Evaluation of hypertrophic cardiomyopathy: new horizons for CMR?

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Cardiac magnetic resonance imaging (CMR) is an accurate and reliable means of evaluating cardiac morphology, ventricular function, and myocardial perfusion, both for the left and the right ventricle thereby covering a whole spectrum of cardiac diseases.¹ CMR is therefore very well suited for identifying and characterising patients with various manifestations of left ventricular hypertrophy (LVH).² For instance, CMR can resolve the question whether training-induced LVH in athletes is a physiological rather than a pathophysiological phenomenon. In a previous meta-analysis, involving 59 studies and 1451 athletes (both endurance-trained and strength-trained athletes), it was confirmed that the athlete's heart demonstrated normal systolic and diastolic cardiac function, implying that training-induced LVH in athletes is predominantly a physiological phenomenon.³ CMR has also been shown of great value in patients with hypertrophic cardiomyopathy (HCM).⁴⁻⁷ By virtue of its high spatial and temporal resolution, CMR is capable of accurately identifying regional deformities in patients with HCM. In particular, CMR tagging allows the evaluation of regional systolic and diastolic function after therapeutic interventions.⁸ In recent years, late gadolinium enhancement CMR has been used to visualise myocardial interstitial abnormalities in patients with different forms of cardiomyopathies.⁹ Late gadolinium enhancement was present in cardiomyopathy patients who showed a mean signal intensity 3-4 times higher than in normal regions.¹⁰ The affected areas included papillary muscles (sarcoïd), the mid-myocardium (Anderson-Fabry disease, glycogen storage disease, myocarditis, Becker and Duchenne muscular dystrophy) and the global

subendocardium (systemic sclerosis, Loeffler's endocarditis, amyloid, Churg-Strauss).¹¹ In these specific cardiomyopathies, a typical pattern of late gadolinium enhancement can be found which is overtly distinct from the enhancement patterns seen in myocardial infarction.¹²⁻¹⁸

In an interesting review article in the current issue of the *Netherlands Heart Journal*, Germans et al.¹⁹ address the clinical applicability of CMR to determine the underlying cause of different forms of LVH, in particular in patients with HCM. According to the recent position statement of the European Society of Cardiology, HCM can be subdivided into familial and nonfamilial forms.²⁰ It is well known that HCM is characterised by myocyte disarray, hypertrophy and interstitial fibrosis, which predominantly results in diastolic dysfunction and may serve as substrates for ventricular arrhythmias.²⁰ In the familial form, HCM accounts for at least 25% of sudden cardiac death in young athletes, and is sometimes the first symptom of disease.^{22,23} Interestingly, in HCM patients with documented ventricular arrhythmias on Holter monitoring, late enhancement is more often visible, whereby the extent of delayed enhancement is increased in patients with two or more clinical risk factors for sudden cardiac death.²⁴ As a result, late enhancement CMR can be used in assessing risk for sudden cardiac death in HCM patients. Importantly, Germans et al. were the first to show structural abnormalities described as crypts of the inferoseptum in 80 to 90% of HCM mutation carriers, in whom no hypertrophy was present.^{25,26} As pointed out by the authors, these crypts can be considered a sign of probable HCM mutation carriership but should be carefully discriminated from hypertrabecularisation of the myocardium, which is not typically found in HCM mutation carriers.²⁷

In nonfamilial HCM, CMR may be of great value in identifying systemic disorders such as diabetes, end-stage renal disease, and amyloidosis. Cardiac involvement of amyloidosis displays a characteristic pattern on late enhancement imaging which occurs in approximately 70% of patients.²⁸ The typical pattern of late enhancement together

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with the increased T1 signal of myocardium yield a diagnostic accuracy of 97% in patients with cardiac involvement in biopsy-proven amyloidosis. Lastly, CMR has great value in assessing endocardial fibrosis and measuring the severity of iron overload in patients with cardiomyopathies.

In order to gain more insight into diagnostic capabilities and prognostic implications of LVH using CMR, a European Cardiovascular Magnetic Resonance Registry (EuroCMR Registry) has recently been launched.²⁸ The EuroCMR Registry consists of two parts: 1) a multicentre registry with consecutive enrolment of patients scanned in all participating European CMR centres using web-based online case record forms, and 2) a prospective clinical follow-up of patients with suspected coronary artery disease and HCM every 12 months after enrolment to assess prognostic data. The EuroCMR Registry therefore offers an opportunity to provide information about the clinical utility of routine CMR in a large number of patients and in diverse populations. In particular, it has the potential to gather information about the prognostic value of CMR in patients with LVH and HCM. We are anxiously awaiting prospective data in the near future. The paper by Germans et al. already offers an encouraging prospect.^{19,*} ■

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* P.S. Of note, the above-mentioned article by Germans et al. has been selected as e-learning article with the aim to improve general knowledge on hypertrophic cardiomyopathy and to gather accreditation points. We hope the reader will be stimulated to read the article and respond to the online questions. Instructions will be given at the end of the article.