Liver Transplant Saves 10-Day-Old Baby Boy

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ST. LOUIS -

Keeley Gibbs knew it was risky to get pregnant. Doctors warned that she and her son born in January could have died. The young woman from Eldorado, Ill., like generations of her family's females, carries the gene for a rare metabolic disorder of the liver.

OTC-deficiency is fatal in males, and in utero tests diagnosed the fetus with it. Gibbs wouldn't consider abortion. She survived the pregnancy and birth, and her son, Jacob, received a lifesaving liver transplant at St. Louis Children's Hospital at the youngest of ages, 10 days. Officials say he is perhaps the youngest child ever to receive a liver transplant in the United States. Three months later, the child is doing well.

"Everything happens for a reason," Brian Gibbs said, cradling his tiny son in his lap during a medical checkup at Children's Hospital on Wednesday. Then, looking into Jacob's blue eyes, Gibbs added,

"Ain't that right?"

Ornithine transcarbamylase -- or OTC deficiency -- is the absence or shortage of a critical liver enzyme that helps rid the body of ammonia - a toxic breakdown product of protein. When ammonia accumulates in the blood and travels to the brain, coma, brain damage and death result. Women typically are carriers; their sons with the gene suffer the disease. Peggy Sauls can testify to the disorder's devastation. Sauls, Keeley Gibbs' maternal grandmother, and Jacob's great-grandmother, lost an infant son with the disorder 46 years ago.

"My baby boy at two days old went into seizures and tremors as the ammonia went to his brain," she said. "That was in 1960. We didn't know at the time what he died from." Sauls, her daughter, and Keeley and her sister are carriers. So when in utero tests during Keeley Gibbs' pregnancy revealed high levels of ammonia, intensive preparations were made for the birth and round-the-clock monitoring of Jacob at Children's intensive care unit. His survival depended on a liver transplant.

Jacob, born Jan. 10 at Barnes-Jewish Hospital, was put on the transplant waiting list at 7 days. When a donated liver became available three days later, Dr. Jeffrey Lowell, chief of pediatric transplant surgery at Children's Hospital, performed the surgery on Jan. 20. St. Louis Children's Hospital is one of the busiest pediatric organ centers in the country, with 18 pediatric liver transplants last year alone. But Jacob was so tiny and young, the surgery posed extra challenges. The donated liver from an older child was far larger than what would fit easily in Jacob's tiny cavity. Lowell substantially reduced the size of the organ but it was still too large. Until Jacob's belly could
accommodate the liver, which shrinks with time, a medical-grade rubber patch was placed over the surgical site. The patch was surgically downsized every couple of days for two weeks until the belly could safely be sewn. Forcing the closure would have led to serious problems.

Jacob’s new liver will grow with him."The liver is smart," Lowell said. "It’s the only organ that regenerates."

During Jacob’s checkup Wednesday, Children’s Hospital medical staff marveled at how well he is recovering. Dr. Ross Shepherd said Jacob has avoided complications that often accompany organ transplants.

"The belly looks fine. The scar is fantastic," he said. Next procedure? Jacob's circumcision. Keeley Gibbs was a little worried.

"We don't want him to hurt," she said.