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EDITORIAL

Usefulness of strain imaging echocardiography in arrhythmogenic right ventricular cardiomyopathy: clinical perspective

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Abstract

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is a rare, but severe disease that predisposes patients to ventricular arrhythmias, heart failure, and increased risk of sudden cardiac death. Because of the broad spectrum of phenotypic expressions, diagnostic assessment is often challenging. Speckle-tracking echocardiography has emerged as an alternative technique enhancing the performance of standard echocardiography in ARVC patients, leading to an early detection of subtle ventricular abnormalities and potentially identifying predictors of adverse clinical outcome.

Keywords ARVC · Transthoracic echocardiography · Speckle tracking echocardiography · Myocardial strain

This letter refers to the article "Echocardiographic assessment of right ventricular function: current clinical practice" by Schneider et al. [1], published in Int J Cardiovasc Imaging (2019), with the aim to further underline the promising strength of right ventricle (RV) analysis in clinical practice in subjects affected by ARVC, a rare, but severe disease.

ARVC is an inherited cardiomyopathy, pathologically characterized by the replacement of the cardiac myocytes, of the right and even the left ventricle, with fibro-fatty tissue. This feature predisposes patients to ventricular arrhythmias, heart failure, and increased risk of sudden cardiac death (SCD) [2]. Because of the broad spectrum of phenotypic expressions, diagnostic assessment is often challenging [2].

Since ARVC is frequently marked by a poor outcome, an early correct diagnosis of the index cases and the mutation carriers is critical. Diagnostic criteria may fall in the early stages of the disease when no ECG change occur and the identification of RV abnormalities using echocardiography may be hard due to the subtle abnormalities and the patchy involvement of RV wall.

Moreover, conventional echocardiographic parameters such as the tricuspid annular plane systolic excursion, peak systolic RV annular velocity (s'RV), and RV-fractional area change (RV-FAC) or an excessive ventricular trabeculation lack of specificity.

Speckle-tracking echocardiography (STE; Fig. 1) has emerged as an alternative technique that measures myocardial deformation guaranteeing unique information in detecting clinical and subclinical myocardial dysfunction in different setting such as ischemic heart disease, cardiomyopathies and heart failure.

Its role has been also recently established in the multimodality imaging approach in ARVC [2]. Increasing studies revealed how STE may enhance the performance of echocardiography in ARVC patients, overcoming the previously mentioned limitations of conventional RV echocardiography leading to an early detection of subtle ventricular abnormalities and potentially identifying predictors of adverse clinical outcome.

Since fibro-fatty replacement of the myocardium often precede wall motion impairment, RV peak systolic strain seems to be a reliable measurement of impaired wall motion and has high diagnostic value especially in the absence of RV dilatation, visual wall motion abnormalities and a reduced global RV systolic function. A reduction of both, RV global longitudinal strain and free wall strain has been reported in the early phases of ARVC.

A cut-off value of − 18% peak systolic strain has been proposed to differentiate between normal and abnormal RV segments for identifying ARVC patients with a high diagnostic accuracy in terms of sensitivity and specificity.

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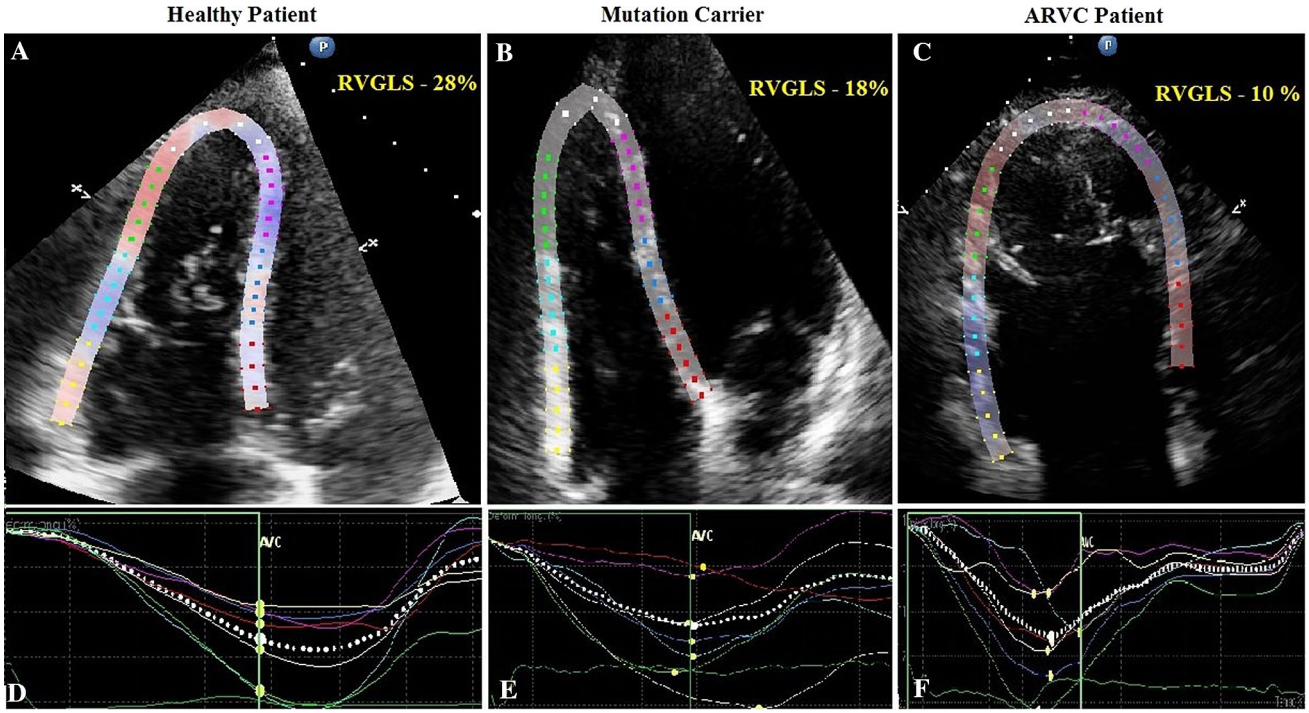

Fig. 1 Head to head comparison of RV global longitudinal strain (RVGLS) and strain curves of a healthy control (**a**, **d**), a mutation carrier (**b**, **e**) and an affected patient (**c**, **f**). RVGLS decrease from the healthy patient to the affected one. Strain curves show a uniform time to peak (yellow dots) indicating a normal mechanical contraction in the healthy patient and a progressively different time to peak indicating mechanical dispersion in the mutation carrier and the affected patient. (Color figure online)

In addition, recent data highlight the potential prognostic role of strain imaging in ARVC. Since electrical abnormalities usually precede structural alterations in ARVC, analysis of mechanical dys-synchrony due to electro-mechanical contraction uncoupling may represent a suitable tool for early detection of the disease. The heterogeneous contraction by myocardial strain has been used as a parameter of mechanical dispersion (Fig. 1). Increased dispersion of time to peak systolic value within the RV and reduced myocardial strain are present in affected patients and asymptomatic mutation carriers and resulted an independent predictor of arrhythmias.

These novel echocardiographic parameters deriving from RV tissue deformation imaging, represent useful tools to improve diagnostic and prognostic approach to ARVC.

These findings highlight the importance of analyzing RV structure and function and the need to increase diagnostic accuracy in clinical practice.

Compliance with ethical standards

Conflict of interest The author declares no conflict of interest.

References

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