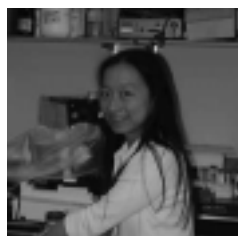


## A Possible Alternative Epithelial Chloride Conductance

By Peiyong Fong, Ph.D.,

Postgraduate Research Scientist in the Department of Physiology at Johns Hopkins University School of Medicine in Baltimore, Maryland

**Editor's Note:** Dr. Fong was awarded a research grant from CFRI in May, 1999 when she was at the University of California at San Diego. Her work is titled, "Functional Characterization of Ion Transport in a Model Epithelium Lacking the Cystic Fibrosis Transmembrane Conductance Regulator."



Dr. Peiyong Fong

As we have come to appreciate, Cystic Fibrosis Transmembrane Conductance Regulator, (CFTR) behaves as a chloride channel important in the directional transport—absorption and secretion—of salt and water. When CFTR activity is lacking, the disruption of transport leads to the symptoms of CF-related disease. The discovery of other chloride channels in the

same cells that normally show CFTR-mediated transport raises the possibility that these other channels can be recruited to compensate for the lack of functional CFTR in CF patients. However, transport is a complicated process that hinges not only on having the key players present and at peak performance, but also on *orchestration*. So a critical evaluation of the potential of alternative channels requires a consideration of not only how they behave *solo*, but also how they normally interact with the other players.

Let's set the stage. First, what does the stage look like, and what's being performed? Directional, or *vectorial*, transport is the distinguishing characteristic of all epithelial tissue. With few exceptions, epithelia have a side that faces the blood supply, at the inside of the body, and another that faces a compartment that is contiguous with the outside world. These sides are called "basolateral" and "apical" respectively. The cells that form epithelia also display sidedness, or *polarity*. This means that epithelial cells have different protein and lipid composition at the membranes at their apical and basolateral poles and consequently can move water and solutes, such as salt, in one direction or the other. Finally, on yet another level of organization, epithelial cells are bundled together snugly so that all their apical sides are facing one direction, and their basolateral ones in the other direction. The specialized structures that join the cells in this fashion are called *tight junctions*. These structures impart epithelia with effective barrier properties, and give a further degree of efficiency to

directional transport as well. So, for instance, digestive juices secreted by the pancreas do not seep back into the tissue, and nutrients absorbed by the cells lining the small intestine are efficiently taken into the bloodstream.

The work of many research groups also point to a role for CFTR as a regulator of other ion channels, including chloride channels, that underlie transport. So CFTR could be the violin soloist who also conducts the orchestra! However, as exciting as the possibility that alternative chloride channels may compensate for CFTR sounds (no pun intended), researchers now face the challenge of unraveling the normal roles of these alternative channels. It could be a violin, but is it a first violin? What is the score that player has in front of it? How to switch it with the soloist's score? Will the final performance sound melodious, or will it grate the ear? Moreover, because CFTR potentially regulates coexisting channels, the distinct possibility exists that these channels may be recalcitrant to activation in the absence of functional CFTR. All of these levels of tissue complexity lead to the obvious conclusion that these factors must be accounted for in studies of alternative chloride channels.

**RESEARCH**  
Your CFRI Dollars at Work

Of the candidate chloride channels that have been cloned, those of the CLC family are arguably among the best characterized. CLC channels are a large family of chloride channels that are structurally

unrelated to the ATP Binding Cassette (ABC) transporter family to which CFTR belongs. One well-studied CLC channel is CIC-2. CIC-2 has a broad distribution and can be found also in epithelial cells. On the biophysical level, the factors that determine CIC-2 channel opening and closing have been probed successfully, and its preference for chloride over other ions established. Its pharmacology, as well as its modulation by intracellular activators, recently has become a focus for several groups. Furthermore, CIC-2 levels are higher in the intestines of asymptomatic CFTR knockout mice.

All told, we have a better understanding of how CIC-2 performs as a soloist than we do for many other alternative channels. However, the role of CIC-2 in normal epithelial

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## Message from the Executive Director

25 Years of Research and Education! Not many agencies can say that! CFRI has an incredible foundation of researchers and volunteers who have dedicated themselves to our cause. We have so much to be proud of! Still, in my line of work it can be easy to forget how far we've come and get bogged down in daily worries. Recently, I have been worrying about our soaring rent costs, research funding costs versus support programs' costs, automating our bookkeeping procedures, soliciting new grants and donors, and recruiting volunteers when most people in this area already work long hours in the workplace. These are the kinds of problems that cause most people to lose sight of accomplishments and run from nonprofits. For me, I love the challenge. Recently though, it all felt very overwhelming. At the lunch hour, I took a long walk to clear my head.

When I returned to the office, a woman I had never met was waiting for me. She introduced herself as the grandmother of a CF child. She was clearly troubled about her situation. We went into my office for privacy. She told me her story. As tears ran down this grandmother's face, my own eyes became moist. I told myself, "Don't start crying. You have to help this grandmother in her time of pain." The details of her story are not important here. What is important is that she needed support and guidance and CFRI was there to help her. Don't get me wrong. We could not fix the troubles she was facing, but we could listen, empathize, and offer advice where appropriate. Mostly she needed to vent in a place where she felt understood. She needed someone to let her know that it was hard to be a grandparent of a CF child, to want to help but not always be welcomed to help, to be aware of the progression of the disease, yet have little control over any intervention. And she was worried about her adult child, the parent of her grandchild, who carries the weight of this diagnosis.

After our conversation, it became clear why I feel a personal bond with CFRI's families. I continue to fight for a cure because I know the importance of living a good life, of smelling flowers, feeling the wind, the joys of family, watching kids grow up, appreciating a good laugh, and wondering what we'll look like in old age. These simple things are essential. I was so touched by the courage this woman possessed to reach out for help on behalf of her family. Despite difficult odds, she was determined to persevere. She gave me tremendous hope.



CFRI's staff at a Staff Appreciation Luncheon given by volunteer Debbie Gerow (whose son Mark Ellis has CF), from left to right: Taylor Bui, Program Services Manager; Calvinia Williams, Executive Director; and Carolyn Easter, Administrative Assistant.

Soon after, another family visited the office. They were from a country on the other side of the world. Their child, age 18, had just been diagnosed with CF. The doctors in their homeland had never seen an 18-year-old with CF. They arrived hurting, but left two hours later with newsletters, fliers, lists of books to buy and take home, and email and website addresses to help get them the long-term support they needed. It was so impressive to see this family determined to get their child on the best possible course toward survival. They thanked us profusely. But once again I wondered if perhaps we should be thanking them. They reminded us of CFRI's underlying purpose: to provide hope and

(Continued on page 10)

# GOOD NEWS

*For People with CF and their Families*

## Heard of Book Clubs? Now There's "Birthday Clubs"

Our donors never seem to run out of fundraising ideas. Recently, a donation of \$365 came our way from June Miller of Yuba City, California. June, along with ten of her friends, gathered to celebrate the birthdays of Gail Montna and Patty Jaeger. In lieu of gifts, both women chose to send a donation to their favorite charity—CFRI—in honor of Brett Bennett of Palm Desert, California, June's 26-year-old nephew who has CF. The women gather regularly either at a local restaurant or at someone's house. This last event was a potluck occasion held at the home of Patty Morrison in Yuba City, CA.



*Brett Bennett, right, with his nephew, Brandyn Bennett.*



*Front row, left to right: June Miller, Patty Morrison, Patricia Rue, Gail Montna—the original birthday person. Back row: Marge Hill, Ginne Rosson and Virginia Belza. Not pictured: Michelle Montna, Susan Eager, Nicole Van Vleck, June Brookin, Karen Jelavich*

## Solvay Pharmaceuticals, Inc. Awards Scholarships

Twenty students from around the US who have cystic fibrosis (CF) were awarded a CREON® Minimicrospheres® Family Scholarship toward their college educations. Students will receive \$2,000 per year for up to four years or until completion of their current degree. Scholarships are awarded to students based on academic achievement, financial need, leadership qualities, and ability to serve as role models to others who have CF. All high school, vocational school and college students who have CF are eligible.

Each year from March 1 through July 15, applications are made available at CF treatment centers. Solvay established the scholarship program in 1993 as part of its "Partners In Caring™" program, and has awarded more than \$1 million in scholarship money since the program began.

*(Continued on page 7)*

## Study Participants Wanted

The International Association of Cystic Fibrosis Adults (IACFA) is seeking adults who were diagnosed with CF at age 18 or older to participate in a confidential study exploring their experience, educational needs, and sources of support at and following their diagnoses. Volunteers will be mailed a fixed-choice and short answer questionnaire, which should take less than 20 minutes to complete. A pre-addressed, stamped return envelope will be provided. We will use the survey results to guide the development of educational materials for those diagnosed in adulthood, and for their physicians and families, and to better understand how CF education impacts adults' outlooks during and following diagnosis. If you were diagnosed with CF at age 18 or older and are willing to participate in this important study, please contact either of us:

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## Attention Packard Children's and Stanford Hospital CF Center Patients!

Despite the nurses' strike, American BioSystems, Inc (ABI) continues to sponsor an incredible opportunity for patients in the CF Centers at Packard and Stanford Hospitals to try out the Vest®. *Regardless of insurance or financial status*, if you try the Vest and it works for you, ABI will make it available to you. Call Chandra at the CF Clinic at 650-497-8841 immediately to set up an appointment. The hospital trial ends 9-15-00.

# Cisapride (Propulsid®) Pulled from the Market

Dr. Dorsey Bass, pediatric GI specialist, spoke at Lucile Salter Packard Children's Hospital at Stanford Medical Center on May 25<sup>th</sup>, 2000 at CFRI's semi-annual General Meeting on the topic of "What's Happening with Cisapride?" Cisapride was pulled from the market by Janssen Pharmaceutica on July 14<sup>th</sup>, 2000 after being available in the U.S. for seven years. Cisapride is prescribed by doctors to treat Gastroesophageal reflux disease (GERD) as well as bowel motility problems. (Many people refer to GERD simply as reflux or severe heartburn.) GERD is the splashing back or refluxing of acidic stomach contents into the esophagus, and is fairly common in people with cystic fibrosis. Cisapride helps your stomach empty more quickly and tightens up the upper esophageal sphincter, thus reducing or eliminating the painful effects of reflux. It can also be very useful for constipation.

Dr. Bass said that over the years, quite a few GI patients at Stanford have been placed on Cisapride because the physicians there thought that Cisapride had a very safe drug profile with few side effects. But after five years of use in the U.S., Janssen Pharmaceutica noted that a cardiac toxicity could develop in a person taking Cisapride. For the most part, this seemed to involve people who were taking other drugs that also increased heart rate (eg. Intraconazole, erythromycin) and therefore in combination with Cisapride became toxic. It was also noted to be unsafe for people who already had cardiac abnormalities. When used improperly, people were noted to have very fast and irregular heartbeats that could lead to death. To date, ninety deaths have occurred. To put this in perspective, over three million prescriptions have been filled in the U.S. alone. 85% of the deaths have occurred in people who had reason NOT to be taking the drug.

Janssen Pharmaceutica for the last two years has been sending out warnings to physicians and clinics in an effort to eliminate or reduce the death rate. No such decline was observed. The FDA continued to be concerned about these deaths. Since Janssen Pharmaceutica determined that further studies into the viability of the drug were not financially feasible, they believed its only option was to pull the drug off the market.

For his patients who are taking the drug, Dr. Bass has recommended coming off the

(Continued on page 5)

# Compliance with CF Treatments: A Daily Battle or a Way of Life?

By Heather N. Lonn

***Editor's note:** Heather Lonn is a 22-year-old with Cystic Fibrosis interested in exploring hot topics for CFRI News. The opinions expressed are those of the author and the individuals she interviewed. CFRI thanks all who contributed to this piece, especially Heather, for sharing their thoughts on a difficult subject.*

For the first twenty years of my life I was an extremely healthy individual who happened to have CF. I didn't take any medication or enzymes. In fact, I was a water aerobics instructor for years. It wasn't necessary for me to do any type of treatments for my CF. Two years ago my whole world flipped upside down. I've been in and out of the hospital five times, had a number of different surgeries, and several bouts with home I.V. therapy.

I am supposed to do my I.P.V. machine (Intrapulmonary Percussive Ventilator), an air compressor that forces air into the lungs to clear mucus twice a day. I'll be honest. I don't do my treatments at all. It's a daily battle between my parents and myself. They don't understand why I refuse to do them. In my case the reason is simple. For twenty years I hardly even remembered I had CF. I went through the normal hardships of the average teenager. I've lived in the same place for 18 years and I've never met another person who has CF or does treatments. I don't like to think that I'm different from other people. And anytime I'm supposed to do a treatment, I feel different.

So, why are some CF patients more compliant than others? Is it because they've been doing treatments all of their lives so it's all they know? I interviewed several people in an effort to understand this uncomfortable issue. My friend Joe lives back East and has had a fairly difficult time with CF. Recently he had a successful lung transplant. When I questioned him about compliance, he said that one of his parents always made him do his treatments. Also, he usually wound up in the hospital when he didn't do his treatments. When asked if the way a doctor talks with him affects his compliance, Joe said, "absolutely it matters." He feels that the doctor should be someone that the patient trusts and can talk to. For Joe, the three main reasons for doing treatments are: "Because the doctor tells you to; because parents insist CF kids do treatments; and because everyone (who has CF) has to do treatments."

It's logical for people with CF to comply with doing treatments in order to increase their chances of staying healthy. But why is it that some still refuse to do treatments? Kevin was diagnosed at age 21 and is not very compliant. He says, "It's not my doctor's fault at all." His doctor has stressed the importance of treatments, but he says "I'm so busy that the time involved for this type of therapy is so limited that I fail to do as I'm asked." Kevin's case of CF isn't extreme right now. Because he doesn't feel ill, it makes it all the harder for him to comply. When asked about the motivation for compliance, Kevin is unequivocal. "I really think the answer to that question lies in the CF individual. I also believe it's a matter of the severity of the disease that has a positive correlation with compliance. When I was sick, of course I did everything that I was told. But once I got back on my feet again, I didn't follow everything that I was supposed to."

Devin, a nine-year-old boy with CF tells what makes him want to do his treatments. "I do them because I HATE to get sick and go to the hospital. I hate I.V.s *ALOT*. Whatever the doctors say, I try to do it. But if it's too hard for me, I ask for something easy. The Flutter is very hard to do so I don't like doing it. When I'm doing something really fun, like playing a game, and my mom says, 'It's treatment time,' and I don't want to do it, I make sure that I'll be able to come back to my fun stuff. Usually, I do

(Continued on page 5)

## Compliance *(Continued from page 4)*

treatments at the same time every day, so I just expect them. Mostly, I want scientists to invent a cure for CF. Mom says I need clean lungs for when there is a cure.”

Isa, a 28-year-old with CF, is rigorous in following her daily treatment and medical routine. She says, “I tend to be more passionate about complying with my medical regimen when I have a doctor who is also passionate about fighting this disease. I think it is imperative that a doctor is aggressive, even for a “mild” case (after all, I believe everyone who is “mild” becomes “severe” some day). We (together) need to do everything in our human power to fight it. I have had passive doctors who say, ‘You can use puffers instead of nebulizers,’ or ‘You don’t have to take all these meds,’ but I know what makes me feel better. A decade ago, I wouldn’t have taken my disease as seriously if my doctor hadn’t. Now I know better. When I have an open and relaxed relationship with my doctor, I can state what medical regimen I am able to follow. Doctors need to recognize that people with CF are tremendously busy with the ordinary demands of life. When a doctor drills me with, ‘Do four treatments a day no matter what!’ I cannot agree. It is impossible when I have to work, run errands, care for family members and so on. It helps me when doctors make time to hear what my life is like. Recently my doctor said, ‘Wow, you really manage a great deal and I am impressed.’ Sadly, things that would help with time management (and therefore with compliance), such as portable nebulizers, have never been recommended to me by doctors, but rather through the CF lay community.” Isa uses the CF community as much as she can for additional support.

Anna is 19 years old and was diagnosed when she was a toddler. Anna’s motivation is very clear-cut. “If I know I’m going to have to go on I.V.s, I’ll do almost anything to avoid it. That’s how much I hate it. It completely stops my life. I can’t work and I feel like I’m sick. This is the only time I really think of myself as different. Treatments are not the same. They are such a part of my life, that I don’t really even notice them. I do two treatments every day, basically always. When I went through rebelling, it was more about food. But I didn’t ever stop doing respiratory treatments. I knew if I did, I’d get sick. I also believe the doctor knows best. Since I really trust my doctor, and my doctor knows how much I hate I.V.s, I know he is trying to help me avoid them. In terms of my parents, it makes a difference what they say. If they stay out of my way and don’t push things, I’ll generally do it myself. But if they’re butting in and saying, ‘well you didn’t do this,’ I get mad and don’t want to do it. I don’t like to be nagged.”

For Larissa, age 23, taking medicines and adhering to her medical regimen is an internal motivational process. She says, “It’s as simple as this: If I don’t take care of myself, I don’t feel good! And no one else can make me healthy—so it’s got to be up to me. Still treating CF requires a ton of motivation, work and willpower on my part. It takes me longer to get ready in the morning, and I repeat the process again at night. I have to work exercise into my schedule, (at least 30 minutes 3-5 times a week) leaving extra time to cough afterwards. At times, keeping up my health is incredibly discouraging. When my internal drive to keep up my health fails me, I’ve got external sources offering me support. When I’m too tired to do my breathing exercises, my boyfriend is there to encourage me. I know that my parents will never scold me for running late if I’m trying to finish up some inhaled antibiotics (even if it’s really my fault for sleeping in!). The last thing they would ever do is make me feel bad for something I can’t help. And last time I visited my wonderful doctor, he called me an “inspiration” because I exercise so often. Now I feel as though I’m failing him (and myself) if I skip the gym after work and sit on the couch instead. Every now and then I’ll skip some medicine because I’m tired, or I don’t take a pill in front of someone who doesn’t know about my CF. I almost always regret it. Physically, I have to care of myself—but emotionally, I could never do it alone.”

Clearly, many CF patients are able to make regular treatments part of their lives. For others, the battle continues between what will help them stay healthy and trying to live

*(Continued on page 7)*

## Cisapride *(Continued from page 4)*

drug and seeing how they feel without it. For many people, discontinuing use was not that difficult.

If your GI health declines after you have discontinued Cisapride, Dr. Bass suggests looking for alternatives. If you are using Cisapride for heartburn, there have been many new, safe and very effective acid-blocking drugs introduced since Cisapride came on the market, such as Prilosec®, Prevacid®, or Pepcid®. For people with stomach emptying problems, the drug Reglan®, the only drug used before Cisapride’s availability, remains the only alternative. While helpful, Dr. Bass points out it has some bothersome side effects (such as involuntary muscle contractions), especially when taken in high doses for long periods of time, and has been less effective than Cisapride.

Finally, you may feel Cisapride is the safest and only effective option for you. For such people, the company has agreed to put a mechanism into place to allow you to get it. While it will be provided to you for free, you must also agree to participate in the equivalent of an ongoing study. After signing a consent, you must have screening laboratory tests done, and then suffer the ongoing inconvenience of frequent heart monitoring. This may be nothing however, compared to the painful GI condition you are experiencing. So, as long as you are willing to follow the mandatory procedures stated above, the drug can be made available to you. It does take some time to start up the process, so talk to your GI doctor about how to get it right away. ■

## CFRI Rummage Sale Date Changed!



In the last newsletter, we asked you to start saving all clean, usable clothing, linens, furniture, toys, books, video tapes, household items, etc. We still hope to have the Rummage Sale, but the date may have to be changed from October 7<sup>th</sup>, 2000. CFRI may be forced to MOVE due to increasing rent cost. Depending on when this occurs, the Rummage Sale may be postponed. Call the office: 650-856-0546 after September 1<sup>st</sup> for more information. ■

# Cystic Fibrosis Research, Inc.

## Summary of the Annual Report for 1999

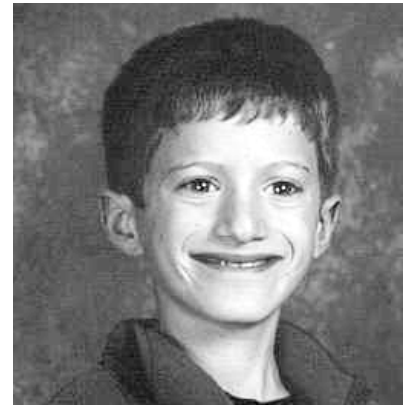
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**Cystic Fibrosis Research, Inc.** (CFRI), an independent nonprofit organization, has been a leader in sponsoring cystic fibrosis biomedical research and education since 1975. Our mission is to end the emotional and physical suffering associated with cystic fibrosis (CF), a fatal genetic disease that affects 70,000 people worldwide and 30,000 people in the United States. Our mission consists of two interrelated efforts: to fund cystic fibrosis research, which will aid in controlling CF and will lead to a cure or control, and to provide educational and support programs for individuals with cystic fibrosis and their families.

The Research Advisory Committee (RAC) evaluates submitted CF research proposals by sending them to outside researchers for peer review, and compiles their input in order to prioritize the proposals. The committee then allocates research funds accordingly. The recommendations of the RAC are sent to the Board of Directors and the general membership for a final vote on the distribution of research funds. CFRI supports basic CF research, clinical research and gene therapy research aimed at improving the quality of life for people with cystic fibrosis.

CFRI provides up-to-date, educational information for our members through a number of important programs: an annual CF Conference for people with CF, their families and health care providers; a website ([www.cfri.org](http://www.cfri.org)); a newsletter circulated to over 12,000 families worldwide; an educational video library; a CF Adult Retreat; and support groups for adults with CF and their families. CFRI recognizes that knowledge and self-management are critical to promoting optimal health for every CF patient.

This year, CFRI celebrates its Silver Anniversary! We are in our 25<sup>th</sup> year of research and education! For fiscal year 1999, we have provided \$406,322 toward research efforts. There is no doubt that cystic fibrosis will be conquered—just how soon depends on us. Your commitment to CFRI's vital work has never been more important than it is today.



*Jeremy, age nine, has cystic fibrosis and requires special care. He takes multiple medications daily to aid in digestion, control infection and reduce inflammation. Exercise and airway clearance therapies are vital parts of his daily health regimen. He requires high-caloric intake to ensure adequate nutrition and weight gain. Unfortunately, these home therapies will not be enough. Jeremy may be hospitalized many times during his shortened lifetime. Help us to help Jeremy and all people with cystic fibrosis.*

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

To Our General Membership and Generous Donors:

CFRI enjoyed a successful year in 1999, lead by revenues of \$799,000. This is a 13 percent increase over last year! Buoyed by this generous level of support, we were able to increase funding during the year for our ongoing educational and support services. Because of your generous donations, CFRI is able to put on an annual educational conference and an adult retreat, offering scholarships to those who otherwise might not be able to attend. Our newsletter is sent out three times 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 increase funding for research. CFRI awarded research grants in the amount of \$406,322 (1998 research grants totaled \$289,000).

Overhead expenses and fundraising costs rose \$28,000 to a total of \$220,000 for the year. Residing as we do in an area enjoying unprecedented economic growth, we must

compete for paid help and volunteers in an extremely difficult market. Adding to this challenge will be the expiration of our office lease early next year. We will need to find new accommodations at an affordable rate.

Net assets stood at \$113,000 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 1999, including the audited financial statements, is available upon request. Please contact the CFRI office.

Sincerely,

Mike Roanhaus  
President of the Board

## Ana Stenzel Receives her Transplant!

Ana Stenzel, a 28 year old with CF and long-time CFRI volunteer, received her long-awaited lung transplant on June 14, 2000! Ana was called at 4:30 a.m., and was prepped and rolled into the O.R. by 2:30 that afternoon. She was in the recovery room by midnight. Her twin sister, Isa, who also has CF, was at her side in Intensive Care by 1 a.m. on June 15<sup>th</sup>. Ana was discharged from Stanford Hospital 12 days later. Despite the nurses' strike at Stanford and Lucile Salter Packard Children's Hospital at Stanford, the family felt that Ana's nursing and physician care was excellent during her stay.

Ana first discussed the possibility of lung transplant with her doctors in 1991. She was eventually listed in 1997, although went on frequent "hold" during periods of improved health (usually during the summer months). Many remember Ana (especially at last year's CFRI conference) eloquently verbalizing the struggles involved with choosing and waiting for a transplant. She (as well as others) shared with many of us the struggles, fear, loss, and ambivalence that come up with a decision of this magnitude and the long waiting period that one must endure. But when the actual moment of truth arrived, Ana was calm and ready. She felt very at peace with her decision for transplant.

Her sister Isa reported that those days immediately following the transplant were the hardest of Ana's life. "She had to really choose to live, and once she did her recovery process took off. After transplant" Isa said, "her blood pressure was very high." (This is normal for a post-transplant situation.) "Ana felt flushed, weak, and had frequent headaches. But now she is doing amazingly well. It is a very happy time for our family. We are all together a lot and having lots of parties, including a huge Fourth of July party. We are really celebrating how well she is doing."

Ana's mom, Hatsuko Arima, who is an L.C.S.W. at Kaiser in Los Angeles, and her father, Reiner Stenzel, who is a professor at the University of California at Los Angeles, both arrived immediately after transplant and have been caring for Ana through her post-transplant climb back to health. Ana's boyfriend, Rob Rohde, has also been very present and supportive throughout the ordeal.

Ana says, "The first two weeks after transplant were very, very challenging. It was hard waiting for the incision to heal, getting adjusted to all the new medications, dealing with being in the I.C.U. and the lack of sleep. My new lungs were not fully inflated, so I felt very short of breath. Fortunately, this is improving daily. With a lot of exercise, my lung capacity three weeks post-transplant is 59%! Pre-transplant I was about 25%. It feels incredibly different to be walking without oxygen. My current anxieties are mostly about germs and remembering the new routine (it's a totally different protocol than before). I take approximately 20 different meds daily and it is a challenge to learn the new routine. The beauty of it is there are no treatments...I can breathe and I have a lot of energy! I look forward to things just getting better and better. Of course, there are some things that are hard to let go of...I still want to sleep with a little oxygen at night. I get anxious. I've been wearing oxygen for 10 years at night and it's hard to sleep without it and trust that the new lungs will work."

We asked Isa how she felt about her twin sister's whole experience. "Well I am just overjoyed at how well Ana's doing and I'm relieved that she is out of danger. I simply cannot find the words to express how grateful I am that she gets this second chance... to breathe well, to NOT do therapy, to travel and do all the things she couldn't do before. There will, of course, be a change in our relationship. As Ana is immuno-compromised, we must be very careful about contact, especially for the next three months, and that will be hard. And we'll never be able to do treatments together like we did before. It changes the dynamic...but that was happening anyway just with the normal events of life like boyfriends and marriage. Mostly, I feel incredibly happy!"

The lungs were donated from a young man in Portland, Oregon. The Stenzel family, as well as the entire CF community, is eternally grateful for the loving gift that family made in an hour of enormous grief for them. ■



*Ana Stenzel smiling happily two days after her double lung transplant. With her are her parents Hatsuko, and Reiner Stenzel.*

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## Compliance

 (Continued from page 5)

what they consider a "normal" life. I am in the process of retraining my brain in an effort to achieve compliance. This is probably most important for people diagnosed later in life, or whose once relatively mild disease suddenly takes a turn for the worse. I'm convinced that it's all in my head. I know that in order to stay alive, I must do my treatments and make them a priority in my life. I need to believe that my treatments will keep my lungs in shape and make them stronger. I'm sure that when I achieve this goal, my lungs (and my parents) will be much happier! ■

## Behind the Scenes at CFRI... Meet the Research Advisory Committee

CFRI's Research Advisory Committee (RAC) consists of seven longtime, dedicated volunteers, and one staff person. They oversee the process of soliciting research proposals, sending them out for peer review, and recommendation of funding.

**Burt Jones, Ph.D.**, holds a degree in astronomy. He has just recently agreed to chair the RAC. He is eager and very willing to go the extra mile needed to handle the review process and evaluate these proposals. We are very lucky to have Dr. Jones. In his own words: "I am a professor at UC Santa Cruz, and Assistant Director of Lick Observatory, south of San Jose on Mount Hamilton. I am married to Mary Jones, and we have two children. Michelle is 13 and has CF, and Kristine is 15 and does not. Outside of the RAC, my hobbies are backpacking (with Kristine) and kayaking (with Michelle). I have served intermittently on the RAC for over ten years. I believe the wise distribution of CFRI's money is equally important to the process of raising it. This is my main interest in serving on the RAC. In the past, I have been successful at obtaining grants from federal agencies, and serving on many review panels for the evaluation of grants. It is these skills I bring to my role on the RAC."



*Burt Jones, Ph.D., and daughter Michelle*



*Meg DeLano, M.D.*

**Meg DeLano, M.D.**, is an allergist and pediatric pulmonologist. She has been very active in CFRI's annual Conference and Retreat over the years serving as our Medical Advisor. She served on the RAC for years and was CFRI's most recent RAC chairperson. CFRI is deeply indebted to Dr. DeLano for her hard work and dedication. She has always supported people with CF. Dr. DeLano says, "I have reviewed the RAC grant proposals every 6 months for over 15 years. During those years CF researchers have been on the cutting edge in multiple fields: organ transplant, genetic analysis, gene therapy, understanding of infectious disease transmission mechanisms, to name a few. The force that keeps me on the RAC is my personal fascination with this unbelievably complicated disease that eventually involves every organ of the body as well as challenging the spirit of those who must cope with it. There is nothing in my medical training that goes unused."

**Martin Kharrazi**, who has a Ph.D. in epidemiology, is another key, long-term player on the RAC. He says, "I have a 9-year-old son, Jeremy, with CF, and four other children without CF. I have served continuously on the RAC since December 1996. I am trained in epidemiology and have worked for the California Department of Health Services for the last ten years. One year ago, I started working for the Genetic Disease Branch. I am now doing research that will help our efforts to add CF to the list of newborn disorders for which California is currently screening. The type and frequency of CF mutations are being determined in Californians who have CF. Then the correct panel of mutations can be selected for use in the screening program." CFRI supports Dr. Kharrazi's important work with newborn screening and has sent a letter to the state of California in support of the screening of newborns for CF.



*Martin Kharrazi, Ph.D., with his son Jeremy who has CF.*

**Joann Blessing-Moore, M.D.**, is an allergist and pulmonologist in private practice in Palo Alto, CA. She says, "Cystic Fibrosis research and patient management has been an important part of my life. I was a Cystic Fibrosis Foundation Fellow at Stanford and later co-directed the Pediatric Allergy/Pulmonary Department at Lucile Packard Children's Hospital at Stanford. When my daughter was young, I moved on to the Palo Alto Medical Foundation and continued in Pediatric Pulmonary work (as well as allergy), following our patients with CF in the community. In 1990 I began my own practice, but continued to be active with our 'shared patients,' as well as committees in the hospital and community. I am interested in working with CFRI because of my interest in cystic fibrosis. There is fantastic research potential in this area and I'm excited that CFRI is helping to make funds available. It is exciting to be involved in this process and work with such a special team on the RAC. The support of CFRI has been greatly appreciated."



*Joann Blessing-Moore, M.D.*

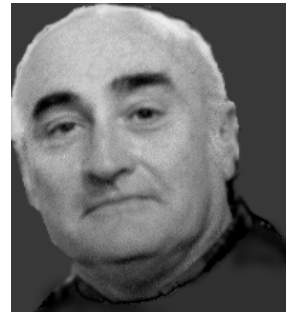
*(Continued on page 9)*

**Doug Modlin**, holds a Ph.D. in electrical engineering. He joined the RAC in 1983, and later served as RAC Chair from 1985 until 1996. He is married to Robin Modlin (a former president of the CFRI Board of Directors). They have two children, Anna, age 19, (who has cystic fibrosis), and Sara, age 12. Doug says, “Initially, I was interested in research, as Anna was sick in the hospital, and I wanted to understand all the medical procedures. I became involved with CFRI six months after Anna’s diagnosis when she was two. I wanted to find something that I could do that was related to my interest in science and could use my academic training. Being involved with CFRI and the RAC was how I could help. Instead of CF just happening to my daughter and me, I had a chance to influence the future. I felt empowered as opposed to sitting on the sidelines watching CF happen to us. If I died tomorrow, I would want the message of my life to be about getting involved and being part of the solution.”



*Doug Modlin, Ph.D.*

**Rob Robinson** holds a Master’s Degree in Product Design. He is married to our former, long-time Executive Director, Ann Robinson. He has two children, Clare, age 27, and Carl, age 22. Carl was diagnosed with CF at birth. Rob has served tirelessly on the RAC, as well as helped to monitor CFRI’s database requirements over the years. In Rob’s words, “17 years ago, when my son Carl was five, I became interested in the process of evaluating research proposals. My training as a mechanical and product development engineer lends understanding to the basic transport and regulation problems in CF. It is great to work with the wonderfully gifted and scientifically oriented volunteer members of the RAC.”



*Rob Robinson*

**Valerie Baldwin** became involved with CFRI in 1983, a year after her family moved from San Francisco to the peninsula of the Bay Area. Her son, Ian (currently 18), who has CF, was two years old at the time. Valerie says, “I joined the CFRI Board as Financial Officer and served for 5 years. I attended (with my son Ian) CFRI’s first CF camp as the Board observer and representative. My husband, Geoff, joined the RAC about the same time and served for 12 years. I replaced Geoff on the RAC in 1998 after returning to school and receiving a Master’s degree in Molecular Biology. I am now working at Dr. Jeff Wine’s Stanford Cystic Fibrosis Research Lab. As CF parents, both Geoff and I have worked hard to keep up on the latest in CF research and ensure that CFRI funds only the best and most promising projects. For me, that is what volunteering on the RAC is about.”



*Valerie Baldwin*

**Marion Rojas**, a CF adult, holds a Ph.D. in neuropsychology and has had a lifelong interest in science. She says, “I am a natural born researcher and I love micro-biology. All of CFRI’s proposals for funding are sent to each member of RAC. We also receive copies of the peer reviews of each proposal well before each meeting. We have a chance to read through the proposed research and evaluations, and confer with each other. We then rank them for suggested funding for the Board and membership. I love getting even a faint glimmer of the possibilities in store for people, especially those with CF. I even pull up the human genome project on my computer and read through the new gene mapping they’ve uncovered. To get this glimpse of the future through my own research and through my involvement with CFRI’s RAC gives me the courage to keep going. Indeed, it is the clue to my longevity.”



*Marion Rojas, Ph.D.*

**Taylor Bui** is CFRI’s new Program Services Manager. In this capacity, she serves as the staff liaison to the RAC. Taylor works very closely with Burt Jones, RAC Chair, handling the administrative end of the research proposal submissions, reviews, and funding. Taylor says, “I’m thrilled to join the CFRI circle of family and friends, and I hope my presence will make a positive impact on its search for a cure for cystic fibrosis.” ■

## **Golfers! August is the Month of your Dreams!**

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Do you enjoy playing golf, good food, fine wines, dinner with friends, and taking your chance at an auction or raffle? Imagine being able to do all that and helping to find the cure for cystic fibrosis! This year there are two great golfing events for you to participate in with your friends. Both events benefit Cystic Fibrosis Research, Inc.

CFRI's Year 2000 Golf Tournament takes place Monday, August 21<sup>st</sup>, 2000 at the beautiful Cinnabar Hills Golf Course in the hills of south San Jose. Player registration begins at 10:30 a.m., with a box lunch at 11:30. Shotgun teeoff is at 12:30 p.m. If you are more the wine and cheese type, join us at 5:30. This is when the Silent Auction starts. Dinner, player awards, and the Live Auction all start at 7:00 p.m. Join us for an incredible day. It's not too late! Call the CFRI office to register at (650) 856-0546.

The Fourteenth Annual Bob Mackey Memorial Golf Tournament and Banquet takes place Friday, August 25<sup>th</sup>, 2000 at the Santa Teresa Gold Club in San Jose, CA. Check-in starts at 11:15, lunch is at 11:45 and the shotgun tee-off begins at 12:30. Wine tasting commences at 5:30 and the banquet, awards and program begin at 6:45. For more information on this fabulous event, or to be a sponsor or donate a prize, call the Robert J. Mackey Foundation at (408) 356-6429 or email them at [BobMacMem@aol.com](mailto:BobMacMem@aol.com) for more information.



## **THANKS TO OUR SPONSORS FOR THEIR GENEROUS SUPPORT!**

CFRI's Annual Educational Conference is upon us. It is important to remember that this event 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. Also we can make scholarships available to those people who might not otherwise be able to attend. Many people describe the conference as the event that keeps them going throughout the year. Remember our sponsors...visit their booths at the conference and thank them for making this invaluable event possible. The following sponsors contributed to this year's Annual Conference:

**American Biosystems, Inc.**  
**Axcan Scandipharm, Inc.**  
**Digestive Care, Inc.**  
**Florida Cystic Fibrosis Pharmacy**  
**General Physiotherapy, Inc.**  
**Genetech, Inc.**  
**PARI Respiratory**  
**PathoGenesis Corporation**  
**Vortran Medical Technology 1, Inc.**

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### **Director's Message** *(Continued from page 2)*

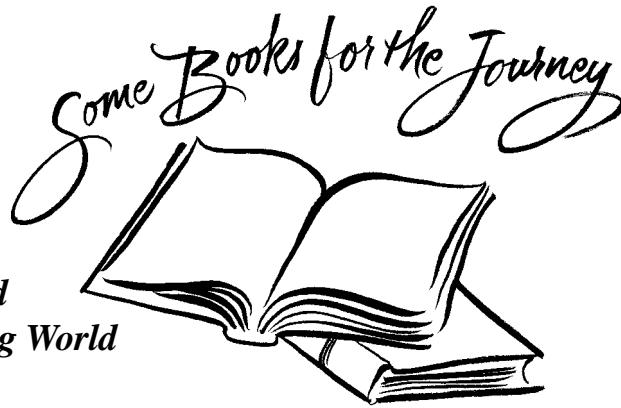
support, and research this disease until there is a cure.

In the midst of running any office, there is a formidable amount of administrative distractions. A growing CFRI is no exception. Still, it is you who live with CF, who call for support, who send tea letters on behalf of someone you know with CF, you who remind me of **why I am here**. And you give me the strength I need to focus on the issues, get them resolved and move on to the single most important issue: YOU. I want to get this disease cured and I want to see that grandmother and her grandson worrying about nothing more than when to go for ice cream, and arranging sleepovers for the weekend and trips to the beach. I won't rest until we're there!

Thanks for all of your support. I look forward to seeing you at the annual Retreat or Educational Conference this August. Please continue to introduce yourselves to me and my staff and share your stories. YOU are our inspiration.

Warmest regards,

Calvinia Williams



## ***Second Opinions: Stories of Intuition and Choice in the Changing World of Medicine***

Jerome Groopman, M.D.,

Viking, © 2000 Penguin Putnam Inc., 243 pages

One of the greatest challenges that patients and their families face is how to evaluate and act on medical advice. In *Second Opinions*, Dr. Jerome Groopman, an eminent physician at Harvard Medical School and leading cancer researcher, explores how patients and their doctors make these critical decisions in the field of medicine where knowledge is imperfect, no therapy is without risks, and no prognosis is fully predictable.

We witness eight gripping clinical dramas and see how the physicians, patients and families deal with conflicting medical opinions, health insurance, and HMOs' rote responses. These real-life stories demonstrate how to navigate the uncertain terrain of illness, and model for the reader the process of balancing intuition and information in order to make the best possible decisions about one's health and future.

The decision to seek another viewpoint on medical treatment might mean the difference between life and death. Yet additional expert advice—at one time required by many health insurers before patients could undergo surgery—is now underused and underappreciated, jeopardizing the quality of care patients receive. Patients may also feel further confused and torn by conflicting diagnoses presented by different experts. This book gives some insight on how to proceed through this fog of uncertainty.

Dr. Groopman understands this from a personal level as much as a professional. After nearly losing his nine-month-old son to multiple medical misjudgments, he has made it his job to encourage patients to question their physicians and ask for expert second opinions. He clearly illustrates that improper diagnoses can be deadly. In some situations, death is inevitable. But how the patient and family face that death is also important.

Dr. Groopman says, "People feel very inhibited in asking for a second opinion". We don't want adversarial relationships. We want good relationships with our doctors. Learning how to cultivate these relationships is an art form worth studying.

## ***Between Husbands and Friends***

Nancy Thayer, © 1999 St. Martin's Press, 241 pages

*Between Husbands and Friends* is a plain, old-fashioned, good summer read with a CF twist. Basically it is an intriguing novel about relationships in families, and the ups and downs these relationships (mainly marriages, but also parent-child) go through over time.

One of the children in the book has cystic fibrosis. The CF is immediately obvious to anyone in the CF community reading this book (connoisseurs of CF as we are), and for myself anyway, that added to the drama. It was just a matter of time before diagnosis. But the spider web of life is tangled in this book and a lot transpires before this child is properly diagnosed and treated. To say anymore would give the plot away.

It's a fun summer read, not too heavy, and not too light. Pack this one in your suitcase for your vacation. And Bon Voyage! ■

## **Giving Gifts of Stock to CFRI**

Giving a gift of stock to Cystic Fibrosis Research, Inc. (CFRI) is easy and can be rewarding in several ways. You will not have to pay capital gains tax on stock that has appreciated over the years. You will receive an income tax charitable deduction for the full-fair-market value of the stock on the date of the gift, and your stock gift will benefit CFRI, making possible further research and educational programs for our families struggling with CF.

### **How can you deliver the stock to CFRI?**

#### ***Securities held by the donor:***

If you hold stock certificates that you wish to donate to CFRI, please let us know and then contact our broker, Mr. Jim Kirwan at Smith Barney, Inc., 1-800-284-2405 for complete instructions. Your gift date is the date you mailed the certificates. The value of your gift is the mean of the stock high and low for that day.

#### ***Securities held by a broker:***

If your broker holds the certificates, instruct your broker in writing that you wish to make a contribution of securities to Cystic Fibrosis Research, Inc. and provide details on the company whose stock you hold and the number of shares you wish to donate. Instruct them to transfer the stock to Smith Barney, Inc., DTC # 0418, for credit to client Cystic Fibrosis Research, Inc., (account #595-11130-1-7-093). **Send a copy of your instructions, along with a letter stating the purpose of the gift to Calvinia Williams, Executive Director, CFRI, 560 San Antonio Rd, Suite 103, Palo Alto, CA 94306.** The gift date is the day the securities are transferred to CFRI's account. The value of your gift is the mean of the stock high and low for that day. If you want to make an end-of-year donation, please begin the process now before the end-of-year rush at your brokerage firm. If you have questions about stock gifts or want to proceed with a gift, please contact the Calvinia Williams, Executive Director, at the CFRI office (650) 856-0546. Thank you for considering a gift of securities for your friends and loved ones with cystic fibrosis.

# CFRI's Year 2000 Educational Conference— **LAST CHANCE!**

This year's Educational Conference will be held August 11-13, 2000 at the DoubleTree Hotel in San Jose, California. The DoubleTree Hotel is located near the San Jose Airport at 2050 Gateway Place, San Jose, California. For complete conference information and registration packets, contact CFRI at 560 San Antonio Road, Suite 103, Palo Alto, CA 94306, by phone at 650-856-0546, by fax at 650-856-0554 or by email at [cfri@cfri.org](mailto:cfri@cfri.org).

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CFRI will begin sending emails for web newsletter readers with our Fall 2000 issue.

## Could your Environment be Making you Sick?

By Ann Robinson

Since the 1970's, homes, schools and office buildings have been built with airtight specifications to conserve energy. This causes airborne particles to remain in a room because they have nowhere to go. According to the Environmental Protection Agency, indoor air is up to 70 times more polluted than outdoor air. The American Lung Association states that people spend about 90% of our time indoors, with 60% of that time at home. Many illnesses such as allergies, asthma, and hay fever are exacerbated by polluted indoor air. These illnesses can aggravate any lung disease—including cystic fibrosis.

For people with allergies, the immune system reacts to different substances known as allergens. Allergens include household dust, dust mites, mold spores, pet dander, trees, grass, weed pollen, tobacco smoke, etc. When the immune system comes in contact with one or more of these allergens (depending on what your system reacts to), it becomes overloaded. This in turn causes stuffiness, nasal discharge, itchy nose and sneezing. By avoiding the allergens that cause this reaction, you are giving your body a chance to recover.

This article will focus only on mold allergies that can be especially devastating to people having cystic fibrosis. Mold is a form of fungus, which is in the plant family. It survives on and eats tiny amounts of plant or animal matter. Mold reproduces by releasing spores into the air. They settle on organic matter and grow into new mold clusters. It is these spores that can trigger allergic reactions when inhaled. These reactions include coughing, wheezing, bronchial spasms, pulmonary infiltrates, central bronchiectasis, and ABPA (allergic bronchopulmonary aspergillus). Aspergillus is a fungus that can grow inside the lungs of people with CF causing medical problems, especially when that person is allergic to the fungus.

**Indoors**, the mold season is year round. And since people spend so much time indoors, it is essential find out if mold is growing in your home, and especially in the bedroom. Within the home, mold can be found growing on houseplant soil, in carpeting, on antique furniture, in the bathroom, and in the air-conditioning system. **Outdoors**, mold spores may be present in the air all year, except when snow is on the ground. People sensitive to mold should avoid exposure to fallen leaves, cut grass, compost piles, rotting damp wood, barns and places that receive little sunshine.

If you have been diagnosed with a mold allergy, the following information is important:

1. Reduce the humidity level in your home to 40%- 45% by using an air conditioner with an electrostatic air filter and/or an electric dehumidifier. This lower humidity prevents mold spores (and dust mites) from reproducing.
2. Get HEPA (High Efficiency Particulate Arresting) Air purifiers to reduce airborne mold spores. These are particularly effective if they run continuously in the bedroom.
3. Clean surfaces where mold grows and use an inhibitor to prevent future mold growth. Keep potted plants that require moist soil outdoors.
4. Never clean carpeting with steam. Moisture trapped in the carpet pad will create a haven for mold growth. It is best to remove any carpeting from the bedroom.
5. Have someone else mow and rake the lawn. If you must work in the yard, wear a high quality mask.
6. Do not hang clothing outdoors. This makes an attractive landing surface for airborne mold spores.

(Continued on page 13)

## Environment *(Continued from page 12)*

7. If you must ride in a convertible or jeep with the top down, wear a high quality mask.
8. If you use a humidifier during the dry season, be careful not to over humidify.
9. Since everyone perspires during the night, buy a new pillow every year. Be sure to use a pillow with an agent that will inhibit the growth of mold. Wash pillows regularly. Get pillow and bed encasings that will protect you from mold in the pillow and mattress. Wash them regularly too.
10. Do aerosol treatments in a room other than the bedroom because aerosol treatments put moisture into the air. Do not shake nebulizer moisture into the bedroom air or on the floor. Completely dry all aerosol equipment between treatments.
11. Keep dirty, sweaty, or damp clothes in a closed hamper or outside the bedroom. They make an attractive damp place for airborne mold spores to multiply.
12. If bed linens become damp with sweat, either change the bed or let them air dry instead of making the bed with damp sheets.
13. Be sure your clothes dryer is vented to the outdoors.
14. While allergy shots are controversial, in many cases they can be helpful. Talk with your allergist (in this case, a specialist is best) about the possibility of allergy shots.

Our son Carl, who has cystic fibrosis and ABPA, was very ill until November of 1999. He was consistently sick, hospitalized three to four times per year, unable to attend college or to work full-time. One of our son's physicians convinced us that Carl was being constantly reinfected by mold spores growing in our home. With her encouragement, I ordered mold test kits to locate where the mold was growing. With this information, we determined that Carl must move to a dryer bedroom that had no mold. If we had known and utilized the above information six years ago, we could have saved several hospitalizations, anguish and costly medical bills. ■

*The above article was gleaned from articles in magazines and catalogues, and from discussions with Carl's physicians. We are especially thankful for the advice of Margaret DeLano, M.D., CFRI's Medical Advisor.*

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## Vehicle Donations!

From January to June, 2000, CFRI made \$1,020.00 on vehicle donations! CFRI has been able to hold vehicle donation expenses down by dealing with an auction company that manages DMV paper work, hires towing companies to pick up the vehicles, and then auctions them. For these services, the auction company charges ten percent of the auction proceeds, and the towing company charges \$75 for the delivery of a vehicle to the auction site. Also, a CFRI volunteer coordinates the donations. This saves the administrative costs other charities incur when they contract with an outside firm to manage vehicle donations.

Beginning January 1, 2000, new California legislation required an additional charge of \$55 for a smog check and a \$75 safety check for every vehicle sold to the public by an auction company. Thus, CFRI earnings have been decreased by \$135 per donated vehicle. Despite the decreased revenue, vehicle donations remain a good source of funding for cystic fibrosis research and education; this method of fundraising requires almost no staff support and it is painless to the donor and to CFRI.

If you have a used car, boat, recreational vehicle or motorcycle, and could use a tax write-off and would like to support CFRI research and education, please consider donating your vehicle to CFRI. You can contact jubilee@sirius.com or call the CFRI office at (650) 856-0546 for additional information. ■

## TOBI® Available Internationally

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The PathoGenesis Corporation drug TOBI® (tobramycin solution for inhalation) has received marketing clearance in a number of countries around the world. The drug, which is popular in the United States for the treatment of lung infections, is now being marketed overseas. In the U.K., the Department of Health has licensed TOBI. The Ministry of Health gave marketing clearance in Israel. And the Therapeutic Goods Administration gave its marketing clearance in Australia. The U.K. is the lead country for seeking regulatory approval of TOBI in the European Union. The other E.U. countries will be asked to approve the inhaled drug through a mutual recognition process. Distributor agreements have been made in a number of other European countries including Germany, the Netherlands, Greece, Cyprus, Italy and France. This is the first step toward the availability of TOBI in these countries. TOBI is used for the long-term management of chronic pulmonary infection due to *Pseudomonas aeruginosa* in people with cystic fibrosis age six years and older.

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## CFRI Greeting Cards: Committee Recruitment

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Everyone who enjoys CFRI Cards and Novelties is urgently asked to help Pat Flynn and participate in their creation. No artistic talent is required. The most difficult part is lining up a card with the grid on a paper cutter. Those who are timid about this can package finished cards (requires the ability to count up to six!). Workshops are planned at the CFRI office: July 22nd or 23rd and October 21st or 22nd. If you have a preference for Saturday or Sunday, morning or afternoon, please call the CFRI office at 650-856-0546. Working together, sharing ideas and experiences, and helping cystic fibrosis research is fun and worthwhile. Bring a friend. Thanks.

# CFRI Poster Child Photo Recruitment

Have you ever wished your child was pictured on the Mother's Day Tea 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 we need a picture of a child with CF by him/herself (usually a more serious pose is appropriate) and another with one or both 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, 560 San Antonio Rd., Ste. 103., Palo Alto, CA 94306-4349.

# Honorariums

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

Sadie Anderson	Jessie Franks	Alex Molle	The Swanson Family
Marilyn Bates-Prator	Emily Fredricks	The Oneto Family	Larissa Swanson
Brett Bennett	Jessica Fredricks	Dale Peterson	Heidi Tegner
Kandra Cady	The Gerow Family	Kent Peterson	Mrs. Benedict Viering
Richard Chartrand	Barbara Greenberg	Erin Phillips	Tom Walton
Cameron Cornell	Alexandria Hampton	Robyn Primack	Jeffrey Wine
Dr. Jill Doran	Elizabeth Hampton	Briauna Red	Marlene Wine
Tess Dunn	Tyler Heavner	Ben Robertson	Timothy Yang
Thomas Evans	Erin Hoyt	Ann Robinson	
Alanah Fink	Will Huston	Carl Robinson	
Kim Fiske	Lori Kipp	Clare Robinson	
Pat Flynn	Dennis Logie	Rob Robinson	
Ryan Foster	Leo Lunine	Jack Rubin	
Cody Franks	Ron Marigian, Jr.	Sarah Skeffington	

# 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.*

David Armknecht	Albert Finston	Lawrence Lavignino	Erik Sena
Jodie Armknecht	Rosalie Goerss	Mamie Lilly	Gloria Sexton
Mariana Avila	Isaac Goldman	Arnie Lowder	Gloria Smerber
Debbie Ware Babbitt	Dora Goldsworth	Robert McAllen	Barbara Smith
Gerald Becker	Brother	Rose Millikan	Helen Stillings
Annie Beltrame	Robert Gray	Jessica Mobley	Gladys Strook
Bimbo Buonamici	Donni Grotte	Amy Morris	David Stuckert
Kyle Butler	Dorothy Hanna	Mrs. Moss	Laurie Stuckert
Tim Calero	Rahbanu Hassung	Thomas Murphy	Mel Stuckert
Ada Cohen	Leonard Hays	Scott Nelson	Howard Tani
Lillian Corda	Betty Heasley	Celia Newberger	Dresden Tingley
Donald Davis	Edith Hesel	May Owerk	John Trask
Keith Davis	George Hoeck	Angie Paganelli	Marge Trask
Kevin Denham	Margaret Howell	Lorraine Parriera	Linda Trojak
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Hilda Dodd	Eva Jones	Tim Prater	Mark Weidenman
William Dougherty	Leslie Kalbach	Gordon Rudel	Tara Wier
Charles Dye	Kathy	Ian Scully	George Wright
Rose Essig			

## 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, 560 San Antonio Rd., Suite 103, Palo Alto, CA 94306-4349

**CFTR** (Continued from page 1)

function and its relationship to the other players participating in that function is not well understood. For example, we do not yet know whether its function when CFTR is around is different from how it works when mutant CFTR is present, or when CFTR is completely lacking in epithelial cells.

One important requirement of addressing such a question is to have a good model system. Such a system ought to 1) lack functional CFTR (that is, either have no wild-type CFTR, or be completely lacking in any form of CFTR at all), 2) retain other epithelial characteristics (ability to either absorb or secrete), 3) express at least one identifiable alternative chloride channel (in this case, CIC-2), and 4) for practical reasons, be robust and easy to propagate.

My work has focused on identifying and characterizing such a system. In addition to specialized transport functions, the epithelial cells of the thyroid gland also have general transport processes that mirror those of epithelia affected in CF. I study an epithelial cell line derived from the Fisher Rat Thyroid (FRT). Although FRT cells express CIC-2 and retain both epithelial morphology and function, they lack CFTR. I use a combination of molecular biological and physiological tools to test whether endogenous CIC-2 can be activated in FRT cell sheets. I have found that several stimuli, previously shown to activate CIC-2 in non-epithelial cells, do indeed increase the total conductance of the epithelium. It will be important to know whether and, if so, how this response is affected when disease-associated and wild-type forms of CFTR are introduced to these cells. Such information will shed light on the question of how CFTR acts as a regulator of other epithelial channels and transporters. A long-term goal is to understand further how CIC-2 activity can be regulated in functional epithelial tissue from transgenic mouse models of CF.

Much remains to be understood about CIC-2, and alternative chloride channels at large. CFRI's support of my individual contribution to this base of knowledge, as well as those of other investigators interested in CF related questions, is very much appreciated. ■

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## 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 that accompanies the Annual Report. 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 2001.
  - Please send me \_\_\_\_\_ invitations!
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Name	Relationship to CF Person	Date of Birth (for CF patients only)
Street		
City	State	Zip
Telephone		

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 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 31.

## 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 EMAIL ADDRESS AND WEB SITE

CFRI's email 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: <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 16 to adult
- Family support groups
- CF support groups for ages 13 to adult
- *CFRI News* newsletter
- 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. Supporters include:

- Executive Director, Program Services Manager, part-time Outreach Specialist and 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 Genentech, Inc., makers of Pulmozyme®, and PathoGenesis Corporation, makers of TOBI®, for their generous support of *CFRI News*.

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