

# Cystic Fibrosis Center News

## New Medication Update

– Deborah “DJ” Kaley, RN, MSN

### Welcome to the newest CFTR modulator therapy: tezacaftor/ivacaftor (Symdeko)

Move over Orkambi, there is a new kid on the block! Symdeko is a combination of the old, reliable drug ivacaftor (Kalydeco) and a new drug tezacaftor. It is indicated for patients who are 12 years old or older and have two copies of the delta F508 gene. It is also indicated for patients with one copy of delta F508 and one copy of any of the 26 other CFTR mutations (see Table 1). Like Orkambi and Kalydeco, Symdeko can alter liver function, so liver function tests should be performed before starting Symdeko, then once every three months during the first year, and then once yearly. Likewise, patients

should have a baseline test for cataracts before starting Symdeko and then once a year thereafter.

Use of Symdeko is not recommended for patients who are also on certain tuberculosis medications (rifampin, rifabutin), antiseizure medications (phenobarbital, phenytoin) or the herbal supplement St. John’s wort. Your Symdeko dosage may need to be adjusted if you are on antifungal medications (ketoconazole, itraconazole, voriconazole, posaconazole, fluconazole) or certain antibiotics (clarithromycin, erythromycin, telithromycin). These medications may reduce the effectiveness of Symdeko. So, why would you want to try Symdeko? In clinical trials, Symdeko

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had fewer side effects than Orkambi. The main side effect was chest tightness. Other adverse reactions included headache, nausea, sinus congestion and dizziness. There were also improvements in lung function (FEV1), and these improvements were maintained over time. Symdeko also reduced the rate of pulmonary exacerbations and improved body mass index (BMI) better than Orkambi.

But wait! My child is under 12 years old – what about us? Phase three studies in children 6 to 11 years old are currently taking place to test the safety and tolerability of Symdeko and determine how the drug is processed in the body. Meanwhile, studies of Orkambi in children ages 2 to 5 are also underway. The Orkambi study in young children is also looking at the safety and tolerability of the drug as well as how the body processes the drug. See the section on current and upcoming research studies to find out which studies are being conducted here at Stanford.

How can I get Symdeko or Orkambi? Talk to your CF care team and they can help determine whether they are appropriate for you/your child. Once it is determined that a patient is a good candidate for either of these drugs, the patient will need to have a baseline eye exam to assess for cataracts

as well as a baseline blood test to check liver function. After these tests have been completed, a prescription will be sent to your specialty pharmacy.

It is likely your insurance will require a prior authorization request, including proof of the patient’s eligible mutations, baseline lung function, eye exam and liver function.

Like Orkambi and Kalydeco, Symdeko should be taken with a meal that contains fat. Likewise, anyone taking Symdeko should avoid grapefruit and Seville oranges.

**Changes in asthma medications**

QVAR (beclomethasone) 40 and 80 are no longer available as a metered dose inhaler to be used with a spacer. QVAR 40 and 80 are now only available as a breath-actuated inhaler, which requires no spacer. However, breath-actuated inhalers are not recommended for anyone under the age of 4 because the patient must be able to take a deep, fast breath in and hold the breath for 10 seconds in order to use the inhaler correctly.

For patients younger than 4 and those who cannot master the breath-holding technique, you may find your doctor has changed the patient’s

**Table 1 – CFTR Mutations**

F508del/F508del (c.1521_1523delICTT)	E56K (c.166G>A)	R347H (c.1040G>A)
A1067t (c.3199G>A)	E831X (c.2491G>T)	R352Q (c.1055G>A)
A455E (c.1364C>A)	F1052V (c.3154T>G)	R74W (c.220C>T)
D110E (c.330C>A)	F1074L (c.3222T>A)	S945L (c.2384C>T)
D110H (c.328G>C)	K1060T (c.3179A>C)	S977F (c.2930C>T)
D1152H (c.3454G>C)	L206W (c.617T>G)	2789+5G->A (c.2657+5G>A)
D1270N (c.3808G>A)	P67L (c.617T>G)	3272-26A->G (c.3140-26A>G)
D579G (c.1736A>G)	R1070W (c.3208C>T)	3849+10kbC->T (c.3718-2477C>T)
E193K (c.577G<A)	R117C (c.349C>T)	711+3A->G (c.579+3A>G)

QVAR prescription to Flovent (fluticasone) 44 or Flovent 110. Flovent is an inhaled corticosteroid like QVAR, but it uses a different molecule. It should be tolerated the same as QVAR, with the same precautions. Inhaled corticosteroids are used to treat asthma-like components of cystic fibrosis (CF) such as wheezing and shortness of breath with activities not directly related to your CF.

For those of you on an Advair metered dose inhaler used with a spacer, which is a combination of salmeterol (a long-acting bronchodilator) and

fluticasone (an inhaled corticosteroid), there is now a generic version available. This new inhaler is called AirDuo. Advair is available in low (45/21), moderate (115/21) and high dose (230/21). The corresponding AirDuo dosing is 55/14, 113/14 and 232/14.

This combination drug is used for CF patients who have an asthma component that requires a short-acting bronchodilator (albuterol or Xopenex) more often. The long-acting bronchodilator reduces the need for additional albuterol or Xopenex not associated with your CF airway clearance therapy.

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## The Lung Clearance Index (LCI): A Novel Type of Lung Function Assessment

– *Jacqueline Zirbes, DNP, PNP, CCRC*

Cystic fibrosis (CF) lung disease is characterized by persistent infection and inflammation, which causes damage to the airways and leads to a progressive decline in lung function. Airway abnormalities are thought to start in the peripheral airways, but they also eventually lead to the destruction of the larger airways (bronchiectasis). Over the past 10 years, groundbreaking research has clearly demonstrated that lung disease starts very early in life for CF patients, possibly shortly after birth. Intensive clinical monitoring of the airway disease and early intervention are needed to delay lung disease progression. This has led to a major shift toward closer monitoring and more aggressive treatment of early CF lung disease. As a result, CF centers have searched for innovative means of assessing and monitoring lung function in young patients.

Spirometry (forced expiratory volume in one second, or FEV1) is still the most widely used method for clinical monitoring of CF

lung disease, and it is believed to be a good predictor of outcomes in patients with moderate to severe CF lung disease. However, two major disadvantages of the use of FEV1 in children with CF have become apparent.

First, FEV1 is not very sensitive to early CF lung disease. Many school-age children with CF may have FEV1 within the normal range or show a slow rate of progression, even though they probably have lung disease that progresses faster than is reflected in spirometric measurements. Second, reliable forced expiratory maneuvers are difficult to obtain in children younger than 5 years old, with testing in the infant and preschool age groups being largely confined to specialized laboratories. As interest in monitoring younger patients has increased, so has the need for more practical and sensitive alternative measures of CF lung disease that can be obtained in children of all ages. As many of our families observe, we

start teaching spirometry to young CF patients at approximately 4 years old as a method to measure their lung function. Typically, for the spirometry test to be reliable, it requires the child to closely follow directions and to be able to force air out of their lungs for several seconds. This skill is learned over time and often not mastered until between 6 and 7 years of age. As a consequence, for the longest time many investigators and care providers have been interested in finding a more practical way to accurately assess the lung function of children. We may now have finally arrived at an answer to this need.

The airway tree is designed to effectively distribute fresh inhaled air to the gas exchange compartments deep down in the peripheral airways of our lungs, where it mixes with the exiting gas present in the lungs. Gas mixing tests reflect peripheral airway function, and because they can be performed during regular relaxed breathing with passive cooperation, these tests may have a role in the CF clinic setting. The main idea is to detect overall inhomogeneity (unevenness) of air distribution in the lungs, often called poor gas mixing. The multiple-breath gas washout (MBW) method is used to measure the efficiency of ventilation distribution in the lungs. A measurement that can be obtained from this method is the lung clearance index (LCI), which represents the cumulative expired volume required to clear a gas from the lungs. We at Stanford, among many other investigators worldwide, have worked for the past 10 years on the refinement of the methodology. Thanks to this effort, it has now been demonstrated that the LCI is more sensitive than spirometry in detecting lung function abnormalities in children with CF.

Making use of the most current technology along with consensus standards for the methodology to which we contributed, the MBW test by nitrogen washout is routinely being performed in our pediatric CF clinic in Palo Alto. The test

starts by letting the child get comfortable while wearing either a face mask or a mouthpiece and breathing room air. When breathing is quiet and regular, 100 percent oxygen is administered through the breathing system. Breath by breath, the equipment measures the concentrations of oxygen and carbon dioxide to estimate the nitrogen left in the lungs until almost all the nitrogen has cleared. The resting lung volume at the end of exhalation (known as functional residual capacity, or FRC) is calculated along with the LCI.

Based on the accumulated evidence, including our own work, we believe the MBW technique provides a very sensitive measure of airway disease through the LCI. In addition, we have already shown that MBW testing can be performed easily and successfully in almost all children who are able to sit quietly while watching a movie. So far, the LCI has primarily been used in research and as an endpoint for clinical trials to demonstrate the efficacy of CF drugs like hypertonic saline, Pulmozyme, Kalydeco and Orkambi. Through these studies, the LCI has consistently proven to be extremely helpful for detecting the beneficial effects of these medications in young CF patients that otherwise would not have been detected by other methods. However, at this point we do not know how useful it could be in the clinical care of patients with CF. With support from the Cystic Fibrosis Foundation (CFF) we have recently embarked on a large study for CF children that is designed to evaluate the feasibility of incorporating LCI into a clinic visit and, more importantly, to investigate the value of the information it provides when used in conjunction with routine CF evaluations like spirometry. We hope to include most of the children that we currently follow at the CF center into this study. Our goal is to provide the CF community with valuable information as to the future prospects of this measurement as a tool for managing CF lung disease.

# Pediatric CF Center Update

– Mary Helmers, RN

Warm summer months are soon approaching. Since people with cystic fibrosis (CF) lose more salt through sweat than the average person, this puts CF patients at a higher risk for dehydration. Drinking water, juice, and sports drinks and consuming salty snacks such as pretzels, salted nuts, and seeds will help replace salt lost through sweat.

Dehydration can occur very quickly, especially in hot, humid weather. Don't wait until you are thirsty to take a drink – always keep up your fluid intake! Carry water or fluids with electrolytes (e.g., Gatorade) with you at all times.

Summer is a great time for exercise and outdoor fun. It is recommended that people with CF drink 6 to 12 ounces of fluid every 20 to 30 minutes during aerobic exercise. Sports drinks with added carbohydrates and salt are a great choice for people with CF. Encourage everyone to take fluid breaks during exercise.

Fluid intake is especially important for children, who are more susceptible to dehydration than adults. Infants with CF will dehydrate more quickly than young children, adolescents or adults with CF. Keep your baby well hydrated and dress your baby in cool clothing. Speak with your CF clinic team for information about supplements and nutrition for infants with CF during the summer months.

Sunscreen is an important addition to your daily routine all year long, but it is particularly imperative during the summer months. When your child is wearing sunscreen, UV rays are absorbed and the skin is protected against damage.

Some medications (e.g., antibiotics and antifungals) have photosensitivity side effects, so children who take these medications can burn more easily if they are not wearing sunscreen.

## Check out the CF Center website

We recently updated the center's website. Take a look and let us know what you think: [med.stanford.edu/cfcenter](http://med.stanford.edu/cfcenter).

The following information is on the CF Center website:

- **CF and dental health:** A fact sheet that gives guidelines and tips on how to teach and encourage your child to practice good oral hygiene.
- **Link to the PG&E website** for Medical Baseline Allowance Application for Medical Baseline Enrollment and Recertification.
- **CF clinic prep form (patient update):** This form was designed to help you get all your questions answered. This is not mandatory, but it is a tool to jog your memory in preparation for your clinic visit. If you typically 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.

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

**Coming soon:** We will be adding a link to the website so you can view the pamphlet describing all the annual studies.

## New podcasts

Please check out our Facebook page and CF Center website. The following three podcasts were added to both sites. If you're planning your summer vacation, make sure you check out the "Traveling with CF" podcast so you are ready for travel.

- **Traveling with CF**, by Mary Helmers, RN, Pediatric CF Nurse Coordinator
- **Gastrostomy Tubes in CF**, by Julie Matel, CRD, CF Dietician
- **Research Participation: The Basics and Beyond**, by Colleen Dunn, CCRC, RRT, Research Administrator

## 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 cystic fibrosis (CF) care team. The CF care team cannot respond to patient or parent emails through MyChart since it is not a secure site. Please note **that any email sent to the team through MyChart will be responded to with a phone call.** We do not always check emails on a daily basis. If you or your child has a clinical need or question, please call the CF RN line at (650) 736-1359. It takes only a minute to sign up for MyChart. One of our front desk staff members will be happy to assist you with the sign up.

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

## 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 reminders

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

**Prescriptions:** Just a reminder that your prescription request can take up to 72 hours to be processed. This has always been our policy, although we do 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 the scrip (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 available.
- If you have a refill, great! Then the pharmacy will process it.
- Your pharmacy should call us if you have no refills left.

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

### Annual labs

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, chest x-ray, bone density scan (for patients 12 years of age and older), full pulmonary function tests (starting at age 7), baseline audiogram (starting at age 6), liver screening and sputum cultures. Please let us know if your child has not had any of these tests done with his or her annuals.

### Wear your mask

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

### 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 result, 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.

**Oliva Davis, who participated in a cystic fibrosis research study at Stanford**

## A Research Experience

– *Oliva Davis*

### How did you hear about your particular study?

My pediatric CF doctor referred me to Zoe to talk to her about participating in Stanford clinical trials.

### What were your main concerns regarding enrolling in a study?

My main concerns were about the time commitment and possible side effects of the study drug.

### What would you share with others who are considering enrolling in a study?

I would tell people who are considering enrolling in a clinical trial that I think the overall risk is lessened by the fact that you can withdraw at any time if you or your health care team decide that you don't want to or shouldn't continue (regardless of the reason and with no penalty).

### How would you describe your research experience?

Participating in the trial was definitely time consuming, but Zoe and Colleen always made the appointments entertaining. I mostly enjoyed it because it makes me feel like I am contributing to the CF community and furthering our cause, which is VERY rewarding.



# Improving Quality of Life through Posture

– Taylor Lewis, MA, CMT, CSCS, PRT

Imagine doing an exercise 16,000 to 24,000 times a day. You would get into good shape very quickly, but over time your body would fatigue, break down and start to compensate. Eventually, this would cause your posture to change. Your body would turn into gelatin, and your joints would let you know how much they do not enjoy the high number of repetitions you're doing.

Over time, your neuromuscular system would fatigue and your posture would adapt. This reprogramming of your posture would ultimately lead to a change in your respiration without you being aware of it.

On average you take in anywhere from 12 to 16 breaths per minute. That number would increase if you exercise and push your threshold. However, that number would also increase if your posture wasn't in line with what you were doing at that very moment. The human system doesn't have a distinct picture of a natural standing, sitting or lying-down posture. It does, however, dissect, adapt, and learn what physiological and psychological parameters are required to sustain and produce these postures and positions.

As you wake up and roll out of bed, your body is using what it has learned to get you from point A to point B as efficiently as it can. In order for this to occur, your respiration has to fall in line. As you take in those 12 to 16 breaths per minute, your diaphragm, abdominals, pelvic floor muscles and ribcage have to synchronize to allow the lungs to receive optimal oxygen.

When your lungs are able to bring in optimal oxygen, they can transfer it to areas that need it, such as muscles, tendons, ligaments and bones.

Posture and breathing go hand in hand. Without oxygen, we cannot survive, and without quality movement, we will not be able to thrive. Improving breathing mechanics can unlock stiffness, improve movement, and down-regulate your stress levels, but most of all it will improve your quality of life. There is a correlation between posture and breathing respiration. Ask yourself what type of correlation you would like to have: positive or negative.

Here are a few bang-for-your-buck exercises that can help improve your breathing mechanics. Over time, they could also help improve your posture and quality of life. Taking a few minutes each week to focus on you and how your body moves will change your life. Remember, you deserve to move well and have a high quality of life, just like anyone else.

You matter. Make your world a better place through the breath of movement.

## 90/90 Breathing





- Lie on your back with your feet flat on a wall and your knees and hips bent at a 90-degree angle.
- Keep your lower back and the back of your head in contact with the ground.
- Tuck your ribcage down and place your palms up and below your shoulders.
- Take a deep breath in through your nose and exhale through your mouth as if you're blowing 42 candles out with a straw.
- Hold for 2 seconds and then repeat.

Recommendation		
<b>Sets</b> 3–4 sets	<b>Reps</b> 4–5 breaths per set	<b>Rest</b> 15–30 seconds between sets

### Side Plank



- Lie on your side with your elbow under your shoulder and your knees and feet stacked on top of each other.
- Drive your hips forward by squeezing your butt, and lift your hips off the ground.
- Take 5 deep breaths in through the nose and out through the mouth.
- Relax, and repeat on the other side.

### Tips

- Maintain a straight line from your head to your feet.
- Try to breathe in slowly and exhale slowly.

### Recommendation

Sets	Reps	Rest
2–4 sets	3–5 breaths per set	30–60 seconds between sets

### Deadbugs



- Lie on your back with your lower back and head in contact with the ground.
- Lift your feet off the ground and bring your knees to a 90-degree angle.
- Bring your arms to a vertical position.
- Extend your right leg out while you bring your left arm above your head.
- Hold for 2 seconds then bring the leg and arm back to the starting position.
- Repeat on the other side.

### Tips

- Keep your lower back in contact with the ground during the whole exercise.

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# Be in Charge: A Study Looking at an Online Tool for Improving Nutrition in Children with CF

– Julie Matel, RD

## **Optimal nutrition improves health outcomes in individuals with CF**

While we know that optimizing nutritional status and growth improves health outcomes and survival in children with cystic fibrosis (CF) and reaching a body mass index (BMI, a measure of weight compared with height) at or above 50 percent for their age is associated with better lung function (Stallings et al., 2008), actually achieving these goals can present a challenge for families living with CF.

Children with CF have much higher calorie needs and need to consume 20 to 50 percent more than peers their age. Unfortunately, studies of dietary intake show that most children with CF do not meet these higher calorie recommendations. (Kawchak et al., 1996)

focus on you and how your body moves will change your life. Remember, you deserve to move well and have a high quality of life, just like anyone else.

You matter. Make your world a better place through the breath of movement.

## **Mealtime frustrations for families living with CF**

Parents of children with CF have reported frustration during mealtimes, including an increase in behaviors that interfere with eating, such as crying, whining, delaying meals with talking, spitting out food and leaving the table. As a result, parents

often resort to countermeasures in an effort to get their kids to eat. These include coercive tactics (“You can’t leave the table until you finish your meal”), commands to get their child to eat (“Take one more bite”), physical prompts (picking up the fork and feeding the child) or delaying mealtimes. Unfortunately, studies show that these efforts do not result in improved calorie intake and in fact correlate with a lower weight percentile in children with CF (Stark et al., 2002).



## **Studies show improvements in nutrition following behavioral and nutrition education**

Studies looking at behavioral interventions have shown promising results. For example, a nine-week clinical trial of behavioral and nutrition education led to less frequent mealtime behavior problems, better calorie intake and improved weight in children with CF (Opipari-Arrigan et al.,

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# Colon Cancer Screening in Cystic Fibrosis

– Meghan Marmor, MD, and Laveena Chhatwani, MD

## What's the news?

In April 2015, an 18-member task force convened at the Cystic Fibrosis Foundation headquarters to develop colon cancer screening recommendations for adults with cystic fibrosis (CF). This led to a recent paper on CF colon cancer screening consensus recommendations (Hadjiliadis et al., 2018).

## Why does it matter?

Individuals with CF are at an increased risk of colon cancer compared to individuals of the same age who do not have CF. How much is the risk increased? It is estimated that in adults with CF, the risk of colon cancer is 5 to 10 times greater compared to the general population of the same age who do not have CF. In adults with CF who have received organ transplantation, this risk is 25 to 30 times greater compared to the general population.

## What do the guidelines recommend?

1. Talk with your CF care team to decide if screening is right for you.
2. If screening is right for you, the best recipe for success is to partner with a gastrointestinal (GI) doctor who understands the needs of individuals with CF.
3. The best, gold-standard screening method for colorectal cancer is a colonoscopy. While CT colonography, stool studies and sigmoidoscopy have been used for colon cancer screening, there is not enough evidence to know if this would work for persons with CF.

4. Start screening at age 40. If the colonoscopy is normal, repeat screening every five years. If the colonoscopy is abnormal, repeat at least every three years or possibly more often for concerning findings.
5. For CF individuals who have undergone organ transplantation, colonoscopy screening should start within two years of transplant for patients older than 30. If the colonoscopy is normal, repeat screening every five years. If the colonoscopy is abnormal, repeat at least every three years or possibly more often for concerning findings.

We have established a collaboration with Stanford Gastroenterology to identify GI specialists with interest and expertise in CF. If your conversation with your CF physician results in a decision to proceed with a colonoscopy, you will be referred to meet the GI specialist for a consultation about the procedure. Once all your questions have been addressed, you will be scheduled for a colonoscopy.

At your next clinic appointment, let's plan to talk more about colon cancer.

## References

Hadjiliadis, D., et al. (2018) Cystic fibrosis colorectal cancer screening consensus recommendations. *Gastroenterology*. 154(3): 736 –745.

Members of the Global Burden of Disease Cancer Collaboration. (2017) Global, regional, and national cancer incidence, mortality, years of

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# Cystic Fibrosis Parent Advisory Council: MyChart

– Kirsten McGowan, parent

MyChart is a secure way to manage your child's health care online from a computer, phone or tablet. There is an app for iPhone and Android, and MyChart is also available in Spanish. Technical support is available in English and Spanish 24 hours a day, 7 days a week, via phone, email or online chat.

## What can I do in MyChart?

- Ask non-urgent medical questions (Don't have time to call the CF nurse line? Message the CF team via MyChart!)
- Appointments (Forgot to schedule your next visit? Request appointments via MyChart! (You can also view and cancel upcoming appointments.)
- Request prescriptions (Out of refills? Send Rx requests via MyChart!)
- View your health records, including:
  - Test results (labs, sputum cultures, x-rays, OGTTs)
  - Medication lists
  - Growth charts
  - Immunizations
  - Visit summaries
- View and pay bills
- Request medical records
- Watch health videos

## Why should I use MyChart?

- It saves time!
  - Messaging questions/requests via MyChart means no waiting for a phone call.
  - Test results typically come faster through MyChart, and you don't have to wait for a phone call with your results.
  - Online bill pay means no more buying stamps or writing checks.
- It's accessible!
  - Anytime, anywhere – access to your child's care is at your fingertips.

If your child is 12–17 years old:

- Teens can sign up for their own MyChart accounts.
- The parent/guardian's account has less access to the teen's medical record to protect the patient's privacy. But don't worry, MyChart is still useful for parents/guardians of teens!
- Parents cannot access the following when their child is between 12 and 17 years old: psychology, FPACT (family planning, OB/gyn and dermatology).

## How do I sign up?

- Download and fill out a consent form on the MyChart website (click Create an Account) and send the completed form to Medical Records.

MyChart access by age

Feature	Parent/Guardian MyChart Account		
	Child (0-11)	Teen (12-17)	Adult (18+)
Secure messaging	Full access	Full access	Patient has their own MyChart account as a legal adult.
View medication list	Full access	Restricted	
Rx refills request	Full access	Via secure messaging	
Allergies/immunizations/growth charts	Full access	Full access	
Request appointments	Full access	Full access	
View upcoming appointments	Full access	Certain appointments only*	
Cancel appointments	Full access	Certain appointments only*	
Test results	Full access	Full access	
Visit summaries	Full access	Certain appointments only*	
Request medial records	Full access	Need signed consent from the patient for certain records*	
View and pay medical bills	Full access	Full access	

■ Full access     
 ■ Restricted     
 ■ Limited access

- Bring a photo ID to your next CF appointment and ask the front desk for MyChart.
  - They will verify you as the legal guardian and ask you to register with an email address.
  - An activation email (with a code) will be sent to the email address you provide, and you will have to click the link and activate the account.
- Activation codes do expire, but you can always ask for another code the next time you are at the clinic.

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

MyChart pre-login page



Pre-login box detail

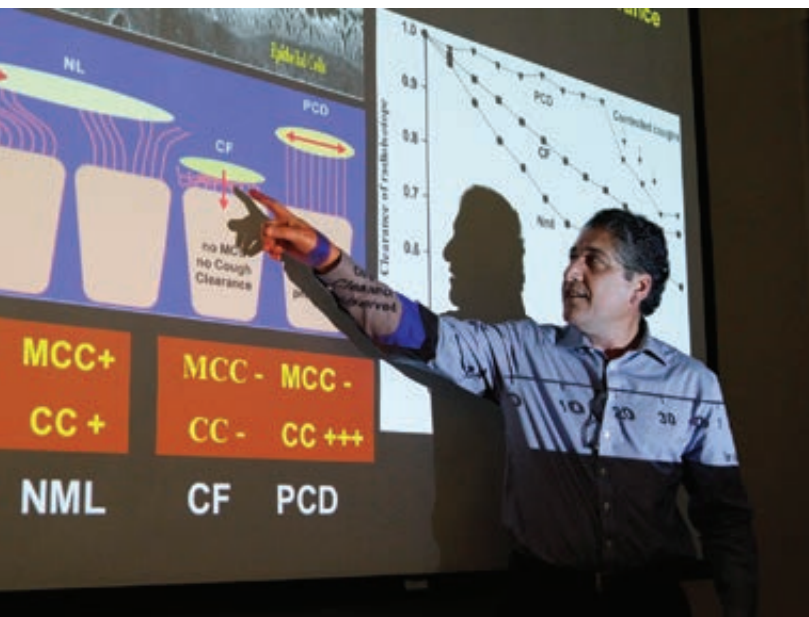
# Stanford CF Education Day: March 10, 2018

– Daniel Alvarez, RC, Sean Ryan, RRT, CCRC, and Wendy Valencia, RC

Our 18th annual CF Education Day started with a general morning session for both the pediatric and adult cystic fibrosis (CF) community at Stanford. Carlos Milla, MD, director of the Stanford pediatric CF clinic, moderated and was the keynote speaker for the morning session. His talk focused on the science of the CFTR protein and the research behind the development of new therapies, specifically Kalydeco, Orkambi and Symdeko. This was followed by a discussion of upcoming therapies such as gene editing.

of fungus in CF lung disease. He began with an overview of the common microbes (germs) found in the CF lung and then focused on fungi, including the prevalence of fungi, the risk factors of fungal colonization and potential treatments.

Next up to the podium were Kirsten McGowan and Colleen Dunn, who provided updates and reviewed past and present projects for the CF Family Advisory Council and the Adult CF Advisory Council. Our last speakers for the morning session were Nicole Hood from the Cystic Fibrosis Foundation (CFF) and Siri Vaeth from Cystic Fibrosis Research, Inc. (CFRI). They reviewed their specific missions and spoke about the resources that are available to families and patients. Following a delicious boxed lunch and great conversation with old and new friends, the afternoon sessions started. They were split into separate pediatric and adult sessions. The pediatric session was moderated by Michael Tracy, MD, and the topics focused on optimizing patient health and therapies. Mary Helmers, RN, started by describing the various tests our patients are required to complete on a yearly basis. More specifically, she reviewed what, why and when we do these annual tests. One key point she touched upon was how children with CF are at risk for developing a number of complications that can easily go unnoticed. Yearly chest x-rays, blood tests, liver ultrasounds, oral glucose tolerance tests, comprehensive pulmonary function tests, and DEXA scans allow us to detect these complications before any significant problems develop. They also help us monitor the progression of the above complications as the patient gets older.



## Carlos Milla, MD

Paul Mohabir, MD, director of the Stanford adult CF clinic, was the second speaker, and he gave an update on the current state of the Stanford CF Center. He went into detail on all of the changes and improvements the Stanford CF Center has made over the past year. The next speaker was Michael Tracy, MD, who talked about the role



**Julie Matel, RD**

The second speaker was our pediatric dietician, Julie Matel, RD, who stressed the importance of growth and weight gain and spoke about the behavioral barriers that may interfere with patients achieving their nutritional goals. She described how optimizing nutritional status and growth improves health outcomes and survival in children with CF and how maintaining a BMI at or above 50 percent for the patient's age group is associated with better overall lung function. One strategy Julie mentioned that can help parents during meals is using praise. She explained how you can use attention to reward your child for eating well and help them develop good eating habits. Some behaviors you should praise include coming to the dinner table right away, taking several bites in a row, chewing and swallowing food, and using good table manners.

The third speaker was Candice Middleton, RCP. Her presentation focused on how patients can get the most out of their daily treatments and explained how to properly maintain the equipment for the treatments. She noted patients can easily be distracted during treatments, so finding the right place to do them can help with the effectiveness

of each session. It is also important to do basic cleaning and maintenance of your home equipment every day to keep it in proper working order. After every use, clean your nebulizer cup with hot water and soap to remove any residue or leftover medicine, and then rinse the cup with sterile water. Disinfect your nebulizer cup daily by placing the nebulizer cup in boiling water for five minutes.

Our final speaker was DJ Kaley, RN, from our CF clinic in Emeryville. She discussed how patients can maximize their office visit by figuring out beforehand what they want out of their clinic visit. For instance, are there any pending lab results that need to be reviewed? Keeping a diary of your symptoms, whether it's increased coughing or difficulty breathing, can help the clinic team provide the best care possible. A diary can also be useful for recording any questions or concerns you may want to share at your next clinic visit. If the visit is for your child, after the visit, review with your child what went well and what did not go so well. Discuss what to work on for the next visit, and ask your child what can be improved. It is important to share these ideas with the clinic team as we are always



**Members of our CF care and research teams**

striving to provide the best care to your child and to make your child feel as comfortable as possible.

The adult session was moderated by Paul Mohabir, MD. The first speaker was Laveena Chhatwani, MD, the associate director of the CF Center. Her topic was colon cancer and cystic fibrosis. She discussed how important it is to screen for colon cancer early. This includes being up to date with your medical history, having your questions ready at the beginning of your clinic visit, making sure that you are aware of the early signs and symptoms of colon cancer, and most importantly, discussing your health with your health care provider.

The second speaker was Marina Basina, MD. She discussed advances in diabetes technology and its role in CF-related diabetes (CFRD) therapy. The technology has really advanced in recent years, which has made dealing with diabetes so much easier for patients and their families. The third speaker was Gauri Pendharkar, the respiratory therapist for the CF adult clinic. Gauri's topic was "Nebulizers and Vests: Which Is Best?" She gave great information on the proper order of inhaled medications, she described the most common bronchodilators that are used in CF and explained

how they work, she discussed airway clearance, and she reviewed the two different types of vests that are commonly used. She also gave information about nebulizers and compressors and how to properly care for and clean them.

Last but not least, she provided great tips about how yoga and exercise can help with airway clearance, as well. The fourth and final speaker was Denise Kwong, PharmD. She discussed pharmacology in CF. She explained how it's important to treat pulmonary exacerbations promptly, she reviewed common pathogens found in the adult CF lung, and she explained how specific antibiotics are selected. She reviewed how patient adherence is very important in maintaining overall health, which medications are commonly used, and which medications are specifically recommended for certain age groups. She also described the new modulator medications (Kalydeco and Orkambi), their risks and who should be taking them.

In closing, this year's CF Education Day was insightful and enjoyable. If you would like a copy of our 18th annual CF Education Day booklet, which includes our speakers' presentations, please call us at (650) 736-0388.





*Improving Quality of Life...*  
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- Breathe in through your nose as you extend out, and exhale as you come back to the starting position.

Recommendation		
<b>Sets</b> 3–4 sets	<b>Reps</b> 2–4 per side	<b>Rest</b> 15–30 seconds between sets

### Standing Cross Crawls



- Stand tall with your weight balanced throughout your feet.
- Drive one knee up to hip height while you bring the opposite hand to the top of the knee.
- Touch the knee and hold for 1 second.
- Repeat on the other side.

#### Tip

- To make it more dynamic, walk, touch and hold for 10 to 20 yards.

Recommendation		
<b>Sets</b> 3–4 sets	<b>Reps</b> 8–10 per side	<b>Rest</b> 20–45 seconds between sets

If you have any questions regarding exercising in general or about a specific exercise for a particular pulmonary disease, please contact us at [cffitnessinstitute@gmail.com](mailto:cffitnessinstitute@gmail.com). We would love to hear from you and see how we can help.

## Farewell to Adult Staff Members

Goodbye to Ronni Wetmore and Julian Yoshizawa, the adult clinic coordinators. We would like to thank both of them for all of their hard work over the past several years and they will be greatly missed. Replacement RN coordinators have been hired. Look for formal introductions in the next newsletter.

## Current Research Studies

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

**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

**Proteostasis 808-01:** Phase 1 modulator study for adults with F508 mutations

**Proteostasis 428-06:** Phase 2 amplifier study for adults with F508 mutations receiving Symdeko

*Be in Charge: A Study...  
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2010). More recently, a pilot study of a web-based behavioral nutrition intervention program (Be in Charge) showed that children of mothers who participated experienced a significant improvement in weight pre-to-post treatment (Stark et al., 2016).

**Be in Charge phase II study**

The Stanford Children’s Health Cystic Fibrosis Center will be participating in a phase II study looking at feasibility and outcomes of a web-based behavioral nutrition intervention program. We are hoping to start study enrollment sometime in the summer of 2018.



Talk to your CF team if you are interested in participating!

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*Colon Cancer Screening...  
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# Cystic Fibrosis Center at Stanford

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

Carlos Milla, MD, Pediatric CF Center Director;  
Sumit Bhargava, MD; Elizabeth Burgener, MD;  
My My Buu, MD; Carol Conrad, MD; David  
Cornfield, 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  
Nanci Martinez, office assistant/  
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  
Teresa Priestley, social work ..... (650) 736-1905  
Jacquelyn Zirbes,  
newborn screening coordinator ..... (650) 721-1132  
Russell Wise, pharmD ..... (650) 736-1905  
Diana Naranjo, PhD, Clinical Psychologist

### 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.

## Adult providers at Stanford

Paul Mohabir, MD, Adult Center Director  
Laveena Chhatwani, MD, Associate Center Director  
**Providers:** Gundeep Dhillon, MD; Jennifer Cannon,  
NP; Elika Rad, NP; Meredith Wiltse, NP  
**Backup providers:** Kelly Johnson, NP; Puja Sarna,  
NP; Julie Hoang, NP

Adult clinic scheduling ..... (650) 736-5400  
Adult CF Center fax ..... (650) 723-3106  
Nurse coordinators ..... (650) 498-6840  
Respiratory therapy ..... (650) 736-8892  
Gauri Pendharkar, RCP; Fernanda Shukla, 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.

### Research

Colleen Dunn, Zoe Davies,  
Sean Ryan, Wendy Valencia (650) 736-0388  
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for more information about our center and CF.

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or by email at [cathyh1@stanford.edu](mailto:cathyh1@stanford.edu).

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

## New Pediatric Staff Members



Russell Wise is from Birmingham, Alabama. He graduated with a bachelor's degree in biomedical sciences from Auburn University and received a Doctor of Pharmacy from Mercer

University in 2014. Prior to moving to the Bay Area, Russell spent a little over three years as a pharmacist at Texas Children's Hospital in Houston. As the first pharmacist to join the Lucile Packard Children's Hospital Stanford CF team, Russell's role includes improving access to medications, optimizing treatment plans and participating in research projects. You will see Russell in many different care settings, including inpatient admissions, clinic appointments and virtual health visits. In his spare time, he enjoys CrossFit, swimming and exploring the outdoors.



Teresa Priestley, MSW, has been a parent-educator for almost 20 years. She has extensive experience working with parents and children who have experienced trauma, separation or both. One of

the main objectives of her work is to empower parents to help their children heal from pain and trauma that they have experienced. She really enjoys assisting families in problem-solving. She looks forward to meeting all of you!

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Erica Oliva, patient service coordinator, has taken a new position in the inpatient setting at Packard Children's. Erica's last day in the CF Center was on January 12, 2018. In the interim, Lashaun Carr covered Erica's role. We would like to thank Lashaun for all her hard work and dedication to our CF families while we search for Erica's replacement.