

Hoping for a future of memories

Rare disorder affects Avon sisters' ability to remember



From left: Amber LeVine, 22; Kathy LeVine; Haley Levine, 20; and Jess Levine. THOMAS P. COSTELLO/STAFF PHOTOGRAPHER

By Ken Serrano :: Staff Writer

It took seven years and five hospitals before Jess and Karen LeVine discovered why the two youngest of their four daughters suffered from severe memory problems and delayed development.

At one point in their quest to attach a name to Haley and Amber LeVine's condition, a doctor who had tested them for a year and a half told the girls' parents it was caused by a disease that attacked the body's organs. The doctor told them the disease was fatal — and then promptly went on vacation, the LeVines said.

That, of course, rattled them both. But ultimately, it amounted to just a temporary setback.

In 1996, they traveled to Johns Hopkins Hospital in Baltimore for more answers. And Jess and Karen LeVine were told by one neurologist that there is no diagnosis for about 30 percent of the patients with disorders like that suffered by their children.

"I started crying," Karen LeVine said. "I said, 'There's got to be an answer

here.'"

They were directed to two geneticists at the hospital. Coincidentally, one had just returned from a conference at which succinic semialdehyde dehydrogenase deficiency, or SSADH, was discussed.

It is a congenital metabolic disorder, a disease involving the degradation of neurotransmitters. It is not fatal.

The testing the children underwent led to the diagnosis of SSADH, something fewer than 400 people in the world

IF YOU GO

The 2011 Wave of Hope Holiday Benefit is from 7 to 11 p.m. Thursday at The Headliner on Route 35 in Neptune.

Live music includes the Pat Roddy Band and New Jersey and Florida Keys musician Paul Cilinski. Tickets cost \$25. For more information, call 732-778-8930.

Succinic semialdehyde dehydrogenase deficiency, or SSADH, is a congenital metabolic disorder, a disease involving the degradation of neurotransmitters. Fewer than 400 people in the world have been diagnosed with the disorder.

have been diagnosed with, according to the Pediatric Neurotransmitter Disease Association. That umbrella nonprofit group provides grants to researchers and advocates for children and families who are affected by any of several rare diseases.

"We just about fell off our chairs," said Jess LeVine, a history professor at Brookdale Community College in the Lineroft section of Middletown and a

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veteran lifeguard in Avon, where the family lives.

They had an answer, but that was only the beginning of their struggle.

Since then, the LeVines have been anxiously awaiting a drug trial that might help their two daughters. Amber is now 22 and Haley is 20.

And they are now turning to their community for help.

They are holding a benefit at The Headliner on Route 35 in Neptune on Thursday. The money will go to the PNDA.

The drug they are pinning their hopes on, SGS742, improves the thinking of patients with mild cognitive impairment, according to one of the pharmaceutical companies that helped develop the drug.

It originally was designed to help patients with Alzheimer's disease and is now being aimed at SSADH sufferers.

The two young women underwent a battery of MRIs and other tests a few years ago in prep-

aration for the study. But regulatory approvals are pending.

"It's not all signed off on yet. It's going to take time to get there," said Dr. K. Michael Gibson, a professor of biological sciences at Michigan Technological University who has spent the last 30 years researching what is now known as SSADH.

"I would hope that it would be started within the year, but I don't know," he said. "It will happen. It's just going to take time."

The drug already has been successfully tested for safety on human subjects.

How well it works to improve mild cognitive impairment is what the drug trial will determine.

He declined to discuss just what he is hoping the drug can do for people like Amber and Haley, other than to say the trial will examine any clinical improvements and that neurobehavioral tests would be part of it.

Jess LeVine's colleague Jim Crowder, a biology professor at Brookdale, explained that SSADH patients get too much

gamma-hydroxybutyric acid, or GHB.

GHB is a naturally occurring substance found in the central nervous system that activates receptors in the brain that interact with neurotransmitters. Neurotransmitters are chemicals that relay information throughout the brain and body.

Large amounts of the substance interfere with the development of the nervous system and lessen the brain's ability to concentrate, process information and use muscles. Fine motor skills and speech are affected, Crowder said.

The first sign of trouble emerged when Amber was 2½. She dragged one of her feet and showed delayed development, Karen LeVine said. Haley was 3 months old at the time.

Now, Haley, 20, and Amber, 22, still require an enormous amount of energy and devotion to help them with daily activities.

"They never grew up, to a large degree," Jess said. "It's still as if they are young children in some ways."

The young women can converse on a simple level and perform daily tasks. Amber even audits classes at Brookdale. Her father hopes the courses, really just a formative experience for her, will serve as a benchmark for cognitive performance when the drug becomes available.

Both women study music — Haley guitar and Amber piano — at Beach Music Studios in Belmar, where they learn basic songs over time.

Both are lively and spirited and act in many ways like any other pair of siblings.

"Don't be rude, Amber," Haley said, scolding her sister for some apparent slip.

Haley attends Hawkswood School, formerly School for Children in Eatontown. It serves children with autism, multiple disabilities and preschool disabilities, according to the state Department of Education.

She cannot read and struggles more than her sister with speech and normal daily activities. Amber's reading level is a source of debate between her parents.

Jess believes she is at a second-grade level, but Karen says she knows only a few common words.

Amber works at the Avon Pavilion snack bar in the summer, stocking shelves, handing out food and sweeping outside, she said. She also holds down a part-time job at TGI Friday's in Eatontown.

"I like it. Nice people," Amber said, struggling to respond and being prodded by her mother.

Memory problems are what the LeVines hope SGS742 can correct. They cannot process time, something the drug might change.

"They could get a sense, a grasp of time," Jess LeVine said, possibly allowing them to learn how to read and perform math problems.

Having two daughters who need assistance with most daily activities has been a strain on both parents.

"When you come home and you're tired, you have to dig deep," he said. "Haley ... might ask me 10 times when a hockey game she is going to is. That can drain you."

The couple's two older

daughters — Jaqui, 29, and Ali, 26 — help take care of the daughters.

There have been family milestones along the way.

When Haley and Amber were 7 and 9, Karen LeVine taught them to ride bicycles. It took six months of 2-foot increments. Karen made it a contest. After six months, they were able to pedal around the block, she said.

"Now we all go out once a week and take a bike ride," she said.

"It's freedom for them," Jess said. "Like the guy who can't read, then can. All those things we take for granted."

The drug may provide some freedom for the parents, but the women most likely will always require supervision.

But the burden of raising awareness about something so rare may not go away any time soon, despite the couple's persistence.

"I'm banging on the door of the 'Today' show, NBC Nightly News," Jess said.

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