Heart Institute

Cincinnati Children's

Division Details

Division Data Summary

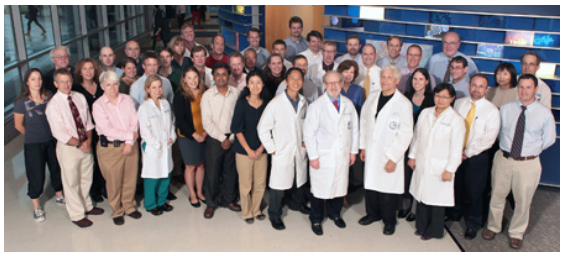
Research and Training Details

Number of Faculty	**50**
Number of Research Fellows	**31**
Number of Research Students	**12**
Number of Support Personnel	**232**
Direct Annual Grant Support	**$6,639,040**
Direct Annual Industry Support	**$59,351**
Peer Reviewed Publications	**189**

Clinical Activities and Training

Number of Clinical Staff	**5**
Number of Clinical Fellows	**13**
Inpatient Encounters	**1,652**
Outpatient Encounters	**16,074**

Division Photo



Row 1: R Siegel, P Goldenberg, A Lorts, C Castleberry, S Rajan, W Zheng, K Hor, J Towbin, J Robbins, E Purevjav
Row 2: M Maillet, S Kirk, K Yutzey, B Blaxall, J van Berlo, A Phillips, S Goldstein, H Ippisch, J Cnota, E Urbina, A Divanovic, J Anderson, D Spar
Row 3: J Gulick, J Waxman, Z Khuchua, S Ware, J Molkentin, C Chin, I Iliopoulos, D Cooper, B Marino, E Michelfelder, B Goldstein C Liu
Row 4: D Nelson, N Madsen, R Hinton, C Learn, R Czosek, T Knilans, G Webb, M Taylor

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

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2. Acharya A, Hans CP, Koenig SN, Nichols HA, Galindo CL, Garner HR, Merrill WH, Hinton RB, Garg V. **Inhibitory role of Notch1 in calcific aortic valve disease**. *PloS one*. 2011; 6:e27743.
3. Ackerman MJ, Priori SG, Willems S, Berul C, Brugada R, Calkins H, Camm AJ, Ellinor PT, Gollob M, Hamilton R, Hershberger RE, Judge DP, Le Marec H, McKenna WJ, Schulze-Bahr E, Semsarian C, Towbin JA, Watkins H, Wilde A, Wolpert C, Zipes DP. **HRS/EHRA expert consensus statement on the state of genetic testing for the channelopathies and cardiomyopathies this document was developed as a partnership between the Heart Rhythm Society (HRS) and the European Heart Rhythm Association (EHRA)**. *Heart rhythm : the official journal of the Heart Rhythm Society*. 2011; 8:1308-39.
4. Ackerman MJ, Priori SG, Willems S, Berul C, Brugada R, Calkins H, Camm AJ, Ellinor PT, Gollob M, Hamilton R, Hershberger RE, Judge DP, Le Marec H, McKenna WJ, Schulze-Bahr E, Semsarian C, Towbin JA, Watkins H, Wilde A, Wolpert C, Zipes DP. **HRS/EHRA expert consensus statement on the state of genetic testing for the channelopathies and cardiomyopathies: this document was developed as a partnership between the Heart Rhythm Society (HRS) and the European Heart Rhythm Association (EHRA)**. *Europace : European pacing, arrhythmias, and cardiac electrophysiology : journal of the working groups on cardiac pacing, arrhythmias, and cardiac cellular electrophysiology of the European Society of Cardiology*. 2011; 13:1077-109.
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Grants, Contracts, and Industry Agreements

Heart Institute

Grant and Contract Awards		Annual Direct
ACCORNERO, F		
PGF Role in Regulating Cardiac Remodeling		
American Heart Association		
11POST7530035	07/01/11-06/30/13	$43,000
ANDERSON, J		
Nutritional Deficiencies in Infants with Single Ventricle		
Thrasher Research Fund		
	10/01/10-09/30/12	$12,500
BHUIYAN, S		
Functional Significance and Regulation of cMyBP-C Binding to Actin		
American Heart Association		
11POST7590181	07/01/11-06/30/13	$43,000
COLE, C		
Myocardial Protection during Fetal Bypass: Role of Calcium Cycling		
National Institutes of Health		
F32 HL 103054	07/01/10-06/30/12	$53,042
COOPER, D		
Outcomes in Patients Requiring More Than a Single Course of ECMO Therapy		
Extracorporeal Life Support Organization		
	04/01/12-03/31/13	$5,000
CORRELL, R		
Regulation of Cardiac Gene Expression by the L-type Calcium Channel, CaV1.2		
National Institutes of Health		
F32 HL 097551	09/07/09-01/06/12	$18,989
CNOTA, J		
Pediatric Heart Network Prairieland Consortium		
National Institutes of Health		
U01 HL 109673	09/01/11-06/30/16	$367,500
GUPTA, M		
Role of Myosin Binding Protein-C Phosphorylation in Cardiac Function		
American Heart Association		
11POST7590026	07/01/11-06/30/13	$43,000
HOR, K		
Early Treatment with Aldosterone Antagonism Attenuates Cardiomyopathy in Duchenne Muscular Dystrophy		
Parent Project Muscular Dystrophy		
	06/01/12-05/31/14	$29,811
Early Treatment with Aldosterone Antagonism Attenuates Cardiomyopathy in Duchenne Muscular Dystrophy - Supplement Funds		
Ohio State University		
	06/01/12-05/31/14	$10,000
HORN, M		
Differential Twist1 Dimer Function and Gene Regulation iin Valve Progenitor Cells		
American Heart Association		

11PRE7240023 07/01/11-06/30/13 $23,000

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

R01 HL 062927 02/01/09-12/31/13 $222,750
Molecular Pathways Controling Cardiac Gene Expression
National Institutes of Health
R37 HL 060562 07/01/08-06/30/13 $250,000
Mechanisms of TGF Regulated Fibrosis in Muscular Dystrophy: Project 3
National Institutes of Health(The University of Chicago)
P01 NS 072027 07/01/11-06/30/16 $225,000
Thrombospondin 4 Regulates Adaptive ER Stress Response
National Institutes of Health
R01 HL 105924 01/01/11-12/31/11 $315,000
Ca Signaling Domains Programming Cardiac Hypertrophy
National Institutes of Health(University of California - Davis)
2P01 HL 080101 08/01/11-05/31/16 $274,517

RAJAN, S

Translational and Post-translational Regulation of Tropomyosin in Normal and Cardiomyopathic Hearts
American Heart Association
11SDG4980029 08/01/11-12/31/14 $102,083

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 $349,278
Proteotoxicity: An Unappreciated Mechanism of Heart Disease
Fondation Leducq
10/01/11-09/30/16 $168,182
Signaling Processes Underlying Cardiovascular Function
National Institutes of Health
P01 HL 069779 01/11/08-12/31/12 $1,219,260

SENGUPTA, A

FOXO Function in Cardiomyocytes During Development and Disease
American Heart Association
11POST7210026 07/01/11-06/30/13 $43,000

SIEGEL, R

Collaborative Community Based Initiative to Improve the Health of Overweight and Obese Children in Cincinnati
Anthem Blue Cross and Blue Shield Foundation, LLC(Wellpoint)
01/01/11-12/31/12 $76,223

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 $379,410
Modifying Dietary Behavior in Adolescents with Elevated Blood Pressure
National Institutes of Health(University of Cincinnati)
R01 HL 088567 02/01/08-01/31/13 $38,672

VAN BERLO, J

Functional Relevance and Extent of Endogenous Cardiac Regeneration by C-Kit Positive Stem Cells
National Institutes of Health
K99 HL 112852 06/04/12-06/03/17 $121,660
GATA-6 Function is Crucial for Cardiac Hypertrophy to Prevent Heart Failure
American Heart Association
07/01/10-06/30/12 $45,000

WARE, S

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 $250,000

Current Year Direct $6,639,040

Industry Contracts

BEEKMAN
JHU $2,229

CHIN
Hoffman-LaRoche Inc $10,372

HIRSCH
AGA Medical , LLC $22,372

KRAWCZESKI
Asklepion Pharmaceuticals, LLC $24,378

Current Year Direct Receipts $59,351

Total $6,698,391

Cardiology

Grant and Contract Awards — Annual Direct

ANDERSON, J

Nutritional Deficiencies in Infants with Single Ventricle
Thrasher Research Fund
10/01/10-09/30/12 $12,500

CNOTA, J / KRAWCZESKI, C

Pediatric Heart Network Prairieland Consortium
National Institutes of Health
U01 HL 109673 | 09/01/11-06/30/16 | $367,500

COOPER, D

Outcomes in Patients Requiring More Than a Single Course of ECMO Therapy
Extracorporeal Life Support Organization
04/01/12-03/31/13 | $5,000

HOR, K

Early Treatment with Aldosterone Antagonism Attenuates Cardiomyopathy in Duchenne Muscular Dystrophy
Parent Project Muscular Dystrophy
06/01/12-05/31/14 | $29,811

Early Treatment with Aldosterone Antagonism Attenuates Cardiomyopathy in Duchenne Muscular Dystrophy - Supplement Funds
Ohio State University
06/01/12-05/31/14 | $10,000

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

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

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

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

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 | $379,410

Modifying Dietary Behavior in Adolescents with Elevated Blood Pressure
National Institutes of Health(University of Cincinnati)
R01 HL 088567 | 02/01/08-01/31/13 | $38,672

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		Annual Direct
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

Grant and Contract Awards		Annual Direct
ACCORNERO, F		
PGF Role in Regulating Cardiac Remodeling		
American Heart Association		
11POST7530035	07/01/11-06/30/13	$43,000
BHUIYAN, S		
Functional Significance and Regulation of cMyBP-C Binding to Actin		
American Heart Association		
11POST7590181	07/01/11-06/30/13	$43,000
CORRELL, R		
Regulation of Cardiac Gene Expression by the L-type Calcium Channel, CaV1.2		
National Institutes of Health		
F32 HL 097551	09/07/09-01/06/12	$18,989
GUPTA, M		
Role of Myosin Binding Protein-C Phosphorylation in Cardiac Function		
American Heart Association		
11POST7590026	07/01/11-06/30/13	$43,000
HORN, M		
Differential Twist1 Dimer Function and Gene Regulation in Valve Progenitor Cells		

American Heart Association
11PRE7240023 07/01/11-06/30/13 $23,000

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

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

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

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 $222,750
Molecular Pathways Controling Cardiac Gene Expression
National Institutes of Health
R37 HL 060562 07/01/08-06/30/13 $250,000
Mechanisms of TGF Regulated Fibrosis in Muscular Dystrophy
National Institutes of Health(The University of Chicago)
P01 NS 072027 07/01/11-06/30/16 $225,000
Thrombospondin 4 Regulates Adaptive ER Stress Response
National Institutes of Health
R01 HL 105924 01/01/11-12/31/11 $315,000
Ca Signaling Domains Programming Cardiac Hypertrophy
National Institutes of Health(The Regents of the University of California)
PO1 HL 080101 08/01/11-05/31/16 $271,987

RAJAN, S

Translational and Post-Translational Regulation of Tropomyosin in Normal and Cardiomyopathic Hearts
American Heart Association
11SDG4980029 08/01/11-12/31/14 $76,370

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 $349,278
Proteotoxicity: An Unappreciated Mechanism of Heart Disease
Fondation Leducq
10/01/11-09/30/16 $168,182
Signaling Processes Underlying Cardiovascular Function
National Institutes of Health

P01 HL 069779	01/11/08-12/31/12	$1,219,260

SENGUPTA, A

FOXO Function in Cardiomyocytes During Development and Disease
American Heart Association

11POST7210026	07/01/11-06/30/13	$43,000

VAN BERLO, J

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

K99 HL 112852	06/04/12-06/03/17	$121,660

GATA-6 Function is Crucial for Cardiac Hypertrophy to Prevent Heart Failure
American Heart Association

	07/01/10-06/30/12	$45,000

WARE, S

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	$250,000

Current Year Direct	**$5,296,767**
Total	**$5,296,767**