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REVIEW

Quantification of systemic right ventricle by echocardiography

Quantification du ventricule droit systémique par échocardiographie

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Summary Improvements in cardiac imaging have recently focused a great interest on the right ventricle (RV). In patients with congenital heart disease, the right ventricle (RV) may support the systemic circulation (systemic RV). There are 2 different anatomic conditions providing such physiology: the congenitally corrected transposition of the great arteries (ccTGA) and the TGA surgically corrected by atrial switch. During the last decades, evidence is accumulating that progressive systemic RV failure develops leading to considerable morbidity and mortality. Various imaging modalities have been used to evaluate the systemic RV, but echocardiography is still predominantly used in clinical practice, allowing an anatomic and functional approach of the systemic RV function and the potential associated anomalies. The goal of this review is to offer a clinical perspective of the non-invasive evaluation of the systemic RV by echocardiography.

Abbreviations: 2D, two-dimensional; 2DE-KBR, 2Dechocardiography with 3D knowledge-based reconstruction; 3D, three-dimensional; ccTGA, congenitally corrected transposition of the great arteries; CHD, congenital heart disease; FAC, fractional area change; IVA, isovolumic myocardial acceleration; LV, left ventricle/ventricular; MRI, magnetic resonance imaging; RT3DE, real-time 3D echocardiography; RV, right ventricle/ventricular; RVEF, right ventricular ejection fraction; SR, strain rate; STE, speckle-tracking echocardiography; TAPSE, tricuspid annular plane systolic excursion; TDI, tissue Doppler imaging; TGA, transposition of the great arteries; TR, tricuspid regurgitation.

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Résumé Les améliorations de l'imagerie cardiaque ont permis de se focaliser récemment sur l'étude du ventricule droit. Chez certains patients porteurs d'une cardiopathie congénitale, le ventricule droit peut se retrouver en position systémique. C'est le cas des patients porteurs d'une double discordance et patients porteurs d'une transposition des gros vaisseaux opérés par un switch à l'étage atrial. Durant les dernières décennies, les données scientifiques ont montré une dégradation progressive du ventricule droit systémique, qui est associée à une morbidité et une mortalité importante. Différentes modalités d'imagerie ont été utilisées pour évaluer le ventricule droit systémique mais l'échographie reste la méthode de choix dans l'activité quotidienne, permettant une analyse anatomique et fonctionnelle. L'objectif de cette revue est d'offrir une perspective clinique de l'évaluation non invasive du ventricule droit systémique par échocardiographie.

Introduction

Improvements in cardiac imaging have recently focused great attention on the right ventricle (RV), emphasising its importance in cardiac physiology. The RV has been studied in many physiopathological conditions, such as pulmonary hypertension and heart failure related to left ventricular (LV) dysfunction.

Specific congenital heart diseases (CHD) may provide an original condition where the RV is pumping through the systemic circulation. In these anatomical and haemodynamic conditions, the RV is called systemic RV (systemic meaning pumping through the systemic circulation as opposed to the pulmonary circulation). There are two different anatomical conditions that provide such physiology.

The first condition is congenitally corrected transposition of the great arteries (ccTGA), where the left atrium is connected to the RV, pumping in the aorta, and where the right atrium is connected to the left ventricle (LV), pumping in the pulmonary artery branches. The other condition is related to atrial switch, which is the surgical treatment developed in the late 1950s by Ake Senning [1] and in the mid-1960s by William Mustard [2] for surgical correction of transposition of the great arteries (TGA). In terms of live birth incidence, TGA is the more common condition (TGA 1:3100 live births vs ccTGA 1:33,000), but the absolute number of Mustard and Senning patients is decreasing because atrial redirection procedures for TGA were superseded by the arterial switch operation in the 1980s. As a consequence, almost all patients with TGA and atrial redirection reach adulthood.

For both populations, right ventricular (RV) failure is an important long-term concern, leading to severe late complications [3—5]. Various imaging modalities have been used to evaluate the systemic RV, including angiography, radionuclide imaging and magnetic resonance imaging (MRI) [6]. Nevertheless, in clinical practice, echocardiography is still used predominantly for the assessment of RV function, as it is non-invasive, widely available, relatively inexpensive and has no adverse side effects. However, because of complex geometry, the assessment of systemic RV function by echocardiography has remained mostly qualitative. Advances in digital echocardiography allow for a more refined assessment of the RV, as demonstrated in patients with pulmonary arterial hypertension [7] and other clinical conditions. These novel echocardiography variables may also be valuable in the functional assessment of the systemic RV [8]. The goal of this review is to offer a clinical perspective of the non-invasive evaluation of the systemic RV by echocardiography.

Echocardiographic assessment of systolic function in the systemic RV

Why the RV is different

By analogy with the LV, RV ejection fraction (RVEF) is considered to be the marker of RV function. However, attempting to extend this to the RV has been problematic. The shape of the RV does not allow the use of geometric formulae to calculate the RVEF. A range of echocardiographic variables has therefore been developed to evaluate RV function in the subpulmonary position, especially simple measurements of long-axis excursion, giving rapid and unambiguous results [8—10], which has proven utility as a measure of LV function in patients with coronary artery disease, valve disease and heart failure [11,12]. The technique is equally straightforward for the RV, and is especially valid because most of the RV myocardial fibres are arranged longitudinally. These variables have been compared with other methods of calculating the RVEF, especially cardiac MRI, which is considered to be the gold standard for RV functional assessment [13]. Logically, these echocardiographic variables have been used subsequently for systemic RV functional assessment (Fig. 1).

Basic longitudinal function variables

The first studies focused on the analysis of longitudinal myocardial fibres using mainly tricuspid annular plane systolic excursion (TAPSE), based on the anatomical hypothesis that the majority of RV myocardial fibres originate at the apex of the heart and insert into the right atrioventricular junction, such that the main bulk of the RV myocardium is composed of longitudinally arranged fibres [14]. Derrick et al. showed that systemic RV long-axis function was notably reduced compared with that of either the normal subpulmonary RV or the systemic LV. The authors hypothesized that impaired systemic RV longitudinal function reflected the response of the longitudinally arranged