

*FREQUENTLY ASKED QUESTIONS ABOUT...***Spinocerebellar Ataxia Type 14 (SCA14)****What is Spinocerebellar Ataxia Type 14?**

Spinocerebellar ataxia type 14 (SCA14) is one of the many types of hereditary cerebellar ataxias. It is an inherited defect in a gene that causes slow degeneration of cells in the cerebellum resulting in ataxia and incoordination.

What are the symptoms of SCA14?

The most common symptom of SCA14 is incoordination (ataxia) of walking (gait). Other symptoms may include poor coordination of speaking (dysarthria) and tremor of the hands when reaching for objects. A few persons with SCA14 have developed brief shaking episodes of the arms or body referred to as myoclonus. Even less common symptoms have included stiffness of the muscles (rigidity), memory loss late in life, and difficulty swallowing (dysphagia).

What is the prognosis of SCA14?

The symptoms of SCA14 typically begin in mid-life (20s-40s), but childhood onset or later adult onset have also been reported. The condition is slowly progressive and may require a cane, walker, or rarely a wheelchair late in life. Life span is not decreased.

How is SCA14 acquired?

SCA14 is inherited as an autosomal dominant genetic disease. This means that each child of an affected parent has a 50% risk of inheriting the gene mutation and developing the disease. Males and females are both affected. The involved gene is located on human chromosome 19 and codes for a protein called protein kinase C γ (PRKCG). It is not yet known how abnormalities of this protein cause degeneration of the cerebellum.

How common is SCA14?

SCA14 is not common. It represents less than 5% of all of the hereditary ataxias.

How is the diagnosis made?

A neurological evaluation by a physician makes the diagnosis of cerebellar ataxia. A CT or MRI scan of the brain may show atrophy of the cerebellum. However, a specific diagnosis of SCA14 can only be made by a genetic test done on a blood sample. This genetic test is sometimes clearly abnormal, but other times is difficult to interpret. The test results often need to be evaluated by an expert in genetic medicine.

What kind of support is available after the diagnosis?

There is no cure or specific treatment for SCA14. Physical therapy and gait training may be of value. Genetic counseling is important for families with SCA14 and genetic counselors can be identified through the website: www.nsgc.org. Further information about SCA14 can also be obtained from www.genereviews.org.

The National Ataxia Foundation provides a wide range of support materials and activities including the publications, *Living with Ataxia: An Information and Resource Guide* and *Evaluation and Management of Ataxic Disorders: An Overview for Physicians* as well as a 48-page quarterly newsletter with the most up-to-date research and medical information on all the types of ataxia. NAF also provides and participates in many local support groups and maintains the website www.ataxia.org which includes an active Bulletin Board and Chat Room.

Contact the National Ataxia Foundation for a more complete listing of resources and support groups affiliated with the National Ataxia Foundation.

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