

Research Highlights

Shelley Kirk, PhD, RD, LD

Dr. Kirk serves as Principal Investigator for the study, "Establishment of the Pediatric Obesity Weight Evaluation Registry (POWER): A prospective pilot project of children and adolescents presenting for weight management." The goal is to establish a national registry of children and adolescent participating in multi-component weight management programs to serve as a resource to promote high quality research as it relates to the evaluation and management of childhood overweight and obesity. Currently there are 28 sites (representing 20 states) that are actively participating in POWER. The POWER database, developed for the Medidata Rave® system by Eileen King, PhD, lead for the Data Coordinating Center for POWER, and her staff at Cincinnati Children's went "live" (i.e. ready for data entry) on June 2, 2014.

Robert Siegel, MD

Dr. Siegel's group facilitated a project entitled "Small Prizes Increase Healthful Food Selection in a School Cafeteria." They were able to increase the selection of the most healthful meal, "The Power Plate," by over 300% by offering elementary students a small prize. The intervention was done at the Frederick Douglass School in collaboration with Cincinnati Public Schools and involved over 200 students.

Jeffrey A. Towbin, MD

The Towbin Lab identified that the cardiovascular disease affecting sickle cell disease and associated sudden death is restrictive cardiomyopathy and not dilated cardiomyopathy with pulmonary hypertension, as is widely described. This has been shown in our patient population and mouse models of sickle cell disease. We are currently determining the causative mechanism(s).

Elaine M. Urbina, MD, MS

Dr. Urbina received an award for one of the top three abstracts submitted to the American Heart Association National meeting in Dallas, TX, November, 2013 for an oral presentation entitled, "Lipoprotein Particle Number and Size Predict Vascular Structure and Function Better than Lipid Concentration in Young Subjects." This work demonstrates that advanced lipid testing is a more powerful predictor of early atherosclerosis in high-risk subjects than traditional cholesterol testing.

Division Summary

RESEARCH AND TRAINING DETAILS

Number of Faculty	46
Number of Joint Appointment Faculty	4
Number of Research Fellows	4
Number of Support Personnel	62
Direct Annual Grant Support	\$3,100,807
Direct Annual Industry Support	\$138,874
Peer Reviewed Publications	110

CLINICAL ACTIVITIES AND TRAINING

Number of Clinical Staff	129
Number of Staff Physicians	1
Number of Clinical Fellows	21
Number of Other Students	2
Inpatient Encounters	1,374
Outpatient Encounters	16,413

Ryan TD, Jefferies JL, Sawnani H, Wong BL, Gardner A, Del Corral M, Lorts A, Morales DL. Implantation of the HeartMate II and HeartWare left ventricular assist devices in patients with duchenne muscular dystrophy: lessons learned from the first applications. *ASAIO J.* 2014 Mar-Apr;60(2):246-8.

Duchenne muscular dystrophy (DMD) is a chronic muscle wasting disease that affects 1:3500 boys and currently has no cure. While recent medical advances have improved quality of life for patients, cardiomyopathy accounts for 20-40% of all deaths. This report documents the first use in the United States of surgically-placed mechanical pumps to assist heart function, and only the 3rd and 4th patients in the world with DMD to undergo such a procedure. Not only does it demonstrate the improvement in symptoms experienced by these two patients, but also the lessons learned by our multidisciplinary care team. The ultimate goal is to alter the natural history of DMD and provide longer symptom-free survival. Since publication, the Heart Institute has continued to use mechanical pumps in patients with DMD, and other centers across the world are also reporting their use.

Urbina EM, de Ferranti S, Steinberger J. Observational studies may be more important than randomized clinical trials: weaknesses in US preventive services task force recommendation on blood pressure screening in youth. *Hypertension.* 2014 Apr;63(4):638-40.

In early 2013, the US Preventive Services Task Force (USPSTF) published the findings of an evidenced-based literature review and concluded that there was insufficient evidence to recommend screening for hypertension in children for the purpose of preventing adult cardiovascular (CV) disease. This editorial represents the response of a group of pediatric hypertension experts from the American Heart Association. After carefully examining the review process and the final product, we concluded that the Task Force's recommendation was flawed because the evidence was too narrowly selected, and available shorter-term, observational data and studies evaluating target organ damage were not considered. We continue to recommend routine (yearly) blood pressure (BP) screening in children and adolescents. We look forward to data soon to come from collaboration among the worlds' largest childhood CV studies that are now evaluating childhood BP and risk for adult CV events. We believe these data will support our recommendation for routine BP screening in youth.

Webb G, Landzberg MJ, Daniels CJ. Specialized adult congenital heart care saves lives. *Circulation.* 2014 May 6;129(18):1795-6.

For over 50 years, growing numbers of practitioners and clinician scientists have created systems for and practiced healthcare delivery within centers of expertise for young and older adults with congenital heart disease. This editorial commented on the first study to provide good evidence that there is a survival advantage to ACHD patients being cared for by ACHD specialists. Mylotte colleagues in the province of Québec showed that soon after the presentation of the first Canadian ACHD guidelines, referrals to ACHD clinics in the province of Québec steadily increased. Soon thereafter, province-wide mortality in ACHD patients declined. Efforts to build excellence in ACHD care need to be redoubled.

Xiong D, He H, **James J**, Tokunaga C, Powers C, Huang Y, Osinska H, **Towbin JA, Purevjav E**, Balschi JA, Javadov S, McGowan FX Jr, **Strauss AW, Khuchua Z.** Cardiac-specific VLCAD deficiency induces dilated cardiomyopathy and cold intolerance. *Am J Physiol Heart Circ Physiol.* 2014 Feb;306(3):H326-38.

The heart requires an enormous amount of energy to support its mechanical function and pump blood through arteries and veins. Energy comes in the form of ATP, which is a universal metabolic currency in the cell. 70% of ATP in healthy adult hearts is derived from oxidation of fatty acids in mitochondria, a cellular powerhouse. A key enzyme of fatty acid oxidation is called very-long-chain-Acyl-CoA dehydrogenase, or VLCAD. Mutations in the VLCAD gene cause clinical conditions including hypoglycemia, heart and liver lipidoses, encephalopathy, skeletal myopathy, cardiomyopathy, arrhythmias and sudden death.

In this paper the Khuchua Lab reported the consequences of VLCAD gene ablation in mouse hearts. They demonstrated that VLCAD gene ablation results in dilated cardiomyopathy, intolerance to physiological stress and reduced ATP production in cardiomyocytes. This work is essential for understanding pathophysiology of metabolic disorders caused by inborn errors fatty acid oxidation genes.

Division Publications

1. Aboulhosn JA, Lluri G, Gurvitz MZ, Khairy P, Mongeon FP, Kay J, Valente AM, Earing MG, Opotowsky AR, Lui G, Gersony DR, Cook S, Child J, Ting J, Webb G, Landzberg M, Broberg CS, Alliance for Adult Research in Congenital C. **Left and right ventricular diastolic function in adults with surgically repaired tetralogy of Fallot: a multi-institutional study.** *Can J Cardiol.* 2013; 29:866-72.
2. Baker-Smith CM, Wilhelm CM, Neish SR, Klitzner TS, Beekman RH, 3rd, Kugler JD, Martin GR, Lannon C, Jenkins KJ, Rosenthal GL. **Predictors of prolonged length of intensive care unit stay after stage I palliation: a report from the National Pediatric Cardiology Quality Improvement Collaborative.** *Pediatr Cardiol.* 2014; 35:431-40.
3. Brown KL, MacLaren G, Marino BS. **Looking beyond survival rates: neurological outcomes after extracorporeal life support.** *Intensive Care Med.* 2013; 39:1870-2.
4. Budts W, Borjesson M, Chessa M, van Buuren F, Trigo Trindade P, Corrado D, Heidbuchel H, Webb G, Holm J, Papadakis M. **Physical activity in adolescents and adults with congenital heart defects: individualized exercise prescription.** *Eur Heart J.* 2013; 34:3669-74.
5. Burch PT, Gerstenberger E, Ravishankar C, Hehir DA, Davies RR, Colan SD, Sleeper LA, Newburger JW, Clabby ML, Williams IA, Li JS, Uzark K, Cooper DS, Lambert LM, Pemberton VL, Pike NA, Anderson JB, Dunbar-Masterson C, Khaikin S, Zyblewski SC, Minich LL, Pediatric Heart Network I. **Longitudinal assessment of growth in hypoplastic left heart syndrome: results from the single ventricle reconstruction trial.** *J Am Heart Assoc.* 2014; 3:e000079.
6. Butts RJ, Zak V, Hsu D, Cnota J, Colan SD, Hehir D, Kantor P, Levine JC, Margossian R, Richmond M, Szwast A, Williams D, Williams R, Atz AM. **Factors associated with serum B-type natriuretic peptide in infants with single ventricles.** *Pediatr Cardiol.* 2014; 35:879-87.
7. Byrnes JW, Fiser RT. **Comparing outcomes in ECMO between roller and centrifugal pumps in the face of evolving technology.** *Ann Thorac Surg.* 2013; 96:376.
8. Byrnes JW, Frazier E, Tang X, Eble B, McKamie A, Gomez A, Imamura M, Prodhan P. **Hemorrhage requiring surgical intervention among children on pulsatile ventricular assist device support.** *Pediatr Transplant.* 2014; 18:385-92.
9. Byrnes JW, Prodhan P, Williams BA, Schmitz ML, Moss MM, Dyamenahalli U, McKamie W, Morrow WR, Imamura M, Bhutta AT. **Incremental reduction in the incidence of stroke in children supported with the Berlin EXCOR ventricular assist device.** *Ann Thorac Surg.* 2013; 96:1727-33.
10. Byrnes JW, Swearingen CJ, Prodhan P, Fiser R, Dyamenahalli U. **Antithrombin III supplementation on extracorporeal membrane oxygenation: impact on heparin dose and circuit life.** *ASAIO J.* 2014; 60:57-62.
11. Calvert PA, Cockburn J, Wynne D, Ludman P, Rana BS, Northridge D, Mullen MJ, Malik I, Turner M, Khogali S, Veldtman GR, Been M, Butler R, Thomson J, Byrne J, MacCarthy P, Morrison L, Shapiro LM, Bridgewater B, de Giovanni J, Hildick-Smith D. **Percutaneous closure of postinfarction ventricular septal defect: in-hospital outcomes and long-term follow-up of UK experience.** *Circulation.* 2014; 129:2395-402.
12. Castleberry C, Ryan TD, Chin C. **Transplantation in the highly sensitized pediatric patient.** *Circulation.* 2014; 129:2313-9.

13. Castleberry C, White-Williams C, Naftel D, Tresler MA, Pruitt E, Miyamoto SD, Murphy D, Spicer R, Bannister L, Schowengerdt K, Gilmore L, Kaufman B, Zangwill S. **Hypoalbuminemia and poor growth predict worse outcomes in pediatric heart transplant recipients.** *Pediatr Transplant.* 2014; 18:280-7.
14. Castleberry CD, Gudauskas TM, Berger S, Tweddell JS, Pelech AN. **Stenting of the right ventricular outflow tract in the high-risk infant with cyanotic teratology of Fallot.** *Pediatr Cardiol.* 2014; 35:423-30.
15. Cnota JF, Hangge PT, Wang Y, Woo JG, Hinton AC, Divanovic AA, Michelfelder EC, Hinton RB. **Somatic growth trajectory in the fetus with hypoplastic left heart syndrome.** *Pediatr Res.* 2013; 74:284-9.
16. Connor JA, Hinton RB, Miller EM, Sund KL, Ruschman JG, Ware SM. **Genetic testing practices in infants with congenital heart disease.** *Congenit Heart Dis.* 2014; 9:158-67.
17. Conway J, Chin C, Kemna M, Burch M, Barnes A, Tresler M, Scheel JN, Naftel DC, Beddow K, Allain-Rooney T, Dipchand AI, Pediatric Heart Transplant Study I. **Donors' characteristics and impact on outcomes in pediatric heart transplant recipients.** *Pediatr Transplant.* 2013; 17:774-81.
18. Costello JM, Pasquali SK, Jacobs JP, He X, Hill KD, Cooper DS, Backer CL, Jacobs ML. **Gestational age at birth and outcomes after neonatal cardiac surgery: an analysis of the society of thoracic surgeons congenital heart surgery database.** *Circulation.* 2014; 129:2511-7.
19. Czossek RJ, Anderson J, Cassidy A, Spar DS, Knillans TK. **Cost-effectiveness of various risk stratification methods for asymptomatic ventricular pre-excitation.** *Am J Cardiol.* 2013; 112:245-50.
20. Dabelea D, Talton JW, D'Agostino R, Jr., Wadwa RP, Urbina EM, Dolan LM, Daniels SR, Marcovina SM, Hamman RF. **Cardiovascular risk factors are associated with increased arterial stiffness in youth with type 1 diabetes: the SEARCH CVD study.** *Diabetes Care.* 2013; 36:3938-43.
21. Daily JA, Tarango C, Czossek RJ, Anderson JB, Spar DS. **Placement of an Implantable Cardioverter Defibrillator in a Pediatric Patient with Heparin-Induced Thrombocytopenia with Thrombosis, Bridging with Argatroban.** *Congenital Heart Today.* 2014; 12:10-12.
22. Dandoy CE, Hirsch R, Chima R, Davies SM, Jodele S. **Pulmonary hypertension after hematopoietic stem cell transplantation.** *Biol Blood Marrow Transplant.* 2013; 19:1546-56.
23. Davies RR, Russo MJ, Reinhartz O, Maeda K, Rosenthal DN, Chin C, Bernstein D, Mallidi HR. **Lower socioeconomic status is associated with worse outcomes after both listing and transplanting children with heart failure.** *Pediatr Transplant.* 2013; 17:573-81.
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26. Dupuis LE, Osinska H, Weinstein MB, Hinton RB, Kern CB. **Insufficient versican cleavage and Smad2 phosphorylation results in bicuspid aortic and pulmonary valves.** *J Mol Cell Cardiol.* 2013; 60:50-9.
27. Everitt MD, Sleeper LA, Lu M, Canter CE, Pahl E, Wilkinson JD, Addonizio LJ, Towbin JA, Rossano J, Singh RK, Lamour J, Webber SA, Colan SD, Margossian R, Kantor PF, Jefferies JL, Lipshultz SE, Pediatric Cardiomyopathy Registry I. **Recovery of echocardiographic function in children with idiopathic dilated cardiomyopathy: results from the pediatric cardiomyopathy registry.** *J Am Coll Cardiol.* 2014; 63:1405-13.
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- Cardiovascular Disease in the Y. **Update: ambulatory blood pressure monitoring in children and adolescents: a scientific statement from the American Heart Association.** *Hypertension*. 2014; 63:1116-35.
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31. Guerrier K, Czosek RJ, Spar DS, Anderson J. **Long QT genetics manifesting as atrial fibrillation.** *Heart Rhythm*. 2013; 10:1351-3.
32. Hange PT, Cnota JF, Woo JG, Hinton AC, Divanovic AA, Manning PB, Ittenbach RF, Hinton RB. **Microcephaly is associated with early adverse neurologic outcomes in hypoplastic left heart syndrome.** *Pediatr Res*. 2013; 74:61-7.
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36. Hor KN, Taylor MD, Al-Khalidi HR, Cripe LH, Raman SV, Jefferies JL, O'Donnell R, Benson DW, Mazur W. **Prevalence and distribution of late gadolinium enhancement in a large population of patients with Duchenne muscular dystrophy: effect of age and left ventricular systolic function.** *J Cardiovasc Magn Reson*. 2013; 15:107.
37. Hornik CP, Krawczeski CD, Zappitelli M, Hong K, Thiessen-Philbrook H, Devarajan P, Parikh CR, Patel UD, Consortium T-A. **Serum brain natriuretic peptide and risk of acute kidney injury after cardiac operations in children.** *Ann Thorac Surg*. 2014; 97:2142-7.
38. Irby K, Swearingen C, Byrnes J, Bryant J, Prodhan P, Fiser R. **Unfractionated heparin activity measured by anti-factor Xa levels is associated with the need for extracorporeal membrane oxygenation circuit/membrane oxygenator change: a retrospective pediatric study.** *Pediatr Crit Care Med*. 2014; 15:e175-82.
39. Irving SY, Medoff-Cooper B, Stouffer NO, Schall JI, Ravishankar C, Compher CW, Marino BS, Stallings VA. **Resting energy expenditure at 3 months of age following neonatal surgery for congenital heart disease.** *Congenit Heart Dis*. 2013; 8:343-51.
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43. Jefferies JL. **Barth syndrome.** *Am J Med Genet C Semin Med Genet.* 2013; 163C:198-205.
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- cardiomyopathy.** *J Card Fail.* 2013; 19:672-7.
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Faculty, Staff, and Trainees

Faculty Members

Jeffrey A. Towbin, MD, FAAP, FACC, FAHA, Professor

Leadership Executive Co-Director, The Heart Institute; Director and Chief, Division of Cardiology

Research Interests Cardiomyopathy and genetics

Jeffrey B. Anderson, MD, MPH, MBA, Assistant Professor

Leadership Chief Quality Officer, Heart Institute

Research Interests Quality and value improvement; nutritional failure in congenital heart disease; syncope

Robert H. Beekman, III, MD, Professor

Research Interests Cardiac catheterization and intervention, quality improvement

Jonathan W. Byrnes, MD, Assistant Professor

Research Interests Coagulation, hemostasis and thrombosis in the critically ill.

Chesney Castleberry, MD, Assistant Professor

Research Interests Immunosuppression therapy in cardiac transplantation; clinical outcomes after transplantation

Clifford Chin, MD, Professor

Leadership Medical Director, Pediatric Heart Transplant Service

Research Interests Cardiomyopathy, heart failure, and cardiac transplantation

James F. Cnota, MD, Associate Professor

Leadership Director, Neonatal Cardiology

Research Interests Fetal cardiology, multicenter clinical trials

David S. Cooper, MD, MPH, Assistant Professor

Leadership Associate Medical Director, Cardiac Intensive Care Unit; Director, Cardiac ECMO Program; Chief Safety Officer, Heart Institute; Co-Director, Center for Acute Care Nephrology

Research Interests Acute kidney injury in critical cardiac disease, anticoagulation on extracorporeal support, outcomes research

Richard J. Czosek, MD, Associate Professor

Leadership Director, Cardiac Rhythm Device Management

Research Interests Cardiac pacing device and arrhythmia management in pediatric patients with congenital heart disease and cardiomyopathy

Allison Divanovic, MD, Assistant Professor

Research Interests Fetal echocardiography

Paula Goldenberg, MD, MSCE, MSW, FAAP, FACMG, Assistant Professor

Research Interests Cardiovascular genetics

Bryan H. Goldstein, MD, Assistant Professor

Research Interests Cardiac catheterization; intervention; hybrid therapies for congenital heart disease; single ventricle outcomes; vascular function in congenital heart disease

Samuel Hanke, MD, MS, Assistant Professor

Leadership Quality Scholar in Healthcare Transformation, James M. Anderson Center for Health Systems Excellence

Research Interests Improvement of hospital care transitions and its impact on hospital readmissions in patients with congenital heart disease

Haleh Heydarian, MD, Assistant Professor

Leadership Medical Director, Heart Institute Single Ventricle Clinic

Research Interests Echocardiography Synchrony/Strain Imaging and Quality Improvement

Robert B. Hinton, MD, Associate Professor

Leadership Director, Heart Institute BioRepository

Research Interests Cardiovascular genetics and developmental biology

Russel Hirsch, MD, Associate Professor

Leadership Director, Pulmonary Hypertension Service; Director, Cardiac Catheterization Laboratory

Research Interests Cardiac Catheterization & Intervention, Device Development

Ilias Iliopoulos, MD, Assistant Professor

Research Interests Hemodynamic management and use of inotropes in cardiac intensive care

Holly M. Ippisch, MD, MS, FAAP, Assistant Professor

Research Interests Pediatric preventative cardiology; lipids; hypertension; cardiovascular effects of pediatric obesity; cardiovascular effects of exercise

John Lynn Jefferies, MD, MPH, FAAP, FACC, FAHA, Associate Professor

Leadership Director, Advanced Heart Failure; Cardiomyopathy; Associate Professor, Human Genetics

Research Interests Cardiomyopathy; advanced heart failure; drug trials; novel gene discovery; quality of life; quality improvement

Thomas R. Kimball, MD, Professor

Leadership Medical Director, Heart Institute

Research Interests Echocardiography, ventricular function, cardiovascular effects of obesity and type II diabetes

Shelley Kirk, PhD, RD, LD, Associate Professor

Research Interests The efficacy, safety and feasibility of interventions for the management of pediatric overweight and obesity

Timothy Knilans, MD, Professor

Leadership Director, Clinical Cardiac Electrophysiology; Director, Pediatric Cardiac Electrophysiology Fellowship; Director, Cardiac Exercise Laboratory

Research Interests Identification and risk stratification of causes of sudden death

Catherine Krawczeski, MD, Associate Professor

Leadership Director, Heart Institute Quality Improvement and Clinical Effectiveness; Co-Director, Center for Acute Care Nephrology

Research Interests Cardiac surgery-associated acute kidney injury, cardiorenal syndromes, biomarkers, quality improvement

Christopher Learn, MD, Assistant Professor

Leadership Co-Director, Adolescent and Adult Congenital Cardiology Fellowship Program

Research Interests Non-invasive imaging in congenital heart disease; adolescent/young adult transition care for persons with congenital heart disease; heart disease in pregnancy

Lisa A. Lee, MD, Assistant Professor

Angela Lorts, MD, Associate Professor

Leadership Director, Ventricular Assist Device Program

Research Interests Mechanical circulatory support; myocardial remodeling; anticoagulation

Nicolas Madsen, MD, MPH, Assistant Professor

Leadership Medical Director, Inpatient Cardiology

Research Interests implications of traditional cardiovascular risk factors such as obesity, hypertension, and sedentary lifestyle on congenital heart disease patients

Peace Madueme, MD, MS, Assistant Professor

Research Interests 3D Heart Modeling; Echo/MRI Quantification

Bradley S. Marino, MD, MPP, MSCE, Associate Professor

Research Interests Outcomes Research

Erik Michelfelder, MD, Associate Professor

Leadership Co-Director, Heart Institute Imaging Service; Director, Fetal Heart Program

Research Interests Fetal cardiology and echocardiography

Erin Miller, MS, LGC, Assistant Professor

Research Interests Genetic etiology of cardiovascular disease; utilization and uptake of genetic testing for diagnosis and management of inherited arrhythmias, cardiomyopathies, aortopathies, and congenital heart disease

David Nelson, MD, PhD, Professor

Leadership Medical Director, CICU

Research Interests Inflammatory injury after cardiac surgery, endocrine dysfunction in critical illness

Enkhsaikhan Purevjav, MD, PhD, Assistant Professor

Research Interests Genetics of Cardiovascular Disease

Thomas D. Ryan, MD, PhD, FACC, Assistant Professor

Research Interests Patients at risk to develop cardiomyopathy, including those with Duchenne muscular dystrophy, malignancy and anthracycline exposure, epidermolysis bullosa

Pirouz Shamszad, MD, Assistant Professor

Research Interests Patient outcomes and quality improvement in pediatric cardiac intensive care

Robert Siegel, MD, Professor

Leadership Medical Director, The Center for Better Health and Nutrition

David Spar, MD, Assistant Professor

Research Interests Pediatric and congenital electrophysiology

Christopher Joseph Statile, MD, Assistant Professor

Research Interests Functional analysis of the fetal heart; quality improvement within cardiac imaging

Joshua James Sticka, MD, Assistant Professor

Leadership Co-Director, Heart Institute Resident; Medical Student Education

Research Interests Non-invasive cardiac imaging; myocardial mechanics

Arnold Strauss, MD, Professor

Leadership BK Rachford Professor and Chair, Department of Pediatrics, University of Cincinnati College of Medicine; Director, Cincinnati Children's Research Foundation; Chief Medical Officer, Cincinnati Children's Hospital Medical Center

Michael Taylor, MD, PhD, Assistant Professor

Leadership Director of Advanced Imaging Innovation and Cardiac MRI (CMR); Co-Director, The Heart Institute Imaging Service

Research Interests Non-invasive study of cardiovascular physiology and metabolism; Pre-clinical imaging of cardiomyopathy models with cardiac MR and positron emission tomography

Elaine M. Urbina, MD, MS, Professor

Leadership Director, Preventive Cardiology

Research Interests Non-invasive assessment of vascular structure and function and relationship to CV risk factors, obesity, diabetes, renal disease and sleep disorders. Treatment of high blood pressure and cholesterol

Gruschen Veldtman, MBChB, FRCP, Professor

Leadership Associate Director, AACHD Program; Inpatient Medical Director, AACHD Program

Research Interests The Fontan circulation and end-organ damage; risk stratification in high-risk pregnancy; pulmonary hypertension

Gary Webb, MD, FRCP(C), FACC, FAHA, Professor

Leadership Director, Adolescent and Adult Congenital Heart Disease Program

Research Interests Adolescent and adult congenital heart disease

Ivan Wilmot, MD, Assistant Professor

Research Interests Quality of life in children with heart failure, transplant, and those requiring mechanical circulatory support

Wenying Zhang, MD, PhD, FACMG, Assistant Professor

Leadership Associate Director, Heart Institute Diagnostic Laboratory

Research Interests Clinical molecular genetics; molecular pathology; molecular diagnosis of heritable cardiovascular disease

Joint Appointment Faculty Members

Stuart L. Goldstein, MD, Professor (Nephrology)

Research Interests Acute kidney injury; continuous renal replacement therapy; end-stage renal disease; health-related quality of life

Jeanne James, MD, Associate Professor (Molecular Cardiovascular Biology)

Research Interests Manifestations and etiologies of misfolded protein response and echocardiography

Stephanie Ware, MD, Associate Professor (Molecular Cardiovascular Biology)

Research Interests Cardiovascular Genetics

Jessica Woo, MHSA, PhD, Associate Professor (Biostatistics and Epidemiology)

Research Interests Molecular epidemiology; pediatric obesity; genetics and early life influences on obesity; insulin resistance; dyslipidemia

Clinical Staff Members

- **Chad Connor, MD**

Trainees

- **Ryan Alexy, MD**, PL7, Cincinnati Children's/University of Cincinnati
- **Catherine Allen, MD**, PL5, A.I. duPont Children's
- **Konstantin Averin, MD**, PL5, Cincinnati Children's/University of Cincinnati
- **Nicole Brown, MD**, PL8, Children's Hospital of Pittsburgh, UPMC
- **Joshua Daily, MD**, PL5, Cincinnati Children's/University of Cincinnati
- **Saul Flores, MD**, PL7, Rainbow Babies; Cincinnati Children's
- **Karine Guerrier, DO**, PL5, Cleveland Clinic
- **Eunice Hahn, MD**, PL7, Columbia and Cornell University Medical Centers
- **Brandon Hays, MD**, PL4, University of Minnesota
- **David Kwiatkowski, MD**, PL6, Cincinnati Children's/University of Cincinnati
- **Benjamin Landis, MD**, PL6, Columbia University
- **Sean Lang, MD**, PL7, Yale School of Medicine

- **Lane Lanier, MD**, PL7, UT Southwestern Medical Center
- **Namheon Lee, PhD**, University of Cincinnati
- **Ryan Moore, MD**, PL6, Cincinnati Children's/University of Cincinnati
- **Charu Munjal, PhD**, University of Louisville
- **John J. Parent, MD**, PL7, Indiana University School of Medicine
- **Preeti Ramachandran, MD**, PL4, Cincinnati Children's/University of Cincinnati
- **Kazuyoshi Saito, MD**, Toyama Medical University, Japan
- **James Starc, MD**, PL4, Mt. Sinai School of Medicine
- **Tuerdi Subati, MD, PhD**, York University, Canada
- **Aashoo Tandon, MD**, PL6, Cincinnati Children's/University of Cincinnati
- **Tamara Thomas, MD**, PL7, Vanderbilt University
- **Alex Verhoeven, MD**, PL4, Vanderbilt University Medical Center
- **Chet Villa, MD**, PL7, Cincinnati Children's/University of Cincinnati

Division Collaboration

Studying immune profiling, especially thymic markers, in patients who have received cardiac transplants and correlation with rates of infection, rejection, and death. (Chesney Castleberry, MD)

Allergy and Immunology » Kimberly A. Risma, MD, PhD

Prospective study measuring oxidized LDL and thromboxane as a marker of inflammation in children with obesity and diabetes. (John Lynn Jefferies, MD, MPH)

Anesthesiology » Nick Pratap, MB Chir, MRCPCH, FRCA

Studying genetic aspects of arrhythmogenic cardiomyopathy. (Jeffrey A. Towbin, MD)

Biomedical Informatics » Bruce Aronow, PhD

Human Genetics » Lisa J. Martin, PhD

Studying cardiovascular morbidity and mortality after 40 years follow-up in international cohorts. (Elaine M. Urbina, MD, MS)

Biostatistics and Epidemiology » Jessica Woo, MHSA, PhD

Evaluation of pulmonary hypertension in transplant mediated micro-angiopathy. (Russel Hirsch, MD)

Bone Marrow Transplantation and Immune Deficiency » Christopher E. Dandoy, MD and Sonata Jodele, MD

Multiple projects including biomarkers in bone marrow failure, cardiac function and imaging in chemotherapy exposure. (Thomas D. Ryan, MD, PhD, John Lynn Jefferies, MD, MPH)

Bone Marrow Transplantation and Immune Deficiency » Christopher E. Dandoy, MD, Stella M. Davies, MBBS, PhD, MRCP, Sonata Jodele, MD, and Kasiani C. Myers, MD

Oncology » Michael J. Absalon, MD, PhD and Rajaram Nagarajan, MD, MS

Studying St2, a novel biomarker predictive of multiple cardiovascular related outcomes. This data is being used to predict morbidity and mortality, specifically TMA. (John Lynn Jefferies, MD, MPH)

Bone Marrow Transplantation and Immune Deficiency » Christopher E. Dandoy, MD, Stella M. Davies, MBBS, PhD, MRCP, Sonata Jodele, MD, and Kasiani C. Myers, MD

Assessing echocardiographic parameters in patients with Diamond-Blackfan syndrome. (John Lynn Jefferies, MD, MPH)

Bone Marrow Transplantation and Immune Deficiency » Kasiani C. Myers, MD

Studying the impact of hybrid procedure for HLHS on acute kidney injury (AKI) and the incidence of chronic kidney disease in patients experiencing AKI after cardiopulmonary bypass. (David S. Cooper, MD, MPH, Nicolas Madsen, MD, MPH)

Center for Acute Care Nephrology (CACN) » Stuart L. Goldstein, MD

Studying the effect of acute kidney injury (AKI) on milrinone pharmacokinetics. (David S. Cooper, MD, MPH)

Center for Acute Care Nephrology (CACN) » Stuart L. Goldstein, MD

Clinical Pharmacology » Alexander A. Vinks, PharmD, PhD, FCP

Studying biomarkers in patients with cardiomyopathy and heart failure, specifically using novel urinary biomarkers to predict future chronic kidney disease and acute kidney injury. Also using a biomarker panel to assess future risk of cardiovascular disease and chronic kidney disease in children with Duchene's Muscular Dystrophy (DMD).

Another study is utilizing existing and novel biomarkers to assess for concomitant renal dysfunction and injury in patients with cardiomyopathy. (Jeffrey A. Towbin, MD, John Lynn Jefferies, MD, MPH)

Center for Acute Care Nephrology (CACN) » Stuart L. Goldstein, MD

Ongoing collaborative work that is focused on the functional cardiovascular findings in the fetus with extracardiac anomalies. Specifically, work is being done on the correlation of ventricular function and brain perfusion pre- and post-intervention for twin-twin transfusion syndrome (James F. Cnota, MD, Christopher J. Statile, MD)

Cincinnati Fetal Center » Foong-Yen Lim, MD, Mounira Habli, MD, and William J. Polzin, MD

Dr. Hanke has been closely collaborating with Dr. Iyer and his entire HNCC quality improvement team focused on hospital-wide readmission reduction. He currently serves on the leadership team and has provided both content and quality improvement expertise to the design of the hospital's readmission reduction interventions. This is an ongoing collaboration that meets weekly for the leadership team and monthly with other units hospital wide including: NICU, Hospital Medicine and Neurology. (Samuel Hanke, MD, MS)

Emergency Medicine » Srikant B. Iyer, MD, MPH

Researching the incidence of elevated left ventricular mass in severely obese toddlers and preschoolers. (Holly M. Ippisch, MD, MS)

Endocrinology » Nancy A. Crimmins, MD

Studying Vitamin D levels and arterial stiffness. (Elaine M. Urbina, MD, MS)

Endocrinology » Lawrence M. Dolan, MD

We are investigating the effects of diabetes on the cardiovascular system, specifically the progression of atherosclerosis in youth with obesity and T2DM and arterial stiffness in T1DM. (Thomas R. Kimball, MD, Elaine M. Urbina, MD, MS)

Endocrinology » Lawrence M. Dolan, MD and Amy Shah, MD

Studying arterial stiffness in Turner syndrome. (Elaine M. Urbina, MD, MS)

Endocrinology » Philippe F. Backeljauw, MD and Sarah Lawson, MD

Studying arterial Stiffness & HDL proteomics, as well as cerebrovascular and arterial change in T2DM. (Elaine M. Urbina, MD, MS)

Endocrinology » Amy Shah, MD

Studying cardiac findings in patients with epidermolysis bullosa. (Thomas D. Ryan, MD, PhD, John Lynn Jefferies, MD, MPH)

Epidermolysis Bullosa Center » Anne W. Lucky, MD

Evaluation of the impact of nutritional status in patients with epidermolysis bullosa. (John Lynn Jefferies, MD, MPH)

Epidermolysis Bullosa Center » Anne W. Lucky, MD

Studying desmoplakin mutation, resulting in epidermolysis bullosa and cardiomyopathy. (John Lynn Jefferies, MD, MPH)

Epidermolysis Bullosa Center » Anne W. Lucky, MD

Studying cardiovascular complications in sickle cell disease. (Enkhsaikhan Purevjav, MD, PhD, Jeffrey A. Towbin, MD)

Experimental Hematology and Cancer Biology-Molecular Genetics » Punam Malik, MD

Researching the presentation and management of hypertriglyceridemia-induced pancreatitis in children at Cincinnati Children's. (Holly M. Ippisch, MD, MS)

Gastroenterology, Hepatology and Nutrition » Maisam A. Abu-El-Haija, MD and Stavra A. Xanthakos, MD, MS

A clinical research study investigating cirrhotic cardiomyopathy in end-stage liver disease and liver transplantation. (John Lynn Jefferies, MD, MPH)

Gastroenterology, Hepatology and Nutrition » Monique Goldschmidt, MD and James E. Heubi, MD

Studying the effect of Antithrombin 3 on anticoagulation during mechanical circulatory support. (David S. Cooper, MD, MPH)

Hematology » Joseph S. Palumbo, MD

Clinical Pharmacology » Alexander A. Vinks, PharmD, PhD, FCP

Pediatric General and Thoracic Surgery » Jason S. Frischer, MD

Quality improvement project improving imaging and reporting of coronary arteries within the echo lab. (Christopher J. Statile, MD)

Hospital Medicine » Angela Statile, MD, Med

Collaboration and research efforts investigating Familial Hypercholesterolemia. The aims of this current effort are threefold: (1) Estimate the prevalence of FH in a pediatric population and further characterize the population by defining other diagnoses, medical therapy, and type of managing healthcare provider; (2) Examine attitudes and knowledge among parents of children with FH and identify patterns of associated factors; (3) Determine if an FH

educational brochure is associated with increased knowledge or interest in genetic counseling and testing or intended cholesterol screening uptake. (Erin Miller, MS, LGC)

Human Genetics » Melanie F. Myers, PhD

A Phase 3/4, prospective, multinational, open-label, noninferiority study of alglucosidase alfa manufactured at the 160 L and 4000 L Scales in treatment naïve patients with infantile-onset Pompe Disease. Also, a Phase 4, open-label, prospective study in patients with Pompe Disease to evaluate the efficacy and safety of alglucosidase ALFA produced at 4000 L scale. (John Lynn Jefferies, MD, MPH)

Human Genetics » Nancy Doan Leslie, MD

Investigation of cardiomyopathy and speech delays in individuals with 1p36 Deletion Syndrome by evaluating genetic, echocardiography, and EKG data. (John Lynn Jefferies, MD, MPH)

Human Genetics » Robert J. Hopkins, MD

A cross-sectional study of renal function in treatment-naïve, young male patients with Fabry Disease. (John Lynn Jefferies, MD, MPH)

Human Genetics » Robert J. Hopkins, MD

Using advanced imaging studying to assess coronary artery disease in individuals with Mucopolysaccharidoses. (John Lynn Jefferies, MD, MPH)

Human Genetics » Carlos E. Prada, MD

Studying activated T cell therapy in murine model of cardiac transplantation. (Chesney Castleberry, MD)

Immunobiology » Jonathan D. Katz, PhD

The National Pediatric Cardiology Quality Improvement Collaborative is a Learning Network of 50 pediatric cardiology centers working together and with families to improve the quality of life for children with complex congenital heart disease. (Jeffrey Anderson, MD, MPH, MBA, Robert Beekman III, MD)

James M. Anderson Center for Health Systems Excellence » Carole Lannon, MD, MPH

Studying fish oil supplementation in pregnancy. (Elaine M. Urbina, MD, MS)

Section of Neonatology, Perinatal and Pulmonary Biology » Christina J. Valentine, MD

Studying blood pressure in CKD. (Elaine M. Urbina, MD, MS)

Nephrology and Hypertension » David K. Hooper, MD, MS

Multicenter, NIH funded study assessing cardiovascular implications of end-stage renal disease (ESRD) in children. Multiple longitudinal projects, including assessment of cardiovascular findings in patients with ESRD at CCHMC, focusing on aortic changes. Another project is studying the effects of chronic kidney disease on the cardiovascular system, specifically aortopathy and arterial stiffness in chronic kidney disease. (Thomas R. Kimball, MD, John Lynn Jefferies, MD, MPH, Peace Madueme, MD, MS, Elaine M. Urbina, MD, MS)

Nephrology and Hypertension » Jens W. Goebel, MD, Ahmad Kaddourah, MD, and Mark M. Mitsniefes, MD

Studying urinary neutrophil gelatinase associated lipocalin (NGAL) levels in patients undergoing ultrafiltration for acute decompensated heart failure. (John Lynn Jefferies, MD, MPH)

Nephrology and Hypertension » Prasad Devarajan, MD

Assessment of changes in novel urinary biomarkers in a mouse model of acute kidney injury receiving serelaxin, a novel heart failure drug. (John Lynn Jefferies, MD, MPH)

Nephrology and Hypertension » Michael R. Bennett, PhD and Prasad Devarajan, MD

Dr. Taylor is working on our VAD national trials to improve stroke care in patients that have had an adverse event on mechanical support. (Angela Lorts, MD)

Nephrology and Hypertension » J. Michael Taylor, MD

This collaboration focuses on the Duchenne Muscular Dystrophy (DMD) population. One study assesses multiple biomarkers, including cardiac, for characterization of non-ambulatory patients. Another study focuses on genotype-phenotype correlations in patients with dystrophinopathies.) (Thomas D. Ryan, MD, PhD, John Lynn Jefferies, MD, MPH)

Neurology » Brenda Wong, MD, MBBS

Using advanced echocardiographic techniques to identify cardiotoxicity in patients receiving anthracyclines and/or cisplatin. Also assessing echocardiographic parameters in patients with hepatoblastoma. (John Lynn Jefferies, MD, MPH)

Oncology » James I. Geller, MD and Rajaram Nagarajan, MD, MS

A Phase I pilot study of CPX-351 for children, adolescents and young adults with recurrent or refractory hematologic malignancies. They are using cardiac biomarkers to assess for cardiotoxicity after receiving this novel AML therapy. (John Lynn Jefferies, MD, MPH)

Oncology » Michael J. Absalon, MD, PhD

Determination of impact of chronic liver failure on endothelial dysfunction. (Russel Hirsch, MD)

Pediatric Liver Care Center » John C. Bucuvalas, MD

Pediatric General and Thoracic Surgery » Gregory M. Tiao, MD

The Clinical Trials in Organ Transplantation in Children (CTOT-C) project is a cooperative research program sponsored by the National Institute of Allergy and Infectious Diseases (NIAID). Dr. Carlos Esquivel at Stanford University is the principal investigator for The Biomarkers for Post-Transplant Lymphoproliferative Disorder (PTLD) in Children project. Dr. Chin is the CCHMC site primary investigator who will be working in collaboration with Dr. Bucuvalas. The primary objective is to assess viral and immunologic biomarkers associated with the development and progression of PTLD after transplantation. (Clifford Chin, MD, Angela Lorts, MD)

Pediatric Liver Care Center » John C. Bucuvalas, MD

Collaboration with the VAD team to solidify preparedness planning for VAD patients. They are publishing on the difficult ethical decisions that accompany these complex patients. (Angela Lorts, MD)

Pediatric Palliative and Comfort Care » Norbert J. Weidner, MD

Ethics Center » Armand H. Antommaria, MD, PhD

Studying cardiac MRI imaging and phenotype correlation in animal models of cardiomyopathies. (Enkhsaikhan Purevjav, MD, PhD, Jeffrey A. Towbin, MD)

Radiology » Janaka P. Wansapura, PhD

Studying cardiac MRI in neuromuscular disease. (Jeffrey A. Towbin, MD)

Radiology » Robert J. Fleck, MD

Conducting a comprehensive follow-up study of adolescent bariatric surgery at 5+ years post-operatively. The goal is to compare the long-term medical and psychosocial outcomes of surgical and non-surgical management of adolescents with severe obesity. This long-term is designed to provide the first systematic prospective data collection for a cohort of severely obese adolescents over a 5+ year period. (Shelley Kirk, PhD, RD, LD, Elaine M. Urbina, MD, MS)

Pediatric General and Thoracic Surgery » Thomas H. Inge, MD, PhD

Studying the outcome of multi-run ECMO. (David S. Cooper, MD, MPH)

Pediatric General and Thoracic Surgery » Jason S. Frischer, MD

Shared project looking at sleep studies in patients with congenital prolonged QT syndrome and with HCM. (Timothy Knilans, MD, John Lynn Jefferies, MD, MPH)

Pulmonary Medicine » Abu Syed Mohammad Shamsuzzaman, MD

Studying arterial structure & function in sleep apnea. (Elaine M. Urbina, MD, MS)

Pulmonary Medicine » Raouf S. Amin, MD

Role of placental angiogenesis in fetal growth restriction associated with congenital heart disease. (James F. Cnota, MD, Robert Hinton, Jr, MD)

Reproductive Services » Helen N. Jones, PhD

Physical activity in the congenital heart disease population. (Nicolas L. Madsen, MD, MPH, Robert Siegel, MD, Elaine M. Urbina, MD, MS)

Sports Medicine » Nicholas M. Edwards, MD, MPH

Grants, Contracts, and Industry Agreements

Grant and Contract Awards	Annual Direct
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CASTLEBERRY, C

mTOR Inhibitors in Pediatric Heart Transplantation: Do the Potential Benefits Outweigh the Risks, Clues from the ISHLT Registry

International Society of Heart & Lung Transplantation

04/01/13-12/31/13

\$5,000

CNOTA, J

Pediatric Heart Network Prairieland Consortium

National Institutes of Health

U10 HL 109673

09/01/11-06/30/16

\$467,005

Trial of Beta Blocker vs Angiotensin II Receptor Blocker Therapy in Individuals with Marfan Syndrome

National Institutes of Health(New England Research Institutes)

U01 HL 068270	05/05/11-12/31/14	\$16,148
GOLDSTEIN, B		
Evaluation of Growth and Puberty in Adolescent and Young Adult Fontan Survivors		
National Institutes of Health(University of Utah)		
U10 HL 068270	07/01/13-06/30/14	\$4,455
Utilizing Vascular Function to Predict Functional Outcomes in Fontan Survivors		
American Heart Association		
14BGIA18740061	01/01/14-12/31/15	\$59,999
HINTON, R		
Genetic Regulation of Thoracic Aortic Aneurysm Progression		
American Heart Association		
14IRG18830027	01/01/14-12/31/15	\$68,182
HIRSCH, R		
Covered Cheatham Platinum Stent for Prevention or Treatment of Aortic Wall Injury Associated with Aortic Coarctation		
Food and Drug Administration(The Johns Hopkins University)		
R01 FD 003898	06/22/11-05/31/15	\$4,203
KWIATKOWSKI, D		
Early Renal Replacement Therapy vs. Furosemide for Neonates with Oliguria After Cardiopulmonary Bypass		
American Heart Association		
13POST1624002	07/01/13-06/30/15	\$50,000
MARINO, B		
Transitional Telehealth Homecare: REACH		
National Institutes of Health(Children's Hospital of Philadelphia)		
R01 NR 002093	09/21/11-06/30/16	\$103,225
MICHELFELDER, E		
Hypoplastic Left Heart Syndrome: Expression of RHD in the Fetus?		
National Institutes of Health(Washington University)		
R01 HL 098634	05/15/11-12/31/14	\$34,672
TOWBIN, J		
Biomarkers in Pediatric Cardiomyopathy		
National Institutes of Health(University of Miami)		
R01 HL 109090	08/06/12-07/31/16	\$34,637
Genetics, Mechanisms and Clinical Phenotypes of Arrhythmogenic Cardiomyopathy		
National Institutes of Health		
R01 HL 116906	08/23/13-06/30/18	\$1,858,137

URBINA, E**Accelerated CV Aging in Youth Related to CV Risk Factor Clusters**

National Institutes of Health

R01 HL 105591	01/01/11-12/31/15	\$354,654
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Early Identification of World Trade Center Conditions in Adolescents

Centers for Disease Control and Prevention(New York University School of Medicine)

U01 OH 010394	07/01/13-06/30/16	\$18,000
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Pediatric HIV/AIDS Cohort Study

National Institutes of Health(Tulane University)

U01 HD 052104	08/01/13-07/31/15	\$12,490
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WEBB, G**ACHD Conference**

Medtronic, Inc.

06/10/14-06/09/15	\$10,000
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Current Year Direct	\$3,100,807
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Industry Contracts**BEEKMAN, R**

The Johns Hopkins University	\$6,044
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COOPER, D

Cadence Pharmaceuticals, Inc.	\$67,783
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HIRSCH, R

AGA Medical , LLC	\$2,832
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Lilly USA, LLC	\$8,701
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Pfizer, Inc.	\$3,608
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United Therapeutics Corporation	\$3,015
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KRAWCZESKI, C

Asklepion Pharmaceuticals, LLC	\$17,500
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LORTS, A

SynCardia Systems, Inc.	\$29,391
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Current Year Direct Receipts	\$138,874
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Total	\$3,239,681
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