The Right Heart and Pulmonary Circulation

The Right Heart and Pulmonary Circulation: Is It Really a Minor Circulation?

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INTRODUCTION

Continued education has traditionally been considered to be of great importance in *Revista Española de Cardiología*. The "Editorial," "Review Article," and "Update" sections are the usual means for transmitting to readers the opinions and points of view of experts of known prestige in the different aspects of cardiovascular disease. In the case of the Update section, over the years we have attempted to select areas of obvious interest to a large part of our readers that, not being the object of attention in more classical reviews, would otherwise be overlooked. Thus, the subjects we have attempted to address in a cross-cutting manner in recent years include: "Cardiovascular disease in women" (2006),[1] "Non-coronary arterial disease" (2007),[2] "Cardiovascular prevention" (2008),[3] and "Cardiovascular translational medicine" (2009).[4]

The subject selected this year for in-depth evaluation, "The right heart and pulmonary circulation," encompasses several aspects that are undoubtedly attractive. The first is its evident implication in the practice of cardiologists of all hues, whether general or specialized, ranging from those devoted to clinical activity to basic researchers. On the other hand, after decades of highly predominant attention to the left chambers of the heart and the systemic vessels, over the last 15 years we have been witnessing a progressive awareness of the importance of right ventricle (RV) dysfunction in different heart diseases,[5] both in those cases in which this chamber plays the leading role in the condition (as in arrhythmogenic right ventricular dysplasia or certain congenital heart diseases) and in those in which it accompanies conditions involving the left heart chambers and valves, significantly modifying the clinical signs and prognosis. To paraphrase the title of a recent report, we now consider that "the left heart will only be able to work as much as the right heart lets it."[6] With respect to the pulmonary circulation, the past decade has witnessed important advances based on a more complete knowledge of the pathophysiology of the different forms of pulmonary hypertension, which has been expressed in a classification that is of evident practical utility, as well as an improved diagnostic approach and, what is more important, the possibility of employing treatments that are useful for the improvement of the symptoms and the prognosis of these patients.

Therefore, this Update arrives just in time to help us learn about the latest findings, incorporate the diagnostic and therapeutic advances into our clinical practice and become aware of the uncertainties and gaps left by our current knowledge. The editors of *Revista Española de Cardiología* can state with pride that among the authors who have agreed to contribute their knowledge to this series of monographs are the major world experts in this field. Approximately half of the reports are written by authors living in the United States, and half were submitted by specialists from different European countries.

REASONS FOR A SECULAR OVERSIGHT

As early as 1553, the Aragonese physician Miguel Servet published his pioneering theories, scientifically confirmed by the work of Harvey in the 17th century, concerning the existence of an independent pulmonary circuit for the oxygenation of the blood, which is conducted and propelled into the pulmonary circulation from the right chambers of the heart. However, the importance of the function of the right heart continued to be the subject of debate until the end of the 20th century. This chamber has traditionally been considered to play a secondary role in the circulatory system, where it was thought to enhance the capacity of the

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pulmonary circulation more than to propel the flow of blood through the lungs. This belief was founded on experimental works based on the ablation or the replacement of the free RV wall in open-chested animals, in which this maneuver produced little impact on the cardiac output.[7] This came to be confirmed decades later with the viability of those patients with congenital heart disease in whom the RV was excluded from the pulmonary circulation by means of a cavopulmonary shunt like that created in the Fontan procedure. The fact that the circulation is far from normal in both cases has been recognized only recently.[8]

On the other hand, the noninvasive study of the anatomy and function of the right chambers has classically been highly difficult, especially if compared with the easy accessibility of the left ventricle (LV) to evaluation using ultrasonographic and isotopic techniques. Moreover, the spatial configuration of the LV makes its easy to estimate its volume by means of models (such as the so-called ellipsoid of revolution) that make it possible to accurately calculate its volume on the basis of two-dimensional measurements. This has facilitated the quantification of the contractile function of the ventricle with measurements such as the LV ejection fraction, which, although it has its limitations, has enabled great advances in the classification and the study of patients with dysfunction of this chamber. In the case of the RV, the opposite occurs since, given its irregular shape, with entry route, apical region and exit route its volume is difficult to analyze on the basis of 2-dimensional measurements.[8] The situation is more or less the same when we consider the physiological aspects: the study of LV function is more accessible because its basic task is to generate high pressures (easy to estimate through the arterial pressure) by means of the contractility (estimable using parameters such as the ejection fraction). In contrast, the function of the RV is basically to maintain pulmonary flow (much more difficult to measure in practice) in a low-pressure system,[6] and the contractility of its free wall does not always reflect its function well because it varies widely depending on the loading conditions.

THE RIGHT VENTRICLE AND PULMONARY CIRCULATION IN CURRENT CARDIOLOGY

Despite the reported difficulties, the growing knowledge of our anatomy and physiology of the RV and of the mechanisms that operate in the presence of disease have brought us to grant the right heart the importance it warrants. Pulmonary circulation, must combine a low pressure (to avoid the formation of pulmonary edema through the thin and highly permeable membranes that permit gas exchange, with the capacity to absorb the entire cardiac output through a circulation characterized by very low resistance. As a consequence, the pump that propels the circulation in this circuit must basically be capable of maintaining an elevated cardiac output without generating high pressures. Thus, the RV presents anatomical characteristics (due to its shape, greater volume and the distribution of the muscle layers) and physiological features (asynchronous, "peristaltic"-like contraction) that differ widely from those of the LV. Substantial differences between RV and LV have been reported in the genes and in the molecular and cellular mechanisms that are observed under normal conditions and in the presence of disease, both in embryonic and fetal development and in the adult individual. Suffice it to say that, in recent years, the most important journals related to this area have devoted considerable space to reviewing these concepts.[9-13] The first article of this series, which the readers will find immediately following this presentation, focuses on a description of our basic knowledge of the right heart.[14]

On the other hand, the advances in cardiovascular imaging techniques have revolutionized the study of the right cardiac chambers and the pulmonary vasculature. While the three-dimensional reconstruction of ultrasonographic images can solve the problem of measuring the volume of the RV, magnetic resonance is the technique that is clearly becoming the "gold standard" in this field as it enables the accurate evaluation of anatomical and physiological parameters of the circulation in the right heart.[8,15,16] We will devote a chapter of this Update to the review of this technique.

However, the fact that perhaps has contributed the most to the present interest in the RV and pulmonary circulation is the clinical confirmation of their impact on different heart diseases. Thus, the presence of both pulmonary hypertension and RV dysfunction are common findings in "left" heart disease and act as powerful predictors of a poorer prognosis. The diagnosis and clinical management of this situation are a challenge that will affect cardiologists in the coming years[17,18] and is the subject of a chapter of this series.

Other aspects of which the clinical cardiologist also be aware are the incidence and the clinical impact of RV both in acute situations (such as RV infarction, pulmonary thromboembolism and graft dysfunction following heart transplantation) and in chronic conditions, especially valve diseases, certain congenital heart diseases and different forms of chronic pulmonary hypertension.

The strategies for the prevention and treatment of RV involvement in these situations will be of great value when put into practice.[12,13,17]

Two articles of this series will be devoted to reviewing the "state of the art" in a field as timely as that of pulmonary hypertension. The first focuses on the pathophysiological and diagnostic aspects as well as the evaluation of the prognosis, while the second provides an update on the possibilities and limitations of the different therapeutic measures developed over the past 15 years, beyond the recently published norms and clinical practice guidelines.[19,20]

Given its incidence and clinical impact, thromboembolic disease warrants particular attention. The spectrum of clinical signs is very broad since the development of pulmonary hypertension and, in advanced phases, RV dysfunction accompany the most severe acute forms. In discussing the recent publication of the European guidelines for pulmonary embolic disease,[21] we will be able to offer the comments of one of the highest authorities in this field.

In contrast to those mentioned previously, there are certain conditions in which RV is the primary substrate of the disease. These include types of cardiomyopathy that predominantly affect the chambers of the right side of the heart, in which the genesis of potentially lethal arrhythmias is not uncommon, and aggressive measures for their prevention and treatment may be required. We have increasingly greater knowledge of the genetic aspects of these diseases, which are associated with anomalies in the genes that encode the desmosomal proteins. Other arrhythmogenic heart diseases that involve the right-sided chambers also appear to have a genetic basis and will be dealt with in the same chapter.[22,23]

There are a number of connections between congenital heart diseases and conditions affecting the chambers of the right side of the heart and the pulmonary vasculatureThis subject will be approached in two separate articles: the first refers to the different forms of structural involvement of the right heart in these heart diseases, their clinical implications and their treatment,[24,25] whereas the second focuses on the subgroup of patients, frequently of adult age, with pulmonary hypertension associated with congenital shunts, a field in which the traditional absence of effective therapeutic measures has given way over the past decade to significant innovations.[26,27]

The series of reviews will close with a chapter devoted to the surgical possibilities in the treatment of right heart disease, with a greater emphasis on the controversial subject of surgery of the tricuspid valve which, despite the fact that it has been carried out for decades, has yet to be associated with an indication that is generally accepted among cardiac surgical teams.[28] Other currently performed interventions, such as the implantation of mechanical circulatory assist devices for situations of RV failure resistant to other measures, will also be discussed.

CONCLUSIONS

It is evident that we cannot ignore the basic aspects of the diagnosis and treatment of the diseases affecting what used to be referred to as the "minor circulation," since the tools that are presently available enable their evaluation and management. We hope that enumerating the different aspects encompassed in the conditions affecting the right chambers of the heart and the pulmonary vessels, given their incidence and clinical importance, has awakened reader interest. The objective of this series of Update chapters is to provide the cardiologist with the knowledge necessary to confront these aspects with confidence when they arise in his or her professional activity. Thus, we invite the readers of *Revista Española de Cardiología* to follow us month after month as we present this series, in which our guides will be experts of known worldwide prestige.

If the publication of these articles contributes to cardiologists granting diseases of the right heart and the pulmonary vasculature their proper importance in our daily practice, the effort will have had its reward: among us, it will never again be a "minor circulation."

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