

Just In:

Long Term Health Care Planning

Have you ever had questions about your health that you didn't know how to ask? Have you ever wanted to talk to your doctor or health care team about what to expect in the future but the time never seemed right? The Adult CF Clinic team knows this is a common and important issue and wants to help. We understand that talking about the future can be hard, and no one wants to think about their health deteriorating. However, we also know that it is important to make your health care wishes known while you are healthy, and that doing so can have many benefits. Studies funded by the Agency for Healthcare Research and Quality (AHRQ) have shown that people who talked with their family, physician, or others about their preferences for long term health care had less fear and anxiety, felt more in control of their own medical care, and believed their doctor had a better understanding of their wishes.

Later this year, we plan to begin offering "Long-Term Health Care Planning" sessions to patients in our clinic. Our hope is that this program will start the conversation about this topic between you, your CF team of providers, and the family and friends who you feel should be involved in these conversations. To accomplish this, we will provide those who are interested with some introductory materials to help you identify topics you want to discuss and help questions you need answers to. After you've had time to look at these materials, we will reserve a time to meet with you and discuss a range of topics that focus on a range of relevant topics, including general information on the course of CF over time, lung transplantation, symptom management, social supports, and documentation of your wishes related to your healthcare. This will be held in the clinic, but will be separate from your usual clinic appointment. It might be on the same day as your clinic appointment or on a different day, whatever works best. Again, this is meant to be a conversation starter which we hope will continue over time. Most of all, we want

you to know that we want to help and support you in any way we can.

In the meantime, if you have questions related to this topic, feel free to reach out to your CF Social Worker:

Brooke Jones, MSW:
 brooke.jones@unchealthcare.unc.edu
 984-974-5128

Marianne Buchanan, MSW:
 Marianne.buchanan@unchealthcare.unc.edu
 984-974-3385

~ Laura Beth Rupcich, P.A.



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CF GREAT STRIDES

On a bright sunny morning in April, more than 100 walkers gathered at Chapel Hill Community Center Park to make Great Strides against cystic fibrosis. Fifteen teams who had raised more than \$30,000 together walked through the rose garden and onto Lake Creek Trail for the scenic 3k stroll. Children and adults living with CF, friends, family, physicians, nurses, caregivers, and other supporters of the CF community celebrated the many successes in CF research and care. Sponsors Abbvie and Chiesi shared their inspiring thoughts with the crowd and Dr. Scott Donaldson updated everyone on the newest promising research toward a cure and control for CF.

The walk was followed by a community picnic with all the trimmings including a DJ to keep everyone on their feet, bubbles and watermelon to drip off everyone's chins.



As one grandmother explained, "It is very fitting that we walk through a rose garden to start the walk. Many years ago Mary Weiss, mother of 3 boys with CF, was frantically calling everyone in the phone book to see if there was anyone who could help, her young son Richard approached her. She was surprised to hear him say that he knew what she had been doing. He matter of factly informed her that she was trying to save the "sixty-five roses." Because of the efforts of Mary Weiss, walkers at Great Strides Chapel Hill and many others, people living with CF are living into their 40s, 50s and beyond and we are getting ever closer to the day when CF will stand for CURE FOUND!

Walkers and teams are welcome to continue fundraising through the end of the year. Please contact Marla Dansky at dansky@cff.org or 919-845-2155 for ideas and support. Thank you for all that you do!

~ Jaimee Watts, R.N.

Spiritus Project Golf Tournament



The 2017 Lake Realty [Spiritus Masters](#) presented by Novant Health was a success! 2017 was another successful event, raising more than \$100,000 for [Spiritus Project](#). There were nearly 200 golfers, sponsors, volunteers and staff who participated in this event on April 10, 2017 at the Arden Ridge Golf and Country Club. The funds raised will assist in the purpose of the Spiritus Project, which is to provide various forms of direct, need-based financial assistance to adult cystic fibrosis patients in an effort to reduce stress and provide an improved quality of life. For more information, visit www.spiritusproject.org.

Urinary Incontinence

Loss of bladder control is embarrassing, inconvenient, and --- very common. In fact, nearly 75% of women with CF report problems with urinary incontinence at some point in their life, compared to 13% of women without CF. For men, it appears that almost twice as many men with CF have issues with incontinence, compared to healthy peers. Incontinence is likely caused by a number of issues, including chronic cough, increased stress on the pelvic muscles during airway clearance, or underlying structural problems. Incontinence can result in increased anxiety and depression, and a decreased quality of life.

Fortunately, there is treatment available. Your doctor can refer you to a "continence physiotherapist" who can help retrain your bladder using exercises and biofeedback to improve the pelvic floor strength. Your airway clearance may even need to be adjusted to take the stress off the pelvic muscles.

If you suffer from urinary incontinence, don't be afraid to talk to your CF provider. We can help!

~ Jennifer Goralski, M.D.

“BREAKTHROUGH” TREATMENTS UPDATE

Clinical research at UNC’s Adult CF Center remains as busy as ever, thanks to your participation! With countless hours, visits, and extra trips to Chapel Hill, our patients are helping to improve the lives of people with CF not just in North Carolina, but worldwide. This year, the CFF supported Therapeutic Development Network, which UNC is a member of, will conduct more than 50 clinical trials aimed at rapidly bringing new therapies to our patients and new knowledge about CF.

In recent months, there have been several recent highlights we’d like to share! In March, Vertex announced results from two phase III trials of tezacaftor (VX-661) in combination with ivacaftor. These studies involved patients with various CF gene mutations. While we await publication of final results, both studies met their objectives. In patients with 2 copies of the common CF gene mutation (delF508), the EVOLVE study showed a clinically meaningful improvement in lung function. In these patients, this new combination would be a potential replacement for Orkambi, rather than an addition to Orkambi. Tezacaftor/ivacaftor may have advantages over Orkambi due to fewer drug interactions and, hopefully, fewer instances of intolerability. In the second study, called EXPAND, patients were required to have one copy of the common CF gene mutation, and one gene copy that led to a partially functioning CF protein. Both the combination of tezacaftor/ivacaftor and ivacaftor by itself were tested. Again, both single drug and combination drug approaches meaningfully improved lung function and reduced CF symptoms. At this point in time, we await the decision by the FDA regarding approval of this new drug combination.

In early May, the FDA expanded the group of individuals who can be treated with ivacaftor (Kalydeco) to include 33 additional mutations. Like in the EXPAND study, these mutations retain a small amount of CF protein function. Interestingly, this FDA decision was made based upon both clinical data and the results laboratory experiments that demonstrate the activity of ivacaftor in these patients. This is important, because many mutations in CF are very rare, which makes it very difficult to conduct large clinical trials. This move by the FDA signals that laboratory tests may play a larger role gaining approval for other rare mutations as well.

Currently underway and/or on the near horizon or many other approaches to treatment. Your UNC CF Center team is working hard on many of these trials, including new 3-drug combinations designed to improve CF protein function in many more patients, and increase the amount of benefit for patients with the common mutation. There are also a number of other non-CFTR modulator studies going on, including some other therapies, such as a drug that blocks over-active sodium channels in CF, a study of the effectiveness of airway clearance devices, and measuring ventilation in different areas of the lung using MRI scans. New studies are being started all of the time, and we invite you to keep track of current studies being performed at your center by:

- ◇ Visit the UNC CF Center website (<https://www.med.unc.edu/marsicolunginstitute/adult-cf-program>) and check out the CF Clinical Studies page
- ◇ Visit the Clinical Trial Finder at CFF.org (<https://www.cff.org/Trials/finder>)
- ◇ Talk to your CF Clinic and Clinical Research team members either in clinic or by contacting us directly with any questions you might have (Marsico Clinical Research Center: 984-974-9198)



~Aaron Trimble, M.D.

PERSONNEL UPDATES

Marianne Buchanan has been a licensed social worker for 16 years and has spent most of this time working with patients who struggle with depression, anxiety and other mental health problems. She joined the UNC CF team in February, 2017 and has been grateful for the warm welcome from both the CF team and CF patients. She lives in Chapel Hill with her husband, 2 daughters, and goofy black lab.



Laura Beth first became interested in CF while working as a clinical research assistant in the UNC Cystic Fibrosis Center as an undergraduate. It was there that she was inspired to take care of patients and then went on to become a Certified Physician Assistant (PA-C) at Wake Forest University. After completing this, she worked as a PA in Hospital Medicine at UNC for 5 years, during which time she took care of many adult CF patients during their hospitalization. She is excited to have joined the UNC Adult CF team full-time in February 2017 and is seeing patients in the Adult CF Clinic as well as providing consults on our patients in the hospital. Laura Beth lives in Durham with her husband and sweet dog Sasha.

Nicole Bingham graduated from nursing school at UNC Chapel Hill in 2003. She has spent the majority of her career caring for pulmonary inpatients, including those with cystic fibrosis. She worked closely with the Adult CF team as a key inpatient liaison for years, and transitioned to the outpatient CF and Pulmonary Clinics in October of 2016. Beginning in June, she is assuming lead Adult CF Nurse position, with all of her time devoted to Adult CF Care. In her personal life, Nicole lives in Chapel Hill with her family, loves the beach, cooking, and coffee.



As we say hello to new staff coming in, we will be also be saying goodbye to a member of our team. Jaimee, the CF nurse coordinator has left to go back to school. While she is sad to go and will be missed, she will not be leaving UNC entirely and you may see her in the hospital one day.

FOOD INSECURITY

Food insecurity is defined as not having reliable access to an adequate supply of affordable, nutritious food. North Carolina has the **8th** highest rate of food insecurity in the nation, with 15.9 percent of households experiencing food insecurity. For some of our patients, maintaining a sufficient supply of food is difficult due to the financial strain of healthcare and overall living expenses. Because poor nutrition status is correlated with a decline in pulmonary function in patients with CF, the UNC CF Center will be asking a few questions at your appointment regarding your ability to access enough food or whether you are stretching or skipping meals. If this is occurring, however, you do not need to wait for an appointment to discuss further as you can contact our SW team, Brooke Jones and Marianne Buchanan, or our Dietician, Courtney Busby. We can assist in identifying resources to assist.

~ Brooke Jones, MSW

CHANGES IN HEALTH WELL

The HealthWell Foundation provides grants to help cover certain CF related medications and treatments. As of April 10, 2017 changes were made to how the fund is structured. There are now 2 funds that you can apply for:

- ◇ **Cystic Fibrosis Vitamins and Supplements Fund** can help with costs associated with nutritional supplements, tube feeding formulas and vitamins. This fund grants up to \$1,500 per year.
- ◇ **Cystic Fibrosis Treatments Fund** can help with costs associated with treatments and medications. This fund grants up to \$15,000 per year.

If you applied for a HealthWell Grant before April 10, 2017 you can continue to use your current funding until you are due to reapply.

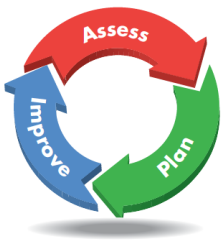
- ◇ **To qualify and apply:**
 - You must be treated for CF.
 - You must have insurance that covers medications.
 - Your income must fall within guidelines (Household income limit: 400-500% of federal poverty level. Adjusted for household size and high cost of living in particular cities/states).
 - By phone, call **1-800-675-8416.**
 - Online, go to www.healthwellfoundation.org

- ◇ **Helpful tips:**
 - A list of covered medications and nutritional products is available on the HealthWell Foundation website.
 - You must use your HealthWell Foundation grant every 120 days or the grant will close.
 - ◇ Contact your CF Social Worker or CF Dietitian if you have questions or would like to learn more.

~ Courtney Busby, R.D.



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



QUALITY IMPROVEMENT @ UNC'S ADULT CF CENTER

Your Quality Improvement Team continues to work hard behind the scenes to improve the quality of care for patients at our CF Center. Most of our time recently has been focused on developing the new Long Term Health Care Planning program (see page 1). In April, we held a half-day retreat to focus on the project and we look forward to rolling it out in the coming months. In addition to this project, we continue to monitor how often patients come to clinic and are working on reaching out and providing support to patients who need to be seen more often. We are also working on establishing a virtual support group for patients, and we will keep you updated as that begins.

As always, if you have questions or concerns for the QI team, email them to AdultCFNurse@unchealth.unc.edu.

~ Katie Reisinger, Adult CF Patient



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UNC Adult Cystic Fibrosis Center

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Social Worker: Marianne Buchanan Brooke Jones	P: 984-974-3385 marianne.buchanan@unchealth.unc.edu P: 984-974-5128 brooke.jones@unchealth.unc.edu
Pharmacist: Emily Wong	P: 984-974-0012 pulmonaryclinicpharm@unchealth.unc.edu

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

KEEP IN TOUCH