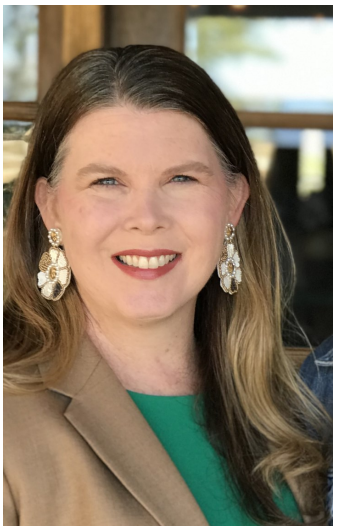
CF FAMILY NEWSLETTER

Fall 2022

UNC HEALTH Children's

New CF Education Committee Member: Jennifer Leandro, RN



Jennifer Leandro, one of our phenomenal CF nurses here at UNC, was recently selected from numerous well-qualified applicants to serve as a member of the Cystic Fibrosis Foundation Education Committee. The CF Education Committee is a group of pediatric and adult specialists in a multidisciplinary group (e.g., an adult with CF, a CF parent, Pulmonologists, Pediatric RN, Psychologist, Registered Dietician, Social Worker, Respiratory Therapist, Physical Therapist, Pharmacist, and Geneticist). Each member serves a 3 year term with a competitive process based on years of experience, innovation and direct patient impact. Once the application is submitted, it is voted on by the current team members. The committee meets monthly and/or quarterly, depending on the deadlines of the authors and volume being reviewed. The committee just completed the 2022 Resource Guide, which took 4 months and will be ready to view at NACFC., and is currently analyzing pain and fatigue in CF patients.

The committee works hard to ensure that quality educational materials are available to people with CF, CF families, and CF Care Centers. They update the content of existing material, draft and edit new materials, review materials for Education Committee approval, and promote awareness of materials available. The Committee also looks at areas of educational need for people living with CF and seeks out experts who can help fill in the gaps that have been identified. They also review submitted abstracts to the NACFC and manage an online resource library for all CF Centers.

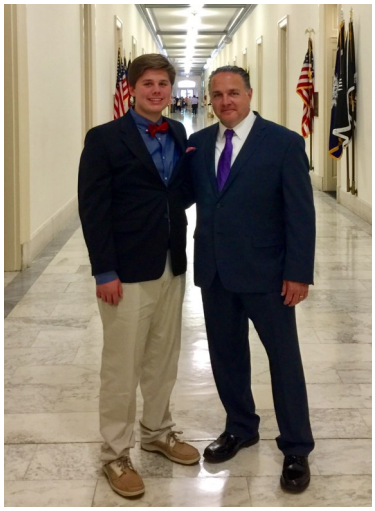
As a parent, you can help the education committee! Please be sure respond to any need assessment surveys that are sent out from the CFF or from our CF center. You can also share with your care team if you are in needs of something not found on the CFF website.

When we asked Jennifer what excited her most about working on this committee, she replied, "I love working with the bigger picture to better our CF Care Centers across the country. I think knowledge is power, so having the same information from center to center is super important, a unified voice to all of the CF community… it is a real privilege representing our center."

Submitted by Jennifer Leandro, RN

Inside this Issue:

An Easy, Effective Way to Support Your Loved One with CF—ADVOCATE!



Everett and Tim Tew on Capitol Hill in 2017.

As parents and caregivers, we manage a lot to help our CF loved ones live healthy lives. A powerful but simple step we can all take is to get involved with the CF Foundation's federal and state advocacy efforts to support the mission of ensuring that everyone with CF has access to high quality, specialized care.

Every year CF volunteers participate in events that help state and federal policymakers and decision-makers understand the complexities of living with CF, and the importance of having access to quality and affordable healthcare and supporting research and drug development.

In 2017, Congress was considering several health care proposals to remove important health care protections included in the Affordable Care Act. Prior to a pivotal vote on one of the proposals, a woman approached U.S. Senator Susan Collins at a Fourth of July parade in Maine and told her, "The bill is terrible, and my grandson has cystic fibrosis – he is going to have it his entire life. It's a pre-existing condition."

Senator Collins cast one of the deciding votes against the bill — a victory for people with pre-existing conditions. When asked, Senator Collins said the courage of that grandmother swayed her vote.

As the mother of a daughter with CF, I took part in the CFF's advocacy for The Affordable Care Act and continue to advocate as NC's Congressional District 5 Captain, recruiting others to support public policy initiatives through call-to-action alerts from the Foundation's public policy group.

My entire family advocates, including my son and husband who walked the halls of Washington, D.C., to advocate as part of the CF Foundation's Teen Advocacy Day.

As advocates, we must continue working to protect adequate affordable insurance and access to high quality care, and to fund CF research and drug development to support progress toward new therapies and a cure.

Start today! Whoever the advocate is, they have a story to share that can spark emotion, ignite conversation, and effect change.

To begin receiving advocacy alerts and information, **text "FIGHTCF" to 96387**. Extend the invitation to your friends and family.

Contact your local CF Chapter's Advocacy Chair for more information on how you can help; become a member of your district's advocacy team or become a congressional captain for your legislative district.

Easter/Central North Carolina Chapter – John Kelly kelly_johnw@yahoo.com

Western North Carolina Chapter – Doughton Horton dhorton@sodomalaw.com

Take Immediate Action Today to Support the Pasteur Act

Drug research is a vital part of the mission. Too many people with CF find themselves battling difficult-to-treat infections for which existing antibiotics are not effective. Funding science that holds promise to address infections, finding ways to pay for antibiotics and rewarding companies who bring these to market is critical.

The Cystic Fibrosis Foundation supports the PASTEUR Act, a bipartisan proposal that, if passed, will support the development of new antibiotics, and promote appropriate use of existing ones and spur vital investment into new antibiotics by addressing the economic disincentives that have long been associated with antibiotic development.

"People with CF need more antibiotic options as do all Americans. The Foundation is making significant investments into the research and development of new approaches to address infections, but more is needed. Congress must take swift action to pass the PASTEUR Act, invigorating a new era of antibiotic research and development that could bring novel treatments into the hands of patients who need them the most."

Take action today! Five minutes is all it takes. **https://act.cff.org/HTadfCF**

Submitted by Revonda Tew.

Specialist Corner:
Meet Dr. Adam Kimple & Dr. Cameron McKinzie

Adam Kimple, MD, PhD

Cameron McKinzie, PharmD

Adam Kimple, MD, PhD is an Associate Professor in the Ear, Nose and Throat department at UNC who is passionate about understanding boogers that accumulate in the noses of individuals with CF and helping rid the nose of these pests. Cameron McKinzie, PharmD is a CF clinical pharmacist whose career is dedicated to improving patients understanding of their medicines and helping them get access to their treatments.

They represented their specialties and UNC on a committee that developed the recently released CF Foundation ENT consensus guidelines. The committee consisted of 29 -person group that included otolaryngologists ("ENT" surgeons), pulmonologists, audiologists, pharmacists, a social worker, a nurse coordinator, a respiratory therapist, two adults with CF, and a caregiver of a child with CF.

As a committee they came up with questions to help guide recommendations that would help improve the lives of individuals with CF. After a comprehensive review of the literature related to these questions, the committee came up with 24 recommendations which are available at https://bit.ly/3Si4Esu.

A few highlights of the recently published guidelines include recommendations for:

1. Hearing screening for all children and adults with CF in anticipation of receiving medications that can cause hearing loss (i.e. tobramycin, amikacin)
2. Nasal saline rinses for children and adults with CF and nasal congestions or nasal drainage
3. Treatment of allergic rhinitis (symptoms like sneezing, congestion, itchy nose, sore throat) to improve nasal symptoms in children and adults with CF allergies
4. Referral to an ENT specialist for children and adults with CF with persistent ear, nose and throat symptoms (i.e. hoarseness).

We hope that these guidelines will help improve care of people with CF by addressing ENT concerns in a more consistent way. Your CF care team is happy to answer questions about the guidelines!

Submitted by Adam Kimple, MD , PhD and Cameron McKinzie, PharmD.

Tips for Packing School Lunch

Packing lunches in 5 easy steps:

1. Protein: hard boiled eggs, lunchmeat, chicken, pepperoni, yogurt, nuts, trail mix, almond butter, beans, canned tuna or chicken
2. Carbohydrate: pretzels, crackers, bagels, bread, English muffin, tortilla, waffles, pancakes, muffins, sweet potatoes, rice, pasta, granola
3. Fruit: banana, strawberries, apples, oranges, canned fruit, dried fruit
4. Vegetable: any cut raw veggies with hummus or ranch, salad, cooked veggies
5. Dairy: cow's or dairy free milk, yogurt, cheese, cottage cheese

Other tips to try:

1. Make lunches the night before and/or pack for 2 days at a time
2. Include kids in making lunches
3. Keep a basket of grab & go items (see list below)
4. Use compartment lunch containers to save time and packaging
5. Ask your child to write a list of their favorite lunch items in each food group

Try some of these grab& go items:

- pouches of canned tuna, salmon or chicken
- Pepperoni sticks or beef/turkey jerky
- Cracker packs with cheese/peanut butter
- Peanut butter pretzels
- Breakfast cookies/bars
- Dark chocolate
- To-go peanut or almond butter packs
- Individual snack packs of nuts, trail mix, etc.
- Dried fruit, fruit leather, apple sauce
- Full sodium soups—hearty or cream base
- Cheese sticks
- Yogurt
- Single serve juice, milks, yogurts, supplements

Submitted by your CF Nutritionists

Medicaid Beneficiaries Update

Since it began in January 2020, the federal Public Health Emergency (PHE) helped prevent NC Medicaid beneficiaries from losing their health coverage during the pandemic, even if someone's eligibility changed.

With the PHE officially ending on Monday, August 15th local DSS offices will begin initiating Medicaid redeterminations. Medicaid will not terminate or reduce benefits without completing a full eligibility determination. They want all eligible North Carolinians to be covered and stay covered. If coverage continues, Medicaid beneficiaries will receive a notice in the mail.

If a Medicaid beneficiary no longer qualifies, they will receive:

- Notice of when their Medicaid coverage will end
- Information on how to request an appeal
- Information about the Health Insurance Marketplace and other affordable health care coverage options

Actions Medicaid Beneficiaries Can Take Today

Don't miss out on important NC Medicaid and NC Health Choice information. Ensure that your mailing address, email, phone number, household size, and income are correct and up to date with your local DSS. You can find your local DSS on the Local DSS Directory: www.ncdhhs.gov/localdss

Report any life changes to the local DSS (www.ncdhhs.gov/localdss) as these may impact coverage. These changes may include marriage or divorce, the birth of a baby, starting a new job, or moving to another state.

Check your mail. Your local DSS will send a notice if any information is needed. It is very important to respond if DSS asks for information.

Other Health Care Options

Medicaid beneficiaries who no longer meet eligibility criteria and lose coverage have options to find affordable health insurance.

- Contact the NC Navigator Consortium at 1-855-733-3711 or go online at www.ncnavigator.net/schedule-assistance to schedule an appointment for free in-person assistance with health insurance applications
- Find a plan on the Health Insurance Marketplace www.healthcare.gov/
- Contact CFF COMPASS and/or your CF social worker.

COMPASS®
CYSTIC FIBROSIS FOUNDATION

Submitted by Amy Sangvai, MSW, LCSW

RESEARCH

Many CF studies are ongoing at UNC! More information on clinical trials being done here at UNC and around the country can be found on the Clinical Trials page at www.cff.org or by searching www.clinicaltrials.gov. You can also contact one of your hard-working Pediatric CF Research Coordinators directly:

Rosie Lainez at Rhlainez@email.unc.edu OR Thomas Shields at shieldt@email.unc.edu.

Research Update

The CF Foundation recently sponsored a Research Conference in Seattle, Washington last June. This conference, separate from the annual North American Cystic Fibrosis Conference, was designed to gather about 200 people who are at the cutting edge of CF research. Several sessions focused on the impact of Trikafta as highly effective modulator therapy (HEMT), showing that people taking it in the "real world" - as part of their regular CF care - are doing as well on it as people who took it during clinical trials which require very close monitoring and follow up A bronchoscopy study revealed that Trikafta reduces airway infection and inflammation, which is exciting since that sort of reduction has not been found consistently with previous modulator therapies. However, some inflammation and infection remained, indicating a need to continue monitoring and treating those taking Trikafta. Other sessions focused on developing new treatment approaches for everyone, including those who are not eligible Trikafta.

Many "novel" (new) strategies are being explored to correct the basic defect in CF. One intriguing approach uses mRNA to promote expression of functional CFTR. This method is similar to the mRNA based vaccines for COVID-19. Another novel approach involves stem cells, which are cells that can be obtained from the blood of an individual with CF, corrected in the lab, then converted into airway cells and put back into the lung. While promising, one challenge is that getting the corrected stem cells back into the lung typically requires damaging the airway to remove the native cells. Researchers at Carnegie Mellon University have developed a method to "shrink wrap" stem cells in a specialized membrane that allows the cells to unwrap and integrate into the airway without damage. Use of mRNA and shrink-wrapped stem cells are only two of the variety of approaches discussed in the conference, which are too numerous to describe in detail but include suppressor tRNA, antisense oligonucleotides, lung targeted peptides, engineered adeno associated viruses, and more. Although most of these strategies are several years away from being tested in the clinic, they demonstrate wide ranging efforts to address the basic defect in CF.

While we wait for a cure for CF, researchers are hard at work developing new methods to treat the mucus, inflammation, and infection associated with CF airways disease. For example, researchers have found that bacteria release extracellular vesicles (EVs) that contribute to their ability to cause infection, and targeting these EVs may serve as a novel treatment strategy. Researchers have also demonstrated that airway mucus and inflammation are barriers to many of the correction strategies described above, so addressing these factors will be critical for everyone.

While the conference included far more than can be summarized even briefly, one of the most important benefits was the opportunity to bring together scientists taking very different approaches towards addressing CF. This gathering resulted in a great deal of idea sharing among different research teams and resulted in new collaborations. These collaborations will undoubtedly accelerate advances in research that will lead to better treatment options for people with CF.

Submitted by Charles Esther, MD.

Want to Help Other Families Affected By Cystic Fibrosis?

Join the CF Family Advisory Board!

Who we are:

We are parents of children with CF who receive their care at the UNC Pediatric CF Center. We are dedicated to working with and getting to know the CF care team and each other in an atmosphere of trust and compassion and with the purpose of improving care and the quality of life for those with CF.

Mission:

We are dedicated to enhancing the medical care and quality of life for those with CF. Through collaborative efforts with the medical community; we seek to promote an open learning environment that results in personal empowerment and individualized care driven by evidence based best practices. In all we do, we seek to deliver the highest standard of safe and comprehensive care, provide compassionate support to CF patients and their families, and strengthen the collaboration between patients, family members and the health care team.

If you have interest in joining our monthly zoom meeting contact Jennifer Leandro at leandro@email.unc.edu.

Contact Us

Scheduling: 984-974-1401
(En Español: 919-966-6669)

Pulmonary Office:
919-966-1055 (8am-4:30pm)

CF Nurses:
919-966-1055 (9am-4pm)

Hospital Operator:
984-974-1000

Visit us on the web at:
www.uncchildrens.org/uncmc/unc-childrens/care-treatment/pulmonary-care/cf

UNC MyChart: https://myuncchart.org

uncpedscfcenter

Help Us Make the Newsletter Better!

If you would like to submit an article or have an idea for the next CF Family Newsletter, please contact Kelly Moormann at kelly.moormann@unc.edu.