

Division Data Summary

Research and Training Details

Number of Faculty	47
Number of Research Fellows	25
Number of Research Students	14
Number of Support Personnel	138
Direct Annual Grant Support	\$5,786,182
Direct Annual Industry Support	\$47,585
Peer Reviewed Publications	155

Clinical Activities and Training

Number of Clinical Staff	4
Number of Clinical Fellows	12
Inpatient Encounters	1,648
Outpatient Encounters	14,231

Division Photo



Row 1: B Siegel, K Hor, C Chin, K Yutzey, J Towbin, J Robbins, H Heydarian
 Row 2: S Goldstein, J Cnota, M Maillet, C Liu, B Goldstein, J Sticka, A Phillips, R Hirsch, P Manning, C Krawczeski, E Urbina, L Cripe, J James, E Purevjav
 Row 3: J Waxman, M Taylor, J Jefferies, Z Khuchua, S Ware, J Molquentin, R Spicer, J Gulick, DW Benson, R Czosek, R Hinton, E Michelfelder, D Cooper, H Ippisch

Significant Publications

Significant Accomplishments

Advanced Heart Failure and Mechanical Circulatory Support Service

The Heart Institute launched the advanced heart failure and mechanical circulatory support service this year to extend the cutting-edge medical therapies we provide to patients of all ages.

A large component of any successful advanced heart failure program is providing mechanical circulatory support specifically in the form of ventricular assist devices (VADs.) Without such devices, many patients who develop end-stage disease would die. We implanted 10 Berlin EXCOR devices within the last year in patients from neonates to adults.

Creating an Environment that Matches Our Level of Care

An extensive renovation of our outpatient clinic area created a more welcoming environment for patients and families. We more than doubled the clinic area to more than 40,000 square feet, adding 16 patient care rooms, separate MRI and echo reading rooms, and separate spaces for pediatric and adult patients with congenital heart disease. New technology allows cardiologists to remotely supervise all echocardiograms.

The new interior features the heart-themed artwork of internationally-known artist Mackenzie Thorpe. Last fall, Thorpe conducted an art workshop with patients and they co-created the sculpture that resides in our pediatric waiting area.

Quality Improvement and Research

Our quality improvement and clinical excellence program implemented an “Executive Dashboard” of crucial measures including safety and quality, family satisfaction, clinical functions and resource use. This dashboard has allowed the Heart Institute to review its status quickly and efficiently with others and has become a model for other institutes and divisions within the hospital.

In cardiac research, faculty findings published October 2010 in the *Journal of the American College of Cardiology* showed that mutations in the nebulin protein isoform is associated with lethal cardiac structural abnormalities, including dilated cardiomyopathy and endocardial fibroelastosis. Study authors included Enkhsaikhan Purevjav, MD, PhD, Michael Taylor, MD, PhD, and Jeffrey Towbin, MD.

Our work in molecular cardiovascular biology also progressed significantly this year. Jeffrey Molkenin, PhD, studies intracellular signaling pathways and transcriptional regulatory circuits that control mammalian cell growth and differentiation. Jeffrey Robbins, PhD, established the means to direct the heart to synthesize normal and mutant proteins and to turn these on and off at will. This allows scientists to establish cause-and-effect relationships between mutant proteins and the development of cardiac disease. Stephanie Ware, MD, PhD, continued work with X-linked heterotaxy (HTX-1, MIM 306955), a rare developmental disorder characterized by disturbances in embryonic laterality and other midline developmental defects. Ware uses this genetic defect to study the underpinnings of early axis formation and its importance for cardiogenesis.

Division Publications

1. Acehan D, Vaz F, Houtkooper RH, James J, Moore V, Tokunaga C, Kulik W, Wansapura J, Toth MJ, Strauss A, Khuchua Z. **Cardiac and skeletal muscle defects in a mouse model of human Barth syndrome**. *The Journal of biological chemistry*. 2011; 286:899-908.
2. Anderson JB, Beekman RH, 3rd, Eghtesady P, Kalkwarf HJ, Uzark K, Kehl JE, Marino BS. **Predictors of poor weight gain in infants with a single ventricle**. *The Journal of pediatrics*. 2010; 157:407-13, 413 e1.
3. Anderson JB, Benson DW. **Genetics of sick sinus syndrome**. *Cardiac electrophysiology clinics*. 2010; 2:499-507.
4. Anderson JB, Kalkwarf HJ, Kehl JE, Eghtesady P, Marino BS. **Low weight-for-age z-score and infection risk after the Fontan procedure**. *The Annals of thoracic surgery*. 2011; 91:1460-6.
5. Atz AM, Trivison TG, Williams IA, Pearson GD, Laussen PC, Mahle WT, Cook AL, Kirsh JA, Sklansky M, Khaikin S, Goldberg C, Frommelt M, Krawczeski C, Puchalski MD, Jacobs JP, Baffa JM, Rychik J, Ohye RG. **Prenatal diagnosis and risk factors for preoperative death in neonates with single right ventricle and systemic outflow obstruction: screening data from the Pediatric Heart Network Single Ventricle Reconstruction Trial(*)**. *The Journal of thoracic and cardiovascular surgery*. 2010; 140:1245-50.
6. Baker-Smith CM, Neish SR, Klitzner TS, Beekman RH, 3rd, Kugler JD, Martin GR, Lannon C, Jenkins KJ, Rosenthal GL. **Variation in postoperative care following stage I palliation for single-ventricle patients: a report from the Joint Council on Congenital Heart Disease National Quality Improvement Collaborative**. *Congenital heart disease*. 2011; 6:116-27.
7. Banchs J, Jefferies JL, Plana JC, Hundley WG. **Imaging for cardiotoxicity in cancer patients**. *Texas Heart Institute journal / from the Texas Heart Institute of St. Luke's Episcopal Hospital, Texas Children's Hospital*. 2011; 38:268-9.
8. Baumgartner H, Bonhoeffer P, De Groot NM, de Haan F, Deanfield JE, Galie N, Gatzoulis MA, Gohlke-Baerwolf C, Kaemmerer H, Kilner P, Meijboom F, Mulder BJ, Oechslin E, Oliver JM, Serraf A, Szatmari A, Thaulow E, Vouhe PR, Walma E, Vahanian A, Auricchio A, Bax J, Ceconi C, Dean V, Filippatos G, Funck-Brentano C, Hobbs R, Kearney P, McDonagh T, Popescu BA, Reiner Z, Sechtem U, Sirnes PA, Tendera M,

- Vardas P, Widimsky P, Swan L, Andreotti F, Beghetti M, Borggrefe M, Bozio A, Brecker S, Budts W, Hess J, Hirsch R, Jondeau G, Kokkonen J, Kozelj M, Kucukoglu S, Laan M, Lionis C, Metreveli I, Moons P, Pieper PG, Pillosoff V, Popelova J, Price S, Roos-Hesselink J, Uva MS, Tornos P, Trindade PT, Ukkonen H, Walker H, Webb GD, Westby J. **ESC Guidelines for the management of grown-up congenital heart disease (new version 2010)**. *European heart journal*. 2010; 31:2915-57.
9. Beery TA, Smith CR, Kudel I, Knilans T. **Measuring sports participation decisional conflict in youth with cardiac pacemakers and/or ICDs**. *Journal of advanced nursing*. 2011; 67:821-8.
 10. Belmont JW, Craigen W, Martinez H, Jefferies JL. **Genetic disorders with both hearing loss and cardiovascular abnormalities**. *Advances in oto-rhino-laryngology*. 2011; 70:66-74.
 11. Bergersen L, Gauvreau K, Marshall A, Kreutzer J, Beekman R, Hirsch R, Foerster S, Balzer D, Vincent J, Hellenbrand W, Holzer R, Cheatham J, Moore J, Lock J, Jenkins K. **Procedure-type risk categories for pediatric and congenital cardiac catheterization**. *Circulation. Cardiovascular interventions*. 2011; 4:188-94.
 12. Boat A, Mahmoud M, Michelfelder EC, Lin E, Ngamprasertwong P, Schnell B, Kurth CD, Crombleholme TM, Sadhasivam S. **Supplementing desflurane with intravenous anesthesia reduces fetal cardiac dysfunction during open fetal surgery**. *Paediatric anaesthesia*. 2010; 20:748-56.
 13. Bolli R, Stein AB, Guo Y, Wang OL, Rokosh G, Dawn B, Molkentin JD, Sanganalmath SK, Zhu Y, Xuan YT. **A murine model of inducible, cardiac-specific deletion of STAT3: its use to determine the role of STAT3 in the upregulation of cardioprotective proteins by ischemic preconditioning**. *Journal of molecular and cellular cardiology*. 2011; 50:589-97.
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 15. Breitbart A, Auger-Messier M, Molkentin JD, Heineke J. **Myostatin from the heart: local and systemic actions in cardiac failure and muscle wasting**. *American journal of physiology. Heart and circulatory physiology*. 2011; 300:H1973-82.
 16. Broberg CS, Aboulhoshn J, Mongeon FP, Kay J, Valente AM, Khairy P, Earing MG, Opotowsky AR, Lui G, Gersony DR, Cook S, Ting JG, Webb G, Gurvitz MZ. **Prevalence of left ventricular systolic dysfunction in adults with repaired tetralogy of fallot**. *The American journal of cardiology*. 2011; 107:1215-20.
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 18. Brunetti-Pierri N, Pignatelli R, Fouladi N, Towbin JA, Belmont JW, Craigen WJ, Wong LJ, Jefferies JL, Scaglia F. **Dilation of the aortic root in mitochondrial disease patients**. *Molecular genetics and metabolism*. 2011; 103:167-70.
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 21. Carlo WF, Nelson DP. **Inhaled nitric oxide improves oxygen saturation in children with pulmonary**

- arteriovenous malformations after the Fontan procedure.** *Pediatric critical care medicine : a journal of the Society of Critical Care Medicine and the World Federation of Pediatric Intensive and Critical Care Societies.* 2011; 12:e152-4.
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38. Das A, Banerjee RK, Gottliebson WM. **Right ventricular inefficiency in repaired tetralogy of Fallot: proof of concept for energy calculations from cardiac MRI data.** *Annals of biomedical engineering.* 2010; 38:3674-87.
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Grants, Contracts, and Industry Agreements

Grant and Contract Awards

Annual Direct / Project Period Direct

ANDERSON, J

Nutritional Deficiencies in Infants with Single Ventricle

Thrasher Research Fund

10/01/10-09/30/12

\$12,500

BENSON , W

Musculoskeletal Ancillary Study

Marfan Foundation(The Johns Hopkins University)

07/01/10-06/30/11

\$3,750

Quality of Life Study-Pediatric Heart Network

National Institutes of Health(New England Research Institutes)

U01 HL 068270

07/01/10-06/30/11

\$500

Trial of Beta Blocker vs Angiotensin Receptor Blocker

National Institutes of Health(New England Research Institutes)

U01 HL 068270

07/01/10-06/30/11

\$72,010

BRAITSCH, C

Tbx18 Regulation of Epicardial-Derived Cell Proliferation, Migration and Differentiation in Cardiac Development

American Heart Association

07/01/09-06/30/11

\$23,000

COLE, C

Myocardial Protection during Fetal Bypass: Role of Calcium Cycling

National Institutes of Health F32 HL 103054	07/01/10-06/30/12	\$54,254
CORRELL, R		
Regulation of Cardiac Gene Expression National Institutes of Health F32 HL 097551	09/07/09-09/06/12	\$50,474
DAVIS, J		
The Non-Hypertrophic Role of Calcineurin in Regulating Cardiac Structure-Function National Institutes of Health F32 HL 095353	12/15/08-12/14/11	\$52,154
EGHTESADY, P		
Hypoplastic Left Heart Syndrome: Expression of RHD in the Fetus ? National Institutes of Health R01 HL 098634	01/15/10-12/31/14	\$269,164
IPPISCH, H		
Effects of Dietary Fat and Exercise Challenges in Obese Children National Institutes of Health K23 HL 091174	09/01/08-07/31/13	\$126,085
KARCH, J		
The Role of Bax and Bak in Necrotic Cell Death American Heart Association	07/01/10-06/30/12	\$23,000
KHUCHUA, Z		
The shRNA-Mediated Tafazzin Knockdown Mouse Model Barth Syndrome Foundation, Inc.	01/01/10-12/31/11	\$16,361
KIMBALL, T		
Chronic Kidney Disease in Children National Institutes of Health(Children's Mercy Hospital) U01 DK 066143	08/01/08-07/31/13	\$55,222
KRISHNAMURTHY, V		
Valve Tissue Mechanics and Cell Phenotype in a Mouse Model of Aortic Valve Disease American Heart Association	07/01/09-06/30/11	\$23,000
MAILLET, M		
Role of IP-3 Mediated Calcium Release in Cardiac Hypertrophic Cardiomyopathy National Institutes of Health R21 HL 097186	04/15/10-03/31/12	\$125,000
MARINO, B		
Understanding Mechanisms of Fontan Failure and Key Predictors of Patient Outcome National Institutes of Health(Georgia Tech Research Corp) R01 HL 098252	02/01/10-01/31/14	\$18,488
MOLKENTIN, J		
Calcium as a Molecular Signal in the Heart		

National Institutes of Health(Temple University School of Medicine)		
R01 HL 089312	08/15/07-06/30/12	\$239,303
Cardiac Hypertrophic Intracellular Signaling Pathways		
National Institutes of Health		
R01 HL 062927	02/01/09-12/31/13	\$250,000
Molecular Pathways Controlling Cardiac Gene Expression		
National Institutes of Health		
R37 HL 060562	07/01/08-06/30/13	\$250,000
Thrombospondin 4 Regulates Adaptive ER Stress Response		
National Institutes of Health		
R01 HL 105924	01/01/11-12/31/11	\$315,000
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RAZZAQUE, A		
Cardiomyopathic Mechanisms in Pediatric Congenital Disease		
American Heart Association		
	07/01/10-06/30/12	\$43,000
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ROBBINS, J		
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	\$356,105
Nikon A1 Confocal Microscope		
National Institutes of Health		
S10 RR 027014	07/01/10-06/30/11	\$388,205
Signaling Processes Underlying Cardiovascular Function		
National Institutes of Health		
P01 HL 069779	01/11/08-12/31/12	\$1,219,260
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SENGUPTA, A		
URC Postdoctoral Fellow Research Grant		
University of Cincinnati		
	01/01/11-12/31/11	\$5,000
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SIEGEL, R		
Collaborative Community Based Initiative to Improve the Health of Overweight and Obese Children		
WellPoint Foundation		
	01/01/11-12/31/11	\$76,223
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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	\$383,528
Modifying Dietary Behavior in Adolescents with Elevated Blood Pressure		
National Institutes of Health(University of Cincinnati)		
R01HL088567	02/01/11-01/31/12	\$37,818
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VAN BERLO, J		
GATA-6 Function is Crucial for Cardiac Hypertrophy to Prevent Heart Failure		
American Heart Association		
	07/01/10-06/30/12	\$43,000
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WARE, S		
Role of the Embryonic Node in Cardiac Development and Congenital Heart Disease		
National Institutes of Health		
R01 HL 088639	04/01/07-03/31/12	\$250,000
Uncovering Novel Genetic Causes and Risk in Congenital Heart Disease Patients		
Burroughs Wellcome Foundation(University of Cincinnati)		

1008496	07/01/09-06/30/14	\$75,000
Genetic Causes of Congenital Heart Defects		
March of Dimes		
1FY10401	06/01/10-05/31/13	\$93,866
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WAXMAN, J		
Elucidation of Molecular Networks Required to Limit Cardiac Cell Number		
National Institutes of Health		
R00 HL 091126	07/15/10-05/31/13	\$163,368
Illumination of Mechanisms Controlling Atrial Cell Formation		
March of Dimes National		
	02/01/11-01/31/13	\$69,327
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YUTZEY, K		
Notch Signaling in Heart Valve Development and Disease		
National Institutes of Health		
R01 HL 094319	07/01/09-06/30/11	\$308,217
The Function of Notch1 in Heart Valve Development		
American Heart Association		
	07/01/09-06/30/11	\$23,000
Twist 1 Regulation of Valve Progenitors		
National Institutes of Health		
R01 HL 082716	07/01/10-05/31/15	\$250,000
Weinstein Cardiovascular Development Conference 2011		
March of Dimes National		
	04/01/11-09/30/11	\$2,000
Student Undergraduate Research Fellowships		
American Heart Association		
10UFEL3580016	06/01/10-05/31/12	\$20,000
		Current Year Direct \$5,786,182
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Industry Contracts		
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BEEKMAN. R		
AGA Medical, LLC		\$4,177
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HIRSCH. R		
AGA Medical , LLC		\$12,199
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KRAWCZESKI. C		
Asklepion Pharmaceuticals, LLC		\$26,208
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YUTZEY, K		
Genentech, Inc		\$5,000
		Current Year Direct Receipts \$47,585
		Total \$5,833,767