

# Cystic Fibrosis Center of Excellence

Volume 6, Issue 3

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Dear friends, patients, families, and colleagues,

As the crazy year that was 2016 draws to a close, we write to send you the latest edition of our e-newsletter. As always, there is much to report on in the following pages. Our main focus for this e-newsletter is reporting on the 2016 North American Cystic Fibrosis Conference, held this past October in Orlando, FL. The CF@LANTA team was very well represented at the conference, which is the largest gathering of scientists, physicians, and others focused on CF in the world. CF@LANTA members presented their research progress, led symposia and workshops, and even presented one of the plenary sessions. See the following pages for a list of ways in which we contributed to the conference.

In addition, we share the news that our own Dr. Daniel Caplan will retire in February, 2017. Dr. Caplan is well known as the “grandfather of CF” in Atlanta, and is much loved by his patients, for whom he has provided care for 48 years. We plan to celebrate his life, his commitment to CF patients, and his contributions to our CF Program at the annual Dr. Dan Caplan CF Family Science Dinner, in the Spring. Please be on the lookout for details, to be announced in the coming months.

Finally, we report on the recent book club that we held on November 30. For the second year in a row, we gathered together to discuss a book that has ties to science and the role of science in the world. This year’s book, “Brave Genius: A Scientist, a Philosopher, and Their Daring Adventures from the French Resistance to the Nobel Prize,” by Sean Carroll, was recommended by Dr. Rabin Tirouvanziam, Assistant Professor of Pediatrics. The book describes the great contributions of Albert Camus and Jacques Monod, both Frenchman, during the invasion

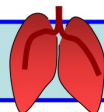
of France by Germany in World War II, and then the important work of both men following the war: giants in philosophy and molecular biology, respectively. Camus was awarded the Nobel Prize in Literature in 1957, and Monod shared the Nobel Prize in Physiology or Medicine with two other colleagues in 1965. A mixture of discussion of the fight for justice and the scientific search for the fundamental nature of life was shared by about ten team members, over (of course) French wine and cheese.

We hope that each of you has a wonderful holiday season, and are able to spend quiet time with family and friends. As we enter a new year, we recall the many blessings that we share – great friends, great colleagues, great excitement for what lies ahead in 2017. Those of us who are healthy will remember those whose health is threatened by CF, asthma, and many other diseases, and we will become even further motivated to do all we can on behalf of our patients – those we know and the many who count on us for their futures.

With best wishes,  
Nael A. McCarty, PhD, [CF@LANTA](mailto:CF@LANTA)



Center for Cystic Fibrosis  
and Airways Disease Research  
Advancing Wellness in Patients Through Research

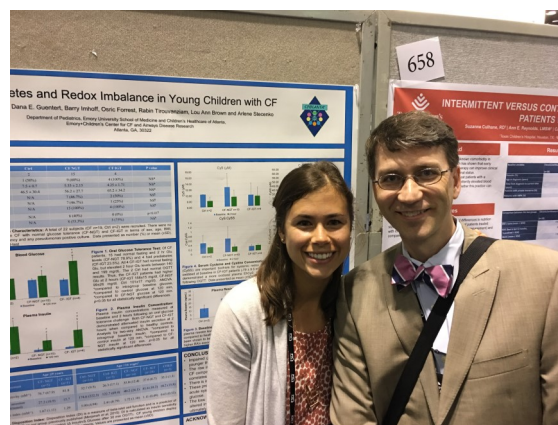


## NACFC Presentations 2016

The 2016 North American Cystic Fibrosis Conference was held in Orlando, Florida. **CF@LANTA** was represented at all levels of presentations, sharing all of the great work happening in our center.

### Posters

- Sabusap, C.P.; Fu, L.; McNicholas-Bevensee, C.M.; Wang, W.; Chung, W.; Wen, H.; Mazur, M.; Kirk, K.; Collawn, J.F.; Hong, J.S.; Sorscher, E.J. CHARACTERIZATION OF P67L CFTR AS A MODEL OF "PRECISION" THERAPEUTICS FOR RARE CFTR ALLELES
- Cui, G.; Khazanov, N.; Imhoff, B.; Senderowitz, H.; McCarty, N.A. NOVEL POTENTIATORS OF CYSTIC FIBROSIS TRANSMEMBRANE CONDUCTANCE REGULATOR IDENTIFIED WITH PHARMACOPHORE MODELING AND EXPERIMENTAL SCREENING
- Goeckeler-Fried, J.; Estabrooks, S.K.; Chiang, A.; Chung, W.; Ye, Z.; Denny, R.A.; Weissman, A.M.; Camacho, C.J.; Sorscher, E.J.; Brodsky, J.L. IDENTIFICATION OF NOVEL INHIBITORS OF UBIQUITINATION THAT IMPROVE RESCUE OF F508DEL-CFTR BY VX-809
- Liang, F.; Wong, E.; Jordan, N.J.; Haibo, S.; Fitzpatrick, L.M.; Hong, J.S.; Rab, A.; Sorscher, E.J.; Mense, M. COMBINATIONS OF TRAFFICKING/FOLDING CORRECTORS AND READTHROUGH MODULATORS AS THERAPEUTIC APPROACH FOR CFTR PTC MUTATIONS
- Oliver, K.E.; Wang, W.; Wolpert, M.J.; Maya, J.; Icyuz, M.; Hong, J.S.; Hartman, J.L.; Sorscher, E.J. RIBOSOMAL TARGETING CORRECTS CFTR CLASS I AND II DEFECTS
- Chung, W.; Fortenberry, J.A.; Sokirniy, I.; Wu, J.; Chiang, A.; Goeckeler-Fried, J.; Sterner, D.; Mattern, M.; Brodsky, J.L.; Sorscher, E.J.; Kumar, S. AUGMENTATION OF THE AVAILABLE POOL OF CFTR FOR SMALL MOLECULE CORRECTION BY INHIBITING THE E3 LIGASE, GP78
- Day, B.J.; Min, E.; Nichols, D.; Chandler, J.D. BACTERIAL THIOREDOXIN REDUCTASE IS A TARGET FOR HYPOTHIOCYANITE-MEDIATED ANTIMICROBIAL EFFECTS
- Stauffer, B.; Cui, G.; Infield, D.T.; McCarty, N.A. S. AUREUS SPHINGOMYELINASE IS A STATEDEPENDENT INHIBITOR OF CFTR
- Beaty, T.C.; Brown, M.R.; Forrest, O.A.; Ingersoll, S.; Stecenko, A.; Tirouvanziam, R. EFFECT OF INHALED CORTICOSTEROID THERAPY WITHDRAWAL ON CF LUNG FUNCTION AND INFLAMMATORY MARKERS
- Chandler, J.D.; Horati, H.; Scholte, B.J.; Jones, D.P.; Peng, L.; Gaggar, A.; Janssens, H.; Tirouvanziam, R. UNTARGETED METABOLOMICS OF CF INFANT BALF IDENTIFIES SIGNATURES ASSOCIATED WITH EARLY AIRWAY DISEASE BASED ON PRAGMA CT SCORING
- Bedi, B.; Yuan, Z.; Varga, J.J.; Wei, J.W.; Goldberg, J.B.; Stecenko, A.; Sadikot, R.T. PSEUDOMONAS AERUGINOSA ISOLATES FROM PATIENTS WITH CYSTIC FIBROSIS INHIBIT PPAR $\gamma$  EXPRESSION IN MACROPHAGES AND EPITHELIAL CELLS
- Forrest, O.A.; Ingersoll, S.; Preininger, M.; Laval, J.; Limoli, D.; Brown, M.R.; Lee, F.; Bedi, B.; Sadikot, R.T.; Goldberg, J.B.; Tangpricha, V.; Tirouvanziam, R. IN VITRO TRANSMIGRATION OF NEUTROPHILS INTO CF AIRWAY FLUID INDUCES FUNCTIONAL AND METABOLIC REPROGRAMMING CHARACTERISTIC OF CF AIRWAY NEUTROPHILS IN VIVO
- Rada, B.; Floyd, M.; Yoo, D.; Cullen, C. SWIMMING MOTILITY MEDIATES NET FORMATION STIMULATED BY FLAGELLATED PSEUDOMONAS AERUGINOSA
- Molina, S.A.; Vance, R.J.; Imhoff, B.; McCarty, N.A.; Koval, M. ACTIVATION OF AKT SIGNALING IN HUMAN AIRWAY CELLS EXPRESSING MUTANT CFTR DIFFERS COMPARED TO WILD-TYPE



### NACFC Presentations 2016 continued

- Icyuz, M.; Mutyam, V.; Rowe, S.M.; Sorscher, E.J.; Hartman, J.L. GENOME-WIDE ANALYSIS IN YEAST TO IDENTIFY MOLECULAR TARGETS PROMOTING READTHROUGH
- Plyler, Z.E.; Schoeb, T.R.; Hong, J.S.; Tuggle, K.L.; Birket, S.; Schultz, B.D.; Sorscher, E.J. ANALYSIS OF VAS DEFERENS ATRESIA IN THE CF RAT
- Rab, A.; Wen, H.; Manfredi, C.; Ehrhardt, A.; Joshi, D.; Sorscher, E.J.; Hong, J.S. MODEL SYSTEMS FOR INVESTIGATING MOLECULAR PATHOGENESIS OF RARE CFTR ALLELES
- Bihler, H.; Harrington, J.; Musisi, I.; Mercadante, D.; Hong, J.S.; Rab, A.; Sorscher, E.J.; Saltz, J.; Mahiou, J.; Wong, E.; Liang, F.; Mense, M. FUNCTIONAL CHARACTERIZATION OF ESTABLISHED AND NOVEL NONSENSE MUTATION READTHROUGH MODULATORS IN ENGINEERED FRT CELL LINES AND PRIMARY HUMAN BRONCHIAL EPITHELIAL (HBE) CELLS
- Scholte, B.J.; Veltman, M.; Horati, H.; Koval, S.; Vreeken, R.; Harms, A.; Tiddens, H.; Janssens, H.; Tirouvanziam, R.; Stick, S.M. BIOACTIVE LIPIDS IN BRONCHOALVEOLAR LAVAGE OF CF INFANTS CORRELATE WITH LUNG CT SCORE, REVEALING THERAPEUTIC TARGETS
- Haibo, S.; Wong, E.; Jordan, N.J.; Fitzpatrick, L.M.; Hong, J.S.; Rab, A.; Sorscher, E.J.; Liang, F.; Mense, M. HIGH-THROUGHPUT SCREENING FOR READTHROUGH MODULATORS OF CFTR PTC MUTATIONS Y122X AND W1282X
- Manfredi, C.; Rab, A.; Joshi, D.; Wang, W.; Kirk, K.; Saltz, J.; Mense, M.; Mahiou, J.; Sorscher, E.J.; Hong, J.S.; Ehrhardt, A. MOLECULAR ANALYSIS OF THE CFTR W1282X MUTATION
- Forrest, O.A.; Chopyk, D.; Ingersoll, S.; Brown, M.R.; Scholte, B.J.; Janssens, H.; Alvarez, J.A.; Tangpricha, V.; Tirouvanziam, R. RESISTIN, A NEUTROPHIL-DERIVED IMMUNOMETABOLIC MEDIATOR, IS A CELLSURFACE ASSOCIATED AND SOLUBLE MARKER OF EARLY AND CHRONIC CF AIRWAY DISEASE
- Walker, S.D.; Henry, B.; Sueblinvong, V.; Hunt, W.R. EXPERIENCE OF A LARGE ACADEMIC CF CENTER WITH LUMACAFTOR/IVACAFTOR
- Zhang, Z.; Ma, H.; Peng, L.; Lai, H.J. GUT MICROBIOTA IN EARLY CHILDHOOD IN CYSTIC FIBROSIS
- Yang, J.; Peng, L.; Zhang, Z.; Rahman, A.; Ma, H.; Lai, H.J. JOINT PROFILE OF RESPIRATORY INFECTIONS AND THEIR ASSOCIATIONS WITH BREASTFEEDING IN INFANTS WITH CF
- Griego, B.; Tataria, S.; Kirchner, K. INVESTIGATING THE IMPROVEMENT IN PHYSICAL AND PULMONARY ENDURANCE AS MEASURED BY THE MODIFIED SHUTTLE WALK TEST AND FEV1 IN PEDIATRIC PATIENTS ADMITTED TO THE HOSPITAL FOR ACUTE CYSTIC FIBROSIS EXACERBATION
- Patel, K.; Britt, E.; Duval, M.A.; Kirchner, K. VITAMIN D SUPPLEMENTATION: HOW MUCH IS ADEQUATE? OUTCOMES FROM A QUALITY IMPROVEMENT PROJECT AT A PEDIATRIC CYSTIC FIBROSIS CENTER
- Lindsay, K.L.; Revilla, E.; Tangpricha, V.; Stecenko, A. EFFECTIVENESS OF WEIGHT-BASED, WEEKLY DOSING OF VITAMIN D3 IN CHILDREN WITH CYSTIC FIBROSIS
- Ling, S.; Ye, W.; Leung, D.H.; Weymann, A.; Karnsakul, W.; Freeman, A.J.; Magee, J.; Narkewicz, M.R. SIMPLE BIOMARKERS CORRELATE WITH LIVER ULTRASOUND SCAN ABNORMALITIES IN CHILDREN WITH CF
- Kanhere, M.; Ziegler, T.R.; Alvarez, J.A.; Walker, S.D.; Gewirtz, A.T.; Chassaing, B.; Tangpricha, V. PILOT STUDY EVALUATING THE ROLE OF VITAMIN D REPLETION ON GUT MICROBIOTA IN ADULT CF
- Hunt, W.R.; Guentert, D.E.; Brown, L.S.; Stecenko, A. PREDIABETES AND REDOX IMBALANCE IN YOUNG CHILDREN WITH CF
- Cohen, L.A.; Walker, S.D.; Kempker, J.; Howard, D. EFFECT OF CLINIC VISIT FREQUENCY ON CYSTIC FIBROSIS PATIENTS WITH SEVERE LUNG DISEASE: A SINGLE-CENTER OBSERVATION
- Blair, S.D.; Gardner, L. ETHICAL CONSIDERATIONS FOR IMPLEMENTING CAREGIVER MENTAL HEALTH SCREENING WITHIN PEDIATRIC INSTITUTIONS



### NACFC Presentations 2016 continued

- Middour-Oxler, B.J.; Bergman, S.; Blair, S.D.; Pendley, S.; Crews, B.; Siragusa, A.; Walker, S.D.; Stecenko, A. A RETROSPECTIVE SURVEY OF PATIENT EXPERIENCE WITH PEDIATRIC TO ADULT TRANSITION OF CF CARE
- Kaplan, E.T.; Blair, S.D.; Fitzpatrick, A.; Hunt, W.R.; Pendley, S.; Wei, J.W.; Stecenko, A. DISEASE-SPECIFIC GENETIC KNOWLEDGE IN ADOLESCENTS WITH CYSTIC FIBROSIS AND THEIR PARENTS

#### Lunch Roundtable Leaders

- Dahlman, J.: NEXT GENERATION STRATEGIES FOR CF GENE THERAPY
- Freeman, A.J.: WORKING UP ELEVATED LIVER ENZYMES
- Stephenson, K.: INCREASING CALORIES FOR INFANTS

#### Workshop Session Chair

- DiMagno, M.J.; Freeman, A.J. Gastroenterology & Hepatology
- Larson-Ode, K.; Tangpricha, V. Endocrine (Research)
- Crews, B.: Mid-level Primary Care
- Pendley, S.: Nurse Mentoring & Education
- Hong, J.: CFTR 2016



#### Workshop or Symposium

- Blair, S.: MENTAL HEALTH SCREENING
- Freeman, A.J.: GASTROENTEROLOGY & HEPATOLOGY, INTRODUCTION
- Stecenko, A.A.: ROLE OF INSULIN SECRETION & OXIDATIVE STRESS ON THE DEVELOPMENT OF CFRD: A CLINICAL PERSPECTIVE
- Goldberg, J.B.: VACCINES FOR CF INFECTIONS: EPISODE IV - A NEW HOPE

#### Plenary

- Drumm, M.L.; Sorscher E.J. A CURE FOR ALL: LEAVING NO ONE BEHIND

#### Selected as one of the top 5 poster presenters

- Chandler, J.
- Plyler, Z.E.

#### Chair of CFTR Folding Consortium Meeting

- Sorscher, E.J.



## New Faculty: Jeong Hong, PhD



Jeong Hong, PhD

Dr. Hong trained as a molecular virologist with a focus on adenovirus fiber protein responsible for binding to cell surface receptor. Her work led to development of strategies to modify cellular tropism of adenovirus and formed the basis for studies of therapeutic viral

retargeting by numerous laboratories.

A long standing interest of her laboratory has pursued studies of cystic fibrosis (CF). For more than 20 years, she has directed laboratory aspects

of a highly collaborative and productive joint CF research program with Dr. Eric Sorscher, former head of the UAB CF Research Center and now a professor at Emory and a member of [CF@LANTA](#). Her group investigates structural and functional aspects of the CFTR gene product in relation to disease associated mutations, with the goal of promoting new 'precision-type' treatments. She has served as a member of the international CF therotyping initiative since its inception in 2014, and developed epithelial cell models encoding clinically relevant CF alleles.

Outside lab, Dr. Hong enjoys gardening and spending time with her family.

~submitted by Jeong Hong

## New Faculty: Joon Chung, PhD



Joon Chung, PhD

Joon Chung is a new faculty of the Center for Cystic Fibrosis and Airways Disease research (CF-AIR). After receiving his PhD and postdoctoral fellowship from the University of Alabama at Birmingham, he had joined the cystic fibrosis research center at UAB where his

research had been focused on new approaches to develop drugs that improves rescue of defective CFTR proteins. He joined the Emory University Pediatrics Department as an assistant professor in September 2016. Dr. Chung's research is focused

on understanding mechanisms underlying defects of CFTR mutations and developing experimental therapeutics. He has developed a novel biochemical drug screening method targeting intra molecular domain interactions to improve the chloride channel activity of F508del CFTR. Currently, he is developing a novel class of CFTR modulators ('amplifiers') that complement the substrate limitations of other CF therapeutic modalities such as 'correctors' and 'potentiators'. In particular, he is evaluating experimental drugs that target the ubiquitination steps in the protein degradation pathway to augment pools of CFTR mutant proteins for pharmacologic correction.

Outside of his research, he is enjoying a variety of cultural and culinary experiences with his family and learning to navigate busy streets in Atlanta.

~submitted by Joon Chung

## Recent CF-AIR Publications

- Alvarez JA, Grunwell JR, Gillespie SE, Tangpricha V, Hebbar KB. Vitamin D deficiency is associated with an oxidized plasma cysteine redox potential in critically ill children. *J Steroid Biochem Mol Biol*. 2016 Sep 15. pii:S0960-0760(16)30249-7. doi:10.1016/j.jsbmb.2016.09.013. [Epub ahead of print] PubMed PMID: 27641738.
- Balekian DS, Linnemann RW, Hasegawa K, Thadhani R, Camargo CA Jr. Cohort Study of Severe Bronchiolitis during Infancy and Risk of Asthma by Age 5 Years. *J Allergy Clin Immunol Pract*. 2016 Aug 11. pii: S2213-2198(16)30270-7. doi:10.1016/j.jaip.2016.07.004. [Epub ahead of print] PubMed PMID: 27523277.
- Bedi B, Yuan Z, Joo M, Zughair SM, Goldberg JB, Arbiser JL, Hart CM, Sadikot RT. Enhanced Clearance of *Pseudomonas aeruginosa* by Peroxisome Proliferator-Activated Receptor Gamma. *Infect Immun*. 2016 Jun 23;84(7):1975-85. doi:10.1128/IAI.00164-16. PubMed PMID: 27091928; PubMed Central PMCID: PMC4936346.
- Burnham EL, McNally A, Gaydos J, Brown LA. The Relationship Between Airway Antioxidant Levels, Alcohol Use Disorders, and Cigarette Smoking. *Alcohol Clin Exp Res*. 2016 Oct;40(10):2147-2160. doi:10.1111/acer.13201. PubMed PMID: 27627733.
- Chung WJ, Goeckeler-Fried JL, Havasi V, Chiang A, Rowe SM, Plyler ZE, Hong JS, Mazur M, Piazza GA, Keeton AB, White EL, Rasmussen L, Weissman AM, Denny RA, Brodsky JL, Sorscher EJ. Increasing the Endoplasmic Reticulum Pool of the F508del Allele of the Cystic Fibrosis Transmembrane Conductance Regulator Leads to Greater Folding Correction by Small Molecule Therapeutics. *PLoS One*. 2016 Oct 12;11(10):e0163615. doi:10.1371/journal.pone.0163615. PubMed PMID: 27732613; PubMed Central PMCID: PMC5061379.
- Cui G, Khazanov N, Stauffer BB, Infield DT, Imhoff BR, Senderowitz H, McCarty NA. Potentiators exert distinct effects on human, murine, and *Xenopus* CFTR. *Am J Physiol Lung Cell Mol Physiol*. 2016 Aug 1;311(2):L192-207. doi:10.1152/ajplung.00056.2016. PubMed PMID: 27288484.
- Fitzpatrick AM, Stephenson ST, Brown MR, Nguyen K, Douglas S, Brown LA. Systemic Corticosteroid Responses in Children with Severe Asthma: Phenotypic and Endotypic Features. *J Allergy Clin Immunol Pract*. 2016 Sep 21. pii: S2213-2198(16)30376-2. doi:10.1016/j.jaip.2016.08.001. [Epub ahead of print] PubMed PMID: 27665382.
- Floyd M, Winn M, Cullen C, Sil P, Chassaing B, Yoo DG, Gewirtz AT, Goldberg JB, McCarter LL, Rada B. Swimming Motility Mediates the Formation of Neutrophil Extracellular Traps Induced by Flagellated *Pseudomonas aeruginosa*. *PLoS Pathog*. 2016 Nov 17;12(11):e1005987. doi:10.1371/journal.ppat.1005987. PubMed PMID: 27855208.
- Gauthier TW, Guidot DM, Kelleman MS, McCracken CE, Brown LA. Maternal Alcohol Use During Pregnancy and Associated Morbidities in Very Low Birth Weight Newborns. *Am J Med Sci*. 2016 Oct;352(4):368-375. doi:10.1016/j.amjms.2016.06.019. PubMed PMID: 27776718; PubMed Central PMCID: PMC5098418.
- Gauthier TW, Brown LA. In utero alcohol effects on foetal, neonatal and childhood lung disease. *Paediatr Respir Rev*. 2016 Aug 19. pii:S1526-0542(16)30080-X. doi:10.1016/j.prrv.2016.08.006. [Epub ahead of print] Review. PubMed PMID: 27613232.
- Grisanti LA, Gumpert AM, Traynham CJ, Gorsky JE, Repas AA, Gao E, Carter RL, Yu D, Calvert JW, García AP, Ibáñez B, Rabinowitz JE, Koch WJ, Tilley DG. Leukocyte-Expressed  $\beta$ 2-Adrenergic Receptors Are Essential for Survival After Acute Myocardial Injury. *Circulation*. 2016 Jul 12;134(2):153-67. doi:10.1161/CIRCULATIONAHA.116.022304. PubMed PMID: 27364164.
- Hasegawa K, Linnemann RW, Mansbach JM, Ajami NJ, Espinola JA, Fiechtner LG, Petrosino JF, Camargo CA Jr. Association of household siblings with nasal and fecal microbiota in infants. *Pediatr Int*. 2016 Sep 16. doi:10.1111/ped.13168. [Epub ahead of print] PubMed PMID: 27638139.
- Hasegawa K, Linnemann RW, Mansbach JM, Ajami NJ, Espinola JA, Petrosino JF, Piedra PA, Stevenson MD, Sullivan AF, Thompson AD, Camargo CA Jr. The Fecal Microbiota Profile and Bronchiolitis in Infants. *Pediatrics*. 2016 Jul;138(1). pii: e20160218. doi:10.1542/peds.2016-0218. PubMed PMID: 27354456; PubMed Central PMCID: PMC4925084.
- Hunt WR, Helfman BR, McCarty NA, Hansen JM. Advanced glycation end products are elevated in cystic fibrosis-related diabetes and correlate with worse lung function. *J Cyst Fibros*. 2016 Sep;15(5):681-8.

doi:10.1016/j.jcf.2015.12.011. PubMed PMID: 26817932.

- Klingensmith NJ, Yoseph BP, Liang Z, Lyons JD, Burd EM, Margoles LM, Koval M, Ford ML, Coopersmith CM. Epidermal Growth Factor Improves Intestinal Integrity and Survival in Murine Sepsis Following Chronic Alcohol Ingestion. *Shock*. 2016 Jul 25. [Epub ahead of print] PubMed PMID: 27465753.
- Ku DN, Ku SK, Helfman B, McCarty NA, Wolff BJ, Winchell JM, Anderson LJ. Ability of device to collect bacteria from cough aerosols generated by adults with cystic fibrosis. *Fl000Res*. 2016 Aug 5;5:1920. PubMed PMID: 27781088; PubMed Central PMCID: PMC5054809.
- McClure ML, Barnes S, Brodsky JL, Sorscher EJ. Trafficking and function of the cystic fibrosis transmembrane conductance regulator: a complex network of posttranslational modifications. *Am J Physiol Lung Cell Mol Physiol*. 2016 Oct 1;311(4):L719-L733. doi:10.1152/ajplung.00431.2015. Review. PubMed PMID: 27474090.
- Meisner J, Goldberg JB. The Escherichia coli rhaSR-PrhaBAD Inducible Promoter System Allows Tightly Controlled Gene Expression over a Wide Range in Pseudomonas aeruginosa. *Appl Environ Microbiol*. 2016 Oct 27;82(22):6715-6727. PubMed PMID: 27613678; PubMed Central PMCID: PMC5086565.
- Owings JP, McNair NN, Mui YF, Gustafsson TN, Holmgren A, Contel M, Goldberg JB, Mead JR. Auranofin and N-heterocyclic carbene gold-analogs are potent inhibitors of the bacteria Helicobacter pylori. *FEMS Microbiol Lett*. 2016 Jul;363(14). pii: fnw148. doi:10.1093/femsle/fnw148. PubMed PMID: 27279627.
- Qadi M, Lopez-Causapé C, Izquierdo-Rabassa S, Mateu Borrás M, Goldberg JB, Oliver A, Albertí S. Surfactant protein A recognizes outer membrane protein OprH on Pseudomonas aeruginosa chronic infection isolates. *J Infect Dis*. 2016 Aug 19. pii: jiw387. [Epub ahead of print] PubMed PMID: 27543671.
- Sabusap CM, Wang W, McNicholas CM, Chung WJ, Fu L, Wen H, Mazur M, Kirk KL, Collawn JF, Hong JS, Sorscher EJ. Analysis of cystic fibrosis-associated P67L CFTR illustrates barriers to personalized therapeutics for orphan diseases. *JCI Insight*. 2016 Sep 8;1(14). pii: e86581. PubMed PMID: 27660821; PubMed Central PMCID: PMC5029786.
- Sathe MN, Freeman AJ. Gastrointestinal, Pancreatic, and Hepatobiliary Manifestations of Cystic Fibrosis. *Pediatr Clin North Am*. 2016 Aug;63(4):679-98. doi:10.1016/j.pcl.2016.04.008. Review. PubMed PMID: 27469182.
- Schlingmann B, Overgaard CE, Molina SA, Lynn KS, Mitchell LA, Dorsainvil White S, Mattheyses AL, Guidot DM, Capaldo CT, Koval M. Regulation of claudin/zonula occludens-1 complexes by hetero-claudin interactions. *Nat Commun*. 2016 Jul 25;7:12276. doi:10.1038/ncomms12276. PubMed PMID: 27452368; PubMed Central PMCID: PMC4962485.
- Sulton C, McCracken C, Simon HK, Hebbar K, Reynolds J, Cravero J, Mallory M, Kamat P. Pediatric Procedural Sedation Using Dexmedetomidine: A Report From the Pediatric Sedation Research Consortium. *Hosp Pediatr*. 2016 Sep;6(9):536-44. doi:10.1542/hpeds.2015-0280. PubMed PMID: 27516413.
- Van Avermaete AE, Trac PT, Gauthier TW, Helms MN. Real-time X-ray Imaging of Lung Fluid Volumes in Neonatal Mouse Lung. *J Vis Exp*. 2016 Jul 18;(113). doi:10.3791/52751. PubMed PMID: 27500410.
- Yeligar SM, Chen MM, Kovacs EJ, Sisson JH, Burnham EL, Brown LA. Alcohol and lung injury and immunity. *Alcohol*. 2016 Sep;55:51-59. doi:10.1016/j.alcohol.2016.08.005. Review. PubMed PMID: 27788778.
- Yeligar SM, Mehta AJ, Harris FL, Brown LA, Hart CM. Peroxisome Proliferator-Activated Receptor  $\gamma$  Regulates Chronic Alcohol-Induced Alveolar Macrophage Dysfunction. *Am J Respir Cell Mol Biol*. 2016 Jul;55(1):35-46. doi:10.1165/rcmb.2015-0077OC. PubMed PMID: 26677910; PubMed Central PMCID: PMC4942203.

*If you have a publication you would like in the next newsletter, email Karen Kennedy, [kmurra5@emory.edu](mailto:kmurra5@emory.edu).*

### Recent CF-AIR visitors

- **Marshall Plaut, PhD**, August 30, *NIAID Funding opportunities: Research focused on asthma, allergy (including food allergy and atopic dermatitis) and airway biology*
- **Joshua Chandler, PhD**, October 11, *Targeting small molecules that drive big problems in cystic fibrosis and other inflammatory diseases*
- **Ineke Braakman, PhD**, November
- **Peter van der Sluijs, PhD**, November



## Events for Researchers

These events will be taking a break for the summer months, check the websites below for the upcoming schedule.

Each month there are several opportunities for CF-AIR researchers to get together to discuss their work.

- **CF-AIR Faculty and Trainees Research (CF-TR):**  
On the first Tuesday of the month, faculty chalk talks on either the overall work in their lab, or on a grant proposal planned for submission soon. On the third Tuesday of the month trainee chalk talks discussing planned manuscripts or fellowship proposals. Meet at noon in ECC 302
- **CF-AIR Workshop:**  
A weekly Wednesday meeting for research-in-progress and journal club presentations. Meet at 4:30 pm in ECC 302
- **CF Scholars Meetings:**  
A monthly program for CF Scholars, Friday afternoons, see website schedule

More information and current schedules can be found on

[www.pedsresearch.org/research/centers/cf-air/seminars-workshops/](http://www.pedsresearch.org/research/centers/cf-air/seminars-workshops/)  
and

[www.pedsresearch.org/research/centers/cf-air/cf-center-of-excellence/education-outreach/cf-scholars-program/](http://www.pedsresearch.org/research/centers/cf-air/cf-center-of-excellence/education-outreach/cf-scholars-program/)

### Clinics:

Children's Healthcare of Atlanta  
CF Care Center:  
Children's at North Druid Hills  
1605 Chantilly Drive NE  
Atlanta, GA 30324  
404-785-2000

Children's at Scottish Rite  
Cystic Fibrosis Affiliate Program  
5455 Meridian Mark Road, Suite 200  
Atlanta GA 30342  
404-785-2898

Emory Adult CF Clinic: 404-778-7929

### Website:

[www.pedsresearch.org/research/centers/cf-air](http://www.pedsresearch.org/research/centers/cf-air)

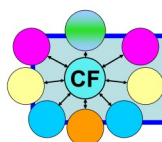
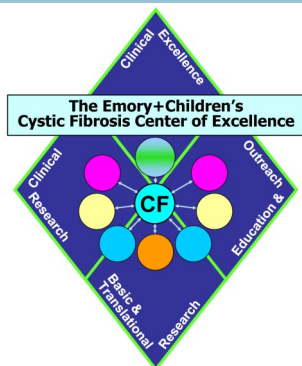
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### Contact:

Nael McCarty, PhD  
Program Director  
[namccar@emory.edu](mailto:namccar@emory.edu)

Karen Kennedy, PhD  
Program Coordinator  
[kmurra5@emory.edu](mailto:kmurra5@emory.edu)



**Center for Cystic Fibrosis  
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Advancing Wellness in Patients Through Research

