A GUIDE TO TAKING KALYDECO® (ivacaftor) Tablets
For people age 6 years and older and their caregivers

Use of KALYDECO in children aged 1 month to less than 6 months born from a pregnancy lasting (gestational age) less than 37 weeks has not been evaluated.

What is KALYDECO?
KALYDECO is a prescription medicine used for the treatment of cystic fibrosis (CF) in people aged 1 month and older who have at least one mutation in the cystic fibrosis transmembrane conductance regulator (CFTR) gene that is responsive to KALYDECO.

Talk to your doctor to learn if you have an indicated CF gene mutation.

It is not known if KALYDECO is safe and effective in children under 1 month of age.

IMPORTANT SAFETY INFORMATION
Before taking KALYDECO, tell your doctor about all of your medical conditions, including if you:

- have liver or kidney problems
- are allergic to KALYDECO or any ingredients in KALYDECO. See the Patient Information for a list of ingredients

Please see Important Safety Information and full Prescribing Information, including Patient Information.
Before taking KALYDECO®, tell your doctor about all of your medical conditions, including if you:

- have liver or kidney problems
- are allergic to KALYDECO or any ingredients in KALYDECO. See the Patient Information for a list of ingredients
- are pregnant or plan to become pregnant. It is not known if KALYDECO will harm your unborn baby. You and your doctor should decide if you will take KALYDECO while you are pregnant
- are breastfeeding or planning to breastfeed. It is not known if KALYDECO passes into your breast milk. You and your doctor should decide if you will take KALYDECO while you are breastfeeding

Tell your doctor about all the medicines you take, including prescription and over-the-counter medicines, vitamins, and herbal supplements.

KALYDECO may affect the way other medicines work, and other medicines may affect how KALYDECO works. Ask your doctor or pharmacist for a list of these medicines if you are not sure.

Especially tell your doctor if you take:

- the antibiotics rifampin (RIFAMATE®, RIFATER®) or rifabutin (MYCOBUTIN®)
- seizure medicines such as phenobarbital, carbamazepine (TEGRETOL®, CARBATROL®, EQUETRO®), or phenytoin (DILANTIN®, PHENYTEK®)
- St. John’s wort
- antifungal medicines such as ketoconazole, itraconazole (SPORANOX®), posaconazole (NOXAFIL®), voriconazole (VFEND®), or fluconazole (DIFLUCAN®)
- antibiotics such as telithromycin, clarithromycin (BIAXIN®), or erythromycin (ERY-TAB®)

What should I avoid while taking KALYDECO?

- KALYDECO can cause dizziness in some people who take it. If you experience dizziness, do not drive or operate machines until symptoms improve
- Avoid food or drink containing grapefruit while you are taking KALYDECO

Please see Important Safety Information and full Prescribing Information, including Patient Information.
What are the possible side effects of KALYDECO®?

KALYDECO can cause serious side effects, including:

- **High liver enzymes in the blood**, which have happened in people receiving KALYDECO. Your doctor will do blood tests to check your liver:
  - before you start KALYDECO
  - every 3 months during your first year of taking KALYDECO
  - every year while you are taking KALYDECO

For people who have had high liver enzymes in the past, your doctor may do blood tests to check the liver more often.

Call your doctor right away if you have any of the following symptoms of liver problems:

- pain or discomfort in the upper right stomach (abdominal) area
- yellowing of your skin or the white part of your eyes
- loss of appetite
- nausea or vomiting
- dark, amber-colored urine

- **Serious allergic reactions** have happened to people who are treated with KALYDECO. Call your healthcare provider or go to the emergency room right away if you have symptoms of an allergic reaction. Symptoms of an allergic reaction may include:

  - rash or hives
  - tightness of the chest or throat or difficulty breathing
  - light-headedness or dizziness

- **Abnormality of the eye lens (cataract)**, which has happened in some children and adolescents receiving KALYDECO. Your doctor should perform eye examinations before and during treatment with KALYDECO to look for cataracts.

**The most common side effects of KALYDECO include:**

- headache
- upper respiratory tract infection (common cold), including:
  - sore throat
  - nasal or sinus congestion
  - runny nose
- stomach (abdominal) pain
- diarrhea
- rash
- nausea
- dizziness

Tell your doctor if you have any side effect that bothers you or that does not go away. These are not all the possible side effects of KALYDECO. For more information, ask your doctor or pharmacist.

Use of KALYDECO in children aged 1 month to less than 6 months born from a pregnancy lasting (gestational age) less than 37 weeks has not been evaluated.

Call your doctor for medical advice about side effects. You may report side effects to FDA at 1-800-FDA-1088.
Taking KALYDECO® (ivacaftor) tablets

Take 1 KALYDECO tablet (150 mg) every 12 hours with fat-containing food or as directed by your healthcare provider

- Taking KALYDECO every 12 hours helps keep enough medicine in the body
- Remember, taking KALYDECO every 12 hours is not the same as taking it twice a day
- Your healthcare provider may give you different instructions on how much KALYDECO to take and when to take it
- Keep in mind that every dose matters, so it’s important to make sure that you take KALYDECO exactly as your doctor tells you to take it

Continue to take KALYDECO plus all your other CF therapies as directed by your healthcare provider

- Be sure to talk to your healthcare provider before starting, changing, or stopping any CF therapies or other medications, including prescription and non-prescription medicines, vitamins, and herbal supplements

Please see Important Safety Information and full Prescribing Information, including Patient Information.
Taking KALYDECO® (ivacaftor) tablets (continued)

What to do if you miss a dose of KALYDECO

≤6 hrs

• If you miss a dose of KALYDECO and it is **within 6 hours** of when you usually take it, take the dose of KALYDECO as prescribed with fat-containing food as soon as possible

≥6 hrs

• If you miss a dose and it is **more than 6 hours** after the time you usually take it, **skip that dose only** and take the next dose with fat-containing food when you usually take it. Do **not** take 2 doses at the same time to make up for your missed dose

Avoid foods and drinks that contain grapefruit—these may affect the amount of KALYDECO in the body.

You need to take KALYDECO with foods that contain fat. Learn more on the next page. ▶

Please see Important Safety Information and full Prescribing Information, including Patient Information.
Always take KALYDECO with foods that contain fat

KALYDECO should always be taken with a fat-containing meal or snack because the fat helps the body absorb the medicine better. KALYDECO can be taken right before or right after a fat-containing meal or snack.

You may have heard that not all fats are created equal. Here are some basic facts on fat:

- There are a variety of fats to choose from
- Some sources of fat may be better for you than others

Talk to your healthcare provider or dietitian about a fat-containing diet that is right for you.

Choosing fats for your diet

Get to know the two main kinds of fat

**Unsaturated fats**
- Vegetable oils, such as olive, canola, peanut, soybean, corn, and safflower oils
- Avocados
- Many nuts and seeds, such as walnuts and sunflower seeds
- Fatty fish, such as mackerel and herring

**Saturated fats**
- Fatty beef, chicken with skin, and bacon
- Palm oil and coconut oil

Like peanut butter?
It likes you, too! Peanut butter has some saturated fat, but approximately 80% is unsaturated. And it tastes great with a banana.
Consider these meals and snacks when choosing fat-containing foods to take with KALYDECO® (ivacaftor)

Some of these meals may also provide vitamins, minerals, and other nutrients. Remember to talk to your healthcare provider or dietitian about a fat-containing diet that is right for you before making any changes to your diet.

Here are suggestions for fat-containing foods:

<table>
<thead>
<tr>
<th>Morning meals</th>
<th>Evening meals</th>
<th>Snacks</th>
</tr>
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<tbody>
<tr>
<td>• Bagel with peanut butter</td>
<td>• Omelet with ham and vegetables</td>
<td>• Hard-boiled eggs</td>
</tr>
<tr>
<td>• Sausage, egg, and cheese biscuit*</td>
<td>• Cheese pizza*</td>
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<tr>
<td></td>
<td>• Cheddar cheeseburger*</td>
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<tr>
<td></td>
<td>• Macaroni and cheese*</td>
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*Be sure the cheeses are made with whole milk.

If you have any questions about healthy fat-containing foods, talk to your healthcare provider.

Get delicious recipes and food ideas on Everyday-CF.com.
**Q:** How should I take KALYDECO tablets?

Take KALYDECO tablets exactly as your doctor tells you to take them. Most people take 1 tablet by mouth every 12 hours with fat-containing food. Your dose may be different. Examples of fat-containing foods include eggs, butter, peanut butter, cheese pizza, and whole-milk dairy products.

**Q:** What are some ways to remember to take KALYDECO every 12 hours?

Here are some ideas:
- Set an alarm on your clock, watch, cell phone, or computer
- Take KALYDECO when you are doing something you do at the same time every day
- Choose times that are convenient every day of the week, even on the weekend

**Q:** Why must I take KALYDECO with fat-containing food?

Your body will absorb KALYDECO better when you take it with fat-containing food.

**Q:** What should I avoid while taking KALYDECO?

KALYDECO can cause dizziness in some people who take it. If you experience dizziness, do not drive or operate machines until symptoms improve. You should avoid foods and drinks containing grapefruit while you are taking KALYDECO.

**Q:** Do I still need to take my other CF therapies with KALYDECO?

Take KALYDECO and your other CF therapies exactly how your doctor tells you. Talk to your healthcare provider before starting, changing, or stopping any of your CF therapies.

**Q:** Can I drink alcohol while taking KALYDECO?

It is not known how alcohol interacts with KALYDECO. Talk to your healthcare provider about consuming alcohol while taking KALYDECO.

**Q:** What if my dose of KALYDECO is reduced or stopped?

Your healthcare provider may reduce or stop your dose of KALYDECO for medical reasons. Take KALYDECO exactly as your doctor tells you to take it.

Please see Important Safety Information and full Prescribing Information, including Patient Information.
We’re here to help you get there
Wherever life with cystic fibrosis (CF) takes you, Vertex GPS™: Guidance & Patient Support is here to help. We offer personalized, one-on-one support to help you start and stay on track with treatment. Once you’re enrolled, you’ll be assigned a dedicated Support Specialist who will be with you every step of the way.

Here are just some of the ways your Support Specialist can help:

**Get you started on treatment** by verifying your coverage and out-of-pocket costs with your insurance company. They’ll also connect with your healthcare provider to discuss any requirements or questions your insurance company may have while determining coverage.

**Help you explore financial assistance options,** regardless of your insurance coverage. And if you have commercial insurance, the Vertex GPS Co-pay Assistance Program may be able to lower your co-pay to as little as $0 per fill.*

*Eligibility restrictions and limitations apply. Annual assistance is limited to a maximum of $20,000.

**Keep you on track with your treatment** by coordinating shipments with your specialty pharmacy and reminding you when it’s time to refill your Vertex medicine. And if your daily routine changes, they can help you pre-plan refills, ship your medicine to a new address, and share tips to help you stay motivated.

**Meet your everyday needs** with information on nutrition and tips for staying physically active and maintaining a healthy mindset. And if you’re caring for someone with CF, they’ll send educational resources to help you teach your loved one about the importance of their daily treatment routine.

**Plan for what’s ahead** as you approach big life changes. They can help you prepare for your next chapter and give you tips on staying on track with treatment. They can also share advice from others living with CF.

**Not enrolled in Vertex GPS?**
If you have been prescribed a Vertex medicine, ask your healthcare provider to complete an enrollment form for you.

**Already enrolled?**
If you are currently enrolled in GPS, you can call or text your Support Specialist at 1-877-752-5933 (press 2 when calling). Monday through Friday, from 8:30 am to 7 pm ET.

Discover more about GPS and the support resources available at VertexGPS.com.