Cystic Fibrosis Newsletter

Summer, 2017
Volume 2, Issue 3

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In This Issue

Back to School

As we prepare for a fun summer also think about those back to school forms that are needed for the return for the the 2017-2018 school year.

Orange County Hospital Homebound forms need to be obtained by a parent or guardian at:

3130 Edgewater Drive
Orlando FL, 32804.

All other counties please contact your school guidance counselor for assistance with obtaining forms. Forms can be faxed to 321-843-6659.

All form requests can be made to APHCFCENTER@orlandohealth.com please include the following:

- Your child's name
- School Name and County
- What medications are needed for school such as Enzymes, Inhalers, etc.

IEPs should be requested the first few weeks of school to set a meeting and plan for the entire school year. Resource information on CF and School can be found at www.cff.org or please feel free to contact our center Social Worker Melissa Zeno-Quinn, LCSW at 321-841-6306 for resources, questions, and or concerns.

Vogmask

There has been controversy in the news in regards to Vogmasks utilized for patients with cystic fibrosis in the clinical settings. The Cystic Fibrosis Foundation has published that the Vogmask is NOT approved to protect patients against the spread of pathogens in the clinic and hospital setting. The masks are only intended to protect patients against environmental particles such as dust pollen, and other particles found in air pollution. The CF Foundations' infection prevention and control guideline recommends that "all people with CF wear a surgical mask when in a health care facilitate to reduce the risk of transmission of acquisition of CF pathogens".

In summary: Vogmasks DO NOT provide protection against infection. Please wear a surgical mask when in the clinic or hospital setting.

Research Study Involvement

Vertex is a pharmaceutical company that works on developing new medications for patients with CF to allow for prolonged and improved quality of life. In order to develop new medications, clinical trials are performed to assess the safety and effectiveness of the investigational medicines. Based on the results of the clinic trial, regulation authorities determine if the medication can become available to all patients. Our physicians closely monitor patient safety and effectiveness while on these medications through lab work and clinical visits. If a patient is eligible for the research study, our physicians will reach out to your family to discuss the trials in greater detail. The purpose of the clinical trials is to improve the flow of salt and water into and out of the cells. Here is a list of our current clinical trials at the APH CF Center:

Study name: VX16-440-102

Qualifications: Heterozygous F508del and a minimal function mutation (F508del/MF), 12 years and older

Length of Study: 24 weeks with screening prior to initiation of study and a safety follow up post study

Medication: Patient would take new medication (VX-440) in combination with Tezacaftor (TEZ)and Ivacaftor (IVA), half of patient’s obtain placebo and half obtain VX-440, TEZ, and IVA.

Study name: VX16-661-114

Qualifications: Homozygous F508del, 12 years and older

Length of Study: 56 days with screening prior to initiation of study and a safety follow up post study

Medication: Tezacaftor (VX-661) and Ivacaftor

Study name: VX16-152-102

Qualifications: Heterozygous F508del and minimal function CFTR, 18 years and older

Length of Study: 2 part study, initially 2 weeks followed by 8 weeks

Medication: VX-152 in triple combination with Tezacaftor and Ivacaftor

Study name: VX15-371-101

Qualifications: Homozygous F508del currently being treated with Orkambi, 12 years and older

Length of Study: 2 trials of 28 days

Medication: VX-371, Lumacaftor, and Ivacaftor

Medication Update

The FDA has approved expanding guidelines for some Cystic Fibrosis medications. Kalydeco (Ivacaftor) is now offered to patients 2 years and older, and Orkambi (Lumacaftor and Ivacaftor combo) is now offered for patients 6 years and older.

To be eligible for Kalydeco you must have one of the following CF variants: G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N, or S549R. The new CF variants that qualify for this medication are: A1067T, A455E, D110E, D110H, D1152H, D1270N, D579G, E193K, E56K, F1052V, F1074L, G1069R, K1060T, L206W, P67L, R1070Q, R1070W, R117C, R347H, R352Q, R74W, S945L, S977F.

To be eligible for Orkambi you must have two copies of the F508del mutation.

For more information, please talk to your provider at your next Cystic Fibrosis visit.

Vitamins

We have been hearing from families that the pharmacies have been dispensing different vitamins than what have been prescribed. We want to make sure everyone is getting what they are supposed to. So, please bring in your vitamins or take a picture of them and show to Allison Brinkley at your next clinic visit. Another option is to e-mail her at Allison.Brinkley@orlandohealth.com.

We just received an update that MVW vitamins should be available in all retail pharmacies now, so this should make getting these easier. AquADEK vitamins are no longer in production, so if you are currently taking these and needing a refill soon, please notify Allison about that as well.

Taking the right vitamins is an important part of staying healthy with Cystic Fibrosis. Enzymes help digest food within the digestive system so nutrients can be absorbed. In Cystic Fibrosis, thick mucus can block the ducts and the body's enzymes cannot reach the food. Without enzymes the calories and nutrients in the food you eat flushes away with each bowel movement. It is important to keep enzymes with you so you do not skip them, but keep in mind that capsules need to be kept in a cool, dry place.

Labs

Summer is a great time of year to do annual labs. This way, less school is missed. Annual labs assist the Cystic Fibrosis team to determine if you have sufficient nutrient intake, checks the function of your liver, and kidneys as well. In addition, it is recommended that patient's over the age of 8 obtain oral glucose tolerate testing (OGTT) annually. The OGTT measures the body's ability to use a type of sugar (glucose) which provides the body with energy. This is a test to screen for diabetes. Patients with cystic fibrosis are at increased risk for a specific kind of diabetes called Cystic Fibrosis Related Diabetes; that is why this screening is so important. Annual Chest x-ray is used to monitor your lung health, and look for any changes or progression of cystic fibrosis disease, as well as monitor for infection. Patients over the age of 18 obtain a dual-energy X-ray Absorptiometry (DXA). The DXA is utilized to determine the density of the bones. Decreased bone density is an indicator of poor vitamin and mineral absorption. It is important to bring orders for bloodwork to the lab, even if test will be done at Arnold Palmer Hospital. Please call our office at (321)841-6317 if you do not have the lab or radiology orders needed.

We also wanted to inform families that we are no longer going to call results of normal lab work or if the sputum culture is normal or consists of the patient's usual CF bacteria, unless a new or different treatment is indicated. If there are new symptoms after a culture, please call the office to obtain results and further information and recommendations. Accessing the Orlando Health Patient Portal is an option. This was mentioned in the previous newsletter.

An Introduction to Cystic Fibrosis for

The CF Foundation has announced a new DVD titled An Introduction to Cystic Fibrosis for Patient and Families, 6th Edition. Clinicians, people with CF and their families discuss the basics of CF and other pediatric and adult CF related issues. The DVD can be viewed on CFF's YouTube Channel or ordered from the CF Foundation. The content complements the book of the same title, which is available in both English and Spanish on CFF.org.

To order hard copies of the DVD (currently only available in English) or the book (available in English and Spanish), email resources@cff.org with the title of the resource, the quantity, your name, street address (including room/building number), and phone number. If you need assistance with ordering the book or DVD, please let us know during your next office visit and we can assist you through the process.

Ranch Dip

Mix following ingredients in a bowl and serve

- *1 package regular cream cheese*
- *½ cup sour cream*
- *½ cup mayonnaise*
- *1 package ranch dressing mix*
- *2 Tbsp parmesan cheese*

Airway Clearance and Aerosol Therapy Video Contest

We wanted to say thank you to all of our families who participated in the FUNLLC quality improvement project last year. We learned a lot about which airway clearance and aerosol therapy modalities our center population is using.

We were also able to identify some confusion around nebulizer device disinfection. Remember, do NOT use tap water as a final rinse for your nebulizer.

We are going to continue the airway clearance and aerosol therapy questionnaire on a yearly basis in an effort to identify educational opportunities.

We are continuing to ask patients to bring in their airway clearance and aerosol compressor once a year for a check-up to make sure it is operating properly.

If you are satisfied with the operation of your equipment, instead of bringing in your respiratory equipment you may perform a short video documentary on a mobile device.

Show us what you do during a routine airway clearance and aerosol therapy session including:

1. Name and order of medications
2. How long it takes to do each treatment
3. Vest settings include: pressure, frequency, duration of each setting
4. Aerosol nebulizer device cleaning and disinfection

Winners will be eligible for A NEW CAR!!!!!!!!!!!!! Courtesy of Dr. Weatherly and Faverio. Just kidding, but there will be a prize for the best video.

CF R.I.S.E.

For our patients 15 and older that are enrolled in CF R.I.S.E., our transition program, we ask you to start the next module when you arrive at your next clinical visit. If you do not have a mobile device, we can provide an I-pad mini, to use while in clinic, to assist with this.

If you are 15 years or older and have not registered for CF R.I.S.E. yet, please be ready to do so at your next clinic visit. We'll explain then.

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