Winter 2020

Stanford Children's Health | Lucile Packard Children's Hospital Stanford

Stanford MEDICINE

Cystic Fibrosis Center **News**

Wildfires and Lung Disease

—Karen Hardy, MD

Sadly, with climate change, the Western United States suffers a fire season every year. This occurs when hot, dry winds coincide with very dry landscape and spontaneous fires as well as those caused by lightning strikes, electrical mishaps, fireworks, arson, etc. These fires are extremely dangerous and result in loss of life, loss of property, and very poor air quality.

All persons, especially those with lung diseases, need to know about air quality importance, monitor the air quality in their area, and respond appropriately to the air quality index (AQI). It is helpful to have an app on your smart-phone or computer where you can easily access this information:

- **IQAir AirVisual:** This app's icon is a square with a sketched face of a person wearing a mask in the middle of a blue background. AirVisual displays results from monitors throughout the world and, based on GPS, will access the data from the monitor closest to your position at any time. The app runs very much like a weather app, showing a large banner that is colored to display the current status of the air quality and a seven-day forecast providing more detail about what to expect in the coming week. The banners correlate to the air quality index. This is arranged as shown below and helps you know how to respond on certain days.

Continues on page 2

In this issue

Wild fires and...continued from page 1

- **AirNow:** From the U.S. Environmental Protection Agency (EPA), this is a simpler app, showing the AQI with corresponding color for the day and the coming week, and is restricted to the United States.

Zone	AQI	What it means
Green	0 – 50	**Good:** May keep windows open, play outside.
Yellow	51 – 100	**Moderate:** Close windows if able, especially over 75; no running outside; minimize walking outside; must be wearing a mask all the time if outside.
Orange	101 – 150	**Unhealthy for Sensitive Groups:** Close all your windows; don't go outside at all if possible; use an N-95 or KN-95 mask if you have one all the time you are outside if you must go anywhere. No exercising outside at all. Use an air filter inside.
Red	151 – 200	**Unhealthy:** Use same advice as for the orange zone.
Purple	201 – 300	**Very Unhealthy:** Do not go outside. Use air filters inside your home.
Maroon	301–500	**Hazardous:** Use same advice as for the purple zone.

***Please note that during the COVID19 pandemic masks should be worn outside regardless of the air quality index reading.**

EPA AirNow Air Quality Index (AQI) Basics (https://cfpub.epa.gov/airnow/index.cfm?action=aqibasics.aqi): This is a helpful color-coded chart you can print and post somewhere in your home.

It is a good idea to keep your car on recirculation of internal air all the time during fire season unless we have a green day. After fire season, you should get your car filtration checked, and cleaned or replaced as appropriate.

Do your part to help California conserve resources: plant drought-tolerant plants in your yard; if you replace grass, you may be eligible for a rebate; and of course, don't waste water!

If you would like more information about the effects of fire smoke:

"Health Effects of Wildfire Smoke in Children and Public Health Tools: A Narrative Review" (https://www.nature.com/articles/s41370-020-00267-4)

"Protecting Children From Wildfire Smoke and Ash" (https://www.pehsu.net/_Library/facts/PEHSU_Protecting_Children_from_Wildfire_Smoke_and_Ash_FACT_SHEET.pdf)

"Protect Your Health During Wildfires" (https://www.lung.org/getmedia/695663e2-bdb8-4a61-9322-02657f530b99/protecting-lung-health-during.pdf.pdf)

Path to a Cure—Many Routes, One Mission: Genetic-Based Therapies

—Sean Ryan, RRT, CCRC

It is an exciting time for the cystic fibrosis community. Over the course of decades, we have gone from treating the signs and symptoms of CF to finally treating the basic defect. The 2019 approval of the highly effective modulator Trikafta has given the majority of our community the hope and promise of finally living a healthy, normal life. Data across the board has been remarkable: improved quality of life and lung function, weight gain, and decreases in exacerbations and hospital stays. In fact, patients report feeling so well that the Cystic Fibrosis Foundation (CFF) is funding a study called SIMPLIFY, which will determine if hypertonic saline and/or dornase alfa can be reduced or eliminated from daily therapy. However, the work does not stop there, as the CFF has committed $500 million towards the Path to a Cure initiative.

Currently, the CFF is working with 40 companies toward genetic-based therapies and has already funded 15 industry programs. The ultimate goal is to provide a cure for everyone with CF regardless of their CFTR mutation, but how exactly? What we know is that CF is a single-gene disease, which makes it an ideal candidate for genetic-based therapies. Additionally, the lungs are a large organ that is easily accessible to therapies. The question scientists have faced for years since the discovery of the mutated CFTR gene was, How do we repair, restore, and replace the CF gene? Fortunately, dedicated research and breakthrough discoveries are providing the answers we have sought for so long. The 2020 North American Cystic Fibrosis Conference gave us an opportunity to see what progress has been made toward a cure—specifically, the methodology and the challenges we are currently facing.

Two potential therapies toward a cure are RNA therapy and gene (DNA) therapy. Both approaches can deliver a correct copy of the genetic instructions to cells in the lungs. With the correct messenger RNA or DNA sequence, cells would be able to produce normal CFTR protein that moved to the cell surface, where it could adjust the amount of salt and fluids flowing through the cell membrane. Over time, the correct copy of the genetic instructions is degraded or lost, as the cells naturally die and are replaced. Thus, regular repeated treatments would be needed to ensure that enough correct CFTR protein was present to hydrate mucus.

Gene editing and cell-based therapy are also potential therapies. Gene editing would permanently repair the CFTR gene mutation in cellular DNA. The goal is to specifically target basal cells in the lung because they have the ability to divide and differentiate into other cell types to replenish lost cells. As cells died off, the basal cells with permanently repaired DNA would then generate new cells that contained the correct CFTR sequence. If enough basal cells were successfully edited, cells with normal CFTR would constantly be supplied to the airway surface. Repeat treatments would be rarely required, if ever. Cell-based therapy involves removing cells from the airway to create space for the delivery of gene-edited cells. Cells would be taken from the same patient, corrected via gene editing in the lab, and then delivered into the airway, where they would generate new basal cells and specialized cells that create CFTR protein. Much like gene editing, this approach would permanently restore CFTR function in people with CF.

Continues on page 7

Nutrition for Individuals With Cystic Fibrosis During COVID-19

—Julie Matel, MS, RD, CDE

Optimizing nutrition and maintaining a healthy weight can sometimes be difficult for people with cystic fibrosis. Add to that issues surrounding living in the COVID-19 pandemic, and the struggle to stay well nourished may be even more challenging. During these times, depending on individual health circumstances and medication management, individuals with CF may struggle with either under- or overnutrition.

More widespread treatment with modulator therapies, including Kalydeco, Orkambi, Symdeko, and the most recent FDA-approved highly effective modulator, Trikafta, have had a welcomed benefit of making it easier for some individuals with CF to gain weight. In a study looking at treatment for three months with ivacaftor (Kalydeco), researchers found a decrease in energy expenditure, improved fat absorption, and decreased gut inflammation, which may provide a possible mechanism for weight gain.1

Although improved weight gain while taking modulator therapies occurs, it is not everyone's experience.2 Individuals may continue to struggle with inadequate weight gain, while others are trying to manage weight gain that may lead to overweight or obesity. Make sure to track and discuss individual weight trends with your CF Center dietitian before implementing significant diet changes.

Existence in a pandemic may contribute to nutrition challenges related to stress, contributing to poor food choices, food insecurity related to financial hardship, or simply the inability to feel safe grocery shopping. In addition, the ability to exercise may be limited due to sheltering indoors, loss of access to gyms, or simply losing connection with an exercise partner.

While nutrient and calorie needs vary among individuals with CF, there are many diet and exercise strategies that have a broader benefit. The following tips and resources may help individuals meet nutrition and exercise goals during this unusually stressful time!

Ideas for healthy living during a pandemic:

Make half of your daily grain choices whole grain

- Choose cereal with at least 5 g of fiber per serving.
- Choose breads with "whole grain" in the title and with "whole grain" as the first ingredient.
- Give these five whole grains a try: amaranth, barley, oats, quinoa, teff.

Choose more fruits and vegetables

- Choose brightly colored varieties (orange, red, purple, dark green) for valuable antioxidants, vitamins, and minerals to preserve immune function.
- Focus on whole fruits rather than juice (they provide fewer calories and no added sugar); when drinking juice, look for 100% juice.
- Keep frozen, canned (packed in its own juice), and dried fruit and vegetables on hand for convenience. These items have the benefit of lasting longer during a pandemic!
- Choose five servings a day for optimal health (serving sizes: 1 tennis-ball-size piece of fruit, 1 cup of raw veggies, ½ cup of cooked vegetables).

Get plenty of exercise

- Turn on some music and just dance (like no one is watching!).
- Search YouTube for exercise videos (e.g., Yoga with Adrienne, PopSugar, Self).
- Resources for kids, including exercise challenges and videos: Active Schools (http://www.activeschoolsus.org and https://www.activeschoolsus.org/wp-content/uploads/2020/06/Resources-for-Kids-to-Stay-Active-at-Home-6.19.20.pdf).
- CF-specific exercise ideas: CFF Fitness (https://www.cff.org/Life-With-CF/Daily-Life/Fitness-and-Nutrition/Fitness/), Beam Cystic Fibrosis (https://beamfeelgood.com/cystic-fibrosis).

If you need extra calories for weight gain, try these healthy high-calorie tips

- Add oil instead of butter (such as canola, olive, avocado) to sauces, soups, rice, and vegetables.
- Choose high-calorie fruits and vegetables such as avocado, olives, dried fruits, smoothies, banana or apple with nut butters.
- Choose these high-calorie foods for meals and snacks: salmon, tuna packed in oil, sweet potato fries, yogurt with granola, nuts, hummus and vegetables, trail mix.

Ideas for navigating the grocery store in a pandemic

- Keep a well-stocked pantry and freezer with shelf-stable items to reduce grocery store trips (dried or canned beans, peas, and lentils; canned or frozen vegetables/fruits; oats and other grains; cereal; rice; nuts, seeds, nut butters; pouches or cans of fish or chicken, oils, dried herbs and spices).
- Plan your grocery trip before you go (organize food list according to store layout).
- Bring wipes and hand sanitizer to wipe down grocery cart handle and hands before entering the store (most stores have these items available at the store entrance or wipe carts down between each customer).
- Wash hands when returning home.
- Rinse produce (even if the peel will not be eaten) under running water and dry with a paper towel before storing. Firm produce, such as melons or cucumbers, can be scrubbed with a clean produce brush. Soap, bleach, or commercial cleaning products should never be used when washing fresh produce.

Food resources

- Local food banks, SNAP (Supplemental Nutrition Assistance Programs), WIC (Women, Infants, and Children), and local school lunch programs all provide food assistance to struggling families.
- CF-specific programs, such as HealthWell and enzyme assistance programs.
- Your CF Center social worker can provide area-specific resources to help with food assistance.

References

[1] Stallings VA et al. Energy balance and mechanisms of weight gain with ivacaftor treatment of cystic fibrosis gating mutations. *J Pediatr*. 2018; 201:229–237e224.

[2] Bailey J, et al. Effect of CFTR modulators on anthropometric parameters in individuals with cystic fibrosis: An evidence analysis center systemic review. *J Acad Nutr Diet*. June 9, 2020; S2212–2672.

Home Spirometry During COVID-19

—Gauri Pendharkar, RCP

Pulmonary functions tests (PFTs) are a very important part of every patient's clinic visit. COVID-19 presented a new challenge for our cystic fibrosis patients because we had to stop in-person clinic visits for a few months. During the months of video visits, patients did not have PFTs, which made it difficult to get a good assessment of how patients were doing.

The Cystic Fibrosis Foundation (CFF), in conjunction with ZephyRx, allocated home spirometers to centers all over the United States so that patients would have the ability to do a PFT at home in order to assess their lung function. The Stanford Adult CF Center has been able to get approximately 90 patients a home spirometer (some of our patients had previously self-purchased spirometers before the COVID-19 pandemic). This has helped us in understanding the lung health of our patients and guided us in treating patients accordingly.

This MIR SpiroBank Smart spirometer, provided by the CFF and ZephyRx, is very easy to use. It works with batteries and is the size of a TV remote, making it easy to hold and use to do a self-test.

How does this work?

The patient downloads the Breathe Easy App on his or her smartphone and then enters his or her height, date of birth, and gender in the Demographics section. There is a dropdown menu from which to select the CF center to which the patient is affiliated.

The app pairs with the spirometer and then gives the patient instructions on starting the test. The steps in using the home spirometer are as follows:

1. Put the device in the mouth.
2. Take a fast, deep breath in.
3. Blow out as fast and hard as you can, and keep blowing for six to seven seconds.
4. Take another fast, deep breath in to end the test.
5. Contact your respiratory therapist if you are having trouble doing the spirometry.

MIR SpiroBank Smart spirometer

ZephyRx app screen

When should I do a home spirometry?

1. Ideally the patient should do a PFT in the morning after his or her first respiratory treatment. It is not necessary to do a PFT every day or several times a day, as it may cause fatigue.
2. We recommend doing three test attempts; the device will pick the best test as your result. A screen shot of this test should be sent to the CF team via MyHealth, and then we can input the data in the patient's medical record.
3. Now that we are seeing patients in person in clinic, the home spirometers are a great tool to have for patients to check their PFT between clinic visits or if they are not feeling well.
4. If a patient has a video visit scheduled, he or she should do a home spirometry the morning of the video appointment. This will help the care team to assess lung health and provide appropriate treatment if needed.

Will the home PFT be the same as in clinic?

While the method is the same as in clinic, it is a shorter and quicker version of the test as compared with a PFT in clinic. It is important to get the demographic data entered correctly and use proper technique to get accurate results. If the test is set up correctly, we see PFT values correlate very well to the numbers in clinic.

Going forward, home spirometers are a great tool for our patients to have to assess their lung function remotely. We thank the CFF and ZephyRx for their generosity in helping our patients during this pandemic.

Path to a Cure...
continued from page 3

As exciting as these advancements sound, there are also barriers facing genetic-based therapies. One is a physical barrier, the thick and sticky mucus layer in the airway. This mucus layer can prevent the entry of foreign inhaled particles into the cell, thus preventing translation of the corrected gene. Second, at the top of the airway epithelium lies the cilia, hairlike structures that line the bronchus, which coordinate the mucociliary clearance, a mechanism by which the lungs clear anything inhaled that is foreign and potentially damaging to the airway. There are also biological barriers to effective airway-directed gene therapy. Once the gene bypassed the sticky mucus layer and entered the airway cell, it could lead to an immune response that resulted in the activation of antibodies that worked against CFTR and the gene therapeutic, ultimately destroying the translated cell. Lastly, defining the airway-specific formulations that can effectively deliver these inhaled therapies with the correct delivery device continues to be studied to provide the most optimal form of therapy.

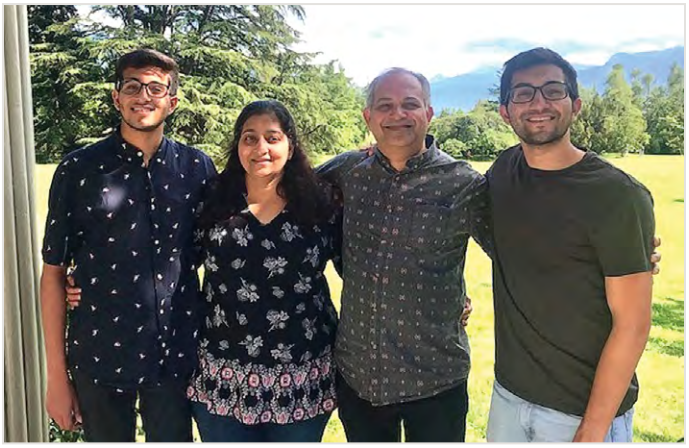
Despite the noted barriers, great strides have been made over the course of the year, providing extraordinary insight into the biology of the CF airway. These discoveries have opened doors to therapies that seemed unattainable just a few years ago. Much work remains to be done, but the path to a cure seems closer than ever. With the continued effort and dedication by scientists, researchers, and, most important, our CF community, there is now hope that one day we can live in a world free of cystic fibrosis.

Cystic Fibrosis Among People of Indian Origin: The Need for Heightened Awareness

—Siri Vaeth, MSW, Executive Director Cystic Fibrosis Research, Inc. (CFRI)

Harini and Sesh Tirumala's first son was born in 1995, seemingly in perfect health. Yet Srivas was not growing. When Harini questioned why Srivas had not gained weight between the age of 3 and 4 years, the pediatrician was initially not concerned. But when her son began complaining of excruciating stomach and gastrointestinal cramps, an ultrasound was ordered. The results led to an appointment with a gastroenterologist and a nine-month diagnostic journey. After viruses were excluded and prescribed medications were found to be ineffective, the G.I. physician observed that Srivas frequently had a cough and sinus issues. A sweat test was ordered, and at the age of 6 years, Srivas was diagnosed with cystic fibrosis (CF) and referred to Stanford, one of only a small number of CF patients of Indian origin.

Until that moment, Harini and Sesh did not know anything about cystic fibrosis. "It was a mix of fear and denial at that point," says Harini, "more about information processing than emotion. I was confident that it was not something to be that concerned about. The medical articles that I read did not indicate CF being prevalent in Asians or Indians, and I felt that there was some sort of mistake in testing." After Harini and Sesh reached out to a pulmonologist in India, who confirmed that there were indeed CF cases in their native country, reality began to set in. When their second son, Abhijit, was diagnosed with CF two years later, any remnant of denial was gone. It has now been twenty years since her older son's diagnosis and Harini shares, "My family and I are very grateful for all the wonderful care and



Tirumala Family

support that we have received from the Stanford CF team, be it at the clinic or at the hospital."

Cystic fibrosis is relatively rare in Asian populations. While 1 in 2,500–3,000 Caucasians per year are diagnosed with CF, the rate for all Asians is believed to be 1 in 35,000. It is hypothesized that this number is underestimated in many countries due to a lack of testing and socioeconomic conditions that lead to high rates of disease that mask CF symptoms, including malnutrition, dysentery, and tuberculosis. While the term Asian is used broadly, there is a wide range in rates of cystic fibrosis based on country of origin, with many studies indicating that rates are higher among those of South Asian (from India, Pakistan, Sri Lanka, Bangladesh, Nepal, Bhutan, and Afghanistan) descent than those of East Asian (China, Japan, South Korea, Taiwan) origin. Indeed, the estimated prevalence of CF in South Asia has been estimated to be approximately 1 in 10,000 people compared with approximately 1 in 100,000 people in East Asia.

These numbers hold true at Stanford, where Sriram Vaidyanathan, PhD, and Zachary Sellers, MD, PhD, are conducting research on incidence rates and mutation status among Asian CF patients. According to their research, currently only 4% of the 429 CF patients at Stanford (both pediatric and adult) are identified as Asian. As shared by Dr. Vaidyanathan, the 2010 U.S. census found that within the five San Francisco Bay Area counties neighboring Stanford, Asians make up 17%–38% of the population and that South Asians make up approximately 5% of this total.

These numbers are not reflective of the patient population at Stanford Hospital and Lucile Packard Children's Hospital Stanford, where more than 80% of Asian patients are of South Asian origin. After identifying patients who were treated at Stanford in the past, Dr. Vaidyanathan and Dr. Sellers found that in total, 29 Asian patients (24 South Asian and five East Asian) with CF (or CFTR-related metabolic syndrome—CRMS) were treated at Stanford between 2000 and 2020.

Dr. Vaidyanathan and Dr. Sellers found that more South Asian patients were affected by the F508del mutation (40%) than East Asian patients (20%), noting, "These results are consistent with our analysis of literature since both CF and the F508del has been reported much more frequently in South Asians." They found only one report of the F508del mutation in a patient of East Asian origin. Many of the mutations present in the patients studied would never be detected through California's newborn screening panel. As described in their research abstract shared at the recent North American Cystic Fibrosis Conference, "In our cohort, 40%–50% of alleles present in South Asian and East Asian patients were not part of commonly used screening panels, and we identified 20%–30% of patients had mutations in both alleles that are not part of common screening panels. Our data suggests the need for additional awareness of CF and improved characterization of CF in the South Asian and East Asian population."

The need for heightened awareness and improved diagnostics is shared by clinicians, researchers, patient advocacy groups, patients, and patient families. Dr. Vaidyanathan became interested in investigating CF in people of Indian origin in 2018 after learning about Indian patients from Jackie Spano, PNP, PhD. When he was next in India, he spoke to a hospital pediatrician who was treating approximately 30 CF patients at the time. This furthered his awareness that CF in Indians might be underrecognized and exposed the need for further investigation. Dr. Vaidyanathan reached out to Carlos Milla, MD; Jeff Wine, PhD; Dr. Sellers; and Dr. Spano.

Thanks to the Stanford CF care team's commitment to research that results in improved patient outcomes, and to their engagement with the broader CF community, a meeting was convened, which also included Harini and Sesh Tirumala and CFRI's executive director. Efforts are now focused on exploring ways to update information on the prevalence of CF in North American patients of Indian origin. In addition, the group seeks to build a network of families of Indian origin living with CF in order to determine unmet needs as well as to create tools and resources to educate, raise awareness, and provide support.

Cystic fibrosis has long been perceived as a disease of people of European descent, and this outdated perception may lead to misdiagnosis among children of Indian origin. Dr. Vaidyanathan and Dr. Sellers' work emphasizes the importance of expanded research in this area. Because those with CF of Indian origin are more likely to have mutations that may be missed by newborn screening panels, it is vital that CF clinicians, researchers, patients, families, and CF organizations work in partnership to raise awareness and advance understanding of cystic fibrosis among this community.

The Impact of Exercise on Mental Health

—Taylor Lewis, MA, CSCS, CMT, PRT

Mental health is a very important component that affects many aspects of our daily life. The accumulation of acute and chronic fluctuations in emotions and behaviors plays an important role in how energy (mental and physical) is utilized during daily activities. The human body is continuously burning energy to keep us alive. Every thought, breath, and movement we do requires a level of energy requirement. Since the emotions and behaviors we express utilize energy, the accumulation of energy expended directly plays a role in an individual's motivation and determination to stay physically active.

The regulation and balance of an individual's mental health is a very important component when discussing mental and physical health. Chronic and acute fluctuations in emotions and behaviors can play a toll on energy levels in cystic fibrosis. Individuals with CF are two to three times more likely to experience bouts of depression or anxiety compared with people in the general population ("Cystic Fibrosis Foundation" n.d.). The level and magnitude of the impact that mental health takes on an individual with CF can't be categorized or expressed in a value that is sufficient to tell each individual story. Mental health is unique and specific to each person. Problems with it can't be solved using a broad-brushed approach. Exercise or physical activity, however, is a tool that people can use to manage and address mental health during day-to-day life. Exercise improves properties within the body that regulate mental health and can help build a strong foundation to help aid symptoms of stress, depression, and anxiety.

Across many populations and ages, exercise has shown to decrease symptoms of stress and improve mental health benefits (Warburton, Nicol & Bredin 2006). What is not often considered is that mental health can also affect physical health. Research has shown that individuals who consistently experience depression are more likely to develop cardiovascular disease, and this can adversely affect nutrition habits and sleep patterns (Thompson 2019). What this tells us is that physical activity and exercise can directly improve mental health symptoms. This means you can improve sleep patterns; improve neuroendocrine regulation and function, an area that deals with emotions and behavioral outcomes; and, most important, improve your overall quality of life. Exercise facilitates the reduction of unwanted stress, while further facilitating the reaping of positive physical benefits that come with it, such as, but not limited to, decreased resting heart rate, increased aerobic capacity, increase muscle mass, and improved bone mineral density.

Some important questions that tend not to be directly answered: How much exercise should people do? How long should they exercise for? How many days a week would they need to work out to improve physical and mental health outcomes? The American College of Sports Medicine recommends three to five days of ≥150 minutes of moderate exercise or ≥75 minutes of vigorous intensity (Thompson 2019). Now, this doesn't mean you have to work out continuously for 30–60 minutes each training session, and it also doesn't mean to focus on one form of training such as only aerobic-based training (walking, running, cycling, etc.). Aerobic training, short bouts of HITT training (anaerobic: ≤15 minutes), resistance training, flexibility training, and balance training all fall into physical activity guidelines, along with other recreational activities such as, but not limited to, hiking, golfing, walking, and park days with the family.

It is a misconception that exercise is different from physical activity, such as walking. Walking is exercise. Exercise focuses on four areas that can

overlap with a lot of recreational activities. The four areas of focus in physical fitness are resistance training, endurance training, flexibility training, and balance training—all of which can be done in an informal environment outside of a gym.

The best approach to physical fitness or physical activities is to cross-train and spend time in each category to keep the body enriched with proprioceptive feedback. All four categories work in unison to improve physical and mental health. Improving overall physical health can be accomplished through a consistent weekly exercise regimen. Many biological mechanisms benefit from acute and consistent physical activity and/or exercise. This doesn't mean someone has to work out every day or spend the same amount of time exercising at each workout. Life and responsibilities will impact how much time and energy you can put forth toward a workout. On days that are challenging your mental health, it is OK to cut back on your typical routine, but don't stop. Do a walk instead. Channeling times of hardship and despair through exercise is an opportunity to build up physical strength and exercise capacity to help equip you for when the times get tough. It doesn't mean you have to push the limits each time.

Mental health isn't an easy journey, and there is no approach that will solve every outcome. Exercise, however, is a tool that everyone can use to help reduce symptoms, improve overall quality of life, and increase physical strength and endurance. One insight that I would like to leave everyone with is, the approach to physical activity and exercise is specific to the person, and it will look different from person to person. Try not to put pressure on yourself to exercise like everyone else. Pick a physical activity that you enjoy to do and start there. Enjoy the time you have exercising, make it fun, and change it up every so often. Don't get so strict on the rules that you lose sight of the great benefits that exercise has to offer to your life.

"If you move better, you will feel better, and if you feel better, you can accomplish anything."™

—Taylor Lewis MA, CSCS, CMT, PRT

References

Cystic Fibrosis Foundation. (n.d.). Depression, Anxiety, and Cystic Fibrosis: What the Guidelines Mean for You. https://www.cff.org/Living-with-CF/Emotional-Wellness/Depression-Anxiety-and-Cystic-Fibrosis-What-the-Guidelines-Mean-for-You.pdf.

Thompson, WR. (2019). *ACSM'S Clinical Exercise Physiology*. Wolters Kluwer.

Warburton, DE, CW Nicol, and SSD Bredin. (2006). Health benefits of physical activity: The evidence. *CMAJ: Canadian Medical Association Journal*, 174(6), 801–809.

Congratulations

To our own Colleen Dunn, MS, RRT, CCRC, and Mary Helmers RN. In recognition of outstanding contributions made to the CF community, Colleen was named as Cystic Fibrosis Research, Inc. (CFRI), Professional of the Year, and Mary received the CFRI Champion Award. Congratulations again to you both. These are very well-deserved awards.

Colleen Dunn, MS, RRT, CCRC

Mary Helmers RN

Cystic Fibrosis Parent Advisory Council

http://med.stanford.edu/cfcenter/advisory-councils/pediatric-advisory-board.html

—Kirsten McGowan

Please find some activities listed below to help keep you and your children entertained (and safe) during the COVID-19 pandemic:

Fun activities

- Art for Kids Hub (https://www.youtube.com/user/ArtforKidsHub).
- Go on an indoor scavenger hunt with clues.
- Decorate a cardboard box (race car, spaceship, castle).
- Repurpose cardboard for toys (ramps, forts, stuffed animal hideouts).
- Build a tent or a fort with sheets and chairs and read or have a sleepover in it.
- Backyard camping (pitch a tent and sleep outside, make s'mores).
- Plant some flowers or a garden.
- Learn paper folding (origami, paper airplanes, newspaper hats).
- Build a fairy garden.
- Sidewalk chalk.
- Write letters to friends or family they cannot visit in person.
- Family game or puzzle night.
- Build a tower out of playing cards—see who can build the tallest.
- Learn new jokes and tell them to the family.
- FaceTime or Zoom with friends and family.
- Family movie night.

Learn new chores (age-appropriately, organized youngest to oldest)

- Picking up or organizing toys.
- Dusting.
- Vacuuming.
- Mopping the floor.
- Loading or unloading the dishwasher or washing dishes.
- Walking the dog.
- Cleaning bathrooms.
- Yardwork (mowing the lawn, raking, watering plants).
- Wash the car.

Cooking

- Bake dessert or a treat (cookies, brownies, cupcakes, banana bread, muffins).
- Younger kids can assist making meals.
- Older kids can be responsible for preparing one meal per week for the family.

Fitness activity

- Gonoodle.com (free activity/fitness/dancing videos for kids; requires a login/password set-up).
- Do a family workout together (https://www.youtube.com/watch?v=5if4cjO5nxo).
- Have a stair running race.
- Dribble a basketball up and down the sidewalk (or around the block).
- Ride bicycles or scooters.
- Set up an obstacle course around your house and have an American Ninja Warrior competition.
- Play catch.
- Find an empty tennis or basketball court and bring your own racket, ball, or bat (have kids make up their own game—doesn't have to be actual tennis).

CF Medication Practice (use the extra time at home to educate your CF child about his or her medications and responsibilities)

- Get his or her own meds ready for each meal and snack.
- Set up treatment (plug in equipment, put on vest, put in hoses, etc.).
- Assemble his or her nebulizer and add meds.
- Sterilize his or her nebulizer sets.
- Practice good hand hygiene.

Pediatric CF Center Update

—Mary Helmers, RN

COVID-19 update

We hope 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, or if you feel you need some additional support during these uncertain times. Feel free to call the CF RN phone line at (650) 736-1359.

We started seeing patients back in clinic on May 4, 2020, and visits have gone well. Your CF team members—MD/Advanced Practice Provider, RN, RT, social worker, dietitian, and/or pharmacist—are now all accessible in person during your clinic visit. We continue to enforce the rule that only one family member may accompany his or her child to the visit. No siblings or additional family members are allowed in clinic at this time. Upon request, some providers are offering telehealth visits.

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

We encourage you to refer to the Centers for Disease Control and Prevention (CDC) website, CDC.gov, for the most up-to-date information, and the Cystic Fibrosis Foundation website, CFF.org.

Transition Quality Improvement (QI) project

Over the past two years, the Pediatric and Adult CF teams have collaborated to review and redefine our transition process from pediatric to adult CF program. We have reinstated Quarterly Transition meetings with the two teams and started virtual visits where some of the Adult team members join in during your quarterly Pediatric visits in your last year at the Pediatric Clinic. At the final Pediatric visit, you will meet some of the Adult team members in person, and some of your Pediatric team will join you at your first Adult CF Clinic appointment.

Starting at age 8, Transition Booklets are given to patients and families. We ask that you keep the booklet in your CF binder and bring it with you to all your visits!

Check out the CF Center website: http://med.stanford.edu/cfcenter.html

Helpful tips

You can get assistance with your PG&E bill: PG&E forms for medical equipment and devices can be found on the PG&E website under "Medical Baseline Allowance Application for Medical Baseline Enrollment and Recertification." All you need to do is print the form, 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. Your provider will sign the form, and then you can mail it to PG&E.

CF Clinic Prep form (patient update): This form was designed to help you get all your questions answered. This is not mandatory, but 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 a question"? You can now fill out this form ahead of time and bring it to your clinic appointment.

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. Due to COVID-19, we have minimized our annual studies to labs and CXR; however, we have restarted checking all annual studies. If you did not get all your testing done in 2020, we will catch up in 2021.

New Pediatric Providers

Belinda Pagarigan, CPhT

Belinda recently joined our CF team at Stanford Children's Health as the CF pharmacy technician. Originally from Michigan, Belinda has been a pharmacy technician for 10 years and has been with Packard Children's since 2014, with experience in many areas within the pharmacy department. Sponsored through a CF Foundation grant, Belinda dedicates about one day per week directly to working with the CF team. You may have the opportunity to meet Belinda during clinic or over the telephone during an outreach call. She is passionate about pharmacy and truly cares about expanding access and reducing costs to our CF patients and families. When not in the pharmacy, Belinda loves to travel, swim, hike, sing karaoke, and explore new food venues.

Aditi Gupta

Aditi majored in cell and molecular biology at San Francisco State University and is currently working as a life science technician supporting Dr. Elizabeth Burgener's lab. Her research focuses on the development of novel diagnostic tools that permit the early detection of lung disease manifestations.

New Adult Providers

Theresa Kinney, RN (CF Nurse Coordinator)

Theresa is from Phoenix, Arizona. She received her nursing degree in 2010. Her professional experience includes inpatient rehabilitation, as well as teaching nursing assistant students in the clinical and didactic setting. Most recently, she worked as a traveling nurse, helping hospitals around the country in the emergency, trauma, orthopedics/spine, neurology, oncology, and heart and kidney departments. In her spare time, she enjoys cooking, traveling, and many outdoor activities. Theresa is very excited to be a part of the cystic fibrosis family at Stanford.

Sheldon (Shelli) Porter, RCP, RRT, BSRT

Shelli was born in Denver, Colorado, and now lives in Livermore, California. She received her respiratory therapy degree in 1991 and went on to get her bachelor of science degree in respiratory therapy in 2015. Her professional experience includes critical care, pulmonary rehabilitation, home care, and teaching respiratory students in the clinical and didactic setting, as well as critical care patient transport. Most recently, she worked in inpatient care for Stanford Health Care. This included work in the intensive care units, on general medical floors, and on the rapid response teams. In her spare time, she enjoys traveling, outdoor activities, and shopping for antiques. Shelli loves working with cystic fibrosis patients and is happy to be joining our team.

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 (Zirbes), DNP, RN, CPNP
Clinic Scheduling ..... (650) 724-4788
Clinic and Prescription Refill Fax ..... (650) 497-8791
Office Assistant/Patient Service Coordinator: Laura Banuelos ..... (650) 498-2655
Nurse Coordinator: Mary Helmers ..... (650) 736-1359
CF Clinic Nurse: Liz Beken ..... (650) 736-1359
Respiratory Therapist: Jessica King ..... (650) 724-0206
Nutritionist, Dietitian: Julie Matel ..... (650) 736-2128
Social Work: Teresa Priestley ..... (650) 736-1905
Newborn Screening: Jacquelyn Spano (Zirbes) ..... (650) 721-1132
PharmD: 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) ..... (510) 587-9631
Nutritionist, Dietitian: Ayah El-Beshbeeshy (Tues & Thurs) ..... (510) 457-4232
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
Adult Clinic Scheduling ..... (650) 498-6840
Adult CF Center Fax ..... (650) 723-3106
Nurse Coordinators: Theresa Kinney, RN and Kristel Fallon, RN ..... (650) 498-6840
Respiratory Therapy: Gauri Pendharkar, RCP and Sheldon Porter, CRT, RRT, BSRT ..... (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) 725-7061
- 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
Provider: Carolyn C. Hruschka, ANP-BC
Adult Clinic Scheduling ..... (415) 923-3421
Adult CF Center Fax ..... (415) 243-8666
Program 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.

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

STANFORD ADULT
Cystic Fibrosis
Advisory Council

As many of you know, sometimes it is necessary for individuals with cystic fibrosis to spend time in the hospital for what is commonly referred to as "a tune-up." The length of stay can be anywhere from a couple of days to several weeks. I know everyone is different, but here is a list of things you might consider packing in your hospital go-bag:

- If you are admitted during a holiday season, maybe bring some small decorations and/or a favorite blanket and pillow, as these might help make your room feel more like home.
- Electronic devices such as your laptop, cell phone, or tablet. Make sure you have your earphones and chargers for these items.
- Books or even e-books help pass the time.
- Bring your favorite pajamas or clothing. Pack footwear like slippers, flip-flops, or sneakers, and bring your own favorite socks.
- Family photos help as well. During the pandemic, these photos can bring a little peace and comfort to us.
- Snacks and treats are helpful. Sometimes you just need a little pick-me-up.
- Toiletries and skin care products are important to feel fresh.
- Earplugs might be necessary, depending where you stay. (The new hospital is quiet.)
- Bring things that help you deal with your stress. I bring my ukulele. Essential oils can help reduce the stress as well as help the room smell better. I have seen some people with small keyboards, coloring books, knitting materials, and sketching pads and pencils.
- Make sure you have your specialty medications. There are things that the hospital might not have that you regularly take.

Newsletter Contact Information

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

Editor: Zoe A. Davies, RN, MS, PNP, CCRC **Assistant editor:** Colleen Dunn, MS, RRT, CCRC