

## Fetal Alcohol Syndrome with Hydrocephalus

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### ABSTRACT

Exposure to alcohol in the fetal period leads to various structural abnormalities and this is called as fetal alcohol syndrome. Cranial and facial anomalies are the common features of fetal alcohol syndrome. Microcephaly is frequently associated with fetal alcohol syndrome, but hydrocephalus may be rarely seen.

The female baby at a gestational age of 33 weeks and with a weight of 2000 g was born prematurely at our hospital from a mother of a foreign nationality. The baby had dysmorphic features and macrocephalus. The mother drank 1 L alcohol per day during pregnancy. The baby was admitted to the neonatal intensive care unit for prematurity, hydrocephalus, and fetal alcohol syndrome.

In our country, alcohol use in women, particularly pregnant women, is a very rare condition because of community rules and beliefs. In this report, we highlighted fetal alcohol syndrome and its coexistence with hydrocephalus.

**Keywords:** Fetal alcohol syndrome, hydrocephalus, prematurity

### Introduction

In recent years, the prevalence of the use of alcohol has been increasing among women (1). The use of alcohol among pregnant women and women in the reproductive age group is an important public concern in the United States of America (2). The rate of planned pregnancy is low among women living in the United States of America, and most women drink alcohol without being aware of their pregnancy. The prevalence of alcohol consumption among parturients has been reported to vary between 12% and 15% in recent years. Prenatal alcohol exposure is a frequent teratogenic cause of developmental disorders, congenital defects, and mental diseases (3).

### Case Presentation

A female baby weighing 2000 g was born through normal spontaneous vaginal delivery at the 33<sup>rd</sup> gestational week in the first pregnancy of her 20-year-old mother. The baby was macrocephalic, and her head circumference was 45 cm (>95 percentile). Her face was dysmorphic, and she had micrognathia, a smooth philtrum, and a thin upper lip (Figure 1). The patient was hypoactive, and her newborn reflexes were poor. She could not close her eyelids, and her cornea was blurred. It was learned that her mother, who was of foreign nationality, drank approximately 1 L of alcohol per day before and during pregnancy. The baby was admitted to the neonatal intensive care unit for the pre-diagnoses of prematurity and fetal alcohol syndrome (FAS).

In her laboratory evaluation, complete blood count and biochemistry values were normal. Moreover, the results of examinations performed for intrauterine viral infections (TORCH infections) were found to be normal. No numerical and structural chromosome anomaly was observed in the chromosome analysis (46XX). On cranial computed tomography, ventricles were observed to be severely dilated and the corpus callosum could not be viewed. The brain parenchyma could be monitored in partially frontal area (Figure 2). The result of the echocardiographic examination was normal. The neurosurgery department was consulted, and ventricular tapping was performed. The mother rejected the insertion of a ventriculoperitoneal shunt in the baby. In the evaluation performed by the department of ophthalmology, bilateral exposure keratopathy, bilateral central corneal opacity, and corneal edema were detected. On the 17<sup>th</sup> day of the follow-up period, the baby had cardiac arrest and became exitus.

Necessary permission was received from the mother for using the pictures of the baby.

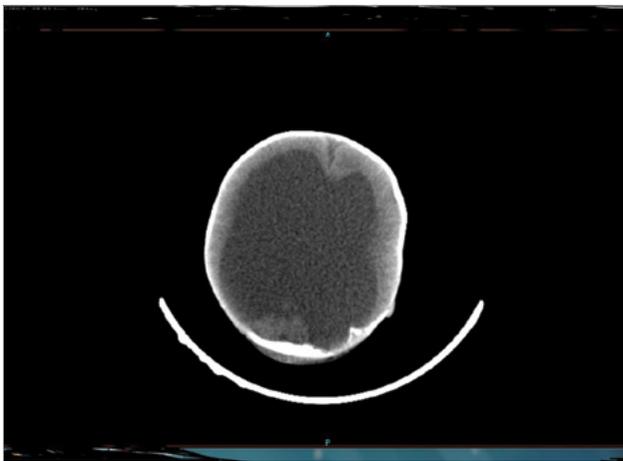
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Received : 11.01.2015  
Accepted : 15.06.2015

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**Figure 1.** Facial appearance of the hydrocephalic baby with fetal alcohol syndrome



**Figure 2.** Cranial computed tomography image of the hydrocephalic baby with fetal alcohol syndrome

## Discussion

Alcohol addiction in mothers causes abnormal fetal development; this condition is defined as FAS. FAS is characterized by central nervous system dysfunction, pre- and post-natal growth retardation, cranial and facial anomalies, and organ anomalies. When the central nervous system is involved, the most serious disorders are mental retardation, motor dysfunction, and ataxia. Exposure to alcohol during the intrauterine period is the most commonly known cause of mental retardation in the United States of America.

The prevalence of fetal alcohol spectrum disorders is 9.1 per 1000 live births (4). Recently, it has been determined that prenatal alcohol exposure is an important factor for mortality during the fetal period, infancy, and childhood (3).

FAS was first defined by Jones and Smith in 1973 (5). Prenatal alcohol exposure is a cause of congenital brain anomalies (6). Hydrocephalus is an uncommon anomaly among patients with FAS; microcephaly is more frequently seen (7). The causes and neuropathological aspects of hydrocephalus

are not well known. The most common and well-defined type of hydrocephalus is X-linked, and it is related to the mutation in L1 gene coding region. Brain dysfunction and malformations associated with FAS occur in patients having a mutation in this region. Ethanol is teratogen, and it has been revealed that it affects the brain by blocking the molecular interaction formed by L1 (8). The exposure of fetal cells to ethanol can lead to dysmorphic features (smooth philtrum, thin upper lip, small palpebral fissures, upturned nostrils, and microcephaly), other congenital anomalies, neurodevelopmental anomalies, and neuroanatomic defects (4).

The most harmful form of alcohol consumption for the fetus is heavy episodic drinking. In babies of pregnant women who continue to drink alcohol during pregnancy and consume more alcohol than usual, the occurrence of FAS with severe dysmorphic features is highly probable. Moreover, the age and social status of mothers can affect the development of FAS (9). In children with FAS, neuronal–glial heterotopias, cerebellar dysplasia, corpus callosum agenesis, and microcephaly from neuropathological abnormalities can be encountered (10). In our case, the mother had a history of heavy alcohol consumption during pregnancy, and severe FAS findings were detected in the baby.

## Conclusion

In Turkey, the use of alcohol among pregnant women is uncommon. Therefore, we could not find a study conducted on this issue in Turkey in literature. Moreover, in literature, there are a few studies on cases of FAS coexisting with hydrocephalus. With this case, we aimed to highlight FAS, which is rarely found among the causes of hydrocephalus in Turkey.

**Informed Consent:** Written informed consent was obtained from the patient's parents.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Concept - B.T.B., S.B.; Design - B.T.B.; Supervision - S.B.; Funding - M.H.; Materials - M.K.; Data Collection and/or Processing - M.H., M.K.; Analysis and/or Interpretation - S.B.; Literature Review - M.H., M.K.; Writer - B.T.B.; Critical Review - S.B.

**Conflict of Interest:** The authors declared no conflict of interest.

**Financial Disclosure:** The authors declared that this study has received no financial support.

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