Middle interhemispheric variant of holoprosencephaly

Marcela Posada · Mauricio Castillo

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A girl was born at gestational week 34 from a pregnancy complicated by pre-eclampsia and insulin-dependent diabetes mellitus. Multiple fetal anomalies were found on US (holoprosencephaly, cardiac situs inversus, ventricular septal defect and polyhydramnios). Fetal (Fig. 1) and postnatal (Fig. 2) brain MRIs show midline fusion of posterior frontal and anterior parietal regions with callosal dysgenesis and lack of midline thalamic fusion.

Middle interhemispheric holoprosencephaly consists of abnormal midline hemispheric fusion of the posterior frontal and parietal regions. It differs from classic holoprosencephaly because there is interhemispheric separation of the basal forebrain and the anterior frontal lobes [1]. A

Fig. 1 Fetal MRI. Coronal T2-W image

Fig. 2 Postnatal MRI. Axial T2-W image

previous report shows an association between middle interhemispheric holoprosencephaly and a mutation (ZIC2 gene) related to neural tube closure and differentiation of the embryonic roof plate [2]. Patients with middle interhemispheric holoprosencephaly have fewer motor and cognitive disabilities than those with classic holoprosencephaly [2]. Prenatal MRI provides valuable information in counseling of parents [2].

References

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M. Posada · M. Castillo (✉)
Department of Radiology,
University of North Carolina School of Medicine,
222 Old Fayetteville Road,
Chapel Hill, NC 27510, USA
e-mail: mauricio_castillo@med.unc.edu