

MLD

Sisters get special moment before seclusion



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OREM -- Victoria, Madelena and Emma Campbell donned new formal dresses, did up their long hair and put on a touch of makeup in preparation for a dance on Friday night. They danced the night away, laughing and celebrating with friends and family who were similarly attired. They swapped smiles, hugs and dances in the packed church gymnasium as the Mountain View High School Jazz Band played favorite songs.

The three girls, ages 16, 14 and 12, were the guests of honor at the dance, a sort of prom for Victoria and Madelena. It was the last time in months the girls would be able to party, see friends or even leave the house, as both are preparing to enter treatment for metachromatic leukodystrophy, a rare and potentially fatal genetic disease.

"I'm so excited, beyond happy. I'm excited everyone came for me," Victoria said. "I want to have a good attitude. I feel like if I don't go in with a good attitude the experience will be wasted. I want to be the girl that gives everybody hope."

What is it?

Aaron and Emily Campbell had no idea when they got married that both were carriers of MLD. There is about a one-in-40,000 chance of that. In fact, the genetic strain can jump several generations.

"It was a fluke in the genetic lottery and we won," Emily Campbell said. "We've really struggled with what will happen."

Metachromatic leukodystrophy causes the degeneration of the white matter of the brain and central nervous system. MLD affects the nerves so signals can't get through between hemispheres of the brain and from the brain to the body. She added that typically an MLD patient has between two and seven years life expectancy as their world digresses. People first start acting and feeling younger than they are. Ultimately they are confined to a wheelchair, dementia sets in and they lose their ability to control body functions like swallowing.

There is no cure for MLD. The only treatment is a bone marrow transplant or stem cell transplant that slows the disease's effects, or in some cases, halts it.

"There is a significant mortality rate," Emily Campbell said. "But we have to get it done."

What's next

The girls' best chance for survival most likely will come through little sister Emma and her matching bone marrow. Victoria and Madelena will go through radical chemotherapy treatments, which will kill their bone marrow. It will then be replaced with Emma's transplanted bone marrow, assuming she is not a carrier for the disease.

"Emma is a bone marrow match," Emily Campbell said. "We're waiting to hear if she is free of the disease. If the genetic tests come in positive, the girls will go in together. If not, they will be staggered by a couple of weeks."

While the process for retrieving the bone marrow is much less invasive than it used to be, it still is not pleasant. Emma will be in the hospital at least overnight as she goes through a dialysis process. The 12-year-old says she would kind of like some of the attention.

"I'm not mad about it 'cause I get it," Emma said.

Victoria is starting the process off. Within the next couple of weeks she will go through a battery of tests on all of her organs to allow doctors to develop baselines. Then Madelena goes in. Both then will go through seven to 10 days of radical chemotherapy. This also will destroy the girls' immune systems, meaning any exposure to germs could prove deadly. They'll be stuck inside, communicating electronically and trying not to get sick.

"Then we start with day zero when the chemo is done and they get the transplant," Emily Campbell said.

The girls will spend between 20 and 40 days in Primary Children's Medical Center for the marrow to engraft and start producing clean blood and marrow. The old will be completely wiped out. The girls also will need to be on immunosuppressants for months. They'll need multiple blood transfusions. The girls will be very weak. After that it's two months or more of seclusion. It will be nearly a year from start to finish.

One of the biggest concerns from doctors is that the most intense part of the procedure will happen during the height of the RSV season.

During their seclusion, which will be in their grandmother's basement apartment close to the hospital, the girls will not be able to see their friends and extended family members. They won't be going to girl's camp, school dances or parties with friends. Their mother will live with them, but she will always be masked. Their grandmother, who is a nurse, will stay close by. If the girls get the slightest temperature, they must be at the hospital within an hour.

Aaron Campbell will stay with the rest of the family in Orem and they will visit by phone and Skype. The girls' environment must be kept clean and as germ free as possible -- not completely enclosed in a bubble but as close as possible. Extended family will have to communicate through electronic means or letters.

How did they know?

Just a short time ago, Victoria Campbell was an extroverted teenager who enjoyed traveling and learning. She was a year ahead in school and the family had been living in South America

for two years. That's where the signs first appeared.

After returning to Utah, Emily and Aaron Campbell noticed Victoria was showing signs of digression in her muscle control and motor skills. For instance her walking gait had changed. It started as little things. She also had compulsive behavior problems. At first her parents thought she was just being a goofy teenager. People with MLD revert to their younger baseline personality. Her mother said Victoria is kind of going backward now.

"Victoria is vivacious, fun and optimistic all the time," Emily Campbell observed. "It's kind of a mercy. It's amazing this isn't depressing her."

The Campbells waited months just to be seen, then they waited even longer for a diagnosis from specialists in Utah County and at Primary Children's Medical Center. A neurologist discovered the white matter disorder. They went back to Primary Children's, where they met with the foremost expert on MLD west of the Mississippi. The MLD diagnosis came in early December, along with the news that the condition was genetic.

Frightened, Emily Campbell watched her other children closely and noticed that Madelena was becoming more subdued, acting younger and more innocent. She had become more childlike. Her changes were less physical and more psychological. She was tested two weeks before Christmas.

What scared her parents excited the teenage girl, however. Emily Campbell recalled how Madelena came rushing out of the doctor's office after her diagnosis. She ran right to Victoria and gave her a huge hug.

"I was just happy Victoria didn't have to go through it alone," Madelena said. "Both of us are growing younger. When she goes through the tough times I can make her laugh. I didn't want Tori to go through this alone."

Since then all of the children have been tested. Emma is waiting to hear if she can donate bone marrow. A younger brother, Eli, 8, may be a potential donor. Ike, 7, also was tested.

Despite all the needles, testing, fear and uncertainty that fill their days, the family is staying positive. Even Victoria and Madelena, facing pain, treatment and isolation, have managed to stay happy -- most of the time.

"I started bawling when I heard I was going to lose my hair," Victoria said.

Hair loss is one of the side effects of chemotherapy, meaning both girls will lose their long, light brown hair. They're sad, they admitted, and scared. It's one more thing their disease is taking away from them, one they'll have to face every time they look in the mirror.

So the Campbell sisters decided to outwit the chemotherapy. Since they weren't going to keep the hair, they decided to give it to somebody else. Before they go into Primary Children's, Victoria and Madelena will shave their hair and donate it to Locks of Love. Additionally, Aveda Institute in Provo has chosen Jan. 18 as a day to honor the girls. Anyone donating hair to Locks of Love that day can have it donated in Victoria's and Madelena's name.

Support back home

Fellow students at Mountain View High School found out about Victoria's diagnosis just before leaving for Christmas break. Vice principal Peter Glahn said the teenagers contacted Aaron Campbell and asked how to remain connected with Victoria.

"Kids just volunteered," Glahn said. "She knows a lot of students and what's cool is there has been more positiveness around this. Students are helping her to not be alienated from them socially. It's a team effort."

Technology provided ways for the girls to not only keep in touch with friends but also keep up on their schoolwork. Victoria and Madelena each have a tablet computer for studies and to communicate with friends. They'll have a tutor and will have school via the Internet. Their peers plan to write letters, blog, chat and Skype every day to keep the girls up-to-date on what's happening on the social scene. Plus, Mountain View students are working on a life-size cutout of Victoria to take to all the school events. There will be pictures, they've said.

"I've got good friends and lots of family," Victoria said. "There's always hope. Surprisingly, I have no fears. Everything will work out the way it's supposed to."

Glahn went on to say the public often undervalues the social aspect of high school. "Tori is in Bruin Crew, a service group for people in need. It's cool all these people are coming together to help her."

Of course, a digital presence or a cardboard presence doesn't completely make up for hanging out with friends on Friday night. Madelena says she'll miss the day-to-day connection with friends. She said when she's depressed she goes shopping with her friends and they talk about important things and have fun eating.

"I want my friends to send me messages. I'll write poems about being in the hospital," Madelena said.

The girls believe they will be just fine as long as their friends are supporting them through the long road to recovery.

Emily Campbell said that although she doesn't like it she believes it will all work out. "We believe we will be a family forever."

"I'm pretty much ready," Victoria said. "Bring it on."