INCIDENCE OF MYELOMENINGOCELE WITH ANENCEPHALY IN VIZIANAGARAM DISTRICT OF NORTH COASTAL ANDHRA PRADESH

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ABSTRACT
Anencephaly is a condition that prevents the normal development of the brain and the bones of the skull. This condition results when the neural tube fails to close during the first few weeks of embryonic development. The neural tube is a layer of cells that ultimately develops into the brain and spinal cord. Anencephaly is caused by abnormality of the neural tube which fails to approximate from the sides at the rostral end between the 20th and 28th days of intrauterine life (IUL). The present study showed an incidence of 5.084 neural tube defects per 1000 births and an incidence of 0.84 cases of Anencephaly associated with myelomeningocele per 1000 births. The present study once again reiterates the significance of periconceptional folic acid supplementation intake. Hence a good nutritional status with supplementation of folic acid will go a long way in the prevention of NTDs in this region of Andhra Pradesh.

Key Words: Neural Tube Defects, Anencephaly, Myelomeningocele and Folic Acid

INTRODUCTION
Congenital malformations are among the most distressing diseases that affect mankind. Anencephaly 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 day of conception, resulting in the absence of a major portion of the brain, skull and scalp. Many investigations of the geographical distribution of anencephaly and defects of closure of the neural tube have been reported. Many of these reports, however, group all meningoceles of the spine and head together. The incidence of spina bifida shows a peculiar geographical pattern. The highest rates were found in Belfast Stevenson et al., (1966) and Dublin Coffey (1955), being 4.69 and 4.2 per 1,000 births respectively. It is recommended that all women of child-bearing age consume 0.4 mg of folic acid daily, especially those attempting to conceive or who may possibly conceive, as this can reduce the risk to 0.03%. It is not advisable to wait until pregnancy has begun, since by the time a woman knows she is pregnant, the critical time for the formation of neural tube will elapse. In the present instance anencephaly is associated with myelomeningocele which is the most probable cause of still birth. Anencephaly has an incidence of 1 in 1000 still births according to various studies and in the established literature.

MATERIALS AND METHODS
The data of 1000 live births available in the medical records of government and private hospitals of vizianagaram district was reviewed, 180 still born foetuses during the same period were obtained following the protocol and congenital abnormalities were evaluated with emphasis on the incidence of NTDs spanning three years (2009-2012). These were embalmed by standard method and preserved in formalin and their morphological features were studied. Six cases of Anencephaly were observed. Of these two cases were of 24 weeks gestation and the remaining four were of 38 weeks gestation, one of these is an Anencephaly with Myelomeningocele.
Observations
Spina bifida malformations fall into three categories: spina bifida occulta, spina bifida cystica with meningocele, and spina bifida cystica with myelo meningocele. The most common location of the malformations is the lumbar and sacral areas. Myelomeningocele is the most significant and common form, and this leads to disability in most affected individuals. The foetus weighed 2900 grams and the length was 50cms (CROWN-HEEL). Normal limb pattern was observed, the thorax and abdomen were of normal proportions. The anencephaly was very prominent and the spine showed a typical 2 inches long/1 inch wide open mylomeningocele in the lumbar region which is an open type of NTD. There was no skin and the meninges were intact and the spinal cord was visible underneath (Figure 1). Ears were approximated from the pinna to the upper helix. Neck is very short and eyes were exophthalmic (Figure 2). The karyotypes of these entire foetuses were done and were 46xy. Following a dissection of this foetus region wise no associated congenital abnormalities were detected in the viscera.

DISCUSSION
Embryological Considerations
In the human, the first part of the neural tube to close is the region of the caudal rhombencephalon or cranial spinal cord, usually when five pairs of somites are present. Closure of the neural tube occurs
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during a 4- to 6-day period. Although it was previously thought to close in a linear fashion, like a zipper extending cranially and caudally from the point of initial closure, both observations of human NTDs and experimental manipulations in mice suggest that neurulation appears to involve several waves of closure along the craniocaudal neuraxis. The cranial neural tube closes from the coordinated interaction of at least four waves of discontinuous neural tube closure. The spinal cord closes in a linear manner from a caudal wave of neurulation that extends from the point of initial contact at the cranial end of the spinal cord to the posterior neuropore.

Anencephaly (cranioschisis) is the most representative example of a large heterogeneous group of developmental malformations characterized by abnormalities of the central nervous system and surrounding mesodermal structures. Many of these malformations have been known since ancient times and today many can be reproduced experimentally either by the administration of various teratogens or by mechanical means. Some of these malformations, e.g. anencephaly (cranioschisis) and complete cranio-myeloschisis represent the most severe abnormalities known to man; others, e.g. localized open myeloschisis (rhachischisis) and meningomyelocele (cystic rhachischisis), although compatible with life, require extensive surgical and medical management. Miguel Marin-Padilla (1970) and Kulkarni (1989) reported a high incidence of anencephaly cases in his study in Southern India. Neural tube defect (NTD) is the most common congenital malformation of central nervous system. However, its prevalence varies greatly from region to region in the same country and in populations of different countries. The highest incidence of NTD has been reported from Ireland and Wales (6.38-10.92 per 1000 births), whereas its incidence in other European countries has been only 0.1-0.6 per 1000 births (Charas Suwanwela, 1972). The prevalence of NTD in the US and worldwide is about 1 per 1000 (Charas Suwanwela, 1972). Other parts of the world with high prevalence of NTD are northern India (Coffey and Jessop, 1955), northern China (Dudin, 1997), Egypt and Lebanon (Charas Suwanwela, 1972). Its prevalence in Palestinians too has been reported to be high (Lemire, 1988). Interestingly, compared to Northern provinces in China, the prevalence of NTD in southern China is only 1 per 1000. Its prevalence in Japan also is low. The prevalence of NTD from different parts of India has been reported to vary from 0.5 to 11 per 1000 births (Miguel Marin Padilla, 1970; Kulkarni, 1989; Moore et al., 1997; Agarwal, 1999). The present study showed an incidence of 5.084 neural tube defects per 1000 births and an incidence of 0.84 cases of Anencephaly associated with Myelomeningocele per 1000 births which is consistent with the findings of earlier workers. Anencephaly associated with myelomeningocele of the present study was found to be an extremely rare entity. However less than 1% incidence of NTDS would be an indication of good maternal nutritional status. Consanguinity, parity and maternal age were considered to be precipitant factors resulting in NTDS Kulkarni (1989). However from the present study it is evident that none of the factors mentioned by the earlier workers resulted in the present condition, as the parents were non-consanguinous, mother being primi and of early twenties of age. From the present study it appears that environment did not have a role in NTDS.

Conclusion

Though several reasons and causative factors were implicated in anencephaly the single most consistent etiology remains to be a deficiency of folic acid in the diet of expectant mothers. In this case also the etiology of anencephaly with associated myelomeningocele appears to be folic acid deficiency. Hence a good nutritional status with supplementation of folic acid will go a long way in the prevention of NTDS in this region.

ACKNOLEDGMENT

I dedicate this article to my beloved teacher and mentor Dr. B Narasinga Rao Professor and Head of the Department of Anatomy, MIMS, Vizianagaram whose lectures in neuro anatomy were enthralling.
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