

Just In:

BE PART OF THE CURE!

We don't want to sound like a broken record, but this is truly an incredible time for CF research! We hope you've heard about the incredible progress that is occurring during clinic visits and are as excited as we are. If not, here are a few updates, and be sure to ask about these next time you see us in clinic.

1. *Restoring the function of the CF protein:*

When Kalydeco (ivacaftor) was approved in 2012 for ~4% of patients with specific CFTR (the CF gene/protein) mutations, we realized what an amazing improvement in health typically results following treatment with a highly effective CFTR "modulator". Since this time, the CF community has been pushing hard to bring similarly effective treatments for all patients with CF. Approval of Orkambi, followed more recently by Symdeko, were 2-drug combinations that moved us toward this goal. These drugs are currently being used by many patients with 2 copies of the common CFTR mutation (F508del). Despite being less effective than Kalydeco, they have been shown to yield small improvements in lung function and significant reductions in how often pulmonary exacerbations occur. They also slow the rate of decline in lung function over time.

Over the last year, testing of 3-drug combinations from multiple pharmaceutical companies have been tested in phase 2 trials (Vertex, Proteostasis, Galapagos). These results have generally been extremely positive, with large improvements reported in press releases. Currently, phase 3 trials of Vertex compounds (the final step before approval) are underway at UNC and across the world. Perhaps most importantly, not only do these regimens appear to be much more effective than existing regimens (i.e. Symdeko, Orkambi) for patients with 2 copies of the common mutation (about 45% of the CF population), but also appear to be equally effective for patients with only a single copy of this mutation (~40% of the CF population). This means that in the near future, we hope to be able to offer highly effective CFTR modulator therapies for more than 90% of adult patients. This is a total game changer that we couldn't be more excited about!

2. *Non-tuberculous mycobacterial infections:*

17% of adult CF patients at UNC grow mycobacteria (typically *Mycobacterium avium* or *Mycobacterium abscessus*) in their sputum. These unusual bacteria are not always problematic or need treatment, but can cause lung destruction in some individuals. Treatment can be difficult, and typically requires at least a year of therapy using combinations of multiple drugs. Understanding when treatment is needed, and how best to treat these organisms, has been identified as one of the most important priorities for CF clinical research by experts in the CF community. Investments to identify and test novel treatments are already underway. Your team at UNC is involved in a national consortium of ~10 sites (led by University of Colorado) that is aimed at answering these questions. If you happen to grow one of these organisms, we will talk to you in detail about the best approach to diagnosis and treatment, and may invite you to participate in these studies.

3. *"Therotyping" for unusual CFTR mutations:*

Scientists at UNC have been leaders in the development of assays (i.e. tests) that assess the ability of medications to restore CFTR function using cells collected from patients. Because there are nearly 1900 different CFTR mutations that can cause CF, many patients have at least one poorly characterized mutation, and these may not be included in treatment trials of CFTR corrector drugs that are being carried out in patients with more common mutations. Further, this work is important because it provides a potential means of comparing the effects of different combinations of CFTR modulators. We do not yet know whether these assays can accurately predict the response that a patient will have in response to a CFTR modulator, but because we want to provide the most effective CFTR modulators to every patient who will benefit from them, we may invite you to participate in some of these studies. Involvement is typically very short (one visit), and may require collection of cells either from the nose (via a brush) or the rectum (via a simple biopsy procedure done in the research clinic). Stay tuned!

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Thanks for your interest and involvement! You can learn how to get involved by calling the Marsico Clinical Research Center – 984-974-9198 – and asking for one of our CF Research Coordinators. You can also visit CFF.org and look at the Clinical Trials Finder to see research that is going on across the country – since we can't take on all the important studies that need to be done, we are happy to refer you to a research center that is conducting other studies which may be of interest to you!

~ Scott Donaldson, M.D.



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DID YOU KNOW?

The order in which you take your Inhaled Medications matters! Here's why.....

The Cystic Fibrosis Foundation and the UNC CF Center recommend the following sequence for inhaled medications:

1. **Bronchodilators** (Albuterol, Xopenex) to open the airways, decrease spasm, and aid other inhaled medications in working more effectively.
2. **Hypertonic Saline** (3%, 7% or 10%) draws water into your secretions and in turn thins and mobilizes mucus and improves airway clearance.
3. **Airway clearance techniques** loosen the thick, sticky mucus so it can be cleared from the lungs. Clearing the airways may help decrease lung infections and improve lung function.
 - ***Aerobika, IPV, Vibralong** all can be used while inhaling bronchodilators and hypertonic saline, but never in combination with pulmozyme or inhaled antibiotics.
 - ***The Chest Vest** can be used during or after the inhaled bronchodilator and hypertonic saline.
 - ***Flutter and CPT** should be completed after inhaled bronchodilator and hypertonic saline.
 - ***The huff cough** is also a useful tool during and after hypertonic saline to clear secretions.
4. **Pulmozyme (DNase)** is a mucolytic that breaks down the structure of the mucus to make it easier to cough out.
5. **Inhaled Antibiotics (TOBI, Colistin, Cayston)** are used after your previous therapies to allow the antibiotics to penetrate as deeply as possible to treat the bacteria.
6. **Inhaled Steroids (Flovent, Pulmicort, Advair, QVAR, Symbicort)** are medications to help reduce lung inflammation in people with CF and asthma. They need to stay in the airways as long as possible to be most effective, so take them after completing all the therapies that make you cough.

If you start coughing up blood (more than a teaspoon), you should stop pulmozyme, hypertonic saline, and airway clearance and contact your physician. If you sense an exacerbation take all inhaled medications and airway clearance as ordered, consider increasing the frequency of your airway clearance and hypertonic saline, and contact your physician.

DON'T GET BEHIND!

Although there is no magic solution for managing life with CF, consistency is one of the best ways to keep yourself as healthy as possible. Although daily consistency with complicated and time-consuming treatments is difficult, keeping up with regular clinic visits is a great way to make a commitment to being as healthy as possible. We know from data from the CF registry that keeping up with regular clinic visits is important for optimal health. The CFF recommends routine visits every three months, and more frequently if you have been ill. If you've had a pulmonary exacerbation, then the focus of the visit will be to try to get you better, and less about planning healthy routines. It's easy to get into a cycle of constantly playing "catch-up" with your health, rather than working with your provider to come up with strategies to forge ahead with your health. In these situations, many things get overlooked, such as screening for diabetes, liver disease, bone health, colorectal cancer screening, vitamin deficiencies, and other treatable complications of CF that, if left un-treated, can make you even worse off. If it's been more than three months since you've been in to clinic, we'd like to see you back as soon as possible. If there are issues that make getting into clinic difficult (employer difficulties, transportation, etc), we may be able to assist.

Spiritus Project Golf Tournament / Great Strides 2018

Spiritus Project Golf Tournament:

On April 9, 2018, several members of our adult team attended the 3rd annual Spiritus Masters golf tournament in Denver, NC. It was predicted to rain all day, but luckily it cleared up just in time! There were about 200 participants in total, including golfers, volunteers and sponsors. Participants were able to bid on items in a silent auction, as well as a live auction later on. At the end of the evening, a reception was held where several people spoke, including one of our very own Marianne Buchannan, adult CF social worker. She reminded us of all the ways that Spiritus has helped our patients throughout the year. In total, the golf tournament raised \$100,000 to use toward providing financial assistance for CF patients throughout North Carolina.

As a reminder, Spiritus Project is a non-profit organization that was started in honor of one of our patients who passed away in 2015. Not only do they provide hospital admission bags at UNC, but they also provide various forms of financial support to patients in need.



Great Strides Chapel Hill 2018

On April 14, 2018, we celebrated the 30th anniversary of Great Strides by walking a 5k through Community Center Park in Chapel Hill, NC. It was a fun-filled day with members of the UNC Pediatric, Adult, and Research teams, as well as family and friends. Dr. Scott Donaldson gave a short speech about advances in CF research and the need for continued support through Great Strides. Becky Woodruff, walk chair and leader of Holly's Heroes, spoke about what Great Strides means to her and her family. As we walked through the rose garden, we were again reminded of why we walk. At the end, participants enjoyed music from DJ Billy and food from local vendors. To date, the walk has raised about \$28,500, which will go toward research and care for children and adults living with cystic fibrosis in North Carolina.

PERSONNEL UPDATES



Sallie Barrett, RN-BSN, received her nursing degree from UNC-CH in 2002 and worked as a neonatal ICU nurse at Duke Hospital before switching to a career in pharmaceutical sales. After over a decade working in psychiatric specialty sales, she decided to return to her passion of helping patients in a nursing capacity. Sallie joined the Adult CF team in May 2018 and is thrilled to have found a nursing role where she can contribute

as part of an interdisciplinary team. Sallie lives in Chapel Hill with her husband and stays busy “raising little women” with their 3 beautiful girls.



Donna Enloe, RRT, received her degree in 2012 and has worked as a respiratory therapist at UNC Chapel Hill her entire career. She has spent 5 of those years working primarily with cystic fibrosis patients in the hospital setting and has recently accepted the role of Cystic Fibrosis Clinical Specialist which has expanded her role to the clinic setting as well.

She is excited to become a part of the clinical team and to continue to learn and grow in her new role. She is currently enrolled at UNC-Charlotte working toward her BSRT. Donna is married and lives in Ramseur with her husband and stays busy with school, her family and “fur babies”.

CF FALL FUNDRAISER and LOCAL FINANCIAL ASSISTANCE PROGRAMS

- o **SPIRITUS:** Spiritus is the Latin term for breath, often used figuratively to mean spirit. The Spiritus Project is a non-profit organization whose concept was created by a UNC CF patient and was launched by her family and friends in 2015 in order to provide UNC CF patients financial support in order to reduce stress and increase quality of life. If you or someone you know may benefit from this program, or to make a donation, please check out the Spiritus Project (www.spiritusproject.org) for further information.

- o **The Garrett Thomas Foundation:** The Garrett Thomas Foundation was established to help individuals and their families who are living with Cystic Fibrosis in the states of North Carolina and South Carolina with transportation, medical, housing, educational, and family leave assistance. If you or someone you know may benefit from this program please contact your CF Social Worker [Woker](#) or if you would like to make a donation to The Garrett Thomas Foundation please visit: www.garrettthomasfoundation.org.

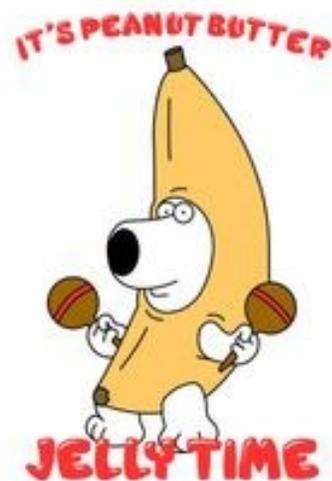
- o **UNC CF Patient Assistance Fund:** The UNC CF Patient Assistance Fund is used for airway clearance devices, transportation assistance, and most recently grocery assistance. We hope to raise awareness around food insecurity among those living with Cystic Fibrosis and support patients who have an increased caloric need but who are unable to afford meals in order to support that need. We have been able to raise funds for these types of assistance via local CF Awareness Nights as well as our Annual Fall Fundraiser; If you are having difficulty affording food or transportation to clinic appointments, [please](#) contact your CF Social Worker. Also, donations are always greatly appreciated! If you would like to donate, please visit: <https://go.unc.edu/cf>.

- o **CF Fall 2018 Fundraiser:** Our annual UNC CF Fall Fundraiser and Corn Hole Tournament will be held on Sunday, September 23rd to raise awareness surrounding our Grocery Assistance Program and to raise money for UNC's Patient Assistance Fund. The program was piloted in the fall of 2017 and created in order to serve patients who are food insecure. After last year's fundraiser, we were able to expand the program to include more patients, but do not yet have enough funding to serve all patients in our [clinic](#) who have trouble affording food. We're currently organizing our second annual event with the hopes of being able to do more for our patients who have difficulty being able to pay for their basic needs. For further information please contact Katie Howe at katie_howe@med.unc.edu or (984) 974-2965. You may also visit: <https://go.unc.edu/CFCornhole18>.

MAKE YOUR SNACK COUNT

300-500 Calorie Snack Ideas

- ◆ Turkey and cheese pinwheels (flour tortilla, 2 slices cheddar cheese, 2-3 slices deli turkey)
- ◆ Peanut butter & jelly sandwich (2 slices bread, 1 tablespoon grape jelly, 2 table spoons peanut butter)
- ◆ 1 large muffin with 1 cup whole milk
- ◆ 3 ounces cheddar cheese, 6 saltine crackers
- ◆ 1 cup Cherrios, 1 cup whole milk, 1 banana
- ◆ 2 ounces of peanuts
- ◆ 1 bagel with 1 tablespoon cream cheese
- ◆ 1 cup vanilla ice cream with 1 tablespoon chocolate syrup
- ◆ 1 pack Nature's Valley Granola Bar and 1-2 tablespoons peanut butter or almond butter



A POEM - MORNING FEEDING

by Evana Bodiker

Agonized, like a baby bird screeching
 For its mother to deliver it an earthworm,
 The toddler's hunger sounds set the nursery
 Going, warned a sleeping house that the time's
 Arrived yet another fruitless meal.

Mother Sisyphus climbed the stairs,
 Nesting the milk bottle between hands
 To warm it, hoping it would start the day
 With her colic child pacified.

She broke the enzyme capsules in half,
 Spilling their beans onto the child's tongue.
 Then waiting for sobs to turn to suckling,
 For her daughter's stomach to announce
 With gurgling consent, its wordless gratitude for her.

In the wicker chair, rocking for hours,
 She keeps the bottle firm between forming teeth,
 The ones she hopes her daughter will soon
 Learn to use, to chew and to not starve.

BE PART OF THE CURE!

Continued.....

4. Developing better ways to test new CF treatments: One of the factors that slows down how rapidly we can test new CF medications is the absence of tests that clearly predict whether that drug will be a success or not. Typical lung function tests, for example, vary over time and can miss important changes in the lung. As a result, hundreds of patients need to be enrolled in a trial in order to learn whether the medication is likely to be effective. At UNC, we are working to develop tests that are more informative and more sensitive to treatments. We have recently developed an imaging method that allows us to "see" where inspired air goes in the lung, and where the airways are too blocked to allow air to pass. This test relies upon MRI, which uses a strong magnet but no radiation to create images, and an inhaled gas that can be visualized by the MRI machine, but isn't absorbed (so is simply exhaled after the test is completed). Our early results show that we can see clear abnormalities in these images, even in patients who have normal lung function tests. Several studies are ongoing, and we need your help to make them ready for future clinical trials and/or clinical care. Please ask us about this work if you're interested!



SPIRITUS PROJECT

For more information regarding the Spiritus Project please visit www.spiritusproject.org or contact Helen or Robert Link.
 Helen Link: hml@spiritusproject.org
 Robert Link: rl@spiritusproject.org

UNC CF Online Support Group

The UNC CF Center is now offering online support groups that meet for 8 weeks at a time. If you're interested, please contact Brooke Jones (brooke.jones@unchealth.unc.edu) or Marianne Buchanan (Marianne.buchanan@unchealth.unc.edu) for more information. Here is what one patient had to say about their experience in group:

"I recently completed an online support group, specifically for people with CF at UNC and cannot say enough positive things about the experience. Before I joined, I was skeptical to say the least. I was worried it would be depressing and I would leave each group meeting feeling like CF was all that mattered in my life. I was so wrong! I was able to meet people that helped me to know that the feelings I had about life, CF, and managing everything was completely normal. I ended the support group with friends I didn't expect on making and a sense of strength that I really needed at this point. If you are hesitant about joining, don't be. If you are worried about judgement, don't be. I would recommend prioritizing the hour a week for a few weeks and joining one of the support groups being offered! You won't regret it!"



Follow us on Facebook!!

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