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**Magazine**

# Why Was Her Vision Jerky and Blurry if There Was Nothing Wrong With Her Eyes?

## Diagnosis

By Lisa Sanders, M.D. May 1, 2018

The young woman rubbed her eyes. The numbers and letters on her computer screen jumped erratically. So did the world around her. This had happened before, but late at night when she was tired, never in the middle of the day. The light from the screen suddenly seemed too bright. And her headache, the one that was always present these days, tightened from a dull ache to a squeezing pressure on the back of her head and neck. Nearly in tears from pain and frustration, the 19-year-old called her mother. She couldn't see; she couldn't drive. Could her mother pick her up from work?

The problems with her eyes began in grade school. Two years earlier, she nearly went blind. All she could see on the left was a rim of light. Everything else was blocked by a big black spot. And then a black dot appeared in her right eye as well. Her parents took her to see many eye doctors, only to be told that there was nothing wrong. One doctor told them that she had "emotional blindness." The young woman's vision somehow got a lot better on its own, and though the black dot still obstructed some of her vision, for the last eight months she'd been able to drive — so important in this small mountain town an hour north of San Diego.

### Problems in the Brain?

Now she couldn't see for what seemed like a different reason. The young woman's mother arranged for her to go to San Diego to see a neuro- ophthalmologist — a doctor who specializes in vision problems that originate in the brain. When they got to the office, though, the young woman's vision and headache had returned to their imperfect but baseline state. She told the doctor that her symptoms were least intrusive in the morning; standing and walking seemed to make everything worse. Come back later, the doctor instructed. Mother and daughter walked around and shopped.

When a couple of hours later the daughter's eyes started jumping and her headache worsened, they hurried back to the office.

The doctor took one look at the young woman's eyes and told her she had nystagmus. It's a failure in the parts of the brain that allow our eyes to stay focused when the object being observed or the observer moves. Problems in the inner ear — where head position is perceived — are the most common cause of nystagmus, usually accompanied by vertigo. Persistent nystagmus is worrisome because it can indicate abnormalities in other parts of the brain, primarily the cerebellum — the chief coordinator of all movement. The doctor sent the patient to

the Sharp Memorial Hospital emergency room for an M.R.I.; it would reveal if a clot or mass in the brain was causing the nystagmus.

There was no clot, no mass, but there was an abnormality. At the lowest part of the young woman's brain, where the spinal cord emerges, a tiny sliver of cerebellum was visible just below the skull. A little slippage of brain tissue into the spinal column can be normal as long as it is no more than five millimeters below the skull; anything more is considered pathological. Hers was right at five millimeters.

**How Serious Is Brain Slippage?**

This downward displacement of the brain, known as a Chiari malformation (after the 19th-century Austrian pathologist who identified the types of malformations) is a common abnormality. Imaging studies suggest that it may be present in one in every 200 of us, and many times it causes no symptoms at all. For these patients, the discovery of the Chiari malformation is usually accidental — noted on a scan obtained for some other reason.

The symptoms, for those who get them, are caused by compression of the brain tissue and nerves into the small space of the spinal canal. The specific symptoms will depend on what is being crushed. The most common is headache, usually located at the back of the head and down the neck, but a wide variety of other symptoms can occur, ranging from weakness, fainting and difficulty swallowing to hearing loss, curvature of the spine and insomnia. The question for patients with small Chiari malformations, like this young woman's, is whether it is the cause of her symptoms or an incidental finding. It's an important determination, because if the malformation is the cause, then surgery is needed to create room for the brain.

The young woman made an appointment with a neurosurgeon, but before she could see him, she awoke one day too weak to get out of bed. Her legs muscles refused to hold her up. When helped to her feet, bolts of pain shot from the back of her head down her spine into her legs. Her mother took her back to the E.R. at Sharp Memorial, where she was admitted.

Were the weakness, pain and nystagmus caused by the crowding in her brain? The neurosurgeon reviewed her scan. He said he did not think the tiny malformation visible on the M.R.I. could cause any of her symptoms; it was simply too small. She did need to be evaluated by a neurologist. That doctor was not sure what was going on, either.

**A Different Ailment**

The parents mentioned to the neurosurgeon that the year before, their daughter was diagnosed with Ehlers-Danlos syndrome (E.D.S.), an inherited disorder of the connective tissues that causes — in its most benign form — hypermobility in the joints and unusually stretchy skin. It can also cause repeated joint dislocations and injuries of skin, muscle and blood vessels. She already had four shoulder operations to stabilize the joints and prevent additional dislocations. Could the headaches and the nystagmus be related in some way to her E.D.S.? the parents asked. Probably not, the neurosurgeon told them. The patient's mother scoured the internet for a link between these two disorders. She came across several papers referring to a surgeon, Dr. Ulrich Batzdorf in Los Angeles, who described Chiari as a disorder that can be complicated by E.D.S. She called his office and was referred to Dr. Aria Fallah, a pediatric neurosurgeon at U.C.L.A. Mattel Children's Hospital.

**A Standing Test**

Fallah listened carefully as the young woman and her parents described her horrible past weeks — the jerky, blurred vision, the light sensitivity, the terrible headaches and now the weakness and pain in her legs.

After examining the young woman, Fallah then reviewed her M.R.I. Her symptoms were classic for a Chiari malformation, but her scan was not. While size alone did not determine how significant the symptoms would be, the bit of tissue slipping into the spinal cord on her M.R.I. seemed too small to cause the symptoms she described.

Following their appointment, Fallah took the case to Batzdorf, a mentor to him and a surgeon widely considered a "guru" in Chiari malformations and their repair. Batzdorf recommended an

M.R.I. done while the patient was standing upright. The patient had noticed that her symptoms got worse while standing; perhaps that reflected a change in the severity of the malformation. He'd certainly seen this in the past with some patients who also had E.D.S. He wasn't sure why.

Standing M.R.I.s are not widely available, but Batzdorf knew of one in a facility nearby. These images of the malformation were different. The bottom of the cerebellum extended nine millimeters into the spinal canal. And the compression of brain and nerve tissue was clearly visible in this scan.

**Necessary Surgery**

The patient was scheduled for surgery the next month. It was late morning when the young woman was taken to the recovery room after the procedure. The first thing she noticed when she woke was that her jiggling, blurred vision was now stilled and sharp. The black dot was also gone. The light no longer stabbed her eyes. She saw her mother and father come in and burst into tears. I can see, she sobbed. I can see.

It took the young woman many months of physical therapy to get her strength back. She will start college this summer. She's planning to be a physical therapist.

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