

# Cystic Fibrosis Center News

## Bone Health in Cystic Fibrosis

– Joy Wu, MD, PhD

As a testament to the outstanding cystic fibrosis (CF) care provided by centers such as Stanford Health Care and Lucile Packard Children's Hospital Stanford, people with CF are living longer and healthier lives. One consequence, however, is that people with CF increasingly encounter health issues associated with aging, potentially at earlier ages than those without CF.

### What is osteoporosis?

**Osteoporosis** – literally “porous bone” – is a disease where low bone mass leads to increased risk of fractures. In the United States, half of women and one-quarter of men age 50 and older will fracture a bone due to osteoporosis. Unlike fractures in children, which heal quickly while encased in colorful casts, fractures in older adults can be devastating. Among older adults who suffer a hip fracture, 1 in 4 will die within a year, and fewer than half

will ever walk independently again. Although osteoporosis is often considered a disease of older women, it also affects men, who account for up to 1 in 3 hip fractures.

### Why are people with CF at risk for osteoporosis?

Many factors can affect bone health, including genetics, diet, physical activity, medications and diseases. Bone mass typically increases during childhood and adolescence, reaching a peak around age 30. Because people with CF may experience nutritional deficiencies and impaired growth throughout childhood, their peak bone mass may not be as high. After age 30, bone mass gradually declines with age, and in women it can decline rapidly around the time of menopause. In people with CF, chronic infections can lead to increased inflammation, which can accelerate bone loss. People with CF may also be treated with high doses of medications that cause bone loss,

### *In this issue*

Bone Health in Cystic Fibrosis .....	1
Vitamins and Minerals for Optimal Bone Health in Cystic Fibrosis .....	3
Cystic Fibrosis Family Advisory Council: CF and Dental Health .....	5
Know Your Mutations: A CFTR Mutation Fact Sheet .....	6

Pediatric CF Center Update .....	8
Adult CF Center Update .....	9
A Research Experience .....	10
Farewell to Dr. Richard Moss .....	11
Current Research Studies .....	11

such as glucocorticoids. Lower levels of hormones such as estrogen and testosterone as well as limited physical activity can further reduce bone mass. People with CF who receive lung transplants need immune-suppressing medications that can lead to rapid bone loss in the first few months after transplant. Finally, some studies suggest that the CFTR protein, which does not function normally in CF, may have an important function in bone health.

Osteoporosis and fractures are very common in CF patients. Up to half of adult CF patients, even at an average age of only 30, have low bone density, and many have already suffered a fracture. Fractures can be especially troublesome for people with CF. For example, spinal fractures of the vertebrae are not only painful but can worsen lung function.

### Who should be tested for osteoporosis?

The amount of bone in the body is determined by the balance between bone breakdown and bone formation, and it can be estimated using a DXA (dual energy x-ray absorptiometry) scan, more commonly known as a bone density scan. A bone density scan assigns a T-score to a person's bone density. The World Health Organization defines osteoporosis as a T-score lower than -2.5, while less severe bone loss (osteopenia) is defined as a T-score of between -1 and -2.5.

Because low bone density is so common among people with CF, the European Cystic Fibrosis Society recommends that people with CF have their first bone density scan as early as between the ages of 8 and 10. In children and adolescents under the age of 20, bone density is measured at the spine and total body. In adults, bone density is measured at the spine and hip. If the patient's bone density is normal, the bone density scan does not need to be repeated for five years. If the patient's bone density is low, the scan should be repeated every one to two years. In addition, anyone who has experienced a fracture, for example at the spine or wrist, should undergo bone density testing every one to two years.

### What can be done to prevent osteoporosis?

Many things can be done to optimize bone health in people with CF. Good nutrition and maintaining a normal BMI are a priority, as is good control of lung infections. People with CF often require high doses of fat-soluble vitamins such as vitamin D and should have their blood levels checked

to adjust the doses as needed. People with CF should aim for a daily calcium intake of 1,000 to 1,200 milligrams, or the equivalent of three to four servings of dairy products, such as one cup of milk or one ounce of cheese. Information on the calcium content of common foods can be found here: <https://www.nof.org/patients/treatment/calciumvitamin-d/a-guide-to-calcium-rich-foods/>. Regular exercise, especially weight-bearing and muscle-strengthening exercise, can also improve bone health.

### What treatments are available for osteoporosis?

In addition to the lifestyle recommendations above — a healthy diet, adequate calcium and vitamin D, and regular exercise — other important approaches to improving bone health in people with CF include using the lowest possible doses of glucocorticoids (steroids) and diagnosing and treating low levels of testosterone in men with CF.

People with CF who have had fragility fractures, have been diagnosed with low bone density by DXA scan, are losing bone rapidly, are awaiting or have already undergone a transplant, or are on high doses of glucocorticoids may need specific medications to treat osteoporosis.

Medications that increase bone mass work by either preventing bone breakdown or promoting bone formation. The most commonly used medications for the treatment of osteoporosis are the bisphosphonates, which include alendronate (Fosamax), risedronate (Actonel) and ibandronate (Boniva). These medications are taken either weekly or monthly by mouth, and they are very effective at increasing bone strength and preventing fractures. They are very safe and generally well-tolerated, although the most common side effect is gastroesophageal reflux disease, with symptoms like heartburn, indigestion or nausea. If these symptoms occur, then a similar medication called zoledronic acid (Reclast) can be given as an intravenous infusion to avoid irritating the GI tract. These medications appear to be effective in improving bone density in people with CF.

Other medications are available for the treatment of osteoporosis, including denosumab (Prolia), which also prevents bone breakdown, and teriparatide (Forteo) and abaloparatide (Tymlos), which both promote bone formation. However, these medications have not been tested extensively in people with CF and should be used in consultation with an osteoporosis specialist.

## What is on the horizon for bone health in CF?

Several interesting laboratory studies have suggested that the CFTR protein (which does not function normally in CF patients) may have an important function in bone health. If so, newer medications for CF that improve the function of CFTR may be beneficial to the bones, as well. A small study showed that patients treated with ivacaftor (Kalydeco) had improvements in bone density.

Of course, these patients also experienced improved lung function and weight gain, which themselves can lead to improved bone density. More work lies ahead, but your CF team at Stanford Health Care and Lucile Packard Children's Hospital Stanford are committed to your overall health, including your bones!

# Vitamins and Minerals for Optimal Bone Health in Cystic Fibrosis

– Julie Matel, RD

People with CF are living longer than ever! This makes maintaining optimal bone health very important. Certain medications that are sometimes used in CF (such as steroid therapies), low body weight, delayed puberty, and malabsorption of fat-soluble vitamins (including vitamins D and K) can all contribute to inadequate bone mineralization. Individuals with CF can take important steps to promote strong bones, including maintaining a healthy weight and participating in weight-bearing and strength-training exercises such as walking, jogging, and weightlifting. Another important way to maintain strong bones is to consume a well-balanced diet that contains the optimal amounts of bone-making vitamins and minerals, specifically vitamins D and K, calcium, and magnesium. These important diet and exercise modifications can help people with CF protect their bones. The following is a closer look at bone-building vitamins and minerals.

**Vitamin D** is known as the sunshine vitamin because the body's process for making active vitamin D begins in the skin after it has been exposed to UV rays. Being in the sun for five to ten minutes, two to three times a week, can raise your vitamin D levels. To avoid sunburn, it is important not to stay out in the sun too long. Another important

source of vitamin D is from certain foods, such as vitamin D-fortified milk, eggs and fatty fish. People with CF who are pancreatic insufficient have difficulty absorbing fat and fat-soluble vitamins, including vitamin D. Getting enough vitamin D in the diet alone is difficult for people with CF. In order to meet higher vitamin D requirements, they should supplement their diet with vitamins that are made for them, like MVW Complete Formulation, DEKAs, or AquADEK. These vitamin preparations contain water-soluble forms of vitamin D, which are easier to absorb.

**Vitamin K** is another fat-soluble vitamin that's important for both blood clotting and bone mineralization. In addition to vitamin D, vitamin K is a component of CF-specific vitamin supplements, which should be taken daily by individuals with CF who are pancreatic insufficient and require enzymes. Vitamin K is found in dark green leafy vegetables, such as spinach, broccoli, turnip greens and Swiss chard. The healthy bacteria in the intestines also produce vitamin K. However, antibiotics can destroy these bacteria, so it is important to consume both foods and vitamin supplements that are high in vitamin K.

**Calcium** is a mineral that's well known for its importance in bone health. Children and teens require the most

calcium because their bones are growing rapidly. Some medications, such as certain antibiotics, cannot be taken at the same time as calcium supplements or calcium-containing foods. If your CF team recommends a calcium supplement, your pharmacist and CF team can provide guidance as to the best time to take your calcium supplement. Milk and milk-based products are excellent sources of calcium. Despite what you may have heard in anecdotes, drinking milk does not produce more mucus! Green leafy vegetables are good sources of calcium, as well. However, these sources tend to contain compounds that prevent optimal calcium absorption. Since calcium levels in the blood are maintained in a very narrow range, the only way to determine whether you need more calcium is to measure bone mineral density using a DEXA study.

**Magnesium** is another mineral that's involved in bone health because low levels of magnesium block the body's ability to use calcium. Magnesium is naturally present in many foods, such as nuts, seeds, legumes, fruits and vegetables, and it is added to other food products, such as bread and cereal. There are no specific recommended magnesium intake amounts for individuals with CF.

Remember, there are steps you can take to maintain your healthy bones! These are some foods to include in your diet to get the bone-building vitamins and minerals your bones need every day.

### Recommended Vitamin D intake

- 11 years and older, take 800 – 2000 IU
- 1 – 10 years, take 800 – 1000 IU
- 12 months and younger, take 400 – 500 IU
- Make sure the form of vitamin D is cholecalciferol or D3
- Make sure to take enzymes with vitamin D supplements
- Your team may recommend increased amounts of vitamin D supplementation depending on your blood results

### Recommended Vitamin K intake

- 8 years and older, take 300 – 500 mcg
- 4 – 8 years, take 300 – 500 mcg
- 1 – 3 years, take 300 – 500 mcg
- 0 –12 months, take 300 – 500 mcg
- It has been shown that many individuals with CF need more than 500 mcg per day to maintain adequate vitamin K status. Consult with your care team for specific amounts of supplemental vitamin K that may be right for you.

### Recommended Calcium Intake

- 9 years and older, take 1300 mg per day
- 4-8 years, take 1000 mg per day
- 1-3 years, take 700 mg per day
- 0-12 months, take 200-260 mg per day

### Bone-Building Foods

- Nuts
- Sunflower seeds
- Legumes
- Canned salmon with bones
- Sardines
- Tuna
- Milk
- Eggs
- Calcium-fortified orange juice
- Smoothies
- Yogurt
- Spinach salad with nuts and mandarin oranges
- Calcium-fortified soy or almond milk
- Calcium-fortified breakfast cereals



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# Cystic Fibrosis Family Advisory Council: CF and Dental Health

– *Kirsten McGowan, parent*

The CF Family Advisory Council seeks to address the needs of all families seen at Stanford Children’s Health’s CF clinics. We recently had a pediatric dentist, Dr. Donald Chi, visit the FAC from the University of Washington and present his research on oral hygiene in kids with CF. What did we learn? Taking care of your teeth and gums can impact CF health! We wanted to share some recommendations based on his research.

## Regular dental visits (checkups/cleanings)

- Always tell the dentist that your child has cystic fibrosis.
- Your child should have a dental checkup and cleaning 1–2 times per year (every 6–12 months).
- Consider doing airway clearance before dental cleanings (especially if your child is coughing).
- Floss and brush 2 times per day (morning and night) for 2 minutes at a time.
  - Get a fun timer.
  - Use chewable tablets to highlight areas to brush, such as Butler G-U-M Red-cote dental tablets.
  - Use a fun song to encourage good habits, such as “Healthy Teeth, Healthy Me: Brushy Brush” from Sesame Street.
  - Help a favorite toy “brush” their teeth.
- Limit your juice and soda intake.
- Always rinse your mouth with water after taking oral steroids.
- No pacifier, bottle or thumb-sucking after turning 1 year old.

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

## Dental treatments or procedures

- Always do airway clearance before any dental procedure (for example, tooth extraction, root canal, etc.).
- Make sure you know what to do if your child needs to take antibiotics. Notify your dentist of interactions or concerns with CF medications or therapies.
- Communicate with CF team if extra dental care is needed.
  - May need a note from the CF team for any dental procedure beyond a regular cleaning (such as root canals, oral surgery, etc.) that will involve anesthesia (such as nitrous oxide or general anesthesia).
- Discuss with the CF team any instance where anesthesia is needed.
- Be aware of transitioning to the mouthpiece and its impacts on orthodontia.



# Know Your Mutations: A CFTR Mutation Fact Sheet

– *Cystic Fibrosis Foundation*

Cystic fibrosis is caused by mutations, or changes, in the CFTR gene. This gene provides the code that tells the body how to make the cystic fibrosis transmembrane conductance regulator (CFTR) protein. The protein controls the salt and water balance in the lungs and other tissues. All people have two copies of the CFTR gene, and there must be mutations in both copies to cause CF. More than 1,700 mutations of the CFTR gene have been identified. Although some are common, others are rare and found in only a few people.

CFTR mutations are grouped into classes based on the way the mutations affect the CFTR protein. The reverse side of this sheet shows the most common CFTR mutation classes. In the future, mutations may also be classified by “theratype,” meaning which type of CFTR modulator therapy they respond to best. This is because mutations within the same class may

respond to therapies differently, and not every mutation can be neatly assigned to one mutation class.

Certain types of CFTR mutations are associated with different disease complications. For example, some mutations are more likely to affect the pancreas than others. However, this correlation is not perfect, and knowing an individual’s CFTR mutations cannot always tell you how severe that person’s CF symptoms will be.

Although the potential therapies described on this sheet can be very effective for some people with CF, others may not experience the exact same benefit. Researchers continue to work in the lab and in clinical trials to find the best therapeutic approaches to target specific CFTR mutations or classes of mutations to improve the health of all individuals living with CF.

## How is CFTR made?

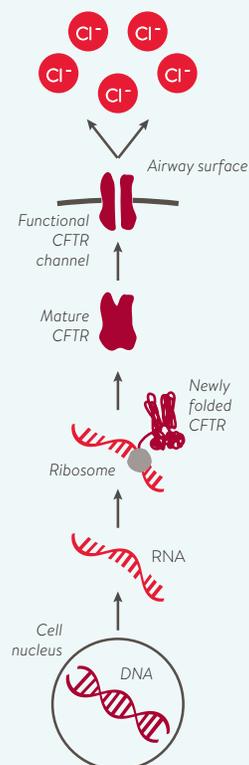
Once at the cell surface, the CFTR protein functions as a chloride channel. This channel helps maintain the right balance of fluid in the airways.

Once complete, the CFTR protein moves through the cell to the cell surface. This process is called **trafficking**.

Ribosomes are tiny molecular machines that read the instructions in the RNA and use them to make the CFTR protein. This process is called **translation**.

RNA acts as a template to make proteins. RNA is created by matching the coded instructions in the DNA. This process is called **transcription**.

DNA in the cell nucleus provides instructions to make proteins. The CFTR gene contains instructions to make the cystic fibrosis transmembrane conductance regulator (CFTR) protein.



## Potential therapies for CFTR mutations

Potentiators are drugs that help open the CFTR channel at the cell surface and increase chloride transport.

Correctors are drugs that help the defective CFTR protein fold properly so that it can move to the cell surface.

Read-through compounds aim to allow full-length CFTR protein to be made, even when the RNA contains a mutation telling the ribosome to stop.

RNA therapies aim to either fix the incorrect instructions in defective RNA, or provide normal RNA directly to the cell.

Gene-editing techniques aim to repair the underlying genetic defect in the CF gene DNA. Gene replacement techniques aim to provide a correct copy of the CFTR gene.

# CFTR Mutation Classes

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DESCRIPTION	Normal	Class I	Class II	Class III	Class IV	Class V
MUTATION EXAMPLES	No mutation	<p><b>22%</b></p> <p>G542X W1282X R553X</p> <p>aka "production mutations," which include nonsense mutations, some splice mutations and deletions</p>	<p><b>88%</b></p> <p>F508del N1303K I507del</p> <p>aka "processing mutations"</p>	<p><b>6%</b></p> <p>G551D S549N</p> <p>aka "gating mutations"</p>	<p><b>6%</b></p> <p>D1152H R347P R117H</p> <p>aka "conduction mutations"</p>	<p><b>5%</b></p> <p>3849+10kbCG→T 2789+5Cg→A A455E</p> <p>includes some splice mutations</p>
WHAT'S HAPPENING IN THE CELL						
POTENTIAL THERAPIES		Read-through compounds may allow production of full-length CFTR for nonsense mutations	Correctors such as lumacaftor or tezacaftor help defective CFTR fold correctly	Potentiators such as ivacaftor help open the CFTR channel, and also help increase the function of normal CFTR		

# Pediatric CF Center Update

– Mary Helmers, RN

Please check out our Facebook page, Cystic Fibrosis Center at Stanford, and CF Center website, [cfcenter.stanford.edu](http://cfcenter.stanford.edu).

## Two podcasts were added to the FB page

- **Traveling with CF**, by Mary Helmers, RN, Pediatric CF Nurse Coordinator
- **Gastrostomy Tubes in CF**, by Julie Matel, CRD, our CF Dietician

## The following information was added to our CF Center website

- **CF and Dental Health** fact sheet, which gives guidelines and tips on how to teach good oral hygiene and encourage your child to practice it
- **Link to the PG&E website** for Medical Baseline Allowance application for medical baseline enrollment and recertification
- **CF Clinic Prep form (patient update)**, which is designed to help you get all your questions answered. This is not mandatory, but you can use it as a tool to assist you in jogging your memory in preparation for your clinic visit. If you usually drive away from the 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.

Keep an eye out for more new topics and informational tips in the next few months on our FB page and CF Center website.

## 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 through MyChart, since it is not a secure site. Please note that any email sent to the team will be responded to with a phone call.

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

It takes only a minute to sign up. A member of our front desk staff will be happy to assist you with the sign up.

## CF Passport

Make sure you bring your **CF PASSPORT** with you! Use the PASSPORT in the hospital whenever you have an appointment, test or procedure.

**Parents/patients:** Please remember to carry your child’s **CF PASSPORT** in your wallet. If, for some reason, you do not have one or tossed it, please ask for one when you come to your next clinic appointment. We now have them in English and Spanish.

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

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

## 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

**Helpful hints for requesting refills:**

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

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

**Annual labs**

Remember our goal is to get all annual testing done on or around your child's birthday. Included in the annuals are your child's lab work, CXR, bone density scan (starting at age 12), full PFTs (starting at age 7), baseline audiogram (starting at age 6), liver screening and sputum cultures. Please let us know if any of these tests weren't done with your child's annuals.

**Wear your mask**

We are asking all CF patients to wear the turquoise-colored masks that are being handed out at the front desk. These masks have smaller filters that provide more protection when walking outside during all the construction that's happening at the hospital. We would like all patients to wear them to and from all clinic and hospital visits and when you walk outside the medical center. They should fit snugly around the nose and mouth. If you have not received the new mask, ask the front desk staff or anyone from the CF team for one.

**Genotypes needed**

With all the exciting research being done in CF, new drug advances for CF patients are genotype specific. We need to have copies of all our CF Center patients' genotype/ sweat chloride test results. If you have a copy or the original results, please bring it with you to your next clinic appointment. If your child had these tests done at an outside lab or another CF Center, please contact them and ask them to fax the results to (650) 497-8791, ATTN: Mary Helmers, RN, Pediatric CF Coordinator. We need these test results for all our patients! Our plan is to have patients re-genotyped if there is no documentation on file. Thank you for your help with this task.

# Adult CF Center Update: Tips for your Home Respiratory Supplies (Nebulizer Cups/Compressors)

– *Gauri Pendharkar, RCP*

1. Pari LC nebulizer cups can be used for up to six months IF they are cleaned and disinfected DAILY.
2. When you start using a new nebulizer cup, it is a good idea to put a reminder on your calendar or phone to order new cups when the old ones are close to being six months old.
3. If you have had a prescription for nebulizer cups filled before, call the pharmacy and see if there are refills available on the prescription. This saves time, and the pharmacy can simply fill the prescription for you if you have refills.
4. If there is no refill, send an email or MyHealth message to your RT coordinator to have a new prescription sent in for you.
5. To have a new prescription sent, it is very important to know the name of the pharmacy that accepts your insurance. This saves a LOT of time.
6. Most compressors come with a warranty of up to five years (depending on the brand). If your compressor is not working properly, call the pharmacy that sent it to you. Most of the time, if it is within the warranty period, they can replace it for you. If it is very old, you can send a message to your RT coordinator and a new one will be ordered for you.
7. Remember to change the filters on your compressor once a year. Most compressors come with a few new replacement filters.
8. In order to be regular with your treatments, do not wait until the last minute to get your supplies ordered. For example, if you are going to be travelling, plan ahead to see what you need and have it ordered well before your trip. Especially if you're travelling overseas, there may be other considerations, such as obtaining a travel compressor.

# A Research Experience

— McKay and Stacy Allen, parents

We are McKay and Stacy Allen. We have been married for 10 years and currently live in Salt Lake City, Utah. In 2011, we had our first handsome “big” boy, Jaxson, who weighed in at a healthy 10 pounds. Two years later, we added our second handsome boy, Evan, to our family. Evan was healthy at birth and grew normally. We were in newborn paradise! Five weeks later, our pediatrician called us into his office to let us know that Evan’s newborn screen had come back positive for cystic fibrosis. Our world, like many other CF parents, was turned upside down. Evan’s pulmonologist ended our first overwhelming appointment with the encouraging words that although CF is a tough diagnosis, there has never been a better time to have CF. Research and advancements are providing hope for a brighter and healthier future for those living with CF.

**HOPE.** Those four letters got us through that first tough year and every year after. After our initial CF clinic appointment, we sat down together and made a vow to each other that we will do all we can to provide Evan with the best care and medication possible, despite the sacrifice or cost. We spend our free time listening to CF podcasts, reading the latest research study literature, being involved in our local CF parent advisory board and participating in the Salt Lake chapter of the Cystic Fibrosis Foundation (CFF). We believe in the power of research and, most importantly, the hope it provides to those living with CF.

Below, we answer some common questions about participating in research.

## 1. How did you hear about the study that Evan is presently participating in?

The online trial finder on the CFF website. We had been following the older age groups trials closely since Evan was born, hoping he would be able to get into the trial when the age range dropped. We saw Stanford was chosen as a site for the trial and called as soon as his age range opened.

## 2. What were your concerns regarding enrolling your child in a study?

Obviously, you worry about side effects. But for us, the biggest worry was the travel. We travel from Salt Lake City to Stanford for the study, but it’s not been bad.



It’s been convenient. The flight is short, and the team is great.

## 3. What would you share with parents of young children with CF who are considering enrolling their child in a study?

It takes a lot of work, but it is worth it. For parents of a child with CF, just getting your child to take one more medication each day seems daunting, on top of the normal regimen of pills, inhaled drugs and therapies. But seeing Evan’s health improve and knowing that we’re on the cutting edge of finding a cure for cystic fibrosis is really important to us. When Evan was diagnosed, we sat down as a family and promised each other that we would do anything to help Evan and prolong his life. ANYTHING. So when we had an opportunity to fly to Stanford, it was just a small inconvenience. We’re committed to his health and finding a cure for CF. And, to top it off, all of his study visits have made him a pro for his normal clinical visits!

## 4. How would you describe your research experience?

It’s been great! Sure, the day-long appointments and tests aren’t fun, but Evan loves his team. The research team at Stanford is absolutely amazing and have become part of our family! We’re so hopeful and excited for the studies that are ongoing and those still in the pipeline.

## Current Research Studies

### Current studies:

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

**Vertex 661-113** – Phase 2 study for patients 6-11 years of age studying the Tezacaftor/Ivacaftor combination drug.

**Vertex 809-116** – Phase 3 roll-over study for children 2-5 years of age with F508 mutations.

**OPTIMIZE** – For treatment of newly acquired pseudomonas.

**Prospect** – Enrolling now, observational study for healthy normal people and CF patients.

**SHIP** – Enrolling now, use of hypertonic saline in children.

**Celtaxsys** – Anti-inflammatory study in adults.

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

**Vertex 770-126** – Phase 3, rollover study for children less than 2 years of age investigating the long-term effects of Ivacaftor.

**Utility of Lung Clearance Index: LCI** study to be done in clinic for pediatric patients.

**STOP 2:** A study designed to standardize the best length of time to treat a pulmonary exacerbation.

**RARE:** Rare CFTR mutation cell collection protocol.

### 5. Are there any other thoughts you'd like to share?

As parents of a child with CF, we are grateful for all the advancements taking place right now to find a cure for CF and improve the lives of those living with CF. We believe strongly in the importance of research. We are grateful for those that have participated in clinical trials to advance the medications for CF thus far, and we are excited to do our part to help the work along! It's a small sacrifice to help improve our son's life and increase our hope that someday we will see a cure for our little boy!

## Farewell to Dr. Richard Moss

– *David Cornfield, MD*



While much about the care of patients with cystic fibrosis has changed over the past three decades, Richard Moss, MD, has been a constant, always delivering kind, compassionate and wise care to patients and

families. Rick came to train at Stanford in the 1980s, and he became intellectually engaged by the biology of CF and emotionally, perhaps even spiritually, attracted to the palpable humanity of people living with CF and the caregivers who support them. Through countless conversations, exams and letters, Rick never failed to give every patient, family and trainee his very best. Each patient was a whole new world for him. Providers, patients and families from across the globe are elevated for knowing and working with Rick Moss. He has made us all better, and we will miss his wit, wisdom and grace. We wish him the very best in retirement.

## Cystic Fibrosis Center at Stanford

### Pediatric providers at Lucile Packard Children's Hospital Stanford

Pediatric CF Center Director: Carlos Milla, MD

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

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

### Urgent issues

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

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

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

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### Research

Colleen Dunn, Zoe Davies, Sean Ryan,

Wendy Valencia..... (650) 736-0388

### Adult providers at Stanford

Adult CF 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: Kelly Johnson, NP; Puja Sarna, NP; Glenna Monk, NP

Adult clinic scheduling ..... (650) 736-5400

Adult CF Center fax ..... (650) 723-3106

Nurse coordinators ..... (650) 498-6840

Patient last name A-K: Julian Liang, RN, BS

Patient last name L-Z: Ronni Wetmore, RN, MS

Respiratory therapy ..... (650) 736-8892

Gauri Pendharkar, RCP

Registered dietitian ..... (650) 529-5952

Michelle Stroebe, MS, RD

Social work

Meg Dvorak, LCSW ..... (650) 518-9976

Anastasia Kaiser, MSW..... (650) 444-6512

Mental health coordinator: Liza Sher, MD

### Urgent issues

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

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

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

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

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

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

Visit our website at [cfcenter.stanford.edu](http://cfcenter.stanford.edu) for more information about our center and CF.

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

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