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To access Texas Children’s Heart Center outcomes information online, please visit [texaschildrens.org/heartoutcomes](http://texaschildrens.org/heartoutcomes).
TEXAS CHILDREN’S HEART CENTER®

Texas Children’s Hospital has been a leader in pediatric heart care for more than 60 years. Led by Denton Cooley, M.D., and Dan McNamara, M.D., the hospital started performing heart surgery on children in 1956. Dr. McNamara, a patriarch of pediatric cardiology, and Dr. Cooley, a pioneer heart surgeon, were among the first to demonstrate that small children could safely undergo heart surgery.

Today, Texas Children’s Heart Center is a global leader in pediatric cardiac care, treating some of the rarest and most complex heart cases. With several multidisciplinary teams working in conjunction with pediatric subspecialists throughout the hospital, the Heart Center strives to provide unparalleled care at every point from diagnosis through treatment and follow-up, in order to achieve the best possible result for each patient.

For the third consecutive year, Texas Children’s Hospital is ranked as the #1 place in the nation for children to receive cardiology and heart surgery care by U.S. News & World Report in their 2019-20 edition of Best Children’s Hospitals. Texas Children’s Heart Center is committed to consistently achieving outcomes that are among the best in the country.

HEART CENTER MOVES TO NEW LESTER AND SUE SMITH LEGACY TOWER

Texas Children’s Heart Center had a monumental year in 2018 with the move into the new state-of-the-art Lester and Sue Smith Legacy Tower. The milestone came just months after the historic May 22 move of our pediatric intensive care and progressive care units into the spacious, high-tech tower.

The Move

Over the course of about eight hours on Sept. 25, six specially trained clinical teams comprised of more than 200 members transported 64 heart patients, some critically ill, safely to their new, state-of-the-art rooms in the tower. The patients ranged in age from 3 days to 22 years.

The following day, an 11-year-old patient became the first patient to undergo a cardiac catheterization procedure and an intra-cath MRI in the Heart Center’s new Charles E. Mullins, M.D. Cardiac Catheterization Laboratories.

On Sept. 27, the Heart Center’s new cardiovascular operating rooms were officially opened, ushering in a new era of cardiac surgery at Texas Children’s. Later that morning, a 4-year-old patient underwent the first surgical intervention in the expansive new space – a Fontan procedure.

And on Oct. 1, patients received treatment for the first time in the tower’s new therapy gym. The gym is a powerful resource to help children and parents learn and focus on what they can do, rather than what they can’t. Features include machines for building core strength and balance, exercise bikes and a bathtub and set of stairs for parents to practice everyday tasks at home with their children.

A week before the move, patients and families entered the doors of the Heart Center’s new outpatient clinic for the first time. Situated on the 21st and 22nd floors of the tower, the clinic is designed top to bottom with Texas Children’s families in mind. The bright, welcoming space was specially configured to offer a more personal approach to care, and to handle high clinical volume.
The Purpose
Texas Children’s started planning for the Lester and Sue Smith Legacy Tower more than five years ago in an effort to reinvest in the needs of our most critically ill patients. Demand for the level of care Texas Children’s Hospital provides continues to grow — here in our community and far beyond Houston. And prior to this expansion, our core areas – Critical Care, Emergency Center, ORs and PACU – were often at capacity.

As an organization, we saw an opportunity to advance quality, service, safety and strategic growth; broaden our expertise; better coordinate care to improve the patient/family experience; and expand access to make certain we do not have to turn children away when they need us most.

The tower is helping Texas Children’s accomplish all of this and more in the 640,000-square-foot space that includes:

• Eight floors for Texas Children’s Heart Center
• Seven floors of intensive care patient rooms
• One radiology suite
• Six high-intensity surgical operating rooms
• Four cardiovascular operating rooms
• Two intraprocedural MRIs
• Four cardiac catheterization labs
• One helistop

To learn more, visit legacytower.org.
Texas Children’s Hospital is affiliated with Baylor College of Medicine® in the areas of pediatrics, pediatric surgery and obstetrics and gynecology. Baylor is ranked by U.S. News & World Report as one of the nation’s top 10 medical schools for pediatrics. Currently and throughout our partnership, Texas Children’s Hospital serves as Baylor’s primary pediatric training site. The collaboration between Texas Children’s Hospital and Baylor is one of the top five such partnerships for pediatric research funding from the National Institutes of Health.

Physicians are employees of Baylor College of Medicine, not Texas Children’s Hospital. Because they practice at Texas Children’s Hospital, they may be referred to as “our team” or “Texas Children’s physicians” throughout this report.

**TEXAS CHILDREN’S HOSPITAL®**

Texas Children’s Hospital is one of the nation’s largest and most comprehensive specialty pediatric hospitals, with 4.3 million patient encounters in 2018. With a staff of more than 13,500 employees and 2,000 physicians, pediatric subspecialists, pediatric surgical subspecialists and dentists, Texas Children’s offers more than 40 subspecialties, programs and services.

The 2019-20 edition of Best Children’s Hospitals by U.S. News & World Report ranked Texas Children’s Hospital third in the country among nearly 200 pediatric centers. For the eleventh straight year, Texas Children’s Hospital was placed on the Best Children’s Hospital Honor Roll of best pediatric institutions, one of only 10 hospitals nationally and the only hospital in Texas awarded this distinction.

Texas Children’s Hospital is located near downtown Houston in the Texas Medical Center, the largest medical center in the world. This campus includes nearly 800 licensed beds; the Mark A. Wallace Tower for outpatient visits; the Feigin Tower for pediatric research; Texas Children’s® Pavilion for Women, a comprehensive OB/GYN facility with a focus on high-risk births; and the Texas Children’s Hospital® Jan and Dan Duncan Neurological Research Institute®, a basic research institute dedicated to solving childhood neurological diseases. Additionally, two community hospitals, Texas Children’s Hospital® West Campus and Texas Children’s Hospital® The Woodlands, bring specialty pediatric care – including an additional 160 licensed acute care and critical care beds, two 24/7 pediatric emergency centers, full-service surgical suites and more than 20 subspecialty clinics a piece – to a rapidly expanding population of children across the west and north of Houston.

Texas Children’s also operates Texas Children’s® Health Plan, the nation’s first health maintenance organization created just for children, and Texas Children’s® Pediatrics, the nation’s largest pediatric primary care network, with over 50 locations across Houston. Texas Children’s® Specialty Care and Texas Children’s® Urgent Care locations provide additional enhanced access to care throughout the Greater Houston community.

In 2018, we expanded our care even further, all the way to the capital city of Austin. The first Texas Children’s Urgent Care opened in March 2018 to bring convenient, affordable, high-quality after-hours pediatric care to Austin area families. Additionally, three Texas Children’s Pediatrics practices and a Texas Children’s Specialty Care location staffed by Texas Children’s board-certified physicians, therapists and clinical staff, opened in Austin in 2018.
Texas Children’s Heart Center is committed to setting a new standard for the treatment of children and adults with congenital heart disease and to pioneering innovative therapies for patients with cardiac conditions.
ADULT CONGENITAL HEART DISEASE

The Adult Congenital Heart Disease (ACHD) program at Texas Children’s Hospital, the largest ACHD program in Texas, allows patients with congenital heart disease to receive seamless continuity of care from birth throughout adulthood. As pediatric patients with congenital heart defects transition into adulthood, our multidisciplinary team of experienced congenital heart disease specialists advises them on health and lifestyle choices for their adult needs, including physical challenges, exercise options and family planning.

Our program offers comprehensive medical and surgical care backed by state-of-the-art technologies and research from our world-class institutions. We provide the full range of ACHD services and specialties including surgery, diagnostic and interventional cardiac catheterization, noninvasive imaging (including cardiac MRIs and coronary CT angiography), arrhythmia services and more.

The ACHD program is accredited by the Adult Congenital Heart Association (ACHA) and is one of only three programs accredited in Texas.

To achieve this distinction, Texas Children’s had to meet ACHA’s criteria for medical/surgical services and personnel requirements and go through a rigorous accreditation process developed over a number of years through a collaboration with doctors, physician assistants, nurse practitioners, nurses and adult congenital heart disease patients. We are also one of only 21 programs in the country with an Accreditation Council for Graduate Medical Education ACHD training program and the only program in Texas that offers this training opportunity.
ACHD CLINIC ENCOUNTERS
Volume by year

CATHETERIZATION CASES
Patient age >18
The Cardiac Developmental Outcomes program (CDOP) provides routine longitudinal neurodevelopmental follow up by board-certified developmental-behavioral pediatricians, neurodevelopmental disability pediatricians and child psychologists for all children who undergo surgery for congenital heart disease during the first three months of life. The CDOP team also provides longitudinal monitoring for children and adolescents with congenital heart disease and neurodevelopmental concerns who are referred by pediatric cardiologists and other providers. Our program is the most mature and comprehensive developmental outcomes program in Texas, and we see not only Texas Children’s Hospital patients, but children from other programs across the state.

Over the past year, our clinic has continued to grow, as has our clinic team and the number of clinic days. We welcomed our new research coordinator who has worked with the clinicians to further develop our state-of-the-art database. In addition to providing an outstanding clinical service, we are participating in national quality initiatives and increasing our research platform with multiple current and upcoming projects. These projects will evaluate outcomes for our patients as well as interventions taking place and resources being provided as part of their routine care within the CDOP clinic.
CARDIAC IMAGING

Our noninvasive imaging lab performs more than 31,000 echocardiograms annually for fetuses, infants, children and adults with congenital or acquired heart disease. We provide transesophageal, epicardial, intracardiac and transthoracic echocardiographic support in the catheterization labs, operating rooms and intensive care units on a daily basis. Our imaging team now consists of 38 highly trained sonographers and more than 35 faculty members, a dedicated advanced practice provider for sedation, as well as nurses, medical assistants and clerical staff.

In September 2018, our team moved into its new home in the Lester and Sue Smith Legacy Tower. We now have 15 transthoracic echo imaging rooms with a six-bed sedation bay. In the new building, we created unique pods where echo imaging rooms are nested adjacent to clinic rooms to promote communication between imaging and clinical teams and improve our patient/family experience. State-of-the-art multiphysician “Mission Control” reading rooms are embedded between our pods, giving echo faculty the ability to watch live imaging from within the echo lab and in patient care units throughout the hospital.

Echocardiographic services are also provided at all of the Texas Children’s Specialty Care locations and we have additional imaging centers at Texas Children’s Hospital West Campus and Texas Children’s Hospital The Woodlands to support our services in the community.

ECHOCARDIOGRAMS
Volume by location

- Texas Children’s Hospital
- Texas Children’s Hospital West Campus
- Texas Children’s Hospital The Woodlands
- Texas Children’s Specialty Care locations
- Community hospitals
**Cardiac MRI & CT**

The diagnostic imaging capabilities of cardiac MRI and CT provide an excellent complement to the primary imaging modality, echocardiography. With advances in technology that allow rapid imaging with minimal radiation, cardiac CT has become vital in the diagnosis and surgical planning for many of our patients with congenital heart disease. For neonates with complex congenital heart disease, we use cardiac CT to provide rapid and excellent diagnostic information, typically without the need for sedation. For our single ventricle patients, we often employ cardiac CT prior to each surgical endeavor to supply information about the extracardiac anatomy to our surgeons.

Our cardiac MRI program continues to grow and is currently one of the largest in North America. This modality provides a wealth of information in multiple areas, including anatomy, morphology, volumetric and functional data, as well as physiologic information regarding blood flow. This imaging modality is frequently used in our teenage and adult congenital populations because of the valuable information it provides and the limitations of transthoracic echocardiography among this population.

In late 2018, we opened our new hybrid cardiac MRI/interventional cardiac catheterization suite. This addition allows us to meld these two procedures together to combine the strengths of both modalities, decrease the use of radiation, and eventually perform MRI-guided cardiac catheterizations.
Pharmacologic Cardiac Stress MRI
At Texas Children’s Hospital, we are proud to have one of the busiest pediatric stress MRI programs in the world. There are numerous pediatric cardiac diseases that may be a cause of myocardial ischemia. Cardiac MRI is an excellent diagnostic tool to examine the cardiac function, perfusion and viability in these unique populations. As part of our robust Coronary Artery Anomalies program, the majority of patients undergo a dobutamine stress MRI as part of their diagnostic work-up. For patients at risk of coronary ischemia, including those with an arterial switch operation or a diagnosis of Kawasaki disease, we are one of only a few pediatric centers in the U.S. to offer regadenoson as a pharmacologic stress agent. These tests require a dedicated, coordinated effort from a team of experts including a pediatric cardiologist, pediatric radiologist, cardiac nurse, cardiac MRI technologist and MRI physicist. In 2018, our program performed 113 cardiac stress MRI examinations.

Advances in Cardiac Imaging
• Hypoplastic left heart syndrome (HLHS) is a complex cyanotic heart defect requiring three surgical palliations in the first three years of life. To better assess for changes in function of the single right ventricle in these patients over time, we instituted a prospective imaging protocol over a more than two year period using cardiac strain technology, which assesses deformation of the cardiac muscle. The advantage of strain is that it is a quantitative measure with high reproducibility. The outcomes of this work have been accepted for publication in an upcoming issue of the Journal of the American Society of Echocardiography. We found that strain indices in patients between the Norwood and Glenn stages are worse in HLHS infants who later have a poor cardiac outcome. This may help us better identify at-risk patients and has the potential to change practice as we incorporate this modality into the care of these patients.

• With industry support, we are currently evaluating the clinical utility of blood speckle imaging (BSI). Blood cells traveling through a heart chamber or valve produce flow variations such as vortices and helix formations. We are looking at ascertaining normal and abnormal intracardiac flow patterns without using injected contrast or invasive methods. We are also now collecting simultaneous 4D data evaluation by MRI to assist in the evaluation of flow through the cardiac valves.

• The impact of chemotherapy on ventricular function in children is being increasingly recognized. As part of our Cardio-Oncology initiative, we have adopted strict standards for detailed ventricular functional analysis and included myocardial strain. Our goals are to improve accuracy and consistency of functional assessment, detect earlier identification of cancer therapy-related cardiac dysfunction and determine best practices for coordinating care with our Hematology-Oncology colleagues. Upon recent early review of this initiative, we found we have been successful in decreasing variability in assessing cardiac function through these changes to our operating procedure.

• Evaluating diastolic dysfunction (abnormalities in relaxation of the heart) in pediatric patients is a challenge. We are investigating left atrial strain analysis as a potentially important tool to assess for early diastolic abnormalities in patients who have had cardiac transplant and in those treated with anthracyclines as part of their cancer therapy. Both projects are currently under consideration for publication.

• 3D evaluation of cardiac anatomy and function is performed to provide more complete imaging details essential in preoperative or precatheterization intervention planning, and to assist in functional assessment over time.
CARDIAC INTENSIVE CARE UNIT

The Cardiac Intensive Care Unit (CICU) at Texas Children’s Hospital strives for continued excellence in the care of infants, children and adults with complex cardiac disease. The patient- and family-focused multidisciplinary care delivery model is supported by some of the best specialists and care providers in the nation, ensuring the highest level of clinical support for some of the sickest children in our region and beyond. In September 2018, together with the entire Heart Center, the CICU moved to our new home – the Lester and Sue Smith Legacy Tower – which houses 48 state-of-the-art CICU rooms, all with dedicated family space. The unit spans three floors, in direct adjacency or vertical proximity with the cardiovascular operating rooms, catheterization laboratory, MRI scanners, interventional radiology suites and all other support services. The new CICU is also collocated with the 84-bed Pediatric Intensive Care Unit.

CICU PATIENT ADMISSIONS

The CICU Team

The CICU is staffed by a highly experienced, multidisciplinary team of physicians, advanced practice providers, nurses, respiratory therapists and pharmacists, as well as a nutrition team, physical therapists and occupational therapists. All members of the team and the patient families are included in multidisciplinary rounding with shared decision-making. We are actively involved in numerous collaborative clinical, quality and research initiatives. Our growing group of dedicated CICU advanced practice providers, together with more than 40 ACGME fellows in cardiology and critical care medicine, provide patient care under the supervision of our 33 cardiac intensivists with subspecialty training in pediatric critical care, cardiology, neonatology and cardiac anesthesia. The Cardiac Critical Care Instructorship program is nationally recognized for innovations and leadership in training the next generation of pediatric cardiac intensivists. Specialized complex care patient teams monitor patient progress, developing plans for interventions and optimizing supportive therapies to achieve the best outcomes in some of the most complex patient populations, such as patients with hypoplastic heart syndrome. A dedicated Heart Center Rapid Response Team is staffed by intensive care attendings, fellows and nurses, ensuring timely escalation of care and transport of patients to a higher level of care.
Subspecialty Cardiac Intensive Care
Our new environment includes increased capacity and four pods with 12 beds each, allowing us to further expand our subspecialty focus for critically ill children with heart disease. We also have a 12-bed dedicated Heart Failure Intensive Care Unit, the first of its kind in the nation, which focuses on the treatment of children with heart failure and those requiring intensive care before and after heart transplant including both durable and temporary mechanical circulatory support strategies. In addition, we have a new neonatal CICU to which we admit newborns before and after surgery and are able to focus on their developmental care while attending to the specific needs of their families.

Quality and Outcomes
The CICU’s outcomes and performance are closely monitored through participation in national registries including Pediatric Critical Care Consortium (PC4), Extracorporeal Life Support Organization (ELSO), Society of Thoracic Surgery (STS) and PediMACS. Our Heart Failure ICU is an active participant in the Advanced Cardiac Therapies Improving Outcomes Network (ACTION) collaborative focusing on improving outcomes for pediatric ventricular assist device patients. Patient safety and quality are approached by our quality team, with regular reviews of all of our practices and procedures. Internal audit routinely includes benchmarking our performance alongside other high volume U.S. centers.

One of our recent focuses has been on reducing the incidence of catheter-associated bloodstream infections. Our multidisciplinary team of critical care clinical nurse specialists, intensive care physicians, quality specialists and the Texas Children’s Infection Control team have developed a central line stewardship team with 40 team members. Through this, we introduced multiple initiatives including new care bundles, rapid multidisciplinary reviews of any suspected hospital-acquired infection, nurse-led rounding tools and dress codes in all ICU areas.

CICU COMPLICATIONS

- Unplanned Extubations: 2.59 (Texas Children’s Hospital), 2.86 (High volume centers)
- Cardiac Arrest after Surgery: 2.07 (Texas Children’s Hospital), 2.89 (High volume centers)
- Cardiac Arrest in Medical Admissions: 1.69 (Texas Children’s Hospital), 2.45 (High volume centers)
- Necrotizing Enterocolitis in Surgical Patients: 0.48 (Texas Children’s Hospital), 1.08 (High volume centers)

Data from PC4 registry
Extracorporeal Cardiopulmonary Resuscitation

Extracorporeal cardiopulmonary resuscitation (ECPR) refers to the initiation of lifesaving extracorporeal membrane oxygenation (ECMO) in the setting of cardiac arrest. Successful ECPR outcomes require rapid decision-making and careful team training to ensure that patients receive ECMO as quickly as possible. Texas Children’s Heart Center is committed to working towards more rapid initiation of the support with an improved call-out and response system, and enhancement of the availability of key personnel around the clock.

In early 2018, the Heart Center and Perioperative teams embarked upon a major multidisciplinary initiative to develop a robust, unified response to minimize the time taken to initiate ECMO. One important step was to introduce round-the-clock in-house ECMO circuit primers – highly trained experts in the setup of the complex ECMO circuits – helping us to avoid any delays in readiness to initiate ECMO. In addition, we introduced a unified call-out mechanism that placed the responsibility of decision-making on the CICU medical and nursing team with simultaneous notification of all key team members. Finally, between January and March 2018, we conducted a series of high-fidelity simulation exercises to test the system and train the team.

International Collaborations

Our CICU team has established strong links with cardiac teams in several countries within Latin America. Through videoconferences with our colleagues in Mexico City, we have established regular virtual ward rounds and additional communication forums, rounding with the local cardiac surgeons and intensivists in their two main cardiac centers. In 2018, we held more than 70 video-conference ward rounds in the two CICUs in Mexico City, assisting with patient management, sharing of protocols, establishing new or innovative therapies including ECMO, and recommending quality improvement initiatives. In addition, together with many Heart Center colleagues, our critical care team has active educational and simulation-based collaborations with several centers in Latin America (Mexico, Chile, Colombia, Argentina and Costa Rica) aimed at developing advanced perioperative cardiac care and to establish and maintain ECMO programs.
We are committed to helping our patients with congenital heart disease (CHD) who are approaching adulthood make a smooth transition from pediatric to adult health care. We have developed a unique transition program that serves adolescents with CHD by performing individualized evaluations that reveal individual patient needs surrounding CHD knowledge and disease management skills. Our program works with cardiologists to teach adolescents to better understand their specific CHD and practice the skills needed to manage their CHD in an adult setting, in order to facilitate transfer to adult care by 21 years of age. Our program additionally helps patients meet an adult congenital cardiologist, facilitates communication about patients between the pediatric and adult cardiologists, and creates medical summaries for these adult providers so that they may understand the unique needs of our patients.

Serial educational sessions and assessments are conducted to ensure appropriate knowledge and skill base prior to transferring to adult care. The assessments focus on promoting independence in the following areas:

- **Education**: CHD knowledge for lifelong care
- **Skill building**: Tools needed to become a successful adult with a CHD
- **Enhancing connections**: Providing an environment to foster social networks for patients and advocates with CHD
- **Creating a portfolio**: Including a medical summary and lists of financial, psychosocial and adult CHD resources
- **Community building**: Building a community of young adults with CHD

**WHAT PATIENTS CAN EXPECT**

- **14 years**: Meet the cardiology transition team and learn about the process and opportunities.
- **15/16 years**: Serial educational visits, targeted resources, encourage shared decision making.
- **17 years**: Take ownership of your health care and identify lifelong care plan.
- **18-21 years**: Mastery of disease knowledge, health care skills and transfer to adult care.

**WHAT PARENTS CAN EXPECT**

- **14 years**: Meet the team, discuss individual learning plan and transition expectations.
- **15/16 years**: Do transition readiness assessments, individualized CHD education and health care skills training.
- **17 years**: Transition to a supportive role and prepare for child’s health care changes at 18 years old.
- **18-21 years**: Adult planning visit and prepare for transfer of care to adult specialist.
CARDIOVASCULAR ANESTHESIOLOGY

The Cardiovascular Anesthesiology program at Texas Children’s Hospital, the largest of its kind in the country, provides sedation and anesthesia services for all congenital cardiac operations and for children with congenital heart disease undergoing various non-cardiac procedures.

Our team of more than 15 board-certified pediatric anesthesiologists and 10 nurse anesthetists also provides anesthesia services for catheterization and electrophysiology procedures as well as imaging studies in MRI, CT and interventional radiology.

CARDIOVASCULAR ANESTHESIOLOGY CASES

Volume by year

<table>
<thead>
<tr>
<th>Year</th>
<th>Catheterizations and electrophysiology patients</th>
<th>Cardiovascular operating room patients</th>
<th>Interventional radiology patients</th>
<th>MRI patients</th>
</tr>
</thead>
<tbody>
<tr>
<td>2014</td>
<td>1,094</td>
<td>1,002</td>
<td>252</td>
<td>152</td>
</tr>
<tr>
<td>2015</td>
<td>1,129</td>
<td>1,129</td>
<td>264</td>
<td>252</td>
</tr>
<tr>
<td>2016</td>
<td>1,226</td>
<td>1,051</td>
<td>264</td>
<td>428</td>
</tr>
<tr>
<td>2017</td>
<td>1,177</td>
<td>1,017</td>
<td>394</td>
<td>340</td>
</tr>
<tr>
<td>2018</td>
<td>1,331</td>
<td>1,085</td>
<td>359</td>
<td>436</td>
</tr>
</tbody>
</table>

Catheterizations and electrophysiology patients
Cardiovascular operating room patients
Interventional radiology patients
MRI patients
Cerebral Autoregulation

The Cardiovascular Anesthesiology team at Texas Children’s Hospital, in collaboration with the Cardiac Intensive Care Unit, is active in the study of risk factors and possible interventions to avoid or decrease the potential changes in cerebral autoregulation and the impact this may have on neurologic outcomes. Using standard cerebral oxygen saturation (rScO2 by NIRS) and invasive blood pressure monitoring, algorithms are developed to identify periods of change in cerebral autoregulation.

Our team has demonstrated the importance of blood pressure control during and immediately following cardiopulmonary bypass in children undergoing the bidirectional cavo-pulmonary shunt repair to preserve cerebral autoregulation and thus decrease the risk of neurologic injury in the perioperative period.

Hypertension is associated with increased hemoglobin volume index (HVx) signifying cerebral dysautoregulation.\(^1\)

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Dexmedetomidine Bolus and Infusion in Corrective Infant Cardiac Surgery

Led by principal investigator Dean Andropoulos, M.D., and working within the National Institutes of Health-funded Pediatric Heart Network, investigators in the Division of Pediatric Cardiovascular Anesthesiology published a landmark study\(^2\): "Results of a Phase I Multicentre Investigation of Dexmedetomidine Bolus and Infusion in Corrective Infant Cardiac Surgery." This research established a robust pharmacokinetic model for dosing of this very important and increasingly utilized drug and recommended safe dexmedetomidine dosing from the start of anesthesia, and during cardiopulmonary bypass, for the first time.

The study also established that dexmedetomidine, when used in correct doses for neonates and young infants, is a safe drug with very low incidence and severity of side effects. The study was published in the British Journal of Anaesthesia, and was accompanied by an editorial describing the importance of the research and recognizing the excellence of the study design, which included validation of the pharmacokinetic model with a separate study. Among the major findings are that neonates require a dexmedetomidine dose 50 percent less than older infants, and that metabolism of dexmedetomidine on cardiopulmonary bypass decreases by greater than 90 percent, necessitating significant dose reduction. Dosing recommendations by patient age and desired plasma level are displayed in the table below.

### DOSING RECOMMENDATIONS FOR DEXMEDETOMIDINE

#### STEADY STATE CONCENTRATIONS

<table>
<thead>
<tr>
<th>Age Group (days)</th>
<th>Target Css (pg/ml)</th>
<th>Initial Loading Dose (mcg/kg)</th>
<th>Infusion 1: pre-CPB, first 60 minutes of CPB (mcg/kg/hour)</th>
<th>Loading Dose to CPB Prime Volume (mcg/ml)</th>
<th>Infusion 2: after 60 minutes of CPB until end of CPB (mcg/kg/hour)</th>
<th>Infusion 3: 60 minutes after CPB (mcg/kg/hour)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neonatal (0-21)</td>
<td>200</td>
<td>0.24</td>
<td>0.22</td>
<td>0.004</td>
<td>0.04</td>
<td>0.14</td>
</tr>
<tr>
<td>Neonatal (0-21)</td>
<td>500</td>
<td>0.6</td>
<td>0.55</td>
<td>0.01</td>
<td>0.1</td>
<td>0.35</td>
</tr>
<tr>
<td>Neonatal (0-21)</td>
<td>700</td>
<td>0.84</td>
<td>0.77</td>
<td>0.014</td>
<td>0.14</td>
<td>0.49</td>
</tr>
<tr>
<td>Neonatal (0-21)</td>
<td>1000</td>
<td>1.2</td>
<td>1.1</td>
<td>0.02</td>
<td>0.2</td>
<td>0.7</td>
</tr>
<tr>
<td>Infant (22-180)</td>
<td>200</td>
<td>0.29</td>
<td>0.26</td>
<td>0.005</td>
<td>0.05</td>
<td>0.17</td>
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<tr>
<td>Infant (22-180)</td>
<td>500</td>
<td>0.72</td>
<td>0.66</td>
<td>0.012</td>
<td>0.12</td>
<td>0.42</td>
</tr>
<tr>
<td>Infant (22-180)</td>
<td>700</td>
<td>1.01</td>
<td>0.92</td>
<td>0.017</td>
<td>0.17</td>
<td>0.59</td>
</tr>
<tr>
<td>Infant (22-180)</td>
<td>1000</td>
<td>1.44</td>
<td>1.32</td>
<td>0.024</td>
<td>0.24</td>
<td>0.84</td>
</tr>
</tbody>
</table>

Erector Spinae Block
Thoracotomy for pediatric cardiac surgery can be associated with significant pain. Regional anesthesia techniques such as epidurals and paravertebral blocks can reduce the pain of thoracotomy but can also be associated with significant risk due to the proximity of such techniques to the neuraxial (epidural) and pleural (paravertebral) spaces.

The erector spinae plane (ESP) block, first described in 2016, targets the dorsal and ventral rami of spinal nerve roots and has been used extensively for a host of adult surgical procedures including chest wall and abdominal surgeries.3-4 The ESP block is a less invasive option due to the avoidance of the neuraxial and paravertebral spaces. The Cardiovascular Anesthesiology team at Texas Children’s Hospital has initiated a program to utilize regional anesthetic techniques, including the ESP block for the management of postoperative pain in pediatric cardiac patients. The ESP block was used successfully in several children undergoing thoracotomy for cardiac surgical repairs (Table 1). The blocks were performed after induction of general anesthesia and catheters were placed following the initial block. The patients were followed by the pain service postoperatively. Intraoperative and postoperative opioid use was decreased compared to our usual doses at our institution, and early mobilization and feeding were achieved successfully.

Table 1

<table>
<thead>
<tr>
<th>Age, Gender, Wt (kg)</th>
<th>Surgery</th>
<th>Ropivacaine Dose</th>
<th>Post Operative FLACC Scores</th>
<th>Post OP Pain Meds in First 24 Hours</th>
<th>Opiod Usage in First 24 Hours (Morphine Equivalent)</th>
<th>Discharge (POD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>A 4 year old male, 19.7 kg</td>
<td>Coarctation Repair via Left Thoracotomy</td>
<td>0.2%, 15 ml bolus, 0.2% at 5 ml/h</td>
<td>0-4</td>
<td>Acetaminophen scheduled Morphine pm Diazepam pm</td>
<td>1 mg (0.05 mg/kg)</td>
<td>3</td>
</tr>
<tr>
<td>B 4 year old male, 16.1 kg</td>
<td>Vascular Ring Division via Left Thoracotomy</td>
<td>0.5%, 8 ml bolus, 0.1% at 4 ml/h</td>
<td>0-4</td>
<td>Acetaminophen scheduled Ketorolac scheduled Morphine pm Diazepam pm</td>
<td>3.6 mg (0.225 mg/kg)</td>
<td>5 (Chylothorax)</td>
</tr>
<tr>
<td>C 9 year old female, 30.4 kg</td>
<td>Vascular Ring Division via Left Thoracotomy</td>
<td>0.5%, 12 ml bolus, 0.1% at 6-8 ml/h</td>
<td>0-3</td>
<td>Acetaminophen scheduled Morphine pm Diazepam pm</td>
<td>14.4 mg (0.5 mg/kg)</td>
<td>3</td>
</tr>
<tr>
<td>D 5 year old female, 20.5 kg</td>
<td>Vascular Ring Division via Left Thoracotomy</td>
<td>0.2%, 15 ml bolus, 0.2% at 4 ml/h</td>
<td>0</td>
<td>Acetaminophen scheduled Morphine pm Diazepam pm</td>
<td>3 mg (0.15 mg/kg)</td>
<td>16 (Chylothorax)</td>
</tr>
<tr>
<td>E 6 year old female, 21.9 kg</td>
<td>Coarctation Repair via Left Thoracotomy</td>
<td>0.2%, 20 ml bolus, 0.2% at 6 ml/h</td>
<td>2-7</td>
<td>Acetaminophen scheduled Morphine pm Diazepam pm</td>
<td>6.4 mg (0.3 mg/kg)</td>
<td>5</td>
</tr>
</tbody>
</table>


The erector spinae plane block can potentially lead to decreased intraoperative and postoperative opioid use for pediatric patients undergoing thoracotomy for cardiac surgery and thus lead to a decrease in the side effects associated with opioids. The erector spinae plane block is also less technically challenging than an epidural or paravertebral block and is potentially safer as it avoids the neuraxial and pleural spaces (Figure 1). We hope to continue to use this relatively new block to provide safer and better pain control to our pediatric cardiac patients.

Figure 1
**Intubation Route**

Children presenting for cardiac surgery require endotracheal intubation to provide oxygen and anesthetic gases and remove carbon dioxide throughout the procedure and afterwards into the ICU. Depending on the age of the patient and the type of procedure, the endotracheal tube may either be placed orally or nasally into the trachea. There are advantages and disadvantages to both routes and there is not a consistent practice throughout the United States and elsewhere. The Congenital Cardiac Anesthesia Society recently undertook a national study in conjunction with the Society of Thoracic Surgeons looking at oral versus nasal intubation to determine if there were risks and benefits to each route and whether the age of the patient made a difference.\(^5\)

This study found that patients less than one year of age appeared to benefit more from nasal intubation while those older than one year of age had benefits from oral intubation. These relative benefits and risks are likely related to the development of the sinus cavities around the nose that begin to form in the first year of life, as the sinus cavities may serve as a source for bacteria that may subsequently impact surgical infections. The younger patients may benefit from the increased stability of a nasal tube during imaging in the operating room with transesophageal echocardiography (TEE) as well as postoperatively in the ICU. Our practice is consistent with these findings as almost all newborns and infants are nasally intubated while toddlers and older children are typically orally intubated unless specific circumstances warrant placing it in one place or the other due to anatomic problems, prolonged intubations or family preference based on prior experience.

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CONGENITAL HEART SURGERY

The Congenital Heart Surgery team provides individualized and comprehensive surgical care for all aspects of pediatric and adult congenital heart disease. With a team of six surgeons, we are experienced in the rarest and most common cases and perform more than 900 surgical procedures annually, with outcomes among the best in the country. We treat patients of all ages, from preterm and low-birth-weight newborns to adults with congenital heart disease, and we personalize treatments and procedures to best suit the situation of each patient and family. This tailored approach includes cardiopulmonary bypass and neuroprotection strategies focused on the patient’s condition and needs, helping to achieve optimal functional outcomes. The Heart Center’s Heart and Heart-Lung Transplant programs are integrated with the ventricular assist device (VAD) program and together comprise one of the largest and most successful pediatric programs in the nation. Our lung transplant program is also one of the largest in the country.

Texas Children’s Welcomes Two Esteemed Congenital Heart Surgeons

Two congenital heart surgeons joined the Texas Children’s Heart Center team last fall – Dr. Christopher Caldarone and Dr. E. Dean McKenzie. Caldarone is an internationally recognized leader in congenital heart surgery and most recently served as surgeon-in-chief at The Hospital for Sick Children in Toronto. He joined Texas Children’s in September as the chief of congenital heart surgery and professor of congenital heart surgery at Baylor College of Medicine. Caldarone received his undergraduate degree from Johns Hopkins University and his medical degree from Columbia University. He completed his general surgery and cardiothoracic surgery residencies at Beth Israel Deaconess Medical Center/Harvard Medical School. He also completed a fellowship in congenital heart surgery at The Hospital for Sick Children.

McKenzie, who joined Texas Children’s in October, is a world-renowned leader in congenital heart surgery. He was previously a member of Texas Children’s Heart Center team for more than 15 years. Most recently, he served as chief of pediatric congenital cardiothoracic surgery at Children’s Healthcare of Atlanta, and professor and chair of the division of cardiothoracic surgery at Emory University School of Medicine. McKenzie received his undergraduate degree from The University of Texas at Austin and his medical degree from Baylor. He completed his residency in general surgery at the University of Louisville and his residency in cardiovascular and thoracic surgery at The University of Florida College of Medicine.

CARDIOVASCULAR AND THORACIC SURGERY CASES

Volume by year
CARDIOVASCULAR AND THORACIC SURGERY CASES
Percent by age

TOTAL CARDIOPULMONARY BYPASS OPERATIONS
Volume by year
### MORTALITIES BY STAT CLASSIFICATION

**in 2018**

<table>
<thead>
<tr>
<th>Primary Procedure</th>
<th>Number of Procedures</th>
<th>Number of Discharge Mortalities</th>
<th>Percent Mortality at Texas Children’s Hospital</th>
<th>STS National Benchmark&lt;sup&gt;6&lt;/sup&gt;</th>
</tr>
</thead>
<tbody>
<tr>
<td>STAT 1</td>
<td>183</td>
<td>0</td>
<td>0%</td>
<td>0.4%</td>
</tr>
<tr>
<td>STAT 2</td>
<td>197</td>
<td>2</td>
<td>1%</td>
<td>1.3%</td>
</tr>
<tr>
<td>STAT 3</td>
<td>83</td>
<td>0</td>
<td>0%</td>
<td>2.1%</td>
</tr>
<tr>
<td>STAT 4</td>
<td>157</td>
<td>4</td>
<td>2.6%</td>
<td>6.2%</td>
</tr>
<tr>
<td>STAT 5</td>
<td>33</td>
<td>3</td>
<td>9.1%</td>
<td>11.9%</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>653</strong></td>
<td><strong>9</strong></td>
<td><strong>1.4%</strong></td>
<td><strong>2.7%</strong></td>
</tr>
</tbody>
</table>


### MORTALITIES BY AGE AND OPERATION TYPE

**in 2018**

<table>
<thead>
<tr>
<th>Age</th>
<th>CPB Cases</th>
<th>Non-CPB Cases</th>
<th>CPB Discharge Mortalities</th>
<th>Non-CPB Discharge Mortalities</th>
<th>Discharge Mortality</th>
<th>Percent Mortality at Texas Children’s Hospital</th>
<th>STS National Benchmark&lt;sup&gt;7&lt;/sup&gt;</th>
</tr>
</thead>
<tbody>
<tr>
<td>Neonate (0d - 30d)</td>
<td>84</td>
<td>28</td>
<td>4</td>
<td>2</td>
<td>6/112</td>
<td>5.4%</td>
<td>7.4%</td>
</tr>
<tr>
<td>Infant (31d - 1y)</td>
<td>185</td>
<td>31</td>
<td>1</td>
<td>1</td>
<td>2/216</td>
<td>0.9%</td>
<td>2.6%</td>
</tr>
<tr>
<td>Child (&gt;1y - &lt;18y)</td>
<td>264</td>
<td>27</td>
<td>0</td>
<td>1</td>
<td>1/291</td>
<td>0.3%</td>
<td>1.1%</td>
</tr>
<tr>
<td>Adult (18y+)</td>
<td>32</td>
<td>2</td>
<td>0</td>
<td>0</td>
<td>0/34</td>
<td>0%</td>
<td>1.3%</td>
</tr>
<tr>
<td><strong>Total</strong></td>
<td><strong>565</strong></td>
<td><strong>88</strong></td>
<td><strong>5</strong></td>
<td><strong>4</strong></td>
<td><strong>9/653</strong></td>
<td><strong>1.4%</strong></td>
<td><strong>2.7%</strong></td>
</tr>
</tbody>
</table>

**ARTERIAL SWITCH OPERATION**

*Volume by year*

Overall hospital discharge mortality rate*: 0%
STS national benchmark*: 4.2%

---

**ATRIAL SEPTAL DEFECT REPAIR**

*Volume by year*

Overall hospital discharge mortality rate*: 0%
STS national benchmark*: <1%

---

**ATRIOVENTRICULAR CANAL REPAIR**

*Volume by year*

Overall hospital discharge mortality rate*: 0%
STS national benchmark*: 1.9%

---

* Hospital mortality is calculated over the last four years from 2015-2018.
* Source for STS national benchmark is the Society of Thoracic Surgeons Data Harvest Report January 2014 to December 2017.
NORWOOD OPERATION

Volume by year

Overall hospital discharge mortality rate\(^8\): 8.8%  STS national benchmark\(^9\): 13.9%

TETRALOGY OF FALLOT REPAIR

Volume by year

Overall hospital discharge mortality rate\(^8\): 0.8%  STS national benchmark\(^9\): 1.8%

VENTRICULAR SEPTAL DEFECT REPAIR

Volume by year

Overall hospital discharge mortality rate\(^8\): 0%  STS national benchmark\(^9\): <1%
Since 2012, Texas Children’s Coronary Artery Anomalies program has provided multidisciplinary care for patients with rare congenital heart defects associated with coronary ischemia, myocardial infarction and sudden death. Coronary artery anomalies are the second-leading cause of sudden cardiac death in children and young adults. Diagnosing these anomalies can be challenging because many individuals with the conditions have no symptoms, and their first manifestation can be sudden cardiac arrest or death. Those who do have symptoms complain most often of chest pain, palpitations, dizziness or fainting during or just after exercise.

How to best treat children and young adults with coronary artery anomalies is a subject of debate in the medical community. Most physicians agree that surgery is necessary for patients who show evidence of decreased blood flow to the heart muscle, but how to treat patients who have no physical complaints and show no evidence of reduced blood flow to the heart is unclear.

Symposium on Coronary Artery Anomalies
In December 2018, experts from across the country convened for the fourth Symposium on Coronary Artery Anomalies, hosted by Texas Children’s Hospital and Children’s Hospital of Philadelphia.

The event was founded by leaders of Texas Children’s Coronary Anomalies program – the first of its kind in the nation – as a way for providers, researchers and others to gather and discuss the diagnosis and management of patients with coronary artery anomalies.

The symposium featured comprehensive presentations by members of Texas Children’s Coronary Anomalies Program, as well as 11 visiting faculty from some of the country’s preeminent pediatric health care institutions, including Boston Children’s, CHOP, Stanford University, Columbia University, UT Southwestern, and the Heart & Vascular Institute at Hartford Hospital in Connecticut. Major topics included AAOCA, Kawasaki disease and myocardial bridges.

An addition to this year’s conference was a special Patients and Families Symposium. During this day-long event, parents and children attended talks that addressed topics such as the impact of coronary anomalies on families and the importance of counseling and shared decision-making when determining a plan of care. Visitors had an opportunity to hear from Texas Children’s patients and families who have had similar experiences of unknowingly living with these life-threatening conditions, receiving crucial diagnoses, making tough decisions, undergoing open-heart surgery and living a normal life in the aftermath. This emotional and powerful session was followed by discussions about the importance of networking and building a sense of community and collective support for people affected by coronary anomalies.
The Electrophysiology program at Texas Children’s Hospital offers cutting-edge treatment for patients with heart rhythm abnormalities. Most of these abnormalities cause the heart to beat either too fast or too slow and can be dangerous if they cause a significant decrease in the heart’s ability to pump blood. There are many causes of arrhythmias including congenital heart disease, heart muscle disease, genetic disorders and others.

At Texas Children’s Heart Center, we offer a broad complement of diagnostic and therapeutic options to evaluate and manage heart rhythm abnormalities. We are one of the highest volume pediatric centers in the country for invasive electrophysiology studies and pacemaker/defibrillator implantations, and we maintain success rates for ablations that exceed the national average. Additionally, Texas Children’s Heart Center is one of the leading centers in the world for the diagnosis and treatment of arrhythmias in children and adults with congenital heart disease.

**Treatment Of Arrhythmias and Sudden Death in Children**

Electrophysiology has developed a multipronged approach to tackle and study arrhythmias and sudden death in children in the Houston community. First, educating the public and potential responders regarding arrhythmias and sudden cardiac arrest is paramount. As such, we have embarked on an ambitious campaign to educate and prepare the community and schools in the Houston area via our Project ADAM affiliate. The campaign’s goal is to certify all schools as “Heart Safe.” This past year, with the help of a grant from the American Heart Association (AHA), we were able to double the number of Houston schools that are certified as Heart Safe and provided schools with new Automated External Defibrillators.

Our team is also at the forefront of ongoing research to gain insight into the molecular mechanisms of these arrhythmias and develop new diagnostic approaches and therapies. Using novel technologies, we are able to convert blood samples from patients into their own stem cells and subsequently into their own cardiac cells. When coupled with experimental platforms that allow for measurement of electrical changes and ion flow, we can explore the molecular causes of sudden death-predisposing arrhythmias in an unprecedented fashion. This also allows us to test pharmacologic agents to treat their arrhythmias on what is effectively their own individualized heart tissue. This may open the door for truly individualized, precision medicine that is custom tailored to these children.

**Stereotaxis**

As one of only a few pediatric hospitals in the U.S. with a remote magnetic navigation system for catheter manipulation, we have used Stereotaxis on over 250 ablation cases in recent years with excellent results. Magnetic navigation offers several theoretic advantages in the treatment of children. The catheter is more flexible and atraumatic than standard catheters, potentially eliminating the risk of heart perforation. It can also be navigated with more precision, allowing for movements as small as one millimeter or deflections as small as one degree. This may result in enhanced accuracy and safety during mapping and ablation of certain arrhythmias in this patient population.
Radiation Reduction
In order to decrease morbidity associated with radiation exposure during ablation cases, the Electrophysiology program has established protocols utilizing nonfluoroscopic mapping techniques to minimize radiation. This has resulted in a nearly 20-fold reduction in fluoroscopy times, with most procedures being performed with minimal radiation (<1 minute) over the past four years.

SUPRAVENTRICULAR TACHYCARDIA ABLATION OUTCOMES ACUTE SUCCESS RATE in 2018

CHANGES IN FLUOROSCOPY TIME FOR ABLATION PROCEDURES

Based on data from the Pediatric Radiofrequency Catheter Ablation Registry.
Awards and Grants

- Three Electrophysiology team members received Baylor College of Medicine’s Fulbright Educational Excellence Award in Leadership and Teaching
- National Institutes of Health K23 Grant for study of the genetic basis of pediatric sudden arrhythmic death syndromes
- Cancer Prevention Grant for study of genetic causes of cardiotoxicity with anthracycline and radiation exposure
- Pediatric Pilot Grant Award for study of Junctophilin type 2 and cardiac nodal dysfunction
- American Heart Association grant to provide AEDs for schools that are certified “Heart Safe”
FETAL CARDIOLOGY

For more than 30 years, Texas Children’s Fetal Cardiology program has provided comprehensive fetal cardiac care to expecting families when there is a concern for or risk of heart disease in the fetus. In partnership with Texas Children’s Fetal Center, this care includes diagnosis, counseling, delivery planning and then transition to treatment at Texas Children’s Heart Center after birth. Our collaborative model allows patients to receive the best care from a team of specialists, all in one place.

We have significant expertise and experience in fetal cardiology given our long history and high volume. We currently have 16 fetal cardiologists highly trained in fetal cardiac imaging. Our vast experience following fetuses through gestation, delivery, infancy and adulthood gives us unique insight and ability to make accurate diagnoses and formulate the best care plans.

We strive to develop new cutting-edge treatments and therapies for fetal cardiac anomalies and have many promising clinical trials and research efforts in progress. Our program is heavily involved with the Fetal Heart Society, which is an international fetal cardiology research organization. We are leading and contributing to multiple fetal studies through this collaboration. We are also members of the International Fetal Cardiac Intervention Registry, which studies techniques and outcomes of fetal cardiac intervention across the world.

FETAL ECHOCARDIOGRAMS

Volume by year

<table>
<thead>
<tr>
<th>Year</th>
<th>Total number of patients</th>
<th>Total number of visits</th>
</tr>
</thead>
<tbody>
<tr>
<td>2014</td>
<td>1,019</td>
<td>1,803</td>
</tr>
<tr>
<td>2015</td>
<td>1,188</td>
<td>1,832</td>
</tr>
<tr>
<td>2016</td>
<td>1,308</td>
<td>2,107</td>
</tr>
<tr>
<td>2017</td>
<td>1,566</td>
<td>2,335</td>
</tr>
<tr>
<td>2018</td>
<td>1,650</td>
<td>2,396</td>
</tr>
</tbody>
</table>

FETAL ECHOCARDIOGRAMS IN 2018

Volume by condition

<table>
<thead>
<tr>
<th>Condition</th>
<th>Total number of patients</th>
<th>Total number of visits</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arrhythmia</td>
<td>46</td>
<td>46</td>
</tr>
<tr>
<td>Atrioventricular Septal Defects</td>
<td>29</td>
<td>29</td>
</tr>
<tr>
<td>Double Outlet</td>
<td>38</td>
<td>38</td>
</tr>
<tr>
<td>Fallot’s Anomaly</td>
<td>59</td>
<td>59</td>
</tr>
<tr>
<td>Ebstein’s Anomaly</td>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>Heterotaxy</td>
<td>21</td>
<td>21</td>
</tr>
<tr>
<td>Hypoplastic Left Heart Syndrome and Related Disorders</td>
<td>17</td>
<td>17</td>
</tr>
<tr>
<td>Horseshoe</td>
<td>20</td>
<td>20</td>
</tr>
<tr>
<td>Pulmonary Ven Anomaly</td>
<td>44</td>
<td>44</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>89</td>
<td>89</td>
</tr>
<tr>
<td>Transposition of Great Arteries</td>
<td>12</td>
<td>12</td>
</tr>
<tr>
<td>Tetralogy of Fallot</td>
<td>63</td>
<td>63</td>
</tr>
<tr>
<td>Transposition of Great Arteries</td>
<td>15</td>
<td>15</td>
</tr>
</tbody>
</table>

Total number of patients: 864 Total number of visits: 1,300
**Fetal Cardiac Interventions**
For fetuses with some cardiovascular conditions, fetal cardiac intervention is available at Texas Children’s Hospital to try to improve the physiology of the heart before birth. Texas Children’s Fetal Center is one of only a few centers in the world capable of providing the full array of fetal cardiac interventions, which involves a coordinated effort among a large, multidisciplinary team of fetal cardiologists, interventional pediatric cardiologists, congenital heart surgeons, maternal fetal medicine experts, maternal and fetal anesthesiologists, and other clinical specialists.

We offer the following fetal cardiac interventions:

**Fetal Ebstein anomaly** – Ebstein anomaly of the fetus is an abnormality of the tricuspid valve than can result in severe tricuspid regurgitation and heart failure in the fetus. For the most severe cases, we currently offer two experimental interventions: oxygen therapy and indomethacin therapy, to try to restrict the ductus arteriosus, decrease circular shunting, and improve flow to the fetal lungs.

**Fetal arrhythmia evaluation and treatment** – While some abnormal fetal rhythm problems are benign, others may result in fetal heart failure and thus pose a risk to the fetus and mother. Medical therapy is customized for each fetus and mother. We collaborate closely with adult cardiologists to protect the safety of both fetus and mother during treatment.

**Fetal hypoplastic left heart syndrome (HLHS) and related conditions** – We offer three fetal cardiac interventions for babies with small left heart structures, with the type of therapy depending on the fetal heart anatomy.

- For the most severe form of fetal aortic valve stenosis, which may progress to HLHS, we offer a catheter-based intervention called aortic valvuloplasty. Performed by a multidisciplinary team, this procedure consists of placing a small balloon across the fetal aortic valve to enlarge it and to promote blood flow through and growth of the left side of the heart.

- For fetuses with HLHS or mitral valve dysplasia and a restrictive or intact atrial septum, we offer fetal atrial septal intervention. In this catheter-based intervention, either a balloon or a stent is placed across the atrial septum of the heart to decrease the pressure in the left atrium and to help the fetal lungs recover before birth.

- For fetuses with small left-sided structures who may need neonatal heart surgery but do not meet the criteria for the above procedures, we are conducting an experimental study protocol called chronic maternal hyperoxygenation. This treatment involves providing daily home oxygen for expectant mothers to promote fetal left heart growth.
HEART FAILURE

As the number of diagnoses for pediatric heart failure increases, so has our experience in treating patients. Each year, our dedicated team of physicians, nurse coordinators and administrative personnel cares for hundreds of cardiomyopathy and heart failure patients.

In many hospitals, the only option for pediatric heart failure patients is extracorporeal membrane oxygenation (ECMO). Although ECMO is a necessary intervention in some cases – and one with which we have particular expertise – it is not a long-term solution for heart failure. At Texas Children’s Heart Center, we offer a full range of treatment options from medication to heart transplantation and beyond.

Treatment is tailored to the individual needs of each patient. The majority of cardiomyopathy and heart failure patients are managed with medication. A small percentage of patients will go on to require a heart transplant. Since its inception in 1984, Texas Children’s Heart Center has become one of the largest and most successful heart transplant programs in the nation.

Some patients awaiting a heart transplant may require mechanical circulatory support with a ventricular assist device (VAD) as a bridge to transplantation. Since we implanted our first VAD in 1985, we have become one of the most comprehensive pediatric VAD programs in the world, implanting 20-30 VADs annually. Texas Children’s Hospital offers a comprehensive range of both short- and long-term mechanical devices for children of all ages and sizes. Our program is also leading the way in using a VAD as a bridge to recovery.
Innovations in the Use of Ventricular Assist Devices for Children

For patients with heart failure, a ventricular assist device (VAD) can buy valuable time until a suitable heart is located. In some rare cases, a VAD can be used as a permanent therapy for heart failure or can even improve heart function to such a degree that it makes a transplant unnecessary. VADs can either simulate heart function with pulsing action or allow a continuous stream of blood to flow through the heart. Continuous-flow VADs tend to be smaller and quieter but also more durable, and in recent years have yielded improved results in adult heart failure patients.

Led by Dr. Iki Adachi, a world-renowned leader in the development of innovative VAD therapies, Texas Children’s Hospital assisted with the design of the Jarvik 2015, which is the first and only implantable continuous-flow VAD designed specifically for small children. The development of the device took more than a decade. The Jarvik 2015 is an investigational device in the United States. Its safety and effectiveness have not been established and are currently being evaluated under the PumpKIN trial.

Dr. Adachi and the VAD team have also developed innovative therapies with other VAD devices, positioning Texas Children’s Hospital as a cardiac center where the newest VAD technology can be brought to the bedside to match the needs of each patient.
HEART TRANSPLANT

The Heart Transplant program at Texas Children’s Hospital provides complex, multifaceted medical and surgical care for patients from newborns to young adults in need of heart transplants. Since 1984, our team has performed more than 400 pediatric heart transplants. We provide a comprehensive, interdisciplinary team approach through all aspects of the transplant process, from initial referral to hospitalization and outpatient management. Our heart transplant team works closely with patients, families and referring physicians to help make the evaluation process as convenient and efficient as possible.

HEART TRANSPLANTS

Volume by year

HEART TRANSPLANTS IN 2018

By patient age

HEART TRANSPLANT GRAFT SURVIVAL RATES

Pediatric age <18

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1 Scientific Registry of Transplant Recipients (SRTR). Program Specific Reports. Table 11 – srtr.org.
2 Based on transplants performed from July 1, 2015 through December 31, 2017.
3 Based on transplants performed from January 1, 2013 through June 30, 2015.
INTERVENTIONAL CARDIOLOGY

Interventional pediatric cardiology continues to be a rapidly advancing field, with an increasing number of heart conditions able to be treated through minimally invasive, catheter-based interventions. Interventional Cardiology at Texas Children’s Hospital is the largest program of its kind in the region, performing over 1,300 cardiac catheterization procedures in 2018. The Charles E. Mullins Cardiac Catheterization Laboratories were included in the Lester & Sue Smith Legacy Tower expansion and the Heart Center now operates four catheterization labs and an adjacent intra-cath MRI suite. This state-of-the-art environment allows for greater access to care for patients needing cardiac interventional and imaging expertise. Our catheterization labs and advanced MRI diagnostics can be used in a single episode of care to determine how to best intervene in a patient’s condition, but to also immediately determine the effectiveness of the intervention.

Our expansion into Legacy Tower allowed for the introduction of a new generation of catheterization lab systems. The use of this technology directly reduces radiation exposure for our patients without a compromise in imaging. The application of best practices and the innovation of techniques has positioned the Interventional Cardiology service at the forefront of cardiac medicine. Our faculty is actively involved with research that allows for new treatment strategies and device usage that will lead to enhanced patient outcomes. Much of the research work is multi-center and multi-year clinical trials. Our current works includes, but is not limited to:

- Clinical trial of closure of patent ductus arteriosus using the new Amplatzer® Duct Occluder II Additional Sizes (ADO-II AS)
- Clinical trial of closure of atrial septal defects with the new ASSURED trial using the Gore Cardioform ASD device (the highest enrolling site in the country)
- Clinical trial of transcatheter implantation of the new Medtronic Harmony® valve in patients with pulmonary insufficiency after repair of tetralogy of Fallot or after treatment of pulmonary valve stenosis
- Clinical trial of transcatheter implantation of the new Edwards Alterra® pre-stent system in patients with pulmonary insufficiency after repair of tetralogy of Fallot
- Post-approval study of transcatheter closure of muscular ventricular septal defects using the Amplatzer® Muscular Septal Defect Occluder (the highest enrolling site in the country)

The Interventional Cardiology team also has a clear focus on outcomes and quality. The team’s expertise spans from common disease management to less common, highly complex patient conditions. Implementation of pre and post procedure risk assessment has become a standard for every patient we care for. That assessment, combined with close monitoring of our complication data, allows our team to compare our outcomes to national benchmarks. Our adverse event rate has been consistently less than the national benchmark in all risk categories.
2018 ADVERSE EVENT RATE\textsuperscript{14}
Texas Children’s Hospital Actual Compared to Jayaram Expected by Risk Category

The Preventive Cardiology program at Texas Children’s Hospital supports our mission to provide excellence in patient care and improve outcomes for our patients. Our purpose is to prevent future cardiac disease and events such as heart attack and stroke through early detection and intervention, and by educating children and families on how to better care for themselves.

The program uses a collaborative approach in treating patients, drawing on the expertise of a multidisciplinary team of specialists, including experts in pediatric cardiology, nephrology, endocrinology, nutrition and physical therapy. We care for patients with a strong personal or family history of cardiac disease, elevated cholesterol, high blood pressure, obesity or Kawasaki disease. We strive to provide quality service and cost-effective care that will enhance the health and well-being of children locally and regionally.

The Preventive Cardiology program has a commitment to quality service, which is the foundation of ongoing research. Our goal is to provide cutting-edge therapy for patients with modifiable cardiovascular risk factors, increase knowledge through research, and translate research findings to clinical care.

Reasons for Early Intervention
Most adult forms of heart disease are the result of a long process that begins in early childhood. Abnormal cholesterol and high blood pressure are well recognized conditions in adults. It surprises many people that these conditions can also arise in childhood, and that they lay the foundation for heart disease in adulthood.

Some children have liver disease, diabetes or metabolic syndrome in childhood or early adolescence, increasing their risk of early acquired (adult forms) heart disease. While others are at risk because of past chemotherapy, congenital heart disease, Kawasaki or kidney disease. Still another group of children are at risk because of genetic factors. Childhood obesity, poor dietary habits and today’s lifestyle factors (video gaming, high screen time, lack of physical exercise) contribute to risk for early acquired heart disease.

Research shows that maintaining normal weight, consuming a healthy diet and maintaining adequate physical activity reduces risk for adult heart disease. Our research shows that healthy diet and adequate exercise result in improved cholesterol levels and achievement of normal weight. Normalizing cholesterol values and lowering blood pressure in children prevents or slows the development of heart disease as they reach adulthood.
The Single Ventricle program at Texas Children’s Hospital is a multidisciplinary team dedicated to caring for infants with single ventricle congenital heart defects such as hypoplastic left heart syndrome (HLHS), unbalanced atrioventricular septal defects, complex heterotaxy syndromes and related conditions. Our dedicated clinic provides specialized care for patients and their families.

During the critical interstage period – the months between the first operation (Norwood procedure) and the second operation (bidirectional Glenn) – these infants are still at risk of certain problems and conditions such as growth and neurodevelopmental delays, viral illnesses, residual defects and arrhythmia, among others. Parents and caregivers actively participate in monitoring and addressing these issues when they arise. Our home monitoring program provides support, resources and clinical care to help families transition successfully to the second surgery.

We are also actively involved with the National Pediatric Cardiology Quality Improvement Collaborative, working with over 60 centers nationwide towards the goal of improving overall outcomes in infants with single ventricle heart disease from fetal diagnosis to their first birthday.
OUTCOMES & IMPACT SERVICE

The Outcomes & Impact Service at Texas Children’s Hospital measures important clinical outcomes and the impact of care on the quality of life and functional status of our patients over time. Our team’s goal is to deliver the highest value of health care to the patient.

By tracking outcomes, we learn about what happens to our patients, and we also learn about our performance as a health care delivery organization, as we constantly strive to improve.

Our team includes dedicated outcomes nurses in several clinical specialties, computer programmers, data architects and specialists and a statistician who works closely with clinical and administrative teams to measure, improve and share our outcomes.

Our goals include:

• Track and improve our clinical outcomes
• Understand the impact of those outcomes on the lives of our patients and families over time
• Make our outcomes data available in a form accessible by the general public
• Partner with patients and families in understanding outcomes data
• Help patients and families know what questions to ask when seeking medical treatment

For more information, please visit texaschildrens.org/outcomes-and-impact-service.

REFERRALS

To refer a patient to Texas Children’s Heart Center, please visit texaschildrens.org/refer or call us at 832-824-3278.

Texas Children’s Hospital
6621 Fannin St.
Houston, TX 77030
texaschildrens.org/heart