

Cystic Fibrosis Center News

Cystic fibrosis liver disease: What is it and what are we doing about it?

— Zachary M. Sellers, MD, PhD

Cystic fibrosis (CF) affects the gastrointestinal tract in numerous ways (e.g., exocrine pancreatic insufficiency, constipation or distal intestinal obstruction syndrome, etc.), but I find that many families are surprised to hear that CF can involve the liver. While often touted as a “lung disease,” CF is actually a whole-body disease, with CFTR (the impaired protein in CF) being expressed in almost every tissue and organ of the body. That being said, the lungs and gastrointestinal system are generally the most affected. Each patient’s unique genetics (and possibly other unidentified factors) influences whether one has lung-predominant or GI-predominant CF.

In this article, I hope to help you learn more about liver involvement in CF, tell you what we are doing to detect and monitor CF liver disease (CFLD), review treatment options and discuss what the future may hold.

What causes liver disease in CF?

The long-standing thought on how CFLD develops is that it’s similar to what seems to occur in the lungs and pancreas in CF: thick secretions get trapped in the liver, causing inflammation and damage. The liver consists of liver cells (hepatocytes) and bile ducts. The hepatocytes make proteins, process drugs, and detoxify and secrete

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bile. As seen in Figure 1, bile is then transported out of the liver and into the gallbladder or intestines via the bile ducts. Bile is important for the digestion and absorption of dietary fat and vitamins, and CFLD is thought to originate in the bile ducts. CFTR is expressed in the bile ducts (not the hepatocytes). As bile is secreted into the bile ducts, CFTR adds chloride, bicarbonate and water to help create the right environment for bile duct flow and fat digestion in the intestines. When CFTR does not work properly, it is thought that the bile becomes thick and gets stuck within the bile ducts, leading to inflammation. Inflammation starts in the bile ducts and then spreads to the neighboring hepatocytes. Recent research also suggests that intestinal health and the bacterial flora in your intestines may also influence this process, but this idea is still being explored. When inflammation occurs in the liver, the liver can heal itself if the inflammation is minor and transient. However, if it is severe or persistent,

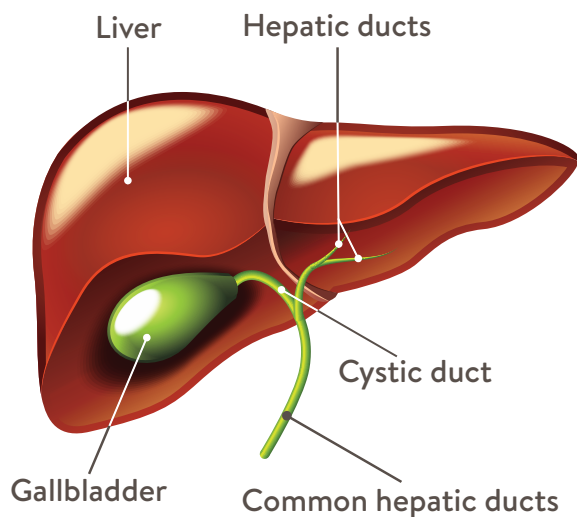


Figure 1

scarring (fibrosis) may occur, and without resolution, the inflammation and scarring can spread to involve the entire liver. This is similar to what happens with a cut on your skin. A scab will first develop, and if the cut is small, the skin will heal without a scar. But if the cut is larger, a scar may remain. CFLD first develops as liver fibrosis, which then becomes focal biliary cirrhosis (local disease), which then progresses to multilobular cirrhosis (entire liver).

When diagnosing a child with CFLD, I am often asked, “Is it something I did? Is it from all of the medications, or is it something I could have prevented?” While medications can cause liver damage separate from CFLD, CFLD occurs because CFTR is not doing its job effectively in the bile ducts. I can reassure parents that there is nothing they did to cause it. Like other problems related to CF, our job as providers, patients and families is to identify liver involvement in CF and help improve or prevent further disease.

Will I (or my child) develop liver disease?

CFLD does not occur in every CF patient, which is why many people don’t know about it. We are working to fully understand the different degrees of liver involvement among CF patients (i.e., how many have liver fibrosis only, how many have localized liver disease and how many have multilobular cirrhosis). Current research shows that about 5 to 10 percent of individuals with CF will develop cirrhosis. However, the number of CF patients that have fibrosis may be as high as 50 percent. Ongoing research is being conducted here at the Stanford Children’s CF Center and elsewhere to understand which CF patients will develop liver fibrosis, and more importantly, who is at risk for progressing from fibrosis to cirrhosis.

How do you diagnose CF liver disease?

The current recommendations from the Cystic Fibrosis Foundation are to check liver function tests (LFTs) annually to screen for CFLD. LFTs show levels of AST and ALT, which are enzymes released by hepatocytes, and GGT, which is released by the bile ducts. However, our research (and that done by others) has found that LFTs may be abnormal when there is no CFLD, and CFLD may be present when LFTs appear normal. At the Stanford Children’s Cystic Fibrosis Center, we have been working on identifying patterns in LFTs that may be more reliable in detecting CFLD. We also recently introduced a new component to our CFLD screening protocol – abdominal ultrasounds – to try and improve our detection of CFLD. Thus, for all of our CF patients that are at least 6 years old and are exocrine pancreatic insufficient (i.e., needing pancreatic enzymes), an abdominal ultrasound will be done to examine the liver. This doesn’t automatically mean your child has CFLD, but it is intended to screen for it. Right now, this is being done every

year, but depending on the findings, it may be done less or more frequently. Ultrasound can identify the nodularity that occurs in the liver when there are areas of cirrhosis.

This helps us to identify local cirrhosis when LFTs may still be normal. In addition to a standard ultrasound, at Lucile Packard Children's Hospital Stanford, we have the ability to use an advanced ultrasound technique, called elastography, to measure the stiffness of the liver. Healthy livers are soft; those with fibrosis or cirrhosis are stiffer. In other liver diseases, this test is useful to detect fibrosis, prior to cirrhosis. We are adapting this technique for CF so that we can identify early signs of fibrosis before cirrhosis develops. Our hope is that the addition of these tests will help us identify CFLD earlier and more accurately, and will lead to improvements in the liver and gastrointestinal health of our CF patients. It is for this reason that you may be asked to get your ultrasound done at LPCHS, even if your labs and clinic visits are done elsewhere. Ultrasound elastography is done using a similar method as regular ultrasounds, with most patients not even knowing the difference.

If I am diagnosed with CFLD, what next?

If you have lab results or imaging tests that indicate CFLD, you will likely be referred to see a gastroenterologist. At Stanford, our Division of Pediatric Gastroenterology works closely with our pediatric pulmonologists. The gastroenterologist will discuss how well the liver is working and if any additional blood or imaging tests are necessary. They will make sure there are no other medical causes beyond CF that may result in liver disease. They may also discuss new medicines for you.

Are there any treatments for CFLD?

Once we detect CFLD, what can we do about it? Our most important task is to watch out for complications from advanced cirrhosis (multilobular cirrhosis). This can cause what's known as "portal hypertension," where blood

flow through the liver is impaired because the liver is too stiff, which causes the spleen and blood vessels in the gastrointestinal tract to enlarge (i.e., esophageal varices). This can lead to gastrointestinal bleeding that cannot resolve easily. This can, in fact, be life threatening. If someone develops this degree of liver disease, endoscopic or surgical options (even liver transplant) may be necessary. Generally, this occurs in a minority of patients. However, we need to monitor for it in all patients with CFLD. Our goal is to identify CFLD early and prevent progression to cirrhosis. It is our hope that the CFTR modulators and correctors will not only prevent worsening lung disease, but that they will also prevent liver disease. Stay tuned for more on that theory. In the meantime, there is a medicine called ursodiol that improves bile flow and has been used in patients with CFLD. Results are mixed as to whether it is beneficial for CFLD. However, because it is a generally safe medicine, many people are using it in the hopes that early intervention with this medicine may be helpful. Should there be concern for CFLD, your gastroenterologist can discuss if this medicine may be helpful. As new medicines are developed for CF and other liver diseases, we hope to apply these to our patients to improve the health of their livers.

What does the future hold?

The future for CF is bright. Certainly the new CFTR modulators and correctors have brought a new energy to CF care. Many new treatments are in development and in clinical trials. For CFLD, we are actively working to improve diagnosis through optimizing the use of LFTs and applying new imaging tools to CF. While being diagnosed with another CF-related problem can be daunting, we hope our efforts will actually make the liver healthier and lead to overall improvements in the health of our CF patients. As always, our goal is to work with our patients and families to keep them healthy while minimizing the burden of health care on them. We remain committed to working with you as a team.

If you have any questions about CFLD or other gastrointestinal symptoms associated with CF, email Dr. Sellers at zsellers@stanford.edu. Dr. Sellers is also available for GI consultation for children with CF at Stanford Children's Health in Palo Alto.

The 2017 Therapeutic Development Network spring meeting

— Sean Ryan, RRT

This year's 2017 Therapeutic Development Network (TDN) spring meeting was held on April 3 and 4 in Nashville, TN. A total of 466 members from 89 sites from across the United States gathered at the Sheraton Nashville Downtown hotel to discuss ongoing and upcoming clinical trials and to attend breakout sessions on various topics centered on education and training to improve clinical research expertise. There were a range of breakout choices, including "History of the TDN," "Ensuring research strategies align with cystic fibrosis community priorities," "Managing multiple projects," "Child life services in clinical research," "Practices and traits of successful coordinators," and "Hidden gems in CF CRNet" (a research website) led by our very own Zoe Davies.

During the opening plenary session, members learned about the various upcoming studies. It was especially exciting to hear about the next generation of CFTR modulator studies. The hope that these novel drugs will provide to the CF community is remarkable and represents a giant leap forward in drug development. Additionally, 82 new research coordinators had the opportunity to attend the one-day clinical research bootcamp that was held the day prior to the meeting. The bootcamp touched on many subjects and was designed to provide a CF-specific overview and in-depth training to those transitioning into clinical research. Some areas that were covered include the informed consent process, the difference between researcher and coordinator, study budgeting and negotiation, and optimizing your study organization. Daniel Alvarez, our newest research coordinator, was among the attendees. I, myself, attended bootcamp just two years ago, and I am amazed to reflect on how much I have learned

from these meetings and the Stanford research team. Indeed, I've learned enough to be invited to actually teach a breakout session, "Sputum induction training," alongside the world-renowned Sputum Queen herself, Colleen Dunn!

The closing plenary session hit a more personal note. Those living with cystic fibrosis gave voice to their own experiences participating in clinical research. It was inspiring to hear of the commitment and dedication these individuals show on a daily basis to finding new treatments and, ultimately, a cure. It was especially gratifying to know that the work we do is continuing to benefit these individuals.

Not only is the TDN spring meeting a place for learning and education, but it is also a place where we can network with others from different sites. It is an opportunity for us to share our stories, experiences, and successes along with our mistakes and how we learned from them. Sharing these ideas, strategies and tools with one another plays an important role in helping us evolve and become better clinical researchers. We also get to have fun together, too, and this year's social event included a photo booth, dinner and dancing. The TDN always does a great job with these events, and this year they invited a professional dancer to teach all of us the latest country line dances! It was a nice way to let loose after a full day of breakout sessions.

To conclude, the spring meeting was very informative and enjoyable. It was nice to see familiar faces from around the country and to meet some new ones. Next year's TDN spring meeting will be held in Philadelphia, PA. If you would like to know more about the spring meeting or Stanford's ongoing clinical trials, please call us at (650) 736-0388.

Spreading CF awareness in Sacramento

On May 8, 2017, Cystic Fibrosis Research Inc. took their advocacy to another level. Ten people — including some of the adult CF clinic staff — traveled to the state senate in Sacramento, California, to address some of the main concerns of the CF community. Awareness is the key to research funding and, hopefully, the eradication of this orphan disease.

Consider these facts:

- Approximately 30,000 children and adults in the United States have CF.
- More than 10 million people are symptomless carriers of the defective CF gene.
- People with this disease are living into their 30s, 40s and beyond.
- The life expectancy for CF patients has doubled in the last 30 years, but CF patients still lose their lives every day.
- CFRI will not rest until we find a cure and CF stands for “cure found.”

On that day in Sacramento, the people most affected by CF met with their representatives to voice their frustrations about dealing with insurance companies, government agencies and pharmaceutical companies. The group was given a standing ovation in the senate chamber because of their commitment to eradicating this insidious disease, and May was declared National Cystic Fibrosis Awareness Month!



Recipe

— Julie Matel, RD

Tropical Banana Shake

(480 calories)

Ingredients:

- 2 ripe bananas
- ½ c. canned peaches
- 1 c. cup mango or guava nectar
- ½ c. whole milk
- 3 ice cubes



Instructions:

1. Wash all of the fruit you'll be using before getting started.
2. Prepare your ingredients, and place them in blender.
3. Blend the ingredients together to the desired consistency, adding additional liquid if needed.

Tip: Frozen fruit can be used for a thicker smoothie or shake.

Managing cystic fibrosis–related diabetes during hospitalization

— *Carole Nakamura, MSN, RN, PCCN, CMSRN*

Cystic fibrosis–related diabetes (CFRD) is very common and affects about 20 percent of adolescents and 40 to 50 percent of adults with CF. The presence of CFRD is linked to worsening lung function and nutritional status, regardless of age. CFRD shares features with both type 1 and type 2 diabetes, but it is different. It is primarily caused by insulin deficiency that is most likely due to scarred pancreas tissue and the subsequent decrease in insulin-producing cells and insulin secretion. Studies also show fluctuating levels of insulin resistance in those with CFRD, especially during acute and chronic illness. Insulin resistance may worsen with acute pulmonary exacerbation, chronic severe lung disease and glucocorticoid therapy (i.e., prednisone). You may require more insulin doses during your hospital stay.

As an outpatient, you will be screened regularly to diagnose CFRD, following the Cystic Fibrosis Foundation guidelines. A diagnosis of CFRD may be made while you are hospitalized if you have any of these blood glucose results that persist for > 48 hours: fasting serum glucose is ≥ 126 mg/dl, two-hour blood glucose test after a meal is ≥ 200 mg/dl, or the mid- or after-tube feeding blood glucose test is ≥ 200 mg/dl.

Insulin is the standard therapy for maintaining blood glucose control in CFRD. The goal for treating CFRD is to keep blood glucose at normal levels to preserve lung function and to optimize nutritional status. CFRD is managed by the CF multidisciplinary team with expert consultation from endocrine doctors and diabetes nurse-educators that specialize in CFRD. Individuals with CFRD still need to maintain a very high-calorie and high-protein diet to meet their metabolic needs for weight maintenance or weight gain. You may be taught how to do carbohydrate (CHO) counting with an individualized insulin-to-CHO ratio (e.g., 1 unit of insulin for each 15 grams of CHO eaten) to calculate insulin dosing.

Usually a “basal-bolus” insulin regimen will be prescribed, with a single daily injection of a “basal” or continuous, non-peaking insulin (e.g., glargine, detemir) plus multiple injections of rapid-acting insulin (e.g., lispro, aspart) to cover meals and snacks. Use of your own subcutaneous insulin pump is permitted in the hospital if prescribed by your provider. We do have a specific hospital protocol to follow if you want to continue to use your own insulin pump.

The nurses will be checking your fingerstick blood glucose (FSBG) tests in the early morning, before meals, two hours after meals, at bedtime and sometimes at 2 a.m. to check for low blood-sugar levels (hypoglycemia). The timing of the FSBG tests is important for accuracy and dosing of insulin, so please inform your nurse when you are going to eat your meals and snacks. You may check your blood sugar with your own glucometer, but we must use the hospital glucometer to officially record the glucose results in your hospital record. Usually, we can “share” that fingerstick drop of blood to test on both glucometers if you are curious to see the difference in glucometer results.

Hypoglycemic (low blood sugar) reactions are a serious complication that can occur with insulin therapy. It is important to consume the carbohydrates that were counted and dosed with insulin because the insulin will work to bring down the blood glucose level. Hypoglycemia is a level < 70mg/dl and severe hypoglycemia is < 50. If your appetite is unpredictable, we can give the CHO-insulin dose after you eat the meal or snack.

Any combination of being hospitalized with a worsening infection, receiving new medications, experiencing increased stress, having surgery or procedures or having other acute medical problems can cause your blood glucose levels to fluctuate, usually causing higher levels (hyperglycemia). Don't be dismayed — your team will work with you to manage your CFRD.

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Back-to-school tips from the Cystic Fibrosis Parent Advisory Council

— *Kirsten McGowan*

The CF Parent Advisory Council seeks to address the needs of all families seen at the Stanford Children's Health CF clinics. As we round out the summer, we thought it would be a good time to share some tips for getting ready to go back to school.

1. Hot, Hot, Hot!

- a. People with CF lose more salt through sweat and dehydrate more easily than people who don't have CF. Make sure your child consumes extra water, fluids and salt to stay hydrated and safe. Some good salty options are:
 - i. Salted nuts
 - ii. Potato chips
 - iii. Pretzels
- b. Watch out for medications
 - i. Many meds are temperature sensitive (like enzymes!), so don't keep them in risky areas such as pockets or cars or leave them in direct sunlight.

2. Educate the educators

- a. Starting a new school year can be daunting, so it is well worth taking the time to speak to your child's teacher and school nurse about your child's needs.
- b. Meeting with school staff before school starts ensures everyone is up to date and minimizes surprises.

3. Update paperwork

- a. Now's the time to review IEPs, 504s and health plans. Update the forms with new medications, changes in your child's care plan or other accommodations.
- b. Consider asking your child's teacher if you can speak with or write a short letter or email to the parents of the other children in your child's class about the basics of CF so the entire class can be supportive.

4. Find your rhythm

- a. Routine is so important and can make life easier once school starts. Even during the summer, it is good to maintain routines so your child doesn't get out of the habit by the time school starts again. Getting your child back into a regular rhythm before school starts can help ensure those first few weeks of school are not as difficult.

If you have input for the CF Parent Advisory Council, please email **Kirsten McGowan** or **Amy Baugh** at kmcgowan@stanfordchildrens.org or abaugh@stanfordchildrens.org.



Pediatric CF Center Update

— Mary Helmers, RN

Some helpful tips

Did you know that you can get assistance with your PG&E bill? PG&E forms for medical equipment and devices can be found on the PG&E website under Medical Baseline Allowance Application for Medical Baseline Enrollment and Recertification. All you need to do is print the form and fill it out, including all your medical devices (e.g., nebulizer/compressor, if you use oxygen, CPAP or BIPAP). Then, bring the form with you to your next CF clinic visit, and your provider will sign it. Finally, mail the completed form to PG&E.

Our onsite urgent care clinic is located on the first floor of 730 Welch Road, across from the clinical lab.

Hours of operation:

Monday – Friday, 5:30 p.m. – 9:30 p.m.

Weekends and Holidays, 10:00 a.m. – 4:00 p.m.

If your child is sick after clinic hours and you would like your child assessed, you can use the urgent care clinic. The clinic is staffed with medical personnel and providers.

MyChart (secure electronic correspondence)

If you have not signed up already, please sign up for **MyChart at your next clinic visit**.

MyChart is a secure way to communicate with your provider and CF care team. The CF care team cannot respond to patient or parent emails in MyChart, since it is not a secure site. Please note that any email sent to the team will be responded to with a phone call. We do not always check emails on a daily basis. If you or your child has a clinical need or question, please call the CF RN line at (650) 736-1359.

It takes only a minute to sign up. At your next clinic visit, one of the front desk staff will be happy to assist you with sign up.

CF Passport

Parents and patients: please remember to carry your child's **CF PASSPORT** in your wallet. The CF Clinic sent the PASSPORTS out in the mail to each family.

If for some reason you tossed it or did not receive it, please ask for one when you come to your next clinic appointment. We now have them in English and Spanish.

Helpful reminders

To help expedite your clinic visit, please remember to bring your CF binder with you to the clinic, along with your most recent CF action plan.

Prescriptions: Just a reminder that your prescription request can take up to 72 hours to be processed. This has always been our policy, although we strive to turn them around sooner. Please keep in mind that even after we send the scrip to the pharmacy, it can still take another 48 to 72 hours for the pharmacy to process it (especially mail-order pharmacies). It is important for you to stay on top of your refills and request them at least one week before you are due to run out.

Helpful hints for requesting refills:

- Call your pharmacy first to find out if you have refills.
- If you have a refill, great! Then they will process it.
- If you have no refills, your pharmacy should call us.

Remember: We cannot guarantee your request will be filled the same day or within 24 hours.

CYSTIC FIBROSIS PASSPORT

- Please escort me to a private room
- Please follow contact/droplet precautions (see CF Isolation Policy)
- Gown, mask, gloves for all health care providers
- Clean all surfaces after patient contact
- Please remember to use good hand washing/gel/foam cleanser before and after patient contact

CF Passport sample

Wearable sweat sensor can diagnose cystic fibrosis, study finds

A wristband-like wearable sweat sensor could transform diagnostics and drug evaluation for cystic fibrosis, according to a trial led by members of the CF Center and researchers at Stanford University School of Medicine and UC Berkeley.

The system of flexible sensors and microprocessors sticks to the skin, stimulates the sweat glands and then detects the presence of different molecules and ions based on their electrical signals. The more chloride in the sweat, for example, the more electrical voltage is generated at the sensor's surface. The team used the sensor to detect chloride ion levels — high levels are an indicator of cystic fibrosis.

Current research studies

Vertex 661-110 — Open label extension study for subjects who participated in the Vertex 661-103 and 661-108 study protocols

OPTIMIZE — For treatment of newly acquired pseudomonas

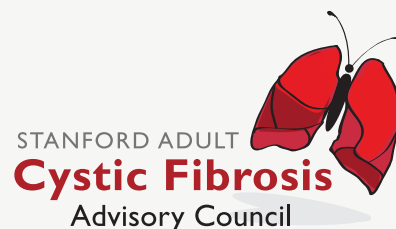
Prospect — Observational study for healthy people without CF and CF patients; enrolling now

Vertex 809-115 — Phase 3 study for children 2 to 5 years of age with the F508 mutations

Vertex 770-124 — Phase 3 study for children less than 2 years of age who have a gating mutation

Vertex 440-101 — Phase 2 study for patients 12 years old and older looking at the next-generation potentiator/corrector combination

Concert — Phase 2, open label comparator for patients with gating mutations



Stanford Adult Cystic Fibrosis Advisory Council

— *Brian Eddy*

It has been another interesting and productive quarter for the adult advisory council here at Stanford. We're excited to announce our first council-driven research study! Six volunteers, primarily council members, have started monitoring their FEV1 scores at home using small, handheld measuring devices, which have been donated by either the manufacturer or the Stanford CF research team. The goal of this first phase is to see what value the patients see in capturing this type of information for their own use. Our primary concern is detecting and monitoring a change in FEV1 score, not the actual values.

Potential benefits include the early detection of a potential health issue and tracking improvement due to a change in treatment plan. Similar studies are underway in other parts of the United States, so it's clear that others see the same potential here that we do. We look forward to completing this study and will continue to look for ways to contribute to the adult CF health care experience at Stanford.

If you would like information about the council or are interested in participating, please contact us at stanfordcfac@gmail.com.

Clinical Trial Finder:

A new tool to help staff, patients and families find clinical trials

— Zoe Davies PNP, CCRC

The CF research coordinators at Stanford often hear from patients and families about the frustration of not being adequately informed of ongoing and upcoming clinical studies and whether or not they qualify to participate in them. So, we are excited to let you know that the Cystic Fibrosis Foundation, in partnership with the Therapeutics Development Network, has developed a new Clinical Trial Finder tool. It was made available to everyone in the CF community on July 27, 2016.

This tool is designed to help staff and patients track down studies that are happening across the country and find out more information about drugs that are currently in

development. It lets you use search and filter options to specifically tailor results for individuals (e.g., by zipcode, genotype, age, and FEV1).

Users can also sign up for email alerts that will notify them when there are new trials in their region or when new study results become available.

Where can you find this tool? Go to **cff.org**, click on the Research tab and the Clinical Trial Finder tool can be found under the “Developing New Treatments” header (Image 1).

Also, remember you can always call the Stanford Research office at (650) 736-0388 if you have any questions or concerns pertaining to CF research.

CLINICAL TRIAL FINDER
Be a part of the movement transforming the future of cystic fibrosis treatment.

Search (Protocol, drug name, CFTR mutation, etc.)

Enter keyword

Clear search

Filter
Tailor your results by using the filters below.

STUDY STATUS

- Enrolling ☐
- Closed to enrollment ☐
- Completed with results ☐

Clinical trials are sorted below by therapeutic approach. Use the search and filter options to tailor the results. [Learn more about therapeutic approaches](#)

Showing 1-7 of 125 studies

RESTORE CFTR FUNCTION | ENROLLING

Phase 3 study of ivacaftor in babies who have a CFTR gating mutation | Vertex VX15-770-124

This two-part, open-label study is taking place at multiple care centers across the U.S. It will look at the safety and effectiveness of ivacaftor, as well as how the body processes the drug, in babies who have a CFTR gating mutation.

AGE	MUTATION(S)	FEV1% PREDICTED	NUMBER OF VISITS	LENGTH OF PARTICIPATION
0 Months to 24 Months	One Copy F508del or No Copies F508del	No FEV1 Limit	8	24 weeks

MUCOCILIARY CLEARANCE | ENROLLING

Cystic Fibrosis Foundation clinical trial finder tool

Adult CF Center Update: Traveling with cystic fibrosis

— Julian Liang, RN, BSN, and Anastasia Kaiser, MSW

With long security lines, delays, unpredictable weather and tiny seats, air travel can be challenging and full of headaches. Unfortunately, traveling with CF can add complications to an already frustrating experience. With preparation and awareness, it is possible to avoid some of the hassles that a person living with CF might face. Please know that our care team supports your desire to travel, and we are here to help. The Cystic Fibrosis Foundation also offers helpful guidelines for traveling, and we encourage you to visit their website (cff.org). People living with CF often ask the treatment team about the best way to prepare for travel, and so we thought we'd share a bit of advice.

To have the most stress-free travel experience, please contact your CF center social worker at least two weeks before your trip for a travel letter. This letter explains any medical needs and can help certify and explain why you need to have your vest and medications carried onto the flight. The team advises that necessary medications, your vest and other expensive medical equipment is always carried onto the plane and never in checked baggage. The key is to plan and coordinate your documents, medications and supplies in advance so that you're not scrambling at the last minute.

Please consider the differences between an international or domestic trip, and the duration of time spent abroad. If you're traveling for a long period of time, research local CF centers or pulmonologists who may be able to help you, and don't forget to alert your local center of the time and duration of your journey. It might be hard to find CF providers in remote areas. Please check in with your primary care provider on whether your destination warrants getting any vaccinations or special medications beforehand. When you return, please let the CF team know if you have taken any oral antibiotics that were prescribed for travel.

For any oxygen needs, please give extra attention to each airline's specific list of approved products. If you're already on portable oxygen, altitude simulation tests may be required to determine the amount of oxygen needed in flight. This needs to be scheduled in advance with the PFT

lab by calling (650) 723-6371. If you're already on oxygen, please contact your provider to set up oxygen needed for flights. We want to make sure you have enough battery packs and tanks for your entire trip! Oxygen rentals are available through companies such as Oxygentogo.com. Please know that it is your responsibility to rent, and it may not be covered by insurance. You can always call one of our respiratory therapists (available Tuesday through Friday) at (650) 736-8892 for questions and clarification.

Lastly, please remember to stay hydrated and take infection-control precautions seriously (hand washing, mask wearing). Please remember to consider how your medications may interact with alcohol. CFF also advises that people with CF avoid certain risky activities, including bungee jumping, scuba diving and hot tubbing. Most importantly, have a wonderful trip, and embrace the adventure!

*Managing cystic fibrosis,
continued from page 6*

While hospitalized, feel free to ask for our diabetes education folder, which has helpful reading material. Also, feel free to speak to a diabetes nurse educator or CF dietitian and to view basic diabetes education videos on your Skylight TV in your room.

References:

- Kelly, A., and Moran, A. (2013). "Update on cystic fibrosis-related diabetes." *Journal of Cystic Fibrosis*, 12:318–331.
- Moran, A., et.al. (2010). "Clinical care guidelines for cystic fibrosis-related diabetes: A position statement of the American Diabetes Association and a clinical practice guideline of the Cystic Fibrosis Foundation, endorsed by the Pediatric Endocrine Society." *Diabetes Care*, 33:2697–2708.
- O'Riordan, S.M., Robinson, P.D., Donaghue, K.C., and Moran, A. (2008). "Management of cystic fibrosis-related diabetes." *Pediatric Diabetes*, 9:338–344.

Cystic Fibrosis Center at Stanford

Pediatric providers at Lucile Packard Children's Hospital Stanford

Pediatric CF Center Director: Carlos Milla, MD

Providers: Sumit Bhargava, MD; My My Buu, MD; Carol Conrad, MD;
David Cornfield, MD; Richard Moss, MD; Terry Robinson, MD; Michael
Tracy, MD; Jacquelyn Zirbes, DNP, RN, CPNP

Clinic scheduling	(650) 724-4788
Clinic and prescription refill fax	(650) 497-8791
Erica Oliva, patient services coordinator	(650) 498-2655
Mary Helmers, nurse coordinator	(650) 736-1359
Liz Beken, CF clinic nurse	(650) 736-1359
Candice Middleton, respiratory therapist	(650) 724-0206
Julie Matel, nutritionist and dietitian	(650) 736-2128
Sruthi Veeravalli, social work	(650) 736-1905
Jacquelyn Zirbes, newborn screening coordinator	(650) 721-1132

Urgent issues

Monday – Friday, 8:00 a.m. – 4:00 p.m.

Contact the nurse coordinator at (650) 736-1359.

After-hours and weekends, call the main hospital number,
(650) 497-8000, to ask for the on-call pulmonary doctor.

Research

Colleen Dunn, Zoe Davies, Sean Ryan,

Wendy Valencia (650) 736-0388

Adult providers at Stanford

Adult CF Center director: Paul Mohabir, MD

Providers: Laveena Chhatwani, MD; Gundeep Dhillon, MD; Jennifer
Cannon, NP; Erika Rad, NP; Meredith Wiltse, NP

Backup providers: Kelly Johnson, NP; Laura Starr, NP; Puja Sarna, NP;
Glenna Monk, NP

Adult clinic scheduling	(650) 736-5400
Adult CF Center fax	(650) 723-3106
Nurse coordinators	(650) 498-6840
Patient last name A-K: Julian Liang, RN, BS	
Patient last name L-Z: Ronni Wetmore, RN, MS	
Respiratory therapy	(650) 736-8892
Carol Power, RCP; Gauri Pendharkar, RCP	
Registered dietitian	(650) 529-5952
Michelle Stroebe, MS, RD	
Social work	
Meg Dvorak, LCSW	(650) 518-9976
Anastasia Kaiser, MSW	(650) 444-6512
Mental health coordinator: Liza Sher, MD	

Urgent issues

Monday – Friday, 8:00 a.m. – 5:00 p.m.

Call the nurse coordinator at (650) 498-6840.

Monday – Sunday, 5:00 p.m. – 7:00 a.m.

Call (650) 723-4000 and ask for the on-call pulmonary fellow.

Saturday – Sunday, 7:00 a.m. – 5:00 p.m.

Call (650) 723-4000 and ask for the Adult CF ghost pager.

Visit our website at cfcenter.stanford.edu for more information about our center and CF.

To subscribe to this newsletter, please contact Cathy Hernandez by phone at (650) 724-3474 or by email at cathyh1@stanford.edu.

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