The Stanford CF Center has taken another important step in its evolution as a comprehensive program dedicated to the care of CF patients. In keeping with the growth and aging of our patient population, a major expansion of the adult CF care team has been implemented. The enhanced adult program was introduced in September 2008 with a significant expansion of staff, and adult program resources that include the addition of three physicians, a coordinator and two nurse practitioners, in addition to Paul Mohabir, MD, and the CF center clinical and research team. This evolution of the adult program has also included the identification of David Weill, MD, as its new director. The coordinated program offers Stanford patients one of the most comprehensive spectrums of CF disease management, clinical care and bench-to-bedside research in the US, with specialized resources starting with newborn screening through lung transplantation.

This issue of the Cystic Fibrosis Center News is dedicated to introducing you to members of the Stanford adult CF team, to sharing our philosophy of care, and to explaining how best to access our services.

Our Mission and Philosophy
The Stanford Cystic Fibrosis Center provides state-of-the-art care for a patient population that comes from Northern California and beyond. The clinical care component of the Center is comprised of three main programs: the CF Newborn Screening Program, the Pediatric CF Program and the Adult CF Program. Each program has its own clinical coordinator. The philosophical approach of the Stanford CF Center is that CF is a life-long problem and that best survival

Our Center’s mission is to excel in cystic fibrosis care, to be partners with those we care for, and to be leaders in the discovery process that will produce the cure for cystic fibrosis.
and best health will come when CF care is given in continuity by a multidisciplinary team of CF caregivers from the moment of diagnosis through the course of life. The new born, pediatric and adult programs function as independent components of the CF Center, but share core staff and facilities. As stated in our mission statement, we are committed to excel in CF care in partnership with those we care for. Our approach to the care of our patients is therefore fully consistent with our mission.

Comprehensive, Continuous Care Management

The CF Center at Stanford operates as a comprehensive center with coordination across ages, stages of disease, and facilities. Outpatient care and telephone triage are conducted at the same site, allowing close communication among team members, particularly during transitions between sites and programs. A weekly CF Team conference brings together physicians, staff and the research team to discuss all CF inpatients and patients scheduled for clinic visits that week. Patients and team members benefit from the dozens of years of clinical experience of the team, while enabling strong communication between programs to facilitate continuity of care. Our team members collectively represent extensive expertise in management of the most complex cases. The process is particularly valuable during transition to the adult program, or transition between physicians. An active transitional process is in place to assure that ultimately every adult patient followed at this Center is under the care of the Adult Program.

How the Adult CF Team Works

The adult CF program has grown from approximately 125 adults to more than 200 adults since 1999 when it was formally launched. New patients continue to outpace growth in the pediatric program, a result of pediatric patients “graduating” to the adult program, new diagnoses and referrals, and new patients seeking care at our specialty center. It is one of the largest adult CF programs in the country. The multidisciplinary team includes four board certified adult pulmonologists, a nurse coordinator, two hospital-based nurse practitioners, a nutritionist, a respiratory therapist and a social worker.

The Adult Team Expansion

In September 2008 the Stanford Lung Transplant Program physician team joined Paul Mohabir, MD, to establish a robust adult CF physician team. Under the directorship of David Weill, MD, a team of four physicians trained in internal medicine and adult pulmonary and critical medicine now oversee the care of adults with CF at Stanford. The team’s success in leading one of the top lung transplant centers in the world will be applied to the adult CF program. As experts in advanced lung disease, all four adult CF physicians have cared for numerous adults with CF throughout their careers. Weill is one of the first recipients of a CFF Program for Adult Care Excellence grants to support further career development in CF clinical and research activities. The team seeks to distinguish Stanford as a leading center for the clinical care and training of adult CF pulmonary and transplant physicians.

Adult Outpatient Clinics

Adult CF clinics are scheduled two days a week. The attending physicians, Drs. Weill, Mohabir, Sista and Dhillon each have at least two clinic days a month. Because they are also members of the transplant and critical care teams, when they are on service for those programs, clinic time may be more limited. Nurse coordinators Kathy Gesley, RN, and Nicole Eden, RN, attend the adult clinics, providing continuity of care for follow-up, telephone triage and port flushes. Patients are encouraged to choose the same physician for their care, or they may choose any available physician. The center is developing a team approach to care similar to the transplant program in which all physicians become familiar with each patient so that inpatient and outpatient care is coordinated, regardless of the physician on service or in the clinic. Weekly team meetings are held to discuss all patients, and the physicians actively share notes and advice on treatment plans and patient needs.

Respiratory therapist Kristin Shelton, RRT, and social worker Joanne Asano, LCSW, are also part of the team. Nutritionist Sabina Martinelli, RD, joins Julie Matel, MS, RD, CDE, to oversee nutritional aspects of care. Ms. Martinelli, RD, received her Registered Dietitian degree from Cal State Northridge and worked at Children’s Hospital of Los Angeles before moving to Packard Children’s.
Inpatient Care

One of the four adult pulmonologists is always on service to care for the adult CF hospitalized patients. In addition, one of the two inpatient nurse practitioners, Camille Washowich, RN, MSN, ASNP, and Elika Derakshandeh, RN, MSM, NP, is on duty to facilitate care plans and coordination of services between physicians, staff and patients.

David Weill, MD

David Weill, MD, is the Adult CF Program Director at Stanford and an Associate Professor in the Pulmonary and Critical Care Division of the Department of Medicine. He has been the Medical Director of the Stanford Lung Transplant Program since 2005. Weill trained at Tulane University, the University of Texas and the University of Colorado, where he pursued fellowship training in pulmonary and critical care medicine at one of the largest adult CF and advanced lung disease centers in the world.

Weill undertakes direction of the adult CF program with a commitment to excellence in patient care, education and research. He seeks to develop a premier adult CF program, distinguished by outstanding health outcomes and patient satisfaction, as well as advancement in the training of future pulmonologists in the care of the growing population of adults with CF. His experience in directing and enhancing the organizational structure of the Stanford Lung Transplant Program positions him well to firmly establish a model of care that will lead our Adult CF program to preeminence. His training and experience with CF patients with advanced lung disease give him a solid foundation for understanding the unique set of medical problems present in adults with CF.

In his short time as director of the program, Weill has overseen the training of a new team of nurse practitioners and the restructuring of adult inpatient care. He has ongoing strong relationships with adult medicine specialists in gastroenterology and liver disease, endocrinology and diabetes education, Ear, Nose and Throat and thoracic surgeons, and infectious diseases, which he is planning to leverage to facilitate consultative services and collaborative care management for CF patients.

In addition to clinical care expertise, Weill has been involved in clinical research throughout his career. He is co-principal investigator of the new Cystic Fibrosis Foundation Translational Research that will expand translational CF research at Stanford. Other research includes a broad array of lung transplant research that includes CF-specific studies on sinus disease and gene expression. He collaborates with Jeff Wine, PhD, in the study of airway glands and epithelial cells in CF lungs following transplant.

Weill is passionate in his commitment to build an outstanding adult CF center as part of a leading CF clinical and research center. He is pleased to join the current adult and pediatric teams in their pursuit of excellence.

Paul Mohabir, MD

Paul Mohabir, MD, served as the director and has been a key provider in the adult CF program since 2005. He trained at Michigan State University and completed fellowship training in pulmonary and critical care medicine at California Pacific Medical Center and Stanford. He divides his duties between critical care and the adult CF program. Mohabir has an active interest in clinical research in CF and interstitial lung disease.

Patients and team members benefit from the dozens of years of clinical experience of the team. Our team members collectively represent extensive expertise in management of the most complex cases.
He grew up in Canada and is fluent in French. CF treatment was a passion of Mohabir’s while he was in training. He is increasingly enthusiastic about efforts to fight the disease – especially the advances of the past ten years. As more patients live into adulthood, Mohabir notes that the disease evolves with them, and so must treatments. He welcomes the three new pulmonologists to the adult CF program as the program grows to meet increasing demand. The new resources should enable the team to strive toward an even greater focus on clinical and research opportunities to improve quality of care and collaboration among physicians trained in adult medicine.

Gundeep Dhillon, MD, MPH

Gundeep Dhillon, MD, was recruited to join the Stanford Lung Transplant team in 2005, following five years of pulmonary/critical care and lung transplant experience at Louisiana State University and the Ochsner Clinic. He is currently an Assistant Professor of Medicine in the Pulmonary Division at Stanford. Dhillon trained at Wayne State and Louisiana State Universities. He has been board certified in Internal Medicine since 1996, in Pulmonary Medicine since 1998 and in Critical Care Medicine since 2001. In addition to his fellowship training in adult Pulmonary/Critical Care Medicine, Dhillon received advanced training in lung transplantation at Stanford, and subsequently joined the program as faculty.

During his fellowship he received a Masters in Public Health, concentrating on epidemiology and biostatistics, from Tulane University. Dhillon has had an interest and expertise in the management of advanced lung diseases, including CF, as well as transplantation. He has extensive experience in the management of cystic fibrosis patients before and after lung transplantation, and has published studies on CF and non-CF advanced lung disease.

Dhillon has been actively involved in clinical and translational research throughout his career. He is currently site principal investigator for two multicenter clinical trials in lung transplantation. In addition, he is examining roles of airway hypoxia and CMV specific T-cell immunity on outcomes after lung transplantation. Dhillon also serves on the lung review board for the United Network for Organ Sharing (UNOS), the national organization for quality assurance and oversight of solid organ transplant.

Ramachandra R. Sista, MD

Ramachandra R. Sista, MD, is a clinical instructor of Medicine in the Pulmonary Division at the Stanford University School of Medicine. Sista trained in internal medicine at Wayne State University, and practiced at Louisiana State University. He completed fellowship training in Pulmonary and Critical Care Medicine at Stanford, and has worked as a member of the critical care and lung transplant teams for the past few years. Sista received advanced training in Lung and Heart-Lung Transplantation at Stanford and subsequently joined the program as a faculty member in 2008. Sista has developed interest and expertise in the management of advanced lung diseases, including CF, through his treatment of CF patients before and after lung transplantation.

Sista is actively involved in clinical research. He is currently co-principle investigator for a multicenter trial in lung transplantation. In addition, he is examining the role of airway hypoxia in subsequent airway fibrosis and chronic lung rejection after lung transplantation. Throughout his training and career Sista has received numerous awards for clinical teaching and a Young Investigator Award for his transplant-related research in pathophysiology of advanced cardiopulmonary disease.

Adult Nurse Practitioners

Care coordination for the adult CF patients has been significantly upgraded to include two nurse coordinators, Kathy

Kathy Gesley, RN, MSN, PNP, (left) and Nicole Eden, RN, MSN, ACNP, (right) nurse coordinators for the adult CF Program
Gesley, RN, MSN, PNP, and Nicole Eden, RN, MSN, CPNP, and two inpatient nurse practitioners, Camille Washowich, RN, MSN, ACNP, and Elika Derakshandeh, RN, MSN, NP. Kathy Gesley joined the adult CF team with twenty-plus years of experience that includes a broad spectrum of pediatric, childbirth education, allergy and hospice care. She has participated in several state and regional policy task forces dealing with hospice and maternal and child health. As the principal nurse coordinator for adult CF patients, she plays a pivotal role in communicating and coordinating care among the inpatient and outpatient teams. Nicole Eden shares the adult nurse coordinator role. Eden’s depth of experience with CF includes several years as the pediatric coordinator prior to training at UCSF to earn an advanced practice nursing degree. Eden and Gesley manage the outpatient adult coordination duties, seeing patients in clinic, arranging admissions, and coordinating telephone advice. Mary Helmers, RN, the prior Adult program coordinator, has assumed the role of pediatric CF program nurse coordinator, as well as mentor to the new adult CF team members.

Two nurse practitioners also dedicated to adult CF, Washowich and Derakshandeh, are based at Stanford Hospital to coordinate hospital admissions, inpatient care, discharge and follow-up. They provide case management for adult inpatients and provide a communication bridge between inpatients, physicians, residents and fellows. Stationed at the adult inpatient nursing units, they provide unprecedented access to CF expertise seven days a week, to oversee medications, facilitate treatments, and work with the adult CF physician team to manage inpatient stays. Both Washowich and Derakshandeh have many years of critical care and hospital nursing experience. Both share a strong commitment to working with patients, caregivers and the CF team to optimize patient outcomes and offer Stanford patients the best possible care management and hospital experience.

**Adult Hospital Team**

Expansion of the adult physician and clinical team has enabled the CF center to commit experienced adult pulmonology physicians to continuous coverage of the adult hospital service. This allows for the care of adults with CF to be completely supervised and managed by a team of specialists with training and expertise in adult medicine. As the largest adult CF center in California, and one of the top ten in the country, Stanford provides an unprecedented level of coverage, commitment and experience for adults with CF, from first transition as a young adult through chronic care management and transplant. The dedicated adult CF team, combined with Stanford Medical Center’s experienced nursing, respiratory and dietary team provide Stanford CF patients with the expertise needed to optimize patient care.

### Current Research Studies

Development of new drugs and therapies requires people with CF to participate in clinical trials. Be a part of the cure! Volunteer for a study today. To learn more, visit [http://cfcenter.stanford.edu](http://cfcenter.stanford.edu), contact our research coordinators or talk to your physician. The following trials are currently underway:

- Inspire Phase 3 “Tiger 2” drug for correction of salt and water abnormalities (closed)
- NAC Phase IIb (enrolling)
- Pulmonary exacerbation (enrolling)
- Vertex potentiator VX-770 (closed)
- MPEX 204 inhaled levofloxacin (closed)
- Kalobios anti-Pseudomonas antibody study (enrolling)
- EPIC trial early treatment of Pseudomonas (closed)
- Sweat testing in newborns with CF (enrolling)
- Chest CT and natural history of CF lung disease (closed)
- New trials to begin later this year
  - Vertex corrector VX-809
  - PTC124 Phase III for stop mutations (e.g. G542X)
  - Vertex potentiator VX-770 Phase III for mutation G551D
  - GSK oral anti-inflammatory SB-656933
  - Gilead FTI (fosfomycin-tobramycin inhalation)
The Cystic Fibrosis Foundation established the Therapeutics Development Network (TDN) in 1998 to facilitate the implementation of clinical studies exclusively focused on CF. Stanford was a member of the original network and rapidly gained prominence as a high-performing center. The success of the CF TDN in its first 10 years is best reflected in the high-quality research studies conducted including the early studies with Denufosol (now in Phase III studies) and Aztreonam (now awaiting FDA approval) as well as studies that led to the standardization of methodologies used to evaluate CF therapies such as Nasal Potential Difference (NPD), infant Pulmonary Function Testing (iPFT) and Induced Sputum (IS), among many other accomplishments. As the Foundation expanded its drug development pipeline (more than 30 therapies are now in development for CF) for early phase clinical trials and development of cutting-edge technologies and new outcome measures the need to significantly expand the number of TDN centers, became clearly apparent. At the same time, the CFF identified a need to maintain a core group of Translational Research Centers for early phase clinical trials and development of cutting-edge technologies and new outcome measures. These centers are expected to be highly specialized sites where the most advanced methodologies are in place for the execution of groundbreaking discoveries and the implementation of the most sophisticated studies. Only 13 centers in the US hold this high level designation. Stanford is proud to be a member of this elite group. Through our 10 years as a TDN center, we have not only established a solid research team but also have garnered the necessary resources within the medical center and our community at large to maintain a state-of-the-art center that is on the leading edge of CF research.

The Stanford CF Center sees this designation as a natural step in our evolution as a research center of excellence focused on CF. We clearly recognize that part of our mission is to be leaders in the discovery process that will produce the cure for CF. In many aspects our current activities reflect the spirit of what a clinical translational research center is meant to be. We have in place a highly interactive community of basic and clinical scientists that have produced a unique environment at Stanford. Further, our efforts are not in a vacuum. Stanford University at large has embraced the concept of clinical translational research and provides an environment that is fully supportive of our efforts. The Departments of Pediatrics and Medicine, Lucile Packard Children’s Hospital and Stanford Hospital, the School of Medicine, and the Lucile Packard Foundation for Children’s Health have all signaled strong support for the growth of the Stanford CF program and clearly consider it a high-profile program. Our CF FRD designation synergizes with Stanford’s recent receipt of a major Clinical and Translational Science Award (CTSA) from the National Institute of Health.

As CF clinical research continues to evolve and new potential therapies are identified, we have identified the following priorities to meet the needs and challenges ahead:

- **Improving the way novel therapies developed for CF are evaluated.** Currently there is a large consensus among CF researchers that more sensitive and dynamic outcome measurements are needed. We are committed to the development of new outcome measures for the most sensitive assessment and monitoring of lung disease in CF patients.

- **Apply the power of genomic medicine down to the level of the individual patient.** Novel therapies to control and perhaps completely correct the underlying basic CF defect will likely be specific to the defect in CF gene function that each patient carries. However, of the more than 1,500 disease-associated CF gene mutations reported, only a minority have been fully characterized. We are attempting to address this issue by establishing what would likely become one of the largest efforts for the longitudinal tracking of outcomes in CF patients from the time of birth.

- **Identify effective therapeutic interventions for complications of CF other than lung disease.** As CF patients live longer it has become apparent that CF is clearly a multisystemic process and a number of complications are becoming a serious problem for our patients. We believe that to address these different problems we need to gain a better
understanding of their basic mechanisms and embark on studies to identify abnormalities before the patient has overt signs of a complication. Our current work is geared toward gaining a better understanding of these defects and hopefully identifying not only better biomarkers for the presence of CF associated complications but also potential targets for intervention.

• Train the next generation of clinical and translational CF researchers. We envision translational CF Centers as entities that attract the best basic and clinical investigators, as well as members of the community and industry to facilitate the development of new knowledge and treatments for CF. We are committed to creating a fully integrated program that stimulates and educates all levels of trainees interested in investigational careers focused on cystic fibrosis.

In summary, Stanford University provides a rich environment that will be conducive to the seamless evolution of our highly successful TDN center into a translational CF research center. By engaging our colleagues from across disciplines in the basic sciences, clinical sciences, and bioinformatics fields at Stanford, we have established the most successful collaborative, cohesive effort in CF translational research in California. Our strong institutional support and, as importantly, the support from our patients, families and community at large, provides us the impetus to continue with our work on the path to the discovery of the cure for CF.

Richard Moss, MD, was honored by the local chapter of the CF Foundation as the Provider of the Year in recognition for his years of leadership in the regional and national CF. Moss has been appointed to the Executive Committee overseeing Stanford’s CTSA, the recently designated NIH Stanford Center for Clinical and Translational Education and Research. His wife Jill Kaplan and pulmonary nurse Deb Robinson join Moss at the celebration. Photo by Kymberli Brady.

David Weill, MD, was awarded a Program for Adult Care Excellence grant by the Cystic Fibrosis Foundation.

Colleen Dunn Receives CFF Award from Bob Beall, President of the Cystic Fibrosis Foundation

Colleen Dunn, RRT, CCRC, Research Coordinator received the national CFF Judy Williams Award in October, 2008, recognizing her outstanding contributions to CF research, education and clinical care. Dunn joined the CF research team at Stanford in 2000 following more than ten years as a respiratory therapist working with CF patients. She is actively involved in managing clinical trials at Stanford with her co-workers Zoe Davies and Jacqueline Zirbes. In her work with the CFF and the Therapeutics Development Network she is widely recognized as a leading advocate for CF research and a mentor to other centers. Dunn has always been a strong advocate for her patients and the Stanford CF Center, serving on institutional and national committees, most recently on the program committee for the 2008 and 2009 North American CF Conferences (NACFC). Dunn contributes to national CF research educational projects, including the development of curriculum for CF Research 101, study management tools, and best practices guidelines. Dunn’s recognition as an expert in care coordination issues specific to CF has lead to participation in site visits and selection committees for research studies and participants. Currently, Dunn is partnering with the CFF to establish a Research Coordinator mentoring program.

2008 NACFC

The 2008 North American Cystic Fibrosis Conference in Orlando, held in October, brought together over 3,000 clinicians and scientists dedicated to CF. The Stanford team lead an unprecedented number of sessions, including Jeff Wine, PhD, who delivered the main plenary lecture. Other Stanford program leaders and topics included:

Carlos Milla, MD, California Newborn Screening and Airway Clearance
Carol Conrad, MD, Oral Antioxidant Therapies
Colleen Dunn, RRT, CCRC, and Zoe Davies, PNP, Recruiting for Clinical Trials Management
Jacquelyn Zirbes, DRN, MSN, Complex Care for Newborns
Kristin Shelton, RRT, Spirometry for 3- to 4-year olds

In The News
Who to Call When You Are Sick, Need a Refill, Need Advice

All calls during regular clinic hours (8 am to 4 pm) should go to the CF Nurse Coordinator listed below. This is the only number families and patients should call during regular hours when sick or if you have questions. The call usually goes straight to voicemail, but messages are checked throughout the day and are returned the same day, unless otherwise stated. The coordinator’s voicemail will give the hours they are available and after hours/weekend numbers to call if there is an urgent need.

**Pediatric CF:** Mary Helmers, (650) 736-1359

**Adult CF:** Kathy Gesley/Nicole Eden, (650) 736-1358

If you or your child needs to go to the emergency department (ED) on your own without letting the Nurse Coordinator know or without calling the on-call MD first, it is possible that the CF team will not be notified that same day, which can result in delays. Please tell the ED staff that you are a CF patient, give them the name of your regular physician and ask them to page the pulmonary MD on call (for children). For adults with CF the Pulmonary/Transplant Fellow is the person who is first contacted for after-hours/weekend sick calls. Because the physician on call changes weekly, and you need to be specific with the emergency department staff to let them know your physician’s name and that you have CF.

Remember, all calls Monday through Friday should be directed to the nurse coordinators during clinic hours.

**For scheduling appointments:** call (650) 497-8841

For prescriptions: call your pharmacy first. If there are no more refills the pharmacy should call the physician office or fax the request to the prescription refill line at (650) 497-8791. Please allow up to 72 hours for refills. It is very difficult to get prescriptions called in the same day you make a request due to the paperwork and volume of calls.

If you or your child is sick, the CF nurse coordinator will triage the call. If it is determined that a clinic visit is necessary the nurse will schedule a visit if there is a clinic opening. If the regular physician cannot see the patient, another physician is not available and it is necessary for the patient be seen, the patient may be asked to go to the ED. The nurse coordinator will call the ED to let them know you are coming and why. For children the physician on call will be notified. For adults, the inpatient nurse practitioners (Camille or Elika) as well as the physician on call will be notified. However, you should follow the instructions above to ensure they know you have CF and that they should consult with the appropriate attending physician.

**CYSTIC FIBROSIS CENTER AT STANFORD**

**Pediatric Providers:** Richard Moss, MD, Center Co-Director; Carlos Milla, MD, Center Co-Director; Carol Conrad, MD; David Cornfield, MD; John Mark, MD; Terry Robinson, MD; Lauren Witcoff, MD; Nanci Yuan, MD; Jacquelyn Zirbes, DNP, RN, CPNP.

**Adult Providers:** David Weill, MD, Program Director; Paul Mohabir, MD, Associate-Program Director; Gundeep Dhillon, MD; Rama Sista, MD

**Clinic Scheduling** (650) 497-8841

**Clinic and Prescription Refill (Fax)** (650) 497-8837

**Miguel Huerta, Patient Services Coordinator** (650) 498-2655

**Mary Helmers, Pediatric Coordinator** (650) 736-1359

**Kathy Gesley and Nicole Eden, Adult Coordinators** (650) 736-1358

**Jacquelyn Zirbes, Newborn Screening Coordinator** (650) 721-1132

**Kristin Shelton, Respiratory Coordinator** (650) 724-0206

**Julie Matel, Sabrina Martinelli, Nutritionists, Dieticians** (650) 736-2128

**Joanne Asano, Social Work** (650) 736-1905

**Research Coordinators** (650) 736-0388

**For Urgent Issues:**

Monday-Friday 8:30 am - 5 pm, contact RN coordinator

All other times call (650) 497-8000, ask for pulmonary physician on call

**Visit our Web site at http://cfcenter.stanford.edu for more information about our center and CF.**

To subscribe to this newsletter please contact Cathy Hernandez by phone at (650) 724-3474 or by email at cathyh1@stanford.edu.

We gratefully acknowledge the leadership of friend and parent Penny Stroud in producing this publication.