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EDITORIAL COMMENT

Right ventricular speckle tracking echocardiography: A new tool for decision-making after surgical repair of tetralogy of Fallot?

Ecocardiografia de *speckle tracking* do ventrículo direito: uma nova ferramenta para a tomada de decisão após a reparação cirúrgica da Tetralogia de Fallot?

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Surgical correction of tetralogy of Fallot (TOF) has evolved in recent years from correction after childhood to early surgery, even in the neonatal period. Before and after correction the right ventricle is mainly affected, experiencing persistent pressure and/or volume overload and intrinsic or acquired right ventricular (RV) myopathy. On follow-up, these patients can present RV enlargement, dysfunction and fibrosis, all with significant clinical impact, such as exercise limitation, RV failure and a substrate for lethal ventricular arrhythmias and sudden cardiac death (SCD).[1] A combination of varying degrees of long-standing pulmonary regurgitation and residual stenosis has been identified as a major issue after TOF correction that over time triggers RV remodeling, affecting ventricular synchrony and coupling and also impairing left ventricular function.[2]

In long-term follow-up, arrhythmias are a common problem in more than 30% of patients with repaired TOF, sometimes causing adverse outcomes and even SCD, which has an estimated incidence of 0.2% per year in these patients. There is thus a need for accurate risk predictors in the follow-up of TOF patients.[3] Various clinical factors have been investigated as predictors of late arrhythmic events, but they still lack strong positive predictive accuracy. This situation has prompted considerable changes in the surgical approach to TOF in recent decades. The current approach favors repair at a younger age (<2 years), efforts to preserve pulmonary valve competence and to avoid ventriculotomy, and extensive removal of infundibular muscle.[4] Nevertheless, definitive evidence on the optimal surgical strategies to protect RV function and reduce arrhythmias and risk of SCD is lacking.[5]

One of the hot topics in TOF is active long-term preservation of RV function. Ways to prevent significant pulmonary regurgitation and to avoid RV dilatation remain the main target, but the optimal timing for therapeutic intervention remains uncertain, although there is a trend towards early reintervention and acceptance of certain approaches is emerging.[6,7] Careful imaging surveillance of RV size and function is essential to determine optimal timing of additional corrective interventions.[7] Which imaging parameters are the most reliable for decision-making in clinical assessment remains a crucial question.

Although there have been many advances in noninvasive RV imaging during the past decade, echocardiography

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remains the most accessible and easiest method to assess anatomical anomalies and RV volumes and function during follow-up. However, this technique for assessing subclinical RV dysfunction has limited accuracy because of its inability to determine the geometric shape of the entire ventricle, the load dependency of the measured parameters, and the lack of specialized software for analyzing the right ventricle. For these reasons cardiac magnetic resonance (CMR) is considered the gold-standard tool to assess RV anatomy and function in both acquired and congenital heart disease (CHD), when accuracy matters.[8]

In 2010, the American Society of Echocardiography developed guidelines for quantitative assessment of RV function in adults, providing standardized measures for serial follow-up and aiming to improve the accuracy and reproducibility of echocardiographic imaging.[9] Strain and strain rate measurements obtained by tissue Doppler imaging (TDI) and two-dimensional (2D) speckle-tracking echocardiography (STE) have emerged in the last decade as tools to assess RV myocardial deformation and to detect subclinical myocardial dysfunction in patients with normal conventional ventricular function indices, and before RV ejection fraction decreases.[9,10] An increasing number of publications support 2D-STE as a primary modality for quantitative assessment of myocardial deformation in different clinical settings, including repaired TOF. Notwithstanding, it is still a research tool and needs further validation and normative values in CHD patients before it can be integrated into everyday clinical practice.[10–13]

In this issue of the *Journal*, Timóteo et al.[14] present a retrospective study assessing 2D-STE right atrial (RA) and RV peak systolic longitudinal strain and conventional echocardiographic parameters of RV function as predictors of arrhythmias in a cohort of adult patients with repaired TOF. They compare data from two groups of TOF patients, those with and without arrhythmic events, in order to determine whether RV and/or RA speckle tracking strain are associated with previous arrhythmic events. They found that patients with arrhythmias were older, surgical correction was performed later in life, and RA and RV strain were significantly reduced. Age at correction and the presence of residual defects were the only independent predictors for arrhythmias. RA and RV strain measurements were significantly decreased, but they failed to establish strain as an independent predictor for arrhythmias in the overall study group. This was overcome by performing a subanalysis after excluding patients with a second surgical correction. They conclude that strain measurements of the right heart may be useful for risk stratification of arrhythmias in patients with repaired tetralogy of Fallot, although these data need to be supported by more extensive prospective data.

Overall, the findings of this study corroborate a previous pilot study by the same group and are in line with other studies addressing tachyarrhythmia risk predictors in operated TOF patients. Of note, the studied patients with arrhythmias were older (12 years older than those without arrhythmias), and were operated later in life (twice the age of those without arrhythmias), reflecting the expected evolution of the disease in a longer follow-up and the use of older surgical strategies. Additionally, these patients may also present structural abnormalities of the RV myocardium, such as fibrosis, which can trigger RV dysfunction and arrhythmias. Investigation of these anomalies using late enhancement CMR could add interesting data on the studied population.

The authors also studied RV function using conventional echocardiographic measurements and found that neither RV fractional area change nor tricuspid annular plane systolic excursion (TAPSE) predicted the presence of arrhythmias. Both are simple routine measures of RV systolic function, but they do not take into account the global geometry of the RV, as the authors point out. Three-dimensional (3D) echocardiography was expected to overcome some of these issues. Dragulescu et al.[15] demonstrated that 3D echocardiography provided more accurate measurements of RV function and a better correlation with CMR in repaired TOF patients. They also showed that TAPSE was useful as an indicator of RV pressure and volume overload, with a good correlation with RV ejection fraction on CMR.[15] Nevertheless, 3D echocardiography is time-consuming and when image quality is suboptimal may lead to incomplete data sets, as the technique requires specialized software and training.

RV peak systolic longitudinal strain is an index of RV myocardial function, but its measurement in clinical practice has been hampered by a lack of uniformity in software, methods and reference values. Recently, a large study provided reference values and methods adjusted for demographics and gender in healthy subjects.[16] These authors also demonstrated that LV and RA longitudinal strain were correlates of RV longitudinal strain, suggesting that 2D-STE is able to assess the functional coupling of the right ventricle with the left ventricle and the right atrium.[16] In the present study, the addition of LV longitudinal strain analysis could have shed light on the relationships governing ventricular coupling in these patients. It would also be interesting to analyze the presence of RV dyssynchrony and its relation with arrhythmic events, although Moon et al. failed to demonstrate differences in ventricular dyssynchrony in TOF patients with decreased longitudinal strain of both ventricles, which was associated with arrhythmic events.[17]

The use of RV longitudinal strain as a parameter of systolic dysfunction in TOF patients should, however, be treated with caution, because it only measures the shortening of the ventricle along the base-to-apex direction, reflecting the contraction of longitudinal myocardial fibers. Studies have demonstrated that RV ejection fraction depends on multiple mechanisms which include the movement of the RV free wall, the involvement of the interventricular septum during contraction and the circumferential contraction of the right outflow tract, each of which makes varying contributions under different conditions. Repaired TOF patients with pulmonary regurgitation may present some degree of outflow tract obstruction or hypertrophy, which causes RV myocardial remodeling that favors circumferential displacement of myocardial fibers, and for this reason RV longitudinal strain lacks accuracy to measure global RV function.[18]

Finally, the study by Timóteo et al.[14] adds important evidence on the usefulness of 2D-STE-derived longitudinal strain as a superior marker of early myocardial dysfunction compared to conventional echocardiographic measurements of RV function. However, the importance of these changes in clinical practice and their value as predictors of late arrhythmic events in patients with repaired TOF are still to be determined.

Speckle tracking echocardiography is here to stay; extending its use to CHD may enhance our understanding of long-term functional changes in these patients. Our ability to predict deterioration of ventricular function in patients with repaired TOF, as in other operated CHD patients, remains limited, since we still do not understand why RV function deteriorates in some patients while in apparently similar patients it remains stable. There are likely to be unknown factors, such as genetic phenotype, that play critical roles in mediating deterioration of the disease, and this requires further investigation.

Conflicts of interest

The author has no conflicts of interest to declare.

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