Lucile Packard Children's Hospital at Stanford

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STANFORD SCHOOL OF MEDICINE
Stanford University Medical Center

CYSTIC FIBROSIS CENTER NEWS

A Word About Exercise: YES

BY JULIE DESCH, MD

There is little controversy regarding the importance of exercise to children and adults with cystic fibrosis. In the twenty years since Nixon et al. showed that higher levels of aerobic fitness were associated with a significantly lower risk of dying, numerous studies have documented the benefits of exercise, including: improved airway clearance, weight gain, improved cardiorespiratory fitness, increased work capacity, improved bone density, increased exercise tolerance, decreased feeling of breathlessness, and improved body image and quality of life. Even those who are awaiting transplantation are encouraged to exercise as tolerated, since studies have shown that exercise both before and after lung-transplant surgery tends to make cardiopulmonary function more efficient, strengthen respiratory muscles, and ensure good bone density.

But perhaps the most significant reason to incorporate exercise in the CF care regimen was shown in the longest published study to date regarding this issue. A three-year home based program including aerobic exercise three times weekly concluded that "*pulmonary function declined more slowly in the exercise group than in the control group*".[2] Certainly with the exciting prospect of CFTR correctors and potentiators around the corner, it is extremely important to maintain as much lung function as possible until the day comes when CF becomes a controllable disease.

Undoubtedly, we (adults with CF) should exercise regularly, and parents and clinicians should do what they can to encourage children with CF to introduce this healthy habit as soon as possible. But just as in the case of CFTR-able people, it is no easy task to convince those with lung disease to comply with an exercise prescription. The first obstacle is one of motivation, and the second is the very real issue of time-management. Neither of these are simple problems with simple resolutions; indeed, possible solutions change with the age of the patient and with the severity of the illness. Additionally, the recommendations for mode of exercise are slightly amended as a patient ages.[3]

PRE-ADOLESCENTS

The good news about young children is there is never an easier time to get them to move. Furthermore, a caregiver's power to promote long lasting behaviors at this stage is enormous. Children are wired to be active in short bursts; they play "anaerobically." They love to sprint or climb or do explosive movements, and then walk around to recover. This is a perfect for children with cystic fibrosis, because research has shown that this type of exercise trains both the anaerobic and aerobic energy systems. This type of play can also be performed for longer periods of time, due to the frequent rest periods. This is significant because the most recent research in exercise physiology shows that it seems to be total volume of daily activity that has the strongest effect on maintaining lung function. In other words, the more daily physical activity that a child is involved in, the slower

FDA APPROVES KALYDECO!

The entire CF community took a major step forward in finding a cure for cystic fibrosis when the Food and Drug Administration announced its approval of Kalydeco™ (VX-770) for CF patients with the G551D mutation. This breakthrough drug is the result of a decade-long collaboration between the Cystic Fibrosis Foundation and Vertex Pharmaceuticals. As the first drug to address the defective CFTR protein, Kalydeco represents a major treatment advance for the cystic fibrosis community. By attacking this disease-causing mutation at its core, the drug dramatically improves lung function, lowers sweat chloride levels and helps patients gain the weight they need to stay healthy. Not only is Kalydeco a major treatment breakthrough for people with the G551D mutation, the science behind the drug has opened exciting new doors to research and development that may eventually lead to a cure for all people living with CF.

1 Nixon, et al, N Engl J Med 1992; 327: 1785-8., 2 Schneiderman-Walker, J Pediatr 2000, 136: 304-10.
3 eCystic Fibrosis Review, March 2012: Vol 3, Number 8.

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.

A Word About Exercise story continued from page 1

their lung function will decline. Encourage anything that your child thinks is fun, and if possible, expose them to as many forms of exercise and sport as you can. This not only keeps it interesting for them, it also is beneficial to train their growing bodies in multiple ways. Some favorites are swimming, soccer, basketball, biking, jumping rope, rollerblading, games like tag or capture the flag, jumping on a pogo stick (one of my favorites), or a rebounder (mini-trampoline). The critical thing is consistent daily participation. The recommendation for children with CF is no different from that for healthy children, at least 60 minutes accrued throughout the day. At this age, it is also important that the intensity of the exercise be relatively high, as in a sport such as soccer, where there are frequent bursts of intense running, followed by longer periods of a walking recovery.

Motivation is usually not an issue with this age group, but if it is, delaying favored activities (screen time, reading, shopping) until after playtime works wonders. Some children are extremely motivated by team sports and group activities, while others shy away from them. If a child falls into the latter group, a pedometer may do the trick, or giving them exercise "points" or stickers and offering a great prize for accumulating a target number. It is also very helpful (and healthy for everyone) to establish family rituals that involve exercise. Weekend family hikes or bike rides are not just good for the children, they also establish exercise and time together as family values.

ADOLESCENTS

In both girls and boys, aerobic capacity begins to plateau during adolescence. For this reason, it is important to train the aerobic energy system during this stage of development. Aerobic exercise works the lungs, heart, and muscles, thus having a very powerful effect on the oxygen delivery systems in the body. Historically, girls have been at a greater risk for a sudden drop in FEV1 during adolescence, so while exercise is important for both sexes, this is a critical period for girls in terms of health maintenance. Unfortunately, it is also during adolescence that a significant difference in activity level develops between children with CF and healthy children, and this difference is even more marked in girls than boys. Low-level aerobic exercise, such as walking, biking, or leisurely swimming can be extremely beneficial for building aerobic capacity. In addition, this type of exercise can increase mucus clearance by up to 30%. This type of aerobic activity can be increased substantially by simply having teens walk or ride bikes to school, or to a friend's house or other planned activity. Fitting as much simple, low-level activity into daily life as possible can slow lung function decline. This is both time-efficient and can be a motivating fact to share with a teenager.

Exercising at a higher intensity is also beneficial, and can be much more tolerable and even enjoyable if done with *interval training*. This is characterized by two to three minutes of exercise at a moderate intensity, such as jogging quickly or swimming a few laps, followed by a short period of rest for the heart rate and breathing to return back towards baseline, and then repeating this for several intervals. This has the benefit of shortening the exercise session, a very important factor for busy teenagers.

Anaerobic training is also beneficial for adolescents. In sports like soccer or basketball, there are periods of intense exercise (anaerobic) followed by lower level recovery periods (aerobic), so both energy systems are being stressed and therefore, trained. Interestingly, adherence to this form of exercise has been shown to be very high, so kids actually enjoy this type of activity.

Finally, I would be remiss not to mention the myriad benefits of strength training for young men and women with CF. A study recently completed at the Hospital for Sick Children in Toronto shows that upper body strength and anaerobic capacity are significantly related to FEV1[3] Increased muscle mass, which results from weight training, leads to positive health benefits, both physical and psychological. Just as in anaerobic-based sports described above, when lifting weights, the anaerobic system is tasked during the lifting set, and the aerobic system is pushed during the rest phase between sets. On a personal note, it was when I discovered strength training as a young college student that my lifelong "addiction" to exercise took root. Understanding that this was one area where I felt control over my body, where my cystic fibrosis could not hold me back, was by far the strongest motivator to exercise I have ever felt. And I am female, lacking the benefit of testosterone! Imagine how significant it could be for a young man, perhaps smaller than his peers, to discover an activity that he could do to reliably increase strength and muscle mass. As I'm sure we all painfully remember, self-esteem is at its most vulnerable state during adolescence, and when you have a chronic illness, this is magnified. Depression and increased stress are definitely issues to contend with as teens with CF navigate increased time pressures, possible delayed puberty, and increasing health challenges. Exercise is known to increase self-esteem in children with cystic fibrosis, and is also a very effective way to deal with depression and stress.

Just as with younger children, variety improves the interest level and the overall benefits of an exercise program. If a teenager is inclined toward team or individual sports, this is ideal. If not, encouraging as much low-level activity in daily life, introducing interval training, and encouraging resistance training is my recommendation. Whatever the choice of activity, adolescence is the time to emphasize the importance of habitual, daily exercise as a central component to the routine management of their cystic fibrosis. Hopefully, as they notice that they feel and look better when they exercise, your teenager will develop the internal motivation to continue this habit into adulthood.

ADULTHOOD

The most exciting piece of information I've seen in researching this topic, comes from Toronto, where new, interesting and not yet published research that shows that in a seven year follow-up, FEV1, lung function, and habitual physical activity are closely related, and *the people with the highest levels of physical activity have a 50 percent slower*

rate of decline of lung function (emphasis mine).[3] For adults with CF, this means one thing: move more! Incorporating physical activity into your daily routine should be as important as your other therapies. Walk as much as you can. Ride a bike. Swim. All of these activities have well-documented positive effects, both CF-specific and non-CF specific. For example, regular exercise both helps with airway clearance and has been shown to be as effective as medication in treating depression and anxiety. For women especially, weight-bearing exercise such as walking, jogging, or weight lifting can help stave off osteoporosis. In non-CF subjects, weight lifting improves insulin sensitivity. It isn't known if this is true in CF, but it certainly could be. Most importantly, regular exercise improves the quality of life and perception of well being, and what can be more important as an adult with CF?

For adults with CF, the exercise recommendation is for "concurrent exercise," which is medical-speak for cross training. This means it is best to incorporate many different forms of exercise throughout your week. Strength train at least twice a week, work aerobically, at both low and moderate intensity as tolerated, at least three or four days a week, and try to incorporate flexibility and mobility work into your routine on several days. I recommend that people aim for 30 minutes per exercise session, but then also incorporate more movement throughout the day. Obviously, if you have more severe lung disease, you should consult with your doctor as to the need for O2 supplementation or any contraindications to exercise.

There are three important exercise considerations that apply to all age groups. First, if you are sick, don't work out. While it may seem macho to do so, and healthy people can get away with it, exercise would be an added stressor to an already-taxed system. If you can, continue with low-level activity such as walking, so that you don't decondition quite as much, and then begin again when you are healthy. Secondly, make sure that with increased physical activity, you or your child with CF are also taking in enough good quality calories to maintain the appropriate body-mass index. Even though exercise correlates strongly with lung function, so does nutritional status, and it would be counterproductive to sacrifice an optimal BMI in your pursuit of fitness. Finally, with increased exercise comes the need for fluid and electrolyte replacement. Especially when exercising in heat or for a long duration, this cannot be overstressed.

Clearly, I am a big fan of exercise. I firmly believe that one of the reasons I have enjoyed such good health as a DDf508 is that I made it a daily habit over 35 years ago. But now, it is not just me and other adults with CF who have seen exercise change their lives, preaching about the need for everyone with CF to *add this to their treatment regimen*. Research is proving us right, and even giving clues into the mechanisms of the benefits of exercise. As a fifteen-year-old, I didn't know that my jogging was causing my ENAC channels to be down regulated, thereby increasing airway surface liquid. All I knew was that it made me feel great! I became hooked, and I'm still here, writing about it.

STARS-2

ZOE DAVIES AND COLLEEN DUNN

In 2010, the research team decided to conduct a survey of all CF patients who had participated in at least one research study since 1992 in order to determine which areas of the program could be improved. This project was called **STARS**: **S**urvey **T**o **A**ssess **R**esearch **S**atisfaction at Stanford. Overall, study subjects were found to be very satisfied with their research experience, and we were able to identify specific areas that could easily be improved through research-focused education of patients, their families, and staff. During **STARS** data analysis, we found a large number of individuals who had either never participated in research or had never participated in a study at Stanford. Thus, **STARS-2** evolved!! We developed a brief survey to investigate the reasons for non-participation. Individuals were also asked if there was interest in participating in future studies, and if so, how would they like to hear about them and who would they like to hear about them from.

A total of 289 adult and pediatric CF patients were identified through the Stanford database as eligible to participate: 199 (69%) completed the survey. A non-biased interviewer asked the questions over the phone or in person at a clinic visit: 55% (n = 110) of respondents reported participating in at least one study and 45% (n = 89) reported not participating in any type of research study at Stanford. Of the non-participators: 55% (n = 49) said the primary reason for nonparticipation was because they were not asked, 27% (n = 24) reported that they didn't participate because of scheduling conflicts, 12% (n = 11) reported that they were not eligible, 9% (n = 8) were not interested in research, 4% (n = 4) felt that research participation would negatively impact their health, and 3% (n = 3) felt the length and complexity of studies made participation difficult. However, 93% of the respondents said they would consider research participation in the future if they were contacted. 94% had no preference as to who made the initial contact (physician, nurse practitioner, research coordinator, or clinic staff). 31% (n = 62) wanted to hear about studies during a routine clinic visit, 28% (n = 56) wanted to be contacted via e-mail, 3% (n = 6) wanted to be contacted via phone, and 45% (n = 90) had no preference how they heard about potential studies.

The percentage of individuals who did not participate in research studies at Stanford because they were never asked (55%) is strikingly similar to the survey that was completed by the Cystic Fibrosis Foundation (CFF) back in 2006 (54%). The research team was disappointed to learn that their increased education and recruitment efforts over the past several years were not recognized. However, this may have been due, in part, to the relocation of the adult CF program and the decreased research team presence in the new clinic. These results clearly indicate that there is a continued need to educate, and perhaps the face-to-face interaction between the patients, families, and the research staff is crucial.

We would like to thank Cathy Hernandez and Cassie Everson for all of their work contacting the patients and families and, of course, all the CF patients/families who took the time to complete the survey. These results were presented at the NACFC research coordinator workshop in November 2011.

The Potential Benefits of Including Omega-3 Fatty Acids in a High-Calorie, High-Protein Diet

BY LARA FREET, RD

A high-calorie, high-protein diet is the standard recommendation for people with cystic fibrosis to ensure that they are receiving enough calories and protein daily to meet estimated metabolic needs for weight maintenance or weight gain. Food sources that are high in fat are considered good choices as they are calorically dense, providing nine calories per gram of fat, therefore a person can eat a smaller quantity of food but achieve a higher caloric benefit than from protein or carbohydrate sources.

The focus on a high-calorie, and often high-fat diet, does not necessarily mean that a person has to eat butter, fried foods, or other sources of saturated fats and trans fats to achieve goal calorie needs daily for weight gain. Some types of fats are healthier choices for the body and research has shown that those fats have an anti-inflammatory benefit for the body. Poly unsaturated fats, also known as omega-3 and omega-6 fatty acids (FA), have been shown to have anti-inflammatory benefits in cardiovascular diseases. Omega-3 fatty acids, which have two metabolically active derivatives, eicosapentaenoic acid (EPA) and docosahexaenoic acid (DHA), is the focus of studies with the CF population for its potential anti-bacterial, anti-microbial, and anti-inflammatory properties.

Research published over the past few years examined the possible anti-inflammatory benefits of omega-3 fatty acids on infections in people with chronic diseases including cystic fibrosis. There is no conclusive evidence that consuming omega-3 fatty acids, either from diet or supplements, can definitely lower infection rates in people with cystic fibrosis however it is noted there are no significant negative side effects to omega-3 fatty acids [3]. A study conducted by Mil-Homens et al showed that there may be some benefit to increasing omega-3 fatty acids in the diet to support cellular membrane integrity and promote anti-inflammatory benefit [3]. Most recent research indicates that DHA may be the best form of omega-3 FA derivative to promote anti-inflammatory and antibacterial activity in people with cystic fibrosis. It is noted that omega-3 fatty acid supplement had beneficial nutraceutical activity in combination with antibiotic therapy for Pseudomonas aeruginosa and antibiotic resistant Burkholderia cenocepacia [4]. Both studies concluded that further research should be conducted to study and confirm potential benefits and better define dosing recommendations.

Foods sources of omega-3 fatty acids have been researched and marketed over the past several years for their heart health benefits, primarily due to their anti-inflammatory properties. Eating sources of omega-3 fatty acids on a daily basis will provide the same calories as eating saturated fat sources but will have more desirable health benefits. Food choices that have high concentrations of omega-3 fatty acids includes fatty fishes (like trout, herring, sardines, albacore tuna, and salmon), nuts (especially walnuts and almonds), soybean oil, canola oil, flaxseed oil, and dark leafy greens. The American Heart Association recommends eating sources of fatty fish (if it is appropriate with personal diet choices) two to three times weekly to optimize the nutritional benefit. Appropriate pancreatic enzyme dosing regimen will help ensure that

Pediatric CF Center Update

CF Clinic days have changed as of January 1, 2012. CF Newborn Clinic is held Monday mornings. All other patients are seen either on Monday afternoons, Tuesday or Friday mornings. When you call to schedule an appointment, the schedulers will let you know which day your provider has clinic.

Starting October, 2011 our CF Binder project begun. The binders are being given to all families. The goal of the new system is that the binders will enhance adherence in our patients/families with regards to their care and increase knowledge of the disease with the hopes of better outcomes for patients/families. The CF Action Plan is one component of the binders and it was developed to assist families to keep track of their CF information. Some items included on the form are your child's current mediations, spirometry results, nutritional needs and a plan of care. If any changes occur at the visit they will be placed on the form. In addition, we have asked you to fill out several questionnaires which many of you have already filled out and we appreciate all your efforts.

We have new turquoise-colored masks that we will we ask all CF patients to wear. They are being handed out at the front desk. These masks have smaller filters which allow for more protection when walking outside during all the construction. We would like all patients to wear them to and from all clinics/hospital and when you walk outside the medical center. They should fit snug around the nose and mouth. If you have not received the new mask, ask the front desk staff or anyone from the CF Team.

a person with exocrine pancreatic insufficiency may have improved success in digestion and absorption of fats in the diet, including omega-3 fatty acids.

There are also a variety of omega-3 fatty acid supplements available in stores, pharmacies, and online. The challenge is that further research is needed to study different doses of omega-3 fatty acids and any dose-specific potential benefits. There is no regulatory body that oversees the herbal and nutritional supplement industry therefore unless a company finances its own testing it is challenging to have a guarantee of potency and efficacy of a supplement. Discuss with your cystic fibrosis team if you want to start taking omega-3 fatty acids supplements or increase your intake of food-rich sources within your diet. Through the American Heart Association website there is a database for recipe ideas for how to incorporate more unsaturated fats, especially those rich in omega-3 fatty acids, into a high calorie diet.

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3) Mil-Homens D, Bernardes N, Fialho AM. The antibacterial properties of docosahexaenoic omega-3 fatty acid against the cystic fibrosis multiresistant pathogen Burkholderia cenocepacia. FEMS Microbiology Letters. 2012,328(1):61-9.

4) Oliver C, Jahnke N. Omega-3 fatty acids for cystic fibrosis. Cochrane Database Systematic Reviews. 2011,(8):CD002201.

Adult CF Center Update

Stanford's adult cystic fibrosis patients come from as far north as Oregon and as far south as Twenty Nine Palms, California. Driving to the clinic takes hours and coming to clinic when sick may require a family member to drive the patient. The nonprofit agency Angel Flight has started assisting our far flung patients with transportation to Adult CF clinic by flying them in prescheduled flights to clinic. Patients must be able to walk and be medically stable. Each patient is evaluated clinically for safety and an oxygen test is done to ensure they can tolerate the flight. This pulmonary test must be done at SUH before medical clearance is given. Volunteer pilots using their own aircraft arrange directly with each patient to ensure that the patient can be picked up and returned home after the clinic visit. Our patients love the flights and this volunteer group has helped some of our very sick patients return to clinic more often and improves care coordination with other SUH specialists. Angel Flight 888 4 An Angel accepts patients for flight under 1000 miles. Need more details? Contact Meg Dvorak, MSW at 650-723-6273, Adult Cystic Fibrosis Clinic.

Cystic Fibrosis and Hemoptysis

BY PAUL MOHABIR, MD

The pulmonary manifestations of Cystic Fibrosis (CF) are related to the combination of chronic airway inflammation, viscous mucous obstruction, and infection with a multitude of organisms. It is proposed that these three entities contribute to the destruction of airways and ultimate erosion into nearby vessels leading to bleeding and bloody sputum, also known as hemoptysis.

Hemoptysis is commonly described in CF patients. One study reported that over a five-year period, approximately 9.1% experienced some degree of hemoptysis[1]. Although most patients will have mild to moderate bleeding, it is estimated that 4.1% of all CF patients will have at least one episode of massive hemoptysis in their lifetime[2]. Of these patients, 74% will have one episode and 26% will have greater than one episode[2]. Majority (75%) will have first episode after the age of eighteen[2].

The definition of massive hemoptysis is the coughing of blood that is greater than 250ml of blood over 24 hours or recurrent bleeding with >100 ml/ day over several days. Massive hemoptysis is unpredictable and can occur at any time. Risk factors include:

1. Older age
2. FEV1<30% predicted
3. Pseudomonas aeruginosa
4. Staphyloccus aureus
5. Burkholderia cepacia
6. Tube Feeds
7. CF-related liver disease with cirrhosis.

Even with immediate intervention/treatment massive hemoptysis carries a significant mortality. Symptoms can include the obvious varying degrees of bloody sputum, shortness of breath, increased heart rate, chest tightness, massive hemoptysis leading to low blood pressure, respiratory failure, and cardiac arrest.

Treatment of hemoptysis is tailored to the degree of bleeding. It should also be considered that a smaller bleed might be a sign of an upcoming larger episode. Bronchial artery embolization (BAE) by Interventional Radiology should be immediately considered in patients with massive hemoptysis. BAE was first reported in 1973 and is the recommended procedure of choice for massive hemoptysis. BAE controls hemoptysis in approximately 90% and 10-52% may require repeat BAE. This procedure remains controversial regarding efficacy in non-massive hemoptysis; however, each patient should be assessed based on his/her clinical situation. BAE procedure involves: Catheter insertion into groin (femoral artery), contrast dye is injected into the arterial system, polyvinyl alcohol particles are injected into potential bleeding sites to occlude the blood supply, coils also used to occlude the bronchial arterial supply.

Recently, CF guidelines were published regarding a consensus statement on the treatment of patients with hemoptysis[3]

1 Contact health care provider if: mild hemoptysis (≥5ml) or (< 5ml) and first-ever episode or if persistent or minimal concern and concerned

Although most patients will have mild to moderate bleeding, it is estimated that 4.1% of all CF patients will have at least one episode of massive hemoptysis in their lifetime. Of these patients, 74% will have one episode and 26% will have greater than one episode. Majority (75%) will have first episode after the age of eighteen.

2 Admission to hospital if: moderate to massive hemoptysis or respiratory tract exacerbation with hemoptysis. Each patient should be assessed individually regarding admission.

3 Treatment with antibiotics: Considered if ≥5ml of blood.

4 Non-steroidal anti-inflammatory drugs (NSAIDS) should be discontinued if ≥5ml of blood

5 Airway clearance should be discontinued if: Moderate to massive hemoptysis. No general consensus on strategy if mild- moderate bleed. In these cases, patient should consult CF center for advice,

6 Aerosolized treatments (bronchodilators, inhaled antibiotics, hypertonic saline, Dornase alfa) should be discontinued if: Moderate to Massive hemoptysis. Consider continued treatment if scant hemoptysis

7 Treatment of Massive hemoptysis should include: bronchial artery embolization (BAE) asap. Emergent lung resection has been considered for uncontrolled bleeding after BAE. Bronchoscopy is not recommended before BAE.

In conclusion, hemoptysis is commonly seen in Cystic Fibrosis. It is often unpredictable. However, education, early detection and evaluation, treatment, and early referral for Bronchial Artery Embolization in the setting of massive hemoptysis can create control in the majority of patients.

1. Efrati O, Harash O, Rivlin J, Bibi H, Meir M-Z, Blau H, Mussaffi H, Barak A, Levy I, Vilozni D, et al. Hemoptysis in Israeli CF patients. Prevalence, treatment, and clinical characteristics. J Cyst Fibros 2008;7: 301-306

2. Flume PA, Yankaskas JR, Ebeling M, Hulsey T, Clark LL. Massive hemoptysis in cystic fibrosis. Chest 2005;128: 729–738.

3. Flume PA, Mogayze PJ et al. Cystic Fibrosis Pulmonary Guidelines. Pulmonary Complications: Hemoptysis and Pneumothorax. AJRCCM 2010; 182: 299-306.

New Staff Members

PEDIATRIC

SUMIT BHARGAVA, M.D

Dr Bhargava went to medical school in India. He completed his residency in pediatrics at the University of Kansas Medical Center and trained in pediatric pulmonology at the Children's Hospital of Philadelphia. Prior to coming to Stanford and Packard Children's Hospital, he served as Assistant Professor of Pediatrics at Yale University and the Director of the Yale New Haven Children's Hospital Pediatric Sleep Center. At Yale he established the first pediatric Sleep Clinic in the Department of Pediatrics. His research interests include the metabolic correlates of sleep disordered breathing in obese and diabetic children and sleep disordered breathing in chronic pulmonary and neuromuscular disease.

COLE SOUZA, BA

Please join us in welcoming Cole who is our new Pediatric Pulmonary Clinic Coordinator at Packard Children's Hospital. Cole has been working for LPCH for 3 years as one of the schedulers for Respiratory Specialties/ENT Clinics. In his new role he will be responsible for managing clinic flow, creating the Physician's schedules and coordinating the clerical staff for Pulmonary, CF and Transplant. In the interim, Cole will also be assuming the role as the Patient Services Coordinator, until we hire a replacement for Miguel Huerta's position.

ADULT

SUSAN CASSIDY, MS, RN, ACNP-BC, CCRN

Susan spent the first 5 years of her nursing career working in an ICU and immediately developed an interest in organ donation. She had the opportunity to work for 2 different organ procurement organizations in Michigan and California for 10 years. She graduated from UCSF in 2010 with her MS, and started her NP career here at Stanford in IR, transitioned to neurosurgery, and is now very happy to be part of Advanced Lung Disease.

LAURA STARR, RN, MSN, NP

Laura was born and raised in the San Francisco Bay Area. After completing her B.A. in Kinesiology at Occidental College in Los Angeles she attended UCSF for both her R.N and Master's degree specializing as an Acute Care Nurse Practitioner.

Laura joined the Advanced Lung Disease program in 2008 and her role includes inpatient and outpatient medical management as well as patient and caregiver teaching. During her time off, Laura enjoys spending time with her husband and two sons.

Goodbye and Thank You

NICOLE EDEN, RN, MS, CPNP

Nicole started in the CF Center in 2001 as the Pediatric CF Nurse Coordinator. After working full time for 2 years, Nicole went back to school for her advanced degree and took on a per diem role as the Adult CF Nurse Coordinator. She had two children who are now 6 and 4 years old, Zachary and Chloe. Nicole and her family have relocated to the East Bay, where she will take on her full time role as Mom for a while. She will no doubt fill her day with many activities; besides continue on her training for triathlons and half marathons. She is quite the athlete!

MIGUEL HUERTA

Miguel started in the CF Center in 2006 as the Patient Services Coordinator for CF and Transplant. He has been a valuable resource for all our CF patients and families and will be greatly missed by all. Miguel has accepted a position with the IT department at Packard Children's Hospital working on developing a new medical information system called EPIC.

We will miss Nicole and Miguel's professionalism and dedication to the CF patients and their families. We wish them both the best in their new endeavors.

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Adult CF Advisory Council (ACFAC)

STANFORD ADULT Cystic Fibrosis Advisory Council

In partnership with the CF Center, the Patient Care Services Program, and other clinic Advisory Councils at Stanford, the Adult CF Advisory Council is developing a groundbreaking peer-to-peer mentoring program. Research shows that peer support and one-to-one mentoring can be of tremendous help for people coping with disease and chronic illness. In its initial trial phase, the mentor program will consist of a small group of patients; to track the efficacy of the project, specific data will be gathered, such as FEV1, compliance, and return hospitalizations. This is an exciting opportunity to involve patients in their own care in a new and more direct way.

Website: http://cfcenter.stanford.edu/acfac/

Email: stanfordcfac@gmail.com

Phone: (650) 549-5102

Current Research Studies

Be a part of the cure for CF! Volunteer for a clinical trial today. To learn more, visit http://cfcenter.stanford.edu, contact our research coordinators, or talk to your physician. The following trials are currently underway:

- Sweat testing in newborns with CF
- Phase II study of VX-809 and VX-770
- Advanced Diagnostic Testing for Lung Disease
- Exercise study
- ABPA study
- EPIC trial for early treatment of Pseudomonas
- Lung Clearance Index
- PTC124 Phase III

CYSTIC FIBROSIS CENTER AT STANFORD
Pediatric Providers at Packard Children's: Carlos Milla, MD, Center Director; Carol Conrad, MD; David Cornfield, MD; John Mark, MD; Richard Moss, MD,; Terry Robinson, MD; Nanci Yuan, MD; Jacquelyn Zirbes, DNP, RN, CPNP.

Clinic Scheduling	(650) 497-8841
Clinic & Prescription Refill FAX	(650) 497-8791
Cole Souza, Patient Services Coordinator	(650) 498-2655
Mary Helmers, Nurse Coordinator	(650) 736-1359
Kristin Shelton, Respiratory Therapist	(650) 724-0206
Julie Matel, Nutritionist, Dietitian	(650) 736-2128
Lindsey Evans, Social Work	(650) 736-1905
Jacquelyn Zirbes, Newborn Screening Coordinator	(650) 721-1132

For Urgent Issues:
Monday-Friday, 8:00 AM to 4 PM, contact RN Coordinator
All other times, for children call (650) 497-8000

Adult providers at Stanford: David Weill, MD, Adult Program Director; Paul Mohabir, MD; Gundeep Dhillon, MD; Camille Washowich, MSN, ACNP; Elika Rad, RN, MSN, NP, Kelly Johnson RN MSN NP, Susan Cassidy RN, MSN, NP, Laura Starr RN, MSN, NP

Clinic Scheduling		(650) 736-5400
Clinic & Prescription Refill FAX		(650) 723-3106
Kathy Gesley, Nurse Coordinator	Office	(650) 498-6840
	Patient Line	(650) 736-1358
Carol Power, Respiratory Therapist		(650) 736-8892
Lara Freet, Registered Dietitian		(650) 721-6666
Meg Dvorak, Social Work		(650) 723-6273

For Urgent Issues:
Monday-Friday, 9:00 AM to 4:00 PM, after hours call SUH, (650) 723-4000 and ask for Pulmonary Fellow on-call.

Research:

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