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PERSPECTIVE

Right heart failure: Toward a common language

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In this perspective, the *International Right Heart Foundation Working Group* moves a step forward to develop a common language to describe the development and defects that exemplify the common syndrome of right heart failure. We first propose fundamental definitions of the distinctive components of the right heart circulation and provide consensus on a universal definition of right heart failure. These definitions will form the foundation for describing a uniform nomenclature for right heart circulatory failure with a view to foster collaborative research initiatives and conjoint education in an effort to provide insight into echanisms of disease unique to the right heart.
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"The right ventricle may be said to be made for the sake of transmitting blood through the lungs, not for nourishing them."

–William Harvey, Exercitatio Anatomica de Motu Cordis et Sanguinis in Animalibus, 1628

Even as progress in understanding left heart failure has ensued unfettered, the right heart has, for many years, been relegated to a bystander chamber. This downgrade has been accepted for decades under the presumption that the right heart is a conduit structure that is a secondary actor in the interplay of heart failure, with primacy accorded to the left ventricle. Yet, the sentinel role of right heart failure in determining functional, end-organ and clinical outcomes has become the subject of increasing recent inquiry.[1] We now recognize that the right heart is structurally discrete in its anatomic, electrical and cellular configuration; develops failure often due to distinct pathobiologic pathways that are separate from the left ventricle; and plays a central role in determining prognosis even as therapeutic success in addressing left ventricular dysfunction is demonstrated.[2] Uniquely, therapy that influences the left ventricle favorably may not impact the dysfunctional right ventricle and vice versa.[3]

The right ventricle and left ventricle are anatomically, physiologically and functionally distinct. The right ventricle is anatomically composed of 3 distinct portions[4]: the first portion is the inlet, which includes the tricuspid valve, the chordae tendinae and papillary muscles; the second portion is the trabeculated apical myocardium; and finally the infundibulum or conus constitutes the outlet region. The shape of the right ventricle is complex: it appears triangular from the side, and is crescent shaped in cross-section. There are 2 layers of right ventricular myocardium. The fibers of the superficial layer of the right ventricle are arranged circumferentially in a direction that is parallel to the atrioventricular (AV) groove in continuity with the left ventricle. The deep muscle fibers of the right ventricle are longitudinally aligned base to apex (in contrast to the left ventricle where oblique fibers are found superficially, longitudinal fibers on the endocardium and circumferential fibers in between). Haddad and colleagues[4] elegantly described the morphologic differences between the left and right ventricles as follows: (1) a more apically situated hinge point of the septal leaflet of the tricuspid valve relative to the anterior leaflet of the mitral valve; (2) the presence of a moderator band in the right ventricle cavity; (3) more than 2 papillary muscles; (4) a tri-leaflet atrioventricular valve with septal attachments; (5) predominantly coarse trabeculations; and (6) a ventriculoinfundibular fold that separates the tricuspid valve from the pulmonic valve (as opposed to the aortomitral continuity seen on the left). These anatomic structural differences indicate that the

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designs are intended for distinct purposes on an evolutionary scale. Importantly, they provide insight into the biologic diversity that explains the varied phenotypic reactions to hemodynamic stressors.

Physiologically, the right ventricle is more uniquely sensitive to after-load, demonstrates a trapezoid pressure–volume curve (as opposed to a rectangular pressure volume loop for the left ventricle), and develops dysfunction by many distinct pathways.[5] In the setting of an acute increase in pulmonary arterial impedance, as encountered in pulmonary embolism, the right ventricle demonstrates evidence of a severe reduction in stroke volume with a narrow window of pressure increase.[6] When a left ventricular assist device is placed and ventricular suction applied such that the septum is moved into the left ventricular cavity, one frequently has significant right ventricular dysfunction even with reduced after-load.[7] Extrinsic compression as with pericardial constriction or effusion can impede right ventricular function by compressive dynamics and lead to manifestations of heart failure.[8] In clinical syndromes of congenital heart disease one can exhibit the syndrome of right-sided failure due to changes in flow and anatomic defects located within the right ventricle or in anatomic areas preceding blood entry into this chamber.[5] In chronically raised after-load and pulmonary impedance, the right ventricle demonstrates rather diverse responses with variable expression of dysfunction over time. Despite these complex attributes, it is clear that development of right-sided dysfunction portends a steep decline in prognosis accompanied by multisystem organ failure reflected variably in clinical expression as the cardiorenal syndrome, protein-losing enteropathy and cardiac cachexia.[9–11]

It has become obvious that the complex nature of right-sided heart failure, the diverse pathways and multispecialty involvement among distinct clinicians, such as cardiologists, pulmonologists, congenital heart disease experts and cardiothoracic surgeons, has enforced the development of varying definitions which are uniformly resident within silos and consequently cover a limited scope and clinical need. To address this impediment to research and innovation in right heart failure, leading experts from around the world in the fields of congenital heart disease, pulmonary vascular disease, congestive heart failure and cardiothoracic surgery came together under the aegis of the newly founded International Right Heart Foundation with the core mission of bringing this distinguished interdisciplinary group of expert physician scientists together to develop an integrative language that effectively captures and describes right heart disease. The development of a common language relevant to scientists and clinicians alike was designed to foster collaborative research initiatives and conjoint education in an effort to provide insight into mechanisms of disease unique to the right heart while advancing patient care.

This group reached united consensus on the basic definitions concerning the right heart and further emphasized that confusion between the commonly used nomenclature of "right ventricular failure" and "right heart failure" must be clarified as follows and not used interchangeably.

Definition 1: Distinction between right heart failure and right ventricular failure

Right heart failure represents a disturbance or dysfunction in any of the components that constitute the right heart circulatory system (defined below). Thus, right ventricular failure, in contradistinction, is one component (albeit major) of a pathophysiological entity that can result in right heart circulatory failure.

Definition 2: Components of the right heart system

The *right heart circulatory system* is comprised of the systemic veins up to the pulmonary capillaries–at which point deoxygenated blood transitions to oxygenated blood. The right heart system can be classified into systemic and pulmonary circuits. The systemic circuit includes the systemic veins, right atrium, coronary sinus (and cardiac venous drainage), tricuspid valve, right ventricular free wall, right ventricular outflow tract and pulmonic valve. The pulmonary circuit includes the main pulmonary artery post-pulmonic valve and secondary and tertiary branches of the pulmonary arteries.

The *left heart circulatory system* is comprised of the post-pulmonary capillaries to the systemic arteries–at which point oxygenated blood begins to shift to deoxygenated blood. The left heart circulatory system is comprised of the pulmonary veins, left atrium, mitral valve, left ventricle, aortic valve, aorta and systemic arteries (including the coronary arteries). The pulmonary and systemic capillary beds are shared between the two compartments on the right- and left-sided circulatory system.

Definition 3: What is right heart failure?

We define right heart failure *as a clinical syndrome due to an alteration of structure and/or function of the right heart circulatory system that leads to sub-optimal delivery of blood flow (high or low) to the pulmonary circulation and/or elevated venous pressures–at rest or with exercise*.

Distinctively, this definition is broad and classifies right heart failure as a syndrome, which may result from anatomic or physiologic aberrations, or both, from a variety of etiologies that are not restricted to the right ventricle. Importantly, this definition allows for abnormalities to manifest themselves clinically during exercise alone while remaining quiescent during resting conditions. Furthermore, we believe that it is important to allow for a broad definition that encompasses most disorders (ranging from those that *always* involve the right ventricle to those that may spare the right ventricle yet result in the manifest clinical syndrome such as a pre-tricuspid lesion). However, we do recognize that clinical exceptions to the rule always exist (e.g., occlusive disease of the inferior vena cava). Thus, it is our contention that this definition as proposed may meet the goal of widest incorporation of pathologic substrates and their clinical manifestation that influence clinical expression in the right heart circulatory system.