Lucile Packard Children's Hospital at Stanford

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STANFORD SCHOOL OF MEDICINE
Stanford University Medical Center

CYSTIC FIBROSIS CENTER NEWS

Integrative Medicine and Cystic Fibrosis

BY JOHN MARK, MD

The National Institutes of Health (NIH) defines complementary and alternative medicine (CAM) as a group of diverse medical and health care systems, practices, and products that are not generally considered part of conventional medicine. Integrative medicine is defined as relationship-centered care that focuses on the whole person, makes use of all appropriate therapeutic approaches to achieve optimal health and healing, and includes the best of evidence-based CAM therapies and evidenced-based conventional therapies.

Rates of reported CAM usage among children vary between studies, but are approximately 10-40 percent of healthy children and more than 50 percent of children with chronic, recurrent, or incurable conditions. The use CAM is most often in conjunction with conventional care. It is estimated that 33 percent to 66 percent of the CF population use or have used non-traditional therapies. Use of herbal or dietary supplements (so called natural products) alone is the most popular non-conventional therapy which is similar to the general population. One questionnaire study in adults with CF asked about dietary supplements use found 19 percent of patients were currently using dietary supplements and 10 percent had reported past use. An area of concern for health care team is that the intake of such supplements (in addition to other non-conventional therapies) is not always discussed with their health care providers. This could be a problem since many supplements may have interactions with other medications that are also being taken so-called "drug/herb" interaction.

An approach using integrative medicine for children, adolescents and adults with chronic lung problems such as cystic fibrosis may provide an opportunity to decrease symptoms such as cough, shortness of breath, difficulty maintaining weight and help increase an overall feeling of wellness. This integrative approach will not "cure" their CF, but by using these various therapies and lifestyle changes, one could possibly improve breathing, increase exercise capability and decrease the need for some conventional medications (which may decrease the chances for potential side effects).

The integrative approach of using conventional therapies along with complementary and alternative medicine (CAM) would ideally start at a young age or when the disorder is first diagnosed. Since there is a significant inflammatory component to CF, conventional medications and therapies can often rapidly improve these symptoms. CAM therapies such as exercise, nutritional changes, environmental control and other such therapies tend to act over a much longer period of time but with conventional therapies may help decrease or slow the progression of CF, especially lung function, and help maintain better nutrition. These various approaches would optimize the goals of therapy and allow the

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.

Integrated Medicine story continued from page 1

person with CF to live as active and normal a life as possible.

The degree of impact that some CAM therapies-such as acupuncture and dietary supplements-may have over a prolonged period of time is not yet clear. However, studies to assess their safety and efficacy are being conducted and will hopefully assist health care providers in counseling patients and families as to which ones may best be suited to that person's particular cystic fibrosis issues. A study that is being considered here at Stanford and Packard Children's is using acupuncture to help chronic pain in older children and adults with cystic fibrosis. The occurrence of pain in adult CF patients is well documented with approximately 85 percent of adult patients reporting significant pain in the previous month of being asked. Of the adult CF patients experiencing pain, 70 percent reported that pain altered their quality of life. Recent studies have also shown that up to 59 percent of pediatric CF patients are also experiencing significant pain. Despite the high incidence of chronic pain, few patients reported this to their physicians resulting in infrequent use of pain medications. The infrequent use of pain medication is also partially explained by the fact that many physicians are hesitant to prescribe certain medications such as opioids to CF patients because the side effects of these medications may exacerbate pre-existing gastrointestinal problems such as constipation, which could lead to distal intestinal obstruction syndrome (DIOS). It seems that a more effective treatment strategy would be helpful in order to manage chronic pain in CF patients.

Medical acupuncture has been used for years in order to help alleviate chronic pain various parts of the body. Medical acupuncture has also been shown to be effective as an adjunctive therapy to help alleviate pain in adult CF patients. In a small 2005 study acupuncture was well tolerated and no side effects were reported. However, few studies have been conducted to examine medical acupunctures' ability to reduce pain in pediatric CF patients. In the study being considered here, funded by a grant from the Tracie Lawlor Cystic Fibrosis Foundation, we are hypothesizing that medical acupuncture can be used as an adjunctive treatment in pediatric and adult patients with CF in order to help alleviate pain and potentially could help maintain or improve lung function (FEV1 – a measurement of lung function in CF patients). The idea behind this proposed study was supported by a recent 2007 report using non-segmental acupuncture in patients with asthma who improved their in FEV1. Recently in a larger study in the journal Chest (2012), 68 adults with chronic obstructive lung disease (COPD) were studied using acupuncture to improve shortness of breath- so-called dyspnea on exertion (DOE). This study clearly demonstrated that acupuncture was a useful adjunctive therapy in reducing DOE in patients with COPD. The Stanford study also hopes to show that medical acupuncture will improve the quality of life of CF patients.

The specific goals of the proposed Stanford study are to determine if four weekly treatments of acupuncture as compared to four weekly treatments of sham (fake) acupuncture is effective in improving pain as measured by the visual analog scale (using numbers 1 to 5 to represent severity) in teenagers and adults with CF. In addition to this goal other areas to be studied are to determine if acupuncture treatments are effective in improving quality of life as measured by the cystic fibrosis questionnaire-revised, to determine if acupuncture treatments are effective in improving weight gain and weight maintenance, to determine if lung function is maintained or improved, and to determine if acupuncture treatments are effective in improving sleep as measured by actigraphy (wrist watch worn by subject to measure restlessness during sleep) along with sleep diaries.

In the study being considered here at Stanford, we are hypothesizing that medical acupuncture can be used as an adjunctive treatment in pediatric and adult patients with CF in order to help alleviate pain and potentially could help maintain or improve lung function.

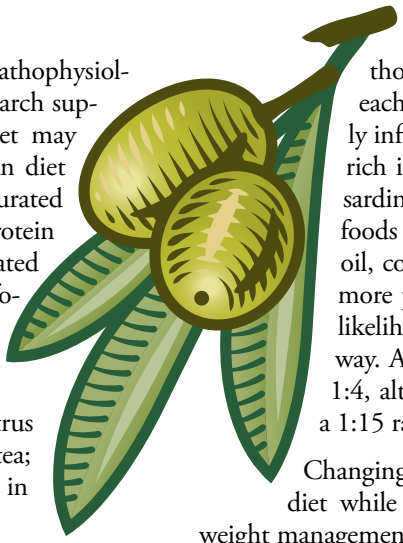
It is important to remember, integrative medicine uses all aspects of health care: preventive, psycho-social, mind-body, natural substances, energy, conventional medications and techniques in reaching the ultimate goal of improving health and wellbeing. Using an integrative approach in the treatment of cystic fibrosis patients may help in many ways that are now starting to be studied. One example is the use of acupuncture as discussed earlier. Patients and families are welcome to discuss integrative approaches with their CF health care provider and, in addition may make an appointment with John Mark, MD, in his "integrative medicine pulmonary clinic" at (650) 724-4788 (adults welcome).

One common question may be: where does one start? For children through adults who have mild to even severe disease, a step-wise approach in addition to conventional care should be considered. These therapies may include lifestyle (improving sleep and exercise programs), nutrition (consider an anti-inflammatory diet and increased intake of "good fats"), cautious use of certain dietary supplements, mind-body therapies (such as relaxation, imagery and biofeedback), manipulative therapies (osteopathic manipulation, chiropractic manipulation and massage) and alternative health care approaches (traditional chinese medicine/acupuncture and homeopathy). Remember, it is important to discuss all types of therapies being used (especially any type of supplements) with the CF health care team. A more in-depth review of such CAM therapies will be forthcoming in future newsletters.

The Role of the Anti-Inflammatory Diet and Cystic Fibrosis

BY LARA FREET, RD

Inflammation is a potential contributor to the pathophysiology of many chronic diseases and cancers. Research supports that following a Mediterranean-style diet may have anti-inflammatory effects. The Mediterranean diet encourages a higher consumption of monounsaturated fats, whereas the traditional high calorie, high protein diet is notable for a higher intake of polyunsaturated and saturated fats. An anti-inflammatory diet also focuses on an increased intake of food sources rich in: omega-3 fatty acids such as salmon, ground flaxseed, sardines, and walnuts; antioxidants including red, yellow, and orange colored vegetables, citrus fruits, dark leafy green vegetables, green and black tea; and consuming foods that are high in fiber found in whole grain, fruits, and vegetables.

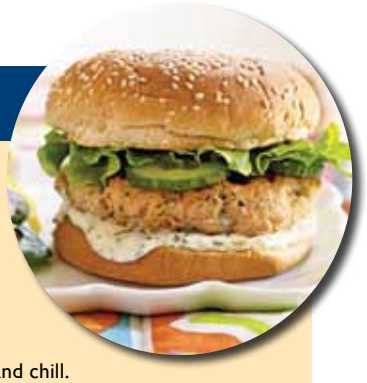


There is concern that following a more traditional high calorie diet promotes a pro-inflammatory response. However, there is no research for patients who have CF to assess the role of the high calorie diet, particularly the fat composition, and the inflammatory response. There is some research on the role of omega-3 fatty acid supplements in the CF population. The most common omega-3 fatty acid supplements used within the United States are fish oil tablets, ground flaxseed, or flaxseed oil. Omega-3 fatty acid supplementation may provide some anti-inflammatory benefit, but given food containing omega-3 fatty acids are higher in fat the pancreatic enzyme regimen may need to be adjusted.

In trying to increase consumption of foods rich in omega-3 fatty acids, the ratio of omega-3 to omega-6 fatty acids in the diet is thought to be more significant than quantity of each fatty acid consumed. The ratio can be positively influenced by increasing the intake of food sources rich in omega-3 fatty acids such as: cold water fish, sardines, and flaxseed, and decreasing the intake of foods rich in omega-6 fatty acids including: rapeseed oil, corn oil, and soybean oil as well as avocado. The more processed and fried food consumed the greater likelihood of the ratio increasing in an undesirable way. A desirable ratio (omega-3:omega-6) is less than 1:4, although the North American diet may closer to a 1:15 ratio.

Changing eating habits to follow an anti-inflammatory diet while trying to maintain a high caloric intake for weight management with CF may be challenging but is achievable with planning. The basis of the anti-inflammatory diet focuses on whole grains and produce; particularly the red, orange, yellow, and green hues, compared to processed flours and grains. It also highlights decreasing intake of whole fat dairy products, butter, margarine, and higher fat meats because of saturated fat composition. Instead protein sources may be achieved through weekly consumption of: cold water fish, soy, beans, legumes, lentils, nuts, and lean meats. Using higher calorie food items such as: olive oil, walnut oil, nuts, nut butters, seeds, and fish the calorie intake can be comparable. The challenges that may arise include if the appropriate foods are available, affordable, early satiety from higher fiber intake, and decreased variety with the limit on high calorie sources and processed foods.

Recipe



SALMON-CILANTRO BURGER

BY JACKIE MILLS, MS, RD FROM *COOKING LIGHT*, MAY 2009

INGREDIENTS

- 1/4 cup reduced-fat mayonnaise
- 1 tablespoon chopped fresh cilantro
- 1 tablespoon fresh lime juice
- 1/8 teaspoon salt
- 1/8 teaspoon freshly ground black pepper
- 1 (1-pound) salmon fillet, skinned and cut into 1-inch pieces
- 1/4 cup dry breadcrumbs
- 2 tablespoons cilantro leaves
- 2 tablespoons chopped green onions
- 1 tablespoon chopped seeded jalapeño pepper
- 2 tablespoons fresh lime juice
- 1/2 teaspoon salt
- 1/4 teaspoon freshly ground black pepper
- Cooking spray
- 4 (1 1/2-ounce) hamburger buns with sesame seeds, toasted
- 12 (1/4-inch-thick) slices English cucumber
- 4 leaf lettuce leaves

PREPARATION

1. Combine first 5 ingredients in a small bowl; cover and chill.

2. Place salmon in a food processor; pulse until coarsely chopped. Add breadcrumbs and next 6 ingredients (through 1/4 teaspoon black pepper); pulse 4 times or until well blended. Divide salmon mixture into 4 equal portions, shaping each into a 3/4-inch-thick patty.

3. Heat a grill pan over medium-high heat. Coat pan with cooking spray. Add patties to pan; cook 2 minutes. Carefully turn patties over; cook 2 minutes or until done.

4. Spread about 1 tablespoon mayonnaise mixture over bottom half of each hamburger bun. Top each serving with 1 salmon patty, 3 cucumber slices, 1 lettuce leaf, and top half of bun.

Enjoy with avocado slices, a spinach salad, and baked sweet potato fries with your favorite dipping sauce.

(http://www.myrecipes.com/recipe/fresh-salmon-cilantro-burgers-10000001891952/)

In Memoriam Clyde Mosier (1941 - 2012)

BY RICHARD MOSS, MD

Clyde Mosier was born and grew up in Memphis and was educated at the University of Mississippi. He married Lillian Link in 1963. He served in the US Air Force from 1964 to 1969, doing three tours of duty in Vietnam. After leaving the military he moved with Lillian to San Jose and worked in the fledgling tech industry at Fairchild Semiconductor, but soon turned his intellectual and interpersonal talents to finance as a stockbrocker for 41 years, both in firms and independently.

Clyde and Lillian's son Ross was born in September 1967, and was soon diagnosed with cystic fibrosis. The Mosiers moved to California to give Ross the best care possible. He received his CF care at the Children's Hospital at Stanford on Sand Hill Road (CH@S), the forerunner to today's Lucile Packard Children's Hospital. In 1972 Ross, not yet five years old, became the national CF Poster Child, and on May 9 of that year Ross and his parents were received in the White House by First Lady Pat Nixon. But such was the state of things in those years that Ross only made it to age 12, and he passed away at CH@S in 1980. I remember those sad days clearly, and know that no child was ever loved better than Ross was loved by Clyde and Lillian.

After Ross' passing Clyde Mosier labored with a fierce and persistent determination to honor Ross in the best way he could conceive: to raise funds to, as he was fond of putting it - "make CF history" - by helping researchers find better ways to control and eventually cure CF. It has been our enduring good fortune at the Packard/Stanford CF Center that Clyde and Lillian decided to place their faith in our Center and devote their fund-raising to help our scientists and clinicians battle CF and give our patients a better and longer life. Over the past 32 years the Ross Mosier Classic golf tournament and banquet, hosting hundreds of participants every last Monday in July, has raised over $3 million for CF research at Lucile Packard Children's Hospital and the Ross Mosier Laboratory for CF Research at Stanford University Medical Center. These funds have been used to support both bench research and clinical trials of new drugs, outcome measures, and modalities of treatment for infants, children and adults with CF in our Center, community and region. Our fundamental orientation as a Center has always been a commitment to translational discovery (i.e., bringing scientific discoveries from the laboratory as quickly and safely as possible to the patient in early and later phase clinical trials) and has encompassed projects from both academic investigator-initiated protocols to biotech/pharmaceutical industry-sponsored programs. The Ross Mosier Classic has played a huge role in this effort for over three decades.

I also must acknowledge the inspiration of Clyde's high school sweetheart, wife and life partner of almost 50 years, Lillian. Besides her dedication to the annual Ross Mosier Classic, as a longtime teacher at St. Andrew's School in Saratoga Lillian also has led a Community Service Cystic Fibrosis Collection and Walk based at the school, and daughter Emily always lent her talents and help to her parents and friends in the fund-raising campaigns.

Clyde was a man of deep faith, a pillar of his Episcopal church. He also had faith in us to advance care of patients with CF and eventually conquer this cruel disease. May we be worthy of it, and him.

CF Parent Advisory Council News

In recent months, the Cystic Fibrosis Parent Advisory Council has worked in partnership with the CF Care Team on many projects, including the development of resources to improve the transition process for teens and young adults; the provision of support groups; the creation of the "Binder Project;" and the offering of "CF College 101," a workshop hosted by the CF Center and the Cystic Fibrosis Foundation (CFF) to help prepare teens and their parents for the transition to college.

CF College 101 offered invaluable tips for college-bound students, most notably the importance of exploring in advance the ability of prospective schools to accommodate students' health needs. Once accepted, it was strongly recommended that students with CF register with the school's office of disabled student services. By doing so, students with CF are often able to have a single room, and will not be penalized for missing class due to illness or doctors' appointments. Often, the office of disabled student services will provide a note-taker to registered students with CF.

There are many scholarship opportunities available to students with CF, notably through the Boomer Esiason Foundation, The Living Breath Foundation and the Cystic Fibrosis Scholarship Foundation. For a complete list, go to http://www.cfri.org/scholarship.shtml.

The Cystic Fibrosis Foundation filmed the workshop, and it will be posted on-line soon. Check the CF Center website and Facebook page for updates. Due to the response to the workshop, it is likely that it will be offered again at CF Education Day in March.

For more information about the Parent Advisory Council, email Siri Vaeth at svaeth@lpch.org.

Distal Intestinal Obstruction Syndrome (DIOS)

BY ELIKA RAD, RN, MS, NP

When referring to gastrointestinal manifestations in cystic fibrosis (CF), patients and caregivers are most familiar with neonatal meconium ileus, exocrine pancreatic insufficiency, and periodic constipation. With the increased longevity of CF patients, we are observing an increased incidence of constipation and distal intestinal obstruction syndrome (DIOS) in the adolescent and adult populations. This is not surprising as meconium ileus, constipation, and DIOS are all interrelated in that they are due to the increase in the thickness and stickiness of the mucus in the intestines as well as prolongation of food transit through the intestines. The lifetime prevalence of DIOS is 8 percent in pediatric CF patients and 16 percent in adult CF patients[5].

Constipation vs DIOS: Distal intestinal obstruction syndrome (DIOS) is a type of constipation caused by a buildup of mucus that lines the intestinal walls. This buildup reduces the inner diameter of the intestines making blockage more likely. You may also hear this condition referred to as "meconium ileus equivalent." The interchangeable use of the terms "constipation" and "DIOS" has made the approach to treatment difficult for clinicians and at times confusing for the patient. DIOS is characterized by complete or incomplete obstruction of the intestine by thick and adhesive fecal material. The classic location is the terminal ileum (end of the small intestine) and proximal colon (beginning of the large intestine). Characteristically DIOS patients have abdominal pain with or without distension as well as vomiting in combination with a right lower quadrant mass, which is palpable and usually seen on plain X-ray of the abdomen. An important differential diagnosis of DIOS is constipation. However, in contrast to DIOS, symptoms of constipation are usually milder and more chronic in nature; most often with constipation regular stooling is not interrupted, but is less frequent or of smaller amount.

In 2011, the European Society for Pediatric Gastroenterology, Hepatology, and Nutrition Cystic Fibrosis Working Group suggested consensus definitions to distinguish DIOS (Table 1) and constipation (Table 2); they also went a step further to differentiate between complete and impending DIOS[5].

TABLE 1

European society for pediatric gastroenterology, hepatology, and nutrition CF working group definition for distal intestinal obstruction syndrome (DIOS) in cystic fibrosis

1. Complete intestinal obstruction as evidenced by vomiting of bilious material and/or fluid levels in small intestine on abdomincal radiography
2. Fecal mass in ileocecum
3. Abdominal pain and/or distension

Compete DIOS: 1, 2, and 3

Incomplete/impedning DIOS: 2 and 3, without 1

TABLE 2

European society for pediatric gastroenterology, hepatology, and nutrition CF working group definition for constipation in cystic fibrosis

1. Abdominal pain and/or distension

2a. Reduced frequency of bowel movements in the past few weeks or months

2b. Increased consistency of stools in the past few weeks or months

3. Symptoms 1 and 2 are relieved by the use of laxatives

Constipation: 1 or 2a or 2b and 3

Contributing Factors: There are multiple factors that can result in both constipation and DIOS.

- Dehydration (systemic and intestinal)
  - Lack of fluid intake
  - Infection/fevers
  - Diarrhea related to antibiotics
  - Surgery/immobility
  - Abnormal regulation of sodium and chloride absorption (expression of CFTR mutation)
  - Medications (anti-cholinergics: Benadryl, etc.)
- Fat malabsorption
  - Frequently missed doses of enzymes
  - Under-dosing of enzymes
- Slow intestinal transit
  - CF diabetes
  - Narcotic use
  - Previous intestinal surgery such as meconium ileus re pair or partial colectomy
  - Intestinal inflammation (new area of research)
- Abnormal intestinal mucins (proteins that line the intestinal wall)
- Defective bile acid uptake
- Dysmotility (abnormal movement of the intestines)

The link between DIOS and a history of meconium ileus surgery has been well described in literature as an alteration in motility as well as narrowing of the bowel[5]. Therefore, when seeing a new CF provider, it is important for the patient to report a history of meconium ileus at birth. This history may affect management of and future prevention of developing DIOS.

DIOS is frequently seen in 10-20 percent of CF patients in the early post-operative lung transplantation period[1]. This is a time marked by dehydration, decreased mobility and use of narcotics for post-operative pain control. A history of meconium ileus or prior bowel surgery (with the presence of scar tissue and adhesions) can easily contribute to this problem[5]. A bowel cleanout out prior to surgery and prophylactic laxative therapy during the peri- and post-transplant period is recommended.

DIOS story continued from page 5

Diagnosis: Plain abdominal Xray of the abdomen is the first line diagnostic tool, which can differentiate between constipation and DIOS. In DIOS (Figure 1)[6] a characteristic bubbly fecal mass can be seen in the right lower quadrant (ileocecum); air fluid levels and dilated small bowel loops (indicative of complete obstruction) may be seen in an upright film. In constipation (Figure 2)[7], the X-ray will show a moderate to large amount of fecal burden throughout the colon.

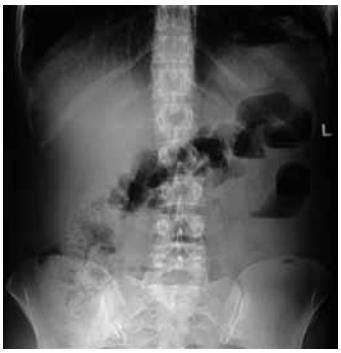


Figure 1: DIOS

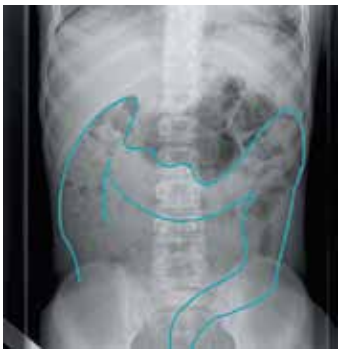


Figure 2: Constipation

To rule out other causes of acute abdominal pain, such as intussusception, ovarian cysts in females, or appendicitis your provider may order a CT scan of your abdomen. An increased incidence of appendicitis in the CF patient, which has not been observed in the literature[5], is a misconception among providers unfamiliar with CF bowel abnormalities. This can lead to unnecessary abdominal surgery, so giving your provider a good history is important. However, the presence of other symptoms, such as peritonitis (which may be seen on CT) and fever with or without a high white blood cell count, may warrant surgical intervention/appendectomy.

Complications, Treatment & Prevention: If left untreated, DIOS with complete obstruction can lead to bowel perforation requiring invasive surgery to include adhesiolysis (removal of adhesions or scar tissue), hemicolectomy (partial removal of the colon) and small bowel resection. It is important to note that invasive surgery is highly contraindicated in patients with poor lung function due to high mortality. With early aggressive medical management by a provider familiar with CF and DIOS surgery can be avoided and is often the last resort.

Medical treatment of DIOS is primarily aimed at reliving the obstructing mass of stool often with a combination of therapies, in a methodological manner[1],[3-5]. If mild, DIOS can be treated at home with oral agents tailored to the patient. Moderate to severe cases will require hospitalization.

- Hydration is essential throughout acute treatment
  - Orally if patient is not nauseated or vomiting
  - Intravenously (IV) in the presence of nausea/vomiting or abdominal distention.
- Decompression
  - Nasogatric (NG) tube
- Colonic lavage fluid orally/NG tube and/or rectally
  - Osmotic laxative containing polyethylene glycol (PEG) – Golytely, Klean-Prep, high dose Miralax.
  - Senna
  - Lactulose and/or acetylcysteine (Mucomyst)
  - Large volume rectal enemas/lavage solutions
- Enema under fluoroscopy - Gastrograffin or hypaque enemas
  - Used for patients at risk for aspiration or those failed above therapies
  - Moves fluid into the intestinal lumen, softening and helping to evacuate hardened stool
  - Should be performed by an experienced radiologist, who can assure that the material reaches the terminal ileum
- Replacement of Electrolytes (potassium, magnesium and phosphorus)

Once a patient has had an episode of DIOS they are at risk for recurrence, thus daily prevention and maintenance of good bowel function using a laxative regimen tailored by your CF provider is essential. Many CF patients self-treat with ineffective methods, use medications that their cohorts use, or try to ameliorate their symptoms to avoid hospitalization. Many of the medications, if taken incorrectly, have side effects that may contribute to or worsen symptoms thus consulting your CF team is key. As DIOS and constipation have multifactorial causes, their prevention also requires a multifactorial and interdiscipinary approach[1],[5]:

- Avoidance of dehydration
- Daily laxative (Miralax, Senna, lactulose)
- Optimization of pancreatic enzyme dosing
- Optimization of blood sugar control in diabetic patients
- Treatment of infections
- Control of acid reflux
- May require referral to GI specialist for further diagnostics

Constipation and DIOS are significant issues that occur in the pediatric CF patient, but are more prevalent in the adult CF population. Early intervention during acute symptoms and prevention of reoccurring symptoms are important in maintaining your bowel health. For prevention of symptoms, please consult your CF provider to discuss an approach that is tailored to you and your medical history.

1. Colombo C, Ellemunter H, Houwen R, et al. *Guidelines for the diagnosis and management of distal intestinalobstruction syndrome in cystic fibrosis patients.* Journal of Cystic Fibrosis. 2011; 10(2): S24–S28

2. Dray X, Bienvenu T, Desmazes-Dufeu N, et al. *Distal intestinal obstruction syndrome in adults with cystic fibrosis.* Clin Gastroenterol Hepatol. 2004;2:498–503.

3. Houwen RH, van der Doef HP, Sermet I, et al; ESPGHAN *Cystic Fibrosis Working Group. Defining DIOS and constipation in cystic fibrosis with a multicentre study on the incidence, characteristics, and treatment of DIOS.* J Pediatr Gastroenterol Nutr. 2010; 50(1):38-42.

4. Khan K, Schwarzenberg SJ. *Gastrointestinal Disease Associated with Cystic Fibrosis.* US Respiratory Disease. 2007 Oct: 42-47.

5. van der Doef HP, Kokke FT, van der Ent CK, Houwen RH. *Intestinal Obstruction Syndromes in Cystic Fibrosis: Meconium Ileus, Distal Intestinal Obstruction Syndrome, and Constipation.* Curr Gastroenterol Rep. 2011 Jun; 13(3):265-70.

6. http://www.health-reply.com/distal-intestinal-obstruction-syndrome/

7. https://www.mja.com.au/journal/2005/182/5/5-constipation-and-toileting-issues-children

Pediatric CF Center Update

MARY HELMERS, RN

Our **Quality Improvement Project** (aka: the CF Binder project) has started the second phase: addressing the educational needs of our patients and families. Once again the CF Team would like to say "thank you" to all our families and patients who have filled out the questionnaires. These questionnaires help us to evaluate the areas in which we need to focus our patient/family teaching. We appreciate all your efforts! In the coming weeks we will start our educational piece of the project. After review of all the educational questionnaires, our teaching efforts will focus on respiratory and nutritional aspects of CF. We will provide handouts and/or internet links of teaching materials to assist our patients with their learning.

Transition Update: The Pediatric and Adult CF Programs have been working together improve our patient transition process. We have quarterly meetings to discuss the patients who will be transitioned within the following month or two. Members of the Adult Program: Paul Mohabir, MD, Kathy Gesley, RN, and Meg Devorak, LCSW come to the last visit at the Pediatric Clinic, and Mary Helmers, RN attends the first clinic visit in the Adult Clinic. Once it has been determined that the pediatric patient is ready for transition, a referral is sent over to the Adult clinic to help expedite the process. It is our hope that the patient will be seen within one month from their last pediatric appointment.

Reminder: We have new turquoise colored masks that 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 walking 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.

We would like to acknowledge the Parent Advisory Council for all of their help with our QI project, College 101 day and numerous other projects that the Pediatric CF Center has embarked upon. Thank you!

Adult CF Center Update

KATHY GESLEY, RN

- Teaching is available every Tuesday 12 -1pm and Friday 11am -12pm through the Dietician and Coordinator for a variety of education topics related to Cystic Fibrosis. You may request an appointment by contacting or emailing either team member.
- My Health sign up with your next clinic visit will permit us to review and respond to your questions about your clinic lab work and request medication refills. Completed annual diagnostic tests will be available for review online.
- After each clinic visit you should receive a copy of your patient instructions and medication list. The instructions will include needed tests to be completed for the calendar year.
- Flu shots are recommended and available at each clinic visit.
- Peer2Peer is a new Stanford Hospital sponsored program to provide one on one support in navigating your chronic illness. Trained patient volunteers can regularly contact you for support and suggest resources for you. Please contact an Adult Team member to be referred to this program.
- Our Stanford Cystic Fibrosis Center is participating in a Leadership Development Program through the Cystic Fibrosis Foundation. The leadership team completed self-assessments and peer reviews this summer, and met with the Center mentor in October at the NACFC national conference. The Leadership Program includes regular conference calls and online tasks. Our center goal is to improve mutual respect and communication within the entire team. Carlos Milla, MD, Paul Mohabir, MD, and both clinic Coordinators are working to improve and promote joint projects related to assessment and support of patients. Our center was one of ten other programs selected for this yearlong effort.

Current and Upcoming Research Studies

- Sweat testing in newborns with CF
- Advanced Diagnostic Testing for Lung Disease
- Exercise study
- ABPA study
- EPIC trial for early treatment of Pseudomonas
- Lung Clearance Index
- E-ICE, study of Pulmonary Exacerbation utilizing home FEV1 monitoring
- Vertex 770-110/12 (for the R117H genotype only)
- Saliva Testing
- KaloBios (IV Anti Pseudomonas antibody study) – enrolling Feb 2013
- Gilead (cycling antibiotic study) – enrolling Feb 2013
- Phase III study of VX-809 and VX-770 -enrolling March 2013

For more information please contact our research coordinators or talk to your physician.

New Pediatric Staff Member

Elizabeth Foley, MD is a bay area native that grew up in San Carlos. She attended UCLA and completed her undergraduate studies in psychobiology. She then moved to New York city, where she attended New York Medical College for medical school, and completed her residency in general pediatrics at Cohen children's medical center at Long Island Jewish hospital. She is very excited to be returning to the bay area with her husband and baby boy. She cannot wait to meet you and work with you all in the future. When not in the office, Dr. Liz enjoys shopping, hiking and reading (her recent favorite is Goodnight Moon).

CF Center at Stanford
770 Welch Road, Suite 350
Palo Alto, CA 94304

Adult CF Advisory Council

Do you have trouble getting to clinic? Need assistance with transportation? In partnership with the CF clinic, as part of Stanford's Patient/Family-Centered Care initiative, the Adult CF Advisory Council (ACFAC) is developing a project to help patients get to their all-important quarterly appointments. This project involves fundraising for patient transportation and information on transit options.

ACFAC is also launching the Peer2Peer mentor program, the first of its kind at Stanford, in partnership with the heart transplant advisory council and Guest/Volunteer Services. Adult CF patients will be matched together for one-to-one, confidential, telephone mentorship on any issues that interest them: challenges and triumphs of chronic illness, adjusting to new diagnoses, and living with CF. Stay tuned for further details or contact ACFAC to learn more.

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

Email: stanfordcfac@gmail.com

Phone: (650) 307-3714
(Laura Steur, Chairperson, ACFAC)

CYSTIC FIBROSIS CENTER AT STANFORD

Pediatric Providers at Packard Children's: Carlos Milla, MD, Pediatric CF Center Director; Sumit Bhargava, MD; My My Buu, MD; Carol Conrad, MD; David Cornfield, MD; Christin Kuo, MD; John Mark, MD; Richard Moss, MD; Terry Robinson, MD; Nanci Yuan, MD; and Jacquelyn Zirbes, DNP, RN, CPNP.

Clinic Scheduling	(650) 724-4788
Clinic & Prescription Refill	FAX (650) 497-8791
Erica Oliva, 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 Martins, Social Work	(650) 736-1905
Jacquelyn Zirbes, Newborn Screening Coordinator	(650) 721-1132

For Urgent Issues:
Monday-Friday, 8am to 4pm, contact RN Coordinator (650) 736-1359
All other times, for children call (650) 497-8000 main hospital number

Adult providers at Stanford: Paul Mohabir, MD, Adult CF Center Director; David Weill, 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) 725-7061
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, 9am to 4pm, after hours call SUH, (650) 723-4000 and ask for Pulmonary Fellow on-call.

Research:

Colleen Dunn, Zoe Davies, Cassie Everson	(650) 736-0388

Visit our Website 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

Editor: Zoe A. Davies, RN, MS, PNP, CCRC · Assistant Editor: Colleen Dunn, RRT, RPFT, CCRC