



PVDOMICS STUDY

PVD Assessment - Form #102

Instructions: This form is completed and entered for all participants. Database will skip over sections that do not apply.

For Comparators and PH Groups, complete this form after the right heart catheterization and other tests are completed. Use all medical and surgical history related to pulmonary vascular disease (PVD). Information should be obtained from medical records, local physician records, and participant interview. It should include any current diagnoses and past pertinent history.

Medical records might include, but are not limited to: hospital discharge summaries, consultation letters, MD progress notes, problem lists, medication records and imaging reports [i.e. x-ray, ultrasound, CT].

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1. Identification Number

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2. Alphacode

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3. Date of review: mm/dd/yyyy

4. Identify PVD status of this participant.....
- 1=Control, go to Q200.
2=Comparator, go to Q5.
3=PH, go to Q11.

COMPARATORS

If Q4=2-Comparator, complete the following items Q5-Q10:

5. Was this participant diagnosed with any of these WHO Group **1** comparator diagnoses?
- For Q5: 0=No, skip to Q6; 1=Yes, *complete Q5a-d.*

For Q5a-d: use 0=No, 1=Yes

- 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

6. Was this participant diagnosed with any of these WHO Group **2** comparator diagnoses?
- For Q6: 0=No, skip to Q7; 1=Yes, *complete Q6a-b2*

For Q6a-b2: use 0=No, 1=Yes

- 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)?
- Retired: Q6b.2. Provocable mPCW >18 or mPCW >15 with large (> 5 mmHg) v waves following challenge and represents significant change from resting hemodynamics?*

6b, continued,

6.b.3. Provocable mPCW >18 following challenge, V waves < 5 mmHg, and represents significant change from resting hemodynamics? (0=No, 1=Yes) _____

6.b.4. Provocable mPCW >15 with large (> 5 mmHg) V waves following challenge and represents significant change from resting hemodynamics? (0=No, 1=Yes) _____

7. Was this participant diagnosed with any of these WHO Group 3 comparator diagnoses? _____
For Q7: 0=No, skip to Q8; 1=Yes, complete Q7a-f, h-i.

For Q7a-g: 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

g. Mild to no PVD risk (mPAP <21) or Moderate PVD risk (mPAP 21 to <25) (question retired as of 01/10/2019)

h. Mild to no PVD risk (mPAP <21)? _____

i. Moderate PVD risk (mPAP 21 to <25)? _____

j. Moderate-significant restrictive lung physiology without ILD? _____

8. Was this participant diagnosed with this WHO Group 4 comparator diagnosis? _____
For Q8: 0=No, skip to Q9; 1=Yes, complete Q8a.

a. Chronic PE with mild to no PVD (mPAP <25)? (0=No, 1=Yes) _____

9. What is the Primary WHO Group Comparator classification? _____

1=WHO Group 1

3=WHO Group 3

5=WHO Group 5

2=WHO Group 2

4=WHO Group 4

10. What is the Secondary WHO Group Comparator classification? _____

1=WHO Group 1

3=WHO Group 3

5=WHO Group 5

2=WHO Group 2

4=WHO Group 4

8=Not applicable

*If Q5-10 were answered, skip to Q19 – Comments for Adjudication Committee***PULMONARY HYPERTENSION (PH) – Answer Q11-end of form.**

11. a. How was participant diagnosed with pulmonary hypertension (PH) at time of enrollment (Form 100 enroll date)? _____

1=Diagnosed by right heart cath (RHC)

2=Diagnosed by testing but not including RHC

3=PH suspected

b. Was a WHO PH Group previously identified for this participant? _____
(0=No, skip to 11d, 1=Yes, complete Q11c)

5/17/2018 Q12d7-undifferentiated CTD added. 06/28/2018 Q14j-cystic fibrosis. 01/10/2019: Q7g. Mild to no PVD risk (mPAP <21) or Moderate PVD risk (mPAP 21 to <25) split into two separate questions. Sarcoid Q7 and 16a response codes added-1=Yes, unknown if diagnosis was by biopsy, 2=Yes, diagnosed by biopsy as per protocol; Q9 & 10: Comparator codes added: 5=WHO Group 5; DCC only 'pure' inquiry number. 04/22/2020: added: Note: for the purposes of this study, PVDOMICS is not differentiating between Familial PAH (FPAH) and Heritable PAH (HPAH). 8/3/2021: "Other" added as Q12k, Q13e, Q14k, Q15d, Q16d.

WHO Group 2 Review – PH due to Heart Diseases

13. Was this participant 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:

- a. Valvular heart disease?
- a.1. Stenotic?.....
- a.2. Regurgitant?
- b. Systolic heart failure?
- c. Heart failure with preserved ejection fraction?.....
- 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. Was this participant diagnosed with any of these WHO Group 3 diagnoses?.....

For Q14: 0=No, skip to Q15; 1=Yes, *complete Q14a-k*.

For Q14a-k: 0=No, 1=Yes:

- 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. Was this participant diagnosed with any of these WHO Group 4 diagnoses?.....

For Q15: 0=No, skip to Q16; 1=Yes, *complete Q15a-d*.

5/17/2018 Q12d7-undifferentiated CTD added. 06/28/2018 Q14j-cystic fibrosis. 01/10/2019: Q7g. Mild to no PVD risk (mPAP <21) or Moderate PVD risk (mPAP 21 to <25) split into two separate questions. Sarcoid Q7 and 16a response codes added-1=Yes, unknown if diagnosis was by biopsy, 2=Yes, diagnosed by biopsy as per protocol); Q9 & 10: Comparator codes added: 5=WHO Group 5: DCC only 'pure' inquiry number. 04/22/2020: added: Note: for the purposes of this study, PVDOMICS is not differentiating between Familial PAH (FPAH) and Heritable PAH (HPAH). 8/3/2021: "Other" added as Q12k, Q13e, Q14k, Q15d, Q16d.

For Q15: 0=No, skip to Q16; 1=Yes, *complete Q15a-d.*

- a. Chronic thromboembolic pulmonary hypertension (CTEPH) (0=No, 1=Yes).....
- b. Pulmonary thromboendarterectomy (PTE) (0=No, 1=Yes).....
- c. Balloon pulmonary angioplasty (BPA) (0=No, 1=Yes).....
- d. Other Group 4 condition not listed above? (*Specify Q15d and condition in Comments*) (0=No, 1=Yes)....

WHO Group 5 Review

16. Was this participant diagnosed with any of these WHO Group 5 diagnoses?.....

For Q16: 0=No, skip to Q17; 1=Yes, *complete Q16a-d.*

For Q16a-d: Use 0=No, 1=Yes:

- 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?
c.1. Sick Cell?
c.2. Thalassemia?
- d. Other Group 5 condition not already listed? (*specify Q16d with condition in Comments*)

17. What WHO Group does the PVDOMICS Study Team think this participant falls

into **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 Q17a-c, Q18a-e, and Q19-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

- a. What is the Primary WHO Group classification?.....
- b. What is the Secondary WHO Group classification?.....
- c. What is the Tertiary WHO Group classification?.....

If Q17 **a and b** = WHO Groups 1 and/or 2 and/or 3, complete Q18:

Note: Participant can only be classified once in Q18a-e:

- 18. a. Is this participant's PH considered Mixed WHO Groups 2 & 3 as defined below (0=No, skip to 18c, 1=Yes)

5/17/2018 Q12d7-undifferentiated CTD added. 06/28/2018 Q14j-cystic fibrosis. 01/10/2019: Q7g. Mild to no PVD risk (mPAP <21) or Moderate PVD risk (mPAP 21 to <25) split into two separate questions. Sarcoid Q7 and 16a response codes added-1=Yes, unknown if diagnosis was by biopsy, 2=Yes, diagnosed by biopsy as per protocol); Q9 & 10: Comparator codes added: 5=WHO Group 5: DCC only 'pure' inquiry number. 04/22/2020: added: Note: for the purposes of this study, PVDOMICS is not differentiating between Familial PAH (FPAH) and Heritable PAH (HPAH). 8/3/2021: "Other" added as Q12k, Q13e, Q14k, Q15d, Q16d.

Definition:•PCW > 18, LAH, LAE, atrial fib **AND**

•COPD, IPF, SSc-ILD, OHS, OSA, non-parenchymal restrictive lung diagnosis or CPFE

b. What is mixed WHO Group for 2 and 3? _____

1=WHO 2, 3: acute worsening in PCW with fluid challenge, exercise, pulmonary vasodilator

2=WHO 3, 2: acute worsening of oxygenation or only mild elevation of PCW

c. Is this participant's PH considered WHO 1 & 3 as described below (0=No, 1=Yes) _____

Features of WHO 1: i.e,

• autoimmune serology, positive family history **AND**

• mild (COPD, IPF, SSc-ILD, OHS, OSA, non-parenchymal restrictive lung diagnosis, CPFE); ie. evidence of progressive PH in response to activity and/or over time with stability of hypoxemia or mild parenchymal lung disease

d. Is this participant's PH considered WHO 1, 2 as described below (0=No, 1=Yes) _____

• WHO 1, 2: PAH (WHO 1, autoimmune serology) with provocation elevation in PCW

e. Is this participant's mixed PH described as 'other'? (0=No, 1=Yes) _____

Please provide rationale for decision in Comments below.**19. Comments/Rationale for Adjudication Committee (required).** *Continue on 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 RHC 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 _____

DCC Only: For PH and Comparator WHO Groups:

DCC Inq # of 'pure' WHO Group assigned but has other 'mixed' WHO Group condition(s):..... _____

DCC Comments

5/17/2018 Q12d7-undifferentiated CTD added. 06/28/2018 Q14j-cystic fibrosis. 01/10/2019: Q7g. Mild to no PVD risk (mPAP <21) or Moderate PVD risk (mPAP 21 to <25) split into two separate questions. Sarcoid Q7 and 16a response codes added-1=Yes, unknown if diagnosis was by biopsy, 2=Yes, diagnosed by biopsy as per protocol); Q9 & 10: Comparator codes added: 5=WHO Group 5: DCC only 'pure' inquiry number. 04/22/2020: added: Note: for the purposes of this study, PVDOMICS is not differentiating between Familial PAH (FPAH) and Heritable PAH (HPAH). 8/3/2021: "Other" added as Q12k, Q13e, Q14k, Q15d, Q16d.