

# CYSTIC FIBROSIS CENTER NEWS

## New Mental Health Screening Guidelines recommended by the CF Foundation

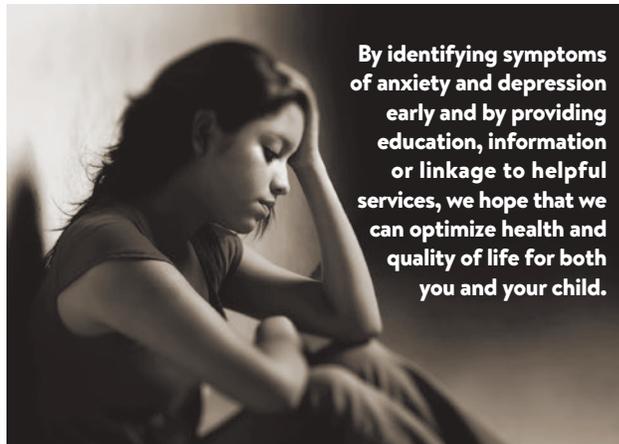
BY SRUTHI VEERAVALLI, MSW, DIANA NARANJO, PHD, SABRINA KARCZEWSKI, PHD

In 2013, the Cystic Fibrosis Foundation and the European Cystic Fibrosis Society convened an expert committee, the Guidelines Committee on Mental Health (GCMH), to develop clinical care recommendations for anxiety and depression in individuals with cystic fibrosis (CF) and parent caregivers. The recommendations are based on the significant findings of The International Depression Epidemiological Study (TIDES), released in 2014 (Quittner et al, Thorax 2014; 69:12 1067-1068).

TIDES was conducted in Europe and the USA over a three-year period. Two brief screening measures—the Depression and Anxiety (HADS) and the Center for Epidemiological Studies–Depression (CES-D)—were administered to individuals with CF, ages 12 and older, and caregivers of children with CF, birth to 18. Measures were completed during a stable, routine clinic visit; demographic and health information were collected and verified via chart review.

Psychological screening measures were completed by 6,088 individuals with CF and 4,102 parents. Elevated symptoms of depression were found in 10 percent of adolescents, 19 percent of adults, 37 percent of mothers, and 31 percent of fathers. Elevations in anxiety were found in 22 percent of adolescents, 32 percent of adults, 48 percent of mothers and 36 percent of fathers. Overall, elevations were 2-3 times the rates reported in community samples (Quittner et al.).

Analyses of comorbid symptoms indicated that adolescents reporting depression were 14.97 times more likely to report anxiety; adults elevated on depression were 13.64 times more likely to report anxiety; mothers with elevated depression were 15.52 times more likely to report anxiety; fathers with elevated depression were 9.20 times more likely to report elevated anxiety. Significant differences were found by patient age (depression: adolescents 19 percent vs. adults 29 percent; anxiety: adolescents 22 percent vs adults 32 percent). Mothers reported more symptoms of depression and anxiety than fathers, respectively (depression 37 percent vs. 31 percent; anxiety 48 percent vs. 36 percent). Con-



**By identifying symptoms of anxiety and depression early and by providing education, information or linkage to helpful services, we hope that we can optimize health and quality of life for both you and your child.**

cordance between 1,122 parent-teen dyads indicated that adolescents were 2.32 and 2.22 times more likely to be elevated on depression and anxiety, respectively, if a parent was elevated.

Perhaps the most significant finding was that elevated depression of either adolescent or parent was associated with decreased quality of life, decreased adherence to therapies, and decreased respiratory status. Depression was also associated with increased number of hospitalizations, increased health utilization, and increased healthcare costs. Clearly, mental health is an area that needs

more attention in CF programs.

To better understand the context of mental health care delivery, the GCMH distributed an online survey to approximately 4,000 CF health professionals of which there were 1,454 respondents. In the US, responsibility for mental health issues was predominantly undertaken by social workers, whereas in Europe, psychologists handled this responsibility. However, the majority did not have a colleague trained to manage mental health issues and over 20 percent of respondents had no one on their team whose primary role was mental health.

Additionally, 73 percent of respondents did not have any experience in screening for anxiety and depression. The survey highlights the importance of standardizing the screening of mental health in CF programs around the world as well as training staff, if necessary, to conduct the screenings.

Starting this year, the CF Foundation recommends that CF

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*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.*

*Mental Health Screening Guidelines story continued from page 1*

centers screen every CF patient ages 12 and up, as well as parent caregivers for anxiety and depression, on an annual basis and additionally as needed. The screening tools to be used are the Patient Health Questionnaire (PHQ-9) and the Generalized Anxiety Disorder (GAD-7) screening tool, both of which are well validated tools used frequently in primary care settings. These tools are easy to understand and take very little time to complete. They can be used as a platform to discuss concerns and increase awareness of the strong impact of mental health on medical treatment. Patients with concerning screening scores can be referred to treatment and monitored more closely in follow up visits. In addition, our clinic will be assessing quality of life, as research has demonstrated how chronic illness can impact the quality of life across multiple domains.

In order to support this continued focus on mental health in each center, the CF Foundation has committed funds toward implementation and the sustainability of these guidelines. Out of the 150 applications submitted to the CF Foundation Stanford, across both pediatric and adult centers was awarded this prestigious award for three years to build the program.

Our new clinical psychologist, Dr. Diana Naranjo will work with the team to build a comprehensive screening program that includes not only identifying symptoms of anxiety and depression early, but also providing education, information and linkages to helpful services. This promotes a more comprehensive standard of care for all CF patients and ensures that those patients most at risk for depression and anxiety get the treatment they need. If you have any questions about these new guidelines or would like to request a screening at any time, please ask your CF team.

## Stanford Adult Cystic Fibrosis Advisory Council BY BRIAN EDDY



### STANFORD ADULT Cystic Fibrosis Advisory Council

The adult advisory council has completed several projects to assist the CF community at Stanford, including the publication of an Issue Management Tree to assist patients on escalating issues they may experience with the healthcare team, as well as a pilot program providing valet parking for patients requiring oxygen. Work continues on exciting projects such as the use of patient virtual clinic visits to complement the normal clinic visits and the collection of real life stories to be utilized as a tool to help clinic staff learn from these particular incidents. If you are interested in participating on the council, we are always looking for new members and would love to have you join us. For more information please go to <http://cfcenter.stanford.edu/acfac/>

## The Cystic Fibrosis Parent Advisory Council BY KIRSTEN MCGOWEN

### Cystic Fibrosis Parent Advisory Council: What Do You Need? We Can Help!

*The CF Parent Advisory Council seeks to address the needs of all families seen at the Pediatric CF Clinic at Lucile Packard Children's Hospital Stanford. Here are some reminder and tips for successfully managing CF.*

#### 1. Bring your CF Binder AND Action Plan to your Appointments.

Think of your CF Binder as a one-stop shop for your child's care. It contains contact names and phone numbers, calendars to track illnesses and appointment dates, dividers for supplemental information by category (Action Plans, GI/Nutrition, PFTs, etc.), as well as educational handouts. It is critical to bring your most recent Action Plan to clinic to discuss any changes in care since your last visit.

#### 2. Carry your CF Passports: Available in English and Spanish

Don't forget to use your purple CF Passports whenever you visit another clinic or the hospital to ensure proper infection control. Additional passports are available in clinic.

#### 3. Get Your Voice Heard! We Need Your Input!

Do you have feedback for the council or a project you would like to be addressed? Is there something in the binders that you want to see that would help you with your child's care? Is there something that you do that might benefit all the CF families at Stanford that you want to share? We are always seeking input and feedback; nothing is too big or small. Please contact Kirsten McGowan at [kmcgowan@stanfordchildrens.org](mailto:kmcgowan@stanfordchildrens.org).

#### 4. CF Parent Tip of the Day

It's cold and flu season! Travel safely during the holidays by requesting to pre-board your flight and sanitize your seat area with disinfecting wipes. Think seats, armrests, tray tables, windows, media touch screens – anything your child might touch. Remember seatback pockets are one of the dirtiest areas on a plane! Carrying hand sanitizer for bathroom trips is always a plus, too.

*If you have feedback or an idea/topic for the CF Parent Advisory Council, please email Kirsten McGowan, Co-Lead Parent, at [kmcgowan@stanfordchildrens.org](mailto:kmcgowan@stanfordchildrens.org)*

# Adult Mental Health Coordinator By Meg Dvorak, LCSW

The adult program is excited to announce the award of a prestigious grant from the CF Foundation to fund a new Mental Health Coordinator (MHC). The MHC will ensure the full implementation of the current mental health prevention, screening and treatment of depression and anxiety in CF patients. A faculty psychiatrist, specializing in Psychosomatic Medicine with expertise in CF and lung transplant, will be supported in her role as an embedded psychiatrist within the CF clinic and act as the MHC via this grant.

In terms of prevention, the MHC will oversee and ensure implementation of the following interventions:

- Monthly teleconference CF support group
- Monthly web-based writing group for interested individuals
- Dissemination of the literature on behavioral approaches to reduce distress and anxiety surrounding medical procedures
- Dissemination both in written and seminar form of education regarding depression and anxiety

Our MHC will also ensure that all patients receive at minimum once annual screening with GAD-7, PHQ-9, CF Questionnaire, and Adherence Questionnaire. To streamline the procedure, screening will take place:

1. During the first quarter of the year for all CF patients attending regular clinic appointments

2. During annual visit for transplant patients

3. Towards the end of hospital admission for patients who are lost to follow up in clinic

4. When there is clinical suspicion for depression or anxiety

In addition, the MHC will ensure that all patients will receive appropriate psychological care. Patient needs will be established by screening and immediate in-depth evaluation when indicated and services provided based on acuity. Care will consist of:

- Evidence-based psychotherapy provided by embedded psychiatrist and/or social worker
- Evidence-based medication management provided by our embedded psychiatrist
- Identification and coordination of additional resources when needed (e.g., inpatient hospitalization, partial hospitalization program, identification of support groups and systems)
- Referral to psychological/mental health providers closer to patient's residence if preferred and indicated



## New Staff Members:

### New Pediatric Staff Member



**Diana M. Naranjo, PhD**, is an Assistant Professor of Psychiatry at Stanford, Lucile Packard Children's Hospital. As a trained pediatric and adult psychologist working in behavioral medicine, Dr. Naranjo focuses on the psychosocial needs of patients and families with diabetes. She is a licensed clinical psychologist and a part of both the diabetes care team and cystic fibrosis care team at Lucile Packard Children's Hospital. In both research and clinical work, she aims to understand barriers and facilitators to care, work toward an integrative mental health screening program, understand what developmental demands are important as adolescents transition to adulthood, and how best to provide services that engage youth and their families. Furthermore, as a Latino-American fluent in Spanish, much of Dr. Naranjo's clinical work focuses on bridging the health care gap for underserved ethnic minority youth with chronic illness.

### New Adult Staff Members



**Natalie Chan, RD**: Natalie is originally from the Bay Area, born and raised in San Jose. She received her degree in Clinical Nutrition from UC Davis, before completing her dietetic internship in Sarasota, Florida. She worked with Cardiac and Thoracic patients at UC Davis Medical Center for 2 years before she started working at Stanford Health Care this past April. Her hobbies include: cooking, yoga, hiking,

and arts and crafts. Outside of work you can find her outdoors or trying new restaurants. She is excited to work with the Cystic Fibrosis population because it is rewarding to work with such a tight knit community, and fun, engaged patients and their families!



**Laveena Chhatwani, MD**: Dr. Chhatwani recently joined the Adult Cystic Fibrosis Team at Stanford. A graduate of the Stanford Lung and Heart-Lung Transplantation fellowship, Dr. Chhatwani returns to Stanford with a clinical focus in CF, advanced lung disease and lung transplantation. She looks forward to meeting each of our patients in clinic and working with

them as a team to manage CF. Along with her rich clinical experience and background in clinical research, she brings her strong commitment to excellence and to patient-centered care.



**Meredith Wiltse, NP**: Meredith joins us from Montana where she worked for many years in internal medicine. She earned her Master's in Nursing at the University of Washington and has traveled extensively around the world with her husband, a photojournalist. They have two grown sons and now live near the coast. Meredith misses skiing, but is happy to have traded

long, snowy winters for perennial gardening in a more temperate climate and year-round hiking.

# Reproductive Health in Cystic Fibrosis Part I:

## THE BASICS: ANATOMY AND FERTILITY BY JENNIFER CANON, NP

Given the advancing lifespan of our Cystic Fibrosis (CF) patients, the issues concerning fertility and reproduction are increasingly important. With this in mind, this three-part series is dedicated to everything patients may need to know regarding reproductive health. This first section will discuss the general basics and will include:

1. Reproductive anatomy and hormonal cycles
2. The changes in anatomy that may occur as a result of CF mutations
3. Overall fertility in CF patients

The second and third sections, which will be included in future newsletters, will discuss Contraception, Reproductive Health Screening, Family Planning (for example, In Vitro Fertilization and Genetic Counseling) and Pregnancy in CF.

### THE BASICS: ANATOMY

#### The Female Reproductive System

The female reproductive system includes both internal and external structures:

##### INTERNAL STRUCTURES:

- o **Vagina:** A fibro-muscular tract, which leads from the uterus to the exterior of the body. It is also known as the birth canal.
- o **Uterus:** Also known as the womb, the uterus is a pear-shaped organ that holds a developing pregnancy. It is made of two parts: the cervix, which is the lower part of the uterus that opens to the vaginal canal, and the main body of the uterus, which holds a developing pregnancy. The lining of the uterus is called the endometrium.
- o **Fallopian Tubes:** Two tubes (one on the left and one on the right), which lead from the ovaries to the uterus. During intercourse, sperm from the male travel up through the vagina and uterus to the fallopian tubes where fertilization of an egg occurs. The fallopian tubes then act as the pathway by which a fertilized egg travels back down to the uterus for implantation.
- o **Ovaries:** Two organs (one on the left and one on the right of the uterus), which are responsible for the production and release of eggs (ova) as well as the secretion of hormones.

##### EXTERNAL STRUCTURES:

- o **Labia Major and Minora:** Skin that encloses and protect the vaginal and urethral openings.
- o **Bartholin's Glands:** Located next to the vaginal opening, these glands produce fluid secretions.
- o **Clitoris:** A small, sensitive protrusion where the labia majora meet. The clitoris is an important part of sexual stimulation.

#### The Male Reproductive System

The male reproductive system includes both external and internal structures

##### EXTERNAL STRUCTURES

- o **Penis:** The male reproductive organ for sexual intercourse. It serves as the passageway for urine, pre-ejaculate and semen out of the body.
- o **Scrotum:** Skin that encases the testicles

##### INTERNAL STRUCTURES: Please refer to figure 1 below

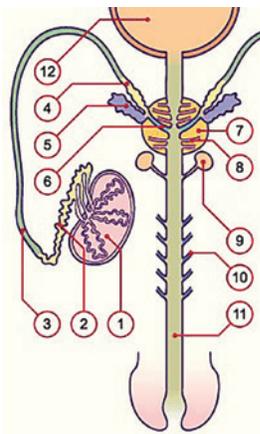


figure #1

- o **Testicles (1):** Located within the scrotum, the testicles are two glands that produce immature sperm as well as sex hormones.
- o **Epididymis (2):** A long, coiled tube that sits atop the testicles. It is the location where sperm from the testicles mature (become more compact and obtain the ability to be mobile). The mature semen is then stored in the epididymis until ejaculation.

- o **Vas Deferens (3+4):** A long, narrow tube that carries sperm from the epididymis to the seminal vesicles.

- o **Seminal Vesicles (5):** Two small organs that produce seminal fluid, which is a sugar-rich fluid that provides sperm with energy and helps with the sperms' ability to move. The fluid from the seminal vesicles makes up the majority of ejaculate.

- o **Ejaculatory Duct (6):** A duct, which is formed by the union of the Vas Deferens and the duct of the seminal vesicle. The ejaculatory duct passes through the prostate to the urethra.

- o **Prostate (7+8):** A walnut-sized gland located below the urethra, the prostate produces a fluid that helps sperm move through the male reproductive tract

- o **Cowper's Glands (9):** Located beneath the prostate, these glands also produce a fluid that prepares the urethra for ejaculation/passage of sperm.

- o **Urethra (10+11):** The tube that carries urine from the bladder to the outside of the body. In men, it serves the additional purpose of expelling semen during ejaculation.

### THE BASICS: HORMONAL CYCLES

#### The Menstrual Cycle

The menstrual cycle occurs within the female reproductive system monthly. Its purpose is to prepare a woman's body for a potential pregnancy. The cycle progresses in response to very specific hormones, including follicle-stimulating hormone, luteinizing hormone, estrogen and progesterone. A healthy menstrual cycle is imperative to fertility and the ability to become pregnant. The phases of the menstrual cycle include: (Please refer to figure 2 on next page)

1. **The Follicular Phase:** This phase starts on the first day of your period and results in the maturing of approximately 15 to 20 eggs in the ovaries. Each egg is encased in a small shell, which is called a follicle. At the end of this cycle, one dominant follicle has developed.
2. **Ovulation:** Occurring on approximately day 14 of the menstrual cycle, the dominant follicle releases its egg into the fallopian tube, where it awaits sperm for fertilization.
3. **The Luteal Phase:** During this phase, the uterus thickens and prepares itself for pregnancy. If intercourse has occurred and sperm has fertilized the egg, the fertilized egg travels through the fallopian tube to implant in the uterus. If the egg is not fertilized, it passes through the uterus. When the uterus is not going to support

Reproductive Health story continued from page 4

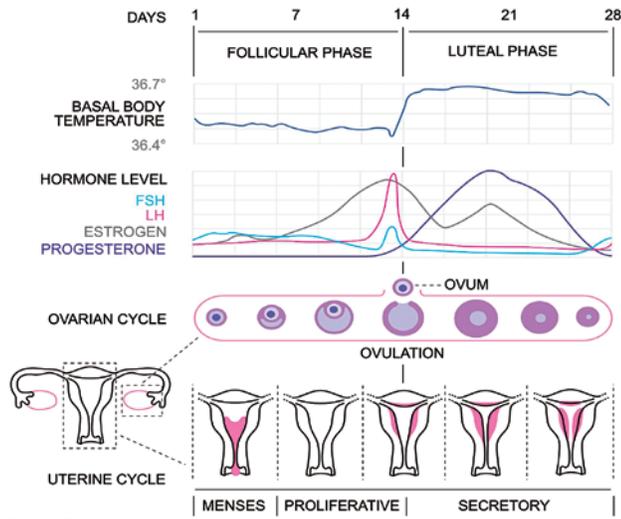


figure #2

a pregnancy, the lining breaks down and sheds. This lining is made up mostly of blood, as well as cervical mucus, vaginal secretions, and endometrial tissue, and takes a few days to shed completely each month. When this period is complete, the cycle begins again.

**THE BASICS:**

**Cystic Fibrosis Mutations and the Effect on the Reproductive System**

**INTRODUCTION**

Cystic Fibrosis arises from a genetic mutation on chromosome 7 resulting in defective production of the cystic fibrosis trans membrane conductance regulator protein (CFTR protein). The CFTR is a protein channel that regulates the passage of chloride (and as a result, sodium and water) across cell membranes. This mutation results in thicker secretions throughout the respiratory system, sweat glands, GI tract as well as reproductive organs.

**EFFECT ON THE FEMALE REPRODUCTIVE TRACT**

CFTR protein is found throughout the female reproductive system, including the cervix, the endometrium, and the fallopian tubes.<sup>2</sup>

- o **Cervix:** CFTR protein is found in large quantities on the cervix. The cervix produces mucus, which changes in consistency throughout the menstrual cycle. In patients without CF, this mucus becomes thinner during ovulation to allow easier passage of semen into the uterus. In patients with CF, there is evidence to support that this change in consistency may not occur. This results in a persistent, thick mucus plug throughout the menstrual cycle, which can make it more difficult for sperm to travel into the uterus<sup>2</sup>.
- o **Endometrium and Fallopian tubes:** CFTR protein is present but little data exists to support that its presence makes a significant impact on fertility. This may be related to a low expression of CFTR protein in these organs<sup>2</sup>.

**FEMALE NUTRITION AND THE REPRODUCTIVE SYSTEM**

It is important to include the role of nutrition in the development of a healthy reproductive system, specifically a monthly menstrual cycle. The relationship between nutrition, body composition and menstruation is fairly complex. Research supports that CF patients on average experience a 12-month delay in the onset of menstruation (average

age of menarche in CF is approximately 14.2-14.9 years of age versus 13 in non-CF patients). The strongest risk factor for delayed menstruation is a patient's weight.<sup>2</sup> Malabsorption and malnutrition is a substantial concern in CF, which can result in difficulty gaining weight, which may ultimately result in delay of menstruation. The good news is that although the menstrual cycles may be delayed in CF patients, once menstruation does begin, the majority of patients develop normal monthly cycles. It is important to note that, as with patients who do not have CF, menstrual irregularities (such as irregular cycles and absence of menses) can always occur.

**EFFECT ON THE MALE REPRODUCTIVE TRACT**

The CFTR mutation can result in several anatomical changes in men with CF.

- o **Vas Deferens:** The most widely known anatomical abnormality in men with CF is Congenital Bilateral Absence of the Vas Deferens (CBAVD). As discussed above, the Vas Deferens transports mature semen from the testicles to the urethra; however, without an intact Vas Deferens, the semen does not have a way to travel out of the body, resulting in an obstructive azoospermia (lack of sperm in ejaculate).<sup>1</sup>
- o **Epididymis:** There is a large amount of CFTR protein located in the head of the epididymis, which is where the majority of secretions are made. It is thought that epididymal CFTR dysfunction results in abnormal secretions and obstruction, which eventually leads to atrophy and possible absence of the epididymis.<sup>3</sup>
- o **Seminal Vesicles:** CFTR dysfunction in the seminal vesicles is not well understood; however, many men with CF and CBAVD also have atrophy or absence of the seminal vesicles.<sup>1</sup>

**THE BASICS:**

**Fertility in CF Patients**

**OVERVIEW**

As this article has discussed, there are many aspects that determine a healthy reproductive system; however, in order for fertilization of an egg by sperm to occur, it comes down to two important factors:

- o A patent (unobstructed) reproductive tract in both men and women.
- o An intact hormonal cascade allowing the uterus to be prepared for a pregnancy in women.

Breakdown of either of these processes will prevent pregnancy and can contribute to infertility.

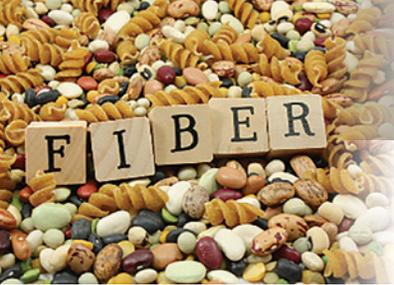
**WOMEN**

While there is well-established data regarding fertility in male CF patients, the data surrounding fertility in female patients is less clear. When applying the two major factors for fertility mentioned above:

- o A patent (unobstructed) reproductive tract: The cervical mucus during ovulation may be thicker than in a non-CF patient, which can make it more difficult for sperm to travel to the uterus and fallopian tube.
- o An intact hormonal cascade: Women with CF are at risk for abnormal menstrual cycles related to malnutrition/malabsorption.

The good news is that despite these potential challenges, there appears to be little evidence that fertility is reduced in healthy women with CF. It was previously reported that women with CF had decreased fertility by approximately 20 percent; however, with improved lifespan

Continued on page 7



# Fiber: An Important Part of a Healthy High Calorie Diet

BY JULIE MATEL, RD

When we think of a healthy diet for individuals with CF, we don't often think of fiber as being an important component. But in fact, it is. Fiber is an important yet under-appreciated nutrient. Here is why upping your fiber consumption should be one of your health priorities this year:

## FIBER FACTS

- Also called "nature's broom," fiber is a carbohydrate found only in whole plant foods, such as fruits, vegetables, nuts, seeds, whole grains, sprouts, and legumes.
- Unlike other types of carbohydrates, fiber cannot be absorbed by your body.
- Fiber helps keep the bowel movements softer and easier to pass, and can add bulk to loose stools.
- Eating enough fiber can promote healthy bacteria in the intestines.

## WAYS TO ADD FIBER TO YOUR DIET

- Read food labels and aim for foods with three or more grams of fiber per serving.
- Choose breakfast cereals with five or more grams of fiber per serving.
- Choose whole grains
  - Whole wheat pasta, ½ c. (5 g fiber)
  - Raisin bran cereal, ½ c. (3 g fiber)
  - 100 percent whole wheat bread, 1 slice (3 g fiber)
  - Whole wheat tortillas, 1 (3 g fiber)
- Eat beans, peas, and lentils
  - Baked beans, ½ c. (8 g fiber)
  - Ham and bean soup, 1 c. (8 g fiber)
  - Chili with beans, 1 c. (8 g fiber)
  - Green peas, ½ c. (4 g fiber)

- Choose these high fiber high calorie snacks
  - Handful of almonds (170 calories, 3 g fiber)
  - Handful of dried apricots (100 calories, 3 g fiber)
  - Peanut butter and banana sandwich on whole wheat bread (420 calories, 7 g fiber)
  - Half-cup of granola cereal and ¼ cup whole milk (340 calories, 5 g fiber)
  - Fig cookies, 2 (2 g fiber)
  - Raw apple with 1 tbsp peanut butter (5 g fiber)



If you have trouble eating whole grain foods all of the time, try these tricks

- Use one slice whole wheat and one slice white bread in a sandwich.
- Top whole fat yogurt or ice cream with nuts and/or raisins.
- Try mixing a high fiber cereal with your favorite cereal.

## HOW MUCH FIBER DO I NEED IN A DAY?

There are no specific fiber recommendations for individuals with CF. The recommended daily fiber intake for persons who don't have CF is:

Age (years)	Total Fiber (grams daily)
1-2	19
4-8	25
9 and older	26-38

## CAN I GET TOO MUCH FIBER?

Yes, adding too much fiber in one day and increasing your fiber intake too quickly can cause gas and stomach pain. Kids can become too full when too much fiber is added to their diet. This may make it difficult for them to eat enough calories. To avoid these difficulties, add fiber into your diet slowly and be sure to drink plenty of fluids.

## Pediatric CF Center Update

BY MARY HELMERS, RN

**MYCHART (Secure Electronic Correspondence):** If you have not signed up already, please sign up for MyChart at your next clinic visit.

MyChart is a secure way to communicate with your provider and CF care team. The CF Care Team cannot respond to patient/parent emails, since general email is not secure. To ensure your security and privacy, our staff will respond to emails with a phone call. We do not always check emails on a daily basis, so if you or your child has a clinical need or question please call the CF nurse line at (650) 736-1359.

It takes only a minute to sign up for MyChart. One of our front desk staff will be happy to assist you.

**ANNUALS:** Remember our goal is to get all annual testing done on or around your child's birthday. Included in the annuals are your lab work, CXR, Bone density scan (12 years of age and older), Full PFT's (starting at age 7), Baseline Audiogram (starting at age 6). Please let us know if you have not had any of these tests done with your annuals.

**WEAR YOUR MASK:** We have new turquoise colored masks that we ask all CF patients to wear. They are available at our front desk. These masks have smaller filters that 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 our front desk staff or anyone from the CF Team.

Lastly, with all the exciting research being done in CF, new drug advances for CF patients are genotype-specific. We need to have copies of all our CF Center patients Genotype/Sweat Chloride test results. If you have a copy or the original result please bring it with you to your next clinic appointment. If your child had these tests done at an outside lab or another CF Center, please contact them and ask them to fax to (650) 497-8791 ATTN: Mary Helmers, RN, Pediatric CF Coordinator. We need these test results for all our patients. Our plan is to have patients re-genotyped if there is no documentation on file.

# Adult Cystic Fibrosis Center Update

BY RONNI WETMORE, RN, MS

As the New Year begins, we will be implementing changes in our program. We are confident these changes will bring about good results, smoother and more effective clinics and patient care.

Our primary goal is to provide you with the best CF care we can, and to maintain all the “gold” standards of care that have been established by the CF Foundation. We are proud of our accreditation status and our ranking amongst one of the top 10 CF centers in the country. In our efforts to maintain our high standards we are reviewing our protocols and reminding you of our commitment to your CF health.

In 2016 we will expect ALL of our patients to establish care with a primary care physician. This is required by many insurance companies as prior authorizations are required for many medications, procedures and therapies. Our physicians are specialists in their field and cannot be considered by insurance companies as primary care physicians.

In an effort to maintain the CF Guidelines established by the CF Foundation, we expect to see each patient a minimum of four times per year. This assures that we are within the national standard of care for our CF Center, which assures adequate monitoring of your health. We plan to schedule your appointment every eight to 10 weeks, to assure this requirement. We know there are times when you may need to reschedule as well as times when we may need to reschedule—working together we can be assured you will be seen regularly for routine clinic visits.

As always, if you are ill, we will do our best to bring you into CF clinic on a Wednesday or Thursday afternoon. However, there may be times when our advice is for you to come to the Stanford Emergency Department for evaluation.

The CF Foundation guidelines also require what we call “Annual Labs,” which are due yearly. We will be ordering these between January and March. The testing includes Complete Blood Count (CBC), Comprehensive Metabolic Panel (CMP), Fructosamine, Hemoglobin A1c, Magnesium, Phosphorous, Gamma-Glutamyl Transferase (GGT), Total IGE, Vitamin Levels A, E and D, and PT/PTT/INR. A two-hour Traditional Oral Glucose Tolerance Test (OGTT) is required annually unless you already have this diagnosis. Please note, you must be NPO (nothing by mouth after midnight) for the OGTT test, which is a 3-hour test during which you must remain seated and may not walk around or run errands between the lab draws. At your clinic visit, we will also collect CF Respiratory and Fungal sputum quarterly, or as needed, as well as and Acid Fast Bacteria (AFB) culture twice annually or as needed.

In an effort to promote better communications and follow through after an admission to Stanford Health Care, nurse coordinators Julian Liang and Ronni Wetmore will visit each of you prior to your discharge. We will schedule your hospital follow-up clinic visit at this time. Also, if you are required to have follow-up labs and/or PFTs, please do so, and call this office with any changes in the discharge plan.

Please call (650)498-6840 to reach Julian and Ronni, Nurse Coordinators. Press Option #1 if your last name begins with A-K, and Option #2 if your last name begins with L-Z. We make every effort to answer your phone calls and messages in a timely manner. If we have called you to follow up on labs, PFT’s, etc., please return our call at your earliest convenience. Both Julian and Ronni see patients in CF Clinic on Wednesday and Thursday afternoons and therefore may not be able to return your call until the following day.

Our primary goal is to provide you with the best CF care we can, and to maintain all the “gold” standards of care that have been established by the CF Foundation.

Both Julian and Ronni meet with our nurse practitioners daily to consult regarding your phone messages and calls. We also have a weekly meeting with our entire team to discuss our practice, our clinics and our protocols. We work together to bring you the best and most comprehensive CF care and maintain our high standards of care. Please do not hesitate to contact us with your concerns or suggestions as we all work together in this effort.

Our entire team looks forward to working together with you in 2016 and making this our best year yet.

## Reproductive Health continued from page 5

and an increasing number of women reaching reproductive age, it is becoming more clear that with good nutrition and lung function, fertility rates can match that of the general population.<sup>2</sup>

### MEN

Up to 98 percent of men with CF are infertile, in part due to CBAVD, as well as atrophy or absence of the seminal vesicles and epididymis.<sup>3</sup> Although most men with CF do not have an intact Vas Deferens, sperm production can remain intact in patients with CF. This is an important point when discussing assistive reproductive technology (to be discussed in Part III).

It is also important to note that fertility and the risk for sexually transmitted infections (STI) are two separate entities in both men and women. Although a patient with CF may be at higher risk for infertility, this does not reduce the risk of obtaining or transmitting a STI. Women and men should continue to practice safe sexual practices to protect themselves from STIs.

## THE BASICS:

### Conclusion

Education and awareness regarding reproductive anatomy, hormonal cycles and the impact CF may have on your fertility are an essential foundation for reproductive health. This article is meant to serve as a general overview of fertility in CF patients as well as a starting point for conversations with your health care team. Please stay tuned for the next articles in this series, which will delve further into reproductive health in CF patients.

<sup>1</sup> Blau, H, Freud E, Mussaffi, H et al. Urogenital abnormalities in male children with cystic fibrosis. *Arch Dis Child.* 2002;87: 135-138

<sup>2</sup> Edenborough, F.P, Women with cystic fibrosis and their potential for reproduction. *Thorax.* 2001;56:649-655

<sup>3</sup> Lewis-Jones, D.I., Gazvani, M.R and Mountford, R. Cystic fibrosis in infertility: screening before assisted reproduction. *Oxford Journals.* 2000;15: 2415-2417

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## Current Research Studies

### CURRENT STUDIES:

**Vertex 661-110** – Open label extension study for subjects who participated in the Vertex 661-103 and 661-108 study protocols.

**Vertex 809-110** – Open label extension study for subjects who participated in the Vertex 809 Part B and the 809-109 study protocols.

**OPTIMIZE** – For treatment of newly acquired Pseudomonas

**PTC study** – Enrolling now for Stop mutations

**Lung CT study** – Enrolling now, young children

**ProQR study** – Enrolling now for F508/F508 adult patients

**Prospect** – Enrolling now, Observational study for healthy normal people and CF patients

**SHIP** – Use of Hypertonic saline in children; enrolling children now.

**Nivalis** – A novel therapy to be taken in combination with Orkambi for adult F508/F508 patients. Enrolling now.

**Vertex 661-108 – 661** in combination with Ivacaftor – Enrollment closed (for limited number of genotypes)

**Vertex 809-109** – Lung clearance index in children 6-12 years with the F508/F508 mutations. (Blinded study with Vertex 809 and 770 combination therapy).

### UPCOMING STUDIES:

**Celtaxsys** – Anti-inflammatory study in adults

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