

Breath Sounds

The newsletter of The Antonio J. and Janet Palumbo Cystic Fibrosis Center

From the Director



David Orenstein, MD

Hey! It has been a while since we have distributed a newsletter, and we are glad to be able to update you on the many exciting developments in our Center. Some of these include:

- New staff
- Quality Improvement activities
- New specialty clinics
- Update on Infection Control
- Changes in clinic schedules

I am very pleased to report that we have been awarded a \$1.5 million grant from the National Institutes of Health to continue our research on the role of exercise in patients with CF. This, along with my work as Chair of the Children's Hospital Ethics Committee, is exciting for me. The down side is that it leaves me with fewer available clinic sessions and appointments.

I am very happy to introduce Dr. Daniel Weiner, who joins our Center as the Associate Director... (it's taken us a long time to find someone with the appropriate training, personality, and neck-wear to fill this position)...

From the Associate Director



Daniel Weiner, MD

Greetings! I am delighted to join this outstanding Cystic Fibrosis Center. I moved to Pittsburgh from the Children's Hospital of Philadelphia in August 2006, along with my wife Aviva (another doc at Children's), and 4 kids (Gabe, Sam, Channah, & Shoshana). The kids have quickly become Steelers and Pirates fans. Like Dr. O, I've been very interested in CF for a long time, and worn bow-ties for a long time. I do, however, wear shoes! ☺

Along with the many activities and projects within the Center, one of my other professional interests is infant lung function testing, and more information about this is inside.

There are so many exciting projects going on within the Center, and we hope to be able to give you a taste of that excitement via this newsletter.

I look forward to meeting each of you, and working with you in your care or the care of your family. Please do not hesitate to contact me with your ideas or concerns.

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Quality Improvement: Working to Improve our CF Center

The Cystic Fibrosis Foundation is committed to providing every CF Foundation-accredited care center throughout the United States with the opportunity to improve the quality of care delivered to patients with CF and their families. A standardized training program is being offered to assist centers in looking at how their Center can accelerate the rate of improvement in CF care.

We have joined in this process at our CF Center by developing a Quality Improvement (QI) Lead Team which represents both the adult and pediatric programs. It is composed of 19 representatives from the CF interdisciplinary health care team, 3 parents from the Pediatric Clinic, and 2 patients from the Adult Clinic.



Back row: Lynne Arrington, Iris Yann, Donna Wilding, Denise Boring, Louise Bauer, Jenny Haglund; Front Row: Daniel Weiner, Jonathan Spahr, Sandy Hurban, Reid Masi; Not pictured: David Orenstein, Kathy Bliss, Connie Richless, Sandra Scharding, Megan Weiss, Joe Pilewski, Carolyn Walker, Judy Fulton and Nancy Smizik

Our first meeting occurred on December 1, 2006, with just the health care team. We decided to meet twice monthly, with a great deal of homework to be done between meetings. Our Patient/Parent representatives joined us at our meeting on 4/17/07, bringing with them lots of practical ideas about living with cystic fibrosis.

This partnership of patients, parents, and health care team working together is exciting.

The mission of the Center's QI Lead Team is to work together to develop skills to improve our Center's outcomes by assessing our current ways of operating, and developing quality improvement goals and projects.

We have chosen to focus our efforts on improving pulmonary function in our patients. There are two specific projects underway, one regarding airway clearance, and the other to improve regularity of office visits. You'll be getting more information about these projects, both in clinic and in the mail.

Our goal -- in cooperation with you! -- will be to improve the quality of life for our patients. We greatly appreciate your input through your responses to the Patient Survey. The time you have taken to share your ideas has helped the QI Lead Team to determine our current outcomes. There will be further opportunities for participation.

This is only the beginning of a formal partnership of patients, families, and the CF Care Team working together to provide the very best medical care to our patients.

Jenny Haglund, Social Worker

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Research Corner

You are the Key to Evaluating New Treatments for Cystic Fibrosis!

Significant progress has been made in recent years towards understanding how lung disease in CF develops. As a result, a number of new therapies have been developed and are in various stages of clinical trials. The CF Foundation supports the development of new treatments, and a number of potential new medications are being tested now, and many more are likely to be studied in the near future.

Developing new treatments requires that patients participate in research studies. Without clinical studies, we will never know whether new treatments are effective. As a CF Center, we are committed to helping to develop and evaluate new treatments while keeping patient care and safety as our primary concern. Your CF center is currently involved in many studies, both as one of the 18 centers in the Cystic Fibrosis Foundation Therapeutic Development Network (TDN), and on our own here in Pittsburgh. The TDN was developed in 1997 for the purpose of conducting early phase clinical studies of new therapies.

Our goal is to contribute all that we can to evaluating new therapies, so we encourage you to participate in research studies. We have had a great response from many of you who have participated in these studies. We always need more patients to participate, as very soon progress may be slowed if sufficient research participants are not identified.

To make research study participation more convenient, we are able to have research study visits Monday through Friday from 7 am to 7 pm, and on Saturday mornings. You will be meeting the research staff in clinic, but please ask your doctor or call Liz Hartigan (412-692-7060), Sandy Hurban (412-692-7042), Judy Fulton (412-692-5677), or Adrienne Horn (412-692-8069) if you are interested in learning more about the research studies available at our center.

Are there different kinds of clinical trials?

There are many kinds of clinical trials, but most of them test new forms of treatment. Clinical trials are carried out in phases, each of which is set up to gather information such as finding a safe dose or whether a new treatment may be effective. After the trial is over, and if the new treatment proves to be effective, it may be a while before the new drug is generally available. Note that one group of patients in some trials are given a placebo, which has no active medication. This group serves as a comparison group. Other studies compare different kinds of active medication. Without comparisons, it would be impossible to show that the new treatment is effective.

Am I eligible for a clinical trial?

Clinical trials are designed to answer specific questions, so they enroll only patients with specific characteristics. The researchers will make sure that you meet all the entry criteria before they enroll you. If you don't qualify for one trial, you may be eligible for others. Usually, you can participate in only one clinical trial at a time.

What are the benefits of participating in a clinical trial?

People who take part in clinical trials may have the first chance to benefit from new treatments. Whether the new treatment turns out to be helpful or not, the information gathered in the clinical trial will help researchers and can ultimately benefit all patients with CF.

Before participating in a clinical trial, Ask...

- What is the purpose of the study?
- What kinds of tests and treatments?
- How long will the study last?
- How will the study affect my daily life?
- What side effects can I expect?
- How many visits will I have to make?
- Will I be compensated for participating?
- Whom do I contact if I have questions?

By Elizabeth Hartigan, MPH, RN, CCRC

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Respiratory Corner

While you're packing for that trip to the beach or to Grandma's, don't forget to include your airway clearance device! Here are a few ideas that may make this a little easier.

- If you use a vest, it can be powered by a UL approved power inverter that plugs into the cigarette lighter of your car or truck. According to one of the vest companies, it MUST be "continuous power rating group of minimum 400 watts". One family told me that they purchased one at Cabella's for under \$80.
- If you have one of the larger, heavier Hill-Rom (formerly American Biosystems) generators for your vest, now may be the time to see about upgrading to the more compact unit. The company is currently working on a new, easier upgrade program.
- Another helpful tool for some patients may be a hand-held device, such as an Acapella, Flutter, or Quake. If your doctor recommends one of these for you, the respiratory therapists in our clinics will teach you how to use it.

Off to college this Fall?

- Again, you may want to think about trying to upgrade to the smaller, lighter weight vest. And don't forget about your compressor and nebulizer set-ups.
- Change the filters on your compressor if necessary. Get a new compressor if your compressor is more than 5 yrs old.
- Have ample supply of nebulizer set-ups.
- Work out a plan for properly cleaning of nebulizer set-ups at college.

Please contact the CF Center and ask us about any these things.

Have a wonderful summer!

by Becky Mutich, RRT

Summer Nutrition Update

With warmer weather now here we thought it would be a good time to review some summer CF nutrition tips. So, take a minute and complete the quiz below:

Q: People with CF lose more salt when they sweat than people without CF.

A: True. People with CF lose more salt than normal when they sweat and therefore need to take in more salt in their diet. This is especially true during hot, humid weather and with increased outdoor activities (i.e. long walks, playing outside, jogging, trips to the zoo or amusement park etc).

Q: Salt can be replaced by eating high-salt foods.

A: True. You should be able to get all the salt you need from the foods you eat. Eat salty foods such as potato chips, pretzels, salted nuts or pickles. Make liberal use of the salt shaker during meals. Salt tablets are not recommended.

Q: It is safe to store enzymes in your car's glove compartment.

A: False. Keep enzymes with you at all times and at room temperature. This means you cannot store enzymes in your car's glove compartment, next to windows or next to appliances that emit heat. Heat may cause the enzymes to break down and not work properly.

Q: People with CF need to drink more fluids in the summer.

A: True. People with CF do need to drink more fluids in the summer. Drink plenty of water, fruit juice or sports drinks such as Gatorade or PowerAde. Avoid caffeinated fluids. During aerobic exercise encourage fluid breaks every 20 minutes. Carry a water bottle with you at all times! People with CF may underestimate their fluid needs and this puts them at higher risk for dehydration.

By Iris Yann, RD

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New and Revised CF Infection Control Guidelines



One important goal of our center and Children's Hospital of Pittsburgh is to create an environment that is safe and comfortable for all CF families by reducing or halting the spread of organisms and germs in the hospital and clinics.

As many of you know, in 2001, the Cystic Fibrosis Foundation developed recommendations for infection control based on research, theory, and/or state and federal regulations. Many of these recommendations were implemented at our center and Children's Hospital in 2002. We sent a letter informing you of these changes and stated that as we learn more and gain more experience we would be revising and updating these changes.

For the past year, a team of infection control doctors and nurses, CF center doctors and nurses, as well as in-patient and out-patient respiratory therapists, have met to review our center data, as well as current research, to make sure we were doing everything possible to provide the best infection control possible.

As a result of this work, we have updated and revised our CF infection control policy. We have included the new out-patient and in-patient changes with this newsletter. Additional information about infection control is available on the Cystic Fibrosis Foundation website (www.cff.org), or ask your CF team in clinic. Please let us know if you have any questions or concerns.

By Louise Bauer, CF Nurse

Parent Corner

When my daughter was diagnosed with CF my husband and I took a look at our home to figure out what we could do to help protect against the spread of germs and keep her healthy. Some things we implemented were:

- We placed antibacterial gel by the front door so visitors could wash their hands before entering
- We daily wipe down surfaces (e.g. door-knobs, light switches, phone, and other "frequently touched" areas) with Lysol wipes. It only takes about 5-10 minutes to do the whole house. We do this more frequently if someone in our house is sick.
- We put antibacterial gel in our cars and always wash our hands with it when we leave a store and before coming home.
- We stopped using sponges, dish cloths, and towels in the kitchen and switched to paper towels to dry hands and Lysol wipes to clean counters, high chair, etc.

When we were told our daughter had *Pseudomonas* at 7 months old it made us take a look once again at what we could do differently around the house to protect her from this pesky bacterium. We realized we had bath toys that retained water even after we drained the tub (eg squeeze toys). Any place that water sits is a possible breeding ground for bacteria. We made sure to buy toys that can completely dry out between baths. We stopped using the "wipes warmer" we had since that always stays wet. Obviously we cannot protect completely against these bacteria, but we feel better knowing we are taking precautions where we can.

These are just some of the things we do to help keep the germs and bacteria at bay. What are some of the things that you do?

By Melissa Wiggins, parent

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Adult Corner

Appointments

Our Antonio J. and Janet Palumbo Cystic Fibrosis Center now cares for more than 450 individuals. Nearly one-half of them are adults seen in the adult program. Consequently, our adult program is growing quickly. It is just as important as you grow older to have regular clinic follow-up. A consensus of CF patient care leaders have determined that by getting frequent, close follow-up, patients have better outcomes and stay well longer. The Clinical Practice Guidelines for Cystic Fibrosis from the CF Foundation recommends quarterly (every 3 months), routine follow-up visits for all CF patients. Frequent visits allow for the assessment of lung function and nutritional status, and facilitate earlier intervention. You will hopefully be able to make your follow-up appointment before leaving clinic. If not - here are some important phone numbers for you to have at your finger-tips:

Scheduling: 412-648-6161, Option 2

Social Worker: 412-692-5590

Nutritionist: 412-692-5677

Nurse Call Line: 412-692-8764 or -5630
(Monday - Friday; 8:30 - 3:30 p.m.)

After Hours Sick or Urgent: 412-621-1200

You must identify yourself as a CF patient when you call. An adult CF physician is on-call for your sick or urgent needs after hours.

Emergency Care: UPMC Shadyside Hospital
Emergency Department

Remember that the PFT appointment is scheduled separately and is usually 15 to 30 minutes prior to your clinic appointment.

Because our clinics are always full, we often have individuals on a waiting list to get an appointment. It is essential that if you must cancel your appointment, you call at least 24 to 48 hours

before the scheduled appointment, so that someone else may be able to come in your place.

Pharmacy news

As you probably know, Children's Pharmacy has closed. Your insurance coverage may dictate what pharmacy you are allowed to use. Falk Pharmacy has agreed to mail prescriptions to our patients. If you choose to use Falk Pharmacy, their hours are Monday through Friday from 7:30 a.m. to 5:30 p.m.

Falk Pharmacy, Falk Medical Building

3601 Fifth Avenue, Room 211

Pittsburgh, PA 15213

Ph: 412-473-7427 (PHAR), Fax: 412-648-1838

A Note Regarding Refills: When calling to request refills, remember to give yourself ample time so that you do not run out. We recommend 2 weeks turn around time as many insurance companies now require prior authorization for many medications.

Fun in the Sun: A reminder to those taking antibiotics this summer. Cipro, Bactrim, Doxycycline, and many other oral antibiotics may make you particularly sensitive to sun exposure. Remember sun screen!

Welcome!

Carolyn Walker joined the adult team last fall. She is a Pennsylvania native and went to nursing school in Pittsburgh. She is an experienced nurse who came to the CF Center in November after working with CF and other pulmonary patients at Shadyside Hospital. Please be sure to say hello!

By Connie Richless, Adult CF Nurse Practitioner



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New Faces

Jonathan Spahr, MD, grew up in Pennsylvania where he went to college, medical school and did his residency training. He recently moved from Wisconsin, where he completed an Adult/Pediatric Pulmonary Fellowship at the University of Wisconsin. He is actively involved in the Lung Transplant program.

Shean Aujla, MD recently completed her fellowship here at CHP and joins the faculty, continuing her research in the laboratory of Dr. Kolls, and seeing patients.

Carrie Falvo, RD, joins the Center as a dietitian from Heinz where she worked as a Research Assistant. She is a 2004 graduate of the University of Pittsburgh and is currently working on her MPH at Pitt's Graduate School of Public Health.

Adrienne Horn, RN, BSN, is a graduate of Duquesne University. She previously worked on a surgical unit and in the OR. As a Research Coordinator for the Center, she recruits patients for studies, and sees them through the study process. She is looking forward to a trip to Italy next August.

Mark Patterson, RRT, RPFT, joined the Pulmonary Function Lab in January. He has worked for many years in the Respiratory Therapy Department, and is actively involved in the new Infant Pulmonary Function Lab.

Danielle Goetz, MD, joins the Division as a first year Fellow in Pediatric Pulmonology. She completed her residency at CHP and served as Chief Resident in Pediatrics. **Heather Strawbridge, MD**, is our other first year Fellow, and joins the Division after completing her residency in Pediatrics at Tod Children's Hospital in Youngstown, Ohio.

A "fellow" is a doctor who has finished training in their specialty (i.e. pediatrics) and is doing 3 years more training and research (in pediatric pulmonology).

Beth Butler is the Administrative Assistant to Drs. Finder and Kurland as well as the Pulmonology Fellowship Coordinator. Outside of the office, she is the mother of three sons and the grandmother of a 1 and 1/2 year old, Arionna ("Pumpkin"). **Terri Fisher** is a secretary for the Sleep program, and interacts with patients to confirm sleep appointments, greeting guests, answering phones, etc. She says that she "is fascinated with the care the doctors and nurses provide their patients, and has much respect for what they do."

New Services

We are pleased to announce the formation of a **CF-Gastroenterology clinic**! This clinic will be held monthly, and staffed by **Dr. Doug Lindblad**, a staff Gastroenterologist. Dr. Lindblad is available for consultation for problems including liver disease, constipation, gastroesophageal reflux, and uncontrolled malabsorption, and will work in concert with our nutritionists and other staff.

Genetic counselors from the **Division of Medical Genetics** are available for counseling to all our families at CHP. **Jacquelyn M. Hoover, MS, CGC**, and **Marianne McGuire, MS**, serve CF clinics in Oakland by appointment. Ask your CF team for more information if you are interested.

Coming Soon! **Dr. Jonathan Spahr** will be piloting a new Adolescent Clinic, the first of which will be held August 30th at CHP North. More information will be coming.

The Pulmonology Lab is excited to announce the opening of the **Infant PFT Laboratory**. Patients able to cooperate (usually 5-6 years of age) perform "voluntary" tests at each office visit. In order to get an objective assessment of lung function in infants and toddlers, specialized tests can be performed and were developed for young children with CF. This testing requires sedation and more time than standard testing. Ask your CF doctor for more information.