

Cystic Fibrosis Center of Excellence

Emory University - Children's Healthcare of Atlanta - Georgia Tech

Volume 12, Issue 1

September, 2023

Inside this issue:

NIH funding to CF@LANTA	2
Welcome New Faculty Members	3,4
Adult Program Celebrates 20 years!	5
Corporate Partnership	5
David McKenney—Family, Devotion, and Dedication	6,7
Impact of Philanthropy—Genetic Counseling	8
New Grant for Dr. Chandler	9
Dr. Jones Joins as Associate Director for CF@LANTA	9
Stay in Touch	10

Dear friends and supporters of CF@LANTA,

We are writing with a lot of great news, hoping this finds you well. There is much to tell you, with so many great things having happened since we last wrote.

CF@LANTA has continued to grow and reach great new heights. In this newsletter, you will hear about new faculty, new grants, new leadership in both CF@LANTA and CF-AIR, new accolades for our Center. On page 2, we tell you about the new very large NIH grant that our Center has been awarded. On pages 3 and 4, we introduce you to two new faculty: Drs. Kathryn Oliver and Ben Kopp. On page 5, we reflect on the 20th anniversary of the launch of Emory's Adult CF Clinical Program. Also on page 5, we recognize a long-standing corporate partnership that has brought visibility and support to CF@LANTA for over a decade. On page 6-7, we introduce you to Mr. David McKenney, a member of CF@LANTA's Community Advisory Board. On page 8, we tell you how a philanthropic gift from the Patrick Flynn Memorial Foundation has supported the work of our star Genetic Counselor, Ms. Eileen Barr. On page 9, we tell you about a new NIH grant to the lab of Dr. Joshua Chandler, and introduce CF@LANTA's new Associate Director, Dr. Kymry Jones.

Our team received two great awards at the 2022 North American CF Conference, held in Philadelphia, PA, Nov 3-6, 2022. (1) The Children's+Emory CF Care Center was selected to receive the CF Foundation's 2022 Outstanding Partnership Award! (2) Our own Ms. Shaina Blair, Mental Health Coordinator for the Center, received the Carolyn and C. Richard Mattingly Leadership in Mental Health Care Award from the CF Foundation.



We are so blessed to have Shaina as a key member of our clinical team!

Dr. Nael McCarty, Director, **CF@LANTA**

Dr. Guglani Named Associate Director for the CF Biospecimen Repository (CF-BR)



Lokesh Guglani, MD, Associate Professor of Pediatrics, is a CF clinician researcher in the Emory Pediatric CF Program and leads the Primary Ciliary Dyskinesia center at Children's Healthcare of Atlanta. He will help lead the CF-BR, a key component of our CF research infrastructure. His main areas of research focus are related to various aspects of CF, ranging from advanced lung function techniques such as Lung Clearance Index, to imaging (using scoring techniques to quantify regional lung damage), to CF diagnostics (novel medical device for sweat testing), to translational studies related to lung inflammation in early stages of CF lung disease (IMPEDE-CF Study). He has been involved in mentoring several pulmonary fellows and pediatric residents for their research and has authored more than 70 research publications and book chapters. In his spare time, he loves to pursue 3D printing, running, traveling, and going on long hikes with his wife and two teenaged kids.



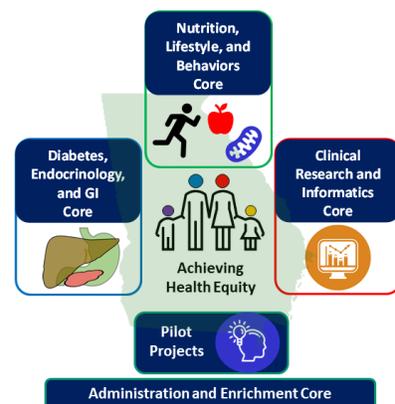
NIH P30 Grant — Renewed for Five Years

In September, 2020, the NIH awarded our Center a large research grant, called a “P30” mechanism, that launched the Georgia CF Research and Translation Core Center (a.k.a., the Georgia CF Core Center). We were one of only eight CF centers funded by the P30 mechanism across the country – putting us into a very select group. The 2020 grant was funded for only three years, and at a somewhat reduced budget, meaning that we would be required to submit a renewal application in July 2022, less than 2 years after the initial grant was launched. Of course, during that time, we also were fighting back a global pandemic, which made it a real challenge to get this new effort up and running in time to make a big impression in the renewal application. Thanks to a massive effort from several team members, which started only 14 months after the initial grant was awarded, we put together an excellent renewal application which the NIH funded – this time, for all five years, and for full budget!

Like its predecessor, the renewal for the Georgia CF Core Center includes CF researchers all across Georgia, at premier institutions including Emory, Children’s, Georgia Tech, Augusta University, the University of Georgia, and Georgia State University. The Center includes three biomedical research Cores, as shown in the adjacent figure, plus a pilot grant mechanism, an enrichment program, and an administrative core. The research supported by this grant focuses on the non-pulmonary aspects of CF.

The P30 grant renewal represents the largest single CF research grant awarded to our Georgia institutions in history. We are grateful to the NIH for this funding, and very much look forward to the advances in care that will come from the research that this grant enables.

The Georgia Cystic Fibrosis Core Center



National Institutes of Health Ranks Pediatrics Department #1, Again! Message from Dr. Lucky Jain, Chair, Dept. of Pediatrics

Dear Friends,

Many of you may have already heard that the Blue Ridge Institute for Medical Research has ranked Emory University’s Department of Pediatrics (DOP), the primary academic partner of Children’s Healthcare of Atlanta, No. 1 in the country for federal research dollars from the National Institutes of Health (NIH) in 2022 for pediatrics departments. Congratulations to all! This is an honor and reflection of the hard work you put into research and helping to advance medical discoveries every day. The Emory Department of Pediatrics received more than \$77 million in NIH funds last year... and extramural funding for the DOP totaled more than \$111 million. The new figures show the pediatrics department ranking #1 again, for the second time in three years! The program has maintained a top five ranking since 2016, up from #49 in 2004.

I hope you know how proud I am of our department and I want to express my deepest gratitude to all of you for your commitment to pediatric health research.

Best wishes to all!

Lucky Jain, MD, MBA

Pediatrician in Chief, Children’s Healthcare of Atlanta

George W. Brumley Jr. Professor and Chair

Emory University School of Medicine, Department of Pediatrics

Executive Director, Emory+Children’s Pediatric Institute

Welcome Kathryn Oliver, PhD — New Faculty Member

Please join us in welcoming Kathryn Oliver, PhD, to CF@LANTA as Assistant Professor in the PACS Division of the Department of Pediatrics, and a member of the CF-AIR Center!

Dr. Oliver has pursued studies of cystic fibrosis pathogenesis since 2009. She began her scientific career as a master's student at Auburn University, where she characterized adaptive metabolic strategies by which *Pseudomonas aeruginosa* chronically establishes itself in the lungs of individuals with CF. After graduating with an M.S. in Microbiology, she worked one year as Lead Instructor for the "Genetics and Evolution" course at the University of Alabama in Huntsville.

In 2012, Dr. Oliver enrolled in the Genetics graduate program at the University of Alabama at Birmingham to study CF disease mechanisms. Her dissertation project utilized high-throughput yeast phenomics to identify novel genetic modifiers of the most common CF-causing variant (F508del-CFTR) and elucidated, for the first time, effects of ribosomal perturbation on CFTR synthesis, trafficking, and function. Dr. Oliver began postdoctoral training at Emory University in 2017 and continued to work towards a career in basic and translational science. Her studies concentrated on evaluating changes in translation kinetics – both in vitro and in vivo – following suppression of specific ribosomal proteins.

In September, 2021, Dr. Oliver was promoted to Assistant Professor of Pediatrics at Emory. Her current research program employs interdisciplinary approaches in molecular genetics, RNA biology, biochemistry, cellular physiology, and functional genomics to investigate the impact of translational speed, ribosome fidelity, and mRNA surveillance on protein synthesis in the context of CF. Particular emphasis is placed on premature termination codons (PTCs) and other rare defects that occur in the CFTR gene, specifically evaluating underlying mechanism(s) responsible for genotype-phenotype correlation, pharmacologic responsiveness, and heterogeneity of patient outcomes. Dr. Oliver's scientific objectives also include better delineation

of genetic factors that influence severity of refractory CFTR variants, including attention to mutations that occur in Black, Indigenous, or People of Color (BIPOC) with the disease. Her overarching goals are to provide new insights relevant to CF clinical intervention and to extend research findings to other pediatric conditions with similar etiology. Dr. Oliver presently serves on the national CFF Guidelines Committee for CF Related Metabolic Disorder (CRMS) and CF Screen Positive Inconclusive Disorder (CFSPID), whose mission is to develop consensus recommendations for CRMS/CFSPID genetic testing, diagnosis, and treatment.

ORCID: <https://orcid.org/0000-0002-7781-7006>

LinkedIn: <https://www.linkedin.com/in/oliverke/>

Dr. Oliver is a highly accomplished basic/translational researcher, and we are convinced that she will be a very successful independent scientist. The Oliver lab is located in the Emory-Children's Center building. We look forward to her team adding to the vibrant research environment on that wing and in our Center and Division.

Please join us in welcoming Dr. Oliver to the CF@LANTA faculty!



Introducing: Benjamin Kopp, MD, MPH — New Faculty Member

Please join us in welcoming Ben Kopp, MD, to CF@LANTA as Associate Professor in the PACS Division of the Department of Pediatrics, and Co-Director of CF-AIR.

Dr. Ben Kopp recently (Aug 2022) joined the PACS Division and CF@LANTA as a physician-scientist and Co-Director of the CF-AIR research center. He also leads the clinical Pulmonary Sickle Cell Program.

He received his MD and MPH degrees from The Ohio State University and was faculty at OSU and Nationwide Children's Hospital from 2012 – 2022. At Nationwide he directed the CF Immune Core as part of a CFF-funded Research Development Program grant. His overall approach is to blend clinical, research, and advocacy efforts to holistically improve patient care. A major focus of this work has been on translational research models utilizing human-derived specimens. Specifically, his lab seeks to understand how innate immunity contributes to the pathogenesis of chronic lung diseases. His lab is focused on host-pathogen-environment interactions that impact respiratory diseases such as cystic fibrosis (CF) and lung disease related to sickle cell disease.

One of his main CF projects is focused on understanding the pathophysiologic mechanisms that lead to defective macrophage phagocytosis, how these mechanisms can be altered by CFTR modulation, and the relation of resulting macrophage function to clinical outcomes. He hopes that this, combined with microbiome and deep sequencing data, can lead to better 'immunophenotyping' of individuals to better contextualize their cellular disease. Related to understanding how CFTR regulates innate immune function, a second project his lab is studying how related ion channels such as ENaC could be CFTR targeted in CFTR-agnostic approaches to improving macrophage function. Similar to the airway epithelium, ENaC is over-expressed in CF immune cells and regulates certain aspects of macrophage function. Finally, his lab is very interested in environmental influences on cellular consequences of CF. These include indoor air pollutants such as tobacco or e-cigarette exposure as well as an emerging interest in environmental health inequities and how these influence overall health.

Outside of work, Dr. Kopp enjoys running and outdoor activities with his wife and 2 children (15 & 12) who are avid soccer players. He loves sports and is looking forward to exploring all the great parks and events in Georgia!

Please join us in welcoming Dr. Kopp to the CF@LANTA faculty!



Benjamin Kopp, MD, MPH
Associate Professor of Pediatrics
Emory University School of Medicine

We note with gratitude the many and outstanding contributions that Dr. Arlene Stecenko has made as Co-Director of the CF-AIR research center, since its inception in 2010.

With Dr. Kopp joining the CF@LANTA faculty, Dr. Stecenko has decided to focus both on her own research projects and on serving as Co-Director for the Georgia CF Core Center and Director of the Clinical Research & Informatics Core within the P30.

Thanks, Dr. Stecenko!



Emory Adult CF Clinical Program Celebrates 20 Years

Twenty years ago, the Cystic Fibrosis Center formally launched an adult-specific CF clinical program which, along with our pediatric CF clinical program, comprises the Children's+Emory CF Care Center. Most children transition from the pediatric program to the adult program by age 21. Until just the past couple of years, CF was considered a "pediatric disease" since most patients did not live into late adulthood. However, due to the transformative advanced treatments recently made available, the nation now has more adults with CF than children!

Twenty years ago, Dr. Arlene Stecenko (the Marcus Professor of Pulmonology and Associate Professor of Pediatrics), worked with Emory Healthcare to establish our adult clinical program. The following video celebrates the anniversary of this excellent partnership between Emory and Children's, on behalf of our CF patients. Dr. Stecenko is interviewed by Dr. Randy Hunt, Director of the Emory Adult CF Clinical Program.

[Emory - CF Center 20th Anniversary Video](#)



Shout-out to a Corporate Partner

For over a decade, CF@LANTA has had a great relationship with the Wealth Management team from Merrill Lynch. The team provides retirement planning classes, which financially support the Center – funds from all "tuition" charges for the classes come to our philanthropy accounts. The classes are led by an outstanding group, including Mr. Ernest "Skip" Guilliams (Senior Vice President and Wealth Management Advisor) and Mr. Adam Sitz (First Vice President and Senior Financial Advisor, who also serves on CF@LANTA's Community Advisory Board). Guilliams and Sitz invite CF@LANTA Director, Dr. Nael McCarty, to meet with the class attendees to share the Center's story, which has led to some good connections.

The Merrill Lynch team recently shared that they had been recognized on the 2023 Forbes "Best-in-State Wealth Management Teams" list, published January 12, 2023. Their announcement stated:

Our mission has always been simple – to bring simplicity and transparency to the complexities of investing. We are committed to helping clients pursue their personal goals – from planning for and funding their children's educations to helping ensure that their money lasts in retirement or even managing the transfer of wealth to the next generation.

We are very happy for the team in this important accomplishment, and are grateful for our continued partnership. If interested, the team can be reached [here](#).



Community Advisory Board Member David McKenney—Family, Devotion, and Dedication

David McKenney is a second-generation native of the Atlanta area. He attended high school at Georgia Military Academy in College Park (now Woodward Academy), where he earned appointments to national service academies, as well as a Naval ROTC full scholarship to Georgia Tech. His experiences as an Eagle Scout and high school ROTC led him into military service, where he spent three years as a Marine Corps officer, serving at Quantico, Virginia, and primarily at Camp Pendleton, California. During those three years he decided to become an engineer rather than follow his degree in Physics, so he returned to Georgia Tech to earn a degree in Industrial Engineering in one year.



Much like his college years when David was very engaged in extracurricular activities, David has been actively engaged with Georgia Tech (Alumni Association president in 2000, member of the GT Foundation board of trustees, fraternity chapter advisor); Young Presidents Organization; active member and president of his three professional and trade associations; co-founding board member, chairman, and now emeritus board member of Bobby Dodd Institute; board member of Georgia Community Trust; member of the downtown Atlanta Rotary Club; board member of The University Financing Foundation; founding member and four-time elder and finance committee member of his church; advisory board member of start-up technology firm iFolio; investor/supporter of non-profits The Warrior Alliance and HINRI (High Impact Network of Responsible Innovators/Healthcare Institution for National Renewal and Innovation); parent and supporter of Jacob's Ladder Neurodevelopmental Learning Center; and community advisory board member of CF@LANTA.

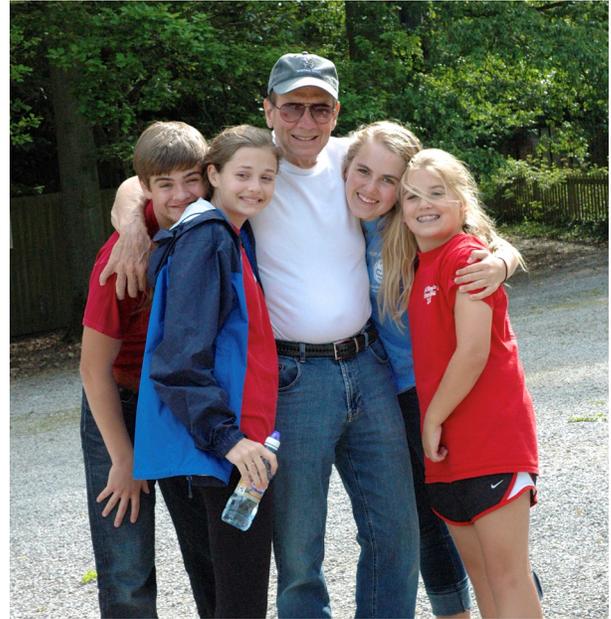


After graduation, David married Sarah, a second generation native of Pasadena, California, and joined The Trane Company as a sales engineer, where he enjoyed eight successful years while starting a family, becoming a registered Professional Engineer, and earning an MBA at Georgia State University. In 1972 he joined his family's business, McKenney's, Inc., and became its president and CEO in 1973. In 1972 the company was a \$4-5 million mechanical contractor with about 100 employees. When he retired at the end of 2009 its revenue was \$200 million, with 800 employees. His son John now operates the business and has doubled its size and scope.



David McKenney—Family, Devotion, and Dedication

In 2004 David created a family foundation to encourage and engage his children and grandchildren in supporting worthy charitable institutions. Each year after receiving annual donations from the three families, the directors meet near the end of the year to discuss the foundation's mission and specific charitable organizations for that year's donations. As each of his four grandchildren has become a teenager, they are included in the annual meeting, where they are encouraged to actively participate. One of the primary charitable interests of the family and the foundation is the health and well-being of children, which has resulted in annual gifts to Children's Healthcare of Atlanta, recently earmarked in part for CF@LANTA. David's connection there was through his church, where Nael McCarty is also a member and at one time held an appointment with the School of Biology at Georgia Tech.



Since becoming involved with Nael in the early days of CF@LANTA, an amazing amount of progress in support, staffing, research, groundbreaking results, and reputation has been made through the joint efforts of Emory University School of Medicine, Children's Healthcare of Atlanta, and Georgia Tech. David hopes to see the time when this disease can be prevented before birth, and successfully treated to further extend the lives of people with this disease.



Genetic Counselor — Impact of Philanthropy

Support for our work at CF@LANTA comes from a variety of sources, including institutional support from Children's and Emory, external grants, and gifts from donors. We use these gifts to support activities that often fall through the cracks of our institutional budgets. A great example is our Genetic Counseling Services, supported in part by a philanthropic gift from the **Patrick Flynn Memorial Foundation**. In this article, we outline the incredible impact that support from the Foundation has had, so far, through the work of the wonderful Ms. Eileen Barr, our Licensed Genetic Counselor.

First, here's an overview of our CF Genetic Counseling Services:

- Provide genetic counseling at both the pediatric and adult CF clinics
- Increase accessibility to genetic testing
- Provide counseling in cases of newborns diagnosed with CF or CFTR-related metabolic syndrome (CRMS), and CF carriers
- Provide guidance to families interesting in genetic counseling prior to conceiving
- Counseling for late diagnoses in adults and adolescents
- Review of all unknown/missing genetic results for pediatric and adult patients in the CFF patient registry to ensure that correct information is available for continuity of care

Over the past two years, this funding enabled Ms. Barr to launch the following initiatives.

1. Established a CRMS clinic: worked with nurse practitioners (NPs) and a fellow to review all patients with CFTR-related metabolic syndrome (CRMS) diagnosis at Children's; created a work-flow and management guideline based on provider survey and recent literature to ensure consistent care across providers; developed a CRMS clinic for annual follow-up, launched in Oct 2022; and continue to collaborate with providers and NPs for improved work-flow for establishing CRMS diagnosis and subsequent management.
2. Established Prenatal Genetic Counseling for parents of a baby with confirmed CF diagnosis: Ms. Barr and NPs work together to see couples who have received a prenatal diagnosis of CF a few weeks prior to delivery, which allows the couple to meet the team and see the clinic prior to delivery, and also helps with comfort and transition for the family.
3. Pregnancy and Trikafta: Recent literature identified that for some babies born to mothers with CF, Trikafta can impact the sensitivity of newborn screening, causing the babies with CF to be missed on newborn screening. Ms. Barr and a NP established a care pathway for newborns born to mothers with CF to coordinate genetic testing shortly after birth to ensure cases are not missed.

Ms. Barr was a co-author on four abstracts presented at the North American CF Conferences 2020-2022. She also published the following article: Hutchins K, **Barr E**, Bellcross C, Ali N, Hunt WR (2022) Evaluating Differences in the Disease Experiences of Minority Adults with Cystic Fibrosis. *Journal of Patient Experience* 4:23743735221112629.

Ms. Barr also achieved the following accolades: Winner of the Outstanding Friend in Care Award, 2022, GA CFF GA Chapter; CFF Peachtree Society Inductee, 2022; Emory Healthcare Physician Group Practice Service Hero Award, 2021; member of GA CF Newborn Screening Subcommittee, 2021-present.

We are very proud of Ms. Barr, and are grateful for all that she does for our patients and families. We also appreciate the donors that enable Ms. Barr to provide such great service!

Eileen Barr, MS, Licensed Genetic Counselor



Exciting New NIH Grant for Dr. Joshua Chandler

Lung inflammation typified by high neutrophil counts is a serious complication of CF that begins in infancy and progressively contributes to pulmonary failure. Although highly effective modulator drugs targeting the CFTR protein are available and have conferred significant lung function gains in some patients, an increasing body of evidence suggests that modulators are failing to treat the inflammatory complications of CF, especially after pulmonary disease has been established. Additionally, other pulmonary diseases that lack such highly effective therapies, such as primary ciliary dyskinesia (PCD), exhibit similar modalities of neutrophil-dominated lung inflammation and progressive lung structural damage. Thus, it is imperative that research is conducted to better understand how inflammation contributes to lung disease, so that better monitoring and interventions can be developed.

One such program is currently ongoing at Emory: The Chandler Laboratory was awarded an R01 grant from the National Heart, Lung, and Blood Institute of the National Institutes of Health, entitled “*Neutrophil hyperexocytosis and hypochlorous acid exposure in early cystic fibrosis lung disease.*” This study is designed to test the hypothesis that the cellular process of neutrophils called exocytosis is primarily responsible for releasing high levels of the enzyme myeloperoxidase (MPO) into the CF airway lumen as a direct consequence of pathological changes to neutrophils (a form of white blood cells) upon entering CF airways. Additionally, the proposal will test whether released MPO associates with extracellular vesicles from neutrophils that may enhance its activity. Finally, the proposal will test the impact of MPO activity on lung cell metabolism and viability and whether anti-MPO interventions block these effects.

Each of these aims uses state of the art methods including high resolution metabolomics, a signature technique of the Chandler Lab, to study redox biochemistry and metabolism critical to the underlying causes of CF lung damage. Additionally, the neutrophil transmigration model developed by the Lab of Dr. Tirouvanziam (also in CF@LANTA) has proven to be a critical technique in the success of this project.

This research is expected to yield new biomarkers and potential drug targets to better treat and monitor inflammation in CF. Indeed, early results indicating a novel pathway relevant to lung damage already have been validated in a cohort of CF toddlers. Furthermore, these same cellular and molecular processes are likely active in related diseases, including PCD, so that knowledge gained from this proposal may suggest monitoring and therapeutic strategies that will benefit patients well beyond CF. For questions or more information, please send an email to Dr. Chandler at joshua.chandler@emory.edu.



Joshua Chandler, PhD
Assistant Professor of Pediatrics
Emory University School of Medicine

Dr. Kymry Jones Joins CF@LANTA as Associate Director

We also are pleased to announce the recruitment of Dr. Kymry Jones as CF@LANTA’s Associate Director.



Dr. Jones earned her PhD in Molecular and Cell Biology at Georgia Tech, where she completed her studies in Dr. Nael McCarty’s lab in 2007. She then pursued postdoctoral training in inflammation at the Norwegian University of Science & Technology in Norway, and then in Biochemistry and Molecular Pharmacology, at NYU Langone Medical Center in New York, where she became Research Assistant Professor. She then became manager of the Neuropathology Core of the Alzheimer’s Disease Research Center at the Univ. of Southern California in Los Angeles. Dr. Jones will manage all of the Center’s research cores, oversee communications, serve as the co-chair of the Research Oversight Committee, develop infrastructure for high complexity clinical trials for CF, and coordinate activities with philanthropic and finance teams for CF@LANTA and CF-AIR. Welcome to the team, Kymry!

Events for Researchers

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

[Full CF-AIR Calendar of Events](#)

- [CF-AIR Research Workshop](#):
Meets every Thursday at 1:00 pm, hybrid format. Research-in-progress and journal club presentations.
- [CF Scholars Meetings](#):
A monthly program for CF Scholars, Friday afternoons; see website schedule.

Interested in more information? Send an email to Dr. McCarty (namccar@emory.edu) or to our Associate Director, Dr. Kymry Jones (ktjone8@emory.edu)

Clinics:

Pediatric Clinics at the CAP
1400 Tullie Road NE
Atlanta, GA 30329
404-785-KIDS

Emory Adult CF Clinic
1605 Chantilly Dr. NE
Atlanta, GA 30324
404-778-7929

Website:

www.cfatl.org

If you are interested in supporting our research and outreach programs please visit: www.pedsresearch.org/research/centers/cf-air/donors-visitors/

Contact:

Nael McCarty, PhD
Director
namccar@emory.edu

