

10-Year Ache

Singer/songwriter Rosanne Cash opens up about her long road to diagnosis and treatment for Chiari malformation and syringomyelia.

BY TODD FARLEY

Neurologic conditions can be very difficult to diagnose—just ask Rosanne Cash. The Grammy award-winning singer/songwriter and daughter of legendary country singer Johnny Cash lived with painful and often debilitating headaches for most of her adult life. It wasn't until more than a decade of searching that Cash finally discovered her symptoms were caused by a Chiari I malformation, in which part of the brain is forced downward into the spinal cavity, and an associated syringomyelia, which is a hole that forms in the spinal cord and can fill with fluid.

Never heard of a Chiari I malformation? You're not alone. Vineeta Singh, M.D., associate professor of neurology at the University of California, San Francisco, and a member of the American Academy of Neurology (AAN), says lack of awareness among patients and doctors is common. "The symptoms of a Chiari I malformation are quite nonspecific, quite vague. It can go undiagnosed for many, many years," she says.

The condition occurs when there's not enough space in the back part of the brain, forcing it downward. "A part of the brain called the cerebellum, which is in the back part of the head, leaves its normal location within the skull and descends into the spinal cavity in the neck," explains neurosurgeon John Heiss, M.D., of the Surgical Neurology Branch of the National Institutes of Health in Bethesda, MD.

As a result, the cerebellar tonsils (the lowest part of the cerebellum) are pushed out of the skull. "They slip out of the hole in the base of the skull and hang down, pushing against the lower part of the brain stem and the top part of the spinal cord," Dr. Singh explains. (See box, "The Basics: Chiari I Malformation.")

It's not easy to say with any precision how common Chiari I malformation is, according to Dr. Singh, because many people who have no symptoms will discover that they have Chiari I malformation incidentally, when it is seen on an MRI performed for another reason. Others may go through life with such a Chiari I malformation and never know it.



SINGING OUT Cash performing earlier this year in Newcastle, England.

PHOTO BY SAM REYNER/EMI MUSIC



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— ROSANNE CASH



EXTENDED FAMILY

Top to Bottom: Rosanne Cash's parents, Vivian and Johnny; Cash with her father, Johnny, in 1957; Cash with her sisters and mother, Vivian, in Nashville in 2003 (left to right: Tara, Rosanne, Vivian, Kathy, and Cindy).

ONE LONG HEADACHE

"I've had headaches for as long as I can remember," says Cash, who lives in New York City and has made a name for herself over the last four decades as a musician and a writer. She has been nominated eleven times for a Grammy and won the 1985 award for Best Female Country Vocal Performance. Her 2010 memoir, *Composed*, was critically acclaimed.

Cash's headaches worsened during her second pregnancy. By 1994, they were so severe that she finally consulted with a neurologist. Still, it wasn't until 2007 that Cash's Chiari I malformation was accurately diagnosed. The first neurologist Cash went to thought the singer was experiencing cluster headaches—an exceedingly painful and relatively rare kind of headache that tends to occur in a cyclical pattern—but the medications she prescribed offered little help.

The second neurologist, a headache specialist, diagnosed Cash with migraines. When the headaches continued and intensified, the diagnosis changed to atypical migraines.

"This went on for a decade," Cash says. "A decade!"

Dr. Singh says such a delay in diagnosis can be common in these cases. "It is not unusual because the headache is not persistent, often brought on only by certain activities" she says. "The condition is extremely rare and has very indistinct or mild symptoms that are not present all the time."

Rosanne Cash even wondered if she might have a Chiari I malformation after discovering the term online. She discussed it with the headache specialist, but an MRI came back negative. Although most experts consider MRI to be the best way of diagnosing Chiari I malformation, it isn't flawless, according to Dr. Singh, in part because a malformation can change over time.

The singer frequently experienced neck pain and stiffness, and at times her headaches were severe enough to knock her off her feet. "Sometimes it felt like someone had hit me in the back," she says. "Once I even dropped to my knees, the pain was so intense." Her general practitioner determined that she had Lyme disease. "But after she treated me for Lyme disease, nothing changed," Cash says.

During these years, Cash tried a number of treatments and approaches to managing her pain, including migraine medications, yoga, acupuncture, massage, and chiropractic adjustments. Most offered temporary help at best. "Sometimes not even temporary," Cash recalls.

A KARMIC PHONE CALL

By 2007, Cash's discomfort was such that she could no longer even do yoga, a practice she had enjoyed for 15 years. On a day when she felt such pain that she was hunched over and

PHOTOS COURTESY ROSANNE CASH

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barely able to walk, Cash searched the Internet for a neurologist specializing in Lyme disease. She found Norman Latov, M.D., Ph.D., director of the Peripheral Neuropathy Clinical and Research Center at Weill Cornell Medical College in New York City, member of the AAN, and author of the AAN patient book *Peripheral Neuropathy: When the Numbness, Weakness, and Pain Won't Stop* (Demos, 2006).

Dr. Latov had treated her father, Johnny Cash, for peripheral neuropathy—and had even approached Johnny about doing a public service announcement to raise awareness of the condition. When Cash recalled these family connections to Dr. Latov, she thought, “This is too weird. I’ve got to call him.”

That karmic call would change Cash’s life. Two weeks after an initial visit, he made a diagnosis based on her symptoms of chronic headaches and regular pain and stiffness across the upper back, arms, and legs. “The cervical spine MRI revealed a Chiari I malformation with syringomyelia,” Dr. Latov says. “The diagnosis was made based on her symptoms, the MRI results, and the absence of other explanations for her symptoms.”

TREATMENT OPTIONS

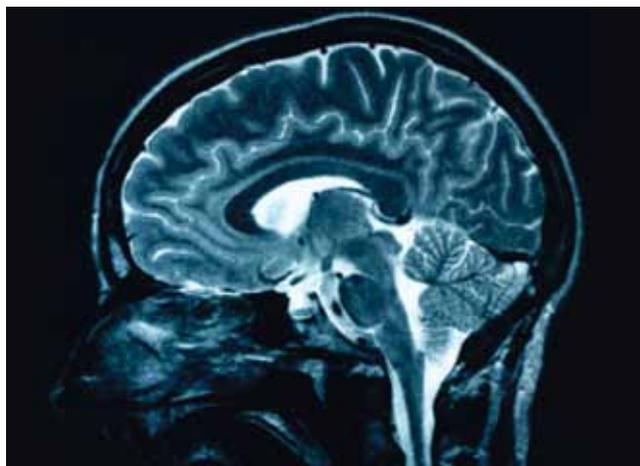
Treatments for Chiari I malformations can vary depending on their severity. Some patients never even know they have the condition; others do but don’t experience any symptoms. Patients with mild symptoms might opt to be treated with pain relievers such as acetaminophen or ibuprofen or migraine medications, especially if there is no associated syringomyelia. But for someone like Cash—that is, who has a Chiari I malformation and a syringomyelia with severe disabling headache and/or other neurologic symptoms—surgery may be the only choice.

“I was so relieved,” Cash says of her diagnosis. “Even though I figured I would have to undergo brain surgery, I was flooded with relief that someone finally knew what was wrong with me.”

Guy McKhann II, M.D., associate professor of neurological surgery at Columbia University Medical Center/New York Presbyterian Hospital, believes that unless a patient is debilitated by the condition, surgery may not be worth the risk. “I think that surgery for Chiari I malformation should be restricted to people who have clear-cut symptoms from a syrinx, which is when the hole in the spinal cord fills with fluid, or a syrinx that is worrisome and enlarging at a significant pace, or who have symptoms caused by the pain at the back of the head,” Dr. McKhann says.

“Of all the surgeries I perform—for tumors, epilepsy, Parkinson’s disease—the procedure for Chiari I malformation involves the most difficult recovery,” says Dr. McKhann.

In surgery for a Chiari I malformation, a cut is made in the back of the skull and a piece of bone is removed. A piece of tis-



Chiari I Malformation: The Basics

- ▶ **HOW COMMON IS CHIARI I MALFORMATION?** According to a study published in the *New England Journal of Medicine* in 2007, 0.9 percent of normal adults had the minimal criteria to diagnose Chiari I malformation based on an MRI. That would equate to nearly three million people in the United States.
- ▶ **WHAT CAUSES IT?** The displacement of the cerebellum into the spinal cord results from a lack of space in the skull, but the reasons for that lack of space vary. In many cases, the cause is unknown.
- ▶ **WHAT ARE THE SYMPTOMS?** Chiari I malformation can cause headaches, neck pain, dizziness and numbness, and problems with sight or hearing.
- ▶ **WHAT IS SYRINGOMYELIA?** A syringomyelia is a hole that forms in the spinal cord, most frequently resulting from trauma to the spinal cord, tumors in the spinal cord, or Chiari I malformation. It often affects the hands first, leading to weakness or numbness and an inability to feel temperature or control fine motor skills. Because a Chiari I malformation can interrupt the normal flow of cerebral spinal fluid between the brain and spinal cord, the hole in the spinal cord can fill with fluid, resulting in a cyst (syrinx). If the syrinx expands or enlarges, it can cause additional compression in the spinal cavity, resulting in pain, muscle weakness, and stiffness.

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sue is then added to the membrane around the brain (called the dura) to make the space larger. This alleviates some of the pressure and allows the cerebellum and its tonsils to stay inside the skull where they belong. It also frees the cerebral spinal fluid in the area to circulate normally. A normalized circulation of cerebral spinal fluid should also lead to the shrinking of the syrinx, decreasing the pressure on the spinal cord and lowering the chances that neurologic symptoms would continue to get worse.

CHOOSING SURGERY

For Cash, surgery was her last hope. After her diagnosis, she had tried acetazolamide (Diamox), an altitude-sickness medicine that is supposed to help alleviate pressure in the head. It didn't help. The pain medication pregabalin (Lyrica) offered her some relief, but it caused unwelcome side effects, including weight gain. So after her husband, John, went into “perfect patient advocate mode,” as Cash puts it, researching this country's preeminent experts on this condition, Cash found herself scheduled to go under Dr. McKhann's knife in November 2007.

The singer prepared herself for surgery with hypnosis and dark humor. “I called up a friend who had ovarian cancer, and we made sick jokes to each other about illness and death,” Cash laughs. “It was helpful.”

During surgery, Dr. McKhann found an obvious Chiari I malformation. “In many patients, there is a compression line right across the cerebellar tonsils where you can see they have been compressed by the lack of room,” Dr. McKhann explains. “When I performed Rosanne's surgery, I could see very clearly how much compression there had been.”

To alleviate that compression, Dr. McKhann grafted a piece of synthetic material to the membrane covering Cash's brain. “The graft holds all the spinal fluid in yet allows room for the tonsils where there wasn't before,” Dr. McKhann says. Cash's surgery was completed when her head was sewn up and her skull shut with nineteen staples. In keeping with her dark sense of humor, Cash often referred to herself as a “zipperhead” after the surgery.

RECOVERING FROM SURGERY

As predicted, the recovery was difficult. “It was hard, really hard,” Cash says. She spent a week in the hospital, on morphine and steroids, trying to relearn some of the basic skills she'd once taken for granted.

At home, Cash tried to recuperate with rest and medications, but the process was slow. Too many people or too much noise would overwhelm her, leading her to seek refuge in the quiet and the dark. “I would spend a lot of time on this sofa,” she says in her townhouse in New York's artsy Chelsea neighborhood. “I just hurt all the time.”

The singer had scheduled her first performance after the surgery for April—a full four months later—but even that schedule was too optimistic.

“That plane flight just about killed me,” she says. It would be a year before Cash took the stage again, when the next October she travelled to Germany for the Century of Song Festival, part of the annual Ruhr Triennale.

Although Cash once scoffed when a fellow zipperhead explained that her complete recovery took seven years, Cash concedes she wasn't exaggerating. More than four years after her surgery, she still feels its effects. She has yet to begin yoga or exercise again (other than mild physical therapy). She still experiences some headaches from certain low noises and changes in barometric pressure, although they are much less severe and less frequent than before. And she continues to have discomfort in her neck—probably because her top vertebrae had to be broken during surgery.

Difficult recovery or not, Cash's surgery was a success. According to Dr. McKhann, the purpose of the surgery was two-fold: to “help with her debilitating pain” and to “stabilize and allow her syrinx to shrink.” Cash says, “It's much better than it was.”

Looking back on it all, the singer has only one regret: she wishes she hadn't placed so much faith in so many “new age” practitioners of alternative

therapies to deal with her problems. “I still have a lot of residual anger towards some of these people—many of them basically told me it was my fault for 10 years,” Cash says. “You know, that it was my hormones, or my stress, or my holding in my anger.”

In the end, Rosanne is glad to have entrusted her health to the neurologists and neurosurgeons. “I have nothing but the utmost respect for the people who fix brains,” she says. NN



THE NEW LIFE

Top to Bottom: Rosanne Cash with her husband, Jon Leventhal; with daughter, Carrie, in Montana, 2007; and with her son, Jake, six weeks after her surgery for Chiari.

PHOTOS COURTESY ROSANNE CASH