3/19/26

1) Personal Information:

Wanda K. O'Neal, Ph.D.
Research Associate Professor

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

2) 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

3) Professional Employment and Employment History:

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	**Master's Degree Student** and **Graduate Teaching Fellow** Advisor: Calvin Messersmith, Ph.D., Department of Crop Science. North Dakota State University, Fargo, North Dakota

4) Honors:

None

5) Bibliography and Products of Scholarship.

Published Refereed Papers/Articles *(from newest to oldest)*

1. **O'Neal WK**, Boucher RC. Molecular Echoes of Mucus Plugging in COPD. Am J Respir Crit Care Med. 2026 Jan 23:aamaf101. doi: 10.1093/ajrccm/aamaf101. Epub ahead of print. PMID: 41738146.
2. Cao S, Shrivastav A, Bleecker ER, Madapoosi S, Erb-Downward J, Zein J, Chen X, Howard TD, Hawkins GA, Barjaktarevic IZ, Bowler R, Barr RG, Comellas A, Cooper CB, Couper D, Han M, Hansel NN, Hastie AT, Kaner R, Kanner RE, Kim V, Martinez FJ, Moore WC, **O'Neal WK**, Paine R, Smith B, Hoffman EA, Meyers DA, Woodruff PG, Christenson S, Curtis JL, Huang YJ, Ortega VE. Epigenomic study of the lower airway reveals COPD-associated methylation patterns and potential microbiota links. BMJ Open Respir Res. 2025 Dec 18;12(1):e003572. doi: 10.1136/bmjresp-2025-003572. PMID: 41419234; PMCID: PMC12716595.
3. Hoesterey DT, Dang H, Markovic D, Buhr RG, Tashkin DP, Barr RG, Belperio JA, Bowler RP, Bleecker ER, Couper DJ, Criner GG, Cooper CB, Doerschuk CM, Dransfield MT, Drummond MB, Fawzy A, Freeman CM, Han MK, Hansel NN, Hastie AT, Hoffman EA, Huang YJ, Kaner RJ, Kanner RE, Kim V, Krishnan JA, Martinez FJ, **O'Neal WK**, Ortega VE, Paine R 3rd, Shrivastav AK, Wells JM, Woodruff PG, Curtis JL, Barjaktarevic I. Neutrophil-to-Lymphocyte Ratio (NLR) as a Biomarker in Clinically Stable Chronic Obstructive Pulmonary Disease: SPIROMICS cohort. Ann Am Thorac Soc. 2025 Dec;22(12):1881-1890. doi: 10.1513/AnnalsATS.202412-1265OC. PMID: 40920896. PMCID: PMC12700239.
4. Sun L, Walls SA, Dang H, Quinney NL, Sears PR, Sadritabrizi T, Hasegawa K, Okuda K, Asakura T, Chang X, Zheng M, Mikami Y, Dizmond FU, Danilova D, Zhou L, Deshmukh A, Cholon DM, Radicioni G, Rogers TD, Kissner WJ, Markovetz MR, Guhr Lee TN, Gutay MI, Esther CR Jr, Chua M, Grubb BR, Ehre C, Kesimer M, Hill DB, Ostrowski LE, Button B, Gentzsch M, Robinson C, Olivier KN, Freeman AF, Randell SH, Vladar E, **O'Neal WK**, Boucher RC Jr, Chen G. STAT3-dependent Regulation of CFTR and Ciliogenesis Is Essential for Mucociliary Clearance and Innate Airway Defense in Hyper-IgE Syndrome. Am J Respir Crit Care Med. 2025 Oct;211(10):1951-1969. doi: 10.1164/rccm.202407-1415OC. PMID: 40315437. PMCID: PMC12555045.
5. Schworer SA, Murano H, Dang H, Markovetz MR, Saito M, Kato T, Asakura T, Chen G, Gilmore RC, Morton LC, van Heusden C, Chua M, Strickler E, Wisniewski ZY, Crisp G, Mitchell E, Doherty KA, Mastan S, Trejo Bittar HE, Cody BA, Trudeau JB, De la Cruz G, Ralph LM, Askin FB, Panettieri RA Jr, Koziol-White CJ, Byrd KM, Livraghi-Butrico A, **O'Neal WK**, Randell SH, Wenzel SE, Okuda K, Boucher RC Jr. Airway Epithelial Heterogeneity and Mucus Plugging in Asthmatic Bronchioles. Am J Respir Crit Care Med. 2025 Sep 23. doi: 10.1164/rccm.202409-1849OC. Epub ahead of print. PMID: 40986379. PMCID: PMC12668797.
6. Lee-Ferris RE, Okuda K, Galiger JR, Schworer SA, Rogers TD, Dang H, Gilmore R, Edwards C, Crisp G, Nakano S, Cawley AM, Pickles RJ, Gallant SC, Crisci E, Rivier L, Hagood JS, **O'Neal WK**, Baric RS, Grubb BR, Boucher RC, Randell SH. Prolonged airway explant culture enables study of health, disease, and viral pathogenesis. Sci Adv. 2025 Apr 25;11(17):eadp0451. doi: 10.1126/sciadv.adp0451. PMID: 40279421; PMCID: PMC12024639.
7. DeMeis JD, Roberts JT, Delcher HA, Godang NL, Coley AB, Brown CL, Shaw MH, Naaz S, Dahal A, Alqudah SY, Nguyen KN, Nguyen AD, Paudel SS, Shell JE, Patil SS, Dang H, **O'Neal WK**, Knowles MR, Houserova D, Gillespie MN, Borchert GM. Long G4-rich enhancers target promoters via a G4 DNA-based mechanism. Nucleic Acids Res. 2025 Jan 11;53(2):gkae1180. doi: 10.1093/nar/gkae1180. PMID: 39658038. PMCID: PMC11754661.
8. Stonebraker JR, Pace RG, Gallins PJ, Dang H, Aksit MA, Faino AV, Gordon WW, MacParland S, Bamshad MJ, Gibson RL, Cutting GR, Durie PR, Wright FA, Zhou YH, Blackman SM, **O'Neal WK**, Ling SC, Knowles MR. Genetic variation in severe cystic fibrosis liver disease is associated with novel mechanisms for disease pathogenesis. Hepatology. 2024 Nov; 80(5):1012-1025. doi: 10.1097/HEP.0000000000000863. PMID: 38536042. PMCID: PMC11427593.
9. Curtis JL, Bateman LA, Murray S, Couper DJ, Labaki WW, Freeman CM, Arnold KB, Christenson SA, Alexis NE, Kesimer M, Boucher RC, Kaner RJ, Barjaktarevic I, Cooper CB, Hoffman EA, Barr RG, Bleecker ER, Bowler RP, Comellas A, Dransfield MT, Freedman MB, Hansel NN, Krishnan JA, Marchetti N, Meyers DA, Ohar J, **O'Neal WK**, Ortega VE, Paine Iii R, Peters SP, Smith BM, Wedzicha JA, Wells JM, Woodruff PG, Han MK, Martinez FJ. Design of the SPIROMICS Study of Early COPD Progression:

SOURCE Study. Chronic Obstr Pulm Dis. 2024 Sep 27;11(5):444-459. doi: 10.15326/jcopdf.2023.0490. PMID: 39159077. PMCID: PMC11548966.

10. Plender EG, Prodanov T, Hsieh P, Nizamis E, Harvey WT, Sulovari A, Munson KM, Kaufman EJ, **O'Neal WK**, Valdmanis PN, Marschall T, Bloom JD, Eichler EE. Structural and genetic diversity in the secreted mucins MUC5AC and MUC5B. Am J Hum Genet. 2024 Aug 8;111(8):1700-1716. doi: 10.1016/j.ajhg.2024.06.007. PMID: 38991590. PMCID: PMC11344006.

11. Sheridan MB, Aksit MA, Pagel K, Hetrick K, Shultz-Lutwyche H, Myers B, Buckingham KJ, Pace RG, Ling H, Pugh E, **O'Neal WK**, Bamshad MJ, Gibson RL, Knowles MR, Blackman SM, Cutting GR, Raraigh KS. The clinical utility of sequencing the entirety of CFTR. J Cyst Fibros. 2024 Jul;23(4):707-715. doi: 10.1016/j.jcf.2024.04.018. PMID: 38734509.

12. Sun Q, Yang Y, Rosen JD, Chen J, Li X, Guan W, Jiang MZ, Wen J, Pace RG, Blackman SM, Bamshad MJ, Gibson RL, Cutting GR, **O'Neal WK**, Knowles MR, Kooperberg C, Reiner AP, Raffield LM, Carson AP, Rich SS, Rotter JI, Loos RJF, Kenny E, Jaeger BC, Min YI, Fuchsberger C, Li Y. MagicalRsq-X: A cross-cohort transferable genotype imputation quality metric. Am J Hum Genet. 2024 May 2;111(5):990-995. doi: 10.1016/j.ajhg.2024.04.001.PMID: 38636510. PMCID: PMC11080605.

13. Asakura T, Okuda K, Chen G, Dang H, Kato T, Mikami Y, Schworer SA, Gilmore RC, Radicioni G, Hawkins P, Barbosa Cardenas SM, Saito M, Cawley AM, De la Cruz G, Chua M, Alexis NE, Masugi Y, Noone PG, Ribeiro CMP, Kesimer M, Olivier KN, Hasegawa N, Randell SH, **O'Neal WK**, Boucher RC. Proximal and Distal Bronchioles Contribute to the Pathogenesis of Non-Cystic Fibrosis Bronchiectasis (NCFB). *Am J Respir Crit Care Med*. 2024 Feb 15;209(4):374-389. doi: 10.1164/rccm.202306-1093OC. PMID: 38016030. PMCID: PMC10878387.

14. Ngo D, Pratte KA, Flexeder C, Petersen H, Dang H, Ma Y, Keyes MJ, Gao Y, Deng S, Peterson BD, Farrell LA, Bhambhani VM, Palacios C, Quadir J, Gillenwater L, Xu H, Emson C, Gieger C, Suhre K, Graumann J, Jain D, Conomos MP, Tracy RP, Guo X, Liu Y, Johnson WC, Cornell E, Durda P, Taylor KD, Papanicolaou GJ, Rich SS, Rotter JI, Rennard SI, Curtis JL, Woodruff PG, Comellas AP, Silverman EK, Crapo JD, Larson MG, Vasan RS, Wang TJ, Correa A, Sims M, Wilson JG, Gerszten RE, O'Connor GT, Barr RG, Couper D, Dupuis J, Manichaikul A, **O'Neal WK**, Tesfaigzi Y, Schulz H, Bowler RP. Systemic Markers of Lung Function and Forced Expiratory Volume in 1 Second Decline across Diverse Cohorts. *Ann Am Thorac Soc*. 2023 Aug;20(8):1124-1135. doi: 10.1513/AnnalsATS.202210-857OC. PMID: 37351609; PMCID: PMC10405603.

15. Ryu MH, Yun JH, Morrow JD, Saferali A, Castaldi P, Chase R, Stav M, Xu Z, Barjaktarevic I, Han M, Labaki W, Huang YJ, Christenson S, **O'Neal W**, Bowler R, Sin DD, Freeman CM, Curtis JL, Hersh CP. Blood Gene Expression and Immune Cell Subtypes Associated with Chronic Obstructive Pulmonary Disease Exacerbations. *Am J Respir Crit Care Med*. 2023 Aug 1;208(3):247-255. doi: 10.1164/rccm.202301-0085OC. PMID: 37286295; PMCID: PMC10395718.

16. Izquierdo M, Marion CR, Genese F, Newell JD, **O'Neal WK**, Li X, Hawkins GA, Barjaktarevic I, Barr RG, Christenson S, Cooper CB, Couper D, Curtis J, Han MK, Hansel NN, Kanner RE, Martinez FJ, Paine R 3rd, Tejwani V, Woodruff PG, Zein JG, Hoffman EA, Peters SP, Meyers DA, Bleecker ER, Ortega VE; National Heart, Lung and Blood Institute's SubPopulations and InteRmediate Outcome Measures In COPD Study (SPIROMICS) investigators. Impact of Bronchiectasis on COPD Severity and Alpha-1 Antitrypsin Deficiency as a Risk Factor in Individuals with a Heavy Smoking History. *Chronic Obstr Pulm Dis*. 2023 Jul 26;10(3):199-210. doi: 10.15326/jcopdf.2022.0388. PMID: 37199731. PMCID: PMC10484491.

17. Van Buren E, Radicioni G, Lester S, **O'Neal WK**, Dang H, Kasela S, Garudadri S, Curtis JL, Han MK, Krishnan JA, Wan ES, Silverman EK, Hastie A, Ortega VE, Lappalainen T, Nawijn MC, Berge MVD, Christenson SA, Li Y, Cho MH, Kesimer M, Kelada SNP. Genetic regulators of sputum mucin concentration and their associations with COPD phenotypes. *PLoS Genet*. 2023 Jun 23;19(6):e1010445. doi: 10.1371/journal.pgen.1010445. PMID: 37352370; PMCID: PMC10325042.

18. Hill AC, Guo C, Litkowski EM, Manichaikul AW, Yu B, Konigsberg IR, Gorbet BA, Lange LA, Pratte KA, Kechris KJ, DeCamp M, Coors M, Ortega VE, Rich SS, Rotter JI, Gerzsten RE, Clish CB, Curtis JL, Hu X, Obeidat ME, Morris M, Loureiro J, Ngo D, **O'Neal WK**, Meyers DA, Bleecker ER, Hobbs BD, Cho MH, Banaei-Kashani F, Bowler RP. Large scale proteomic studies create novel privacy considerations. *Sci*

Rep. 2023 Jun 7;13(1):9254. doi: 10.1038/s41598-023-34866-6. PMID: 37286633; PMCID: PMC10247808.

19. Mikami Y, Grubb BR, Rogers TD, Dang H, Asakura T, Kota P, Gilmore RC, Okuda K, Morton LC, Sun L, Chen G, Wykoff JA, Ehre C, Vilar J, van Heusden C, Livraghi-Butrico A, Gentzsch M, Button B, Stutts MJ, Randell SH, **O'Neal WK**, Boucher RC. Chronic airway epithelial hypoxia exacerbates injury in muco-obstructive lung disease through mucus hyperconcentration. *Sci Transl Med*. 2023 Jun 7;15(699):eabo7728. doi: 10.1126/scitranslmed.abo7728. PMID: 37285404. PMCID: PMC10664029.

20. DiLillo KM, Norman KC, Freeman CM, Christenson SA, Alexis NE, Anderson WH, Barjaktarevic IZ, Barr RG, Comellas AP, Bleecker ER, Boucher RC, Couper DJ, Criner GJ, Doerschuk CM, Wells JM, Han MK, Hoffman EA, Hansel NN, Hastie AT, Kaner RJ, Krishnan JA, Labaki WW, Martinez FJ, Meyers DA, **O'Neal WK**, Ortega VE, Paine R 3rd, Peters SP, Woodruff PG, Cooper CB, Bowler RP, Curtis JL, Arnold KB; SPIROMICS investigators. A blood and bronchoalveolar lavage protein signature of rapid FEV1 decline in smoking-associated COPD. *Sci Rep*. 2023 May 22;13(1):8228. doi: 10.1038/s41598-023-32216-0. PMID: 37217548; PMCID: PMC10203309.

21. Zhou YH, Gallins PJ, Pace RG, Dang H, Aksit MA, Blue EE, Buckingham KJ, Collaco JM, Faino AV, Gordon WW, Hetrick KN, Ling H, Liu W, Onchiri FM, Pagel K, Pugh EW, Raraigh KS, Rosenfeld M, Sun Q, Wen J, Li Y, Corvol H, Strug LJ, Bamshad MJ, Blackman SM, Cutting GR, Gibson RL, **O'Neal WK**, Wright FA, Knowles MR. Genetic Modifiers of Cystic Fibrosis Lung Disease Severity: Whole Genome Analysis of 7,840 Patients. *Am J Respir Crit Care Med*. 2023 May 15;207(10):1324-1333. doi: 10.1164/rccm.202209-1653OC. PMID: 36921087. PMCID: PMC10595435.

22. LaFon DC, Woo H, Fedarko N, Azar A, Hill H, Tebo AE, Martins TB, Han MK, Krishnan JA, Ortega VE, Barjaktarevic I, Kaner RJ, Hastie A, **O'Neal WK**, Couper D, Woodruff PG, Curtis JL, Hansel NN, Nahm MH, Dransfield MT, Putcha N; SPIROMICS investigators. Reduced quantity and function of pneumococcal antibodies are associated with exacerbations of COPD in SPIROMICS. *Clin Immunol*. 2023 May;250:109324. doi: 10.1016/j.clim.2023.109324. PMID: 37030524. PMCID: PMC10171244.

23. LeMaster WB, Quibrera PM, Couper D, Tashkin DP, Bleecker ER, Doerschuk CM, Ortega VE, Cooper C, Han MK, Woodruff PG, **O'Neal WK**, Anderson WH, Alexis NE, Bowler RP, Barr RG, Kaner RJ, Dransfield MT, Paine R 3rd, Kim V, Curtis JL, Martinez FJ, Hastie AT, Barjaktarevic I. Clinical Implications of Low Absolute Blood Eosinophil Count in the SPIROMICS COPD Cohort. *Chest*. 2023 Mar;163(3):515-528. doi: 10.1016/j.chest.2022.10.029. PMID: 36343688; PMCID: PMC10083128.

24. Esther CR Jr, **O'Neal WK**, Alexis NE, Koch AL, Cooper CB, Barjaktarevic I, Raffield LM, Bowler RP, Comellas AP, Peters SP, Hastie AT, Curtis JL, Ronish B, Ortega VE, Wells JM, Halper-Stromberg E, Rennard SI, Boucher RC. Prolonged, Physiologically Relevant Nicotine Concentrations in the Airways of Smokers. *Am J Physiol Lung Cell Mol Physiol*. 2023 Jan 1;324(1):L32-L37. doi: 10.1152/ajplung.00038.2022. PMID: 36342131. PMCID: PMC9829458.

25. Kato T, Asakura T, Edwards CE, Dang H, Mikami Y, Okuda K, Chen G, Sun L, Gilmore RC, Hawkins P, De la Cruz G, Cooley MR, Bailey AB, Hewitt SM, Chertow DS, Borczuk AC, Salvatore S, Martinez FJ, Thorne LB, Askin FB, Ehre C, Randell SH, **O'Neal WK**, Baric RS, Boucher RC; NIH COVID-19 Autopsy Consortium. Prevalence and mechanisms of mucus accumulation in COVID-19 lung disease. *Am J Respir Crit Care Med*. 2022 Dec 1;206(11):1336-1352. doi: 10.1164/rccm.202111-2606OC. PMID: 35816430. PMCID: PMC9746856.

26. Dinnon KH§, Leist SR§, Okuda K§, Dang H§, Fritch EJ§, Gully KL, De la Cruz G, Evangelista MD, Asakura T, Gilmore RC, Hawkins P, Nakano S, West A, Schäfer A, Gralinski LE, Everman JL, Sajuthi SP, Zweigart MR, Dong S, McBride J, Cooley MR, Hines JB, Love MK, Groshong SD, VanSchoiack A, Phelan SJ, Liang Y, Hether T, Leon M, Zumwalt RE, Barton LM, Duval EJ, Mukhopadhyay S, Stroberg E, Borczuk A, Thorne LB, Sakthivel MK, Lee YZ, Hagood JS, Mock JR, Seibold MA, **O'Neal WK**, Montgomery SA, Boucher RC*, Baric RS*. SARS-CoV-2 infection produces chronic pulmonary epithelial and immune cell dysfunction with fibrosis in mice. *Sci Transl Med*. 2022 Sep 28;14(664):eabo5070. doi: 10.1126/scitranslmed.abo5070. PMID: 35857635. PMCID: PMC9273046. §These authors contributed equally. *Joint senior authorship.

27. Biering SB, Sarnik SA, Wang E, Zengel JR, Leist SR, Schäfer A, Sathyan V, Hawkins P, Okuda K, Tau C, Jangid AR, Duffy CV, Wei J, Gilmore RC, Alfajaro MM, Strine MS, Nguyenla X, Van Dis E, Catamura C, Yamashiro LH, Belk JA, Begeman A, Stark JC, Shon DJ, Fox DM, Ezzatpour S, Huang E, Olegario N,

Rustagi A, Volmer AS, Livraghi-Butrico A, Wehri E, Behringer RR, Cheon DJ, Schaletzky J, Aguilar HC, Puschnik AS, Button B, Pinsky BA, Blish CA, Baric RS, **O'Neal WK**, Bertozzi CR, Wilen CB, Boucher RC, Carette JE, Stanley SA, Harris E, Konermann S, Hsu PD. Genome-wide bidirectional CRISPR screens identify mucins as host factors modulating SARS-CoV-2 infection. *Nat Genet*. 2022 Aug;54(8):1078-1089. doi: 10.1038/s41588-022-01131-x. PMID: 35879412. PMCID: PMC9355872.

28. Esther CR Jr, **O'Neal WK**, Anderson WH, Kesimer M, Ceppe A, Doerschuk CM, Alexis NE, Hastie AT, Barr RG, Bowler RP, Wells JM, Oelsner EC, Comellas AP, Tesfaigzi Y, Kim V, Paulin LM, Cooper CB, Han MK, Huang YJ, Labaki WW, Curtis JL, Boucher RC; SPIROMICS. Identification of Sputum Biomarkers Predictive of Pulmonary Exacerbations in Chronic Obstructive Pulmonary Disease. *Chest*. 2022 May;161(5):1239-1249. doi: 10.1016/j.chest.2021.10.049. PMID: 34801592. PMCID: PMC9131049.

29. Raraigh KS, Aksit MA, Hetrick K, Pace RG, Ling H, **O'Neal W**, Blue E, Zhou YH, Bamshad MJ, Blackman SM, Gibson RL, Knowles MR, Cutting GR. Complete CFTR gene sequencing in 5,058 individuals with cystic fibrosis informs variant-specific treatment. *J Cyst Fibros*. 2022 May;21(3):463-470. doi: 10.1016/j.jcf.2021.10.011. Epub 2021 Nov 12. PMID: 34782259.

30. Morrison CB, Edwards CE, Shaffer KM, Araba KC, Wykoff JA, Williams DR, Asakura T, Dang H, Morton LC, Gilmore RC, **O'Neal WK**, Boucher RC, Baric RS, Ehre C. SARS-CoV-2 infection of airway cells causes intense viral and cell shedding, two spreading mechanisms affected by IL-13.*Proc Natl Acad Sci USA.* 2022 Apr 19;119(16):e2119680119. doi: 10.1073/pnas.2119680119. Epub 2022 Mar 30. PMID: 35353667. PMCID: PMC9169748.

31. Kato T, Radicioni G, Papanikolas MJ, Stoychev GV, Markovetz MR, Aoki K, Porterfield M, Okuda K, Barbosa Cardenas SM, Gilmore RC, Morrison CB, Ehre C, Burns KA, White KK, Brennan TA, Goodell HP, Thacker H, Loznev HT, Forsberg LJ, Nagase T, Rubinstein M, Randell SH, Tiemeyer M, Hill DB, Kesimer M, **O'Neal WK**, Ballard ST, Freeman R, Button B, Boucher RC. Mucus concentration-dependent biophysical abnormalities unify submucosal gland and superficial airway dysfunction in cystic fibrosis. *Sci Adv.* 2022 Apr;8(13):eabm9718. doi: 10.1126/sciadv.abm9718. Epub 2022 Apr 1. PMID: 35363522.

32. Morrison CB, Shaffer KM, Araba KC, Markovetz MR, Wykoff JA, Quinney NL, Hao S, Delion MF, Flen AL, Morton LC, Liao J, Hill DB, Drumm ML, **O'Neal WK**, Kesimer M, Gentzsch M, Ehre C. Treatment of cystic fibrosis airway cells with CFTR modulators reverses aberrant mucus properties via hydration. *Eur Respir J*. 2022 Feb 3;59(2):2100185. doi: 10.1183/13993003.00185-2021. PMID: 34172469; PMCID: PMC8859811.

33. Georas SN, Wright RJ, Ivanova A, Israel E, LaVange LM, Akuthota P, Carr TF, Denlinger LC, Fajt ML, Kumar R, **O'Neal WK**, Phipatanakul W, Szefler SJ, Aronica MA, Bacharier LB, Burbank AJ, Castro M, Crotty Alexander L, Bamdad J, Cardet JC, Comhair SAA, Covar RA, DiMango EA, Erwin K, Erzurum SC, Fahy JV, Gaffin JM, Gaston B, Gerald LB, Hoffman EA, Holguin F, Jackson DJ, James J, Jarjour NN, Kenyon NJ, Khatri S, Kirwan JP, Kraft M, Krishnan JA, Liu AH, Liu MC, Marquis MA, Martinez F, Mey J, Moore WC, Moy JN, Ortega VE, Peden DB, Pennington E, Peters MC, Ross K, Sanchez M, Smith LJ, Sorkness RL, Wechsler ME, Wenzel SE, White SR, Zein J, Zeki AA, Noel P; PrecISE Study Team. The Precision Interventions for Severe and/or Exacerbation-Prone (PrecISE) Asthma Network: An overview of Network organization, procedures, and interventions. *J Allergy Clin Immunol*. 2022 Feb;149(2):488-516.e9. doi: 10.1016/j.jaci.2021.10.035. Epub 2021 Nov 29. PMID: 34848210; PMCID: PMC8821377.

34. Sun Q, Liu W, Rosen JD, Huang L, Pace RG, Dang H, Gallins PJ, Blue EE, Ling H, Corvol H, Strug LJ, Bamshad MJ, Gibson RL, Pugh EW, Blackman SM, Cutting GR, **O'Neal WK**, Zhou YH, Wright FA, Knowles MR, Wen J, Li Y; Cystic Fibrosis Genome Project. Leveraging TOPMed imputation server and constructing a cohort-specific imputation reference panel to enhance genotype imputation among cystic fibrosis patients. *HGG Adv.* 2022 Jan 11;3(2):100090. doi: 10.1016/j.xhgg.2022.100090. eCollection 2022 Apr 14. PMID: 35128485; PMCID: PMC8804187.

35. Radicioni G, Ceppe A, Ford AA, Alexis NE, Barr RG, Bleecker ER, Christenson SA, Cooper CB, Han MK, Hansel NN, Hastie AT, Hoffman EA, Kanner RE, Martinez FJ, Ozkan E, Paine R 3rd, Woodruff PG, **O'Neal WK**, Boucher RC, Kesimer M. Airway mucin MUC5AC and MUC5B concentrations and the initiation and progression of chronic obstructive pulmonary disease: an analysis of the SPIROMICS cohort. *Lancet Respir Med*. 2021 Nov;9(11):1241-1254. doi: 10.1016/S2213-2600(21)00079-5. Epub 2021 May 28. PMID: 34058148; PMCID: PMC8570975.

36. Carpenter J, Wang Y, Gupta R, Li Y, Haridass P, Subramani DB, Reidel B, Morton L, Ridley C, **O'Neal WK**, Buisine MP, Ehre C, Thornton DJ, Kesimer M. Assembly and organization of the N-terminal region of mucin MUC5AC: Indications for structural and functional distinction from MUC5B. *Proc Natl Acad Sci U S A.* 2021 Sep 28;118(39): e2104490118. doi: 10.1073/pnas.2104490118. PMID: 34548396; PMCID: PMC8488587.

37. Eastman AC, Pace RG, Dang H, Aksit MA, Vecchio-Pagán B, Lam AN, **O'Neal WK**, Blackman SM, Knowles MR, Cutting GR. SLC26A9 SNP rs7512462 is not associated with lung disease severity or lung function response to ivacaftor in cystic fibrosis patients with G551D-CFTR. *J Cyst Fibros*. 2021 Sep;20(5):851-856. doi: 10.1016/j.jcf.2021.02.007. Epub 2021 Mar 2. PMID: 33674211; PMCID: PMC8410892.

38. Dunican EM, Elicker BM, Henry T, Gierada DS, Schiebler ML, Anderson W, Barjaktarevic I, Barr RG, Bleecker ER, Boucher RC, Bowler RP, Christenson SA, Comellas A, Cooper CB, Couper D, Criner GJ, Dransfield M, Doerschuk CM, Drummond MB, Hansel NN, Han MK, Hastie AT, Hoffman EA, Krishnan JA, Lazarus SC, Martinez FJ, McCulloch CE, **O'Neal WK**, Ortega VE, Paine R 3rd, Peters S, Schroeder JD, Woodruff PG, Fahy JV. *Am J Respir Crit Care Med*. 2021 Apr 15;203(8):957-968. doi: 10.1164/rccm.202006-2248OC. PMID: 33180550; PMCID: PMC8048745.

39. Fortis S, Comellas A, Bhatt SP, Hoffman EA, Han MK, Bhakta NR, Paine R 3rd, Ronish B, Kanner RE, Dransfield M, Hoesterey D, Buhr RG, Barr RG, Dolezal B, Ortega VE, Drummond MB, Arjomandi M, Kaner RJ, Kim V, Curtis JL, Bowler RP, Martinez F, Labaki WW, Cooper CB, **O'Neal WK**, Criner G, Hansel NN, Krishnan JA, Woodruff P, Couper D, Tashkin D, Barjaktarevic I. Ratio of Forced Expiratory Volume in 1 second /Slow Vital Capacity (FEV1/SVC)<0.7 is associated with clinical, functional, and radiologic features of obstructive lung disease in smokers with preserved lung function. *Chest*. 2021 Jul;160(1):94-103. doi: 10.1016/j.chest.2021.01.067. Epub 2021 Feb 1. PMID: 33539837; PMCID: PMC8295909.

40. Zou C, Li F, Choi J, Haghighi B, Choi S, Rajaraman PK, Comellas AP, Newell JD, Lee CH, Barr RG, Bleecker E, Cooper CB, Couper D, Han M, Hansel NN, Kanner RE, Kazerooni EA, Kleerup EC, Martinez FJ, **O'Neal W**, Paine R 3rd, Rennard SI, Smith BM, Woodruff PG, Hoffman EA, Lin CL. Longitudinal Imaging-Based Clusters in Former Smokers of the COPD Cohort Associate with Clinical Characteristics: The SubPopulations and Intermediate Outcome Measures in COPD Study (SPIROMICS). Int J Chron Obstruct Pulmon Dis. 2021 May 31;16:1477-1496. doi: 10.2147/COPD.S301466. eCollection 2021. PMID: 34103907. PMCID: PMC8178702.

41. Okuda K, Dang H, Kobayashi Y, Carraro G, Nakano S, Chen G, Kato T, Asakura T, Gilmore RC, Morton LC, Lee RE, Mascenik T, Yin WN, Barbosa Cardenas SM, O'Neal YK, Minnick CE, Chua M, Quinney NL, Gentzsch M, Anderson CW, Ghio A, Matsui H, Nagase T, Ostrowski LE, Grubb BR, Olsen JC, Randell SH, Stripp BR, Tata PR, **O'Neal WK**, Boucher RC. Secretory Cells Dominate Airway CFTR Expression and Function in Human Airway Superficial Epithelia. *Am J Respir Crit Care Med*. 2021 May 15;203(10):1275-1289. doi: 10.1164/rccm.202008-3198OC. PMID: 33321047; PMCID: PMC8456462.

42. Israel E, Denlinger LC, Bacharier LB, LaVange LM, Moore WC, Peters MC, Georas SN, Wright RJ, Mauger DT, Noel P, Akuthota P, Bach J, Bleecker ER, Cardet JC, Carr TF, Castro M, Cinelli A, Comhair SAA, Covar RA, Alexander LC, DiMango EA, Erzurum SC, Fahy JV, Fajt ML, Gaston BM, Hoffman EA, Holguin F, Jackson DJ, Jain S, Jarjour NN, Ji Y, Kenyon NJ, Kosorok MR, Kraft M, Krishnan JA, Kumar R, Liu AH, Liu MC, Ly NP, Marquis MA, Martinez FD, Moy JN, **O'Neal WK**, Ortega VE, Peden DB, Phipatanakul W, Ross K, Smith LJ, Szefler SJ, Teague WG, Tulchinsky AF, Vijayanand P, Wechsler ME, Wenzel SE, White SR, Zeki AA, Ivanova A. PrecISE: Precision Medicine in Severe Asthma: An adaptive platform trial with biomarker ascertainment. *J Allergy Clin Immunol*. 2021 May;147(5):1594-1601. doi: 10.1016/j.jaci.2021.01.037. Epub 2021 Mar 2. PMID: 33667479; PMCID: PMC8113113.

43. Pratte KA, Curtis JL, Kechris K, Couper D, Cho MH, Silverman EK, DeMeo DL, Sciurba FC, Zhang Y, Ortega VE, **O'Neal WK**, Gillenwater LA, Lynch DA, Hoffman EA, Newell JD Jr, Comellas AP, Castaldi PJ, Miller BE, Pouwels SD, Hacken NHTT, Bischoff R, Klont F, Woodruff PG, Paine R, Barr RG, Hoidal J, Doerschuk CM, Charbonnier JP, Sung R, Locantore N, Yonchuk JG, Jacobson S, Tal-Singer R, Merrill D, Bowler RP. Soluble receptor for advanced glycation end products (sRAGE) as a biomarker of COPD. *Respir Res*. 2021 Apr 27;22(1):127. doi: 10.1186/s12931-021-01686-z. PMID: 33906653; PMCID: PMC8076883.

44. Opron K, Begley LA, Erb-Downward JR, Freeman C, Madapoosi S, Alexis NE, Barjaktarevic I, Graham Barr R, Bleecker ER, Bowler RP, Christenson SA, Comellas AP, Cooper CB, Couper DJ, Doerschuk CM, Dransfield MT, Han MK, Hansel NN, Hastie AT, Hoffman EA, Kaner RJ, Krishnan J, **O'Neal WK**, Ortega VE, Paine R 3rd, Peters SP, Michael Wells J, Woodruff PG, Martinez FJ, Curtis JL, Huffnagle GB, Huang YJ. Lung microbiota associations with clinical features of COPD in the SPIROMICS cohort. *NPJ Biofilms Microbiomes*. 2021 Feb 5;7(1):14. doi: 10.1038/s41522-021-00185-9. PMID: 33547327; PMCID: PMC7865064.

45. Xu J, Livraghi-Butrico A, Hou X, Rajagopalan C, Zhang J, Song J, Jiang H, Wei HG, Wang H, Bouhamdan M, Ruan J, Yang D, Qiu Y, Xie Y, Barrett R, McClellan S, Mou H, Wu Q, Chen X, Rogers TD, Wilkinson KJ, Gilmore RC, Esther CR Jr, Zaman K, Liang X, Sobolic M, Hazlett L, Zhang K, Frizzell RA, Gentzsch M, **O'Neal WK**, Grubb BR, Chen YE, Boucher RC, Sun F. Phenotypes of CF rabbits generated by CRISPR/Cas9-mediated disruption of the CFTR gene. *JCI Insight*. 2021 Jan 11;6(1):e139813. doi: 10.1172/jci.insight.139813. PMID: 33232302; PMCID: PMC7821608.

46. Gillenwater LA, Pratte KA, Hobbs BD, Cho MH, Zhuang Y, Halper-Stromberg E, Cruickshank-Quinn C, Reisdorph N, Petrache I, Labaki WW, **O'Neal WK**, Ortega VE, Jones DP, Uppal K, Jacobson S, Michelotti G, Wendt CH, Kechris KJ, Bowler RP. Plasma Metabolomic Signatures of Chronic Obstructive Pulmonary Disease and the Impact of Genetic Variants on Phenotype-Driven Modules. *Netw Syst Med*. 2020 Dec 1;3(1):159-181. doi: 10.1089/nsm.2020.0009. Epub 2020 Dec 31. PMID: 33987620; PMCID: PMC8109053.

47. Putcha N, Fawzy A, Matsui EC, Liu MC, Bowler RP, Woodruff PG, **O'Neal WK**, Comellas AP, Han MK, Dransfield MT, Wells JM, Lugogo N, Gao L, Talbot CC Jr, Hoffman EA, Cooper CB, Paulin LM, Kanner RE, Criner G, Ortega VE, Barr RG, Krishnan JA, Martinez FJ, Drummond MB, Wise RA, Diette GB, Hersh CP, Hansel NN. Clinical Phenotypes of Atopy and Asthma in COPD: A Meta-analysis of SPIROMICS and COPDGene. *Chest*. 2020 Dec;158(6):2333-2345. doi: 10.1016/j.chest.2020.04.069. Epub 2020 May 23. PMID: 32450244; PMCID: PMC7768932.

48. Dang H, Polineni D, Pace RG, Stonebraker JR, Corvol H, Cutting GR, Drumm ML, Strug LJ, **O'Neal WK**, Knowles MR. Mining GWAS and eQTL data for CF lung disease modifiers by gene expression imputation. *PLoS One*. 2020 Nov 30;15(11):e0239189. doi: 10.1371/journal.pone.0239189. PMID: 33253230; PMCID: PMC7703903.

49. Kim V, Jeong S, Zhao H, Kesimer M, Boucher RC, Wells JM, Christenson SA, Han MK, Dransfield M, Paine R 3rd, Cooper CB, Barjaktarevic I, Bowler R, Curtis JL, Kaner RJ, O'Beirne SL, **O'Neal WK**, Rennard SI, Martinez FJ, Woodruff PG. Current smoking with or without chronic bronchitis is independently associated with goblet cell hyperplasia in healthy smokers and COPD subjects. *Sci Rep*. 2020 Nov 18;10(1):20133. doi: 10.1038/s41598-020-77229-1. PMID: 33208859; PMCID: PMC7674445.

50. Mock JR, Dial CF, Tune MK, Gilmore RC, **O'Neal WK**, Dang H, Doerschuk CM. Impact of Regulatory T Cells on Type 2 Alveolar Epithelial Cell Transcriptomes during Resolution of Acute Lung Injury and Contributions of IFN-γ. *Am J Respir Cell Mol Biol*. 2020 Oct;63(4):464-477. doi: 10.1165/rcmb.2019-0399OC. PMID: 32543909; PMCID: PMC7528916.

51. Kato T, Hiraishi Y, Kaseda K, Suzuki S, Tanaka G, Yamauchi Y, Yasui M, Shinozaki-Ushiku A, Ushiku T, Yamaji O, **O'Neal WK**, Nagase T. A 79-Year-Old Man With Progressive Dyspnea and Multiple Pulmonary Nodules. *Chest*. 2020 Aug;158(2):e79-e84. doi: 10.1016/j.chest.2020.03.067. PMID: 32768080.

52. Hou YJ, Okuda K, Edwards CE, Martinez DR, Asakura T, Dinnon KH 3rd, Kato T, Lee RE, Yount BL, Mascenik TM, Chen G, Olivier KN, Ghio A, Tse LV, Leist SR, Gralinski LE, Schäfer A, Dang H, Gilmore R, Nakano S, Sun L, Fulcher ML, Livraghi-Butrico A, Nicely NI, Cameron M, Cameron C, Kelvin DJ, de Silva A, Margolis DM, Markmann A, Bartelt L, Zumwalt R, Martinez FJ, Salvatore SP, Borczuk A, Tata PR, Sontake V, Kimple A, Jaspers I, **O'Neal WK**, Randell SH, Boucher RC, Baric RS. SARS-CoV-2 Reverse Genetics Reveals a Variable Infection Gradient in the Respiratory Tract. *Cell*. 2020 Jul 23;182(2):429-446.e14. doi: 10.1016/j.cell.2020.05.042. Epub 2020 May 27. PMID: 32526206; PMCID: PMC7250779.

53. Zhang WZ, Oromendia C, Kikkers SA, Butler JJ, O'Beirne S, Kim K, **O'Neal WK**, Freeman CM, Christenson SA, Peters SP, Wells JM, Doerschuk C, Putcha N, Barjaktarevic I, Woodruff PG, Cooper CB, Bowler RP, Comellas AP, Criner GJ, Paine R 3rd, Hansel NN, Han MK, Crystal RG, Kaner RJ, Ballman KV, Curtis JL, Martinez FJ, Cloonan SM. Increased airway iron parameters and risk for exacerbation in

COPD: an analysis from SPIROMICS. *Sci Rep*. 2020 Jun 29;10(1):10562. doi: 10.1038/s41598-020-67047-w. PMID: 32601308; PMCID: PMC7324559.

54. Raffield LM, Dang H, Pratte KA, Jacobson S, Gillenwater LA, Ampleford E, Barjaktarevic I, Basta P, Clish CB, Comellas AP, Cornell E, Curtis JL, Doerschuk C, Durda P, Emson C, Freeman CM, Guo X, Hastie AT, Hawkins GA, Herrera J, Johnson WC, Labaki WW, Liu Y, Masters B, Miller M, Ortega VE, Papanicolaou G, Peters S, Taylor KD, Rich SS, Rotter JI, Auer P, Reiner AP, Tracy RP, Ngo D, Gerszten RE, **O'Neal WK**, Bowler RP; NHLBI Trans-Omics for Precision Medicine (TOPMed) Consortium. Comparison of Proteomic Assessment Methods in Multiple Cohort Studies. *Proteomics*. 2020 Jun;20(12):e1900278. doi: 10.1002/pmic.201900278. PMID: 32386347; PMCID: PMC7425176.

55. Rao W, Wang S, Duleba M, Niroula S, Goller K, Xie J, Mahalingam R, Neupane R, Liew AA, Vincent M, Okuda K, **O'Neal WK**, Boucher RC, Dickey BF, Wechsler ME, Ibrahim O, Engelhardt JF, Mertens TCJ, Wang W, Jyothula SSK, Crum CP, Karmouty-Quintana H, Parekh KR, Metersky ML, McKeon FD, Xian W. Regenerative Metaplastic Clones in COPD Lung Drive Inflammation and Fibrosis. *Cell*. 2020 May 14;181(4):848-864.e18. doi: 10.1016/j.cell.2020.03.047. Epub 2020 Apr 15. PMID: 32298651; PMCID: PMC7294989.

56. McCarron A, Cmielewski P, Reyne N, McIntyre C, Finnie J, Craig F, Rout-Pitt N, Delhove J, Schjenken JE, Chan HY, Boog B, Knight E, Gilmore RC, **O'Neal WK**, Boucher RC, Parsons D, Donnelley M. Phenotypic Characterization and Comparison of Cystic Fibrosis Rat Models Generated Using CRISPR/Cas9 Gene Editing. *Am J Pathol*. 2020 May;190(5):977-993. doi: 10.1016/j.ajpath.2020.01.009. Epub 2020 Feb 18. PMID: 32084371.

57. **O'Neal WK**, Ribeiro CMP. "Shocking" the System to Achieve Efficient Gene Targeting in Primary Human Airway Epithelia. *Am J Respir Cell Mol Biol*. 2020 Mar;62(3):279-280. doi: 10.1165/rcmb.2019-0360ED. PMID: 31633992; PMCID: PMC7055690.

58. Ortega VE, Li X, **O'Neal WK**, Lackey L, Ampleford E, Hawkins GA, Grayeski PJ, Laederach A, Barjaktarevic I, Barr RG, Cooper C, Couper D, Han MK, Kanner RE, Kleerup EC, Martinez FJ, Paine R 3rd, Peters SP, Pirozzi C, Rennard SI, Woodruff PG, Hoffman EA, Meyers DA, Bleecker ER; NHLBI Subpopulations and Intermediate Outcomes Measures in COPD Study (SPIROMICS). The Effects of Rare SERPINA1 Variants on Lung Function and Emphysema in SPIROMICS. *Am J Respir Crit Care Med*. 2020 Mar 1;201(5):540-554. doi: 10.1164/rccm.201904-0769OC. PMID: 31661293; PMCID: PMC7047460.

59. van Heusden C, Button B, Anderson WH, Ceppe A, Morton LC, **O'Neal WK**, Dang H, Alexis NE, Donaldson S, Stephan H, Boucher RC, Lazarowski ER. Inhibition of ATP hydrolysis restores airway surface liquid production in cystic fibrosis airway epithelia. *Am J Physiol Lung Cell Mol Physiol*. 2020 Feb 1;318(2):L356-L365. doi: 10.1152/ajplung.00449.2019. Epub 2019 Dec 4. PMID: 31800264; PMCID: PMC7052677.

60. Barjaktarevic IZ, Buhr RG, Wang X, Hu S, Couper D, Anderson W, Kanner RE, Paine Iii R, Bhatt SP, Bhakta NR, Arjomandi M, Kaner RJ, Pirozzi CS, Curtis JL, **O'Neal WK**, Woodruff PG, Han MK, Martinez FJ, Hansel N, Wells JM, Ortega VE, Hoffman EA, Doerschuk CM, Kim V, Dransfield MT, Drummond MB, Bowler R, Criner G, Christenson SA, Ronish B, Peters SP, Krishnan JA, Tashkin DP, Cooper CB; NHLBI SubPopulations and InteRmediate Outcome Measures In COPD Study (SPIROMICS). Clinical Significance of Bronchodilator Responsiveness Evaluated by Forced Vital Capacity in COPD: SPIROMICS Cohort Analysis. *Int J Chron Obstruct Pulmon Dis*. 2019 Dec 20;14:2927-2938. doi: 10.2147/COPD.S220164. Erratum in: Int J Chron Obstruct Pulmon Dis. 2020 Apr 23;15:901. PMID: 31908441; PMCID: PMC6930016.

61. Hobbs BD, Putman RK, Araki T, Nishino M, Gudmundsson G, Gudnason V, Eiriksdottir G, Zilhao Nogueira NR, Dupuis J, Xu H, O'Connor GT, Manichaikul A, Nguyen J, Podolanczuk AJ, Madahar P, Rotter JI, Lederer DJ, Barr RG, Rich SS, Ampleford EJ, Ortega VE, Peters SP, **O'Neal WK**, Newell JD Jr, Bleecker ER, Meyers DA, Allen RJ, Oldham JM, Ma SF, Noth I, Jenkins RG, Maher TM, Hubbard RB, Wain LV, Fingerlin TE, Schwartz DA, Washko GR, Rosas IO, Silverman EK, Hatabu H, Cho MH, Hunninghake GM. Overlap of Genetic Risk between Interstitial Lung Abnormalities and Idiopathic Pulmonary Fibrosis. *Am J Respir Crit Care Med*. 2019 Dec 1;200(11):1402-1413. doi: 10.1164/rccm.201903-0511OC. PMID: 31339356; PMCID: PMC6884045.

62. Chen G, Sun L, Kato T, Okuda K, Martino MB, Abzhanova A, Lin JM, Gilmore RC, Batson BD, O'Neal YK, Volmer AS, Dang H, Deng Y, Randell SH, Button B, Livraghi-Butrico A, Kesimer M, Ribeiro CM, **O'Neal WK**, Boucher RC. IL-1β dominates the promucin secretory cytokine profile in cystic fibrosis. *J Clin Invest*. 2019 Oct 1;129(10):4433-4450. doi: 10.1172/JCI125669. PMID: 31524632; PMCID: PMC6763234.

63. Labaki WW, Gu T, Murray S, Curtis JL, Yeomans L, Bowler RP, Barr RG, Comellas AP, Hansel NN, Cooper CB, Barjaktarevic I, Kanner RE, Paine R 3rd, McDonald MN, Krishnan JA, Peters SP, Woodruff PG, **O'Neal WK**, Diao W, He B, Martinez FJ, Standiford TJ, Stringer KA, Han MK. Serum amino acid concentrations and clinical outcomes in smokers: SPIROMICS metabolomics study. Sci Rep. 2019 Aug 6;9(1):11367. doi: 10.1038/s41598-019-47761-w. PMID: 31388056; PMCID: PMC6684630.

64. Halper-Stromberg E, Gillenwater L, Cruickshank-Quinn C, **O'Neal WK**, Reisdorph N, Petrache I, Zhuang Y, Labaki WW, Curtis JL, Wells J, Rennard S, Pratte KA, Woodruff P, Stringer KA, Kechris K, Bowler RP. Bronchoalveolar Lavage Fluid from COPD Patients Reveals More Compounds Associated with Disease than Matched Plasma. *Metabolites*. 2019 Jul 25;9(8):157. doi: 10.3390/metabo9080157. PMID: 31349744; PMCID: PMC6724137.

65. 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. XBP1S Regulates MUC5B in a Promoter Variant-Dependent Pathway in Idiopathic Pulmonary Fibrosis Airway Epithelia. *Am J Respir Crit Care Med*. 2019 Jul 15;200(2):220-234. doi: 10.1164/rccm.201810-1972OC. Erratum in: Am J Respir Crit Care Med. 2019 Oct 15;200(8):1074. PMID: 30973754; PMCID: PMC6635783.

66. 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. Systemic Markers of Inflammation in Smokers With Symptoms Despite Preserved Spirometry in SPIROMICS. *Chest*. 2019 May;155(5):908-917. doi: 10.1016/j.chest.2018.12.022. Epub 2019 Jan 23. PMID: 30684474; PMCID: PMC6533449.

67. Okuda K, Chen G, Subramani DB, Wolf M, Gilmore RC, Kato T, Radicioni G, Kesimer M, Chua M, Dang H, Livraghi-Butrico A, Ehre C, Doerschuk CM, Randell SH, Matsui H, Nagase T, **O'Neal WK**, Boucher RC. Localization of Secretory Mucins MUC5AC and MUC5B in Normal/Healthy Human Airways. *Am J Respir Crit Care Med*. 2019 Mar 15;199(6):715-727. doi: 10.1164/rccm.201804-0734OC. PMID: 30352166; PMCID: PMC6423099.

68. Gong J, Wang F, Xiao B, Panjwani N, Lin F, Keenan K, Avolio J, Esmaeili 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. Genetic association and transcriptome integration identify contributing genes and tissues at cystic fibrosis modifier loci. *PLoS Genet*. 2019 Feb 26;15(2):e1008007. doi: 10.1371/journal.pgen.1008007. PMID: 30807572; PMCID: PMC6407791.

69. 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. An airway epithelial IL-17A response signature identifies a steroid-unresponsive COPD patient subgroup. *J Clin Invest*. 2019 Jan 2;129(1):169-181. doi: 10.1172/JCI121087. Epub 2018 Nov 26. PMID: 30383540; PMCID: PMC6307967.

70. Kesimer M, Smith BM, Ceppe A, Ford AA, Anderson WH, Barr RG, **O'Neal WK**, Boucher RC; SPIROMICS Investigative Group. Mucin Concentrations and Peripheral Airway Obstruction in Chronic Obstructive Pulmonary Disease. *Am J Respir Crit Care Med*. 2018 Dec 1;198(11):1453-1456. doi: 10.1164/rccm.201806-1016LE. PMID: 30130124; PMCID: PMC6290944.

71. 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. Association between acute respiratory disease events and the MUC5B promoter polymorphism in smokers. *Thorax*. 2018 Nov;73(11):1071-1074. doi: 10.1136/thoraxjnl-2017-211208. Epub 2018 Feb 13. PMID: 29440587; PMCID: PMC6089672.

72. 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. Role of Spdef in the Regulation of Muc5b Expression in

the Airways of Naive and Mucoobstructed Mice. *Am J Respir Cell Mol Biol*. 2018 Sep;59(3):383-396. doi: 10.1165/rcmb.2017-0127OC. PMID: 29579396; PMCID: PMC6189647.

73. 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; Current and former investigators of the SPIROMICS sites and reading centers. Rural Residence and Chronic Obstructive Pulmonary Disease Exacerbations. Analysis of the SPIROMICS Cohort. *Ann Am Thorac Soc*. 2018 Jul;15(7):808-816. doi: 10.1513/AnnalsATS.201710-837OC. PMID: 29584453; PMCID: PMC6207115.

74. 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. 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*. 2018 Jun;15(6):710-717. doi: 10.1513/AnnalsATS.201708-687OC. PMID: 30726108; PMCID: PMC6207135.

75. 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. Lower serum IgA is associated with COPD exacerbation risk in SPIROMICS. *PLoS One*. 2018 Apr 12;13(4):e0194924. doi: 10.1371/journal.pone.0194924. PMID: 29649230; PMCID: PMC5896903.

76. Saini Y, Lewis BW, Yu D, Dang H, Livraghi-Butrico A, Del Piero F, **O'Neal WK**, Boucher RC. Effect of LysM+ macrophage depletion on lung pathology in mice with chronic bronchitis. *Physiol Rep*. 2018 Apr;6(8):e13677. doi: 10.14814/phy2.13677. PMID: 29667749; PMCID: PMC5904692.

77. 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. Lung disease phenotypes caused by overexpression of combinations of α-, β-, and γ-subunits of the epithelial sodium channel in mouse airways. *Am J Physiol Lung Cell Mol Physiol*. 2018 Feb 1;314(2):L318-L331. doi: 10.1152/ajplung.00382.2017. Epub 2017 Oct 26. PMID: 29074490; PMCID: PMC5866504.

78. 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. 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*. 2018 Jan 24;5(1):46-56. doi: 10.15326/jcopdf.5.1.2017.0141. PMID: 29629404; PMCID: PMC5870739.

79. Rowson-Hodel AR, Wald JH, Hatakeyama J, **O'Neal WK**, Stonebraker JR, VanderVorst K, Saldana MJ, Borowsky AD, Sweeney C, Carraway KL 3rd. Membrane Mucin Muc4 promotes blood cell association with tumor cells and mediates efficient metastasis in a mouse model of breast cancer. *Oncogene*. 2018 Jan 11;37(2):197-207. doi: 10.1038/onc.2017.327. Epub 2017 Sep 11. PMID: 28892049; PMCID: PMC5930013.

80. 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. Alveolar eosinophilia in current smokers with chronic obstructive pulmonary disease in the SPIROMICS cohort. *J Allergy Clin Immunol*. 2018 Jan;141(1):429-432. doi: 10.1016/j.jaci.2017.07.039. Epub 2017 Sep 12. PMID: 28916185; PMCID: PMC5758424.

81. Polineni D, Dang H, Gallins PJ, Jones LC, Pace RG, Stonebraker JR, Commander LA, Krenicky JE, Zhou YH, Corvol H, Cutting GR, Drumm ML, Strug LJ, Boyle MP, Durie PR, Chmiel JF, Zou F, Wright FA, **O'Neal WK**, Knowles MR. Airway Mucosal Host Defense Is Key to Genomic Regulation of Cystic Fibrosis Lung Disease Severity. *Am J Respir Crit Care Med*. 2018 Jan 1;197(1):79-93. doi: 10.1164/rccm.201701-0134OC. PMID: 28853905; PMCID: PMC5765386.

82. 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. Electronic Cigarette Use in US Adults at Risk for or with COPD: Analysis from Two Observational Cohorts. *J Gen*

Intern Med. 2017 Dec;32(12):1315-1322. doi: 10.1007/s11606-017-4150-7. Epub 2017 Sep 7. PMID: 28884423; PMCID: PMC5698219.

83. 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. 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. doi: 10.1016/S2213-2600(17)30432-0. Epub 2017 Nov 13. PMID: 29146301; PMCID: PMC5849066.

84. 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, Kazerooni EA, Kleerup EAC, Martinez FJ, **O'Neal WK**, Woodruff PG, Lin CL. Differentiation of quantitative CT imaging phenotypes in asthma versus COPD. *BMJ Open Respir Res*. 2017 Nov 9;4(1):e000252. doi: 10.1136/bmjresp-2017-000252. Erratum in: *BMJ Open Respir Res*. 2018 Mar 6;5(1):e000252corr1. PMID: 29435345; PMCID: PMC5687530.

85. Donoghue LJ*, Livraghi-Butrico A*, McFadden KM, Thomas JM, Chen G, Grubb BR, **O'Neal WK**, Boucher RC, Kelada SNP. Identification of trans Protein QTL for Secreted Airway Mucins in Mice and a Causal Role for Bpifb1. *Genetics*. 2017 Oct;207(2):801-812. doi: 10.1534/genetics.117.300211. Epub 2017 Aug 29. PMID: 28851744; PMCID: PMC5629340.

86. 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. Variability in objective and subjective measures affects baseline values in studies of patients with COPD. *PLoS One*. 2017 Sep 21;12(9):e0184606. doi: 10.1371/journal.pone.0184606. PMID: 28934249; PMCID: PMC5608200.

87. Kesimer M, Ford AA, Ceppe A, Radicioni G, Cao R, Davis CW, Doerschuk CM, Alexis NE, Anderson WH, Henderson AG, Barr RG, Bleecker ER, Christenson SA, Cooper CB, Han MK, Hansel NN, Hastie AT, Hoffman EA, Kanner RE, Martinez F, Paine R 3rd, Woodruff PG, **O'Neal WK**, Boucher RC. Airway Mucin Concentration as a Marker of Chronic Bronchitis. *N Engl J Med*. 2017 Sep 7;377(10):911-922. doi: 10.1056/NEJMoa1701632. PMID: 28877023; PMCID: PMC5706541.

88. 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. Frequency of exacerbations in patients with chronic obstructive pulmonary disease: an analysis of the SPIROMICS cohort. *Lancet Respir Med*. 2017 Aug;5(8):619-626. doi: 10.1016/S2213-2600(17)30207-2. Epub 2017 Jun 28. PMID: 28668356; PMCID: PMC5558856.

89. Livraghi-Butrico A, Grubb BR, Wilkinson KJ, Volmer AS, Burns KA, Evans CM, **O'Neal WK***, Boucher RC*. Contribution of mucus concentration and secreted mucins Muc5ac and Muc5b to the pathogenesis of muco-obstructive lung disease. *Mucosal Immunol*. 2017 May;10(3):829. doi: 10.1038/mi.2017.29. PMID: 28435155. ***co-senior authors; equal contribution**. Correction: *Mucosal Immunol*. 2017;10(3):829. PMID: 28435155.

90. 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. Cigarette smoke disrupts monolayer integrity by altering epithelial cell-cell adhesion and cortical tension. *Am J Physiol Lung Cell Mol Physiol*. 2017 Sep 1;313(3):L581-L591. doi: 10.1152/ajplung.00074.2017. Epub 2017 Jun 22. PMID: 28642260; PMCID: PMC5625260.

91. **O'Neal WK**. Lung Cell-Specific Cre Deleter Mouse Strains: Going Back to Move Forward. *Am J Respir Cell Mol Biol*. 2017 Aug;57(2):149-150. doi: 10.1165/rcmb.2017-0099ED. PMID: 28762768.

92. Dang H, Gallins PJ, Pace RG, Guo XL, Stonebraker JR, Corvol H, Cutting GR, Drumm ML, Strug LJ, Knowles MR, **O'Neal WK**. Corrigendum: Novel variation at chr11p13 associated with cystic fibrosis lung disease severity. *Hum Genome Var*. 2017 May 25;4:17016. doi: 10.1038/hgv.2017.16. Erratum for*: Hum Genome Var*. 2016 Jul 07;3:16020. PMID: 28674633; PMCID: PMC5477836.

93. Keene JD, Jacobson S, Kechris K, Kinney GL, Foreman MG, Doerschuk CM, Make BJ, Curtis JL, Rennard SI, Barr RG, Bleecker ER, Kanner RE, Kleerup EC, Hansel NN, Woodruff PG, Han MK, Paine R

3rd, Martinez FJ, Bowler RP*, **O'Neal WK***; COPDGene and SPIROMICS Investigators ‡. Biomarkers Predictive of Exacerbations in the SPIROMICS and COPDGene Cohorts. *Am J Respir Crit Care Med*. 2017 Feb 15;195(4):473-481. doi: 10.1164/rccm.201607-1330OC. PMID: 27579823; PMCID: PMC5378424. ***co-senior authors; equal contribution**.

94. 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, Kleerup 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. Common Genetic Polymorphisms Influence Blood Biomarker Measurements in COPD. *PLoS Genet*. 2016 Aug 17;12(8):e1006011. doi: 10.1371/journal.pgen.1006011. PMID: 27532455; PMCID: PMC4988780. ***co-senior/corresponding authors**.

95. Yu D, Davis RM, Aita M, Burns KA, Clapp PW, Gilmore RC, Chua M, **O'Neal WK**, Schlegel R, Randell SH, C Boucher R. Characterization of Rat Meibomian Gland Ion and Fluid Transport. *Invest Ophthalmol Vis Sci*. 2016 Apr 1;57(4):2328-43. doi: 10.1167/iovs.15-17945. PMID: 27127933; PMCID: PMC4855829.

96. Saini Y, Wilkinson KJ, Terrell KA, Burns KA, Livraghi-Butrico A, Doerschuk CM, **O'Neal WK**, Boucher RC. Neonatal Pulmonary Macrophage Depletion Coupled to Defective Mucus Clearance Increases Susceptibility to Pneumonia and Alters Pulmonary Immune Responses. *Am J Respir Cell Mol Biol*. 2016 Feb;54(2):210-21. doi: 10.1165/rcmb.2014-0111OC. PMID: 26121027; PMCID: PMC4821038.

97. Lubamba BA, Jones LC, **O'Neal WK**, Boucher RC, Ribeiro CM. X-Box-Binding Protein 1 and Innate Immune Responses of Human Cystic Fibrosis Alveolar Macrophages. *Am J Respir Crit Care Med*. 2015 Dec 15;192(12):1449-61. doi: 10.1164/rccm.201504-0657OC. PMID: 26331676; PMCID: PMC4731720.

98. 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. Genome-wide association meta-analysis identifies five modifier loci of lung disease severity in cystic fibrosis. *Nat Commun*. 2015 Sep 29;6:8382. doi: 10.1038/ncomms9382. PMID: 26417704; PMCID: PMC4589222.

99. **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. Gene expression in transformed lymphocytes reveals variation in endomembrane and HLA pathways modifying cystic fibrosis pulmonary phenotypes. *Am J Hum Genet*. 2015 Feb 5;96(2):318-28. doi: 10.1016/j.ajhg.2014.12.022. Epub 2015 Jan 29. PMID: 25640674; PMCID: PMC4320265.

100. 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. 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*. 2015 Jan 27;13:19. doi: 10.1186/s12967-014-0374-z. PMID: 25622723; PMCID: PMC4314767.

101. Fossum SL, Mutolo MJ, Yang R, Dang H, **O'Neal WK**, Knowles MR, Leir SH, Harris A. Ets homologous factor regulates pathways controlling response to injury in airway epithelial cells. *Nucleic Acids Res*. 2014 Dec 16;42(22):13588-98. doi: 10.1093/nar/gku1146. Epub 2014 Nov 20. PMID: 25414352; PMCID: PMC4267623.

102. Guo X, Pace RG, Stonebraker JR, **O'Neal WK**, Knowles MR. Meconium ileus in cystic fibrosis is not linked to central repetitive region length variation in MUC1, MUC2, and MUC5AC. *J Cyst Fibros*. 2014 Dec;13(6):613-6. doi: 10.1016/j.jcf.2014.05.005. Epub 2014 Jun 8. PMID: 24920497; PMCID: PMC4253052.

103. 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. The association of plasma biomarkers with computed tomography-assessed emphysema phenotypes. *Respir Res*. 2014 Oct 12;15(1):127. doi: 10.1186/s12931-014-0127-9. PMID: 25306249; PMCID: PMC4198701.

104. Saini Y, Dang H, Livraghi-Butrico A, Kelly EJ, Jones LC, **O'Neal WK**, Boucher RC. Gene expression in whole lung and pulmonary macrophages reflects the dynamic pathology associated with airway surface dehydration. *BMC Genomics*. 2014 Sep 10;15(1):726. doi: 10.1186/1471-2164-15-726. PMID: 25204199; PMCID: PMC4247008.

105. Bove PF, Dang H, Cheluvaraju C, Jones LC, Liu X, **O'Neal WK**, Randell SH, Schlegel R, Boucher RC. Breaking the in vitro alveolar type II cell proliferation barrier while retaining ion transport properties. *Am J Respir Cell Mol Biol*. 2014 Apr;50(4):767-76. doi: 10.1165/rcmb.2013-0071OC. PMID: 24191670; PMCID: PMC4068919.

106. 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. Muc5b is required for airway defence. *Nature*. 2014 Jan 16;505(7483):412-6. doi: 10.1038/nature12807. Epub 2013 Dec 8. PMID: 24317696; PMCID: PMC4001806.

107. **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 SM; SPIROMICS Investigators. 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 Transl Med*. 2014 Jan 8;12:9. doi: 10.1186/1479-5876-12-9. PMID: 24397870; PMCID: PMC3928911.

108. 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*. Genome reference and sequence variation in the large repetitive central exon of human MUC5AC. Am J Respir Cell Mol Biol. 2014 Jan;50(1):223-32. doi: 10.1165/rcmb.2013-0235OC. PMID: 24010879; PMCID: PMC3930937. ***Equal Co-Senior authors.**

109. Sesma JI, Kreda SM, Okada SF, van Heusden C, Moussa L, Jones LC, **O'Neal WK**, Togawa N, Hiasa M, Moriyama Y, Lazarowski ER. Vesicular nucleotide transporter regulates the nucleotide content in airway epithelial mucin granules. *Am J Physiol Cell Physiol*. 2013 May 15;304(10):C976-84. doi: 10.1152/ajpcell.00371.2012. Epub 2013 Mar 6. Erratum in: *Am J Physiol Cell Physiol*. 2014 Feb 15;306(4):C415. PMID: 23467297; PMCID: PMC3651637.

110. Martino MB, Jones L, Brighton B, Ehre C, Abdulah L, Davis CW, Ron D, **O'Neal WK**, Ribeiro CM. The ER stress transducer IRE1β is required for airway epithelial mucin production. *Mucosal Immunol*. 2013 May;6(3):639-54. doi: 10.1038/mi.2012.105. Epub 2012 Nov 21. PMID: 23168839; PMCID: PMC4031691.

111. Livraghi-Butrico A, Kelly EJ, Wilkinson KJ, Rogers TD, Gilmore RC, Harkema JR, Randell SH, Boucher RC, **O'Neal WK**, Grubb BR. Loss of Cftr function exacerbates the phenotype of Na(+) hyperabsorption in murine airways. *Am J Physiol Lung Cell Mol Physiol*. 2013 Apr 1;304(7):L469-80. doi: 10.1152/ajplung.00150.2012. Epub 2013 Feb 1. PMID: 23377346; PMCID: PMC3627939.

112. Ehre C, Worthington EN, Liesman RM, Grubb BR, Barbier D, **O'Neal WK**, Sallenave JM, Pickles RJ, Boucher RC. Overexpressing mouse model demonstrates the protective role of Muc5ac in the lungs. *Proc Natl Acad Sci U S A*. 2012 Oct 9;109(41):16528-33. doi: 10.1073/pnas.1206552109. Epub 2012 Sep 24. Erratum in: Proc Natl Acad Sci U S A. 2014 Apr 15;111(15):5753. PMID: 23012413; PMCID: PMC3478656.

113. Livraghi-Butrico A, Kelly EJ, Klem ER, Dang H, Wolfgang MC, Boucher RC, Randell SH, **O'Neal WK**. Mucus clearance, MyD88-dependent and MyD88-independent immunity modulate lung susceptibility to spontaneous bacterial infection and inflammation. *Mucosal Immunol*. 2012 Jul;5(4):397-408. doi: 10.1038/mi.2012.17. Epub 2012 Mar 14. PMID: 22419116; PMCID: PMC3377774.

114. 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, Boëlle PY, Cutting GR, Knowles MR, Durie PR, Strug LJ. Multiple apical plasma membrane constituents are associated with

susceptibility to meconium ileus in individuals with cystic fibrosis. *Nat Genet*. 2012 May;44(5):562-9. doi: 10.1038/ng.2221. PMID: 22466613; PMCID: PMC3371103.

115. Livraghi-Butrico A, Grubb BR, Kelly EJ, Wilkinson KJ, Yang H, Geiser M, Randell SH, Boucher RC, **O'Neal WK**. Genetically determined heterogeneity of lung disease in a mouse model of airway mucus obstruction. *Physiol Genomics*. 2012 Apr 15;44(8):470-84. doi: 10.1152/physiolgenomics.00185.2011. Epub 2012 Mar 6. PMID: 22395316; PMCID: PMC3339860.

116. Jones LC, Moussa L, Fulcher ML, Zhu Y, Hudson EJ, **O'Neal WK**, Randell SH, Lazarowski ER, Boucher RC, Kreda SM. VAMP8 is a vesicle SNARE that regulates mucin secretion in airway goblet cells. *J Physiol*. 2012 Feb 1;590(3):545-62. doi: 10.1113/jphysiol.2011.222091. Epub 2011 Dec 5. PMID: 22144578; PMCID: PMC3379700.

117. Grubb BR, **O'Neal WK**, Ostrowski LE, Kreda SM, Button B, Boucher RC. Transgenic hCFTR expression fails to correct β-ENaC mouse lung disease. *Am J Physiol Lung Cell Mol Physiol*. 2012 Jan 15;302(2):L238-47. doi: 10.1152/ajplung.00083.2011. Epub 2011 Oct 14. PMID: 22003093; PMCID: PMC3349361.

118. Nguyen Y, Procario MC, Ashley SL, **O'Neal WK**, Pickles RJ, Weinberg JB. Limited effects of Muc1 deficiency on mouse adenovirus type 1 respiratory infection. *Virus Res*. 2011 Sep;160(1-2):351-9. doi: 10.1016/j.virusres.2011.07.012. Epub 2011 Jul 26. PMID: 21816184; PMCID: PMC3163747.

119. 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. Rho signaling regulates pannexin 1-mediated ATP release from airway epithelia. *J Biol Chem*. 2011 Jul 29;286(30):26277-86. doi: 10.1074/jbc.M111.260562. Epub 2011 May 23. PMID: 21606493; PMCID: PMC3143590.

120. 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, Paré 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. Genome-wide association and linkage identify modifier loci of lung disease severity in cystic fibrosis at 11p13 and 20q13.2. *Nat Genet*. 2011 Jun;43(6):539-46. doi: 10.1038/ng.838. Epub 2011 May 22. PMID: 21602797; PMCID: PMC3296486.

121. Guo X, Pace RG, Stonebraker JR, Commander CW, Dang AT, Drumm ML, Harris A, Zou F, Swallow DM, Wright FA, **O'Neal WK***, Knowles MR*. Mucin variable number tandem repeat polymorphisms and severity of cystic fibrosis lung disease: significant association with MUC5AC. *PLoS One*. 2011;6(10):e25452. doi: 10.1371/journal.pone.0025452. Epub 2011 Oct 6. PMID: 21998660; PMCID: PMC3188583. *** Co-Senior authors.**

122. Bove PF, Grubb BR, Okada SF, Ribeiro CM, Rogers TD, Randell SH, **O'Neal WK**, Boucher RC. Human alveolar type II cells secrete and absorb liquid in response to local nucleotide signaling. *J Biol Chem*. 2010 Nov 5;285(45):34939-49. doi: 10.1074/jbc.M110.162933. Epub 2010 Aug 27. PMID: 20801871; PMCID: PMC2966108.

123. Greenberg J, Deshmukh R, Huang L, Mostafa J, LaVange L, Carretta E, **O'Neal W.** The COPD Ontology and toward empowering clinical scientists as ontology engineers. *J Library Metadata.* 2010 Oct 20;10:173-87. doi: 10.1080/19386389.2010.520604.

124. 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, Grubb BR, Boucher RC. Airway surface liquid volume regulation determines different airway phenotypes in liddle compared with betaENaC-overexpressing mice. *J Biol Chem*. 2010 Aug 27;285(35):26945-26955. doi: 10.1074/jbc.M110.151803. Epub 2010 Jun 21. PMID: 20566636; PMCID: PMC2930694.

125. Ostrowski LE, Yin W, Rogers TD, Busalacchi KB, Chua M, **O'Neal WK**, Grubb BR. Conditional deletion of dnaic1 in a murine model of primary ciliary dyskinesia causes chronic rhinosinusitis. *Am J Respir Cell Mol Biol*. 2010 Jul;43(1):55-63. doi: 10.1165/rcmb.2009-0118OC. Epub 2009 Aug 12. PMID: 19675306; PMCID: PMC2911571.

126. Kreda SM, Seminario-Vidal L, van Heusden CA, **O'Neal W**, Jones L, Boucher RC, Lazarowski ER. Receptor-promoted exocytosis of airway epithelial mucin granules containing a spectrum of adenine nucleotides. *J Physiol*. 2010 Jun 15;588(Pt 12):2255-67. doi: 10.1113/jphysiol.2009.186643. Epub 2010 Apr 26. PMID: 20421285; PMCID: PMC2911224.

127. Cholon DM, **O'Neal WK**, Randell SH, Riordan JR, Gentzsch M. Modulation of endocytic trafficking and apical stability of CFTR in primary human airway epithelial cultures. *Am J Physiol Lung Cell Mol Physiol*. 2010 Mar;298(3):L304-14. doi: 10.1152/ajplung.00016.2009. Epub 2009 Dec 11. PMID: 20008117; PMCID: PMC2838667.

128. Seminario-Vidal L, Kreda S, Jones L, **O'Neal W**, Trejo J, Boucher RC, Lazarowski ER. Thrombin promotes release of ATP from lung epithelial cells through coordinated activation of rho- and Ca2+-dependent signaling pathways. *J Biol Chem*. 2009 Jul 31;284(31):20638-48. doi: 10.1074/jbc.M109.004762. Epub 2009 May 12. PMID: 19439413; PMCID: PMC2742828.

129. Ribeiro CM, Hurd H, Wu Y, Martino ME, Jones L, Brighton B, Boucher RC, **O'Neal WK**. Azithromycin treatment alters gene expression in inflammatory, lipid metabolism, and cell cycle pathways in well-differentiated human airway epithelia. *PLoS One*. 2009 Jun 5;4(6):e5806. doi: 10.1371/journal.pone.0005806. PMID: 19503797; PMCID: PMC2688381.

130. Kesimer M, Scull M, Brighton B, DeMaria G, Burns K, **O'Neal W**, Pickles RJ, Sheehan JK. Characterization of exosome-like vesicles released from human tracheobronchial ciliated epithelium: a possible role in innate defense. *FASEB J*. 2009 Jun;23(6):1858-68. doi: 10.1096/fj.08-119131. Epub 2009 Feb 3. PMID: 19190083; PMCID: PMC2698655.

131. Martino ME, Olsen JC, Fulcher NB, Wolfgang MC, **O'Neal WK**, Ribeiro CM. Airway epithelial inflammation-induced endoplasmic reticulum Ca2+ store expansion is mediated by X-box binding protein-1. *J Biol Chem*. 2009 May 29;284(22):14904-13. doi: 10.1074/jbc.M809180200. Epub 2009 Mar 25. PMID: 19321437; PMCID: PMC2685672.

132. Sesma JI, Esther CR Jr, Kreda SM, Jones L, **O'Neal W**, Nishihara S, Nicholas RA, Lazarowski ER. Endoplasmic reticulum/golgi nucleotide sugar transporters contribute to the cellular release of UDP-sugar signaling molecules. *J Biol Chem*. 2009 May 1;284(18):12572-83. doi: 10.1074/jbc.M806759200. Epub 2009 Mar 10. PMID: 19276090; PMCID: PMC2673323.

133. Livraghi A, Grubb BR, Hudson EJ, Wilkinson KJ, Sheehan JK, Mall MA, **O'Neal WK**, Boucher RC, Randell SH. Airway and lung pathology due to mucosal surface dehydration in {beta}-epithelial Na+ channel-overexpressing mice: role of TNF-{alpha} and IL-4R{alpha} signaling, influence of neonatal development, and limited efficacy of glucocorticoid treatment. *J Immunol*. 2009 Apr 1;182(7):4357-67. doi: 10.4049/jimmunol.0802557. PMID: 19299736; PMCID: PMC2659461.

134. Rock JR, **O'Neal WK**, Gabriel SE, Randell SH, Harfe BD, Boucher RC, Grubb BR. Transmembrane protein 16A (TMEM16A) is a Ca2+-regulated Cl- secretory channel in mouse airways. *J Biol Chem*. 2009 May 29;284(22):14875-80. doi: 10.1074/jbc.C109.000869. Epub 2009 Apr 10. PMID: 19363029; PMCID: PMC2685669.

135. Mall MA, Harkema JR, Trojanek JB, Treis D, Livraghi A, Schubert S, Zhou Z, Kreda SM, Tilley SL, Hudson EJ, **O'Neal WK**, Boucher RC. Development of chronic bronchitis and emphysema in beta-epithelial Na+ channel-overexpressing mice. *Am J Respir Crit Care Med*. 2008 Apr 1;177(7):730-42. doi: 10.1164/rccm.200708-1233OC. Epub 2007 Dec 13. PMID: 18079494; PMCID: PMC2277210.

136. Rowe SM, Jackson PL, Liu G, Hardison M, Livraghi A, Solomon GM, McQuaid DB, Noerager BD, Gaggar A, Clancy JP, **O'Neal W**, Sorscher EJ, Abraham E, Blalock JE. Potential role of high-mobility group box 1 in cystic fibrosis airway disease. *Am J Respir Crit Care Med*. 2008 Oct 15;178(8):822-31. doi: 10.1164/rccm.200712-1894OC. Epub 2008 Jul 24. PMID: 18658107; PMCID: PMC2566793.

137. Kreda SM, Okada SF, van Heusden CA, **O'Neal W**, Gabriel S, Abdullah L, Davis CW, Boucher RC, Lazarowski ER. Coordinated release of nucleotides and mucin from human airway epithelial Calu-3 cells. *J Physiol*. 2007 Oct 1;584(Pt 1):245-59. doi: 10.1113/jphysiol.2007.139840. Epub 2007 Jul 26. PMID: 17656429; PMCID: PMC2277076.

138. Ostrowski LE, Yin W, Diggs PS, Rogers TD, **O'Neal WK**, Grubb BR. Expression of CFTR from a ciliated cell-specific promoter is ineffective at correcting nasal potential difference in CF mice. *Gene Ther*. 2007 Oct;14(20):1492-501. doi: 10.1038/sj.gt.3302994. Epub 2007 Jul 19. PMID: 17637798.

139. Morin-Kensicki EM, Boone BN, Howell M, Stonebraker JR, Teed J, Alb JG, Magnuson TR, **O'Neal W**, Milgram SL. Defects in yolk sac vasculogenesis, chorioallantoic fusion, and embryonic axis elongation in mice with targeted disruption of Yap65. *Mol Cell Biol*. 2006 Jan;26(1):77-87. doi: 10.1128/MCB.26.1.77-87.2006. PMID: 16354681; PMCID: PMC1317614.

140. Neuringer IP, Sloan J, Budd S, Chalermskulrat W, Park RC, Stonebraker JR, **O'Neal WK**, Aris RM, Randell SH. Calcineurin inhibitor effects on growth and phenotype of human airway epithelial cells in vitro. *Am J Transplant*. 2005 Nov;5(11):2660-70. doi: 10.1111/j.1600-6143.2005.01071.x. PMID: 16212625.

141. Ribeiro CM, Paradiso AM, Schwab U, Perez-Vilar J, Jones L, **O'Neal W**, Boucher RC. Chronic airway infection/inflammation induces a Ca2+i-dependent hyperinflammatory response in human cystic fibrosis airway epithelia. *J Biol Chem*. 2005 May 6;280(18):17798-806. doi: 10.1074/jbc.M410618200. Epub 2005 Mar 3. PMID: 15746099.

142. Barnes AP, Livera G, Huang P, Sun C, **O'Neal WK**, Conti M, Stutts MJ, Milgram SL. Phosphodiesterase 4D forms a cAMP diffusion barrier at the apical membrane of the airway epithelium. *J Biol Chem*. 2005 Mar 4;280(9):7997-8003. doi: 10.1074/jbc.M407521200. Epub 2004 Dec 15. PMID: 15611099.

143. Stonebraker JR, Wagner D, Lefensty RW, Burns K, Gendler SJ, Bergelson JM, Boucher RC, **O'Neal WK**, Pickles RJ. Glycocalyx restricts adenoviral vector access to apical receptors expressed on respiratory epithelium in vitro and in vivo: role for tethered mucins as barriers to lumenal infection. *J Virol*. 2004 Dec;78(24):13755-68. doi: 10.1128/JVI.78.24.13755-13768.2004. PMID: 15564484; PMCID: PMC533903.

144. Okada SF, **O'Neal WK**, Huang P, Nicholas RA, Ostrowski LE, Craigen WJ, Lazarowski ER, Boucher RC. Voltage-dependent anion channel-1 (VDAC-1) contributes to ATP release and cell volume regulation in murine cells. *J Gen Physiol*. 2004 Nov;124(5):513-26. doi: 10.1085/jgp.200409154. Epub 2004 Oct 11. PMID: 15477379; PMCID: PMC2234005.

145. Mall M, Grubb BR, Harkema JR, **O'Neal WK**, Boucher RC. Increased airway epithelial Na+ absorption produces cystic fibrosis-like lung disease in mice. *Nat Med*. 2004 May;10(5):487-93. doi: 10.1038/nm1028. Epub 2004 Apr 11. PMID: 15077107.

146. Zariwala M*, **O'Neal WK***, Noone PG, Leigh MW, Knowles MR, Ostrowski LE. Investigation of the possible role of a novel gene, DPCD, in primary ciliary dyskinesia. *Am J Respir Cell Mol Biol*. 2004 Apr;30(4):428-34. doi: 10.1165/rcmb.2003-0338RC. Epub 2003 Nov 20. PMID: 14630615. ***authors contributed equally to manuscript.**

147. Coakley RD, Grubb BR, Paradiso AM, Gatzy JT, Johnson LG, Kreda SM, **O'Neal WK**, Boucher RC. Abnormal surface liquid pH regulation by cultured cystic fibrosis bronchial epithelium. *Proc Natl Acad Sci U S A*. 2003 Dec 23;100(26):16083-8. doi: 10.1073/pnas.2634339100. Epub 2003 Dec 10. PMID: 14668433; PMCID: PMC307696.

148. Ostrowski LE, Hutchins JR, Zakel K, **O'Neal WK**. Targeting expression of a transgene to the airway surface epithelium using a ciliated cell-specific promoter. *Mol Ther*. 2003 Oct;8(4):637-45. doi: 10.1016/s1525-0016(03)00221-1. PMID: 14529837.

149. Zhang YJ, **O'Neal WK**, Randell SH, Blackburn K, Moyer MB, Boucher RC, Ostrowski LE. 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*. 2002 May 17;277(20):17906-15. doi: 10.1074/jbc.M200348200. Epub 2002 Mar 4. PMID: 11877439.

150. Morral N, **O'Neal WK**, Rice K, Leland MM, Piedra PA, Aguilar-Córdova E, Carey KD, Beaudet AL, Langston C. Lethal toxicity, severe endothelial injury, and a threshold effect with high doses of an adenoviral vector in baboons. *Hum Gene Ther*. 2002 Jan 1;13(1):143-54. doi: 10.1089/10430340152712692. PMID: 11779418.

151. Lazarowski ER, Rochelle LG, **O'Neal WK**, Ribeiro CM, Grubb BR, Zhang V, Harden TK, Boucher RC. Cloning and functional characterization of two murine uridine nucleotide receptors reveal a potential target for correcting ion transport deficiency in cystic fibrosis gallbladder. *J Pharmacol Exp Ther*. 2001 Apr;297(1):43-9. PMID: 11259526.

152. **O'Neal WK**, Rose E, Zhou H, Langston C, Rice K, Carey D, Beaudet AL. Multiple advantages of alpha-fetoprotein as a marker for in vivo gene transfer. *Mol Ther*. 2000 Dec;2(6):640-8. doi: 10.1006/mthe.2000.0198. PMID: 11124066.

153. **O'Neal WK**, Zhou H, Morral N, Langston C, Parks RJ, Graham FL, Kochanek S, Beaudet AL. Toxicity associated with repeated administration of first-generation adenovirus vectors does not occur with a helper-dependent vector. *Mol Med*. 2000 Mar;6(3):179-95. PMID: 10965494; PMCID: PMC1949942.

154. Morral N, **O'Neal W**, Rice K, Leland M, Kaplan J, Piedra PA, Zhou H, Parks RJ, Velji R, Aguilar-Córdova E, Wadsworth S, Graham FL, Kochanek S, Carey KD, Beaudet AL. 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*. 1999 Oct 26;96(22):12816-21. doi: 10.1073/pnas.96.22.12816. PMID: 10536005; PMCID: PMC23112.

155. Morris AP, Scott JK, Ball JM, Zeng CQ, **O'Neal WK**, Estes MK. NSP4 elicits age-dependent diarrhea and Ca(2+)mediated I(-) influx into intestinal crypts of CF mice. *Am J Physiol*. 1999 Aug;277(2):G431-44. doi: 10.1152/ajpgi.1999.277.2.G431. PMID: 10444458.

156. **O'Neal WK**, Zhou H, Morral N, Aguilar-Cordova E, Pestaner J, Langston C, Mull B, Wang Y, Beaudet AL, Lee B. Toxicological comparison of E2a-deleted and first-generation adenoviral vectors expressing alpha1-antitrypsin after systemic delivery. *Hum Gene Ther*. 1998 Jul 20;9(11):1587-98. doi: 10.1089/hum.1998.9.11-1587. PMID: 9694157.

157. Morral N, **O'Neal W**, Zhou H, Langston C, Beaudet A. 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*. 1997 Jul 1;8(10):1275-86. doi: 10.1089/hum.1997.8.10-1275. PMID: 9215744.

158. Zhou H, **O'Neal W**, Morral N, Beaudet AL. Development of a complementing cell line and a system for construction of adenovirus vectors with E1 and E2a deleted. *J Virol*. 1996 Oct;70(10):7030-8. doi: 10.1128/JVI.70.10.7030-7038.1996. PMID: 8794347; PMCID: PMC190753.

159. Hasty P, **O'Neal WK**, Liu KQ, Morris AP, Bebok Z, Shumyatsky GB, Jilling T, Sorscher EJ, Bradley A, Beaudet AL. Severe phenotype in mice with termination mutation in exon 2 of cystic fibrosis gene. *Somat Cell Mol Genet*. 1995 May;21(3):177-87. doi: 10.1007/BF02254769. PMID: 7482032.

160. **O'Neal WK**, Hasty P, McCray PB Jr, Casey B, Rivera-Pérez J, Welsh MJ, Beaudet AL, Bradley A. A severe phenotype in mice with a duplication of exon 3 in the cystic fibrosis locus. *Hum Mol Genet*. 1993 Oct;2(10):1561-9. doi: 10.1093/hmg/2.10.1561. PMID: 7505691.

161. Yorifugi T, **Lemna** (**O'Neal) WK**, Ballard CF, Rosenbloom CL, Roxmahel R, Plavsic N, Tsui L-C, Beaudet, AL. Molecular cloning and sequence analysis of murine cDNA for the cystic fibrosis transmembrane conductance regulator. *Genomics.* 1991 Jul;10(3):547-50. doi: 10.1016/0888-7543(91)90434-g. PMID: 1716243.

162. **Lemna** (**O'Neal) WK**, Feldman GL, Kerem B, Fernbach SD, Zevkovich EP, O'Brien WE, Riordan JR, Collins FS, Tsui, L-C, Beaudet, AL. Mutation analysis for heterozygote detection and the prenatal diagnosis of cystic fibrosis. *New Engl J Med.* 1990 Feb 1;322(5):291-6. doi: 10.1056/NEJM199002013220503. PMID: 2296270.

Published Abstracts *(from newest to oldest):*

1. Chen G, Sun L, Dang H, Hasegawa K, Volmer AS, Zhou L, Deshmukh AA, Gilmore RC, Guhr Lee TN, Snead JY, Livingston EW, Yuan H, Livraghi-Butrico A, Esther Jr CR, Okuda K, **O'Neal WK**, Boucher RC, Hagood JS. Epithelial Xbp1s Expression Leads to Distal Lung MUC5B Hyperproduction and Progression of Pulmonary Fibrosis Following Injury. *Am J Respir Crit Care Med.* 2024;209:A5186.

2. Fawzy A, Hansel NN, Putcha N, Curtis JL, Labaki WW, **O'Neal WK**, Martinez FJ, Tejwani V, Wells JM, Woodruff P, Barr RG, Pistenmaa CL. Platelet Activation Is Associated With Respiratory Exacerbation Rate and Symptoms in SPIROMICS. *Am J Respir Crit Care Med.* 2024;209:A5107.

3. Hawkins P, Leist SR, Schåfer A, Dang H, Gilmore RC, Rock JR, Okuda K, Pappu R, Pestka JJ, Harkema JR, Livraghi-Butrico A, Baric RS, **O'Neal WK**, Boucher RC. Epithelial IL33 Contributes to the Development of Post SARS-CoV-2 Eosinophilic Pneumonia in a Model of Chronic Bronchitis *Am J Respir Crit Care Med.* 2024;209:A6854.

4. Mikami Y, Van Heusden C, Dang H, Burns E, Ribeiro CM, Gilmore RC, Okuda K, Randell SH, **O'Neal WK**, Boucher RC. Interactions Between Airway Epithelial Hypoxia and Inflammatory Stimuli in Airways Disease. *Am J Respir Crit Care Med.* 2024;209:A3289.

5. Richert ME, Kong H, Balasubramanian S, Jang MK, Andargie T, Brower R, Bleecker ER, Bowler RP, Christenson SA, Meyers DA, **O'Neal WK**, Ortega VE, Woodruff P, Martinez FJ, Curtis JL, Agbor-Enoh S.

Cell-Free DNA to Predict Severe Exacerbations in Chronic Obstructive Pulmonary Disease. *Am J Respir Crit Care Med.* 2024;209:A2798.

6. Saito M, Asakura T, Gilmore RC, Chua M, Randell SH, **O'Neal WK**, Boucher RC. Morphological Analysis of Bronchiectasis in Primary Ciliary Dyskinesia. *Am J Respir Crit Care Med.* 2024;209:A4432.

7. Schworer S, Okuda K, Dang H, Markovetz M, Kato T, Asakura T, Chen G, Gilmore R, Van Heusden C, Chua M, Trejo Bittar H, Cody B, Trudeau J, **O'Neal WK**, Randell SH, Wenzel SE, Boucher RC. Transcriptional pathways in small airway basal cells define niches of goblet cell metaplasia in asthma. *Am J Respir Crit Care Med.* 2024;209:A5015.

8. Stevens NC, Clair GC, Cawley AM, van Heusden C, Wisniewski ZY, Singh A, Burns EF, Dang H, Randell SH, **O'Neal WK**, Mikami Y, Boucher RC, Okuda K. Regional Susceptibility to Hypoxia-induced Changes in Airway Lipid Composition and Implications in Muco-obstructive Lung Diseases. *Am J Respir Crit Care Med.* 2024;209:A6358.

9. Asakura T, Dang H, Okuda K, Chen G, Kato T, Mikami Y, Gilmore RC, Hawkins P, Schworer SA, Masugi Y, Hasegawa N, Randell SH, **O'Neal WK**, Boucher RC. Spatial Transcriptional Profiling of the Bronchiolectatic Lungs with Non-cystic Fibrosis Bronchiectasis (NCFB). *Am J Respir Crit Care Med.* 2023;207:A4239.

10. Cawley A, Nakano S, Singh A, Wisniewski Z, Dang H, Morton L, Gilmore R, Asakura T, Quinney N, Boyles S, Cholon D, Kelly S, Fulcher, Gentzch M, Randell S, **O'Neal W**, Boucher R, Okuda K. 201 SCGB3A2-positive distal airway secretory cells are a therapeutic target for CFTR function restoration. *J Cyst Fibros*. 2023;22:S105.

11. Gomez JC, Okuda K, Jones LC, Martin JR, Dang H, **O'Neal WK**, Doerschuk CM. Heterogeneity in Neutrophil Transcriptomes During Streptococcus Pneumoniae Pneumonia Suggests Diverse Neutrophil Functions During Infection. *Am J Respir Crit Care Med.* 2023;207:A4267.

12. Kato T, Asakura T, Dang H, Strickler ER, Okuda K, Randell SH, **O'Neal WK**, Boucher RC. Ectopic MUC5B Expression in SFTPC-Positive Alveolar Epithelial Cells in Subjects Without Lung Fibrosis. *Am J Respir Crit Care Med.* 2023;207:A2800.

13. Labaki WW, Huang YJ, Freeman CM, Gurczynski SJ, Erb-Downward J, Bowler RP, Barr RG, Comellas AP, Criner GJ, Wells JM, Hansel NN, Barjaktarevic I, Paine R, Krishnan JA, Peters SP, Woodruff P, **O'Neal WK**, Martinez FJ, Curtis JL, Stringer KA, Han MK. Serum Kynurenine/Tryptophan Ratio Is a Marker of Lung Disease Severity in Current and Former Tobacco Cigarette Users With and Without COPD. *Am J Respir Crit Care Med.* 2023;207:A4206.

14. Schworer S, Okuda K, Dang H, Gilmore R, **O'Neal W**, Randell S, Wenzel S, Boucher R. Mucus Plugging and Regional Heterogeneity are Features of Asthmatic Small Airways. *J Allergy Clin Immunol*. 2023;151(2):AB223.

15. Schworer S, Okuda K, Dang H, Kato T, Chen G, Gilmore R, Chua M, Trejo Bittar H, Cody B, Trudeau J, **O'Neal WK**, Randell SH, Wenzel SE, Boucher RC. Steroid-resistant Severe Asthma and Fatal Asthma Small Airways Are Defined by Mucus Plugging and Heterogeneous MUC5AC Expression. *Am J Respir Crit Care Med.* 2023;207:A4535.

16. Singh A, Nakano S, Cawley AM, Caitlin EE, Barnett KC, Brocke SA, Dang H, Takanori A, Gilmore RC, Morton LC, Randell SH, Pickles RJ, Ting JP, **O'Neal WK**, Baric RS, Boucher RC, Okuda K. The Role of the IL-1β Pathway in Severe Acute Respiratory Syndrome Coronavirus 2 Infection of Human Airway Epithelia. *Am J Respir Crit Care Med.* 2023;207:A3972.

17. Asakura T, Okuda K, Chen G, Kato T, Mikami Y, Rodney GC, Barbosa Cardenas SM, Chua M, Masugi Y, Noone PG, Ribeiro CM, Doerschuk CM, Olivier KN, Hasegawa N, Randell SH, **O'Neal WK**, Boucher RC. MUC5B-Dominated Mucus Hyperproduction in the Bronchiolectatic Airway of Non-Cystic Fibrosis Bronchiectasis (NCFB). *Am J Respir Crit Care Med*. 2022;205:A2003.

18. Gomez Ladron De Guevara D, Anderson WH, Couper D, Quibrera PM, Barjaktarevic IZ, Barr RG, Bhatt SP, Bleecker ER, Bowler R, Christenson S, Comellas AP, Curtis JL, Dransfield MB, Fortis S, Han MK, Hansel NN, Hoffman EA, Kanner RE, Kim V, Krishnan JA, Martinez FJ, Ortega VE, **O'Neal WK**, Paine III R, Peters SP, Putcha N, Rennard SI, Smith B, Tashkin DP, Woodruff PG, Cooper CB. SPIROMICS COPD Patients with Decline of Lung Function Have Thinner Airway Walls at Baseline and Are More Likely to Show Bronchodilator Responsiveness. *Am J Respir Crit Care Med*. 2022;205:A3727.

19. Kato T, Asakura T, Edwards CE, Dang H, Mikami Y, Okuda K, Chen G, Sun L, Gilmore RC, Hawkins PE, Hewitt SM, Chertow DS, NIH COVID-19 Autopsy Consortium, Borczuk A, Martinez FJ, Thorne LB, Askin FB, Ehre C, Randell SH, **O'Neal WK**, Baric RS, Boucher RC. Prevalence and mechanisms of bronchiolar mucus plugging in COVID-19 lung disease. *Am J Respir Crit Care Med*. 2022;205:A3612.

20. Kelada SN, Van Buren E, Radicioni G, **O'Neal WK**, Dang H, Kasela S, Garudadri S, Curtis JL, Han MK, Krishnan JA, Wan ES, Silverman EK, Hastie AT, Ortega VE, Lappalainen T, Christenson S, Li Y, Cho MH, Kesimer M. Genetic Regulators of Sputum Mucin Concentration Revealed by GWAS and Their Associations with Chronic Bronchitis and Acute Exacerbations of COPD. *Am J Respir Crit Care Med*. 2022;205:A4666.

21. Martin JR, Gomez JC, Kim YH, Volmer A, **O'Neal WK**, Gilmour MI, Doerschuk CM. The Effect of a Single Exposure to Simulated Burn Pit Products in Mice with Healthy and Inflamed Airways and Their Response to Influenza A. *Am J Respir Crit Care Med*. 2022;205:A2194.

22. Mikami Y, Grubb B, Rogers T, Dang H, Kota P, Asakura T, Vilar J, Gilmore R, Okuda K, Morton L, Chen G, Sun L, Gentzsch M, Randell SH, **O'Neal WK**, Boucher RC. Airway Obstruction Produces Chronic Hypoxia-Dependent Cellular Adaptation and Mucus Dehydration in Human Airway Epithelial Cells. *Am J Respir Crit Care Med*. 2022;205:A1999.

23. Okuda K, Dinnon III KH, Leist SR, Dang H, Gully K, De la Cruz G, Evangelista MD, Asakura T, Gilmore RC, Hawkins PE, Nakano S, A. West, Schaefer A, Gralinski LE, Zweigart MR, Dong S, McBride J, Cooley MR, Love MK, Sakthivel M, Lee YZ, Hagood JS, Mock JR, **O'Neal WK**, Montgomery SA, Boucher RC, Baric RS. Spatial Transcriptional Profiling of Chronic Pulmonary Disease in a Mouse-Adapted Model of SARS-CoV-2. *Am J Respir Crit Care Med*. 2022;205:A3621.

24. Stonebraker JR, Pace RG, Gallins PJ, Dang H, **O'Neal WK**, Blackman SM, Aksit MA, Faino AV, Bamshad MJ, Gibson RL, Cutting GR, Wright FA, Zhou YH, Durie PR, Ling SC, Knowles MR. Discovery of novel genetic variants in severe CF liver disease as determined by whole genome sequencing. *J Cyst Fibros*. 2022;21;S132-3.

25. Sun L, Quinney NL, Sears PR, Zhou L, Dizmond FU, Cholon DM, Rogers TD, Gilmore RC, Chang CX, Ceppe AS, Ramsey KA, Dang H, Esther CR, Chua M, Grubb BR, Kesimer M, Hill DB, Ostrowski LE, Button B, Gentzsch M, **O'Neal WK**, Freeman AF, Robinson C, Bogdanovski K, Olivier KN, Boucher RC, Chen G. STAT3 Mutations Lead to Defective Mucociliary Clearance in the Autosomal Dominant Hyper IgE Syndrome (AD-HIES) Airways. *Am J Respir Crit Care Med*. 2022;205:A1977.

26. Tejwani V, Putcha N, Woo H, Liu C, Lafon D, Dransfield MT, Comellas AP, Azar A, **O'Neal WK**, Barr RG, Ortega VE, , Paine R, Barjaktarevic I, Woodruff P, Bowler RP, Martinez FJ, Peters SP, Han MK, Ohar JA, Krishnan JA, Criner GJ, Alexis NE, Stampfli MR, Kaner RJ, Hansel NN, Freeman CM, Huang YJ, Curtis JL. Associations of Serum and Bronchoalveolar Immunoglobulins with Lung Microbiota Diversity, B-Cell Memory Phenotypes, and COPD Morbidity and Exacerbations. *Am J Respir Crit Care Med*. 2022;205: A3489.

27. Lemaster WB, Quibrera PM, Couper D, Tashkin DP, Bleecker ER, Doerschuk CM, Ortega VE, Cooper CB, Han MK, Woodruff P, **O'Neal WK**, W. H. Anderson, Alexis NE, Curtis JL, Bowler RP, Barr R, Kaner J, Dransfield MT, Paine R, Kim V, Martinez FJ, Hastie AT, Barjaktarevic I. Clinical Implications of Low Absolute Blood Eosinophil Count in the SPIROMICS COPD Cohort. *Am J Respir Crit Care Med*. 2021;203: A1245.

28. O'Beirne S, Salit J, Alexis NE, Arjomandi M, Barjaktarevic I, Basta P, Bowler RP, Christenson SA, Cloonan SM, Curtis JL, Doerschuk CM, Drummond MB, Hastie AT, Huang YJ, Kim V, **O'Neal WK**, Ortega VE, Postow L, Putcha N, Rennard SI, Wells JM, Woodruff P, Zhang WZ, Crystal RG, Kaner RJ. Secondhand Smoke Alters Alveolar Macrophage Gene Expression. *Am J Respir Crit Care Med.* 2021;203:A2318.

29. Ortega VE, Paulin LM, Kaufman JD, Ampleford E, Woo H, Hawkins GA, Li X, Barjaktarevic I, Bhatt SP, Bowler RP, Barr RG, Cooper CB, Couper D, Curtis JL, Gassett AJ, Han MK, Kanner RE, Kim V, Martinez FJ, Moore WC, **O'Neal WK**, Paine R, Smith BP, Smith B, Woodruff P, Hoffman EA, Peters SP, Meyers DA, Bleecker ER, Hansel NN. Air Pollutant Exposure-by-Gene Interactions Associated with Emphysema, Lung Structure, and Lung Function in SPIROMICS. *Am J Respir Crit Care Med*. 2021;203:A1095.

30. 27. Zhou Y, Gallins P, Pace R, Dang H, **O'Neal W**, Li Y, Ling H, Corvol H, Strug L, Bamshad M, Gibson R, Cutting G, Blackman S, Wright F, Knowles M. Genetic variants that modify severity of CF lung disease: Update from the CF Genome Project. *J Cyst Fibros*. 2021;20:S306.

31. Chen G, Sun L, Esther CR, Wolfgang M, Kesimer M, Hill D, Ramsey KA, Button B, Quinney N, Gentzsch M, **O'Neal WK**, Freeman A, Robinson C, Bogdanovski K, Olivier KN, Boucher RC. Defective Epithelial Mucociliary Differentiation Leads to Mucin Hyperconcentration in the Autosomal Dominant Hyper Ige Syndrome (AD-HIES) Airways. *Am J Respir Crit Care Med*. 2020;201:A6395.

32. Eastman AC, Pace RG, Dang H, Aksit MA, Vecchio-Pagán B, Lam AT, **O'Neal WK**, Blackman SM, Knowles MR, Cutting GR. SLC26A9 SNP rs7512462 is not associated with lung disease severity or lung function response to ivacaftor in cystic fibrosis patients with G551D-CFTR. *Pediatr Pulmonol*. 2020;55:S117-S118.

33. Esther CR, **O'Neal WK**, Koch AL, Cooper CB, Barjaktarevic I, Raffield L, Bowler RP, Comellas AP, Peters SP, Curtis JL, Ronish B, Ortega VE, Wells JM, Halper-Stromberg E, Rennard SI, Boucher RC. Prolonged, Physiologically Relevant Concentrations of Nicotine and Nicotine Metabolites in Airways of Smokers. *Am J Respir Crit Care Med*. 2020;201:A1907.

34. Fawzy A, Woo HJ, Balasubramanian A, Barjaktarevic I, Barr R, Bowler RP, Comellas AP, Criner GJ, Dransfield MT, Han MK, Hoffman EA, Kanner RE, Krishnan JA, **O'Neal WK**, Paine R, Peters SP, Woodruff P, Hansel NN, Putcha N. Phenotypic Characteristics and Clinical Outcomes Among Individuals with COPD and Polycythemia. *Am J Respir Crit Care Med*. 2020;201:A2855.

35. Freeman CM, Brown JP, Kady MR, Stolberg VR, Toma MS, Alexis NE, Arjomandi M, Barjaktarevic I, Barr R, Bleecker ER, Bowler RP, Christenson S, Comellas AP, Couper D, Criner GJ, Doerschuk CM, Dransfield MT, Han MK, Hastie AT, Hoffman EA, Kaner RJ, Kaner RJ, Krishnan JA, Labaki WW, Martinez FJ, Meyers DA, **O'Neal WK**, Paine R, Tejwani V, Wells JM, Woodruff P, Curtis JL. Intermediate (CD14++CD16+) Monocytes Correlate with Parametric Response Mapping of Functional Small Airways Disease: Spiromics Immunophenotyping Sub-Study. *Am J Respir Crit Care Med*. 2020;201:A1063.

36. Hoesterey D, Dang H, Barr R, Belperio JA, Bowler RP, Buhr RG, Couper D, Criner GJ, Doerschuk CM, Dransfield MT, Drummond M, Freeman CM, Han MK, Hansel NN, Hastie AT, Hoffman EA, Kaner RJ, Kanner RE, Kim V, Krishnan JA, Martinez FJ, **O'Neal WK**, Ortega VE, Paine R, Tashkin DP, Wells JM, Woodruff P, Curtis JL, Barjaktarevic I, Spiromics Investigators. Neutrophil to Lymphocyte Ratio (NLR) Is a Potential Biomarker in Clinically Stable Chronic Obstructive Pulmonary Disease: SPIROMICS Cohort. *Am J Respir Crit Care Med*. 2020;201:A2857.

37. Kato T, Okuda K, Cardenas BS, Gilmore RC, Thacker H, Goodell HP, Loznev HT, Radicioni G, Kesimer M, Randell S, Ballard ST, **O'Neal W**, Button B, Boucher R. Altered biophysical properties of SMG mucus and decreased prr4 in induced sputum suggest impaired SMG secretion in CF. *Pediatr Pulmonol*. 2020;55:S40.

38. Masters B, Curtis JL, Han MK, Martinez FJ, **O'Neal WK**, Freeman CM, Criner GJ, Dransfield MT, Bowler RP, Couper D, Labaki WW, Hansel NN, Peters SP, Woodruff P, Bhatt SP, Siler TM, Miller M, Herrera JE. Plasma GDF-15 Levels Improve Risk Assessments for Severe COPD Outcomes. *Am J Respir Crit Care Med*. 2020;201:A2860.

39. Morrison CB, Delion M, Markovetz MR, Quinney NL, Morton L, Gentzsch M, Hill DB, **O'Neal W**, Kesimer M, Ehre C. Modulator therapy normalizes CF mucus in vitro. *Pediatr Pulmonol*. 2020;55:S40.

40. Ngo D, Pratte KA, Flexeder C, Petersen H, Dang H, Ma Y, Keyes MJ, Petersonl BD, Sitars V, Gillenwater LA, Xu H, Emson C, Gieger C, Suhre K, Graumann J, Jain D, Conomos MP, Tracy RP, Guo X, Liu Y, Johnson W, Cornell E, Durda P, Taylor KD, Papanicolaou GJ, Rich SS, Rotter JI, Rennard SI, Curtis JL, Woodruff P, Comellas AP, Silverman EK, Crapo JD, Larson MG, Ramachandran V, Wang TJ, Gerszten RE, O'Connor GT, Barr R, Couper D, Dupuis J, Manichaikul A, **O'Neal WK**, Tesfaigzi Y, Schulz H, Bowler RP. Systemic Biomarkers of Lung Function and FEV1 Decline Across Multiple Cohorts. *Am J Respir Crit Care Med*. 2020;201:A5527.

41. O'Beirne SL, Salit J, Alexis NE, Arjomandi M, Barjaktarevic I, Bowler RP, Christenson S, Curtis JL, Doerschuk CM, Drummond M, Hastie AT, Huang YJ, Kim V, **O'Neal WK**, Ortega VE, Wells JM, Woodruff P, Basta P, Crystal RG, Kaner RJ, Spiromics Investigators. Current Smoking Down-Regulates Specific Alveolar Macrophage Host Defense Gene Expression Pathways. *Am J Respir Crit Care Med*. 2020;201:A1217.

42. Okuda K, Dang H, Kobayashi Y, Nakano S, Cardenas SMB, O'Neal VK, Kato T, Chen G, Gilmore RC, Quinney NL, Gentzsch M, Anderson C, Chua M, Andrew G, Randell S, Tata P, **O'Neal W**, Boucher R. Secretory cells dominate airway CFTR expression and function in superficial epithelia lining human airways. *Pediatr Pulmonol*. 2020;55:S63.

43. Okuda K, Kato T, Chen G, Gilmore RC, Chua M, Bittar HT, **O'Neal WK**, Wenzel SE, Boucher RC. Altered Secretory Mucin Expression in Systemic Steroid-Dependent Severe Asthma Small Airways*. Am J Respir Crit Care Med*. 2020;201:A7466.

44. Okuda K, Kobayashi Y, Dang H, Nakano S, Barbosa S, O'Neal VK, Kato T, Chen G, Gilmore RC, Quinney NL, Gentzsch M, Radicioni G, Kesimer M, Anderson C, Chua M, Ghio AJ, Randell SH, Tata P, **O'Neal WK**, Boucher RC. Mucin-Competent Secretory Cells Are a Major Cell Type for CFTR Expression in Normal Human Airway Epithelia. *Am J Respir Crit Care Med*. 2020;201:A2663.

45. Ortega VE, Barjaktarevic I, Li X, Ampleford E, **O'Neal WK**, Hawkins GA, Bowlers RP, Moore WC, Barr R, Cooper CB, Couper D, Han MK, Hansel NN, Kanner RE, Martinez FJ, Paine R, Woodruff P, Hoffman EA, Peters SP, Bleecker ER, Meyers DA, Nhlbi S. Genome-Wide Association Study of Short-Acting Bronchodilator Response Identifies Novel Pharmacogenetic Loci in SPIROMICS. *Am J Respir Crit Care Med*. 2020;201:A7826.

46. Ortega VE, Kim D, Moore WC, Zein JG, Li X, Ampleford E, **O'Neal WK**, Bowler RP, Barr R, Barjaktarevic I, Castro M, Cooper CB, Couper D, Denlinger LC, Erzurum SC, Fahy JV, Fitzpatrick AM, Gaston BM, Han MK, Hansel NN, Hastie AT, Hoffman EA, Israel E, Jarjour NN, Kanner RE, Martinez FJ, Mauger D, Paine R, Levy BD, Wenzel SE, Woodruff P, Peters SP, Bleecker ER, Meyers DA, Liggett SB, Hawkins GA, Nhlbi S, Nhlbi S. Deep Resequencing of the Beta-2 Adrenergic Receptor Gene (ADRB2) in COPD and Asthma Cohorts Identifies Functional Rare Variants. *Am J Respir Crit Care Med*. 2020;201:A4220.

47. Putcha S, Gu T, Murray S, Meldrum CA, Cardenas-Garcia J, Curtis JL, Martinez FJ, Criner GJ, Bowler RP, Barr R, Hoffman EA, Fawzy A, Putcha N, Hansel NN, Bleecker ER, Barjaktarevic I, Kanner RE, Paine R, Dransfield MT, Krishnan JA, Ortega VE, Peters SP, **O'Neal WK**, Arjomandi M, Woodruff P, Han MK, Labaki WW. Patient-Reported Clinical Variables Associated with Significant Air Trapping in Smokers: An Analysis of the SPIROMICS Cohort. *Am J Respir Crit Care Med*. 2020;201:A4564.

48. Vameghestahbanati M, Hoffman EA, Couper D, Hansel NN, Oelsner E, Sack C, Woodruff P, **O'Neal WK**, Hamid Q, Barr R, Smith BM. Airway Tree Structure Modifies Cigarette Smoke-Associated Lung Function Decline. *Am J Respir Crit Care Med*. 2020;201:A6407.

49. Winters C, Radicioni G, Wilkinson K, Kesimer M, Boucher R, **O'Neal W**, Livraghi-Butrico A. Age and host microbiota affect the abundance and composition of airway mucus. *Pediatr Pulmonol*. 2020;55:S45-S6.

50. Yee N, Markovic D, Buhr RG, Tashkin DP, Bhatt SP, Fortis S, Arjomandi M, Ronish B, Couper D, Anderson WH, Kanner RE, Paine R, Kaner RJ, **O'Neal WK**, Woodruff P, Han MK, Martinez FJ, Barr R, Wells JM, Ortega VE, Hoffman EA, Kim V, Drummond M, Bowler RP, Criner GJ, Curtis JL, Coopers CB, Barjaktarevic I. Significance of FEV3/FEV6 in Recognition of Early Airway Disease in Smokers at Risk of Development of COPD: Analysis of the SPIROMICS Cohort. *Am J Respir Crit Care Med*. 2020;201:A6418.

51. Zhou Y, Gallins P, Pace RG, Dang H, **O'Neal W**, Li Y, Ling H, Bamshad M, Gibson R, Cutting GR, Blackman SM, Wright F, Knowles M. Genetic modifiers of cystic fibrosis lung disease: Initial results from the Cystic Fibrosis Genome Project. *Pediatr Pulmonol*. 2020;55:S117.

52. 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. Accelerated ATP hydrolysis in airway surface liquid (ASL) provides a mechanism for mucus dehydration in chronic bronchitis. *Am J Respir Crit Care Med*. 2019;199:A3847.

53. 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. Altered alveolar macrophage expression of Axl and Mertk in COPD: SPIROMICS Immunophenotyping Sub-Study. *Am J Respir Crit Care Med*. 2019;199:A4040.

54. 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. 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.* 2019;199:A4858.

55. Chen G, Sun L, Kato T, Okuda K, Martino ME, Abzhanova A, Lin JM, Gilmore RC, Batson B, O'Neal VK, Volmer A, Dang H, Deng Y, Randell SH, Button B, Livraghi-Butrico A, Kesimer M, Ribeiro CM, **O'Neal W**, Boucher R. IL1 beta is the dominant pro-mucin secretory cytokine in CF airway secretions. *Pediatr Pulmonol*. 2019;54:S182-S3.

56. Cmielewski P, McCarron A, Reyne N, Finnie J, Rout-Pitt N, Schjenken J, Chen H, McIntyre C, Craig F, **O'Neal W**, Parsons D, Donnelley M. An update on the phenotype characterisation of Phe508del and CFTR knockout rats. *Pediatr Pulmonol*. 2019;54:S168.

57. Dang H, Polineni D, Pace RG, Stonebraker JR, Corvol H, Cutting GR, Drumm ML, Strug LJ, **O'Neal W**, Knowles M. Novel CF modifier genes by expression imputation from GWAS and EQTL data. *Pediatr Pulmonol*. 2019;54:S243.

58. Esther CR, **O'Neal WK**, Anderson WH, Kesimer M, Hastie AT, Barr RG, Bowler RP, Wells JM, Oelsner E, Boucher RC. Impaired mucus hydration and hydration signaling pathways in COPD. *Am J Respir Crit Care Med.* 2019;199:A1126.

59. 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. 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.* 2019;199:A3849.

60. 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. Lung microbiota associations with clinical features of COPD in the SPIROMICS cohort. *Am J Respir Crit Care Med.* 2019;199:A5578.

61. Kamath D, Budden T, Kim M, Baumlin N, **O'Neal W**, Knowles M, Weinman S, Salathe M, Polineni D. Anti-inflammatory effects of methylthioadenosine in cystic fibrosis. *Pediatr Pulmonol*. 2019;54:S172.

62. Kato T, Okuda K, Cardenas SMB, Gilmore RC, Das J, Sun L, Lin JM, Nakano S, Chen G, Button B, Randell SH, Ballard ST, **O'Neal W**, Boucher R. Identification of submucosal gland-specific proteins as a marker of gland secretion. *Pediatr Pulmonol*. 2019;54:S179.

63. Kelada SN, Li Y, Van Buren E, Radicioni G, Ford A, **O'Neal WK**, Ortega VE, Woodruff P, Bleecker ER, Boucher RC, Kesimer M. Genome-wide association study of sputum mucin concentrations in the Subpopulations and Intermediate Outcomes in COPD Study (SPIROMICS). *Am J Respir Crit Care Med;*2019.199:A2149.

64. Okuda K, Kobayashi Y, Dang H, Nakano S, Cardenas SMB, O'Neal VK, Kato T, Chen G, Gilmore RC, Chua M, Livraghi-Butrico A, Doerschuk C, Grubb B, Randell SH, Tata P, **O'Neal W**, Boucher R. Regional regulation of CFTR and ionocyte expression in normal human airways. *Pediatr Pulmonol*. 2019;54:S173.

65. **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. Distinct blood biomarkers associate with visual evidence of interstitial lung abnormality on HRCT in both SPIROMICS and COPDGene. *Am J Respir Crit Care Med.* 2019;199:A5840.

66. 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. 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.* 2019;199:A4861.

67. 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. Genome-wide association study of acute exacerbations of COPD identifies novel loci in SPIROMICS. *Am J Respir Crit Care Med.* 2019;199:A4862.

68. 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. 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.* 2019;199:A1580.

69. Radicioni G, Ford A, Ceppe A, Alexis NE, **O'Neal WK**, Boucher RC, Kesimer M, SPIROMICS Investigative Group. Cigarette smoke driven MUC5AC increase and airway obstructions in COPD. *Am J Respir Crit Care Med.* 2019;199:A1127.

70. Subramani DB, Chen G, Okuda K, Haridass P, Wang B, Markovetz MR, Radicioni G, Christine LJ, Morrison CB, Fontana NC, Garbarine I, Winkler SS, Livraghi-Butrico A, Hill DB, Button B, **O'Neal W**, Kesimer M, Boucher R, Ehre C. CRISPR/CAS9 based cell lines (A549 and CALU-3 cells at single cell level) for human MUC5ACM and MUC5BM and its role in lung diseases. *Pediatr Pulmonol*. 2019;54:S327.

71. 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. 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.* 2019;199:A5369.

72. 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. Association between acute respiratory disease events and the MUC5B promoter polymorphism in smokers. *Am J Respir Crit Care Med.* 2018;197:A2410.

73. 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. Bronchodilator responsiveness in COPD: results from SPIROMICS. *Am J Respir Crit Care Med.* 2018;197:A6395.

74. 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. Airway epithelial microRNAs associated with small airway dysfunction and Interleukin-17-driven inflammation in chronic obstructive pulmonary disease. *Am J Respir Crit Care Med.* 2018;197:A7441.

75. 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. Mucin-based metrics to quantitate and identify mucus component of peripheral airways obstruction in the SPIROMICS cohort. *Am J Respir Crit Care Med.* 2018;197:A1209.

76. 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. 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.* 2018;197:A7442.

77. 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. 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.* 2018;197:A6387.

78. 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.

Increased endothelial cell damage and decreased endothelial cell repair are implicated in emphysema pathogenesis: SPIROMICS Immunophenotyping Sub-Study. *Am J Respir Crit Care Med*. 2018;197:A7439.

79. 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. 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*. 2018;197:A3544.

80. 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. Imaging-based cluster analysis using novel metrics in COPD for Sub-Phenotyping of Current Smokers: SPIROMICS. *Am J Respir Crit Care Med*. 2018;197:A1033.

81. 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. 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*. 2018;197:A2283.

82. 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. Higher levels of growth differentiation factor 15 (GDF-15) are associated with lower walking distance and exercice capacity in COPD: an analysis of the SPIROMICS cohort. *Am J Respir Crit Care Med*. 2018;197:A3124.

83. 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. Genomic and transcriptomic analysis of pulmonary function and emphysema on CT scan. *Am J Respir Crit Care Med*. 2018;197:A4187.

84. 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. Blood biomarker algorithms as an aide to describe exacerbations activity in symptomatic smokers and early staged COPD patients. *Am J Respir Crit Care Med*. 2018;197:A2747.

85. 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. Regional expression of secretory mucins MUC5AC and MUC5B in normal human airways. *Am J Respir Crit Care Med*. 2018;197:A7636.

86. 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. Comprehensive gene resequencing of SERPINA1 in SPIROMICS reveals novel rare loci for alpha 1-antitrypsin deficiency and emphysema. *Am J Respir Crit Care Med*. 2018;197:A4184.

87. 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. Allergen sensitization and exposure is associated with exacerbations in COPD. *Am J Respir Crit Care Med*. 2018;197:A2750.

88. Dang H, Polineni D, Pace RG, Stonebraker JR, Li Q, Corvol H, Cutting GR, Drumm ML, Strug LJ, **O'Neal W**, Knowles M. Identification of novel CF modifier genes and pathways by expression imputation from GWAS. *Pediatr Pulmonol.* 2017;52:S263.

89. Esther CR, Polineni D, Mahon A, Isaacman S, Bonfield TL, Knowles M, **O'Neal W**. A novel therapeutic targeting the methionine salvage pathway reduces airway inflammation. *Pediatr Pulmonol.* 2017;52:S328-S329.

90. 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. Active smoking is associated with decreased Cd4+T cells in bronchoalveolar lavage: Spiromics Immunophenotyping Sub-Study. *Am J Respir Crit Care Med.* 2017;195:A2629.

91. Okuda K, Chen G, Wolf M, Burns K, Chua M, Livraghi-Butrico A, Ehre C, Doerschuk C, Randell S, **O'Neal W**, Boucher RC. Localization of secretory mucins MUC5AC and MUC5B in normal human airways. *Pediatr Pulmonol.* 2017;52:S238-S239.

92. Okuda K, Chen G, Wolf M, Burns K, Chua M, Livraghi-Butrico A, Ehre C, Doerschuk C, Randell S, **O'Neal W**, Boucher RC. Localization of secretory mucins MUC5AC and MUC5B in normal human airways. *Pediatr Pulmonol.* 2017;52:S238-S239.

93. 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. Nasal transcriptomic studies combined with genomic analyses identify gene networks associated with cystic fibrosis lung disease severity. *Pediatr Pulmonol.* 2017;52:S263-S264.

94. 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**. EXPLORING the biologic basis of the chr11p13 CF lung disease modifying locus using CF airway epithelial cells. *Pediatr Pulmonol.* 2017;52:S265.

95. 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. Adhesive, cohesive and viscoelastic properties of MUC5AC play a role in CF pathogenesis. *Pediatr Pulmonol.* 2017;52:S279.

96. Wells JM, Parker M, Osteri RA, Bowler RP, Dransfield MT, Cho MH, Woodruff PG, Kim V, Curtis JL, Martinez FJ, Paine R, Barri RG, Hani MK, **O'Neal WK**, Castaldi PJ, Gaggar A. Elevated matrix metalloprotease 9 in moderate to severe COPD: results from Spiromics and Eclipse. *Am J Respir Crit Care Med.* 2017;195:A2732.

97. 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. How 'stable' is stable COPD? Daily symptom variability of subjects enrolled in the SPIROMICS Exacerbation Sub-Study. *Am J Respir Crit Care Med.* 2016;193:A3527.

98. 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. 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*. 2016;193:A3515.

99. 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. Biomarker characterization of a debilitated phenotype in Spiromics. *Am J Respir Crit Care Med*. 2016;193:A1426.

100. 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. Identification of a debilitated phenotype in Spiromics. *Am J Respir Crit Care Med.* 2016;193:A3519.

101. 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. 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*. 2016;193:A1432.

102. Chen G, **O'Neal WK**, Deng Y, Yu D, Jones L, Ribeiro C, Randell SH, Boucher RC. Il1r1 regulates Muc5b production and inflammatory responses in the airway epithelium. *Am J Respir Crit Care Med*. 2016;193:A5559.

103. Chen G, Volmer A, Wilkinson K, Terrell K, **O'Neal WK**, Livraghi-Butrico A, Boucher RC. 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*. 2016;193:A5558.

104. Ford AA, Radicioni G, Cao R, Ceppe A, Doerschuk CM, **O'Neal WK**, Anderson WH, Boucher RC, Kesimer M. Mucin hypersecretion associated with chronic bronchitis and not emphysema in sputum from COPD patients from the SPIROMICS Cohort. *Am J Respir Crit Care Med*. 2016;193:A3518.

105. 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. 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.* 2016;193:A6612.

106. Kelada S, Livraghi-Butrico A, McFadden K, Thomas J, **O'Neal WK**, Boucher RC. Quantitative genetic analysis of MUC5AC and MUC5B in a mouse model of asthma. *Am J Respir Crit Care Med*. 2016;193:A2916.

107. 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. 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*. 2016;193:A6345.

108. 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. Scnn1b-TG mice: a unique model to test airway mucus-mobilizing therapies in vivo. *Pediatr Pulmonol.* 2016;51:284-5.

109. 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. 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*. 2016;193:A3510.

110. 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. Rare *SERPINA1* variants in the multi-ethnic Spiromics population. *Am J Respir Crit Care Med*. 2016;193:A4433.

111. 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. 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*. 2016;193:A6458.

112. 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. Age of onset of chronic P. aeruginosa pulmonary infection is associated with differential gene expression in CF nasal epithelia in vivo. *Pediatr Pulmonol.* 2016;51 Suppl 45:244.

113. 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. Association between blood biomarkers, CT phenotype and COPD stage in SPIROMICS. *Am J Respir Crit Care Med*. 2016;193:A4066.

114. Tew W, Chung C, Manser P, Holweg C, Christenson SA, Rosenberger C, **O'Neal WK**, Woodruff PG, Neighbors M. 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*. 2016;193:A7873.

115. 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, Kleerup EC, Martinez FJ, Paine R, Peters SP, Woodruff PG, Rennard SI, Couper D, **O'Neal WK**. 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*. 2015;191:A2935.

116. 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. 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*. 2015;191:A2930.

117. Carlson TD, Kinney GL, Woodruff PG, Hansel NN, Han MK, **O'Neal WK**, Tashkin DP, Bowler RP. The significance and clinical impact of marijuana use in the SPIROMICS Cohort. *Am J Respir Crit Care Med*. 2015;191:A4487.

118. Dang H, Gallins PJ, Rhonda PG, Stonebraker JR, Fossum SL, Leir S, Harris A, **O'Neal WK**, Knowles MR. Re-sequencing GWAS region on chromosome 11 identified novel candidate genetic variants associated with modification of CF lung disease. *Pediatr Pulmonol*. 2015;50 Suppl 41:250.

119. Fossum SL, Mutolo MJ, Yang R, Dang H, **O'Neal WK**, Knowles MR, Leir S, Harris A. Ets homologous factor regulates pathways controlling response to injury in the airway epithelial cells. *Pediatr Pulmonol*. 2015;50 Suppl 41:253.

120. Gupta R, Abdelwahab S, Jones L, Dang H, **O'Neal WK**, Kesimer M. Intercellular communication between the airway epithelial cells is mediated by exosomes like vesicles. *Am J Respir Crit Care Med*. 2015;191:A3660.

121. 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, Kleerup EC, Martinez FJ, Meyers DA, **O'Neal WK**, Peters SP, Rennard SI, Woodruff PG, Bowler RP. Blood biomarker quantitative trail loci in chronic obstructive pulmonary disease. *Am J Respir Crit Care Med*. 2015;191:A3627.

122. 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, Kleerup EC, Kanner R, Hansel NN, Jacobson S, Martinez FJ, Bowler RP. Blood biomarkers of exacerbations in the Spiromics and COPDgene Cohorts. *Am J Respir Crit Care Med*. 2015;191:A5118.

123. Nguyen T, Chung F, **O'Neal WK**, Jones L, Gentzsch M, Boucher R, Kreda SM. The effect of CFTR activity, bicarbonate, and pH on airway mucin secretion. *Pediatr Pulmonol*. 2015;50 Suppl 41:214-5.

124. Polineni D, Esther CR, Pace RG, Stonebraker JR, Jones LC, Guo X, Boyle MP, Chmiel J, Durie PR, **O'Neal WK**, Knowles MR. In vivo human nasal lavage metabolomics suggest biomarkers of lung disease severity in cystic fibrosis. *Pediatr Pulmonol*. 2015;50 Suppl 41:277.

125. Sesma JI, Livraghi-Butrico A, Wilkinson KJ, Weitzer C, Saini Y, Harden TK, **O'Neal WK**, Lazarowski ER. Blockage of the P2y14-receptor inhibits neutrophil infiltration in chronic lung diseases. *Am J Respir Crit Care Med*. 2015;191:A6144.

126. 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. Patterns of biomarker expression in common comorbidities of COPD in the Spiromics population. *Am J Respir Crit Care Med*. 2014;189:A5877.

127. Chen G, Martino ME, **O'Neal WK**, Boucher RC, Ribeiro CM. Role of the UPR in airway epithelial mucin production relevant to cystic fibrosis. *Pediatr Pulmonol*. 2014;49 Suppl 38:255-6.

128. Duncan EA, Burns EN, Dang H, Jania CM, Tilley SL, Boucher RC, **O'Neal WK**, Doerschuk CM. 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*. 2014;189:A4289.

129. 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, Kleerup E, Kanner RE, Barr G, Bleecker ER, Martinez FJ, Woodruff PG, Hoffman EA. Blood biomarkers and CT phenotypes in the Subpopulations and Intermediate Outcome Measures in COPD Study (Spiromics). *Am J Respir Crit Care Med*. 2014;189:A5915.

130. Nguyen T, Jones L, **O'Neal WK**, Boucher RC, Kreda SM. Airway mucin secretion is affected only indirectly by CFTR activity. *Pediatr Pulmonol*. 2014;49 Suppl 38:265.

131. **O'Neal WK**, Anderson WH, Bowler R, Peters SP, Couper D, Rennard SI. Persistent systemic inflammation in the Spiromics COPD population. *Am J Respir Crit Care Med*. 2014;189:A5918.

132. **O'Neal WK**, Wolf W, Jones LC, Gallins PJ, Pace RG, Stonebraker JR, Dang H, Wright F, Knowles MR. 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*. 2014;189:A6717.

133. Pirozzi CS, Scholand MB, Han MK, Cui G, Carretta E, **O'Neal WK**, Barr RG, Paine R, Kanner R. Low and high body mass index are associated with specific COPD phenotypes. *Am J Respir Crit Care Med*. 2014;189:A5936.

134. 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. In vivo human nasal epithelia inform transcriptomic studies in cystic fibrosis (CF) complementary to genomic variation studies. *Pediatr Pulmonol*. 2014;49 Suppl 38:272.

135. 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. 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*. 2014;189:A5519.

136. Terryah S, Wilkinson K, **O'Neal WK**, Taylor J, Tarran R. A SPLUNC1-derived peptide reduces lung disease in SCNN1B mice. *Pediatr Pulmonol*. 2014;49 Suppl 38:285-6.

137. Bove PF, Cheluvaraju C, Dang H, Jones LC, Liu X, **O'Neal WK**, Randell SH, Schlegel R, Boucher RC. 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*. 2013;187:A3399.

138. Doerschuk CM, Carretta E, **O'Neal WK**, Anderson WH, Couper D, Curtis JL, Woodruff PG, Rennard SI, Barr G. Circulating leukocytes, erythrocytes, and platelets in SPIROMICS. *Am J Respir Crit Care Med*. 2013;187:A1513.

139. Duncan EA, Burns E, Prabhu S, Livraghi-Butrico A, Hua X, Dang H, Tilley SL, Boucher RC, **O'Neal WK**, Doerschuk CM. The role of airway surface dehydration and tobacco smoke in the pathogenesis of chronic obstructive pulmonary disease. *Am J Respir Crit Care Med.* 2013;187:A4086.

140. Fletcher A, Livraghi-Butrico A, **O'Neal WK**, Grubb BR, Boucher RC, Evans C. Required roles for Muc5B, but not Muc5ac in mucociliary clearance and anti-bacterial defenses in the lungs. *Pediatr Pulmonol.* 2013;48 Suppl 36:451.

141. Kreda SM, Moussa L, Nguyen T, Jones LC, Wilkinson KJ, Kelly EJ, **O'Neal WK**, Lazarowski ER, Boucher RC. Mucin secretion inhibition in vivo reduces air way mucus obstruction, infection, and Inflammation. *Pediatr Pulmonol.* 2013;48 Suppl 36:238.

142. **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. Differential gene expression identifies CF genetic modifiers associated with severity of lung disease. *Pediatr Pulmonol.* 2013;48 Suppl 36:264.

143. 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. Transforming growth factor beta 1 (TGFB1) expression in vivo is correlated with interleukin 8 (IL8) expression in CF airway (nasal) epithelia. *Pediatr Pulmonol.* 2013;48 Suppl 36:267.

144. Button B, Cai L, **O'Neal WK**, Ehre C, Rubinstein M, Boucher RC. The role of cell surface-attached mucins in airway health. *Pediatr Pulmonol*. 2012;47 Suppl 35:252.

145. Dang H, Guo X, Polineni D, Jones LC, Pace RG, Knowles MR, **O'Neal WK**. MUC5AC reference genomic and MRNA structures from high-throughput sequencing data. *Pediatr Pulmonol*. 2012;47 Suppl 35:282.

146. Kreda SM, Jones L, Moussa L, Fulcher L, Yunxiang Z, Hudson E, **O'Neal WK**, Randell SH, Lazarowski ER, Boucher RC. VAMP8 is the vesicle SNARE for mucin secretion in airway goblet cells. American Thoracic Society, San Francisco CA. *Am J Respir Crit Care Med.* 2012;185:A6299.

147. Kreda SM, Moussa L, Jones L, Clapp P, **O'Neal WK**, Lazarowski ER, Boucher RC. Rock signaling is an important mediator of airway goblet cell mucin secretion. *Pediatr Pulmonol*. 2012;47 Suppl 35:256.

148. Livraghi-Butrico A, Cao R, Kelly EJ, Evans CM, Boucher RC, **O'Neal WK**, Kesimer M. Airway mucin interactome composition in response to defective mucus clearance and genetic deletion of secreted mucins. *Pediatr, Pulmonol.* 2012;47 Suppl 35:253.

149. Livraghi-Butrico A, Duncan EA, Saini Y, Dang H, Doerschuk CM, **O'Neal WK,** Boucher RC. 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*. 2012;9(2):82-3.

150. Saini Y, Dang H, Livraghi-Butrico A, **O'Neal WK,** Boucher RC. Macrophage activation patterns and development of airway inflammation in Scnn1b-transgenic mice. *Pediatr Pulmonol.* 2012;47 Suppl 35:229.

151. Guo X, Dang H, Jones LC, Dang AT, **O'Neal WK**, Knowles MR. Relating respiratory mucin genes to CF lung disease severity: obtaining more accurate genetic maps and identification of functional variants. *Pediatr Pulmonol*. 2011;46 Suppl 34:270.

152. Kreda SM, Jones L, Moussa, L, Fulcher L, **O'Neal WK**, Lazarowski ER, Boucher RC. VAMP8 is the v-SNARE that regulates airway mucin secretion. The 25th Annual North American Cystic Fibrosis Conference. *Pediatr Pulmonol*. 2011;46 Suppl 34:246.

153. Kreda SM, **O'Neal WK**, Jones L, Moussa, L, Boucher RC, Lazarowski ER. 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, 2011. ***Carl Storm Award***

154. LaVange L, Greenberg J, Carretta E, Mostafa J, Deshmukh R, **O'Neal WK**, Rennard SI. The COPD ontology and its role in the Subpopulations and Intermediate Outcome Measures in COPD Study (SPIROMICS). *Am J Respir Crit Care Med*. 2011;183:A6011.

155. Livraghi-Butrico A, Duncan EA, Kelly EJ, Waxer DL, Gilmore RC, Evans CM, Doerschuk CM, Boucher RC, **O'Neal WK.** Deletion of *Muc5b* in *Scnn1b* transgenic mice reduces airway mucus obstruction but perturbs lung immune homeostasis: a new role for secreted mucins? *Pediatr Pulmonol*. 2011;46 Suppl 34:239.

156. Livraghi-Butrico A, Grubb BR, Kelly EJ, Yang H, Geiser M, Boucher RC, **O'Neal WK.** 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.* 2011;46 Suppl 34:271.

157. Livraghi-Butrico A, Kelly EJ, Klem E, Fulcher NB, Wolfgang MC, Boucher RC, Randell SH, **O'Neal WK.** 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.* 2011;183:A1072.

158. Martino M, Brighton B, Dang H, **O'Nneal WK**, Ribeiro CP. IRE1β regulates transcription of airway mucin genes and genes involved in mucin production and glycosylation. *Pediatr Pulmonol*. 2011;46 Suppl 34:248-9.

159. **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. From genes to function: exploring the mechanism of association between CF lung disease severity and genetic variation at Chr11p13. *Pediatr Pulmonol*. 2011;46 Suppl 34:271.

160. Saini Y, Terrel K, Waxer D, Livraghi-Butrico A, **O'Neal WK**, Boucher RC. The roles of macrophages in determining the initiation, progression and resolution of lung inflammation in a mouse model of chronic bronchitis. *Pediatr. Pulmonol.* 2011;46 Suppl 34:258.

161. Bove PF, Grubb BR, Okada SF, Randell SH, **O'Neal WK**, Boucher RC. Human alveolar type II cells secrete and absorb liquid in response to local luminal environmental signals. *Pediatr Pulmonol.* 2010;45 Suppl 33:264.

162. Bove PF, Rogers TD, Brighton B, Ribiero CM, Randell SH, **O'Neal WK**, Grubb BR, Boucher RC. Ion transport and alveolar surface liquid regulation in human alveolar type II cells. *Am J Respir Crit Care Med*. 2010;181:A6460.

163. Gilmore RC, Watson M, Jones L, **O'Neal WK**, Tarran R. A novel, high-throughput fret assay to monitor NF kappa B activation. *Pediatr Pulmonol.* 2010;45 Suppl 33:295.

164. Kinev A, Clapp P, Tatreau JR, Jones L, Gilmore R, **O'Neal WK**, Randell SH. Differential involvement of NF-kappa B and C/EBP beta in acute versus chronic responses of human airway epithelium to P. aeruginosa. *Pediatr Pulmonol.* 2010;45 Suppl 33:282.

165. Kreda SM, **O'Neal WK**, Jones L, Boucher RC, Lazarowski ER. VAMP-8 regulates mucin secretion in airway goblet cells. Poster. The 24th Annual North American Cystic Fibrosis Conference, Baltimore MD. *Pediatr Pulmonol*. 2010;45 Suppl 33:248.

166. Kreda SM, Vidal-Seminario L, van Heusden CA, **O'Neal WK**, Jones L, Boucher RC, Lazarowski ER 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, 2010.

167. Livraghi-Butrico A, Kelly EJ, Evans CE, Boucher RC, **O'Neal WK**. Deletion of Muc16, Muc5ac, Muc5b in *Scnn1b* transgenic mice: implications for CF lung disease pathogenesis and therapy. *Pediatr Pulmonol*. 2010;45 Suppl 33:258.

168. Livraghi-Butrico A, Kelly EJ, Klem E, Fulcher NB, Wolfgang MC, Boucher RC, Randell SH, **O'Neal WK**. Increased susceptibility to bacterial infection/colonization in Scnn1b transgenic mice. *Pediatr Pulmonol.* 2010;45 Suppl 33:295.

169. Livraghi-Butrico A, Kelly EJ, Boucher RC, **O'Neal WK**, Randell SH. 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.* 2010;45 Suppl 33:281.

170. Martino MEB, Jones L, Brighton B, **O'Neal WK**, Ribeiro CMP. The ER stress transducer IRE1β is a key regulator of airway mucin production. *Pediatr Pulmonol.* 2010;45 Suppl 33:256.

171. Watson M, Gilmore R, **O'Neal WK**, Gentzsch M, Tarran R. Adenosine 2B receptor signaling with CFTR is regulated by PDZ binding motifs on the C-terminus. *Pediatr Pulmonol.* 2010;45 Suppl 33:226.

172. Cholon DM, **O'Neal WK**, Randell SH, Riordan JR, Gentzsch M. Apical recyling in primary human airway cells requires N-linked glycans and correct folding of CFTR. *Pediatr Pulmonol*. 2009;44 Suppl 32:219.
173. Kreda SM, Vidal-Seminario L, van Heusden CA, **O'Neal WK**, Jones L, Boucher RC, Lazarowski ER. Thrombin and neutrophil elastase promote coordinated ATP and mucin release from airway epithelial cells. *Pediatr Pulmonol*. 2009;44 Suppl 32:248-9.
174. Livraghi A, Hudson EJ, Wilkinson KJ, Boucher RC, Doerschuk CM, **O'Neal WK**. Identification of congenic *Scnn1b*-Tg lines markedly susceptible to pulmonary hemorrhage and right ventricular hypertrophy. *Pediatr Pulmonol.* 2009;44 Suppl 32:279.
175. Livraghi A, Klem ER, Hudson EJ, Wilkinson KJ, Wolfgang MC, **O'Neal WK**, Boucher RC, Randell SH. Role of MyD88 and environmental bacteria in normal and pathologic airway development. *Am J Respir Crit Care Med*. 2009;179:A2838.
176. Livraghi A, Wilkinson KJ, Hudson EJ, Randell SH, Boucher RC, **O'Neal WK**. Deletion of membrane-tethered mucins 1 and 4 does not alter the phenotype of *Scnn1b* transgenic mice: implications for airway mucin function. *Pediatr Pulmonol.* 2009;44 Suppl 32:249.
177. Martino MB, Ehre C, **O'Neal WK**, Ribeiro CP. Role of the ER stress transducer IRE1 beta in airway inflammation-induced mucin production. *Pediatr Pulmonol*. 2009;44 Suppl 32:259.
178. Bove PF, Trout L, Brighton B, **O'Neal WK**, Boucher RC. Coupling of alveolar to airway surfaces: studies using a transgenic CC10/EGFP secreted protein in alveolar epithelium. *Pediatr Pulmonol.* 2008;43 Suppl 31:230.
179. Braun M, **O'Neal WK,** Xie M, Hurd H, Ribeiro CP. Selective inhibition of EIF2 alpha dephosphorylation reveals a key role for ER stress in airway inflammation. *Pediatr Pulmonol.* 2008;43 Suppl 31:259.
180. Cholon DM, **O'Neal WK**, Riordan JR, Gentzsch M. Modulation of apical stability of rescued ΔF508 CFTR in primary human airway epithelial cells. *Pediatr Pulmonol.* 2008;43 Suppl 31:203.
181. Ehre C, **O'Neal WK,** Boucher RC. A mouse model to study the mechanisms and treatment of mucus adhesion in dehydrated airways. *Pediatr Pulmonol.* 2008;43 Suppl 31:247.
182. Livraghi A, Grubb BR, **O'Neal WK**, Mall M, Boucher RC, Randell SH. Signaling pathways regulating the airway phenotype of mice exhibiting airway surface dehydration. Experimental Biology Meeting 2008, San Diego, CA. *FASEB J.* 2008;22 Suppl 1:328-7.
183. Livraghi A, Klem ER, Hudson, EJ, Wilkinson KJ, Wolfgang MC, **O'Neal WK**, Boucher RC, Randell SH. Genetic deletion of MyD88-mediated signaling in βENaC-overexpressing mice reduces airway neutrophilia but promotes spontaneous, mucus-associated bacterial infection. *Pediatr. Pulmonol.* 2008;43 Suppl 31:262.
184. Livraghi A, Rogers TD, Brighton BW, Hudson EJ, Wilkinson KJ, Boucher RC, **O'Neal WK**, Grubb BR. Increased airway epithelial necrosis and massive airway neutrophilia in double CF/βENaC mice highlights role for CFTR-mediated Cl^- secretioin in neonatal airways. *Pediatr. Pulmonol.* 2008;43 Suppl 31:247.
185. Murray SA, Livraghi A, Sharp JE, Grubb BR, Hudson EJ, Wilkinson KJ, Jones LC, Boucher RC, **O'Neal WK.** Analysis of genetic modifiers of phenotypic severity in the βENaC mouse model of cystic fibrosis. *Pediatr. Pulmonol.* 2008;43 Suppl 31:270.
186. **O'Neal WK**, Hurd H, Wu Y, Braun M, Jones LC, Brighton B, Boucher RC, Ribeiro CM. Azithromycin alters the gene expression and inflammatory responses of human airway epithelia *Pediatr. Pulmonol.* 2008;43 Suppl 31:243.
187. Sesma JI, Esther CR, Kreda SM, Jones LC, **O'Neal WK**, Nicholas R, Lazarowski E. Cellular release of UDP-sugars from the secretory pathway. *Pediatr. Pulmonol.* 2008;43 Suppl 31:245.
188. Braun M, **O'Neal WK**, Ribeiro CP. Expansion of epithelial ER calcium stores during airway infection/inflammation is mediated by Upr-dependent Xbp-1. *Pediatr Pulmonol*. 2007;42 Suppl 30:259-60.
189. Braun M, Tatreau J, Fulcher N, Wolfgang MC, Randell SH, **O'Neal WK**, Ribeiro CP. Pseudomonas aeruginosa activates the unfolded protein response in airway epithelia: relevance for airway inflammation. *Pediatr Pulmonol*. 2007;42 Suppl 30:260.
190. Gentzsch M, **O'Neal WK**, Randell SH, Riordan JR. Endocytic trafficking of wild-type and Δf508 CFTR in highly differentiated primary cultures of human airway epithelia. *Pediatr Pulmonol*. 2007;42 Suppl 30:217.
191. Grubb BR, Livraghi A, Rogers TD, Wilkinson KJ, Hudson EJ, Boucher RC, **O'Neal WK**. Enhancing Na+ absorption in the airways of the CF mouse results in increased mortality. *Pediatr Pulmonol*. 2007;42 Suppl 30:280-1.

192. 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. Assessing mucin gene polymorphisms to determine their role as modifiers of different CF phenotypes. *Pediatr Pulmonol*. 2007;42 Suppl 30:265-6.

193. 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. Novel mediators of persistent neutrophilic inflammation in cystic fibrosis. *Pediatric Pulmonol*. 2007;42 Suppl 30:229-30.

194. Livraghi A, Grubb BR, Hudson EJ, Wilkinson KJ, Mall M, **O'Neal WK,** Boucher RC, Randell SH. TNFα is not a key mediator of airway inflammation and remodeling in βENaC-transgenic mice. *Pediatr Pulmonol*. 2007;42 Suppl 30:257-8.

195. Livraghi A, Grubb BR, Threadgill DW, Mall MA, Hudson EJ, Boucher RC, **O'Neal WK.** Assessment of genetic modifiers for phenotypic severity of *Scnn1b* transgenic mice. *Pediatr Pulmonol*. 2007;42 Suppl 30:266.

196. Livraghi A, Mall M, Rogers TD, Hudson EJ, **O'Neal WK**, Boucher RC, Grubb BR. Generation and phenotypic characterization of transgenic mice over-expressing multiple subunits of EnaC in the airways. *Pediatr Pulmonol.* 2007;42 Suppl 30:239.

197. Livraghi A, Rogers TD, Wilkinson KJ, Hudson EJ, Boucher RC, **O'Neal WK.** Enhancing Na+ absorption in the airways of the CF mouse results in increased mortality. *Pediatric Pulmonol.* 2007;42 Suppl 30:280-1.

198. Mall M, Harkema JR, Trojanek JB, Treis D, Schubert S, Zhou Z, Tilley SL, Livraghi A, **O'Neal WK**, Boucher RC. Initial pulmonary lesions and spontaneous course of lung disease caused bv airway surface liquid depletion in βENaC overexpressing mice. *Pediatr Pulmonol*. 2007;42 Suppl 30:279-80.

199. **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. Deficiency of transmembrane mucin MUC4 reveals a potential role for MUC4 in determination of ciliary beat frequency. *Pediatr Pulmonol*. 2007;42 Suppl 30:240.

200. Rowe SM, Solomon GM, Livraghi A, **O'Neal WK**, Gaggar A, Blalock JE, Clancy JP, Sorscher EJ, Abraham EA, Liu G. Evidence for pathogenic role of HMGB1 in cystic fibrosis. American Thoracic Society International Conference, San Francisco, CA. *Am J Respir Crit Care Med.* 2007;175:A448.

201. Livraghi A, Hudson EJ, DeMaria GC, Sheehan JK, **O'Neal WK**, Boucher RC, Randell SH. Absence of IL-4Rα in Scnn1b-transgenic mice enhances neonatal survival but does not mitigate adult lung pathology. *Pediatr Pulmonol*. 2006;41 Suppl 29:246-7.

202. Livraghi A, **O'Neal WK**, Mall M, Boucher RC, Grubb BR. Enhanced Na^+ absorption in the double transgenic ENaC mouse Scnn1b/c correlates with severity of lung pathology. *Pediatr Pulmonol.* 2006;41 Suppl 29:284.

203. Livraghi A, **O'Neal W,** Mall M, Boucher RC. Airway inflammation in *Scnn1b* transgenic mice. *Pediatr Pulmonol.* 2005;Suppl 28:261.

204. Holda JR, Pickles RJ, Burns KA, Gendler SJ, Boucher RC, **O'Neal WK.** Muc1 is a significant component of mouse airway lumenal glycocalyx. North American Cystic Fibrosis Conference, 2001.

205. Mall M, Boucher RC, **O'Neal WK.** 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, 2001.

206. Randell SH, **O'Neal WK**, Martsen EO, Noone PG, Zhou Z, Plonk MK, Wu Q, Knowles MR, Boucher RC, Gabriel SE. mRNA expression profiling of human airway epithelium. North American Cystic Fibrosis Conference, 2001.

207. Zhang YJ, **O'Neal WK**, Blackburn K, Moyer M, Moseley R, Boucher R, Ostrowski L. Identification and characterization of an axonemal dynein heavy chain reduced in cilia from a patient with primary ciliary dyskinesia. American Thoracic Society, 2001.

208. Lazarowski E, Grubb BR, **O'Neal WK**, Ribeiro CP, Burch N, Boucher RC. Regulation of gastrointestinal ion secretory responses by uridine nucleotide receptors. North American Cystic Fibrosis Conference, 2000.

209. Morse KM, **O'Neal WK**, Patel M, Davis CW, Olsen JC. An ecdysone-inducible expression system for use with retroviral vectors. *Mol Ther.* 2000;1:S186-7.

210. **O'Neal WK**, Wang Y, Taylor K, Beaudet AL. Advantages of a-fetoprotein (AFP) as a reporter gene for expression in lung and liver in animals and humans. *Pediatr Pulmonol.* 1997;Suppl 14:187.

211. Lee B, Timme TL, Shine HD, **O'Neal WK**, Morral N, Zhou H, et.al. 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, 1996.
212. Morral N, **O'Neal WK**, Zhou H, Langston C, Beaudet AL. Host responses to reporter genes complicate the interpretation of studies using adenoviral vectors. *Pediatr Pulmonol*. 1996;Suppl 13:168.
213. **O'Neal WK**, Beaudet AL. Adenovirus infection in MDCK polarized epithelial cells. Ninth Annual North American Cystic Fibrosis Conference, Dallas, TX, October 12-15, 1995.
214. Zhou H, Morral N, **O'Neal, WK**, Beaudet AL. Development of an adenoviral vector with deletions of E1 and E2a genes for CF gene therapy. *Pediatr Pulmonol.* 1995;Suppl 12:164.
215. **O'Neal WK**, Ecay T, Katkin J, Dubinsky W. Adenovirus-mediated expression of CFTR in MDCK cells. *Pediat. Pulmonol*. 1994;Suppl 10:164.
216. **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. Characterization of mice with mutations in exons 2 and 3 or the cystic fibrosis gene. *Pediatr Pulmonol.* 1993;Suppl 9:138.
217. Beaudet AL, Wilson RW, Sligh JE, Ballantyne CM, Lemna **(O'Neal) WK**, Patejunas G, Hasty P, Bradley A, O'Brien WE. Applications of murine gene targeting to human genetics. *J Cell Biochem.* 1992;Suppl 16F:6.
218. Lemna **(O'Neal) WK**, Hasty EP, Beaudet AL, Bradley A. Toward a mouse model for cystic fibrosis. *Pediatr Pulmonol.* 1991;Suppl 6:50.
219. Beaudet AL, Feldman GL, Kobayashi K, Lemna **(O'Neal) WK**, Fernbach SD, O'Brien WE. Strategies for mutation analysis for cystic fibrosis. Fifth International Congress of Inborn Errors of Metabolism. Asilomar, June 1-5, 1990.

Published Reviews, peer reviewed *(from newest to oldest):*

1. **O'Neal WK**, Knowles MR. Cystic Fibrosis Disease Modifiers: Complex Genetics Defines the Phenotypic Diversity in a Monogenic Disease. *Annu Rev Genomics Hum Genet*. 2018 Aug 31;19:201-222. doi: 10.1146/annurev-genom-083117-021329. Epub 2018 Apr 25. PMID: 29709203.
2. Ribeiro CM, **O'Neal WK**. Endoplasmic reticulum stress in chronic obstructive lung diseases. *Curr Mol Med*. 2012 Aug;12(7):872-82. doi: 10.2174/156652412801318791. PMID: 22697344.
3. Randell SH, Fulcher ML, **O'Neal W**, Olsen JC. Primary epithelial cell models for cystic fibrosis research. *Methods Mol Biol*. 2011;742:285-310. doi: 10.1007/978-1-61779-120-8_18. PMID: 21547740.
4. **O'Neal WK**, Beaudet AL. Somatic gene therapy for cystic fibrosis. *Hum Mol Genet*. 1994;3 Spec No:1497-502. doi: 10.1093/hmg/3.suppl_1.1497. PMID: 7849744.
5. Beaudet AL, Feldman GL, Kobayashi K, **Lemna (O'Neal) WK**, Fernbach SD, Knowles MR, Boucher RC, O'Brien WE. Mutation analysis for cystic fibrosis in a North American population. *Adv Exp Med Biol*. 1991;290:53-4. doi: 10.1007/978-1-4684-5934-0_7. PMID: 1950757.

6) Teaching Activities

No formal teaching.

7) Grants (Present and Completed)

ACTIVE

1 U01 HL156655-01A1 (Boucher, Chen, Olivier) 3/5/22-2/28/26
NIH/NHLBI
The molecular and cellular mechanisms of the STAT3 mutation-mediated pulmonary disorder in Autosomal Dominant Hyper IgE Syndrome (AD-HIES)

This application proposes to identify the molecular and cellular mechanisms underlying mutant STAT3 functional defects in innate immunity in airway epithelia and provide therapeutic options for new therapies for the pulmonary manifestations of autosomal dominant hyper IgE syndrome (AD-HIES).

Role: Co-Investigator

5 P30 DK 065988-17 (Boucher) 8/1/20-5/31/25
NIH/NIDDK
UNC Cystic Fibrosis Research and Translation Core Center, Core B: Molecular/Functional Measurement Core

The major goal of the overall P30 is to synergize and accelerate cystic fibrosis research by creating and supporting four research cores, a pilot and feasibility program and an administrative core to coordinate the activities. Cores include 1) Molecular and Functional Measurement Core focused on CFTR biogenesis/function and cystic fibrosis pathophysiology, 2) Cell Models Core focused on generation/provision of relevant airway and GI epithelial cell models, 3) Mucus Biochemistry/Biophysics Core, and 4) Human Translational Studies Core. Core B will provide support for translational cystic fibrosis research by evaluating pre-clinical drug candidates in vitro in cell cultures, ex vivo in organoid models, and in vivo in mouse models.
Role: Core Co-Director

2021-237918 (Hagood/Boucher/Hernandez/Kimple/Randell) 10/1/21-9/30/24
The Chan Zuckerberg Initiative
Mapping the Pediatric Inhalation Interface: Nose, Mouth and Airways

We will integrate respiratory system (RS) cellular transcriptomes and epigenomes, RS fluid secretomes, and RS microbiomes to describe and understand the healthy ecological landscape of the inhaled RS interface throughout childhood.
Role: Co-Investigator

No Number (Moorman, Baric, Heise) 1/1/22-12/31/23
State of North Carolina
Rapidly Emerging Antiviral Drug Development Initiative (READDI)

Funding requested to support READDI, which is designed to serve as a non-profit drug research and development organization with a unique open science component that is focused on the viral families that cause the majority of epidemics and pandemics. The goal is to be prepared in anticipation of future viral pandemics.
Role: Co-Investigator

AWD00002557 (134922-1) (Wenzel) 7/1/20-6/30/24
Univ of Pittsburgh sub on NIH R01 HL153058
Mucin sialylation drives epithelial cell senescence and severe asthma

This application explores the paradigm shifting hypothesis that post-translational modification (sialylation) of a cell surface (tethered) mucin, MUC4, drives terminal differentiation and senescence of airway epithelial cells (AECs) through inhibition of epidermal growth factor receptor (EGFR) family pathways, worsening epithelial wound repair and asthma severity. Our proposed studies will be the first to specifically test tethered mucins and their post-translational N-glycosylation/sialylation for a role in AEC terminal differentiation and senescence.
Role: Co-Investigator

5 U24 HL141762-04 (Couper & O'Neal) 8/15/18-7/31/23 2.4 calendar
NIH/NHLBI $1,074,023 annual direct costs
SPIROMICS GIC Support

This application proposes infrastructure support for the Genomics and Informatics Center (GIC) and assistance to the clinical sites and investigators for the multi-center Sub-Populations and InteRmediate Outcome Measures in COPD Study (SPIROMICS). The GIC will continue to manage SPIROMICSA data

and biospecimens, undertake statistical analyses of study data, collaborate in operational and scientific aspects of the study, and provide study management and regulatory oversight.
Role: MPI

5 U24 HL138998-04 (Ivanova/LaVange) 9/23/17-6/30/23
NIH/NHLBI
Data, Modeling, and Coordination Center for PrecISE Network

The goal of this U24 application is to establish the Data, Modeling, and Coordination Center (DMCC) for the NHLBI's Precision Interventions for Severe and/or Exacerbation-Prone Asthma (PrecISE) Network. The objective of PrecISE is to conduct sequential, adaptive, phase II/proof of concept trials with precision interventions in stratified patient with severe asthma populations.
Role: Co-Investigator

BOUCHE19R0 (Boucher) 7/1/19-6/30/23 0.60 calendar
Cystic Fibrosis Foundation $163,625 (Core E) annual direct costs
Epithelial Function in Cystic Fibrosis, Core E: Molecular Biology and Animal Models Core

The major goal of this core is to provide molecular biology materials, technical expertise and training and genetically modified mice relevant to cystic fibrosis research. These materials and support will further research efforts into CF within UNC and also by outside collaborators.
Role: Core Leader

KNOWLES21XX0 (Knowles) 5/1/21-4/30/23 (NCE)
Cystic Fibrosis Foundation
Whole Genome Sequencing to Define Gene Modifiers in CF

The major goal of this project is to carry out whole genome sequencing on CF patients enrolled in multiple CFF studies from three separate sites.
Role: Co-Investigator

BOUCHE19XX0 (Boucher) 2/1/20-1/31/23
Cystic Fibrosis Foundation
Abrogation of Airway Epithelial Barriers to Transduction, Project 1: Regions/Cell Types as Targets for CFTR Therapy

The UNC Collaborative Research Grant is designed to: 1) identify airway regional and cellular targets for CFTR nucleic acid therapy; 2) develop novel systems for their study; and 3) innovate techniques to overcome barriers. Three integrated projects attack these issues. Project 1 will 1) test findings that the Club cell is the dominant CFTR-expressing cell in small airways; 2) characterize the complex Club cell ion transport/mucin secretory functions; and 3) characterize Club cell nucleic acid transducibility.
Role: Co-Investigator

PICKLE21G0 (Pickles) 2/1/21-1/31/23
Cystic Fibrosis Foundation
Do mucus secretions and airway inflammation protect the CF Lung from SARS2?

Because CF lung disease is so complex, with dehydrated mucus accumulating in the airways due to loss of the CFTR protein with inflammation and infection, we will use models of CF in mice infected with a SARS virus strain specifically adapted to infect mice. Using these models, we will explore three of the major features of CF lung disease: mucus, inflammation, and CFTR-deficiency. Increased mucus secretion might restrict access to target cells by inhaled viruses; the inflamed airway epithelium present in CF lungs might result in resistance of the epithelium to respiratory virus infection; and CFTR deficiency alone could alters cell and tissue homeostasis, which may render the cells more resistant or susceptible to virus infection and

spread. We hypothesize excessive accumulation of dehydrated mucus secretions, pre-existing airway inflammation, and/or loss of CFTR function, as present in CF patients, alters SARS2 responses.
Role: Co-Investigator

22-0377 (O'Neal) 7/1/21-6/30/22 0.12 calendar
NHMRC (Synergy Australia) subcontract $17,062 annual direct costs
Comparison of fresh versus frozen epithelial cells for scRNA-seq analysis

Goal is to test fidelity of the freezing technique vs freshly isolated cell technique to inform the Synergy group as to best practices with respect to how to handle fresh transbrochoscopic brush epithelial and BAL samples for scRNAseq, and the congruence of culture sample scRNAseq data to data generated from deconvoluted bulk seq spatial transcriptomics analyses.
Role: PI

5 P01 HL108808-10 (Peden/Boucher) 7/1/20-6/30/22
NIH/NHLBI
Development of Novel Mycolytic Therapies for Lung Disease, Project 2: PK/PD requirements for mucolytic therapeutic agents in vitro and in vivo

The overarching therapeutic goal for the UNC tPPG renewal is to clear the hyperconcentrated, adherent mucus that promotes airways obstruction, inflammation, and infection. Project 2 has focused on the hypotheses that: (1) hyperconcentrated (dehydrated) mucus produces airway mucus adhesion/plaques that drive the progression of muco-obstructive lung diseases; and, (2) that clearance of these plugs/plaques will be therapeutically useful in patients with the muco-obstructive phenotype. Thus the overarching goal of the 2b Project is to identify the optimal drug properties, balancing rate of mucin thiol reduction, duration of activity, and safety, to develop a new class of muco-clearance assisting agents that focus on mucin MW reduction that can be delivered alone or in combination with hydrating agents.
Role: Co-Investigator

Subcontract on R01 HL137995-01A1 (Bowler) 6/1/21-5/31/22 1.28 calendar
National Jewish Health sub on NIH/NHLBI $20,000 (sub only) annual direct costs
Biomarkers of Lung Disease in African-Americans

The goals of the overall proposal are: (1) identify and replicate proteomic signatures for chronic obstructive pulmonary disease (COPD) disease progression (airflow limitation and emphysema) in African American (AA) and Whites and (2) integrate these proteomic signatures with other existing omics data to identify molecular networks associated with disease progression.
Subcontract PI

10454sc (Couper UNC, Woodruff, UCSF) 9/15/17-5/31/22
UCSF, subcontract from NIH U01 HL137880
SPIROMICS II

The UNC Collaborative Studies Coordinating Center will serve as the Data Coordinating Center (DCC) for the SPIROMICS II project. The DCC will be responsible for developing and maintaining study documents as well as creation/curation of the web-based data entry system.
Role: Co-Investigator

5 R01 HL136961-04 (Boucher) 8/1/17-5/31/22
NIH/NHLBI
Multiscale Biochemical/Biophysical Integration of Pulmonary Mucus Transport

We have selected four key gaps in our understanding of the mucociliary system for investigation. These gaps constitute our four specific aims. Specific Aim 1 How is mucin secretion organized intraregionally in superficial epithelia and proximally with submucosal gland secretion? Specific Aim 2: Are there functional

differences in mucus secreted by superficial epithelia vs glands? Specific Aim 3: How are mucins released from granules onto the airway surface? Specific Aim 4: Why are mucins organized into gigantic (>100MD) higher-order multimers? The hypotheses tested are that gigantic molecules are required to generate mucus gels with biophysical properties commensurate with transport in dilute solutions and that such large molecules impose unique cell biologic packaging constraints on the cell. Importantly, resolution of the questions/hypotheses generated by this analysis of "gaps" in knowledge of the MCC system may lead to paradigm shifts in our understanding of normal MCC and how to approach novel therapies for bronchitic diseases.
Role: Co-Investigator

PENDING

1 P01 HL164320-01 (Boucher, Rubinstein) 7/1/22-6/30/27
NIH/NHLBI
Multi-Scale Investigations of Respiratory Mucus/Mucin Structure and Function in Health and Disease, Project 2: Why are mucins so gigantic and is it safe/effective to sever them therapeutically?

The goal is to search for potential favorable effects of reducing multimer length on the mucus viscosity required for cough clearance and identify off-target effects mediated by reduction of other intra-mucin cysteines that may produce untoward mucin aggregation/stickiness. The deliverables of the project are to: 1) quantitate the relationships between mucin multimer size, space-occupying characteristics (c*), and mucus function in health; and 2) characterize the mucolytic agent therapeutic index with respect to on- vs off-target effects and provide a roadmap for development of novel mucolytic therapeutics for patients in need.
Role: Co-Investigator

3 R01 HL155951-02S1 (Chen) 4/1/22-3/31/23
NIH/NHLBI
Role of alveolar KRT8+ transitional cells in promoting pulmonary fibrosis in response to SARS-CoV-2 infection

Our central hypotheses are that: 1) an abnormal AT2 transitional Krt8+ cell with features of hyperinflammation and reduced cell cycle/reparative capacity drives PF progression; and 2) this cell type is targetable by novel senolytic compounds that may prevent disease progression. We propose to test the hypothesis that persistence of alveolar Krt8+ transitional cells derived from ER stressed airway and/or AT2 progenitors drive late-stage PF and PASC. We will test this hypothesis by the following specific aims. Specific Aim1: Investigations of the cellular origin(s) and fates of the alveolar Krt8+ transitional cells following SARS-CoV-2 infection. Specific Aim2: Determine whether infected or non-infected (bystander) cells give rise to Krt8+ transitional cells in alveoli post SARS-CoV-2 infection.
Role: Co-Investigator

Admin Supp (Chen, Stripp) 4/1/22-3/31/23
Cedars-Sinai Medical Center sub on NHLBI
Chronic lung fibrosis as a sequalae of SARS-CoV-2 infection

The UNC subcontract team will conduct mouse exposure experiments, scRNAseq analyses, histological analyses, viral titers, and Luminex cytokine panels. They will perform 10x Genomcis scRNA analyses of single cells and generate libraries and perform quality control and initial bioinformatics analyses of all scRNA seq data.
Role: Co-Investigator

3 R01 HL150541-03S1 (Livraghi-Butrico) 4/1/22-3/31/23
NIH/NHLBI

Mouse models to identify risks imposed by preexisting pulmonary diseases for development of prolonged COVID-19 Pulmonary sequelae

The major goals of this grant are to identify and characterize models of lung disease that: 1) predict lung disease-specific risks for COVID-19 pulmonary sequelae; and 2) accelerate therapeutics testing.
Role: Co-Investigator

COMPLETED

NH19-OKUDA-1 (Okuda) 6/1/19-5/31/21
Cystic Fibrosis Research, Inc. New Horizons Grant
Regional regulation of CFTR and ionocyte expression in airways

The first aim reflects the source and distribution of CFTR in airways. We will systematically assess the regional distribution of pulmonary ionocytes and other possible cell types for CFTR expression in the proximal-distal axis of the human lung by RNA in situ hybridization, complemented by studies on rabbit and mice airways. The second aim reflects the potential regulation of pulmonary ionocyte expression under environmental stresses experienced by the airway epithelium. We will seek to clarify how the frequency of pulmonary ionocyte is altered under hyperosmotic, hypoxic and shear stress, utilizing primary airway epithelial cell cultures obtained from normal and CF human lungs.
Role: Co-Investigator

KNOWLE18XX0 (Knowles) 6/1/18-4/30/21 (NCE)
Cystic Fibrosis Foundation
Discovery of CF Modifiers using Whole Genome Sequencing UNC

To discover CF modifiers, we will carry out whole genome sequencing (WGS) in 5,200 individuals with CF from the Gene Modifier Study (UNC), the Twin and Sibling Study (JHU), and the EPIC Observational Cohort Study (EPIC).
Role: Co-Investigator

COVID-19 Collaborative Grant (Boucher/O'Neal) 7/1/20-12/31/20 0.3 calendar
State of North Carolina $557,899
Therapeutics II: Preclinical Studies of Novel Therapeutic Agents in Mouse Models for Target/Drug Validation, Pharmacokinetic (PK) Studies, and Efficacy Using Mouse-Adapted SAS-CoV-2 Virus, Project 1

We propose to create with the Baric lab a world-class in vivo mouse facility to test therapies to treat both components of COVID-19 disease: 1) the virus-dominated early damage phase; and 2) the late inflammation/repair phase. The Baric lab will focus on anti-viral approaches. The O'Neal lab component of a mouse therapeutics core focuses on the inflammation/repair components of COVID-19.
Role: Co-PI

5 UH3 HL 123645-05 (Boucher) 9/22/14-6/30/20 (NCE)
NIH/NHLBI
Synthesis of Effective and Safe Mucolytics for Pulmonary Disease

Our goal is to develop a novel mucolytic to be used as a single agent, or in combination of hydrating agents, to treat mucus retention in patients in need thereof. Strategies to optimize a lead compound and generate a clinical candidate are outlined in a four-tier approach in Specific Aim 1 that focus on both increases in safety and efficacy. Processes required to move the clinical lead to an IND are outlined in Specific Aim 2, including all of the IND requiring medicinal chemistry, toxicology, ADME, and PK studies.
Role: Co-Investigator

GRUBB17XX0 (Grubb) 4/1/17-3/31/20 (NCE) 0.60 calendar
Cystic Fibrosis Foundation Therapeutics, Inc. $349,909

Characterizing the CF Rabbit

Our goal is to establish the feasibility of the CF rabbit as a model of spontaneous CF airway disease by extending the life and evaluating the longitudinal development of lung pathology.
Role: Co-PI

5 P30 DK 065988-15 (Boucher) 4/23/15-3/31/20
NIH/NIDDK
UNC Cystic Fibrosis Research and Translation Core Center, Core B: Pre-Clinical Core

The major goal of the overall P30 is to synergize and accelerate cystic fibrosis research by creating and supporting four research cores, a pilot and feasibility program and an administrative core to coordinate the activities. Cores include 1) Pre-Clinical Core focused on CFTR biogenesis/function, 2) Cell Models Core focused on generation/provision of relevant airway and GI epithelial cell models, 3) Mucus Biochemistry/Biophysics Core, and 4) Human Translational Studies Core. Core B will provide support for translational cystic fibrosis research by evaluating pre-clinical drug candidates in vitro in cell cultures, ex vivo in organoid models, and in vivo in mouse models.
Role: Core Co-Leader

1 R56 HL 136909-01A1 (Lazarowski) 9/21/18-8/31/19
NIH/NHLBI
Regulation of Airway Mucus Hydration and Clearance by Released Nucleotides

The objective of this proposal is to investigate the contribution of nucleotide release and metabolism to mucin hydration in healthy and CB-stressed primary cultures of human and murine airway epithelial cells. Based on our preliminary data, we hypothesize that perturbations in ATP release and/or ecto-metabolic pathways govern mucus hydration responses to CB-related insults in airway epithelial cells.
*Effort was 0.6 cm 9/21/18-8/31/19
Role: Co-Investigator

5 R01 HL 125280-04 (Button) 7/3/15-6/30/19
NIH/NHLBI
The Role of Mucus and Pulmonary Surface Intersections in Lung Disease

Our goal is to understand how the mucus and periciliary layers are maintained in health and why they fail in disease. In Aim 1 we will investigate the role of the PCL in airway defense; in Aim 2 we will perform studies to understand how mucus dehydration and neutrophil elastase alter the osmotic and cohesive properties of the mucus layer; and in Aim 3, we will combine the knowledge gained to understand how the mucus and PCL layers interact to maintain cilia- and cough-mediated mucus clearance and why they fail in disease.
Role: Co-Investigator

BOUCHE15R0 (Boucher) 7/1/15-6/30/19 0.60 calendar
Cystic Fibrosis Foundation $172,287 (Core E)
Epithelial Function in Cystic Fibrosis, Core E: Molecular Biology and Mouse Core

The major goal of this core is to provide molecular biology materials, technical expertise and training and genetically modified mice relevant to cystic fibrosis research. These materials and support will further research efforts into CF within UNC and also by outside collaborators.
Role: Core Leader

Research Contract (O'Neal) 12/10/18-5/31/19 0.24 calendar
Spirovation $19,321
Identification of TMEME16a in the Human Airway using RNAscope

The goal of this contract to conduct tests on the detection of RNA within fixed cells/tissue.
Role: PI

KNOWLE17G0 (Knowles) 11/1/17-10/31/18
Cystic Fibrosis Foundation
Exploring miRNAs for Regulating CF Lung Disease Severity

The goal of the project is to provide key information on miRNA expression that can be integrated with other datasets to develop further insight into the link between non-CFTR genetic variation and CF lung disease severity.
Role: Co-Investigator

DANG16I0 (Dang) 11/1/16-10/31/18
Cystic Fibrosis Foundation
Discovering CF Modifier Genes & Pathways by Expression Imputation from GWAS

We propose to impute genetically regulated gene expression from the CF GWAS data of 6,350 patient samples, and test CF disease phenotype association with much improved power to accelerate the discovery of modifier genes, which maybe targeted for intervention.
Role: Co-Investigator

4 P01 HL 108808-05 (Boucher) 6/15/12-8/31/18
NIH/NHLBI
Novel Therapies for Muco-Obstructive Lung Diseases, Project II: Mucus Obstructed Mice for Biomarker and Drug Development

The major goals of this project are (1) to test in vivo the novel biophysical/biochemical formulation of a "two-gel" mucus clearance system, focusing on the durability of the response to inhaled hypertonic saline, (2) utilizing βENaC mouse lines of differing airways Na+ transport/airway surface dehydration, search for in vivo mucus biomarkers denoting the pharmacodynamic activities of drugs and identify novel "downstream" markers of muco-obstructive/dehydration-induced pathogenesis, e.g., inflammatory cell and epithelial cell markers, (3) test the hypothesis that secreted mucins (MUC5AC, MUC5B) or transmembrane tethered mucins (MUC1, 4, 16) are therapeutic targets with favorable risk:benefit ratios by genetic studies of MUC5AC and MUC5B -/-crosses with βENaC mice; and (4) test the hypothesis that bacterial infection of mucus-obstructed COPD airways can be prevented or reversed by inhaled hydration/mucolytic therapies.
Role: Co-Investigator

5 P50 HL 120100-05 (Tarran) 9/19/13-6/30/18
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?

The major goal of the overall P50 is to measure the potential adverse impact of tobacco alternatives ("little cigars" and Hookah) on the lung's innate defense system. Project 3 proposes development of a novel animal model of smoke exposure that more closely mimics the chronic bronchitis phenotype seen in humans with COPD. This model will be used to validate tobacco exposure biomarkers seen in Projects 1 and 2 as well as to determine epigenetic changes following in vivo exposure to alternative tobacco.
Role: Co-Investigator

4 R01 HL 117843-04 (Harris/Knowles/O'Neal) 7/15/13-5/31/18 1.20 calendar
Case Western Reserve Univ (sub on NIH/NHLBI) $137,869 (sub only)
Mining open chromatin to define molecular mechanisms of CF modifier genes

The goals of this project are to define important mechanistic links between genetic variation and severity of CF lung disease and offer opportunity for therapeutic insights. This application focuses specifically on a region of chromosome 11 between the two genes, *EHF* and *APIP*, that shows strong genetic association to cystic fibrosis lung disease severity.

Role: MPI

1 R43 HL 132646-01 (Isaacman/Esther) 4/1/16-12/31/17 (nce)
PHD Biosciences, SBIR sub on NIH
A Targeted Drug for the Treatment of Inflammation in Cystic Fibrosis Lung Disease

The goal of this Phase I SBIR is to examine the potential of an MTAP inhibitor as a therapeutic for CF using a mouse model of CF airways disease. We will test the effects of different single dose concentrations as well as longer term (7-day) treatment on multiple outcome measures including airway and systemic levels of inflammatory cells (neutrophils, macrophages), inflammatory markers (KC, MIP2, LIX), and methionine salvage pathway metabolites (MTA, polyamines). We will also assess the impact of treatment on macrophage activity and lung histology. The primary goal of this proposal is to identify the minimum efficacious dose and determine the pharmacological benefits of a repeat dosing schedule.
Role: Co-Investigator

None Assigned (Couper) 8/1/16-9/30/17
COPD Foundation
SPIROMICS Bridge Funding

The CSCC will serve as the Data Coordinating Center (DCC) for the SPIROMICS Bridge project. The DCC is responsible for maintaining study documents and the data entry system. We will be retrieving data from the data entry system and creating reports, data checks, and analysis datasets. The DCC is also responsible for site training and certification as well as IRB tracking.
Role: Co-Investigator

4 P01 HL 110873-05 (Boucher) 5/15/12-4/30/17 3.70 calendar
NIH/NHLBI $141,351 (Core B)
Pulmonary Surface Liquid Homeostasis, Core B: Molecular Biology Core

The major goal of this core is to provide molecular services to the projects of the PPG: cloning, RNA expression analysis and siRNA development, and transgenic mouse development and mouse genotyping.
Role: Core Leader

5 R01 HL 103940-05 (Kesimer) 7/21/10-4/30/17 (nce)
NIH/NHLBI
The Role of Mucin-Protein Interactions in the Innate Defense of the Lung

The goals of this project are 1) To determine our target panel of proteins that show evidence of specific mucin binding; 2) To determine the domains required for the mucin-protein interactions and 3) To assess the effects of mucin-protein interactions on the surface and bulk rheological properties of the mucus. If the goals of this proposal are achieved, they will increase our knowledge and understanding of the relationship between protein composition and the function of airway secretions. Such knowledge is an essential pre-requisite for informed therapeutic intervention.
Role: Co-Investigator

Research Contract (O'Neal) 11/3/15-9/30/16 0.12 calendar
Spirovation $16,000
Target Validation using CRISPR/Cas9 Technology

The goal of this contract to test target compounds on cell cultures for influence on correction of deltaF508-CFTR.
Role: PI

HHSN268200900020C (Couper) 2/1/09-7/31/16

NIH/NHLBI
SPIROMICS - Genomics and Informatics Center

The major goals of this contract are to establish at UNC-Chapel Hill the Genomics and Informatics Center for the Subpopulations and Intermediate Outcome Measures in COPD study (SPIROMICS) and to serve in this capacity for the course of the study.
Role: Co-Investigator

5 R01 HL 068890-13 (Knowles) 8/1/12-6/30/15
NIH/NHLBI
Gene Modifiers in CF Lung Disease

The major goal of this project is to test the hypothesis that identification of genetic modifiers of CF lung disease will help to clarify disease pathogenesis and suggest therapeutic targets.
Role: Co-Investigator

R026-CR11 (Boucher) 7/1/11-6/30/15 0.6 calendar
Cystic Fibrosis Foundation $162,500 (Core E)
Epithelial Function in Cystic Fibrosis, Core E: Molecular Biology and Mouse Core

The major goal of this core is to provide molecular biology materials, technical expertise and training and genetically modified mice relevant to cystic fibrosis research. These materials and support will further research efforts into CF within UNC and also by outside collaborators.
Role: Core Leader

5 P30 DK 065988-10 (Boucher) 4/27/09-3/31/15* 1.2 calendar
NIH/NIDDK $83,390 (Core C)
Molecular Therapy Core Center, Core C: Molecular Biology and Mouse Core

The major goal of this core is to support the gene transfer community by providing mouse models relevant to cystic fibrosis gene transfer and access to molecular biology equipment and expertise. *4/1/14-3/31/15 is bridge funding.
Role: Core Leader

Early Excellence Grant (Ribeiro) 7/1/10-6/30/13
American Asthma Foundation
IRE1beta-Dependent Airway Mucin Production and ATP Release: A New Pathway in Asthma

The major goal of this project is to test the hypothesis that IRE1β is required for mucin production by airway mucous cells, it stimulates mucin transcription and/or regulates genes involved in mucin production or glycosylation, its overexpression potentiates mucin production, and it regulates airway ATP release by regulating the ATP content in mucin granules.
Role: Co-Investigator

5 R01 HL102371-03 Subcontract (Gaggar) 7/2/10-5/31/13 0.6 calendar
UAB subcontract on NIH grant $23,000 (sub only)
A Novel Proteolytic System of Pulmonary Inflammation

The goal of this project is to use the expertise of UNC-CH investigators with regards to the Scnn1b-transgenic over-expressing mouse model along with the UAB group's expertise on the prolylendopeptidase pathway, which is thought to be a novel regulator in pulmonary neutrophilic inflammation, to study the potential for the praline-glycine-proline (PGP)-containing sequences, found to be chemotactic for neutrophils, as a pathway in pulmonary disease.
Role: Subcontract PI

5 R01 HL 095396-04 (Knowles/Wright) 9/24/08-7/31/12

NIH/NHLBI
Molecular Phenotypes for Cystic Fibrosis Lung Disease

The major goal of this project is to study the role of gene expression variation in CF lung disease and the integrated analysis of SNPs/CNVs and expression data.
Role: Co-Investigator

5 P01 HL 034322-25 (Boucher) 2/1/07-1/31/12 4.80 calendar
NIH/NHLBI $139,833 (sub only)
Pulmonary Epithelia in Health and Disease, Core B: Molecular and Protein Core

The major goal of this core is to provide Molecular Biology and Protein services to the PPG projects including cloning, RNA expression analysis and siRNA development, and transgenic mouse development and mouse genotyping.
Role: Core Leader

HIRSH01I0 (O'Neal) 6/1/10-7/31/11 0.60 calendar
Cystic Fibrosis Foundation $40,000
Feasibility of Na^+ Channel Blocker Therapy for CF

The major goal of this project is to test the hypothesis that the duration of action of Na^+ channel blockers is influenced by their affinity towards Na^+ channels and the rate of absorption by the airway epithelium.
Role: PI

R026-CR07 (Boucher) 7/1/07-6/30/11 0.60 calendar
Cystic Fibrosis Foundation $152,474 (sub only)
Epithelial Function in Cystic Fibrosis, Core E: Molecular Biology and Mouse Core

The major goal of this core is to provide molecular biology materials, technical expertise and training and genetically modified mice relevant to cystic fibrosis research. These materials and support will further research efforts into CF within UNC and also by outside collaborators.
Role: Core Leader

Subcontract on 3 P40 RR 016049-09S1 (Donahue) 7/1/09-6/30/10 1.20 calendar
NIH/NCRR subcontract from Jackson Labs $175,000
Development of the bENaC Model of Cystic Fibrosis for Translational Research

The major goal of this project is to develop and improve the *Scnn1b* mouse, an existing mouse model of cystic fibrosis (CF), for use in translational research and preclinical drug test protocols. The *Scnn1b* mouse represents an ideal animal model to both identify novel modifier genes and pathways of CF lung disease and to demonstrate the effect of genetic background on preclinical outcomes, paving the way for improved protocol design.
Role: Subcontract PI

RANDEL07P0 (Randell) 3/1/07-6/30/09 (NCE)
Cystic Fibrosis Foundation
Dysregulated Airway Physiology in Scnn1b Transgenic Mice

The major goal of this core is to elucidate key mechanisms by which defective mucus clearance results in chronic airway injury by 1) studying the interaction between tethered and secreted mucins to examine mechanisms of mucus adhesion, plaque formation and airway obstruction, 2) determining causes and effects of airway inflammation and goblet cell metaplasia in Scnn1b mice, and 3) examining the pathogenesis of respiratory virus-induced airway disease in Scnn1b mice.
Role: Co-Investigator

R026-CR07 (Boucher) 7/1/07-6/30/09 0.30 calendar

Cystic Fibrosis Foundation $40,000 (sub only)
Epithelial Function in Cystic Fibrosis, Project 2: Transmembrane Mucin Function in Mucociliary Clearance

The major goal of this project is to test the hypothesis that in the dehydrated environment of a CF lung, the secreted mucus sticks to the cells by interacting with mucin molecules, called transmembrane mucins, that are attached to the cell surface, by developing a mouse model deficient for the transmembrane mucin Muc 16.
Role: Project Leader

ONEAL07G0 (O'Neal) 4/1/07-3/31/09 1.80 calendar
Cystic Fibrosis Foundation $90,000
CF Modifiers Defined by Scnn1b Over-expressing Mice

The major goals of this project are to (1) evaluate the strain dependent differences with respect to survival, lung pathology, and inflammatory status by studying these phenotypes in congenic inbred lines of Scnn1b over-expressing mice, (2) document and characterize the short-circuit current between lines and among strains in both tracheal and bronchial tissues, and (3) begin to define the modifying effects between the two lines and strains at a genetic level by establishing an F2 recombinant panel of DNA's for genetic linkage studies and by cloning the transgene insertion site for line 6047, which is hypothesized to be linked to modifying locus.
Role: PI

5 R01 HL 080322-04 (Randell) 4/1/05-3/31/09
NIH/NHLBI
Airway Epithelial Adaptation to Infectious Stimuli

The goal of this project is to test the hypothesis that adaptation is a key determinant of airway inflammation and that underlying mechanisms can be exploited as novel approaches for anti-inflammatory therapy.
Role: Co-Investigator

5 P30 DK 065988-05 (Boucher) 4/1/04-3/31/09 1.20 calendar
NIH/NIDDK $94,621 (sub only)
Molecular Therapy Core Center, Core C: Molecular Core

The major goal of this Core is to provide a mouse with CF-like lung disease (the betaENaC mouse) and toxicogenomics capabilities to the UNC-CH molecular therapeutics community.
Role: Core Leader

5 P50 HL 060280-10 (Boucher) 9/16/03-8/31/08 1.20 calendar
NIH/NHLBI $157,297 (sub only)
SCOR in Airway Biology/Pathogenesis of Cystic Fibrosis, Core B: Molecular Biology and Monoclonal Antibody Core

The major goal of this core is to provide centralized facilities for development of molecular and monoclonal antibody reagents and services as required for SCOR projects.
Role: Core Leader

5 P30 DK 065988-04 (Boucher) 4/1/06-3/31/08 0.60 calendar
NIH/NIDDK $43,314 (sub only)
Molecular Therapy Core Center, Project 6: Interfering RNA for Modulation of ENaC Function

The major goals of this pilot project are to generate small interfering RNA to αENaC to study the role of ENaC in CF well-differentiated human airway cultures and in the development of CF disease, and to generate a murine model to evaluate loss of ENaC function *in vivo*.
Role: Project Leader

R026-CR02 (Boucher) 7/1/02-6/30/07 0.60 calendar
Cystic Fibrosis Foundation $101,537 (sub only)
Epithelial Function in Cystic Fibrosis, Core F: Molecular Core

The major goal of this core is to provide molecular services and analysis to CF investigators.
Role: Core Leader

OSTROW04G0 (Ostrowski) 4/1/05-3/31/07
Cystic Fibrosis Foundation
The Minimal Level of CFTR Necessary for Correction of ENaC Function *in Vivo*

The major goal of this project is to use an inducible, cell-type specific promoter to express different levels of normal mouse CFTR protein in the airways of CF mice to determine in both *in vivo* and *in vitro* models the amount of CFTR protein needed to correct the sodium absorption defect in CF.
Role: Co-Investigator

5 P01 HL 034322-20 (Boucher) 4/1/01-3/31/07 (NCE) 2.4 calendar
NIH/NHLBI $144,444 (sub only)
Pulmonary Epithelia in Health and Disease, Core B: Molecular and Protein Core

The major goal of this Core is to provide a centralized facility for development of molecular reagents and antibodies required for the projects within the Program Project “Pulmonary Epithelia in Health and Disease”, including vector construction, production of tagged proteins, protein expression, synthesis and analysis of antibodies, RNA analysis, and maintenance of cDNA clones and cloning of cDNA sequences.
Role: Core Leader

MALL03G0 (O’Neal) 9/1/04-8/31/06 1.2 calendar
Cystic Fibrosis Foundation $159,120
Chronic Pseudomonas aeruginosa Infection Model in Sodium Hyperabsorbing Mouse

The major goal of this project is to develop a mouse model for chronic lung infection with Pseudomonas aeruginosa and other CF related pathogens. We will test the hypothesis that chronic Pseudomonas aeruginosa infection of beta-ENaC transgenic mice requires (1) chronic bacterial exposure with Pseudomonas aeruginosa, and/or (2) co-infection with respiratory viruses and/or other bacterial CF pathogens like Staphylococcus aureus or Haemophilus influenzae.
Role: PI

5 P01 HL 066973-05 (Samulski) 9/30/01-7/31/06
NIH/NHLBI
Gene Therapy for Pulmonary & Hematologic Disorders, Project 3: Extra Cellular Barriers to Gene Transfer in the Lung

The major goals of this project are to investigate (1) whether the transported mucus layer is a vector barrier, (2) identification of the components of the glycocalyx of human bronchial epithelia *in vitro* that act as a barrier to gene transfer, and (3) whether mouse models can be used to evaluate the role of the glycocalyx as a barrier to gene transfer *in vivo*.
Role: Co-Investigator

5 R01 HL 070199-04 (Ostrowski) 4/1/02-3/31/06
NIH/NHLBI
A Ciliated Cell-Specific Promoter for Gene Therapy of CF

The major goal of this project is to develop a ciliated cell-specific promoter that will improve the effectiveness of gene therapy for cystic fibrosis.
Role: Co-Investigator

RIBEIR03FG0 (Ribeiro) 1/1/04-12/31/05
Cystic Fibrosis Foundation
Azithromycin Effect on Airway Epithelial Gene Transfer

The major goals of this project are to evaluate the effect of azithromycin on the inflammatory response of normal airway epithelia triggered by acute *in vitro* mucosal SMM exposure, and to evaluate the effect of azithromycin on the inflammatory response of CF airway epithelia chronically exposed *in vivo* to airway bacterial infection during the course of disease.
Role: Co-Investigator

5 R01 HL 58342-08 (Johnson) 5/1/01-4/30/05
NIH/NHLBI & NIDDK
Enhanced Gene Transfer to Lung Epithelia

The major goals of this project are to (1) characterize the intercellular junctional proteins in respiratory epithelia, (2) identify rapidly acting reversible agents that modulate paracellular permeability and determine their mechanism of action, and (3) use regulators of junctional permeability and agents that stimulate surfactant uptake to enhance gene transfer to respiratory epithelia by a variety of gene transfer vectors.
Role: Co-Investigator

ONEAL00V0 (O'Neal) 3/1/01-2/28/05 (NCE) 1.8 calendar
Cystic Fibrosis Foundation $406,001
CF Therapeutic Targets Revealed by Expression Arrays

The major goal of this project is to identify novel therapeutic targets for CF lung disease by using gene expression arrays to reveal genes important in the pathogenesis of CF.
Role: PI

S880 (Boucher) 12/1/01-11/30/02 0.6 calendar
Cystic Fibrosis Foundation $50,000 (sub only)
Gene Therapy Center, Project XV: Evaluation of Helper-dependent Adenoviral Vectors for Delivery to Lung Epithelium

The major goals of this project are to study the role of the protein MUC4 in the prevention of gene transfer vector introduction by generating mice lacking MUC4 and testing gene transfer in them.
Role: Project PI

R026 (Boucher) 3/1/00-6/30/02 1.8 calendar
Cystic Fibrosis Foundation $106,550 (sub only)
Core F: Molecular Core

The major goal of this core is to provide molecular services and analysis to CF investigators.
Role: Core Leader

S880 (Boucher) 12/1/99-11/30/01 0.6 calendar
Cystic Fibrosis Foundation $50,000 (sub only)
Gene Therapy Center, Project III: MUC4 as a Barrier to *In Vivo* Gene Transfer

The major goals of this project are to study the role of the protein MUC4 in the prevention of gene transfer vector introduction by generating mice lacking MUC4 and testing gene transfer in them.
Role: Project Leader

ONEAL00I0 (O'Neal) 4/1/00-3/31/01 3.0 calendar
Cystic Fibrosis Foundation $40,000
In Vitro Cell Models for CFTR Expression

The major goal of this project is to provide flexible, reliable systems for introducing wild-type and mutant CFTR genes into a variety of relevant *in vitro* culture models.
Role: PI

5 R21 DK 53927-02 (O'Neal) 8/1/98-7/31/00 7.2 calendar
NIH/NIDDK $100,000
Large Deletion Adenoviral Vectors for Cystic Fibrosis

The major goal of this project is to develop improved vectors for use in clinical trials for cystic fibrosis gene therapy.
Role: PI

8) Service

2020-present	Cardiovascular and Respiratory Sciences Integrated Review Group, Ad-Hoc Reviewer
2020-present	NHLBI LCMI Study Section, Permanent Member
2020-present	Co-Chair, Cystic Fibrosis Foundation Path to a Cure Review Panel
2020-present	Chair, PreCISE Network (NHLBI) Quality Control and Biospecimens Committee
2019-present	NHLBI, F10A-K Fellowship Review Panel, Ad Hoc Reviewer
2019-present	Advisory Committee, Cystic Fibrosis Foundation Animal Resource Core
2018-present	Chair, Biospecimen and Biomarker Committee, Co-Chair Quality Control Committee, NHLBI PrecISE Trial Network
2012-present	Chair, Biomarker Working Group and Ancillary Studies Committee, SPIROMICS
2018	NHLBI LCMI Study Section Ad-Hoc Reviewer
2015-2018	NHLBI K99 Study Section, Permanent Member
2010-2012	NHLBI K99 Award Study Section Ad-Hoc Reviewer
2004-2007	NHLBI SBIR Study Section Ad-Hoc Reviewer

9) Research Statement

As an undergraduate at North Dakota State University, I trained in the basics of disease pathogenesis and genetics related to agriculture. The focus on agriculture in these early years stemmed from my love of the farm and my love of farmers. My switch from agricultural to "medical" sciences developed over time as I realized my desire to study cystic fibrosis (CF), which was devastating to my own siblings. This new understanding led me to pursue my PhD in Human and Molecular Genetics. I was accepted as a PhD candidate at Baylor College of Medicine, now one of the premier institutes in the country tackling this topic, under the direction of Dr. Arthur Beaudet. During this time, I transitioned into a large and competitive Howard Hughes Medical Institute funded laboratory. In retrospect, my training at NDSU was excellent. It provided me a very solid background in disease pathogenesis and genetics, and I thrived in Dr. Beaudet's laboratory. I was able to come out of it a well-trained molecular geneticist capable of technically working with animal models, DNA, and RNA.

At the time of my move to North Carolina, I was focused on gene therapy for CF in my postdoctoral work, following the development of one of the first mouse models for CF and some interesting human genetics related to the F508del mutation as my PhD dissertation. Throughout my training, I was aware of the work of the Cystic Fibrosis Research and Treatment Center (now the Marsico Lung Institute) directed by Dr. Richard Boucher. The work of this group in CF disease pathogenesis was (and still is) world-renowned. Again, by luck or fate, and the help of great people, I was introduced to Dr. Boucher through Dr. Beaudet. Modern molecular biology, with the advent of cloning and polymerase chain reaction and next-generation sequencing, was at its blossoming stage, and my training nicely complemented the skills of the faculty (physiology/cell biology) at the Center at the time. I took on a role as "resident molecular biologist," which morphed into Director of the Molecular Biology and Animal Models Core, a position that I still hold today. I acknowledge my colleague, Dr. Alessandra Livaraghi-Butrico, who now thankfully serves as Co-Director of the Animals Models side of the Core.

As a Core Director, I am working with the top CF researchers. Through the years, I have been able to participate in a wide array of research efforts, often on the periphery, but always with appreciation. I hope that a brief review of the titles of the papers for which I serve as co-author provides a sense of the breadth of the years. In short, I have worked to develop and/or phenotype a number of animal models (over 20 at last count), introduce the Center to new techniques (from PCR, to quantitative PCR, to RNA/DNA microarrays, and now to single-cell RNA sequencing and spatial transcriptomics) that have been successfully applied to multiple manuscripts and research projects. The efforts of the Core have supported multiple grant applications and trained a constant stream of new post-docs, fellows, technicians, and students throughout the last 20-plus years. My hope through all of this is that I have played some small part in the incredible success the field as a whole, which has seen a dramatic increases in life span and quality of life brought about by modulator therapies. When I started in CF research, the goal was to make progress. What has been accomplished instead are massive gains. The humbling reality is that treatments available today would have allowed my own siblings to survive and live productive lives. I have been extremely lucky to have been a small part of this progress.

Another defining feature of my research career has been the expansion of research interests beyond CF. This of course has everything to do with the Marsico Lung Institute and its incredible faculty who have far-reaching research interests beyond CF, to include asthma, COPD, bronchiectasis, bronchopulmonary dysplasia, primary ciliary dyskinesia, and (of relevance today) respiratory pathogens (including viruses and SARS-CoV-2). What I have learned is that what is on the forefront of CF biology and physiology is often on the forefront of respiratory biology, and what is learned in CF can often apply to a multitude of disease situations, providing an opportunity for even broader impact. Thus, we see that cigarette smoking affects CFTR function; mucus accumulation in the CF lung brought about by loss of CFTR affects patients with asthma and bronchitis in similar ways; disease manifestations related to airway inflammation/infection and alterations in physiology seen in CF lung disease apply across a multitude of respiratory conditions. As the connections keep growing, through my interactions with large cohorts, such as SPIROMICS (COPD) and PreCISE (asthma) the impact becomes exponential and the opportunities for understanding compound. SARS-CoV2 has recently provided another opportunity for important collaborations where my input has been of some value. The challenge moving forward, as it has been in the past, is “keeping up” with the technology and the insights it provides – never a dull moment.

10) Teaching Statement

As a research scientist in a clinical institute, I do not have any formal teaching responsibilities. My teaching, then, involves training. One of the primary purposes of the Core is to train in molecular biology methods and the use of mouse models. The training occurs in the environment of regular lab meetings, departmental seminars, manuscript preparation, grant support, and one-on-one meetings with postdocs, faculty, and associated staff. For non-faculty trainees, my philosophy is to generate independent research scientists capable of running their own projects with high integrity. For faculty, it is important to help integrate methods into ongoing research efforts. High scientific rigor and careful consideration of experimental design is a goal, coupled with an appreciation for the benefits and risk of each method. In addition, it is important to me to disseminate the information as much as possible by encouraging publication and public presentation of data. It is hoped that my efforts to teach and train are producing high quality manuscripts that stand the test of time and highly successful independent scientists who are careful thinkers and doers. Formally, I have taken my turn to lead, and present at, the departmental seminar series have run the Animal Models research meeting. In these types of settings, my ability and willingness to teach is hopefully apparent.

11) Diversity Statement

I have not been directly involved in formal DEI activities. I do actively seek to promote a positive, inclusive learning/working environment within my laboratory for the staff, trainees, and students. We provide our Core services without regards to individual characteristics or beliefs, and we base the provision of these services purely on scientific interest and necessity within our resource limitations. All staff are expected to undergo the

necessary DEI training and are encouraged to participate in DEI activities. The excellent communication that our administrative staff provides leads to seamless communication of all university and School of Medicine DEI-focused activities to the staff, which provides the necessary awareness. On the peer review side, we as reviewers are asked to evaluate grants and manuscript proposals for inclusion of diverse populations to ensure that, as much as possible and scientifically reasonable, diverse populations (children, women, and minorities) are included in research studies. This is critical for scientific equity, and it is one of the important aspects of my personal reviews. As an employer, I have had the great privilege of supporting a diverse employee base throughout the years, and I am grateful to have worked with them all.