Love saves daughter's life: a Cleveland Clinic liver transplant story (The Plain Dealer)

SHAKER HEIGHTS, Ohio -- There's an old adage that emerged thousands of years ago: "Greater love has no one than this, than to lay down one's life for his friends."

This ancient proverb is still exercised today. Every year in the U.S., nearly 200 reports surface about courageous individuals who risk their lives in a rare surgical procedure known as a living-donor liver transplant. Such operations consist of removing a piece of the liver from the donor, so it can be transferred to the recipient. This procedure differs from a cadaver-donor liver transplant, in which the donor is deceased.

The first successful living-donor liver transplant happened at the University of Chicago Medical Center on Nov. 27, 1989. At Cleveland Clinic, such a procedure was performed on March 17, the eighth living-donor transplant this year and the 75th at the health system since 1999. The transplant took place between 39-year-old Shelley Hunt and her 22-month-old daughter Kenace Hunt, both of Shaker Heights.

Shelley was the donor. Kenace, the recipient.

On Friday, May 16, Kenace turned two years old, and her mother says she's more active than ever. "Now, she takes more risks than she did before. Before the surgery, we would look at other kids her age, and they far advanced her physically," Shelley explains. "Now, we're seeing her catch up. She's like a normal two-year-old."

Although, for more than 20 months prior to her second birthday, Kenace's body gradually spiraled downward as a result of a dysfunctional liver. But evidence shows that love is what saved her life.

It's common for newborns to enter the world with high levels of bilirubin in their blood. Many times it's an easy fix. With the use of phototherapy, bilirubin levels are quickly lowered.

At first, that's how it all appeared for Kenace two years ago. Just like other newborns, her bilirubin levels were high. Yet, not high enough for doctors to consider phototherapy. But for six weeks, Kenace's levels never dropped. That's when Shelley and her husband, Sean, realized something wasn't right.

"Hers just kind of remained steady," said Shelley. "We knew that something was really wrong, because we were watching it closely."
Soon after, Kenace was referred to a gastrointestinal specialist. She was given a series of tests. The child also showed low coagulation levels - or blood clotting levels. For about six months, uncertainty lingered about the cause of Kenace's health problems. Then there came the day Shelley picked Kenace up from daycare. She noticed visible changes on the infant's body.

"She had a bruise on the forehead and on the following day she had more bruising on the torso. We immediately took her to the pediatrician," said Shelley.

The pediatrician diagnosed Kenace with an ear infection and ordered lab work for the six-month-old. Soon afterward, Kenace started vomiting and Sean and Shelley took her to the hospital. There, it was discovered Kenace had bleeding on the brain as a result of her body's failure to coagulate her blood.

A new complication arose: Her body was unable to absorb Vitamin K, a nutrient responsible for coagulation. Likewise, her body failed to soak up just about all fat-soluble vitamins.

A gastrointestinal specialist realized it was crucial to get to the bottom of Kenace's problems. More aggressive tests were ordered, including genetic evaluations. Nearly three months later, the specialist reached a diagnosis: Kenace had a rare disorder known as **Progressive Familial Intrahepatic Cholestasis, or PFIC**.

She inherited the disease from Sean and Shelley, who were carriers of a gene mutation that prevented Kenace's bile ducts from forming normally. Her liver lacked the ability to release bile - a greenish-fluid that helps with digestion. Kenace's bile built up in her liver, causing the organ to scar severely.

PFIC affects about 1 in 50,000 people worldwide. It's prognosis: liver failure and eventually death.

In addition to Kenace's inability to absorb fat-soluble vitamins, she suffered from other signs of illness: severe itching and the failure to thrive. To help her daughter gain weight, Shelley began working with a dietician. Kenace eventually gained weight.

However, finding a solution to the severe itching seemed impossible.

"She had scars all over her body from constantly scratching," Shelley explained. "Kenace had to be fully covered or she would scratch if her skin was exposed. She didn't get sleep at night, which means we didn't get any sleep."
For several months, the Hunt family tried different treatments, to no avail. After speaking with doctors, there was only one choice left.

"As soon as they said she needed a liver, I said, 'I have to do this,'" said Shelley. "This was one of those situations where you don't think about it. My initial thought was, I have to try and give my daughter life."

Though the Hunts came to accept that their young daughter needed a new liver, they didn't want her on the cadaver-donor wait list. Instead, they chose the living-donor option, a risky procedure that's very costly.

"Kenace would have been on that list a long time had I not donated my liver," said Shelley. "She would have been on that list a really long time and eventually died."

According to the American Liver Foundation, in the U.S., there are more people in need of a transplant than there are donated livers. About 16,000 Americans are on the cadaver-donor list waiting for a liver transplant. Health systems use what's called a Model for End-Stage Liver Disease - or MELD - score to determine which patients get priority.

The higher the score, the greater chance of being chosen. The lower, the longer the wait. About 1,700 people die each year before receiving a transplant.

As desperately as Kenace needed a new liver, her MELD score was just too low.

Sean and Shelley were both tested. Eventually, Shelley was the best candidate for evaluation. A few months later, she found out she was a perfect match.

Though a liver transplant looked promising, Shelley spent months dealing with insurance companies which tried to convince the Hunts to add Kenace to the cadaver-donor wait list. Some told her that based on her insurance coverage, the living-donor liver transplant could only be performed in certain hospitals located out-of-state. In the eyes of Shelley's insurance companies, having the surgery done at Cleveland Clinic was not an option.

But Shelley was adamant. "I said no, my team is at Cleveland Clinic, I don't think you understand. You've got top of the line surgeons right up the street," she said. "The insurance companies want you to do everything but the most expensive thing."
Expensive is right - a living-donor liver transplant can accrue costs up to $580,000. "It's expensive, but it's not like it can't be done," said Shelley.

From November of 2013 to January of 2014, Shelley spent hours on the phone, arguing with insurance companies. "It took me screaming and yelling everyday, calling them everyday," said Shelley. "But my thing was, 'look you don't seem to understand. My daughter needs a liver. It doesn't matter who I talk to, she's going to need a liver and I don't understand why you guys are dragging your feet.'"

Finally, by February 2014, the transplant was signed and approved. On March 17, Shelley and Kenace underwent their long-awaited procedure at Cleveland Clinic Main Campus.

Kenace's entire liver was removed, then replaced with a small piece of Shelley's liver.

Dr. Koji Hashimoto, transplant surgeon at Cleveland Clinic, explained that when it comes to a delicate procedure like a living donor liver transplant, size matters.

"You have to have a good size match. The mother donated 20 percent of her liver, and that was just the right fit for the baby," he said. "If you give too much of a liver, it's not going to work. If it's too small, it won't work either."

The liver is remarkably unique. It's the only internal organ able to regenerate itself. Shelley's liver is expected to automatically regenerate within six to eight weeks.

Many risks are involved for both the donor and recipient. According to Hashimoto, one donor death occurred years ago in the U.S. In another instance, the donor had liver failure and needed a transplant.

"There is the risk of death for the donor, but it's at 0.2 percent," he explained. "In 2001, living donor transplants reached 500 per year in this country, but after the donor death, the number declined." He hopes to see more living-donor liver transplants happen in 2014.

Shelley and Kenace are recovering at home in Shaker Heights. Kenace stays busy and is curious about her surroundings. Before the transplant, walking was painful. Now she runs.

Twice a week, Kenace gets her blood drawn. Doctor visits are once a week. Ultrasound visits once a month. She takes several medications. Kenace has to take anti-rejection medicine for the rest of her life.
There is an expensive hospital bill to pay. The family is working to raise money, but the challenge doesn't beat having a playful two-year-old around.

Shelley said she would do this all over again for a loved one. To those considering becoming living donors, she says, "The people who get high up on the cadaver-donor wait list are on their deathbed. Why wait until someone is too weak to even go through the process? When your liver is not working, so many other things go wrong."

About Kenace, Shelley says, "She sleeps through the night, which is good for the whole family.  "I'm glad I had a good recovery because I have to run around chasing her."