

Patient Information
KALYDECO® (kuh-LYE-deh-koh)
(ivacaftor) tablets, for oral use
(ivacaftor) oral granules

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.

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 end of this patient information leaflet for a complete list of ingredients in KALYDECO.
- 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 such as 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®)

Know the medicines you take. Keep a list of them to show your doctor and pharmacist when you get a new medicine.

How should I take KALYDECO?

- Take KALYDECO exactly as your doctor tells you to take it.
- Take your doses of KALYDECO 12 hours apart.
- **Always take KALYDECO tablets or oral granules with food that contains fat.** Examples of fat-containing foods include eggs, butter, peanut butter, cheese pizza, and whole-milk dairy products such as whole milk, cheese, yogurt, breast milk, or infant formula.
- KALYDECO Tablets (ages 6 years and older):
 - Each KALYDECO box contains 4 individual blister cards.
 - Each blister card contains 14 tablets: 7 morning doses and 7 evening doses.
 - In the morning, unpeel the paper backing from a blister card to remove 1 KALYDECO tablet and take it with food that contains fat.
 - In the evening, 12 hours later, open another blister card to remove 1 KALYDECO tablet and take it with food that contains fat.
 - Swallow the KALYDECO tablets whole.
 - You may cut along the dotted line to separate your doses from the blister card.
- KALYDECO Oral Granules (ages 1 month to under 6 years old):
 - Hold the packet 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 in the packet into 1 teaspoon of soft food or liquid. Food or liquid should be at or below room temperature. Some examples of soft foods or liquids include puréed fruits or vegetables, yogurt, applesauce, water, breast milk, infant formula, milk, or juice.
 - Mix the KALYDECO granules with soft food or liquid.
 - After mixing, give KALYDECO within 1 hour. Make sure all medicine is taken.

- Give a child fat-containing food just before or just after the KALYDECO granules dose. Examples of fat-containing foods include eggs, butter, peanut butter, cheese pizza, and whole-milk dairy products such as whole milk, cheese, yogurt, breast milk and infant formula.
- If you miss a dose of KALYDECO and it is **within 6 hours** of when you usually take it, take your dose of KALYDECO as prescribed with fat-containing food as soon as possible.
- If you miss a dose of KALYDECO and it is **more than 6 hours** after the time you usually take it, **skip that dose only** and take the next dose when you usually take it. Do **not** take 2 doses at the same time to make up for your missed dose.

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 food or drink containing grapefruit while you are taking KALYDECO.

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** can happen to people who are treated with KALYDECO. Call your healthcare provider or go to the emergency room right away if you have any 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 | • diarrhea |
| • upper respiratory tract infection (common cold) including sore throat, nasal or sinus congestion, and runny nose | • rash |
| | • nausea |
| • stomach (abdominal) pain | • 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.

Call your doctor for medical advice about side effects. You may report side effects to FDA at 1-800-FDA-1088.

How should I store KALYDECO?

- Store KALYDECO at room temperature between 68°F to 77°F (20°C to 25°C).

Keep KALYDECO and all medicines out of the reach of children.

General information about the safe and effective use of KALYDECO.

Medicines are sometimes prescribed for purposes other than those listed in a Patient Information leaflet. Do not use KALYDECO for a condition for which it was not prescribed. Do not give KALYDECO to other people, even if they have the same symptoms that you have. It may harm them.

You can ask your pharmacist or doctor for information about KALYDECO that is written for health professionals.

What are the ingredients in KALYDECO?

Ivacaftor tablets:

Active ingredients: ivacaftor.

Inactive ingredients: colloidal silicon dioxide, croscarmellose sodium, hypromellose acetate succinate, lactose monohydrate, magnesium stearate, microcrystalline cellulose, and sodium lauryl sulfate.

The tablet film coat contains: carnauba wax, FD&C Blue #2, PEG 3350, polyvinyl alcohol, talc, and titanium dioxide.

The printing ink contains: ammonium hydroxide, iron oxide black, propylene glycol, and shellac.

Ivacaftor oral granules:

Active ingredients: ivacaftor.

Inactive ingredients: colloidal silicon dioxide, croscarmellose sodium, hypromellose acetate succinate, lactose monohydrate, magnesium stearate, mannitol, sucralose, and sodium lauryl sulfate.



Manufactured for: Vertex Pharmaceuticals Incorporated, 50 Northern Avenue, Boston, MA 02210

KALYDECO, VERTEX, and the VERTEX triangle logo are registered trademarks of Vertex Pharmaceuticals Incorporated.

All other trademarks referenced herein are the property of their respective owners.

©2023 Vertex Pharmaceuticals Incorporated

For more information, go to www.kalydeco.com or call 1-877-752-5933.

This Patient Information has been approved by the U.S. Food and Drug Administration.

Revised: 08/2023