

Are problems in the areas of cognition and emotion related to the cerebellar damage in Ataxia?

Yes, they can be. The cerebellum has long been known to be important for motor control, and people with Ataxic disorders experience motor difficulties including wide-based and unsteady gait (Ataxia), incoordination of the arms and legs, slurred speech, and eye movement abnormalities. We now know that the cerebellum, like the cerebral cortex, has some parts that are critical for movement, and other parts that play a role in cognition and emotion. The motor cerebellum is linked with the motor cortex that governs movements; whereas, non-motor regions of the cerebellum are linked with cerebral cortical “association” areas important for thought, reasoning, motivation, memory and feelings. Damage to the non-motor cerebellum disrupts these cerebellar links with cerebral cortical thinking areas and can lead to difficulties in intellectual functions and to changes in mood and personality. Ataxic disorders may also produce neurodegeneration in parts of the cerebral cortex important for intellect, which is another reason why ataxia patients may experience problems with these functions.

What types of psychological problems may be caused by cerebellar damage or disorders?

The cerebellar cognitive affective syndrome is the set of problems that arises when the non-motor cerebellum is damaged. It includes impairments in executive function, visual-spatial analysis and selected deficits in language skills as well as changes in personality and behavior. There may be difficulties with multi-tasking, require intellectual flexibility and which were previously performed automatically and may require conscious effort and new strategies.

There may be difficulty expressing thoughts logically and coherently, and memory problems. Mood changes include depression, apathy, irritability and limited frustration tolerance. Psychosocial interaction may be impaired, particularly in children with cerebellar damage.

Dementia is uncommon in cerebellar disease but can be a problem in those ataxic disorders that affect widespread areas of the cerebral cortex. Currently, the role of the cerebellum is under investigation for the following disorders: dyslexia, attention deficit hyperactivity disorder, autism spectrum disorders, schizophrenia, developmental delay and panic disorder.

Why is this important for the Ataxia patient?

Cerebellar patients and families generally find it helpful to know that cognitive and psychiatric problems may occur as a direct result of the illness. These challenges, beyond motor control, are not “in their head”, rather, they are in their brain. These difficulties are not the fault of the affected individual, family members or health care providers. It is also useful to know that the stress of chronic illness and the social changes that result may place additional burdens on patients and families. Recognizing the brain basis of these mental and mood changes, along with challenging social and psychological issues that come with the illness, is the essential first step leading to treatment. Parents of children with developmental delay from cerebellar disease may have to deal with many special psychosocial and intellectual challenges, even if there is only minimal or no Ataxia.

What can be done for the Ataxia patient?

The ultimate answer is to prevent or cure the cerebellar disease itself. Even in the absence of a cure, it is still possible to substantially improve quality of life by treatment of cognitive and psychiatric disorders through therapies such as medication, cognitive rehabilitation, psychological counseling and environmental interventions. Primary care physicians and neurologists should be capable of managing these aspects of care. In more difficult or complicated cases, consultation with an ataxia specialist, behavioral neurologist, or psychiatrist with experience in the treatment of cognitive and mood issues may be necessary. Successful treatment depends on a thorough evaluation in order to use the most appropriate medications and interventions tailored to the needs of everyone.

Children with cognitive and psychosocial challenges need special attention and may do well with time and intensive rehabilitation.

How can the National Ataxia Foundation help?

The National Ataxia Foundation (NAF) is committed to providing information and education about ataxia, support groups for those affected by ataxia, and promoting and funding research to find the cause for the various forms of ataxia, better treatments, and, hopefully someday, a cure. NAF has been at the forefront funding promising worldwide research to find answers.

Ataxia research has moved into the clinical phase, and pharmaceutical companies have begun recruiting participants for clinical trials. Individuals with Ataxia or who are at-risk for Ataxia are encouraged to enroll in the CoRDS Ataxia Patient Registry. To access the Registry, go to NAF's website www.ataxia.org and click on the "Enroll in the Patient Registry" tab and follow the directions on the CoRDS website.

NAF provides accurate information for you, your family, and your physician about Ataxia. Please visit the NAF website at www.ataxia.org for additional information, including a listing of ataxia support groups, physicians who treat Ataxia, social networks, and more. For questions contact the NAF directly at 763/553-0020 or naf@ataxia.org.

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