I.R.C.C.S.
POLICLINICO SAN DONATO

PUNTO DI ASCOLTO TRA MALATI NEUROMUSCOLARI, MEDICI E RICERCATORI

Dal punto di vista aritmologico

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AF-ABLATION
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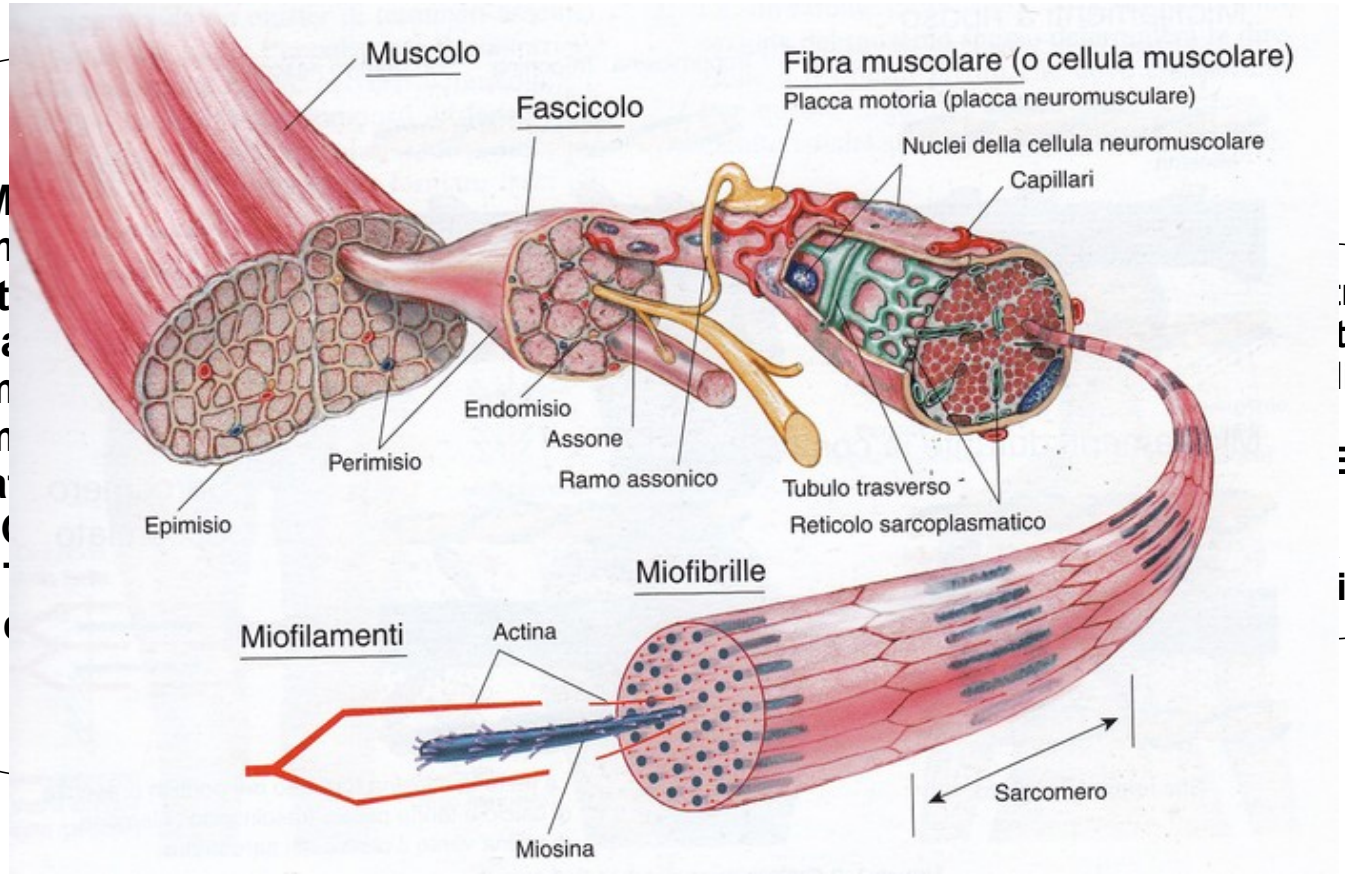
SUDDEN DEATH

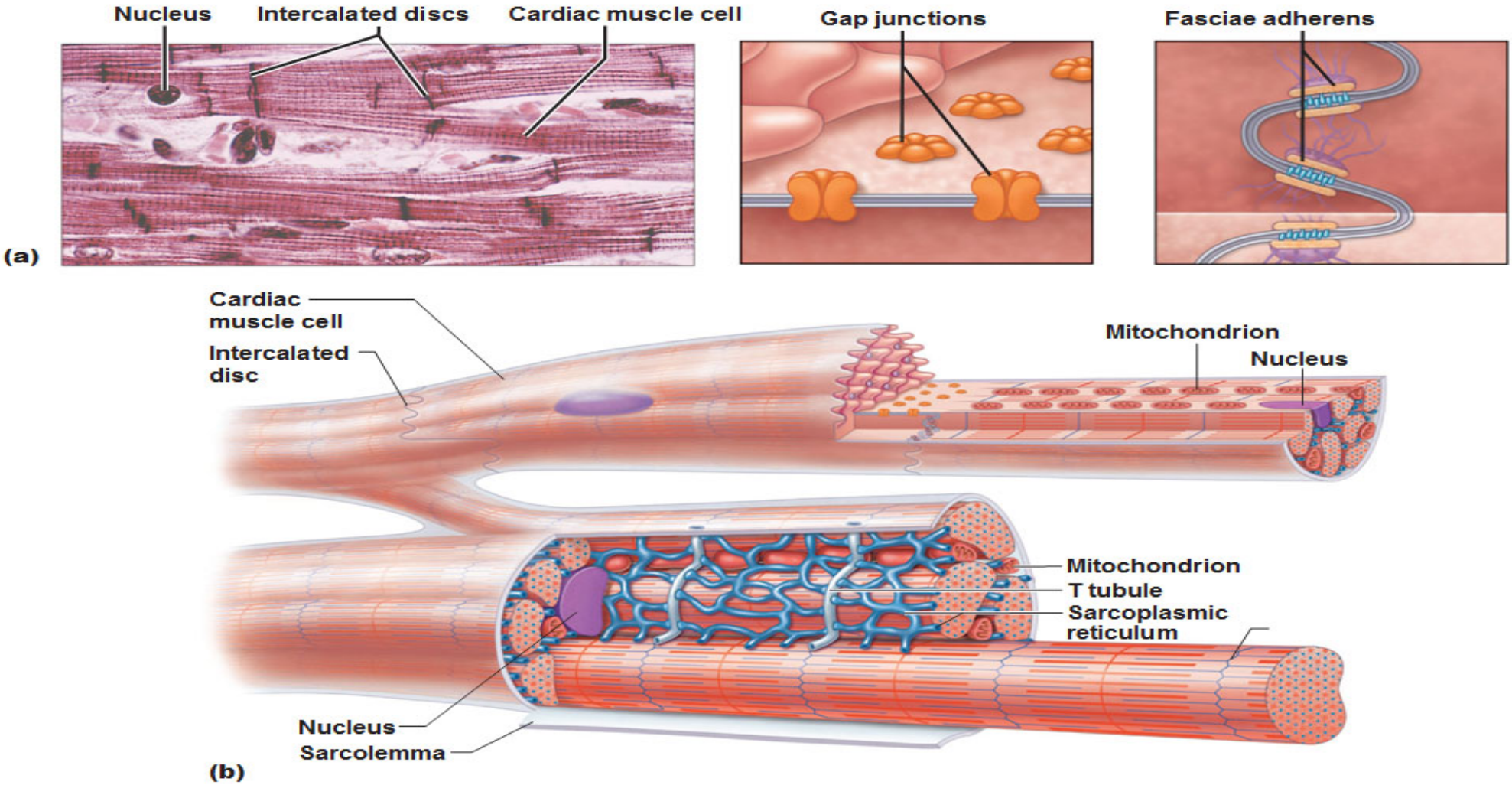
Distrofia M
Disordini m
Laminopat
Desminopa
Distrofia m
Distrofia m
Sclerosi la
Miastenia
Andersen
Sindrome



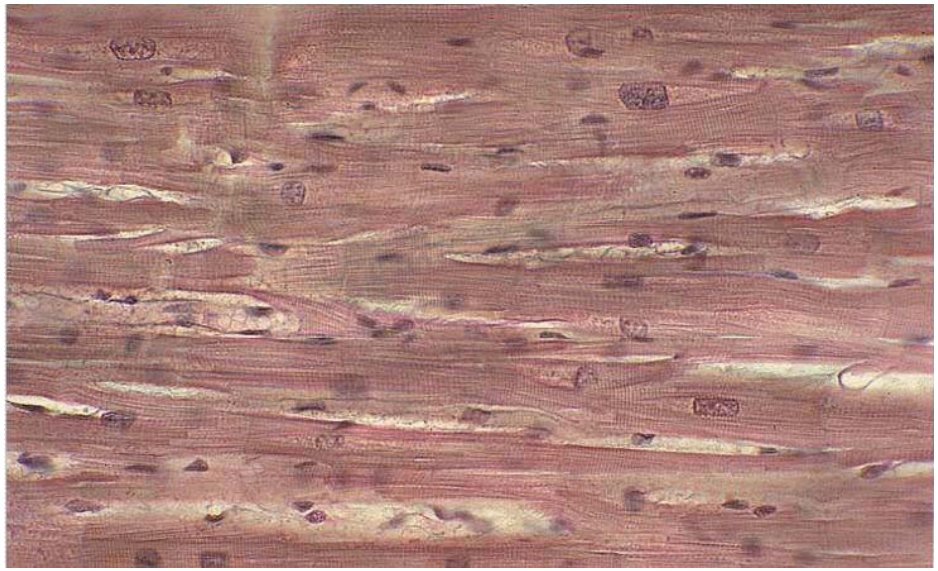
mogena
tion
ilatativa
QT lungo
Brugada

conduzione
icolari

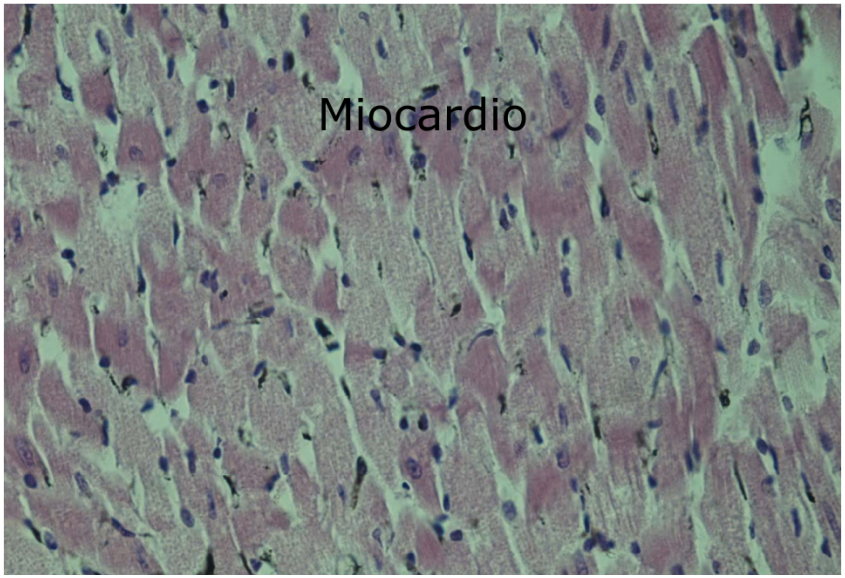
Hystology of the cardiac muscle



Miocardio Comune

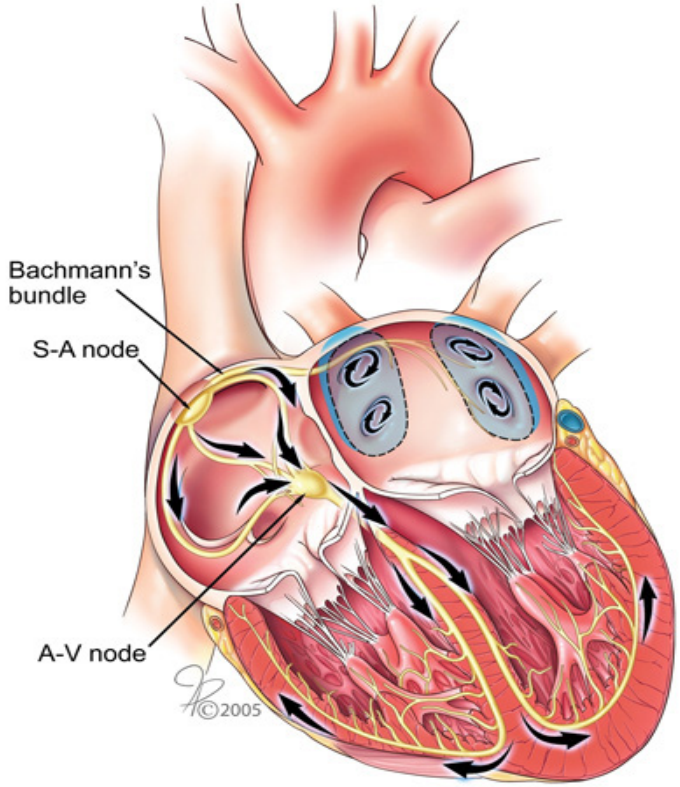


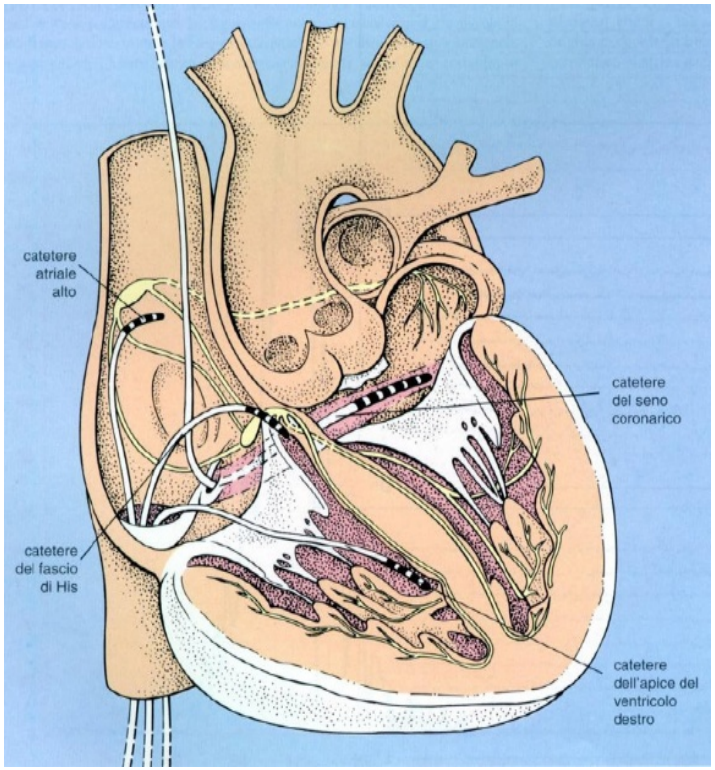
Miocardio Specifico



PUNTO DI ASCOLTO TRA MALATI NEUROMUSCOLARI, MEDICI E RICERCATORI

Dal punto di vista aritmologico





The Heart “Electrical” System

The normal electrical activity of the heart is physiologically made by:

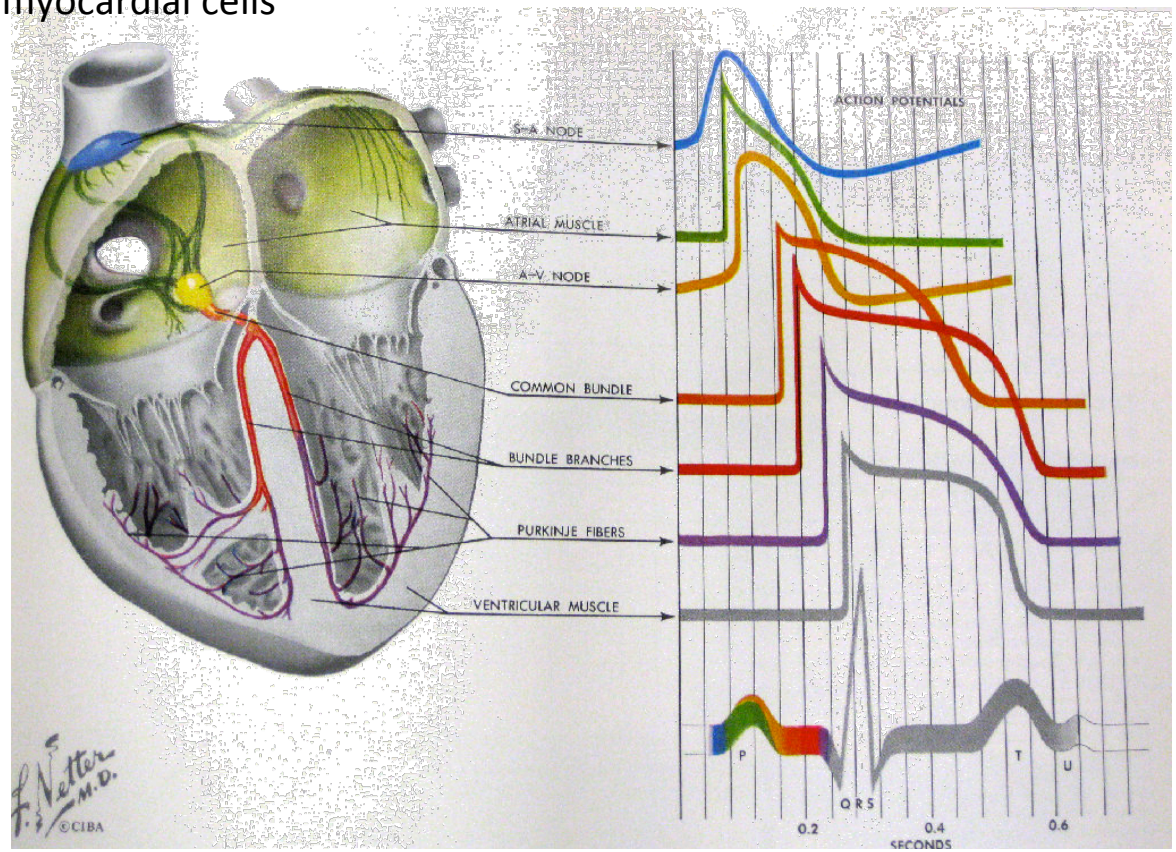
- Sino-Atrial Node (SAN)
- Atrio-Ventricular Node (AVN)
- His bundle
- Right Bundle
- Left Bundle
- Purkinje Network

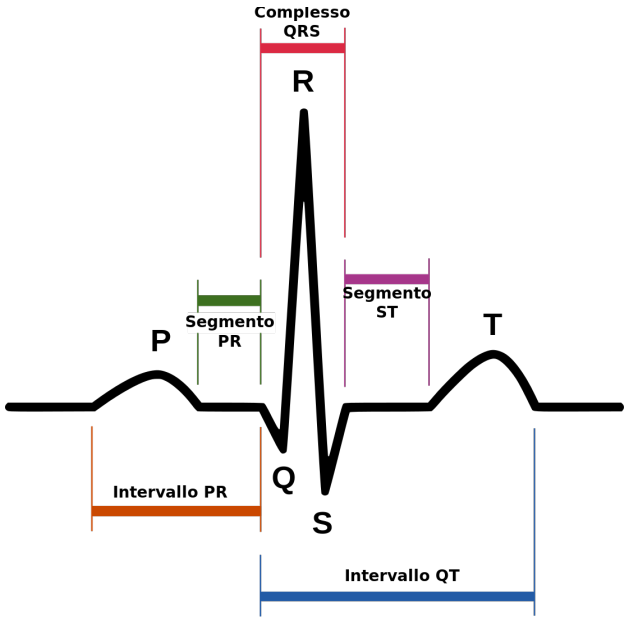
Trough electro-mechanical coupling these structures

coordinate the cardiac pump

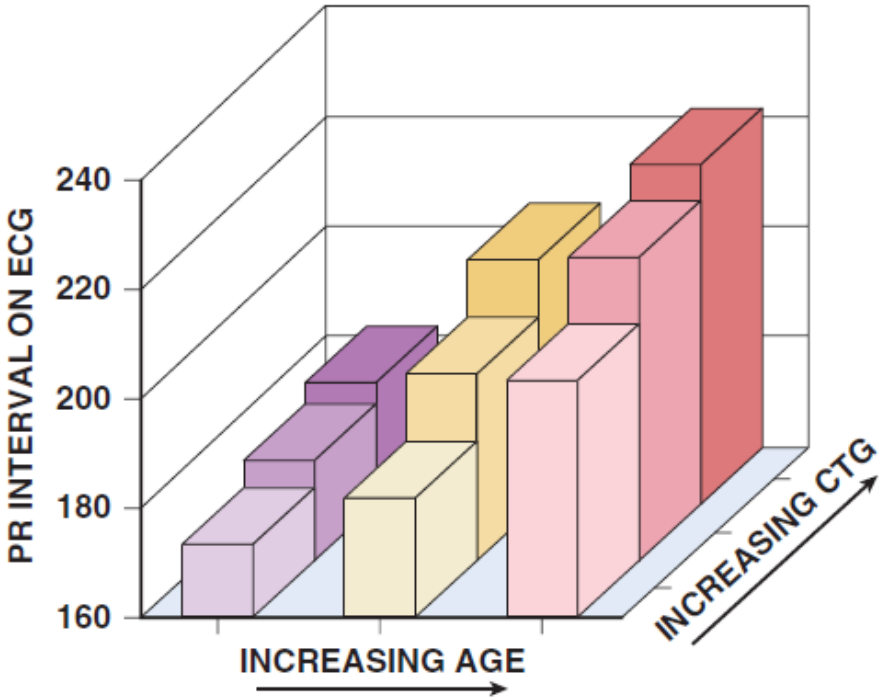
Electrophysiological Properties of Myocardial Cells

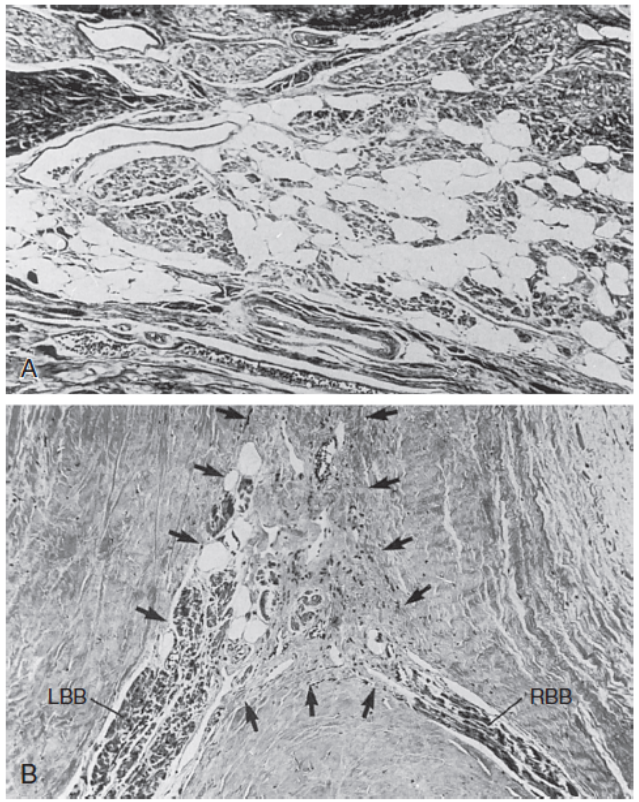
- Every cell type of the heart has a specific ion channel expression and electrical pattern
- Different Ion Currents lead to a specific electrophysiological behavior of the myocardial cells





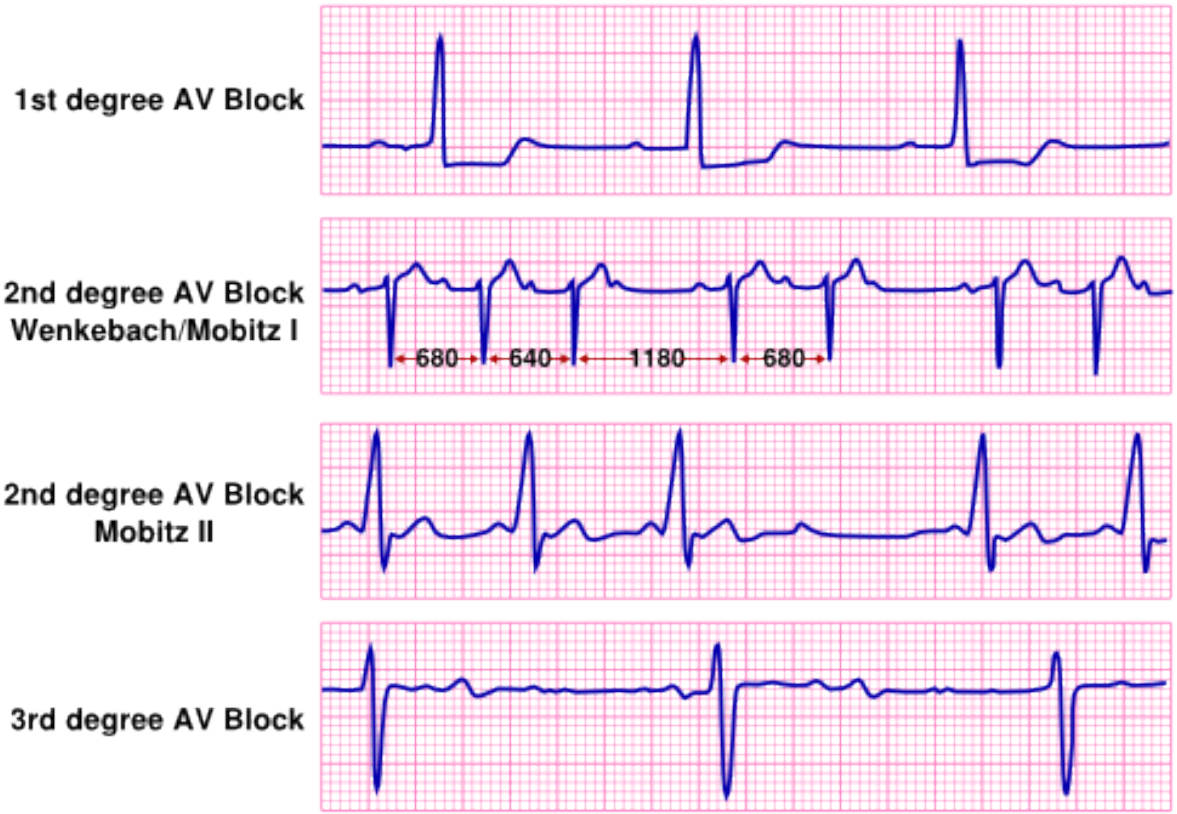
Myopathy	Gene	Heart involvement	Frequency of heart involvement	Ventricular arrhythmia	Atrial arrhythmia	Sudden death reported
Duchenne	Dystrophin	DCM	>90%	PVC	Only at late stage	Yes
Becker	Dystrophin	DCM	60–75%	VT associated with DCM	Associated with DCM	Yes
Myotonic, type 1	CGT repeat expansion	Conduction 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

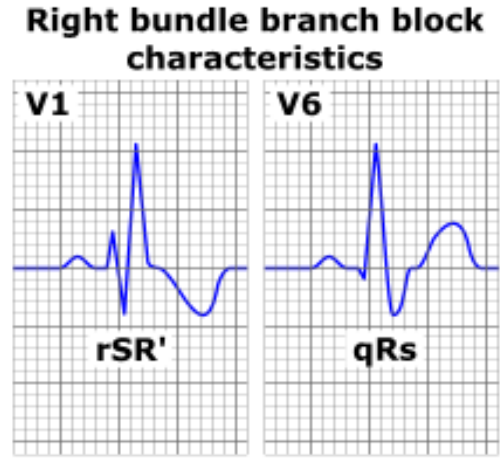


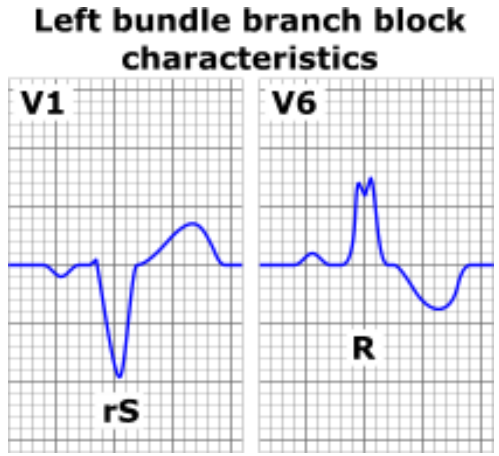


From Groh WJ, Lowe MR, Zipes DP: Severity of cardiac conduction involvement and arrhythmias in myotonic dystrophy type 1 correlates with age and CTG repeat length. J Cardiovasc Electrophysiol 13:444,2002

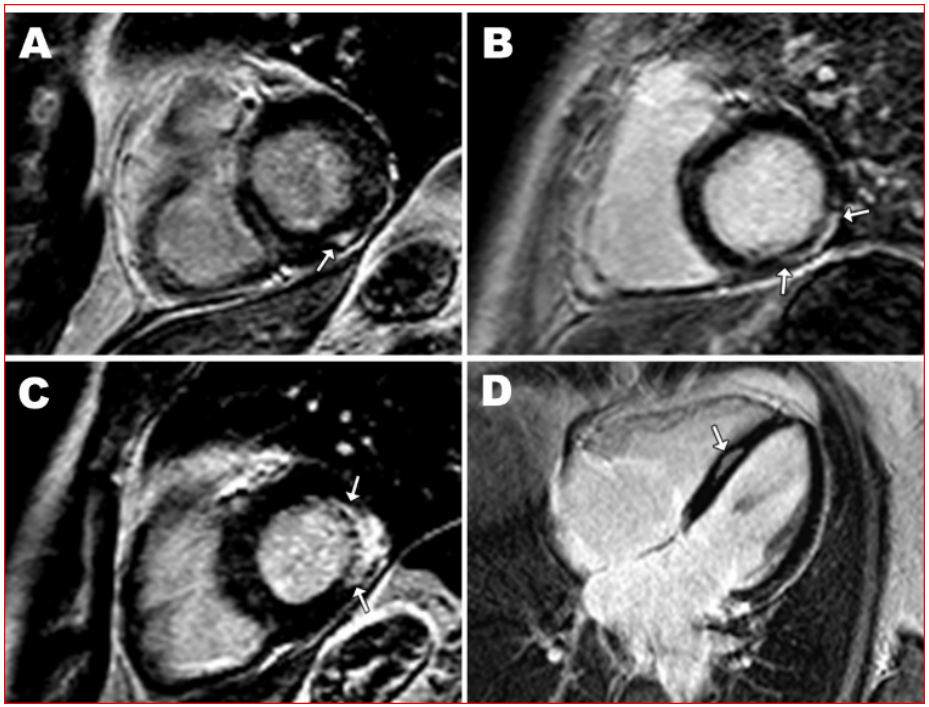
Quando è interessato il sistema di conduzione



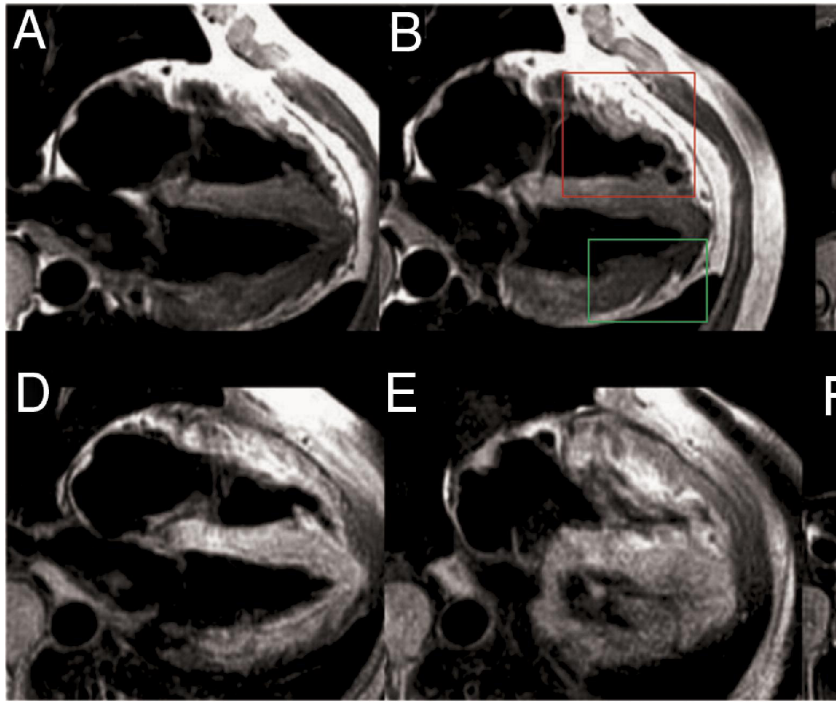




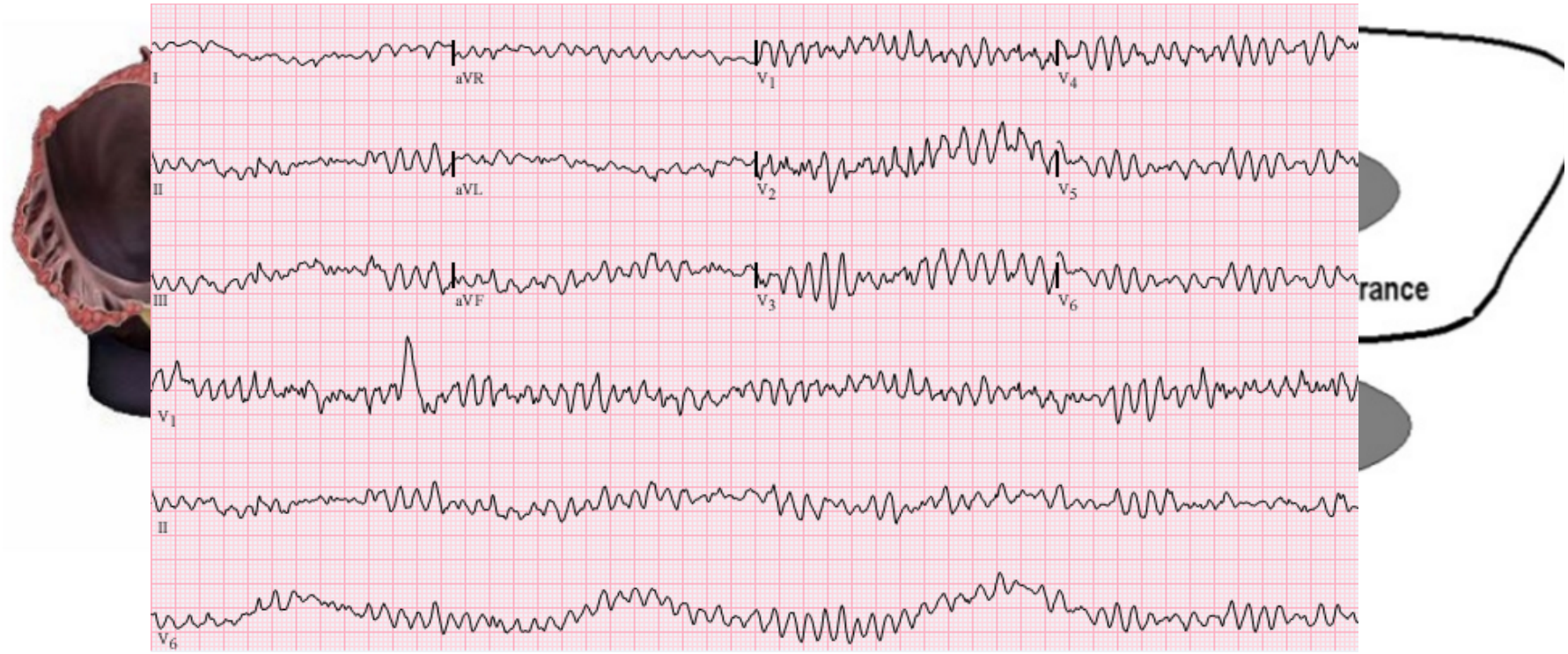
Distrofia miotonica

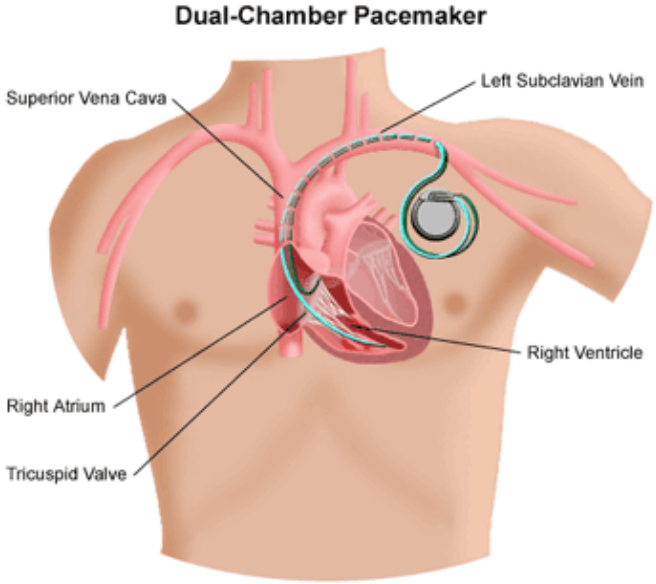


Displasia aritmogena



Ventricular Tachycardia Associated with Structural Heart Disease: Scar Based




Dual-Chamber Pacemaker
Superior Vena Cava
Left Subclavian Vein
Right Ventricle
Right Atrium
Tricuspid Valve

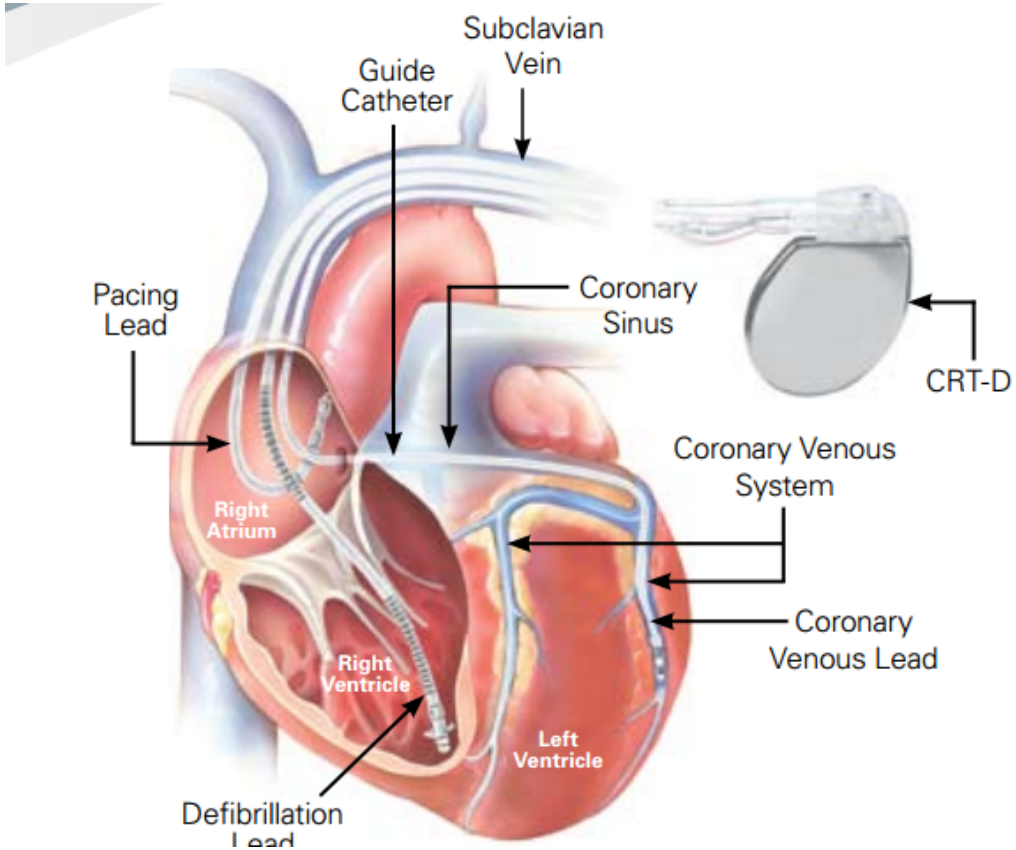
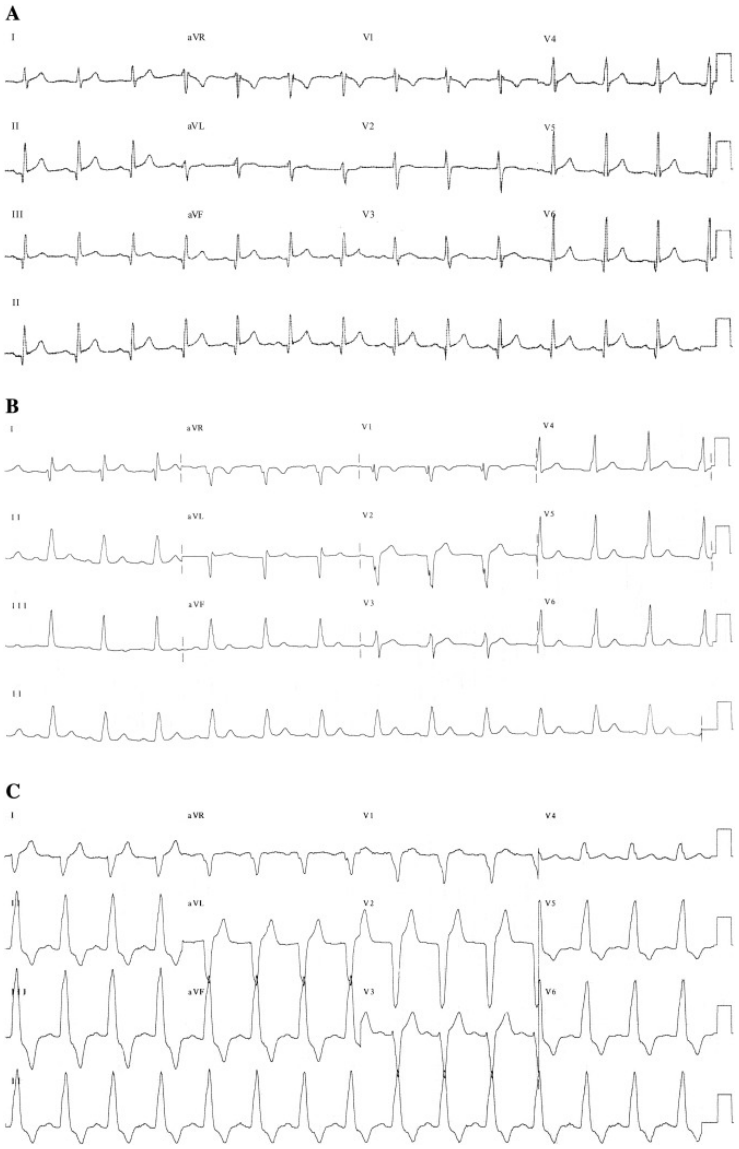
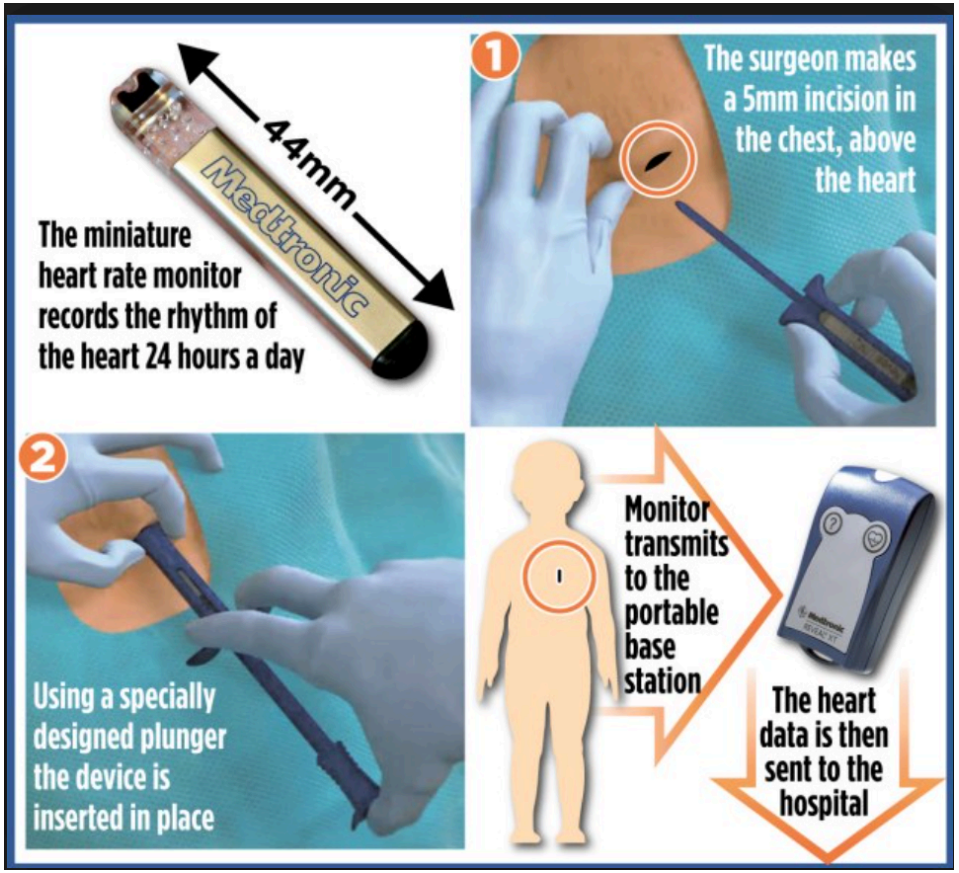

Subclavian Vein
Guide Catheter
Pacing Lead
Coronary Sinus
CRT-D
Coronary Venous System
Right Atrium
Coronary Venous Lead
Right Ventricle
Left Ventricle
Defibrillation Lead

Table 1
Electrocardiogram.

Parameter	Findings in MD	Reference
Conduction intervals and atrial arrhythmias	PR >240 ms, QRS >120 ms, second or third degree heart block ("Groh criteria") or a history of atrial tachyarrhythmias: • Associated with increased risk of mortality (cardiac and all cause) • Conduction abnormalities are associated with LV impairment, heart failure and family history of sudden death • Conduction abnormalities predict pacemaker implantation and atrial tachyarrhythmias	[20,26,28,83,84]
PR and QRS combined	PR and QRS combined duration >320 ms predicted long-term (17 years) mortality	[85]
QTc	QTc interval >450 ms associated >3-fold increased risk of sudden death or need for pacemaker implantation on long-term follow-up	[8]



Int J Cardiol. 2015 Apr 1;184:600-8. doi: 10.1016/j.ijcard.2015.03.069. Epub 2015 Mar 5


44mm
Medtronic
The miniature heart rate monitor records the rhythm of the heart 24 hours a day
1
The surgeon makes a 5mm incision in the chest, above the heart
2
Using a specially designed plunger the device is inserted in place
Monitor transmits to the portable base station
The heart data is then sent to the hospital

Age

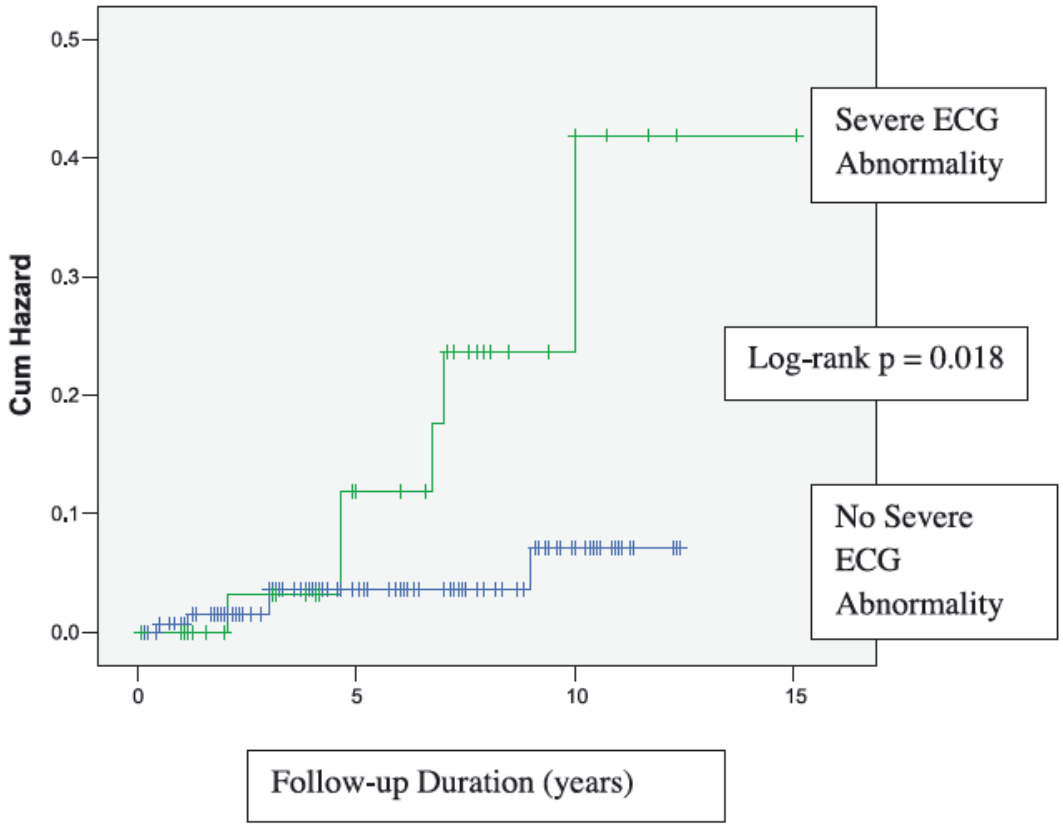
Hypertension

History of atrial tachycardia or atrial fibrillation

Family history of pacemaker
Family history of sudden cardiac death

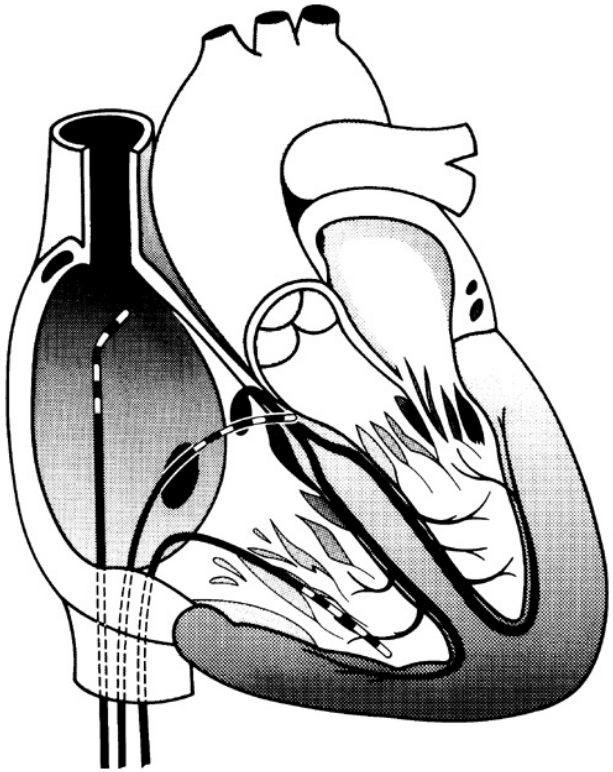
Baseline Characteristics of Patients with Myotonic Dystrophy Type-1 with and without a Severe ECG Abnormality

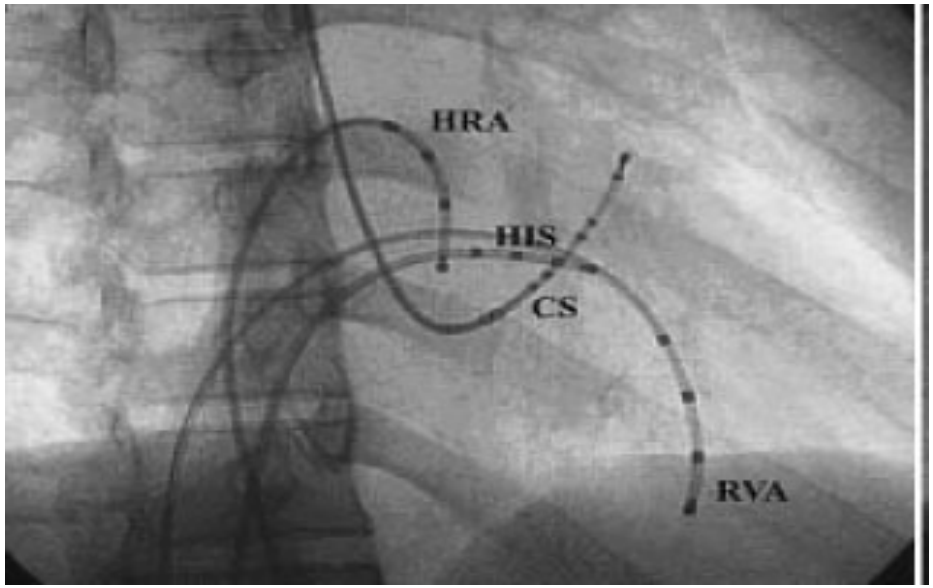
Characteristic	Patients with a Severe ECG Abnormality (n = 45)	Patients without a Severe ECG Abnormality (n = 144)	Univariate P-value
Age	41.6 ± 14.6	35.4 ± 12.6	0.006
Female	48.9%	54.9%	0.48
Hypertension	13.3%	4.2%	0.038
Diabetes mellitus	11.1%	5.6%	0.20
Coronary artery disease	6.7%	1.4%	0.09
History of heart failure	4.4%	0.0%	0.056
History of atrial tachycardia or atrial fibrillation	6.6%	0.7%	<0.001
Cataracts	80.0%	72.9%	0.34
Modafinil	28.9%	34.0%	0.52
Beta-blocker or calcium channel blocker	8.9%	3.5%	0.29
Family history of pacemaker	20.0%	0.7%	<0.001
Family history of sudden cardiac death	26.7%	5.6%	<0.001
Syncope or presyncope	6.7%	2.1%	0.27
Palpitations	6.7%	1.4%	0.01
Leg extension (kg)	116.2 ± 65.6	113.4 ± 63.5	0.83
Right grip strength (kg)	16.7 ± 11.7	17.0 ± 11.9	0.89
Left grip strength (kg)	15.7 ± 11.4	16.6 ± 11.7	0.70
PR interval (ms)	219 ± 64	174 ± 23	<0.001
QRS width (ms)	127 ± 26	96 ± 10	<0.001
QTc* (ms)	437 ± 34	411 ± 24	<0.001
RBBB	20.0%	2.1%	<0.001
LBBB	6.7%	0.0%	0.004
LAFB	13.3%	6.3%	0.13
Average HR—Holter (bpm)	69 ± 9	69 ± 8	0.95
HV Interval	71 ± 17	49 ± 6	0.04
Left ventricular ejection fraction (%)	56.6 ± 5.1	60.1 ± 4.9	0.01
CTG repeat length	689 ± 451	474 ± 322	0.01

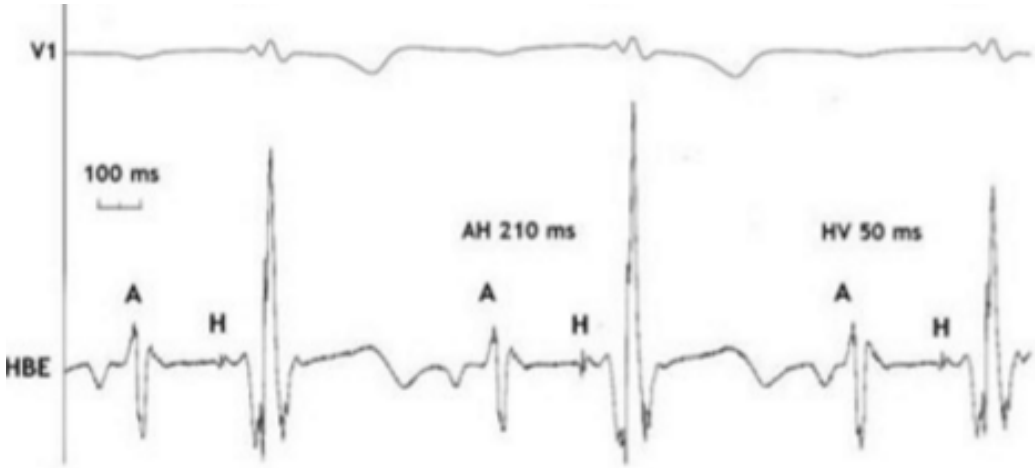


All-cause mortality in patients with myotonic dystrophy type 1, classified by the presence or absence of severe electrocardiographic abnormality.

Lo studio elettrofisiologico

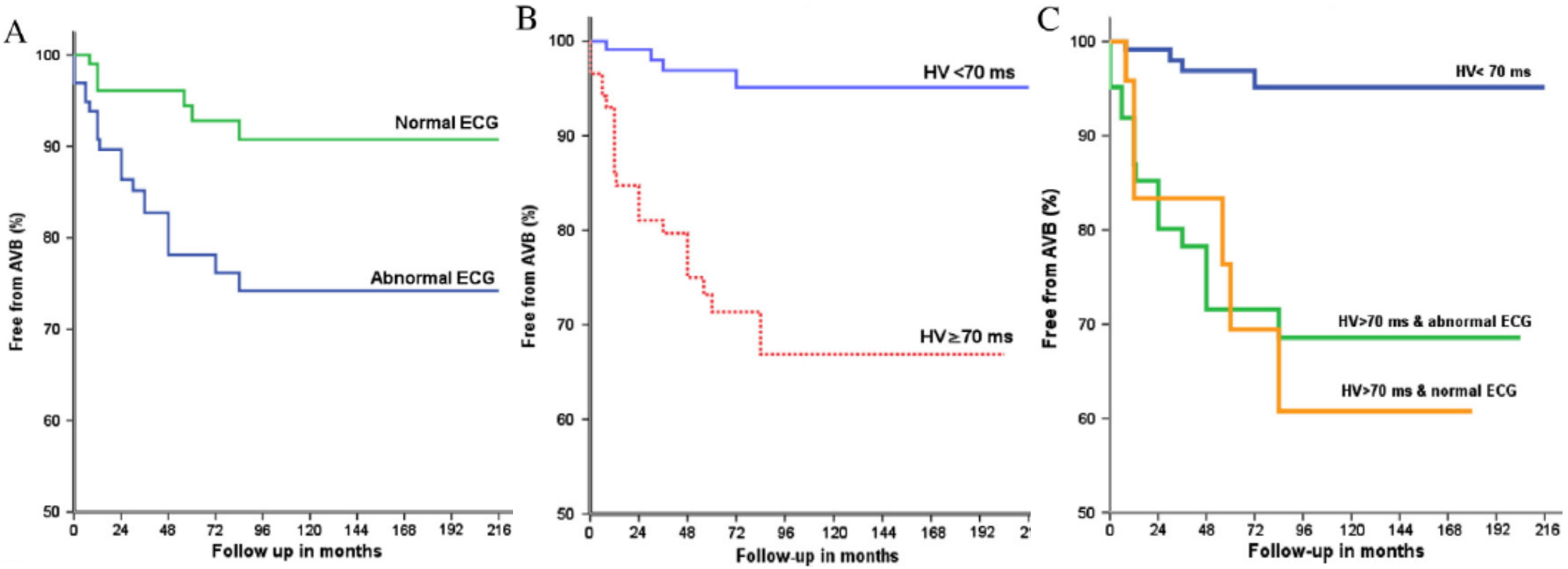






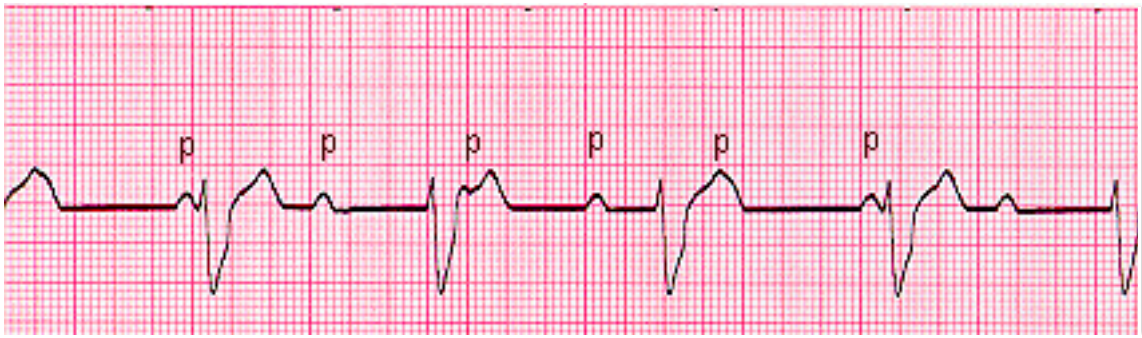
Insufficiency of electrocardiogram alone in predicting infrahisian abnormalities in patients with type 1 myotonic dystrophy

Edouard Siméon [a], Aurélie Patier-Dussauge [b], Anne Bernard-Brunet [a], Nicolas Clémenty [a], Jean-Baptiste Gouraud [b], Béatrice Guyomarch [b], Armelle Magot [b], Vincent Probst [b], Dominique Babuty [a,*]

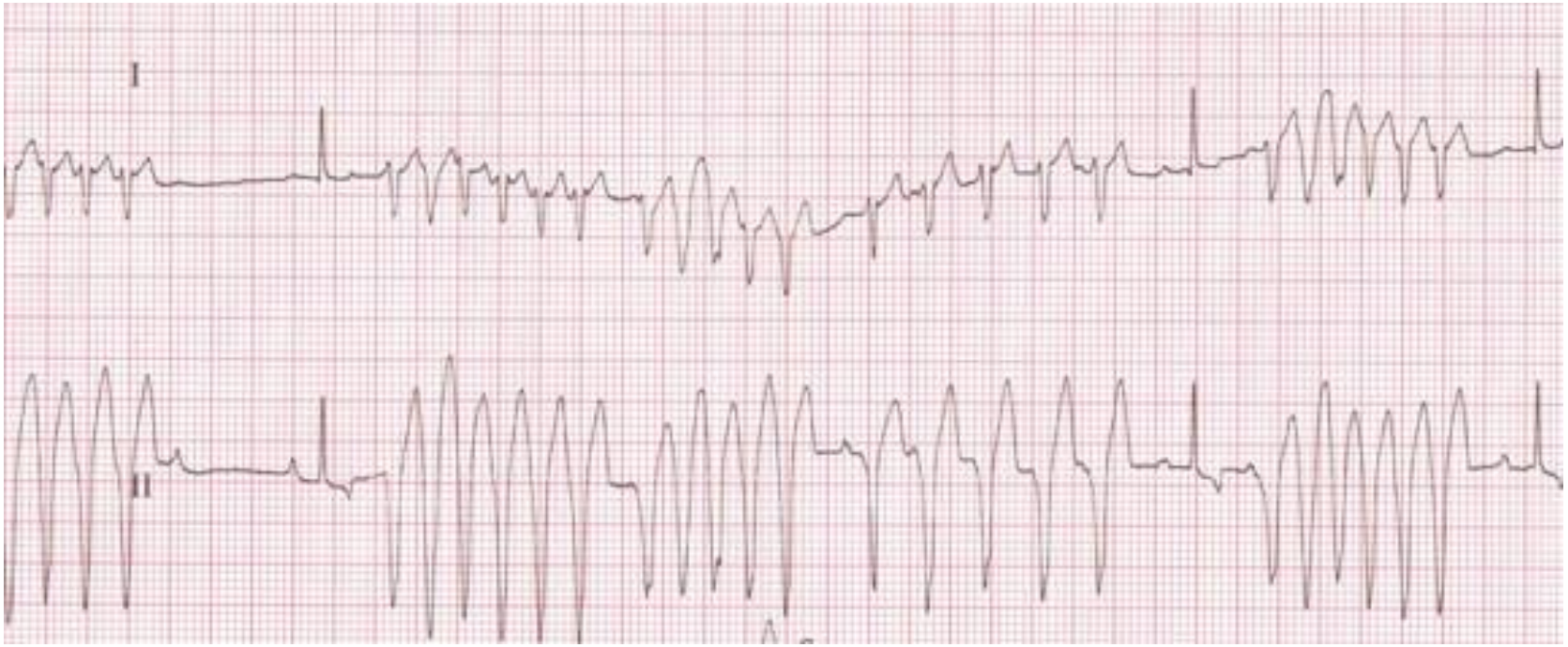


2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

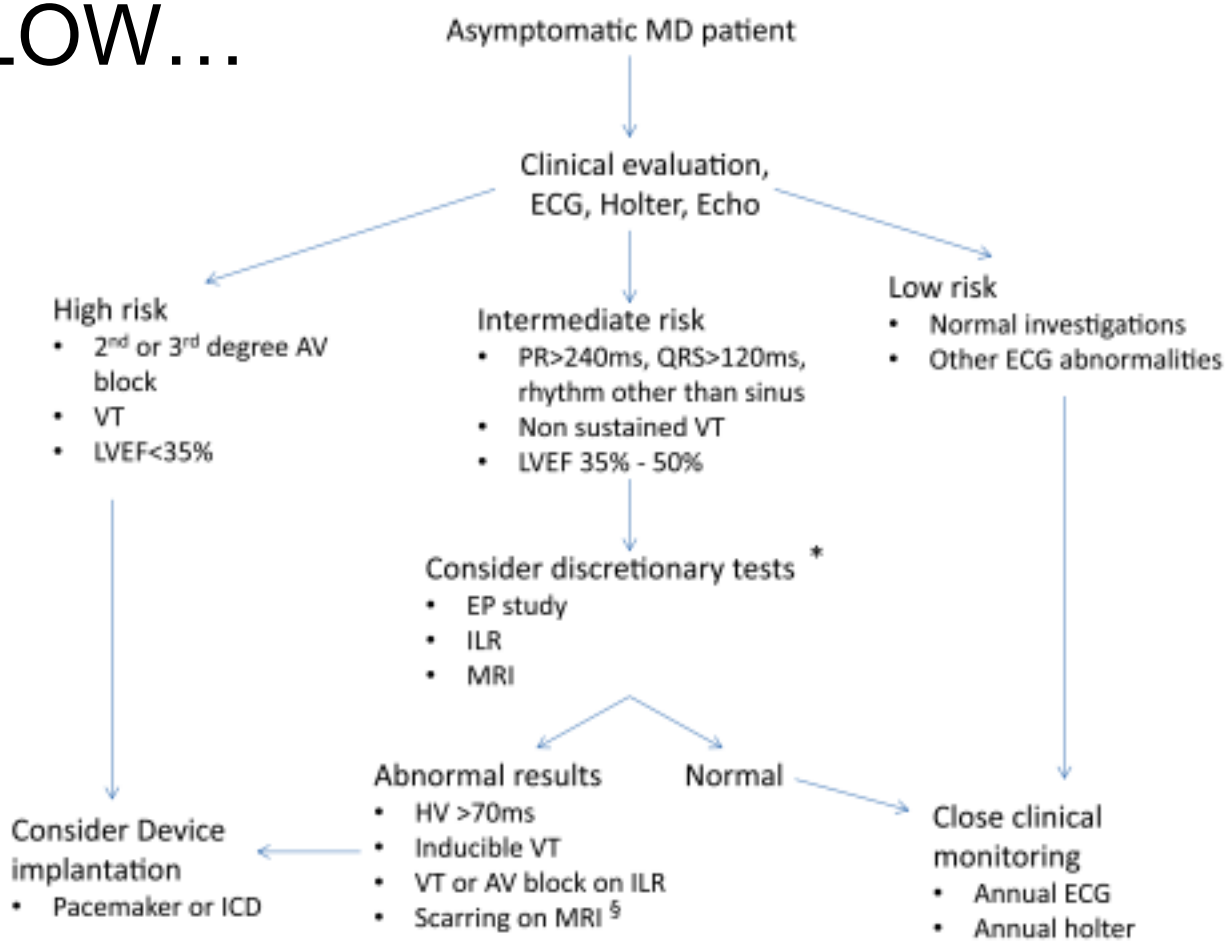
Permanent pacemaker implantation is recommended in patients with neuromuscular diseases and third-degree or advanced second-degree AV block at any anatomical level.	I	B	669
Permanent pacemaker implantation may be considered in patients with myotonic dystrophy type 1 (Steinert disease), Kearns–Sayre syndrome or limb-girdle muscular dystrophy with any degree of AV block (including first-degree) in consideration of the risk of rapid progression.	IIb	B	666, 669–672



The use of an ICD may be considered in myotonic dystrophy type 1 (Steinert disease), Emery–Dreifuss and limb-girdle type 1B muscular dystrophies when there is an indication for pacing and evidence of ventricular arrhythmias.	IIb	B



WORKFLOW…


Asymptomatic MD patient
Clinical evaluation, ECG, Holter, Echo
High risk
• 2nd or 3rd degree AV block
• VT
• LVEF<35%
Intermediate risk
• PR>240ms, QRS>120ms, rhythm other than sinus
• Non sustained VT
• LVEF 35% - 50%
Low risk
• Normal investigations
• Other ECG abnormalities
Consider discretionary tests *
• EP study
• ILR
• MRI
Abnormal results
• HV >70ms
• Inducible VT
• VT or AV block on ILR
• Scarring on MRI §
Normal
Consider Device implantation
• Pacemaker or ICD
Close clinical monitoring
• Annual ECG
• Annual holter

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Dal punto di vista aritmologico

Conclusioni:

- L' ECG rappresenta lo strumento essenziale di valutazione iniziale del paziente
- Il confronto di ECG seriati consente una valutazione semplice e non invasiva di un eventuale coinvolgimento del sistema di conduzione
- In casi selezionati può essere necessario effettuare una valutazione invasiva o richiedere l utilizzo di sistemi di monitoraggio impiantabili
- Il ruolo dell'imaging ed in paritcolare della RMN dovrebbe essere maggiormente ampliato
- Raccolta dei dati e la pubblicazione di registri ci aiuterà in futuro a meglio identificare i pazienti che richiedono un intervento e a scegliere il tipo di terapia o dispositivo che meglio si adatta a loro