



Rachel Knight Hopper

Clinical Professor, Pediatrics - Cardiology

CLINICAL OFFICE (PRIMARY)

- **Pediatric Heart Center**

725 Welch Rd

MC 5912

Palo Alto, CA 94304

Tel (650) 721-2121

Fax (650) 724-5650

Bio

BIO

Dr. Hopper is a Clinical Associate Professor in the Division of Pediatric Cardiology in the Department of Pediatrics. Dr. Hopper's clinical responsibilities are focused on the inpatient and outpatient care of children with pulmonary hypertension. She is the Associate Director of the Pediatric Pulmonary Hypertension Program based within the Children's Heart Center and part of the Vera Moulton Wall Center for Pulmonary Vascular Disease at Stanford. Dr. Hopper has a specific interest in pulmonary hypertension associated with prematurity and co-directs the multidisciplinary Cardiac and Respiratory care for Infants with Bronchopulmonary Dysplasia (CRIB) Program. She also works with the Pediatric Lung and Heart-Lung transplant program and co-directs the Center for Advanced Lung therapies (CEAL). She is also part of the Hereditary Hemorrhagic Telangiectasia (HHT) program at Stanford. Additionally, Dr. Hopper pursues clinical and translational research in the area of pulmonary hypertension in children.

CLINICAL FOCUS

- Pediatric Cardiology
- Pulmonary Hypertension

ACADEMIC APPOINTMENTS

- Clinical Professor, Pediatrics - Cardiology
- Member, Cardiovascular Institute
- Member, Maternal & Child Health Research Institute (MCHRI)

ADMINISTRATIVE APPOINTMENTS

- Associate Director, Pediatric Pulmonary Hypertension Program, Lucile Packard Children's Hospital Stanford, (2017- present)
- Co-director, CRIB Program, Lucile Packard Children's Hospital Stanford, (2018- present)

HONORS AND AWARDS

- Barst Fund Award, Pulmonary Hypertension Association (12/1/2016 - 11/30/2017)

BOARDS, ADVISORY COMMITTEES, PROFESSIONAL ORGANIZATIONS

- Co-chair, Clinical Trials Committee, Pediatric Pulmonary Hypertension Network (2014 - present)
- Member, Pediatric Task Force, Pulmonary Vascular Research Institute (2013 - present)
- Member, American Thoracic Society (2016 - present)
- Member, American Heart Association (2014 - present)

PROFESSIONAL EDUCATION

- BA, Pomona College , Molecular Biology (2001)
- Fellowship: Stanford University Pediatric Cardiology Fellowship (2014) CA
- Fellowship: Stanford University Pediatric Cardiology Fellowship (2013) CA
- Board Certification: Pediatrics, American Board of Pediatrics (2010)
- Residency: Boston Childrens Hospital Pediatric Residency (2010) MA
- Medical Education: University of Michigan Medical School (2007) MI
- Fellowship, Stanford (Lucile Packard Children's Hospital) , Pediatric Pulmonary Hypertension (2014)
- Fellowship, Stanford (Lucile Packard Children's Hospital) , Pediatric Cardiology (2013)
- Residency, Boston Children's Hospital (Boston Combined Residency Program in Pediatrics) , Pediatrics (2010)
- Board Certification: Pediatric Cardiology, American Board of Pediatrics (2014)
- MD, University of Michigan Medical School (2007)

LINKS

- Stanford Wall Center for Pulmonary Vascular Disease: <http://med.stanford.edu/wallcenter/our-team.html>
- Pediatric Pulmonary Hypertension Network: <http://www.pphnet.org>
- CRIB Program: <https://www.stanfordchildrens.org/en/service/cardiac-bpd>

Research & Scholarship

CURRENT RESEARCH AND SCHOLARLY INTERESTS

Current research interests include:

PH related to prematurity and bronchopulmonary dysplasia

Right heart failure in children with pulmonary hypertension, imaging and biomarkers

Pulmonary hypertension in children with congenital heart disease and/or heart failure

Clinical trials in children with PH

CLINICAL TRIALS

- Study to Evaluate Sotatercept (MK-7962) in Children With Pulmonary Arterial Hypertension (PAH) (MK-7962-008), Recruiting

Publications

PUBLICATIONS

- **An interdisciplinary consensus approach to pulmonary hypertension in developmental lung disease.** *The European respiratory journal*
Varghese, N. P., Austin, E. D., Galambos, C., Mullen, M. P., Yung, D., Guillerman, R. P., Vargas, S. O., Avitabile, C. M., Chartan, C. A., Cortes-Santiago, N., Ibach, M., Jackson, E. O., Jarrell, et al
2024; 64 (3)

- **Interleukin(IL)-1/IL-6-inhibitor-associated drug reaction with eosinophilia and systemic symptoms (DReSS) in systemic inflammatory illnesses.** *The journal of allergy and clinical immunology. In practice*
Saper, V. E., Tian, L., Verstegen, R. H., Conrad, C. K., Cidon, M., Hopper, R. K., Kuo, C. S., Osoegawa, K., Baszis, K., Bingham, C. A., Ferguson, I., Hahn, T., Horne, et al
2024
- **Younger age at initiation of subcutaneous treprostinil is associated with better response in pediatric Group 1 pulmonary arterial hypertension.** *Pulmonary circulation*
Kochanski, J. J., Feinstein, J. A., Ogawa, M., Ritter, V., Hopper, R. K., Adamson, G. T.
2024; 14 (1): e12328
- **Pulmonary Vasodilator Therapy in Pediatric Patients on Ventricular Assist Device Support: A Single-Center Experience and Proposal for Use.** *ASAIO journal (American Society for Artificial Internal Organs : 1992)*
Schramm, J. E., Dykes, J. C., Hopper, R. K., Feinstein, J. A., Rosenthal, D. N., Kameny, R. J.
2023
- **Measurement of Physical Activity by Actigraphy in Infants and Young Children with Pulmonary Arterial Hypertension.** *The Journal of pediatrics*
Avitabile, C. M., Yung, D., Handler, S., Hopper, R. K., Fineman, J., Freire, G., Varghese, N., Mullen, M., Krishnan, U. S., Austin, E., Silveira, L., Ivy, D. D.
2023: 113639
- **Editorial: Advances in the care of the pediatric pulmonary hypertension patient: from the neonate to the adolescent-young adult patient.** *Frontiers in pediatrics*
Avitabile, C. M., Hopper, R. K., Handler, S. S., Bates, A.
2023; 11: 1220070
- **International practice heterogeneity in pre-transplant management of pulmonary hypertension related to pediatric left heart disease.** *Pediatric transplantation*
Hopper, R. K., van der Have, O., Hollander, S. A., Dipchand, A. I., Perez de Sa, V., Feinstein, J. A., Tran-Lundmark, K.
2023: e14461
- **Role of left atrial hypertension in pulmonary hypertension associated with bronchopulmonary dysplasia** *Frontiers in Pediatrics*
Sullivan, R. T., Tandel, M. D., Bhombal, S., Adamson, G. T., Boothroyd, D. B., Tracy, M., Moy, A., Hopper, R. K.
2022: 1012136
- **Pulmonary Hypertension in Children with Down Syndrome: Results from the Pediatric Pulmonary Hypertension Network Registry.** *The Journal of pediatrics*
Hopper, R. K., Abman, S. H., Elia, E. G., Avitabile, C. M., Yung, D., Mullen, M. P., Austin, E. D., Bates, A., Handler, S. S., Feinstein, J. A., Ivy, D. D., Kinsella, J. P., Mandl, et al
2022
- **Subcutaneous Treprostinil Improves Surgical Candidacy for Next Stage Palliation in Single Ventricle Patients With High-Risk Hemodynamics.** *Seminars in thoracic and cardiovascular surgery*
Sullivan, R. T., Handler, S. S., Feinstein, J. A., Ogawa, M., Liu, E., Ma, M., Hopper, R. K., Norris, J., Hollander, S. A., Chen, S.
2022
- **Revisiting Arginine Therapy for Sickle Cell Acute Vaso-occlusive Painful Crisis.** *American journal of respiratory and critical care medicine*
Hopper, R. K., Gladwin, M. T.
2022
- **Cardiac Catheterization and Hemodynamics in a Multicenter Cohort of Children with Pulmonary Hypertension.** *Annals of the American Thoracic Society*
Rosenzweig, E. B., Bates, A., Mullen, M. P., Abman, S. H., Austin, E. D., Everett, A., Fineman, J., Feinstein, J., Hopper, R. K., Kinsella, J. P., Krishnan, U. S., Lu, M., Mandl, et al
1800
- **A case of acquired von Willebrand disease in severe pediatric pulmonary hypertension contributing to bleeding following reverse Potts shunt.** *Pulmonary Circulation*
Sullivan, R. T., Lo, C., Martin, E., Kameny, R. J., Hopper, R. K.
2022; 12 (e12042)
- **A case of acquired von Willebrand disease in severe pediatric pulmonary hypertension contributing to bleeding following reverse Potts shunt.** *Pulmonary circulation*

- Sullivan, R. T., Lo, C., Martin, E., Kameny, R. J., Hopper, R. K.
2022; 12 (1): e12042
- **Characterisation of Pediatric Pulmonary Hypertensive Vascular Disease from the PPHNet Registry.** *The European respiratory journal*
Abman, S. H., Mullen, M. P., Sleeper, L. A., Austin, E. D., Rosenzweig, E. B., Kinsella, J. P., Ivy, D., Hopper, R. K., Usha Raj, J., Fineman, J., Keller, R. L., Bates, A., Krishnan, et al
2021
 - **Pulmonary Arterial Hypertension: Diagnosis, Treatment, and Novel Advances.** *American journal of respiratory and critical care medicine*
Maron, B. A., Abman, S. H., Elliott, C. G., Frantz, R. P., Hopper, R. K., Horn, E. M., Nicolls, M. R., Shlobin, O. A., Shah, S. J., Kovacs, G. n., Olschewski, H. n., Rosenzweig, E. B.
2021
 - **Pharmacokinetics of Oral Treprostinil in Children with Pulmonary Arterial Hypertension.** *Journal of cardiovascular pharmacology*
Hopper, R. K., Ivy, D. D., Yung, D., Mullen, M. P., Hanna, B. D., Kirkpatrick, E., Hirsch, R., Austin, E. D., Fineman, J., Solum, D., Deng, C. Q., Feinstein, J. A.
2020
 - **Treprostinil improves hemodynamics and symptoms in children with mild pulmonary hypertension awaiting heart transplantation.** *Pediatric transplantation*
Hollander, S. A., Ogawa, M. T., Hopper, R. K., Liu, E. n., Chen, S. n., Rosenthal, D. N., Feinstein, J. A.
2020: e13742
 - **Pulmonary lung Doppler signals: normative data in a pediatric population compared with adults** *JOURNAL OF CLINICAL MONITORING AND COMPUTING*
Burstein, D. S., Hopper, R. K., McCarthy, E. K., Hall, K., Schatzberger, R., Palti, Y., Feinstein, J. A.
2019; 33 (6): 1055–60
 - **Inhaled Nitric Oxide Is Associated with Improved Oxygenation in a Subpopulation of Infants with Congenital Diaphragmatic Hernia and Pulmonary Hypertension.** *The Journal of pediatrics*
Lawrence, K. M., Monos, S., Adams, S., Herkert, L., Peranteau, W. H., Munson, D. A., Hopper, R. K., Avitabile, C. M., Rintoul, N. E., Hedrick, H. L.
2019
 - **Diminished right ventricular function at diagnosis of pulmonary hypertension is associated with mortality in bronchopulmonary dysplasia.** *Pulmonary circulation*
Altit, G., Bhombal, S., Feinstein, J., Hopper, R. K., Tacy, T. A.
2019; 9 (3): 2045894019878598
 - **Patent Ductus Arteriosus and the Effects of Its Late Closure in Preterm Infants with Severe Bronchopulmonary Dysplasia.** *Neonatology*
Ansems, S. M., Kirpalani, H., Mercer-Rosa, L., Wang, Y., Hopper, R. K., Fraga, M. V., Jensen, E. A.
2019: 1-8
 - **Histopathologic and Genetic Features of Alveolar Capillary Dysplasia with Atypical Late Presentation and Prolonged Survival** *JOURNAL OF PEDIATRICS*
Edwards, J. J., Murali, C., Pogoriler, J., Frank, D. B., Handler, S. S., Deardorff, M. A., Hopper, R. K.
2019; 210: 214+
 - **Diminished right ventricular function at diagnosis of pulmonary hypertension is associated with mortality in bronchopulmonary dysplasia** *PULMONARY CIRCULATION*
Altit, G., Bhombal, S., Feinstein, J., Hopper, R. K., Tacy, T. A.
2019; 9 (3)
 - **EXPRESS: Oral Treprostinil in Transition or as Add-on Therapy in Pediatric Pulmonary Arterial Hypertension.** *Pulmonary circulation*
Ivy, D. D., Feinstein, J., Yung, D., Mullen, M., Kirkpatrick, E. C., Hirsch, R., Austin, E., Fineman, J., Truong, U., Solum, D., Deng, C. Q., Hopper, R. K.
2019: 2045894019856471
 - **Death or resolution: the "natural history" of pulmonary hypertension in bronchopulmonary dysplasia** *JOURNAL OF PERINATOLOGY*
Altit, G., Bhombal, S., Hopper, R. K., Tacy, T. A., Feinstein, J.
2019; 39 (3): 415–25
 - **Time versus treatment: interpreting longitudinal data analysis of treprostinil** *Reply JOURNAL OF PEDIATRICS*
Hopper, R. K., Rogers, R., Lawrence, K. M., Hedrick, H. L.

2019; 206: 305-306

- **Pulmonary lung Doppler signals: normative data in a pediatric population compared with adults.** *Journal of clinical monitoring and computing*
Burststein, D. S., Hopper, R. K., McCarthy, E. K., Hall, K., Schatzberger, R., Palti, Y., Feinstein, J. A.
2019
- **Death or resolution: the "natural history" of pulmonary hypertension in bronchopulmonary dysplasia.** *Journal of perinatology : official journal of the California Perinatal Association*
Altit, G., Bhombal, S., Hopper, R. K., Tacy, T. A., Feinstein, J.
2019
- **Subcutaneous and Intravenous Treprostinil Pharmacokinetics in Children With Pulmonary Vascular Disease.** *Journal of cardiovascular pharmacology*
Hall, K. n., Ogawa, M. n., Sakarovich, C. n., Hopper, R. K., Adamson, G. T., Hanna, B. n., Ivy, D. D., Miller-Reed, K. n., Yung, D. n., McCarthy, E. n., Siehr-Handler, S. L., Feinstein, J. A.
2019; 73 (6): 383-93
- **Use of prostaglandin E1 to treat pulmonary hypertension in congenital diaphragmatic hernia.** *Journal of pediatric surgery*
Lawrence, K. M., Berger, K., Herkert, L., Franciscovich, C., O'Dea, C. L., Waqar, L. N., Partridge, E., Hanna, B. D., Peranteau, W. H., Avitabile, C. M., Hopper, R. K., Rintoul, N. E., Hedrick, et al
2019; 54 (1): 55-59
- **EXPRESS: Acute Vasoreactivity Testing in Pediatric Idiopathic Pulmonary Arterial Hypertension: an international Survey on Current Practice.** *Pulmonary circulation*
Caicedo, L. n., Hopper, R. K., Garcia, H. n., Ivy, D. D., Haag, D. n., Fineman, J. n., Humpi, T. n., Al-Tamimi, O. n., Feinstein, J. n., Berger, R. M., Berman-Rosenzweig, E. n., Kashour, T. n., Diaz, et al
2019: 2045894019857533
- **Racial and Ethnic Differences in Pediatric Pulmonary Hypertension: An Analysis of the Pediatric Pulmonary Hypertension Network Registry.** *The Journal of pediatrics*
Ong, M. S., Abman, S. n., Austin, E. D., Feinstein, J. A., Hopper, R. K., Krishnan, U. S., Mullen, M. P., Natter, M. D., Raj, J. U., Rosenzweig, E. B., Mandl, K. D.
2019
- **Reply.** *The Journal of pediatrics*
Hopper, R. K., Rogers, R., Lawrence, K. M., Hedrick, H. L.
2018
- **Pulmonary Vein Stenosis: Outcomes in Children with Congenital Heart Disease and Prematurity.** *Seminars in thoracic and cardiovascular surgery*
DiLorenzo, M. P., Santo, A., Rome, J. J., Zhang, H., Faerber, J. A., Mercer-Rosa, L., Hopper, R. K.
2018
- **Treprostinil Improves Persistent Pulmonary Hypertension Associated with Congenital Diaphragmatic Hernia** *JOURNAL OF PEDIATRICS*
Lawrence, K. M., Hedrick, H. L., Monk, H. M., Herkert, L., Waqar, L. N., Hanna, B. D., Peranteau, W. H., Rintoul, N. E., Hopper, R. K.
2018; 200: 44-49
- **Settling the Score in Pulmonary Hypertension?** *Pediatric critical care medicine : a journal of the Society of Critical Care Medicine and the World Federation of Pediatric Intensive and Critical Care Societies*
Hopper, R. K., Cornfield, D. N.
2018; 19 (8): 782-83
- **Settling the Score in Pulmonary Hypertension?*** *PEDIATRIC CRITICAL CARE MEDICINE*
Hopper, R. K., Cornfield, D. N.
2018; 19 (8): 782-83
- **Right ventricular function mirrors clinical improvement with use of prostacyclin analogues in pediatric pulmonary hypertension** *PULMONARY CIRCULATION*
Hopper, R. K., Wang, Y., DeMatteo, V., Santo, A., Kawut, S. M., Elci, O. U., Hanna, B. D., Mercer-Rosa, L.
2018; 8 (2): 2045894018759247
- **Central line replacement following infection does not improve reinfection rates in pediatric pulmonary hypertension patients receiving intravenous prostanoid therapy** *PULMONARY CIRCULATION*
McCarthy, E. K., Ogawa, M. T., Hopper, R. K., Feinstein, J. A., Gans, H. A.

2018; 8 (1): 2045893218754886

- **Subcutaneous treprostinil in pediatric patients with failing single-ventricle physiology** *JOURNAL OF HEART AND LUNG TRANSPLANTATION*
Handler, S. S., Ogawa, M. T., Hopper, R. K., Sakarovitch, C., Feinstein, J. A.
2018; 37 (2): 306–7
- **The Left Ventricle in Congenital Diaphragmatic Hernia: Implications for the Management of Pulmonary Hypertension.** *The Journal of pediatrics*
Kinsella, J. P., Steinhorn, R. H., Mullen, M. P., Hopper, R. K., Keller, R. L., Ivy, D. D., Austin, E. D., Krishnan, U. S., Rosenzweig, E. B., Fineman, J. R., Everett, A. D., Hanna, B. D., Humpl, et al
2018; 197: 17–22
- **Evaluation and Management of Pulmonary Hypertension in Children with Bronchopulmonary Dysplasia.** *The Journal of pediatrics*
Krishnan, U. n., Feinstein, J. A., Adatia, I. n., Austin, E. D., Mullen, M. P., Hopper, R. K., Hanna, B. n., Romer, L. n., Keller, R. L., Fineman, J. n., Steinhorn, R. n., Kinsella, J. P., Ivy, et al
2017; 188: 24–34.e1
- **Codependence of Bone Morphogenetic Protein Receptor 2 and Transforming Growth Factor- β in Elastic Fiber Assembly and Its Perturbation in Pulmonary Arterial Hypertension.** *Arteriosclerosis, thrombosis, and vascular biology*
Tojais, N. F., Cao, A. n., Lai, Y. J., Wang, L. n., Chen, P. I., Alcazar, M. A., de Jesus Perez, V. n., Hopper, R. K., Rhodes, C. J., Bill, M. A., Sakai, L. Y., Rabinovitch, M. n.
2017
- **Persistent Challenges in Pediatric Pulmonary Hypertension.** *Chest*
Hopper, R. K., Abman, S. H., Ivy, D. D.
2016; 150 (1): 226-36
- **In Pulmonary Arterial Hypertension, Reduced BMPR2 Promotes Endothelial-to-Mesenchymal Transition via HMGA1 and Its Target Slug** *CIRCULATION*
Hopper, R. K., Moonen, J. A., Diebold, I., Cao, A., Rhodes, C. J., Tojais, N. F., Hennigs, J. K., Gu, M., Wang, L., Rabinovitch, M.
2016; 133 (18): 1783-?
- **Recommendations for the Use of Inhaled Nitric Oxide Therapy in Premature Newborns with Severe Pulmonary Hypertension.** *The Journal of pediatrics*
Kinsella, J. P., Steinhorn, R. H., Krishnan, U. S., Feinstein, J. A., Adatia, I., Austin, E. D., Rosenzweig, E. B., Everett, A. D., Fineman, J. R., Hanna, B. D., Hopper, R. K., Humpl, T., Ivy, et al
2016; 170: 312-4
- **RNA Sequencing Analysis Detection of a Novel Pathway of Endothelial Dysfunction in Pulmonary Arterial Hypertension** *AMERICAN JOURNAL OF RESPIRATORY AND CRITICAL CARE MEDICINE*
Rhodes, C. J., Im, H., Cao, A., Hennigs, J. K., Wang, L., Sa, S., Chen, P., Nickel, N. P., Miyagawa, K., Hopper, R. K., Tojais, N. F., Li, C. G., Gu, et al
2015; 192 (3): 356-366
- **BMPR2 Preserves Mitochondrial Function and DNA during Reoxygenation to Promote Endothelial Cell Survival and Reverse Pulmonary Hypertension** *CELL METABOLISM*
Diebold, I., Hennigs, J. K., Miyagawa, K., Li, C. G., Nickel, N. P., Kaschwich, M., Cao, A., Wang, L., Reddy, S., Chen, P., Nakahira, K., Alcazar, M. A., Hopper, et al
2015; 21 (4): 596-608
- **Neonatal Pulmonary Arterial Hypertension and Noonan Syndrome: Two Fatal Cases with a Specific RAF1 Mutation** *AMERICAN JOURNAL OF MEDICAL GENETICS PART A*
Hopper, R. K., Feinstein, J. A., Manning, M. A., Benitz, W., Hudgins, L.
2015; 167A (4): 882-885
- **FK506 activates BMPR2, rescues endothelial dysfunction, and reverses pulmonary hypertension.** *journal of clinical investigation*
Spiekerkoetter, E., Tian, X., Cai, J., Hopper, R. K., Sudheendra, D., Li, C. G., El-Bizri, N., Sawada, H., Haghghat, R., Chan, R., Haghghat, L., de Jesus Perez, V., Wang, et al
2013; 123 (8): 3600-3613
- **FK506 activates BMPR2, rescues endothelial dysfunction, and reverses pulmonary hypertension.** *journal of clinical investigation*
Spiekerkoetter, E., Tian, X., Cai, J., Hopper, R. K., Sudheendra, D., Li, C. G., El-Bizri, N., Sawada, H., Haghghat, R., Chan, R., Haghghat, L., de Jesus Perez, V., Wang, et al
2013; 123 (8): 3600-3613

- **Use of P-32 To Study Dynamics of the Mitochondrial Phosphoproteome** *JOURNAL OF PROTEOME RESEARCH*
Aponte, A. M., Phillips, D., Hopper, R. K., Johnson, D. T., Harris, R. A., Blinova, K., Boja, E. S., French, S., Balaban, R. S.
2009; 8 (6): 2679-2695
- **Mitochondrial matrix phosphoproteome: Effect of extra mitochondrial calcium** *BIOCHEMISTRY*
Hopper, R. K., Carroll, S., Aponte, A. M., Johnson, D. T., French, S., Shen, R. F., Witzmann, F. A., Harris, R. A., Balaban, R. S.
2006; 45 (8): 2524-2536