

# Ataxia Triggered by Gluten

Hakim Rahmoune<sup>1,2,\*</sup>, Nada Boutrid<sup>1,2</sup>, Mounira Amrane<sup>2,3</sup>, Belkacem Bioud<sup>1,2</sup>

<sup>1</sup>Department of Pediatrics, University Hospital of Setif, Setif 1 University, Algeria

<sup>2</sup>Genetic, Cardiovascular & Nutritional Diseases Laboratory, Setif-1 University, Algeria

<sup>3</sup>Biochemistry Laboratory, University Hospital of Setif, Setif 1 University, Algeria

\*Corresponding author: [rahmounehakim@gmail.com](mailto:rahmounehakim@gmail.com)

**Abstract** The gluten ataxia is one of the commonest neurological manifestations of gluten-related disorders: prevalence was estimated at 15% amongst all ataxias and 40% of all idiopathic sporadic ataxias; its diagnosis should be confirmed by the presence of anti-gliadin antibodies. Gluten ataxia is, by its management, very peculiar: few etiologies, among the large panel of childhood ataxias, may respond to dietary treatments. We present an instructive case of gluten ataxia in a teenage girl.

**Keywords:** ataxia, gluten, anti-gliadin antibodies, diet

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## 1. Introduction

Nutrition-responsive ataxias are specific, rare and treatable causes in children. We present an instructive case report of gluten ataxia, a common and particular neurological presentation in the wide spectrum of gluten related disorders.

## 2. Case Report

A 11-year aged girl is admitted for a second episode of acute ataxia. Her somatic and neurological examination showed no other abnormalities.

She had only dynamic ataxia with no MRI detected abnormalities in the cerebellum.

Reviewing her history, the patient also reported suffering from chronic diarrhea since several years; without prompt investigation neither diagnosis

Celiac disease with gluten ataxia was then suspected; positive serology (IgA anti-tissue transglutaminase type 2 and IgA anti-gliadin) associated to flat intestinal mucosa lead to confirm the gluten intolerance. Once the teenager put on gluten free diet, she has no more presented such ataxias during 3 years of follow-up

## 3. Discussion

Cerebellar ataxia may be an organ-specific autoimmune disease, and evidence comes from the observation that the prevalence of autoimmune diseases in patients with idiopathic sporadic ataxia is significantly higher (47%) than in control populations [1,2]. In the other hand, among the large panel of childhood ataxias, few etiologies may

benefit from dietary or biochemical therapies, such as the gluten ataxia along with other hereditary forms like Refsum, ataxia with vitamin E deficiency, cerebrotendinous xanthomatosis and coenzyme Q10 deficiency [3]. Cerebellar ataxia, with or without associated enteropathy, was one of the first symptoms to be described in connection with CD. Symptoms are similar to other forms of cerebellar ataxia including gait and limb ataxia [4]. Nowadays, gluten ataxia is best defined as a gluten-related disorder, a term that encompasses all pathological conditions related to gluten. [5]. It is considered as one of the commonest neurological manifestations of gluten-related disorders: its prevalence was estimated at 15% amongst all ataxias and 40% of all idiopathic sporadic ataxias. [6]. Its diagnosis should be confirmed by the presence of anti-gliadin antibodies, with or without other serological, autoimmune markers of gluten intolerance (i.e. anti-tissue transglutaminase) [6,7]. Early diagnosis and treatment may protect the first group from the development and/or progression of neurological dysfunction. [7]. Recently, it was reported that the presence of anti-transglutaminase-6 (TG6) antibody can be used as a biomarker to diagnose gluten ataxia. [8]

Gluten free diet remains the only therapy that has proven real efficacy, and the response to diet seems depending on the prior duration of the ataxia before diagnosis. [6,7,9].

## 4. Conclusion

Nutrition-dependent forms of ataxia like gluten ataxia should be investigated in pediatric population at first line option as it dramatically respond to dietary adaptation.

Gluten ataxia is also a peculiar, treatable and reversible form of the expanding spectrum of gluten-related neurological disorders.

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