Save The Date

CF Education Day

Saturday, November 8, 2008
7:30 am - 12:30 pm
Fluno Center for Executive Education
601 University Ave
Madison, Wisconsin

Official announcement and registration information will be mailed in late August.

Due to infection control reasons mandated by the Cystic Fibrosis Foundation we ask that patients with Cystic Fibrosis not attend this function. We welcome parents, family and friends of our patients with Cystic Fibrosis.

American Family Children's Hospital
Cystic Fibrosis Center
600 Highland Ave. K4/938
Madison, WI 53792-9988

Infection Control

For pediatric patients coming to our new clinic facilities in the American Family Children's Hospital, a reminder that our infection control policy is still in place. People with CF should wear a facemask as soon as they arrive at the clinic. If you do not have a facemask, these are available at clinic registration- please ask for a facemask. Please leave the facemask on until you are placed in a clinic exam room. At the conclusion of the visit, please replace the facemask and continue to wear it until you have stepped out of the building. These infection control guidelines are in place for your protection.

See insert for the infection control guidelines.

University of Wisconsin
Clinical Sciences Center

Center Focus

Summer, 2008

Newsletter of the
Cystic Fibrosis Center

Changes Abound at the Cystic Fibrosis Clinic

Over the past few years, you may have noticed changes occurring in your Cystic Fibrosis Center. Perhaps you've been coming to clinic more often and spending more time with the nutritionist, developing nutrition goals and making changes to your diet at each visit. The reasons for these changes are many.

Every time you come to a clinic visit, you get weighed, measured, and examined. All of this information collected during your visit gets entered into a database maintained by the Cystic Fibrosis Foundation (CFF). Each year, the CFF summarizes the data and gives a report to each CF Center. The report lets the Center know how well it performs, compared to other CF Centers, in areas such as lung function and nutrition outcomes.

One thing we discovered after receiving our reports is that we're just above the national average for nutrition outcomes. Being average isn't good enough for us. We want to be among the best because that means you, as patients and families, are getting top care.

Almost 2½ years ago, several members of our CF Center Team visited one of the Centers with the best nutrition outcomes as one of the first steps to improve care. We learned that the patients there come to clinic more often when they are either not gaining or losing weight. We also learned that the patients receive more supplemental and tube feedings and used appetite stimulants to boost calorie intake. Finally, the most important thing we learned is that everyone on the team has a strong desire to improve nutrition because it improves lung function.

Recently, the CFF stated this as well. The CFF established a team of leaders in the CF Community and asked them to study all of the articles they could find on CF and nutrition. After studying those articles, this team discovered that when people with CF are at a good weight for height, they have better lung function and are healthier. To compare weight to height, a tool called the Body Mass Index (BMI) is used. In people less than 20 years, the goal is to have the BMI (calculated starting at age 2 years) at the 50th percentile or above when plotted on a graph. For adults (age 20 years or older), this equals a BMI of 22 for females and a BMI of 23 for males. For children less than 2 years of age, weight to length is plotted on a different graph with the goal being a weight to length at the 50th percentile or above. Information on the calculation and graphs is at the end of the article.

We hope this explains why you're seeing a greater emphasis on nutrition status when you come to clinic. We also hope you understand that we want to work with you to develop a care plan that fits your needs, lifestyle, and condition. Remember, you and your family are a very important part of the team. If we all work together, we'll reach our goals and maximize care.

BMI = weight in pounds/height in inches squared
x 703

For children age 2 to 20 years, the graphs to plot BMI are located at:

http://www.cdc.gov/nchs/about/major/nhanes/growthcharts/clinical_charts.htm

University of Wisconsin Cystic Fibrosis Center Specific Outcomes

We and the Cystic Fibrosis Foundation agree with the concept of data transparency. This means that the averaged outcomes of patients seen in our Center should be available to patients and families. There is publicly available data on all Cystic Fibrosis Centers available at the Cystic Fibrosis Foundation website (*www.cff.org*). Data on that website is adjusted for attained age of patients, gender, pancreatic sufficiency, race/ethnicity, socio-economic status, and age of diagnosis. What follows in this article is the raw data reported to us for the calendar year 2006.

Pediatric Center

A very useful measure of lung function is the FEV_1 percent predicted. For patients 6-12 years of age, the University of Wisconsin CF Center median FEV_1 percent predicted is 95.1%, which is identical to the national average of 95.1%. For patients 13-17 years of age, the median FEV1 percent predicted is 88% at UW compared to a national rate of 87.4%. For patients 6-17 years of age, the UW median FEV_1 percent predicted is 92.3% compared to a national average of 92.1%.

Our nutritional outcomes are expressed in terms of BMI (Body Mass Index) percentile. For pediatric patients 2-20 years of age, we strive for a BMI percentile of at least 50. The median BMI percentile for CF patients 2-20 years of age at UW was 51.6 compared to a national average of 47.3.

Adult Center

For patients 18-30 years of age, the median FEV_1 percent predicted at UW is 58.7% compared to a national average of 68.1%. For CF patients > 30 years of age, the median FEV_1 percent predicted at UW is 50.7% compared to a national average of 54%.

In adults over 20 years of age, one no longer uses BMI expressed as a percentile. In adult patients, BMI is expressed as the actual value with the units of kilograms per meter squared. The Cystic Fibrosis Foundation goal is that males should have a BMI of greater than or equal to 23 kilograms per meter squared and females should have a goal BMI greater than or equal to 22 kilograms per meter squared. In the UW Center, the percentage of adult patients who did not meet those goals was 58.9% compared to national average of 60.2%.

We strongly support the Cystic Fibrosis Foundation Quality improvement Initiative and have no doubt that patient outcomes can become better by a careful collaboration between patients and their healthcare team. If you view the 3rd Plenary Session at the 2007 North American Cystic Fibrosis Conference by Dr. Michael Boyle, you will see that one of the principles of quality improvement is for patients to be seen more frequently in clinic if they are not meeting their pulmonary or nutritional goals. We support this concept to the utmost degree.

Participation in CF Clinical Trials

How are new drugs developed? There are many steps in this process, with one of the final steps being participation of patients in clinical trials. Today, all cystic fibrosis patients are benefiting from the previous participation of patients in trials which has led to the approval of medications such as TOBI, Pulmozyme and the anticipated approval later this year of Aztreonam for inhalation. There are many other new medications that are in development for patients with CF. Here at the University of Wisconsin, we are dedicated to participating in clinical trials and helping to advance the science of CF such that there will be new and better therapies.

Enclosed with this newsletter is a brochure from the CF Foundation entitled "I Am the Key to Finding New Treatments for Cystic Fibrosis." You can find out about clinical trials by calling the clinical trials hotline at 1-877-8CF-JOIN or by visiting the website *www.cff.org/clinicaltrials*. Additionally, ask your care provider about clinical trials here at the University of Wisconsin. Although it can be somewhat of a time commitment to participate in a clinical trial, one can be very proud of your role in studying these new medications. It is only through the participation of people with CF that we can develop new treatments. We hope that almost every patient will chose to participate in a clinical trial.

Quality Improvement Initiative

It has been well known in cystic fibrosis that outcomes vary from one CF Center to another CF Center. A good example of this is illustrated in the below graph, Median BMI (Body Mass Index) percentile in CF patients 2-20 years of age. Each vertical bar represents a different CF Center. Please note that there is a large variability with the Center on the far left having a median BMI percentile of 67 and the Center on the far right having a median BMI percentile of 21.7. (The green bar on the far right is the median of all CF Centers at 47.1. We strive for pediatric CF patients to have a BMI percentile of > 50 as this correlates with good pulmonary functions.) The question arises; Why is there such a variability of outcomes from one Center to another? We all have access to the same therapies (no one has a magic bullet that they are using for their Center only). The answer must lie in how Centers apply these therapies to their patients. The Cystic Fibrosis Foundation has been studying this by visiting Centers with good pulmonary and nutrition outcomes. Although there is not one single answer that results in good outcomes, there are a number of principles that, when applied, result in better outcomes.

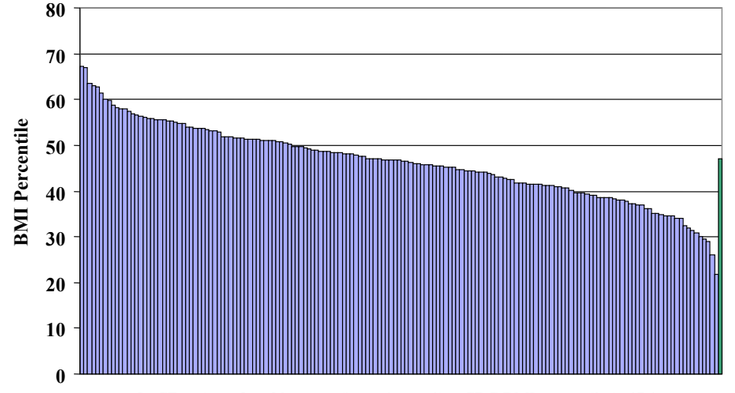
An excellent presentation of the principles of quality improvement was presented at the 2007 North American Cystic Fibrosis Conference. This lecture can be viewed on the internet if you have a broadband connection. Here are the steps to find that lecture:

1. Go to *www.cff.org*
2. Place your cursor over "research overview", and a drop down menu appears.
3. Go to 2007 North American CF Conference and click.
4. Scroll down to "Plenary Session III-SAT.,OCT.6 and click on "click here to view this session".

For those of you who do not have a broadband connection, we can supply a CD of this lecture. If your computer has the latest version of Windows Media Player, then you will be able to view this lecture. Please call our office at 608-263-8555 and request that we mail a CD to you.

In my 20 years of attending the North American Cystic Fibrosis Conference, I found this was the most inspiring lecture that I have ever witnessed. These quality improvement principles can make a difference. It is a partnership between patients and families and the medical care givers that, working together, can make a positive difference.

Median BMI Percentile for CF Patients 2 to 20 Years* by CF Center, 2006



In CF patients 2 to 20 years of age, the median CDC BMI percentile is 47.1.
The center level range is a percentile of 21.7 to 67.1.

*BMI percentile is not calculated for patients less than 2 years of age.