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30 Settembre
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Cardionerologia: le cardiomiopatie

QUALE IL RUOLO DELL'IMAGING NELLE DISTROFIE MIOTONICHE?

Martino Annamaria, MD, PhD

Policlinico Casilino, Rome

Journal Pre-proof

2022 HRS expert consensus statement on evaluation and management of arrhythmic risk in neuromuscular disorders

William J. Groh, MD, MPH, FHRS (Chair),[1] Deepak Bhakta, MD, MBA, FHRS, FACC, FAHA, FACP, CCDS (Vice-Chair),[2] Gordon F. Tomaselli, MD, FHRS (Vice-Chair),[3] Ryan G. Aleong, MD, FHRS,[4] Ricardo Alkmim Teixeira, MD, PhD,[5*] Anthony Amato, MD,[6+] Samuel J. Asirvatham, MD, FHRS,[7] Yong-Mei Cha, MD, FHRS,[7] Domenico Corrado, MD, PhD, FESC,[8‡] Denis Duboc, MD, PhD,[9] Zachary D. Goldberger, MD, FHRS,[10] Minoru Horie, MD, PhD,[11§] Joseph E. Hornyak, MD, PhD,[12¶] John Lynn Jefferies, MD, MPH, FACC, FAHA, FHFSA,[13#] Stefan Kääb, MD, PhD,[14‡] Jonathan M. Kalman, MBBS, PhD, FHRS,[15] Naomi J. Kertesz, MD, FHRS, CEPS-P,[16**] Neal K. Lakdawala, MD,[6++] Pier D. Lambiase, BCH, BM, MBChB, PhD, FHRS,[17‡] Steven A. Lubitz, MD, MPH,[18‡‡] Hugh J. McMillan, MD, MSc,[19§§] Elizabeth M. McNally, MD, PhD,[20#] Margherita Milone, MD, PhD,[7+] Narayanan Namboodiri, MBBS, MD,[21¶¶] Saman Nazarian, MD, PhD, FHRS,[22] Kristen K. Patton, MD, FHRS,[23] Vincenzo Russo, MD, PhD,[24‡] Frederic Sacher, MD, PhD,[25‡] Pasquale Santangeli, MD, PhD,[22] Win-Kuang Shen, MD, FHRS,[26] Dario C. Sobral Filho, MD, PhD,[27##] Bruce S. Stambler, MD, FHRS,[28] Claudia Stöllberger, MD,[29] Karim Wahbi, MD, PhD,[30] Xander H.T. Wehrens, MD, PhD, FHRS,[31] Menachem Mendel Weiner, MD,[32***] Matthew T. Wheeler, MD, PhD,[33#] Katja Zeppenfeld, MD, PhD[34‡]

Heart Rhythm 2022;S1547-5271

4.2 Diagnostic testing and risk stratification in myotonic dystrophy types 1 and 2

Recommendations for diagnostic testing and risk stratification in myotonic dystrophy types 1 and 2

Referenced studies that support recommendations are summarized in Appendix 3

COR	LOE	Recommendations	References
1	C-EO	1. Coordinated care of patients with DM1 or DM2 should be conducted in a medical setting where there is access to expertise in the neurological, cardiac, arrhythmic, pulmonary, and genetic manifestations of these disorders.	
1	B-NR	2. In patients with DM1 or DM2, cardiac evaluation including physical examination, ECG, ambulatory ECG, and cardiac imaging (echocardiography or CMR) at diagnosis with periodic retesting is recommended even in the absence of cardiac symptoms.	24, 26, 104, 106, 108, 110, 114, 115

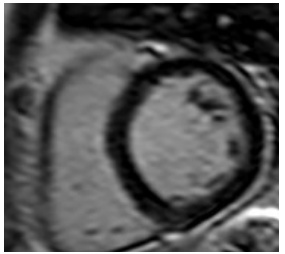
Trans-thoracic echocardiography (2D or 3D)

LV and RV dimension, wall thickness, systolic and dyastolic function

Valves, vessels, pericardium

Subtle localized myocardial contractility impairment (strain)

Contrast enhanced cardiac MRI



may detect eventual myocardial damage suggestive of scar (LGE)

may quantify interstitial fibrosis (extracellular volume fraction)

may detect subtle localized myocardial contractility impairment (strain)

SPECIAL REPORT

American Heart Association.

Clinical Care Recommendations for Cardiologists Treating Adults With Myotonic Dystrophy

Elizabeth M. McNally, MD, PhD; Douglas L. Mann, MD; Yigal Pinto, MD, PhD; Deepak Bhakta, MD; Gordon Tomaselli, MD; Saman Nazarian, MD, PhD; William J. Groh, MD, MPH; Takuhisa Tamura, MD; Denis Duboc, MD; Hideki Itoh, MD, PhD; Leah Hellerstein, LCSW, MPH; Pradeep P. A. Mammen, MD

Abstract—Myotonic dystrophy is an inherited systemic disorder affecting skeletal muscle and the heart. Genetic testing for myotonic dystrophy is diagnostic and identifies those at risk for cardiac complications. The 2 major genetic forms of myotonic dystrophy, type 1 and type 2, differ in genetic etiology yet share clinical features. The cardiac management of myotonic dystrophy should include surveillance for arrhythmias and left ventricular dysfunction, both of which occur in progressive manner and contribute to morbidity and mortality. To promote the development of care guidelines for myotonic dystrophy, the Myotonic Foundation solicited the input of care experts and organized the drafting of these recommendations. As a rare disorder, large scale clinical trial data to guide the management of myotonic dystrophy are largely lacking. The following recommendations represent expert consensus opinion from those with experience in the management of myotonic dystrophy, in part supported by literature-based evidence where available. (***J Am Heart Assoc***. 2020;9:e014006. DOI: 10.1161/JAHA.119.014006.)

AHA scientific statement expert consensus opinion suggests

DM1: cardiac imaging examination at baseline and every 1 to 5 years thereafter

Two steps back…

ECHOCARDIOGRAPHY

LV diastolic dysfunction.

Prevalence 5%-50%
(different selection of subjects and TEE techniques)

Related to:
-AF,
-myocardial fibrotic degenerative changes
-impaired calcium metabolism in cardiomyocytes

Paunic et al. J Chin Med Assoc. 2017;80:408-412

LV systolic dysfunction Prevalence: 0%-21%

.

Different findings:
-retrospective single-center TTE cohort studies
-different characteristics of subjects,
-variety in the definition of LVSD

Review

Prevalence of Left Ventricular Systolic Dysfunction in Myotonic Dystrophy Type 1: A Systematic Review

VINCENZO RUSSO, MD, PhD, MMSc,[1] SIMONA SPERLONGANO, MD,[1] EMANUELE GALLINORO, MD,[1] ANNA RAGO, MD,[1] ANDREA ANTONIO PAPA, MD,[1] PAOLO GOLINO, MD, PhD,[1] LUISA POLITANO, MD, PhD,[2] SAMAN NAZARIAN, MD, PhD,[3] AND GERARDO NIGRO, MD, PhD[1]

Table 2. Echocardiographic Findings Concerning LV Systolic Function

Author	Year of publication	DMI (n)	Mean Age, y ± D	Cardiac Ultrasound Examination (n)	Cut-off LVEF	LV Systolic Dysfunctionn (%)
Paunic	2017	111	42,2 ± 10,9	111	<55%	8 (7.5)
Tanawuttiwat	2016	136	44,46 ± 15,24	136	< 55%	16 (11,8)
Petri	2014	129	44 ± 15	126	< 50%	26 (20,6)
Dhand	2013	27	39,1 ± 14,1	27	< 50%	5 (18,5)
Bhakta	2010	406	42 ± 12	180	< 50%	34 (18,9)
Lindqvist	2009	36	45 ± 10	36	< 50%	0 (0)
Di Cori	2009	31	42 ± 12	31	< 50%	0 (0)
Total		876	42,68	647		89 (13,8)

cut-off LVEF, cut-off considered in the study for the definition of left ventricle systolic dysfunction; DMI (n), number of patients affected by myotonic dystrophy I included in the study; LV, systolic dysfunction n (%), number of patients with systolic dysfunction and percentage in parentheses; LVEF, left ventricle ejection fraction; SD, standard deviation; y, mean age of the patients included in the study.

Journal of Cardiac Failure Vol. 00 No. 00 2019

LV systolic dysfunction due to:

-intra-ventricular (IV) and atrio-ventricular (AV) conduction time delay,

-atrial or ventricular arrhythmias

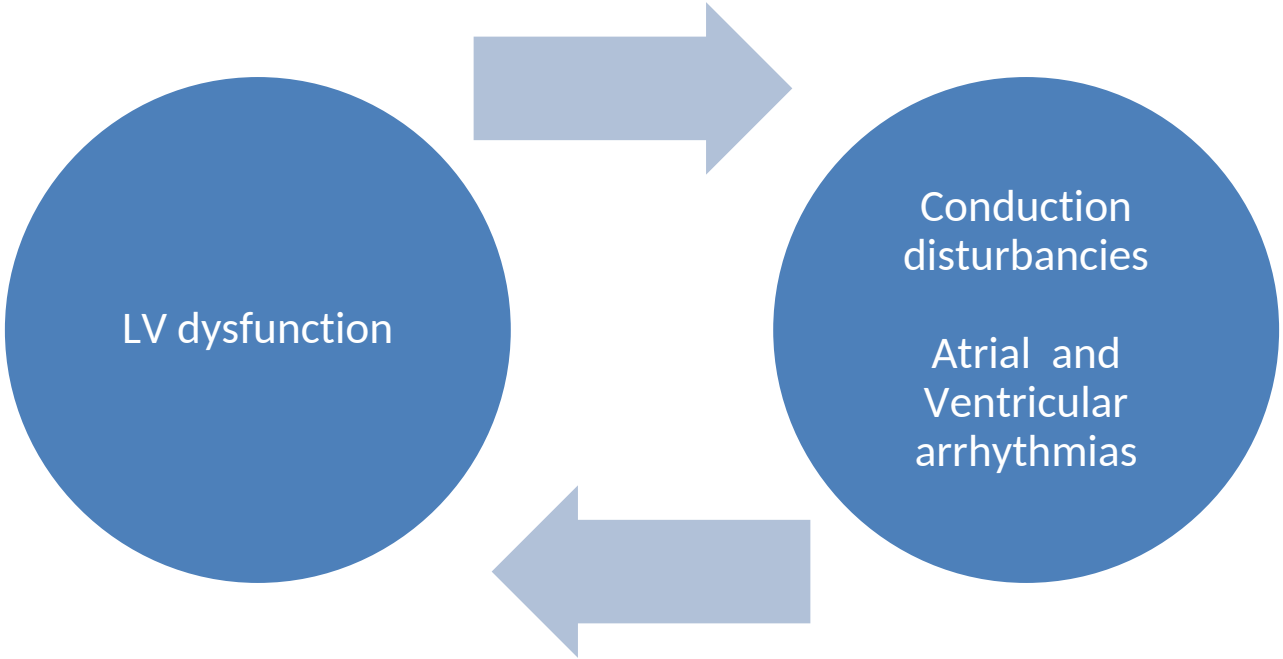
-myocardial fibrosis.

DM1 with prolonged PR or QRS intervals: 4-times higher HF risk

Association between LVSD and AF (prevalence up to 46%)

Paunic et al. J Chin Med Assoc. 2017;80:408-412
Russo V et al, J Cardiac Fail 2019;00:1_8
Bhakta D et al. Am Heart J 2010;160:1137e41
McNally EM et al. J Am Heart Assoc. 2020;9:e014006

Electro-mechanical correlation



McNally EM et al. J Am Heart Assoc. 2020;9:e014006

ESC European Society of Cardiology

European Heart Journal - Case Reports (2019) 3, 1–5
doi:10.1093/ehjcr/ytz095

CASE SERIES
Arrhythmias/Electrophysiology

Ventricular tachycardia in patients with type 1 myotonic dystrophy: a case series

Anish Nikhanj [1,2], Soori Sivakumaran[1,2], Bailey Miskew-Nichols[1,2], Zaeem A. Siddiqi[3], and Gavin Y. Oudit [1,2]*

[1]Division of Cardiology, Department of Medicine, University of Alberta, 8440 112 Street NW, Edmonton, Alberta T6G 2R7, Canada; [2]Mazankowski Alberta Heart Institute, University of Alberta, 11220 83 Avenue NW, Edmonton, Alberta T6G 2J2, Canada; and [3]Division of Neurology, Department of Medicine, University of Alberta, 8440 112 Street NW, Edmonton, Alberta T6G 2R7, Canada

Received 10 December 2018; first decision 5 February 2019; accepted 18 May 2019; online publish-ahead-of-print 9 June 2019

Background Type 1 myotonic dystrophy (DM1) is associated with a variety of cardiac conduction abnormalities and the frequent need for permanent pacing. However, the role of ventricular tachycardia (VT) and the implied risk of sudden cardiac death (SCD) is poorly understood.

Case summary This study examined a 56-patient DM1 cohort of men and women, and identified five patients (two females and three males) with ventricular arrhythmias (8.9%). Patients were reviewed on a case-by-case basis, with their clinical presentation and management of VT and the associated cardiomyopathy indicated. Patient cardiac function was determined by 12-lead electrocardiogram, 48-h Holter monitor, and transthoracic echocardiography. These patients were therefore suitable candidates for implantable cardioverter-defibrillator implantation and received these devices; four of the five patients also received cardiac resynchronization therapy. Medical therapies included angiotensin converting enzyme inhibition, mineralocorticoid receptor antagonist, and following device implantation, beta-blocker therapy was initiated.

Discussion Our case series demonstrates the prevalence of VT in patients with DM1 highlighting the associated risks of SCD in this patient population. The burden of ventricular arrhythmias, advanced conduction disease, and cardiomyopathy are best treated with a combination of device and medical therapies.

Keywords Myotonic dystrophy • Cardiomyopathy • Conduction disease • Arrhythmia • Ventricular tachycardia • Sudden cardiac death • Case series

Table I Clinical characteristics of patients with type 1 myotonic dystrophy presenting with ventricular tachycardia

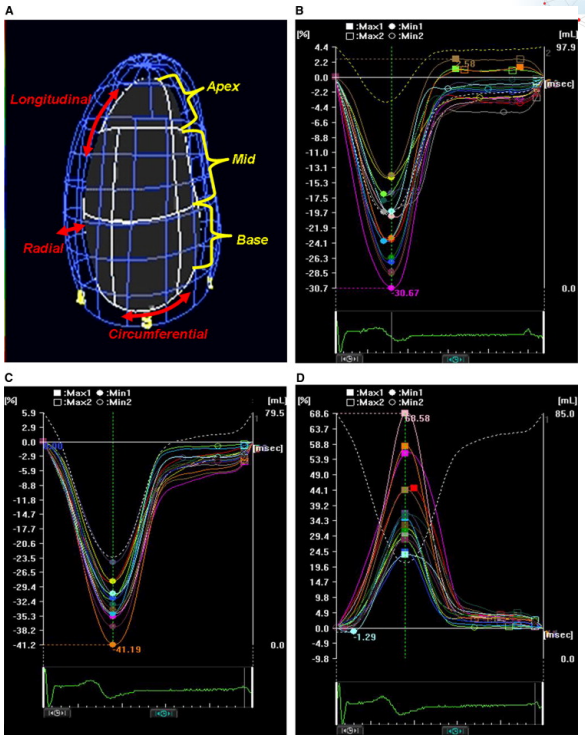
Patient	Age (years)/ gender	HR (b.p.m.)	ECG findings (ms)	Echocardiographic parameters	VT detection	Presentation	Medications
1	44/F	82	• PR: 212; QRS: 105 QTc: 476 • Biventricular paced rhythm	• LVEF: 46% • LVMI: 56.6 g/m^2 • LVIDd: 5.0 cm • LVIDs: 4.3 cm	CRT-P	Cardiac Arrest	• Mexiletine • Perindopril
2	36/F	69	• PR: 179; QRS: 146 QTc: 388 • Atrial-paced rhythm, LBBB	• LVEF: 46% • LVMI: 60.9 g/m^2 • LVIDd: 4.6 cm • LVIDs: 3.7 cm	DC-PPM	Asymptomatic	• Furosemide • Ramipril • Spironolactone
3	49/M	86	• PR: 200; QRS: 107 QTc: 442 • LAFB	• LVEF: 45% • LVMI: 51.5 g/m^2 • LVIDd: 4.0 cm • LVIDs: 3.3 cm	Tele-monitoring	Syncope	• ASA • Metoprolol • Ramipril • Rosuvastatin
4	61/M	64	• PR: 272; QRS: 168 QTc: 446 • Atrial-paced rhythm, LBBB	• LVEF: 45% • LVMI: 69.7 g/m^2 • LVIDd: 4.8cm • LVIDs: 3.5cm	DC-PPM	Palpitations	• Atorvastatin • Candesartan • Spironolactone
5	40/M	71	• QRS: 170; QTc: 474 • Atrial fibrillation, biventricular paced rhythm	• LVEF: 38% • LVMI: 137 g/m^2 • LVIDd: 6.6cm • LVIDs: 5.1cm	CRT-D	ICD shock	• Digoxin • Ramipril • Warfarin

Refer to Methods section for how values were obtained or calculated.
ASA, acetylsalicylic acid; CRT-D, cardiac resynchronization therapy defibrillator; CRT-P, cardiac resynchronization therapy pacemaker; DC-PPM, dual-chamber permanent pacemaker; ECG, electrocardiogram; LBBB, left bundle branch block; LV, left ventricle; LVEF, left ventricular ejection fraction; LVIDd, left ventricle internal diameter end diastole; LVIDs, left ventricle internal diameter end systole; VT, ventricular tachycardia.

3D-TTE and speckle tracking analysis

empower 2D TTE diagnostic and prognostic ability.

Early marker of systolic dysfunction.



Russo V et al, J Cardiac Fail 2019;00:1_8
Lau JK et al. Int J Cardiol 2015;184:600-608
Galderisi M et al. Int Jour Cardiol 2014; 176:1094–1096

International Journal of Cardiology 176 (2014) 1094–1096

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International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Letter to the Editor

Early changes of myocardial deformation properties in patients with dystrophia myotonica type 1: A three-dimensional Speckle Tracking echocardiographic study

Maurizio Galderisi [a,*], Francesco De Stefano [a], Ciro Santoro [a], Agostino Buonauro [a], Daniela De Palma [a], Fiore Manganelli [b], Lucia Ruggiero [b], Lucio Santoro [b], Giovanni de Simone [a]

[a] Hypertension Research Center, Department of Translational Medical Sciences, "Federico II" University Hospital, Naples, Italy
[b] Department of Neurosciences, Reproductive and Odontostatric Sciences, "Federico II" University Hospital, Naples, Italy

DM1 subjects with normal LVEF compared with an healthy control group

impairment of 3D global circumferential strain

(**impaired contraction of the midwall layer of cardiomyocytes**)

Cardiac MRI

Observational studies have observed a certain prevalence of:

-reduced LVEF

-reduced RVEF ,

-LV hypertrophy

-LV non-compaction

Hermans MCE et al.J Cardiovasc Magn Reson 2012;14:48
Luetkens JA et al, Circ Cardiovasc Imaging 2019;12:e009100
Choudhary P et al. Heart 2016; 102:1472–1478

LV LGE:

-non-ischemic distribution pattern
(i.e. in the midwall or subepicardial myocardial layers),

-mostly located in the inter-ventricular septum or in the postero-lateral wall

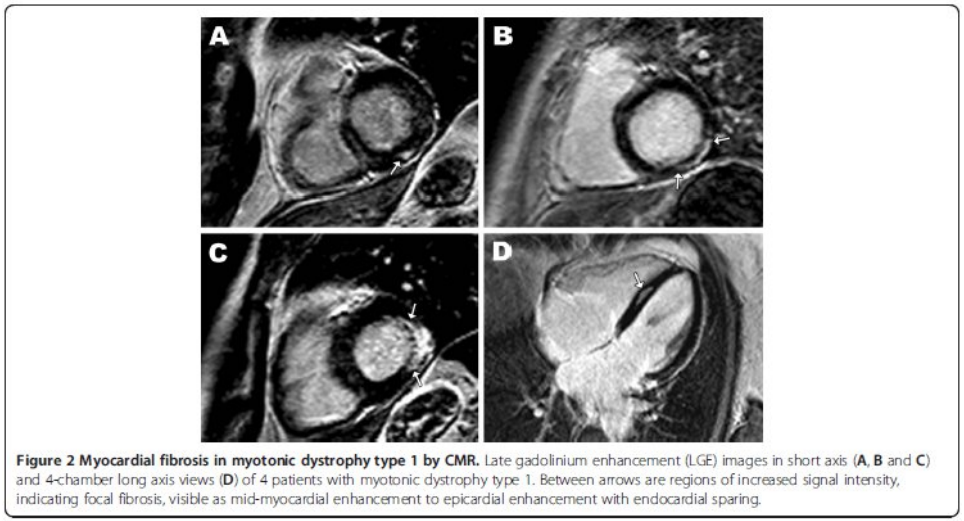


Figure 2 Myocardial fibrosis in myotonic dystrophy type 1 by CMR. Late gadolinium enhancement (LGE) images in short axis (**A**, **B** and **C**) and 4-chamber long axis views (**D**) of 4 patients with myotonic dystrophy type 1. Between arrows are regions of increased signal intensity, indicating focal fibrosis, visible as mid-myocardial enhancement to epicardial enhancement with endocardial sparing.

From Hermans MCE et al.J Cardiovasc Magn Reson 2012;14:48

Hermans MCE et al.J Cardiovasc Magn Reson 2012;14:48
Choudhary P et al. Heart 2016; 102:1472–1478
Chmielewski L et al. Clin Res Cardiol 2019;108:857-867.
Petri H et al. Cardiovasc Magn Reson; 2014:16:59
Cardona A et al. J Cardiovasc Magn Reson. 2019;21:26.

-prevalence of LGE:

12.5-13% (Hermans; Choudhary),

32% (Chmielewski L),

40-42% (Petri; Cardona).

Why different prevalences?

Different LGE extimation methodologies (different sensibility)

Different populations

Hermans MCE et al.J Cardiovasc Magn Reson 2012;14:48
Choudhary P et al. Heart 2016; 102:1472–1478
Chmielewski L et al. Clin Res Cardiol 2019;108:857-867.
Petri H et al. Cardiovasc Magn Reson; 2014:16:59
Cardona A et al. J Cardiovasc Magn Reson. 2019;21:26.

Association between LGE and the number of CTG repeat sequences

(Hermans, Choudhary).

Debated association between LGE and cardiac conduction abnormalities

(Petri, Cardona, Hermans).

Hermans MCE et al.J Cardiovasc Magn Reson 2012;14:48
Choudhary P et al. Heart 2016; 102:1472–1478
Petri H et al. Cardiovasc Magn Reson; 2014:16:59
Cardona A et al. J Cardiovasc Magn Reson. 2019;21:26.

Hermans et al. Journal of Cardiovascular Magnetic Resonance 2012, 14:48
http://www.jcmr-online.com/content/14/1/48

Journal of Cardiovascular Magnetic Resonance

RESEARCH Open Access

Structural and functional cardiac changes in myotonic dystrophy type 1: a cardiovascular magnetic resonance study

Mieke C E Hermans[1*], Catharina G Faber[1], Sebastiaan C A M Bekkers[2], Christine E M de Die-Smulders[4], Monique M Gerrits[4], Ingemar S J Merkies[5], Gabriel Snoep[3], Yigal M Pinto[6] and Simon Schalla[2]

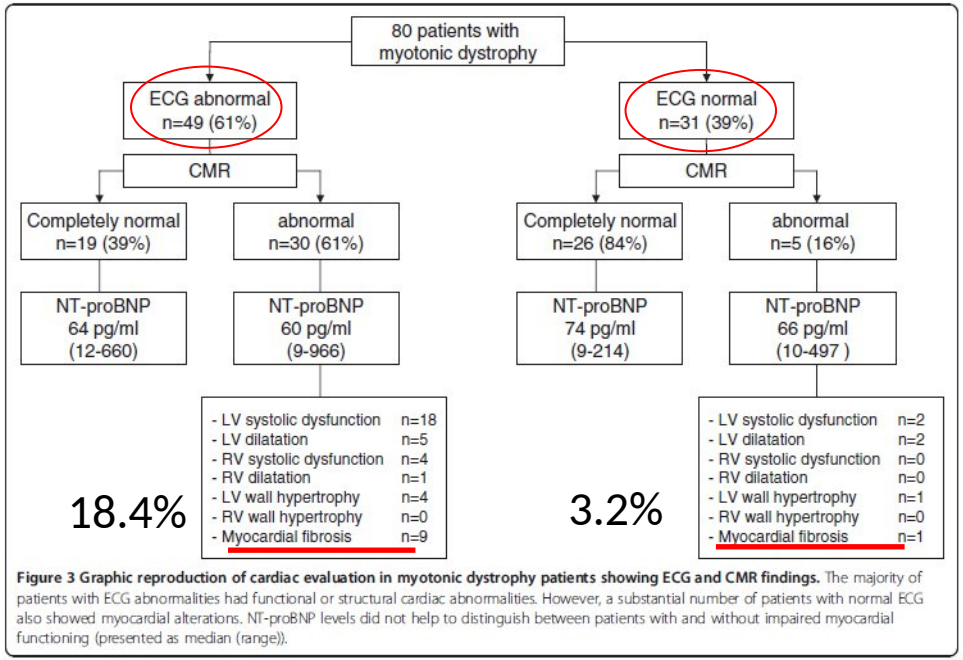


Figure 3 Graphic reproduction of cardiac evaluation in myotonic dystrophy patients showing ECG and CMR findings. The majority of patients with ECG abnormalities had functional or structural cardiac abnormalities. However, a substantial number of patients with normal ECG also showed myocardial alterations. NT-proBNP levels did not help to distinguish between patients with and without impaired myocardial functioning (presented as median (range)).

Myocardial fibrosis by late gadolinium enhancement cardiovascular magnetic resonance in myotonic muscular dystrophy type 1: highly prevalent but not associated with surface conduction abnormality

Andrea Cardona[1,2], William D. Arnold[3], John T. Kissel[3], Subha V. Raman[1] and Karolina M. Zareba[1*]

Abstract

Background: Conduction disease and arrhythmias represent a major cause of mortality in myotonic muscular dystrophy type 1 (MMD1). Permanent pacemaker (PPM) implantation is the cornerstone of therapy to reduce cardiovascular mortality in MMD1. Cardiovascular magnetic resonance (CMR) studies demonstrate a high prevalence of myocardial fibrosis in MMD1, however the association between CMR myocardial fibrosis with late gadolinium enhancement (CMR-LGE) and surface conduction abnormality is not well established in MMD1.
We investigated whether myocardial fibrosis by CMR-LGE is associated with surface conduction abnormalities meeting criteria for PPM implantation according to current guidelines in a cohort of patients with genetically confirmed MMD1.

Methods: Patients with genetically confirmed MMD1 were retrospectively evaluated. 12-lead electrocardiography (ECG) performed within 6 months of CMR was necessary for inclusion. The severity and extent of MMD1 was quantified using a validated Muscular Impairment Rating Scale (MIRS). Based on current guidelines for device-based therapy of cardiac rhythm abnormalities, we defined surface conduction abnormality as the presence of ECG alterations meeting criteria for PPM implant (class I or II indications): PR interval > 200 ms (type I atrioventricular (AV) block) and/or mono or bifascicular block (QRS > 120 ms), or evidence of advanced AV block. Balanced steady-state free precession sequences (bSSFP) were used for assessment of left ventricular (LV) volumes and ejection fraction. MOdified Look-Locker Inversion Recovery (MOLLI) acquisition schemes were used to acquire T1 maps. Patients' charts were reviewed up to 12 months post-CMR for occurrence of PPM implantation.

Results: Fifty-two patients (38% male, 41 ± 14 years) were included. Overall, 31 (60%) patients had a surface conduction abnormality and 22 (42%) demonstrated midwall myocardial fibrosis by CMR-LGE. After a median of 57 days from CMR exam, 15 patients (29%) underwent PPM implantation. Subjects with vs. without surface conduction abnormality had significantly longer disease length (15.5 vs. 7.8 years, $p = 0.015$) and higher disease severity on the MIRS scale ($p = 0.041$). High prevalence of myocardial fibrosis by CMR-LGE was detected in subjects with and without surface conduction abnormality with no significant difference between the two cohorts (42% vs. 43%, $p = 0.999$). By multivariate logistic regression analysis, disease length was the only independent variable associated with surface conduction abnormality (OR 1.071, 95%CI 1.003–1.144, $p = 0.040$); while CMR-LGE was not associated with conduction abnormality ($p = -0.009$, $p = 0.949$).

(Continued on next page)

Table 3 CMR Findings

CMR data	Whole Cohort (N = 52)	Conduction Abnormality Positive (N = 31)	Conduction Abnormality Negative (N = 21)	P value
LVEF (%)	60 ± 6	59 ± 6	60 ± 6	0.687
LV EDVI, ml/m^2	65 ± 15	65 ± 14	66 ± 15	0.728
LV ESVI, ml/m^2	26 ± 8	26 ± 8	26 ± 7	0.940
LV mass index, g/m^2	44 ± 11	45 ± 14	43 ± 6	0.563
LAVI, ml/m^2	30 ± 11	31 ± 10	29 ± 12	0.634
LGE, N (%)	22 (42%)	13 (42)	9 (43)	0.999
LGE mass, g	2.4 ± 3.6	2.4 ± 3.8	2.3 ± 3.4	0.876
LGE, %	3.1 ± 4.6	3.0 ± 4.6	3.1 ± 4.7	0.975
ECV, %	25 ± 3	26 ± 3	24 ± 3	0.050

Data are presented as mean ± SD or N (%). *LVEF* left ventricular ejection fraction, *EDVI* end-diastolic volume index, *ESVI* end-systolic volume index, *LAVI* left atrial volume index, *LGE* late gadolinium enhancement, *ECV* extracellular volume

Cardona et al. Journal of Cardiovascular Magnetic Resonance (2019) 21:26
https://doi.org/10.1186/s12968-019-0535-6

Is association between LGE and cardiac conduction abnormalities a matter of time?

May LGE (septum) preceide development of cardiac conduction abnormalities?

Increased ECV values, suggesting diffuse subclinical LV myocardial interstitial fibrosis,

in comparison with healthy controls

In comparison with normal cut-offs reported in the literature.

Luetkensen JA et al. Circ Cardiovasc Imaging 2019;12:e009100

13 DM1
22 DM2
31 healthy controls

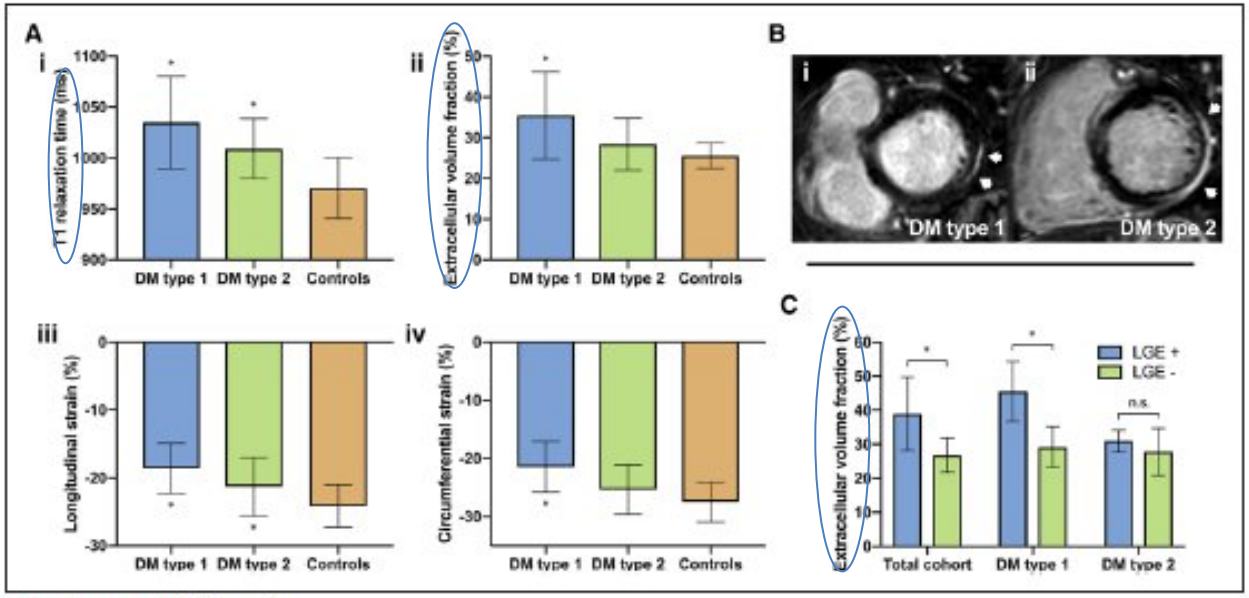


Figure. Summary of study results.

A, Column graphs of different T1 mapping and strain parameters for myotonic dystrophy (DM) type 1, DM type 2, and control subjects. Data are presented as mean with SD error bars. Differences are shown for (**AI**) native T1 relaxation time, (**AII**) extracellular volume fraction, (**AIII**) global peak systolic longitudinal strain, and (**AIv**) global peak systolic circumferential strain. *Statistical significance ($P<0.05$) against control subjects. **B**, Visible myocardial fibrosis in 2 male patients with DM type 1 (47 y old; **BI**) and DM type 2 (56 y old; **BII**). On short-axis late gadolinium enhancement (LGE), image regions of high signal intensity (enhancement) are seen in the subepicardium of the lateral basal wall (arrows) indicating myocardial fibrosis. **C**, Column graph shows differences in extracellular volume fraction between LGE-positive and LGE-negative patients for the total cohort, DM type 1 patients, and DM type 2 patients. Data are presented as mean with SD error bars. NS indicates nonsignificant. *Statistical significance ($P<0.05$) between groups.

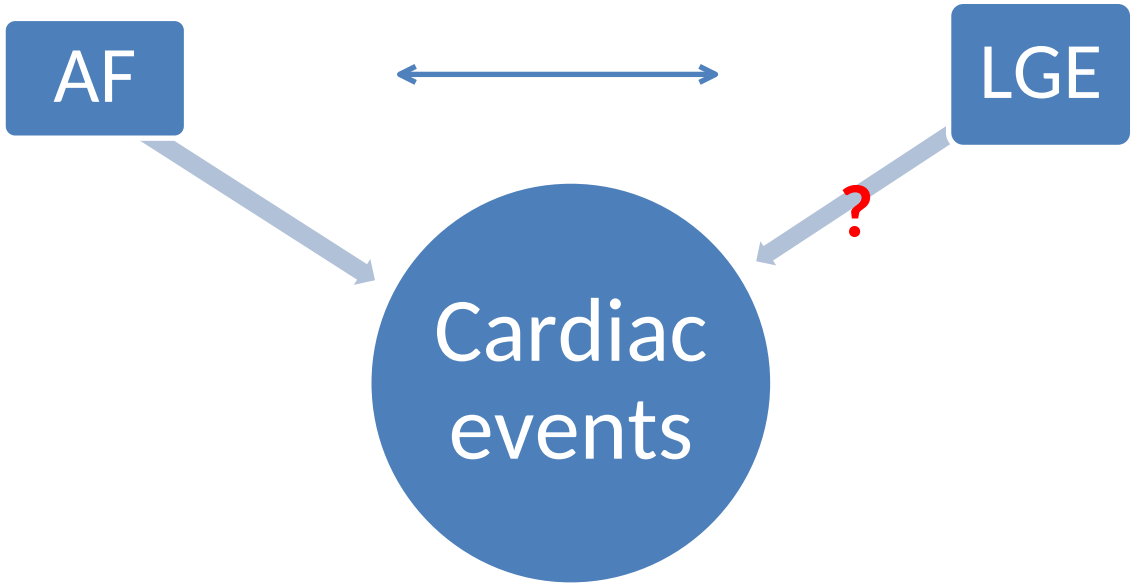
Luetkensen JA et al. Circ Cardiovasc Imaging 2019;12:e009100

Unknown prognostic role of LGE or interstitial fibrosis

Association between LGE (intramural pattern, septum and inferior/inferolateral segments), and AF (Chmielewski L)

AF in turn, predicts adverse cardiac events in DM1 subjects

Chmielewski L et al. Clin Res Cardiol 2019;108:857-867.
Groh et al Cardiovasc Magn Reson; 2014:16:59


AF
LGE
?
Cardiac events

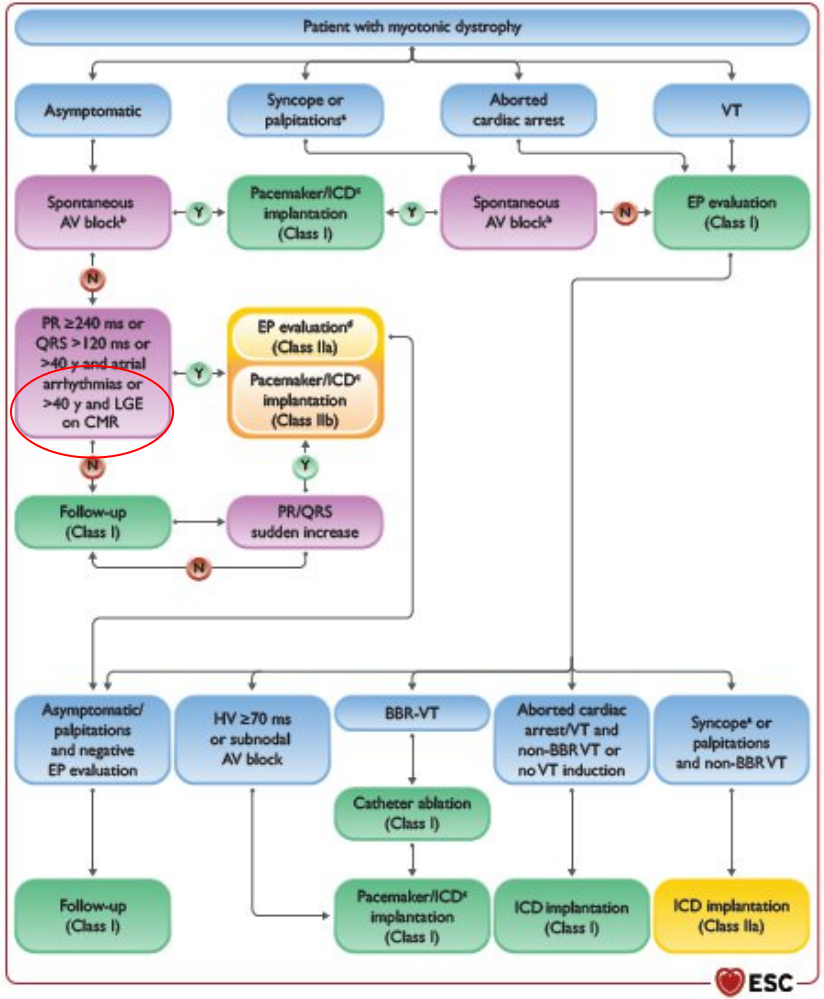
ESC European Society of Cardiology
European Heart Journal (2022) 00, 1–130
https://doi.org/10.1093/eurheartj/ehac262

ESC GUIDELINES

2022 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Developed by the task force for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death of the European Society of Cardiology (ESC)

Endorsed by the Association for European Paediatric and Congenital Cardiology (AEPC)



Invasive electrophysiological evaluation should be considered in patients with myotonic dystrophy and a PR interval ≥240 ms or QRS duration ≥120 ms or who are older than 40 years and have supraventricular arrhythmias[c] or who are older than 40 years and have significant LGE on CMR.[c,5,14,16,766]	IIa	B

Wahbi et al. Eur Heart J 2017;38:751
Hermans MCE et al. J Cardiovasc Magn Reson 2012; 14:48
Wahbi J et al. JAMA 2012; 307; 1292

Future larger studies will help clarifying the role of cardiac MRI for an eventual risk stratification in DM.

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