

# CFRI NEWS

*An Educational Community Contribution by CFRI*

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## Development and Implementation of a High-Throughput Pseudomonal Adhesion Assay

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As antibiotic resistance in cystic fibrosis patients infected with *Pseudomonas aeruginosa* becomes a more ominous problem, novel, non-antibiotic therapies must be discovered. One group of such agents is those that inhibit bacterial adhesion/initial colonization. Although the binding site for bacterial attachment has not been defined, several lines of research suggest that the binding site is some type of carbohydrate moiety. In addition, some evidence has shown that certain carbohydrates can interfere with this interaction and may lead to improved clinical outcomes.

Our lab is developing a high throughput assay to rapidly screen for the ability of similar agents to inhibit adhesion to epithelial cells and/or mucus. It is believed that mucus acts as the lung's physical barrier against infection by capturing the bacteria and clearing them through natural ciliary action. However, in CF patients, the mucus is not efficiently cleared. This allows time for the adherent bacteria to potentially penetrate the mucus layer and invade the epithelium.

As both substrates may play critical roles in initial colonization and/or subsequent dissemination of infection, we will analyze the effectiveness of carbohydrates to block attachment of bacteria to either surface. In our adhesion assay, we have utilized an immortalized, adherent epithelial cell line grown in an opaque 96-well plate in combination with several *P. aeruginosa* laboratory strains carrying a bioluminescent reporter plasmid suitable for constitutive replication in *Pseudomonas*. Bacteria were applied to each well containing epithelial cells and incubated for 1 hour at 37°C. A pre-wash luminometer reading was taken. Non-adherent bacteria were then washed, and the remaining adherent bacteria were counted using a luminometer. The relative light units (RLUs) were then

converted into colony-forming units (CFU)/mL after creating a standard curve for each bacterial strain tested. When we compared the adherence of wild-type *P. aeruginosa* with a pilin mutant (pilin plays a major role in the adhesion of *P. aeruginosa* to epithelial cells), we were able to demonstrate a threefold decrease in adherence over a wide range



Britta Swanson, Ph.D.

of bacterial concentrations ( $P < 0.05$ ). Similar results were achieved using two other strains of *P. aeruginosa* and their specific pilin mutants, which produce different virulence factors and presumably different adhesions.

The addition of certain carbohydrates to the binding reaction inhibited wild-type *P. aeruginosa* from adhering to the epithelium by varying degrees based on concentration.

We are currently preparing multimers of the carbohydrates in order to increase the effect of the carbohydrate inhibition. Next, we will test the ability of this carbohydrate to compete with mucus for binding bacteria, and we will test its effectiveness in our mouse model of pseudomonal lung infection. It is our goal to save mice from lethal infection by *P. aeruginosa* by blocking the initial colonization step in the lung.

In summary, we have created a rapid, sensitive, quantitative screening technique for testing the ability of molecules to inhibit *Pseudomonas* epithelial adhesion. Our future plans include continued carbohydrate screening and broadening the scope of inhibitors. ■

**RESEARCH**  
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## CFRI News

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## Message from the Executive Director

Your assistance and support is encouraging now more than ever. The images from September 11<sup>th</sup> will continue to have a direct impact on the lives of individuals around the world. As families and friends unite across the globe, our thoughts remain with the victims, their families, and relief workers. This is a challenging time for all of us – that is why I have dedicated a collection of my poetry that is befitting for the moment.

Let's all come together for a better tomorrow!

### *Dreams and Visions*

Can't we just stop and smell the aroma of peace  
A place where one goes to stop all order  
A place full of humility and tranquility  
A place of love and joy?  
Life and all its complexities are so hard to endure  
Our bodies are scored with open wounds  
Confusion has taken the minds of many  
Time is the absolute value of what we make it  
We must enjoy the pilot of peace for the time is not long  
Let's scurry through the depths of no return  
My dreams are embraced with the courage of hope and visions  
My visions are made up of things to come  
Many times I can't remember what my thoughts were  
I only know it will come to pass.

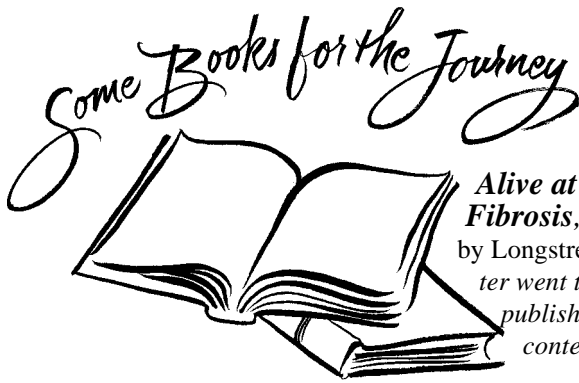
### *A Country of Hope*

As we reflect let's not forget we are a country of hope  
A country that has endured much  
A country of great leaders  
A country made up of past achievements  
Yes we are Americans that strive for better  
As we stand together let's remember we have each other.

### *The Numbness of Life*

My face is soaked with the rivers of tears  
I cry out for help  
The rumble is too thick  
None hear my voice  
Darkness is all around me  
My breathing has gotten short  
I remember an hour ago I kissed my family and said I will see you later  
Don't forget to take the food out of the freezer  
I'll cook it tonight, when I return.  
How can this be?  
Must I keep digging or should I do nothing?  
Reflections of my life soar, faces of my parents  
Family and friends creep into my mind  
I'm confused and scared  
Coldness has crept into my bones as my body lies numb  
I hear echoes of others around me  
God please keep me in this hour of terror  
Protect my loved ones I have left behind

(Continued on page 23)



**Alive at 25: How I'm Beating Cystic Fibrosis**, by Andy Lipman. To be published by Longstreet Press. (Note: When the Newsletter went to press, this book had not yet been published. The author has summarized the contents here.)

When Andy Lipman was seven, he read an encyclopedia article that changed his life. The article read, "People with cystic fibrosis do not normally live to the age of 25." Andy had always known his life was a little more complicated than his peers with his daily chest therapy and his medication, but he had no idea that his life was in such jeopardy. In fact, Andy's life was in jeopardy the moment he was born. His doctors told Andy's parents that he would be lucky to live to be a teenager because of his cystic fibrosis. How would Andy be alive at 25?

Andy lived a relatively normal childhood. He was a skinny boy who never really had any long-term goals. For one, he wasn't supposed to live very long. Secondly, he had to depend on his parents to administer his therapy. When Andy entered the tenth grade, a new type of chest therapy became available to him. It was a vest that permitted Andy to administer his therapy by himself. Now, Andy could consider going away to college.

Andy's dream of independence came true in 1991 when he arrived at the University of Georgia. This was what Andy had wanted for so long, but from the moment he got there, he felt out of place. He had a therapy machine to lug around when he went on trips. He had to remind himself to take medicine while his peers' biggest worries were buying textbooks for classes. Andy, in his own words, felt like a freak, and he began thinking that others felt the same way about him.

Andy became a recluse. He stopped going to class and stopped exercising. He spent weekends just lying on his couch in the dark. He stopped doing his therapy and stopped taking many of his medications. He wanted to die. He was getting sicker every day. His doctor noticed Andy's decline in health and tried to give him stronger medications to get him better. But Andy wasn't interested in taking any more medications. And he wasn't interested in getting better.

Then one day, Andy's life changed. He was playing some basketball, and while he played he was spitting up phlegm and leaning over to cough. He felt that he could go no further. Then Brett, a player from the other team, pushed him down to the ground. Andy lay there, trying to breathe, when Brett grabbed him by his bony arm and lifted him up, then laughed and said, "Joining any weightlifting contests any time soon?" Everyone around Brett laughed. Andy wasn't looking for pity anymore; he was looking for revenge!

That night, Andy decided that he was going to make some changes in his life. He began lifting weights. He attended his college classes again. He regained his stamina. Andy began making it his life mission to show his peers that people with CF can live long lives and that they can not only live with CF, they can beat CF too. Andy's lungs became healthier. He began running and has now run in five 10-kilometer races. He got stronger and has now lifted as much as 300 pounds. He started a softball tournament that benefited CF and has raised more than \$50,000 in 2 years.

On his 25<sup>th</sup> birthday, Andy began writing his story. While writing *Alive at 25*, he began answering several "tough" questions regarding mortality, fertility, relationships, and the dilemma of whether to tell people about his disease. *Alive at 25* is a sincere look at the life of someone who has had his share of ups and downs, but he will not give up.

(Continued on page 16)

## Thanks to Our Conference Sponsors for Their Generous Support!

CFRI's Annual Educational Conference would not be possible without the generous donations of our sponsors. Because of them, CFRI is able to offer this event at an affordable rate for our membership. Many people describe the Conference as informative and educational. The following sponsors contributed to our 14th Annual Educational Conference:

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Thank You



## 2002 CFRI Golf Tournament

The CFRI Golf Committee, chaired by CFRI board member Scott Hoyt, invites all golfers to the 18<sup>th</sup> Annual Golf Tournament in the summer of 2002 at the Cinnabar Hills Golf Club in San Jose, California. Help CFRI raise funds while having a great time! More details and information can be found on our Web site in the coming months.

# Kids Crave the Darndest Things (Grown-ups Too)

Need a new idea for dinner tonight? There's a cookbook just for you! The Lucile Salter-Packard CF Care Center at Stanford, in partnership with Digestive Care, Inc., makers of PANCRECARB® (pancrelipase), the only bicarbonate-buffered delayed-release enzyme, is developing a family-style cookbook with easy-to-prepare and fun-to-eat foods. The cookbook, entitled *Kids Crave the Darndest Things*, provides both high-fat and traditional versions of recipes to satisfy the entire family. Themes include pasta perfections; meaty meals; vegetarian venues; soups, salads and sandwiches; and breads, muffins, and desserts.

Recipes can be mailed, faxed, or submitted online. Along with your list of ingredients please be sure to include preparation time, cooking instructions, cooking temperature and time, number of people the dish serves, and any other pertinent information concerning your recipe. Your name will be printed in the cookbook and you will receive a complimentary copy.

Recipes are being compiled through the end of the year, and the cookbook will be available in early 2002.

## SEND SUBMISSIONS OR REQUESTS TO:

Judy Kirby or Julie Matel  
Stanford CF Care Center  
Attn: Cookbook  
701 Welch Road, #3328  
Palo Alto, CA 94304  
(650) 723-5201 FAX



## The Teen Panel Discussion

By Lori Lindmeier

One of the biggest concerns a parent has for children is their happiness and emotional well-being. For the parent of a child with a chronic illness, that concern is magnified, I believe, in the knowledge that the child will have much more to deal with in the years to come.

Sunday morning, the third day of CFRI's 2001 Annual Educational Conference, started with a teen panel consisting of 6 young adults. They ranged in age from 13 to 20 years old. As I sat down to listen to them talk and answer audience questions, a part of me wondered if I needed to be there. After all, our daughter with CF is only six and a half, so I wondered what they could say that would change anything we already do with her.

The hour-plus exchange had us all, at times, laughing, shaking our heads in agreement, and wiping tears from our eyes. The subjects ranged from school issues and acceptance, to compliance with and responsibility for treatments, to when they first knew that CF could shorten their life.

The majority of the panel had issues regarding being accepted, and many had chosen to avoid talking about CF until they felt their friends wouldn't judge them. Acceptance from peers was a big issue, but a continuing theme always came back—the love and support of family. They knew their family would always be there for them. They all spoke of ways to help pass treatment time, and television was number one. All stressed the importance of exercise and taking the meds, but one girl, Christina Miller, actually recommended that we “allow” a little bit of slack-off time. In allowing our children to shorten or delay a treatment every now and then, we also allow ourselves as parents to release a little stress. It's difficult trying to keep it all together 100% of the time. It not only takes a toll on the families, it also adds to the stress of the child having to be on a strict regimen with no break to feel “normal,” which was Christina's point.

Though most of them spoke of having at least one non-CF best friend who understood and supported them all the way, there was overwhelming agreement that they all need to be with other CF friends. Though they don't attend camps for kids anymore because of cross-infection issues, after listening to this group, I could now understand the need to have friends in the same “place” as they are. When they spoke about meeting other people with CF and feeling as if they had known them forever, it was both heartbreaking and heartwarming at the same time. I don't want my daughter growing up without contact with others with CF, but I feel better armed with knowledge about cross-infection and will try to find a good and comfortable balance. I do believe there are options now for parents who fear physical contact between CF kids. The Internet is a great resource for both information and support groups—and those groups are growing.

The final question asked of the panel was about when they first knew CF could be life-shortening. I find it difficult to write this, as the emotional impact still runs high. We parents don't want to ever think about the possibility of losing our precious child and so we try not to think about that part, and we focus, rather, on how to keep them healthy and strong. To hear each of these kids, one by one, address this subject with so much maturity, so much grace, I have to say I was amazed at their grasp of reality. They all had a sense of assurance that the time they do spend here, no matter how long or how short, will be worthwhile. I will never forget their words of wisdom, the emotional impact they left, and their inspiration. I had a strong twinge of maternal pride for each of them, even though I'd never met them.

These young adults will all make a difference.

They already have with me. ■



Teen Panel with Jim Hampton

# Gene Therapy Study in the Sweat Glands of Individuals with Cystic Fibrosis

Contact Person: Charlene Sharon Correa  
Clinical Research Coordinator

Cystic fibrosis (CF) is a hereditary disorder caused by mutations of the cystic fibrosis gene. It is the most common lethal hereditary disorder in the U.S.A. There is no cure for CF; current management strategies target symptoms only.

In 1989, the identification of the gene whose mutation is responsible for the manifestations of CF opened the door to new strategies for treating CF in which the normal gene is transferred to the most affected cells of individuals with CF. Researchers at the Weill Medical College at Cornell University in New York City are pursuing gene therapy for the lung manifestation of CF. In their prior studies, they have administered the normal CF gene to the cells lining the lung airways (windpipes) and have observed successful transfer of the normal CF gene. However, it was technically impossible to evaluate the function of the normal gene.

An upcoming study by this gene therapy research group is focused on answering the question, does transfer of the normal CF gene result in correction of the abnormal function of the cells lacking the normal gene? It is well known that the sweat glands of individuals with CF produce sweat with abnormally high amount of salt (NaCl) and have a low production of sweat (low sweat rate) upon stimulation with standard medications. Since the sweat glands are easily accessible, the researchers propose to transfer the normal CF gene to the sweat glands. Their hypothesis is that the transfer of the normal CF gene to the sweat glands of individuals with cystic fibrosis will reverse the abnormally high sweat chloride concentration and abnormally low sweat rate that are characteristic of individuals with CF. This is a "proof-of-principle" clinical study to demonstrate that gene therapy can correct the abnormalities resulting from the presence of an abnormal CF gene.

The site of the study is New York Presbyterian Hospital. Its results will represent a very important step in the process of developing strategies to cure or ameliorate the clinical manifestations of CF.

A total of 15 individuals with a clinically confirmed diagnosis of cystic fibrosis are needed. Standard screening and clinical laboratory tests will be employed.

You may qualify as a participant if you have cystic fibrosis, are over the age of 18, and are not taking immunosuppressive medications.

## ***Procedures include:***

Complete physical exam

Blood and urine tests

Electrocardiogram (EKG)

Chest X-ray

Breathing tests

Skin biopsy

Sweat chloride test

Sweat rate test

Participants will be compensated as follows: \$50 per sweat chloride test, \$50 per sweat rate test, \$50 per skin biopsy.

Travel expenses will be reimbursed.

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## ***Participation involves:***

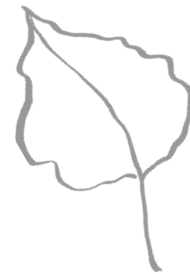
6 to 8 hour screening test

One-week hospital stay at New York  
Presbyterian Hospital

One half-day follow-up, outpatient  
visit after one month

## CFRI's 2002 Annual Educational Conference

CFRI's 2002 Educational Conference will be held August 9-11, 2002, at the Double Tree Hotel in San Jose, California. The DoubleTree Hotel is located near the San Jose Airport at 2050 Gateway Place, San Jose. Please set aside this date on your calendars and look for more information on the Conference in the year 2002.



## Be a Mother's Day Tea Sender!

The Mother's Day Tea last spring brought in close to \$250,000. If you would like to be part of next spring's effort, please call the office immediately at (650) 404-9975 for details on ordering the supplies needed. There are people waiting for a cure. Help us to help them by sending Tea invitations to your friends and family. It's easy and fun!

*It's never too late to be a Tea sender!*



## What is Wrong with Airway Secretions in CF & An Overview of Basic Research –Tomas Ganz, M.D., Ph.D.

By Pam Heilman

Thomas Ganz, M.D., Professor of Medicine and Pathology at UCLA, made two presentations at the CFRI Conference. The first was “What Is Wrong with Airway Secretions in CF?” and the second was “An Overview of Basic Research.” The first was a precursor to the second, as understanding the disorder in the secretions helps to understand where research is headed and why.

The discussion on Friday night, August 10<sup>th</sup>, centered on the fundamental hypothesis that Dr. Ganz adheres to, which is that the secretions in CF lungs cannot kill bacteria and therefore are abnormally susceptible to airway infections. “We see a different colony of bacteria not seen in other diseases.” The biggest problem in fully understanding the CF secretions is that there are no good models to study. Mice do not have the same glands that humans have to make secretions. Consequently, when mice are altered genetically to have CF, they do not have the same type of lung disease that humans have.

The lung secretions are the first line of defense for the lungs, and when they cannot effectively eradicate the bacteria and, with the aid of the cilia, move them up and out of the lungs, the white cells move in and eat the bacteria. Large numbers of white cells are characteristic of CF. Therefore, Dr. Ganz believes that secretions from the nasal epithelial lining can be used for this research as they are essentially the same as those in the lungs. In order to have a complete scientific study, however, researchers need a control group without infections. The only group of CF individuals who would fit the criteria is newborn infants. But newborns are difficult to study for a variety of rather obvious reasons. Dr. Ganz believes a control group of asthmatics might work, as they are similar in many ways to CF patients.

Basic research is currently considering two hypotheses related to the secretions in CF lungs: **1)** The Mechanical Hypothesis, which supposes that the problem is the volume and consistency of the mucus. CF mucus is dehydrated and sticky. Thus, inhaled bacteria cannot be removed and become a stimulus for infection.

**2)** The Chemical Hypothesis supposes the secretions have an antibacterial impairment, which results in an exaggerated inflammatory response. The debate between these two hypotheses may seem a bit like splitting hairs to a parent waiting for a cure. However, correcting the defects, whether chemical or mechanical, will depend on two very different therapies.

So where is CF research heading? The target is going to be the submucosal glands because this is where the antibacterial activity happens. There is more evidence to support the chemical hypothesis at this time.

In conclusion, Dr. Ganz stated, “CF is a complex and subtle disease where patients can remain healthy for a long time. It is a system that almost works.” There is a need for a better animal model, one that has the submucosal glands and can get the same type lung disease when genetically altered. Dr. Ganz also said, “Based on current knowledge, focus on infections is warranted.” ■



Tomas Ganz, M.D., Ph.D.



# Ambry's New Gene Test Identifies 900 CF Mutations

By Charles Dunlop  
President, Ambry Genetics  
Costa Mesa, Calif.

Word has traveled quickly that Ambry Genetics has introduced a test that can identify more than 900 mutations of the cystic fibrosis gene. Other DNA screenings look for 87 mutations at most. The Ambry Test™: CF is also capable of discovering previously unknown mutations that could lead to the disease.

Within days of making the announcement on October 17, Ambry had calls and e-mails from as far away as England and Argentina.

The test employs DNA sequencing technology similar to that used in the Human Genome Project. Improvements we've developed make the analysis faster, more comprehensive, and more cost-effective.

Technical details aside, what has really touched my heart since the announcement is hearing the urgency that people feel who are confronted by this disease.

For instance, I received an e-mail from one mother who has a five-year-old daughter with "questionable CF." The young girl has had two positive sweat tests and has had pneumonia five times in six months, but other gene tests so far show no mutations. "Her doctors won't treat her like she has CF until they know about the genes," her mother wrote. "Please consider [my daughter] for your tests. Thank you from a very desperate Mom!"

Once we receive a blood sample from the daughter, I'm confident we'll be able to specify the mutation the daughter may have, or rule out the disease altogether.

Procedures for detecting gene mutations can be separated into two distinct groups. The first are "single nucleotide polymorphism" (SNP) technologies that efficiently detect common, well-known disease-causing alleles. The second group consists of fully sequencing the gene in search of any mutation that might be there, including known or unknown disease-causing alleles.

Virtually every clinical genetic test today is an SNP technology. These tests have traditionally had the advantage of being able to detect known mutations inexpensively and with high reproducibility. The problem with these technologies is their inability to detect rare disease-causing mutations that can account for a significant number of diseased individuals, especially in different ethnic groups.

Traditionally the scanning and full-sequence methods have been expensive and/or



*Charles Dunlop, President, Ambry Genetics Lab*

## The Experts' Corner

The Experts' Corner is a new addition to the Newsletter: It features articles written by professionals considered experts in their field of knowledge.

### A Moldy World...

Mold has been getting a lot of press lately. From Erin Brockovich's moldy house to a myriad of newspaper, magazine, and Internet articles, this primitive organism has become a popular environmental health concern.

Mold growth can occur in buildings because many building materials are made of organic substances, such as wood, paper, and vinyl, that are food sources for mold. What usually prevents mold growth on building materials is a lack of water. If water becomes available by direct saturation (leaks/floods) or high humidity (greater than 60 percent), mold can begin to grow.

Mold reproduces by microscopic seed-like units called spores that are easily dispersed by air movement. Mold spores that encounter damp conditions produce root-like structures that search out food sources. If sufficient moisture and food are present, mold enters a reproductive phase where it produces more mold spores as well as "microbial volatile organic compounds" (MVOCs). MVOCs are what give mold its musty odor.

Most commonly, spores from excessive mold growth in buildings can cause hay fever or aggravate asthma in sensitized individuals. Exposure to MVOCs can also irritate the eyes, nose, and throat.

The Environmental Protection Agency (EPA) recently released new guidelines entitled "Mold Remediation in Schools and Buildings," which are available on the Internet at <http://www.epa.gov/iaq/molds>, [www.epa.gov/iaq/molds](http://www.epa.gov/iaq/molds).

These guidelines present strategies to:

- respond to flood emergencies, such as pipe breaks or sewage back-ups, to prevent mold growth

*(Continued on page 19)*

*(Continued on page 8)*

- assess mold growth that has occurred
- remediate mold growth under controlled conditions that protect workers and occupants

The most important part of mold assessment is a thorough visual assessment to quantify the amount of mold present and identify the source of water intrusion. The EPA guidelines recommend three levels of remediation control, depending on the amount (square feet) of mold-damaged building materials. The EPA guidelines strongly emphasize the importance of eliminating the source of moisture intrusion; otherwise mold growth can reoccur.

The government does not regulate professionals that engage in old assessment and remediation. It is therefore worthwhile to carefully select mold consultants and remediation contractors. Having a Certified Industrial Hygienist on staff is a good (but not certain) indication that a firm will make a good mold consultant. Mold remediation contractors should be able to show they have appropriate hazard communication, respiratory protection, medical surveillance, and personal protective equipment programs for their worker.

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## The Annual CFRI Teen and Adult Retreat 2001 —Another Year of Survivors

By Ana Stenzel

The Annual CFRI Teen and Adult Retreat of August 5-10, 2001, was outstanding for several highly successful structural innovations and a renewed sense of enthusiasm and energy. Held at Vallombrosa Retreat Center in Menlo Park, California, the Retreat was marked by an influx of first-time attendees. There were 62 people who attended Retreat for at least some of the time, including 28 people with CF (11 teens) and 19 drop-ins. Most of the participants were from the San Francisco Bay Area, although two came from out of state. Individuals without CF included parents, siblings, partners of people with CF, and volunteers. Scholarships were available for those who requested them.

This year's theme was "Survivor: Live, Learn, and Thrive," a takeoff on the hit TV "virtual reality" series. Social activities were structured around the theme and included team competitions, "Survivor" games, a scavenger hunt, and a movie night featuring *Castaway*. A new mentoring program was established, in which youths - boys and girls under the age of 21—were paired with older adults with CF who served as role models. The goal was to provide a "buddy" for young people to ask questions of, share concerns with, and learn coping skills from, as well as to emulate the mentor's compliance with the daily medical care regimen. Mentors would check in daily with their youths to make sure they were getting enough sleep, doing their treatments, and eating well. The mentor system allowed friendships to develop between younger and older people with CF and provided the young with a support system available to them throughout the year.

A newly structured leadership system was implemented in which a "leader of the day" was appointed to oversee activities and supervise Retreat. The Retreat leadership was also restructured into a committee system in which participants volunteered to assist with activities, fund-raising, mailings, obtaining donations, and recruitment. This group effort made Retreat happen with everyone working together, efficiently and productively. In the course of the event, we learned about the importance of collaboration, and the generosity of spirit that exists within a community that strives to improve the lives of those dealing with cystic fibrosis.

Each day was structured around social activities, support groups and educational workshops. Each support group had a particular focus, such as career planning, dating and relationships, exercise, grief/loss, teen issues, and gender-specific concerns. Social workers Isabel Stenzel-Byrnes and Joanne Asano, and



(Continued on page 9)

Retreat Survivor Team

**Teen and Adult Retreat** (continued from page 8)

Robert Rohde, a counselor to adolescents, facilitated the groups. A comfort level was reached quickly, providing an intimate atmosphere for expressing emotions.

Educational workshops were held daily as well. Dr. Meg DeLano held a question-and-answer session on cross-infection issues. Dr. Noreen Henig, CF adult physician from Stanford Hospital, spoke about sexuality, pregnancy, and fertility for persons with CF. Michael Chen, Pharm.D., and Anita Arguello, R.N., facilitated a workshop on the latest advancements in home intravenous therapy. Andrea Thronson conducted an excellent Yoga workshop. All talks were rated “excellent” by most Retreat attendees.

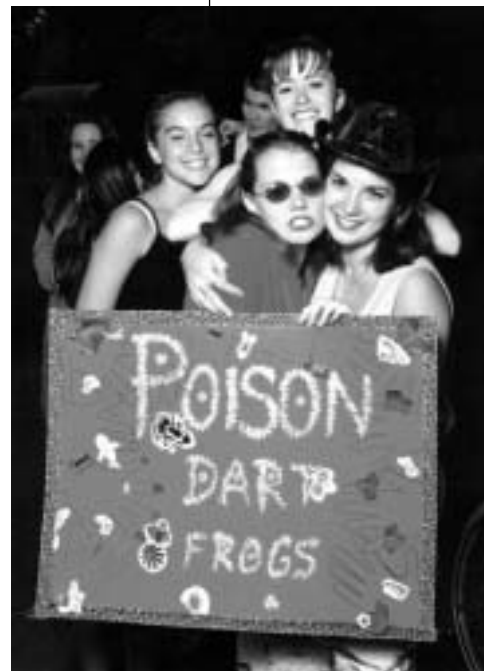
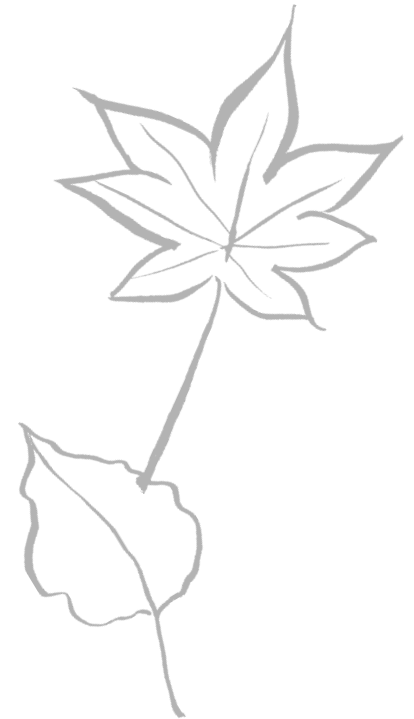
Social activities included arts and crafts, field games, Karaoke, a talent show, and a dance. Almost all the activities were held outdoors to reduce risk for cross-infection reasons. Field trips included a nature hike at Huddard Park in Redwood City, and swimming at Burgess Park in Menlo Park.

We received extremely positive feedback from many attendees. Everyone had an enjoyable time and stated that they wish to return next year. We all know that living with CF can sometimes be an isolating and psychologically difficult experience. Coming to an environment where everyone shares similar issues and fears creates an intimacy and bonding experience unlike any other. One participant stated, “Every year I leave learning more about myself, about how wonderful people at Retreat are, and with more knowledge about CF... Retreat gives me a renewed sense of life....” Another attendee said that Retreat was “the best experience in my CF life.” And another participant stated that the best thing about Retreat was “the feeling of belonging and acceptance, the people and the new friendships formed...”

The Annual CFRI Teen and Adult Retreat would not have been possible without the generous contributions of many donors. We are extremely grateful to the Strobel Family Trust, Lockheed employees, the Summer Youth Project, and the Peninsula Community Foundation for their generous grants that made this wonderful event possible. With the help of the Strobel Family Trust and Lockheed grants, we were able to cover a significant portion of the Retreat costs. The Summer Youth Project grant was awarded to CFRI Retreat by Northern California Grantmakers, an association of foundations and private grantmakers whose mission is “to promote the well-being of people and their communities.” With their help, we were able to purchase an array of enjoyable arts and crafts supplies and recreational equipment. The Peninsula Community Foundation, one of the fastest growing philanthropic foundations in the country, awarded Retreat a generous grant that provided scholarships to Peninsula residents who would otherwise have been unable to attend Retreat because of financial hardship.

In addition, we would like to extend our gratitude to Alex Jenkins for organizing a superb “Rock for Air” benefit concert to raise money for Retreat. Ed Kinney graciously donated to Retreat in honor of his friends who have passed away from CF in the past year. We would also like to express our gratitude to The Gap, Inc. and to Bob Thorpe for the beautiful t-shirts and silk-screening services. Last, we would like to thank Costco, Inc. and Safeway for their generous contribution of snacks and drinks so that Retreat participants were well nourished. A special thank-you to everyone on the Retreat committee and to all participants who worked hard to raise funds for this year’s Retreat.

Plans are already underway for next summer’s Retreat. The Retreat will be open to teens aged 15 and older. We welcome new faces and encourage teens and adults to call CFRI at (650) 404-9975 to get on the Retreat mailing list. Join us for the Annual CFRI Teen and Adult Retreat 2002 and come witness the magic that happens at Retreat! ■



Retreat Attendees

## 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? 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. 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 don't send us your only 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 attach on the back of your photo: 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 written permission!

We can't wait to see the beautiful photographs of your child! Send to CFRI, 2672 Bayshore Parkway, Suite 520, Mountain View, CA 94043 or [www.cfri.org](http://www.cfri.org). ■

2001 ANNUAL CONFERENCE

## Organ Donation – A Talk by Lisa Solinger

*By Lori Lindmeier*

Lisa Solinger works for the California Transplant Donor Network. Established in 1984, this is a nonprofit government-appointed agency. The agency is at work 24 hours a day, 7 days a week, helping families make the decision to donate their organs, and then allocating the organs to recipients who match. The reality is that there isn't a shortage of potential donors, but a shortage of organs donated; only 50% of the population sign up to be donors. This shows the tremendous need to educate the public on the benefits of organ donation.

The subject of organ donation is not one that many wish to discuss. People don't want to talk about dying, so discussing what happens to their organs after they die may seem morbid to them. The fact is that anything can happen to any of us at any given time. This creates the need to have these discussions in advance so that we can all be assured that our wishes, whatever they may be, will be carried out. It also eliminates the emotional burden of having family members decide what they think you would want.

Organ donation can only take place if the giver is brain dead. Though there is no blood supply to the brain, there is still blood going to the other organs. This can often add to the emotions of the grieving families, because to some degree the patient appears to be "alive." Ventilators inflate the lungs; heating blankets keep the body warm. Brain death is not the same as a coma. You will never wake up from brain death.

The criteria involved for organ donors change yearly, so it's important to make the decision to donate regardless of any health problems. Organs not good for transplant can be used instead for research.

United Network for Organ Sharing, or UNOS, is the governing body that oversees all organ procurement agencies, making certain there is fair and equitable distribution of organs. Often we hear about famous people getting a transplant and it may seem that they have gotten top priority. This is not the case at all. The transplant is widely reported because important events in the lives of well-known people make news. Each person listed has the same and equal chance at an organ transplant. UNOS handles all policies of donations, as well as the lists.

Organs currently being transplanted now are heart, lungs, liver, pancreas, kidney, and small bowel. Unfortunately, the number of people waiting for organ transplants has quadrupled in the last 13 years, rising from 16,026 in 1988 to 76,617 in 2001. Of those people, only 30% actually have an organ transplant operation. This number has doubled from 12,786 in 1988 to 22,854 in 2000. The numbers show that the transplants have not kept up with the number of those being listed for them. As it stands now, about half of the listed children and one third of the listed adults will die while waiting for a transplant. The average wait for a pair of lungs is 643 days. The average wait for a heart/lung block (often done for cystic fibrosis patients) averages 759 days; about 4,000 people are waiting for a single or double lung transplant today.

To become listed for transplant, patients must first be recommended by a physician. Following that, there are criteria the patients must meet. They will be evaluated for the surgery itself (are they strong enough to make it through?), anesthesia (will they tolerate it?), and available insurance (without it, the cost of



*(Continued on page 20)*

*Lisa Solinger*

# 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

## CF, Andy, and I

By Genevieve Broom

When I wake up, I sometimes curse the ever-present mucus in my lungs. Usually, once I'm awake, it's difficult to sleep again, as the lungs command me to cough to remove the mucus.

It could be at the same moment that I also think, Couldn't I, just for one morning, wake with a clear chest and breathe easily? In the normal scheme of things, this seems like a fairly simple basic request. But for someone with CF, it would be a gift from heaven! It's a rare moment within my busy life that I feel self-pity, and quickly I reprimand myself for feeling so negative.

Then I sit up to contemplate my day and what is planned. As I look out my window, I gaze across a well-loved garden, the paddocks with their beautiful green grass, and a very special friend who inhabits them. And then I forget my full and aching lungs, and I marvel how I came to own such an amazing animal, who over 12 wonderful years has given me strength, fun and laughter, and a will to live.

Pretty amazing gifts for an animal, isn't it? He can't speak, can't embrace me, but he has an unlimited supply of unconditional love, tolerance, patience, and forgiveness.

Poplars Farm Anniversary, more affectionately known as Andy, or the "Randster," came into our lives at the tender age of 3, a

*(Continued on page 12)*



Genevieve and Andy

Dear Readers,

Hey, what's up?

First of all I'd like to send my condolences to those that have been affected by the recent tragedies in New York, Washington, and Pennsylvania.



Katie Weber, Editor

I am so excited that we're finally getting *Aspirations* on its feet, but I really feel the need to stress that this paper is made by you for you, and YOU are the writer, the poet, the artist, the reporter, YOU are the one who can make the difference. I just help out. So please send in your birth dates, pictures, drawings, poems, articles, endorsements, questions, ideas, songs, recipes, anything, PLEASE! I'm open to anything and everything, and I hope to hear from you soon!

I would like to tell you a little bit about what I did this past summer. I recently attended the 14th Annual CFRI Conference, and I'm so pleased at the positive motivational support that I received from everyone there. I was excited because they had a teen panel this year and I didn't feel like a tagalong. The teen panel was basically for people to ask teens with CF questions on how they cope with school, friends, and family issues. The teen panel was made up of: Joseph G. Batchelder, age 13, who enjoys LOTS of physical activity; Daniel Hendricks, age 15, who enjoys BMX biking; Michelle Jones, age 14, is who also is very active, and was the model for the Vest (see last issue); Christina Miller, age 16, who enjoys spending time at the Retreat; Anna Modlin, age 20, a swim coach and very active with the Retreat; and last but not least, Joey Yerves, age 15, who enjoys art.

Please read my interview with Christina and Joey. It was really interesting. Well, that's about it, guys. Finally, I'm going to beg you again to PLEASE send in anything and everything that you possibly can think of, okay?

Love, Katie

[Cfriteeneditor@aol.com](mailto:Cfriteeneditor@aol.com)

# Dream Surfer Network

Hey readers, there's a great Web site out there for teens with CF. It's called the Dream Surfer Network.

Dream Surfer was started in 1997 by a group at John Hopkins Hospital. They were trying to create a Web site for teens from the Cystic Fibrosis Center. They wanted to make a place where teens could relate to other teens that knew what they were going through. It was launched in 1999, and is now in full operation.

The Dream Surfer Web site is for teens aged 12 to 18 and is fully protected and secure. You can download it from any computer. Those without a computer may be interviewed to get a computer completely free.

The Dream Surfer Network was started for teens with CF, but now is available to teens with heart problems, mobile defects, other lung diseases, and many other genetic diseases.

This is what some teens have to say about the Dream Surfer Network:

*"My compliments on the site!  
IT ROCKS!"*

*—Sally, 18-year-old Dream Surfer member*

*"I feel that the Dream Surfer Network is a great way to bring people together that share a common bond."*

*—Andy, 16-year-old Dream Surfer member*

*"I am thankful for the donation of my computer from The National Cristina Foundation and the Grant-A-Wish Foundation. I have been able to keep in touch and chat on line with other teenagers who have the same illness as myself and many other different ones as well. I use my computer in various ways. A few are: searching the Web site praying that a cure has been found for cystic fibrosis, playing games on the Dream Surfer site, and chatting and visiting the library on the Dream Surfer site to educate myself more about the life-threatening lung disease, cystic fibrosis, with which I'm living today."*

*—Allen, 14-year-old Dream Surfer member*

If you'd like to be a Dream Surfer member, you can contact Carey Wargo, the Dream Surfer Network program manager, for an application, or for more information. Call (800) 933-5470.

E-mail, [info@dreamsurfer.org](mailto:info@dreamsurfer.org).  
Demo site, [www.dreamsurfer.org](http://www.dreamsurfer.org). ■

CF, Andy, and I (continued from page 11)

black, gangly, and kind-looking part Welsh Galloway. Dad had gone to a horse sale to buy a float, but instead came home with a horse! I already owned a pony, but I was growing too big for her, and told Dad (confidently thinking, as children often do, that my request would be granted!) that I needed a bigger horse. I didn't expect this to happen for quite a while.

At the horse sale, some of my friends from Pony Club saw Andy, and told Dad he was the "perfect next horse" for me. Luckily, Dad heeded their advice, bid for Andy successfully, and brought him home. To this day, I marvel at the synchronistic events that brought Andy home to me. Andy and I have had a very successful career together and are known around the district as a winning combination.

Often a horse's temperament and conformation are suited to only one discipline—Dressage, One Day Eventing, or Showing. But not Andy—he is accomplished in all three. In the showing ring, we have placed in hundreds of ridden and led events, resulting in 30 Champion and 30 Reserve Champion ribbons. In the Eventing scene, in Pony Club (at Interzone and State Level), and now in Open Competition, Andy and I have won over 30 One Day Events, and 18 times have won the Dressage Phase. My favorite wins include 1st Place at the 1992 Pony Club State Dressage Championships, 1st Place in the Grade 2 Pony Club Interzone Eventing Championships, and 2nd Place at the Royal Horse Summer Show in 1995 for Best Pony Club Mount, out of over 100 entries.

The best part of the competitive horse world was meeting different people. Growing up in Pony Club was a wonderful social time, and Club members learned the skills of comradeship, support, and the grace of keeping a smile on your face when beaten, or being humble after a good day.

Competing also gave me some treasured memories for my parents and myself. At my 21st birthday, my mum gave a beautiful speech, and expressed great joy when seeing her supposedly "frail little girl" gallop around a cross-country course, arriving back safely at the finish line, flushed, out of breath, but very happy.

As I grew older, I looked forward to my competitions not only for riding, but also for the time I shared with my parents, particularly my mum, as my dad was often still milking cows when we needed to leave around dawn.

I was fortunate that my parents encouraged my horse riding. At that age, I guess they were unsure how long I would be around, so anything that I expressed interest in, they were happy for me to follow it, and encouraged it enthusiastically.

Horses can often be a passing fad for many children but my passion has stayed strong. Yet, over time, and as my health deteriorated, I lost some of my competitiveness and found myself grateful that I was able to ride and enjoy Andy's company and personality. And he certainly has a personality! Recently, he decided that the pumpkins next to his paddock (which he'd never shown an interest in before) looked very nice, and proceeded to eat the pumpkin vine, then a whole pumpkin, and left telltale teeth marks in another before Dad was on to him!

Several weeks later, Andy got in trouble again, when he ate a prized watermelon (only the second one Mum had ever grown) that was unfortunately too close to the fence.

Andy's antics over the years have given us many laughs, and it's surprising how intelligent those tricks can be, particularly if they have to do with food!

Andy turns 15 this year, as I turn 26. As the time rolls by, I become a bit daunted, as it becomes likely that I'll outlive him. I can't imagine my life without Andy, so I try

(Continued on page 14)

## I Have CF

By Patty Weber, age 10

I have CF,  
It's not fun.  
When it's over,  
I have won,  
And the world will know  
Who I am,  
And my family,  
Too,  
They will think,  
"How did she survive?"  
I have CF,  
It's not fun.  
When it's over,  
I have won.



## Pokes 'N Pricks

By Cindy Lynn Ray, age 13  
Durham, North Carolina

Pokes 'n pricks 'n pricks 'n pains  
Lots of hurts with zero gains  
Bloodtests, skintests, IV's too—  
Why just me and why not you?

Needles 'n knives 'n pokey things  
Things with cuts, things with stings  
All part of life when you're like me  
Which—trust me—you wouldn't  
wanna be!

## The Lake

By Katie Weber

The water's clear,  
And all is calm,

I hold a flower in my palm,  
The air is fresh from lily pads,  
Here is where  
I'm not so sad.

I used to come here  
With my true love,  
He's now in heaven, with a dove,

With a dove  
That talks to me,  
Fills my spirit,  
Sets it free.  
He's out there somewhere,  
Watching me.

He knows I love him,  
He knows I care,  
I cannot help but sit and stare

At the lake,  
As ripples form,  
That's why I'm here,  
So I can mourn,  
My heart . . .  
Is torn.

The water's clear,  
And all is calm,  
I hold a flower  
In my palm.



## My Interview with Teen Panel Members Joey and Christina

(Christina Miller, age 16, and Joey Yerves, age 15; August 11, 2001)

**Q# 1: What would you like to see in a youth newsletter for teens with CF, their siblings, and friends?**

**Joey:** "Discussions of how it is for kids in their everyday lives whether it be with friends or family. Updates on new equipment and medical breakthroughs that could lead to a cure."

**Christina:** "I would like to see people, instead of talking about cross-infection with other cystics, I would like to see them talk about how much we need each other in our lives, and how much we love spending time with each other."

**Q#2: Do you find it difficult to find ways to interact safely with other teens with CF?**

**Joey:** "Not really because in my mind I know that they go through the same problems I do, and I know that we can discuss a common experience in hospitals, with treatments, etc. . . With other people, you're scared to tell them because that gives them another reason to judge you on top of personal looks and the way you dress as well as the people you hang out with."

**Christina:** "No, we're pretty careful about cross-infection but we're also very laid back and we love spending time with each other. And if we're sick, we usually tell each other to stay away."

(Continued on page 14)



Christina Miller



Joey Yerves

not to think about the inevitable. Animals are lifelong friends, and have so many gifts to give to their owners. I like to think of myself as a guardian to Andy, to whom I owe responsibility and care.

What we CFers may lack at times in our health, we certainly make up for, with the talents we have been blessed with. Owning, riding, and competing with Andy has given me a chance to forget about CF and focus on something that gives me much joy, pride, and success. Andy and the experiences we have had together helped shape the person I have become today. Never doubt the healing power of our animal friends.

Check out Genevieve Broom's CF Web site at <http://www.angelfire.com/ok4/cfgen> ■

## Words of Wisdom

♦ *Find a penny, pick it up,  
and all day long you'll  
have one cent.*

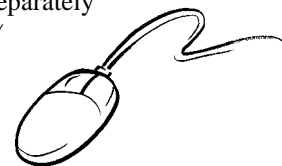
♦ *Good things come in  
small packages, and  
medium, and big ones.*



## Dear Readers,

This is a new thing that we're going to try. If you have any questions about school, family issues, friends, boyfriends, girlfriends, etc., I'd be glad to try to help you out. If it's a medical question, I'll contact a CF specialist, and he or she will answer for me. Now if your question is personal and you don't want it published in *Aspirations*, let me know so I don't slip it in there for all to see! If it's that personal, and you send me your e-mail address, I'll e-mail you privately, and no one has to know. Can't wait to hear from you!

By the way...you'll notice that *Aspirations* is being put right in the middle of the CFRI Newsletter again. This is so you can easily detach it without tearing any pages. That way, you can begin collecting your own newsletter to save separately if you want to. You can also ask to receive *CFRI Newsletter/Aspirations* by e-mail if you will send us your name and address, relationship to CF, date of birth (of the CF patient only) and e-mail address.



And again... this is your newsletter, for and about kids and teens with CF. Please send submissions of articles, poetry, photography, and artwork to Katie Weber, CFRI's teen editor, at [cfriteeneditor@aol.com](mailto:cfriteeneditor@aol.com), or mail to CFRI 2672 Bayshore Parkway, Suite 520, Mountain View, CA 94043. 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!

Web Sites: [www.Pogo.com](http://www.Pogo.com) [www.dreamsurfer.com](http://www.dreamsurfer.com) [www.snoopy.com](http://www.snoopy.com) (just for fun) ■

—Katie Weber  
Editor



Interview with Joey and Christina (continued from page 13)

**Q#3:** *Do you have any comments or words of wisdom for Aspirations readers?*

**Joey:** "Stay strong, come to camp, and be social with some people who are just like you and have some of the same experiences as you. You'll feel welcome the minute you step on the grounds of the camp."

**Christina:** "It's not always a bad thing, and people shouldn't get so freaked out about cross-infection. Be careful, but don't make yourself live in a bubble, because there's more to life than just germs. There's love and bonding."

I would like to personally thank Joey and Christina for their opinions and comments.

Thank you. —Katie Weber. ■

## End-of-Life Issues—A Talk by Walter M. Robinson, M.D., M.P.H.

By Ann Robinson

Dr. Robinson reminded us that a little planning goes a long way – especially in planning how we want to die. Why do we talk about death? Talking doesn't hasten death. Pain cannot be treated unless you talk about it. Studies show that pain is currently undertreated, or unnecessary because drugs are available to alleviate pain.

What are the end-of-life symptoms for people with cystic fibrosis? Some patients experience dyspnea (breathlessness) or a spasmodic cough, headache, chronic sinusitis, hypoxia (low oxygen levels in the body), or chest pain and hemoptysis (bleeding from the lungs). The treatments for these symptoms include morphine, medicines for local pain, and sedation. Patients must keep an open dialogue with the health care team to let them know of their pain and special needs.

A study of CF patients who died at Children's Hospital in Boston showed that their forced expiratory volume (FEV1) scores varied from 14% to 34%. 43% did not write DNR (do not resuscitate) orders until their final 5 days of life. It seems as though we didn't know they would die. DNR orders, living wills, and health care proxies have a unique role in CF and should be discussed in clinic when there is no crisis. Why should patients have blood drawn or take vitamins on their last day of life?

What is appropriate for people with CF? Should we use assisted ventilation (BiPap), which extended the dying process before transplantation was possible? Do we use BiPap until transplanted lungs may become available? During curative therapy, the health care team treats symptoms. During palliative therapy, the team manages symptoms. Death is scripted by the patient.

Where should patients be for end-of-life care? Wherever they want to be. Settings can be in the hospital as an inpatient, in a hospice, in the home, or in a rehabilitation setting. Who should be there? – The family, brothers and sisters? What is the contingency plan? Does the doctor have a script?

What are the best treatments? For dyspnea, we use morphine. For pain, we think locally and treat the location of the pain. For hemoptysis, we use sedation and dyspnea medications. When pain and suffering are emergencies, they should be handled as such.

Lung transplantation has changed end-of-life care for patients with CF. Now there are new antibiotic agents and a replacement model (lung transplantation). Can physicians simultaneously prepare patients for dying and transplantation? What is the role of the medical doctor? Are we cheerleaders or heroic rescuers? What is the role of the patient? Are you a survivor or a beater of odds? When should patients get on a transplant list? If people list too soon, it may shorten lives. If people list too late, many people may not survive.

What are the ethical issues of end-of-life care? The fear of addiction weighed against pain that is soul-destroying presents a dramatic dilemma. The widely held belief that morphine hastens death is unsubstantiated; there are no such findings in the medical literature. There is no consensus on physician-assisted suicide: Oregon is the only state where this is legal.

What are the fears and hopes of the patients? Cross-infection at the end of life is a very contentious issue. Dying alone for this reason is tragic. Think about negotiating a middle ground beforehand, so that family and friends can be present and supportive.

*Walter Robinson's rule – Talking about dying doesn't make it happen. ■*



Walter Robinson, M.D., M.P.H.



## Planned Giving

CFRI is profoundly grateful to those who have supported our children and adults during their lifetime. We deeply appreciate our kind friends who make our mission possible.

The future of CF research and educational programs is important to me!

Please send me information on how I can include CFRI in my will.

I have included CFRI in my will.

Please send me information on how to donate securities or other properties to CFRI.

Please have someone call me to discuss giving opportunities.

Enclosed is my donation of \$ \_\_\_\_\_

Name: \_\_\_\_\_

Address: \_\_\_\_\_

City: \_\_\_\_\_

State /Zip: \_\_\_\_\_

Best time to call: \_\_\_\_\_

E-mail Address: \_\_\_\_\_

*Clip and mail this form into an envelope addressed to:*

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

 \_\_\_\_\_



## Message from the President of the Board of Directors:

CFRI enjoyed a successful year in 2000, led by revenues of \$730,489. This generous support enabled CFRI to maintain a high level of funding during the year for both research and our ongoing educational and support services. (A comparison with the revenues of 1999 reveals a 9% decrease in 2000. This difference is attributable to exceptionally large one-time donations made in 1999; revenues in 2000 were considerably higher than in 1998, however, demonstrating that basic CFRI growth is proceeding at a satisfying pace.) Also because of our supporters' generosity, CFRI is able to put on its Annual Educational Conference and Adult/Teen Retreat, offering scholarships to those who might not otherwise be able to attend. Our Newsletter is sent out twice a year to educate and connect our families. CFRI has also been able to offer more consistent outreach and support. In addition to improving these services, we have also been able to establish funding for ongoing research fellowships to encourage postdoctoral researchers to focus on cystic fibrosis. CFRI awarded research grants in the amount of \$173,787. Research grants in 1999 totaled \$406,322—again marking that untypical year with a total of grant commitments that was exceptionally high for one calendar year, and especially so in view of the increased costs associated with relocating the CFRI offices early in the year and increased staff expenditures. As of 10/1/2001, research grants this year were just under \$300,000.

Overhead expenses and fundraising costs were \$252,479 for the year. Residing as we do in an area that has enjoyed unprecedented economic growth, we must compete for paid help and volunteers in an extremely difficult market. Of particular note was our ability to find an excellent new home for CFRI. Our new offices are near Highway 101 and San Antonio Road in Mountain View (about 1 mile from the old office).

Net assets stood at \$233,989 at year-end. CFRI remains deeply indebted to its generous donors, its cadre of untiring volunteers, and its dedicated staff. CFRI's complete Annual Report for 2000, including the audited financial statements, is available upon request. Please contact the CFRI office.

Sincerely,

Mike Roanhaus, President of the Board

### Some Books for the Journey (continued from page 3)

Andy is now 28 years old, and in December, he'll be running with the Olympic torch. He has a girlfriend and a full-time job, and he spreads his inspiring story wherever he goes. He's even had the help of some famous CF advocates – *Sports Illustrated* writer Frank Deford; former All-Pro quarterback and Monday Night Football announcer, Boomer Esiason; and All-Star third baseman of the Atlanta Braves, Chipper Jones. All of them have written a foreword for Andy's book.

Andy isn't satisfied with just living with CF. Andy's goal is to beat it. And though Andy's story is inspirational, doctors say "beating CF" cannot be done. But Andy Lipman refuses to listen. After all, he wasn't even supposed to be alive at 25! ■

# 2001 Conference Photos



All photos  
courtesy  
of Craig Burleigh



## Second Generation of TOBI Delivery Systems – Pete Challoner, Ph.D.

By Pam Heilman

Pete Challoner, the VP and Director of Inhalation Pharmaceuticals and Technology of the Chiron Corp., was the final presenter on Sunday afternoon, August 12<sup>th</sup>. Mr. Challoner spoke on the “Second Generation of TOBI Delivery Systems,” reporting that there are many products in the works that will make life with CF less stressful.

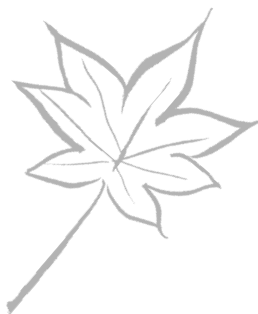
TOBI has been of great benefit to the CF community. Study after study has proven that TOBI can maintain lung function and is relatively safe. Many of the problems associated with TOBI have centered on the delivery system and not the actual drug. TOBI is used with an air jet nebulizer. Air jet nebs are continuous flow devices, which means that most of the medicine is sprayed into the environment. The best nebulizer, the Pari LC Plus, gets only 15% of the medicine to the lungs. This means that large amounts of expensive medicine are wasted. There is the added risk of resistant bacteria developing in the patient’s environment. A docimetric jet nebulizer is soon to be available. These nebulizers actually anticipate a patient’s breathing and only release medicine when the patient inhales. This would also reduce the dose variability that occurs with a regular jet nebulizer. Right now, Pari does have a manual interrupter that can be used with their LC Plus nebulizers. However, the interrupter relies on the patient to press a button while exhaling to prevent the constant flow.



Pete Challoner, Ph.D.

Two companies are currently working on a docimetric, hand-held, battery-operated aerosolized nebulizer. The companies are Pari and Aerogen. Aerogen is the company that Chiron has contracted with to work on the Aerodose, which is a nebulizer especially for TOBI. Two trials have been done on the Aerodose, and the conclusions were positive. The patient received a similar amount of medicine with Aerodose when compared with the Pari LC, although the LC was still slightly superior. This means that because the Aerodose sprays

faster, more medicine would be needed for the patient to get the exact same dose as with the Pari neb. The time for a regular TOBI treatment using the Pari LC is around 20.4 minutes while the Aerodose would be only 7.4 minutes. The study concluded that the drug was being deposited and distributed into the lungs in exactly the same fashion as the Pari LC. Chiron believes their Aerodose will be efficient and decrease delivery time. Mr. Challoner warned, however, that getting FDA approval would take some time.



What can we look forward to in the third generation of aerosolized technologies? The future is a dry powder aerosol technology. It will be a simple, inexpensive, disposable device. The powder will allow for better storage stability (no refrigeration). Best of all, the amount of active compound will be much higher. The powder will be inhaled in one big puff, which would take seconds for a treatment. While this technology is eagerly anticipated, it will be quite a few years before it will be available. The technical aspects of powdered TOBI are complicated, and much more work will need to be done.

In the meantime, the old-fashioned jet nebulizers are available, and Pari has many products that can help to address some of the problems associated with the constant-flow device. The interrupter, as mentioned, can help a patient get a much higher dose of TOBI; however, it does add time to the treatment. Pari also has filters, which will help to eliminate the environmental exposure to drugs. On a related note, in an abstract released recently by Dr. David Gellar, the Pari ProNeb Ultra Compressor and the PulmoAide Compressor were found identical in effectiveness. They are both considered the best compressors to use with the Pari LC Plus nebulizer while administering TOBI. ■

not reproducible. Ambry Genetics has successfully addressed these issues.

Using patent-pending innovations, our geneticists have developed the most sensitive genetic testing service available for use in a clinical setting.

By scanning the entire length of a gene, virtually every genetic anomaly leading to disease will be detected. This is especially important when patients are medium or high risk—where there’s been a history of disease in the family, for instance; or where you need to find whether a spouse is a carrier, or to confirm a diagnosis of CF, as in the case of the young girl I mentioned.

The Ambry Test™:CF is also ideal for patients where other tests have failed to detect one or more disease-causing alleles. Moreover, the Ambry Test™ is able to identify mutations regardless of ethnic background. Cystic fibrosis occurs most commonly in Northern and Eastern Europeans of Caucasian ancestry, which means that the mutations that other tests look for are the mutations that appear most often in Caucasians. The Ambry Test™: CF does not have that bias. It simply looks for *any* mutation that occurs in the critical gene, regardless of ethnicity.

Because laboratories are becoming so much better at identifying people who carry a CF gene mutation, in October the American College of Obstetrics and Gynecology recommended that all expecting couples be provided with information about carrier-status testing for cystic fibrosis. I believe this is an important moment for possibly reducing the incidence of the disease in this country.

The Ambry Test™:CF was validated using “blind” samples provided by the College of American Pathologists. The test correctly identified the mutation in each sample. In other samples, the test found the second, previously unidentified allele in persons who were known to have CF but whose second mutation had not been identified by other tests.

Tests conducted at Ambry Genetics are performed within the guidelines of the American College of Medical Genetics and are approved under the Clinical Laboratory Improvement Amendments Act of 1988 (CLIA).

For more information, clinics may contact Steve Swanson at Ambry: (949) 642-7202 or e-mail [sswanson@ambrygeneticscorp.com](mailto:sswanson@ambrygeneticscorp.com). Steve will arrange to send the Ambry Sample Submission Kit. Ambry’s Customer Service department at (866) 262-7943 will also send a Sample Submission Kit.

The Ambry Sample Submission Kit is easy to use, free of cost (we pay shipping).

Name: \_\_\_\_\_

Address: \_\_\_\_\_

City/State Zip: \_\_\_\_\_

Best time to call: \_\_\_\_\_

Email Address: \_\_\_\_\_

(Currently, we are not accepting samples from Rhode Island, Pennsylvania, Florida, Maryland, and New York, but will be shortly.)

With our ability to accurately and reliably sequence human DNA, Ambry can develop tests for mutations on any human gene. We are poised to introduce tests on other genes where mutations cause disease. The ultimate goal with this sort of genetic analysis is helping doctors help their patients to manage risk factors and anticipate problems before symptoms arise. ■

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## Catch All The Sites: A New Source for CF Web Sites

Have you been struggling to find information on the Internet about Cystic Fibrosis? We were, until CFRI and Digestive Care, Inc., makers of PANCRECARB®, the only bicarbonate-buffered, delayed-release enzyme, came up with the solution. **Catch All The Sites** is a comprehensive resource booklet listing Web sites related to CF. The guide is divided by topics for quick reference. Research, Support, Nutrition, and Financial Information are just a few topics with many informative Web sites listed.

To receive your free guide, please contact CFRI at [www.CFRI.org](http://www.CFRI.org) or 650-404-9975, or Digestive Care, Inc., at [www.digestivecare.com](http://www.digestivecare.com).



### Organ Donation (Continued from page 10)

transplantation is beyond the reach of most people). Can the patients surely handle the psychological aspect of organ replacement—for example, of breathing with another person's lungs? Have they a support system to follow them through recovery and beyond? Additionally, compliance is an important issue: it's important to be sure the candidates for a transplant will abide by all that is necessary to keep them as healthy as possible and ensure the best recovery. Once every criterion is met, the patients are then listed and they begin their wait.

There would be no shortage of organs if everyone made the decision to donate. Make a point of discussing this subject with your loved ones. Share Your Life...Share Your Decision. ■

## Endocrine Disorders in Adults with CF— A Talk by Dana S. Hardin, M.D.

By Ann Robinson

Osteoporosis, osteopenia, and CF-related diabetes are endocrine disorders found in adults with cystic fibrosis. If untreated, these three disorders can severely complicate the severity and well-being of CF patients.

Osteoporosis is defined as bone mineral density (BMD) that is more than 2.5 times below average for people of the same age and gender. Osteoporosis is associated with increased risk of fracture. CF patients experience an increased risk of fracture in the vertebrae and ribs due to increased coughing.

Osteopenia, a prelude to osteoporosis, also causes an increased fracture risk. There is poor nutrition, malabsorption of calcium and vitamin D, and delayed and reduced production of sex steroids in the body. Both osteopenia and osteoporosis are diagnosed in the clinic with conventional x-rays.

**Why do people with CF need to have good nutrition?** Osteoporosis and osteopenia are easier to prevent with good nutrition than to treat as fully developed clinical diseases. Calcium requirements for those with CF include: children 4-8 years old, 800 mg/day; children 9-18, 1500 mg/day; adults, 1200 mg/day. **Just because you have CF doesn't mean you have to have osteoporosis.**

CF-related diabetes is an abnormal glucose intolerance in people with cystic fibrosis. The number of patients with diabetes is higher than in any other age-matched disease group. Although diabetes in CF patients shares some clinical features with both type 1 and type 2 diabetes, it is a unique disease and is called cystic fibrosis-related diabetes (CFRD).

Of all CF patients over the age of 18, 75% have impaired glucose tolerance (a grossly underreported percentage, according to Dr. Hardin); 15% of these patients have indisputable diabetes. Insulin sensitivity can be impaired by the use of steroids such as prednisone. When taking steroids, care should be taken to check glucose levels. Hospital inpatients often have impaired insulin during the time they are sick.

How is CFRD diagnosed? Clinical tests can verify the disease. Physicians use the hemoglobin A1c test (a score of less than 6.5 is normal), fasting blood glucose (less than 126 is normal), postprandial (two hours after eating a large meal) blood glucose (less than 150 is normal), and oral glucose tolerance (below 150 is normal).

What are the symptoms of CFRD? Symptoms include fatigue, extreme thirst, excessive urination, unexplained weight loss, and difficulty maintaining weight. If you have any of these symptoms, please discuss them with your health care team. If CFRD is diagnosed, your clinic team will include an endocrinologist who is an MD, and a nurse educator and a dietician, both of whom have been trained in diabetic care.

**Having diabetes is not a death sentence.** Blood glucose can be monitored, treated, and controlled at home. The benefits of insulin therapy include weight gain, improved pulmonary function, and greater lean body mass. CF patients who control their blood glucose are able to live full, productive lives. ■

# In honor of

(From June 2001 to November 3, 2001)

**"In honor of" lists names of living persons in whose honor a donation has been made.** Do you have a relative or friend whom you would like to celebrate/salute? Are you searching for the perfect birthday present, wedding or anniversary gift, graduation gift, etc.? Give a gift with lasting impact – make a donation to **Cystic Fibrosis Research, Inc.** At your request we will send a special message to the recipient informing him/her of your contribution. 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 the occasion for the person you are honoring to: CFRI, Bayside Business Park, 2672 Bayshore Parkway, Suite 520, Mountain View, CA 94043

Chelsa About	Caroline Daly	Milly Held	Michael Livingston	Kaeti Pierce	Larissa Swanson
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Cameron Cornell	Alyssa Harvey	Stan Lenox	Erin Phillips	Ann Swanson	
Kelly Couch	Tyler Heavner	Barbara Lenssen	Jereme Pierce	Christina Swanson	

# Memorials

(From June 2001 to November 3, 2001)

**A memorial is a donation made in honor of a deceased person.** The memorial gifts 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 that occasionally someone who died had the same name as a living person.) Send name of the deceased, the name, address, city/state and zip of the relative/spouse, etc., whom you would like notified that the donation was made. Send to: CFRI, Bayside Business Park, 2672 Bayshore Parkway, Suite 520, Mountain View, CA 94043

Erica Acm	Kyle Butler	Lillian Hall	Robert Mueller	Michelle Schaeffer	Susan Thibault
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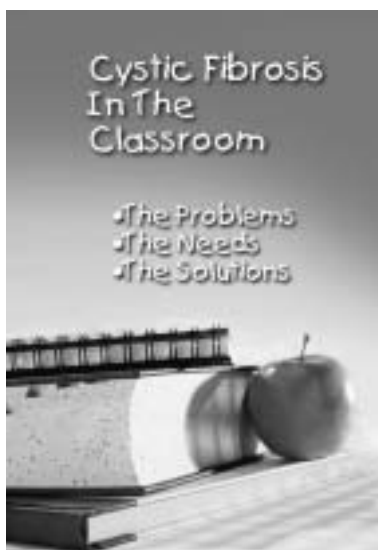
# Cystic Fibrosis In The Classroom

The “premier” guide for those in the school system.

Do you know someone in school who has cystic fibrosis? The “premier” guide, 20 pages highlighting important issues related to children with CF in school, has finally been written. *Cystic Fibrosis in the Classroom* has been issued by CFRI and Digestive Care, Inc., makers of PANCRECARB®, (pancrelipase) the only bicarbonate-buffered delayed-release enzyme.

*Cystic Fibrosis in the Classroom* covers topics pertinent to a CF patient in a school setting. Nutrition, exercise, symptoms, medication, and treatment are all topics addressed in this guide. There are many issues related to CF of which the school faculty may be unaware. This book is for them. In clear and concise terms it describes CF and how it may affect the classroom environment.

Forget about the old, dated brochures you’ve seen. This is the most comprehensive guide to date. For your free copy, please contact CFRI at [www.cfri.org](http://www.cfri.org) or (650) 404-9975 or Digestive Care, Inc. at [www.digestivecare.com](http://www.digestivecare.com).



2001 ANNUAL CONFERENCE

## Newborn Screening In California – Martin Kharazzi, Ph.D.

By Pam Heilman

Dr. Martin Kharrazi started the conference on Friday night with his presentation on the proposed Newborn Screening Program in California. Earlier, at the Doubletree in San Jose, there had been a day-long conference discussing the challenges California must overcome in order to have an equitable and beneficial cystic fibrosis newborn screening program. There are other states with successful programs, such as Colorado and Wisconsin, and California could use them as a model. For instance, in Wisconsin, a blood spot is taken from all newborn babies and tested for IRT (immunoreactive tryptophan). IRT is a chemical released from the pancreas. High levels of IRT would mean that a baby was at risk for CF, and therefore genetic testing would be warranted. This seems simple enough, but with the high number of births in California, establishing the right level of IRT is problematic. California would need to have a cutoff for IRT that would not overburden the system with expensive genetic testing, which poses its own problems. Unfortunately, California’s diverse population makes screening more complicated. At this time the least expensive and most widely used DNA testing has a 90% detection rate for Whites, a 56% detection rate for Hispanics, and a 75% rate for Blacks. With this kind of disparity, minorities with CF could suffer a high false-negative rate, which could prevent them from getting proper care.



Martin Kharazzi, Ph.D.

The benefits of a newborn screening program appear to be innumerable.

Evidence supports the idea that children who are diagnosed early with CF do tend to maintain health longer than those who are diagnosed only after suffering some debilitating symptoms. Often, there is a monetary, emotional, and of course physical cost when there is a long medical “odyssey” before diagnosis, says Dr. Kharazzi. Yet a doctor, who was a conference attendee from Massachusetts, stood up at the question portion of the presentation and made some points about the possible costs of having a newborn screening program at all. He referred to the fact that the programs primarily screen out carriers, and this can place heavy burdens when it comes to counseling and resources. Dr. Kharrazi did speculate that you would find thousands of carriers and maybe 100 actual cases of CF in the newborn babies in any given year. The Massachusetts doctor claimed that even in his state, where there are fewer Hispanics than in California, many minority patients were misdiagnosed because of the screening. Their mutation was simply unknown; but practitioners believe that, because they were screened at birth, there was no chance of these children having CF; therefore a lot of them fell through some dangerous cracks.


Despite the controversy, the program is being planned and is moving forward: Dr. Kharrazi is working as the Chief of the Program Research and Demonstration Unit. He is hopeful that California will be able to overcome the obstacles and have a program in place by 2003. Currently he is conducting a study that will produce a list of CF mutations, which will achieve a 90% detection rate for the three main population racial groups: Hispanics, Whites, and Blacks. This is a huge undertaking that involves two phases. Phase A: create a statewide registry of patients using 12 of the 18 CF clinics in the state as well as other resources. There must also be lab standardization for the IRT sampling as well as a consensus on the best cutoff number for high IRT.

Phase B: identify the types and frequencies of CFTR mutations in the Hispanic CF cases. Dr. Kharrazi maintains that California’s Newborn CF Screening Program will not go forward unless the state can identify 90% of Hispanic, Black, and White CF mutations. ■

## America Stands

As I sit in my corner who could image  
 Our famous twin towers up in flames  
 Now, as I open my doors fear sets in  
 Is the worst yet to come?  
 How could a division of individuals  
 Use their time for destruction?  
 Utter nonsense  
 Yes America's heartland is hurting  
 With fear stamped across our land  
 We wait and watch what will happen next  
 We feel uncomfortable in our home  
 Must we live in fear?  
 Can we collectively rise above this hate?  
 No more, no more  
 Victory will occur when America Stands

## Our Statue of Liberty



Our Magnificent Lady draped in the garment of equal rights  
 And a chance at the American dream.  
 Her arm extended with the torch represents the Flames of Victory.  
 In the air as you draw near you hear her soft voice echoing in the wind.  
 Strength, and equality for all.  
 We must stand together to make a difference.  
 Life, Liberty and Justice.  
 Approaching her makes you feel proud to be an American.

We at CFRI would like to give our heartfelt thanks and appreciation for our friends and families who have given us their dedication and generous support.

—Calvinia Williams, Executive Director

## 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 I like about your Newsletter: \_\_\_\_\_

Improvements I would like to see in your Newsletter: \_\_\_\_\_

Below I have listed my ideas for future articles, regular columns, or subjects I'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 that 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
- Memorial and In Honor Of gifts
- 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](mailto: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](http://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 15 to adult
- Family support groups
- CF support groups for ages 16 to adult
- *CFRI News* and *Aspirations* newsletters
- E-mail 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, two Administrative Assistants
- 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

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

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## *Fall/Winter 2001*

### ***In This Issue***

- Your CFRI Dollars at Work! *Page 1*
- Some Books for the Journey *Page 5*
- Ambry's New Gene Test *Page 7*
- New! CFRI Youth Newsletter—*Aspirations* *Page 11-14*