# Cook Children's Cystic Fibrosis Newsletter

Special Edition


SPECIAL EDITION NEWSLETTER: NACFC 2014

## WORLD'S LARGEST CYSTIC FIBROSIS MEDICAL CONFERENCE KICKS OFF IN ATLANTA

The 28th annual North American Cystic Fibrosis Conference (NACFC) in Atlanta was a gathering of over 4000 researchers and healthcare professionals. We were all there to share and learn with the goal of improving the lives of everyone affected by Cystic Fibrosis. It was a great conference!

This year, Cook's CF Center is bringing you some of the highlights of that conference in a new way. Many of the staff who attended the conference have provided a short summary of a session they attended. We hope you find this helpful and would love to know what you think about getting the conference information this way

CYSTIC FIBROSIS FOUNDATION

My favorite session of the whole conference was the first plenary session (that's where they put all 4000 of us in the same room!!). The title was Scaling the Mountain: The Journey to delivering Transformational CF therapeutics. Mouthful, right? Dr. Boyle talked about climbing Mt. Everest and that each of the significant milestones we reach in CF care is like reaching the each of the basecamps on the mountain.

He ended the talk with the commitment to finding treatments to help each kind of mutation/defect (you know, that "transformational therapeutics" thing.) He reviewed each of the different types of defects and what treatment is available or where we are with the research. He even talked about the next steps we need to take to "summit" the mountain.

The second plenary was CF Microbiology Past, Present, Future . Dr. Dyson will talk a bit about that in his article.

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You can find all of the "Plenary sessions" and other sessions at: https://arc.nacfconference.org/cff/live/2014nacfc# . You will need to register but there is not a fee involved.

You may find the Saturday sessions especially beneficial as they were developed to include families. They include information about the new infection control guidelines, the role of exercise in CF and ways for patients, parents and families to help their centers to improve care.

Enjoy,
Carrie

# Introducing Exercise as a Recommended Airway Clearance Technique (ACT)

A large group of studies on exercise have proven to benefit patients with Cystic Fibrosis (CF). There were several speakers on this topic at the conference.

Exercise can provide improvements to quality of life in CF patients, with benefits including an increase in sputum expectoration and a reduced rate of decline in pulmonary function. In addition, exercise training is known to improve fluid balance; therefore exercise training has the potential to improve the fluid and salt loss due to sweat gland dysfunction in CF patients.

To aid in increasing physical activity during hospitalization, the inpatient unit (PCU) at Texas Health Fort Worth Hospital recently purchased a stepper, stationary bike, resistance bands, foam rollers, and CF certified Yoga DVDs. Now the adult CF patients will not only have the opportunity to stretch and exercise with our specialty trained Physical Therapist, but will also have the capability to exercise any time in their room. With the new equipment, our goal is to educate our patients and their support systems on the importance of an exercise program and provide an alternate mode of therapy to improve their lung function, airway clearance and quality of life.

Sean Clark, RRT
Clinical Coordinator- Cystic Fibrosis & Cardiac Teams
Texas Health Harris Methodist Fort Worth Hospital

Ashley Hodo, MSN, RN
Nurse Manager
Texas Health Harris Methodist Fort Worth Hospital

## Fertility and Sexuality Issues for Adults and Men with CF – Sexual Reproduction Health in Pediatrics (CF)

There is a need to know for both parents and young males with CF the probability of infertility, with that percentage being at 98%. According to surveys, the average age that this is discussed is at 17.4 years of age; however, it has been shown that adolescent boys would prefer to learn more about this at the age of 14 years of age with the most appropriate age being at 13.8 years of age. Patient knowledge should include how one underestimates the prevalence of infertility; confusion between what infertility is and being impotent (infertility does not mean being impotent); and understanding of pathophysiology. Both the CF team and parents of these patients should start the conversation early and attempt to eliminate any barriers that would arise in communicating this information such as: privacy, embarrassment, lack of time and insufficient training. Based on this information, the Cook's CF center provides a pamphlet to parents early in adolescence. That same information is provided to the teen soon thereafter.

Laura Madrid, BSN, RN

# Screening for Depression and Anxiety

-Brooke Tomlin MSN, RN

Many clinics are starting to do routine screening for depression and anxiety. The main group that they would like to focus on is teenagers (ages 14-19). There can be many obstacles to screening in this age group: teenagers not answering correctly or feeling as if they cannot answer truthfully due to a parent or sibling being in the room. One way some clinics have approached this is to offer a screening questionnaire on a sheet of paper that is only given to the adolescent upon arrival. It could also be offered when the patient has their partial independent visit. Both of these methods may provide the privacy needed for the teen to answer the questions more honestly. Then additional screening would be given to determine what the teen needs to help with any issues they may be facing. In a perfect world, each clinic would have a mental health professional on hand to start right then and there so that the patient does not have to wait too long to get in to see someone. Unfortunately, not all clinics have that resource but most have mental health professionals available.

Ultimately, the goal is to have patients develop coping skills and tools needed to deal with the everyday things that they encounter. The CF Foundation is working with CF care centers to improve recognition of mental health issues so the appropriate referrals can be made.

# PSEUDOMONAS AERUGINOSA:

## Let's get rid of that bug!

Staci Ballew, BSN, RN


Pseudomonas aeruginosa (PA) is a type of bacteria that often lives in the lungs of people with CF and causes lung infections. The first PA infection:

1. Generally gotten from the environment (not spread form patient to patient).
2. Present at low levels
3. Highly sensitive to antibiotics
4. "Window of opportunity" to eradicate before the development of chronic infection!

What do we mean by "eradication"? It means we want to get rid of the bacteria. Research has shown that the longer the lungs free of PA, the healthier the lungs will stay.

Current guidelines of care stress finding early and treating early with antibiotics. This is why we culture at most clinic visits and all hospitalizations. Early eradication approaches have included inhaled, oral and IV antibiotics. Sometimes they are used alone or in combination which in general have shown similar eradication rates. Inhaled tobramycin is the most widely recommended treatment and has been reported to work well at clearing PA. Always speak with your pulmonologist, CF coordinator or nurse for specific treatment plans for your child.

# Bugs, Bugs, and More Bugs

One of the plenary speeches was the history of microbiology in CF but in fact it looked into the future as well. This and other presentations look forward to a changing way of viewing the organisms in the lung of the CF patient. Historically the sputum or swab of patients has been smeared on an agar (gelatin) plate and as bacteria start to grow and form colonies they are picked off, identified and tested against antibiotics. This means days of waiting, then identifying those organisms that someone chooses to collect either because they are numerous or recognized as known pathogens.

While this method has taken us far, a new method is coming. Like CSI, the microbiology lab is turning to DNA. Already the lab is using DNA probes to identify some organisms such as MRSA, Mycoplasma and Pertussis. It is anticipated that in the future sputum will be analyzed looking for the DNA of organisms. This will have two benefits. One is speed since the lab will not have to wait for bacteria to grow and the DNA also contains the information to tell which antibiotics the bacteria may be resistant to. The other is as this process is improved labs will be able to identify more organisms. The focus is now shifting to not just a few "pathogens" but thinking of the whole community of organisms or the "biome." The idea is the health of the lung, like we've heard about GI health, may depend not just on one or two organism but the entire microbiological community in the lung.

In the distant future, one company is working on a device that will allow a drop of sputum to be placed on it at the bedside and in a few minutes it will check to the DNA of hundreds of organisms. Of course the point of this is that with more rapid and more complete information about the biome of the lung better therapies can be devised.
-Maynard Dyson, MD

# Antibiotic Use and Taking Medications

The 2014 North American Cystic Fibrosis Conference welcomed pharmacists from a variety of national and international cystic fibrosis centers, academic settings, hospitals, clinics, and research sites around the world. The conference provided many opportunities to exchange a wealth of information on the latest products, research, services, and advances in CF care. Two areas of interest highlighted at this year's CF pharmacist research sessions include:

- **Improving antibiotic use in the CF population** – Advances in CF care mean CF patients are leading longer, healthier lives. As patients are exposed to more antibiotic courses throughout their lifetime, bacteria can become resistant to currently available antibiotics, making them less useful to treat infections. By studying the effects of many different antibiotic doses, pharmacist researchers are taking necessary steps to ensure that patients will continue to receive antibiotic doses that are most effective at killing bacteria while ensuring that resistance to antibiotics is minimized. Additional antibiotic research study information can be found at www.cff.org.

- **Medication adherence support for patients and families** – Medication adherence is defined as the extent to which patients take medications as prescribed by their health care providers. There are many reasons that keep patients with CF from regularly taking all of their medications as prescribed, including insurance or co-pay issues, problems accessing specialty pharmacies, medications not carried on formulary, prescriptions needing prior authorization, taste issues , time-constraints, etc . Improving adherence is key to improving outcomes in CF. Pharmacists are in a unique position to be able to help patients identify and address many of the barriers to medication adherence. Studies addressing how to best assist patients with adherence are currently ongoing. Additionally, many CF centers now employ a dedicated CF pharmacist to help better meet patient needs.

- Denise Pinal, PharmD

# About Clinical Trials

by: Trudy Morris, RN, CCRC

## Cystic Fibrosis Foundation and Cook Children's Research GOAL.

The driving force behind the CF Foundation and our research here at Cook Children's is as follows:

- To find new medicines to fight CF
- Partner with the most successful and innovative biotech companies and scientists in the world.
- Ultimately to cure this disease!!!

## Learn. Ask. Join.

Today, there are approximately 30 potential drugs in development for the treatment of cystic fibrosis. These new therapies can only be made available to the people who need them after they are tested by individuals with CF in a series of clinical trials. Clinical trials that test potential new drugs and therapies to see how well they work are a major part of CF research. People with CF can play a critical role in the development of new treatments and the search for a cure by participating in clinical trials.

## Research Cannot Move Forward Without Your Help.

To help promising drugs move swiftly from the research and testing phase to the people who need them most, many people with cystic fibrosis are needed to participate in clinical trials.

- ***Learn*** about CF clinical trials
- ***Ask*** your care center team about participating in trials.
- ***Join*** the search for new treatments and a potential cure.

## Cook Children's Medical Center Pulmonary Research Updates.

Cook Children's Medical Center CF Foundation Therapeutic Development Network is participating in several research trials including the following:

- VX12 809 104 and 105 Study-- Combination therapy (lumacaftor/ivacaftor) to evaluate the safety and efficacy in homozygous DeltaF508 mutation
- BONUS (baby observational and nutritional study)
- Star too study which looked at rather to treat or observe subjects that were newly colonized with methicillin resistant staph aureus
- TIP 4 which examined the safety and efficacy of inhaled tobramycin powder
- VX12 770 115 study to assess ocular safety of ivacaftor
- CF Registry
- CFFC (fibrosing colonopathy) observational safety study to review the incidence of and risk factors for fibrosing colonopathy in US patients with cystic fibrosis treated with pancreatic enzymes
- EPIC Observational Study 11-15 years which will collect specimens to increase the biorepository
- VX14 661 106 study which is currently in the start- up phase to look at efficacy and safety of the 661 in combination for the homozygous DeltaF508 mutation.

### CF stands for "CURE FOUND"

We are proud of our numerous research accomplishments. Our center was designated as a Therapeutic Development Center in January of 2009. We have focused on growing our research culture and have made great strides in becoming a significant contributor to CF research. Our CF patients and families continue to be enthusiastic regarding participation in research. We are devoted to steadily increasing research efforts at our facility.

It is an exciting time in CF Research. There are more potential therapies to treat CF in development today than in the entire history of Cystic Fibrosis Research. We must recruit more people than ever before to help us test new drugs.

**Without patient volunteers—without people like you—research and progress are not possible**.

# URINARY INCONTINENCE IN THE CF PATIENT

Physical therapists involved in CF care are finding that one of the most common, yet least addressed problems in CF is stress urinary incontinence (UI), AKA, leaking urine when you cough (or sneeze, laugh, or jump)! It can occur in girls as young as 5-6, and roughly 33% of adolescent girls with CF have it. Males are affected too, though less frequently. It becomes more common with increasing age, with up to 80% of adult women with CF reporting UI. Even mild symptoms can affect daily function, including social activities and airway clearance efforts. If you notice crossing of legs and/or weak effort with PFTs, or having to use the toilet before airway clearance or PFTs, UI is probably the culprit! Imagine the muscles between your pubic bone and your tailbone as a sling or hammock. They are always active to "hold in", but have to work harder to overcome the pressure of forceful coughing. After constant stress, something *has to give*, and eventually these muscles are too weak to overcome this force. Bladder irritants like caffeine, carbonated drinks, milk and dairy products, and certain fruits can make it worse. Help is out there! Physical therapists are trained in basic assessment and education strategies, but there are specialists who can target treatments for people with UI. Your clinic has handouts with more information.

In other news, we continue to see research rolling out with evidence for exercise as "medicine." It regulates inflammation and improves immune response, airway clearance, quality of life, and more. You can get these benefits with at least 30 minutes of moderate intensity activity at least three times each week!

*Allison Ewing, PT, DPT*
**Physical Therapist**
**(682) 885-2246**
**CookChildren's** Medical Center

**The Cystic Fibrosis Center at Cook Children's Medical Center is one of more than 115 accredited CF centers throughout the nation. These centers are accredited by the Cystic Fibrosis Foundation, a non-profit organization founded in 1955 dedicated to funding research to find a cure for CF and improving the quality of life for people with the disease**

## Pediatric CF Center Staff:

Center Physicians:

- James Cunningham, MD– Co-Center Director (Nurse: Stacy),
- Nancy Dambro, MD– Co-Center Director (Nurse: Jennifer & Karen),
- Maynard Dyson, MD (Nurse: Stacy),
- Sami Hadeed, MD (Nurse: Sharon),
- John Pfaff, MD (Nurse: Jessica),
- Karen Schultz, MD (Nurse: Paulette),
- Shailendra Das, DO (Nurse: Lisa)
- Erin Newman, DO (Nurse: Lisa)

## Adult CF Center Staff:

Center Physicians:

- John Burk, MD– Adult Center Director,
- Jack Gilbey, MD
- Stuart McDonald, MD
- Randall Rosenblatt, MD
- Cyndy Roger, ACNP-BC **(**Nurse: Laura, adult nurse and newborn screening)

CF Coordinators:

- Janet Garbarz, Carrie Stradley 682-885-6299

Dietitians:

- Staci Brummett, Rachel Hamik, Esther Giezendanner 682-885-7496

Respiratory Therapists:

- Deanna Pinckney, Alex Rasmussen, Crystal Thompson, Cindy Corne, Shonda Thompson 682-885-4189

Child Life Specialist:

- Alex Steward 682-885-4892

Social Services

- Amy Wilson 682-885-3991

## Cook Children's Medical Center and Cystic Fibrosis Center

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Making CF stand for "Cure Found"