

The Role of the Basolateral Chloride Channel in Absorbing Salt

By Beate Illek, Ph.D., Assistant Research Scientist,
Children's Hospital Oakland Research Institute, Oakland, CA.

At birth, fluid filled airways switch from chloride secretion to sodium absorption, resulting in the rapid absorption of airway fluid. Adult airways are lined with a thin film of liquid, which is essential for the normal function of the airways such as ciliary beating, mucociliary clearance and bacterial killing. Defective salt absorption in CF results in abnormal salt content of the airway surface liquid, which in turn is believed to block the action of salt-sensitive antibiotics. My research studies the basic mechanism and underlying chloride channels for the transcellular transport of salt in normal and CF airways.

The absorption of salt and fluid follows the basic mechanism that was proposed over 50 years ago in Denmark by Hans Ussing. He studied the absorption of sodium across the skin of the frog and found that the entry of sodium ions occurs across the apical membrane (the membrane that is exposed to the outer environment) via an epithelial sodium channel. Sodium uptake into the cell is driven by both the chemical (low intracellular sodium concentration) and electrical gradient (the intracellular space is negatively charged, which attracts the positively-charged sodium ions). Sodium is then extruded across the basolateral membrane (the membrane that is facing the bloodside) by the sodium/potassium-ATPase, which pumps sodium out of the cell in exchange for potassium.

The sum of net transepithelial ion movements is always electrically neutral. In other words, sodium has a partner that hitches a ride. The positively-charged sodium ion is accompanied by the negatively-charged chloride ion that may travel across the cell (transcellular), or between the cells (paracellular), or both. Our working hypothesis was that chloride is absorbed across the cell and therefore has to cross both apical and basolateral cell membranes. The chloride entry step is mediated by CFTR chloride channels and is regulated by intracellular cyclic AMP. However the route for chloride exit was not known.

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I explored the possibility that chloride is conducted across the basolateral membrane via chloride channels. This was done by studying the chloride transport properties of the basolateral cell membrane in intact tracheal sheets from humans and cows. I used a pore-forming agent to render the apical cell membrane permeable, which allowed me to isolate and measure chloride currents across the basolateral membrane in Ussing chambers. I found a novel

chloride conductance in the basolateral membrane, which was regulated by cell volume and cyclic AMP and blocked by a variety of chloride channel blockers. We further determined the biophysical properties of this basolateral chloride conduc-

tance by patch clamping the basolateral pole of freshly isolated ciliated cells, which we obtained by nasal brushings from normal volunteers in our lab. We found two types of chloride channel populations, which were distinct from CFTR and resembled chloride channels of the recently cloned CIC family. Using molecular biology techniques we showed the molecular expression of messenger RNA for CIC-type chloride channels in human airways. From our findings we proposed a novel model of epithelial chloride transport: in absorbing epithelia, the basolateral chloride conductance supports transcellular chloride absorption, whereas in chloride secreting epithelia, the basolateral chloride conductance attenuates chloride secretion.



Beate Illek, Ph.D., speaking at CFRI's Conference 2000 on her research efforts.

Future research in my lab will study the regulation of basolateral chloride channels in cystic fibrosis cells and their regulatory role on transepithelial chloride transport. Our findings may eventually point to a novel therapeutic strategy that may circumvent the CFTR defect.

I am very grateful to CFRI and all its members for funding this research. ■

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Letter from the Executive Director

CFRI has become a center of HOPE AND STRENGTH for not only our local neighbors, but for people throughout this country. In 1975, a group of dedicated activists decided they wanted more for their CF community. Their dreams and vision gave birth to CFRI. Their actions determined our future and began our 25 year history. We are deeply grateful to Maxine Eggerth, George and Anne Graham, George and Marilyn Hauptert, Donald and Merrilyn Holmes, Chuck Nelson, Elaine Peterson, Jackie Phelps, Robert C. Stewart (Sr.) and Bernie Stewart, Robert Stewart, Jr., and Laurie Stewart, Mary and Roy Tripp, Heath and Gail Wakelee, and Pat Ware for the courage, creativity, toil and dreams it took to create CFRI.

On our 25th anniversary, I am reflecting on this past year and our accomplishments. I feel proud of the quality of the research that we have funded this year. I feel successful when I see CF adults laughing with friends at the Conference or Retreat. I feel moved when parents of CF children gather in the office to mail fliers or offer their volunteer services and pause to share intimate stories about raising youngsters with CF. I feel honored when someone lets us know how one of our services, the Conference, the Retreat, or the Newsletter, has had an impact on their life. And I feel ecstatic when a donor or sponsor contributes to the mission of CFRI. At our gatherings, I can feel the love in your hearts. And I can feel the disappointment and frustration at how long it is taking us to find a cure for this disease.

Just the other day, I was asked what is the secret of CFRI's success? My response was that CFRI is a group of individuals who love people and life itself. They are determined to live life to the fullest despite statistics, complicated medical regimens and frequent health problems. When we REACH OUT and touch each other with our courage, and share our stories with empathy and compassion, we can hold out for that cure just looming around the corner.

This season is the best time of year to share the love in your hearts and to conquer our fears and disappointments. Let us be generous this season. Let us really look into our hearts and see what we can give to further the mission of CFRI and find that elusive cure. For those of you with small funds, your volunteer time is equally valuable to us. We have so many committees needing man and woman power. I've also compiled a wish list this season. We need additional computer memory or funds to help us build our computer system. Many of you may know companies or individuals that would like to give a major gift this season. Please think of CFRI! Your involvement is critical. **YOU'RE THE SECRET TO OUR SUCCESS.**

For myself, my goal is to continue to learn as much as I can about cystic fibrosis so I can be the appropriate advocate to carry the torch. If I can educate others and open the eyes and hearts of people around the world to the needs of this community, then my gift to CFRI will be adequate. This is a serious mission and I will do what I can to be the best I can be for you. Have a Wonderful Holiday!

Calvinia Williams,

Executive Director, CFRI



Founders in attendance at the 25th Anniversary Celebration at Conference 2000 from left to right: Chuck Nelson, Elaine Peterson, Maxine Eggerth, and Heath and Gail Wakelee.

CFRI Hosts 250 People at Our 13th Annual Educational Conference!

By Kathleen Flynn

Editor's Note: Due to space constraints, several talks will only be available on tapes or on our website: www.cfri.org

CFRI's Conference 2000 was truly an amazing experience for all involved. The speakers, sponsors, facilitators, volunteers, staff and attendees all pulled together for one of our finest educational events yet. In general, attendees relished the opportunity to gather with their friends in a supportive and compassionate environment, all the while checking in on each other's health, each other's children's health and all the health care issues that go with it. They participated in the latest educational discourses on cystic fibrosis, shared their experiences in support groups, and fell into bed exhausted at night not sure they could even remember their own names. I thought I'd never say it...but I can't wait to do it again next year!

CFRI was very blessed to have the support of an excellent audio-visual committee. John and Margaret Myers did almost all of the videotaping at the conference. Scott Wakefield, Frank Flynn and Phillip Howell provided invaluable audio and videotaping support. John Myers, Joe Lindic, and Scott Wakefield have also taken on the process of duplication of all the videotapes so that CFRI could offer tapes of Conference 2000 to our membership at an affordable rate. ■

Excellent Photo Display by Michelle Compton

Thanks to Michelle Compton, a post-transplant CF adult, for sharing with us her beautiful images of adults with cystic fibrosis and her moving poetry. We are truly blessed to have her in our community.



"Through the Looking Glass," Michelle Compton's fabulous images on display at the Conference. Special thanks go to the Compton family, Margaret Myers, and Beth Sufian for help with the displays.



For People with CF and their Families

The Roller Coaster Ride Toward Transplant

While many of us were making our travel arrangements for the conference, **Travis Evans-Hugen**, age 23, was undergoing his transplant surgery. This roller coaster event began appropriately enough, at a popular water theme park, Water World U.S.A., in Concord, CA on August 5th, 2000, an hour's drive from Travis's home in Tracy. Travis wore his pager to the theme park and carried it with him throughout the morning, as he had done religiously for the two years he had his pager. Even when he traveled, Travis's family figured out a way to have his pages forwarded to his long-distance destination. Yet, until that day, Travis had never received a page.

The family had packed a picnic lunch and after a morning on the water rides they went out to the car to eat. After lunch, they thought they would only be in the park for another hour or so, so they decided to leave all of their belongings, including Travis's pager, in the car. Unbeknownst to the family, not five minutes after they left the car the pager went off. When the family returned almost three hours later and saw the page, they were stunned. They immediately called the hospital only to find out that the lungs had been offered to another family. The Evans-Hugen's were devastated. Not knowing what to do, and still in complete shock, the family returned home and called the hospital back to see if the other family had accepted the lungs (they had) and to let the doctors know where they could be reached.

One doctor said that he didn't want to get the family's hopes up, but there was a possibility of another set of lungs. The doctor told the family that he had only just received the call and knew nothing about the quality or match of the lungs, but that he

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Travis Evans-Hugen

Golfers Know How to Have a Good Time!

Special thanks go to Bob and Nadine Mackay for hosting the Robert J. Mackay, Jr. Memorial Golf Tournament, held August 25th, 2000 at the Santa Teresa Golf Club in San Jose, California. The completely sold out event netted close to **\$36,000!** The Mackays put on this annual tourney in memory of their son.

CFRI 2000 Golf Tournament Raises Close to \$60,000!



CFRI's annual fundraising event, the CFRI Golf Tournament, was held on Monday, August 21, 2000 at the breathtaking Cinnabar Hills Golf Club in San Jose, California. We had a record turnout with over 150 golfers, all of whom enjoyed a day at the 27-hole golf course, the fantastic dinner and auction afterwards. We had a fantastic record year, netting close to \$60,000 for cystic fibrosis research and education! Thanks to all the sponsors, prize donors, and Scott Hoyt, General manager of Cinnabar Hills Golf Club and this year's Golf Committee Chair. Special thanks also go to the Cinnabar Hills Golf Club for hosting the event.



From left, Ralph Swanson, Mike Roanhaus, and Scott Hoyt (Chair of the Golf Committee) in action at CFRI's 2000 Annual Golf Tournament.

CFRI Conference 2000: The Current Status of Glutathione (GSH) Research

Speaker: Rabin Tirouvanziam, Ph.D., Post Doctoral Fellow at Cystic Fibrosis Research Lab, Stanford University, Stanford, CA.
Summarized by Eric Lindmeier

In doing research on cystic fibrosis lung disease, Dr. Rabin Tirouvanziam encountered a puzzle. Common theory suggests a specific course for the disease to follow. First, the lungs are obstructed with mucus, which then leads to infection, which in turn causes inflammation. Inflammation is a normal defense mechanism in the lung, assisting in infection control, but in CF, inflammation actually contributes to damaging the lungs rather than preventing the damage. This process begins shortly after birth. The puzzle arises in the current research that shows that newborns without detectable infections still have inflamed lungs. How can an uninfected lung be inflamed? Could the inflammation of the lung be the first step towards obstruction and infection instead of vice versa? What could bring the lung to a normal state, and would that prevent the spiral of damage from occurring?

To delve deeper into solving the puzzle of inflammation, Dr. Tirouvanziam decided to focus on the role of neutrophils in the lungs. These powerful but fragile cells have one job, to kill microbes. They belong to a major subset of white blood cells and are the first cells recruited to the sites of infection to eliminate microbes. They have a very specific microbe-killing mechanism. First they recognize the microbes and engulf them. Then they kill the microbes by exposing them to potent, deleterious molecules. Once microbes have been eliminated, neutrophils finally kill themselves. Though destructive, this mechanism is critical for killing germs and normalization of the lung function. In the CF lung, neutrophils are inefficient in their jobs, and are actually accumulating rather than eliminating themselves and their waste through cell suicide. How and why are these cells impaired? What is the link between the inefficient neutrophil, unable to do its designed function, and inflammation in the CF lung?

Dr. Tirouvanziam postulates that glutathione (GSH), a ubiquitous molecule found in and out of cells, functions abnormally in the CF lung compared to the normal lung. GSH's main purpose is to maintain proper cell function and general metabolism, and more importantly it is used by the neutrophil as a checkpoint that controls most of its functions, including the microbe-killing and cell suicide processes. In the normal lung, external GSH (GSH that lies on the surface of the lung) is in high levels in the small airways with almost no eGSH (external GSH) in the larger airways. The small airway supply of eGSH is provided by the liver and by small airways themselves. In the lung with CF, eGSH levels are low. Recent research has proven that CFTR mediates the transport of GSH. Because of impaired expression of CFTR on liver and small airway epithelial cells in CF, small airway eGSH levels are too low. Because there is little or no eGSH for the neutrophils to use when recruited to the CF lung, these are inefficient in killing microbes and in undergoing cell suicide, which results in chronic infection, inflammation, obstruction and tissue damage.

Convinced that neutrophil impairment is the driving force in early CF lung disease, Dr. Tirouvanziam has started the GSH Project within Dr. Jeff Wine's Cystic Fibrosis Research Lab at Stanford. His goal is to provide a model for the small airways and understand how these use CFTR to transport GSH and support neutrophils during the microbe-killing process. This new route of investigation will help determine whether GSH supplementation, by the oral route and/or inhalation can help bring the lung to a normal state and relieve the neutrophil-dominated inflammation that causes so much of the damage in the CF lung. ■



Dr. Rabin Tirouvanziam (far right) speaking with parents of CF children (also from right) Eric Heilman, Eric and Lori Lindmeier, and Pam Heilman.

CFRI Conference 2000: Spirituality and Health

Speaker: Fred Luskin, Ph.D. is a post-doctoral NIH Fellow in the Preventative Cardiology Department at the Stanford Center for Research in Disease Prevention at the Stanford University School of Medicine.
Summarized by Kathleen Flynn

Perhaps a more appropriate title for this lecture would have been, "Attitudes and Healing." Attitude is something we all struggle with and this talk made it painfully clear how much our thinking and belief systems either interfere with or enhance our health.

Dr. Luskin's own interest in this subject came about from a search for meaning in a world that can be a threatening place. In his review of research on the topic, he learned that what appears to help people cope with life's most difficult aspects is faith in a higher power, Spirit, or the belief in life's goodness, beauty and the power of love. Basically, our own attitudes toward the difficulties that come our way will, to a certain degree, affect our health. People who deal with hardship primarily with anger or depression have higher rates of heart disease and cancer. Whereas people who can take life's difficulties and reframe them in the context of something greater than themselves, whether it be a belief in the goodness of life or God's will, have more positive health outcomes.

Dr. Luskin described what biologically occurs in our bodies when we are thinking of something positive, versus focusing on something that stresses or angers us. With the latter, the body feels alone and in danger, and gears up to protect itself from being attacked. Facing adversity alone takes away the greatest strength we humans have to cope with adversity, which is our connection both to others and/or to a higher being or power. When we connect, every one of our biological symptoms shift. When we are bathing ourselves in thoughts about love, goodness, or things we feel grateful for, the heart function and nervous system are optimized. If we think of life as a struggle for survival, we are constantly forcing our bodies into a chronic state of fight or flight. In this state, we cannot connect with others or any other positive forces in the universe because we are existing behind a wall of protective armor. Simple acts, such as looking up and noticing something beautiful even though we might be feeling very stressed, reorient our thinking and our biology, healing the body.

Dr. Luskin cited numerous, fascinating studies that corroborate his findings. I recommend that everyone get this tape and take in Dr. Luskin's message. In fact, I wish it were mandatory in all of our schools. It may be the best preventative medicine that we have in our culture. ■



Dr. Fred Luskin, speaking at the CFRI Conference.

CFRI Annual Awards

Awards were presented at our Conference's Saturday night banquet. Mike Roanhaus, President of the Board of Directors, presented our **Professional of the Year Award** to Dr. Birgitta Strandvik. Dr. Strandvik was selected for her work as a world-renowned CF physician, including her intensive research into nutritional aspects of CF. Dr. Strandvik runs a CF clinic in Göteborg, Sweden, where she has had a lot of success with the median survival age, and lung function scores. Her patients have higher body weights and heights in comparison to other international CF centers. Dr. Strandvik was instrumental in putting on the XIII International CF Congress this year in collaboration with IACFA. In the midst of all this, at a very late date, she agreed to give up yet another portion of her summer to come and speak to parents of CF children and CF adults at the CFRI conference this August. Dr. Strandvik, quite obviously, cares deeply about people with CF and their families.



Mike Roanhaus, President of the CFRI Board of Directors presenting Dr. Birgitta Strandvik with the CFRI 2000 Professional of the Year Award.

CFRI's Dave Stuckert Memorial Volunteer of the Year Award was awarded to Kathy Russell. Beth Sufian spoke on her behalf. Kathy Russell has dedicated her life to serving people with CF. Kathy's 20-year career as a nurse allowed her to serve the needs of people in the hospital setting. When she retired, she continued to volunteer at the hospital and work on projects to fund CF research. For more than three decades, Kathy and her husband have

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Awards (Continued from page 5)

organized a variety of parties for children with CF. Kathy is best known for her work as a member and now as president of the United States Association of Cystic Fibrosis Adults (USACFA), and as editor of their newsletter, *CF Roundtable*. She was a founding member of the organization and has worked tirelessly for nine years to make sure the organization serves the needs of adults with CF across the country. Kathy works tirelessly, offering her services freely to family members who have questions or need a referral to resources to assist them with a variety of problems people with CF face each day. Kathy is an inspiration to all who meet her. It is fitting to honor her dedication to the CF community with the CFRI award. (Unfortunately, Kathy Russell could not be at the 2000 Conference to receive the award in person.) ■

Good News (Continued from page 3)

would call back. The family waited about thirty minutes close by their phone. The doctor called back and amazingly, the second pair of lungs was a match. The doctor cautioned Travis that everything was still tentative until he determined the quality of the lungs. Even still, Travis and his family packed up and headed to Stanford just in case. The family arrived at 8:30 that evening and all the preliminary health checks were performed on Travis. He went to the O.R. at 7 a.m. the next morning and the transplant was completed by 2 p.m. How odd (and how lucky) that after months of waiting for lungs, two sets had been offered to Stanford within a four hour period and both were matches for Travis. And how blessed Travis was to have not only a second chance at life, but a second chance for lungs in one day!

Travis is currently still at the Homotel next to Stanford. He is doing very well with no signs of rejection. The family eagerly expects to return home in another month. The holidays will truly be wonderful this year!

Defying Gravity and CF!

Janelle Estournes, nearly fourteen years old, lives with cystic fibrosis. Over the summer, she attended an annual family reunion near Tahoe, California. She participated in strenuous hiking and swimming, but had the thrill of her life when offered an opportunity to try out rock climbing. Janelle's cousin, an experienced climber, took Janelle and three other cousins out to the rocks to learn how to climb. Janelle says, "Climbing the side of a steep rock requires that you depend on all of your muscles and strength to pull you up, and it is a lot harder than it sounds. I can say that I made it to the top, with the

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Conference 2000: Managing Living with Cystic Fibrosis

Speaker: Robyn Petras, 34 year old Adult with CF.
Summarized by Francine Bion

Editor's Note: The following synopsis does not do justice to Robyn's talk. To really experience for yourself how inspiring she is, order a copy of her videotape.

Robyn spoke very eloquently about her strategies for living with CF from the adult perspective. CF influences every part of her day and affects all of her decisions. As her disease has progressed, she has developed ways to manage it so that CF is just a part of her and not who she is. Her health was excellent well into her 20's, but a serious decline in her health six years ago required that she make the difficult but necessary decision to end her teaching career. A non-CF health complication, as well as declining lung function brought on by CF, threw her into a severe depression. A friend and doctor challenged her to do three things: to gain 10 to 20 pounds, to meditate, and to exercise every day. She rose to the challenge and developed new goals and priorities to love and care for her family, to grow spiritually and to commit to daily exercises. The two modalities she credits with improving and maintaining her physical health are the American Biosystems, Inc.(ABI) Vest® and exercise, both of which require many hours of daily work.

Over the last two years, with her physical health much improved, she has been searching for healing. She credits the CFRI Conference of 1998 as being the single event that transformed her life, due to the ongoing relationships she established with other attendees. The conference triggered her search for peace in accepting how the disease limits her, peace in accepting the unknown about how CF will affect her body, peace in accepting change, and peace in accepting the circumstances of her life.

Robyn defines loss of dreams as accepting the fact that people with CF live life differently than their family members and friends. Loss of dreams may involve the loss of financial stability, career goals, or the loss of a previous state of health. As people with CF experience losses, they move in and out of phases of grief. Some of the phases include shock, denial, depression, and obsession. Experiencing these phases is natural, but people with CF must work to avoid getting stuck in any one phase. When one gets stuck, one thinks there is no other choice and the disease becomes overwhelming.

To move beyond a phase of grief, it is helpful to develop strategies for survival. Strategies that work for Robyn include daily exercise, airway clearance, meditation, massage therapy, keeping a medical journal, and having a good rapport with her medical team. She recommends that people express emotions through support groups, a journal, or email list groups. Getting rest and relaxation, knowing your limitations, nurturing your spirit and having a positive attitude are other ways that can help one accept losses and give hope for the future.

Her final thoughts centered on practicing compliance and the necessity for thinking of prevention—the whole point in using a Vest today, for example, is to maintain your health for as long as you can, even if you seem healthy now. Her journey for peace in the last two years has taught her to regard CF as her greatest opportunity for growth and development. ■



Robyn Petras at CFRI's Conference 2000.

CFRI Conference 2000: The Swedish Protocol for Treating Cystic Fibrosis

Speaker: Dr. Birgitta Strandvik, CF Center Director, University Hospital, Göteborg, Sweden. Summarized by Eric Lindmeier



Dr. Birgitta Strandvik at the August, 2000 Educational Conference.

Dr. Birgitta Strandvik, who runs the CF center in Göteborg, Sweden, began her presentation by pointing out that from a statistical point of view, members of the CF community have reason to be very optimistic about the future. Dr. Strandvik noted that with only conventional treatments, survival rates for people with CF have improved dramatically. For the CF person born in the 1960's in Sweden, the median life expectancy was 6 to 7 years. For those born in the '70's, the median moved to 30 years. Roughly 85% of the CF patients born in the '80's survive today, and for the CF child born in the '90's, there is no real decrease in life expectancy attributable to CF. She asserts that the Swedish success is not due to a less severe phenotype—in fact the Swedish mutation tends to be a more severe one. Instead, the success is due to 1) centralized care, 2) efficient antibiotic treatment combined with physical activity and physiotherapy, and 3) improved nutrition. Most importantly, Dr. Strandvik emphasized that as a CF caregiver, she does not

accept any decline in lung function in her patients. When indications of distress are observed, she prescribes an aggressive treatment to eliminate the infectious cause.

Dr. Strandvik plays down the focus on the eradication of *Pseudomonas*. She points to patients who have 100% lung function even 10 years after *Pseudomonas* colonization. She asserts that *Staph aureus* is a more problematic pathogen—its presence is more common early on, and it can do a lot of damage.

For children under six, Dr. Strandvik recommends a treatment approach that includes regular inhalation of salbutamol (albuterol), regular usage of oral mucolytics and dietary supplementation with antioxidant vitamins A and E. All her patients also engage in some sort of physiotherapy, whether it be a PEP mask or other devices, huffing, trampoline bouncing or other active playing. She emphasized that these regimens must be started at an early age and adhered to when no illness is present. If indications of *S. aureus* infection are observed (increased mucus or cough, poor appetite, weight loss) she prescribes oral antibiotics. (For older children, she recommends use of a blood test which checks for the *S. aureus* antibody titres, i.e., evidence of the body's own effort to eradicate a *S. aureus* infection.) Reiterating her aggressive treatment stance, Strandvik stated that this antibiotic course must be followed at the appearance of very mild symptoms. The doses must be high and should be administered in various combinations. She noted that her antibiotic of choice, flucloxacillin, has not shown any tendency toward reduced effectiveness due to resistance. The use of mucolytics should be increased during exacerbations.

At the first sign of *Pseudomonas aeruginosa*, Strandvik treats with IV tobramycin for 10 days and repeats that administration at each recurrence. She noted that inhalation of antibiotics make sense, and she is studying TOBI usage now. For IV courses, she recommends a new breed of small pumping devices and surgically implanted subcutaneous infusion ports. The port can last up to six years, and can be very unobtrusive. Because the pumps, which can fit into a pocket, are so easy to use, compliance can be increased.

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CFRI Conference 2000: The Returning to School Program

Speaker: Jeanne Kane, Returning to School Program Coordinator, Children's Health Council. Summarized by Kathleen Flynn

The Children's Health Council in Palo Alto, CA was granted money three years ago to help children with cancer re-enter the school environment after their long-term absence due to illness. Since then, the program has expanded to include all medically fragile children, including children with cystic fibrosis.

This program can help with all sorts of cognitive and learning issues and serves all children anywhere in the San Francisco Bay Area. Best of all, there is no cost to the families of the chronically ill children. It is entirely underwritten by grant monies.

If your child is frequently absent due to illness and has fallen behind, or has other psychosocial issues that are impairing his/her ability to function in the classroom, this program can provide an invaluable service to you and your child. They encourage the local school district to do any necessary assessments and meet the needs brought out in the assessment. Children's Health Council also has staff available to serve these needs. They will meet with your child's teachers and help you to better advocate for your child in the classroom. And they will even meet with your child and his/her classmates to help explain your child's condition and how it manifests itself. This has been found to greatly increase the empathy among classmates for the child.

Ms. Kane urged parents to get their plan of modifications done ahead of time. This is also something they can help you with. She spoke of the necessary timeline required to put these requests into writing and into practice. She felt it was very difficult to draft this sort of document when your child was acutely ill. Once in place, Ms. Kane felt the plan should be reviewed annually and modified as needed.

To find out more call the Returning to School Referral Line: 650-688-3622 or send an email request to Jeanne Kane at: jkane@chconline.org.



Jeanne Kane



Dr. Jeff Wine, with his daughter Jenny, age 14, and his wife, Marlene, at the CFRI conference.

New Newsletter!

CF Links: New Newsletter for the CF Centers of Bay Area Pediatric Pulmonary (BAPP) Medical Group (includes Oakland Children's and Cal Pacific's CF Centers)

The second issue of *CF Links* is out for those families being served by BAPP Medical Group under Dr. Karen Hardy's direction. This is also a helpful newsletter for other Bay Area families as well. A few highlights include:

- CF Journals with highly relevant articles along with comments and clarifications by CF Center Director, Karen Hardy, M.D.
- A CF chat group through the Internet to discuss CF related thoughts and feelings with other parents of and caretakers of children and adults with CF. The chat room is open Monday and Wednesday nights at 10 p.m. EST. The address is www.Delphi.com/cffamily/chat/ Try it out and let us know what you think.
- Late November, Early December: BAPP Family/Holiday Event including a talk on "Highlights of the North American CF Conference."
- To receive your copy of CF Links contact Catherine Delpizzo at Catherinedelpizzo@yahoo.com or at (510) 428-3885, ext: 5299.

CFRI Conference 2000: An Overview of Basic CF Research

Speaker: Jeffrey Wine, Ph.D.,
Director of the Cystic Fibrosis Research Lab at Stanford University, Stanford, California. Summarized by Eric Heilman

Dr. Jeffery Wine, Director of the Cystic Fibrosis Research Lab at Stanford, began his presentation by reviewing the basic facts about CF. It is a single gene disease, caused by a recessive gene mutation of which there are more than 900 known varieties. The CFTR molecule is made by the gene that is affected by the mutation. While experts in the field have had a good basic understanding of the CF defect for over 15 years, recent research has shown that wide variability in the level of severity of the disease in an individual can also be the result of the effect of modifier genes.

Two of these modifier genes seem to merit further examination. One of these produces mannose binding lectin, or MBL, a universal microbial that helps fight off various pathogens. In the general population, 15% have a mutation in this gene, but it has little significant impact on overall health. But the combination of the CF mutation and a mutation in the MBL producing gene tends to shorten life expectancy by about ten years. The other modifier gene is associated with abnormal levels of TGFB1, a compound that makes tissue scar more easily. Internal scarring is the "fibrosis" part of the problem in CF. In summary, a person seems to fare better with CF, if that individual has a high MBL and low TGFB1. Progress is being made at counteracting the effects of variations in these and other modifier genes. MBL can be manufactured, and although the technology for testing its administration exists, it is not readily available.

Dr. Wine then returned to the subject of CFTR, explaining that it is a molecular valve that allows chloride and bicarbonate to cross the cell membrane; that is, it is a transport machine that allows fluids to pass through the organ lining. He then pointed out that some researchers think that CFTR is doing something specifically in the lung that affects inflammatory response. In this light, the investigation of the role of dietary fatty acids is interesting. Specifically, researchers are examining the role of DHA, an essential fatty acid prevalent in fish oil, that is found in many places in the body, but is especially important in cell membranes. In a highly publicized recent study, CF mice were found to have extremely decreased levels of DHA in tissues of CF affected organs, and correspondingly, increased levels of another fatty acid. When fed large doses of DHA, this imbalance was eliminated and CF symptoms were relieved. Why is this interesting? The question is whether a diet rich in DHA can reverse CF symptoms in humans. Unfortunately, warns Dr. Wine, the answer is "We don't know." He points out that with CF, hundreds of physiological alterations occur, and it is not clear whether particular fatty acid imbalances are the problem or the body's response to another problem. There is no evidence that DHA administration actually prolongs life even in CF mice. Clinical trials will be needed to test its effects.

Dr. Wine next focused on CF and lung disease. He pointed out that CF is not a systemic disease. It affects only certain organs and, even then, only certain parts of those organs. In the lung, the CFTR is found at the highest concentration in the system of serous cells that produce the fluid lining the lungs. This fluid is an extremely complex soup full of anti-inflammatory agents and natural antibiotics which acts as a barrier to infection. This CFTR activity is most pronounced in a cellular system that operates in glands in the smallest parts of the lung, the alveoli and alveolar ducts, and therefore it is very difficult to study.

In contrast to the slow rate of progress in research into CFTR function processes, Dr. Wine pointed to a breakthrough related to *Pseudomonas aeruginosa* infection. He explained the process through which an individual *P. aeruginosa* bacterium enters the lung, attaches to the lung surface, grows, and then senses the presence of other

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CFRI Conference 2000: Sinus Treatments for Patients with CF

Speaker: Victoria King, M.D., Otolaryngologist in private practice in Canon City, CO.
Summarized by Eric Lindmeier

Dr. Victoria King, otolaryngologist and CF sinus specialist, has pioneered a treatment for the infected sinuses of CF patients. Under normal circumstances, mucus moves freely through the sinuses. Patients with CF, however, are plagued with sinus problems due to the thick secretions trapped in the sinus cavities. These sludge-like secretions eventually harden into a tar like substance in the sinuses, becoming a breeding ground for organisms and infection. Current methods of treatment for severely diseased sinuses is to surgically clean-out the sinuses. But, 88% of those who undergo conventional sinus surgery will revert to their previously infected state, most likely within one to two years. In an effort to prevent this re-infection rate, Dr. King pioneered a protocol of treatment for the sinuses called the Endoscopic Sinus Surgery and Antibiotic Lavage (ESSAL).

The first step using ESSAL is for a patient with severe sinus disease to undergo endoscopic sinus surgery creating small openings, or windows, into the sinuses. Once the sinuses are cleaned out, cultures and drug sensitivities to the bacteria are obtained. Then, tiny butterfly catheters are placed into each sinus, running out through the nasal passage to the opening of the nostril. After the surgery, Tobramycin (or another appropriate antibiotic) is injected into the catheters three times per day for a seven to ten day period. The catheters are then easily removed, and the patient returns for monthly office visits to have the sinuses flushed with the appropriate antibiotic. Visits can be stepped up if symptoms reoccur more frequently, and, if this is not enough, catheters can be reinstalled for a three to four day period with the patient administering the antibiotic at home three times per day.

Dr. King's statistics showed that her regular topical flushing of the sinuses dramatically reduced the need for repeat surgeries. Compared with 88% of patients that returned for surgery under conventional methods, patients using the ESSAL protocol only had a 19% repeat surgery rate. Lack of compliance with the monthly antibiotic lavage therapy directly correlated to the need for further surgery. ■

CFRI Conference 2000: What You've Always Wanted to Ask Your Pharmacist

Speaker: N. Lois Adams, C.RPH, MBA, President/CEO of Florida Cystic Fibrosis Pharmacy, Inc. Summarized by Francine Bion

Ms. Adams outlined key areas of concern regarding the use of pharmaceuticals.

Compliance and Outcomes

It is important to take your medications as directed by your physician. Also be honest with your doctor about your adherence.

Generic vs. Brand Name Drugs

Generic drugs are drugs that are no longer protected by a patent. In most cases, generic drugs are lower in price. Before you choose generic drugs, be sure you know they are made by reputable manufacturers (less reputable companies may use poorer quality medicines or unreliable quantities of the active ingredient); that they are AB-rated by the ABA; and that you are aware of the rate of recalls by the manufacturer.

For CF patients, Ms. Adams recommends that you not use generic



Lois Adams, second from the right, talking with other conference attendees after her talk.

Thank You

CFRI Appreciates the Generous Donations of our Sponsors!

The following companies made major donations to help CFRI underwrite the cost of our 2000 Conference, our most attended conference to date. Each year these dedicated sponsors help us to reach out to people with CF and their families and to further our mission of education and support. Sales representatives from the following corporations were also on hand all weekend to allow attendees to try out their wares and ask any questions they may have had about the products. Without their support, our Conference would not be possible. Special thanks go to:

American Biosystems, Inc., makers of the ABI Vest ®

Axcan Scandipharm, Inc., makers of Ultrase® Pancreatic Enzymes, ADEKs® Vitamins, and the Flutter®

Digestive Care, Inc., makers of Pancrecarb® Enteric Coated Pancreatic Enzymes

Florida Cystic Fibrosis Pharmacy, an innovative pharmacy catering to the needs of people with CF

General Physiotherapy, Inc., makers of GE Directional-Stroking Percussors

Genetech, Inc., makers of Pulmozyme®

PARI Respiratory, makers of PARI Jet® nebulizers and DuraNeb® portable compressors.

PathoGenesis Corporation, makers of TOBI®, tobramycin prepared for inhalation therapy

Vortran Medical Technology 1, Inc., makers of the Percussive Tech HF™ Intrapulmonary Percussive Airway Clearance Device

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CFRI Conference 2000: 504s and IEPs: Tools For Success at School

Speaker: Beth Sufian, J.D., CF adult and a lawyer specializing in the rights of the disabled. Summarized by Kathleen Flynn

There are two laws available to help parents advocate for children in the schools.

The first law is called the Individuals with Disabilities Education Act (IDEA). It says that any public school must provide a free and appropriate education for students with disabilities. Most schools have interpreted this law to provide for students with learning disabilities, but it is also meant to protect children with Other Health Impairments (O.H.I.), that is, kids with health problems who miss a lot of school for medical reasons. To qualify under this law, you must prove necessity. Additionally, teachers, school nurses or administrators may not understand that this law encompasses O.H.I. and, unknowingly, deny a child coverage under IDEA. If this happens, Ms. Sufian counseled the group to seek protection under the second law.

The second law comes under Section 504 (hence the name) of the Rehabilitation Act of 1973 and offers the same modifications. In general, fewer teachers and school administrators are familiar with this law. This law says that any agency that receives federal funds must make accommodations for people with disabilities and cannot discriminate against someone based on their disability. The advantage of 504 over IDEA is that it covers children in environments other than the public school. If you have a child in a private school that receives any federal funds, they must accommodate for your child. This also includes any college or graduate school in the United States (with the exception of the Virginia Military Institute), as they all receive some kind of federal funding.

With either law, you must set up a meeting with teachers, the school nurse, and administrators (called an Individual Education Plan (I.E.P.) meeting under IDEA, and a 504 meeting under Section 504) to discuss the necessary modifications your child needs. Don't get overwhelmed by all the papers. The schools are interested in having parents sign papers to show that the school is in compliance with the law.

Be creative in setting up modifications. The

(Continued on pag15)

CFRI Conference 2000: Parents' Panel Discussion

Summarized by Ann Robinson

Editor's Note: The following parents of CF children took part in a parent panel moderated by Kathleen Flynn, CFRI's Outreach Specialist. These parents presented honest and inspiring accounts of what it takes to live with cystic fibrosis while maintaining "normalcy" within their families. Each parent related personal strategies for coping with CF while still doing all the things families do to raise children in a loving and supportive environment. While the text below gives a brief summary of the topics discussed and questions raised, this article cannot do justice to the wit, wisdom, and wealth of information provided by the stories of these parents. Get the tape!

- Danny Altano is the father of two girls, Gianna, age 10, who has cystic fibrosis, and Marissa, age 6. Danny and his wife, Sydney, live in Soquel, California.
- Lisa Giuliano is the mother of two daughters, Jenise, age 7, and Larissa, age 9. Both girls have cystic fibrosis. Lisa and her husband, Joe, live in Portland, Oregon.
- Jill Miller is the mother of two children, Jonathan, age 10, who has CF, and Kristen, age 12. Jill and her children live in Connecticut.
- Chuck Nelson is the father of three children. Scott died ten years ago of CF lung disease at the age of 21. Kim, age 35, also has CF. She received a double lung transplant on June 22, 1998. His daughter, Tammy, does not have CF. Chuck and his wife, Jody, live in San Jose, California.
- Beverly Sufian is the mother of three daughters, Beth, age 35, and Sandy, age 32, and Aviva, age 24. Beth and Sandy both have CF. Beverly and her husband, Dave, live in Houston, Texas.

All panelists discussed various school issues. What do you tell the teacher? What accommodations should you request; for example, unlimited bathroom privileges, leaving the classroom to cough, taking medications during school and leaving school for medical appointments. How can the school nurse help your child? How do you deal with educating parents of other students? When do you consider home schooling and how does it work? What happens when your child has had extended absences? Conditions in a classroom may be causing your child to be sick. One parent discovered that mold growing in an improperly installed classroom air conditioner caused his daughter to be hospitalized 11 times in 19 months. When the condition was corrected, her health improved dramatically.

How do you give your child a sense of empowerment? How can you teach your children to dream their dreams? How do you help children learn about cystic fibrosis? How do you teach children to advocate and speak up for themselves? When do you begin to teach children to manage their own illness? Each parent addressed the complex issue of when to push one's child to do their treatments and when to let the child say "no." Teaching cause and effect: "If you do your treatments you will feel better," was easier with some children and lost on others. Sibling issues were also discussed.

Each parent also elaborated on his or her own coping strategies for when a child was sick. One parent was able to use the hospital experience to reach out to others and that eased his own pain. One parent talked about "pretending" that CF was just what normal childhood was, because even though it wasn't her idea of a normal childhood, it was normal for her children.

Finally, a discussion of when to bring up mortality issues with family, friends and, in particular, the child, was particularly poignant. Many people in the audience also had comments on this difficult topic. One thing was clear, no one wanted their child to be limited or defined by the statistics. ■

Retreat 2000 is a Hit!

By Isa Stenzel-Byrnes

The CFRI Annual Teen and Adult Retreat was held from August 7-11, 2000 at Vallombrosa Retreat Center in Menlo Park, California and proved to be a great success. Twenty-seven people with CF turned out for this year's Retreat with 20 of their friends, family members, and significant others. It is difficult to put into words exactly what happens at the retreat, because so much of it includes the indescribable process of building an atmosphere of compassion and empathy where relationships, trust and self-acceptance can flourish. The four days of Retreat included thought-provoking discussions, including topics on relationship issues, managing college and work demands, body image, declining health and lung transplantation.

The retreat also featured numerous and challenging arts and crafts activities, painting, and workshops. Professionals from the community donated their time to help out. Suzanne Flint, who runs workshops through Stanford's Complementary Medicine Clinic on stress management techniques, gave an excellent presentation on guided imagery, relaxation, and deep breathing. Also, Margaret Stein, a local art therapist, provided an art therapy session for all interested participants. The retreat also provided opportunities for exercise and outdoor activities including several recreational sports activities such as swimming and hiking the gorgeous Sawyer Trail in San Carlos.

Of note, several of our older teens have started college and shared their experiences during the retreat. One of the most joyful parts of this retreat was witnessing how these young people have grown up. They have a firm sense of their identity as adults with cystic fibrosis and they have developed into solid, mature and sophisticated people. Sitting outside under a majestic ginkgo tree on the lawn at Vallombrosa and listening to the inspiring words of all of these people living with CF was an invaluable gift and will inspire many of us to keep going throughout the trials of the year ahead. Thank you to all who participated in the retreat and to the volunteers who made it possible. All of us would like to thank CFRI and our sponsors for their generosity in making the retreat possible. ■



(From left) Alex Jenkins, Christina and Jessica Miller, and Anna Modlin.

Swedish Protocol

(Continued from page 7)

Returning again to her emphasis on aggressive treatment, Strandvik said that monthly checkups should be used to watch for signs of low-grade infections, and that clinical symptoms should not be accepted as an inevitable result of CF.

Shifting to the issue of nutrition, she noted the role of essential fatty acid deficiencies in the problem of low weight gain, as well as increased inflammatory response in the patient. Along with the regular pancreatic enzyme usage, she recommended dietary choices that maximize proper fatty acid balance. She noted that it is usually possible to obtain normal height and weight by using well chosen ordinary foods, but that essential fatty acid deficiencies could still occur. She responds to these deficiencies with IV intralipid supplementation when dietary approach fails to achieve the proper balance.

During the fruitful Question and Answer session that followed her main presentation, Strandvik disapproved of the use of Ibuprofen or steroids to reduce inflammation, asserting that these mask the body's signs of infection, and Ibuprofen is only treating the inflammation. Dr. Jeff Wine remarked that the success of 20 years of clinical experience would seem to reduce the need for clinical trials. Strandvik stated that while there are people at her clinic who are not faring as well as she might hope, their decline is due not to genotype factors, but rather due to improper adherence to treatment regimens. Responding to a question about Pulmozyme usage, she stated that careful use of mucolytics usually is sufficient for mucus clearance, but if the patient finds Pulmozyme beneficial, then that should be the guide to its choice.

Because Dr. Standvik's protocol is different from the CFF accredited protocol for treatment which most of us receive at U.S. centers, I found this talk very valuable. I recommend that readers purchase this videotape and listen to the talk. It brought up numerous questions from the audience and caused many of us to rethink some of the clinical practices that we take for granted and perform without thinking, as well as others that we do not have access to because they are not recommended by our physicians. This was a powerfully stimulating lecture. ■

Pharmacy (Continued from page 9)

enzymes. Enzymes vary in their absorption rate and the size of the spheres.

Drug Interactions

Be aware that there can be serious drug/drug interactions. In order to avoid drug/drug interactions you must inform your doctor of all the medications you are taking before he/she prescribes a new drug. Drugs can also interact with food, alcohol, and natural herbs. They can also compromise your skin's ability to protect against sunburn. You and your doctor must work together to avoid any potential problems.

Proper Labeling and Storage of Medications

It is important that your medicines have proper and clear instructions on the bottle. Pay attention to expiration dates. The drugs become ineffective. Separate oral and topical medications in your medicine cabinets and note which ones must be stored in a refrigerator. Drugs that require refrigeration can be shipped, but you must pack them well in ice.

Adverse Reactions to Drugs

Some drugs cause undesirable side effects even though the drug is doing what it needs to do in the body. Some of the more common side effects include rashes, hypertension, headache and dizziness, nausea and vomiting, dry mouth, tachycardia and drowsiness. An allergic reaction is more serious and can bring on anaphylactic shock.

Ms. Adams also offers these medication tips:

- Take all your medications as prescribed.
- Do not skip doses and take the exact amount.
- Take at the correct time of day.

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Conference Tapes

To order CFRI Conference 2000 Videotapes, send \$6.00 per tape (includes shipping and handling) to the CFRI office, 560 San Antonio Rd., Ste. 103, Palo Alto, CA 94306. Be sure to include the number of the tape that you want to buy. This year all tapes are for purchase only (it was too hard to get rentals back), so pass them along to a friend, family member, CF clinic, or hospital library when you are finished with them. Following is a list of the tapes of Conference 2000:

Tap e 1: "Overview of Basic CF Research," Jeff Wine, Ph.D.; "Legal Strategies for Dealing with Cystic Fibrosis," Beth Sufian, J.D.; "IEPs and 504s: Plans for Modifying School Rules and Requirements," Beth Sufian and Jeanne Kane.

Tap e 2: "The Swedish Protocol for Treating Cystic Fibrosis (including Nutritional Issues)," Dr. Birgitta Strandvik, CF Center Director, Goteburg, Sweden; "Ask the Doctors Panel," including Dr. Birgitta Strandvik, Dr. Richard Moss, Pediatric Pulmonologist, Dr. Kenneth Cox, Pediatric Gastroenterologist, and Dr. Victoria King, Otolaryngologist.

Tap e 3: "Parents Panel," including five parents of CF children, Danny Altano, Lisa Giuliano, Jill Miller, Chuck Nelson, and Beverly Sufian; "Managing Living with Cystic Fibrosis," Robyn Petras.

Tap e 4: "The Emotional Impact of Cystic Fibrosis," Dr. Ann Steiner; "Spirituality and Health," Fred Luskin, Ph.D.; "What I Would Like Everyone to Know About CF," Dr. Ruben Diaz.

Tap e 5: "Sinus Treatments for Patients with CF," Dr. Victoria King; "Liver Issues for Patients with CF," Dr. Kenneth Cox; "Ask the Pharmacist," Lois Adams.

Tap e 6: (Warning: the material contained on tape 6 is highly technical.) "GSH (Glutathione) Research Update," Rabin Tirouvanziam, Ph.D.; "Signals and the Trojan Horse: A New Treatment Strategy," Alexis Traynor-Kaplan, Ph.D.; "Post-Doctoral Fellowship Program," Jonathan Widdicombe, Ph.D.

CFRI Conference 2000: The Emotional Impact of Chronic Illness

Speaker: Ann Steiner, Ph.D., M.F.T., Associate Clinical Professor, Department of Psychiatry, University of California at San Francisco, and private practice in Benicia and Walnut Creek.
Summarized by Kathleen Flynn

Dr. Steiner reminded us, "Don't Postpone Joy," an uplifting segue into what can be the more muddy waters of the emotional side of chronic illness. This said, she reminded us that there would be many times during the course of our (or our child's) illness when our concept of freedom and joy would have to be redefined, and that we might have to look for new ways to include the things that bring us joy.

Dr. Steiner noted three traits that would help us cope well with the ups and downs of CF: flexibility, creativity and, most importantly, compassion. These attitudes should be bestowed upon ourselves as well as others as we face daily medical challenges. Dr. Steiner gave several additional tips for coping well with chronic illness:

1. Ask others for help. This might mean what Dr. Steiner calls, "borrowing someone's brain" when we are too tired or stressed to think clearly for ourselves.
2. Those who cope the best emotionally, tend to be more active in their (or their child's) care. Because they understand what is going on medically, they recognize the benefits of and adhere to their regimen.
3. Read literature on strategies for emotional coping. Return to literature that helped you in the past. You are bound to notice new information that resonates, or remind yourself of strategies you've forgotten.
4. Prepare for your doctor's visits with a handy list of questions. Bring an extra copy for the doctor to follow along with you. Bring someone else who can write the answers to your questions.
5. Keep your own medical record.
6. Take charge of feeling better. Learn to listen to the warning signs your body is giving you. Eventually, you'll learn to anticipate your body's needs and slow down before you get very sick.
7. Ask for support when you need it from a friend, family member or a support group. Additional sources of support include self-help groups, church involvement, individual or group psychotherapy, independent living centers, printed material or other support services offered by organizations focused on cystic fibrosis. Get outside help when it's anything more than the blues. If you can't budge from the depression you are feeling or are feeling suicidal, seek clinical help.
8. Dr. Steiner also presented her Medical Information Sheet, available for all to print out from her web site at www.DrSteiner.net. This document needs to be filled out with all the pertinent information necessary in a medical emergency. It then folds up to the size of a credit card for easy carrying in your wallet. This is for anyone on medication who wants to be out in the world, feeling safe and in charge of their life and disease. Giving a copy of an updated sheet to your doctors can save time and reduce the risk of drug interactions.



Dr. Ann Steiner at the conference.

Ultimately, Dr. Steiner reminded us that it is COMPASSION that will help to carry us in times when we have very little left to run on. Whether you have CF yourself or care for someone with the disease, treat yourself to this kindness. ■

CFRI Conference 2000: Liver Issues for People with CF

Speaker: Kenneth Cox, M.D., Professor of Pediatrics, Chief of Pediatric Gastroenterology, Hepatology, and Nutrition, and Medical Director of the Pediatric Liver Transplant Program at Stanford University.
Summarized by Eric Heilman

Kenneth Cox, M.D., pediatric gastroenterologist and renowned head of the Pediatric Liver Transplant Program at Packard Children's Hospital at Stanford, has a long history of leadership in the CF treatment field.

Dr. Cox began his presentation by noting that currently 50% of the CF population has some liver disorder, but most of it is mild, with only 5 to 10% developing into cirrhosis. As increasing CF survival rates lead to an overall aging of the CF population, the incidence of liver disease associated with CF also increases. CF liver disease is caused by two factors. First, the CF gene abnormality leads to the production of bile, normally the consistency of water, however in people with CF it becomes thick and glue-like, resulting in impairment in bile flow. This leads to damage to the bile gland cells, which in turn causes scarring and liver damage. Second, pancreatic enzyme deficiency causes the body to malabsorb fat, and when the body tries to mobilize nutrients needed to form proteins, a fatty liver results. Additionally, complications arising from diabetes and/or from the regular usage of drugs to combat other CF symptoms, increase the potential for damage to the liver. This can be compounded by IV drugs or blood transfusions.

To prevent and treat liver disease in CF, Dr. Cox listed six steps: **1)** Prevent and correct malnutrition; **2)** Immunize against hepatitis A and B; **3)** Administer bile salts (ursodeoxycholic acid, also known as Actigall®) where liver blood tests are abnormal or gallstones are present; **4)** Avoid toxic substances, especially alcohol; **5)** Treat complications of liver disease; and **6)** In extreme cases, liver transplants may be necessary. With regards to liver transplant, Dr. Cox pointed out that liver disease accounts for only about 3% of CF deaths. Transplantation becomes a viable option where liver damage is severe. Because of new drugs that decrease the problem of organ rejection in transplants, the five-year survival rate is now 85%. The biggest problem now is waiting time for donors. For children, parents can serve as living donors (if they are an accurate match) by giving a section or lobe of their liver. The donor's liver, in this case, will regrow to its normal size within about two to three weeks.

Dr. Cox made several recommendations for testing for liver disease in CF. Yearly physical exams should be undertaken, checking the liver and spleen for enlargement. Also, an annual blood cell count and liver panel can be used to see if liver enzyme levels are abnormal. Where abnormalities are discovered, ultrasounds should be performed to look for gallstones and abnormal bile ducts or abnormal liver consistency. Though not often done by CF practitioners for CF patients, this routine should be adopted. It is now the standard of care for CF. ■



Dr. Kenneth Cox speaking at CFRI's annual educational conference.

Overview of Basic Research

(Continued from page 8)

P. aeruginosa bacterium with which it gathers together to form a colony. This colony protects itself from the body's defenses, and from externally introduced antibiotics, by building an external microfilm shield. Stanford researcher Dr. Arthur Kornberg, while doing research not directly related to CF, discovered a gene, PPK, within the *P. aeruginosa* DNA that is used to make this biofilm shield. He found that a PPK mutated *Pseudomonas* did not create this biofilm or raise up the protected colony structures that pose such a threat in the CF lung. If this weakness could be targeted, the danger posed by *Pseudomonas* could be greatly reduced.

Dr. Wine used this last topic as an illustration of the importance of basic research. He pointed out that where the progress in CF specific research trials can be slow and deliberate, basic research can lead to fast unexpected breakthroughs. Basic research allows us to understand the basic defect that is cystic fibrosis and problems that arise from that defect. Once we understand the problem, the road to correcting it is a much more direct route. ■

Good News (Continued from page 6)

coaching of my uncle and cousin. I had the time of my life and I have scars to prove it! I would do it again in a heartbeat."

Janelle also keeps her CF in perspective. "Although I have CF, it has never stopped me from doing things I want to do like rock climbing, hiking at high elevations or playing basketball. If anything, it has made me more determined to do those activities. Now whenever I face a new problem or challenge, I can just think back to that day, and say to myself, 'If I can climb rocks, I can do almost anything!' " ■



Janelle, age 14, scaling the rocks in Tahoe, California.

Thank You, Taylor Bui!

CFRI says good-bye to our well-loved and highly respected Program Services Manager, Taylor Bui. Taylor's energetic and creative strategies helped to make CFRI a better place to work. Taylor, You Will Be Missed! And we wish you every good fortune at your new position.



*Taylor Bui,
CFRI's
former
Program
Services
Manager*

An Easy Way to Help Further Research Efforts!

Children and teens with cystic fibrosis sometimes experience pain, yet health care providers know very little about pain associated with CF. Researchers at the University of Alabama at Birmingham are interested in learning more about the pain experiences of children and teens with CF. We are also interested in what their parents think about the pain and what they do to manage it. We invite children and teens with CF and their parents to complete a 10-minute questionnaire focused on the pain experienced with CF, how the child and parent treated the pain and how their health care provider managed it. If you are interested in receiving a questionnaire, please call Dr. Marion Broome at the University of Alabama at Birmingham (205-975-0343) or e-mail her: broomem@son.uab.edu. We will send you the survey and a stamped self-addressed envelope with which to return the survey. The Institutional Review Board for Human Use at the University of Alabama, Birmingham has approved this study.

Honorariums

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

Rebecca Atkins	Lori Kipp	Tanya Pescht
Lucy Barnes	Leo Lunine	Mrs. Tom Rivers
Brett Bennett	Ruth Lunine	Tom Rivers
Lauran Catron	Wendy Mardigian	Jason Ross
Tess Dunn	Glen Matson	Otis Stracener
Eric Epstein	Helen Matson	Christina Swanson
Jill Epstein	Olga McCoy	Larissa Swanson
Jarrod Fischer	Spank McCoy	Heidi Tegner
Emily Fredricks	Chuck Nelson Family	Adam Thompson
Jessica Fredricks	Jack O'Brien	Laurie Thompson
Cathy Huston	Pat O'Brien	Tom Walton
Will Huston	Erica Pescht	Timothy Yang
William T. Huston		

Memorials

These memorials come to us not only for victims of CF but for their families and relatives as well. We extend our deepest sympathy to their families and friends. These gifts have given new hope to children and adults with CF. *Note: Occasionally someone who has died had the same name as a living person.*

Charlie Anderson	Margaret Favero	Matt Mitchell
Debra Ware Babbitt	Irene Firmstone	Kathy Moon
Lorraine Beach	R. D. Forrester	Eugene Northcott, MD
Lela Blue	Isaac Goldman	Vincent Parko
Donni Boismier	Philip Gordon	Lucia Pfuhl
Clara Borba	Cynthia Haley	Pamela Rockhold
Dorothy Briozza	Wright Helder	Thomas Rolefson
Milton Burnham	Edith Helsel	Gregory Schapiro
Thelma Cady	Maxine Hildert	Celia Schneer
Suzanne Campbell	Rose Horn	Jimmy Sloan
Ann Christen	Jim Irvin	Dave Stuckert
Kevin Denham	George Jenkins, Sr.	Laurie Stuckert
Delia Derrick	Shirley Kelley	Ivah Tegner
Bob Duckart	Christopher Kloss	Norma Mae Valentins
Clara Duckart	Eric Lay	Gordon Vaughan
Anna Earl	Rudy Miller	

How to Submit an Honorarium or a Memorial in Honor of Someone You Love

Do you have a relative or friend that you would like to honor? Are you searching for the perfect birthday present, wedding or anniversary gift or memorial? Give a gift with lasting impact. Make a donation to Cystic Fibrosis Research, Inc. We'll send a special message to the recipient or to the deceased's family informing them of your contribution if you give us the name and address of the family. Your donation not only recognizes your loved one's special occasion, it benefits children and adults with cystic fibrosis and their families as well. Mail your contribution with the name, address and occasion of the person you are honoring or remembering to: CFRI, 560 San Antonio Rd., Suite 103, Palo Alto, CA 94306-4349

Pharmacist (Continued from page 11)

- Take medications at the same time as one of your daily routines, such as at mealtime.
- You can also use an alarm clock to remind you when to take your medications.
- A weekly pill box or bubble pak may help. Ask your pharmacist about them.
- Consult your physician or pharmacist before taking over-the-counter (OTC) drugs. These OTC medications may be working against the goals of your treatment plan.
- Notify your pharmacist or physician of any side effects you may be experiencing.
- Keep a list of your current medications with you.
- Know why you are taking each medication.
- Do not stop any medication without consulting your physician.
- Take your medications with a full glass of water. This will also help you to stay well hydrated.
- Be sure to let your physician or pharmacist know if you cannot afford your medicines. There may be others that cost less.
- Don't be afraid to ask questions. ■

504's and IEPs (Continued from page 10)

bottom line is the child can get anything they need in order to learn. Beth assured us, any rule can be modified. Look to your child to determine what they need and what they can handle. Set up a program that works for them. A CF child's plan might include: child does not get late slips, child carries and takes his own enzymes, move child away from others who are sick, child needs to eat snacks in class, child does not need to ask permission to go to the bathroom or to get a drink, child needs modifications for P.E., child needs homework sent home after one absence, child needs a tutor after three days absence, etc.

Many parents don't want to set up IEPs or 504s for a child that currently appears healthy. However, when your child suddenly becomes ill and needs to be in the hospital, it is not the best time to be putting together a 504 plan. Beth felt that the best plans included provisions for if a child becomes ill. You may never need it, but it is in place if you do. ■

CFRI HAS LOST ITS LEASE AND WILL BE MOVING 12-31-2000! Our new office will be located nearby in the Bayside Business Plaza at 2672 Bayshore Parkway, Suite 520, Mountain View, CA 94043. A full update will appear in the spring newsletter.

Action Coupon – Become a Member of CFRI

CFRI is dedicated to sponsoring CF biomedical research and education about CF. When you become a member of CFRI, you join a community of concerned people who work together to fight cystic fibrosis. Your annual dues of \$15 (or lifetime membership of \$50 per person) entitles you to vote on the allocation of CFRI research funds and run for a Board of Directors position. All members also receive CFRI News, a newsletter providing the latest in CF research information and social news to the CF community. To join CFRI, simply complete the Action Coupon below and mail it to our office.

CFRI's mailing list is confidential. We do not sell our list, nor do we give out any names or addresses under any circumstances. Our mail solicitations include our personalized Mother's Day Tea mailing and a year-end Special Gifts request that accompanies the Annual Report. If you receive other solicitations for money, they are from organizations other than Cystic Fibrosis Research, Inc.

- | | |
|---|--|
| <input type="checkbox"/> Here is my annual membership for \$15 per person.
<small>(Contributions above the \$15 annual subscription rate will help offset costs for those unable to donate.)</small> | <input type="checkbox"/> Yes, I want to send Mother's Day Tea invitations in 2001. |
| <input type="checkbox"/> Yes! I want to help. Here is my check for _____. | <input type="checkbox"/> Please send me _____ invitations! |
| <input type="checkbox"/> Here is my lifetime membership of \$50 per person. | <input type="checkbox"/> Please remove my name from your mailing list. |

Name _____	Relationship to CF Person _____	Date of Birth (for CF patients only) _____
Street _____		
City _____	State _____	Zip _____
Telephone _____		E-mail _____

What do you like about our newsletter? _____

What would you like to see us improve in our newsletter? _____

Please list any ideas for future articles, regular columns or subjects you'd like to see featured here: _____

ABOUT CYSTIC FIBROSIS AND CFRI

CF is the most common life-threatening, hereditary disease in the U.S. Twelve million Americans are symptomless carriers of the CF gene. One in 2,300 American children is born with CF.

Symptoms include:

- persistent cough, wheezing or pneumonia
- loose, foul-smelling stools
- heat prostration
- abdominal pain
- excessive appetite, but poor weight gain
- salty-tasting skin
- clubbed finger tips

CF patients suffer from chronic lung disease and digestive disorders which may require frequent hospitalizations. At present, only half of those with CF survive to age 31.

ABOUT OUR FUNDRAISING

Cystic Fibrosis Research, Inc. raises funds through a number of channels:

- Personal mail solicitations
- Memorials and Honorariums
- Grants
- Special fundraising events
- Membership fees
- General donations

CFRI's EMAIL ADDRESS AND WEB SITE

CFRI's email address is: cfri@cfri.org. Use this address to obtain information about our organization, the latest word on cystic fibrosis, or to communicate with our office. Also check out our Web site: <http://www.cfri.org>. See you on the Internet!

ABOUT OUR SERVICES

CFRI offers the following services:

- Research funds awarded to scientists at major medical research centers
- Educational meetings for the CF community
- Annual CF Educational Conference
- Annual CF Retreat for ages 16 to adult
- Family support groups
- CF support groups for ages 13 to adult
- *CFRI News* newsletter
- Email and telephone referrals and support services
- Videotapes of meetings and conferences

ABOUT OUR SUPPORTERS

CFRI is a nonprofit, volunteer organization not affiliated with any other group. Our support comes from the dedicated volunteers and staff. Supporters include:

- Executive Director, Program Services Manager, Outreach Specialist (part-time) and two Administrative Assistants
- Research Advisory Committee of physicians, scientists and members
- Board of Directors and Executive Committee
- Advisory Committees
- General membership
- Mother's Day Tea senders
- Office volunteers

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



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