'brutal and devastating' ALS pulls Big Island family together

'Every time he loses an ability to do something, it's heartbreaking'

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Nohea Nakaahiki was starting to develop a complex a few years ago.

Her husband, Clarence "Bully" Nakaahiki, would look at her and start to laugh, for apparently no reason.

Months later, as the couple watched videos about the effects of amyotrophic lateral sclerosis, Nohea realized Bully wasn't laughing at her. Spontaneous laughter is one sign of the bulbar form of ALS, a form in which the first muscles to progressively degenerate are those that are used for chewing, swallowing and speaking.

Doctors diagnosed Bully with bulbar ALS in July 2006, more than six months after he began experiencing difficulties swallowing and his speech began to slur. He'll now also cry spontaneously, and the laughter has become more frequent.

"It could still be my face," Nohea said, smiling.

No longer able to talk, Bully just shook his head at the comment.

The Nakaahikis at first didn't believe the diagnosis, Nohea said.

"I said, 'No way, he golfed last week,'" she said, recounting the day a doctor gave the diagnosis. "From what I read, it starts with arms or legs. (At that point) he's still driving, he's cooking, he's golfing."

They requested a second opinion and were sent to the University of California San Francisco Medical Center, where the center's director provided confirmation. Even then, as doctors said the progression could be slower than anticipated because of Bully's size -- he weighed about 245 pounds at the time -- and otherwise good health, the couple's disbelief continued. Bully didn't start taking his prescription, Rilutek, the only FDA-approved medication that slows the disease, right away. He remained active.

Nohea began recording everything about the disease she describes as "brutal and devastating" and credits her legal background with her ability to deal with the insurance providers. Binders filling two bookshelves document her requests for equipment, like the eye response reader Bully recently acquired that, with its preprogrammed phrases, will allow him to communicate again. Not every request has been granted; the couple borrowed some equipment from ALS Association "loan closets" when an insurance provider repeatedly issued denials.

A timeline shows when certain symptoms began, when Bully caught a cold, when he lost certain motor skills and when he felt muscle weaknesses. Things progressed more quickly than the doctors predicted.

"Every time he loses an ability to do something, it's heartbreaking," she said. "We try not to cry in front of him. ... We try to keep the laughter going."
Bully can no longer walk, swallow or speak clearly. He can't write messages like he did when he first lost his ability to talk. Nohea can understand some of what he says; what he can't say, she tries to guess, and he'll nod or shake his head in response. If someone is watching, he can tap a finger on his leg to indicate he needs something; at night, if he needs to use the bathroom, he'll pull a tube forcing oxygen in and out of his lungs free of his nose, which creates a hissing noise that catches Nohea's attention.

Through each of those losses, Bully remains positive, Nohea said. Married for 22 years, the couple, both 44, first lived on Oahu, then moved to California about seven years ago. They wanted to get ahead financially, do some traveling, then move back to Hawaii, back to family. Bully worked as a security manager for a resort in Monterey Bay, and Nohea was director of paralegal services for a law firm.

Nohea was a workaholic, she said. Bully used to tease her, tell her she needed to learn to have more fun. She used to scold him for playing too much golf. After he was diagnosed with ALS, she changed her mind, and encouraged him to golf as much as possible. They, with their children, Mahea, 21, and Max, 19, took their first family vacation back to Hawaii in 2006.

A shift in priorities meant the diagnosis brought one positive thing -- more time spent together as a family. Mahea was living on Kauai when her dad was diagnosed, Max was just out of high school. They put off college to spend time at home. Nohea started working from home a few days a week.

In July 2007, they moved to Waikoloa, a decision based on Nohea's ability to work as a paralegal with a law firm in Kailua-Kona and the proximity to physicians in Waimea and at North Hawaii Community Hospital. The move didn't get off to a great start. The area's neurologist had just left. The pulmonary specialist wasn't taking new patients, and they couldn't find a primary doctor right away, either.

A gastroenterologist, the fourth doctor Bully needed as part of a team of physicians to treat ALS, was available and accepted Bully as a patient.

The lack of formal support for people with ALS and their families also was a frustration, Nohea said. A few months after the move, she contacted the Muscular Dystrophy Association, which helped organize the first Big Island ALS support group late last year.

They've talked about the future, about a time when Nohea and Mahea may no longer be able to care for Bully by themselves. But the thought of bringing a stranger into their home is a hard one to discuss, Nohea said. And they know the average life expectancy after diagnosis is two to five years. Just more than two years have passed since the first symptoms appeared.

They've already decided to do everything at home, right up until the end.

Friends recently started telling Nohea to take time for herself, maybe go for a massage.

"You don't think about yourself," she said. "Where do I find the time to do that?"

Nohea wakes up most mornings between 4 and 4:30 a.m. She drives from their Waikoloa home to Kailua-Kona, where she begins work by 5:30 a.m. Her workday ends between 1:30 and 2 p.m., and she immediately begins the drive back north. She doesn't have time to go to the bank or Costco or even the pharmacy; her daughter needs to leave for work as soon as Nohea arrives. Nohea's son works a security schedule that varies from day to day, so she doesn't count on advanced notice from him that he could stay with his dad while she runs errands.

"This disease has affected us on every level," Nohea said. "It's demoralizing. You get mad. You try to have faith."

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