

# Cardiology

## Division Details

### RESEARCH AND TRAINING DETAILS

Faculty	45
Joint Appointment Faculty	3
Total Annual Grant Award Dollars	\$5,545,328
Total Annual Industry Award Dollars	\$604,252
Total Publications	166

### CLINICAL ACTIVITIES AND TRAINING

Staff Physicians	1
Clinical Fellows	23
Inpatient Encounters	1,075
Outpatient Encounters	18,661

## Significant Publications

**Anderson JB, Beekman RH 3rd, Kugler JD, Rosenthal GL, Jenkins KJ, Klitzner TS, Martin GR, Neish SR, Brown DW, Mangeot C, King E, Peterson LE, Provost L, Lannon C, National Pediatric Cardiology Quality Improvement C. [Improvement in Interstage Survival in a National Pediatric Cardiology Learning Network](#). *Circ Cardiovasc Qual Outcomes*. 2015 Jul;8(4):428-36.**

This project demonstrated for the first time the power of a collaborative learning network to improve outcomes in infants with a rare disease, hypoplastic left heart syndrome (HLHS). Using improvement methods, and the participation of 60 surgical centers, the National Pediatric Cardiology Quality Improvement Collaborative (NPC-QIC) demonstrated improved discharge and outpatient clinic processes that resulted in a 40% reduction in interstage mortality in patients with HLHS in this collaborative. This work was the result of collaboration among cardiologists, nurses, and parents to co-produce improvement efforts that ultimately resulted in these improved outcomes.

Niss O, Quinn CT, Lane A, Daily J, Khoury PR, Bakeer N, **Kimball TR, Towbin JA, Malik P, Taylor MD. [Cardiomyopathy With Restrictive Physiology in Sickle Cell Disease](#). *JACC Cardiovasc Imaging*. 2016 Mar;9(3):243-52.**

Research associates sickle cell disease with significant cardiopulmonary morbidity, including an increased rate of unexplained sudden death. This paper describes a novel paradigm for the cardiac involvement in sickle cell disease as a form of cardiomyopathy with a restrictive phenotype. The paper has two parts: an extensive meta-analysis of previously published cardiac imaging studies, and a detailed echocardiographic study from our institution that together show the unique high output restrictive physiology in this

patient group. These findings are the preliminary data used to design the current clinical trial of cardiac MR and serologic biomarkers to further delineate the cardiopulmonary phenotype of sickle cell disease.

**Towbin JA, Lorts A, Jefferies JL. Left ventricular non-compaction cardiomyopathy.** *Lancet*. 2015 Aug 22;386(9995):813-25.

This report provides a contemporary understanding of left ventricular noncompaction cardiomyopathy (LVNC). The disease is increasingly diagnosed in clinical practice, but many clinicians are not familiar with the diagnosis or treatment. LVNC represents a unique type of cardiomyopathy that can have associated findings, such as heart failure and significant arrhythmias, which can lead to morbidity and mortality. Our paper provides care providers, and researchers, with a comprehensive description of the disease along with potential strategies for clinical surveillance and management.

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## Division Publications

1. Alsaied T, Alsidawi S, Allen CC, Faircloth J, Palumbo JS, Veldtman GR. **Strategies for Thromboprophylaxis in Fontan Circulation: A Meta-Analysis.** Editor, M Rothenberg. *Heart*. 2015; 101:1731-7.
2. Alsaied T, Khan MS, Cnota J. **Database Research for Fellows-in-Training and Early-Career Cardiologists.** *J Am Coll Cardiol*. 2015; 66:1404-7.
3. Alsaied T, Marino B, Esbensen A, Anixt J, Epstein J, Cnota J. **Does Congenital Heart Disease Affect Neurodevelopmental Outcomes in Children with Down Syndrome?** *Congenit Heart Dis*. 2016; 11:26-33.
4. Alsaied T, Vasili Y, Moore R, McPhaul J, Javarayee P, Beekman R. **Asymmetric Pulses in a 5-Year-Old Asian Female: Is It Worth Further Investigations?** *Clin Pediatr (Phila)*. 2016; 55:192-95.
5. Alten JA, Rhodes LA, Tabbutt S, Cooper DS, Graham EM, Ghanayem N, Marino BS, Figueroa MI, Chanani NK, Jacobs JP, Donohue JE, Yu S, Gaies M. **Perioperative Feeding Management of Neonates with Chd: Analysis of the Pediatric Cardiac Critical Care Consortium (Pc4) Registry.** *Cardiol Young*. 2015; 25:1593-601.
6. Anand V, Kwiatkowski DM, Ghanayem NS, Axelrod DM, DiNardo J, Klugman D, Krishnamurthy G, Siehr S, Stromberg D, Yates AR, Roth SJ, Cooper DS. **Training Pathways in Pediatric Cardiac Intensive Care: Proceedings from the 10th International Conference of the Pediatric Cardiac Intensive Care Society.** *World J Pediatr Congenit Heart Surg*. 2016; 7:81-8.
7. Anderson JB, Beekman RH. **Addressing Nutrition and Growth in Children with Congenital Heart Disease.** In: PR Barach, JP Jacobs, SE Lipshultz, PC Laussen, eds. *Pediatric and Congenital Cardiac Care: Volume 2: Quality Improvement and Patient Safety*. New York: Springer; 2015:153-64.
8. Anderson JB, Beekman RH, 3rd, Kugler JD, Rosenthal GL, Jenkins KJ, Klitzner TS, Martin GR, Neish SR, Brown DW, Mangeot C, King E, Peterson LE, Provost L, Lannon C, National Pediatric Cardiology Quality Improvement C. **Improvement in Interstage Survival in a National Pediatric Cardiology Learning Network.** *Circ Cardiovasc Qual Outcomes*. 2015; 8:428-36.
9. Anderson JB, Willis M, Lancaster H, Leonard K, Thomas C. **The Evaluation and Management of Pediatric Syncope.** *Pediatr Neurol*. 2016; 55:6-13.
10. Apers S, Kovacs AH, Luyckx K, Thomet C, Budts W, Enomoto J, Sluman MA, Wang JK, Jackson JL, Khairy P, Cook SC, Chidambarathanu S, Alday L, Eriksen K, Dellborg M, Berghammer M, Mattsson E, Mackie AS, Menahem S, Caruana M, et al. **Quality of Life of Adults with Congenital Heart Disease in 15 Countries: Evaluating Country-Specific Characteristics.** *J Am Coll Cardiol*. 2016; 67:2237-45.
11. Averin K, Michelfelder E, Sticka J, Cash M, Hirsch R. **Changes in Ventricular Geometry Predict Severity of Right Ventricular Hypertension.** *Pediatr Cardiol*. 2016; 37:575-81.
12. Averin K, Villa C, Krawczeski CD, Pratt J, King E, Jefferies JL, Nelson DP, Cooper DS, Ryan TD, Sawyer J, Towbin JA, Lorts A. **Initial Observations of the Effects of Calcium Chloride Infusions in Pediatric Patients with Low Cardiac Output.** *Pediatr Cardiol*. 2016;

13. Bainbridge MN, Davis EE, Choi WY, Dickson A, Martinez HR, Wang M, Dinh H, Muzny DM, Pignatelli R, Katsanis N, Boerwinkle E, Gibbs RA, Jefferies JL. **Loss of Function Mutations in Nnt Are Associated with Left Ventricular Noncompaction.** *Circ Cardiovasc Genet.* 2015; 8:544-52.
14. Banerjee R, D'Souza G, Lee N, Taylor M. **Comparison of Pulmonary Arterial Wall Properties between a Congenital Heart Disease Patient and a Normal Subject Using in Vivo Pressure–Diameter Measurements: A Feasibility Study.** *Prog Pediatr Cardiol* 2016; 41:97-102.
15. Beekman III RH, Goldstein BH, Hirsch R, Whiteside W, Zampi J. **Transcatheter Therapies for Congenital Heart Disease.** In: EJ Topol, PS Teirstein, eds. *Textbook of Interventional Cardiology, Seventh Edition.* Philadelphia, PA: Elsevier Health Sciences; 2015:874-900.
16. Blume ED, Rosenthal DN, Rossano JW, Baldwin JT, Eghtesady P, Morales DL, Cantor RS, Conway J, Lorts A, Almond CS, Naftel DC, Kirklin JK, PediMACS Investigators. **Outcomes of Children Implanted with Ventricular Assist Devices in the United States: First Analysis of the Pediatric Interagency Registry for Mechanical Circulatory Support (Pedimacs).** *J Heart Lung Transplant.* 2016; 35:578-84.
17. Bokma JP, Burchill LJ, Kovacs AH, Oechslin EN, Khairy P, Mulder BJ, Veldtman GR. **Fellows' Perspectives on Training in Adult Congenital Heart Disease: Results of a Survey from the International Society for Adult Congenital Heart Disease (Isachd).** *Int J Cardiol.* 2016; 202:253-5.
18. Brown DW, Mangeot C, Anderson JB, Peterson LE, King EC, Lihn SL, Neish SR, Fleishman C, Phelps C, Hanke S, Beekman RH, Lannon CM, National Pediatric Cardiology Quality Improvement Collaborative. **Digoxin Use Is Associated with Reduced Interstage Mortality in Patients with No History of Arrhythmia after Stage I Palliation for Single Ventricle Heart Disease.** *J Am Heart Assoc.* 2016; 5.
19. Burchill LJ, Redington AN, Silversides CK, Ross HJ, Jimenez-Juan L, Mital S, Oechslin EN, Dragulescu A, Slorach C, Mertens L, Wald RM. **Renin-Angiotensin-Aldosterone System Genotype and Serum Bnp in a Contemporary Cohort of Adults Late after Fontan Palliation.** *Int J Cardiol.* 2015; 197:209-15.
20. Burkett DA, Slorach C, Patel SS, Redington AN, Ivy DD, Mertens L, Younoszai AK, Friedberg MK. **Left Ventricular Myocardial Function in Children with Pulmonary Hypertension: Relation to Right Ventricular Performance and Hemodynamics.** *Circ Cardiovasc Imaging.* 2015; 8.
21. Cai S, Rohailla S, Peng J, Zeng C, Li J, Redington A. **Loss of Tlr4 in a Murine Model of Left Anterior Descending Myocardial Infarction Modifies Early Remodeling, but Does Not Provide Long-Term Benefit.** *Int J Cardiol.* 2016; 212:118-20.
22. Campbell MJ, Czosek RJ, Hinton RB, Miller EM. **Exon 3 Deletion of Ryanodine Receptor Causes Left Ventricular Noncompaction, Worsening Catecholaminergic Polymorphic Ventricular Tachycardia, and Sudden Cardiac Arrest.** *Am J Med Genet A.* 2015; 167A:2197-200.
23. Campbell MJ, Zhou X, Han C, Abrishami H, Webster G, Miyake CY, Knilans TK, Czosek RJ. **Electrocardiography Screening for Hypertrophic Cardiomyopathy.** *Pacing Clin Electrophysiol.* 2016; 39:944-50.
24. Castleberry C, Zafar F, Thomas T, Khan M, Bryant R, Chin C, Morales D, Lorts A. **Allosensitization Does Not Alter Post-Transplant Outcomes in Pediatric Patients Bridged to Transplant with a Ventricular Assist Device.** *Pediatr Transplant.* 2016; 20:559-64.
25. Castleberry CD, Zafar F, Dipchand AI, Edwards L, Chin C. **Early Initiation of Mtor Inhibitors in Children with Heart Transplantation: A Propensity-Based Registry Analysis.** *J Heart Lung Transplant.* 2016; 35:253-5.
26. Chaouki AS, Czosek RJ, Spar DS. **Missing Link: Extrusion of a New-Generation Implantable Loop Recorder in a Child.** *Cardiol Young.* 2016; 26:1445-7.

27. Chen CK, Cifra B, Morgan GJ, Sarkola T, Slorach C, Wei H, Bradley TJ, Manlhiot C, McCrindle BW, Redington AN, Benson LN, Mertens L. **Left Ventricular Myocardial and Hemodynamic Response to Exercise in Young Patients after Endovascular Stenting for Aortic Coarctation.** *J Am Soc Echocardiogr.* 2016; 29:237-46.
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31. Cooper D, Morales D, Corral M, Paden M, Thiagarajan R. **Databases for Extracorporeal Membrane Oxygenation and Ventricular Assist Devices.** In: PR Barach, JP Jacobs, SE Lipshultz, PC Laussen, eds. *Pediatric and Congenital Cardiac Care: Volume 1: Outcomes Analysis.* New York: Springer; 2015:211-18.
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37. Czosek RJ, Jefferies JL, Khoury PR, Anderson JB, Wilmot I, Knilans TK, Spar DS. **Arrhythmic Burden and Ambulatory Monitoring of Pediatric Patients with Cardiomyopathy.** *Pacing Clin Electrophysiol.* 2016; 39:443-51.
38. Czosek RJ, Kaltman JR, Cassidy AE, Shah MJ, Vetter VL, Tanel RE, Wernovksy G, Wray J, Marino BS. **Quality of Life of Pediatric Patients with Long Qt Syndrome.** *Am J Cardiol.* 2016; 117:605-10.
39. Czosek RJ, Spar DS, Anderson JB, Knilans TK, Marino BS. **Cardiac Conduction, Dysrhythmias, and Pacing.** In: DH Shaffner, DG Nichols, eds. *Rogers' Textbook of Pediatric Intensive Care, Fifth Edition.* Lippincott Williams & Wilkins: Philadelphia PA; 2015:1214-40.
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41. Devarajan P, Jefferies JL. **Progression of Chronic Kidney Disease after Acute Kidney Injury.** *Prog Pediatr Cardiol.* 2016; 41:33-40.
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44. Drogalis-Kim D, Jefferies J, Wilmot I, Alejos J. **Right Sided Heart Failure and Pulmonary Hypertension: New Insights into Disease Mechanisms and Treatment Modalities.** *Prog Pediatr Cardiol.* 2016.
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47. Flores S, Daily J, Pratap JN, Cash MC, Hirsch R. **Management of a Child with Pulmonary Arterial Hypertension Presenting with Systemic Hypertension.** *Cardiol Young.* 2016; 26:378-81.
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49. Ford MA, Gauvreau K, McMullan DM, Almodovar MC, Cooper DS, Rycus PT, Thiagarajan R. **Factors Associated with Mortality in Neonates Requiring Extracorporeal Membrane Oxygenation for Cardiac Indications: Analysis of the Extracorporeal Life Support Organization Registry Data.** *Pediatr Crit Care Med.* 2016; 17:860-70.
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53. Gao Z, Khoury P, Mccoy C, Shah A, Kimball T, Dolan L, Urbina E. **Adiposity Has No Direct Effect on Carotid Intima-Media Thickness in Adolescents and Young Adults: Use of Structural Equation Modeling to Elucidate Indirect & Direct Pathways.** *Atherosclerosis.* 2016; 246:29-35.
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56. Goldstein BH, Holzer RJ, Trucco SM, Porras D, Murphy J, Foerster SR, El-Said HG, Beekman RH, 3rd, Bergersen L. **Practice Variation in Single-Ventricle Patients Undergoing Elective Cardiac Catheterization: A Report from the Congenital Cardiac Catheterization Project on Outcomes (C3po).** *Congenit Heart Dis.* 2016; 11:122-35.
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58. Guerrier K, Madueme PC, Jefferies JL, Anderson JB, Spar DS, Knilans TK, Czosek RJ. **Unexpectedly Low Left Ventricular Voltage on Ecg in Hypertrophic Cardiomyopathy.** *Heart.* 2016; 102:292-7.
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60. Guerrier K, Shamszad P, Czosek RJ, Spar DS, Knilans TK, Anderson JB. **Variation in Antiarrhythmic Management of Infants Hospitalized with Supraventricular Tachycardia: A Multi-Institutional Analysis.** *Pediatr Cardiol.* 2016; 37:946-52.

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62. Hehir DA, Easley RB, Byrnes J. **Noncardiac Challenges in the Cardiac Icu: Feeding, Growth and Gastrointestinal Complications, Anticoagulation, and Analgesia.** *World J Pediatr Congenit Heart Surg.* 2016; 7:199-209.
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67. Iliopoulos I, Branco RG, Brinkhuis N, Furck A, LaRovere J, Cooper DS, Pathan N. **Mesenteric near-Infrared Spectroscopy and Risk of Gastrointestinal Complications in Infants Undergoing Surgery for Congenital Heart Disease.** *Cardiol Young.* 2016; 26:772-80.
68. Iliopoulos I, Nelson D. **Cardiopulmonary Interactions in Adults and Children with Congenital Heart Disease.** *Prog Pediatr Cardiol.* 2015; 39:151-56.
69. Jacobs J, Wernovsky G, Cooper D, Karl T. **Principles of Shared Decision-Making within Teams.** *Cardiol Young.* 2015; 25:1631-36.
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## Grants, Contracts, and Industry Agreements

### Annual Grant Award Dollars

Investigator	Title	Sponsor	ID	Dates	Amount
James F Cnota, MD	Effect of Hydrocortisone on the Cardiac Function of Premature Intubated Infants <30 Weeks Gestational Age	Children's Hospital of Michigan Fdtn (Wayne State University)	WSU16034	4/9/2014 - 12/31/2016	\$10,790
James F Cnota, MD	Pediatric Heart Network Prairieland Consortium	National Institutes of Health	U10 HL109673	9/1/2011 - 6/30/2017	\$460,250
James F Cnota, MD	Long-term Outcomes of Children with Hypoplastic Left Heart Syndrome and the Impact of Norwood Shunt Type	National Institutes of Health (New England Research Institutes)	U10 HL109673	3/31/2014 - 6/30/2017	\$42,900
Richard J Czosek, MD	Exercise in Genetic Cardiovascular Conditions	National Institutes of Health (Yale)	R01 HL125918	2/1/2015 - 1/31/2020	\$22,500
Bryan H Goldstein, MD	Do Serum Biochemical and Hematopoietic and Stool Biomarkers Predict Low Cardiac Index in Patients with Fontan Physiology?	Children's Heart Foundation (Lurie Children's Hospital of Chicago)	NITPDA	1/1/2015 - 12/31/2016	\$6,500
Russel Hirsch, MD	TOPP-2 Registry (Tracking Outcomes and Practice in	Assoc Pediatric Pulmonary Hypertension	TOPP2	8/14/2015 - 8/13/2020	\$58,980

Pediatric Pulmonary  
Hypertension)

Russel Hirsch, MD	Covered Cheatham Platinum Stent for Prevention or Treatment of Aortic Wall Injury Associated with Aortic Coarctation (COAST II)	Food and Drug Administration (Johns Hopkins School of Medicine)	R01 FD003898	6/22/2011 - 5/31/2016	\$2,752
John Lynn Jefferies, MD	Assessment of Quality of Life, Anxiety, and Depression in Barth Syndrome: Expanding the Scope of Comprehensive Care	Barth Syndrome Foundation, Inc.	Jefferies, John, Bar	4/1/2015 - 3/31/2017	\$14,566
John Lynn Jefferies, MD	Cardiac Biomarkers in Pediatric Cardiomyopathy	National Institutes of Health (Wayne State University)	R01 HL109090	7/16/2014 - 6/30/2017	\$24,064
Ashley S Parrott	The Impact of Genetic Variant Reclassification on Patients and Families	National Society of Genetic Counselors	Parrott, Ashley NSGC	2/11/2016 - 11/30/2016	\$500
Robyn Wingard Stamm APRN-CNP	Screening infants with Single Ventricle Physiology for Abnormal Cardiopulmonary Patterns during Sleep	The Gerber Foundation	Stamm_Gerber	11/19/2015 - 12/31/2016	\$20,000
Michael Taylor, MD-PHD	Cardiac Magnetic Resonance Imaging Core Laboratory for Long-term Outcomes of Children with HLHS and the Impact of Norwood Shunt Type (SVR III)	National Institutes of Health (New England Research Institutes)	U10 HL068270	1/1/2015 - 12/31/2016	\$151,605
Jeffrey A Towbin, MD	Genetics, Mechanisms and Clinical Phenotypes of Arrhythmogenic Cardiomyopathy	National Institutes of Health	R01 HL116906	8/23/2013 - 6/30/2016	\$2,350,202
Elaine Urbina, MD	Study of Hypertension in Pediatrics - Adult Hypertension Onset in Youth	American Heart Association	15SFRN23680000	4/1/2015 - 3/31/2019	\$941,443
Elaine Urbina, MD	Search Nutrition Ancillary Study 2	National Institutes of Health (University of Cincinnati)	R01 DK077949	4/15/2014 - 3/31/2018	\$17,734
Elaine Urbina, MD Jessica G Woo, PHD	Childhood CV Risk and Adult CVD Outcomes: an International Long-term Follow-up	National Institutes of Health	R01 HL121230	12/1/2014 - 11/30/2019	\$1,279,814
Elaine Urbina, MD	Validation of Circulating	National Institutes of Health	R01 HL110957	8/1/2013 -	\$63,776

	Endothelial Cells and Microparticles in Youth	(University of Minnesota)		11/30/2016	
Elaine Urbina, MD	The Studies to Treat or Prevent Pediatric Type 2 Diabetes	National Institutes of Health (George Washington University)	U01 DK061230	3/1/2012 - 7/31/2016	\$28,765
Elaine Urbina, MD	Pediatric HIV/AIDS Cohort Study (PHACS 3)	National Institutes of Health (Tulane University)	U01 HD52104	8/1/2015 - 7/31/2020	\$21,411
Elaine Urbina, MD	Early Identification of World Trade Center Conditions in Adolescents	Ctr for Disease Control and Prevention (NYU Sponsored Programs Administration)	U01OH010394	7/1/2013 - 6/30/2016	\$26,776

**Total Annual Grant Award Dollars**

**\$5,545,328**

### **Annual Industry Award Dollars**

<b>Investigator</b>	<b>Industry Sponsor</b>	<b>Amount</b>
Bryan H Goldstein, MD	PFM Medical, Inc.	\$38,125
Karine Guerrier DO	Medtronic, Inc.	\$2,820
Russel Hirsch, MD	United Therapeutics Corporation	\$112,973
Jeanne Foose James, MD	Medtronic, Inc.	\$30,000
John Lynn Jefferies, MD	Capricor ATherapeutics, Inc.	\$134,221
John Lynn Jefferies, MD	MyoKardia, Inc.	\$100,000
Robert M Siegel, MD	VIVUS, Inc.	\$95,502
Chet R Villa, MD	Syncardia Systems, Inc	\$90,611
<b>Total Annual Industry Award Dollars</b>		<b>\$604,252</b>