

Neural Tube Defects In A University Teaching Hospital In Southern Nigeria: Trends And Outcome

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Abstract

Background: There has been some increase in the proportion of Neural Tube Defects (NTD) admitted in the University of Port Harcourt Teaching Hospital recently. For a largely preventable birth defect, this increase is both unnecessary and unacceptable.

This study was undertaken to describe the admission patterns and outcome of neural tube defects in University of Port Harcourt Teaching Hospital.

Methods: A retrospective study of babies with neural tube defects who were admitted into Special Care Baby Unit (SCBU) of the University of Port Harcourt Teaching Hospital from 1st May 2002 to 30th April 2005 was carried out. Their case notes were retrieved and information on the sex, maternal drugs during pregnancy, type of defect and associated malformations, prenatal diagnosis, management and outcome were obtained. The admission rate and the incidence were then calculated.

Results: There were 2891 total admissions (1691 males and 1200 females) during the study period of which 37(1.3%) were neonates with NTD. Of those with NTD, 25 were males and 12 female giving a male to female ratio of 2:1 (statistically not significant $p=0.242$.) The total hospital delivery at the study period was 7,388 of which 7 had NTD giving an incidence of 0.95/1000 deliveries. The commonest type of NTD was myelomeningocele in 31 (83.8%), and the commonest site was the thoracolumbar region (93.5%). Frontal encephalocele was seen in 6 (16.2%). All the babies with myelomeningocele presented with flaccid paraparesis and were incontinent of both urine and faeces. Seventeen of the babies had only spina bifida while 14 had additional defects including talipes equinovarus (8), hydrocephalus (2), frontal encephalocele (1), and multiple malformations (3). Ten babies (27%) died, three of them after surgery. All the mothers received folic acid from the second trimester of pregnancy, but none did before pregnancy.

Conclusion: The Incidence of NTD is on the increase in our environment. There is need to formulate/implement the policy of preconceptional folic acid therapy for all woman of childbearing age as a preventive measure

Key words: central nervous system, neural tube defects, folic acid supplementation

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INTRODUCTION

Neural tube defects (NTD) are a group of disorders resulting from failure of the neural tube to close spontaneously between the third and fourth week of in utero development (a time before many women realize that they are pregnant).^{1,2} They constitute the commonest congenital abnormality of the central nervous system. The commonest forms are myelomeningocele, anencephaly and encephalocele.² Spina bifida accounts for over 70% of all the neural tube defects and the commonest sites are the lumbosacral and the thoracolumbar regions.^{1,2}

In Africa, the disease has immense psychological and cultural implications for both the child and the family because of the unsightly deformity and also because of the poor outcome even with repair.³ The disorder is often associated with flaccid paralysis of the lower limbs, hydrocephalus, varying degrees of lower limb deformities and sphincter disturbances.^{1,2}

The causes are multifactorial, ranging from genetic determinants like mutations in folate responsive and folate-dependent pathways^{1,2,4} and chromosomal factors especially trisomy 21 and 18,^{1,5,6} to environmental factors, drugs like valproic acid and carbamazepine,^{1,5,7,8} malnutrition (especially folic acid deficiency),^{1,2} maternal factors (diabetes mellitus, obesity, hyperthermia, alcohol abuse)^{1,5,8,9} and maternal exposure to fumonisin-contaminated maize,^{1,10} chemicals and irradiation.^{1,2} Low socioeconomic status is also associated with increased risk in some population.¹

The incidence of NTD varies greatly between ethnic groups, being highest in whites and Hispanics, and lowest in black Africans and Asians.^{1,5,11-13} It also shows seasonal and geographic variation, being highest in

Great Britain, Ireland and China and lowest in Japan and Africa.¹¹⁻¹³ The incidence of NTD has however declined in the developed countries. Before the eighties, the prevalence in the United States was 1.5-4.5/1000 live births, but this has decreased to 0.74-2.5/1000 live births.¹¹

In China the incidence is between 1-4.8/1000 live births in the northern region and 0.6-1/1000 births in the southern region; the higher incidences being in women who did not take any folic acid.¹⁴ In Africa, the incidence was 0.63-1.74/1000 live births in South Africa,^{12, 13} 1.15/1000 live births in Ghana¹⁵ and 0.5/1000 in Nigeria.^{16,17} In Port Harcourt in the early nineties, the rate was 4.4/1000 admissions.¹⁸ There has however been some increase in the proportion of cases of NTDs admitted in the hospital recently. The objectives of this study are thus to describe the admission patterns and outcome of NTD in University of Port Harcourt Teaching Hospital.

SUBJECTS AND METHOD

A retrospective study of babies with neural tube defects who were admitted into Special Care Baby Unit (SCBU) of the University of Port Harcourt Teaching Hospital from 1st May 2002 to 30th April 2005 was carried out. Their case notes were retrieved and information on sex, maternal drugs during pregnancy, type of NTD and associated malformations, prenatal diagnosis, management and outcome was obtained. The admission rate and the incidence were then calculated. Statistical analysis was carried out using the chi-square test and a p-value of <0.05 was considered as being statistically significant.

RESULTS

Of the 2891 (1691 males and 1200 females) [(M:F = 1.4:1)] neonatal admissions, 37(1.3%) had NTD, 7 (18.9%) of who were inborn and 30 (81.1%) were outborn. Twenty-five of them were males and 12 of them females (M:F = 2.1:1) of which the male preponderance was not statistically significant ($\chi^2 = 1.24$; DF = 1, p=0.242). There were a total of 7388 deliveries within the period giving an incidence of 0.95/1000 deliveries. None of the mothers had a previous child with neural tube defect nor was any of them on anticonvulsants. There were no cases of consanguinity between the parents. Eleven of the mothers had prenatal ultrasound, but only in 3 was the defect detected.

Thirty-one (83.8%) of the neural tube defects were myelomeningocele and 29 (93.5%) were located in the thoracolumbar region. Six of the babies (16.2%) had frontal encephalocele and 2 of them had microcephaly. No case of anencephaly was seen (Table I). All the

patients with myelomeningocele presented with flaccid paraparesis, bladder and rectal dysfunction.

Seventeen (46.0%) had only myelomeningocele, whereas 14 (37.8%) had additional defects (Table II). Three of those with additional defects had multiple malformations. The first child had imperforate anus, camptodactyly, epispadias and low set ears. The second child had uterovaginal prolapse, bilateral inguinoscrotal hernia and polydactyly, while the third child had ventricular septal defect, imperforate anus, polydactyly, uterovaginal prolapse and low set ears. Surgery was done in 17 (46%) patients, 4 (11%) neonates had their surgery deferred and were discharged, while in 9(24%), the parents asked for discharge against medical advice before surgical intervention. Out of a total of 10 (27%) deaths, 3 occurred post surgery and 7 occurred before surgery. All the mothers received folic acid from the second trimester of pregnancy, but none did before pregnancy.

Table I. Types and site of the neural tube defects in 37 neonates

Type of lesion	Site	No.	%
Myelomeningocele	Thoracolumbar	29	78.4
	Lumbosacral	2	5.4
Encephalocele	Frontal	6	16.2
Total		37	100.0

Table II. Type of neural tube defects with the associated anomalies

Type of NTD	No	%
Myelomeningocele only	17	46.0
Myelomeningocele with talipes equinovarus	8	21.6
Myelomeningocele with multiple malformations	3	8.1
Myelomeningocele with hydrocephalus	2	5.4
Myelomeningocele with frontal encephalocele	1	2.7
Frontal encephalocele	6	16.2
Total	37	100.0

DISCUSSION

The overall admission rate of neural tube defect in this study was 13/1000 admissions. This is much higher than the rate of 4.4/1000 previously reported in Port

Harcourt in 1991¹⁸ and the 8/1000 admissions reported in Abuja.¹⁷ It is however lower than the admission rate of 19/1000 admissions reported from Jos.¹⁶ The incidence of 0.95/1000 deliveries found in this study is similar to the 0.94/1000 live births reported in Lagos.¹⁹ It is however higher than the 0.46/1000 live births reported in Ibadan²⁰ and the 0.5/1000 live births reported by Uba et al¹⁶ and Audu et al¹⁷ in North central Nigeria and Abuja respectively. Airede²¹ reported an incidence rate of 7/1000 live births in middle belt of Nigeria. In Saudi Arabia, the incidence was 0.78/1000 births, 50% of them being offsprings of consanguineous marriage.²² There was no case of consanguinity in this study as was also the case in other studies in Abuja¹⁷ and Jos.²¹

In Ghana the commonest type of NTD reported was anencephaly (73%),¹⁵ whereas in this study and in previous studies in Jos,^{21,23} Saudi Arabia,²² Ilorin²⁴ and Port Harcourt,^{2,5} the commonest type was meningomyelocele. In China, 20-35% had anencephaly.¹⁴ No case of anencephaly was seen in this study. The commonest site in most studies was the lumbosacral region,^{2,11,16,17,23,24} however in this study and in Saudi Arabia,²² the commonest site was the thoracolumbar region. There is increasing neurologic deficits as the myelomeningocele extends into the thoracic region. This may explain the severity of the neurological deficits seen in this study. That all the encephalocoels seen in this study were frontal differs from previous report in Ilorin²⁴ which showed a predominance of occipital encephalocoel. The sex predilection varies from place to place. Some studies done in United States of America¹, China¹⁴ and Jos²¹, Nigeria found that females outnumbered the males, but studies in Ibadan²⁰ and Ilorin²⁴ reported a male to female ratio of 1.2:1 and 1.3:1 respectively. Asindi and Al-Sheri found no sex predilection in Saudi Arabia.²² In this study, there was a male preponderance of 2.1:1.

For some unexplained reason, neonates with myelomeningocele in Africa show reduced incidence of pre-operative hydrocephalus determined by occipitofrontal circumference measurement and transfontanelle ultrasound, whereas reports from the developed countries show as much as 80-90% of them with pre-operative hydrocephalus.^{2,5,11} In Saudi Arabia, 87% had hydrocephalus at birth.²² These values are clearly much higher than the 5.4% of cases that presented with pre-operative hydrocephalus found in this study and the 5 out of 18 babies (27.8%) reported in Abuja.¹⁷

The incidence of NTD has been shown to be on the decline in Scotland²⁶ and United States^{1,2,5,11,27} due to the improvement in the general health and nutritional status of the populace as well as the prenatal diagnosis and termination of affected pregnancies. In Africa, the incidence has remained relatively the same. NTD has been conclusively shown to be aetiologically linked to folic acid deficiency²⁸ and supplementation with folic acid has led to a dramatic decline in the incidence in many parts of the world.^{14,27-31} A randomized double-blind prevention trial showed a 72% protective effect of folic acid supplementation.³¹ Folic acid supplementation not only reduces the risk of occurrence but also the risk of recurrence of NTD.^{29,31} Prevention using folic acid by all women of child bearing age before pregnancy is still not a practice in Africa despite its proven efficacy in other parts of the world. Knowledge and use of preconceptional folic acid is still very low.³² In this study all the mothers had some form of antenatal care and received folic acid, but after 12 weeks of gestation (at a time when neural tube formation would have been completed and supplementation will be of no benefit), with no mother receiving pre-conceptional folic acid supplements. This is similar to findings by other workers where majority of the pregnant women either have no antenatal care or they register in the 2nd or 3rd trimester.^{16,17,22,24}

Because of its proven protective effects, it has been recommended that all women of child bearing age who are capable of becoming pregnant should receive a daily dose of 0.4mg of folic acid in order to reduce their risk of having pregnancies complicated by NTD.^{33,34} To be effective, folic acid supplementation should be initiated before conception and continued until at least 12th week of gestation when neurulation is complete. There is thus a great need for the Ministry of Health of all African countries to make preconceptional folic acid supplementation mandatory if Africa is going to witness a decline in the incidence of NTD and thus reduce the immense psychological and neurological effects of this disabling birth defect.

CONCLUSION

The Incidence of NTD is on the increase in our environment. The overall morbidity associated with this serious and disabling birth defect calls for concerted efforts to identify aetiological factors and preventive actions. There is need to implement the policy of preconceptional folic acid therapy for all woman of childbearing age as a preventive measure.

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