Stanford

Edda Spiekerkoetter

Professor of Medicine (Pulmonary and Critical Care Medicine)

Medicine - Pulmonary, Allergy & Critical Care Medicine

CLINICAL OFFICE (PRIMARY)

- **Critical Care Medicine**

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  Stanford, CA 94305

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Bio

CLINICAL FOCUS

- Pulmonary Arterial Hypertension
- Hereditary Hemorrhagic Telangiectasia
- Chronic Right Heart Failure
- Arteriovenous Malformations
- Investigator Initiated Clinical Trials
- Rare Genetic Diseases
- Pulmonary Disease

ACADEMIC APPOINTMENTS

- Professor - University Medical Line, Medicine - Pulmonary, Allergy & Critical Care Medicine
- Member, Bio-X
- Member, Cardiovascular Institute
- Member, Maternal & Child Health Research Institute (MCHRI)

ADMINISTRATIVE APPOINTMENTS

- Director, HHT Center of Excellence, Stanford University, (2019- present)

HONORS AND AWARDS

- Mid-Career Women Faculty Leadership Seminar (MidWIMS), Association of American Medical Colleges (Dec 2022)
- High-Potential Women Leaders Program, Stanford University Graduate School of Business (Jan-Feb 2022)
- R01 - Understanding and targeting right ventricular adaptation and failure, NIH / NHLBI (2021 - 2025)
- Peer Reviewed Medical Investigator-Initiated Research Award: Arteriovenous Malformations, Department of Defense (2019 -2022)
- Peer Reviewed Medical Investigator-Initiated Research Award: BMPR2 and RV Function in CHD, Department of Defense (2017 - 2020)

- R01 - Targeting Novel BMPR2 modifiers in Pulmonary Hypertension with Repurposed Drugs, NIH / NHLBI (2016 - 2021)
- Young Physician Scientist Award, American Society of Clinical Investigation (April 2015)
- K08 Career development award, NIH (2011-2016)
- Supplemental award of the Pulmonary Hypertension Association (PHA), Pulmonary Hypertension Association (2011 - 2016)
- Helmholtz International Research Group Award, Helmholtz Zentrum Muenchen, Germany (2013-2016)
- Seed Grant - Phase II Clinical Trial, SPARK and Spectrum Stanford (2012-2014)
- Seed Grant - Phase II Clinical Trial, Wall Center of Pulmonary Vascular Disease (2012-2014)
- Seed Grant- BMP signaling in the RV, Cardiovascular Institute Stanford (2013-2014)
- Manuscript Award, Cardiovascular Institute at Stanford (Feb 2014)
- Poster Award, Excellence Cluster Cardio-Pulmonary System (ECCPS) and Pulmonary Vascular Research Institute (PVRI) (Jan 2014)
- Winner of Poster competition, Cardiovascular Institute Stanford (Sept 2012)
- Seed Grant - Small Molecule High Throughout Screen, Wall Center of Pulmonary Vascular Disease (2012)
- Postdoctoral Research Fellowship, Pulmonary Hypertension Association (2003-2005)

BOARDS, ADVISORY COMMITTEES, PROFESSIONAL ORGANIZATIONS

- Chair, 3CPR Early Career Committee, American Heart Association (2021 - 2023)
- Member of Cure HHT Research Network _ CZI Rare as ONE, Cure HHT.org (2022 - present)
- Nominating Committee, American Thoracic Society (2020 - 2021)
- Pulmonary Circulation Program Committee, American Thoracic Society (2013 - 2019)
- Member, Pulmonary Vascular Research Institute (2014 - present)
- Member at large, European Respiratory Society (2000 - present)
- Member at large, American Thoracic Society (1998 - present)

PROFESSIONAL EDUCATION

- Fellowship: Stanford University Pulmonary and Critical Care Fellowship (2009) CA
- Fellowship: Stanford University (2008) CA
- Fellowship: Lucile Packard Children's Hospital (2006) CA
- Residency: Medizinische Hochschule Hannover (2002) Germany
- Medical Education: University Hospital Freiburg (1995) Germany

PATENTS

- Edda Spiekerkoetter, Md Khadem Ali, Adam Andruska. "United States Use of Tyrosine Kinase Inhibitor for the Treatment of Hereditary Hemorrhagic Telangiectasia and Pulmonary Arterial Hypertension", Leland Stanford Junior University, Nov 22, 2022
- Edda Spiekerkoetter, Svenja Dannewitz, Xuefei Tian, Purvesh Khatri. "United States Patent CA3064275A1 Enzastaurin and Fragile Histidine Trial (FHIT) Increasing Agents for the Treatment of Pulmonary Hypertension", Leland Stanford Junior University, May 18, 2018
- Edda Spiekerkoetter, Marlene Rabinovitch, David Solow-Cordero, Phil Beachy. "United States Patent 61481317 Low-Dose FK506 for the treatment of Pulmonary Arterial Hypertension", Leland Stanford Junior University, May 2, 2012

LINKS

- My Lab site: http://med.stanford.edu/spiekerkoetterlab.html

Research & Scholarship

CURRENT RESEARCH AND SCHOLARLY INTERESTS

The limited treatment options for patients with end-stage pulmonary arterial hypertension (PAH) and right heart failure that I observed as a Pulmonary and Critical Care fellow at Hannover Medical School in Germany in the early 2000s were the reason I sought out basic research training in vascular biology under the mentorship of Dr. Marlene Rabinovitch at Stanford University.

As a physician scientist I strive to better understand the pathogenesis and underlying pathobiology of pulmonary and cardiovascular diseases such as PAH, arteriovenous malformations in hereditary hemorrhagic telangiectasia (HHT) and right heart failure to develop more effective treatments for these diseases. We use 3-D deep tissue imaging, mouse mutants and lineage tracing approaches to answer questions about the molecular and anatomic structure of blood vessels in concert with the extracellular matrix in the lung and the heart in health and disease.

A particular focus in my laboratory is the involvement of the BMPR2/TGF-b pathway in vascular biology. We use High-Throughput Screening techniques, induced pluripotent stem cells and bioinformatic approaches to identify and test repurposed and repositioned drugs that modulate BMPR2 signaling. By testing compounds in vitro and in vivo models of HHT and PAH, our ultimate goal is to identify candidates that would be promising to move forward into clinical trials.

Our discoveries have led to the initiation of a phase II clinical trial to test the safety, tolerability and efficacy of low-dose FK506 in PAH at Stanford (http://www.clinicaltrials.gov NCT01647945) as well as three patents for repurposed and repositioned drugs for the treatment of PAH and HHT.

My laboratory values close collaboration of clinicians, translational as well as basic scientists to apply biological concepts to disease models, driven by the notion that we first need to understand processes in health and disease before we can intervene. The ultimate objective of the lab is to successfully realize bench-to-bedside research for our patients.

CLINICAL TRIALS

- FK506 (Tacrolimus) in Pulmonary Arterial Hypertension, Not Recruiting

Teaching

COURSES

2025-26

- Cardiovascular and Pulmonary Sciences Seminar: MED 223 (Aut, Win)

2024-25

- Cardiovascular and Pulmonary Sciences Seminar: MED 223 (Aut, Win)

2023-24

- Cardiovascular and Pulmonary Sciences Seminar: MED 223 (Aut, Win)

2022-23

- Cardiovascular and Pulmonary Sciences Seminar: MED 223 (Aut, Win)

STANFORD ADVISEES

Postdoctoral Faculty Sponsor

Yue Qi

Publications

PUBLICATIONS

- **Somatic PIK3CA Activating Mutation in a Plexiform Lesion of a Patient With HHT and PAH.** *JACC. Case reports*
  Schimmel, K., Hallmark, T., DeBose-Scarlett, E., Qi, Y., Tan, S., Mastrodicasa, D., Hopper, R. K., Wu, J., Marchuk, D., Spiekerkoetter, E.
  2026: 106786

- **Recurrent somatic copy number alterations in resected cerebral cavernous malformations.** *Human genomics*
  Ressler, A. K., Debose-Scarlett, E., Fuenzalida, A., Lightle, R., Weinsheimer, S., Faughnan, M. E., Spiekerkoetter, E., Schimmel, K., Lawton, M., Kim, H., Awad, I., Marchuk, D. A.
  2025

- **Executive summary of the 15th HHT international scientific conference.** *Angiogenesis*
  Droege, F., Guilhem, A., Ricard, N., Spiekerkoetter, E., Hermann, R., Rossi, E., Bailly, S., Dupuis-Girod, S., Clancy, M., Friday, C.
  2025; 28 (Suppl 1): 61

- **Somatic activating mutation in Phosphoinositide 3-kinase in a plexiform lesion of a patient with hereditary hemorrhagic telangiectasia and pulmonary arterial hypertension**
  Schimmel, K., DeBose-Scarlett, E., Qi, Y., Tan, S., Mastrodicasa, D., Peruzzi, N., Tran-Lundmark, K., Wu, J., Marchuk, D., Spiekerkoetter, E.
  LIPPINCOTT WILLIAMS & WILKINS.2025

- **The effect of Microvascular Remodeling on Fluid Dynamics in the Pressure Overloaded Right Ventricle.** *American journal of physiology. Heart and circulatory physiology*
  Essafri, I., Ichimura, K., Ivy, D., Stenmark, K., Turton, H. A., Pyle, L., Spiekerkoetter, E., Kheyfets, V. O.
  2025

- **Transcriptional changes of the extracellular matrix in chronic thromboembolic pulmonary hypertension govern right ventricle remodeling and recovery.** *Nature cardiovascular research*
  Jafari, L., Wiedenroth, C. B., Kriechbaum, S. D., Grün, D., Chelladurai, P., Guenther, S., Kuenne, C., Späth, A. M., Cherian, A. V., Troidl, C., Wilhelm, J., Keranov, S., Keller, et al
  2025

- **Pathogenic Concepts in Pulmonary Arterial Hypertension Revisited - A Multigenerational Perspective.** *American journal of respiratory cell and molecular biology*
  Kwapiszewska, G., Rhodes, C., Rabinovitch, M., Simpson, C., Hassoun, P., Ichimura, K., Humbert, M., Spiekerkoetter, E.
  2025

- **Macrophages: key conductors behind perivascular inflammation and vascular remodeling in hypoxiainduced pulmonary hypertension** *JOURNAL OF CLINICAL INVESTIGATION*
  Spiekerkoetter, E.
  2025; 135 (6)

- **Developing Benchmarks in the Diagnosis and Treatment of Pulmonary Arterial Hypertension in a Tertiary, Academic Medical Center.** *Pulmonary circulation*
  Kholdani, C. A., Lee, J. H., Swenson, K. E., Liu, J., Hsi, A., Kudelko, K. T., Sweatt, A. J., Spiekerkoetter, E. F., De Jesus Perez, V., Rigdon, J., Hedlin, H., Andruska, A. M., Lyn, et al
  2025; 15 (1): e70063

- **Protocol for efficient generation of human artery and vein endothelial cells from pluripotent stem cells.** *STAR protocols*
  Loh, K. M., Zheng, S. L., Liu, K. J., Yin, Q., Amir-Ugokwe, Z. A., Jha, S. K., Qi, Y., Wazny, V. K., Nguyen, A. T., Chen, A., Njunkeng, F. M., Cheung, C., Spiekerkoetter, et al
  2024; 6 (1): 103494

- **Abnormalities in Coronary Microvasculature in Pulmonary Atresia With Intact Ventricular Septum.** *Circulation. Cardiovascular imaging*
  Vincent, A. G., Ichimura, K., Ichimura, S., Alden, A. O., Price, E., Salmi, D., Kushwaha, A., Spiekerkoetter, E., Reddy, S.
  2024: e017415

- **Increased BMP-Responsive Transcription Factors in Distinct Endothelial and Mesenchymal Cells in PAH.** *American journal of respiratory and critical care medicine*
  Andruska, A. M., Cantu Valadez, R., Ichimura, K., Chu, P., Zhang, T., Schimmel, K., Wang, L., Cao, A., Aldred, M. A., Spiekerkoetter, E.
  2024

- **Case report: A finding of PVOD and PAH in first degree relatives suggests shared heritable risk and overlapping features of both pulmonary vascular diseases.** *Respirology case reports*
  Winters, R., Forbes, L. M., Ivy, D., Cool, C., Park, B. D., Hountras, P., Badesch, D., Gu, S., Spiekerkoetter, E., Zamanian, R., LeierGluck, S., Bull, T. M.
  2024; 12 (11): e70064

- **Somatic mutations in arteriovenous malformations in hereditary hemorrhagic telangiectasia support a bi-allelic two-hit mutation mechanism of pathogenesis.** *American journal of human genetics*
  DeBose-Scarlett, E., Ressler, A. K., Gallione, C. J., Sapisochin Cantis, G., Friday, C., Weinsheimer, S., Schimmel, K., Spiekerkoetter, E., Kim, H., Gossage, J. R., Faughnan, M. E., Marchuk, D. A.
  2024

- **Tricuspid annular plane systolic excursion in pulmonary hypertension-Moving beyond the sector plane.** *Pulmonary circulation*
  Ichimura, K., Celestin, B. E., Bagherzadeh, S. P., Zamanian, R. T., Salerno, M., Spiekerkoetter, E., Haddad, F.
  2024; 14 (3): e12416

- **Cardiorenal Syndrome in Right Heart Failure Due to Pulmonary Arterial Hypertension-The Right Ventricle as a Therapeutic Target to Improve Renal Function.** *Cardiovascular drugs and therapy*
  Ichimura, K., Gross, A., Mathew, R. O., Salman, L., Reddy, S., Spiekerkoetter, E., Sidhu, M. S.
  2024

- **3D Imaging Reveals Complex Microvascular Remodeling in the Right Ventricle in Pulmonary Hypertension.** *Circulation research*
  Ichimura, K., Boehm, M., Andruska, A. M., Zhang, F., Schimmel, K., Bonham, S., Kabiri, A., Kheyfets, V. O., Ichimura, S., Reddy, S., Mao, Y., Zhang, T., Wang, et al
  2024

- **Rat microbial biogeography and age-dependent lactic acid bacteria in healthy lungs.** *Lab animal*
  Zhao, L., Cunningham, C. M., Andruska, A. M., Schimmel, K., Ali, M. K., Kim, D., Gu, S., Chang, J. L., Spiekerkoetter, E., Nicolls, M. R.
  2024; 53 (2): 43-55

- **MicroRNA-34a-Dependent Attenuation of Angiogenesis in Right Ventricular Failure.** *Journal of the American Heart Association*
  Reddy, S., Hu, D. Q., Zhao, M., Ichimura, S., Barnes, E. A., Cornfield, D. N., Alejandre Alcázar, M. A., Spiekerkoetter, E., Fajardo, G., Bernstein, D.
  2024: e029427

- **Characterizing the Spatiotemporal Transcriptomic Response of the Right Ventricle to Acute Pressure Overload.** *International journal of molecular sciences*
  Kheyfets, V. O., Kumar, S., Heerdt, P. M., Ichimura, K., Brown, R. D., Lucero, M., Essafri, I., Williams, S., Stenmark, K. R., Spiekerkoetter, E.
  2023; 24 (11)

- **Repetitive Schistosoma Exposure Causes Perivascular Lung Fibrosis and Persistent Pulmonary Hypertension.** *Clinical science (London, England : 1979)*
  Kumar, R., Lee, M. H., Kassa, B., Fonseca Balladares, D. C., Mickael, C., Sanders, L., Andruska, A., Kumar, M., Spiekerkoetter, E., Bandeira, A., Stenmark, K. R., Tuder, R. M., Graham, et al
  2023

- **Novel left ventricular mechanical index in pulmonary arterial hypertension.** *Pulmonary circulation*
  Ichimura, K., Santana, E. J., Kuznetsova, T., Cauwenberghs, N., Sabovčik, F., Chun, L., Francisco, N. L., Kheyfets, V. O., Salerno, M., Zamanian, R. T., Spiekerkoetter, E., Haddad, F.
  2023; 13 (2): e12216

- **Decreasing ELK3 expression improves Bone Morphogenetic Protein Receptor 2 signaling and pulmonary vascular cell function in PAH.** *bioRxiv : the preprint server for biology*
  Ali, M. K., Zhao, L., Perez, V. d., Nicolls, M. R., Spiekerkoetter, E. F.
  2023

- **PTPN1 Deficiency Modulates BMPR2 Signaling and Induces Endothelial Dysfunction in Pulmonary Arterial Hypertension.** *Cells*

Ali, M. K., Tian, X., Zhao, L., Schimmel, K., Rhodes, C. J., Wilkins, M. R., Nicolls, M. R., Spiekerkoetter, E. F.
2023; 12 (2)

- **The Human Respiratory Microbiome: Current Understandings and Future Directions.** *American journal of respiratory cell and molecular biology*
  Zhao, L., Luo, J. L., Ali, M. K., Spiekerkoetter, E., Nicolls, M. R.
  2022

- **The role of circular RNAs in pulmonary hypertension.** *The European respiratory journal*
  Ali, M. K., Schimmel, K., Zhao, L., Chen, C. K., Dua, K., Nicolls, M. R., Spiekerkoetter, E.
  2022

- **Relationship between impaired BMP signalling and clinical risk factors at early-stage vascular injury in the preterm infant.** *Thorax*
  Heydarian, M., Oak, P., Zhang, X., Kamgari, N., Kindt, A., Koschlig, M., Pritzke, T., Gonzalez-Rodriguez, E., Forster, K., Morty, R. E., Hafner, F., Hubener, C., Flemmer, et al
  2022

- **Cardiac Fibrosis in the Pressure Overloaded Left and Right Ventricle as a Therapeutic Target.** *Frontiers in cardiovascular medicine*
  Schimmel, K., Ichimura, K., Reddy, S., Haddad, F., Spiekerkoetter, E.
  2022; 9: 886553

- **Shunt-type plexiform lesions identified in the Sugen5416/Hypoxia rat model of pulmonary arterial hypertension using SPCT.** *The European respiratory journal*
  van der Have, O., Westoo, C., Ahrne, F., Tian, X., Ichimura, K., Dreier, T., Norvik, C., Kumar, M. E., Spiekerkoetter, E., Tran-Lundmark, K.
  2022

- **Arteriovenous Malformations-Current Understanding of the Pathogenesis with Implications for Treatment.** *International journal of molecular sciences*
  Schimmel, K., Ali, M. K., Tan, S. Y., Teng, J., Do, H. M., Steinberg, G. K., Stevenson, D. A., Spiekerkoetter, E.
  2021; 22 (16)

- **Role of Long Non-Coding RNAs in Pulmonary Arterial Hypertension.** *Cells*
  Han, Y., Ali, M. K., Dua, K., Spiekerkoetter, E., Mao, Y.
  2021; 10 (8)

- **Pulmonary arterial banding in mice may be a suitable model for studies on ventricular mechanics in pediatric pulmonary arterial hypertension.** *Journal of cardiovascular magnetic resonance : official journal of the Society for Cardiovascular Magnetic Resonance*
  Dufva, M. J., Boehm, M., Ichimura, K., Truong, U., Qin, X., Tabakh, J., Hunter, K. S., Ivy, D., Spiekerkoetter, E., Kheyfets, V. O.
  2021; 23 (1): 66

- **Improving Right Ventricular Function by Increasing BMP Signaling with FK506.** *American journal of respiratory cell and molecular biology*
  Boehm, M., Tian, X., Ali, M. K., Mao, Y., Ichimura, K., Zhao, M., Kuramoto, K., Dannewitz Prosseda, S., Fajardo, G., Dufva, M. J., Qin, X., Kheyfets, V. O., Bernstein, et al
  2021

- **Comparative analysis on the anti-inflammatory/immune effect of mesenchymal stem cell therapy for the treatment of pulmonary arterial hypertension.** *Scientific reports*
  Oh, S. n., Jang, A. Y., Chae, S. n., Choi, S. n., Moon, J. n., Kim, M. n., Spiekerkoetter, E. n., Zamanian, R. T., Yang, P. C., Hwang, D. n., Byun, K. n., Chung, W. J.
  2021; 11 (1): 2012

- **Severe Pulmonary Arterial Hypertension is Characterized by Increased Neutrophil Elastase and Relative Elafin Deficiency.** *Chest*
  Sweatt, A. J., Miyagawa, K., Rhodes, C. J., Taylor, S., Del Rosario, P. A., Hsi, A., Haddad, F., Spiekerkoetter, E., Bental-Roof, M., Bland, R. D., Swietlik, E. M., Gräf, S., Wilkins, et al
  2021

- **Promising therapeutic approaches in pulmonary arterial hypertension.** *Current opinion in pharmacology*
  Ali, M. K., Ichimura, K., Spiekerkoetter, E.
  2021; 59: 127-139

- **Novel Advances in Modifying BMPR2 Signaling in PAH.** *Genes*
  Dannewitz Prosseda, S., Ali, M. K., Spiekerkoetter, E.

2020; 12 (1)

- **Repurposing of medications for pulmonary arterial hypertension.** *Pulmonary circulation*
  Toshner, M., Spiekerkoetter, E., Bogaard, H., Hansmann, G., Nikkho, S., Prins, K. W.
  2020; 10 (4): 2045894020941494
- **The left ventricle undergoes biomechanical and gene expression changes in response to increased right ventricular pressure overload.** *Physiological reports*
  Kheyfets, V. O., Dufva, M. J., Boehm, M., Tian, X., Qin, X., Tabakh, J. E., Truong, U., Ivy, D., Spiekerkoetter, E.
  2020; 8 (9): e14347
- **The Hallmarks of Severe Pulmonary Arterial Hypertension: The Cancer Hypothesis - Ten years later.** *American journal of physiology. Lung cellular and molecular physiology*
  Cool, C. D., Kuebler, W. M., Bogaard, H. J., Spiekerkoetter, E., Nicolls, M. R., Voelkel, N. F.
  2020
- **A Notch3-Marked Subpopulation of Vascular Smooth Muscle Cells is the Cell of Origin for Occlusive Pulmonary Vascular Lesions.** *Circulation*
  Steffes, L. C., Froistad, A. A., Andruska, A. n., Boehm, M. n., McGlynn, M. n., Zhang, F. n., Zhang, W. n., Hou, D. n., Tian, X. n., Miquerol, L. n., Nadeau, K. n., Metzger, R. J., Spiekerkoetter, et al
  2020
- **Targeting BMPR2 Trafficking with Chaperones - An Important Step Towards Precision Medicine in Pulmonary Arterial Hypertension.** *American journal of respiratory cell and molecular biology*
  Andruska, A. n., Ali, K. n., Spiekerkoetter, E. n.
  2020
- **Targeted Proteomics of Right Heart Adaptation to Pulmonary Arterial Hypertension.** *The European respiratory journal*
  Amsallem, M. n., Sweatt, A. J., Arthur Ataam, J. n., Guihaire, J. n., Lecerf, F. n., Lambert, M. n., Ghigna, M. R., Ali, M. K., Mao, Y. n., Fadel, E. n., Rabinovitch, M. n., de Jesus Perez, V. n., Spiekerkoetter, et al
  2020
- **Delineating the molecular and histological events that govern right ventricular recovery using a novel mouse model of PA de-banding.** *Cardiovascular research*
  Boehm, M., Tian, X., Mao, Y., Ichimura, K., Dufva, M. J., Ali, K., Prosseda, S. D., Shi, Y., Kuramoto, K., Reddy, S., Kheyfets, V. O., Metzger, R. J., Spiekerkoetter, et al
  2019
- **Drug repositioning in pulmonary arterial hypertension: challenges and opportunities** *PULMONARY CIRCULATION*
  Grinnan, D., Trankle, C., Andruska, A., Bloom, B., Spiekerkoetter, E.
  2019; 9 (1)
- **EXPRESS: Drug Repositioning in Pulmonary Arterial Hypertension: Challenges and Opportunities.** *Pulmonary circulation*
  Grinnan, D., Trankle, C., Andruska, A., Bloom, B., Spiekerkoetter, E. F.
  2019: 2045894019832226
- **Hot topics in the mechanisms of pulmonary arterial hypertension disease: cancer-like pathobiology, the role of the adventitia, systemic involvement, and right ventricular failure.** *Pulmonary circulation*
  Spiekerkoetter, E. n., Goncharova, E. A., Guignabert, C. n., Stenmark, K. n., Kwapiszewska, G. n., Rabinovitch, M. n., Voelkel, N. n., Bogaard, H. J., Graham, B. n., Pullamsetti, S. S., Kuebler, W. M.
  2019; 9 (4): 2045894019889775
- **FHIT, a Novel Modifier Gene in Pulmonary Arterial Hypertension** *AMERICAN JOURNAL OF RESPIRATORY AND CRITICAL CARE MEDICINE*
  Prosseda, S., Tian, X., Kuramoto, K., Boehm, M., Sudheendra, D., Miyagawa, K., Zhang, F., Solow-Cordero, D., Saldivar, J. C., Austin, E. D., Loyd, J. E., Wheeler, L., Andruska, et al
  2019; 199 (1): 83-98
- **EXPRESS: Myocardial Bridge - An Unrecognized Cause of Chest Pain in Pulmonary Arterial Hypertension.** *Pulmonary circulation*
  Rajmohan, D. n., Sung, Y. K., Kudelko, K. n., Perez, V. i., Haddad, F. n., Tremmel, J. n., Schnittger, I. n., Zamanian, R. T., Spiekerkoetter, E. F.
  2019: 2045894019860738

- **New and Emerging Therapies for Pulmonary Arterial Hypertension.** *Annual review of medicine*
  Spiekerkoetter, E. n., Kawut, S. M., de Jesus Perez, V. A.
  2019; 70: 45–59

- **Exome data clouds the pathogenicity of genetic variants in Pulmonary Arterial Hypertension** *MOLECULAR GENETICS & GENOMIC MEDICINE*
  Abbasi, Y., Jabbari, J., Jabbari, R., Glinge, C., Izadyar, S., Spiekerkoetter, E., Zamanian, R. T., Carlsen, J., Tfelt-Hansen, J.
  2018; 6 (5): 835–44

- **Consequences of BMPR2 Deficiency in the Pulmonary Vasculature and Beyond: Contributions to Pulmonary Arterial Hypertension** *INTERNATIONAL JOURNAL OF MOLECULAR SCIENCES*
  Andruska, A., Spiekerkoetter, E.
  2018; 19 (9)

- **Consequences of BMPR2 Deficiency in the Pulmonary Vasculature and Beyond: Contributions to Pulmonary Arterial Hypertension.** *International journal of molecular sciences*
  Andruska, A., Spiekerkoetter, E.
  2018; 19 (9)

- **Fragile Histidine Triad (FHIT), a Novel Modifier Gene in Pulmonary Arterial Hypertension.** *American journal of respiratory and critical care medicine*
  Dannewitz Prosseda, S., Tian, X., Kuramoto, K., Boehm, M., Sudheendra, D., Miyagawa, K., Zhang, F., Solow-Cordero, D., Saldivar, J. C., Austin, E. D., Loyd, J. E., Wheeler, L., Andruska, et al
  2018

- **Update in Pulmonary Vascular Disease 2016 and 2017** *AMERICAN JOURNAL OF RESPIRATORY AND CRITICAL CARE MEDICINE*
  Brittain, E. L., Thennapan, T., Maron, B. A., Chan, S. Y., Austin, E. D., Spiekerkoetter, E., Bogaard, H. J., Guignabert, C., Paulin, R., Machado, R. F., Yu, P. B.
  2018; 198 (1): 13–23

- **New and Emerging Therapies for Pulmonary Arterial Hypertension** *Annual Reviews in medicine*
  Spiekerkoetter, E., Kawut, S., de Jesus Perez, V.
  2018

- **A Pro - Con debate: Current Controversies in PAH Pathogenesis at the American Thoracic Society International Meeting in 2017.** *American journal of physiology. Lung cellular and molecular physiology*
  Kuebler, W. M., Nicolls, M. R., Olschewski, A. n., Abe, K. n., Rabinovitch, M. n., Stewart, D. J., Chan, S. Y., Morrell, N. W., Archer, S. L., Spiekerkoetter, E. n.
  2018

- **Career Development of Young Physician-Scientists in the Cardiovascular Sciences: Perspective and Advice From the Early Career Committee of the Cardiopulmonary, Critical Care, and Resuscitation Council of the American Heart Association.** *Circulation research*
  Agarwal, S. n., Spiekerkoetter, E. n., Austin, E. D., de Jesus Perez, V. n., Dezfulian, C. n., Maron, B. A., Ryan, J. J., Starks, M. A., Yu, P. B., Bonnet, S. n., Perman, S. M.
  2018; 122 (10): 1330–33

- **Randomised placebo-controlled safety and tolerability trial of FK506 (tacrolimus) for pulmonary arterial hypertension** *EUROPEAN RESPIRATORY JOURNAL*
  Spiekerkoetter, E., Sung, Y. K., Sudheendra, D., Scott, V., Del Rosario, P., Bill, M., Haddad, F., Long-Boyle, J., Hedlin, H., Zamanian, R. T.
  2017; 50 (3)

- **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
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