What is KALYDECO?

KALYDECO is a prescription medicine used for the treatment of cystic fibrosis (CF) in patients age 6 months and older who have at least one mutation in their CF gene that is responsive to KALYDECO.

Talk to your doctor to learn if your child has an indicated CF gene mutation.

It is not known if KALYDECO is safe and effective in children under 6 months of age.

Please see Important Safety Information on pages 4-5 and full Prescribing Information for KALYDECO, including Patient Information.
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Please see Important Safety Information on pages 4-5 and full Prescribing Information for KALYDECO, including Patient Information.
Who should not take KALYDECO® (ivacaftor)?

Patients should not take KALYDECO if they take certain medicines or herbal supplements, such as:
- the antibiotics rifampin (Rifamate®, Rifater®) or rifabutin (Mycobutin®)
- seizure medicines such as phenobarbital, carbamazepine (Tegretol®, Carbatrol®, and Equetro®), or phenytoin (Dilantin®, Phenytek®)
- St. John’s wort (Hypericum perforatum)

Talk to the patient’s doctor before he/she takes KALYDECO if he/she takes any of the medicines or supplements listed above.

What should I tell the patient’s doctor before taking KALYDECO?

Before taking KALYDECO, tell the doctor if the patient:
- has liver or kidney problems
- drinks grapefruit juice, or eats grapefruit or Seville oranges

KALYDECO may affect the way other medicines work, and other medicines may affect how KALYDECO works.

Tell the patient’s doctor about all the medicines he/she takes, including prescription and non-prescription medicines, vitamins, and herbal supplements, as the dose of KALYDECO may need to be adjusted when taken with certain medications.

Especially tell the doctor if the patient takes:
- antifungal medications such as ketoconazole (e.g., Nizoral®), itraconazole (e.g., Sporanox®), posaconazole (e.g., Noxafil®), voriconazole (e.g., Vfend®), or fluconazole (e.g., Diflucan®)
- antibiotics such as telithromycin (e.g., Ketek®), clarithromycin (e.g., Biaxin®), or erythromycin (e.g., Ery-Tab®)

What should the patient avoid while taking KALYDECO?

- KALYDECO can cause dizziness in some people who take it. The patient should not do anything that would need the patient to be alert until it is known how KALYDECO affects the patient
- Avoid food containing grapefruit or Seville oranges while taking KALYDECO

What are the possible side effects of KALYDECO® (ivacaftor)?

KALYDECO can cause serious side effects.

High liver enzymes in the blood have been reported in patients receiving KALYDECO. The doctor will do blood tests to check the liver:
- before starting KALYDECO
- every 3 months during the first year of taking KALYDECO
- every year while taking KALYDECO

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

Call the doctor right away if the patient has any of the following symptoms of liver problems:
- pain or discomfort in the upper right stomach (abdominal) area
- yellowing of the skin or the white part of his/her eyes
- loss of appetite
- nausea or vomiting
- dark, amber-colored urine

Abnormality of the eye lens (cataract) has been noted in some children and adolescents receiving KALYDECO. The doctor should perform eye examinations prior to 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 the doctor if the patient has any side effect that bothers him/her or that does not go away. These are not all the possible side effects of KALYDECO. For more information, ask the doctor or pharmacist.

Call the doctor for medical advice about side effects. You are encouraged to report side effects to FDA at 1-800-FDA-1088.
Taking KALYDECO® (ivacaftor) oral granules

**KALYDECO oral granules**

KALYDECO comes in single-use, oral granule packets for children age 6 months to less than 6 years with cystic fibrosis (CF) who have at least one mutation in their CF gene that is responsive to KALYDECO. These small granules should be mixed with 1 teaspoon (5 mL) of soft food or liquid.

KALYDECO oral granules are prescribed based on weight, and come in 3 different strengths

<table>
<thead>
<tr>
<th>WEIGHT-BASED DOSE</th>
<th>EACH DOSE</th>
<th>TOTAL DAILY DOSE</th>
</tr>
</thead>
<tbody>
<tr>
<td>11 to &lt;15.4 pounds</td>
<td></td>
<td></td>
</tr>
<tr>
<td>~11 pounds to less than ~15 pounds (5 kg to &lt;7 kg)</td>
<td>25-mg packet (1 packet every 12 hours)</td>
<td>50 mg (2 packets per day)</td>
</tr>
<tr>
<td>15.4 to &lt;31 pounds</td>
<td></td>
<td></td>
</tr>
<tr>
<td>~15 pounds to less than ~31 pounds (7 kg to &lt;14 kg)</td>
<td>50-mg packet (1 packet every 12 hours)</td>
<td>100 mg (2 packets per day)</td>
</tr>
<tr>
<td>≥31 pounds</td>
<td></td>
<td></td>
</tr>
<tr>
<td>~31 pounds or more</td>
<td></td>
<td></td>
</tr>
<tr>
<td>(14 kg or more)</td>
<td>75-mg packet (1 packet every 12 hours)</td>
<td>150 mg (2 packets per day)</td>
</tr>
</tbody>
</table>

Your child’s dose may be different. Your healthcare provider will tell you how much KALYDECO your child should take and when to take it.

**Remember**—having your child take KALYDECO every 12 hours is not the same as having him or her take it twice a day.

**How to give your child KALYDECO® (ivacaftor) oral granules**

**Step 1: Preparation**

- Hold 1 packet of KALYDECO oral granules with the cut line on top
- Shake the packet gently to settle the KALYDECO granules
- Tear or cut the packet open along the cut line
- Carefully pour all of the KALYDECO granules into 1 teaspoon (5 mL) of age-appropriate soft food or liquid such as puréed fruits or vegetables, yogurt, applesauce, water, breast milk, prepared infant formula, milk, or juice
  - Food or liquid should be at or below room temperature
- Then, mix the granules with the 1 teaspoon of food or liquid

**Step 2: Administration**

- Within 1 hour of mixing, give KALYDECO to your child
- Make sure the entire medicine mixture is taken

**STEP 3: Fat-containing food before or after the dose**

Always take KALYDECO with fat-containing food

- Give your child fat-containing food just before or after the dose of KALYDECO granules. This helps the body absorb KALYDECO better
- Examples of fat-containing food include:
  - Eggs
  - Peanut butter
  - Whole milk
  - Whole-milk yogurt
  - Butter
  - Cheese pizza
  - Whole-milk cheese
- Your doctor can help you choose healthy fat-containing meals and snacks

Continued ▶
What to do if your child misses a dose of KALYDECO?

- If your child misses a dose of KALYDECO and it is **within 6 hours** of when your child usually takes it, give your child the dose of KALYDECO as prescribed with fat-containing food as soon as possible.
- If your child misses a dose and it is **more than 6 hours** after the time your child usually takes it, **skip that dose only** and give the next dose with fat-containing food when your child usually takes it. Do **not** give 2 doses at the same time to make up for your child’s missed dose.

Helpful reminders and tips for taking KALYDECO oral granules

- Take KALYDECO oral granules, plus all other CF therapies, exactly as prescribed.
- Take KALYDECO oral granules every 12 hours* or as directed by your child’s healthcare provider.
- Take KALYDECO oral granules at the same times every day.
- Set an alarm on your clock, watch, cell phone, or computer to remind you to give your child KALYDECO.

*Your child’s dose of KALYDECO may be different. Take KALYDECO granules as directed by your child’s healthcare provider.

Learn more about KALYDECO and how to take KALYDECO oral granules at KALYDECO.com/pediatric-video.
Frequently asked questions

Q: What is the recommended dose of KALYDECO® (ivacaftor) oral granules?

Please see page 6 for weight-based dose recommendations for KALYDECO oral granules. Your child’s dose may be different. Your healthcare provider will tell you how much KALYDECO your child should take and when to take it.

Q: Can I mix KALYDECO oral granules in breast milk or prepared infant formula?

Yes, KALYDECO oral granules can be mixed with 1 teaspoon (5 mL) of breast milk or prepared infant formula at or below room temperature. Be sure that the entire mixture is taken by the child within 1 hour. Remember, your child should always take KALYDECO with fat-containing food.

Q: Can I mix KALYDECO granules in soft food or liquid that contains fat? If so, do I still need to give fat-containing food to my child?

Yes, you can mix KALYDECO granules in fat-containing food, for example, 1 teaspoon (5 mL) of whole-milk yogurt or milk, to give to your child. But your child should still eat fat-containing food just before or after taking the spoonful of the mixture.

Q: Do breast milk and prepared infant formula qualify as fat-containing foods that my child can take with KALYDECO?

Yes, breast milk and prepared infant formula qualify as fat-containing foods.

Q: Does the temperature of the food that I mix with KALYDECO oral granules matter? Can I mix oral granules in breast milk or prepared infant formula that I’ve warmed?

The granules should be mixed with soft food or liquid at room temperature or below. The granules should not be mixed in items that are frozen or hot.

Q: After the granules are mixed in soft food or liquid, how quickly does my child need to take the medicine?

The granules should be taken within 1 hour of mixing with soft food or liquid. Make sure all medicine is taken.

Q: Can I mix KALYDECO oral granules in breast milk or prepared infant formula?

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Explore how Vertex GPS™: Guidance & Patient Support can help

Vertex GPS provides eligible patients with reimbursement support, information and financial resources, refill reminders, and ongoing educational materials.

If you are currently enrolled and want to learn more about how GPS can support you and your child, you can speak to your Case Manager at 1-877-752-5933 (press 2), Monday through Friday from 8:30 AM to 7:00 PM ET. If your child has been prescribed KALYDECO® (ivacaftor) and you are not enrolled, please speak with your child’s healthcare provider.

To find out more about GPS and the support resources available to you, visit VertexGPS.com.

Please see full Prescribing Information for KALYDECO, including Patient Information.