The Time Is Now: The Critical Need for Increased Research Funding

By Paul Quinton, Ph.D.

The time to increase funding for cystic fibrosis (CF) research is now. Not tomorrow. Now.

I am extremely honored to serve as the honorary co-chair for CFRI’s “2014 CF Research Challenge Circle Fund.” As a scientist who has worked for decades on the challenges of cystic fibrosis (CF), I am alarmed by the decrease in available funds for researchers, most notably from the National Institutes of Health (NIH). I fear setbacks that we can – and must – avoid. CF researchers have achieved what many would call brilliant successes. Not long ago children with CF rarely reached their teens, but now there are as many adults with CF as children. Don’t get me wrong – cystic fibrosis is not solved, and will not solve itself. It has, does, and will need you to support advanced research to understand how to control – and ultimately cure – the disease.

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CF Research Challenge Match

CFRI is embarking on a critical campaign to raise additional funds for cystic fibrosis (CF) research programs, at a time when CF research funding is declining. There is no cure for CF. There is much to be done.

You can help.

Our many thanks to honorary co-chairs of the CF Research Challenge Circle, Paul Quinton, Ph.D., a preeminent CF researcher living with CF, and Jim and Barbara Curry, longtime friends of CFRI, grandparents to Cameron who has CF, and strong advocates for increased CF research. Their strong belief that the time is now for CFRI to increase funding for CF research has enabled us to bring together additional strong supporters to offer a large pool of funds that CFRI can use to match donations throughout the year to sponsor CF research.

Our goal for this newsletter is to raise $15,000 in gifts so that we can access $15,000 from the pool of matching funds so as to reach $30,000 in additional funds for our CF research projects by April 30. You can help us meet our goal as follows: If you have not given a gift in the past 12 months, your gift will be matched dollar for dollar, doubling the impact of your giving! If you have given a gift within the past 12 months, please increase your gift and the increased amount will be matched dollar for dollar!

Our goal for 2014 is to raise a minimum of $150,000. These funds will be restricted to CFRI’s research programs, which include the Elizabeth Nash Memorial Fellowship Program, the New Horizons Research Program.

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Paul Quinton, Ph.D.
Notes from the Executive Director

Dear CFRI Friends,

“Symbiosis: A relationship of mutual benefit or dependence.” Life at CFRI is springing ahead with a multitude of exciting projects that are cause for both celebration and reflection on how CFRI and the CF community are linked symbiotically. CFRI exists to serve the needs of you: our community of individuals living with cystic fibrosis (CF) and their loved ones, as well as the caregivers, researchers and healthcare professionals involved with providing support and professional expertise to those with CF. Because of you, we are here, and we continuously strive to meet the current and changing needs of our community.

Our new look, with the fresh new logo, incorporates our philosophy and serves as the foundation for our name. I want to recognize CFRI’s staff, editor Bridget Barnes, and graphic designer Marina Ward for the collective effort that led to our inspiring design. The infinity symbol represents something without any limit. It is a natural addition to CFRI’s name as our purpose is to provide education, research, awareness, and support to our community, now, and into the future. We want to recognize that the symbiotic relationship between CFRI and our beloved CF community knows no bounds. It is infinite.

Our new look also marks the kick-off of our 2014 CF Research Challenge matching fund program. We must do more - we must do it now - so that we can move closer to a cure. It is imperative that CFRI funds more CF research projects at the basic cellular level to increase knowledge about the basic CF genetic defects, to engage new and promising scientists in the field of CF, and to fund CF scientists that have years of valuable experience. We are all working toward the same goal – to touch the future and touch lives with new drug therapies and a cure for CF.

Warmly,

Sue Landgraf
Executive Director
Mother of an adult daughter with CF

News from the Board

CFRI’s Board of Directors is excited and optimistic about the opportunities for the organization in 2014. We’re also deeply aware of the challenges that face those with CF, and are positioning ourselves to meet the changing needs of our community. Through the leadership of our Executive Director, Sue Landgraf, we have established a formal Finance Committee, which includes experienced accountants and CPAs, and provides an improved focus on our budget process. The first task of the Finance Committee was to review CFRI’s accounting system and recommend improvements. In order to better maintain the financial elements of Board fiduciary responsibilities, we have approved and monitored the installation of a more appropriate and robust accounting system that facilitates clear understanding and sound audit processes.

The Board is determined to develop a strategy for balancing the distribution of funds between research, education and support programs,
Development and Characterization of Novel 3D Airway Cell Models for Cystic Fibrosis Research

By Carolin Boecking, M.D.

Cell culture models are an integral component of cystic fibrosis (CF) research. They enable us to study, understand and modify biochemical processes of the airways in a laboratory environment. Furthermore, cell culture models aid in the discovery and development of novel therapies by providing an in vitro platform for testing the effects of potential new treatments.

Human airway cells have been widely studied using an air-liquid interface cell culture model. This model is based on growing cells on permeable filters and exposing their apical surface to air. This system mimics the airway mucosal lining and provides a practical cell model for the airway surface cells. However, cultures of submucosal gland (SMG) cells grown this way lack their normal three-dimensional (3D) orientation within the airway connective tissues, and fail to develop full differentiation. Hypersecretion of SMGs is believed to be a major contributing factor of airway destruction in cystic fibrosis patients. Additionally, airway CFTR has been predominantly localized in serous cells of gland acini in the human bronchus.

Our goal is to provide highly differentiated 3D cell culture models, producing a vast number of airway cells for use in high throughput screening and functional studies of gland cell biology.

Our first goal is to develop and characterize 3D organotypic airway surface epithelial (“epitheliods”) and SMG (“acinoids”) cultures. Organotypic cultures closely resemble parent tissue in morphology (form and structure) and functionality. To achieve cultivation of organoids, airway cells will be grown suspended in 3D gels (Matrigel™). The gel mixture resembles the complex extracellular environment found in many tissues including the lung, and supports cell growth. Its porous matrix allows for absorption of the growth medium to test for their effects on the development of airway-derived cell spheres. The added stem cell factors, including R-spondin, Noggin and BMP4, are believed to be involved in the process of lung branching, and promote airway cell differentiation.

Our second goal is the development and characterization of 3D rotating wall vessel bioreactor cultures of human airway epithelial and SMG cells. Rotary bioreactors, developed to mimic the zero gravity in space, may influence cellular functions such as differentiation, proliferation, viability, and cell cycle. Airway cells are grown in a simulated microgravity, providing a low-shear environment in suspension and simulating in vivo conditions. The environment within the rotary cell culture system enables cells to assemble into tissue-like aggregates with high mass transport of nutrients, oxygen and waste. Growth in 3D aggregates can be encouraged by either the introduction of microcarrier beads, e.g. Cytodex 3, or encapsulation of cells in alginate beads, all of which simulate the airway connective tissues.

We are optimistic that the development of both novel 3D cell culture models could potentially advance, expedite and even expand treatment options for cystic fibrosis. Organoids, as well as cultures cultivated in the rotary bioreactor, will provide an increased level of cell differentiation as well as sufficient numbers of cells for drug discovery studies.
I know research is the answer. We must remain committed to unraveling the complicated disease processes of the most common fatal genetic disease in North America. Unless we understand the basis of the abnormal cellular function, we cannot restore it to normal. But it is becoming more difficult to solve the mysteries of cystic fibrosis. I am alarmed by the diminishing prospects for “young” investigators to work on CF, and for “old” investigators to maintain laboratories dedicated to solving this disease. It is time for a greater community investment in CFRI’s research funding. Over the past 37 years, CFRI has played a vital role in supporting both veteran researchers and those new to the field, and the impact of this support is immense.

One has only to look at the wealth of CFRI-funded research breakthroughs to know that every dollar makes a difference. My peer and colleague, Dr. Jeffrey Wine, a CF researcher and CFRI grant recipient, has had a slew of discoveries that have advanced the scientific world’s understanding of CF, including the identification of the first heterozygote effect of the CFTR mutation, and numerous landmark studies of the airway submucosal gland. In 1986, CFRI-funded researcher Dr. Jonathan Widdicombe was the first to demonstrate that cultures of airway epithelium from CF patients lacked chloride secretion. With CFRI’s support from 1981 to 1983, my lab discovered that the basic functional problem was with chloride ions not moving across CF epithelial tissues, thereby pointing to the malfunction of an as-then-undefined chloride transport channel. Since then, we have found that defective movement of bicarbonate ions largely explains why mucus in CF is abnormally thick. CFRI-supported research brings understanding of CF – and the conviction that a cure will come.

In addition to being a CF researcher, I am also an “old” adult with cystic fibrosis. As a scientist focused on unraveling this disease, I know that living with CF means sometimes having to set limits and make difficult choices in order to live. I have seen the relief, the hope, and the brighter futures that CF research brings. Although we have no cure, there will be better control of the disease process as we observe and test for changes and improvements in cell, organ and patient functions. We have developed new therapies that have enhanced and prolonged lives. Please, don’t let this progress stop. With the community’s support, CFRI will continue to champion cutting-edge research that will touch the future. Our improved survival rates are linked to discoveries that emerged with CFRI’s support. I urge your full commitment to “fixing” CF. Please contribute to the 2014 CF Research Challenge.

The time is here. The time is now.

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For information on how you can join our 2014 CF Challenge Circle, please contact Sue Landgraf, Executive Director, at 650-404-9975.
Marianela Fajardo knew something was wrong with her baby, Maria. The mother of nine children, she was well versed in reading the signs, and Maria was seriously ill. From birth, Maria had such severe coughing spasms that she turned blue from lack of oxygen. Four months of medical tests failed to identify what was ailing her, and Maria was admitted to the intensive care unit of a San Jose hospital. A consulting doctor from Lucile Packard Children’s Hospital at Stanford (LPCH) then recommended the sweat chloride test that solved the mystery. Maria had cystic fibrosis (CF).

“I was so distraught. I didn’t know what cystic fibrosis was, I didn’t know anyone who had it – I couldn’t even pronounce it!” recalls Marianela. “Everything happened so fast. We were transferred to LPCH, and all of a sudden there were so many people – the pulmonologist, nurses, dietician, respiratory therapist, social worker – it was overwhelming.”

The first days after diagnosis, Marianela, “felt numb and confused,” but then shifted her focus to educate herself about CF, and to learn about the complex medical regimen that Maria would follow when she came home.

Thus began the Fajardo family’s journey with cystic fibrosis. Two years later, baby Francisco arrived, and was also diagnosed with CF. At the time, Marianela’s children ranged in age from infancy to 16 years old, and the older children rose to meet the responsibilities created by their siblings’ diagnoses. Says Marianela proudly, “It was hard at first, but they adjusted. It brought them closer. They wanted to be hands on. I remember they’d pass Maria around and ‘play the drums’ on her back for her percussion therapy. When they got older and Maria and Francisco were hospitalized, they all helped to do chores and laundry. My kids are very caring and responsible.”

Marianela quickly became a hospital veteran. Within 13 years there have been at least 20 inpatient stays, including a three-week stint after Maria’s double lung transplant in November 2012. “Honestly, I have lost count,” says Maria. “We’ve been there a lot.”

From her own experience, Marianela encourages people to work closely with their healthcare team, and to be vocal about their care. By the time she was 10 years old, Maria’s health had significantly worsened, and she needed oxygen around the clock. Says Marianela, “From the beginning, I told Dr. Conrad (Maria’s doctor), ‘Do not beat around the bush with me. Whatever is going on, tell me straight.’”

This open communication helped Marianela to work with Maria’s care team and create a plan to best support her daughter through transplant. A medical assistant by training, she secured a job outside of her profession that provided excellent benefits, and from which she was able to take a three-month leave during Maria’s recovery. Marianela’s older daughter left her job in order to help provide the round-the-clock care required post-transplant. While each hospitalization brings tremendous stress to the family, the long days spent as an in-patient, especially in light of cross-infection protocols that require those with CF to remain in isolation, create a special closeness.

While Marianela is adept at juggling her children’s healthcare with work and family responsibilities, at times this means that she must leave the hospital when one of her children is an inpatient. While confident in the quality of care her children receive, she wants to be there. Marianela draws comfort from the knowledge that her children have become their own health advocates. “Francisco always has a lot of questions for the doctors, and he’s only 10,” says Marianela. “Maria listens carefully, and is very vocal about her health.”

In addition to advocating for her own children, Marianela is an unofficial mentor, translator and advocate for Spanish-speaking families whose children have been diagnosed with cystic fibrosis (fibrosis quística). Every state in the U.S. now tests for CF through newborn screening, and in California nearly half the babies diagnosed are Hispanic. Many CF centers are now struggling to meet the needs of their Spanish-speaking patients and families. “Imagine how
Diagnosis Doesn’t Determine Destination

AKA: Preparing for a Fulfilling Life in a World with No Guarantees

By David R. Cartnal – Architect, Husband, Father, Grandfather, Mentor, Coach, Fellow CF Sojourner

If we allow ourselves to be driven by statistical averages (the median CF age of survival is 37 years), there would be no reason to spend time or effort planning for things like college, career, marriage, family, and retirement. My hope in sharing my personal 71-year CF journey and the lessons learned, is that children, teens, young adults, and their families will not simply mark time, but seize the day and consider a much more fulfilling alternative. None of us has any guarantee we will see tomorrow. Life’s journey is worth the preparation and exploration, if we simply understand the premise: Our attitude largely determines our altitude.

When I was first diagnosed with CF, my FEV-1 (lung function) was 28% of normal. Today, after many hospitalizations, clinic visits, medications, therapies, and prayers, my FEV-1 is 46%. My determination to live a full and productive life remains undaunted, even though my health condition, like any person with CF, may still be at risk. Our birthright, genetics and family are chosen well before we arrive. We needn’t be defined by our DNA, nor a chronic disease. Instead, we should be defined by our character, relationships and contributions. This article is a humble revelation of one person’s attempts to not simply survive CF, but to thrive in all aspects of life’s rich journey.

My childhood consisted of life on a farm and all the accompanying irritants to my respiratory system. When I was two years old, I had my first serious episode, but the doctors thought it was just allergies. As the only boy in our family, I didn’t wish to be confined indoors, so I often suffered the consequences of too much dust, animal dander, fertilizer chemicals, and poor decisions.

During adolescence, I wanted to be like every other kid, active and engaged, not sitting on the sidelines watching others participate. In the 7th grade, I had my first collapsed lung, though not my first hospitalization. My small-town doctors ran a battery of tests and concluded I had chronic respiratory allergies that I might grow out of if I endured two years of desensitization injections. My natural tendency was to hide this “defect” from my friends and classmates, but my frail physique and chronic cough were a dead give-away (I was 4’8” and weighed 78 lbs. in the 8th grade). Lesson Learned: Face reality head on; accept what you cannot change; find alternative ways to your goal and always profit from your experience — especially your mistakes.

Moving into high school, but still wanting to be like all the other guys, I decided to participate in sports. I didn’t have the body mass to play football, so I chose basketball. A strenuous running game was not a wise choice for a kid who wheezed with every trip down the court, but I learned to become a skill-player and focused on my free-throw shooting and jump shots. As I grew and matured, I re-focused my competitive desires toward sports that I could play for life, like swimming, tennis and golf. Lesson Learned: Learn to excel at things you can master — you can still make an invaluable contribution. Always take your ‘A’ game, but never begin without a Plan ‘B’ in your hip-pocket.

Career choices are hugely important in a person’s life, providing mental stimulation, financial independence, self-esteem, and personal fulfillment. As a young boy, I was forced to learn the necessity of pursuing an indoor career. During my many confinements, I learned to sketch, draw and write as forms of communication and enjoyment. Little did I realize it might ultimately point me toward a career in architecture, after a chance encounter with a Chicago architect who explained the possibilities and fulfillment from such a career.

After dealing with such major life milestones as health, self-esteem, education, and career choice, I moved to the next big plateau: dating, marriage and family. After eight years of marriage, my wife and I discovered we couldn’t have children. Instead of resigning ourselves to the notion, “I guess we weren’t meant to have children,” we adopted an infant son; two years later, an infant daughter. Lesson Learned: Never accept ‘no’ as the final answer. Had we not adopted, we would have lost a lifetime of family experiences, including the joys shared with two healthy (non-CF) children and six talented, healthy grandchildren.

As life unfolded, I became convinced there was very little I couldn’t attempt, including hiking, biking, skiing (snow
Marianela with two of her children, Francisco and Maria, both of whom have CF. 

Maria Fajardo, Advocate
Continued from page 5

hard this is,” says Marianela, “to have to cope with the diagnosis, and learn about the disease when you don’t speak English.” Marianela has made herself available to help other families, even when Maria has been in the hospital. “I’m happy to offer whatever help and support I can. To this day I bump into people that I met and talked with when their children were first diagnosed. I will always be an advocate.”

Marianela hopes that in addition to expanded resources for Spanish-speaking families, there will soon be better understanding of why Hispanic children with CF tend to be so sick. Numerous studies have found that Hispanics often have more severe disease expression than Caucasian patients, and researchers are exploring the many factors that may contribute to this, including CF genotypes, modifier genes, and economic and environmental issues.

Marianela’s advice for children and families entering the hospital or clinic: “Stay focused, and ask as many questions as you can. And ask for help, it’s OK.” An advocate for her children and the broader CF community, Marianela Fajardo helps others to cope with a frightening diagnosis, thereby embodying her favorite quote by the writer Maria Anderson, “Nobody can go back and start a new beginning, but anyone can start today and make a new ending.”

Tunes and Brews
Raise Awareness & Funds for a Cure

By Bridget Barnes

On a chilly evening last December, Tess Dunn and her band heated up a packed house while performing her original songs in a benefit concert for cystic fibrosis (CF) and organ donation awareness. Honoring Gianna Altano, who last spring became a donor after losing her fight with CF at 23, Tess (who has CF) shared her remarkable spirit, humor and musical talent, leaving the audience aglow. Generously hosted by Bill Welch of Moe’s Alley in Santa Cruz, the event facilitated the registration of many new organ donors and raised over $9,500 for CFRI. It also featured opening bands Urban Theory and The Royal Oui, a successful raffle and silent auction, cake to celebrate Tess’ 19th birthday, and an official proclamation declaring it “Tess Dunn Day” in Santa Cruz County.

Two days earlier at Tap 25, a pub in Livermore, Diana Heppe organized “Brews for a Cure,” featuring nine new beers from New Belgium Brewing Company. A portion of the cost of each pint was donated to CFRI from both “Tap” and Altamont Beer Works between 12 noon and 10 p.m. New Belgium Brewing Company donated their signature bike, which was raffled among several other prizes. The event raised $2,600 and brought together many supporters of adults and children living with cystic fibrosis to raise a glass for a cure.

CFRI is very appreciative of the goodwill of these volunteers. Events like theirs are the lifeblood of our organization, both for the financial support they provide, and the strength and spirit they bring to our community.
In Honor of

Tributes

Our “In Honor of” and “In Memory of” pages provide the opportunity to honor a person, family, or special event, or to remember a loved one.

If you want your donation to honor or remember someone special, please include the person’s name and address with your donation.

At your request, we will send an acknowledgement of your gift to the person you designate.

Please mail your contributions to:
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2672 Bayshore Pkwy.
Suite 520
Mountain View, CA
94043
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Inflammation and CF

By Laura Tillman

Inflammation is a major contributor to lung damage in cystic fibrosis (CF). The inflammatory process irreversibly damages the airways, causing bronchiectasis and, ultimately, respiratory failure. The safety and effectiveness of numerous anti-inflammatory therapies for CF, including oral corticosteroids, ibuprofen and azithromycin have been studied, showing that several treatments are beneficial, particularly in younger patients with mild disease. Ibuprofen and azithromycin are generally recommended as “standard therapies,” due to research demonstrating their impact on preserving, and even improving, lung function for some patients.

Cystic Fibrosis Pulmonary Guidelines: Chronic Medications for Maintenance of Lung Health.
The authors developed CFF recommendations related to pulmonary medications. For patients with CF, 6+ years of age, without asthma or allergic bronchopulmonary aspergillosis (ABPA), the routine use of inhaled corticosteroids to improve lung function and reduce exacerbations is not recommended. For patients 6 to 18 years, without asthma or ABPA: oral corticosteroids should not be used chronically. For adult patients with CF, 18+ years of age, without asthma or ABPA: evidence is insufficient to recommend for or against the continuous use of oral corticosteroids. For patients with CF, 6+ years, with an FEV1 greater than 60% of predicted, the ongoing use of oral ibuprofen is recommended to slow the loss of lung function.

http://tinyurl.com/nyvijpmj

Antibiotic and Anti-Inflammatory Therapies for Cystic Fibrosis.
Review of current and future antibiotic and anti-inflammatory therapies for CF patients concludes that high-dose continuous ibuprofen treatment is the best-proven anti-inflammatory drug thus far. The authors emphasize that more studies are necessary to determine which therapy provides the most benefit, and to establish the optimal patient age and stage of lung disease at which to begin treatment.

http://tinyurl.com/izlc4dj

Anti-Inflammatory Therapies for Cystic Fibrosis: Past, Present and Future.
Besides ibuprofen and azithromycin, this article examines other treatments such as inhaled corticosteroids, colchicine, methotrexate, montelukast, pentoxifylline, nutritional supplements, and protease replacement, all of which have not had a significant impact on CF. Oral corticosteroids, ibuprofen and fish oil therapies have been found to be limited due to adverse effects.

http://tinyurl.com/mq3j2yr

Oral Non-Steroidal Anti-Inflammatory Drug Therapy for Lung Disease in Cystic Fibrosis.
Review indicates that high-dose ibuprofen can slow the progression of lung disease in people with CF, especially in children.

http://tinyurl.com/lzlc4dj

Anti-Inflammatory Medications for Cystic Fibrosis Lung Disease: Selecting the Most Appropriate Agent.
Concludes that correction of the basic CFTR defect must also correct inflammation in order to be effective. Until therapies repairing the basic defect are fully achieved, limiting the effects of the inflammatory process is important in slowing pulmonary decline, thus prolonging survival in patients with CF.

http://tinyurl.com/k38t8qm

Anti-Inflammatory Therapies for Cystic Fibrosis-Related Lung Disease.
Among proven therapies for inflammation of the airways, high-dose ibuprofen and azithromycin are presently the most widely used agents. Other compounds have been studied, or are under investigation, to identify an effective treatment that is more acceptable to the CF community. If realized, this would be a significant advance in the care of CF patients.

http://tinyurl.com/kjwzpdd
Elyse Elconin-Goldberg was three years old when she was diagnosed with cystic fibrosis (CF). The year was 1961, and at that time, children with CF rarely survived to their teens. Elyse grew up in West Los Angeles with a younger brother, and parents who raised her with a “can do” attitude. She received her Bachelor’s in Urban Planning from UC Davis, and her Master’s in Urban and Regional Studies from the University of Southern California. In 1985, Elyse married her husband Craig and relocated to the Bay Area. Elyse worked in the private sector until 1993, when she and her husband adopted their first child, Stephanie, followed by their son, Bryan, in 1997. Until late 2012, Elyse was managing to raise her family and pursue her many other interests. Yet after battling two debilitating viruses, her health began a steady decline. Elyse was listed for transplant in June 2013, and received new lungs on September 17, 2013. In the following interview, Elyse shares the story of her road to transplant and the new path she follows.

What changes have you have experienced in CF care over your lifetime?
There are so many! DNase, inhaled antibiotics and hypertonic saline just to mention a few. When I was young, I slept in a mist tent until doctors realized that the moist environment was a breeding ground for pseudomonas. The early care I received was CPT (chest physiotherapy) and oral antibiotics. I didn’t start using nebulized medications until I was 21.

Tell me about the decision to be listed for transplant.
I had lost lung function from two back-to-back viral infections. I was diligent about my respiratory treatments (“compliant” is my middle name), exercised daily and ate well, yet my health wasn’t improving. By April 2013 I was using oxygen 24/7, and Dr. Mohabir asked if I would consider meeting the Stanford lung transplant team. I had never imagined myself going through a lung transplant. Soon after this idea was presented to me, I was admitted yet again for another CF exacerbation.
I asked the medical team if they could arrange for me to start the pre-transplant testing while I was an inpatient. Things started moving at an incredibly rapid pace.

What about your emotional response?
I will admit to being scared, nervous, and at times uncertain of this decision. I have since learned that I’m not alone in these feelings. Once listed, you think, “I’m not sick enough to need a transplant.” I tended to minimize each step down in health, so that it became my “new normal,” then I took one step lower and that became my “normal,” and so on. Ultimately, it comes down to the truism that “you have to be sick enough to need a transplant, and well enough to survive it.”

How did you prepare for transplant?
I trained for my transplant like I was training for a marathon. Working out was not only a huge plus in preparation for transplant, but it really boosted my emotional well-being. Between exercising and doing four respiratory treatments, I spent seven hours a day on my healthcare. Also, the Stanford lung transplant team really prepares you for your upcoming transplant. You have to attend sessions with a social worker as well as attend a comprehensive class of what to expect pre, during and post transplant.

Tell me about the call.
I was listed for two and a half months, which I recognize was a very short time. I am very grateful that a match became available so soon. I was playing Mah Jong at a friend’s house and my phone rang. It was Kelly, one of the Stanford transplant nurse practitioners. She asked me several questions about how I was feeling, and I suddenly realized the point of her call. I broke out into a sweat, and when she said that a donor set of lungs had become available for me and asked if I was willing to accept them, I just yelled out “YES!”

Is the post-surgery period difficult?
I had an amazingly easy and fast recovery. I recognize this is not always the norm, and I am very thankful that what few bumps in the road I had weren’t major ones. Post-transplant, being so immunosuppressed, I have a similar, yet greater, “protective dome” than I did pre-transplant. It feels so different from pre-transplant days because I feel so healthy. But, I really need to protect myself now by wearing a mask outdoors, in large crowds, and in a hospital environment. Hand sanitizer accompanies me wherever I go!

Continued on page 12
Elyse’s Journey  
Continued from page 11

What CF issues do you still cope with?  
I still have to do sinus flushes daily, keep on top of GI issues, as well as manage my CFRD (cystic fibrosis related diabetes).

What is life like post-transplant?  
Fantastic! I am embarking on a journey of re-inventing myself. What this will be, I am yet to be sure of. I’m full of energy and excitement for life. For me, I’ve traded hours of respiratory treatments for a huge number of pills to be taken throughout the day. I am eternally grateful to the Stanford Hospital lung transplant team, the nurses, the respiratory therapists, and to my husband and kids. I so hope to be able to thank my donor’s family for the gift of life they have given me.

Diagnosis Doesn’t Determine Destination  
Continued from page 6

and water), swimming, golf, and travel. I disclose this in the hopes some youngster, young adult or parent may find encouragement in their pursuit of a “normal” life, despite a CF diagnosis. While you may not be able to pick your DNA, you most certainly can pick your attitude, core values, outlook, sense of humor, mate, and lifestyle, and surround yourself with interesting, supportive friends. What more could anyone ask from this amazing, exploratory journey?

Lesson Learned: Listen to your body (and your doctors, nurse practitioners and respiratory therapists); use common sense and good judgment; be compliant with your medications, proper diet, adequate sleep, and regular exercise; find ways to give to others; don’t take ‘NO’ for a final answer, and enJOY every day of this unpredictable, but wondrous journey.

Note: To read Dave’s expanded list of ‘Lessons Learned’ go to: www.cfri.org
Coping With CF: A Warrior’s Perspective

By Darlene Batchelder

A wise warrior prepares for battle. A wise warrior is clear about the nature of the enemy, assessing its strengths and weaknesses. Even if the enemy is bigger and stronger, a warrior imagines what it will feel like to win the battle and is excited, knowing that it is possible, even though it is not clear exactly how the battle will be won.

I believe this is what it takes to confront cystic fibrosis (CF) when it arrives with your child.

My son Joseph is now 26 years old. He was diagnosed with CF in 1989 when he was 12 months old, before the CF gene was identified, before Pulmozyme and the other therapies that exist today were developed. My husband and I were shocked by the diagnosis, and then determined. I wasn’t a trained life coach at the time, and have since learned a great deal about what to do – and not to do – as the parent and primary caretaker of a child with CF. When I realized the magnitude of our opponent, I did something very powerful: I declared that our son would live as long as I did. At that time, children with CF were dying as young as 5 or 6 years old, and there were no promising therapies on the horizon. I had absolutely no idea how to realize this seemingly preposterous declaration. Still, I was resolved.

As a life coach, I now know that this is the single most effective thing to do when confronted by the impossible: Declare that it is possible, and be determined and willing to work to discover how. This is empowering because it requires you to focus on a desirable future result, and to be active in this process of discovery. The alternative is to constantly focus on a frightening future, and work from a place of fear to avoid it.

I knew when I took on Joseph’s battle with CF that my well-being was the foundation on which my ability to mentally and emotionally handle CF was built. Warriors are in it for the long haul, because a relentless opponent will always be lurking. So a wise warrior takes exquisite care of him or herself, with plenty of rest, time for reflection, respite, excellent nutrition, and emotional and spiritual support. A caregiver’s well-being is paramount.

CF inevitably means pain, for both the person with CF and the family. While there is no way to escape this reality, the worst thing we can do is to deny it. It is possible to have and hold a vision or commitment that is bigger than our fear, one that inspires us as we battle CF.
Save the Dates

**CFRI Teen & Adult Retreat**
Relax – Recharge – Rejoice at the CF Zen Retreat
July 27 – August 1, 2014
Vallombrosa Center
Menlo Park, CA

**CFRI 27th National Family CF Education Conference**
The Changing Faces of Cystic Fibrosis: Inspiring Hope
August 1 – 3, 2014
Sofitel San Francisco Bay
Redwood City, CA

**29th Annual Golf Tournament**
Benefitting CFRI
Monday, August 4, 2014
Pasatiempo Golf Club
Santa Cruz, CA

**CF Discovery Series**
Tuesday, March 11, 2014
Tuesday, April 8, 2014
6:00 p.m. – 7:30 p.m.
Crowne Plaza Cabana Hotel
Palo Alto, CA
Watch and participate online at www.cfri.org

Support CFRI and Become a Partner in Living!

Your support is invaluable and enables CFRI to offer vital education, research and awareness programs to the CF community. Please consider making a gift today through the options below.

- The 2014 CF Research Challenge: Your gift will be matched 1:1 if you have not given in the past 12 months; if you have given within the past 12 months, any increase in your gift will be matched. All contributions will be restricted to our CF research programs. See page 1 for more details.
- A donation of stocks or other marketable securities
- Your company’s Workplace Giving program
- A vehicle donation
- Our online donation program at www.cfri.org

Thank you for keeping CFRI in your charitable donation plans! Become a Partner in Living. You can touch the future, by touching a life.

and CFRI’s operating reserves to address opportunities and challenges. To this end, we will develop a Strategic Planning Committee to create a necessary plan to guide our growth so that we can continue to meet the needs of our CF community.

In closing, the Board is mindful of the loss of many friends in 2013, including Ana Stenzel, a long-standing Board member and good friend to CFRI, and we wish peace for the families and friends of those who are no longer with us.

Peace and good health,
Bill Hult, Board President

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Thank you for keeping CFRI in your charitable donation plans! Become a Partner in Living. You can touch the future, by touching a life.
As the mother of two lovely adult daughters, the oldest with cystic fibrosis (CF), each year I look forward to celebrating Mother’s Day and reflecting on the joy they have given me. CFRI inspires the same. Every year, the outpouring of support that CFRI receives through its Tea brings a deep contentment, knowing that friends, family and countless others are actively supporting the search for a cure through our biggest annual fundraiser. Our goal this year is to raise $205,000 for CFRI’s vital programs.

By volunteering as a Tea Sender, you can join this devoted group of advocates who send invitations to their family and friends, encouraging them to enjoy a cup of tea on Mother’s Day while reflecting on their loved ones living with cystic fibrosis. Donors give in their honor to further CFRI’s outstanding research and education programs, and improve and expand the lives of people with CF.

This year’s brochure features 12-month-old Kareese Wilson, who was diagnosed through California’s newborn screening program. As the future for Kareese and others living with CF continues to brighten, CFRI is proud to play such a crucial supporting role in the lives of those impacted by CF, from infants to seniors.

If you would like to become a Tea Sender, now is the time! Please call our office at 650.404.9975, email cfri@cfri.org, or sign-up online at www.cfri.org. We will provide you with everything you need, including invitations, tea bags and mailing labels. In turn, you will have the joy of being a key supporter of CFRI and the CF community.

The Seasons Go Round and Round... It’s Tea Time Again!
By Bridget Barnes

To ensure good health for all, please use proper hygiene practices. All participants and guests with CF must comply with CFRI’s Infection Control Guidelines. See www.cfri.org for specifics.

For more information, visit www.cfri.org or call 1.855.cfri.now
CFRI’s General Membership Meeting:
Let Your Voice Be Heard!

By Siri Vaeth Dunn, M.S.W.

CFRI’s General Membership Meeting will be held on Wednesday, May 28, providing our community with the opportunity to vote on organizational issues, including the election of CFRI’s Board of Directors and approval of research grant funding. In addition, members will hear from the esteemed Jeffrey Wine, Ph.D., about recent updates in cystic fibrosis (CF) research.

To be a member of CFRI is to join a community of people that includes those diagnosed with CF, parents, spouses, partners, researchers, and clinicians. There are two levels of membership: voting and general. There is no membership fee, and anyone may apply.

The key responsibility of voting members is to review the CFRI materials under consideration and to vote by mail, or at the meeting. For those who do not wish to vote, general membership is the perfect alternative. All members receive CFRI updates, our newsletter, and invitations to events.

The meeting will be held from 6:30 p.m. to 8:30 p.m. at Bayside Business Plaza, 2665 Marine Way, Conference Room C, in Mountain View, CA (off Garcia and Bayshore Parkway). Become a member and help lead CFRI into the future.

For their generous support of CFRI Community, special thanks to:

Novartis • Cornerstone Therapeutics • Genentech

Call toll free: 1.855.cfri.now

Cystic Fibrosis Research, Inc. a 501(c)(3) nonprofit organization Federal EIN# 51-0169988