

The episodic ataxias are a relatively rare group of conditions, which as their name suggests tend to affect people in bouts or attacks of unsteadiness. There are several types but 2 are the most well understood, and they are called episodic ataxia type 1 and 2. Both of these occur in families and are inherited in what is known as an autosomal dominant manner. This means that an affected individual has a 50% chance of passing the gene on to their children.

Episodic ataxia type 2 (EA-2)

It involves people having episodes of ataxia that last hours to days. Exertion and stress often trigger the episodes. Common symptoms are vertigo, nausea and vomiting, nystagmus (involuntary eye movements) and some people have migraine. It is a rare disorder, representing less than 1% of the inherited ataxia syndromes. Acetazolamide often dramatically helps with the symptoms during the episodes of ataxia. Please note that the use of any medicines would of course always need to be discussed with a doctor or neurologist who would know the specific circumstances of the patient. Not all people respond well to acetazolamide and some people may be allergic to it. If acetazolamide is ineffective or causes side effects there are other drugs that can be tried. As stress often triggers attacks, stress management techniques (e.g. meditation) can be helpful in controlling symptoms. Alcohol and caffeine should be avoided, and regular but modest exercise should be encouraged. In some cases patients with this form of ataxia develop a slowly progressive ataxia, this most frequently coincides with the attacks lessening in frequency. EA-2 is caused by mutations in a calcium channel found in cells mainly in the cerebellum. Calcium channels normally are important for maintaining the correct amount of calcium in cells which is important for the cells to perform their normal functions. It is important to see a neurologist, who will monitor the condition, on a regular basis.

Episodic ataxia type 1 (EA-1)

EA-1 is very rare - in a relatively recent publication only a few families had been identified. It is characterised by short attacks (seconds or minutes) of incoordination and dysarthria (speech problem) that generally last minutes. Myokymia (twitching of muscles) is present in most people. It can be twitching of muscles in hand, fingers, sometimes the tongue. Myokymia is frequently accentuated during attacks. The severity of the symptoms varies greatly between individuals, both in terms of the severity and frequency of attacks and also the amount of myokymia. Most people do not have symptoms between attacks but others may have slight ataxia and tremor between attacks. Attacks can occur spontaneously or they can be triggered by rapid sudden movements, or if one is startled. Anxiety, and fatigue also increase the susceptibility to an attack. Basically, EA-1 is a non-progressive disorder, but some elderly people show slight permanent ataxia and tremor. Once a diagnosis is confirmed there are a number of drugs that can be tried to help with the symptoms. EA-1 is caused by mutations in potassium channels in cells that may lead to problems with transmitting nervous impulses. 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 Professor Nicholas Wood, Neurologist at National Hospital for Neurology and Neurosurgery, London.

Disclaimer

This leaflet is for guidance purposes only and, while every care is taken to ensure its accuracy, no guarantee of accuracy can be given. Individual professional advice should be sought before taking or refraining from taking any action based on the information contained in this leaflet and nothing should be construed as professional advice given by Ataxia UK or any of its officers, trustees or employees. No person shall have any claim of any nature whatsoever arising out of or in connection with the contents of this leaflet against Ataxia UK or any of its officers, Trustees or employees

Ataxia UK, Lincoln House, Kennington Park, 1-3 Brixton Road, London SW9 6DE

**www.ataxia.org.uk
Tel: 020 7582 1444**

**helpline@ataxia.org.uk
Helpline: 0845 644 0606**

Disclaimer

This leaflet is for guidance purposes only and, while every care is taken to ensure its accuracy, no guarantee of accuracy can be given. Individual professional advice should be sought before taking or refraining from taking any action based on the information contained in this leaflet and nothing should be construed as professional advice given by Ataxia UK or any of its officers, trustees or employees. No person shall have any claim of any nature whatsoever arising out of or in connection with the contents of this leaflet against Ataxia UK or any of its officers, Trustees or employees.