

- a. Mild PH (mPAP 21 to <25)?.....
- b. ePAH (exercise mPAP $\geq$ 30, flow <10 L/min and
mPAP-Q slope >3) (mmHg*min/L)
- c. Relative of heritable pulmonary arterial hypertension (HPAH)
- d. CTD with mild (mPAP 21 to <25) or no PH.....

7. **Newly diagnosed** with any of these WHO Group **2** comparator diagnoses?.....
 For Q7: 0=No, skip to Q8; 1=Yes, *complete Q7a-b.4*: use 0=No, 1=Yes **New Dx**
- a. Mild PVD risk associated with LHD (left heart disease with mPAP <25)?.....
- b. Moderate PVD risk associated with LHD with no PVD but meets one of the following criteria?.....
- b.1. Isolated post-capillary pulmonary hypertension (Ipc-PH) mPAP $\geq$ 25, PVR <3, DPG <7 (mmHg)?.....
- b.3. Provocable mPCW >18 following challenge, V waves < 5 mmHg, and represents significant change from resting hemodynamics? (0=No, 1=Yes).....
- b.4. Provocable mPCW >15 with large (> 5 mmHg) V waves following challenge and represents significant change from resting hemodynamics? (0=No, 1=Yes).....
8. **Newly diagnosed** with any of these WHO Group **3** comparator diagnoses?.....
 For Q8: 0=No, skip to Q9; 1=Yes, *complete Q8a-f, h-i*: use 0=No, 1=Yes
- a. Chronic Obstructive Pulmonary Disease (COPD)?.....
- b. Idiopathic Pulmonary Fibrosis (IPF)?.....
- c. Other Interstitial Lung Diseases (ILD) including CPFE and SSc-ILD?.....
- d. Obstructive Sleep Apnea (OSA)?.....
- e. Obesity Hypoventilation Syndrome (OHS)?.....
- f. Sarcoidosis? (*use codes below*).....
 0=No, 1=Yes, unknown if diagnosis was by biopsy, 2=Yes, diagnosis made by biopsy as per protocol
- h. Mild to no PVD risk (mPAP <21)?.....
- i. Moderate PVD risk (mPAP 21 to <25)?.....
- j. Moderate-significant restrictive lung physiology without ILD?.....
9. **Newly diagnosed** with this WHO Group **4** comparator diagnosis?.....
 For Q9: 0=No, skip to Q10; 1=Yes, *complete Q9a*.
- a. Chronic PE with mild to no PVD (mPAP <25)? (0=No, 1=Yes).....
10. Did final Comparator WHO Group classification change from parent study? (0=No, 1=Yes).....
- a. If Yes, what is the Primary WHO Group Comparator classification now?
 1=WHO Group 1
 2=WHO Group 2
 3=WHO Group 3
 4=WHO Group 4
 5=WHO Group 5
- b. What is the Secondary WHO Group Comparator classification now?.....

PULMONARY HYPERTENSION (PH) – Answer Q11-end of form.11. Were there **newly diagnosed** PAH conditions since the end of the PVDOMICS

(Parent) Study? (0=No, 1=Yes) _____

Any of the following PH associated medical conditions SINCE THE LAST PVDOMICS VISIT?:**WHO Group 1 Review – Pulmonary arterial hypertension (PAH)**12. **Newly diagnosed** with any of these WHO Group 1 diagnoses? _____

For Q12: 0=No; 1=Yes, complete Q12a-k: use 0=No, 1=Yes

New Dx

- a. IPAH (Idiopathic pulmonary arterial hypertension)? _____
- b. FPAH (Familial pulmonary arterial hypertension)? _____
 - b.1. Hereditary hemorrhagic telangiectasia (HHT) _____
- c. Drug-induced pulmonary arterial hypertension? _____
- d. Connective tissue disease (CTD)? _____
 - d.1. Systemic lupus erythematosus (Lupus) (SLE)? _____
 - d.2. Sjogrens? _____
 - d.3. Rheumatoid Arthritis (RA)? _____
 - d.4. Mixed connective tissue disease (MCTD)? _____
 - d.5. Systemic sclerosis (SSc)? _____
 - d.6. Antisynthetase syndrome? _____
 - d.7. CTD, undifferentiated? _____
- e. Human immunodeficiency virus (HIV) infection induced PH? _____
- f. Portal hypertension? _____
- g. Congenital heart disease (CHD)? _____
 - g.1 Was the shunt repaired? _____
- h. Schistosomiasis? _____
- i. Pulmonary-capillary hemangiomatosis (PCH)? _____
- j. Pulmonary veno-occlusive disease (PVOD)? _____
- k. Other Group 1 condition not listed above? (Specify Q12k and condition in Comments) _____

WHO Group 2 Review – PH due to Heart Disease13. **Newly diagnosed** with any of these WHO Group 2 diagnoses? _____For Q13: 0=No, skip to Q14; 1=Yes, complete Q13a-e. For Q13a-e: use 0=No, 1=Yes: **New Dx**

- a. Valvular heart disease? _____
 - a.1. Stenotic? _____
 - a.2. Regurgitant? _____
- b. Systolic heart failure? _____
- c. Heart failure with preserved ejection fraction (HFpEF)? _____

- d. Cardiomyopathy?.....
- d.1. Hypertrophic?
- d.2. Restrictive?
- e. Other Group 2 condition not listed above? (*Specify Q13e and condition in Comments*).....

WHO Group 3 Review – PH due to Lung Disease and/or Hypoxemia

14. **Newly diagnosed** with any of these WHO Group 3 diagnoses?.....
For Q14: 0=No, 1=Yes, complete Q14a-k. For Q14a-k: 0=No, 1=Yes: **New Dx**
- a. Chronic obstructive pulmonary disease (COPD)?
- b. Idiopathic pulmonary fibrosis (IPF)?
- c. Combined pulmonary fibrosis and emphysema (CPFE)?
- d. Other interstitial lung disease (ILD)?
- e. Obstructive sleep apnea (OSA)?.....
- f. Non-parenchymal restrictive lung disease (RLD)?
- g. Obesity hypoventilation syndrome (OHS)?.....
- h. Hypersensitivity pneumonitis?.....
- i. Thoracic cage abnormality?.....
- j. Cystic fibrosis?
- k. Other Group 3 condition not listed above? (*Specify Q14k and condition in Comments*)

WHO Group 4 Review

15. **Newly diagnosed** with any of these WHO Group 4 diagnoses?.....
For Q15: 0=No, 1=Yes, complete Q15a-d. **New Dx**
- a. Chronic thromboembolic pulmonary hypertension (CTEPH).....
- b. Pulmonary thromboendarterectomy (PTE).....
- c. Balloon pulmonary angioplasty (BPA)
- d. Other Group 4 condition not listed above? (*Specify Q15d and condition in Comments*)

WHO Group 5 Review

16. **Newly diagnosed** with any of these WHO Group 5 diagnoses?.....
For Q16b-d: Use 0=No, 1=Yes: **New Dx**
- a. Sarcoidosis? (*use codes below*).....
 0=No, 1=Yes, unknown if diagnosis was by biopsy, 2=Yes, diagnosis made by biopsy as per protocol
- b. Myeloproliferative Disease?
- c. Hemoglobinopathy?) (0=No, 1=Yes)
- c.1. Sickle Cell) (0=No, 1=Yes)
- c.2. Thalassemia?
- d. Other Group 5 condition not listed above? (*Specify Q16 and condition in Comments*)

17. Did final PH classification change from parent study? (0=No, 1=Yes).....

a. If yes, what is PH classification now?

1=WHO Group 1

2=WHO Group 2

3=WHO Group 3

4=WHO Group 4

5=WHO Group 5

6=Mixed (must complete Q17c-e
and Q18-Comments)If response to **Q17=6=Mixed**, please identify WHO Groups using the codes below:

1=WHO Group 1

2=WHO Group 2

3=WHO Group 3

4=WHO Group 4

5=WHO Group 5

8=Not applicable

b. What is the Primary WHO Group classification now?.....

c. What is the Secondary WHO Group classification now?.....

d. What is the Tertiary WHO Group classification now?.....

*If the status (comparator vs PH) or the WHO Group classification changed from the parent study the PI needs to provide comments below.*18. **Comments/Rationale** (required). Continue on back of next page, if needed.

199. Username of physician* reviewing/completing this form

(Username is the first 6 letters of physician's last name and first initial.)

*This should be the center PI or other study physician trained on the PVDOMICS protocol.

200. Date this form completed (mm/dd/yyyy) ____/____/____

201. Username of person reviewing completeness of this form

Clinical Center Use Only

Date form entered (mm/dd/yyyy) ____/____/____

Username of person entering this form ____