Agenesis of the corpus callosum

Agenesis of the corpus callosum (ACC) is a rare congenital disorder in which there is a complete or partial absence of the corpus callosum. It occurs when the corpus callosum, fails to develop normally, typically during pregnancy. The development of the fibers that would otherwise form the corpus callosum become longitudinally oriented within each hemisphere and form structures called Probst bundles.

General information

A failure of commissuration occurring ≈ 2 weeks after conception. Results in an expansion of the third ventricle and separation of the lateral ventricles (which develop dilated occipital horns and atria, and concave medial borders).

The corpus callosum (CC) forms from rostrum (genu) to splenium, in agenesis there may be an anterior portion with absence of the posterior segment (the converse occurs infrequently). The absence of the anterior CC with the presence of some posterior CC is indicative of some form of holoprosencephaly.

In addition to agenesis of the corpus callosum, other callosal disorders include hypogenesis (partial formation), dysgenesis (malformation) of the corpus callosum, and hypoplasia (underdevelopment) of the corpus callosum.

Differential diagnosis

May occasionally be associated with hydrocephalus, but more often merely represents an expansion of the third ventricle and separation of the lateral ventricles.

Case reports

Kho et al., report a patient with Parkinson's disease in whom imaging revealed a complete agenesis of the corpus callosum. Although this co-occurrence is probably coincidental, this finding suggests that the bilateral degenerative changes in Parkinson's disease may occur independent of the interhemispheric connections ¹).

Tijssen et al. describe the neuroimaging findings of an 11-year-old boy who presented with mild occipital headache and precocious puberty. This child was found to have a combination of various midline anomalies including a Chiari type 1 malformation, corpus callosum agenesis and patent craniopharyngeal canal with adjacent intracranial dermoid cyst ²).


²) Tijssen MP, Poretti A, Huisman TA. Chiari type 1 malformation, corpus callosum agenesis and patent

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