

# Cystic Fibrosis Center News

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## *Pseudomonas aeruginosa* in the Cystic Fibrosis Lung: Inevitable or Preventable?

By Stacy VandenBranden, APN, CNP

Bacteria, or microbes are one of the smallest living organisms of life, and are a purposeful part of every environment. However, when certain bacteria enter the human body they can cause a disruption in health and are then referred to as germs, or pathogens. Bacteria need the right environment to grow and flourish. Having cystic fibrosis changes the environment of the lungs, making them an inviting environment for bacteria to grow and flourish.

*Staphylococcus aureus*, *Haemophilus influenzae*, and *Pseudomonas aeruginosa* are some of the most common bacteria found in the lungs of people with CF. *Staphylococcus aureus* and *Haemophilus influenzae* are also found in children without CF. These bacteria are often responsible for familiar childhood respiratory illnesses such as ear infections, sinusitis, and pneumonias.

*Pseudomonas aeruginosa* (*Pa*) is by far the most common bacteria affecting individuals with CF over the life span. As children with CF get older, *Pa* is more likely to be present in the lungs. We know from the CF Foundation Registry (a data bank that collects information on CF patients in the US), that *Pa* is found in 15-30% of infants, 30-40% of children 2-10 years of age, about 60% of adolescents, and about 80% of adults with CF. An initial *Pa* infection may not have any noticeable immediate impact on health, but over time chronic infection causes loss of lung function and damage to the lungs. At the time of first *Pa* acquisition recorded in the CF Registry, 60% of patients had respiratory signs and symptoms, 15% had a decline in nutritional status, and 34% had no clinical symptoms. Many questions surround the role of *Pa* in CF. Some of these questions can be answered, while others will require additional research to understand.

**What do we know about *Pa*?** We know that *Pa* is common in the environment and is found living in soil and water in most climates and countries. Eliminating exposure to *Pa* in the

environment would be impossible. There are several different strains of *Pa*. It is thought that most of the first time *Pa* infections in children are from environmental *Pa* strains. However, *Pa* can mutate or change when it finds a suitable environment, and can become resistant to antibiotics making it more difficult to treat. In addition, *Pa* bacteria have a special talent for creating a gel-like layer around itself that protects it even more from antibiotics. We call this type of bacteria a "mucoid *Pa*" strain.

**How do we know if *Pa* is present in the lungs?** Regular cultures from the throat can identify that *Pa* is somewhere in the respiratory tract (throat, nasal passages, sinuses or lungs). A sputum sample is better at identifying what is actually in the lungs, so we encourage children to learn how to cough up ("expectorate") mucous for a sample at as young an age as possible. The third method of identifying bacteria in the lungs is through a procedure called bronchoscopy, which is done under sedation. Bronchoscopy involves using a special tube threaded through the nose and into the lungs to obtain a culture directly from the lungs.

**Why do some kids get *Pa* at an early age and some not until they are adults?** Little is known about individual or environmental risk factors for getting *Pa* in the lungs, or what keeps some children from getting *Pa*. A few small research studies have been done that have identified possible risk

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# Helping Children Cope with Medical Procedures

By Dora Castro-Ahillen, LCPC, ATR-BC, CCLS

Dora Castro-Ahillen, child life specialist and art therapist at Children's Memorial, presented this topic at our Annual CF Research Update.

For many children, anxiety surrounding being hurt can be one of their greatest fears. Preparing children for a hospital or clinic visit can help them have less stress regarding medical procedures. Caregivers can help children cope by doing the following:

- Gather information: Know why your child needs a procedure, how the procedure may feel and

how long it will last. Consider being with your child during the procedure and role-model positive attitudes and behaviors.

- Be honest and sensitive: Explain to your child why the procedure is needed and what the child can expect to feel, see and hear.
- Encourage curiosity and exploration: Becoming familiar with the facility and understanding the medical equipment that will be used during a treatment is very important to a child. Help your child learn about the purpose of the procedure and the equipment



that the child will come in contact with.

- Reassure your child: Make sure your child knows that the hospital, doctor's office or clinic is not a punishment and it is not necessarily a place where children will experience pain.
- Use simple language: When describing a medical procedure, try to use words that do not have double meanings or are threatening.
- Listen to your child's concerns: Let your child know that it is

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## Pseudomonas in the CF Lung

factors. These risk factors include long term, non-pseudomonal antibiotic use, having the  $\Delta F508$  homozygous genotype (two copies of the most common CF gene), female gender, and previously having *Staphylococcus aureus* in the respiratory tract. Other factors that have been speculated about but not studied include environmental tobacco smoke exposure, number of siblings with CF, breastfeeding, and daycare attendance.

**How can we better understand how to treat *Pa* infection?** The CF Center at CMH is participating in two national studies that have recently been launched to better understand *Pa* infection and how best to treat it. The first study is called **EPIC 002: Longitudinal Assessment of Risk Factors For and Impact of *Pseudomonas aeruginosa* Acquisition and Early Anti-Pseudomonal Treatment in Children.** This is an observational study sponsored by the CF Foundation Therapeutic Development Network to observe young CF patients to see what factors contribute to them



*Pseudomonas aeruginosa* under the microscope

acquiring *Pa*. Patients with CF <13 years of age, who participate in the CFF Registry and do not grow *Pa* in their respiratory cultures will be asked to participate and will be followed for 2.5 to 5 years. Normal data will be collected at CF visits. The only additional testing required will be an annual blood test for antibodies to *Pa* which can be drawn at the same time as the annual labs. Patients/parents will also be asked to complete a questionnaire annually.

The second study is called **EPIC 001: Effectiveness and Safety of Intermittent Antimicrobial Therapy for the Treatment of New Onset**

### *Pseudomonas aeruginosa* Airway Infection in Young Patients with CF.

This study also sponsored by the CFF Therapeutic Development Network will enroll patients with CF <13 years of age who grow *Pa* in their respiratory cultures for the first time in at least 2 years. The aim of this study is to evaluate different approaches to treating these patients. Participants will be randomized to one of four different treatment regimens. All study medications will be provided by the sponsor. Subjects will be monitored for 18 months. Some additional blood work and respiratory cultures will be required during the course of the study.

The question, "**Is *Pseudomonas* inevitable or preventable?**" has yet to be answered. The CFF and research community are clearly committed to understanding the role of *Pa* in CF and how best to treat it. For now, we will continue to utilize the knowledge we have to emphasize good infection control and prevention, and early treatment of respiratory infections when they occur.

# Advancing Science Through Research

by Cathy Powers, RD, CCRC

Our research team is actively coordinating multiple CF trials. Any patients with CF are welcome to participate if they meet the study criteria and their physician agrees. Participation in any study is completely voluntary. If you have questions about any of the clinical trials or would like more information please talk to your CF Center doctor or contact our clinical research manager, Cathy Powers at 773.880.8223 by phone or e-mail at cpowers@childrensmemorial.org.

## NEW STUDIES

**CP-AI-005: A Phase 3, Double-Blind, Multicenter, Randomized, Placebo-Controlled Trial with Aztreonam Lysinate for Inhalation in Cystic Fibrosis Patients with Pulmonary *P. aeruginosa* Requiring Frequent Antibiotics (AIR-CF2).** The purpose of this phase 3 study sponsored by Corus Pharma, Inc. is to evaluate how safe and effective an inhaled antibiotic under investigation (aztreonam lysinate for inhalation) is in treating patients/subjects with CF lung infections. Following their normal course of TOBI® subjects will receive a 28-day course of study drug or placebo to take 2 or 3 times a day. Subjects will then be followed for 2 months to see how long they are able to go before their lung infection requires treatment with additional antibiotics.

**CP-AI-007: A Phase 3, Double-Blind, Multicenter, Multinational, Randomized, Placebo-Controlled Trial Evaluating Aztreonam Lysinate for Inhalation in Cystic Fibrosis Patients with Pulmonary *P. aeruginosa* (AIR-CF1).** The purpose of this phase 3 study sponsored by Corus Pharma, Inc. is to evaluate how safe and effective an inhaled antibiotic under investigation (aztreonam lysinate for inhalation) is in treating patients/subjects with CF lung infections. Participants in this study will administer 28 days of the study drug or placebo 3 times a day and will be followed during the study and for 14 days after the last dose. This study involves 5 visits over 2 months and will measure changes in CF disease and lung function.

**CP-MCID-001: Evaluation of CF Patient Perception of Symptom Improvement Following Inhaled Antibiotic Treatment.** The purpose of this study sponsored by Corus Pharma, Inc. is to evaluate how useful 2 questionnaires, the Cystic Fibrosis Questionnaire (CFQ-R) and the Global Rate of Change Questionnaire, are at assessing patients' perception of changes in their well being during treatment. This is a data collection study with no blinding and no controls. Patients will be asked to participate if they come to clinic with symptoms of a lung infection and are prescribed TOBI® at the visit.

**EPIC 002: Longitudinal Assessment of Risk Factors**

**for and Impact of *Pseudomonas aeruginosa* Acquisition and Early Anti-Pseudomonal Treatment in Children with CF and EPIC-001: Effectiveness and Safety of Intermittent Antimicrobial Therapy for the Treatment of New Onset *Pseudomonas aeruginosa* (Pa) Airway Infection in Young Patients with Cystic Fibrosis.** A description of these studies can be found in the article "*Pseudomonas aeruginosa*: Inevitable or Preventable?" on page 1.

**Phase II, Multicenter, Randomized, Controlled Open-Label Study of the Safety and Efficacy of Nutropin AQ for the Treatment of Growth Restriction in Children with Cystic Fibrosis.** This study sponsored by Genentech aims to evaluate the effect and safety of daily injections of growth hormone on the growth and lung function of children with CF known to be growth restricted. Subjects will have a 50/50 chance of being selected to receive growth hormone. The length of treatment will be 12 months, but subjects will be followed for a total of 18 months. The study does require additional testing; however most of the visits can coincide with regular CF clinic care.

**A Multi-Center, Double-Blind, Randomized, Placebo-Controlled, 28-Day Study of Denufosol Tetrasodium (INS37217) Inhalation Solution in Patients with Cystic Fibrosis Lung Disease,** sponsored by Inspire Pharmaceuticals. Cystic Fibrosis (CF) is characterized by thickened mucus secretions that lead to infection and damage to the lungs. The basic problem is related to abnormal transport of water and salt (sodium and chloride ions) across airway cells. This study will evaluate an inhaled solution under investigation that may improve movement of chloride and water out of airway cells; therefore it may make airway secretions thinner and easier to cough up. This solution also makes cilia, hair-like fibers on the airway surface, beat more frequently, which may give additional benefit in secretion clearance. This phase 2 study sponsored by INSPIRE will investigate the safety and efficacy (or health benefit) of two strengths of this investigational drug compared to placebo (solution with no drug in it). The drug or placebo is administered via a nebulizer three times per day for up to 28 days to subjects with mild to moderate CF lung disease.

## ONGOING STUDIES:

**Project on Adult Care for Cystic Fibrosis,** sponsored by Boston Children's Hospital/NIH. The aim of this 3 year survey/interview study of adult patients with CF is to better understand the quality of life, social and logistical needs of adult patients and their families.

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## Advancing Science

**Genetic Modifiers of Cystic Fibrosis (CF Twin/Sib Study).** This study, done in collaboration with Johns Hopkins University, is attempting to discover what genes other than the CF gene may cause differences or similarity of illness between CF children in the same family. All families with one or more person affected with CF in the immediate family are eligible to participate. This study involves one blood draw from affected siblings and their parent(s). Medical history and clinical symptoms will also be reviewed and compared to DNA to look for additional genes that affect the course of the disease.

**Genetic Modifiers in Cystic Fibrosis Lung Disease and Liver Disease.** This study done in conjunction with researchers at Case Western Reserve and the University of North Carolina-Chapel Hill is looking at other genes that might explain why patients with the same CF mutations might display a wide range of disease severity. Identifying these genes that modify CF lung disease and/or CF liver disease could help direct new therapies.

***Pseudomonas Aeruginosa* Type III Secretion in CF.** *Pseudomonas aeruginosa* (*Pa*) infection, present in 60% of persons with CF, leads to worsening lung function in some but not others. *Pa* has the ability to secrete molecules that can damage human cells. One such family of molecules is “Type III” molecules. The purpose of this research study conducted by researchers at Children’s Memorial Hospital and Northwestern University is to find out if strains that secrete these molecules lead to more rapid worsening of lung function in CF patients.

**CFF Patient Registry.** Data is collected on individuals

with CF to better understand the illness, guide in the planning of research studies and ultimately improve the care and survival of those with CF. The CFF Registry data helps CF clinicians improve care, including nutritional status, infection control and treatment of lung disease.

**The Epidemiologic Study of Cystic Fibrosis.** This study, sponsored by Genentech, is a phase 4 (post-marketing) study of Pulmozyme® and is designed to gather information about the routine care of patients with CF, their lung function, and the frequency of lung infections.

### STUDIES RECENTLY COMPLETED

**A Phase 2, Randomized, Double Blind Parallel Dose Ranging Study of Oral TheraCLEC-Total In Cystic Fibrosis Subjects with Exocrine Pancreatic Insufficiency TC-2A.** This Phase II study sponsored by Altus Pharmaceuticals enrolled 125 patients from 26 CF care centers across the U.S. Patients with CF and pancreatic insufficiency were randomized to receive one of three fixed doses of a new enzyme replacement (TheraCLEC) for one month. This study is the only trial of its kind to concurrently evaluate the impact of an enzyme replacement therapy on the absorption of fats, proteins and carbohydrates. Preliminary results showed that TheraCLEC was well tolerated by patients and the trial achieved its primary efficacy endpoint, a statistically significant improvement in fat absorption. Altus plans to release the full Phase II results in October 2005 at the North American Cystic Fibrosis Conference.

*This article was approved by the Institutional Review Board of Children’s Memorial Hospital.*

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## Helping Children Cope

okay to cry, ask questions and talk about feelings. Give your child an opportunity to tell their feelings about what they have experienced. Use this information to prepare for the next healthcare visit.

- Give your child choices: Allowing children to take a more active role in the procedure, such as deciding to sit on the examining table or on a parent’s lap, can lessen anxiety and pain.
- Help your child manage pain: Many coping strategies can be used to help reduce anxiety and perceptions of pain and discomfort (i.e. deep steady breathing, squeezing a ball or your hand, distracting with books, songs, bubbles, etc.).
- Comfort your child: Touch is an important part of healing. If medical needs prevent you from holding or rocking your child, you may still stroke your child

or hold his/her hand. Give lots of praise for achieving even a small success.

- Encourage play when appropriate: Children learn about their world and how to cope by playing. Play gives children control and a way to work out and understand their feelings. Playing with puppets, painting pictures and telling stories before and after a procedure are some meaningful ways to teach children about their health care needs and experiences.

I hope this information can help create a path to more positive healthcare experiences for you and your child.

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*Editor’s Note: Dora has recently been reassigned to the cardiology unit. Children that are hospitalized on 9West will meet Brittney Woodruff, CCLS.*

# Your View

## Contributions from our readers

A very good resource for information and advice about living and coping with CF is each other. “Your View”, a new addition to the *CF Center News*, contains articles written by our readers who want to share information and help others who live with CF. If you have a story, a topic, or a “view” that can benefit others, please contact your CF Team, or send your article to Cathy O’Malley ([comalley@childrensmemorial.org](mailto:comalley@childrensmemorial.org)) or Eileen Potter ([epotter@childrensmemorial.org](mailto:epotter@childrensmemorial.org)).

## And Baby Makes Three...

by Debbie Jackson



Debbie and Madeline enjoying a mommy-daughter hug.

Goals in life...college, career, marriage, babies!!! Three out of four came easy. Babies didn’t. It was an easy decision to make to have children; it was difficult to achieve it. After a year of trying on our own my husband Bill and I saw a fertility specialist. We went through five unsuccessful artificial inseminations.

We took a few years off from trying to regain some sanity and had a multitude of things that came up that took precedence over trying to conceive. However, the inability to conceive naturally and then with medical help started to wear on our minds and souls. Tension and bitterness set in and then life got in the way. Life also got in the way with my CF, which landed me in the hospital for the very first time. I neglected my treatments and didn’t eat well. I was also diagnosed with CF-related diabetes. I had my first 10 day “tune up” at Northwestern Memorial Hospital. The hospital was a true wake up call for me to not let the frustration of not having children and life affect my health. Trust me though the thought was still in the back of my mind.

My health improved and I started to enjoy life again. When things were stable we approached in vitro fertilization and were successful. I was referred to Northwestern’s maternal fetal medicine group and saw them every 2 weeks along with the high risk pregnancy endocrinologist. I saw the CF team every 6 weeks. During my pregnancy my health was fairly good up until the 22<sup>nd</sup> week. I cracked a rib from just a slight cough and landed in the ER. I went in thinking I had a cracked rib and to everyone’s surprise I had pneumonia. When I did my vest I had to unbuckle the last buckle because of my belly and I was missing clearing the lower lobes so mucous built up. I was hospitalized for 5 days and had a PICC line for 10 days. My mother and my husband had to do the traditional CPT on me for the remainder of the pregnancy. The hospitalization

and PICC line were hard for me because I wanted to be able to get through the pregnancy without having the CF get in the way. We really didn’t have proof that CF affected our ability to get pregnant, but like any other CF patient I wanted some sense of normalcy especially when it came to having a baby. Later we came to find that the pregnancy made my bones osteopenic (low bone density), which I now know can be a factor with CF. My pregnancy progressed

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## On Pregnancy And Parenting With CF

As people with CF are living longer and longer, many are contemplating having children. Having children is an enormous undertaking for anyone, more so for parents when one partner has a chronic illness. Many women with CF have successful pregnancies, but risk worsening of their lung disease, new onset or worsening of diabetes, or worsening of nutritional status as they progress through pregnancy. There is also the risk of having a baby with CF which can be decreased by testing the partner for CF mutations. Following delivery there is the challenge of balancing the needs of a newborn with one’s own CF therapies. Our Adult CF Team has taken care of 11 CF couples (9 women and 2 men) through their pregnancies. Needless to say, having a baby is a complicated decision for people with CF and each experience is unique.

In this article, Debbie Jackson, a 29 year old with CF, kindly shares her experiences of having a child. While each individual is different, she expresses some thoughts that may apply to many, if not all, prospective CF parents.

—Manu Jain, MD,

Director, Adult CF Program  
Northwestern University

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## And Baby Makes Three

as did my belly and at 35 weeks my rib cracked again. I stopped working and stayed home for the remainder of the pregnancy.

At 38 weeks I went into labor naturally. However, I think it may have been from the ultrasound technician telling me my baby would be 10 pounds. I was told with the CF-related diabetes my baby may be big. Luckily Madeline only weighed in at a healthy 8 lbs 7 ounces. I was in labor for 24 hours and spent one hour pushing only to have my daughter delivered C-section as a result of her turning while I was pushing. I did have my respiratory treatments while I was in labor. Up until the epidural the treatments were hard but well worth it. I was able to breathe free and clear during labor and then lie on my back during the C-section without coughing.

In all honesty there were no fears of my daughter having CF. My husband was carrier tested before we did any fertility treatments and came back negative. She was so healthy and I just knew in my heart she was fine. She has brought such

joy and happiness in our lives. Even with little sleep. Since her birth I have had minor respiratory infections that were treated orally. I did my treatments religiously and tried to eat as best as I could. I was able to breastfeed my daughter for 9 months. I am truly blessed to have the healthcare team that I do. Without their care and support I could not have gotten through this.

For other CF patients considering pregnancy/parenthood, **make sure your health is in the best shape ever.** It's real easy to forget about yourself when you're taking

care of your baby. It's also easy to want to blow off treatments when you're pregnant, tired and have morning sickness. I am speaking from experience. **Make sure you have a good support system like a spouse, boyfriend/girlfriend and/or family members** who will remind you what it's like to be in the hospital and missing out on life.

When I do my morning treatment I have my daughter in her saucer with her toys, books and favorite TV shows (I have had to sacrifice Oprah for Roli

Poli Oli) to keep her occupied so I can do my vest. However, there have been times when she gets upset from being in there so long and I have had to stand up and dance and sing, with the vest, to make her laugh.

**There are drawbacks though...** It can be challenging when your baby is teething, screaming and not sleeping, and you're congested, want to sleep, and feel miserable. However, you remember you and your significant other brought this child into the world and when you go in to their room and they look up at you with the toothless grin it makes it all worthwhile and you wouldn't change it for the world.

Last but not least...**get help when and where you can.** Remember you can't do it all on your own. Have someone else get up with the baby or do the work around the house, or watch the baby while you do your treatments. Let the dust go another week. Your life revolves around this bundle of joy. Was it a piece of cake...no way! Would I do it again... in a heartbeat!!!!

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## Vacationing with CF

by Brenda Ruf

After a family member has been diagnosed with cystic fibrosis, taking a family vacation may be the last thing on anyone's mind. Eventually, as the months of doctor visits, treatments, medicines and tests roll by, and as the family begins to adjust to the new order of things, a family vacation may be just what everyone needs.

There definitely is an art to taking a family vacation with a child with CF. Making sure that the treatments and medicines are administered without making CF the center focus of your family vacation takes some planning. In the 3½ years since our youngest daughter Eleanor was diagnosed with CF, we have taken several trips. With each vacation adventure Ellie gets a bit older, we parents get a bit wiser, and as a family we all get better at having fun. What follows are some vacationing tips we have collected over several family outings.

Keep it short ...

A weekend at a nearby resort makes an ideal first trip out. Our first family vacation with Ellie was an overnight hotel stay with amusement park admission, a package that my husband had won in a raffle at his company picnic. Our hotel was no more than 10 miles from our home. This short trip allowed us to figure out how to pack medications, how to fit in treatments, and to determine what unexpected extras we might need. Knowing that we could easily run home for something took the pressure off, and allowed us to really enjoy ourselves as a family.



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## Vacationing With CF..



Ellie, Elizabeth and John in their pop-up camper.

### ...and “suite”

Overnight vacationing with CF requires the following basic necessities:

1. Electricity (a grounded outlet if you use The Vest)
2. Facilities to boil your nebulizers
3. Reliable refrigeration

Refrigeration and facilities to boil water are not always available in an ordinary hotel room. Many vacation spots have family suites that typically have kitchen facilities, including a refrigerator and stove. These suites are ideal for CF care on vacation. Call ahead to make certain which amenities you can expect in your room.

### ...or go RVing

Take your kitchen with you! Do your airway clearance under the stars! Count the calories in a S'more! This is our family's favorite way to take a trip. We purchased a pop-up camper a year ago, and we were amazed at how well suited it is for CF care. Our tiny little pop-up has two gas burners, a sink, a small refrigerator, and an electrical hookup. We can do manual clearance on one of the bunks, or Ellie can use her vest. You always know you'll have what you need, and it'll be just as clean as you left it! (Just make sure your campsite has electricity available.)

Note: State Parks tend to be much more secluded, less smoky, and at \$10-20/night, less expensive than private campgrounds. To minimize smoke exposure avoid campfires as they are being built or as they are dying out, as these are the times when the most smoke is generated. Everyone should always stay upwind of a campfire.

### ...but plan ahead for CF care on the road!

A stress-free vacation means easy access to medications

and treatments when and where you need them.

**Enzymes** Long boring car trips are perfect for packing in a few extra calories. Bring many more enzymes than you anticipate you will need, and pack them in more than one spot, in case you misplace a bag or purse. Make sure you have applesauce and spoons handy if you need them. When Ellie was a toddler we always had a few plastics spoons in the glove compartment, and a few in my purse. I also kept some ketchup packages around just in case we ran out of applesauce. Ellie always sat in the seat just behind the driver, and I'd prepare her applesauce and enzymes and give them to her from the front passenger seat. Older children might keep their own enzymes in a fanny pack or purse. Remember not to expose enzymes to extreme heat.

**Refrigerated Medicines** Keep the Styrofoam coolers and ice packs from your Pulmozyme® delivery for use on vacation. If you start with a frozen ice pack, the cooler can give you as much as 24 hours travel time between refrigerators. These coolers are also a good place to keep applesauce after it has been opened

**Nebulizers** Take as many nebulizers as you have doses per day, and take really good care of them. I once allowed the water to boil down and was horrified to see that I had melted all our nebulizers just hours before we were to leave for the Black Hills. Times like these are when you appreciate your resourceful CF Team the most. (Thanks Carolyn!)

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***If you prefer not to clean and disinfect your nebulizers, then pack disposable nebulizers, which can be thrown away after use!***

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**Airway Clearance** Don't you worry, airway clearance *will* fit in your busy vacation schedule. I wasted a lot of time worrying about this before our first vacation, and it turned out to be an absolute non-issue. There is always a little downtime between activities, and most likely your family will be more than happy to cool their feet for the duration of an airway clearance session. Sure, the schedule might vary a bit from what it is at home, but you'll get them in.

Above all, whatever you do, and wherever you decide to go, don't forget to bring your CF Team's contact information... so you can send them a postcard! Think positive and have fun!

# Meet Our Staff



Dr. Jain at the Loire Valley South of Paris

**Manu Jain, MD** is Director of our Adult CF Program at Northwestern University (NU). Dr. Jain began his medical education at NU where he received a Bachelor of Arts degree in Biochemistry. He completed his medical degree, internal medicine internship and residency, and Pulmonary & Critical Care Medicine fellowship at the University of Chicago. In 1997, Dr. Jain became a member of the Pulmonary Division at NU and his specialties include CF care and the Intensive Care Unit. In 1998, Dr. Jain teamed with Joanne Cullina, MSN, RN, and they launched the Adult CF Program.

Dr. Jain is committed to CF patient care, research, and teaching. He and his team help their patients balance their CF care with the many challenges of young adulthood, such as college, employment, family planning, pregnancy and parenthood. Dr. Jain finds working with the CF team and helping patients face these challenges very gratifying. He has a proactive approach to CF care, in which he feels that preventive steps are well worth the effort, and he welcomes new therapies with proven clinical benefit.

Dr. Jain recently received a Master's Degree in Clinical Investigation from NU Graduate School. His interest is in translational research – that is, taking information gained from laboratory bench research and studying its impact in the clinical setting. One example of this type of research is the *Pseudomonas Aeruginosa* Type III Secretion Study. In addition, he is studying sputum in the adult CF population for the presence of mediators that may contribute to scarring of lung tissue.

Dr. Jain is a new member of the CF Foundation's Center Committee. One of the tasks for him in this position will be conducting accreditation visits of other CF Care Centers in the United States. Other interesting endeavors of Dr. Jain's include being a reviewer for many pulmonary critical care journals and an editor of the web-based critical care journal of the American Thoracic Society.

Beyond the university, Dr. Jain enjoys an active social life. He and his wife like to travel and have taken memorable trips to far away places, such as South Africa, Tahiti, India and Bali. Dr. Jain is also an avid runner, and has played *Ultimate Frisbee* since his college days. Ask him if he is competitive, and he says he plays for fun, but "It's always more fun to win!" Despite this, along with many of his fellow Chicagoans, he remains a "long-suffering" fan of the Chicago Cubs!

## Staff Update

Warm best wishes to **Emily Griffith, RD** as she leaves Chicago and relocates to Dallas this summer to be closer to her family. Leaving the CF Center was a difficult choice for Emily and a sad one for us, but we wish her well. **Megan Hart, MS, RD** joins us in October from Phoenix Children's Hospital where she was a pediatric nutritionist involved with the multidisciplinary care of chronic illness. **Wendy Estrellado, MD** completed her Pediatric Pulmonology Fellowship and will head to Philadelphia to complete the U.S. residency requirements – we wish Wendy continued success! **Oren Lakser, MD** has accepted a position at the University of Chicago where he will continue with his research, and be on staff in their Pulmonary Division. We will miss Dr. Lakser and take comfort in knowing he will be close by. We are happy to welcome **Juan Pablo Jorba, MD**, who joins our Pulmonary Division as a Pulmonology Fellow, and **Cheryl Ma, RN** who joined our CMH nursing staff this spring. Congratulations to **Kim Watts, MD!** She has a new addition to her family; a baby boy named Augustine! The Adult CF Team welcomes **Melissa Hewitt, LCSW**. Melissa comes to us from the inpatient side at NMH. She has been a social worker for 8 years, primarily working in neuroscience, cardiovascular and orthopedics. This is her first experience working with CF and she is enthusiastic about her new role.

## New Director of Human Molecular Genetics Program is International Expert in CFTR



**Ann Harris, Ph.D.**, has been named director of the Human Molecular Genetics

Program at Children's Memorial Research Center (CMRC). Dr. Harris is a full professor at the University of Oxford with more than 130 peer-reviewed articles to her credit. Her research focuses on the regulatory domains and mechanisms of the human cystic fibrosis transmembrane conductance regulator (CFTR) gene and potential applications to novel therapies for CF. Dr. Harris will join Children's Memorial in September.

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