University of Wisconsin
Clinical Sciences Center

Center Focus

Newsletter of the Pediatric and Adult Cystic Fibrosis Centers

Winter, 2018

How Often Should Patients Be Seen in Clinic?

Michael Rock, M.D.

The first time that I attended the CF Conference was in 1985 when I was finishing my second year of pediatric residency. (At that time, the conference was called "Cystic Fibrosis Club"– a gathering of physicians, basic scientists and other health professionals who had an interest in CF. The attendance at that 1985 meeting was approximately 200 people. Compare that to the almost 5,000 people in attendance at the 2017 North American Cystic Fibrosis Conference in Indianapolis.) There was a very interesting presentation at the 1985 conference by Dr. Robert Wood, a physician who cared for CF patients at that time at the University of North Carolina in Chapel Hill. Dr. Wood examined the median survival of patients from three different CF Centers. (Median survival is the point where 50% of patients pass away prior to a specific age, and 50% of patients live beyond that specific age.) The three centers were similar in the number of patients, size of the metropolitan area and continuity of the center director. Characteristics of the patient population at the three centers were also similar (age at diagnosis and age at first referral to the center). The results of respiratory cultures, hospitalization patterns, number of days in the hospital and treatment (chest physiotherapy and antibiotics) were similar amongst the three centers.

Center A had a median survival of 8 years; center B had a median survival of 16 years; and center C had a median survival of 24 years. (As an aside, thank goodness that the health and survival of patients with CF has markedly improved since 1985.) What accounted for the difference in survival of patients? The number of outpatient clinic visits varied markedly between the three centers. Center C, which had the best survival, saw patients in clinic more frequently than the other two centers.

The standard of care as set by the CF Foundation is that for everyone above 1 year of age, patients should be seen in clinic a minimum of every 3 months. For infants, the standard of care is to be seen monthly in the first 6 months of life, and every other month between 6 months and 1 year of age. These frequencies of visits are based on the patient doing well with no growth or pulmonary problems. If a patient is not at their best baseline, then they should be seen more often than these guidelines.

If you look at pages 6 and 7 of our winter 2017 CF Center newsletter, you will see tables of the specific tests that are obtained. *https://www.uwhealthkids.org/files/kids/docs/cysticfibrosis/Center_Focus_Winter_2017.pdf*

Today compared to 1985, we have so many more tools to use to keep patients healthy (some of which are: early diagnosis by newborn screening, better enzymes, improved oral antibiotics and the use of inhaled antibiotics if necessary, emphasis of the importance of airway clearance and a menu of choices to achieve airway clearance, hypertonic saline, DNase, Kalydeco®, Orkambi®, improved attention to infection control, etc.). By working together via seeing patients frequently and intervening early before problems become more advanced, we can make a difference with improved outcomes.

Another "…caftor" is coming

A few basics about classes of CF mutations before moving on to the protein modulator therapies. CF mutations have different characteristics and have been placed into five (or six) different classes. Class 1 mutations are those that produce no CFTR protein. Many of these mutations have an "X" at the end of the name which signifies that the production of protein stops prematurely. Class 2 mutations are folding/trafficking defects in which very little protein reaches the cell surface. The most frequent mutation, F508del, is a class 2 mutation. Class 3 mutations are also known as gating defects. These mutations result in abnormal CFTR protein reaching the membrane of the cell and the chloride channel does not work properly. The G551D mutation is a class 3 mutation. Class 4 mutations are known as having decreased conductance. The CFTR protein is at the cell surface, but there is decreased movement of chloride (a slang term for this is a narrowed channel).

Class 5 mutations result in decreased production of the CFTR protein and thus decreased amount of protein at the cell surface. Some splicing mutations such as 3849 + 10kb C>T and 3272-26A>G are class 5 mutations.

Ivacaftor is the generic name for Kalydeco®. This was the first protein modulator therapy approved for CF in 2012. This was initially studied in patients with the G551D mutation, a class 3 gating defect. The results of that study were published in the New England Journal of Medicine on November 3, 2011 (volume 365, pages 1663-1672). There was a significant improvement in the FEV1 and significant decrease in the sweat chloride value. Initially, this was approved for patients with the G551D mutation who were 12 years of age and older. The FDA later changed the approval down to 2 years of age. (Currently, we are a study site for the use of ivacaftor in infants who are under 2 years of age.) Additionally, the FDA has approved a number of other mutations that are responsive to ivacaftor. You can go to *www.kalydeco.com* to see which mutations have FDA approval for the use of Kalydeco®.

Approximately 50% of people with CF have two copies of the F508del mutation. This mutation does not respond to ivacaftor alone. Why? Ivacaftor activates the chloride channel. In people with two copies of the F508del mutation, there are very few CFTR chloride channels in the membrane of the cell. A second drug is needed to traffic the CFTR protein to the cell surface, and then the protein can be activated with ivacaftor. Lumacaftor is a drug that traffics the CFTR protein to the surface of the cell. The combination drug lumacaftor/ivacaftor (Orkambi®) was studied in people with CF who have two copies of the F508del mutation. The results of the study were published in the July 16, 2015 issue of the New England Journal of Medicine (volume 373, pages 220-231). The patients who received Orkambi® had significantly improved pulmonary function values (although not as impressive as the improvement in G551D patients who received Kalydeco®) compared to those who receive placebo. The FDA approved Orkambi® for patients above 12 years of age who have two F508del mutations. Subsequently, the FDA has decreased the age of approval down to 6 years of age.

There are some potential drawbacks to lumacaftor: 1) Ivacaftor and lumacaftor can interact with each other to cause a decrease in each other's drug level. 2) For patients with more advanced lung disease, when first starting lumacaftor, there can be a sensation of chest tightness. 3) Lumacaftor activates an enzyme called cytochrome P-450-3A. This can cause issues with certain foods (such as Seville oranges) and there are drug interactions (such as decreased effectiveness of oral contraceptives in people who are receiving Orkambi®).

There is a new "caftor" that has been studied and the results were published in the November 23, 2017 issue of the New England Journal of Medicine. Tezacaftor/ivacaftor was studied in two different populations (and thus, there are two different articles in this issue of the New England Journal of Medicine). Tezacaftor/ivacaftor was studied in people with CF who have two copies of the F508del mutation and was shown to significantly improve pulmonary function and the quality of life score (New England Journal of Medicine 2017, volume 377, pages 2013-2023). The second publication in the same issue of the New England Journal was the use of the tezacaftor/ivacaftor product in patients who had one F508 mutation and a second mutation that is predicted to have residual CFTR chloride channel function. (There were 27 mutations on this list; 25 of these mutations have already been approved by the FDA for Kalydeco®.) The patients who received tezacaftor/ivacaftor had a 6.8% improvement in FEV1 compared to a 4.7% improvement in FEV1 for patients who received ivacaftor alone.

The advantage of tezacaftor is that it does not have any of the above issues of lumacaftor… no drug-drug interaction between tezacaftor and ivacator, no chest tightness and no activation of cytochrome P-450-3A.

Vertex has applied to the FDA for approval of tezacaftor/ivacaftor. The FDA must submit a response by February 28, 2018. Stay tuned… we may have a new modulator therapy to prescribe in 2018.

Trailblazer

Do you want to be a trailblazer? Go to *cff.org/trailblazer* to watch a video and click on other links that discuss how you and the care center can partner together to bring new therapies to the market. Working together, we look forward to the day when CF is no longer cystic fibrosis; instead, CF will mean Cure Found.

Update from the 2017 North American Cystic Fibrosis Conference

Almost 5,000 care providers and researchers gathered in Indianapolis for the 2017 North American Cystic Fibrosis Conference. The highlight of the conference were the three plenary sessions:

- Plenary 1: Matching Medicines with Mutations
- Plenary 2: Lung transplantation: Challenges & Opportunities for Advanced CF Lung Disease
- Plenary 3: Partnering With the Community

You can watch these plenary sessions by going to: *https://www.cff.org/Research/Researcher-Resources/North-American-CF-Conference/2017-North-American-CF-Conference/*

All three of these sessions were important, but we will highlight the information from the first plenary. In this plenary session, it was stated that 2017 will be known as the breakthrough year for modulator therapies. There are several reasons for this. In 2017, the FDA approved additional mutations that are responsive to ivacaftor. The significance of this is that some of these additional mutations are rare and do not occur in a large number of people. Thus, it is not practical to conduct clinical trials with so few available subjects. The FDA approved these additional mutations based on the response of these medications to ivacaftor in the laboratory. This is a huge shift for the FDA and opens the door to approval of future medications by this method.

Another reason for 2017 being a breakthrough year for modulator therapy is the publication of the results of tezacaftor/ivacaftor (see "Another '…caftor' is coming" article in this issue of Center Focus). Tezacaftor/ivacaftor is an improved version of lumacaftor/ivacaftor and it is hoped that there will be FDA approval in 2018.

Lastly, in 2017, the results of phase II trials of triple combination therapy were released by Vertex. There are four "next generation" correctors which are being studied as part of triple combination therapy with tezacaftor/ivacaftor: VX-440, VX-152, VX-659 and VX-445. These four next generation correctors, in combination with tezacaftor/ivacaftor, have been shown to result in improved pulmonary function and decreased sweat chloride values in patients with two F508del mutations and patients with an F508del mutation and a minimal function mutation. Vertex is continuing to collect data from phase II trials and in 2018, they will start a larger phase III trial of one or more of these triple combination regimens (and we will be one of the study sites). You can read the results of the phase II trials at: *http://investors.vrtx.com/releasedetail.cfm?ReleaseID=1033559*

Although Vertex has taken the lead on small molecule drugs that target defective CFTR, there are other companies that are developing drugs to improve CFTR chloride channel activity.

Pediatric Parent Advisory Group

The Pediatric Parent Advisory Group will have its first meeting in January 2018. After sending out invitations to you all as well as our invitation at CF Education day, we have heard back from several interested families and are excited to begin our work together. We will keep you posted on our projects and progress!

Keeping Healthy during Cold and Flu Season

It is that time of year where cold and flu viruses begin to peak in our area. There are many ways to ensure that you and your family stay healthy this winter season.

Get vaccinated: Make sure to stay up to date with all immunizations.

Good hand hygiene: It is important to clean your hands often, either with soap and water or by utilizing hand sanitizer, especially after coughing or sneezing. Doing so will help reduce the spread of illness to you and others around you!

Reduce the spread: Make sure you are covering your nose and mouth with a tissue when coughing or sneezing. Tissue should be thrown away in a waste basket. It is important to clean hands afterwards to help prevent the spread of germs to others.

Hands off: Avoid rubbing or touching eyes, nose, and mouth. Germs are easily spread from surfaces to you through the eyes, nose, and mouth.

Give yourself some space: It is important to avoid close contact with others who are sick. Keeping an arm's length away will help reduce the spread of illness. If you are sick, try to keep your distance from other to protect them from illness.

Stay home: The best way to prevent the spread of illness is to stay home when you are sick!

Take care of yourself: By taking care of yourself you will help protect your body against illness. It is important to maintain good hydration, eat healthy, nutritious foods, and get good quality sleep. Doing so will help your body defend itself against germs and infection.

Wishing you a healthy winter season!

Social Work Changes and Welcome

Some news and changes in the social work coverage have started as of December 1, 2017. Tristan Verbeten has joined the pediatric pulmonary team. Tristan trained here in our CF Center and has been back here at American Family Children's Hospital since December of 2015. She will primarily cover the general pulmonary population and Craig will focus more on the CF population. With the changes and increase in coverage, we will begin the depression screening which has been recommended by the CF Foundation.

Infection Control Safety Tips for at Home and in your Environment from the CFF Infection Control Guidelines

Below is a list of some of the safety tips that you may or may not know in regards to infection control:

1. All people with CF who do not live in the same household should avoid activities and risk factors that are associated with transmission of CF pathogens such as:
   a. Social contact between people with CF
   b. Physical contact between people with CF (handshakes/kissing/intimate contact)
   c. Car rides with another person with CF
   d. Sharing hotel rooms with another person with CF
   e. Fitness class with another person with CF
2. All people with CF, including those who live in the same household, should avoid the following:
   a. Sharing personal items (toothbrush, drinking utensils)
   b. Sharing respiratory therapy equipment
3. Tap water or well water that meets local public health standards, distilled water or bottled water may be use by people with CF for drinking, bathing and cleaning nebulizers and other respiratory equipment (airway clearance devices, spacers and netipots) if followed by disinfection, and for the water needed for heat disinfection (boiling, microwaving, and steam sterilizing). Only sterile water should be used for nasal rinses (netipots) filling of humidifier reservoirs, and as a final rinse of respiratory equipment (after cold disinfection)
4. All people with CF and their family members/close contacts receive recommended vaccines at the recommended schedule/dose/age/route of administration unless there is a medical contraindication
5. Use of antiviral medications (Tamiflu) for prevention or treatment of influenza according to ACIP recommendations
6. People with CF who live in the same household perform airway clearance with only 1 person with CF in the room during treatment
7. People with CF attending the same daycare and/or school should not be in the same room at the same time unless they live in the same household. Education should be provided to the staff so they can work with people with CF and/or parents to develop strategies to minimize contact between people with CF (assignment to separate classrooms and separation during other schedule common activities including lunch physical education and recess.)
8. Only 1 person with CF attend CF Foundation/Center sponsored indoor events, unless they live in the same household, to reduce the risk of person-to-person transmission of CF pathogens
9. People with CF can attend CF Foundation/Center sponsored outdoor events providing they maintain a distance of at least 6 feet from others with CF
10. People with CF should avoid direct contact with people with skin and soft-tissue infections caused by MRSA unless wounds are covered, hand hygiene is performed frequently, personal items (towels) are not shared, sports equipment is cleaned between use, and cleaning protocols for environmental surfaces are established to reduce the risk of MRSA transmission
11. People with CF and respiratory cultures positive for MRSA should not be restricted from contact with people without CF in congregate settings (sports teams, classrooms and the workplace) if the person with CF performs appropriate hand and respiratory hygiene.
12. People with CF should limit prolonged and/or repeated exposure to activities that generate dust from soil and organic matter (gardening and lawn mowing) to decrease exposure to potential soil borne pathogens (Burkholderia and Aspergillus.)
13. People with CF should avoid exposure to construction and renovation activities that generate dust to decrease exposure to potential pathogens (Aspergillus.)
14. People with CF can swim in pools or water parks with adequate disinfection (chlorination.)
15. People with CF should avoid activities in hot tubs, whirlpool spas, and stagnant water.
16. People with CF should perform hand hygiene after changing the litter, handling feces, cleaning and disinfecting the cages or fish tanks of their pets, or interacting with farm animals.
17. People with CF should avoid cleaning stalls, pens, or coops
18. Healthcare Personnel with CF should not provide care for other people with CF
19. People with CF interested in a career in healthcare should receive counseling from their CF Care team regarding specialty areas wherein job duties minimize the risk of transmission or acquisition of potential pathogens

Rebecca Hays, Mental Health Coordinator

My name is Rebecca Hays, and I am the new Mental Health Coordinator for the adult CF clinic.

I come to the adult CF clinic from the UW transplant program, where I was a social worker for 15 years. While there, I enjoyed helping people fit their treatments into the rest of a good quality life, and supporting programs that helped patients learn from one another. I helped start a patient mentoring program, built education materials in partnership with patients, and supported a patient/ family advisory council to increase the strength of patient voice.

I'm looking forward to helping build a mental health and quality of life program for the adult CF clinic. This will include designing an individualized care plan with each of you, and sharing the latest mental health and quality of life science findings. I hope to learn from you about your goals, & explore approaches and tools to help you feel your best.

I have been learning so much by shadowing the experienced CF team, as of December 2017, and look forward to meeting you! You can read more information about the mental health initiative elsewhere in this newsletter.

See you soon!

Rebecca Hays, MSW

A few snippets about me:

- I love the water: kayaking, hiking to waterfalls, stand-up paddle-boarding, and reading at the beach. In the winter, I skate (poorly!) on the lake.
- Before I became a social worker, I was a musician.
- I like to try new foods, the spicier the better.

Tristan Verbeten

Hello everyone! I am so happy to be back with the pulmonary team at the American Family Children's Hospital! A few of you may recognize me as I completed my social work traineeship with the Pediatric Pulmonary Center in 2012-2013. I am excited to see some familiar faces and meet new patients and families!

I attended the wonderful UW-Madison for both my undergraduate and graduate education (*Go Badgers!*). I believe they say that makes me a "Double Badger." I have a Bachelor of Science degree in Psychology, a Masters of Social Work degree and am a Certified Advanced Practice Social Worker in the state of Wisconsin. After graduating I worked with adults for a few years before coming back to AFCH in 2015 to continue my passion – serving children and families. I worked in various pediatric speciality clinics prior to accepting the position with the pulmonary team!

Hope to see many of you soon!

Andrew Braun, MD

Dr. Andrew Braun is the new adult CF director, and a native of Wisconsin. He attended the University of Wisconsin-Madison for both his undergraduate and medical school training. He subsequently completed internal medicine residency at the University of California–San Francisco, and his clinical and research fellowship in pulmonary, critical care, and sleep medicine at the Johns Hopkins Hospital in Baltimore, Maryland. He has a Master of Health Science degree in clinical investigation from the Johns Hopkins Bloomberg School of Public Health.

Dr. Braun has been working with the CF community since medical school and his initial research mentor was Dr. Phil Farrell. Dr. Braun is currently funded by the CF Foundation to continue research investigating sleep disorders in patients with CF. He has specific clinical interests in advanced CF lung disease, metabolic complications of CF, and lung transplantation.

Tracy Vaughan, Social Worker

Hello! I'm so glad to have this opportunity to introduce myself to you! Many of you have already met me, but for those that haven't, my name is Tracy Vaughan. I am the new(ish) social worker at the UWHC adult CF clinic. I'm so thrilled to be a part of this team and to have the opportunity to work with such an amazing group of both colleagues and patients! I joined the team in February 2017 and have learned so much from you already; I'd like to express my sincere thanks for taking the time to meet with me during our clinic days!

Just a little bit about me, so we can continue to get to know each other better: My social work practice has always been centered around healthcare. I've chosen this path because I feel that the healthcare system can be complex, expensive, and confusing. This, in combination with the stress of living with a healthcare condition, can be overwhelming for so many. I feel passionate about helping patients with their individual needs and connecting patients and families with resources that will lessen burdens and worries and increase joyful days. I often help patients and families with questions and resources including, but not limited to, insurance, finding affordable medications, transportation, as well as financial and disability related concerns.

While I'm not at work I enjoy traveling with my husband, walking my goofy dog, reading, and biking. My favorite book of all time is The Art of Racing in the Rain. I love the ocean, sunshine, and gardening.

I'm looking forward to our continued work together!

To Our Patients and Families:

As we continue to strive to provide the best possible care for people with CF, we will be implementing new recommendations offered by the CF Foundation to offer all patients screening for anxiety and depression once per year, with follow-up available as indicated.

Research shows that people with CF (kids and adults) sometimes experience anxiety and depression. People experiencing anxiety and/or depression may have a harder time enjoying quality of life, and getting treatments done. Their health tends to be poorer, and they are more likely to struggle in work, school, and relationships. Though we all get worried or sad at times, anxiety and depression that doesn't go away can interfere with daily activities and worsen overall quality of life.

What does this mean for you?

- At an upcoming clinic visit, we will invite you to complete two screening tools, the PHQ-9 (Patient Health Questionnaire) and the GAD-7 (Generalized Anxiety Disorder screen). The questionnaires have 9 and 7 questions, and should take no more than 3-5 minutes to complete.
- In the adult clinic, our new mental health coordinator, Rebecca Hays, will share the tools and results with you. If there are concerns based on the results, or if you have questions, she will work with you to find the best solution to address these concerns.
- In the peds clinic, we will be updating you soon with ideas about how to implement the new guidelines.
- These questionnaires are optional, but we hope that by identifying symptoms of anxiety and depression early through screening, and then providing supportive education and connections to helpful services, we can contribute to your health and well-being.

We are glad to answer questions you may have about these new guidelines, and are eager for your thoughts on how to make this care meaningful. Please do not hesitate to contact us, as we value your input. For those of you in the adult clinic, you may contact the mental health coordinator, Rebecca Hays (608-263-8214) for more information, or discuss this with the team in clinic. For those in the peds clinic, please call the team at 608-263-6420 for more information.

We look forward to seeing you at your next visit!

Sincerely,

Your CF Team

Quittner et al. Thorax. 2014: 69(12): 1090-7

Upcoming Changes for Annual Labs/Chest X-Ray/Oral Glucose Tolerance Test in the Pediatric CF Center

Starting in 2018 we will be requesting all annual labs, chest x-rays and oral glucose tolerance tests to be performed in the first quarter of the year (January –March). The reason behind this change is to hopefully improve our success with obtaining all required testing so we can report the required data to the Cystic Fibrosis Foundation. This is part of the nationwide registry data collection which is something each CF Center is graded on. Obtaining all required testing in a timely manner allows us to work towards our goal of being a top 10 performing center. When testing is missed on any patient at our CF Center not only is our center penalized making it more difficult to reach our goal of being a top 10 performing center, but it more importantly does not provide the best care possible.

If your child is 10 years of age or greater, an Oral Glucose Tolerance Test is required. We will be calling you prior to the clinic visit in which this test is ordered to remind your child to come fasting to clinic (nothing to eat or drink 8 hours before the test) in hopes of improving success. We will also be handing out educational information on obtaining an Oral Glucose Tolerance Test at the clinic visit prior to when this test is ordered.

On the next page is an example of what the education information looks like. (*Health Facts for You*). If your insurance mandates obtaining labs and chest x-rays locally we will be providing you printed orders to take to your local lab. If we have not received the lab/x-ray report from your local facility, we will call you to remind you that tests are still needed. All the above efforts are meant to improve success in obtaining the necessary tests and most importantly optimize your child's care.

By obtaining annual labs in the first quarter of the year, this will also provide us with a better picture of your child's Vitamin D level. One's vitamin D level is at its lowest during the winter months. Obtaining the level at this time will allow us to recommend the best dose of vitamin D supplementation.

UW Health

Health Facts *for you*

Screening for Cystic Fibrosis Related Diabetes (CFRD) using the Oral Glucose Tolerance Test

What is cystic fibrosis related diabetes (CFRD)?

CFRD occurs when people with cystic fibrosis (CF) have a unique type of diabetes. CFRD is not the same as diabetes found in people who do not have CF. CFRD is a mix of resisting insulin and not having enough of it in the body. Therefore, the finding and treating of CFRD is not quite like that of other types of diabetes.

What are the common symptoms of CFRD?

- Weight loss or trouble maintaining weight
- Unexplained decrease in lung function
- Excess fatigue
- Frequent urination
- Frequent drinking

How common is CFRD?

CFRD is common in people with CF, even more so as they get older. It occurs in about 20% of adolescents and 40-50% of adults.

How is CFRD diagnosed?

First, a blood glucose level will be drawn. This is a blood test that is done without paying attention to the time of day or to when a meal was last eaten. If this level is less than 126 mg/dl, it is not likely that a person has diabetes. The CF clinical care guidelines suggest this level should be checked yearly.

Second, an oral glucose tolerance test (OGTT) will be performed every year starting at the age of 10 years. The test should be performed during a stable period of health, at least 6 weeks from a recent respiratory infection. The OGTT may also be performed on people younger than 10 years if the health care provider suspects CFRD. A person must have nothing to eat or drink for at least 8 hours before this test is done. Water is permitted.

To do an OGTT, blood is drawn to measure the "baseline" glucose level. Then the person will be asked to drink a 10 ounce flavored drink (Glucola) over 5-10 minutes. The blood sugar will be measured again two hours later. Any person who has a blood sugar level greater than 200mg/dL at two hours or a blood sugar drawn at baseline greater than 126 mg/dL, has a positive test and will require a repeat test at least six weeks apart from the first. If both of these tests are positive, a diagnosis of CFRD is made. Insulin treatment should be prescribed.

When and where should screening be done for CFRD?

Screening for CFRD should occur every year starting at the age of 10 years. This test may be performed as part of your routine clinic visit at your CF Center, or if you prefer, the test may be performed closer to your home.

If you have any questions or concerns, call the CF Center at **608-263-6420**

The toll free number is **1-800-323-8942**. Ask for the CF Center.

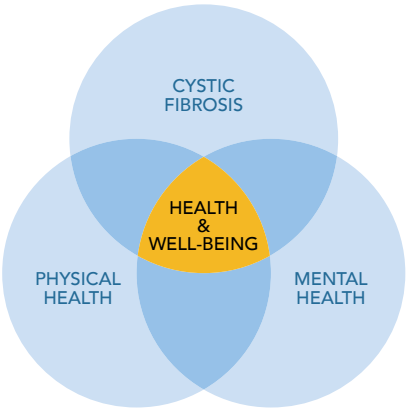
DEPRESSION, ANXIETY *and* CYSTIC FIBROSIS

WHAT THE GUIDELINES MEAN FOR YOU

People with cystic fibrosis and parents who take care of children with CF are two to three times more likely to experience depression, anxiety or both, compared to people in the general population.*

Guidelines were published to help CF care teams provide effective care for people with CF and their families with depression, anxiety or both.

The following recommendations are intended to help you understand that although moments of sadness and anxiety may come and go, **depression and persistent anxiety should be treated as part of your overall health and emotional wellness.**



WHAT IS DEPRESSION?

Depression is a common medical illness that negatively affects the way you feel, think and act. Unlike occasional sadness or feeling blue, clinical depression can last for a long time—weeks, months or years—if not treated. People who have depression can have extended periods where they feel hopeless and lose interest in things they normally would enjoy.

WHAT IS ANXIETY?

Anxiety is a normal emotion that comes and goes in response to fears or worries about changes in health, work, relationships or money. A person may have an anxiety disorder if the anxiety does not go away, gets worse over time and prevents them from participating in ordinary daily activities.

In addition to the generalized anxiety many people with CF and their family caregivers may experience, some people also experience a very specific form of anxiety centered on medical procedures.

WHEN LEFT UNTREATED:

Untreated depression, anxiety or both can affect both your physical and emotional health and interfere with your ability to take care of yourself or your child.

People with untreated depression, anxiety or both:

- Are less likely to manage their treatment plans effectively.
- Tend to have worse lung function.
- Have a lower body mass index (BMI).
- Experience more hospitalizations.
- Often have higher health care costs.
- Experience a lower quality of life.

WHAT YOU CAN DO IF YOU THINK YOU ARE DEPRESSED OR HAVE ANXIETY:

While feelings of depression and anxiety can be a normal response to living with CF, there are things that you can do to prevent, get help for and reduce the risk of either depression or anxiety from returning.

The Cystic Fibrosis International Guidelines Committee (sponsored by the Cystic Fibrosis Foundation and the European Cystic Fibrosis Society) recommends the following for screening and treating depression and anxiety as part of comprehensive CF care:

- **Learn New Coping Skills:** Your CF care team will work with you on effective ways to manage stress and provide ongoing age-appropriate information on how to cope with a chronic disease like CF.
- **Get Screened:** If you have CF and are at least 12 years old, or if you are caring for someone with CF who is age 17 or younger, you will be asked to complete two short screening surveys.
- **Get Help:** If the survey results suggest you are struggling with depression, anxiety or both, your CF care team will recommend further evaluation and may recommend treatment.

*Quittner AL, Goldbeck L, Abbott J, Duff A, Lambrecht P, Solé A, Tiboshc MM, Brucefors AB, Yüksel H, Catastini P, Blackwell L, Barker D. *Prevalence of depression and anxiety in patients with cystic fibrosis and parent caregivers: results of The International Depression Epidemiological Study across nine countries*. Thorax. 2014;69:1090–1097. doi:10.1136/thoraxjnl-2014-205983.

WHAT YOU CAN DO TO STAY WELL:

If you have mild symptoms or recognize the beginning signs of depression, anxiety or both, talk to your CF care team. They can work with you to maintain your emotional health.

The following are ways that you can prevent problems with depression or anxiety from getting worse and limit their impact on your life or that of your loved ones.

- Talk with somebody, preferably in person. Many people with depression withdraw and isolate themselves from other people.
- Spend time with people who lift your spirits.
- Avoid alcohol or drugs.
- Continue your CF treatment plan.
- Practice good sleep habits. Do your best to get enough sleep. Go to bed and wake up on a consistent schedule. Avoid staying in bed when you are not sleeping.
- Get outside in nature for 30 minutes every day.
- Make time for things you enjoy.
- Be physically active. Exercise can help reduce stress.
- Practice relaxation techniques.
- Avoid caffeine and cigarettes, which can increase anxiety levels.
- Join a support group. Talking about your problems with people who have the same experience can help you feel less alone.

These activities are not a substitute for professional care, but can make a real difference in your mood.

WHAT ABOUT INSURANCE COVERAGE?

The Affordable Care Act expanded coverage for conditions like depression and anxiety, and most insurance providers cover preventive services, such as depression and anxiety screening. Most large insurance plans cover mental health screenings for adults and children, age 12 and older.

All state Medicaid programs must provide some mental health coverage for treatments, while Medicare covers a range of mental health services. Medicare Part A provides coverage for inpatient care, and Medicare Part B covers outpatient care, such as diagnosis and treatment.

Insurance providers often have different policies regarding coverage for treatment of mental health issues like depression and anxiety. Some cover a limited number of talk therapy visits. Some cover only medication. Check your insurance enrollment materials or call your insurance provider for information about treatment for your depression or anxiety.

The CF Foundation can help people with CF and their families understand their insurance coverage options and connect them to the right resources.

Call **844-COMPASS** (844-266-7277) or email **compass@cff.org**.

TO LEARN MORE

View the CF Foundation and ECFS Guidelines at **cff.org/Care-Guidelines/Depression-Anxiety**

View the CF Foundation's website on emotional wellness at **cff.org/Living-with-CF/Emotional-Wellness**

If you are considering suicide, or if someone you know may be planning to harm him or herself, immediately call **911** or **800-273-8255** (National Suicide Prevention Lifeline) or visit **suicidepreventionlifeline.org**.

For questions, call **1-800-FIGHT-CF** (800-344-4823) or email **info@cff.org**.

CYSTIC FIBROSIS FOUNDATION®
ADDING TOMORROWS™

When you come to our CF clinic, we take special precautions to make sure your child stays safe and avoids exposure to illness.

We have created a CF passport that can be used when you bring your child to the local pediatrician, urgent care, emergency room or dentist.

It is important to remember that no matter how much we want to keep our kids in a bubble to protect them, they are kids first. The idea of the passport is to minimize exposure in high risk situations.

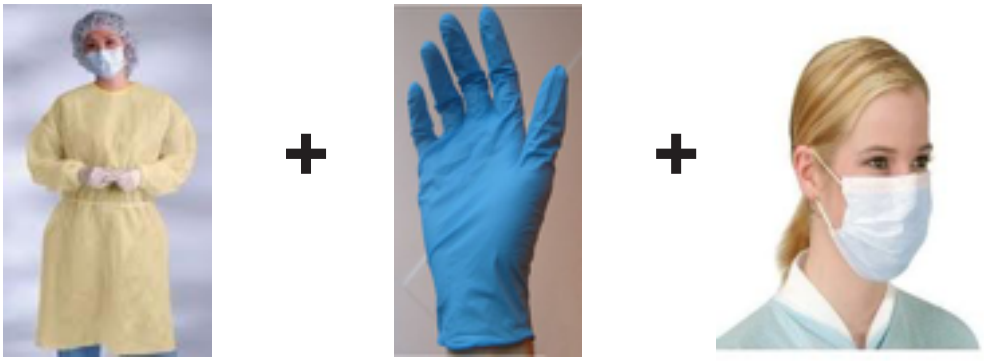
CF PASSPORT

- Please remind me to wear a mask throughout the clinic or hospital.
- Please make sure the room surfaces have been thoroughly cleaned with Cavi Wipes before I am placed in a clinic room.
- Please place me into a room as soon as possible to minimize my exposure.
- Please follow our CF care team recommendations to gown, glove and mask.
- Please thoroughly clean your stethoscope with a Cavi Wipe before and after using.
- Please remember to use good handwashing/cleanser before and after contact with me.

American Family Children's Hospital CF Center

If you have questions, please contact my CF care team:

American Family Children's Hospital and Clinics CF Center at 608.263.6420



Thanks for helping me stay healthy!

Cystic Fibrosis Foundation Pharmacist Award

A belated congratulations to Catherine Decker, our adult CF pharmacist, for receiving a "Clinical Pharmacist Award" from the CF Foundation in early 2017. This is a prestigious and competitive award that she is honored to have received. Catherine has prepared a summary of the goals of this award:

"There are two main goals we are hoping to achieve with this award:

First, we are identifying opportunities for improving the medical knowledge of our adult CF patients with regards to their CF care. Many of our patients (70 so far!) have taken a survey to assess their knowledge of medical issues of which many adult CF patients experience. We were happy to learn (and not surprised) that many of our patients have a solid baseline of core knowledge with regards to best CF care. However, specific knowledge gaps were identified that we can target for educational interventions throughout the year. For example, some patients were not aware of commonly used medical acronyms such as BMI, CFRD, and DIOS (if you don't know the meaning of these acronyms – see definitions below). Additionally, some patients were not aware of the therapeutic goal of some of our frequently prescribed medications such as Pulmozyme, or Hypersal. This information is helping us to target ways that our care team can address and "close knowledge gaps" so that both patients and care providers can fully engage in medical and pharmaceutical care.

Second, we are looking to provide guidance to the national community of CF care centers, what is the ideal role and scope of practice for a pharmacist who has expertise and specialization in the care of patients with CF. Some of the roles that the pharmacist can provide in the clinic include: preparing "CF Care Plans"; reviewing and discussing airway clearance therapies; information on new medications additions, review of medication regimens; vaccination recommendations; and ensuring the optimal laboratory monitoring for potential side effects. The CF Foundation does not yet require that all centers have a clinical pharmacist, but we hope that the expertise from our center will help to provide national guidelines for the critical role that once can add to a CF care center."

Acronym definitions:

BMI: body mass index

CFRD: CF-related diabetes

DIOS: distal intestinal obstruction syndrome (a severe form of constipation)

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