

*FREQUENTLY ASKED QUESTIONS ABOUT...***Cognition and Emotion in Cerebellar Disorders****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, 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,

planning, and organizing. Tasks of every day life that require intellectual flexibility and that were previously performed automatically may require conscious effort and new strategies. There may be difficulty expressing thoughts logically and coherently, and memory problems, particularly with working, or scratch pad, memory. 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. The role of the cerebellum in dyslexia, attention deficit hyperactivity disorder, autism spectrum disorders, schizophrenia, developmental delay and panic disorder, is under active investigation.

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

Is there on-going research?

Yes. There is a new and vibrant area of research: the cognitive neuroscience of the cerebellum. It includes anatomical and behavioral studies in animal models, as well as functional imaging studies and clinical investigations in patients and healthy controls. The National Ataxia Foundation (NAF) can direct you to the appropriate laboratories if you would like to participate or contribute to these studies. An essential

component for rare disease research is the availability of people with ataxia to participate in research. If you have ataxia or are at risk of having ataxia, you are encouraged to sign-up on the ataxia patient registry. Go to www.nationalataxiaregistry.org to register.

How can the National Ataxia Foundation help?

The National Ataxia Foundation has been assisting families affected by ataxia for over 50 years. NAF is committed to education about ataxia, service to individuals through neurologist referrals, support groups and a comprehensive website that is kept current with information on clinical trials and research in ataxia. Also available on the NAF website are online chat groups, bulletin boards and social networking opportunities as well as links to pertinent and credible websites.

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12/08

Reprinted 7/10