UPMC CHILDREN'S
HEART INSTITUTE

2024 OUTCOMES AND HIGHLIGHTS

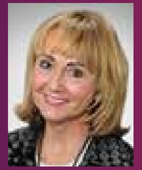
UPMC | CHILDREN'S
HOSPITAL OF PITTSBURGH

The Heart Institute at UPMC Children's Hospital of Pittsburgh is a destination center for pediatric heart care. Our multidisciplinary team of specialists brings together experts from cardiac surgery, cardiology, interventional cardiology, cardiac imaging, and heart and lung transplant to provide the full scope of care to our patients. Through a collaborative effort between UPMC Magee-Womens Hospital, UPMC Children's, and the UPMC Heart and Vascular Institute, we care for patients with congenital heart disease in every stage of life.

Each year, the Heart Institute welcomes more than 18,000 patients from around the world to receive care with our team. We consistently achieve outstanding surgical outcomes that earn our program recognition from prestigious national forums, including the Society of Thoracic Surgeons (STS) and *U.S. News and World Report*. Our team's dedication to research and innovation continually advances the care that we provide patients.

OUR LEADERSHIP TEAM

Victor O. Morell, MD
Eugene S. Wiener Endowed Professor
of Pediatric Cardiothoracic Surgery
Chair, Department of Cardiothoracic Surgery
Co-Director, Heart Institute at
UPMC Children's Hospital of Pittsburgh



Jacqueline Kreutzer, MD
Peter and Ada Rossin Endowed Professor
of Pediatric Cardiology
Chief, Division of Cardiology,
Department of Pediatrics
Co-Director, Heart Institute at
UPMC Children's Hospital of Pittsburgh

Avihu Z. Gazit, MD
Patrick Dick Endowed Chair in Cardiac Critical Care
Chief, Pediatric Cardiac Critical Care Medicine
Director, Cardiac Critical Care Services
Co-Director, Heart Institute at
UPMC Children's Hospital of Pittsburgh

AT A GLANCE

- **The Heart Institute is home to the Da Silva Center for Ebstein's Anomaly**, where **Luciana da Fonseca da Silva, MD**, and **José Pedro da Silva, MD**, the creator of the Cone procedure, operate on patients from around the world with Ebstein's anomaly.
- **The Heart and Lung Transplant Program at UPMC Children's has performed 544 organ transplants** in children to date, including 404 heart transplants, 95 lung transplants, and 45 heart-lung transplants.
- **The Heart Institute is a member of the Pediatric Heart Network (PHN)**, an international consortium of nine congenital heart programs across North America.
- **UPMC Children's has more than 30 years of experience** using ventricular assist devices (VADs) to help children in heart failure.
- **We are an Extracorporeal Life Support Organization (ELSO) Platinum Center of Excellence** – the highest designation for Excellence in Life Support by ELSO.
- **The UPMC Adult Congenital Heart Disease Program is accredited by the Adult Congenital Heart Association (ACHA)** for the program's excellence in clinical care and patient support, one of only 50 centers across the nation to receive this recognition.

PROGRAM HIGHLIGHTS

THE SINGLE VENTRICLE CLINIC AT THE HEART INSTITUTE

The Single Ventricle Program at the Heart Institute at UPMC Children's Hospital of Pittsburgh was launched in March 2024 to address the complex and multidisciplinary needs of infants with single ventricle congenital heart defects during the high-risk interstage period.

Leading the development of the program and its ongoing work is a dedicated team, including pediatric cardiologists **Lizabeth Lanford, MD**, associate professor of Pediatrics, and **Evonne Morell, DO**, associate professor of Pediatrics and director of Inpatient General Pediatric Cardiology.

Navigating the High-Risk Interstage Period

Single ventricle congenital heart defects (SV) are among the most complex congenital heart conditions faced by pediatric cardiology and surgery teams. These defects require multiple staged surgical interventions, with the interstage period — the time between the patient's first procedure (Norwood procedure) and second (Glenn procedure) surgeries — being particularly precarious.

"The interstage period carries significant risks, with historical mortality rates as high as 20% two decades ago," says Dr. Morell. "Families were frustrated by prolonged hospital stays, but the lack of structured follow-up and technological advancements in care and remote monitoring made home care too risky in most cases in prior years. We knew we needed a better solution."

The primary goal of the Single Ventricle Program is to ensure infants receive the necessary medical oversight and care while benefiting from the developmental and emotional advantages of being at home instead of the hospital. Families of single ventricle patients play a central role in this effort.

Core Elements of the Program

The Single Ventricle Program is built around a multidisciplinary team that includes pediatric cardiologists, advanced practice providers (APPs), neurodevelopmental specialists, social workers, and therapists. Together, the team collaborates to ensure that each family entering the program is fully equipped and trained to care for their child at home while they prepare for their next surgery.

Families undergo extensive preparation for caring for their child at home, beginning weeks before discharge. The training process is rigorous, covering medication administration, feeding techniques for both oral and tube-fed patients, and CPR certification. Parents are also trained to identify critical red flags for when either an emergency room visit is necessary or a call to emergency medical services. To support families, the program provides each with a personalized binder tailored to their child's specific needs and plan of care.

"The training is intense but necessary," says Dr. Lanford. "We ask a lot from these families. The preparation is comprehensive because the safety and well-being of the patient depend on their readiness and ability to respond to the needs of their child."

Another part of the preparation family caregivers must go through and successfully navigate is a live-in trial at the Heart Institute, during which families spend 24 hours providing, managing, and ordering all aspects of care for their child under hospital supervision. This trial ensures that parents are confident and capable of managing the complex care their child requires at home.

Community integration is another element of the program that makes it possible for single ventricle patients and families to be at home instead of the hospital. Upon discharge, local emergency medical services (EMS) and emergency departments (EDs) are notified about the presence of these high-risk patients in their area.

"This proactive communication ensures that first responders and local health care providers are familiar with the specific care protocols required for these interstage patients, further enhancing their safety," says Dr. Lanford. "We make it a priority that these groups know what the plans of action are should their services be needed, and it's specific to the individual case."

Improved Quality of Life and Outcomes Through Care at Home

The benefits of home care for interstage single ventricle patients are profound. Infants who transition to home often demonstrate better feeding and developmental outcomes than those who remain hospitalized for the entirety of the interstage period.

"At home, babies receive the individualized attention and stimulation that are critical for their optimal development," says Dr. Morell. "The improvements in feeding that we see in patients living at home alone can make a significant difference in their overall health and ability to tolerate their subsequent surgical needs."

Evidence in the literature and also from data tracking at UPMC Children's suggests that sending patients home during the interstage period may also extend the time between the Norwood and Glenn procedures.

"By allowing patients to grow stronger and healthier at home, we've been able to delay the Glenn procedure until the baby is more developed, often closer to five or six months of age," says Dr. Lanford. "This additional time can improve surgical outcomes and overall recovery, and it's one of the areas in which we hope to study rigorously."

Expanding the Program's Reach

The Single Ventricle Program at the Heart Institute at UPMC Children's exemplifies how a multidisciplinary, family-centered approach can transform outcomes for a complex and high-risk patient population. By addressing the known and anticipated challenges of the interstage period, the program ensures that patients receive the best possible care while empowering families to play an active role in their child's health care journey.

THE PITTFON PROGRAM AT UPMC CHILDREN'S HOSPITAL OF PITTSBURGH

Optimizing Long-Term Health for Fontan Circulation Patients

The Fontan procedure is a lifesaving surgery for patients born with single ventricle congenital heart disease. While the procedure has allowed many individuals to survive and thrive into adulthood, the long-term consequences of Fontan circulation can result in downstream morbidities potentially affecting multiple organ systems. Recognizing the complexity of managing Fontan patients as they age, the Heart Institute at UPMC Children's Hospital of Pittsburgh, along with colleagues from other divisions, has developed PittFON—the Pittsburgh Innovative Therapeutic Approach for Patients with Fontan Circulation. This comprehensive, multidisciplinary initiative is designed to optimize care, enhance outcomes, and address the many health challenges that can be associated with Fontan circulation.

Fontan Circulation: A Multisystem Clinical Challenge

The Fontan procedure creates a unique circulatory path where blood bypasses what would normally be the right ventricle, instead relying on passive flow from the body's veins directly to the lungs via the pulmonary arteries. While effective in managing single ventricle congenital heart defects, this abnormal circulation results in increased central venous pressure and decreased cardiac output, both of which can lead to complications affecting the liver, kidneys, lungs, and neurodevelopmental systems over time.

"While the Fontan procedure is primarily a cardiac solution, the long-term effects extend far beyond the heart," says **Tarek Alsaied, MD, MSc, FACC**, pediatric cardiologist at the Heart Institute and one of the leaders of the PittFON program. "We can see multi-organ involvement in these patients, and that's what we're working to address with PittFON. The goal is to catch morbidities early and prevent them from leading to significant deterioration."

A Comprehensive Approach to Complex Care

The PittFON program is structured to provide ongoing, comprehensive care for Fontan patients, focusing on both routine surveillance and early detection of complications. Currently, the program follows approximately 350 to 400 patients across the UPMC system. These patients are closely monitored through standardized testing protocols that focus on cardiac function and the health of other organ systems, particularly the liver, kidneys, lungs, and brain.

The testing protocol includes regular evaluations such as liver ultrasound, cardiac MRI, stress tests, and neurodevelopmental assessments to monitor for early signs of organ dysfunction.

"Some patients may need early intervention, whether that's a heart transplant, a heart-liver transplant, or other surgical procedures," says Dr. Alsaied. "Our multidisciplinary team works together to identify these patients early and ensure timely care. My colleagues, **Lizabeth Lanford, MD,** and **Evonne Morell, DO**, continue to follow many Fontan patients through our comprehensive single ventricle program."

The program also benefits from the neurodevelopmental expertise of **Lauren Lorenzi, PhD**, and her team, who provide comprehensive evaluations and interventions for any neurodevelopmental concerns in Fontan patients.

Given the frequency at which Fontan patients require cardiac catheterization interventions, the PittFON Program works in tandem with the Heart Institute's interventional cardiology team, including **Bryan H. Goldstein, MD**, **Jacqueline Kreutzer, MD,** and **Sara Trucco, MD**. This collaboration ensures seamless care for patients requiring catheter-based therapies.

"Additionally, we have a close collaboration with our pediatric heart transplant team, referring patients as needed based on their clinical picture for heart transplant evaluation," says Dr. Alsaied.

Regular "Tune-Ups" and Long-Term Monitoring

Patients are seen in the PittFON clinic every three to five years for a full evaluation, also referred to as a "tune-up". These visits allow the care team to review all test results and make any necessary adjustments to the patient's care plan.

Between these visits, patients continue to follow up with their primary cardiologists. The program also supports different patterns of care, allowing some patients to co-manage their care between the PittFON clinic and their local providers. This flexible model accommodates patients who live far from UPMC Children's, ensuring they still receive expert guidance and monitoring even when they cannot visit the clinic regularly.

A Lifelong Commitment

Fontan patients need lifelong care, and the Heart Institute at UPMC Children's and the UPMC Heart and Vascular Institute have a robust Adult Congenital Heart Disease (ACHD) program, led by **Arvind Hoskoppal, MD, MHS**, **Anita Saraf, MD, PhD**, and **Natasha Wolfe, MD**, ensuring a seamless transition as patients grow older.

"Once patients reach their mid-20s, they transition to our ACHD team, but we continue to collaborate closely," says Dr. Alsaied. "This is crucial because many complications, particularly involving the liver and kidneys, often manifest later in life."

Collaborating with the ACHD team ensures that Fontan patients continue to receive expert care tailored to their evolving needs into adulthood.

LEADERSHIP NEWS AT THE HEART INSTITUTE

The Heart Institute at UPMC Children's Hospital of Pittsburgh is excited to welcome Avihu Gazit, MD, as Patrick Dick Endowed Chair in Cardiac Critical Care, chief of Pediatric Cardiac Critical Care Medicine, director of Cardiac Critical Care Services, and co-director of the Heart Institute. Dr. Gazit specializes in pediatric critical care medicine and is board-certified in pediatric critical care medicine, pediatric cardiology, and pediatrics by the American Board of Pediatrics.

Clinical and Research Interests

Dr. Gazit's primary clinical focus is on the care of critically ill patients in the cardiac intensive care unit. His current research interests include:

- Validation of near real-time physiological algorithms in pediatric intensive care.
- Utilizing pre-extubation IDO2 dose, heart rate, and blood pressure variability to predict extubation readiness in infants with critical congenital heart disease following cardiopulmonary bypass operations.
- Analysis of patterns of clinical deterioration in critically ill neonates with congenital heart disease.
- Utilizing cerebral near-infrared spectroscopy saturations and arterial blood pressure data to investigate postoperative patterns of cerebral autoregulation in neonates with CHD who underwent cardiac surgery with cardiopulmonary bypass in the first four weeks of life and to identify clinical factors associated with impaired cerebral autoregulation.

Dr. Gazit serves on several critical care medicine and pediatric cardiac intensive care society committees and on the editorial boards of multiple critical care and cardiology journals. His numerous publications reflect his dedication to advancing the field of pediatric cardiac critical care medicine.

Dr. Gazit brings decades of experience, expertise, and first and foremost dedication to patient- and family-centered care to the Heart Institute at UPMC Children's Hospital of Pittsburgh, a nationally recognized institution for pediatric and congenital heart care.

THE HEART INSTITUTE AT UPMC CHILDREN'S CENTER FOR COMPLEX TRANSPOSITION

The Heart Institute at UPMC Children's Hospital of Pittsburgh recently unveiled the Center for Complex Transposition. This center provides specialized care and treatment for complex congenital heart defects, with a special focus on transposition of the great arteries (TGA).

TGA is a group of congenital heart defects which occurs when the two main blood vessels leaving the heart are reversed. In typical TGA, the aorta is connected to the right ventricle and the pulmonary artery is connected to the left ventricle. This reduces the amount of oxygen-rich blood sent to the body.

Babies with TGA can require different treatment plans depending on the specific structural defects present in their hearts. Our team of experts specialize in treating TGA and collaborate to create personalized treatment plans for each patient.

A Comprehensive Approach to TGA Care

Led by **Victor Morell, MD**, co-director of the Heart Institute at UPMC Children's, chair of the Department of Cardiothoracic Surgery, and VP and Chief Strategy Officer of the UPMC Heart and Vascular Institute, and **Laura Olivieri, MD, FAHA, FSCMR**, director of noninvasive cardiac imaging at the Heart Institute, the Center for Complex Transposition is a destination for specialized surgical care and advanced cardiac imaging for patients with TGA.

At the Heart Institute, our team achieves excellent surgical outcomes that are among the best in the country. Our surgeons work closely with our imaging specialists to determine the best treatment plan for each patient. Our expertise in all aspects of complex congenital heart defects brings patients from around the world to our program to receive care.

Our advanced cardiac imaging program at UPMC Children's plays a major role in the center's approach to care. Through innovations like 3D modeling technology, we are able to enhance the diagnosis and evaluation process for TGA. Advanced imaging enhances our team's view of the condition and can improve our surgical approach.

New or expecting parents whose baby is diagnosed with transposition of the great arteries (TGA) have access to a multidisciplinary team of experts at the Center for Complex Transposition. Teams across UPMC Children's collaborate to provide specialized, comprehensive care to patients with TGA. Our multidisciplinary approach brings together experts from fetal and perinatal cardiology, advanced imaging, the CICU, and the Adult Congenital Heart Disease Center.

THE DA SILVA CENTER FOR EBSTEIN'S ANOMALY

Led by surgical director and creator of the Cone procedure, **José Pedro da Silva, MD**, alongside **Luciana da Fonseca da Silva, MD**, the Da Silva Center for Ebstein's Anomaly at the Heart Institute provides world-class care to patients of all ages with Ebstein's anomaly, a rare congenital heart disease.

Over three decades ago, Dr. da Silva created the Cone procedure to repair Ebstein's anomaly. The Cone procedure creates a durable repair of the malformed tricuspid valve (TV) using the patient's own tissue, which creates a TV capable of appropriate growth.

Cone Repair After Starnes Procedure

Our team has had great success in performing Cone repair following the Starnes procedure. The Starnes procedure is a surgical technique used to treat Ebstein's anomaly in which the surgeon closes the malfunctioning right ventricle, creating a single ventricle circulation to improve the patient's heart function. The Da Silva Center team recently published new research on Cone repair following the Starnes procedure in *The Journal of Thoracic and Cardiovascular Surgery: "The Cone Repair Allows Right Ventricle Rehabilitation with Excellent Tricuspid Valve Function Following the Starnes Procedure"*.

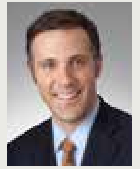
Outcomes

Our team's commitment to surgical excellence, combined with our dedication to patient-centered care, has allowed us to achieve great outcomes for many patients with Ebstein's anomaly.

- Our program has performed more than 140 Cone procedures to date.
- We average 30 Cone procedures per year.
- We have maintained an all-time 0.00% mortality rate for Cone procedures.
- In the last two years, we have performed 18 Cone procedures following Starnes procedures.

STUDY FROM UPMC CHILDREN'S HEART INSTITUTE: TOP 10 MOST CITED ARTICLES IN PEDIATRIC TRANSPLANTATION

A study published by the Heart Institute at UPMC Children's Hospital of Pittsburgh in the journal *Pediatric Transplantation* on "Early Findings After Integration of Donor-Derived Cell-Free DNA Into Clinical Care Following Pediatric Heart Transplantation" was one of the top 10 most cited articles from the journal in 2022-2023.



The study was led by **Brian Feingold, MD, MS**, professor of Pediatrics and Clinical and Translational Science, and medical director of the Heart Failure and Transplantation Programs at UPMC Children's.

This study, conducted in a group of pediatric heart transplant patients at UPMC Children's, provided the first real-world clinical data on the feasibility, reliability, and success of using donor-derived cell-free DNA (dd-cfDNA) testing in lieu of endomyocardial biopsy as part of routine post-transplantation care and surveillance for signs of rejection in this population.

Pediatric Transplantation Study Reference

Feingold B, Rose-Felker K, West SC, Zinn MD, Berman P, Moninger A, Huston A, Stinner B, Xu Q, Zeevi A, Miller SA. Early Findings After Integration of Donor-Derived Cell-Free DNA Into Clinical Care Following Pediatric Heart Transplantation. Pediatric Transplant. 2022; 26: e14124.

Further Reading and Research by Dr. Feingold and Colleagues on the Use of dd-cfDNA

Feingold B, Rose-Felker K, West SC, Miller SA, Zinn, MD. Short Term Clinical Outcomes and Predicted Cost-Savings of dd-cfDNA-led Surveillance After Pediatric Heart Transplant. Clin Transplant. 2023 May; 37(5): e14933. Open Access.

OUTCOMES

Benchmark Operation Mortality

Our congenital cardiac surgery program consistently receives prestigious ratings from the Society of Thoracic Surgeons. The following data represents UPMC Children's mortality percentage compared to the STS overall performance mortality percentage for the listed series of benchmark procedures.

Overall Mortality for All STAT Mortality Categories as reported by the STS:
2.57% Observed vs 2.93% Expected

Children's Hospital of Pittsburgh Society of Thoracic Surgeons Benchmark Procedures versus STS Overall Performance-2024 Fall Report (7/1/20-6/30/24)

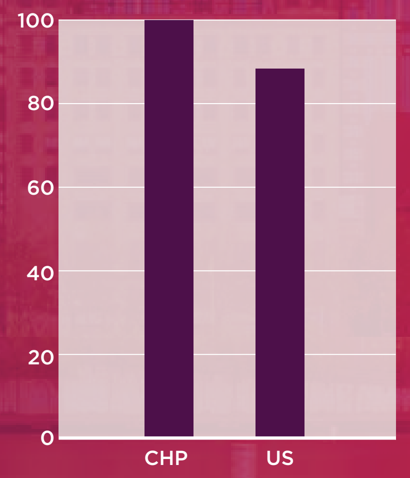
Benchmark Procedure	Children's Hospital of Pittsburgh Mortality Percentage	STS Overall Performance Mortality Percentage (IQR)
Off Bypass Coarctation	0%	1.04% (0-1.75)
VSD	0%	0.30% (0-0.52)
TOF	0%	0.75% (0-1.01)
AVC	2.86%	2.10% (0-2.86)
ASO	0%	1.91% (0-2.70)
ASO+VSD	0%	4.13% (0-3.70)
Glenn/Hemi-Fontan	0%	1.50% (0-2.41)
Fontan	2.17%	1.02% (0-1.69)
Truncus	0%	6.99% (0-11.11)
Norwood	11.43%	12.35% (7.14-15)

POST TRANSPLANT PEDIATRIC OUTCOMES

*Based on SRTR published results January 2025

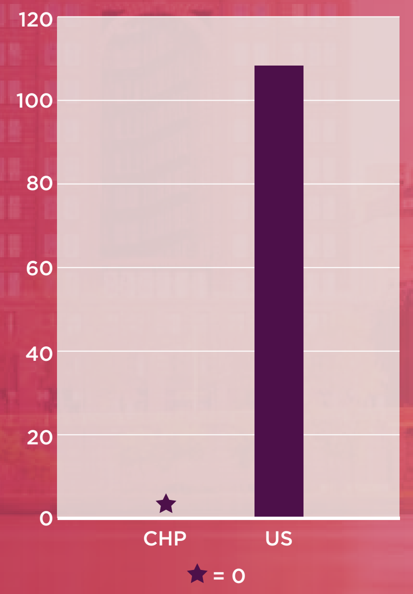
Estimated probability of surviving with a functioning graft at 3 years (unadjusted for patient and donor characteristics)

CHP 100% vs US 86.99%



Number of observed graft failures (including deaths) during the first 3 years after transplant

CHP 0 vs US 101



UPMC
LIFE CHANGING MEDICINE

OUR REFERRAL TEAM

At the Heart Institute at UPMC Children's, collaboration with referring providers is key to our mission of providing comprehensive care to children and adults with congenital heart disease. Our referral team maintains a robust second opinion program and welcomes the opportunity for consultations to carefully evaluate each case. We will work with you to create a plan specific to your patient's needs and maintain open communication throughout each stage of your patient's journey.

CONTACT US

For providers or patients within the United States, contact the Heart Institute at **412-692-5218** or email us at **CHPHeartReferral@chp.edu.**

For providers or patients outside the United States, contact our International Services Department at **+1-412-692-3000** or **international@chp.edu.**

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