

## Exciting updates about CFTR modulators

On Thursday May 30th, Vertex Pharmaceuticals announced that it will be pursuing FDA approval for the triple combination therapy VX-445 (elexacaftor) plus tezacaftor/ivacaftor. We have been awaiting to hear which compound was to be selected after two phase 3 trials of the compounds VX-659 and VX-445. VX-445 (elexacaftor) was selected due to its favorable safety and tolerability profile when compared to VX-659.

In patients with one F508del mutation and one minimal function mutation, the VX-445 triple combination therapy was associated with an average increase in the ppFEV1 of 14.3%, and a reduction in CF exacerbations by 63% when compared to those who were taking a placebo. Additionally, those on the triple combination therapy were found to have improved respiratory symptoms and improved weight (body mass index improvement of 1.04 kg/m<sup>2</sup>). Similar improvements were also seen in those with two copies of the F508del mutation. In comparison, Orkambi<sup>®</sup> and Symdeko<sup>®</sup> were found to improve ppFEV1 by approximately 4%.

The FDA approval process typically takes 3 to 6 months and we are hopeful that this new therapy will be available by early 2020. We will need to wait and see if the FDA chooses to approve this for ANY patient with an F508del mutation, or ONLY those with the specific mutation pairs that were tested in the clinical trial. All patients 12 and older, who are currently taking Orkambi<sup>®</sup> and Symdeko<sup>®</sup>, should likely be eligible for this new therapy as well. Once approved, it may take some time for insurance companies to make decisions about who they will cover; our clinic expects many insurers will require prior authorization for the new triple combination therapy.

Clinical trials are underway for those ages 6 to 11. We hope that if approved by the FDA, over time eligibility for this therapy could expand to up to 90 percent of people with CF.

## Back to School

It is hard to believe that back to school time is already upon us! As you and your child get ready for the start of the school year, do not forget to ask the school if there are any forms that need to be completed by your child's medical provider. This includes a school medication administration form for enzymes, albuterol, or any other medications your child takes during the school day. If you are seeing your provider prior to school starting, please ask for a school medication form to be completed. If you will not be seen in clinic prior to school starting, please contact our office at 608-263-6420 option 2, and request that a form be completed and faxed to the school.

If your child is newly heading off to school, please ask us about our school resource folders. These folders provide useful information for schools and teachers, including information regarding nutrition, enzymes, general CF information, and the importance of access to sports drinks during gym and other physical activity. It is important to have a conversation with school staff at the beginning of the school year to discuss the above information available in the folder, so any questions can be addressed right away.

If your school has further questions, the Cystic Fibrosis Foundation has many great resources for schools and teachers to learn more about CF. You can find this information by going to: <https://www.cff.org/Life-With-CF/Daily-Life/CF-and-School/>

If any questions arise, please contact our office at 608-263-6420 option 2 to discuss further. *Have a great school year!*

## Behind the Curtain – Medication Prior Authorization

Some medications require prior authorization (PA) through the insurance company. Each insurance company has their preferred medications, their non-preferred medications, and specialty medications. Non-preferred medications and many specialty medications will require a PA.

Insurance companies may require you or your child to change from a non-preferred medication to a preferred medication and will not approve a PA until the preferred medication is trialed. Insurance companies can change their formularies frequently and so these preferred medications may change, requiring a PA or a change in medication again.

Typically, when an insurance company is requesting a PA the pharmacy is notified and will contact our office via fax or phone. When we receive this notification, we will review the electronic medical record to determine if alternative medications have been trialed in the past, if a PA has already been started, or if there was a PA completed in the past for this medication. If we determine a PA is truly needed, we will send a message on to our UW Health Prior Authorization Specialists to assist with the PA process.

Our PA specialists will complete the needed paperwork and fax to your provider for signature. Once the paperwork is signed, the PA specialist will gather any needed documentation from the chart to submit to insurance. Some insurance companies will require clinic notes, labs, or newborn screening results to approve medications. Certain insurance companies may require that this documentation be from within the last 3 months.

With commercial insurances, our PA specialist can submit the documentation directly to the insurance company. Wisconsin Medical Assistance requires that the pharmacy that will be dispensing the medication submit the PA. In these instances, our PA team will provide the completed paperwork to the pharmacy for submission.

Once a PA is submitted to insurance, the insurance typically allows themselves up to 30 days to make a determination. Once approved, our PA specialist will be notified and will contact the filling pharmacy. If the PA was submitted by the filling pharmacy, they will be notified by the insurance. You may receive a call from our PA specialists, the pharmacy, or a letter in the mail from your insurance company indicating the medication was approved. Often PAs are approved for 1 year at a time. Certain insurance may approve some medications for a lifetime. If you are newly starting a modulator therapy, some insurance companies may only approve a couple weeks at a time and may request more regular lab and visit documentation.

With more expensive therapies entering the market, such as modulator medications, insurance companies are closely watching refill history of these medications. Some insurance companies will start denying PAs for these medications if they feel the refill history is poor or will ask the care team to discuss at the next visit the importance of refilling and taking the medication as prescribed.

If you receive a denial from insurance for a medication, there are typically steps that can be taken to appeal the decision. Sometimes insurance companies will request a written Letter of Medical Necessity, or a “Peer to Peer” phone call from your provider. Insurance companies may also ask for you to appeal the decision.

### Steps you can take:

- Stay in contact with your pharmacy to check in on the PA process
- You can call your insurance company to check on PA process and if any documentation is required from you.
- If you have medications that are requiring yearly PAs, keep track of when those PAs will expire and reach out to the pharmacy or your care team to work on those renewals before you run out of medication.
- Make sure you are seeing your care team and obtaining labs in the recommended timeframes to reduce any delay in medication approvals.

We understand that prior authorizations can be frustrating and confusing. We are here to help you navigate this process and if there are questions that arise, please contact your care team to discuss further.

*-Your Peds Pulmonary Nursing Staff*

## Did You Know? AKA Silver Linings

*authored by our Parent Advisory Group*

Children with CF may qualify to attend one of the week-long summer Serious Fun Children's Network camps started by Paul Newman. Your whole family may also qualify to attend the spring or fall weekend family camps. The Midwest camps are North Star Reach in Ann Arbor, MI and Flying Horse Farms near Columbus, OH. It is no cost to campers while onsite, including food and housing. It is funded by the Newman's Own Foundation. They cater to all dietary needs, have strict infectious disease policies and will only allow one child with CF per camping session.

## SAVE THE DATE: CF Education Day October 19, 2019

We are excited this year to have Gunnar Esiason as our key note speaker! More details and registration information will follow.

## Good Bye and Well Wishes

We are sad to say that two of our nurses, Jennifer Harmelink and Erin Henter, have accepted new positions and will no longer be part of our pediatric CF care team. We wish them all the best and will miss them! We are recruiting for their replacements, but in the meantime, it might take us a little longer to get back to you when you call. Please be patient while we search, hire and train our new nurses... Jen and Erin leave behind some big shoes to fill!

## Flu Reminder

As fall in Wisconsin approaches, it is time to start thinking about getting your flu shot! The flu vaccine is available at UW Health clinics and pharmacies starting in September. If your insurance does not allow for you to receive the vaccine at our clinics, you can contact your Primary Care Provider's (PCP) office to ask about flu shot clinic dates, or to set-up a nurse visit. Many pharmacies also can give walk-in flu shots. *Wishing you a healthy fall and winter season!*

## Adult CF Center Inpatient Airway Clearance Changes

The UW Inpatient Respiratory Therapy Department will be modifying its airway clearance workflow starting on September 2nd, 2019. After reviewing practices at leading national CF centers, and partnering with the adult CF care team including Dr. Braun, we have developed a robust therapy plan for all patients admitted with a diagnosis of CF.

- The respiratory therapist will assist with ALL airway clearance therapy 4 times daily
- The respiratory therapist will be present to initiate and complete ALL therapy sessions
- Percussive therapy options: patients will have TWO primary choices:
  1. Patients will receive either VEST or Metaneb 4 times daily, OR
  2. Patients will receive either VEST or Metaneb 3 times daily and a 4th session of either:
- PEP
- Aerobika/Acapella
- Manual chest physiotherapy

We value the care our respiratory therapists provide and strive to see as much improvement as possible in your lung function during an inpatient stay. We understand that these changes may be a difficult transition for some but ask that our patients work with us as we strive to provide you with the best quality care. If you have any questions regarding these changes, please contact Kristin Stephenson, Respiratory Therapy Supervisor at 608-265-7908.

## Nutritional Information for the Administration of CF Modulating Drugs

As more people with cystic fibrosis become eligible for gene modifying drugs such as Kalydeco®, Orkambi®, and Symdeko®, it's nice to have information about how best to take these medications. It is important to take your prescribed medication with fat containing food (and enzymes if needed). Studies have shown absorption increases 2.5-4 fold. A general rule is to take the medications with 15-20 grams of fat, although a specific number has not been determined. If your child is using the granules, these may be mixed into a sweet food (to mask the bitter taste) and are stable in that food for up to an hour.

Below are some ideas of fat containing foods to try with your medications. The foods are labeled GF (Gluten Free), DF (Dairy Free), NF (Nut Free) to accommodate anyone with food allergies/intolerances. If you do not have any food allergies, simply make the appropriate substitution (ex: regular waffle, bagel, or pasta instead of gluten free or regular pasta or whole cow's milk yogurt instead of coconut milk). You can also combine different foods to get adequate fat.

Food	Grams of Fat
1 cup whole milk (GF, NF)	8 grams
1 carton So Delicious® Dairy Free Coconut yogurt (GF, NF, DF)	6 grams
Blueberry Muffin Lara Bar (GF, DF)	9 grams
10 corn tortilla chips with 2 Tbsp guacamole (GF, NF, DF)	10 grams
2 eggs (GF, NF, DF)	11 grams
3 slices bacon (GF, NF, DF)	11 grams
½ cup Udi's® gluten free granola (GF, DF) *only certain flavors are NF	11 grams
1 Tbsp Earth Balance® butter (GF, DF) *contains pea protein if allergic to legumes	11 grams
¼ cup hummus (GF, NF, DF) *chick peas usually tolerated with nut allergy	12 grams
2 Tbsp Nutella (GF)	12 grams
2 string cheese (GF, NF)	12 grams
1 Udi's® gluten free tortilla with ¼ cup melted cheese (GF, NF)	13 grams
1 Jack Links® beef stick (GF, NF, DF)	13 grams
2 pork sausage links (GF, NF, DF)	14 grams
½ avocado (GF, NF, DF)	15 grams
1 Van's® gluten free waffle with 1 Tbsp butter (GF, NF)	15 grams
1 Tbsp sunflower butter (GF, NF, DF)	15 grams
¼ cup sunflower seeds (GF, NF, DF)	16 grams
2 Tbsp peanut butter (GF, DF)	16 grams
1 Udi's® gluten free bagel with 2 Tbsp cream cheese (GF, NF)	16 grams
2 Jimmy Dean® pork sausage links (GF, NF, DF)	16 grams
¼ cup peanuts (GF, DF)	18 grams
GeeFree® Sausage, egg, and cheese pocket (GF, NF)	28 grams

## Teen Connections

The Cystic Fibrosis Foundation is creating opportunities for teenagers with CF, ages 13-18, to meet other teens their age, share stories, and talk about what's important to them. Beginning this summer, online group calls for teens will be hosted on a monthly basis, including separate groups for middle school and high school ages. Teen Groups will be social groups - not support groups - and will be facilitated by an adult with CF and a CF Foundation staff member.

### What to Share with Teens

Want to connect with other teens who have CF? The CF Foundation is starting monthly online groups for people with CF, age 13-18, to meet others your age, share stories, and connect about what's important to you. There will be separate groups for middle school and high school ages. Sign up for Teen Connections in order to receive information about these online groups and other future connection opportunities. Parental consent will be required for participants <18 years. [www.cff.org/TeenSignup](http://www.cff.org/TeenSignup)

For questions, please contact Aimee Jeffrey: [ajeffrey@cff.org](mailto:ajeffrey@cff.org).

*Aimee*

Aimee Jeffrey  
Senior Manager, Community Support  
Cystic Fibrosis Foundation

## Teen Connections FAQ

### What is Teen Connections?

The goal of Teen Connections is to provide opportunities for teenagers with CF (ages 13-18) to connect virtually with others their age, share stories, and socialize with someone who is in the same shoes.

### What are Teen Groups?

Over the next year, the CF Foundation will pilot monthly online groups for teens. These groups will be casual, virtual meetings where teens can socially interact. Teen Groups will be hosted on Blue Jeans or a similar platform so that participants can engage via audio and video. Calls will be held monthly and are open-ended; teens can join one or more calls and regular participation is not required.

### Who is moderating the Teen Groups?

An adult with CF and a CF Foundation staff member, trained in facilitation, will moderate each call. The facilitators' role is to create a positive environment and help enable conversations, not to direct or lead conversations.

### Will all ages of teens be mixed together?

No. Separate groups will be hosted for teens in middle school (age 13-15) and high school (age 15-18).

### Are Teen Groups support groups?

No. While teens may find support in connecting with their peers, the groups are intended to be social and are not for counseling of any type. Facilitators will establish group norms with all participants at the start of every call to ensure a common understanding of the group's purpose and boundaries.

### What if a teen is depressed or very negative during the group call?

Facilitators will be trained to redirect conversation if it veers toward the negative or towards mental health conversations. In addition, the facilitator can follow-up with the teen's parent if there is concern for the individual's safety or well-being.

### How do teens signup?

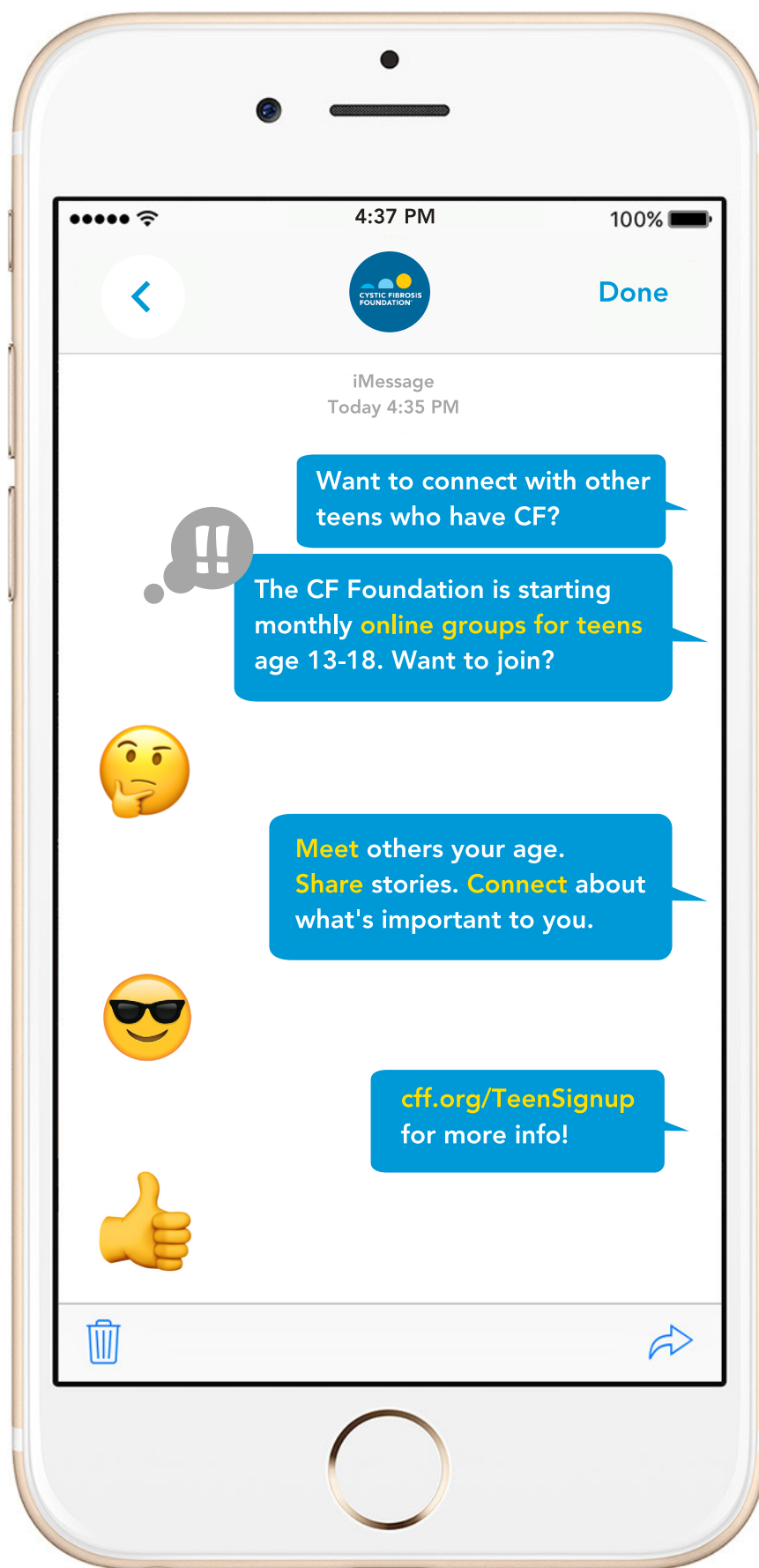
Teenagers with CF who are 13-18 years of age can sign up for Teen Connections at [cff.org/TeenSignup](http://cff.org/TeenSignup) (parent consent required for those <18 years). After signing up for Teen Connections, teens will receive information and invites to Teen Groups and any other connection opportunities in the future. It does not obligate anyone to participate in any online group.

### What other opportunities are available for teens?

Teen Groups are the first connection activity being piloted specifically for this age group. If there is interest among teens, additional connection opportunities may be added in the future. Online groups will be a great way for teens to share their ideas with us. In addition, teens age 16+ can request a mentor through CF Peer Connect and can attend some virtual events.

# TEEN CONNECTIONS

[cff.org/TeenSignup](http://cff.org/TeenSignup)

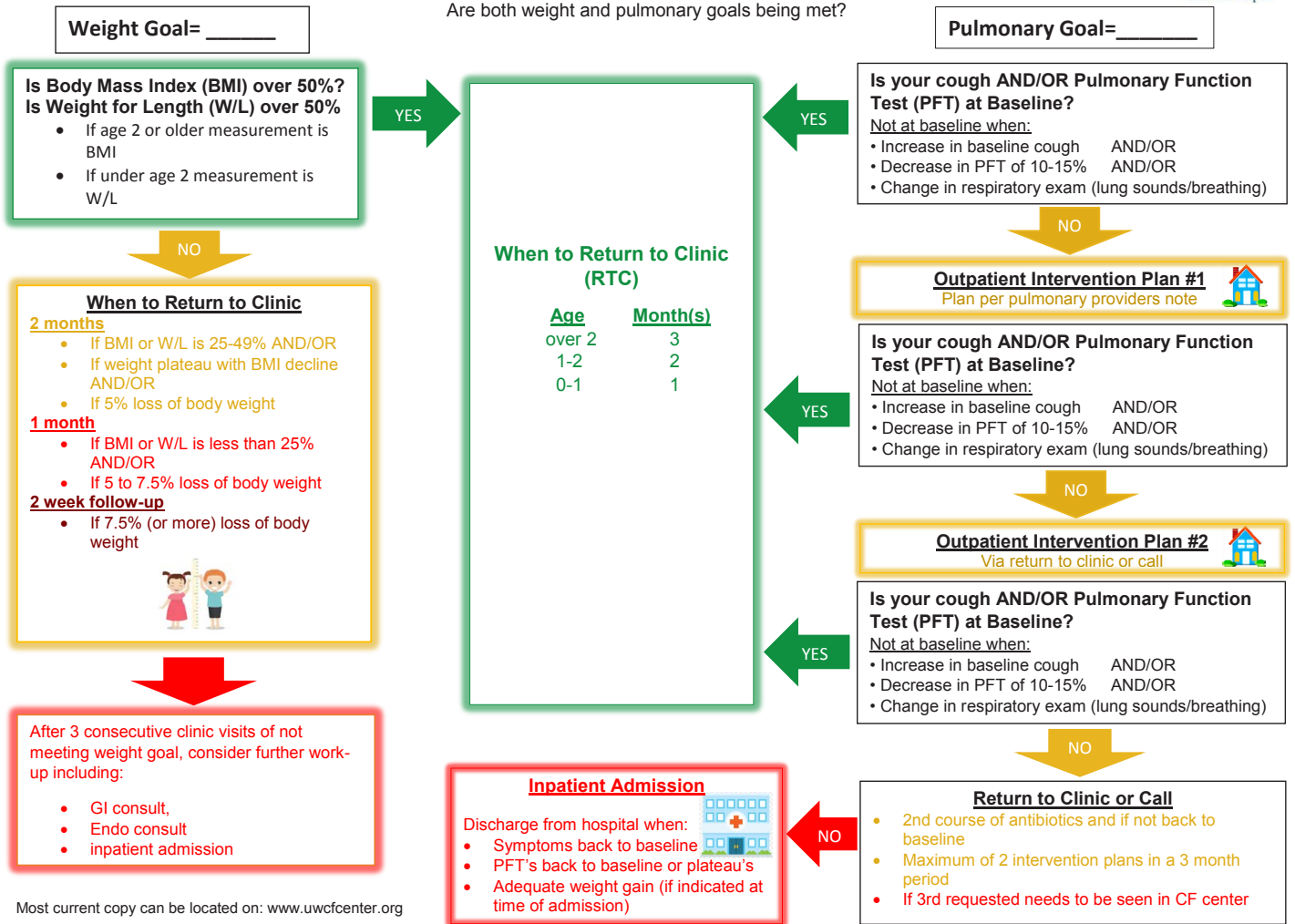




## Return to Clinic Guidelines for Pediatric CF Center

As a CF care team, we work together with patients and families to ensure you stay in optimal health. We have guidelines that we use as providers and recently collaborated with our parents and parent advisory group to have a family friendly version of our guidelines. Our feeling is that knowledge is power! We want you to know what guides us in our thinking and practice. Please see the attached return to clinic guidelines. You can also access them by going to our CF Center website.

### Return to Clinic Guidelines for Patients with Cystic Fibrosis



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