Case Report

Concomitant anencephaly and hydrocephalus in twin

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Abstract
Multiple gestation accounts for about 1 percent of all pregnancies. The major congenital anomaly develops in about 2% of cases in the multiple gestations. The incidence of central nervous system anomalies as hydrocephalus and anencephaly are 0.2% & 0.02% respectively in singleton gestation. Concordant central nervous system malformations in multiple gestation is extreme rarity. A case of prenataly detected concomitant anencephaly and hydrocephalus with meningo-myelocoel in twin pregnancy is being reported.

Introduction
Multiple gestation accounts for about 1 percent of all pregnancies.1 Congenital anomalies are more common, particularly in monochorionic twins, than singletons but in the majority, etiology is not known.2 Neural tube defects (NTDs), including spina bifida and anencephaly, are the second most common birth defect with an incidence of 1/1000.3

Case report:
A rural pregnant woman G5P4L4 aged 32 years presented in our out patient department at seven and half months gestation without any prior antenatal checkups. All previous four babies were live and healthy and delivered vaginally at home. Last child birth was one and half years back. On examination abdomen was over-distended with fundal height of 36 weeks / 37 cm, abdominal girth was 83cm and multiple fetal parts were felt. On sonography twin live fetuses, one anencephaly and another hydrocephalus (fig. 1) of 29 weeks gestational age were detected. There
was no history of iron folic acid tablet intake, any chronic illness, and drug or radiation exposure in this pregnancy. Labor was induced with prostaglandin E1 and she delivered a 1000 gm stillborn anencephalic female as cephalic presentation followed by 1500 gm live female baby as breech with hydrocephaly and meningo-myelocoele vaginally. The baby expired on day 2 of birth. Placenta was diamniotic dichorionic type. Her postpartum period was uneventful and discharged home after 48 hours.

Discussion:
Population based data on Multiple pregnancies and congenital anomalies had found that twins particularly monochorionic twins have higher risk of congenital anomalies. In twins it has been postulated that delayed ovulation or delayed fertilization results in over ripe ova, which are believed to lack cohesion, thus causing splitting of zygote. Such decreased cohesiveness of neural cells produces neural tube defect. Both anencephaly and hydrocephalus have higher incidence in same sex twins compared to unlike sex twins and single births. Occurrence in families and higher association with monozygotic twinning suggest that some Genetic factors contribute to NTD risk.

Anencephaly occurs in 1.4-4.7 per 10,000 deliveries and is thought to be result from failed closure of the anterior neuropore at 24-26 days post fertilization. A baby born with anencephaly is without a forebrain, the largest part of the brain consisting mainly of the cerebral hemispheres, including the isocortex, responsible for higher-level cognition, i.e., thinking. They are usually blind, deaf, unconscious, and unable to feel pain but reflex actions such as breathing and responses to sound or touch are present. Research has suggested that, overall, female babies are more likely to be affected by the disorder.

Congenital hydrocephalus occurs during fetal development and is present at birth. Causes include infections (e.g., cytomegalovirus, toxoplasmosis, rubella) and hemorrhage in the brain. It affects one in every 500 live births and more common in male babies. It may also be a heritable condition and runs in certain families. About 80-90% of fetuses with spina bifida, often associated with meningocele or meningomyelocele, develop hydrocephalus.

The results of the British Medical Research Council’s randomized controlled trial had proved that folic acid can prevent spina bifida and anencephaly. US Public Health Service issued the recommendation that all women of child-bearing age who are capable of becoming pregnant should consume 0.4 mg of folic acid per day to prevent NTD’s. Higher dosage of folic acid (4 mg/day) should be prescribed for women who have had a previous pregnancy with a neural tube defect.

References:

**Figure 1:** abdominal sonography showing anencephalic head in one and hydrocephalic head in second twin