

SPINOCEREBELLAR ATAXIA TYPE 11 (SCA11)



Spinocerebellar ataxia type 11

Spinocerebellar ataxia type 11 (SCA11) is a type of inherited cerebellar ataxia, caused by an abnormal gene. It results in damage and wasting away (atrophy) in the part of the brain that controls movement and coordination (the cerebellum).

What are the symptoms?

Common symptoms include a general loss of control of bodily movements (ataxia), unsteadiness when walking, clumsy movement in the upper and lower limbs, slurring of speech (dysarthria) and possibly experience double vision or blurred vision. [Houlden et al. Nature genetics 2007; 39 (12):1434-36; Worth et al. Am J Hum Gen 1999; 65: 420-426]

The disease has a relatively slow progression and normal life expectancy.

What causes SCA11?

SCA11 is caused by a fault in a gene known as a mutation. Recent research at the National Hospital for Neurology and Neurosurgery in London has led to the identification of the exact location of the single gene responsible for SCA11 [Houlden et al. Nature genetics 2007; 39 (12):1434-36].

How is SCA11 inherited?

SCA11 is inherited in an autosomal dominant way. For more information on inheritance see Ataxia UK's '*Ataxia: what's that?*' leaflet.

When do symptoms start?

The age of onset for the disease has been reported to range from 11 to over 60 years of age. These figures were obtained from the study of eight generations of two families with SCA11.

How is SCA11 diagnosed?

As the discovery of SCA11 is so recent, there is as yet no laboratory that offers SCA11 testing. There are, however, plans to make SCA11 testing available at the National Hospital for Neurology and Neurosurgery in the near future. Also, there is a research project currently taking place at this Hospital, and patients attending the Ataxia Clinic there will be tested for SCA11 if clinically relevant.

How common is SCA11?

There is currently little information on the prevalence of the SCA11 in the UK.

Ataxia UK is funding research to rectify this lack of knowledge. Most of the current information on the disease has come from the study of two affected families, in which the disease stretches over eight generations and so far includes 34 affected individuals.

Management of SCA11

As with other cerebellar ataxias, physiotherapy and speech therapy can be helpful. A visit by an occupational therapist will be useful in order to assess the need for items such as adaptations to the home. It is important to see a neurologist, who will monitor the condition, on a regular basis.

This information leaflet was written by Ataxia UK in collaboration with Dr Giunti, Neurologist at National Hospital for Neurology and Neurosurgery, London.

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