



## **2018 Annual Report**

### ***Division of Cardiology***

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#### **DIVISION DESCRIPTION**

The Division of Pediatric Cardiology and the Congenital Heart Program of the American Family Children's Hospital (AFCH) provide outstanding comprehensive care to children and adults with congenital and acquired heart disease throughout Wisconsin and the region.

Clinical specialties include diagnosis and management of pediatric and adult congenital heart disease, including care of fetuses and newborns with complex structural heart problems; noninvasive cardiac imaging; interventional catheterization; inherited arrhythmias and interventional electrophysiology; exercise physiology; and preventive cardiology. The division partners with pediatric cardiothoracic surgery and critical care services to provide complete interventional and surgical care for complex congenital heart disease.

Current research investigates the biology of inherited cardiomyopathies, diagnosis and management of complex arrhythmias, pulmonary vascular abnormalities associated with congenital heart disease, and effective strategies to diagnose and treat familial hypercholesterolemia in children.

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#### **2018 HIGHLIGHTS**

- Medical student Carolina Larrain (mentor: **Luke Lamers, MD**) received a Shapiro Summer Research Award for the project, "Assessment of Pulmonary Vascular Function Following Catheter Interventions for Branch Pulmonary Artery Stenosis in a Swine Model of Congenital Heart Disease."
- Medical student Connor Enright (mentor: **Amy Peterson, MD**) received a Shapiro Summer Research Award for the project, "Safety and Efficacy of Statin Medications for Cardiovascular Risk Reduction in Pediatric Populations."
- **J. Carter Ralphe, MD**, received a one-year, \$73,460 Fall Competition grant from the UW-Madison Office of the Vice Chancellor for Research and Graduate Education for the project, "Human Engineered Cardiac Tissue Modeling of Familial Hypertrophic Cardiomyopathy."
- **Nicholas Von Bergen, MD**, launched Atrility, LLC, in partnership with a several UW-Madison bioengineering students to develop cost-effective novel tools to identify and treat arrhythmias.
- In 2018, *U.S. News and World Report* ranked UW Health's American Family Children's Hospital in the [top 50 children's hospitals for cardiology and heart surgery](#).

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## RECENT PUBLICATIONS

Alshwabkeh L, Opotowsky AR, Carter KD, Givertz MM, Landzberg MJ, Urey MA, **Bartlett H**. disparities in wait-list outcomes for adults with congenital heart disease listed for heart transplantation before and since revision of status I listing. *Am J Cardiol*. 2018 Nov 15;122(10):1761-1764. doi: 10.1016/j.amjcard.2018.08.013. Epub 2018 Aug 21. PubMed PMID: 30236623.

Bates ML, Anagnostopoulos PV, Nygard C, Torgeson J, Reichert J, Galambos C, Eldridge MW, **Lamers LJ**. Consequences of an early catheter-based intervention on pulmonary artery growth and right ventricular myocardial function in a pig model of pulmonary artery stenosis. *Catheter Cardiovasc Interv*. 2018 Mar 30. doi: 10.1002/ccd.27593. [Epub ahead of print] PubMed PMID: 29602248. \*

Chen X, Weber C, **Farrell ET**, Alvarado FJ, Zhao YT, Gómez AM, Valdivia HH. Sorcin ablation plus  $\beta$ -adrenergic stimulation generate an arrhythmogenic substrate in mouse ventricular myocytes. *J Mol Cell Cardiol*. 2018 Jan;114:199-210. doi: 10.1016/j.yjmcc.2017.11.017. Epub 2017 Nov 22. PubMed PMID:29174767; PubMed Central PMCID: PMC5800963. \*\*

**Farrell E**, Armstrong AE, Grimes AC, Naya FJ, **de Lange WJ**, **Ralphe JC**. Transcriptome analysis of cardiac hypertrophic growth in MYBPC3-null mice suggests early responders in hypertrophic remodeling. *Front Physiol*. 2018 Oct 25;9:1442. doi: 10.3389/fphys.2018.01442. eCollection 2018. PubMed PMID: 30410445; PubMed Central PMCID: PMC6210548.

Fenster ME, **Hokanson JS**. Heart murmurs and echocardiography findings in the normal newborn nursery. *Congenit Heart Dis*. 2018 Sep;13(5):771-775. doi: 10.1111/chd.12651. Epub 2018 Jul 24. PubMed PMID: 30039518.

Field ME, Laffin JJ, Langberg JJ, **Von Bergen NH**. Isolated Wolff-Parkinson-White syndrome in identical twins. *HeartRhythm Case Rep*. 2018 Feb 9;4(4):138-140. doi: 10.1016/j.hrcr.2018.01.013. eCollection 2018 Apr. PubMed PMID: 29755940; PubMed Central PMCID: PMC5944030.

Goss KN, Beshish AG, Barton GP, Haraldsdottir K, Levin TS, Tetri LH, Battiola TJ, Mulchrone AM, Pegelow DF, Palta M, **Lamers LJ**, Watson AM, Chesler NC, Eldridge MW. Early pulmonary vascular disease in young adults born preterm. *Am J Respir Crit Care Med*. 2018 Jun 26. doi: 10.1164/rccm.201710-2016OC. [Epub ahead of print] PubMed PMID: 29944842. \*

Kipp RT, Boynton JR, Field ME, Wang JF, Bares A, Leal MA, **Von Bergen NH**, Eckhardt LL. Outcomes during intended fluoroscopy-free ablation in adults and children. *J Innov Card Rhythm Manag*. 2018 Sep;9:3305-3311. PubMed PMID: 30568847; PubMed Central PMCID: PMC6296490.

**Lamers L**, Jimenez EE, **Allen C**, **Hoyme D**, Lushaj EB, Anagnostopoulos PV. Intraoperative completion angiogram may be superior to transesophageal echocardiogram for detection of pulmonary artery residual lesions in congenital heart surgery. *Pediatr Cardiol*. 2018 Jun;39(5):884-891. doi: 10.1007/s00246-018-1837-4. Epub 2018 Mar 10. PubMed PMID: 29525903.

Legare JM, Modaff P, Strom SP, Pauli RM, **Bartlett HL**. Geleophysic dysplasia: 48 year clinical update with emphasis on cardiac care. *Am J Med Genet A*. 2018 Nov;176(11):2237-2242. doi: 10.1002/ajmg.a.40377. Epub 2018 Sep 8. PubMed PMID: 30195254.

Moses KL, Seymour M, Beshish A, Baker KR, Pegelow DF, **Lamers LJ**, Eldridge MW, Bates ML. Inspiratory and expiratory resistance cause right-to-left bubble passage through the foramen ovale. *Physiol Rep*. 2018 Jun;6(12):e13719. doi: 10.14814/phy2.13719. PubMed PMID: 29952137; PubMed Central PMCID: PMC6021277.

Nellis JR, Fitch ZW, Choi AY, Meza JM, Spector ZZ, **Von Bergen NH**, Torres JE, Klapper JA, Sauer JS, Turek JW. A minimally invasive approach for placing sew-on epicardial leads in the child. *Innovations (Phila)*. 2018 Nov/Dec;13(6):455-457. doi: 10.1097/IMI.0000000000000568. PubMed PMID: 30540590.

Qureshi AM, Bansal N, McElhinney DB, Boudjemline Y, Forbes TJ, Maschietto N, Shahnavaz S, Cheatham JP, Krasuski R, **Lamers L**, Chessa M, Morray BH, Goldstein BH, Noel CV, Wang Y, Gillespie MJ. Branch pulmonary artery valve implantation reduces pulmonary regurgitation and improves right ventricular size/function in patients with large right ventricular outflow tracts. *JACC Cardiovasc Interv*. 2018 Mar 26;11(6):541-550. doi: 10.1016/j.jcin.2018.01.278. PubMed PMID:29566799.

Roston TM, Haji-Ghassemi O, LaPage MJ, Batra AS, Bar-Cohen Y, Anderson C, Lau YR, **Maginot K**, Gebauer RA, Etheridge SP, Potts JE, Van Petegem F, Sanatani S. Catecholaminergic polymorphic ventricular tachycardia patients with multiple genetic variants in the PACES CPVT Registry. *PLoS One*. 2018 Nov 7;13(11):e0205925. doi: 10.1371/journal.pone.0205925. eCollection 2018. PubMed PMID: 30403697; PubMed Central PMCID: PMC6221297.

Roston TM, Yuchi Z, Kannankeril PJ, Hathaway J, Vinocur JM, Etheridge SP, Potts JE, **Maginot KR**, Salerno JC, Cohen MI, Hamilton RM, Pflaumer A, Mohammed S, Kimlicka L, Kanter RJ, LaPage MJ, Collins KK, Gebauer RA, Temple JD, Batra AS, Erickson C, Mischak-Knecht M, Kubuš P, Bar-Cohen Y, Kantoch M, Thomas VC, Hessling G, Anderson C, Young ML, Choi SHJ, Cabrera Ortega M, Lau YR, Johnsrude CL, Fournier A, Van Petegem F, Sanatani S. The clinical and genetic spectrum of catecholaminergic polymorphic ventricular tachycardia: findings from an international multicentre registry. *Europace*. 2018 Mar 1;20(3):541-547. doi: 10.1093/europace/euw389. PubMed PMID: 28158428; PubMed Central PMCID: PMC6059141. \*\*

Ruedinger KL, Zhou H, Trampe B, Heiser T, **Srinivasan S**, Iruretagoyena JI, Roldán-Alzate A. Modeling fetal cardiac anomalies from prenatal echocardiography with 3-dimensional printing and 4-dimensional flow magnetic resonance imaging. *Circ Cardiovasc Imaging*. 2018 Sep;11(9):e007705. doi: 10.1161/CIRCIMAGING.118.007705. PubMed PMID: 30354680.

Smelter DF, **de Lange WJ**, Cai W, Ge Y, **Ralphe JC**. The HCM-linked W792R mutation in cardiac myosin-binding protein C reduces C6 FnIII domain stability. *Am J Physiol Heart Circ Physiol*. 2018 Jun 1;314(6):H1179-H1191. doi: 10.1152/ajpheart.00686.2017. Epub 2018 Feb 16. PubMed PMID: 29451820; PubMed Central PMCID: PMC6032085.

**Srinivasan S**, Howley LW, Cuneo BF, Chatfield KC. In-utero idiopathic ductal constriction: a prenatal manifestation of Alagille and Williams syndrome arteriopathy. *J Perinatol*. 2018 Nov;38(11):1453-1456. doi: 10.1038/s41372-018-0221-9. Epub 2018 Sep 10. PubMed PMID: 30202046.

Yang KC, Breitbart A, **De Lange WJ**, Hofsteen P, Futakuchi-Tsuchida A, Xu J, Schopf C, Razumova MV, Jiao A, Boucek R, Pabon L, Reinecke H, Kim DH, **Ralphe JC**, Regnier M, Murry CE. Novel adult-onset systolic cardiomyopathy due to MYH7 E848G mutation in patient-derived induced pluripotent stem cells. *JACC Basic Transl Sci*. 2018 Dec 31;3(6):728-740. doi: 10.1016/j.jacbs.2018.08.008. eCollection 2018 Dec. PubMed PMID: 30623132; PubMed Central PMCID: PMC6314962.

Zawacki A, Dodge A, Eickhoff J, Sun W, Marriott E, **Ralphe JC**, **Peterson AL**. novel lipid thresholds for screening predict the need for pharmacotherapy. *J Pediatr*. 2018 Nov;202:220-225.e2. doi: 10.1016/j.jpeds.2018.07.008. Epub 2018 Aug 29. PubMed PMID: 30172432.

Zawacki AW, Dodge A, Woo KM, **Ralphe JC, Peterson AL**. In pediatric familial hypercholesterolemia, lipoprotein(a) is more predictive than LDL-C for early onset of cardiovascular disease in family members. *J Clin Lipidol*. 2018 Nov - Dec;12(6):1445-1451. doi: 10.1016/j.jacl.2018.07.014. Epub 2018 Jul 31. PubMed PMID: 30150142.

*\*ePub only; no print citation available when report was compiled*

*\*\* Publication had previously appeared in 2017 report as an ePub*

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## GRANT SUPPORT

Principal Investigator	Sponsor	Title	Co-Investigators
Hokanson, John S	Wisconsin Department of Health Services	SHINE	
Lamers, Luke J	UW - Institute for Clinical & Translational Research	Assessment of early vs. delayed catheter intervention for the management of pulmonary artery stenosis	Cardiology: J. Carter Ralphe (Mentor)
Speidel, Michael (UW Department of Medical Physics)	National Institutes of Health (NIH)	Dual-energy subtraction angiography for transcatheter interventions	Cardiology: Luke Lamers
Peterson, Amy	Familial Hypercholesterolemia Foundation	CASCADE-FH registry	
Peterson, Amy	Wisconsin Partnership Program	Improved diagnosis of pediatric familial hypercholesterolemia through the Wisconsin pediatric lipid consortium	
Ralphe, John Carter	National Institutes of Health (NIH)	The role of the C3 domain of myosin binding protein-C in the pathogenesis of hypertrophic cardiomyopathy	
Ralphe, John Carter	Wisconsin Alumni Research Foundation	Human engineered cardiac tissue modeling of familial hypertrophic cardiomyopathy	
Ge, Ying (UW Department of Cell and Regenerative Biology)	National Institutes of Health (NIH)	Top-down proteomics of myofilaments in heart failure	Cardiology: J. Carter Ralphe
Wakai, Ronald (UW Department of Medical Physics)	National Institutes of Health (NIH)	Optimized measurement and signal processing of fetal MCG	Cardiology: Shardha Srinivasan

Von Bergen, Nicholas H	Medtronic, Inc. & Washington University	Recovery time and patient satisfaction in pediatric patients following implantable loop recorder implant: LINQ vs. REVEAL	
Von Bergen, Nicholas H	Hospital for Sick Children	Defibrillation testing at the time of cardioverter defibrillation implantation in pediatric and congenital heart disease patients	
Von Bergen, Nicholas H	Altathera Pharmaceuticals	Intravenous sotalol in pediatric and congenital patients: A multi-center registry study	