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'I've got to see this through': Michigan woman dying of Lou Gehrig's disease fights to make her final days meaningful

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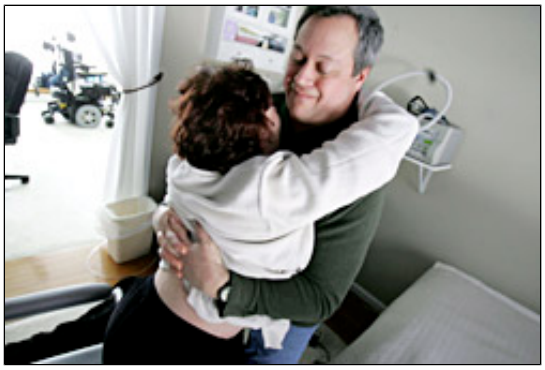
Akiller is stalking Deneane Chiplock and her family.



Photos by ERIC SEALS/DFP
Jerry Chiplock, 47, lifts Deneane Chiplock so he can place her in bed. He is now staying at home full time. "I'm just trying to get through the day to day," he says.

She watched as a rare hereditary form of amyotrophic lateral sclerosis (ALS) -- often referred to as Lou Gehrig's disease -- killed her mother at age 36 when Chiplock was 16. Ten years later, she watched an aunt who was like a second mother to her die the same death. Then there were her great aunts, numerous cousins, her grandfather.

Then, in 1994, Chiplock got the news she'd been waiting years to hear. She could finally take a test to learn whether she carried the defective gene responsible for her family's affliction.

"I didn't hesitate," says Chiplock, then pregnant with her second child. She sent her blood sample to researchers the next day. "I had responsible decisions to make -- housing, my family, our finances, everything." She waited months. It was a beautiful September day when the genetics counselor from Northwestern University called with the good news: She did not have the defect.

The dark cloud that had hovered over her since she was a teen suddenly lifted.

Chiplock drove home from work, music blaring, tears of joy streaming down her face as she wondered: "How did I get this lucky?"

But her thoughts soon turned to her two siblings, Dennis and Anita. After all, she thought, if she had dodged this bullet, one of them might have it. But mostly Chiplock was elated for her sons, who would never inherit the disease.

"I didn't have to worry about ALS anymore."

Disheartening news

Two months later, the genetics counselor called back.

"We made a grave error," Chiplock recalls hearing. "They mixed my test up with my remaining aunt's," she says of her mother's only sibling who escaped the curse of ALS.

She said she felt a rush of "unbelievable guilt" and vowed never to have another child.

"What have I done to my boys? What if they inherited the gene?"

It's a question that still haunts Chiplock of Saginaw Township, who in November 2003 became the 25th person in three generations of her family to be diagnosed with hereditary ALS.

Each of her sons, Sean, 14, and Evan, 10, has a 50 percent chance of inheriting the disease.

"People have asked, 'If you knew this was in your family, why did you have kids?' I wrestle with that all the time," she says. "I didn't want to find out 40 years down the line I never had the gene."

Now she focuses on the conviction that medical conditions will be better for them in the future and, with her time growing short, she is doing everything she can to be part of an answer.

A promising drug

HOW TO HELP

The Chiplock Family ALS Fund is auditable and registered with the Saginaw Medical Federal Credit Union.

Donations help pay only for ALS-related expenses. After Deneane Chiplock's death, any money left after those expenses are paid will be split among three organizations: Project ALS, the ALS Association and the ALS division of the Muscular Dystrophy Association.

To help, send donations to:

Chiplock Family ALS Fund

Saginaw Medical Federal Credit Union

4550 State Street

Saginaw, MI 48603

Call 989-791-7070

For more information on ALS, contact the ALS Association at 800-782-4747 or go to www.ALSA.org.

Chiplock has been using herself as a human petri dish since May in an independent trial of an experimental drug that, in lab studies, has shown promise in slowing the nerve degeneration of ALS. She also has become a vocal advocate for legislative changes for in-home care that would benefit ALS patients and their families.

Physical therapist Sandy Ellery massages Deneane Chiplock. Families pay up to $200,000 a year for the care of a loved one in the advanced stages of ALS.

It has not been easy. She even contemplated suicide in August. "I've had so many people tell me I'm one of the strongest people they know and how courageous I am. Those words aren't even in my vocabulary for myself. I was barely coping with this disease. I still feel that way," says Chiplock. "But once I made the decision that I've got to see this through, it's like now I've got to live with that decision."

'Buried alive'

ALS destroys motor neurons, typically debilitating its victims through progressive wasting and paralysis of the muscles. Patients ultimately lose the ability to swallow and breathe. The brain, meanwhile, remains alert.

"I equate (it) with being buried alive," says Chiplock.

Of the 30,000 Americans who have ALS at any one time, most cases are sporadic, or of unknown origin. From 5 percent to 10 percent are hereditary, affecting about 700 families nationwide.

The gene mutation that causes ALS in Chiplock's family and in some others involves a genetic defect on chromosome 21 that causes a mutation of the enzyme known as SOD1. The mutation, which alters the enzyme's ability to protect against free radical damage to motor neurons, was discovered in 1993 by Dr. Teepu Siddique, a genetics researcher at Northwestern University.

Chiplock's family was integral to that discovery, having been one of the first to submit blood samples for testing.

According to Siddique, who knows Chiplock well, most people with the mutation get the disease by age 47. Chiplock was 39. (The average age of onset for ALS is 55.)

"It's a very, very bad outlook," he says. "This comes in the middle of life, at the most productive, happy period of people's lives. It's a horrible thing. And the mind is intact, especially with this form of ALS."

Since the discovery of the defective gene, it's been left up to individuals to decide whether to be tested. Chiplock's sister waited four years before getting tested; her brother doesn't want to know. As for her sons, she says, "We made the decision to let the boys decide" later in life.

Jerry Chiplock displays a photo of his wife and sons. Deneane Chiplock has made a three-hour video for the boys to watch when she is gone and is working on memory trunks for each of them.

Although the average remaining life for a person diagnosed with ALS is two to five years, no one in Chiplock's family has survived more than 13 months after the onset of symptoms, she says. The youngest victim died at age 21, the oldest at 70.

The reasons she has lived longer than 13 months are unclear. Perhaps it's because she knew the symptoms well and was diagnosed early. Or perhaps the experimental drug is having some effect.

The drug trial

Because of her own efforts, Chiplock is the most closely monitored patient currently on Rocephin (generic name: Ceftriaxone), an antibiotic approved for treating meningitis that showed promise in slowing the progression of hereditary ALS in scientific studies last year.

Last May, already six months into her diagnosis, Chiplock learned of the research and the human trial that will start later this year, too late for her.

Eager to get on the drug while she was still able to help prove its possible benefits, she contacted Dr. Jeffrey Rothstein, professor of Neurology and Neuroscience at Johns Hopkins University, who led the lab studies. According to Rothstein, a collaborator on the upcoming human drug trial, the drugs being used to treat ALS, including Rocephin, won't reverse damage done by the disease. They can only slow its progression.

"It doesn't give you anything back. The best possibility would be for it to stop the disease."

Chiplock started the drug in May -- intravenously twice a day -- under the care of her neurologist, Dr. David Simpson, director of the Muscular Dystrophy Clinic at Michigan State University in East Lansing. Simpson, who encouraged Chiplock to get on the drug, monitors her progress through weekly blood samples and monthly appointments.

He says that with Chiplock, he has not seen any of the expected side effects, such as kidney failure, gallstones and infections.

"We've learned a lot," he says. "Clinically, she's slowed down from where I'd expect her to be."

Indeed, Chiplock says: "Everyone is stunned that my bulbar skills (speaking, chewing, swallowing) are still intact. I'm not slurring my words."

Meanwhile, the failure of her right diaphragm several weeks ago lessened Chiplock's expectation that she would live until February. Her breathing is now aided by a bipap machine that assists her breathing. It's one step away from a ventilator, she says, something she insists she will not go on.

Living with ALS

A visit with the Chiplocks one Sunday in late November revealed a life that seemed almost normal. Chiplock's husband, Jerry, 47, dressed in jeans and a gray Heritage Hawks T-shirt from his son's high school, answered the door with a smile, apologizing for their two barking dogs, Nikki and Pepper.

Evan played video games in the kitchen of the two-story, four-bedroom brick home, while his mother, then able to sit up in a motorized chair at the table, munched on a bagel.

Sean was off practicing for a school play.

The progression of the symptoms of ALS differs from person to person.

For Chiplock, it started with the loss of the muscles in her right leg. She cannot stand on her own or use the bathroom without help from her husband, the only one strong enough to lift her.

She last used a pen in early September when, within a matter of days, she lost the grip in her right hand. Her left arm is now going numb, although she can still feed herself and cradle a phone. "There are things I'm learning I never knew before, like the amount of pain involved," says Chiplock. "It's claustrophobic."

Sharing her double hospital-style bed with Jerry is more than an emotional comfort, she says. Jerry needs to be there in case she needs a pat on the back. "There've been times when I've nearly choked on my own saliva," says Chiplock.

The bed also serves the whole family on Sundays, when everyone, including the two dogs, hops in to cuddle. Although Chiplock can't reciprocate, she says, "It's a blessing that you can still feel somebody holding your hand or kissing you." Although she's able to use her motorized wheelchair on occasion to see her sons' basketball games and swim meets, she's mostly trapped in bed.

"It's pissing me off," says Chiplock, who can be demanding. She'll order Jerry or the boys to do things she's frustrated that she cannot do for herself, such as looking something up on the computer.

"We bicker," she says. "We all yell. Everyone's exhausted. Jerry and I will have blowouts, and we'll get past it. You just blow off steam."

Chiplock's sister, Anita Crawley, 38, a pediatric nurse practitioner from Buffalo, N.Y., sees the strain.

"I'm seeing moments where she is not handling it well," she says, but notes, "She's got a ton of strength." Crawley, who tested positive for the defective gene, is a single mom with 7-year-old twins.

"She's taught me to keep doing what you can to try to find an answer for our kids. That's the biggest thing."

Aside from Chiplock's fears that her sons will inherit the disease, she worries about what it's doing to them now.

She's glad Evan is sharing his feelings. "We talk about how scared we are. We'll have a really good cry, and we decide to just say: 'OK, we've had enough crying. We love each other.' We hug and try to just move on."

Sean is causing them more concern. Although he's coming to terms with the disease, Chiplock says she sees a lot of herself in him.

"He was in major league denial. He (was) doing a lot of what I did when my mom was dying, secluding himself and dealing with it on his own." Sean admits he at times tells himself that something will happen and his mom will get better.

"I know that's probably not going to happen," he says, "but my mind refuses to let me get pessimistic."

The cost of ALS

ALS takes a crippling financial toll.

Depending on costs, the average family can expect to pay up to $200,000 a year for the care of a loved one in the advanced stages of the disease, according to the ALS Association.

The Chiplocks are no exception.

Although they did what they could to plan ahead, Deneane Chiplock's family history and positive gene test did them no favors when it came to qualifying for long-term care and life insurance policies. They are paying $1,500 out of pocket every month for a home health aide, who works 40 hours a week. When Chiplock gets to the point of needing round-the-clock care -- and she's arguably there -- the nursing care cost alone will be more than $10,000 a month out of pocket.

"We haven't even come to that yet, and we can't even pay for what we have now," she says.

The family also pays about $1,440 out of pocket a month for Chiplock's ALS drug cocktail -- 34 pills she takes each day, including Rilutek, the only approved drug to help slow the progression of ALS, and a box filled with pharmaceutical-grade vitamins and supplements.

Rocephin is another story. The experimental drug costs $9,000 a month. The price is exorbitant because the dose is much higher than what the drug is approved for. Because it's considered experimental, insurance won't cover it.

Right now, Chiplock says, the family owes more than $80,000 for the drug.

The Chiplocks have used what retirement funds they could tap.

They've re-mortgaged their home and cashed in what they could of a $70,000 life insurance policy.

They put what they can on Visa but have to pay off as much as they can each month to maintain a $14,000 credit card limit.

Some relief came last year when family and friends in Buffalo and Saginaw started fund-raising efforts.

The efforts, since consolidated as the Chiplock Family ALS Fund, have raised about $29,000, says Chiplock, who insisted the fund be auditable. After her death, she says, any money left over after paying for ALS-related expenses will go to three ALS charities.

Loving husband, busy dad

Much of the load for his wife's care falls to Jerry Chiplock, who is exhausted by running the household and chauffeuring his sons to school and other activities.

Meanwhile, he worries about his job. He's been taking time off from his job as vice president for human resources at St. Mary's Medical Center in Saginaw through the Family Medical Leave Act, which guarantees 12 weeks off work with job protection but no pay.

Since August, when his wife had a nasty fall on the kitchen floor, he's been home, at first two days a week and now full time. He's kept up his salary by using sick days and vacation time, but by the middle of this month, his 12 weeks will be up, he says.

Technically, he could lose his job or be forced to return to work, which would require hiring help. It also could force him to be away from his wife at the most critical time. Despite all he is doing, Jerry Chiplock feels guilty for not doing enough.

"I feel like I'm not doing my job justice, not doing my home life justice, not doing Deneane's situation justice. I'm not acting as an advocate for her the way she's acting on her own behalf. I'm not calling senators and writing letters," he says. "I'm just trying to get through the day to day. I live in constant fear of getting sick. If I spend one day in bed with a cold or flu, she is stuck."

Although many friends have fallen away or just don't know what to say or do, the Chiplocks are thankful for friends like Sandy Arbitter. "She has been my rock," says Chiplock. "She was here four days a week initially just helping me get through the shock of everything." These days, Arbitter arrives each Thursday to clean house and keep up on the laundry.

"I'm not sure what to do to help her other than to be there. It bothers her to be dependent on other people," says the retired nurse, who is in awe of Chiplock. "It continually amazes me how she maintains such control over her environment and how she stays on top of things."

The future

As Chiplock looks ahead, she's unable to focus on her death.

"I'm really afraid about the end. It's the unknown," she says.

What causes her the most anguish, however, is leaving her family.

"I don't know if there's anything beyond this. But if there is something more of me existing without them, it breaks my heart. I know that their lives will go on. I know they're going to have joy and happiness and love and laughter. I just wish I could be a part of it."

And Chiplock is finding ways to be part of their future. She made a three-hour video for the boys to watch when she is gone and is working on memory trunks for each of them, chests filled with things like the Bible, family cookbooks and their first teddy bears.

Meanwhile, she has kept up as best she can with e-mails, and she posts her views on message boards like BrainTalk, an online patient support forum regarding neurology. More recently, she has offered what support she can to ALS patients who have called to ask about her drug trial.

Despite the unimaginable stresses in the Chiplock household, family members remain remarkably strong. They joke, laugh and even poke fun at the disease itself.

Chiplock, who is donating certain body parts to science and wants the rest to be cremated, has been making special arrangements for her ashes. She tells her husband and her sons she has three adventures planned for them to dispose of the ashes, including taking some on her favorite amusement park ride, the Millennium Force roller coaster at Cedar Point in Ohio.

She has a fourth adventure for her husband, although she is still figuring out the details.

"You're going to be a pain in the ass in death, too, aren't you?" he says.

"Yeah," she answers. "Pretty much."