

Optimizing Experimental Research in Respiratory Diseases: an ERS Statement Online supplement

References listed in the tables from the main published paper

1. Jiang H, Zhao PJ, Su D, Feng J, Ma SL. Paris saponin I induces apoptosis via increasing the Bax/Bcl-2 ratio and caspase-3 expression in gefitinib-resistant non-small cell lung cancer in vitro and in vivo. *Mol Med Rep.* 2014;9(6):2265-72. doi:10.3892/mmr.2014.2108.
2. Alberini JL, Boisgard R, Guillermet S, Siquier K, Jego B, Theze B et al. Multimodal In Vivo Imaging of Tumorigenesis and Response to Chemotherapy in a Transgenic Mouse Model of Mammary Cancer. *Mol Imaging Biol.* 2016;18(4):617-26. doi:10.1007/s11307-015-0916-7.
3. Sinclair SE, Molthen RC, Haworth ST, Dawson CA, Waters CM. Airway strain during mechanical ventilation in an intact animal model. *Am J Respir Crit Care Med.* 2007;176(8):786-94. doi:10.1164/rccm.200701-088OC.
4. Ebner B, Behm P, Jacoby C, Burghoff S, French BA, Schrader J et al. Early assessment of pulmonary inflammation by 19F MRI in vivo. *Circ Cardiovasc Imaging.* 2010;3(2):202-10. doi:10.1161/CIRCIMAGING.109.902312.
5. Kaushik SS, Freeman MS, Yoon SW, Liljeroth MG, Stiles JV, Roos JE et al. Measuring diffusion limitation with a perfusion-limited gas--hyperpolarized 129Xe gas-transfer spectroscopy in patients with idiopathic pulmonary fibrosis. *J Appl Physiol (1985).* 2014;117(6):577-85. doi:10.1152/jappphysiol.00326.2014.
6. Kuebler WM, Parthasarathi K, Lindert J, Bhattacharya J. Real-time lung microscopy. *J Appl Physiol (1985).* 2007;102(3):1255-64. doi:10.1152/jappphysiol.00786.2006.
7. Hoffman RM, Yang M. Dual-color, whole-body imaging in mice. *Nat Biotechnol.* 2005;23(7):790; author reply 1. doi:10.1038/nbt0705-790.
8. El-Amouri SS, Cao P, Miao C, Pan D. Secreted luciferase for in vivo evaluation of systemic protein delivery in mice. *Mol Biotechnol.* 2013;53(1):63-73. doi:10.1007/s12033-012-9519-6.
9. Gaertner M, Cimalla P, Meissner S, Kuebler WM, Koch E. Three-dimensional simultaneous optical coherence tomography and confocal fluorescence microscopy for investigation of lung tissue. *J Biomed Opt.* 2012;17(7):071310. doi:10.1117/1.JBO.17.7.071310.
10. Meissner S, Tabuchi A, Mertens M, Kuebler WM, Koch E. Virtual four-dimensional imaging of lung parenchyma by optical coherence tomography in mice. *J Biomed Opt.* 2010;15(3):036016. doi:10.1117/1.3425654.
11. Gerstmair A, Fois G, Innerbichler S, Dietl P, Felder E. A device for simultaneous live cell imaging during uni-axial mechanical strain or compression. *J Appl Physiol (1985).* 2009;107(2):613-20. doi:10.1152/jappphysiol.00012.2009.
12. Martin C, Uhlig S, Ullrich V. Videomicroscopy of methacholine-induced contraction of individual airways in precision-cut lung slices. *Eur Respir J.* 1996;9(12):2479-87.
13. Nickles HT, Sumkauskaitė M, Wang X, Wegner I, Puderbach M, Kuebler WM. Mechanical ventilation causes airway distension with proinflammatory sequelae in mice. *Am J Physiol Lung Cell Mol Physiol.* 2014;307(1):L27-37. doi:10.1152/ajplung.00288.2013.
14. Tabeling C, Yu H, Wang L, Ranke H, Goldenberg NM, Zabini D et al. CFTR and sphingolipids mediate hypoxic pulmonary vasoconstriction. *Proc Natl Acad Sci U S A.* 2015;112(13):E1614-23. doi:10.1073/pnas.1421190112.
15. Kizub IV, Strielkov IV, Shaifta Y, Becker S, Prieto-Lloret J, Snetkov VA et al. Gap junctions support the sustained phase of hypoxic pulmonary vasoconstriction by facilitating calcium sensitization. *Cardiovasc Res.* 2013;99(3):404-11. doi:10.1093/cvr/cvt129.
16. Huh D, Matthews BD, Mammoto A, Montoya-Zavala M, Hsin HY, Ingber DE. Reconstituting organ-level lung functions on a chip. *Science.* 2010;328(5986):1662-8. doi:10.1126/science.1188302.
17. Kuebler WM, Wittenberg C, Lee WL, Reppien E, Goldenberg NM, Lindner K et al. Thrombin stimulates albumin transcytosis in lung microvascular endothelial cells via activation of acid

- sphingomyelinase. *Am J Physiol Lung Cell Mol Physiol.* 2016;310(8):L720-32. doi:10.1152/ajplung.00157.2015.
18. Uhlig S, Yang Y, Waade J, Wittenberg C, Babendreyer A, Kuebler WM. Differential regulation of lung endothelial permeability in vitro and in situ. *Cellular physiology and biochemistry : international journal of experimental cellular physiology, biochemistry, and pharmacology.* 2014;34(1):1-19. doi:10.1159/000362980.
19. Dabral S, Tian X, Kojonazarov B, Savai R, Ghofrani HA, Weissmann N et al. Notch1 signalling regulates endothelial proliferation and apoptosis in pulmonary arterial hypertension. *Eur Respir J.* 2016;48(4):1137-49. doi:10.1183/13993003.00773-2015.
20. Kuebler WM. Vascular Calcification in Pulmonary Hypertension. Another Brick in the Wall. *Am J Respir Crit Care Med.* 2016;194(10):1187-9. doi:10.1164/rccm.201606-1170ED.
21. Eickelberg O, Kohler E, Reichenberger F, Bertschin S, Woodtli T, Erne P et al. Extracellular matrix deposition by primary human lung fibroblasts in response to TGF-beta1 and TGF-beta3. *Am J Physiol.* 1999;276(5 Pt 1):L814-24.
22. Yin J, Michalick L, Tang C, Tabuchi A, Goldenberg N, Dan Q et al. Role of Transient Receptor Potential Vanilloid 4 in Neutrophil Activation and Acute Lung Injury. *Am J Respir Cell Mol Biol.* 2016;54(3):370-83. doi:10.1165/rcmb.2014-0225OC.
23. Amatullah H, Shan Y, Beauchamp BL, Gali PL, Gupta S, Maron-Gutierrez T et al. DJ-1/PARK7 Impairs Bacterial Clearance in Sepsis. *Am J Respir Crit Care Med.* 2017;195(7):889-905. doi:10.1164/rccm.201604-0730OC.
24. Huang RT, Wu D, Meliton A, Oh MJ, Krause M, Lloyd JA et al. Experimental Lung Injury Reduces Kruppel-like Factor 2 to Increase Endothelial Permeability via Regulation of RAPGEF3-Rac1 Signaling. *Am J Respir Crit Care Med.* 2017;195(5):639-51. doi:10.1164/rccm.201604-0668OC.
25. Michalick L, Erfinanda L, Weichelt U, van der Giet M, Liedtke W, Kuebler WM. Transient Receptor Potential Vanilloid 4 and Serum Glucocorticoid-regulated Kinase 1 Are Critical Mediators of Lung Injury in Overventilated Mice In Vivo. *Anesthesiology.* 2017;126(2):300-11. doi:10.1097/ALN.0000000000001443.
26. Estrada R, Giridharan GA, Nguyen MD, Roussel TJ, Shakeri M, Parichehreh V et al. Endothelial cell culture model for replication of physiological profiles of pressure, flow, stretch, and shear stress in vitro. *Anal Chem.* 2011;83(8):3170-7. doi:10.1021/ac2002998.
27. Rahaman SO, Grove LM, Paruchuri S, Southern BD, Abraham S, Niese KA et al. TRPV4 mediates myofibroblast differentiation and pulmonary fibrosis in mice. *J Clin Invest.* 2014;124(12):5225-38. doi:10.1172/JCI75331.
28. Gerasimovskaya EV, Ahmad S, White CW, Jones PL, Carpenter TC, Stenmark KR. Extracellular ATP is an autocrine/paracrine regulator of hypoxia-induced adventitial fibroblast growth. Signaling through extracellular signal-regulated kinase-1/2 and the Egr-1 transcription factor. *J Biol Chem.* 2002;277(47):44638-50. doi:10.1074/jbc.M203012200.
29. Wang L, Taneja R, Wang W, Yao LJ, Veldhuizen RA, Gill SE et al. Human alveolar epithelial cells attenuate pulmonary microvascular endothelial cell permeability under septic conditions. *PLoS One.* 2013;8(2):e55311. doi:10.1371/journal.pone.0055311.
30. Colvin KL, Cripe PJ, Ivy DD, Stenmark KR, Yeager ME. Bronchus-associated lymphoid tissue in pulmonary hypertension produces pathologic autoantibodies. *Am J Respir Crit Care Med.* 2013;188(9):1126-36. doi:10.1164/rccm.201302-0403OC.
31. Charbonney E, Speight P, Masszi A, Nakano H, Kapus A. beta-catenin and Smad3 regulate the activity and stability of myocardin-related transcription factor during epithelial-myofibroblast transition. *Mol Biol Cell.* 2011;22(23):4472-85. doi:10.1091/mbc.E11-04-0335.
32. Tan Q, Choi KM, Sicard D, Tschumperlin DJ. Human airway organoid engineering as a step toward lung regeneration and disease modeling. *Biomaterials.* 2017;113:118-32. doi:10.1016/j.biomaterials.2016.10.046.
33. Stabler CT, Lecht S, Mondrinos MJ, Goulart E, Lazarovici P, Lelkes PI. Revascularization of decellularized lung scaffolds: principles and progress. *Am J Physiol Lung Cell Mol Physiol.* 2015;309(11):L1273-85. doi:10.1152/ajplung.00237.2015.

34. Looney MR, Thornton EE, Sen D, Lamm WJ, Glenny RW, Krummel MF. Stabilized imaging of immune surveillance in the mouse lung. *Nat Methods*. 2011;8(1):91-6. doi:10.1038/nmeth.1543.
35. Tabuchi A, Nickles HT, Kim M, Semple JW, Koch E, Brochard L et al. Acute Lung Injury Causes Asynchronous Alveolar Ventilation That Can Be Corrected by Individual Sighs. *Am J Respir Crit Care Med*. 2016;193(4):396-406. doi:10.1164/rccm.201505-0901OC.
36. Muders T, Luepschen H, Zinserling J, Greschus S, Fimmers R, Guenther U et al. Tidal recruitment assessed by electrical impedance tomography and computed tomography in a porcine model of lung injury*. *Crit Care Med*. 2012;40(3):903-11. doi:10.1097/CCM.0b013e318236f452.
37. Perez JR, Ybarra N, Chagnon F, Serban M, Lee S, Seuntjens J et al. Tracking of Mesenchymal Stem Cells with Fluorescence Endomicroscopy Imaging in Radiotherapy-Induced Lung Injury. *Sci Rep*. 2017;7:40748. doi:10.1038/srep40748.
38. Westphalen K, Gusarova GA, Islam MN, Subramanian M, Cohen TS, Prince AS et al. Sessile alveolar macrophages communicate with alveolar epithelium to modulate immunity. *Nature*. 2014;506(7489):503-6. doi:10.1038/nature12902.
39. Mertens M, Tabuchi A, Meissner S, Krueger A, Schirrmann K, Kertzsch U et al. Alveolar dynamics in acute lung injury: heterogeneous distension rather than cyclic opening and collapse. *Crit Care Med*. 2009;37(9):2604-11. doi:10.1097/CCM.0b013e3181a5544d.
40. Judkewitz B, Yang C. Axial standing-wave illumination frequency-domain imaging (SWIF). *Opt Express*. 2014;22(9):11001-10. doi:10.1364/OE.22.011001.
41. Shui B, Lee JC, Reining S, Lee FK, Kotlikoff MI. Optogenetic sensors and effectors: CHROMus-the Cornell Heart Lung Blood Institute Resource for Optogenetic Mouse Signaling. *Front Physiol*. 2014;5:428. doi:10.3389/fphys.2014.00428.
42. Rackley CR, Stripp BR. Building and maintaining the epithelium of the lung. *The Journal of clinical investigation*. 2012;122(8):2724-30. doi:10.1172/JCI60519.
43. Parent RA. *Comparative Biology of the Normal Lung*. Elsevier Science & Technology Books; 2015.
44. Linder CC. The influence of genetic background on spontaneous and genetically engineered mouse models of complex diseases. *Lab animal*. 2001;30(5):34-9.
45. Holmdahl R, Malissen B. The need for littermate controls. *European journal of immunology*. 2012;42(1):45-7. doi:10.1002/eji.201142048.
46. Florez-Vargas O, Brass A, Karystianis G, Bramhall M, Stevens R, Cruickshank S et al. Bias in the reporting of sex and age in biomedical research on mouse models. *eLife*. 2016;5. doi:10.7554/eLife.13615.
47. Sellers RS, Clifford CB, Treuting PM, Brayton C. Immunological variation between inbred laboratory mouse strains: points to consider in phenotyping genetically immunomodified mice. *Veterinary pathology*. 2012;49(1):32-43. doi:10.1177/0300985811429314.
48. Beura LK, Hamilton SE, Bi K, Schenkel JM, Odumade OA, Casey KA et al. Normalizing the environment recapitulates adult human immune traits in laboratory mice. *Nature*. 2016;532(7600):512-6. doi:10.1038/nature17655.
49. Wu BG, Segal LN. Lung Microbiota and Its Impact on the Mucosal Immune Phenotype. *Microbiology spectrum*. 2017;5(3). doi:10.1128/microbiolspec.BAD-0005-2016.
50. Birky TL, Bray MS. Understanding circadian gene function: animal models of tissue-specific circadian disruption. *IUBMB life*. 2014;66(1):34-41. doi:10.1002/iub.1241.
51. Speakman JR, Keijer J. Not so hot: Optimal housing temperatures for mice to mimic the thermal environment of humans. *Molecular metabolism*. 2012;2(1):5-9. doi:10.1016/j.molmet.2012.10.002.
52. Jaber SM, Hankenson FC, Heng K, McKinstry-Wu A, Kelz MB, Marx JO. Dose regimens, variability, and complications associated with using repeat-bolus dosing to extend a surgical plane of anesthesia in laboratory mice. *J Am Assoc Lab Anim Sci*. 2014;53(6):684-91.
53. Terrell RC. The invention and development of enflurane, isoflurane, sevoflurane, and desflurane. *Anesthesiology*. 2008;108(3):531-3. doi:10.1097/ALN.0b013e31816499cc.
54. Lawrance CC, Lucas EA, Clarke SL, Smith BJ, Kuvibidila S. Differential effects of isoflurane and CO2 inhalation on plasma levels of inflammatory markers associated with collagen-induced arthritis in

- DBA mice. *International immunopharmacology*. 2009;9(7-8):807-9. doi:10.1016/j.intimp.2009.03.017.
55. Angus DW, Baker JA, Mason R, Martin IJ. The potential influence of CO₂, as an agent for euthanasia, on the pharmacokinetics of basic compounds in rodents. *Drug metabolism and disposition: the biological fate of chemicals*. 2008;36(2):375-9. doi:10.1124/dmd.107.018879.
56. Carbone L, Carbone ET, Yi EM, Bauer DB, Lindstrom KA, Parker JM et al. Assessing cervical dislocation as a humane euthanasia method in mice. *J Am Assoc Lab Anim Sci*. 2012;51(3):352-6.
57. Fernandes CA, Vanbever R. Preclinical models for pulmonary drug delivery. *Expert opinion on drug delivery*. 2009;6(11):1231-45. doi:10.1517/17425240903241788.
58. Geraghty RJ, Capes-Davis A, Davis JM, Downward J, Freshney RI, Knezevic I et al. Guidelines for the use of cell lines in biomedical research. *British journal of cancer*. 2014;111(6):1021-46. doi:10.1038/bjc.2014.166.
59. Hughes P, Marshall D, Reid Y, Parkes H, Gelber C. The costs of using unauthenticated, over-passaged cell lines: how much more data do we need? *BioTechniques*. 2007;43(5):575, 7-8, 81-2 passim.
60. Booth AJ, Hadley R, Cornett AM, Dreffs AA, Matthes SA, Tsui JL et al. Acellular normal and fibrotic human lung matrices as a culture system for in vitro investigation. *American journal of respiratory and critical care medicine*. 2012;186(9):866-76. doi:10.1164/rccm.201204-0754OC.
61. Shojaie S, Ermini L, Ackerley C, Wang J, Chin S, Yeganeh B et al. Acellular lung scaffolds direct differentiation of endoderm to functional airway epithelial cells: requirement of matrix-bound HS proteoglycans. *Stem cell reports*. 2015;4(3):419-30. doi:10.1016/j.stemcr.2015.01.004.
62. Gilpin SE, Ren X, Okamoto T, Guyette JP, Mou H, Rajagopal J et al. Enhanced lung epithelial specification of human induced pluripotent stem cells on decellularized lung matrix. *The Annals of thoracic surgery*. 2014;98(5):1721-9; discussion 9. doi:10.1016/j.athoracsur.2014.05.080.
63. Ghaedi M, Calle EA, Mendez JJ, Gard AL, Balestrini J, Booth A et al. Human iPSC cell-derived alveolar epithelium repopulates lung extracellular matrix. *The Journal of clinical investigation*. 2013;123(11):4950-62. doi:10.1172/JCI68793.
64. Bonvillain RW, Scarritt ME, Pashos NC, Mayeux JP, Meshberger CL, Betancourt AM et al. Nonhuman primate lung decellularization and recellularization using a specialized large-organ bioreactor. *Journal of visualized experiments : JoVE*. 2013(82):e50825. doi:10.3791/50825.
65. Maniotis AJ, Chen CS, Ingber DE. Demonstration of mechanical connections between integrins, cytoskeletal filaments, and nucleoplasm that stabilize nuclear structure. *Proceedings of the National Academy of Sciences of the United States of America*. 1997;94(3):849-54.
66. Guillame-Gentil O, Semenov O, Roca AS, Groth T, Zahn R, Voros J et al. Engineering the extracellular environment: Strategies for building 2D and 3D cellular structures. *Advanced materials*. 2010;22(48):5443-62. doi:10.1002/adma.201001747.
67. Nikolic MZ, Rawlins EL. Lung Organoids and Their Use To Study Cell-Cell Interaction. *Current pathobiology reports*. 2017;5(2):223-31. doi:10.1007/s40139-017-0137-7.
68. Rubashkin MG, Ou G, Weaver VM. Deconstructing signaling in three dimensions. *Biochemistry*. 2014;53(13):2078-90. doi:10.1021/bi401710d.
69. Kirschvink N, Reinhold P. Use of alternative animals as asthma models. *Current drug targets*. 2008;9(6):470-84.
70. Reinero CR, DeClue AE, Rabinowitz P. Asthma in humans and cats: is there a common sensitivity to aeroallergens in shared environments? *Environmental research*. 2009;109(5):634-40. doi:10.1016/j.envres.2009.02.001.
71. Kirschvink N, Leemans J. Feline allergic asthma and experimental models: the sky is the limit. *Veterinary journal*. 2011;190(1):9-10. doi:10.1016/j.tvjl.2011.02.005.
72. Reinero CR. Advances in the understanding of pathogenesis, and diagnostics and therapeutics for feline allergic asthma. *Veterinary journal*. 2011;190(1):28-33. doi:10.1016/j.tvjl.2010.09.022.
73. Barrett EG, Rudolph K, Bowen LE, Muggenburg BA, Bice DE. Effect of inhaled ultrafine carbon particles on the allergic airway response in ragweed-sensitized dogs. *Inhalation toxicology*. 2003;15(2):151-65. doi:10.1080/08958370304474.

74. Royer CM, Rudolph K, Barrett EG. The neonatal susceptibility window for inhalant allergen sensitization in the atopically predisposed canine asthma model. *Immunology*. 2013;138(4):361-9. doi:10.1111/imm.12043.
75. Barrett EG. Maternal influence in the transmission of asthma susceptibility. *Pulmonary pharmacology & therapeutics*. 2008;21(3):474-84. doi:10.1016/j.pupt.2007.06.005.
76. Abraham WM. Modeling of asthma, COPD and cystic fibrosis in sheep. *Pulmonary pharmacology & therapeutics*. 2008;21(5):743-54. doi:10.1016/j.pupt.2008.01.010.
77. Leclere M, Lavoie-Lamoureux A, Gelinias-Lymburner E, David F, Martin JG, Lavoie JP. Effect of antigenic exposure on airway smooth muscle remodeling in an equine model of chronic asthma. *American journal of respiratory cell and molecular biology*. 2011;45(1):181-7. doi:10.1165/rcmb.2010-0300OC.
78. Bullone M, Lavoie JP. Asthma "of horses and men"--how can equine heaves help us better understand human asthma immunopathology and its functional consequences? *Molecular immunology*. 2015;66(1):97-105. doi:10.1016/j.molimm.2014.12.005.
79. Leclere M, Lavoie-Lamoureux A, Lavoie JP. Heaves, an asthma-like disease of horses. *Respirology*. 2011;16(7):1027-46. doi:10.1111/j.1440-1843.2011.02033.x.
80. Davis MS, McKiernan B, McCullough S, Nelson S, Jr., Mandsager RE, Willard M et al. Racing Alaskan sled dogs as a model of "ski asthma". *American journal of respiratory and critical care medicine*. 2002;166(6):878-82. doi:10.1164/rccm.200112-142BC.
81. Davis M, Williamson K, McKenzie E, Royer C, Payton M, Nelson S. Effect of training and rest on respiratory mechanical properties in racing sled dogs. *Medicine and science in sports and exercise*. 2005;37(2):337-41.
82. Gehlen H, Oey L, Rohn K, Bilzer T, Stadler P. Pulmonary dysfunction and skeletal muscle changes in horses with RAO. *Journal of veterinary internal medicine*. 2008;22(4):1014-21. doi:10.1111/j.1939-1676.2008.01111.x.
83. Theegarten D, Sachse K, Mentrup B, Fey K, Hotzel H, Anhenn O. Chlamydomphila spp. infection in horses with recurrent airway obstruction: similarities to human chronic obstructive disease. *Respiratory research*. 2008;9:14. doi:10.1186/1465-9921-9-14.
84. Hunter KS, Albiets JA, Lee PF, Lanning CJ, Lammers SR, Hofmeister SH et al. In vivo measurement of proximal pulmonary artery elastic modulus in the neonatal calf model of pulmonary hypertension: development and ex vivo validation. *Journal of applied physiology*. 2010;108(4):968-75. doi:10.1152/jappphysiol.01173.2009.
85. Tian L, Lammers SR, Kao PH, Albiets JA, Stenmark KR, Qi HJ et al. Impact of residual stretch and remodeling on collagen engagement in healthy and pulmonary hypertensive calf pulmonary arteries at physiological pressures. *Annals of biomedical engineering*. 2012;40(7):1419-33. doi:10.1007/s10439-012-0509-4.
86. Su Z, Tan W, Shandas R, Hunter KS. Influence of distal resistance and proximal stiffness on hemodynamics and RV afterload in progression and treatments of pulmonary hypertension: a computational study with validation using animal models. *Computational and mathematical methods in medicine*. 2013;2013:618326. doi:10.1155/2013/618326.
87. Bartels K, Brown RD, Fox DL, Bull TM, Neary JM, Dorosz JL et al. Right Ventricular Longitudinal Strain Is Depressed in a Bovine Model of Pulmonary Hypertension. *Anesthesia and analgesia*. 2016;122(5):1280-6. doi:10.1213/ANE.0000000000001215.
88. Holt TN, Callan RJ. Pulmonary arterial pressure testing for high mountain disease in cattle. *The Veterinary clinics of North America Food animal practice*. 2007;23(3):575-96. vii. doi:10.1016/j.cvfa.2007.08.001.
89. Naeije R. Pulmonary circulation at high altitude. *Respiration; international review of thoracic diseases*. 1997;64(6):429-34.
90. Niemarkt HJ, Kuypers E, Jellema R, Ophelders D, Hutten M, Nikiforou M et al. Effects of less-invasive surfactant administration on oxygenation, pulmonary surfactant distribution, and lung compliance in spontaneously breathing preterm lambs. *Pediatric research*. 2014;76(2):166-70. doi:10.1038/pr.2014.66.

91. Caminita F, van der Merwe M, Hance B, Krishnan R, Miller S, Buddington K et al. A preterm pig model of lung immaturity and spontaneous infant respiratory distress syndrome. *American journal of physiology Lung cellular and molecular physiology*. 2015;308(2):L118-29. doi:10.1152/ajplung.00173.2014.
92. Ballard-Croft C, Wang D, Sumpter LR, Zhou X, Zwischenberger JB. Large-animal models of acute respiratory distress syndrome. *The Annals of thoracic surgery*. 2012;93(4):1331-9. doi:10.1016/j.athoracsur.2011.06.107.
93. Pohlmann JR, Brant DO, Daul MA, Reoma JL, Kim AC, Osterholzer KR et al. Total liquid ventilation provides superior respiratory support to conventional mechanical ventilation in a large animal model of severe respiratory failure. *ASAIO journal*. 2011;57(1):1-8. doi:10.1097/MAT.0b013e3182018a9f.
94. Rogers CS, Stoltz DA, Meyerholz DK, Ostedgaard LS, Rokhlina T, Taft PJ et al. Disruption of the CFTR gene produces a model of cystic fibrosis in newborn pigs. *Science*. 2008;321(5897):1837-41. doi:10.1126/science.1163600.
95. Elferink RO, Beuers U. Are pigs more human than mice? *Journal of hepatology*. 2009;50(4):838-41. doi:10.1016/j.jhep.2008.12.014.
96. Welsh MJ, Rogers CS, Stoltz DA, Meyerholz DK, Prather RS. Development of a porcine model of cystic fibrosis. *Transactions of the American Clinical and Climatological Association*. 2009;120:149-62.
97. Yan Z, Stewart ZA, Sinn PL, Olsen JC, Hu J, McCray PB, Jr. et al. Ferret and pig models of cystic fibrosis: prospects and promise for gene therapy. *Human gene therapy Clinical development*. 2015;26(1):38-49. doi:10.1089/humc.2014.154.
98. Hendricks JC, Kline LR, Kovalski RJ, O'Brien JA, Morrison AR, Pack AI. The English bulldog: a natural model of sleep-disordered breathing. *Journal of applied physiology*. 1987;63(4):1344-50.
99. Lonergan RP, 3rd, Ware JC, Atkinson RL, Winter WC, Suratt PM. Sleep apnea in obese miniature pigs. *Journal of applied physiology*. 1998;84(2):531-6.
100. Otto P, Elschner M, Reinhold P, Kohler H, Streckert HJ, Philippou S et al. A model for respiratory syncytial virus (RSV) infection based on experimental aerosol exposure with bovine RSV in calves. *Comparative immunology, microbiology and infectious diseases*. 1996;19(2):85-97.
101. Gershwin LJ, Schelegle ES, Gunther RA, Anderson ML, Woolums AR, Larochele DR et al. A bovine model of vaccine enhanced respiratory syncytial virus pathophysiology. *Vaccine*. 1998;16(11-12):1225-36.
102. Gershwin LJ. Immunology of bovine respiratory syncytial virus infection of cattle. *Comparative immunology, microbiology and infectious diseases*. 2012;35(3):253-7. doi:10.1016/j.cimid.2012.01.005.
103. Grandin C, Lucas-Hourani M, Clavel M, Taborik F, Vabret A, Tangy F et al. Evidence for an intranasal immune response to human respiratory syncytial virus infection in cynomolgus macaques. *The Journal of general virology*. 2015;96(Pt 4):782-92. doi:10.1099/vir.0.000039.
104. Van Rhijn I, Godfroid J, Michel A, Rutten V. Bovine tuberculosis as a model for human tuberculosis: advantages over small animal models. *Microbes and infection*. 2008;10(7):711-5. doi:10.1016/j.micinf.2008.04.005.
105. Vanden Bush TJ, Rosenbusch RF. Characterization of the immune response to *Mycoplasma bovis* lung infection. *Veterinary immunology and immunopathology*. 2003;94(1-2):23-33.
106. Prysliak T, van der Merwe J, Lawman Z, Wilson D, Townsend H, van Drunen Littel-van den Hurk S et al. Respiratory disease caused by *Mycoplasma bovis* is enhanced by exposure to bovine herpes virus 1 (BHV-1) but not to bovine viral diarrhoea virus (BVDV) type 2. *The Canadian veterinary journal = La revue veterinaire canadienne*. 2011;52(11):1195-202.
107. Hermeyer K, Buchenau I, Thomasmeyer A, Baum B, Spargser J, Rosengarten R et al. Chronic pneumonia in calves after experimental infection with *Mycoplasma bovis* strain 1067: characterization of lung pathology, persistence of variable surface protein antigens and local immune response. *Acta veterinaria Scandinavica*. 2012;54:9. doi:10.1186/1751-0147-54-9.
108. Reinhold P, Ostermann C, Liebler-Tenorio E, Berndt A, Vogel A, Lambert J et al. A bovine model of respiratory *Chlamydia psittaci* infection: challenge dose titration. *PloS one*. 2012;7(1):e30125. doi:10.1371/journal.pone.0030125.

109. Ostermann C, Schroedl W, Schubert E, Sachse K, Reinhold P. Dose-dependent effects of *Chlamydia psittaci* infection on pulmonary gas exchange, innate immunity and acute-phase reaction in a bovine respiratory model. *Veterinary journal*. 2013;196(3):351-9. doi:10.1016/j.tvjl.2012.10.035.
110. Ostermann C, Ruttger A, Schubert E, Schrodl W, Sachse K, Reinhold P. Infection, disease, and transmission dynamics in calves after experimental and natural challenge with a Bovine *Chlamydia psittaci* isolate. *PloS one*. 2013;8(5):e64066. doi:10.1371/journal.pone.0064066.
111. Ostermann C, Linde S, Siegling-Vlitakis C, Reinhold P. Evaluation of pulmonary dysfunctions and acid-base imbalances induced by *Chlamydia psittaci* in a bovine model of respiratory infection. *Multidisciplinary respiratory medicine*. 2014;9(1):10. doi:10.1186/2049-6958-9-10.
112. Sachse K, Grossmann E, Berndt A, Schutt C, Henning K, Theegarten D et al. Respiratory chlamydial infection based on experimental aerosol challenge of pigs with *Chlamydia suis*. *Comparative immunology, microbiology and infectious diseases*. 2004;27(1):7-23. doi:10.1016/S0147-9571(02)00079-6.
113. Reinhold P, Hartmann H, Constable PD. Characterisation of acid-base abnormalities in pigs experimentally infected with *Chlamydia suis*. *Veterinary journal*. 2010;184(2):212-8. doi:10.1016/j.tvjl.2009.02.005.
114. Reinhold P, Kirschvink N, Theegarten D, Berndt A. An experimentally induced *Chlamydia suis* infection in pigs results in severe lung function disorders and pulmonary inflammation. *Veterinary research*. 2008;39(3):35. doi:10.1051/vetres:2008012.
115. Collie D, Govan J, Wright S, Thornton E, Tennant P, Smith S et al. A lung segmental model of chronic *Pseudomonas* infection in sheep. *PloS one*. 2013;8(7):e67677. doi:10.1371/journal.pone.0067677.
116. Martinez-Olondris P, Sibila O, Agusti C, Rigol M, Soy D, Esquinas C et al. An experimental model of pneumonia induced by methicillin-resistant *Staphylococcus aureus* in ventilated piglets. *The European respiratory journal*. 2010;36(4):901-6. doi:10.1183/09031936.00176709.
117. Soerensen KE, Olsen HG, Skovgaard K, Wiinberg B, Nielsen OL, Leifsson PS et al. Disseminated intravascular coagulation in a novel porcine model of severe *Staphylococcus aureus* sepsis fulfills human clinical criteria. *Journal of comparative pathology*. 2013;149(4):463-74. doi:10.1016/j.jcpa.2013.04.003.
118. Martinez-Olondris P, Rigol M, Torres A. What lessons have been learnt from animal models of MRSA in the lung? *The European respiratory journal*. 2010;35(1):198-201. doi:10.1183/09031936.00113908.
119. Sadowitz B, Roy S, Gatto LA, Habashi N, Nieman G. Lung injury induced by sepsis: lessons learned from large animal models and future directions for treatment. *Expert review of anti-infective therapy*. 2011;9(12):1169-78. doi:10.1586/eri.11.141.
120. Chalkias A, Spyropoulos V, Koutsovasilis A, Papalois A, Kouskouni E, Xanthos T. Cardiopulmonary Arrest and Resuscitation in Severe Sepsis and Septic Shock: A Research Model. *Shock*. 2015;43(3):285-91. doi:10.1097/SHK.0000000000000285.
121. Shull MM, Ormsby I, Kier AB, Pawlowski S, Diebold RJ, Yin M et al. Targeted disruption of the mouse transforming growth factor-beta 1 gene results in multifocal inflammatory disease. *Nature*. 1992;359(6397):693-9.
122. Kallapur S, Ormsby I, Doetschman T. Strain dependency of TGFbeta1 function during embryogenesis. *Mol Reprod Dev*. 1999;52(4):341-9. doi:10.1002/(SICI)1098-2795(199904)52:4<341::AID-MRD2>3.0.CO;2-N.
123. Dickson MC, Martin JS, Cousins FM, Kulkarni AB, Karlsson S, Akhurst RJ. Defective haematopoiesis and vasculogenesis in transforming growth factor-beta 1 knock out mice. *Development*. 1995;121(6):1845-54.
124. Harris KS, Zhang Z, McManus MT, Harfe BD, Sun X. Dicer function is essential for lung epithelium morphogenesis. *Proc Natl Acad Sci U S A*. 2006;103(7):2208-13. doi:10.1073/pnas.0510839103.
125. Xu Q, Tam M, Anderson SA. Fate mapping Nkx2.1-lineage cells in the mouse telencephalon. *J Comp Neurol*. 2008;506(1):16-29. doi:10.1002/cne.21529.

126. Rock JR, Barkauskas CE, Crouce MJ, Xue Y, Harris JR, Liang J et al. Multiple stromal populations contribute to pulmonary fibrosis without evidence for epithelial to mesenchymal transition. *Proc Natl Acad Sci U S A*. 2011;108(52):E1475-83. doi:10.1073/pnas.1117988108.
127. Okubo T, Knoepfler PS, Eisenman RN, Hogan BL. Nmyc plays an essential role during lung development as a dosage-sensitive regulator of progenitor cell proliferation and differentiation. *Development*. 2005;132(6):1363-74.
128. Perl AK, Wert SE, Nagy A, Lobe CG, Whitsett JA. Early restriction of peripheral and proximal cell lineages during formation of the lung. *Proc Natl Acad Sci U S A*. 2002;99(16):10482-7. doi:10.1073/pnas.152238499.
129. Takeda N, Jain R, LeBoeuf MR, Wang Q, Lu MM, Epstein JA. Interconversion between intestinal stem cell populations in distinct niches. *Science*. 2011;334(6061):1420-4. doi:10.1126/science.1213214.
130. Jain R, Barkauskas CE, Takeda N, Bowie EJ, Aghajanian H, Wang Q et al. Plasticity of Hopx(+) type I alveolar cells to regenerate type II cells in the lung. *Nat Commun*. 2015;6:6727. doi:10.1038/ncomms7727.
131. Li H, Cho SN, Evans CM, Dickey BF, Jeong JW, DeMayo FJ. Cre-mediated recombination in mouse Clara cells. *Genesis*. 2008;46(6):300-7. doi:10.1002/dvg.20396.
132. Perl AK, Wert SE, Loudy DE, Shan Z, Blair PA, Whitsett JA. Conditional recombination reveals distinct subsets of epithelial cells in trachea, bronchi, and alveoli. *Am J Respir Cell Mol Biol*. 2005;33(5):455-62. doi:10.1165/rcmb.2005-0180OC.
133. Rawlins EL, Okubo T, Xue Y, Brass DM, Auten RL, Hasegawa H et al. The role of Scgb1a1+ Clara cells in the long-term maintenance and repair of lung airway, but not alveolar, epithelium. *Cell Stem Cell*. 2009;4(6):525-34. doi:10.1016/j.stem.2009.04.002.
134. Zhang Y, Huang G, Shornick LP, Roswit WT, Shipley JM, Brody SL et al. A transgenic FOXJ1-Cre system for gene inactivation in ciliated epithelial cells. *Am J Respir Cell Mol Biol*. 2007;36(5):515-9. doi:10.1165/rcmb.2006-0475RC.
135. Rawlins EL, Ostrowski LE, Randell SH, Hogan BL. Lung development and repair: contribution of the ciliated lineage. *Proc Natl Acad Sci U S A*. 2007;104(2):410-7. doi:10.1073/pnas.0610770104.
136. Rock JR, Onaitis MW, Rawlins EL, Lu Y, Clark CP, Xue Y et al. Basal cells as stem cells of the mouse trachea and human airway epithelium. *Proc Natl Acad Sci U S A*. 2009;106(31):12771-5. doi:10.1073/pnas.0906850106.
137. Yu K, Xu J, Liu Z, Sosic D, Shao J, Olson EN et al. Conditional inactivation of FGF receptor 2 reveals an essential role for FGF signaling in the regulation of osteoblast function and bone growth. *Development*. 2003;130(13):3063-74.
138. Chen H, Zhuang F, Liu YH, Xu B, Del Moral P, Deng W et al. TGF-beta receptor II in epithelia versus mesenchyme plays distinct roles in the developing lung. *Eur Respir J*. 2008;32(2):285-95. doi:10.1183/09031936.00165407.
139. Holtwick R, Gotthardt M, Skryabin B, Steinmetz M, Potthast R, Zetsche B et al. Smooth muscle-selective deletion of guanylyl cyclase-A prevents the acute but not chronic effects of ANP on blood pressure. *Proc Natl Acad Sci U S A*. 2002;99(10):7142-7. doi:10.1073/pnas.102650499.
140. Wu Z, Yang L, Cai L, Zhang M, Cheng X, Yang X et al. Detection of epithelial to mesenchymal transition in airways of a bleomycin induced pulmonary fibrosis model derived from an alpha-smooth muscle actin-Cre transgenic mouse. *RespirRes*. 2007;8:1.
141. Wirth A, Benyó Z, Lukasova M, Leutgeb B, Wettschureck N, Gorbey S et al. G12-G13-LARG-mediated signaling in vascular smooth muscle is required for salt-induced hypertension. *Nat Med*. 2008;14(1):64-8. doi:10.1038/nm1666.
142. Zhang W, Menke DB, Jiang M, Chen H, Warburton D, Turcatel G et al. Spatial-temporal targeting of lung-specific mesenchyme by a Tbx4 enhancer. *BMC Biol*. 2013;11:111. doi:10.1186/1741-7007-11-111.
143. Kumar ME, Bogard PE, Espinoza FH, Menke DB, Kingsley DM, Krasnow MA. Mesenchymal cells. Defining a mesenchymal progenitor niche at single-cell resolution. *Science*. 2014;346(6211):1258810. doi:10.1126/science.1258810.

144. Foo SS, Turner CJ, Adams S, Compagni A, Aubyn D, Kogata N et al. Ephrin-B2 controls cell motility and adhesion during blood-vessel-wall assembly. *Cell*. 2006;124(1):161-73. doi:10.1016/j.cell.2005.10.034.
145. Greif DM, Kumar M, Lighthouse JK, Hum J, An A, Ding L et al. Radial construction of an arterial wall. *Dev Cell*. 2012;23(3):482-93. doi:10.1016/j.devcel.2012.07.009.
146. Zhu X, Bergles DE, Nishiyama A. NG2 cells generate both oligodendrocytes and gray matter astrocytes. *Development*. 2008;135(1):145-57. doi:10.1242/dev.004895.
147. Que J, Wilm B, Hasegawa H, Wang F, Bader D, Hogan BL. Mesothelium contributes to vascular smooth muscle and mesenchyme during lung development. *Proc Natl Acad Sci USA*. 2008;105(43):16626-30.
148. Kisanuki YY, Hammer RE, Miyazaki J, Williams SC, Richardson JA, Yanagisawa M. Tie2-Cre transgenic mice: a new model for endothelial cell-lineage analysis in vivo. *Dev Biol*. 2001;230(2):230-42.
149. Pham TH, Baluk P, Xu Y, Grigorova I, Bankovich AJ, Pappu R et al. Lymphatic endothelial cell sphingosine kinase activity is required for lymphocyte egress and lymphatic patterning. *J Exp Med*. 2010;207(1):17-27. doi:10.1084/jem.20091619.
150. Srinivasan RS, Dillard ME, Lagutin OV, Lin FJ, Tsai S, Tsai MJ et al. Lineage tracing demonstrates the venous origin of the mammalian lymphatic vasculature. *Genes Dev*. 2007;21(19):2422-32. doi:10.1101/gad.1588407.
151. Freem LJ, Escot S, Tannahill D, Druckenbrod NR, Thapar N, Burns AJ. The intrinsic innervation of the lung is derived from neural crest cells as shown by optical projection tomography in Wnt1-Cre;YFP reporter mice. *J Anat*. 2010;217(6):651-64. doi:10.1111/j.1469-7580.2010.01295.x.
152. Kim EJ, Ables JL, Dickel LK, Eisch AJ, Johnson JE. *Ascl1* (*Mash1*) defines cells with long-term neurogenic potential in subgranular and subventricular zones in adult mouse brain. *PLoS One*. 2011;6(3):e18472. doi:10.1371/journal.pone.0018472.
153. Li Y, Linnoila RI. Multidirectional differentiation of Achaete-Scute homologue-1-defined progenitors in lung development and injury repair. *Am J Respir Cell Mol Biol*. 2012;47(6):768-75. doi:10.1165/rcmb.2012-0027OC.
154. McKee AS, Munks MW, MacLeod MK, Fleenor CJ, Van Rooijen N, Kappler JW et al. Alum induces innate immune responses through macrophage and mast cell sensors, but these sensors are not required for alum to act as an adjuvant for specific immunity. *J Immunol*. 2009;183(7):4403-14. doi:10.4049/jimmunol.0900164.
155. Besnard AG, Guillou N, Tschopp J, Erard F, Couillin I, Iwakura Y et al. NLRP3 inflammasome is required in murine asthma in the absence of aluminum adjuvant. *Allergy*. 2011;66(8):1047-57.
156. Maes T, Provoost S, Lanckacker EA, Cataldo DD, Vanoirbeek JA, Nemery B et al. Mouse models to unravel the role of inhaled pollutants on allergic sensitization and airway inflammation. *Respir Res*. 2010;11:7.
157. Eisenbarth SC, Piggott DA, Huleatt JW, Visintin I, Herrick CA, Bottomly K. Lipopolysaccharide-enhanced, toll-like receptor 4-dependent T helper cell type 2 responses to inhaled antigen. *J Exp Med*. 2002;196(12):1645-51.
158. Delayre-Orthez C, de Blay F, Frossard N, Pons F. Dose-dependent effects of endotoxins on allergen sensitization and challenge in the mouse. *Clin Exp Allergy*. 2004;34(11):1789-95. doi:10.1111/j.1365-2222.2004.02082.x.
159. Garn H, Neves JF, Blumberg RS, Renz H. Effect of barrier microbes on organ-based inflammation. *J Allergy Clin Immunol*. 2013;131(6):1465-78. doi:10.1016/j.jaci.2013.04.031.
160. Bihouee T, Bouchaud G, Chesne J, Lair D, Rolland-Debord C, Braza F et al. Food allergy enhances allergic asthma in mice. *Respir Res*. 2014;15:142. doi:10.1186/s12931-014-0142-x.
161. Zhang Z, Hener P, Frossard N, Kato S, Metzger D, Li M et al. Thymic stromal lymphopoietin overproduced by keratinocytes in mouse skin aggravates experimental asthma. *Proc Natl Acad Sci U S A*. 2009;106(5):1536-41. doi:10.1073/pnas.0812668106.
162. Saunders SP, Moran T, Floudas A, Wurlod F, Kaszlikowska A, Salimi M et al. Spontaneous atopic dermatitis is mediated by innate immunity, with the secondary lung inflammation of the atopic

- march requiring adaptive immunity. *J Allergy Clin Immunol.* 2016;137(2):482-91. doi:10.1016/j.jaci.2015.06.045.
163. Deckers J, Sichien D, Plantinga M, Van Moorleghem J, Vanheerswynghels M, Hoste E et al. Epicutaneous sensitization to house dust mite allergen requires interferon regulatory factor 4-dependent dermal dendritic cells. *J Allergy Clin Immunol.* 2017;in press, doi:10.1016/j.jaci.2016.12.970. doi:10.1016/j.jaci.2016.12.970.
164. Sonar SS, Ehmke M, Marsh LM, Dietze J, Dudda JC, Conrad ML et al. Clara cells drive eosinophil accumulation in allergic asthma. *Eur Respir J.* 2012;39(2):429-38. doi:10.1183/09031936.00197810.
165. Nolin JD, Ogden HL, Lai Y, Altemeier WA, Frevert CW, Bollinger JG et al. Identification of Epithelial Phospholipase A2 Receptor 1 as a Potential Target in Asthma. *Am J Respir Cell Mol Biol.* 2016;55(6):825-36. doi:10.1165/rcmb.2015-01500C.
166. de Vries M, Hesse L, Jonker MR, van den Berge M, van Oosterhout AJ, Heijink IH et al. Pim1 kinase activity preserves airway epithelial integrity upon house dust mite exposure. *Am J Physiol Lung Cell Mol Physiol.* 2015;309(11):L1344-53. doi:10.1152/ajplung.00043.2015.
167. Le Cras TD, Acciani TH, Mushaben EM, Kramer EL, Pastura PA, Hardie WD et al. Epithelial EGF receptor signaling mediates airway hyperreactivity and remodeling in a mouse model of chronic asthma. *Am J Physiol Lung Cell Mol Physiol.* 2011;300(3):L414-21. doi:10.1152/ajplung.00346.2010.
168. Anagnostopoulou P, Dai L, Schatterny J, Hirtz S, Duerr J, Mall MA. Allergic airway inflammation induces a pro-secretory epithelial ion transport phenotype in mice. *Eur Respir J.* 2010;36(6):1436-47. doi:10.1183/09031936.00181209.
169. Loser S, Gregory LG, Zhang Y, Schaefer K, Walker SA, Buckley J et al. Pulmonary ORMDL3 is critical for induction of Alternaria-induced allergic airways disease. *J Allergy Clin Immunol.* 2017;139(5):1496-507 e3. doi:10.1016/j.jaci.2016.07.033.
170. Jacobsen EA, Lesuer WE, Willetts L, Zellner KR, Mazzolini K, Antonios N et al. Eosinophil activities modulate the immune/inflammatory character of allergic respiratory responses in mice. *Allergy.* 2014;69(3):315-27. doi:10.1111/all.12321.
171. Jacobsen EA, Ochkur SI, Doyle AD, LeSuer WE, Li W, Protheroe CA et al. Lung Pathologies in a Chronic Inflammation Mouse Model Are Independent of Eosinophil Degranulation. *Am J Respir Crit Care Med.* 2017;195(10):1321-32. doi:10.1164/rccm.201606-1129OC.
172. Mesnil C, Raulier S, Paulissen G, Xiao X, Birrell MA, Pirottin D et al. Lung-resident eosinophils represent a distinct regulatory eosinophil subset. *J Clin Invest.* 2016;126(9):3279-95. doi:10.1172/JCI85664.
173. Nabe T, Hosokawa F, Matsuya K, Morishita T, Ikedo A, Fujii M et al. Important role of neutrophils in the late asthmatic response in mice. *Life Sci.* 2011;88(25-26):1127-35. doi:10.1016/j.lfs.2011.04.003.
174. Hogan MB, Weissman DN, Hubbs AF, Gibson LF, Piktel D, Landreth KS. Regulation of eosinophilopoiesis in a murine model of asthma. *J Immunol.* 2003;171(5):2644-51.
175. Webb DC, McKenzie AN, Matthaei KI, Rothenberg ME, Foster PS. Distinct spatial requirement for eosinophil-induced airways hyperreactivity. *Immunol Cell Biol.* 2001;79(2):165-9. doi:10.1046/j.1440-1711.2001.00989.x.
176. Daubeuf F, Frossard N. Eosinophils and the ovalbumin mouse model of asthma. *Methods Mol Biol.* 2014;1178:283-93. doi:10.1007/978-1-4939-1016-8_24.
177. Schmit D, Le DD, Heck S, Bischoff M, Tschernig T, Herr C et al. Allergic airway inflammation induces migration of mast cell populations into the mouse airway. *Cell Tissue Res.* 2017. doi:10.1007/s00441-017-2597-9.
178. Williams CM, Galli SJ. Mast cells can amplify airway reactivity and features of chronic inflammation in an asthma model in mice. *J Exp Med.* 2000;192(3):455-62.
179. Liu T, Kanaoka Y, Barrett NA, Feng C, Garofalo D, Lai J et al. Aspirin-Exacerbated Respiratory Disease Involves a Cysteinyl Leukotriene-Driven IL-33-Mediated Mast Cell Activation Pathway. *J Immunol.* 2015;195(8):3537-45. doi:10.4049/jimmunol.1500905.

180. Li S, Aliyeva M, Daphtary N, Martin RA, Poynter ME, Kostin SF et al. Antigen-induced mast cell expansion and bronchoconstriction in a mouse model of asthma. *Am J Physiol Lung Cell Mol Physiol*. 2014;306(2):L196-206. doi:10.1152/ajplung.00055.2013.
181. Sugimoto K, Kudo M, Sundaram A, Ren X, Huang K, Bernstein X et al. The alphavbeta6 integrin modulates airway hyperresponsiveness in mice by regulating intraepithelial mast cells. *J Clin Invest*. 2012;122(2):748-58. doi:10.1172/JCI58815.
182. Yu M, Eckart MR, Morgan AA, Mukai K, Butte AJ, Tsai M et al. Identification of an IFN-gamma/mast cell axis in a mouse model of chronic asthma. *J Clin Invest*. 2011;121(8):3133-43. doi:10.1172/JCI43598.
183. Nakae S, Ho LH, Yu M, Monteforte R, Iikura M, Suto H et al. Mast cell-derived TNF contributes to airway hyperreactivity, inflammation, and TH2 cytokine production in an asthma model in mice. *J Allergy Clin Immunol*. 2007;120(1):48-55. doi:10.1016/j.jaci.2007.02.046.
184. Reuter S, Heinz A, Sieren M, Wiewrodt R, Gelfand EW, Stassen M et al. Mast cell-derived tumour necrosis factor is essential for allergic airway disease. *Eur Respir J*. 2008;31(4):773-82. doi:10.1183/09031936.00058907.
185. Zaslona Z, Przybranowski S, Wilke C, van Rooijen N, Teitz-Tennenbaum S, Osterholzer JJ et al. Resident alveolar macrophages suppress, whereas recruited monocytes promote, allergic lung inflammation in murine models of asthma. *J Immunol*. 2014;193(8):4245-53. doi:10.4049/jimmunol.1400580.
186. Bang BR, Chun E, Shim EJ, Lee HS, Lee SY, Cho SH et al. Alveolar macrophages modulate allergic inflammation in a murine model of asthma. *Exp Mol Med*. 2011;43(5):275-80. doi:10.3858/em.2011.43.5.028.
187. Hammad H, Plantinga M, Deswarte K, Pouliot P, Willart MA, Kool M et al. Inflammatory dendritic cells--not basophils--are necessary and sufficient for induction of Th2 immunity to inhaled house dust mite allergen. *J Exp Med*. 2010;207(10):2097-111. doi:10.1084/jem.20101563.
188. Yang M, Kumar RK, Foster PS. Interferon-gamma and pulmonary macrophages contribute to the mechanisms underlying prolonged airway hyperresponsiveness. *Clin Exp Allergy*. 2010;40(1):163-73. doi:10.1111/j.1365-2222.2009.03393.x.
189. Bedoret D, Wallemacq H, Marichal T, Desmet C, Quesada Calvo F, Henry E et al. Lung interstitial macrophages alter dendritic cell functions to prevent airway allergy in mice. *J Clin Invest*. 2009;119(12):3723-38. doi:10.1172/JCI39717.
190. van Rijt LS, Jung S, Kleinjan A, Vos N, Willart M, Duez C et al. In vivo depletion of lung CD11c+ dendritic cells during allergen challenge abrogates the characteristic features of asthma. *J Exp Med*. 2005;201(6):981-91. doi:10.1084/jem.20042311.
191. Lambrecht BN, Salomon B, Klatzmann D, Pauwels RA. Dendritic cells are required for the development of chronic eosinophilic airway inflammation in response to inhaled antigen in sensitized mice. *J Immunol*. 1998;160(8):4090-7.
192. Moran TP, Nakano K, Whitehead GS, Thomas SY, Cook DN, Nakano H. Inhaled house dust programs pulmonary dendritic cells to promote type 2 T-cell responses by an indirect mechanism. *Am J Physiol Lung Cell Mol Physiol*. 2015;309(10):L1208-18. doi:10.1152/ajplung.00256.2015.
193. Le DD, Rochlitz S, Fischer A, Heck S, Tschernig T, Sester M et al. Allergic airway inflammation induces the migration of dendritic cells into airway sensory ganglia. *Respir Res*. 2014;15:73. doi:10.1186/1465-9921-15-73.
194. Soroosh P, Doherty TA, Duan W, Mehta AK, Choi H, Adams YF et al. Lung-resident tissue macrophages generate Foxp3+ regulatory T cells and promote airway tolerance. *J Exp Med*. 2013;210(4):775-88. doi:10.1084/jem.20121849.
195. Marichal T, Bedoret D, Mesnil C, Pichavant M, Goriely S, Trottein F et al. Interferon response factor 3 is essential for house dust mite-induced airway allergy. *J Allergy Clin Immunol*. 2010;126(4):836-44 e13. doi:10.1016/j.jaci.2010.06.009.
196. Jandl K, Stacher E, Balint Z, Sturm EM, Maric J, Peinhaupt M et al. Activated prostaglandin D2 receptors on macrophages enhance neutrophil recruitment into the lung. *J Allergy Clin Immunol*. 2016;137(3):833-43. doi:10.1016/j.jaci.2015.11.012.

197. Lee YG, Jeong JJ, Nyenhuis S, Berdyshev E, Chung S, Ranjan R et al. Recruited alveolar macrophages, in response to airway epithelial-derived monocyte chemoattractant protein 1/CCl₂, regulate airway inflammation and remodeling in allergic asthma. *Am J Respir Cell Mol Biol*. 2015;52(6):772-84. doi:10.1165/rcmb.2014-0255OC.
198. Tang C, Inman MD, van Rooijen N, Yang P, Shen H, Matsumoto K et al. Th type 1-stimulating activity of lung macrophages inhibits Th2-mediated allergic airway inflammation by an IFN-gamma-dependent mechanism. *J Immunol*. 2001;166(3):1471-81.
199. Matsuoka K, Shitara H, Taya C, Kohno K, Kikkawa Y, Yonekawa H. Novel basophil- or eosinophil-depleted mouse models for functional analyses of allergic inflammation. *PLoS One*. 2013;8(4):e60958. doi:10.1371/journal.pone.0060958.
200. Nabe T, Matsuya K, Akamizu K, Fujita M, Nakagawa T, Shioe M et al. Roles of basophils and mast cells infiltrating the lung by multiple antigen challenges in asthmatic responses of mice. *Br J Pharmacol*. 2013;169(2):462-76. doi:10.1111/bph.12154.
201. Vissers JL, van Esch BC, Hofman GA, van Oosterhout AJ. Macrophages induce an allergen-specific and long-term suppression in a mouse asthma model. *Eur Respir J*. 2005;26(6):1040-6. doi:10.1183/09031936.05.00089304.
202. Ballesteros-Tato A, Randall TD, Lund FE, Spolski R, Leonard WJ, Leon B. T Follicular Helper Cell Plasticity Shapes Pathogenic T Helper 2 Cell-Mediated Immunity to Inhaled House Dust Mite. *Immunity*. 2016;44(2):259-73. doi:10.1016/j.immuni.2015.11.017.
203. Gavett SH, Chen X, Finkelman F, Wills-Karp M. Depletion of murine CD4⁺ T lymphocytes prevents antigen-induced airway hyperreactivity and pulmonary eosinophilia. *Am J Respir Cell Mol Biol*. 1994;10(6):587-93. doi:10.1165/ajrcmb.10.6.8003337.
204. Dullaers M, Schuijs MJ, Willart M, Fierens K, Van Moorlegheem J, Hammad H et al. House dust mite-driven asthma and allergen-specific T cells depend on B cells when the amount of inhaled allergen is limiting. *J Allergy Clin Immunol*. 2017;140(1):76-88 e7. doi:10.1016/j.jaci.2016.09.020.
205. Peters M, Kohler-Bachmann S, Lenz-Habijan T, Bufe A. Influence of an Allergen-Specific Th17 Response on Remodeling of the Airways. *Am J Respir Cell Mol Biol*. 2016;54(3):350-8. doi:10.1165/rcmb.2014-0429OC.
206. Vroman H, Bergen IM, Li BW, van Hulst JA, Lukkes M, van Uden D et al. Development of eosinophilic inflammation is independent of B-T cell interaction in a chronic house dust mite-driven asthma model. *Clin Exp Allergy*. 2017;47(4):551-64. doi:10.1111/cea.12834.
207. Ghosh S, Hoselton SA, Asbach SV, Steffan BN, Wanjara SB, Dorsam GP et al. B lymphocytes regulate airway granulocytic inflammation and cytokine production in a murine model of fungal allergic asthma. *Cell Mol Immunol*. 2015;12(2):202-12. doi:10.1038/cmi.2014.103.
208. Meyts I, Vanoirbeek JA, Hens G, Vanaudenaerde BM, Verbinnen B, Bullens DM et al. T-cell mediated late increase in bronchial tone after allergen provocation in a murine asthma model. *Clin Immunol*. 2008;128(2):248-58. doi:10.1016/j.clim.2008.03.515.
209. Braza F, Chesne J, Durand M, Dirou S, Brosseau C, Mahay G et al. A regulatory CD9(+) B-cell subset inhibits HDM-induced allergic airway inflammation. *Allergy*. 2015;70(11):1421-31. doi:10.1111/all.12697.
210. Halim TY, Krauss RH, Sun AC, Takei F. Lung natural helper cells are a critical source of Th2 cell-type cytokines in protease allergen-induced airway inflammation. *Immunity*. 2012;36(3):451-63. doi:10.1016/j.immuni.2011.12.020.
211. Gold MJ, Antignano F, Halim TY, Hirota JA, Blanchet MR, Zaph C et al. Group 2 innate lymphoid cells facilitate sensitization to local, but not systemic, TH2-inducing allergen exposures. *J Allergy Clin Immunol*. 2014;133(4):1142-8. doi:10.1016/j.jaci.2014.02.033.
212. Kim HY, Chang YJ, Subramanian S, Lee HH, Albacker LA, Matangkasombut P et al. Innate lymphoid cells responding to IL-33 mediate airway hyperreactivity independently of adaptive immunity. *J Allergy Clin Immunol*. 2012;129(1):216-27 e1-6. doi:10.1016/j.jaci.2011.10.036.
213. Christianson CA, Goplen NP, Zafar I, Irvin C, Good JT, Jr., Rollins DR et al. Persistence of asthma requires multiple feedback circuits involving type 2 innate lymphoid cells and IL-33. *J Allergy Clin Immunol*. 2015;136(1):59-68 e14. doi:10.1016/j.jaci.2014.11.037.

214. Chang YJ, Kim HY, Albacker LA, Baumgarth N, McKenzie AN, Smith DE et al. Innate lymphoid cells mediate influenza-induced airway hyper-reactivity independently of adaptive immunity. *Nat Immunol.* 2011;12(7):631-8. doi:10.1038/ni.2045.
215. Lambrecht BN, Hammad H. The immunology of asthma. *Nat Immunol.* 2015;16(1):45-56. doi:10.1038/ni.3049.
216. Takeda T, Unno H, Morita H, Futamura K, Emi-Sugie M, Arae K et al. Platelets constitutively express IL-33 protein and modulate eosinophilic airway inflammation. *J Allergy Clin Immunol.* 2016;138(5):1395-403 e6. doi:10.1016/j.jaci.2016.01.032.
217. Perkins C, Yanase N, Smulian G, Gildea L, Orekov T, Potter C et al. Selective stimulation of IL-4 receptor on smooth muscle induces airway hyperresponsiveness in mice. *J Exp Med.* 2011;208(4):853-67. doi:10.1084/jem.20100023.
218. Yang M, Kumar RK, Foster PS. Pathogenesis of steroid-resistant airway hyperresponsiveness: interaction between IFN-gamma and TLR4/MyD88 pathways. *J Immunol.* 2009;182(8):5107-15. doi:10.4049/jimmunol.0803468.
219. Alcorn JF, Rinaldi LM, Jaffe EF, van Loon M, Bates JH, Janssen-Heininger YM et al. Transforming growth factor-beta1 suppresses airway hyperresponsiveness in allergic airway disease. *Am J Respir Crit Care Med.* 2007;176(10):974-82. doi:10.1164/rccm.200702-334OC.
220. Spinelli AM, Liu Y, Sun LY, Gonzalez-Cobos JC, Backs J, Trebak M et al. Smooth muscle CaMKII δ promotes allergen-induced airway hyperresponsiveness and inflammation. *Pflugers Arch.* 2015;467(12):2541-54. doi:10.1007/s00424-015-1713-5.
221. Chesne J, Braza F, Chadeuf G, Mahay G, Cheminant MA, Loy J et al. Prime role of IL-17A in neutrophilia and airway smooth muscle contraction in a house dust mite-induced allergic asthma model. *J Allergy Clin Immunol.* 2015;135(6):1643- e3. doi:10.1016/j.jaci.2014.12.1872.
222. Daubeuf F, Frossard N. Acute Asthma Models to Ovalbumin in the Mouse. *Curr Protoc Mouse Biol.* 2013;3(1):31-7. doi:10.1002/9780470942390.mo120202.
223. Reber LL, Daubeuf F, Plantinga M, De Cauwer L, Gerlo S, Waelput W et al. A dissociated glucocorticoid receptor modulator reduces airway hyperresponsiveness and inflammation in a mouse model of asthma. *J Immunol.* 2012;188(7):3478-87. doi:10.4049/jimmunol.1004227.
224. Xisto DG, Farias LL, Ferreira HC, Picanco MR, Amitrano D, Lapa ESJR et al. Lung parenchyma remodeling in a murine model of chronic allergic inflammation. *Am J Respir Crit Care Med.* 2005;171(8):829-37. doi:10.1164/rccm.200408-997OC.
225. Hackett TL, Ferrante SC, Hoptay CE, Engelhardt JF, Ingram JL, Zhang Y et al. A Heterotopic Xenograft Model of Human Airways for Investigating Fibrosis in Asthma. *Am J Respir Cell Mol Biol.* 2017;56(3):291-9. doi:10.1165/rcmb.2016-0065MA.
226. Jaffer OA, Carter AB, Sanders PN, Dibbern ME, Winters CJ, Murthy S et al. Mitochondrial-targeted antioxidant therapy decreases transforming growth factor-beta-mediated collagen production in a murine asthma model. *Am J Respir Cell Mol Biol.* 2015;52(1):106-15. doi:10.1165/rcmb.2013-0519OC.
227. Hardy CL, Nguyen HA, Mohamud R, Yao J, Oh DY, Plebanski M et al. The activin A antagonist follistatin inhibits asthmatic airway remodelling. *Thorax.* 2013;68(1):9-18. doi:10.1136/thoraxjnl-2011-201128.
228. Wakahara K, Tanaka H, Takahashi G, Tamari M, Nasu R, Toyohara T et al. Repeated instillations of *Dermatophagoides farinae* into the airways can induce Th2-dependent airway hyperresponsiveness, eosinophilia and remodeling in mice: effect of intratracheal treatment of fluticasone propionate. *Eur J Pharmacol.* 2008;578(1):87-96. doi:10.1016/j.ejphar.2007.09.005.
229. Doyle TM, Ellis R, Park HJ, Inman MD, Sehmi R. Modulating progenitor accumulation attenuates lung angiogenesis in a mouse model of asthma. *Eur Respir J.* 2011;38(3):679-87. doi:10.1183/09031936.00133210.
230. Lee KS, Park SJ, Kim SR, Min KH, Lee KY, Choe YH et al. Inhibition of VEGF blocks TGF-beta1 production through a PI3K/Akt signalling pathway. *Eur Respir J.* 2008;31(3):523-31. doi:10.1183/09031936.00125007.

231. Johnson JR, Wiley RE, Fattouh R, Swirski FK, Gajewska BU, Coyle AJ et al. Continuous exposure to house dust mite elicits chronic airway inflammation and structural remodeling. *Am J Respir Crit Care Med*. 2004;169(3):378-85. doi:10.1164/rccm.200308-1094OC.
232. Saglani S, Lui S, Ullmann N, Campbell GA, Sherburn RT, Mathie SA et al. IL-33 promotes airway remodeling in pediatric patients with severe steroid-resistant asthma. *J Allergy Clin Immunol*. 2013;132(3):676-85 e13. doi:10.1016/j.jaci.2013.04.012.
233. Daubeuf F, Jung F, Douglas GJ, Chevalier E, Frossard N. Protective effect of a Protein Epitope Mimetic CCR10 antagonist, POL7085, in a model of allergic eosinophilic airway inflammation. *Respir Res*. 2015;16:77. doi:10.1186/s12931-015-0231-5.
234. Schuijs MJ, Willart MA, Vergote K, Gras D, Deswarte K, Ege MJ et al. Farm dust and endotoxin protect against allergy through A20 induction in lung epithelial cells. *Science*. 2015;349(6252):1106-10. doi:10.1126/science.aac6623.
235. Qiu S, Fan X, Yang Y, Dong P, Zhou W, Xu Y et al. *Schistosoma japonicum* infection downregulates house dust mite-induced allergic airway inflammation in mice. *PLoS One*. 2017;12(6):e0179565. doi:10.1371/journal.pone.0179565.
236. Sabatel C, Radermecker C, Fievez L, Paulissen G, Chakarov S, Fernandes C et al. Exposure to Bacterial CpG DNA Protects from Airway Allergic Inflammation by Expanding Regulatory Lung Interstitial Macrophages. *Immunity*. 2017;46(3):457-73. doi:10.1016/j.immuni.2017.02.016.
237. Shim JU, Lee SE, Hwang W, Lee C, Park JW, Sohn JH et al. Flagellin suppresses experimental asthma by generating regulatory dendritic cells and T cells. *J Allergy Clin Immunol*. 2016;137(2):426-35. doi:10.1016/j.jaci.2015.07.010.
238. Patel PS, Kearney JF. Neonatal exposure to pneumococcal phosphorylcholine modulates the development of house dust mite allergy during adult life. *J Immunol*. 2015;194(12):5838-50. doi:10.4049/jimmunol.1500251.
239. Finlay CM, Walsh KP, Mills KH. Induction of regulatory cells by helminth parasites: exploitation for the treatment of inflammatory diseases. *Immunol Rev*. 2014;259(1):206-30. doi:10.1111/imr.12164.
240. Chang YJ, Kim HY, Albacker LA, Lee HH, Baumgarth N, Akira S et al. Influenza infection in suckling mice expands an NKT cell subset that protects against airway hyperreactivity. *J Clin Invest*. 2011;121(1):57-69. doi:10.1172/JCI44845.
241. Arnold IC, Dehzad N, Reuter S, Martin H, Becher B, Taube C et al. *Helicobacter pylori* infection prevents allergic asthma in mouse models through the induction of regulatory T cells. *J Clin Invest*. 2011;121(8):3088-93. doi:10.1172/JCI45041.
242. Hagner S, Harb H, Zhao M, Stein K, Holst O, Ege MJ et al. Farm-derived Gram-positive bacterium *Staphylococcus sciuri* W620 prevents asthma phenotype in HDM- and OVA-exposed mice. *Allergy*. 2013;68(3):322-9. doi:10.1111/all.12094.
243. Fuchs B, Knothe S, Rochlitz S, Nassimi M, Greweling M, Lauenstein HD et al. A Toll-like receptor 2/6 agonist reduces allergic airway inflammation in chronic respiratory sensitisation to Timothy grass pollen antigens. *Int Arch Allergy Immunol*. 2010;152(2):131-9. doi:10.1159/000265534.
244. Peters M, Kauth M, Schwarze J, Korner-Rettberg C, Riedler J, Nowak D et al. Inhalation of stable dust extract prevents allergen induced airway inflammation and hyperresponsiveness. *Thorax*. 2006;61(2):134-9. doi:10.1136/thx.2005.049403.
245. Preston JA, Thorburn AN, Starkey MR, Beckett EL, Horvat JC, Wade MA et al. *Streptococcus pneumoniae* infection suppresses allergic airways disease by inducing regulatory T-cells. *Eur Respir J*. 2011;37(1):53-64. doi:10.1183/09031936.00049510.
246. Vital M, Harkema JR, Rizzo M, Tiedje J, Brandenberger C. Alterations of the Murine Gut Microbiome with Age and Allergic Airway Disease. *J Immunol Res*. 2015;2015:892568. doi:10.1155/2015/892568.
247. Calixto MC, Lintomen L, Schenka A, Saad MJ, Zanesco A, Antunes E. Obesity enhances eosinophilic inflammation in a murine model of allergic asthma. *Br J Pharmacol*. 2010;159(3):617-25. doi:10.1111/j.1476-5381.2009.00560.x.

248. Takeda M, Tanabe M, Ito W, Ueki S, Konno Y, Chihara M et al. Gender difference in allergic airway remodelling and immunoglobulin production in mouse model of asthma. *Respirology*. 2013;18(5):797-806. doi:10.1111/resp.12078.
249. Birmingham JM, Gillespie VL, Srivastava K, Li XM, Busse PJ. Influenza A infection enhances antigen-induced airway inflammation and hyperresponsiveness in young but not aged mice. *Clin Exp Allergy*. 2014;44(9):1188-99. doi:10.1111/cea.12365.
250. Park HJ, Lee JH, Park YH, Han H, Sim da W, Park KH et al. Roflumilast Ameliorates Airway Hyperresponsiveness Caused by Diet-Induced Obesity in a Murine Model. *Am J Respir Cell Mol Biol*. 2016;55(1):82-91. doi:10.1165/rcmb.2015-0345OC.
251. Brandenberger C, Li N, Jackson-Humbles DN, Rockwell CE, Wagner JG, Harkema JR. Enhanced allergic airway disease in old mice is associated with a Th17 response. *Clin Exp Allergy*. 2014;44(10):1282-92. doi:10.1111/cea.12388.
252. Everaere L, Ait-Yahia S, Molendi-Coste O, Vorng H, Quemener S, LeVu P et al. Innate lymphoid cells contribute to allergic airway disease exacerbation by obesity. *J Allergy Clin Immunol*. 2016;138(5):1309-18 e11. doi:10.1016/j.jaci.2016.03.019.
253. Herbst T, Sichelstiel A, Schar C, Yadava K, Burki K, Cahenzli J et al. Dysregulation of allergic airway inflammation in the absence of microbial colonization. *Am J Respir Crit Care Med*. 2011;184(2):198-205. doi:10.1164/rccm.201010-1574OC.
254. Melgert BN, Oriss TB, Qi Z, Dixon-McCarthy B, Geerlings M, Hylkema MN et al. Macrophages: regulators of sex differences in asthma? *Am J Respir Cell Mol Biol*. 2010;42(5):595-603. doi:10.1165/rcmb.2009-0016OC.
255. Verheijden KA, Willemsen LE, Braber S, Leusink-Muis T, Delsing DJ, Garssen J et al. Dietary galacto-oligosaccharides prevent airway eosinophilia and hyperresponsiveness in a murine house dust mite-induced asthma model. *Respir Res*. 2015;16:17. doi:10.1186/s12931-015-0171-0.
256. Castanhinha S, Sherburn R, Walker S, Gupta A, Bossley CJ, Buckley J et al. Pediatric severe asthma with fungal sensitization is mediated by steroid-resistant IL-33. *J Allergy Clin Immunol*. 2015;136(2):312-22 e7. doi:10.1016/j.jaci.2015.01.016.
257. Van Hove CL, Maes T, Cataldo DD, Gueders MM, Palmans E, Joos GF et al. Comparison of acute inflammatory and chronic structural asthma-like responses between C57BL/6 and BALB/c mice. *Int Arch Allergy Immunol*. 2009;149(3):195-207.
258. Gueders MM, Paulissen G, Crahay C, Quesada-Calvo F, Hacha J, Van Hove C et al. Mouse models of asthma: a comparison between C57BL/6 and BALB/c strains regarding bronchial responsiveness, inflammation, and cytokine production. *Inflamm Res*. 2009;58(12):845-54. doi:10.1007/s00011-009-0054-2.
259. Blacquiere MJ, Timens W, Melgert BN, Geerlings M, Postma DS, Hylkema MN. Maternal smoking during pregnancy induces airway remodelling in mice offspring. *Eur Respir J*. 2009;33(5):1133-40. doi:10.1183/09031936.00129608.
260. Dinger K, Kasper P, Hucklenbruch-Rother E, Vohlen C, Jobst E, Janoschek R et al. Early-onset obesity dysregulates pulmonary adipocytokine/insulin signaling and induces asthma-like disease in mice. *Sci Rep*. 2016;6:24168. doi:10.1038/srep24168.
261. Daubeuf F, Hachet-Haas M, Gizzi P, Gasparik V, Bonnet D, Utard V et al. An antedrug of the CXCL12 neutraligand blocks experimental allergic asthma without systemic effect in mice. *J Biol Chem*. 2013;288(17):11865-76. doi:10.1074/jbc.M112.449348.
262. Fattouh R, Midence NG, Arias K, Johnson JR, Walker TD, Goncharova S et al. Transforming growth factor-beta regulates house dust mite-induced allergic airway inflammation but not airway remodeling. *Am J Respir Crit Care Med*. 2008;177(6):593-603. doi:10.1164/rccm.200706-958OC.
263. Bouchaud G, Braza F, Chesne J, Lair D, Chen KW, Rolland-Debord C et al. Prevention of allergic asthma through Der p 2 peptide vaccination. *J Allergy Clin Immunol*. 2015;136(1):197-200 e1. doi:10.1016/j.jaci.2014.12.1938.
264. Castan L, Magnan A, Bouchaud G. Chemokine receptors in allergic diseases. *Allergy*. 2017;72(5):682-90. doi:10.1111/all.13089.

265. Maes T, Joos GF, Brusselle GG. Targeting interleukin-4 in asthma: lost in translation? *Am J Respir Cell Mol Biol.* 2012;47(3):261-70. doi:10.1165/rcmb.2012-0080TR.
266. Paulissen G, Rocks N, Gueders MM, Bedoret D, Crahay C, Quesada-Calvo F et al. ADAM-8, a metalloproteinase, drives acute allergen-induced airway inflammation. *Eur J Immunol.* 2011;41(2):380-91. doi:10.1002/eji.200940286.
267. Moriwaki A, Inoue H, Nakano T, Matsunaga Y, Matsuno Y, Matsumoto T et al. T cell treatment with small interfering RNA for suppressor of cytokine signaling 3 modulates allergic airway responses in a murine model of asthma. *Am J Respir Cell Mol Biol.* 2011;44(4):448-55. doi:10.1165/rcmb.2009-0051OC.
268. Durk T, Duerschmied D, Muller T, Grimm M, Reuter S, Vieira RP et al. Production of serotonin by tryptophan hydroxylase 1 and release via platelets contribute to allergic airway inflammation. *Am J Respir Crit Care Med.* 2013;187(5):476-85. doi:10.1164/rccm.201208-1440OC.
269. Muller T, Vieira RP, Grimm M, Durk T, Cicko S, Zeiser R et al. A potential role for P2X7R in allergic airway inflammation in mice and humans. *Am J Respir Cell Mol Biol.* 2011;44(4):456-64. doi:10.1165/rcmb.2010-0129OC.
270. McGee HS, Edwan JH, Agrawal DK. Flt3-L increases CD4+CD25+Foxp3+ICOS+ cells in the lungs of cockroach-sensitized and -challenged mice. *Am J Respir Cell Mol Biol.* 2010;42(3):331-40. doi:10.1165/rcmb.2008-0397OC.
271. Al-Shami A, Spolski R, Kelly J, Keane-Myers A, Leonard WJ. A role for TSLP in the development of inflammation in an asthma model. *J Exp Med.* 2005;202(6):829-39. doi:10.1084/jem.20050199.
272. Honda K, Arima M, Cheng G, Taki S, Hirata H, Eda F et al. Prostaglandin D2 reinforces Th2 type inflammatory responses of airways to low-dose antigen through bronchial expression of macrophage-derived chemokine. *J Exp Med.* 2003;198(4):533-43. doi:10.1084/jem.20022218.
273. Maes T, Tournoy KG, Joos GF. Gene therapy for allergic airway diseases. *Curr Allergy Asthma Rep.* 2011;11(2):163-72. doi:10.1007/s11882-011-0177-8.
274. Maher SA, Birrell MA, Adcock JJ, Wortley MA, Dubuis ED, Bonvini SJ et al. Prostaglandin D2 and the role of the DP1, DP2 and TP receptors in the control of airway reflex events. *Eur Respir J.* 2015;45(4):1108-18. doi:10.1183/09031936.00061614.
275. Chevalier E, Lagente V, Dupont M, Fargeau H, Palazzi X, Richard V et al. Lack of involvement of type 7 phosphodiesterase in an experimental model of asthma. *Eur Respir J.* 2012;39(3):582-8. doi:10.1183/09031936.00102610.
276. Park SJ, Lee KS, Kim SR, Min KH, Moon H, Lee MH et al. Phosphoinositide 3-kinase delta inhibitor suppresses interleukin-17 expression in a murine asthma model. *Eur Respir J.* 2010;36(6):1448-59. doi:10.1183/09031936.00106609.
277. Suzaki Y, Hamada K, Nomi T, Ito T, Sho M, Kai Y et al. A small-molecule compound targeting CCR5 and CXCR3 prevents airway hyperresponsiveness and inflammation. *Eur Respir J.* 2008;31(4):783-9. doi:10.1183/09031936.00111507.
278. Reber LL, Daubeuf F, Nemska S, Frossard N. The AGC kinase inhibitor H89 attenuates airway inflammation in mouse models of asthma. *PLoS One.* 2012;7(11):e49512. doi:10.1371/journal.pone.0049512.
279. Nials AT, Uddin S. Mouse models of allergic asthma: acute and chronic allergen challenge. *Dis Model Mech.* 2008;1(4-5):213-20. doi:10.1242/dmm.000323.
280. Eddens T, Campfield BT, Serody K, Manni ML, Horne W, Elsegeiny W et al. A Novel CD4+ T Cell-Dependent Murine Model of Pneumocystis-driven Asthma-like Pathology. *Am J Respir Crit Care Med.* 2016;194(7):807-20. doi:10.1164/rccm.201511-2205OC.
281. Goplen N, Karim MZ, Liang Q, Gorska MM, Rozario S, Guo L et al. Combined sensitization of mice to extracts of dust mite, ragweed, and *Aspergillus* species breaks through tolerance and establishes chronic features of asthma. *J Allergy Clin Immunol.* 2009;123(4):925-32 e11. doi:10.1016/j.jaci.2009.02.009.
282. Grundstrom J, Saarne T, Kemi C, Gregory JA, Waden K, Pils MC et al. Development of a mouse model for chronic cat allergen-induced asthma. *Int Arch Allergy Immunol.* 2014;165(3):195-205. doi:10.1159/000369066.

283. Lloyd CM. Building better mouse models of asthma. *Curr Allergy Asthma Rep.* 2007;7(3):231-6.
284. Haworth O, Cernadas M, Levy BD. NK cells are effectors for resolvin E1 in the timely resolution of allergic airway inflammation. *J Immunol.* 2011;186(11):6129-35. doi:10.4049/jimmunol.1004007.
285. Boudousquie C, Pellaton C, Barbier N, Spertini F. CD4+CD25+ T cell depletion impairs tolerance induction in a murine model of asthma. *Clin Exp Allergy.* 2009;39(9):1415-26. doi:10.1111/j.1365-2222.2009.03314.x.
286. Lu M, Dawicki W, Zhang X, Huang H, Nayyar A, Gordon JR. Therapeutic induction of tolerance by IL-10-differentiated dendritic cells in a mouse model of house dust mite-asthma. *Allergy.* 2011;66(5):612-20. doi:10.1111/j.1398-9995.2010.02526.x.
287. Van Hove CL, Maes T, Joos GF, Tournoy KG. Prolonged Inhaled Allergen Exposure Can Induce Persistent Tolerance. *Am J Respir Cell Mol Biol.* 2007;36(5):573-84.
288. Ravanetti L, Dijkhuis A, Sabogal Pineros YS, Bal SM, Dierdorp BS, Dekker T et al. An early innate response underlies severe influenza-induced exacerbations of asthma in a novel steroid-insensitive and anti-IL-5-responsive mouse model. *Allergy.* 2017;72(5):737-53. doi:10.1111/all.13057.
289. Mahmutovic Persson I, Akbarshahi H, Menzel M, Brandelius A, Uller L. Increased expression of upstream TH2-cytokines in a mouse model of viral-induced asthma exacerbation. *J Transl Med.* 2016;14:52. doi:10.1186/s12967-016-0808-x.
290. Hobbs BD, de Jong K, Lamontagne M, Bossé Y, Shrine N, Artigas MS et al. Genetic loci associated with chronic obstructive pulmonary disease overlap with loci for lung function and pulmonary fibrosis. *Nature genetics.* 2017;49(3):426-32. doi:10.1038/ng.3752.
291. Starkhammar M, Larsson O, Kumlien Georen S, Leino M, Dahlen SE, Adner M et al. Toll-like receptor ligands LPS and poly (I:C) exacerbate airway hyperresponsiveness in a model of airway allergy in mice, independently of inflammation. *PLoS One.* 2014;9(8):e104114. doi:10.1371/journal.pone.0104114.
292. Dahl ME, Dabbagh K, Liggitt D, Kim S, Lewis DB. Viral-induced T helper type 1 responses enhance allergic disease by effects on lung dendritic cells. *Nat Immunol.* 2004;5(3):337-43. doi:10.1038/ni1041.
293. Nguyen TH, Maltby S, Simpson JL, Eysers F, Baines KJ, Gibson PG et al. TNF-alpha and Macrophages Are Critical for Respiratory Syncytial Virus-Induced Exacerbations in a Mouse Model of Allergic Airways Disease. *J Immunol.* 2016;196(9):3547-58. doi:10.4049/jimmunol.1502339.
294. Reuter S, Dehzad N, Martin H, Bohm L, Becker M, Buhl R et al. TLR3 but not TLR7/8 ligand induces allergic sensitization to inhaled allergen. *J Immunol.* 2012;188(10):5123-31. doi:10.4049/jimmunol.1101618.
295. Bartlett NW, Walton RP, Edwards MR, Aniscenko J, Caramori G, Zhu J et al. Mouse models of rhinovirus-induced disease and exacerbation of allergic airway inflammation. *Nature medicine.* 2008;14(2):199-204. doi:10.1038/nm1713.
296. Sjoberg LC, Nilsson AZ, Lei Y, Gregory JA, Adner M, Nilsson GP. Interleukin 33 exacerbates antigen driven airway hyperresponsiveness, inflammation and remodeling in a mouse model of asthma. *Sci Rep.* 2017;7(1):4219. doi:10.1038/s41598-017-03674-0.
297. De Alba J, Ota R, Calama E, Domenech A, Prats N, Gozzard N et al. Double-stranded RNA evokes exacerbation in a mouse model of corticosteroid refractory asthma. *Clin Sci (Lond).* 2015;129(11):973-87. doi:10.1042/CS20150292.
298. Ramadan A, Pham Van L, Machavoine F, Dietrich C, Alkan M, Karasuyama H et al. Activation of basophils by the double-stranded RNA poly(A:U) exacerbates allergic inflammation. *Allergy.* 2013;68(6):732-8. doi:10.1111/all.12151.
299. Clarke DL, Davis NH, Majithiya JB, Piper SC, Lewis A, Sleeman MA et al. Development of a mouse model mimicking key aspects of a viral asthma exacerbation. *Clin Sci (Lond).* 2014;126(8):567-80. doi:10.1042/CS20130149.
300. Schutze N, Lehmann I, Bonisch U, Simon JC, Polte T. Exposure to mycotoxins increases the allergic immune response in a murine asthma model. *Am J Respir Crit Care Med.* 2010;181(11):1188-99. doi:10.1164/rccm.200909-1350OC.

301. Kumar RK, Herbert C, Foster PS. Mouse models of acute exacerbations of allergic asthma. *Respirology*. 2016;21(5):842-9. doi:10.1111/resp.12760.
302. Murakami D, Yamada H, Yajima T, Masuda A, Komune S, Yoshikai Y. Lipopolysaccharide inhalation exacerbates allergic airway inflammation by activating mast cells and promoting Th2 responses. *Clin Exp Allergy*. 2007;37(3):339-47. doi:10.1111/j.1365-2222.2006.02633.x.
303. Lanckacker EA, Tournoy KG, Hammad H, Holtappels G, Lambrecht BN, Joos GF et al. Short cigarette smoke exposure facilitates sensitisation and asthma development in mice. *Eur Respir J*. 2013;41(5):1189-99.
304. Moerloose KB, Pauwels RA, Joos GF. Short-term cigarette smoke exposure enhances allergic airway inflammation in mice. *Am J Respir Crit Care Med*. 2005;172(2):168-72.
305. De Grove KC, Provoost S, Hendriks RW, McKenzie AN, Seys LJ, Kumar S et al. Dysregulation of type 2 innate lymphoid cells and TH2 cells impairs pollutant-induced allergic airway responses. *J Allergy Clin Immunol*. 2017;139(1):246-57 e4. doi:10.1016/j.jaci.2016.03.044.
306. Brandt EB, Khurana Hershey GK. A combination of dexamethasone and anti-IL-17A treatment can alleviate diesel exhaust particle-induced steroid insensitive asthma. *J Allergy Clin Immunol*. 2016;138(3):924-8 e2. doi:10.1016/j.jaci.2016.03.037.
307. Brandt EB, Biagini Myers JM, Acciani TH, Ryan PH, Sivaprasad U, Ruff B et al. Exposure to allergen and diesel exhaust particles potentiates secondary allergen-specific memory responses, promoting asthma susceptibility. *J Allergy Clin Immunol*. 2015;136(2):295-303 e7. doi:10.1016/j.jaci.2014.11.043.
308. Martin RA, Ather JL, Lundblad LK, Suratt BT, Boyson JE, Budd RC et al. Interleukin-1 receptor and caspase-1 are required for the Th17 response in nitrogen dioxide-promoted allergic airway disease. *Am J Respir Cell Mol Biol*. 2013;48(5):655-64. doi:10.1165/rcmb.2012-0423OC.
309. Min MG, Song DJ, Miller M, Cho JY, McElwain S, Ferguson P et al. Coexposure to environmental tobacco smoke increases levels of allergen-induced airway remodeling in mice. *J Immunol*. 2007;178(8):5321-8.
310. Liang L, Li F, Bao A, Zhang M, Chung KF, Zhou X. Activation of p38 mitogen-activated protein kinase in ovalbumin and ozone-induced mouse model of asthma. *Respirology*. 2013;18 Suppl 3:20-9. doi:10.1111/resp.12189.
311. Farraj AK, Boykin E, Ledbetter A, Andrews D, Gavett SH. Increased lung resistance after diesel particulate and ozone co-exposure not associated with enhanced lung inflammation in allergic mice. *Inhal Toxicol*. 2010;22(1):33-41. doi:10.3109/08958370902862434.
312. Inoue K, Koike E, Takano H, Yanagisawa R, Ichinose T, Yoshikawa T. Effects of diesel exhaust particles on antigen-presenting cells and antigen-specific Th immunity in mice. *Exp Biol Med (Maywood)*. 2009;234(2):200-9.
313. Hadebe S, Kirstein F, Fierens K, Chen K, Drummond RA, Vautier S et al. Microbial Ligand Costimulation Drives Neutrophilic Steroid-Refractory Asthma. *PLoS One*. 2015;10(8):e0134219. doi:10.1371/journal.pone.0134219.
314. Duechs MJ, Tilp C, Tomsic C, Gantner F, Erb KJ. Development of a novel severe triple allergen asthma model in mice which is resistant to dexamethasone and partially resistant to TLR7 and TLR9 agonist treatment. *PLoS One*. 2014;9(3):e91223. doi:10.1371/journal.pone.0091223.
315. McGovern TK, Goldberger M, Allard B, Farahnak S, Hamamoto Y, O'Sullivan M et al. Neutrophils mediate airway hyperresponsiveness after chlorine-induced airway injury in the mouse. *Am J Respir Cell Mol Biol*. 2015;52(4):513-22. doi:10.1165/rcmb.2013-0430OC.
316. Mizutani N, Nabe T, Yoshino S. IL-17A promotes the exacerbation of IL-33-induced airway hyperresponsiveness by enhancing neutrophilic inflammation via CXCR2 signaling in mice. *J Immunol*. 2014;192(4):1372-84. doi:10.4049/jimmunol.1301538.
317. De Vooght V, Smulders S, Haenen S, Belmans J, Opendakker G, Verbeken E et al. Neutrophil and eosinophil granulocytes as key players in a mouse model of chemical-induced asthma. *Toxicol Sci*. 2013;131(2):406-18. doi:10.1093/toxsci/kfs308.

318. Devos FC, Boonen B, Alpizar YA, Maes T, Hox V, Seys S et al. Neuro-immune interactions in chemical-induced airway hyperreactivity. *Eur Respir J*. 2016;48(2):380-92. doi:10.1183/13993003.01778-2015.
319. Matheson JM, Johnson VJ, Luster MI. Immune mediators in a murine model for occupational asthma: studies with toluene diisocyanate. *Toxicol Sci*. 2005;84(1):99-109. doi:10.1093/toxsci/kfi051.
320. Manni ML, Mandalapu S, McHugh KJ, Elloso MM, Dudas PL, Alcorn JF. Molecular Mechanisms of Airway Hyperresponsiveness in a Murine Model of Steroid-Resistant Airway Inflammation. *J Immunol*. 2016;196(3):963-77. doi:10.4049/jimmunol.1501531.
321. Kraneveld AD, van der Kleij HP, Kool M, van Houwelingen AH, Weitenberg AC, Redegeld FA et al. Key role for mast cells in nonatopic asthma. *J Immunol*. 2002;169(4):2044-53.
322. Bonvini SJ, Birrell MA, Grace MS, Maher SA, Adcock JJ, Wortley MA et al. Transient receptor potential cation channel, subfamily V, member 4 and airway sensory afferent activation: Role of adenosine triphosphate. *The Journal of allergy and clinical immunology*. 2016;138(1):249-61 e12. doi:10.1016/j.jaci.2015.10.044.
323. Kistemaker LE, Bos IS, Menzen MH, Maarsingh H, Meurs H, Gosens R. Combination therapy of tiotropium and ciclesonide attenuates airway inflammation and remodeling in a guinea pig model of chronic asthma. *Respir Res*. 2016;17:13. doi:10.1186/s12931-016-0327-6.
324. Yu L, Liu Q, Canning BJ. Evidence for autocrine and paracrine regulation of allergen-induced mast cell mediator release in the guinea pig airways. *Eur J Pharmacol*. 2017. doi:10.1016/j.ejphar.2017.11.017.
325. Kloek J, Mortaz E, van Ark I, Lilly CM, Nijkamp FP, Folkerts G. Glutathione prevents the early asthmatic reaction and airway hyperresponsiveness in guinea pigs. *J Physiol Pharmacol*. 2010;61(1):67-72.
326. Aun MV, Bonamichi-Santos R, Arantes-Costa FM, Kalil J, Giavina-Bianchi P. Animal models of asthma: utility and limitations. *J Asthma Allergy*. 2017;10:293-301. doi:10.2147/JAA.S121092.
327. Laurell CB, Eriksson S. The electrophoretic alpha1-globulin pattern of serum in alpha1-antitrypsin deficiency. 1963. *Copd*. 2013;10 Suppl 1:3-8. doi:10.3109/15412555.2013.771956.
328. Paterson T, Moore S. The expression and characterization of five recombinant murine alpha 1-protease inhibitor proteins. *Biochemical and biophysical research communications*. 1996;219(1):64-9. doi:10.1006/bbrc.1996.0182.
329. Borriello F, Krauter KS. Multiple murine alpha 1-protease inhibitor genes show unusual evolutionary divergence. *Proceedings of the National Academy of Sciences of the United States of America*. 1991;88(21):9417-21.
330. Alam S, Li Z, Atkinson C, Jonigk D, Janciauskiene S, Mahadeva R. Z alpha1-antitrypsin confers a proinflammatory phenotype that contributes to chronic obstructive pulmonary disease. *American journal of respiratory and critical care medicine*. 2014;189(8):909-31. doi:10.1164/rccm.201308-1458OC.
331. Lao T, Jiang Z, Yun J, Qiu W, Guo F, Huang C et al. Hhip haploinsufficiency sensitizes mice to age-related emphysema. *Proceedings of the National Academy of Sciences of the United States of America*. 2016;113(32):E4681-7. doi:10.1073/pnas.1602342113.
332. Zhou X, Qiu W, Sathirapongsasuti JF, Cho MH, Mancini JD, Lao T et al. Gene expression analysis uncovers novel hedgehog interacting protein (HHIP) effects in human bronchial epithelial cells. *Genomics*. 2013;101(5):263-72. doi:10.1016/j.ygeno.2013.02.010.
333. Jiang Z, Lao T, Qiu W, Polverino F, Gupta K, Guo F et al. A Chronic Obstructive Pulmonary Disease Susceptibility Gene, FAM13A, Regulates Protein Stability of beta-Catenin. *American journal of respiratory and critical care medicine*. 2016;194(2):185-97. doi:10.1164/rccm.201505-0999OC.
334. Zhou X, Baron RM, Hardin M, Cho MH, Zielinski J, Hawrylkiewicz I et al. Identification of a chronic obstructive pulmonary disease genetic determinant that regulates HHIP. *Human molecular genetics*. 2012;21(6):1325-35. doi:10.1093/hmg/ddr569.
335. Bonniaud P, Kolb M, Galt T, Robertson J, Robbins C, Stampfli M et al. Smad3 null mice develop airspace enlargement and are resistant to TGF-beta-mediated pulmonary fibrosis. *Journal of immunology*. 2004;173(3):2099-108.

336. Chen H, Sun J, Buckley S, Chen C, Warburton D, Wang XF et al. Abnormal mouse lung alveolarization caused by Smad3 deficiency is a developmental antecedent of centrilobular emphysema. *American journal of physiology Lung cellular and molecular physiology*. 2005;288(4):L683-91. doi:10.1152/ajplung.00298.2004.
337. Chuang PT, Kawcak T, McMahon AP. Feedback control of mammalian Hedgehog signaling by the Hedgehog-binding protein, Hip1, modulates Fgf signaling during branching morphogenesis of the lung. *Genes & development*. 2003;17(3):342-7. doi:10.1101/gad.1026303.
338. Hokuto I, Perl AK, Whitsett JA. Prenatal, but not postnatal, inhibition of fibroblast growth factor receptor signaling causes emphysema. *The Journal of biological chemistry*. 2003;278(1):415-21. doi:10.1074/jbc.M208328200.
339. Jakkula M, Le Cras TD, Gebb S, Hirth KP, Tudor RM, Voelkel NF et al. Inhibition of angiogenesis decreases alveolarization in the developing rat lung. *American journal of physiology Lung cellular and molecular physiology*. 2000;279(3):L600-7.
340. Jin N, Cho SN, Raso MG, Wistuba I, Smith Y, Yang Y et al. Mig-6 is required for appropriate lung development and to ensure normal adult lung homeostasis. *Development*. 2009;136(19):3347-56. doi:10.1242/dev.032979.
341. Tang K, Rossiter HB, Wagner PD, Breen EC. Lung-targeted VEGF inactivation leads to an emphysema phenotype in mice. *Journal of applied physiology*. 2004;97(4):1559-66; discussion 49. doi:10.1152/jappphysiol.00221.2004.
342. Maki JM, Sormunen R, Lippo S, Kaarteenaho-Wiik R, Soininen R, Myllyharju J. Lysyl oxidase is essential for normal development and function of the respiratory system and for the integrity of elastic and collagen fibers in various tissues. *The American journal of pathology*. 2005;167(4):927-36. doi:10.1016/S0002-9440(10)61183-2.
343. Neptune ER, Frischmeyer PA, Arking DE, Myers L, Bunton TE, Gayraud B et al. Dysregulation of TGF-beta activation contributes to pathogenesis in Marfan syndrome. *Nature genetics*. 2003;33(3):407-11. doi:10.1038/ng1116.
344. Drummond D, Baravalle-Einaudi M, Lezmi G, Vibhushan S, Franco-Montoya ML, Hadchouel A et al. Combined Effects of in Utero and Adolescent Tobacco Smoke Exposure on Lung Function in C57Bl/6J Mice. *Environmental health perspectives*. 2017;125(3):392-9. doi:10.1289/EHP54.
345. Xiao R, Perveen Z, Paulsen D, Rouse R, Ambalavanan N, Kearney M et al. In utero exposure to second-hand smoke aggravates adult responses to irritants: adult second-hand smoke. *American journal of respiratory cell and molecular biology*. 2012;47(6):843-51. doi:10.1165/rcmb.2012-0241OC.
346. Meyer KF, Krauss-Etschmann S, Kooistra W, Reinders-Luinge M, Timens W, Kobzik L et al. Prenatal exposure to tobacco smoke sex dependently influences methylation and mRNA levels of the Igf axis in lungs of mouse offspring. *American journal of physiology Lung cellular and molecular physiology*. 2017;312(4):L542-L55. doi:10.1152/ajplung.00271.2016.
347. Singh SP, Gundavarapu S, Smith KR, Chand HS, Saeed AI, Mishra NC et al. Gestational exposure of mice to secondhand cigarette smoke causes bronchopulmonary dysplasia blocked by the nicotinic receptor antagonist mecamylamine. *Environmental health perspectives*. 2013;121(8):957-64. doi:10.1289/ehp.1306611.
348. John-Schuster G, Gunter S, Hager K, Conlon TM, Eickelberg O, Yildirim AO. Inflammation increases susceptibility to cigarette smoke-induced COPD. *Oncotarget*. 2016;7(21):30068-83. doi:10.18632/oncotarget.4027.
349. Zhou S, Wright JL, Liu J, Sin DD, Churg A. Aging does not enhance experimental cigarette smoke-induced COPD in the mouse. *PloS one*. 2013;8(8):e71410. doi:10.1371/journal.pone.0071410.
350. Mercer PF, Abbott-Banner K, Adcock IM, Knowles RG. Translational models of lung disease. *Clinical science*. 2015;128(4):235-56. doi:10.1042/CS20140373.
351. Jones B, Donovan C, Liu G, Gomez HM, Chimankar V, Harrison CL et al. Animal models of COPD: What do they tell us? *Respirology*. 2017;22(1):21-32. doi:10.1111/resp.12908.

352. Ni K, Serban KA, Batra C, Petrache I. Alpha-1 Antitrypsin Investigations Using Animal Models of Emphysema. *Annals of the American Thoracic Society*. 2016;13 Suppl 4:S311-6. doi:10.1513/AnnalsATS.201510-675KV.
353. Stevenson CS, Birrell MA. Moving towards a new generation of animal models for asthma and COPD with improved clinical relevance. *Pharmacology & therapeutics*. 2011;130(2):93-105. doi:10.1016/j.pharmthera.2010.10.008.
354. Nikota JK, Stampfli MR. Cigarette smoke-induced inflammation and respiratory host defense: Insights from animal models. *Pulmonary pharmacology & therapeutics*. 2012;25(4):257-62. doi:10.1016/j.pupt.2012.05.005.
355. Dvorkin-Gheva A, Vanderstocken G, Yildirim AO, Brandsma CA, Obeidat M, Bosse Y et al. Total particulate matter concentration skews cigarette smoke's gene expression profile. *ERJ open research*. 2016;2(4). doi:10.1183/23120541.00029-2016.
356. Hodge-Bell KC, Lee KM, Renne RA, Gideon KM, Harbo SJ, McKinney WJ. Pulmonary inflammation in mice exposed to mainstream cigarette smoke. *Inhalation toxicology*. 2007;19(4):361-76. doi:10.1080/08958370601144076.
357. Tam A, Churg A, Wright JL, Zhou S, Kirby M, Coxson HO et al. Sex Differences in Airway Remodeling in a Mouse Model of Chronic Obstructive Pulmonary Disease. *American journal of respiratory and critical care medicine*. 2016;193(8):825-34. doi:10.1164/rccm.201503-0487OC.
358. Tam A, Bates JH, Churg A, Wright JL, Man SF, Sin DD. Sex-Related Differences in Pulmonary Function following 6 Months of Cigarette Exposure: Implications for Sexual Dimorphism in Mild COPD. *PloS one*. 2016;11(10):e0164835. doi:10.1371/journal.pone.0164835.
359. Thorne D, Adamson J. A review of in vitro cigarette smoke exposure systems. *Experimental and toxicologic pathology : official journal of the Gesellschaft fur Toxikologische Pathologie*. 2013;65(7-8):1183-93. doi:10.1016/j.etp.2013.06.001.
360. Hu G, Zhou Y, Hong W, Tian J, Hu J, Peng G et al. Development and systematic oxidative stress of a rat model of chronic bronchitis and emphysema induced by biomass smoke. *Experimental lung research*. 2013;39(6):229-40. doi:10.3109/01902148.2013.797521.
361. Sussan TE, Ingole V, Kim JH, McCormick S, Negherbon J, Fallica J et al. Source of biomass cooking fuel determines pulmonary response to household air pollution. *American journal of respiratory cell and molecular biology*. 2014;50(3):538-48. doi:10.1165/rcmb.2013-0201OC.
362. Triantaphyllopoulos K, Hussain F, Pinart M, Zhang M, Li F, Adcock I et al. A model of chronic inflammation and pulmonary emphysema after multiple ozone exposures in mice. *American journal of physiology Lung cellular and molecular physiology*. 2011;300(5):L691-700. doi:10.1152/ajplung.00252.2010.
363. Wiegman CH, Li F, Clarke CJ, Jazrawi E, Kirkham P, Barnes PJ et al. A comprehensive analysis of oxidative stress in the ozone-induced lung inflammation mouse model. *Clinical science*. 2014;126(6):425-40. doi:10.1042/CS20130039.
364. March TH, Barr EB, Finch GL, Nikula KJ, Seagrave JC. Effects of concurrent ozone exposure on the pathogenesis of cigarette smoke-induced emphysema in B6C3F1 mice. *Inhalation toxicology*. 2002;14(12):1187-213. doi:10.1080/08958370290084818.
365. Vernooij JH, Dentener MA, van Suylen RJ, Buurman WA, Wouters EF. Long-term intratracheal lipopolysaccharide exposure in mice results in chronic lung inflammation and persistent pathology. *American journal of respiratory cell and molecular biology*. 2002;26(1):152-9. doi:10.1165/ajrcmb.26.1.4652.
366. Stolk J, Rudolphus A, Davies P, Osinga D, Dijkman JH, Agarwal L et al. Induction of emphysema and bronchial mucus cell hyperplasia by intratracheal instillation of lipopolysaccharide in the hamster. *The Journal of pathology*. 1992;167(3):349-56. doi:10.1002/path.1711670314.
367. Savov JD, Brass DM, Berman KG, McElvania E, Schwartz DA. Fibrinolysis in LPS-induced chronic airway disease. *American journal of physiology Lung cellular and molecular physiology*. 2003;285(4):L940-8. doi:10.1152/ajplung.00102.2003.

368. Brass DM, Hollingsworth JW, Cinque M, Li Z, Potts E, Toloza E et al. Chronic LPS inhalation causes emphysema-like changes in mouse lung that are associated with apoptosis. *American journal of respiratory cell and molecular biology*. 2008;39(5):584-90. doi:10.1165/rcmb.2007-0448OC.
369. Birrell MA, McCluskie K, Wong S, Donnelly LE, Barnes PJ, Belvisi MG. Resveratrol, an extract of red wine, inhibits lipopolysaccharide induced airway neutrophilia and inflammatory mediators through an NF-kappaB-independent mechanism. *FASEB journal : official publication of the Federation of American Societies for Experimental Biology*. 2005;19(7):840-1. doi:10.1096/fj.04-2691fje.
370. Onnervik PO, Lindahl M, Svitacheva N, Stampfli M, Thim K, Smailagic A et al. The role of the CCR1 receptor in the inflammatory response to tobacco smoke in a mouse model. *Inflammation research : official journal of the European Histamine Research Society [et al]*. 2010;59(10):817-25. doi:10.1007/s00011-010-0193-5.
371. Churg A, Sin DD, Wright JL. Everything prevents emphysema: are animal models of cigarette smoke-induced chronic obstructive pulmonary disease any use? *American journal of respiratory cell and molecular biology*. 2011;45(6):1111-5. doi:10.1165/rcmb.2011-0087PS.
372. Plopper CG, Hyde DM. The non-human primate as a model for studying COPD and asthma. *Pulmonary pharmacology & therapeutics*. 2008;21(5):755-66. doi:10.1016/j.pupt.2008.01.008.
373. Shapiro SD. The use of transgenic mice for modeling airways disease. *Pulmonary pharmacology & therapeutics*. 2008;21(5):699-701. doi:10.1016/j.pupt.2008.01.006.
374. Shapiro SD. Animal models for chronic obstructive pulmonary disease: age of klotho and marlboro mice. *American journal of respiratory cell and molecular biology*. 2000;22(1):4-7. doi:10.1165/ajrcmb.22.1.f173.
375. Brusselle GG, Bracke KR, Maes T, D'Hulst A I, Moerloose KB, Joos GF et al. Murine models of COPD. *Pulmonary pharmacology & therapeutics*. 2006;19(3):155-65. doi:10.1016/j.pupt.2005.06.001.
376. Drannik AG, Pouladi MA, Robbins CS, Goncharova SI, Kianpour S, Stampfli MR. Impact of cigarette smoke on clearance and inflammation after *Pseudomonas aeruginosa* infection. *American journal of respiratory and critical care medicine*. 2004;170(11):1164-71. doi:10.1164/rccm.200311-1521OC.
377. Gaschler GJ, Skrtic M, Zavitz CC, Lindahl M, Onnervik PO, Murphy TF et al. Bacteria challenge in smoke-exposed mice exacerbates inflammation and skews the inflammatory profile. *American journal of respiratory and critical care medicine*. 2009;179(8):666-75. doi:10.1164/rccm.200808-1306OC.
378. Nikota JK, Shen P, Morissette MC, Fernandes K, Roos A, Chu DK et al. Cigarette smoke primes the pulmonary environment to IL-1alpha/CXCR-2-dependent nontypeable *Haemophilus influenzae*-exacerbated neutrophilia in mice. *Journal of immunology*. 2014;193(6):3134-45. doi:10.4049/jimmunol.1302412.
379. Harvey CJ, Thimmulappa RK, Sethi S, Kong X, Yarmus L, Brown RH et al. Targeting Nrf2 signaling improves bacterial clearance by alveolar macrophages in patients with COPD and in a mouse model. *Science translational medicine*. 2011;3(78):78ra32. doi:10.1126/scitranslmed.3002042.
380. Phipps JC, Aronoff DM, Curtis JL, Goel D, O'Brien E, Mancuso P. Cigarette smoke exposure impairs pulmonary bacterial clearance and alveolar macrophage complement-mediated phagocytosis of *Streptococcus pneumoniae*. *Infection and immunity*. 2010;78(3):1214-20. doi:10.1128/IAI.00963-09.
381. Ganesan S, Comstock AT, Kinker B, Mancuso P, Beck JM, Sajjan US. Combined exposure to cigarette smoke and nontypeable *Haemophilus influenzae* drives development of a COPD phenotype in mice. *Respiratory research*. 2014;15:11. doi:10.1186/1465-9921-15-11.
382. Bauer CMT, Morissette MC, Stampfli MR. The influence of cigarette smoking on viral infections: translating bench science to impact COPD pathogenesis and acute exacerbations of COPD clinically. *Chest*. 2013;143(1):196-206. doi:10.1378/chest.12-0930.
383. Del Vecchio AM, Branigan PJ, Barnathan ES, Flavin SK, Silkoff PE, Turner RB. Utility of animal and in vivo experimental infection of humans with rhinoviruses in the development of therapeutic agents for viral exacerbations of asthma and chronic obstructive pulmonary disease. *Pulmonary pharmacology & therapeutics*. 2015;30:32-43. doi:10.1016/j.pupt.2014.10.005.

384. Donovan C, Bourke JE, Vlahos R. Targeting the IL-33/IL-13 Axis for Respiratory Viral Infections. *Trends in pharmacological sciences*. 2016;37(4):252-61. doi:10.1016/j.tips.2016.01.004.
385. Robbins CS, Bauer CM, Vujicic N, Gaschler GJ, Lichty BD, Brown EG et al. Cigarette smoke impacts immune inflammatory responses to influenza in mice. *American journal of respiratory and critical care medicine*. 2006;174(12):1342-51. doi:10.1164/rccm.200604-561OC.
386. Kang MJ, Lee CG, Lee JY, Dela Cruz CS, Chen ZJ, Enelow R et al. Cigarette smoke selectively enhances viral PAMP- and virus-induced pulmonary innate immune and remodeling responses in mice. *The Journal of clinical investigation*. 2008;118(8):2771-84. doi:10.1172/JCI32709.
387. Gualano RC, Hansen MJ, Vlahos R, Jones JE, Park-Jones RA, Deliyannis G et al. Cigarette smoke worsens lung inflammation and impairs resolution of influenza infection in mice. *Respiratory research*. 2008;9:53. doi:10.1186/1465-9921-9-53.
388. Botelho FM, Bauer CM, Finch D, Nikota JK, Zavitz CC, Kelly A et al. IL-1 α /IL-1R1 expression in chronic obstructive pulmonary disease and mechanistic relevance to smoke-induced neutrophilia in mice. *PloS one*. 2011;6(12):e28457. doi:10.1371/journal.pone.0028457.
389. Kearley J, Silver JS, Sanden C, Liu Z, Berlin AA, White N et al. Cigarette smoke silences innate lymphoid cell function and facilitates an exacerbated type I interleukin-33-dependent response to infection. *Immunity*. 2015;42(3):566-79. doi:10.1016/j.immuni.2015.02.011.
390. Vlahos R, Bozinovski S. Preclinical murine models of Chronic Obstructive Pulmonary Disease. *European journal of pharmacology*. 2015;759:265-71. doi:10.1016/j.ejphar.2015.03.029.
391. Lo Sasso G, Schlage WK, Boue S, Veljkovic E, Peitsch MC, Hoeng J. The Apoe(-/-) mouse model: a suitable model to study cardiovascular and respiratory diseases in the context of cigarette smoke exposure and harm reduction. *Journal of translational medicine*. 2016;14(1):146. doi:10.1186/s12967-016-0901-1.
392. Plump AS, Smith JD, Hayek T, Aalto-Setälä K, Walsh A, Verstuyft JG et al. Severe hypercholesterolemia and atherosclerosis in apolipoprotein E-deficient mice created by homologous recombination in ES cells. *Cell*. 1992;71(2):343-53.
393. Libby P, Lichtman AH, Hansson GK. Immune effector mechanisms implicated in atherosclerosis: from mice to humans. *Immunity*. 2013;38(6):1092-104. doi:10.1016/j.immuni.2013.06.009.
394. Goldklang M, Golovatch P, Zelonina T, Trischler J, Rabinowitz D, Lemaitre V et al. Activation of the TLR4 signaling pathway and abnormal cholesterol efflux lead to emphysema in ApoE-deficient mice. *American journal of physiology Lung cellular and molecular physiology*. 2012;302(11):L1200-8. doi:10.1152/ajplung.00454.2010.
395. Lietz M, Berges A, Lebrun S, Meurrens K, Steffen Y, Stolle K et al. Cigarette-smoke-induced atherogenic lipid profiles in plasma and vascular tissue of apolipoprotein E-deficient mice are attenuated by smoking cessation. *Atherosclerosis*. 2013;229(1):86-93. doi:10.1016/j.atherosclerosis.2013.03.036.
396. Moore BB, Hogaboam CM. Murine models of pulmonary fibrosis. *American journal of physiology Lung cellular and molecular physiology*. 2008;294(2):L152-60. doi:10.1152/ajplung.00313.2007.
397. Cipriani P, Di Benedetto P, Ruscitti P, Verzella D, Fischietti M, Zazzeroni F et al. Macitentan inhibits the transforming growth factor-beta profibrotic action, blocking the signaling mediated by the ETR/TbetaRI complex in systemic sclerosis dermal fibroblasts. *Arthritis research & therapy*. 2015;17:247. doi:10.1186/s13075-015-0754-7.
398. Roman J, Brown KK, Olson A, Corcoran BM, Williams KJ, Group ATSCBoLFW. An official American thoracic society workshop report: comparative pathobiology of fibrosing lung disorders in humans and domestic animals. *Annals of the American Thoracic Society*. 2013;10(6):S224-9. doi:10.1513/AnnalsATS.201309-321ST.
399. Miele A, Dhaliwal K, Du Toit N, Murchison JT, Dhaliwal C, Brooks H et al. Chronic pleuropulmonary fibrosis and elastosis of aged donkeys: similarities to human pleuroparenchymal fibroelastosis. *Chest*. 2014;145(6):1325-32. doi:10.1378/chest.13-1306.
400. Bonvillain RW, Danchuk S, Sullivan DE, Betancourt AM, Semon JA, Eagle ME et al. A nonhuman primate model of lung regeneration: detergent-mediated decellularization and initial in vitro

recellularization with mesenchymal stem cells. *Tissue engineering Part A*. 2012;18(23-24):2437-52. doi:10.1089/ten.TEA.2011.0594.

401. Daly AB, Wallis JM, Borg ZD, Bonvillain RW, Deng B, Ballif BA et al. Initial binding and recellularization of decellularized mouse lung scaffolds with bone marrow-derived mesenchymal stromal cells. *Tissue engineering Part A*. 2012;18(1-2):1-16. doi:10.1089/ten.TEA.2011.0301.

402. Wagner DE, Bonenfant NR, Sokocevic D, DeSarno MJ, Borg ZD, Parsons CS et al. Three-dimensional scaffolds of acellular human and porcine lungs for high throughput studies of lung disease and regeneration. *Biomaterials*. 2014;35(9):2664-79. doi:10.1016/j.biomaterials.2013.11.078.

403. Wagner DE, Bonenfant NR, Parsons CS, Sokocevic D, Brooks EM, Borg ZD et al. Comparative decellularization and recellularization of normal versus emphysematous human lungs. *Biomaterials*. 2014;35(10):3281-97. doi:10.1016/j.biomaterials.2013.12.103.

404. Alsafadi HN, Staab-Weijnitz CA, Lehmann M, Lindner M, Peschel B, Konigshoff M et al. An ex vivo model to induce early fibrosis-like changes in human precision-cut lung slices. *American journal of physiology Lung cellular and molecular physiology*. 2017;312(6):L896-L902. doi:10.1152/ajplung.00084.2017.

405. Hansen NU, Karsdal MA, Brockbank S, Cruwys S, Ronnow S, Leeming DJ. Tissue turnover of collagen type I, III and elastin is elevated in the PCLS model of IPF and can be restored back to vehicle levels using a phosphodiesterase inhibitor. *Respiratory research*. 2016;17(1):76. doi:10.1186/s12931-016-0394-8.

406. Tatler AL, Barnes J, Habgood A, Goodwin A, McAnulty RJ, Jenkins G. Caffeine inhibits TGFbeta activation in epithelial cells, interrupts fibroblast responses to TGFbeta, and reduces established fibrosis in ex vivo precision-cut lung slices. *Thorax*. 2016;71(6):565-7. doi:10.1136/thoraxjnl-2015-208215.

407. Suki B, Majumdar A, Nugent MA, Bates JH. In silico modeling of interstitial lung mechanics: implications for disease development and repair. *Drug discovery today Disease models*. 2007;4(3):139-45. doi:10.1016/j.ddmod.2007.10.002.

408. Briles DE, Crain MJ, Gray BM, Forman C, Yother J. Strong association between capsular type and virulence for mice among human isolates of *Streptococcus pneumoniae*. *Infect Immun*. 1992;60(1):111-6.

409. Azoulay-Dupuis E, Vallee E, Veber B, Bedos JP, Bauchet J, Pocard JJ. In vivo efficacy of a new fluoroquinolone, sparfloxacin, against penicillin-susceptible and -resistant and multiresistant strains of *Streptococcus pneumoniae* in a mouse model of pneumonia. *Antimicrobial agents and chemotherapy*. 1992;36(12):2698-703.

410. Tateda K, Takashima K, Miyazaki H, Matsumoto T, Hatori T, Yamaguchi K. Noncompromised penicillin-resistant pneumococcal pneumonia CBA/J mouse model and comparative efficacies of antibiotics in this model. *Antimicrobial agents and chemotherapy*. 1996;40(6):1520-5.

411. Orihuela CJ, Gao G, McGee M, Yu J, Francis KP, Tuomanen E. Organ-specific models of *Streptococcus pneumoniae* disease. *Scandinavian journal of infectious diseases*. 2003;35(9):647-52.

412. Gingles NA, Alexander JE, Kadioglu A, Andrew PW, Kerr A, Mitchell TJ et al. Role of genetic resistance in invasive pneumococcal infection: identification and study of susceptibility and resistance in inbred mouse strains. *Infect Immun*. 2001;69(1):426-34. doi:10.1128/IAI.69.1.426-434.2001.

413. Anas AA, de Vos AF, Hoogendijk AJ, van Lieshout MH, van Heijst JW, Florquin S et al. Endoplasmic reticulum chaperone gp96 in macrophages is essential for protective immunity during Gram-negative pneumonia. *The Journal of pathology*. 2016;238(1):74-84. doi:10.1002/path.4637.

414. de Stoppelaar SF, Van't Veer C, Roelofs JJ, Claushuis TA, de Boer OJ, Tanck MW et al. Platelet and endothelial cell P-selectin are required for host defense against *Klebsiella pneumoniae*-induced pneumosepsis. *Journal of thrombosis and haemostasis : JTH*. 2015;13(6):1128-38. doi:10.1111/jth.12893.

415. Hackstein H, Lippitsch A, Krug P, Schevtschenko I, Kranz S, Hecker M et al. Prospectively defined murine mesenchymal stem cells inhibit *Klebsiella pneumoniae*-induced acute lung injury and improve pneumonia survival. *Respiratory research*. 2015;16:123. doi:10.1186/s12931-015-0288-1.

416. Rosen DA, Hilliard JK, Tiemann KM, Todd EM, Morley SC, Hunstad DA. *Klebsiella pneumoniae* FimK Promotes Virulence in Murine Pneumonia. *J Infect Dis.* 2016;213(4):649-58. doi:10.1093/infdis/jiv440.
417. Xiong H, Carter RA, Leiner IM, Tang YW, Chen L, Kreiswirth BN et al. Distinct Contributions of Neutrophils and CCR2+ Monocytes to Pulmonary Clearance of Different *Klebsiella pneumoniae* Strains. *Infect Immun.* 2015;83(9):3418-27. doi:10.1128/IAI.00678-15.
418. Pereira MS, Morgantetti GF, Massis LM, Horta CV, Hori JI, Zamboni DS. Activation of NLRC4 by flagellated bacteria triggers caspase-1-dependent and -independent responses to restrict *Legionella pneumophila* replication in macrophages and in vivo. *J Immunol.* 2011;187(12):6447-55. doi:10.4049/jimmunol.1003784.
419. Amer A, Franchi L, Kanneganti TD, Body-Malapel M, Ozoren N, Brady G et al. Regulation of *Legionella* phagosome maturation and infection through flagellin and host Ipaf. *The Journal of biological chemistry.* 2006;281(46):35217-23. doi:10.1074/jbc.M604933200.
420. Zamboni DS, Kobayashi KS, Kohlsdorf T, Ogura Y, Long EM, Vance RE et al. The Birc1e cytosolic pattern-recognition receptor contributes to the detection and control of *Legionella pneumophila* infection. *Nature immunology.* 2006;7(3):318-25. doi:10.1038/ni1305.
421. Pereira MS, Marques GG, Dellama JE, Zamboni DS. The Nlrc4 Inflammasome Contributes to Restriction of Pulmonary Infection by Flagellated *Legionella* spp. that Trigger Pyroptosis. *Frontiers in microbiology.* 2011;2:33. doi:10.3389/fmicb.2011.00033.
422. Ren T, Zamboni DS, Roy CR, Dietrich WF, Vance RE. Flagellin-deficient *Legionella* mutants evade caspase-1- and Naip5-mediated macrophage immunity. *PLoS pathogens.* 2006;2(3):e18. doi:10.1371/journal.ppat.0020018.
423. Rouse MS, Steckelberg JM. Animal models of gram-negative bacillary experimental pneumonia. In: Zak O, Sande MA, editors. *Handbook of animal models of infection: Experimental models in antimicrobial chemotherapy.* London: Academic Press; 1999. p. 495-500.
424. Feng N, Wang Q, Zhou J, Li J, Wen X, Chen S et al. Keratinocyte growth factor-2 inhibits bacterial infection with *Pseudomonas aeruginosa* pneumonia in a mouse model. *Journal of infection and chemotherapy : official journal of the Japan Society of Chemotherapy.* 2016;22(1):44-52. doi:10.1016/j.jiac.2015.10.005.
425. Starke JR, Edwards MS, Langston C, Baker CJ. A mouse model of chronic pulmonary infection with *Pseudomonas aeruginosa* and *Pseudomonas cepacia*. *Pediatric research.* 1987;22(6):698-702. doi:10.1203/00006450-198712000-00017.
426. van Heeckeren AM, Schluchter MD. Murine models of chronic *Pseudomonas aeruginosa* lung infection. *Laboratory animals.* 2002;36(3):291-312.
427. Stotland PK, Radzioch D, Stevenson MM. Mouse models of chronic lung infection with *Pseudomonas aeruginosa*: models for the study of cystic fibrosis. *Pediatric pulmonology.* 2000;30(5):413-24.
428. Stevenson MM, Kondratieva TK, Apt AS, Tam MF, Skamene E. In vitro and in vivo T cell responses in mice during bronchopulmonary infection with mucoid *Pseudomonas aeruginosa*. *Clinical and experimental immunology.* 1995;99(1):98-105.
429. Tam M, Snipes GJ, Stevenson MM. Characterization of chronic bronchopulmonary *Pseudomonas aeruginosa* infection in resistant and susceptible inbred mouse strains. *American journal of respiratory cell and molecular biology.* 1999;20(4):710-9. doi:10.1165/ajrcmb.20.4.3223.
430. Sapru K, Stotland PK, Stevenson MM. Quantitative and qualitative differences in bronchoalveolar inflammatory cells in *Pseudomonas aeruginosa*-resistant and -susceptible mice. *Clinical and experimental immunology.* 1999;115(1):103-9.
431. Guilbault C, Stotland P, Lachance C, Tam M, Keller A, Thompson-Snipes L et al. Influence of gender and interleukin-10 deficiency on the inflammatory response during lung infection with *Pseudomonas aeruginosa* in mice. *Immunology.* 2002;107(3):297-305.
432. Yang ZP, Kuo CC, Grayston JT. A mouse model of *Chlamydia pneumoniae* strain TWAR pneumonitis. *Infect Immun.* 1993;61(5):2037-40.

433. Yang ZP, Kuo CC, Grayston JT. Systemic dissemination of *Chlamydia pneumoniae* following intranasal inoculation in mice. *J Infect Dis.* 1995;171(3):736-8.
434. Moazed TC, Kuo CC, Grayston JT, Campbell LA. Evidence of systemic dissemination of *Chlamydia pneumoniae* via macrophages in the mouse. *J Infect Dis.* 1998;177(5):1322-5.
435. Moazed TC, Kuo C, Grayston JT, Campbell LA. Murine models of *Chlamydia pneumoniae* infection and atherosclerosis. *J Infect Dis.* 1997;175(4):883-90.
436. Dutow P, Lingner S, Laudeley R, Glage S, Hoymann HG, Dittrich AM et al. Severity of allergic airway disease due to house dust mite allergen is not increased after clinical recovery of lung infection with *Chlamydia pneumoniae* in mice. *Infect Immun.* 2013;81(9):3366-74. doi:10.1128/IAI.00334-13.
437. Janik K, Bode J, Dutow P, Laudeley R, Geffers R, Sommer K et al. Temperature and host cell-dependent changes in virulence of *Chlamydia pneumoniae* CWL029 in an optimized mouse infection model. *Pathogens and disease.* 2015;73(1):1-8. doi:10.1093/femspd/ftu001.
438. Sommer K, Njau F, Wittkop U, Thalmann J, Bartling G, Wagner A et al. Identification of high- and low-virulent strains of *Chlamydia pneumoniae* by their characterization in a mouse pneumonia model. *FEMS immunology and medical microbiology.* 2009;55(2):206-14. doi:10.1111/j.1574-695X.2008.00503.x.
439. Thangavel RR, Bouvier NM. Animal models for influenza virus pathogenesis, transmission, and immunology. *Journal of immunological methods.* 2014;410:60-79. doi:10.1016/j.jim.2014.03.023.
440. Bouvier NM, Lowen AC. Animal Models for Influenza Virus Pathogenesis and Transmission. *Viruses.* 2010;2(8):1530-63. doi:10.3390/v20801530.
441. Fukushi M, Ito T, Oka T, Kitazawa T, Miyoshi-Akiyama T, Kirikae T et al. Serial histopathological examination of the lungs of mice infected with influenza A virus PR8 strain. *PloS one.* 2011;6(6):e21207. doi:10.1371/journal.pone.0021207.
442. Prokopyeva EA, Sobolev IA, Prokopyev MV, Shestopalov AM. Adaptation of influenza A(H1N1)pdm09 virus in experimental mouse models. *Infection, genetics and evolution : journal of molecular epidemiology and evolutionary genetics in infectious diseases.* 2016;39:265-71. doi:10.1016/j.meegid.2016.01.022.
443. Pica N, Iyer A, Ramos I, Bouvier NM, Fernandez-Sesma A, Garcia-Sastre A et al. The DBA.2 mouse is susceptible to disease following infection with a broad, but limited, range of influenza A and B viruses. *Journal of virology.* 2011;85(23):12825-9. doi:10.1128/JVI.05930-11.
444. Srivastava B, Blazejewska P, Hessmann M, Bruder D, Geffers R, Mauel S et al. Host genetic background strongly influences the response to influenza A virus infections. *PloS one.* 2009;4(3):e4857. doi:10.1371/journal.pone.0004857.
445. Force ADT, Ranieri VM, Rubenfeld GD, Thompson BT, Ferguson ND, Caldwell E et al. Acute respiratory distress syndrome: the Berlin Definition. *Jama.* 2012;307(23):2526-33. doi:10.1001/jama.2012.5669.
446. Matute-Bello G, Downey G, Moore BB, Groshong SD, Matthay MA, Slutsky AS et al. An official American Thoracic Society workshop report: features and measurements of experimental acute lung injury in animals. *American journal of respiratory cell and molecular biology.* 2011;44(5):725-38. doi:10.1165/rcmb.2009-0210ST.
447. Germain M, Eyries M, Montani D, Poirier O, Girerd B, Dorfmüller P et al. Genome-wide association analysis identifies a susceptibility locus for pulmonary arterial hypertension. *Nature genetics.* 2013;45(5):518-21. doi:10.1038/ng.2581.
448. Vattulainen S, Aho J, Salmenperä P, Bruce S, Tallila J, Gentile M et al. Accurate genetic diagnosis of Finnish pulmonary arterial hypertension patients using oligonucleotide-selective sequencing. *Molecular genetics & genomic medicine.* 2015;3(4):354-62. doi:10.1002/mgg3.147.
449. Austin ED, Ma L, LeDuc C, Berman Rosenzweig E, Borczuk A, Phillips JA, 3rd et al. Whole exome sequencing to identify a novel gene (caveolin-1) associated with human pulmonary arterial hypertension. *Circulation Cardiovascular genetics.* 2012;5(3):336-43. doi:10.1161/CIRCGENETICS.111.961888.

450. Gomez J, Reguero JR, Alvarez C, Junquera MR, Arango A, Moris C et al. A Semiconductor Chip-Based Next Generation Sequencing Procedure for the Main Pulmonary Hypertension Genes. *Lung*. 2015;193(4):571-4. doi:10.1007/s00408-015-9736-4.
451. Aldred MA, Comhair SA, Varella-Garcia M, Asosingh K, Xu W, Noon GP et al. Somatic chromosome abnormalities in the lungs of patients with pulmonary arterial hypertension. *American journal of respiratory and critical care medicine*. 2010;182(9):1153-60. doi:10.1164/rccm.201003-0491OC.
452. Yeager ME, Halley GR, Golpon HA, Voelkel NF, Tudor RM. Microsatellite instability of endothelial cell growth and apoptosis genes within plexiform lesions in primary pulmonary hypertension. *Circulation research*. 2001;88(1):E2-E11.
453. Machado RD, James V, Southwood M, Harrison RE, Atkinson C, Stewart S et al. Investigation of second genetic hits at the BMPR2 locus as a modulator of disease progression in familial pulmonary arterial hypertension. *Circulation*. 2005;111(5):607-13. doi:10.1161/01.CIR.0000154543.07679.08.
454. Gu M, Shao NY, Sa S, Li D, Termglinchan V, Ameen M et al. Patient-Specific iPSC-Derived Endothelial Cells Uncover Pathways that Protect against Pulmonary Hypertension in BMPR2 Mutation Carriers. *Cell Stem Cell*. 2017;20(4):490-504 e5. doi:10.1016/j.stem.2016.08.019.
455. Sa S, Gu M, Chappell J, Shao NY, Ameen M, Elliott KA et al. Induced Pluripotent Stem Cell Model of Pulmonary Arterial Hypertension Reveals Novel Gene Expression and Patient Specificity. *American journal of respiratory and critical care medicine*. 2017;195(7):930-41. doi:10.1164/rccm.201606-1200OC.
456. Rabinovitch M. Combining induced pluripotent stem cell with next generation sequencing technology to gain new insights into pathobiology and treatment of pulmonary arterial hypertension. *Pulmonary circulation*. 2013;3(1):153-5. doi:10.4103/2045-8932.109963.
457. Kataoka M, Aimi Y, Yanagisawa R, Ono M, Oka A, Fukuda K et al. Alu-mediated nonallelic homologous and nonhomologous recombination in the BMPR2 gene in heritable pulmonary arterial hypertension. *Genetics in medicine : official journal of the American College of Medical Genetics*. 2013;15(12):941-7. doi:10.1038/gim.2013.41.
458. Mutlu Z, Kayikcioglu M, Nalbantgil S, Vuran O, Kemal H, Mogulkoc N et al. Sequencing of mutations in the serine/threonine kinase domain of the bone morphogenetic protein receptor type 2 gene causing pulmonary arterial hypertension. *Anatolian journal of cardiology*. 2015. doi:10.5152/AnatolJCardiol.2015.6297.
459. Deng Z, Morse JH, Slager SL, Cuervo N, Moore KJ, Venetos G et al. Familial primary pulmonary hypertension (gene PPH1) is caused by mutations in the bone morphogenetic protein receptor-II gene. *American journal of human genetics*. 2000;67(3):737-44. doi:10.1086/303059.
460. Lane KB, Machado RD, Pauculo MW, Thomson JR, Phillips JA, 3rd, Loyd JE et al. Heterozygous germline mutations in BMPR2, encoding a TGF-beta receptor, cause familial primary pulmonary hypertension. *Nature genetics*. 2000;26(1):81-4. doi:10.1038/79226.
461. Chaouat A, Coulet F, Favre C, Simonneau G, Weitzenblum E, Soubrier F et al. Endoglin germline mutation in a patient with hereditary haemorrhagic telangiectasia and dexfenfluramine associated pulmonary arterial hypertension. *Thorax*. 2004;59(5):446-8.
462. Harrison RE, Berger R, Haworth SG, Tulloh R, Mache CJ, Morrell NW et al. Transforming growth factor-beta receptor mutations and pulmonary arterial hypertension in childhood. *Circulation*. 2005;111(4):435-41. doi:10.1161/01.CIR.0000153798.78540.87.
463. Abdalla SA, Gallione CJ, Barst RJ, Horn EM, Knowles JA, Marchuk DA et al. Primary pulmonary hypertension in families with hereditary haemorrhagic telangiectasia. *The European respiratory journal*. 2004;23(3):373-7.
464. Ma L, Roman-Campos D, Austin ED, Eyries M, Sampson KS, Soubrier F et al. A novel channelopathy in pulmonary arterial hypertension. *The New England journal of medicine*. 2013;369(4):351-61. doi:10.1056/NEJMoa1211097.
465. Shintani M, Yagi H, Nakayama T, Saji T, Matsuoka R. A new nonsense mutation of SMAD8 associated with pulmonary arterial hypertension. *Journal of medical genetics*. 2009;46(5):331-7. doi:10.1136/jmg.2008.062703.

466. Chida A, Shintani M, Nakayama T, Furutani Y, Hayama E, Inai K et al. Missense mutations of the BMPR1B (ALK6) gene in childhood idiopathic pulmonary arterial hypertension. *Circulation journal : official journal of the Japanese Circulation Society*. 2012;76(6):1501-8.
467. Chida A, Shintani M, Matsushita Y, Sato H, Eitoku T, Nakayama T et al. Mutations of NOTCH3 in childhood pulmonary arterial hypertension. *Molecular genetics & genomic medicine*. 2014;2(3):229-39. doi:10.1002/mgg3.58.
468. Yang Y, Chen D, Yuan Z, Fang F, Cheng X, Xia J et al. Megakaryocytic leukemia 1 (MKL1) ties the epigenetic machinery to hypoxia-induced transactivation of endothelin-1. *Nucleic acids research*. 2013;41(12):6005-17. doi:10.1093/nar/gkt311.
469. Navas Tejedor P, Tenorio Castano J, Palomino Doza J, Arias Lajara P, Gordo Trujillo G, Lopez Meseguer M et al. An homozygous mutation in KCNK3 is associated with an aggressive form of hereditary pulmonary arterial hypertension. *Clin Genet*. 2017;91(3):453-7. doi:10.1111/cge.12869.
470. Navas P, Tenorio J, Quezada CA, Barrios E, Gordo G, Arias P et al. Molecular Analysis of BMPR2, TBX4, and KCNK3 and Genotype-Phenotype Correlations in Spanish Patients and Families With Idiopathic and Hereditary Pulmonary Arterial Hypertension. *Rev Esp Cardiol (Engl Ed)*. 2016;69(11):1011-9. doi:10.1016/j.rec.2016.03.029.
471. Levy M, Eyries M, Szezepanski I, Ladouceur M, Nadaud S, Bonnet D et al. Genetic analyses in a cohort of children with pulmonary hypertension. *The European respiratory journal*. 2016;48(4):1118-26. doi:10.1183/13993003.00211-2016.
472. Kerstjens-Frederikse WS, Bongers EM, Roofthoof MT, Leter EM, Douwes JM, Van Dijk A et al. TBX4 mutations (small patella syndrome) are associated with childhood-onset pulmonary arterial hypertension. *Journal of medical genetics*. 2013;50(8):500-6. doi:10.1136/jmedgenet-2012-101152.
473. Nimmakayalu M, Major H, Sheffield V, Solomon DH, Smith RJ, Patil SR et al. Microdeletion of 17q22q23.2 encompassing TBX2 and TBX4 in a patient with congenital microcephaly, thyroid duct cyst, sensorineural hearing loss, and pulmonary hypertension. *Am J Med Genet A*. 2011;155A(2):418-23. doi:10.1002/ajmg.a.33827.
474. Evans JD, Girerd B, Montani D, Wang XJ, Galie N, Austin ED et al. BMPR2 mutations and survival in pulmonary arterial hypertension: an individual participant data meta-analysis. *Lancet Respir Med*. 2016;4(2):129-37. doi:10.1016/S2213-2600(15)00544-5.
475. Ghigna MR, Guignabert C, Montani D, Girerd B, Jais X, Savale L et al. BMPR2 mutation status influences bronchial vascular changes in pulmonary arterial hypertension. *The European respiratory journal*. 2016;48(6):1668-81. doi:10.1183/13993003.00464-2016.
476. Eichstaedt CA, Song J, Viales RR, Pan Z, Benjamin N, Fischer C et al. First identification of Kruppel-like factor 2 mutation in heritable pulmonary arterial hypertension. *Clin Sci (Lond)*. 2017;131(8):689-98. doi:10.1042/CS20160930.
477. Song J, Eichstaedt CA, Viales RR, Benjamin N, Harutyunova S, Fischer C et al. Identification of genetic defects in pulmonary arterial hypertension by a new gene panel diagnostic tool. *Clin Sci (Lond)*. 2016;130(22):2043-52. doi:10.1042/CS20160531.
478. Meloche J, Potus F, Vaillancourt M, Bourgeois A, Johnson I, Deschamps L et al. Bromodomain-Containing Protein 4: The Epigenetic Origin of Pulmonary Arterial Hypertension. *Circulation research*. 2015;117(6):525-35. doi:10.1161/CIRCRESAHA.115.307004.
479. Yang Q, Sun M, Ramchandran R, Raj JU. IGF-1 signaling in neonatal hypoxia-induced pulmonary hypertension: Role of epigenetic regulation. *Vascular pharmacology*. 2015;73:20-31. doi:10.1016/j.vph.2015.04.005.
480. Zelko IN, Folz RJ. Regulation of Oxidative Stress in Pulmonary Artery Endothelium. Modulation of Extracellular Superoxide Dismutase and NOX4 Expression Using Histone Deacetylase Class I Inhibitors. *American journal of respiratory cell and molecular biology*. 2015;53(4):513-24. doi:10.1165/rcmb.2014-0260OC.
481. Chen D, Yang Y, Cheng X, Fang F, Xu G, Yuan Z et al. Megakaryocytic leukemia 1 directs a histone H3 lysine 4 methyltransferase complex to regulate hypoxic pulmonary hypertension. *Hypertension*. 2015;65(4):821-33. doi:10.1161/HYPERTENSIONAHA.114.04585.

482. Zhang L, Tang L, Wei J, Lao L, Gu W, Hu Q et al. Extrauterine growth restriction on pulmonary vascular endothelial dysfunction in adult male rats: the role of epigenetic mechanisms. *Journal of hypertension*. 2014;32(11):2188-98; discussion 98. doi:10.1097/HJH.0000000000000309.
483. Chen D, Fang F, Yang Y, Chen J, Xu G, Xu Y et al. Brahma-related gene 1 (Brg1) epigenetically regulates CAM activation during hypoxic pulmonary hypertension. *Cardiovascular research*. 2013;100(3):363-73. doi:10.1093/cvr/cvt214.
484. Wang Y, Kahaleh B. Epigenetic repression of bone morphogenetic protein receptor II expression in scleroderma. *Journal of cellular and molecular medicine*. 2013;17(10):1291-9. doi:10.1111/jcmm.12105.
485. Xu XF, Lv Y, Gu WZ, Tang LL, Wei JK, Zhang LY et al. Epigenetics of hypoxic pulmonary arterial hypertension following intrauterine growth retardation rat: epigenetics in PAH following IUGR. *Respiratory research*. 2013;14:20. doi:10.1186/1465-9921-14-20.
486. Perros F, Cohen-Kaminsky S, Gambaryan N, Girerd B, Raymond N, Klingelschmitt I et al. Cytotoxic cells and granulysin in pulmonary arterial hypertension and pulmonary veno-occlusive disease. *American journal of respiratory and critical care medicine*. 2013;187(2):189-96. doi:10.1164/rccm.201208-1364OC.
487. Yang Q, Lu Z, Ramchandran R, Longo LD, Raj JU. Pulmonary artery smooth muscle cell proliferation and migration in fetal lambs acclimatized to high-altitude long-term hypoxia: role of histone acetylation. *American journal of physiology Lung cellular and molecular physiology*. 2012;303(11):L1001-10. doi:10.1152/ajplung.00092.2012.
488. Zhao L, Chen CN, Hajji N, Oliver E, Cotroneo E, Wharton J et al. Histone deacetylation inhibition in pulmonary hypertension: therapeutic potential of valproic acid and suberoylanilide hydroxamic acid. *Circulation*. 2012;126(4):455-67. doi:10.1161/CIRCULATIONAHA.112.103176.
489. Yang Q, Lu Z, Singh D, Raj JU. BIX-01294 treatment blocks cell proliferation, migration and contractility in ovine foetal pulmonary arterial smooth muscle cells. *Cell proliferation*. 2012;45(4):335-44. doi:10.1111/j.1365-2184.2012.00828.x.
490. Aljubran SA, Cox R, Jr., Tamarapu Parthasarathy P, Kollongod Ramanathan G, Rajanbabu V, Bao H et al. Enhancer of zeste homolog 2 induces pulmonary artery smooth muscle cell proliferation. *PloS one*. 2012;7(5):e37712. doi:10.1371/journal.pone.0037712.
491. Li M, Riddle SR, Frid MG, El Kasmi KC, McKinsey TA, Sokol RJ et al. Emergence of fibroblasts with a proinflammatory epigenetically altered phenotype in severe hypoxic pulmonary hypertension. *Journal of immunology*. 2011;187(5):2711-22. doi:10.4049/jimmunol.1100479.
492. Archer SL, Marsboom G, Kim GH, Zhang HJ, Toth PT, Svensson EC et al. Epigenetic attenuation of mitochondrial superoxide dismutase 2 in pulmonary arterial hypertension: a basis for excessive cell proliferation and a new therapeutic target. *Circulation*. 2010;121(24):2661-71. doi:10.1161/CIRCULATIONAHA.109.916098.
493. Chen F, Li X, Aquadro E, Haigh S, Zhou J, Stepp DW et al. Inhibition of histone deacetylase reduces transcription of NADPH oxidases and ROS production and ameliorates pulmonary arterial hypertension. *Free radical biology & medicine*. 2016;99:167-78. doi:10.1016/j.freeradbiomed.2016.08.003.
494. . !!! INVALID CITATION !!! 51-63.
495. Zhang H, Xu M, Xia J, Qin RY. Association between serotonin transporter (SERT) gene polymorphism and idiopathic pulmonary arterial hypertension: a meta-analysis and review of the literature. *Metabolism: clinical and experimental*. 2013;62(12):1867-75. doi:10.1016/j.metabol.2013.08.012.
496. Ulasli SS, Eyuboglu FO, Verdi H, Atac FB. Associations between endothelial nitric oxide synthase A/B, angiotensin converting enzyme I/D and serotonin transporter L/S gene polymorphisms with pulmonary hypertension in COPD patients. *Molecular biology reports*. 2013;40(10):5625-33. doi:10.1007/s11033-013-2664-6.
497. Li JF, Lin Y, Yang YH, Gan HL, Liang Y, Liu J et al. Fibrinogen Aalpha Thr312Ala polymorphism specifically contributes to chronic thromboembolic pulmonary hypertension by increasing fibrin resistance. *PloS one*. 2013;8(7):e69635. doi:10.1371/journal.pone.0069635.

498. Koumakis E, Wipff J, Dieude P, Ruiz B, Bouaziz M, Revillod L et al. TGFbeta receptor gene variants in systemic sclerosis-related pulmonary arterial hypertension: results from a multicentre EUSTAR study of European Caucasian patients. *Annals of the rheumatic diseases*. 2012;71(11):1900-3. doi:10.1136/annrheumdis-2012-201755.
499. Byers HM, Dagle JM, Klein JM, Ryckman KK, McDonald EL, Murray JC et al. Variations in CRHR1 are associated with persistent pulmonary hypertension of the newborn. *Pediatric research*. 2012;71(2):162-7. doi:10.1038/pr.2011.24.
500. Baloira A, Nunez M, Cifrian J, Vilarino C, Ojeda M, Valverde D. Polymorphisms in the serotonin transporter protein (SERT) gene in patients with pulmonary arterial hypertension. *Archivos de bronconeumologia*. 2012;48(3):77-80. doi:10.1016/j.arbres.2011.10.008.
501. Calabro P, Limongelli G, Maddaloni V, Vizza CD, D'Alto M, D'Alessandro R et al. Analysis of endothelin-1 and endothelin-1 receptor A gene polymorphisms in patients with pulmonary arterial hypertension. *Internal and emergency medicine*. 2012;7(5):425-30. doi:10.1007/s11739-011-0643-2.
502. De Jesus LC, Kazzi SN, Dahmer MK, Chen X, Quasney MW. Role of angiotensin-converting enzyme gene polymorphism in persistent pulmonary hypertension of the newborn. *Acta paediatrica*. 2011;100(10):1326-30. doi:10.1111/j.1651-2227.2011.02277.x.
503. Vadapalli S, Katta S, Sastry BK, Nallari P. MAO-A promoter polymorphism and idiopathic pulmonary arterial hypertension. *Journal of genetics*. 2010;89(4):e43-5.
504. Chen Z, Nakajima T, Tanabe N, Hinohara K, Sakao S, Kasahara Y et al. Susceptibility to chronic thromboembolic pulmonary hypertension may be conferred by miR-759 via its targeted interaction with polymorphic fibrinogen alpha gene. *Human genetics*. 2010;128(4):443-52. doi:10.1007/s00439-010-0866-8.
505. Wipff J, Dieude P, Guedj M, Ruiz B, Riemekasten G, Cracowski JL et al. Association of a KCNA5 gene polymorphism with systemic sclerosis-associated pulmonary arterial hypertension in the European Caucasian population. *Arthritis and rheumatism*. 2010;62(10):3093-100. doi:10.1002/art.27607.
506. Agarwal SK, Gourh P, Shete S, Paz G, Divecha D, Reveille JD et al. Association of interleukin 23 receptor polymorphisms with anti-topoisomerase-I positivity and pulmonary hypertension in systemic sclerosis. *The Journal of rheumatology*. 2009;36(12):2715-23. doi:10.3899/jrheum.090421.
507. Cao H, Gu H, Qiu W, Zuo W, Zheng L, Wang Z et al. Association study of serotonin transporter gene polymorphisms and ventricular septal defects related possible pulmonary arterial hypertension in Chinese population. *Clinical and experimental hypertension*. 2009;31(7):605-14. doi:10.3109/10641960902993061.
508. Rodriguez-Murillo L, Subaran R, Stewart WC, Pramanik S, Marathe S, Barst RJ et al. Novel loci interacting epistatically with bone morphogenetic protein receptor 2 cause familial pulmonary arterial hypertension. *The Journal of heart and lung transplantation : the official publication of the International Society for Heart Transplantation*. 2010;29(2):174-80. doi:10.1016/j.healun.2009.08.022.
509. Ulrich S, Szamalek-Hoegel J, Hersberger M, Fischler M, Garcia JS, Huber LC et al. Sequence variants in BMPR2 and genes involved in the serotonin and nitric oxide pathways in idiopathic pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension: relation to clinical parameters and comparison with left heart disease. *Respiration; international review of thoracic diseases*. 2010;79(4):279-87. doi:10.1159/000250322.
510. Ulrich S, Hersberger M, Fischler M, Nussbaumer-Ochsner Y, Treder U, Russi EW et al. Genetic polymorphisms of the serotonin transporter, but not the 2a receptor or nitric oxide synthetase, are associated with pulmonary hypertension in chronic obstructive pulmonary disease. *Respiration; international review of thoracic diseases*. 2010;79(4):288-95. doi:10.1159/000226243.
511. Yu Y, Keller SH, Remillard CV, Safrina O, Nicholson A, Zhang SL et al. A functional single-nucleotide polymorphism in the TRPC6 gene promoter associated with idiopathic pulmonary arterial hypertension. *Circulation*. 2009;119(17):2313-22. doi:10.1161/CIRCULATIONAHA.108.782458.
512. Chung WK, Deng L, Carroll JS, Mallory N, Diamond B, Rosenzweig EB et al. Polymorphism in the angiotensin II type 1 receptor (AGTR1) is associated with age at diagnosis in pulmonary arterial

hypertension. *The Journal of heart and lung transplantation : the official publication of the International Society for Heart Transplantation*. 2009;28(4):373-9. doi:10.1016/j.healun.2009.01.016.

513. Roberts KE, Fallon MB, Krowka MJ, Benza RL, Knowles JA, Badesch DB et al. Serotonin transporter polymorphisms in patients with portopulmonary hypertension. *Chest*. 2009;135(6):1470-5. doi:10.1378/chest.08-1909.

514. Manetti M, Liakouli V, Fatini C, Cipriani P, Bonino C, Vettori S et al. Association between a stromal cell-derived factor 1 (SDF-1/CXCL12) gene polymorphism and microvascular disease in systemic sclerosis. *Annals of the rheumatic diseases*. 2009;68(3):408-11. doi:10.1136/ard.2008.098277.

515. Phillips JA, 3rd, Poling JS, Phillips CA, Stanton KC, Austin ED, Cogan JD et al. Synergistic heterozygosity for TGFbeta1 SNPs and BMPR2 mutations modulates the age at diagnosis and penetrance of familial pulmonary arterial hypertension. *Genetics in medicine : official journal of the American College of Medical Genetics*. 2008;10(5):359-65. doi:10.1097/GIM.0b013e318172dcdf.

516. Katta S, Vadapalli S, Sastry BK, Nallari P. t-plasminogen activator inhibitor-1 polymorphism in idiopathic pulmonary arterial hypertension. *Indian journal of human genetics*. 2008;14(2):37-40. doi:10.4103/0971-6866.44103.

517. Cua CL, Cooke G, Taylor M, Hayes J, Waldon L, Lipowski P et al. Endothelial nitric oxide synthase polymorphisms associated with abnormal nitric oxide production are not over-represented in children with Down syndrome. *Congenital heart disease*. 2006;1(4):169-74. doi:10.1111/j.1747-0803.2006.00029.x.

518. Ashley-Koch AE, Elliott L, Kail ME, De Castro LM, Jonassaint J, Jackson TL et al. Identification of genetic polymorphisms associated with risk for pulmonary hypertension in sickle cell disease. *Blood*. 2008;111(12):5721-6. doi:10.1182/blood-2007-02-074849.

519. Suntharalingam J, Goldsmith K, van Marion V, Long L, Treacy CM, Dudbridge F et al. Fibrinogen Aalpha Thr312Ala polymorphism is associated with chronic thromboembolic pulmonary hypertension. *The European respiratory journal*. 2008;31(4):736-41. doi:10.1183/09031936.00055107.

520. Izikki M, Hanoun N, Marcos E, Savale L, Barlier-Mur AM, Saurini F et al. Tryptophan hydroxylase 1 knockout and tryptophan hydroxylase 2 polymorphism: effects on hypoxic pulmonary hypertension in mice. *American journal of physiology Lung cellular and molecular physiology*. 2007;293(4):L1045-52. doi:10.1152/ajplung.00082.2007.

521. Remillard CV, Tigno DD, Platoshyn O, Burg ED, Brevnova EE, Conger D et al. Function of Kv1.5 channels and genetic variations of KCNA5 in patients with idiopathic pulmonary arterial hypertension. *American journal of physiology Cell physiology*. 2007;292(5):C1837-53. doi:10.1152/ajpcell.00405.2006.

522. Wipff J, Kahan A, Hachulla E, Sibilia J, Cabane J, Meyer O et al. Association between an endoglin gene polymorphism and systemic sclerosis-related pulmonary arterial hypertension. *Rheumatology*. 2007;46(4):622-5. doi:10.1093/rheumatology/kel378.

523. Tanabe N, Amano S, Tatsumi K, Kominami S, Igarashi N, Shimura R et al. Angiotensin-converting enzyme gene polymorphisms and prognosis in chronic thromboembolic pulmonary hypertension. *Circulation journal : official journal of the Japanese Circulation Society*. 2006;70(9):1174-9.

524. Eddahibi S, Chaouat A, Tu L, Chouaid C, Weitzenblum E, Housset B et al. Interleukin-6 gene polymorphism confers susceptibility to pulmonary hypertension in chronic obstructive pulmonary disease. *Proceedings of the American Thoracic Society*. 2006;3(6):475-6. doi:10.1513/pats.200603-038MS.

525. Kawaguchi Y, Tochimoto A, Hara M, Kawamoto M, Sugiura T, Katsumata Y et al. NOS2 polymorphisms associated with the susceptibility to pulmonary arterial hypertension with systemic sclerosis: contribution to the transcriptional activity. *Arthritis research & therapy*. 2006;8(4):R104. doi:10.1186/ar1984.

526. Marasini B, Cossutta R, Selmi C, Pozzi MR, Gardinali M, Massarotti M et al. Polymorphism of the fractalkine receptor CX3CR1 and systemic sclerosis-associated pulmonary arterial hypertension. *Clinical & developmental immunology*. 2005;12(4):275-9. doi:10.1080/17402520500303297.

527. Machado RD, Koehler R, Glissmeyer E, Veal C, Suntharalingam J, Kim M et al. Genetic association of the serotonin transporter in pulmonary arterial hypertension. *American journal of respiratory and critical care medicine*. 2006;173(7):793-7. doi:10.1164/rccm.200509-1365OC.
528. Koehler R, Olschewski H, Hoepfer M, Janssen B, Grunig E. Serotonin transporter gene polymorphism in a cohort of German patients with idiopathic pulmonary arterial hypertension or chronic thromboembolic pulmonary hypertension. *Chest*. 2005;128(6 Suppl):619S. doi:10.1378/chest.128.6_suppl.619S.
529. Willers ED, Newman JH, Loyd JE, Robbins IM, Wheeler LA, Prince MA et al. Serotonin transporter polymorphisms in familial and idiopathic pulmonary arterial hypertension. *American journal of respiratory and critical care medicine*. 2006;173(7):798-802. doi:10.1164/rccm.200509-1361OC.
530. Vachharajani A, Saunders S. Allelic variation in the serotonin transporter (5HTT) gene contributes to idiopathic pulmonary hypertension in children. *Biochemical and biophysical research communications*. 2005;334(2):376-9. doi:10.1016/j.bbrc.2005.06.107.
531. Amano S, Tatsumi K, Tanabe N, Kasahara Y, Kurosu K, Takiguchi Y et al. Polymorphism of the promoter region of prostacyclin synthase gene in chronic thromboembolic pulmonary hypertension. *Respirology*. 2004;9(2):184-9. doi:10.1111/j.1440-1843.2004.00568.x.
532. Solari V, Puri P. Genetic polymorphisms of angiotensin system genes in congenital diaphragmatic hernia associated with persistent pulmonary hypertension. *Journal of pediatric surgery*. 2004;39(3):302-6; discussion -6.
533. Yildiz P, Oflaz H, Cine N, Erginel-Unaltuna N, Erzen F, Yilmaz V. Gene polymorphisms of endothelial nitric oxide synthase enzyme associated with pulmonary hypertension in patients with COPD. *Respiratory medicine*. 2003;97(12):1282-8.
534. Eddahibi S, Chaouat A, Morrell N, Fadel E, Fuhrman C, Bugnet AS et al. Polymorphism of the serotonin transporter gene and pulmonary hypertension in chronic obstructive pulmonary disease. *Circulation*. 2003;108(15):1839-44. doi:10.1161/01.CIR.0000091409.53101.E8.
535. Kanazawa H, Okamoto T, Hirata K, Yoshikawa J. Deletion polymorphisms in the angiotensin converting enzyme gene are associated with pulmonary hypertension evoked by exercise challenge in patients with chronic obstructive pulmonary disease. *American journal of respiratory and critical care medicine*. 2000;162(4 Pt 1):1235-8. doi:10.1164/ajrccm.162.4.9909120.
536. Gonzalez Ordonez AJ, Fernandez Carreira JM, Medina Rodriguez JM, Martin Sanchez L, Alvarez Diaz R, Alvarez Martinez MV et al. Risk of venous thromboembolism associated with the insertion/deletion polymorphism in the angiotensin-converting enzyme gene. *Blood coagulation & fibrinolysis : an international journal in haemostasis and thrombosis*. 2000;11(5):485-90.
537. Fang M, Huang Y, Zhang Y, Ning Z, Zhu L, Li X. Interleukin-6 -572C/G polymorphism is associated with serum interleukin-6 levels and risk of idiopathic pulmonary arterial hypertension. *Journal of the American Society of Hypertension : JASH*. 2017. doi:10.1016/j.jash.2017.01.011.
538. Zhuo Y, Zeng Q, Zhang P, Li G, Xie Q, Cheng Y. VEGF Promoter Polymorphism Confers an Increased Risk of Pulmonary Arterial Hypertension in a Chinese Population. *Yonsei Med J*. 2017;58(2):305-11. doi:10.3349/ymj.2017.58.2.305.
539. Trittmann JK, Jin Y, Chicoine LG, Liu Y, Chen B, Nelin LD. An arginase-1 SNP that protects against the development of pulmonary hypertension in bronchopulmonary dysplasia enhances NO-mediated apoptosis in lymphocytes. *Physiological reports*. 2016;4(22). doi:10.14814/phy2.13041.
540. Zhuo Y, Zeng Q, Zhang P, Li G, Xie Q, Cheng Y. Functional polymorphism of lncRNA MALAT1 contributes to pulmonary arterial hypertension susceptibility in Chinese people. *Clin Chem Lab Med*. 2017;55(1):38-46. doi:10.1515/cclm-2016-0056.
541. Nishimura R, Tanabe N, Sekine A, Kasai H, Suda R, Kato F et al. Synergistic Effects of ACE Insertion/Deletion and GNB3 C825T Polymorphisms on the Efficacy of PDE-5 Inhibitor in Patients with Pulmonary Hypertension. *Respiration; international review of thoracic diseases*. 2016;91(2):132-40. doi:10.1159/000443772.
542. Trittmann JK, Gastier-Foster JM, Zmuda EJ, Frick J, Rogers LK, Vieland VJ et al. A single nucleotide polymorphism in the dimethylarginine dimethylaminohydrolase gene is associated with

lower risk of pulmonary hypertension in bronchopulmonary dysplasia. *Acta paediatrica*. 2016;105(4):e170-5. doi:10.1111/apa.13296.

543. Ventetuolo CE, Mitra N, Wan F, Manichaikul A, Barr RG, Johnson C et al. Oestradiol metabolism and androgen receptor genotypes are associated with right ventricular function. *The European respiratory journal*. 2016;47(2):553-63. doi:10.1183/13993003.01083-2015.

544. Li XF, Song CH, Sheng HZ, Zhen DD, Pan M, Zhu JH. P-selectin gene polymorphism associates with pulmonary hypertension in congenital heart disease. *International journal of clinical and experimental pathology*. 2015;8(6):7189-95.

545. Penaloza D, Banchemo N, Sime F, Gamboa R. The Heart in Chronic Hypoxia. *Biochemical clinics*. 1963;2:283-98.

546. Sanchis-Gomar F, Vina J, Lippi G. Intermittent hypobaric hypoxia applicability in myocardial infarction prevention and recovery. *Journal of cellular and molecular medicine*. 2012;16(5):1150-4. doi:10.1111/j.1582-4934.2011.01508.x.

547. Millet GP, Roels B, Schmitt L, Woorons X, Richalet JP. Combining hypoxic methods for peak performance. *Sports medicine*. 2010;40(1):1-25. doi:10.2165/11317920-000000000-00000.

548. Herrera EA, Farias JG, Ebensperger G, Reyes RV, Llanos AJ, Castillo RL. Pharmacological approaches in either intermittent or permanent hypoxia: A tale of two exposures. *Pharmacological research*. 2015;101:94-101. doi:10.1016/j.phrs.2015.07.011.

549. Gassmann M, Muckenthaler MU. Adaptation of iron requirement to hypoxic conditions at high altitude. *Journal of applied physiology*. 2015;119(12):1432-40. doi:10.1152/jappphysiol.00248.2015.

550. Bishop T, Ratcliffe PJ. HIF hydroxylase pathways in cardiovascular physiology and medicine. *Circulation research*. 2015;117(1):65-79. doi:10.1161/CIRCRESAHA.117.305109.

551. Huetsch J, Shimoda LA. Na(+)/H(+) exchange and hypoxic pulmonary hypertension. *Pulmonary circulation*. 2015;5(2):228-43. doi:10.1086/680213.

552. Evans AM, Lewis SA, Ogunbayo OA, Moral-Sanz J. Modulation of the LKB1-AMPK Signalling Pathway Underpins Hypoxic Pulmonary Vasoconstriction and Pulmonary Hypertension. *Advances in experimental medicine and biology*. 2015;860:89-99. doi:10.1007/978-3-319-18440-1_11.

553. Kholdani C, Fares WH, Mohsenin V. Pulmonary hypertension in obstructive sleep apnea: is it clinically significant? A critical analysis of the association and pathophysiology. *Pulmonary circulation*. 2015;5(2):220-7. doi:10.1086/679995.

554. Niermeyer S, Andrade MM, Vargas E, Moore LG. Neonatal oxygenation, pulmonary hypertension, and evolutionary adaptation to high altitude (2013 Grover Conference series). *Pulmonary circulation*. 2015;5(1):48-62. doi:10.1086/679719.

555. Lai N, Lu W, Wang J. Ca(2+) and ion channels in hypoxia-mediated pulmonary hypertension. *International journal of clinical and experimental pathology*. 2015;8(2):1081-92.

556. Stenmark KR, Tudor RM, El Kasmi KC. Metabolic reprogramming and inflammation act in concert to control vascular remodeling in hypoxic pulmonary hypertension. *Journal of applied physiology*. 2015;119(10):1164-72. doi:10.1152/jappphysiol.00283.2015.

557. Veit F, Pak O, Brandes RP, Weissmann N. Hypoxia-dependent reactive oxygen species signaling in the pulmonary circulation: focus on ion channels. *Antioxidants & redox signaling*. 2015;22(6):537-52. doi:10.1089/ars.2014.6234.

558. Pugliese SC, Poth JM, Fini MA, Olschewski A, El Kasmi KC, Stenmark KR. The role of inflammation in hypoxic pulmonary hypertension: from cellular mechanisms to clinical phenotypes. *American journal of physiology Lung cellular and molecular physiology*. 2015;308(3):L229-52. doi:10.1152/ajplung.00238.2014.

559. Jernigan NL. Smooth muscle acid-sensing ion channel 1: pathophysiological implication in hypoxic pulmonary hypertension. *Experimental physiology*. 2015;100(2):111-20. doi:10.1113/expphysiol.2014.081612.

560. Papamatheakis DG, Chundu M, Blood AB, Wilson SM. Prenatal programming of pulmonary hypertension induced by chronic hypoxia or ductal ligation in sheep. *Pulmonary circulation*. 2013;3(4):757-80. doi:10.1086/674767.

561. Robinson JC, Graham BB, Rouault TC, Tuder RM. The crossroads of iron with hypoxia and cellular metabolism. Implications in the pathobiology of pulmonary hypertension. *American journal of respiratory cell and molecular biology*. 2014;51(6):721-9. doi:10.1165/rcmb.2014-0021TR.
562. Shimoda LA, Laurie SS. HIF and pulmonary vascular responses to hypoxia. *Journal of applied physiology*. 2014;116(7):867-74. doi:10.1152/jappphysiol.00643.2013.
563. Nathan SD, Hassoun PM. Pulmonary hypertension due to lung disease and/or hypoxia. *Clinics in chest medicine*. 2013;34(4):695-705. doi:10.1016/j.ccm.2013.08.004.
564. Papamatheakis DG, Blood AB, Kim JH, Wilson SM. Antenatal hypoxia and pulmonary vascular function and remodeling. *Current vascular pharmacology*. 2013;11(5):616-40.
565. Caminati A, Cassandro R, Harari S. Pulmonary hypertension in chronic interstitial lung diseases. *European respiratory review : an official journal of the European Respiratory Society*. 2013;22(129):292-301. doi:10.1183/09059180.00002713.
566. Semenza GL. Oxygen sensing, hypoxia-inducible factors, and disease pathophysiology. *Annual review of pathology*. 2014;9:47-71. doi:10.1146/annurev-pathol-012513-104720.
567. Aggarwal S, Gross CM, Sharma S, Fineman JR, Black SM. Reactive oxygen species in pulmonary vascular remodeling. *Comprehensive Physiology*. 2013;3(3):1011-34. doi:10.1002/cphy.c120024.
568. Welsh DJ, Peacock AJ. Cellular responses to hypoxia in the pulmonary circulation. *High altitude medicine & biology*. 2013;14(2):111-6. doi:10.1089/ham.2013.1016.
569. Swenson ER. Hypoxic pulmonary vasoconstriction. *High altitude medicine & biology*. 2013;14(2):101-10. doi:10.1089/ham.2013.1010.
570. Lim CS, Kiriakidis S, Sandison A, Paleolog EM, Davies AH. Hypoxia-inducible factor pathway and diseases of the vascular wall. *Journal of vascular surgery*. 2013;58(1):219-30. doi:10.1016/j.jvs.2013.02.240.
571. Naeije R, Dedobbeleer C. Pulmonary hypertension and the right ventricle in hypoxia. *Experimental physiology*. 2013;98(8):1247-56. doi:10.1113/expphysiol.2012.069112.
572. Poor HD, Girgis R, Studer SM. World Health Organization Group III pulmonary hypertension. *Progress in cardiovascular diseases*. 2012;55(2):119-27. doi:10.1016/j.pcad.2012.08.003.
573. Neubauer JA, Sunderram J. Heme oxygenase-1 and chronic hypoxia. *Respiratory physiology & neurobiology*. 2012;184(2):178-85. doi:10.1016/j.resp.2012.06.027.
574. Zhu D, Ran Y. Role of 15-lipoxygenase/15-hydroxyeicosatetraenoic acid in hypoxia-induced pulmonary hypertension. *The journal of physiological sciences : JPS*. 2012;62(3):163-72. doi:10.1007/s12576-012-0196-9.
575. Sylvester JT, Shimoda LA, Aaronson PI, Ward JP. Hypoxic pulmonary vasoconstriction. *Physiological reviews*. 2012;92(1):367-520. doi:10.1152/physrev.00041.2010.
576. Maxova H, Herget J, Vizek M. Lung mast cells and hypoxic pulmonary hypertension. *Physiological research / Academia Scientiarum Bohemoslovaca*. 2012;61(1):1-11.
577. Kent BD, Mitchell PD, McNicholas WT. Hypoxemia in patients with COPD: cause, effects, and disease progression. *International journal of chronic obstructive pulmonary disease*. 2011;6:199-208. doi:10.2147/COPD.S10611.
578. Lapointe A, Barrington KJ. Pulmonary hypertension and the asphyxiated newborn. *The Journal of pediatrics*. 2011;158(2 Suppl):e19-24. doi:10.1016/j.jpeds.2010.11.008.
579. Shimoda LA, Semenza GL. HIF and the lung: role of hypoxia-inducible factors in pulmonary development and disease. *American journal of respiratory and critical care medicine*. 2011;183(2):152-6. doi:10.1164/rccm.201009-1393PP.
580. Gao Y, Raj JU. Hypoxic pulmonary hypertension of the newborn. *Comprehensive Physiology*. 2011;1(1):61-79. doi:10.1002/cphy.c090015.
581. Evans AM, Hardie DG, Peers C, Mahmoud A. Hypoxic pulmonary vasoconstriction: mechanisms of oxygen-sensing. *Current opinion in anaesthesiology*. 2011;24(1):13-20. doi:10.1097/ACO.0b013e3283421201.
582. Gorr TA, Wichmann D, Hu J, Hermes-Lima M, Welker AF, Terwilliger N et al. Hypoxia tolerance in animals: biology and application. *Physiological and biochemical zoology : PBZ*. 2010;83(5):733-52. doi:10.1086/648581.

583. Resta TC, Broughton BR, Jernigan NL. Reactive oxygen species and RhoA signaling in vascular smooth muscle: role in chronic hypoxia-induced pulmonary hypertension. *Advances in experimental medicine and biology*. 2010;661:355-73. doi:10.1007/978-1-60761-500-2_23.
584. Sakao S, Tatsumi K, Voelkel NF. Reversible or irreversible remodeling in pulmonary arterial hypertension. *American journal of respiratory cell and molecular biology*. 2010;43(6):629-34. doi:10.1165/rcmb.2009-0389TR.
585. Luks AM. Can patients with pulmonary hypertension travel to high altitude? *High altitude medicine & biology*. 2009;10(3):215-9. doi:10.1089/ham.2009.10306.
586. Ward JP, McMurtry IF. Mechanisms of hypoxic pulmonary vasoconstriction and their roles in pulmonary hypertension: new findings for an old problem. *Current opinion in pharmacology*. 2009;9(3):287-96. doi:10.1016/j.coph.2009.02.006.
587. Weir EK, Obrezhtchikova M, Vargese A, Cabrera JA, Peterson DA, Hong Z. Mechanisms of oxygen sensing: a key to therapy of pulmonary hypertension and patent ductus arteriosus. *British journal of pharmacology*. 2008;155(3):300-7. doi:10.1038/bjp.2008.291.
588. Gupte SA, Wolin MS. Oxidant and redox signaling in vascular oxygen sensing: implications for systemic and pulmonary hypertension. *Antioxidants & redox signaling*. 2008;10(6):1137-52. doi:10.1089/ars.2007.1995.
589. Nozik-Grayck E, Stenmark KR. Role of reactive oxygen species in chronic hypoxia-induced pulmonary hypertension and vascular remodeling. *Advances in experimental medicine and biology*. 2007;618:101-12.
590. Archer SL, Gomberg-Maitland M, Maitland ML, Rich S, Garcia JG, Weir EK. Mitochondrial metabolism, redox signaling, and fusion: a mitochondria-ROS-HIF-1 α -Kv1.5 O₂-sensing pathway at the intersection of pulmonary hypertension and cancer. *American journal of physiology Heart and circulatory physiology*. 2008;294(2):H570-8. doi:10.1152/ajpheart.01324.2007.
591. Pak O, Aldashev A, Welsh D, Peacock A. The effects of hypoxia on the cells of the pulmonary vasculature. *The European respiratory journal*. 2007;30(2):364-72. doi:10.1183/09031936.00128706.
592. Bonnet S, Archer SL. Potassium channel diversity in the pulmonary arteries and pulmonary veins: implications for regulation of the pulmonary vasculature in health and during pulmonary hypertension. *Pharmacology & therapeutics*. 2007;115(1):56-69. doi:10.1016/j.pharmthera.2007.03.014.
593. Preston IR. Clinical perspective of hypoxia-mediated pulmonary hypertension. *Antioxidants & redox signaling*. 2007;9(6):711-21. doi:10.1089/ars.2007.1587.
594. Hanze J, Weissmann N, Grimminger F, Seeger W, Rose F. Cellular and molecular mechanisms of hypoxia-inducible factor driven vascular remodeling. *Thrombosis and haemostasis*. 2007;97(5):774-87.
595. Esteve JM, Launay JM, Kellermann O, Maroteaux L. Functions of serotonin in hypoxic pulmonary vascular remodeling. *Cell biochemistry and biophysics*. 2007;47(1):33-44.
596. Penalzoza D, Arias-Stella J. The heart and pulmonary circulation at high altitudes: healthy highlanders and chronic mountain sickness. *Circulation*. 2007;115(9):1132-46. doi:10.1161/CIRCULATIONAHA.106.624544.
597. Shimoda LA, Wang J, Sylvester JT. Ca²⁺ channels and chronic hypoxia. *Microcirculation*. 2006;13(8):657-70. doi:10.1080/10739680600930305.
598. Moudgil R, Michelakis ED, Archer SL. The role of K⁺ channels in determining pulmonary vascular tone, oxygen sensing, cell proliferation, and apoptosis: implications in hypoxic pulmonary vasoconstriction and pulmonary arterial hypertension. *Microcirculation*. 2006;13(8):615-32. doi:10.1080/10739680600930222.
599. Stenmark KR, Fagan KA, Frid MG. Hypoxia-induced pulmonary vascular remodeling: cellular and molecular mechanisms. *Circulation research*. 2006;99(7):675-91. doi:10.1161/01.RES.0000243584.45145.3f.
600. Aaronson PI. TRPC Channel upregulation in chronically hypoxic pulmonary arteries: the HIF-1 bandwagon gathers steam. *Circulation research*. 2006;98(12):1465-7. doi:10.1161/01.RES.0000231254.58548.b4.

601. Ghofrani HA, Voswinckel R, Reichenberger F, Weissmann N, Schermuly RT, Seeger W et al. Hypoxia- and non-hypoxia-related pulmonary hypertension - established and new therapies. *Cardiovascular research*. 2006;72(1):30-40. doi:10.1016/j.cardiores.2006.07.025.
602. Weir EK, Olschewski A. Role of ion channels in acute and chronic responses of the pulmonary vasculature to hypoxia. *Cardiovascular research*. 2006;71(4):630-41. doi:10.1016/j.cardiores.2006.04.014.
603. Weissmann N, Sommer N, Schermuly RT, Ghofrani HA, Seeger W, Grimminger F. Oxygen sensors in hypoxic pulmonary vasoconstriction. *Cardiovascular research*. 2006;71(4):620-9. doi:10.1016/j.cardiores.2006.04.009.
604. Morrell ED, Tsai BM, Crisostomo PR, Hammoud ZT, Meldrum DR. Experimental therapies for hypoxia-induced pulmonary hypertension during acute lung injury. *Shock*. 2006;25(3):214-26. doi:10.1097/01.shk.0000191380.44972.46.
605. Semenza GL. Involvement of hypoxia-inducible factor 1 in pulmonary pathophysiology. *Chest*. 2005;128(6 Suppl):592S-4S. doi:10.1378/chest.128.6_suppl.592S.
606. Derchi G, Forni GL. Therapeutic approaches to pulmonary hypertension in hemoglobinopathies: Efficacy and safety of sildenafil in the treatment of severe pulmonary hypertension in patients with hemoglobinopathy. *Annals of the New York Academy of Sciences*. 2005;1054:471-5. doi:10.1196/annals.1345.070.
607. Remillard CV, Yuan JX. High altitude pulmonary hypertension: role of K⁺ and Ca²⁺ channels. *High altitude medicine & biology*. 2005;6(2):133-46. doi:10.1089/ham.2005.6.133.
608. Rhodes J. Comparative physiology of hypoxic pulmonary hypertension: historical clues from brisket disease. *Journal of applied physiology*. 2005;98(3):1092-100. doi:10.1152/jappphysiol.01017.2004.
609. Zielinski J. Effects of intermittent hypoxia on pulmonary haemodynamics: animal models versus studies in humans. *The European respiratory journal*. 2005;25(1):173-80. doi:10.1183/09031936.04.00037204.
610. Howell K, Ooi H, Preston R, McLoughlin P. Structural basis of hypoxic pulmonary hypertension: the modifying effect of chronic hypercapnia. *Experimental physiology*. 2004;89(1):66-72.
611. Maggiorini M. Cardio-pulmonary interactions at high altitude. Pulmonary hypertension as a common denominator. *Advances in experimental medicine and biology*. 2003;543:177-89.
612. McMurtry IF, Bauer NR, Fagan KA, Nagaoka T, Gebb SA, Oka M. Hypoxia and Rho/Rho-kinase signaling. Lung development versus hypoxic pulmonary hypertension. *Advances in experimental medicine and biology*. 2003;543:127-37.
613. Novotna J, Herget J. Possible role of matrix metalloproteinases in reconstruction of peripheral pulmonary arteries induced by hypoxia. *Physiological research / Academia Scientiarum Bohemoslovaca*. 2002;51(4):323-34.
614. Tucker A, Rhodes J. Role of vascular smooth muscle in the development of high altitude pulmonary hypertension: an interspecies evaluation. *High altitude medicine & biology*. 2001;2(2):173-89. doi:10.1089/152702901750265288.
615. Neubauer JA. Invited review: Physiological and pathophysiological responses to intermittent hypoxia. *Journal of applied physiology*. 2001;90(4):1593-9.
616. Aldashev AA. High-altitude pulmonary hypertension and signal transduction in the cardiovascular system. *Journal of receptor and signal transduction research*. 2000;20(4):255-78. doi:10.3109/10799890009150647.
617. Stenmark KR, Bouchez D, Nemenoff R, Dempsey EC, Das M. Hypoxia-induced pulmonary vascular remodeling: contribution of the adventitial fibroblasts. *Physiological research / Academia Scientiarum Bohemoslovaca*. 2000;49(5):503-17.
618. Herget J, Wilhelm J, Novotna J, Eckhardt A, Vytasek R, Mrazkova L et al. A possible role of the oxidant tissue injury in the development of hypoxic pulmonary hypertension. *Physiological research / Academia Scientiarum Bohemoslovaca*. 2000;49(5):493-501.
619. Pierson DJ. Pathophysiology and clinical effects of chronic hypoxia. *Respiratory care*. 2000;45(1):39-51; discussion -3.

620. Stenmark KR, Frid M, Nemenoff R, Dempsey EC, Das M. Hypoxia induces cell-specific changes in gene expression in vascular wall cells: implications for pulmonary hypertension. *Advances in experimental medicine and biology*. 1999;474:231-58.
621. Scherrer U, Sartori C, Lepori M, Allemann Y, Duplain H, Trueb L et al. High-altitude pulmonary edema: from exaggerated pulmonary hypertension to a defect in transepithelial sodium transport. *Advances in experimental medicine and biology*. 1999;474:93-107.
622. Egermayer P, Town GI, Peacock AJ. Role of serotonin in the pathogenesis of acute and chronic pulmonary hypertension. *Thorax*. 1999;54(2):161-8.
623. Karamsetty VS, Kane KA, Wadsworth RM. The effects of chronic hypoxia on the pharmacological responsiveness of the pulmonary artery. *Pharmacology & therapeutics*. 1995;68(2):233-46.
624. Kourembanas S, Bernfield M. Hypoxia and endothelial-smooth muscle cell interactions in the lung. *American journal of respiratory cell and molecular biology*. 1994;11(4):373-4. doi:10.1165/ajrcmb.11.4.7917305.
625. Vender RL. Chronic hypoxic pulmonary hypertension. *Cell biology to pathophysiology*. *Chest*. 1994;106(1):236-43.
626. Higenbottam T, Cremona G. Acute and chronic hypoxic pulmonary hypertension. *The European respiratory journal*. 1993;6(8):1207-12.
627. Stenmark KR, Aldashev AA, Orton EC, Durmowicz AG, Badesch DB, Parks WC et al. Cellular adaptation during chronic neonatal hypoxic pulmonary hypertension. *The American journal of physiology*. 1991;261(4 Suppl):97-104.
628. Cremona G, Dinh Xuan AT, Higenbottam TW. Endothelium-derived relaxing factor and the pulmonary circulation. *Lung*. 1991;169(4):185-202.
629. Peacock A. Pulmonary hypertension due to chronic hypoxia. *Bmj*. 1990;300(6727):763.
630. Meyrick BO, Perrett EA. The sequence of cellular and hemodynamic changes of chronic pulmonary hypertension induced by hypoxia and other stimuli. *The American review of respiratory disease*. 1989;140(5):1486-9. doi:10.1164/ajrccm/140.5.1486.
631. Barer GR. Hypoxia and the pulmonary circulation: a brief review. *Zeitschrift fur Erkrankungen der Atmungsorgane*. 1989;173(2):109-15.
632. Suggett AJ, Barer GR. Experimental prevention of hypoxic pulmonary hypertension in animals by drugs. *European heart journal*. 1988;9 Suppl J:13-8.
633. Herget J. Mechanism of hypoxic pulmonary hypertension. *Pneumonologia polska*. 1988;56(3):89-93.
634. Flenley DC, Muir AL. Cardiovascular effects of oxygen therapy for pulmonary arterial hypertension. *Clinics in chest medicine*. 1983;4(2):297-308.
635. Lockhart A, Saiag B. Altitude and the human pulmonary circulation. *Clin Sci (Lond)*. 1981;60(6):599-605.
636. Ostadal B, Urbanova D, Ressler J, Prochazka J, Pelouch V, Widimsky J. Changes of the right and left ventricles in rats exposed to intermittent high altitude hypoxia. *Cor et vasa*. 1981;23(2):111-20.
637. Herget J, Palecek F. Experimental chronic pulmonary hypertension. *International review of experimental pathology*. 1978;18:347-406.
638. Heath D. Hypoxia and the pulmonary circulation. *Journal of clinical pathology Supplement*. 1977;11:21-9.
639. Harris P, Barrie E, Gibson K, Gloster J. Effects of chronic hypoxia on myocardial metabolism. *Recent advances in studies on cardiac structure and metabolism*. 1975;8:265-70.
640. Burrows B. Arterial oxygenation and pulmonary hemodynamics in patients with chronic airways obstruction. *The American review of respiratory disease*. 1974;110(6 Pt 2):64-70. doi:10.1164/arrd.1974.110.6P2.64.
641. Reeves JT. Pulmonary vascular response to high altitude residence. *Cardiovascular clinics*. 1973;5(1):81-95.
642. Hultgren HN, Grover RF. Circulatory adaptation to high altitude. *Annual review of medicine*. 1968;19:119-52. doi:10.1146/annurev.me.19.020168.001003.

643. Hussain A, Suleiman MS, George SJ, Loubani M, Morice A. Hypoxic Pulmonary Vasoconstriction in Humans: Tale or Myth. *Open Cardiovasc Med J.* 2017;11:1-13. doi:10.2174/1874192401711010001.
644. Dai Z, Zhao YY. Discovery of a murine model of clinical PAH: Mission impossible? *Trends in cardiovascular medicine.* 2016. doi:10.1016/j.tcm.2016.12.003.
645. Dunham-Snary KJ, Wu D, Sykes EA, Thakrar A, Parlow LR, Mewburn JD et al. Hypoxic Pulmonary Vasoconstriction: From Molecular Mechanisms to Medicine. *Chest.* 2017;151(1):181-92. doi:10.1016/j.chest.2016.09.001.
646. Kylhammar D, Radegran G. The principal pathways involved in the in vivo modulation of hypoxic pulmonary vasoconstriction, pulmonary arterial remodelling and pulmonary hypertension. *Acta Physiol (Oxf).* 2017;219(4):728-56. doi:10.1111/apha.12749.
647. Mohsenin V. The emerging role of microRNAs in hypoxia-induced pulmonary hypertension. *Sleep & breathing = Schlaf & Atmung.* 2016;20(3):1059-67. doi:10.1007/s11325-016-1351-y.
648. Rowan SC, Keane MP, Gaine S, McLoughlin P. Hypoxic pulmonary hypertension in chronic lung diseases: novel vasoconstrictor pathways. *Lancet Respir Med.* 2016;4(3):225-36. doi:10.1016/S2213-2600(15)00517-2.
649. Montani D, Seferian A, Savale L, Simonneau G, Humbert M. Drug-induced pulmonary arterial hypertension: a recent outbreak. *European respiratory review : an official journal of the European Respiratory Society.* 2013;22(129):244-50. doi:10.1183/09059180.00003313.
650. Seferian A, Chaumais MC, Savale L, Gunther S, Tubert-Bitter P, Humbert M et al. Drugs induced pulmonary arterial hypertension. *Presse medicale.* 2013;42(9 Pt 2):e303-10. doi:10.1016/j.lpm.2013.07.005.
651. de Jesus Perez V, Kudelko K, Snook S, Zamanian RT. Drugs and toxins-associated pulmonary arterial hypertension: lessons learned and challenges ahead. *International journal of clinical practice Supplement.* 2011(169):8-10. doi:10.1111/j.1742-1241.2010.02606.x.
652. Abenheim L, Humbert M. Pulmonary hypertension related to drugs and toxins. *Current opinion in cardiology.* 1999;14(5):437-41.
653. Egermayer P. Epidemics of vascular toxicity and pulmonary hypertension: what can be learned? *Journal of internal medicine.* 2000;247(1):11-7.
654. Szymanski C, Andrejak M, Peltier M, Marechaux S, Tribouilloy C. Adverse effects of benfluorex on heart valves and pulmonary circulation. *Pharmacoepidemiology and drug safety.* 2014;23(7):679-86. doi:10.1002/pds.3642.
655. Leung YY, Tang KS, Tsang CC, Chan CK, Wong KK, Yu AW. Pulmonary hypertension, hyperthyroidism, and fenfluramine: a case report and review. *MedGenMed : Medscape general medicine.* 2006;8(4):29.
656. Tellier P. Fenfluramines, idiopathic pulmonary primary hypertension and cardiac valve disorders: facts and artifacts. *Annales de medecine interne.* 2001;152(7):429-36.
657. Eddahibi S, Adnot S. Anorexigen-induced pulmonary hypertension and the serotonin (5-HT) hypothesis: lessons for the future in pathogenesis. *Respiratory research.* 2002;3:9.
658. Humbert M, Nunes H, Sitbon O, Parent F, Herve P, Simonneau G. Risk factors for pulmonary arterial hypertension. *Clinics in chest medicine.* 2001;22(3):459-75.
659. Michelakis ED, Weir EK. Anorectic drugs and pulmonary hypertension from the bedside to the bench. *The American journal of the medical sciences.* 2001;321(4):292-9.
660. MacLean MR. Pulmonary hypertension, anorexigens and 5-HT: pharmacological synergism in action? *Trends in pharmacological sciences.* 1999;20(12):490-5.
661. Simonneau G, Fartoukh M, Sitbon O, Humbert M, Jagot JL, Herve P. Primary pulmonary hypertension associated with the use of fenfluramine derivatives. *Chest.* 1998;114(3 Suppl):195S-9S.
662. McCann UD, Seiden LS, Rubin LJ, Ricaurte GA. Brain serotonin neurotoxicity and primary pulmonary hypertension from fenfluramine and dexfenfluramine. A systematic review of the evidence. *Jama.* 1997;278(8):666-72.

663. Ranchoux B, Gunther S, Quarck R, Chaumais MC, Dorfmuller P, Antigny F et al. Chemotherapy-induced pulmonary hypertension: role of alkylating agents. *The American journal of pathology*. 2015;185(2):356-71. doi:10.1016/j.ajpath.2014.10.021.
664. Shah NP, Wallis N, Farber HW, Mauro MJ, Wolf RA, Mattei D et al. Clinical features of pulmonary arterial hypertension in patients receiving dasatinib. *American journal of hematology*. 2015;90(11):1060-4. doi:10.1002/ajh.24174.
665. Godinas L, Guignabert C, Seferian A, Perros F, Bergot E, Sibille Y et al. Tyrosine kinase inhibitors in pulmonary arterial hypertension: a double-edge sword? *Seminars in respiratory and critical care medicine*. 2013;34(5):714-24. doi:10.1055/s-0033-1356494.
666. Galbally M, Gentile S, Lewis AJ. Further findings linking SSRIs during pregnancy and persistent pulmonary hypertension of the newborn: clinical implications. *CNS drugs*. 2012;26(10):813-22. doi:10.2165/11630310-000000000-00000.
667. t Jong GW, Einarson T, Koren G, Einarson A. Antidepressant use in pregnancy and persistent pulmonary hypertension of the newborn (PPHN): a systematic review. *Reproductive toxicology*. 2012;34(3):293-7. doi:10.1016/j.reprotox.2012.04.015.
668. Koren G, Nordeng H. Antidepressant use during pregnancy: the benefit-risk ratio. *American journal of obstetrics and gynecology*. 2012;207(3):157-63. doi:10.1016/j.ajog.2012.02.009.
669. Occhiogrosso M, Omran SS, Altemus M. Persistent pulmonary hypertension of the newborn and selective serotonin reuptake inhibitors: lessons from clinical and translational studies. *The American journal of psychiatry*. 2012;169(2):134-40. doi:10.1176/appi.ajp.2011.11040553.
670. Galbally M, Snellen M, Lewis AJ. A review of the use of psychotropic medication in pregnancy. *Current opinion in obstetrics & gynecology*. 2011;23(6):408-14. doi:10.1097/GCO.0b013e32834b92f3.
671. Hsu CH, Gomberg-Maitland M, Glassner C, Chen JH. The management of pregnancy and pregnancy-related medical conditions in pulmonary arterial hypertension patients. *International journal of clinical practice Supplement*. 2011(172):6-14. doi:10.1111/j.1742-1241.2011.02711.x.
672. Udechuku A, Nguyen T, Hill R, Szego K. Antidepressants in pregnancy: a systematic review. *The Australian and New Zealand journal of psychiatry*. 2010;44(11):978-96. doi:10.3109/00048674.2010.507543.
673. Bedard E, Dimopoulos K, Gatzoulis MA. Has there been any progress made on pregnancy outcomes among women with pulmonary arterial hypertension? *European heart journal*. 2009;30(3):256-65. doi:10.1093/eurheartj/ehn597.
674. Nagasaki J, Aoyama Y, Nomoto Y, Ido K, Ichihara H, Mugitani A. Reversible dasatinib-related pulmonary arterial hypertension in a CML patient. *Rinsho Ketsueki*. 2016;57(5):618-23. doi:10.11406/rinketsu.57.618.
675. Gunther S, Behr J, Knoop H. [Drug-induced Pulmonary Hypertension - a Current Review]. *Pneumologie*. 2016;70(5):320-7. doi:10.1055/s-0041-108315.
676. Grunig G, Marsh LM, Esmail N, Jackson K, Gordon T, Reibman J et al. Perspective: ambient air pollution: inflammatory response and effects on the lung's vasculature. *Pulmonary circulation*. 2014;4(1):25-35. doi:10.1086/674902.
677. Babu AS, Padmakumar R, Maiya AG, Mohapatra AK, Kamath RL. Effects of Exercise Training on Exercise Capacity in Pulmonary Arterial Hypertension: A Systematic Review of Clinical Trials. *Heart, lung & circulation*. 2015. doi:10.1016/j.hlc.2015.10.015.
678. Arena R, Lavie CJ, Borghi-Silva A, Daugherty J, Bond S, Phillips SA et al. Exercise Training in Group 2 Pulmonary Hypertension: Which Intensity and What Modality. *Progress in cardiovascular diseases*. 2015. doi:10.1016/j.pcad.2015.11.005.
679. Sahni S, Capozzi B, Iftikhar A, Sgouras V, Ojrzanowski M, Talwar A. Pulmonary rehabilitation and exercise in pulmonary arterial hypertension: An underutilized intervention. *Journal of exercise rehabilitation*. 2015;11(2):74-9. doi:10.12965/jer.150190.
680. Arena R, Cahalin LP, Borghi-Silva A, Myers J. The effect of exercise training on the pulmonary arterial system in patients with pulmonary hypertension. *Progress in cardiovascular diseases*. 2015;57(5):480-8. doi:10.1016/j.pcad.2014.03.008.

681. Marra AM, Egenlauf B, Bossone E, Eichstaedt C, Grunig E, Ehlken N. Principles of rehabilitation and reactivation: pulmonary hypertension. *Respiration; international review of thoracic diseases.* 2015;89(4):265-73. doi:10.1159/000371855.
682. Yuan P, Yuan XT, Sun XY, Pudasaini B, Liu JM, Hu QH. Exercise training for pulmonary hypertension: a systematic review and meta-analysis. *International journal of cardiology.* 2015;178:142-6. doi:10.1016/j.ijcard.2014.10.161.
683. Kim CH, Jae SY, Johnson BD. Pulmonary Hypertension and Cardiopulmonary Exercise in Heart Failure. *Pulse.* 2014;1(3-4):143-51. doi:10.1159/000360964.
684. Zafir B. Exercise training and rehabilitation in pulmonary arterial hypertension: rationale and current data evaluation. *Journal of cardiopulmonary rehabilitation and prevention.* 2013;33(5):263-73. doi:10.1097/HCR.0b013e3182a0299a.
685. Babu AS, Padmakumar R, Maiya AG. A review of ongoing trials in exercise based rehabilitation for pulmonary arterial hypertension. *The Indian journal of medical research.* 2013;137(5):900-6.
686. Babu AS, Myers J, Arena R, Maiya AG, Padmakumar R. Evaluating exercise capacity in patients with pulmonary arterial hypertension. *Expert review of cardiovascular therapy.* 2013;11(6):729-37. doi:10.1586/erc.13.33.
687. Naeije R, Vanderpool R, Dhakal BP, Saggat R, Saggat R, Vachiery JL et al. Exercise-induced pulmonary hypertension: physiological basis and methodological concerns. *American journal of respiratory and critical care medicine.* 2013;187(6):576-83. doi:10.1164/rccm.201211-2090CI.
688. Paolillo S, Farina S, Bussotti M, Iorio A, PerroneFilardi P, Piepolil MF et al. Exercise testing in the clinical management of patients affected by pulmonary arterial hypertension. *European journal of preventive cardiology.* 2012;19(5):960-71.
689. Naeije R, Chesler N. Pulmonary circulation at exercise. *Comprehensive Physiology.* 2012;2(1):711-41. doi:10.1002/cphy.c100091.
690. Waxman AB. Exercise physiology and pulmonary arterial hypertension. *Progress in cardiovascular diseases.* 2012;55(2):172-9. doi:10.1016/j.pcad.2012.07.003.
691. Bossone E, Naeije R. Exercise-induced pulmonary hypertension. *Heart failure clinics.* 2012;8(3):485-95. doi:10.1016/j.hfc.2012.04.007.
692. Arena R. Exercise testing and training in chronic lung disease and pulmonary arterial hypertension. *Progress in cardiovascular diseases.* 2011;53(6):454-63. doi:10.1016/j.pcad.2011.02.003.
693. Khajali F, Wideman RF. Nutritional approaches to ameliorate pulmonary hypertension in broiler chickens. *Journal of animal physiology and animal nutrition.* 2016;100(1):3-14. doi:10.1111/jpn.12315.
694. Sahni S, Palkar AV, Rochelson BL, Kepa W, Talwar A. Pregnancy and pulmonary arterial hypertension: A clinical conundrum. *Pregnancy hypertension.* 2015;5(2):157-64. doi:10.1016/j.preghy.2015.01.004.
695. Gei A, Montufar-Rueda C. Pulmonary hypertension and pregnancy: an overview. *Clinical obstetrics and gynecology.* 2014;57(4):806-26. doi:10.1097/GRF.0000000000000076.
696. Obican SG, Cleary KL. Pulmonary arterial hypertension in pregnancy. *Seminars in perinatology.* 2014;38(5):289-94. doi:10.1053/j.semperi.2014.04.018.
697. Pieper PG, Lameijer H, Hoendermis ES. Pregnancy and pulmonary hypertension. *Best practice & research Clinical obstetrics & gynaecology.* 2014;28(4):579-91. doi:10.1016/j.bpobgyn.2014.03.003.
698. Olsson KM, Jais X. Birth control and pregnancy management in pulmonary hypertension. *Seminars in respiratory and critical care medicine.* 2013;34(5):681-8. doi:10.1055/s-0033-1355438.
699. Van Marter LJ, Hernandez-Diaz S, Werler MM, Louik C, Mitchell AA. Nonsteroidal antiinflammatory drugs in late pregnancy and persistent pulmonary hypertension of the newborn. *Pediatrics.* 2013;131(1):79-87. doi:10.1542/peds.2012-0496.
700. Safdar Z. Pulmonary arterial hypertension in pregnant women. *Therapeutic advances in respiratory disease.* 2013;7(1):51-63. doi:10.1177/1753465812461680.
701. Martinez MV, Rutherford JD. Pulmonary hypertension in pregnancy. *Cardiology in review.* 2013;21(4):167-73. doi:10.1097/CRD.0b013e318275cf01.

702. Lane CR, Trow TK. Pregnancy and pulmonary hypertension. *Clinics in chest medicine*. 2011;32(1):165-74. x. doi:10.1016/j.ccm.2010.10.006.
703. Cotrim SC, Loureiro MJ, Avillez T, Simoes O, Cordeiro P, Almeida S et al. Three cases of pregnancy in patients with severe pulmonary arterial hypertension: experience of a single unit. *Revista portuguesa de cardiologia : orgao oficial da Sociedade Portuguesa de Cardiologia = Portuguese journal of cardiology : an official journal of the Portuguese Society of Cardiology*. 2010;29(1):95-103.
704. Sanchez O, Marie E, Lerolle U, Wermert D, Israel-Biet D, Meyer G. Pulmonary arterial hypertension in women. *Revue des maladies respiratoires*. 2010;27(8):e79-87. doi:10.1016/j.rmr.2009.12.001.
705. Higton AM, Whale C, Musk M, Gabbay E. Pulmonary hypertension in pregnancy: two cases and review of the literature. *Internal medicine journal*. 2009;39(11):766-70. doi:10.1111/j.1445-5994.2009.02051.x.
706. Madden BP. Pulmonary hypertension and pregnancy. *International journal of obstetric anaesthesia*. 2009;18(2):156-64. doi:10.1016/j.ijoa.2008.10.006.
707. Thorne S, Nelson-Piercy C, MacGregor A, Gibbs S, Crowhurst J, Panay N et al. Pregnancy and contraception in heart disease and pulmonary arterial hypertension. *The journal of family planning and reproductive health care / Faculty of Family Planning & Reproductive Health Care, Royal College of Obstetricians & Gynaecologists*. 2006;32(2):75-81. doi:10.1783/147118906776276486.
708. Warnes CA. Pregnancy and pulmonary hypertension. *International journal of cardiology*. 2004;97 Suppl 1:11-3. doi:10.1016/j.ijcard.2004.08.004.
709. Roberts NV, Keast PJ. Pulmonary hypertension and pregnancy--a lethal combination. *Anaesthesia and intensive care*. 1990;18(3):366-74.
710. McCaffrey RM, Dunn LJ. Primary Pulmonary Hypertension in Pregnancy. *Obstetrical & gynecological survey*. 1964;19:567-91.
711. Mocumbi AO, Thienemann F, Sliwa K. A global perspective on the epidemiology of pulmonary hypertension. *The Canadian journal of cardiology*. 2015;31(4):375-81. doi:10.1016/j.cjca.2015.01.030.
712. Bigna JJ, Sime PS, Koulla-Shiro S. HIV related pulmonary arterial hypertension: epidemiology in Africa, physiopathology, and role of antiretroviral treatment. *AIDS research and therapy*. 2015;12:36. doi:10.1186/s12981-015-0078-3.
713. Medarov BI, Jogani S, Sun J, Judson MA. Readdressing the entity of exercise pulmonary arterial hypertension. *Respiratory medicine*. 2017;124:65-71. doi:10.1016/j.rmed.2017.02.012.
714. Morris NR, Kermeen FD, Holland AE. Exercise-based rehabilitation programmes for pulmonary hypertension. *The Cochrane database of systematic reviews*. 2017;1:CD011285. doi:10.1002/14651858.CD011285.pub2.
715. Madonna R, De Caterina R, Geng YJ. Aerobic exercise-related attenuation of arterial pulmonary hypertension: A right arrow targets the disease? *Vascular pharmacology*. 2016;87:6-9. doi:10.1016/j.vph.2016.10.002.
716. Pinkstaff SO, Burger CD, Daugherty J, Bond S, Arena R. Cardiopulmonary exercise testing in patients with pulmonary hypertension: clinical recommendations based on a review of the evidence. *Expert review of respiratory medicine*. 2016;10(3):279-95. doi:10.1586/17476348.2016.1144475.
717. L'Huillier AG, Posfay-Barbe KM, Pictet H, Beghetti M. Pulmonary Arterial Hypertension among HIV-Infected Children: Results of a National Survey and Review of the Literature. *Frontiers in pediatrics*. 2015;3:25. doi:10.3389/fped.2015.00025.
718. Butrous G. Human immunodeficiency virus-associated pulmonary arterial hypertension: considerations for pulmonary vascular diseases in the developing world. *Circulation*. 2015;131(15):1361-70. doi:10.1161/CIRCULATIONAHA.114.006978.
719. Correale M, Palmiotti GA, Lo Storto MM, Montrone D, Foschino Barbaro MP, Di Biase M et al. HIV-associated pulmonary arterial hypertension: from bedside to the future. *European journal of clinical investigation*. 2015;45(5):515-28. doi:10.1111/eci.12427.

720. Graham BB, Kumar R. Schistosomiasis and the pulmonary vasculature (2013 Grover Conference series). *Pulmonary circulation*. 2014;4(3):353-62. doi:10.1086/675983.
721. Dai HL, Zhang M, Xiao ZC, Guang XF, Yin XL. Pulmonary arterial hypertension in HIV infection: a concise review. *Heart, lung & circulation*. 2014;23(4):299-302. doi:10.1016/j.hlc.2013.10.088.
722. Taichman DB, Mandel J. Epidemiology of pulmonary arterial hypertension. *Clinics in chest medicine*. 2013;34(4):619-37. doi:10.1016/j.ccm.2013.08.010.
723. Mirrakhimov AE, Ali AM, Barbaryan A, Prueksaritanond S. Human immunodeficiency virus and pulmonary arterial hypertension. *ISRN cardiology*. 2013;2013:903454. doi:10.1155/2013/903454.
724. Barnett CF, Hsue PY. Human immunodeficiency virus-associated pulmonary arterial hypertension. *Clinics in chest medicine*. 2013;34(2):283-92. doi:10.1016/j.ccm.2013.01.009.
725. Cicalini S, Chinello P, Petrosillo N. HIV infection and pulmonary arterial hypertension. *Expert review of respiratory medicine*. 2011;5(2):257-66. doi:10.1586/ers.11.10.
726. Cool CD, Voelkel NF, Bull T. Viral infection and pulmonary hypertension: is there an association? *Expert review of respiratory medicine*. 2011;5(2):207-16. doi:10.1586/ers.11.17.
727. Cicalini S, Almodovar S, Grilli E, Flores S. Pulmonary hypertension and human immunodeficiency virus infection: epidemiology, pathogenesis, and clinical approach. *Clinical microbiology and infection : the official publication of the European Society of Clinical Microbiology and Infectious Diseases*. 2011;17(1):25-33. doi:10.1111/j.1469-0691.2010.03286.x.
728. Janda S, Quon BS, Swiston J. HIV and pulmonary arterial hypertension: a systematic review. *HIV medicine*. 2010;11(10):620-34. doi:10.1111/j.1468-1293.2010.00829.x.
729. Derchi G, Lai ME, Marcaccini P, Carta MP, Vacquer S. Human immunodeficiency virus and beta-thalassemia major: A "competition of guilt" for pulmonary arterial hypertension. Report of a case and a review of the literature. *Hemoglobin*. 2010;34(1):61-6. doi:10.3109/03630260903547765.
730. Cicalini S, Chinello P, Grilli E, Petrosillo N. Treatment and outcome of pulmonary arterial hypertension in HIV-infected patients: a review of the literature. *Current HIV research*. 2009;7(6):589-96.
731. Talwar A, Sarkar P, Rosen MJ. Pulmonary arterial hypertension in human immunodeficiency virus infection. *Postgraduate medicine*. 2009;121(5):56-67. doi:10.3810/pgm.2009.09.2053.
732. Degano B, Sitbon O, Simonneau G. Pulmonary arterial hypertension and HIV infection. *Seminars in respiratory and critical care medicine*. 2009;30(4):440-7. doi:10.1055/s-0029-1233313.
733. Murphy D, Girgis RE. HIV-associated pulmonary arterial hypertension: a clinical problem that is here to stay? *International journal of clinical practice Supplement*. 2009(161):19-21. doi:10.1111/j.1742-1241.2008.01959.x.
734. Sitbon O. HIV-related pulmonary arterial hypertension: clinical presentation and management. *Aids*. 2008;22 Suppl 3:S55-62. doi:10.1097/01.aids.0000327517.62665.ec.
735. Voelkel NF, Cool CD, Flores S. From viral infection to pulmonary arterial hypertension: a role for viral proteins? *Aids*. 2008;22 Suppl 3:S49-53. doi:10.1097/01.aids.0000327516.55041.01.
736. Humbert M. Mediators involved in HIV-related pulmonary arterial hypertension. *Aids*. 2008;22 Suppl 3:S41-7. doi:10.1097/01.aids.0000327515.55041.da.
737. Lederman MM, Sereni D, Simonneau G, Voelkel NF. Pulmonary arterial hypertension and its association with HIV infection: an overview. *Aids*. 2008;22 Suppl 3:S1-6. doi:10.1097/01.aids.0000327509.30385.3b.
738. Barnett CF, Hsue PY, Machado RF. Pulmonary hypertension: an increasingly recognized complication of hereditary hemolytic anemias and HIV infection. *Jama*. 2008;299(3):324-31. doi:10.1001/jama.299.3.324.
739. Limsukon A, Saeed AI, Ramasamy V, Nalamati J, Dhuper S. HIV-related pulmonary hypertension. *The Mount Sinai journal of medicine, New York*. 2006;73(7):1037-44.
740. Grubb JR, Moorman AC, Baker RK, Masur H. The changing spectrum of pulmonary disease in patients with HIV infection on antiretroviral therapy. *Aids*. 2006;20(8):1095-107. doi:10.1097/01.aids.0000226949.64600.f9.

741. Pellicelli AM, D'Ambrosio C, Vizza CD, Borgia MC, Tanzi P, Pino P et al. HIV-related pulmonary hypertension. From pathogenesis to clinical aspects. *Acta cardiologica*. 2004;59(3):323-30. doi:10.2143/AC.59.3.2005190.
742. Burkart KM, Farber HW. HIV-associated pulmonary hypertension: diagnosis and treatment. *Advances in cardiology*. 2003;40:197-207.
743. Klings ES, Farber HW. The pathogenesis of HIV-associated pulmonary hypertension. *Advances in cardiology*. 2003;40:71-82.
744. Pellicelli AM, Palmieri F, Cicalini S, Petrosillo N. Pathogenesis of HIV-related pulmonary hypertension. *Annals of the New York Academy of Sciences*. 2001;946:82-94.
745. Petrosillo N, Pellicelli AM, Boumis E, Ippolito G. Clinical manifestation of HIV-related pulmonary hypertension. *Annals of the New York Academy of Sciences*. 2001;946:223-35.
746. Seoane L, Shellito J, Welsh D, de Boisblanc BP. Pulmonary hypertension associated with HIV infection. *Southern medical journal*. 2001;94(6):635-9.
747. Pellicelli AM, Barbaro G, Palmieri F, Girardi E, D'Ambrosio C, Rianda A et al. Primary pulmonary hypertension in HIV patients: a systematic review. *Angiology*. 2001;52(1):31-41.
748. Mehta NJ, Khan IA, Mehta RN, Sepkowitz DA. HIV-Related pulmonary hypertension: analytic review of 131 cases. *Chest*. 2000;118(4):1133-41.
749. Fiorencis R, Zoncin P, Carraro M, Zampieri P, Roncon L, Baracca E et al. Pulmonary hypertension associated with human immunodeficiency virus infection. Report of two cases and review of the literature. *Giornale italiano di cardiologia*. 1998;28(12):1404-8.
750. Golpe R, Fernandez-Infante B, Fernandez-Rozas S. Primary pulmonary hypertension associated with human immunodeficiency virus infection. *Postgraduate medical journal*. 1998;74(873):400-4.
751. Mesa RA, Edell ES, Dunn WF, Edwards WD. Human immunodeficiency virus infection and pulmonary hypertension: two new cases and a review of 86 reported cases. *Mayo Clinic proceedings*. 1998;73(1):37-45. doi:10.1016/S0025-6196(11)63616-1.
752. Weiss JR, Pietra GG, Scharf SM. Primary pulmonary hypertension and the human immunodeficiency virus. Report of two cases and a review of the literature. *Archives of internal medicine*. 1995;155(21):2350-4.
753. de Chadarevian JP, Lischner HW, Karmazin N, Pawel BR, Schultz TE. Pulmonary hypertension and HIV infection: new observations and review of the syndrome. *Modern pathology : an official journal of the United States and Canadian Academy of Pathology, Inc*. 1994;7(6):685-9.
754. Mani S, Smith GJ. HIV and pulmonary hypertension: a review. *Southern medical journal*. 1994;87(3):357-62.
755. Polos PG, Wolfe D, Harley RA, Strange C, Sahn SA. Pulmonary hypertension and human immunodeficiency virus infection. Two reports and a review of the literature. *Chest*. 1992;101(2):474-8.
756. Voelkel NF, Cool C, Taraseviciene-Stewart L, Geraci MW, Yeager M, Bull T et al. Janus face of vascular endothelial growth factor: the obligatory survival factor for lung vascular endothelium controls precapillary artery remodeling in severe pulmonary hypertension. *Critical care medicine*. 2002;30(5 Suppl):S251-6.
757. Tuder RM, Cool CD, Yeager M, Taraseviciene-Stewart L, Bull TM, Voelkel NF. The pathobiology of pulmonary hypertension. *Endothelium. Clinics in chest medicine*. 2001;22(3):405-18.
758. Giannakoulas G, Mouratoglou SA, Gatzoulis MA, Karvounis H. Blood biomarkers and their potential role in pulmonary arterial hypertension associated with congenital heart disease. a systematic review. *International journal of cardiology*. 2014;174(3):618-23. doi:10.1016/j.ijcard.2014.04.156.
759. Merkus D, de Beer VJ, Houweling B, Duncker DJ. Control of pulmonary vascular tone during exercise in health and pulmonary hypertension. *Pharmacology & therapeutics*. 2008;119(3):242-63. doi:10.1016/j.pharmthera.2008.04.003.
760. Jeffery TK, Morrell NW. Molecular and cellular basis of pulmonary vascular remodeling in pulmonary hypertension. *Progress in cardiovascular diseases*. 2002;45(3):173-202. doi:10.1053/pcad.2002.130041.

761. Xing XQ, Li YL, Zhang YX, Xiao Y, Li ZD, Liu LQ et al. Sphingosine kinase 1/sphingosine 1-phosphate signalling pathway as a potential therapeutic target of pulmonary hypertension. *International journal of clinical and experimental medicine*. 2015;8(8):11930-5.
762. Lenna S, Han R, Trojanowska M. Endoplasmic reticulum stress and endothelial dysfunction. *IUBMB life*. 2014;66(8):530-7. doi:10.1002/iub.1292.
763. Huertas A, Perros F, Tu L, Cohen-Kaminsky S, Montani D, Dorfmuller P et al. Immune dysregulation and endothelial dysfunction in pulmonary arterial hypertension: a complex interplay. *Circulation*. 2014;129(12):1332-40. doi:10.1161/CIRCULATIONAHA.113.004555.
764. Klinger JR, Abman SH, Gladwin MT. Nitric oxide deficiency and endothelial dysfunction in pulmonary arterial hypertension. *American journal of respiratory and critical care medicine*. 2013;188(6):639-46. doi:10.1164/rccm.201304-0686PP.
765. Sakao S, Tatsumi K. Crosstalk between endothelial cell and thrombus in chronic thromboembolic pulmonary hypertension: perspective. *Histology and histopathology*. 2013;28(2):185-93.
766. Amabile N, Guignabert C, Montani D, Yeghiazarians Y, Boulanger CM, Humbert M. Cellular microparticles in the pathogenesis of pulmonary hypertension. *The European respiratory journal*. 2013;42(1):272-9. doi:10.1183/09031936.00087212.
767. Hagan G, Pepke-Zaba J. Pulmonary hypertension, nitric oxide and nitric oxide-releasing compounds. *Expert review of respiratory medicine*. 2011;5(2):163-71. doi:10.1586/ers.11.5.
768. Shao D, Park JE, Wort SJ. The role of endothelin-1 in the pathogenesis of pulmonary arterial hypertension. *Pharmacological research*. 2011;63(6):504-11. doi:10.1016/j.phrs.2011.03.003.
769. Hui-li G. The management of acute pulmonary arterial hypertension. *Cardiovascular therapeutics*. 2011;29(3):153-75. doi:10.1111/j.1755-5922.2009.00095.x.
770. Bunn HF, Nathan DG, Dover GJ, Hebbel RP, Platt OS, Rosse WF et al. Pulmonary hypertension and nitric oxide depletion in sickle cell disease. *Blood*. 2010;116(5):687-92. doi:10.1182/blood-2010-02-268193.
771. Morrell NW. Role of bone morphogenetic protein receptors in the development of pulmonary arterial hypertension. *Advances in experimental medicine and biology*. 2010;661:251-64. doi:10.1007/978-1-60761-500-2_16.
772. Jurasz P, Courtman D, Babiak S, Stewart DJ. Role of apoptosis in pulmonary hypertension: from experimental models to clinical trials. *Pharmacology & therapeutics*. 2010;126(1):1-8. doi:10.1016/j.pharmthera.2009.12.006.
773. Morris CR, Gladwin MT, Kato GJ. Nitric oxide and arginine dysregulation: a novel pathway to pulmonary hypertension in hemolytic disorders. *Current molecular medicine*. 2008;8(7):620-32.
774. Humbert M, Montani D, Perros F, Dorfmuller P, Adnot S, Eddahibi S. Endothelial cell dysfunction and cross talk between endothelium and smooth muscle cells in pulmonary arterial hypertension. *Vascular pharmacology*. 2008;49(4-6):113-8. doi:10.1016/j.vph.2008.06.003.
775. Guignabert C, Tu L, Le Hiress M, Ricard N, Sattler C, Seferian A et al. Pathogenesis of pulmonary arterial hypertension: lessons from cancer. *European respiratory review : an official journal of the European Respiratory Society*. 2013;22(130):543-51. doi:10.1183/09059180.00007513.
776. Xu W, Erzurum SC. Endothelial cell energy metabolism, proliferation, and apoptosis in pulmonary hypertension. *Comprehensive Physiology*. 2011;1(1):357-72. doi:10.1002/cphy.c090005.
777. Sakao S, Tatsumi K, Voelkel NF. Endothelial cells and pulmonary arterial hypertension: apoptosis, proliferation, interaction and transdifferentiation. *Respiratory research*. 2009;10:95. doi:10.1186/1465-9921-10-95.
778. Goldthorpe H, Jiang JY, Taha M, Deng Y, Sinclair T, Ge CX et al. Occlusive Lung Arterial Lesions in Endothelial-targeted, Fas-Induced Apoptosis (FIA) Transgenic Mice. *American journal of respiratory cell and molecular biology*. 2015. doi:10.1165/rcmb.2014-0311OC.
779. Chen H, Strappe P, Chen S, Wang LX. Endothelial progenitor cells and pulmonary arterial hypertension. *Heart, lung & circulation*. 2014;23(7):595-601. doi:10.1016/j.hlc.2014.02.007.
780. Sato Y, Nakanuma Y. Role of endothelial-mesenchymal transition in idiopathic portal hypertension. *Histology and histopathology*. 2013;28(2):145-54.

781. Jane-Wit D, Chun HJ. Mechanisms of dysfunction in senescent pulmonary endothelium. *The journals of gerontology Series A, Biological sciences and medical sciences*. 2012;67(3):236-41. doi:10.1093/gerona/blr248.
782. Diller GP, Thum T, Wilkins MR, Wharton J. Endothelial progenitor cells in pulmonary arterial hypertension. *Trends in cardiovascular medicine*. 2010;20(1):22-9. doi:10.1016/j.tcm.2010.03.003.
783. Eickelberg O, Morty RE. Transforming growth factor beta/bone morphogenic protein signaling in pulmonary arterial hypertension: remodeling revisited. *Trends in cardiovascular medicine*. 2007;17(8):263-9. doi:10.1016/j.tcm.2007.09.003.
784. Nicod LP. The endothelium and genetics in pulmonary arterial hypertension. *Swiss medical weekly*. 2007;137(31-32):437-42. doi:2007/31/smw-11668.
785. Black SM, Kumar S, Wiseman D, Ravi K, Wedgwood S, Ryzhov V et al. Pediatric pulmonary hypertension: Roles of endothelin-1 and nitric oxide. *Clinical hemorheology and microcirculation*. 2007;37(1-2):111-20.
786. Barst RJ. A review of pulmonary arterial hypertension: role of ambrisentan. *Vascular health and risk management*. 2007;3(1):11-22.
787. Lahm T, Crisostomo PR, Markel TA, Wang M, Lillemoe KD, Meldrum DR. The critical role of vascular endothelial growth factor in pulmonary vascular remodeling after lung injury. *Shock*. 2007;28(1):4-14. doi:10.1097/shk.0b013e31804d1998.
788. Mathew R, Huang J, Gewitz MH. Pulmonary artery hypertension: caveolin-1 and eNOS interrelationship: a new perspective. *Cardiology in review*. 2007;15(3):143-9. doi:10.1097/01.crd.0000249381.49138.b9.
789. Arciniegas E, Frid MG, Douglas IS, Stenmark KR. Perspectives on endothelial-to-mesenchymal transition: potential contribution to vascular remodeling in chronic pulmonary hypertension. *American journal of physiology Lung cellular and molecular physiology*. 2007;293(1):L1-8. doi:10.1152/ajplung.00378.2006.
790. Hooper WC, Mensah GA, Haworth SG, Black SM, Garcia JG, Langleben D. Vascular endothelium summary statement V: Pulmonary hypertension and acute lung injury: public health implications. *Vascular pharmacology*. 2007;46(5):327-9. doi:10.1016/j.vph.2006.10.017.
791. Morrell NW. Pulmonary hypertension due to BMPR2 mutation: a new paradigm for tissue remodeling? *Proceedings of the American Thoracic Society*. 2006;3(8):680-6. doi:10.1513/pats.200605-118SF.
792. Black SM, Fineman JR. Oxidative and nitrosative stress in pediatric pulmonary hypertension: roles of endothelin-1 and nitric oxide. *Vascular pharmacology*. 2006;45(5):308-16. doi:10.1016/j.vph.2006.08.005.
793. Haworth SG. Role of the endothelium in pulmonary arterial hypertension. *Vascular pharmacology*. 2006;45(5):317-25. doi:10.1016/j.vph.2006.08.006.
794. Opitz CF, Ewert R. Dual ET(A)/ET(B) vs. selective ET(A) endothelin receptor antagonism in patients with pulmonary hypertension. *European journal of clinical investigation*. 2006;36 Suppl 3:1-9. doi:10.1111/j.1365-2362.2006.01691.x.
795. Ghofrani HA, Pepke-Zaba J, Barbera JA, Channick R, Keogh AM, Gomez-Sanchez MA et al. Nitric oxide pathway and phosphodiesterase inhibitors in pulmonary arterial hypertension. *Journal of the American College of Cardiology*. 2004;43(12 Suppl S):68S-72S. doi:10.1016/j.jacc.2004.02.031.
796. Napoli C, Loscalzo J. Nitric oxide and other novel therapies for pulmonary hypertension. *Journal of cardiovascular pharmacology and therapeutics*. 2004;9(1):1-8.
797. Budhiraja R, Tuder RM, Hassoun PM. Endothelial dysfunction in pulmonary hypertension. *Circulation*. 2004;109(2):159-65. doi:10.1161/01.CIR.0000102381.57477.50.
798. Michelakis ED, McMurtry MS, Sonnenberg B, Archer SL. The NO - K⁺ channel axis in pulmonary arterial hypertension. Activation by experimental oral therapies. *Advances in experimental medicine and biology*. 2003;543:293-322.
799. Michel RP, Langleben D, Dupuis J. The endothelin system in pulmonary hypertension. *Canadian journal of physiology and pharmacology*. 2003;81(6):542-54. doi:10.1139/y03-008.

800. Tuder RM, Voelkel NF. Angiogenesis and pulmonary hypertension: a unique process in a unique disease. *Antioxidants & redox signaling*. 2002;4(5):833-43. doi:10.1089/152308602760598990.
801. Chen YF, Oparil S. Endothelial dysfunction in the pulmonary vascular bed. *The American journal of the medical sciences*. 2000;320(4):223-32.
802. Veysier-Belot C, Cacoub P. Role of endothelial and smooth muscle cells in the physiopathology and treatment management of pulmonary hypertension. *Cardiovascular research*. 1999;44(2):274-82.
803. Abman SH. Abnormal vasoreactivity in the pathophysiology of persistent pulmonary hypertension of the newborn. *Pediatrics in review / American Academy of Pediatrics*. 1999;20(11):e103-9.
804. Sperling RT, Creager MA. Nitric oxide and pulmonary hypertension. *Coronary artery disease*. 1999;10(5):287-94.
805. Higenbottam TW, Laude EA. Endothelial dysfunction providing the basis for the treatment of pulmonary hypertension: Giles F. Filley lecture. *Chest*. 1998;114(1 Suppl):72S-9S.
806. McQuaid KE, Keenan AK. Endothelial barrier dysfunction and oxidative stress: roles for nitric oxide? *Experimental physiology*. 1997;82(2):369-76.
807. Higenbottam T. Pathophysiology of pulmonary hypertension. A role for endothelial dysfunction. *Chest*. 1994;105(2 Suppl):7S-12S.
808. Lang IM, Gaine SP. Recent advances in targeting the prostacyclin pathway in pulmonary arterial hypertension. *European respiratory review : an official journal of the European Respiratory Society*. 2015;24(138):630-41. doi:10.1183/16000617.0067-2015.
809. Jimenez SA, Piera-Velazquez S. Endothelial to mesenchymal transition (EndoMT) in the pathogenesis of Systemic Sclerosis-associated pulmonary fibrosis and pulmonary arterial hypertension. Myth or reality? *Matrix biology : journal of the International Society for Matrix Biology*. 2016. doi:10.1016/j.matbio.2016.01.012.
810. Keller RL. Pulmonary Hypertension and Pulmonary Vasodilators. *Clinics in perinatology*. 2016;43(1):187-202. doi:10.1016/j.clp.2015.11.013.
811. Montani D, Chaumais MC, Guignabert C, Gunther S, Girerd B, Jais X et al. Targeted therapies in pulmonary arterial hypertension. *Pharmacology & therapeutics*. 2014;141(2):172-91. doi:10.1016/j.pharmthera.2013.10.002.
812. de Man FS, Handoko ML, Guignabert C, Bogaard HJ, Vonk-Noordegraaf A. Neurohormonal axis in patients with pulmonary arterial hypertension: friend or foe? *American journal of respiratory and critical care medicine*. 2013;187(1):14-9. doi:10.1164/rccm.201209-1663PP.
813. Rabinovitch M. PPARgamma and the pathobiology of pulmonary arterial hypertension. *Advances in experimental medicine and biology*. 2010;661:447-58. doi:10.1007/978-1-60761-500-2_29.
814. Rabinovitch M. Structure and function of the pulmonary vascular bed: an update. *Cardiology clinics*. 1989;7(2):227-38.
815. Perrin S, Chaumais MC, O'Connell C, Amar D, Savale L, Jais X et al. New pharmacotherapy options for pulmonary arterial hypertension. *Expert opinion on pharmacotherapy*. 2015;16(14):2113-31. doi:10.1517/14656566.2015.1074177.
816. Humbert M, Ghofrani HA. The molecular targets of approved treatments for pulmonary arterial hypertension. *Thorax*. 2016;71(1):73-83. doi:10.1136/thoraxjnl-2015-207170.
817. Ghofrani HA, Humbert M. The role of combination therapy in managing pulmonary arterial hypertension. *European respiratory review : an official journal of the European Respiratory Society*. 2014;23(134):469-75. doi:10.1183/09059180.00007314.
818. Clozel M, Maresta A, Humbert M. Endothelin receptor antagonists. *Handbook of experimental pharmacology*. 2013;218:199-227. doi:10.1007/978-3-642-38664-0_9.
819. Bienertova-Vasku J, Novak J, Vasku A. MicroRNAs in pulmonary arterial hypertension: pathogenesis, diagnosis and treatment. *Journal of the American Society of Hypertension : JASH*. 2015;9(3):221-34. doi:10.1016/j.jash.2014.12.011.

820. Guignabert C, Tu L, Girerd B, Ricard N, Huertas A, Montani D et al. New molecular targets of pulmonary vascular remodeling in pulmonary arterial hypertension: importance of endothelial communication. *Chest*. 2015;147(2):529-37. doi:10.1378/chest.14-0862.
821. Chaumais MC, Guignabert C, Savale L, Jais X, Boucly A, Montani D et al. Clinical pharmacology of endothelin receptor antagonists used in the treatment of pulmonary arterial hypertension. *American journal of cardiovascular drugs : drugs, devices, and other interventions*. 2015;15(1):13-26. doi:10.1007/s40256-014-0095-y.
822. Mathew R. Pathogenesis of pulmonary hypertension: a case for caveolin-1 and cell membrane integrity. *American journal of physiology Heart and circulatory physiology*. 2014;306(1):H15-25. doi:10.1152/ajpheart.00266.2013.
823. Antoniu SA. Targeting PDGF pathway in pulmonary arterial hypertension. *Expert opinion on therapeutic targets*. 2012;16(11):1055-63. doi:10.1517/14728222.2012.719500.
824. Sutliff RL, Kang BY, Hart CM. PPARgamma as a potential therapeutic target in pulmonary hypertension. *Therapeutic advances in respiratory disease*. 2010;4(3):143-60. doi:10.1177/1753465809369619.
825. Ghofrani HA, Voswinckel R, Gall H, Schermuly R, Weissmann N, Seeger W et al. Riociguat for pulmonary hypertension. *Future cardiology*. 2010;6(2):155-66. doi:10.2217/fca.10.5.
826. Montani D, Chaumais MC, Savale L, Natali D, Price LC, Jais X et al. Phosphodiesterase type 5 inhibitors in pulmonary arterial hypertension. *Advances in therapy*. 2009;26(9):813-25. doi:10.1007/s12325-009-0064-z.
827. Davie NJ, Schermuly RT, Weissmann N, Grimminger F, Ghofrani HA. The science of endothelin-1 and endothelin receptor antagonists in the management of pulmonary arterial hypertension: current understanding and future studies. *European journal of clinical investigation*. 2009;39 Suppl 2:38-49. doi:10.1111/j.1365-2362.2009.02120.x.
828. Price LC, Howard LS. Endothelin receptor antagonists for pulmonary arterial hypertension: rationale and place in therapy. *American journal of cardiovascular drugs : drugs, devices, and other interventions*. 2008;8(3):171-85.
829. Fukumoto Y, Tawara S, Shimokawa H. Recent progress in the treatment of pulmonary arterial hypertension: expectation for rho-kinase inhibitors. *The Tohoku journal of experimental medicine*. 2007;211(4):309-20.
830. Xing XQ, Gan Y, Wu SJ, Chen P, Zhou R, Xiang XD. Rho-kinase as a potential therapeutic target for the treatment of pulmonary hypertension. *Drug news & perspectives*. 2006;19(9):517-22. doi:10.1358/dnp.2006.19.9.1050426.
831. Jacobs A, Preston IR, Gomberg-Maitland M. Endothelin receptor antagonism in pulmonary arterial hypertension--a role for selective ET(A) inhibition? *Current medical research and opinion*. 2006;22(12):2567-74. doi:10.1185/030079906X158020.
832. Patel MD, Katz SD. Phosphodiesterase 5 inhibition in chronic heart failure and pulmonary hypertension. *The American journal of cardiology*. 2005;96(12B):47M-51M. doi:10.1016/j.amjcard.2005.10.005.
833. Barst RJ. PDGF signaling in pulmonary arterial hypertension. *The Journal of clinical investigation*. 2005;115(10):2691-4. doi:10.1172/JCI26593.
834. Clozel M. Effects of bosentan on cellular processes involved in pulmonary arterial hypertension: do they explain the long-term benefit? *Annals of medicine*. 2003;35(8):605-13.
835. Travadi JN, Patole SK. Phosphodiesterase inhibitors for persistent pulmonary hypertension of the newborn: a review. *Pediatric pulmonology*. 2003;36(6):529-35. doi:10.1002/ppul.10389.
836. Mandegar M, Remillard CV, Yuan JX. Ion channels in pulmonary arterial hypertension. *Progress in cardiovascular diseases*. 2002;45(2):81-114.
837. Michelakis ED, Weir EK. The pathobiology of pulmonary hypertension. *Smooth muscle cells and ion channels*. *Clinics in chest medicine*. 2001;22(3):419-32.
838. Boucherat O, Chabot S, Antigny F, Perros F, Provencher S, Bonnet S. Potassium channels in pulmonary arterial hypertension. *The European respiratory journal*. 2015;46(4):1167-77. doi:10.1183/13993003.00798-2015.

839. Dasgupta A, Bowman L, D'Arsigny CL, Archer SL. Soluble guanylate cyclase: a new therapeutic target for pulmonary arterial hypertension and chronic thromboembolic pulmonary hypertension. *Clinical pharmacology and therapeutics*. 2015;97(1):88-102. doi:10.1002/cpt.10.
840. Kao CC, Wedes SH, Hsu JW, Bohren KM, Comhair SA, Jahoor F et al. Arginine metabolic endotypes in pulmonary arterial hypertension. *Pulmonary circulation*. 2015;5(1):124-34. doi:10.1086/679720.
841. Fessel JP, West JD. Redox biology in pulmonary arterial hypertension (2013 Grover Conference Series). *Pulmonary circulation*. 2015;5(4):599-609. doi:10.1086/683814.
842. Voelkel NF, Tuder RM. Cellular and molecular biology of vascular smooth muscle cells in pulmonary hypertension. *Pulmonary pharmacology & therapeutics*. 1997;10(5-6):231-41. doi:10.1006/pupt.1998.0100.
843. Tu L, Guignabert C. Emerging molecular targets for anti-proliferative strategies in pulmonary arterial hypertension. *Handbook of experimental pharmacology*. 2013;218:409-36. doi:10.1007/978-3-642-38664-0_17.
844. Guignabert C, Dorfmüller P. Pathology and pathobiology of pulmonary hypertension. *Seminars in respiratory and critical care medicine*. 2013;34(5):551-9. doi:10.1055/s-0033-1356496.
845. de Jesus Perez V, Yuan K, Alastalo TP, Spiekeroetter E, Rabinovitch M. Targeting the Wnt signaling pathways in pulmonary arterial hypertension. *Drug discovery today*. 2014;19(8):1270-6. doi:10.1016/j.drudis.2014.06.014.
846. Rabinovitch M. Molecular pathogenesis of pulmonary arterial hypertension. *The Journal of clinical investigation*. 2012;122(12):4306-13. doi:10.1172/JCI60658.
847. Umar S, Rabinovitch M, Eghbali M. Estrogen paradox in pulmonary hypertension: current controversies and future perspectives. *American journal of respiratory and critical care medicine*. 2012;186(2):125-31. doi:10.1164/rccm.201201-0058PP.
848. Stenmark KR, Rabinovitch M. Emerging therapies for the treatment of 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*. 2010;11(2 Suppl):S85-90. doi:10.1097/PCC.0b013e3181c76db3.
849. He J, Bao Q, Yan M, Liang J, Zhu Y, Wang C et al. The Role of Hippo/YAP Signaling in Vascular Remodeling and Related Diseases. *British journal of pharmacology*. 2017. doi:10.1111/bph.13806.
850. Tuder RM. Pulmonary vascular remodeling in pulmonary hypertension. *Cell Tissue Res*. 2017;367(3):643-9. doi:10.1007/s00441-016-2539-y.
851. Hayabuchi Y. The Action of Smooth Muscle Cell Potassium Channels in the Pathology of Pulmonary Arterial Hypertension. *Pediatr Cardiol*. 2017;38(1):1-14. doi:10.1007/s00246-016-1491-7.
852. Abe K, Shinoda M, Tanaka M, Kuwabara Y, Yoshida K, Hirooka Y et al. Haemodynamic unloading reverses occlusive vascular lesions in severe pulmonary hypertension. *Cardiovascular research*. 2016;111(1):16-25. doi:10.1093/cvr/cvw070.
853. Abe K, Toba M, Alzoubi A, Ito M, Fagan KA, Cool CD et al. Formation of plexiform lesions in experimental severe pulmonary arterial hypertension. *Circulation*. 2010;121(25):2747-54. doi:10.1161/CIRCULATIONAHA.109.927681.
854. Tuder RM, Voelkel NF. Plexiform lesion in severe pulmonary hypertension: association with glomeruloid lesion. *The American journal of pathology*. 2001;159(1):382-3. doi:10.1016/S0002-9440(10)61705-1.
855. Hirose S, Hosoda Y, Furuya S, Otsuki T, Ikeda E. Expression of vascular endothelial growth factor and its receptors correlates closely with formation of the plexiform lesion in human pulmonary hypertension. *Pathol Int*. 2000;50(6):472-9.
856. Cool CD, Stewart JS, Werahera P, Miller GJ, Williams RL, Voelkel NF et al. Three-dimensional reconstruction of pulmonary arteries in plexiform pulmonary hypertension using cell-specific markers. Evidence for a dynamic and heterogeneous process of pulmonary endothelial cell growth. *The American journal of pathology*. 1999;155(2):411-9. doi:10.1016/S0002-9440(10)65137-1.
857. Jamison BM, Michel RP. Different distribution of plexiform lesions in primary and secondary pulmonary hypertension. *Human pathology*. 1995;26(9):987-93.

858. Tuder RM, Groves B, Badesch DB, Voelkel NF. Exuberant endothelial cell growth and elements of inflammation are present in plexiform lesions of pulmonary hypertension. *The American journal of pathology*. 1994;144(2):275-85.
859. Ogata T, Iijima T. Structure and pathogenesis of plexiform lesion in pulmonary hypertension. *Chinese medical journal*. 1993;106(1):45-8.
860. Hsieh SP, Wang JS, Tsui CY, Liu HC. Plexogenic pulmonary vascular lesions in primary pulmonary arteriopathy--report of two autopsy cases. *Zhonghua Yi Xue Za Zhi (Taipei)*. 1990;45(2):134-8.
861. Smith P, Heath D. Electron microscopy of the plexiform lesion. *Thorax*. 1979;34(2):177-86.
862. Kanjuh VI, Sellers RD, Edwards JE. Pulmonary Vascular Plexiform Lesion. *Pathogenetic Studies*. *Arch Pathol*. 1964;78:513-22.
863. Galambos C, Sims-Lucas S, Abman SH, Cool CD. Intrapulmonary Bronchopulmonary Anastomoses and Plexiform Lesions in Idiopathic Pulmonary Arterial Hypertension. *American journal of respiratory and critical care medicine*. 2016;193(5):574-6. doi:10.1164/rccm.201507-1508LE.
864. Corte TJ, Wort SJ, Wells AU. Pulmonary hypertension in idiopathic pulmonary fibrosis: a review. Sarcoidosis, vasculitis, and diffuse lung diseases : official journal of WASOG / World Association of Sarcoidosis and Other Granulomatous Disorders. 2009;26(1):7-19.
865. Rabinovitch M. Pathobiology of pulmonary hypertension. Extracellular matrix. *Clinics in chest medicine*. 2001;22(3):433-49, viii.
866. Farkas L, Gauldie J, Voelkel NF, Kolb M. Pulmonary hypertension and idiopathic pulmonary fibrosis: a tale of angiogenesis, apoptosis, and growth factors. *American journal of respiratory cell and molecular biology*. 2011;45(1):1-15. doi:10.1165/rcmb.2010-0365TR.
867. Rabinovitch M. EVE and beyond, retro and prospective insights. *The American journal of physiology*. 1999;277(1 Pt 1):L5-12.
868. Reviron-Rabec L, Girerd B, Seferian A, Campbell K, Brosseau S, Bergot E et al. Pulmonary complications of type 1 neurofibromatosis. *Revue des maladies respiratoires*. 2016. doi:10.1016/j.rmr.2014.09.010.
869. Di Wang H, Ratsep MT, Chapman A, Boyd R. Adventitial fibroblasts in vascular structure and function: the role of oxidative stress and beyond. *Canadian journal of physiology and pharmacology*. 2010;88(3):177-86. doi:10.1139/Y10-015.
870. Harvey LD, Chan SY. Emerging Metabolic Therapies in Pulmonary Arterial Hypertension. *J Clin Med*. 2017;6(4). doi:10.3390/jcm6040043.
871. Lopez-Crisosto C, Pennanen C, Vasquez-Trincado C, Morales PE, Bravo-Sagua R, Quest AF et al. Sarcoplasmic reticulum-mitochondria communication in cardiovascular pathophysiology. *Nat Rev Cardiol*. 2017. doi:10.1038/nrcardio.2017.23.
872. Liu N, Parry S, Xiao Y, Zhou S, Liu Q. Molecular targets of the Warburg effect and inflammatory cytokines in the pathogenesis of pulmonary artery hypertension. *Clin Chim Acta*. 2017;466:98-104. doi:10.1016/j.cca.2017.01.015.
873. Sharma S, Ruffenach G, Umar S, Motayagheni N, Reddy ST, Eghbali M. Role of oxidized lipids in pulmonary arterial hypertension. *Pulmonary circulation*. 2016;6(3):261-73. doi:10.1086/687293.
874. Lewis GD, Ngo D, Hemnes AR, Farrell L, Doms C, Pappagianopoulos PP et al. Metabolic Profiling of Right Ventricular-Pulmonary Vascular Function Reveals Circulating Biomarkers of Pulmonary Hypertension. *Journal of the American College of Cardiology*. 2016;67(2):174-89. doi:10.1016/j.jacc.2015.10.072.
875. Berger G, Azzam ZS, Hoffman R, Yigla M. Coagulation and anticoagulation in pulmonary arterial hypertension. *The Israel Medical Association journal : IMAJ*. 2009;11(6):376-9.
876. Lopes AA. Pathophysiological basis for anticoagulant and antithrombotic therapy in pulmonary hypertension. *Cardiovascular & hematological agents in medicinal chemistry*. 2006;4(1):53-9.
877. Herve P, Humbert M, Sitbon O, Parent F, Nunes H, Legal C et al. Pathobiology of pulmonary hypertension. The role of platelets and thrombosis. *Clinics in chest medicine*. 2001;22(3):451-8.
878. Farber HW, Loscalzo J. Prothrombotic mechanisms in primary pulmonary hypertension. *The Journal of laboratory and clinical medicine*. 1999;134(6):561-6.

879. Chaouat A, Weitzenblum E, Higenbottam T. The role of thrombosis in severe pulmonary hypertension. *The European respiratory journal*. 1996;9(2):356-63.
880. Rich S, Levitsky S, Brundage BH. Pulmonary hypertension from chronic pulmonary thromboembolism. *Annals of internal medicine*. 1988;108(3):425-34.
881. Ezedunukwe IR, Enuh H, Nfonoyim J, Enuh CU. Anticoagulation therapy versus placebo for pulmonary hypertension. *The Cochrane database of systematic reviews*. 2014;6:CD010695. doi:10.1002/14651858.CD010695.pub2.
882. Lannan KL, Phipps RP, White RJ. Thrombosis, platelets, microparticles and PAH: more than a clot. *Drug discovery today*. 2014;19(8):1230-5. doi:10.1016/j.drudis.2014.04.001.
883. Klok FA, Mos IC, van Kralingen KW, Vahl JE, Huisman MV. Chronic pulmonary embolism and pulmonary hypertension. *Seminars in respiratory and critical care medicine*. 2012;33(2):199-204. doi:10.1055/s-0032-1311800.
884. Lang I. Advances in understanding the pathogenesis of chronic thromboembolic pulmonary hypertension. *British journal of haematology*. 2010;149(4):478-83. doi:10.1111/j.1365-2141.2010.08142.x.
885. Opitz CF, Kirch W, Mueller EA, Pittrow D. Bleeding events in pulmonary arterial hypertension. *European journal of clinical investigation*. 2009;39 Suppl 2:68-73. doi:10.1111/j.1365-2362.2009.02122.x.
886. Lee MT, Rosenzweig EB, Cairo MS. Pulmonary hypertension in sickle cell disease. *Clinical advances in hematology & oncology : H&O*. 2007;5(8):645-53, 585.
887. Johnson SR, Granton JT, Mehta S. Thrombotic arteriopathy and anticoagulation in pulmonary hypertension. *Chest*. 2006;130(2):545-52. doi:10.1378/chest.130.2.545.
888. Egermayer P, Peacock AJ. Is pulmonary embolism a common cause of chronic pulmonary hypertension? Limitations of the embolic hypothesis. *The European respiratory journal*. 2000;15(3):440-8.
889. Schultze AE, Roth RA. Chronic pulmonary hypertension--the monocrotaline model and involvement of the hemostatic system. *Journal of toxicology and environmental health Part B, Critical reviews*. 1998;1(4):271-346. doi:10.1080/10937409809524557.
890. Hassell KL. Altered hemostasis in pulmonary hypertension. *Blood coagulation & fibrinolysis : an international journal in haemostasis and thrombosis*. 1998;9(2):107-17.
891. Palevsky HI. Pulmonary hypertension and thromboembolic disease in women. *Cardiovascular clinics*. 1989;19(3):267-83.
892. Robinson JC, Pugliese SC, Fox DL, Badesch DB. Anticoagulation in Pulmonary Arterial Hypertension. *Current hypertension reports*. 2016;18(6):47. doi:10.1007/s11906-016-0657-2.
893. Kadavath S, Zapantis E, Zolty R, Efthimiou P. A novel therapeutic approach in pulmonary arterial hypertension as a complication of adult-onset Still's disease: targeting IL-6. *International journal of rheumatic diseases*. 2014;17(3):336-40. doi:10.1111/1756-185X.12324.
894. Kato M, Kataoka H, Odani T, Fujieda Y, Otomo K, Oku K et al. The short-term role of corticosteroid therapy for pulmonary arterial hypertension associated with connective tissue diseases: report of five cases and a literature review. *Lupus*. 2011;20(10):1047-56. doi:10.1177/0961203311403347.
895. Kherbeck N, Tamby MC, Bussone G, Dib H, Perros F, Humbert M et al. The role of inflammation and autoimmunity in the pathophysiology of pulmonary arterial hypertension. *Clinical reviews in allergy & immunology*. 2013;44(1):31-8. doi:10.1007/s12016-011-8265-z.
896. Hassoun PM, Mouthon L, Barbera JA, Eddahibi S, Flores SC, Grimminger F et al. Inflammation, growth factors, and pulmonary vascular remodeling. *Journal of the American College of Cardiology*. 2009;54(1 Suppl):S10-9. doi:10.1016/j.jacc.2009.04.006.
897. El Kasmi KC, Stenmark KR. Contribution of metabolic reprogramming to macrophage plasticity and function. *Seminars in immunology*. 2015;27(4):267-75. doi:10.1016/j.smim.2015.09.001.
898. Stenmark KR, Davie NJ, Reeves JT, Frid MG. Hypoxia, leukocytes, and the pulmonary circulation. *Journal of applied physiology*. 2005;98(2):715-21. doi:10.1152/japplphysiol.00840.2004.

899. Durmowicz AG, Stenmark KR. Mechanisms of structural remodeling in chronic pulmonary hypertension. *Pediatrics in review / American Academy of Pediatrics*. 1999;20(11):e91-e102.
900. Stenmark KR, Mecham RP. Cellular and molecular mechanisms of pulmonary vascular remodeling. *Annual review of physiology*. 1997;59:89-144. doi:10.1146/annurev.physiol.59.1.89.
901. Voelkel NF, Cool C, Lee SD, Wright L, Geraci MW, Tuder RM. Primary pulmonary hypertension between inflammation and cancer. *Chest*. 1998;114(3 Suppl):225S-30S.
902. Tuder RM, Voelkel NF. Pulmonary hypertension and inflammation. *The Journal of laboratory and clinical medicine*. 1998;132(1):16-24.
903. Rabinovitch M, Guignabert C, Humbert M, Nicolls MR. Inflammation and immunity in the pathogenesis of pulmonary arterial hypertension. *Circulation research*. 2014;115(1):165-75. doi:10.1161/CIRCRESAHA.113.301141.
904. Nicolls MR, Taraseviciene-Stewart L, Rai PR, Badesch DB, Voelkel NF. Autoimmunity and pulmonary hypertension: a perspective. *The European respiratory journal*. 2005;26(6):1110-8. doi:10.1183/09031936.05.00045705.
905. Rabinovitch M. Elastase and the pathobiology of unexplained pulmonary hypertension. *Chest*. 1998;114(3 Suppl):213S-24S.
906. Rabinovitch M. It all begins with EVE (endogenous vascular elastase). *Israel journal of medical sciences*. 1996;32(10):803-8; discussion 9-10.
907. Rabinovitch M. Elastase and cell matrix interactions in the pathobiology of vascular disease. *Acta paediatrica Japonica; Overseas edition*. 1995;37(6):657-66.
908. Gashouta MA, Humbert M, Hassoun PM. Update in systemic sclerosis-associated pulmonary arterial hypertension. *Presse medicale*. 2014;43(10 Pt 2):e293-304. doi:10.1016/j.lpm.2014.06.007.
909. Cohen-Kaminsky S, Hautefort A, Price L, Humbert M, Perros F. Inflammation in pulmonary hypertension: what we know and what we could logically and safely target first. *Drug discovery today*. 2014;19(8):1251-6. doi:10.1016/j.drudis.2014.04.007.
910. Collard HR, Morris A, Daley CL, Humbert M, Meyer KC. Clinical year in review IV: HIV, mycobacterial disease, pulmonary hypertension, and interstitial lung disease. *Proceedings of the American Thoracic Society*. 2012;9(4):204-9. doi:10.1513/pats.201207-035TT.
911. Lefevre G, Dauchet L, Hachulla E, Montani D, Sobanski V, Lambert M et al. Survival and prognostic factors in systemic sclerosis-associated pulmonary hypertension: a systematic review and meta-analysis. *Arthritis and rheumatism*. 2013;65(9):2412-23. doi:10.1002/art.38029.
912. O'Connell C, O'Callaghan DS, Humbert M. Novel medical therapies for pulmonary arterial hypertension. *Clinics in chest medicine*. 2013;34(4):867-80. doi:10.1016/j.ccm.2013.08.002.
913. Price LC, Wort SJ, Perros F, Dorfmuller P, Huertas A, Montani D et al. Inflammation in pulmonary arterial hypertension. *Chest*. 2012;141(1):210-21. doi:10.1378/chest.11-0793.
914. Dorfmuller P, Perros F, Balabanian K, Humbert M. Inflammation in pulmonary arterial hypertension. *The European respiratory journal*. 2003;22(2):358-63.
915. Walker KM, Pope J, participating members of the Scleroderma Clinical Trials C, Canadian Scleroderma Research G. Treatment of systemic sclerosis complications: what to use when first-line treatment fails--a consensus of systemic sclerosis experts. *Seminars in arthritis and rheumatism*. 2012;42(1):42-55. doi:10.1016/j.semarthrit.2012.01.003.
916. Le Pavec J, Humbert M, Mouthon L, Hassoun PM. Systemic sclerosis-associated pulmonary arterial hypertension. *American journal of respiratory and critical care medicine*. 2010;181(12):1285-93. doi:10.1164/rccm.200909-1331PP.
917. Ranque B, Mouthon L. Geoepidemiology of systemic sclerosis. *Autoimmunity reviews*. 2010;9(5):A311-8. doi:10.1016/j.autrev.2009.11.003.
918. Meloche J, Renard S, Provencher S, Bonnet S. Anti-inflammatory and immunosuppressive agents in PAH. *Handbook of experimental pharmacology*. 2013;218:437-76. doi:10.1007/978-3-642-38664-0_18.
919. Mirrakhimov AE, Hill NS. Primary antiphospholipid syndrome and pulmonary hypertension. *Current pharmaceutical design*. 2014;20(4):545-51.

920. Agmon-Levin N, Selmi C. The autoimmune side of heart and lung diseases. *Clinical reviews in allergy & immunology*. 2013;44(1):1-5. doi:10.1007/s12016-012-8335-x.
921. El Chami H, Hassoun PM. Inflammatory mechanisms in the pathogenesis of pulmonary arterial hypertension. *Comprehensive Physiology*. 2011;1(4):1929-41. doi:10.1002/cphy.c100028.
922. Sanges S, Yelnik CM, Sitbon O, Benveniste O, Mariampillai K, Phillips-Houlbracq M et al. Pulmonary arterial hypertension in idiopathic inflammatory myopathies: Data from the French pulmonary hypertension registry and review of the literature. *Medicine (Baltimore)*. 2016;95(39):e4911. doi:10.1097/MD.0000000000004911.
923. Voelkel NF, Tamosiuniene R, Nicolls MR. Challenges and opportunities in treating inflammation associated with pulmonary hypertension. *Expert review of cardiovascular therapy*. 2016;14(8):939-51. doi:10.1080/14779072.2016.1180976.