CF Connection

Summer 2007

University of Florida Pediatric Pulmonary Center

When to Call-Part II: Digestive System Problems

In the last newsletter, you learned about lung changes that should cause a phone call to your CF Care Team. Now, let's talk about changes in the digestive system that should also prompt a phone call. Some of these will sound familiar because they are part of the routine questions asked in clinic.

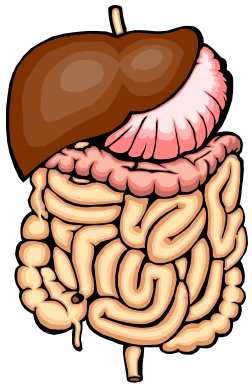
You already know that in most people with CF, enzymes that break down food can't get from the pancreas to the intestine. That is why people with CF take digestive enzymes (Creon, Pancrease, Ultrase) to replace what the body usually makes. These enzymes, taken immediately before eating, will help digest the food into bits that the body can use for energy. If enzyme doses are missed or the dose is too low, the food isn't broken down enough and the intestines are left to manage much bigger loads than they are made for. Then you would notice pain, gas, bigger stools, or more frequent, loose stools. These are things to call about.

On the other hand, if you/your child stop having regular bowel movements or all that's coming out is liquid, that's another cause for concern. This will likely be associated with a feeling of pressure or pain because of an over-full intestine. Call and we can help decide how best to manage. The earlier you call, the easier the solution.

Good nutrition is very important for growth and good lung health. Weight loss is a bad thing and we want to know about it. Our CF nutritionists are experts in finding ways to help regain or maintain good weight. This is another time when calling sooner is better than waiting.

In summary, weight loss, belly aches or pains, and changes in bowel movements should all prompt a call to your CF Care Team.

Cindy Capen, MS, RN



Inside this issue:

We Need You!

Please let us know if you have any tips or stories you would like to share. We would love to include them in our Family Corner! Send them to us at the address on page 6 or e-mail them to hillajb@peds.ufl.edu.

The Calcium Corner

Calcium is the most abundant mineral in the body and is needed to support the structure of bones and teeth, help with muscle contraction, and aid in the transmission of messages through the nervous system. Getting enough calcium can be especially important for people with cystic fibrosis (CF) because they have an increased risk for certain bone diseases such as osteoporosis (porous bones) and osteopenia (weak bones).

In addition, not getting enough calcium can interfere with important CF treatments, lead to potential bone fractures, and even prevent or delay lung transplantation in the future.

The best source of calcium is through the diet, but many times, people with CF need additional supplementation. When looking for a supplement, you will notice that there are many different varieties to choose from. Sources of calcium in supplements include calcium carbonate, calcium citrate, calcium glubionate, and tricalcium phosphate. Not all sources of calcium are the same; some can be better absorbed in the body than others. If you are on medication such as Prevacid, Pepcid, Nexium, or other antacids to reduce stomach acid, calcium citrate (such as Citracal) may be the best choice since it is absorbed well regardless of stomach acidity. Other sources of calcium require stomach acid for absorption and therefore must be taken with food. In addition, to increase the amount of calcium absorbed, take your calcium supplements at different times throughout the day instead of all at once. Make sure that you are not taking more than 500 mg of calcium at one time. Also, make sure to avoid taking your calcium supplements with calcium-rich foods such as milk, yogurt, ice cream, and cheese.

As with other supplements, calcium is available in many different forms including pills, chewable tabs, and flavored chews. Each form has advantages based on your preferences and individual needs. There are also brand name and generic supplements available. The generic supplements will be slightly cheaper, but they may not dissolve as well in the stomach as the brand name supplements. A good way to test how well a supplement dissolves is to put it in a glass of warm water. If it dissolves within 20 minutes, it will most likely dissolve in your stomach. Although supplements are not regulated by the Food and Drug Administration (FDA), there are voluntary methods of regulation. The USP (US Pharmacopeia) is a voluntary set of standards for supplements to check for quality and purity. When looking for a supplement, be sure to check for the USP stamp.

Current calcium intake recommendations are 500 mg/day for children 1-3 years, 800 mg/day for children 4-8 years, and 1,300 mg/day for children and teenagers 9-18 years. Since bones continue to gain more mass until about age 30, it is especially important for children and teenagers to meet these guidelines.

Before taking any new supplement, talk with your CF care team. There is the potential for calcium supplements to interact with other medications you may be taking and you want to be sure not to exceed the upper limit for calcium intake (2,500 mg/day). If you are concerned about your bone health, talk with your doctor and CF care team about screening options.

For more information on calcium, visit these websites:

NIH Office of Dietary Supplements:
http://ods.od.nih.gov/factsheets/calcium.asp#h1

Cystic Fibrosis Foundation:
www.cff.org

MyCysticFibrosis.com

Cristin Cuozzo and Jessica McIntire,
Master of Science-Dietetic Internship Students

A Patient's Perspective: The Differences Between Shands at UF and AGH

I was one of the first people to move over from Shands at UF when the transfer started to move pediatric patients to AGH. The transition went smoothly, and when I got to AGH, I was impressed to say the least. All the rooms are private, unlike Shands at UF, where you could get stuck in a room where it was difficult to get any sleep.

In addition, the food at AGH is considerably better than Shands at UF. All of the food at AGH is a more home style type of food. They have 2 chef's specials every day that you can choose from and also a pediatric or adult menu that can be ordered from for any meal. CF patients also get access to a "Grill Pass" and can order unlimited hamburgers and other grilled items from the cafeteria. Examples of meals are meatloaf and potatoes or turkey and dressing, something for those people who get tired of or don't really like chicken nuggets and pizza all the time.

The nursing care at AGH seems much better to me as well. Many of the nurses moved over from Shands at UF to AGH. Even though there are quite a few new nurses, they provided great care. There are still plenty of activities at AGH just as there were at Shands at UF. ChildLife comes by almost every day to see if you want anything like board games, videogames, or just a volunteer to talk to or play with. Arts In Medicine also comes by to see if you want anything to color, paint, or some craft activities to do. There is a computer room on each of the 3 floors that you can use if you have a volunteer or someone from ChildLife with you.

These are just a few of the differences between Shands at UF and AGH, but I have to say I like AGH much better.

James Woods

Great Strides: A Great Success!

The Great Strides walk to raise money for the Cystic Fibrosis Foundation was a great success! Over $36,000 was raised this year and about 235 people participated in the event. This was a great team effort! Buchholz High School and Natasha Gaziano raised the most money on the team and individual levels, respectively. Following the walk, the PPC held an education session for anyone interested, and plans to continue this in future years. Thank you to everyone who participated and made this a positive experience!

Information Contributed by: Carrie Godfrey

What You Need to Know About CFRLD!

Most of the care for people with CF focuses on maintaining lung function. However, CF affects many organs and frequently leads to significant gastrointestinal and nutritional problems. Cystic fibrosis-related liver disease (CFRLD) is a complication that has been gaining more attention.

About 1/3 of people with CF will develop CFRLD. Damage is caused by thickened bile in the ducts of the liver. It is not known why some people with CF have liver disease and others do not. In addition, among those with liver disease, it is not known why some have mild disease and others have severe disease (liver failure). CFRLD shows clinical symptoms in most affected people by age 10.

CFRLD is under-diagnosed because signs of liver disease often cannot be detected until the problem is severe. Detecting CFRLD early is important because research suggests only early disease is likely to be reversible. Unfortunately, there is no gold standard for screening for liver disease, making early detection difficult. At this time, diagnosis relies on a combination of the doctor's physical examination, blood tests, and imaging studies. A physical examination of the abdomen is part of every clinic visit. Annual blood tests are recommended by the Cystic Fibrosis Foundation. However, routine imaging (such as an annual liver ultrasound) is currently not recommended.

So what can be done for CFRLD? Only one medication, ursodiol (also known as Actigall), is currently available that has proven benefit. Studies suggest that ursodiol may be able to reverse early liver disease. In addition, ursodiol is free of serious side effects. It is important to point out that we do not yet know if starting all people with CF on ursodiol at a young age could prevent the development of liver disease. Correcting malnutrition from CFRLD is a second area of focus for treatment. People with CFRLD may need to exceed calorie requirements for CF patients without liver disease by 20 to 40%. Monitoring and correcting deficiencies in fat-soluble vitamins (A, D, E, and K) is needed more often in people with CFRLD.

Lots of questions about the prevention, diagnosis, and management of CFRLD are still unanswered. The biggest of these is how effective ursodiol is in preventing liver disease. Routine liver ultrasound to help make an earlier diagnosis should also be a future consideration. If you have any questions about CFRLD, please talk with the doctor at your next visit.

Thomas Horsman, MD

PPC Family Website

The UF PPC is developing a website for families. The website will have educational information and will also provide the opportunity for families to interact online (via a discussion board and blogs). At present, the site has some general parenting materials on it as well as some educational materials related to CF. We are planning to add more information over time. Please let us know what kinds of information you would like to have available. Also, if you have materials, information or useful links, please e-mail Susan Horky (chaunst@peds.ufl.edu). Please also feel free to start a blog or discussion.

To access the site, go to: http://elearning.mchtraining.net/. Then click the "Pediatric Pulmonary Centers" link on the left. This will bring you to a page with three PPC listings. Click on the third one ("PPC Family Community"). This brings you to a login page. You can simply click on "log in as a guest" and you will get to the site. If you have any questions, please feel free to email or call Susan Horky (352-392-4458).

Susan Horky, LCSW

Inhalers Go Green!!

Your inhalers are changing!

I am sure you've heard about greenhouse gases and damage to the stratospheric ozone layer. Well, you may not know that the stuff that has been used in your inhalers to "propel" the medicine out of its canister is one of the chemicals that damages the environment, known as chlorofluorocarbon or CFC. The pharmaceutical companies are mandated to change that chemical by 2008 and most have already done so. The new, environmentally-friendly propellant is hydrofluoroalkane (in case you want to toss around a big name). The easy version is "HFA" and that's what you will see on your inhaler.

What you will notice about HFA inhalers

- The spray will feel warmer and will be a bit slower.
- It will taste different…but not "bad" different.
- It will cost more! Because these are "new" drugs, they will all be marketed brand name for several years.
- They will clog more easily because HFA attracts moisture.

Changes in your inhaler care

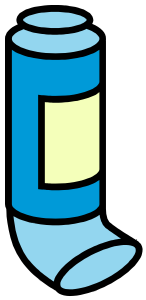
- MDIs are designed to deliver a perfectly measured dose of medicine each time they are puffed. Priming assures that a full dose will be puffed out when you inhale the medicine. Prime the MDI by squirting the inhaler into the air before taking your prescribed dose.
- Prime the MDI four times when you open a new one, or if you haven't used the inhaler for two weeks (for most meds including Proair®, Ventolin® and Proventil® or three days for Xopenex ®).
- Once a week, remove the canister from the mouthpiece and run warm water through the mouthpiece for at least 30 seconds. The opening gets clogged and has to be cleaned out so the "metered" dose of medicine can be released. DO NOT get the canister wet!
- Get in the habit of reading package inserts and checking expiration dates because these new products have more "rules" for use. For example, the brand name Ventolin® (albuterol) now comes with a very nifty dose counter so you can tell just how much is left, but it expires two months after you remove it from the foil packaging.

Which medications does this affect?

- All the albuterol products have now converted to HFA propellant. The brand names for these include Proventil®, Ventolin®, and Proair®.
- Most of the inhaled steroids that are MDIs have converted to HFA. These include Flovent®, Advair®, and Symbicort®.
- Some inhaled steroids, such as Advair® and Asthmanex®, come in a "dry powder inhaler" (DPI) and so are not propellant driven. These medications are not changing.

If you have any questions about HFA products, please give us a call.

Cindy Capen, MS, RN



Breathe Strong: Pilates for CF

Exercise is beneficial to everyone. It improves overall fitness and quality of life, builds strength and endurance, promotes bone health, and extends longevity. For individuals with CF, exercise is also believed to stimulate coughing, aid in airway clearance, and even maintain or slow the decline of pulmonary function. Although individuals with CF who are physically active enjoy a greater degree of general health with fewer respiratory complications, most individuals with CF exercise less than their peers who are not affected by a chronic illness. With all of this in mind, how can we encourage and integrate therapeutic exercise into the daily CF regimen?

For this dilemma, Pilates is just what the doctor ordered! Recognized as a movement based exercise that can be tailored to any level of fitness or health, Pilates is particularly effective in improving posture and lung capacity because it focuses on the abdominals, spine, and breath. Additionally, Pilates engages the mind, body, and spirit thus enhancing body awareness, body image, self-esteem, relaxation, and concentration.

As a 26-year-old living with CF, Natasha Gaziano was so encouraged by the benefits she experienced first hand that it inspired her to become a Pilates practitioner and develop a program designed to meet the needs of others living with this disease. Supported by the University of Florida Pediatric Pulmonary Center and Shands Arts in Medicine, *Breathe Strong* is a comprehensive and innovative program that includes private sessions for inpatients, group sessions for outpatients, a 45 minute exercise DVD, and a website with blogs, chats, and streamlined video. Natasha invites individuals with CF of all ages and health conditions to join this exciting program. Don't just breathe, breathe STRONG!

For more information, contact BreatheStrong@mac.com.

Natasha Gaziano, BFA and Lauren Arce, RN, BSN

Natasha working with David Brown.

Natasha working with Sarah Tanner.

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