August 2016

The CF Breeze

"CF wisdom & inspiration for CF families"

Emotional Wellness – Mental Health Screenings

The Cystic Fibrosis Foundation has implemented new guidelines for the screening and treatment of depression and anxiety in CF patients and families. Many downplay the emotional toll that CF can have, and this is one step towards integrating emotional wellbeing with specialized CF care.

Studies have shown that people with CF and parent caregivers are at increased risk for depression and anxiety, which negatively impacts adherence to CF treatments and overall health, including lower lung function, lower body mass index, and more frequent hospitalizations.

To address these needs, our CF social worker will be administering a depression and anxiety screen for all patients 12 and older at your annual or clinic visit, as well as parent caregivers with children between birth and 17 years. If the screening suggests that a person may have depression or anxiety the social worker will then talk with the patient and family about resources and referral options. Treatment may include an assessment with a mental health professional, talk therapy, medication, or a combination of both.

Please do not wait until your annual visit to talk about your mental health needs. The CF social worker is available throughout the year to discuss any mental health concerns, questions, and provide brief counseling to patients and families.

If you have any questions regarding this new process, or would like to discuss your mental health, contact Tami Vance, LICSW at 612-813-6839.

Upcoming Events

CF Community Parent Support Group meets from 7 p.m. to 8:30 p.m. at Children's Minneapolis. (*A call in number is also available at 612-215-9496 Password 12345.)*

- Monday, Sept 26

For CF Foundation activities, please visit:

www.cff.org/Chapters/minnesota

The CF Breeze will be distributed electronically for patients and families of Children's Hospital and CRCCS.

To be added to the distribution list or to update your e-mail address, please complete a listserv form available in the lobby of CRCCS or by contacting Mary Sachs or Sandy Landvik.

How You Can Help Yourself

In addition to care provided by a mental health specialist, you can do the following things to help yourself manage your mental health:

- 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.
- Make sure you do your CF treatments every day.
- 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 or in nature for 30 minutes each day.
- Make time for things you enjoy.
- Exercise every day.

Although these activities are not a substitute for professional care, they can make a real difference in your mood.

How to Help your Child Get through a Long Day

Suggestions from Child Life Specialists at Children's Hospital

Recently, the CF research team expanded a medical trial to children 2-3 years old. Some of the days were very long with many procedures for children this young. In efforts to make these long days successful for the kids, the team reached out to the Child Life department for support. Child Life Specialists (CLS) Christi Dady and Melissa Haun were excited to partner with the CF research department.

We asked them to share a little bit about their profession and some tips for parents bringing their children in for medical experiences.

What is a Child Life Specialist?

For those who are not familiar with child life services, we are professionals trained in child development who specialize in working with children and siblings in a health care setting. Child Life Specialists build on children's strengths and interests to help them adjust to their medical experience. We work in a variety of areas throughout the hospital including the emergency department, sedation and procedural services, pre-surgery area, medical surgical units and intensive care areas.

Setting Up for Success

While collaborating with the research team we identified a few tips to share that parents can keep in mind to support children of all ages in a clinical trial or clinic visit.

- It's important that your child knows how long the visit will be, what is expected of them and is able to ask questions about their care
- Bring toys to keep them busy during downtime. Items such as a favorite stuffed animal or blanket provide comfort during tests and procedures
- Provide simple explanations about what test is being done and why.
- Build in acceptable choices during the visit. Medical tasks are best made as statements. Example: "It is time to take your blood pressure to see how strong your heart is. Are you going to sit in mom/dad's lap or on the chair all by yourself?" vs. "Do you want to take your blood pressure?"
- Play and distraction are great tools to alleviate pain, calm fears and pass time
- It's normal for your child to be sad, scared or even cry; focus on what he/she did well
- Practice coping and calming skills when outside of the medical setting as well to build these up for future visits

A caregiver's support and guidance teaches children life skills and how to cope with challenging situations. Building resilience will be an asset to them for future medical experiences.

Does CF Impact Puberty?

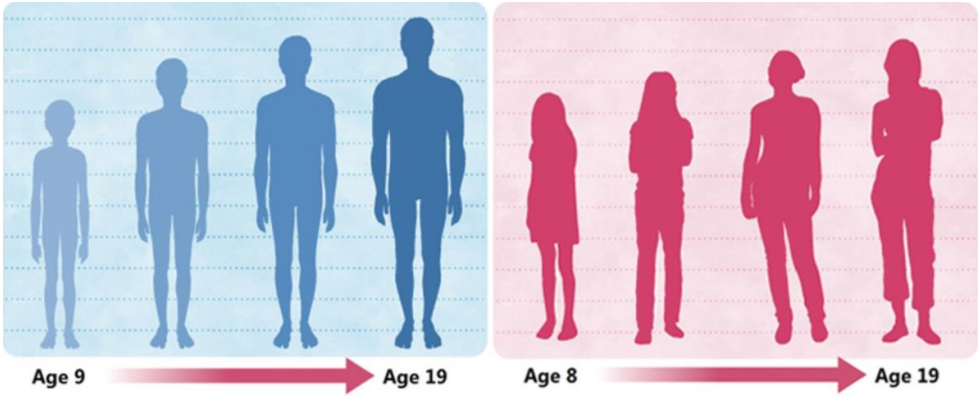
Both young men and women with CF may go through puberty later than many of their peers.

For young men with CF, delayed puberty often stems from the impaired production of sexual development hormones. The timing of puberty can also depend on the overall health of the young man with CF, particularly in terms of nutrition, malabsorption, diabetes and lung disease. Once their hormones reach their peak levels, the majority of young men with CF do eventually go through puberty. In addition, most men with CF have normal testosterone production and often grow to achieve normal height, muscle mass, sex drive and other typical sexual features.

Young women with CF may also experience delayed puberty or get their first period after many of their friends and peers. Poor nutrition, low body weight and compromised lung function all affect the body's ability to ovulate and menstruate regularly. For this reason, some women with CF may have absent or irregular periods associated with being underweight or malnourished. Most women with CF have normal hormonal function, reproductive tracts and sexual development. However, in addition to absent or irregular periods, many women with CF experience other common reproductive health issues such as fungal vaginitis, and stress incontinence.

If your son or daughter is experiencing a lack of confidence due to delayed puberty, it may be comforting for them to understand that every adolescent goes through puberty at his/her own rate, whether they have CF or not. The CF care team is an excellent resource to help adolescents manage anxiety, insecurity and other emotional issues associated with delayed puberty.

Since delayed puberty is also associated with inadequate nutrition, low body weight, poor lung function, hormonal issues and other health concerns, it is important for parents and their adolescents to together talk to the CF care team about their development. This is an excellent opportunity for young adults to begin to take more responsibility of their own CF care, and be engaged in the discussion. The CF care team may be able to help jump-start puberty by finding ways to improve nutrition and to monitor or adjust treatment plans. By working with your CF care team, young adults can learn more about taking steps toward gaining weight, increasing caloric intake and improving general health to help encourage long-term growth.



Clinical Practice Guidelines

Pediatric CF Patients aged 2-5

In March of 2016, the CF Foundation published clinical practice guidelines for the management of pediatric CF patients aged 2-5 years. The CF Foundation invited CF experts and parents to develop key questions to be addressed in the extensive literature review.

A summary of some key recommendations include:

- Continue annual seasonal influenza vaccine and age appropriate vaccines/ well child exams through the primary care practitioner.
- Add the Pneumococcal 23 valent vaccine to be given at least 8 weeks after the last dose of the pneumococcal conjugate vaccine (the last dose is typically given at about 18 months of age).
- PFT's or spirometry should be attempted in children as young as 3 years.
- Chest x-rays at diagnosis and every other year. CT scans may be used as an alternative to chest x-rays and should be done every 2-3 years (this would replace the CXR every other year).
- Continue obtaining throat cultures every 3 months
- Continue to recommend airway clearance techniques to improve lung function and reduce exacerbations

- More intensive focus on nutrition management is recommended if BMI < 50 percentile; rate of weight gain < 50 percentile for age; weight-for-age < 10 percentile; or there is inappropriate weight loss. Nutritional supplements are encouraged.
- Dornase alfa (pulmozyme) and hypertonic saline may be continued to be offered to patients based on their needs.
- Continue anti-pseudomonal antibiotics in children with persistent pseudomonas infection.
- Add Ivacaftor in eligible patients with certain CF mutations.
- Salt should be added to meals and snacks especially during the summer months

The complete guidelines are published on CFF.org

How Are CF Clinical Care Guidelines Developed?

To help inform the standard of care delivered at accredited care centers, the CF Foundation brings together committees of subject matter experts to write guidelines on topics related to the care of people with cystic fibrosis. Committee members include doctors, nurses, respiratory therapists, dietitians, social workers and people with CF and their families. We are excited to say that Cindy Brady, DNP, was a co-author on the above guidelines.

Where appropriate, the Foundation may refer to or modify existing guidelines from other professional organizations. The Foundation supports the development of the following types of guidelines:

- Evidence-based guidelines developed from a systematic review of the best available evidence.
- Consensus-based guidelines developed primarily from the opinions and experience of experts.

CF clinical care guidelines are developed for members of the CF community, which includes health care professionals at CF care centers, other health care professionals who provide care to people with CF, as well as people with CF and their families.

Featured Recipes

Dog Days of Summer Ice Cream Sandwich Treats

Ingredients

- One 12-pack ice cream sandwiches
- One 12 ounce jar of caramel topping
- One 1.5 quart container ice cream of your choice. Examples: chocolate peanut butter, chocolate caramel fudge, double caramel cookie crunch
- Crushed up candy of your choice. Example: chocolate peanut butter cups, toffee, chocolate covered pretzels, chocolate covered crispy wafer cookies

Instructions:

- Line a 13-by-9 inch baking pan with foil, leaving about 1 inch of foil extending over the edges of the pan.
- Layer ice cream sandwiches in the bottom of the pan. Spread caramel topping on sandwich layer. Freeze for 1 hour.
- Spread ice cream on top and sprinkle with crushed candy bars. Cover and freeze for several hours or overnight.
- Use foil to lift uncut treats out of pan. Place on cutting board and cut into squares/portion size of your choice.

Adapted from Hy-Vee Summer 2016 recipe

Medication 101

Exercise Fun for the Whole Family

Regular exercise is tough and we all find reasons to avoid it. But it makes sense, for some pretty basic and powerful reasons:

Better overall health: Our bodies were made to be active. When we become inactive, we put ourselves at increased risk for heart and bone disease, diabetes and cancer.

More energy: When you improve your heart, lung and muscle function, you have more energy for daily tasks, as well as daily CF treatments.

Better lung function: Regular exercise can improve your ability to get mucus out of your airways.

More time with others: Taking part in physical activities with other people is a good way to motivate yourself and a great way to maintain social relationships.

The bottom line is that our bodies were made to move. Being inactive is unnatural for your body and can give rise to disease and disability.

To get started, consider building a family fitness plan where you write goals and track your family's progress. Remember that physical activity is a fun, healthy habit that makes you feel good, not a chore to be endured.

Abridge version from www.cff.org

Meet Our New Team Member Tami Vance

I started in my role as Cystic Fibrosis Social Worker in January, however took a hiatus in March to have my first baby, a boy named Sonny Wilder. He has quickly become my favorite person, next to my husband. I'm back from leave and excited to fully join the CF team here at Children's.

I received my Master's in Social Work from New York University and have my clinical license with the Minnesota Board of Social Work. I have worked with children, adolescents and adults in a variety of settings including non-profit, schools, and hospitals. I have clinical experience working with people suffering with a severe mental illness, anxiety, depression, eating disorders, trauma, as well as childhood and adolescent development.

My role on the CF team is to not only help patients and families navigate the many resources to manage their CF care, but integrate mental health care to support the emotional toll CF may have. I am here to listen, offer support and counseling, and provide resources and referrals when needed. I have enjoyed my time here so far and look forward to meeting you all at your next visit. In the meantime, do not hesitate to call with any needs or concerns.

Medical Research Updates

Studies Open for Enrollment

1. Saline Hypertonic In Preschoolers (SHIP-001): Patients with CF ages 36-72 months may be eligible. **Open for enrollment!**
2. Vertex14-661-107: A Phase 3, Randomized, Double Blind, Placebo Controlled, Parallel Group Study to Evaluate the Efficacy and Safety of VX-661 in Combination With Ivacaftor in Subjects Aged 12 Years and Older With Cystic Fibrosis, Heterozygous for the F508del CFTR Mutation and With a Second CFTR Mutation That Is Not Likely to Respond to VX-661 and/or Ivacaftor Therapy. Will Re-open for enrollment in October!
3. High-Frequency Chest Wall Oscillation Therapy Adherence: Utilization of Novel Data Reporting Technology to Measure Adherence among Children and Adolescents with Cystic Fibrosis. **Open for enrollment!**

To learn more about the Cystic Fibrosis Research Program contact:
Mahrya Johnson at 612-813-6384 or via email at *mahrya.johnson@childrensmn.org*
Anne Mills at 612-813-7756 or via email at anne.mills@childrensmn.org