

Ventavis is indicated for the treatment of pulmonary arterial hypertension (PAH) (WHO Group 1) to improve a composite endpoint consisting of exercise tolerance, symptoms (NYHA Class), and lack of deterioration. Studies establishing effectiveness included predominately patients with NYHA Functional Class III-IV symptoms and etiologies of idiopathic or heritable PAH (65%) or PAH associated with connective tissue disease (23%).

Ventavis Patient Enrollment Forms

Patient name:

IMPORTANT SAFETY INFORMATION: Hypotension leading to syncope has been observed; Ventavis should therefore not be initiated in patients with systolic blood pressure less than 85 mmHg. Stop Ventavis immediately if signs of pulmonary edema occur; this may be a sign of pulmonary venous hypertension. Ventavis inhalation may cause bronchospasm and patients with a history of hyperreactive airway disease may be more sensitive. Serious adverse events reported at a rate of less than 3% included congestive heart failure, chest pain, supraventricular tachycardia, dyspnea, peripheral edema, and kidney failure. Vital signs should be monitored while initiating Ventavis. In clinical studies, the most common adverse events occurring more often ($\geq 6\%$) in Ventavis-treated patients than in patients taking placebo included vasodilation (flushing) (27% vs 9%), cough (39% vs 26%), headache (30% vs 20%), trismus (12% vs 3%), and insomnia (8% vs 2%). Ventavis has the potential to increase the hypotensive effect of vasodilators and antihypertensive agents. Ventavis also has the potential to increase risk of bleeding, particularly in patients maintained on anticoagulants.

Please see accompanying full prescribing information.





TO: PAH Pathways®

FAX NUMBER: 1-866-279-0669

FAXED FROM: _____

DATE/TIME: _____

FROM: _____

NUMBER OF PAGES (INCLUDING THIS ONE): _____

COMMENTS: _____

CHECKLIST

- 1) COMPLETE PATIENT ENROLLMENT
- 2) DOCUMENT PAH DIAGNOSIS
- 3) DETERMINE PAH CLINICAL STATUS
- 4) COMPLETE CCB TRIAL
- 5) PROVIDE REQUIRED DOCUMENTATION:
RIGHT HEART CATHETERIZATION,
ECHOCARDIOGRAM RESULTS, AND
HISTORY AND PHYSICAL NOTES

REMINDER: PLEASE INCLUDE PHOTOCOPY OF
BOTH SIDES OF PATIENT INSURANCE CARD.

**FAX COMPLETED FORMS TO:
1-866-279-0669**

**FOR MORE INFORMATION, CALL
PAH PATHWAYS: 1-866-ACTELION
1-866-228-3546**

*Submission of the Ventavis® (iloprost)
enrollment form is not a guarantee of
patient approval.*

*Additional testing and clinical information
may be requested in some cases, including:*

- ANA results
- PFTs
- V/Q perfusion scan
- Chest CT

Ventavis® (iloprost) Inhalation Solution

Fax To: 1-866-279-0669

PO Box 826, South San Francisco, CA 94083-0826
 Phone 1-866-ACTELION (1-866-228-3546) or Fax 1-866-279-0669

Prescription	Ventavis (iloprost) Inhalation Solution 2.5 mcg or 5 mcg (10 mcg/mL) inhalation as tolerated. 6 to 9 times per day during waking hours. Start with 2.5 mcg × 1. If tolerated, go to 5 mcg (10 mcg/mL) ongoing. If not tolerated, resume 2.5 mcg. If patient is maintained at 5 mcg (10 mcg/mL) dose and repeatedly experiences extended treatment times, transition to 5 mcg (20 mcg/mL). <input type="checkbox"/> If patient is maintained at Ventavis 5 mcg (10 mcg/mL) for 1 month, transition to Ventavis 5 mcg (20 mcg/mL) starting at month 2, unless contacted by physician. Or please provide dosing instructions: _____ _____
	Dispense 1-month supply. Refills (select 1): 0 1 2 3 4 5 6 7 8 9 10 11 Send one (1)* I-neb® AAD® System if this is an initial order. <i>*If the patient resides in a remote area that does not allow for timely delivery (delivery within 8 hours), two (2) I-neb AAD Systems will be dispensed.</i>
	Nursing services requested. Skilled nursing visit for patient education related to therapy and disease state, administration of medication as prescribed, and assessment of general status and response to therapy. One to 3 visits to be provided for patient training. Patient training: <input type="checkbox"/> Specialty pharmacy to conduct initial patient training; initial training with I-neb Insight® breathing monitor required. <input type="checkbox"/> PAH treatment center to conduct initial patient training; initial training with I-neb Insight® breathing monitor required. Or please provide patient training instructions: _____ _____
	Follow-up nursing visits as ordered by physician to ensure patient is proficient in medication use and I-neb AAD administration.

Ship-to directions: <input type="checkbox"/> Physician office <input type="checkbox"/> Patient's home <input type="checkbox"/> Hospital If shipped to physician's office, physician accepts delivery on behalf of patient for administration in office.	
Address (no PO Box):	
City:	
State:	Zip:
Ship Attn:	

I certify that I am prescribing Ventavis for this patient as a medically appropriate treatment. By signing, I certify that the therapy is medically necessary and that I authorize the designated specialty pharmacy to be my designated agent to (1) provide any information on this form to the insurer of the named patient and (2) to forward the prescription by fax or other mode of delivery to the designated specialty pharmacy. This Appointment and Authorization shall be in force until cancelled in writing by the prescribing physician.

PRESCRIBER SIGNATURE _____ **DATE** _____

REQUIRED: PLEASE PROVIDE COPIES OF PATIENT'S CURRENT MEDICAL INSURANCE AND PRESCRIPTION CARDS.

Specialty Pharmacy	Indicate specialty pharmacy preference: If no preference is indicated, this referral will be sent to the appropriate specialty pharmacy based on the patient's existing insurance benefits.	<input type="checkbox"/> Benefit verification only. Do not send drug at this time. <input type="checkbox"/> Request pre-training demonstration visit only at this time.
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Physician Information	Name:	DEA #:	NPI #:	
	Name of facility:	MD specialty:	Tax ID #:	
	Contact name and phone #:	State license #:	Phone #:	
	Address:	City:	State:	Zip:

Patient Information	Name:	SSN:	DOB:	
	Address:	City:	State:	Zip:
	Preferred language, if not English:	Phone #:	Sex: <input type="checkbox"/> Male <input type="checkbox"/> Female	
	Caregiver name:	Relationship:	Alternate phone #:	

Insurance Information	Primary insurance company:	Phone #:	
	Policy holder name:	ID #:	Group/policy #:
	Secondary insurance name:	Phone #:	
	Policy holder name:	ID #:	Group/policy #:
	Prescription coverage name:	Phone #:	ID #:

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By signing below, I authorize Actelion and other entities involved with its medication-access program, and their employees, distributors, or agents, to use and give out my health information to run the medication-access program and any related education and patient-assistance programs. I also allow my health plans, other payers, pharmacies, and other healthcare providers to give my health information to Actelion as needed to help find ways to pay for Ventavis, or for treatment or healthcare operations purposes. I agree that my health information may be given to insurance companies, the Food and Drug Administration, or other government agencies (to comply with state and federal regulation or coverage eligibility requirements), charities, or other parties as necessary to participate in the medication-access program and run the program. I know that this program may be changed or stopped at any time. I may also cancel this authorization in the future by notifying PAH Pathways in writing and submitting the notification by fax to 1-866-279-0669. I know that completing this form does not ensure that I will receive therapy. I understand that Actelion does not promise to find ways to pay for my Ventavis, and I know that I am responsible for the costs of my care. I understand that signing this form does not commit me to paying for Ventavis. I also certify that the information I have set forth in this application is true, correct, and complete.

Patient/guardian signature: _____

Date: _____

Patient: _____ DOB: _____

Physician: _____

Prescriber signature: _____

Date: _____

It is the responsibility of the Prescriber to complete this form with information that most accurately and completely describes the condition of the patient, regardless of the potential impact on insurance coverage or reimbursement. Actelion makes no representation that the diagnosis information printed on this form is accurate or complete or that it will support insurance coverage or reimbursement.

Please select the diagnosis information that most accurately and completely describes the signs, symptoms, and condition of the patient:

WHO GROUP 1 PAH DIAGNOSES

- ICD-9: 416.0 Primary Pulmonary Arterial Hypertension (Idiopathic PAH)
- ICD-9: 416.0 Familial Pulmonary Arterial Hypertension (FPAH)
- ICD-9: 416.8 Secondary Pulmonary Hypertension (Associated PAH)

Please specify one:

___ Connective Tissue Disease (eg, CREST, MCTD, Scleroderma, Lupus)

___ Other: _____

OTHER

- ICD-9: Description: _____

MEDICAL RATIONALE FOR OTHER

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*From Proceedings of the 3rd World Symposium on Pulmonary Arterial Hypertension. Venice, Italy, June 23-25, 2003. *J Am Coll Cardiol.* 2004;43:1S-90S.

Patient: _____ DOB: _____

Physician: _____

Prescriber signature: _____

Date: _____

NYHA/WHO Functional Class: (Check only one)

- Class 3
- Class 4
- Other: _____

Clinical Signs and Symptoms: (Check all appropriate)

- Fatigue
- Shortness of breath or dyspnea on exertion
- 6-minute walk: _____ meters Date of evaluation: _____
- Chest pain or pressure
- Syncope or near syncope
- Edema or fluid retention
- Increasing limitation of physical activity
- Other: _____

Course of Illness: (Check all appropriate)

- Evidence of worsening heart failure (eg, rales on physical exam, worsening edema, increased NT-proBNP, increased CRP)
- Worsening pulmonary hemodynamics (eg, mPAP, RAP, PVR, CO)
- Decreasing 6-minute walk test
- Change in functional class
- Worsening dyspnea on exertion
- Change in patient-reported symptoms (eg, increased fatigue)
- Other: _____

Fax To: 1-866-279-0669

Patient: _____ DOB: _____

Physician: _____

Prescriber signature: _____

Date: _____

Prior to the initiation of Ventavis® (iloprost), Medicare policy requires documentation that a calcium channel blocker (CCB) has been tried, failed, or considered and ruled out.¹ Because many PAH patients may become disabled and Medicare-eligible, the following documentation is generally required prior to initiation of treatment with Ventavis.

The above named patient was trialed as follows:

A CCB WAS NOT TRIALED BECAUSE:

- Patient did not meet ACCP Guidelines² for Vasodilator Response (ie, a fall in mPAP ≥ 10 mm Hg to ≤ 40 mm Hg, with an unchanged or increased cardiac output)
- Patient is hemodynamically unstable or has history of postural hypotension
- Patient has systemic hypotension (SBP ≤ 90 mm Hg)
- Patient has depressed cardiac output (cardiac index ≤ 2.4 L/min/m²)
- Patient has known hypersensitivity
- Patient has documented bradycardia or second- or third-degree heart block
- Patient has signs of right-sided heart failure
- Other: _____

OR

THE FOLLOWING CCB WAS TRIALED:

CCB: _____

With the following response:

- Pulmonary arterial pressure continued to rise
- Disease continued to progress or patient remained symptomatic
- Patient hypersensitive or allergic
- Adverse event: _____
- Patient became hemodynamically unstable
- Other: _____

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1. Centers for Medicare and Medicaid Services. Coverage for iloprost for pulmonary artery hypertension. <http://www.cms.hhs.gov/>. Accessed March 26, 2008. **2.** Badesch DB, Abman SH, Simonneau G, Rubin LJ, McLaughlin VV. Medical therapy for pulmonary arterial hypertension: updated ACCP evidence-based clinical practice guidelines. *Chest*. 2007;131:1917-1928.

Patient: _____ DOB: _____

Physician: _____

PLEASE CHECK EACH BOX ONCE COMPLETED.

Right heart catheterization has been performed. Results form is attached.

The right heart catheterization report should include:

- Mean pulmonary artery pressure (or systolic and diastolic pressure)
- Cardiac output (CO)
- Pulmonary vascular resistance (PVR)
- Pulmonary capillary wedge pressure (or LVEDP)

Echocardiogram has been performed to rule out left-sided heart or valvular disease. Results form is attached.

Current history and physical notes with need for therapy and PAH symptoms (eg, dyspnea on exertion, fatigue, angina, syncope) documented. Notes are attached.

Prescriber Initials: _____ Date: _____

SAMPLE RIGHT HEART CATHETERIZATION RESULTS FORM

*PPH Hemodynamic DATA COLLECTION SHEET
Acute Study - Cardiac Catheterization Lab*

Patient Name: _____ M.R. #: _____ Date: _____
 Ht: _____ cm Wt: _____ kg BSA: _____
 Physician: _____ Age: _____
 Diagnosis: R/O PPH _____ Tech: _____ Birthday: _____

	Baseline	Nitro/O ₂	Exercise	End Ex	Dose 1	Dose 2	Baseline	Comments
Time Measured								
Heart Rate								
Body Temp								
Resp. rate								
FiO ₂ %								
SaO ₂ %								
RV								
PA sys/dias								
PA mean								
PA wedge								
AO sys/dias								
AO mean								
CVP								
Id C.O./C.I								
Id SVR/SVRI								
PVR/PVRI/dwgs								
TPR								
PVR/wood								
Stroke Vol. ml/b								
Hepatic wedge								
hepatic vein								
PAw Sat%								
RA Sat%								
IVC Sat%								
SVC Sat%								
RV Sat%								
PA% O ₂ Sat								
Art.%O ₂ Sat								
BSA								

SAMPLE ECHOCARDIOGRAM RESULTS FORM

Echocardiogram Report

Patient: _____ Age: _____
 Procedure Date: _____ ID #: _____
 Referring Physician: _____ Clinic ID: _____
 Reviewing Physician: _____ Procedure: _____
 Technician: _____ Tape Number: _____
 Echo Chart: _____

Indication:
Measurements: (Normal in Parentheses)

Estimated Ejection Fraction: _____ (55-75%)

Left Ventricular Dimensions:
 End diastole: _____ cm Septal wall: _____ cm (0.6 - 1.1 cm)
 End systole: _____ cm Posterior wall: _____ cm (0.6 - 1.1 cm)

Right Ventricular Dimensions:
 End diastole: _____ cm Lateral wall: _____ cm
 End systole: _____ cm

Aorta: _____ cm (2.0 - 3.7 cm) **Left Atrium:** _____ cm (1.9 - 4.0 cm)

Hemodynamics:
 Pulmonary acceleration time: _____ msec
 Systolic right ventricular pressure (estimated): _____
 Diastolic pulmonary pressure (estimated): _____
 Mitral inflow deceleration time: _____ msec
 Pulmonary vein "A" wave duration: _____ msec
 Pulmonary vein "A" wave velocity: _____ m/sec
 Mitral inflow "A" wave duration: _____ msec
 TR jet velocity: _____ m/sec

Findings:

Conclusions:

Ventavis is indicated for the treatment of pulmonary arterial hypertension (PAH) (WHO Group 1) to improve a composite endpoint consisting of exercise tolerance, symptoms (NYHA Class), and lack of deterioration. Studies establishing effectiveness included predominately patients with NYHA Functional Class III-IV symptoms and etiologies of idiopathic or heritable PAH (65%) or PAH associated with connective tissue disease (23%).