Current use of cardiac magnetic resonance in tertiary referral centres for the diagnosis of cardiomyopathy: the ESC EORP Cardiomyopathy/Myocarditis Registry

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ESC European Society of Cardiology

European Heart Journal - Cardiovascular Imaging (2021) **22**, 781–789
doi:10.1093/ehjci/jeaa329

Current use of cardiac magnetic resonance in tertiary referral centres for the diagnosis of cardiomyopathy: the ESC EORP Cardiomyopathy/Myocarditis Registry

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Received 7 April 2020; editorial decision 9 November 2020; accepted 12 November 2020; online publish-ahead-of-print 8 January 2021

Aims Cardiac magnetic resonance (CMR) is recommended in the diagnosis of cardiomyopathies, but it is time-consuming, expensive, and limited in availability in some European regions. The aim of this study was to determine the use of CMR in cardiomyopathy patients enrolled into the European Society of Cardiology (ESC) cardiomyopathy registry [part of the EURObservational Research Programme (EORP)].

Methods and results Three thousand, two hundred, and eight consecutive adult patients (34.6% female; median age: 53.0 ± 15 years) with cardiomyopathy were studied: 1260 with dilated (DCM), 1739 with hypertrophic (HCM), 66 with restrictive (RCM), and 143 with arrhythmogenic right ventricular cardiomyopathy (ARVC). CMR scans were performed at baseline in only 29.4% of patients. CMR utilization was variable according to cardiomyopathy subtypes: from 51.1% in ARVC to 36.4% in RCM, 33.8% in HCM, and 20.6% in DCM ($P < 0.001$). CMR use in tertiary referral centres located in different European countries varied from 1% to 63.2%. Patients undergoing CMR were younger, less symptomatic, less frequently had implantable cardioverter-defibrillator (ICD)/pacemaker implanted, had fewer cardiovascular risk factors and comorbidities ($P < 0.001$). In 28.6% of patients, CMR was used along with transthoracic echocardiography (TTE); 67.6% patients underwent TTE alone, and 0.9% only CMR.

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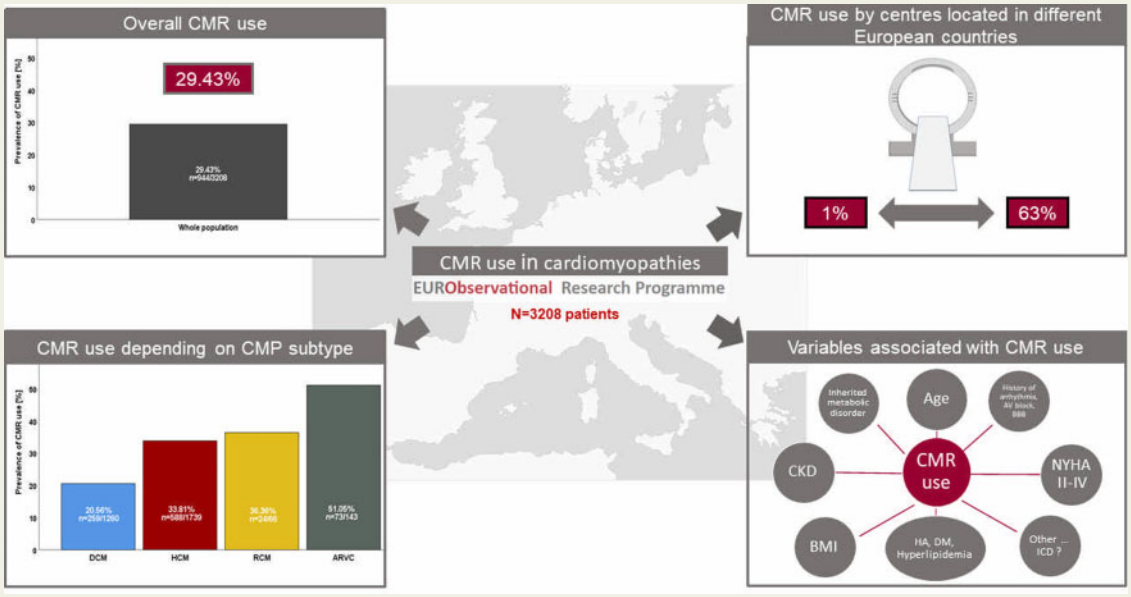
Conclusion Less than one-third of patients enrolled in the registry underwent CMR and the use varied greatly between cardiomyopathy subtypes, clinical profiles of patients, and European tertiary referral centres. This gap with current guidelines needs to be considered carefully by scientific societies to promote wider availability and use of CMR in patients with cardiomyopathies.

Graphical Abstract



Keywords cardiomyopathy • cardiac magnetic resonance • transthoracic echocardiography • registry

Introduction

Cardiomyopathies are a heterogeneous group of disorders defined by structural and functional abnormalities of the myocardium unexplained by loading conditions or coronary artery disease.[1–7] Cardiac imaging is a prerequisite for the diagnosis and management of cardiomyopathy and international guidelines on the assessment of cardiomyopathies recommend a multimodality imaging approach to diagnosis, including transthoracic echocardiography (TTE) and cardiac magnetic resonance (CMR).[8–10] Some limitations of TTE may be overcome using CMR. The great advantage of CMR imaging is not only the important role in patients with poor echo windows but also the crucial role in evaluating tissue characteristics and myocardial perfusion.[11,12] However, CMR is a time-consuming and expensive method with limited availability in some European regions. It can be anticipated that the real-life choice of the appropriate technique is based on expert knowledge, cost–benefit ratio and, most importantly, its availability. However, no data are available regarding the current use of CMR in cardiomyopathies, especially at a European level.

The Cardiomyopathy/Myocarditis Registry is part of the EURObservational Research Programme (EORP) and is designed to collect prospective clinical data on patients with a confirmed diagnosis of cardiomyopathy.[13] The aim is to provide insights into the contemporary features, diagnostic process, and management of patients with cardiomyopathy across Europe.[13]

In this study of adult patients with cardiomyopathy enrolled in the registry, we analysed the use of CMR and the potential CMR determinants according to cardiomyopathy subtypes and clinical profile of patients in different European tertiary cardiology centres.

Methods

General design

The design and protocol for the EORP Cardiomyopathy/Myocarditis Registry as well as the mandatory criteria for the participating centres have been reported previously.[13] The study was approved by local ethics committees and all participants gave written informed consent to registry enrolment. The diagnostic work-up and therapy as documented by the registry reflected the local approach to management of cardiomyopathy patients. The data on patients' demographics and clinical characteristics were gathered by means of structured electronic case report form accessible via a secure website.[13] The study was coordinated and supervised by the EORP department of ESC. The data were anonymously processed. The statistical analyses were performed by the core EORP statistical unit.

Patients and cardiomyopathies subtypes

Three thousand, two hundred and eight consecutive adult patients (34.9% female; median age at enrolment 53.0 ± 15 years; mean age the first evaluation in the centre: 49 ± 15 years) with cardiomyopathy were

studied: 1260 had dilated cardiomyopathy (DCM); 1739 hypertrophic cardiomyopathy (HCM); 66 restrictive cardiomyopathy (RCM); and 143 arrhythmogenic right ventricular cardiomyopathy (ARVC). Patients were recruited in 68 centres located in 18 countries. Participating centres should have the expertise in management of cardiomyopathies and were selected using pre-specified criteria, that is, Cardiac Magnetic Resonance Imaging Lab with experience in diagnosis of typical and atypical cardiomyopathies.[13] The number of enrolled patients was diverse from 27 up to 659 per country. The inclusion criteria comprised: age >18 years, consent to study participation and unequivocal diagnosis of cardiomyopathy consistent with diagnostic criteria for either probands or relatives. All the definitions applied for the study population were formerly specified in the core manuscript.[13] For the purpose of the study, the whole cardiomyopathy population was divided into subjects with CMR (CMR population) and without CMR used in the diagnostic process (non-CMR population).

Diagnostic tests

Data on the following tests regarding cardiomyopathy diagnosis were noted in the CRF: electrocardiogram, transthoracic echocardiography (TTE) with Doppler assessment, CMR, 24 h ECG Holter monitoring, exercise test, and genetics. Data on all tests were recorded at two time points: at baseline and at 1-year follow-up. The analysis presented here was focused on CMR application in the cardiomyopathy diagnostic process; TTE was used as a comparator for CMR applicability.

Among detailed TTE and CMR parameters registered in the CRF protocol, the study presented information on whether CMR and TTE were performed and on conclusion of the CMR scanning (normal, abnormal, and inconclusive). According to the EORP Registry CRF, the following reasons for diagnosis were taken into account: incidental, symptoms, history of cardiac arrest, family screening, and so on.

Statistical analysis

Univariate analysis was applied to both continuous and categorical variables. Continuous variables were reported as mean ± standard deviation (SD) and/or as median and interquartile range (IQR) when appropriate. Among-group comparisons were made using the non-parametric test (Kruskal–Wallis test). Categorical variables were reported as percentages. Among-group comparisons were made using the χ^2 test or a Fisher's exact test if any expected cell count was <5. A univariate logistic regression analysis was performed to identify variables associated with CMR use in study population. Odds ratio (OR) and 95% confidence interval (95% CI) were obtained. A two-sided *P*-value <0.05 was considered as statistically significant. All analyses were performed using SAS statistical software version 9.4 (SAS Institute Inc., Cary, NC, USA).

Results

Prevalence of CMR use in cardiomyopathy patients

Baseline CMR scans were performed in 944 (29.4%) patients. The prevalence of CMR use in different cardiomyopathies was as follows: 20.6% in DCM, 33.8% in HCM, 36.4% in RCM, and 51.1% in ARVC ($P < 0.001$) (*Table 1* and *Figure 1*). Abnormal CMR results were present in 93.4% of patients, with the highest percentage in RCM (95.8%) and HCM (94.9%) followed by DCM (91.5%) and ARVC (87.7%) ($P = 0.030$); normal CMR results were registered in 5.6% of patients, and only 1.0% the CMR results were inconclusive for diagnosis (*Table 1*).

In 83 subjects without a baseline CMR imaging, the CMR evaluation was completed during 1-year follow-up. Finally, the total prevalence of CMR use at baseline and at 1-year follow-up raised to 32.0% with the highest prevalence in ARVC (53.9%) (*Table 1*).

The prevalence of CMR use (baseline+1-year follow-up data) in patients with cardiomyopathies varied from 1% to 63.2% in centres located in different European countries (*Figure 2*).

Demographic and clinical characteristics of CMR and non-CMR populations

Some demographic features differed in patients diagnosed using CMR compared to the non-CMR population. Age at enrolment (50.0 ± 15.7 vs. 54.8 ± 14.6 years, $P < 0.001$) and age at the first evaluation in the centre (46.8 ± 16.4 vs. 50.5 ± 15.5 years, $P < 0.001$) were lower in the CMR population. The CMR group had lower BMI (26.5 ± 4.6 vs. 27.3 ± 5.0 kg/m^2, $P < 0.001$). Inherited metabolic disorders were more frequently observed in CMR subjects (2.0 vs. 0.7%, $P = 0.012$). NYHA class was less advanced in the CMR population (NYHA I/II/III/IV: 37.0/43.0/16.5/3.4%) than in non-CMR subjects (NYHA I/II/III/IV: 23.5/47.9/24.3/4.4%, $P < 0.001$). History of arrhythmias, atrial fibrillation (20.3% vs. 36.2%, $P < 0.001$), sustained VT (8.2% vs. 12.8%, $P < 0.001$), and AV block (6.1% vs. 10.5%, $P = 0.003$) were less frequent in CMR than in non-CMR population.

The percentage of the patients with implanted ICD was lower in the CMR as compared with the non-CMR population (18.01% vs. 29.67%, $P < 0.001$). The ICDs were implanted in approximately 80% patients for primary and in 20% for secondary prophylaxis of sudden cardiac death (SCD) both in CMR and non-CMR subjects. Among patients implanted for the primary prophylaxis of SCD ($n = 677$): 148 (21.86%) subjects were examined by CMR. Among patients implanted for the secondary prophylaxis of SCD ($n = 155$): 37 (23.87%) underwent CMR.

The following comorbidities were less prevalent in CMR population: arterial hypertension (29.8% vs. 39.3%, $P < 0.001$), diabetes mellitus (9.8% vs. 13.9%, $P = 0.001$), hyperlipidaemia (28.8% vs. 39.3%, $P < 0.001$), and renal impairment (6.5% vs. 12.8%, $P < 0.001$).

Univariate logistic regression analysis of different demographic, clinical, and imaging variables associated with the CMR use in the whole population confirmed the above-mentioned results and quantified the magnitude of effects through odds ratio (*Table 2*).

CMR use and reason for cardiomyopathy diagnosis

In patients in whom the CMR imaging was performed at baseline and/or follow-up, incidental, history of cardiac arrest, family screening, and other reasons for diagnosis, were registered more frequently than in non-CMR population. On the other hand, in non-CMR subjects, the presence of symptoms dominated as a reason for diagnosis (71.7% vs. 57.3% in CMR population, $P < 0.001$) (*Table 3*). Similar observations were obtained in patients with DCM ($P < 0.001$) and HCM ($P = 0.001$) (*Table 4*).

Table 1 Prevalence of CMR use in a whole cardiomyopathy population and in different types of cardiomyopathies

Variables		Type of cardiomyopathy					P value
		All (N = 3208)	DCM (N = 1260)	HCM (N = 1739)	RCM (N = 66)	ARVC (N = 143)	
Baseline evaluation							
CMR scan performed		944/3208 (29.43%)	259/1260 (20.56%)	588/1739 (33.81%)	24/66 (36.36%)	73/143 (51.05%)	<0.001
CMR scan	Normal	53/944 (5.61%)	19/259 (7.34%)	27/588 (4.59%)	0/24 (0.00%)	7/73 (9.59%)	
	Abnormal	882/944 (93.43%)	237/259 (91.51%)	558/588 (94.90%)	23/24 (95.83%)	64/73 (87.67%)	0.030
	Inconclusive	9/944 (0.95%)	3/259 (1.16%)	3/588 (0.51%)	1/24 (4.17%)	2/73 (2.74%)	
TTE		3086/3208 (96.20%)	1221/1260 (96.90%)	1666/1739 (95.80%)	63/66 (95.45%)	136/143 (95.10%)	0.387
Baseline+12-month follow-up							
CMR scan performed[a]		1027/3208 (32.01%)	280/1260 (22.22%)	644/1739 (37.03%)	26/66 (39.39%)	77/143 (53.85%)	<0.001
CMR scan[a]	Normal	69/1027 (6.72%)	21/280 (7.50%)	39/644 (6.06%)	2/26 (7.69%)	7/77 (9.09%)	0.046
	Abnormal	948/1027 (92.31%)	256/280 (91.43%)	602/644 (93.48%)	23/26 (88.46%)	67/77 (87.01%)	
	Inconclusive	10/1027 (0.97%)	3/280 (1.07%)	3/644 (0.47%)	1/26 (3.85%)	3/77 (3.90%)	

[a]Data were collected from baseline evaluation if CMR was performed at baseline, and from 12-month follow-up evaluation if CMR was not performed at baseline.
ARVC, arrhythmogenic right ventricular CM; CM, cardiomyopathy; CMR, cardiac magnetic resonance; DCM, dilated CM; HCM, hypertrophic CM; RCM, restrictive CM; TTE, transthoracic echocardiography.

Application of TTE/CMR for cardiomyopathy diagnosis

At baseline, CMR was used as a single diagnostic method in only 0.9% of patients. In 28.6% of patients, the CMR was used along with transthoracic echocardiography. TTE was the only diagnostic imaging method at baseline in 67.6% of patients (*Table 5*).

Comparison of the TTE and CMR application among different cardiomyopathies shows that CMR constitutes a single diagnostic method in a limited number of patients: 0.5% in DCM, 1.2% in HCM, 0.0% in RCM, and 1.4% in ARVC. On the other hand, TTE was used without CMR imaging in 76.8% of patients with DCM, in 63.1% of patients with HCM, in 59.1% of patients with RCM, and in 45.6% of patients with ARVC (*Table 5*).

Discussion

This study shows that less than one-third of adult patients enrolled in the ESC EORP Cardiomyopathy Registry underwent CMR and that the CMR use varied greatly between cardiomyopathy subtypes, clinical profiles of patients, and European centres.

Expert consensus statements and ESC guidelines recommend that CMR scanning should be performed both for diagnosis, prognosis as well as for further therapeutic options in patients with a suspected cardiomyopathy.[5,14–17] In the ESC Cardiomyopathy Registry, most cardiomyopathy diagnoses were made on the basis of TTE imaging alone, although there were difference between cardiomyopathies and different regions. Indeed, TTE is a non-invasive, cost-effective, and widely available method; however, it has limited application in myocardial tissue evaluation.[8–10] In contemporary cardiology practice, TTE followed by CMR as complementary modality should be implemented.

The Registry provided data also on the conclusions of the CMR imaging (normal, abnormal, and inconclusive). Most patients presented an abnormal CMR scanning and only 1.0% of the results were inconclusive. However, as many as 5.6% of CMR results were assessed as normal and the final cardiomyopathy diagnosis was based on other diagnostic methods (i.e. TTE, genetic tests). We should be aware that every diagnostic method has some false negative results as well as we cannot exclude some biases in the evaluation process.

CMR in different cardiomyopathy subtypes

CMR has an important clinical role in all cardiomyopathies in terms of diagnosis,[6,8,10,11,18,19] treatment strategy, and the prediction of prognosis.[20–22] However, the Registry revealed unsatisfactory availability of CMR in every cardiomyopathy sub-type.

In our study, CMR was used most frequently (51.1%) in patients with ARVC, probably reflecting the limitations of TTE in assessing the right ventricle as well as the importance of tissue characterisation in this disease. In contrast, the use of CMR was low in all other subtypes, in spite of clear guidance on the importance of CMR in characterising phenotypes and in assessing sudden death risk. For example, CMR characterization of myocardial tissue is fundamental in cases of suspected amyloidosis, sarcoidosis, Fabry disease, and

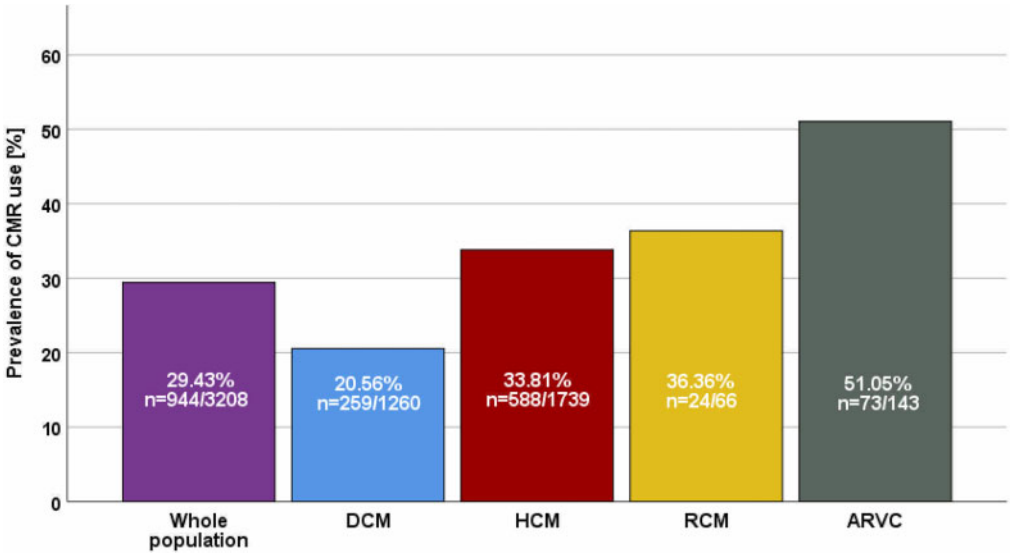


Figure 1 Prevalence of CMR use in patients with cardiomyopathies—whole cardiomyopathy population and different cardiomyopathy subtypes—baseline data. ARVC, arrhythmogenic right ventricular cardiomyopathy; DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy; RCM, restrictive cardiomyopathy.

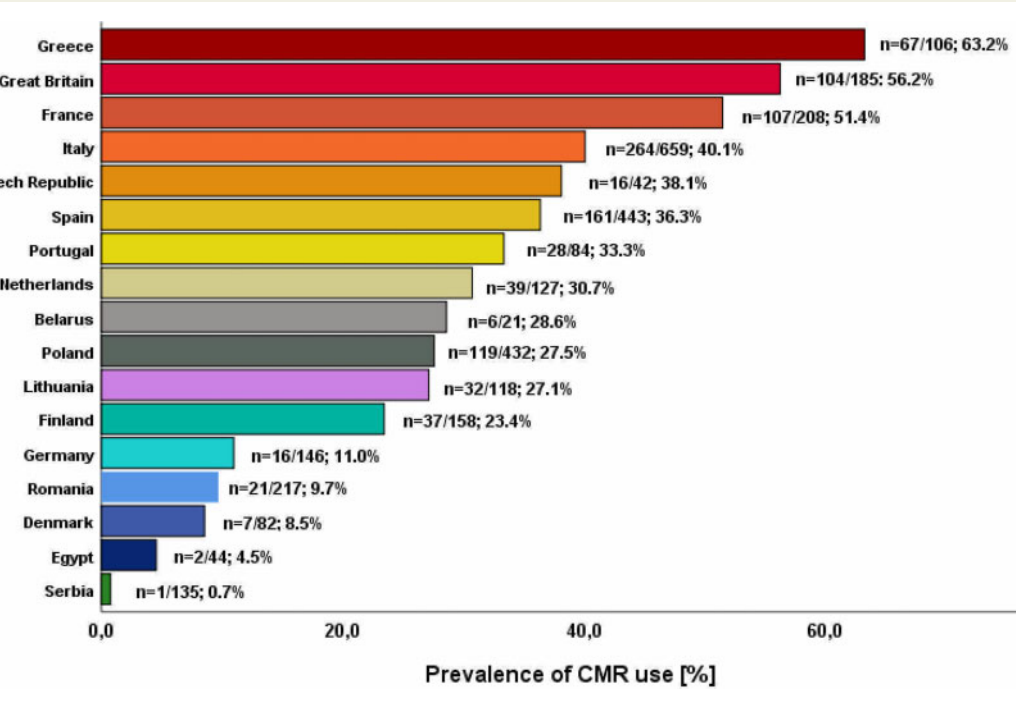


Figure 2 Prevalence of CMR use in whole cardiomyopathy population in centres located in different European countries—baseline+12-month follow-up data.

haemochromatosis[16]; however, CMR was used only in 36.4% of RCM subjects. Access to CMR assessment was even less in HCM (33.8%) and DCM (20.6%). These data contrast with current ESC guidelines for HCM,[5] where CMR is a class I, level B recommendation in the evaluation of heart disease and Class I, Level C recommendation in patients with suspected HCM, who have inadequate

Table 2 Demographic and clinical characteristics of patients with cardiomyopathy associated with the use of CMR (based on baseline and follow-up data)—simple comparison and logistic regression analysis

Variables		CMR (*N* = 1027)	Non-CMR (*N* = 2181)	*P*-value	OR (95% CI)	OR *P*-value
Age at enrolment (years), mean ± SD		50.0 (±15.7)	54.8 (±14.6)	<0.001	0.979 (0.974–0.984)	<0.001
Age at first evaluation in the centre (years), mean ± SD		46.8 (±16.4)	50.5 (±15.5)	<0.001	0.986 (0.981–0.990)	<0.001
Gender: female		355/1027 (34.57%)	764/2181 (35.03%)	0.797	0.980 (0.838–1.145)	0.798
Body mass index (kg/m^2), mean ± SD		26.5 (±4.6)	27.3 (±5.0)	<0.001	0.970 (0.955–0.986)	<0.001
Inherited metabolic disorder		10/507 (1.97%)	11/1586 (0.69%)	0.012	2.879 (1.215–6.819)	0.016
NYHA class	NYHA I	314/848 (37.03%)	419/1784 (23.49%)	<0.001	/	/
	NYHA II	365/848 (43.04%)	854/1784 (47.87%)		0.570 (0.471–0.690)	<0.001
	NYHA III	140/848 (16.51%)	433/1784 (24.27%)		0.432 (0.339–0.549)	<0.001
	NYHA IV	29/848 (3.42%)	78/1784 (4.37%)		0.496 (0.316–0.779)	0.002
History of arrhythmias						
History of atrial fibrillation		208/1027 (20.25%)	790/2181 (36.22%)	<0.001	0.557 (0.410–0.773)	<0.001
History of sustained VT		84/1027 (8.18%)	278/2181 (12.75%)	<0.001	0.610 (0.472–0.788)	<0.001
History of resuscitated VF/cardiac arrest		32/1027 (3.12%)	99/2181 (4.54%)	0.057	0.676 (0.451–1.015)	0.059
History of AV block		31/507 (6.11%)	166/1586 (10.47%)	0.003	0.557 (0.374–0.829)	0.004
History of BBB		83/507 (16.37%)	373/1586 (23.52%)	<0.001	0.637 (0.490–0.828)	<0.001
Family history of sudden death		170/983 (17.29%)	354/1986 (17.82%)	0.721	0.964 (0.788–1.179)	0.722
Cardioverter defibrillator implanted		185/1027 (18.01%)	647/2181 (29.67%)	<0.001	0.521 (0.433–0.626)	<0.001
Primary prophylaxis		148/185 (80.00%)	529/647 (81.76%)	0.587	0.892 (0.591–1.347)	0.587
Secondary prophylaxis		37/185 (20.00%)	118/647 (18.24%)			
Pacemaker implanted		45/1027 (4.38%)	279/2181 (12.79%)	<0.001	0.309 (0.224–0.428)	<0.001
History of stroke: TIA or Stroke		59/1019 (5.79%)	144/2172 (6.63%)	0.365	0.866 (0.633–1.183)	0.365
Comorbidities						
Arterial hypertension		306/1027 (29.80%)	858/2181 (39.34%)	<0.001	0.654 (0.558–0.767)	<0.001
Diabetes mellitus I or II		101/1027 (9.83%)	302/2181 (13.85%)	0.001	0.679 (0.535–0.862)	0.001
Hyperlipidaemia/dyslipidaemia		296/1027 (28.82%)	857/2181 (39.29%)	<0.001	0.626 (0.533–0.734)	<0.001
Renal impairment		67/1027 (6.52%)	278/2181 (12.75%)	<0.001	0.478 (0.362–0.631)	<0.001
Chronic obstructive pulmonary disease		44/1027 (4.28%)	114/2181 (5.23%)	0.250	0.812 (0.568–1.159)	0.250

AV, atrioventricular; BBB, bundle branch block; CI, confidence interval; CMR, cardiac magnetic resonance imaging; NYHA, New York Heart Association classification; OR, odds ratio; TIA, transient ischaemic attack; VF, ventricular fibrillation; VT, ventricular tachyardia.

Table 3 CMR use depending on the reason for diagnosis of cardiomyopathies (based on baseline and follow-up data)

Reason for diagnosis	Whole population of CM patients		
	CMR (*N* = 1027)	Non-CMR (*N* = 2181)	*P*-value
Incidental	214/995 (21.51%)	285/2025 (14.07%)	<0.001
Symptoms	570/995 (57.29%)	1452/2025 (71.70%)	
History of cardiac arrest	18/995 (1.81%)	29/2025 (1.43%)	
Family screening	147/995 (14.77%)	201/2025 (9.93%)	
Other	46/995 (4.62%)	58/2025 (2.86%)	
ND	32	156	

CMR, cardiac magnetic resonance; ND, no data.

echocardiographic window in order to confirm the diagnosis. In the case of HCM and DCM, myocardial scar burden is also an important consideration when assessing prognosis and risk for sudden death risk.[19,21]

CMR in different centres/countries

Our data show that CMR has variable and limited availability in some European centres. The differences between centres localized in different countries need to be interpreted cautiously as the ESC registry

Table 4 CMR use depending on the reason for diagnosis of different cardiomyopathies (based on baseline and follow-up data)

Reason for diagnosis	DCM			HCM		
	CMR (*N* = 280)	Non-CMR (*N* = 980)	*P*-value	CMR (*N* = 644)	Non-CMR (*N* = 1095)	*P*-value
Incidental	36/273 (13.19%)	80/925 (8.65%)	<0.001	168/620 (27.10%)	196/996 (19.68%)	0.001
Symptoms	194/273 (71.06%)	776/925 (83.89%)		309/620 (49.84%)	595/996 (59.74%)	
History of cardiac arrest	4/273 (1.47%)	16/925 (1.73%)		7/620 (1.13%)	11/996 (1.10%)	
Family screening	26/273 (9.52%)	31/925 (3.35%)		107/620 (17.26%)	161/996 (16.16%)	
Other	13/273 (4.76%)	22/925 (2.38%)		29/620 (4.68%)	33/996 (3.31%)	
ND	7	55		24	99	

Reason for diagnosis	RCM			ARVC		
	CMR (*N* = 26)	Non-CMR (*N* = 40)	*P*-value	CMR (*N* = 77)	Non-CMR (*N* = 66)	*P*-value
Incidental	3/26 (11.54%)	2/40 (5.00%)	0.738	7/76 (9.21%)	7/64 (10.94%)	0.467
Symptoms	22/26 (84.62%)	36/40 (90.00%)		45/76 (59.21%)	45/64 (70.31%)	
History of cardiac arrest	0/26 (0.00%)	0/40 (0.00%)		7/76 (9.21%)	2/64 (3.13%)	
Family screening	0/26 (0.00%)	1/40 (2.50%)		14/76 (18.42%)	8/64 (12.50%)	
Other	1/26 (3.85%)	1/40 (2.50%)		3/76 (3.95%)	2/64 (3.13%)	
ND	0	0		1	2	

P-value for comparison between CMR and non-CMR patients separately for DCM, HCM, RCM, and ARVC.
ARVC, arrhythmogenic right ventricular CM; CM, cardiomyopathy; CMR, cardiac magnetic resonance; DCM, dilated CM; HCM, hypertrophic CM; ND, no data; RCM, restrictive CM.

Table 5 Application of different imaging modalities for the diagnosis of cardiomyopathies (based on baseline data)

Variables	CMs (*N* = 3208)	DCM (*N* = 1260)	HCM (*N* = 1739)	RCM (*N* = 66)	ARVC (*N* = 143)
TTE (+)/CMR (-)	2170/3208 (67.64%)	968/1260 (76.83%)	1098/1739 (63.14%)	39/66 (59.09%)	65/143 (45.45%)
TTE (+)/CMR (+)	916/3208 (28.55%)	253/1260 (20.08%)	568/1739 (32.66%)	24/66 (36.36%)	71/143 (49.65%)
TTE (-)/CMR (+)	28/3208 (0.87%)	6/1260 (0.48%)	20/1739 (1.15%)	0/66 (0.00%)	2/143 (1.40%)

ARVC, arrhythmogenic right ventricular CM; CM, cardiomyopathy; CMR, cardiac magnetic resonance; DCM, dilated CM; HCM, hypertrophic CM; RCM, restrictive CM; TTE, transthoracic echocardiography.

is, by definition, limited to a small number of selected centres that may not be representative of local healthcare systems. Nevertheless, the frequency of CMR use varied substantially between centres localized in European countries with the highest percentage reaching 63.2% and the lowest 1%. Indications for CMR assessment are well-established,[5,14–19] thus it may be assumed that the low frequency of CMR use relates to local restraints, for example, relatively high costs and incomplete reimbursement. Limited access to scanners with cardiac dedication and lack of skills to interpret CMR images may also partly explain our findings. Importantly, most patients in the registry were enrolled at tertiary reference centres and teaching hospitals, and thus the use of CMR in general cardiological practice is likely to be even lower.

CMR and clinical profile of patients

In addition to the variation between cardiomyopathies, we observed differences in the characteristics of patients that were scanned . The CMR population was younger, less symptomatic, with a lower prevalence of cardiovascular risk factors and associated cardiovascular diseases compared to the non-CMR population. This may be explained by the perceived need for CMR in patients with milder disease in whom the diagnosis was less secure or in whom risk assessment was more challenging. On the other hand, concomitant renal impairment as the limitation for contrast administration may explain less-frequent CMR use in this population. In support of this, the reason for diagnosis in patients undergoing CMR was more likely to be incidental or prompted by family screening rather than symptoms. Another explanation is that many patients with the most severe disease had ICDs implanted as thus were difficult to scan. It should be noted that electrotherapy could be a limitation for CMR use in cardiomyopathy patients. On the other hand, the decision of ICD implantation is still too rarely taken based on the reference imaging modality of CMR.

The indications (primary vs. secondary) for SCD prophylaxis did not determine the percentage of patients undergoing CMR imaging in the EORP Registry. The CMR use was similar in population with the urgent indications for ICD implantation (secondary prophylaxis of SCD) as well as in population with the elective ICD implantations (primary prophylaxis of SCD).

Study limitations

There are limitations intrinsic to all registries including selection bias and lack of adjudication. As most patients were enrolled in tertiary referral centres, our results may not be generally applicable, and CMR use could be even lower in less expert centres. In relation to the considerable disparities among centres located in different countries on prevalence of CMR use, it should be emphasised that our results pertain primarily to centres with high CMR utilization. Participating centres should have the expertise in management of cardiomyopathies, that is, CMR Imaging Lab with experience in diagnosis of typical and atypical cardiomyopathies. Therefore, the presented data may even overestimate the actual CMR use. The considerable disparities among the number of enrolled patients should also be noted. Especially low-number centres may mismatch analysis of the percentage of CMR use. Additionally <50% of the ESC affiliated counties have been enrolled into the Registry and it constitutes the next limitation of the study. It should be noted that some patients had ICD at baseline or underwent ICD implantation during the follow-up, thus limiting CMR use. Generally, a lot of therapeutic decisions, that is, ICD implantation for primary prevention of SCD, have been made without 'gold-standard' like CMR imaging.

The follow-up was as short as 12 months and a waiting time for CMR might have been longer in some countries due to reimbursement issues. The study presents only information on whether CMR was performed and on conclusion of the CMR, without details of the CMR imaging. The registry was not dedicated to study a temporal sequence of the use of CMR and TTE in cardiomyopathy diagnostic process and an impact of the CMR result on the management. This is an inherent limitation of all registries.

Conclusion

The EORP Cardiomyopathy/Myocarditis Registry provides real-life data on the use of CMR in patients with cardiomyopathies. Regardless of the potential value of CMR in this setting, the overall use of CMR in Europe is limited. Less than one-third of patients enrolled in the registry underwent CMR and the use varied greatly between cardiomyopathy subtypes, clinical profiles of patients, and European tertiary referral centres. This gap between society recommendations and clinical practice needs to be better understood and should be considered more deliberately in the drafting of practice guidelines. An improvement regarding access, training, and reimbursement is necessary to provide wider application of CMR in diagnosis of cardiomyopathies.

Supplementary data

Supplementary data are available at *European Heart Journal - Cardiovascular Imaging* online.

Acknowledgements

EORP Oversight Committee, The Registry Executive Committee of the EURObservational Research Programme (EORP). Data collection was conducted by the EORP department from the ESC by Rachid Mir Hassaine as Clinical Project Manager, Emanuela Fiorucci, Myriam Glemot and Patti-Ann McNeill as Project Officers, Marème Konté and Sebastien Authier as Data Managers. Statistical analyses were performed by Cécile Laroche. Overall activities were coordinated and supervised by Doctor Aldo P. Maggioni (EORP Scientific Coordinator). All investigators are listed in the Supplementary data online, *Appendix S1*.

Funding

Since the start of EORP, the following companies have supported the programme: Abbott Vascular Int. (2011–21), Amgen Cardiovascular (2009–18), AstraZeneca (2014–21), Bayer AG (2009–18), Boehringer Ingelheim (2009–19), Boston Scientific (2009–12), The Bristol Myers Squibb and Pfizer Alliance (2011–19), Daiichi Sankyo Europe GmbH (2011–20), The Alliance Daiichi Sankyo Europe GmbH and Eli Lilly and Company (2014–17), Edwards (2016–19), Gedeon Richter Plc. (2014–16), Menarini Int. Op. (2009–12), MSD-Merck & Co. (2011–14), Novartis Pharma AG (2014–20), ResMed (2014–16), Sanofi (2009–11), SERVIER (2009–21), and Vifor (2019–22).

Conflict of interest: none declared.

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