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CFRI news™

38 Years of Research, Education & Support

New Lungs and New Opportunities: An Overview of Transplantation

By Siri Vaeth-Dunn

Artist Eva Markvoort did much to bring global awareness to organ donation during her lifetime (www.65redroses.com). Photo by Cyrus McEachern, Copyright Provincial Health Services Authority, BC Transplant.

While significant progress has been made in developing new treatments for cystic fibrosis (CF), lung transplantation is the only life-extending option for many with advanced disease. As presented by Gundeep Dhillon, M.D., Assistant Professor of Medicine, Division of Pulmonary and Critical Care Medicine at Stanford University Medical Center, transplantation is a complex world unto itself. While there are risks and potential complications, lung transplantation can give people with CF the opportunity to live their lives free from the physical limitations previously imposed by advanced lung disease.

The first organ transplants were performed in the 1960s, but due to acute rejection soon after transplant there were "uniformly poor outcomes." The use of anti-rejection drugs – notably cephalosporin – as well as the study of kidney transplants led to the first successful double lung transplant in 1987, and since then the number of transplants has "exploded," said Dr. Dhillon. While there are approximately sixty transplant centers in the U.S., the ten centers that perform the largest number tend to have better outcomes.

Cystic fibrosis is the third most common reason for lung transplantation in the U.S., and CF patients almost exclusively have double lung transplants. Dr. Dhillon discussed the criteria involved with "candidate selection," including those who are under age 65, for whom medical therapies are ineffective or unavailable, and who have a life expectancy of less than eighteen months. Optimal candidates must

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RESEARCH

Focus on CF Lung Infection: *P. aeruginosa*

Pradeep Singh, MD

Despite concerns about drug resistance, conventional antibiotics generally work well in acute bacterial infections, provided that organisms are antibiotic-sensitive and treatment is timely. In contrast, chronic bacterial infections are inherently resistant to treatment. This is true even if organisms are antibiotic-sensitive, and high concentrations of drugs reach infection sites.

The *Pseudomonas aeruginosa* infections that afflict patients with cystic fibrosis (CF) are a prime example. Patients develop lung infections with this organism early in life, and the strain that establishes chronic infection persists in the airways for decades, despite countless courses of antibiotics. Even the combined use of two to three intravenous (IV) drugs only marginally reduces bacterial counts, and counts quickly rebound. This occurs not only in those with CF: chronic infections of the prostate, sinuses, wounds, medical devices, and other organs show similar resistance.

The mechanisms producing treatment resistance of chronic infections are poorly understood. In acute infections the antibiotic sensitivity of infecting bacteria, as assayed in clinical labs, correlates well with treatment response. Thus, if the pathogen is sensitive, the patient is likely to improve.

Pradeep Singh, MD

In contrast, the association between antibiotic sensitivity measurements and clinical response does not hold for CF

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Notes *from our Executive Director – Spring 2013*

Options for cystic fibrosis clinical care and treatments are expanding, and we want to keep you informed about this exciting progress. Bridget Barnes follows the development of Bronchitol, a medication from Pharmaxis which helps to clear mucus from the lungs. Clinical trials have been completed, and Bronchitol was considered by the FDA for US approval early this year. Although it is not yet available, there will be continued study of this new treatment option (see p. 3).

The number of centers that perform lung transplants for cystic fibrosis patients have continued to grow. In our cover story, Siri Vaeth-Dunn discusses key issues related to transplantation, as presented by Gundeep Dhillon, M.D., of the Stanford University Medical Center, at a recent CF Discovery Series™.

Dr. Dhillon describes several considerations when approving candidates for transplants, criteria for organ donations, potential complications during surgery and post-operative issues that impact best outcomes for patients. Adhering to a treatment plan before transplant, and setting up a strong support network for the months following surgery are important strategies for optimizing positive outcomes.

How important is body image for healthy outcomes and a better quality of life? As discussed on page 5, for some patients with cystic fibrosis the pursuit of unrealistic standards of "beauty" can be at direct odds with adherence to best CF health practices. Adolescent girls with CF may compromise their health in a quest to be thin, while the self esteem of adolescent boys may suffer due to the effects of delayed puberty. It is increasingly recognized that people with CF need to feel comfortable with their self-image to improve their quality of life.

Also in this issue, we share recommended "Books for the CF Journey." And, meet Maggie Williamson, an adult with CF, who shares her adventure of moving across the continent to pursue her dreams.

Exciting events are planned for the summer – mark your calendars! The CFRI Golf Tournament at Pasatiempo, our National CF Education Conference, and the Teen and Adult Retreat are all in place; we are very grateful to our sponsors who support these events.

Finally, your support of our Mothers' Day Tea (see page 15) is critical this year. New treatments promise a healthier future, and your contributions will lead to more of these, and to a cure. Please give generously. Your donations support us in our mission to fund CF research, to provide educational and personal support and to spread awareness of cystic fibrosis. Thank you!

Warmly,

Carroll Jenkins

Carroll

Save the Dates

CFRI 26th National CF Family Education Conference
August 2 – 4, 2013
Sofitel San Francisco Bay
Redwood City,

CFRI Teen and Adult Retreat
August 11 – 16, 2013
Vallombrosa Center
Menlo Park, CA

CFRI Golf Tournament
August 5, 2013
Pasatiempo Golf Club
Santa Cruz, CA

Bronchitol®: Latest CF Drug Waiting for FDA Approval

By Bridget Barnes

Recently approved in Australia and the United Kingdom as an effective treatment to clear mucus from the lungs of people with cystic fibrosis (CF), Bronchitol awaits approval in the United States by the Food and Drug Administration (FDA). In January, the FDA's Pulmonary-Allergy Drugs Advisory Committee (PADAC) met in Washington, D.C. to discuss Pharmaxis' new drug application for Bronchitol. At the public hearing, the PADAC received supportive testimony from CF physicians, including Drs. Felix Ratjen, Michael Boyle, and Moira Aitken; CF patients, Jerry Cahill and Ronnie Sharpe; as well as from organizational leaders, Carroll Jenkins (CFRI) and Bruce Marshall (CF Foundation). After hearing the testimonies and reviewing the data gathered from clinical trials, the PADAC ruled that the statistics regarding efficacy and safety failed to provide enough evidence, at least for now, to approve the drug. The committee did, however, acknowledge that for some patients, Bronchitol was very effective, with improvements in FEV1, and reductions in the significant time associated with cystic fibrosis treatments, thus enhancing adherence and quality of life.

In the next three months, the FDA will release "a complete response letter" addressing the application for approval of Bronchitol. At that time, according to Stephen Beckman, President of Pharmaxis, "the pathway forward for Bronchitol will take shape." He added, "Pharmaxis remains committed to getting Bronchitol into the hands of those living with cystic fibrosis." If approved, Bronchitol, which has an "orphan drug designation," could be available for people with CF six years of age and older. The FDA considers CF an orphan disease as it affects under 100,000 people in the United States.

Bronchitol is an inhalable dry-powder mannitol (a sugar alcohol) and osmotic agent, which increases mucociliary clearance. Once inhaled, Bronchitol's dynamic mode of action on mucus reduces its viscosity and enhances airway clearance. Among its other desirable features is the

ease of administration using a simple, disposable, capsule-based dry powder inhaler without the need for refrigeration, nebulization, equipment cleaning, or sterilization. As for where Bronchitol fits into the current "suite" of airway clearance therapies, Beckman said, "It's another key option in airway clearance. It's portable, discreet, no power source is needed, and a great alternative for patients who are highly active. It facilitates independence and compliance, and lessens the time burden of breathing treatments."

The FDA submission for Bronchitol was supported by two large Phase III clinical trials conducted with 600 patients with cystic fibrosis six years of age and older. Combined data of these two pivotal, randomized, double blind, controlled multi-center trials reported improvements in pulmonary function (FEV1) combined with reductions in pulmonary exacerbations.

The two studies were similar in design: 26 weeks in duration, they were comprised of patients from Europe, Australia, New Zealand, and North and South America. Patients treated with Bronchitol averaged a 7.3% improvement in lung function compared to baseline and a highly significant improvement compared to patients in the control group. The sub-group of patients who were also on DNase (pulmozyme) while on Bronchitol showed a 5.3% improvement from baseline that was again superior to the control group. Those who were not on DNase while on Bronchitol showed a 9.44% improvement from baseline and had superior lung function than the control group. The study also found a reduced incidence of pulmonary exacerbations for patients using Bronchitol. Adverse events reported in the Phase III trials were mild in nature. The most common side effect related to treatment was cough, occurring in 6% of the Bronchitol group and 3.3% of the control group. Sustained improvement in lung function was seen over an 18-month period. Other positive results of Bronchitol include a good safety profile and patient adherence.

Dr. Moira Aitken, Professor of Medicine at the Division of Pulmonary and Critical Care at the University of Washington in Seattle, and recipient of CFRI's 2012 Professional of the Year Award, was lead principal investigator on the second international Bronchitol Phase III trials and is excited by the findings. "Across both trials, inhaled mannitol was well tolerated and demonstrated an early and sustained improvement in lung function. The improvement in FEV1 was achieved on top of aggressive use of concomitant medications such as inhaled antibiotics and rhDNAse. These results, coupled with Bronchitol's novel formulation and portable dry powder inhaler administration, suggest that it will have a significant impact on CF patient well-being."

The positive results learned from the two large-scale Phase III trials of Bronchitol all support the efficacy, safety and tolerability of this new mucolytic therapy. The simplicity of the storage and delivery system, the lower cost, and the short five-minute administration time are attractive features of this new drug that may not only enhance compliance and utilization, but quality of life as well. Perhaps the most compelling testimony comes from an 18-year-old study participant who shared, "Bronchitol was a portable medication and that was certainly important…I could go on a trip and didn't have to drag along the nebulizer."

CFRI Executive Director Carroll Jenkins testified at the FDA public hearing on January 30. To view this hearing, go to: **http://tinyurl.com/d2wuy6j**

TRANSPLANTATION *(continued from page 1)*

have "rehabilitation potential," with acceptable nutritional status and adequate psychosocial support.

Dr. Dhillon presented "absolute contraindications" for transplant. These include an active malignancy other than skin cancer, untreatable advanced dysfunction of another major organ, coronary artery disease, chronic extra-pulmonary infections such as HIV, hepatitis C, and active viral hepatitis B. A history of non-adherence to medical therapies, an absence of a reliable social support system, active psychiatric illness and substance abuse are additional contraindications. Dr. Dhillon stressed that these rules exist, "not because we are mean," but because experience shows that outcomes are consistently poor for people with these issues.

When is the best time to be referred and listed for transplant? The answer to this question can be elusive. As Dr. Dhillon noted, "the natural history of the disease can be unpredictable." CF's clinical course "tends to have ups and downs, making it difficult to assess when something is an exacerbation that will respond to therapy, or a true decline that will not respond to antibiotics and therapies." Deciding when to list somebody for transplant is when the "less science, more art part of medicine comes in," said Dr. Dhillon, adding that there is a "transplantation window," when a person is "sick enough to need the transplantation, but healthy enough to survive the operation."

Ideally, noted Dr. Dhillon, the team at the transplant center will see patients early enough to get to know them before the operation. Criteria for a transplant evaluation referral include FEV1 (lung function) consistently under 30% of predicted, a rapid decline in FEV (especially in young women), an intensive care unit admission, increasing frequency of exacerbations, refractory and/or recurrent pneumothorax, and recurrent hemoptysis that requires medical attention. Any of these issues indicate that it may be time to start being seen in a transplant center. Dr. Dhillon stressed that this does not necessarily mean that transplantation is imminent, noting that the Stanford center has patients who have been followed for years who still do not need the surgery.

Patients are listed for transplant when they experience oxygen dependent respiratory failure, hypercapnia (when carbon dioxide rises in the blood) and pulmonary hypertension. There are often special considerations prior to listing, including respiratory infections common to those with CF. Only two or three of the large centers will transplant someone with *Burkholderia cepacia*, while others will not transplant those with multi-drug resistant *Pseudomonas aeruginosa*, or the newly emerging mycobacteriums. Liver disease is another contraindication. While still risky, patients on a ventilator who have already been fully evaluated and are on the waiting list may still be transplanted.

Gundeep Dhillon, MD, MPH

Dr. Dhillon noted that each center has its own specific criteria, and it is important to research these in advance.

In the past, patients received lungs on a first-come, first-served basis. People were encouraged to get on the list early, because the wait could be very long. In 2005, a new system was instituted nationally based on a Lung Allocation Score (LAS), which measures the most urgent need, as well as the probability of survival. The higher the person's LAS, the higher their priority for available lungs. Noted Dr. Dhillon, transplantation "is a waiting game." Listed patients must be available 24 hours per day, and are expected to be at the center within four hours of being called. Some patients move closer to the transplant centers; others make prior arrangements with air ambulances, so that when the call comes, they are ready.

Addressing the ideal criteria for organ donors, Dr. Dhillon said that they must be under 55 years old, with a clear chest X-ray, no evidence of chronic infection, and a normal bronchoscopic exam. Donors are matched to recipients based on blood type, size, and the absence of "recipient preformed antibodies to donor," which would lead to organ rejection.

Post-operative care is critical. "After the transplant happens, that is when the real work begins," said Dr. Dhillon. The surgery is probably the easiest part: we can control things." He noted that, "Patients must take care of the organs, or they will go bad." Immediately after transplant, patients receive immunosuppressants. Because these medications affect the immune system's ability to prevent opportunistic infections, antibiotics are used post-surgery, driven by pre-transplant cultures. Once discharged, most patients will take three different types of immunosuppressants and three infection prophylaxes, including anti-fungals, sulfa antibiotics, and anti-virals. Complete adherence to the prescribed medications is required.

At Stanford, most patients are discharged within 10 to 15 days. Close follow-up is vital, and it is hoped that patients will live within a 30 - 45 minute drive for the first three months, as they are seen weekly during this time. Recommended follow up includes regular monitoring of pulmonary function tests, chest X-Rays, and blood tests measuring creatinine levels, complete blood count, liver function and cytomegalovirus (CMV) infection. In addition, post-transplant bronchoscopy, surveillance with bronchoalveolar lavage, and trans-bronchial biopsy are used to monitor for rejection.

There are many complications associated with lung transplantation. Primary graft dysfunction occurs in 5% to 10% of patients. There are surgical, or mechanical complications, such as when sutures come loose, blood vessels are too loose or constricted, or there are nerve injuries. Other complications are immunosuppression-related, including infections or drug toxicity. Many infections come from the donor, although hospital-acquired *aspergillus* and

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Body Image and Cystic Fibrosis: Finding a Healthy Balance

Siri Vaeth-Dunn

Body image is an important issue for most everyone. As defined by Merriam-Webster, body image is "a subjective picture of one's own physical appearance established both by self-observation and by noting the reactions of others." This self-image can impact a person's self-esteem and in turn, his or her behavior. We are bombarded by media images that promote standards of beauty nearly impossible to attain. Cystic fibrosis (CF), with its systemic impact on the body, can cause greater challenges with body image. Whether it is low body weight, a steroid-induced "moon face," a gastronomy-tube, clubbed fingertips, or an ever-present cough, the manifestations of CF can deeply affect the body image and self-esteem of those who live with the disease.

In a society where female models are often so thin that they meet the clinical criteria for malnourishment, it is all too common for girls to internalize the message that being thin is desirable. According to Dr. Kathleen Kara Fitzpatrick, of Stanford University School of Medicine, somewhere between 40% and 60% of girls diet while in high school, and it is estimated that a minimum of 25% of adolescent girls and women have some form of eating disorder. The effort to be lean can have negative impacts on otherwise healthy girls, but for girls living with cystic fibrosis, a low body mass index (BMI) can lead to a correlating drop in lung function, with frightening outcomes.

Jonny Simpson, English Bodybuilder with CF

Researchers in the United Kingdom who reviewed a range of studies exploring body image and cystic fibrosis found that girls with CF tended to have a better body image than boys, but this was likely related to the fact that girls with CF tended to be thin, fitting with societal "ideals" for women. Conversely, boys had a more negative body image due to standards for men that include having larger body mass. Because of this, boys were more likely to adhere to nutritional recommendations. In another study, U.S. researchers found that 45% of girls with CF surveyed were "dissatisfied with their body size and desired a thinner figure," even when their BMI was lower than recommended.

The ideal BMI for those with cystic fibrosis may seem higher than the societal "norm," and many – especially girls – may be reluctant to achieve it. In a survey of 160 people with CF living in Scotland, between 12% and 18% of the female respondents said that they had tried to lose weight by omitting insulin, overusing laxatives, and not complying with supplements, while nearly 25% noted they had skipped enzymes to achieve this. As the authors of another study on eating disorders and cystic fibrosis noted, "Adolescents with CF have been found to subscribe to the same cultural values regarding shape and weight as their non-CF peers. It is clear that CF adolescents may pay with their lives for trying to live up to the(se) current cultural ideals."

Conversely, for many with cystic fibrosis, the extreme effort required to keep one's weight up can take away the joy of eating. Every visit to the CF clinic includes a weigh-in, and detailed questions about one's eating habits. These questions often extend into the daily routines of those with CF. Because of problems with nutrient absorption, people with CF often need to significantly increase the number of calories they consume. In both research studies and anecdotal stories, many people with cystic fibrosis describe the frustration of listening to people comment that they are "lucky" they can eat without gaining weight. As one study respondent wrote, "My general reply is, 'I'll swap my lungs for your fat!'"

When efforts to gain weight are unsuccessful, a gastrostomy tube (G-tube) may be placed. This is a tube placed directly into the stomach through a small incision, through which high-calorie liquid supplements can be administered, usually at night while a person sleeps. While the resulting weight gain can be esteem building, the visual presence of the tube can cause embarrassment. Similarly, the placement of

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CFRI a Winner in Nina Storey's Successful PledgeMusic Campaign

Singer-songwriter Nina Storey recently released her latest album, *Think Twice*, funded through a PledgeMusic drive in which fans pledged money to support the project in exchange for fun incentives (think signed CDs, personal messages on voice mails, bowling, and picnics with Storey under the Hollywood sign). As part of her campaign, Storey committed a percentage of the total to CFRI in honor of her fellow singer-songwriter, Tess Dunn, who has cystic fibrosis. In the years since she first met Dunn, Storey has served as a mentor to her, and has participated in numerous CFRI benefit concerts. The two recently performed together in Santa Cruz, after which Storey presented Dunn with nearly $1,000 in support of CFRI's services. Many thanks to Nina Storey, and to all who supported her PledgeMusic campaign!

photo by Francis Battaglia

Nina Storey and Tess Dunn

RESEARCH *(continued from page 1)*

Katie Hisert and Ben Staudinger are physician-scientists in training in the Singh Lab at the University of Washington in Seattle. Both are pulmonologists who specialize in the care of adult CF patients, and perform cutting-edge research on CF infections with the support of CFRI.

and other chronic infections. For example, early in CF infections, bacteria cannot be eradicated, even though they are highly sensitive *in vitro*. Later in disease, treatment can improve symptoms even though strains are resistant. These observations have spurred the search for new mechnisms that produce treatment resistance in chronic infections.

One possible mechanism is the biofilm growth mode. Biofilms are clusters of bacteria encased in an extracellular matrix, and biofilm growth produces marked antibiotic tolerance *in vitro*. Evidence for biofilm growth in CF consists primarily of the identification of biofilm-like cell clusters at chronic infection sites, and biofilm-like antibiotic resistance to chronic infections. However, many labs have observed that *P. aeruginosa* isolates from chronic CF infections fail to form biofilms when grown in the lab. Thus, the contribution of biofilm growth to treatment resistance *in vivo* remains unclear.

Another mechanism to explain resistance in chronic infections is antibiotic-tolerant "persister" cells. Persisters are the small fraction of bacterial populations that are phenotypically tolerant to bactericidal antibiotics. The exact mechanism of persister-cell tolerance is poorly understood, but it may be linked to low metabolic activity of some subpopulations. *P. aeruginosa* isolates from advanced CF lung disease were recently found in large numbers of persister cells. Additional direct evidence linking "persisters" to resistance in chronic infections could advance this idea.

Finally, recent work shows that extensive population diversity evolves in CF *Pseudomonas* strains during infection. Population diversity may contribute to treatment resistance if subpopulations with any kind of antibiotic tolerance phenotype increase significantly during treatment. The relative contribution of these proposed mechanisms to antibiotic tolerance in CF infections (*in vivo*) remains unknown.

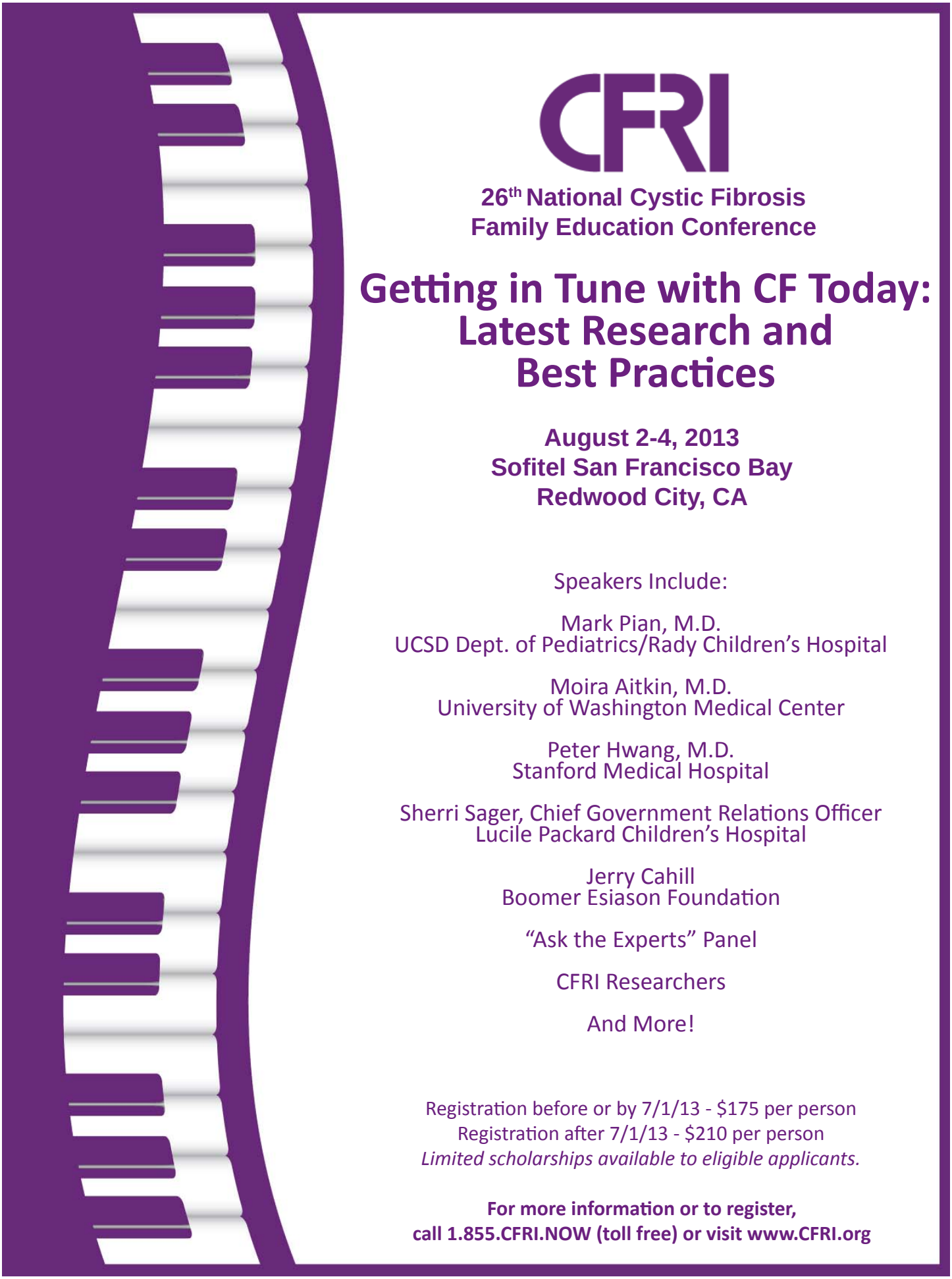
Progress in understanding treatment resistance in chronic infections has been stymied for several reasons. First, experimental infections that model chronic infections and their resistance have been difficult to generate.

Second, the bacterial mechanisms producing resistance remain elusive. As noted previously, the long-standing assumption that antibiotic sensitivity is a key parameter affecting treatment outcomes in chronic infections is likely incorrect.

Third, identifying bacterial functions critical for resistance is hindered by the fact that bacterial gene and protein expression are highly dependent on conditions. Thus, bacterial physiology changes rapidly when cells are removed from infection sites, and this could alter expression of functions important *in vivo*. Together, these limitations have made studies of *in vivo* resistance mechanisms very difficult to perform.

Our lab and others are using new approaches to better understand treatment resistance in CF. One approach is to use advanced technology to measure the proteins produced by bacteria living in the lungs of CF patients before and during antibiotic treatment. These studies could allow us to identify the functions that mediate bacterial antibiotic resistance *in vivo*.

Another approach exploits changes in the composition of the P. *aeruginosa* population that occur during treatment to identify resistance functions. Advances in understanding treatment resistance in chronic infections are essential to making progress with CF lung infections. New technology, new thinking, and a new generation of researchers make prospects for progress very bright.



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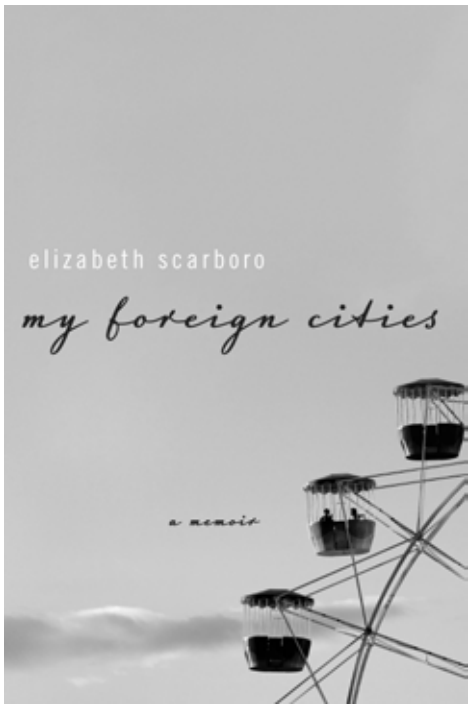
My Foreign Cities: A Much Needed Read for the CF Community

By Anabel Stenzel

In the prologue to Elizabeth Scarboro's memoir, *My Foreign Cities,* she and her husband Stephen, who has cystic fibrosis (CF), climb to a rock ledge that perches thirty feet above a swimming hole. Stephen is relaxed as he steps off the rock and plunges to the water below. But Scarboro hesitates, noting, "It was a delicate matter, to land in the spot where the river ran deep. Too far and I would hit the bank, not far enough and I would tumble down the rocks." Ultimately, she steps off the ledge and plummets to join her waiting husband. This literal leap to join Stephen in the narrow depths of the river serves as an excellent metaphor for her life with him, and their shared life with CF.

My Foreign Cities, is a much-needed book for the CF community. Sharing her experiences as a teenager who falls in love with a young man with CF, and their 10-year journey as life partners, Elizabeth's writing deeply moved me. Rarely have I come across a story so powerful and insightful in providing the spouse's viewpoint on the CF experience. The book is beautifully written, humorous at many points and painfully honest at others, and gives the reader an in-depth personal account of the joys and frustrations of loving a spouse who happens to have CF. Unlike some CF-related books, it did not leave me feeling down, critical or frustrated, but rather infused with love and appreciation for those who are able to love people with CF unconditionally, "for better or for worse, in sickness and in health."

This is a story of young love and devotion of two best-friends-turned-lovers surviving the challenges of young adulthood together despite the horrendous nature of cystic fibrosis. Elizabeth's writing style brings the reader deeply into her experience and emotions and allows the reader to get to know Stephen in an intimate way. She touches upon important issues facing CF adults, such as the fine balance between pursuing career/educational goals and health, the risks of addiction from prescription pain medications, the desire to



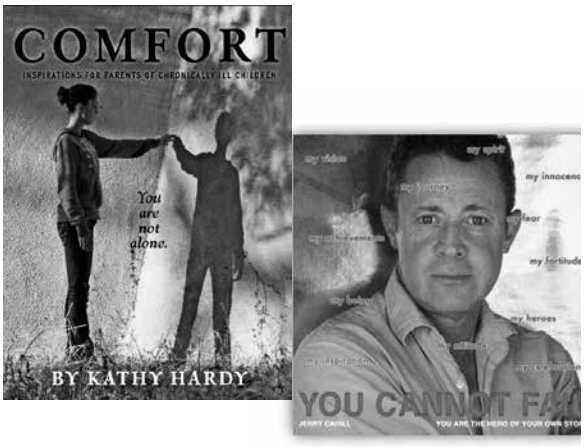
delay hospitalizations because of knowing too well the song-and-dance red tape that awaits once admitted, and that final surrender to medical care when illness takes over and there is no choice.

This book is essential reading for adults with CF, to recognize and appreciate the experience of our supportive life partners, and the joys and tears of loving someone until the end, and beyond. As a married person with CF, I saw myself in Stephen and can only imagine the frustrations and joys that my own spouse experiences. I felt that Stephen was a kindred spirit, with whom I share some common characteristics that result from being raised with CF. Like Stephen, I can be obstinate, am highly determined, possess a keen self awareness of my physical well-being, and am all too aware of my limited time on earth. Like so many of us with CF, Stephen grabbed onto life and cherished every day, even if it meant putting off that visit to the ER, or minimizing CF complications like hemoptysis and lung collapses, despite the concern of those around him. More than once I identified with Stephen–and probably would have chosen the same course of action as him– in balancing health, education, job pursuits, and family needs. Only through reading *My Foreign Cities,* did I truly grasp how frustrating, yet strangely comical, it must be to witness the choices made by those of us with CF from our loved ones' perspectives.

Elizabeth eloquently shares the reality of arranging life plans to fit Stephen's health and medical schedule, and points to the valuable life lessons that she received despite some sacrifice. I strongly recommend this book for those with CF and their partners/spouses/spouses-to-be. Those of us with CF will have greater awareness of the personal struggles and joys of our partners, and our partners will likely feel that they are not alone in their experience. You may laugh, you may cry, you may reflect on your own experience. Most likely, you will come away with appreciation that love is not chosen – it just happens. And when it does, it lasts forever.

For more information, contact Elizabeth Scarboro via Facebook at **https://www.facebook.com/myforeigncities?fref=ts**

My Foreign Cities can be ordered on Amazon.com and will be in bookstores this April.



Two Books for the Journey

These two books offer thoughtful insight and inspiration to the CF community:

Comfort by Kathy Hardy helps others understand the day-to-day-struggles and joys involved in raising a chronically-ill child, and the comfort one's personal faith can provide. Kathy is the mother of eight children, three of whom have CF.

You Cannot Fail by Jerry Cahill provides inspiration to face difficult days and life-changing decisions with strength and grace. He encourages us to recognize our inner strength. Jerry will be speaking at the CFRI Conference in August.

My Big Adventure: Moving 2,000 Miles to Chase a Dream

By Maggie Williamson

I stepped off the plane at the San Francisco airport in late spring of 2012, and found myself in familiar territory, in an unfamiliar situation. I had just moved 2,000 miles away from everything I knew. My husband Tom and I had decided months before that we wanted an adventure, and we both loved California for many different reasons. I had never lived outside of the land of Lincoln, the great state of Illinois; it was the only place I had ever called home.

In 2006, I was introduced to the CFRI Teen and Adult Retreat, and it was one of the best events I had ever attended for young adults living with cystic fibrosis (CF). I knew from then on that not only did I need to go to Retreat every summer; I also needed to move to California. This was only a pipe dream, as I had always thought CF would hold me back from ever moving away from my support system, my family.

After Tom and I had our "destination wedding" in San Francisco, we talked seriously about our dream to move and experience new things. Only a few months after our wedding, the stars seemed to align and everything pointed towards us moving out west.

My whole life I had been afraid of change. CF was always changing my health status and my plans, and it made me scared of the unknown. But picking up my life and moving across the country made me feel at ease. I was at peace with this change; maybe because it was a dream of mine, or maybe it was because I wanted to prove to myself that CF was not going to stand in the way of my dreams. I wanted to prove I could do anything anyone else could.

Maggie and Tom Williamson

We had our bags packed and knew roughly our plans for residency once arriving in the Bay Area, but nothing was set in stone completely. I am a real planner, and this move was planned as well as it could be from far away, but Tom and I had never seen the apartment we rented on Craigslist. We had no idea if our furniture would fit, and we had no idea how steep the hill was on our street. We arrived at our new apartment twenty minutes before the moving truck got there, and had to decide right then where all of our belongings would go. There was a bit of anxiety that came with this experience of not really knowing what we were getting ourselves into, as online photos and Skype video-chats with our landlord took us only so far in preparing for our new town and home.

I had not even decided on a new CF center before I left Chicago. I had done my research, but wanted to wait until I arrived in California to see the various centers before fully making my decision. It felt very strange not being under anyone's care for a whole two weeks! After researching the options, I chose my adult center and am really happy with my selection.

Change is not always easy, and realizing one's dream can be a challenge, but this challenge was one I was ready to take head on. Having my husband by my side made this transition easier, but not having family close by has been difficult at times. Modern technology has helped ease the distance, and I am grateful for the support of my family and friends back in Illinois.

Cystic fibrosis is a challenging disease to live with. I have always had to adjust my life for hospitalizations, more treatments, and new routines, but I was not ready to let it stand in the way of my biggest dream. I wanted to be able to say that at least I tried and gave it my all. I can now say, I am succeeding in this unfamiliar situation of living in a new place, and am enjoying every minute of my decision to not let my disease hold me back.

CF and Cancer

By Laura Tillman

While improvements in the treatment of cystic fibrosis (CF) have greatly improved the quality of life and survival of people living with CF, there is an emergence of new symptoms and diagnoses as the effects of CF are manifested in other parts of the body. Some of the conditions now seen in CF are specific to the underlying disease process, others are side effects from taking long-term medications, and some are associated with patients' advanced age.

Cancers of the digestive tract, colon and pancreas were previously rare in people living with CF, as patients were unlikely to live to an age at which these malignancies would occur. Recently, these types of cancers are being seen more frequently and research has shown that their incidence is higher in patients with CF as compared to the general population. The reason for the increased risk of cancer of the intestinal tract in patients with CF is unknown, but various theories have been proposed. Possible explanations relate to the effect that CF has on organs of the intestinal tract. Barrett's esophagus has been reported in patients with CF and is known to be associated with cancer of the esophagus.

Liver cancer and cancers of the biliary tract often occur in patients with gallstones, which are frequently found in patients with CF. The presence of excess fat in feces, decreased expression of mucin genes (MUC3, MUC11 and MUC12), and malabsorption that leads to nutritional/antioxidant deficiencies have also been linked to the development of cancer. Additionally, an increase in the incidence of digestive tract tumors is seen in patients who have undergone transplant procedures, which is likely related to ongoing and harsh immunosuppressive therapy.

The American Cancer Society, as well as the Cancer Treatment Centers of America, publishes information in which risk factors for small intestine cancer are discussed. Both state that small intestine adenocarcinoma is uncommon but that CF is one risk factor: "People with CF have an increased risk of adenocarcinoma of the ileum," according to the American Cancer Society.

http://tinyurl.com/ba3vxws, http://tinyurl.com/auqd92p and http://tinyurl.com/a3l4lyn

Other Articles that Describe Cancer Risks in CF Patients:

The Epidemiology of Cancer of the Small Bowel. Neugut AI; Jacobson JS; Suh S; et al. Cancer Epidemiol Biomarkers Prev. 1998 Mar;7(3):243-51. The authors discuss various risk factors for small bowel cancer, including cystic fibrosis.
http://tinyurl.com/a7g6rb6

Association of Pancreatic Adenocarcinoma, Mild Lung Disease and Delta F508 Mutation in a Cystic Fibrosis Patient.
Tsongalis GJ; Faber G; Dalldorf FG; et al. Clin Chem. 1994 Oct;40(10):1972-4. At the time of publication, only five cases of pancreatic cancer in CF patients had been reported. The authors suggested a possible association of CF with this type of malignancy.
http://tinyurl.com/bdm49ow

The Risk of Cancer Among Patients with Cystic Fibrosis.
Neglia JP; FitzSimmons SC; Maisonneuve P; et al. N Engl J Med. 1995 Feb;332:494-9. This is the largest study to assess the risk of cancer in patients with CF over a seven-year period. The overall risk of cancer was similar to that of the general population, but the risk of cancer of the digestive tract was significantly increased in CF patients and was most dramatic in those aged 20-29 years for cancer affecting the pancreas, esophagus and bowel.
http://tinyurl.com/a2jjcf6

Cancer Risk in Nontransplanted and Transplanted Cystic Fibrosis Patients: A 10-Year Study.
Maisonneuve P; FitzSimmons SC; Neglia JP; et al. J Natl Cancer Inst. 2003 March; 95(5):381-7. This comprehensive article shows the risk of cancer is particularly elevated for the small bowel, colon, and biliary tract in CF patients. Results also suggest that CF, when initially diagnosed due to failure to thrive, is associated with a higher risk of digestive tract cancer and also indicates that the risk of digestive tract cancer appears more pronounced after transplantation.
http://tinyurl.com/aw3y27s

Cancer Risk in Cystic Fibrosis: A 20-Year Nationwide Study From the United States. Maisonneuve P; Marshall BC; Knapp EA; et al. J Natl Cancer Inst. 2013 Jan;105(2);122-9. This article states that CF patients have an increased risk of digestive tract cancer, lymphoid leukemia and testicular cancer, while the risk of melanoma remains low.
http://tinyurl.com/csstljk

Intussusception and Metastatic Colon Cancer in an Adult with Cystic Fibrosis.
Gilchrist FJ; Jones AM; Bright-Thomas RJ. J R Soc Med. 2012 Jun;105 Supp 2:S40-3. An extremely comprehensive article describing the first reported case of intussusception in a patient with CF in which a malignancy has been identified as the cause. It also highlights a number of other important clinical, psychological and social issues related to the disease.
http://tinyurl.com/bydorof

Risk of Pancreatic Cancer in Patients with Cystic Fibrosis.
Maisonneuve P; Marshall BC; Lowenfels AB. Gut. 2007 Sep;56(9):1327-8. The authors found that pancreatic cancer seems to be age-related and therefore relatively rare in younger CF patients. While the risk of this type of malignancy is five to six times greater than in the general population, it still is not significant in the CF population.
http://tinyurl.com/akle927

A Cohort Study of Cystic Fibrosis and Malignancy.
Sheldon CD; Hodson ME; Carpenter LM; et al. Br J Cancer. 1993 Nov;68(5):1025-8. Based on the researchers' findings, as well as previous case reports, cancer of the pancreas and of the terminal ileum may be linked to CF.
http://tinyurl.com/affzjzk

(continued on page 15)

CFRI Teen and Adult Retreat

August 11th - August 16th, 2013 at the
Vallombrosa Center in Menlo Park, CA
www.cfri.org/TeenAndAdultRetreat.shtml

- **Meet some great friends.**
- **Feel that you're not alone.**
- **Learn more about CF self care.**
- **Experience a place for hope & healing.**

Who Can Come: Teens and adults 15 years and older with cystic fibrosis*, their family members, friends and health care providers.

Purpose of the Retreat: The retreat provides a safe and welcoming environment aimed at enhancing positive coping skills, social support and education for people who share common experiences with CF.

What We Do: Activities that promote health include daily exercise, arts and crafts, rap sessions, and educational workshops with guest speakers. Fun group-bonding activities include a talent show, games, and just hanging out getting to know others.

Cost: $85 per person for the entire week. Daily fees are $15 per day for visitors or $10 per meal. Hotel and transportation fees are not included. On-site private rooms/baths are available at $55 per night. Scholarships are available.

Safety: All people with CF are required to comply with CFRI's cross-infection guidelines and on-site protocols. A medical advisor is available at the Retreat. Participants with CF must 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 the retreat.

CFRI™ | **We'd love to see you!**

For more information: 650.404.9975 | www.CFRI.org | cfri@cfri.org
http://www.facebook.com/home.php#!/group.php?gid=2342719557

BODY IMAGE *(continued from page 5)*

a port for IV antibiotics can also lead to self- consciousness, depending on where it is placed. In the Scottish survey of 160 people with CF, 60% of those with a port commented that it impacted their body image. Most responded that placement location was key, and that patients with CF should actively express where they prefer the port to be placed.

Adolescence can be one of the most difficult periods for positive body image, and cystic fibrosis can add unique challenges. Many with CF experience puberty that is delayed, likely caused by low body weight which delays hormonal activity. While this is temporary, it is important to recognize the anxiety and depression delayed puberty can cause. The body image of some men with CF may be impacted by the reality that they are unable to father children without medical intervention. Girls with CF may experience chronic yeast infections due to antibiotic use, and suffer in silence due to the embarrassment they feel.

The chronic cough that most people with CF live with can significantly impact self-esteem and body image. Many people with CF have experienced unwanted comments from strangers about their cough, who assume they are sick with a cold, or who joke about a "smoker's hack." Despite knowing that coughing and clearance is critical for the lungs, many people with CF try to stifle their cough, to avoid the attention it draws. For girls and women, chronic coughing places stress on the pelvic floor muscles that can lead to another source of shame – urinary incontinence – which studies have found to affect over half of young women with CF. Studies have also found that although there are treatments available, urinary incontinence is a source of tremendous embarrassment, and is therefore underreported to CF caregivers. In one study conducted in Australia, one-third of respondents said that this issue impacted their social life, and nearly half of the teenaged girls had not told anyone about the problem.

The Scottish survey results were published in 2010 as a booklet, "Cystic Fibrosis and Body Image," by Dr. Jacqueline Didsbury, a psychiatrist, and Emily Thackray, the chair of an organization that promotes organ donation. Both women have CF, and both have had double lung transplants. In addition to the issues referenced above, their survey explored people's embarrassment over clubbed fingers, bloated stomachs, osteoporosis, weight gain and facial puffiness from steroid use, the use of oxygen in public, noticeable salt crystals when perspiring, and scarring from CF-related surgeries, such as those from meconium ileus and transplantation. In addition to reporting survey results, the authors –as well as the respondents – share strategies to address many of the issues. Wrote the authors, "We know that it is not only physical health that is important, because what is the benefit of good health when we do not feel comfortable with our own image?"

Both patients and care teams must be encouraged to explore the many ways in which cystic fibrosis impacts self-image, as having a positive body image directly impacts both health outcomes, and quality of life.

TRANSPLANTATION *(continued from page 4)*

candida are common. It is now understood that acid reflux is more likely to damage transplanted lungs, as are respiratory viral infections. Dr. Dhillon noted that his center is "very aggressive" about treating these, due to concern they will damage the graft down the road.

Bronchiolitis Obliterans Syndrome (BOS) affects over half of transplant patients after five years, and nearly 80% after ten years. Chronic allograft dysfunction (rejection) usually appears as a decline in FEV, and treatments are not usually effective, leaving re-transplantation as the only option. Dr. Dhillon noted that outcomes are not as good for re-transplantation.

Currently, the one-year survival rate for first-time lung transplant recipients is 90%. Almost half of these patients will develop high blood pressure, and 80% will be on high blood pressure medications within five years due to the immunosuppression drugs. About half a percent will develop kidney disease, half of the patients will develop hyperlipidemia, and approximately one-third will develop diabetes.

Cancer risk increases as well, due to the immunosuppression drugs that interfere with the body's ability to fight cancer. Of those who do develop cancer post-transplant, for most patients it is skin cancer. There is also some increase in transplant-related lymphomas. The team at Stanford is currently discussing potential protocols for colon cancer screening due to increased risk.

Dr. Dhillon stressed that CF patients still need to take care of the issues they had prior to transplant, including sinus disease, pancreatic insufficiency, psychosocial issues, gastroesophageal reflux disease (GERD), and distal intestinal obstruction syndrome (DIOS). Some of the post-transplant medications can make conditions worse, such as osteoporosis and diabetes.

Having a strong post-transplant support system is critical. Said Dr. Dhillon, "It truly takes a village to go through this operation, and almost nobody can go through it alone." Transplant patients must have a caretaker with them 24 hours per day, seven days per week. Dr. Dhillon noted that depression often emerges in patients between six weeks and six months, as medications can unmask depression in people who are predisposed to it. "First there is euphoria that you have survived, then the medications (prednisone) taper down, and most people need support. Beyond social workers and psychiatrists, people need help from loved ones." It is estimated that over half the patients post-transplantation are on antidepressants.

Lung transplantation outcomes are improving, with 50% of patients living for 6.5 years. Dr. Dhillon credits the increased survival rates to a better understanding of nutrition, diabetes and high blood pressure, as well as improvements in surgical techniques.

Most importantly, stressed Dr. Dhillon, after successful transplantation, patients tend to have "very good functional outcomes, with no activity limitations." This opens many opportunities to those whose lives were dramatically limited by their lung disease.

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September 1, 2012 – January 31, 2013

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29th Annual CFRI Golf Tournament

Pasatiempo Golf Club
Santa Cruz, California
Monday, August 5, 2013

Join us at the award-winning, "Top 100 Course" Pasatiempo Golf Club in Santa Cruz, California for a wonderful day of golf, great food and good friends. Tournament registration includes lunch, wine reception, silent and live auctions, and a delicious celebration dinner – all with stunning views of the Monterey Bay. Proceeds from this fundraiser support Cystic Fibrosis Research, Inc.

To register, or to inquire about sponsorship opportunities, contact Scott Hoyt at dscott.hoyt@gmail.com. We look forward to seeing you there!

"Becca's Team" 2012 tournament

Spread Awareness and Hope with the Mothers' Day Tea!

The annual Mothers' Day Tea (MDT) provides CFRI with its largest source of funding to support our services to the cystic fibrosis (CF) community. This year's MDT brochure features Jamie Beasley Killinger, a beautiful little girl who was diagnosed with cystic fibrosis when she was two weeks old. Jamie is a happy, active toddler who is growing and thriving thanks to an early diagnosis and the support of her extended family.

Jamie

By signing up to be a Tea Sender, you will join a dedicated group of parents, family members, friends and those living with CF who send tea invitations to their family, friends and colleagues. These invitations encourage supporters to take a moment, enjoy a cup of tea, and think of those living with cystic fibrosis. Your participation in the Mothers' Day Tea will help CFRI expand its funding of cutting-edge research, and continue to provide education and support to the CF community, free of charge. These programs improve the quality of life of those with CF, as well as their families and caregivers.

Our goal this year is to raise $200,000. Barbara and Jim Curry, grandparents to Cameron, who has CF, once again graciously hosted the Mothers' Day Tea kick-off reception at their home. R.C. Bigelow, Inc. has generously provided the English Breakfast Tea included in our invitations. Dedicated CFRI supporter Pat Flynn donated a beautiful teapot that was raffled at the MDT Kickoff. A long-time tea sender Ethel Harder was the lucky winner. Congratulations, Ethel!

If you want to become a Tea Sender, now is the time! For complete packets including invitations, envelopes, tea bags and mailing labels, please call our office at 855.CFRI.NOW (toll free) or email: **CFRI@cfri.org.** You can also order online at **www.CFRI.org**.
Thank you!

CF AND CANCER *(continued from page 10)*

Colon Cancer in Lung Transplant Recipients with CF: Increased Risk and Results of Screening.
Meyer KC; Francois ML; Thomas HK; et al. J Cyst Fibros. 2011 Sep;10(5):366–9.

The authors report that the risk of colorectal carcinoma occurs at a younger age in lung transplant recipients with CF. They surmised that this was due to intense immunosuppression drugs and suggest that lung transplant programs should be screening CF lung transplant patients early on, regardless of age.
http://tinyurl.com/argozpr

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It's easy! Follow the link on our website to find instructions and the appropriate form. Your vehicle will be picked up, sold at auction and a portion of the proceeds will be sent to CFRI. That's all there is to it!

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Need more information?
Contact JoAnn Davis at 650.404.9979 or via email: **jdavis@cfri.org.**

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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.