

The Gift of Life

Children's dedicated transplant team gives patients with end-stage organ disease another chance at life.

Pediatric surgeon and division chief of Transplantation, Dr. Patrick Healey performs many of the liver transplants at Children's and also provides follow up care for his patients.



CLOCKWISE FROM TOP:
*Matt, Madeline and
Lorie Hunt*



WORRIED THAT THEIR 6-DAY-OLD DAUGHTER, MADELINE, SEEMED UNUSUALLY LETHARGIC, LORIE AND MATT HUNT VISITED THEIR PEDIATRICIAN IN PUYALLUP. BY EVENING, MADDY BECAME COMATOSE AND WAS AIRLIFTED TO CHILDREN'S WHERE DOCTORS DIAGNOSED HER WITH CITRULLINEMIA, A RARE INHERITED DISORDER IN WHICH HER LIVER LACKED AN ENZYME TO METABOLIZE SOME PROTEINS, WHICH RESULTS IN A TOXIC ACCUMULATION OF AMMONIA IN THE BLOOD.

Maddy's doctors and nurses at Children's stabilized her condition and taught her parents to control her symptoms through diet and medication. However, her parents would have to remain on guard against sudden rises in her ammonia level for the rest of her life — any spike could potentially result in brain damage and even death.

A new liver, which would produce the missing enzyme, would correct the problem. "It was a clear decision," her mom recalls.

Because organs are scarce, it was nearly six months before the Hunts received the call that a matching organ was available. They rushed to Children's, where Dr. Patrick Healey, division chief of Pediatric Transplantation, performed the 10-hour operation to give Maddy, then 10 months old, her new liver. Unfortunately, a clot developed in her portal vein, blocking the primary blood supply to the liver, and Maddy's new liver failed within five days.

Even though the most advanced care for liver failure is available at Children's, there was nothing that could be done. Without a second liver, Maddy would likely die within 48 hours. Her parents went public with their story to encourage others to consider organ donation.

At the same time, a different family faced an agonizing situation of their own — their teenage son was on life support with no chance of recovery. They decided to honor his life by choosing for him to be an organ donor. Nearly 40 hours after Maddy's first liver failed, the teen's

adult-sized organ was offered for transplant. Healey performed a split liver transplant — Maddy received the smaller left lobe of the liver, and the larger right lobe was transplanted into an adult.

"It was bittersweet to realize what another family had to lose in order for us to gain the world," recalls Lorie Hunt.

Saving lives

Children's highly regarded liver transplantation program was launched in 1990 and remains the only pediatric liver transplantation program in the Northwest. The survival rates of our patients are among the nation's best, and they spend less time on the organ waiting list than the national average, according to the Scientific Registry of Transplant Recipients.

Children's began performing split-liver transplants in the early 1990s to help address the national shortage of organs and shorten wait times. This advanced surgical option is usually only performed at centers with both adult and pediatric programs. However, our close relationship with the University of Washington Medical Center enables us to offer this service to patients of any age in our region.

Living-donor liver transplant, in which doctors remove a segment or lobe of a relative's liver and transplant it into a child, is another advanced surgical option performed at Children's.

Kelly Thurlow, RN, coordinates the care of children before and after transplant.



Becoming an Organ Donor

Nearly 90,000 people nationwide, including more than 1,200 of our Northwest neighbors, are waiting for life-saving organ transplants. You can document your personal decision to be an organ and tissue donor by:

1. Saying 'yes' to organ donation when renewing your driver's license
2. Registering online at www.livinglegacyregistry.org
3. Calling the Living Legacy Foundation toll-free at 1 (877) 275-5269

Wait ... and hurry up!

Though the wait can be long, things move fast once an organ becomes available. A transplant team has one hour to contact a family and determine if the donor organ is best for their waiting patient. If the family can't be contacted, this very important opportunity may be missed and the patient's life remains in the balance as they continue on the waiting list.

Maddy's mom remembers this time vividly. "We lived as if the call would come any minute. I never left the house without thinking, 'What if it's today?' When the liver arrived, it was a hallelujah moment. We were scared, but so full of hope."

Transplanting an organ is a painstaking process that involves many hours of preparation before the surgery, and patients typically stay in the hospital for several weeks after receiving their transplant.

"There's a lot of excitement and anxiety leading up to the transplant, but the real challenge begins as patients learn to live with their new organ," says Healey.

It's a lifelong process. Medications are taken to suppress the immune system so that it won't reject or attack the transplanted organ. Other medications prevent infections and the side effects of the anti-rejection medications. The comprehensive team at Children's teaches families how to follow complex regimens of care — which can include up to 60 pills a day — at home.

"We see our transplant kids quite a bit their first year after transplant," says Kelly Thurlow, RN, who coordinates the care of children before and after their transplant. "Once some of the anti-rejection and infection issues subside, we can lower the doses of medication these kids take and they can begin to live more normal lives."

Coordinating with a patient's primary care provider and providing follow-up care after a transplant are among the unique aspects of Children's program, Healey says. "When our kids finally leave us,



it's because they've made the transition to an adult provider. It's kind of like a graduation."

For Maddy's parents, this continuity of care makes an incredible difference. "Her team's expertise, the fact that they know her history, even the loving way they greet her, gives us a real sense of security and faith in Maddy's future," says Lorie Hunt.

New hope, new program

By the early 2000s, Children's had developed a multidisciplinary transplant team whose excellent patient outcomes and surgical and clinical expertise positioned it to expand into new frontiers. The decision was made to expand the liver, cardiac and kidney programs and to add the emerging field of small intestine transplant.

"A variety of factors — including trauma, infection and congenital defect — can cause permanent intestinal failure," says Healey. "Many of these kids have never been able to eat by mouth — they receive all their nutrition intravenously and miss out on all the physical, emotional and social nourishment that eating provides. Small intestine transplant offers these kids a chance to eat, and live, more normally."

To jump-start the program, Children's and the University of Washington recruited Dr. Jorge Reyes, one of the physician-scientists who helped pioneer this new field at the University of Pittsburgh's Thomas Starzl Transplant Center. Though the first small intestine transplant was performed in 1987, the advances in anti-rejection protocols and surgical and infection-control techniques have recently made this procedure more successful.

"Building an intestinal program from the ground up is an exciting prospect. Within the last 12 years, intestinal transplant has progressed from a completely experimental procedure to one that may replace

Children's and the University of Washington have developed the first small intestine transplant programs on the West Coast under the leadership of Dr. Jorge Reyes, (left) who helped pioneer this field.

long-term intravenous nutrition (TPN) as the preferred method for treating kids with irreversible intestinal failure," says Reyes. "A few years ago, our main challenge was controlling organ rejection. Today, we are exploring ways to decrease immunosuppressive drug therapy while still protecting against organ rejection. Reducing the side effects of anti-rejection therapy lets us offer children a better quality of life."

After joining Children's and the University of Washington Medical Center as leader of both transplant programs, Reyes recruited internationally renowned hepatologist Dr. Simon Horslen to help him build the program.

"In deciding to create a small bowel program, you really are accepting responsibility to run it on a national level," says Horslen. "There are only about five programs in the United States

doing a significant volume of small bowel transplants. We will be the first on the West Coast."

Dr. Horslen oversees Children's new Intestinal Care Center, which treats children from around the nation with complex digestive conditions. Children in the center participate in an innovative program designed to restore bowel function by dietary, medical and non-transplant surgical therapies. Patients and their families receive extensive counseling and education throughout the process.

"Our goal," says Dr. Horslen, "is to gradually wean our patients from TPN. Although TPN is a lifesaver, its long-term complications include both liver failure and life-threatening infections. When weaning isn't possible, we offer state-of-the-art transplantation. But we want to prevent that whenever we can."

Next great challenge

Having achieved significant improvements in survival after transplantation of all organs in children, the next challenge is to improve their quality of life. The initial research to reduce the use of immunosuppressant drugs has shown promising results, and many children have been able to stop medications altogether. This strategy essentially removes any potential toxicity that immunosuppressive drug therapy may cause. But there is more work to be done. Under the direction of Reyes, Children's transplant team will be leaders in finding ways to improve the lives of children who have had organ transplants here, and around the nation.

END



PICTURED LEFT:

Maddy Hunt is a social 3-year-old who started preschool this fall. "Her care team's expertise and even the loving way they greet her give us a real sense of security and faith in her future," says her mom, Lorie Hunt.



PICTURED RIGHT:

Dr. Simon Horslen leads Children's innovative, new Intestinal Care Center. The center aims to restore patients' bowel function, which enables them to live more healthy, normal lives.