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**SHORT REPORT**

# Identical Twins Presenting With Hydrocephalus

Ahmet ÇOLAK, Murat KUTLAY, Burak O. BORAN, Nusret DEMİRCAN
Department of Neurosurgery, GATA Haydarpaşa Training Hospital, İstanbul - Turkey

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Congenital hydrocephalus is an etiologically diverse disorder, including infectious, teratogenic, and genetic factors as the underlying mechanism (1). Information regarding the occurrence of hydrocephalus in twins is especially important in establishing the significance of environmental factors as well as the genetic basis (2). In this paper we report a pair of monozygotic twins, both presenting with hydrocephalus, but one with triventricular and the other with tetraventricular hydrocephalus.

## Case Report

Identical female twins were delivered to a gravida-1, para-2 mother, by cesarean section at the 32nd week of gestation. Neither abnormal episodes nor histories were observed during the pregnancy. Weights at birth were 1490 g and 1710 g. Body lengths were 39 cm and 41 cm. Head circumferences were 36 cm and 37 cm, which were both more than 2 standard deviations above the estimated values for the gestational age.

Twin A experienced respiratory insufficiency at birth, and was therefore admitted to the ICU and connected to a ventilator. She had a fever. Cultures revealed urinary tract infection and she was put on appropriate antibiotics. Cerebrospinal fluid (CSF) and blood cultures were sterile. She was disconnected from the ventilator on the third day. Her head circumference increased 1 cm in 3 days and the anterior fontanelle was bulging. Cranial ultrasonography revealed ventricular enlargement with no sign of hemorrhage. Computerized tomography was performed (Figures 1a and b), and triventricular hydrocephalus was demonstrated. Soon after, the patient experienced a convulsion. Phenobarbital infusion was initiated and emergency surgery was performed to insert a ventricular reservoir. Intermittent CSF removal was performed for about 2 weeks, until her general status improved and she was free of fever. Afterwards she was operated on and a ventriculoperitoneal shunt was inserted while the ventricular reservoir was removed.

Twin B experienced an uneventful delivery. Her physical examination was within normal limits except for a bulging anterior fontanelle. Cranial ultrasonography was performed and hydrocephalus was noted. Computerized tomography (Figure 2a and b) demonstrated the dilatation of all 4 ventricles, but the subarachnoid space was not widened. She was operated on and a ventriculoperitoneal shunt was inserted.

## Discussion

Hydrocephalus can be briefly described as the abnormal accumulation of CSF in the cranium (3). Although the underlying cause in acquired hydrocephalus has generally been determined, the etiology of congenital hydrocephalus is not well established (4). It may be associated with chromosomal abnormalities (trisomy 13, trisomy 18, triploidy, etc.), Mendelian conditions

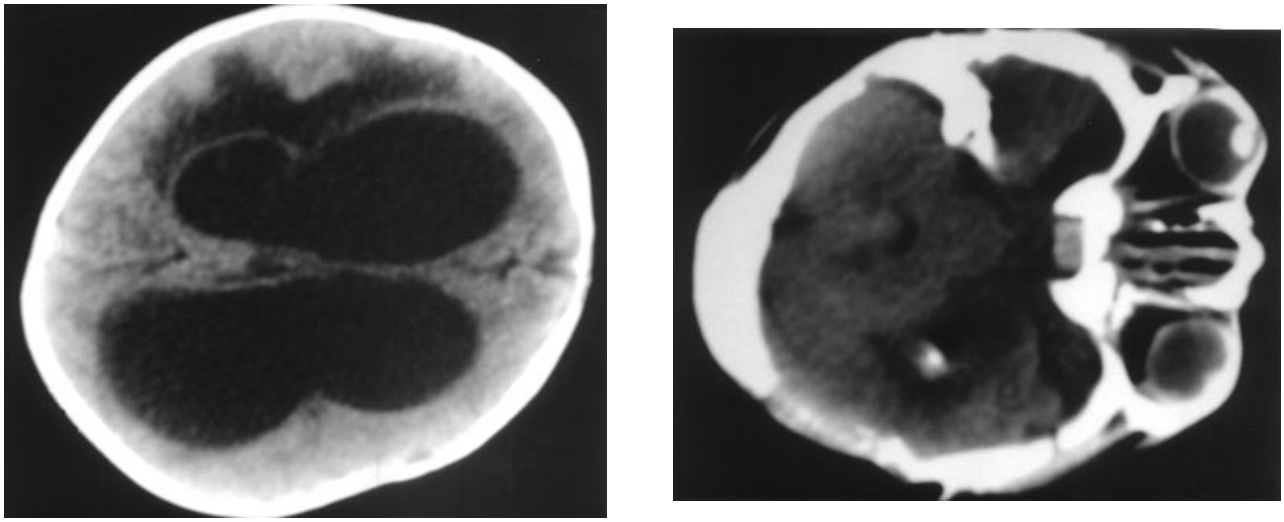

Figure 1a, b. Computerized tomography of twin A demonstrating triventricular hydrocephalus. Note that the 4th ventricle is small.

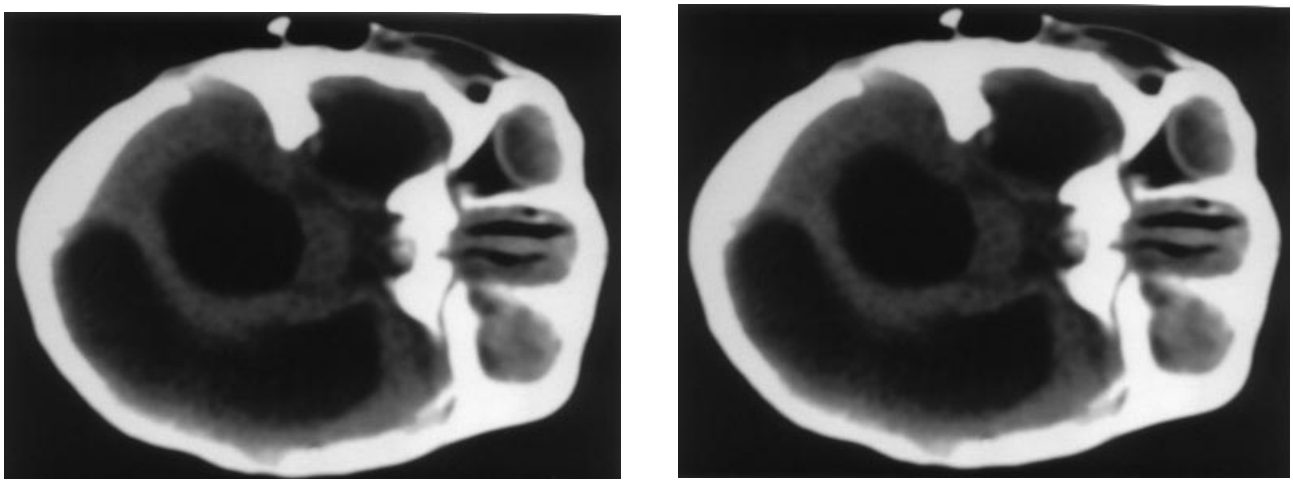

Figure 2 a, b. Computerized tomography of twin B demonstrating tetraventricular hydrocephalus. The 4th ventricle is large but the subarachnoid space is not widened.

(Walker-Warburg syndrome, Meckel syndrome, Fanconi anemia, etc.), other central nervous system (CNS) malformations (Dandy-Walker malformation, Chiari malformation, neural tube defects, etc.) or it may be isolated (5). In the cases presented, no genetic studies could be carried out because of technical difficulties, but there were no recognizable patterns of malformation.

In isolated cases of congenital hydrocephalus, a genetic basis is strongly suggested (6-9). Information regarding the occurrence of hydrocephalus in twins is therefore important. Wachi et al. (9) reported a pair of identical twins presenting with external hydrocephalus. Neuroimaging, doppler sonography, and intracranial pressure (ICP) monitoring were performed, delineating the anatomical and biomechanical similarities of the intracranial space, and thereby noting the importance of the genetic basis. On the other hand, Piatt (10), reporting monozygotic twins discordant for external hydrocephalus, stated that, whatever the genotype of the twins, their different phenotypes show that hydrocephalus is not set in motion during a critical window of fetal development.

Imaizumi (2) reported 107 twin pairs from Japan, of which at least 1 in each pair had congenital hyrocephalus. The rate of concordance for hydrocephalus among these twins was 15%. The rate was 28% in like-sex male twins, probably due to X-linked hydrocephalus with aqueductal stenosis. Similarly, Record et al. (11) reported 17 twin pairs and the rate of concordance for hydrocephalus was 11.8%. Aside from research on twins, Lorber (7) carried out a longitudinal study on 270 babies with isolated congenital hydrocephalus; they had 453 siblings of whom 5 were hydrocephalic (1.1%).

The role of environmental factors in the pathogenesis of congenital hydrocephalus is also well documented in the literature (5). Prenatal infections such as toxoplasmosis, rubella, syphilis and cytomegalovirus seem to demonstrate the strongest association (8). The risk of hydrocephalus may be increased by the maternal consumption of drugs containing misoprostol, dextromethorphan, nalidixic acid, cephalosporins, etc. (12,13). Maternal smoking does not appear to affect hydrocephaly rates (14). Although the association between hydrocephaly and maternal diabetes is not clear, a recent study demonstrated an increased risk among infants born to obese but non-diabetic women (15). Interestingly, hydrocephaly risk seems to be lower in infants born to women living at high altitudes (16).

In conclusion, congenital hydrocephalus seems to be a multifactorial disorder, triggered by environmental factors in genetically predisposed individuals.

*Corresponding author:*
*Ahmet ÇOLAK*
*GATA Haydarpaşa Teaching Hospital,*
*Tıbbiye Cad, 81327, Haydarpaşa*
*Üsküdar, İstanbul - Turkey*
*e-mail: ahmetcolak@ixir.com*

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