ARIZONA PEDIATRIC PULMONARY CENTER

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NEWSLETTER

WHAT'S NEW WITH MEDICATIONS?

An Update on Kalydeco® (ivacaftor)

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What is Kalydeco® (ivacaftor) and how does it work?
Kalydeco® (ivacaftor) is an oral medication that can be used for specific patients with cystic fibrosis. It works by improving the transport of salt and water across membranes which helps to hydrate and clear mucus in the airways.

Which CF mutation(s) is Kalydeco® (ivacaftor) approved for use?
G551D, G1244E, G1349D, G178R, G551S, S1251N, S1255P, S549N, S549NR, R117H

What is NEW about Kalydeco® (ivacaftor)?
Previously, Kalydeco® (ivacaftor) was only approved for patents 6 years of age and older with specific mutations. It was only available as a 150 mg tablet given orally every 12 hours. On March 18, 2015, the food and drug administration (FDA) approved the use of Kalydeco® (ivacaftor) for children 2 to 5 years of age who have at least one of the above listed mutations. Now 50 mg and 75 mg oral granules are now available for young children. Dosing is weight-based as follows:

Children less than 14 kg (< 31 lbs): One 50 mg packet mixed with 5 mL of soft food or liquid given every 12 hours by mouth with fat-containing food.

Children at least 14 kg (≥31 lbs): One 75 mg packet mixed with 5 mL of soft food or liquid given every 12 hours by mouth with fat-containing food.

If I am started on Kalydeco (ivacaftor), what can I expect as far as possible side effects and needed monitoring by the CF Team?
Kalydeco® (ivacaftor), is broken down by the liver to an inactive drug. One of the main side effects that may be seen is an increase in liver enzymes. Prior to starting the medication, baseline tests will be performed to check your liver enzymes. After starting the therapy, your liver enzymes will be monitored every 3 months for the first year and then yearly if they are normal. Other side effects that can be seen with Kalydeco® (Ivacaftor) include headache, sore throat, head cold, diarrhea, nausea, and dizziness. If after starting Kalydeco® (ivacaftor) you notice any of these side effects, please inform your doctor. Kalydeco® (ivacaftor) may interact with other medications. It is important you talk to your doctor or pharmacist before starting any new medications.

UPDATES FROM OUR TRAINEES

My name is Mailin Rivera, and I am a graduate social work student and a University of Arizona Pediatric Pulmonary Center (PPC) social work trainee. I recently distributed an assessment to several cystic fibrosis (CF) patients covered by Medicaid and Medicare, looking at their nutritional needs because appropriate nutrition is associated with better CF health outcomes. Most patients diagnosed with CF need to consume high fat/high calorie diets in order to maintain a healthy weight, which positively impacts their lung function and overall health.

I chose to focus on individuals covered by Medicaid and Medicare to assess whether the resources provided were meeting both the nutritional and financial needs of the patients. The results showed many families, regardless of whether they were receiving assistance from the Arizona Nutrition Assistance Program, were not able to provide the high calorie/high fat foods recommended to meet their CF nutritional needs due to finances. Several patients mentioned how beneficial it would be to have a food bank that would cater to their CF nutritional needs. The results also showed that some patients have turned to food pantries/food banks in order to supplement with foods that they cannot afford to purchase. Although these food pantries/food banks were helpful in providing more food in their homes, many of the foods specifically needed as a patient with CF were not being provided.

Given these results, I am working on collaborating with local food banks, asking them to consider providing high fat/high calorie foods specifically for patients diagnosed with CF, in order to help promote healthier growth and development of children and improve health outcomes for all CF children and adults. My hope is to have at least one food bank collaborate to meet these specific financial and nutritional needs. Please feel free to contact me with any questions.

My name is Sofhia Ytuarte, and I am a Nurse Practitioner intern. During my internship with the University of Arizona Pediatric Pulmonary Center, a need arose for cystic fibrosis (CF) education at a k through 8 primary school in Benson, AZ. Based on the need, I developed a PowerPoint presentation to educate students and faculty on CF. Besides the PowerPoint presentation, visual aids were used, demonstration of the vest and breathing through a straw were done. Faculty were also given educational handouts on CF. Due to the need for education on CF in schools, I am developing a curriculum to be used by schools in the future. To develop the curriculum, I would like to know what you think should be included. If you have a child or children with CF, what is the most important thing(s) that you would want to see presented to students and teachers? Please send comments to nonnie521@msn.com.

ONLINE RESOURCES

www.uappc.peds.arizona.edu
The Pediatric Pulmonary Center's website

www.CysticLife.org
A social network for the cystic fibrosis community

CONTACT INFORMATION

Medical Questions
(520)694-5132

Newsletter Questions
(520)626-2962

SOME HELPFUL REMINDERS ABOUT ALL YOUR MEDICATIONS

• **Maintain a medication list with all your medications names, doses, and how often or when you take them.**
- Take it with you to the pharmacy to remind your pharmacists of what you are currently taking.
- Take it to clinic to help your CF team assess your current treatment.
- Keep it with you in case of medical emergencies.
- You can ask the CF clinical pharmacist for a handy medication list form or smartphone app suggestion to create your list.

• **Try to keep all your prescriptions at one pharmacy chain.**
- With all your medication history at one location, the pharmacist can best assess for possible drug interactions.
- Use auto-refill programs at your pharmacy to keep your medication supply steady.
- If you cannot consolidate your medications to one pharmacy, try to have only one community pharmacy (e.g., Walgreens) and one specialty pharmacy (e.g., Optum) for your medication needs.

• **Ask your pharmacist before starting any new medications, both prescription and over-the-counter, to prevent drug interactions.**

• **Ask your CF clinical pharmacist any questions you may have about your medications – They're happy to help!**

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