

CFRI *news*

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The Importance of Cultures: The Critical Role of the Accredited Cystic Fibrosis Center Laboratory

By Siri Vaeth-Dunn

As the parent of a teen with cystic fibrosis (CF), I am accustomed to holding my breath while waiting for the results of her latest sputum culture, hoping that she has not acquired a new pathogen in her lungs. When my daughter recently developed a fever accompanied by a deep “gunky” cough, I worried that she had a bacterial infection that would potentially damage her airways and impact her lung function. In hopes of avoiding a fifty-mile drive to our CF center, I asked that her pulmonologist call in orders for a sputum culture at a hospital closer to our home. While sympathetic to the inconvenience, Tess’ doctor requested that we travel to our CF center, where the laboratory is prepared to screen for pathogens prevalent in the cultures of those with cystic fibrosis.

The role of the laboratory at CF centers cannot be overstated. With continued progress in cystic fibrosis care, the median age of survival continues to rise. A key aspect of improved health outcomes for those with cystic fibrosis is the identification and treatment of infections. The lungs of those with CF provide an ideal environment for opportunistic pathogens. It is these pathogens, as well as the body’s immune response to them, that lead to inflammation and scarring of the airways, and the resulting progressive decline in lung function. Numerous studies have correlated lung function with frequency of CF Center clinic visits, at which clinical status is reviewed, lung function is measured and cultures are conducted to detect pathogens in the

lungs. Early intervention is critical, and the greater the number of visits, the better the outcomes.

The pathogens that are common within the CF lung differ from those of the general population. Numerous bacteria have complex DNA that allows them to adapt and flourish in CF lungs. *Pseudomonas aeruginosa* serves as a prime example of this. It is estimated that nearly 80% of people with CF are colonized with various strains of this bacteria, which undergoes many genetic adaptations in the lungs, making eradication difficult. In 2003, the clinical lab at Stanford identified 300 strains of *Pseudomonas* in cultures from CF patients. When it comes to diagnosis and treatment, time is of the essence, as

(Continued on page 5)

RESEARCH

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Spring Update

Research on Diverse Bacteria in CF Lungs

Weidong Kong, Ph.D., University of California, San Francisco, is investigating the relationship between pulmonary health in cystic fibrosis (CF) patients and the diverse mix of bacterial species that constitute the airway microbiome. Recently, several studies have demonstrated that the airways of CF patients are colonized by a complex polymicrobial community, and that the composition of this microbiome is associated with pulmonary health status. By integrating high-resolu-

tion phylogenetic microarray (16S rRNA PhyloChip) and metaproteomic approaches, Dr. Kong is currently exploring the dynamics of the airway microbiome and host immune response during periods of pulmonary stability, acute exacerbation, antimicrobial treatment and recovery in CF patients. His current research demonstrates that specific changes in airway microbiota occur immediately prior to pulmonary exacerbation and following antimicrobial treatment, and that these changes occur at the same time as distinct and characteristic protein expression from specific host defense mechanisms. Moreover, in one patient studied, the recovery to stable

pulmonary health coincided with the return of the airway microbiome and the protein expression profiles of host responses to pre-exacerbation conditions. Dr. Kong aims to build upon these findings by using next generation sequencing to characterize global gene expression of the airway microbiome and to determine the relationships between the activities of these assemblages and pathogenic processes in CF patients.



Weidong Kong, Ph. D.

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Notes from our Executive Director

CFRI has once again earned Charity Navigator's highest rating: 4 stars! This reflects not only our careful fiscal management but also effective governance of the organization and incredible volunteer support. We maximize the value of every donation made to CFRI. Thank you!



live and connect with CF experts by checking our website (www.CFRI.org) and clicking on the UStream link provided.

Meet volunteer Barbara Curry who is active on both the Conference Committee and the Mothers' Day Tea Committee (See page 6).

Also, read about Ron and Pat Ware, who helped found CFRI shortly after their daughter Debbie was diagnosed with CF in the early 1970's.

Don't forget to mark your calendar for our 24th National Cystic Fibrosis Family Education Conference and our Teen and Adult Retreat!

Finally, we share a notice from California's GHPP, a health care program for adults with genetic diseases. Do you have a similar program in your state? If so, let us know, and we will post the information.

Warm wishes, and thank you again for your support,

Carroll Jenkins
Executive Director
(650) 404-9977 • cjenkins@CFRI.org

CFRI Membership: A Key To Community

By Siri Vaeth-Dunn

Since its inception, CFRI has been a member-driven organization. To be a member of CFRI is to join a community of people living with CF, which extends to parents, spouses, partners, grandparents, researchers, and clinicians. The gathering of CFRI's community at its General Membership Meeting is vital to the organization. At this meeting, voting members have a say in electing CFRI's Board of Directors, approving grant funding requests, and revising the organization's bylaws.

Our General Membership Meeting is held annually on the fourth Wednesday in May. In addition to participating in CFRI's business planning, members benefit from educational speakers whose presentations cover a wide range of topics. Previous guests have addressed such issues as CF

care, research updates, CF genetics, insurance options and special needs trusts.

There are two levels of membership: voting and general. There is no membership fee, and anyone may apply to be a voting member. The key responsibility of voting members is to review the CFRI materials under consideration, and then to vote by mail-in ballot, or to cast one's vote in person at the meeting. For those who do not wish to have this responsibility, general membership is the perfect alternative. All members receive CFRI updates, our newsletter, and invitations to CFRI events.

Our next meeting will be on May 25th. For more information contact JoAnn Davis, CFRI's Development Manager, at jdavis@cfri.org, or call 650-404-9979.

VX-770 Phase 3 Trials Yield “Profound and Sustained” Results

By Bridget Barnes

Imagine taking one pill twice a day and experiencing a 10 to 17% improvement in lung function, as well as a greater overall sense of well being. On February 23, 2011, Vertex Pharmaceuticals brought the cystic fibrosis community one step closer to that possibility. Vertex announced that the Phase 3 STRIVE study of the oral medication VX-770 demonstrated profound and sustained improvements in lung function as well as other disease symptoms associated with CF. Designed to evaluate patients age 12 and older with a mutation in their CF gene known as G551D, VX-770 targets the underlying defective protein that causes thicker mucus and repeated lung infections in cystic fibrosis. While only 4% of the CF population carries the G551D mutation, the results of this study are particularly encouraging because Vertex and others are looking at ways to correct the way the CFTR protein is made and transported to the cell. Vertex is also running two other studies that could benefit those who carry the Delta F508 mutation, which affects 70% of the CF population.

The G551D mutation causes the CF gene to create a defective CFTR protein, which closes off the passageway moving chloride to the surface of the cells. Serving as a “potentiator,” VX-770 markedly improved the dysfunctional properties of CFTR channels in patients with the G551D defect by restoring the normal function of the mutated protein and allowing chloride to get through to the surface of the cells.

The trial included 161 patients who received either VX-770 or a placebo for 48 weeks. After 24 weeks, those who received the drug gained approximately 10 to 17 percentage points in lung function as compared to those getting the placebo, a statistically significant difference. Trial participants continued to take VX-770 or the placebo for another 24 weeks and the improvement in lung function was sustained, a key marker of success for Phase 3 trials. The study met its primary goal, the mean absolute improvement in lung

function from baseline compared to placebo through week 24, which was sustained through week 48.

Study participants also improved in other areas critical to the health of those with CF. They were 55% less likely to experience a pulmonary exacerbation, while self-reporting improvements in respiratory symptoms such as coughing, congestion, and wheezing. Participants also gained an average weight of seven pounds, which is very beneficial for people with cystic fibrosis, as weight gain is linked to improved lung function. In addition, sweat chloride levels, a measure used



to diagnose CF, decreased in patients on VX-770 towards normal levels, indicating that the drug was truly effecting the faulty CFTR function, the underlying defect in CF. Sweat Chloride levels in those on placebo experienced no change.

VX-770 is being developed by Vertex and was discovered in collaboration with the Cystic Fibrosis Foundation, which contributed \$75 million towards the effort. “Treating the underlying cause of cystic fibrosis with VX-770 led to clinical improvements that were far beyond our expectations, providing support for an entirely new approach to the treatment of this disease,” said Peter Mueller, Executive Vice President and Chief Scientific Officer for Vertex.

The data collected from the Phase 3 trials support Vertex’s plan to seek approval for VX-770 from the U.S. Food and Drug Administration (FDA) in the second half of 2011 and, with FDA

approval, availability for those patients with the G551D mutation. “Due to the significance of these data and the great need for new, more effective medicines, we will work with regulatory agencies to determine the fastest way to get VX-770 approved for people with this specific type of CF,” Mueller explained.

Vertex also reported the results of the Phase 2 DISCOVER study using VX-770, which was designed for those with two copies of the Delta F508. This mutation causes the CFTR protein to fold itself in such a way that it is prevented from inserting itself into the cell membrane. The primary goals of the DISCOVER study were safety and absolute change of baseline lung function through 16 weeks. The results were not statistically significant.

The Vertex Phase 2a trial is testing VX-770 in combination with a drug known as VX-809. VX-809 is a CFTR “corrector,” which helps the defective CFTR protein move from where it is generated to the proper location in the cell membrane, and increases the amount of CFTR at the cell surface. “Based on the results of DISCOVER, we continue to believe the combination of a potentiator and a corrector may be the best approach to treating people with two copies of the F508 Delta mutation,” said Dr. Robert Kauffman, Senior Vice President and Chief Medical Officer for Vertex. Results from this clinical trial are expected later this year.

These studies underline the importance of participation in CF clinical trials. The sooner eligible CF patients enroll, the sooner the results will be available and advances made toward better CF care. For more information on CF clinical trials, please visit the Cystic Fibrosis Foundation’s website at www.cff.org.

Nutrition and Cystic Fibrosis

By Laura Tillman

Good nutritional status in cystic fibrosis (CF) patients improves long-term survival. Up to 85% of patients with CF cannot produce the enzymes necessary to digest and absorb fats, proteins and starch (pancreatic insufficiency). The failure to digest (maldigestion) and to absorb food, nutrients, and vitamins (malabsorption) leads to poor outcomes. While a deficiency of pancreatic enzymes is the primary cause of malabsorption, other factors may include bicarbonate deficiency, abnormalities of bile salts, mucosal transport and motility, and anatomical structural changes. Energy expenditure may be as high as 199% of predicted for CF patients, usually due to increased energy demands (the body needs more calories to cough, breathe and fight infections). Because of this, those with CF may need 120–150% more calories per day than those without CF.

Symptoms of malabsorption and maldigestion include:

- Poor weight gain despite a good, often ravenous, appetite;
- Failure to thrive;
- Weight loss;
- Frequent, loose, and/or very large bowel movements;
- Foul-smelling bowel movements;

- Mucus or oil in bowel movements;
- Abdominal pain and/or excessive gas;
- Distention or bloating;
- Vitamin deficiencies, especially the fat-soluble vitamins A, B, D, E, K;
- Poor bone health due to malabsorption of Vitamin D and calcium — leading to increased risk for broken bones and osteoporosis;
- Cystic Fibrosis Related Diabetes (CFRD);
- Increased salt loss;
- Essential fatty acid deficiency.

Patients who receive optimal nutrition have better growth, maintain better nutritional reserves, and have better pulmonary function. There are numerous pamphlets and articles available that discuss the nutritional needs of the CF patient, as well as strategies to add the needed calories. The following provide beneficial information:

The New York Times Health Guide. Cystic Fibrosis — Nutritional Considerations. A Brighter Outlook for Cystic Fibrosis Patients. Carolyn Sayer. Feb. 11, 2011.

This article discusses the nutritional needs of those with CF, and provides strategies for adding protein and calories to their diet. <http://tinyurl.com/cy2abu>

Nutrition — Cystic Fibrosis: Changes Through Life. 2006 Cystic Fibrosis Foundation (CFF).

This pamphlet discusses issues related to nutrition in the CF patient, such as Cystic Fibrosis Related Diabetes, CF and Pregnancy, and Osteoporosis. The pamphlet may be obtained through the CFF: (800) FIGHT CF; www.cff.org; 6931 Arlington Rd. Bethesda, MD 20814

Nutrition in Cystic Fibrosis.

Julia L. Matel, Carlos E. Milla. *Semin Respir Crit Care Med* 2009; 30(5): 579-586. Several factors contribute to impaired nutritional status in CF such as: pancreatic insufficiency, chronic malabsorption, recurrent sinopulmonary infections, chronic inflammation, increased energy expenditure, suboptimal intake. Progressive lung disease further increases calorie requirements by increasing the work of breathing. Treatment programs that place an emphasis on higher caloric intake and more aggressive nutritional management in CF patients report better outcomes. <http://tinyurl.com/4tkhp8h>

Improving Nutrition in the Cystic Fibrosis Patient. Judy Pitts, MS, CPNP; Jennifer Flack, MS; Jessica Goodfellow, MS. *Journal of Pediatric Health Care.* 2008; 22(2):137-140.

Inadequate nutrition causes an overall loss of muscle mass as well as a decrease in the contractility and endurance of muscles, including respiratory muscles. This weakening in the respiratory muscles contributes to ineffective airway clearance and leads to increased risk for infection in persons with CF. The nutritional status of the patient with CF is crucial in the overall prognosis, especially in terms of pulmonary function.

<http://tinyurl.com/69tfhzq>

Nutrition in Cystic Fibrosis. John Pohl. *Practical Gastroenterology.* March 2010, pgs. 20-27.

This comprehensive article discusses nutrition therapies in CF and provides an update on pancreatic enzyme replacement therapy.

<http://tinyurl.com/4e6eon3>

GHPP: Important Update

The Genetically Handicapped Persons Program (GHPP) provides health care services for California adults with genetic diseases, including cystic fibrosis. Applicants must be 21 years of age or older, residents of California, and meet GHPP's income and medical eligibility requirements. If you are applying, or re-applying, please read the following updates:

- You must **re-enroll annually** with current information. Otherwise, medical services may cease! You will receive renewal information ONLY if your **current address** is on file.
- You must submit your **prior year's Federal Tax Form 1040** with your application. (If you are a dependent on another person's tax return, that return must be submitted.) If you need to obtain a copy from the IRS, visit <http://www.irs.gov/taxtopics/tc156.html?portlet=1>. ALWAYS save copies of this and other important financial information. If you do NOT file a Tax Form 1040, you will need Form 4506: <http://www.irs.gov/pub/irs-pdf/f4506t.pdf>.
- You may contact your **social worker or case manager** at your CF clinic to assist you with your GHPP application. For a short list of social workers and case managers in California, please visit www.CFRI.org.
- For a **GHPP application and requirements**, call 1(800) 639-0597, or visit <http://www.dhcs.ca.gov/services/ghpp/pages/default.aspx>.

Cystic Fibrosis Lab (Continued from page 1)

Pseudomonas can develop into a mucoid strain which is nearly impossible to destroy.

While in the past, *Staphylococcus aureus*, *Pseudomonas aeruginosa*, *Burkholderia cepacia* and *Haemophilus influenzae* were the most commonly identified pathogens, those with CF are increasingly culturing other bacteria, including *Achromobacter xylosoxidans*, *Stenotrophomonas maltophilia*, *Pandora apista*, and *Non-tuberculous mycobacteria*. In addition to bacterial infections, those with CF often experience fungal infections in their lungs, culturing various species of *Aspergillus*.

There are over 110 CF care centers in the United States that are accredited by the Cystic Fibrosis Foundation (CFF). Centers undergo a rigorous annual screening by the Foundation, which includes an audit of the labs used to grow and screen cultures. Laboratories that are not certified by the CFF often do not have the capacity to properly screen for the lesser-known pathogens that are now common in those with CF, but rare in the general population. Dr. Carol Conrad, Associate Professor of Pediatrics and Director of the Pediatric Lung and Heart-Lung Transplant Program at Stanford University, notes that most of the patients at the Stanford CF center have their cultures conducted there because, “It requires a special sort of growth medium to select out for *Pseudomonas* growth, lab personnel trained in knowing what else to look for, and specific protocols to search for *Burkholderia* and other pathogens. It is extremely expensive. Smaller community hospitals with limited budgets often can’t afford to do this testing.” In addition to identifying pathogens, CFF accredited labs have the ability to test the bacteria for antibiotic susceptibility or resistance.

While people living closer to urban centers have the advantage of direct access to their CF centers, there are thousands of patients living in remote communities who are hundreds of miles away from their care centers. Lucile Packard Children’s Hospital at Stanford has patients who travel over 300 miles for their clinic visits. For these families, driving to the CF center for a sputum culture outside of

their quarterly visit is often not an option. Knowing that local labs may not accurately identify CF pathogens, physicians at the CF Centers must rely on patients’ previous cultures. Says Dr. Conrad, “For people who live far away, we will usually treat on the basis of the last culture, and ask them to come down for recheck and re-culture at some point. Cultures don’t usually change so rapidly that we are missing a diagnosis of a new organism growing.”

What does the future hold? Someday, the standard agar plates and media used in most CF labs may be supplemented by



advances in the field of microbiology. The understanding of pathogens endemic to the CF population has been advanced due to the use of molecular technology, which utilizes nucleic acid gene sequencing. This technology was developed by Norman Pace, a professor at the University of Colorado at Boulder (who in 2001, won the prestigious MacArthur Foundation Fellowship — known as the “genius grant”). Professor Pace and his colleagues at CU-Denver’s Health Sciences Center and Denver’s Children’s Hospital examined cultures from 28 CF patients in Denver, ultimately identifying more than 60 species of bacteria. Notably, thirteen samples contained bacteria that would never have been identified with standard cystic fibrosis lab cultures.

This work is progressing due to the efforts of Kirk Harris, Ph.D., Assistant Professor of Pediatrics at the University of Colorado Anschutz Medical Campus, who initiated the CF microbiome research in the Pace lab, and now continues it in his own lab at Children’s Hospital in Aurora, Colorado.

Dr. Harris’ background in environmental microbiology and bacterial diversity led him to his study of bacteria in the CF lung. Says Dr. Harris, “It was clear that micro-

“The role of the laboratory of CF centers cannot be overstated.”

biology beyond the traditional pathogens was present in the CF airways, and that emerging pathogens were a problem. The sequence-based identification approaches that were developed in the Pace Lab offered a direct way to identify the diversity present more efficiently than culture. We quickly obtained results that showed bacteria in sputum that had not been identified by standard culture techniques.” In fact, in the study of the 28 CF patients in Denver, one child cultured only bacteria that would not be identified with the standard agar plates and media used in most CF labs.

According to Dr. Harris, the sequence-based identification approach to studying microbes, “is the most comprehensive — it can identify dozens of species per sample — but the turnaround is still slow.” This approach can take months to complete. Dr. Harris’ lab has another tactic for studying the microbes in CF airways. Says Dr. Harris, “The second approach is based on quantitative polymerase chain reaction (qPCR) that tracks specific organisms. This approach is much faster, but is only helpful if the targeted organism is important.” This technology is still in the research stage and is not yet available for use in clinical care. Says Dr. Harris, “We have several translational projects that may lead to improved care, but we need to collect additional data.”

In theory, the use of gene sequencing will be extremely useful, particularly when a CF patient’s health is declining, and standard cultures are not illuminating the cause. Until the current research translates to clinical practice, the use of CFF accredited labs is vital to CF health maintenance. With consistent cultures conducted by an accredited laboratory, clinicians can accurately diagnose and treat pathogens, leading to improved health outcomes.

OUR FOUNDERS Ron and Pat Ware

By Ann Robinson

Debbie, the oldest of Ron and Pat Ware's three children, was born in 1970 in San Jose, California. When Debbie was two years old, her persistent cough led to a diagnosis of cystic fibrosis (CF) by Dr. Norm Lewiston, who was then Director of the Cystic Fibrosis Treatment Center at Children's Hospital at Stanford.

Dr. Lewiston told the family that Debbie's life expectancy was 10 years, and referred them to the CFF chapter in San Jose. They spoke with Bob Stewart, Sr. who invited them to a meeting. After joining the group and helping to raise money, Pat was elected Assistant Corresponding Secretary. Ron and Pat became founding members of CFRI in 1975 when the organization was incorporated as a nonprofit. Ron says, "We began CFRI because we wanted more of our money to go to CF research."

Debbie was an artistic child who enjoyed CF camp. She was especially good at portraits and won several art awards in Junior High and High School. Even though Debbie took piano and ballet lessons, she was a tomboy and loved mowing the pasture with a ride-on lawn mower and trailing her younger brothers behind in the wagon. Scott and James did not have CF.

Pat related, "Our family life was fairly normal. We took vacations and traveled by train to see relatives in Ohio and California, and traveled to Disneyland. We took Debbie's medicine and extra snacks and were always careful about germs." Debbie even went with her Dad to Europe.

Debbie was an A student throughout school and on the Dean's List at Boise State University where she majored in art and social science. Her sense of humor, determination, ability to accept her life



Debbie Ware, with husband Adam Babbitt

and move forward, and her care and concern for others made an impression on everyone.

Debbie started taking oxygen off and on in 1996. Before her transplant at Stanford in 1997, she was using it 24 hours a day. At the time of Debbie's surgery, Hewlett Packard's insurance only covered the cost

(Continued on page 14)

The Heart of a Volunteer: Barbara Curry

By Bridget Barnes

Barbara Curry is the living definition of "devoted grandmother" and "dedicated volunteer." Ever since her grandson, Cameron, was diagnosed with cystic fibrosis at age two, Curry has been an integral part of most every aspect of CFRI. "I became a Board member and acquainted myself with CFRI," she humbly explained. "I volunteered in the office, became a tea sender and later co-chair of the Mother's Day Tea committee, and served on the conference committee."

Curry has been a member of the Mothers' Day Tea committee for many years. "Obviously I want to help our grandson and others with CF," she explained. "CFRI does so much good, not only by financing research but also in helping support those with this devastating genetic disease and their caregivers. It is an extremely caring organization from the staff down to the volunteers. My husband and I send out tea invitations with a friend who has a granddaughter with CF."



Barbara Curry

Curry has also been involved with CFRI's Conference Committee since 1997, when she attended her first conference. "I realized the importance of the support groups; the information being imparted by doctors, scientists and social workers about new research; nurses who spoke about hygiene, infection-control and cross-infection; and the sponsors who set up booths showing their products which help treat CF." Curry's role on the Conference Committee is significant: she helps create the conference

theme, suggests topics to be covered, and finds speakers. She explained, "We listen to those who have attended past conferences and review conference evaluations as to what subjects attendees would like to hear about in the future. We recruit speakers who will help make the annual conference a dynamic experience."

In addition to helping with conference planning, Barbara Curry also contributes her aesthetic expertise. She orchestrates the conference flower arrangements, which are donated from the garden of her friend and Tea collaborator, Peggy Jones.

As for her favorite "conference moment," one took place last summer. "In 2010, two Canadian couples who lived in communities remote from one other attended the conference for the first time. Both of them had daughters of a similar age with CF and they found they had so much in common. It was fun to hear them discover the similarities in their lives."

(Continued on page 12)



24th National Cystic Fibrosis Family Education Conference

***Cystic Fibrosis in the Spotlight:
Taking A Leading Role***

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Pradeep Singh, M.D.
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Seattle, Washington*

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Understanding CF Organisms and

By Siri Vaeth-Dunn

The complex path from the identification of pathogens in a cystic fibrosis patient's sputum sample to the selection of proper antibiotic therapies was compellingly presented by Ellen Jo Baron, Ph.D., and Deepak Sisodiya, Pharm.D., BCPS, at CFRI's monthly CF Discovery Series, held in partnership with Stanford University.

Dr. Baron is Professor Emeritus of Pathology and Medicine at Stanford University, as well as the Director of Medical Affairs for Cepheid, a high-technology molecular diagnostics company in Sunnyvale, California. In her presentation, Dr. Baron discussed the detailed process by which microbiology labs analyze sputum cultures



Ellen Jo Baron, Ph.D.

from CF patients. CF sputum cultures require special handling and procedures. A culture set utilizing special media is required to diagnose CF pathogens such as *Staphylococcus aureus*, *Pseudomonas aeruginosa*, and *Burkholderia cepacia*, which is not utilized with standard sputum samples. In addition, labs employ special treatment for CF mycobacteria using acid-fast cultures, and more stringent decontamination procedures.

As described by Dr. Baron, after collection, cultures are quickly delivered by courier to the lab. Once they are logged in, up to ten different culture plates and media are prepared. Dr. Baron presented fascinating slides of various plates and described the purpose of each one. For example, sheep's blood grows most organisms; CAN plates (colistin-nalidixic acid) kill gram-negative organisms, while allowing gram-positive organisms to grow; MacConkey plates grow gram-negative rods; and chocolate plates grow *Haemophilus* species as well as most other organisms that may be present in the sample. To

diagnose yeasts and fungus in the cultures, three plates are used: Chromagar candida, potato dextrose agar and Mycosel agar.

Once the samples are introduced onto culture plates, they are placed in an incubator and checked each day by a certified clinical lab scientist. On day one, the lab technologists often see colonies developing, but typically there are not enough bacteria to work on. By day two, bacterium may have reproduced enough to allow it to be isolated and identified. After five days, if nothing has grown on a plate, it is discarded.

The spectrum of colors represented by CF pathogens was vividly demonstrated by Dr. Baron's slides. *Staph* is mauve, *H. influenza* is grey, *B. cepacia* is pinkish, and mucoid *Pseudomonas* appears to be a "gloppy," metallic, silvery green, and often looks sugar-coated. (Dr. Baron added that *Pseudomonas* even has a particular smell — like "concord grapes.") The plates containing mold develop fuzzy colonies. Technologists have to keep the mold plates taped closed because the hazardous spores can be easily inhaled.

According to Dr. Baron, "painstaking care" is taken to ensure that lab technologists have a pure colony of the organisms for identification and further study. Once colonies are growing, technologists do a gram stain on each colony, examine them through a microscope, and then do a manual or automated biochemical spot test. While Stanford has two types of machines to identify pathogens and check susceptibility, Dr. Baron shared that analysis of mucoid *Pseudomonas* is not reliable with machines and is usually tested manually.

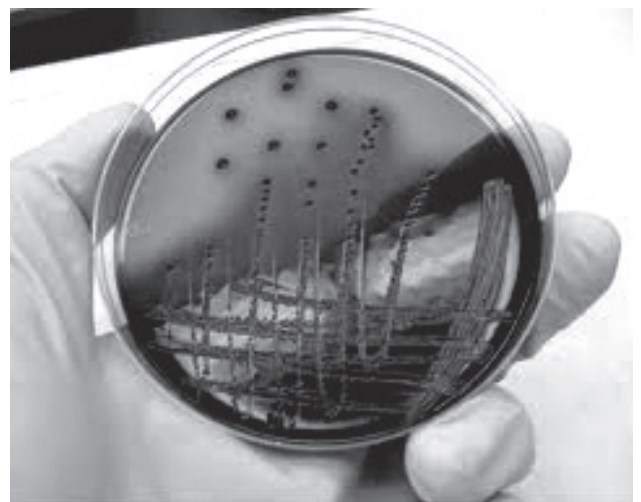
As Dr. Baron emphasized, "A lot of brainwork goes into interpreting what we learn from a culture result." When an organism's susceptibility patterns are not clear, often as a result of mixed cultures, technologists need to start

over, which can delay results. While this can be frustrating for CF patients and families, the labs are committed to accuracy.

The pathogens most common in CF cultures include *Staphylococcus aureus*, *Pseudomonas aeruginosa*, *Burkholderia cepacia*, and *Burkholderia gladioli*. In addition, Dr. Baron discussed other non-fermenting gram-negative rods that are important pathogens for those with CF, including *Acinetobacter*, which is often resistant to antibiotics. Dr. Baron said, "While it has low virulence, it is an important cause of nosocomial (hospital acquired) infections." *Stenotrophomonas maltophilia* is also usually multiply resistant. Fungal cultures are common. These are more difficult to identify and require a longer time to grow.

Dr. Baron discussed mycobacteria, another type of pathogen common to CF patients that "have a special cell wall composed of fats and lipids that need a special kind of acid-fast stain to show their true colors." The process requires decontaminating the culture and then liquefying it with a mucolytic agent. The regular bacteria are killed off — including *Pseudomonas*. The acid-fast organisms require more time to grow, and it can take one month or longer to make a diagnosis.

After identification, each pathogen is tested for "susceptibility," following stringent clinical lab standards. Once completed, all the results are entered in the



Antibiotic Therapies

computer. Typically, the time required for culture results is as follows: 4 – 6 days for simple cultures; 2 – 4 weeks for bacteriology; and 4 – 8 weeks for mycology. Dr. Baron shared that CF cultures require the most labor-intensive microbiology: two-and-a-half hours of “hands-on time,” versus 15 minutes for non-CF cultures. Dr. Baron stressed that even post transplant, physicians should still request a CF panel screening, because in the lab, “the same organisms are growing after the transplant as were growing before.”

Once the pathogens have been identified and tested for susceptibilities, the proper medication can be prescribed. Dr. Deepak Sisodiya’s presentation, “Cystic Fibrosis Antibiotic Therapies,” dovetailed effec-



Lab Technologists at work.

tively with Dr. Baron’s. Dr. Sisodiya, a clinical pharmacist specializing in internal medicine and infectious diseases at Stanford Hospital and Clinics, provided a helpful overview of the primary medications used with CF patients, and the pathogens that these medications are used to treat.

Aminoglycosides have been used for several decades, including three used primarily in the U.S.: gentamicin, tobramycin, and amikacin. Their mechanism of action is to inhibit protein synthesis, and they are useful for treating gram-negative bacteria, including *Pseudomonas*. They

must be given by IV, and are administered either with three smaller doses in a day (traditional) or as a larger dose once a day (once-daily). As they may have adverse effects, including renal (kidney) damage, and ototoxicity (hearing loss), serum levels must be closely monitored and specific labs for kidney function should be run. Hearing tests should be conducted regularly as well.

Quinolones have also been used for decades, and include ciprofloxacin and levofloxacin. A flat dose is typically prescribed for all and levels do not require monitoring. Quinolones may be taken by IV, but are also easily absorbed by mouth. Their adverse effects include gastrointestinal issues, photosensitivity, arthropies (damage to joints and tendons), and their use can lead to the development of *C. difficile colitis*, an infection that grows after normal gut bacteria has been wiped out. Quinolones are effective in combating MSSA (Methicillin susceptible *Staphylococcus aureus*), MRSA (Methicillin resistant *Staphylococcus aureus*), and *Pseudomonas*.

Beta-lactams constitute a whole family of drugs used to treat *Pseudomonas*, MSSA, MRSA, *B. cepacia*, and *S. maltophilia*. As explained by Dr. Sisodiya, “Beta-lactams share a commonality in their chemical structure,” and include the Penicillins, the Cephalosporins, and the Carbapenems (which Dr. Sisodiya referred to as “the big guns — a shotgun approach”). Aztreonam is also included in the family though it is structurally different. Allergenicity is an issue with beta-lactams, as well as gastrointestinal problems. Because Aztreonam has a different chemical structure, it sometimes can be used with people who are allergic to the other beta-lactams.

Clinicians and their patients may have limited treatment options based on sensitivities and allergic

reactions. In these cases, desensitization may be used, which entails slowly introducing small amounts of the needed drug to the patient, to allow the body to get used to it. The amounts of the drug are then slowly increased. Dr. Sisodiya stressed that this practice is used often at Stanford, and has been very successful.

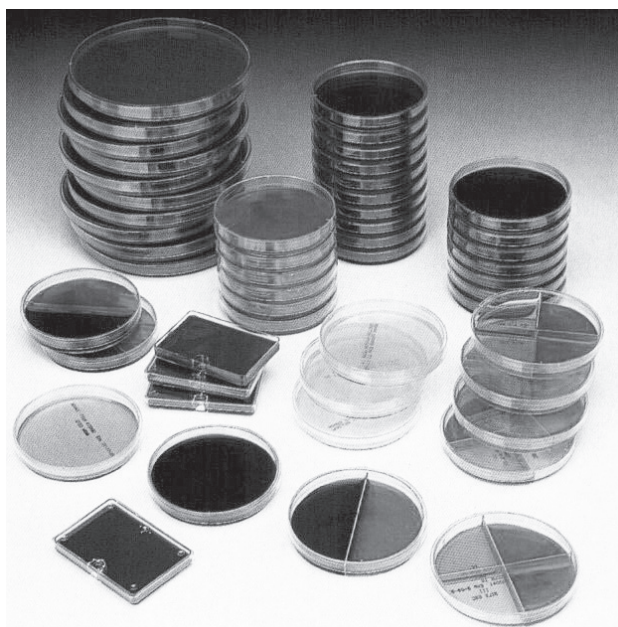
Vancomycin has been in use since the 1950’s, since its discovery in the jungles of Borneo. It is used solely for gram-positive bacteria such as MRSA. It is administered by IV, with dosing based on weight. According to Dr. Sisodiya, “Target dosing for ‘steady state levels’ is very difficult to achieve” with vancomycin as people with CF tend to clear drugs from their systems very quickly and often need higher doses.

Linezolid is a synthetic drug that can be taken by IV or orally, and is used to treat only gram-positive bacteria, such as MRSA. It has potential for both short-term side effects (gastrointestinal, drop in platelet counts) as well as long-term ones such as neuropathy,

(Continued on page 10)



Deepak Sisodiya, Pharm. D.



College Assistance for Disabled Students — California Department of Rehabilitation

By Katalin Rogers

There is a little known program provided by the California Department of Rehabilitation for disabled students – in our case students with cystic fibrosis – who plan to pursue their education at either a California-based vocational school, state college, or university, after having completed high school. The goal of this program is to ensure that disabled students receive either job training or education that will allow them to live independently and become productive members of society. This program is not income-based, and parents do not have to provide financial statements and tax returns.

The program provides tuition for the first two years of college for California students who study at community colleges, California State University or University of California campuses. After the first two years, the assistance will increase to cover the tuition at a state college or university, whichever is less. However, if the student is in a special program that is provided only by the University of California, and not by any of the California State colleges, the department covers the full UC fees, including textbooks for the last two years of a four-year program. Students might receive the same level of assistance at

private institutions, however there could be more obstacles to obtain this kind of help.

During the initial appointment with a Department of Rehabilitation counselor, the student must submit a summary of his or her career goals, fill out program forms and write an essay about his or her future plans. This essay has to include the student's research on job opportunities and pay scale for the chosen profession. After being accepted to this program, the process for obtaining the assistance is quite simple.

Before the beginning of each quarter, the student e-mails the upcoming quarter's course selection to the appointed counselor, and in return, the Department sends a voucher based on the number of units at \$26 per unit. The voucher has to be submitted to the school's financial aid office so that the money can be applied toward the tuition. The formula is slightly different starting with the junior year. The Department of Rehabilitation faxes an "authorization for payment" to the student's financial aid office that covers the tuition and fees. For the textbooks, the student has to submit his or her textbook

requirements indicating prices and taxes. The Department then sends the student a check that can be used as payment in the school's bookstore. In order to remain in the program, the student must send his or her semester or quarterly final grades to the Department counselor.

It may be difficult to obtain an initial assessment appointment with a counselor in the local Department of Rehabilitation office by phone, so it is advisable to go in person to the office and ask for an appointment. In case there is a difficulty with the Department of Rehabilitation, the client assistants at the Independent Living Center Programs can steer students in the right direction and provide help. To find out about your local advocates, call the main office in Sacramento and ask for a local referral at (916) 558-5390 or (800) 952-5544.

The regulation related to this subject can be found under "Barclays Official California Code of Regulations, Title 9. Rehabilitative and Developmental Services, Division 3. Department of Rehabilitation (FNA1), Chapter 3. Vocation Rehabilitation Services for Individuals with Disabilities, Article 3. Training and Job Coaching Services, California Administrative Code, Title 9, paragraph 7156. College Level Training."

CF Organisms and Antibiotic Therapies

(Continued from page 9)

and lactic acidosis — an acid-base disorder.

Colistin is another drug developed in the 1950's which can be used by IV or inhaled. Because of its negative renal and neurological effects, it fell out of favor, though it has made a re-emergence in the past few years due to resistance. Dr. Sisodiya expressed that, "Resistance is prevalent in the antibiotic world — a battle we are losing." One of the benefits of colistin is that resistance is relatively uncommon, though this is likely due to lack of usage.

Dr. Sisodiya then gave an overview of the primary pathogens affecting CF patients and the antibiotics to treat them.

Some newer agents have reached the market in the past year. Of note was the approval of telavancin by the Food and Drug Administration last year for the treatment of *Staph*, as well as the approval of ceftaroline, the first cephalosporin to have MRSA coverage.

The range of inhaled drugs available to CF patients was also reviewed, from antibiotics such as Tobi, colistin and aztreonam, to mucolytics such as hypertonic saline and DNase. Dr. Sisodiya noted that these drugs are often used to manage infections, not to cure them.

(Continued on page 12)

Because College Assistance Programs vary in each state, here are some helpful resources:

Disability.gov

<https://www.disability.gov/education>

College Funding For Students with Disabilities

<http://www.washington.edu/doiit/Brochures/PDF/financial-aid.pdf>

State Rehabilitation Services

http://wdcrobcolp01.ed.gov/Programs/EROD/org_list.cfm?category_ID=SVR

CFRI Scholarship Resources

<http://www.cfri.org/scholarship.shtml>



CF Teen and Adult Retreat

July 31st - August 7th, 2011
Vallombrosa Center
Menlo Park, CA



Meet some great friends.



Learn more about CF self care.



Feel that you're not alone.



Experience a place for hope and 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 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. Overnight accommodations and transportation are the responsibility of participants. Scholarships are available for those unable to pay fees.

Safety: All people with CF are required to comply with cross infection behavioral precautions. A medical advisor is available at all times, and volunteers are available to assist with respiratory treatments. 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.*

We'd love to see you!

For more information:

(650) 404-9975 www.CFRI.org

<http://www.facebook.com/home.php#!/group.php?gid=2342719557>

CFRI Golf, Auction And More...

If you missed last year's Golf Tournament and Auction at Cinnabar Hills, you missed a great time! Nearly 100 golfers spent a beautiful summer day out on the course vying for tee prizes and engaging in good-natured one upmanship.

Over 100 items were donated to our silent and live auctions, and bidding was fast and furious. If you have never witnessed a live auction run by Scott Hoyt, CFRI board member and General Manager of Cinnabar Hills, it is an experience! The weather was perfect, the golf unbeatable, the food delicious, and — as always — the company unsurpassed!

Thanks to the generous support of Cinnabar Hills Golf Club, the Kirkorian Foundation, the players, and their friends and family, over \$50,000 was raised for CFRI! Please plan to join us at the 2011 CFRI Golf Tournament, which will be held on Monday, August 1, 2011. For more information, please call Scott Hoyt at (408) 323-7803.

CF Organisms and Antibiotic Therapies

(Continued from page 10)

What are the “hot topics” in CF treatment and care? Dr. Sisodiya referenced allergic bronchopulmonary aspergillosis (ABPA), which is not an acute infection, but an allergic response, in which patients have a hypersensitivity to aspergillus. Diagnosis can be difficult, and usually entails skin tests and blood tests to check IgE levels (immune response measures). Treatment for ABPA includes steroids and itraconazole, which control, but do not eradicate, ABPA.

Dosing is another hot topic. “How many infusions a day are necessary for optimal dosing?” asked Dr. Sisodiya. He shared data demonstrating that dosing goals for beta-lactams can be better reached when infusions are given over a prolonged period (4 hours), and have the advantage

Help Us Grow

Your Donations Support Vital CF Research and Education

Mothers' Day Tea

Join our largest annual fundraiser by becoming a Tea Sender! We provide everything you need to send invitations to your friends, relatives and colleagues. It is a fun and unique way to raise funds for CFRI and spread awareness of CF.

Annual CFRI Golf Tournament – Monday, August 1st, 2011

Spend the day in golf heaven at beautiful Cinnabar Hills Golf Club in San Jose; then stay for the Silent and Live Auctions and delicious dinner buffet that evening. For more information contact Scott Hoyt at (408) 323-7803.

Need more information? Call JoAnn Davis at (650) 404-9979 or e-mail jdavis@cfri.org

Donate a Vehicle

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

Gifts in “Honor” or “Memory” of

Any donation may be designated in honor or in memory of someone special to you. Tributes are published in the *CFRInews* and if requested, an acknowledgement will be sent to the person or family being honored.

of less drug being used, but noted that this can make treatment even more challenging for the patient who is hooked up to an IV for extended periods of time. Dr. Sisodiya is also concerned with the risk of medication shortages, due to a lack of raw materials for their production. These materials often come from outside the U.S. and procurement can be difficult. In addition, regulatory issues have impacted the ability to obtain drugs.

Dr. Sisodiya expressed great concern about the dwindling drug development pipeline. Since the 1980's, there has been a significant decrease in the development of new drugs, “due to the lower profitability of anti-infectives.” According to Dr. Sisodiya, there are “only fifteen or so in the pipeline, and only eight target the worst pathogens.” Unfortunately, new antimicrobials are increasingly hard to come by due to focus on other drug therapies (e.g. blood pressure, asthma) by the pharmaceutical industries.

What's next? Dr. Sisodiya discussed the growing movement “to increase public and governmental awareness of the need for novel therapies to battle resistant

pathogens.” With the slogan “Bad Bugs Need Drugs,” a task force of the Infectious Diseases Society of America is actively advocating for an increase in drugs within the development pipeline to treat increasingly resistant bacteria that are significantly impacting the CF community.

The CF Discovery Series is generously sponsored by Genentech and A-Med Health Care. To view this presentation, go to: <http://www.cfri.org/CFDiscoveryVideos2011.shtml>

Barbara Curry

(Continued from page 6)

Barbara Curry is an extraordinary human being and the personification of all that CFRI exists to do: she helps raise funds for research; she provides educational and personal support; she spreads awareness of cystic fibrosis, a life-threatening disease. “I want to do all I can to see progress in finding a cure for CF,” she explained. In turn, CFRI wishes to honor Barbara Curry and thank her for the remarkable contributions she makes to our CF community.

April Showers Bring May Flowers...and Mother's Day!

By Bridget Barnes

Spring is here and that means Mother's Day isn't far away! It's time to help CFRI cultivate a brighter future for those with cystic fibrosis by participating in our biggest annual fundraiser, the Mothers' Day Tea!

By volunteering to be a Tea Sender you can join many others who devotedly send tea invitations to their family and friends year after year. These invitations encourage supporters to enjoy a cup of tea while thinking about a loved one living with cystic fibrosis. Your donation will further



Gina and Gianna Serrato

CFRI's outstanding research and programs, and improve and expand the lives of people with CF.

This year's brochure features Gianna Serrato, an adorable little girl who started her journey living with cystic fibrosis eighteen months ago. Ann Robinson, one of the original founders of CFRI and mother of Carl, a 33 year old with CF, photographed the colorful image on the invitation. As the population of adults living with cystic fibrosis expands, CFRI is proud to play such a crucial supporting role in the lives of parents with infants, children, teenagers, adults and even seniors.

Our longtime sponsor, R.C. Bigelow, has once again kindly provided the Orange & Spice Herb teabags to include in our invitations. Loyal supporter Pat Flynn donated a fine china teapot from Staffordshire, England, featuring painted pastel cupcakes, which will be raffled at the "kick-off" reception, where the lucky winner will be announced.

Devoted CFRI volunteers Barbara and Jim Curry, grandparents to 17-year-old Cameron who has CF, will host the Tea's kick-off reception on March 20th, from 2 to 4 pm, at their home in Hillsborough. Our goal this year is to raise \$247,000. With your help, CFRI will be able to



continue the New Horizons Research Program and the Elizabeth Nash Memorial Fellowship Program, educational programs such as the National Family Education Conference and Teen & Adult Retreat, as well as the production of outreach materials including *CF in the Classroom*, the *CF Website Guide*, and *CFRInews*.

If you want to become a Tea Sender, now is the perfect time! For complete packets including invitations, envelopes, tea bags and mailing labels, please call our office at (650) 404-9975 or email: CFRI@cfri.org. You can also sign-up online at <http://www.cfri.org/formMDT2011.shtml>. We will be delighted to provide you with everything you need and in turn you will have the pleasure of participating in CFRI's biggest fundraising event.

Yes, I would like to send Tea invitations. Please send me _____ (number of invitations)

Your Name (please PRINT)

(Area Code) Telephone Number

PO Box/Street Address

City/State/Zip

Email Address

Relationship to CF

(Complete and mail to CFRI)

In Honor of

Joanna Cryan Aiken
 Claire Alexander
 Gianna Altano
 Sadie Anderson
 Frank and Dorothy Andrews
 Jessica Arvidson
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 Kyle Baker
 Lucy Larkin Barnes
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 Barbara Greenberg
 Alec and Lizzy Hampton
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 The Hardy Family
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 Santosh Krishnan
 Steven Kusalo
 Dylan Leaphart
 Kaeti Pierce Lillibridge
 Michael Livingston

“In Honor of” and “In Memory of”

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

If you wish your donation to honor or remember someone special, please include the person’s name and address with your donation. At your request, we will send an acknowledgement of your gift to the person you designate.

Mail your contributions to:
 CFRI, 2672 Bayshore Parkway, Suite 520,
 Mountain View, CA 94043

Contributions listed here were received from
 September 16, 2010 through January 31, 2011.

Joseph Arthur Lopez
 Alyson Lowery
 Marc Anthony Maciel
 Larissa Marocco
 Helen Maschino
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 Claire McCabe
 Caryl McColly
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 Rachel and Rebecca McMullen
 Carly McReynolds
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 Payton Walker
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 Clare Webster
 Christopher Wernli
 Eliza Ming Williams
 Ricky Jon Williams
 Lauren Williams
 Nina Wine

Our Founders (Continued from page 6)
 of transplanting one lung.

Thanks to Ron’s persistence, the company changed its insurance policy to provide coverage for double lung transplants, thus helping future patients in need of this surgery.

Debbie married in 1998. Sadly, she developed a lung infection and died in 1999, at the age of 29.

Ron and Pat are retired and live in Boise, Idaho enjoying their seven grandchildren.

They are amazed at the progress in CF research, and that CF adults are often living into their 40’s and 50’s. CFRI thanks the Wares for their role in establishing CFRI and their fundraising efforts to support biomedical research and education.

In Memory of

Tommy Abrams	Mrs. Edmunds	Blake Kelley	Kim Nelson	Tammy Smerber
Marcus, Kimberley and Carol Adelman	Lesley Eiferle	Bridget Klein	Adam and Naomi Oneto	Danny Smith
Rosemary Altano	Hank Enge	Edmond Klein	Jennifer Ortman	Maxine Spruiell
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Melissa Reyes Dodson	Jim Kadow	Kate Nelson		
	Kevin Keaveney			

Please Save These Dates

For more information: www.CFRI.org

July 29-31, 2011

CFRI 24th National CF
Family Education Conference
Sofitel San Francisco Bay
Redwood City, CA

August 1, 2011

27th Annual CFRI
Golf Tournament
Cinnabar Hills Golf Course
San Jose, CA

July 31 – August 7, 2011

CFRI Teen & Adult Retreat
Vallombrosa Center
Menlo Park, CA

In this issue...

Critical Role of CF Labs	page 1
VX 770 — Profound Results	page 3
Nutrition and CF	page 4
CF Organisms and Therapies	page 8
College Assistance	page 10

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For their generous support of CFRInews.*

CFRI's Mission

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

CFRI's Vision

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



Get to know us:

www.CFRI.org 650.404.9975

