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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Makson, Age 7, R117H

Makson at home with his mom, Kristen, practicing karate

Please see Important Safety Information on pages 4-5 and full Prescribing Information for KALYDECO, including Patient Information.
Who should not take KALYDECO?
Do not take KALYDECO if you 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 your doctor before taking KALYDECO if you take any of the medicines or supplements listed above.

What should I tell my doctor before taking KALYDECO?
Before you take KALYDECO, tell your doctor if you:
- have liver or kidney problems
- drink grapefruit juice, or eat grapefruit or Seville oranges
- 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

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

Tell your doctor about all the medicines you take, 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 your doctor if you take:
- 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 I avoid while taking KALYDECO?
- KALYDECO can cause dizziness in some people who take it. Do not drive a car, use machinery, or do anything that needs you to be alert until you know how KALYDECO affects you
- You should avoid food containing grapefruit or Seville oranges while you are 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.
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 patients who have had high liver enzymes in the past, the 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

Abnormality of the eye lens (cataract) has been noted in some children and adolescents receiving KALYDECO. Your doctor should perform eye examinations prior to and during treatment with KALYDECO to look for cataracts.

The most common side effects 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.

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

Please see full Prescribing Information for KALYDECO, including Patient Information.
Taking KALYDECO® (ivacaftor) tablets

<table>
<thead>
<tr>
<th>Take 1 KALYDECO tablet (150 mg) every 12 hours with fat-containing food or as directed by your healthcare provider</th>
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<tbody>
<tr>
<td>• Taking KALYDECO every 12 hours helps keep enough medicine in the body</td>
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<tr>
<td>• Remember, taking KALYDECO every 12 hours is not the same as taking it twice a day</td>
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<tr>
<td>• Your dose may be different—your healthcare provider will tell you how much KALYDECO to take and when to take it</td>
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<tr>
<td>• 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</td>
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<tr>
<th>What to do if you miss a dose of KALYDECO® (ivacaftor)</th>
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<tbody>
<tr>
<td>• If you miss a dose of KALYDECO and it is <strong>within 6 hours</strong> of when you usually take it, take your dose of KALYDECO as prescribed with fat-containing food as soon as possible</td>
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<tr>
<td>• If you miss a dose of KALYDECO and it is <strong>more than 6 hours</strong> 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</td>
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<tr>
<th>Continue to take KALYDECO plus all your other CF therapies as directed by your healthcare provider</th>
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<tr>
<td>• 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</td>
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<tr>
<th>Avoid foods or drinks that contain grapefruit or Seville oranges—these may affect the amount of KALYDECO in the body.</th>
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Learn about different types of fats on the next page. 

Please see Important Safety Information on pages 4-5 and full Prescribing Information for KALYDECO, 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 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 and olives
- Many nuts and seeds, such as walnuts and sunflower seeds
- Fatty fish, such as salmon, mackerel, herring, and trout

**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 actually unsaturated. That makes it about as healthy a fat source as olive oil! And it tastes great with a banana.

“In the morning I get Makson what he wants for breakfast. Usually toast with peanut butter or butter, something high fat.”

—Makson’s mom, Kristen

Makson, Age 7, R117H
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 beneficial substances. 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.

<table>
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<tr>
<th>MEAL</th>
<th>IF YOU HAVE TIME</th>
<th>IF YOU’RE IN A RUSH</th>
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| **Breakfast** | • Scrambled or poached egg with avocado on multigrain toast  
• Chocolate chip pancakes  
• Peach yogurt shake† | • Bagel with peanut butter  
• Egg and cheese on an English muffin |
| **Snack**  | • English muffin pizza  
• Peanut butter and jelly smoothie  
• Chocolate pudding† | • Cookies with whole milk  
• Trail mix  
• Tortilla chips with guacamole |
| **Dinner** | • Macaroni and cheese†  
• Turkey burger with sliced avocado and tomatoes  
• Salmon and rice  
• Tater tot bake† | • Ham and cheese sandwich with tomatoes  
• Grilled cheese sandwich |

*These lists are not intended to be complete.
†Find these recipes on the facing pages.

Looking for new recipes?
Visit VertexGPS.com for Delicious Dishes recipe videos featuring our very own Case Managers. For more information on Vertex GPS™: Guidance & Patient Support, see the back cover of this brochure or visit VertexGPS.com.
**Simple mac and cheese**

**INGREDIENTS**
1 (8 ounce) box of elbow macaroni  
¼ cup butter  
¼ cup all-purpose flour  
½ teaspoon salt  
2 cups milk  
2 cups shredded cheddar cheese  
Black pepper to taste

**DIRECTIONS**
1. Bring a large pot of lightly salted water to a boil.
2. Cook elbow macaroni in the boiling water, stirring occasionally until cooked through but firm to the bite (about 8 minutes). Drain.
3. Melt butter in a saucepan over medium heat; stir in flour, salt, and pepper until smooth (about 5 minutes).
4. Slowly pour milk into butter-flour mixture while continuously stirring until mixture is smooth and bubbling (about 5 minutes).
5. Add cheddar cheese to milk mixture and stir until cheese is melted (2 to 4 minutes).
6. Fold macaroni into cheese sauce until coated.

**Tater tot bake**

**INGREDIENTS**
1 pound ground beef  
1 onion, chopped  
Salt and pepper to taste  
16 ounces tater tots  
1 (10.75 ounce) can condensed cream of mushroom soup  
½ cup milk  
½ cup shredded cheddar cheese

**DIRECTIONS**
1. Preheat oven to 350°F.
2. In a large skillet over medium-high heat, brown the ground beef with the onions.
3. Drain excess fat, and season with salt and pepper to taste.
4. Spread the beef mixture evenly over the bottom of a 2-quart casserole dish.
5. Arrange tater tots evenly over beef layer. In a small bowl, stir the soup into the milk until smooth; pour over tater tot and beef layers.
6. Sprinkle cheddar cheese evenly over the top.
7. Bake for 30 to 40 minutes, until cheese is bubbly and slightly brown.
**Peach yogurt shake**

**INGREDIENTS**

1 cup sliced peaches
1 cup plain whole-milk yogurt
1 cup whole milk
1 tablespoon honey

**DIRECTIONS**

Combine all ingredients in a blender and blend until smooth.

Variations: Instead of peaches, use 1 cup of sliced bananas, fruit cocktail, strawberries, raspberries, or blackberries.

Calories per serving: 460 | Fat: 16 g | Carbohydrates: 66 g | Protein: 16 g
Total servings: 1

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**Chocolate pudding**

**INGREDIENTS**

1/2 cup white sugar
3 tablespoons unsweetened cocoa powder
1/4 cup cornstarch
1/4 teaspoon salt
2 3/4 cups milk
2 tablespoons butter, room temperature
1 teaspoon vanilla extract

**DIRECTIONS**

1. In a saucepan, stir together sugar, cocoa, cornstarch, and salt. Place over medium heat, and stir in milk.
2. Bring to a boil and cook, stirring constantly, until mixture thickens enough to coat the back of a metal spoon.
3. Remove from heat, and stir in butter and vanilla. Let cool briefly, and serve warm, or chill in refrigerator until serving.

Calories per serving: 274 | Fat: 10 g | Carbohydrates: 42.5 g | Protein: 6 g
Total servings: 6

Source: CFF.org

Source: Allrecipes.com
How should I take KALYDECO® (ivacaftor) 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.

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

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

What should I avoid while taking KALYDECO?
KALYDECO can cause dizziness in some people who take it. Do not drive a car, use machinery, or do anything that needs you to be alert until you know how KALYDECO affects you. You should avoid food containing grapefruit or Seville oranges while you are taking KALYDECO.

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

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.

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

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

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

To find out more about Vertex GPS support resources, including Delicious Dishes, visit VertexGPS.com.

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