

Agensis (Dysgenesis) of the Corpus Callosum

Symptoms or Behaviors

Seizures

Abnormal head and facial features

Brain anomalies (e.g. hydrocephalus, Arnold-Chiari malformation, or migration disorders)

Syndromes (e.g., Aicardi or Andermann)

Other genetic disorders

Cognitive impairments

Behavioral or neurobiological disorders (ADHD, OCD, ASD or autistic-like behaviors)

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About the Disorder

Agensis (absence) or dysgenesis (malformation) of the corpus callosum are brain abnormalities involving the large bundle of nerve fibers that connect the two hemispheres of the brain (the corpus callosum). These fibers may be completely absent, partially absent, thin, or malformed in some way. 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.

It is unclear how often disorders of the corpus callosum actually occur since no one knows how many individuals have callosal conditions but are not diagnosed. Current research suggests callosal conditions may occur in as many as 4 persons per 1,000 in the general population. They occur more frequently among persons with developmental disabilities (22-24 per 1,000).

Abnormalities of the corpus callosum can be diagnosed by computerized tomography (CT) scan or magnetic resonance imaging (MRI). They are sometimes detected during pregnancy through routine prenatal ultrasound. When detected prenatally, a CT scan or MRI is often ordered to confirm the diagnosis and obtain additional information.

Causes

Potential causes include chromosome errors or inherited genetic factors, prenatal infections, injuries, or toxic exposures, structural blockages (e.g. cysts), metabolic disorders, or other unknown factors.

Treatment

Callosal disorders are lifelong conditions. Individuals may benefit from early intervention services, supportive therapies, special education, and adult support services based on their individual needs. Evaluations and therapies should begin early in life and continue throughout childhood and into adult life.

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Educational Considerations

There are numerous learner characteristics associated with ACC, many of which are reported more commonly in individuals with callosal disorders. An individual with a callosal condition may have few or many of these potential learning issues. These areas of need can include any of the following: developmental delays, motor delays (may be late to crawl, walk or talk), sensory issues (visual impairments, hearing deficits, increased sensitivity to touch), low muscle tone, poor motor coordination, feeding/eating difficulties, gastric reflux, High tolerance to pain, sleep-related difficulties (e.g. getting to sleep, nighttime waking, bed-wetting) Academically, students with ACC may have difficulties attending to tasks, may have ADHD. They may also experience difficulty with complex tasks, abstract reasoning, and problem solving. Socially and emotionally, they may have difficulty imagining the consequences of their own behavior, and may appear socially immature and lacking in self-awareness. Students with ACC may have difficulty with understanding idiomatic language (slang, sarcasm, metaphors) and reading social cues. Individuals with ACC generally have a pleasant disposition, enjoy being with others, demonstrate a willingness to learn, etc.

Instructional Strategies & Classroom Accommodations

- Modify curriculum and instructional methods as needed, keeping in mind that student may have difficulty with abstract concepts, memory recall, application of new concepts, problem solving, focused attention to tasks, etc.
 - Provide accommodations for fine motor tasks, such as providing alternatives to handwriting, modifying amount of written work, etc.
 - Provide accommodations for large motor tasks that may involve coordinated motor planning
 - Monitor student for possible sensory impairments
 - Check for understanding when giving auditory directions, or when involved in conversations
- Note:** Document all accommodations on the IEP.

Resources

National Organization for Rare Disorders (NORD)

P.O. Box 1968
(55 Kenosia Avenue)
Danbury, CT 06813-1968
orphan@rarediseases.org
<http://www.rarediseases.org>

National Institute of Neurological Disorders and Stroke

NIH Neurological Institute

P.O. Box 5801
Bethesda, MD 20824
<http://www.ninds.nih.gov/>

ACC Network

5749 Merrill Hall Room 337
University of Maine
Orono, ME 04469-5749
UM-ACC@maine.edu
<http://www.umaine.edu/edhd/research/accnetwork.htm>