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The prognostic role of cardiac magnetic resonance in restrictive cardiomyopathy

Restrictive cardiomyopathy (RCM) is a challenging myocardial disorder, characterized by restrictive left and/or right diastolic pathophysiology in the setting of normal or small diastolic volumes, normal or small systolic volumes, and preserved ventricular wall thickness, as defined by the European Society of Cardiology (ESC).[1,2] This umbrella phenotype encompasses genetic myocyte disorders and desminopathies, as well as endomyocardial and extracellular matrix diseases, including endomyocardial fibrosis, hypereosinophilia, hyperoxaluria, and radiation injury. Importantly, diseases with only occasional restrictive physiology do not meet the criteria for RCM.[1]

Cardiac magnetic resonance (CMR) has shown a key role in the evaluation of cardiomyopathies, offering unique insights into morphology, function, and tissue characterization. However, its prognostic role in RCM remains less well established. Recent reports examining patients with preserved left ventricular ejection fraction (LVEF) and restrictive physiology illustrate the usefulness of CMR in risk stratification but also reveal the challenges of applying CMR in this context.[3]

In heart disease with restrictive physiology, LVEF may remain preserved despite advanced disease, limiting its utility for risk stratification. Global longitudinal strain (GLS) derived from feature-tracking CMR has emerged as a more sensitive parameter, detecting subclinical systolic dysfunction before this is reflected in the LVEF and independently predicting mortality and heart failure (HF) hospitalization, even when LVEF appears normal. Thresholds around -9.5% have shown prognostic value, highlighting the need to incorporate strain analysis into standard practice.[3]

Late gadolinium enhancement (LGE) adds complementary prognostic information by detecting myocardial fibrosis. While quantification of LGE burden is not consistently predictive, the presence of LGE alone has been repeatedly associated with adverse outcomes across non-ischemic cardiomyopathies.[3,4] Additionally, clarifying the relevance of specific enhancement patterns, such as isolated junctional versus diffuse or patchy fibrosis, remains an important research question.

Native T1 mapping and extracellular volume quantification provide non-invasive measures of diffuse myocardial involvement and interstitial expansion. Their prognostic value has been extensively validated in cardiac amyloidosis, where elevated native T1 and ECV have been associated with all-cause mortality and HF hospitalization.[5] However, the prognostic role in the remaining spectrum of restrictive heart disease is less well established, as data come mainly from HF with preserved ejection fraction (HFpEF) studies, where ECV has been shown to be an important progrnostic parameter for adverse outcomes.[6]

Despite encouraging findings, several challenges restrict the translation of CMR prognostic markers into practice. Terminological inconsistency, such as the interchangeable use of “idiopathic” or “non-specific” RCM, does not align with ESC classification and complicates cross-study comparisons. Similarly, heterogeneous criteria for defining restrictive physiology and incomplete reporting of diastolic or valvular parameters limit reproducibility. In the absence of standardized definitions, the integration of imaging biomarkers into clinical decision-making will be impeded.

For patients with RCM and preserved LVEF, therapeutic strategies remain largely extrapolated from HFpEF guidelines.[5] Sodium-glucose co-transporter 2 (SGLT2) inhibitors are the only pharmacological agents with a proven prog-

Key words

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nostic benefit in reducing the risk of HF hospitalization or cardiovascular death,[5] while diuretics help in relieving congestion. In this context, refining prognostic assessment through CMR and its implications in patient management is particularly valuable, but requires harmonized methodologies and validation in multicentre cohorts.

Although CMR demonstrates promising potential in RCM through markers such as GLS and LGE, the clinical implementation of those prognostic markers faces several challenges. Future work should focus on standardizing RCM terminology, validating CMR strain thresholds, and defining the prognostic impact of specific LGE patterns, with the ultimate goal of integrating imaging markers into therapeutic strategies.

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ΠΕΡΙΛΗΨΗ

Ο προγνωστικός ρόλος της μαγνητικής τομογραφίας καρδιάς στην περιοριστική μυοκαρδιοπάθεια

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