

- An abstract summarizing the results of the PAAC survey will be presented at the National CF Conference this November in Denver, CO.
- The CF Center is currently enrolling for several CF studies. Ask your provider if you or your child qualify for any of them at your next visit.

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CF News

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PAAC Survey Results

The Parent Advocacy and Advisory Council (PAAC) at the University of MN CF Center developed a survey last Fall to identify key concerns that CF parents and patients have with the pediatric clinic, and implement quality improvements wherever possible.

Of the 198 surveys that were sent, 54 were completed, returned and analyzed. The most common clinic concerns listed by parents and patients included: visit efficiency, infection control, interpretation of lab results, lack of overall CF

education, and staff professionalism.

The PAAC presented these results to clinic administration, and in response the fol-



PAAC members: Front row Karin Shelstad; Suzy Cossette; Jill Christensen; Debbie Tharp; Karen Reigstad, RN. Back row Joe Harris; Jackie Zirbes, NP; Brooks Shelstad; Carlos Milla, MD; Pam Mertz, Lou Mertz.

lowing interventions occurred: clinic visit efficiency was improved, a new protocol was developed for cleaning clinic space and exam rooms, a quarterly newsletter (ta-dah!) addressing labs and patient education was created, and new staff was hired at the front desk and given additional training.

The PAAC would like to thank each and every one of you that took the time to give your feedback. With your help, we will continue to make improvements to our clinic.

Bacteria of the Day: *Staphylococcus aureus*



Staphylococcus aureus, also known as Staph or S. aureus, is often the first bacteria, or pathogen, to infect the respiratory tract of CF patients. Before antibiotics were available, Staph infections significantly shortened the life expectancy of infants with CF. Now that so many great antibiotics are available, the average life ex-

pectancy of patients with CF has increased to 36.8 years!

Different strains of S. aureus spread easily within families with or without a member with CF. However, it is not uncommon for CF patients to be infected or colonized with the same strain of Staph for at least 1 to 2 years. In fact, for children under the age of 6, it is not uncommon to give prophylactic antibiotics to prevent infection with Staph.

Transmission of Staph is person to person, therefore good infection control practices reduce the spread of this bacteria. Ways to prevent the transmission of Staph include: good hand hygiene (keep your hands clean), keeping your linens, towels and clothing clean and dry, never sharing personal items (such as razors, toothbrushes, towels), and taking care of a Staph infection with antibiotics.

Infection Control Improvements



Josh Olson

After 6 years of service, we announce Josh Olson's departure from the CF Center Team. Josh decided to take a new job opportunity, and he will be greatly missed. The CF Office has hired Tracy Killoren to fill the open position, but Josh will never be forgotten. BEST WISHES, JOSH!

The Pediatric Specialty Clinic continues to work on improving the experience for children and families that come to the University of MN CF Center for care. Although the CF Center infection control procedures have always met or exceeded the Cystic Fibrosis Foundation guidelines, the results of the PAAC survey indicated that infection control in our clinic waiting room and exam rooms is a big concern for parents and patients. Because your feedback matters to us, we have made the following changes to our infection control procedures:

- The chairs in the waiting area have been rearranged to prevent crowding.
- The chairs are wiped off with a disinfectant each morning before clinic begins.
- A Respiratory Hygiene Station has been ordered for the waiting area that will hold hand sanitizing foam, Kleenex, "Cover your Cough" signs, and masks.
- The wall toys in the lobby and the children's table and chairs get wiped down with a disinfectant each morning.
- The climb-on-animals, mirror, books, end tables, lobby phone, door handles, water fountain, and television are disinfected each day.
- All toys brought into the exam rooms by Child Family Life to entertain/distract your child have been cleaned thoroughly and wiped down with a disinfectant before each use.
- The front desk staff disinfects the registration desk areas, keyboards, mice, phones, clipboards, and pens frequently throughout the day with single-use sanitizing wipes. In particular, staff will wipe all of these areas after a child with respiratory symptoms has checked in.
- The pulmonary function waiting area, including the water fountain, chair arms, and countertop with brochures, is wiped down with a disinfectant daily.
- When a patient has a known infection, this information is passed on to the clinic providers, increasing awareness and assessment for using contact precautions.
- The nurses wipe off exam tables after each patient. After new white exam table paper is pulled for the next patient, the nurse also places a sign on the bed stating: "This room has been cleaned and is ready for use."
- We cleaned the vents and carpets, and although we have seen an overall improvement, we still plan to continue to work on this.

- We are looking forward to a clinic facelift later this year. We should see new carpet, fresh paint, and a friendlier environment!

With all of the changes that have been made, there is only one thing left for you to do. Although all clinic providers have been thoroughly trained to use sanitizer hand foam before or after entering an exam room, if you don't see them do so, you need to ask whether or not they did. It is critical to all of the clinic providers that parents and patients feel comfortable with the care received, so you will never offend them.

We welcome any additional comments or suggestions you may have. Please let us



Look around the clinic to see the infection control improvements that have been made

know how we can continue to meet your needs.

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Classes of CF Mutations

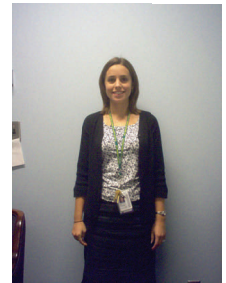
Upon diagnosis, a blood sample is drawn to determine a patient's CF mutations. Over 1400 different CF mutations have been discovered thus far. The production of a protein called CFTR, which is found in the cells lining the nose, sinuses, esophagus, lungs, stomach, intestines, pancreas, liver and reproductive tract, is 'broken' in people that have 2 CF mutations. This leads to overly thick, sticky mucous production in all of these areas of the body.

There are 5 different ways CFTR protein production can 'break'; therefore there are 5

different classes of CF mutations. In classes 1, 2, and 3 the CFTR protein is not produced in the cells, or it is unable to make it to the cell's surface where mucous production and regulation takes place. Since classes 1, 2, and 3 keep the CFTR protein from doing its job (making thin and viscous mucous), these classes are typically associated with lung involvement and pancreatic insufficiency. Some common examples of these mutations include: G542X, DeltaF508, and G551D. In classes 4 and 5, the CFTR protein is produced in the cell

and makes it to the cell's surface, however the cell's surface is unable to properly regulate 'chemicals' (sodium and chloride) the body needs to produce 'good' mucous. Since the CFTR protein is produced and able to do its job to some extent, class 4 and 5 mutations are usually associated with less lung involvement and some degree of pancreatic sufficiency. Examples of these mutations include: R117H, A455E and 3849+10KbC>T.

If you have questions about CF mutations, please contact Amy Powers at 612-624-5721.



Amy Powers is the genetic counselor for the U of MN CF Center

Message From the CF Office

Ever get frustrated when you try to contact the CF Center Office and are unable to reach a live person? Although Karen and Evelyn wish they could personally answer each and every phone call, it just isn't possible. With over 500 patients and more than 100 phone calls received each day, Karen and Evelyn must triage, or prioritize, the calls they receive. So, which calls are given which priority?

Highest Priority: Sick patients always have and always will get first priority! If a patient is sick, make sure you leave a message stating what the symptoms are, and how long they have been present. Patients that need to be seen right away, or that need IV medication, will get the quickest response times.

Moderate Priority: Issues that

take second priority include: changes in health status (increased cough, colds, adjusting enzymes, etc), laboratory test results, medication changes, insurance issues, and school communications.

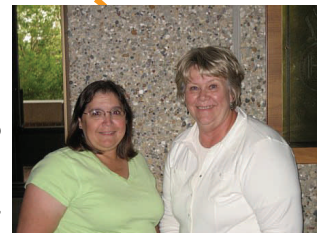
Lowest Priority: Issues that hold the lowest priority, and will therefore take the longest to respond to, include: prescription requests, faxes for prescription renewals, prior authorizations for medications not covered by insurance, and various forms that need to be filled out.

In order to help Karen and Evelyn operate at maximum efficiency, try to follow these tips:

- Leave a voicemail message. Your feedback is important, that is why voicemail is checked frequently. Karen

or Evelyn will get back to you.

- Make appointments for future tests and visits as you leave the clinic each time. If you do not do this, routine scheduling unnecessarily falls into Karen and Evelyn's hands.
- Give your pharmacy, Karen or Evelyn plenty of advanced warning when you need a new prescription or refill.
- Give at least 3 weeks advanced notice for any letters needed (home health forms, travel letters, letters to insurance companies, Tefra updates, etc).
- Bring school forms (usually permission to give meds in school) with you to clinic so they can be signed while you are there.



Evelyn McKee and Karen Reigstad

"We hope some of this information is helpful to you, and increases your understanding of why we need things like voicemail. We really appreciate your patience as we work together to keep your child and family healthy."

Karen Reigstad

Understanding Lab Results

Sure, we know a lot of tests take place come check-up time, but what do all of the tests mean?

Throat or sputum culture: Throat cultures check for germs, also known as bacteria, that may be present in the airways of the lungs. Following a good cough, the bacteria present in the back of the throat, if any, is collected and sent to a lab. The lab tests the sample and identifies which bacteria are present and how certain antibiotics affect the bacteria found. By doing this, the CF Center learns which antibiotics the bacteria are sensitive to, and thus knows which antibiotic to use for treatment. This process can take just a few days for bacteria like beta strep (strep throat), or up to 2 weeks for pseudomonas. After the doctor or nurse practitioner has looked at the throat culture results, any orders for medication changes are written in the medical chart and given to a nurse. The nurse will call you with the results if a medication change is required. If no changes are required, the results will be mailed to you. If your child has an increased cough or isn't well when you are in clinic, you may be told to call the CF Center in a few days to check on the throat culture results, or you may be

given medication before the results are available. The CF airways are frequently colonized by bacteria, therefore if a throat culture shows an organism is growing, it does not mean you or your child have a pneumonia or lung infection. A positive throat culture indicates airway colonization that needs to be treated with antibiotics to keep the bacteria count low; it does not mean you or your child are in any immediate danger.

Additional Tests: At times additional tests may be ordered during a routine visit. For example, if you or your child are not well, blood tests that check for infections and the body's response to infections may be performed.

Annual Labs: Although CF primarily affects the respiratory and gastrointestinal (GI) systems, it is considered a "multisystem" condition. Therefore, the purpose of annual labs is to check all systems of the body in an attempt to find problems early and intervene quickly.

Oral Glucose Tolerance (blood test): From age 6 and on, glucose levels are monitored since we know that CFRD (Cystic Fibrosis Related Diabetes) affects 30-40% of adult patient and 15% of adolescent patients.

Nutritional Markers (blood test): Since good nutrition plays an important role in CF, the amount of albumin and total proteins is monitored.

Liver Function (blood test): Approximately 15% of CF patients can develop liver disease, therefore Alkaline Phosphatase and AST levels are checked.

DEXA scans: DEXA scans are performed to study bone density and check for malabsorption of Vitamin D and Calcium, or changes in bone density due to steroid use.

Lung Status: We perform a chest x-ray, aerosol scan, and/or CT scan to monitor lung status in our patients.

All of the Annual Lab results are received and reviewed within approximately one week. The doctor or nurse practitioner is very watchful when it comes to abnormal results, and will write new orders for medication changes in the medical chart promptly. The chart will be given to a nurse that will contact you with the changes. If there are no abnormal results, you will only receive the lab results in the mail.

If you ever have questions about lab results, or feel that you or your child need treatment, please call the CF Center at 612-625-5995 to speak with a nurse.



DEXA Scan

DEXA Scans use a fine thin beam of x-rays to measure bone density, typically of the hips and lumbar vertebrae. The procedure is painless and lasts about 10 minutes. The test is important in assessing the degree of bone thinning in osteoporosis. This test is useful because, as they age, most CF patients develop some degree of osteoporosis.

CAM Meets CF

Complimentary and Alternative Medicine (CAM) includes a wide variety of treatments available to patients worldwide. These can be separated into biomechanical (massage, osteopathic manipulation), lifestyle and mind-body (environmental changes, nutrition), bioenergetic (acupuncture, therapeutic touch, prayer) and biochemical (herbs, nutritional supplements) treatments. Many of these therapies can compliment the traditional therapies prescribed by your doctor and should never replace them without discussion. It is estimated that up to 60% of CF patients/families utilize CAM. We will focus on herbal therapies to make you aware of the possible benefits and risks involved.

Herbal therapies have been utilized to prevent and cure disease throughout history. Echinacea had been used by American Indians to treat snake bites and colds. Many of our modern medicines derive from herbs. For example, Digoxin, a heart medicine, comes from the foxglove plant. Unfortunately, many of the herbal supplements available to us have

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not been studied to evaluate their effectiveness. In fact, since many of these supplements are considered food additives, they are not regulated by the US Food and Drug Administration. Currently, this billion dollar industry is based primarily on the historical uses of these herbs as well as subjective assessments of their efficacy.

Beware of claims regarding miracle cures for you or your child's ailments. Although there are many potential benefits, herbal supplements act as medicines and may interact with CF medicines. Herbal therapies contain complex chemical ingredients, and many are yet to be identified. Thus, it is very important to discuss all supplements with the nurses and doctors when you come to your regular clinic visits!

The CF Foundation has recognized the potential benefits of herbal therapies and has supported research projects in their fight to cure and control this disease. One of these studies examines the effects of curcumin (a substance found in the spice turmeric) on the

CFTR gene. These are promising therapies and more research needs to be completed before their use becomes recommended and widespread. Other therapies that patients have asked about in clinic include garlic, ginseng, dandelion, chamomile and nettle. Garlic and ginseng appear to have interesting antibacterial properties, dandelion has high levels of vitamin A, chamomile is a mild sleep aid, and nettle has been used as an allergy remedy. Many of these therapies may sound enticing but there are no recommendations to go out and start them, especially in children! I encourage you to ask about therapies so that we can work as a team to make sure there are no dangerous interactions with other CF medications before starting something new. These are complimentary therapies that should not replace the routine medications that are the standard of care for CF. Please keep an open mind and an open ear since a new therapy may be in your kitchen cabinet. Hopefully, we will find it in a scoop of ice cream!

Q & A With the Experts

Q: My daughter was diagnosed with CF a year ago; is there any way to predict whether her course with this disease will be 'mild' or 'severe?'

A: Since there are over one thousand different CF mutations, we are not able to definitively predict the severity of the clinical course. However, based on the different classes of CF mutations, we can provide information on the symptoms that are typically associated

with a particular mutation. In addition, studies show that poor pulmonary function that cannot be stabilized, malnutrition, colonization with pseudomonas, diabetes, and the female sex can be associated with a more severe (lung and gastrointestinal involvement) clinical course.

Q: I always have a lot of questions for the doctor when I come in for my children's clinic visits. When would the doctor prefer I

ask them?

A: It is helpful for the provider to know your questions at the beginning of the visit. This way, the answers can be incorporated as the visit progresses, which results in a better understanding for both the patient and family. Several families write their questions on the Interval History Form that is handed out by the front desk staff. Item #15 on this form asks if there are any questions for the provider.



**Jackie Zirbes, NP
from the University
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University of MN CF Center

Our mission is to provide diagnosis, care, teaching and research for cystic fibrosis.

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PAAC VISION

To be a body of parents advocating and advising the University of MN Pediatric CF Center on the practice of family-centered care from the patient and family perspective.

PAAC MISSION

To promote family-centered care and to advise the University of MN Pediatric CF Center and the University of MN Children's Hospital Fairview administration, faculty, and staff on patient and family needs and priorities.

WHAT'S NEXT?

We need your feedback! Let us know what you think the PAAC should focus on to improve care at the University of MN Pediatric CF Center and the University of MN Children's Hospital Fairview.

MARK YOUR CALENDARS	
JCs Wild Game Dinner	10/09/06
National CF Conference	11/01/06-11/05/06
Breath of Life Gala	11/04/06
CF Education Day	11/18/06



Newborn Screening

As you may already know, the Minnesota Department of Health (MNDOH) recently added cystic fibrosis (CF) to the list of disorders tested for in its newborn screening program. We are proud to have been involved in the effort to have CF added to newborn screening. We believe there is already strong and compelling evidence that demonstrates the great benefit that children with CF and their families get from early diagnosis and treatment.

It is important to remember that the screening strategy currently available for CF should be seen as

a first step in the detection of infants possibly affected, since the test is not diagnostic. Thus, infants identified as possibly affected by CF should undergo definitive diagnostic testing at an accredited CF Center according to the recommendations set forth by the Centers for Disease Control and Prevention (CDC).

The University of MN CF Center, building on its longstanding tradition of excellence in CF care, has developed a comprehensive program for the diagnostic evaluation of infants identified through newborn screening. This program includes a comprehensive thera-

peutic program for those infants that are ultimately diagnosed with the disease, and genetic counseling services to help families that discover they are carriers of the disease.

Our ultimate goal is to be a resource for comprehensive care for those affected infants and their families, as well as to be a resource for carrier families and the community at large.

Since the newborn screening program began in March of 2006, 7 infants have been diagnosed with cystic fibrosis in the state of Minnesota.