

Cystic Fibrosis Center News

COVID-19 and Cystic Fibrosis

— Richard B. Moss, MD

The worldwide pandemic of the SARS-CoV-2 virus continues, with over 6 million cases and 370,000 deaths. First recognized in the city of Wuhan, China, in late 2019, this “novel” (new to humans) coronavirus infection is now present in almost all areas of the globe and all 50 of these United States. America is now the world epicenter, with nearly 1.85 million cases and 107,000 deaths. These terrible numbers will increase every day for months to come.

The virus was long present in bats but probably mutated, perhaps in an intermediary animal such as the pangolin, and became able to infect humans. It does so by binding to a molecule called ACE2 on the surface of lung, blood vessel,

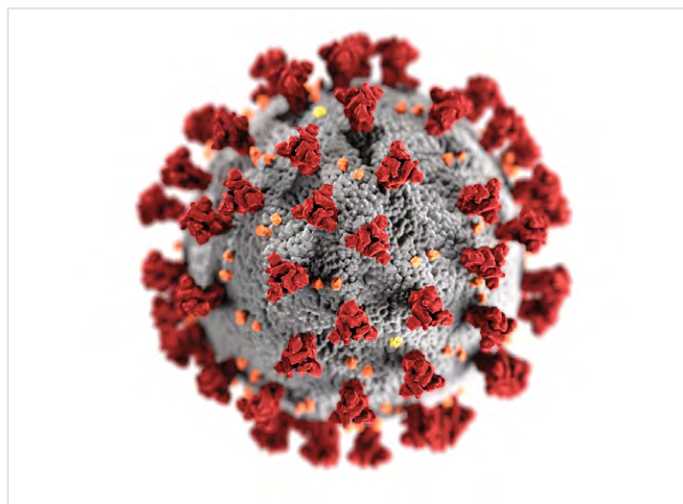


Illustration of SARS-CoV-2

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and other cells, aided ironically by a human enzyme present on those same cells that activates the binding spike (S) protein of the virus. Once in the cell, the genetic material in the virus (in this case RNA rather than DNA) takes over the cell and makes it produce more virus until the cell bursts, releasing the new viral particles. It should be noted that SARS-CoV-2 is very similar to some other coronaviruses that jumped from animals to humans, namely SARS in 2002 in East Asia and MERS in 2012 in the Arabian Peninsula. However, SARS-CoV-2 is distinguished from these forerunners by two insidious features: It is less virulent but more infectious. Being less virulent means fewer infected people get seriously ill—this allows for silent spread by people who don't know they're sick or think they only have a common cold or mild flu. Being more infectious means it can spread more easily, in droplets exhaled by coughing, sneezing, talking, or even just breathing, and it can survive for hours and perhaps days on surfaces that the tiny airborne virus-containing liquid droplets settle upon.

Finally, it needs to be remembered that these recent animal-to-human coronaviruses are also related to other coronaviruses that long ago adapted to humans and are not particularly troublesome, causing 10 to 30 percent of seasonal common colds. These older human seasonal coronaviruses have been included in many conventional respiratory virus test panels, and early in the COVID-19 pandemic there was confusion when some labs reported positive “coronavirus” results to people actually infected with one of the older benign common cold types and not SARS-CoV-2, leading to unnecessary alarm or panic. This has been corrected in lab reporting. Today some labs will test for conventional respiratory viruses (like influenza A and B, parainfluenza, RSV, and seasonal coronaviruses) as well as SARS-CoV-2 specifically.

The disease that SARS-CoV-2 causes goes by the name of COVID-19. It attacks mainly the

respiratory tract, including lung cells lining the air sacs (alveoli) where oxygen from air enters the blood, thus starving the body of oxygen. The heart, kidneys, intestines, and nervous system are also involved in many cases. The earliest cases nationally were recognized in Washington state and shortly thereafter in Northern California. Early action in these states to implement physical distancing has flattened the exponential growth curve of infection, but other regions, such as the New York metro area, acting later, were not so fortunate, relatively speaking.

Identification of cases by symptoms and testing for the genetic RNA material of the virus using nasal or oral swabs has badly lagged in the United States, and our federal system has dealt poorly with the need for a uniform national response. Recently, many commercial antibody tests have become available, but all await validation; these show only a small percentage of the population has been exposed to SARS-CoV-2, meaning the vast majority remains vulnerable.

Shortages of personal protective equipment (PPE) for health care workers and other essential workers, and of certain medical supplies and equipment such as ventilators, have led to more misery. In some locales, hospitals have been overwhelmed. Today nearly all of us across the nation are under stay-at-home orders under local or state emergency decrees. Much social life has migrated to teleconferencing apps like Zoom, while much non-emergency-medicine health care has migrated to telemedicine encounters. The nation struggles to improve its detection systems, to measure exposure and (hopefully) immunity in the population using blood antibody tests, and to find the necessary supplies of masks, other PPE, and needed medical equipment and drugs. Realizing that prior neglect has gutted our national public health capacity, we now search for ways to train the hundreds of thousands of personnel needed to detect cases and then reach and quarantine contacts of each case.

Ironically, the cystic fibrosis (CF) community has been better prepared for this pandemic than almost any other group in society. For years we have hammered home the importance of infection control, especially hand hygiene and mask-wearing in any setting where cross-infection between people with CF exists. Now the world has come to live like us. If there is any silver lining in all this, it is that people with CF, their loved ones, and their care providers know what to do and how to do it. Of course, since the risk is essentially universal rather than restricted to health care facilities or social activities involving patients, more vigilance is necessary—the handwashing, the disinfecting of surfaces, the wearing of masks in public, and so on. But we get it; we are not the ones needing reminding, cajoling, arguing.

Because of the primarily respiratory nature of this infection, and the profiles of groups at higher risk of more severe COVID-19 disease, preexisting chronic lung disease as a category is a major risk factor. In COVID-19 reports, because of numbers, this shows up mainly as people with COPD rather than identifying CF. The Cystic Fibrosis Foundation, the European CF Society, and an international group have begun reporting COVID-19 cases. There are only very few cases worldwide (well under 100 to date) of people with CF who have come down with COVID-19. So far, based on this scanty data, it appears that people with CF fare similarly to others, with age, diabetes, and transplant seeming to be greater risk factors. All these factors increase the sense of vulnerability. Nevertheless, we do know from early reports that even those with advanced lung disease and low baseline lung function have survived COVID-19.

Treatment remains supportive. This means no specific medicine has been proven effective; however, there have been early promising results in sicker patients receiving remdesivir, an intravenous antiviral medicine. Giving oxygen, fluids, nutrition, and medicine for fever, and if necessary supporting exhausted bodies with failing lungs by using

noninvasive or invasive ventilation, is the current standard of care. Increasingly, increased blood clotting and vessel blockages (thrombi) have been seen in very sick patients, responding to anticoagulation treatment. Thankfully, only a small minority of infected people are sick enough to require admission to the hospital, and only a small fraction of those need ICU care. Unfortunately, of those sickest needing ICU care, many will not survive. This tragic group, whose terrible fate is so devastating to us all, is mainly composed of the elderly with coexisting chronic illnesses, especially hypertension, diabetes and obesity. Health disparities contribute greatly to COVID-19 severity, disproportionately affecting African-Americans and Latino individuals. About 10 percent of those hospitalized, however, are 20 to 50 years old. Children beyond infancy are at low risk, but not zero, and most have mild illness. Overall, remember that, depending on where specifically one looks, deaths represent less than 1 to at most 10 percent of those identified with COVID-19 infection, and the vast majority are over 60 years of age.

“Ironically, the cystic fibrosis (CF) community has been better prepared for this pandemic than almost any other group in society.”

A very large number of medicines are being tested, and hopefully by the time you read this we will know for sure if any besides the modestly effective remdesivir work. Hydroxychloroquine with or without azithromycin has not produced consistent benefit, and it has some dangers. Our very first dictum in medicine, “First, do no harm,” reminds us to do our best to establish safety and efficacy before widely using new therapies, all of which inevitably have some side effect/toxicity profile, not to mention cost and

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Nontuberculous Mycobacteria

— Cissy (Xin) Si, MD

What is NTM?

Nontuberculous mycobacteria (NTM) are a group of organisms found throughout our environment. The name helps differentiate it from its sibling, *Mycobacterium tuberculosis*, an organism that can cause serious human disease. NTM naturally live in water and soil and are commonly found in the home—in our showers, water heaters, plumbing systems, and garden soil. We are all exposed to these organisms on a daily basis, but our bodies are normally able to remove them before they cause harm. Disease occurs rarely—sometimes in healthy individuals and sometimes in those who have underlying immune deficiencies or lung disease. There are more than 100 species of NTM; the species that commonly cause disease in humans include *Mycobacterium avium* complex and *Mycobacterium abscessus* complex. Unlike *Mycobacterium tuberculosis*, NTM disease is contracted through environmental exposure, not from person-to-person spread.

What kind of disease do NTM cause?

NTM can cause infections of the lungs, skin/soft tissues, and lymph nodes, which are glands that help fight off illnesses. In instances where there is an underlying immune defect, NTM can cause disseminated disease—an infection of the entire body. Lung disease occurs when we inhale these organisms from the air into our lungs; other forms of disease occur when these organisms enter our body through a cut in our skin. In patients with cystic fibrosis, NTM most commonly cause lung disease, which is further divided into two subtypes: nodular bronchiectatic disease, in which inflammation and scarring leads to lung nodules and widening of the airways (most common); and cavitary disease, in which holes are formed in the lungs.



Cissy (Xin) Si, MD

Why are patients with CF at higher risk for NTM lung disease?

People with CF have underlying structural lung disease, bronchiectasis (widening of the airways), and difficulty clearing mucous; these create an increased risk for NTM lung disease. Older patients with poor nutrition are also at increased risk. Having a positive sputum culture for NTM does not necessarily mean a person has NTM lung disease. Some people can be colonized with NTM in their airways without any damage to their lungs.

How common is NTM lung disease?

Over the last few decades, the rates of NTM lung disease have increased. According to the CF Foundation, the rates of NTM were about 1 percent in 1984 but up to 12 percent by 2012. The rising rates are due to a combination of environmental changes, increased longevity, and better detection modalities. We estimate that

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Medications in the Event of a Natural Disaster

— Roy C. Lee, PharmD, BCPS

Natural disasters seem to be all too common now, and with each new disaster, more intense. As the recent COVID-19 outbreak and the recent fires in California have laid bare, such disasters can leave us vulnerable and cause much anxiety when we are left without our lifesaving medications. In such scenarios, we should all be prepared to know what to do. Planning starts before a disaster strikes. The following guidelines are essential:

- Keep a current list of your medications, including your dosages.
- Keep a log of how much medication you have.
- Obtain early refills in the event that your access to pharmacies may be limited (this will also allow you to keep a limited emergency supply on hand).
- Have your medications easily accessible so that you can quickly grab them in the event that you have to evacuate.

If disaster strikes and you find yourself without your lifesaving medications, and you cannot go to your usual pharmacy, you should go to the nearest pharmacy available and transfer your prescriptions. In the case of major drugstore chains such as Walgreens and CVS, you can also change your pickup location online. In cases where this is not possible, many states permit pharmacists to make medically necessary exceptions during a declared state of emergency. The California



Remember to keep track of how much medication you have left.

State Board of Pharmacy permits pharmacies to provide care by waiving requirements that may be impossible to meet during an emergency and to refill a prescription without a prescriber's authorization if failure to refill the prescription might have a significant adverse impact on the patient's well-being. However, depending on your insurance, you may have to pay out of pocket and seek reimbursement later. Other insurances will allow for emergency overrides once the pharmacy explains what is occurring.

A website that may be useful if disaster strikes is Rx Open ([rxopen.org](https://www.rxopen.org)), which can help you find nearby pharmacies in areas that are impacted by the disaster. Rx Open displays the precise locations on Google Maps of open pharmacies, closed pharmacies, and those whose status is unknown.

Pediatric CF Center Update

— Mary Helmers, RN

Mask changes

Since construction is complete here for both Lucile Packard Children's Hospital Stanford and Stanford Hospital, patients have switched back to wearing the yellow masks. Patients should wear them to and from all clinics and in the hospital, and when you walk outside the medical center. They should fit snugly around the nose and mouth. Patients can also wear a Vogmask (a microfiber, high-filter mask with vents).

COVID-19 update

We are aware that the coronavirus (COVID-19) outbreak is causing significant concern, particularly within the cystic fibrosis community. We hope that all our patients and families are doing well and staying safe. We want our families to know that we are here to answer all your concerns and any questions you may have. Feel free to call the CF RN phone line at (650) 736-1359 with questions or if you need some additional support during these uncertain times.

On May 4, we started to see patients back in the CF clinic. For the time being, visits will involve seeing the provider, the RN, and the RT (if applicable). If needed, the social worker, dietitian, and/or pharmacist will contact you by phone or telehealth after your clinic visit. We ask that only one family member accompany their child to the visit. No siblings or additional family members are allowed in the clinic at this time.

Everyone who enters the building and/or hospital will be screened at the entrance before coming up to the clinic.

We encourage you to refer to the **Centers for Disease Control and Prevention (CDC)** website, [CDC.gov](https://www.cdc.gov), for the most up-to-date information, the **Cystic Fibrosis Foundation** website, [CFF.org](https://www.cff.org), and our website, med.stanford.edu/cfcenter.

Helpful tips and reminders

Did you know that you can get assistance with your PG&E bill?

The PG&E form for medical equipment/devices can be found on the PG&E website: www.pge.com/en_US/residential/customer-service/other-services/brochures-and-forms/brochures-and-forms.page. Click on the "Medical Baseline Allowance Application."

All you need to do is print the form, fill it out including all your medical devices (e.g., nebulizer/compressor if you use oxygen, CPAP, or BIPAP), bring the form with you to your next CF clinic visit, have your provider sign it, and mail the form to PG&E.

CF clinic prep form (patient update)

This form was designed to help you get all your questions answered. Filling it out is not mandatory—it is a tool to assist you in jogging your memory in preparation for your clinic visit. Do you drive away from clinic thinking, "Oh no, I forgot to ask something?" You can now fill out this form ahead of time and bring it to your clinic appointment. Find it on our website, med.stanford.edu/cfcenter.

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 with a phone call. We do not always check emails on a daily basis, so if you or your child has a clinical need or question, please call the CF RN line at **(650) 736-1359**.

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

To help expedite your clinic visit, please remember to bring your CF binder and the most recent CF Action Plan with you to clinic.

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 take another 48 to 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! They will process it.
- 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.

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

Parents: Be sure to carry your child's CF PASSPORT in your wallet. The CF clinic sent the PASSPORTS out in the mail to each family; however, that was over a year ago. If for some reason you did not see it, tossed it, or may not have received it, please ask for one when you come to your next clinic appointment. We now have them in English and Spanish.

COVID-19...

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access issues. Properly designed clinical trials are underway for those mentioned above and many more potential COVID-19 treatments. Answers will come in the next few weeks to months. Once the spread of COVID-19 disease has come under some check by physical distancing (mitigation), America as we knew it all our lives until now will slowly emerge by carefully returning certain groups to activity outside the home.

Many are very concerned that the economic shutdown creates its own health risks as well as obvious other hardships, so there is tremendous pressure to do so. But this needs to be guided by fact and science—identifying cases, reaching their recent contacts and isolating that group (containment), and thus starving the virus, which needs the human body to come to life and multiply. Ultimately this pandemic will end when enough people are immune, either through surviving natural infection (likely to remain a very small minority of the entire population) or acquiring immunity via vaccination. Dozens of vaccine candidates have already been designed, and several have already entered early clinical trials. The timelines of viral spread and vaccine development are not in sync—the former moving exponentially in days, the latter moving to availability much more slowly. Even the often-mentioned “12 to 18 months” would represent the fastest vaccine program by far in history.

Stay informed by checking reliable sources, such as websites of the Centers for Disease Control and Prevention and your local county public health department. We need to be very patient, very consistent, very determined—and very hopeful. Finally, the CF community needs to be very mindful that the gains we've together experienced in recent years must remain in place by preserving all the elements of working together, ensuring that all those weapons that have pushed CF into a corner are not now neglected.

Nutrition Considerations in the Age of Modulator Therapies

— Julie Matel, MS, RD, CDE

Maintaining good nutrition has always been a cornerstone of care for individuals with cystic fibrosis (CF). We know that an early focus on achieving optimal growth and a normal body mass index (BMI) can improve outcomes for people with CF. For example, infants with CF who achieve weight-gain goals by 2 years of diagnosis are more likely to sustain better growth and better lung function through 12 years of age.¹ In addition, greater weight at age 4 is associated with greater height, better pulmonary function, and better survival through age 18.²

With the advent of new modulator therapies for CF, many individuals are seeing that it is less of a struggle to gain weight. Several studies have shown an improved weight and/or BMI³⁻⁷ as well as growth velocity⁶ in individuals on ivacaftor. Similar although less robust results were seen in individuals receiving a combination therapy of ivacaftor plus lumacaftor.^{8,9} With the new FDA-approved triple therapy, ivacaftor, tezacaftor, elexacaftor, promising results regarding improved weight and BMI have been demonstrated.^{10,11}

It is thought that perhaps reduced energy expenditure, improved fat absorption, decreased gastrointestinal inflammation,¹² and improved glucose intolerance^{13,14} may be contributing to these positive trends.

You are probably wondering, “How does this affect me or my child with CF?” Now that folks with CF are living longer, is there a greater risk for developing chronic diseases such as obesity,

type 2 diabetes, and heart disease? While the jury is still out on these topics, it may be prudent to consider dietary changes that promote health and longevity, with dietary adjustments depending on your individual nutrition goals. A well-balanced diet with foods from all of the food groups, including whole grains (whole grain rice, pasta, breads, and cereals), protein sources (meats, fish, poultry, dairy), and five to seven servings of fruits and vegetables per day will help ensure adequate intake of macronutrients as well as key vitamins and minerals.

A few tips and things to consider

- Your CF dietitian can help you monitor your weight and help you set goals.
- If weight gain is desired, consider incorporating healthy high-calorie options.
- Remember to take your modulator with fat-containing meals for maximum absorption.
- Talk to your team before adjusting your enzyme dose.
 - GI symptoms may subside after a few weeks.
 - Keep a poop log to observe changes.
- Talk to your endocrinology team before making any insulin dose changes.
 - Monitor blood sugars as directed.
 - Consider a continuous glucose monitor, which can help determine glucose fluctuations in real time.

Some examples of fat-containing meals to include when taking your modulator

Morning meal	Evening meal
Scrambled egg, avocado on multigrain toast	Chicken Caesar salad
Bagel with cream cheese or peanut butter	Pasta with pesto sauce
Oatmeal with whole milk, almonds, and fruit	Macaroni and cheese
Full-fat yogurt with fruit	Turkey burger with sliced avocado and tomato

Healthy snack and meal options

- Fill pita bread with hummus and vegetables
- Peanut butter-filled pretzels
- Trail mix
- Smoothie
- Crackers and cheese
- Yogurt parfait topped with granola
- Guacamole and tortilla chips
- Dried fruit (peaches, mangoes, raisins)
- Peanut butter on banana or apple slices
- Sports bar
- Oatmeal cookies and whole milk
- Mini whole grain bagels topped with cream cheese
- Add olive or canola oil to rice, pasta, or soup
- Turkey or chicken on whole grain bread topped with pesto sauce, cheese and/ or avocado, spinach, and tomatoes
- Cheese tortellini mixed with pesto
- Tuna or egg salad sandwich mixed with olive oil-based mayonnaise
- Salmon with olive or avocado oil
- Sweet potato mashed with butter; roasted potatoes made with olive oil
- Salad with toppings (cranberries, nuts, shredded cheese, dressing)
- Homemade pizza (whole grain dough with pesto or tomato sauce, olives, cheese)
- Hamburger, bison, turkey, or plant-based burger topped with avocado and cheese, served with sweet potato fries



Pair bananas and peanut butter for a healthy snack

Black bean–quinoa burgers with spicy mayo, avocado, and mango

Recipe from eatthismuch.com

Makes 4 servings.

Ingredients

1 cup quinoa	1 extra-large egg
1½ cups black beans	½ cup light mayonnaise
¼ c fresh cilantro	1½ tsp Old Bay Seasoning*
1 tsp ground cumin	1 tsp lemon juice
1 tsp coriander seeds	½ tsp hot sauce
¼ tsp cayenne pepper	1 tbsp olive oil
1 clove minced garlic	4 hamburger buns
Dash of salt	1 avocado
Dash of pepper	1 mango
1 tbsp arugula	

*Substitute for Old Bay Seasoning: Combine 1 tbsp celery salt, 3 whole bay leaves, ¾ tsp brown mustard seeds, ½ tsp black peppercorns, 10 allspice berries, 10 whole cloves, ½ tsp paprika. Grind mixture with a mortar and pestle or coffee grinder.

Directions

1. Preheat oven to 375 degrees F. Prepare quinoa as per package directions; set aside.
2. In a food processor combine beans, cilantro, cumin, coriander, cayenne, and garlic, and pulse until well combined but still slightly chunky. Transfer mixture into a large bowl and add cooked quinoa. Season with salt and pepper.
3. Add egg into bean mixture and mix until fully combined. Using your hands, divide mixture into 4 balls. Place on a large plate and gently flatten slightly to form patties. Cover with plastic wrap and transfer to the refrigerator for 10 minutes.



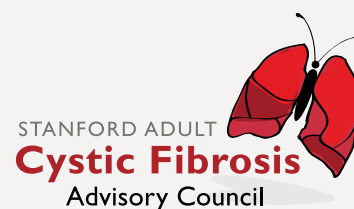
Black bean-quinoa burger

4. In a small bowl, whisk together mayonnaise, Old Bay Seasoning, lemon juice, and hot sauce, and season to taste with salt and pepper.
5. Heat olive oil in a large, ovenproof nonstick pan. Sear burgers for 2 minutes per side, then place in oven for 10–12 minutes (or until cooked through).
6. Serve burgers on hamburger buns with a dollop of spicy mayonnaise, and top with avocado, mango, and arugula. Enjoy!

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Stanford Adult Cystic Fibrosis Advisory Council

— Shawn Taylor

Items we are working on

Mask information: Developing an information pamphlet on appropriate selection and usage of masks based on health status and need.

Virtual clinic visits: Promoting the use of virtual tools to improve the quality of health care for CF patients.

Bodywork and CF: Researching how bodywork like massage and chiropractic can help relieve CF symptoms, by talking to people with expertise in the field and interviewing CF patients. Bringing together information for the CF community about easy ways to incorporate bodywork into their medical regimen.

Future activities

Stanford Health Care patient encounters: Capturing anonymous stories from patients on interactions with health care professionals, both positive and negative, which can be used as a teaching tool for caregivers.

Please join us: If you hear the call to serve your CF community and are interested in participating in the council, please contact our membership chair, Colleen Dunn, at cedunn@stanford.edu or visit our website at med.stanford.edu/cfcenter/advisory-councils/acfac.

Latino Family Resource Program

Programa de recursos para familias Latinas

– *Dani Rey-Ardila, Program Manager*

The Department of Family Centered Care launched the Latino Family Resource Program (LFRP) in August 2019 to address the needs of the Latino population at Lucile Packard Children's Hospital Stanford. The mission of LFRP is to close existing gaps in services and care for Spanish-speaking patients and their families.

LFRP was started following extensive discovery that included a review of the literature, parent interviews, and input from both front-line staff and leaders across the organization. This work brought to light the issues that Latino families face when they have a child admitted to Packard Children's, such as unmet resource needs, communication barriers during medical updates, and limited information about our existing services. The program also takes into account the special needs of Latino families who have children with cystic fibrosis.

In order to meet our families' needs, LFRP is focusing on three areas:

- Inpatient rounding
- Support for asylum-seeking families
- Broadcast studio activities

Inpatient rounding with monolingual Spanish-speaking families

At least three times each week, LFRP's manager meets with the monolingual Spanish-speaking families who have children admitted to Packard Children's. This service is provided in 10 units covering Acute Care, Intensive Care, and the Bass Center for Childhood Cancer and Blood Diseases. In March 2020, rounding in the Neonatal

Intensive Care Units was initiated. Inpatient rounds consist of providing important information to families about the hospital services most relevant to their situation and strategies to navigate the health care system. During rounds, we identify communication issues, discharge-planning barriers, and psychosocial needs. Any identified needs are escalated to the appropriate staff member.

Surveys with Spanish-speaking, English-speaking, and bilingual families

Through various surveys and polling of the inpatient population, we are learning about the diverse needs of monolingual Spanish-speaking families. A total of six surveys have been launched: Services and Places, Wayfinding, Communication, Technology, Hot Topics in the Latino Community, and Smartphone Utilization Among Spanish-Speaking and Bilingual Families. The survey results from more than 300 families have been used to continuously adapt and expand LFRP to best serve Latino patients and their families.

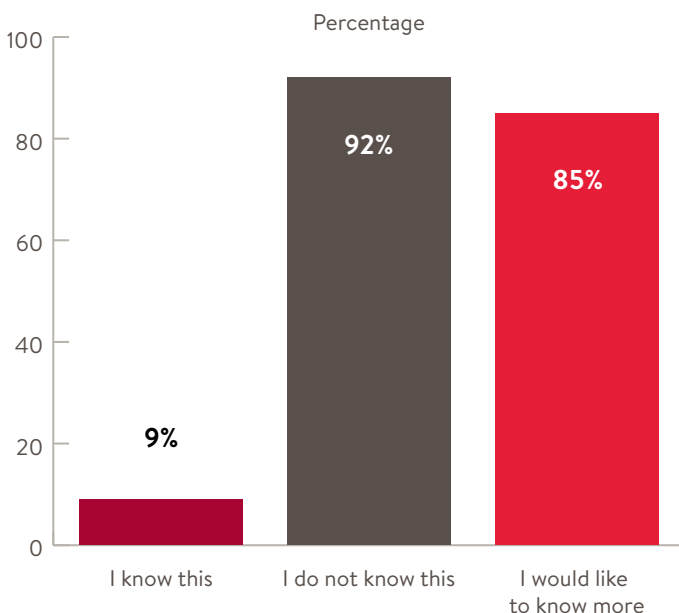
Inpatient Asylum Latino Initiative (ALI)

LFRP also provides a unique service to families who have come through immigration detention centers and may still be under active supervision by Immigration and Customs Enforcement. This initiative consists of a multidisciplinary team that includes a social worker, nurse coordinator, parent mentor, and medical director. LFRP provides care coordination and psychosocial support, as well as specific self-management tips concerning immigration, navigating the U.S. health care system, and information about life in the United States. LFRP continues to support families referred to ALI even after discharge from the hospital.

Broadcast studio spanish programming

One of the surveys showed that only 9 percent of monolingual Spanish-speaking families knew about the services of the Broadcast Studio. As a result, LFRP provides Spanish-language programming and activities every Thursday at 4:00 p.m. in the Broadcast Studio. All Spanish/English bilingual shows are on channel 71 of the GetWell TVs, and the most requested activity is Lotería (Bingo).

Sophie's Place knowledge among Spanish speakers



LFRP future endeavors

Although our current focus is on families of children who are admitted to our hospital, we understand that there are additional needs in the outpatient setting. There are plans to expand the Latino Family Resource Program to the Latino population who are routinely seen in our clinics and other outpatient services.

For more information about LFRP, contact **Dani Rey-Ardila** at Drardila@stanfordchildrens.org.

Tai Chi Training Videos

— *John Mark, MD*

The Tracie Lawlor Trust for Cystic Fibrosis (TLT4CF) was started in Wexford, Ireland, in 2008 as a memorial to Tracie Lawlor, who passed away at age 24 of CF. TLT4CF has since conducted research and provided resources for children and adults with CF throughout the world. They funded an acupuncture study in young adults with CF here at Stanford almost 10 years ago and have initiated a study utilizing tai chi. As part of their work to improve the lives of all children and adults with CF, they have made tai chi training videos available for all to use, which is part of their ongoing study. TLT4CF hopes to bring a mindset of hope to all affected by CF. Check out the videos here: <http://www.tracielawlortrust.com/2020/03/22/cf-cats-research-by-tlt4cf-free-training-video/>

Cystic Fibrosis Parent Advisory Council

— *Amy Baugh*

The Pediatric Advisory Council (med.stanford.edu/cfcenter/advisory-councils/pediatric-advisory-board) is a group of concerned parents whose children receive care from the Cystic Fibrosis Center at Stanford, who work in partnership with members of the pediatric CF clinic care team to provide the highest quality of care and service to patients and families. Do you have suggestions? Comments? Questions? Please don't hesitate to contact **Amy Baugh** at amycnbaugh@gmail.com or **Kirsten McGowan** at kmcgowan4@gmail.com. We'd love to hear from you!

Adult CF Center Update

— Jennifer Cannon, NP

In an effort to reduce COVID-19 transmission, the adult clinic in the Cystic Fibrosis Center at Stanford has transitioned to video visits in lieu of in-person clinic visits.

What is a video visit?

A video visit is similar to your regular in-person clinic visit. You will still be able to see the health care provider for this visit; however, it will be conducted via computer or smartphone.

What do I need for my video visit?

You will need either a computer or a smartphone; however, using a smartphone via the MyHealth application is strongly preferred.

Prior to your visit, you will receive instructions for accessing your visit through MyHealth.

You will need a photo ID.

If you have the supplies, it is appreciated if you can take your temperature, heart rate, blood pressure, weight, and/or oxygen saturation prior to your visit.

Currently, you must reside in California to participate in a video visit; however, if you live outside of California, you do not need to cancel your visit. We will convert your visit to a telephone visit.

What does this mean for my currently scheduled appointment?

Rest assured, our team will convert your currently scheduled appointment to a video

visit automatically. There is nothing you need to do to convert your visit.

What about my pulmonary function test (PFT)?

For the time being, the Stanford Pulmonary Function Lab is not performing PFTs on scheduled clinic patients. The need for a PFT will be determined on a case-by-case basis.

Our team is working with the Cystic Fibrosis Foundation to determine whether home spirometry can be used as an alternative. We will provide you with more information as it becomes available.

What about my sputum sample?

For the time being, we are not ordering sputum specimens for scheduled clinic patients. This will be determined on a case-by-case basis.

When will the adult clinic at the Stanford CF Center begin to offer in-person clinic visits?

We currently do not have an official date for when the CF clinic will resume in-person clinic visits. We continue to work with Stanford Health Care and public health officials to determine the most appropriate time frame.

We greatly appreciate your understanding and partnership in helping ensure the safety of our cystic fibrosis community. We look forward to providing you care virtually at your scheduled appointment.

*Nontuberculous...**continued from page 4*

approximately 10 percent of the CF population seen at Stanford has NTM lung disease.

How is NTM lung disease diagnosed?

Diagnosis of NTM lung disease relies on three components: clinical symptoms, imaging, and positive sputum cultures. Symptoms of NTM lung disease include worsening cough and sputum production, coughing up blood, increasing frequency of CF exacerbations, unexplained decline in lung function, weight loss, fatigue, low-grade fevers, or night sweats. New changes on imaging are usually evaluated with a CT scan of the chest. Since NTM are common environmental organisms, sputum cultures can be contaminated—lung disease is suspected only when repeated sputum cultures are positive. Sometimes, differentiating between being a carrier of NTM (colonized) and having active lung disease can also be difficult. It is important that you talk to your CF providers about any changes in respiratory symptoms.

How do we screen for NTM lung disease?

Our goal at the Stanford CF clinics is to screen for NTM lung disease with a sputum sample at least once yearly. If your sputum sample returns positive, you will be asked to provide additional sputum samples for closer monitoring. Screening for NTM can only occur with a coughed up sputum sample. We have noticed that the amount of sputum produced by many patients decreases after they start modulator therapies such as Trikafta. Because of this, you may be asked to provide an induced sputum sample (breathing in hypertonic saline to aid in coughing up sputum) for both screening and monitoring of disease progression.

How is NTM lung disease treated?

It is important to know which type of NTM caused your lung disease, since treatment is different for

different species. NTM treatment requires three or more antibiotics for usually greater than a year in duration. Lung disease caused by *Mycobacterium avium* complex is usually treated with three oral antibiotics; lung disease caused by *Mycobacterium abscessus* will also include an initial period of IV antibiotics before transitioning to maintenance inhaled and oral antibiotics. These antibiotics commonly include azithromycin, ethambutol, rifampin, amikacin, imipenem, cefoxitin, and linezolid. Due to side effects related to certain antibiotics, you may be asked to obtain a vision and hearing exam before treatment is started.

There are many drug interactions with these antibiotics and common CF medications. Our center has a dedicated CF pharmacist (Russell Wise, PharmD) at the pediatric center as well as a CF infectious diseases specialist (Joanna Nelson, MD) at the adult center, who help with monitoring of NTM therapy. You will be regularly monitored using blood work, sputum samples, and spirometry while on treatment. Treatment typically stops after your sputum cultures are repeatedly negative for NTM for about a year. In addition to antibiotics, airway clearance and overall good CF care are key components of NTM therapy.

Is there any research on NTM lung disease at Stanford?

There is an ongoing study looking at why NTM causes different degrees of lung disease severity in different people, with the hypothesis that an individual's immune response to this organism plays a major role. In our CF clinics, we are actively collecting sputum samples of patients with a history of positive NTM sputum cultures. Enrollment in the study is optional and includes obtaining a series of sputum samples over time.

Build Your Own Home Exercise Program

– Taylor Lewis, MA, CMT, CSCS, PRT

Don't let limited space hinder your ability to work out or exercise. During the current COVID-19 pandemic, it is hard to get outside without also having the fear of getting sick. However, it should not stop you from getting in some form of exercise. Exercising can burn unwanted stress and can help you feel rejuvenated. Having your own home workout plan is a great way to occupy some time, it is an opportunity to have fun with your significant other, or you can even set up an online workout session with your best friend.

Science has shown that increasing your heart rate and ventilation, and moving in multiple directions under stress, are great for the body. This also means that you do not need a gym to get a good workout in. All you need to do is get creative, integrate full body movements, change up the intensity and duration of your workouts, and, most important, show up and stay consistent. A quality workout consists of a warm-up, strength and/or conditioning exercises, and a cool down.

Warm-up

Performing a 5-to-15-minute warm-up before you start strength training or endurance training helps reduce the risk of injury by preparing the muscles and joints for what you are about to do in your training.

Strength and/or conditioning

If you are working out several days a week (four to five), integrate a couple of days of strength training and a couple of days of endurance training. If you are limited to two to three days a week, do strength training and conditioning in each workout.

Cool down

Always integrate a five-to-10-minute cool-down. Taxing the body during a workout increases stress on the lungs, muscles, and heart. Allowing the body

to down-regulate through low-intensity stretching and breathing is important in the transition from exercise to work and/or family responsibilities.

Now, let's focus on exercises you can do at home that you can integrate into your workout:

Push-ups are a great exercise to target the upper body (chest, shoulders, triceps) and core, all at the same time. To build strength, you want to keep the reps low to medium and complete six to 12 repetitions. Once you can complete three to four sets of 12 reps, increase the difficulty by lifting one leg off the ground, alternating legs off the ground, or elevating the feet onto a chair or step.

We want to make sure to work the whole upper body, and while push-ups target the front side of the upper body, you also want to target the back side of the upper body. If you have weights at home, you can never go wrong with a one-arm row. However, not everyone has weights, so if you do not have them, try seated wall slides as an alternate option. If you are using weights, go eight to 12 reps of three to four sets, and if you are performing wall slides, do as many reps as possible for the same amount for three to four sets.



Push-ups



Wall slides

Now that the upper body is taken care of, we need to focus on the lower body and core. Lunges, rear-foot-elevated split squats, and hip hinging exercises such as deadlifts and hip thrusters are great for targeting all aspects of the legs. They work the quads, glutes, and hamstrings in a synchronized fashion. For the core you can't go wrong with performing planks, side planks, bird dogs, or bear crawls. Your core is built to control your body's movement by slowing down how much force is distributed to the spine during movement. What this means is that performing bracing exercises like planks, side planks, and bird dogs will target many of your core muscles at once. Increasing core strength could help improve breathing.

We can't forget about the endurance aspect of exercise. First, you need to think about what your goal is and, most important, what you have been cleared to do. If you have been cleared to perform any form of exercise training, then you have two options: anaerobic-based training or aerobic-based training. One focuses on short-duration (anaerobic) training, and the other focuses on longer-duration (aerobic) training. If you are trying to achieve conditioning and you do not have a lot of time (less than 15 minutes), think about doing interval training. This means you would pick a couple of exercises that elevate your heart rate, such as, but not limited to, jumping jacks or side shuffles, and perform them for a particular amount of time,

then rest and repeat—for example, 20 seconds of jumping jacks, 40 seconds of rest, 20 seconds of side shuffles, 40 seconds of rest; and repeat. You would do this for four to 10 rounds, or five to 10 minutes, and then you would cool down.

If you decide to do intervals, start with a work/rest ratio that has you resting more than working. For example, complete a 1:4 ratio of work to rest (20 seconds of work and 80 seconds of rest). When that starts to get easier, decrease the rest but keep the work the same, so now the ratio is 1:3 (20 seconds of work, 60 seconds of rest). The goal should be to improve the recovery between sets. When you start with a higher amount of rest, it allows your body to get used to the stimulus and keep the quality of work higher.

If you are more of a long-duration exercise enthusiast, taking short walks throughout the day allows you to get outside and will add to the volume of exercise per day—for example, a 15-minute walk in the morning, a 15-minute walk after lunch, and a 15-minute walk before or after dinner. That equals 45 minutes of aerobic training. Walking is a very powerful aerobic mechanism that doesn't get the recognition it deserves. Walking is low-impact, which means you can do more over time while limiting the stress on the joints. Walking can improve cardiopulmonary performance (decrease resting heart rate, lower



Lunges

blood pressure) and even increase your creative productivity (Oppezzo & Schwartz, 2014).

If you want to dial it up a little bit, you can take the interval mentality and perform a walk-jog/run. Walk-jog/run gives you the opportunity to push the limits of a walk and introduce a higher stressor when you do not want to or do not have the capacity to run consistently at a particular pace. Once you are warmed up and ready to go, start by jogging or running for a particular amount of time (20 seconds), then walking for the rest (60 seconds), then repeat; you do this for four to 10 rounds. This allows you change it up and integrate jogging/running with your walking.

Last but not least, make sure you integrate recovery days into your weekly workout routine. A recovery day could involve light stretching, going for a walk, or meal prepping for the days ahead; and some of the most beneficial choices are sleeping and relaxing. When you work out, you stress the body. The more you stress the body, the more time the body needs to recover. All the positive results from exercising come from allowing the body to recover and rebuild. Having adequate rest between workouts will allow the brain and body to recharge and reboot to give them a better platform to push the limits next time.

Just remember to integrate full body exercises into your warm-up, work out, and make sure to cool down. If you are looking to increase the difficulty in the strength portion, start by making each rep slower. For example, if it takes you three seconds to do a push-up, then perform a six-second push-up by descending for three seconds and ascending for three seconds. The next phase is increasing the number of reps (start with eight, and then go to 10, then 12); then increase the difficulty of the exercise, elevating the hands or feet (feet elevated push-ups) or making the exercise dynamic (bear crawls), or performing a single-leg or single-arm version of the exercise (single-leg push-ups, single-leg deadlifts). Whatever exercise you decide to incorporate, make sure you keep the quality of movement at the highest standard. As the muscles fatigue, they are going to try to recruit other muscles to help out. This can cause faulty movement patterns and over time can lead to stiffness and unwanted soreness in the joints. Quality always trumps quantity when training.

Most important, stay consistent. Progress takes time, and staying consistent and doing some form of exercise throughout the week will set you up for bigger gains (e.g., increased strength, endurance, quality of life, lung function) down the road.

Link to sample workout:

<https://www.youtube.com/watch?v=IFmenVA4M-g>

Reference

Oppezzo, M., and D.L. Schwartz. 2014. Give your ideas some legs: The positive effect of walking on creative thinking. *Journal of Experimental Psychology* 40(4): 1142–52.



Planks

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 Zirbes, DNP, RN, CPNP

Clinic Scheduling(650) 724-4788

Clinic and Prescription Refill Fax(650) 497-8791

Patient Services Coordinator(650) 498-2655

Nurse Coordinator: Mary Helmers.....(650) 736-1359

CF Clinic Nurse: Liz Beken.....(650) 736-1359

Respiratory Therapy: Candice Middleton(650) 724-0206

Nutrition: Julie Matel.....(650) 736-2128

Social Work: Teresa Priestley.....(650) 736-1905

Newborn Screening: Jacquelyn Zirbes(650) 721-1132

Pharmacy: Russell Wise, PharmD.(650) 724-4788

Clinical Psychology: 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 Children's Health

Specialty Services – Emeryville

Providers: Karen Hardy, MD; Eric Zee, MD;

Manisha Newaskar, MD; and Rachna Wadia, MD

CF Clinic Scheduling.....(650) 724-8414

Clinic and Prescription Refill Fax(510) 457-4236

Nurse Coordinator: DJ Kaley, RN.....(650) 724-8414

Respiratory Therapy: Lorraine MacPhee (Tues–Fri)

.....(650) 587-9631

Nutrition: Ayah El-Beshbeeshy (Tues & Thurs)

(leave message with DJ Kaley, RN)(650) 724-8414

Social Work: Cleo Rice-Hodge (Tues, Thurs & Fri a.m.)

.....(510) 362-7504

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: Laveena Chhatwani, MD

Providers: Gundeep Dhillon, MD; Jennifer Cannon, NP;

Elika Rad, NP; Meredith Wiltse, NP

Backup Providers: Julie Hoang, NP; PA; Jenny Christensen, PA;

Feifei Yang, NP; Amy Milliken, NP

Adult Clinic Scheduling(650) 725-7061

Adult CF Center Fax(650) 723-3106

Nurse Coordinators: Theresa Kinney, RN

and Kristel Fallon, RN.....(650) 498-6840

Respiratory Therapy: Gauri Pendharkar, RCP

.....(650) 736-8892

Registered Dietitian: Michelle Stroebe, MS, RD

.....(650) 529-5952

Social Work: Meg Dvorak, LCSW.....(650) 518-9976

Social Work: Kate Yablonsky, MSW(650) 444- 6512

Mental Health Coordinator: Liza Sher, MD

Routine issues/concerns during business hours, 8 a.m. to 5 p.m.

CF Nurse Coordinator Line(650) 498-6840

• Please leave a voicemail if no answer. These calls will be answered within 24-48 business hours

• Alternatively, you can utilize MyHealth messaging. These messages will be answered within 24-48 business hours. This is NOT to be used for urgent issues. MyHealth is NOT checked on the weekends

Urgent issues/concerns during business hours, 8 a.m. to 5 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) 723-4000

• Nurse triage is available 4:30 p.m. – 7:30 a.m. A message will be generated and sent to the CF Team ASAP

Adult providers at Sutter Health CPMC

Adult Center Director: Ryan Dougherty, MD

Associate Center Director: Vinayak Jha, MD

Providers: 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

Colleen Dunn, Zoe Davies, Sean Ryan, Jackie Zirbes

.....(650) 736-0388

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

New Staff Members



Pediatric Clinic

Jessica King, RRT, NPS, AS, a Bay Area native, became a part of the Stanford family 11 years ago while working in the division office for Pediatric

Gastroenterology, Hepatology and Nutrition. During that time, she discovered her passion for working with patients and their families, which led her to pursue the career path of respiratory care. Most recently, Jessica has worked with a variety of patient populations in both the acute and ICU levels of Lucile Packard Children's Hospital Stanford. During her free time, she enjoys cooking, gardening, and fostering animals in need. She is excited to transition over to the Pulmonology Clinic as the new CF Respiratory Coordinator and looks forward to building new relationships within the CF community.



Adult Clinic

Kristel Fallon was born in the Philippines, and earned her baccalaureate degree from University of Baguio in 2007. Kristel applied her knowledge of cultural competence and nursing

skills in various care settings such as hospice, home care, assisted living and hospitals. With her passion in teaching, she earned her teaching credentials in 2014, mentoring and teaching nursing assistants. Kristel joined Stanford in 2016 as a RN Transitional Coordinator at Aging Adult Services, then transitioned to the Stanford Cystic Fibrosis Clinic in April 2020 as a CF Clinic Nurse Coordinator. In her free time, Kristel enjoys dancing, eating new foods, video editing, and photography.

Newsletter Contact Information

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

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