Pulmonary Hypertension and Right Ventricular Failure in Emergency Medicine

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Pulmonary hypertension is a hemodynamic condition, defined as a mean pulmonary artery pressure by right-sided heart catheterization of at least 25 mm Hg at rest. It is classified into 5 general groups based on the underlying cause, with left ventricular failure and chronic obstructive pulmonary disease being 2 of the most common causes in the United States. Although the specifics of the pathophysiology will vary with the cause, appreciating the risks of pulmonary hypertension and right ventricular failure is critical to appropriately evaluating and resuscitating pulmonary hypertension patients in the emergency department (ED). Patients may present to the ED with complaints related to pulmonary hypertension or unrelated ones, but this condition will affect all aspects of care. Exertional dyspnea is the most common symptom attributable to pulmonary hypertension, but the latter should be considered in any ED patient with unexplained dyspnea on exertion, syncope, or signs of right ventricular dysfunction. Patients with right ventricular failure are often volume overloaded, and careful volume management is imperative, especially in the setting of hypotension. Vasopressors and inotropes, rather than fluid boluses, are often required in shock to augment cardiac output and reduce the risk of exacerbating right ventricular ischemia. Intubation should be avoided if possible, although hypoxemia and hypercapnia may also worsen right-sided heart function. Emergency physicians should appreciate the role of pulmonary vasodilators in the treatment of pulmonary arterial hypertension and recognize that patients receiving these medications may rapidly develop right ventricular failure and even death without these therapies. Patients may require interventions not readily available in the ED, such as a pulmonary artery catheter, inhaled pulmonary vasodilators, and mechanical support with a right ventricular assist device or extracorporeal membrane oxygenation. Therefore, early consultation with a pulmonary hypertension specialist and transfer to a tertiary care center with invasive monitoring and mechanical support capabilities is advised. [Ann Emerg Med. 2015;66:619–628.]

A **podcast** for this article is available at www.annemergmed.com.

0196-0644/$-see front matter

http://dx.doi.org/10.1016/j.annemergmed.2015.07.525

INTRODUCTION

During the last 30 years, physicians have been increasingly recognizing the risks of pulmonary hypertension and right ventricular failure, with an increasing focus on pulmonary hypertension research and clinical considerations.[1] Quantifying the burden of pulmonary hypertension is difficult because it is a heterogeneous condition, with demographics varying according to the underlying cause. The most common cause of pulmonary hypertension in the United States is left-sided heart failure, but many cases of pulmonary hypertension remain undocumented.[2] Pulmonary arterial hypertension, a distinct category of pulmonary hypertension described below, is a relatively rare disease, with estimates of 5 to 15 cases per 1 million adults,[3] although a recent study estimates more than 64,400 ED visits for it in the United States in a 5-year period.[4] There are few studies of assessment or treatment of pulmonary hypertension or right ventricular failure in the emergency department (ED), with only a few case reports[5,6] and observational studies,[7-9] but no randomized controlled trials, to our knowledge.

Although dyspnea is one of the most common chief complaints in the ED,[10] pulmonary hypertension has not traditionally been considered on the differential diagnosis.[8] Numerous high-quality studies,[11-13] reviews,[14-16] and clinical policies[17,18] have been developed for assessment and management of the patient with acute decompensated heart failure in the ED, but these primarily focus on the left side of the heart. This article will emphasize the importance of the right ventricle and describe the unique pathophysiology and treatment of pulmonary hypertension and right ventricular failure in patients presenting to the ED.

The pulmonary circulation is a low-pressure, low-resistance system, with thin-walled vessels and a large reserve of unperfused vessels. The right ventricle is typically a thin-walled structure[19] and can accommodate large changes in volume, or preload, but acutely has a limited contractile reserve to tackle increased impedance to ejection, or afterload.[20] The time course and degree of pulmonary vascular resistance affect the clinical severity of pulmonary hypertension.[21] Increased pressure in the pulmonary system

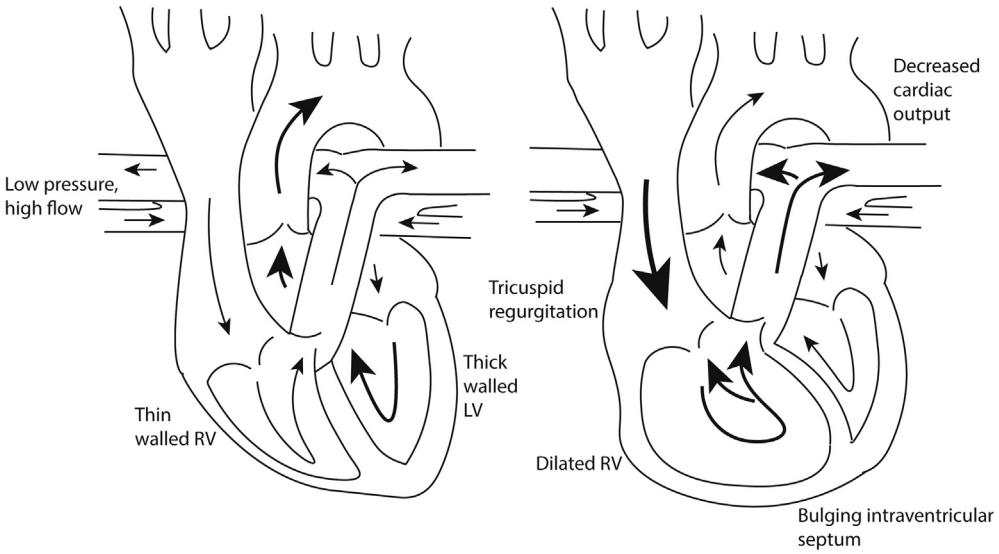

Figure 1. Normal circulation (left) and changes in pulmonary hypertension (right). *RV*, Right ventricular.

will decrease right ventricular stroke volume and right ventricular output and will increase right ventricular volume (Figure 1). When the right ventricle is overloaded, the interventricular septum may bulge into the left ventricle (LV), leading to decreased LV filling and decreased cardiac output,[1,19,22] a relationship known as "interventricular dependence."[23] Moreover, elevated pressure and volume leading to right ventricular dilatation may increase tricuspid regurgitation,[24] further reducing cardiac output and decreasing end-organ perfusion.[25]

Normal mean pulmonary artery pressure is approximately 15 mm Hg,[1,26] and pulmonary hypertension is defined as a mean pulmonary artery pressure, measured by right-sided heart catheterization, of at least 25 mm Hg at rest.[22,27] By echocardiography, a right ventricular systolic pressure of more than 35 mm Hg suggests pulmonary hypertension, although confirmation requires invasive measurement.

The right ventricle can fail in both acute and chronic pulmonary hypertension. In patients with chronic pulmonary hypertension, pulmonary vascular resistance increases gradually, allowing the right ventricle to progressively compensate.[1,28] Normally, right ventricular ejection fraction depends on right ventricular preload and wall tension increases by the Frank-Starling mechanism. However, beyond a certain point of myocardial distention, ventricular function will fail, with a reduced cardiac output and an elevation in right ventricular filling pressure.[29,30]

Appreciating coronary blood flow is also crucial to understanding the risk of pulmonary hypertension. Unlike the LV, which is fed by the coronaries during diastole, the right ventricle is perfused during both diastole and systole because of low right ventricular wall tension. In pulmonary hypertension, right ventricular perfusion by the coronary arteries decreases in proportion to increases in right ventricular pressure. If the pulmonary artery pressure surpasses the systemic pressures, the right ventricle cannot be well perfused and will become ischemic,[21,24] further diminishing right ventricular contractility and worsening right ventricular overload, beginning a catastrophic spiral (Figure 2). Because these patients have little physiologic reserve, any superimposed illness, hypo- or hypervolemia, tachyarrhythmias, or changes in oxygenation or ventilation may upset their homeostatic balance, precipitating acute on chronic right ventricular failure.[31,32]

Pulmonary hypertension is divided into 5 classifications, World Health Organization (WHO) groups 1 to 5, based on underlying cause.[33] Recognizing the various origins of pulmonary hypertension is important in the ED because not all patients with pulmonary hypertension are treated in the same manner (Table 1). However, an individual may have multiple factors contributing to development of pulmonary hypertension.

Pulmonary arterial hypertension is a group of diseases characterized by vascular remodeling of the small pulmonary arteries, with abnormal proliferation of the vascular smooth muscle and endothelial cells, inflammation, and fibrosis,[34] and is a progressive and fatal disease if untreated. It is idiopathic, formerly known as "primary pulmonary hypertension" or associated with other disease states, including connective tissue diseases, drug or toxin exposures,[3] and others.[13,34] Specific medical therapies for pulmonary hypertension are approved only for patients with group 1 disease, although pulmonary hypertension specialists may prescribe these medications for other patients as well.