

# CFRI news

34 Years of Research, Education & Support

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## VX Drug Trials Offer Hope

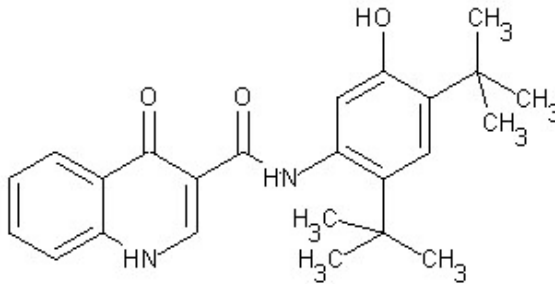
Potentiators and Correctors: Vertex Continues Exciting Clinical Trials With VX-770 and VX-809

By Siri Vaeth-Dunn

Cystic fibrosis is caused by genetic mutations that lead to either a malfunctioning or missing cystic fibrosis transmembrane conductance regulator (CFTR) protein on the cell surfaces. This creates an imbalance of salt and water in the epithelial cells that line every open surface of the body, including the lungs, digestive tract, pancreas, liver, and reproductive tract. In the airways, this imbalance causes the dangerous cycle of mucus plugging, infection and inflammation that characterizes CF. In exciting research pursued by Vertex Pharmaceuticals, in collaboration with Cystic Fibrosis Foundation Therapeutics, Inc. (CFFT), two new compounds, VX-770 and VX-809, have been developed to address both the malfunctioning and the missing CFTR protein. Initial clinical trials offer hope that the underlying defect causing the disease can be corrected.

VX-770 is a CFTR “potentiator.” Approximately 4% of CF patients carry the G551D mutation on at least one allele. This mutation creates a defective CFTR protein which can insert itself into the membrane of the cell, but which closes off the passage of chloride to the cell surface (In a recent *New Yorker* article, Dr. Jerome Groopman characterized this defect as a “rusty gate.”). Taken orally, VX-770 serves as a potentiator, to restore the normal function of the mutated protein and increase chloride ion transport through the defective CFTR protein.

Initial results, presented at the North American Cystic Fibrosis Conference last October, were very exciting, with 19 participants who received the drug in a placebo-controlled study demonstrating a



renewed ability to transport chloride, and experiencing an average 10% increase in lung function. Based on the promising results from the Phase 2a trial, Vertex will conduct three new clinical trials: one for people aged 12 and older with the G551D mutation on at least one allele; one for children aged 6 to 11 with this same mutation; and for the first time, a trial which

will evaluate the clinical activity of VX-770 for CF patients with the Delta F 508 mutation on both alleles.

VX-809 is a CFTR “corrector.” The majority of CF patients have the delta F508 mutation, which causes the CFTR protein to fold itself in such a way that it cannot insert itself into the cell membrane. VX-809 may help the defective CFTR protein to move to the proper location in the cell membrane, resulting in an increase in chloride transport across the cell surface

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### RESEARCH

Your CFRI Dollars at Work

### Summer Update

## Study May Unscramble CF Male Infertility Puzzle

By Bridget Barnes

Ninety-eight percent of men with cystic fibrosis are infertile. This is a major consideration for men with CF when planning a family. However, Carole Ober, a geneticist at the University of Chicago may have an answer. According to data presented this spring at the Sackler Colloquium on Evolution and Health in Medicine and published in *The Scientist*, Ober has identified a gene that boosts fertility in humans and may hold a new key to unlocking fertility in CF males.

For over twenty years, Ober has studied fertility in an insular group of people called the Hutterites who live in South Dakota and do not limit their family size, wait less than two years between births, and rarely marry outside their community, which reduces outside genetic input that can scramble studies of inheritance.

Ober’s most recent research has focused on the cystic fibrosis transmembrane conductance regulator (CFTR) gene which codes for a chloride ion channel and has a few known alleles that have been implicated in both cystic fibrosis and male fertility. One of these alleles is a single amino acid difference, either a valine or a methionine. Ober found from genotyping that a valine residue is significantly correlated with increased male birth rate. She also found

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## CFRI NEWS 2009 Summer Issue

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# Notes from our Executive Director

CFRI's office has been a hub of activity during the past several months, and I am very pleased to share an update of all that has been going on.

CFRI will soon have an extraordinary new website to reflect the many facets of our organization and serve as an inviting, useful resource to our community. We are extremely grateful to the Taproot Foundation, which provided an in-kind grant to make this possible. Join us: [www.CFRI.org](http://www.CFRI.org).

I want to welcome everyone to our 22<sup>nd</sup> National Cystic Fibrosis Family Education Conference, July 31 – August 2 at Sofitel San Francisco Bay. This year's theme is "The Power of New Possibilities: Growing Stronger and Living Longer with CF." Our list of speakers is on page four, and I am confident that you will find many topics of interest. We are able to offer this high quality event thanks to the generosity of our sponsors as well as our volunteers.

Our annual CFRI Teen and Adult Day Retreat will be held from August 2 – 9, at the beautiful Vallombrosa Center in Menlo Park. The Retreat provides a safe and welcoming environment for people who share common experiences with CF.

August 3<sup>rd</sup> brings CFRI's 25<sup>th</sup> Annual Golf Tournament at the spectacular Cinnabar Hills Golf Course in San Jose, where participants can enjoy



18 holes of golf, wine tasting and a silent auction. We appreciate our volunteers who organize this phenomenal event.

Four new post-doctoral fellowships have recently been approved! These researchers will pursue innovative studies that will increase our understanding of cystic fibrosis, and move us closer to a cure. The value of CFRI funded research cannot be understated. The recent Vertex trials are the result of research conducted many years ago, some of which was funded through CFRI.

We have heard the saying, "It takes a village to raise a child." In these challenging economic times, the same is true for an organization such as CFRI. Thanks to the generosity of our volunteers, corporate sponsors, foundation funders and individual donors, we are steadfast in our commitment to funding cutting edge research, while providing meaningful education and support services. Thank you for being a part of our CFRI village.

Warm Regards,

*Carroll Jenkins*

Carroll Jenkins,  
Executive Director  
(650) 404-9977 • [cjenkins@cfri.org](mailto:cjenkins@cfri.org)

## Of Note –

**July 31 – August 2, 2009**  
**CFRI 22nd National CF Family**  
**Education Conference**  
Sofitel San Francisco Bay

**August 2 – 9, 2009**  
**CFRI Teen & Adult Day Retreat**  
Vallombrosa Center, Menlo Park, CA

**Monday, August 3, 2009**  
**CFRI Golf Tournament**  
Cinnabar Hills Golf Course, San Jose, CA  
Call Scott Hoyt (408-323-7803)

## Send Us Your Video Clips and Photos!

For this year's National Family Education Conference, we are creating an exhibit of children and adults with CF engaging in activities such as: dancing, skateboarding, playing the drums, etc. If you would like to submit a photo (no larger than 5"x7" in hard copy or jpg format) or a video clip (no longer than 2 minutes in length on a DVD or flash drive), send your entry to CFRI, 2672 Bayshore Parkway, Suite 520, Mountain View, CA 94043 or [cfri@cfri.org](mailto:cfri@cfri.org). Please identify your video clips and photos with names, contact info, and the approximate dates of when they were taken. Photos and videos may be edited and cannot be returned. If you have questions, contact Mary at (650) 404-9975 or via e-mail: [cfri@cfri.org](mailto:cfri@cfri.org).

**JUNE 21 DEADLINE**

# Cystic Fibrosis and Lung Transplantation: The Rewards and Challenges

By Siri Vaeth-Dunn

Tremendous progress has been made in the treatment of cystic fibrosis and the predicted age of survival. However, for many the damage to the lungs becomes too significant, and transplant becomes the most viable option. While not a cure for CF, lung transplantation can extend the lives of those who would otherwise lose their battle with the disease.

Such was the case for Clare Webster, 39

years old, who had a double lung transplant in 2004. After years of declining lung function, her FEV1 dropped to near 20% of predicted and her doctor encouraged her to be listed for transplant. Clare says, "It's a hard decision to make. It may not be for everyone, but my realization was either to try and live, or die." Scott Petersen, 52, faced a similar situation.

A saxophonist who has performed around the world with many of the world's most notable jazz musicians, Scott was still performing with an FEV1 hovering near 30% of predicted. But in 2008, his health began to decline rapidly. "Without a transplant, I am sure that I would not have lived to see 2010," he says. He received a double lung transplant in January 2009.

In general, the criteria for inclusion on the national transplant wait list includes: severe lung disease with an FEV1 of less than 40% of predicted; a life expectancy (without transplant) of 1 to 2 years; decreased ability to function in daily life; increasing frequency of pulmonary exacerbations or occurrence of life threatening pulmonary complications; the ability to undergo a very major medical procedure;

and the ability to comply with post-transplant medications and a new life-long health regimen.

The decision to be added to the wait list for transplant involves medical assessment, familial and community support, psychological fortitude, and for many, spiritual guidance. Each individual must undergo a thorough review by the transplant center's team which generally includes a medical referral, several phases



Clare Webster, holding her son Jason, enjoying the day with her husband Justin. Clare had a double transplant in 2004 and earlier this year completed a half marathon.

of medical assessment, a psychosocial evaluation, and parent and family education. Some centers exclude transplants for those culturing *Burkholderia cepacia*, which infects 5% of CF patients, and dramatically decreases survival rates. While the evaluation is arduous, Scott says it didn't seem that hard. "During the process, you just become a CF warrior. You do what you need to do."

The length of time one waits for a transplant is determined by his or her Lung Allocation Score (LAS), a system put in place in 2005 by the United Network for Organ Sharing (UNOS). Prior to 2005, one's position on the waiting list was based on length of time waiting. Everyone started at the bottom, and worked their way up. With the new LAS system, each patient is evaluated and given a score between 0 and 100, based on disease severity and likelihood of survival. Those with the highest scores are placed at the top of the waiting list. Scores must be updated every six months. This system applies

only to those aged 12 years and older. The transplant list for children under 12 is still chronologically-based.

Statistics show that people with CF tend to do better post transplant than people with other lung diseases, and therefore they may score higher under the new LAS system. With this change in the evaluation process, people with CF are more proactive. Delays for them are often related to size match and blood type. Scott waited eleven months to receive his transplant. "My wait on the list was a long eight months, all the while on oxygen," says Clare. "It was an emotional roller coaster."

Lung transplantation is not a cure for cystic fibrosis. CF is caused by a mutation on chromosome 7, and its impact is systemic. The newly transplanted lungs will not have the CFTR mutation and will effectively transport salt and water, thus "curing" a person of severe lung disease. However, the CFTR mutation will continue to impact the pancreas, sinuses, liver, sweat glands and reproductive tract.

Individuals who needed enzymes before the surgery will still need them post-transplant. Pre-existing CF-related sinus and/or liver disease will remain. Those with cystic fibrosis related diabetes will have to work harder to control their sugars, due to the large doses of steroids that are necessary to prevent rejection. These immunosuppressive drugs will also decrease the body's ability to fight bacteria such as *Pseudomonas aeruginosa* and *Burkholderia cepacia* (*B. cepacia*), which can then infect the new lungs.

"You trade lung disease for immunosuppression management, says Scott. "I take more pills now, about 30 a day," says Clare,



(Continued on page 9)



**The Power of New Possibilities:  
Growing Stronger  
and Living Longer  
With Cystic Fibrosis**



**22<sup>nd</sup> National Cystic Fibrosis Family Education Conference  
Sofitel San Francisco Bay, Redwood City, CA  
July 31 - August 2, 2009**

**Topics and Speakers Include:**

**“A Swedish Care Model: What We Can Learn”**

Birgitta Strandvik, MD, PhD, Göteborg University, Göteborg, Sweden

**“Cystic Fibrosis: The Future Begins Now”**

Jeff Wagener, MD, University of Colorado School of Medicine, Denver, CO

**“Clinical Research Update: Keeping Up with New CF Treatments”**

Mark Dovey, MD, St. Christopher Hospital, Philadelphia, PA

**“New Findings to Address Tough Psychosocial Challenges in CF”**

Alexandra Quittner, PhD, University of Miami, FL

**“Sex Hormones: How They Play A Role in CF Health”**

Marcia Katz, MD, Baylor College of Medicine, Houston, TX

**“Drug Therapy in CF: Treating Today’s Problems While Investing in the Future”**

Robert Kuhn, PharmD, University of Kentucky College of Pharmacy, Lexington, KY

**“Transition of Care in Cystic Fibrosis”**

K. Randall Young, Jr., MD, University of Alabama Birmingham Pulmonary, Allergy and Critical Care Medicine, Birmingham, AL

**“Make the Medical System Work for You”**

Julie Biller, MD, Medical College of Wisconsin, Milwaukee, WI

**“A Sister’s Story: My Path to Becoming a CF Nurse”**

Veronica (Ronni) Wetmore, RN, MS, Adult Cystic Fibrosis Center, Jacksonville, FL

**Continuing Education Units (CEU’s) are now available for coordinators, nurse practitioners, nurses, social workers, dieticians, respiratory therapists, other healthcare providers and students. For CEU information and forms, visit [www.cfri.org](http://www.cfri.org) or call (650) 404-9975.**

★*Early Bird Registration on or before 7/1/09: \$150 per person*

★*Regular Registration after 7/1/09: \$175 per person*

★*All meals included*

★*Precautions to avoid cross-infection are rigorously followed by CFRI and hotel staff*

★*Scholarships are available for eligible applicants*

★*Significant hotel room discounts via CFRI’s web site: [www.cfri.org](http://www.cfri.org)*

**For more information, contact CFRI at (650) 404-9975 or [cfri@cfri.org](mailto:cfri@cfri.org)**

# TRANSITIONS – The Changing CF World

By Robin Modlin

Recent improvements in the treatment of CF have created a period of long hoped for changes, with the median age of survival now at 37 years plus. With over half the population over 18 years, many CF Centers are having to adjust to the growing number of young adults. This change has led to a host of new issues, and has inspired a future Transitions series in *CFRI News*. Through your responses to our inquiry about your adult care, we want to look closer at this change and its impact.

In an article titled “Transition and Transfer of Patients Who Have Cystic Fibrosis to Adult Care” (2007) H. Worth Parker, MD, reported the following statistics demonstrating that developments in research and therapy have resulted in improvements in survival.

According to Dr. Parker, there has been a dramatic increase in the number of adults with cystic fibrosis. The number of adults with CF aged 18 years or older has grown more than 400% since the early 1970s. Approximately 43% of all individuals with cystic fibrosis are now 18 years or older. In the next decade, 50% of all individuals with cystic fibrosis will be older than 18 years, and CF will become a condition seen more commonly in adult medicine than in pediatrics. The majority of the adults with CF are between the ages of 18 and 30 years but, there are a growing number of older adults, with 25% being

between the ages of 30 and 39 years, and 10% being older than 40 years. In 2006 there were 10 known individuals older than 70 years in the U.S. Cystic Fibrosis Foundation National Data Registry.

Even with this increase in longevity the disease takes its toll as a chronic and life threatening condition. Different rates of decline in lung function result in a wide spectrum of lung disease severity in adults with cystic fibrosis, with approximately 25% having severe lung disease, 40% with a more moderate disease, and 35% having mild lung disease or normal lung function.

Cystic fibrosis is a systemic disease that impacts the entire body. With increased longevity come other complications beyond reduced lung function, including diabetes, liver disease and osteoporosis. In addition, adults with CF often have specialized needs in terms of reproductive issues.

About 150 individuals with cystic fibrosis undergo lung transplantation each year, which constitutes approximately 1.5% of the adult cystic fibrosis population annually. However there is still only a 5-year survival rate of 50% after this procedure and little evidence that the advent of lung transplantation has affected overall median predicted survival.

Concerns about adult care have led many to push for the development of a care

model comparable to that of pediatrics, where so many strides have been made in coordination of services. Inspiring Internists and Adult Pulmonologists to pursue cystic fibrosis care has been a challenge, but is vital to continue the excellent care that the pediatric centers have provided.

Among the greatest issues for adults with CF are problems with adherence to regimens due to full adult lives, not having enough time, being underinsured and recognizing symptoms of depression and chronic grief. Clearly there is work to be done to improve care for our growing CF adult population.

## We'd like to hear from you.

These improved statistics are testament to what we've been striving for and give us hope, but they also represent challenges and new considerations. We want to hear from you and how you feel this transition to adult care is going and how it has affected care.

What has worked and not worked in your center and your life? What recommendations do you have to the patient or the medical communities about getting older with CF? Please tell us, so together we can help each other as we age and break the odds of this disease. Send your comments to: [DBatchelder@CFRI.org](mailto:DBatchelder@CFRI.org) or mail them to our office. We will present some of your responses in a future issue of *CFRI News*.

## In Retreat, Board Looks Forward!

By Bridget Barnes

At an all-day retreat in February, CFRI's Board of Directors got together to review and reaffirm the organization's short and long-term goals. A variety of topics were covered in depth, including the key issues of organizational and financial sustainability during this year's economic downturn, expanding outreach and advocacy, and governance of our research programs.

As with non-profits everywhere, the Board looked at ways for CFRI to further its commitment to being a lean and efficient

organization as well as ensuring ongoing excellence and continuity in the governance and oversight of the research programs it supports. As for expanding CFRI's "niche" as an advocate for the CF community, Board members agreed to expand new outreach possibilities over time for the Spanish speaking population.

The Board of Directors is made up of parents of children with cystic fibrosis, an adult with CF, and others who are friends and advocates for the CF community. This annual retreat, with its blend of focus and friendship, provided Directors with a renewed commit-



Board President Bill Hult and Vice-President Jessica Martens planning CFRI goals earlier this year.

ment and enthusiasm about overseeing and helping to accomplish the goals ahead for CFRI. At the close of the day, the Board unanimously agreed how fortunate CFRI is to have the committed and visionary leadership of Executive Director Carroll Jenkins.

# Make a Wish Foundation: A Gift to the CF Community

By Siri Vaeth-Dunn

Since 1980, the Make-A-Wish Foundation® has granted over 178,000 wishes to children with life-threatening diseases, many of whom are children and teens with cystic fibrosis. The mission of the Make-A-Wish Foundation is simple. *“We grant the wishes of children with life-threatening medical conditions to enrich the human experience with hope, strength and joy.”*

For children and teens struggling with the daily tedium of treatments, frequent hospitalizations and the stress of a progressive disease, the granting of a wish provides a much needed relief from the norm, and often, the realization of a dream.

An example of this is Devin, 17, who has had numerous health challenges related to his cystic fibrosis. Last fall, Devin was granted his wish to visit Stonehenge in England. His Make-A-Wish volunteers learned that he was an avid history buff, and his ten-day trip to England included other historical sites. *“Going to Stonehenge was really fun,”* says Devin, *“I’ve always loved history and so I wanted to see one of the oldest and most mysterious monuments in the world. But I didn’t expect London to be so awesome. The Churchill War Rooms, the British Museum and the Tower of London --- well, I could have spent a week in each one!”* His parents, Kathleen and Scott, as well as his brother Brendan accompanied him. It was a life-changing journey for all. As Kathleen shared, *“Devin and our entire family were going through a hard time. We came back from England with a different mindset. It was the perfect trip for Devin and the most incredible trip of our lives.”*

For families who have children with CF between the ages of two and a half and



*Brendan, Devin, Kathleen and Scott photographed in front of Stonehenge, England.*

eighteen years, starting the wish process is simple. There are no income requirements or income limitations. Referrals can be made from any of three sources: the child, his or her parent or guardian, or a member of the CF medical care team (usually the social worker). Once a referral has been made, the process moves very quickly. After a child has been determined to be eligible, a team of volunteers visits the family to learn more about the child and his/her wish. Of utmost importance to the volunteers is that they hear directly from the child about his or her dreams. The information is then presented to Foundation staff, which honors the needs and wishes of the child above all else.



My own 14-year-old daughter, Tess, was granted her wish last winter. A singer-songwriter, she recorded her first album in a North Hollywood recording studio, thanks to the Greater Bay Area chapter. As Tess puts it, *“I will always be grateful to everyone at the Make-A-Wish Foundation. Making my album was more than a wish – it was a dream. I had a blast, and it has helped me reach the next milestone in my music career.”*

The Greater Bay Area chapter grants about three to five percent of wishes each year to

children with cystic fibrosis. Some parents hesitate to utilize the services of the Make-A-Wish Foundation, due to the mistaken perception that it is for kids who are perceived as “terminal.” This is not the case. Any child with a life-threatening medical condition is eligible.

When reflecting upon the best time for your child to enjoy a wish, Kathleen recommends, *“Don’t wait too long. It is best for your child to enjoy the wish when he or she is in optimal health. There might be some things that your child won’t be able to do if you wait too long.”*

Lisa McIntire, Program Associate for the Greater Bay Area Make-A-Wish Foundation shares, *“Working here and being able to plan wishes for inspiring kids is unreal! It’s an honor to be a part of the magic that happens for our kids who are going through so much.”* Whether through shopping sprees, room makeovers, meetings with celebrities, journeys to far away places, or the realization of musical dreams, The Make-A-Wish Foundation has made it possible for thousands of children with CF to have a meaningful respite from the tedium of their daily routines and health challenges and to experience a wish come true.

*To find out more about the Make-A-Wish Foundation, or to locate a local chapter, go to [www.wish.org](http://www.wish.org).*

# What's New?

## The latest CF research about Neutrophils on the Internet

By Laura Tillman

**N**eutrophils are the most common type of white blood cell, comprising between 50-70% of all white blood cells. Neutrophils are present in the bloodstream until signaled by chemical cues in the body to attack infection, and are the first immune cells to arrive at a site of infection, reaching the site within an hour. Before ingesting invasive bacteria, neutrophils can release a net of fibers called a neutrophil extracellular trap (NET), which serves to trap and kill microbes outside the cell. When neutrophils ingest microbes, they release a number of proteins in primary, secondary, and tertiary granules that help kill the bacteria. They also release superoxide, which becomes converted into hypochlorous acid, or chlorine bleach, which is theorized to play a part in killing microbes as well. CF patients experience progressive airway destruction caused in part by chronic neutrophilic inflammation. The following links will take you to abstracts for more information:

**Activation of critical, host-induced, metabolic and stress pathways marks neutrophil entry into cystic fibrosis lungs.** Megha Makam, et al. *PNAS*. April 7, 2009, vol. 106, no. 14 5779-5783

<http://tinyurl.com/d41689>

**Summary:** The data demonstrates there is an early and sustained activation of host-responsive metabolic and stress pathways upon neutrophil entry into CF airways, thus suggesting potential targets for therapeutic modulation.

**Insights Give New Hope Against Cystic Fibrosis.** Stanford University, news release, March 16, 2009

<http://tinyurl.com/cqk3fe>

**Summary:** It was long believed that neutrophils attacked and destroyed bacteria that became trapped in the excess mucus that collects in the lungs of CF patients. Contrary to this belief, research shows that neutrophils quickly die in the lungs,

releasing tissue-destroying enzymes as they expire. As a result, the neutrophils in CF patients may release large amounts of an enzyme called human neutrophil elastase, which destroys the elastic fiber of the lungs. In healthy people, neutrophils never release this enzyme into nearby tissues.

**Neutrophils in cystic fibrosis.** D G Downey, et al. *Thorax*, 2009; 64:81-88

<http://thorax.bmj.com/cgi/content/abstract/64/1/81>

**Summary:** This review of neutrophils in cystic fibrosis describes the cellular mechanisms involved in their migration into the airways and their role in bacterial phagocytosis, which is the destruction of foreign matter by white blood cells which serves as an important bodily defense mechanism against infection. The authors discuss the inflammatory process and its resolution and ultimately how neutrophil function can be modulated.

**Lung inflammation: Disarming neutrophils in cystic fibrosis.** Olive Leavy. *Nature Reviews Immunology* 2008, Vol 8; Number 1, page 8

<http://www.nature.com/nri/journal/v8/n1/full/nri2244.html>

**Summary:** This article proposes that in the lungs of patients with CF the chemokine interleukin-8, which is found at the site of infection or inflammation, continuously recruits neutrophils to the lung. There CXC-chemokine receptor 1 is immediately cleaved from the neutrophil's cell surface by free elastase, thereby disabling the antibacterial function of these cells. This cleavage begins a chemical process initiating a vicious cycle in which interleukin-8 releases chemokine receptor 1 which, in turn, produces more of the chemokine interleukin-8 and increases airway inflammation.

**Editor's note:** Some of these URLs do not work via "Google". Enter directly into your URL browser.

## Sodium Channel Blocker Shows Promise

**C**ystic fibrosis patients may benefit from a new therapy that increases airway hydration, preventing the buildup of mucus which is a key factor in the disease, according to researchers. "Our results suggest we have identified a new agent that acts directly on a specific pathway, which is involved in the development of CF," said lead author Andrew Hirsh, Ph.D., senior director of drug discovery and preclinical development for Parion Sciences.

In individuals with CF, the hydration level of the airway is altered, impacting their ability to effectively clear mucus. One of the mechanisms causing this involves the body's natural homeostasis of sodium which, when absorbed too rapidly from the surface of the airway, subsequently causes moisture to be absorbed too quickly. The new aerosol-based therapy uses a specific epithelial sodium channel-blocking agent called GS-9411 which prevents sodium from being absorbed across the airway. The resulting increase in moisture allows individuals to more effectively clear mucus and infectious agents.

During the study, researchers applied GS-9411 to airway surface cells grown in the laboratory and assessed the potency and reversibility of the drug. Results of the study indicated GS-9411 allowed the cells to retain liquid for more than eight hours. Concurrent animal studies revealed that the agent enhanced mucus clearance for more than four hours.

The results offer new hope for CF patients. Currently, GS-9411 is in Phase I clinical trials being conducted by Parion's development partner, Gilead Sciences, Inc. The clinical phase of the drug development cycle will enable researchers to refine the treatment for eventual distribution.

*Adapted from materials provided by American Thoracic Society, via EurekAlert!, a service of AAAS.*

## OUR FOUNDERS George and Anne Graham

By Ann Robinson

George and Anne Graham were living in Aberdeen, Scotland, when their son Douglas was born in 1954. They soon realized that he was sickly, and began taking him to doctors who were initially unable to discover why he was so ill. They finally found a doctor who diagnosed Douglas with cystic fibrosis. Their other son, Ron, did not have CF.

Because Douglas was not doing well in Scotland, George and Anne decided to immigrate to the United States where better medical care was available. George arrived first to secure a job then went back to Scotland to move his family to California. The Graham family settled in their new home near Palo Alto in 1964 where Douglas could be treated at Children's Hospital at Stanford.

Douglas' treatments included taking antibiotics, sleeping in a mist tent and taking enzymes to help him digest food. Douglas' infections were so numerous that he spent weeks in the hospital getting I.V. antibiotics and chest physiotherapy. Even though



he was receiving the best care possible, Douglas died in 1974 at the age of 19 years. The family was devastated.

George and Anne became founding members of CFRI after meeting Marilyn Holmes, an active parent in the Cystic Fibrosis Foundation's peninsula chapter. George and Anne began attending meetings and raising money for the local CFF where George served as Treasurer and Anne served as Secretary. Members of the peninsula chapter wanted to have more

say in how their research dollars were allocated and the seeds of CFRI were sown. When CFRI was incorporated, the Grahams were founding members. Anne has sent Mother's Day Tea invitations in memory of Douglas every year since 1975. George was active in the Scottish Highland Games which raised money for various charities including CFRI.

The Grahams were pleased to learn that children and adults living today with cystic fibrosis are doing so well. They never thought that new treatments could extend the lives of CF adults so they could graduate from college, have families and careers.

*Sadly, George had a heart attack and died in November, 2008. Anne continues to live in San Jose and keeps in touch with other CFRI members. We thank you, Anne and George; your dedication to helping everyone with cystic fibrosis is greatly appreciated. You have contributed to the happiness and well being of many.*

## The Heart of a Volunteer: Terry Nelson

By Bridget Barnes

Terry Nelson is a devoted, long-time champion of CFRI who enthusiastically volunteers in a variety of capacities, some visible, some not. She has given of her time and of herself literally for over 21 years. "I started volunteering at CF camp when my sister couldn't find a "thumper" for my nephew. We talked about the best way to handle this and decided I would go to camp with my nephew and be his "thumper," while she watched my three young children." I had so much fun I went for 15 years straight. I was like a fish in water." Because of her background as a teacher and someone who naturally brims with creativity, Terry taught oil painting, paper mâché and introduced a variety of new "theme-based" activities to the campers every year. She was also part of the leadership team. When asked to reveal her favorite memory, she quickly replied, "the cherry drop, but

you'll have to ask someone from camp about that!"

Once her camp days were over, Nelson's creative juices for another idea started flowing. She founded the Hospitality Room at the CFRI Annual Conference. "I wanted to set up a room with tables and chairs where people could visit, talk and unwind," Terry explains. "I wanted a way of bringing camp to the conference." She turned her idea into a reality and for the past several years she has transformed a conference room into a welcoming space offering individuals and families a place to connect with each other, bid on silent auction items, read a variety of authors' poetry from "The Breathing Room" and view a selection of



creative endeavors made by people living with cystic fibrosis. Perhaps most important of all, the Hospitality Room provides a quiet place to reflect on the advances and challenges of this multi-faceted disease.

*(Continued on page 11)*

**Lung Transplantation** (continued from page three)

who was recently put back on nebulized medications for a lung infection. “It’s a small price to pay to stay healthy, but it’s still a bummer to be on nebs again!”

The transplant recovery process varies widely. Clare struggled for three months, as her newly transplanted lungs took time to fully expand. The physical and emotional challenges can take their toll, and depression for post-transplant patients is common. In addition to “incredible pain,” Scott experienced “survivor guilt.” While understanding intellectually that the lungs he received were the best match for him, it posed an emotional challenge, knowing that others had waited longer for transplant. Having a strong support system is critical. Says Clare “It’s so important...it’s a big life change, and it takes getting used to.” Scott’s circle of friends helped arrange housing close to the hospital for Scott and his wife, and took weekly caregiving shifts.

Survival rates for lung transplant have improved over the past two decades. According to the 2007 Transplant Registry of The International Society for Heart and Lung Transplantation, 87% of CF patients receiving lung transplants were alive after one year, and 68% were alive after three years. Still, there are risks to this procedure, and the life expectancy post-transplant declines over the years. According to the Cystic Fibrosis Foundation, approximately 50% of CF transplant patients are living after five years.

Would survival rates improve if the wait for transplant was shorter? According to the Cystic Fibrosis Foundation registry, approximately 120 to 150 people with cystic fibrosis receive lung transplants each year. Unfortunately, more people are waiting for transplants than can receive them. In 2003, 126 people with CF had lung transplants, while 368 people were assessed and accepted for the procedure. There is a shortage of available organs, and the perilous wait for much needed lungs might be shortened if more people would identify themselves as donors. This is especially the case for pediatric donors.



Scott Petersen received a lung transplant earlier this year and is already playing “gigs” with his saxophone. —Photo courtesy of Michelle Compton

In a survey conducted for Donate Life America in February 2009, it was found that 90% of American adults support organ donation, but only 38% of licensed drivers have registered as donors. In the meantime, every 12 minutes another name is added to the national organ transplant waiting list (which includes all organ transplants, not solely those related to CF), and an average of 18 people die each day waiting for transplants.

A team of researchers in Toronto has recently developed a system to maintain donated lungs outside the body, using a ventilator, pump and filters. This technique gives surgeons a longer window of time to assess and repair any damage to the lungs before transplanting them. While still in the clinical trial stage, seven patients have received lungs using this technique, and all are doing well. This exciting breakthrough has the potential to double the number of acceptable donor lungs available for transplant.

“The number of people awaiting organ transplants has climbed to more than 100,000” said Donate Life America Chair, Sara Pace Jones. “It is more important than ever for those who support donation to legally document those wishes – and registering through the DMV or state donor registries are the most important ways to do that.”

To register in your state, visit Donate Life America, a not-for-profit alliance of national organizations and state teams across the United States that advocates for

organ donor registration: [www.donatelife.net](http://www.donatelife.net).

For those considering transplant, both Scott and Clare have some advice. “Talk with others who have gone through it. Everyone’s experience is different, so it’s good to get other people’s perspective,” advises Clare. Says Scott, “It’s a big decision, but when the time comes, just go with it.”

More advice comes from Dr. Carol Conrad, Director of the Pediatric Lung and Heart-Lung Transplant Program at Lucile Packard Children’s Hospital at Stanford. “Every child and adolescent I have transplanted has said they achieved an excellent quality of life after their transplant and would go through the same thing again, even if it was for a month, six months, or six years. I advise considering quality of life rather than length of life attained.”

A successful lung transplant opens the door to previously unimaginable experiences for individuals who had spent years struggling to breathe. Clare became a mother in 2007, and recently completed a half marathon. Scott is back to a full schedule of musical gigs, and looks forward to traveling again. When asked how his life has changed, Scott’s initial reply was “At the most obvious level, I can breathe. It is as simple as that. I can inhale and exhale deeply, and do it over and over.” He adds, “I feel so much better than I ever have. It is a miracle.” Through it all, both have a deep appreciation for their donors, and their donors’ families. Says Clare, “Every day is precious, and every day I think of my donor and her family. If it weren’t for them, I wouldn’t be here.” For those whose lung disease has progressed beyond repair, lung transplantation offers the opportunity to breathe deeply and live fully.



# Keeping Hope Alive During Tough Times

By Lisa C. Greene



**I** pull into the carpool lane to pick my kids up from school and am yet again touched by the sight of a service dog with a bright pink lunch pail hanging from her mouth as she trots alongside a young girl in a wheelchair.

And then there is the computer-generated voice that says “Amen!” at just the right moment during church. I am inspired by the young family in the front row as they minister to the physical and spiritual needs of their disabled son who is barely visible amidst the medical equipment that surrounds him. It would be so easy not to go to church but there they are, week after week. How do these families and others like them keep their hope alive in the face of such daunting challenges?

I am no stranger to hope – or hopelessness for that matter. Both of our children were born with cystic fibrosis. The median life-expectancy is currently about 37 years of age and rapidly climbing due to great advances in medical technology. There is great *hope* for the future of those living with cystic fibrosis and I am generally optimistic about my children’s future. But I know of a beautiful young lady who recently died from the disease at the tender age of twelve. And the many pills, medical treatments and hospital visits that my two children endure serve as a bleak reminder of our race against time with this progressive disease.

Sometimes the icy fingers of hopelessness hide in the shadows around my heart, waiting for a beat to falter so they can enter. But I don’t let hopelessness sneak in. I can’t. My children depend on me. Not only am I their caregiver, but also their role model. They will pick up on my cues as they learn to navigate their own way through a lifetime filled with the trials that their illness will impose upon them. If I model hope, they will be hopeful. If I

model despair, then they will be hopeless. So, how do we keep our hope alive even when we don’t feel very hopeful?

Throughout my CF journey I have had many teachers encourage me, inspire me and lead the way. And I have noticed they all have one thing in common: gratitude. A little research on gratitude turned up some interesting things. It is not clear why some people are naturally more positive than others. Perhaps it’s genetic. But *it is* clear that purposely focusing on what we are thankful for, instead of what we are upset about, will help us feel more optimistic about the future as well as more loving, forgiving, joyful, healthy and, of course, hopeful in the present.

Anabel Stenzel, co-author with twin sister Isabel of the book “The Power of Two,” says: “Because of my CF, I have received many gifts: resilience, closeness with loved ones, the amazing people one meets in the CF community, and the maturity and depth that living with a chronic and life-threatening disease can bring. Despite living with progressive lung disease that required me to have a lung transplant at age 28, I have learned, loved, seen, heard, eaten, walked, talked, touched, thanked, hoped and dreamed in my lifetime. And I have no regrets.” Ana and Isa are living examples of counting our blessings one by one. The good news is that an attitude of gratitude is a choice. Anyone can be more thankful with a little awareness and effort. Here’s how:

**1. Pay attention to your thoughts.** Make the decision to replace negative thoughts with positive ones. For example, say “I am thankful for having medical insurance” rather than “What a hassle this insurance company is!” Repeat over and over as necessary!

**2. Stay in the present.** Focusing on the challenges of the past or on what might happen in the

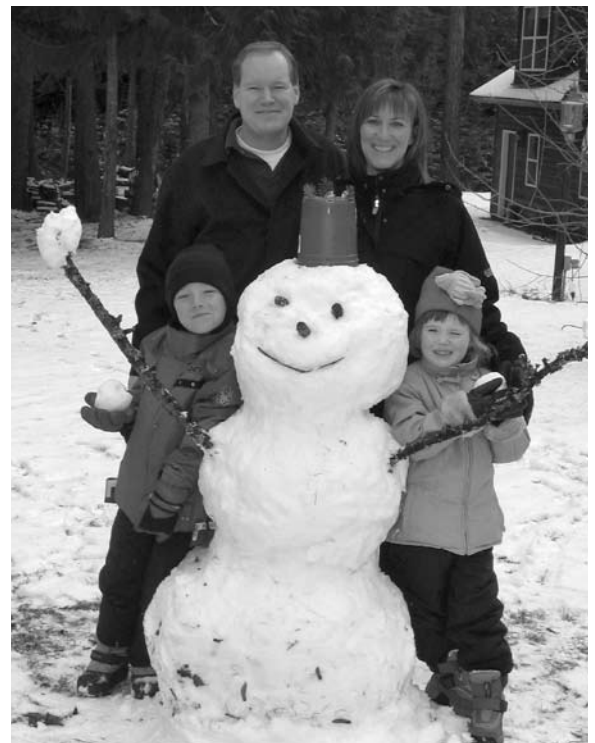
future can rob your joy today. What are you grateful for *right now*? Clean clothes? A loving family? Food on the table? Start with the simple things in life we often take for granted.

**3. Take five minutes each day to meditate** on one or two things you are thankful for. What blessings did your day hold? A meaningful moment with a loved one? A beautiful sunset? A word of encouragement given or received? A job well done?

**4. Begin a “gratitude journal.”** Write down one different thing that you are thankful for each day for three weeks. Notice how much happier you feel. Then, keep going.

**5. Share your blessings each day** with your family -- especially your children. Ask them what their blessings were, too. An attitude of gratitude is contagious!

Sometimes it can be difficult to muster up the will to count our blessings. Here are some tips for keeping our hope alive during those especially hard times:



Lisa Greene, her husband and their two children, both of whom have CF.

**Keep your expectations reasonable.**

Hoping for a cure can keep us going but can be devastating when it doesn't happen. While miracles can happen, it's also important to stay grounded in reality. Having faith with reasonable expectations helps us stay balanced.

**Take the time to grieve:** Bad things happen. And when they do, we understandably respond with shock, anger and despair. The way to acceptance is through these emotions by allowing ourselves the time and space to grieve. The key is to find a balance and not get stuck. Elise Free, mom of a two-year-old girl with CF nicknamed Froggy explains, "There are levels to my own grief. One day I feel elation that Froggy lives in a time of progressive medicine, and in the next second, sorrow that we were so unlucky to be a carrier of a defective gene, passing on something dreadful to our child. And then there is the warm wash of gratefulness leading me to appreciate every moment with a monumental love powered by the knowledge that life is fragile and our time together is measured in breaths. We have been given an opportunity: to see life in a dark light, a constant balance of pain and peace, hope and devastation, sorrow and joy. And maybe that is the purpose of life, to find the balance, to accept the reality, but live in the hope."

**Faith-filled folks have more hope!**

When times are really tough, we need something to hold on to – some sort of anchor in the storm. We can find comfort in knowing that there is something bigger than us in all of this; that there is some meaning and purpose for it all – even if we don't know what that might be. So, how do we keep our hope alive? Marla, mom of sweet baby Emma (who has cystic fibrosis), sums this up for us perfectly: "I look at my daughter's face. Seeing her beautiful smile gives me hope."

*Originally published in PARENTGUIDE, December 2008; reprinted with permission.* Lisa C. Greene is the mom of two kids with cystic fibrosis, a parent educator and co-author with Foster Cline, MD, of the book "Parenting Children with Health Issues: Essential Tools, Tips and Tactics for Raising Kids with Chronic Illness, Medical Conditions and Other Special Needs." Visit <http://www.ParentingChildrenWithHealthIssues.com>.

## Sign Up Today

### For Our 25<sup>th</sup> Annual CFRI Golf Tournament

Golf enthusiasts! Get ready to enter a friendly game of competition and raise money for CFRI at the same time. Join the 100 or so golfers who cheerfully descend upon Cinnabar Hills Golf Club in San Jose on the first Monday in August each year.

In addition to 18 holes of golf, there will be tee prizes, wine and cheese tasting, silent and live auction items including fantastic sports memorabilia, and a celebration banquet. You may attend as a non-golfer.



For more information, contact Scott Hoyt, General Manager at Cinnabar Hills at (408) 323-7803 or [shoyt@cinnabarhills.com](mailto:shoyt@cinnabarhills.com).



More than 200 people attend our CFRI Golf Tournament banquet each year. Here a golfer tees off on one of the beautiful eighteen holes played during the day at Cinnabar Hills Golf Club in San Jose.



Golfers and non-golfers alike participate in our silent auction and raffle prize drawings. Here one "winner" comes from the dining area to accept her prize. Scott Hoyt is CFRI's Treasurer and serves on our Board of Directors. Scott is a golf professional and general manager at Cinnabar Hills. He and his wife have a teenage daughter with CF.

**Terry Nelson** (continued from page 8)

In addition to being the "hostess with the mostest" Terry also serves on the CFRI Conference Committee. She is responsible for all the posters, flyers and graphics displayed at the Conference, lends a hand on the Retreat, is a Mothers' Day Tea sender, and is the primary "go to" person for all things creative. "You want to get things done, ask a busy person," she laughs. Kidding aside, Terry revealed why she volunteers so robustly for CFRI, "It's my commitment to my family and how I deal with the disease. I deal with things

face-to-face. I get a lot of joy from people and those years at camp were so much fun – they were my life, they were my summer." Now as she begins brainstorming about this year's "sports bar theme" for the Hospitality Room, Terry Nelson lightheartedly reaffirms her commitment to her family and to CFRI in saying "that's me, I'm a permanent fixture." And, we are so grateful!

*Terry Nelson's oil painting also graces our Mothers' Day Tea invitation this year (see page 12).*

# At CFRI It's "Tea Time" All Year Round!

By Bridget Barnes

Although Mother's Day has come and gone, the spirit of the Mothers' Day Tea lives on throughout the year at CFRI. Taking a moment to pause and reflect on a loved one living with cystic fibrosis is a rewarding thing to do any time of the year, as is making a donation to CFRI in their honor. If you have not yet participated in the Tea this year, it's not too late!



Peggy Jones and Maxine Eggerth admire the oil painting created by Terry Nelson while attending the 2009 Mothers' Day kick-off event.

Our goal is to raise \$250,000 to support first-rate research and programs instrumental in improving and expanding the lives of people with CF. Your donations, which mean even more this year in light of the current economic climate, continue to be crucial in funding CFRI's New Horizons Research Program and the Elizabeth Nash Memorial Fellowship, as well as educational programs

such as the annual Education Conference and Teen/Adult Day Retreat, and outreach materials including *CF in the Classroom*, the *CF Website Guide* and *CFRI News*.

We received our first donation of 2009 in March. Since then, 212 tea senders have sent out invitations and over 1,700+

people have responded from over thirty-one states.

If you have not had the satisfaction this year of participating in our largest fundraising event, you can still do so in either of the following ways: mail your donation to CFRI, 2672 Bayshore Parkway, Suite 520, Mountain View, CA 94043, or visit our newly designed website at [www.cfri.org](http://www.cfri.org) and click on the "donate now" link.

Thank you for your support. It helps so many children and young adults living with cystic fibrosis.

***A Special Thanks to Our Sponsors:***  
**R.C. BIGELOW, Inc.**  
**Digestive Care, Inc.**

## Mothers' Day Tea 2009

Just clip and send us this order form, call us at 650-404-9975 or email us: [cfri@cfri.org](mailto:cfri@cfri.org). We will provide you with invitations and envelopes, information about CF, and teabags, generously donated by R.C. Bigelow®

Send these to your family and friends, and join others across the country for a satisfying cup of tea. It's that easy. With your support, we fund important research and provide current information to those with cystic fibrosis and their families.



Valerie Schneider won the beautiful Wedgwood teapot donated by longtime CFRI volunteer Pat Flynn.

### Mothers' Day Tea 2009!



**Yes**, I would like to send Tea invitations. Please send \_\_\_\_\_ (number of invitations)

\_\_\_\_\_  
 Your Name (please PRINT)

\_\_\_\_\_  
 (Area Code) Telephone Number

\_\_\_\_\_  
 PO Box/Street Address

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

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

\_\_\_\_\_  
 Relationship to CF

# CYSTIC FIBROSIS TEEN & ADULT DAY RETREAT THE GREAT OUTDOORS: A BREATH OF FRESH AIR!



Last year's attendees pose in costume.

Need support in dealing with the challenges of CF? Looking for some guidance in the CF journey? Come to the CFRI Retreat! This is an amazing opportunity to connect with others with CF in a fun and SAFE atmosphere.

## SAVE THE DATE! AUGUST 2 (SUNDAY) TO AUGUST 9 (SUNDAY), 2009

We would like to invite you to an exciting summer event; the annual CYSTIC FIBROSIS TEEN & ADULT DAY RETREAT sponsored by Cystic Fibrosis Research, Inc., which will take place at the Vallombrosa Center in Menlo Park, California.

This retreat is a place for hope and healing as we learn more about CF and each other. The retreat features educational workshops and support groups covering medical issues, relationships, and coping. We also have lots of fun events including arts and crafts, sports, talent shows and offsite activities.

The CFRI Day Retreat welcomes teens and adults 15 years and older with CF, their spouses, family members and friends. However, due to cross infection concerns, all people with CF are required to obtain a sputum culture before the start of the retreat. People who have ever cultured *Burkholderia cepacia*, cultured Methicillin-resistant *Staphylococcus aureus* (MRSA) within the past 2 years, or are currently resistant to all antibiotics will not be allowed to attend. Participants must comply with cross infection behavioral precautions.

*“Retreat has been a transforming experience in my life. I have learned so much about how to take care of myself, and it inspires me to keep fighting. The friends I have made here understand my life in a way no one else I know does.”*  
—adult with CF, age 27

*“CFRI Retreat is such an important part of my life. It motivates me to take care of myself, to live, love and cherish those around me. The friends I met there are as close as family.”*  
—adult with CF, age 36

*“ This was my first year... what I did experience was possibly life-altering... the whole week was such an incredibly therapeutic experience for me and my girlfriend. The hardest part was leaving.”*  
—man with CF, age 31

Join us for a week of camaraderie, support, learning, and love. You are guaranteed to leave touched by this fulfilling experience, having spent the the days enriched by our community, sense of humor, depth and creativity. We hope to see you there!



For more information, please contact Jessica Martens, Retreat Chair, at [jemartens@earthlink.net](mailto:jemartens@earthlink.net) or call 650-404-9445, email [cfri@cfri.org](mailto:cfri@cfri.org) or visit [www.cfri.org](http://www.cfri.org)

# In Honor of

(Contributions listed were received from  
February 1, 2009 to April 30, 2009)

The Adelman Family  
The Batchelder Family  
The Pierce Family  
The Westley Family  
Claire Alexander  
Gianna Rose Altano  
Sadie Anderson  
Jessica Arvidson  
Rebecca J. Atkins  
Haleigh Baker  
Jaimee Baker  
Mat Baptiste  
Robin Baptiste  
Lucy Barnes  
Joe Batchelder  
Makinnon Baugh  
Marin Baugh  
Mikele Baugh  
Rebecca Boyer  
Brian Burks  
Ryan Coelho  
Lauren Colonna  
Barbara Curry  
Maxine Eggerth  
Elyse Elconin Goldberg  
Danny Ellett  
Hayden Ellett  
Alanah Fink  
Jarrod Fischer  
Casey Flaherty  
Ryan Foster  
Scott Foulger  
Jacob Fraker

## *"In Honor of" lists names of living persons in whose honor a donation has been made.*

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.

Mail your contribution with the name, address, and the occasion for the person you are honoring to:  
CFRI, 2672 Bayshore Parkway,  
Suite 520, Mountain View, CA 94043.

Joseph Fraker  
Emily Fredrick  
Jessica Fredrick  
Tara Goodearly  
Emily Gorsky  
Barbara Greenberg-Harwood  
Jacob Greene  
Kasey Greene  
Westley Hampbrecht  
Will Harbison  
Julianna Harding  
Monica Harding  
Brendan Harrigan  
Alyssa Harvey  
Stacy Hawes Melle  
Clark Huddleston

Robert Hunziker  
Arthur Johnson  
Peggy Jones  
Alex Karwowski  
Franny Kiles  
Don King  
Lori Kipp  
Eleanor Kolchin  
Maia Kolchin Miller  
Susan Lane Hoffman  
Tim Laufenberg  
Ann Laye  
Cole Lefebvre  
Barbara Lenssen  
Maeve Leonard  
Joseph A. Librers  
Rose Logue Harting

Larissa Marocco  
Jessica Martens  
Dave Martin  
Gary Masching  
Alex Maschino  
Helen Maschino  
Marsha McElwin  
Rachel McMullen  
Rebecca McMullen  
Carly McReynolds  
Nancy Melvin  
Jackie Merrill  
Linda Meyer  
Jonathan Miller  
Nancy M. Miller  
Fiona Mischel  
Matt Mitchell

A.M. Murphy  
George Nijmeh  
Lindsay Nijmeh  
Mr. and Mrs. Pierce  
Melissa Predny  
Robyn Primack  
Justin Raines  
Libby Richland  
Rebecca Roanhaus  
Ben Robertson  
Ann Robinson  
Carl Robinson  
Cortney Roeder  
Kathy Rolefson  
Myranda K. Salvage  
Linda Jeanne Scherschel  
Erika Schlotterbeck Harrington  
Joseph Sinnaeve  
Lisa Steiding  
Ana Stenzel  
Isa Stenzel Byrnes  
Brian Stone  
Emily Talbot  
Heidi Tegner  
Tara Telford  
Adam Thompson  
Todd Trisch  
Lyle Tusk  
Betty Vitousek  
Devin Wakefield  
Kassi Watkins  
Denise Wold

### **Vertex** (continued from page one)

in patients. Based on initial encouraging results, Vertex has initiated a Phase 2a clinical trial for VX-809 to evaluate the safety and tolerability of multiple doses of VX-809 in approximately 90 patients with the double Delta F508 mutation. In addition, the trial will evaluate the potential effect of VX-809 on CFTR function.

There are many years of research behind every current clinical trial. In the early 1980s, CFRI funded research that led to a significant breakthrough in the understanding of chloride impermeability in CF. This groundbreaking discovery, conducted by Paul Quinton, Ph.D., laid the foundation for future research, most notably the current Vertex trials.

Through the years, there have been numerous advancements in cystic fibrosis medica-

tions and therapies, but these have largely focused on the treatment of symptoms and complications. With the advancement of compounds that can act as potentiators and

correctors of the CFTR protein, there is hope that the underlying cause of the disease can be rectified.

### **Male Infertility** (continued from page one)

that men who carried two copies of the valine allele were almost three times more likely to conceive than men with two copies of the methionine allele.

In an effort to understand how valine increases a male's fertility, Ober is currently analyzing the semen of approximately 100 men. "Presumably there's something different but whether it's number, or morphology, or what, I don't know what to expect," Ober said. "I'm not sure that a standard semen analysis is going to be enough to identify valine's effect, but if it is it will be amazing."

For men with cystic fibrosis who are infertile, Ober's research on fertility looks promising and perhaps will provide a solution for those who wish to have their own children.

It is important to note infertility in men with cystic fibrosis is primarily due to the absence of the vas deferens tube which prevents sperm from being released. A minor surgical procedure may be required in order to harvest sperm for in vitro fertilization.

For further information on Ober's study please visit the following link:<http://www.the-scientist.com/blog/display/55597/>.

# In Memory of

(Contributions listed were received from  
February 1, 2009 to April 30, 2009)

*"In Memory of" lists names of loved ones we have lost, and in whose memory a donation has been made.*

We extend our deepest sympathy to their families and friends. These gifts continue to give hope to children and adults with cystic fibrosis. "In Memory of" is not only for those with CF but for their families and relatives as well.

**Note:** *Occasionally a deceased person will have the same name as a living person.*

Please send the name of your loved one with an address so that we may send an acknowledgement of your donation.

Send to: CFRI, 2672 Bayshore Parkway,  
Suite 520, Mountain View, CA 94043.

Carol Carey	John Holmes	Jim Neal	Greg Thul
Sonya Chartrand	Leslie Hotson	Kate Nelson	Mark Tillet
Cindy Clark	Sean Hyland	Kim Nelson	Renee Topol
Ryan Coelho	Orzella Jemas	Scott Nelson	John Trask
Sid Cohn	Brian Jensen	Celia Newberger	Maureen Sazio
Rachel Crocker	Dean Joy	Bobby L. Newport	Linda Trojak
Beth Dauphinais Selser	Peter Judge	Jennifer Ortman	Margaret Vasquez
Sandy De La Porta	Kathy Judge Morse	Ernest M. Phipps	Jody Voller
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Elayne Dougherty	Dorothy Kenney	Tim Prater	Sean Waltrip
Barbara DuBose	Kathleen Kiouss	Holli Pratt	Tara Weir
Trevor Eisenman	Bridget Teresa Klein	Catherine Rawlings	Hayley Wester
Boyd Faulk	Bernice Irene Kusalo	Jack Roche	
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Jessie Marie Franks	Dawn Longero	Randy Ruprecht	
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Dawn Gonzales	Margaret McFadden	Dhea Schalles	
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Given Grimsman	Janie Meeker	Jesse Schuh	
Stephanie Southworth	Pietta Menosse	Lindy Sheppard	
Halling	Anna Mesquita	Millicent G. Silver	
Gloria Halton	Jessica Mobley	Mary V. Stengel	
Virgil Hanson	Kathy Moore	Jordan Sterling	
Warren A. Harden	Betty Morgan	Robert C. Stewart, Jr.	
Carl Hardy	Roger Morris	Nancy Teese	
Muriel Hardy	Linette Irene Moulton	Hazel Thompson	

Carol Adelman	Jodi Armknecht	Helen Bergin
Kim Adelman	Estel Back	James W. Bertolini
Marcus Adelman	Ann Baldwin	Ingeborg Bitter
Esther Anderson	Ron Baldwin	Myrtie Boore
David Armknecht	Anne C. Beltrame	Greg Brazil



## Help Us Grow - Donate to CFRI

*Your Donations Support Vital CF Research and Education*

### Become a Member

CFRI depends on its membership to help meet the needs of the cystic fibrosis community. As a member, you will have an active voice in the organization's delivery of services. A large and active membership is key to CFRI's organizational health and responsiveness. Annual dues are just \$25.

### Life Settlements & Planned Giving

Planned giving offers benefits for donors that often include increased income and substantial tax savings. Life Settlements, and the donation of your life insurance policy if you no longer need it, may also be to your advantage and benefit CFRI. Both events create the opportunity to meet your philanthropic goals and provide positive tax benefits.

### Giving Stock

Giving a gift of appreciated stock is easy and can be rewarding in several ways. You will not 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 make possible our research and educational programs that are helping everyone face the challenges of cystic fibrosis. Please call 650-404-9975.

### In Honor of, In Memory of

Any gift given can be made in honor or in memory of a loved one. Their name will appear in our newsletter, and an acknowledgement will be sent to the person honored or to their family. Many choose to send in a group contribution to celebrate occasions such as birthdays and anniversaries.

### Vehicle Donations

For many years CFRI has received donations of vehicles. If you have a used car, boat, recreational vehicle or motorcycle, please consider donating it to CFRI. This contribution is tax-deductible and we will coordinate the transfer of property and handle the paperwork from any state in the U.S.A.

### CFRI Golf

At the CFRI Tournament, golfers spend a rewarding day at the course and are treated to a fantastic dinner and auction in the evening, all to support cystic fibrosis research! This year's tournament will be held at Cinnabar Hills Golf Club in San Jose, California, on Monday, August 3, 2009. If you're interested in participating, please contact Scott Hoyt 408-323-7803, [shoyt@cinnabarhills.com](mailto:shoyt@cinnabarhills.com).

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**Genentech, Inc.**  
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## CFRI's Mission

*Cystic Fibrosis Research, Inc., exists to fund research, to provide educational and personal support, and to spread awareness of cystic fibrosis, a life-threatening genetic disease.*

## CFRI's Vision

*As we work to find a cure for cystic fibrosis, CFRI envisions informing, engaging and empowering the CF community to help all who have this challenging disease attain the highest possible quality of life.*



**Get to know us:**  
**www.cfri.org 650.404.9975**

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