CF Care at Nemours
Nemours Children’s Clinic, Pensacola | Your child. Our promise.

MyNemours: Online Tool For Patients & Families

At Nemours, one of our goals is to improve the delivery and coordination of health care to children. As part of this effort, Nemours has developed MyNemours, a confidential, easy-to-use, internet-based tool that gives parents and guardians secure electronic access to select portions of their child’s health records. Best of all, MyNemours can be used from the comfort of your own home.

With MyNemours online tool, you can view some of the health records and lab tests. Patients and their families can communicate with their doctor and request appointments and prescription refills. Stop by the registration desk for more information or visit the MyNemours link at: www.nemours.org/mynemours.

Director’s Corner
Okan Eldemir, MD, Division Chief & Pediatric Pulmonologist

Hello everyone! It gives me great excitement to write my very first corner in our brand new CF newsletter. These are extraordinary days with a lot of great news coming out for our CF community. In this section, I am going to try to share new developments and new medications with you.

This first corner is dedicated to Ivacaftor (Kalydeco). Many of you might already have heard about this revolutionary drug. It became FDA approved last year and is a pill taken by mouth and is effective in only a small group of CF patients who carry the mutation G551D. This mutation is found in only 2% of all CF patients. G551D mutation causes the chloride channel responsible for CF disease to stay closed or locked. Ivacaftor works like a key and opens it up so the channel can function normally. As a result, the patient’s lung function improves, the infection frequency decreases, the nutrition improves and even the sweat test normalizes.

The same company who developed Ivacaftor is now working on another product for patients with Delta-F508 mutation. This new medicine is called VX-809 in research studies, and we expect to hear some preliminary results within the next few years. I will write about VX-809 in more detail in the next newsletter.

Arikase and MP-376 are two new inhaled antibiotics waiting for FDA approval. The first one is made from amikacin and the second from levofloxacin. A dry powder inhalation form of tobramycin is also currently being studied. My final update on new medications is regarding hypertonic saline. A study performed in infants and young children with cystic fibrosis showed very small benefit from the use of this medicine. This does not mean that it should not be used, but we should know that this is not the miracle maker we were hoping it would be. I will give you the latest update on all these and other new medications in the next newsletter.

CF Assistance Program
Need Help with Co-Pays?

The Cystic Fibrosis Patient Assistance Foundation (CFPAF) is a non-profit organization created to help lessen the financial burden costly medications have on CF patients and their families. It was started in response to the high cost of CF medications, and it is the only national patient assistance program designed specifically for the CF community. The CFPAF can work with private insurances to help cover expensive prescriptions co-pays. The program is designed to help cover FDA-approved inhaled medications (like Pulmozyme and TOBI). If you are taking other medications they may also be able to help you find other resources that offer assistance.

To learn more about the CFPAF or to see if you qualify for assistance, please call (888) 315-4154 or go to http://www.cfpaf.org.
Taking Great Strides for CF

Great Strides is largest national fundraising event for the Cystic Fibrosis Foundation (CFF). Tens of thousands of co-workers, friends and families come together each year as one community for one cause...to help find a cure for CF. Last year, nearly $40 million was raised to fund life-saving research, quality care, and education programs. Real progress toward a cure has been made, but the lives of young people with CF are still cut far too short. We urgently need the public’s continued support to fulfill our mission and help extend the lives of those with the disease.

**Pensacola**

Seville Square .......................................................... May 25, 8:00 a.m.

**Crestview - Race for Lace**

Main Street Medical ..................................................... Nov 2, 2013

**Panama City**

Aaron Besant Park (adjacent to Pier Park)...................... Nov 9, 2013

Sign up today for a Great Strides walk near you and join us in adding tomorrows.

www.cff.org/greatstrides.

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Before You Push The Elevator Button

The number of bacteria present on an elevator button is almost 40 times higher than on a public toilet seat, according to new findings. Research carried out in hotels, restaurants, banks, offices and airports, showed that the level of bacteria on elevator buttons averaged 313 “bugs” per square centimeter, compared to 8 bugs on the average public toilet seat. Among the common bacteria likely to be found are E. coli, Staphylococcus aureus and methicillin-resistant Staphylococcus aureus (MRSA).

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Virtual CF Education Days

**Research and the Future of CF Care**

Produced by the Cystic Fibrosis Foundation (CFF), this innovative series of webcasts brings the cystic fibrosis (CF) community together in a forum to learn from experts about living with CF and the latest in CF research.

Providing an opportunity to answer questions from the CF community, this educational tool helps to empower people with CF, families and friends to take an active part in CF care through increased knowledge about the research and the highest quality care. Learn more and log on at www.cff.org/LivingWithCF/Webcasts.
Meet Our New CF Program Coordinator: Sandie Hudson

As of January 2013, Sandie is our dedicated Cystic Fibrosis Coordinator for the Nemours Children's Clinic. Sandie started the medical field in 1992 as a Respiratory Therapist at Sacred Heart Hospital and held that position until 1999 when she graduated from USA as a Registered Nurse.

She continued to work at Sacred Heart (on School Age/Adolescent) until 2001 when she graduated from USA with a Masters degree and began working in a private primary care clinic as a Nurse Practitioner. Sandie worked in Primary Care until 2005 when she came to Nemours as a Nurse Practitioner working in the Endocrine Department.

For the past 2+ years, she has been working in both the Endocrine and Pulmonology departments. “I am very excited to be the coordinator for our CF Program and want to help make our site the best in the country,” says Sandie.

Some of her responsibilities as CF coordinator are:
• Coordinate care for the patients to ensure all needs of the patient and family are met.
• Review of each patient’s medical record to ensure all recommended annual labs are requested. Ensuring a Registered Dietitian, Social Services and Respiratory Therapy visits are scheduled as needed.
• Ensure all patients and families have the opportunity to be a part of the CF Registry.
• Place all data from patient encounters into the CF Registry.
• Ensure family is notified if appointments are overdue and assist in overcoming barriers which may be causing missed appointments.
• Assist with several Quality Assurance programs in the clinic to improve patient care.

We are very glad to have a dedicated CF Coordinator to help in overseeing your child’s care. Please call anytime with questions.

High Calorie Recipes for CF Patients

Emilee Shelton, Licensed Dietician

Swisshed Cubed Steak

Ingredients:
Vegetable oil-enough to fry the steak
1 package cubed steak
Flour-enough to coat steak
Salt & Pepper to taste
Pepper-to taste
Shredded Swiss cheese (or use your favorite flavor or mix of shredded cheese)

Directions:
Season raw steak with salt, pepper, and garlic powder. Coat seasoned steak with flour. Heat ½ of vegetable oil in frying pan. When oil is hot enough for flour to bubble place coated pieces of seasoned steak in frying pan and cook until browned on both sides. Remove from frying pan and place on paper towels to drain off oil. Meanwhile, in a large casserole dish, spray with Pam cooking spray to eliminate sticking. Spread out pieces of butter across bottom of dish and layer sliced onion. Bake in 350 degree oven until onion starts to caramelize. Next layer browned steak on top of caramelized onion. Place a healthy dollop of sour cream on top of each piece of steak. Then top sour cream with a generous amount of shredded cheese. Return to oven and bake until sour cream and cheese melts in to the steak and onions. Serve with mashed potatoes and green beans.

Clam Dip

Ingredients:
1 package cream cheese
1 can minced or chopped clams (drained with reserved juice)
1 tbsp clam juice
1 tbsp lemon juice
1 tbsp worcestershire sauce

Directions:
Soften cream cheese in microwave for one minute on high. Add clams, clam juice, lemon juice, and worcestershire sauce. Mix thoroughly and serve with pretzels, chips or your favorite crackers.

Homemade Onion Dip

Ingredients:
1 package cream cheese
1 onion

Directions:
Soften cream cheese in the microwave for one minute. Stir cream cheese. Grate onion into cream cheese with a very fine grater. Grate over cream cheese so that the juice from the grated onion goes into the cream cheese. You can stir and taste to adjust how much onion flavor you want in your onion dip. Serve with pretzels, chips or your favorite crackers.

Above recipes donated by William Potter and Family

Candid Camera:
Nemours Pensacola CF Care Team

Left to Right: Glenn Hildreth, RRT, Respiratory Therapist; Jackie Gantzhorn, RN, Pulmonary Nurse; Jenny Phillips, Patient Service Specialist; Dr. Okan Eldemir, Program Director; Ashley Tubb, LPN, Pulmonary Nurse; Sandie Hudson, ARNP, CF Clinic Coordinator; Melanie Fryou, LCSW, Social Worker; Dr. Alicia Della Volpe, Pediatric Pulmonologist; Emilee Shelton, Licensed Dietician.
Traveling with Cystic Fibrosis

So you want to see the world… or Disneyworld. Traveling, whether for vacation or business, can be fun, but people with Cystic Fibrosis need to take a few extra precautions. First, you need to make sure that you are healthy enough to travel – this is especially important if you are planning to travel by air and have significantly reduced pulmonary function. You should discuss your plans with your doctor to make sure there are no exceptional concerns.

If you will be staying in a hotel, make sure that you book a non-smoking room. If you have medications that need to be refrigerated, find out in advance if your room has a refrigerator to keep meds in. You can use a cooler and ice, but that’s not ideal.

Take a list of all the medications that you use regularly, as well as those that you use on occasion. Make certain that you have an adequate supply of medicines and medical supplies, both for your trip and until you can obtain refills when you return. (Don’t forget your inhaler – your physician won’t do you much good sitting in your cupboard at home if you need it!)

If you will be traveling by air, the FAA requires that all medications be “properly labeled (professionally printed label identifying the medication or a manufacturer’s name or pharmaceutical label).” Some people recommend having a letter from your doctor listing your diagnosis and medications; you may or may not need such a letter if you are traveling to another country. It is advisable to put all medications in a zip-lock plastic bag to make inspection easier. Given that medications and supplies can add up, it is important to know that “The limit of one carry-on bag and one personal item (e.g. purse or briefcase) for each traveler does not apply to passengers with disabilities; medical supplies, equipment, mobility aids, or assistive devices.” However, you will need to identify yourself as having a disability to qualify for this, and you should check with your airline in advance to ensure that it cooperates with ADA guidelines regarding this. Additionally, when packing your meds, make sure you have a separate and ample supply in a carry-on other than what is packed away. If you have layovers, cancellations or something happens to your bags which contain your main supply, you’ll need an emergency supply until you and your bags are reunited. Remember once your bags go through security and your medications are checked, you won’t see them until you get off the plane.

Security is something else to think about. When going through security, give yourself and extra 20-30 more than anyone else would. Here are some important tips from the FAA:

- Notify the screener that you have a medical condition and are carrying your supplies with you.
- Make sure insulin (vials or outer box of individual doses), jet injectors, pens, infusers, and pre-loaded syringes are marked properly (professionally printed label identifying the medication or manufacturer’s name or pharmaceutical label).
- There is no limitation on the number of empty syringes that you will be allowed to carry through the security checkpoint; however you must have insulin with you in order to carry empty syringes through the checkpoint.
- Lancets, blood glucose meters, blood glucose test strips can be carried through the security checkpoint.
- Notify screeners if you’re wearing an insulin pump and ask if they will visually inspect the pump since it cannot be removed from your person.
- Insulin pumps and supplies must be accompanied by insulin with professionally printed labels described above.
- You can ask for your items to be visually screened, but nebulizers and other medical devices may be tested for chemical residues and X-rayed.

Make sure that you are prepared to keep medications refrigerated if necessary (e.g., Tobi, Pulmozyme, insulin). Pack a quick freeze pack to keep refrigerated medicines cold and use an insulated container. If you will be traveling without access to refrigeration, you might consider buying a portable 12 volt refrigerator. Several models are available for $50.00 or so that can be plugged into the 12 volt electrical outlet in your automobile, and some have adaptors that will allow it to be plugged into a standard 120 volt outlet. If you are traveling to Europe, remember that you will need a step-down transformer (not just an adaptor) or you will overload the appliance (unfortunately, we know this from experience). We took a refrigerator with us as a carry on last summer, and plugged it into standard outlets in the airports in between flights.

Travel can often leave a lot of sedentary “down time,” so make sure that you find some ways to be active and build some exercise into your trip. Just as importantly, many of us try to pack too much into our vacations. Rest and relaxation are especially important; you don’t want to come home exhausted because you overdid it. Oh yeah… and have fun!

A Day With Cystic Fibrosis
by Destin Johnson

What does your daily routine consist of? Is there enough time in your day to accomplish your tasks? Could you handle additional tasks with the same time schedule?

For me, day and time is uncertain. My name is Destin Johnson. Twenty-seven years ago, I was diagnosed with a genetic disease called Cystic Fibrosis (CF). CF is a disease that causes thick mucus to be released in the airway and or duct of different organ systems in the body. Since the time of my diagnosis my daily routine has been as follows;

I start with an hour of breathing treatments and chest percussion which I perform twice a day to open and clear my airway of mucus blockages. With a food requirement of 2,500 calories per day, I eat heavy well-balanced meals to maintain energy, weight and strength, along with two multi-vitamins. Before each meal, I consume six capsules of pancreatic enzymes and three capsules before snacks to help with food digestion and absorption. I eat every two hours to maintain adequate calorie intake and blood sugar levels. I also monitor blood glucose (sugar) levels and take insulin day and night to fight diabetes that developed through CF as I reached fifteen years of age.

As I tackle the tasks of my daily life, I am never too burdened that I can’t help improve the lives of others. I am a Respiratory Therapist who strive daily to help others who face similar lifestyles.

We’d Love to Hear from You!
We’d love to hear from you concerning this newsletter and other Cystic Fibrosis issues that are of concern to our fellow parents and patients. Please send an email to cowens@nemours.org if:
- You have a newsletter story idea.
- You have a topic you’d like for us to cover in the newsletter.
- Would consider writing or sharing your story.
- You have a recipe to share.
- You want to share a CF Event.

CF Mom’s Support Group Mtg
For a schedule of when the CF Moms will get together, contact Laurie Hutchison at lhutch1@cox.net or (850) 582-4738.