Management of Crashing Patients with Pulmonary Hypertension

John C. Greenwood, MD[a,*], Ryan M. Spangler, MD[b]

KEYWORDS

• Pulmonary hypertension • Right ventricular failure • Cardiogenic shock

KEY POINTS

- Management goals for patients with pulmonary hypertension (PH) are to optimize preload and volume status, maintain right ventricular function, prevent right coronary artery malperfusion, reduce right ventricular afterload, and reverse the underlying cause whenever possible.
- Right ventricular failure is a hallmark finding in patients with decompensated PH.
- The bedside echocardiogram is the most useful tool when evaluating patients with PH and suspected right heart failure.
- Atrial fibrillation, atrial flutter, and atrioventricular nodal reentrant tachycardia are the most common dysrhythmias in patients with PH. Rhythm control is preferred over rate control in patients with severe PH.
- Unmonitored continuous fluid administration should be avoided in patients with PH because it often worsens pressure overload of the right heart.

INTRODUCTION

Critically ill patients with pulmonary hypertension (PH) often seem well, but they can decompensate dramatically in a short time. PH has several causes, classes, and complications; but the natural progression eventually leads to right ventricular (RV) failure, which can be extraordinarily difficult to manage. The purpose of this review is to discuss the causes, signs, and symptoms of PH as well as its management strategies and emergent complications. Treatment options are often limited, so it is imperative

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[a] Department of Emergency Medicine, Ground floor, Ravdin Hospital of the University of Pennsylvania, 3400 Spruce Street, Philadelphia, PA 19104, USA; [b] Department of Emergency Medicine, University of Maryland School of Medicine, 110 South Paca Street, 6th Floor, Baltimore, MD 21201, USA
* Corresponding author.
E-mail address: johncgreenwood@gmail.com

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that the emergency department (ED) physician can recognize and manage these patients in a timely fashion.

CLASSIFICATION

PH is defined by an elevated mean pulmonary artery pressure (PAP) (≥25 mm Hg) during right heart catheterization (normal, 14–20 mm Hg).[1,2] The first classification of PH was published by the World Health Organization (WHO) in 1973. The classification has been revised several times since then. The current structure, with 5 groups (**Table 1**), is based on cause, physiology, pathology, and treatment (**Fig. 1**).

Group 1 PH is defined by a pulmonary wedge pressure of 15 mm Hg or less, indicating isolated pulmonary arterial hypertension (PAH) and normal left ventricular (LV) function.[2] This group has a much lower prevalence than the others.[3] In 1970, the histologic differences between this group and the others were delineated.[4] The difference is thought to be the result of a variety of homeostatic imbalances related to vasoactive chemicals, growth factors, and prothrombotic and antithrombotic

Table 1
Current classification of PH

Group 1	PAH 1.1 Idiopathic PAH 1.2 Heritable PAH 1.3 Drug and toxin induced 1.4 Associated with connective tissue disorders, HIV infection, portal hypertension, congenital heart disease, schistosomiasis
Group 1′	Pulmonary venoocclusive disease and/or pulmonary capillary hemangiomatosis
Group 2	PH caused by left heart disease 2.1 Left ventricular systolic dysfunction 2.2 Left ventricular diastolic dysfunction 2.3 Valvular disease 2.4 Congenital/acquired left heart inflow/outflow tract obstruction and congenital cardiomyopathies
Group 3	PH caused by lung diseases and/or hypoxia 3.1 Chronic obstructive pulmonary disease 3.2 Interstitial lung disease 3.3 Other pulmonary diseases with mixed restrictive and obstructive pattern 3.4 Sleep-disordered breathing 3.5 Alveolar hypoventilation disorders 3.6 Chronic exposure to high altitude 3.7 Developmental lung diseases
Group 4	Chronic thromboembolic PH
Group 5	PH with unclear multifactorial mechanisms 5.1 Hematologic disorders: chronic hemolytic anemia, myeloproliferative disorders, splenectomy 5.2 Systemic disorders: sarcoidosis, pulmonary histiocytosis, lymphangioleiomyomatosis 5.3 Metabolic disorders: glycogen storage disease, Gaucher disease, thyroid disorders 5.4 Others: tumoral obstruction, fibrosing mediastinitis, chronic renal failure, segmental PH

Abbreviations: HIV, human immunodeficiency virus; PAH, pulmonary arterial hypertension.