

THE LINK

Sutter Cystic Fibrosis Center Newsletter



May 2006

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Clinic News:

The Sutter CF Team strives to make each of your clinic visits worthwhile and thorough. We welcome your questions and concerns and encourage you to write them down in preparation for your appointment. For maximum assistance with your airway clearance therapies, it is best to bring with you your vest and any other airway clearance equipment. If possible, it would be very helpful to bring a written list of the medications and respiratory treatments you have. Providing us with the name of the Lab where you obtain blood work and cultures is also very important. Finally, we would very much appreciate obtaining the name of the Radiological Facility, where you obtain chest x-rays. All this information helps us evaluate your health and treatment plan at your clinic visit optimally.

Behind the scenes, Susan O’Bra, our Clinical Data and Research Assistant, has been very busy on several projects. She is designing a new clinical database to track the data of our patients; i.e. spirometries, lab values, heights and weights, etc. Susan has also been working with John Hopkins providing data for the Sibling Study, for which we currently have nine sets of siblings enrolled. Lastly, she is collaborating with Dr. Chipps, our CF Center Medical Director, on an ongoing basis, analyzing patient data in relation to research studies.

From the Editor:

I will admit that I am not a “computer person”, however, I am a curious person who likes answers and has lots of questions (to the dismay of my husband and family, at times, but that’s another story...) Almost everyone who comes to our center has questions and wants information related to Cystic Fibrosis. With the wealth of information available on the Internet, people in general often don’t know what is true and what is the “best” information. On behalf of our CF team, I would like to promote the Cystic Fibrosis Foundation website, cff.org. As I was recently scanning over the home page, several topics caught my eye, which I think would be helpful to many of our patients and families. “Living with CF”, “Recent CF News,” and “Legislative Action” are some examples found on the Home Page. Click on CF Services Pharmacy and you’ll find such topics as “Patient Education,” “Insurance,” “Patient Assistance Programs” and of course “Products and Prices.” So, I encourage you to sign on and click away and the knowledge you gain could make you stronger, both mentally and physically. Give it a try!

Wendy Hubbs, RN, MSN, CNS

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The following article was written by one of our patients, Patrick Brown, and was featured in the "Teens Writing for Teens" column in the January 25, 2006 edition of The Paradise Post Newspaper.

Young at Heart

By Patrick Allen Brown

It is sad when children are forced to grow up way too fast. It's even sadder when they are not allowed a childhood at all. Today more and more children are losing their innocence at such a young age, which is wrong. Children should just stay innocent and play without the fear and worries of life. In today's society, children are subjected to violence, sex and drugs wherever they look.

You know it's bad when children as young as 5 years old are playing violent video games and see nothing wrong with games they are playing. I don't favor banning video games or even the claim video games lead to violent behavior. I believe parents who allow their kids to play these types of games are the ones to blame.

Looking back on my own childhood, I wonder if I allowed my innocence to fade too quickly. Did I grow up way too fast? I think I did. I always wanted to be an adult -- as do a lot of children. I used to look up to adults and admire how free they were.

Now that I am 18 and have to face some of the pressures adults must deal with, although to a lesser degree, I have realized we're not free but stuck in a never-ending battle with money to survive. Had I known then what I know now, I wouldn't have wanted to grow up at all. Instead of growing up and having to face this, I would have stayed in never-never land, where I could have remained a kid forever, where money wasn't a worry and playing was my only responsibility.

I think I have found my never land recently. I have begun a relationship with a girl who has shown me money isn't important for happiness -- as long as you love what you do. This is good, because if I become a writer, like I want to, I am going to have to learn to be happy while I am poor. I am the luckiest man in the world because even though my life has been full of lemons, I have made it full of happiness. My buddies and I are still young at heart, and we can play like we were 10 again. If everyone was



to just play like a kid again, I know for a fact this place would be a better place. Just play without heartache or pain; play without worry or care for money or life. If all of us forgot about the stress of life just for a day, or just for a minute, we would be happier and healthier.

I want my readers to do this for me: When you're stressed or worried about life, go to the park -- who cares if you have children or not? -- And play like a little kid; play tag, play hide-and-go-seek.

Just stay young at heart and let go of the worries of life, just for a moment. I am not saying to forget about your worries forever, but just long enough to clear your mind and wash away the stress of a hard day. If we all did this, then we'd be all happier for it. Who cares if you look like a fool?

Stress is the leading causes of heartache, so I say let go of the fear of looking foolish and play like a kid plays. We can have a longer, happier life if we all do as kids do.

I hope my readers will remember that life is full of responsibilities and if you are so focused on them, then you might miss all the greatness life has to offer. We need to play and enjoy life before we miss it. Play on a daily basis and let go of the stress, but remain responsible at the same time. I thank you for your time and attention ... and I hope you will take my advice and let go of your stress.

Just play ... that's all I can say. Just play.

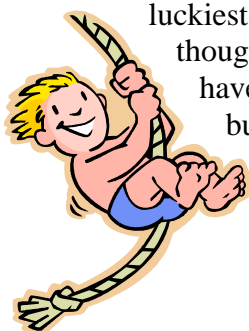
**CFRI: We Invite Your Participation in
A Special Poster Exhibition:**

Look At Me Now!

CFRI's 19th Annual Educational Conference
August 4-6, 2006

***Send us your posters* sharing your life
experiences, activities and adventures as children
and adults living with CF**

For details go to the CFRI website at www.cfri.org
or call the office at: (650) 404-9975



Hypertonic Saline Therapy for Cystic Fibrosis: Is it Right for You?

The Cystic Fibrosis Foundation funded a study in Australia to find out if inhaling a mist of hypertonic saline twice a day would help people with cystic fibrosis (CF). Hypertonic saline is extra-salty water that is sterile, so there are no germs in it. Because CF airways are known to lack enough salt and water, researchers thought a hypertonic saline mist would help clear the thick mucus from the lungs.

The results of this study were first published at the 2004 North American Cystic Fibrosis Conference and now have been published at the *New England Journal of Medicine* with another article on the same topic from the University of North Carolina at Chapel Hill. This fact sheet will help you learn about this Australian study. Please talk with your child's CF care center team to see if a hypertonic saline treatment should be added to you or your child's routine CF care.

How was the trial designed?

People in the study were put into one of two groups. One group inhaled normal saline that was a 0.9% salt solution. The other group inhaled hypertonic saline that was a 7% salt solution. Both groups inhaled a bronchodilator drug (to open airways) then inhaled a sterile salt-water mist using a nebulizer twice a day for a year. During the study, the patients and investigators did not know who was inhaling normal saline and who was inhaling hypertonic saline. Patients were watched closely for any health benefits or any problems while inhaling either solution.

What were the results of the trial?

Both groups had better lung function during the study. However, those taking hypertonic saline had even better lung function than the persons taking normal saline. Also, it was found that the people in the hypertonic saline group had fewer lung infections than the other group.

Were there any side effects?

The side effects that were noted by some people included coughing more, sore throat and chest tightness. (It is known that hypertonic saline can irritate the airways.) Your CF care center team may

want you to take your first dose of hypertonic saline while at the care center. This is to ensure your or your child's lungs will not have problems from hypertonic saline.

Is hypertonic saline right for me or my child?

Your CF care team can help answer this question. Knowing who was chosen for the clinical trial might help you decide if hypertonic saline is right for you or your child. The people with CF in the study were 6 years old and older and had mild-to-moderate lung disease. People with *Burkholderia cepacia* lung infection were not included in the study. Your CF care team can provide you with more information and may do some tests (sputum cultures) to see if hypertonic saline is right for you or your child.

Who should take hypertonic saline?

We do not know if hypertonic saline is safe for everyone. We do know that people who are 6 years of age and older, or who have an FEV₁ greater to or equal to 40% predicted might be able to take hypertonic saline. Before it can be prescribed, your CF care team will assess you or your child.

Can I make my own hypertonic saline to inhale?

To help prevent any germs from getting into your child's lungs, and to make sure the solution contains the right amount of salt, it is strongly recommended that you only use hypertonic saline prepared by a pharmacy. Ask your CF care center team which pharmacy in your area can fill a prescription for inhaled hypertonic saline.

The CF Services Pharmacy, a national mail order pharmacy, also can supply hypertonic saline with a doctor's prescription. Also, do not forget to clean and disinfect your nebulizer. If you have questions about nebulizer care, ask your CF care team or read "Respiratory: Stopping the Spread of Germs," available on the CF Foundation's Web site at www.cff.org.

How much hypertonic saline will be taken?

In the study, 4 ml of hypertonic saline was inhaled twice a day. A Pari PARI LC Plus® jet nebulizer

and a PariPARI Proneb® Turbo compressor were used to inhale hypertonic saline during the study. Your CF doctor will prescribe how much and how often you should take hypertonic saline. Ask your CF care team whether you can use your nebulizer and compressor.

If hypertonic saline is added to my or my child's CF care, should other drugs be stopped?

Hypertonic saline is one more helpful "tool" in CF care. It may be used as a part of you or your child's regular CF treatment. It is not meant to replace other proven treatments. **Do not stop any therapy before you talk with your CF care team.** It is always a good idea to talk about all of your therapies with your CF care team to make sure that you or your child are always getting the proper treatment.

Can I save time and mix my other inhaled medications with hypertonic saline?

You should not mix any other medications with hypertonic saline. Unless your CF doctor or

therapist tells you to do it, do not put two medications into your nebulizer at the same time.

What is in the future for hypertonic saline and CF?

We need to improve our understanding of how hypertonic saline is to be used. For example, we need to know if a different nebulizer would work better. Also, we need to find out if people who are sicker or younger than 6 years of age will benefit by taking hypertonic saline.

What other drugs are being developed by the CF Foundation?

To find a cure for this disease, the CF Foundation is pursuing several different strategies. It has committed close to \$131 million in science that uses the latest technology to discover new CF-specific drugs. At the same time, the CF Foundation is working to identify "low-hanging fruit," such as hypertonic saline, which are therapies already on the market for other diseases that may have a benefit for people with CF.

New Research Explores Genes that May Affect Severity of CF Disease

By Allison Handler, R.N., B.S.N., C.C.R.C. and Michael Knowles, M.D.,
University of North Carolina at Chapel Hill

The terms "gene" and "genotype" are fast becoming part of everyday language for people with CF and their families. Genetics are an important part of the CF puzzle. In fact, genes other than the CF gene may prove important for individuals with this disease. Therefore, researchers have been studying "modifier genes" and the impact they may or may not have on the severity of CF.

Q. What are DNA and genes?

A. The genetic code is made up of DNA (deoxyribonucleic acid) which is contained in long double strands. The DNA strands are wrapped together to make up chromosomes, which are contained in the nucleus of each cell. Inside the DNA strands, there is a genetic code for each gene, which provides the directions for a cell to produce specific components (proteins) for that cell to function. There are two copies of every gene; one copy is inherited

from the mother and one copy is inherited from the father. Some genes are similar for every human being, like the ones that direct the development of five fingers on each hand. The genetic code in other genes determines characteristics that differ among individuals – for instance, who will have blue eyes or brown eyes. There are 30,000 different known genes in the human genome.

Q. What is the CF Gene?

A. In CF, the DNA code in the CF gene is altered or "mutated." This mutation alters the gene's production of its protein. (The protein created by the CF gene is called the Cystic Fibrosis Transmembrane Conductance Regulator, or CFTR. The alteration in the genetic code is similar to how a change in just a few letters of a

sentence can alter the meaning of that sentence. Consider this example:

- Jim went to the school today.
- Jim went to the pool today.

The meaning of these two sentences changes entirely with the alteration of only one word or four letters.

CF develops because there has been an alteration (mutation) to the CF genetic code. If a child inherits one altered gene from each parent, then both the child's CF genes are altered. This leads to CF.

Most people diagnosed with CF have had their blood collected for DNA testing and have been told the code (name) of their CF genotypes. Scientists have identified more than 1,000 different mutations in the CF gene, which cause CF. Some alterations in the CF genetic code may cause people to be sicker. Other alterations in the CF gene may cause patients to have a milder course of the disease. However, it is not yet known which genotypes lead to a more severe or less severe "case" of CF. Research continues in this important area.

Q. What are modifier genes?

A. Many times the severity and symptoms of patients with CF are very different, even when they have the same alteration of the CF gene. The differences in severity among patients with the same CF genotypes may be caused by a combination of environmental and genetic factors. Some of the genetic factors come from other (non-CF) genes called modifier genes, which may affect or change the course of the CF disease. Researchers believe that modifier genes may play a role in the severity and symptoms of CF. By themselves, these modifier genes cannot cause a patient to have CF. However, modifier genes may work in conjunction with the CF gene to determine the severity of CF. For example, two patients may have the exact same CF genotype, but one patient

may have a mild case of CF, whereas the other patient may have a more severe course. These patients may have different alterations in their other modifier genes. Additionally, there may be various environmental causes, such as second-hand smoke, which bring about the differences in the disease severity.

Q. How do modifier genes work?

A. Research to advance our understanding of modifier genes is still in the early phase. However, researchers do know that the roles of modifier genes are different than the role of the CF gene. In the lung, some modifier genes are probably involved in fighting off infection and controlling inflammation. Other modifier genes may predispose the patient to the development of liver disease (cirrhosis) or intestinal problems. Ongoing research at the University of North Carolina and other places is aimed at studying the DNA from CF subjects with known CF genotypes to see which other genes modify the symptoms and severity of the disease. This will lead to a better understanding of the CF disease and hopefully point the way to new therapies. Modifier genes are only being studied in the laboratory and are not yet available as a part of clinical care or clinical trials.

Q. How is the study of modifier genes different from gene therapy?

A. In gene therapy studies, researchers attempt to place the healthy CF gene into the human body to see if the normal copy of the gene can correct the CF defect. The goal for that research is to help cure the disease or relieve symptoms. In the study of modifier genes, a blood sample is collected and the DNA is studied from that sample in the laboratory. No treatments are administered.

Q. What are the future goals for research with modifier genes?

A. Modifier gene research is very new. The short term goal for this research is to identify some of the modifier (non-CF)

genes that may play important roles in determining the symptoms and severity of CF. The long-term goal is to catalogue all the non-CF genes that affect CF disease severity. This will lead to more knowledge about CF, help define the type of therapy for individual patients and help develop improved treatments.

Conclusion

The severity and course of CF may be determined by a variety of factors, including the type of CF mutation, environmental causes and modifier genes.

The study and science of modifier genes is relatively new and has not yet entered into the clinical testing phase. **Therefore, this research currently has no clinical impact for people with CF.**

However, researchers believe a better understanding of modifier genes will be useful in the development of new treatments for CF in the future. For information about modifier gene studies, please ask your CF physician or CF nurse coordinator or visit the CF Foundation's Web site at

www.cff.org/research/clinical_trials/ongoing_trials/.



Stanford Education Day

By Jana Newman (Parent)

On March 11, 2006 three families from the Sutter Memorial Center attended the Stanford CF education day. The theme of the day was nutrition and the importance of eating right and maintaining good weight. Over and over again presenters stressed the importance of having a well balanced diet.

Dr. Carolos Milla from Minneapolis focused on the correlation of weight/nutrition and lung function. Weight gain in children of 3 oz per month resulted in better pulmonary function, the weight gain needs to be consistent not gain 4 oz one month and then lose two the next and then gain two the next month...try to avoid a zigzag pattern on the growth charts. The doctor from Minneapolis also said that at his clinic they use the parent's height to predict what the child's height should be and then put that on the growth charts. They realized that by doing this it helped see how the child was doing and if they were way under the predicted then they would look at the treatments and make adjustments. He said this helped keep them on track a little bit better and resulted in earlier treatment intervention.

Dr. Jackie Fridge and Julie Matel RD, presented on the use/placement of gastrointestinal tubes (G-tubes). The use of a G-tube is great for getting weight on and for maintenance of weight. A G-tube can be very beneficial and it is something that is worth considering prior to a dramatic decrease in weight. The studies indicated that it was better to start nutrition intervention early and do what it takes to keep the person weight above the 50%.

Dr. John Mark focused on complementary and alternative therapies in CF. There is an array of options for a CF patient or family to consider, but treatment decisions should be discussed with the CF Team and doctors.

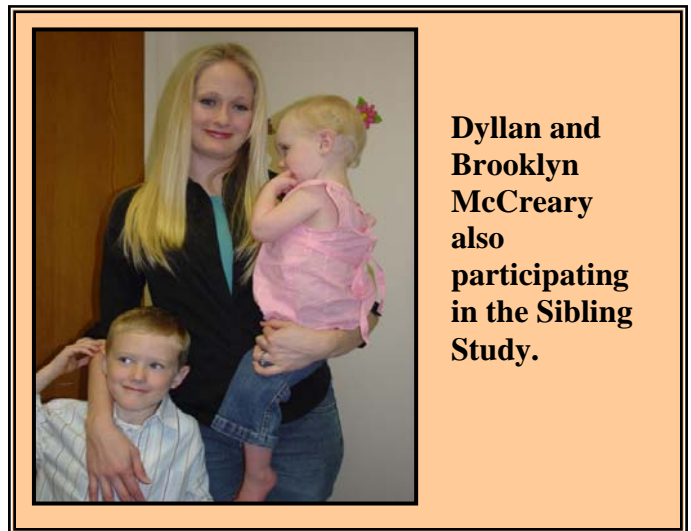
Dr. Terry Robinson provided an update on CT Imaging and the progress in making the equipment see the entire lungs not just segments and the time it takes to do an image. It was interesting that a number of people who had really high PFT's (FV1 in the high 90's) still showed more lung involvement than expected on the CT scans.

Ron Gibson, a doctor from Seattle came down and to speak about Pseudomonas and the ways to treat it. He is researching which medications are the most effective to treat the first acquisition of Pseudomonas.

General Information:

- Deductions for federal taxes: The government has published a document that helps with calculating deductions. The 2005 Medical and Dental Expenses document can be found at: www.irs.gov/pub/irs-pdf/p502.pdf. Now is the time to start saving receipts and documenting mileages for 2006.
- *HomeLine* Archive: Past issues of *HomeLine* are available at www.cfservicespharmacy.com. *HomeLine* is a quarterly newsletter published by the Cystic Fibrosis Pharmacy.
- **SOLVAY PHARMACEUTICALS, INC. TO AWARD 40 SCHOLARSHIPS TO STUDENTS WITH CYSTIC FIBROSIS** Solvay Pharmaceuticals, Inc. will award 40 students who have cystic fibrosis (CF) with college scholarships through its 2006 CREON[®] Family Scholarship Program. Applications are now available online at www.solvaypharmaceuticals-us.com and at CF centers across the country.
- The Bonnie Strangio Education Scholarship was established in 2005 to honor the memory of Bonnie Strangio. Please go to: www.cfscholarships.com/bonnie_strangio_scholarship.html for application information, deadline and criteria for the scholarship opportunity.

Our Center Photo Gallery:



Faith Davis celebrating Valentines Day.



Summer Tips: *Dehydration*

Understanding Dehydration

People with cystic fibrosis are more susceptible to dehydration than other adults. Often this is because there has been a reduction in kidney function or because the medications being taken (such as some antibiotics) require greater fluid intake. Dehydration, an abnormally low level of fluids and electrolytes (salts, such as sodium, potassium, and chloride) in your body, can result from diarrhea, severe vomiting, excessive sweating, and inadequate fluid intake. For people with CF, dehydration and excessive salt loss may also occur with exercising, especially in a hot or humid environment. Dehydration and salt loss can contribute to distal intestinal obstruction syndrome (DIOS) or heatstroke.

It is important to replace the lost fluids:

Recommended Fluids and Foods

Fluids	Fresh* Fruits and Vegetables	Other Foods
Broth	Cantaloupe	Gelatin
Coffee (iced or hot Decaffeinated preferred)	Grapefruit	Ice cream
Electrolyte-replacement fluids	Honeydew melon	Ice milk
Frozen pops made from juice or soft drinks	Orange	Sorbet
Fruit juices	Peach	Soup
Lemonade	Watermelon	
Punch	Cabbage	
Soft drinks	Celery	
Caffeine-free tea (iced or hot)	Cucumber	
Water	Lettuce	
	Spinach	
	Watercress	

*Not cooked, canned, or frozen

This article was prepared by

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