

Cough it Up, Spit it Out!

Pediatric Pulmonology CF Center Updates

A Division of Prisma Health Children's Hospital-Upstate (864) 454-5530

Special Interest Articles:

- Pot or NOT.
- Phase 3 clinical trial looks good.
- Hypersal really works.

Individual Highlights:

Movie Review: Five Feet Apart	2
Mental Health	3
Recipe Corner	4
Birthdays!	5
Facility Dogs	6

We're excited to share BIG NEWS!!!

The Cystic Fibrosis Foundation has awarded our CF Center with the "Outstanding Partnerships Between Care Centers and CF Foundation Chapters Award!"

This is such a wonderful honor, as there are currently 282 CF Care Programs and we were one of 14 to be distinguished with this award. The award was initially announced at the North American CF Conference in November

of 2018. Our team was presented the award by our own CF Foundation representatives; Jen Nielson, Megan Onysko, Jamie Bierdz & Annie Carenbauer.

This award is in a large part to our families. YOU are the reason we fight so hard to deliver the care that will improve your child's and family's quality of life. Our affiliation with the Cystic Fibrosis Foundation only helps to cement that care. The

CFF provides us with guidelines, education, resources and events all working toward improved care and the eventual cure. Our team THANKS the CFF, especially our SC staff, and each of you for trusting us with the care of your loved ones. If you have not already gotten involved with the CFF consider reaching out via: 1-843-388-5968 or monysko@cff.org.





“This whole time I’ve been living for my treatments, instead of doing my treatments so that I can live, and I want to live.”



Hmmmm, is it POT or not?

Our social work listserv shared the article below:

The CFF Mental Health Advisory Committee would like to bring to your attention a recent publication in the Journal of CF showing a very high rate of false positive urine drug screens in people taking Orkambi. Of 22 adults taking Orkambi, 21 tested positive for cannabinoids by immunoassay.

Confirmatory gas chromatography-mass spectrometry analysis showed these to be false positive screens. CF Clinicians and patients should be aware of this, as it may have important implications for those undergoing urine drug screening, including for employment or transplant evaluation purposes.

If you have further questions regarding this or

any mental health question please email, mentalhealth@cff.org.

Five Feet Apart, a review from a Nurse Practitioner

Okay, so full and honest disclosure. I am the person at our clinic who keeps the Infection Prevention & Control policies up to date and I coordinate with hospital staff to be sure other departments and facilities follow the CFF guidelines.

Okay, now that is off my chest and y’all can realize where my initial thoughts on the movie stem from. My younger daughter Kate, age 18, who has met many of our patients at Great Strides and other events was with me and within the first 10 minutes she had to fuss at me because I was completely **UNDONE** that the children were **NOT** wearing their masks while in the hospital. Okay, stop laughing, you know I am the “mask queen” in our office. Once I got **OVER** the mask issue and the

children spending time together in the atrium (an enclosed indoor room), I did settle down.

I will not spoil the story but I will share what I liked about the movie and what I have read others with CF liked.

CF is **NOW** on the forefront, people actually know what it is. The movie showed the characters coughing up secretions and **SPITTING THEM OUT!** YAY! This is a hard concept for some of our kids to grasp as they often want to suppress their coughing/secretions.

The movie showed the treatment regimens, the medications, the gtube supplements and the isolation of being hospitalized.

NOBODY realizes the lives our kids lead,

because most of the time when you see them, it is not obvious they have a disease which literally takes **HOURS** a day to manage with treatments, medications, etc.

I liked how the patients would make their rooms their own and have a fairly independent relationship with the hospital staff. This is an area we work on with your kids through our CF Rise program.

CFF is recommending your children **NOT** go to the movie and instead wait until it is on Netflix, etc., again due to infection concerns. Family and friends who know about CF will likely find some aspects of the movie are not at all realistic, but remember, this is Hollywood and we just got a lot of free publicity!

Promising Phase 3 clinical trial results

November 27, 2018 was an exciting day, not because Christmas shopping was in high swing, BUT because CF Foundation made the announcement that Vertex pharmaceuticals had a promising Phase 3 clinical trial for VX-659 combined with ivacaftor and tezacaftor. There is also another modulator VX-445 combined with ivacaftor and tezacaftor currently being studied and hopes are for results very soon.

The clinical trials are currently being done on those patients age 12 and

older who have 2 copies of the F508del mutation and another study on those with only 1 copy of F508del and another allele.

Studies are being done at a variety of centers across the country and the entire CF Community is anxiously awaiting the findings of which combination is providing the most benefit. Plans will then be for Vertex to submit a new drug application to the FDA in late 2019 for potential approval.

The implications for the triple therapy combination are quite exciting as it means that about 90% of those affected with Cystic Fibrosis would have a modulator treatment option.

If you are interested in more information please look at the CFF website at:

www.cff.org.



Promising results with Potential triple-combination therapy with CFTR modulators

Mental Health Minute courtesy of Christy Clarke, LMSW

Parents, do you have to nag your kids to get them to do their treatments? Do your kids try to push your buttons and test the limits? You are not alone!

We want to provide you with some resources that include some tried and true tips for you to gain cooperation from your kids. We have ordered for you the condensed version of Parenting Children with Health Issues and Special Needs written by Foster Cline, MD and Lisa Greene (certified parent coach and parent of two kids with CF).

We'll be giving out the booklets at clinic the next several months. We are hopeful that these booklets will give you some tips on engaging, communicating and

ultimately gaining cooperation from your kids. Here are a few of the practical tips in the booklet:

Share your thoughts and ideas with your child and allow them to make affordable mistakes. Model taking care of yourself.

Give your child choices - would you like to do your treatment now or after dinner? Would you like butter or half and half in your potatoes?

Say "Yes!" with limits. For example, "Yes, you may have a cookie right after you finish your dinner!"

Validate feelings of frustration and encourage

thinking about wise choices. For example, "I'd rather do something else than a breathing treatment too if I were in your shoes but what does the doctor say about treatments?"

Kids make mistakes – be sad for them, not mad at them. Try to respond with empathy rather than anger.

Respond with curiosity rather than blame. "What could you have done differently? What have you learned from this?"

Never underestimate the power of love to bring hope.

In addition to this booklet, Lisa Greene has written multiple CF specific pamphlets for each stage that includes many of the same tips. We have the pamphlets in clinic so feel free to ask for one. Also, check out these websites for additional parenting tips:

<http://www.parentingchildrenwiththehealthissues.com/index.html>

<https://www.loveandlogic.com/blog/love-and-logic-blog-for-parents-and-teachers>

<http://www.happyheartfamilies.com/TipsForCFParents.html>

Lastly, we think YOU are doing a great job! We see the sacrifices you make and the perseverance that you show which inspires us all!

Hypersal REALLY does work!



How many of you think about the number of treatments your children must take each day? I think about it and I don't have a child with CF. The time it takes, the taste and the ongoing question of, "Is this REALLY worth it?" A recent study looking at the effect of hypertonic saline in the Journal of

Cystic Fibrosis noted a **PROLONGED** effect on mucociliary clearance in adults and adolescents. (Study was not open to children). The study wanted to show that this affect occurs in the CF lung, as opposed to a healthy lung. It showed that a **SINGLE** dose of hypersal improved

mucociliary clearance for up to 4 hours! Previous studies did not show this effect in a healthy lung. This study supports continued use of hypertonic saline.

Remember, hypersal should **NOT** be mixed with albuterol **BUT** instead albuterol given prior to hypersal.

Recipe Corner: Nacho Skillet Casserole

Ingredients:

- 1 lb. ground beef
- 1 medium onion, chopped
- 1 tsp sugar
- ½ tsp oregano leaves
- ½-1 tsp chili powder
- 1 15.5 oz. can kidney beans, drained
- 1 5 oz. can tomato sauce
- 1 12 oz. can whole kernel corn with red peppers, undrained
- 2 cups slightly broken tortilla chips
- 4 oz. (1 cup) cheddar cheese, shredded

Stir in sugar, oregano, chili powder, kidney beans, tomato sauce and corn.

Simmer 10-20 minutes, stirring occasionally, until most of the moisture is absorbed.

Sprinkle tortilla chips evenly over meat mixture. Top with cheese.

Cover and simmer 2-3 minutes or until cheese is melted.

Serve immediately.

Makes approximately 6 servings.

Nutrition Information:

Serving size: 1 cup
 Calories per serving: 558
 Protein: 32 g

Fat: 31 g
 Carbohydrates: 40 g

This is such a great dish to add even **MORE** calories. Adding guacamole (155 cal/1/2 cup) and sour cream (30 cal/1 TBS) will give extra taste too!

Make this dish your own using a variety of chips or other add-ons like olives, black beans, or hot sauce!

Please let me know if you and your family have a great recipe to share and I can add it to our next Recipe Corner.



Directions:
 In large skillet, brown ground beef and onion. Drain well.

Book donations continue...

We have been extremely blessed over the last several years to be able to provide most of our younger CF patients with books to take home.

The books are a wonderful asset to our clinic as we are well aware our visits are often lengthy as we work to have everyone follow-up as indicated.

Reading will ALWAYS be recommended even as young as infancy. (I read to my babies in utero!) The act of sharing a story and the time we spend holding and reading to our children is a very special kind of bonding.

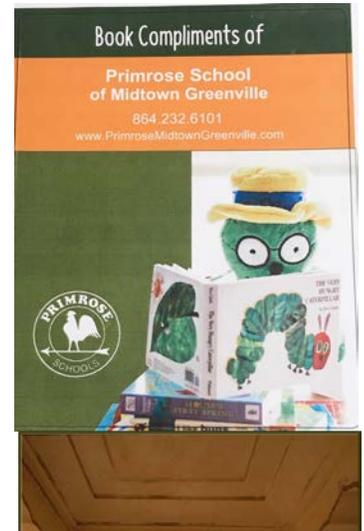
It is not unusual to ask an older child what their favorite childhood memories are, and their recollection includes reading time with family.

All of my teacher colleagues are of course huge proponents of reading and the effects it will have on your child's ability to perceive language and typically improve their writing skills. Reading increases vocabulary which in turn can assist children with word recognition and spelling as they enter school.

We just wanted to share with each of you how very much we appreciate the Primrose School of Midtown Greenville and their willingness to support your children.

Maybe consider a thank you note to their program if you have a minute:

Primrose School of Midtown Greenville
404 Houston St.
Greenville, SC 29601
(864) 370-8118



Book Donations for our CF Patients.

Birthday Bonanza for those celebrating Dec-May!

Austin R.	12/21	Heath R.	12/5	Madison M.	4/26
Kyleigh C.	1/11	Nathan P.	3/22	Cameron D.	4/2
Emily S.	3/29	Brynlee G.	4/14	Adaley C.	5/26
Cameron B.	4/4	Emilee M.	3/12	Konnor W.	4/19
Sylvester H.	12/31	Blaise B.	12/25	Gianna F.	5/16
Brianna H.	03/04	Owen G.	1/20	Khlyha L.	5/13
Heather B.	1/21	Kaiden B.	2/13	Jocelyn S.	4/13
Riley K.	12/18	Adam S.	5/10	Juliatt F.	4/19
Are'ya L.	2/17	Karson C.	3/1		
Jessica D.	2/5	Jacob H.	1/21		
Lily W.	3/15	Raelyn D.	3/28		
Logan T.	1/26	Ava W.	5/12		
Izabella C.	3/13	Jude T.	12/13		

Happy Birthday to all our sweeties over the last several months! Hope you ate or will eat LOTS OF CAKE!



Angels with Roses

Division of Pediatric
Pulmonology
200 Patewood Dr.
Suite A 300
Greenville, SC 29615

Phone:
(864) 454-5530

Fax:
(864) 241-9246

We're on the Web!

See us under:

Children's Hospital at:

www.ghs.org

We're also on FACEBOOK
So check us out and "like us"
www.facebook.com/GHSCchildrens

Our own Miss Lily appeared on the CanineFetchUnit feed of Instagram in her support of our facility dog program.

The facility dog program currently has 4 dogs but we are soon to acquire a fifth. These dogs are specially trained to interact with patients in a way that can allay fears and nerves and allow a child to feel comforted. We are very blessed to have such a wonderful

About Our Organization...

Dr. Steve Snodgrass is the Medical Director of the Division of Pulmonology of Prisma Health Children's Hospital-Upstate and is also our CF Center director. Our purpose:

Inspire health. Serve with compassion. Be the difference.

program. You can find out more information here: at www.ghsgiving.org

Check out Miss Lily and Kalle!



Our CF center is a CORE CF Center accredited by the National CF Foundation. We are continually striving to maintain the high level of care we deliver to our CF families. We have multiple subspecialists involved in our care center. Prisma Health

Children's Hospital-Upstate is here to support our CF families to the best of our abilities.

