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

Stanford MEDICINE

Cystic Fibrosis Center **News**

Reproductive health in cystic fibrosis, Part III

By Jennifer Canon, MS, RN, NP-C

For the third and final installment of our Reproductive Health in Cystic Fibrosis (CF) series, we will be discussing pregnancy in CF patients. Although there is an absence of comprehensive pregnancy guidelines for CF patients, the good news is that there is an increasing amount of research dedicated to this topic. From pregnancy preparation to postpartum life, this article will provide a good overview of this amazing journey. As with all things related to your health care, specific details should always be discussed with your health care team.

Facts and figures

As the age of our CF population continues to increase, the topic of pregnancy has become more prominent. Although not all of the data surrounding pregnancy in CF is in agreement, there is a good deal of evidence that the majority of CF women who wish to become pregnant have successful pregnancies. According to a recent review of the United States CF Registry, CF women who became pregnant experienced similar respiratory trends as non-pregnant CF women. (McMullen, et al., 2006) This review also found that long-term survival is not negatively impacted by pregnancy. Additionally, there is little evidence that CF women experience a higher percentage of obstetric complications as compared to non-CF pregnant women. (Edenborough, 2002) However, with these findings in mind, the data does suggest that there are a few medical conditions that can place the baby or the mother at higher risk for poor outcomes. These include pulmonary hypertension, a body-mass index (BMI) of less than 19, CF-related diabetes, Burkholderia cepacia infection and an FEV-1 below 50 percent. (Thorpe-Beeston, 2009)

In this issue

Pregnancy prep

The first and most important step in the pregnancy journey comes back to the basics — preparation. The ultimate goal of pregnancy preparation is to ensure a healthy and safe pregnancy for both the mom and the baby. For the most part, the preparation that CF women take is very similar to that of non-CF women when planning a pregnancy. However, there are circumstances that are unique to CF families that should ideally be addressed prior to pregnancy.

Family considerations: The first piece of preparation is open and honest discussions with your family members about what you each see as a part of your family plan: child care, finances, employment, potential for lung transplantation and end-of-life care are all essential topics of discussion.

Medical considerations: This is when your CF team comes into the picture. Prior to pregnancy, take the opportunity to be really honest with your care team and discuss the hard stuff. The most important thing to remember is that your CF team is there to support and guide you.

Ultimately, the decision of whether or not to start a family is yours. The team is there to ensure that you are able to do it safely.

Genetic screening: If your partner has not been screened for CF, it is recommended that he or she does so prior to pursuing pregnancy. Stanford has genetic counselors who will meet with both you and your spouse to guide you through this process.

Health optimization: The prenatal (pre-pregnancy) time is the perfect opportunity to get your health in order. This is detailed further in table 1 below.

Table 1: Health optimization prior to pregnancy

Item	Examples and notes
Complete all outdated annual labs	Complete blood count (anemia), metabolic panel (kidney health), vitamin labs, liver testing, screening for ABPA, etc.
Complete your annual glucose tolerance test	Women with CF are at higher risk for developing gestational diabetes, and this can lead to increased pregnancy complications (McArdle, 2011).
Optimization of your inhaled treatments and airway clearance	Have you established a good schedule or routine? Does your vest fit correctly? Are you experiencing any benefit from your treatments? If not, are there other options that would work better?
Optimization of remaining CF medications	Bowel regimen, sinus treatments, etc.
Establishment of a regular exercise routine	To strengthen the lungs and accessory muscles so that you are better conditioned during pregnancy
Ensure healthy nutrition	Weight, BMI and vitamin levels
Optimize mental health	Ensure that all psychiatric medications are safe during pregnancy. If not, discuss safe alternatives. Pregnancy is stressful. Ensure that you have the necessary tools to manage stress in a healthy way.

What to expect when you are expecting

Pregnancy can be an overwhelming time for anyone. The good news is that your CF care will likely change very little.

CF clinic visits: As you will be seeing your CF team more often, there is increased attention to your health and CF regimen during this time period. Early in pregnancy, you will be expected to see the CF team monthly, and they will want to see you weekly closer to your due date.

Hospital stays and antibiotic courses: As there is a lower threshold to treat pulmonary infections during pregnancy, most women will ultimately require an increase in the number of antibiotic courses (both oral and IV), as well as an increase in the number of hospital stays. (McMullen et al., 2006)

Medication changes: Unfortunately, there is very limited data regarding CF medications and their safety during pregnancy. The decision regarding which medications to continue versus discontinue should be discussed with the CF team as well as your OBGYN. Ultimately, the OBGYN will decide which medications are safe to use while pregnant. The good news here is that the majority of maintenance medications will remain unchanged.

Symptomatic changes: As your body changes with pregnancy, you may start to notice a change in your day-to-day symptoms. The most common complaints are a sensation of increased shortness of breath, nasal congestion, postnasal drip, increased acid reflux, constipation and urinary incontinence. These symptoms are universal for all pregnant women. However, given that CF already predisposes patients to these symptoms, they may be more exaggerated in CF patients.

The importance of nutrition in pregnancy: The importance of proper nutrition and weight gain cannot be emphasized enough during pregnancy. Low BMI is associated with premature delivery and low fetal birth weight. (Thorpe-Beeston, 2009) Weight gain of at least 10 to 12 kg is recommended for pregnant CF women, along with proper PERT and vitamin intake. This is to ensure that your body has enough fuel to properly maintain and grow a pregnancy. However, the answer here is not always, "more is better." For example, taking too high a dose of Vitamin A (>10,000 units per day) is associated with fetal anomalies. Therefore, it is essential that you work with the CF RD to ensure proper weight gain and appropriate vitamin intake. Closely tied to nutrition is diabetes screening. You will likely complete several oral glucose tolerance tests throughout your pregnancy to ensure that you have not developed gestational diabetes.

I've delivered. Now what?

Don't delay! It is already time to resume your full treatments. This is usually the hardest point of adjustment for all new CF moms. The most important thing to remember is that a healthy mom equals a healthy baby equals a healthy family. This is where all of that preparation pays off. Falling behind on your CF care doesn't help anyone. It may be a temporary fix, but in the long term it will start to take a toll.

The decision to start a family is an exciting one.

As discussed, there are a lot of topics to review with your family and CF care team. The goal is always to set you up for success. Research has demonstrated that pregnancy in the majority of CF patients is well tolerated. However, preparation is strongly encouraged. You are encouraged to talk to your CF team if this is something that you and your partner are thinking about.

References:

Edenborough, F. P. 2002. "Pregnancy in women with cystic fibrosis." *Acta Obstetricia et Gynecologica Scandinavia* (81:689–692).

McArdle, J. 2011. "Pregnancy in cystic fibrosis." *Clinics in Chest Medicine* (32:111–120).

McMullen, A. H., Pasta, D. J., Frederick, P. D., et al. 2006. "Impact of pregnancy on women with cystic fibrosis." *Chest* (129:706–711).

Thorpe-Beeson, J. G. 2009. "Contraception and pregnancy in cystic fibrosis." *Journal of the Royal Society of Medicine* (102:3–10).

Our Center's mission is to excel in cystic fibrosis care, to be partners with those we care for and to be leaders in the discovery process that will produce the cure of cystic fibrosis.

CFTR modulator therapies: Ivacaftor (Kalydeco®) and lumacaftor/ivacaftor (Orkambi®) are making an impact on the nutritional status of our patients

By Julie Matel, RD

With advances in the medical management of cystic fibrosis (CF) in recent years, we have seen an improvement in our patients' nutritional status. Ivacaftor (Kalydeco®) was FDA approved in 2012 for treatment of about 5 percent of the U.S. cystic fibrosis community, specifically those with the following genotypes: G551D, G1244E, G1349D, G178R, G551S, R117H, S1251N, S1255P, S549N and S549R. More recently, it was extended to include pediatric patients older than 2 years of age. Since ivacaftor's approval and more widespread use, research has shown that patients have reported experiencing fewer respiratory illnesses and substantial weight gain. One study, published in the *New England Journal of Medicine* by Acurso et al. in 2010, looked at the effect of ivacaftor versus a placebo in persons who have CF with the G551D-CFTR mutation. They found an average weight gain of 2.8 kg (6 lb.) in children taking ivacaftor versus children taking a placebo. The weight gain was rapid in the first 2 months for participants taking ivacaftor, while the BMI declined in those receiving a placebo. It has been hypothesized that an improvement in gut pH, which may make enzymes work more effectively, may be responsible for the improvements in weight. In some patients, an improvement in fecal elastase (an indirect measure of pancreatic status) has been found, although it does not completely normalize. Finally, there may also be a decrease in the energy needs of individuals taking ivacaftor.

The drug combination of lumacaftor/ivacaftor, known as Orkambi®, was FDA approved in 2016 for individuals with CF who are older than 6 years of age and have the delta F508 homozygous gene mutation. Modest improvements in symptoms, lung function and weight gain have been seen in individuals taking the drug combination compared to participants taking a placebo. Common side effects that occurred during the phase 3 trial included nausea (13% vs. 8% placebo), diarrhea (12% vs. 8% placebo) and flatulence (7% vs. 3% placebo). (Source: Wainwright, et al; 2015)

Commonly asked questions

What should I do if I experience side effects associated with Orkambi®?

If you experience diarrhea, frequent stools and/or abdominal pain, talk to your CF health care provider. Sometimes these symptoms will resolve without any intervention. However, if they persist, your team may consider recommending an adjustment in your enzymes, proton pump–inhibitor medication or bowel regimen.

With the improvements seen in fecal elastase, should I stop taking my enzymes when I start Kalydeco® or Orkambi®?

It is not recommended to discontinue your enzyme therapy without first talking to your CF health care provider. The decrease in fecal elastase levels seen in studies have been small, with levels below the normal range, requiring individuals to continue using their pancreatic enzymes.

I have been told to take Kalydeco® and Orkambi® with fat. How much fat is needed? What are some examples of high-fat foods that would be appropriate?

For better absorption, you should take the medications with fat-containing food, but there is no specific amount of fat that needs to be consumed. Choose high-fat foods for meals and snacks when taking Kalydeco® and Orkambi®.

Some examples of fat-containing meals include (some ideas here are from the medication's package insert):

Breakfast ideas:

- Oatmeal with almonds and dried cranberries
- Whole-milk yogurt
- Scrambled eggs with avocado on multigrain toast
- Bagel with almond butter or peanut butter
- Breakfast burrito (containing eggs and ham or bacon and cheese)

Lunch/dinner ideas:

- Caesar salad with salmon
- Pasta with cream or pesto sauce
- Fish tacos with avocado and cheese
- Tuna melt with mayo and carrots on whole wheat bread
- A slice of combination pizza

On-the-go snack ideas:

- Nuts, granola bars, peanut butter, cheese-filled crackers, whole-milk string cheese

Remember, when taking your Kalydeco® or Orkambi®

- Do not take St. John's wort (supplement used for depression, anxiety, sleep)
- Do not eat grapefruit or Seville oranges with Kalydeco (these foods are OK with Orkambi)

http://www.kalydeco.com/how-to-take-kalydeco
http://pi.vrtx.com/files/patientpackageinsert_ivacaftor.pdf

Recipe

Avocado pasta

(prep time: 10 min.; cook time: 10 min.;
total time: 20 min.; yield: 4 servings)

Ingredients:
12 oz. spaghetti
2 ripe avocados, halved, seeded and peeled
½ c. fresh basil leaves
2 cloves garlic
2 tbsp. freshly squeezed lemon juice
Kosher salt and freshly ground black pepper, to taste
⅓ c. olive oil
1 c. cherry tomatoes, halved
½ c. canned corn kernels, drained and rinsed

Instructions:
1. Cook pasta in a large pot of boiling salted water according to package instructions. Drain well.
2. To make the avocado sauce, combine avocados, basil, garlic and lemon juice in a food processor. Season with salt and pepper to taste.
3. With the food processor running, add the olive oil in a slow stream until emulsified. Set aside.
4. In a large bowl, combine pasta, avocado sauce, cherry tomatoes and corn.
5. Serve immediately.
6. Calories: 670.7 per serving (4 servings per recipe); total carbohydrates: 80.7 g per serving; dietary fiber: 10.5 g per serving; protein: 14.3 g per serving.

Recipe adapted from Comfort of Cooking.

Adult mental health update

By Meg Dvorak, LCSW

One year ago, the CF Foundation announced that mental health grants would be available for select CF centers to fund a mental health coordinator (MHC) position and expand mental health services to all CF patients. This movement stems from results of the TIDES study (Quittner et al., 2014) and the diligent work of the International Committee on Mental Health (ICMH). The CF Foundation has recognized the vital importance of mental health screening and care as part of a CF treatment plan to ensure the holistic approach to patient care and to maximize the physical and emotional well-being of each patient. The Stanford Adult CF Program immediately had a vision for this mental health grant, as the care team has had a unique partnership with Stanford faculty psychiatrist Dr. Liza Sher. As an expert in psychosomatic pulmonary medicine, Dr. Sher had already established a large practice of CF and lung transplant patients in her psychiatry clinic, and she was our gold standard for mental health referrals.

In early 2016, the Stanford Adult Program was selected as one of the 84 CF centers to receive the MHC grant. The funds from the grant were used to establish an embedded psychiatrist, Dr. Sher, in all CF clinics starting in April 2016. In addition to helping with mental health screenings and education in the clinics, Dr. Sher offers real-time psychiatric evaluations, medication management, psychotherapy and suicide assessment, as needed. Under the grant, Dr. Sher can see any of the clinic's CF patients regardless of their insurance status. Additionally, Dr. Sher can follow CF patients during hospitalizations, which allows for seamless continuity and tailored interventions based on an established relationship. In 6 short months, we have already begun to see the bridging of a large gap between CF patients and mental health services, as well as a reduction in the number of patients receiving psychotropic medications through a non-mental health provider.

As part of the MHC grant, the mental health coordinator provides prevention as well as education around mental health topics. Starting in June, we began to offer the "Wellness Newsletter" to all CF patients for every clinic visit. The newsletter is given out to patients by the respiratory therapists after receiving PFTs, which serves as a nice filler for long wait times in the clinic. The newsletter's topics have covered a variety of themes, including mindfulness, procedure anxiety and guided imagery.

A new edition of the newsletter comes out every 2 months. We presented our newsletter at NACFC in Orlando in October, and it was very well received and recognized. In fact, other CF centers around the world would like to borrow the idea and even use the same content.

As recommended by the experts from ICMH, the mental health coordinator (Dr. Sher) is responsible for tracking the scores of the mental health (MH) screenings (including the PHQ9 for depression and GAD7 for anxiety) for every patient and providing intervention for patients with elevated scores. Thus, every patient will be offered MH screenings at least once per year, and more frequently if indicated. Additionally, Dr. Sher can see any patient for MH needs regardless of the screening score. She has met approximately 40 percent of the 162 adult CF patients who attended a clinic appointment between April and October. We expect this number to rise next year as the multiple needs of our patients are just being discovered through screenings and deeper discussions about mental health. There has been a significant reduction in screening scores (with a lower number indicating less depression or anxiety) for those patients who have been re-screened after an intervention from Dr. Sher.

Our goals for next year are to get the paper screenings put onto tablets for easier use and tracking and to extend our screening and patient surveys to include adherence and patients' perception of mental health attitudes, needs and interventions. We want to know what is most important for our patients so that we can address these concerns.

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The Cystic Fibrosis Parent Advisory Council

By Kirsten McGowan

The CF Parent Advisory Council seeks to address the needs of all families seen at CF clinics that are part of Stanford Children's Health.

New members, hand hygiene and travel tips!

- **Welcome new council members!**
  We'd like to welcome two new parents who have joined the CF Parent Advisory Council in 2016. Arek is the father of a 2-year-old daughter with CF who comes to us from the Emeryville clinic (formerly Bay Area Pediatric Pulmonary), which is now part of Stanford Children's Health. Arek enjoys international travel, skiing, the outdoors and investments. Niamh is the mother of two boys, an 8-year-old with CF and a 5-year-old without CF. Her 8-year-old son was diagnosed with CF during her pregnancy and has received care at Stanford Children's Health since he was born. Niamh enjoys climbing, surfing, hiking and cooking. We welcome their experiences and insights in helping our council impact all CF families.

- **Hand hygiene: Do you know the 7 steps?**
  Hand hygiene remains the #1 way to reduce the spread of infection, which is important in the CF community! As recommended by the World Health Organization (WHO), Lucile Packard Children's Hospital Stanford encourages families to know the 7 steps of hand hygiene so they can stay healthy.
  - Going through all 7 steps ensures you clean ALL surfaces of your hands, including hard-to-clean areas like knuckles, thumbs and wrists.
  - The 7 steps work for BOTH cleaning hands with hand sanitizer and cleaning hands with soap and water.
  - Don't forget to dry! Hand sanitizer is only effective after it is dry, and wet or damp hands (after cleaning with soap and water) can spread 1,000 times more germs than dry hands.

- **CF travel tip of the day**
  Traveling with CF can be a major concern. What if my child gets sick? Will the doctors know how to care for someone with CF? If you are traveling and need to see a medical provider at another location (whether it's ER, urgent care, another CF care center or a pediatrician's office) make sure that the provider ALWAYS calls the hospital at (650) 497-8000 and asks to speak to the on-call pulmonologist. It is important that the CF team understands what is happening with your child so they can help coordinate care with all other medical providers.

If you have feedback or an idea/topic for the CF Parent Advisory Council, please email **Kirsten McGowan**, co-lead parent, at **kmcgowan@stanfordchildrens.org.**

Beat the bugs

7 steps to clean hands

*As recommended by the World Health Organization

Rub palms together.

Rub the back of both hands.

Interlace fingers and rub hands together.

Interlock fingers and rub the back of fingers of both hands.

Rub thumb in a rotating manner followed by the area between index finger and thumb for both hands.

Rub fingers on palm for both hands.

Rub the wrists in a rotating manner. Rinse thoroughly.

Pediatric CF Center Update

By Mary Helmers, RN

Did you know that 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 out the information for all your medical devices (e.g., 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 you will mail it to PG&E.

Our on-site urgent care clinic is located at 730 Welch Road on the first floor across from the clinical lab.
Hours of operation:
Monday–Friday, 5:30 p.m.–9:30 p.m.
Weekends and Holidays, 10:00 a.m.–4:00 p.m.

If your child is sick after clinic hours and you would like your child assessed, you can use the urgent care clinic. The clinic is staffed with medical personnel, including medical doctors, nurse practitioners, registered nurses and medical assistants. Make sure you bring your **CF PASSPORT** with you! The team has been instructed on the latest CF isolation policy.

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 for patients and parents to communicate with their provider and CF care team through email. Please note that any email sent to the team will be responded to with a phone call. The CF care team cannot respond by email because MyChart is not a secure site. We do not always check emails on a daily basis, so if you or your child has a clinical need or question, please call the CF RN line at **(650) 736-1359**. The front desk will be happy to assist you with the sign up.

Patients and parents: Remember to carry your **CF PASSPORT** in your wallet. The CF Clinic sent the PASSPORTS out in the mail to each family. However, it has been more than a year. If you did not receive one, or if you may have misplaced it or thrown it out, please ask for one when you come to your next clinic appointment. We now have them in English and Spanish.

Helpful reminders!
To help expedite your clinic visit, please remember to bring your CF binder with you to the clinic along with the 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, but 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 (this is especially true for mail-order pharmacies). It is important for you to stay on top of your refills and request them at least 1 week before you are due to run out.

Helpful hints for requesting refills:

- Call your pharmacy first to find out if you have refills.
- If you have a refill, great! Then they will process 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.

Annuals: Remember our goal is to get all annual testing done on or around your child's birthday. Included in the annual tests are lab work, CXR, a bone-density scan (for patients 12 years of age and older), full PFTs (starting at age 7) and audiograms (baseline at age 6 and then yearly). Please let us know if you have not had any of these tests done with your annuals.

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

Wear your mask: We have new turquoise-colored masks that we will ask all CF patients to wear. 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 visits and when walking 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.

Lastly, 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 patients' genotype/sweat chloride test results. If you have a copy or the original test 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 that lab or center and ask them to fax the results to **(650) 497-8791**, ATTN: Mary Helmers, RN, Pediatric CF Coordinator. We need these 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.

Current and upcoming 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 809-110 — Open-label extension study for subjects who participated in the Vertex 809 Part B and the 809-109 study protocols.
OPTIMIZE — For treatment of newly acquired pseudomonas.
PTC study — Open-label extension study for patients with stop mutations.
ProQR study — Enrolling now for F508/F508 adult patients NOT on Orkambi.
Prospect — Enrolling now. Observational study for healthy, normal people and CF patients.
SHIP — Study on the use of hypertonic saline in children. Enrolling children now.

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STANFORD ADULT
Cystic Fibrosis
Advisory Council

Stanford Cystic Fibrosis Patient & Family Advisory Council

By Brian Eddy

It has been another productive quarter for the CF PFAC here at Stanford. Two council-initiated projects were completed and delivered to the clinic for distribution, and we are one step closer to initiating an important study with the hope of enabling patients to better understand and manage their health. The *Inpatient Issue Management Decision Tree* brochure provides assistance to patients who have experienced an issue with their health care as an inpatient at the hospital. The brochure explains the options the patient has to safely report the issue without worrying that their care may be adversely affected. The CF Passport is a small, laminated, wallet-size information card that provides critical health care guidance to health care workers who may not be familiar with CF care when patients have an emergency situation that may prevent them from ensuring their needs are understood. In both cases, well-informed patients are better able to clearly advocate for their health care needs. Beginning in January 2017, another initiative is looking into the value of increasing the patient's role in the management of his or her health. The initiative's first step will be to evaluate the perceived value of patients monitoring their lung function at home to better understand their health and identify the onset of potential exacerbations.

If you would like information about the council or are interested in participating, please contact us at **stanfordcfac@gmail.com**.

New pediatric care staff members

Daniel Alvarez graduated from the University of California, Los Angeles, in 2016 with a Bachelor of Science degree in biology. He worked at UCLA Medical Center during his junior and senior years in between classes as a clinical research assistant in the Hematology-Oncology Department. After graduation, Daniel decided to move back to the Bay Area and was given the opportunity to join the Cystic Fibrosis Research team at Stanford as a research assistant. Daniel enjoys baseball and spending time with family and friends.

Karl Engel is from southern Maryland and graduated from the University of Maryland in 2014 with a bachelor's degree in chemical and biomolecular engineering. After graduation Karl worked as a postbaccalaureate research fellow at the National Institutes of Health in Bethesda, MD, before moving to California in 2015 to work as a research assistant with the Gurtner Lab in the Division of Plastic Surgery at Stanford University School of Medicine. In November 2016 Karl joined the CF Research Team at Lucile Packard Children's Hospital as clinical research assistant. In his spare time, he enjoys running, photography, traveling, and backpacking in the Sierras.

New adult care staff members

Dr. Yelizaveta (Liza) Sher received her Bachelor of Arts degree in molecular and cell biology with an emphasis in neuroscience from UC Berkeley. She finished medical school at Washington University in St. Louis, where she was honored to receive the merit-based, full-tuition Olin Scholarship. During her medical school training, she received several awards and was elected into AOA. She completed her residency in psychiatry and a fellowship in psychosomatic medicine at Stanford University Medical Center, and she was elected to serve as a chief resident during her final year of residency training. She has been a part of the psychosomatic medicine faculty at Stanford since 2013, where she also serves as an associate director of psychosomatic medicine fellowship. Her clinical and research interests include psychiatric co-morbidities in patients with pulmonary disorders. In particular, she specializes in the mental health needs of patients with cystic fibrosis as well as lung and heart transplant patients. She consults on patients who are hospitalized on medical and surgical units and sees patients in outpatient clinics. She combines psychopharmacological and psychotherapeutic interventions for her patients based on each patient's unique needs. Dr. Sher is honored to serve as a mental health coordinator for the Adult Cystic Fibrosis Clinic. Dr. Sher has been publishing in scientific journals, writing book chapters and presenting at national meetings. She is currently editing a new book on psychosocial care of patients with end-stage disease and transplantation. Dr. Sher is most passionate about connecting with each patient and finding the unique attributes, needs and strengths of each patient. She finds her patients very inspiring and appreciates their connections and trust.

Anastasia joins the Adult CF team as a social worker, alongside Meg Dvorak. She comes to the center after spending her first year as a float social worker in the hospital, where she was able to get a variety of experiences in many different areas. A native Californian, Anastasia has a Bachelor of Arts degree in psychology from UC Santa Cruz, and she went on to get a master's degree in social work from Boston University. She has a distinct interest in working in the mental health field and finds great inspiration from being in nature, from exercising, and from CF patients and their amazing families!

Congratulations Julie

The Grace Awards were established to recognize outstanding achievement and dedication in caring for children and expectant mothers. Congratulations to our own Julie Matel, MS, RD, for her outstanding contributions to our patients and families as she was named one of the Grace Awards "special honorees."

Adult CF Center Update

By Julian Liang, BSN, RN, BSE

As we look forward to 2017, the Adult Cystic Fibrosis team has a few reminders and recommendations for maintaining a healthy new year.

1) Establish care with a primary care physician (PCP)
We highly recommend visiting a primary care physician to take care of any non-CF issues. There are many adult health concerns that we are not properly equipped to care for. It is especially important for patients who live far from our clinic to find primary care and pulmonary care providers in their local area, if available. Our team is always happy to collaborate with your local providers.

2) Plan for Cystic Fibrosis clinic visits every 2 to 3 months
Our goal is to keep our patients out of the hospital as much as possible. We believe that proactive medicine and follow-up care is an essential part of keeping our patients healthy and at home. Of note, as a Cystic Fibrosis Foundation accredited center, we are required to see all patients a minimum of four times a year. Furthermore, insurance companies often require recent clinic notes to cover medications and equipment.

3) Annual labs and tests
Anticipate completing annual lab work, bone density scans (if indicated) and oral glucose tolerance tests (if not currently diabetic) at the first visit of the year. Our goal is to have all lab work completed by the first quarter of the year.

4) Insurance plan and pharmacy changes
Many insurance plans change what medications they cover from year to year. This often results in a need for a prior authorization. Insurance companies ask the office for recent clinic visit notes and data supporting the patient's need for the medication. Please be mindful of this and request refills at least 2 weeks in advance, if possible. Also, if your insurance plan changes, be aware that the preferred mail-order pharmacy may change, as well.

5) Sign up for MyHealth
https://myhealth.stanfordhealthcare.org
Stay in communication with your health care team! With the MyHealth app, you are able to message providers, view appointments and see most lab results.

Adult mental health update, continued from page 6

As we continue to collect data, we can see the year-by-year trends and adapt our practice to what is working. We will continue to screen, provide mental health support and follow patients when they are admitted to the hospital. This has been a long overdue focus in the CF care model, and we are excited about the future possibilities.

According to the recommendation put forth by the CF Foundation, starting in January 2017 we will re-start our routine annual screenings for depression (using the PHQ-9 tool) and for anxiety (using the GAD-7 tool). We are also introducing a new tool that will include six to seven scaled questions about adherence. This tool will help us gain a deeper understanding about patients' successes, needs and challenges related to adherence. The questions are not meant to judge or criticize, but rather to facilitate our conversation about how we can help patients manage their CF better.

Current and upcoming research studies, continued from page 9

Celtaxsys — Anti-inflammatory study in adults.
AbbVie — GI symptom tracker. Observational study.

Upcoming studies:

Vertex 809-115 — Phase 3 study for children 2 to 5 years of age with the F508 mutations.
Vertex 770-124 — Phase 3 study for children less than 2 years of age who have a gating mutation.
Vertex 440-101 — Phase 2 study for patients 12 year of age and older looking at the next-generation potentiator/corrector combination.
Concert — Phase 2, open-label comparator for patients with gating mutations.

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Palo Alto, CA 94304

Cystic Fibrosis Center at Stanford

Pediatric providers at Lucile Packard Children's Hospital Stanford: Carlos Milla, MD, Pediatric CF Center director; Sumit Bhargava, MD; My My Buu, MD; Carol Conrad, MD; David Cornfield, MD; Richard Moss, MD; Terry Robinson, MD; Jacquelyn Zirbes, DNP, RN, CPNP

Pediatric clinic scheduling: (650) 724-4788
Pediatric 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.**
All other times: For children's needs, call **(650) 497-8000**
(Lucile Packard Children's Hospital main number).

Adult providers at Stanford:

Paul Mohabir, MD, Adult CF Center director; Laveena Chhatwani, MD; Gundeep Dhillon, MD; Jennifer Cannon, NP; Elika Rad, NP; Meredith Wiltse, NP. Backup providers: Kelly Johnson, NP; Laura Starr, NP; Puja Sarna, NP; Matt Conlin, PA

Adult clinic scheduling: (650) 736-5400
Adult CF Center fax: (650) 723-3106

Nurse coordinator: (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
Carol Power, RCP; Gauri Pendharkar, RCP
Registered dietitian: (650) 529-5952
Michelle Stroebe, MS, RD; Marianne Schroeder, RD
Social work: (650) 518-9976
Meg Dvorak, LCSW
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
Visit our website at **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**
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