### Division Data Summary

#### Research and Training Details

<table>
<thead>
<tr>
<th>Details</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of Faculty</td>
<td>47</td>
</tr>
<tr>
<td>Number of Research Fellows</td>
<td>25</td>
</tr>
<tr>
<td>Number of Research Students</td>
<td>14</td>
</tr>
<tr>
<td>Number of Support Personnel</td>
<td>138</td>
</tr>
<tr>
<td>Direct Annual Grant Support</td>
<td>$5,786,182</td>
</tr>
<tr>
<td>Direct Annual Industry Support</td>
<td>$47,585</td>
</tr>
<tr>
<td>Peer Reviewed Publications</td>
<td>155</td>
</tr>
</tbody>
</table>

#### Clinical Activities and Training

<table>
<thead>
<tr>
<th>Details</th>
<th>Number</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of Clinical Staff</td>
<td>4</td>
</tr>
<tr>
<td>Number of Clinical Fellows</td>
<td>12</td>
</tr>
<tr>
<td>Inpatient Encounters</td>
<td>1,648</td>
</tr>
<tr>
<td>Outpatient Encounters</td>
<td>14,231</td>
</tr>
</tbody>
</table>

### 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 Molkentin, 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 nebulette 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 Molkentin, 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**


29. Crowley DI, Khoury PR, Urbina EM, Ippisch HM, Kimball TR. Cardiovascular impact of the pediatric obesity epidemic: higher left ventricular mass is related to higher body mass index. The Journal of pediatrics. 2011; 158:709-714 e1.


78. Kleinman ME, Chameides L, Schexnayder SM, Samson RA, Hazinski MF, Atkins DL, Berg MD, de Caen...


Genotype Influences Ventricular Remodeling in Infants With Single Ventricle.


31. Stone GW, Goldberg S, O'Shaughnessy C, Midei M, Siegel RM, Cristea E, Dangas G, Lansky AJ, Mehran R. 5-year follow-up of polytetrafluoroethylene-covered stents compared with bare-metal stents in aortocoronary saphenous vein grafts the randomized BARRICADE (barrier approach to restenosis:
restrict intima to curtail adverse events) trial. JACC. Cardiovascular interventions. 2011; 4:300-9.

132. Sugden PH, Markou T, Fuller SJ, Tham el L, Molkentin JD, Paterson HF, Clerk A. Monophosphothreonyl extracellular signal-regulated kinases 1 and 2 (ERK1/2) are formed endogenously in intact cardiac myocytes and are enzymically active. Cellular signalling. 2011; 23:468-77.


147. Ware SM. Genetic diagnosis in pediatric cardiomyopathy: clinical application and research


### Grants, Contracts, and Industry Agreements

<table>
<thead>
<tr>
<th>Grant and Contract Awards</th>
<th>Annual Direct / Project Period Direct</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>ANDERSON, J</strong></td>
<td></td>
</tr>
<tr>
<td>Nutritional Deficiencies in Infants with Single Ventricle</td>
<td></td>
</tr>
<tr>
<td>Thrasher Research Fund</td>
<td>10/01/10-09/30/12</td>
</tr>
<tr>
<td><strong>BENSON , W</strong></td>
<td></td>
</tr>
<tr>
<td>Musculoskeletal Ancillary Study</td>
<td>07/01/10-06/30/11</td>
</tr>
<tr>
<td>Marfan Foundation(The Johns Hopkins University)</td>
<td></td>
</tr>
<tr>
<td>Quality of Life Study-Pediatric Heart Network</td>
<td>07/01/10-06/30/11</td>
</tr>
<tr>
<td>National Institutes of Health(New England Research Institutes)</td>
<td></td>
</tr>
<tr>
<td>U01 HL 068270</td>
<td>07/01/10-06/30/11</td>
</tr>
<tr>
<td><strong>BRAITSCH, C</strong></td>
<td></td>
</tr>
<tr>
<td>Tbx18 Regulation of Epicardial-Derived Cell Proliferation, Migration and Differentiation in Cardiac Development</td>
<td>07/01/09-06/30/11</td>
</tr>
<tr>
<td>American Heart Association</td>
<td></td>
</tr>
<tr>
<td><strong>COLE, C</strong></td>
<td></td>
</tr>
<tr>
<td>Myocardial Protection during Fetal Bypass: Role of Calcium Cycling</td>
<td>07/01/09-06/30/11</td>
</tr>
<tr>
<td>Researcher</td>
<td>Project Title</td>
</tr>
<tr>
<td>------------------</td>
<td>-------------------------------------------------------------------------------</td>
</tr>
<tr>
<td>CORRELL, R</td>
<td>Regulation of Cardiac Gene Expression</td>
</tr>
<tr>
<td>DAVIS, J</td>
<td>The Non-Hypertrophic Role of Calcineurin in Regulating Cardiac Structure-Function</td>
</tr>
<tr>
<td>EGHTESADY, P</td>
<td>Hypoplastic Left Heart Syndrome: Expression of RHD in the Fetus?</td>
</tr>
<tr>
<td>IPPISCH, H</td>
<td>Effects of Dietary Fat and Exercise Challenges in Obese Children</td>
</tr>
<tr>
<td>KARCH, J</td>
<td>The Role of Bax and Bak in Necrotic Cell Death</td>
</tr>
<tr>
<td>KHUCHUA, Z</td>
<td>The shRNA-Mediated Tafazzin Knockdown Mouse Model</td>
</tr>
<tr>
<td>KIMBALL, T</td>
<td>Chronic Kidney Disease in Children</td>
</tr>
<tr>
<td>KRISHNAMURTHY, V</td>
<td>Valve Tissue Mechanics and Cell Phenotype in a Mouse Model of Aortic Valve Disease</td>
</tr>
<tr>
<td>MAILLET, M</td>
<td>Role of IP-3 Mediated Calcium Release in Cardiac Hypertrophic Cardiomyopathy</td>
</tr>
<tr>
<td>MARINO, B</td>
<td>Understanding Mechanisms of Fontan Failure and Key Predictors of Patient Outcome</td>
</tr>
<tr>
<td>MOLKENTIN, J</td>
<td>Calcium as a Molecular Signal in the Heart</td>
</tr>
<tr>
<td>Project Title</td>
<td>Responsible Institution</td>
</tr>
<tr>
<td>------------------------------------------------------------------------------</td>
<td>-------------------------------------------------------------</td>
</tr>
<tr>
<td>Cardiac Hypertrophic Intracellular Signaling Pathways</td>
<td>National Institutes of Health</td>
</tr>
<tr>
<td>Molecular Pathways Controlling Cardiac Gene Expression</td>
<td>National Institutes of Health</td>
</tr>
<tr>
<td>Thrombospondin 4 Regulates Adaptive ER Stress Response</td>
<td>National Institutes of Health</td>
</tr>
<tr>
<td>Thrombospondin 4 Regulates Adaptive ER Stress Response</td>
<td>National Institutes of Health</td>
</tr>
<tr>
<td>RAZZAQUE, A</td>
<td>Cardiomyopathic Mechanisms in Pediatric Congenital Disease</td>
</tr>
<tr>
<td>ROBBINS, J</td>
<td>Cardiac Myosin Binding Protein-C: Structure, Function and Regulation</td>
</tr>
<tr>
<td>ROBBINS, J</td>
<td>Cardiac Myosin Binding Protein-C: Structure, Function and Regulation</td>
</tr>
<tr>
<td>SENGUPTA, A</td>
<td>URC Postdoctoral Fellow Research Grant</td>
</tr>
<tr>
<td>SIEGEL, R</td>
<td>Collaborative Community Based Initiative to Improve the Health of Overweight and Obese Children</td>
</tr>
<tr>
<td>URBINA, E</td>
<td>Accelerated CV Aging in Youth Related to CV Risk Factor Clusters</td>
</tr>
<tr>
<td>URBINA, E</td>
<td>Modifying Dietary Behavior in Adolescents with Elevated Blood Pressure</td>
</tr>
<tr>
<td>VAN BERLO, J</td>
<td>GATA-6 Function is Crucial for Cardiac Hypertrophy to Prevent Heart Failure</td>
</tr>
<tr>
<td>WARE, S</td>
<td>Role of the Embryonic Node in Cardiac Development and Congenital Heart Disease</td>
</tr>
<tr>
<td>WARE, S</td>
<td>Uncovering Novel Genetic Causes and Risk in Congenital Heart Disease Patients</td>
</tr>
<tr>
<td>Research Project</td>
<td>PI Name</td>
</tr>
<tr>
<td>----------------------------------------------------------------------------------</td>
<td>--------------</td>
</tr>
<tr>
<td><strong>Genetic Causes of Congenital Heart Defects</strong></td>
<td>March of Dimes</td>
</tr>
<tr>
<td><strong>Elucidation of Molecular Networks Required to Limit Cardiac Cell Number</strong></td>
<td>WAXMAN, J</td>
</tr>
<tr>
<td><strong>Illumination of Mechanisms Controlling Atrial Cell Formation</strong></td>
<td>YUTZEY, K</td>
</tr>
<tr>
<td><strong>Notch Signaling in Heart Valve Development and Disease</strong></td>
<td>YUTZEY, K</td>
</tr>
<tr>
<td><strong>The Function of Notch1 in Heart Valve Development</strong></td>
<td>YUTZEY, K</td>
</tr>
<tr>
<td><strong>Twist 1 Regulation of Valve Progenitors</strong></td>
<td>YUTZEY, K</td>
</tr>
<tr>
<td><strong>Weinstein Cardiovascular Development Conference 2011</strong></td>
<td>YUTZEY, K</td>
</tr>
<tr>
<td><strong>Student Undergraduate Research Fellowships</strong></td>
<td>YUTZEY, K</td>
</tr>
</tbody>
</table>

**Current Year Direct** $5,786,182

**Industry Contracts**

<table>
<thead>
<tr>
<th>Industry Contract Details</th>
<th>Amount</th>
</tr>
</thead>
<tbody>
<tr>
<td>BEEKMAN, R AGA Medical, LLC</td>
<td>$4,177</td>
</tr>
<tr>
<td>HIRSCH, R AGA Medical, LLC</td>
<td>$12,199</td>
</tr>
<tr>
<td>KRAWCZESKI, C Asklepion Pharmaceuticals, LLC</td>
<td>$26,208</td>
</tr>
<tr>
<td>YUTZEY, K Genentech, Inc</td>
<td>$5,000</td>
</tr>
</tbody>
</table>

**Current Year Direct Receipts** $47,585

**Total** $5,833,767