"FREQUENCY AND MANAGEMENT OF ANENCEPHLY AT SHEIKH ZAYED HOSPITAL, RAHIM YAR KHAN"

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ABSTRACT

Anencephaly is the commonest neural tube defect. The world incidence is 1-5/1000.

Objective: To ascertain the frequency and management protocol of Anencephaly at Sheikh Zayed Hospital, Rahim Yar khan.

Materials and Methods: This prospective study was carried out over a period of one year from January, 2013 to December, 2013 at Sheikh layed Hospital, Rahim Yar Khan. All the patients having fetuses with anencephaly admitted in labour room / obstetric ward of Sheikh layed Hospital 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 analyzed on SPSSversion 16.

Results: Twenty one patients were enrolled during the study period. The mean age was 30.5 years range (18-40). Twelve patients (57.14%) were multigravida. Nineteen patients (90.47%) were unbooked. The mean gestational age was 28+2weeks. Induction was palned in 85.71%. Most of the cases 71.43% were delivered through normal vaginal delivery after induction. Additional malformation such as meningocele, cleft lip, spina bifida, cleft palate hepatosplenomegaly, talipes aquina valgus occur in 42.86% cases.

Conclusion: An encephaly is the most common congenital anomaly reported in Sheikh laved Hospital, Rahim Yar Khan. Complication can be prevented by implementing screening program for early diagnosis, treatment and managements. Prevalence can be decreased by folic acid supplementation periconceptionally.

Keywords: Anencephaly, folic acid supplements, Nerual tube defects.

INTRODUCTION

Anencephaly is the absence of a major protion of the brain skull and scalp that occurs during embryonic development 1. It is a cephalic disorder that results from a neural tube defect that occurs when the rostral (head) end of the neural tube fails to close, usually between the 23rd and 26th days of conception 2. The defect is almost always continuous with an open cord in the cervical region. The vault of the skull is absent. The eyes bulge forward, the neck is absent and the surface of the face and chest form a continuous plane, since the fetus lacks the central mechanism for swallowing and the last two months of pregnancy are commonly characterized by polyhydramnios. Anencephaly is a common abnormality (1:100) and is seen four times more frequently in females than males fetuses, similarly it is seen four times more frequently in whites than in blacks 2,3. Anencephaly is uniformly lethal (50% of

infants with this anomaly are still born and only 5% survive for more than one week. Termination of pregnancy is appropriate at any stage 3. The cause of anencephaly is doputed. In general, neural tube defects do not follow direct patterns of heredity, though there is some indirect evidence of inheritance and recent animal models indicate a possible association with deficiencies of the transcription factor TEAD2 4. Studies show that a woman who has had one child with a neural tube defects such as an encephaly has about 3% risk of having another child with a neural tube defects. It is known that women taking certain medications for epilepsy and women with insulin dependent diabetes have a higher risk of having a child with a neural tube defect 5. Genetic counseling is usually offered to women at a higher risk of having a child with a neural tube defect to discuss available testing 6. Recent studies have shown that the addition of folic acid to the diet of

women of child bearing age may significantly reduce, although not eliminate, the incidence of neural tube defect 6,7,8. Therefore it is recommended that all women of child bearing age consume O.4mg of folic acid daily 9,10. An obstetrician may prescribe even higher dosage of folic acid (4mg/day) for women having had a previous pregnancy with neural tube defect 11. Anencephaly can often be diagnosed before birth by maternal serum alpha-Fetoprotein (AFP) and detailed fetal ultrasound 12.

The diagnosis can probably be made as early as the 12 - 13 weeks on ultrasonography 13. The present study was conducted to find out the frequency, its birth management and morbidity and mortality associated with those management protocols in our region.

Methodology:

This study was carried out at Sheikh Zayed Hospital, Rahim Yar Khan from 1st January, 2013 to 31st December, 2013. Twenty one patients fulfilling the inclusion criteria were enrolled in the study period. In OPD detailed history, regarding gravidity, parity, presenting complaints, duration of gestation, past history of anencephaly fetuses, number of normal babies, mode of deliveries, family history of anencephalic babies and cousin marriages were inquired. Method of induction was decided according to bishop score. If the bishop score was unfavourable prostaglandins was used for cervical ripening (Bishop score <4). If bishop score was favourable then oxytocin (syntocinon) in 1000ml drip started with the dose off 2m iu/minutes increasing at intervals of 30 minutes according to the strength and frequency of uterine contractions to a maximum of 32 m iu/mint. Fetal outcome included APGAR score, sex of fetuses and additional malformation associated with anecephaly were noted. The results obtained were recorded in SPSS system and statistical analysis was carried out.

Inclusion Criteria:

Diagnosed cases of an encephaly were included in the study.

Exclusion Criteria:

Other neural tube defects such as spina bifida, hydrocephaly meningocele, meningocle, meningomyelocele were excluded because these fetuses had normal skull development.

RESULTS

A total of 6698 subject were included in this study, out of them 21 were had fetuses with anencephaly. Prevalence of anencephaly was 3.1 / 1000 births in Sheikh Zayed Hospital, Rahim Yar Khan. Maximum age of presentation was 35 years i.e. 0.4 / 1000 (Table 1). Out of twenty one patients, three were primigravida, 12 were muligravida (57.14%). Majority were unbooked patients 90.47% and belonged to lower socioeconomic class. The mean gestational age was 28 \pm 2 weeks range was (26-30 weeks) 100% diagnosis of anencephaly on ultrasonography.

85.71% were induced with different methods, out of 18 eleven were induced with prostaglandin E2. One patient with mechanical traction by extraamniotic catheter. Four patients were induced with misoprostole (Table II). Majority of fetuses 76.19% were female (Table III). Additional malformation such as meningocle cleft lip, spina bifida, cleft palate, hepatosplenomegaly, Talipses aquina valgus occur in 42.86% (Table IV). Morbidity observed in 33.33% such as retained placenta 9.52% fever 4.76%, postpartum haemorrhage 4.76% and polyhydramnios 14.29% (Table V).

Character	Subgroup	No. of patients	% age
Ages in years	18-25 years	2	9.52%
	26-35 years	18	85.71%
	36-40 years	1	4.76%
Gravidity	Primigravida	3	14.29%
	Multigravida	12	57.14%
	Grandmultigravida	6	28.57%
Mode of admission	Booked	2	9.52%
	Un-booked	19	90.47%
Gestational age in weeks	20-25 weekly	3	14.28%
	26-30 weekly	15	71.42%
	31-35 weekly	1	4.76%
	36-40 weekly	2	9.52%

Table 1:

Character	Subgroup	No. of patients	% age
Induction done in 18 patients 85.71%	Prostaglandin E2	11	52.38%
	Mechanical traction Intra-amniotic catheter	1	4.76%
	Spontaneous labour augmentation with oxytocin	2	9.52%
	Cytotec misoprostole	4	19.04%
Mode of delivery	NVD	15	71.43%
	NVD with episiotomy	2	9.52%
	Elective LSCS	3	14.29%
	Emergency LSCS	1	4.76%

Table 2: Mode of delivery and management of labours

NVD - Normal vaginal delivery

LSCS - Lower segment caesarean section

Table 4: Additional malformation with Annecephaly found in 9 fetuses

Malformation	No. of patients	% age
Maningocele	2	9.52%
Cleft lip	2	9.52%
Spina bifida	2	9.52%
Cleft Palate	1	4.76%
Hepatosplenomegaly	1	4.76%
Talipes Aquino Valgus	1	4.76%

Maternal	Туре	No. of Patients	% age
Morbidity	Retained placenta	2	9.52%
	Fever	1	4.76%
	PPH	1	4.76%
	Hydramnios	3	14.29%
Mortality	Maternal mortality	Nil	0%

PPH – postpartum haemorrhage

DISCUSSION

Frequency of an encephaly in our study was 3.1 / 1000 deliveries. Whereas in another Pakistani studies prevalence were reported to be 3.2 - 13.9 / 1000 14,15. A study in Swat Saidu teaching hospital incidence was 11.54/ 1000 16. Incidence of anencephaly in UK, USA, Denmark and Oman is around 1-5 / 1000 deliveries 5. 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 17. In our study the prevalence of anencephaly was max at the age of 26-35 years i.e. 8.5 / 1000 and lower in women of 36-40 years i.e. 0.4/ 1000. These prevalence did not correlates with maternal age specific prevalence of NTD in UK. The exact reason for this is unclear. The sensitivity of ultrasound screening overall is higher than other screening tests 12. Primary ultrasound screening . achieved 100% sensitivity for an encephaly 10,11,12 .

Majority of the cases 76.19% were female fetuses i.e. similar to other study conducted in SWAT 16. Tanne J Hand Deam SC showed anencephaly and other neural tube defects can be prevented by women who daily consume vitamin supplements containing folic acid 9. In our study women could not take folic acid periconceptionally. As they belonged to lower socioeconomic class, noneducated, has lower income and not used vitamin containing folic acid. 71.42% were diagnosed anencephaly at 26-30 weeks gestation whereas study by James DK Steer PJ early diagnosis and termination at 19.6weeks gestation was found⁵.

CONCLUSION

In Pakistan there are no screening programs for those patients who are high risk for neural tube defect.

Complication can be prevented by implementing screening program for early diagnosis and treatment. Prevalence can be decreased by folic acid supplementation peri-conceptionally.

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