



# Gluten ataxia

It is exactly four years since my last article in *The Ataxian* on gluten ataxia. At that time Ataxia UK had awarded us a grant to investigate the mechanism by which gluten sensitivity (ie: sensitivity to a protein found in wheat, barley and rye) can manifest with cerebellar dysfunction, causing ataxia (gluten ataxia). This work has resulted in a various publications and has advanced our understanding of this relatively common form of ataxia. To put it in perspective, gluten ataxia accounts for up to 40% of the idiopathic sporadic ataxias (those with no known cause and in whom there is no family history of ataxia).

We have advanced our knowledge on two fronts: firstly on the mechanism of the production of ataxia and secondly on the effect of gluten-free diet in patients with gluten ataxia.

We have demonstrated that antigliadin antibodies (these are the antibodies against the gluten protein) recognise antigens (proteins) found on Purkinje cells - the most important cells of the cerebellum (balance centre). This suggests that there is some part of the Purkinje cell that shares similar structure with part of the gluten protein. Antigliadin antibodies are found also in patients with gluten sensitivity who have small bowel inflammation (coeliac disease) but no ataxia. To explain why it is the cerebellum that is affected in gluten ataxia and not the bowel, we went on to demonstrate that patients with gluten ataxia have additional antibodies that are targeting the Purkinje cells. We are now looking for the precise protein that these antibodies are recognising. This may enable us to develop a blood test that will exclusively diagnose gluten ataxia and distinguish it from other forms of gluten sensitivity.

### Gluten-free diet

The second aspect of our research was to investigate the effectiveness of a

gluten-free diet in patients with gluten ataxia. We would like to take this opportunity to thank all those who participated in this trial. All patients underwent validated tests to assess their balance at the start of the study and after one year on the diet. Twenty six patients (treatment group) were strict with the diet, as demonstrated by the elimination of the antigliadin antibodies within one year. Fourteen patients decided not to go on the diet (control group).

After one year, there was improvement in ataxia reflected in all of the ataxia tests in the treatment group. This was significant when compared to the control group. We concluded that gluten ataxia responds to a strict gluten-free diet. The prompt diagnosis of gluten ataxia is vital. The sooner the diagnosis is made, the more likely that the diet will be effective. If a patient with gluten ataxia has remained undiagnosed for years, the damage to the cerebellum may be permanent and the improvement minimal. However, the diet may prevent deterioration.

We therefore suggest that all patients with ataxia of unknown cause (idiopathic ataxia) should be tested for gluten sensitivity. The testing should be done using antigliadin antibodies (IgG type). We have offered, and will continue to offer this testing facility for those members of Ataxia UK who are unable to access the test through their doctors locally.

### New imaging project

Thanks to a grant from Ataxia UK we are now able to start our next project which focuses on imaging the balance centre (cerebellum). Readers will be familiar with Magnetic Resonance Imaging (MRI) as the best imaging technique for patients with ataxia. MRI is capable of showing shrinkage (atrophy) of the balance centre often seen in patients with gluten ataxia. However, quantifying the degree of

atrophy is not easy and changes occurring over a period of a year may be so small that they remain undetected. Magnetic Resonance Spectroscopy (MRS) is a more sensitive technique. It uses the same scanning equipment as MRI. But instead of producing images of the structure of the brain, it provides information on the way the cerebellum is functioning, based on the chemical products of its cells. Pilot data collected so far suggest that even those patients with gluten ataxia with no evidence of cerebellar abnormalities on MRI may have abnormal MRS.

We intend to monitor improvement or deterioration in patients with gluten ataxia of cerebellar function by doing MRS at baseline and then a year after the introduction of a gluten-free diet. We hope that this may confirm the results of the trial described above and provide further evidence for the beneficial use of a gluten-free diet. There will be two control groups: healthy controls and an ataxia control group (consisting of people with either Friedreich's ataxia or SCA6). We are looking for people with FA or SCA6 to take part so if you are interested in helping (by travelling to Sheffield twice for MRS scans), please contact the Ataxia UK office. As well as acting as a control group to this study we will also be able to get information about the rate of progression (in MRS terms) of Friedreich's ataxia and SCA 6.

Thanks to the efforts of Ataxia UK and its members, ataxia is no longer a neglected neurological disease, largely ignored by researchers and clinicians alike. It has been transformed to a challenging neurological condition that deserves all our attention.

**Dr Marios Hadjivassiliou**

Consultant Neurologist

The Royal Hallamshire Hospital,  
Sheffield.