

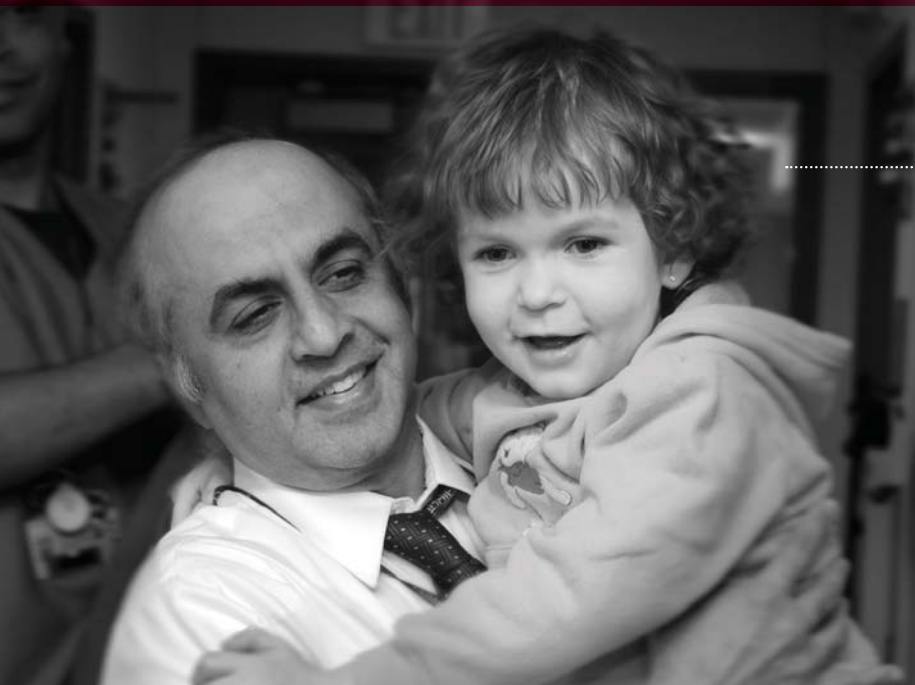
HILLMAN CENTER FOR PEDIATRIC TRANSPLANTATION

Pediatric Liver and Intestine Transplantation  
Referring Physician's Guide

UNMATCHED OUTCOMES.



UNCOMMON CARE.





“It takes a team to help a kid through a transplant. What they did for him is really like a miracle. Peter has a future now.”

*Catherine Herridge, mother of Peter.*

*In dual surgeries at Children’s Hospital of Pittsburgh of UPMC and the University of Pittsburgh Medical Center, Fox News correspondent Catherine Herridge underwent surgery June 6, 2006, to donate part of her liver to her infant son, Peter.*

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91%

... patient survival for liver transplant recipients in the past 10 years

## A WORD FROM THE DIRECTOR

Dear Colleague,

At Children's Hospital of Pittsburgh of UPMC's Hillman Center for Pediatric Transplantation, we have a rich history in the field of pediatric transplantation. From the early years when Thomas E. Starzl, MD, PhD, pioneered a new field of medicine, to recent progress in immunosuppressive therapies, these advancements have given hope to thousands of children.

Opening the nation's first comprehensive pediatric transplant center in 1981, Children's Hospital of Pittsburgh has continued to be a leader in improving solid organ transplantation. Over the years, we've made great strides in developing and improving surgical techniques. New strategies have offered children improved opportunities for long-term survival and a normal quality of life. This progressive approach has helped Children's achieve some of the nation's highest patient survival rates among pediatric transplant centers.

Our success is due to understanding the unique needs of our young transplant patients. At Children's Hospital of Pittsburgh, we provide a comprehensive approach to treatment, which includes addressing the medical, emotional and social needs of our patients and their families. For our patients, transplantation is a lifelong journey, and we have built a program that is committed to providing ongoing optimal care.

Our transplant team is multi-faceted and includes transplant surgeons, infectious disease specialists, hepatologists, gastroenterologists, physician assistants, nurses, transplant RN coordinators, nutritionists, social workers, child life specialists, physical and occupational therapists and many others who provide care specifically tailored to each child's needs.

I hope you find this guide on liver and intestine transplantation at the Hillman Center for Pediatric Transplantation to be a useful resource as you make the best recommendations for your patients. I invite you to contact me or Linda Boig, RN, BSN, CCTC, supervisor, at the numbers listed in this guide should you have any questions or need any additional information.

Sincerely,



George V. Mazariegos, MD  
*Director, Pediatric Transplantation  
Hillman Center for Pediatric Transplantation  
Children's Hospital of Pittsburgh of UPMC  
Jamie Lee Curtis Professor of Surgery and Critical Care Medicine  
University of Pittsburgh School of Medicine*





## Partnering With Referring Physicians Provides Key for Success

Children's Hospital of Pittsburgh of UPMC's successful Hillman Center for Pediatric Transplantation combines a high level of medical expertise with a team approach to evaluating, supporting and managing our liver and intestine transplant recipients. Patients are taking fewer medications, living longer lives and experiencing fewer side effects. This allows our young patients to do what they do best — be kids.

With pediatric survival rates that are among the best in the world, Children's Hospital of Pittsburgh's Liver and Intestine Transplant Program continues to offer young patients — like liver transplant survivor Carleigh Plotts (right) — and their families, not only health, but hope. Christine Plotts gave life to Carleigh, not once, but twice. The first time was in 2000: a delivery, not unlike any other birth. The second was a little more than a year later, when Carleigh was suffering a downward spiral due to a failing liver. A living-organ donation from Christine — part of her own liver — gave her daughter the chance at life she so desperately needed.

### Comprehensive Care

Children's Hospital of Pittsburgh's unique group of dedicated clinicians, surgeons and scientists, who also are affiliated with the prestigious Thomas E. Starzl Transplantation Institute at UPMC, are just a few of the members of a comprehensive team dedicated to providing the very best in pediatric transplant care. The team also works closely with the referring physician of every patient. Many different liver and intestine experts are available around the clock for any questions that referring physicians may have. In addition, a variety of referral resources are also available to assist throughout the process.



## A RICH HISTORY OF INNOVATION

Children's Hospital of Pittsburgh of UPMC holds a prominent place in the history of pediatric transplantation. Children's Hospital performed its first pediatric kidney transplant in 1964. And, in 1981, the hospital opened the country's first pediatric transplant center under the guidance of transplant pioneer Thomas E. Starzl, MD, PhD. Dr. Starzl and his team were instrumental in the development of FK506 (tacrolimus) and were the first to use the antirejection agent, which has significantly improved patient and graft survival rates for liver and other organ transplants, including the intestine.

Groundbreaking research and advanced training programs at Children's have developed innovative therapies for previously fatal liver and intestinal disorders. Treatment protocols for pediatric intestinal transplants, both alone or in combination with other organs, now provide a viable surgical solution for children with intestinal diseases. Children's Hospital of Pittsburgh's Transplant Program performed the world's first and most successful series of small intestine transplants.

As the outcomes for liver transplant patients have improved significantly over the last two decades, access to deceased donor organs continues to be a major problem. To help overcome the organ shortage, living-donor liver transplantation has been developed as a life-saving alternative. Children's Hospital of Pittsburgh of UPMC is the leading center for pediatric living-donor liver transplantation. Potential advantages may include the increased ability to lower immunosuppression and further improve the quality of life.

The first living-donor liver transplant was performed in 1997 on an 18-month-old boy with biliary atresia, who successfully received a portion of his father's liver. Surgeons are skilled in left and right hepatectomies, which offer a child of any size or age the option of a living-donor transplant.

# REFERRAL AND TRANSPLANTATION PROCESS

## Referral

A demographic and clinical summary (available in the back of this booklet) should be completed and sent to the liver and intestine transplant team. The pre-transplant RN coordinator will collect the medical history, radiology studies, recent blood work and current history and physical for the patient. The information is reviewed by the surgical and hepatology/gastroenterology team and sent to our business office for insurance authorization. After authorization is obtained, a transplant evaluation is scheduled.

## Evaluation

The evaluation for a liver transplant is conducted on an outpatient basis and typically requires three to four days in Pittsburgh. The evaluation for an intestine or liver/intestine transplant is conducted on an inpatient basis and typically requires a Monday-through-Friday hospital visit. The evaluation includes diagnostic testing, consultations by Anesthesia, Cardiology, Hepatology/Gastroenterology, Psychology, Social Work and education on the entire transplant process. At the conclusion of the evaluation, the patient is presented to the multi-disciplinary team for consideration of transplant.

## Pre-Transplant

After the evaluation, the patient is discharged to his/her local community. Children's will list the patient with the United Network for Organ Sharing (UNOS) and obtain authorization for the transplant procedure from the insurance company. The patient's listing status with UNOS is based on his or her current medical condition and blood work. During the waiting period, the pre-transplant RN coordinator will be in communication with the referring physician to keep him or her informed about the patient's condition and ensure the patient's score with UNOS is accurate. The hepatologist (for liver transplants) and the Intestinal Care and Rehabilitation Center (ICARE) (for intestine transplants) work with the referring physician to develop a comprehensive plan of treatment.

## Transplantation

When an organ is made available, the transplant coordinator notifies the patient for transport to Pittsburgh. The patient is admitted to the Transplant Center and is prepped for surgery. During the surgery, the family will be updated about the patient's condition and progress of the surgery. Immediately after surgery, patients go to the intensive care unit (ICU). The length of time spent in the ICU varies based on each child's needs. Once the patient is stable, he/she will be sent back to the Transplant Center.

## Post-Transplant and Follow-Up Care

The transplant educator will review all medications, activity restrictions, diet, and any other specific instructions the family will need to care for the child. The child will remain in the Pittsburgh area until he/she is stable. The staff will educate the family on the frequency of blood work and dates for clinic appointments. Medications will be adjusted and wound care continued.

At a typical clinic visit, the doctor and the nurse will discuss the child's current health, check the child's weight and blood pressure, and perform a physical examination. Blood work will be done to monitor the patient's acceptance of the organ. One of the most important aspects of ensuring the health of a transplant patient is the careful maintenance of immunosuppressant medications to avoid rejection and prevent infection.

Throughout the transplant process, the referring physician will be continually updated by the transplant surgeons and/or the transplant coordinator on the patient's progress. When the patient is discharged, the transplant coordinator will send the referring physician a packet of information outlining a treatment regimen, including medical records, current medications and recommended laboratory schedule. Families will be advised to make an appointment with the child's referring physician. Transplant surgeons will continue to keep the referring physician aware of any changes to immunosuppression medications and annual evaluations.





“This is a modern-day miracle ...  
Children’s doctors gave Haden a  
chance at life.”

*Nanette Thomas, mother of Haden Thomas.*

*At 18 months old, Haden received a portion of his father’s (Lynn Thomas) liver during the area’s first living-related pediatric liver transplant. The long-term survival rate for patients who have undergone living-donor liver transplants is more than 90 percent.*

Children's Hospital of Pittsburgh of UPMC's transplant program treats a variety of disorders. In addition to addressing acute and end-stage liver disease, Children's offers transplants for conditions that, while not immediately life-threatening, impair development and diminish the child's quality of life.

For example, Children's Hospital of Pittsburgh is among the most accomplished centers in the use of transplantation to address metabolic disorders. In 2004, Children's developed the first and most comprehensive, multidisciplinary medical protocol for liver transplants to cure maple syrup urine disease (MSUD), a potentially deadly metabolic disorder. Children's has performed more successful transplants for MSUD than any other center in the country.

In the case of portal hypertension, the patient does not often need transplantation. Children's offers alternative surgeries and treatments to treat this condition. Examples include: meso-*rex* shunt, distal spleno-renal shunt and transjugular portosystemic shunt (TIPPS).

For patients diagnosed with liver cancers such as hepatocellular carcinoma (HCC) or hepatoblastoma, Children's also offers liver resection surgery as an alternative to transplant. Children's is the national leader in treating children with hepatoblastoma.

Children's Hospital of Pittsburgh has taken intestinal transplantation from a procedure considered prohibitive just a decade ago to one that today offers patients improved chances of long-term survival and a much higher quality of life. Children's performed the first and most successful series of pediatric small bowel transplants beginning in 1990. Today, the preconditioning of intestinal transplant patients with thymoglobulin or Campath® has greatly improved outcomes. Graft rejection has decreased, hospital stays

Children's Hospital of Pittsburgh has taken intestinal transplantation from a procedure considered prohibitive just a decade ago, to one that today offers patients improved chances of long-term survival and a much higher quality of life.

are shorter, and patients experience fewer post-surgical complications than ever before.

Children's Hospital of Pittsburgh also has developed tests that enable specialists to detect complicating infections early and to treat pre-emptively threats such as cytomegalovirus and Epstein-Barr virus.

For short-gut syndrome, Children's offers lengthening procedures including Bianchi and STEP, aggressive attempts at nutritional rehabilitation and innovative therapies such as intravenous omega 3 fatty acid as a part of institutional review board-approved protocols.

# Diseases Children's Hospital of Pittsburgh of UPMC has treated with liver and intestinal transplantation

## Diagnosis for Liver Transplant (isolated)

- 54% Biliary atresia
- 14% Metabolic disease
- 11% Acute hepatic necrosis
- 7% Miscellaneous diseases
- 7% Autoimmune and other cirrhosis
- 4% Cholestatic liver disease
- 2% Malignancy
- 1% Congenital hepatic fibrosis



## Diagnosis for Intestine Transplant (isolated and multi-organ)

- 25.0% Volvulus
- 24.4% Gastroschisis
- 13.3% Other
- 10.6% Pseudo-obstruction
- 10.0% Necrotizing enterocolitis
- 8.9% Jejunal atresia
- 7.8% Hirschsprung's



## UNMATCHED VOLUMES AND OUTCOMES

To date, Children’s Hospital of Pittsburgh of UPMC has performed more than 1,400 pediatric liver and intestine transplants. It is this large volume of procedures that has made the Hillman Center one of the most active pediatric transplant centers in the nation. The hospital receives referrals from transplant centers, hospitals and physicians all over the world. These referrals include the most complex and difficult cases. Despite these challenging cases, Children’s outcomes remain among the best in the world.

### Three-Year Patient Survival

Liver

**96%**

(National average: 88%)

Intestine

**91%**

(National average: 66%)

### Transplant Program Volumes\*

Type-Abdominal	2006	2007	Program Inception through 12/31/2007
Liver and Liver/Kidney	22	21	1,240
Living-Donor Liver	10	15	55
Liver/Small Bowel	4	7	97
Small Bowel/Multivisceral	11	12	104
<b>Total</b>	<b>47</b>	<b>55</b>	<b>1,496</b>

\* Sources: Internal data; Scientific Registry of Transplant Recipients ([www.ustransplant.org](http://www.ustransplant.org))



### A Note on Children’s Outcomes Data

Many aspects of transplantation care comprise a transplant center’s actual outcomes. There are many factors to take into consideration, including the patient’s condition, the donor, etc. Some centers perform transplants on critically ill children, like Children’s, but all define “critical” differently. Our most critical patients include those with acute liver failure, those under the age of 2, as well as living donor and patients requiring re-transplantation (most often initially transplanted at other centers). Children’s receives referrals from all over the world, including many highly complex cases.

Additional outcomes information can be found at [www.unos.org](http://www.unos.org)



# Did you know that Children's Hospital of Pittsburgh of UPMC ...

... was the nation's first pediatric transplant center?

... pioneered pediatric multiorgan transplantation, including the world's first pediatric heart-liver transplant in 1984?

... runs Camp Chihopi, an annual summer camp for liver and intestine transplant recipients since 1994?

... features more than 55 experts in a variety of subspecialties, and that our four transplant surgeons have nearly 40 years of combined experience?







Tracey was an extremely sick toddler in 1990 when her life was saved by a liver and intestine transplant at Children's.

It's sometimes hard to fully comprehend the impact that Children's Hospital of Pittsburgh of UPMC's transplant surgeons have had over the last 27 years. Hundreds of patients with little hope as infants or toddlers are now young adults. The program has expanded to offer operations for children who need intestinal transplant or multivisceral transplant procedures previously thought to have been impossible.

Tracey Gonzalez was one such patient. Tracey was an extremely sick toddler in 1990 when her life was saved by a liver and intestine transplant at Children's Hospital. She was Children's — and the nation's — first such successful combined liver and intestine transplant. Today, Tracey (left) is a vibrant 20-year-old who is attending college and even volunteers as a counselor at Children's Hospital's Camp Chihopi, an annual summer camp for organ transplant survivors.



1,500+

... liver and intestine transplants have been performed by Children's, more than any other pediatric center in the world.



“Children’s Hospital has given so much to Renee. We’re so proud that she wants to return the favor.”

*Gayle Williams, mother of Renee Williams.*

*Renee Williams received a liver transplant in 1985. Renee regularly volunteers at the Hillman Center for Pediatric Transplantation.*



## Improving Quality of Life After Transplantation

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Improved survival rates have focused more attention on finding ways to make recovery less difficult and give children a quality of life vastly better than what patients could expect a decade ago.

Following surgery, the quality of life Children's Hospital of Pittsburgh of UPMC patients will enjoy, often managed under our steroid-free simplified immunosuppressive regimens, is exceptional. Their body image is better, their growth is superior, and the risk of infection is very low. In fact, Children's surgeons have developed tests that enable them to detect complicating infections early and to treat pre-emptively threats such as cytomegalovirus and Epstein-Barr virus. More than 90 percent of Children's long-term pediatric intestinal patients no longer need total parenteral nutrition and the number of patients with long-term need of a catheter, ileostomy, gastrostomy tube, physical therapy or other post-transplant therapy has dramatically decreased.

Experience at Children's Hospital of Pittsburgh suggests that the better families are prepared to deal with the challenges of transplantation, the better their children's outcomes are likely to be. Children's offers comprehensive services to help children and their families cope with transplantation and all that recovery demands. Services include instruction on caring for a child while in the hospital, training for caring for the child at home, and access to social workers, psychologists, occupational therapists, physical therapists and others who can help with the physical, emotional, social, educational and even the financial challenges of transplantation.

*Children's Transplant Program offers access to a wide variety of world-renowned specialists including Robert H. Squires Jr., MD (above) specializing in gastrointestinal diseases and Benjamin L. Schneider, MD (right) who specializes in hepatology.*

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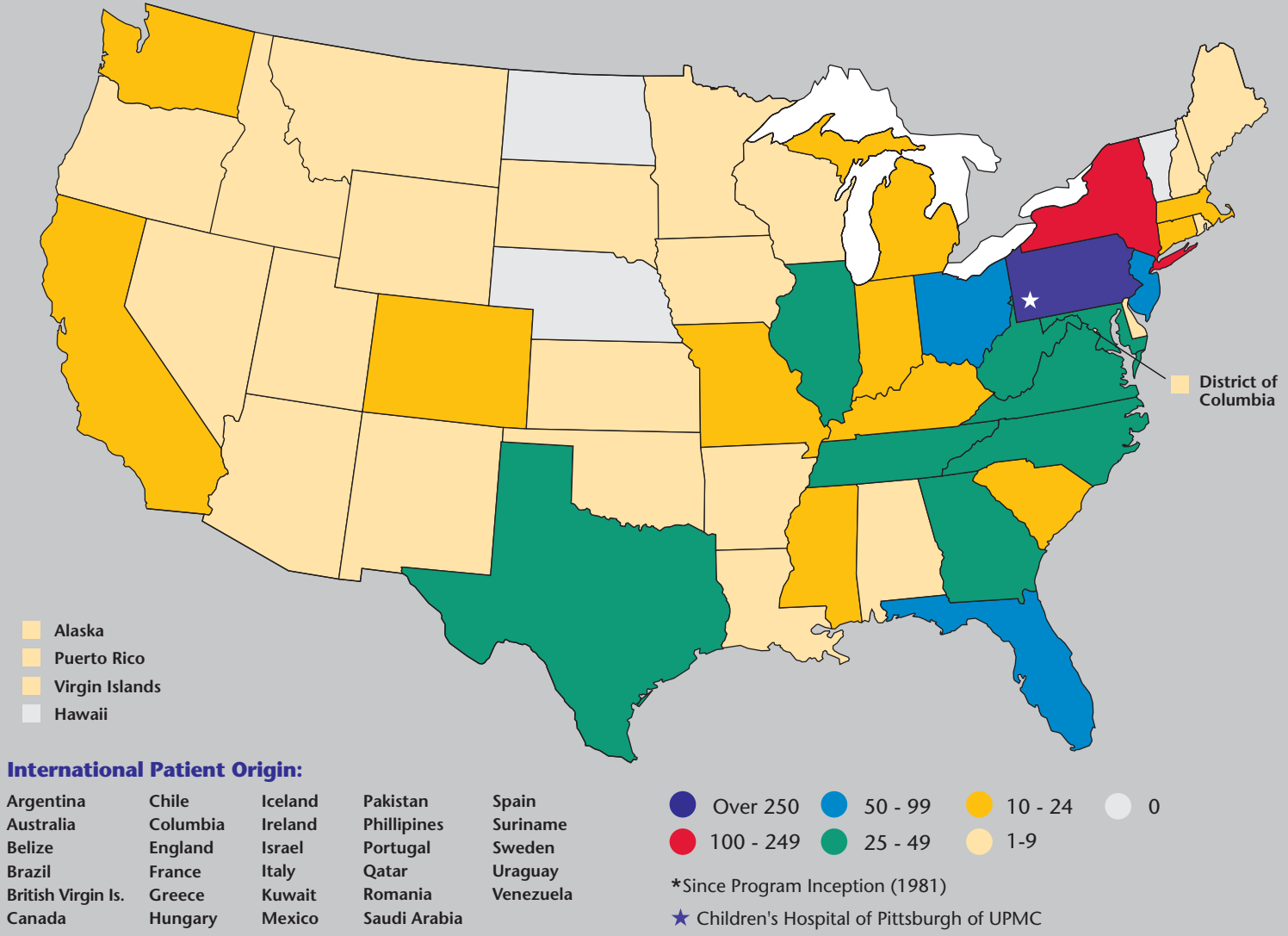


88%

... 10-year patient survival rate after liver transplantation for biliary atresia [Shneider; Mazariegos: "Biliary Atresia: A Transplant Perspective," *Liver Transplantation*, 13(11): 1482-95,2007.]



# Liver and Intestine Transplants: Patient Origin by State by Volume\*



## RESEARCH AND INNOVATION: THE FOREFRONT OF OUR APPROACH TO TRANSPLANT MEDICINE

The basic science and clinical research under way at Children's Hospital of Pittsburgh of UPMC's Hillman Center for Pediatric Transplantation follows a rich tradition of innovation that has improved the lives of pediatric transplant patients dramatically through advancements in surgical technique, organ procurement and the use of immunosuppression.

Today, researchers in Children's Hospital's transplant programs are improving drug and diagnostic protocols that will allow them to deliver personalized medicine. Anti-rejection regimens are being evaluated for their ability to bring about selective suppression of the host immune system toward the graft. General immune processes that protect against life-threatening infections may be spared by these approaches. To improve care after transplantation, experimental testing is being evaluated that will predict the risk of rejection. Once known, minimization of anti-rejection therapy can be timed to individual need, so that rebound rejection is prevented.

Directed by Rakesh Sindhi, MD (right), the Pediatric Transplant Research Laboratory is identifying genomic "fingerprints" that differentiate children prone to rejection from those who never experience rejection on the usual anti-rejection medicines. Prior knowledge of such patterns will allow tailored therapy to be initiated at the time of transplantation, prior to the occurrence of any rejection event. For example, less medicine may be given to children with better tolerance for the organ, and more may be given to those predicted to have a greater risk of rejection.

Currently, our physicians and scientists have studies under way totaling more than \$13.6 million in grants, awards and endowments.

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Children's Hospital of Pittsburgh of UPMC participates in National Institutes of Health-funded, multicenter studies investigating acute pediatric liver failure, biliary atresia, neonatal cholestasis and rare liver diseases such as alpha-1 antitrypsin deficiency and mitochondrial diseases that involve the liver.



Rakesh Sindi, MD  
Transplant Surgery



“We feel so lucky to have been given another chance for Jakob to have his life back.”

*Susan Jasin, mother of Jakob.*

*Jakob was born with Maple Syrup Urine Disease (MSUD).*

*At the age of 5, Jakob was transplanted. Today, he is a vibrant, 8-year-old boy free of all symptoms.*

**UNMATCHED OUTCOMES. UNCOMMON CARE.**

**Pediatric Liver and Intestine Transplantation**



[www.chp.edu](http://www.chp.edu)

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