CF Family Advisory Council
ATLANTA, GEORGIA

The purpose of the CF Advisory Council is to work together with staff to advance the practice of family-centered care — the belief that healthcare providers and families are partners working together to improve quality care for patients. This is accomplished by designing better programs, policies and procedures that provide the best possible care and meet the needs of the CF patient and their families.

Let us hear from you.

- If you have newsletter story ideas
- If you or your child would like to be featured in the spotlight section
- Share your favorite recipe
- Nominate a CF Superhero
- Have questions or comments you would like FAC to address

Email to: stickynotesnewsletter@gmail.com

To the parents of:

CF Family Advisory Council
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Sticky Notes

Fall 2015

CF Foundation Celebrates FDA Approval of Orkambi

By Dr. Seth Walker, MD, FCCP, Director of Emory Adult CF Clinic

Orkambi (lumacaftor/ivacaftor) is the latest drug in our fight against CF. It only appears affective in patients with 2 copies of the deltaF508 mutation. It works by allowing the deltaF508 protein to escape the cell's quality control mechanisms and be placed at the edge of the cells membrane where it needs to be to function. It also increases the activity of the protein once it is there. In a large study that led to its approval, there was an average improvement of 6% FEV1 and average decrease in exacerbations by 35%. A decrease in exacerbation rate is important, as we believe exacerbations cause a significant amount of the lung function loss over time in CF patients. All approved CF therapies have been shown a persistent decrease in exacerbations, and this may be the greatest benefit patient actually receive from their preventative CF medications.

The drug was well tolerated in the study, but some patients did have elevations in the liver enzymes. No patient had liver failure or liver injury, and often the drug was restarted after liver enzymes returned to normal without any further elevations, suggesting there may have been another cause for the abnormal labs. Many patients noticed some chest tightness when starting the mediation. In most patients, this was insignificant. In some, they used albuterol with benefit and this symptom went away after 2-3 weeks, but less than 1% of patients had decline in lung function and were unable to take the medication. In my experience and in talking with colleagues around the nation, this has been happening more frequently than in the study, especially in patients whose FEV1 is 40-5-% predicted. Lastly, Orkambi did have interactions with several drugs, most notably anti-fungals that are taken orally or through IV. It will also make hormonal contraceptives ineffective, so female patients will need to use another form of birth control.

Orkambi is not a cure. The benefits seen were in patients who were believed to have been adhering to their regular CF therapies. This pill was designed to be an add-on to current treatment, not a replacement for any therapy. It also is likely an intermediary until we find better compounds. Currently there are other drugs that are being evaluated for a better side effect profile, less drug interactions, and applicable to more genotypes.

CALENDAR OF EVENTS

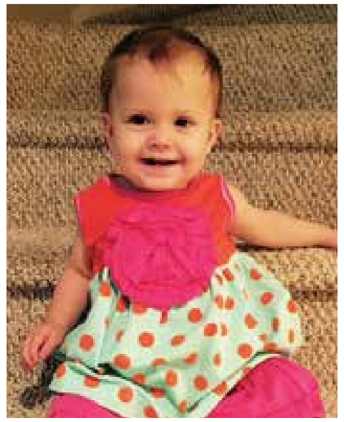
January 21, 2016
CF Nutrition Night

March 12, 2016
ShamRockin' For A Cure

April 11, 2016
Danile B. Caplan Family Science Dinner

May 21, 2016
Great Strides Atlanta

Patient Spotlight



Name: Parker
Age: 1
Hometown: Woodstock, Ga.

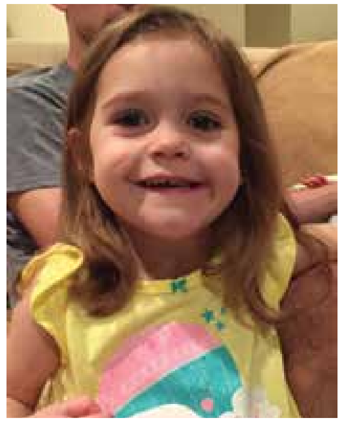
FAVORITES:

Food: Bananas, chicken nuggets, puffs, yogurt, and much more!
Book: Where is baby's belly button
One word that describes you: Vivacious

Accomplishment: Starting to walk

One of your favorite things to do with your family: Playing with my sissy on our bouncy house

Your role model and why: My sissy, Riley because she always encourages me to take my enzymes and eat good. And because my sissy and I have a very special bond.



Name: Riley
Age: 3
Hometown: Woodstock, Ga.

FAVORITES:

Food: I can't just pick one. I have lots of favorites! Mac and cheese, hotdogs, spaghetti, chicken nuggets, and eggs and sausage
Movie: "Frozen"
Book: Who I am (the cf book with Patti)

Favorite Sport: Golf because my daddy is an avid golfer and he teaches me how to putt and even made me my own putter.

Accomplishment you are proud of: I sleep In a big girl bed now that is Minnie Mouse.

Advice you'd give other kids who have CF: Always do your "shake shake" and take your enzymes. Also being active and playing outside.

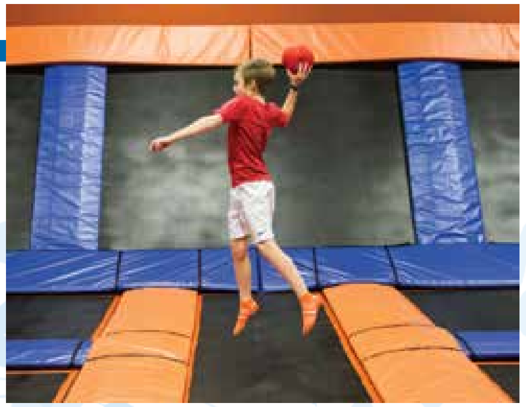
One of your favorite things to do with your family: I like to go to my Mimi's beach house in Fort Myers Beach, FL. I also like to go to the park and jump in my bouncy house.

What are you looking forward too next year? I look forward going to preschool.

Your role model and why: my daddy, he is always there for me and pushes me to be active. He also teaches me to be strong and tough and never let CF bring me down or define who I am.

Kid's Corner

Get Creative in the Cold



When it gets cold outside, you need to turn creative to keeping your family active.
Here is a list of the top five activities for both inside and outside fun.
Tell us what you think - post your favorites on our Facebook page.

1. Get Active - SkyZone Indoor Trampoline Park, Atlanta Rocks or Stone Summit Climbing Gym.
2. Visit a Museum - Fernbank Museum of Natural History, High Museum of Art, Imagine it! The Children's Museum of Atlanta, Tellus or Booth Western Museum (both in Cartersville).
3. Build Something - Home Depot or Lowe's Workshops.
4. Brave the cold outside - visit the Atlanta Zoo, hike Kennesaw Mountain, ice skate in Centennial Olympic Park or Piedmont Park or visit Stone (Snow) Mountain.
5. Play tourist - Georgia Aquarium or the World of Coca Cola.

CF SUPERHERO

Timothy Beaty, MD, MSc
Assistant Professor
Emory University School of Medicine
Department of Pediatrics
Division of Pulmonology, Allergy/Immunology, Cystic Fibrosis, and Sleep

Where did you grow up?
I grew up in the once small town of Trussville, AL, population of around 2000 people when I was young. I remember when the McDonald's opened and the whole town celebrated, because before we only had a Hardee's!

Who is your family?
My wife of 8 years Ayumi and two children; Joshua – 2 years, and Penelope 4 months.

How long have you been in the field of CF?
I've been taking care of patients with CF for 6 years, but most of that in training as a pediatric resident, then pulmonology fellow.

What was the decision that led you into the field of CF?
I always imagined myself as a small-town Doc, because I wanted to have strong connections with my patients, to really make a difference in someone's life. During my residency, my experience was that the strongest relationships any physicians had were the pulmonary doctors with their CF patients. (If I may tell a story) The first day of clinic a 3 year old with CF saw her doctor from far away down the hall, gave a huge smile, and ran into his open arms. The next patient was graduating from college and asked the same pulmonologist to write a letter of recommendation for law school. He then went on to tell a story of her 9 year old soccer team – he really KNEW her. The opportunity to be a part of someone's life like that and help them achieve their dreams really pulled me into pulmonology and working with CF in particular.

Where did you attend college?
Auburn University (War Eagle!)

If you could have one superpower what would it be?
This is toss-up between being able to reverse time to take back idiotic things I say and the ability to eat all the pizza I want without gaining weight (or hardening my arteries!).

What is one thing you miss about being a kid?
Having too much time on my hands!

Name one word that describes you:
Patient.

What is one accomplishment that you are proud of?
Despite being one of the smallest and slowest players on the team, earning a starting spot in football through hard work.

List one thing that most people do not know about you?
When I had time to practice (see question #7), I was a pretty good guitarist. 15 years ago, I was actually paid to be the entertainment at the Alabama Tax Collector Convention, so I still consider myself a "professional" musician.

Links of Interest

cff.org
georgia.cff.org
milesforcysticfibrosis.org
reachingoutfoundation.org
choa.org/cf
happyheartfamilies.com
cfvoice.com
cysticlife.org
shamrockinforacure.com

White Cheddar Mac and Cheese

Ingredients:
1 pound farfalle pasta
2 tablespoons unsalted butter
2 tablespoons all-purpose flour
2 cups milk
1/2 teaspoon salt
1/4 teaspoon black pepper
1/4 teaspoon ground nutmeg
1 pound white cheddar cheese, shredded (about 4 cups)
1 package (10 ounces) frozen peas, thawed
1/2 pound finely diced ham

Make It:

1. Heat oven to 350 degrees . Coat 13 x 9 x 2-inch baking dish with nonstick cooking spray. Cook pasta following package directions. Drain.

2. While pasta is cooking, melt butter in a medium-size saucepan over medium heat. Stir in flour until well blended; cook 1 minute. Whisk in 1 cup of milk and cook, whisking continuously, until smooth. Whisk in remaining 1 cup milk and continue to cook, whisking until thickened and smooth, about 5 minutes. Remove from heat; stir in salt, pepper and nutmeg. Whisk in 2 cups of the cheese until smooth.

3. In a large bowl, mix together the pasta, cheese sauce and 1 cup of the remaining cheese. Stir in the peas and ham. Pour into prepared baking dish. Scatter remaining 1 cup cheese evenly over the top.

4. Bake at 350 degrees for 30 minutes or until bubbly. Remove from oven; allow to cool 10 minutes. May be served warm or at room temperature.

Nutritional analysis (per serving)
Servings Per Recipe: 12; Calories: 372, Fat: 17, sat. fat (g): 10, carb. (g): 34, fiber (g): 3, protein 21 gram, sodium (mg): 625, Percent Daily Values are based on a 2,000 calorie diet.