



**KHUYẾN CÁO
QUẢN LÝ CÁC BỆNH CƠ TIM
ESC 2023
CÁCH NHÌN HỆ THỐNG**

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American Heart Association

2019 HRS expert consensus statement on evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy  

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Diagnosis and Treatment of Cardiomyopathy
American Heart Association Joint

STATE OF THE ART REVIEW
Heart failure and cardiomyopathies

Restrictive cardiomyopathy: definition and diagnosis

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Khuyến cáo đầu tiên thống nhất về các bệnh cơ tim



cardiomyopathy
phenotypes and their
aetiology features that should



ESC GUIDELINES

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2023 ESC Guidelines for the management of cardiomyopathies

Developed by the task force on the management of cardiomyopathies of the European Society of Cardiology (ESC)

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2023 ESC Guidelines for the management of cardiomyopathies		
Noonan syndrome	I	B
Ident genetic diagnosis (i.e. a P/LP variant) with cardiomyopathy in the family (starting with cascading out sequentially).	IIa	B
Post-test counselling should be considered in genetic diagnosis (i.e. a P/LP variant) has cardiomyopathy in the family (starting with cascading out sequentially), considering the age of onset, presentation in the family, and		

32 key messages

- ❖ What to do
- ❖ What not to do

Những điểm chính

1. Phân loại BCT
2. Cách tiếp cận chẩn đoán các BCT
3. Tiếp cận người bệnh theo từng tình huống
4. Khác biệt trong quản lý BN BCT so với BN ST / RL nhịp
5. Vai trò của các phương pháp chẩn đoán hình ảnh
6. BCT: một biểu hiện trong một hội chứng
7. Vai trò của xét nghiệm gen – tư vấn di truyền

Những điểm chính ...

8. BCT ở trẻ nhỏ và trẻ sơ sinh
9. Cập nhật về BCT phì đại có tắc nghẽn ĐRTT
10. Cập nhật điều trị BCT giãn
11. BCT do chuyển hóa: giá trị của xác định bệnh căn
12. BCT và thai nghén
13. BCT và vận động thể lực
14. BCT nguy cơ cao và đột tử

4 nội dung nổi bật



1. Phân loại BCT

Định nghĩa bệnh cơ tim

- Tình trạng bất thường về cấu trúc và chức năng cơ tim không do nguyên nhân BMV, THA, bệnh van tim, bệnh TBS
- Áp dụng ở cả người lớn và trẻ em, không phân biệt nguyên nhân do di truyền/ mắc phải

Table 3 Morphological and functional traits used to describe cardiomyopathy phenotypes

Morphological traits

Ventricular hypertrophy: left and/or right

Ventricular dilatation: left and/or right

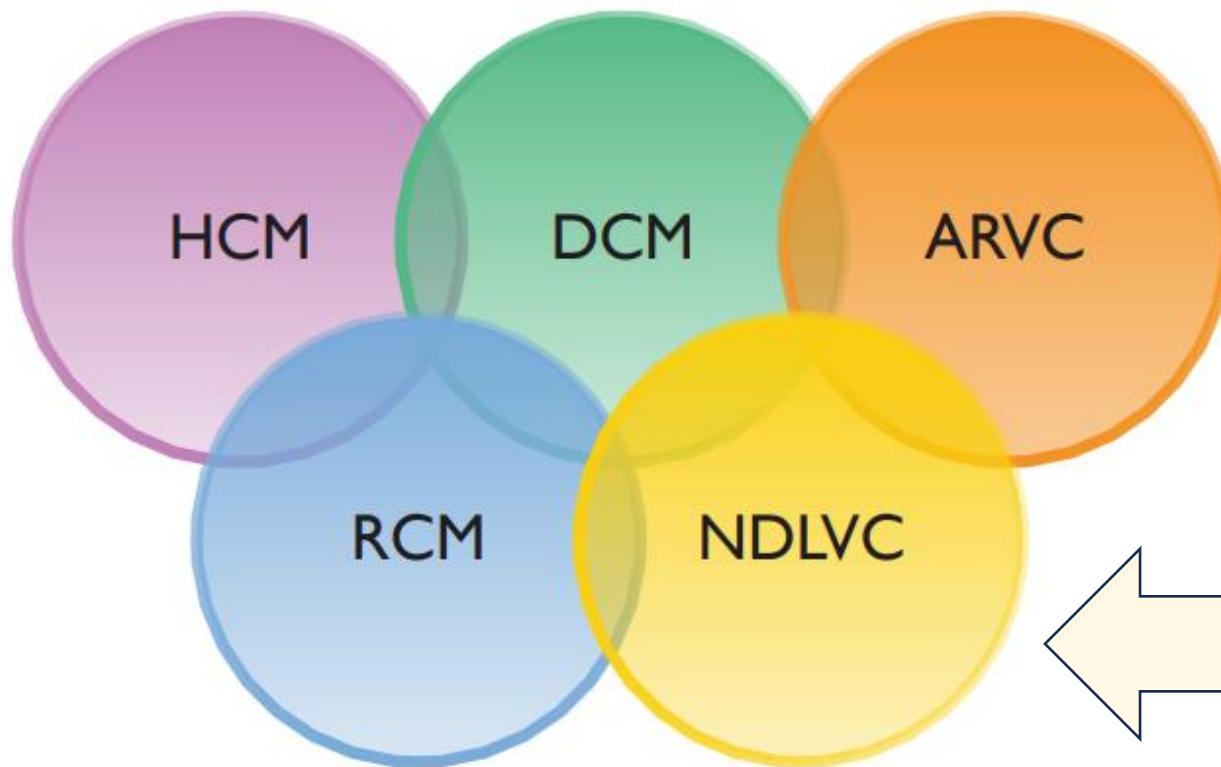
Non-ischaemic ventricular scar and other myocardial tissue characterization features on cardiac magnetic resonance

Functional traits

Ventricular systolic dysfunction (global, regional)

Ventricular diastolic dysfunction (restrictive physiology)

5 nhóm kiểu hình bệnh cơ tim



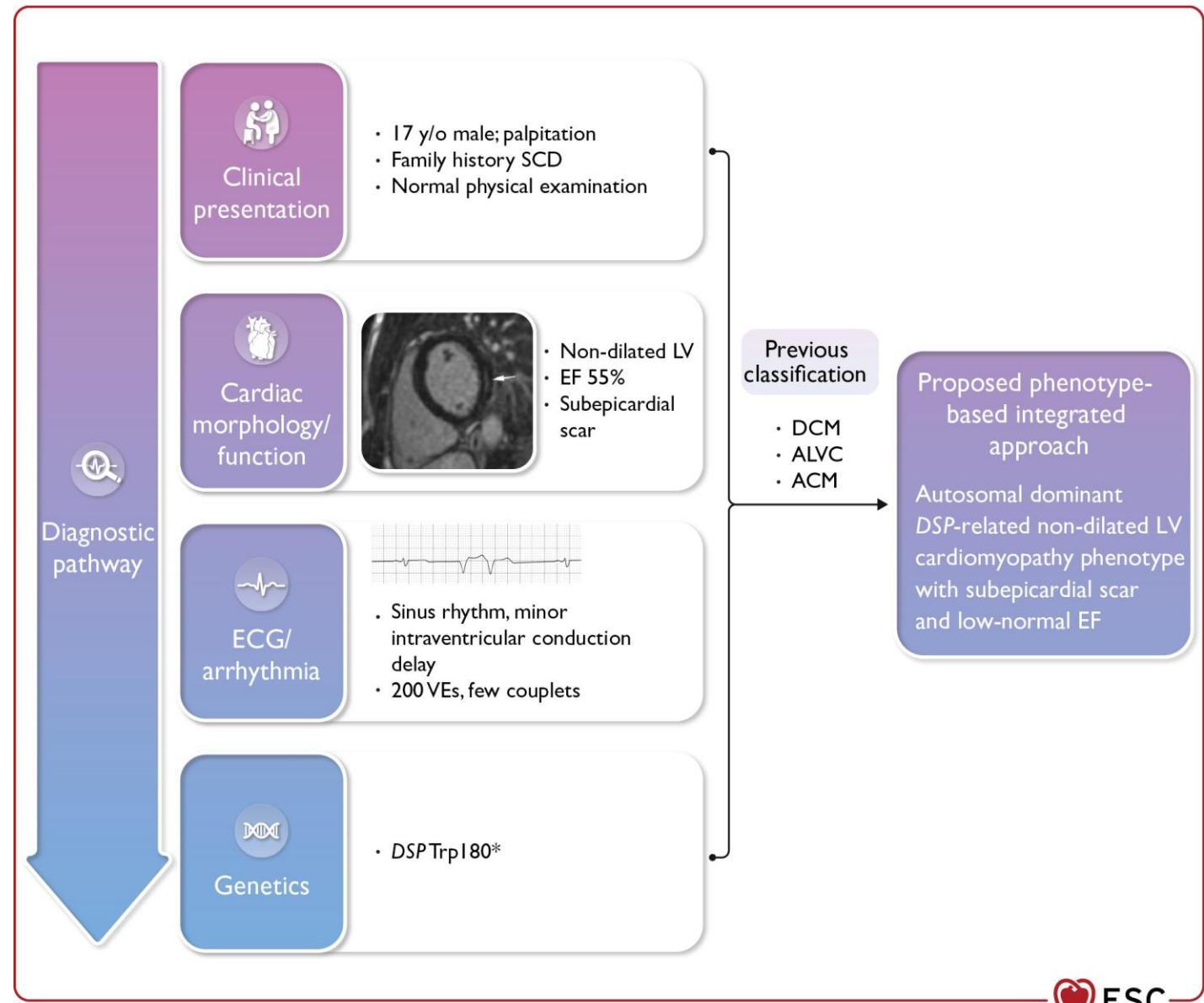
Non-dilated left ventricular cardiomyopathy:
Bệnh cơ tim có thất trái không giãn

Bệnh cơ tim có thất trái không giãn (NDLVC)

- Có tổn thương sẹo cơ tim hoặc cơ tim bị thay thế bởi tổ chức mỡ nhưng không giãn buồng thất trái
- Bao gồm những thể trước đây:
 - BCT giãn nhưng không giãn thất trái
 - BCT thất trái do rối loạn nhịp / BCT do RL nhịp có tổn thương chủ yếu bên thất trái / BCT giãn có rối loạn nhịp nhưng không đủ tiêu chuẩn chẩn đoán BCT do RL nhịp

Figure 4

Worked example of the non-dilated left ventricular cardiomyopathy phenotype



Những bệnh cảnh không phân loại trong BCT:

- BCT xốp:
 - Thiếu bằng chứng mô, phôi thai học về tình trạng “kết đặc” (compaction) cơ tim ở người
 - Nhóm bệnh BCT giãn/ phì đại/ RL chức năng thất trái
 - Khái niệm “phì đại cột cơ” (đặc biệt ở vận động viên/ PNCT)
- Hội chứng Takotsubo:
 - Mang tính chất thoáng qua, có hồi phục

2. Cách tiếp cận hệ thống:

chẩn đoán, quản lý theo quá trình bệnh

Đánh giá có hệ thống

Mục đích:

- Chẩn đoán, phân loại nhóm BCT
- Xác định bệnh nguyên: theo “cardiomyopathy mindset”

→ Chẩn đoán sớm, chính xác
→ Điều trị sớm

Recommendation Table 2 — Recommendations for diagnostic work-up in cardiomyopathies

Recommendations	Class ^a	Level ^b
<p>It is recommended that all patients with suspected or established cardiomyopathy undergo systematic evaluation using a multiparametric approach that includes clinical evaluation, pedigree analysis, ECG, Holter monitoring, laboratory tests, and multimodality imaging.⁶³</p>	I	C
<p>It is recommended that all patients with suspected cardiomyopathy undergo evaluation of family history and that a three- to four-generation family tree is created to aid in diagnosis, provide clues to underlying aetiology, determine inheritance pattern, and identify at-risk relatives.^{64–66}</p>	I	C

Figure 2

Clinical diagnostic workflow of cardiomyopathy

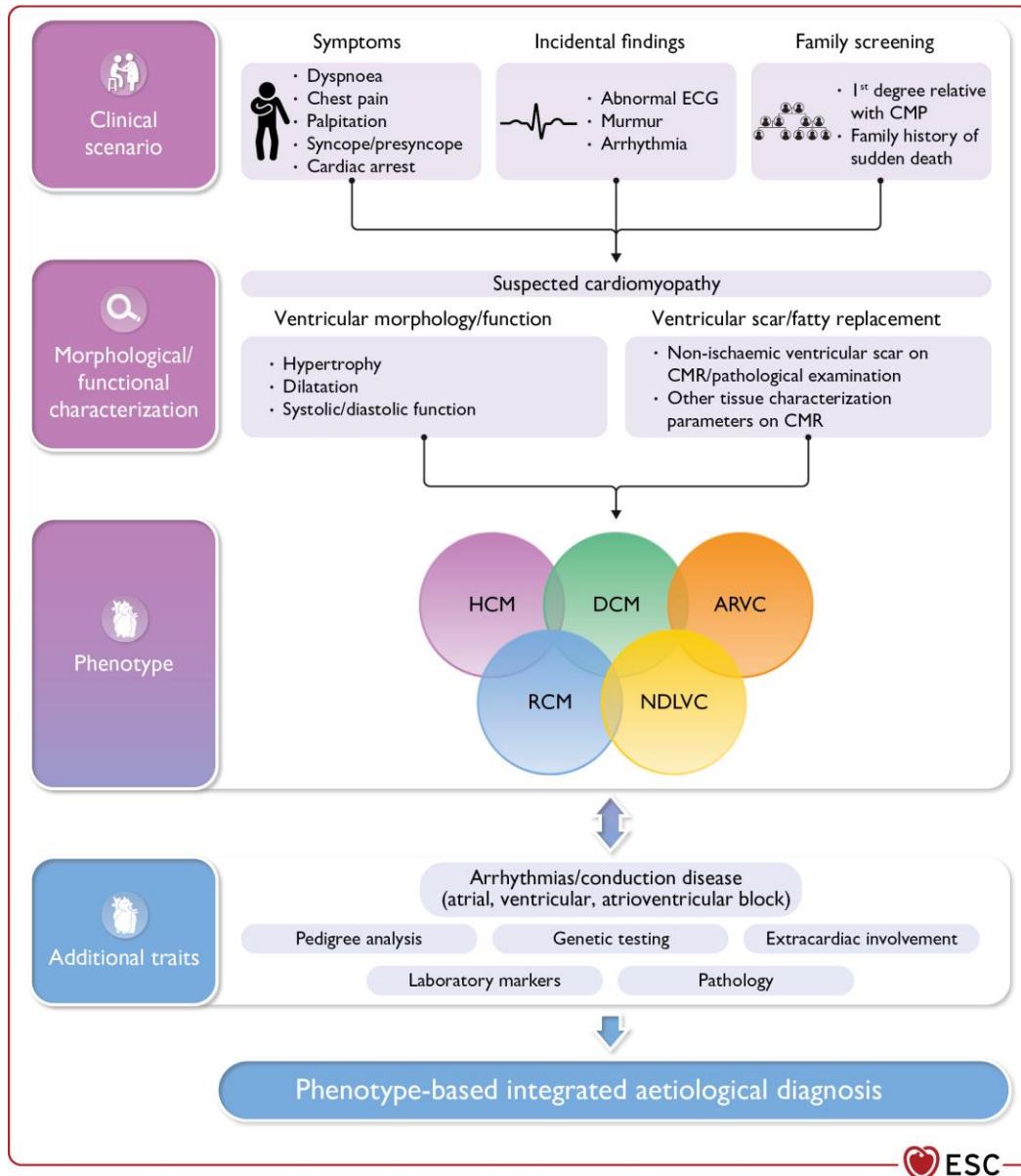


Figure 6
Multimodality imaging process in cardiomyopathies

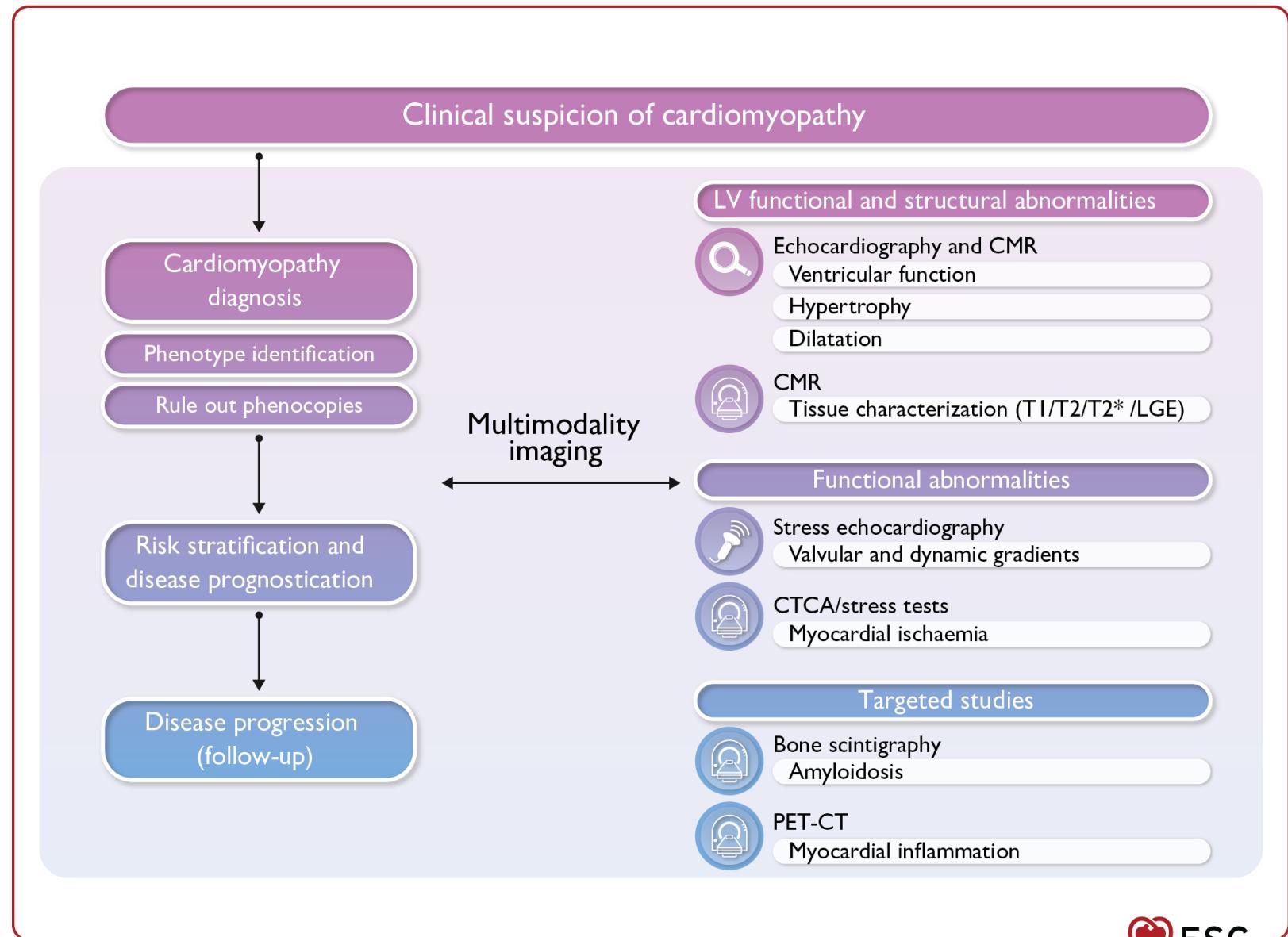


Figure 7

Examples of cardiac magnetic resonance imaging tissue characterization features that should raise the suspicion of specific aetiologies, grouped according to cardiomyopathy phenotype

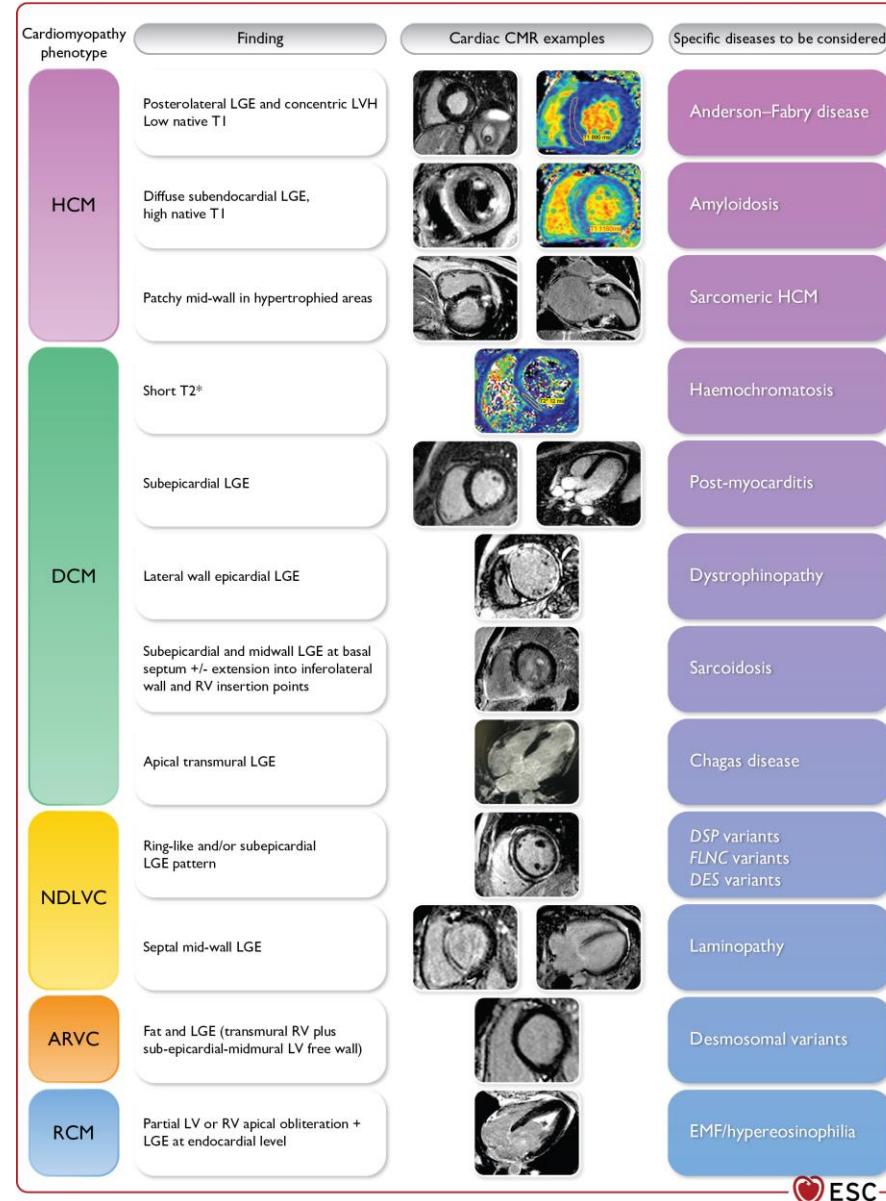
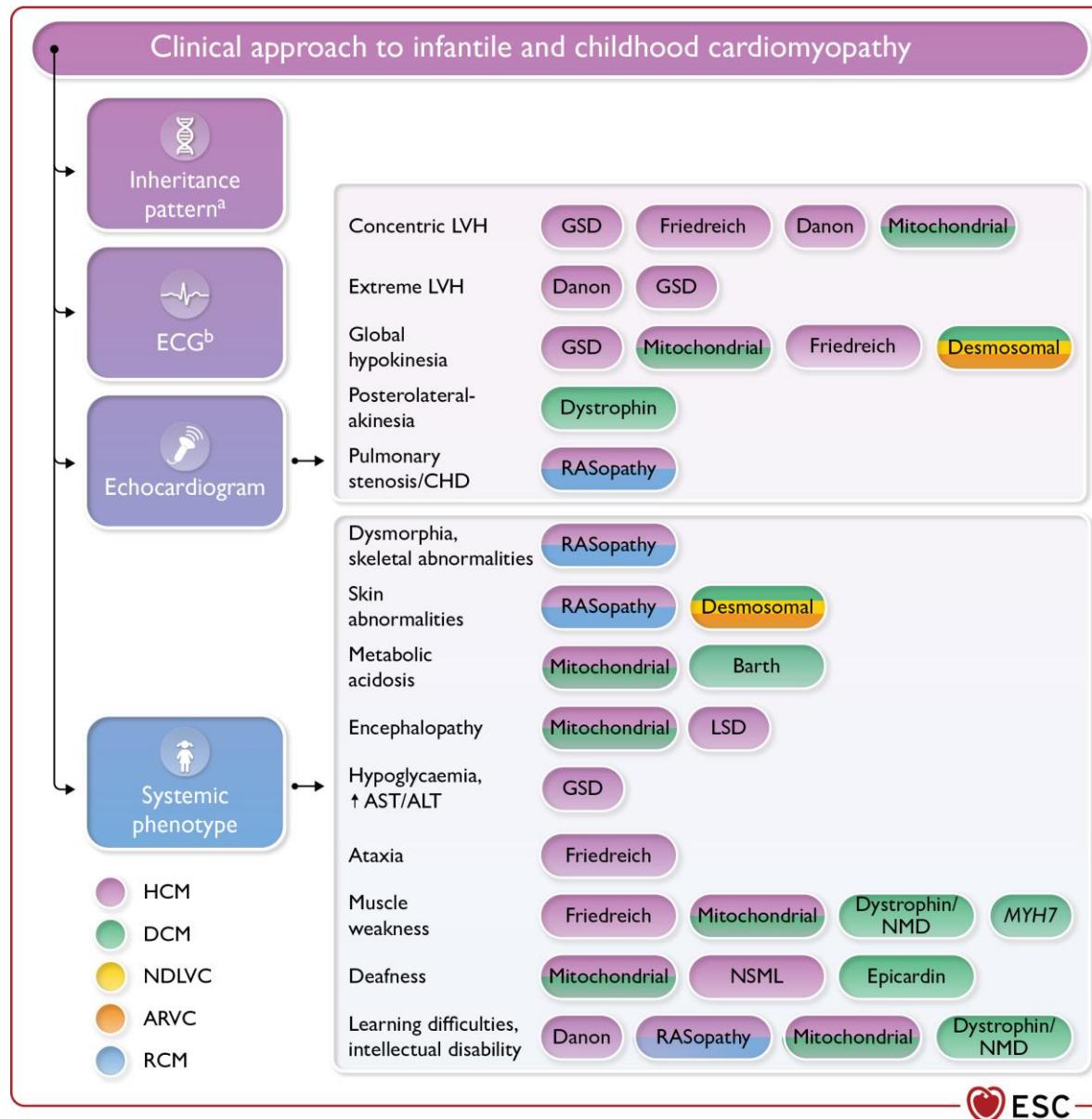


Figure 10

Clinical approach to infantile and childhood cardiomyopathy



Nhìn nhận BCT trong bối cảnh bệnh toàn thể

Examples of signs and symptoms that should raise the suspicion of specific aetiologies, grouped according to cardiomyopathy phenotype (1)

Finding	Cardiomyopathy phenotype				
	HCM	DCM	NDLVC	ARVC	RCM
Learning difficulties, developmental delay	Mitochondrial diseases	Dystrophinopathies	-	-	Noonan syndrome
	Noonan syndrome	Mitochondrial diseases	-	-	-
	Danon disease	Myotonic dystrophy	-	-	-
	-	<i>FKTN</i> variants	-	-	-
Sensorineural deafness	Mitochondrial diseases	Epicardin variants	-	-	-
	NSML	Mitochondrial diseases	-	-	-

Examples of signs and symptoms that should raise the suspicion of specific aetiologies, grouped according to cardiomyopathy phenotype (2)

Finding	Cardiomyopathy phenotype				
	HCM	DCM	NDLVC	ARVC	RCM
Visual impairment	Mitochondrial diseases	<i>CRYAB</i>	-	-	-
	ATTRv or hereditary ATTR	Type 2 myotonic dystrophy	-	-	-
	Danon disease	-	-	-	-
	Anderson–Fabry disease	-	-	-	-
Gait disturbance	Friedreich ataxia	Dystro-phinopathies	Myofibrillar myopathies	-	-
	-	Sarco-glycanopathies	-	-	-
	-	Myofibrillar myopathies	-	-	-

Examples of signs and symptoms that should raise the suspicion of specific aetiologies, grouped according to cardiomyopathy phenotype (3)



Finding	Cardiomyopathy phenotype				
	HCM	DCM	NDLVC	ARVC	RCM
Myotonia	-	Myotonic dystrophy	-	-	-
Paraesthesia/ sensory abnormalities/ neuropathic pain	Amyloidosis	-	-	-	Amyloidosis
	Anderson–Fabry disease	-	-	-	-
Carpal tunnel syndrome	TTR-related amyloidosis	-	-	-	-

Examples of signs and symptoms that should raise the suspicion of specific aetiologies, grouped according to cardiomyopathy phenotype (4)

Finding	Cardiomyopathy phenotype				
	HCM	DCM	NDLVC	ARVC	RCM
Muscle weakness	Mitochondrial diseases	Dystrophinopathies	Laminopathies	-	Desminopathies
	Glycogenoses	Sarco-glycanopathies	Desminopathies	-	-
	<i>FHL1</i> variants	Laminopathies	-	-	-
	-	Myotonic dystrophy	-	-	-
	-	Desminopathies	-	-	-
Palpebral ptosis	Mitochondrial diseases	Mitochondrial diseases	-	-	-
		Myotonic dystrophy	-	-	-
Lentigines	NSML	-	-	-	-

Examples of signs and symptoms that should raise the suspicion of specific aetiologies, grouped according to cardiomyopathy phenotype (5)



Finding	Cardiomyopathy phenotype				
	HCM	DCM	NDLVC	ARVC	RCM
Angiokeratomata	Anderson–Fabry disease	-	-	-	-
Pigmentation of skin and scars	-	Haemo-chromatosis	-	-	-
Palmoplantar keratoderma and woolly hair	-	Carvajal syndrome	-	Naxos and Carvajal syndromes	-
	-	DSP variants	DSP variants	DSP variants	-

BCT + gợi ý trên điện tim → tìm hội chứng bao trùm

Examples of electrocardiographic features that should raise the suspicion of specific aetiologies, grouped according to cardiomyopathy phenotype (1)



Cardiomyopathy phenotype	Finding	Specific diseases to be considered
HCM	Short PR interval/pre-excitation	Glycogenosis Danon disease <i>PRKAG2</i> cardiomyopathy Anderson–Fabry disease Mitochondrial disease
	AV block	Amyloidosis Anderson–Fabry disease (late stage) Danon disease Sarcoidosis <i>PRKAG2</i> cardiomyopathy
	Extreme LVH	Danon disease Glycogenosis (e.g. Pompe disease) <i>PRKAG2</i> cardiomyopathy

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Examples of electrocardiographic features that should raise the suspicion of specific aetiologies, grouped according to cardiomyopathy phenotype (2)



Cardiomyopathy phenotype	Finding	Specific diseases to be considered
HCM (continued)	Low QRS voltage	Amyloidosis Friedreich ataxia
	Superior QRS axis ('northwest axis')	Noonan syndrome
	Q waves/pseudoinfarction pattern	Amyloidosis
DCM	AV block	Laminopathy Emery–Dreifuss 1 Myocarditis (esp. Chagas disease, Lyme disease, diphtheria) Sarcoidosis Desminopathy Myotonic dystrophy
	Low P wave amplitude	Emery–Dreifuss 1 and 2
	Atrial standstill	Emery–Dreifuss 1 and 2

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Examples of electrocardiographic features that should raise the suspicion of specific aetiologies, grouped according to cardiomyopathy phenotype (3)



Cardiomyopathy phenotype	Finding	Specific diseases to be considered
DCM (continued)	Posterolateral infarction pattern	Dystrophinopathy Limb-girdle muscular dystrophy Sarcoidosis
	Extremely low QRS amplitude	<i>PLN</i> variant
NDLVC	AV block	Laminopathy Desminopathy
	Extremely low QRS amplitude	<i>PLN</i> variant
	Low QRS voltage + atypical RBBB	Desmosomal variants
ARVC	T wave inversion V1-V3 + terminal activation delay +/- low right ventricular voltages +/- atypical RBBB	-
RCM	AV block	Desminopathy Amyloidosis

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Những thăm dò/ xét nghiệm cơ bản (first: I-C) và chuyên sâu (second: IIa-C) với từng nhóm BCT

First-level (to be performed in each patient) and second-level (to be performed in selected patients following specialist evaluation to identify specific aetiologies) laboratory tests, grouped by cardiomyopathy phenotype (1)



Level	HCM	DCM	NDLVC	ARVC	RCM
First	CK	Calcium	Calcium	C-reactive protein	CK
	Liver function	CK	CK	Liver function	Ferritin
	NT-proBNP	Ferritin	C-reactive protein	NT-proBNP	Full blood count
	Proteinuria	Full blood count	Full blood count	Renal function	Liver function
	Renal function	Liver function	Liver function	Troponin	NT-proBNP
	Troponin	NT-proBNP	NT-proBNP		Proteinuria
		Phosphate	Phosphate		Renal function
		Proteinuria	Proteinuria		Serum angiotensin-converting enzyme
		Renal function	Renal function		Serum iron
		Serum iron	Troponin		Troponin
		Thyroid function			Urine and plasma protein
		Troponin			immunofixation, free light chains
		Vitamin D (children)			

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First-level (to be performed in each patient) and second-level (to be performed in selected patients following specialist evaluation to identify specific aetiologies) laboratory tests, grouped by cardiomyopathy phenotype (2)



Level	HCM	DCM	NDLVC	ARVC	RCM
Second	Alpha-galactosidase A levels (males) and lyso-Gb3	Carnitine profile Free fatty acids Lactic acid	Organ- and non-organ-specific serum autoantibodies Viral serology	-	Organ- and non-organ-specific autoantibodies Serum angiotensin-converting enzyme
	Carnitine profile	Organ- and non-organ-specific serum			
	Free fatty acids	Organ- and non-organ-specific serum			
	Immunofixation and free light chains	autoantibodies			
	Lactic acid	Serum angiotensin-converting enzyme			
	Myoglobinuria	Thiamine			
	Pyruvate	Viral serology			
	PTH	Urine organic acids and plasma amino acids			
	Urine and plasma protein				
	Urine organic acids and plasma amino acids				

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Chỉ định của các kỹ thuật CĐHA

Recommendations for cardiac magnetic resonance indication in patients with cardiomyopathy



Recommendations	Class	Level
Contrast-enhanced CMR is recommended in patients with cardiomyopathy at initial evaluation.	I	B
Contrast-enhanced CMR should be considered in patients with cardiomyopathy during follow-up to monitor disease progression and aid risk stratification and management.	IIa	C
Contrast-enhanced CMR should be considered for the serial follow-up and assessment of therapeutic response in patients with cardiac amyloidosis, Anderson–Fabry disease, sarcoidosis, inflammatory cardiomyopathies, and haemochromatosis with cardiac involvement.	IIa	C
In families with cardiomyopathy in which a disease-causing variant has been identified, contrast-enhanced CMR should be considered in genotype-positive/phenotype-negative family members to aid diagnosis and detect early disease.	IIa	B
In cases of familial cardiomyopathy without a genetic diagnosis, contrast-enhanced CMR may be considered in phenotype-negative family members to aid diagnosis and detect early disease.	IIIb	C

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Recommendations for computed tomography and nuclear imaging



Recommendations	Class	Level
DPD/PYP/HMDP bone-tracer scintigraphy is recommended in patients with suspected ATTR-related cardiac amyloidosis to aid diagnosis	I	B
Contrast-enhanced cardiac CT should be considered in patients with suspected cardiomyopathy who have inadequate echocardiographic imaging and contraindications to CMR.	IIa	C
In patients with suspected cardiomyopathy, CT-based imaging should be considered to exclude congenital or acquired coronary artery disease as a cause of the observed myocardial abnormality.	IIa	C
18F-FDG-PET scanning should be considered for the diagnostic work-up in patients with cardiomyopathy in whom cardiac sarcoidosis is suspected.	IIa	C

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Recommendation for endomyocardial biopsy in patients with cardiomyopathy



Recommendation	Class	Level
In patients with suspected cardiomyopathy, EMB should be considered to aid in diagnosis and management when the results of other clinical investigations suggest myocardial inflammation, infiltration, or storage that cannot be identified by other means.	IIa	C

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Hệ thống lại ý nghĩa của những dấu hiệu trong thăm dò/CĐHA

Frequently encountered actionable results on multimodality imaging



Parameter/Finding	Action
RWMAs on echocardiography or CMR	Raise suspicion of concomitant CAD, myocarditis, ARVC, NDLVC, or sarcoidosis
Systolic impairment on echocardiography or CMR	Assessment of risk in DCM, NDLVC, and ARVC; evaluation of treatment efficacy
Measurement of the wall thickness on echocardiography or CMR	Diagnosis of HCM (when echocardiography is inconclusive); risk stratification in HCM
Diastolic dysfunction on echocardiography	Explain symptoms; evaluation of treatment efficacy
Left atrial size on echocardiography	SCD risk prediction in HCM; systematic screening for AF in case of left atrial enlargement
LVOTO in HCM on resting/exercise echocardiography	Explain symptoms; guide management
Non-invasive evaluation of pulmonary pressures	Explain symptoms; guide management
Tissue characterization on CMR	Diagnosis; risk assessment
Inflammation on CMR or 18F-FDG-PET	Diagnosis; evaluation of treatment efficacy in inflammatory cardiomyopathies

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Figure 1

Central illustration. Key aspects in the evaluation and management of cardiomyopathies

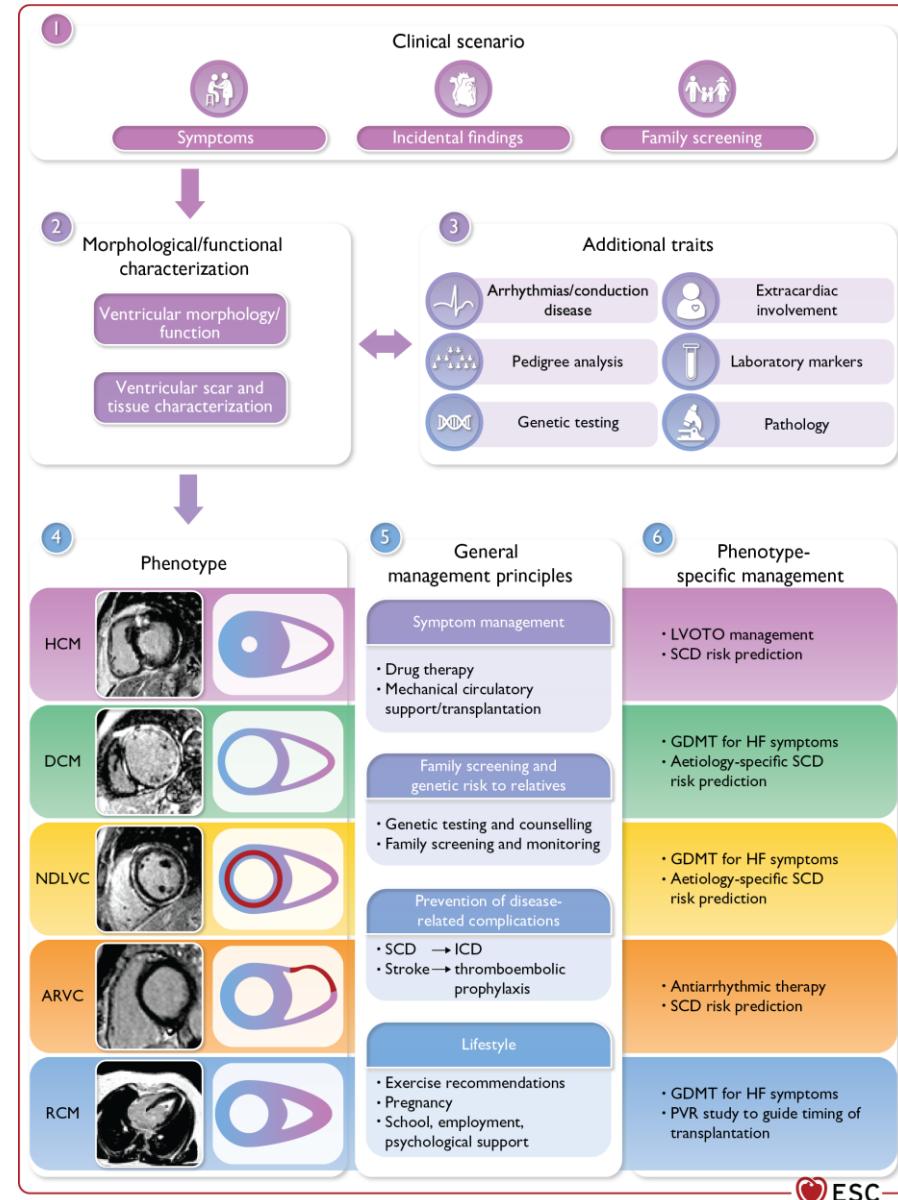
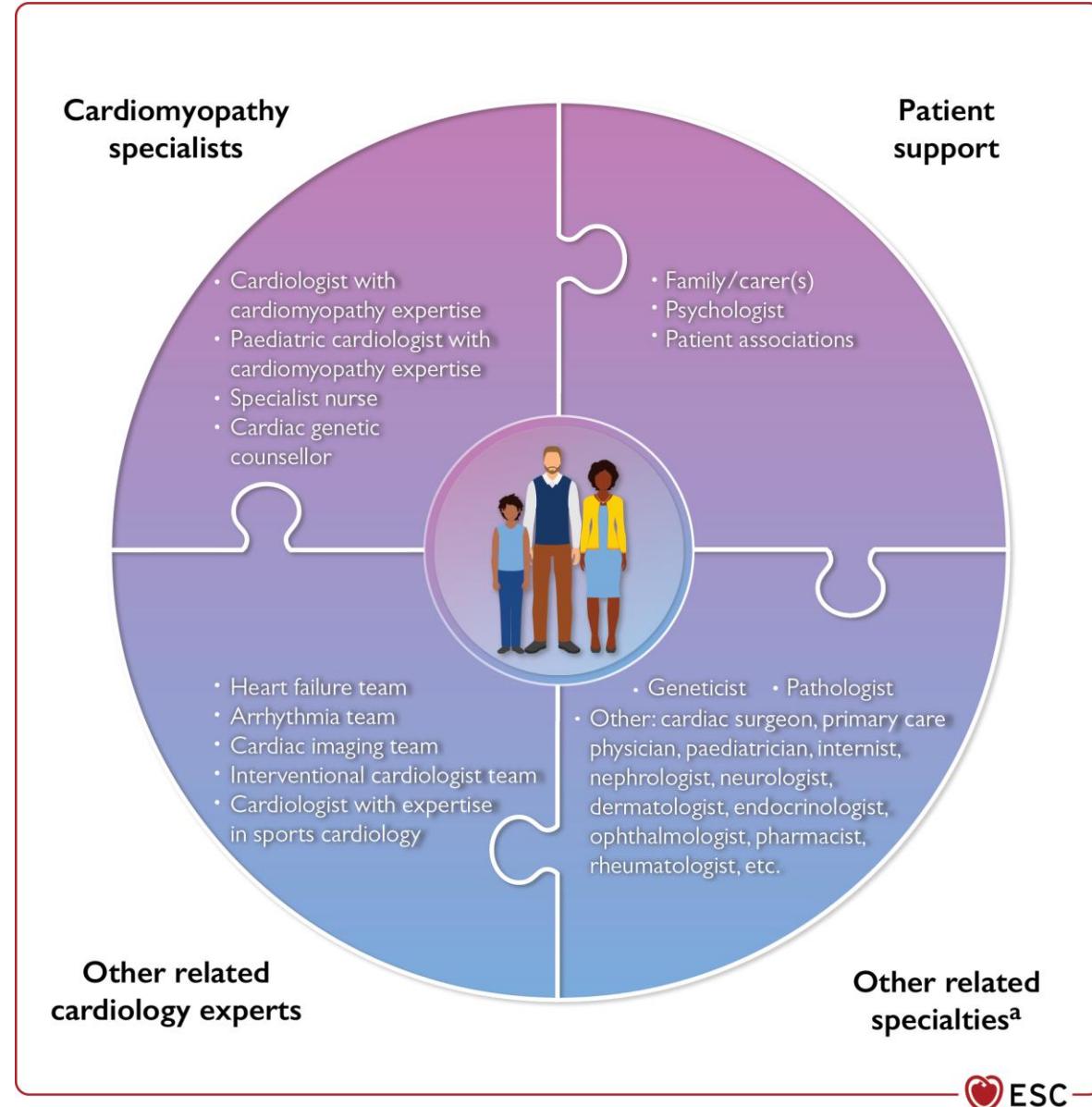


Figure 5
**Multidisciplinary care
of cardiomyopathies**



3. Gen di truyền và tư vấn

Hệ thống lại các đột biến liên quan và cơ chế
di truyền trong các nhóm BCT

Examples of inheritance patterns that should raise the suspicion of specific genetic aetiologies, grouped according to cardiomyopathy phenotype (1)



Cardiomyopathy phenotype		AD	AR	X-linked	Matrilineal
HCM	Sarcomeric	X			
	Anderson–Fabry			X	
	Danon			X	
	TTR amyloidosis	X			
	RASopathy	X	(X)		
	Friedreich ataxia		X		
	Mitochondrial				
	Mitochondrial DNA				X
	Nuclear DNA	X	X	X	

Examples of inheritance patterns that should raise the suspicion of specific genetic aetiologies, grouped according to cardiomyopathy phenotype (2)

Cardiomyopathy phenotype	AD	AR	X-linked	Matrilineal
DCM	<i>LMNA</i>	X		
	<i>RBM20</i>	X		
	Sarcomeric	X		
	Dystrophin		X	
	Emerin		X	
	Barth syndrome		X	
	Mitochondrial			
	Mitochondrial DNA			X
	Nuclear DNA	X	X	X

Examples of inheritance patterns that should raise the suspicion of specific genetic aetiologies, grouped according to cardiomyopathy phenotype (3)

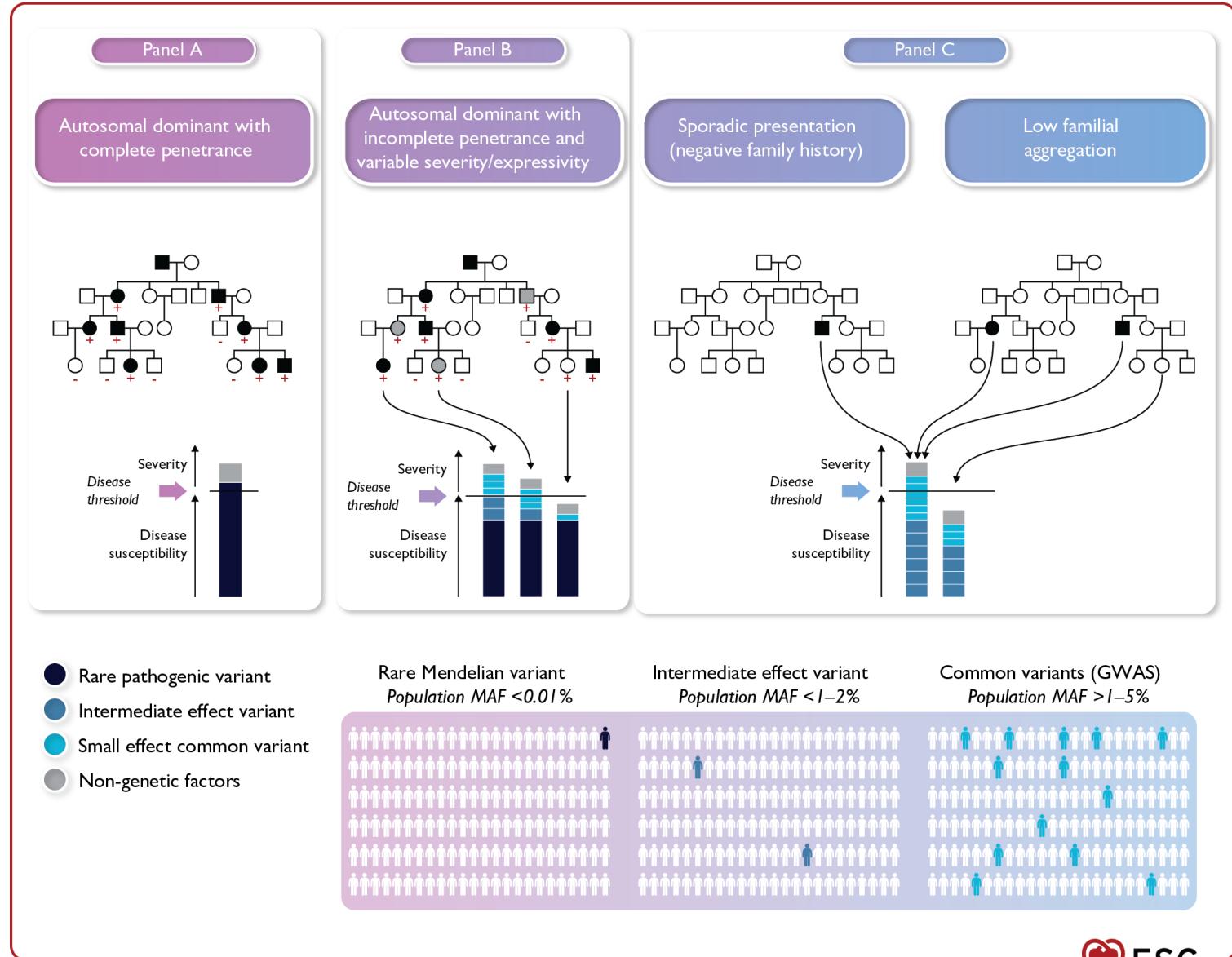


Cardiomyopathy phenotype	AD	AR	X-linked	Matrilineal
NDLVC	<i>LMNA</i>	X		
	<i>DES</i>	X	X	
	<i>FLNC</i>	X		
	<i>PLN</i>	X		
	<i>TMEM43</i>	X		
	<i>RBM20</i>	X		
ARVC	<i>PLN</i>	X		
	Desmosomal	X	X	
	<i>TMEM43</i>	X		
RCM	Sarcomeric	X		
	<i>DES</i>	X	X	
	<i>FLNC</i>	X		
	<i>BAG3</i>	X		
	RASopathy	X	(X)	

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Figure 8

The genetic architecture of the cardiomyopathies



Hay kiểu hình BCT tương ứng với từng đột
biến gen cụ thể

Overview of genes associated with monogenic, non-syndromic cardiomyopathies, and ESC their relative contributions to different cardiomyopathic phenotypes (1)

Gene	Cardiomyopathy phenotype					Associated phenotype
	HCM	DCM	NDLVC	ARVC	RCM	
<i>ABCC9</i>	●	○				Cantu syndrome
<i>ACTA1</i>	○					
<i>ACTC1</i>	●	○	○	○	○	
<i>ACTN2</i>	○	○	○			
<i>ALPK3</i>	●					
<i>ANKRD1</i>	○	○				
<i>BAG3</i>	●	● ●		○		Myofibrillar myopathy
<i>CACNA1C</i>	●					Timothy syndrome
<i>CACNB2</i>	○					
<i>CALR3</i>	○					
<i>CASQ2</i>	○					
<i>CAV3</i>	●					Caveolinopathy
<i>CDH2</i>				○		

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Overview of genes associated with monogenic, non-syndromic cardiomyopathies, and ESC their relative contributions to different cardiomyopathic phenotypes (2)

Gene	Cardiomyopathy phenotype					Associated phenotype
	HCM	DCM	NDLVC	ARVC	RCM	
<i>COX15</i>	●					Leigh syndrome
<i>CRYAB</i>	●					Alpha-B crystallinopathy
<i>CSRP3</i>	●	○				
<i>CTF1</i>		○				
<i>CTNNA3</i>				○		
<i>DES</i>	●	●	●	●	●	Desminopathy
<i>DMD</i>		●	●			X-linked progressive MD
<i>DMPK</i>			●			
<i>DSC2</i>				● ●		
<i>DSG2</i>		○		● ●		
<i>DSP</i>	○	● ●	●	●		
<i>DTNA</i>	○		●			
<i>EYA4</i>	○					

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Overview of genes associated with monogenic, non-syndromic cardiomyopathies, and ESC their relative contributions to different cardiomyopathic phenotypes (3)

Gene	Cardiomyopathy phenotype					Associated phenotype
	HCM	DCM	NDLVC	ARVC	RCM	
<i>FHL1</i>	●					Emery–Dreifuss MD
<i>FLNC</i>	●	● ●	●	●	●	Myofibrillar myopathy
<i>FHOD3</i>	●					
<i>FXN</i>	●					Friedreich ataxia
<i>GAA</i>	●					Pompe disease
<i>GATA4</i>			●			
<i>GATAD1</i>		○				
<i>GLA</i>	●					Anderson–Fabry disease
<i>HCN4</i>			●			
<i>ILK</i>		○	●			
<i>JPH2</i>	●	●				
<i>JUP</i>				●		Naxos disease (cardiocutaneous syndrome)

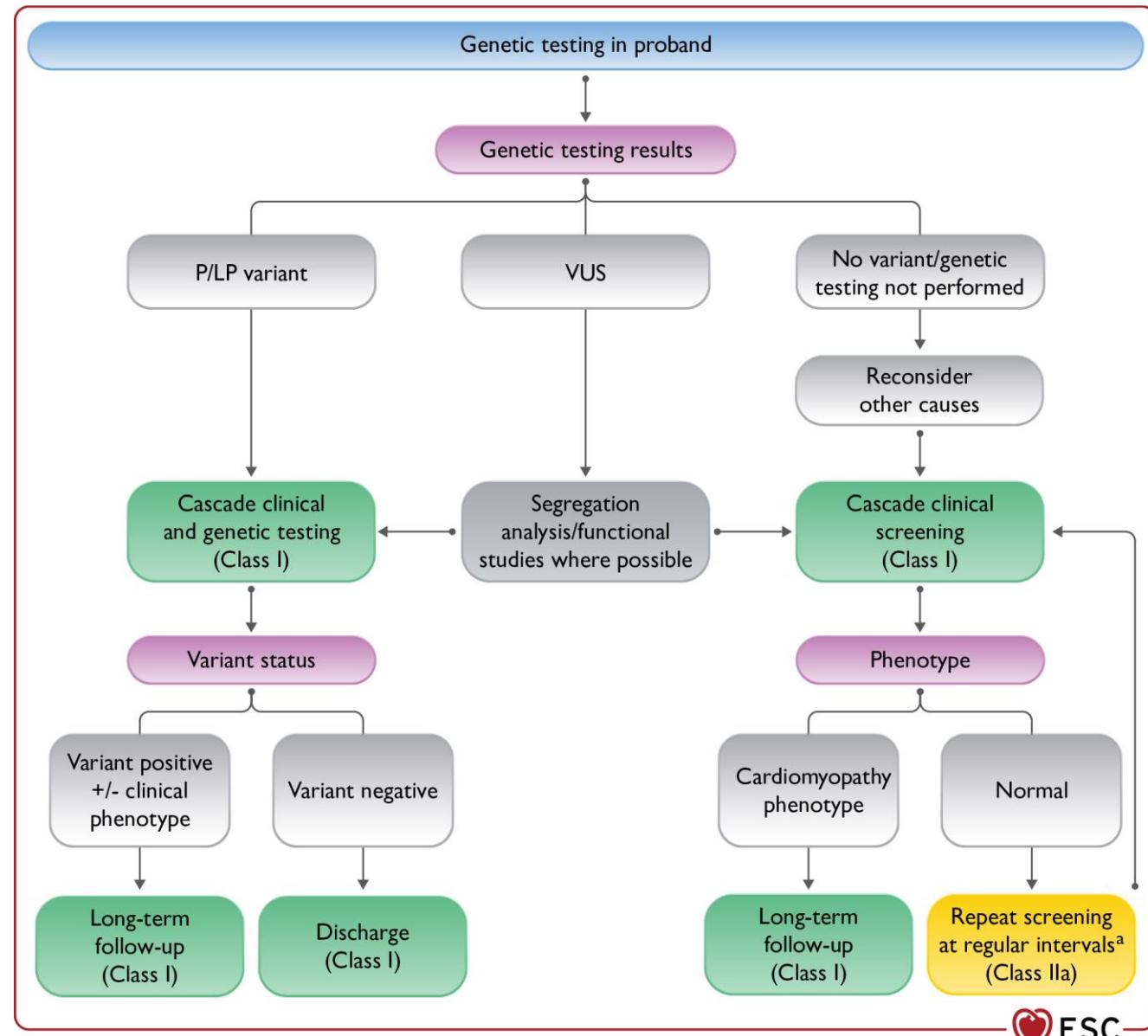
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Vai trò của xét nghiệm di truyền trong BCT

- Chẩn đoán: bệnh cảnh LS chưa rõ ràng (VD: hình ảnh gợi ý BCTPD ở người THA → phát hiện BCTPD thể sarcome)
- Tiên lượng: VD: BCT giãn có đột biến LMNA → cân nhắc chỉ định ICD
- Điều trị: trong Amyloid
- Tư vấn trước sinh: trường hợp hỗ trợ sinh sản
- Sàng lọc trong gia đình

Figure 11

Algorithm for the approach to family screening and follow-up of family members



4. Cập nhật khuyến cáo về đột tử ở BN BCT

Hệ thống những trường hợp BCT có nguy cơ đột tử
cao, các gen nguy cơ cao và chỉ định ICD

Additional recommendations for prevention of sudden cardiac death in patients with hypertrophic cardiomyopathy (1)

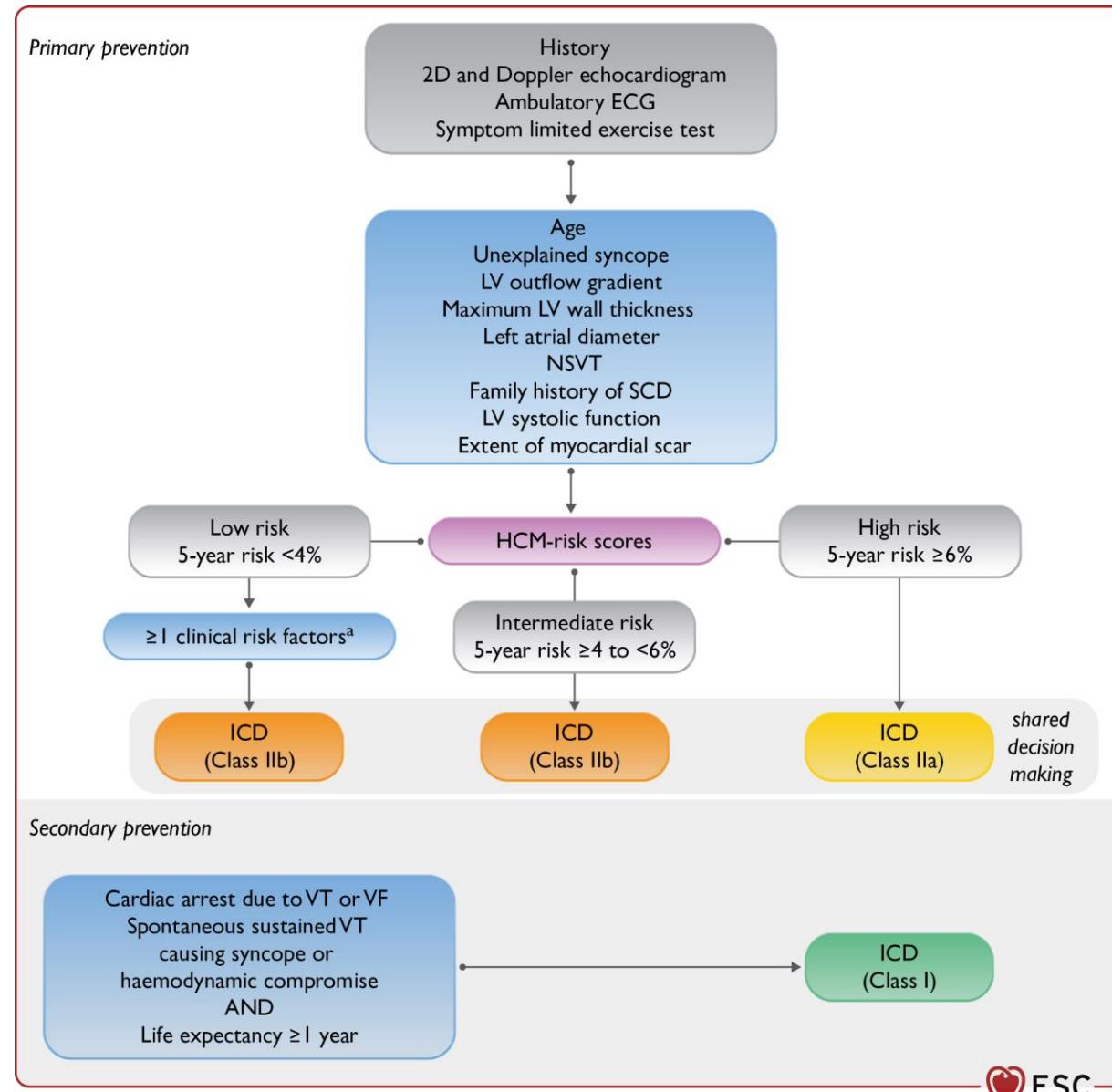


Recommendations	Class	Level
<i>Secondary prevention</i>		
Implantation of an ICD is recommended in patients who have survived a cardiac arrest due to VT or VF, or who have spontaneous sustained VT with haemodynamic compromise.	I	B
<i>Primary prevention</i>		
The HCM Risk-SCD calculator is recommended as a method of estimating risk of sudden death at 5 years in patients aged ≥ 16 years for primary prevention.	I	B
Validated paediatric-specific risk-prediction models (e.g. HCM Risk-Kids) are recommended as a method of estimating risk of sudden death at 5 years in patients aged <16 years for primary prevention.	I	B
It is recommended that the 5-year risk of SCD be assessed at first evaluation and re-evaluated at 1–2 year intervals or whenever there is a change in clinical status.	I	B

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Figure 16

Flow chart for implantation of an implantable cardioverter defibrillator in patients with hypertrophic cardiomyopathy



High-risk genotypes and associated predictors of sudden cardiac death (1) ESC

Gene	Annual SCD rate	Predictors of SCD
<i>LMNA</i>	5–10%	Estimated 5-year risk of life-threatening arrhythmia using LMNA risk score https://lmna-risk-vta.fr
<i>FLNC-truncating variants</i>	5–10%	LGE on CMR LVEF<45%
<i>TMEM43</i>	5–10%	Male Female and any of the following: LVEF <45%, NSVT, LGE on CMR, >200 VE on 24h Holter ECG

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High-risk genotypes and associated predictors of sudden cardiac death (2) ESC

Gene	Annual SCD rate	Predictors of SCD
PLN	3–5%	Estimated 5-year risk of life-threatening arrhythmia using <i>PLN</i> risk score https://plnriskcalculator.shinyapps.io/final_shiny LVEF<45% LGE on CMR NSVT
DSP	3–5%	LGE on CMR LVEF<45%
RBM20	3–5%	LGE on CMR LVEF<45%

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Recommendations for an implantable cardioverter defibrillator in patients with dilated cardiomyopathy (1)

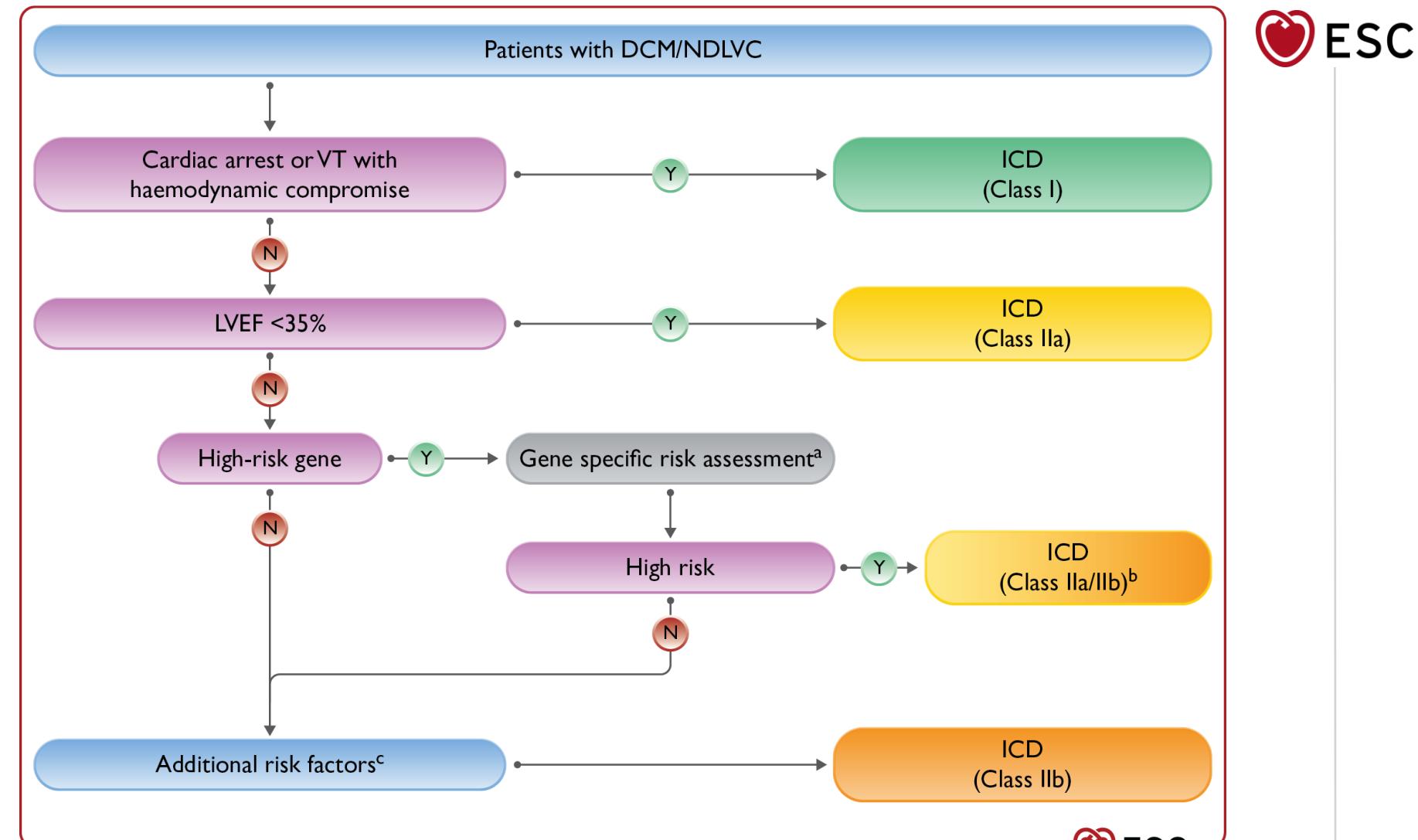


Recommendations	Class	Level
<i>Secondary prevention</i>		
An ICD is recommended to reduce the risk of sudden death and all-cause mortality in patients with DCM who have survived a cardiac arrest or have recovered from a ventricular arrhythmia causing haemodynamic instability.	I	B
<i>Primary prevention</i>		
An ICD should be considered to reduce the risk of sudden death and all-cause mortality in patients with DCM, symptomatic heart failure, and LVEF ≤35% despite >3 months of OMT.	IIa	A
The patient's genotype should be considered in the estimation of SCD risk in DCM.	IIa	B
An ICD should be considered in patients with DCM with a genotype associated with high SCD risk and LVEF >35% in the presence of additional risk factors.	IIa	C

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Figure 17

Implantation of implantable cardioverter defibrillators in patients with dilated cardiomyopathy or non-dilated left ventricular cardiomyopathy flowchart



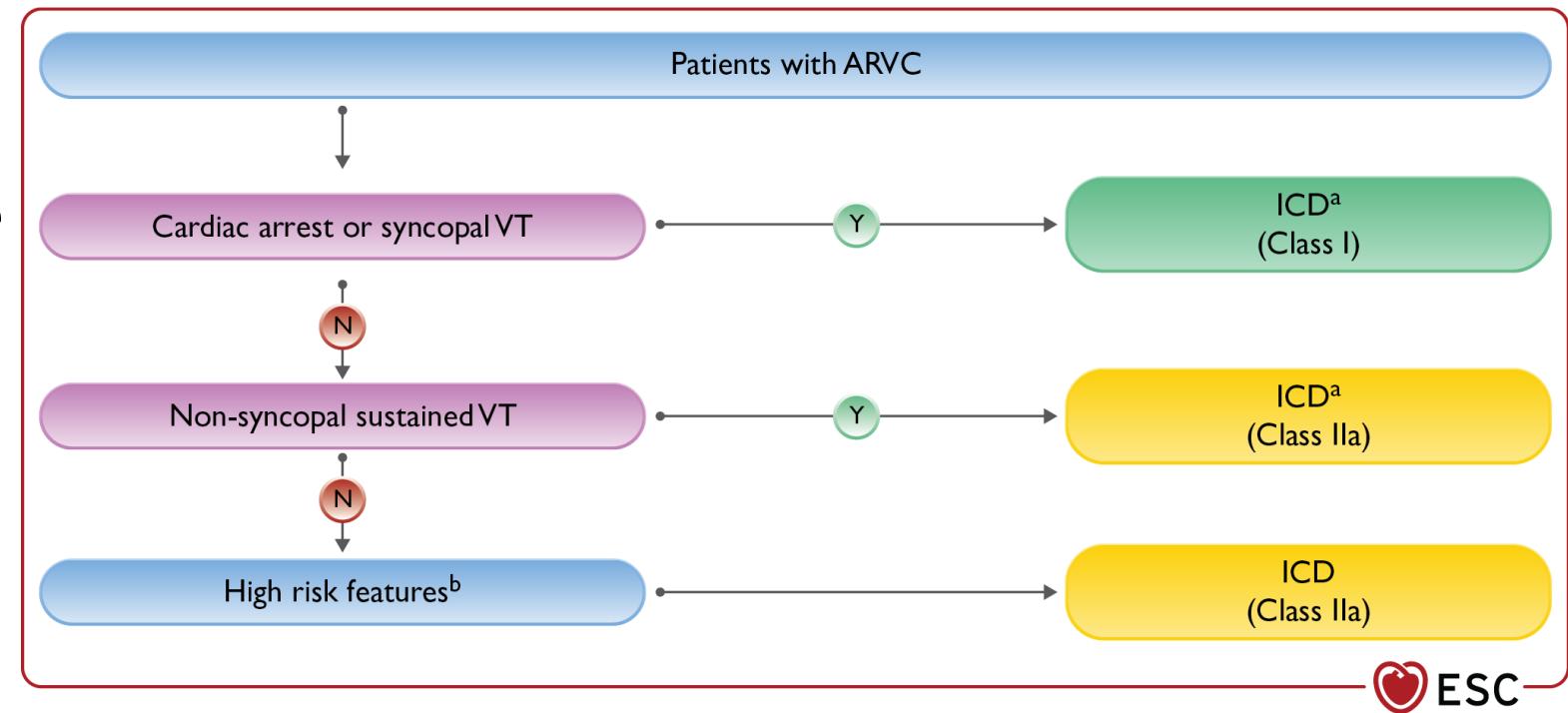
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Recommendations for an implantable cardioverter defibrillator in patients with non-dilated left ventricular cardiomyopathy (1)

Recommendations	Class	Level
<i>Secondary prevention</i>		
An ICD is recommended to reduce the risk of sudden death and all-cause mortality in patients with NDLVC who have survived a cardiac arrest or have recovered from a ventricular arrhythmia causing haemodynamic instability.	I	C
<i>Primary prevention</i>		
An ICD should be considered to reduce the risk of sudden death and all-cause mortality in patients with NDLVC, heart failure symptoms, and LVEF ≤35% despite >3 months of OMT.	IIa	A
The patient's genotype should be considered in the estimation of SCD risk in NDLVC.	IIa	C
An ICD should be considered in patients with NDLVC with a genotype associated with high SCD risk and LVEF >35% in the presence of additional risk factors.	IIa	C

Figure 18

Algorithm to approach implantable cardioverter defibrillator decision-making in patients with arrhythmogenic right ventricular cardiomyopathy



Recommendations for implantable cardioverter defibrillator in patients with cardiomyopathy (2)



Recommendations	Class	Level
<i>Secondary prevention</i>		
Implantation of an ICD is recommended:		
• in patients with HCM, DCM, and ARVC who have survived a cardiac arrest due to VT or VF, or who have spontaneous sustained ventricular arrhythmia causing syncope or haemodynamic compromise in the absence of reversible causes.	I	B
• in patients with NDLVC and RCM who have survived a cardiac arrest due to VT or VF, or who have spontaneous sustained ventricular arrhythmia causing syncope or haemodynamic compromise in the absence of reversible causes.	I	C
ICD implantation should be considered in patients with cardiomyopathy presenting with haemodynamically tolerated VT, in the absence of reversible causes.	IIa	C

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Recommendations for implantable cardioverter defibrillator in patients with cardiomyopathy (3)



Recommendations (continued)	Class	Level
<i>Primary prevention</i>		
Comprehensive SCD risk stratification is recommended in all cardiomyopathy patients who have not suffered a previous cardiac arrest/sustained ventricular arrhythmia at initial evaluation and at 1–2 year intervals, or whenever there is a change in clinical status.	I	C
The use of validated SCD algorithms/scores as aids to the shared decision-making when offering ICD implantation, where available:		
• is recommended in patients with HCM.	I	B
• should be considered in patients with DCM, NDLVC, and ARVC.	IIa	B
If a patient with cardiomyopathy requires pacemaker implantation, comprehensive SCD risk stratification to evaluate the need for ICD implantation should be considered.	IIa	C

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Recommendations for implantable cardioverter defibrillator in patients with cardiomyopathy (4)



Recommendations (continued)	Class	Level
<i>Choice of ICD</i>		
When an ICD is indicated, it is recommended to evaluate whether the patient could benefit from CRT.	I	A
Subcutaneous defibrillators should be considered as an alternative to transvenous defibrillators in patients with an indication for an ICD when pacing therapy for bradycardia, cardiac resynchronization, or antitachycardia pacing is not anticipated.	IIa	B
The wearable cardioverter defibrillator should be considered for adult patients with a secondary prevention ICD indication who are temporarily not candidates for ICD implantation.	IIa	C

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