From the Desk of CF Medical Director Dr. Shatha Yousef:

Kids today have jam-packed schedules, often getting up early to catch the bus, participating in after school activities and, of course, doing homework. That means fitting daily CF therapies into the day can be a challenge. Winter months mean extra challenges to kids with CF. In addition to it being cold and flu season, colder temperatures may reduce helpful exercise activities — and a few extra calories to protect against the cold would be fantastic.

We know that without parental support and careful planning, CF children will not achieve their daily therapy that is needed to prevent lung infections and changes in lung capacity. Even if your child knows what to do, it’s important that you offer reminders and check-ins every day.

I also wanted to update you on two recent news items:

- In November, members of our Nemours CF care team attended the North American Cystic Fibrosis Conference (NACFC) in Indianapolis. In recognition of our collaboration with the national Cystic Fibrosis Foundation, we received the 2017 Outstanding Partnership Award! We are humbled by this honor and we’ll continue to maintain this award-winning relationship that makes such a difference to patients and families.

- We anticipate a new CFTR regulating medication will be approved in 2018. We will continue to track FDA approvals of these medications to determine if your child meets the indications for the new therapy. Read more in our “About CFTR Medication” article. These therapies hold the promise of advancing future treatments for CF and we will continue our journey together with you until there is a cure.

As always, we are committed to supporting you and your child.

All the best for a happy and healthy 2018,

Dr. Yousef

Always Plan Ahead

Hurricane Irma was devastating to our area, and it meant challenges for many CF families. Here are some important “lessons learned” from Irma and reminders, so you and your family will be well-prepared for any future hurricane or other unforeseen event:

- **Sign up in advance for the medical shelters in your area.** Visit [www.Floridaisaster.org](http://www.Floridaisaster.org) to find the medical shelter in your area and information on how to sign up.

- **Keep your prescriptions filled.** If a storm is predicted, refill your daily medication as soon as possible since unforeseen events can delay vital medications.

- **Refresh your manual chest PT skills.** Remember, when you lose power, you can always do manual chest physical therapy for airway clearance. CPT is easy to do. For a child, CPT can be done by anyone, including parents, siblings and even friends. Visit the CFF Airway Clearance section at [www.cff.org](http://www.cff.org) to refresh your CPT skills.
There's Still Time to Get Vaccinated for the Flu

We strongly recommend all of our CF patients and their families get a flu vaccine every year. Note that timing is everything when it comes to the flu vaccine. That's because it takes about two weeks for antibodies to develop and provide protection against the influenza virus. It's ideal to get vaccinated early in the fall — before the flu season really gets underway — but there's still time! This season, the CDC recommends injectable flu shots, not nasal sprays. Read what the Cystic Fibrosis Foundation has to say about CF and the flu: https://www.cff.org/Life-With-CF/Daily-Life/Germs-and-Staying-Healthy/What-Are-Germs/The-Flu/.

CF Research Opportunities

Your child may be eligible to participate in one of the following trials in 2018:

- **Savara (AeroVanc) Study**: The Savara (AeroVanc) study is officially called the AVAIL study. For more, visit AVAILstudy.com.
- **TEACH Study**: Testing the effect of adding oral azithromycin to inhaled tobramycin in people with CF (TEACH-IP-15). This study will look at the effect of adding oral azithromycin to inhaled tobramycin. (Ages 12 and older.)
- **Vertex VX-440-101 Study**: (Vertex VX-440-101) Phase 2 study of VX-440 combination drug in people with cystic fibrosis. This study will look at the safety and effectiveness of the drug VX-440 when combined with ivacaftor and/or tezacaftor (VX-661). (Ages 12 and older.)
- **Savara SAV005-02 Study**: (Savara SAV005-02) AeroVanc for the treatment of MRSA in people with CF. This study examines the safety and effectiveness of AeroVanc in the treatment of persistent MRSA lung infections in people with CF. (Ages 12 and older.)

For more information about these trials, contact Amanda Darling, clinical research coordinator for Nemours, at (407) 650-7966 or Amanda.Darling@nemours.org. For additional CF-related clinical trials, visit the Cystic Fibrosis Foundation’s clinical trial finder.

New CF Videos Available

We teamed up with the Orlando Cystic Fibrosis Foundation chapter and produced a series of videos on how to live with CF. The videos provide information and helpful tips on topics such as:

- heading back to school with CF
- improving nutrition
- infection control
- current studies and CF research
- CF and school socialization

Click here to watch the videos now.

Additionally, Genentech released a video on how CF medications work on the lungs. We encourage you to watch it and continue to build your knowledge about CF. Watch the video.
10 Do’s and Don’ts for Managing Mealtimes
Here are 10 do’s and don’ts for making mealtime less stressful and more enjoyable, especially when siblings are involved.

**DO**

1. **DO explain why servings can’t always be equal.** If a sibling complains that portion sizes are unequal, use simple words and phrases to explain why this is happening. For example: “Your brother has CF. He needs more food than we do in order to grow and play and be happy.”

2. **DO remind siblings that you care for them, too.** Siblings need reassurance that you love them. Whenever you explain why your child with CF needs more or special foods, make sure to also remind your child who does not have CF of special things you do for them.

3. **DO give your child choices.** Whenever possible, offer choices when it comes to foods. However, two are usually fine, as more than that can be overwhelming to a child.

4. **DO include your child in meal planning and cooking.** It can help him become more interested in the meal and more willing to eat a variety of foods.

5. **DO praise siblings after a good meal experience.** Also, point out times when they made efforts to share foods or portions with their brother or sister with CF.

**DON’T**

6. **DON’T add “extras” at the dinner table.** Before serving food or calling your family to the dinner table, add butter, cream or coconut oil to your child’s food. Adding calories when you are alone is not being sneaky, it’s being smart and respectful of your entire family, including your child with CF, so everyone can enjoy a pleasant meal together.

7. **DON’T stop offering new foods.** Studies show that a child may need to be exposed to a food 10–15 times before they are willing to try it.

8. **DON’T call your child a “picky” eater.** Your child with CF may overhear you and decide they like being known as a picky eater.

9. **DON’T draw attention to your child’s eating.** Instead, compliment your child with CF when she finishes a meal or tries a new food.

10. **DON’T fulfill special requests or immediately give snacks after uneaten meals.** Remember that calories are important, but healthy eating habits and a consistent meal schedule are important, too.

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**Add Calories to Everyday Foods**

Try adding some of the following high-calorie items to your child’s foods:

- avocado
- bacon bits
- breading (good on meats)
- brown sugar
- canned french-fried onion rings
- caramel dip
- Carnation Breakfast Essentials® powder
- coconut or olive oil
- cream cheese
- heavy cream or half-and-half
- honey
- jam
- mayonnaise
- melted or shredded cheese
- Nutella®
- nuts, such as walnuts or pecans
- peanut butter (or other nut butters)
- powdered sugar
- ranch or cream-based salad dressings

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This article was adapted with permission from Karen Maguiness, board-certified specialist in pediatric nutrition, specializing in the nutritional care of patients with cystic fibrosis. As a dietitian/nutrition leader at the Cystic Fibrosis Foundation, Maguiness participated in developing national clinical care guidelines, nutrition education materials and research that documented improved pulmonary outcomes. She has also received regional and national awards for her leadership in the field of nutrition and cystic fibrosis.

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**What to Expect When Your Teen Goes to Nemours Children’s Hospital**

When your teen is admitted to Nemours Children’s Hospital for CF care, your child will have four treatments per day, which are delivered by one of our respiratory therapists. In order to accommodate meal times and other medical testing or therapy, sessions are scheduled for 11 a.m., 3 p.m., 8 p.m. and midnight.

At Nemours, a vest is our first choice for airway clearance therapy, but we can also use MetaNeb®, intrapulmonary percussive ventilation (IPV) and Acapella® if the vest isn’t working well. You can help us understand what’s best for your teen by letting us know what has worked in the past.

You are welcome to bring your home airway clearance device if it’s more comfortable, but your device will require an electrical check.

During these sessions, the respiratory therapist will encourage your child to produce an effective “huff” cough. This will help mobilize the secretions that are being shaken loose.

PARI reusable nebulizers (a drug delivery device that administers medication in the form of a mist inhaled into the lungs) are changed by the respiratory therapist or a clinician every 24 hours to help prevent infection.

If your teen has been doing lung function testing, a clinician from our pulmonary lab will likely come to your room to do pulmonary function tests (PFTs) to measure how well your child’s lungs are working and evaluate progress.
About CFTR Medication

Cystic fibrosis transmembrane conductance regulator (CFTR) modulator therapies are designed to correct the function of the defective protein made by the CF gene. As different mutations cause different defects in the protein, the medications that have been developed so far are effective only in people with specific mutations. There are currently two FDA-approved CFTR modulators:

1. ivacaftor (Kalydeco®)
2. lumacaftor/ivacaftor (Orkambi®)

How to Use CFTR Medications

If your child is on a CFTR medication, here are some things you need to keep in mind:

- The medication helps produce thinner mucus, but is not a cure.
- Your child must continue regular airway clearance and aerosol therapy.
- If you skip doses or stop taking the medication, the effects on your child's lungs will stop.
- The effects of CFTR modulators only last for as long as the medication is in your child's system. Therefore, your child needs to take the medication every 12 hours, or as your care team recommends.
- CFTR modulator medications should be taken with fat containing foods, so the medication can be absorbed properly. Talk to your CF Care Team about fat dosing recommendations.

How Do CFTR Medications Work?

The CFTR protein regulates the proper flow of fluids and sodium (salt) in and out of cell linings in the lungs and other organs. In children with CF, mutations in the CF gene cause the protein to malfunction or to not be produced at all. This causes lung mucus to be thick and sticky, which can lead to infection and lung damage.

Note: We anticipate a new CFTR regulating medication will be approved in 2018. We will continue to track FDA approval of this medication to determine if your child meets the indications for this new therapy.

To learn more about CFTR, go to the Cystic Fibrosis Foundation website at www.cff.org and view Treatments and Therapies.