What is Agenesis of the Corpus Callosum?

Agenesis of the corpus callosum (ACC) is a birth defect in which the structure that connects the two hemispheres of the brain (the corpus callosum) is partially or completely absent. ACC can occur as an isolated condition or in combination with other cerebral abnormalities, including Arnold-Chiari malformation, Dandy-Walker syndrome, Andermann syndrome, schizencephaly (clefts or deep divisions in brain tissue), and holoprosencephaly (failure of the forebrain to divide into lobes.) Girls may have a gender-specific condition called Aicardi's syndrome, which causes severe mental retardation, seizures, abnormalities in the vertebra of the spine, and lesions on the retina of the eye. ACC can also be associated with malformations in other parts of the body, such as midline facial defects. The effects of the disorder range from subtle or mild to severe, depending on associated brain abnormalities. Intelligence may be normal with mild compromise of skills requiring matching of visual patterns. But children with the most severe brain malformations may have intellectual retardation, seizures, hydrocephalus, and spasticity.

Is there any treatment?

There is no standard course of treatment for ACC. Treatment usually involves management of symptoms and seizures if they occur.

What is the prognosis?

Prognosis depends on the extent and severity of malformations. ACC does not cause death in the majority of children. Mental retardation does not worsen. Although many children with the disorder have average intelligence and lead normal lives, neuropsychological testing reveals subtle differences in higher cortical function compared to individuals of the same age and education without ACC.

What research is being done?

The NINDS conducts and supports a wide range of studies that explore the complex mechanisms of normal brain development. The knowledge gained from these fundamental studies helps researchers understand how the process can go awry and provides opportunities for more effectively treating, and perhaps even preventing, developmental brain disorders such as ACC.

NIH Patient Recruitment for Agenesis of the Corpus Callosum Clinical Trials

- At NIH Clinical Center
- Throughout the U.S. and Worldwide
- NINDS Clinical Trials

Organizations

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