Tips to help make the most of your treatment with VENTAVIS

INDICATION
What is VENTAVIS?
VENTAVIS® (iloprost) Inhalation Solution is a prescription medicine used to treat adults with certain kinds of severe pulmonary arterial hypertension (PAH), a condition in which blood pressure is too high in the blood vessels between the heart and the lungs. VENTAVIS may improve your ability to exercise and your symptoms for a short time by lowering your blood pressure and opening up the blood vessels in your lungs.

The study showing VENTAVIS is effective included mainly patients with NYHA Functional Class III-IV PAH. In these patients, PAH was caused by unidentified or hereditary factors (65%) or connective tissue diseases (23%). VENTAVIS has not been studied in children younger than 18 years old.

IMPORTANT SAFETY INFORMATION
What are the possible side effects of VENTAVIS?
VENTAVIS may cause side effects, including feeling dizzy, lightheaded and faint. If you have any of these side effects, you should stand up slowly when you get out of chairs or bed. Tell your doctor if your fainting gets worse during treatment with VENTAVIS. Your doctor may need to change your dose or your treatment.

Do not drive a car or operate any tools or machines if dizziness or fainting from low blood pressure is a problem for you.

You may have trouble breathing after taking VENTAVIS because it may cause the muscles around your airway to tighten (bronchospasm). Get emergency help right away if you have trouble breathing.

Please see accompanying full Prescribing Information and Patient Information. Please see Important Safety Information continued on back.
Your I-neb® AAD® System

Learn the ABCs—Angle, Breathing, Cleaning—to make the most of your VENTAVIS® (iloprost) Inhalation Solution treatment

Angle

Holding the I-neb AAD System at the proper angle ensures continuous drug delivery.

- Stand or sit in a comfortable, upright position.
- Hold the I-neb AAD System at a 90-degree angle to your mouth, like you would when eating a sandwich. Resting your elbows on the table while holding your I-neb AAD System makes it easy.
- The I-neb AAD System will remind you with 4 short beeps if you are not holding it at a 90-degree angle.
- Holding the I-neb AAD System at the wrong angle will increase the length of your treatment.

If you would like additional training on how to breathe correctly with your I-neb AAD System, please contact an Actelion Case Manager at 1-866-ACTELION (1-866-228-3546), or contact your specialty pharmacy. Your specialty pharmacy nurse has a breathing monitor that can help you learn to breathe your best with the I-neb AAD System.

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Breathing

Proper breathing is the single most important part of your treatment.

Keep your lips closed around the I-neb AAD System mouthpiece as you breathe in and out.

- Breathe in and out through your mouth, not your nose.
- The I-neb AAD System will use your first 3 breaths in and out to adapt to your own breathing pattern. As you begin your 4th breath in, VENTAVIS will be delivered.
- The I-neb AAD System will vibrate as you begin to take a breath in to let you know VENTAVIS is being delivered.

Relax and breathe in and out in a slow and steady manner.

- While breathing, try counting in your head, “one one-thousand, two one-thousand, three one-thousand.”
- The longer you can breathe in, the more VENTAVIS is delivered on each breath, and your treatment times may decrease.
- If you need a break, take one. Rest a minute or two, then restart your treatment. Remember, the I-neb AAD System will take 3 breaths in and out to adapt to your breathing pattern before VENTAVIS will be delivered again.
To keep your I-neb AAD System in the best working condition, clean it once a day and boil it weekly.

Once-a-day cleaning

- At the end of the day, remove the chamber lids with mesh and place them in the mesh wash basket.
- Place the mouthpiece, medication chamber, and chamber lids in the main wash basket. Using only one drop of dishwashing liquid, wash all the pieces in distilled water. (Do not use dishwashing liquid that contains bleach, fragrance, or antibacterial ingredients.)
- Rinse the pieces with more distilled water (never reuse the distilled water). Shake off the water and allow to air dry for 2 hours.

Weekly boiling

- Put all of the parts (not the body) in the cleaning basket.
- Boil in distilled water for 6 to 10 minutes. Do not microwave—some of the parts are metal. Also, do not wash in the dishwasher because it may damage the parts.
- Rinse with distilled water.
- Shake off the water and then allow to air dry.

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Dosing options

Take your VENTAVIS® (iloprost) Inhalation Solution treatments 6 to 9 times each day during waking hours, at least 2 hours apart, or as prescribed by your doctor.

VENTAVIS is available in 3 dosing options

<table>
<thead>
<tr>
<th>Dosing Option</th>
<th>Description</th>
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<tbody>
<tr>
<td>2.5 mcg with 10 mcg/mL—Red*</td>
<td>The specialty pharmacy will start you with this dose, which uses the red-latched medication chamber. If tolerated.</td>
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<tr>
<td>5.0 mcg with 10 mcg/mL—Purple*</td>
<td>Nearly all patients transition to this dose, which uses the purple-latched medication chamber.</td>
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<tr>
<td>5.0 mcg with 20 mcg/mL—Gold*</td>
<td>If you are experiencing long treatment times, your doctor may transition you to this dosing option, which may decrease your treatment times.</td>
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VENTAVIS 20 mcg/mL†

Higher concentration of medicine lowers the amount of solution you need to take by 50%, for shorter treatment times.

†For patients who are maintained at the 5 mcg dose and who have repeatedly experienced extended treatment times which could result in incomplete dosing.
• Concentrating on **angle**, **breathing**, and **cleaning** can help you make the most of your treatment with VENTAVIS® (iloprost) Inhalation Solution.

• If you have any questions about the I-neb® AAD® System or VENTAVIS, call Actelion Pathways® at 1-866-ACTELION (1-866-228-3546) or visit [www.4VENTAVIS.com](http://www.4VENTAVIS.com).

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**IMPORTANT SAFETY INFORMATION**

**What should I tell my doctor before taking VENTAVIS?**

VENTAVIS may not be right for you. Before taking VENTAVIS, tell your doctor about all of your medical conditions, including if you:

- have liver or kidney problems. Your doctor may need to give you a lower dose of VENTAVIS.
- are pregnant, or plan to become pregnant. It is not known if VENTAVIS can harm your unborn baby. VENTAVIS should only be used during pregnancy if the benefit to you is worth the possible risk to your baby.
- are breastfeeding. It is not known if VENTAVIS passes into your breast milk. You and your doctor should decide if you will take VENTAVIS or breastfeed.

Tell your doctor about all the medicines you take, including prescription and nonprescription medicines, vitamins, and herbal supplements. VENTAVIS and other medicines may affect each other, causing side effects. VENTAVIS may affect the way other medicines work, and other medicines may affect how VENTAVIS works.

Especially tell your doctor if you take:

- medicines used to treat high blood pressure or heart problems
- medicines that lessen blood clotting (warfarin, Coumadin, Jantoven)

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

**How should I take VENTAVIS?**

- Take VENTAVIS exactly as your doctor tells you to take it. Your doctor may change your dose if needed.
- You should not take VENTAVIS more than every 2 hours. The benefits of VENTAVIS may not last 2 hours, so you may adjust the times that you use it to cover planned activities.
- Do not drink VENTAVIS.
- Do not let VENTAVIS solution come into contact with your skin or eyes. If it does, rinse your skin or eyes with water right away.
- Do not allow other people to be exposed to VENTAVIS while you are breathing it, especially babies.
- If you take too much VENTAVIS, you may have a headache, red face, dizziness, nausea, vomiting and diarrhea. If this happens stop taking VENTAVIS. If your symptoms do not go away, call your doctor or get emergency help right away.

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Other important side effects of VENTAVIS include:

- bleeding
- low blood pressure
- red face (flushing)
- headaches
- increased cough
- nausea
- spasm of your jaw muscles that makes it hard to open your mouth

Talk to 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 VENTAVIS. 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.**

**Practice your ABCs every day**

- angle
- breathing
- cleaning