# Spontaneous Pneumocephalus Associated with Open Myelomeningocele

In the present day of antenatal screening, ultrasound, amniocentesis and folate therapy, we still are unfortunate to see babies with open neural tube defects being born occasionally. We saw a 2150 grams girl who was born caesarian section at 35 weeks gestation. An open leaking myelomeningocele was observed on her lumbo- sacral area. Both lower extremities were paralytic. Other systemic examinations were normal. A cranial ultrasonography showed dilated both lateral ventricles and hyperechoic materials in frontal horns of lateral ventricle, which suggested air. A cranial computerized tomography (CT) scan revealed apparent hydrocephalus and pneumocephalus on lateral ventricles. A considerable brain edema was also present (*Fig. 1*). *Staphylococcus aureus* was grown in blood and CSF cutlure. Teicoplanine (10 mg/kg/d) and amikacin (15 mg/kg/d) were started intra-venously for the treatment of meningitis and sepsis. The patient died at the 14th day of therapy.

Pneumocephalus (pneumocranium) means air in cranium. It is associated with several neurosurgical procedures, lumbar puncture, or cranial trauma and rarely with open neural tube defects(1). Spontaneous pneumocephalus has been previously reported in four cases(2-5) in babies with hydrocephalus and open sacral myelomeningocele.

An *X*-ray film, cranial ultrasound or CT scan can detect pneumocephalus. Cranial ultrasonography can pickup smaller amounts of air than conventional *X*-rays.

Massive pneumocephalus can increase the intracranial pressure and behave as life threatening condition. Drainage of air by a needle or a subdural drain may be performed(1). We performed transencephalic puncture for three times for meningitis and drainage of pneumocephalus.

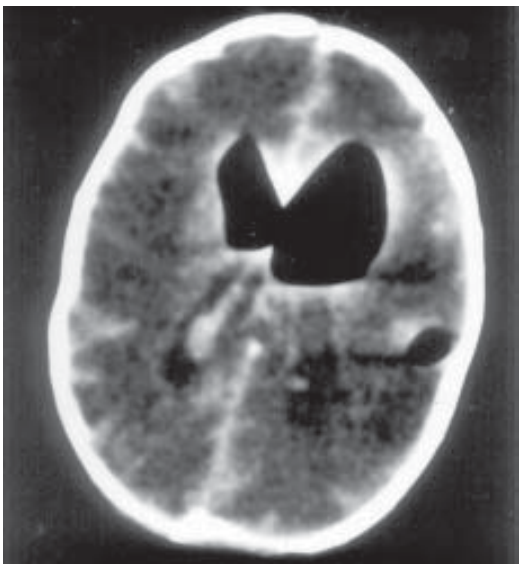

Fig. 1. Nonenhanced Cranial CT Scan shows Prominent air (pneumocephalus) in the frontal horns of lateral ventricles and left sylvian fissure.

Myelomeningocele is usually complicated with intracranial infections such as meningitis and ventriculitis. Spontaneous pneumocephalus should be kept in mind as a rare complication of open meningomyelocele. Any patient with open meningomyelocele should be carefully evaluated for pneumocephalus with a cranial USG. This information is useful prognostically as well as has therapeutic implications.

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## Duchenne Muscular Dystrophy in Monozygotic Twins

Duchenne muscular dystrophy (DMD), an X-linked disorder, is a rare occurrence in monozygotic twins, previously reported on five occasions(1-5). We report a rare case of DMD in a pair of monozygotic twins confirmed by DNA analysis for both monozygosity and mutation in the dystrophin gene.

Five-year-old twin boys were admitted to our hospital with history of progressive difficulty in walking, climbing stairs and frequent falls. There was delay in the attainment of motor milestones. Both boys had bilateral calf muscle hypertrophy with weakness of proximal muscles of lower and upper limbs. Serum creatinine kinase levels were grossly elevated (>11,000 IU/L) in both boys, and muscle biopsy showed dystrophic changes. DNA analysis for mutation detection in the dystrophin gene revealed intragenic deletion of exons 45-48 in both children. The monozygosity was confirmed by DNA analysis. Mother's CPK level was within normal limits. Four generation pedigree revealed no other involved member in the family suggesting that the deletion in the dystrophin gene represented a new mutation.

The rare occurrence of such a genetic disorder in monozygotic twins gives us an opportunity to study to what extent genetic and environmental factors control the different manifestations of the disease.

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