

Wanda K. O'Neal, Ph.D.

OFFICE ADDRESS:

Marsico Lung Institute/ Cystic Fibrosis Center
 7212 Marsico Hall, CB # 7248
 University of North Carolina at Chapel Hill
 Chapel Hill, North Carolina 27599-7248
 Phone: (919) 962-9866
 FAX: (919) 966-6821
 Email: woneal@med.unc.edu

Professional Employment and Training:

2002-Present	Research Associate Professor and Director of Molecular Biology Core Laboratory CF/Pulmonary Research and Treatment Center, Department of Medicine, University of North Carolina at Chapel Hill
1998-2002	Research Assistant Professor and Director of Molecular Biology Core Laboratory - CF/Pulmonary Research and Treatment Center, Department of Medicine, University of North Carolina at Chapel Hill
1997-1998	Assistant Professor - Department of Molecular and Human Genetics Baylor College of Medicine, Houston, Texas
1993-1997	Postdoctoral Fellow - Supervising Professor: Arthur L. Beaudet, M.D. Department of Molecular and Human Genetics, Baylor College of Medicine, Houston, Texas. Development of Adenovirus Vectors for Gene Therapy with Emphasis on Treatment of Genetic Disorders of the Lung and Liver
1988-1993	Graduate Student - Advisor: Arthur L. Beaudet, M.D. Department of Molecular and Human Genetics, Baylor College of Medicine, Houston, Texas. Thesis Title: "Generation and Characterization of a Murine Model for Cystic Fibrosis."
1984-1987	Masters Degree Student and Graduate Teaching Fellow Advisor: Calvin Messersmith, Ph.D., Department of Crop Science. North Dakota State University, Fargo, North Dakota

Education:

Doctor of Philosophy 1993	Baylor College of Medicine - Department of Molecular and Human Genetics, Houston, Texas
Master of Science 1987	North Dakota State University Crop Science Department, Fargo, North Dakota
Bachelors of Science 1984	North Dakota State University Plant Pathology and Crop Science, Fargo, North Dakota

Present and Completed Research Support

2016-2017	NIH/NHLBI SBIR sub, "A Targeted Drug for the Treatment of Inflammation in Cystic Fibrosis Lung Disease," Co-investigator
2015-2019	NIH/NHLBI "The Role of Mucus and Pulmonary Surface Intersections in Lung Disease," Co-investigator
2015-2019	Cystic Fibrosis Foundation "Epithelial Function in Cystic Fibrosis, Core E: Molecular Biology and Mouse Core, Core Leader
2015-2010	NIH/NIDDK "UNC Cystic Fibrosis Research and Translation Core Center, Core B: Pre-clinical Core," Core Co-Leader
2014-2019	NIH/NHLBI "Synthesis of Effective and Safe Mucolytics for Pulmonary Disease," Co-investigator

2013-2018 NIH/NHLBI "The Impact of Tobacco Exposure on the Lung's Innate Defense System, Project 3: Mouse Models of Human Smoking-related Diseases: What is the Best Mimic of Human Disease?" Co-investigator

2013-2018 NIH/NHLBI "Mining Open Chromatin to Define Molecular Mechanisms of CF modifier Genes," Co-Principal Investigator

2012-2017 NIH/NHLBI "Pulmonary Surface Liquid Homeostasis, Core B: Molecular Biology Core," Core Leader

2012-2017 NIH/NHLBI Novel Therapies for Muco-Obstructive Lung Diseases, Project II: Mucus Obstructed Mice for Biomarker and Drug Development," Co-Investigator

2012-2015 NIH/NHLBI "Gene Modifiers in CF Lung Disease," Co-Investigator

2010-2015 NIH/NHLBI "The Role of Mucin-Protein Interactions in the Innate Defense of the Lung," Co-Investigator

2009-2014 NIH/NIDDK "Molecular Therapy Core Center, Core C: Molecular Core," Core Director

2010-2013 NIH/NHLBI "Role of Prolyl Endopeptidase as a Novel Regulator in Pulmonary Neutrophilic Inflammation," Co-Investigator

2010-2013 American Asthma Foundation "IRE1beta-Dependent Airway Mucin Production and ATP Release: A New Pathway in Asthma," Co-Investigator

2010-2011 Cystic Fibrosis Foundation, "Feasibility of Na⁺ Channel Blocker Therapy for CF," Principal Investigator

2007-2017 NIH/NHLBI "Pulmonary Epithelia in Health and Disease, Core B: Molecular and Protein Core," Core Director

2007-2015 Cystic Fibrosis Foundation "Epithelial Function in Cystic Fibrosis, Core E. Molecular Biology and Mouse Core," Core Director

2009-2016 NIH/NHLBI "SPIROMICS Genomics and Informatics Center" Co-investigator

2008-2012 NIH/NHLBI "Molecular Phenotypes for Cystic Fibrosis Lung Disease," Co-investigator.

2009-2010 NIH/NCRR "Development of the betaENaC Model of Cystic Fibrosis for Translational Research," Sub-contract Principal Investigator

2001-2004 Cystic Fibrosis Foundation Research Grant "CF Therapeutic Targets Revealed by Expression Arrays," Principal Investigator.

2007-2009 Cystic Fibrosis Foundation "Epithelial Function in Cystic Fibrosis, Project 2. Transmembrane Mucin Function in Mucociliary Clearance," Principal Investigator.

2007-2009 Cystic Fibrosis Foundation, "Dysregulated Airway Physiology in Scnn1b Transgenic Mice." Co-investigator.

2007-2009 Cystic Fibrosis Foundation, "CF Modifiers Defined by Scnn1b Over-expressing mice," Principal Investigator.

2006-2008 NIH/NIDDK "Molecular Therapy Core Center, Project 6: Interfering RNA for Modulation of ENaC Function," Project Leader.

2001-2006 NIH/NHLBI "Pulmonary Epithelia in Health and Disease", Core B, Molecular Core, Principal Investigator

2001-2006 "Gene Therapy for Pulmonary and Hematologic Disorders, Project 3: Extra Cellular Barriers to Gene Transfer in the Lung", Co-Investigator (10% effort)

2002-2006 NIH/NHLBI "A Ciliated Cell-specific Promoter for Gene Therapy of CF", Co-Investigator (10% effort)

2002-2006 Cystic Fibrosis Foundation "Molecular Core", Principal Investigator

2001-2005 NIH/NHLBI & NIDDK "Enhanced Gene Transfer to Lung Epithelia", Co-investigator (10% effort)

2000-2002 Cystic Fibrosis Foundation, Gene Therapy Pilot and Feasibility Grant "MUC-4 as a Barrier to Lung Gene Transfer," Principal Investigator.

2000-2001 Cystic Fibrosis Foundation Research Grant "In Vitro Cell Models for CFTR Expression," Principal Investigator.

1998-2000 NIDDK R21 Grant: "Large Deletion Adenoviral Vectors for Cystic Fibrosis," Principal Investigator.

1994-1997 Cystic Fibrosis Foundation Postdoctoral Fellowship

1993-1994 Postdoctoral Fellowship - NIH Training Grant

1989-1993 March of Dimes Graduate Student Fellowship

1985-1988 Graduate Teaching Fellowship, North Dakota State University

NIH Study Section Membership:

2004-2007
2010-2012

NHLBI SBIR Study Section Ad-Hoc Reviewer
NHLBI K99 Award Study Section Ad-Hoc Reviewer

Research Publications, peer reviewed (from newest to oldest):

1. Chen G, Ribeiro CMP, Sun L, Okuda K, Kato T, Gilmore RC, Martino MB, Dang H, Abzhanova A, Lin JM, Hull-Ryde EA, Volmer AS, Randell SH, Livraghi-Butrico A, Deng Y, Scherer PE, Stripp BR, **O'Neal WK**, Boucher RC. 2019. XBP1S regulates MUC5B in a promoter variant-dependent pathway in IPF airway epithelia. *Am J Respir Crit Care Med*. [Epub ahead of print]. PMID: 30973754.
2. Christenson SA, van den Berge M, Faiz A, Inkamp K, Bhakta N, Bonser LR, Zlock LT, Barjaktarevic IZ, Barr RG, Bleecker ER, Boucher RC, Bowler RP, Comellas AP, Curtis JL, Han MK, Hansel NN, Hiemstra PS, Kaner RJ, Krishnanm JA, Martinez FJ, **O'Neal WK**, Paine R 3rd, Timens W, Wells JM, Spira A, Erle DJ, Woodruff PG. 2019. An airway epithelial IL-17A response signature identifies a steroid-unresponsive COPD patient subgroup. *J Clin Invest*. 129(1):169-181. PMID: 30383540; PMCID: PMC6307967.
3. Garudadri S, Woodruff PG, Han MK, Curtis JL, Barr RG, Bleecker ER, Bowler RP, Comellas A, Cooper CB, Criner G, Dransfield MT, Hansel NN, Paine R 3rd, Krishnan JA, Peters SP, Hastie AT, Martinez FJ, **O'Neal WK**, Couper DJ, Alexis NE, Christenson SA. 2019. Systemic markers of inflammation in smokers with symptoms despite preserved spirometry in SPIROMICS. *Chest*. 155(5):908-917. PMID: 30684474.
4. Gong J, Wang F, Xiao B, Panjwani N, Lin F, Keenan K, Avolio J, Esmaili M, Zhang L, He G, Soave D, Mastromatteo S, Baskurt Z, Kim S, **O'Neal WK**, Polineni D, Blackman SM, Corvol H, Cutting GR, Drumm M, Knowles MR, Rommens JM, Sun L, Strug LJ. 2019. Genetic association and transcriptome integration identify contributing genes and tissues at cystic fibrosis modifier loci. *PLoS Genet*. 15(2):e1008007. PMID: 30807572; PMCID: PMC6407791.
5. Okuda K, Chen G, Subramani DB, Wolf M, Gilmore RC, Kato T, Radicioni G, Kesimer M, Chua M, Livraghi-Burtrico A, Ehre C, Doerschuk CM, Randell SH, Matsui H, Nagase T, **O'Neal WK**, Boucher RC. 2019. Localization of secretory mucins MUC5AC and MUC5B in normal human airways. *Am J Respir Crit Care Med*. 199(6):715-727. PMID: 30352166; PMCID: PMC6423099.
6. Ash SY, Harmouche R, Putman RK, Ross JC, Martinez FJ, Choi AM, Bowler RP, Regan EA, Curtis JL, Han MK, Boucher RC, **O'Neal WK**, Hatabu H, Lynch DA, Rosas IO, Hunninghake GM, San Jose Estepar R, Washko GR; COPDGene Investigators. 2018. Association between acute respiratory disease events and the MUC5B promoter polymorphism in smokers. *Thorax*. 73(11):1071-1074. PMID: 29440587; PMCID: PMC6089672.
7. Burkes RM, Gassett AJ, Ceppe AS, Anderson W, **O'Neal WK**, Woodruff PG, Krishnan JA, Barr RG, Han MK, Martinez FJ, Comellas AP, Lambert AA, Kaufman JD, Dransfield MT, Wells JM, Kanner RE, Paine R 3rd, Bleecker ER, Paulin LM, Hansel NN, Drummond MB; SPIROMICS Investigators. 2018. Rural residence and COPD exacerbations: analysis of the SPIROMICS Cohort. *Ann Am Thorac Soc*. [Epub ahead of print]. PMID: 29584453; PMCID: PMC6207115.
8. Chen G, Volmer AS, Wilkinson KJ, Deng Y, Jones LC, Yu D, Bustamante-Marin XM, Burns KA, Grubb BR, **O'Neal WK**, Livraghi-Butrico A, Boucher RC. 2018. Role of Spdef in the regulation of Muc5b expression in the airways of naïve and muco-obstructed mice. *Am J Respir Cell Mol Biol*. 59(3):383-396. PMID: 29579396; PMCID: PMC6189647.
9. Kesimer M, Smith BM, Ceppe A, Ford AA, Anderson WH, Barr RG, **O'Neal WK**, Boucher RC; SPIROMICS Investigative Group. 2018. Mucin concentrations and peripheral airway obstruction in chronic obstructive pulmonary disease. *Am J Respir Crit Care Med*. 198(11):1453-1456. PMID: 30130124; PMCID: PMC6290944.
10. Livraghi-Butrico A, Wilkinson KJ, Volmer AS, Gilmore RC, Rogers TD, Caldwell RA, Burns KA, Esther CR Jr, Mall MA, Boucher RC, **O'Neal WK**, Grubb BR. 2018. Lung disease phenotypes caused by over-expression of combinations of alpha, beta, and gamma subunits of the epithelial sodium channel in mouse airways. *Am J Physiol Lung Cell Mol Physiol*. ajplung.00382.2017. 314(2):L318-L331. PMID: 29074490; PMCID: PMC5866504.
11. Martinez CH, Li SX, Hirzel AJ, Stolberg VR, Alexis NE, Barr RG, Bleecker ER, Carretta EE, Christenson SA, Cooper CB, Couper DJ, Doerschuk CM, Han MK, Hansel NN, Hastie AT, Hoffman EA, Kaner RJ, Martinez FJ, Meyers DA, **O'Neal WK**, Paine R 3rd, Putcha N, Rennard SI, Woodruff PG, Zeidler M, Curtis JL, Freeman CM; SPIROMICS Investigators. 2018. Alveolar eosinophilia in current smokers with chronic obstructive pulmonary disease in the SPIROMICS cohort. *J Allergy Clin Immunol*. 141(1):429-432. PMID: 28916185; PMCID: PMC5758424.

12. Morris MA, Jacobson SR, Kinney GL, Tashkin DP, Woodruff PG, Hoffman EA, Kanner RE, Cooper CB, Drummond MB, Barr RG, Oelsner EC, Make BJ, Han MK, Hansel NN, **O'Neal WK**, Bowler RP. 2018. Marijuana use associations with pulmonary symptoms and function in tobacco smokers enrolled in the Subpopulations and Intermediate Outcome Measures in COPD Study (SPIROMICS). *Chronic Obstr Pulm Dis.* 5(1):46-56. PMID: 29629404; PMCID: PMC5870739.
13. Polineni D, Dang H, Jones L, Gallins P, Pace R, Stonebraker J, Commander L, Krenicky J, Zhou Y, Corvol H, Cutting G, Drumm M, Strug L, Boyle M, Durie P, Chmiel J, Zou F, Wright F, **O'Neal W**, Knowles M. 2018. Airway mucosal host defense is key to genomic regulation of cystic fibrosis lung disease severity. *Am J Respir Crit Care Med.* 197(1):79-93. PMID: 28853905; PMCID: PMC5765386.
14. Putcha N, Fawzy A, Paul GG, Lambert AA, Psoter KJ, Sidhaye VK, Woo J, Wells JM, Labaki WW, Doerschuk CM, Kanner RE, Han MK, Martinez C, Paulin LM, Martinez FJ, Wise RA, **O'Neal WK**, Barr RG, Hansel NN; SPIROMICS investigators. 2018. Anemia and adverse outcomes in a chronic obstructive pulmonary disease population with a high burden of comorbidities. An analysis from SPIROMICS. *Ann Am Thorac Soc.* 15(6):710-717. PMID: 30726108; PMCID: PMC6207135.
15. Putcha N, Paul GG, Azar A, Wise RA, **O'Neal WK**, Dransfield MT, Woodruff PG, Curtis JL, Comellas AP, Drummond MB, Lambert AA, Paulin LM, Fawzy A, Kanner RE, Paine R 3rd, Han MK, Martinez FJ, Bowler RP, Barr RG, Hansel NN; SPIROMICS investigators. 2018. Lower serum IgA is associated with COPD exacerbation risk in SPIROMICS. *PLoS One.* 13(4):e0194924. PMID: 29649230; PMCID: PMC5896903.
16. Rowson-Hodel AR, Wald JH, Hatakeyama J, **O'Neal WK**, Stonebraker JR, VanderVorst K, Saldana MJ, Borowsky AD, Sweeney C, Carraway KL Rd. 2018. Membrane Mucin Muc4 promotes blood cell association with tumor cells and mediates efficient metastasis in a mouse model of breast cancer. *Oncogene.* 37(2):197-207. PMID: 28892049; PMCID: PMC5930013.
17. Saini Y, Lewis BW, Yu D, Dang H, Livraghi-Butrico A, Del Piero F, **O'Neal WK**, Boucher RC. 2018. Effect of LysM+ macrophage depletion on lung pathology in mice with chronic bronchitis. *Physiol Rep.* 6(8):e13677. PMID: 29667749; PMCID: PMC5904692.
18. Anderson WH, Ha JW, Couper DJ, **O'Neal WK**, Barr RG, Bleecker ER, Carretta EE, Cooper CB, Doerschuk CM, Drummond MB, Han MK, Hansel NN, Kim V, Kleerup EC, Martinez FJ, Rennard SI, Tashkin D, Woodruff PG, Paine R 3rd, Curtis JL, Kanner RE; SPIROMICS Research Group. 2017. Variability in objective and subjective measures affects baseline values in studies of patients with COPD. *PLoS One.* 12(9):e0184606. PMID: 28934249; PMCID: PMC5608200.
19. Bowler RP, Hansel NN, Jacobson S, Graham Barr R, Make BJ, Han MK, **O'Neal WK**, Oelsner EC, Casaburi R, Barjaktarevic I, Cooper C, Foreman M, Wise RA, DeMeo DL, Silverman EK, Bailey W, Harrington KF, Woodruff PG, Drummond MB; for COPDGene and SPIROMICS Investigators. 2017. Electronic cigarette use in US adults at risk for or with COPD: analysis from two observational cohorts. *J Gen Intern Med.* 32(12):1315-1322. PMID: 28884423; PMCID: PMC5698219.
20. Choi S, Haghighi B, Choi J, Hoffman EA, Comellas AP, Newell JD, Wenzel SE, Castro M, Fain SB, Jarjour NN, Schiebler ML, Barr RG, Han MK, Bleecker ER, Cooper CB, Couper D, Hansel N, Kanner RE, Kazeroni EA, Kleerup EAC, Martinez FJ, **O'Neal WK**, Woodruff PG, Lin CL. 2017. Differentiation of quantitative CT imaging phenotypes in asthma versus COPD. *BMJ Open Respir Res.* 4(1):e000252. PMID: 29435345; PMCID: PMC5687530.
21. Donoghue LJ*, Livraghi-Butrico A*, McFadden KM, Thomas JM, Chen G, Grubb BR, **O'Neal WK**, Boucher RC, Kelada SNP. 2017. Identification of trans protein QTL for secreted airway mucins in mice and a causal role for Bpifb1. *Genetics.* 207(2):801-12. PMID: 28851744; PMCID: PMC5629340.
22. Han MK, Quibrera PM, Carretta EE, Barr RG, Bleecker ER, Bowler RP, Cooper CB, Comellas A, Couper DJ, Curtis JL, Criner G, Dransfield MT, Hansel NN, Hoffman EA, Kanner RE, Krishnan JA, Martinez CH, Pirozzi CB, **O'Neal WK**, Rennard S, Tashkin DP, Wedzicha JA, Woodruff P, Paine R 3rd, Martinez FJ; SPIROMICS investigators. 2017. Frequency of exacerbations in patients with chronic obstructive pulmonary disease: an analysis of the SPIROMICS cohort. *Lancet Respir Med.* 5(8):619-26. PMID: 28668356; PMCID: PMC555885.
23. Hastie AT, Martinez FJ, Curtis JL, Doerschuk CM, Hansel NN, Christenson S, Putcha N, Ortega VE, Li X, Barr RG, Carretta EE, Couper DJ, Cooper CB, Hoffman EA, Kanner RE, Kleerup E, **O'Neal WK**, Paine R 3rd, Peters SP, Alexis NE, Woodruff PG, Han MK, Meyers DA, Bleecker ER; SPIROMICS investigators. 2017. Association of sputum and blood eosinophil concentrations with clinical measures of COPD

- severity: an analysis of the SPIROMICS cohort. *Lancet Respir Med*. 2017 Dec;5(12):956-967. PMID: 29146301; PMCID: PMC5849066.
24. Kesimer M, Ford AA, Ceppe A, Radicioni G, Cao R, Davis WC, Doerschuk CM, Alexis NE, Anderson WH, Henderson AG, Barr G, Bleecker ER, Christenson SA, Cooper CB, Han MLK, Hansel NN, Hastie AT, Hoffman EA, Kanner RE, Martinez F, Paine R, Woodruff PG, **O'Neal WK**, Boucher RC. 2017. Airway mucin concentration as a marker of chronic bronchitis. *N Engl J Med*. 377(10):911-22. PMID: 28877023; PMCID: PMC5706541.
 25. Livraghi-Butrico A, Grubb BR, Wilkinson K, Volmer AS, Burns KA, Evans C, **O'Neal WK***, Boucher RC*. 2017. Contribution of mucus concentration and secreted mucins Muc5ac and Muc5b to the pathogenesis of muco-obstructive lung disease. *Mucosal Immunol*. 10(2):395-407. PMID: 27435107; PMCID: PMC5250616. ***co-senior authors; equal contribution. Correction:** *Mucosal Immunol*. 2017;10(3):829. PMID: 28435155.
 26. Nishida K, Brune KA, Putcha N, Mandke P, **O'Neal WK**, Shade D, Srivastava V, Wang M, Lam H, An SS, Drummond MB, Hansel NN, Robinson DN, Sidhaye VK. 2017. Cigarette smoke disrupts monolayer integrity by altering epithelial cell-cell adhesion and cortical tension. *Am J Physiol Lung Cell Mol Physiol*. 313(3):L581-91. PMID: 28642260; PMCID: PMC5625260.
 27. **O'Neal WK**. 2017. Lung cell-specific Cre deleter mouse strains: going back to move forward. *Am J Respir Cell Mol Biol*. 57(2):149-50. PMID: 28762768.
 28. Dang H, Gallins PJ, Pace RG, Guo XL, Stonebraker JR, Corvol H, Cutting GR, Drumm ML, Strug LJ, Knowles MR, **O'Neal WK**. 2016. Novel variation at chr11p13 associated with cystic fibrosis lung disease severity. *Hum Genome Var*. Jul 7;3:16020. PMID: 27408752; PMCID: PMC4935763. **Corrigendum:** *Hum Genome Var*. 2017;4:17016. PMID: 28674633; PMCID: PMC5477836.
 29. Keene JD, Jacobson S, Kechris K, Kinney GL, Foreman MG, Doerschuk CM, Make BJ, Curtis JL, Rennard SI, Barr RG, Bleecker ER, Kanner RE, Kleeup EC, Hansel NN, Woodruff PG, Han MK, Paine III R, Martinez FJ, Bowler RP*, **O'Neal WK***; for COPDGene and SPIROMICS Investigators. 2016. Biomarkers predictive of exacerbations in the SPIROMICS and COPDGene Cohorts. *Am J Respir Crit Care Med*. 195(4):473-81. PMID: 27579823; PMCID: PMC5378424. ***co-senior authors; equal contribution**
 30. Saini Y, Wilkinson KJ, Terrell KA, Burns KA, Livraghi-Butrico A, Doerschuk CM, **O'Neal WK**, Boucher RC. 2016. Neonatal pulmonary macrophage depletion coupled to defective mucus clearance increases susceptibility to pneumonia and alters pulmonary immune responses. *Am J Respir Cell Mol Biol*. 54(2):210-21. PMID: 26121027; PMCID: PMC4821038.
 31. Sun W, Kechris K, Jacobson S, Drummond MB, Hawkins GA, Yang J, Chen TH, Quibrera PM, Anderson W, Barr RG, Basta PV, Bleecker ER, Beaty T, Casaburi R, Castaldi P, Cho MH, Comellas A, Crapo JD, Criner G, Demeo D, Christenson SA, Couper DJ, Curtis JL, Doerschuk CM, Freeman CM, Gouskova NA, Han MK, Hanania NA, Hansel NN, Hersh CP, Hoffman EA, Kaner RJ, Kanner RE, Kleeup EC, Lutz S, Martinez FJ, Meyers DA, Peters SP, Regan EA, Rennard SI, Scholand MB, Silverman EK, Woodruff PG, **O'Neal WK***, Bowler RP*; SPIROMICS Research Group; COPDGene Investigators. 2016. Common genetic polymorphisms influence blood biomarker measurements in COPD. *PLoS Genetics*. 12(8):e1006011. PMID: 27532455; PMCID: PMC4988780. ***co-senior/corresponding authors**
 32. Yu D, Davis RM, Aita M, Burns KA, Clapp PW, Gilmore RC, Chua M, **O'Neal WK**, Schlegel R, Randell SH, C Boucher R. 2016. Characterization of rat meibomian gland ion and fluid transport. *Invest Ophthalmol Vis Sci*. 57(4):2328-43. PMID: 27127933; PMCID: PMC4855829.
 33. Corvol H, Blackman SM, Boëlle PY, Gallins PJ, Pace RG, Stonebraker JR, Accurso FJ, Clement A, Collaco JM, Dang H, Dang AT, Franca A, Gong J, Guillot L, Keenan K, Li W, Lin F, Patrone MV, Raraigh KS, Sun L, Zhou YH, **O'Neal WK**, Sontag MK, Levy H, Durie PR, Rommens JM, Drumm ML, Wright FA, Strug LJ, Cutting GR, Knowles MR. 2015. Genome-wide association meta-analysis identifies five modifier loci of lung disease severity in cystic fibrosis. *Nat Commun*. 6:8382. PMID: 26417704; PMCID: PMC4589222.
 34. Freeman CM, Crudgington S, Stolberg VR, Brown JP, Sonstein J, Alexis NE, Doerschuk CM, Basta PV, Carretta EE, Couper DJ, Hastie AT, Kaner RJ, **O'Neal WK**, Paine R 3rd, Rennard SI, Shimbo D, Woodruff PG, Zeidler M, Curtis JL. 2015. Design of a multi-center immunophenotyping analysis of peripheral blood, sputum and bronchoalveolar lavage fluid in the Subpopulations and Intermediate

- Outcome Measures in COPD Study (SPIROMICS). *J Transl Med.* 13:19. PMID: 25622723; PMCID: PMC4314767.
35. Lubamba BA, Jones LC, **O'Neal WK**, Boucher RC, Ribeiro CM. 2015. X-box-binding protein 1 and innate immune responses of human cystic fibrosis alveolar macrophages. 2015. *Am J Respir Crit Care Med.* 192(12):1449-61. PMID: 26331676; PMCID: PMC4731720.
 36. **O'Neal WK**, Gallins P, Pace RG, Dang H, Wolf WE, Jones LC, Guo X, Zhou YH, Madar V, Huang J, Liang L, Moffatt MF, Cutting GR, Drumm ML, Rommens JM, Strug LJ, Sun W, Stonebraker JR, Wright FA, Knowles MR. 2015. Gene expression in transformed lymphocytes reveals variation in endomembrane and HLA pathways modifying cystic fibrosis pulmonary phenotypes. *Am J Hum Genet.* 96(2):318-28. PMID: 25640674; PMCID: PMC4320265.
 37. Bove PF, Dang H, Cheluvvaraju C, Jones LC, Liu X, **O'Neal WK**, Randell SH, Schlegel R, Boucher RC. 2014. Breaking the in vitro alveolar type II cell proliferation barrier while retaining ion transport properties. *Am J Respir Cell Mol Biol.* 50(4):767-76. PMID: 24191670; PMCID: PMC4068919.
 38. Carolan BJ, Hughes G, Morrow J, Hersh CP, **O'Neal WK**, Rennard S, Pillai SG, Belloni P, Cockayne DA, Comellas AP, Han M, Zemans RL, Kechris K, Bowler RP. 2014. The association of plasma biomarkers with computed tomography-assessed emphysema phenotypes. *Respir Res.* 15:127. PMID: 25306249; PMCID: PMC4198701.
 39. Fossum SL, Mutolo MJ, Yang R, Dang H, **O'Neal WK**, Knowles MR, Leir SH, Harris A. 2014. Ets homologous factor regulates pathways controlling response to injury in airway epithelial cells. *Nucleic Acids Res.* 42(22):13588-98. PMID: 25414352; PMCID: PMC4267623.
 40. Guo X, Pace RG, Stonebraker JR, **O'Neal WK**, Knowles MR. 2014. Meconium ileus in cystic fibrosis is not linked to central repetitive region length variation in MUC1, MUC2, and MUC5AC. *J Cyst Fibros.* 13(6):613-6. PMID: 24920497; PMCID: PMC4253052.
 41. Guo X, Zheng S, Dang H, Pace RG, Stonebraker JR, Jones CD, Boellmann F, Yuan G, Haridass P, Fedrigo O, Corcoran DL, Seibold MA, Ranade SS, Knowles MR, **O'Neal WK***, Voynow JA*. 2014. Genome reference and sequence variation in the large central repetitive central exon of human MUC5AC. *Am J Respir Cell Mol Biol.* 50:223-32. PMID: 24010879; PMCID: PMC3930937. ***Equal Co-Senior authors.**
 42. **O'Neal WK**, Anderson W, Basta PV, Carretta EE, Doerschuk CM, Barr RG, Bleecker ER, Christenson SA, Curtis JL, Han MK, Hansel NN, Kanner RE, Kleerup EC, Martinez FJ, Miller BE, Peters SP, Rennard SI, Scholand MB, Tal-Singer R, Woodruff PG, Couper DJ, Davis S. 2014. Comparison of serum, EDTA plasma and P100 plasma for luminex-based biomarker multiplex assays in patients with chronic obstructive pulmonary disease in the SPIROMICS study. *J Trans Med.* 12:9. PMID: 24397870; PMCID: PMC3928911.
 43. Roy MG, Livraghi-Butrico A, Fletcher AA, McElwee MM, Evans SE, Boerner RM, Alexander SN, Bellinghausen LK, Song AS, Petrova YM, Tuvim MJ, Adachi R, Romo I, Bordt AS, Bowden MG, Sisson JH, Woodruff PG, Thornton DJ, Rousseau K, De la Garza MM, Moghaddam SJ, Karmouty-Quintana H, Blackburn MR, Drouin SM, Davis CW, Terrell KA, Grubb BR, **O'Neal WK**, Flores SC, Cota-Gomez A, Lozupone CA, Donnelly JM, Watson AM, Hennessy CE, Keith RC, Yang IV, Barthel L, Henson PM, Janssen WJ, Schwartz DA, Boucher RC, Dickey BF, Evans CM. 2014. Muc5b is required for airway defence. *Nature.* 505:412-6. PMID: 24317696; PMCID: PMC4001806.
 44. Saini Y, Dang H, Livraghi-Butrico A, Kelly EJ, Jones LC, **O'Neal WK**, Boucher RC. 2014. Gene expression in whole lung and pulmonary macrophages reflects the dynamic pathology associated with airway surface dehydration. *BMC Genomics.* 15:726. PMID: 25204199; PMCID: PMC4247008.
 45. Livraghi-Butrico A, Kelly EJ, Wilkinson KJ, Rogers TD, Gilmore RC, Harkema JR, Randell SH, Boucher RC, **O'Neal WK**, Grubb BR. 2013. Loss of CFTR function exacerbates the phenotype of Na(+) hyperabsorption in murine airways. *Am J Physiol Lung Cell Mol Physiol.* 304:L469-480. PMID: 23377346; PMCID: PMC3627939.
 46. Martino MB, Jones L, Brighton B, Ehre C, Abdulah L, Davis CW, Ron D, **O'Neal WK**, Ribeiro CM. 2013. The ER stress transducer IRE1 β is required for airway epithelial mucin production. *Mucosal Immunol.* 6:639-54. PMID: 23168839; PMCID: PMC4031691.
 47. Sesma JI, Kreda SM, Okada SF, van Heusden C, Moussa L, Jones LC, **O'Neal WK**, Togawa N, Hiasa M, Moriyama Y, Lazarowski ER. 2013. Vesicular nucleotide transporter regulates the nucleotide content in

- airway epithelial mucin granules. *Am J Physiol Cell Physiol.* 304:C976-84. PMID: 23467297; PMCID: PMC3651637.
48. Ehre C, Worthington EN, Liesman RM, Grubb BR, Barbier D, **O'Neal WK**, Sallenave JM, Pickles RJ, Boucher RC. 2012. Overexpressing mouse model demonstrates the protective role of Muc5ac in the lungs. *Proc Natl Acad Sci U S A.* 109:16528-33. PMID: 23012413; PMCID: PMC3478656.
 49. Livraghi-Butrico A, Kelly EJ, Klem ER, Dang H, Wolfgang MC, Boucher RC, Randell SH, **O'Neal WK**. 2012. Mucus clearance, MyD88-dependent and MyD88-independent immunity modulate lung susceptibility to spontaneous bacterial infection and inflammation. *Mucosal Immunol.* 5:397-408. PMID: 22419116; PMCID: PMC3377774.
 50. Livraghi-Butrico A, Grubb BR, Kelly EJ, Wilkinson KJ, Yang H, Geiser M, Randell SH, Boucher RC, **O'Neal WK**. 2012. Genetically determined heterogeneity of lung disease in a mouse model of airway mucus obstruction. *Physiol Genomics.* 44:470-84. PMID: 22395316; PMCID: PMC3339860.
 51. Sun L, Rommens JM, Corvol H, Li W, Li X, Chiang TA, Lin F, Dorfman R, Busson, PF, Parekh RV, Zelenika D, Blackman SM, Corey M, Doshi VK, Henderson L, Naughton KM, **O'Neal WK**, Pace RG, Stonebraker JR, Wood SD, Wright FA, Zielenski J, Clement A, Drumm ML, Boelle PY, Cutting GR, Knowles MR, Durie PR, Strug LJ. 2012. Multiple apical plasma membrane constituents are associated with susceptibility to meconium ileus in individuals with cystic fibrosis. *Nat Genet.* 44a:562-9. PMID: 22466613; PMCID: PMC3371103.
 52. Grubb BR, **O'Neal WK**, Ostrowski LE, Kreda SM, Button B, Boucher RC. 2011. Transgenic hCFTR expression fails to correct β ENaC mouse lung disease. *Am J Physiol Lung Cell Mol Physiol.* 302:L238-47. PMID: 22003093; PMCID: PMC3349361.
 53. Guo X, Pace RG, Stonebraker JR, Commander CW, Dang AT, Drumm ML, Harris A, Zou F, Swallow DM, Wright FA, Knowles MR*, **O'Neal WK***. 2011. Mucin variable number tandem repeat polymorphisms and severity of cystic fibrosis lung disease: significant association with MUC5AC. *PLoS One.* 6:e25452. PMID: 21998660; PMCID: PMC3188583. * **Co-Senior authors.**
 54. Jones LC, Moussa L, Fulcher LM, Zhu Y, Hudson EJ, **O'Neal WK**, Randell SH, Lazarowski ER, Boucher RC, Kreda SM. 2011. VAMP8 is a vesicle SNARE that regulates mucin secretion in airway goblet cells. *J Physiol.* 590:545-62. PMID: 22144578; PMCID: PMC3379700.
 55. Nguyen Y, Procario MC, Ashley SL, **O'Neal WK**, Pickles RJ, Weinberg JB. 2011. Limited effects of Muc1 deficiency on mouse adenovirus type 1 respiratory infection. *Virus Res.* 160:351359. PMID: 21816184; PMCID: PMC3163747.
 56. Seminario-Vidal L, Okada SF, Sesma JI, Kreda SM, van Heusden CA, Zhu Y, Jones LC, **O'Neal WK**, Penuela S, Laird DW, Boucher RC, Lazarowski ER. 2011. Rho signaling regulates pannexin 1-mediated ATP release from airway epithelia. *J Biol Chem.* 286:26277-86. PMID: 21606493; PMCID: PMC3143590.
 57. Wright FA, Strug LJ, Doshi VK, Commander CW, Blackman SM, Sun L, Berthiaume Y, Cutler D, Cojocaru A, Collaco JM, Corey M, Dorfman R, Goddard K, Green D, Kent JW Jr, Lange EM, Lee S, Li W, Luo J, Mayhew GM, Naughton KM, Pace RG, Pare P, Rommens JM, Sandford A, Stonebraker JR, Sun W, Taylor C, Vanscoy LL, Zou F, Blangero J, Zielenski J, **O'Neal WK**, Drumm ML, Durie PR, Knowles MR, Cutting GR. 2011. Genome-wide association and linkage identify modifier loci of lung disease severity in cystic fibrosis at 11p13 and 20q13.2. *Nat Genet.* 43:539-46. PMID: 21602797; PMCID: PMC3296486.
 58. Bove P, Grubb BR, Okada SF, Ribeiro CM, Rogers TD, Randell SH, **O'Neal WK**, Boucher RC. 2010. Human alveolar type II cells secrete and absorb liquid in response to local nucleotide signaling. *J Biol Chem.* 285:34939-49. PMID: 20801871; PMCID: PMC2966108.
 59. Cholon DM, **O'Neal WK**, Randell SH, Riordan JR, Gentsch M. 2010. Modulation of endocytic trafficking and apical stability of CFTR in primary human airway epithelial cultures. *Am J Physiol Lung Cell Mol Physiol.* 298:L304-14. PMID: 20008117; PMCID: PMC2838667.
 60. Greenberg J, Deshmukh R, Huang L, Mostafa J, LaVange L, Carretta E, **O'Neal W**. 2010. The COPD Ontology and toward empowering clinical scientists as ontology engineers. *J Library Metadata.* 10:173-87.
 61. Kreda SM, Seminario-Vidal L, van Heusden CA, **O'Neal W**, Jones L, Boucher RC, Lazarowski ER. 2010. Receptor-promoted exocytosis of airway epithelial mucin granules containing a spectrum of adenine nucleotides. *J Physiol.* 588:2255-67. PMID: 20421285; PMCID: PMC2911224.

62. Mall MA, Button B, Johannesson B, Zhou Z, Livraghi A, Caldwell RA, Schubert SC, Schultz C, **O'Neal WK**, Pradervand S, Hummler E, Rossier BC, Brubb BR, Boucher RC. 2010. Airway surface liquid volume regulation determines different airway phenotypes in Liddle compared with beta ENaC-overexpressing mice. *J Biol Chem*. 285:26945-55. PMID: 20566636; PMCID: PMC2930694.
63. Ostrowski LE, Yin W, Rogers TD, Busalacchi KB, Chua M, **O'Neal WK**, Grubb BR. 2010. Conditional deletion of Dnaic1 in a murine model of primary ciliary dyskinesia causes chronic rhinosinusitis. *Am J Respir Cell Mol Biol*. 43:55-63. PMID: 19675306; PMCID: PMC2911571.
64. Kesimer M, Scull M, Brighton B, Demaria G, Burns K, **O'Neal W**, Pickles RJ, Sheehan JK. 2009. Characterization of exosome-like vesicles released from human tracheobronchial ciliated epithelium: a possible role in innate defense. *FASEB J*. 23:1858-68. PMID: 19190083; PMCID: PMC2698655.
65. Livraghi A, Grubb BR, Hudson EJ, Wilkinson KJ, Sheehan JK, Mall MA, **O'Neal WK**, Boucher RC, Randell SH. 2009. Airway and lung pathology due to mucosal surface dehydration in {beta}-epithelial Na⁺ channel overexpressing mice: role of TNF- α and IL-4R α signaling, influence of neonatal development, and limited efficacy of glucocorticoid treatment. *J Immunol*. 182:4357-67. PMID: 19299736; PMCID: PMC2659461.
66. Martino ME, Olsen JC, Fulcher NB, Wolfgang MC, **O'Neal WK**, Ribeiro CM. 2009. Airway epithelial inflammation-induced endoplasmic reticulum Ca²⁺ store expansion is mediated by X-box binding protein-1. *J Biol Chem*. 284:14904-13. PMID: 19321437; PMCID: PMC2685672.
67. Ribeiro CMP, Hurd H, Wu Y, Martino MEB, Jones L, Brighton B, Boucher RC, **O'Neal WK**. 2009. Azithromycin treatment alters gene expression in inflammatory, lipid metabolism, and cell cycle pathways in well-differentiated human airway epithelia. *PLoS ONE*. 4:e5806. PMID: 19503797; PMCID: PMC2688381.
68. Rock JR, **O'Neal WK**, Gabriel SE, Randell SH, Harfe BD, Boucher RC, Grubb BR. 2009. Transmembrane protein 16a (Tmem16a) Is A Ca⁺⁺ regulated Cl⁻ secretory channel In mouse airways. *J Biol Chem*. 284:14875-80. PMID: 19363029; PMCID: PMC2685669.
69. Seminario-Vidal L, Kreda S, Jones L, **O'Neal W**, Trejo J, Boucher RC, Lazarowski ER. 2009. Thrombin promotes release of ATP from lung epithelial cells through coordinated activation of Rho- and Ca²⁺-dependent signaling pathways. *J Biol Chem*. 284:20638-48. PMID: 19439413; PMCID: PMC2742828.
70. Sesma JI, Esther CR Jr, Kreda SM, Jones L, **O'Neal W**, Nishihara S, Nicholas RA, Lazarowski ER. 2009. ER/golgi nucleotide sugar transporters contribute to the cellular release of UDP-sugar signaling molecules. *J Biol Chem*. 284:12572-83. PMID: 19276090; PMCID: PMC2673323.
71. Mall MA, Harkema J, Trojanek JB, Treis D, Schubert S, Zhou Z, Hudson EJ, Kreda S, Livraghi A, **O'Neal WK**, Boucher RC. 2008. Development of chronic bronchitis and emphysema in β ENaC overexpressing mice. *Am J Respir Crit Care Med*. 177:730-42. PMID: 18079494; PMCID: PMC2277210.
72. Rowe SM, Jackson PL, Liu G, Hardison M, Livraghi A, Solomon GM, McQuaid DB, Noerager BD, Gaggari A, Clancy J, **O'Neal W**, Sorscher EJ, Abraham E, Blalock JE. 2008. Potential role of high mobility group box 1 in cystic fibrosis airway disease. *Am J Respir Crit Care Med*. 178:822-31. PMID: 18658107; PMCID: PMC2566793.
73. Kreda SM, Okada SF, van Heusden CA, **O'Neal W**, Gabriel S, Abdullah L, Davis CW, Boucher RC, Lazarowski ER. 2007. Coordinated release of nucleotides and mucin from human airway epithelial Calu-3 cells. *J Physiol*. 584:245-59. PMID: 17656429; PMCID: PMC2277076.
74. Ostrowski, LE, Yin W, Diggs PS, Rogers TD, **O'Neal, WK**, and Grubb, BR. 2007. Expression of CFTR from a ciliated cell-specific promoter is ineffective at correcting nasal potential difference in CF mice. *Gene Ther*. 14:1492-1501. PMID: 17637798.
75. Morin-Kensicki EM, Boone BN, Howell M, Stonebraker JR, Teed J, Alb JG, Magnuson TR, **O'Neal W**, Milgram SL. 2006. Defects in yolk sac vasculogenesis, chorioallantoic fusion, and embryonic axis elongation in mice with targeted disruption of yap65. *Mol Cell Biol*. 26:77-87. PMID: 16354681; PMCID: PMC1317614.
76. Barnes AP, Livera G, Huang P, Sun C, **O'Neal WK**, Conti M, Stutts MJ, Milgram SL. 2005. Phosphodiesterase 4D forms a cAMP diffusion barrier at the apical membrane of the airway epithelium. *J Biol Chem*. 280:7997-8003. PMID: 15611099.
77. Neuringer IP, Sloan J, Budd S, Chalermkulrat W, Park RCW, Stonebraker JR, **O'Neal WK**, Aris RM, Randell, SH. 2005. Calcineurin inhibitor effects on growth and phenotype of human airway epithelial cells in vitro. *Am J Transplantation*. 11:2660-70. PMID: 16212625.

78. Ribeiro CM, Paradiso AM, Schwab U, Perez-Vilar J, Jones L, **O'Neal WK**, Boucher RC. 2005. Chronic airway infection/inflammation induces a Ca²⁺ independent hyperinflammatory response in human cystic fibrosis airway epithelia. *J Biol Chem.* 280:17798-806. PMID: 15746099.
79. Mall M, Grubb BR, Harkema JR, **O'Neal WK**, Boucher RC. 2004. Increased airway epithelial Na⁺ absorption produces cystic fibrosis-like lung disease in mice. *Nat Med.*10:487-93. PMID: 15077107.
80. Okada SF, **O'Neal WK**, Huang P, Nicholas RA, Craigen WJ, Lazarowski ER, Boucher RC. 2004. A role of plasmalemmal voltage-dependent anion channel-1 (pI-VDAC-1) in ATP release and regulatory volume decrease in airway epithelial cells. *J Gen Physiol.*124:513-26. PMID: 15477379.
81. Stonebraker JR, Wagner D, Lefensty RW, Burns K, Gendler SJ, Bergelson JM, Boucher RC, **O'Neal WK**, Pickles RJ. 2004. Glycocalyx restricts adenoviral vector access to apical receptors expressed on respiratory epithelium in vitro and in vivo: role for tethered mucins as barriers to luminal infection. *J Virol.* 78: 13755-68. PMID: 15564484; PMCID: PMC533903.
82. Zariwala M*, **O'Neal WK***, Noone PG, Leigh MW, Knowles MR, Ostrowski LE. 2004. Investigation of the possible role of a novel gene, DPCD, in Primary Ciliary Dyskinesia. *Am J Respir Cell Mol Biol.* 30:428-34. PMID: 14630615. ***authors contributed equally to manuscript.**
83. Coakley RD, Grubb BR, Paradiso AM, Gatzky JT, Johnson LG, Kreda SM, **O'Neal WK**, Boucher RC. 2003. Abnormal surface liquid pH regulation by cultured cystic fibrosis bronchial epithelium. *Proc Natl Acad Sci U S A.* 100:16083-8. PMID: 14668433; PMCID: PMC307696.
84. Ostrowski LE, Hutchins JR, Zakel K, **O'Neal WK**. 2003. Targeting expression of a transgene to the airway surface epithelium using a ciliated cell-specific promoter. *Mol Ther.* 8:637-45. PMID: 14529837.
85. Morral N, **O'Neal WK**, Rice K, Leland M, Piedra PA, Aguilar-Cordova E, Carey KD, Beaudet AL, Langston C. 2002. Lethal toxicity, severe endothelial injury, and a threshold effect with high doses of an adenoviral vector in baboons. *Hum Gene Ther.* 13:143-54. PMID: 11779418.
86. Zhang YJ, **O'Neal WK**, Randell SH, Blackburn K, Moyer MB, Boucher RC, Ostrowski LE. 2002. Identification of dynein heavy chain 7 as an inner arm component of human cilia that is synthesized but not assembled in a case of primary ciliary dyskinesia. *J Biol Chem.* 17:17906-15. PMID: 11877439.
87. Lazarowski E, Rochelle LG, **O'Neal WK**, Ribeiro CP, Grubb BR, Harden TK, Boucher RC. 2001. Cloning and functional characterization of two murine uridine nucleotide receptors reveal a potential target for correcting ion transport deficiency in cystic fibrosis gallbladder. *J Pharm Exp Ther.* 297:43-9. PMID: 11259526.
88. **O'Neal WK**, Rose E, Velji R, Zhou H, Quinones J, Langston C, Rice K, Carey D, Beaudet AL. 2000. Multiple advantages of alpha-fetoprotein as a marker for in vivo gene transfer. *Mol Ther.* 6:640-8. PMID: 11124066.
89. **O'Neal WK**, Zhou H, Morral N, Langston C, Parks R, Graham F, Kochanek S, Beaudet AL. 2000. Toxicity associated with repeated administration of first-generation adenoviral vectors is not induced by a helper-dependent vector. *Mol Med.* 6:179-95. PMID: 10965494; PMCID: PMC1949942.
90. Morral N, **O'Neal WK**, Rice K, Leland M, Kaplan J, Piedra PA, Zhou H, Parks R, Velji R, Quinones J, Aguilar-Cordova E, Wadsworth S, Graham F, Kochanek S, Langston C, Carey KD, Beaudet AL. 1999. Administration of helper-dependent adenoviral vectors and sequential delivery of different vector serotype for long-term liver-directed gene transfer in baboons. *Proc Natl Acad Sci U S A.* 96:12816-21. PMID: 10536005; PMCID: PMC23112.
91. Morris AP, Ball JM, Zeng Q-Y, Scott JK, **O'Neal WK**, Estes MK. 1999. NSP4 elicits age-dependent diarrhea and Ca²⁺ mediated I⁻ efflux into intestinal crypts of CF mice. *Am J Physiol.* 40:G431-44. PMID: 10444458.
92. **O'Neal WK**, Zhou H, Morral N, Aguilar-Cordova E, Pestaner J, Langston C, Mull B, Wang Y, Beaudet AL, Lee B. 1998. Toxicological comparison of E2a-deleted and first-generation adenoviral vectors expressing alpha1-antitrypsin after systemic delivery. *Hum Gene Ther.* 9:1587-98. PMID: 9694157.
93. Morral N, **O'Neal WK**, Zhou H, Langston C, Beaudet AL. 1997. Immune responses to reporter proteins and high viral dose limit duration of expression with adenoviral vectors: comparison of E2a wild type and E2a deleted vectors. *Hum Gene Ther.* 8:1275-86. PMID: 9215744.
94. Zhou H, **O'Neal WK**, Morral N, Beaudet AL. 1997. Development of a complementing cell line and a system for construction of adenovirus vectors with E1 and E2a deleted. *J Virol.* 70:7030-8. PMID: 8794347.

95. Hasty P, **O'Neal WK**, Liu, K-Q, Morris AP, Bebok Z, Shumyatsky GB, Jilling T, Sorscher EJ, Bradley A, Beaudet AL. 1995. Severe phenotype in mice with termination mutation in Exon 2 of cystic fibrosis gene. *Somat Cell Mol Genet.* 21:177-87. PMID: 7482032.
96. **O'Neal WK**, Hasty EP, McCray PB Jr., Casey B, Rivera-Perez J, Welsh MJ, Beaudet AL, Bradley A. 1993. A severe phenotype in mice with a duplication of exon 3 in the cystic fibrosis gene. *Hum Mol Genet.* 2:1561-9. PMID: 7505691.
97. Yorifugi T, **Lemna (O'Neal) WK**, Ballard CF, Rosenbloom CL, Roxmahel R, Plavsic N, Tsui L-C, Beaudet, AL. 1991. Molecular cloning and sequence analysis of murine cDNA for the cystic fibrosis transmembrane conductance regulator. *Genomics.* 10:547-50. PMID: 1716243.
98. **Lemna (O'Neal) WK**, Feldman GL, Kerem B, Fernbach SD, Zevkovich EP, O'Brien WE, Riordan JR, Collins FS, Tsui, L-C, Beaudet, AL. 1990. Mutation analysis for heterozygote detection and the prenatal diagnosis of cystic fibrosis. *New Engl J Med.* 322:291-6. PMID: 2296270.

Published Reviews (from newest to oldest):

1. **O'Neal WK**, Knowles MR. 2018. Cystic fibrosis disease modifiers: complex genetics defines the phenotypic diversity in a monogenic disease. *Annu Rev Genomics Hum Genet.* 19:201-222. PMID: 29709203.
2. Ribeiro CM, **O'Neal WK**. 2012. Endoplasmic reticulum stress in chronic obstructive lung disease. *Curr Mol Med.* 12:872-82. PMID: 22697344.
3. Randell SH, Fulcher ML, **O'Neal WK**, Olsen JC. 2011. Primary epithelial cell models for cystic fibrosis research. *Methods Mol Biol.* 742:285-310. PMID: 21547740.
4. **O'Neal WK**, Beaudet AL. 1994. Somatic gene therapy for cystic fibrosis. *Hum Mol Genet.* 3:1497-502. PMID: 7849744.
5. Beaudet AL, Feldman GL, Kobayashi K, **Lemna (O'Neal) WK**, Fernbach SD, Knowles MR, Boucher RC, O'Brien WE. 1991. Mutation analysis for cystic fibrosis in a North American population. *Adv Exp Med Biol.* 290:53-4. PMID: 1950757.

Selected Abstracts (from newest to oldest):

1. Anderson WH, Quibrera M, **O'Neal WK**, Drummond MB, Alexis NE, Barjaktarevic I, Couper D, Bateman L, Paine R, Kaner RJ, Curtis JL, Woodruff P, Han MK, Martinez FJ, Barr RG, Hansel NN, Bleecker ER, Hoffman EA, Ortega VE, Doerschuk CM, Kim V, Dransfield MT, Bhatt SP, Bowler RP, Criner G, Cooper CB, Kanner RE, Boucher RC, Lazarowski E. 2019. Accelerated ATP hydrolysis in airway surface liquid (ASL) provides a mechanism for mucus dehydration in chronic bronchitis. *Am J Respir Crit Care Med.* 199:A3847.
2. Bates M, Brown JP, Freeman CM, Stolberg VR, Kady M, Wang Y, Murray S, Alexis NE, Barjaktarevic I, Barr RG, Bleecker ER, Bowler RP, Christenson S, Comellas AP, Cooper CB, Couper D, Criner GJ, Doerschuk CM, Han MK, Hansel NN, Hastie AT, Hoffman EA, Huang YJ, Kaner RJ, Krishnan JA, Martinez FJ, Meyers DA, **O'Neal WK**, Paine R, Peters SP, Putcha N, Wells JM, Wilson N, Woodruff P, Curtis JL. 2019. Altered alveolar macrophage expression of Axl and Mertk in COPD: SPIROMICS Immunophenotyping Sub-Study. *Am J Respir Crit Care Med.* 199:A4040.
3. Brehm CE, Genese F, Marion CR, Li X, **O'Neal WK**, Newell JD, Hawkins GA, Barr RG, Cooper CB, Couper D, Han MK, Hansel NN, Kanner RE, Kleerup EC, Martinez FJ, Paine R, Woodruff P, Peters SP, Hoffman EA, Bleecker ER, Meyers DA, Ortega VE. 2019. Pathogenic primary ciliary dyskinesia pathway variants associate with disease severity in at-risk smokers and COPD subjects from SPIROMICS. *Am J Respir Crit Care Med.* 199:A4858.
4. Esther CR, **O'Neal WK**, Anderson WH, Kesimer M, Hastie AT, Barr RG, Bowler RP, Wells JM, Oelsner E, Boucher RC. 2019. Impaired mucus hydration and hydration signaling pathways in COPD. *Am J Respir Crit Care Med.* 199:A1126.
5. Genese F, Newell JD, **O'Neal WK**, Marion CR, Li X, Hawkins GA, Brehm CE, Barjaktarevic I, Barr RG, Cooper CB, Couper D, Han MK, Hansel NN, Kanner RE, Kleerup EC, Martinez FJ, Paine R, Woodruff P, Hoffman EA, Peters SP, Meyers DA, Bleecker ER, Ortega VE. 2019. The rare PI Z variant associates

- with bronchiectasis in non-Hispanic White at-risk current and ex-smokers and COPD subjects from SPIROMICS. *Am J Respir Crit Care Med.* 199:A3849.
6. Huang YJ, Opron K, Erb-Downward J, Begley L, Alexis NE, Barjaktarevic I, Barr RG, Bleecker ER, Bowler RP, Christenson S, Comellas AP, Cooper CB, Couper D, Doerschuk CM, Dransfield MT, Han MK, Hansel NN, Hastie AT, Hoffman EA, Kaner RJ, Krishnan JA, **O'Neal WK**, Paine R, Peters SP, Wells JM, Woodruff P, Martinez FJ, Bates M, Freeman CM, Curtis JL, Huffnagle GB. 2019. Lung microbiota associations with clinical features of COPD in the SPIROMICS cohort. *Am J Respir Crit Care Med.* 199:A5578.
 7. Kelada SN, Li Y, Van Buren E, Radicioni G, Ford A, **O'Neal WK**, Ortega VE, Woodruff P, Bleecker ER, Boucher RC, Kesimer M. 2019. Genome-wide association study of sputum mucin concentrations in the Subpopulations and Intermediate Outcomes in COPD Study (SPIROMICS). *Am J Respir Crit Care Med.* 199:A2149.
 8. **O'Neal WK**, Putman RK, Wang JJ, Barjaktarevic I, Cooper CB, Cho MH, Couper DJ, Hansel NN, Hatabu H, Rosas IO, Nishino M, Ortega VE, Lynch DA, Woodruff PG, Barr RG, Curtis JL, Han MK, Bleecker ER, Kaner RJ, Paine R, Sidhaye VK, Silverman EK, Hoffman EA, Martinez FJ, Washko GR, Bowler RP, Hunninghake GM, Newell, JD; SPIROMICS (Spiromics); COPDgene. 2019. Distinct blood biomarkers associate with visual evidence of interstitial lung abnormality on HRCT in both SPIROMICS and COPDgene. *Am J Respir Crit Care Med.* 199:A5840.
 9. Ortega VE, **O'Neal WK**, Newell JD, Li X, Hawkins GA, Barjaktarevic I, Barr RG, Cooper CB, Couper D, Han MK, Hansel NN, Kanner RE, Kleerup EC, Martinez FJ, Paine R, Scholand M, Woodruff P, Peters SP, Hoffman EA, Meyers DA, Bleecker ER. 2019. Genome-wide association study identifies novel loci and recapitulates known loci for interstitial lung disease in non-Hispanic Whites from SPIROMICS. *Am J Respir Crit Care Med.* 199:A4861.
 10. Ortega VE, Li X, Ampleford E, **O'Neal WK**, Hawkins GA, Manichaikul A, Barjaktarevic I, Barr RG, Cooper CB, Couper D, Han MK, Hansel NN, Kanner RE, Kleerup EC, Martinez FJ, Paine R, Woodruff P, Hoffman EA, Peters SP, Bleecker ER, Meyers DA. 2019. Genome-wide association study of acute exacerbations of COPD identifies novel loci in SPIROMICS. *Am J Respir Crit Care Med.* 199:A4862.
 11. Putcha N, Dransfield MT, LaFon D, Woo J, Azar A, Fawzy A, Cooper CB, Bowler RP, Comellas AP, Krishnan JA, Han MK, Couper D, Peters SP, Drummond MB, **O'Neal WK**, Criner GJ, Martinez FJ, Curtis JL, Barr RG, Woodruff P, Hansel NN. 2019. BAL and serum immunoglobulin G levels are associated with risk for exacerbations, clinical and CT phenotypes, an analysis of SPIROMICS. *Am J Respir Crit Care Med.* 199:A1580.
 12. Radicioni G, Ford A, Ceppe A, Alexis NE, **O'Neal WK**, Boucher RC, Kesimer M, SPIROMICS Investigative Group. 2019. Cigarette smoke driven MUC5AC increase and airway obstructions in COPD. *Am J Respir Crit Care Med.* 199:A1127.
 13. Zhang W, Oromendia C, Kikkers SA, Kim K, **O'Neal WK**, Freeman CM, Christenson S, Peters SP, Wells JM, Doerschuk CM, Putcha N, Woodruff P, Cooper CB, Comellas AP, Criner GJ, Paine R, Hansel NN, Han MK, Han MK, Curtis JL, Kaner RJ, Ballman KV, Martinez FJ, Cloonan SM. 2019. Extracellular bronchoalveolar lavage fluid ferritin and risk for exacerbation in COPD: an analysis from the Subpopulations and Intermediate Outcome Measures in COPD Study (SPIROMICS). *Am J Respir Crit Care Med.* 199:A5369.
 14. Ash SY, Harmouche R, Putman RK, Ross JC, Martinez FJ, Choi AM, Bowler RP, Regan EA, Curtis JL, Han MK, Boucher RC, **O'Neal WK**, Hatabu H, Lynch DA, Rosas IO, Hunninghake GM, Estepar RS, Washko GR. 2018. Association between acute respiratory disease events and the MUC5B promoter polymorphism in smokers. *Am J Respir Crit Care Med.* 197:A2410.
 15. Barjaktarevic I, Hu S, Couper D, Anderson WH, Kanner RE, Paine R, Kaner RJ, Curtis JL, **O'Neal WK**, Woodruff P, Postow L, Han MK, Martinez FJ, Barr R, Wells JM, Ortega VE, Hoffman EA, Doerschuk CM, Kim V, Dransfield MT, Bhatt SP, Bhakta NR, Drummond MB, Bowler RP, Criner GJ, Martinez C, Tashkin D, Cooper CB. 2018. Bronchodilator responsiveness in COPD: results from SPIROMICS. *Am J Respir Crit Care Med.* 197:A6395.
 16. Bolourchi S, **O'Neal WK**, Kaner RJ, Doerschuk CM, Boucher RC, Criner GJ, Couper D, Bowler RP, Comellas AP, Dransfield MT, Krishnan JA, Barjaktarevic I, Paine R, Bleecker ER, Peters SP, Barr RG, Rennard SI, Raman SM, Hansel NN, Hoffman EA, Galban CJ, Curtis JL, Freeman CM, Martinez FJ, Han MK, Christenson S, Woodruff PG, Bhakta NR. 2018. Airway epithelial microRNAs associated with small

- airway dysfunction and Interleukin-17-driven inflammation in chronic obstructive pulmonary disease. *Am J Respir Crit Care Med.* 197:A7441.
17. Boucher RC, Ceppe AS, **O'Neal WK**, Ford A, Barr RG, Bleecker ER, Curtis JL, Cooper CB, Han MK, Hoffman EA, Martinez FJ, Paine R, Woodruff P, Smith BM, Kesimer M. 2018. Mucin-based metrics to quantitate and identify mucus component of peripheral airways obstruction in the SPIROMICS cohort. *Am J Respir Crit Care Med.* 197:A1209.
 18. Burkes RM, Ceppe AS, **O'Neal WK**, Han MK, Curtis JL, Comellas AP, Criner GJ, Krishnan JA, Christenson S, Barr RG, Cooper CB, Barjaktarevic I, Paine R, Ortega VE, Wise RA, Hansel NN, Drummond M. 2018. Low plasma cathelicidin levels are associated with reduced lung function and prior COPD exacerbations: an analysis of the SPIROMICS cohort. *Am J Respir Crit Care Med.* 197:A7442.
 19. Choi S, Haghighi B, Hoffman EA, Newell JD, Comellas AP, Barr R, Han MK, Cooper CB, **O'Neal WK**, Woodruff P, Martinez FJ, Bleecker ER, Bowler RP, Krishnan JA, Kanner RE, Paine R, Peters SP, Dransfield MT, Hansel NN, Criner GJ, Couper D, Bhatt SP, Lin C. 2018. QCT imaging-based clusters of former smokers identify clinical phenotypes in the SubPopulations and Intermediate Outcome Measures in COPD Study (SPIROMICS). *Am J Respir Crit Care Med.* 197:A6387.
 20. Freeman CM, Hoffman EA, Alikaj H, Stolberg VR, Alexis NE, Bleecker ER, Bowler RP, Christenson S, Comellas AP, Cooper CB, Couper D, Doerschuk CM, Han MK, Hansel NN, Hastie AT, Kaner RJ, Martinez FJ, Meyers DA, **O'Neal WK**, Paine R, Peters SP, Wells JM, Woodruff P, Barr RG, Curtis JL. 2018. Increased endothelial cell damage and decreased endothelial cell repair are implicated in emphysema pathogenesis: SPIROMICS Immunophenotyping Sub-Study. *Am J Respir Crit Care Med.* 197:A7439.
 21. Genese F, Ortega VE, Marion CR, **O'Neal WK**, Newell JD, Barr RG, Cooper CB, Couper D, Han MK, Hansel NN, Kanner RE, Martinez FJ, Meyers DA, Woodruff P, Peters SP, Hoffman EA, Bleecker ER. 2018. Risk factors and implications of bronchiectasis on COPD severity in the Subpopulations and Intermediate Outcome Measures in COPD Study (SPIROMICS). *Am J Respir Crit Care Med.* 197:A3544.
 22. Haghighi B, Choi S, Choi J, Li F, Hoffman EA, Comellas AP, Newell JD, Barr R, Han MK, Bleecker ER, Cooper CB, Couper D, Hansel NN, Kanner RE, Kazerooni E, Martinez FJ, **O'Neal WK**, Paine R, Rennard SI, Woodruff P, Lin C. 2018. Imaging-based cluster analysis using novel metrics in COPD for Sub-Phenotyping of Current Smokers: SPIROMICS. *Am J Respir Crit Care Med.* 197:A1033.
 23. Kraft J, Jeong S, Zhao H, Kesimer M, Boucher RC, Christenson S, Comellas AP, Bateman LA, Britt A, Doerschuk CM, Dransfield MT, Han MK, Paine R, Cooper CB, Huang YJ, Barjaktarevic I, Moore WC, Nguyen CP, Morris C, Crystal RG, Curtis JL, Hastie AT, Kaner RJ, **O'Neal WK**, Ortega VE, Peters SP, Postow L, Rennard SI, Woodruff P, Kim V. 2018. Current smoking with or without chronic bronchitis is independently associated with goblet cell hyperplasia in healthy smokers and COPD Subjects: an analysis of the SPIROMICS Cohort. *Am J Respir Crit Care Med.* 197:A2283.
 24. Labaki WW, Martinez CH, Freeman CM, Martinez FJ, Wells JM, Bhatt SP, Dransfield MT, Cooper CB, Putcha N, Hansel NN, Bleecker ER, Meyers DA, Pirozzi CS, Kanner RE, Paine R, Couper DJ, **O'Neal WK**, Barr RG, Woodruff PG, Ampleford EJ, Ortega VE, Curtis JL, Han MK. 2018. Higher levels of growth differentiation factor 15 (GDF-15) are associated with lower walking distance and exercise capacity in COPD: an analysis of the SPIROMICS cohort. *Am J Respir Crit Care Med.* 197:A3124.
 25. Manichaikul A, Nguyen JN, Christenson S, Tew W, Neighbors M, Garudadri S, Nerella S, Hoffman EA, Farber CR, Cho MH, **O'Neal WK**, Woodruff P, Wain LV, Rotter JI, London S, Im H, Rich SS, Barr RG. 2018. Genomic and transcriptomic analysis of pulmonary function and emphysema on CT scan. *Am J Respir Crit Care Med.* 197:A4187.
 26. Masters B, Han MK, Martinez FJ, Woodruff P, Criner GJ, Peters SP, Hansel NN, Comellas AP, Curtis JL, Paine R, **O'Neal WK**, Herrera JE, Miller M, Copeland K. 2018. Blood biomarker algorithms as an aide to describe exacerbations activity in symptomatic smokers and early staged COPD patients. *Am J Respir Crit Care Med.* 197:A2747.
 27. Okuda K, Chen G, Kato T, Wolf M, Gilmore R, Burns K, Chua M, Livraghi-Butrico A, Ehre C, Doerschuk CM, Randell SH, Matsui H, Nagase T, **O'Neal WK**, Boucher RC. 2018. Regional expression of secretory mucins MUC5AC and MUC5B in normal human airways. *Am J Respir Crit Care Med.* 197:A7636.
 28. Ortega VE, Hawkins GA, Li X, **O'Neal WK**, Manichaikul A, Barr R, Cooper CB, Couper D, Curtis JL, Han MK, Hansel NN, Kanner RE, Martinez FJ, Paine R, Wells JM, Woodruff P, Hoffman EA, Peters SP, Meyers DA, Bleecker ER. 2018. Comprehensive gene resequencing of SERPINA1 in SPIROMICS

- reveals novel rare loci for alpha 1-antitrypsin deficiency and emphysema. *Am J Respir Crit Care Med.* 197:A4184.
29. Putcha N, Fawzy A, Matsui E, Bowler RP, Woodruff P, **O'Neal WK**, Comellas AP, Han MK, Dransfield MT, Lugogo N, Hoffman EA, Cooper CB, Hersh CP, Paulin LM, Drummond M, Wise RA, Diette GB, Hansel NN. 2018. Allergen sensitization and exposure is associated with exacerbations in COPD. *Am J Respir Crit Care Med.* 197:A2750.
 30. Dang H, Polineni D, Pace RG, Stonebraker JR, Li Q, Corvol H, Cutting GR, Drumm ML, Strug LJ, **O'Neal W**, Knowles M. 2017. Identification of novel CF modifier genes and pathways by expression imputation from GWAS. *Pediatr Pulmonol.* 52:S263.
 31. Esther CR, Polineni D, Mahon A, Isaacman S, Bonfield TL, Knowles M, **O'Neal W**. 2017. A novel therapeutic targeting the methionine salvage pathway reduces airway inflammation. *Pediatr Pulmonol.* 52:S328-S329.
 32. Mok H, Freeman CM, Martinez CH, Stolberg VR, Alexis NE, Barr RG, Bleecker ER, Bowler RP, Carretta EE, Christenson SA, Cooper CB, Couper DJ, Doerschuk CM, Han MK, Hansel NN, Hastie AT, Hoffman EA, Kaner RJ, Martinez FJ, Meyers DA, **O'Neal WK**, Paine R, Putcha N, Rennard SI, Woodruff PG, Curtis JL. 2017. Active smoking is associated with decreased Cd4+T cells in bronchoalveolar lavage: Spiromics Immunophenotyping Sub-Study. *Am J Respir Crit Care Med.* 195:A2629.
 33. Okuda K, Chen G, Wolf M, Burns K, Chua M, Livraghi-Butrico A, Ehre C, Doerschuk C, Randell S, **O'Neal W**, Boucher RC. 2017. Localization of secretory mucins MUC5AC and MUC5B in normal human airways. *Pediatr Pulmonol.* 52:S238-S239.
 34. Okuda K, Chen G, Wolf M, Burns K, Chua M, Livraghi-Butrico A, Ehre C, Doerschuk C, Randell S, **O'Neal W**, Boucher RC. 2017. Localization of secretory mucins MUC5AC and MUC5B in normal human airways. *Pediatr Pulmonol.* 52:S238-S239.
 35. Polineni D, Dang H, Gallins P, Jones L, Pace RG, Stonebraker JR, Corvol H, Cutting GR, Drumm ML, Strug LJ, Boyle MP, Durie P, Chmiel J, Zhou Y, Zou F, Wright F, **O'Neal W**, Knowles M. 2017. Nasal transcriptomic studies combined with genomic analyses identify gene networks associated with cystic fibrosis lung disease severity. *Pediatr Pulmonol.* 52:S263-S264.
 36. Stonebraker JR, Dang H, Pace RG, Boyles S, Quinney N, Cholon DM, Harris A, Li Q, Randell S, Gentzsch M, Knowles M, **O'Neal W**. 2017. EXPLORING the biologic basis of the chr11p13 CF lung disease modifying locus using CF airway epithelial cells. *Pediatr Pulmonol.* 52:S265.
 37. Subramani DB, Shenoy SK, Wang B, Markovetz MR, Chen G, Radicioni G, Haridass P, Jones L, Garbarine IC, Winkler SS, Sears PR, Ostrowski LE, Livraghi-Butrico A, Esther CR, Hill DB, **O'Neal W**, Kesimer M, Button B, Boucher RC, Ehre C. 2017. Adhesive, cohesive and viscoelastic properties of MUC5AC play a role in CF pathogenesis. *Pediatr Pulmonol.* 52:S279.
 38. Wells JM, Parker M, Osteri RA, Bowler RP, Dransfield MT, Cho MH, Woodruff PG, Kim V, Curtis JL, Martinez FJ, Paine R, Barri RG, Han MK, **O'Neal WK**, Castaldi PJ, Gaggari A. 2017. Elevated matrix metalloproteinase 9 in moderate to severe COPD: results from Spiromics and Eclipse. *Am J Respir Crit Care Med.* 195: A2732.
 39. Ancy KM, Leidy NK, Malley KG, Anderson WH, Barr RG, Bleeker E, Bowler RP, Carretta EE, Cooper CB, Couper DJ, Doerschuk CM, Dransfield MT, Hansel NN, Hoffman EA, Kanner R, **O'Neal WK**, Paine R, Peters SP, Scholand M, Woodruff PG, Han MK, Martinez FJ. 2016. How 'stable' is stable COPD? Daily symptom variability of subjects enrolled in the SPIROMICS Exacerbation Sub-Study. *Am J Respir Crit Care Med.* 193:A3527.
 40. Anderson WH, **O'Neal WK**, Doerschuk CM, Carretta EE, Couper DJ, Tashkin D, Paine, III R, Cooper CB, Bleecker ER, Barr RG, Hansel NN, Han MK, Martinez FJ, Curtis JL, Woodruff PG, Kleerup E, Kanner R. 2016. Short-term stability of pulmonary function and clinical measures in COPD using a cohort from Spiromics (Subpopulations and Intermediate Outcome Measures in COPD Study). *Am J Respir Crit Care Med.* 193:A3515.
 41. Barjaktarevic I, Martinez CH, Curtis JL, Bowler RP, **O'Neal WK**, Hoffman EA, Carretta EE, Quibrera M, Barr RG, Bleecker E, Couper DJ, Criner GJ, Dransfield MT, Han MK, Hansel NN, Kanner R, Lazarus SC, Martinez FJ, Paine R, Tashkin DP, Woodruff PG, Cooper CB. 2016. Biomarker characterization of a debilitated phenotype in Spiromics. *Am J Respir Crit Care Med.* 193: A1426.
 42. Barjaktarevic I, Martinez CH, Curtis JL, Hoffman EA, Carretta EE, Bowler RP, **O'Neal WK**, Quibrera M, Barr RG, Martinez FJ, Couper DJ, Criner GJ, Dransfield MT, Han MK, Hansel NN, Kanner R, Lazarus

- SC, Bleecker ER, Paine R, Tashkin DP, Woodruff PG, Cooper CB. 2016 Identification of a debilitated phenotype in Spiromics. *Am J Respir Crit Care Med.* 193:A3519.
43. Brown JP, Freeman CM, Crudgington SW, Stolberg VR, Sonstein J, Alexis NE, Basta PV, Carretta EE, Christenson S, Couper DJ, Doerschuk CM, Hansel NN, Han MK, Hastie AT, Kaner RJ, Martinez FJ, **O'Neal WK**, Paine R, Rennard SI, Shimbo D, Woodruff PG, Zeidler M, Curtis JL. 2016. Current smoking increases macrophage numbers and immunoregulatory receptor expression in bronchoalveolar lavage but not sputum in chronic obstructive pulmonary disease: Spiromics Immunophenotyping Sub-Study. *Am J Respir Crit Care Med.* 193:A1432.
 44. Chen G, **O'Neal WK**, Deng Y, Yu D, Jones L, Ribeiro C, Randell SH, Boucher RC. 2016. Il1r1 regulates Muc5b production and inflammatory responses in the airway epithelium. *Am J Respir Crit Care Med.* 193:A5559.
 45. Chen G, Volmer A, Wilkinson K, Terrell K, **O'Neal WK**, Livraghi-Butrico A, Boucher RC. 2016. Spdef-independent mucin production during non-Th2-dominated pulmonary inflammation revealed in the Scnn1b-Tg muco-obstructive mouse model. *Am J Respir Crit Care Med.* 193:A5558.
 46. Ford AA, Radicioni G, Cao R, Ceppe A, Doerschuk CM, **O'Neal WK**, Anderson WH, Boucher RC, Kesimer M. 2016. Mucin hypersecretion associated with chronic bronchitis and not emphysema in sputum from COPD patients from the SPIROMICS Cohort. *Am J Respir Crit Care Med.* 193:A3518.
 47. Haghighi B, Choi S, Hoffman EA, Newell J, Barr RG, Han MK, Cooper CB, **O'Neal WK**, Woodruff PG, Bleecker ER, Martinez FJ, Lin CL. 2016. Multiscale imaging-based clusters in the COPD Cohort associates with clinical characteristics: the Subpopulations and Intermediate Outcome Measures in COPD Study (SPIROMICS). *Am J Respir Crit Care Med.* 193:A6612.
 48. Kelada S, Livraghi-Butrico A, McFadden K, Thomas J, **O'Neal WK**, Boucher RC. 2016. Quantitative genetic analysis of MUC5AC and MUC5B in a mouse model of asthma. *Am J Respir Crit Care Med.* 193:A2916.
 49. Li SX, Hirzel AJ, Freeman CM, Crudgington SW, Stolberg VR, Sonstein J, Alexis NE, Basta PV, Carretta EE, Christenson SA, Couper DJ, Doerschuk CM, Han MK, Hansel NN, Hastie AT, Kaner RJ, Martinez FJ, **O'Neal WK**, Paine R, Rennard SI, Shimbo D, Woodruff PG, Zeidler M, Curtis JL. 2016. Active smoking is associated with increased eosinophils in bronchoalveolar lavage but not in sputum or peripheral blood: SPIROMICS Immunophenotyping Sub-Study. *Am J Respir Crit Care Med.* 193:A6345.
 50. Livraghi-Butrico A, Grubb B, Ehre C, Ostrowski LF, Button B, Rogers T, Wilkinson K, Samir S, Villalon DG, Hale A, **O'Neal WK**, Thelin W, Boucher RC. 2016. Scnn1b-TG mice: a unique model to test airway mucus-mobilizing therapies in vivo. *Pediatr Pulmonol.* 51:284-5.
 51. Martinez CH, Freeman CM, Nelson JD, Anderson WH, Bowler RP, Cooper CB, Couper DJ, Han MK, Martinez FJ, **O'Neal WK**, Ortega VE, Paine R, Woodruff PG, Curtis JL. 2016. Chronic bronchitis and airway thickening are associated with higher levels of growth differentiation factor-15 (GDF-15) in smokers without airflow obstruction: an analysis of the SPIROMICS Cohort. *Am J Respir Crit Care Med.* 193:A3510.
 52. Ortega VE, Li X, **O'Neal WK**, Pennington EJ, Barr RG, Couper DJ, Han MK, Kanner R, Kleerup E, Martinez FJ, Peters SP, Rennard SI, Woodruff PG, Hoffman EA, Meyers DA, Bleecker ER. 2016. Rare *SERPINA1* variants in the multi-ethnic Spiromics population. *Am J Respir Crit Care Med.* 193:A4433.
 53. Polineni D, Dang H, Pace RG, Stonebraker JR, Guo XG, Jones LC, Esther CR, Boyle MP, Chmiel JF, Durie PR, Gallins PJ, Wright FA, **O'Neal WK**, Knowles M. 2016. In vivo cystic fibrosis (CF) nasal epithelial transcriptomic studies reveal non-CFTR differential gene expression associated with CF Lung disease severity. *Am J Respir Crit Care Med.* 193:A6458.
 54. Polineni D, Dang H, Pace R, Stonebraker JR, Jones L, Boyle MP, Chmiel J, Durie PR, Gallins, p C., Wright F, **O'Neal WK**, Knowles MK. 2016. Age of onset of chronic *P. aeruginosa* pulmonary infection is associated with differential gene expression in CF nasal epithelia in vivo. *Pediatr Pulmonol.* 51 Suppl 45:244.
 55. Taylor J, Degtiar I, Murray S, Wang X, Martinez CH, Erb-Downward JR, Marchetti N, Criner GJ, **O'Neal WK**, Couper DJ, Bowler RP, Scholand M, Pirozzi C, Cooper CB, Kanner R, Paine R, Barr RG, Bleecker E, Hansel NN, Woodruff PG, Hoffman EA, Galban C, Ross BD, Kazerooni E, Martinez FJ, Han MK. 2016. Association between blood biomarkers, CT phenotype and COPD stage in SPIROMICS. *Am J Respir Crit Care Med.* 193:A4066.
 56. Tew W, Chung C, Manser P, Holweg C, Christenson SA, Rosenberger C, **O'Neal WK**, Woodruff PG, Neighbors M. 2016. Serum type 2 chemokines in chronic obstructive pulmonary disease (COPD) subjects in the Subpopulations and Intermediate Outcomes in COPD Study (SPIROMICS). *Am J Respir Crit Care Med.* 193:A7873.

57. Anderson WH, Basta P, Carretta E, Cui G, Davis S, Doerschuk CM, Barr RG, Bleecker ER, Bowler RP, Curtis JL, Drummond MB, Han MK, Hansel NN, Kanner RE, Kleeerup EC, Martinez FJ, Paine R, Peters SP, Woodruff PG, Rennard SI, Couper D, **O'Neal WK**. 2015. Reliability of multiplex-based blood biomarkers in COPD subjects enrolled in Spiromics (Subpopulation and Intermediate Outcome Measures in COPD Study). *Am J Respir Crit Care Med*. 191:A2935.
58. Brown JP, Freeman CM, Crudgington S, Stolberg VR, Sonstein J, Alexis NE, Basta P, Carretta E, Couper D, Doerschuk CM, Hastie AT, Kaner RJ, **O'Neal WK**, Paine R, Rennard SI, Shimbo D, Woodruff PG, Zeidler MR, Curtis JL. 2015. Monocytoid macrophages comprise an increased percentage of both mononuclear phagocytes and all leukocytes in bronchoalveolar lavages of smokers: SPIROMICS Immunophenotyping Sub-Study. *Am J Respir Crit Care Med*. 191:A2930.
59. Carlson TD, Kinney GL, Woodruff PG, Hansel NN, Han MK, **O'Neal WK**, Tashkin DP, Bowler RP. 2015. The significance and clinical impact of marijuana use in the SPIROMICS Cohort. *Am J Respir Crit Care Med*. 191:A4487.
60. Dang H, Gallins PJ, Rhonda PG, Stonebraker JR, Fossum SL, Leir S, Harris A, **O'Neal WK**, Knowles MR. 2015. Re-sequencing GWAS region on chromosome 11 identified novel candidate genetic variants associated with modification of CF lung disease. *Pediatr Pulmonol*. 50 Suppl 41:250.
61. Fossum SL, Mutolo MJ, Yang R, Dang H, **O'Neal WK**, Knowles MR, Leir S, Harris A. 2015. Ets homologous factor regulates pathways controlling response to injury in the airway epithelial cells. *Pediatr Pulmonol*. 50 Suppl 41:253.
62. Gupta R, Abdelwahab S, Jones L, Dang H, **O'Neal WK**, Kesimer M. 2015. Intercellular communication between the airway epithelial cells is mediated by exosomes like vesicles. *Am J Respir Crit Care Med*. 191:A3660.
63. Jacobson S, Kechris K, Sun W, Yang J, Chen TH, Barr RG, Basta P, Bleecker ER, Couper D, Curtis JL, Doerschuk CM, Drummond MB, Han MK, Hansel NN, Hawkins G, Hoffman EA, Kanner R, Kleeerup EC, Martinez FJ, Meyers DA, **O'Neal WK**, Peters SP, Rennard SI, Woodruff PG, Bowler RP. 2015. Blood biomarker quantitative trait loci in chronic obstructive pulmonary disease. *Am J Respir Crit Care Med*. 191:A3627.
64. Keene J, Curtis JL, Doerschuk CM, Kinney GL, Foreman MG, Black-Shinn JL, Han MK, Kechris K, Woodruff PG, **O'Neal WK**, Barr RG, Rennard SI, Bleecker ER, Kleeerup EC, Kanner R, Hansel NN, Jacobson S, Martinez FJ, Bowler RP. 2015. Blood biomarkers of exacerbations in the Spiromics and COPDgene Cohorts. *Am J Respir Crit Care Med*. 191:A5118.
65. Nguyen T, Chung F, **O'Neal WK**, Jones L, Gentsch M, Boucher R, Kreda SM. 2015. The effect of CFTR activity, bicarbonate, and pH on airway mucin secretion. *Pediatr Pulmonol*. 50 Suppl 41:214-5.
66. Polineni D, Esther CR, Pace RG, Stonebraker JR, Jones LC, Guo X, Boyle MP, Chmiel J, Durie PR, **O'Neal WK**, Knowles MR. 2015. In vivo human nasal lavage metabolomics suggest biomarkers of lung disease severity in cystic fibrosis. *Pediatr Pulmonol*. 50 Suppl 41:277.
67. Sesma JI, Livraghi-Butrico A, Wilkinson KJ, Weitzer C, Saini Y, Harden TK, **O'Neal WK**, Lazarowski ER. 2015. Blockage of the P2y14-receptor inhibits neutrophil infiltration in chronic lung diseases. *Am J Respir Crit Care Med*. 191:A6144.
68. Bailey KL, Han MK, Sayles HR, Barr RG, Doerschuk CM, Peters SP, Putcha N, Hansel NN, Woodruff PG, **O'Neal WK**, Couper D, Rennard SI. 2014. Patterns of biomarker expression in common comorbidities of COPD in the Spiromics population. *Am J Respir Crit Care Med*. 189:A5877.
69. Chen G, Martino ME, **O'Neal WK**, Boucher RC, Ribeiro CM. 2014. Role of the UPR in airway epithelial mucin production relevant to cystic fibrosis. *Pediatr Pulmonol*. 49 Suppl 38:255-6.
70. Duncan EA, Burns EN, Dang H, Jania CM, Tilley SL, Boucher RC, **O'Neal WK**, Doerschuk CM. 2014. Tobacco smoke causes oxidative stress and alterations in immune regulation in a murine model of chronic obstructive pulmonary disease. *Am J Respir Crit Care Med*. 189:A4289.
71. Han MK, Wang X, Murray S, **O'Neal WK**, Doerschuk CM, Anderson WH, Basta P, Couper D, Bowler RP, Weir M, Scholand MB, Pirozzi CS, Kleeerup E, Kanner RE, Barr G, Bleecker ER, Martinez FJ, Woodruff PG, Hoffman EA. 2014. Blood biomarkers and CT phenotypes in the Subpopulations and Intermediate Outcome Measures in COPD Study (Spiromics). *Am J Respir Crit Care Med*. 189:A5915.
72. Nguyen T, Jones L, **O'Neal WK**, Boucher RC, Kreda SM. 2014. Airway mucin secretion is affected only indirectly by CFTR activity. *Pediatr Pulmonol*. 49 Suppl 38:265.
73. **O'Neal WK**, Anderson WH, Bowler R, Peters SP, Couper D, Rennard SI. 2014. Persistent systemic inflammation in the Spiromics COPD population. *Am J Respir Crit Care Med*. 189:A5918.
74. **O'Neal WK**, Wolf W, Jones LC, Gallins PJ, Pace RG, Stonebraker JR, Dang H, Wright F, Knowles MR. 2014. Differential gene expression identifies genetic modifiers associated with lung disease severity and

- age of onset of persistent pseudomonas infection in cystic fibrosis. *Am J Respir Crit Care Med*. 189:A6717.
75. Pirozzi CS, Scholand MB, Han MK, Cui G, Carretta E, **O'Neal WK**, Barr RG, Paine R, Kanner R. 2014. Low and high body mass index are associated with specific COPD phenotypes. *Am J Respir Crit Care Med*. 189:A5936.
 76. Polineni D, Dang H, Gallins PJ, Pace RG, Stonebraker JR, Jones LC, Guo X, Boyle MP, Chmiel JF, Drumm ML, Durie PR, Wright FA, **O'Neal WK**, Knowles MR. 2014. In vivo human nasal epithelia inform transcriptomic studies in cystic fibrosis (CF) complementary to genomic variation studies. *Pediatr Pulmonol*. 49 Suppl 38:272.
 77. Polineni D, Dang H, Pace RG, Stonebraker JR, Guo XL, Jones LC, Boyle MP, Chmiel J, Drumm M, Durie PR, Gallins PJ, Wright FA, **O'Neal WK**, Knowles MR. 2014. Correlation of inflammatory markers in human nasal epithelia in vivo informs transcriptomic studies in cystic fibrosis (CF) complementary to genomic variation studies. *Am J Respir Crit Care Med*. 189:A5519.
 78. Terryah S, Wilkinson K, **O'Neal WK**, Taylor J, Tarran R. 2014. A SPLUNC1-derived peptide reduces lung disease in SCNN1B mice. *Pediatr Pulmonol*. 49 Suppl 38:285-6.
 79. Bove PF, Cheluvvaraju C, Dang H, Jones LC, Liu X, **O'Neal WK**, Randell SH, Schlegel R, Boucher RC. 2013. Alveolar type 2 cells co-cultured with fibroblasts and a rho kinase inhibitor promotes proliferation and maintains cell specific ion/liquid transport properties. *Am J Respir Crit Care Med*. 187:A3399.
 80. Doerschuk CM, Carretta E, **O'Neal WK**, Anderson WH, Couper D, Curtis JL, Woodruff PG, Rennard SI, Barr G. 2013. Circulating leukocytes, erythrocytes, and platelets in SPIROMICS. *Am J Respir Crit Care Med*. 187:A1513.
 81. Duncan EA, Burns E, Prabhu S, Livraghi-Butrico A, Hua X, Dang H, Tilley SL, Boucher RC, **O'Neal WK**, Doerschuk CM. 2013. The role of airway surface dehydration and tobacco smoke in the pathogenesis of chronic obstructive pulmonary disease. *Am J Respir Crit Care Med* 187:A4086.
 82. Fletcher A, Livraghi-Butrico A, **O'Neal WK**, Grubb BR, Boucher RC, Evans C. 2013. Required roles for Muc5B, but not Muc5ac in mucociliary clearance and anti-bacterial defenses in the lungs. *Pediatr Pulmonol*. 48 Suppl 36:451.
 83. Kreda SM, Moussa L, Nguyen T, Jones LC, Wilkinson KJ, Kelly EJ, **O'Neal WK**, Lazarowski ER, Boucher RC. 2013. Mucin secretion inhibition in vivo reduces air way mucus obstruction, infection, and Inflammation. *Pediatr Pulmonol*. 48 Suppl 36:238.
 84. **O'Neal WK**, Wolf WE, Jones LC, Mayhew GM, Gallins PJ, Pace RG, Stonebraker JR, Dang H, Zhou Y, Sun W, Zou F, Wright FA, Knowles MR. 2013. Differential gene expression identifies CF genetic modifiers associated with severity of lung disease. *Pediatr Pulmonol*. 48 Suppl 36:264.
 85. Polineni D, Guo X, Jones LC, Patrone MV, Pace RG, Stonebraker JR, Boyle MP, Chmiel JF, Durie PR, Drumm ML, **O'Neal WK**, Knowles MR. 2013. Transforming growth factor beta 1 (TGFB1) expression in vivo is correlated with interleukin 8 (IL8) expression in CF airway (nasal) epithelia. *Pediatr Pulmonol*. 48 Suppl 36:267.
 86. Button B, Cai L, **O'Neal WK**, Ehre C, Rubinstein M, Boucher RC. 2012. The role of cell surface-attached mucins in airway health. *Pediatr Pulmonol*. 47 Suppl 35:252.
 87. Dang H, Guo X, Polineni D, Jones LC, Pace RG, Knowles MR, **O'Neal WK**. 2012. MUC5AC reference genomic and mRNA structures from high-throughput sequencing data. *Pediatr Pulmonol*. 47 Suppl 35:282.
 88. Kreda SM, Jones L, Moussa L, Fulcher L, Yunxiang Z, Hudson E, **O'Neal WK**, Randell SH, Lazarowski ER, Boucher RC. 2012. VAMP8 is the vesicle SNARE for mucin secretion in airway goblet cells. American Thoracic Society, San Francisco CA. *Am J Respir Crit Care Med*. 185: A6299.
 89. Kreda SM, Moussa L, Jones L, Clapp P, **O'Neal WK**, Lazarowski ER, Boucher RC. 2012. Rock signaling is an important mediator of airway goblet cell mucin secretion. *Pediatr Pulmonol*. 47 Suppl 35:256.
 90. Livraghi-Butrico A, Cao R, Kelly EJ, Evans CM, Boucher RC, **O'Neal WK**, Kesimer M. 2012. Airway mucin interactome composition in response to defective mucus clearance and genetic deletion of secreted mucins. *Pediatr, Pulmonol*. 47 Suppl 35:253.
 91. Livraghi-Butrico A, Duncan EA, Saini Y, Dang H, Doerschuk CM, **O'Neal WK**, Boucher RC. 2012. Mechanisms of host defense during mucus obstruction in *Scnn1b* transgenic mice, a mouse model of COPD. 54th Thomas L. Petty Aspen Lung Conference, "COPD and Lung Cancer. A Continuum of Airway Disease", Aurora, CO. *Proc Am Thorac Soc*. 9(2):82-3.
 92. Saini Y, Dang H, Livraghi-Butrico A, **O'Neal WK**, Boucher RC. 2012. Macrophage activation patterns and development of airway inflammation in *Scnn1b*-transgenic mice. *Pediatr Pulmonol*. 47 Suppl 35:229.

93. Guo X, Dang H, Jones LC, Dang AT, **O'Neal WK**, Knowles MR. 2011. Relating respiratory mucin genes to CF lung disease severity: obtaining more accurate genetic maps and identification of functional variants. *Pediatr Pulmonol.* 46 Suppl 34:270.
94. Kreda SM, Jones L, Moussa, L, Fulcher L, **O'Neal WK**, Lazarowski ER, Boucher RC. 2011. VAMP8 is the v-SNARE that regulates airway mucin secretion. The 25th Annual North American Cystic Fibrosis Conference. *Pediatr Pulmonol.* 46 Suppl 34:246.
95. Kreda SM, **O'Neal WK**, Jones L, Moussa, L, Boucher RC, Lazarowski ER. 2011. Rho signaling and VAMP-8 are important mediators of airway goblet cell mucin secretion. Oral presentation; Gordon Research Conference "Cilia, Mucus & Mucociliary Interactions", Ventura CA, USA. **Carl Storm Award**
96. LaVange L, Greenberg J, Carretta E, Mostafa J, Deshmukh R, **O'Neal WK**, Rennard SI. 2011. The COPD ontology and its role in the Subpopulations and Intermediate Outcome Measures in COPD Study (SPIROMICS). *Am J Respir Crit Care Med.* 183:A6011.
97. Livraghi-Butrico A, Duncan EA, Kelly EJ, Waxer DL, Gilmore RC, Evans CM, Doerschuk CM, Boucher RC, **O'Neal WK**. 2011. Deletion of *Muc5b* in *Scnn1b* transgenic mice reduces airway mucus obstruction but perturbs lung immune homeostasis: a new role for secreted mucins? *Pediatr Pulmonol.* 46 Suppl 34:239.
98. Livraghi-Butrico A, Grubb BR, Kelly EJ, Yang H, Geiser M, Boucher RC, **O'Neal WK**. 2011. Identification of congenic variant of *Scnn1b* transgenic mice reveals contribution of early lesions to the development of obstructive lung disease due to airway surface dehydration. *Pediatr Pulmonol.* 46 Suppl 34:271.
99. Livraghi-Butrico A, Kelly EJ, Klem E, Fulcher NB, Wolfgang MC, Boucher RC, Randell SH, **O'Neal WK**. 2011. Impaired mucus clearance correlates with increased susceptibility to spontaneous bacterial infection/colonization in *Scnn1b*-transgenic mice. American Thoracic Society International Conference, Denver, Colorado. *Am J Respir Crit Care Med.* 183:A1072.
100. Martino M, Brighton B, Dang H, **O'Neal WK**, Ribeiro CP. 2011. IRE1 β regulates transcription of airway mucin genes and genes involved in mucin production and glycosylation. *Pediatr Pulmonol.* 46 Suppl 34:248-9.
101. **O'Neal WK**, Guo X, Dang H, Pace RG, Stonebraker JR, Polineni D, Gallins PJ, Jones LC, Dang AT, Doerschuk CM, Wright FA, Harris A, Knowles MR. 2011. From genes to function: exploring the mechanism of association between CF lung disease severity and genetic variation at Chr11p13. *Pediatr Pulmonol.* 46 Suppl 34:271.
102. Saini Y, Terrel K, Waxer D, Livraghi-Butrico A, **O'Neal WK**, Boucher RC. 2011. The roles of macrophages in determining the initiation, progression and resolution of lung inflammation in a mouse model of chronic bronchitis. *Pediatr. Pulmonol.* 46 Suppl 34:258.
103. Bove PF, Grubb BR, Okada SF, Randell SH, **O'Neal WK**, Boucher RC. 2010. Human alveolar type II cells secrete and absorb liquid in response to local luminal environmental signals. *Pediatr Pulmonol.* 45 Suppl 33:264.
104. Bove PF, Rogers TD, Brighton B, Ribiero CM, Randell SH, **O'Neal WK**, Grubb BR, Boucher RC. 2010. Ion transport and alveolar surface liquid regulation in human alveolar type II cells. *Am J Respir Crit Care Med.* 181:A6460.
105. Gilmore RC, Watson M, Jones L, **O'Neal WK**, Tarran R. 2010. A novel, high-throughput fret assay to monitor NF kappa B activation. *Pediatr Pulmonol.* 45 Suppl 33:295.
106. Kinev A, Clapp P, Tatreau JR, Jones L, Gilmore R, **O'Neal WK**, Randell SH. 2010. Differential involvement of NF-kappa B and C/EBP beta in acute versus chronic responses of human airway epithelium to *P. aeruginosa*. *Pediatr Pulmonol.* 45 Suppl 33:282.
107. Kreda SM, **O'Neal WK**, Jones L, Boucher RC, Lazarowski ER. 2010. VAMP-8 regulates mucin secretion in airway goblet cells. Poster. The 24th Annual North American Cystic Fibrosis Conference, Baltimore MD. *Pediatr Pulmonol.* 45 Suppl 33:248.
108. Kreda SM, Vidal-Seminario L, van Heusden CA, **O'Neal WK**, Jones L, Boucher RC, Lazarowski ER. 2010. Thrombin and neutrophil elastase promote nucleotide release from mucin granules in airway epithelial cells. Poster. The 10th International Symposium on Adenosine and Adenine nucleotides, Tarragona, Spain.
109. Livraghi-Butrico A, Kelly EJ, Evans CE, Boucher RC, **O'Neal WK**. 2010. Deletion of *Muc16*, *Muc5ac*, *Muc5b* in *Scnn1b* transgenic mice: implications for CF lung disease pathogenesis and therapy. *Pediatr Pulmonol.* 45 Suppl 33:258.
110. Livraghi-Butrico A, Kelly EJ, Klem E, Fulcher NB, Wolfgang MC, Boucher RC, Randell SH, **O'Neal WK**. 2010. Increased susceptibility to bacterial infection/colonization in *Scnn1b* transgenic mice. *Pediatr Pulmonol.* 45 Suppl 33:295.

111. Livraghi-Butrico A, Kelly EJ, Boucher RC, **O'Neal WK**, Randell SH. 2010. Non-redundant role of Toll-like receptor 2 and 4 in host defense and development of lung pathology when mucus clearance is impaired. *Pediatr Pulmonol.* 45 Suppl 33:281.
112. Martino MEB, Jones L, Brighton B, **O'Neal WK**, Ribeiro CMP. 2010. The ER stress transducer IRE1 β is a key regulator of airway mucin production. *Pediatr Pulmonol.* 45 Suppl 33:256.
113. Watson M, Gilmore R, **O'Neal WK**, Gentzsch M, Tarran R. 2010. Adenosine 2B receptor signaling with CFTR is regulated by PDZ binding motifs on the C-terminus. *Pediatr Pulmonol.* 45 Suppl 33:226.
114. Cholon DM, **O'Neal WK**, Randell SH, Riordan JR, Gentzsch M. 2009. Apical recycling in primary human airway cells requires N-linked glycans and correct folding of CFTR. *Pediatr Pulmonol.* 44 Suppl 32:219.
115. Kreda SM, Vidal-Seminario L, van Heusden CA, **O'Neal WK**, Jones L, Boucher RC, Lazarowski ER. 2009. Thrombin and neutrophil elastase promote coordinated ATP and mucin release from airway epithelial cells. *Pediatr Pulmonol.* 44 Suppl 32:248-9.
116. Livraghi A, Hudson EJ, Wilkinson KJ, Boucher RC, Doerschuk CM, **O'Neal WK**. 2009. Identification of congenic *Scnn1b*-Tg lines markedly susceptible to pulmonary hemorrhage and right ventricular hypertrophy. *Pediatr Pulmonol.* 44 Suppl 32:279.
117. Livraghi A, Klem ER, Hudson EJ, Wilkinson KJ, Wolfgang MC, **O'Neal WK**, Boucher RC, Randell SH. 2009. Role of MyD88 and environmental bacteria in normal and pathologic airway development. *Am J Respir Crit Care Med.* 179:A2838.
118. Livraghi A, Wilkinson KJ, Hudson EJ, Randell SH, Boucher RC, **O'Neal WK**. 2009. Deletion of membrane-tethered mucins 1 and 4 does not alter the phenotype of *Scnn1b* transgenic mice: implications for airway mucin function. *Pediatr Pulmonol.* 44 Suppl 32:249.
119. Martino MB, Ehre C, **O'Neal WK**, Ribeiro CP. 2009. Role of the ER stress transducer IRE1 beta in airway inflammation-induced mucin production. *Pediatr Pulmonol.* 44 Suppl 32:259.
120. Bove PF, Trout L, Brighton B, **O'Neal WK**, Boucher RC. 2008. Coupling of alveolar to airway surfaces: studies using a transgenic CC10/EGFP secreted protein in alveolar epithelium. *Pediatr Pulmonol.* 43 Suppl 31:230.
121. Braun M, **O'Neal WK**, Xie M, Hurd H, Ribeiro CP. 2008. Selective inhibition of EIF2 alpha dephosphorylation reveals a key role for ER stress in airway inflammation. *Pediatr Pulmonol.* 43 Suppl 31:259.
122. Cholon DM, **O'Neal WK**, Riordan JR, Gentzsch M. 2008. Modulation of apical stability of rescued Δ F508 CFTR in primary human airway epithelial cells. *Pediatr Pulmonol.* 43 Suppl 31:203.
123. Ehre C, **O'Neal WK**, Boucher RC. 2008. A mouse model to study the mechanisms and treatment of mucus adhesion in dehydrated airways. *Pediatr Pulmonol.* 43 Suppl 31:247.
124. Livraghi A, Grubb BR, **O'Neal WK**, Mall M, Boucher RC, Randell SH. 2008. Signaling pathways regulating the airway phenotype of mice exhibiting airway surface dehydration. Experimental Biology Meeting 2008, San Diego, CA. *FASEB J.* 22 Suppl 1:328-7.
125. Livraghi A, Klem ER, Hudson, EJ, Wilkinson KJ, Wolfgang MC, **O'Neal WK**, Boucher RC, Randell SH. 2008. Genetic deletion of MyD88-mediated signaling in β ENaC-overexpressing mice reduces airway neutrophilia but promotes spontaneous, mucus-associated bacterial infection. *Pediatr. Pulmonol.* 43 Suppl 31:262.
126. Livraghi A, Rogers TD, Brighton BW, Hudson EJ, Wilkinson KJ, Boucher RC, **O'Neal WK**, Grubb BR. 2008. Increased airway epithelial necrosis and massive airway neutrophilia in double CF/ β ENaC mice highlights role for CFTR-mediated Cl⁻ secretion in neonatal airways. *Pediatr. Pulmonol.* 43 Suppl 31:247.
127. Murray SA, Livraghi A, Sharp JE, Grubb BR, Hudson EJ, Wilkinson KJ, Jones LC, Boucher RC, **O'Neal WK**. 2008. Analysis of genetic modifiers of phenotypic severity in the β ENaC mouse model of cystic fibrosis. *Pediatr. Pulmonol.* 43 Suppl 31:270.
128. **O'Neal WK**, Hurd H, Wu Y, Braun M, Jones LC, Brighton B, Boucher RC, Ribeiro CM. 2008. Azithromycin alters the gene expression and inflammatory responses of human airway epithelia *Pediatr. Pulmonol.* 43 Suppl 31:243.
129. Sesma JL, Esther CR, Kreda SM, Jones LC, **O'Neal WK**, Nicholas R, Lazarowski E. 2008. Cellular release of UDP-sugars from the secretory pathway. *Pediatr. Pulmonol.* 43 Suppl 31:245.
130. Braun M, **O'Neal WK**, Ribeiro CP. 2007. Expansion of epithelial ER calcium stores during airway infection/inflammation is mediated by Upr-dependent Xbp-1. *Pediatr Pulmonol.* 42 Suppl 30:259-60.
131. Braun M, Tatreau J, Fulcher N, Wolfgang MC, Randell SH, **O'Neal WK**, Ribeiro CP. 2007. Pseudomonas aeruginosa activates the unfolded protein response in airway epithelia: relevance for airway inflammation. *Pediatr Pulmonol.* 42 Suppl 30:260.

132. Gentsch M, **O'Neal WK**, Randell SH, Riordan JR. 2007. Endocytic trafficking of wild-type and Δ F508 CFTR in highly differentiated primary cultures of human airway epithelia. *Pediatr Pulmonol.* 42 Suppl 30:217.
133. Grubb BR, Livraghi A, Rogers TD, Wilkinson KJ, Hudson EJ, Boucher RC, **O'Neal WK**. 2007. Enhancing Na⁺ absorption in the airways of the CF mouse results in increased mortality. *Pediatr Pulmonol.* 42 Suppl 30:280-1.
134. Guo XL, **O'Neal WK**, Yeatts JP, Swallow DM, Pace RG, Stonebraker JR, Zariwala MA, Harris A, Wright FA, Zou F, Perez-Vilar J, Knowles MR. 2007. Assessing mucin gene polymorphisms to determine their role as modifiers of different CF phenotypes. *Pediatr Pulmonol.* 42 Suppl 30:265-6.
135. Jackson PL, Livraghi A, **O'Neal WK**, Noerager BD, Solomon GM, McQuaid DB, Gaggar A, Clancy JP, Sorscher EJ, Liu G, Abraham EA, Blalock JE, Rowe SM. 2007. Novel mediators of persistent neutrophilic inflammation in cystic fibrosis. *Pediatric Pulmonol.* 42 Suppl 30:229-30.
136. Livraghi A, Grubb BR, Hudson EJ, Wilkinson KJ, Mall M, **O'Neal WK**, Boucher RC, Randell SH. 2007. TNF α is not a key mediator of airway inflammation and remodeling in β ENaC-transgenic mice. *Pediatr Pulmonol.* 42 Suppl 30:257-8.
137. Livraghi A, Grubb BR, Threadgill DW, Mall MA, Hudson EJ, Boucher RC, **O'Neal WK**. 2007. Assessment of genetic modifiers for phenotypic severity of *Scnn1b* transgenic mice. *Pediatr Pulmonol.* 42 Suppl 30:266.
138. Livraghi A, Mall M, Rogers TD, Hudson EJ, **O'Neal WK**, Boucher RC, Grubb BR. 2007. Generation and phenotypic characterization of transgenic mice over-expressing multiple subunits of EnaC in the airways. *Pediatr Pulmonol.* 42 Suppl 30:239.
139. Livraghi A, Rogers TD, Wilkinson KJ, Hudson EJ, Boucher RC, **O'Neal WK**. 2007. Enhancing Na⁺ absorption in the airways of the CF mouse results in increased mortality. *Pediatric Pulmonol.* 42 Suppl 30:280-1.
140. Mall M, Harkema JR, Trojanek JB, Treis D, Schubert S, Zhou Z, Tilley SL, Livraghi A, **O'Neal WK**, Boucher RC. 2007. Initial pulmonary lesions and spontaneous course of lung disease caused by airway surface liquid depletion in β ENaC overexpressing mice. *Pediatr Pulmonol.* 42 Suppl 30:279-80.
141. **O'Neal WK**, Winters SL, Stonebraker JR, Godfrey VL, Brighton BW, Jones LC, Hartwell HJ, Gilmore RC, Pickles RJ, Sheehan J, Davis C, Boucher RC. 2007. Deficiency of transmembrane mucin MUC4 reveals a potential role for MUC4 in determination of ciliary beat frequency. *Pediatr Pulmonol.* 42 Suppl 30:240.
142. Rowe SM, Solomon GM, Livraghi A, **O'Neal WK**, Gaggar A, Blalock JE, Clancy JP, Sorscher EJ, Abraham EA, Liu G. 2007. Evidence for pathogenic role of HMGB1 in cystic fibrosis. American Thoracic Society International Conference, San Francisco, CA. *Am J Respir Crit Care Med* 175:A448. 25.
143. Livraghi A, Hudson EJ, DeMaria GC, Sheehan JK, **O'Neal WK**, Boucher RC, Randell SH. 2006. Absence of IL-4R α in *Scnn1b*-transgenic mice enhances neonatal survival but does not mitigate adult lung pathology. *Pediatr Pulmonol.* 41 Suppl 29:246-7.
144. Livraghi A, **O'Neal WK**, Mall M, Boucher RC, Grubb BR. 2006. Enhanced Na⁺ absorption in the double transgenic ENaC mouse *Scnn1b/c* correlates with severity of lung pathology. *Pediatr Pulmonol.* 41 Suppl 29:284.
145. Livraghi A, **O'Neal W**, Mall M, Boucher RC. 2005. Airway inflammation in *Scnn1b* transgenic mice. *Pediatr Pulmonol.* Suppl 28:261.
146. Holda JR, Pickles RJ, Burns KA, Gendler SJ, Boucher RC, **O'Neal WK**. 2001. Muc1 is a significant component of mouse airway luminal glycocalyx. North American Cystic Fibrosis Conference.
147. Mall M, Boucher RC, **O'Neal WK**. 2001. The novel ubiquitin-protein ligase Nedd 4.2 is co-expressed with the amiloride-sensitive Na⁺ channel ENaC in human normal and CF airway epithelial cells. North American Cystic Fibrosis Conference.
148. Randell SH, **O'Neal WK**, Martsen EO, Noone PG, Zhou Z, Plonk MK, Wu Q, Knowles MR, Boucher RC, Gabriel SE. 2001. mRNA expression profiling of human airway epithelium. North American Cystic Fibrosis Conference.
149. Zhang YJ, **O'Neal WK**, Blackburn K, Moyer M, Moseley R, Boucher R, Ostrowski L. 2001. Identification and characterization of an axonemal dynein heavy chain reduced in cilia from a patient with primary ciliary dyskinesia. American Thoracic Society.
150. Lazarowski E, Grubb BR, **O'Neal WK**, Ribeiro CP, Burch N, Boucher RC. 2000. Regulation of gastrointestinal ion secretory responses by uridine nucleotide receptors. North American Cystic Fibrosis Conference.
151. Morse KM, **O'Neal WK**, Patel M, Davis CW, Olsen JC. 2000. An ecdysone-inducible expression system for use with retroviral vectors. *Mol Ther.* 1:S186-7.

152. **O'Neal WK**, Wang Y, Taylor K, Beaudet AL. 1997. Advantages of α -fetoprotein (AFP) as a reporter gene for expression in lung and liver in animals and humans. *Pediatr Pulmonol. Suppl* 14:187.
153. Lee B, Timme TL, Shine HD, **O'Neal WK**, Morral N, Zhou H, et.al. 1996. Toxicological studies of adenovirus mediated gene transfer via a) intracerebral and intraprostatic injection in non-human primates and cotton rats and b) systemic delivery into non-human primates, cotton rats, and mice. National Institute of Health, Gene Therapy Conference, Bethesda, MD, July 11-12.
154. Morral N, **O'Neal WK**, Zhou H, Langston C, Beaudet AL. 1996. Host responses to reporter genes complicate the interpretation of studies using adenoviral vectors. *Pediatr Pulmonol. Suppl* 13:168.
155. **O'Neal WK**, Beaudet AL. 1995. Adenovirus infection in MDCK polarized epithelial cells. Ninth Annual North American Cystic Fibrosis Conference, Dallas, TX, October 12-15.
156. Zhou H, Morral N, **O'Neal WK**, Beaudet AL. 1995. Development of an adenoviral vector with deletions of E1 and E2a genes for CF gene therapy. *Pediatr Pulmonol. Suppl* 12:164.
157. **O'Neal WK**, Ecay T, Katkin J, Dubinsky W. 1994. Adenovirus-mediated expression of CFTR in MDCK cells. *Pediatr. Pulmonol. Suppl* 10:164.
158. **O'Neal WK**, Hasty EP, Liu K-Q, Casey BM, McCray PB Jr., Rivera-Perez J, Welsh MJ, Doyle NA, Doerschuk C, Bradley A, Beaudet AL. 1993. Characterization of mice with mutations in exons 2 and 3 or the cystic fibrosis gene. *Pediatr Pulmonol. Suppl* 9:138.
159. Beaudet AL, Wilson RW, Sligh JE, Ballantyne CM, Lemna (**O'Neal**) **WK**, Patejunas G, Hasty P, Bradley A, O'Brien WE. 1992. Applications of murine gene targeting to human genetics. *J Cell Biochem. Suppl* 16F:6.
160. Lemna (**O'Neal**) **WK**, Hasty EP, Beaudet AL, Bradley A. 1991. Toward a mouse model for cystic fibrosis. *Pediatr Pulmonol. Suppl* 6:50.
161. Beaudet AL, Feldman GL, Kobayashi K, Lemna (**O'Neal**) **WK**, Fernbach SD, O'Brien WE. 1990. Strategies for mutation analysis for cystic fibrosis. Fifth International Congress of Inborn Errors of Metabolism. Asilomar, June 1-5.