ORIGINAL ARTICLE PREVALENCE AND MANAGEMENT OF ANENCEPHALY AT SAIDU TEACHING HOSPITAL, SWAT

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Background: Anencephaly is a fatal congenital abnormality characterised by absence of cerebral hemisphere and cranial vault. Overall incidence of an encephaly is 3.8–6.0/1,000 births. This study was conducted to determine the prevalence, diagnosis and management of an encephaly in District Swat. Methods: This Prospective study was carried out over a period of one year from January 2007 to December 2007 at Saidu Teaching Hospital/Saidu Medical College Swat. Pregnant women having foetuses with neural tube defects (NTDs) admitted in Labour Room/Obstetric Ward of the Saidu Teaching Hospital Swat were selected as subjects. Those women who delivered normal babies were taken as control subjects. A proper history was taken from each subject and all the relevant information were recorded on a proforma. Results: A total of 5,560 subjects were included in this study. Out of them, 69 were having NTDs, 63 had anencephaly, 4 had spina *bifida*, and 2 foetuses were having multiple neural tube defects. Prevalence of an encephaly was 11.33/1000 births and that of *spina bifida* was 0.72/1000 births. Eighty percent (80%) anencephaly and 50% spina bifida were diagnosed by ultrasonography. Most of the cases (52, 75.36%) were delivered through normal vaginal delivery after induction. Twelve (17.40%) had spontaneous vaginal delivery, 4 (5.80%) had caesarean section, and hysterectomy was done in 1 (1.45%). Conclusion: Anencephaly is common congenital anomaly in Swat. Complications can be prevented by implementing screening program for early diagnosis, treatment, and management. Prevalence can be decreased by folic acid supplementation periconceptionally.

Keywords: Anencephaly, Neural tube defects, prevalence, *spina bifida*

INTRODUCTION

Anencephaly is a fatal congenital developmental abnormality characterized by absence of cerebral hemisphere and cranial vault.^{1,2} Approximately 80% foetuses die before birth and none survive beyond two weeks post-natally. Overall incidence of anencephaly is 3.8–6.0 per 1,000 births with considerable variation throughout the world.³ In neonates the anomaly is more frequent in females than males.⁴

Anencephaly as well as *spina bifida* has multi factorial aetiology as some genetic and environmental factors like diet are said to be involved.⁵ A non-genetic factor (Nutritional) has a marked influence on prevalence of neural tube defects.⁶ A large protective effect of folic acid supplementation on recurrence of NTDs has been demonstrated.⁷ Known risk factors for NTDs are maternal age, weight, racial factors, history of previous affected child, un-controlled diabetes mellitus and intake of drugs like anti epileptics.⁷

The commonest associated malformations with an encephaly are *spina bifida*, cleft lip, cleft palate and club foot. Omphalocele has also been described.⁸ An encephaly was the first congenital anomaly identified *in utero* with ultrasound. The diagnosis relies on the failure to demonstrate the cranial vault. Also has frog like appearance and usually have short neck (bulging eyes and large tongue).⁸

The diagnosis can probably be made as early as the 12–13 weeks on ultrasonography.² Prevention and management of anencephaly include dealing with known risk factors and prenatal counselling of affected couple regarding termination of pregnancy.⁹ Birth prevalence of NTDs has continued to decline due to early ante natal diagnosis and elective termination of affected pregnancies.¹⁰ Peri-conceptual use of folic acid reduces first occurrence as well as recurrence of NTDs.¹¹

The present study was conducted to find out the prevalence, predisposing factors and its birth management in our region.

MATERIAL AND METHODS

This prospective study was conducted in Gynaecology Unit of Saidu Teaching Hospital, Swat for a period of one year from 1st Jan 2007 to 31st Dec 2007. During this period 5,560 babies were delivered and were included in this study. The subjects for the study were selected from the women admitted through emergency or through outpatient department, labour room, and obstetric ward of the hospital. The cases were diagnosed as anencephalic foetus based on clinical evaluation and ultrasonography. Those women who were carrying a neural tube defect foetus were taken as subjects and the women who were having normal babies delivered were taken as control subjects. A proper history was taken from each subject and all the relevant informations were recorded on a proforma. They were specifically asked about intake of folic acid and any other drugs. Past history of neural tube defect pregnancy was taken into account.

The diagnosis of the neural tube defect in the current pregnancy was investigated. The diagnostic procedure for neural tube defect was recorded. Complications which occurred in the present pregnancy were noted.

RESULTS

A total of 69 babies had NTDs out of 5,560 deliveries. The total prevalence being 12.41/1,000 deliveries. Sixty-three (63) out of 69 foetuses had anencephaly with the prevalence of 11.33/1,000 births. Four (4) had *spina bifida* and 2 foetuses had multiple defects. The prevalence of anencephaly was maximum at the age of 30–35 years of women, i.e., 20 (31.75%) or 3.60/1,000, and lowest in women of 36 years and above, i.e., 9 (13.05%) or 1.62/1,000 (Table-1).

Among 69 pregnant patient 14 were primigravida, 31 were multigravida and 24 were grand multigravida. Sixty (87%) patients with NTDs belonged to poor socioeconomic class. Four 4 (5.80%) patients had previous history of NTDs and 10 (14.50%) women had consanguineous marriage. Intake of sodium valporate were noted in 3 (4.35%) of patients. Eightyseven percent of the patients were from poor socioeconomic class.

Majority of neural tube defects (53, 76.82%) were diagnosed by ultrasonography, 12 (17.40%) were diagnosed during labour and 4 (5.80%) during caesarean section. The sensitivity of ultrasonography for NTDs was highest (80%) for anencephaly, and lowest (50%) for *spina bifida* (Table-2).

Twelve (17.40%) patients had spontaneous vaginal delivery, 52 (74.36%) had vaginal delivery after induction. Four 4 (5.80%) patients had caesarean section and 1 (1.45%) had subtotal abdominal hysterectomy (Table-3). Among 69 affected foetuses 28 (40.5%) were male and 41 (59.42%) were female.

DISCUSSION

Prevalence of anencephaly in our study was 11.54/1,000 deliveries, whereas in another Pakistani study prevalence was reported to be 3.2–13.9/1,000.^{1,12} Incidence of anencephaly in UK, USA, Denmark, and Oman is around 1–5/1,000 deliveries.^{13–15} This higher incidence could be multifactorial, dietary deficiency of folates, multiparity, consanguineous marriages, failure of early diagnosis and termination of affected pregnancies and referral to tertiary care hospital from periphery.^{1,12}

Women who belong to poor socioeconomic class were more prone to have babies with NTDs. This predisposition may be due to lack of proper diet which may be deficient in folic acid. A survey conducted in UK, where Indian and Pakistani women aged 16–50 years had significantly lower concentration of red cell folates as compared to general population.¹⁶ In our study the relative risk was higher (8.29) for development of NTDs in women who had not taken folic acid.

In this study prevalence of an encephaly has increased up to 35 years of maternal age, which did not correlates with maternal age specific prevalence of NTDs in UK.¹⁷ The exact reason for this is unclear.

The sensitivity of ultrasound screening overall is higher than other screening test. Primary ultrasound screening achieved 100% sensitivity for anencephaly.¹⁷ In this study anencephaly was diagnosed with ultrasonographically by 80%.

Intake of antiepileptic drugs such as sodium valporate increase the risk by 1-1.5%.^{18,19} In our study 4.35% of patients had history of intake of antiepileptic drugs.

More foetuses were of female sex among foetuses with an encephaly as observed by others and it is comparable with this study but is in contrast to those of Eurocat study in which no difference was noted.^{20–22}

Table-1: Flevalence of N1DS with Maternal Age per 1,000 bit ths					
Type of NTDs	<25 yrs	25-29 yrs	30-35 yrs	36-40 yrs	Total
Anencephaly	18 (3.24/1,000)	16 (2.88/1,000)	20 (3.60/1,000)	9 (1.62/1,000)	63 (11.33/1,000)
Spina bifida	1 (0.18/1,000)	2 (0.36/1,000)	1 (0.18/1,000)	-	4 (0.72/1,000)
Multiple		1 (0.18 /1,000)	1 (0.18/1,000)	-	2 (0.36/1,000)

Table-1: Prevalence	of NTDs with Maternal	Age per 1.000 Births
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Table-2: Diagnosis of NTDs					
Congenital Malformation	Cases	Ultrasonography	In Labour	Caesarean/Others	
NTDs	69	53 (76.81%)	12 (17.39%)	4 (5.80%)	
Anencephaly	63	50 (79.37%)	10 (15.87%)	3 (4.35%)	
Spina bifida	4	2 (50%)	1 (25.0%)	1 (25.0%)	
Multiple	2	1 (50.0%)	1 (50.0%)	0 (0%)	
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Table-3: Management (Mode of Delivery) In NTDs

Termination of pregnancy (Induction)	Spontaneous delivery	Spontaneous abortion	Caesarean Section	Hysterectomy
52 (75.36%)	12 (17.40%)	NIL	4 (5.80%)	1 (1.45%)

CONCLUSION

Anencephalies are common congenital anomalies in our setup. Improved maternal services and early diagnosis and termination of affected foetuses will help to reduce the prevalence and complications of the anencephaly.

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