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Neural Tube Defects- Demography in a Tertiary Care Center

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Abstract
Purpose: *Neural tube defects stands as a varied and relatively common emergency admitted to the pediatric surgery intensive care. The presentation varies ranging from anencephaly, encephalocele to meningomyelocele at different spinal levels. The present study aims to determine the demographic profile of all neural tube defect patients with respect to available literature.*
Methods: *A retrospective analysis of all patients attending the neonatal emergency with a diagnosis of neural tube defect was undertaken. The study period was two years. The demographic pattern of these patients with respect to different parameters was determined.*
Results: *The demography of different types of neural tube defects was obtained. The results were comparable in certain parameters and higher in others when compared to the values in world literature.*
Conclusion: *Neural tube defect is a preventable defect in today's world. The decreased incidence relates to the better maternalawareness and justifies the provisions for maternal health services.*
Key words: *Neural tube defects, encephalocele, meningomyelocele, demography.*

I. Introduction

Neural tube defects are one of the commonly admitted patients to the neonatal surgical emergency. The incidence ranges from 1-3 per 1000 live births worldwide[1]. They are among the most common of all birth defects. The etiology is poorly understood and believed to be multifactorial. Both genetic and environmental factors are implicated[1,2,3]. Embryologically, they are believed to result from failure of closure of the neural tube during neurulation at 21 to 28 gestational days. Primary neurulation involves formation of a neural plate and morphogenetic changes converting it to a neural tube. Secondary neurulation forms the medullary cord with subsequent cavitation forming the neural tube[3]. Any stagnation or alteration in the above processes results in a neural tube defect. They are usually classified as open or closed. In Open neural tube defects , the neural tissue is exposed and includes anencephaly, meningocele, meningomyelocele, myeloschisis and rachischisis. Closed neural tube defects have an exposed membranous covering with intact overlying skin. These include lipomyelocele, diplomyelia, diastematomyelia, neuroenteric cyst, dermal sinus, tethered cord and sacral agenesis[4,5]. The demography of the various defects vary with ethnicity, geography and maternal age groups. World literature suggests the highest incidence in Ireland and Wales with 6 cases per 1000 live births[1,5,6]. As far known, there are no demographic studies available on neural tube defects from the current population based region included in this study.

II. Materials and Methods

A retrospective study was undertaken in the Department of Pediatric Surgery, SMS Medical College, Jaipur over a duration of two years from April 2014 to March 2016. All patients who were admitted to the neonatal emergency in this duration with a diagnosis of neural tube defect were included in the study. The total number of patients were 340. The demographic profile of these patients wereanalysed. The parameters included the type of the defect, the location, sex ratio, maternal age group, social status and religion. In addition, the overall incidence of neural tube defects in our center was determined. The values were correlated to world literature.

III. Results

DEFECT TYPE	MALE	FEMALE	TOTAL	percentage
Meningomyelocele	108	88	196	58%
Meningocele	33	27	60	18%
Encephalocele	33	19	52	14%
Spina bifida occulta	05	07	12	4%
Rachischisis	07	01	08	2%
Anencephaly	10	02	12	4%
	196	144	340	

Table 1- Type of defects and sex ratio

DEFECT LOCATION	meningomyelocele	meningocele	Spina bifida occulta
Cervical	05	05	00
Thoracic	46	15	00
Lumbar	142	38	11
sacral	03	02	01

Table 2- Location of neural tube defects.

SOCIAL STATUS	No of patients	RELIGION	No of patients
Below poverty	262	Hindu	293
Above poverty	78	Muslim	32
		Others	15

Table 3- Patients below poverty line and religion ratio

DEFECT TYPE	Age<20	Age 20-29	Age30-39	Age>40
Meningomyelocele	11	102	58	25
Meningocele	11	28	12	09
Encephalocele	13	28	10	01
Spina bifida occulta	04	06	02	00
Rachischisis	06	01	01	00
Anencephaly	06	06	00	00

Table 4- Maternal age distributions of different defects.

The results show an increased incidence of patients diagnosed with meningomyelocele amongst all neural tube defects (Table 1). The meningomyelocele accounts for 58 % of the total patients which is more than double of the rest subtypes. If the patients with meningocele are clubbed together, both the entities constitute 76 % of all neural tube defects. Meningomyelocele occurs most commonly in the lumbar spine accounting for more than 80 % of all cases. Similar results are also valid for meningocele and spin bifida occulta which are also more common in the lumbar region (Table 2).

Demographically, the defects are much more common in patients belonging to low socio economic status. In fact, more than 80 % of the parents borning a neural tube defect baby belonged to the below poverty line population (Table 3). Similarly, a predisposition to Hindu population was also noted where all the anomalies were more evident.

Finally, we attempted to determine a relation of these patients to the maternal age at presentation. No significant correlation could be found. However, majority of the defects , other than rachischisis was more common in mothers presenting between 20 to 29 years age group. It is to be noted that a large number of mingomyelocele also occurred in mothers more than 40 years age (Table 4).

IV. Discussion

Neural tube defects are relatively common and a worldwide incidence of 3-4 cases per 1000 live births has been quoted[1,6,7]. Patients account for a major burden of all congenital diseases with multiple associated anomalies and delayed sequelae following treatment. A wide spectrum of neural tube defects have been described. Different studies carried out in India describe a variable but high incidence rate of neural tube defects. The rates vary from 8.7 in Delhi to 6.25 in Agra to 8.8 in Chandigarh per 1000 live births. A study from Davangere quotes a rate as high as 11 per 1000 live births[8,9]. Our study had an incidence rate of 3 per 1000 live births which was comparable to Indian studies. The absolute number of patients with neural tube defects were comparatively high (340 in two years) but SMS medical college hospital has approximately 60000 deliveries in a year.

Present western literature supports a downward trend in the incidence probably related to maternal folic acid usage. Lumbar meningomyelocele was the most common presentation and this result largely corroborates with studies from Orissa and Davangere in India[8,9]. Shantakumari et al obtained a 67 % incidence of lumbar meningomyelocele as compared to nearly 56 % in current study[5]. If meningoceles were added to this , the incidence of the two lumbar defects approached more than 75 %. However this result contradicts the values obtained by JP Bernard et al[6]. Rachischisis and anencephaly account for very less proportion of all defects partially due to the reasons of non surgical intervention in these defects. The sex ratio of nearly all neural tube defects is in favour of males which might be partially due to female babies with defects not attending tertiary centers as a result of poor awareness. Kulkarni et al in his detailed study from Davangere, India however tilted the sex ratio in favour of females though the difference was minimal[3,10].

The parents of these babies attending our set up were mostly from below poverty line status. Studies undertaken by ShawGM.et.al ,R.Schultz.et.al ,D.A.Hansen,N.N.Dissanayak,S.Halwachs have all pointed towards low socio economic status to be an associated factor[7,9,10,11]. The religion ratio is tilted towards Hindus but this might be similar to the population strata. There are reports citing neural tube defects to be more

common in muslims citing consanguinity as a cause[3,12,13]. Most mothers presented in the age group of 20 to 29 years. However, rachischisis and anencephaly were equal or more common in young mothers less than 20 years age. There are conflicting reports relating the maternal age and parity to the neural tube defects[14,15]. We did not study parity but the clinical history suggested an increased occurrence of miscarriages and other birth defects in babies born to these mothers. Studies by Menasinkai et al and Golalipour et al have considered 20 to 24 years to be the maternal age group with highest presentation[11].

Finally, an association that was not taken in this study was the synchronous occurrence of other birth defects in these babies. These defects ranged from club foot, congenital dislocation of hip, CTEV, contractures and lung hypoplasia. Some of them were responsible for early mortality of these patients.

V. Conclusion

Neural tube defects are common presentation demanding surgical correction in majority of cases. The incidence in India is still high as compared to Western population. Better awareness of the parents and proper antenatal care will go a long way in prophylactic care of this congenital anomaly.

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