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The 6th World Symposium on PH: Hemodynamic Definitions and Updated Clinical Classification of PH (Part 1)

Oct 30, 2019 | [Saurabh Rajpal, MD, FACC](#)

Expert Analysis

Note: This is **Part 1** of a 6-part series on the 6th World Symposium on PH.

Part 1 | [Part 2](#) | [Part 3](#) | [Part 4](#) | [Part 5](#) | [Part 6 \(Coming Soon!\)](#)

The 6th World Symposium on Pulmonary Hypertension proposed some fundamental changes to the hemodynamic and clinical classification of pulmonary hypertension (PH). We have summarized these changes below.

Change in Criteria for Diagnosis of PH

Based on the mean pulmonary artery pressure (mPAP) of 20 mmHg being 2 standard deviations above the mean value of 14.0 ± 3.3 mmHg, which was the normal value of mPAP observed in recent published data, and the fact that there have been multiple studies across different clinical types of PH documenting poor outcomes in patients with mPAP between 20 and 25 mmHg, it was proposed **to define PH as mPAP >20 mmHg** instead of mPAP ≥ 25 mmHg at rest, measured by right heart catheterization.

Pulmonary Vascular Resistance (PVR) Added to Definition of all Forms of Pre-Capillary PH

To identify pre-capillary PH as representative of pulmonary vascular disease, it was proposed to include PVR of ≥ 3 Wood units in the definition of all forms of pre-capillary PH associated with mPAP >20 mmHg.

Combined Pre- and Post-Capillary PH

The cutoff of ≥ 3 Wood units was also proposed to identify the group of patients that has combined pre- and post-capillary PH in addition to the mandatory pulmonary capillary wedge pressure of >15 mmHg and mPAP >20 mmHg, which is a prerequisite to be classified as having any type of post-capillary PH. This group of patients has worse prognosis compared with other patients with post-capillary PH.

Exercise PH

A definition for exercise PH could not reintroduced. The challenges of getting accurate right heart catheterization measurements and the lack of diagnostic discrimination power whether exercise PH is due to elevated pulmonary capillary wedge pressure (left heart disease) or pulmonary vascular disease makes it difficult to define exercise PH as an entity.

Drug- or Toxin-Associated PH

Drugs and toxins were divided into those with definitive association (epidemiologic outbreaks, case control and randomized control trials; e.g., dasatinib and methamphetamines) and those with possible association (e.g., cocaine).

Pulmonary Arterial Hypertension Long-Term Responders to Calcium Channel Blockers

These patients were introduced as a distinct group within Group I because they have been shown to have significantly better prognosis, unique management, and different pathophysiology. These patients are defined by a reactive vasodilators stress (a reduction of mPAP ≥ 10 mmHg to reach an absolute value of mPAP ≤ 40 mmHg with an increased or unchanged cardiac output) and a sustained hemodynamic response a year after being on calcium channel blockers and New York Heart Association Functional Class I/II.

Pulmonary Veno-occlusive Disease/Pulmonary Capillary Hemangiomatosis

These were defined as pulmonary arterial hypertension with overt features of venous/capillaries involvement was included in World Health Organization (WHO) Group I because their clinical presentation and hemodynamic profile are similar to PH.

WHO Group 4 Updates: PH due to Pulmonary Artery Obstructions

In addition to chronic thromboembolic PH, the entities included in this group were expanded to include other etiologies of pulmonary artery obstruction such as congenital pulmonary artery stenosis (Alagille syndrome) and malignant and non-malignant tumors.

WHO Group 5 Simplified

In the WHO Group 5 group, splenectomy and thyroid disease was removed as a subgroup and is considered as more of a risk factor for PH. Lymphangi leiomyomatosis was reclassified as WHO Group 3 because almost all cases of PH in lymphangi leiomyomatosis are associated with only mild PH but severe pulmonary parenchymal disease.

Table 1: Updated Hemodynamic Definition of PH

Definitions	Characteristics
Pre-Capillary PH	mPAP >20 mmHg
	Pulmonary artery wedge pressure ≤15 mmHg
	PVR ≥3 Wood units
Isolated Post-Capillary PH	mPAP >20 mmHg
	Pulmonary artery wedge pressure >15 mmHg
	PVR <3 Wood units
Combined Pre- and Post-Capillary PH	mPAP >20 mmHg
	Pulmonary artery wedge pressure >15 mmHg
	PVR ≥3 Wood units

Modified from Simonneau et al.¹

References

1. Simonneau G, Montani D, Celermajer DS, et al. Haemodynamic definitions and updated clinical classification of pulmonary hypertension. *Eur Respir J* 2019;53:1801913.

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