

A young man's ear pain was constant and debilitating, but doctor after doctor could not find the cure.

**B**y the time Stephen came to my office complaining of painful spasms and ringing in both ears, he had already been to several physicians about his problem. He had been given an array of diagnoses; most doctors said he had acute outer or middle ear infections and prescribed oral antibiotics or ear drops. These measures had not helped, though. He was 21 years old and had suffered with this pain for nearly a year.

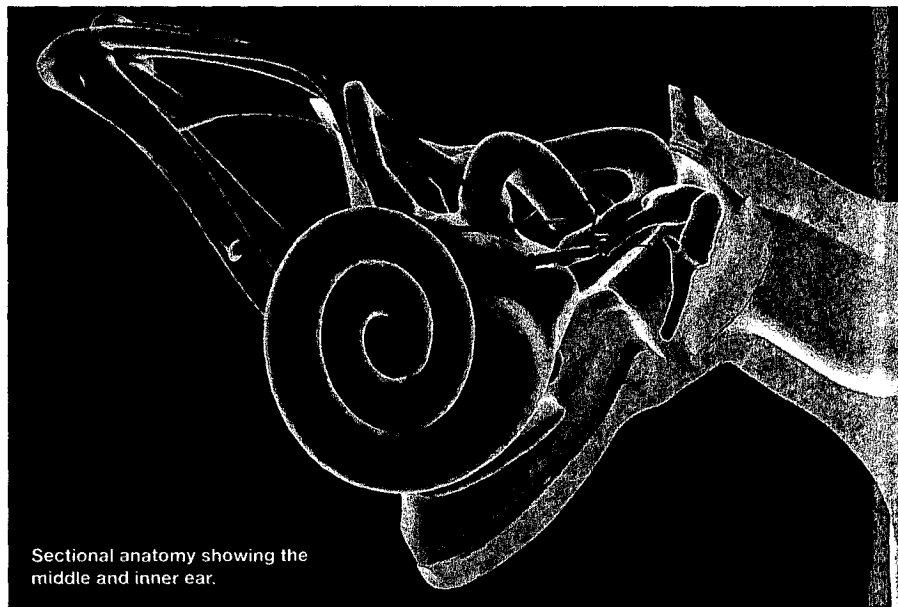
Like many people with ear or head sounds—which we lump into one word, *tinnitus*—Stephen was distraught. Knowing that there is no magic drug for ringing, I mentally prepared my usual sermonette: Avoid noise, aspirin, and methylxanthines (stimulants of the central nervous system, such as caffeine). And embrace the tried-and-true helper, sound substitution, or “masking.” This technique, in my experience, is a remedy for almost every patient with tinnitus.

Then Stephen delivered the bombshell. “I’ve been treated twice with extended courses of intravenous antibiotics for Lyme disease,” he told me. “In fact, I have an intravenous line in my arm right now and am just finishing an eight-week course of Rocephin [an antibiotic] for my second bout of Lyme. I’ve had to drop out of college because the pulses in my ears are so bad I can’t concentrate on anything. Could it be related to Lyme?” The spasms were occurring every few seconds in both of his ears throughout his entire waking day, he continued. The associated pain was so severe that his whole life revolved around dealing with it. Clearly this was not the typical case of tinnitus.

Stephen was first exposed to Lyme disease, a tickborne infection, as a child while playing in the woods near his home in Westchester County, New York. He was reinfected as a college student in 2007. Severe cases of Lyme disease can involve cardiac or neurological symptoms and are often treated with the intravenous antibiotic ceftriaxone. But I was unaware of any link between Lyme disease and pulsatile tinnitus. Otherwise, Stephen was in stable health with no other contributory medical or surgical problems.

I examined his right ear with a microscope and an ear speculum. There it was, apparent right away in an otherwise normal eardrum: The drum pulsed and appeared to go into spasm in an irregular way. I observed the exact same thing in the left ear. There was no mass behind either drum, and the middle ear clefts on each side were normal. My exam did find spasms of his soft palate (palatal myoclonus) and twitching around his eyes (blepharospasm), but these were not as bothersome to him as the pulsing. I had never before seen a patient like this.

It was time to put Stephen through some proper hearing tests to find out what was going on. The middle ear has two muscles, the tensor tympani and the stapedius. The stapedius and, to a lesser extent, the tensor tympani engage in the acoustic reflex (or the middle ear muscle reflex), a contraction in response to loud sounds. A reflex is an involuntary motor response, so it is present both while a person is awake and while unconscious. All reflexes follow a looping nerve response: an arc toward the central nervous system, called the afferent limb, and an arc away from the central nervous system to the end organ, termed the efferent limb. For the middle ear muscle reflex, the afferent limb is the auditory nerve; the efferent limb is the facial nerve for the



Sectional anatomy showing the middle and inner ear.

stapedius muscle and the trigeminal nerve for the tensor tympani muscle. The reflex itself occurs in the brain stem, one of the deepest and most basic parts of the brain.

The stapedius responds by tightening the mobility of the stapes (or “stirrup”), and the tensor tympani tightens the eardrum and pulls the malleus (“hammer”). The stapes and malleus are tiny bones that transmit hearing through the inner ear. By restricting the motion of the stapes, the stapedius may be able to dampen loud sounds coming toward the ear, but that response does not happen quickly enough to prevent acoustic trauma from fast-acting sounds, such as a gun blast. The acoustic reflex is also thought to play a role in preventing us from hearing our own voices as we speak. Although we know that the reflex is present in healthy individuals and we evaluate it as part of every complete hearing test, its ultimate function is not entirely understood.

To investigate Stephen's ear spasms, I ordered a complete hearing test. Any neurological process involving two or more cranial nerves almost always points the finger of diagnostic suspicion to the brain stem, which is the anatomical takeoff point of the cranial nerves. So I also ordered an MRI of my patient's brain, with and without contrast enhancement, to get a good look at that area.

The tests showed that Stephen's hearing was completely normal, but the acoustic reflexes could not be tested because of the ongoing spasms in each ear. The MRI of his brain was entirely normal as well. At this point, my most likely diagnosis was myoclonus (muscle spasm) of either or both the stapedius and tensor tympani muscles. The painful eardrum spasms were the result of the contractions of one or both of the middle ear muscles.

All physicians are trained to begin by thinking broadly and then apply reasonable medical and surgical measures to the most likely cause of the clinical problem. Physicians are also trained to treat medically first and to reserve surgery for cases that fail medical management. Because Stephen had already been treated for Lyme with intravenous antibiotics, I suggested a consultation with a neurologist and perhaps a short trial of common medications used for either seizure or peripheral neuropathy, such as tegretol or gabapentin. Tegretol decreases the spread of seizure; it is also used to treat bipolar disorder and trigeminal neuralgia, a condition causing severe facial pain. Gabapentin is a molecule related to gamma-aminobutyric acid, or GABA, a common neurotransmitter. It has well-studied analgesic and anticonvul-

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sant effects, although its exact mechanism of action is unknown. Gabapentin is used for a variety of neurological disorders, including spasm and extremity pain.

Stephen began with a short course of tegretol, prescribed by his neurologist. It did not lessen the symptoms or block the spasms, so I started Stephen on gabapentin. After a few weeks, he reported that it had no beneficial effect either.

Understandably anxious, Stephen had sought a few other consultations, with conflicting recommendations. I suggested he see a colleague of mine in Boston who had a great deal of experience with eustachian tube dysfunction. The Boston doctor made a small opening in Stephen's left eardrum and inserted a microscopic telescope attached to a video camera to visualize the middle ear muscles. All of this was done under topical anesthesia while Stephen was awake. The results matched my diagnosis but added an important new piece of information: The offending muscle was the tensor tympani muscle, not the stapedius.

Now that we had identified the culprit, how to treat it? There were two possibilities: injecting a neuromuscular-blocking medication, such as botulinum toxin (Botox), into the tensor tympani muscle in the middle ear, or cutting the muscle surgically. The former would be a temporary trial that would wear off in about 6 to 12 weeks. The second option would be permanent. There

were potential risks associated with either choice. First, either temporarily or permanently blocking the muscle might not solve the problem. Second, loud sounds coming toward the treated ear might seem even louder than before. Stephen considered his options and, after speaking with his parents, decided to have the muscle cut. He had been suffering with the spasms for so long that he did not want to try a temporary solution.

I started with the left ear because it was the more bothersome. The surgery was all done through the outer ear channel; it involved making small incisions around the eardrum so it could be turned aside like the page of a book. The eardrum was then dissected off the hammer bone to allow me to see the tensor tympani muscle as it approaches the hammer bone at its narrow neck. The nerve to Stephen's face runs in a bony channel near the tensor tympani muscle. We monitored his face with an electromyography system, which allowed me to electrically stimulate the facial nerve and make sure it was not being injured by the surgery. I then found and divided the tensor tympani muscle, first partially with a small knife and then completely with a scalpel-like laser. I placed a small amount of packing under the eardrum, turned it back into its usual place, put some packing material over the eardrum, and bandaged the ear. Stephen was awakened and transferred to the recovery room.

About an hour later, my patient was fully awake and ready to go home. I went to see him in the patient recovery area and he exclaimed, "It's gone! I think it's really gone!" His parents were as overjoyed as he was. It appeared that his long nightmare might finally be ending. Fast-forward several weeks: The surgery did in fact

abolish the spasm in Stephen's left ear. His hearing remained normal, and the spasms around his left eye subsided.

About six weeks after the left side, I operated on the right ear in a similar fashion. This side also healed, and the spasms went

away with no significant side effects. Today Stephen has resumed his university studies and is getting his life back on track.

I still had not addressed Stephen's original question: Was Lyme disease responsible for his ear agonies? I would say yes. The disease certainly has many far-reaching neurological symptoms. As Sir William Osler, regarded by many as the father of modern medicine, said many years ago, "He who knows syphilis knows medicine." Of course, syphilis was not Stephen's problem, but the cause of Lyme disease, *Borrelia burgdorferi*, is in the same family as the causative agent of syphilis, *Treponema pallidum*. They are both spirochetes, bacteria that cause an insidious range of health issues.

For me, Stephen's case reinforced the notion that the medical history, as detailed by the patient himself or herself, is the most important part of any medical or surgical encounter. Patients who have difficult medical conditions are usually good historians: They live with the problem all day, every day, and can reflect upon its origins and its clinical course. The patient almost always provides all of the diagnostic clues to the solution, guiding the physician in where to look and how to treat. **□**

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