Summer 2023

Stanford MEDICINE
Children's Health

Cystic Fibrosis Center **News**

25th Anniversary—The Cystic Fibrosis Therapeutics Development Network

—Carlos Milla, MD

This year we celebrate the 25th anniversary of the Cystic Fibrosis Therapeutics Development Network (or CF TDN for short). The CF TDN was established in 1998 to promote quality, safety, and efficiency in CF clinical trials. At the time of the network's inception, it was clear that to be able to succeed in the development of novel therapies to benefit CF patients, we needed to have in place a highly coordinated and standardized research process focused on CF. This was not only to have a more efficient research process but also to make CF drug development attractive for industry partners.

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CYSTIC FIBROSIS FOUNDATION
THERAPEUTICS
DEVELOPMENT NETWORK

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Since its founding, the CF TDN has conducted more than 150 clinical studies for CF in a wide range of therapeutic areas, including CFTR modulators, anti-infectives, anti-inflammatories, nutritional therapies, airway surface liquid hydrators, and gene therapies. Many of these studies eventually led to drug approvals, as well as extending the indications for many therapies to our youngest CF patients. In addition, a tremendous amount of new knowledge has been generated, with CF TDN studies resulting in more than 200 publications in the medical literature. As it stands today, the CF TDN is the largest cystic fibrosis clinical trials network in the world and among the most successful research networks dedicated to a specific condition. The success of the CF TDN has not only helped make new therapies available to people with CF but also allowed the CF TDN to serve as a role model for many other clinical research networks focused on genetic conditions.

The CF TDN functions as a nonprofit, self-governed network of specialized clinical research centers, core laboratories, interpretation centers, and a coordinating center, which collaborate to develop and conduct clinical trials involving new therapeutic agents for CF, as well as clinical research to identify novel outcomes for future studies. The network's clinical research centers, called Therapeutic Development Centers (TDCs), are selected through a rigorous competitive process. The Stanford Cystic Fibrosis Center has been a proud member of the CF TDN since its early days. Center staff have not only been active participants in the function of the CF TDN but also made numerous contributions to the standardization of processes and procedures that are now in common use across the network. The Stanford Cystic Fibrosis Center has a longstanding tradition of productive collaborations between both basic and clinical science investigators, and in 2009 we were one of a few centers to receive a designation as a Translational CF TDN Center. By engaging our colleagues from across disciplines in the basic sciences, clinical sciences, and bioinformatics fields at Stanford, we have established the most successful collaborative, cohesive effort in CF translational research in California. Our strong institutional support and, as important, the support from our patients, families, and community at large, provide to us the impetus to continue with our work on the path to the discovery of the cure for CF.

We have and will continue to provide strong leadership in clinical trials design and conduct, novel outcomes measurements development, identification of novel targets for CF therapeutics, and training the future generations of CF investigators and research team members. In recognition of our expertise, we were recently tapped to be the Lead Site for the Regional Cooperative initiative of the CF TDN. This will help us establish a regional collaborative of centers in Northern California that will focus on increasing access to clinical trials and enhancing the research experience for patients and families across the region. We very much look forward to our continued work with our patients, families, and CF community at large to eventually reach our goal of providing a cure for all people with CF.

Everything You Need to Know About Mental Health Therapy

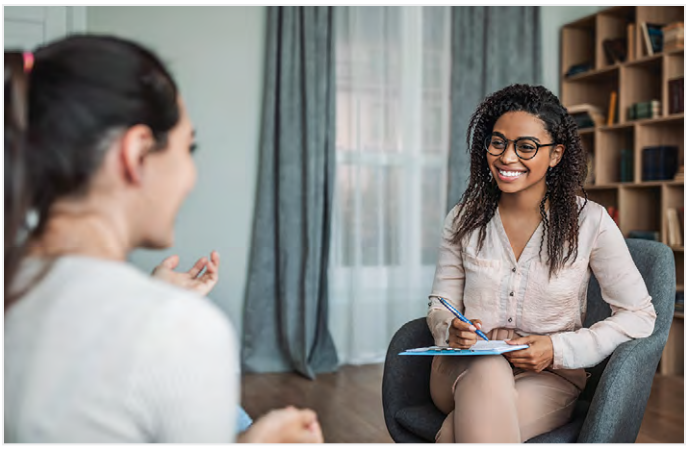
—Yelizaveta Sher, MD

People living with cystic fibrosis (CF) experience a two to three times greater risk of having depression or anxiety. They also have higher rates of attention deficit hyperactivity disorder (ADHD). And although great medical breakthroughs, in particular highly effective modulators, are changing the landscape of living with CF, it is still a chronic medical condition that adds stress to the already-existing everyday stress of life. Psychotherapy is one effective way to help address and cope with these psychological challenges.

So what is therapy? How does it work? How do you find it? And how do you know if your therapist is the right fit for you?

Psychotherapy, or talk therapy, or simply therapy, is a type of psychological treatment to help address a variety of mental health challenges, such as depression and anxiety, and to learn how to more effectively cope with the stress of navigating life stressors, including living with a chronic medical condition. In fact, the CF Foundation recommends therapy as the first-line treatment for mild to moderate depression and anxiety, when it is available. Therapy can of course be combined with psychotropic medications, when needed, and at times this combination is the most effective treatment for a particular condition, as compared with therapy or medications alone.

Psychotherapy utilizes a partnership between a patient and a mental health professional and builds on this foundation to help the patient identify problematic thought patterns and behaviors, work on gaining insights into their origin and function, reframe them, develop healthier behavioral patterns, increase distress tolerance, and improve quality of life overall and decrease burdensome psychological symptoms.



There are many kinds of therapies, based on different ideologies and skill sets. At the end of the day, they have overlapping features, and many can provide relief to a patient with a certain challenge. There are times when a therapist will use a very particular type of therapy to work on a certain mental health challenge—for example, using exposure response prevention (ERP) for treatment of obsessive-compulsive disorder (OCD). There are other times when many approaches can work, and/or the therapist combines several different techniques from different modalities.

Some of the most frequently known psychotherapies in the CF community are evidence-based and are cognitive behavioral therapy (CBT) and acceptance commitment therapy (ACT). CBT is based on the idea that our thoughts or cognitions, our feelings or emotions, our physical functioning, and our behaviors are interconnected. Patients learn

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how to become more aware and identify these connections and to develop a variety of skills to cope with painful emotions. Some of the tools include behavioral activation, relaxation practices, and reframing of cognitive distortions. Behavioral activation addresses the behavior of shutting down or isolating that is common in depression. This tool allows the patient to recognize the harmful patterns and to create a strategy to engage in small behaviors that can increase joy, meaning, and positive emotions during difficult times. Relaxation practices allow the patient to address uncomfortable sensations that accompany anxiety and stress and include guided imagery and progressive muscle relaxation. Finally, reframing, a cognitive tool, involves first listening to the internal dialogue that accompanies strong emotions, identifying negative automatic thoughts (thoughts that are usually extreme or judgmental), naming distortions, and finding an alternative way to think about the challenges.

Another approach is ACT, which is therapy is based on mindfulness and teaches one to accept uncomfortable thoughts and emotions without judging or changing them. It invites you to push through these difficult experiences, stay present in the moment, be resilient, and work toward your identified goals and values.

Other therapy modalities include psychodynamic psychotherapy (evaluating how one's past influences their current way of thinking and being), dialectical behavior therapy (DBT) (based on CBT and aimed to help the patient tolerate distress of particularly strong emotions as well as cultivate interpersonal effectiveness), and interpersonal psychotherapy (looks at transitions and role changes in life).

Who is a therapist/psychotherapist? A psychotherapist is a licensed mental health professional trained to provide psychological therapies. Psychotherapists can be psychiatrists (doctors who further specialize in mental health), psychologists (professionals who undergo graduate school training), social workers, and marriage and family therapists (MFTs). Psychiatrists can also prescribe medications, since they are physicians.

How do you find a therapist? The first step would be to ask your CF team to find out if they have contacts of therapists they already work with. At Stanford, you can be referred to our Psychiatry Department if your insurance allows. You can also call the behavioral line on your insurance card to ask for insurance-covered therapists in your area. If you have medical or county insurance, your best bet would be to contact the behavioral services via your county. You might also research therapists with a preferred background or in your preferred location via Psychologytoday.com. The National Alliance on Mental Illness (NAMI) has additional resources and ideas about how to find a therapist that is right for you.

What can you expect when you contact a therapist? They will likely call you and conduct a short phone intake. If the fit is right, your appointment will be scheduled. The good development after the COVID-19 pandemic is that most, if not all, therapists now conduct virtual appointments in addition to in-person ones. This really increases the number of therapists available to you. The first few appointments are to get to know you better and to come up with a plan of treatment for you. Think about your particular goals for therapy and communicate them with your therapist. Feel free to ask your therapist about their school of training and for how long they envision your treatment.
Some therapies are short-term based, with 12 to 20 sessions being enough to work through an issue at hand, while others take longer.

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It probably will take you five or six sessions to determine if your therapist is the right fit, meaning you feel comfortable with them and able to identify a comfortable flow to your work together, but always feel free to speak up. Frequently, the discomfort you might feel in the session is the best material to bring to your therapist. While the long-term goal of therapy is to ease your distress, actively working through therapy might be distressing and uncomfortable. Your therapist would love to know about it, and it might help them be of even better help to you.

Finally, have fun! While I am biased and think that anyone can benefit from therapy, therapy can certainly provide a safe space to be vulnerable, learn more about yourself, grow, and heal.

Coping With a Chronic Condition

—Debbie Menet, LCSW

As a social worker in Stanford Medicine Children's Health's CF Center, I've had people with CF and their caregivers share with me the various strategies they use for coping with cystic fibrosis. There is one certainty—there will be ups and downs. "Chronic condition" means a condition that requires ongoing medical attention. For people with CF, that likely means attention throughout the life span. So how do we cope? What tools and strategies do we have to bounce back from the challenges? Following are some suggestions based on your report, research, and literature.

1. **Acknowledge and understand how you have power.**
   People with CF have a genetic mutation. This is nobody's fault. Nobody did anything wrong to cause this mutation. This can leave some people feeling powerless. As a person with CF or their caregiver, you do, however, have power over how you respond to this condition. Completing treatments, taking medications and enzymes, eating and exercising according to your needs, a positive mindset—are all ways you have power over your condition. How can you, as person with CF or a caregiver, harness YOUR power?
2. **Understand your condition.**
   Your CF team is available to answer any questions. Doctors, nurses, dietitian, pharmacist, physical therapist, respiratory therapist, social worker … we are eager to help. The more you understand your condition—and how your unique body responds to the condition, treatments, medications—the more confidence you will have and the better equipped you will be to move into your future. What questions do you have for your medical team?
3. **Have a toolbox full of coping strategies/ tools that you can access when needed.**
   Suggestions:

- **Talking to a trusted friend or relative.** Texting, talking on the phone, FaceTime, meeting up … have a list of three or four people. Who can you reach out to? Who is on YOUR list?

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- **Movement.** Taking a short walk outside, dancing in your bedroom, sitting on the floor and stretching, a brief yoga video. Movement helps us release emotions that are physically stuck. It releases feel-good hormones. It helps us think more clearly. It does not have to be intense or long; it can be simple and brief. What types of movement do you like?
- **Mindfulness.** Mindfulness is simply paying attention to this very moment. Not worrying about the future and not thinking about the past. Simply paying attention to your breath, paying attention to your various senses (smell, touch, hearing, taste, vision), writing in a journal. It can be five minutes long or 60 minutes long. Mindfulness takes us out of fight-or-flight mode and calms our nervous system. There are many mindfulness apps that are free and can help you connect to this moment. What type of mindfulness are you willing to try?
- **Taking a gratitude break.** There is evidence that regularly taking a few moments to identify a few things for which we are grateful rewires our brain toward happiness and contentment! No need to go big; we can kind find gratitude in the simple. Can you identify a few things for which you are grateful?

Your toolbox will naturally change over time and change with different circumstances. I encourage you to occasionally consider what tools you have in your toolbox, be open to exploring new ones, and find what works for you. Familiarity with your toolbox makes it easier to access when you most need it.

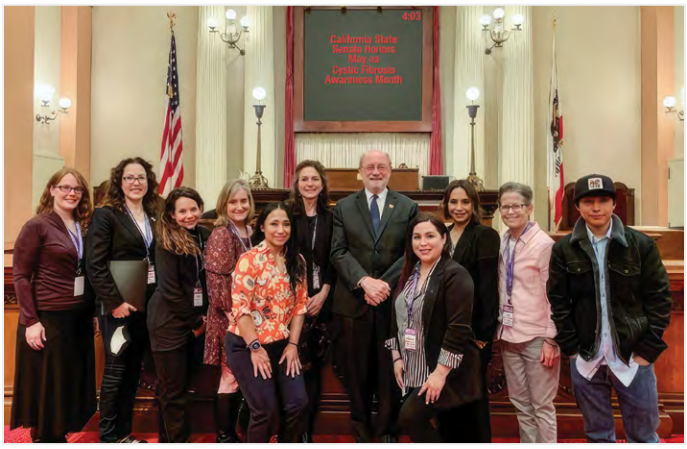
Advocacy Day at the State Capitol on May 8

—Mary Helmers, RN, BSN

On April 11, 2023, Senate Concurrent Resolution No. 49 was introduced by state Sen. John Laird to declare May to be Cystic Fibrosis Awareness Month in California.

On May 7, I traveled to Sacramento, along with board members from CFRI (Cystic Fibrosis Research, Inc.), parents of CF children, and three CF patients for in-person visits with our elected representatives.

Advocacy Day was on Monday, May 8, where we met and attended several meetings with Senate representatives and/or their aides. While we were at the Capitol, we were notified that SCR 49 would be presented on the floor for a vote.



CF advocates in Sacramento

All of us were witness to this momentous moment, when May was declared CF awareness month in California. It was fun and inspiring!

Cystic Fibrosis Registry

—Cathy Hernandez, AD

Our center, as well as other centers, keeps a database for the Cystic Fibrosis Foundation called the Cystic Fibrosis Registry. This registry includes patients with cystic fibrosis as well as patients who have been transplanted. When you come to a clinic visit, you might be approached to consent for the registry. Once consented, you are entered into the database and given a unique number that helps us identify you. This is a number that is used instead of your name; everything is kept completely confidential. When you come to a clinic visit, some of the data collected and entered into the database includes:

- PFTs (pulmonary function tests)
- Medications
- Microbiology
- Height/weight

At the end of every year, we collect information for an annual review, which needs to be completed by February of the following year. This includes economic information, labs, x-rays, and other important facts.

The registry is constantly improving by adding information, as well as combining new programs such as the CF SmartReports. The registry data is transferred into CF SmartReports, which are then compiled into important lists and graphs that can help with quality improvement and guide us to improve our center overall.

The data in the registry is helpful for our physicians as they prepare for research. They are able to search the database for key information that aids in their research project.

Thank you for your continued support of programs like this. When you take the time to do this, it improves not only our center, but centers around the country.

Frozen Yogurt Banana Pops

—Julie Matel, MS, RD, CDE

Prep time: 10 mins. Freezing time: 2 hrs.

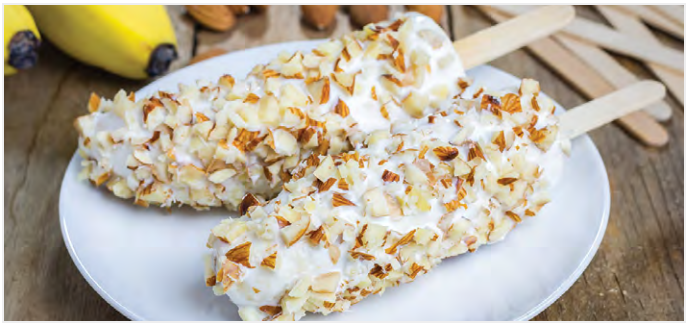
Cook time: 0 mins. Total time: 2 hrs., 10 mins.

Ingredients

- 3 medium bananas
- 1 cup yogurt
- 1 cup toppings of choice—shredded coconut, chopped nuts, sprinkles, mini chocolate chips, peanut butter drizzle, crushed pretzels

Instructions

1. Peel bananas and cut in half. Place a Popsicle stick in the end of each banana half and insert about halfway into the banana.
2. Lay bananas on a baking sheet with a piece of parchment paper.



Frozen yogurt banana pops

3. Freeze for about an hour. Dip each banana into yogurt and then roll into the toppings.
4. Place back on the baking sheet and freeze for another hour. If you want to store them longer, place in an airtight container and enjoy for up to two to three months.

See the recipe online **here**.

Pediatric CF Center Update

—Mary Helmers, RN, BSN

Social Media Updates

Website:
http://med.stanford.edu/cfcenter.html

Like us on Facebook:
Cystic Fibrosis Center at Stanford

Twitter account:
Stanford CF Center—@cf_stanford

Helpful Tips!

Did you know that you can get assistance with your PG&E bill? PG&E forms for medical equipment/devices can be found on the PG&E website under **Medical Baseline Allowance Application for Medical Baseline Enrollment and Recertificatio**n, and **you can now apply online**.

Or you can print the form, fill it out including all your medical devices (i.e., nebulizer/compressor, if you use oxygen, CPAP or BIPAP), and bring the form with you to your next CF clinic visit, and your provider will sign it; then you mail the form to PG&E.

Helpful Reminders!

To help expedite your clinic visit: Please remember to bring your **CF Binder** with you to clinic, along with the most recent CF Action plan.

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/parent emails, since it is not a secure site. Please note that any email sent to the team will be responded to by a phone call. Your CF care team can only communicate with you via MyChart or by phone. If you/your child has a clinical need/question, please call the CF RN line at (650) 736-1359.

It takes only a minute to sign up—one of the front desk staff will be happy to assist you with the sign-up.

Prescriptions: Just a reminder that your prescription request can take up to 72 hours to be processed. This has always been our policy; however, 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 48–72 hours for the pharmacy to process (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.
- Your pharmacy should call us if you have no refills.

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

Annuals: Remember, our goal is to get all annual testing done on or around your child's birthday. At your clinic visit three months prior to when your annuals are due, the CF RN will review with you what is due. Please feel free to ask us, too.

Included in the annuals are your lab work, CXR, bone density scan (12 years of age and older), full PFTs (starting at age 7), baseline audiogram (starting at age 6), liver screening, and sputum cultures. Please let us know if you have not had any of these tests done with your annuals.

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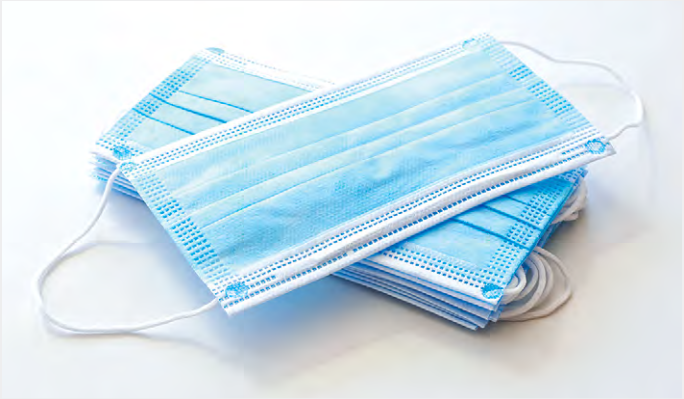
Infection Control
Patients should wear surgical masks (yellow, blue, or white) to and from all clinics and the hospital. They should fit around the nose and mouth.



Surgical masks

COVID-19 Updates
Masks are no longer required to be worn here in the clinics or hospital for anyone who does not have CF. For all our CF patients, however, the infection control guidelines as stated above remain the same and have not changed. Masks are always required when walking to any area in the clinics and hospital. CF patients can now take their mask off in the clinic room, but when leaving the room, a mask must be worn.

We hope all our patients and families are doing well. We understand that the past three years have been a challenge for everyone, and we want our families to know that we are here to answer all your concerns and questions. Feel free to call the CF RN phone line at (650) 736-1359 if you have any questions or concerns, or if you feel that you need some additional support.

Make sure that you bring your **CF PASSPORT** with you! Use the **PASSPORT** around the hospital wherever you have an appointment, test, or procedure. Remember, parents/patients, to carry your child's **CF PASSPORT** in your wallet. **If for some reason you do not have one or lost it, please ask for one when you come to your next clinic appointment**. We now have them in English and Spanish.

CF PASSPORT

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**

XOC (Excellence of Care) Surveys

The Cystic Fibrosis Foundation (CFF) will be sending quarterly surveys (every three months) to all our patients and families after a clinic visit. Surveys (in English and Spanish) are currently being sent via text or email.

Purpose of the survey: To hear directly from our patients and families about their care experience, the Cystic Fibrosis Foundation convened a multidisciplinary steering committee to create the new survey and include perspectives of people with CF, parents of children with CF, representatives from the CF Foundation, and physicians, nurses, and other care team disciplines, including social work, respiratory therapy, and nutrition. The XOC survey focuses on shared decision-making, relationship with the care team, infection control, and overall quality of care. It also includes special questions to capture the experience of telehealth visits.

We appreciate your taking the time to fill out the surveys so that we can best serve your needs. The CF team wants to provide all of our patients and families with the best care, so we value your constructive feedback.

Movement Before Airway Clearance Treatments

—Taylor Lewis, PhD, CSCS, CMT, PRT

Airway clearance techniques aim to enhance mucus mobilization, reduce airway resistance, improve ventilation and gas exchange, and optimize breathing cycles (Lauwers et al., 2020).

In airway clearance treatments, the musculoskeletal system plays a vital role in the mechanical function of breathing. During inspiration, the diaphragm and external intercostal muscles contract, causing the rib cage to expand and move outward, further expanding the thoracic cavity, thereby increasing lung volume. In addition, the diaphragm and intercostals relax during expiration, causing the thorax and lungs to recoil.

Research has shown that movement-based training increases mucociliary transit time (Silva et al., 2019). This is important because preparing the skeletal tissue for airway clearance treatments through low-grade movement can help improve the treatment outcome. A movement-based warm-up allows the body to transition and adjust to the biomechanical and physiological demands that occur during chest therapies. Essentially, when you move the body, the need for oxygen increases. Movement further increases our cardiorespiratory rate and blood flow to muscles, which prepares the respiratory muscles and the surrounding musculoskeletal systems for the task at hand. This is one of the reasons why warm-ups are essential and integrated into exercise programs.

So how can you optimize airway clearance treatments?

Introduce a five-minute full-body movement-based warm-up before treatments, targeting upper- and lower-body multijoint exercises that take the body through multiple ranges of motions, such as cat-camels, dead bugs, glute bridges, and lunges. Multijoint exercises allow you to spend less time warming up and increase blood flow and core temperature faster and more efficiently than single-joint exercises that isolate one muscle group. The progressive increase in respiration frequency and intensity that occurs during the warm-up will prepare the respiratory muscles for the airway clearance technique. In addition, the rhythmic vibration will loosen up the mucus to be able to move out of your system. This will set you up to maximize your airway clearance treatment because you will have progressively prepared the body for chest wall expansion.

Bonus Tip

Water accounts for 75% of muscle mass (Sawka, 1992). Therefore, drinking a glass of water within 30 minutes of waking up can help reduce dehydration that accumulates overnight. This will start your day in a good position and set your respiratory muscles up for quality performance during the next treatment.

References

Lauwers, E., Ides, K., Van Hoorenbeeck, K., & Verhulst, S. (2020). Outcome measures for airway clearance techniques in children with chronic obstructive lung diseases: a systematic review. *Respiratory Research*, 21(1), 217. https://doi.org/10.1186/s12931-020-01484-z.

Sawka, M.N. (1992). Physiological consequences of hypohydration: exercise performance and thermoregulation. *Medicine and Science in Sports and Exercise*, 24(6), 657–70.

Silva, B.S.A., Ramos, D., Bertolini, G.N., Freire, A.P.C.F., Leite, M.R., Camillo, C.A., Gobbo, L.A., & Ramos, E.M.C. (2019). Resistance exercise training improves mucociliary clearance in subjects with COPD: A randomized clinical trial. *Pulmonology*, 25(6), 340–47. https://doi.org/10.1016/j.pulmoe.2019.01.001.

Current Research Studies

Active Research Studies

Vertex 21-121-105: A Phase 3 Study Evaluating the Pharmacokinetics, Safety, and Tolerability of VX-121/ Tezacaftor/Deutivacaftor Triple Combination Therapy in Cystic Fibrosis Subjects 6 Through 11 Years of Age

Vertex21-445-112: A Phase 3 Open-label Study Evaluating the Long-term Safety and Efficacy of Elexacaftor/Tezacaftor/Ivacaftor Triple Combination Therapy in Cystic Fibrosis Subjects 2–5 Years

MCC: Mucociliary Clearance Study: Study evaluating the use of adrenergic and cholinergic agents to increase mucociliary clearance in ages 18+.

Peripheral Biochemical Monitoring Study: Study to evaluate if certain molecules that are secreted in sweat and saliva can be used to diagnose and monitor health conditions in ages 18+.

Pf Bacteriophage and Clinical Outcomes in Cystic Fibrosis: Monitoring of patients with cystic fibrosis, chronic pseudomonas, and presence of Pf bacteriophage with banking of sputum samples at clinical encounters in ages 18+.

Collection of Gene Mutation for Laboratory Quality Assurance: Newborn Screening Accuracy Project: Study collecting blood samples from patients with rare CF mutations to ensure that newborn screening tests are accurate in all ages.

The PROMISE Study: A prospective study to evaluate biological and clinical effects of significantly correct CFTR function in ages 12+.

Utility of Lung Clearance Index: LCI study to be done in clinic for pediatric patients less than 18 years old.

Innovative Strategies for the Study of Disorders of the Respiratory Tract for pediatric patients less than 18 years old.

Pulmonary Function in People with Viral Infections: Study to evaluate if certain molecules that are secreted in sweat and saliva can be used to diagnose and monitor health conditions in ages 18+.

Upcoming Research Studies

Vertex 21-522-001: A Phase 1 Single Dose Escalation Study Evaluating the Safety and Tolerability of VX-522 in Subjects 18 Years of Age and Older with Cystic Fibrosis and a CFTR Genotype Not Responsive to CFTR Modulator Therapy.

BEGIN-OB-19: A Prospective Study to Evaluate Biological and Clinical Effects of Significantly Corrected CFTR Function in Infants and Young Children (BEGIN).

Longitudinal Characterization of Respiratory Tract Exacerbations and Treatment Responses in Primary Ciliary Dyskinesia: To provide critical data needed to inform the design of future interventional trials of respiratory exacerbation prevention and treatment in children and adults with primary ciliary dyskinesia (PCD).

NICE-CF: Observational study of adults with cystic fibrosis for colorectal cancer screening. Study will compare stool-based testing to colonoscopy for colorectal cancer screening in people with CF ages 18+.

RESPIR-102: A Double-Blind, Active-Controlled, Multiple-Ascending Dose, Phase 1b Study of Aerosolized RSP-1502 Delivered VIA the PARI LC Plus Nebulizer in Subjects with Cystic Fibrosis and Chronic Pseudomonas Aeruginosa Lung Infection.

Struggling With Your Home Spirometer?

—Jennifer Kwok, RCP, and Gauri Pendharkar, RCP

Home Spirometers

A significant number of patients in our center have home spirometers from ZEPHYRx. Instructions on how to use them are included in the package and were sent by your CF RT. If you have trouble using it, a good resource is the customer support center at (888) 452-6269 or the ZEPHYRx Home Spirometer User Guide video on https://www.youtube.com/watch?v=VCW34jrvgLE.

The spirometer, **the MIR Spirobank Smart**, requires that you have the **ZEPHYRx Breathe Easy mobile application**, which you will need to install on an Apple or Android cell phone or tablet.

To establish a baseline, we recommend that you take a test when you are feeling well. If you are scheduled with a telemedicine—video or telephone—visit, it is imperative that you do a pulmonary function test (PFT) just before your visit.

After you have completed your tests, you need to take a screenshot of your results and upload the image to us via MyHealth as an attachment.

The results are not automatically sent to your provider, as the spirometry indicates.

Useful spirometry tips:

1. Use the **Breathe Easy** app.
2. Enter your height, weight, etc.

3. Exhale for as long as the device prompts you, or as much as possible.
4. Perform no more than three or four attempts. More attempts (per device instructions) can cause fatigue and distort numbers.
5. Remember that you **DO NOT** do three normal breaths **on the spirometer** before the deep inhalation (as we do in clinic). Just put the device in your mouth, take a fast deep breath in, and blow out all the way.
6. End the test with another fast deep inhalation.
7. Take a screen shot of your result and send it to us via MyHealth.

> “To establish a baseline, we recommend that you take a test when you are feeling well.”

CF Family Advisory Council Fun in the Sun—Travel Tips

—Kirsten McGowan

Summer is here, and with that comes possible vacations or trips. As parents of kids with CF, we understand that traveling can be challenging, but with the right preparation, you can make it less stressful! We put together our best tips and hacks for travel that we've learned over the years (some the hard way).

If you have input or an idea for the CF Family Advisory Council (FAC), please email Kirsten McGowan at kmcgowan@stanfordchildrens.org.

Before You go

- Refill meds early to have plenty for the trip (plus always pack extra in case of unexpected delays!).
- Will the place you're staying have a place to:
    - Wash/sterilize nebulizer sets? PRO TIP! Medela microwave steam bags work great for nebulizer sets (except the soft plastic masks for infants/toddlers). You can also ask about disposable nebulizer sets.
    - Refrigerate meds that need to stay cold?
- Request a travel letter from the CF care team. PRO TIP! This helps getting through TSA much easier and allows for pre-boarding of plane flights.
- If traveling outside the U.S.: Make sure you have the correct plug adapter so that you don't damage any equipment.
- It helps to know the location and phone numbers of the closest pharmacy and CF center at your destination (just in case!). PRO TIP! Double-check with your health insurance that they will be "in network" so you know what kind of coverage you'll have.

Traveling

- If flying, always try to carry on all your meds and equipment. It takes away any risk of anything getting lost or damaged, and liquid limits for medicine do not apply to the TSA limit. PRO TIP! Medical devices (vest/compressors) and medicines do NOT count toward carry-on limits (especially with that handy travel letter from the CF care team).
- Small cooler bags with ice packs are handy to transport cold medicine.
- Give yourself extra time to get through security. Sometimes the medical equipment needs extra screening. You can call TSA Cares three days before your flight to ask any questions: (855) 787-2227.
- Wear masks on the airplane, and bring sanitizing wipes to wipe high-touch areas (armrests, tray, seat belt, etc.). PRO TIP! This is why preboarding is huge—it gives your family more time to settle in.
- Pack extra snacks. Not all flights have high-calorie options that your child will like.

If Your Child Needs Medical Care

- It's happened to all of us: Someone gets sick, gets hurt, or otherwise needs to see a doctor while you're on vacation—that's OK!!
- It's also normal to worry about seeing a doctor who might not know CF. If you see someone at an urgent care or emergency room, or even if it is another CF center, have them call Stanford Medicine Children's Health at (650) 497-8000 and ask to speak with the pulmonologist on-call. It helps the CF care team understand what is happening with your child, and they can talk with the other doctors to get your child the best care possible.

Meet Our New Team Members

Pediatric Clinic

Amanda Keen, MSN, RN, is joining the CF Research team as a Research Nurse. She completed her bachelor's degree in psychology at UC Berkeley in 2010 and her master's degree in nursing at UC Davis in 2020. After working on various research studies at UCSF and the San Francisco Department of Public Health around substance abuse and sexual/reproductive health, Amanda entered the fertility and reproductive endocrinology field. Most recently, she worked as nurse case manager at Stanford's Fertility and Reproductive Endocrinology department in Sunnyvale. In this capacity, she guided patients through the IVF treatment process, assisting couples who were carriers of genetic conditions like CF in their family planning. Amanda enjoys reading, going on hikes with her husband, and birdwatching with her cat, Daphne, in her free time.

Neetu Perumpel, MSN, RN, is a Chicago native who started her career as a nursing assistant on the Pediatric Surgery/Transplant unit at Ann and Robert H. Lurie Children's Hospital of Chicago in 2013. She became a nurse on the General Medicine, Infectious Disease, and Pulmonary unit at Lurie in 2014, where she initially found her passion working with patients with cystic fibrosis. In 2019, she obtained her master's degree in nursing and transitioned into a clinical quality coordinator position for the same unit, where she oversaw many hospital improvement projects and safety initiatives. She moved to the Bay Area with her husband and dog in 2023 and is excited to continue working with the CF community as a CF Nurse Coordinator at the Cystic Fibrosis Center in Emeryville.

Adult Clinic

Jennifer Mori, RCP, has been a respiratory therapist at Stanford for 18 years. She has experience working with CF patients in the inpatient and outpatient settings, and also in the Pulmonary Function lab. She continues to work alongside patient care, including the ICU and ED. In her spare time, she loves spending time with her family and two kids, ages 10 and 7. She also enjoys trying new restaurants and traveling to warm places. She is looking forward to working with the CF care team and is excited for this opportunity!

Cystic Fibrosis Center at Stanford

Pediatric providers at Lucile Packard Children's Hospital Stanford

Pediatric Center Director: Carlos Milla, MD
Providers: Sumit Bhargava, MD; MyMy Buu, MD; Elizabeth Burgener, MD; Carol Conrad, MD; David Cornfield, MD; Michael Tracy, MD; Jacquelyn Spano, DNP, RN, CPNP; Cissy Si, MD; Nick Avdimiretz, MD
Clinic Scheduling (650) 724-4788
Clinic and Prescription Refill Fax (650) 497-8791
Office Assistant/Patient Services Coordinator:
Laura Banuelos (650) 498-2655
Nurse Coordinator: Mary Helmers, RN, MSN (650) 736-1359
CF Clinic Nurse: Liz Beken, RN (650) 736-1359
Respiratory Therapist: Jessica King, RT (650) 724-0206
Nutritionist, Dietitian:
Julie Matel, MS, RD, CDE (650) 736-2128
Social Worker: Debbie Menet, LCSW (650) 796-5304
Newborn Screening Coordinator:
Jacquelyn Spano, DNP, CPNP-AC/PC, CCRC (650) 721-1132
Clinical Pharmacist:
Jake Brockmeyer, PharmD, BCPS (650) 505-9419
Clinical Psychologist: Diana Naranjo, PhD

For urgent issues:

Monday to Friday, 8 a.m. to 4 p.m.
Call the CF Clinic Nurse (650) 736-1359
After hours and weekends: Call the main hospital and ask for the on-call pulmonology doctor (650) 497-8000

Pediatric providers at Stanford Medicine Children's Health Specialty Services – Emeryville

Providers: Karen Hardy, MD; Eric Zee, MD; Manisha Newaskar, MD; Rachna Wadia, MD
CF Clinic Scheduling (844) 724-4140
Clinic and Prescription Refill Fax (510) 457-4236
Nurse Coordinator: Neetu Perumpel, MSN, RN (650) 724-8414
Respiratory Therapist: Lorraine MacPhee, RT (510) 587-9631
Nutritionist, Dietitian: Mikaela Burns, CRD, MPH (510) 806-3659
Social Worker: Teresa Priestley, MSW (925) 357-0733

For urgent issues:

Monday to Friday, 8 a.m. to 4 p.m.
Call the CF Clinic Nurse (650) 724-8414
After hours and weekends: Call the main hospital and ask for the on-call pulmonary doctor (844) 724-4140

Adult providers at Stanford Health Care

Adult Center Director: Paul Mohabir, MD
Associate Center Director: Alicia Mirza, MD
Pulmonologists (MDs): Laveena Chhatwani, MD; Alicia Mirza, MD; Paul Mohabir, MD
Director of Psychiatric and Psychological Services: Liza Sher, MD
Infectious Disease Consultant: Joanna Nelson, MD
Advanced Practice Providers: Elika Rad, NP; Meredith Wiltse, NP
Clinical Pharmacist: Denise Kwong, PharmD
Adult Clinic Scheduler/Patient Care Coordinator:
Patricia Morales (650) 723-0798
Adult CF Center Fax (650) 723-3106
Nurse Coordinators: Theresa Kinney, RN and Kristel Fallon, RN (650) 498-6840
Respiratory Therapy: Erica Collins, RCP IV; Jenny Kwok, RCP IV; Jennifer Mori, RCP; Gauri Pendharkar, RCP (CF RT Coordinator) (650) 736-8892
Registered Dietitian:
Marion Seabaugh, MPH, RD, CNSC, CCTD (650) 529-5952
Social Work: Meg Dvorak, LCSW (650) 518-9976
Social Work: Kate Yablonsky, MSW (650) 444- 6512
Routine issues/concerns during business hours (Monday–Friday, 8:00 a.m.–4:30 p.m.)

- CF Nurse Coordinator Line (650) 498-6840
- Voicemails will be answered within 24–48 business hours, or sooner based on clinical priority.
- Alternatively, you can utilize MyHealth messaging for NON-URGENT NEEDS ONLY. MyHealth messages are NOT checked after hours or on the weekends

Urgent issues/concerns DURING business hours (Monday–Friday, 8:00 a.m.–5:00 p.m.)
Chest Clinic Call Center (650) 725-7061
• A message will be generated and sent to the CF Team ASAP
Urgent Issues/concerns AFTER business hours:
Chest Clinic Call Center (650) 725-7061

- A message will be generated and sent to the covering CF provider ASAP.
- MyHealth messages are NOT checked after hours, weekends, or holidays.

Adult providers at CPMC

Adult Center Director: Ryan Dougherty, MD
Associate Center Director: Vinayak Jha, MD
Provider: Christopher Brown, MD; Carolyn C. Hruschka, ANP-BC
Adult Clinic Scheduling (415) 923-3421
Adult CF Center Fax (415) 243-8666
Nurse Coordinator:
Carolyn C. Hruschka, ANP-BC (415) 923-3421
Respiratory Therapy:
Bryan Ellis, RCP; Arthur Pundt, RCP (415) 600-3424
Registered Dietitian: Elena Zidaru, RD (415) 923-3997
Social Work: Amy Greenberg, LSW (650) 518-9976
Mental Health Coordinator:
Amy Greenberg, LSW (415) 923-3854

For urgent issues:

Monday to Friday, 9 a.m. – 5 p.m.
Call the nurse coordinator (415) 923-3421
Evenings/weekends: Call and ask for the on-call pulmonary provider (415) 923-3421

Research

Tina Conti, BSRC, RRT-NPS (650) 498-8701
Lani Demchak, MBA (650) 725-1087
Monica Elazar, DDS (650) 723-5193
Cathy Hernandez, AD (650) 724-3474
Amanda Keen, MSN, RN (650) 723-4670
Jacquelyn Spano, DNP, CPNP-AC/PC, CCRC (650) 721-1132

CF Center at Stanford
770 Welch Rd Ste 350
Palo Alto, CA 94304

Where Can I Find NEB Cups?

—Jennifer Kwok, RCP, and Gauri Pendharkar, RCP

Ordering Supplies for Your Home RT Treatments
Lately, ordering respiratory therapy supplies such as compressors, nebulizers, and filters has been a challenge for many patients. Many of the regularly used pharmacies are no longer working with insurance companies to supply durable medical equipment (DME). We are still able to order supplies from specialty pharmacies such as Foundation Care and Alliance Walgreens Specialty Pharmacy. Other vendors, such as CVS and Walgreens, do not carry the recommended Pari nebulizers and compressors but do have generic or different brands.

One option is to contact patient assistance services; here is a list of commonly used programs:

- Compass: Compass@cff.org; (844) 266-7277.
- HealthWell grant through the HealthWell Foundation: https://www.healthwellfoundation.org/.
- The Living Breath Foundation: https://www.livingbreathfoundation.org/.

Unfortunately, some patients may still end up paying out of pocket for DME. If you end up purchasing, it is important to ensure that the nebulizer cups are **reusable and not disposable**. Some reputable vendors are:

- **Amazon:** Pari LC Plus (reusable), Pari LC Sprint (reusable), Pari Trek S compressor (rechargeable battery not included).
- **Justnebulizer.com:** A large variety of compressors, including Pari products, are available, along with Aerobika. Sometimes a prescription may be required for these; please contact your CF RT.
- **Nebology.com:** A large variety of compressors, including Pari products, are available. Pari E-Flow compressors and E-Rapids neb cups are available. Sometimes a prescription may be required for these; please contact your CF RT.

Newsletter Contact Information
Editors: Lani Demchak
Visit our website at http://cfcenter.stanford.edu for more information about our center and cystic fibrosis. To subscribe to this newsletter, please contact Cathy Hernandez at cathyh1@stanford.edu. Follow us on Facebook: Cystic Fibrosis Center at Stanford.