

oral granules

kalydeco[®]

(ivacaftor) oral granules
5.8 mg • 13.4 mg • 25 mg • 50 mg • 75 mg

A GUIDE TO GIVING KALYDECO[®] (ivacaftor)

For caregivers of children age
1 month to less than 6 years

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.



People with CF pictured may or may not be taking KALYDECO.

What is KALYDECO[®] (ivacaftor)?

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](#).

kalydeco[®]
(ivacaftor) tablets 150 mg
oral granules 5.8-13.4-25-50-75 mg

Click below to get the information you're looking for.

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Please see [Important Safety Information](#) and [full Prescribing Information](#), including [Patient Information](#).

What is the Important Safety Information for KALYDECO® (ivacaftor)?

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 additional [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.

What is the recommended dose of KALYDECO® oral granules?

KALYDECO oral granules

KALYDECO comes in single-use, oral granule packets for children age 1 month 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.

Recommended dose of KALYDECO oral granules

If your loved one is:	And weighs:	The recommended dose is:
1 month to less than 2 months old	3 kg or more (~7 lb or more)	One 5.8-mg packet every 12 hours Total daily dose: 11.6 mg
2 months to less than 4 months old	3 kg or more (~7 lb or more)	One 13.4-mg packet every 12 hours Total daily dose: 26.8 mg
4 months to less than 6 months old	5 kg or more (~11 lb or more)	One 25-mg packet every 12 hours Total daily dose: 50 mg
6 months to less than 6 years old	5 kg to less than 7 kg (~11 lb to less than ~15 lb)	One 25-mg packet every 12 hours Total daily dose: 50 mg
	7 kg to less than 14 kg (~15 lb to less than ~31 lb)	One 50-mg packet every 12 hours Total daily dose: 100 mg
	14 kg or more (~31 lb or more)	One 75-mg packet every 12 hours Total daily dose: 150 mg

Your healthcare provider may give you different instructions on how much KALYDECO to give your loved one and when you should give it.



Remember—having your loved one take KALYDECO every 12 hours is not the same as having them take it twice a day.



Please see [Important Safety Information](#) and [full Prescribing Information](#), including [Patient Information](#).

3 steps for giving your loved one KALYDECO oral granules

STEP 1: Preparation



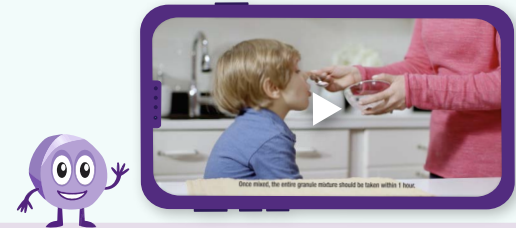
- Hold 1 packet of KALYDECO oral granules with the cut line on top
- Shake the packet gently to settle the KALYDECO granules
- Tear or cut the packet open along the cut line
- Carefully pour all of the KALYDECO granules into 1 teaspoon (5 mL) of age-appropriate soft food or liquid and mix

Here are some examples of soft foods and liquids you can mix KALYDECO granules into. Remember, every child is different, so be sure to talk to your healthcare provider about what the best options are for your loved one.

- Breast milk
 - Applesauce
 - Infant formula
 - Water
 - Puréed fruits or vegetables
 - Milk
 - Yogurt
 - Juice
- Food or liquid should be at or below room temperature
- Even if the food or liquid you mix the granules into contains fat, you still need to give your loved one fat-containing food. See steps 2 and 3 on the next page



Not actual size. Your loved one's KALYDECO carton and packets may look different than what is shown.



Join Grant the Granule and a family as they learn each step for preparing KALYDECO. Visit [KALYDECO.com/granules](https://www.kalydeco.com/granules).

Continued ►

STEP 2: Administration



- Give KALYDECO to your loved one within one hour of mixing
- Make sure the entire medicine mixture is given

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

Always give your loved one KALYDECO with a fat-containing food



- Give your loved one fat-containing food just before or just after the dose of KALYDECO granules. This helps the body absorb KALYDECO better

Here are suggestions for fat-containing foods to give your loved one. Please discuss with your healthcare provider what may be age-appropriate.

- Breast milk
- Infant formula
- Whole milk
- Whole-milk yogurt
- Eggs
- Whole-milk cheese
- Butter
- Peanut butter
- Cheese pizza

What to do if your loved one misses a dose of KALYDECO



If a dose of KALYDECO is missed and it is **within 6 hours** of when it is usually given, give that dose of KALYDECO as prescribed with fat-containing food as soon as possible



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



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

Helpful reminders and tips for giving KALYDECO oral granules to your loved one



• Give KALYDECO oral granules, plus all other CF therapies, exactly as prescribed



• Give **KALYDECO oral granules every 12 hours*** or as directed by your loved one's healthcare provider

• Give KALYDECO oral granules at the same times every day



• Set a reminder on your smartphone, watch, or calendar to help you remember when to give your loved one KALYDECO

*Your loved one's dose of KALYDECO may be different. Give KALYDECO granules as directed by your loved one's healthcare provider.

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.

Everyday^{CF}

Get delicious recipes and food ideas on [Everyday-CF.com](https://www.everyday-cf.com).



Please see [Important Safety Information](#) and [full Prescribing Information](#), including [Patient Information](#).

Q: What is the recommended dose of KALYDECO oral granules?

Please see [page 5](#) for weight- and age-based dose recommendations for KALYDECO oral granules. Your loved one's dose may be different. Your loved one's healthcare provider will tell you how much KALYDECO they should take and when to take it.

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

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

Q: If I mix the KALYDECO oral granules with a fat-containing food, do I still need to give my loved one fat-containing food afterwards?

Yes, your loved one should still eat fat-containing food just before or just after taking the entire mixture.

Q: Does the temperature of the food that I mix with KALYDECO oral granules matter? Can the granules be mixed in foods that are hot or cold?

The granules should be mixed with soft food or liquid at room temperature or below.

Q: What happens if my loved one gains weight and becomes eligible for a higher dose?

The dose of KALYDECO is based on your loved one's age and weight. As they get older, your healthcare provider may adjust their dose depending on their weight.

Frequently asked questions about KALYDECO® oral granules (Continued)

Q: My loved one spit up their dose of KALYDECO oral granules. What should I do?

You should contact your loved one's healthcare provider to discuss this. Your pharmacist may also be a good resource.

Q: If my loved one is napping when it is time for KALYDECO oral granules, should I wait until they're awake to administer them?

You should give your loved one KALYDECO oral granules every 12 hours or as prescribed by their healthcare provider.

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

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

Q: Can my loved one swallow the granules without mixing them in soft food or liquid?

The entire contents of each packet should be mixed with 1 teaspoon (5 mL) of age-appropriate soft food or liquid. The mixture should be taken within 1 hour of being mixed. Make sure all the medicine is taken.

Q: Do the granules have a taste?

The granule formulation is sweetened but unflavored.

Q: How should KALYDECO oral granules be stored?

KALYDECO should be stored at 68°F to 77°F (20°C to 25°C). Do not use KALYDECO after the expiration date on the package. Keep KALYDECO and all medicines out of the reach of children.

Q: Should the granules be removed from the foil packet and stored elsewhere (for example, a pillbox)?

No, granules should be kept in the foil packet until they are ready to be used.

Q: Are the oral granules packets child proof?

The packets are child-resistant.



We're here to help you get there

Wherever life with cystic fibrosis (CF) takes you, Vertex GPSTM: 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.



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