MSK Kids

PATIENT & CAREGIVER EDUCATION

Lumacaftor and Ivacaftor

This information from UpToDate® Lexidrug™ explains what you need to know about this medication, including what it's used for, how to take it, its side effects, and when to call your healthcare provider.

Brand Names: US

Orkambi

Brand Names: Canada

Orkambi

What is this drug used for?

- It is used to treat cystic fibrosis.

What do I need to tell the doctor BEFORE my child takes this drug?

- If your child is allergic to this drug; any part of this drug; or any other drugs, foods, or substances. Tell the doctor about the allergy and what signs your child had.
- If your child has had an organ transplant.
- If your child is taking any of these drugs: Carbamazepine, phenobarbital, phenytoin, rifabutin, rifampin, or St. John's wort.
- If your child is taking any of these drugs: Cyclosporine, everolimus, midazolam, sirolimus, tacrolimus, or triazolam.
- If your child is taking birth control pills or some other hormone-based birth control.

This is not a list of all drugs or health problems that interact with this drug.

Tell the doctor and pharmacist about all of your child's drugs (prescription or OTC, natural products, vitamins) and health problems. You must check to make sure that it is safe to give this drug with all of your child's other drugs and health problems. Do not start, stop, or change the dose of any drug your child takes without checking with the doctor.

What are some things I need to know or do while my child takes this drug?

- Tell all of your child's health care providers that your child is taking this drug. This includes your child's doctors, nurses, pharmacists, and dentists.
- Have your child's blood work and lung function checked as you have been told by the doctor.
- High blood pressure has happened with this drug. Have your child's blood pressure checked as you have been told by the doctor.
- Cataracts have happened in children taking this drug. Be sure your child has an eye exam before starting this drug and while taking it. Talk with the doctor.
- Liver problems have happened with this drug. Liver problems have also gotten worse in people who already have them. Rarely, this has been deadly. If you have questions, talk with the doctor.

If your child is or may be sexually active:

- Birth control pills and other hormone-based birth control may not work as well to prevent pregnancy. Be sure your child uses some other kind of birth control also, like a condom, when taking this drug.

If your child is pregnant or breast-feeding a baby:

- Talk with the doctor if your child is pregnant, becomes pregnant, or is breast-feeding a baby. You will need to talk about the benefits and risks to your child and the baby.

What are some side effects that I need to call my child's doctor about right away?

WARNING/CAUTION: Even though it may be rare, some people may have very bad and sometimes deadly side effects when taking a drug. Tell your child's doctor or get medical help right away if your child has any of the following signs or symptoms that may be related to a very bad side effect:

- Signs of an allergic reaction, like rash; hives; itching; red, swollen, blistered, or peeling skin with or without fever; wheezing; tightness in the chest or throat; trouble breathing, swallowing, or talking; unusual hoarseness; or swelling of the mouth, face, lips, tongue, or throat.
- Signs of liver problems like dark urine, tiredness, decreased appetite, upset stomach or stomach pain, light-colored stools, throwing up, or yellow skin or eyes.
- Signs of high blood pressure like very bad headache or dizziness, passing out, or change in eyesight.
- Signs of new or worsening mental or mood problems like anxiety, depression, suicidal thoughts or actions, or trouble sleeping.
- Change in eyesight.
- Chest pain or pressure.
- Shortness of breath.
- Trouble breathing that is new or worse.
- Feeling confused.
- Raised pressure in the brain has happened during treatment with this drug and other drugs like it. The risk may be raised in people with high vitamin A levels. Call the doctor right away if your child has a headache; dizziness; upset stomach; ringing in the ears; eye pain; or eyesight problems like blurred eyesight, seeing double, or loss of eyesight.

What are some other side effects of this drug?

All drugs may cause side effects. However, many people have no side effects or only have minor side effects. Call your child's doctor or get medical help if any of these side effects or any other side effects bother your child or do not go away:

- Stomach pain or diarrhea.
- Upset stomach.
- Feeling tired or weak.
- Gas.
- Flu-like signs.
- Signs of a common cold.
- Nose or throat irritation.
- Cough.
- More sputum.
- Headache.

If your child has menstrual periods:

- Period (menstrual) changes.

These are not all of the side effects that may occur. If you have questions about side effects, call your child's doctor. Call your child's doctor for medical advice about side effects.

You may report side effects to your national health agency.

How is this drug best given?

Give this drug as ordered by your child's doctor. Read all information given to you. Follow all instructions closely.

All products:

- Give this drug just before or just after fatty foods. There are many fatty foods. Some of these are eggs; butter; peanut butter; cheese pizza; whole-milk dairy products like whole-milk, cheese, or yogurt; breast milk; or infant formula. If you are not sure what types of foods are fatty, talk with your doctor.
- Keep giving this drug to your child as you have been told by your child's doctor or other health care provider, even if your child feels well.
- Do not let your child eat grapefruit or drink grapefruit juice during the first week your child is taking this drug.

Granules:

- Before you open the packet, shake gently so the granules move to the bottom of the packet.
- Mix the contents of the packet with 1 teaspoon (5 mL) of soft food or liquid. This includes pureed fruits or vegetables, flavored yogurt or pudding, applesauce, water, breast milk, baby formula, milk, or juice. Food or liquid needs to be at or below room temperature.
- Give this drug within 1 hour of mixing.

What do I do if my child misses a dose?

- Give a missed dose as soon as you think about it.
- If it has been 6 hours or more since the missed dose, skip the missed dose and go back to your child's normal time.
- Do not give 2 doses at the same time or extra doses.
- If you miss giving your child 7 days of this drug, call your doctor to find out what to do.

How do I store and/or throw out this drug?

- Store at room temperature in a dry place. Do not store in a bathroom.
- Keep all drugs in a safe place. Keep all drugs out of the reach of children and pets.
- Throw away unused or expired drugs. Do not flush down a toilet or pour down a drain unless you are told to do so. Check with your pharmacist if you have questions about the best way to throw out drugs. There may be drug take-back programs in your area.

General drug facts

- If your child's symptoms or health problems do not get better or if they become worse, call your child's doctor.
- Do not share your child's drug with others and do not give anyone else's drug to your child.
- Some drugs may have another patient information leaflet. If you have any questions about this drug, please talk with your doctor, nurse, pharmacist, or other health care provider.
- If you think there has been an overdose, call your poison control center or get medical care right away. Be ready to tell or show what was taken, how much, and when it happened.

Consumer Information Use and Disclaimer

This generalized information is a limited summary of diagnosis, treatment, and/or medication information. It is not meant to be comprehensive and should be used as a tool to help the user understand and/or assess potential diagnostic and treatment options. It does NOT include all information about conditions, treatments, medications, side effects, or risks that may apply to a specific patient. It is not intended to be medical advice or a substitute for the medical advice, diagnosis, or treatment of a health care provider based on the health care provider's examination and assessment of a patient's specific and unique circumstances. Patients must speak with a health care provider for complete information about their health, medical questions, and treatment options,

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If you have questions or concerns, contact your healthcare provider. A member of your care team will answer Monday through Friday from 9 a.m. to 5 p.m. Outside those hours, you can leave a message or talk with another MSK provider. There is always a doctor or nurse on call. If you're not sure how to reach your healthcare provider, call 212-639-2000.

For more resources, visit www.mskcc.org/pe to search our virtual library.

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