

# Just In:

## NACFC 2016 Highlights

The 30<sup>th</sup> annual North American Cystic Fibrosis Conference was held in Orlando, FL from October 27-29, 2016. Many of the team members from UNC were able to attend and share ideas with over 4600 other health care providers and researchers from around the world.

Highlights from the conference included the information presented at the plenary sessions, which are available to view online at <https://www.nacfcconference.org>. *In the first plenary, Drs. Eric Sorscher (Emory University) and Mitchell Drumm (Case Western Reserve University) reviewed exciting data on medications that correct the basic genetic defect in CF (CFTR modulators and correctors), while also discussing gene editing as a new method of “fixing” the CF gene regardless of the type of mutation. In the second plenary, Drs. Patrick Flume (MUSC, Charleston; a former UNC fellow) and Christiane De Boeck (from Belgium), joined by nurse Patricia Burks (CF Foundation) and a panel of CF patients, presented information regarding clinical trials in CF and how important patient involvement is to moving forward with life-saving therapies. Finally, in the third plenary, Dr. Wayne Morgan (University of Arizona) reviewed data available through the CFF Patient Registry, one of the most comprehensive registries that helps us to understand the epidemiology of CF and outcomes over time.*

Research studies were a major focus at this year’s conference. The CFF Therapeutics and Development Network (TDN) is undertaking **51 clinical trials** over the next year! While everyone is naturally excited about the studies that look at the new CFTR correctors and modulators, there is very important work also going on looking at medications that affect inflammation, increase mucus hydration, and new ways of treating infections. The key to the success of these trials is the participation of you, the patient! To this end, the CFF has created the Clinical Trials Finder (which is on their website [www.cff.org](http://www.cff.org)). With this tool, you can find information on different trials that are being conducted around the country. If you find a trial in which you are interested, but UNC is not a participating site, we are happy to help you arrange a consultation at a nearby center to see if you are eligible for participation.

Additionally, our team members attended sessions focused on mental health, proposed changes in disability coverage, food insecurity, urinary incontinence,

quality improvement practices, physical therapy, new imaging modalities, care of the critically ill CF patient, lung transplantation, palliative care and others. A major theme of all sessions was how to engage people with CF and their families into the care center so we move towards more “patient centered care.” Many of the other topics in this newsletter address new ideas and information that our team brought home from the conference. You can expect to see changes at UNC, both in the outpatient clinic and in our inpatient work, reflecting some of the new collaborations and the latest data.

We are pleased to announce that our care center won two awards at the conference this year. We were awarded “Best Rolling Data” for our Quality Improvement Project surrounding transition of adolescents and young adults to the Adult CF Clinic, and the “Partnership in Caring” award for our collaborative work with the local CFF Raleigh’s Chapter.

~ Jennifer Goralski, M.D.



### IN THIS ISSUE:

NACFC 2016 Highlights	1
Mental Health Update	2
Personal Assessment / Changes in Disability	2
“Breakthrough” Treatments Updates	3
Personnel Update/6BT Inpatient Corner	3
Shared Services/QI Meetings @UNC	4

# MENTAL HEALTH UPDATES

It is common for those living with chronic illness to experience depression or anxiety. In one study, the rates of patients living with CF and parent caregivers in the United States and Europe were 2-4 times higher than in the general population. Some of you may read this and think, "Isn't being sad or down just a normal part of being human?" Yes. It is normal to experience a full range of emotions including sadness, anger, or joy. However, some emotions, like sadness, can become more than a normative response.



## Here is how to tell the difference between depression and normal blues or sadness:

- Depressed mood lasts for at least two weeks or more for most of the days, more time than not in the day
- It is associated with decreased interests, motivation, and enjoyment (for example, you don't want to do things that usually bring you pleasure; you have been isolating)
- It is associated with changes in your bodily functions: sleep, appetite, energy
- It is associated with decreased concentration and ability to make decisions
- It is associated with increased thoughts about death
- It affects your functioning: decreases your ability to take care of yourself and your health, has a negative impact on your relationships, school, work and/or hobbies

For those living with chronic illness, anxiety surrounding medical interventions is very common. For patients living with cystic fibrosis, these anxiety-producing interventions can include: PFTs, throat cultures, venipuncture, PICC line placement, NG tube insertion, manipulation with the G/J tube, or physiotherapy/ nebulizer treatments, just to name a few. This can add to the anxiety of coming to the clinic appointments or staying in the hospital, and for some patients might lead to avoidance of these anxiety-producing situations and thus some essential care.

Because we know that depression and anxiety can have a direct effect on a patient's quality of life and their ability to keep up with daily treatments or doctor's appointments, the UNC staff started screening patients at both outpatient appointments and during a hospitalization. We hope that by doing this, we can have richer conversations with our patients about how CF is impacting your mood and resources available for support. Since implementing the screenings in August 2016, we have screened 116 patients. Of the patients we have screened, 48.3% screened positive for depression, 41.3% of patients screened were positive for anxiety and 39.7% of patients screened positive for both depression and anxiety. For those patients who are interested, we have been offering counseling resources either through UNC or within their local community to address symptoms and build or strengthen coping strategies. We appreciate the honest dialogue that has been occurring and the trust that you put in your team when you disclose how CF is impacting the many facets of your life. If you have any questions, please reach out to either social worker in clinic, Jennifer Pagel or Brooke Jones.

~ Brooke Jones, MSW

## New Personal Assessment

The UNC CF Adult Clinic is implementing a new Personal Assessment that patients will complete upon arrival to clinic at each of their visits every 3 months. The assessment is a short, one page series of questions regarding patient concerns, current health status and treatment therapies, as well as patient health goals and barriers and should take approximately 5 minutes to complete. We are asking you to complete this assessment as a means of better understanding your current health status and to create an action plan to better your overall health as a CF patient. Additionally, this information will aid in providing important information for available patient resources.

~ Katie L. Howe, M.S.

## Social Security Disability Changes

As you may or may not be aware, there have recently been changes in the social security disability guidelines related to cystic fibrosis. These changes may directly affect your current social security disability benefits as well as for anyone that will be applying for social security disability in the future. We are working on gathering information related to the changes and will provide this information as soon as it is available. In the meantime, you can contact the CF Legal Hotline at 800-622-0385 or COMPASS with the CF Foundation 844-266-7277 if you have specific questions or concerns about your social security disability benefits. You can also reach social workers: Brooke Jones (984) 974-5128 or Jennifer Pagel (984) 974-3385.

~ Jenn Pagel, MSW

## “BREAKTHROUGH” TREATMENTS UPDATE

2015 was a big year for CF when Orkambi received FDA approval. This meant that many patients were able to receive treatment directed at the underlying cause of CF – not just control of the downstream effects. However, Orkambi is only indicated for people with two copies of the F508del mutation, the most common mutation that causes CF. That’s only about half of the patients in our clinic. Although no additional phase 3 trials were reported at the meeting this year, multiple ongoing studies of a medication called VX-661 (Vertex Pharmaceuticals) in different genotype groups are nearing completion and should be reported this year. VX661 is a CFTR corrector that has shown promise to replace lumacaftor (one of the two medication components that make up Orkambi). VX-661 is quite similar to lumacaftor and, therefore, it is reasonable to expect that its clinical performance will be similar to its predecessor, but with fewer drug interactions. Even more exciting are new studies that will test the combination of **three** medication combinations (anticipated they will begin this year). With roughly 9 companies now competing in this area, we expect faster development of increasingly effective drugs over the next few years. As these drugs become better at restoring CFTR function, patients with only a single one copy of F508del are predicted to be helped as well. Once achieved, this will then bring the number of patients with an available CFTR modulator to 95% of the CF population. While this still does not reach the CFF goal of having disease modifying therapies for every patient, additional efforts are already underway to identify compounds that will bypass abnormal stop mutations in the CFTR gene (most of the remaining 5% of patients). Clearly there has never been a more exciting time for all of us who are working to bring, and have access to, new therapies for CF!!



~Aaron Trimble, M.D.

### PERSONNEL UPDATES



Brooke Jones is a native to Raleigh, NC and has been with UNC since 2012. She started in July 2016 as a new CF clinic social worker as well as the mental health coordinator. She currently lives in Raleigh with her husband, daughter, and pitbull.

Nicole Bingham has been a nurse for 13 years, all of which was spent on the pulmonary and infections disease medicine floor (aka 6 Bedtower) at UNC. She started training with Jaimee Watts in October as the CF Nurse Coordinator. She will take a position in the Pulmonary Specialty Clinic as a nurse in January 2017. She lives in Chapel Hill with her nurse husband and son.

### FOOD INSECURITY

Food insecurity exists all across our country. North Carolina rates among the top ten states with a high rate of people experiencing food shortages; nearly 1 in 6 families according to some sources. The role of nutrition in individuals with cystic fibrosis is well known; however, it can be challenging for people to meet nutritional needs if there are barriers to accessing food sources. There are resources available to help meet nutritional requirements during times of need – these can include food pantries, Food and Nutrition Services (Food Stamps), Supplemental Nutrition Assistance Program, and Women, Infants, and Children Supplemental Program thru NC Health and Human Services. Your CF team is available to help identify and navigate these resources if you are interested.

~ Courtney Busby, R.D.

# 6BT

# Inpatient

# Corner

CF patients *only* on contact precautions may ambulate **independently** outside their rooms on the unit in which they are housed provided the following is done:

- Ideally, patients change in to clean clothes or gown before leaving room.
- Patients perform hand hygiene before leaving their room and put on a mask.
- Patients maintain a greater than 6 foot distance from other CF patients
- Patients are instructed to not touch objects in the environment, environmental surfaces, or other patients.
- Patients must remain only within the unit corridors and may not enter other common areas, including but not limited to: visitor waiting rooms, nutrition areas, nursing stations, and other patient rooms.
- If the patient wishes to leave the unit, **they must be accompanied by healthcare personnel.**—
- Patients who cannot or will not follow these requirements must be accompanied by trained healthcare personnel when ambulating in the hallway. During outbreak situations, Hospital Epidemiology may temporarily suspend these privileges.

## UNC SHARED SERVICES CENTER PHARMACY

The pulmonary clinic has a full-time clinical pharmacist, Emily Wong. Through the new service, Emily acts as a liaison between your provider and our pharmacy care team at the UNC Shared Services Center Pharmacy for all eligible **pulmonary specialty medications**. Once approved, your prescriptions will be mailed directly to your home at no additional cost to you.

**In addition to fulfillment of your prescriptions, our pharmacy care team will also:**

- Request insurance approvals
- Offer financial guidance
- Serve as your advocate to the insurance company and medication manufacturer
- Call you on a monthly basis to set up your next medication refill
- Provide additional clinical oversight to ensure your safety and care
- Answer any questions or concerns

If you reside in North Carolina and your insurance allows you to fill at our pharmacy, we will begin your personalized pharmacy care service by sending your specialty medication prescription to UNC Shared Services Center Pharmacy.

You may also be able to consolidate your other chronic medications and have them fill at one pharmacy.

**CF specific medications available at UNC:** Pulmozyme®, Inhaled tobramycin nebulizer, TOBI® nebulizer solution, TOBI PodHaler®, inhaled colistin, pancreatic enzymes, multivitamins, hypertonic saline, Orkambi®, Kalydeco®

*\*If you have HealthWell Grant Assistance, the pharmacy can also directly bill your copays\**

**Our team believes in this interdisciplinary approach and has seen the benefits in our patients. We would enjoy offering this opportunity to you!**

If you would like to learn more about this program, please contact Emily Wong at (984) 974-0012 or [pulmonaryclinicpharm@unchealth.unc.edu](mailto:pulmonaryclinicpharm@unchealth.unc.edu).

~ Emily Wong, Pharm. D



**REMINDER:**

FLU VACCINES ARE STILL AVAILABLE IN CLINIC.



### QUALITY IMPROVEMENT @ UNC'S ADULT CF CENTER

The QI Team is an interdisciplinary group of doctors, nurses, social workers, as well as our dietician, pharmacist, and respiratory therapist who are working together to improve the quality of care for us both inpatient and outpatient. It has been inspiring to see how much time and effort they are putting in behind the scenes. I am grateful to have recently joined the team to offer a patient's perspective. Together, the QI team has developed the new Personal Assessment you'll fill out at clinic visits, so that your caregivers can better address all of your needs. The QI team is also working on ways to track and encourage patients to stay on schedule with appointments, so that problems can be identified and addressed quickly. The team is also working hard to better meet the mental health needs of CF patients at UNC (see Mental Health Update in this newsletter!). If you have any suggestions for the QI team, let us know by emailing the [AdultCFNurse@unchealth.unc.edu](mailto:AdultCFNurse@unchealth.unc.edu), who will bring your ideas back to the QI team.

~ Katie Reisinger, Adult CF Patient



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*UNC Adult Cystic Fibrosis Center*

**KEEP IN TOUCH**

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**TO MAKE OR CHANGE A CLINIC APPOINTMENT PLEASE CONTACT THE CLINIC DIRECTLY AT 984 974 5703**