

Flu Season

Dear Families,

Flu season is once again upon us. This letter is a gentle reminder of the recommendations regarding vaccines to keep your child as healthy as possible throughout the upcoming flu season. The Wisconsin Immunization Registry (WIR) is an excellent resource to check your child's immunization status. It identifies all vaccines received in the state of WI and what vaccines your child is in need of. The link to the website is: <https://www.dhswir.org/PR/clientSearch.do?language=en>. If your child receives immunizations in a state other than WI, please check with your state's immunization registry.

Listed below are the vaccines that are recommended for your child with cystic fibrosis:

- Annual influenza vaccine (flu shot). This vaccine is recommended for all patients seen at AFCH Pulmonary/CF Specialty clinic each year starting at the age of 6 months. The vaccine can be obtained at your PCP office, AFCH Pulmonary/CF Specialty clinic or in your community. The vaccine is listed as "influenza" in the WIR system.
- Pneumovax vaccine (PPSV 23). This vaccine is recommended for all patients seen at AFCH Pulmonary/CF Specialty clinic starting at the age of 2. If your child has already received this vaccine they will not need a booster until the age of 65. The vaccine is listed as "pneumo-poly" in the WIR system. If your child has never received this vaccine they will need to have proof of first receiving 2 doses of Prevnar 13. This vaccine is listed as "pneumococcal" in the WIR system. It is then further described as Prevnar or Prevnar 13. This is a vaccine that most all children received in the first 2 years of life. It is typically a 4 shot series. If your child only received Prevnar, and not Prevnar 13, they will require 2 doses of Prevnar 13 prior to receiving the Pneumovax vaccine. Prevnar 13 can be obtained at your PCP office or at AFCH Pulmonary/CF Specialty clinic. However your child must receive their last dose of Prevnar 13 8 weeks prior to receiving the Pneumovax vaccine.

Therefore you may want to inquire in advance as to whether your child has received at least 2 doses of Prevnar 13 due to the 8 week waiting period before the Pneumovax vaccine can be administered. This may be something you may want to explore as soon as possible to prevent any further delay in immunizations. If your child is in need of 2 doses of Prevnar 13 prior to receiving the Pneumovax vaccine, this can be obtained at your PCP office.

If you have any questions or concerns regarding the information listed above, please do not hesitate to contact the AFCH Pulmonary/CF Specialty clinic or your PCP office.

Sincerely,

*AFCH Pulmonary/
CF Specialty Clinic Staff*

New policy for Vest Therapy

Beginning June 1, 2017, American Family Children Hospital and UW Hospital and Clinics have changed their practice for delivering vest therapy. If a patient who uses vest therapy in their home requires hospitalization, Respiratory Therapy will only be able to provide vest therapy if the patient brings in their personal vest unit (vest, machine and hoses) from home. If a patient is unable to bring in their personal vest unit from home they will receive manual CPT until the vest can be brought to the hospital.

Bringing in the personal vest unit is for the safety of each patient and will also allow the Respiratory Therapist to ensure the vest is fitting properly and answer any questions about vest therapy. If you have any questions, please feel free to contact your Pulmonary Team.

Regards,

Respiratory Therapy

Introducing Dr. Carrie Barker

We are pleased to welcome Dr. Carrie Barker to our pediatric pulmonology division. Dr. Barker earned undergraduate degrees in comparative literature and music (violin performance) at Northwestern University in Evanston, Illinois. She earned her medical degree at the University of Iowa in Iowa City and was a pediatric resident at the University of Wisconsin from 2005 to 2008. She returned to Iowa to complete a pediatric pulmonology fellowship in 2011. She then was Assistant Professor of Pediatric Pulmonology at the University of Nebraska Medical Center in Omaha from 2011 to 2015. She returned to Iowa City to be on faculty at the University of Iowa.

We are so very pleased that Dr. Barker is returning to her Badger roots and joining our pediatric pulmonology division.



Tips for Traveling Abroad with Cystic Fibrosis

Traveling abroad is an exciting adventure and we would like to help you prepare for your trip to make sure you can spend more time enjoying yourself. The key to a successful trip is all in the planning. We've compiled a list of travel tips offered by the Cystic Fibrosis Foundation and tips from patients with CF who have traveled abroad for you to consider.

1. First things first, see your CF team and notify them of your travel plans several months in advance. Your CF team can help you with a variety of travel related needs including: determining if you're healthy enough to fly, arrange for any tests or treatments you may need ahead of time, prescribe antibiotics for you to take along, help you to understand if you'll need any special travel accommodations, give you necessary vaccinations, put together a health history to take with you, and complete necessary travel documents for travelling with medications or equipment.
2. Oxygen: **For our second tip, please read on, even if you don't use oxygen on a regular basis.** Most people with CF can fly without oxygen, but some will require on flight oxygen due to a change in altitude when flying. Your CF team can conduct a high altitude test prior to your trip to let you know if you'll need oxygen. Airlines have different policies for oxygen use, so check with each airline you're flying with for their requirements. Please note that oxygen canisters are never allowed on flights, only concentrators. Here are some points to consider:
 - a. Do you need a prescription for the oxygen?
 - b. Are any additional airline forms required to use oxygen in flight?
 - c. Are portable concentrators allowed on flight? If so, which ones?
 - d. Is in flight oxygen offered? Is there a charge for this?
 - e. How far in advance do you have to notify the airline of need for oxygen?
 - f. Will your insurance cover special oxygen equipment during travel?
3. Medications: You'll want to be sure that you have all of the medications that you need, and an extra supply in case of any delays in your travel.
 - a. Avoid losing/damaging meds by packing your medications as a carry on, never pack them in checked luggage. Your medications don't count toward carry-on bag limits.
 - b. Written prescriptions may not be valid in some countries. Your pharmacist can help you to order extra meds for your trip. You may be able to ship meds to your destination.

Back to School!

It is the time of year that children are heading back to school. Now is the time to discuss with the school any paperwork or forms that need to be completed by your child's medical provider. This includes school medication administration forms for enzymes, albuterol, or any other medications your child takes during the school day. If you are seeing your provider prior to school starting, please ask for a school medication form to be completed. If you will not be seen in clinic prior to school starting, please contact our office at 608-263-6420 option 2, and request that a form be completed and faxed to the school.

If your child is newly heading off to school, please ask us about our school resource folders. These folders provide useful information for schools and teacher, including information regarding nutrition, enzymes, general CF information, and the importance access to sports drinks during gym and other physical activity. It is important to have a conversation with school staff at the beginning of the school year to discuss the above information available in the folder, so any questions can be addressed right away.

If any questions arise, please contact our office at 608-263-6420 option 2 to discuss further.

College

Some of your children may also be heading off to college this fall. What an exciting time! We have taken advice from patients and families who have already experienced college as well as advice from the CF Center in Chicago, IL. We have some helpful hints that you may want to consider:

- See your current CF care team about a month before school starts to review your current health status, intervention plan and make sure all prescriptions are up to date.
- Identify the location and phone number for the student health services and a place where you can be seen if you become ill or if there is an emergency. Check with your insurance company about out of network benefits.
- Contact the college's office of disability to let them know that you have CF. You may never need their services, but in the event of an illness or hospitalization, they can help guide you and interface with professors or create and individualized education plan (IEP). They may also allow you to register early to ensure you have the most flexible schedule to accommodate your daily treatments.
- Think about who needs to know about your CF to help if you become ill. Many students share this information with their resident advisor, the student health services, the office of disabilities and a friend or roommate to help you in an emergency.

- If possible, request a private room. If not, we recommend that you consider telling your roommate that you have CF and need to take medication and perform daily treatments.
 - If possible, request a room with air-conditioning.
 - Make sure there is access to a refrigerator to store medication and extra food.
 - A microwave is helpful to sterilize airway treatment supplies. If that is not possible, create a plan of how you will sterilize and dry your supplies.
 - Does the electrical wiring in the room meet the needs of your equipment? You may want to consider surge protectors for your air-conditioner, refrigerator and airway clearance equipment.
 - Make sure you have enough medication with you or access to your pharmacy for refills. Identify a pharmacy near campus that can fill medication and provide supplies. When traveling to and from home, remember to bring your medication and supplies with you.
 - Always have the number of the CF center with you in case you have questions or concerns. Now that you are away from home, you can call us directly and speak with our staff.
 - Our CF center can work with you to schedule clinic visits during breaks and days off to continue being seen in clinic at least four times a year.
 - If you are going to school out of state, we can help you identify a CF center that is close to you. We encourage you to check with your insurance company to find out what services will be covered... you don't want to be stuck with a big bill.
 - Try to balance your CF care with your school and social life. We know what college is like, but want you to remember to get plenty of rest and keep up with your CF cares!
 - Remember, college scholarships are available for people with CF. The following website can help navigate CF scholarships. <http://cfscholarships.com>
- c. Some meds are not allowed in some countries. Check with the U.S. Embassy in the country you're travelling to for more information.
 - d. Pack your meds in their original containers to show customs officials.
 - e. If your meds do not fit in a quart size bag, TSA regulations state you must declare them to a TSA officer.
4. Medical Equipment: Be prepared to use your equipment during your travel. Consider:
 - a. To avoid damage/lost equipment, carry your equipment on, do not check it. Your medical equipment does not count towards your carry on limitations, but it must fit in the overhead or under the seat.
 - b. Charge equipment before leaving.
 - c. Does your equipment have the right voltage for the country you're visiting? You may need a converter.
 - d. What type of electrical adaptors will you need to plug equipment in?
 - e. Request to pre-board the plane to give you extra time/space to stow your bags.
 - f. Check with your physician to see if doing chest CPT or using a device such as Acapella or Flutter would be ok instead of using your vest.
 - g. Can you buy sterilizing liquids in the country you're visiting instead of packing them?
 5. Documentation: You will need certain documentation when travelling out of the country. Check with your airline to find out what you'll need. Common forms include:
 - a. Customs certificate signed by your doctor to explain why you're travelling with a large supply of medications, syringes, and equipment. It should include your name, date of birth, list of meds, quantities, and how often taken as well as list of medical supplies and reason for bringing a large quantity of liquids.
 - b. Medical certificate: Written statement saying you are healthy enough to fly.
 - c. Health summary: Name, date of birth, CF center contact information, allergens, description of CF and health status, meds, and therapies. You can give this to doctors if you have a medical emergency.

6. Contact information to bring with you in case of emergency:
 - a. Your CF Center phone number.
 - b. Your insurance company phone number. (Also bring cards with you)
 - c. Phone number for the U.S. Embassy in the country you're visiting. Call the following numbers to locate the nearest Embassy: From the U.S. or Canada: 1-888-407-4747, from overseas: +1-202-501-4444. For information on making international phone calls or for help locating country codes, visit the Federal Communications Commission's website at: <https://www.fcc.gov/consumers/guides/international-long-distance-calling-made-simple-tip-sheet>
 - d. Location of nearest CF center to where you're visiting. Check this website for international CF Center locations: <https://www.cfww.org/global-associations>. Don't wait to seek medical care if you're having health concerns.
 - e. Bring a prepaid phone card with you in case your cell phone doesn't work.
 - f. Your emergency contact's information
7. Insurance: Make sure you're covered.
 - a. Call your health insurance company to check international benefits.
 - b. Consider purchasing travel insurance to cover emergency expenses.
8. Destination: Location, location, location:
 - a. Consider traveling to a county that offers CF specialty care.
 - b. Consider health risks of traveling to certain destinations. For example- warm, wet areas, such as areas affected by monsoon season can harbor more harmful bacteria.

While this is quite a bit of information, we hope that you will find it helpful in planning a hassle free trip. Have fun, enjoy, and Bon Voyage!

Research News

Tezacaftor/Ivacaftor

On March 28, 2017, Vertex Pharmaceuticals, Inc. released the results of two studies of the combination drug tezacaftor/ivacaftor. The first study, called EVOLVE, evaluated the combination treatment over 24 weeks in people who have two copies of the F508del mutation. Subjects who received active drug had an improvement of FEV1 of 4% from baseline compared to subjects who received placebo. The second study, called EXPAND, evaluated the combination treatment over 8 weeks in people with CF who have one F508del mutation and a second mutation that results in residual CFTR function. (Eligible mutations for the study included: 2789+5G->A, D110E, R352Q, 3849+10kbC->T, D110H, A455E, 3272-26A->G, R117C, D579G, R1070W, 711+3A->G, E193K, S945L, F1074L, E56K, L206W, S977F, D1152H, P67L, F1052V, D1270N, R74W, R347H, K1060T and E831X.) The improvement in FEV1 was 6.8 percentage points from baseline compared to placebo for those receiving the tezacaftor/ivacaftor combination.

Based on these results, Vertex announced plans to submit a New Drug Application to the FDA in the fall 2017 for review and potential approval of the combination treatment for people with two copies of the F508del mutation ages 12 years and older as well as for individuals with one copy of the F508del mutation and a residual function mutation, also for ages 12 years and older.

Triple combination therapy with next generation corrector drugs

On July 18, 2017, Vertex announced positive results from phase 1 and phase 2 studies of three different triple combination regimens in people with CF who have one F508del mutation and one minimal function mutation. (A minimal function mutation are those mutations that result in no CFTR protein. Many of these are nonsense mutations, also known as “stop” mutations or “x” mutations [x is at the end of the name of the mutation, such as R553X]). You can read the full press release from Vertex at:

http://files.shareholder.com/downloads/VRTX/4877987110x0x949882/D22E7B23-C446-4A80-A017-C7ED96E161C2/VRTX_News_2017_7_18_General_Business.pdf

A summary of the results of the studies are:

- In the phase 2 study of VX-440 in combination with tezacaftor and ivacaftor, there were 18 subjects with an F508del and minimal function mutation who received study drug. Those subjects who received the 600 mg dose of VX-440 had a 12 percent increase in FEV1. There were 20 subjects with two copies of the F508del mutation who received the triple combination study drug. These subjects had a 9.5 percent increase in FEV1.

- In the phase 2 study of VX-152 in combination with tezacaftor and ivacaftor, there were 10 subjects with and F508del and minimal function mutation who received study drug. The subjects who received the 200 mg dose of VX-152 had an improvement in FEV1 of 9.7 percent. There were 10 subjects with two copies of the F508 who received the triple combination study drug. These subjects had a 7.3 percent increase in FEV1.
- In the phase 1 study of VX-659, there were 9 subjects with CF who had an F508del and minimal function mutation who received study drug. These subjects had 9.6 percent increase in FEV1.

Vertex has a fourth next generation corrector, VX-445, which is also in clinical trials.

Our CF Center will be participating in a phase 3 study of one of these next generation correctors. How can you be involved in a clinical trial?

- Ask your provider at your next clinic visit about clinical trial participation.
- Ask to speak to one of our research coordinators.
- Explore the clinical trial finder tool at the CF Foundation’s website: <https://www.cff.org/Trials/finder>
- Watch the listings of our clinical trials at our website: <http://www.uwhealthkids.org/cf-center/cystic-fibrosis-cf-research-studies/45153>

Vogmask... Did You Know?

- The Vogmask is not classified as a mask to prevent disease.
- The average size of a virus particle is 0.01 to 0.5 microns.
- The average size of a bacterial particle is 0.3 to 10 microns.
- The intended use of the Vogmask is to protect from inhalation of airborne particles over 0.3 microns.
 - Helps protect from non-oil airborne particles such as dust, pollen, shavings, and particles found in air pollution.
 - It does not eliminate the risk of illness, diseases, or infection.
- The maintenance instructions are to gently rinse outer and inner layer, add a drop of liquid soap (like dishwashing soap), rinse again, and hang dry. The filtering efficiency of the middle layer nano fiber will decline if undetected damage (like a tear in the filtering textile) in the middle layer filtering layer occurs. This is why hand washing is recommended.



CF Clinic Recommendations

- Vogmasks are great for airborne pollutants such as smog, smoke, pollen, or dust.
- Vogmasks do not protect against viruses.
- Vogmasks do not protect against all bacteria.
- Vogmasks cannot be disinfected or washed to rid of bacteria.
- Please wear yellow surgical masks to clinic and on the floor.

Our goal is to keep you healthy and free of germs!

* Document originated from Pediatric CF Center in Dallas Texas.

Relative Size Chart of Common Air Contaminants (Shown in Micrometers)

