

**READ THIS FOR SAFE AND EFFECTIVE USE OF YOUR MEDICINE
PATIENT MEDICATION INFORMATION**

**Pr KALYDECO[®]
Ivacaftor tablets
Ivacaftor granules**

Read this carefully before you/your child start taking **KALYDECO** and each time you get a refill. This leaflet is a summary and will not tell you everything about this drug.

Remember, this does not take the place of your/your child's doctor's instructions. Talk to your healthcare professional about your/your child's medical condition and treatment and ask if there is any new information about **KALYDECO**.

What is KALYDECO used for?

KALYDECO tablets (150 mg) are for the treatment of cystic fibrosis (CF):

- in patients age 6 years and older and weighing 25 kg or more who have one of the following mutations in their CF gene: *G551D*, *G1244E*, *G1349D*, *G178R*, *G551S*, *S1251N*, *S1255P*, *S549N* or *S549R*.
- in patients age 18 years and older who have an R117H mutation in their CF gene.

KALYDECO granules (50 mg and 75 mg) are for the treatment of children with cystic fibrosis (CF):

- in patients age 2 years and older and weighing less than 25 kg who have one of the following mutations in their CF gene: *G551D*, *G1244E*, *G1349D*, *G178R*, *G551S*, *S1251N*, *S1255P*, *S549N* or *S549R*.

KALYDECO is not for use in people with CF with two copies of the *F508del* mutation.

It is not known if KALYDECO is safe and effective in children under 2 years of age.

How does KALYDECO work?

KALYDECO belongs to a group of medicines called “*cystic fibrosis transmembrane conductance regulator (CFTR) potentiators*.” The CFTR protein is a channel at the surface of the cell that allows the movement of particles such as chloride in and out of the cell, contributing to salt and water balance. KALYDECO helps this CFTR protein channel open more often allowing more chloride to pass through.

KALYDECO comes in the following dosage forms:

Tablets: 150 mg

Granules: 50 mg and 75 mg

What are the ingredients in KALYDECO?

Medicinal ingredients: ivacaftor

Non-medicinal ingredients in 150 mg tablets:

carnauba wax, colloidal silicon dioxide, croscarmellose sodium, hypromellose acetate succinate, indigo carmine aluminum lake, lactose monohydrate, magnesium stearate, microcrystalline cellulose, PEG 3350, polyvinyl alcohol, sodium lauryl sulfate, talc, and titanium dioxide. The printing ink contains ammonium hydroxide, iron oxide black, propylene glycol, and shellac.

Non-medicinal ingredients in 50 mg and 75 mg granules:

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

Do not use KALYDECO if:

You are allergic to ivacaftor or any of the non-medicinal ingredients.

To help avoid side effects and ensure correct use, talk to your healthcare professional before you take KALYDECO. Talk about any health conditions or problems you may have, including if you:

- have kidney or liver disease.
- have a problem with the lens of your eye (cataract). Your doctor may recommend eye exams before and after your treatment with 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.

Other warnings you should know about:

Driving and using machines: You may get dizzy when you take KALYDECO. Wait to see how you feel after taking KALYDECO before you drive or use machines. **For children:** you or your child's caregiver should supervise your child when they take KALYDECO. Wait to see if your child is dizzy after taking KALYDECO before they ride their bikes or do anything else that needs their full attention.

Abnormal blood test results:

KALYDECO can cause abnormal blood test results. Your doctor will decide if blood tests are needed.

If you have had high liver enzymes in the past, your doctor may order blood tests to check your liver more often.

Tell your healthcare professional about all the medicines you take, including any drugs, vitamins, minerals, natural supplements or alternative medicines.

The following may interact with KALYDECO:

- antifungal medicines, such as ketoconazole, itraconazole, posaconazole, voriconazole, fluconazole
- antibiotics used for the treatment of bacterial infection, such as clarithromycin, erythromycin, rifampin, rifabutin
- phenobarbital, carbamazepine, phenytoin used for the treatment of epileptic seizures
- oral midazolam, alprazolam, diazepam, triazolam used for the treatment of anxiety, insomnia, agitation, etc.
- digoxin is used for the treatment of mild to moderate congestive heart failure and an abnormal heart rhythm called atrial fibrillation
- cyclosporine, tacrolimus are used after an organ transplantation
- warfarin is an anticoagulant used to prevent heart attacks, stroke, and blood clots
- St. John's wort (*Hypericum perforatum*) is an herbal medicine
- grapefruit juice, grapefruit, and Seville oranges

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

How to take KALYDECO:

- Always take KALYDECO exactly how your doctor tells you.
- Take your doses of KALYDECO 12 hours apart.
- Even if you feel well, keep taking KALYDECO exactly how your doctor tells you.
- Check with your doctor if you are not sure about your dose.
- Do not change the dose or stop taking the medicine without first talking to your doctor.
- Your doctor may need to adjust your dose if you have liver disease or if you are taking medications that may interact with KALYDECO.

KALYDECO Tablets (for patients aged 6 years and older and weighing 25 kg or more):

One tablet every 12 hours by mouth with fat-containing food (see below). Do not crush or chew the tablet. Swallow the tablet whole.

- Each KALYDECO box contains 4 individual blister cards.
- There are 7 days of tablets for each blister card. Each blister card contains 14 tablets - 7 morning doses and 7 evening doses.
- You may cut along the dotted line to separate your doses from the blister card.
- **Morning Dose:** unpeel the paper backing from a blister card. Do not push the tablet through the paper backing because the tablets could break. Remove one KALYDECO tablet and swallow it whole with food that contains fat.

- **Evening Dose:** 12 hours after your first dose, unpeel the paper backing from another blister card to remove one KALYDECO tablet and swallow it whole with food that contains fat.

KALYDECO Granules (for children aged 2 years and older and weighing less than 25 kg): Your doctor will prescribe the appropriate amount based on weight. The usual dose is to take 1 packet of granules every 12 hours with fat-containing food.

How to prepare the granules:

- Each KALYDECO box contains 4 individual wallets.
- There are 7 days of granules in each wallet. Each wallet contains 14 packets of granules - 7 morning doses and 7 evening doses.
- Finish all doses from one wallet before starting a new one.
- **Morning Dose:** Remove the first dose from the wallet.
- Hold the packet with cut line on top.
- Shake the packet gently to settle the KALYDECO granules.
- Tear or cut packet open along 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. Each packet is for single use only. Some examples of soft foods or liquids include puréed fruits or vegetables, yogurt, applesauce, water, milk, or juice (except for grapefruit juice).
- Mix the KALYDECO granules with food or liquid.
- **After mixing, give KALYDECO within 1 hour. Make sure all medicine is taken. This is very important for KALYDECO to work properly and be effective.**
- If mixed with food, KALYDECO granules must not be stored for future use.
- **Evening Dose:** This dose should be taken 12 hours after your **Morning Dose**. Follow the same steps listed above under **Morning Dose**.

KALYDECO should always be taken with a fat-containing food:

- Taking KALYDECO with fat-containing food is important to get the right amount of medicine in your body.
- Each dose should be taken just before or just after fat-containing food.
- Meals and snacks recommended in CF guidelines or meals recommended in standard nutritional guidelines contain adequate amounts of fat. A serving size of foods appropriate for age from a typical CF diet should be given.
- Examples of meals that contain fat:
 - meals prepared with butter or oils
 - meals that have eggs, nuts, whole-milk dairy products (such as whole milk, cheese, and yogurt) or meats.

Overdose:

If you think you have taken too much KALYDECO, contact your healthcare professional, hospital emergency department or regional Poison Control Centre immediately, even if there are no symptoms.

Missed Dose:

- 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.

Refilling your prescription:

Remember to get a new prescription from your doctor or a refill from your pharmacy before all your tablets or granules are taken.

What are possible side effects from using KALYDECO?

These are not all the possible side effects you may feel when taking KALYDECO. If you experience any side effects not listed here, contact your healthcare professional.

- Diarrhea
- Nausea
- Stomach (abdominal) pain

- Common cold
- Runny nose
- Upper respiratory tract infection
- Changes in the type of bacteria in your sputum

- Headache
- Dizziness

- Nasal congestion
- Sinus congestion
- Sore throat

- Rash

- Joint pain

Serious Side Effects and What to do About Them

Symptom/Effect	Talk to your healthcare professional		Stop taking drug and get immediate medical help
	Only if severe	In all cases	
UNCOMMON			
Liver disorder: yellowing of the skin or eyes, dark urine, abdominal pain, nausea, vomiting, loss of appetite, pale stools, itchy skin		√	
Abdominal pain (stomach)	√		
Low blood sugar (glucose)	√		
UNKNOWN			
Allergic reaction: rash; hives; swelling of the face, lips, tongue or throat; difficulty swallowing or breathing			√

If you have other symptoms or side effects that are not listed here or becomes bad enough to interfere with your daily activities, talk to your healthcare professional.

Reporting Side Effects

You can help improve the safe use of health products for Canadians by reporting serious and unexpected side effects to Health Canada. Your report may help to identify new side effects and change the product safety information.

3 ways to report:

- Online at [MedEffect](#);
- By calling 1-866-234-2345 (toll-free);
- By completing a Consumer Side Effect Reporting Form and sending it by:
 - Fax to: 1-866-678-6789 (toll-free), or
 - Mail to: Canada Vigilance Program
Health Canada,
Postal Locator 0701E
Ottawa, ON
K1A 0K9

Postage paid labels and the Consumer Side Effect Reporting Form are available at [MedEffect](#).

NOTE: Contact your healthcare professional if you need information about how to manage your side effects. The Canada Vigilance Program does not provide medical advice.

Storage:

Store at 20-25°C; short periods to 15-30°C are permitted.

Keep out of reach and sight of children.

You may need to read this leaflet again. Please do not throw this away.

If you want more information about KALYDECO:

- talk to your healthcare professional.
- find the full product monograph that is prepared for healthcare professionals and includes this Patient Medication Information by visiting the Health Canada website (<http://www.hc-sc.gc.ca/index-eng.php>); the manufacturer's website, <http://www.vrtx.ca>; or by calling: 1-877-634-VRTX (8789).

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