UCSD/Rady Children's Hospital

Fall 2023

SALTY TIMES

CF News

Why Do My Visits Take So Long?

Dr. Meerana Lim

The national Cystic Fibrosis Foundation (CFF) mandates that patients with cystic fibrosis (CF) be seen every 3 months. These regular visits have been proven to improve both quantity and quality of life for people with CF. In order for these visits to be useful, you are seen by different people who work together to make sure your child's health is being optimized in every way possible.

When you arrive, the check in person will verify your contact information and your insurance. This is important so that we can reach you in between visits if needed and so that your requests for labs, procedures, equipment and prescriptions all go to the right place from the start. If this information is not correct, it can lead to delays in care for your child.

Next, the medical assistant (MA) will measure your child and obtain vitals signs (blood pressure, temperature, pulse, oxygen level and breathing rate). These measurements are important for each visit and also important to track over time. Once these measurements are obtained, you will be seen by other members of the CF care team. You may see different team members at each visit depending on your child's specific needs.

At least once a year, you will be seen by a registered dietician who will take a detailed history of your child's nutritional intake and review things like vitamin levels, calorie needs and whether your child may benefit from supplements. If your child has been struggling to gain weight, you will likely see the dietician more often than once a year and if concerns come up during a visit, we may ask the dietician to see you even if that was not planned at the start of the visit. If you have questions, you can always request to see the dietician as well.

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At least once a year, you will be seen by the social worker. Our social workers perform screening for anxiety and depression for patients 12 years of age and older as well as for caregivers of our patients with CF. This is a CFF requirement as it has been shown to directly impact patients' physical health. In addition to yearly screening, our social workers are a resource for you as your child enters school, participates in extracurricular activities or has other needs that come up in the course of their ever changing lives. Our social workers stay up to date on community resources (local and national) that are available to your child and may be able to provide assistance in ways that you did not anticipate, just ask.

Almost all visits will involve seeing the respiratory therapist (RT). When your child is old enough, the RT is the team member that guides your child through pulmonary function testing. Before then, the RT will ask you about how secretion clearance is going at home and how things are going with your respiratory treatments. They can provide hands-on demonstration of various forms of secretion clearance and provide guidance for your equipment you use at home. Sometimes we even set up telemedicine visits with the RT so you can have real time treatments with your RT. Another option is to bring in your home equipment if you have questions or concerns about how to use anything or if it fits.

At least once a year, you will see the pharmacist. As you all know, CF medications can be quite specialized and not every local pharmacist will be up to date on all of your child's medications. Our clinic pharmacists regularly assess what medications your child may be eligible for and talk to you about medication side effects, drug interactions and things you should be on the look out for with specific drugs. They are also a great resource for potential patient assistance programs you may be eligible for. If you are having trouble obtaining your child's medications or have question about any of the medications your child takes, be sure to ask to speak with our pharmacist. Our pharmacists also frequently assist with insurance authorizations and can speak to your insurance company or your local pharmacy to help clarify prescriptions for you.

We are a teaching hospital, so you may see more than one doctor as part of your visit. We have medical students, residents and fellows rotating through our clinics at different times and it is a critical part of their training to see patients with CF. The medical student you see at your next visit could be your child's CF doctor 5 years from now. The fellow you see could be doing research that leads to a new drug for CF. Any of the doctors in training will report back to one of the regular CF doctors who will always see you at a routine CF visit.

If you are attending a CF-GI (gastroenterology) visit, you will also see a GI doctor; and if you are attending a CF-Endo visit, you will also see an Endocrinology doctor. That doctor may also have a nurse or doctor in training with them as well.

If you are participating in one of our research trials, you may get a visit from the research staff as a part of your visit and will likely have gotten a reminder about that through MyChart a few days before your visit. If we are recruiting for a new trial as part of the CFF therapeutic development network, we may ask you if you are interested in hearing details about that trial at your visit or set up a separate time to get that information.

And last, but certainly not least, you will be seen by a nurse or the nurse coordinator. Our nurse coordinates with all of the other team members to make sure your child is getting everything they need. Any concerns about labs, medications, equipment, visits, forms… can start or end with the clinic nurse. If you're not sure who to ask your questions to, it can start with the nurse.

The other question that comes up regularly is, why do so many people ask me the same questions?

One example of this is your child's medication list. When the MA asks, it is so that we all have the basic list of what you have been prescribed and if it has changed since the last visit (by your pediatrician or another physician you saw outside of our clinic). When the RT asks, they want to know how often you are actually doing the albuterol, if you are using a spacer with the inhaler, or which specific nebulizer cup you are using and what sorts of issues are you having with delivery. When the pharmacist asks, they want to know if you were affected by the supply chain shortage and if your co-pay changed unexpectedly and was it related to an insurance issue. When the social worker asks, it is because at the last visit you let us know that your child was starting to ask why they had to do medications that their brother does not have to do.

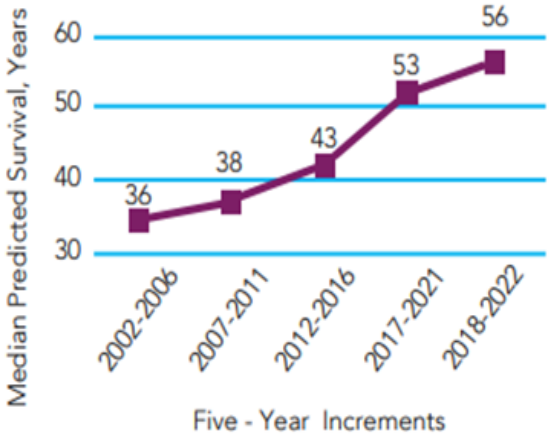
Because there are so many people on your child's CF team, it can take a while to get through the visit. We do talk to each other and review the medical record, but often we still have questions or need further clarification, so please be patient with us when we seem to be asking you the same questions over and over again. We are constantly working on ways to be more efficient, and we welcome your ideas and suggestions for how we can achieve this.

"In order for these visits to be useful, you are seen by different people who work together to make sure your child's health is being optimized in every way possible."

SURVIVAL

56
YEARS
2018 - 2022

Among people with CF born between 2018 and 2022, half are predicted to live to 56 years of age or more. This does not reflect individual variability in survival among people with CF.



CYSTIC FIBROSIS FOUNDATION

CYSTIC FIBROSIS FOUNDATION
4550 Montgomery Ave., Suite 1100N
Bethesda, MD 20814
800-FIGHT-CF

SOURCE DATA
Cystic fibrosis patients under care at CF Foundation-accredited care centers in the United States, who consented to have their data entered.

SUGGESTED CITATION
2022 Cystic Fibrosis Foundation Patient Registry Highlights
Bethesda, Maryland

FYI

Information and Reminders

Mask Reminder

Caregivers are no longer required to wear a face mask when coming clinic. However, we would like to remind you of the pre-pandemic policy that ***all persons with CF need to wear a face mask when in our hallways.*** Mask may be removed once in the clinic room.

This is consistent with CFF guidelines for infection control and provides protection for your child. This rule applies to any of our buildings and indoor areas. Please make sure your child is wearing a mask when at lab, radiology or the cafeteria. If eating indoors at the cafeteria, masks should only be removed once sitting at a table to eat.

Under Construction

Rady Children's Hospital is currently undergoing a campus development plan which includes construction of a new ICU and Emergency Services tower. This investment will help us continue to serve the community for generations to come. During this time, on-campus parking is being prioritized for patients to ensure minimal disruption to patient care.

Construction will continue through 2027. During this time, Sharp will also be adding to their existing facilities. We will do our best to provide updates as they effect clinic visits but do not anticipate any major disruptions for our patients and families.

During demolition, excavation and construction, all precautions are being taken to minimize dust and mold particles in the air. However, there may be increased levels of inhaled particulates in the air as you visit the Rady campus over the next few years. If you want to reduce your risk of inhaling these construction-related particulates, please consider wearing a mask outdoors as you walk around the campus.

RT Corner

Cold and flu season is fast approaching and while things are a little different since the pandemic, we wanted to remind you what to do when your child gets sick with a cold or even just has an increase in cough.

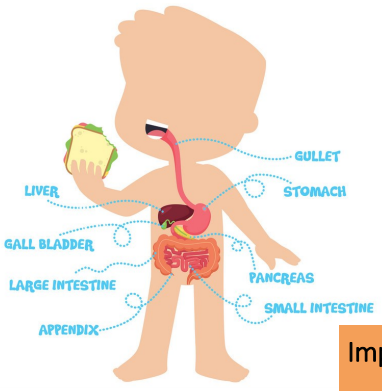
Sick Plan: What to do if your child becomes ill with cough.

- Increase breathing treatments up to 4 times a day.
- Increase vest/cpt/Aerobika treatments up to 4 times a day.
- Typical vest settings are Hz 11-14 Pressure of 5. When sick, increase to Hz 12,14,16,18 and Pressure of 6, as tolerated.
- Call for cough, congestion and/or any other symptoms that are interfering with sleep or eating/drinking or for symptoms that are not improving in 5-7 days.
- If there are lingering symptoms lasting longer than 2 weeks, please call the nurse coordinator. DO NOT USE MYCHART FOR MESSAGES.
- For urgent matters on nights or weekends, please call the operator at (858) 576-1700 ext.0 and let them know that you are followed in Cystic Fibrosis Clinic and need to speak with the Pulmonary doctor on call.

Enzymes: All you need to know

Sandy Salzedo, MS, RDN,CLEC

Digestive enzymes, essential for food digestion, are produced in the pancreas, which is connected to the small intestine where most of the digestion takes place. However, in people with CF, thick mucus obstructs the pancreatic ducts, preventing the digestive enzymes from reaching the intestine and causing poor digestion, absorption, poor nutrition and overall health. Fortunately, there are various enzyme brands, such as Pertyze, Creon, Zenpep and Pancreaze, available to help mitigate this issue and improve the quality of life of people with CF.



What are they?

Enzymes come in capsule form. Inside each capsule, there are small beads that contain digestive enzymes, amylase (helps break down carbohydrates), protease (helps breaks down protein) and lipase (helps break down fat). These enzymes also have a protective coating that allows the beads to dissolve in the intestine, precisely where food is undergoing digestion.

How do I take them?

Enzymes should be taken before every meal and snack. For infants, parents can open the beads and mix the contents with a small amount of applesauce, which helps preserve the protective coating, and give them via spoon. Children and adults, who capable of swallowing whole capsules, should swallow the capsules with enough liquid (for example juice or water).

Why are they important for me?

Ensuring a healthy absorption of the food you eat will impact your nutrition and the health of your lungs. Enzymes play a crucial role in breaking down the food you eat. If enzymes are not taken, then your body is not able to properly break down the foods you eat, leading to potential issues such as poor weight gain, weight loss, and overall compromised health.

What are the signs of malabsorption?

Malabsorption can manifest through various symptoms. These may include frequent large bowel movements or intestinal blockage. Additionally, individuals may experience foul-smelling stools, visible oil, orange fluffy colored stools. Other signs may include, larger quantity of stool, frequent gas, belly pain, cramps, distention or bloating. Others may also experience increased hunger. In severe cases, malabsorption can lead to rectal prolapse, which occurs when the lining of the rectum protrudes through the anus and outside the body.

Important general information:

- Enzymes should be taken before eating. They are needed with all meals and snacks, milk, breast milk, formula, nutritional supplements.
- Enzymes are most effective for about an hour after taking them.
- Do not change your enzyme dose on your own, taking too little or to much can lead to problems. Always reach out to your CF team for guidance
- For infants and young children who need to open capsules, do not crush or chew the enzyme beads.
- Enzymes should not be added to food ahead of time. The enteric coating will start dissolving and they will not work well.
- Enzymes need to be stored in a cool dry area (at room temperature 59-86°F).
- Enzymes should NOT be heated (for example: add to food before cooking, stored near oven or in a trunk of a car).
- Enzymes should not be stored in the refrigerator or cooler.
- Often check the expiration date on the bottle to make sure enzymes are fresh.

Find the Fats!

Can you find the fatty foods that are good choices to have with your Trikafta, Kalyedco, Orkambi or Symdeko?

S T B L V Z C R O I S S A N T S Y F O A
F I K U Q G R E E K Y O G U R T K H U N
D C D M T G T A P E A N U T B U T T E R
F D H W M T L U Z W M T M I L K J W U L
S V U A Q C E J B E A O J L Q P X Y C M
V K V W D B A R N U I L N U T S G K H A
Y V D M M Z A S M K T A N K S S X P I C
J S X W S O I L H I X T U U O I D Q C A
G H E H O M N F M E L D E K T C J M K D
Q J G C O O L T P O W K V R B S A T E A
K F G H I Z G F E I N S B S T E A K N M
G T P E L Z T P N R S D A I Q V U B D I
E T H E S A K C Z Q R T B G S I R L R A
A N B S P R U M J H B E A U K C V V U Q
V N D E M E W J G C M F Y C T T U U M I
O O H R P L B I C Y S C E J H T G I S L
C L B Y E L C H E D D A R M A I E G T M
A Z T N L A M N C O C O N U T C O R I S
D A L M O N D S J Z O U A N K I K S C S
O J O L I V E S Y V E E J T V M P H K V

Egg	Chicken Drumstick	Steak	Milk
Cheese	Mozzarella	Cheddar	Monterrey Jack
Greek Yogurt	Nuts	Almonds	Macadamia
Cashews	Walnuts	Pistachios	Coconut
Peanut Butter	Almond Butter	Oils	Butter
Avocado	Olives	Croissants	Buttermilk Biscuits

Egg (1 item) = 5 g fat
Chicken drumstick (3.5oz) = 5.7g fat
Steak (3 oz) = 16g fat
Whole milk (8 oz) = 8 g fat
2% fat milk (8 oz) = 5 g fat
Whole milk mozzarella (1 oz) = 6 g fat
Part-skim mozzarella (1 oz) = 4.5g fat
Cheddar/Mont Jack Cheese (1oz) = 9g fat

5% fat Greek yogurt (7 oz) = 10g fat
2% fat Greek yogurt (7 oz) = 4g fat
Nuts (1 oz):
- Almonds = 14 g fat
- Macadamia= 22 g fat
- Cashews = 13 g fat
- Walnuts = 19 g fat
- Pistachios = 13 g fat

Peanut Butter (1 Tablespoon) = 8g fat
Almond Butter (1 Tablespoon) = 9 g fat
Butter/oils (1 teaspoon) = 4-4.5 g fat
Avocado (¼ cup sliced) = 5g fat
Olives (10 large) = 5 g fat
Coconut (⅓ cup fruit) = 13.5g
Croissants (medium size) = 12 g fat
Buttermilk biscuits (medium) = 5 g fat

Parent Perspective

Monica Villalobos, *CF Parent*

One of the things that have been clearest to us as a family since we came into the world of CF, is that we must treat our daughter as we treat her other siblings. This is so that she can lead as normal a life as possible, and this is what we have done as a family. This is clear at home, but what about the other members of the family? Does the extended family know how to behave after a heavy treatment?

I have noticed that based on the love that one feels as a family member of a person with a medical condition, and the feelings that this situation can generate, some family members may have behaviors that based on love or compassion can be counterproductive for our CF children. That is why after a
hospitalization or a long treatment I recommend having a talk with the extended family so that they can lovingly support the situation without having harmful behaviors for our children. It is important to emphasize that it is not necessary to overprotect our children, that there be no favoritism or advantages, that all treatment be equitable among all family members, also that they be considered for responsibilities and be allowed to make their own decisions, this so that they can face their mistakes and learn conflict resolution.

"In general, what we want is for our children to have the opportunity to lead as normal a life as possible."

MyChart: changes as your child ages

Access by Ages:

For children under 12 years old:

Parents are given complete access to their child's health information.

For teens 12 to 17 years old:

Parents are given limited access to their child's health information.

For adult patients 18 years and older:

Parents/caregivers are **NOT** given access to their child's health information.

Why does parent access to their child's information change by age?
Parent access to their child's information changes over time, based on privacy concerns.

When a child becomes a teenager **(defined as 12 to 17)**, California confidentiality laws dictate what health information can or cannot be fully shared with his/her/their parent. While we encourage teens to discuss all medical concerns with their parents, teen privacy rights must be taken into consideration.

For more information on what is visible to parents/caregivers please visit:
www.rchsd.org/mcyhar-information/faq/

When a child becomes an adult **(defined as 18 and older)**, by law, parental access to the chart is revoked and only the now adult patient has full access to his/her/their health information. For this reason, because ONLY the now-adult patient will have access, **it is essential that a teen account be setup prior to the 18th birthday** so access to the medical record is not lost at age 18 years old.

Can adult patients grant their parent/caregiver access to their health information?
Yes For caregivers/parents to receive access to their child's health information, the adult patient must grant parents/caregivers access via a standard process.

Not signed up?

Please visit: www.rchsd.org/mychart-information/enrollment/
Or
Sign up at your next visit at the check in desk

Get to know your CF Team!

Meet Savannah

Hi! My name is Savannah and I have been a Respiratory Therapist for over 13 years now. I began my career working in the adult world for the first 7 years. I have worked at Rady's for 6 years now and I absolutely LOVE working in pediatrics! I initially started in the long term care unit at Rady's up until I was accepted into the Adult to Pediatrics program. Upon completion of the program I worked in the inpatient care setting in CTICU, PICU, Med/Surg, ED, and of course, on the Pulmonary unit. Although now I am more than ready to embark on my next journey within my career as I have taken on the role as CF RT at Rady Children's CF Center. I was recently accepted into the CF RT Mentorship program and have been paired with a CF Respiratory Mentor from The University of Texas Health Science Center San Antonio Pediatric and Adult CF Center. Over the course of the next few months I will be participating in conference calls with my mentor, educational webinars, and will be preparing for a site visit to the San Antonio, Texas CF Center. I am grateful beyond measure for the opportunity to continue learning and growing professionally.

A little bit about me personally, I was born and raised in San Diego. I absolutely love being outdoors, trying new things and pretending to be a tourist in my own hometown. I am a firefighter wife and we share 4 pretty amazing kids: Aaliyah 14, Kalea 12, Chance 8, and our little wild child Ace is almost 2. I really look forward to getting to know you and your families more and I am so grateful for the opportunity to work alongside such an incredible team!

Good-bye and Good Luck!

We are sad to announce that our CF Nurse Dae has left for a new and different professional opportunity. We would like to thank Dae for her dedication to our CF Center and patients while she was here. Good luck Dae!

Get Involved with your CF Center

Become a Parent Advisor to the CF Team !

Interested? Email Stephanie at Slord@paloaltou.edu for more info.

Submit an article!

We accept submissions from families to share their perspective in living with CF.

You can also share a cool experience your child had or any of their artwork.

If interested, please send a MyChart message to your team!

Cross word answer key

Contact us

Stacey Mount, Program Coordinator
(858) 966-1700 ext 224562
Denay Gonzales, Social Worker
(858) 966-1700 ext 225340
Courtney Larson, Social Worker
(858) 966-1700 ext 224212
Sandy Salzedo, Dietitian
(858) 966-1700 ext 225016

Martha, Savannah and Cathy
Respiratory Therapists 858) 966-5982
Dr. Lim or Dr. Akong
(858) 966-5846, option 1
Lisa Ramos, Research Coordinator
(858) 966-1700 ext 224127

CF Foundation

San Diego and Imperial Counties

www.cff.org/chapters/san-diego-chapter

2023 EVENTS

9/9 Surf for CF
9/30 Grand Chef Throw Down

CF FOUNDATION CARES | CYSTIC FIBROSIS FOUNDATION

CF Foundation Cares Dinner – Friday, October 6 @ 5:30pm, North County location TBA
As a family member, caregiver, or friend of someone living with CF, we invite you to join others to connect, encourage, and share experiences. CF Foundation Cares is a casual gathering created to help individuals caring for people with cystic fibrosis find support within other members of the community. It is intended for conversation, building relationships, and encouraging families, friends, and caregivers along the CF journey. This CF Foundation Cares event will be held on Friday, October 6 at 5:30pm in North County. Dinner will be provided and there is no cost to attend this event. If you are interested in attending, please contact Jamie Weissburg at the Cystic Fibrosis Foundation at jweissburg@cff.org.