2014 Research Annual Report

Heart Institute

Cincinnati Children's

Institute Summary

RESEARCH AND TRAINING DETAILS	
Number of Faculty	**64**
Number of Joint Appointment Faculty	**4**
Number of Research Fellows	**37**
Number of Research Students	**17**
Number of Support Personnel	**145**
Direct Annual Grant Support	**$9,575,313**
Direct Annual Industry Support	**$213,898**
Peer Reviewed Publications	**169**

CLINICAL ACTIVITIES AND TRAINING	
Number of Clinical Staff	**189**
Number of Staff Physicians	**1**
Number of Clinical Fellows	**22**
Number of Other Students	**12**
Inpatient Encounters	**1,814**
Outpatient Encounters	**20,369**

Institute Photo

Row 1: H DeSena, W Whiteside, W Zhang, E Purevjav, A Lorts, C Chin, M Maillet, J Towbin, J Robbins, B Siegel, J Anderson, E Hahn, Z Khuchua
Row 2: M Schecter, D Millay, J Waxman, S Kirk, E Urbina, B Blaxall, J Gulick, J Sticka, L Rogers, M Taylor
Row 3: J Jefferies, E Miller, I Iliopoulos, B Goldstein, G Veldtman
Row 4: I Wilmot, T Ryan, C Statile, C Learn, R Czosek, T Knilans, G Webb, E Michelfelder, T Kimball, C Villa

Significant Accomplishments

Can the Heart Regenerate

Worms can do it, tadpoles can do it; can humans? After a heart attack, large areas of heart muscle are lost and replaced with scar tissue. Not only does this tissue not effectively pump blood, it can actively interfere with the remaining, functional muscle tissue. Researchers have been working to replace this scar tissue with new, contracting muscle grown from stem cells. Data in animal models suggest that certain types of cardiac stem cells have the capacity to regenerate damaged heart muscle. However, clinical trials so far have been controversial and the results have been mixed.

Stem cells used in most human clinical trials have been identified by a certain type of protein they produce, known as “c-kit.” Identifying stem cells for subsequent injection into humans on the basis of this marker has been widely used for the clinical trials. Jeffery Molkentin, PhD, and colleagues recently published a paper in the journal *Nature* that definitively shows that endogenous stem cells in the heart, expressing c-kit, are not a significant source for making new heart cells in the mouse. The Molkentin lab used sophisticated genetic tracing techniques to demonstrate that c-kit expressing stem cells have extremely limited capacity to make new cardiac myocytes and contribute to new, beating muscle in the heart, even after heart attack injury. These results suggest that any potential benefit of injecting c-kit cells into the hearts of human patients is unlikely to result from generation of new contracting heart tissue.

3D Heart Disease Models Help With Surgery Planning, Educating Families

During the past year, the Heart Institute developed the infrastructure and expertise to create patient-specific 3-

dimensional (3D) complex congenital heart disease models. These 3D heart models have proven to be extremely valuable to our surgical team for both patient education and pre-surgical planning. In a recent case, a 3D-printed model of a patient with interrupted aortic arch was needed to help understand the basic anatomy prior to surgery. We delivered the model to the surgeon within six hours, and the resulting pre-surgical planning ultimately changed the surgeon's approach. The model also was used to educate the family about their child's heart condition, providing them with additional insight into the surgical approach. We have begun tracking the impact 3D heart modeling has upon surgical outcomes. If there is a proven benefit, this would represent the first evidence to support the paradigm shift of using 3D-printed heart models for routine surgical planning.

In a related project aimed at patients, parents, and trainees, we have created "Heartpedia," the first fully interactive 3D congenital heart disease mobile application. The initial version of this free app contains seven 3D heart models of the most common and complex congenital heart defects. Each defect also has an interactive model of the most commonly used surgical palliation. All the detailed information from the popular Heart Institute Encyclopedia website is contained within the app and it links to the recently updated Heart Encyclopedia animated videos. The primary aim of the Heartpedia app is to help patients and parents understand these defects. We hope it also helps parents share this complex information with relatives and friends. The Heartpedia app is being used clinically for counseling families about diagnoses and therapy options. It has received excellent initial reviews from parents and practitioners. The team responsible for both projects is headed by Michael Taylor, MD; and includes Peace Madueme, MD, (Cardiology faculty); Ryan Moore, MD, (Cardiology fellow); Ken Tegtmeyer, MD, (Critical Care faculty); and Jeff Cimprich (Critical Care Media Lab animator).

Community Outreach: Healthy Kids

More than 40 percent of Norwood Public School children are overweight or obese. With this in mind, we have collaborated with the school system to create the Norwood Schools Center for Better Health and Nutrition Clinic, which opened in November 2013. The clinic delivers a comprehensive pediatric weight management program directly to the students of an entire city. The clinic features the services of a pediatrician, dietitian, exercise physiologist, nurse, and social worker. These complete services are offered twice a month at Norwood High School.

The Norwood clinic is the centerpiece of "Healthy Kids Norwood," a comprehensive initiative between Cincinnati Children's and the City of Norwood to lower obesity rates in children. This effort has several community-based interventions, including Fun2BFit, a youth exercise program offered at the Richard E. Lindner YMCA, and Norwood Grows, an augmentation of the Woven Oak Initiative's student gardening program. Efforts to improve nutrition and activity for Norwood Public School students include redesigning cafeteria serving strategies, cooking classes, student taste testing and 100-mile walking/running clubs. The clinic also issues a "Healthy Kids Norwood" newsletter with health information and activities for the entire community. To further these health efforts outside of school, the clinic has been working to increase the availability of fruits, vegetables and other healthy foods at the Zion Food Pantry and Lydia's House (a residential facility for single parent families in distress). In August 2014, we also launched a pilot Norwood Farmer's Market. All of these initiatives will be monitored closely. Once we know what the most effective approaches are, we can scale those efforts up so that more children in our region are reached.

Division Publications

1. Abonia JP, Wen T, Stucke EM, Grotjan T, Griffith MS, Kemme KA, Collins MH, Putnam PE, Franciosi JP, von Tiehl KF, Tinkle BT, Marsolo KA, Martin LJ, Ware SM, Rothenberg ME. **High prevalence of**

eosinophilic esophagitis in patients with inherited connective tissue disorders. *J Allergy Clin Immunol*. 2013; 132:378-86.

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3. Aldoss O, Arain NI, Vinocur JM, Menk J, Ameduri RK, Bryant R, 3rd, Kochilas LK, Gruenstein DH. **Frequency of superior vena cava obstruction in pediatric heart transplant recipients and its relation to previous superior cavopulmonary anastomosis**. *Am J Cardiol*. 2013; 112:286-91.
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5. 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.
6. Belmonte SL, Ram R, Mickelsen DM, Gertler FB, Blaxall BC. **Cardiac overexpression of Mammalian enabled (Mena) exacerbates heart failure in mice**. *Am J Physiol Heart Circ Physiol*. 2013; 305:H875-84.
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8. Braitsch CM, Kanisicak O, van Berlo JH, Molkentin JD, Yutzey KE. **Differential expression of embryonic epicardial progenitor markers and localization of cardiac fibrosis in adult ischemic injury and hypertensive heart disease**. *J Mol Cell Cardiol*. 2013; 65:108-19.
9. Braitsch CM, Yutzey KE. **Transcriptional Control of Cell Lineage Development in Epicardium-Derived Cells**. *Journal of Developmental Biology*. 2013; 1:92-111.
10. Brown KL, MacLaren G, Marino BS. **Looking beyond survival rates: neurological outcomes after extracorporeal life support**. *Intensive Care Med*. 2013; 39:1870-2.
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12. 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.
13. Burr AR, Millay DP, Goonasekera SA, Park KH, Sargent MA, Collins J, Altamirano F, Philipson KD, Allen PD, Ma J, Lopez JR, Molkentin JD. **Na+ dysregulation coupled with Ca2+ entry through NCX1 promotes muscular dystrophy in mice**. *Mol Cell Biol*. 2014; 34:1991-2002.
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15. 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.
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Grants, Contracts, and Industry Agreements

Cardiology

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

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

Pediatric HIV/AIDS Cohort Study
National Institutes of Health(Tulane University)

U01 HD 052104	08/01/13-07/31/15	$12,490

WEBB, G

ACHD Conference
Medtronic, Inc.

	06/10/14-06/09/15	$10,000

Current Year Direct	**$3,100,807**

Industry Contracts

BEEKMAN, R

The Johns Hopkins University	$6,044

COOPER, D

Cadence Pharmaceuticals, Inc.	$67,783

HIRSCH, R

AGA Medical , LLC	$2,832
Lilly USA, LLC	$8,701
Pfizer, Inc.	$3,608

United Therapeutics Corporation	$3,015

KRAWCZESKI, C

Asklepion Pharmaceuticals, LLC	$17,500

LORTS, A

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

Molecular Cardiovascular Biology

Grant and Contract Awards		Annual Direct

ACCORNERO, F

BEX1 and the Control of Protein Translation in Cardiac Hypertrophy
National Institutes of Health
K99 HL 121284 12/20/13-11/30/15 $121,375

BHUIYAN, S

Sigma-1 Receptor and Cardioprotection
National Institutes of Health
K99 HL 122354 04/09/14-03/31/19 $121,750

BLAXALL B

Extracellular Matrix Remodeling and Fibrosis
National Institutes of Health(University of Rochester)
R01 GM 097347 08/15/12-11/30/15 $38,513

Small Molecule Targeting of MLK3 for Heart Failure
American Heart Association
13IRG14670079 01/01/13-12/31/14 $68,183

DAVIS, J

MBNL1's Function in Myofibroblast Transformation and Fibrosis
National Institutes of Health
K99 HL 119353 08/09/13-07/31/15 $121,375

FANG, M

Wnt/beta-catenin Signaling in Heart Valve Development
American Heart Association
13PRE16410009 07/01/13-06/30/15 $26,000

GOMEZ, M

BMP Signaling in the Progression of Calcific Aortic Valve Disease
American Heart Association
13PRE1623006 07/01/13-06/30/15 $26,000

KAMAL, F

Targeting Adrenal and Cardiac GPCR Signaling in Heart Failure: A Novel Therapeutic Strategy
American Heart Association
13POST16670003 07/01/13-06/30/15 $46,000

KHUCHUA, Z

A Mouse Model of Barth Syndrome, a Mitochondrial Cardiolipin Disorder
National Institutes of Health
R01 HL 108867 07/07/11-03/31/15 $245,000

Bezafibrates Pre-Trial on Mice
Barth Syndrome Foundation, Inc.
05/01/14-04/30/15 $38,500

KWONG, J

Defining the Role of SLC25a35 as a Regulator of the Mitochondrial Permeability Transition Pore and Cardiomyocyte Death
American Heart Association
12POSTDOC11950000 07/01/12-06/30/14 $47,999

MOLKENTIN, J / ROBBINS, J (MPI)

Thrombospondin 4 Regulates Adaptive ER Stress Response
National Institutes of Health
R01 HL 105924 01/01/11-12/31/14 $305,366

MOLKENTIN, J

CaMKII and InsP3-Mediated Signaling in Cardiac Myocytes
National Institutes of Health(The Regents of the University of California)
P01 HL 080101 08/01/11-05/31/16 $271,987

Improving Cardiac Function after Myocardial Infarction
National Institutes of Health(Temple University School of Medicine)
P01 HL 108806 05/07/12-03/31/17 $260,000

Regulating Fibrosis and Muscle Growth in the Muscular Dystrophies
National Institutes of Health(The University of Chicago)
P01 NS 072027 07/01/11-06/30/16 $215,000

Molecular Pathways Controlling Cardiac Gene Expression
National Institutes of Health
R37 HL 060562 07/01/13-06/30/18 $238,000

RAJAN, S

Translational and Post-Translational Regulation of Tropomyosin in Normal and Cardiomyopathic Hearts
American Heart Association

11SDG4980029	08/01/11-12/31/14	$70,000

ROBBINS , J

A TG Rabbit Model for the Functional Effects of FHC Mutations in B-Cardiac Myosin
National Institutes of Health(University of Vermont)

R21 HL 111847	07/15/12-06/30/14	$24,960

Cardiac Myosin Binding Protein-C: Structure, Function and Regulation
National Institutes of Health(University of Vermont)

P01 HL 059408	02/01/10-01/31/15	$304,920

Proteotoxicity: An Unappreciated Mechanism of Heart Disease
Fondation Leducq

	10/01/11-09/30/16	$247,636

Signaling Processes Underlying Cardiovascular Function
National Institutes of Health

P01 HL069779	06/06/02-05/31/18	$1,149,912

TARIQ, M

Identification of Novel Human X-Linked Heterotaxy Genes
American Heart Association

12POSTDOC10370002	07/01/12-02/28/14	$30,667

VAN BERLO, J

Functional Relevance and Extent of Endogenous Cardiac Regeneration by C-Kit Positive Stem Cells
National Institutes of Health

K99 HL 112852	06/04/12-07/31/13	$32,000

WARE, S

Genetic and Epigenetic Mechanisms in Cardiomyopathy
American Heart Association

13EIA13460001	01/01/13-12/31/14	$72,727

Genetic Registry for Pediatric Heart Disease: The CCVM Consortium
March of Dimes National

	06/01/13-05/31/16	$79,182

Genotype-Phenotype Association in Pediatric Cardiomyopathy
National Institutes of Health(University of Miami)

R01 HL 111459	04/01/12-03/31/14	$987,826

Uncovering Novel Genetic Causes and Risk in Congenital Heart Disease Patients
Burroughs Wellcome Foundation(University of Cincinnati)

	07/01/09-02/28/14	$150,000

WAXMAN, J

Coup-tf Dependent Mechanisms of Ventricular and Hemangioblast Specification
National Institutes of Health

R01 HL 112893	01/15/13-02/28/18	$220,500

Mechanisms of Retinoic Acid Signaling Induced Congenital Heart Defects
March of Dimes

6FY14389	06/01/14-05/31/17	$113,636

WIRRIG, E

The Role of COX2 in the Progression of Human Calcific Aortic Valve Disease
National Institutes of Health

F32 HL 110390	07/01/12-06/30/15	$53,942

YUTZEY, K

Cell Signaling Mechanisms of Calcific Aortic Valve Disease
National Institutes of Health

R01 HL 114682	08/23/12-06/30/16	$238,000

Cincinnati Children's SURF Program
American Heart Association

14UFEL19160000	02/01/14-01/31/16	$20,000

Twist 1 Regulation of Valve Progenitors
National Institutes of Health

R01 HL 082716	07/01/10-05/31/15	$242,550

Wnt Signaling in Heart Valve Development and Disease
National Institutes of Health

R01 HL 094319	04/15/12-02/28/16	$245,000

Current Year Direct **$6,474,506**

Industry Contracts

ROBBINS, J / MORALES, D

CorMatrix Cardiovascular, Inc	$75,024

Current Year Direct Receipts **$75,024**

Total **$6,549,530**