TWO NEW PEDIATRIC PULMONARY FELLOWS

We are pleased to welcome two pediatric pulmonary fellows to the CF Center. A fellow is a physician who has completed a resident training program. Fellows receive additional training in pulmonology in order to care for cystic fibrosis patients and other pediatric pulmonary diseases.

Don Hayes, MD

Dr. Hayes is originally from Kentucky. He received a Bachelor of Science in Biology and Chemistry from Morehead State University in Morehead, Kentucky, and received his Medical Degree from the University of Kentucky College of Medicine in Lexington, Kentucky. He completed a four-year residency in combined Internal Medicine and Pediatrics at the East Carolina University Brody School of Medicine in Greenville, North Carolina. Dr. Hayes is based full-time in the Pediatric Pulmonary and CF Center and will be completing a three-year pediatric pulmonary fellowship.

Jonathan Spahr, MD

Dr. Spahr attended Dickinson College in Carlisle, Pennsylvania, receiving a Bachelor of Science in Biology. He received his Medical Degree from Jefferson Medical College in Philadelphia and completed a four-year combined training program in Internal Medicine and Pediatrics at the Geisinger Medical Center in Danville, Pennsylvania. Dr. Spahr is pursuing a combined fellowship in Pediatric Pulmonology and Adult Pulmonology/Critical Care Medicine. Dr. Spahr's fellowship will be five years in length.

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Center Focus

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Newsletter of the
Cystic Fibrosis Center
University of Wisconsin
Clinical Sciences Center

Dear Patients and Parents:

Our last Cystic Fibrosis Center newsletter was produced in the winter of 2000. We have been very remiss at not producing CF Center newsletters since then and for that, we apologize. We do have a number of exciting developments occurring in the CF Center and want to inform you of these developments. We hope to be able to send out brief newsletters to you on a regular basis in the future.

PEDIATRIC CLINIC REMODELING

For patients in the pediatric program, many of you may have noticed that one-half of the pediatric clinic module is hidden behind temporary walls. This first half of the clinic is undergoing remodeling and is expected to be available for clinic visits in March 2003. We will begin using clinic rooms in the remodeled half in March and the current half of the clinic that is being used for visits will be temporarily closed for remodeling. The end result will be a beautiful new remodeled clinic space. In the meantime, we are carrying out our regular clinics with the least amount of inconvenience and certainly aim to continue to provide the highest quality care.

CHANGES IN LABORATORIES

The Cystic Fibrosis Foundation has convened a number of Consensus Conferences to aid CF Center personnel in the monitoring and treatment of people with cystic fibrosis. These conferences have been in the areas of Infection Control, Pediatric Nutrition, Screening for Allergic Bronchopulmonary Aspergillosis, and Bone Health in CF. Previously in the pediatric program, our laboratory monitoring has consisted of respiratory secretion cultures twice a year and annual blood work that includes a vitamin E level, liver enzyme ALT, and a blood sugar for patients above ten years of age. Based on recommendations from the Cystic Fibrosis Foundations Consensus Conferences, we have changed our laboratories to the following:

1. A respiratory secretion culture is recommended at every single clinic visit.
2. Blood work obtained on an annual basis is recommended as the following:
   a. Vitamin levels consisting of vitamins A, E, and D.
   b. A complete blood count with differential cell count.
   c. Liver enzymes ALT, AST, GGT, and Alkaline phosphatase. Also measurement of bilirubin.
   d. For patients above ten years of age, a blood sugar measurement.
   e. For patients above six years of age, a serum IgE level. IgE is an allergy antibody that can be elevated in the condition Allergic Bronchopulmonary Aspergillosis.

STUDIES

The University of Wisconsin Cystic Fibrosis Center is participating in a number of studies currently and we will be adding new studies throughout 2003. Your participation is our most valuable resource as this will help us understand CF better and will lead to improved treatments and therefore improved health in people with cystic fibrosis.

1. Gene Modifier Studies

We are participating in three different gene modifier studies. As you all know, cystic fibrosis is caused by a mutation in the cystic fibrosis gene on chromosome number 7. There are over 1000 different mutations in the CF gene with the most common being the ΔF508. The clinical symptoms of CF vary from individual to individual. Some of this is due to different CF gene mutations. However, people can have identical CF gene mutations and have very different disease. This leads researchers to conclude that there must be other genes that modify the cystic fibrosis gene. The National Institutes of Health have funded a number of gene modifier studies and we are participating in three of these studies. They are as follows:

a. Determinants of early pancreatic injury in cystic fibrosis.

This is a study being conducted in the states of Wisconsin and Colorado. Both of these states perform newborn screening for cystic fibrosis by measuring a substance in a baby's blood called trypsinogen. Trypsinogen levels are elevated in babies with cystic fibrosis and thus, this is useful as a newborn screening test for cystic fibrosis. Later in life, these trypsinogen levels are actually lower than the rest of the population. This may reflect injury to the pancreas in cystic fibrosis. We are studying gene modifiers as they relate to trypsinogen levels over time in patients with CF. This study is available for patients who were born after 1985. Extra blood is obtained at the same time as a routine blood draw for the usual laboratory work. We also obtain blood on the parents in order to assess how genes are transmitted from parents to children.

b. Genetic modifiers of cystic fibrosis.

This is a study sponsored by Johns Hopkins University in Baltimore, Maryland. This is looking at gene modifiers in siblings with cystic fibrosis. In order to be eligible for this study, there must be siblings with CF in the family. Blood is obtained from the person with CF at the same time as a routine blood draw for blood work. There is also a blood draw on the parents to assess the transmission of genes from parents to children.

c. Genetic modifiers in cystic fibrosis lung disease.

This is a study that is sponsored by Case Western Reserve University in Cleveland, Ohio and the University of North Carolina in Chapel Hill. This study is examining gene modifiers in cystic fibrosis patients who have two copies of the ΔF508 mutation. In addition to having two copies of the ΔF508 mutation, another criteria for being in the study is certain levels of pulmonary function measurements. This involves obtaining blood at the same time as routine blood work from the patient only.

2. Diagnosis of Pseudomonas aeruginosa infection by ELISA

Pseudomonas aeruginosa is a common bacteria that is present in respiratory secretions in cystic fibrosis patients. This bacteria can be detected in coughed-up sputum from CF patients, however many CF patients do not have a productive cough. This bacteria may or may not be detected by performing a throat swab. Antibodies to this bacteria can be detected in the blood, however it is less than ideal to obtain blood frequently from pediatric patients (and many adults do not like having blood drawn either!) This study is looking at developing a test for Pseudomonas antibodies in the saliva from patients with cystic fibrosis. This study will involve patients in the pediatric program and will obtain saliva and blood from the child.

3. A study of the oral drug BIIL284 in adult and pediatric cystic fibrosis patients.

This study is looking at a drug called BIIL284, a medication that can decrease inflammation in the lungs. This drug prevents a chemical called leukotriene B4 from attaching to a receptor on cells. This will be a double-blind placebo controlled study in which half of the patients receive the active drug and half of the patients receive placebo. Neither the medical caretakers nor the patient knows who is receiving active drug versus placebo.

4. Early Pseudomonas infection in cystic fibrosis (EPIC).

There is interest in using medications (oral or inhaled or both) to treat new onset Pseudomonas aeruginosa in young patients with CF. This study will involve patients with CF who are between six months and twelve years of age.

Some of the above studies involve payment for your participation. This is to reimburse you for your time and inconvenience. We do not coerce anyone to participate in studies. Ultimately, the benefit for cystic fibrosis patients to participate in studies is to allow new understanding of the disease and new treatments to be developed. When you are contacted to participate in a study, we hope that you will say "yes". We will thoroughly explain the study to you and go over the consent form with you.

CYSTIC FIBROSIS FOUNDATION PATIENT REGISTRY AND ESCF STUDIES

The Cystic Fibrosis Foundation has a patient registry which tracks health information from cystic fibrosis patients. A similar registry is sponsored by Genentech and their study is called the Epidemiologic Study of Cystic Fibrosis (ESCF). Because of new regulations that protect the private health information of individuals, we will be asking for signed informed consent for these studies in the near future. These studies involve obtaining information from your routine clinic visits and sending them to the respective patient registries. No other action is needed on your part. The confidentiality of your health information is strictly protected.

UPCOMING INFORMATION

The Infection Control Consensus Conference has made recommendations to assist us in diminishing the chance of cystic fibrosis patients sharing infectious organisms. A policy is being created and we will share that with you in a future CF Center newsletter. We are also working on a transition document that will assist in the transition of pediatric CF patients to the adult CF team

BEWARE GENERIC PANCREATIC ENZYMES

We would like to reiterate the enzyme warning that was published in the fall of 1998 issue of Center Focus. We have recently seen more pharmacy substitutions of generic enzymes for brand name enzymes that has been driven by the insurance companies. Generic pancreatic enzymes have significantly different formulations and may be over-filled or under-filled with the proper amount of enzymes in each capsule. There have been treatment failures in CF patients who have been unknowingly switched to generic pancreatic enzymes. The Food and Drug Administration (FDA) has published a final rule on enzymes as of April 25, 1995. The generic enzymes that are now on the market were introduced after April 25, 1995 and, in effect, they are illegal in that they have not been adequately studied and submitted to the FDA approval process. For CF patients, you should do the following when you receive enzymes from the pharmacy. Examine the bottles and pills upon receipt and verify that they have a recognizable brand name such as Creon, Pancrease, Ultrase, or Pancrecarb. If you have received a generic and you were prohibited from receiving the brand name by your pharmacist or insurance company, please contact your physician at the CF Center. Also, if anyone has received generic enzymes and has had poor response such as an increased number of bowel movements or weight loss, please contact your CF Center physician. We are collecting these treatment failures to submit them to the Cystic Fibrosis Foundation and ultimately to the Food and Drug Administration.