Division Details

Division Data Summary

Research and Training Details

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Significant Accomplishments

Advanced Cardiomyopathy and Heart Failure Diagnosis and Treatments

The Heart Institute has become an international leader in heart muscle disease (cardiomyopathies) and its associated treatments. We have developed the largest Advanced Pediatric and Young Adult Cardiomyopathy and Heart Failure Program in the US, directed by John Lynn Jefferies, MD, MPH, and Jeffrey Towbin, MD. Our Heart Transplant Program, under the medical direction of Clifford Chin, MD, and surgical leadership of Alistair Phillips, MD, is one of the largest and most successful in the US. We performed 14 transplants in the past year and 10 the previous year, with 100 percent survival. More recently, the Heart Institute has become a leader in mechanical circulatory support with the addition of David Morales, MD, a world-renowned pediatric cardiothoracic surgeon and new chief of cardiothoracic surgery. Eight children with end-stage heart failure received mechanical circulatory support this year; six of whom were successfully transplanted and are doing well. Members of the Heart Institute also published more than 170 manuscripts this year.

Heart Institute Leads an International Research Consortium

Under the leadership of Jeffrey Robbins, PhD, the Heart Institute was awarded one of four grants given worldwide by the Fondation Leducq to study how proteotoxicity – cell malfunctions caused by misfolded proteins – can lead to heart disease and failure. The consortium for this study is led by Cincinnati Children’s and includes laboratories at the University of Texas-Southwest, University of North Carolina at Chapel Hill and premier academic medical centers in Germany, Great Britain and Italy. These proteotoxicity studies were recognized by the Leducq Foundation as having the potential to lead to novel treatments for a variety of heart
diseases, with new therapeutic targets being identified in our pediatric population.

**Pediatric Heart Network Consortium**

The Heart Institute successfully competed for a renewal grant to continue serving as one of nine “core” sites in the Pediatric Heart Network (PHN) through 2016. This national competition was open to academic centers across North America. The National Heart, Lung and Blood Institute created this collaborative network to conduct clinical studies in children with congenital or acquired heart disease. Since its inception, the PHN has completed more than 10 multi-center studies that have had significant impact on the field of pediatric cardiovascular disease. The most recent study, evaluating the use of therapies for individuals with Marfan syndrome, began in 2007 and will have a major impact on how we treat these patients. These complex, randomized trials are the only way of carrying out high quality clinical research in rare diseases. Cincinnati Children’s has been a core site in the PHN since 2006. James Cnota, MD and Catherine Krawczeski, MD, serve as principal investigators.

**Division Publications**


23. Burnside J, Gomez D, Preston TJ, Olshove VF, Jr., Phillips A. In-vitro quantification of gaseous


64. Hehir DA, Cooper DS, Walters EM, Ghanayem NS. Feeding, growth, nutrition, and optimal interstage surveillance for infants with hypoplastic left heart syndrome. Cardiol Young. 2011; 21 Suppl 2:59-64.


77. Jacobs JP, Jacobs ML, Austin EHI, Mavroudis C, Pasquali SK, Lacour-Gayet FG, Tchervenkov CI, Walters Hl,


93. Larsen RL, Canter CE, Naftel DC, Tressler M, Rosenthal DN, Blume ED, Mahle WT, Yung D, Morrow WR, Orav EJ, Wilkinson JD, Towbin JA, Lipshultz SE. The impact of heart failure severity at time of listing for


97. Lee MP, Yutzey KE. Twist1 directly regulates genes that promote cell proliferation and migration in developing heart valves. PloS one. 2011; 6:e29758.


110. Marino BS, Jefferies JL. Myocarditis. The 5 Minute Pediatric Consult. Philadelphia: Lippincott Williams &


156. Sengupta A, Chakraborty S, Paik J, Yutzey KE, Evans-Anderson HJ. *FoxO1 is required in endothelial but not myocardial cell lineages during cardiovascular development.* *Developmental dynamics : an official


166. Tariq M, Belmont JW, Lalani S, Smolarek T, Ware SM. SHROOM3 is a novel candidate for heterotaxy identified by whole exome sequencing. Genome Biol. 2011; 12:R91.


Grants, Contracts, and Industry Agreements
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<th>Name</th>
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<td>HORN, M</td>
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IPPISCH, H
Cardiovascular Effects of Dietary Fat and Exercise Challenges in Obese Children
National Institutes of Health
K23 HL 091174  09/01/08-07/31/13  $126,135

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
A Mouse Model of Barth Syndrome, a Mitochondrial Cardiolipin Disorder
National Institutes of Health
R01 HL 108867  07/07/11-03/31/15  $250,000

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
Aortic Root Structure-Function Relationships in a Mouse Model of Aortic Valve Disease and Aortopathy
American Heart Association
11PRE7210044  07/01/11-06/30/12  $23,000

LATTIN, B
Weight Management Program for Pre-school Children: A Feasibility Study for Group Treatment
Aramark(Academy of Nutrition and Dietetics)
06/01/12-05/31/13  $10,000

MARINO, B
Do Biochemical, Hematopoietic, and Stool Biomarkers Predict Low Cardiac Index in Patients with Fontan Physiology
Children's Heart Foundation
01/01/12-12/31/13  $100,000

Understanding Mechanisms of Fontan Failure and Key Predictors for Patient Outcome
National Institutes of Health(Georgia Tech Research Corp)
R01 HL 098252  02/01/10-01/31/14  $16,667

MCLENDON, P
The Role of Impaired Protein Degradation Pathways in CryABR120G-Mediated Desmin-R
National Institutes of Health
F32 HL 112558  01/11-12-01/10/14  $51,326

MICHELFELDER, E
Hypoplastic Left Heart Syndrome: Expression of RHD in the Fetus?
National Institutes of Health(Washington University)
05/15/11-12/31/14  $33,848

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
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<td>Proteotoxicity: An Unappreciated Mechanism of Heart Disease</td>
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<td>GATA-6 Function is Crucial for Cardiac Hypertrophy to Prevent Heart Failure</td>
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### Genetic causes of congenital heart defects

- **March of Dimes National**
  - **06/01/10-05/31/13**
  - **$79,590**

### 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)**
  - **BWF #1008496**
  - **07/01/09-06/30/15**
  - **$150,000**

### 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**
  - **$162,745**

- **Illumination of Mechanisms Controlling Atrial Cell Formation**
  - **March of Dimes National**
  - **02/01/11-01/31/13**
  - **$69,327**

### YUTZEY, K

- **Student Undergraduate Research Fellowship**
  - **American Heart Association**
  - **12UFEL9990000**
  - **02/01/12-01/31/14**
  - **$20,000**

- **Twist 1 Regulation of Valve Progenitors**
  - **National Institutes of Health**
  - **R01 HL 082716**
  - **07/01/10-05/31/15**
  - **$250,000**

- **Wnt Signaling in Heart Valve Development and Disease**
  - **National Institutes of Health**
  - **R01 HL 094319**
  - **04/15/12-02/28/16**
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### Cardiology

#### Grant and Contract Awards

- **ANDERSON, J**
  - **Nutritional Deficiencies in Infants with Single Ventricle**
  - **Thrasher Research Fund**
  - **10/01/10-09/30/12**
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<td>National Institutes of Health, Children's Mercy Hospital</td>
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**Current Year Direct** $1,184,765
Industry Contracts

BEEKMAN, R
JHU $2,229

CHIN, C
Hoffman-LaRoche, Inc $10,372

HIRSCH, R
AGA Medical, LLC $22,372

KRAWCZESKI, C
Asklepion Pharmaceuticals, LLC $24,378

Current Year Direct Receipts $59,351

Total $1,244,116

Cardiothoracic Surgery

Grant and Contract Awards

COLE, C
Myocardial Protection during Fetal Bypass: Role of Calcium Cycling
National Institutes of Health
F32 HL 103054 07/01/10-06/30/12 $53,042

Current Year Direct $53,042

Total $53,042

Molecular Cardiovascular Biology

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American Heart Association
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BHUIYAN, S
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