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CFRI NEWS

An Educational Community Contribution by CFRI

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CF Basic Research: The Work of One CFRI Postdoctoral Fellow

By Terry Machen, Ph.D., Department of Molecular and Cell Biology, University of California at Berkeley

Editor's Note: *For the past two years, CFRI has made a significant financial commitment to the CFRI Fellowship Program in which local labs, primarily specializing in basic CF research, receive financial support to hire research Fellows. Each of the next few issues will feature a glimpse into the lab of a local researcher and the work of his or her Fellows.*

Grischa Chandy, CFRI Postdoctoral Fellow, was supervised by Professors Terry Machen and Hsiao-Ping Moore, Department of Molecular and Cell Biology, University of California at Berkeley. He worked on two projects.

RESEARCH
Your CFRI Dollars at Work

The role of Cystic Fibrosis Transmembrane conductance Regulator (CFTR), in controlling organelle acidity in airway epithelial cells.

Epithelia form a barrier between the inside of the human body and its contact to the outer environment, for example, the mucosal lining of the gastro-intestinal tract, and more importantly for us here, the airways of the lungs. The main function of epithelial cells is to transport a variety of biomolecules, including salts, water and nutrients. To do this, genes in the epithelial cells express a large number of highly specialized transport proteins in their cell membranes. One of these proteins is the CFTR. CFTR is the protein that is missing or broken (depending on your CF mutations) in people with CF. In non-CF people, this protein creates a kind of molecular valve or transport machine that allows chloride and bicarbonate to cross the cell membrane.

One hypothesis to explain how a reduction of chloride permeability could lead to colonization of CF lungs by *Pseudomonas aeruginosa* and other opportunistic bacteria is that it alters the biochemical processing of intracellular organelles. This could occur indirectly in the following way.

Left: Terry Machen, Ph.D., with Grisha Chandy, CFRI Post-doctoral fellow, during his tenure in the Department of Molecular and Cell Biology at the University of California at Berkeley.

The main biochemical processing organelle in cells is the Golgi apparatus**,** a set of membrane-bound vesicles and tubules which are normally acidic relative to the rest of the inside of the cell (the cytosol) due to the accumulation of hydrochloric acid by a proton pump and a chloride channel. The organelle-acidity hypothesis proposed that CF cells had lower chloride permeability (due to lack of CFTR), reduced ability to accumulate acid, altered function of the key biochemical processing enzymes in the Golgi, and altered cell surface properties that cause high bacterial binding. The goal was to test this hypothesis in normal and CF-airway epithelial cells.

Chandy performed molecular engineering to generate a novel acidity-sensitive version of a green fluorescent protein (originally isolated from jellyfish) that could be genetically targeted to the inside of the Golgi vesicles and tubules. He then focused a digital imaging microscopy at this Golgi-localized genetic probe. He saw that the Golgi of CF-airway epithelial cells was more acidic than the cytosol, and a bit more acidic (opposite direction from hypothesis) than the Golgi of CFTR-corrected cells, though the small difference between the levels of acidity in the two cell types was unlikely to be important physiologically. Since CFTR seemed not to be involved in regulating Golgi acidity, the question then arose: What ion transport pathways are important? Further work showed that the Golgi has other chloride (and potassium) channels, and that the most crucial ion transporters are the H pump and an H leak that balance each other's activities to yield the characteristic Golgi acidity that is likely to be similar in all cells, including those in CF. Other explanations for the

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Letter from the Executive Director

The Couage to Make a Difference

Someone approaches you with the news...
You have five minutes remaining in your life!
What do you do?
Thoughts of yesteryears flash before your eyes.
Anticipation creeps slowly through your body.
You must accomplish something,
but what?
You continue to ponder…
but what?
If I leave now without making a difference…
what will happen?
So many people, so little time!
It's not fair…there just is not enough time.
And yet…
One moment in time can change the world!

Calvinia Williams

Craig Burleigh Photography

Calvinia Williams,

Do YOU have the courage to make a difference? Have you tested your determination? Does all appear to be lost at times? Do you have the will to succeed? Just the thought of time not going any further than five minutes for any individual makes all the difference in the world in how we set our priorities. Bring a smile to those that hurt. Give laughter to a world that is full of sadness. Spending time with our families encourages those living daily with CF. The joy of knowing you have given of yourself is priceless. CFRI's fundamental mission is to assist CF families through education and research. CFRI continues to expand because of our wonderful donors. Their actions set the tone for our success.

By the time this publication is read, we will be in the process of putting on our 14th Annual CFRI Conference. The theme this year is "Visions of Hope… Carrying the Torch." CFRI's upcoming conference has an impressive cast of knowledgeable CF scientists and doctors. After a recent conference, one CF family stated, *"Once a year we can look forward to our CF weekend. CFRI brings you in direct contact with the latest in treatment and CF research. The conference attendees are warm and we are able to build lifetime friendships. This is a must-attend conference*."

For those of you who have never attended a CFRI Conference please take this opportunity to do so, you won't regret it. This Conference offers an opportunity to network with CF families across the country. Your presence will make a difference, to your child, your family and the CF community!

Fondly,

Calvinia Williams
Executive Director, CFRI

P.S. Check out our brand new newsletter for teens with CF, their siblings and friends! It's called **Aspirations** *and appears on page 7.*

GOOD NEWS

For People with CF and their Families

Todd Trish Cycles Toward Health

By Todd Trish

I will never forget a medical appointment I had. A certain doctor (who shall remain nameless) stated, "because you have CF, don't expect to be competitive in any type of endurance event." I started out the appointment by telling him that I am a competitive cyclist and train with very fit individuals. He condescendingly replied that my training partners must be far from *fit* if I could keep up with them. Needless to say, this was my first and last appointment with him. He was misled on so many different levels!

Todd Trisch during his race last April.

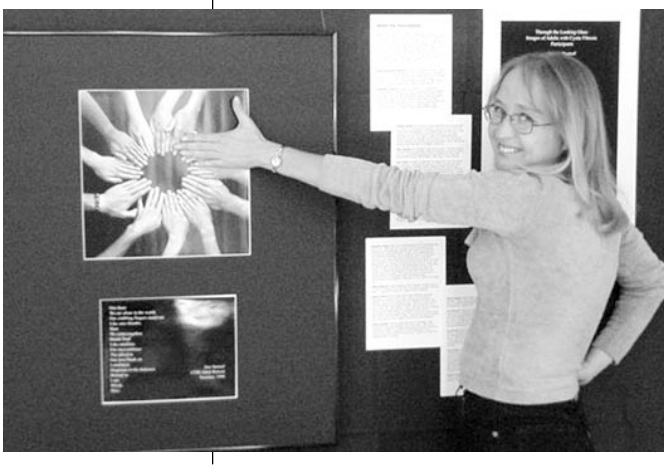
My goal is to go through life with the state of mind that I can accomplish anything I want to and to overlook as much as possible the effects of my disease. Cycling is a passion of mine and I ride for the enjoyment of it. Luckily, I benefit from the exercise with increased health.

Last April, I entered a mountain bike race and did quite well. It was the World Cup series and I competed in the amateur class. The course was an extremely difficult 14 miles of long rocky climbs and descents. It is safe to say that one must be fit to even complete the course let alone race on it. Ironically a year prior to my race I attended the event from the sidelines with an IV in my arm. Watching enviously, I made it a goal to return in a year and race. I did it, and did it well! There were 37 racers in my class and I came in 11th. No I did not win, but winning is relative to your goal, correct? It was a good day! My next goal is to get a top five finish in 2002 and grab a medal. I'll keep you posted…

However, the purpose of this letter is not to announce my accomplishment, but to encourage those with CF to exercise and hopefully relay some peace of mind that we can compete regardless of what some people may say to us. For the past fifteen years, my physician, Dr. Bradley Chipps, has always emphasized that I can do anything I want to do and that it's important not to give up. I wish all of you the same kind of positive relationship and support from your doctor that I have been given.

Todd Trisch is a 28 year old adult with CF and lives in Davis, CA

Kathy Schaal, an adult with CF from San Luis Obispo, California, "putting a hand in" to the beautiful Hands photo at the opening.



Breathing Room's "Through the Looking Glass" Opens

By Ana Stenzel and Kathleen Flynn

It was a great event on Sunday, April 29th, 2001 at the Lucie Stern Community Center in Palo Alto, California. The Breathing Room sponsored "Through the Looking Glass: Images of Adults with Cystic Fibrosis," its first annual and largest photographic exhibit to

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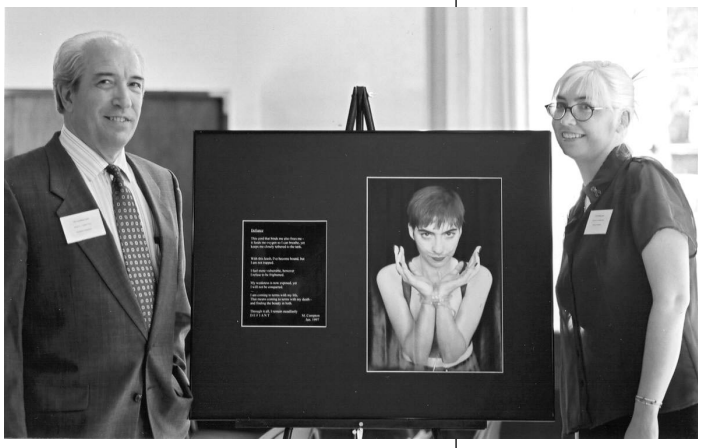
date. The Breathing Room is a community of support for CF adults. Its mission is to facilitate candid and open communication between adults with cystic fibrosis, to support the development of a community of adults with CF and to provide education and insight for families, caregivers, and medical professionals who impact our lives.



Michelle Compton, a post-lung-transplant adult with CF and director of "Through the Looking Glass," standing with a photo of herself from her pre-transplant days, along with her father, Scott.

The "Through the Looking Glass" exhibit is just one medium in which The Breathing Room celebrates CF adult expression. It came to fruition after months of planning by Michelle Compton, director of the Breathing Room, Stephen Boyer, production manager, and many other volunteers and Board members of the Breathing Room. The exhibit currently consists of 22 photographs of adults with cystic fibrosis.

Each CF adult is allowed to personally choreograph his or her own image. The adults use analogy, metaphor, or props from life with CF to depict how they see CF affecting their life, emotionally, spiritually or physically, providing an intense personal reflection of each unique experience. Poetry or prose written by the photographed person accompanies each photographic image.

One might initially respond to some of the photographs with mild discomfort. The images are very compelling and do not let the viewer look away. So often when one tries to verbally describe life with CF, the speaker can feel the listener withdrawing or changing the subject when the speaker treads onto painful ground. It can be very isolating for the person with CF. The photographs do not let you hide from the truth of another's experience. Other responses to the exhibition were very supportive. Some attendees had little knowledge of the emotional impact of cystic fibrosis and found the experience educational, profound and enlightening. As members of the CF community, we were immensely impressed with The Breathing Room's commitment to artistically demonstrate this often-neglected and sometimes dark side of the CF experience—the psychosocial and emotional impact of the disease.

Attendees included adults with CF, both pre- and post-transplant, parents of individuals with CF, friends and family members of the Breathing Room community, health care providers, and members of the local CF community. Attendees also included Kathy and Paul Russell, from Portland, Oregon. Kathy is the director of USACFA, a nonprofit community of CF adults which has agreed to be the fiscal sponsor of The Breathing Room until they acquire their own nonprofit status.

Ed Kinney, a long-time volunteer in the CF community, primarily at Stanford University Hospital, studying the images and poetry at the exhibit.

The highlight of the afternoon occurred when Michelle Compton—art director for Through the Looking Glass—presented a short speech about the history of the Breathing Room and gave thanks to all her sponsors, supporters and those who helped make this memorable event possible. Michelle expressed special gratitude to her parents, Diana and Scott Compton, who have tirelessly supported Michelle through illness, transplant and in her rebirth.

The Breathing Room hopes to further expand "Through the Looking Glass: Images of Adults with Cystic Fibrosis" and plans are underway for more photographic exhibits in the future. For more information and a look at some of these images, visit their website at: **www.thebreathingroom.org.** ■

Dealing with Intestinal Blockages

Editor's Note: *This article is not a substitute for careful monitoring by your physician. Please do not undertake any of these measures without checking with your doctor first!*

By Ann Robinson

Intestinal blockages or distal intestinal obstruction syndrome (DIOS) can be quite a nightmare for people with cystic fibrosis. DIOS may be brought on by too few enzymes (causing large and bulky stools), sudden changes in diet, dehydration, and in rare cases, too many enzymes. But frequently, one may never know what brought on the obstruction and this can leave the person who is recovering feeling anxious and vulnerable.

Our son was first diagnosed with an intestinal blockage as a newborn. At the young age of 10 hours he had a bowel resection because he had meconium ileus, an intestinal blockage (occurring at birth) of very thick, sticky meconium almost always at the site of the ileum in the small intestine. This occurs in about 10% of infants with CF. Later blockages in life, called DIOS, can be more frequent in people who have had meconium ileus, especially if a section of the bowel was removed. Since our son has had repeated incidences of blockages over the past 23 years, we have worked with his doctors at Packard Children's Hospital at Stanford to develop the following protocol both to prevent and deal with blockages.

Preventative Measures

1. Keep hydrated by drinking plenty of fluids especially during hot weather. Our son typically drinks 80-100 ounces per day of water or Gatorade diluted with water.

2. When playing sports or participating in other activities during hot weather, consider taking salt tablets. Our son took salt tablets while playing soccer, football and baseball upon recommendation from his physician.

3. Talk with your physicians about the appropriateness of taking a daily stool softener such as Colace, or a less expensive drugstore brand available over the counter.

4. Determine the efficacy of your enzymes. Most people can determine this by watching weight gain and monitoring stools. If stools are frequent and bulky that might be a sign that you should check in with your physician and CF Center dietician.

5. If enzyme usage is high, you might want to ask your or your child's physician to see if stomach acidity is destroying the digestive enzymes before they are able to work in the small intestine. Our son's physicians recommended an endoscopy and then determined that his stomach acidity was indeed too high. The drug Prilosec was then prescribed. Prilosec lowers stomach acidity while improving the effectiveness of enzymes and lowering the risk of intestinal blockages, not to mention other problems related to high stomach acidity.

6. We talked with our son's physician about an alternative digestive enzyme we had heard of called Pancrecarb® (made by Digestive Care, Inc.). The bicarbonate in the enzyme aids in keeping the stomach acid from destroying the effectiveness of the enzyme in the small intestine. This really improved our son's digestion and ability to absorb calories.

7. Try to eat a high fiber diet, avoiding the BRAT diet foods of Bananas, Rice, Apples and Toast. Our son does eat the BRAT diet items, but only in moderation and not while he is feeling sluggish. (Instead of eating rice, we cook riso or orzo, a small pasta shaped like rice.) Avoid or eat in moderation all foods that are binding.

(Continued on page 14)

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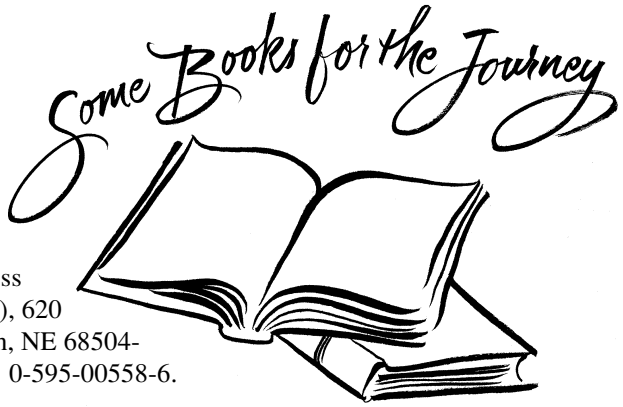
Fat and Loving It! Digestive Care, Inc. Supports Nutrition in People with CF.

The second printing of Gail Farmer's *Fat and Loving It!* has been completed. This book is much more than a cookbook. It is a nutritional guide to living with CF with all the information you need to know about fat, essential fatty acids, proteins, and carbohydrates. The book makes extensive use of handy charts on vitamins, minerals, fiber, caloric analysis and meal planning. Other interesting topics include pancreatic enzyme replacement, whole chapters dedicated to age-appropriate nutrition, snack ideas and menus, not to mention helpful one-page hints thoughout. This is a must have for every high-fat CF-conscious cook. Digestive Care, Inc. is giving away complimentary copies of this cookbook. To receive your copy you must submit a completed ORIGINAL order form, available at your CF Center. If your CF Center is out of forms, contact Rich Holland, Digestive Care sales representative at **rholland926@aol.com.** Digestive Care is also sponsoring the costs of the Packard CF Center Cookbook mentioned on page 14.



Blow the House Down: The Story of My Double Lung Transplant, by Charles Tolchin, © 2000, Writers Club Press (an imprint of iUniverse.com, Inc.), 620 North 48th Street, Ste. 201, Lincoln, NE 68504-3467, **www.iuniverse.com.** ISBN: 0-595-00558-6.

Review by Kathleen Flynn

I discovered this book while reading an article on the questionable nature of e-publishers and their potential for both inundating the public as well as diluting the quality of literature available to the public. (An e-publisher allows you to publish your book on-line for very low cost, but also with almost no editorial support.) Oddly enough, the author of the article ended by noting that if such a medium for books were not available, then he would not have experienced *Blow the House Down* by Charles Tolchin, the excellent account of Tolchin's battle with CF and his double lung transplant. I had to find a copy.

This book is another must-read. First of all, Tolchin's medical account of his transplant story is quite detailed, medically articulate, and very inspiring. Like most transplant recipients, he had plenty of complications to contend with, and the road to recovery was frequently bumpy. He also was determined to hold onto the parts of his life that were not about CF, his writing, his ice hockey, his family and friends. I recommend this book for teens and adults awaiting transplant, as well as their families and support people. Tolchin is very honest about what you are getting yourself in for and he gives the reader a pretty good idea of the inner strength and discipline required to survive and recuperate. Initially, I found the author's style a little awkward, especially some of his descriptions of people. I tired of some of his asides, especially those involving Tolchin's rather consistent pursuit of women. What made me tolerate this was the remarkable maturity and serious reflection with which he presents the rest of his story. Perhaps it is this maturity, determination, persistence, and ultimately gratitude in the face of his illness that is truly incongruous. An incredible story.

Managing Cystic Fibrosis Related Diabetes (CFRD): An Instruction Guide For Patients & Families, Dana S. Hardin, M.D., Carol Brunzell, R.D., C.D.E., Kathleen Schissel, R.D., Terri Schindler, R.D., M.S., Antoinette Moran, M.D., Published by the Cystic Fibrosis Foundation, Bethesda, MD 20814, © 1999, 72 pages. To obtain a copy of this free guide, contact your local Cystic Fibrosis Center or the Cystic Fibrosis Foundation at (800) FIGHT CF.

Review by Ann Robinson

This informative instruction guide presents detailed medical material in terms that are easy to understand. It is intended to supplement information provided by diabetes caregivers and physicians treating patients with cystic fibrosis. Each chapter focuses on different aspects of managing cystic fibrosis related diabetes (CFRD). A recent study found that up to 75 percent of CF adults have some form of glucose intolerance and 15 percent have CFRD.

People with CF have insulin deficiency probably due to thickened secretions that have caused pancreatic scarring. When a person has CF and diabetes, this type of diabetes is called cystic fibrosis related diabetes because this type of diabetes is different from the diabetes found in people who do not have CF. The treatment for CFRD is also different

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Summer 2001

CFRI Aspirations

A Newsletter for Teens with CF and Their Siblings and Friends

Published by Cystic Fibrosis Research Inc., Bayside Business Plaza, 2672 Bayshore Parkway, Suite 520, Mountain View, CA 94043, Phone: 650-404-9975, Fax: 650-404-9981, Email: cfri@cfri.org

14-year-old Michelle Jones Gets to be a Model for the Vest!

Michelle Jones, age 14, got the Vest last winter. Her family set up the Vest next to the piano, so Michelle found herself sitting for 30 minutes two times a day on the piano bench. Michelle had played piano when she was younger, but as she entered her teens, she became less interested in piano and had stopped playing. She had started up again just before getting her Vest, but wasn't especially passionate about practicing. But with so much time on the piano bench, she found herself actively practicing piano again (and enjoying it) while she was doing her Vest treatments.

Michelle with her Vest.

Once Advanced Respiratory (formerly American BioSystems) supplies patients with the Vest, they call the family to make certain it is being used. Patients are asked to read off the numbers on the bottom of the machine. This tracks how much the machine is used. When Advanced Respiratory called the Joneses, the reps remarked on Michelle's usage numbers. They asked her mother how she got her daughter to be so compliant. Mary responded, "I don't do anything…I just put it by the piano and Michelle got hooked on piano. She plays every time she puts on her Vest."

Advanced Respiratory was impressed with Michelle's way of getting herself through her treatments (although Michelle doesn't think of them as a chore). They felt her story would inspire other teens learning to use the Vest. What if more people thought of the time on the Vest as an opportunity to sit down and develop a skill?

Last April, they asked her mother if Michelle would be interested in participating in a photo shoot and being featured in some of their advertisements and informational fliers. Michelle was excited with the idea of being a model.

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Dear Reader,

Hello! My name is Katie Weber and I am the teen editor for *Aspirations*, a newsletter from CFRI for young people. I am thirteen years old and just finished eighth grade. I have two younger siblings with CF. Patty is ten and Stephen is seven. I don't have CF.

I got involved with CFRI last year when my mom took me with her to the CFRI conference in San Jose. I wanted to learn more about what was new in CF technology and medicines. When I first arrived, I felt a little out of place. I was just twelve, surrounded by adult strangers. Once I met some people though, I felt more confident. Later, I met Calvinia, the Executive Director of CFRI. She and I hit it off. I told her how I wanted to be more involved and that there weren't many opportunities for young people. That got Calvinia thinking. We started brainstorming about ways I could get involved. All that led to this newsletter!

So, here's our premiere issue of *Aspirations*! We chose this name because it says how important breathing is to people with CF. But, it also says how much we want, plan and hope for in life. I want to thank everyone I've met so far. I can't wait to hear from more of you. I need your help now, Reader! These next few months are going to be a bit bumpy, but you can help smooth the path. Send us your pictures, questions, poems, funny stories and any idea that you think other 9 to 16 year old people with CF or their siblings might be interested in.

Can't Wait!
Katie Weber
CFRI Teen Editor, Aspirations

P.S. Seize every opportunity!

Michelle Jones *(Continued from page 7)*

Michelle says the whole thing was very exciting. She says, "I got to take a day off school. The photographers came to my house and they changed the living room around to make it more like a photographic studio and to make room for their equipment. I felt very nervous and self-conscious at first, but soon I became even a little full of myself and started smiling for the cameras. The photographers were really nice and explained what they were doing and they took their time. It helped me to relax. They explained that the photos would help to inspire other kids and that made me happy. Then we took a walk down to some open fields with my dog named Sandy and took a few more photos. Those pictures are my favorites.

"Just recently I got the final pamphlets, fliers and folders from the Vest company with my picture on them. I felt really proud of myself (and also of my dog!). I realized that modeling is really tiring even though it doesn't feel like you are doing anything. I also realized that one-half hour twice a day really adds up and can be really tedious, but in the end there are rewards. It's helping my health and helping me to live longer *and,* in my case, I got to be a model too!"

Michelle Jones is 14 years old and just graduated from eighth grade. She lives in Montara, California. ■

Michell and her dog Sandy.

For Patrick Who In the 90's

For Patrick who in the 90's eyes shone clear, deep blue and unflickering.
Powdery, ashy locks swept aside. Bubbling, tickling laughter spilling
from baby teeth and tongue.

For Patrick who in the 90's lives with death, floating around on tainted cells.
Under perfect ivory soap skin live poisoned genes.

For Patrick who in the 90's lives on roads paved in vials and bottles and medicine.
Tiptoeing around boundaries. Watching through glass walls at what he can not do

. We wear the armor of youth, but mine is rusted.

For Patrick who in the 90's struggles forward while I, stagnant, pray for a cure.

By Alice Ryan, now age 19, big sister to Patrick who is currently 8, has CF, and is doing well.

Goodbye Summer

Goodbye summer,
Hello dazzling cold winter rain that
Splatters on the dark cement.

Goodbye summer,
Hello to the holiday season that is so
Much fun for me.

Goodbye summer,
Hello to the nice fluffy tissues
For my nose.

Goodbye summer,
Hello to lots and lots of homework
That is taking over my life!

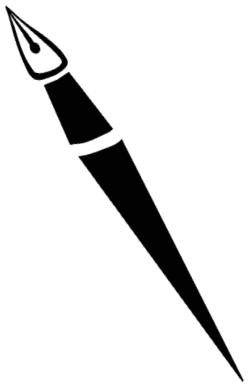
Goodbye summer,
Hello to reddish leaves falling swiftly and
Silently through the air.

By Devin Wakefield
Age 10 and has cystic fibrosis.

A Teen with CF

Being a teenager with CF isn't very easy. Everyone thinks I have it bad but I feel sorry for the people who say that. I have to admit I complain sometimes but it isn't all that bad. Through CF I have come much closer to my family than most other people without CF. I've been taking medicine so long that I'm really very used to it. I just have to remember to set my mind to it and maybe that will help make me better. I don't have to go into the hospital very much. I think the reason for that is because my family and I try to do everything the doctor says. Lately, I have had to go in more often, probably because I'm getting older. I'm still pretty healthy for someone with CF. My doctor said that I could take horseback-riding lessons, and that might help me with loosening the mucus in my lungs. I would still have to do my therapy, but anything that my doctor suggests might help, I will try it. I hope that in my lifetime, hopefully sometime in the near future, I will be able to see a cure for CF.

By Krista McCarty
Age 14 and has cystic fibrosis.



For Larissa

Would you like to have
her disease?
No.
I would like to have
her trust.

Would you want
her struggle?
No.
I want
her compassion,
her courage,
her beauty.

Would you want
her pain?
No.
I want her love.
Most of all,
I want you to love me
for me,
and not for
the disease I was born
without.

By Christina Swanson
Age 19. Her sister Larissa has cystic fibrosis.

I Am

I am smart and athletic
I wonder if dead people get bored
I hear pets talking
I see pictures looking at me
I want a million dollars

I am smart and athletic
I pretend that I am a movie star
I feel the rocks moving around me
I touch the stars
I worry I might get very sick
I cry about my step-dad being dead

I am smart and athletic
I understand that you need an education
I say I believe in a cure for CF
I dream about fairies
I try to be respectful
I hope to win the lottery

I am smart and athletic

By Michelle Rose Sawyer, age 12. Michelle and her brother have cystic fibrosis. Michelle wrote this when she was nine.

Aspirations is a newsletter for adolescents with CF and their siblings and friends. It will be printed inside *CFRI News* for the Summer and Fall 2001 issues. After that it will be sent out to subscribers separately as its own newsletter. Would you like to receive ***Aspirations?*** Send an E-mail to **cfri@cfri.org** with your name, CF connection, and E-mail address, or clip out the following coupon and mail it in**.**

Yes! I would like to receive *Aspirations,* a newsletter for teens with CF!
Subscriber information:

Name: _____

Address: _____

City: _____ State: _____ Zip code: _____

Relationship to CF: _____ Date of Birth (CF patients only) _____

E-mail: _____

Would you like to receive *Aspirations* in the mail, or by E-mail? (Please circle your choice.)
Please clip out and mail this coupon to:

CFRI
2672 Bayshore Parkway, Suite 520
Mountain View, CA 94043

Send submissions of articles, poetry, photography, and artwork to Katie Weber, CFRI's teen editor, at **cfriteeneditor@aol.com,** or mail to Kathleen Flynn, CFRI, 2672 Bayshore Parkway, Suite 520, Mountain View, CA 94043. This is your newsletter, for and about kids and teens with CF. Your submissions can be about living and growing up with CF, or they can be about anything else that interests you. Let's make this newsletter your G-R-E-A-T newsletter!

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Summer 2001

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Maureen Lenssen
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Ella Morris
Patricia Moylan
Terri Muir
Shaynee Muldoon
Terri Mulreaddy
Janet Nanry
Solange Nascimento
Christal Nelson
Judy Nelson
Wanda Olson
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Paula Patty
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Nancy Pecor
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Heather Pescht
Barbara Peterson
Kelley Phillips
Jeanette Pierce
Arlene Podell-Barr
Kathleen Pollard-Vithanage
Charis Powell
Marie Powers
Florence Prater
Bonnie Predny
Dorothy Price
Terri Price
Bune Primack
Pascale Randolph
Heidi Red
Ann Reuscher
Sharon Riddle
Dea Roanhaus
Genevieve Robbe
Ann Robinson
Barbara Robinson
Marianne Robinson
Vicki Robinson
Renee Roeder
Katalin Rogers
Robert Rohde
Donna Romano
Phyllis Ruffner
Siobhan Ryan
Julie Salas
Betz Salmont
Bob Sample
Carolyn Samuels
Linda Sanford
Nancy Sawynsky
Kim Schlotterbeck
Shirley Schmeyer
Ruth Schneider
Valerie Schneider
Kathy Schoenholzer
Carolyn Seaton
Marily Senn
Gail Shak
Janice Shaul
Della Mae Smith
Judybeth Smith
Nicki Smith
Peggy Stacy
Bette Standon
Lisa Steiding
Anabel Stenzel
Isa Stenzel-Byrnes
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Allison Stockley
Pam Stone
Mary Stori
Tammy Striebeck
Shana Strohn
Susie Stuart
Norma Stuckert
Ann Swanson
Margaret Swanson
Sara Swanson
Delinda Syme
Chris Tacke
Anne Tarvin
Mary Kay Taylor
Leisa Tegner
Pat Thibault
Joanne Thompson
June Thompson
Lauri Thompson
Shell Trask
Mary Tripp
Tory Trisch
Kelly Tucker
Connie Turner
Judy Ussher
Aurore Vaeth
Siri Vaeth
Jane Van Dyke
Betty Vitousek
Sarah Vogt
Wendy Von Oech
Linda Walton
Claudia Watkins
Simone Wernli
Lucy West
Doris Wester
Rebekah Whicker
Angela Whipkey
Marie White
Nancy Wilkins
Calvinia Williams
Pat Williams
Tammie Williams
Reynel Wilson
Cindy Witman
Laura Woods
Lena Woods
Rhonda Woodworth
Jean Worley
Linda Yerves

Thank you for being a tea sender!

NOTE: *If you were a tea sender and do not see your name on this list, please know that this is just an oversight on our part and we really appreciate your efforts. Please call or E-mail the CFRI office immediately and let us know if we left you off our list.*

CFRI Poster Child Photo Recruitment

Have you ever wished your child was pictured on the Mother's Day Tea invitation or insert? Would you like your friends to see your child on our Special Gifts Holiday mailing or on the Annual Report? If you have a photo of your child that you think would be perfect for one of our mailings, **please send it in!** We cannot promise that it will be in the mailing of your choice, but we hope to use as many children, teens and adults with CF as possible in our mailers.

Requirements: For the Mother's Day Tea Invitation we need a happy picture of a child with CF either alone or with friends or siblings. For the insert we need several varied poses, alone and with parents, grandparents or guardians. For the Annual Report we need a head and shoulders shot of a child or adult with CF. In the Winter Holiday mailing, we hope to compose a collage of children and adults with CF (with or without family members). Color or black and white photographs are acceptable. No slides, no color xeroxes, please. Photo must be of good quality, clear and crisp. Please send a copy!

CFRI cannot be responsible for lost photographs, and we cannot promise to return photographs (we may want to save them to use in a mailing at a later date). Please mark the back of your photo with your name, your child's name, your address and phone number. We will contact you before use in one of our fliers in order to finalize photo release paperwork. Please do NOT send in a photo of your older child or adult child with CF without his or her permission! We can't wait to see the beautiful photographs of your child! Send to CFRI, attention Kathleen Flynn, 2672 Bayshore Parkway, Suite 520, Mountain View, CA 94043. ■

CFRI's 2001 Educational Conference— LAST CHANCE TO REGISTER!

This year's Educational Conference will be held August 10-12, 2001 at the DoubleTree Hotel in San Jose, California, located conveniently near the San Jose Airport. This year's highlights include:

- Frank Accurso, M.D., Denver Children's Hospital,
- Dana Hardin, M.D., University of Utah
- Tomas Ganz, M.D., Ph.D., University of California at Los Angeles
- Myra Bluebond-Langnor, Ph.D., Rutgers University
- Walter Robinson, M.D. Boston Children's Hospital
- Daina Kalnins, R.D., Toronto's Sick Children's Hospital
- Pete Challoner, Ph.D., Chiron Corp.
- Lisa Solinger, California Transplant Donor Network
- Panel of Teens with CF

***Please Note**: Discounts for hotel rooms are not available after July 12th.*

*To register for the conference visit our website at **www.cfri.org** or call the CFRI office at 650-404-9975.*

Postdoctoral Fellow *(Continued from front page)*

selective accumulation of *P. aeruginosa* in the CF airways must be sought.

Measuring Gene Expression in Living Airway Epithelial Cells Using Imaging Microscopy

Chandy also began to test whether *P. aeruginosa* bacteria regulate expression of the interleukin-8 (IL-8) gene (one of the most important genes involved in triggering the inflammatory response) through the release of soluble factors or through bacterial binding. We used a method that would allow visualization of both bacterial binding and gene expression in living cells.

Chandy and graduate student Kevin Hybiske generated DNA that leads to expression of a special enzyme (beta-lactamase) coupled to the portion of the IL-8 gene that controls its activation. Epithelial cells that have been transfected with this chimera are loaded with a fluorescent dye that changes color when it has been cleaved by the beta-lactamase, which is activated only when the cellular machinery responsible for activating the IL-8 gene has been turned on. They expected that if *P. aeruginosa* binding to the epithelial cells was involved in activating the IL-8 gene, then cells would change color only when they bound the bacteria. In contrast, if soluble factors released from the bacteria were involved, *P. aeruginosa* would trigger IL-8 gene expression in all the cells of the epithelial cell layer.

Early results indicate that *P. aeruginosa* binding is indeed important for up-regulating the IL-8 gene in CF airway epithelia, but this response occurs most prominently only when the bacteria bind the epithelial cell membrane that is closest to the blood stream (i.e., the membrane on the opposite side from the apical side that borders the airways). These results indicate that bacteria will elicit the largest inflammatory response when the barrier function of the airway epithelia has been compromised.

Kevin Hybiske is pursuing this project now that Chandy has moved to a permanent position at Stanford University. ■

Some Books for the Journey *(Continued from page 6)*

from the treatment for the more common type 1 and type 2 diabetes.

The guide lists symptoms of diabetes such as frequent urination, excessive thirst and consumption of liquids, excess fatigue, weight loss and worsening of pulmonary function. Anytime a CF patient has unexplained weight loss or difficulty gaining weight, the book suggests that the physician should test for diabetes. The Cystic Fibrosis Foundation recommends that adults with CF have a casual blood glucose test taken at least once per year. I wonder if this is aggressive enough. As researchers learn more about CF and its effects on the body, they have begun to notice cases where pulmonary decline has its roots in problems in another organ. This can certainly be true of CFRD. Dr. Niels Hoiby noted at a talk recently that youngsters at his clinic are screened for CFRD annually from the age of ten on. This protocol was developed after doctors in Denmark noticed a two-year pulmonary decline usually preceded a diagnosis for diabetes, and that the chance of developing CFRD started to increase after the age of ten.

The book explains the role of insulin in the metabolism of nutrients in food, the effect of insulin deficiency in CF, and the different types of insulin and how they work. One chapter is devoted to nutrition and CFRD. Reading this book is helpful in understanding the routine management of CFRD. The goal of this diabetes therapy is to maintain blood sugar levels as close to normal range as possible. As more adults with cystic fibrosis are living longer and being diagnosed with CFRD, it is important for everyone to be informed about this disease and to be on the lookout for the warning signs. ■

Honorariums

An honorarium is a donation made in honor of a living person.

Chelsa Aboud
Becky Aikins
Gianna Altano
Joseph Amato
Sadie Anderson
Beth Arvidson
Jessica Arvidson
Debra Babbitt
Jaimee Baker
Kyle Baker
Lucy Barnes
Jamie Barry
Brent Bennett
Earl Beydler
Ryan Bortz
Marci Boots
Sierra Boyles
Agnes Calleri
Kerry Canavan
Lauran Catron
Kelly Colgan
Nancy Collins
Shaun Collins
Cameron Cornell
Caroline Daly
Marty Detrick
Gordon DeVore
Jason Dolin
Nancy Dumke
Tess Dunn
Denise Farrar
Alana Fink
Lorin Fink
Scott Folger
Jason Foster
Ryan Foster
Emily Fredrick
Jessica Fredrick
Rhonda Fredrick
Rosalyn Goldman
Barbara Greenberg
Jate Haga
David Hardy
McKenna Hardy
TJ Hardy
Brendon Harrigan
Alyssa Harvey
Tyler Heavner
Mildred Held
Taylor Holm
Haley Horn
Michael Hospodar
Clark Huddleson
Christopher Ireland
Peter Judge
Mickey Kinsley
Lori Kipp
Norma Kipp
Eleanor Kolchin
Philip Kolchin
Santosh Krishnan
Steven Kusalo
John Lenox
Michael Livingston
Gail Lowenthal
Virginia Lyons
Sandra Mardigian
Jim Marocco
David Martin
Claire McCabe
Sam McDonough
Carly McReynolds
Dolores Mendoza
Jackie Merrill
Kaylee Mincey
Fiona Mischel
Alex Molle
Marco Morrill
Kathy Morse
Jacob Osterholt
Aly Maris O'Reilly
Madden O'Reilly
Erica Pescht
Tanza Pescht
Erin Phillips
Kim Piscitello
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Robin Primack
Cynthia Rappaport
Larry Rappaport
Briauna Red
Melissa Reta
Michael Reuscher
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Ben Robertson
Carl Robinson
Randy Rupraat
Siobhan Ryan
Ben Sanford
Erika Schlotterbeck
Mary Schoti
Janice Shawl
Sarah Skeffington
Brian Smith
Robin Stephenson
Rachel Stori
Ewen Syme
Larissa Swanson
Allison Tarvin
Heidi Tegner
Adam Thompson
Johnny Tucker
Jennifer Uskouski
Tess Vaeth
Betty Vitousek
Mat Vitousek
Robert Vogel
Devin Wakefield
Christopher Walton
Helen Weiss
Nicky Westell
Josh Westgate
Kay Wilson
Kelly Wilson

Memorials

These memorials come to us not only for victims of CF but for their families and relatives as well. We extend our deepest sympathy to their families and friends. These gifts have given new hope to children and adults with CF. *Note: Occasionally someone who has died had the same name as a living person.*

Kimberly Adleman
Erika Alm
William Anderson
Eric Bathen
Bob Beach
Anne Beltrame
Amy Bienenstock
Jack Black
Cecil Bond
Greg Brazil
Kristi Burnham
Kyle Butler
Tim Calero
Bernice Chappell
Cindy Clark
Louise Clarke
Sharon Collins
James Contzen
Jim Curry, Sr.
Nancy Curry
Joshua Dadami
Donald Davis
Judy Dawson
Neva DeVore
Jimmy Ervin
Pam Faulk
Marietta Ferreira
Dr. Max Filling
Mrs. Max Filling
Stephanie Gaggs
Dennis Gallagher
Russell Gallaway
Harold Goff
Isaac Goldman
Diana Goodman
Douglas Graham
Leisha Graves
Cynthia Haley
Stephanie Halling
Troy Hampton
Dorothy Herchelroth
Asheligh Hindman
Rose Horn
Stephanie Huff
Phyllis Jones
Kathy Judge
Jeanai Kodis
Michael Kozlowski
Glenn Linebaugh
Mamie Lilly
Duane Little
Dawn Longero
Donald Lucia
Alex MacDonald
Lucy Marsh
Jason Marshall
Jordan Marshall
Clint McDonnell
Barb Ann McGhee
Jessica Mobley
Emma Montna
Cecelia Morris
Scott Nelson
Bernice Newsome
Dick Odgers
Michele Olson
Mary Ann Pedersen
John Prater
Tim Prater
Antonia Prokop
Myrna Ratkovick
Mildred Redding
Pat Rice
Harry Rinsky
Sandy Robbe
Ben Roberts
Pamela Rockhold
William Ruffner
Michelle Sanderson
Tony Saso
Dhea Schalles
Sandra Schukle
Larry Shaw
Tammy Smerber
Adrianus 'Ed' Sneep
Tony Stewart
Dave Stuckert
Laurie Stuckert
Dresden Tingly
John Trask
Marge Trask
Tony Trigueiro
Linda Trojak
Katherine Urbani
Cindy Vidak
Joy Villasenor
Keegan Wahler
Charles Wallace
Thomas Walton
Viola Wastell
Helen Weil
Tara Weir
Hayley Wester
Rudolph Wiesemann
Herbie Wilfong
Rachel Williams
David Wolf
Agnes Woltmanns

How to Submit an Honorarium or a Memorial in Honor of Someone You Love

Do you have a relative or friend that you would like to honor? Are you searching for the perfect birthday present, wedding or anniversary gift or memorial? Give a gift with lasting impact. Make a donation to Cystic Fibrosis Research, Inc. We'll send a special message to the recipient or to the deceased's family informing them of your contribution if you give us the name and address of the family. Your donation not only recognizes your loved one's special occasion, it benefits children and adults with cystic fibrosis and their families as well. Mail your contribution with the name, address and occasion of the person you are honoring or remembering to: CFRI, 2672 Bayshore Parkway, Suite 520, Mountain View, CA 94043

Put Your Favorite Recipe into Print!

The Lucile Salter Packard CF Center at Stanford is developing a family style cookbook with easy to prepare and fun to eat foods. Please share your favorite recipes with us in any category! Along with your list of ingredients and cooking instructions, be sure to include the number of people the dish serves, the serving size per person, preparation time, cooking temperature and time, and any other pertinent comments about the recipe.

Simply cut out your recipe information below and either send or fax to…

Fax:
Attention: Judy Kirby
(650) 723-5201
Address: Judy Kirby or Julie Matel
701 Welch Rd. Suite #3328
Palo Alto, CA 94304

Intestinal Blockages *(Continued from page 5)*

When our son feels a blockage is coming on he knows from past experience that it is serious and he knows that the consequences can be quite painful if he ignores the signs. He feels sluggish. His digestive system slows down. He misses having stools at his usual frequency and loses his appetite which is normally quite large. At this time he makes certain all of the preventative measures listed above are being followed and he starts to monitor himself very closely.

1. He begins eating smaller, more frequent meals. He includes more roughage, while drinking more fluids.

2. He takes PeriColace, which in addition to being a stool softener also includes a bowel motility drug.

3. He takes the laxative Magnesium Citrate Oral Solution, a saline laxative that is available over the counter in several flavors.

4. He looks for signs of improvement in his condition.

When a full or partial intestional blockage occurs, the CF child or adult is in acute pain and the above procedures have not been effective. It is imperative to notify a physician at this point. Sometimes the physician will prescribe a milk and molasses enema. However, these have never been effective for our son because his blockages usually occur higher up in the intestine. His physician then prescribes a treatment of GoLytely®. GoLytely is a powder that you add water to right before using. You drink glasses of it over a prescribed time period. This is difficult as abdominal pain may be present and no one would feel like eating or drinking. The taste is also not especially encouraging! But it really works as a very effective cleanout if you can get it down. We keep a prescription of this on hand and have even taken it with us on vacation.

The availability of Miralax (polyethylene glycol) has introduced an important alternative to GoLytely. It is much more palatable. You just dissolve a packet in a big glass of water and drink it down. GoLytely, however, is the preferred method for more acute situations. Miralax takes longer to clear an obstruction. It is usually taken over several days and it may be as long before one is truly cleaned out. This may be ok if you are having some uncomfortable constipation, but it would not be an option if you truly have a partial obstruction. Some patients with chronic propensity to DIOS take Miralax on a regular schedule (eg. once or twice a week) to prevent DIOS.

If the above treatments are not effective, our son then calls his physician who recommends that he proceed immediately to the hospital. X-rays confirm the blockage and a gastrograffin enema is administered. In extreme cases, surgery may be required.

It is very important that your or your child's physician be involved in the protocol that you create for yourself or your child in dealing with intestinal blockages. The younger or smaller the individual, the easier it is for dehydration to be both a contributing factor to the blockage and a resulting factor. If your youngster is not stooling and not eating, get help quickly. Dehydration is dangerous. Additionally, if left untreated, the bowel can erupt and the poisons in the bowel can spill into other parts of the body. This is a life-threatening situation. Time is essential in treating these obstructions, but with careful management and a hand-crafted protocol developed with your physician, many of these early symptoms can be handled more quickly and unobtrusively from home. ■

Genentech, Inc. is Supporting Faster Delivery System for Pulmozyme!

By Kathleen Flynn

Invacare Corp., a company out of Elyria, OH has joined forces with Genentech and Medic-Aid (an English company) to make and supply a nebulizer that cuts aerosol treatment times in half! Pulmozyme® delivery with the Invacare® Sidestream® Nebulizer, when used with either the Porta-Neb® compressor or the Mobilaire ™ 50 PSI compressor takes on average only two minutes and 16 seconds! The Sidestream Nebulizer has been designed to reduce medication particle size and boost respirable output. It produces medication droplets as small as 2 microns. The smaller the particle size, the better chance the medication has of reaching the smallest airways.

A year ago an agreement between Genentech and Aerogen for developing a secondary delivery system for Pulmozyme fell through. It is welcome news to hear that Genentech has found another way to support a faster Pulmozyme delivery. Many people are using these nebs and compressors for faster TOBI® delivery as well. People with CF have all struggled with adherence issues with their numerous daily aerosols because the delivery process takes so long. The pressure to find the time for treatments between school, work and family life is enormous. Any reduction in time required for these therapies is a welcome break.

Here's the catch. These products are not cheap. The nebulizers run about $25.00 each. (They last approximately one year and they can be cleaned in the dishwasher.) The list price for the Mobilaire is $383.00. Your insurance company (if they will cover the cost) will probably get a contracted (therefore reduced) rate for both items. Kristin Shelton, Head of Respiratory Therapy for the CF Center at Stanford, has found that some insurance companies are covering this equipment as Genentech is recommending their use with Pulmozyme. If you are interested, start by talking with the respiratory therapist at your CF Center and see if the RT can help you get these through your insurance company. If they won't cover these (and you have the funds), you can still purchase them on your own without a prescription (but *please* discuss this with your CF physician first). Both the nebulizer and the compressor are being distributed in the United States to local suppliers by Invacare Corp. Call Invacare's customer service people (800-333-6900). The Sidestream Nebulizer product number is MS2400 and the Mobilaire product number is IRC607. They will ask for your zip code and send you to the dealer nearest you. If you're lucky, the dealer may give you a better price than I quoted above (which is list price). The dealer may have to order these and the Sidestream Nebulizers can only be ordered in boxes of ten nebs. Check ahead to make sure you only have to purchase one or two from the box. For more information, visit Genentech's website at **www.gene.com** or Invacare's website at **www.invacare.com.** ■

Action Coupon – Become a Member of CFRI

CFRI is dedicated to sponsoring CF biomedical research and education about CF. When you become a member of CFRI, you join a community of concerned people who work together to fight cystic fibrosis. Your annual dues of $15 (or lifetime membership of $50 per person) entitles you to vote on the allocation of CFRI research funds and run for a Board of Directors position. All members also receive CFRI News*, a newsletter providing the latest in CF research information and social news to the CF community. To join CFRI, simply complete the Action Coupon below and mail it to our office.*

CFRI's mailing list is confidential. We do not sell our list, nor do we give out any names or addresses under any circumstances. Our mail solicitations include our personalized Mother's Day Tea mailing and a year-end Special Gifts request. If you receive other solicitations for money, they are from organizations other than Cystic Fibrosis Research, Inc.

☐ Here is my annual membership for $15 per person.
(Contributions above the $15 annual subscription rate will help offset costs for those unable to donate.)

☐ Yes! I want to help. Here is my check for_____.

☐ Here is my lifetime membership of $50 per person.

☐ Yes, I want to send Mother's Day Tea invitations in 2002.

☐ Please send me _____ invitations!

☐ Please remove my name from your mailing list.

Name | Relationship to CF Person | Date of Birth (for CF patients only)

Street

City | State | Zip | Telephone | E-mail

What do you like about our newsletter?_____

What would you like to see us improve in our newsletter? _____

Please list any ideas for future articles, regular columns or subjects you'd like to see featured here:_____

ABOUT CYSTIC FIBROSIS AND CFRI

CF is the most common life-threatening, hereditary disease in the U.S. Twelve million Americans are symptomless carriers of the CF gene. One in 2,300 American children is born with CF. Symptoms may include:

- persistent cough, wheezing or pneumonia
- loose, foul-smelling stools
- heat prostration
- abdominal pain
- excessive appetite, but poor weight gain
- salty-tasting skin
- clubbed finger tips

CF patients suffer from chronic lung disease and digestive disorders which may require frequent hospitalizations. At present, only half of those with CF survive to age 32.

ABOUT OUR FUNDRAISING

Cystic Fibrosis Research, Inc. raises funds through a number of channels:

- Personal mail solicitations
- Memorials and Honorariums
- Grants
- Special fundraising events
- Membership fees
- General donations

CFRI's E-MAIL ADDRESS AND WEB SITE

CFRI's E-mail address is: **cfri@cfri.org**. Use this address to obtain information about our organization, the latest word on cystic fibrosis, or to communicate with our office. Also check out our Web site: **www.cfri.org.** See you on the Internet!

ABOUT OUR SERVICES

CFRI offers the following services:

- Research funds awarded to scientists at major medical research centers
- Educational meetings for the CF community
- Annual CF Educational Conference
- Annual CF Retreat for ages 16 to adult
- Family support groups
- CF support groups for ages 16 to adult
- *CFRI News* and *Aspirations* newsletters
- Email and telephone referrals and support services
- Videotapes of meetings and conferences

ABOUT OUR SUPPORTERS

CFRI is a nonprofit, volunteer organization not affiliated with any other group. Our support comes from the dedicated volunteers and staff including:

- Executive Director, Program Services Manager, Outreach Specialist (part-time) and one Administrative Assistant
- Research Advisory Committee of physicians, scientists and members
- Board of Directors and Executive Committee
- Advisory Committees
- General membership
- Mother's Day Tea senders
- Office volunteers

Special Thanks: CFRI wishes to thank PathoGenesis Corporation, makers of TOBI®, for their generous support of *CFRI News*.

CFRI
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Mountain View, CA 94043
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