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Novedades de las nuevas guías ESC de manejo de miocardiopatías (2023)

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2023 ESC Guidelines for the Management of Cardiomyopathies

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ESC
European Society of Cardiology

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2023 ESC Guidelines for the management of cardiomyopathies

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Guideline structure

- Phenotypic approach to cardiomyopathies
- Integrated patient management
- The patient pathway

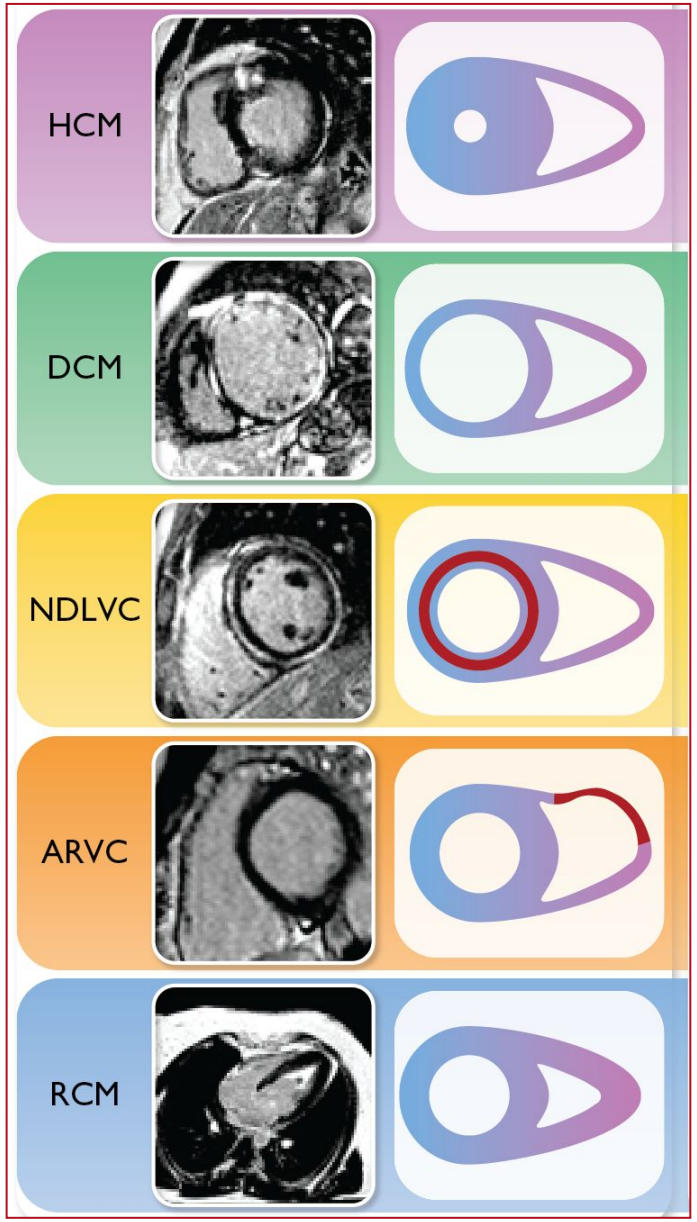
- Specific cardiomyopathy phenotypes

- Other recommendations: sports, reproductive issues, recommendations for non-cardiac surgery
- Requirements for specialized cardiomyopathy units
- Living with cardiomyopathy: advice for patients
- Sex differences in cardiomyopathies
- Comorbidities and cardiovascular risk factors in cardiomyopathies
- Coronavirus disease (COVID-19) and cardiomyopathies

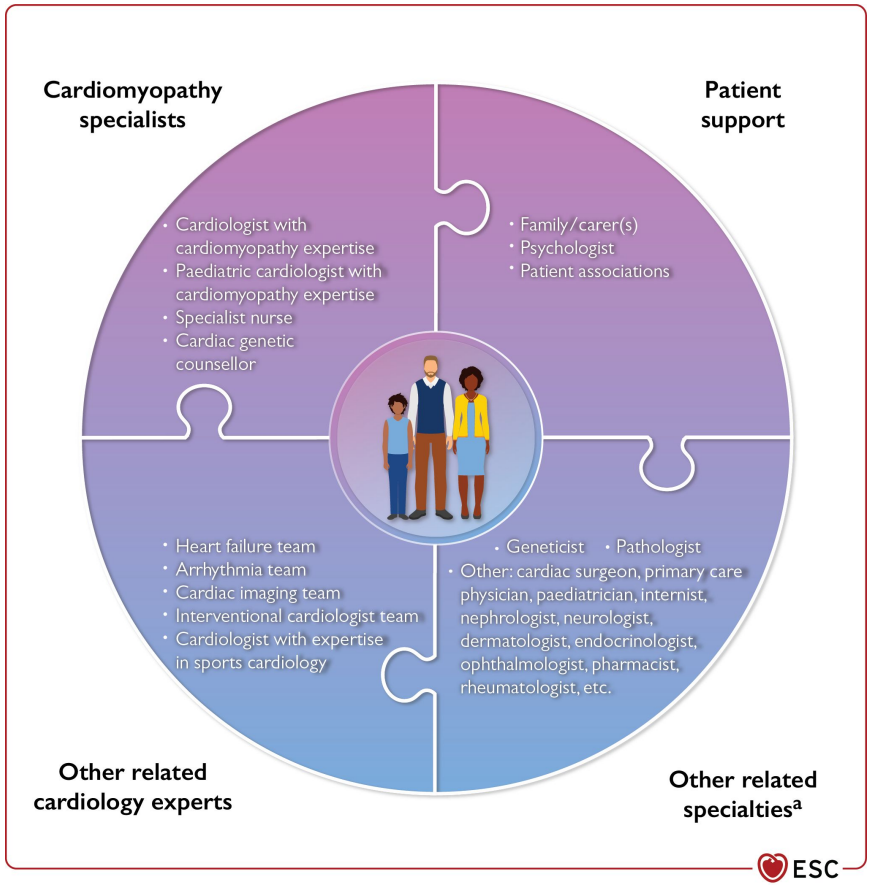
Phenotypic approach to cardiomyopathies

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)



Multidisciplinary care of cardiomyopathies



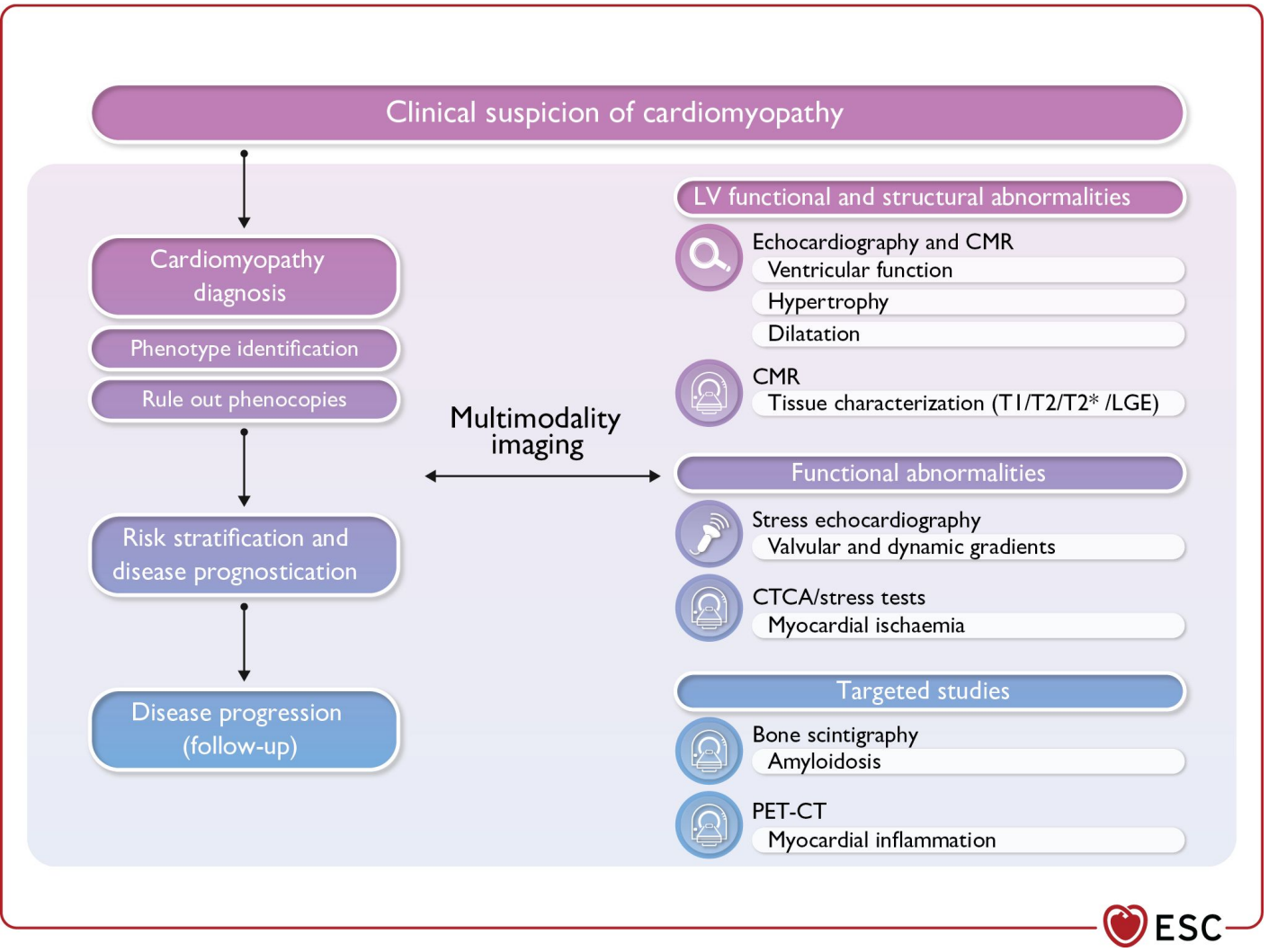
Recommendations	Class	Level
It is recommended that all patients with cardiomyopathy and their relatives have access to multidisciplinary teams with expertise in the diagnosis and management of cardiomyopathies.	I	C
Timely and adequate preparation for transition of care from paediatric to adult services, including joint consultations, is recommended in all adolescents with cardiomyopathy.	I	C

A shared and coordinated care approach between cardiomyopathy specialists and general adult and paediatric cardiology centres is strongly recommended

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			
		FKTN variants			
Sensorineural deafness	Mitochondrial diseases	Epicardin variants			
	NSML	Mitochondrial diseases			
Visual impairment	Mitochondrial diseases	*CRYAB*			
	ATTRv or hereditary ATTR	Type 2 myotonic dystrophy			
	Danon disease				
	Anderson–Fabry disease[a]				
Gait disturbance	Friedreich ataxia	Dystrophinopathies	Myofibrillar myopathies		
		Sarcoglycanopathies			
		Myofibrillar myopathies			
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

Multimodality imaging

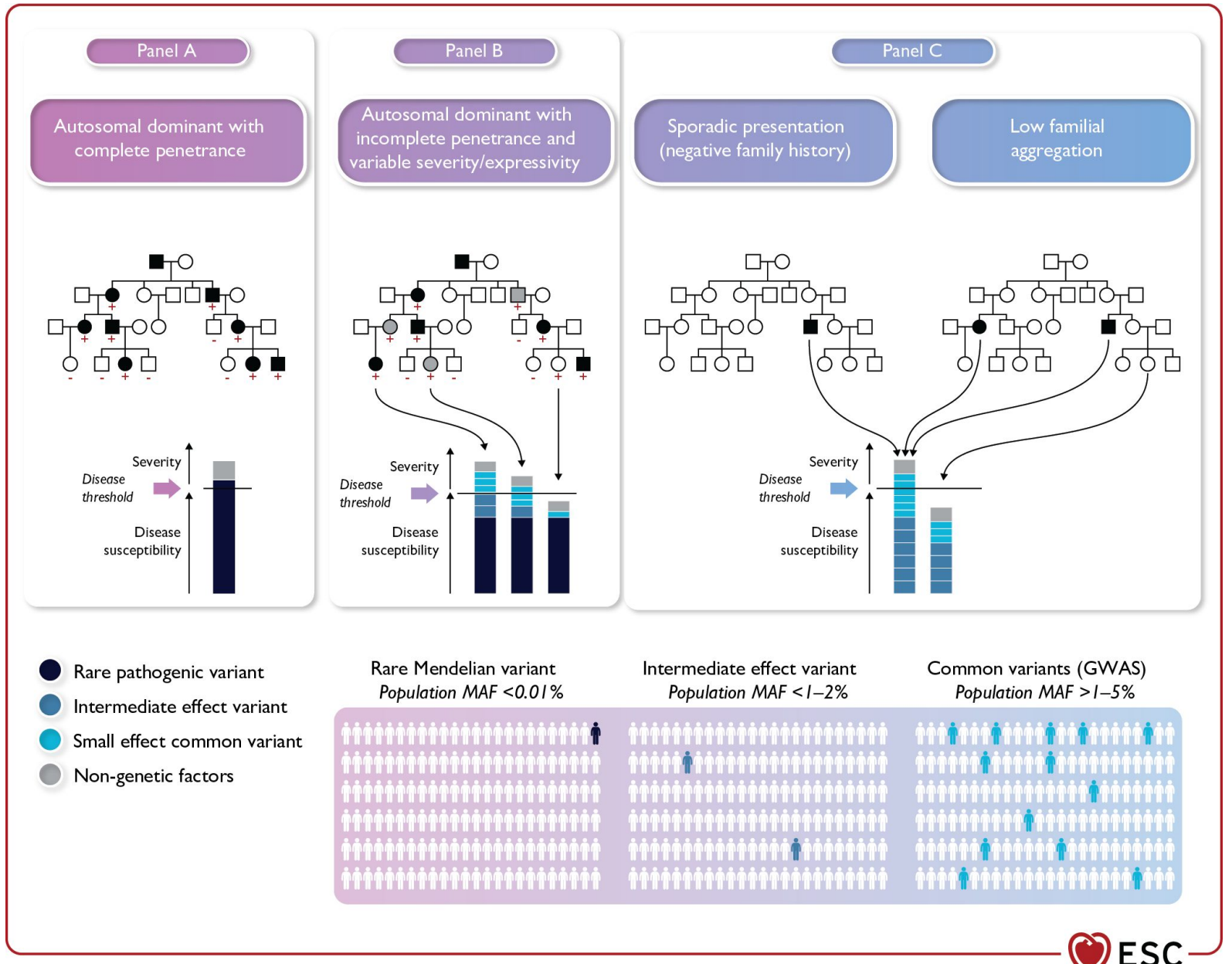


Recommendation	Class	Level
A comprehensive evaluation of cardiac dimensions and LV and RV systolic (global and regional) and LV diastolic function is recommended in all patients with cardiomyopathy at initial evaluation, and during follow-up, to monitor disease progression and aid risk stratification and management.	I	B

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.	IIb	C

The genetic architecture of the cardiomyopathies



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

Gene	Cardiomyopathy phenotype					Associated phenotype
	HCM	DCM	NDLVC	ARVC	RCM	
ABCC9	●[a]	○				[a] Cantu syndrome
ACTA1	○					
ACTC1	●	◐	◐	○	◐	
ACTN2[b]	◐	◐	◐			
ALPK3	●					
ANKRD1	○	○				
BAG3	●[a]	●●			◐	[a] Myofibrillar myopathy
CACNA1C	●[c]					[c] Timothy syndrome
CACNB2	○					
CALR3	○					
CASQ2	○					
CAV3	●[a]					[a] Caveolinopathy

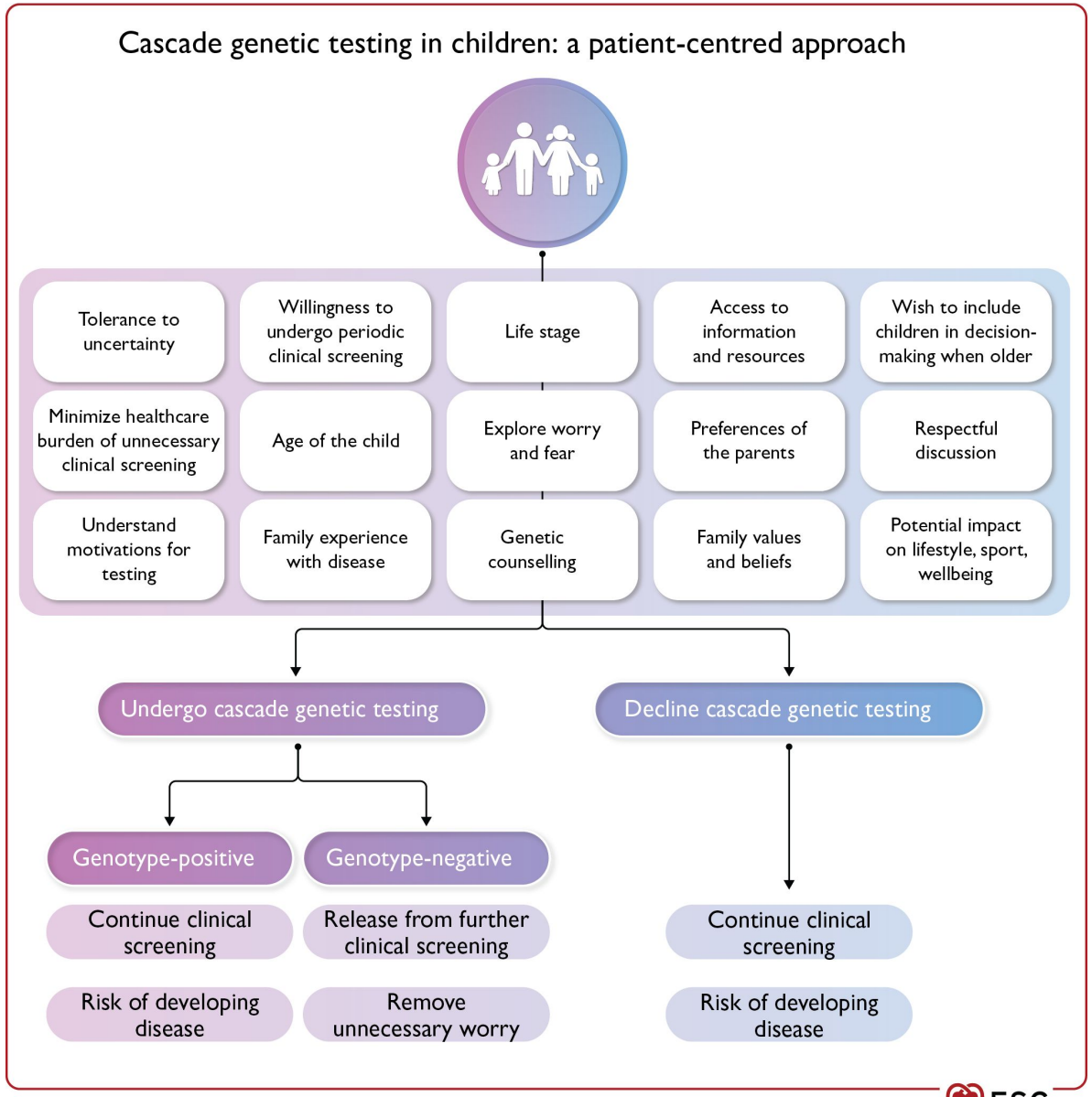
… continued

Genetic testing in cardiomyopathies

For the patient	For relatives
Diagnosis	An individual who does not carry the genetic variant proved to be responsible for disease in their family can be confidently reassured and discharged without surveillance, while an individual who carries a disease-causing variant can be followed closely, and potentially treated early
Prognosis	
Therapy	
Reproductive advice	

Recommendations	Class	Level
Index patients		
Genetic testing is recommended in patients fulfilling diagnostic criteria for cardiomyopathy in cases where it enables diagnosis, prognostication, therapeutic stratification, or reproductive management of the patient, or where it enables cascade genetic evaluation of their relatives who would otherwise be enrolled into long-term surveillance.	I	B

A patient-centred approach to cascade genetic testing of children

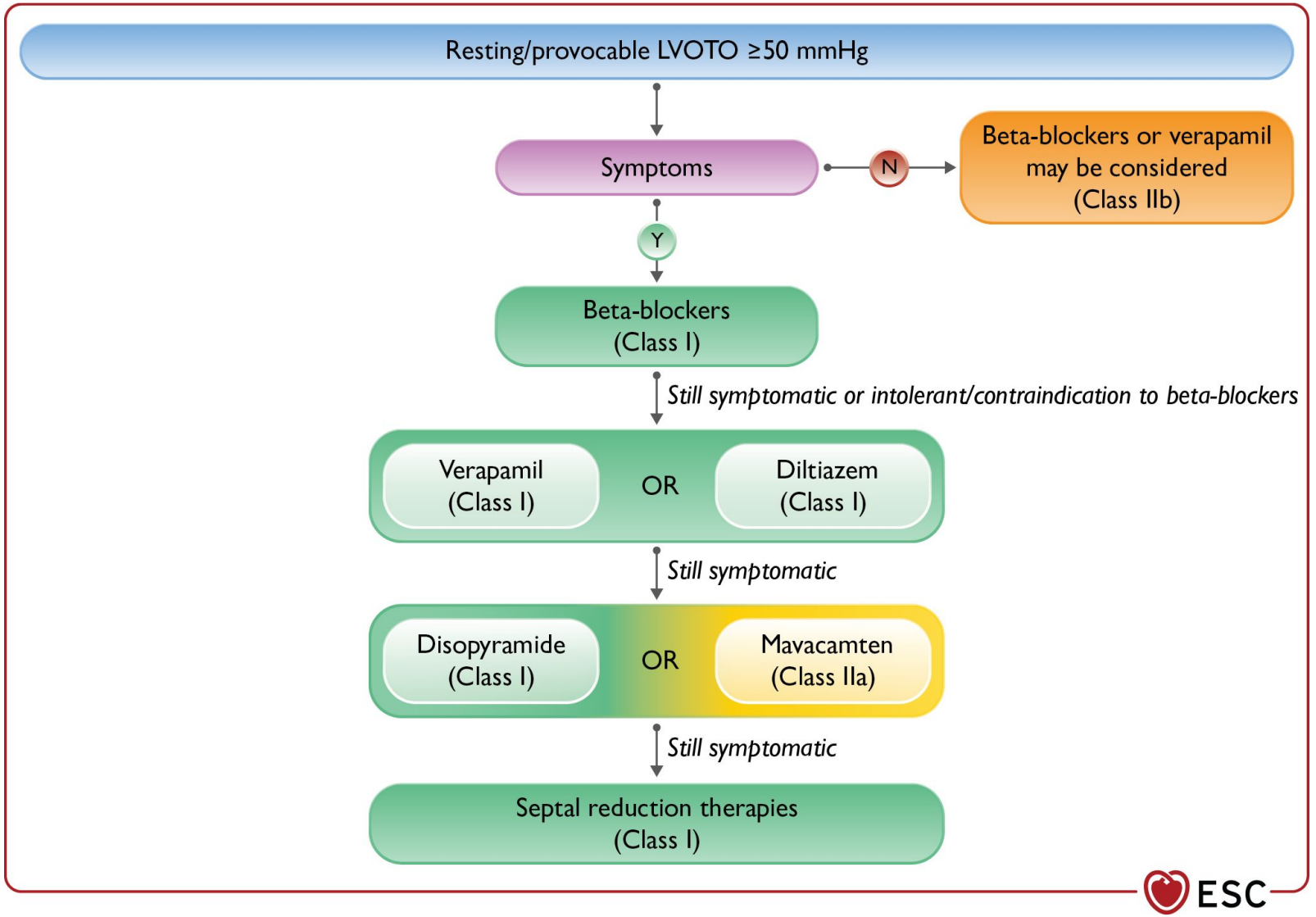


HYPERTROPHIC CARDIOMYOPATHY

Recommendations for medical treatment of LVOTO

Recommendations	Class	Level
Non-vasodilating beta-blockers, titrated to maximum tolerated dose, are recommended as first-line therapy to improve symptoms in patients with resting or provoked LVOTO.	I	B
Verapamil or diltiazem, titrated to maximum tolerated dose, are recommended to improve symptoms in symptomatic patients with resting or provoked LVOTO who are intolerant or have contraindications to beta-blockers.	I	B
Disopyramide, titrated to maximum tolerated dose, is recommended in addition to a beta-blocker (or, if this is not possible, with verapamil or diltiazem) to improve symptoms in patients with resting or provoked LVOTO.	I	B
Cardiac myosin ATPase inhibitor (mavacamten), titrated to maximum tolerated dose with echocardiographic surveillance of LVEF, should be considered in addition to a beta-blocker (or, if this is not possible, with verapamil or diltiazem) to improve symptoms in adult patients with resting or provoked LVOTO.	IIa	A
Cardiac myosin ATPase inhibitor (mavacamten), titrated to maximum tolerated dose with echocardiographic surveillance of LVEF, should be considered as monotherapy in symptomatic adult patients with resting or provoked LVOTO (exercise or Valsalva manoeuvre) who are intolerant or have contraindications to beta-blockers, verapamil/diltiazem, or disopyramide.	IIa	B
Oral or i.v. beta-blockers and vasoconstrictors should be considered in patients with severe provocable LVOTO presenting with hypotension and acute pulmonary oedema who do not respond to fluid administration.	IIa	C
Disopyramide, titrated to maximum tolerated dose, may be considered as monotherapy in patients who are intolerant to or have contraindications to beta-blockers and verapamil/diltiazem to improve symptoms in patients with resting or provoked LVOTO.	IIb	C

Management of left ventricular outflow tract obstruction



SUDDEN DEATH RISK PREVENTION IN CARDIOMYOPATHIES

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

Recommendations	Class	Level
Secondary prevention		
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
Primary prevention		
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

Additional recommendations for prevention of sudden cardiac death in patients with hypertrophic cardiomyopathy (2)

Recommendations	Class	Level
Primary prevention (continued)		
Implantation of an ICD should be considered in patients with an estimated 5-year risk of sudden death of ≥6%, following detailed clinical assessment that considers: (i)the lifelong risk of complications; (ii)competing mortality risk from the disease and comorbidities; AND (i) the impact of an ICD on lifestyle, socio-economic status, and psychological health.	**IIa**	**B**
In patients with LV apical aneurysms, decisions about primary prevention ICD based on an assessment of risk using the HCM Risk-SCD or a validated paediatric risk prediction (e.g. HCM Risk-Kids) tool and not solely on the presence of the aneurysm should be considered.	**IIa**	**B**

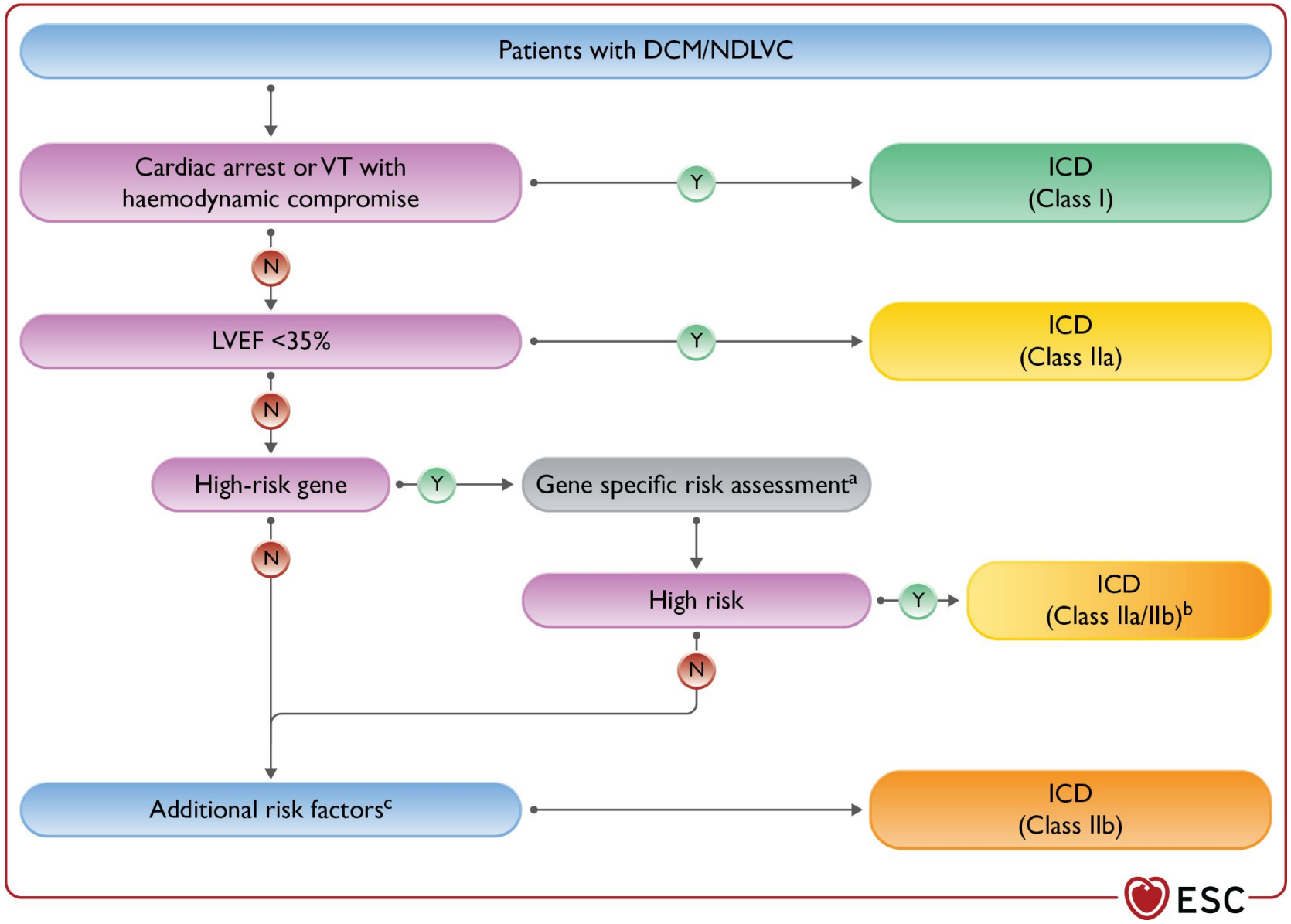
Existen importantes diferencias en el proceso de estratificación del riesgo entre las guías Europeas y Americanas

Usar como referencia las guías ESC para MS MCH

MS: muerte súbita, MCH: miocardiopatía hipertrófica

DILATED CARDIOMYOPATHY

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



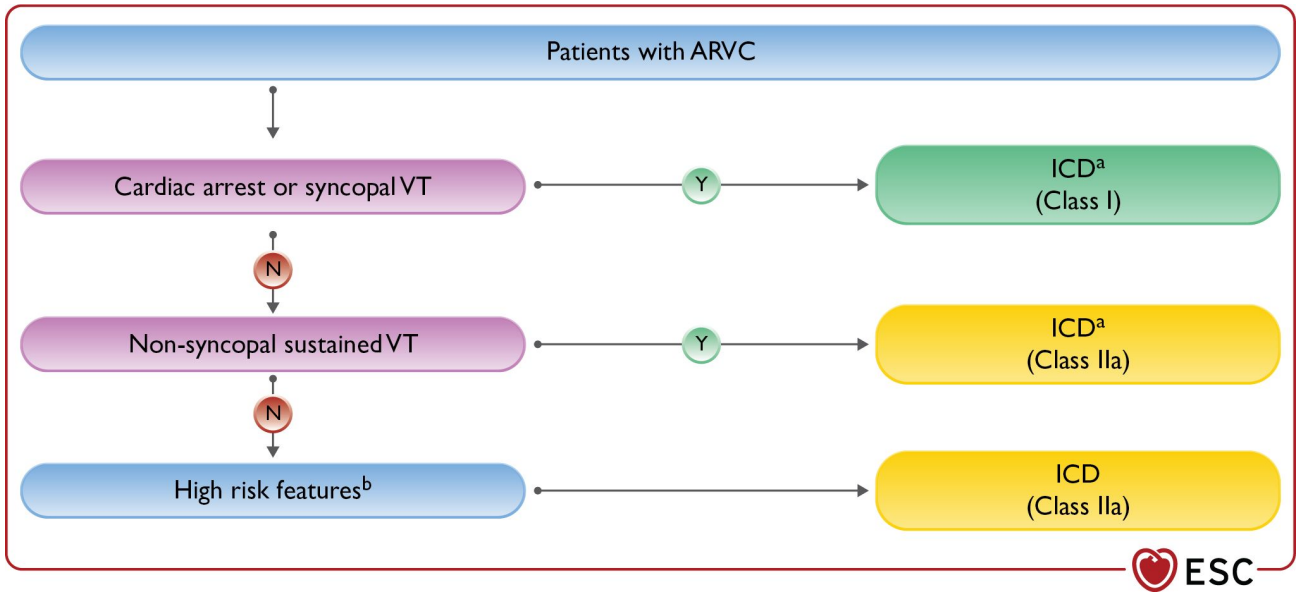
2023 ESC Guidelines for the management of cardiomyopathies
(European Heart Journal; 2023 – doi:10.1093/eurheartj/ehad 194)

High-risk genotypes and associated predictors of sudden cardiac death

Gene	Annual SCD rate	Predictors of SCD
LMNA	5–10%	Estimated 5-year risk of life-threatening arrhythmia using LMNA risk score https://lmna-risk-vta.fr
***FLNC*-truncating variants**	5–10%	LGE on CMR LVEF<45%
TMEM43	5–10%	Male Female and any of the following: LVEF <45%, NSVT, LGE on CMR, >200 VE on 24h Holter ECG
PLN	3–5%	Estimated 5-year risk of life-threatening arrhythmia using *PLN* 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%

ARRHYTHMOGENIC RIGHT VENTRICULAR CARDIOMYOPATHY

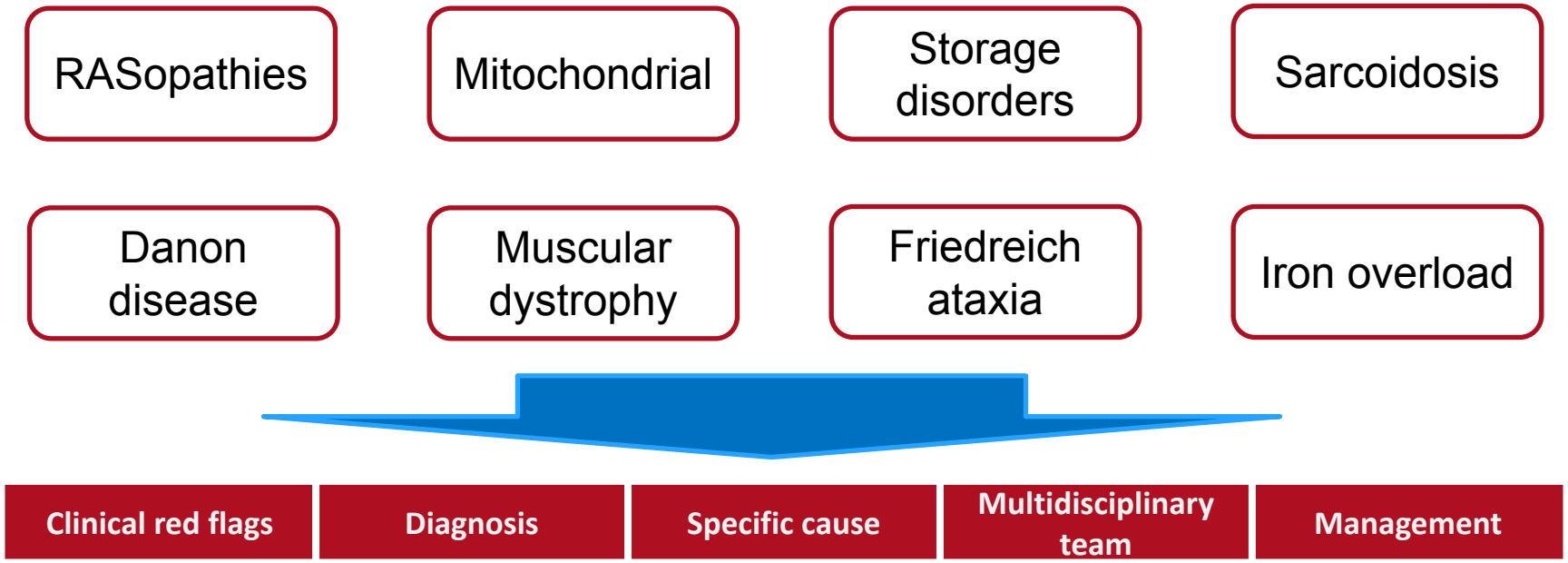
Prevention of sudden cardiac death in patients with arrhythmogenic right ventricular cardiomyopathy



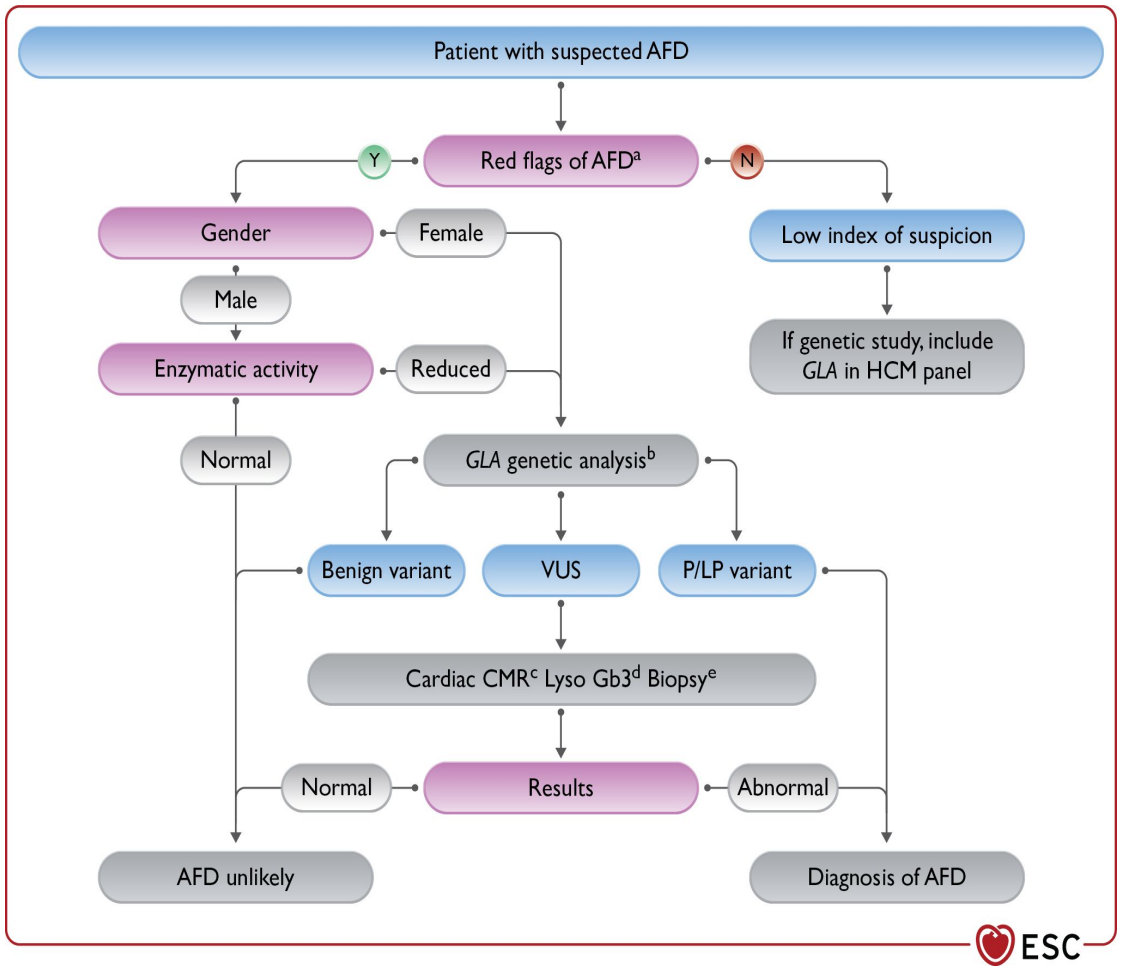
Recommendations	Class	Level
Secondary prevention		
An ICD is recommended to reduce the risk of sudden death and all-cause mortality in patients with ARVC who have survived a cardiac arrest or have recovered from a ventricular arrhythmia causing haemodynamic instability.	I	A
An ICD should be considered in ARVC patients who have suffered a haemodynamically tolerated VT.	IIa	B
Primary prevention		
High-risk features should be considered to aid individualized decision-making for ICD implantation in patients with ARVC.	IIa	B
The updated 2019 ARVC risk calculator should be considered to aid individualized decision-making for ICD implantation in patients with ARVC.	IIa	B

SYNDROMIC FORMS OF CARDIOMYOPATHY

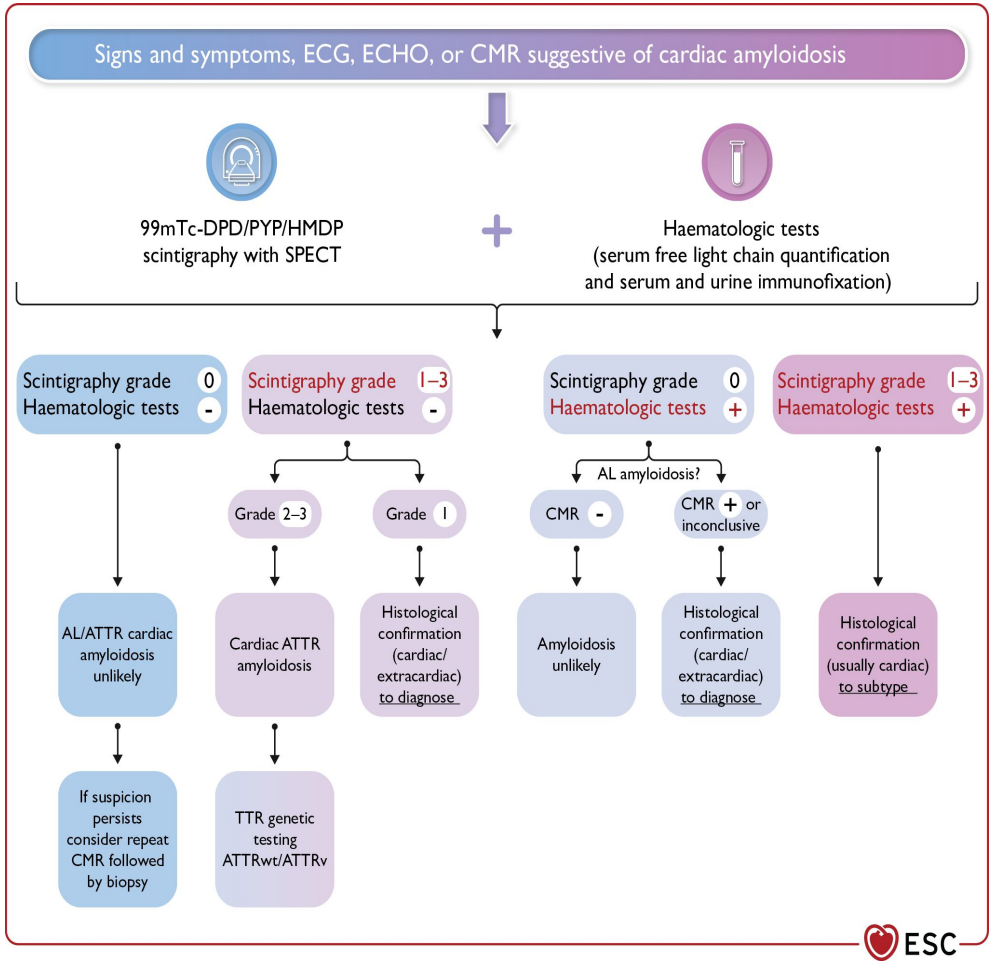
Syndromic and metabolic cardiomyopathies



Anderson–Fabry disease



Cardiac amyloidosis



Additional recommendations

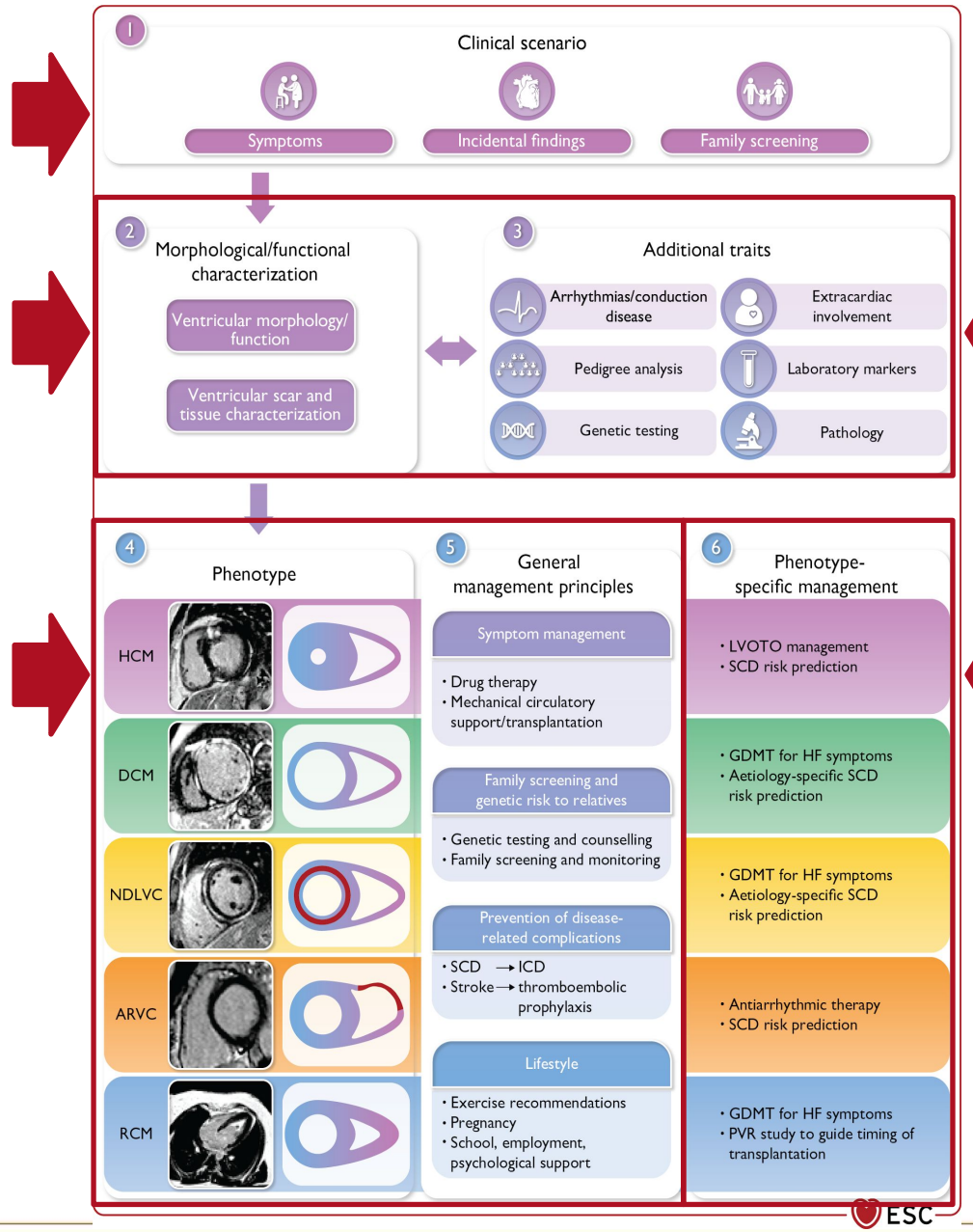
- Reproductive issues
- Non-cardiac surgery
- Cardiovascular risk factors
- Guidance for daily activities for patients

The patient pathway

‘Cardiomyopathy mindset’

Phenotype and general management

Key aspects in the evaluation and management of cardiomyopathies



Evaluation

Specific management

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