

New Cook Children's Hyperinsulinism Center becomes destination spot for children with genetic condition

A quality life

It's easy for his mommy to laugh today as spunky little Ty Hammons commands attention in the way most 3 ½ year olds do for parents. But there was a time, when Denise Hammons faced the possibilities of a world without him.

The odds were against Ty from the moment he was born. He was given a 50/50 chance of surviving at birth. He beat them only to end up at Cook Children's at 4 months old with a seizure.

During this stay, Ty was diagnosed with hyperinsulinism (HI), a rare and severe disorder that frequently causes persistent hypoglycemia in newborns and children. He was then sent to Children's Hospital of Philadelphia (CHOP) where he received a 95 percent pancreatectomy. At the time, CHOP was the only pediatric facility that could determine the exact portion of the pancreas that needed to be removed.

Although Ty did receive some treatment at CHOP, he was mainly cared for at Cook Children's. He is currently on overnight feeds and Octreotide (a drug used to stop insulin secretion) and is doing well, with follow-up visits at Cook Children's every three to four months and a yearly more extensive checkup.

From here on out, local children and kids from throughout the South will no longer have to travel to Philadelphia for top-quality care of hyperinsulinism.

The Cook Children's Hyperinsulinism Center opened in October 2010 and is the second of its kind in the country and the only one in the South. It is the primary referral center for the south. Paul Thornton, M.D., is a renowned physician for patients with HI, helping to establish the program at CHOP before joining Cook Children's.

"While other major pediatric centers may have the ability to treat patients with hyperinsulinism, we go above and beyond by providing a dedicated multidisciplinary team to offer a huge amount of knowledge and support to families," Dr. Thornton said.

We see many patients a year, currently following 35 kids, and are better equipped to provide care because of our experience with hyperinsulinism."

Hypoglycemia, also known as low blood sugars, may be caused by a variety of medical conditions. Once a baby has been diagnosed with hypoglycemia, the next step is to find what caused the hypoglycemia and how to treat it. Congenital hyperinsulinism (CHI) is the most common cause of hypoglycemia in infants more than 3 days old.

CHI is a rare and serious disease that affects nearly one in every 50,000 babies born in the U.S. each year. The condition can cause severe hypoglycemia, which could lead to brain damage in 20 to 40 percent of the affected children.

There are many forms of CHI and the treatment of each form is different, making it crucial to diagnose the exact form of CHI.

Since the opening of the hyperinsulinism center, Cook Children's has consulted, evaluated and treated an extensive amount of children with hypoglycemia from unknown causes to recurrent hypoglycemia due to hyperinsulinism.

In the last few years, a new test has become available in Europe to diagnose and guide treatment. The test is called the 18FDOPA PET SCAN. In the USA, the 18FDOPA is still considered experimental and Cook Children's HI Center is in the process of applying to the FDA for permission to use this agent. At press time, more than 200 infants and children in the entire world have had this test and it has changed the way HI patients have been treated.

Dr. Thornton is a pioneer in HI research and established the first ever multidisciplinary team approach to the management of patients with HI in the United States. He has now set up a similar approach at Cook Children's and will continue to bring leading-edge research to our children.



TOP: Ty Hammons; BOTTOM: Dr. Paul Thornton examines Ty Hammons during one of his follow-up visits.

To contact the
Cook Children's
Hyperinsulinism Center,
call 682-885-7960.

A normal life

It's a Monday night in small town, Colorado City, Texas. Everything seems normal for single mother, Kristian Morris, except she has to work a little later than usual, helping out with a middle school basketball game. The next morning she wakes her 3-year old daughter, Caroline, for the day, and then heads off to the local elementary school where she is a family service worker for the Head Start program.

What a difference time can make.

Three years earlier, on the night before Caroline's birth, Kristian slept in her car, due to an ice storm. She was on shaky ground with her family and had been living in a homeless shelter. She only wanted to make sure that she and her unborn child would have a healthy delivery at a good hospital.

The next day, Jan. 27, 2009, Kristian gave birth to a beautiful baby girl, three weeks earlier than expected, at Medical Center of Lewisville. Morris was shocked to learn that her newborn weighed in at 11 pounds, 3 ounces.

The physicians who delivered Caroline knew that something was wrong because of her labored breathing that was attributed to her patent ductus arteriosus (PDA), and the extreme fluctuations in her blood glucose levels. Her physician suggested she be transferred to a hospital in Dallas. The endocrinologist on staff at that hospital suggested that Caroline be sent to a specialist, Paul Thornton, M.D., at Cook Children's. At 7 days old, Caroline was transported to Cook Children's Medical Center, where she and her mother first met Dr. Thornton, who continues to oversee Caroline's treatment and progress.

"To this day when I see Cook Children's Teddy Bear Transport, I start crying," Morris said. "That day was very emotional for me, knowing something was desperately wrong, worrying that all would be ok for my daughter."

Dr. Paul Thornton is an endocrinologist who is internationally respected for his treatment of the rare condition known as hyperinsulinism, also referred to as (HI). Morris was impressed with how quickly Dr. Thornton diagnosed her daughter with this condition.

Genetic testing revealed that Caroline had autosomal dominant hyperinsulinism, which is usually milder than a recessive form of HI. Early identification and treatment of genetic forms of congenital hyperinsulinism is critical in preventing the 20 to 40 percent of brain damage that is seen in children with this condition.

Caroline was born with a pancreas where all beta cells are abnormal and secrete insulin. When medical therapy fails, surgery is usually required

to remove up to 98 percent of the pancreas. Most of these patients develop diabetes or persistent hypoglycemia and need to be managed by an endocrinologist long term.

James Miller, M.D., performed Caroline's surgery at Cook Children's to remove 98 percent of her pancreas when she was only 3 weeks old. Caroline also received a gastrostomy button (g-button), which was placed into her stomach for additional feedings and medications that she requires.

"It was extra overwhelming for me at the time with my living situation and my family situation," Morris said. "I would not have made it through if not for Dr. Thornton. He definitely went the extra mile to make sure I understood everything that was going on with my baby. He brought me a whole folder to read about hyperinsulinism. I had never even heard of it before Caroline was born. He told me not to go online because there was a lot of inaccurate information. He got me involved with a group to talk to other parents who had babies with hyperinsulinism. Dr. Thornton saved my child's life. I love him. He doesn't just pretend. He knows what he is doing and really cares. To this day, anytime I email him, he picks up the phone and calls me right back."

While Dr. Thornton and the endocrinology team at Cook Children's cared for Caroline, others at the medical center also helped Kristian get her life together. She credits a social worker, a Neonatal Intensive Care Unit nurse, and a Home Health representative, who helped her to find ways to get financial help from Medicaid for Caroline's long term medical care. Also, this has allowed Caroline to be cared for by two skilled nurses while Kristian has continued to pursue her education even further. It has also given her daily support, and simply a shoulder to cry on when she needed it.

"I remember when I got home, I cried because it looked like a hospital had exploded and was all over my living room," Morris said. "It's been an amazing journey. But when I look at Caroline, it's all worth it. Sure, there are days where I'm overwhelmed, but I rarely even think about it anymore. It's routine. Feeding pump at night. Two injections a day. Check sugar every three hours. But if I could offer anyone advice, it gets easier and it's manageable. It's almost second nature."

And most importantly, "it gets easier and your life becomes more normal."

"At 3 years old, Caroline amazes me at how much she already understands about what she can eat, and the importance of following our regimen. My biggest hurdle was the fact that she would not eat. Eating was the only way to keep the blood sugar up and not have the huge



TOP: Today, at 3 years old, Caroline is happy and energetic.
BOTTOM: Caroline in Cook Children's Pediatric Intensive Care Unit following her surgery is held by her mom, Kristian.

swings, and she would not eat. With help from ECI, the nurses, and lots of coaxing, she is doing much better with this."

Morris was told in the beginning that Caroline would be below her age group in learning and in her growth, and that she could experience seizures.

"If you sat and carried on a conversation with Caroline, you would realize she is above her age group in every way," Morris said. "She is such a bright and inquisitive child. Her smile and personality says it all."

In talking with her mom, it is clear that this has been a life changing experience for her. Morris has done an outstanding job with all of the obstacles and changes of HI and getting Caroline to where she is now. She is an outspoken and positive advocate for Cook's Children's and Dr. Paul Thornton. She and Caroline have also been instrumental in helping others understand this condition and have success with their own HI experience.

What a difference time can make.