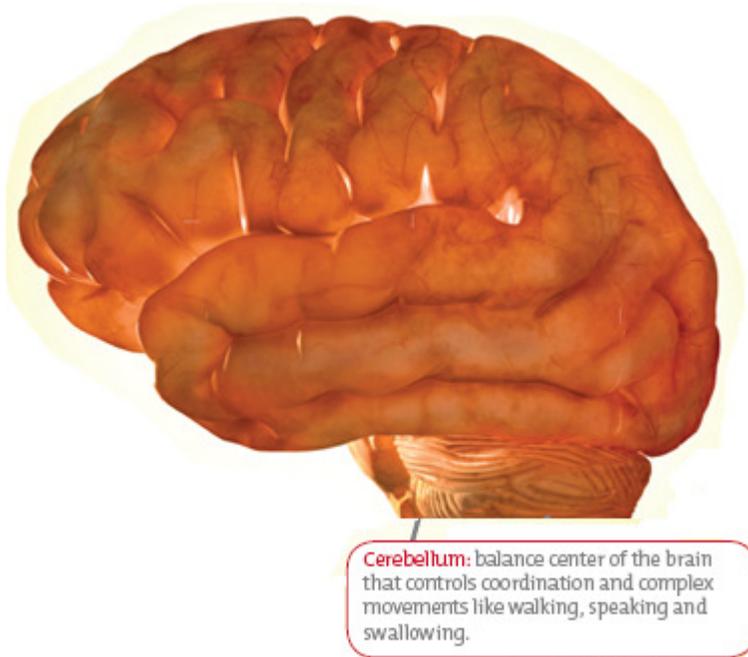


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Gluten Attack: Ataxia

Is gluten attacking your brain?



Cerebellum: balance center of the brain that controls coordination and complex movements like walking, speaking and swallowing.

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Not long after her diagnosis, Sarah Bosse realized her case of celiac disease was not typical. Bosse had joined a support group for newly diagnosed celiacs and was surprised by what she saw at the first few meetings.

“They were working full-time jobs, had families and went out at night. Not to discount the severity of their symptoms and experiences but they had a much more normal life.”

Bosse had expected them to be as sick as she was.

The 26 year old from Raleigh, North Carolina, can trace her celiac symptoms all the way back to elementary school when she spent countless hours holed up in the nurse’s office, nibbling on saltines for her diarrhea, nausea and stomach cramps.

“Looking back, it’s so ironic. Those saltines were making me worse,” recalls Bosse. Her symptoms continued into her teen years and were chalked up to anxiety and irritable bowel syndrome. The bloating, diarrhea and cramping were such a constant part of her youth that when she looks at childhood pictures, she can see the discomfort and embarrassment on her face.

“I didn’t realize it at the time but my life has been about managing my stomach problems,” she says.

Shortly after finishing college, Bosse began experiencing frightening new symptoms. She was increasingly dizzy and disoriented, frequently stumbling while rounding a corner or changing directions. She’d even fall out of a chair or her own bed at times, blaming it on klutziness until balance was no longer her only new complaint. Her head started to feel thick and foggy and she had trouble concentrating, likening it to an extreme case of ADHD. She also began having unusual problems with her vision, seeing bright flashes and noticing that her eyes seemed to bounce, rather than rest on what she was looking at.

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“I could be at the grocery store, staring at a display of ketchup. I’d try to reach for one but my eyes wouldn’t stay still so that I could pick it up,” she says.

Before Bosse was able to see a specialist, a far scarier symptom struck. She was asleep in the early hours of the morning when she woke with a start, gasping for breath. She felt paralyzed, as if her body had forgotten how to breathe. Also unable to swallow, she was choking on her own saliva. Petrified, she managed to call for help and was rushed to the emergency room.

Concerned she might have multiple sclerosis, Bosse underwent a series of brain MRIs at the hospital. When they came back clear, she was tested for Lyme disease, lupus, rheumatoid arthritis and countless other conditions over the next several weeks. Results were normal and doctors didn’t have an answer for her.

Then a close friend was diagnosed with celiac disease and Bosse wondered if it could help explain her digestion symptoms. She asked to be tested and just days later, the diagnosis was nailed. Within weeks of eating gluten free, her stomach troubles were turning around but her balance problems were worse than ever.

“I couldn’t walk anything close to a straight line. By that time, I was spending most of my day in a wheelchair. I had to stop working outside my home. It was hard to appreciate the fact that my stomach wasn’t hurting as much because I was housebound,” she says.

Losing hope, Bosse was hospitalized for depression. While there, on a hunch one of her doctors suggested she could have a little-known condition called gluten ataxia.

A Controversial Call

Gluten ataxia is a neurologic condition characterized by the loss of balance and coordination. However it can also affect fingers, hands, arms, legs, speech and even eye movements. Typical symptoms include difficulty walking or walking with a wide gait, frequent falls, difficulty judging distances or position, visual disturbances and tremor.

Experts believe gluten ataxia may be a form of gluten sensitivity, a wide spectrum of disorders marked by an abnormal immunological response to gluten.

Different organs can be affected by different types of gluten sensitivity. In celiac disease, sometimes called gluten-sensitive enteropathy, the small bowel is affected. In dermatitis herpetiformis, the skin is targeted, resulting in an itchy rash. With gluten ataxia, damage takes place in the cerebellum, the balance center of the brain that controls coordination and complex movements like walking, speaking and swallowing.

Purkinje cells in the cerebellum, key in maintaining balance, are thought to be lost in gluten ataxia.



Brain Freeze

Celiac disease is not just a gastrointestinal disorder, emphasizes Marios Hadjivassiliou, MD, a neurologist at Royal Hallamshire Hospital in Sheffield, England. According to an estimate published in *Neurology* in 1980, up to 10 percent of celiac patients develop neurologic complications. More recently, Hadjivassiliou and colleagues found that for every seven celiac patients presenting to a gastroenterologist, two have neurologic manifestations—and most of these have no bowel symptoms.

Viewing celiac disease as a systemic disorder, not just a gastrointestinal disorder, would improve recognition of its frequent neurologic symptoms, such as peripheral neuropathy (numbness and tingling in the hands and feet) and ataxia (disturbance in balance).

According to author Peter HR Green, MD, director of the Celiac Disease Center at Columbia University, in his book *Celiac Disease, A Hidden Epidemic*, other symptoms include epileptic seizures, particularly in children, migraines, brain atrophy and dementia.

“It’s best to describe gluten ataxia using the term gluten sensitivity because it takes one away from the misconception that you must have celiac disease to have any of these

diverse manifestations,” says Marios Hadjivassiliou, MD, a neurologist at Royal Hallamshire Hospital in Sheffield, England.

Hadjivassiliou first described gluten ataxia in the 1990s. After seeing a number of patients with unexplained balance and coordination problems, he began systematically testing them for gluten sensitivity using antigliadin antibodies, which point to a heightened immune response to gluten but not necessarily to a diagnosis of celiac disease. Hadjivassiliou found a very high prevalence of antibodies in patients with ataxia, coining the condition, gluten ataxia.

But not all neurologists are on board with gluten ataxia. Although several studies support Hadjivassiliou’s findings, at least one small study, published in *Neurology* in 2000, failed to find a link between antigliadin antibodies and cerebellar ataxia. None of the 32 patients in the study tested positive for the antibodies.

But this study is just part of the problem. Casting more doubt on the condition is its so-called ‘soft’ diagnostic criteria. Gluten ataxia is currently diagnosed when antigliadin tests suggest gluten sensitivity and other causes of ataxia are ruled out. (Small bowel biopsy is advisable in patients with positive antigliadin and celiac blood tests). It’s a diagnosis of exclusion.

“There’s fairly good evidence why celiacs could have neurologic problems like ataxia,” says Joseph Murray, MD, a gastroenterologist and celiac expert at the Mayo Clinic. Vitamin deficiencies or a phenomenon called molecular mimicry could be to blame. In molecular mimicry, something in the brain may look enough like gluten that antibodies directed at the small bowel cross-react against part of the brain.

“The bottom line is that when celiac disease and gluten ataxia occur together, gluten ataxia can be a robust diagnosis. But when gluten ataxia occurs on its own, we have less certainty of the diagnosis,” says Murray.

A new screening tool may soon help. Hadjivassiliou and his team recently identified an antibody, transglutaminase TG6, which may be a better marker for gluten ataxia. TG6 is similar to the antibody TG2, detected in the widely used tTG screening for celiac disease—but TG6 is primarily expressed in the brain. Although promising, a test for TG6 is not yet ready for clinical use.

Awareness Lacking

Unfortunately, familiarity with gluten ataxia in the medical community tends to be the exception, not the rule, says Murray.

Bosse’s experience echoes this. “I’m not sure many of my doctors understand when I tell them I have gluten ataxia,” she says. “If I had to go to the hospital, I’m not sure the medical staff would know what I’m talking about.”

Bob Hunter, 63, a patent agent from the Big Island of Hawaii, also has gluten ataxia. His wife first noticed back in 2004 that his hands were shaking when he moved them, like reaching for his coffee cup.

“If you want to grab something, you don’t think about all the things necessary to make that happen. You just close your hand around it and do it. But if that part of the brain isn’t working well, you have to be more conscious of every muscle needed to perform the task,” says Hunter.

Like Bosse, Hunter experienced brain fog and problems with his balance, stubbing his toes and ramming his shoulders on door frames as he tried to walk, clumsily, through his house. He appeared drunk. Embarrassed, he was quick to reassure those around him that wasn’t the case.

Hunter traveled from his home in Hawaii to the Mayo Clinic in Minnesota before he was finally diagnosed with both celiac disease and gluten ataxia.

Carolyn Davison, 39, a mother of two from New South Wales, Australia, traveled halfway around the world for her diagnosis. Davison had been hospitalized half a dozen times following frightening bouts of paralysis and numbness in her legs. When her doctors timed how long she could walk before collapsing or losing her balance, the best she could do was four minutes. She experienced other strange neurologic symptoms, as well. She’d write and her letters would come out backwards, she’d get lost in the supermarket and she’d forget she was talking, mid-sentence. Before long, Davison couldn’t work, giving up her job as a therapist specializing in children with developmental disabilities and autism. Unable to negotiate stairs, she moved to a one-story house.



Defining Gluten Sensitivity

The term "gluten sensitivity" was once synonymous for celiac disease, a condition originally known as "gluten sensitive enteropathy." The precise definition of gluten sensitivity is currently being debated among experts. An umbrella term, its definition has expanded as more gluten-associated disorders, such as dermatitis herpetiformis and gluten ataxia, are recognized and included.

Neurologist Marios Hadjivassiliou, MD, an expert in gluten ataxia, describes gluten sensitivity as a state of heightened immunological responsiveness to ingested gluten in genetically susceptible people.

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On the long list of conditions Davison was tested for, including Guillain–Barré syndrome and multiple sclerosis, blood work was run for celiac disease. Her antibodies were raised but a biopsy failed to find anything. Although Davison’s neurologist knew that in some celiacs there can be neurologic symptoms, without a clear-cut case of celiac disease, she was out of ideas.

Meanwhile, Davison started to question her sanity. Her doctors were giving up on her but her symptoms were getting worse. Like Bosse, she began experiencing frightening episodes where she couldn’t catch her breath and couldn’t swallow, choking on her own saliva. Terrified, Davison took matters into her own hands. She got online and came across Hadjivassiliou’s research. She showed it to her neurologist, who encouraged her to travel halfway across the world to see him.

Armed with binders full of her medical records, Davison went to England and met with Hadjivassiliou. He ran just one additional test, the genetic screen for celiac disease. The test can’t diagnose celiac disease but some experts believe when positive, it can suggest a genetic predisposition toward gluten sensitivity. For Davison, the genetic test helped provide even more evidence she’s gluten sensitive and after three long and trying years, she was finally diagnosed with gluten ataxia.

Help Through Diet

Delayed diagnosis of gluten ataxia is the norm, says Hadjivassiliou, particularly for those patients like Davison, who don’t have celiac disease or gastrointestinal symptoms. Clinicians may look for gluten sensitivity only if gastrointestinal symptoms are present and they’re unlikely to think of gluten sensitivity in the context of ataxia, says

Hadjivassiliou. But his research, published in *Brain* in 2003, found up to 40 percent of patients with unexplained ataxia have gluten sensitivity. Hadjivassiliou recommends neurologists routinely screen patients with unexplained ataxia for gluten sensitivity.

Those with gluten ataxia have no time to waste, he warns. The gluten-free diet—the mainstay of treatment for gluten ataxia—can result in a stabilization of symptoms. But often, significant damage is already done.

The neurologic system tends to heal very poorly and very slowly, says Murray. Unlike the small bowel lining, Purkinje cells of the cerebellum have no capacity for regenerating, explains Hadjivassiliou. Once ataxia is well established, which can happen in as few as six months, it's rare to make a full recovery.

Extensive damage helped explain why, even after going gluten free, Bosse continued to have debilitating symptoms. Her ataxia was advanced. It's not unusual to be wheelchair-bound at diagnosis, Murray says.

However, the outlook isn't grim. In addition to dietary intervention, physical and occupational therapy can make a big difference.

As a former occupational therapist assistant, Bosse practices many of the skills she used to teach. She spends hours each day 'retraining' her brain and muscles to perform everyday tasks. Physical therapy and regular exercise at the gym help strengthen her weakened muscles.

Like Bosse, Hunter began a rigorous physical therapy program shortly after his diagnosis. He also cut out alcohol, which can exacerbate ataxia symptoms. Having read about vitamin D's many health attributes, including possible benefit in multiple sclerosis and other autoimmune and neurologic conditions, Davison began supplementing her diet with vitamin D. Although Davison feels it improves her symptoms, particularly in winter, experts don't yet endorse vitamin D supplementation for gluten ataxia. Celiac disease can cause nutritional malabsorption (of copper and vitamins B6, B12 and E, for example) that may affect balance but gluten ataxia on its own is not thought to cause vitamin deficiencies.

The overwhelming evidence is that gluten ataxia is immune-mediated, say experts. Hadjivassiliou and his team are currently studying how the condition damages neural cells, with the hope of one day developing better targeted therapies. For now, it's most important for patients to repeat blood testing, usually after six months of treatment with the gluten-free diet, to ensure elimination of all antibodies. Symptoms can stabilize and improve once antibodies are gone.

Slow But Steady

Thanks to her hard work, Bosse has made big gains since going gluten free last year. Her celiac is in check and her ataxia symptoms have improved significantly. Fortunately, she

hasn't experienced any subsequent episodes where she struggles to breathe or swallow. Although she still needs her wheelchair much of the day, it's often the burning pain caused by peripheral neuropathy, another neurologic complication of celiac disease, that keeps her off her feet. Bosse recently picked up her piccolo and flute again, favorite hobbies of hers, and found a job she can do part-time from home.

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But recovery is slow—and part of her healing strategy is to simply slow down.

“I have to be willing to slow down and work with my body, especially on days when I'm having more problems. When my emotions get worked up, the ataxia gets worse. I have to calm down and move more slowly, focusing on everything I'm doing,” she says.

Davison also has to do one thing at a time. There's no multitasking. After two years on the gluten-free diet, her coordination has improved substantially and she's able to walk and do gentle yoga exercises. But she has to pace herself.

“My main problem today is fatigue. I'm not working yet but I've started volunteering several mornings a week in my field. Those afternoons I spend in bed.” Although it's sometimes a struggle to stay positive, looking back she can see steady improvement overall.

Dietary Vigilance

If Bosse had any doubts about having gluten ataxia, they were put to rest the first time she accidentally ate gluten. Her roommate had purchased a loaf of spelt bread, and mistaking it for gluten-free bread, Bosse ate it. Within an hour, she was doubled over with diarrhea that lasted for two weeks. Then the joint pain set in. Dizziness, confusion and vision problems kept her bed-ridden for several more weeks.

With this level of sensitivity, a dietary slip is a major setback. The last time it happened for Bosse was at her brother's wedding. She ate a bite of roast beef and was sick 30 minutes later. She thinks the knife used to slice the meat may have doubled to cut rolls. Three months later, she was still suffering from an exacerbation of her ataxia symptoms.

Eventually the symptoms plateaued, leveling out to a point she describes as “just a little lower” than where she was before.

“Every time I’m exposed to gluten, I lose a little bit of something I used to be able to do, probably permanently,” she says.

It’s hard, if not impossible, to fully recover from an incident of getting inadvertent gluten, agrees Davison. “I can’t take any risks with food. The consequences for me are far more serious than for lots of others on gluten-free diets.”

For her own well-being, Bosse doesn’t dine at restaurants. She takes a packed cooler when she goes out with friends. She doesn’t eat food that others prepare. Instead, she invites friends over to her kitchen where they cook using her utensils and ingredients.

“Although all celiacs need to be vigilant with their diet, I really counsel patients who also have gluten ataxia that they can’t take any chances at all,” says Murray. “They’ve got to protect their balance as much as possible.”

Battle for Health

“Every day is a fight with gluten ataxia,” says Bosse. “You have to constantly fight to get better.” In Bosse’s case, she also has to fight against occasional bitterness. She can’t help wondering, what if her celiac disease and gluten ataxia had been diagnosed promptly? When her mood droops, she turns to her music and her faith.

She also takes pride in finding creative solutions to the challenges presented by her ataxia. She’s arranged the furniture in her apartment so she can negotiate corners more easily and quickly stabilize herself, if necessary. Given the lapses in her short-term memory, she knows that good organization and a regular schedule are essential. She takes advantage of technology, wearing a watch synched with her computer that reminds her of appointments or that water is boiling on the stove. Her computer works with voice commands so that she doesn’t have to type by hand, a big help with the new job.

“I’m doing everything I possibly can to improve my quality of life,” she says, adding that an even bigger goal is to improve the lives of others with disabilities. She’s currently an active member of an online support group where she offers problem-solving ideas for others suffering from ataxia. She also ministers to those with disabilities at her church.

“If I hadn’t struggled with gluten ataxia, I wouldn’t have the empathy to really care for others,” she says, renewed by her outreach. “I’ve learned that in every life experience, no matter how difficult, there’s hope and joy.”

Medical writer Christine Boyd lives in Baltimore.

