Nemours CF Team Staff Updates

If you’ve visited recently, you may have noticed the absence of a familiar face; Suzanne Sheres was part of the CF team for nine years. Recently, she and her husband relocated to Atlanta to be closer to their adult children and extended family. Suzanne’s now the clinical dietitian for Option Care Home Infusion Services, where she continues to advocate for the nutritional needs of kids with complex medical needs. We’ve included in this newsletter a message Suzanne wanted to share with you that outlines how being open about eating struggles can help your child stay healthy.

With change comes the opportunity for growth. We’re happy to have Kaitlyn Horinko, RD, as our new clinical dietitian. Kaitlyn joined Nemours in June 2014 as an inpatient hospital dietitian. Prior to Nemours, she was a hospital dietitian in North Carolina for three years, working with both kids and adults. Kaitlyn is working toward completing her Master of Science in Nutrition. Originally from Pennsylvania, she now resides with her husband, dog and cat in downtown Orlando. In her spare time, Kaitlyn enjoys boating, running, exploring different areas of her new home state and traveling to see family and friends.

We’re also pleased to welcome Lindsey Granger, ARNP, to the Nemours pulmonology team. Lindsey is a pediatric nurse practitioner who comes to us from the Children’s Healthcare of Atlanta Cystic Fibrosis Center. She’s passionate about caring for kids with CF, and looks forward to meeting all of our great patients and families. In her personal time, Lindsey enjoys spending time at the beach with her husband and two small children.

Shatha Yousef, MD, is a pulmonologist who joined Nemours Children’s Hospital in November 2015. She earned her medical degree at Jordan University of Science and Technology in Irbid, Jordan. She completed a residency in pediatrics at King Hussein Cancer Center in Amman, Jordan. After completing a Michigan State University residency in pediatrics at the Hurley Medical Center in Flint, Mich., she specialized in pediatric pulmonary medicine with a fellowship at the University of Miami Miller School of Medicine program at Jackson Memorial Hospital, with an additional year of fellowship dedicated to clinical research. Dr. Yousef is certified by the American Board of Pediatrics.

Tarig Ali-Dinar, MD, joined the Division of Pulmonology at Nemours Children’s Hospital in November 2015. Prior to joining Nemours, Dr. Ali-Dinar was an assistant professor and associate director of pediatric pulmonology, and the director of the pediatric sleep center at the University of Miami Miller School of Medicine. Dr. Ali-Dinar earned his medical degree from the University of Szeged Faculty of Medicine at the Albert Szent-Györgyi Clinical Centre in Szeged, Hungary. After completing a residency in pediatrics at Michigan State University’s Hurley Medical Center, he specialized in pediatric pulmonology with a fellowship in the University of Miami Miller School of Medicine program at Jackson Memorial Hospital in Miami. Dr. Ali-Dinar continued his training with a fellowship in pediatric sleep medicine at Harvard Medical School’s Boston Children’s Hospital. Dr. Ali-Dinar is certified by the American Board of Pediatrics and the American Board of Sleep Medicine. He speaks English, Arabic and Hungarian.
Nemours CF Program Awarded The Fundamentals, a Learning and Leadership Collaborative Grant

The Nemours CF program was awarded a grant from the Cystic Fibrosis Foundation called The Fundamentals, a Learning and Leadership Collaborative — or Fun LLC for short. Led by health care professionals, this quality improvement initiative will connect CF programs for 16 months to gain fundamental improvement in knowledge, skills and practice for better care and outcomes for patients across the country.

The CF team has been busy preparing for our Fun LLC, which we call REACT (Re-Education in Airway Clearance Techniques). As part of our initiative, we will ask you to complete a short survey about your child’s current home therapies. We’ll also ask you to bring all of your airway clearance equipment (vest devices, acapella, IPV, nebulizer machines, etc.) to one of your appointments so we can review therapy goals and techniques with your family, and make adjustments as needed (like making it easier to fit therapies into your daily schedule!).

Did You Say “Research”?

Did you know that Nemours Children’s Hospital is involved in cystic fibrosis research? The CF research team, in partnership with your child’s doctors to identify studies your child may qualify for. Many of these studies are supported by the CF Foundation and are used to identify, test, evaluate and improve new CF treatments. In addition, outcomes are used to shape the development of best practice guidelines for the CF care team and your doctors, and most importantly, trial medications have resulted in improved outcomes, which translate to improved quality of life.

Currently, Nemours is participating in one of the most cutting-edge research studies with Vertex Pharmaceuticals. This study is exploring the combination of Kalydeco™, a recently approved FDA drug for CF, with another drug to yet again raise the bar on CF outcomes.

Current CF Studies at Nemours:

A Phase 3, Double-Blind, Placebo-Controlled, Parallel-Group Study to Evaluate the Efficacy and Safety of Lumacaftor in Combination with Ivacaftor in Subjects Aged 6 Through 11 Years With Cystic Fibrosis, Homozygous for the F508del-CFTR Mutation. Enrollment started in November 2015; limited number of available slots available.

Fibrosing Colonopathy (FC) is a Long-Term Prospective Observational Safety Study of the Incidence of and Risk Factors for Fibrosing Colonopathy in U.S. Patients with Cystic Fibrosis Treated with Pancreatic Enzyme Replacement Therapy. Eligible participants must be enrolled in the CF registry and have an FC diagnosis. (Enrollment Closed) AIR-CF 5, A Prospective, 5-year Registry Study to Monitor the Susceptibility to Aztreonam of Pseudomonas Aeruginosa (PA). Final year of data collection is 2016.

Upcoming CF Studies at Nemours:

A Phase 3, Rollover Study to Evaluate the Safety and Efficacy of Long-term Treatment With Lumacaftor in Combination With Ivacaftor in Subjects Aged 6 Years and Older With Cystic Fibrosis, Homozygous for the F508del CFTR Mutation.

Upcoming CF Studies at Nemours:

A Phase 3, Randomized, Double-Blind, Placebo-Controlled, Crossover Study to Evaluate the Efficacy and Safety of Ivacaftor and VX-661 in Combination With Ivacaftor in Subjects Aged 12 Years and Older With Cystic Fibrosis, Heterozygous for the F508del-CFTR Mutation, and a Second Allele With a CFTR Mutation Predicted to Have Residual Function.

“TEACH Trial: Testing the Effect of Adding Chronic Azithromycin to inhaled Tobramycin. A randomized, placebo-controlled, double-blinded trial of azithromycin 500mg thrice weekly in combination with inhaled tobramycin.” This study will be reviewed by the TDN PRC.

For questions in regard to Nemours CF studies, please contact Omar Oquendo at (407) 650-7880 or omar.oquendo@nemours.org.
The Importance of Nutrition in CF Treatment
by Suzanne Sheres, MMSc, RD, LD/N, CDE

If you’re like many parents trying to keep up with various treatments and medicines your child needs, nutrition therapy often falls to the bottom of your long “to-do” list. We understand that adhering to a CF plan takes a toll on the whole family, and you might feel like your child goes through enough with this disease that controlling what, when and how much food he or she eats is a little unfair.

Good nutrition is important for your child to maintain healthy growth and development, and to support the body’s natural defense against infections and lung problems. In short, nutrition therapy can be life-saving. In fact studies show that weight and lung health are connected, and that kids with a BMI (body mass index) in the 50th percentile or higher have the highest FEV1 (expiratory volume) values. That’s why we want you to know we’re here to help.

Talk Openly to Your Child’s Care Team
At your Nemours CF center, you have access to an entire team of health care professionals, including doctors, nurses, dietitians, respiratory therapists and social workers who are here to support you and your child. We empathize with your situation and hope that we can have frank, honest conversations about your child’s eating struggles and adherence so we can better serve you. Please let us know if:

- your child doesn’t like, tolerate or take prescribed supplements
- regularly misses enzymes or vitamins
- your child’s food choices have changed
- your child’s appetite is poor
- the family budget is affecting nutrition care

We’re partners in your child’s health care, and when we know what’s really happening, we can develop an alternative plan to overcome specific problems, no matter what they are. Keeping your child healthy is our top priority.

For more information:
When Tube Feeding is an Option

Generally, children with CF need more calories than their peers to maintain optimal health, but that's sometimes easier said than done! Some children can keep a healthy weight with a good meal and snack plan that includes nutrient-dense, calorie-rich foods and pancreatic enzymes (to help digest foods, especially fat). Others, despite their best effort, still have a difficult time taking in enough calories. If weight gain is a continuous struggle for your child, talk to your care team about tube feeding.

Tube feeding is typically prescribed when dietary interventions, calorie-boosting strategies and enzymes aren’t helping a child gain weight. It’s also a good option for children with infections, breathing problems and poor nutrient absorption, which all factor into the need for more calories.

Benefits of Tube Feeding
Feeding tubes offer the possibility of weight gain, and they can certainly reduce stress around meals. But the extra calories from tube feeds can have additional positive effects, including their ability to:

- increase lung function
- enhance the ability to fight infection
- promote healthy growth
- improve energy
- maximize self-image

Types of Feeding Tubes
Gastrostomy tubes (G-tubes) and nasogastric tubes (NG-tubes) are the most common types of tube feeds.

A G-tube is inserted directly into the stomach with a small valve that looks similar to the valve on a beach ball. Tube insertion is not complicated and may require just a short hospital stay. An NG-tube is a long, thin tube that’s inserted through the nose and into the stomach.

Both G-tubes and NG-tubes can deliver liquid supplements (such as Pediasure® or Boost®) at night while your child is sleeping or during the day as a high-calorie snack. Enzymes can also be delivered through the tube if your child needs them for nutrient absorption.

We want you to view tube feeding as part of the overall cystic fibrosis treatment plan, and not as a failure or because your child isn’t a good eater. Feeding tubes will help your child stay healthy and thrive.
In the Words of Our Families

Dinnertime was constantly stressful for our family. My son had always been a picky eater and getting him to eat enough was a task. He never really cared for fruits and vegetables, and while my husband and I tried different tricks and techniques, nothing worked. We did everything to encourage him to eat high-calorie foods — even bribes (“Eat your pasta. You’ll get ice cream after two more bites!”) — to put on a few precious pounds before the next judgment day: The weigh-in at the doctor’s office.

At one appointment, my son gained two (hard-fought) pounds and we were ecstatic! You can imagine our surprise when the doctor then suggested we consider tube feeding to improve his overall health. My heart stopped and tears welled up in my eyes. I remember thinking, “Hold it in, don’t let my boy see me, he can’t see that I’m scared … Is this when his health starts to decline?” I thought we’d been doing so good and here, in my mind at the time, I’ve failed to nourish my own child (what kind of mother am I?)!

To help my son gain weight, the doctor recommended a gastrostomy tube (or G-tube). I was of course nervous about what this might mean, and I had many questions. Like many moms before me, I put my brave face on and began the journey for answers. Here’s what I learned.

This isn’t my fault — or his fault. It simply is what it is.
Some people with CF have a hard time gaining weight and no matter what they do, they can’t keep up with the amount of calories they need. It doesn’t mean I have failed as a mother, or that my child is a bad eater.

Tube feeding doesn’t mean my child’s health is declining.
On the contrary, tube feeding keeps my child healthy by strengthening his immune system. Extra weight and nourishment helps the body tremendously when fighting off infection or illness, and he needed this advantage.

G-tube placement is more common than I thought.
The procedure, placing a mini-button in the belly, was fairly simple. I was worried it would impact my son’s activity, but he’s free to play sports, swim and just be a normal kid.

Adjusting to anything new takes a little time.
After the surgery, it was a bit nerve-wracking learning how to use the device. But, just like every family who has a child with extra needs, we quickly gained confidence and adjusted to this new routine.

Tube feeding has given my son a better fighting chance against this horrible disease that threatens his every breath. Now, two years since the G-tube placement, he’s an eight-year-old in the 75th percentile for weight, and his lung function topped out at his max ever with PFT scores of 107 percent. My son’s overall health has been good, and we’ve discovered how to make taking medications easier by delivering liquid forms through the G-tube whenever possible.

Every little bit helps. And we know we made the right choice for our son.
We Invite You to Join the Nemours CF Family Collaborative

We believe that being involved helps improve the quality of life for all our CF patients and families, which is why we’re inviting parents and patients 18 and older to join our new CF Family Collaborative. The collaborative will foster feedback and idea-sharing to help us better our program initiatives and quality improvement processes.

If you’re interested, please contact Amanda Montgomery at Amanda.Montgomery@Nemours.org.

MyNemours: An Online Tool for Health Communication

At Nemours, one of our goals is to improve the delivery and coordination of health care to all children. As part of this effort, we’ve developed MyNemours, a confidential, easy-to-use, Internet-based tool that gives parents and legal guardians secure electronic access to select portions of their child’s medical records. With MyNemours you can view medical records, labs and test results. You can also communicate with your doctor and request appointments and prescriptions. Best of all, MyNemours can be used from the comfort of your own home.

Stop by the registration desk for more information or visit the link at www.Nemours.org/MyNemours.

Nemours CF Team Wins Spirit Award at the 2015 Great Strides Walk

The walk was held at University of Central Florida. And our team picture shows there’s no question Nemours has spirit!