PLACE

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ROMA
Centro Congressi
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9ª Edizione

30 Settembre
1 Ottobre
2022

Stratificazione del rischio e terapia nelle malattie neuromuscolari

applicazione delle linee guida nella pratica clinica

Università degli Studi della Campania *Luigi Vanvitelli*

OSPEDALI DEI COLLI
MONALDI - COTUGNO - C.T.O.

Vincenzo Russo ***MD PhD MMSc***

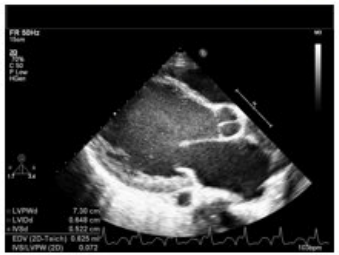
Myopathy	Gene	Heart involvement	Frequency of heart involvement	Ventricular arrhythmia	Atrial arrhythmia	Sudden death reported
Duchenne	Dystrophin	DCM	>90%	PVC	Only at late stage	Yes
Becker	Dystrophin	DCM	60–75%	VT associated with DCM	Associated with DCM	Yes
Myotonic, type 1	CGT repeat expansion	Conduction disease and DCM	60–80%	VT, ICD indicated	Age dependent	Yes, 30% of death
Myotonic, type 2	CGT repeat expansion	Conduction disease	10–25%	Uncommon	Uncommon	Yes
Emery-Dreifuss	Emerin, lamin A and C	Conduction disease and DCM	>90%	VT, ICD indicated	Common, atrial standstill	Yes, 30% of death
Limb-girdle type 1B	Lamin A and C	Conduction disease and DCM	>90%	VT, ICD indicated	Common	Yes, 30% of death
Limb-girdle type 2C–2F	Sarcoglycans	DCM	<25%	Uncommon	Limited data	Unknown
Limb-girdle type 2I	Fukutin-related protein	DCM	20–80%	Uncommon	Not reported	Unknown
Facioscapulohumeral	D4Z4 repeat contraction	Conduction disease	5–15%	Rare VTs	Rare	No

Muscular Dystrophies are an heterogeneous group of inherited diseases characterized by muscular and cardiac involvement, that occurs as a degenerative process with fibrosis and fatty replacement of the myocardium

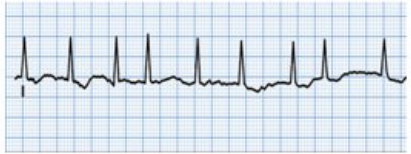
Duchenne Muscular Dystrophy Becker Muscular Dystrophy Limb Girdle type 2 Muscular Dystrophy	Myotonic Dystrophy type I Emery Dreifuss Muscular Dystrophy Erb's Muscular Dystrophy

Early Stage

Dilated cardiomyopathy



Conduction Disorders
Atrial Fibrillation /Flutter
Ventricular Tachycardia / Fibrillation



Late Stage

Arrhythmias and conduction disorders

Arrhythmia management, Cardiac Pacing and ICD indications follow the same rules for non ischemic DCM patients

Dilated cardiomyopathy

Special Indications for PMK/ICD implantation

International Journal of Cardiology 207 (2016) 284–285

Contents lists available at ScienceDirect

International Journal of Cardiology

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

Correspondence

Sudden cardiac death in neuromuscolar disorders: Time to establish shared protocols for cardiac pacing

Vincenzo Russo *, Anna Rago, Gerardo Nigro

Chair of Cardiology, Second University of Naples – Monaldi Hospital, Italy

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

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Claudia Stöllberger, MD,[29] Karim Wahbi, MD, PhD,[9]
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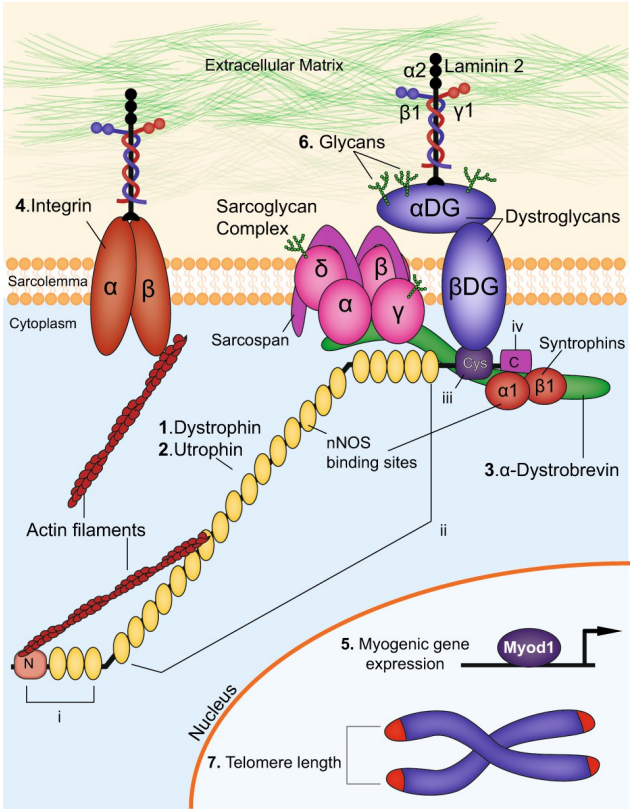

Duchenne Muscular Dystrophy
Becker Muscular Dystrophy
Limb Girdle type 2 Muscular Dystrophy

1	B-NR	3. In patients with DMD, BMD, or LGMD2, cardiac evaluation including physical examination, ECG, ambulatory ECG, and cardiac imaging (echocardiography or cardiac magnetic resonance imaging [CMR]) at diagnosis with periodic retesting is recommended even in the absence of cardiac symptoms.	40-44,46,47,59-69

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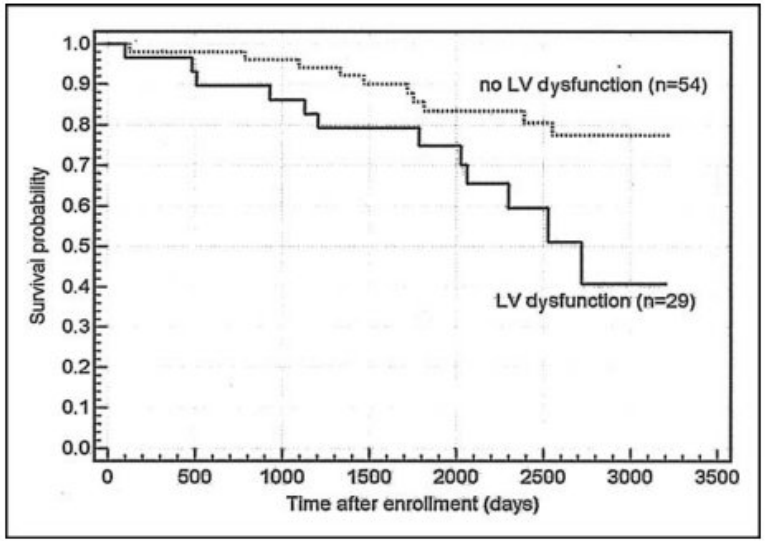
The presence of systolic dysfunction was a powerful predictor of mortality but ECG abnormalities, late potentials or ventricular arrhythmias were not predictive



Corrado G et al. Am J Cardiol. 2002;89:838

Contents lists available at ScienceDirect

Trends in Cardiovascular Medicine

journal homepage: www.elsevier.com

ACE inhibition to slow progression of myocardial fibrosis in muscular dystrophies

Vincenzo Russo MD, PhD, MMSc[a, *], Andrea Antonio Papa MD[a, b], Emmanuel Ato Williams MD[c], Anna Rago MD[a], Alberto Palladino MD[b], Luisa Politano MD, PhD[b], Gerardo Nigro, MD, PhD[a]

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- Use of ACEI or ARB for all neuromuscular disorder patients with reduced systolic cardiac function and suggests to consider such treatment before the onset of reduced systolic cardiac function in boys with DMD age ≥10.

- The relatively low risk of usage of ACE-Is and ARBs should encourage the consideration of earlier therapy.

Pacemaker

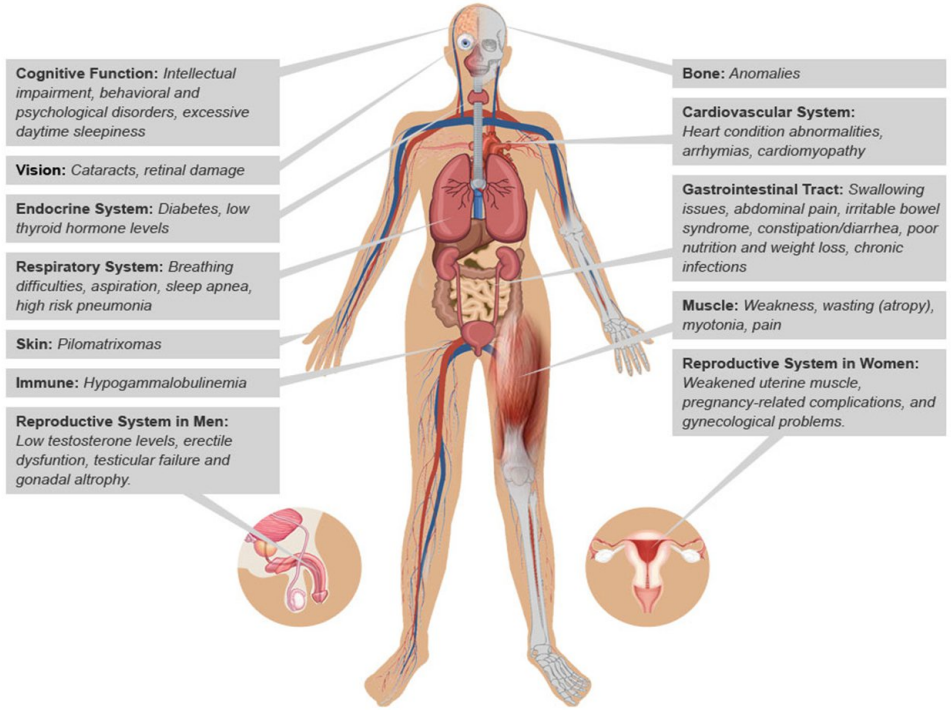
1	B-NR	1. In patients with DMD, BMD, or LGMD2 with documented symptomatic bradycardia due to any degree of sinus node dysfunction or AV block, permanent pacemaker implantation is indicated if concordant with the patient's goals of care and clinical status.	79-82
1	B-NR	2. In patients with DMD, BMD, or LGMD2 and third-degree or advanced second-degree AV block at any anatomical level, with or without symptoms, permanent pacemaker implantation is indicated if concordant with the patient's goals of care and clinical status.	79-81

ICD

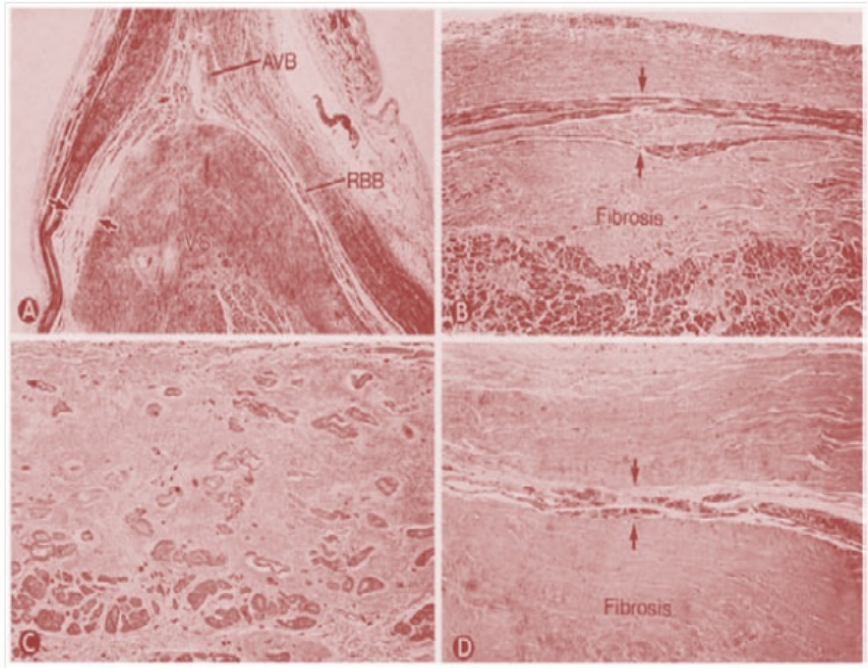
2a	B-NR	2. In patients with DMD, BMD, or LGMD2 with an LVEF ≤35% despite guideline-directed medical therapy, ICD therapy is reasonable if concordant with the patient's goals of care and clinical status.	68,97-99

Major Effects of Myotonic Dystrophy Type 1

www.myotonicdystrophy.com



Cardiac involvement, often preceding the skeletal muscle one, occurs in 80% of DM1 patients and represents the second most common cause of death, after respiratory causes



Cardiac involvement is due to interstitial fibrosis and fatty replacement in the specialized conduction system and in both atria and ventricles

- Atrio-Ventricular Blocks
  - *first degeee*
  - *second degree*
  - *third degree*
- Supraventricular Arrhytmias
  - *atrial fibrillation*
  - *atrial flutter*
  - *atrial tachycardia*
- Ventricular Dysfunction
- Ventricular Tachycardia

30%

3%

Petri H at al. Int J Cardiol 2012;160:82–88

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

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
1	C-LD	3. In patients with DM1 or DM2 and cardiac conduction disorder, close monitoring for arrhythmic complications is recommended when using mexiletine (or other sodium channel blockers).	116-119

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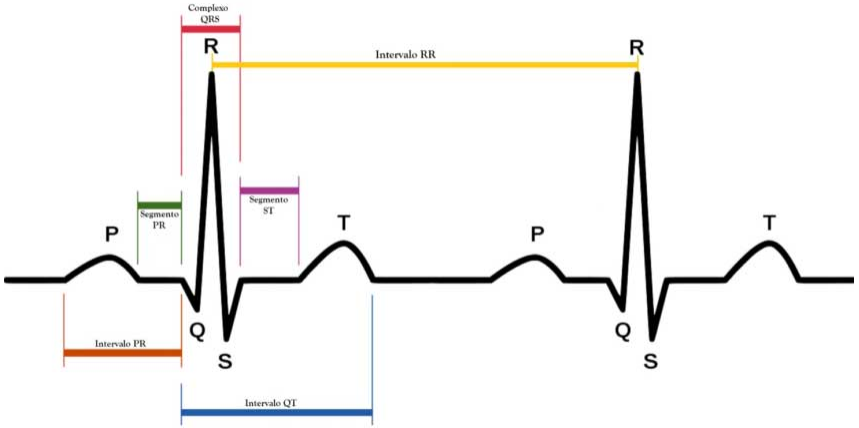
ANTOINE DE SAINT-EXUPÉRY
Il Piccolo Principe
BOMPIANI


Complesso QRS
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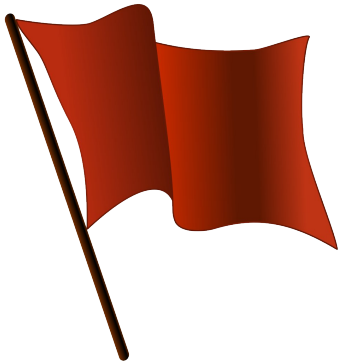
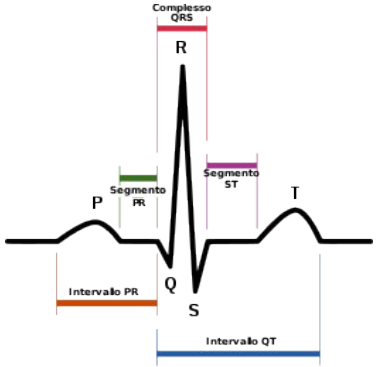
Intervallo QT





PR≥ 240 ms

QRS≥ 120 ms

The NEW ENGLAND JOURNAL of MEDICINE

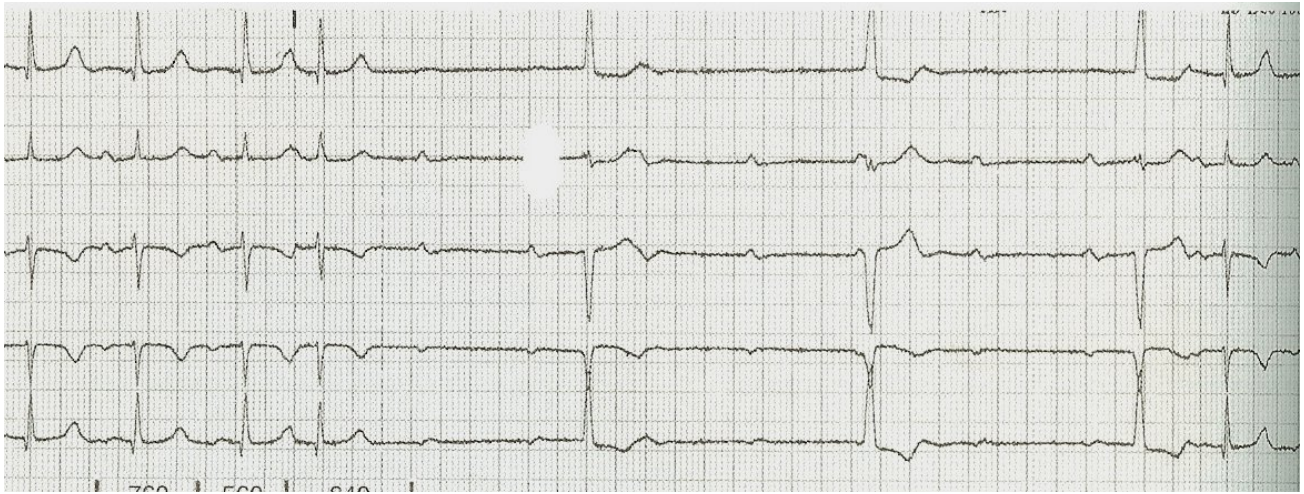
ORIGINAL ARTICLE

Electrocardiographic Abnormalities and Sudden Death in Myotonic Dystrophy Type 1

William J. Groh, M.D., M.P.H., Miriam R. Groh, M.S., Chandan Saha, Ph.D., John C. Kincaid, M.D., Zachary Simmons, M.D., Emma Ciafaloni, M.D., Rahman Pourmand, M.D., Richard F. Otten, M.D., Deepak Bhakta, M.D., Girish V. Nair, M.D., M.S., Mohammad M. Marashdeh, M.D., Douglas P. Zipes, M.D., and Robert M. Pascuzzi, M.D.

Characteristic	Sudden Death		Death from Progressive Neuromuscular Respiratory Failure		Death from Any Cause	
	Relative Risk (95% CI)	P Value	Relative Risk (95% CI)	P Value	Relative Risk (95% CI)	P Value
Age†	1.16 (0.76–1.75)	0.50	1.81 (1.20–2.74)	0.005	1.46 (1.13–1.87)	0.003
Muscular-impairment score‡						
1 or 2			1.0		1.0	
3			1.28 (0.20–8.19)	0.80	1.13 (0.47–2.73)	0.79
4			2.85 (0.60–13.55)	0.19	1.47 (0.67–3.19)	0.33
5			13.07 (2.77–61.61)	0.001	4.54 (2.02–10.19)	<0.001
Heart failure			5.39 (1.46–19.82)	0.01	2.85 (1.20–6.74)	0.02
Atrial tachyarrhythmia	5.18 (2.28–11.77)	<0.001	2.41 (1.10–5.31)	0.03	2.59 (1.55–4.32)	<0.001
Pacemaker	1.35 (0.51–3.56)	0.54	1.80 (0.77–4.20)	0.17	1.46 (0.82–2.60)	0.19
Ventricular tachyarrhythmia					6.41 (2.30–17.89)	<0.001
Severe ECG abnormality§	3.30 (1.24–8.78)	0.02	1.33 (0.53–3.33)	0.55	1.58 (0.90–2.81)	0.11

Groh *et al.* found that DM1 patients – even when asymptomatic – presenting with prolonged PR intervals (≥ 240 ms), wide QRS complex (≥ 120 ms), or atrial tachyarrhythmias were at higher risk of SCD when compared to those with normal ECGs



Because the risk of SCD most often appears to be associated to paroxysmal complete AV block, it has become common practice to implant a PM in DM1 patients when they have ECG abnormalities suggestive of advance conduction system disease, even if they are asymptomatic.

Groh WJ, N Engl J Med. 2008 Jun 19;358(25):2688-97

European Heart Journal (2017) 38, 751–758
doi:10.1093/eurheartj/ehw569

CLINICAL RESEARCH
Heart failure/cardiomyopathy

Incidence and predictors of sudden death, major conduction defects and sustained ventricular tachyarrhythmias in 1388 patients with myotonic dystrophy type 1

Karim Wahbi[1,2,3], Dominique Babuty[4], Vincent Probst[5], Ludivine Wissocque[6], Fabien Labombarda[7], Raphaël Porcher[8], Henri Marc Bécane[2], Arnaud Lazarus[9], Anthony Béhin[2], Pascal Laforêt[2,10], Tanya Stojkovic[2], Nicolas Clementy[4], Aurélie Pattier Dussauge[5,11], Jean Baptiste Gourraud[5], Yann Pereon[12], Arnaud Lacour[13], Françoise Chapon[14], Paul Milliez[7], Didier Klug[6], Bruno Eymard[2,10], and Denis Duboc[1,2,3]

Table 3 Multiple variable analysis of association between baseline characteristics and clinical outcomes

	Overall survival		Sudden death		Sustained ventricular tachyarrhythmia		Major conduction defect	
	Adjusted HR (95% CI)	P	Adjusted HR (95% CI)	P	Adjusted HR (95% CI)	P	Adjusted HR (95% CI)	P
Age, per year	1.07 (1.06–1.08)	<0.0001	1.05 (1.02–1.08)	0.0006	–	–	1.02 (1.01–1.04)	0.0005
Male sex	1.47 (1.12–1.93)	0.005	–	–	–	–	1.49 (1.05–2.11)	0.024
Family history of sudden death	0.99 (0.63–1.57)	0.98	2.82 (1.21–6.53)	0.016	–	–	1.07 (0.58–1.95)	0.84
History of								
Coronary artery disease	0.72 (0.40–1.30)	0.27	–	–	2.77 (0.64–11.9)	0.17	0.68 (0.30–1.52)	0.35
Atrial fibrillation	0.99 (0.69–1.42)	0.95	1.69 (0.76–3.77)	0.20	2.46 (0.89–6.77)	0.081	2.12 (1.41–3.17)	0.0003
Ventricular tachycardia								
Non-sustained	0.94 (0.45–1.96)	0.86	2.36 (0.69–8.08)	0.17	8.57 (2.59–28.3)	0.0004	1.86 (0.91–3.80)	0.090
Sustained	1.65 (0.51–5.33)	0.40	–	–	2.13 (0.36–12.7)	0.41	–	–
Major conduction defect	1.30 (0.65–2.62)	0.46	–	–	–	–	–	–
Syncope	1.60 (1.13–2.28)	0.008	–	–	–	–	2.39 (1.61–3.56)	<0.0001
Heart rate, per 10 bpm	1.17 (1.08–1.26)	<0.0001	1.16 (0.97–1.39)	0.099	–	–	1.10 (0.98–1.24)	0.095
1st degree atrioventricular block	1.73 (1.31–2.28)	0.0001	1.55 (0.79–3.04)	0.21	1.37 (0.54–3.48)	0.51	1.60 (1.12–2.30)	0.010
Bundle branch block				0.012	1.78 (0.57–5.55)	0.32	1.85 (1.18–2.91)	0.007
Left	1.40 (0.98–2.01)	0.063	2.57 (1.23–5.37)					0.010
Right	1.27 (0.84–1.93)	0.26	–	–	–	–	1.91 (1.17–3.11)	
Left ventricular ejection fraction <50%	1.06 (0.64–1.75)	0.82	0.93 (0.27–3.17)	0.90	1.92 (0.57–6.50)	0.30	1.59 (0.90–2.79)	0.11

CI = confidence interval; HR = hazard ratio.

Age, family history of SD and LBBB were independent predictors of SD

2a	B-NR	4. In patients with DM1 or DM2 and marked first-degree AV block (PR interval ≥240 ms) or intraventricular conduction delay (native QRS duration ≥120 ms), permanent pacemaker implantation is reasonable if concordant with the patient's goals of care and clinical status.	106

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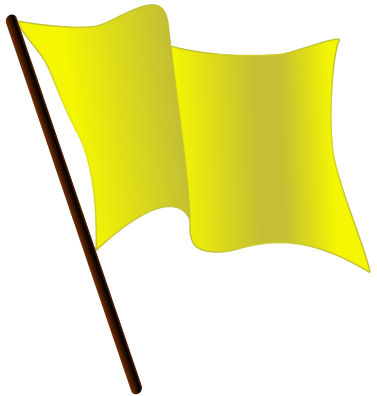
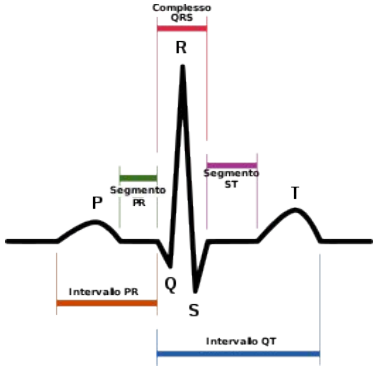
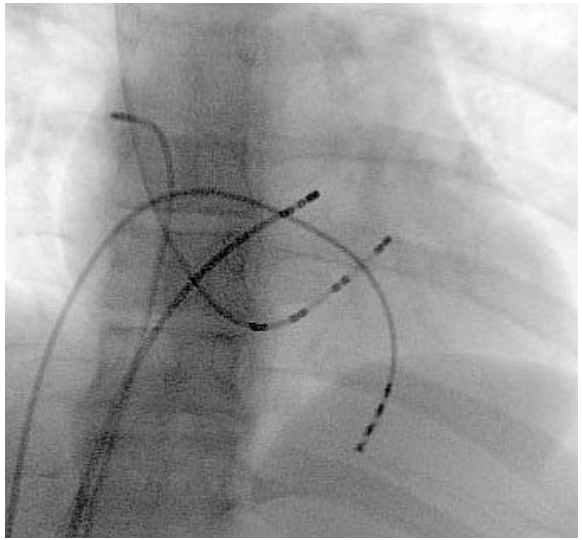


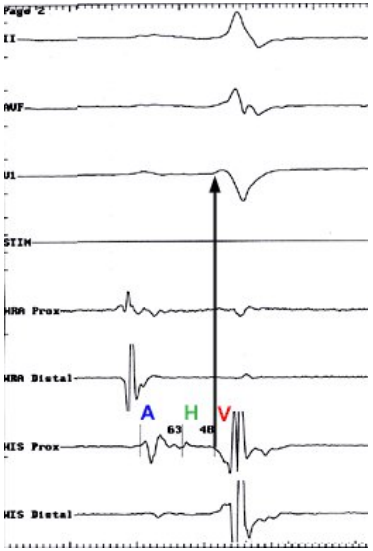


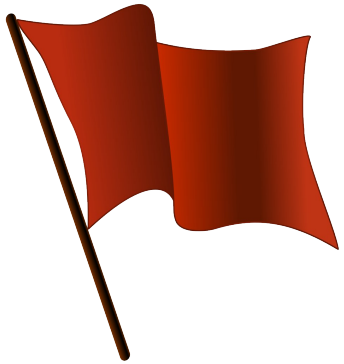
PR> 200 ms

QRS> 100 ms

2a	B-NR	4. In patients with DM1 or DM2 with symptoms consistent with bradycardia and with ECG evidence of mild to moderate conduction disorder and when noninvasive testing is nondiagnostic, electrophysiological testing is reasonable for risk stratification for AV block and sudden cardiac death.	26,120-123







HV> 70 ms

2a	B-NR	5. In patients with DM1 or DM2 with HV interval ≥70 ms on electrophysiological study, permanent pacemaker implantation is reasonable if concordant with the patient's goals of care and clinical status.	120,123-126

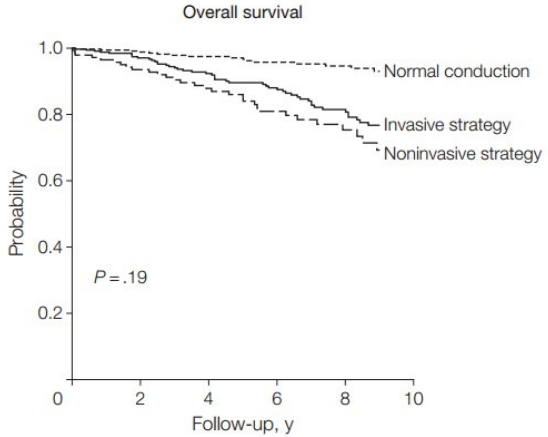
JAMA The Journal of the American Medical Association

JAMA. 2012 Mar 28;307(12):1292-301. doi: 10.1001/jama.2012.346.

Electrophysiological study with prophylactic pacing and survival in adults with myotonic dystrophy and conduction system disease.

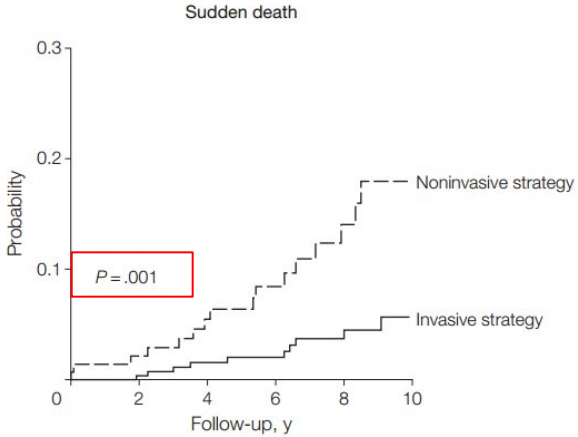
Wahbi K, Meune C, Porcher R, Bécane HM, Lazarus A, Laforêt P, Stojkovic T, Béhin A, Radvanyi-Hoffmann H, Eymard B, Duboc D.

Pierre et Marie Curie-Paris 6 University, Myology Institute, Pitié-Salpêtrière Hospital, 75013 Paris, France. karim.wahbi@cch.aphp.fr



No. at risk

Normal conduction	373	317	257	192	136
Invasive strategy	341	270	205	165	106
Noninvasive strategy	145	124	98	67	43



No. at risk

Invasive strategy	341	270	205	165	106
Noninvasive strategy	145	124	98	67	43

Article in Press

Role of electrophysiological evaluation for the best device choice to prevent sudden cardiac death in patients with Myotonic Dystrophy Type 1 and Emery Dreifuss Muscular Dystrophy

Vincenzo Russo, Luisa Politano, Gerardo Nigro

Pacemaker and Implantable Cardioverter-Defibrillator Use in a US Myotonic Dystrophy Type 1 Population

DEEPAK BHAKTA, M.D.,* CHANGYU SHEN, PH.D.,† JACK KRON, M.D.,‡
ANDREW E. EPSTEIN, M.D.,§ ROBERT M. PASCUZZI, M.D.,¶
and WILLIAM J. GROH, M.D., M.P.H.*

In the largest registry published to date, 406 DM1 were followed for 9.5 years; forty-six patients received pacemakers for conduction abnormalities and twenty one patients received ICDs primarily for LV dysfunction. During follow-up period the authors showed seven sudden cardiac deaths in the pacemaker group, and 6.5% of patients had sudden death due to ventricular tachyarrhythmias compared with no patients in the group who received ICDs.

For the high risk of VT/VF and sudden death the ICDs rather than pacemakers should be considered for these patients.

B. Bhakta et al. J Cardiovasc Electrophysiol 22 (2011) 1369–1375

1	B-NR	4. In patients with DM1 or DM2 in whom clinically relevant ventricular arrhythmias are induced during electrophysiological study, ICD therapy is recommended if concordant with the patient's goals of care and clinical status.	110,124-126,132,133
2b	B-NR	5. In patients with DM1 or DM2 in whom permanent pacemaker implantation is indicated, ICD therapy may be considered if concordant with the patient's goals of care and clinical status.	106,121,132

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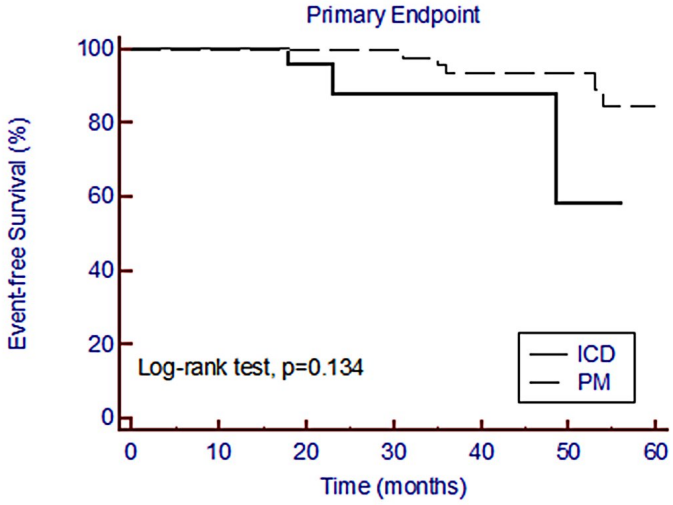
JOURNAL ARTICLE

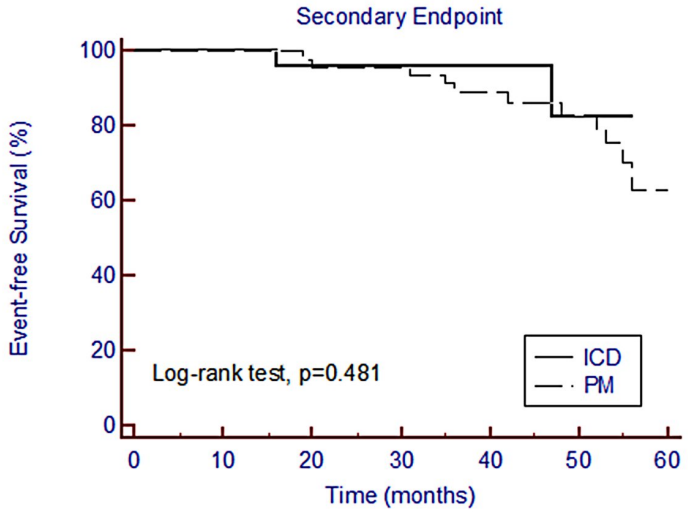
Arrhythmic CArdiac DEath in MYotonic dystrophy type 1 patients (ACADEMY 1) study: the predictive role of programmed ventricular stimulation

Get access

Vincenzo Russo, Andrea Antonio Papa ✉, Anna Rago, Carmine Ciardiello, Anna Maria Martino, Alessandra Stazi, Paolo Golino, Leonardo Calò, Gerardo Nigro

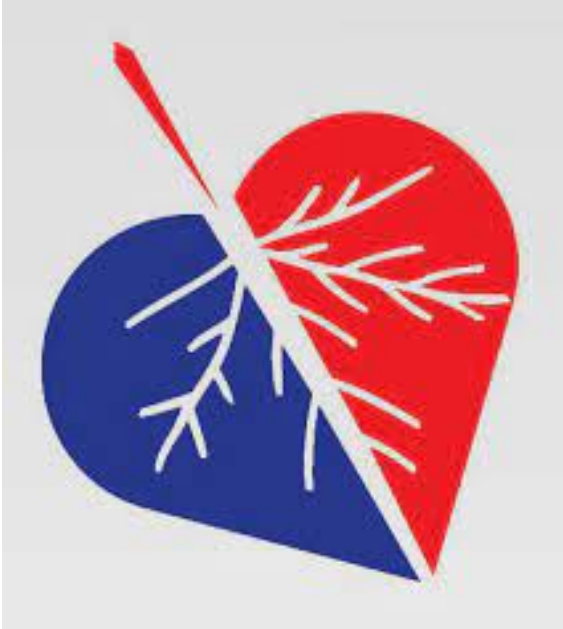
EP Europace, Volume 24, Issue 7, July 2022, Pages 1148–1155,
https://doi.org/10.1093/europace/euab282



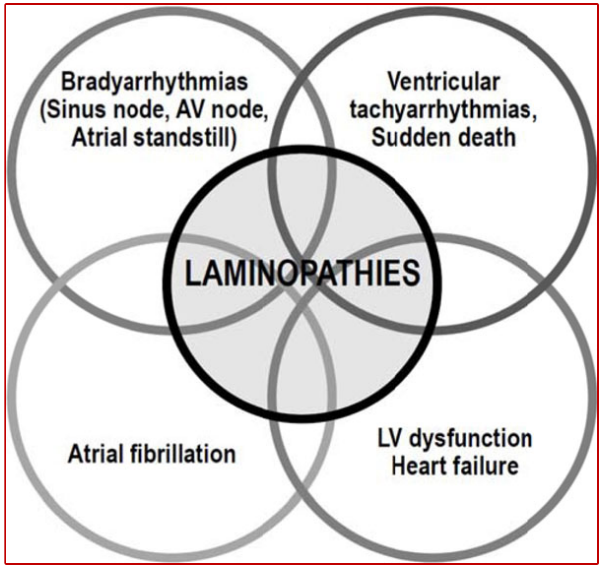


Low positive predictive value (16%) and sensitivity (44%)

High negative predictive value (90%) and specificity (67%)



LMNA-related Myopathies: *Cardiac Arrhythmias*



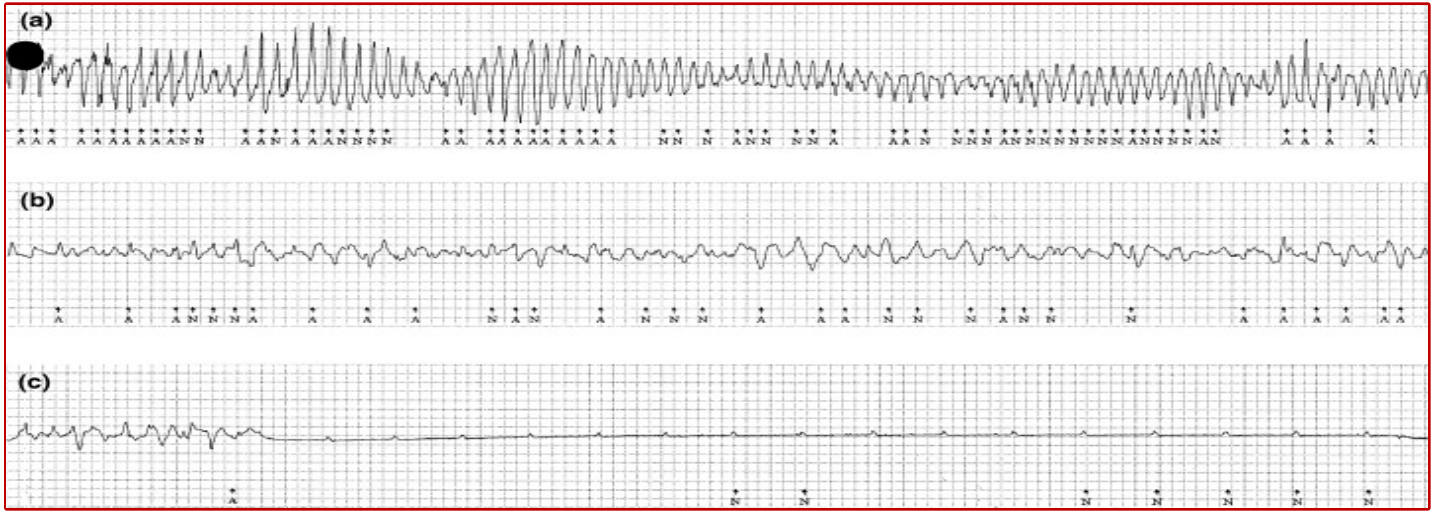
Patients with cardiolaminopathies may present a wide range of arrhythmic disorders, which include either:

Bradyarrhythmias
Atrial fibrillation
Ventricular tachyarrhtymias

in variable combinations, and with frequent association with left ventricular dysfunction and heart failure

➢Ventricular Tachyarrhythmias (VT/VF) can also occur in the early phase of the disease, before and independently of the development of ventricular dilatation.

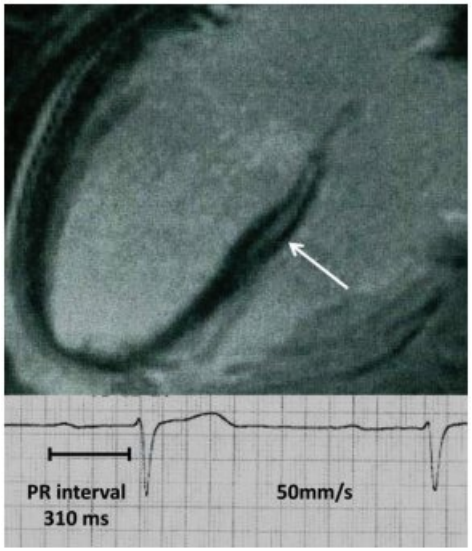
➢ Sudden death can be the first manifestation in some cases of VT/VF



ICD

1	B-NR	3. In patients with EDMD or LGMD1B with at least one of the following: second-degree or third-degree AV block, PR interval ≥230 ms, or spontaneous HV ≥70 ms, ICD therapy is recommended if concordant with the patient's goals of care and clinical status.	141,148,161-163
1	B-NR	5. In patients with EDMD or LGMD1B in whom clinically relevant ventricular arrhythmias are induced during electrophysiological study, ICD therapy is recommended if concordant with the patient's goals of care and clinical status.	124-126,141,155

Heart Rhythm Society SM



Europace (2014) 16, 563–571
doi:10.1093/europace/eut291

CLINICAL RESEARCH
Channelopathies

Risk prediction of ventricular arrhythmias and myocardial function in Lamin A/C mutation positive subjects

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Prolonged PR-interval was the best predictor of Ventricular Arrhythmias in LMNA mutation-positive subjects.

Prolonged PR-interval was associated with VA, reduced myocardial function and septal fibrosis in Lamin A/C mutation-positive subjects.

ICD

2a	B-NR	6. In patients with EDMD or LGMD1B with LVEF <45% and nonsustained VT, an ICD is reasonable if concordant with the patient's goals of care and clinical status.	29,146

Journal of the American College of Cardiology
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Published by Elsevier Inc.

Vol. 59, No. 5, 2012
ISSN 0735-1097/$36.00
doi:10.1016/j.jacc.2011.08.078

Genetic Disorders

Risk Factors for Malignant Ventricular Arrhythmias in Lamin A/C Mutation Carriers

A European Cohort Study

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Jens Mogensen, MD, PHD,# Johanna F. Hermans-van Ast, MS, PHD,*
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Arthur A. M. Wilde, MD, PHD,*† Andreas Perrot, MS,§§ Sabine Pankuweit, PHD,‖‖
Aeilko H. Zwinderman, MS, PHD,§ Philippe Charron, MD, PHD,‡‡ Yigal M. Pinto, MD, PHD*†

LMNA mutation carriers presenting with at least two of these four risk factors
Non-sustained VT, LVEF <45%, Male sex, Non-missense mutations
were at high risk to develop malignant ventricular arrhythmias

Recommendations for shared decision-making and end-of-life decisions			
COR	LOE	Recommendations	References
1	C-EO	1. In patients with NMDs who are considering or have a pacemaker or ICD, education on function including deactivation should be periodically discussed with the patient, their family members, and/or health care decision makers.	
1	C-EO	2. In patients with NMDs in whom the presence of conduction disorder portends a risk of ventricular arrhythmias, the decision of whether to implant a pacemaker or ICD should be concordant with the patient's overall medical care goals and clinical status.	
1	C-EO	3. In patients with NMDs who are considering ICD replacement and are undertaking advanced care planning, discussing the options of deferring ICD replacement is recommended.	
1	C-EO	4. In patients with NMDs who have an ICD and are undertaking advanced care planning, discussing the option of deactivation of ICD shock therapy is recommended.	
1	C-EO	5. In patients with NMDs who have an ICD and are experiencing ventricular arrhythmias with shocks refractory to available therapies, discussion of management of ICD therapy including shock deactivation is recommended, with careful attention to the patient's goals of care.	
2a	C-EO	6. In patients with NMDs who have a pacemaker or ICD and are nearing the end of life, if the patient or their health care decision maker requests pacing inactivation, it is reasonable to comply after education on the consequences of inactivation, with careful attention to the patient's goals of care.	

PLACE

PLATFORM OF LABORATORIES FOR ADVANCES IN CARDIAC EXPERIENCE

ROMA
Centro Congressi
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9ª Edizione

30 Settembre
1 Ottobre
2022

GRAZIE PER L'ATTENZIONE

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