CF Connection special edition!

Summer 2015

University of Florida Pediatric Pulmonary Center

Time for Summer Vacation!

Summer is coming and with that, a change in routine. Here are some ways to make the most of vacation without letting care…and health, slide.

Stay wet and salty

People with CF lose more salt in their sweat and you might have noticed that summer is a sweaty time. It's important to eat plenty of salt and salty foods and drink enough fluids to prevent dehydration. Choose foods high in salt (sodium) and be generous with the saltshaker. People with CF who are active in sports or play outside in the hot weather have even higher salt and fluid needs.

Sports drinks such as Gatorade® or Powerade® do not have enough salt for a person with CF during times of increased activity, so it is recommended to add 1/8 teaspoon of salt to every 12 ounces of sports drink.

Keep up the airway clearance

You may be on vacation, but your lungs are not. A sure way to mess up a nice vacation is to let go the lung therapies and get sick. You might need to switch up your airway clearance technique to fit the plans. If you can't take your vest system, you might try using an Acapella or active cycle breathing technique. Along with all that great summer exercise (hiking, swimming, beach volleyball?), you can keep your lung clearance up and have fun!

Sunlight...the good, the bad, and the ugly

Sunscreen! We know that a few minutes of direct sun exposure helps your body make Vitamin D. More than a few minutes and you need to remember the sunscreen so you don't burn. Now, that might seem like advice that isn't particular to CF care but it might just be. Some of the antibiotics you might need this summer create a "photosensitivity," making your skin more sensitive to the sun than usual. You should keep your skin covered from sun exposure if you are taking Cipro, Septra, Zithromax, or Levofloxacin. It would be a good plan to have your local pharmacist look at your full medication list to see if there are other things that might make the sun a hazard.

Keep 'em cool

If your daily regimen includes either TOBI or Pulmozyme,

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Inside this issue:

We Need You!

Please let us know if you have any tips or stories you would like to share. We would love to include them in our Family Corner! Send them to us at the address on page 4 or e-mail them to hillajb@peds.ufl.edu.

Tips for Summer Vacation (continued)

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some advance travel planning will be needed. Both of these medications MUST STAY REFRIGERATED all the time, no matter what! We recommend that you only take along the amount needed for the trip so the remaining supply stays safely cool in the fridge at home. You might also need an alternative approach to disinfecting nebulizer kits. The choices for disinfecting are:

- ✓ Boiling for 5 minutes (either stovetop or microwave)
- ✓ Dishwasher with water over 150 degrees
- ✓ 3 minutes in a 1:50 mixture of bleach (roughly 1 oz bleach to 6 cups water)
- ✓ 5 minutes soak in 70% isopropyl alcohol
- ✓ 30 minutes in 3% hydrogen peroxide

Do NOT use vinegar! It does not do the job.

Not in the car! Not in hot weather!

There are other medications that need some care. Those enzymes that go with you everywhere…ready for the summer cheeseburger and milkshake stop…do not love being left in a hot car. They are not effective when overheated. Same goes for the inhaler you use before airway clearance, only in addition to the medication becoming "inactivated," the canister can actually burst! They would be happy at room temperature or if you are going to the beach, you could seal them in a watertight container and put them in a cooler. They will also appreciate that watertight container if you are doing the water rides. The propellant in your inhaler loves water and will soak it right up, making your inhaler useless.

Keep food safe

One way to spoil your summer vacation is to eat spoiled food and get foodborne illness! Here are some tips to keep your food safe this summer:

- ✓ Plan ahead—if you are traveling with perishable food, keep it in a cooler with plenty of ice or freezer packs. Keep drinks in a separate cooler so that the cooler with the food is not opened frequently.
- ✓ Pack safely—pack meat and poultry while it is still frozen so that it stays colder longer. Keep raw meat and poultry separate from other foods.
- ✓ When outside, keep the cooler in a shady spot and cover it with a light-colored blanket or towel to reflect the heat.
- ✓ Don't leave perishable food out of the cooler. Throw away food that has been out of the cooler for two hours or more (one hour when the temperature is over 90° F).
- ✓ Wash hands with hot, soapy water before and after handling food. If you don't have soap and water, use hand sanitizer.
- ✓ When grilling, use a meat thermometer to be sure the food reaches a safe temperature. Checking the color of the meat is not enough to be sure it is cooked to a safe temperature.
- ✓ Serve grilled meat on a clean plate—not the same plate that held raw meat, poultry, or fish.

Stay healthy and HAVE FUN this summer!

Cindy Capen, MSN, RN

Condiments Count!

Looking for a way to increase calories without trying to fit in another snack? Try adding high-calorie condiments to meals and snacks you are already eating! Here are some ideas:

Sour cream (2 Tbsp = 50 calories) – Put on baked potatoes, burritos, and tacos; stir into cream soups.

Hummus (2 Tbsp = 50 calories) – Spread on pita bread or crackers; use as a dip for raw veggies.

Wheat germ (2 Tbsp = 50 calories) – Sprinkle on yogurt, pudding, oatmeal, and ice cream; mix into milkshakes.

Large black or green olives (5 olives = 50 calories) – Add to salads, tacos, nachos, and pasta.

Shredded coconut (3 Tbsp = 55 calories) – Add to trail mix, cereal, oatmeal, ice cream, yogurt.

Cheese dip (2 Tbsp = 60 calories) – Melt over cooked veggies; use as dip for raw fruits and veggies.

Heavy cream (2 Tbsp = 100 calories) – Add to mashed potatoes, macaroni and cheese, cream soups, oatmeal, eggs; mix with whole milk.

Mayonnaise (1 Tbsp = 60 calories) – Spread on sandwiches and burgers; add to fruit and veggie salads.

Guacamole (2 Tbsp = 60 calories) – Add to tacos and burritos; use as veggie dip.

Cream cheese (2 Tbsp = 100 calories) – Spread on bagels; use flavored cream cheese as a fruit dip.

Honey (1 Tbsp = 60 calories) – Serve on bagels, toast, warm rolls, and cornbread; add to cereal and oatmeal; mix with peanut butter; use as a fruit dip. **Not for children under one year old.

Nut butters (1 Tbsp = 100 calories) – Spread on breads, crackers, apples, bananas, and celery. Try Sunflower seed butter, almond butter, or pecan butter.

Butter (1 Tbsp = 100 calories) – Add to vegetables, breads, muffins, pastas, casseroles, hot cereals, eggs.

Nuts, seeds, ground flax seeds (2 Tbsps = 100 calories) – Add to trail mix, salads, cereal, ice cream, yogurt; mix into muffins, cookies, and quick breads.

Vegetable oil (1 Tbsp = 120 calories) – Drizzle over noodles and vegetables; use in salad dressings; use as a dip for breads and rolls.

Salad dressing (2 Tbsps = 145 calories) – Serve with salads; use as a vegetable dip; spread on sandwiches.

Smoking and CF

Everybody knows that cigarette smoke (or cigar…or pipe tobacco) is bad for the lungs. People without CF can resist the damage from smoking better than people with CF. That's because when the bad stuff from smoke gets stuck in the mucus inside the lungs, the natural cleaning system in healthy lungs sweeps out the mucus and the bad stuff with it. Once it gets swept up to the throat, healthy people either swallow it or cough it out.

In a person with CF, the natural cleaning system doesn't work. The mucus in a CF lung is thick and sticky and the "mucociliary elevator" that would normally carry mucus up and out, can't move that thick and sticky stuff. This means that when the junk in smoke gets stuck in the mucus of a CF lung, the only way it gets out is with that daily airway clearance and exercise we are always asking you to do. Airway clearance techniques like CPT, vest therapy, IPV, autogenic drainage, active cycle breathing, flutter or acapella work well but are far from the perfection of the natural clearance of a healthy lung. In other words, you can't just knock all the smoke right back out…only some of it.

It's not just a matter of keeping the lungs clean. People who breathe in smoke have more inflammation in their lungs and have a harder time fighting lung infections. These are both problems for someone with CF and smoke makes that worse. In other words, breathing tobacco smoke will make their disease worse. All this is to say, it's a really, really bad idea to smoke if you have CF and a really bad idea to smoke around a person with CF.

NO ONE should smoke in the house where someone with CF lives. A smoking room doesn't solve the problem since smoke floats in the air and sticks to every surface. It's like trying to have a peeing area in a swimming pool!

NO ONE should smoke in the car a person with CF rides in. An open window doesn't prevent smoke exposure. It still circulates through the air in the car. If you are a parent who smokes, please smoke outside away from the house. It's important that you do this for your child.

Quitting tobacco is really hard but it is so important. For help with quitting, call 1-877-848-6696….and consider the health benefits for everyone around you!

Cindy Capen, MSN, RN

Let's Talk about Tune-Ups

Some of you are well acquainted with the 10 day- 2 week hospitalizations we call "tune-ups," while others of you have not yet had this experience. Here are some things to keep in mind about why you come in for tune-ups and how to handle the experience.

CF lung disease involves an ongoing battle against bacteria, and sometimes fungus or mycobacteria, which make you feel worse at times. You may have more cough, more mucus, change in sputum color to yellow or green, less energy, and/or less appetite. Pulmonary function tests (PFTs) will be down. Our first response is, almost always,

Let's Talk about Tune-Ups (continued)

to try to treat at home. We usually prescribe an oral antibiotic, an inhaled antibiotic, and more airway clearance. If this doesn't work, the next step is admission into the hospital to get you back to your best.

During your hospital stay, you will get IV antibiotics, airway clearance four times a day, lots of food, and plenty of rest. It takes all four to get your lungs better and recover lost weight. Let's talk about getting this done because it is a team effort!

If you don't have a port, you will have a PICC line placed. This is a long, flexible tube that enters a vein in the arm and slides all the way up that vein, around the curve of the shoulder, and empties into a large vessel leading to the heart. PICC lines usually last through a whole tune-up making repeated IV placements unnecessary. PICC lines are used to give IV antibiotics and to draw blood to check that levels of some medicines are high enough to work and not high enough to harm.

Next is the four times a day airway clearance, and you have choices. If you have a favorite technique, you can continue doing that in hospital. We can also order a variety of techniques so you can try new things. You can choose from chest physiotherapy (CPT), vest, acapella or flutter, IPV, autogenic drainage, or PEP. The respiratory staff can teach you and let you try them all. This is a great idea because you might find a technique that works better than what you had been doing at home.

Food is the next important step in a tune-up. You will have double portions, snacks, and maybe nutrition shakes delivered to your room. You can also seek food elsewhere…lots of places in Gainesville deliver and the staff can help you if you want to order out. Good nutrition is important to help heal your lungs, so eat as much as you can and take your enzymes if they are prescribed.

Rest is the last critical part of a tune-up. Sleep while you are in the hospital to help your body heal.

The hospital staff needs your help to get you all the care necessary to regain your best possible health. Here's what you can do to make it all work:

- ✓ Be in the room when medicine or treatment is due.
- ✓ Get lots of rest.
- ✓ Eat as much as you can.
- ✓ Disinfect or wash your hands every time you enter or exit your room.
- ✓ Wear a mask when you leave your room or if another person with CF is visiting your room while you are there.
- ✓ Tell one of the pediatric pulmonary staff if you aren't getting the care you should:
- ✓ If you don't get four airway clearance therapies every day
- ✓ If the airway clearance isn't done well
- ✓ If nebulizer kits aren't changed daily
- ✓ If you aren't getting double portion meals and snacks
- ✓ If you aren't getting nutrition shakes (if you use them)
- ✓ If you aren't getting enzymes delivered in time for meals and snacks
- ✓ If you aren't getting all your IV medicines
- ✓ If anyone breaks sterile technique when doing your PICC meds
- ✓ If people aren't handwashing/disinfecting and wearing gowns and gloves when they enter your room

When all those symptoms that caused you to come in the hospital are gone and pulmonary function tests are back to baseline or your personal best, you can go home! Tune-ups may seem scary but they really are just the best way to keep you healthy.

Cindy Capen, MS, RN

When to Call About Changes in Lungs

Many people aren't sure when to call the CF Center and when to wait and see. Here are some tips for determining when to call. Let's start with lungs and then we will discuss digestive system changes.

An "exacerbation" or worsening of lung disease in CF is sometimes an all-of-a-sudden event, but more often is just a sneaky increase in cough over days or weeks along with a decrease in energy or appetite. Either one can be confusing. A cough that is suddenly worse can usually be watched for a couple of days. If it continues, please call us to discuss antibiotics.

Noticing the sneaky cough means getting a clear picture of what "best" looks like in your case and then checking the daily or nightly cough against that best. For example, when does the cough usually happen? Lots of people notice more cough first thing in the morning, with airway clearance or exercise. Some people cough off and on all day but not usually at night. Notice your/your child's cough right after a course of antibiotics when the cough is as good as it gets and then measure against that standard. If you notice more cough than usual for a few days, call us. A night time cough should also prompt a call.

Whether it comes on fast or slow…it's important to treat an increased cough so it doesn't get worse. Remember that in CF, the lung's ability to clear secretions is not good and the worse an infection gets, the longer it takes to get back to "best."

Another lung symptom to be aware of is change in the color of the sputum (mucus) that usually happens with the increased cough. This may be from clear to yellow or green. We also want to know if there is blood in the sputum. If you see blood, it may just be streaks in the mucus or it may look like it's all blood. This is a sign that inflammation has damaged the walls of little blood vessels in the lungs. Don't panic because it's not unusual in CF, but please call us.

We also want to hear from you if you/your child have pain that is sudden, bad, or gets worse with a deep breath or cough. Pain is most often due to sore muscles from coughing a lot, but it could be something else. If you call, we can help you decide what is causing it.

To sum up, lung things to call about include increase in cough and mucus, changes in color of mucus, pain, and of course, difficulty breathing. If you are concerned, call. We don't want you to worry. Don't feel like you are bothering us—we are here to help!

Cindy Capen, MSN, RN

When to Call About Digestive System Problems

Now, let's talk about changes in the digestive system that should prompt a phone call. Some of these will sound familiar because they are part of the routine questions asked in clinic.

You already know that in most people with CF, enzymes that break down food can't get from the pancreas to the intestine.That is why people with CF take digestive enzymes (such as Creon or Zenpep) to replace what the body usually makes. These enzymes, taken immediately before eating, will help digest the food into bits that the body can use for energy. If enzyme doses are missed or the dose is too low, the food isn't broken down enough and the intestines are left to manage much bigger loads than they can do well. Then you would notice pain, gas, bigger stools, or more frequent, loose stools. These are things to call about.

On the other hand, if you/your child stop having regular bowel movements or all that's coming out is liquid, that's another cause for concern. This will likely be associated with a feeling of pressure or pain because of an over-full intestine. Call and we can help decide how best to manage. The earlier you call, the easier the solution.

Good nutrition is very important for growth and good lung health. Weight loss is a bad thing and we want to know about it. Our CF nutritionists are experts in finding ways to help regain or maintain good weight. This is another time when calling sooner is better than waiting.

In summary, weight loss, belly aches or pains, and changes in bowel movements should all prompt a call to your CF Care Team.

Cindy Capen, MSN, RN

Exercise

Watch this! http://livingxtreme.org/

The guidelines for pulmonary care published by the CF Foundation do recommend exercise in addition to airway clearance and for overall health benefits. It is clear that aerobic exercise (such as jogging, swimming, dance) improves pulmonary function and overall health while more isolated exercises are recommended to maintain flexibility and muscle strength (such as weight training, yoga).

It is never too early to lay the foundation for a lifetime of fitness! Recommendation for exercises for people of all ages can be found at http://www.cff.org/UploadedFiles/LivingWithCF/StayingHealthy/LungHealth/Exercise/Day-to-Day-Exercise-and-CF.pdf

The bottom line? Exercise for people with CF is super important and it is never too early to start. Get moving!

Milk Myths and Facts

Have you heard that milk causes mucus production? Think you don't need enzymes when drinking just a glass of milk? Do you drink milk because it has calcium and vitamin D? Read on to find out if these are myths or facts!

Drinking milk leads to increased mucus production and makes mucus thicker.
FALSE—Milk is an emulsion (drops of one liquid dispersed in another liquid), which can create a feeling in the mouth that could be mistaken for mucus. Milk does NOT increase mucus production or make mucus thicker.

The calcium in milk can increase the risk of developing kidney stones.
FALSE—The calcium in milk binds with a salt, called oxalate, in the intestine. This binding actually reduces the risk of kidney stones.

Milk is a good source of vitamin D and calcium.
TRUE—Milk is an excellent source of vitamin D, calcium, protein, and other important nutrients. Milk is fortified with vitamin D (it's added during processing), but not all dairy foods have vitamin D. Read the label to find good sources. Whole milk is great for people who have CF because it has 150 calories in each cup. Add some heavy cream and chocolate or strawberry syrup for even more calories!

Milk is a beverage, so people with CF don't need to take enzymes with it.
FALSE—Milk has protein, fat, and complex carbohydrates. People with CF who take enzymes should take a snack dose of enzymes with milk.

You can tell if you are getting enough calcium by checking the calcium level in your blood.
FALSE—To keep your blood calcium level normal, your body takes calcium from your bones if you are not getting enough in your diet. This can put you at increased risk for bone disease.

Adapted from Milk, Calcium and CF: True or False? From Rainbow Babies and Children's Hospital.

References:

Review article: Milk Consumption Does Not Lead to Mucus Production or Occurrence of Asthma. Wunthrich et al. Journal of the American College of Nutrition, Vol. 24, No. 6, 547S-555S (2005).

Curhan G, Willett WC, Rimm E, Stampher MJ. A prospective study of dietary calcium and other nutrients and the risk of symptomatic kidney stones. N Engl J Med 1993;328:833-8.

Airway Clearance—A Menu of Choices!

You all know that airway clearance is an essential part of daily care for a person with CF. Do you know that you have a bunch of choices in how you accomplish that?

Airway clearance for children under 18 months is pretty much limited to chest percussion and postural drainage using either a cupped hand or a palm cup percussor to clap on the chest wall while the baby is moved through different positions: Sitting leaned forward, sitting leaned back, on the tummy, on the back and on each side. This drains the different branches of the lungs. Once children are old enough to cough after the therapy, that should be added in. Tickling and getting very active (crawlers and toddlers love chase games) also help to mobilize the secretions loosened. A good time for play!

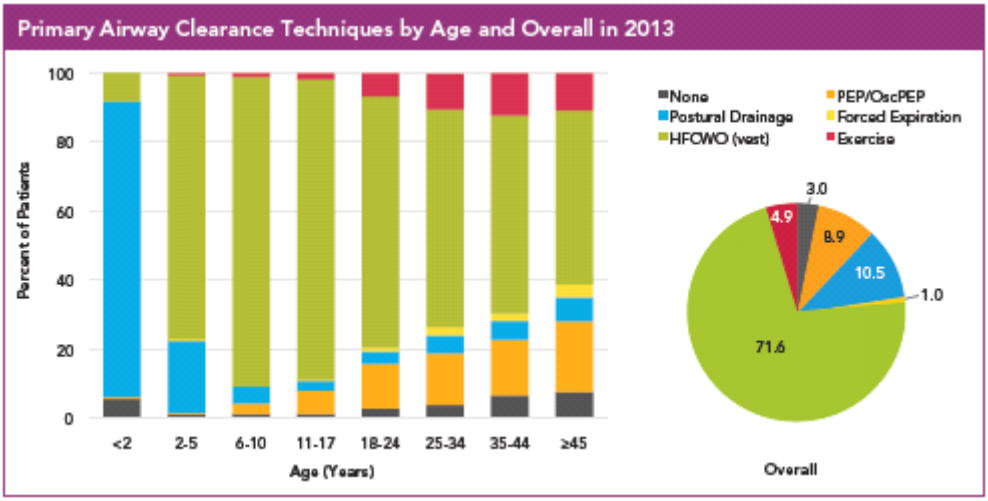
Once children reach 18 or more months, they can usually fit into a jacket or vest for a "chest wall oscillation system" often just called a vest. This is a machine that runs on electricity and attaches by hoses to a jacket and where pulses of air vibrate the chest to loosen secretions. Throughout the treatment it is still essential to get them to cough to move the secretions that have been loosened. Shaking without clearing isn't very useful!

Flutter, acapella and PEP are techniques that use handheld devices that children can usually master when they reach school age, around 6-8. These techniques only work if the child is paying attention during therapy and children do need supervision or make sure they are really putting in an effective effort.

ACAPELLA

Active cycle breathing and autogenic drainage are techniques that are a bit more complicated and so more fitted to older children and adults. They are entirely breath control methods of airway clearance, requiring no equipment at all which makes them well suited to working around busy and flexible schedules.

Data gathered by the CF Foundation in 2013 shows increased use of the more mobile and flexible airway clearance techniques in the older age ranges. All of these techniques are demonstrated to be effective and so it is simply a matter of picking the best for you!



An IEP or 504 Plan—Does My Child Need One?

If your child is in school, make sure that he or she has a plan in place for any needed accommodations. These accommodations may include getting treatments at school (for example, enzymes, albuterol, insulin or nutritional supplements, or even g-tube feeds or vest treatments). Accommodations may also include permission to miss school (and not be marked with an unexcused absence) for clinic visits or hospitalizations, make up missed work/tests, have water or sports drinks if exercising in heat, or use the bathroom when needed.

Some schools provide these types of accommodations willingly, with a verbal agreement, after talking with parents. However, often a more formalized plan is needed.

If your child attends a public school, there are laws that mandate that the school make the accommodations necessary for your child to receive the same quality of education that other children receive. Public schools must meet these requirements if they wish to continue receiving federal funding. These accommodations are stated in either a IEP (Individualized Education Plan) or a 504 plan.

When developing an IEP or 504 plan, school professionals (usually teachers, school nurse, or guidance counselors) meet with parents (and after age 14, the student) to list goals and then to outline the accommodations needed to meet those goals.

Ideally, the school identifies children in need of an IEP or 504 plan. However this does not always occur and sometimes it is up to parents to request a plan. You can make this request verbally or we will be happy to write a letter asking the school to consider your child for an IEP or 504 plan. You should be invited to the IEP or 504 plan meeting and you are welcome to take a friend or other professional for support.

It's best to get an IEP or 504 plan in place at the start of the school year. If an IEP or 504 plan is in place, you have something to fall back on if you are having challenges with the school. If you don't have a plan already in place, the school doesn't have to make accommodations. For example, if a teacher won't allow your child a make up test because your child was in the hospital, but this accommodation is in the IEP or 504, you can advocate with the school board. However, if you didn't have this accommodation previously outlined in an IEP or 504, you may not get very far in arguing your child's case. It is also helpful to remember that even though a school may be helpful and responsive one year, this can change the next year if school personnel change. There can also be big changes between elementary school and middle school or between middle school and high school. So it is most helpful to have a plan in place.

Private schools most often are not required to do 504 plans or IEPs. However, it is still a good idea to request a meeting with school personnel to educate them about CF and your child's needs. If you meet with major barriers from the school, the American's With Disabilities Act (ADA) may be helpful to you. The ADA protects people with disabilities from discrimination. If you feel your child is being discriminated against in getting an education, you can contact the US Department of Justice or the US Office of Civil Rights.

If you would like more information about accommodations in school or would like to talk more about your child's situation, please contact Susan Horky.

Family Corner

Patient and Family Centered Care—The New Normal

Patient and Family Centered Care refers to a standard in healthcare that aims to put the patient and their family at the center of all healthcare interactions. Healthcare professionals are more aware than ever before that patients and their families have a very important role to play in their own and their loved ones continued health and wellbeing.

A patient and family centered approach means changes for all of us, both healthcare providers and patients and their families. Healthcare providers need to listen closely to what patients and families want from their healthcare team but just as importantly, patients are encouraged to actively engage with our team.

Specifically, what does this mean? Healthcare is now very focused on providing patients with high quality, safe care. You may also hear the words "the patient experience" used. As well as keeping you safe and giving you quality care, hospitals also want patients and their families to have a "very good experience." Patients and their families can help in these processes by being active participants in their care: 1) being well prepared for outpatient visits, 2) asking the team questions so that the plan of care is clearly understood, 3) staying in touch between visits, 4) being an active patient (or having someone with you) while in the hospital. At another level, patients and families are encouraged to provide feedback by completing after visit or hospital surveys, complaining or commenting in a constructive manner, being a part of a hospital or CF specific advisory council or volunteering to participate as a family leader on committees developing new projects and initiatives.

The Pediatric Pulmonary Center at the University of Florida is committed to a patient and family centered approach to care. Currently, the division employs a part-time family partner, Angela Miney. The family partner's goal is to help the team give you the best care we can all the time. Angela does this by bringing the family perspective to the team and also teaching healthcare students what dealing with the healthcare system is like from the point of view of the patient and family. If you would like to share any of your experiences with us, share your healthcare story, be a family leader by participating on a committee or advisory council, please contact Angela at: aminey@peds.ufl.edu

Angela Miney, Family Partner

Pediatric Pulmonary Center
University of Florida
Health Science Center
P.O. Box 100296
Gainesville, Florida 32610-0296

Phone: 352-273-8380
Toll Free: 1-800-749-7424
Fax: 352-392-4450

Physicians and Nurse Practitioners:
Mutasim Abu-Hasan, MD
Mary H. Wagner, MD
Pamela Schuler, MD
Sreekala Prabhakaran, MD
Yanerys Colon-Cortes, MD
Pritish Mondal, MD
Dawn Baker, ARNP
Elizabeth LeFave, ARNP

Pharmacy:
Leslie Hendeles, PharmD

Nurses:
Cindy Capen, MSN, RN
Brenda O'Neal, RN
Rhoda Reed, SRNS

Nutritionists:
Ellen Bowser, MS, RN, RD
Jennifer Hillan, MSH, RD, LD/N

Social Worker:
Susan Horky, LCSW

Respiratory Care:
Shands/UF Pediatric Respiratory Care Team

Office Staff:
Brenda Martin
Lisa Harvey

Family Partner:
Angela Miney—
aminey@peds.ufl.edu