HEART

PEDIATRIC CARDIAC CARE AT CHILDREN'S MERCY

2016 REPORT

James E. O'Brien Jr., MD, FACS

Girish Shirali, MBBS, FACC, FASE

Children's Mercy KANSAS CITY

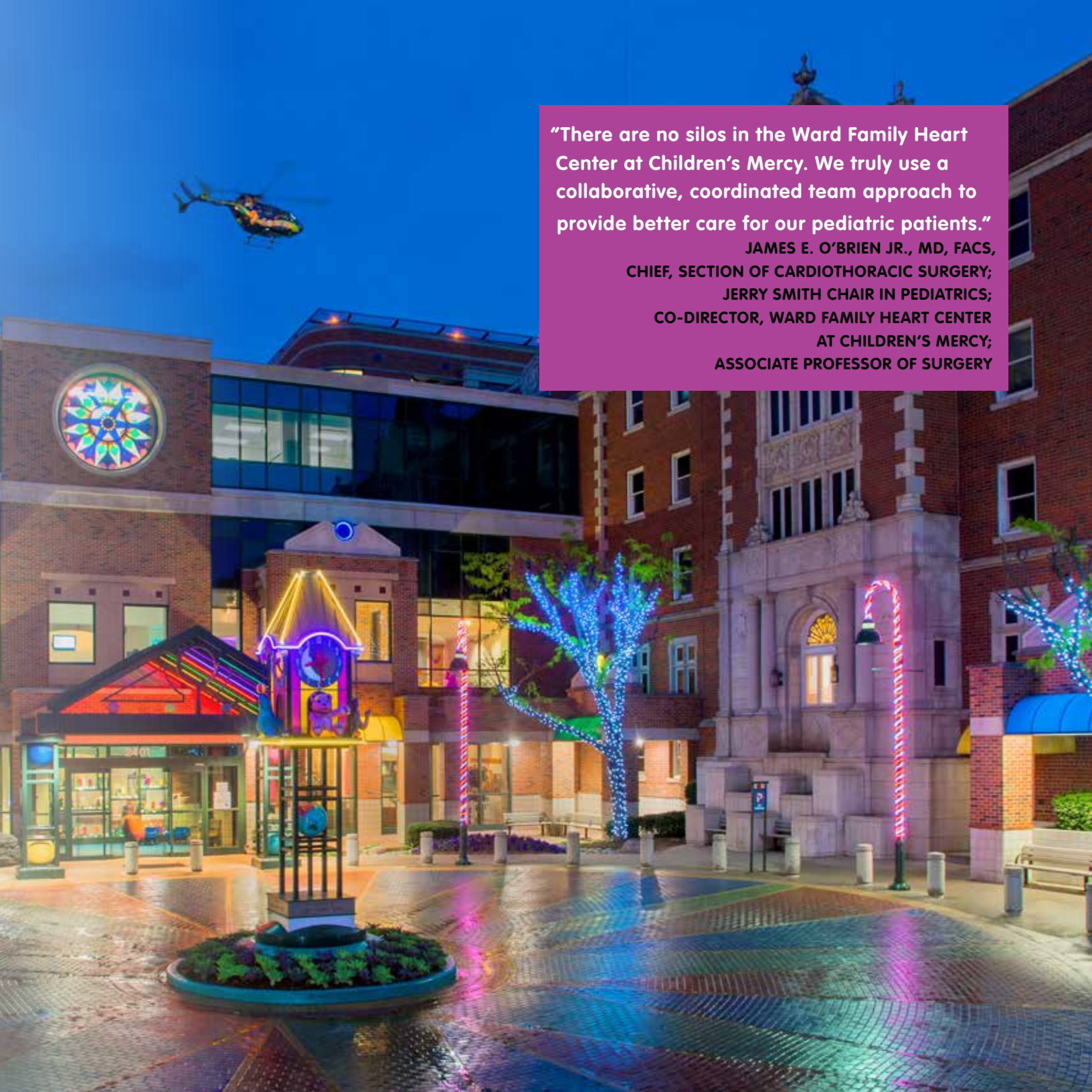
CHILDREN'S MERCY KANSAS CITY

The Ward Family Heart Center at Children's Mercy Kansas City is comprised of a diverse group of professionals with a singular focus—to find answers to the difficult questions our patients and families face each and every day. Questions like. . .

- **How can we improve and simplify at-home monitoring for children born with single-ventricle and their families?**
- **When is three-dimensional echocardiography the right imaging choice?**
- **Do children metabolize statins differently than adults, and if so, why?**
- **Does second-hand smoke contribute to increased risk of heart disease for children?**

Our robust research efforts, service to national and international organizations, creation of innovative technologies and programs and dedication to improved patient outcomes/transparency all contribute to our standing as one of the nation's top-rated pediatric cardiology programs, as recognized by U.S. News & World Report.

Please join us as we share the important progress our team has made in 2016—progress that's resulting in answers for patients and families at Children's Mercy, and for children around the globe.



"There are no silos in the Ward Family Heart Center at Children's Mercy. We truly use a collaborative, coordinated team approach to provide better care for our pediatric patients."

JAMES E. O'BRIEN JR., MD, FACS,
CHIEF, SECTION OF CARDIOTHORACIC SURGERY;
JERRY SMITH CHAIR IN PEDIATRICS;
CO-DIRECTOR, WARD FAMILY HEART CENTER
AT CHILDREN'S MERCY;
ASSOCIATE PROFESSOR OF SURGERY

WE ARE SHARING CHAMP TECHNOLOGY WITH PEDIATRIC CARDIAC PROGRAMS NATIONALLY

Girish Shirali, MBBS, FACC, FASE; Division Director, Cardiology; Melva and Randall L. O'Donnell PhD Chair in Pediatric Cardiology; Co-Director, Ward Family Heart Center at Children's Mercy; Professor of Pediatrics

With the proven success of the Cardiac High Acuity Monitoring Program (better known as the CHAMP App), Children's Mercy is sharing this technology with other pediatric cardiology programs across the nation.

CHAMP uses an innovative PC tablet-based application that automates and simplifies at-home monitoring for children born with single-ventricle heart defect and their families.

The hope is other programs will experience the same level of success as Children's Mercy. In the two-plus years since implementing CHAMP, there have been no deaths in the interstage period for the 60 patients using it.

"We would like to create a single ventricle heart defect registry housed in the cloud that will allow us to track and monitor this low-volume, high-risk population and collaborate with other programs using CHAMP," Dr. Shirali said.

For more information about CHAMP, or to see if your center qualifies for this multi-site project, visit childrensmercy.org/CHAMP or email champapp@cmh.edu.

Recent Awards and Publications

October 2016, CHAMP nurses Bryan Beaven, RN, Amy Ricketts, RN, and Lori Erickson, APRN, accepted "The Magnet Prize® for innovation and nursing excellence worldwide, sponsored by Cerner: http://www.nursecredentialing.org/2016-MagnetPrizeWinner-ChildrensMercyKansasCity.

Harnessing Teams and Technology to Improve Outcomes in Infants with Single Ventricle. Shirali GS, Erickson LA, Goggin K, Apperson J, Bingler M, Tucker D, Williams D, Bradley-Ewing A, Spertus J, Rabbitt, L, Stroup R. (2016) Circulation: Cardiovascular Quality and Outcomes, 9, 303-311; http://circoutcomes.ahajournals.org/content/9/3/303.short.

Cardiac High Acuity Monitoring Program: Early Recognition of Clinical Changes Improves Interstage Resource Utilization among Infants with Single Ventricles. Erickson L, Williams D, Ricketts A, Beaven B, Apperson J, Reid K, Marshall J, Bradley-Ewing A, Stroup R, Bingler M, Lay A, Goggin K, Shirali GS. Cardiology 2016, The Children's Hospital of Philadelphia Cardiac Center Annual Conference, Feb. 2016, Orlando, Fla. Winner of the Nurse Scientist Award.

WE ARE AMONG THE FIRST TO PARTICIPATE IN NATIONAL STS PUBLIC REPORTING OF OUR OUTCOMES

James E. O'Brien Jr., MD, FACS, Chief, Section of Cardiothoracic Surgery; Jerry Smith Chair in Pediatrics; Co-Director, Ward Family Heart Center at Children's Mercy; Associate Professor of Surgery

Children's Mercy was one of the first pediatric cardiac surgery programs in the nation to voluntarily share surgical outcomes with the public via the Society of Thoracic Surgeons' (STS) website.

"That level of transparency is critical to quality improvement for patients, families and staff, as well as for pediatric heart surgery programs nationwide," Dr. O'Brien said.

Dr. O'Brien is a member of the Society's Congenital Heart Disease Database Task Force, as well as one of two auditors for the STS database. Today, of the approximately 120 pediatric heart surgery programs belonging to the database, about half voluntarily share data publicly.

"At Children's Mercy, we have been early adopters of transparency," Dr. O'Brien said. "By evaluating this objective information, we have improved the care we provide patients and their families, helping us rank among the top programs in the country."

For more information go to **childrensmercy.org/heart** or visit **sts.org/congenital-public-reporting-module-search**.

Surgical Mortality Percentage Categorized by Complexity of Procedure

STAT* Category	CM** 1-Year Average	STS*** 1-Year Average	CM 4-Year Average	STS 4-Year Average
1 (least complex)	0.0	0.5	0.3	0.5
2	1.9	1.7	0.7	1.7
3	0.0	2.1	2.2	2.6
4	1.2	6.8	3.9	6.9
5 (most complex)	11.1	17.3	10.4	16.1

*Society of Thoracic Surgeons - EACTS Congenital Heart Surgery Mortality Categories
**Children's Mercy, 2015
***Society of Thoracic Surgeons

This information is based on data submitted by Children's Mercy Hospital to the Society of Thoracic Surgeons National Congenital Heart Surgery Database and compared with combined averages from 114 North American children's cardiac surgery programs.

"Thanks to the STS database, everyone's outcomes have improved, resulting in better patient care across the board."
JAMES E. O'BRIEN JR., MD, FACS

OUR EXPERT IMAGER CO-CHAIRS THREE-DIMENSIONAL ECHOCARDIOGRAPHY GUIDELINES

Girish Shirali, MBBS, FACC, FASE; Division Director, Cardiology; Melva and Randall L. O'Donnell PhD Chair in Pediatric Cardiology; Co-Director, Ward Family Heart Center; Professor of Pediatrics

"As with many modalities, the growth of 3DE has been organic. These guidelines establish a common logic and structure for the display of images."

GIRISH SHIRALI, MBBS, FACC, FASE

Dr. Shirali, an expert in three-dimensional echocardiography (3DE), recently served as the North American co-chair of the joint European-North American writing group responsible for the first set of pediatric 3DE guidelines.

"As clinicians, we encounter a wide range of structural abnormalities in pediatric cardiology that lend themselves superbly to 3D echocardiography, but until now, we have not had expert consensus on their use," Dr. Shirali said. "These guidelines will enhance the clinical adoption of 3DE for the management of patients with congenital heart disease."

The guidelines:

- Outline technical considerations and imaging techniques, particularly for pre-surgical planning.
- Detail the use of 3DE to guide catheter-based interventions.
- Address the need for specific training and educational pathways related to 3DE.
- Summarize techniques and advantages of 3DE for specific types of congenital heart disease, and include 17 figures to illustrate various concepts.

Recent Publications

Three-dimensional Echocardiography in Congenital Heart Disease: An Expert Consensus Document from the European Association of Cardiovascular Imaging (EACVI) and the American Society of Echocardiography. Simpson J, Lopez L, Acar P, Friedberg MK, Khoo NS, Ko HH, Marek J, MD, Marx G, McGhie JS, Meijboom F, Roberson D, Van den Bosch A, Miller O, Shirali GS. J Am Soc Echocardiogr. 2017;30:1-27. PMID 27838227

EAE/ASE Recommendations for Image Acquisition and Display Using Three-Dimensional Echocardiography. Lang RM, Badano LP, Tsang W, Adams DH, Agricola E, Buck T, Faletra FF, Franke A, Hung J, Pérez de Isla L, Kamp O, Kasprzak JD, Lancellotti P, Marwick TH, McCulloch ML, Monaghan MJ, Nihoyannopoulos P, Pandian NG, Pellikka PA, Pepi M, Roberson DA, Shernan SK, Shirali GS, Sugeng L, Ten Cate FJ, Vannan MA, Zamorano JL, Zoghbi WA. J Am Soc Echocardiogr. 2012 Jan;25(1):3-46.PMID 22183020.

WE ARE EARLY ADOPTERS OF ADVANCED MULTI-MODALITY CARDIAC IMAGING

Sanket Shah, MD, MHS, FAAP, Assistant Professor of Pediatrics

Children's Mercy serves as a regional referral center for western Missouri and the state of Kansas, providing advanced multi-modality cardiac imaging, including CT-MRI, 3D echocardiography and 3D printing/modeling.

"We have been early adopters of these advanced imaging techniques, including 3D modeling technology," Dr. Shah said.

By combining CT and MRI data with 3D printing capabilities, the division is creating 3D models for select, complex anatomies including:

- Double outlet right ventricle
- Aortopulmonary collateral arteries
- Aortic arch anomalies
- Anomalous systemic and pulmonary venous return

"Our cardiac surgeons find these models very helpful in improving surgical planning and reducing time in the operating room," Dr. Shah said.

WE ARE ADVANCING THE FIELD OF QUANTITATIVE ECHOCARDIOGRAPHY FOR OUR PATIENTS

Daniel Forsha, MD, MHS, Assistant Professor of Pediatrics
Suma Goudar, MD, Assistant Professor of Pediatrics

Dr. Forsha and Dr. Goudar are using strain analysis to evaluate ventricular mechanics in patients diagnosed with congenital heart defects, including:

- Tetralogy of Fallot
- Single ventricle
- Transposition of the great arteries
- Heart failure

Most of Dr Forsha's work focuses on children diagnosed with congenital heart disease who develop bundle branch block of their systemic ventricle resulting in dyssynchrony. "Predicting response to cardiac resynchronization therapy is especially challenging in this population, but strain pattern analysis may prove useful in identifying the best candidates whose only other option may be transplant."

Dr. Goudar's interest is in examining the correlation between strain and exercise performance in children with single ventricle disease.

Recent Publications

Frequent Activation Delay Induced Mechanical Dyssynchrony and Dysfunction in the Systemic Right Ventricle. Forsha D, Risum, N, Smith PB, Kanter RJ, Samad Z, Barker P, Kisslo J. J Am Soc Echocardiogr. 2016. PMID 27624591. Epub ahead of print.

Patterns of Mechanical Inefficiency in Pediatric Dilated Cardiomyopathy and their Relation to Left Ventricular Function and Clinical Outcomes. Forsha D, Slorach C, Chen CK, Sherman A, Mertens L, Barker P, Kisslo J, Friedberg MK. J Am Soc Echocardiogr. 2016; 29(3): 226-36. PMID 26711366.

The Influence of Angle of Insonation and Target Depth on Speckle Tracking Strain. Forsha D, Risum N, Rajagopal S, Dolgner S, Hornik C, Barnhart H, Kisslo J, Barker P J. Am Soc Echocardiogr. 2015; 28(5): 580-6. PMID 25655402.

Interpreting measurements of cardiac function using vendor-independent speckle tracking echocardiography in children: a prospective, blinded comparison with catheter-derived measurements. Goudar SP, Baker GH, Chowdhury SM, Reid KJ, Shirali G, Scheurer MA. Echocardiography 2016 Dec;33(12):1903-1910.

Speckle-Tracking Echocardiographic Measures of Right Ventricular Diastolic Function Correlate with Reference-Standard Measures Before and After Preload Alteration in Children. Chowdhury SM, Goudar SP, Baker GH, Taylor CL, Shirali GS, Friedberg MK, Dragulescu A, Chessa KS, Mertens L. Pediatr Cardiol. 2016 Sep 21. [Epub ahead of print]

OUR FETAL CHECKLIST ENSURES COMPREHENSIVE PRENATAL EDUCATION FOR FAMILIES OF FETUSES DIAGNOSED WITH CONGENITAL HEART DEFECTS

Tara Swanson, MD, Director of Fetal Cardiology

At Children's Mercy, our goal is not only to improve prenatal diagnosis of congenital heart defects, but to also provide the expertise, counseling and state-of-the-art care necessary to support these children and their families prenatally and beyond.

"We created a comprehensive curriculum to educate and prepare parents before the birth of their child. Our checklist ensures our counseling is standardized and complete but also tailored to the individual needs of each family."

TARA SWANSON, MD

To be certain each concern is addressed, the program's nurse coordinators maintain a fetal checklist, a record of everything the team discusses with the family.

Following Dr. Swanson's presentation of the checklist at a national conference, it has become widely requested by other fetal cardiology and counseling programs across the nation. The National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC), a group of parents and providers who are working to improve outcomes for children with heart defects, is using the checklist to help improve prenatal counseling.

Programs are welcome to download a copy and use it. For a copy of the Fetal Cardiac Program Counseling Checklist, visit: childrensmercy.org/fetalcardiology.

WE ARE INVESTIGATING THE IMPACT OF PHARMACOGENOMICS ON PEDIATRIC STATIN DISPOSITION

Jon Wagner, DO, Pediatric Cardiologist/Clinical Pharmacology

Dr. Wagner was recently selected by the American Heart Association Council on Cardiovascular Disease in the Young as a finalist for the Early Career Investigator Award.

Dr. Wagner's work focuses on how genetic variation of the liver-specific transporter OATP1B1 impacts the amount of statin in the blood after a dose is given. Increased amount of statin in the blood places patients at higher risk of side effects and treatment failure.

Two studies into this important topic are concluding:

- The first investigated the cholesterol-lowering drugs pravastatin and simvastatin. Study participants are 8 to 21 years old with LDL cholesterols greater than 130 mg/dl.
- A second trial looked at the drugs rosuvastatin and atorvastatin.

In his presentation at the American Heart Association's Scientific Sessions in November 2016, Dr. Wagner revealed a novel finding. "Twenty-five percent of the children in our simvastatin study didn't receive anything close to adequate exposure to the medicine. This could result in treatment failure for these patients," Dr. Wagner said. "Our next step in the research process is to understand why these children didn't form to the active drug."

PI Grant Support

American Heart Association National Affiliate Clinical Research Program, July 2013-July 2016

Project: Pharmacokinetics of pravastatin and simvastatin in pediatric dyslipidemia patients: Clinical impact of genetic variation in statin disposition

Award amount: $150,000

Children's Mercy Hospital Marion Merrell Dow Clinical Scholar Award, July 2014-present

Project: Pharmacokinetics of rosuvastatin and atorvastatin in pediatric dyslipidemia patients: Clinical impact of genetic variation in statin disposition

Award Amount: $100,000

Children's Mercy Hospital Clinical Fellowship Research Award, July 2013-June 2015

Project: Cardiology Pharmacogenomics Repository

Award amount: $15,000

Recent Publications and Presentations

Impact of SLCO1B1 Genotype on Pediatric Simvastatin Pharmacokinetics. American Heart Association Scientific Sessions CVDY Early Career Award Finalist Presentation, New Orleans, La., Nov. 13, 2016.

Pediatric Statin Administration: Navigating a Frontier with Limited Data. Wagner J, Rahman S. J Pediatr Pharmacol Ther. (Accepted).

Quantification of Pravastatin Acid, Lactone, and Isomers in Human Plasma by UHPLC-MS/MS and Its Application to a Pediatric Pharmacokinetic Study. Van Haandel L, Gibson K, Leeder JS, Wagner JB. J Chromatogr B Analyt Technol Biomed Life Sci. 2016 Feb 15;1012-1013:169-77. doi: 10.1016/j.jchromb.2016.01.038. Epub 2016 Jan 24. PMID: 26849185

OUR DAILY MANAGEMENT SYSTEM IMPROVES PATIENT-CENTERED CARDIAC CARE FOR INPATIENTS

Amy Lay, MD, Director, Inpatient Cardiology

The Ward Family Heart Center at Children's Mercy has a robust inpatient care unit with excellent outcomes. Our daily management system is being used to standardize inpatient pediatric cardiac care at Children's Mercy. By using this unique system, we are able to improve patient safety.

"We focus on what we need to care for the patients, from simple things such as supplies to more complex items such as appropriate bedside nursing coverage," Dr. Lay said. "This organizes the day-to-day care, allowing us to focus on the big picture of patient safety and the quality of care we are delivering."

The Ward Family Heart Center utilizes cardiac advanced practice nurses (APNs) who anchor the system. "They provide the consistency and continuity of care on the cardiac units," Dr. Lay said. "Our APNs are our clinical experts, present from 7 a.m. to 7 p.m., seven days a week and available 24/7. They are the glue that holds the system together."

Multiple quality improvement efforts have risen from the cardiac inpatient unit. The team discharges patients to home by 11 a.m., in order to safely get patients home during daylight hours. This also allows the team to easily assist families if problems arise while they are settling in at home. "We also evaluate and learn from any transfers that occur in order to improve patient safety," Dr. Lay said. "This is now becoming the standard for the hospital and this process evaluation started on the cardiac floor."

OUR CARDIAC NEURODEVELOPMENTAL PROGRAM IMPROVES PATIENTS' QUALITY OF LIFE

Elizabeth Willen, PhD, Neuropsychologist

The Cardiac Neurodevelopmental Program at Children's Mercy has grown into one of the most comprehensive, robust multidisciplinary programs in the nation, having evaluated nearly 350 patients since 2013.

This unique, multi-faceted program is designed to meet the neurodevelopmental needs of infants, children and adolescents with congenital or acquired heart conditions, including sudden cardiac arrest.

"Our program is a neurodevelopmental safety net," Dr. Willen said. "Our goal is to reduce the impact of congenital heart defects and related conditions on child development by standardizing care and intervening as early as possible."

Key program elements include:

- Serial neurodevelopmental/ neuro psychological assessment beginning at 6 months of age
- Targeted evaluation by CND specialists, including physical, occupational and speech/ language therapies, neurology, psychology and social work
- Development of 504 and individual education plans (IEPs).
- National and international outreach and advocacy through participation in the Cardiac Neurodevelopmental Outcomes Collaborative (CNOC)

Becky Gregory, RN, MSN, CNOR, Cardiac Neurodevelopmental Program Coordinator, serves as the CNOC Treasurer and has been a member of the Steering Committee since 2013. Children's Mercy will host the Cardiac Neurodevelopmental Symposium in 2018.

Dr. Willen has served on the committee's infant working group, which recently published a survey of developmental care practices in pediatric cardiac intensive care units in North America.

Recent Publication

Developmental Care in North American Pediatric Cardiac Intensive Care Units. Sood E, Berends, WM, Butcher JL, Lisanti AJ, Medoff-Cooper B, Singer J, Willen E, and Butler S. Advances in Neonatal Care. Vol. 16, No. 3, pp. 211-219.2016.

OUR PREVENTIVE CARDIOLOGIST CHAIRED THE AHA SCIENTIFIC STATEMENT ON SECOND-HAND SMOKE

Geetha Raghuveer, MD, MPH, FAAP, FACC, FAHA; Director, Preventive Cardiology
David White, PhD, Exercise Physiologist

Dr. Raghuveer and Dr. White, both with the Children's Mercy Preventive Cardiology Clinic, convened an international panel of experts to develop the American Heart Association Scientific Statement on the Cardiovascular Consequences of Childhood Second-hand Tobacco Smoke Exposure: Prevailing Evidence, Burden, and Racial and Socioeconomic Disparities. The statement was published in the September 2016 issue of *Circulation*.

"Our committee included researchers who have spent a lifetime looking at the cardiovascular risks of second-hand or cigarette smoke," Dr. Raghuveer said. Evidence points to early and significant damage to the blood vessels in children who have been exposed to cigarette smoke. It is thus important to educate, counsel and support families to decrease these ill-effects.

Children remain vulnerable as they have little control over their environments, minority children and children from low socio-economic backgrounds are especially affected.

"There is some evidence second-hand smoke is even more damaging than smoking because it is unfiltered," Dr. White added. Dr. Raghuveer serves as Vice Chair of the Atherosclerosis, Hypertension, Obesity in the Youth Committee of the AHA whose mission is to to advance the science related to recognition and treatment of atherosclerosis, hypertension and obesity in children.

Recent Publications

Cardiovascular Consequences of Childhood Second-hand Tobacco Smoke Exposure - Prevailing Evidence, Burden, Racial and Socioeconomic Disparities. American Heart Association Scientific Statement Raghuveer G, White D, Hayman L, Woo J, Villafane J, Celermajer D, Ward K, de Ferranti S, Zachariah J. Circulation September 2016.

Lifelong Cardiovascular Adverse Effects of Childhood Tobacco Smoke Exposure. Spear C, Raghuveer G. Curr Cardiovasc Risk Rep (2016) 10: 26, DOI 10.1007/s12170-016-0508-3 June 2016.

Major Depressive Disorder and Bipolar Disorder Predispose Youth to Accelerated Atherosclerosis and Early Cardiovascular Disease - A Scientific Statement from the American Heart Association on Behalf of the American Heart Association Atherosclerosis, Hypertension and Obesity in Youth Committee of the Council on Cardiovascular Disease in the Young. Goldstein B, Carnethon M, Matthews K, McIntyre R, Miller G, Raghuveer G, Stoney C, Wasiak H, McCrindle B. Circulation. 2015; Sep;132(10):965-86. doi: 10.1161/CIR.0000000000000229. Epub 2015 Aug 10. Review. PMID:26260736.

Vitamin D, Low-grade Inflammation and Cardiovascular Risk in Young Children – A Pilot Study. Singh J, Merrill E D, Sandesara P B, Schoeneberg L, Dai H, Raghuveer G. Pediatr Cardiol.2015; Oct;36(7):1338-43. doi: 10.1007/s00246-015-1162-0. Epub 2015 Apr 3. PMID:25832850.

WE HAVE STANDARDIZED ECHOCARDIOGRAPHY TO IMPROVE THE PATIENT EXPERIENCE

Anitha Parthiban, MD, FAAP, FACC, FASE; Director, Echocardiography

The Echocardiography Laboratory at Children's Mercy is one of the busiest in the nation, providing nearly 18,000 pediatric echoes in 2015 at nine locations in Kansas and Missouri.

"With such a high volume of patients and so many locations, quality improvement processes are critical to improving performance," Dr. Parthiban said. "In our labs, we emphasize a culture of quality improvement involving both our sonographers and physicians."

One of the lab's recent initiatives, which was published in the Journal of the American Society of Echocardiography, involves improving the quality of imaging in post-operative congenital heart disease patients. Using sonographer education, focused echo protocols and increased use of sedation, the echo team improved image quality and completeness of echocardiograms in patients after congenital heart disease surgery.

Recognizing that timeliness of care is an important aspect of patient care, the team implemented a quality improvement project to improve wait time for echo resulting in significant improvement in wait times. Most patients now wait less then 20 minutes from the time that the echocardiogram is ordered by the provider to the start of image acquisition.

Recent Publications

Implementation of a Quality Improvement Bundle Improves Echocardiographic Imaging in Postoperative Congenital Heart Disease. Parthiban A, Levine JC, Nathan M, Marshall J, Shirali G, Simon S, Newburger J, Colan SD, and Raghuveer G. Journal of American Society of Echocardiography 2016 Oct 11. pii: S0894-7317(16)30482-5. doi: 10.1016/j.echo.2016.09.002.

Implementation of a Quality Improvement Program Improves Wait Time for Patients in a Busy Pediatric Echocardiography Laboratory. Parthiban A, Swanson T, Marshall J, Shirali G, Warta A. Journal of American Society of Echocardiography 2015 June; 28:6, B87.

WE PARTICIPATE IN NIH FUNDED PEDIATRIC HEART NETWORK RESEARCH

Geetha Raghuveer, MD, MPH, FAAP, FACC, FAHA; Director, Preventive Cardiology
Jon Wagner, DO, Pediatric Cardiologist/Clinical Pharmacology

Two physician-scientists with Children's Mercy are serving as site lead investigators for Pediatric Heart Network (PHN) studies.

- Dr. Raghuveer is the principal investigator at Children's Mercy for a 17-center multi-institutional collaborative study examining the impact of residual lesions following heart surgery and their effect on outcomes beyond mortality.

"This large study will provide us with a lot of data surrounding issues such as ICU length of stay or complications that will help us better understand the changes we need to make to improve outcomes for these patients," Dr. Raghuveer said.

- Dr. Wagner is the primary investigator for Children's Mercy as one of 24 sites in the U.S., Canada and South Korea to enroll 400 patients 12 to 19 years of age in the Fontan Udenafil Exercise Longitudinal (FUEL) Assessment Trial.

"This trial is novel because it is evaluating the clinical efficacy and safety of udenafil for the treatment of subjects 12 to 19 years old who have undergone the Fontan procedure, but who are otherwise healthy," Dr. Wagner said.

Participants receive a baseline assessment of markers for exercise tolerance and cardiac function, will take the medicine over a six-month period, then will be re-evaluated at the conclusion of the trial for the markers.

OUR ELECTROPHYSIOLOGY TEAM CONTRIBUTES TO INTERNATIONAL GUIDELINES, MULTICENTER STUDIES

John Papagiannis, MD, Director of Electrophysiology

A team of three physicians, John Papagiannis, MD, CEPS, CCDS; Svjetlana Tisma-Dupanovic, MD, CEPS, CCDS; Lindsey Malloy-Walton, DO, MPH, and six EP nurses and technicians performs all levels of interventional EP procedures in children, even in the smallest patients, including infants with hypoplastic left heart syndrome and patients on mechanical circulatory support. More than 200 interventional procedures have been performed in our lab in 2016.

Our team serves patients with genetic arrhythmias, arrhythmias with cardiomyopathy, complex syncope, fetal arrhythmias, with the multidisciplinary care that is required for our most complex patients.

Dr Papagiannis recently served as a member of an international committee of pediatric and adult congenital electrophysiologists who revised the guidelines for catheter ablation in children. He is also the lead investigator of a collaborative international 16-center study examining the outcome of catheter ablation in AV node reentry tachycardia in patients with congenital heart disease.

Recent Publications

PACES/HRS Expert Consensus Statement on the Use of Catheter Ablation in Children and Patients with Congenital Heart Disease: Developed in partnership with the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American Academy of Pediatrics (AAP), the American Heart Association (AHA), and the Association for European Pediatric and Congenital Cardiology (AEPC). Philip Saul J, Kanter RJ; Writing Committee, Abrams D, Asirvatham S, Bar-Cohen Y, Blaufox AD, Cannon B, Clark J, Dick M, Freter A, Kertesz NJ, Kirsh JA, Kugler J, LaPage M, McGowan FX, Miyake CY, Nathan A, Papagiannis J, Paul T, Pflaumer A, Skanes AC, Stevenson WG, Von Bergen N, Zimmerman F. Heart Rhythm. 2016 Jun;13(6):e251-89.

Permanent junctional reciprocating tachycardia in children: a multicenter experience. Kang KT, Potts JE, Radbill AE, La Page MJ, Papagiannis J, Garnreiter JM, Kubus P, Kantoch MJ, Von Bergen NH, Fournier A, Côté JM, Paul T, Anderson CC, Cannon BC, Miyake CY, Blaufox AD, Etheridge SP, Sanatani S. Heart Rhythm. 2014 Aug;11(8):1426-32.

Current management of focal atrial tachycardia in children: a multicenter experience. Kang KT, Etheridge SP, Kantoch MJ, Tisma-Dupanovic S, Bradley DJ, Balaji S, Hamilton RM, Singh AK, Cannon BC, Schaffer MS, Potts JE, Sanatani S. Circ Arrhythm Electrophysiol. 2014 Aug;7(4):664-70.

Early Somatic Mosaicism is a Rare Cause of Long-QT Syndrome. Priest JR, Gawad C, Kahlig KM, Yu JK, O'Hara T, Boyle PM, Rajamani S, Clark MJ, Garcia ST, Ceresnak S, Harris J, Boyle S, Dewey FE, Malloy-Walton L, Dunn K, Grove M, Perez MV, Neff NF, Chen R, Maeda K, Dubin A, Belardinelli L, West J, Antolik C, Macaya D, Quertermous T, Trayanova NA, Quake SR, Ashley EA. Proc Natl Acad Sci U S A. 2016 Oct 11;113(41):11555-11560.

OUR HEART FAILURE AND TRANSPLANT PROGRAM IS FIRST TO JOIN INTERNATIONAL REGISTRY

Aliessa Barnes, MD, Medical Director of Cardiac Transplantation

The Children's Mercy Heart Failure and Transplant Program was the first approved to join the International Pediatric Heart Failure Registry, a collaborative database that can be used to study important questions facing this patient population.

"We believe participation in this registry is the best way to reach the volume of patients necessary to scientifically investigate pediatric transplant and heart failure issues," Dr. Barnes said.

The Children's Mercy program also is a member of the ventricular assist device database, PEDIMACS, and is an active member of the International Pediatric Heart Transplant Study. Dr. Barnes just completed a term on the Board of Directors for the International Pediatric Heart Transplant Study/Foundation. She and her colleague, Brian Birnbaum, MD, are both beginning important prospective trials for new investigational medicines.

In addition to participating in research to expand the field, the heart failure program is going to be piloting an adapted version of the Cardiac High Acuity Monitoring Program (CHAMP) application applying the PC tablet to track data on critical patients waiting for heart transplantation at home. The application will track clinical, imaging and laboratory data allowing real-time trending and interventions to decrease clinic visits, hospitalizations and complications.

Recent Publications

Ethics Rounds: Genomic Contraindications for Heart Transplantation. Char D, Lazaro-Munoz G, Barnes A, Magnus D, Deem M, and Lantos, J. Accepted for publication in Pediatrics.

Viral Cardiac Infections (including Myocarditis, Pericarditis, Endocarditis and Immune Pathologies) Canter CE, Birnbaum BF. Viral Infections in Children Volume 1. Ed: Robin Green. Springer.

OUR CARDIOLOGIST HELPS AUTHOR IMPACT REGISTRY REPORTS

Natalie Jayaram, MD, MSB

Dr. Jayaram authors several of the first reports released by the National Cardiovascular Data Registry's (NCDR) IMPACT Registry (Improving Pediatric and Adult Congenital Treatment).

The multi-center clinical registry collects data on pediatric and adult patients undergoing diagnostic or interventional cardiac catheterization procedures.

Participation in the registry is voluntary, with more than 90 hospitals nationwide taking part, including Children's Mercy. The registry collects data on patients' demographics, medical history and risk factors, detailed procedural information, hemodynamic data, information related to adverse events, and detailed data for six commonly performed interventional procedures. The goal is to gather data to set national benchmarks for diagnostic and certain specific interventional procedures.

Dr. Jayaram's most recent publication evaluates the relationship between hospital annual procedure volume and occurrence of adverse events; it revealed that hospitals performing a threshold volume for congenital cardiac catheterizations may achieve improved patient outcomes.

Recent Publications

Relationship Between Hospital Procedure Volume and Complications Following Congenital Cardiac Catheterization: A Report from the IMPACT® Registry. Jayaram N, Spertus JA, O'Byrne M, Chan PS, Kennedy K, Bergersen L, Glatz AG. Am Heart J. 2017 Jan;183:118-128.

Adjusting for Risk Associated with Pediatric and Congenital Cardiac Catheterization: A Report from the NCDR IMPACT Registry. Jayaram N, Beekman RH, Benson L, Holzer R, Jenkins K, Li Y, Martin GR, Moore J, Ringel R, Rome J, Spertus JA, Vincent R, Bergersen L. Circulation. 2015 Nov 17; 1332 (20): 1863-70.

Characteristics and Safety of Interventions and Procedures Performed During Catheterization of Patients with Congenital Heart Disease; Early Report from the National Cardiovascular Data Registry. Holzer R, Beekman R, Benson L, Bergersen L, Jayaram N, Jenkins K, Kennedy K, Moore J,Ringel R, Rome J, Vincent R, Martin GR. Cardiology in the Young. 2015 Oct 12:1-11.

Procedural Characteristics and Adverse Events for Diagnostic and Interventional Catheterizations in Pediatric and Adult CHD: Initial Report from the IMPACT Registry. Vincent RN, Moore J, Beekman RH, Benson L, Bergersen L, Holzer R, Jayaram N, Jenkins K,Li Y, Ringel R, Rome J, Martin GR. Cardiology in the Young. 2016 Jan; 26 (1): 70-8.

Procedural Results and Safety of Common Interventional Procedures in Congenital Heart Disease (Device Closure of ASD, Device Closure of PDA, Pulmonary Valvuloplasty, Aortic Valvuloplasty, Coarctation of the Aorta Angioplasty and Stenting, and Pulmonary Artery Stenting): Initial Report from the IMPACT Registry®. Moore J, Vincent R, Jayaram N, Li Y, Beekman R, Benson L, Bergersen L, Holzer R, Jenkins K, Ringel R, Rome J, Martin G. Journal of the American College of Cardiology. 2014 Dec 16; 64 (23):2439-51.

OUR SURGEON CONTRIBUTES TO FUTURE ICD CODING AND WORLD SURGICAL DATABASE CREATION

James St. Louis, MD, Surgical Director Cardiac Transplantation and Joseph Boon Gregg/Missouri Endowed Chair in Pediatric Surgery; Professor of Surgery

Dr. St. Louis serves as a co-chair for the Archiving Working Group (AWG) with the International Society for Nomenclature for Pediatric and Congenital Heart Disease.

His role has been to identify, develop and maintain a virtual encyclopedia of representative images and videos that have been acquired from cardiac morphologic specimens, and other image modalities including echocardiography, angiography, CT, MRI and intraoperative videos.

The representative images illustrate the ICD11 codes and definitions as established by the International Pediatric Congenital Cardiac Code (IPCCC). The codes are being released in early 2017 to the World Health Organization and will be utilized for ICD11.

Dr. St. Louis is serving as the co-chair and secretary of the World Society of Pediatric and Congenital Heart Surgery, which is establishing a web-based database that will be available at no charge to pediatric heart surgery programs across the world.

"This is a unified database focused on quality improvement," Dr. St. Louis said. "It is a common platform that programs throughout the world can use to collect everything from diagnosis to procedure, to detailed information about surgery and outcomes."

The database is the first anywhere to include longitudinal follow-up over a 12-month period. Programs will be able to access the database in real-time. Dr. St. Louis presented the database at the World Society for Pediatric and Congenital Heart Surgery meeting in 2016, and the platform is launching in 2017.

Recent Publication

Databases in Pediatric Cardiac Surgery: A Nexus of Opportunity and Obligation. World Journal for Pediatric and Congenital Heart Surgery. Kirklin J, St. Louis J. 7(6): 675-676, 2016.

MAKING A DIFFERENCE THROUGH IMPLEMENTATION OF PULSE OXIMETRY SCREENING

Stephen F. Kaine, MD, Director, Cardiovascular Laboratory; Associate Director, Ward Family Heart Center; Associate Professor of Pediatrics

In September 2011, Secretary of Health and Human Services Kathleen Sebelius recommended that pulse oximetry screening for critical heart defects be added to the uniform screening panel for newborns. Since each state handled the implementation of this screening differently, Dr. Kaine helped spearhead Children's Mercy's efforts with Kansas and Missouri.

"Many states, like Missouri, handled this as a legislative mandate, so we helped provide training for birth facilities and follow up for children with positive screens," Dr. Kaine said. "In the State of Kansas, there was no legislative or regulatory mandate. Instead, we worked with the Kansas Department of Health and Environment to implement screening as a quality improvement project."

The Kansas Department of Health and Environment task force went out to birth facilities and provided education from materials that Children's Mercy provided. Over the course of two years, pulse oximetry screening by birth facilities in the state of Kansas increased from 30 percent to 100 percent.

"Even without a legislative mandate in the State of Kansas, we were able to get babies screened using the quality improvement initiative," Dr. Kaine said.

Now that most states have implemented pulse oximetry screening, developing a standard for reporting the data is necessary. In that way, positive screens can be tracked to define the true impact of pulse oximetry screening to detect unsuspected critical heart defects.

"The state of Kansas now has a field in the electronic birth registry for oximetry screening," Dr. Kaine said. "That allows us to know for sure the number of babies whose heart defects are being detected by this technique. In the state of Missouri, we need to work on better standardized reporting on a patient level, so that we can understand the impact of screening."

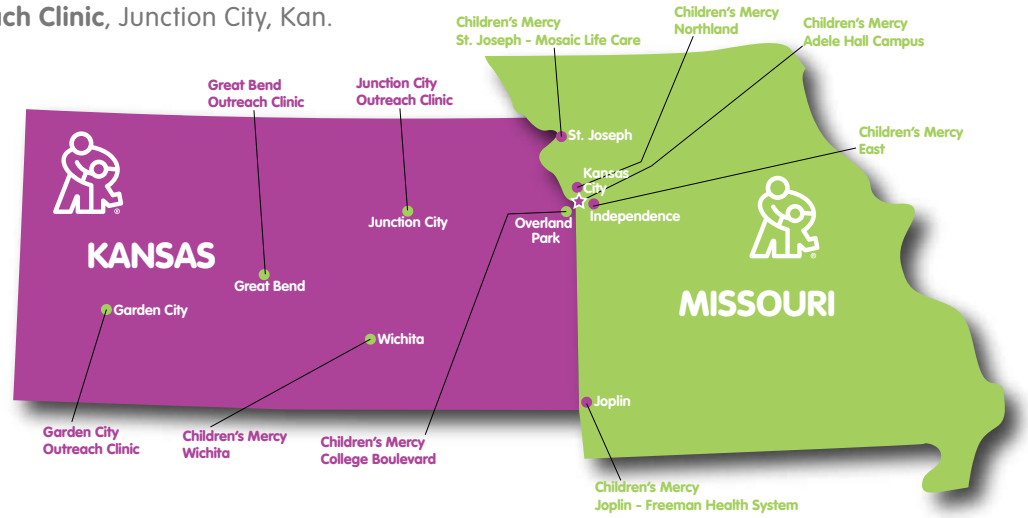
CHILDREN'S MERCY EXTENDS CARDIAC CARE CLOSE TO HOME

The Ward Family Heart Center at Children's Mercy Kansas City is committed to providing outpatient cardiac care close to home for patients in its 150-county service area, from all of Kansas to Western and Southern Missouri. These outreach clinics provide thousands of patients the opportunity to receive the advanced care they need in their own community.

Outpatient Cardiology Clinic locations include:

- **Children's Mercy Adele Hall Campus**, Kansas City, Mo.
- **Children's Mercy College Boulevard**, Overland Park, Kan.
- **Children's Mercy East**, Independence, Mo.
- **Children's Mercy Northland**, Kansas City, Mo.
- **Children's Mercy Wichita**, Wichita, Kan.*
- **Children's Kansas City Joplin-Freeman Health System**, Joplin, Mo.
- **Children's Mercy St. Joseph-Mosaic Life Care**, St. Joseph, Mo.
- **Garden City Outreach Clinic**, Garden City, Kan.
- **Great Bend Outreach Clinic**, Great Bend, Kan.
- **Junction City Outreach Clinic**, Junction City, Kan.

* Specialty clinics in Wichita include preventive cardiology and electrophysiology.



Cardiovascular Surgeons

James E. O'Brien Jr., MD, FACS
Chief, Section of Cardiothoracic Surgery; Jerry Smith Chair in Pediatrics; Co-Director, Ward Family Heart Center

Peter Pastuszko, MD
Congenital Cardiovascular Surgeon

James St. Louis, MD
Surgical Director, Cardiac Transplantation, Joseph Boon Gregg/Missouri Endowed Chair in Pediatric Surgery

Cardiologists

Girish Shirali, MBBS, FASE, FACC
Division Director, Cardiology; Melva and Randall O'Donnell, PhD Chair in Pediatric Cardiology; Co-Director, Ward Family Heart Center

Michael Artman, MD
Joyce C. Hall Eminent Scholar in Pediatrics; Chair, Department of Pediatrics; Senior Vice President and Pediatrician-in-Chief

Aliessa Barnes, MD
Medical Director, Heart Failure and Cardiac Transplantation; Director, Pediatric Cardiology Fellowship Training

Daniel Beissel, MD

Brian Birnbaum, MD
Director, Pulmonary Hypertension

Karina Carlson, MD

Abhay Divekar, MD
Director, Interventional Cardiology

Arpan Doshi, MD
Wichita, Kan.

Unnati Doshi, MD, MPH
Wichita, Kan.

Daniel Forsha, MD, MHS

Mark Gelatt, MD
Director, Outpatient Services

Suma Goudar, MD

Hayley Hancock, MD

Howard Heching, MD

Natalie Jayaram, MD, MSB

Stephen Kaine, MD
Associate Director, Ward Family Heart Center; Director, Cardiovascular Laboratory; Associate Director, Fellowship Training Program

Maria Kiaffas, MD

Amy Lay, MD
Director, Inpatient Cardiology and CHAMP Program

Luisa Madroñero Waitman, MD
Director, Cardiovascular Genetics

Lindsey Malloy-Walton, DO, MPH

John Papagiannis, MD, CCDS, CEPS
Director, Electrophysiology

Anitha Parthiban, MD
Director, Echocardiography

Geetha Raghuveer, MD
Director, Preventive Cardiology

Sanket Shah, MD

Catherine Simon, MD

Tara Swanson, MD
Director, Fetal Cardiology

Svjetlana Tisma-Dupanovic, MD, CCDS, CEPS

Louay Toni, MD
Director, Wichita Cardiology, Wichita, Kan.

Jonathan Wagner, DO

Cardiac Anesthesiologists

Carrie M. Clarke, MD

William B. Daniels, DO

John Erkmann, DO

Joseph Huffman, MD

Kathy M. Perryman, MD, FAAP

Paul Sheeran, MD

Adam B. Striker, MD

Susan J. Whitney, MD

Cardiac Intensivists

Jeremy Affolter, MD

Geoffrey L. Allen, MD

Bruce Banwart, MD

Jennifer Flint, MD

Laura Miller-Smith, MD

Erica A. Molitor-Kirsch, MD

Brian S. Olsen, MD, FAAP

Laura A. Ortmann, MD

Kelly S. Tieves, DO, MS

Cardiac Neonatologists

Howard W. Kilbride, MD

Michael Nyp, DO

Felix Okah, MBBS, MS

Steven L. Olsen, MD

Eugenia K. Pallotto, MD, MSCE

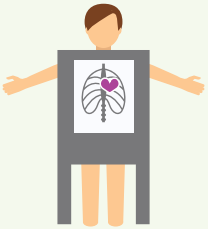
Joshua E. Petrikin, MD

Julie Weiner, DO

19,775
EKGs

18,260
echocardiograms

1,561
Holter Monitor and Event Recorder Exams



543
Cardiac Catheterization and Electrophysiology Procedures

15,704
Outpatient Visits

501
Surgical Procedures

Children's Mercy Kansas City

Children's Mercy Kansas City is an independent, non-profit 367-bed pediatric health system, providing more than half a million patient encounters yearly. Children's Mercy is ranked by U.S. News & World Report as one of "America's Best Children's Hospitals" and has received Magnet recognition four times for excellence in nursing services. In affiliation with the University of Missouri-Kansas City, the medical staff of nearly 750 pediatric subspecialists and scientists is actively involved in clinical care, pediatric research, and educating the next generation of pediatric subspecialists. The hospital's leadership in pediatric genomic medicine and clinical pharmacology is driving research and innovation in nephrology, heart care, cancer treatment and other subspecialties to provide answers for the most difficult cases and challenging pediatric conditions.

Children's Mercy KANSAS CITY

For children. For families. For answers.℠

To learn more about how we are finding answers to improve cardiology care, visit childrensmercy.org/heart

Cardiology Office: (816) 234-3880

For transport, admissions or consults, call:
1 (800) GO MERCY / 1 (800) 466-3729

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