Early on, Jerry Chiplock believed his love for Dee would protect her

The Family Curse

Twenty-four of her relatives had this deadly disease. Dee Chiplock, the 25th, is determined to fight it.

BY JAN GOODWIN

ON A SUNNY DAY LAST JULY, Dee Chiplock hoisted herself into her Honda Odyssey, equipped with hand controls instead of foot pedals. She pulled away from her Saginaw, Michigan, home and headed toward the Canadian border. On the seat next to her was a plastic bag containing a deadly dose of prescription pain medication and tranquilizers. Eight months earlier, the 40-year-old mother of two boys had been diagnosed with ALS, a degenerative nerve disease with no known cure.

Dee wept as she drove. She thought back to the day 18 years before when her husband, Jerry, proposed to her. At an elegant restaurant on the Florida coast, a waiter served Dee her dessert: a velvet ring box containing an exquisite marquise diamond. Amused diners looked on as Jerry got down on one knee, but their smiles faded when Dee started to cry. "I love you so much, but I can't say yes," she said. She was certain she would one day end up with ALS. "I don't want to put you through that."

Amyotrophic lateral sclerosis, also known as Lou Gehrig's disease, which

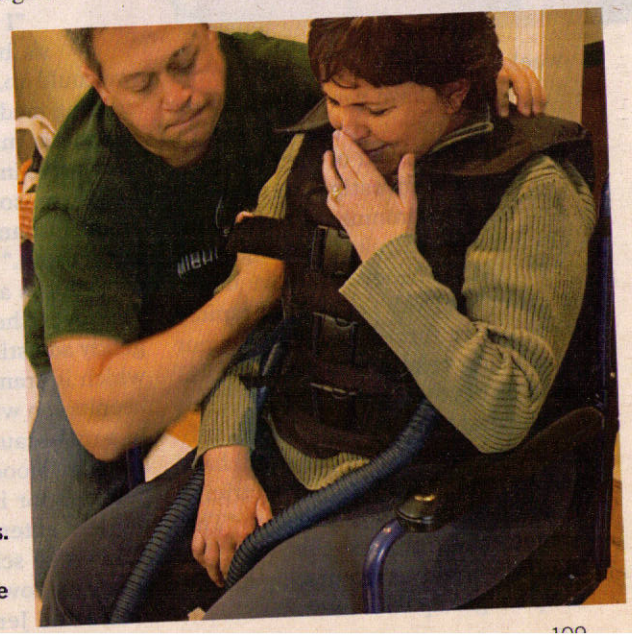
everything Dee wanted it to be. It got even better in 1993 when she became pregnant again, this time with Evan. But in her seventh month, Dee received a call from Northwestern University's Dr. Teepu Siddique, one of the country's leading ALS researchers and someone Dee had been in touch with for years, searching for answers about the disease. His team had made a major breakthrough: They'd identified the gene that, when mutated, causes ALS in 20 percent of familial cases. That gene, he told Dee, had been found in blood her family donated.

"I was stunned," Dee recalls. "The timing couldn't have been worse. Here I was expecting my second child, and the doctors finally figured it out." Dee asked to be tested for the gene.

Seven months later, Dee was working late when Siddique called to tell her the results of her test were negative—she didn't have the flawed gene. "I was elated," she says, "on cloud nine. Suddenly, I had my life before me. I'd be here when my children grew up." On the drive home to tell Jerry, the music on the radio sounded so rich, and the colors of the night sky were so vibrant. When she walked in the front door and shared the news, both she and her husband cried.

Their joyful moment didn't last long. Six weeks later, a genetic counselor from Northwestern phoned. "We got one of your tests back," she told Dee, "and it was positive. Because it conflicted with your previous results, we re-ran it four more times. You *do* have the gene." With the type of testing done, results were subject to false negatives.

"It was like being hit by a tornado," Dee remembers. She felt tremendous guilt. Oh, my God. What have I done to Sean and Evan? she asked herself. "Because of me, they have a 50-50 chance of inheriting the ALS gene. Two



Dee breaks down after being fitted with a vest that stimulates her lungs. "I am sick of being the one everyone has to take ...e of," she says.

Jerry, then 31, and Dee, 23, wed in Long Island, New York, summer 1988.

COURTESY DEE AND JERRY CHIPLOCK

afflicts some 30,000 Americans, refers to the loss of signals that nerves normally send to muscles. A creeping paralysis gradually robs a sufferer's body of the ability to move. As paralysis limits the ability to speak, chew, swallow and ultimately breathe, the person can literally suffocate. Yet the patient's mind usually remains fully functional. Sensation, including physical pain, does not disappear. "It's like being buried alive," says Dee.

Dee was 16 when she watched the disease take her 35-year-old mother's life. Two of her mother's aunts, two of her mother's siblings, and her grandfather all died the same way. She is the 25th member of her family known to have the disease, and among them the only one still living.

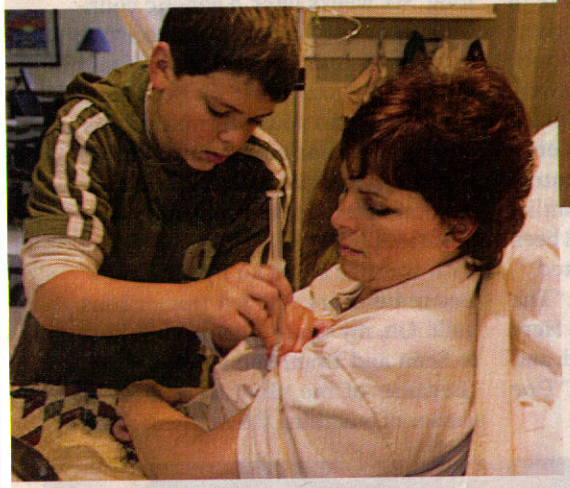
When Dee drove to Canada, she could barely walk with a cane and had no business being behind the wheel of a car. The former senior account executive with a telecommunications corporation had never been a quitter. "But I had reached a breaking point," she remembers. "I didn't want my sons and husband to see what I was about to go through. It's not pretty to watch. I know. The people I loved most in the world have been taken away from me by this disease."

Her plan was to find a peaceful spot in Canada and swallow the pills she'd been saving. "But as I drove around Lake Huron," says Dee, "I knew what I really wanted was for Jerry and the boys to be with me. We had little enough time together as it was." She turned around and headed home.

ON THE NIGHT he proposed, Jerry, an administrator at a small Catholic charity hospital, told Dee, "Whatever we face, we face it together." From the moment he put his arms around her, Dee became hopeful. "I told her my love would protect her from the disease," he says today. "I was wrong."

When Dee and Jerry married, medical researchers believed that ALS might, in some cases, be hereditary. "But I was told over and over that they hadn't identified a cause," she explains. "So, trying to live a normal life, we decided to have kids."

Their first son, Sean, now a witty, articulate teenager, was born in 1990. Life with Jerry and their boy ...



Left to right: Son Evan, 10, flushes a line that delivers an experimental drug; Dee's hairstylist now makes home visits; and the family (son Sean, 14, far rear) still relishes time around the kitchen table.

When Dee's mother was diagnosed in 1980, she completely withdrew, refusing to speak to anyone. Dee, then a teenager, begged her to fight. "Mom, don't give up!" she said. "We'll find the answer."

"I was upset by her diagnosis, but I wanted to know what we could do," she recalls. Dee started reading medical books, talking to researchers, making phone calls. "I was very naive in thinking that somebody out there, somewhere, knew how to treat this, and we just had to get to that person."

"Dee has always been a fighter at whatever she's tackled," says Jerry, gently adjusting one of his wife's legs, which is cramping. "I've had trouble keeping up with her at times." It was in part because of Dee's activism—donating blood to the researchers she pestered for information, encouraging her extended family to do the same—that scientists discovered the gene that now dooms her.

Dee and Jerry believe their so-

weeks later, Jerry had a vasectomy."

Scientists have yet to identify the genes that may cause ALS in the remaining 80 percent of familial cases, and are now investigating whether something environmental triggers the disease. The difficulty in finding a cure is that ALS is believed to have multiple triggers. Dee's family has a rapidly progressive form of the disease; not one of her relatives has survived 13 months beyond onset.

The first signs of Dee's illness surfaced in August 2003. "I started having strange sensations of electricity running down my leg," she says. "The symptom warned me that what I'd always feared was my destiny had finally caught up with me."

should make the decision about testing themselves, when they are 18. "Not everyone wants to know how and when they are going to die," Dee explains. Her sister, a 38-year-old nurse-practitioner and single mother of twins, has learned she also has the gene, but Dee's brother, a steelworker and father of two, refuses to be tested. "I don't want to know," he says. "It's a death sentence. If I tested positive, I would kill myself, drive off a cliff."

In the end, Dee backed out of committing suicide for the sake of her children. She thought about how badly she'd wanted her mother to fight, and knew that Sean and Evan would feel the same way. "All their lives, I've told them never to give up," she notes.

On the way home from Canada, Dee thought about the battles she wanted to wage before she died. She wanted to work for a cure for ALS, but she also wanted to improve conditions for people suffering from all terminal diseases. "I wanted to make my life and death mean something," she says.

So for the last eight months, Dee has jumped ahead of traditional medicine and begun taking ceftriaxone, an antibiotic designed for short-term use in patients with serious infections such as meningitis and pneumonia. It has shown promise in slowing ALS in animals, but has yet to be tested on humans. Clinical trials will begin this spring. "I will be dead by then," Dee says matter-of-factly. Though the drug is not expected to offer a cure, its potential to prolong life is why Dee has agreed to take it "off label."

Twice daily, she receives a 30-minute infusion through a line permanently inserted in a vein in her arm. Often it is administered by Jerry or by Sean, now 14, or Evan, 10. She must

pay a whopping $9,300 per month for the drug, as neither her insurance company nor the drug manufacturer will cover off-label medications.

"Dee is not doing this just for herself," says her neurologist, David Simpson. "We'll gain scientific knowledge from her."

"Why not experiment with me, since I'm dying anyway?" Dee comments. It's encouraging that she has experienced none of the serious side effects—kidney failure, fungal infections, gastrointestinal problems—that some feared would occur with ceftriaxone's long-term use. And Dee believes she is still able to talk this many months into the disease because of the drug. "Had I been able to start it five or six months earlier, when I was first diagnosed, I might still be able to walk and write."

But no drug can help the bankrupting aspect of ALS and other diseases that gradually incapacitate patients. "If I can be a catalyst for change in this one area, it will be my legacy," she says. The average cost to care for an ALS patient can run $220,000 in the first 18 months after diagnosis, according to Dee, but the average insurance program covers just $35,000 to $45,000.

Much of the cost is for custodial nursing care that is not covered by Medicare—"feeding patients, bathing them, wiping the drool off their faces so they can stay at home," Dee explains. "The absolute blackest nightmare for me," she remarks, "is that I will have to go into a care facility. I want to be a part of my family's life, to spend every minute I can with them, to be able to chat, or watch television while cuddling up on my bed together." She pauses, then adds, "Every terminally ill patient feels the same way."

When Dee does start getting around-the-clock care, her family will have to find $10,400 a month to pay for it. With the ceftriaxone, that's $20,000 out of pocket monthly. "How many Americans do you know earn that kind of money?" she asks incredulously. "My boys' and husband's futures are being burned up by the catastrophic expense of this disease. We're going to lose this house. We've already spent my sons' college funds. After I'm gone, Jerry and the kids will be left with a mountain of debt. We're a solid middle-class family with good medical and disability insurance. If we can't do this, how can anyone?"

In her race against time to improve the system for terminally ill patients, Dee has met with legislators, including Sen. Hillary Clinton. She has given speeches advocating a home-care alternative. "I want 15 minutes—*15 minutes*—in front of the full Senate," Dee challenges. "I know I could compel them to make some kind of change."

Dee's breathing is now inhibited by ALS; she's lost her cough reflex, which could cause her to choke to death on her saliva. "I'm afraid of dying, or rather of not living," she admits, her voice quavering. "I'm afraid of not being with the people I love. And I worry about the boys and Jerry [illegible]

I'm not here for them. Jerry's a great dad, but there are just some things that moms do better."

Jerry has taken a leave from work to help care for Dee. They are trying to relish their time together. And Dee is working passionately to prepare Sean and Evan for a future without her. She is filling two large wooden memory chests, one for each boy, with "favorite photographs, family one-liners, Christmas decorations—any little memento that is meaningful to us." She's burned onto CDs recipes of their favorite meals, as well as their drawings from different grades—including one Sean did at age 7, of him playing with his brother under a sky full of hearts. She's also collected papers such as Evan's from the fourth grade about his dreams for the future, which include finding a cure for illnesses like his mom's.

Unlike her mother, who would never discuss ALS, Dee talks freely about her disease. "It's better they know everything," she says. The boys are aware every minute is precious. "I have to get the good times in with Mom now," Evan observes, "because I'm not going to have a lot longer to do it."

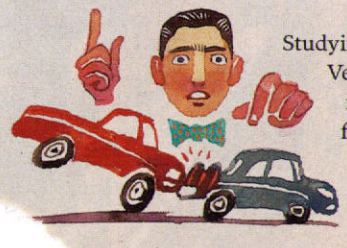
"I'm racing to cram in all the life lessons I thought I'd have a long time to give them," Dee says, tears flowing. She's made a three-hour video telling her sons the things she would have told them as they face milestones: first dates, first jobs, marrying. "I want them to be able to find relationships as good as the one Jerry and I have.

"I also want them to have the courage to know they can make a difference," she continues. "Take me. I'm just another tragic dying person. But if I can effect change before I die, thousands of Americans will benefit—not just ALS patients, but those with Alzheimer's, Parkinson's, end-stage cancer. I want my sons to understand that kind of possibility."

For more information, contact the ALS Association: 800-782-4747; alsa.org.

DRIVE-BY HOOTING

The state of California sure watches out for its citizens. Case in point, the 2004 California Driver's Handbook contains this footnote: "Throughout this handbook, the term 'thumb print' will be used to mean a thumb print or fingerprint, if you have no thumbs." CINDY GUAN



Studying the Oregon Commercial Motor Vehicle manual afforded me the opportunity to learn this important driving factoid: "According to accident reports, the vehicle that trucks and buses most often run into is the one in front of them." KIMBERLY RIDDELL