The Right Ventricle in Pulmonary Hypertension

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Abstract

The main cause of morbidity and mortality in pulmonary arterial hypertension (PAH) is not injury to the lung, but rather progressive right ventricular (RV) dysfunction. Although the pulmonary vasculopathy associated with PAH can impair gas exchange, patients with PAH suffer most from the inability to adequately increase cardiac output, especially during exercise. RV dysfunction in PAH is typically viewed as a secondary event, an unavoidable consequence of the progressive rise in pulmonary vascular resistance. However, the RV response to increased pulmonary afterload varies considerably between individuals, and those variations may significantly impact functional capacity and survival.

This chapter explores the adaptive and maladaptive responses of the RV to elevated pulmonary arterial pressure (PAP) and the cellular and molecular mechanisms responsible for them. The effect of RV overload on left ventricular (LV) function is also reviewed. New techniques used to monitor RV function at baseline and in response to therapy are presented. Finally, an overview of the management of RV failure in PAH is provided along with a discussion of future therapeutic approaches.

Key Words: right ventricle; right heart; pulmonary hypertension; pulmonary circulation; cardiac hypertrophy; echocardiography; magnetic resonance imaging; natriuretic peptides.

1. THE NORMAL RIGHT VENTRICLE AND PULMONARY CIRCULATION

The right ventricle (RV) differs substantially from the left ventricle (LV) in shape and construction, no doubt due in large part to the marked differences in the functional requirements of these adjacent pumps. Unlike the left chamber, which can be considered a true ventricle, the RV consists essentially of a lateral free wall that is attached, almost as a covering, to the more muscularized medial wall (the interventricular septum) of the LV (Fig. 1). Because the lateral free wall curves in parallel to the intraventricular septum, the cross-section of the RV lumen has a crescent shape as opposed to the circular shape of the LV (Fig. 1). The different shapes of the RV and LV chambers result in considerable differences between how force and wall tension are generated (Fig. 2).

The pulmonary circulation is a low-pressure circuit. Even during periods of heavy exertion, the remarkable ability of the lung to recruit partially collapsed or unused vessels results in only modest increases in PAP despite a three- to fourfold increase in cardiac output (1). As a result, mean PAP (mPAP) is only about one-sixth that of the systemic circulation at rest and one-eighth that of the systemic circulation during exercise. Unlike the muscular LV, which responds well