PACA (Primary Autoimmune Cerebellar Ataxia)

PACA

PACA is an autoimmune ataxia, meaning that the body's immune system mistakenly attacks the co-ordination centre of the brain, the cerebellum. **[Hadjivassiliou, M. et al. Diagnostic Criteria for Primary Autoimmune Cerebellar Ataxia – Guidelines from an International Task Force on Immune-Mediated Cerebellar Ataxias. The Cerebellum. 23: 605-610, 2020.]**

What are the symptoms?

Common symptoms of PACA include unsteady gait, limb incoordination, speech difficulties (dysarthria), repetitive involuntary movements of the eyes (nystagmus) and double vision (diplopia).

When do symptoms start?

In a study of 30 people with PACA, onset of symptoms was seen to begin during adulthood, with the average age of onset of symptoms being in a person's 50s, but this ranged from 18 years to 83 years. **[Hadjivassiliou, M. et al. Treatment of Primary Autoimmune Cerebellar Ataxia with Mycophenolate. The Cerebellum. 19: 680-684, 2020.]**

How is PACA diagnosed?

There is not a single definitive test for PACA. In 2017, an International Task Force for Immune-Mediated Cerebellar Ataxias, of which Prof Hadjivassiliou at the Sheffield Ataxia Centre is the lead, wrote a comprehensive list of clues that neurologists should look for when considered whether someone has PACA. **[Hadjivassiliou, M. et al. Diagnostic Criteria for Primary Autoimmune Cerebellar Ataxia – Guidelines from an International Task Force on Immune-Mediated Cerebellar Ataxias. The Cerebellum. 23: 605-610, 2020.]**

This involves a combination of clinical assessments, brain scans, family history and antibody testing. As there is a not a specific trigger for PACA, such as in the case of Gluten ataxia where Gluten is the known trigger, other immune ataxias with known triggers must be ruled out, as well as immune ataxias for which well characterised antibodies are directly involved in the disease.

How common is PACA?

The exact number of people living with PACA is not yet known. However, PACA could be responsible for a number of idiopathic ataxia cases, where the cause is unknown.

Management of PACA

In a study led by the Sheffield Ataxia Centre, 22 people with PACA were treated with the immunosuppressive drug Mycophenolate, which reduces the immune response that causes the ataxia. **[Hadjivassiliou, M. et al. Treatment of Primary Autoimmune Cerebellar Ataxia with Mycophenolate. The Cerebellum. 19: 680-684, 2020.]** This treatment may be able to prevent further damage to the cerebellum and salvage any damaged cells. The study results from brain scans and ataxia clinical rating scales showed that those who received the treatment improved and stabilised, whilst those who did not receive the treatment progressively worsened.

This information leaflet was written by Ataxia UK in collaboration with Prof Marios Hadjivassiliou, Consultant Neurologist at the Sheffield Ataxia Centre.

August 2025.

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