

The Emory+Children's Cystic Fibrosis Center of Excellence

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Coming April 2016: CF@LANTA Symposium and Retreat

On April 11, 2016, we will host a public CF Research Symposium, jointly with our colleagues at the University of Alabama at Birmingham. This will be followed by the Dan Caplan CF Family Science Dinner. On April 12, 2016, we will have our CF Research Retreat, for the local team members, only. These events will emphasize sharing of current work and discussing future directions toward the cure/control of cystic fibrosis. *More information coming soon.*

Update from the Director

Dear friends and supporters of *CF@LANTA*:

As we welcome the arrival of Fall, and the return of moderate weather to Atlanta, we are happy to provide you the next edition of our newsletter. We appreciate your interest in all of the exciting news coming from the CF program in Atlanta. While our most recent newsletters have focused your attention more on our research activities, I want to use this Director's Update to brag about the exciting progress we have made in growing our clinical care program and our research training activities.

One new component related to research training, made possible by our recent funding as an RDP Center, is the establishment of two new fellowships: the *CF@LANTA* RDP Pre-doctoral and Postdoctoral Fellows. These two new fellows will receive partial salary support for two years, along with funds to attend the North American CF Conference. They also are expected to participate in our innovative "CF Scholars" program. Congratulations to Mr. Brandon Stauffer and Dr. Joshua Chandler; these inaugural fellows are introduced on the next page.

We have been very busy in recruiting to our clinical care program more physicians and physician-scientists who will contribute to our goal of improving our patient outcomes. Our goal is to become the "model center" – the CF Care Center that all others use for their benchmarking, the Center with the highest outcomes for patients of all ages, and the Center that sets the bar for quality improvement. One way to accomplish this is to be recognized for having outcome measures for pulmonary function and nutrition, for both our pediatric and adult patient populations, that are ranked in the top ten, nationally. In addition, the clinical programs have several on-going quality improvement projects that are worthy of mention. For example, the Emory+Children's pediatric CF program has developed a program aimed to eliminate Vitamin D deficiency in our CF patients. When we started the program, only 20% of our CF patients had adequate levels of Vitamin D (a sufficiency level consistent with the rest of the country) and now our 2015 data show that 80% have normal levels. The Scottish Rite pediatric CF program has focused on a perfect score in achieving OGTT diabetes screening for CF patients 10 years and older as recommended by the CF Foundation. In 2014, they reached 80% success rate in obtaining the screening test, the best of our three programs. Finally, our two pediatric CF programs have transitioned 90 patients to the Adult CF program at Emory using the Journey to Independence transition program originally developed by Scottish Rite. Dr. Randy Hunt and the team have developed a patient satisfaction survey which they now are asking all 90 transitioned patients to complete so we can make this nationally recognized transition program even better and tailored to our patient's needs.

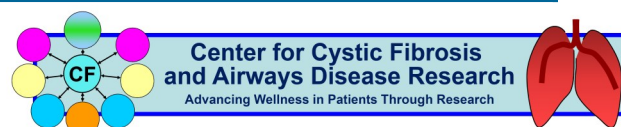
Quality improvement on behalf of our patients is the job of all of members of our care center team: physicians, staff, and trainees. But, to enable quality improvement activities, we needed more physicians. Due to the hard work of Dr. Arlene Stecenko (our Care Center Director, Chief of the PACS Division at Emory, and Director for Clinical Excellence in *CF@LANTA*), we have over the past few months recruited four new physicians to our program. All are pediatric pulmonologists, who will be working primarily in CF and/or asthma: Dr. Timothy Beaty, Dr. Devon Greene, Dr. Rachel Linnemann, and Dr. Lokesh Guglani. We are very glad to have these outstanding new faculty members on the team! Dr. Beaty was highlighted in a recent newsletter; be on the lookout for introductions to our other new faculty in the coming months.

We work closely with the local Chapter of the CF Foundation to make sure that our clinical care programs meet the complex needs of our patients and families. In this regard, we are happy to inform our readers that our Care Center has been awarded an "Outstanding Partnership Award" by the CF Foundation, which was presented to our team at the North American CF Conference in Phoenix, AZ, this month. Congratulations to all!

-Nael McCarty, PhD



Nael A. McCarty, PhD
Center Director



Inaugural RDP Predoctoral Fellow: Brandon Stauffer



Brandon Stauffer

Brandon Stauffer is originally from Lancaster, Pennsylvania. He received his Bachelor of Science in pharmacology from the University of the Sciences in Philadelphia. After graduating, He did a two-year post-baccalaureate program at the NIH in the lab of Steve Ikeda where he studied cannabinoid receptor modulation of neural calcium channels.

Brandon then joined the Molecular and Systems Pharmacology program in Emory's Laney Graduate School and chose to do his thesis in the lab of Nael McCarty, which focuses on epithelial chloride channel physiology, specifically function and regulation of cystic fibrosis transmembrane conductance regulator (CFTR). The goal of

Brandon's thesis work is elucidating the influence of a bacterial virulence factor on CFTR chloride channel activity.

Brandon's hobbies include eating a tasty variety of food (with a focus on cheese and vegetables), playing guitar and riding his bike, learning about finance and cars, and trying craft beers.

Inaugural RDP Postdoctoral Fellow: Joshua Chandler, PhD



Joshua Chandler, PhD

Joshua Chandler received his Ph.D. in Toxicology from the University of Colorado in 2014 and his bachelor's degree with majors in Chemistry and Biology from Drury University in 2009. His interest in research began after interning at the University of Kansas in the lab of C. Russell Middaugh, Ph.D. Soon after joining the University of Colorado Molecular Toxicology program, he joined the lab of Brian J. Day, Ph.D. at the Denver respiratory hospital, National Jewish Health. His research experience includes work in redox biology, respiratory toxicology, immunology, infectious diseases, biopharmaceutical formulation and adjuvant evaluation.

Joshua wrote his thesis on the role of thiocyanate in infectious and inflammatory lung diseases such as cystic fibrosis. This research culminated in the *in vitro*

identification and *in vivo* manipulation of a mammal-specific mechanism of oxidant regulation that promotes selective oxidative pressure on single-celled pathogens, resulting in three first author research publications and two first author reviews. Joshua has also received multiple academic honors including the Occupational and Public Health best abstract award from the Society of Toxicology.

By working with Dr. Dean P. Jones, Ph.D. at Emory, Joshua hopes to extend important results of his thesis research into the world of omics data, with a primary goal of identifying specific targets of inflammatory oxidants with demonstrably different effects on human cells. This research is critical to elucidating distinct *in vivo* functions of inflammatory oxidants and understanding their health implications. A

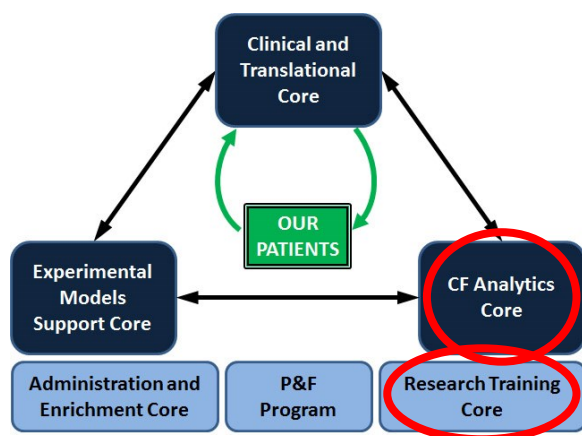
major long-term goal of Joshua's research is to develop a means to fine-tune oxidant generation in human patients to improve health outcomes without disrupting beneficial immunologic processes. CF is an obvious lung disease for targeting in this way due to the role of neutrophils in the disease and the disruption of thiocyanate in CF.

When not in the lab, Joshua is an avid jazz bassist and cyclist. His wife, Megan, is an independent property manager who suffers from multiple chronic illnesses. Her resilience inspires him to continue working on difficult problems in health research.

Research Development Program Grant Highlights: Cores

As we reported in previous newsletters, our CF research program was recently awarded a Research Development Program (RDP) grant from the CF Foundation. This large infrastructure grant enabled the establishment of the **CF@LANTA** RDP Center, with the overall goals of increasing the ability of our existing CF researchers to do their important work and drawing more local investigators into the CF research effort. Our program was established around the objective of enabling research that is close to the patient.

The grant provides funds to support three research cores plus a training core, a pilot & feasibility grant program, and an administrative core, as shown in the figure on this page. We already have made great progress in getting this program up and running. Of course, these Cores and activities will only be useful if our team members know about them; therefore, we are making use of the weekly CF-AIR Research Workshop to roll them out. The first such session was held October 21, where the CF-Analytics Core and the Research Training Core were described by their respective Directors. Brief notes are provided here, and the slides used in their presentations can be found at www.pedsresearch.org/centers/sub-pages/cf-air-cf-research-resources/. Watch for descriptions of the other Cores in future newsletters.



Learn more about the RDP grant using the QR code to the left or at www.pedsresearch.org/centers/sub-pages/cf-air-cf-research-cflanta-core-center/

Research Training Core, Professor Joanna Goldberg, PhD, Director

Dr. Goldberg described the various components available to all trainees:

- CF-AIR Research Workshop - Wednesdays at 4:30
- CF Scholars Program – 9 current members
- CF Academy – updates on state-of-the-art interdisciplinary care, open to researchers
- CF Regional Research Symposium – April 11, 2016, with Univ. Alabama at Birmingham
- **CF@LANTA** Research Retreat – April 12, 2016, for local team only
- Additional career development and training opportunities – *ad hoc*
- **CF@LANTA** RDP Predoctoral and Postdoctoral Fellowships – pages 1 & 2 of this newsletter

CF-Analytics Core (CFAC), Professor Facundo Fernández, PhD, Director

Dr. Fernández described the catalog of sub-cores that will provide consultation and services to CF investigators:

- Metabolomics – at Georgia Tech
- Proteomics – at Georgia Tech
- Immunoinflammation & Redox – at Emory

The capabilities being developed at Georgia Tech are within the brand new Systems Mass Spectrometry Center at Georgia Tech (which Dr. Fernández directs), while those at Emory are being developed by Drs. LouAnn Brown and Rabin Tirouvanziam. Dr. Fernández noted that the RDP grant does not pay for core services, in terms of per-sample costs, but does pay for consultation and supports a portion of the person leading the metabolomics sub-core. The new Core will work in concert with the existing Emory Integrated Proteomics Core. Each Core offers a set of basic assays that are common, and then a range of complementary assays. Special work that can be done at Georgia Tech includes the following: study of complex posttranslational modifications; top-down proteomics; mass spec imaging (proteins and lipids); and kinomics (study of kinases).

NACFC 2015 Recap

An insider's report on the 2015 North American Cystic Fibrosis Conference in Phoenix, Arizona, from Rabin Tirouvanziam, PhD, Assistant Professor at Emory University,



Rabin Tirouvanziam, PhD

Overview. This year's North American Cystic Fibrosis Conference (NACFC) was held in the sunny city of Phoenix, AZ, with a beautiful mountain range as backdrop. Many exciting topics were covered in plenary sessions, symposia, workshops and poster sessions over 3 full days. This year's plenary sessions were dedicated to personalized medicine (Clancy), clinical trials (Retsch-Bogart), and mental health (Quittner, Elborn, Smith). You can view the plenary sessions online at: <https://www.cff.org/2015-NACFC-Plenary-Videos-Available-Online/>. In keeping with my group's main research interests, I primarily attended symposia and workshop talks related to early CF disease.

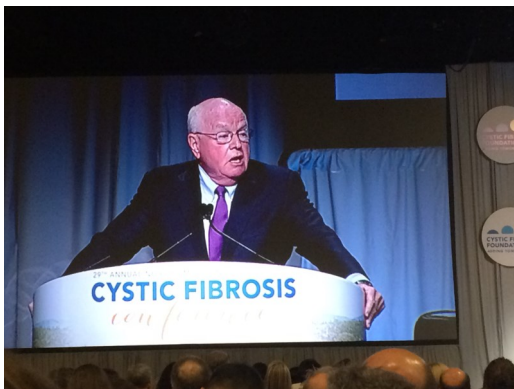
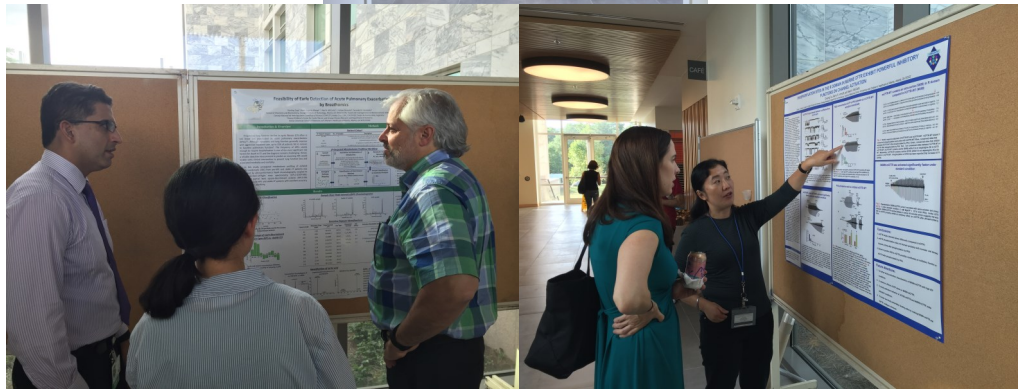
Multiple talks reported on findings from prospective cohorts of CF infants and children followed by structural imaging (by computed tomography -CT-, and magnetic resonance imaging -MRI-), lung function testing (by spirometry in older children, and lung clearance index -LCI- in infants and young children), as well as molecular assays including microbiome, mucus and metabolite analyses. **For MRI**, the main development includes better spatial resolution, close to that of CT (Rosenow). MRI data acquisition is longer than that for CT, however, and requires stillness, so it is not as practical for imaging early disease in infants unless they are sedated. **CT** has seen major improvements, using new programs (PRAGMA, Dutch-Australian collaboration) for detection of structural damage such as bronchial wall thickening, air trapping and bronchiectasis, with very low radiation levels (current CT protocols run in CF children every year for 10 years amount to same cumulative radiation as one half of one adult CT scan, or eating bananas every day for 50 years). **LCI**, which reflects gas flow in the airways, was highly variable and correlated poorly with structural CT scores in infants, but gained in stability and dynamic range (difference between mild and more severe disease) as children aged, showing promising data in school age children (Ratjen). There were questions with regards to the lack of LCI protocol standardization between centers, which still severely hampers its use for routine clinical practice.

From a biomarker standpoint, pilot data on bronchoalveolar lavage fluid (BALF) from the Australian AREST CF infant and children cohort was reported, detailing changes in mucus, microbiome and metabolites occurring in conjunction with disease progression (Esther). With increasing structural damage based on CT measures of bronchial wall thickening and bronchiectasis, BALF samples show changes in mucus (short strands and dominance of the MUC5AC species). Concomitant changes are seen in the BALF microbiome, with progressive switch from a picture dominated by oral flora to a picture including classical CF pathogens, such as *Haemophilus influenzae*, *Staphylococcus aureus* and *Pseudomonas aeruginosa*. Finally, a change in BALF metabolites is also seen with increasing structural lung damage, with a clear metabolic signature (including dipeptides and adenosine) linked to the presence of neutrophils in the airway lumen, which confirms the prominent role of these cells as drivers of disease progression. Interestingly, some of these metabolic markers from BALF were also measurable in exhaled breath condensate (EBC), which opens opportunities for less invasive and more sensitive biomarkers for early CF disease; this is a major unmet need for clinical monitoring and assessment of treatment effects in infants and children (Sagel).

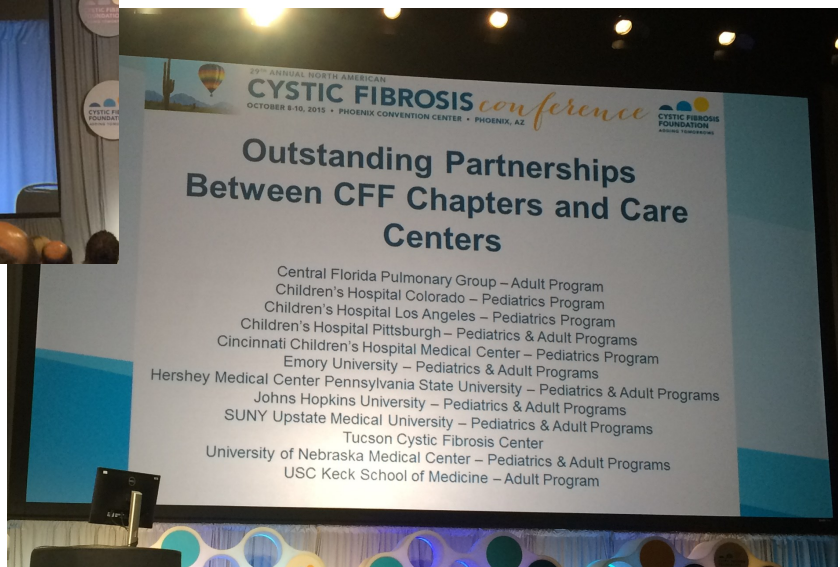
From a basic standpoint, the debate is still raging as to whether airway fluid anomalies (mucus hyperviscosity, abnormal pH) or neutrophilic inflammation occur first in developing CF airways after birth. Data from the CF knockout pig model (Gray) suggest that the CF epithelium primes the lung for neutrophilic inflammation via the abnormally high basal production of proinflammatory molecules S100A8/A9. These were known in the 70s and 80s as the CF antigen and believed by some to be the CF gene, before the gene for CFTR was cloned. The S100A8/A9 burden is further increased when neutrophils, the main known source for these molecules, invade the airways. Also in the CF pig, decreased airway surface liquid pH was measured at an early stage, which is believed to affect bactericidal function of antimicrobial peptides (Shah). An attempt was made to normalize pH by inhibiting the ATP12A transporter, resulting in increased bacterial killing. Interestingly however, researchers from the AREST CF program in Australia reported on their attempt to measure pH directly in CF infants using a pH probe lowered into the small airways via a bronchoscope (Sly). Their data failed to demonstrate any change in pH in CF vs. control infants, unlike what was reported in CF pigs, which emphasizes the important step of always carefully checking the validity of CF models against human disease.

Taken together, data reported at this year's NACFC improved our knowledge of molecular and structural anomalies at the inception of CF airway disease. More work remains to be done in patients and in models to further untangle events of early CF disease and improve on treatment options for infants and children.

NACFC Poster Preview in Atlanta and Conference in Phoenix, AZ



Above: Robert Beall, PhD says farewell as he concludes his 21 year tenure as President & CEO of the Cystic Fibrosis Foundation
 Right: The Emory/CHOA Pediatric & Adult CF Clinics were recognized for their outstanding work with the Georgia Chapter of the Cystic Fibrosis Foundation.



NACFC 2015 Abstracts

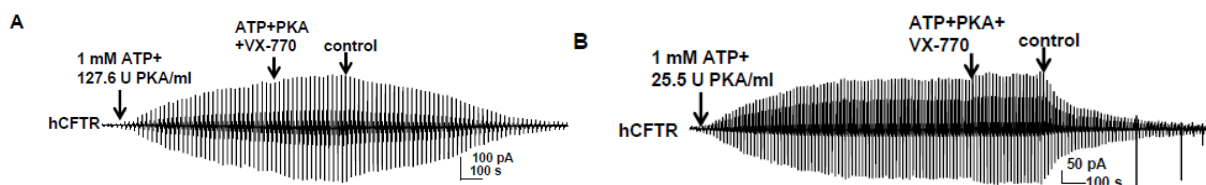
The 29th Annual North American Cystic Fibrosis Conference held on October 8-10, 2015 in Phoenix, Arizona included these abstracts from Atlanta-based CF researchers.

- **MOLECULAR DYNAMICS FLEXIBLE FITTING (MDFF) SIMULATIONS IDENTIFY MODELS OF CLOSE-STATE CFTR** Simchaev, L.¹; McCarty, N.A.²; Ford, B.³; Senderowitz, H.¹; 1. Chemistry, Bar-Ilan University, Ramat Gan, Israel; 2. Center for CF and Airways Disease Research, Emory University, Atlanta, GA, USA; 3. Life Sciences, University of Manchester, Manchester, United Kingdom
- **PHOSPHORYLATION SITES IN THE R DOMAIN IN MURINE CFTR EXHIBIT POWERFUL INHIBITORY FUNCTIONS ON CHANNEL ACTIVATION** Cui, G.; Imhoff, B.R.; McCarty, N.A. Emory University, Atlanta, GA, USA
- **COMPARATIVE MOLECULAR PHARMACOLOGY OF CFTR: TOWARD THE IDENTIFICATION OF NOVEL POTENTIATORS AND THEIR BINDING SITES** Cui, G.¹; Khazanov, N.²; Senderowitz, H.²; McCarty, N.A.¹; 1. Center for CF and Airways Disease Research, Emory Univ. and Children's Healthcare of Atlanta, Atlanta, GA, USA; 2. Bar-Ilan University, Ramat Gan, Israel
- **ATOMISTIC SIMULATIONS VALIDATING A RECENT CFTR HOMOLOGY MODEL** Stock, G.³; McCarty, N.A.¹; Gumbart, J.²; 1. Center for CF and Airways Disease Research, Emory Univ and Children's Healthcare of Atlanta, Atlanta, GA, USA; 2. Physics and Chemistry & Biochemistry, Georgia Institute of Technology, Atlanta, GA, USA; 3. Chemistry & Biochemistry, Georgia Institute of Technology, Atlanta, GA, USA
- **IDENTIFICATION OF SMALL METABOLITES ASSOCIATED WITH METASTABLE AND ACUTE CYSTIC FIBROSIS AIRWAY DISEASE BY HIGH-RESOLUTION METABOLOMICS** Ingersoll, S.¹; Bernardin, N.¹; Tangpricha, V.²; Alvarez, J.A.²; Ziegler, T.R.²; Jones, D.P.²; Tirouvanziam, R.¹; 1. Pediatrics, Emory University, Emory+Children's Center for CF and Airways Disease Research, Atlanta, GA, USA; 2. Medicine, Emory University, Atlanta, GA, USA
- **SPHINGOSINE 1-PHOSPHATE RECEPTOR 3 IS HIGHLY UPREGULATED ON HUMAN NEUTROPHILS UPON MIGRATION TO THE AIRWAYS** Ingersoll, S.^{1,2}; Forrest, O.A.^{2,3}; Brown, M.R.¹; Tirouvanziam, R.^{1,2}; 1. Department of Pediatrics, Emory University School of Medicine, Atlanta, GA, USA; 2. Center for Cystic Fibrosis and Airways Disease Research, Children's Healthcare of Atlanta, Atlanta, GA, USA; 3. Immunology and Molecular Pathogenesis Program, Emory University School of Medicine, Atlanta, GA, USA
- **INSULIN IMPAIRS AIRWAY BARRIER FUNCTION IN CYSTIC FIBROSIS RELATED DIABETES** Molina, S.A.^{1,2}; Moriarty, H.K.¹; Kim, A.H.¹; McCarty, N.A.^{3,2}; Koval, M.^{1,2}; 1. Dept. of Medicine, Emory Univ. School of Medicine, Atlanta, GA, USA; 2. Emory+Children's Center for Cystic Fibrosis and Airways Disease Research, Emory Univ. School of Medicine, Atlanta, GA, USA; 3. Dept. of Pediatrics, Emory Univ. School of Medicine, Atlanta, GA, USA
- **GENERATION OF A BRONCHIAL EPITHELIAL CELL LINE WITH DEFINED CFTR MUTATIONS BY NUCLEASE-ENHANCED SDF-MEDIATED HOMOLOGOUS REPLACEMENT FROM NON-CF I6HBE14O- CELLS** Suzuki, S.¹; Di Geronimo, R.¹; Tasto, C.¹; Guimaraes, A.³; Illek, B.²; Fischer, H.²; Cradick, T.J.⁵; Porteus, M.⁴; Muench, M.^{3,1}; Bao, G.⁵; Gruenert, D.C.^{1,6}; 1. University of California, San Francisco, San Francisco, CA, USA; 2. UCSF Benioff Children's Hospital Oakland, Oakland, CA, USA; 3. Blood Systems of the Pacific Research Institute, San Francisco, CA, USA; 4. Stanford University, Stanford, CA, USA; 5. Georgia Institute of Technology, Atlanta, GA, USA; 6. University of Vermont College of Medicine, Burlington, VT, USA
- **A NOVEL STATISTICAL ANALYSIS ON THE ASSOCIATION BETWEEN RECURRENT PSEUDOMONAS AERUGINOSA AND RECURRENT STAPHYLOCOCCUS AUREUS INFECTIONS IN YOUNG CHILDREN WITH CYSTIC FIBROSIS** Yang, J.¹; Peng, L.¹; Zhang, Z.²; Lai, H.J.²; 1. Biostatistics & Bioinformatics, Emory Univ, Atlanta, GA, USA; 2. Nutritional Sciences, Univ of Wisconsin, Madison, WI, USA
- **FEASIBILITY OF EARLY DETECTION OF ACUTE PULMONARY EXACERBATIONS BY BREATHOMICS** Zang, X.¹; Monge, M.E.^{1,2,3}; McCarty, N.A.^{4,5}; Stecenko, A.^{4,5}; Fernandez, F.¹; 1. School of Chemistry and Biochemistry, Georgia Institute of Technology, Atlanta, GA, USA; 2. National Scientific and Technical Research Council (CONICET), Buenos Aires, Argentina; 3. Center for Bionanoscience Research (CIBION), Buenos Aires, Argentina; 4. Emory+Children's Center for Cystic Fibrosis and Airways Disease Research and Department of Pediatrics, Atlanta, GA, USA; 5. School of Medicine and Children's Healthcare of Atlanta, Emory University, Atlanta, GA, USA
- **ACUTE PULMONARY EXACERBATIONS AT A LARGE CF CENTER** Maliniak, M.L.¹; Salinger, M.²; Stecenko, A.¹; Walker, S.D.³; 1. Pediatrics, Emory University, Atlanta, GA, USA; 2. Emory University School of Medicine, Atlanta, GA, USA; 3. Medicine, Emory University, Atlanta, GA, USA
- **BUILDING A CASE FOR INCREASED CF CENTER STAFF** Walker, S.D.; Martin, M.; Hunt, W.R. Medicine, Emory University, Atlanta, GA, USA
- **VEHICLES FOR THE ABSORPTION OF VITAMIN D IN CYSTIC FIBROSIS: POWDER VS OIL** Hermes, W.A.; Alvarez, J.A.; Lee, M.J.; Tangpricha, V. Division of Endocrinology School of Medicine, Emory University, Atlanta, GA, USA
- **DIETARY AND INFLAMMATION PROFILES OF INDIVIDUALS WITH CYSTIC FIBROSIS BY BODY COMPOSITION STATUS** Alvarez, J.A.^{1,2}; Millson, E.C.³; Ziegler, T.R.^{1,2,3}; Stecenko, A.^{2,4}; 1. Endocrinology, Diabetes, and Metabolism, Emory University School of Medicine, Atlanta, GA, USA; 2. Center for Cystic Fibrosis and Airways Disease Research, Children's Healthcare of Atlanta, Atlanta, GA, USA; 3. Clinical Research Network, Atlanta Clinical and Translational Science Institute, Atlanta, GA, USA; 4. Div. of Pulmonology, Allergy/Immunology, Cystic Fibrosis and Sleep, Dept. of Pediatrics, Emory Univ. School of Medicine, Atlanta, GA, USA

Recent CF-AIR Publications

Recently publications since June 2015. If you have a publication you would like in the next newsletter, please contact CF-AIR program coordinator, Karen Kennedy, kmurra5@emory.edu.

- Boopathy AV, Martinez MD, Smith AW, Brown ME, García AJ, Davis ME. Intramyocardial Delivery of Notch Ligand-Containing Hydrogels Improves Cardiac Function and Angiogenesis Following Infarction. *Tissue Eng Part A*. 2015 Sep;21(17-18):2315-22. doi: 10.1089/ten.TEA.2014.0622. Epub 2015 Jul 13. PubMed PMID: 25982380.
- Carmean A, Fortenberry JD, McCracken C, Hebbar KB. A Survey of Attitudes and Practices Regarding the Use of Steroid Supplementation in Pediatric Sepsis. *Pediatr Emerg Care*. 2015 Oct;31(10):694-8. doi: 10.1097/PEC.0000000000000561. PMID: 26414641.
- Cui G, McCarty NA. Murine and human CFTR exhibit different sensitivities to CFTR potentiators. *Am J Physiol Lung Cell Mol Physiol*. 2015 Jul 24;ajplung.00181.2015. doi: 10.1152/ajplung.00181.2015. [Epub ahead of print] PubMed PMID: 26209275.
- Farkas AE, Hilgarth RS, Capaldo CT, Gerner-Smidt C, Powell DR, Vertino PM, Koval M, Parkos CA, Nusrat A. HNF4 α Regulates Claudin-7 Protein Expression during Intestinal Epithelial Differentiation. *Am J Pathol*. 2015 Aug;185(8):2206-18. doi: 10.1016/j.ajpath.2015.04.023. PubMed PMID: 26216285; PubMed Central PMCID: PMC4530128.
- Koval M. The expanding diversity of roles for claudins. *Semin Cell Dev Biol*. 2015 Jun;42:1-2. doi: 10.1016/j.semcdb.2015.07.005. PubMed PMID: 26343821.
- Lee MJ, Alvarez JA, Smith EM, Killilea DW, Chmiel JF, Joseph PM, Grossmann RE, Gaggar A, Ziegler TR, Tangpricha V; Vitamin D for Enhancing the Immune System in Cystic Fibrosis (DISC) Investigators. Changes in Mineral Micronutrient Status During and After Pulmonary Exacerbation in Adults With Cystic Fibrosis. *Nutr Clin Pract*. 2015 Jun 15. pii: 0884533615589991. [Epub ahead of print] PubMed PMID: 26078287.
- Molina SA, Stauffer B, Moriarty HK, Kim AH, McCarty NA, Koval M. Junctional abnormalities in human airway epithelial cells expressing F508del CFTR. *Am J Physiol Lung Cell Mol Physiol*. 2015 Sep 1;309(5):L475-87. doi: 10.1152/ajplung.00060.2015. Epub 2015 Jun 26. PubMed PMID: 26115671; PubMed Central PMCID: PMC4556929.
- Moustafa DA, Scarff JM, Garcia PP, Cassidy SK, DiGiandomenico A, Waag DM, Inzana TJ, Goldberg JB. Recombinant Salmonella Expressing Burkholderia mallei LPS O Antigen Provides Protection in a Murine Model of Melioidosis and Glanders. *PLoS One*. 2015 Jul 6;10(7):e0132032. doi: 10.1371/journal.pone.0132032. eCollection 2015. PubMed PMID: 26148026; PubMed Central PMCID: PMC4492786.
- Obiany O, Liang Y, Burnham EL, Mehta A, Park Y, Uppal K, Harris FL, Jones DP, Brown LA. Metabolic Consequences of Chronic Alcohol Abuse in Non-Smokers: A Pilot Study. *PLoS One*. 2015 Jun 23;10(6):e0129570. doi: 10.1371/journal.pone.0129570. eCollection 2015. PubMed PMID: 26102199; PubMed Central PMCID: PMC4477879.
- Plyler ZE, Hill AE, McAtee CW, Cui X, Moseley LA, Sorscher EJ. SNP Formation Bias in the Murine Genome Provides Evidence for Parallel Evolution. *Genome Biol Evol*. 2015 Aug 6;7(9):2506-19. doi: 10.1093/gbe/evv150. PubMed PMID: 26253317.
- Schadzek P, Schlingmann B, Schaarschmidt F, Lindner J, Koval M, Heisterkamp A, Preller M, Ngezahayo A. The cataract related mutation N188T in human connexin46 (hCx46) revealed a critical role for residue N188 in the docking process of gap junction channels. *Biochim Biophys Acta*. 2015 Oct 6. pii: S0005-2736(15)00329-6. doi: 10.1016/j.bbamem.2015.10.001. [Epub ahead of print] PMID: 26449341.
- Simek RZ, Prince J, Syed S, Sauer CG, Martineau B, Hofmekler T, Freeman AJ, Kumar A, McElhanon BO, Schoen BT, Tenjarla G, McCracken C, Ziegler TR, Tangpricha V, Kugathasan S. Pilot Study Evaluating Efficacy of two Regimens for Hypovitaminosis D Repletion in Pediatric Inflammatory Bowel Disease. *J Pediatr Gastroenterol Nutr*. 2015 Jul 20. [Epub ahead of print] PubMed PMID: 26196201.
- Tukvadze N, Sanikidze E, Kipiani M, Hebbar G, Easley KA, Shenvi N, Kempker RR, Frediani JK, Mirtskhulava V, Alvarez JA, Lomtadze N, Vashakidze L, Hao L, Del Rio C, Tangpricha V, Blumberg HM, Ziegler TR. High-dose vitamin D3 in adults with pulmonary tuberculosis: a double-blind randomized controlled trial. *Am J Clin Nutr*. 2015 Sep 23. pii: ajcn113886. [Epub ahead of print] PMID: 26399865.



Cui & McCarty
Am J Physiol Lung Cell Mol Physiol 2015
Figure 2 a&b.
See article for full details.

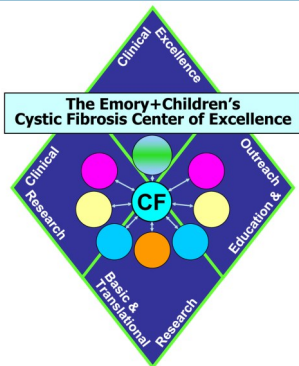
Events for Researchers

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/centers/sub-pages/cf-air-seminars-workshops
and
www.pedsresearch.org/centers/detail/cf-air-cf-education-outreach-cf-scholars-program



Center Brags

There is so much to brag about in the center, here are some recent highlights:

- Welcome to new members Eric Sorscher, MD; Annette Ehrhardt, PhD; Devon Greene, MD; Rachel Linnemann, MD; Tim Beaty, MD; Sam Brown, PhD (GT); Lokesh Guglani, MD
- Mike Koval, PhD was promoted to the rank of Professor
- Eric Sorscher, MD is being inducted in to the American Clinical and Climatological Association.
- Welcome to new students: Bijean Ford of the Immunology & Molecular Pathogenesis Program in the Tirouvanziam lab and Sabrina Lynn of the Biochemistry, Cell and Developmental Biology Program in the Koval lab.
- Claudia Morris, MD garnered press for her paper on the results of the Thalassemia Clinical Research Network, http://news.emory.edu/stories/2015/07/faulty_arginine_metabolism_cardiopulmonary_dysfunction/index.html.
- Sarah Ingersoll, PhD concluded her postdoc in the Tirouvanziam lab, accepting a position at Amgen to study COPD and asthma.
- Sam Molina's abstract "Alpha-type Cx43 function is restored with 4-PBA treatment in Cystic Fibrosis airway epithelial cells that express misfolded F508del-CFTR protein" was selected for oral presentation at the American Society of Cell Biology Annual Meeting, San Diego, December, 2015.
- Eric Sorscher, MD chaired the International CFTR Folding Consortium before the NACFC.
- Rabin Tirouvanziam, PhD was awarded his first R01 as PI, "Contribution of neutrophils to early airway disease in cystic fibrosis children."
- Mike Koval served as Discussion Leader, Gordon Research Seminar (GRS) on Lung Development, Injury & Repair: From Basic Science to Translational Research, Proctor Academy, Andover, NH, August 15-16, 2015.
- Dominique Limoli, PhD, a postdoctoral fellow in the Goldberg lab, visited George O'Toole's lab at Dartmouth College in Hanover, NH to investigate *Pseudomonas aeruginosa* and *Staphylococcus aureus* interactions during biofilm coinfection of bronchial epithelial cells from cystic fibrosis patients. This was supported in part by the Emory+Children's CF Center of Excellence Director's Fund.
- Eric Sorscher, MD will be a visiting professor at the Hebrew University in Hadassah in November.
- Eric Hunter was promoted to Clinical Research Coordinator IV and is managing the CF clinical research coordinator team.
- Troy Kleber, a Georgia Tech BME undergrad working in the McCarty lab, has been accepted into the Petit Undergraduate Research Scholars Program, a competitive scholarship program that serves to develop the next generation of leading bioengineering and bioscience researchers.

Staying in Touch

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/centers/detail/CF-AIR

If you are interested in supporting our research and outreach programs please visit:

www.pedsresearch.org/centers/sub-pages/cf-air-donors-visitors/

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