

Cystic Fibrosis Center News

Bacteriophage: What is it and Why Does it Matter?

— Elizabeth Burgener, MD

Pseudomonas aeruginosa is the most common organism cultured from the sputum of patients with cystic fibrosis (CF). Patients with CF acquire *Pseudomonas* over time, so luckily it is fairly rare in our pediatric patients. However, in the United States approximately 60 to 80 percent of adults with CF have chronic *Pseudomonas* infection. We know that patients with *Pseudomonas* have faster decline in lung function and a higher mortality rate than those without *Pseudomonas* infection.

Pseudomonas infection forms biofilms within the airway, which is one reason why it's so



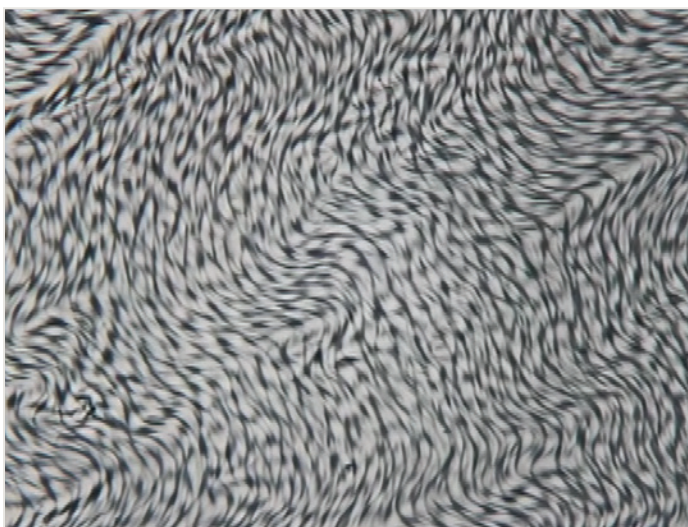
Elizabeth Burgener, MD

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hard to clear with antibiotics. A biofilm is a goopy, sticky mass of molecules that, in this case, protects the *Pseudomonas* from both antibiotics and the immune cells trying to fight the *Pseudomonas* bacteria. Dental plaque, the goop that lives between your teeth and your gums, is another type of biofilm. In the lab of Dr. Paul Bollyky of Stanford University's Division of Infectious Disease, a specific bacteriophage (a virus that infects a bacteria) was found to compose a large part of the biofilms made by *Pseudomonas*. In his laboratory, Dr. Bollyky and his team found that the Pf bacteriophage organizes the molecules and polymers in sputum into a biofilm that is actually a liquid crystal. That means the biofilm is organized and structured so that it will polarize light, just like polarized sunglasses! The lab found that when biofilms take on this liquid crystal structure, they are



Visualization of liquid crystal structure mediated by Pf bacteriophage visualized by light microscopy

stickier, thicker and more resistant to letting antibiotics through. They also decrease the ability of immune cells to engulf *Pseudomonas*.

The laboratory observed that all of the things the Pf bacteriophage does to *Pseudomonas* appear to make *Pseudomonas* harder to remove from the lungs and also make sputum thicker. I wanted to see how this affected our patients with CF. Over the past three years, the lab has been collecting sputum from patients with CF in our Stanford clinics. We have found that about 40 percent of our patients at Stanford who have *Pseudomonas* also have the Pf bacteriophage in their sputum. These patients with Pf bacteriophage are more likely to have chronic *Pseudomonas* infection and to have more antibiotic resistance than patients who have only intermittent infection. Both of these things make us think that Pf bacteriophage may actually help *Pseudomonas* stick around in the lungs and set up chronic infection.

We are continuing to follow patients with *Pseudomonas* (both those with Pf bacteriophage and those without) to track aspects of CF disease such as lung function, nutritional status, and other comorbidities over time to see if Pf bacteriophage affects other outcomes in CF. Additionally, my colleagues in the Bollyky laboratory are studying how the Pf bacteriophage affects the immune system and how antibodies against Pf bacteriophage can help clear (or prevent) a *Pseudomonas* infection. If these efforts are successful, this could lead to an anti-*Pseudomonas* vaccine, which would benefit all infants and children with CF!

Pediatric CF Center Update

— Mary Helmers, RN

Check out the CF Center website, which has recently been updated! Take a look and let us know what you think: med.stanford.edu/cfcenter.

The following information is on our center's 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: Link to the 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 form is not mandatory. Rather, it's a tool for jogging your memory in preparation for your clinic visit. Do you drive away from clinic thinking, "Oh no! I forgot to ask a question." You can now fill out this form ahead of time and bring it to your clinic appointment.

Keep an eye out for more new topics and informational tips in the next few months on our Facebook and center website.

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

Some helpful tips

Please check out our Facebook page and CF Center website.

Three podcasts were added to both sites.

- Traveling with CF, by Mary Helmers, RN, our pediatric CF nurse coordinator
- Gastrostomy Tubes in CF, by Julie Matel, CRD, our CF dietician

- Research Participation: The Basics and Beyond, by Colleen Dunn, CCRC, RRT, our 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 CF care team. The CF care team cannot respond to patient/parent emails on MyChart because 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. 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. One of the front desk staff will be happy to assist you with signing up.

Make sure you bring your **CF PASSPORT** with you to use around the hospital wherever you have an appointment, test or procedure. Parents/caregivers: remember to carry your child's CF PASSPORT in your wallet. If for some reason you do not have one or you tossed it, 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

3 Tips to Improve Your Next Respiratory Treatment

— Taylor Lewis, MA, CMT, CSCS, PRT

The goal of breathing treatments is to maintain or improve lung function, increase mucociliary clearance, improve quality of life and improve overall respiration. Breathing treatments often last 20 to 30 minutes per treatment and should be done 1 to 3 times per day, depending on the patient. This takes time out of your day and adds up over the week. What if we could boost how much you get out of these treatments? What if we could help you improve your next treatment? We can't guarantee anything, but we have put together three strategies for our pulmonary clients that have helped them improve their approach to their treatments.

The goal of respiratory treatments is to help you improve your ability to breathe, right? Well, let's look at how we can help you target that goal and possibly improve your quality of life and breathing mechanics, as well.

Number 1: Hydration

The adult human body is more than 60 percent water. The brain and heart are 73 percent water, the lungs are about 83 percent water, and our muscles are around 79 percent water. If we are dehydrated, our brain isn't going to be as efficient at operating and sending signals to the lungs and muscles for respiration. This could change our physiological chemistry, making it harder to breathe. When something becomes more difficult to accomplish, there is an added stress that is placed upon the body to survive.

This could decrease our energy levels and make us work even harder to breathe, all just because

we are dehydrated. We always recommend drinking water throughout the day, from the moment you get up to 30 minutes before you go to bed. Try having a glass of water 30 minutes before your next treatment. It could help hydrate your brain, lungs and muscles, allowing them to function at a higher output.

Number 2: Posture

Your posture directly affects your breathing mechanics. The human body is a fascinating organism. Our bodies are built to do amazing things. However, when our body does the same thing over and over again, it adapts and can alter our posture. This isn't a bad thing if you are training for a particular goal. However, it can directly affect your breathing mechanics if you do not allow your brain to enrich itself with other postures. For example, when you sit at work all day, your muscles will adapt to



Hydration

the seated position you maintain. In this seated position, your hip, neck and lower back muscles will tighten. These muscles are highly utilized in respiration. When they are kept in a prolonged state of shortening, the brain will tell other muscles such as the hamstrings, glutes, pelvic floor and core muscles to lengthen in order to balance out the adapted seated posture. This causes imbalances throughout the body and changes your breathing mechanics without you even realizing it. This seated posture will cause your head to subtly shift forward and your ribcage to flare up, and it will increase the recruitment of the secondary muscles in the chest, neck and back, which also aid in getting air into your lungs faster when you have an elevated heart rate or an increase in emotional and physiological stress. These muscles are primarily used when there is an increase in stress levels, so when they are recruited for other jobs, like stabilizing our posture for too long, they will become tired and overworked. This causes breathing patterns and respiration rates to change and creates a new pattern for getting air into the lungs.

Our body's number one goal is to survive. If we limit its ability to breathe efficiently, it will compensate and use other muscles, such as the neck, chest and back muscles, to drive your respiration. This can carry over to your breathing treatments, and you may not even realize it. That is why we recommend you stand up and walk around for a minute every hour when you have a job that is highly sedentary. It allows your brain to enrich itself with other postures and creates a balance between all postures (sitting, standing, lying down) that you use throughout your daily life.

Here is a posture strategy for your next treatment

Sit upright, with your ribcage subtly tucked down and your knees at a 90-degree angle under a 2-inch block — Posture A. Sit in this position during your treatment for as long as you feel comfortable. If you have something to squeeze, place it between your knees and subtly squeeze it to increase core activation — Posture B.



Posture A



Posture B

Tips for Caring for Your Home Respiratory Supplies (Nebulizer Cups, Compressors, Vests)

— Gauri Pendharkar, RCP, CPFT

1. Pari LC nebulizer cups can be used for up to six months if they are cleaned and disinfected DAILY.
2. When you use a new nebulizer cup, put a reminder on your calendar or phone so you don't forget 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 your pharmacy and see if there are any refills left. This saves time, and the pharmacy can simply fill the prescription for you. If there are no refills left, send a MyChart or MyHealth message to your RT coordinator to request that a new prescription be sent in for you.
4. To have a new prescription sent, it is very important that you let the RT coordinator know the name of the pharmacy that accepts your insurance.
5. Most compressors come with a warranty for 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, they will replace it for you if it is within the warranty period. If it is very old, you can send a message to your RT coordinator and a new compressor will be ordered for you. You can also always call Pari directly (for Pari compressors). Be sure to give the serial number of your device. Pari's customer service line can be reached at (800) 327-8632.
6. Remember to change the filters on your compressor once a year. Most compressors come with a few new ones.
7. Do not wait until the last minute to order necessary supplies. For example, if you are going to travel, plan ahead to see what you need and have it ordered well before your trip (especially if you're travelling overseas).
8. If you are having mechanical issues with your vest or need a hose or garment replaced, call the vest company directly.
9. Here are some commonly requested phone numbers:
Hill-Rom customer service: (800) 426-4224
Respiritech customer service: (800) 793-1261

A Research Experience

— Andrew Fulwiler

How did you hear about the study?

Zoe and Colleen always tell me about the up-and-coming studies. Also, sometimes Daniel pops in during my clinic visits and lets me know what's going on.

Did you have concerns about enrolling in a study?

I don't really have any concerns about doing CF studies because I've been doing them a lot ever since I was about 18. Plus, they have a detailed consent that describes the potential risks, so you can decide for yourself if you're comfortable after reading it.

What would you share with people with CF who are considering enrolling in a study?

DO IT! Not only is the research helpful for finding new and effective treatments, but they also usually compensate you for your time and effort. If you need to stay close by, they can often put you up in a hotel and pay for meals. All in all, I think it is worth it.

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

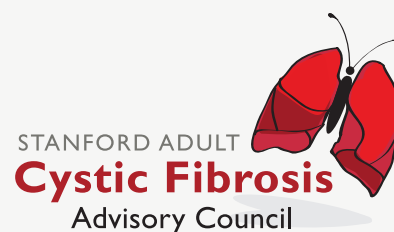
When is the next one?!



Andrew Fulwiler



Andrew's artwork



Stanford Adult Cystic Fibrosis Advisory Council

— Brian Eddy

The council is working on interesting new projects and has been pleased to welcome three new members over the past several months. The most exciting project currently underway is to designate an official piece of artwork in the new Stanford Hospital to memorialize patients we have lost to CF over the years. In addition, we are teaming up with multiple Stanford organizations to pursue other projects initiated by the council, from working with medical students to improve their understanding of the patient's viewpoint to helping define the needs of virtual healthcare. For more information about this council, please visit our website at cfcenter.stanford.edu/acfac.

Cystic Fibrosis Parent Advisory Council

Advisory Council is a group of concerned parents whose children receive care from the Stanford CF Center. We partner with members of the pediatric CF Clinic care team to provide the highest quality of care and service to patients and families. Do you have suggestions? Comments? Questions? Please don't hesitate to contact Amy Baugh at abaugh@stanfordchildrens.org or Kirsten McGowan at kmcgowan@stanfordchildrens.org. We'd love to hear from you!

Recipe

White bean and chicken chili with cheesy garlic bread

Ingredients for the chili:

- | | |
|--|--|
| 2 Tbsp. olive oil | 1 bunch (approx. 1 lb.) Swiss chard or collard greens, stems removed and leaves cut into 1-inch pieces |
| 1 large onion, chopped | |
| 4 garlic cloves, minced | |
| 2 lbs. ground chicken | |
| 1 tsp. salt, plus more for seasoning | 1½ cups frozen corn, thawed |
| 2 Tbsp. ground cumin | 4 cups low-sodium chicken stock |
| 1 Tbsp. fennel seeds | ¼ tsp. (or more to taste) red pepper flakes |
| 1 Tbsp. dried oregano | ½ cup grated Parmesan cheese |
| 2 tsp. chili powder | ¼ cup chopped flat-leaf parsley |
| 3 Tbsp. flour | |
| 2 15 oz. cans cannellini or other white beans, rinse and drained | |

Ingredients for the garlic bread:

- | | |
|----------------------------------|---------------------------|
| 1 loaf crusty bread, sliced | 2–3 cloves garlic |
| | 2–3 Tbsp. unsalted butter |
| 4–6 oz. cheddar cheese, shredded | |

Instructions:

To make the chili: In a large, heavy-bottomed pot or Dutch oven, heat the oil over medium-high heat. Add the onion and cook until translucent, about 5 minutes. Add the garlic and cook for 30 seconds, then add the chicken, 1 tsp. salt, cumin, fennel, oregano and chili powder. Cook, stirring frequently, until the chicken is cooked through, about 8 minutes. Stir in the flour and then add the beans, Swiss chard, corn and chicken stock. Bring the mixture to a simmer, scraping up the browned bits on the bottom of the pot with a wooden spoon. Simmer for about 1 hour, or until the liquid has reduced by half and the chili is thickened. Add the red pepper flakes and simmer an additional 10 minutes. Season with salt and pepper to taste and add the parsley. Ladle the chili into bowls, top with parmesan and serve with cheesy garlic bread.

To make the garlic bread: Blend the cheese, garlic and butter in a food processor and spread onto each slice of bread. Bake in a 400°F oven until golden.



White bean and chicken chili

Feature Article

Update from the 2018 North American Cystic Fibrosis Conference

— Karen Hardy, MD

The North American Cystic Fibrosis Conference was recently held in Denver. I always relish this meeting, and I enjoyed it immensely this year. It provides a feast of information, and happily much of it will now be available online for everyone's consumption. Going to a meeting is always energizing, and I want to share a bit of that enthusiasm with our readers. Nelson Mandela once said, "It always seems impossible until it's done." Each one of us can think about the impossible as a CF patient or a CF caregiver or a CF parent. Over the years, the impossible continues to shift. When I started as a cystic fibrosis physician, the impossible was figuring out the gene. In 1989, it was done. Then it became gene therapy in the next 10 years, which is still on the radar. Then it was modulating the protein rather than attacking the gene itself. In 2012, it was done. Pause for a moment. ... What is your impossible?

Over time, I have reflected more on health and total body wellness as opposed to a single drug or treatment for any of my patients with CF.

To that end, I volunteered to share a bit from the recent meeting.

I believe in another adage: Knowledge is power. How can I communicate some of the information from the meeting and make it understandable for my patients with cystic fibrosis who are in grade school or

high school? I will try with this discussion to offer some information that might help.

Class mutations

One of my favorite symposiums was called "Progress and promise of the CFTR modulator pipeline." The symposium's first speaker united us all with modulator 101. This "class" reviewed basic information about the protein products from different CF mutations. It is important to remember that the cystic fibrosis gene product functions as a chloride channel, and movement of it and sodium (salt) relate to water movement across the cell surface. One does not need perfect 100 percent function. Even around 10 to 25 percent function would be helpful for promoting healthy movement of salt and water at the cell surface. The normal protein structure was flashed on the screen as a very complicated multifolded item — wavy lines and circles. This is the transmembrane protein. Parts of it literally weave into the cell membrane. As many of you have heard or perhaps seen, there are six classes of mutations, and each of them varies in the amount, function or stability of the protein that the gene defect will ultimately produce. The severe mutations are associated with zero to very tiny amounts of CFTR, classes 1–3. The minimal function mutations are class 4–6. Stop here. Do you know the mutations you have? Go ask your parent or call your doctor. Find out what class mutations you want to pay attention to!

It was tested with one class mutation and ultimately approved for the whole group! Newer products in 2015 and 2018 combined Ivacaftor with a second small molecule for the F508del mutation. These medicines (also pills) correct and refold the frogs into swans that then can swim to the cell surface, and helped by Ivacaftor, open their beaks.

The small molecules that are used to treat these gene mutations have been labeled correctors, potentiators and amplifiers. A corrector is a molecule that will correct the protein folding and movement within the cell. A potentiator helps the gating function to open the channel. Amplifiers help prevent the breakdown of a product so that it is still present to continue functioning as a chloride channel.

Until now, large research dollars have been spent working on F508. At this meeting, it was announced that the CF Foundation will be spending half of its research dollars to attack the class 1 mutation – the stop codon problem. This is great news for 11 percent of the CF population.

We have all the class 4, 5 and 6 mutations to address. The CF Foundation vows not to forget anyone. Each of these groups are small and have very few cases. Sometimes there's only one person known in the world with a certain mutation. Standard large research studies will never be possible. How do we treat these rare mutations? Is it impossible? No! We need methods to help individual patients. The speaker likened this to cellphone evolution. The cellphone was a fabulous invention but it keeps changing. Think back to the very heavy, large first cellphones, which led to innovative flip phones and then groundbreaking smart phones. We discarded old cellphones and flip phones and moved to smart phones. Now we are continually

perfecting the smart phone. We're more likely to have upgrades and less likely to toss the entire phone. Likewise, we are perfecting medications for the F508del mutation. Two more small molecules are in clinical trials right now. Thus, the drug or pill being tested has three medications in it. It is an "upgrade," keeping the prior corrector and potentiator but adding a third additional small molecule. The CF Foundation is hoping that if current phase 3 trials are successful, the FDA might approve the new drug in 2019.

The organoid model

Three speakers talked about different models for the rare mutations that need to be fixed. First the organoid model. This is a technique in which a rectal biopsy is taken from a cystic fibrosis patient who has a rare mutation. Stem cells that can continually regenerate are present in the tissue from the biopsy. These cells can be grown in the laboratory on culture plates and will create sheets of tissue that then can form into sacs or spheres. The active cell surface is on the inside of the sphere. The official name for the small sacs of the cells are "miniguts." Once the person's rectal biopsy has been used to create a minigut, it can be used to test available small molecules (hopefully future drugs) from the biologic bank of test molecules to design a personal study just for that patient. The actual minigut is treated so that it will fluoresce and be easily visible when looked at under a microscope. The size of the minigut will enlarge to become a big sphere if the test molecule is causing improved movement of fluid, which will collect inside the sac and enlarge the size of that sac. They showed beautiful pictures of small green miniguts that were then treated with a test drug to improve the function and movement of chloride and water at the cell surface. As this happened, the sac enlarged and

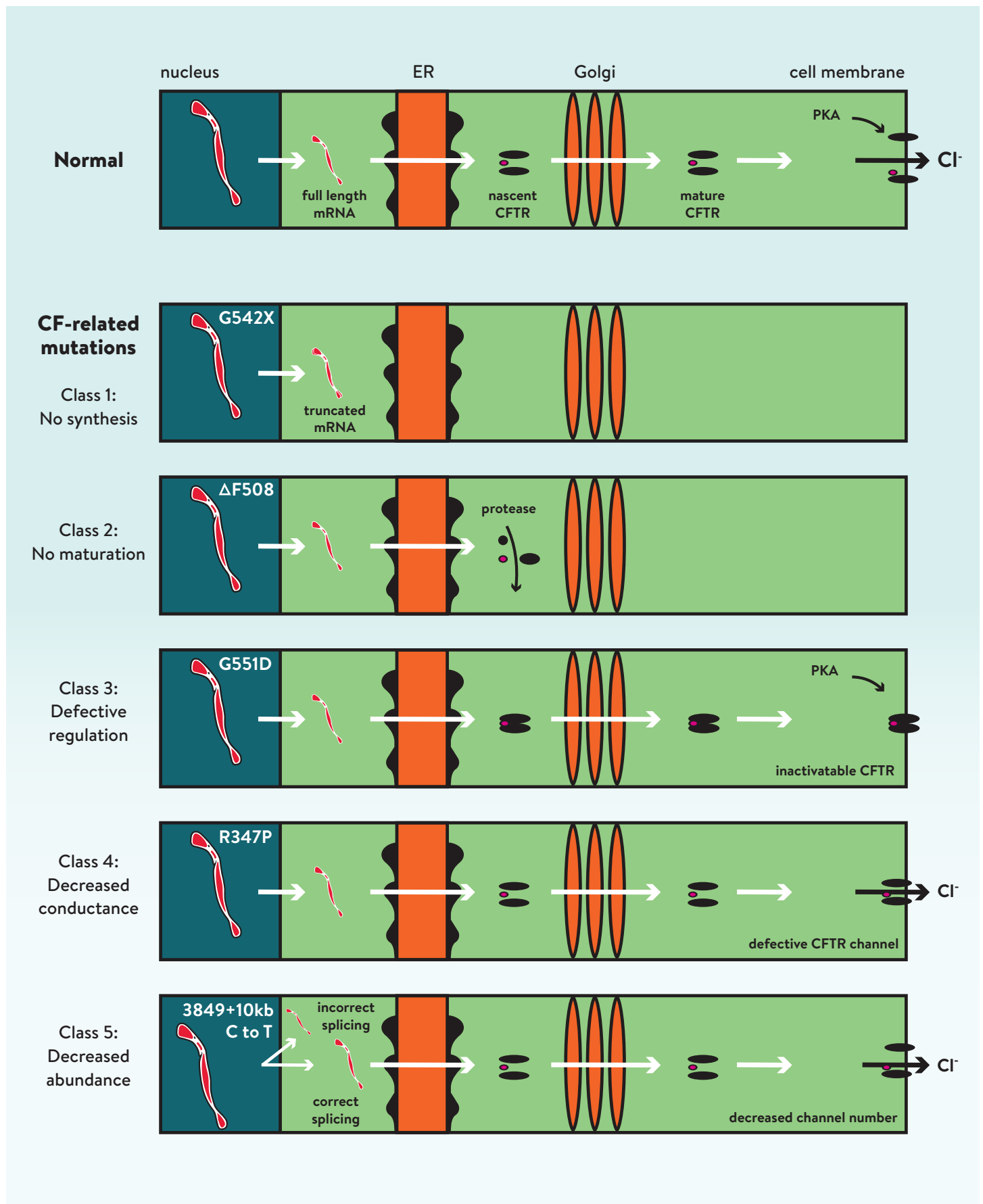
Over the years, this set of grids and diagrams got me thinking about origami, with its complex folding. Most of us have tried origami at some point in our lives. If you've tried it, you very likely managed to produce a swan or at least know what it was supposed to look like. Like the CFTR proteins, origami involves complex folding in order to produce the 3-D image you expect from a finished origami figure. I have applied origami to the grid for different types of gene mutations to help young children get an idea of normal function and the kinds of problems their gene mutations would cause. I present the scientific explanation and the origami analogy, which many people have told me they found helpful over the years:

The grid on *page 12* shows class mutations 1 through 6. Class 1 mutations have extremely little functional protein. The class 1 mutation involves stop codons (that X after the numbers of the gene defect your doctor says some of you have). These folks are not producing any significant amount of protein. Yet, very rarely, a tiny amount of functional protein is produced when the body ignores the stop sign. Overall, patients with these mutations have less than 1 percent of normal functioning protein. Class 2 mutations cause abnormal trafficking of the protein product, which itself is misfolded. They also have very limited function. Class 3 mutations have difficulty regulating (opening) the channel after the protein reaches the cell surface. Classes 4, 5 and 6 have minimal functional protein at the cell surface but do have some residual function.

The idea of modulators is to add a small molecule that can change the structure of the protein so that it becomes more functional. Even trying to write this down to explain it in the newsletter is challenging. If I change this model to origami, it

would run as follows. Imagine that each normal CFTR is a beautiful, intricately folded origami swan. Many of these would be formed in the center of the cell and, like all swans, would know how to swim to the cell surface. At the cell surface, the swans would poke their beaks through the cell wall and open the beak to allow easy flow of chloride. I can then use origami to help children understand the different mutation classes. A class 1 mutation doesn't do any origami. They simply don't know how to do it at all. A class 2 mutation does do origami, but instead of those intricate swans, they fold a frog. The frog only knows how to hop around and can't swim to the cell surface. It doesn't have a beak and cannot form chloride channel. So the body throws away the frog! A class 3 mutation does make an origami swan that does swim to the cell surface and pokes its beak through, but it doesn't know how to open the beak to allow the chloride channel to function. Imagine all those swan beaks stuck in the membrane without being able to open! The class 4 mutation makes origami swans that swim to the cell surface but can barely open their beaks. Hardly any chloride can pass through. A class 5 mutation is the loneliest one. There are so few swans. Even though each one is perfect, there are not enough swans to do the job of moving the chloride. Finally, a class 6 mutation forms swans, but each one is not stable, and they continually unfold or fall apart before they have a chance to work correctly. If I take this model farther, it helps me explain to children how their new medications work.

The impossible happened. This medicine called Ivacaftor was approved in 2012 and has been fabulously successful. It helps the swans open their beaks so chloride flows! The potential was there, and with the medication everything works.



the little green balls became larger green balls. This indicated that the drug tested was effective! Happily, rectal biopsies are not painful and are very easy to obtain. Thus, this model can be used to test drugs to change CFTR function among the class 4, 5 and 6 mutations. If one can predict the outcome using this model, patients with that mutation can then try that drug in the future. The Europeans are generating a large bank of test drugs for this purpose. They're collecting 700 rectal biopsies from cystic fibrosis patients to make the organoids' miniguts. A number of labs across European countries are working together, and each have been getting very consistent data as they replicate these experiments. They hope to be screening drugs throughout 2018 and then moving them into clinical trials by 2019 with usable results by 2020. You can just taste the impossible becoming possible!

Splice mutations

The next exciting talk was an update on splice mutations, or class 5 mutations, which result in a decrease in the amount of functional protein. A gene is transcribed to produce mature messenger RNA in order to produce a protein. Not all of the gene is used to produce the messenger RNA. Some parts of the gene are removed (introns) other parts are kept (exons) in order to make the protein. If the splicing of the transcript is incorrect, the messenger RNA and protein will be incorrect and less functional or non-functional. These types of mutations are more common in the Jewish population, and Dr. Kerem from Israel gave this lecture. She wanted to change splicing to correct it, and she has achieved success using short RNA like antisense oligonucleotides, or ASOs. She has been able to create a number of ASOs that promote normal splicing, and she has been able to prove that

they do change the protein to make it functional. She has achieved 80 percent function with this technique for the first mutation that she addressed. Her model uses human cells that are either from the thyroid or from human nasal epithelium. She published her proof of concept in 2017 and has been able to completely restore function (in vitro) for five patients who had the 3849+10 Kb C ->T mutation. She has applied this to a number of other defects and restored up to 43 percent of wild type function. In summary, splicing with ASOs can result in high levels of full-length CFTR with high levels of function even with different gene combinations. She has started a company called Splisense and is trying to get funding in order to do more in vitro work. She hopes to generate a product that will be inhaled because it is not possible to use ASOs in the bloodstream. Taste that possibility!

Nonsense mutations

Finally, we heard about nonsense mutations, the class 1 stop codon or X mutations. These types of mutations cause many human diseases (over 1,800), such as Duchenne muscular dystrophy, Hurler syndrome, spinal muscular atrophy and certain forms of hemophilia. With this type of CF abnormality, 99.9 percent of the read-through does not occur because of the stop mutation. However, 0.1 percent of the time, CFTR is produced. The body ignored the stop sign (just like people do while driving) and read right on through, making a usable product. Unfortunately, this product is broken down. The speaker's group hopes to amplify the correct work that the body is already doing and stop degradation of CFTR when produced. Long ago, we learned that gentamicin can do this in the test tube and improve the functional product for patients with this mutation. However, this agent is too toxic to

use as a therapy. The speaker has proven there are in vitro agents that will work in conjunction with Ivacaftor on human bronchial epithelium, and he has published this work in the American Journal of Respiratory Cell and Molecular Biology in 2013. He has now screened over three quarters of 1 million compounds and has found working options that, in combination with some of our current correctors, are going into trials with rats. They have developed a G542 X-homozygous

rat that also develops mucous obstruction and lung disease and will be used as a model for this work. His impossible dream is to improve read-through and stop nonsense mediated decay of the CFTR that is produced. It will be done!

So, my final comment is, "Carpe futurum!" Seize the future! Be confident in your hope for tomorrow and dream your impossible dream. I'm looking forward to meeting more of you!

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

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

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New Adult CF Clinic Staff Members



Mary Jane Ramil is a registered nurse with 23 years of experience. She received her bachelor's degree in the Philippines and obtained her Master of Science in Nursing with a specialty in

informatics from Holy Names University in Oakland, California. Her experience includes managing an in-center hemodialysis unit, where she found a passion for helping people with chronic illnesses. She is looking forward to her time here at Stanford and believes it will give her more experience to fuel that passion. Besides helping people, Mary Jane likes to play the piano, cook and make ribbon leis.



Kati Lebowitz was born and raised in Dallas, Texas. She graduated with her BA in medical sociology from The Ohio State University and received her BS in nursing from Nova Southeastern University. Her nursing

background is primarily in acute care, ranging from the emergency department to the OR/PACU to interventional radiology. She moved to California from Austin, Texas, where she was an RN coordinator for interventional radiology. She is excited to be in California and proud to be a part of the cystic fibrosis team here at Stanford!