

Schizencephaly: a rare congenital disorder.

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Schizencephaly is a rare congenital disorder of neuronal migration characterized by the presence of clefts, with heterotopic gray matter, extending from the ependymal surface of the lateral ventricles to the piamater of the cortex. The term originates from the Greek word *schizein*, meaning "to divide," and was introduced by Yakovlev and Wadsworth in 1946. It can be classified into two types: closed-lip and open-lip. The typical triad of most neuronal migration disorders is: motor disorder, intellectual disability, and seizures.

A 29-year-old, primigravida patient presented to the fetal medicine department in the third trimester. She had no relevant medical history. She denied any drug intake. The pregnancy was unplanned but desired, and had taken multivitamins from the beginning of pregnancy. At the ultrasound (image 1.0) the interhemispheric line cannot be delimited due to the presence of severe asymmetric bilateral ventriculomegaly, a hypoechoic clearance in the left frontal region that communicates with the anterior horn of the lateral ventricle, multiple parenchymal calcifications, probable left open lip schizencephaly, subarachnoid cyst and macrocranium.

Conclusion

The etiology of Schizencephaly can be acquired or genetic. The most common cause is vascular disruption and hypoxia during critical periods of neuronal development. It is a congenital malformation characterized by producing a chronic motor disorder. The prognosis is very poor due to severe developmental delay and difficult-to-control epilepsy.

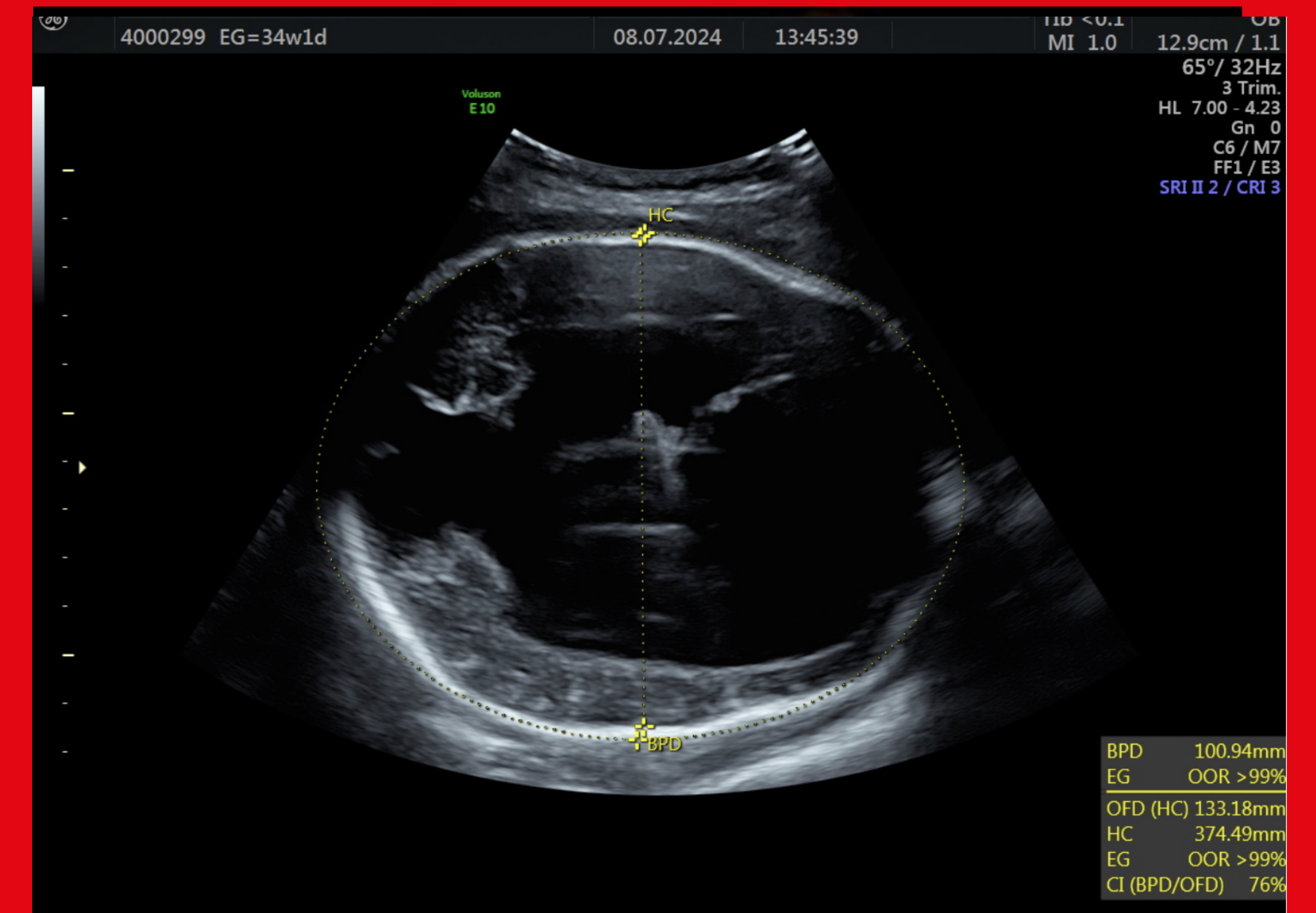


Figure 1. Schizencephaly