Center Focus

Spring, 2005

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

## Developmental Educational Guidelines

*Darci Pfeil, R.N., C.P.N.P*

As you may have noticed in clinic, the CF team has started using a developmental educational checklist to help guide us as CF educators. The checklist was developed by our team to help organize what educational materials should be addressed and at what age.

The developmental educational checklist begins at diagnosis and continues until transition to adult care. In each age group is a list of certain skills for patients and families to focus on and meet. One skill builds on the next, and independence slowly emerges from one stage to the next.

We are currently creating a patient and family version of the developmental educational guidelines to serve as a checklist for you. We look forward to sharing this with you in the near future. We will continue to work towards increasing education and independence through the current use of our developmental educational guidelines. If you have not been introduced to the educational guidelines, please ask to see them at your next clinic visit.

## REPRODUCTIVE HEALTH IN CF

Attention teens and parents of teens...

As we continue to develop our CF education program, reproductive health is a topic we would like address with our teens.

Around the age of 14, we would like to start talking about reproductive health and how it relates to CF.

Some of the topics we would like to talk about are:

- the reproductive system
- fertility
- prevention

Your teen will help guide our discussions over the next several years, as some topics may not be appropriate for all. We hope to individualize this piece of our CF education program to each patient and family so that we are supportive and respectful of each family's wishes.

Most importantly, we would like to get your permission to discuss these issues and talk with you first regarding any questions or concerns you may have. Some of these issues are things that may be discussed with your teen's primary care provider. However, there are some things that relate specifically to CF and that is why we are adding this to our CF education program. If you do not want us to discuss these issues with your teen, you have that choice.

We understand that every family has their own ideas of how and when to discuss these issues. We would like to know how you would like us to approach your teen regarding these topics, and how we can best obtain your permission and answer any questions or concerns you may have.

We want this to be a success and want to make sure your wishes are met. Please let us know what would make you and your teen feel most comfortable. You can reach us via phone or fax. We look forward to hearing from you!

Phone: 608.263.8555

## NEW CF Nutrition Initiative

*Mary Marcus, MS, RD and Erin Tarter, MS, RD*

The UW Pediatric CF center is pleased to announce a new CF Nutrition Initiative. Our goal is to optimize the nutritional health of each and every child with CF followed by the UW Pediatric CF center. The first part of this plan was completed in early January through a generous grant from Digestive Care, Inc., which allowed us to provide you with a complementary copy of the newest edition of "A Way of Life: Good Nutrition Cystic Fibrosis Nutrition Handbook and Cookbook". In the coming months, we will start to use a new CF Nutrition Action Plan with all kids with CF. The CF Nutrition Action Plan will give you a snapshot of how your child is doing nutritionally and provide you with written nutrition goals.

Please feel free to contact Mary Marcus, MS, RD at 608-263-9182 or Erin Tarter, MS, RD at 608-261-1746 if you have any questions about this exciting new CF nutrition initiative!

## Electronic mail

Electronic mail has become a daily part of our lives for many of us. We can communicate with you by e-mail. However, the University of Wisconsin Hospital has a written policy that requires patients to sign an information & consent prior to e-mail communication. If you want to communicate by e-mail with your healthcare provider, please call us (pediatric office: 608-263-8555; adult patients, call Sue Osmond at 608-262-5307) to request a consent form. Please realize that e-mail is not the preferred form of communication. For patients/families who want to communicate by e-mail, this should be for only non-urgent, non-time sensitive issues.

## SCREENING FOR CF RELATED DIABETES IN CLINIC:

**The Oral Glucose Tolerance Test**

We have begun screening for CF related diabetes in clinic per recommendations from the National CF Foundation. CF related diabetes occurs in approximately 26% of patients age 10-19 years. The CF care guidelines state screening for CF related diabetes should occur every other year for patients 10-16 years, and annually for patients >16 years. The screening is performed using the oral glucose tolerance test. In this test, a sample of blood is drawn with the patient having had nothing to eat or drink since the night before. After the initial blood sample is obtained, the patient drinks a 10 oz orange flavored drink called Glucola over 5-10 minutes. (The amount consumed is dependent upon the patient's body weight.) Another blood sample is obtained two hours later. This test may be performed as part of your routine clinic visit, or if you prefer, the test may be performed locally at your convenience.

**Summary:**

**Who:** All patients 10 years of age and older

**What:** Screening for CF related Diabetes

**When:** Every other year for patients 10-16 years
Every year for patients older than 16 years

**Where:** CF center clinic or locally

**How:** Oral glucose tolerance test

## Infection Control

We are continuing to work with our Infection Control team to create an Infection Control policy that will provide the maximal protection for our patients and will be compliant with the Cystic Fibrosis Foundation Infection Control recommendations. In the Summer, 2004 issue of Center Focus, we announced that antibacterial wipes would be available adjacent to the touch screen computer game and desktop computer in the Pediatric Waiting Room. Because of extenuating circumstances, those antibacterial wipes were removed from the Pediatric Waiting Room. We **highly discourage** patients from using the touch screen computer game and desktop computer in the Pediatric Waiting Room. Because this equipment is not being cleaned between patients, there is a very real risk that germs could be transmitted from one patient to another.

**Hospital admissions-Infection Control**

Although we know the bacteria that are in a CF patient's throat swab/sputum culture from the previous clinic visit, it is certainly possible that these bacteria may have changed since the previous clinic visit. The concern is that when patients are admitted to the hospital, new bacteria (such as MRSA: methicillin resistant Staph aureus or multi-drug resistant Pseudomonas aeruginosa) might have been acquired by the patient and there would be the risk of spread of bacteria to other patients. Therefore, we have a new policy for all pediatric and adult CF patients who are admitted to the hospital for a pulmonary exacerbation. The CF patient will need to be in contact isolation until the results of the admission culture are known. Contact isolation means that when the patient comes out of their hospital room, the CF patient should wear a mask, gown and gloves. Healthcare workers who go into the patient's room will also wear a mask, gown and gloves. This contact isolation can be discontinued after the results of the culture are known if the culture shows no MRSA nor multi-drug resistant Pseudomonas.

We are continuing to meet with Infection Control experts at the hospital and we will have additional information about infection control during your clinic visits and in the next Cystic Fibrosis Center newsletter.

## Bracelet for Breath

Show your support for the 30,000 people with CF by wearing this bright blue rubber wristband and raising awareness for the need for a cure. Money generated from the distribution of these "Breathe" bracelets will support the vital research and care programs of the CF Foundation. The word "Breathe" and " www.cff.org" appear in block letters on the wristband.

You must order a minimum of 10 wristbands. Wear one yourself and get your friends and family to wear the others!

This item can only be shipped within the United States.

To order, contact the Cystic Fibrosis Foundation by calling 800-FIGHTCF or visit their website at www.cff.org

Amount: $2.00

Adult size (8") Youth size (7") available also

(Minimum order must be 10; may be combined with youth wristband)

## University of Wisconsin Cystic Fibrosis Center Specific Outcomes

Many of you may have seen an article in the December 6, 2004 issue of the New Yorker magazine entitled "The Bell Curve". This article discusses the sharing of Cystic Fibrosis Center Outcomes with families and patients. We embrace the new thought in this millennium that we should share our Center Specific Outcomes with you.

## Pediatric Center

A very useful measure of lung function is the FEV1 % predicted. In patients 6-12 years of age, the University of Wisconsin CF Center median FEV1 % predicted is 94.4% compared to a national rate of 90.4%. Out of approximately 115 CF Centers, the UW Center is 38th best in the country for patients ages 6-12. For patients 13-17 years of age, the median FEV1 % predicted is 94.5% at UW compared to a national rate of 84.1%. The UW Center is 8th best in the country for median FEV1 in patients 13-17 years of age. The Cystic Fibrosis Foundation has two definitions of nutritional outcomes: patients at risk of nutritional failure, and patients that are in nutritional failure. (In our opinion, the terminology of using the word "failure" is too harsh. Perhaps one could use the term "nutritional issues" instead of nutritional failure.) In patients less than 18 years of age, the UW CF Center has 8.8% of patients at risk of nutritional failure compared to the national rate of 9.3%. For children less than 18 years of age in nutritional failure, the UW CF Center rate is identical to the national rate at 32.8%.

As discussed elsewhere in this CF Center newsletter, we have a new CF nutrition initiative. Our goal is for all patients to be as healthy as possible. This is a collaborative effort between you and your health care team. We have much room for improvement in nutritional outcomes. We are very proud of our pulmonary function outcomes, yet there is room for improvement there also. We want to be the best CF Center in the country. Working together, we can accomplish that goal.

## Adult Center

We now care for 110 adults with CF ages 18-68. The average age is 34 years old. Thirty patients are age 40 or older and 3 are in their sixties. Lung transplantation has now prolonged the life of over 50 patients at our center. The longest surviving CF lung recipient is now 11 years out of transplantation. Our survival rate for CF lung recipients is above the national average. So we are very proud of our transplant program.

As expected, as a group, our adults have worse lung function than our pediatric group.

Our proportion of adults in "Nutritional Failure" (see above) is high (53.8%) compared to the national rate of 42.3%, which is also high. In carefully assessing our patients, we do not find such high rate of nutritional failure based on our own criteria. However, we consider this a target for improvement. Those identified as in "failure" will be closely monitored and supplemented as needed. Patients and family will meet with the dietitian to set up nutritional goals and correct deficiencies more often than the required once a year.

The National data base indicates that were are well above average in the use of maintenance therapy with Azithromycin (UW 87% vs national rate 39%). Although we would appear to be below average for the use of inhaled TOBI, many of our patients are on inhaled Colistin, so in fact a larger proportion of our adults than it would appear are on inhaled antibiotics as suppressive therapy. The use of Pulmozyme is close to the national rate of 67%.

The guidelines recommend at least 4 clinic visits a year. Our adult rate, 19%, is significantly lower than the national rate of 49%. The reasons are variable. Some patients consider themselves "too well" to come every 3 months. Others require frequent hospitalizations; hence there is no need for clinic visits. We should work together to improve the frequency of clinic visits.

Some quality care initiatives recently implemented are: Bone Health and Vitamin D, Electronic CF specific medical record (to improve tracking and flow of patient management according to guidelines), and "Support group" monthly sessions now renamed (at the request of the patients) "CF connections" which is a network for adults with CF and their partners that offers personal support and educational assistance to help lead an active an productive life.

## The Clinical Practice Guidelines for Cystic Fibrosis

The Clinical Practice Guidelines for Cystic Fibrosis were created in 1989 by the Cystic Fibrosis Foundation (CFF) to provide a general framework for the management of patients with CF. The guidelines were distributed to all accredited CF care centers in the United States. The guidelines include diagnostic and treatment recommendations in addition to a comprehensive education program to promote optimum understanding of CF. The guidelines are meant to be flexible in order to be interpreted by CF caregivers to the benefit of individual patients. Below is a summary of the UW CF Center's Clinical Care Guidelines based on the CFF Clinical Care Guidelines for individuals over 6 years of age in stable health. Hopefully, these guidelines can assist you in making knowledgeable decisions that will optimize your health. The CF team can also work with you to tailor additional recommendations based on your health needs over time.

| **Recommended Care** | **Minimum Frequency** |
|---|---|
| Routine Clinic Visit for assessments, interventions, monitoring, education and counseling | Every 3 months or more often as indicated |
| Sputum culture & sensitivity | Every 3 months |
| Pulmonary Function Testing: | |
| Spirometry | Every 3 months |
| Complete pulmonary function (lung volumes etc) | Annual |
| Nutrition Evaluation: | |
| Height and Weight | At each clinic visit |
| Full assessment of growth, dietary habits and nutritional therapies | Annual |
| Nutrition & dietary education | Annual |
| 3 day Fecal Fat Test | As indicated |
| Chest Radiograph (x-ray) | Annual or more often as indicated |
| Psychosocial Evaluation (Social Work): | |
| Addresses concerns such as the impact of chronic disease on the family, insurance, school or other important family issues | Annual or more often as indicated |
| Respiratory Care Evaluation: | |
| Review of airway clearance techniques, inhaled or nebulized medication administration, and equipment use and cleaning | Annual or more often as indicated |

## The Clinical Practice Guidelines for Cystic Fibrosis (continued)

| Recommended Care | Minimum Frequency |
|---|---|
| Laboratory Tests: | |
| Complete Blood count (CBC) | Annual |
| Vitamin A & E | Annual |
| Vitamin D | Annual, preferably in fall |
| Antibody screening for ABPA (IGE) | Annual |
| Liver enzymes | Annual |
| Random Glucose | Annual ages 10+ |
| Albumin | As indicated |
| Electrolytes | As indicated |
| Bone Density (DEXA scan) | Baseline no later than 18, earlier based on risk assessment (steroids, inadequate nutrition) |
| Genetic Counseling | As indicated |
| Oral Glucose Tolerance Test (OGTT) for CF-related diabetes (CFRD) | Ages 10-16: Every 2 years<br>Ages 16+: Annual |
| Patient Education | Ongoing |

## "D"-Lightful Vitamin D and CF

*Written by Michelle Flatt, UW PPC nutrition fellow*

Vitamin D is important for building strong bones and teeth. Without it, bones can become thin and brittle. People with CF need 400-800 IU of Vitamin D everyday. It is important to work with your CF healthcare team to decide how much Vitamin D is right for you. There are not many foods that are naturally high in Vitamin D; however many foods are fortified with Vitamin D, including milk, some yogurts, juices and breakfast cereals. Be sure to take your CF multivitamin every day since it also contains vitamin D. People can also make their own Vitamin D from sunlight which is why Vitamin D is also known as the Sunshine or D-lightful vitamin. Just 10 minutes of sunshine per day on uncovered arms, hands, and face is all you need. However, living in the Midwest makes it hard to get enough Vitamin D during the winter months. Try this recipe rich in vitamin D from A Way of Life: Good Nutrition Cystic Fibrosis Nutrition Handbook and Cookbook.

**"Muscle Milk"**
1 cup whole milk
2 T non-fat milk powder

Add dry milk powder to whole milk and stir until milk powder dissolves. Best served very cold. This can also be used for cooking or served over cereal.

**Nutrition Information:**

| | |
|---|---|
| Yield: | 1 serving |
| Serving size: | 1 cup |
| Calories: | 180 |
| Carbohydrate: | 16 grams |
| Protein: | 11 grams |
| Fat: | 8 grams |
| Calcium: | 395 mg |
| Vitamin D: | 140 IU |

## Sputum induction

Attention CF patients 6 and older. The next time you come for your "C visit" you will see that we have added something to our routine. The "C visit" is the appointment when you meet with our respiratory therapists to review airway clearance techniques and make sure your equipment is in working order. At this visit, we will be having patients perform a sputum induction to help those who cannot cough out sputum on their own. The technique is called the hypertonic saline sputum induction and you may have heard or read about it as an airway clearance technique. Many patients use hypertonic saline on a daily basis in order to help them clear mucous from their lungs. We will be introducing this in our clinic visits in order to help you give an adequate sample of sputum. Additionally, it may be an opportunity to decide if hypertonic saline is a useful airway clearance technique for you.

This is what to expect at your next "C visit":

- Before the visit, you will have pulmonary function testing (PFTs) as usual.

- If you do not produce sputum during PFTs, and you have not produced sputum in the clinic during the last year, then we will have you try the hypertonic saline sputum induction. If you normally take an inhaler like albuterol to open you lungs prior to airway clearance techniques, please bring the inhaler so that you can use it with the hypertonic saline sputum induction.

- The sputum induction consists of inhaling a nebulizer treatment that contains saline (salt water) that breaks up mucous and allows you to cough it out. The sputum induction will make you cough and so the respiratory therapist will be there to coach you through the process and help you to get the phlegm out. The nebulizer treatment lasts about 20 minutes, but you may take a break or stop early if you feel you have cleared all your mucous or you are coughing too hard.

- If you feel that this sputum induction was helpful, let your doctors know and they can prescribe hypertonic saline for use at home as an airway clearance technique.

We are excited to offer this new technique in our clinic visits, because it has been shown to be a useful tool at many CF centers throughout the country. Hopefully, you will find it a useful tool to add to your lung health maintenance program.