

SANTA BARBARA NEWS-PRESS

Rallying around Gwendolyn - Community signs petition to help S.B. infant's fight against muscular disease

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One in every 40 adults carries the recessive gene responsible for spinal muscular atrophy, or SMA.

If two recessive carriers have children, there is a one-in-four chance that their child will have SMA.

Bill and Victoria Strong are one such couple.

In late August, they took a road trip to Stanford University. They packed their then 10-month-old daughter Gwendolyn's breathing machine, her suction machine that helps her swallow, her feeding machine, her coughing machine and her PULSOX, a wristwatch-sized device that monitors heart rate and oxygen levels in the blood, and set off in a borrowed RV.

The Santa Barbara family has been making trips to Northern California ever since their daughter was diagnosed with type 1 spinal muscular atrophy.

The goal of the first trip was to implant a feeding tube in Gwendolyn's stomach. SMA destroys voluntary muscle movement: No longer able to eat on her own, Gwendolyn would be sustained directly through a tube.

"They regress pretty quickly," Mr. Strong, 32, said, referring to his daughter and other children with SMA.

Earlier in the year, he and his wife, 31, a former English teacher, developed a petition in response to their daughter's illness. It is now signed by more than 17,000 people in favor of the SMA Treatment Acceleration Act, which they are lobbying to get on the floor of Congress before the election and ultimately passed. Once passed, the act will provide more funding for the disease and federal resources to organize research and clinical trials.

"Our original goal was 1,000 signatures. We got that in four days. Then it was 10,000, which we got in three weeks. Now our goal is 50,000," said Mr. Strong.

More than just a cry of frustration, the petition has become an effective political tool.

"We'll go into Washington," said Martha Slay, president of an international nonprofit organization called Fight SMA, "into an office that hasn't signed, and say here is a list of your constituents. And in some instances a congressman or senator would be really moved to either sign or take this seriously."

"These are people interested in numbers," she added. "They really are."

Mr. Strong, who works in corporate development, attributes the success of their petition to both his business background and the intense frustration of an SMA community that crosses lines of race and gender.

A few of the statistics that Ms. Slay regularly uses in presentations in Washington, D.C., are:

- No genetic disease kills more babies than SMA.
- SMA is the No. 1 inherited cause of death of children under age 2.

When Gwendolyn Strong took her road trip to Northern California, she did more than just receive medical attention. She also visited the Avenue of the Giants, a scenic drive famous for its redwoods, and the Monterey Bay Aquarium, where she fell in love with the jellyfish.

There was a swimming pool at one of the RV parks in which Gwendolyn swam as well -- Gwendolyn, with her frail body, loves to swim.

"She was born perfectly healthy in October at (Santa Barbara) Cottage Hospital," said her father. "She was diagnosed in April with SMA. Immediately we were told by local doctors that there's nothing we could do."

The fundamental deficiency that causes the disease is a missing protein, appropriately titled the survival of motor neuron protein. Without it, Gwendolyn's motor neurons die.

This deficiency is not addressed by the piles of equipment that force Mr. Strong to borrow an RV from his brother-in-law every time they leave Santa Barbara.

"We call it the symptomatic treatment, meaning you treat the symptoms," said Dr. Ching Wang, a leading SMA researcher at Stanford University, and Gwendolyn's primary neurologist.

"You help them to breathe by giving them some respiratory support. They don't eat -- they cannot eat because they choke. We give them nutrition through a gastronomy tube. They don't walk, so a lot of these kids are in wheelchairs, if they survive longer."

It is an RV full of equipment meant to keep Gwendolyn alive but not well.

In the average type 1 case, time passes and more motor neurons die. The child becomes increasingly frail until pneumonia or another respiratory disease claims his or her life before the age of 2.

While the Strongs are overjoyed with the quality of care they have received at Santa Barbara Cottage Hospital and from local doctors, it is not enough.

"Our goal," said Dr. Wang, "is to hopefully stop the disease progress, or to eventually be able to reverse the disease process. In other words, to stop the motor neurons from dying."

It is the goal of every SMA researcher and, according to Mr. Strong, there are glimmers of hope in his daughter's condition.

They enrolled Gwendolyn in a clinical trial at Stanford under Dr. Wang's watch that aims to administer a drug that may increase the amount of SMN protein in Gwendolyn's body.

Hope can be a dangerous thing when it comes to a disease like spinal muscular atrophy. The changes the Strongs are noticing are not huge. They are slight, subtle changes.

But the Strongs know their daughter, and she is changing.

Gwendolyn has demanded so much of their attention that friends organized a meal-delivery service for them, despite the fact that both of them were taking time off from work, and a constant night nurse was easing their time burden, giving them mostly full nights to sleep. (Mrs. Strong continues to wake up at 2 a.m. and administer a respiratory treatment.)

"I'd say that any parent is in tune with things that their baby is doing, but I think that Victoria and I, people who, on a 24/7 basis, are watching like a hawk, we're so in tune that when we're holding her up, we can just tell that her neck movements (have changed) -- that she can actively move her neck around," said Mr. Strong.

"When she's playing around with fingers," he added, "her grasp seems a lot tighter than it used to be."

Although this is just one incomplete clinical trial, and no conclusions should be drawn, researchers such as Dr. Wang are hoping that in this and similar trials they can detect a new trend out of the relentless statistics of spinal muscular atrophy.

One last statistic that Ms. Slay would like to make known on Capitol Hill is this:

Out of more than 600 neurological diseases and disorders, the National Institutes of Health and the National Institute of Neurological Disorders and Stroke chose SMA as the closest to treatment.

YOU CAN HELP

To read more about Gwendolyn Strong or sign Bill and Victoria Strong's petition, go to www.gwendolynstrong.com

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