Syntelencephaly associated with cystic cochleovestibular malformations

Figure 1 Syntelencephaly

(A) Partial intermediate agenesis of corpus callosum is shown. (B) Failure of cleavage of the posterior frontal and anterior parietal regions with normal cortex bridging the midline and absent septum pellucidum. (C, D) Atretic parietal cephalocele, persistent falcine sinus, and azygous anterior cerebral artery.

A 3-month-old boy presented with bilateral profound hearing loss. Examination of ears was unremarkable. Brain MRI revealed syntelencephaly with associated other anomalies (figures 1 and 2).

Syntelencephaly or middle interhemispheric subtype of holoprosencephaly is characterized by abnormal midline connection of the posterior frontal and parietal lobes with normal separation of rostral forebrain.^{1,2} The coexistence of syntelencephaly and other anomalies such as cystic cochleovestibular malformation, persistent falcine sinus, and parietal encephalocele is unknown and raises the question of pathogenic association or coincidence among them.

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(A) Axial T2-weighted image shows incompletely partitioned bilateral cochleas with dysmorphic vestibules. (B, C) Three-dimensional reformatted images show absent left transverse semicircular canal and enlarged and dysplastic right transverse and left superior semicircular canals. Normal right (D) and absent left (E, circle) cochlear nerves.

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