From the Desk of CF Medical Director Shatha Yousef, MD:

We’ve had a great spring! It included a site visit from the Cystic Fibrosis Foundation, which granted the center continuous accreditation as we grow our team and our patient population. We also had our newborn screen site visit — the team got accolades for all their effort and success with our newborn screening process.

As we stroll into summer, we encourage our families to discuss vacation plans with the team. Whether you’re traveling or planning local activities that are not part of your regular routine, let us know how we can help in assessment, preparation and general considerations.

Our hot Florida climate is great for outdoor activities, but keep in mind that preventing dehydration is key for having a healthy summer. Be sure to read about hydration in our summer tips and then follow the recommendations.

We continue to expand our CF research studies. I encourage you to take a minute to look at our list of studies — consider participating if you qualify and if it works with your summer schedule.

Wishing a wonderful healthy summer to all our CF kids and families!

— Dr. Shatha Yousef

The Experience of Care Survey: Your Voice Can Make a Difference

The CF care team here at Nemours works hard to improve health outcomes for all the patients we care for. The Experience of Care Survey, a CF Foundation initiative, has helped us do just that, helping us gain insight that can improve the quality of your experience in the CF clinic.

How does it work?

- You will be contacted shortly after your CF clinic visit and asked to provide direct feedback via survey. (You will not be surveyed after every visit, only twice per year.)
- There are several options for completing the survey. You can call in to an automated survey or fill it out online. If you do not choose either of those options, you will be contacted to complete the survey by phone.
- All information is confidential, so please be honest! We want to hear the good, the bad and the ugly. It is always our goal to do our very best for our patients and families, so if we are not meeting this standard we need to know!
- Once the survey is complete, this valuable data is then tabulated and distributed back to the CF Program to help inform our quality improvement work.
ATTENTION:
ANNUAL LABS
Summer is the PERFECT time to complete your annual labs, OGTT and chest X-ray!

- CF care can be time consuming!
  Consider getting these tests done over the summer to minimize disruptions during the school year.

- The CF Foundation recommends yearly monitoring for complications of cystic fibrosis. This includes blood work and a chest X-ray for all patients, and an oral glucose tolerance test for those ages 10 and up.

- These tests are extremely important. They help your CF team identify complications early so they can be addressed.

- If you have not completed your annual testing for the year, please contact CF clinic staff. We will be happy to provide orders to get it done!

EQUIPMENT CHECKS
- One of our respiratory therapists will be reaching out to you at least once a year with a reminder to bring your equipment to the clinic to be checked. These check-ins are great opportunities to make sure your equipment is working properly and to address any issues as quickly as possible.

Summer Tips
School is out and the weather is heating up. Your CF team wants you to enjoy your summer! However, some things require special attention for people with CF.

Keep these tips in mind as the temperature rises:

HYDRATION | People with CF always need to drink plenty of fluids. This becomes especially important during the hot, humid months of summer.

Encourage kids not to wait until they feel thirsty. Offer them plenty of water throughout the day. (Caffeinated drinks don’t count — they actually work as diuretics and make you lose more fluid through urination.)

Make sure kids drink before, during and after exercise. Sweat = fluid loss.

Babies need extra fluids, too. Infants can get dehydrated even more quickly than young children. Be sure to keep them hydrated and dress them in clothing that will keep them cool.

SALT | Individuals with cystic fibrosis lose more salt (sodium chloride) in their sweat than those without CF. In the summer months and during exercise, when we sweat more, those with CF are at increased risk of becoming hyponatremic, which means to have a low sodium level in the blood.

Know the signs. Symptoms of hyponatremia include nausea, vomiting, headache, muscle weakness and cramping. It’s important to pay attention to salt intake and to any symptoms.

Use more salt. You can increase the amount of table salt you use, have some salty snacks, and read food labels to choose those with a higher salt content. If kids are exercising, you may want to add 1/8 teaspoon of salt to their 12-ounce sports drinks.

MEDICATIONS | Check the temperature. Some medications will need to be refrigerated while others are fine at room temperature.

Enzymes, for example, need to be kept at room temperature, 59°–86°F. You should not keep some medications in your car or in direct sunlight, as that will cause them to be less effective.

If you’re not sure about the temperature sensitivity of each medication, check with your CF team. You might need to keep medication in a small cooler with an ice pack if you’re going to be out and about during the day.

Enjoy your summer, but don’t take a break from paying attention to your fluids, salt and proper medication storage!

TRAVEL | Check in with your CF team before your trip.

Know your care options. Find out the phone numbers and addresses of the pharmacies, hospitals and urgent care centers at your destination. Bring the information with you.

Know your air travel rights. Additional assistance is available for travelers with medical conditions — it’s the law.

Check out cff.org/Life-With-CF/Daily-Life/Traveling-With-CF/Preparing-to-Travel/ for more information on how to prepare for travel.
Cystic Fibrosis Foundation Events

We always like to keep you up to date on all the events the foundation has planned for the next few months. These events are a fantastic way to engage with the community, raise awareness, and have fun. You can learn more about these events on the foundation’s website. cff.org/CentralFL/

- **Pipeline to a Cure: July 14, New Smyrna Beach**
  This is an annual surf event helping those in the CF community learn how to surf or paddle board, promoting the positive effect that surfing has on the health of children and adults with cystic fibrosis. For more information on this event, visit pipelinetoacurefl.org/

- **Orlando’s Finest Event: August 16, 6:30 p.m., location TBD**
  This event celebrates young professionals in the Central Florida area.

- **CF Climb: September 29, 10:00 a.m., Orlando World Center Marriott**
  512 steps! 28 floors! Participants climb the stairs of the hotel to post their best time. Some years there are also skill competitions between local firefighters. This event is $25 for registration with a minimum fundraising amount of $75.

- **Cycle for Life: November 11, 8:00 a.m., “Artisan Alley,” DeLand**
  Bikers ride a variety of distances throughout Volusia County, with rest stops along the way for refueling and cheering. This event is $35 for registration with a minimum fundraising amount of $150.

- **Greg Warmoth Holiday Classic: December 3, 8:00 a.m., Ritz-Carlton Orlando, Grande Lakes**
  WFTV’s Greg Warmoth hosts a day of golf and fun.

- **Holiday Home Tour: December 12, 11:00 a.m., Alfond Inn, Winter Park**
  Enjoy some holiday cheer with a tour of some beautiful homes in Winter Park. This day also includes a luncheon and live and silent auctions.

2018 CF Great Strides Lake Nona

On May 5, our CF team participated in the Great Strides walk in Lake Nona at Moss Park. Everyone had a fantastic time, and we were so happy to see some of our families present. The Nemours team was in full force and this year our team raised $1,610 for the foundation! Dr. Livingston gave a wonderful talk on advancements in treatments, as well as on the continued support and research to find a cure. We look forward to next year’s walk and hope to see you there!

Jessica Winn, RRT, RPFT, accepts an award from Paul Gloersen, president of the Orlando CFF chapter, and Dr. Floyd Livingston for her commitment and work with CFF.

The Nemours CF Walk Team in their now-famous “Let’s Taco About CF” T-shirts.
Nemours CF Research Opportunities

Currently Enrolling Studies

- **A Long-Term Prospective Observational Study of the Incidence of Fibrosing Colonopathy Cystic Fibrosis Foundation Patient Registry**: This observational study assesses the occurrence and risk factors for a rare bowel disorder called fibrosing colonopathy in patients with CF ages 12 years and older.

- **TEACH Study**: This study looks at the effects of adding oral azithromycin to inhaled tobramycin in patients with CF ages 12 years and older.

- **AVAIL Study**: This study looks at the effectiveness of inhaled vancomycin (AeroVanc) for the treatment of persistent MRSA lung infection in CF patients ages 6 years and older.

- **VX15-770-126**: This study evaluates the safety and effectiveness of long-term ivacaftor (Kalydeco) in patients with CF less than 24 months of age.

- **CHEC Study**: This study looks at changes in sweat chloride levels and their association with clinical outcomes in CF patients 4 months and older who are taking CFTR modulators.

UPCOMING Next Gen Vertex Studies

- **VX17-445-102**: Evaluates the efficacy and safety of VX-445 in triple combination (TC) with tezacaftor and ivacaftor, in patients with CF ages 12 years and older who have one copy of the F508del mutation and a minimal function mutation (F/MF).

- **VX17-445-103**: Evaluates the efficacy and safety of VX-445 in triple combination (TC) with tezacaftor and ivacaftor, in patients with CF ages 12 years and older who have two copies of the F508del mutation.

- **VX17-445-105**: Evaluates the safety and efficacy of VX-445 in triple combination (TC) with tezacaftor and ivacaftor, in patients with CF ages 12 years and older who have one copy of the F508del mutation.

Our CF care team values your thoughts and feedback on how we can best meet your CF care needs. If you have any questions or concerns about the publication or if you have ideas or topics for future articles, please do not hesitate to let a member of your CF care team know. You can also email us at cffeedback@nemours.org.