

Stanford



James Priest

Adjunct Clinical Assistant Professor, Pediatrics - Cardiology

CLINICAL OFFICES

- **LPCH Heart Center**

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Bio

CLINICAL FOCUS

- Pediatrics

ACADEMIC APPOINTMENTS

- Member, Cardiovascular Institute
- Member, Maternal & Child Health Research Institute (MCHRI)
- Member, Wu Tsai Neurosciences Institute

PROFESSIONAL EDUCATION

- Board Certification: Pediatric Cardiology, American Board of Pediatrics (2022)
- Medical Education: Stanford University School of Medicine (2008) CA
- Fellowship: Stanford University Pediatric Cardiology Fellowship (2016) CA
- Residency: University of Washington Pediatric Residency (2011) WA
- Board Certification: Pediatrics, American Board of Pediatrics (2011)
- MA, University of California Berkeley, Molecular Biology (2004)

LINKS

- Priest Lab Website: <https://priestlab.stanford.edu/>
- Priest Lab github repo: <https://github.com/priestlab/>

Research & Scholarship

CURRENT RESEARCH AND SCHOLARLY INTERESTS

Over the last thirty years, our fundamental understanding of the genetics and pathogenesis of congenital heart disease has lagged the tremendous advances in the surgical and clinical care of infants with this group of disorders. With my combined research training in genomics and clinical training in pediatric cardiology I

endeavor to close this gap with investigation into the genetic basis of congenital heart malformations and developing new models of disease. My goal is translate an improved molecular genetic and developmental understanding of congenital heart disease from the laboratory into clinically actionable models, diagnostics, and ultimately therapeutic interventions.

Teaching

GRADUATE AND FELLOWSHIP PROGRAM AFFILIATIONS

- Cardiovascular Medicine (Fellowship Program)
- Pediatric Cardiology (Fellowship Program)

Publications

PUBLICATIONS

- **Adults With Mild-to-Moderate Congenital Heart Disease Demonstrate Measurable Neurocognitive Deficits.** *Journal of the American Heart Association*
Perrotta, M. L., Saha, P. n., Zawadzki, R. n., Beidelman, M. n., Ingelsson, E. n., Lui, G. K., Priest, J. R.
2020: e015379
- **Association of congenital cardiovascular malformation and neuropsychiatric phenotypes with 15q11.2 (BP1-BP2) deletion in the UK Biobank.** *European journal of human genetics : EJHG*
Williams, S. G., Nakev, A. n., Guo, H. n., Frain, S. n., Tenin, G. n., Liakhovitskaia, A. n., Saha, P. n., Priest, J. R., Hentges, K. E., Keavney, B. D.
2020
- **Clonally expanding smooth muscle cells promote atherosclerosis by escaping efferocytosis and activating the complement cascade.** *Proceedings of the National Academy of Sciences of the United States of America*
Wang, Y. n., Nanda, V. n., Direnzo, D. n., Ye, J. n., Xiao, S. n., Kojima, Y. n., Howe, K. L., Jarr, K. U., Flores, A. M., Tsantilas, P. n., Tsao, N. n., Rao, A. n., Newman, et al
2020
- **Maternal Obesity and Diabetes Mellitus as Risk Factors for Congenital Heart Disease in the Offspring.** *Journal of the American Heart Association*
Helle, E. n., Priest, J. R.
2020: e011541
- **Exome-Based Case-Control Analysis Highlights the Pathogenic Role of Ciliary Genes in Transposition of the Great Arteries.** *Circulation research*
Liu, X. n., Chen, W. n., Li, W. n., Priest, J. R., Fu, Y. n., Pang, K. J., Ma, B. n., Han, B. n., Liu, X. n., Hu, S. n., Zhou, Z. n.
2020
- **Ivy: Instrumental Variable Synthesis for Causal Inference**
Kuangy, Z., Sala, F., Sohoni, N., Wu, S., Cordova-Palomera, A., Dunnmon, J., Priest, J., Re, C., Chiappa, S., Calandra, R.
ADDISON-WESLEY PUBL CO.2020: 398–409
- **Association between the 4p16 genomic locus and different types of congenital heart disease: results from adult survivors in the UK Biobank.** *Scientific reports*
Cordova-Palomera, A., Priest, J. R.
2019; 9 (1): 16515
- **Phenome-wide Burden of Copy-Number Variation in the UK Biobank.** *American journal of human genetics*
Aguirre, M., Rivas, M. A., Priest, J.
2019
- **Risk factors associated with the development of double-inlet ventricle congenital heart disease** *BIRTH DEFECTS RESEARCH*
Paige, S. L., Yang, W., Priest, J. R., Botto, L. D., Shaw, G. M., Collins, R., Natl Birth Defects Prevention
2019; 111 (11): 640–48
- **Substantial Cardiovascular Morbidity in Adults With Lower-Complexity Congenital Heart Disease** *CIRCULATION*
Saha, P., Potiny, P., Rigdon, J., Morello, M., Tcheandjieu, C., Romfh, A., Fernandes, S. M., McElhinney, D. B., Bernstein, D., Lui, G. K., Shaw, G. M., Ingelsson, E., Priest, et al
2019; 139 (16): 1889–99

- **Risk factors associated with the development of double-inlet ventricle congenital heart disease.** *Birth defects research*
Paige, S. L., Yang, W., Priest, J. R., Botto, L. D., Shaw, G. M., Collins, R. T., National Birth Defects Prevention Study
2019
- **NEUROCOGNITIVE DEFICITS IN ADULT CONGENITAL HEART DISEASE: DOES CORONARY ARTERY DISEASE ADD INSULT TO INJURY?**
Morello, M. L., Beidelman, M., Saha, P., Ingelsson, E., Shaw, G., Lui, G., Priest, J.
ELSEVIER SCIENCE INC.2019: 566
- **Loss of function, missense, and intronic variants in NOTCH1 confer different risks for left ventricular outflow tract obstructive heart defects in two European cohorts** *GENETIC EPIDEMIOLOGY*
Helle, E., Cordova-Palomera, A., Ojala, T., Saha, P., Potiny, P., Gustafsson, S., Ingelsson, E., Bamshad, M., Nickerson, D., Chong, J. X., Ashley, E., Priest, J. R., Univ Washington Ctr Mendelia
2019; 43 (2): 215–26
- **Substantial Cardiovascular Morbidity in Adults with Lower-Complexity Congenital Heart Disease.** *Circulation*
Saha, P., Potiny, P., Rigdon, J., Morello, M., Tcheandjieu, C., Romfh, A., Fernandes, S. M., McElhinney, D. B., Bernstein, D., Lui, G. K., Shaw, G. M., Ingelsson, E., Priest, et al
2019
- **Expansion of the Human Phenotype Ontology (HPO) knowledge base and resources** *NUCLEIC ACIDS RESEARCH*
Koehler, S., Carmody, L., Vasilevsky, N., Jacobsen, J. B., Danis, D., Gouridine, J., Gargano, M., Harris, N. L., Matentzoglou, N., McMurry, J. A., Osumi-Sutherland, D., Cipriani, V., Balhoff, et al
2019; 47 (D1): D1018–D1027
- **Single-Cell RNA-Seq of the Developing Cardiac Outflow Tract Reveals Convergent Development of the Vascular Smooth Muscle Cells.** *Cell reports*
Liu, X. n., Chen, W. n., Li, W. n., Li, Y. n., Priest, J. R., Zhou, B. n., Wang, J. n., Zhou, Z. n.
2019; 28 (5): 1346–61.e4
- **IMPACT OF CARDIAC ALGORITHM ON CYTOGENETIC TESTING**
Floyd, B. J., Hintz, S. R., Suarez, C. J., Cherry, A., Yu, L., Benitz, W., Priest, J. R., Wright, G. E., Bhombal, S., Davis, A., Chock, V. Y., Weigel, N., Kobayashi, et al
BMJ PUBLISHING GROUP.2019: 207
- **Weakly supervised classification of rare aortic valve malformations using unlabeled cardiac MRI sequences** *Nature Communications*
Fries, J. A., Varma, P., Chen, V. S., Xiao, K., Tejada, H., Saha, P., Dunmon, J., Chubb, H., Maskatia, S., Fiterau, M., Delp, S., Ashley, E., Ré, et al
2019; 10
- **Loss of function, missense, and intronic variants in NOTCH1 confer different risks for left ventricular outflow tract obstructive heart defects in two European cohorts.** *Genetic epidemiology*
Helle, E., Cordova-Palomera, A., Ojala, T., Saha, P., Potiny, P., Gustafsson, S., Ingelsson, E., Bamshad, M., Nickerson, D., Chong, J. X., University of Washington Center for Mendelian Genomics, Ashley, E., Priest, J. R.
2018
- **Expansion of the Human Phenotype Ontology (HPO) knowledge base and resources.** *Nucleic acids research*
Kohler, S., Carmody, L., Vasilevsky, N., Jacobsen, J. O., Danis, D., Gouridine, J., Gargano, M., Harris, N. L., Matentzoglou, N., McMurry, J. A., Osumi-Sutherland, D., Cipriani, V., Balhoff, et al
2018
- **CONGENITAL HEART DISEASE CONFERS SUBSTANTIAL RISK OF ACQUIRED CARDIOVASCULAR DISEASE AMONGST BRITISH ADULTS**
Saha, P., Potiny, P., Tcheandjieu, C., Fernandes, S. M., Romfh, A., Bernstein, D., Lui, G. K., Ingelsson, E., Priest, J.
ELSEVIER SCIENCE INC.2018: 553
- **Ring Finger Protein 207 Degrades T613M Kv11.1 Channel**
Ledford, H. A., Park, S., Sirish, P., Xu, W., Emigh, A. M., Timofeyev, V., Priest, J. R., Perez, M. V., Ashley, E. A., Yarov-Yarovoy, V., Zhang, X., Chiamvimonvat, N.
CELL PRESS.2018: 625A
- **First Trimester Plasma Glucose Values in Women without Diabetes are Associated with Risk for Congenital Heart Disease in Offspring.** *The Journal of pediatrics*

- Helle, E. I., Biegley, P. n., Knowles, J. W., Leader, J. B., Pendergrass, S. n., Yang, W. n., Reaven, G. R., Shaw, G. M., Ritchie, M. n., Priest, J. R.
2018; 195: 275–78
- **Birthweight, Type 2 Diabetes Mellitus, and Cardiovascular Disease: Addressing the Barker Hypothesis With Mendelian Randomization.** *Circulation. Genomic and precision medicine*
Zanetti, D. n., Tikkanen, E. n., Gustafsson, S. n., Priest, J. R., Burgess, S. n., Ingelsson, E. n.
2018; 11 (6): e002054
 - **Beyond Gene Panels: Whole Exome Sequencing for Diagnosis of Congenital Heart Disease.** *Circulation. Genomic and precision medicine*
Paige, S. L., Saha, P. n., Priest, J. R.
2018; 11 (3): e002097
 - **A primer to clinical genome sequencing.** *Current opinion in pediatrics*
Priest, J. R.
2017; 29 (5): 513-519
 - **Transcriptomic Profiling Maps Anatomically Patterned Subpopulations among Single Embryonic Cardiac Cells** *DEVELOPMENTAL CELL*
Li, G., Xu, A., Sim, S., Priest, J. R., Tian, X., Khan, T., Quertermous, T., Zhou, B., Tsao, P. S., Quake, S. R., Wu, S. M.
2016; 39 (4): 491-507
 - **Early somatic mosaicism is a rare cause of long-QT syndrome** *PROCEEDINGS OF THE NATIONAL ACADEMY OF SCIENCES OF THE UNITED STATES OF AMERICA*
Priest, J. R., Gawad, C., Kahlig, K. M., Yu, J. K., O'Hara, T., Boyle, P. M., Rajamani, S., Clark, M. J., Garcia, S. T., Ceresnak, S., Harris, J., Boyle, S., Dewey, et al
2016; 113 (41): 11555-11560
 - **Standards of Evidence and Mechanistic Inference in Autosomal Recessive Hypercholesterolemia** *ARTERIOSCLEROSIS THROMBOSIS AND VASCULAR BIOLOGY*
Priest, J. R., Knowles, J. W.
2016; 36 (8): 1465-1466
 - **Prepregnancy Diabetes and Offspring Risk of Congenital Heart Disease A Nationwide Cohort Study** *CIRCULATION*
Oyen, N., Diaz, L. J., Leirgul, E., Boyd, H. A., Priest, J., Mathiesen, E. R., Quertermous, T., Wohlfahrt, J., Melbye, M.
2016; 133 (23): 2243-2253
 - **De Novo and Rare Variants at Multiple Loci Support the Oligogenic Origins of Atrioventricular Septal Heart Defects.** *PLoS genetics*
Priest, J. R., Osoegawa, K., Mohammed, N., Nanda, V., Kundu, R., Schultz, K., Lammer, E. J., Girirajan, S., Scheetz, T., Waggott, D., Haddad, F., Reddy, S., Bernstein, et al
2016; 12 (4)
 - **Medical implications of technical accuracy in genome sequencing.** *Genome medicine*
Goldfeder, R. L., Priest, J. R., Zook, J. M., Grove, M. E., Waggott, D., Wheeler, M. T., Salit, M., Ashley, E. A.
2016; 8 (1): 24-?
 - **Maternal Midpregnancy Glucose Levels and Risk of Congenital Heart Disease in Offspring** *JAMA PEDIATRICS*
Priest, J. R., Yang, W., Reaven, G., Knowles, J. W., Shaw, G. M.
2015; 169 (12): 1112-1116
 - **Activating Mutations Affecting the Dbl Homology Domain of SOS2 Cause Noonan Syndrome** *HUMAN MUTATION*
Cordeddu, V., Yin, J. C., Gunnarsson, C., Virtanen, C., Drunat, S., Lepri, F., De Luca, A., Rossi, C., Ciolfi, A., Pugh, T. J., Bruselles, A., Priest, J. R., Pennacchio, et al
2015; 36 (11): 1080-1087
 - **Sequence to Medical Phenotypes: A Framework for Interpretation of Human Whole Genome DNA Sequence Data** *PLOS GENETICS*
Dewey, F. E., Grove, M. E., Priest, J. R., Waggott, D., Batra, P., Miller, C. L., Wheeler, M., Zia, A., Pan, C., Karzcewski, K. J., Miyake, C., Whirl-Carrillo, M., Klein, et al
2015; 11 (10)
 - **Sequence to Medical Phenotypes: A Framework for Interpretation of Human Whole Genome DNA Sequence Data.** *PLoS genetics*
Dewey, F. E., Grove, M. E., Priest, J. R., Waggott, D., Batra, P., Miller, C. L., Wheeler, M., Zia, A., Pan, C., Karzcewski, K. J., Miyake, C., Whirl-Carrillo, M., Klein, et al
2015; 11 (10)

- **Molecular diagnosis of long QT syndrome at 10 days of life by rapid whole genome sequencing** *HEART RHYTHM*
Priest, J. R., Ceresnak, S. R., Dewey, F. E., Malloy-Walton, L. E., Dunn, K., Grove, M. E., Perez, M. V., Maeda, K., Dubin, A. M., Ashley, E. A.
2014; 11 (10): 1707-1713
- **Molecular diagnosis of long QT syndrome at 10 days of life by rapid whole genome sequencing.** *Heart rhythm*
Priest, J. R., Ceresnak, S. R., Dewey, F. E., Malloy-Walton, L. E., Dunn, K., Grove, M. E., Perez, M. V., Maeda, K., Dubin, A. M., Ashley, E. A.
2014; 11 (10): 1707-1713
- **Self-reported history of childhood smoking is associated with an increased risk for peripheral arterial disease independent of lifetime smoking burden.** *PloS one*
Priest, J. R., Nead, K. T., Wehner, M. R., Cooke, J. P., Leeper, N. J.
2014; 9 (2)
- **Self-reported history of childhood smoking is associated with an increased risk for peripheral arterial disease independent of lifetime smoking burden.** *PloS one*
Priest, J. R., Nead, K. T., Wehner, M. R., Cooke, J. P., Leeper, N. J.
2014; 9 (2)
- **Ebstein anomaly and Trisomy 21: A rare association.** *Annals of pediatric cardiology*
Siehr, S. L., Punn, R., Priest, J. R., Lowenthal, A.
2014; 7 (1): 67-69
- **Triiodothyronine supplementation and cytokines during cardiopulmonary bypass in infants and children** *JOURNAL OF THORACIC AND CARDIOVASCULAR SURGERY*
Priest, J. R., Slee, A., Olson, A. K., Ledee, D., Morrish, F., Portman, M. A.
2012; 144 (4): 938-?
- **Rare copy number variants in isolated sporadic and syndromic atrioventricular septal defects** *AMERICAN JOURNAL OF MEDICAL GENETICS PART A*
Priest, J. R., Girirajan, S., Vu, T. H., Olson, A., Eichler, E. E., Portman, M. A.
2012; 158A (6): 1279-1284
- **Relationships of the Location and Content of Rounds to Specialty, Institution, Patient-Census, and Team Size** *PLOS ONE*
Priest, J. R., Bereksnyi, S., Hooper, K., Braddock, C. H.
2010; 5 (6)
- **A near null variant of 12/15-LOX encoded by a novel SNP in ALOX15 and the risk of coronary artery disease** *ATHEROSCLEROSIS*
Assimes, T. L., Knowles, J. W., Priest, J. R., Basu, A., Borchert, A., Volcik, K. A., Grove, M. L., Tabor, H. K., Southwick, A., Tabibiazar, R., Sidney, S., Boerwinkle, E., Go, et al
2008; 198 (1): 136-144
- **Common polymorphisms of ALOX5 and ALOX5AP and risk of coronary artery disease** *HUMAN GENETICS*
Assimes, T. L., Knowles, J. W., Priest, J. R., Basu, A., Volcik, K. A., Southwick, A., Tabor, H. K., Hartiala, J., Allayee, H., Grove, M. L., Tabibiazar, R., Sidney, S., Fortmann, et al
2008; 123 (4): 399-408
- **Brucellosis and sacroiliitis: A common presentation of an uncommon pathogen** *JOURNAL OF THE AMERICAN BOARD OF FAMILY MEDICINE*
Priest, J. R., Low, D., Wang, C., Bush, T.
2008; 21 (2): 158-161
- **Polymorphisms in hypoxia inducible factor 1 and the initial clinical presentation of coronary disease** *AMERICAN HEART JOURNAL*
Hlatky, M. A., Quertermous, T., Boothroyd, D. B., Priest, J. R., Glassford, A. J., Myers, R. M., Fortmann, S. P., Iribarren, C., Tabor, H. K., Assimes, T. L., Tibshirani, R. J., Go, A. S.
2007; 154 (6): 1035-1042
- **Comparative genomics: a tool to functionally annotate human DNA.** *Methods in molecular biology (Clifton, N.J.)*
Cheng, J., Priest, J. R., Pennacchio, L. A.
2007; 366: 229-251
- **Genomic sequencing of Pleistocene cave bears** *SCIENCE*
Noonan, J. P., Hofreiter, M., Smith, D., Priest, J. R., Rohland, N., Rabeder, G., KRAUSE, J., Dettler, J. C., Paabo, S., Rubin, E. M.

2005; 309 (5734): 597-600

- **Human-zebrafish non-coding conserved elements act in vivo to regulate transcription** *NUCLEIC ACIDS RESEARCH*

Shin, J. T., Priest, J. R., Ovcharenko, I., Ronco, A., Moore, R. K., Burns, C. G., MacRae, C. A.

2005; 33 (17): 5437-5445

- **The DNA sequence and comparative analysis of human chromosome 5** *NATURE*

Schmutz, J., Martin, J., Terry, A., Couronne, O., Grimwood, J., Lowry, S., Gordon, L. A., Scott, D., Xie, G., Huang, W., Hellsten, U., Tran-Gyamfi, M., She, et al

2004; 431 (7006): 268-274