

Just In:

NACFC 2017 Highlights

The annual North American cystic fibrosis conference was held in November in Indianapolis. Nearly 5000 participants came together to discuss the latest research and evolving care practices. Of interest to everybody in the CF community is the rapidly progressing therapeutic pipeline. Once again this year, the significant progress being made to restore the function of CFTR (the CF protein) headlined many sessions. The results of two phase 3 trials (the final stage before FDA approval) that tested the safety and efficacy of tezacaftor/ivacaftor (Vertex Pharmaceuticals) were presented. In one trial, patients with two copies of the common mutation (F508del) were shown to have clinical improvement similar to, or better, than that previously seen with Orkambi, but without the troublesome side effects and drug interactions that hampers its use in some patients. In the second trial, patients with a partially functioning CFTR protein received either tezacaftor/ivacaftor or ivacaftor by itself. While both were beneficial, the combination drug approach proved to be superior to Ivacaftor by itself and patients experienced substantial improvements in lung function and symptoms. Currently, we await the FDA decision regarding approval of tezacaftor/ivacaftor, which is anticipated in late February, 2018.

Although earlier in development, the results of phase 2 trials of 3-drug CFTR modulator combinations were also discussed. These combinations are based upon the tezacaftor/ivacaftor backbone with a “next generation corrector” added on (also from Vertex Pharmameuticals). These trials showed very large improvements in lung function and symptoms, both in patients with two copies of the common mutation, and also in patients with only a single delF508 copy. We are very excited to enter phase 3 testing of selected 3-drug combinations this summer. We firmly believe this 3-drug approach, once approved, will bring dramatic improvements to more than 90% of our patients. We as a community are working as quickly as possible to make this happen. Importantly, multiple other companies are bringing competing CFTR modulators to trials at the present time, and this competition will ensure that our patients have access to the best therapies possible.

Despite the tremendous progress being made that we know will benefit most CF patients, it is clear that efforts are only intensifying to bring life-changing therapies to *every* CF patient, including those who might not benefit from traditional

CFTR modulators. Towards this end, therapies that don't rely upon a specific CFTR mutation are moving forward in the areas of ion transport modulation (e.g. inhibition of sodium channels and activation of non-CFTR channels), anti-inflammatories, novel antibiotics and mucolytics. Further, high-tech therapies that are aimed to either correct or replace the CFTR gene code, or the message that it produces, are aggressively being sought in the lab. These have the potential for helping patients with any mutation and could even bring a “one-time cure”. While these new approaches will take some time to develop, it is reassuring to know that the investment is being made today to make this possible in the future. The future is bright, indeed!

~ Scott Donaldson, M.D.



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NEW UNC-HILLSBOROUGH CAMPUS

Have you ever been sick and had to wait days, or more, for a bed in the hospital? Unfortunately, this has become a very common situation. As UNC Hospitals become busier and busier serving the residents of North Carolina, the availability of inpatient beds has become a significant issue to our adult patients with CF.

In response to this problem, UNC hospitals recently opened the UNC-Hillsborough campus. This brand new, small, patient-friendly facility contains state-of-the-art medical facilities and is staffed by UNC physicians. For more than six months we have been exploring and preparing for the possibility of caring for some adult CF patients in this new facility. As of January 2018, we are prepared to begin offering this option to our patients.

For those of you who have received their care in Chapel Hill for many years, this probably sounds a bit frightening. Believe me, our team shared your trepidation at the beginning of this process but are now convinced that we can provide care that is equal to that in our main hospital for properly selected patients. Here is a little bit about what to expect:

- 68 bed hospital, with strong radiology, PFT, respiratory therapy, and ICU capabilities
- Located 17 miles (26 minutes) north of the main UNC Hospital (near I-40 and I-85)
- Staffed by UNC Pulmonologists and UNC Hospitalists, currently without involvement of medical residents or students in training
- Seamless integration of medical records, labs and radiology studies between campuses
- Respiratory Therapy, Nutrition, Pharmacy, Social Work, and Nursing staff have received CF-specific training

During the initiation of this process, our plan will be to select patients who are clinically stable and have a straight-forward treatment plan that has been outlined by your CF team. Most patients will begin their hospitalization in Chapel Hill to ensure a good initial response to treatment before transitioning to the Hillsborough campus to complete their course of treatment. Close communication regarding your care between the teams at each campus will be maintained. Transportation between hospital campuses will be provided by UNC without cost to patients, and will be similar to transfers between hospital floors within the main hospital. Once again, we firmly believe that the care we deliver will not be compromised by this plan and we are committed to making it work by remaining very involved in your care during all phases of your hospitalization, including time spent at our Hillsborough campus. We are more than happy to discuss any questions you might have at your next clinic visit, and will formally seek feedback from all patients who get to experience this fresh, new facility this year.

DO YOU NEED A PRIMARY CARE PROVIDER, OTHER THAN YOUR CF DOCTOR?

Do you consider your pulmonologist to be your primary care provider (PCP)?

If you do, you are not alone. Many people feel that since they see their CF providers so often, that they don't need a PCP. However, there are many general medical issues that internists, family practice doctors, and other primary care providers (e.g. some gynecologists) are better prepared to manage. In addition, while the CF Clinic will routinely order tests that are specific to cystic fibrosis care, other screening tests are recommended for all adults and are likely to be missed without the help of a PCP. Seeing a PCP along with your pulmonology will help ensure you receive truly comprehensive care!

In the left-hand column of the table below are examples of screening tests your CF doctor routinely monitors. The right-hand column lists tests that, while important, are not specific to CF, and thus are not routinely monitored by your CF doctor. We ask that you discuss with your primary care provider how frequently you should undergo these tests.

Some Tests with Specific CF Recommendations	Some Tests Not Affected by CF
Vitamin levels yearly	HIV testing and other sexually transmitted infections
Colonoscopy every 5 years starting at age 40	Hepatitis testing
Oral Glucose Tolerance Test every year	Cholesterol checks
Bone density testing every 2 to 5 years	Mammograms (women only)
Liver Function Tests every year	Pap smears (women only)
Screening for depression and anxiety	PSA testing (men only)

If your primary care doctor has questions, we are happy to assist them. If you do not have a primary care doctor, but would like to get one, you can talk to a member of your CF Care Team about options in your area.

Long Term Health Care Planning Update

As you may have seen in our last newsletter, your CF team wants to offer our patients the opportunity to talk as a team about your long-term health care planning. The meetings are generally an hour long and can happen on the same day as your clinic visit. Your CF team will be there, along with other people who are important to you and who you decide to invite. Our first meeting took place in September. We've received positive feedback from patients, families, and team members alike. Here are some questions you may have about Long Term Health Care Planning:

Q: I don't like to think about what's going to happen when my health gets worse.

Why would I think about this in advance?

A: Our goal is to have a conversation that can give us a shared understanding of what matters most to you and your loved ones. This can make it easier to make decisions when the time comes. Talking about health care goals ahead of time can help lessen anxiety for patients, decrease burden for family, and ensure that health care wishes are followed. According to a survey done by the Conversation Project, 60% of people say that making sure their family is not burdened by tough decisions is extremely important. 56% of people have **not** communicated their wishes.

Q: What would we talk about in this meeting?

A: If you're interested in having a meeting, we will provide you with a booklet in advance that covers a range of topics that could be discussed, including general information of the course of CF over time, lung transplantation, symptom management, social supports, and documentation of your wishes related to your healthcare. You'll choose the topics that are most important to you to talk about with your family and team.

Q: When is the right time to schedule a meeting?

A: According to The Conversation Project, "It's always too soon. Until it's too late." It might not seem like it's time to think about your long-term health care, but we think it's best to have these conversations as early as possible, ideally while you're still healthy.

If you're interested in learning more about Long Term Health Care Planning Meetings, please contact your CF Social Worker, either Brooke Jones or Marianne Buchanan.

PERSONNEL UPDATES



Margret, our new CF Data Coordinator, graduated from UNC Chapel Hill in 2016 with a BS in Psychology and Neuroscience and a BA in Political Science. She worked as a research assistant in undergrad in the Cognition and Addiction Biopsychology lab and knew that she wanted to stay in research as a career. She has learned so much about Cystic Fibrosis since starting with the CF team, and has loved every minute of it! She is excited to have

the opportunity to learn even more and work hands on with patients who are helping find the cure for CF! Aside from work, Margret spends most of her time cooking or baking, and fostering dogs for Hope Animal Rescue.

CF FALL FUNDRAISER

The 1st Annual UNC Cystic Fibrosis Fall Fundraiser and Corn Hole Tournament was held on Sunday, November 12th at the Southern Village Market Green to raise awareness surrounding our Grocery Assistance Program. This program was piloted last fall and created in order to serve patients who are food insecure; defined as not having reliable access to an adequate supply of affordable, nutritious, food. The fundraiser was a great way to gather together as a community to support this program. There were approximately 100 family members, friends, and community members in attendance and we were able to raise almost \$4,000. This will aid in expanding the program for other patients who have a similar need in 2018. A sincere Thank You to everyone who supported this event! We are excited for next year!

CF VITAMINS

The 4 “fat-soluble” vitamins (A, D, E, & K) play important roles in the daily functioning of your body:

- Vitamin A – Helps your eyes see well, particularly at night. Food sources include tomatoes, carrots, sweet potatoes, and green leafy vegetables.
- Vitamin D – This vitamin is known by the names cholecalciferol (vitamin D3) and ergocalciferol (vitamin D2). Helps your body absorb calcium for strong bones, as well as plays an important part of your immune system. Food sources include eggs, fatty fish like salmon and tuna, and fortified milks.
- Vitamin E – Helps you fight infections, and helps red blood cells work. Food sources include nuts, vegetable oils, whole grain breads and cereals.
- Vitamin K – Helps your blood clot and is key in building strong bones. Food sources include vegetable oils and green leafy vegetables like spinach.

There are lots of choices for CF vitamins these days. They all provide higher amounts of the fat-soluble vitamins over standard multivitamins. They contain other “water-soluble” vitamins and the mineral zinc. They are also in an easier-to-absorb form for people with CF. The brands on the market currently are AquADEK, MVW Complete Formulation, Choiceful, Multivitamin ABDEK (H2-Pharma), and DEKAs. Most brands have liquid, chewable and softgel forms.

Brands vary slightly in content, but all can meet your CF needs when taken at the correct dose. Your doctor also checks the levels of vitamins in your blood to help assess whether your needs are met. Once brand, form, and dose have been chosen, be sure to *stick with it* and take it every day. It’s important to eat vitamin-rich foods in your diet AND take your CF vitamin every day.

The cost of these specialized vitamins can be significant and they are typically not covered by insurance plans. Check out the resources listed below that might help you obtain these vitamins for free. Ask your CF dietitian or other CF team member for help if needed.

- Creon’s Care Forward Program <https://www.creon.com/cfcareforward>
- Zenpep’s Live2Thrive Program <https://www.live2thrive.org/>
- Healthwell Foundation Grant for CF <https://www.healthwellfoundation.org/>

~ Original by Kimberly Stephenson, RD May 2017. Revised by Courtney Busby, RD November 2017

PATIENT ASSISTANCE PROGRAMS/RESOURCES

CF Foundation Compass

Personalized insurance, financial, and legal service
Call u844-COMPASS (844-266-7277) Monday through Friday, 9 a.m. until 7 p.m. ET, compass@cff.org.

HealthWell Foundation

<https://www.healthwellfoundation.org/fund/cystic-fibrosis-treatments-2/>

AbbVie : CREON® CF Care Forward Patient Support Program offers nutritional services to eligible patients, as well as financial and educational support for patients and families.

<http://www.creon.com/CFCareForward>
1-855-227-3493

Allergan: Zenpep Live 2 Thrive Offers copay assistance, free vitamins and supplements, and nutritional information for eligible patients.

<https://www.live2thrive.org>
1-888-936-7371

Chiesi USA : BETHKIS® (Tobramycin Inhalation Solution) and **PERTZYE®** (pancrelipase) 1-888-865-1222

<http://bethkis.com/support-services/>
<http://www.pertzyecf.com/patient/free-support-and-savings/>

Genentech, Inc.: Pulmozyme® Access Program

<http://www.genentechaccesssolutions.com/portal/site/AS/>
1-866-4-ACCESS

Gilead: Cayston® Access Program

<http://www.gilead.com/responsibility/us-patient-access/cayston%20access%20program>
call 1-877-7CAYSTON (877-722-9786)

Novartis Pharmaceuticals: Tobin Nebs and Podhaler Co-pay assistance program: 1-877-999-TOBI (8624).

<http://www.tobipodhaler.com/info/about/podcare-cf-patient-support.jsp>

PARI: Kitabis (Tobramycin nebs)

<http://kitabis.com/patient-access/>

Vertex Pharmaceuticals

Vertex GPS: Guidance & Patient Support (**Kalydeco®** or **ORKAMBI™**)
<http://www.vertexgps.com>
1-877-752-5933

Abbott Nutrition Patient Assistance Program Call 866-801-5657.

CF Peer Connect

CF Peer Connect is a peer mentoring program that enables one-to-one connections between people with CF. After you complete a mentor request, the CF Foundation will match you with an adult with CF who has experience with the topics you're looking to learn more about. You can connect with a peer mentor to talk about topics that are important to you, including:

- Dating and relationships
- Going to college
- Making work/career decisions, including going on disability
- Having a family, parenting, and infertility
- Getting diagnosed with CF as a teen or adult
- Living with CF-related diabetes
- Considering lung transplantation
- Managing post-transplant issues

Get Started in 4 Simple Steps

1. Complete an online request form at cff.org.
2. The CF Peer Connect team will schedule a call with you within one week to learn more about what you're looking for in a peer mentor.
3. The team will match you with a peer mentor who best fits your request.
4. Connect with your peer mentor over video, phone, email, or text.

If you have any questions about CF Peer Connect, contact peerconnect@cff.org.

*We now offer a local virtual support group, if you're interested in participating or would like further information please contact Brooke Jones brooke.jones@unhealth.unc.edu or Marianne Buchanan Marianne.buchanan@unhealth.unc.edu.



REMINDER:

FLU VACCINES ARE AVAILABLE IN CLINIC.

A Note From Your Pharmacist!

If you have commercial insurance (not Medicare, Medicaid or Tri-care), please check with your CF provider/team about helping you enroll in several copay assistance programs for CF medications (Orkambi®, Kalydeco®, Pulmozyme®, Tobi Podhaler®, Bethkis®, Kitabis®, Cayston®, and select pancreatic enzymes) and Healthwell (income-based). You may qualify for both!

For patients already enrolled with the Healthwell Grant and copay assistance programs, please remind your pharmacies to bill your insurance first, drug copay card second, and then your Healthwell CF Treatment Pharmacy Card.

For Medicare, Medicaid, and Tricare patients, you can still apply for Healthwell Grant Assistance. Don't forget to apply for both open grants: CF Treatment Fund and CF Vitamins and Supplements.



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Cystic Fibrosis
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KEEP IN TOUCH

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