Opinion
Volume 16:01, 2025

Human Genetics & Embryology

ISSN: 2161-0436 **Open Access**

Assessing the Prognostic Impact of Strain Imaging in Arrhythmogenic Right Ventricular Cardiomyopathy Using Speckle Tracking Echocardiography

Betancor Poulin *

Department of Cardiology, University Hospitals Birmingham NHS Foundation Trust, Birmingham B15 2GW, UK

Introduction

Arrhythmogenic Right Ventricular Cardiomyopathy (ARVC) is a rare but serious genetic disorder characterized by progressive degeneration of the right ventricular myocardium, leading to arrhythmias, heart failure, and sudden cardiac death. The disease primarily affects the Right Ventricle (RV), although other parts of the heart may also be involved. ARVC is marked by fibro-fatty replacement of the myocardium, which weakens the cardiac muscle and disrupts electrical conduction pathways, resulting in ventricular arrhythmias. Given the subtle nature of early disease progression, timely diagnosis of ARVC remains challenging. A variety of diagnostic tools are utilized to assess the structural and functional changes associated with the disease, but echocardiography, particularly Speckle Tracking Echocardiography (STE), has emerged as a valuable technique for evaluating myocardial strain and deformation in patients with ARVC. STE offers a non-invasive and quantitative assessment of myocardial function, especially in detecting subclinical impairment in the right ventricle, which is often the first chamber affected in ARVC. This review will explore the role of strain imaging using Speckle Tracking Echocardiography (STE) in assessing the prognostic impact of ARVC. It will discuss the pathophysiology of the disease, the utility of STE in evaluating myocardial deformation, and how strain imaging can help predict clinical outcomes in patients with ARVC [1].

Description

ARVC is primarily a genetically inherited disorder, with autosomal dominant inheritance being the most common mode of transmission. Mutations in several genes, including those encoding desmosomal proteins such as plakophilin-2, desmoglein-2, and desmocollin-2, are implicated in the disease's pathogenesis. These mutations disrupt the intercellular junctions that hold cardiac myocytes together, leading to myocardial apoptosis, inflammation, and replacement of the myocardium by fibrous and fatty tissue. The right ventricle is most commonly affected in ARVC due to its thinner myocardium, which is more susceptible to the deleterious effects of these mutations. As the disease progresses, fibrofatty infiltration of the RV myocardium leads to ventricular arrhythmias, most notably Ventricular Tachycardia (VT) and Ventricular Fibrillation (VF), which can result in sudden cardiac death. The left ventricle can also be involved in later stagesof the disease, though the right ventricle remains the primary site of pathological changes. Structural changes in ARVC, such as right ventricular dilation, regional wall motion abnormalities, and thinning of the myocardium, are often detectable using conventional imaging modalities like echocardiography, Magnetic Resonance Imaging (MRI), and Computed Tomography (CT). However, these structural changes may not always correlate with functional impairment, especially in the early stages of the disease. Therefore, functional imaging techniques like speckle tracking echocardiography, which assesses myocardial strain, have become increasingly important for early detection and risk stratification in ARVC [2].

Speckle Tracking Echocardiography (STE) is an advanced imaging technique that allows for the assessment of myocardial deformation in multiple directions, including longitudinal, circumferential, and radial strain. Unlike conventional echocardiography, which provides information on the motion of the heart walls, STE analyzes the movement of myocardial speckles (natural acoustic markers in the tissue) and quantifies the deformation or strain in the myocardium during the cardiac cycle. The use of STE in ARVC provides a unique advantage because it enables the detection of subtle myocardial dysfunction before significant structural changes are evident. In ARVC, early myocardial involvement often occurs without apparent regional wall motion abnormalities, making conventional echocardiography less sensitive for identifying early disease. However, STE can detect impaired myocardial strain, even in the absence of overt structural abnormalities, and therefore provides critical information for early diagnosis and prognosis.

The prognostic impact of strain imaging in ARVC lies in its ability to assess the severity and extent of myocardial dysfunction, which is crucial for predicting clinical outcomes, including the risk of arrhythmias, heart failure, and sudden cardiac death. Several studies have demonstrated that strain measurements obtained from STE can provide valuable information about the progression of ARVC and help predict adverse events [3].

One of the key challenges in managing ARVC is the ability to detect myocardial dysfunction at an early stage, especially before structural changes are visible on conventional imaging. STE has proven to be sensitive in identifying early myocardial impairment in the right ventricle, even when the patient has no overt symptoms or wall motion abnormalities. Impaired myocardial strain, particularly in the right ventricular free wall, is often one of the earliest indicators of disease progression. Studies have shown that Right Ventricular Longitudinal Strain (RVLS) is significantly reduced in patients with ARVC compared to healthy controls. This reduction in RVLS occurs before any significant dilation or thinning of the right ventricle is detectable, making it a valuable early marker for subclinical disease. Monitoring RVLS over time can help track disease progression and identify patients at higher risk of developing symptomatic heart failure or arrhythmias. Ventricular arrhythmias, including VT and VF, are the hallmark of ARVC and are the leading cause of sudden cardiac death in affected individuals. The ability to predict arrhythmic events is critical for determining appropriate treatment strategies, such as the use of Implantable Cardioverter-Defibrillators (ICDs) or antiarrhythmic therapy. STE can play a role in risk stratification by assessing myocardial strain and identifying areas of the right ventricle that are more prone to arrhythmogenic remodeling. Impaired myocardial strain in the right ventricle is often associated with increased

***Address for Correspondence:** Betancor Poulin, Department of Cardiology, University Hospitals Birmingham NHS Foundation Trust, Birmingham B15 2GW, UK, E-mail: moret@edu.com*

Received: 29 January, 2025, Manuscript No. hgec-25-164245; **Editor Assigned:** 31 January, 2025, PreQC No. P-164245; **Reviewed:** 14 February, 2025, QC No. Q-164245; **Revised:** 19 February, 2025, Manuscript No. R-164245; **Published:** 26 February, 2025, DOI: 10.37421/2161-0436.2025.16.272

arrhythmic risk in ARVC. For example, studies have demonstrated that patients with significant reduction in RVLS have a higher likelihood of experiencing sustained VT or requiring ICD implantation. Additionally, the presence of regional strain abnormalities in the right ventricle, particularly in areas with fibro-fatty infiltration, has been linked to a higher risk of arrhythmias. Therefore, STE can help identify patients who may benefit from more aggressive treatment or closer monitoring [4].

As ARVC progresses, the right ventricle becomes increasingly dilated and dysfunctional, leading to right-sided heart failure. Although conventional echocardiography can identify gross changes in RV size and function, STE offers more sensitive and quantitative data on myocardial deformation, allowing for a better assessment of the severity of functional impairment. This is particularly important for predicting the onset of heart failure and guiding treatment decisions. Studies have shown that reduced RV strain is associated with a worse prognosis in ARVC, including a higher risk of heart failure symptoms and reduced exercise capacity. In some cases, the progression of myocardial dysfunction can be identified through changes in strain imaging before clinical symptoms appear. Monitoring strain over time can help clinicians track the disease's progression and make more informed decisions regarding interventions [5].

Conclusion

Speckle tracking echocardiography represents a significant advancement in the assessment of myocardial function in patients with arrhythmogenic right ventricular cardiomyopathy (ARVC). By evaluating myocardial strain, STE provides a sensitive and non-invasive tool for detecting early myocardial dysfunction, assessing arrhythmic risk, and predicting clinical outcomes such as heart failure and sudden cardiac death. Given its ability to detect subclinical changes in myocardial function, strain imaging has the potential to improve early diagnosis and risk stratification in ARVC, ultimately leading to more personalized and timely treatment strategies. As the understanding of ARVC continues to evolve, strain imaging by STE may become an integral part of clinical practice, helping to guide therapeutic decisions and improve patient outcomes. Further research into the prognostic value of strain imaging, particularly in large, multi-center studies, will be essential to refine its role in managing ARVC and other arrhythmogenic cardiomyopathies.

Acknowledgment

None.

Conflict of Interest

There are no conflicts of interest by author.

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How to cite this article: Poulin, Betancor. "Assessing the Prognostic Impact of Strain Imaging in Arrhythmogenic Right Ventricular Cardiomyopathy Using Speckle Tracking Echocardiography." *Human Genet Embryol* 16 (2025): 272.