6/13/22

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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)*

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

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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

<u>PENDING</u>

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

<u>COMPLETED</u>

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.