

PPH FORM

A. Patient Information

1. State the name of the patient (“Product Recipient”) for whom you are providing the information contained in this form.

(First Name) (Middle Initial) (Last Name)

(Date of Birth)

(Date of Diagnosis with PAH as defined in section C(1-3))

B. Physician Information

1. State your name, office address, telephone number, e-mail address, if any, and medical specialty.

(First Name) (Middle Initial) (Last Name)

(Office Address)

(City) Prov/Terr (Postal Code)

(Area Code and Telephone Number)

(Medical Specialty)

C. Primary Pulmonary Hypertension (“PPH”) Claim

Diagnosis with PAH

1. **If the Product Recipient was diagnosed with pulmonary arterial hypertension (“PAH”) prior to death, did he or she have either of the following clinical findings:**
 - (a) Mean pulmonary artery pressure of >25 mm Hg at rest or >30 mm Hg with exercise during right heart catheterization?
 Yes
 No
 - (b) Peak systolic pulmonary pressure >60 mm Hg estimated during Doppler transthoracic echocardiogram performed in accordance with the criteria set out in s.3.3.1 and 3.3.2 of the Medical Conditions List, where, in the opinion of the attending Certified Cardiologist, Pulmonologist or Respirologist, cardiac catheterization was medically contraindicated?
 Yes
 No
2. **If the Product Recipient was not diagnosed with PAH prior to death, was an autopsy performed including gross and microscopic examination of the heart and lungs conducted on the Product Recipient?**
 Yes
 No
3. **If you checked “Yes” to Question 2, did the Product Recipient’s autopsy report demonstrate histopathological changes in the lung consistent with PAH?**
 Yes
 No

Possible Alternative Causes of PAH

4. **Did the Product Recipient have a diagnosis of PAH prior to first ingestion of the Product?**
 Yes
 No
5. **Does the Product Recipient have evidence of any of the following conditions that may cause PAH? (Please check all that apply. If you need additional space in which to explain your answer please use the attached sheets and be sure to identify which question you are answering by number and letter (e.g. 5(a)).**

(a) Left ventricular failure?

Yes If yes, please specify the basis for your answer:

No If no, please specify the basis for your answer by checking the applicable boxes:

pulmonary capillary wedge pressure or left ventricular end-diastolic pressure ≤ 15 mmHg measured during the same catheterization that established PAH

in the absence of an accurate pulmonary wedge pressure or left ventricular end-diastolic pressure, left ventricular ejection fraction $\geq 60\%$ by echo or $\geq 50\%$ by MUGA and there is no Doppler evidence of elevated left ventricular end-diastolic pressures (using LV in-flow velocities and/or tissue Doppler imaging)

other: _____

(b) Valvular heart disease?

Yes If yes, please specify the basis for your answer:

No If no, please specify the basis for your answer by checking the applicable boxes:

pulmonary capillary wedge pressure or left ventricular end-diastolic pressure ≤ 15 mmHg measured during the same catheterization that established PAH.

no evidence on TTE or TEE of **moderate or greater** mitral stenosis (MVA < 2 cm or transvalvular gradient > 5 mmHg)

no evidence on TTE or TEE of **greater than moderate** aortic or mitral valvular insufficiency

other: _____

(c) congenital cardiac lesion associated with PAH? (Does not include patent foramen ovale.)

Yes If yes, please specify the basis for your answer:

No

(d) pulmonary fibrosis?

Yes If yes, please specify the basis for your answer:

No If no, please specify the basis for your answer by checking the followings boxes:

no **greater than mild to moderate** fibrosis on HRCT and total lung capacity > 80% predicted

other: _____

(e) chronic obstructive lung disease other than asthma?

Yes If yes, please specify the basis for your answer:

No If no, please specify the basis for your answer by checking the applicable boxes:

$FEV1 \leq 80\%$ predicted and $FEV1/FVC \leq 70\%$

other: _____

(f) collagen vascular disease?

Yes If yes, please specify the basis for your answer:

No If no, please specify the basis for your answer by checking the applicable followings boxes:

no clinical/serological evidence of scleroderma

no clinical/serological evidence systemic lupus erythematosus

no clinical/serological evidence of vasculitis

no clinical/serological evidence of mixed connective tissue disease

other: _____

(g) moderate to severe obstructive sleep apnea?

Yes If yes, please specify the basis for your answer:

No If no, please specify the basis for your answer by checking the followings boxes:

less than moderate obstructive sleep apnea

other: _____

(h) pulmonary thrombosis?

Yes If yes, please specify the basis for your answer:

No If no, please specify the basis for your answer by checking the followings boxes:

normal or low probability VQ scan

normal CT angiogram

normal pulmonary angiogram

other: _____

(i) Human Immunodeficiency Viral Infection (HIV)?

Yes

No

(j) Portal Hypertension?

Yes If yes, please specify the basis for your answer:

No If no, please specify the basis for your answer by checking the followings boxes:

no presence of splenomegaly, ascities, esophageal varicies

no evidence of transhepatic gradient ≥ 5 mmHg

other: _____

(k) Schistosomiasis based on exposure, clinical history and/or appropriate non-invasive, laboratory evaluation?

Yes If yes, please specify the basis for your answer:

No

(l) Living at high altitude based on exposure, clinical history and/or appropriate non-invasive, laboratory evaluation?

Yes If yes, please specify the basis for your answer:

No

(m) Ingestion of toxic rapeseed oil, amphetamines, toxic L-tryptophan, meta-amphetamines or cocaine based on exposure, clinical history and/or appropriate non-invasive, laboratory evaluation?

Yes If yes, please specify the basis for your answer:

No

(n) Sickle cell disease based on exposure, clinical history and/or appropriate non-invasive, laboratory evaluation?

Yes If yes, please specify the basis for your answer:

No

(o) Acute respiratory distress syndrome based on exposure, clinical history and/or appropriate non-invasive, laboratory evaluation?

Yes If yes, please specify the basis for your answer:

No

(p) Venous-occlusive disease based on exposure, clinical history and/or appropriate non-invasive, laboratory evaluation?

Yes If yes, please specify the basis for your answer:

No

(q) Pulmonary capillary hemangiomatosis based on exposure, clinical history and/or appropriate non-invasive, laboratory evaluation?

Yes If yes, please specify the basis for your answer:

No

(r) Mediastinal masses compressing the great vessels based on exposure, clinical history and/or appropriate non-invasive, laboratory evaluation?

Yes If yes, please specify the basis for your answer:

No

(s) Sarcoidosis based on exposure, clinical history and/or appropriate non-invasive, laboratory evaluation?

Yes If yes, please specify the basis for your answer:

No

6. If you indicated there is evidence of one or more of the conditions listed in Question 5 by answering yes to any of its the subparts, are any of those conditions the principal cause of the Product Recipient's PAH?

Yes (If yes, the Product Recipient's condition does not qualify for benefits.)

No (Please explain your answer on the attached sheet.)

D. Declaration

I declare under penalty of perjury that it is my opinion, to a reasonable degree of medical certainty, and to the best of my knowledge, that:

(a) the Product Recipient has been diagnosed with PAH;

and

(b) the Product Recipient either does or does not have any of the conditions listed in Question 4, as the case may be, and as indicated above.

(Date: MM/DD/YYYY)

(Signature of Treating Physician)

This form is an official Court document sanctioned by the Court and submitting it to the Ponderal/Redux Settlement Administrator is equivalent to filing it with a Court.