daily wound dressings and weekly bacteriology to aid healing before surgery.

RESULTS

A total of 2,898 patients were admitted into the neurosurgical service from June 2006 to May 2014, total number of all operations was 1253 and paediatric neurosurgical operations 402. Two hundred and thirteen (213) patients were operated for congenital anomalies of the neuraxis.

General

The ages of the patients at time of operation were 18 day to 34 years. The youngest patient had encephalocoele while the oldest patient was late presenting congenital hydrocephalus. There were 115 males and 98 females (ratio 1.4: 1). One hundred and forty-three patients (143) i.e. 67% presented in infancy; only 8% presented in the neonatal period. Four patients, 1.8%, presented between 15 and 34 years. Five patients, 4.6%, presented in the second and third decades. They all had hydrocephalus. Four underwent surgeries for both hydrocephalus and spina bifida Perinatal, Family and Social History. All the patients' mother admitted having ante-natal care of varying duration at health centre, maternities, general or mission hospitals, private clinics and teaching hospitals. 75.5% had pre-natal ultrasonography

which detected hydrocephalus and only 5 of 19 encephalocoele; none of the other anomalies were predicted. None had any known exposure to irradiation (e.g. x-rays) or use of teratogenic medication during pregnancy. Family history of previous congenital anomaly was positive in one case of spina bifida and two siblings with encephalocoele both operated in our facility. Place of Abode and Socio-Economic Status Sixty seven (67) lived in Benin City, 35 (17%) from Edo North, 79 (37.5%) from Warri, Sapele, Port-Harcourt, Bayelsa and other parts of Delta, 16 (7.5%) from the East (Onitsha etc), 8 (3.7%) from the West (Akure, Ondo) and 8 (3.7%) from the Middle belt (Taraba, Lokoja). One hundred and eighteen, 118, patients (i.e. 55.5%) were from very low income/ low socio-economic settings, 81 (38%) were from lower middle class and 14 (6.5%) upper middle class. Twenty, 25, of the patients were from indigent families and were operated by philanthropic groups; 15 patients with hydrocephalus were part sponsored by the unit by providing free ventriculoperitoneal shunts and other surgical materials. Maternal and Paternal Ages Maternal age ranged from 18 to 41 with a mean of 30.6 years (mode 32 years, median 32 years). Summing both maternal and paternal ages, in the group that added up to 35-40 years had 14 of the patients with anomalies, 41-60 years 51, 61-75 years had 81 and 76-90 years had 67 patients. Emergence of congenital anomalies increased with both maternal age and the sum of both maternal and paternal ages.

Hydrocephalus

There were 114 patients, 67 males and 47 females with hydrocephalus; only 64 (56.1%) presented in infancy. Nine had external ventricular drainage for ventriculitis before shunts were carried out. Three, 3, had endoscopic third ventriculostomy (ETV) which failed before ventriculoperitoneal shunts (VPS) were carried out. Ten, 10, (i. e. 8.8%) patients with hydrocephalus died, of intracranial sepsis.

Spina bifida

There were 64 patients (36 male, 28 female); 53 (82.8%) presented in infancy. They had repair of spina bifida.

Encephalocoeles

There were 19 (8.9%) patients with encephalocoeles (7male, 12 female). Two died from brain stem dysfunction.

Others

Seven, 7 (3.3%), patients had Adeloye- Odeku disease, 6 (2.8) porencephalic cysts, 3 (1.4%) craniosynostoses each of which had male to female ratio of 2: 1.

Mortality

Overall, 12 (5.6%) patients died.

DISCUSSION

The patterns of CNS birth defects vary between regions and among different geographical situations in the same country[3 – 6, 13 – 22]. Some studies indicate incidence may exceed 20% in

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newborns in Nigeria[13-15]. Congenital malformations of the CNS rank high among lethal congenital anomalies, hydrocephalus, spina bifida and anaencephaly being the commonest [23,24] Male to female ratio was approximately 1 with slight male preponderance in our study like in most other reports in Nigeria (though some showed a ratio of 2:1) [1,8,9,16,22] The affected patients present to paediatricians much earlier, usually at birth or early neonatal period, but presentation to neurosurgeons is usually much later in infancy, early childhood or much longer after the reasons being ignorance, cultural, socio-economic and other reasons [1,2,8 - 11,14,15] Surgical correction rate varies (38-91%) for spinal bifida [2,9,15]. All cases in our series were operated, though established severe neurological deficits such as in myelomeningocoeles may persist. Operations for congenital anomalies of the neuraxis are the highest number of neurosurgical operations (with Bankole et al reporting spina bifida alone as the commonest indication for neurosurgical referral in Lagos in2012) besides trauma, in many of our settings [11,16,17] Odeku EL operated on 35 congenital anomalies (i. e 26% of a total of 134 operations) at the inception of Nigerian neurosurgery at Ibadan between 1962 October and 1963 October and Binitie on 80 of them representing 60% of total paediatric neurosurgical patients in Jos, Nigeria, between January 1984 and December 1985[17,18]. In our study, CNS anomalies accounted for 17% of all (and 53% of paediatric) neurosurgical interventions, respectively.

Mortality ranges from 1.4 to 5.8% comparable to our finding[2,16,21] Hydrocephalus was the commonest congenital malformation of the CNS in Benin City, Nigeria same as reported by Komolafe et al at Ile - Ife and Marinho at Ibadan; however, Odeku in 1962, Binitie at Jos in 1992 (60%), Bankole at Lagos in 2012 (48%) and Odebode at Ilorin in 2005 at (45%) reported spina bifida to be the commonest [6,10,11,16-18,22]. Other CNS anomalies such as encephalocoeles, craniosynostoses, Adeloye - Odeku disease, were not as common [1,2,22]. The risks of mortality, both perinatal and later, lifelong morbidity from neurological deficits and other systemic complications and sociocultural problems behoves the need for improved perinatal care, early referral, health education by trained experts on issues relating to congenital anomalies and aggressive approach to periconceptional folic acid supplementation for women; also, policies should be formulated to encourage fortification of staple food of staple food with folic acid to reduce neural tube defects [1,10,11] Majority of the causes of congenital anomalies are uncertain, but where the causes are known 25% appear to be multifactorial with complex interractions between genetic and environmental factors [25,26] Folate deficiency has a known teratogenic effect resulting in an increased risk of neural tube defects, observational and intervention studies showing 50-70% protective effect on women taking adequate amounts of folate on NTDS [26,27,28]. Recent advances in medical genetics and molecular biology have revolutionised the understanding of CNS development enabling precise classification of associated congenital anomalies based on the stage of development in which they occur[12]. Literature is replete with studies on neural tube defects because it involves complex interractions of genetic and environmental factors, as well as increased risk of same in relatives of affected members and amenability to multiple vitamin supplementations containing folic acid; relatively less is available anomalies associated with other stages of CNS development[29 – 42]. Careful assessment of patients with these abnormalities is important in order to provide an accurate prognosis and genetic counselling.

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Table I: Age and Sex Distribution of Congenital Anomalies of the CNS

	Hydro		S.B		Encep		Cranio.		P c		A.O		Total		GT
Age	M	F	M	F	M	F	M	F	M	F	M	F	M	F	
0-1mo	-	-	3	6	2	5	1	-	-	-	-	-	6	11	17
>1-3mo	6	4	12	7	4	3	-	-	-	-	-	-	22	14	36
>3-6mo	5	5	9	8	-	-	-	-	-	-	1	-	15	13	28
>6-12 mo	30	14	5	3	-	3	-	2	1	3	1	1	37	26	63
>12-24 mo	12	10	2	2	1	1	-	-	-	-	-	2	15	15	30
>2-3yr	3	2	1	-	-	-	-	-	-	1	-	1	4	4	8
>3-4yr	6	3	-	-	-	-	-	-	-	-		-	6	3	9
>4-5yr	1	2	2	1	-	-	-	-	-	-		-	3	3	6
>5-10yr	2	2	1	-	-	-	-	-	-	-		-	3	2	5
>10-15yr	2	2	1	-	-	-	-	-	-	-		-	3	2	5
>15-20yr	-	1	-	-	-	-	-	-	1	-		-	1	1	2
>20-30yr	-	2	-	-	-	-	-	-	-	-		1	-	3	3
>30yr	-	-	-	1	-	-	-	-	-	-		-	-	1	1
Total	67	47	36	28	7	12	1	2	2	4	2	5	115	98	213
GT	114		64		19		3		6		7		213		