The Best CF Care Takes Time
By Ruth DeVoogd, PNP

If you have been to CF Clinic recently, you have probably noticed that our team has grown and your visits may have lengthened as well. In addition to the provider (MD or PNP), nurse, respiratory therapist, dietitian, social worker, psychologist, behavioral health consultant and research coordinator, we are now fortunate to have physical therapists and a pharmacist too. You don’t need to see each of these people every clinic visit, of course, and we strive to make sure you see the team members you wish to and need to. We will be using our pre-clinic questionnaire form so that you can let us know what’s most important to you on the day of your appointment. There may be times when you don’t especially want to see one of us, but we may want or need to see you!

While we always try to be respectful of your time in clinic, if you have a real time constraint or another appointment, let us know at the beginning of your visit so that we can prioritize. You can also let us know (via MyChart or phone) in advance of your appointment if you have particular concerns you wish to discuss. While we recognize that your visits take a long time, usually between two to three hours, we are excited and proud that we have so many specialists committed to helping you or your child be as healthy as possible and live life to the fullest. Please let us know how we’re doing.

Special Combined Clinics Are Here!

It’s hard enough to come to CF clinic every three months, but when you also have to see another specialist (or sometimes more than one), it can be overwhelming. That’s why we have started a combined CF-Endocrinology clinic for people with CF related diabetes.

The clinic meets the fourth Friday of each month during the morning in the Multidisciplinary (MDC) Clinic space on the 2nd floor. Dr. Christine Chan is the main endocrinologist. The CF physicians will rotate through the clinic, so it is possible you won’t see your primary doctor on that day. Ruth DeVoogd, PNP, will be there regularly.

This type of clinic provides great opportunities for improved communication with you and your team members so that we can develop a more complete plan of care, while decreasing the amount of time you miss for work and school. We are also in the process of developing a combined CF-GI clinic, which we plan to start very soon.
In an effort to improve the transition experience for patients and families, we have formed a working group between National Jewish Health and Children’s Hospital Colorado. The group includes nurses, social workers, psychologists, providers, and patient navigators from both the adult and pediatric CF centers. We all share a common interest in improving this process for patients and families so they feel cognitively and emotionally prepared for the change in their CF care team as children enter late adolescence and adulthood. Since our first meeting, we have already made some changes including how new patient appointments are scheduled at National Jewish, and the way we submit records and patient summaries to National Jewish from Children's.

Some other aims of our group include:

*Standardized practice among Children’s providers of when to introduce varying topics and education to patients.

*Provide opportunities for patients to meet an adult provider/nurse prior to transitioning.

*Increase communication between pediatric and adult CF teams surrounding upcoming transitions and completed transitions.

Please contact us if you have any additional suggestions or feedback that would be valuable to this process.

Meet Ben Hughes, MD—New Grand Junction Outreach Clinic Provider

I attended medical school at the University of Texas Medical Branch and completed pediatric residency and pulmonology fellowship at Children's Hospital Colorado and sleep fellowship at National Jewish Health. I have an amazing wife, Claire, and a handsome baby boy, Findlay, born last June. My hobbies include running, biking, coffee roasting, hiking, camping, and photography.

I am very excited to provide pulmonary and sleep care on the Western slope and look forward to meeting all the families!

Reminder! MyChart messages are not monitored after-hours, on weekends, or on holidays. Responses can take up to two days during regular business hours. If you have an urgent need during these times, please call 720-777-6181.

Improving The Transition Process
By Jill Hammond, BSN, RN, Clinical Nurse III

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Please contact us if you have any additional suggestions or feedback that would be valuable to this process.
Welcome To Our New CF Respiratory Therapists, Pharmacist, and Behavioral Health Clinician!

My name is April Garland, I am one of the CF Respiratory Therapists in the CF Clinic. I started working at Children’s Hospital Colorado in 2001. Outside of work, I love rock climbing, skiing, and running! I also enjoy spending time with my three daughters. I am very excited to be working in the CF Clinic and being part of this team!

My name is Emily Hofmann and I am the newest Respiratory Therapist to join the CF team. I started working at Children’s Hospital Colorado in November 2015. I worked inpatient on the Pulmonary floor and in the Pediatric ICU. In August 2017, I decided I wanted to focus on CF and help our patients in the outpatient CF clinic! Outside of work I enjoy spending time with my fiancé and my family. We have two dogs, I love to hike and to go camping! I am so excited to be a part of the amazing team we have in the CF clinic! I look forward to meeting all of you!

Hello! My name is Laney Brennan, and I am the CF and Pulmonary Pharmacist at Children’s Hospital Colorado. I graduated from Creighton University with my Doctor of Pharmacy degree in 2003, and completed two Pediatric Pharmacy Specialty Residencies here at CHCO in 2004/2005. After that, I practiced in the critical care units before becoming the Breathing Institute Pharmacist in 2007. I truly enjoy working with the CF multidisciplinary team and being involved with the comprehensive medical care of CF patients and their families. Until early 2017 most of my time was spent on the inpatient pulmonary unit in the hospital, but I now spend part of my time in CF clinic assisting with medication therapy and needs in the outpatient setting. When I am not at work I am enjoying our fabulous Colorado mountains, parks and sports with my husband, two children and our Labrador Retriever. I am looking forward to meeting and working with many more CF patients and families!

My name is Jennifer Girard and I am a behavioral health clinician in the CF Center at Children’s Hospital Colorado. I have been working with you and your families since May 2017. I am a Licensed Professional Counselor and I hold a doctoral degree in behavioral health from Arizona State University. I have clinical experience in substance use, domestic violence, as well as integrated primary care and specialty care, addressing chronic disease, adherence issues, lifestyle change and co-morbid mental health conditions. I have deeply enjoyed working with patients and families around the impact of CF, coping, stress management, social-emotional concerns, adherence and simply getting to know you. I am a native Coloradan, so I make sure to take time with my family to enjoy all that this beautiful state has to offer.

Save the Date for CF Family Education Night

Please plan to attend our next CF Family Education Night on Saturday, April 21st from 4:00-7:00pm.

We are very excited to host Gunnar Esiason as our featured speaker for this event!

Gunnar Esiason is 26 years old with cystic fibrosis. After being diagnosed at 2 years old, his parents founded the Boomer Esiason Foundation. Since then Gunnar has grown to live his life despite CF. He played three varsity sports in high school, graduated from Boston College and now works as a cystic fibrosis patient advocate with the very foundation his parents started. He is also the head coach of his high school alma mater’s varsity ice hockey team, making him the youngest head coach on Long Island.

Gunnar has learned that success with cystic fibrosis is possible, and he hopes to share that with our patients and families in April!

We hope to see you all at this exciting event on April 21st! Please RSVP at cysticfibrosis@childrenscolorado.org
Have you gotten your 60 minutes today?

Try these exercises to strengthen your core & upper body, and get your heart rate up!

Physical Therapists Join CHCO CF Team

Physical therapists Abby Good, Jackie Therrien, and Allison Brown are excited to join the CF team. The role of the physical therapist (PT) on the CF team includes assessment of the muscle and skeletal system, posture, cardiac endurance, pain, motor skills and coordination, as well as promoting an active lifestyle.

In clinic, your PT’s are available to encourage exercise and activity and address concerns such as shortness of breath, low endurance, or difficulty keeping up with peers. Strong evidence exists that regular physical activity is important for all patients with CF. Exercise is important in managing CF, including airway clearance, improved lung function and cardiovascular fitness, a positive impact on the lifespan, improved self-confidence and stamina, and increased participation with peers.

Abby, Jackie and Allison will screen and evaluate children in clinic, once they reach school-age. They are looking forward to getting to know families, and encourage you to use physical therapy as a resource during your clinic visits.

Meet the PTs!

Allison Brown
Education and training: Dual Bachelor’s degree in Health Science Education and Public Health from the University of Nebraska-Lincoln and a Doctorate in Physical Therapy from Creighton Medical Center. She has been a physical therapist for over 20 years in many service areas, including the NICU, PICU, general inpatient, and inpatient rehab. Served a CF center in Ohio for 8 years, working with both pediatric and adult CF patients. Currently working: Part time on the Colorado Springs (Briargate) Physical Therapy team, treating patients with orthopedic conditions, injuries, or surgeries, as well as connective tissue disorders, pain, or concussions. In addition to her work at Briargate, Allison teaches high school and junior high biology, as well as homeschooling her children with her husband.

Abby Good
Education and training: Attended University of Vermont. Obtained Bachelor’s degree in Elementary Education and Doctorate in Physical Therapy. Experienced in many pediatric settings, such as early intervention and school-based therapy. Certified infant massage and restorative and vinyasa yoga instructor. Was a collegiate athlete and still enjoys soccer, skiing, hiking, triathlons, and tennis. She provides a holistic approach to care by combining her skills and experiences with her perspective on child development and different models of care. Currently working: at Children’s Parker therapy care clinic with orthopedic patients of all ages, and is part of the foot management team. She runs a yoga group in Parker for patients with orthopedic conditions such as back and neck pain, and postural impairments. Will be working in CF clinic on Wednesdays and Fridays.

Jackie Therrien
Education and training: Attended Quinnipiac University in Connecticut. Obtained Bachelor’s degree in Science and Doctorate in Physical Therapy. She enjoys an active lifestyle including hiking, running, swimming and yoga. Has worked in Boston and at Children’s Colorado in early intervention home care and clinics. Has integrated her experience with early childhood development, transdisciplinary care, and play-based therapy to help children and families in their daily lives.

Currently working: at Children’s Parker therapy care clinic with neurodevelopmental patients, as well as children who toe-walk and infants with torticollis. Will be working in CF clinic on Mondays and Tuesdays.
Just Breathe: Stress Management Tips
By Jennifer Girard, DBH, LPC and Emily Muther, PhD

Stress affects us all and can be healthy in small doses. Stress can propel us forward to take action to improve our lives. However, too much can be harmful. Here are some ideas to address stress for your family. If you feel like stress is an ongoing concern for you or your child, please ask to talk with us or give us a call; we specialize in helping you and your family cope with these challenges.

**Fun:** This may seem obvious, but when we experience a great deal of stress and have no way to release the tension, it builds up. Engage in enjoyable activities by yourself, with your family, and/or with friends. Go outside and play!

**Social Interaction:** It is very important to engage in social interaction with family/friends. As humans, we function best when we have the support of others. Call a friend and make plans!

**Exercise:** It does not matter what you do, but it is important to engage in physical activity to produce natural chemicals in your brain to improve mood, increase relaxation and reduce stress. If you are looking for creative ways to do this with your family, see our CF Physical Therapists!

**Diet:** What we eat affects how we feel, and a balanced diet for you and adherence to the diet you discuss with the CF dieticians on a regular basis for your child with CF, is important in fending off out of control stress.

**Sleep:** This is often the first thing to go by the wayside when we are busy, and among the most important for stress management. The following are useful sleep hygiene tips: If you are not asleep in 20 minutes, get out of bed, create a healthy sleep routine that is the same daily, get up at the same time every day, avoid naps, avoid alcohol and caffeine within 6 hours of bedtime, try to avoid electronics for an hour prior to bedtime, and make sure the bedroom is quiet, dark, and little bit cool.

**Create a daily routine:** We are creatures of habit for a reason. If we know what needs to be done each day and remain organized, we tend to be less stressed. Set achievable goals so you can feel successful, same goes for your children. Reward yourself and your children when goals are accomplished. Celebrating success helps us feel motivated to keep working hard.

**Self-care:** Take time for yourself to do the things that make you, well, you. The same goes for your children. Taking care of ourselves gives us the energy to help care for others around us. Ask for help when you need it!

**Therapy:** Therapy can be preventative and a place to address stress. For children with CF, therapy is often recommended to promote resiliency and explore the impact on well-being.

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**Relaxation Strategies**

| Mindfulness | The practice of focusing awareness on the present moment. |
| Deep Breathing | Put your hand on your stomach (or have your child lie down with a cup/toy on their belly), deep breaths in through your nose and out through your mouth. You should feel your stomach inflate with air and deflate as you exhale. |
| Progressive Muscle Relaxation | A simple technique that involves tensing and relaxing all of the major muscles in your body in order from your head to your feet. |
| Guided Imagery | A relaxation technique in which words, sounds, etc. are used to evoke positive mental images, feelings and thoughts. |
| Yoga/Meditation | Breath control, simple meditation, and the adoption of specific bodily postures, is widely practiced for health and relaxation. |

**Resources**

- Breathe2Relax, Calm, Breathe
  Stress Management Apps

- T2 Mood Tracker App
  Tracks mental health, stress and well-being

- Take a Break! App
  Guided Meditations for Stress Relief

- BellyBio Interactive Breathing App
  Biofeedback device for anxiety and stress

- Children’s Hospital Colorado
  Stress Management Guide

- Cystic Fibrosis Foundation Website
New FDA-Approved Treatment

Tezacaftor-ivacaftor (Symdeko) is a new FDA-approved treatment for people with CF, 12 years and older with two copies of the F508del mutation or one F508del mutation and a second residual function mutation.

Children’s Colorado participated in clinical trials of tezacaftor-ivacaftor and results were presented at the NACFC. While tezacaftor-ivacaftor appears similar in how well it works compared to lumacaftor/ivacaftor (Orkambi), fewer side effects were seen. People with a residual function mutation who took tezacaftor-ivacaftor did better compared to those who just took ivacaftor (Kalydeco).

If you are interested in whether Symdeko is appropriate for you, please talk to your CF provider.

CF Teen Advocacy Day

My name is Micaela Vaughn and I am 18 years old. Over the past two summers, I have participated in the Cystic Fibrosis Teen Advocacy Day on Capitol Hill in Washington, D.C.

While participating in TAD I was able to tour D.C. and Capitol Hill and had the honor to meet with multiple congressmen to discuss and advocate for CF. I have advocated for increased funding for the NIH and FDA along with adequate health care. In addition to meeting congressmen, I met other teens my age, who, like me, have a sibling with CF. It was really refreshing to talk about CF and relate to what they are going through.

The impact of Teen Advocacy Day on both the CF community and myself is unimaginable. I highly encourage any teen who has a family member or friend with CF to participate in TAD because it is truly life changing.

For more information on advocacy in the CF community, please visit https://www.cff.org/Get-Involved/Advocate/Our-Advocacy-Work/

Highlights from the 2017 North American Conference (NACFC)

By Edith Zemanick, MD, MSCS

The annual NACFC was held in Indianapolis, Indiana in November. Over 4,000 medical professionals, researchers, CF care team members and families attended the conference to share updates in CF treatment and care. Members of the CF team from Children’s Hospital Colorado attended the conference and many led sessions on topics such as nutrition in CF (Judy Fulton, RD), Telehealth (Ruth DeVooogd, PNP) and Mental health and CF (Emily Muther, PhD). Drs. Scott Sagel, Edith Zemanick, Stacey Martiniano, Jordana Hoppe and Christine Chan (CF diabetes specialist) presented results of ongoing research projects.

The three main lectures focused on new CFTR modulator treatments, advances in lung transplants for people with CF who develop severe lung disease, and advocacy for policies that improve access to appropriate and affordable health care for people with CF.

CFTR modulators are medications that improve the function of the CF protein (called CFTR) that result in improved respiratory symptoms, lung function and nutrition. Currently available treatments, ivacaftor (Kalydeco), lumacaftor/ivacaftor (Orkambi) and most recently tezacaftor/ivacaftor (Symdeko), work for people with certain CF genetic mutations (two copies of the F508del mutation or rarer mutations known as “gating” or “residual function”). Thus, researchers are working hard to develop treatments that will work for all people with CF, including those with one F508del and those with minimal function mutations.

One of the highlights in CFTR modulators was the release of phase 2 data on triple-combination treatments. The triple combination approach adds a third medication to tezacaftor-ivacaftor with the hope of increasing the amount of CFTR protein that works. Results for phase 2 studies of two compounds (VX-659 and VX-445) were particularly strong with improvements in lung function of ~13% in people with one copy of F508del and one copy of a “minimal function” mutation. This is very important because currently there is no CFTR modulator available for people with F508del plus a minimal function mutation. Two phase 3 studies are planned for 2018. Phase 3 studies are the large clinical trials needed to determine safety and how well a medication works before the FDA considers approving it. These studies will test VX-659 and VX-445 in combination with tezacaftor-ivacaftor in people with CF ages 12 years and older. We are hopeful that these studies will lead to a new treatment option for more people.

Results from other studies were presented, including many of which our site has participated in. The OPTIMIZE study compared treatment of Pseudomonas aeruginosa infection with inhaled tobramycin (TOBI) versus TOBI plus azithromycin 3 times per week. While adding azithromycin did not reduce later infection with Pseudomonas, children who received azithromycin had a longer time before their next respiratory illness (pulmonary exacerbation). Chronic azithromycin given three times a week has previously been shown to decrease the number of pulmonary exacerbations that people with CF experience, and this study supported those findings. Other studies, PROSPECT and GOAL, followed people who were started on lumacaftor-tezacaftor or ivacaftor. Findings from these studies support that these medications improve respiratory health and lung function over time.

The NACFC is a great opportunity for care team members and families to learn from others around the world who are working to improve treatments for CF. We are already planning for the 2018 NACFC conference - to be held in Denver, Colorado!
The Cystic Fibrosis Foundation (CFF) manages an online, nationwide registry, or database, of patients at CF centers around the United States. This registry is known as PortCF, and currently contains information on more than 48,000 CF patients. PortCF tracks patients’ clinic visits and admissions, their physical growth, prescribed medications, pulmonary function test scores, and what kinds of bacteria they are growing in their lungs. If a patient is under 18 years of age, a parent or legal guardian must consent to allow the patient’s information to be entered in the registry. Once the patient is 18, he or she can choose whether to continue to participate in PortCF. The CFF estimates that approximately 84% of all CF patients in the U.S. currently participate. Other countries, such as Canada and Australia, have their own CF registries. The patterns seen in these other countries’ registries sometimes differ from ours in ways that researchers are working to understand.

Dr. Warren Warwick created the original, paper-based registry at the University of Minnesota in the 1960s. In 1986, the CFF took over maintenance of the registry under the leadership of Dr. Bob Beall. In those years, CF care centers would mail annual questionnaires about their patients to the CFF. In the mid-1990s, staff at CF centers started saving patients’ information on floppy disks and mailing the “floppies” to the CFF four times per year. In 2003, the registry moved online and PortCF was born. As the registry has evolved, its usefulness has grown, too.

The registry is an incredible tool in the fight to cure and control CF. Using registry data, we learned the importance of excellent nutrition for lung health, and of regular clinic visits for preventative care. Each year, our center receives a registry report that shows how well our patients are doing compared to other centers. This information helps us to see what we are doing well, and what we need to improve. For example, our center typically has excellent lung function, but we often don’t see children in clinic as frequently as recommended. Because of this, we are now working to make scheduling visits easier to make sure all children can get the routine care that they need. Many other conditions are now trying to develop registries to help understand and treat diseases; in CF, we are fortunate that leaders such as Dr. Warwick and Dr. Beall had the foresight to develop this resource over 40 years ago.

Get To Know Our Research Coordinators!

**Meg Anthony, LCSW, CCRC**
I am a Licensed Clinical Social Worker (LCSW) and a Certified Clinical Research Coordinator (CCRC). I have been working in CF research for 10 years. I was the CF social worker for 14 years and slowly became involved in research through family-centered research studies. Later I transitioned to be a full-time research coordinator. Working with CF patients and families has been so rewarding and an honor to be a part of making people with CF’s lives a little better.
Fun fact: My oldest daughter got married October 1, 2016. She was just 2 years old when I first started working at the CHCO CF Center!

**Dana Coyle, RRT**
I am a Registered Respiratory Therapist (RRT) and I have worked in the CF field for over 25 years, though just started in CF research 9 months ago. I first got interested in CF at Children’s National Medical Center in Washington, DC. My very first CF encounter was with a young lady who told me how much better she felt after I finished her treatment. I was hooked after that. She went on to graduate college and get married. We keep in touch to this day.
Fun fact: My favorite hobby, by far, is traveling! To date, my favorite place is Santorini, Greece

**Mary Cross, RRT, CPFT**
I am a Certified Pulmonary Function Therapist and RRT. I have been working in research for 3 years, in CF as an RT for 6 years. I enjoyed working with the CF patients while working as an inpatient RT and was excited for the chance to get into research with that group.
Fun fact: I love marine critters and enjoy scuba diving with my husband.

**Rebecca Infante, RRT**
I am an RRT and have a bachelor’s degree in Project Management. I have been a CF research coordinator for about two and half years. Working with CF patients as inpatients and getting to know them and their families really got me interested in CF and helping this patient population.
Fun fact: In my free time I enjoy crocheting and scrapbooking, reading, and going to see new places and things with my kids and husband.

**Carol Kopecky, RRT**
I am an RRT in the PFT lab. I have also been working for 5 years as a part-time Research Coordinator but I have been doing research procedures for 9 years. I became interested in CF while working as a respiratory therapist.
Fun fact: I have 4 grandsons!
<table>
<thead>
<tr>
<th>Study Name</th>
<th>Overview</th>
<th>Age</th>
<th>Duration</th>
</tr>
</thead>
<tbody>
<tr>
<td>CF Registry</td>
<td>Collects information on people with CF cared for in CF Foundation-accredited care centers to create CF care guidelines, guide quality improvement initiatives, and inform clinical research and design CF clinical trials.</td>
<td>All ages</td>
<td>No time limit</td>
</tr>
<tr>
<td>GOAL e2</td>
<td>A natural history study of subjects on ivacaftor (Kalydeco) therapy.</td>
<td>6 years and older</td>
<td>Varied</td>
</tr>
<tr>
<td>Nontuberculous mycobacterial (NTM) Infection Diagnosis and Treatment</td>
<td>To study how having a positive NTM respiratory culture affects treatment and CF care.</td>
<td>All ages</td>
<td>No time limit</td>
</tr>
<tr>
<td>GROW</td>
<td>Comparing oral glutathione to placebo in kids 2-10 years old to see if it improves growth in children with CF.</td>
<td>2-10 years</td>
<td>About 6 months</td>
</tr>
<tr>
<td>Outpatient Pulmonary Exacerbation</td>
<td>To study the effectiveness of outpatient oral antibiotic therapy for a pulmonary exacerbation.</td>
<td>8-18 years</td>
<td>Up to 2 months</td>
</tr>
<tr>
<td>Success with Therapies Research Consortium (STRC) Tele-Coaching</td>
<td>A study to obtain patient and parent feedback and input into a Tele-Coaching intervention.</td>
<td>14-18 years</td>
<td>Up to 9 months</td>
</tr>
<tr>
<td>AeroVanc “AVAIL”</td>
<td>A study to test the effectiveness of Inhaled Vancomycin (AeroVanc) for the treatment of chronic MRSA.</td>
<td>6 years and older</td>
<td>Up to 13 months</td>
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<tr>
<td>VX17-659-102</td>
<td>Vertex study for the next CF modulator. Compares three-drug combination against placebo.</td>
<td>12 and older</td>
<td>About 32 weeks</td>
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<tr>
<td>Rare-OB-11</td>
<td>Rare CFTR Mutation Cell Collection Protocol</td>
<td>Over 2 years</td>
<td>1 visit</td>
</tr>
</tbody>
</table>

There are many other active studies that we are working on. If you have any questions about our work or if you are interested in participating in a research study, please contact Meg Anthony at 720-777-2945 or meg.anthony@childrenscolorado.org.