Kardiyomiyopatiler

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TKD Hipertansiyon Yönetim Kurulu

Kardiyomiyopati

- Gözlenen miyokard anormalliğine neden olacak kadar koroner arter hastalığı , hipertansiyon, kapak hastalığı ve konjenital kalp hastalığının yokluğunda, kalp kasının yapısal ve işlevsel olarak anormal olduğu bir miyokard bozukluğu' olarak tanımlanır.

ESC 2023 Kılavuzu

ESC European Society of Cardiology

- Kardiyomiyopatileri kapsayan ilk uluslararası kılavuz
- Yeni fenotipik tanımlama
- Başvurudan tedaviye KMP yönetimi
- KMP ailelerine klinik-genetik tarama
- Pediatrik dönemden erişkinliğe KMP aşamalarına odaklanma
- SCD risk değerlendirme
- HCM tedavisinde yeni öneriler
- Multidispliner yaklaşım

Tanımlama

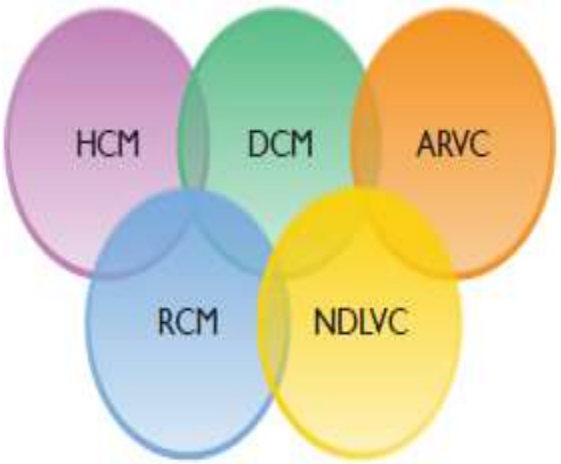
- Tanımlama ilk tanıdaki dominant kardiyak fenotipe göre
- Morfolojik ve fonksiyonel karakteristiklere odaklanma
- Kanalopatileri KMP olarak kabul etmek için yeterli kanıt yok
- Sol ventriküler Nonkompaction
- Takotsubo Sendromu

Table 3 Morphological and functional traits used to describe cardiomyopathy phenotypes

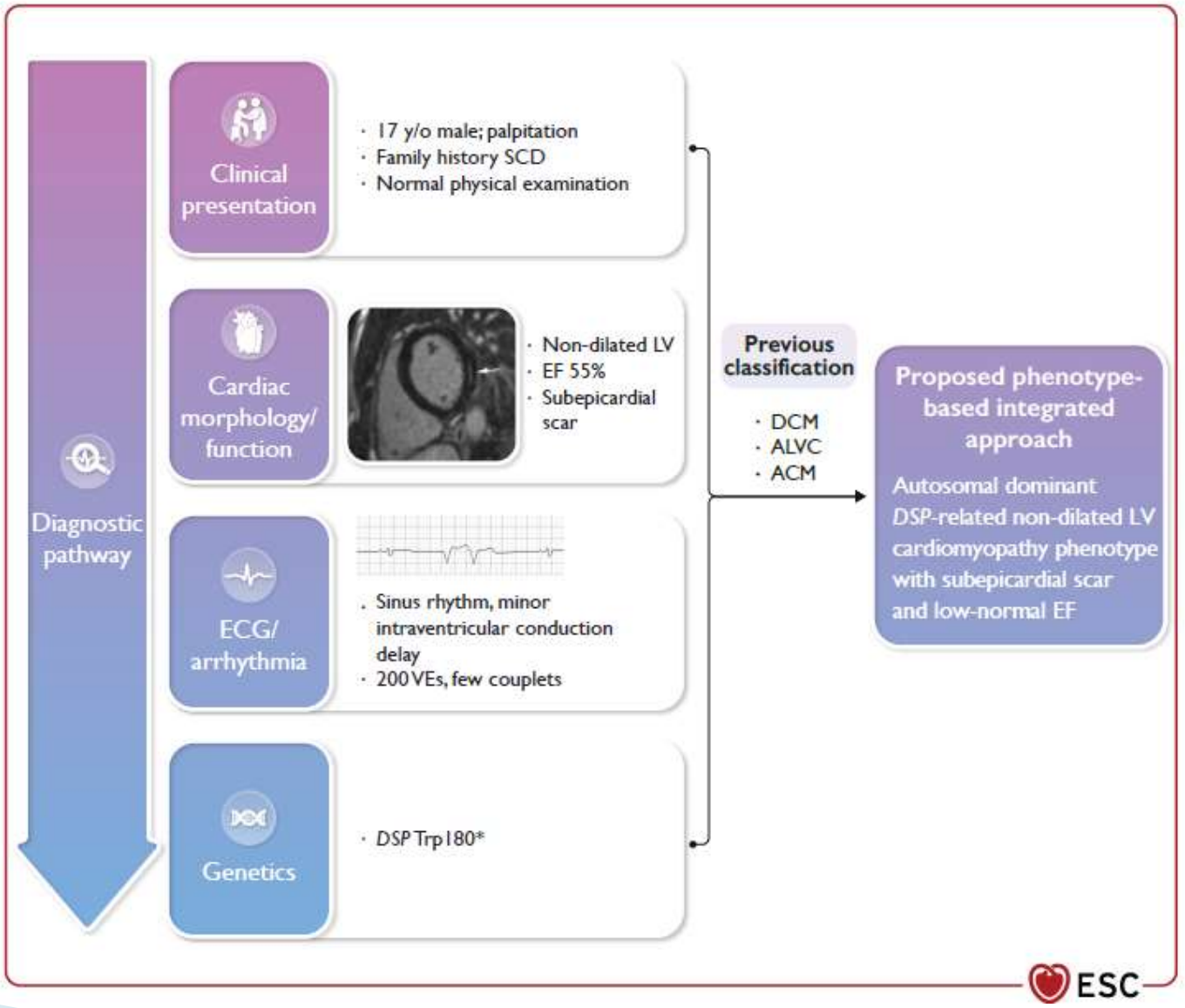
Morphological traits
Ventricular hypertrophy: left and/or right
Ventricular dilatation: left and/or right
Non-ischaemic ventricular scar and other myocardial tissue characterization features on cardiac magnetic resonance
Functional traits
Ventricular systolic dysfunction (global, regional)
Ventricular diastolic dysfunction (restrictive physiology)

Kardiyomiyopati Fenotipleri

- Hipertrofik KMP
- Dilate KMP
- Nondilate Sol Ventriküler KMP
- Aritmojenik Sağ Ventrikül KMP
- Restiriktif KMP

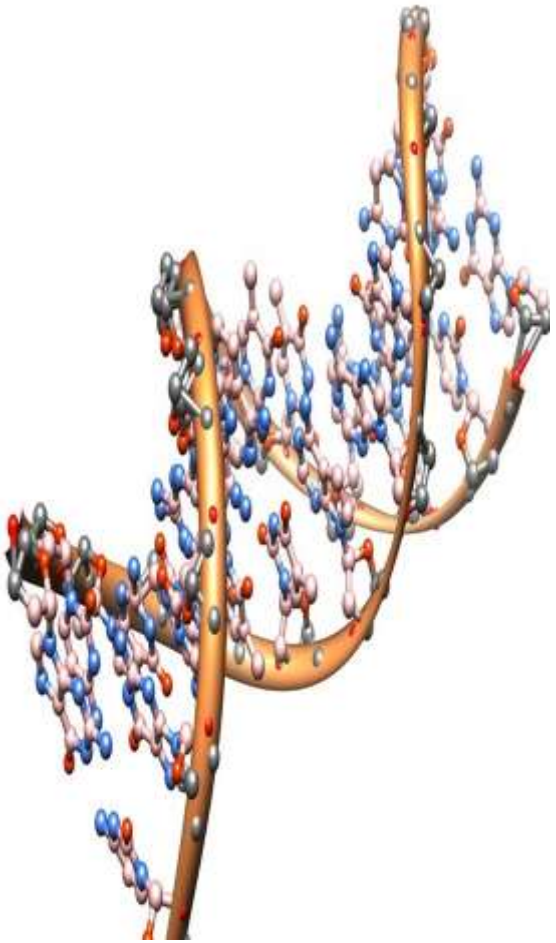


Fenotip Temelli Entegre Yaklaşım

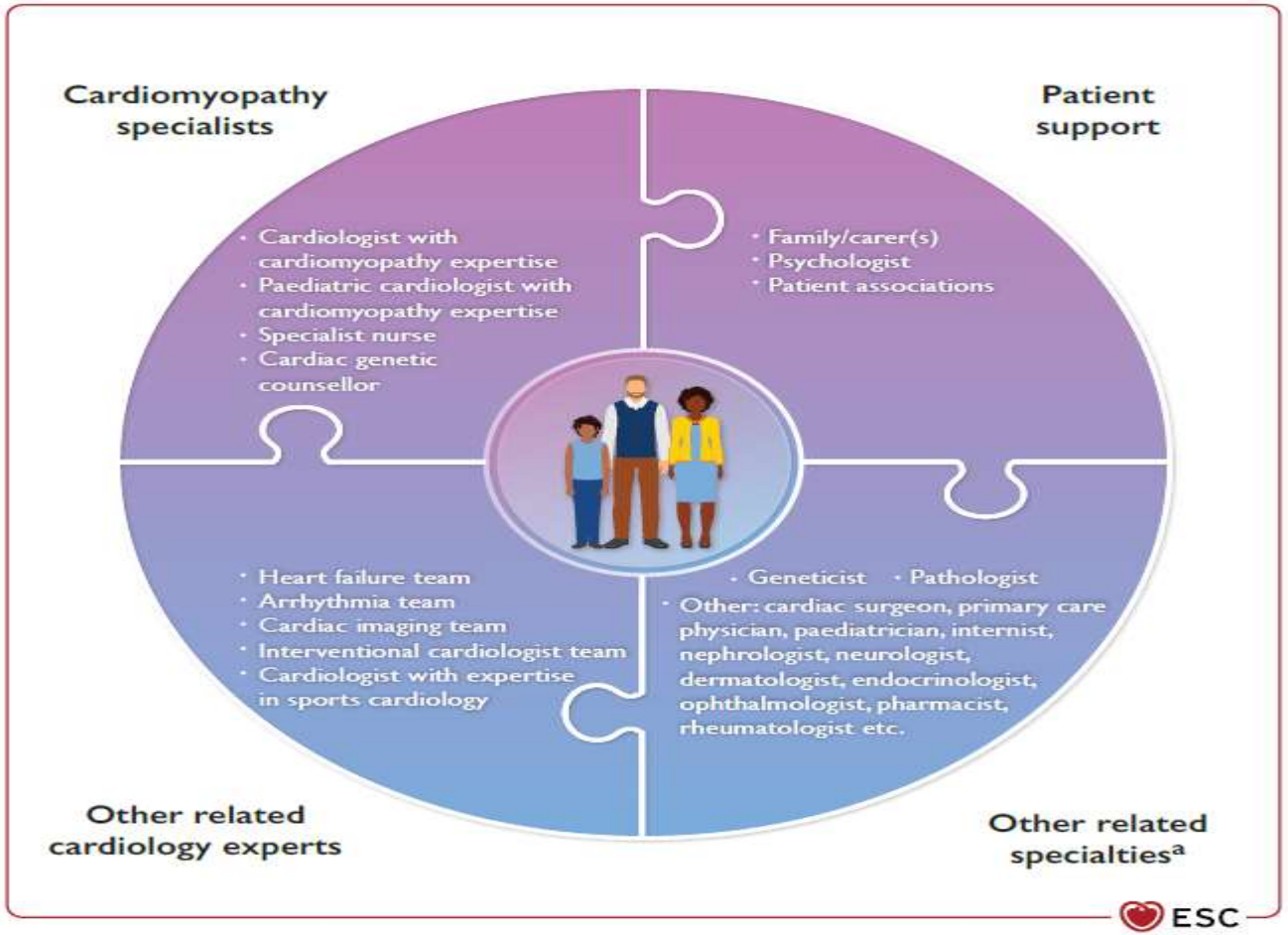


Second Hit Theory

- Titin gene truncating variants (TTNtv) alkolik kmp yatkınlık
- TTNtv kansere bağlı toksisiteye yatkınlık
- PPKMP'de (%15) TTNtv, DSP , FLNC ve BAG3
- LVH ve HCM'lerin %0.9'da FABRY
- ATTTR-CA, Aort stenozunda %8, LVH+HFPEF %12, karpal tünel %7
- Akut miyokarditte desmozomal protein gen varyantları yüksek



Multidispliner Yaklaşım



EKG

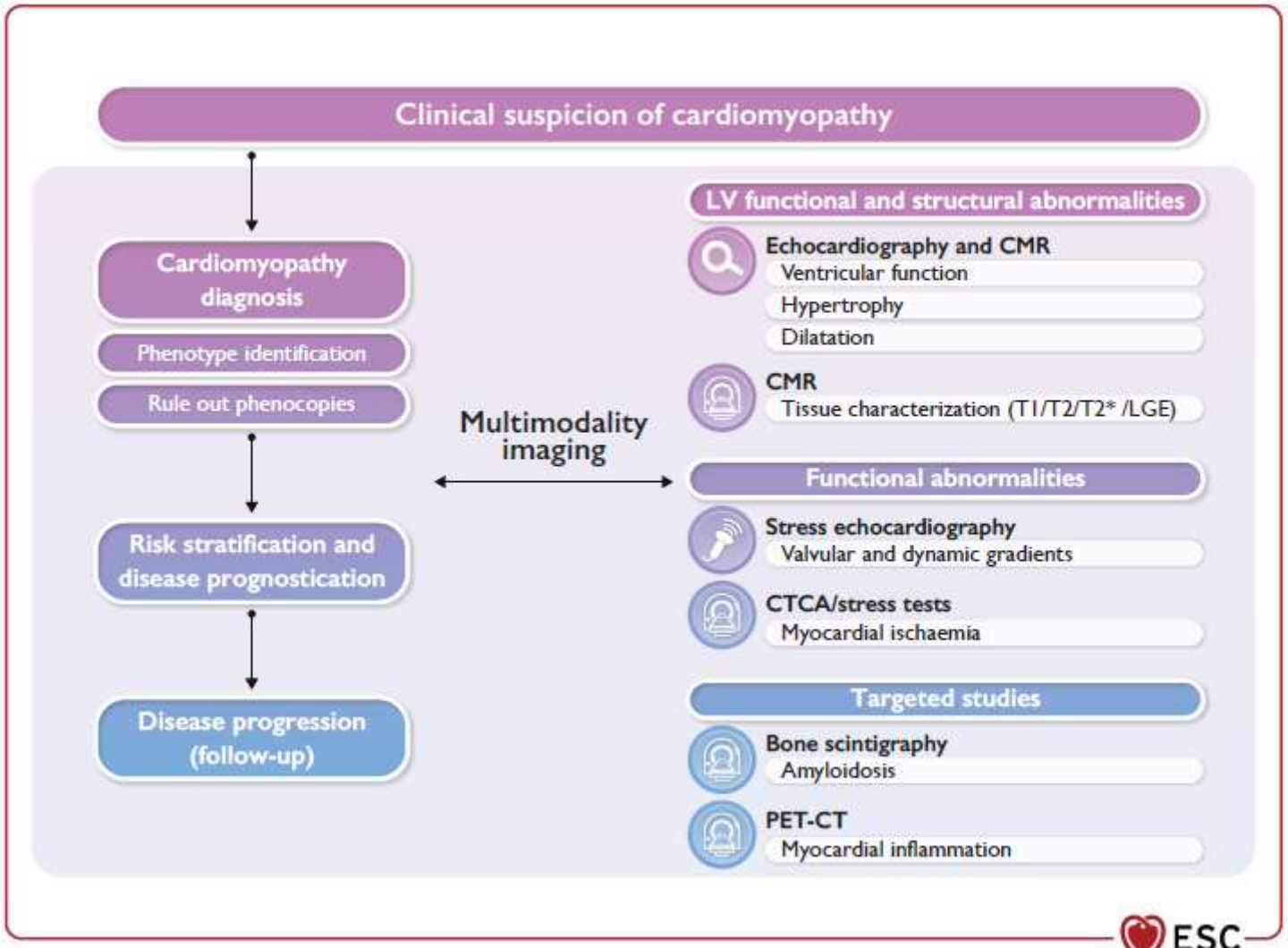
HCM	Short PR interval/pre-excitation	Glycogenosis Danon disease *PRKAG2* cardiomyopathy Anderson–Fabry disease Mitochondrial disease
	AV block	Amyloidosis Anderson–Fabry disease (late stage) Danon disease Sarcoidosis *PRKAG2* cardiomyopathy
	Extreme LVH	Danon disease Glycogenosis (e.g. Pompe disease) *PRKAG2* cardiomyopathy
	Low QRS voltage[a]	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
	Posterolateral infarction pattern	Dystrophinopathy Limb-girdle muscular dystrophy Sarcoidosis
	Extremely low QRS amplitude	*PLN* variant
NDLVC	AV block	Laminopathy Desminopathy
	Extremely low QRS amplitude	*PLN* 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

Tetkik

Kardiyomiyopatilerin tanısında laboratuar testlerine yönelik öneriler		
Etiyolojiyi ve hastalığın ciddiyetini değerlendirmek ve ekstrakardiyak belirtilerin saptanmasında ve sekonder organ fonksiyon bozukluğunun değerlendirilmesine yardımcı olmak için şüpheli veya doğrulanmış kardiyomiyopatisi olan tüm hastalarda rutin (ilk basamak) laboratuar testleri önerilir.	I	C
Kardiyomiyopatili hastalarda ekokardiyografik değerlendirme önerisi		
Kardiyomiyopatili tüm hastalarda ilk değerlendirmede, takip sırasında, hastalığın progresyonunu izlemek ve risk sınıflandırması yönetiminde yardımcı olmak için kardiyak boyutların ve LV ve RV sistolik (global ve bölgesel) ve LV diyastolik fonksiyonunun kapsamlı değerlendirmesi önerilir.	I	B
Kardiyomiyopatili hastalarda kardiyak manyetik rezonans endikasyonuna ilişkin öneriler		
Kardiyomiyopatili hastalarda ilk değerlendirmede kontrastlı kardiyak MR önerilir.	I	B
Bilgisayarlı tomografi ve nükleer görüntülemeye yönelik öneriler		
ATTR ile ilişkili kardiyak amiloidoz şüphesi olan hastalarda tanıya yardımcı olması için DPD/PYP/HMDP ile kemik sintigrafisi önerilir.	I	B
Kardiyomiyopatilerde genetik danışmanlık ve test önerileri		

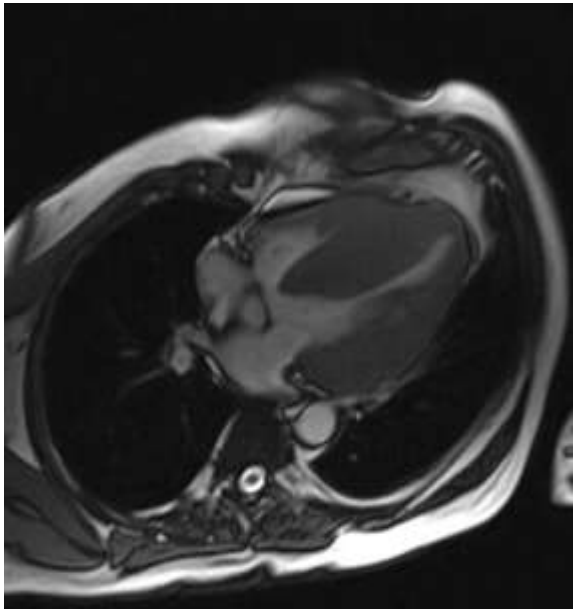
Table 8 First-level (to be performed in each patient) and second-level (to be performed in selected patients following specialist evaluation to identify specific aetiologies) laboratory tests, grouped by cardiomyopathy phenotype

Level	HCM	DCM	NDLVC	ARVC	RCM
First	• CK • Liver function • NT-proBNP[a] • Proteinuria • Renal function • Troponin	• Calcium • CK • Ferritin • Full blood count • Liver function • NT-proBNP[a] • Phosphate • Proteinuria • Renal function • Serum iron • Thyroid function • Troponin • Vitamin D (children)	• Calcium • CK • C-reactive protein • Full blood count • Liver function • NT-proBNP[a] • Phosphate • Proteinuria • Renal function • Troponin	• C-reactive protein • Liver function • NT-proBNP[a] • Renal function • Troponin	• CK • Ferritin • Full blood count • Liver function • NT-proBNP[a] • Proteinuria • Renal function • Serum angiotensin-converting enzyme • Serum iron • Troponin • Urine and plasma protein immunofixation, free light chains
Second	• Alpha-galactosidase A levels (males) and lyso-Gb3 • Carnitine profile • Free fatty acids • Immunofixation and free light chains • Lactic acid • Myoglobinuria • Pyruvate • PTH • Urine and plasma protein • Urine organic acids and plasma amino acids	• Carnitine profile • Free fatty acids • Lactic acid • Organ- and non–organ-specific serum autoantibodies • Serum angiotensin-converting enzyme • Thiamine • Viral serology • Urine organic acids and plasma amino acids	• Organ- and non-organ-specific serum autoantibodies • Viral serology		• Organ- and non–organ-specific autoantibodies • Serum angiotensin-converting enzyme


Clinical suspicion of cardiomyopathy
Cardiomyopathy diagnosis
Phenotype identification
Rule out phenocopies
Risk stratification and disease prognostication
Disease progression (follow-up)
Multimodality imaging
LV functional and structural abnormalities
Echocardiography and CMR
Ventricular function
Hypertrophy
Dilatation
CMR
Tissue characterization (T1/T2/T2* /LGE)
Functional abnormalities
Stress echocardiography
Valvular and dynamic gradients
CTCA/stress tests
Myocardial ischaemia
Targeted studies
Bone scintigraphy
Amyloidosis
PET-CT
Myocardial inflammation
ESC

Hipertrofik Kardiyomiyopati

- **Yetişkinler:** Yüklenme koşullarıyla açıklanamayan herhangi bir segmentte LV duvar kalınlığının ≥15 mm veya 13-14 mm+ genetik,aile öyküsü, EKG bulgusu
- **Çocuklar:** HCM LV duvar kalınlığının ortalamadan 2 standart sapma daha büyük olması (z-puanı >2)
- **Akrabalar:** Erişkin birinci derece akrabalarında LV duvar kalınlığının ≥13 mm
- Çocukta z-skorlarının <2 olması durumunda ilişkili morfolojik veya EKG anormalliklerinin varlığı şüphe uyandırmalıdır ancak tek başına HCM için tanısal değildir.



Görüntüleme Yöntemi

Table 17 **Imaging evaluation in hypertrophic cardiomyopathy**

Item to assess	Primary imaging modality	Comments
LV wall thickness	ECHO/CMR	• All LV segments from base to apex examined in end-diastole, preferably in the 2D short-axis view, ensuring that the wall thickness is recorded at mitral, mid-LV, and apical levels. • CMR is superior in the detection of LV apical and anterolateral hypertrophy, aneurysms,[580] and thrombi,[581] and is more sensitive in the detection of subtle markers of disease in patients with sarcomeric protein gene variants (e.g. myocardial crypts, papillary muscle abnormalities).[159,582,583]
Systolic function (global and regional)	ECHO/CMR	• Ejection fraction is a suboptimal measure of LV systolic performance when hypertrophy is present. • Doppler myocardial velocities and deformation parameters (strain and strain rate) are typically reduced at the site of hypertrophy despite a normal EF and may be abnormal before the development of increased wall thickness in genetically affected patients.
Diastolic function	ECHO	• Routine examination should include mitral inflow assessment, tissue Doppler imaging, pulmonary vein flow velocities, pulmonary artery systolic pressure, and LA size/volume.
Mitral valve	ECHO	• Assess presence and degree of SAM and mitral regurgitation. The presence of a central- or anteriorly directed jet of mitral regurgitation should raise suspicion of an intrinsic/primary mitral valve abnormality and prompt further assessment.
LVOT	ECHO	• See *Figure 12*.
LA dimensions	ECHO/CMR	• Provides important prognostic information.[365,525,584] • Most common mechanisms of LA enlargement are SAM-related mitral regurgitation and elevated LV filling pressures.
Myocardial fibrosis/LGE	CMR	• The distribution and severity of interstitial expansion can suggest specific diagnoses. Anderson–Fabry disease is characterized by a reduction in non-contrast T1 signal and the presence of posterolateral LGE.[134,155] In cardiac amyloidosis, there is often global, subendocardial or segmental LGE and a highly specific pattern of myocardial and blood-pool gadolinium kinetics caused by similar myocardial and blood T1 signals.[585,586]

Geç Gadolinium Tutulumu

- Hastaların %65'inde LGE mevcuttur
- Hipertrofi alanlarında yamalı orta duvar paterninde
- Ön ve arka RV giriş noktalarında
- Hipertrofiye olmayan segmentlerde olağan değil
- Tam kat LGE'nin eşlik ettiği hastalığın ileri evreleri

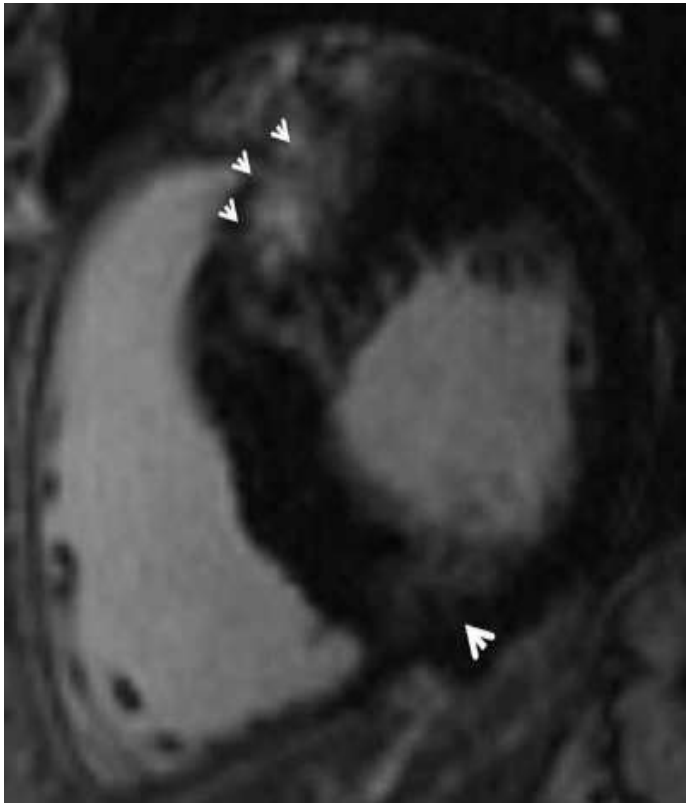
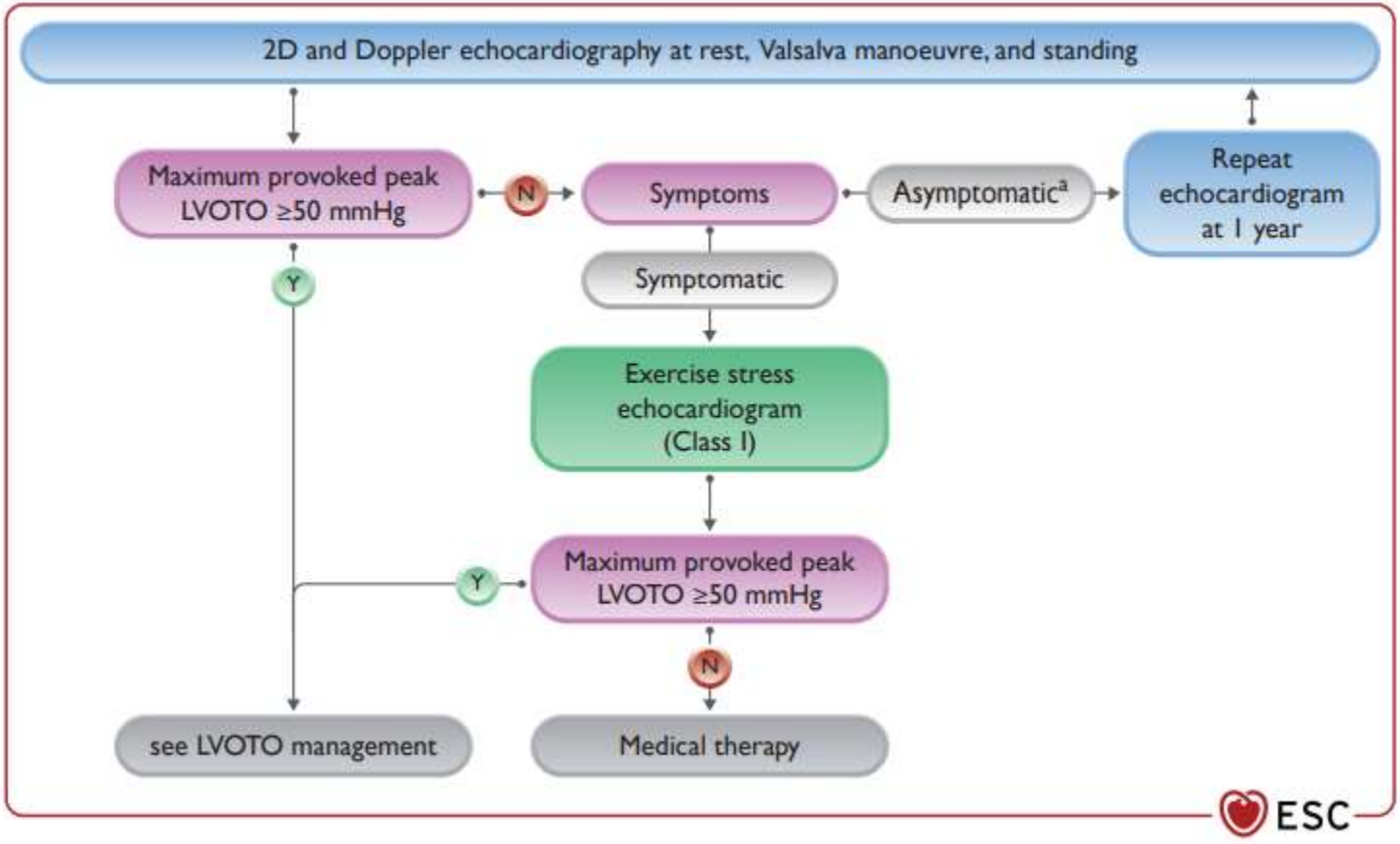


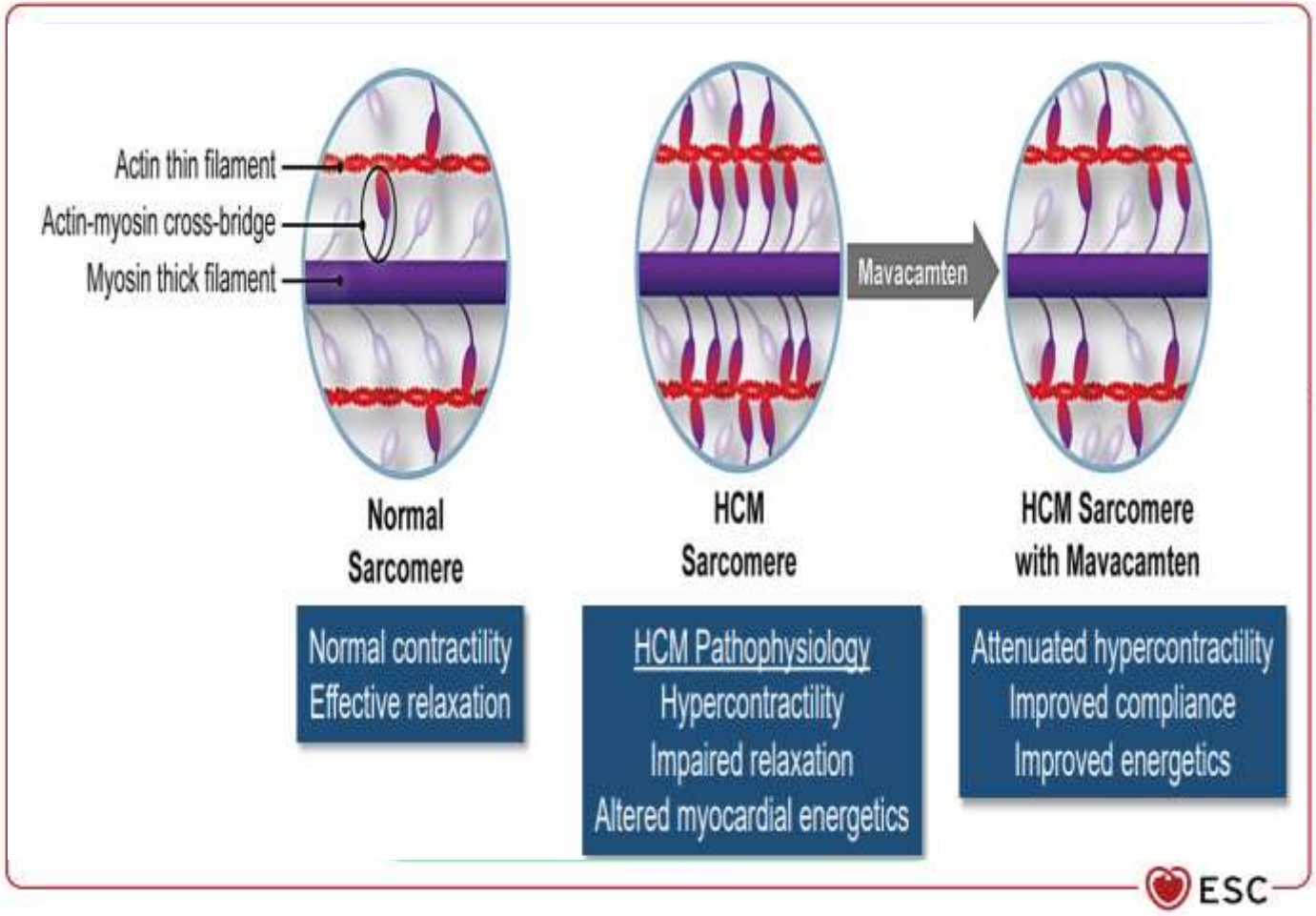
Table 18 **Echocardiographic features that suggest specific aetiologies in hypertrophic cardiomyopathy**

Finding	Specific diseases to be considered
Increased interatrial septum thickness	Amyloidosis
Increased AV valve thickness	Amyloidosis; Anderson–Fabry disease
Increased RV free wall thickness	Amyloidosis, myocarditis, Anderson–Fabry disease, Noonan syndrome, and related disorders
Mild-to-moderate pericardial effusion	Amyloidosis, myocarditis/myopericarditis
Ground-glass appearance of ventricular myocardium on 2D echocardiography	Amyloidosis
Concentric LVH	Glycogen storage disease, Anderson–Fabry disease, *PRKAG2* variants, Friedreich ataxia
Extreme concentric LVH (wall thickness ≥30 mm)	Danon disease, Pompe disease
Global LV hypokinesia (with or without LV dilatation)	Mitochondrial disease, TTR-related amyloidosis, *PRKAG2* variants, Danon disease, myocarditis, advanced sarcomeric HCM, Anderson–Fabry disease, Friedreich ataxia
RVOTO	Noonan syndrome and associated disorders
Apical sparing pattern on longitudinal strain imaging	Amyloidosis

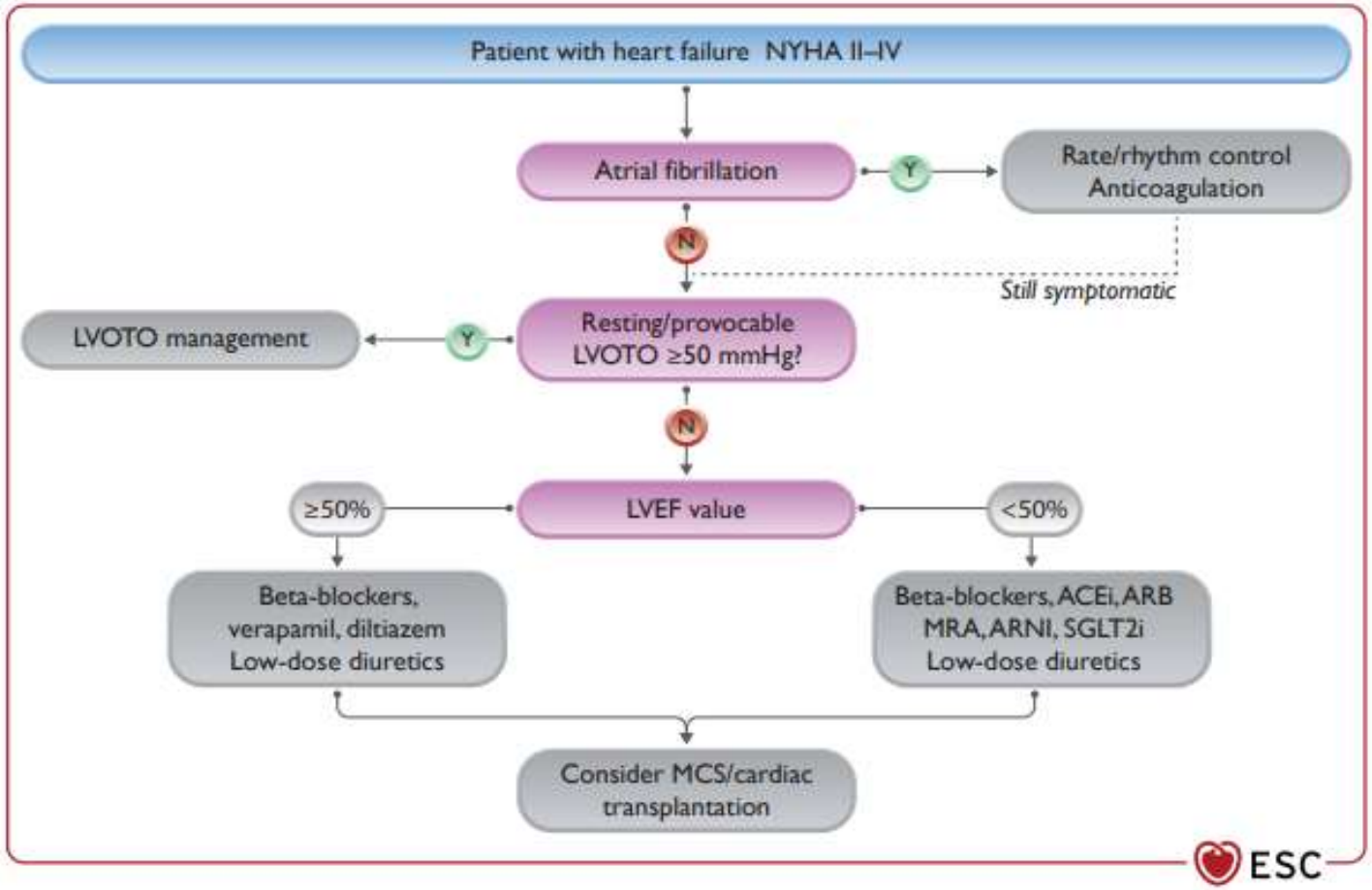
HCM Tedavi Önerileri

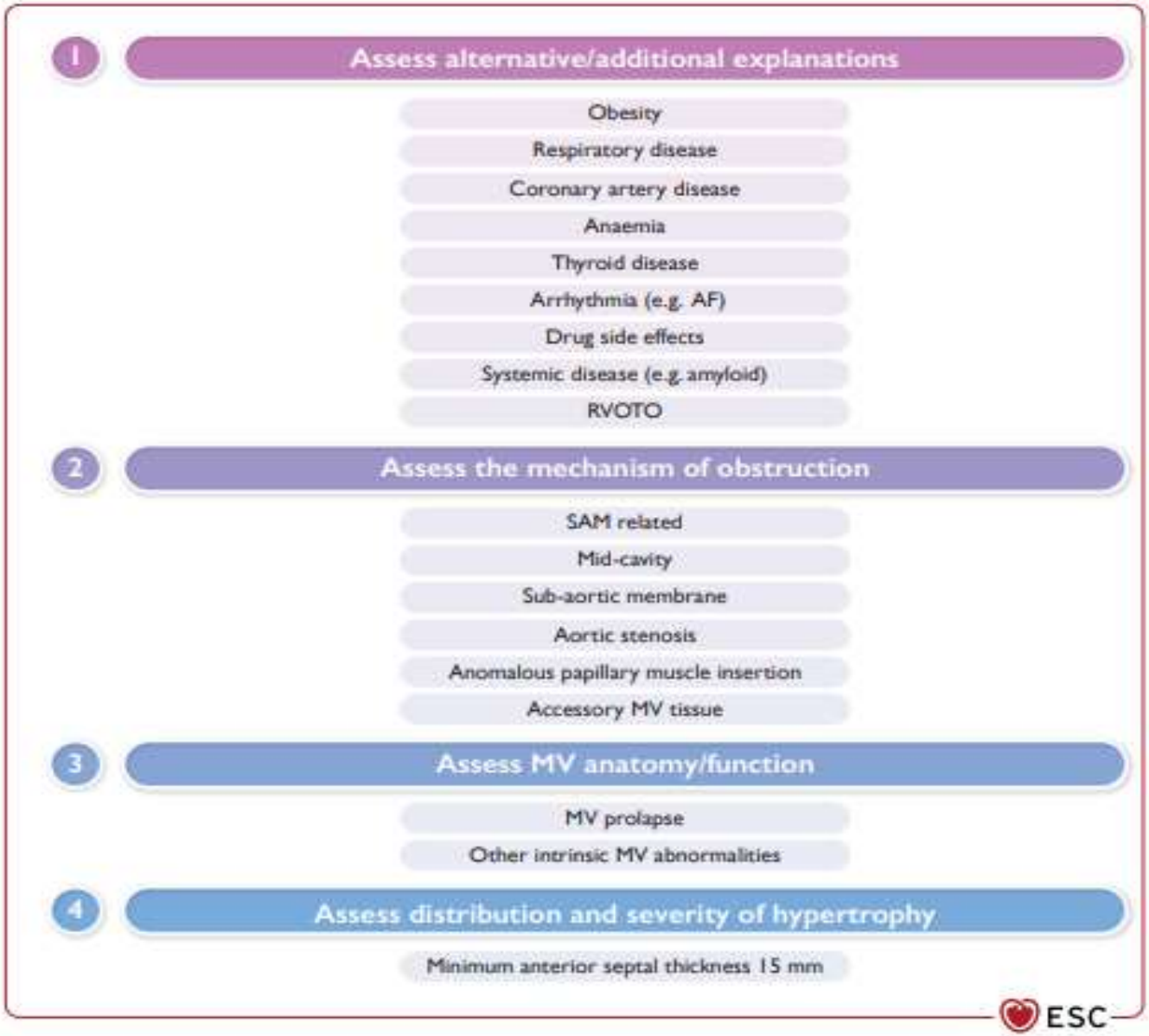


HCM Tedavi Önerileri



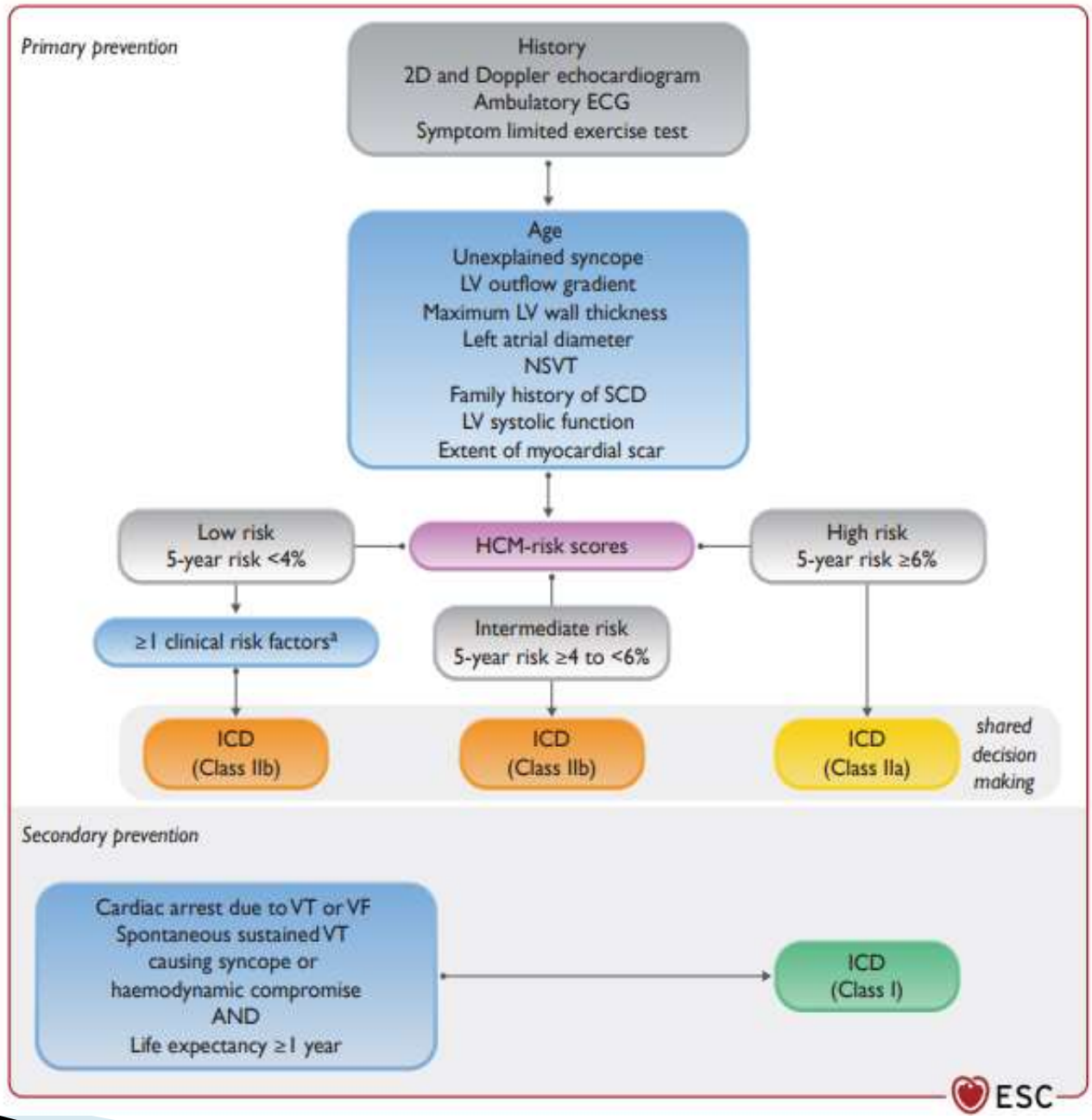
HCM Tedavi Önerileri




1
Assess alternative/additional explanations
Obesity
Respiratory disease
Coronary artery disease
Anaemia
Thyroid disease
Arrhythmia (e.g. AF)
Drug side effects
Systemic disease (e.g. amyloid)
RVOTO
2
Assess the mechanism of obstruction
SAM related
Mid-cavity
Sub-aortic membrane
Aortic stenosis
Anomalous papillary muscle insertion
Accessory MV tissue
3
Assess MV anatomy/function
MV prolapse
Other intrinsic MV abnormalities
4
Assess distribution and severity of hypertrophy
Minimum anterior septal thickness 15 mm
ESC

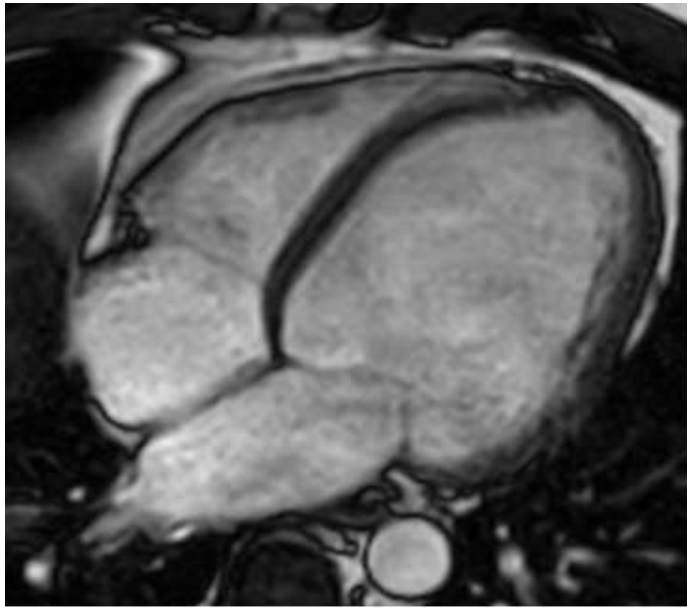
LGE>%15

LVEF <%50



Dilate Kardiyomiyopati

- Anormal yükleme koşulları veya KAH ile açıklanamayan LV dilatasyonu ve sistolik disfonksiyon
- Erkeklerde LVED >58 mm ve kadınlarda >52 mm
- LVEDV indeksinin erkeklerde ≥75 mL/m2 ve kadınlarda ≥62 mL/m2 olması
- LVEF <%50



Kardiyak MR

- Miyokardiyal ödem
- Fibrosis
- Karakterizasyon (demir,amiloid)
- Keskin Koroner Sendrom dışlanması
- Etyolojiyi belirleme
  - Miyokardit >>>>subepikardiyal
  - Sarkoidoz >>>yama şeklinde
  - Distrofinopatiler >>> inferolateral
  - LMNA taşıyıcılarında>>> septal orta duvar
  - DSP ve FLNC varyant >>> halka benzeri

Genetik

Gene	Annual SCD rate	Predictors of SCD
LMNA[185,186,438,541,865,878,879]	5–10%	Estimated 5-year risk of life-threatening arrhythmia using *LMNA* risk score (https://lmna-risk-vta.fr)
FLNC-truncating variants[866,867,880]	5–10%	LGE on CMR LVEF < 45%
TMEM43[868,881]	5–10%	Male Female and any of the following: LVEF <45%, NSVT, LGE on CMR, >200 VE on 24h Holter ECG
PLN[542,882,883]	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[185,186]	3–5%	LGE on CMR LVEF < 45%
RBM20[869]	3–5%	LGE on CMR LVEF < 45%

Search page / RBM20

RBM20

RNA binding motif protein 20 | 10q25.2 | NM_001134363.3

Gene Assessment | Publications | Associated Conditions

- Gene Summary
- Gene Expression
- Curated Information
- Curated Variants Distribution
- Gene Pathogenicity

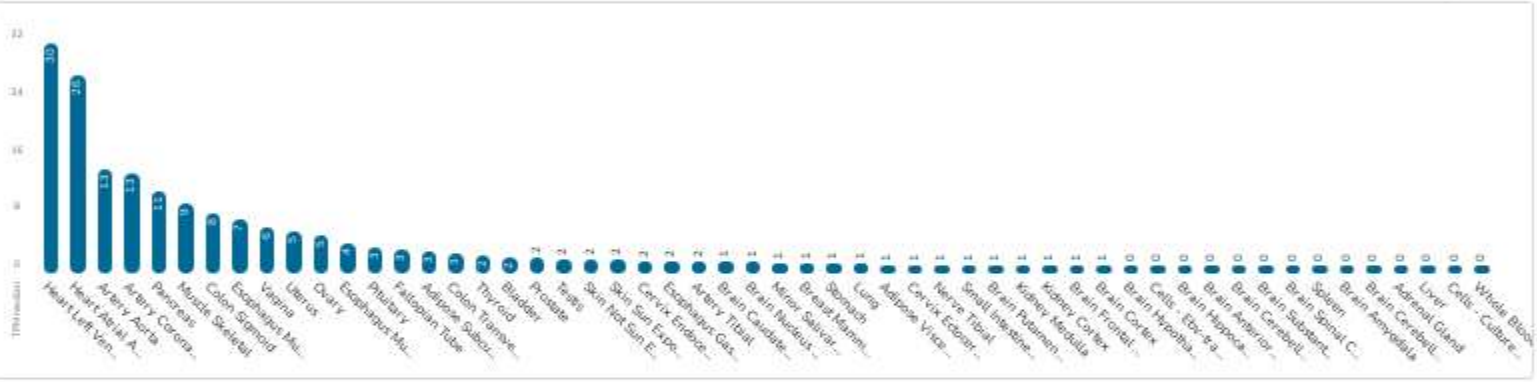
ClinVar | GeneReviews | gnomAD | GTEx | OMIM

Gene Summary

This gene encodes a protein that binds RNA and regulates splicing. Mutations in this gene have been associated with familial dilated cardiomyopathy. [provided by RefSeq, Apr 2014]

Gene Expression

GTEx



Curated Information

This gene was curated on Franklin community

Join the franklin community to find more details Sign up for free

Curated Variants Distribution

Classified variants from ClinVar, UniProt and Franklin Community Database

	LOF	Missense + Inframe Indel	Non-coding	Synonymous	Total
Pathogenic	15	29	2	0	46
VUS	69	756	82	25	932
Benign	3	255	184	403	845
Total	87	1040	268	428	1823

Gene Pathogenicity

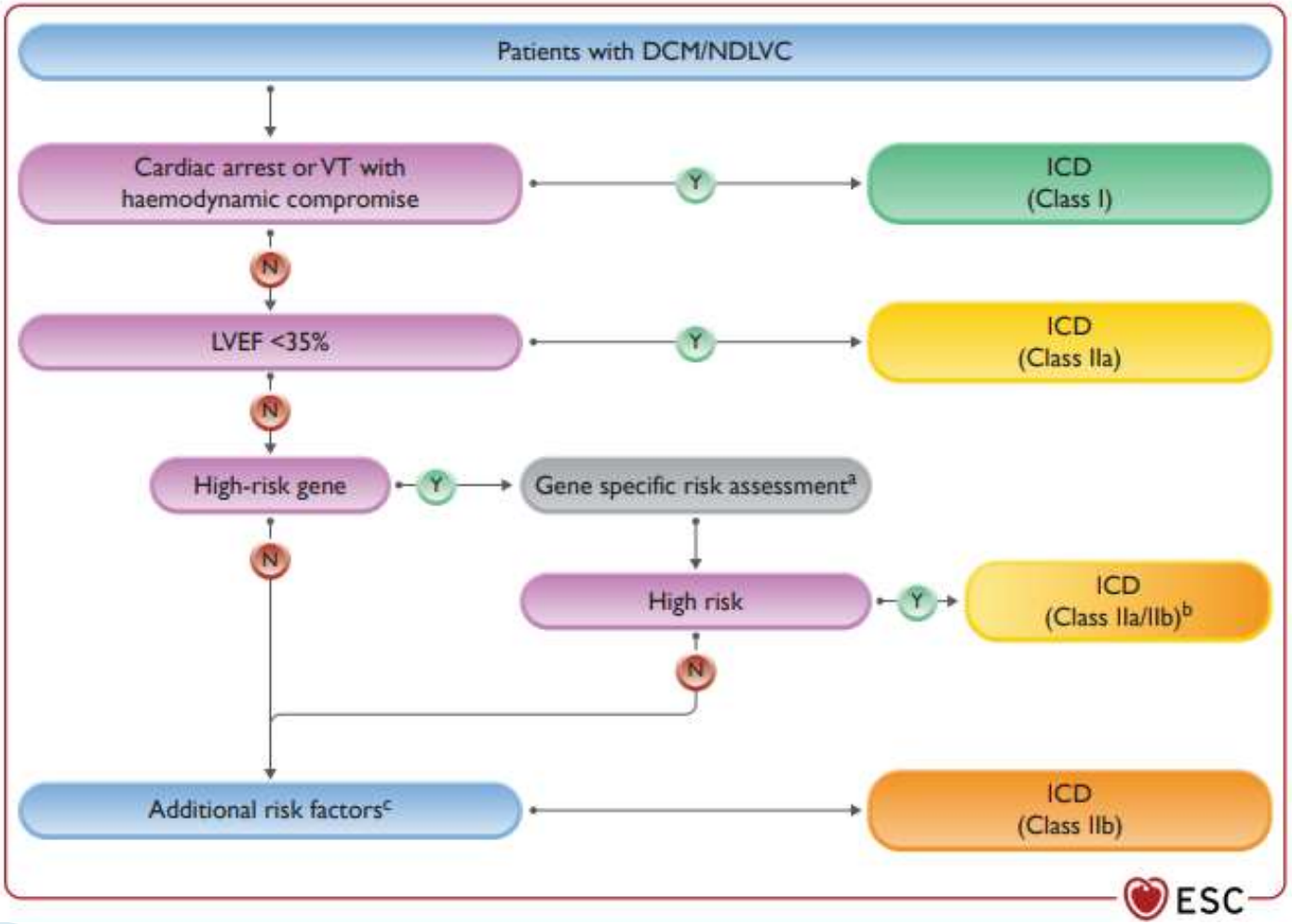
Sensitivity to LOF Mutations PVS1

Genetik olmayan Nedenler

Infection (post-myocarditis)
Viral (enteroviruses, adenoviruses, echoviruses, herpes viruses, parvovirus B19, HIV, SARS-CoV-2, etc.)
Bacterial (Lyme disease)
Mycobacterial
Fungal
Parasitic (Chagas disease)
Toxic and overload
Alcohol (ethanol)
Cocaine, amphetamines, ecstasy
Cobalt
Anabolic/androgenic steroids
Haemochromatosis and other causes of iron overload
Endocrinology
Hypo- and hyperthyroidism
Cushing/Addison disease
Phaeochromocytoma
Acromegaly
Diabetes mellitus
Nutritional deficiency
Selenium deficiency
Thiamine deficiency (Beri-Beri)
Zinc and copper deficiency
Carnitine deficiency
Electrolyte disturbance
Hypocalcaemia
Hypophosphataemia

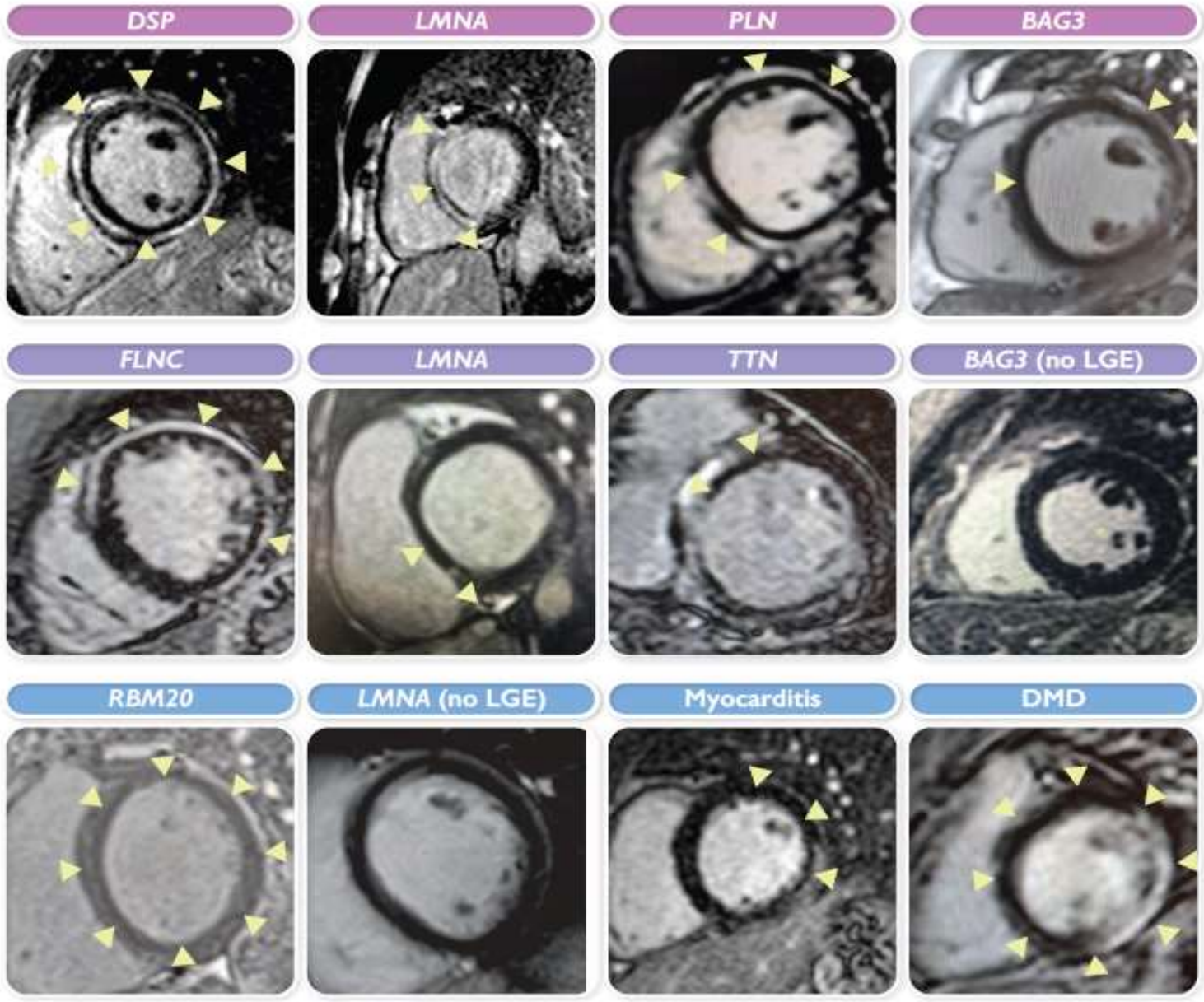
Peripartum	
Autoimmune diseases	
Giant cell myocarditis	
Inflammatory (biopsy-proven, non-infectious myocarditis)	
Eosinophilic granulomatosis with polyangiitis	
Systemic lupus erythematosus	
Sarcoidosis	
Rheumatoid arthritis	
Coeliac disease	
Primary biliary cirrhosis	
Myasthenia gravis	
Pemphigus pemphigoid	
Crohn disease	
Ulcerative colitis	
Polymyositis/dermatomyositis	
Reactive arthritis	
Drugs	
Antineoplastic drugs	Anthracyclines; antimetabolites; alkylating agents; Taxol; hypomethylating agent; monoclonal antibodies; tyrosine kinase inhibitors; immunomodulating agents
Psychiatric drugs	Clozapine, olanzapine; chlorpromazine, risperidone, lithium; methylphenidate; tricyclic antidepressants
Other drugs	All-trans retinoic acid; antiretroviral agents; phenothiazines

SCD Yönetimi

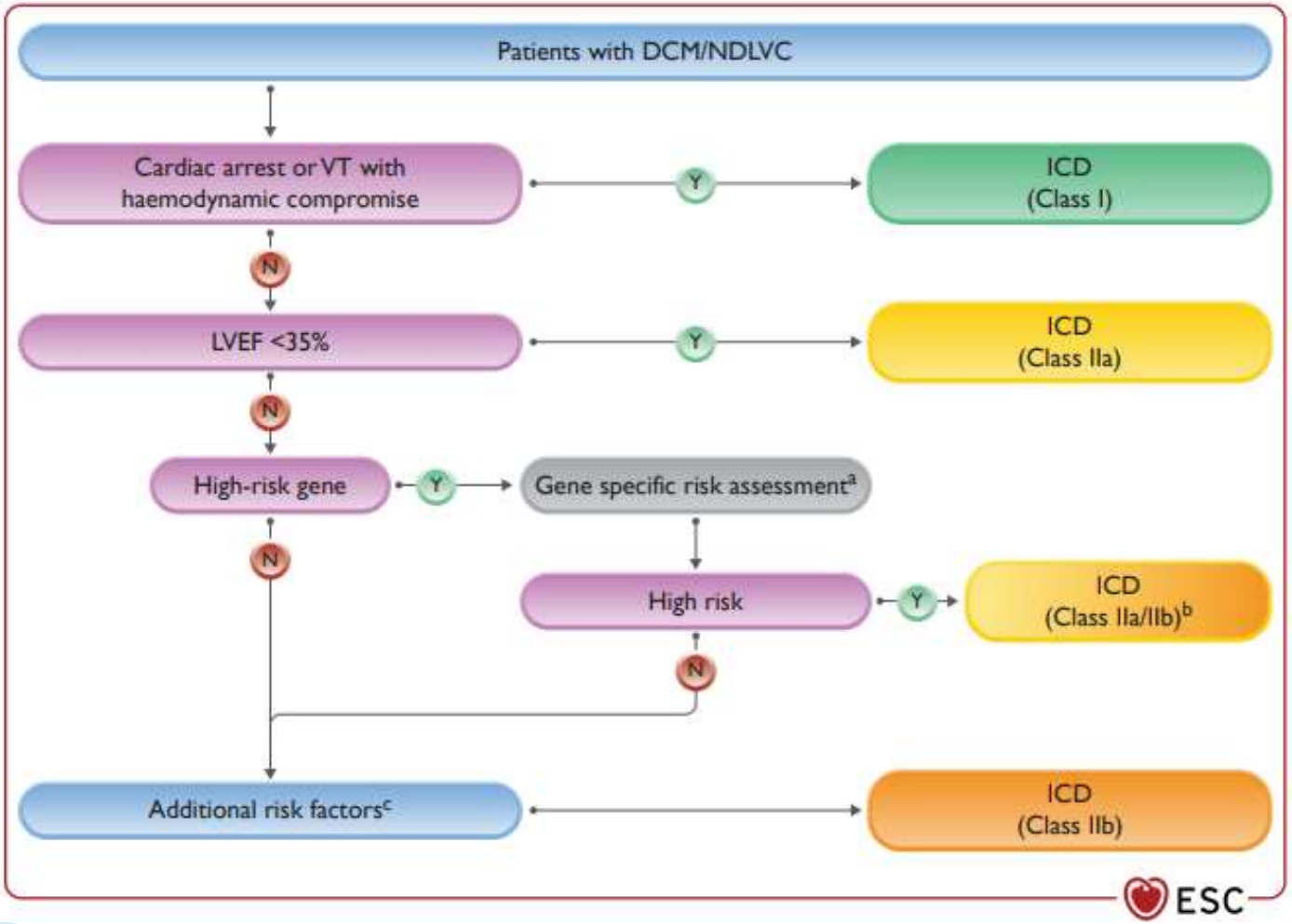


Non-dilated left ventricular cardiomyopathy

- Global veya bölgesel duvar hareketi anormallikleri olsun veya olmasın;
- LV dilatasyonu yokluğunda iskemik olmayan LV skarlaşması veya yağ infiltrasyonu
- Skarlaşma olmaksızın izole global LV hipokinezisi (LVEF<50) ile tanımlanır.
- Anormal yükleme koşulları (hipertansiyon, kapak hastalığı) veya KAH olmaması


DSP
LMNA
PLN
BAG3
FLNC
LMNA
TTN
BAG3 (no LGE)
RBM20
LMNA (no LGE)
Myocarditis
DMD

SCD Yönetimi

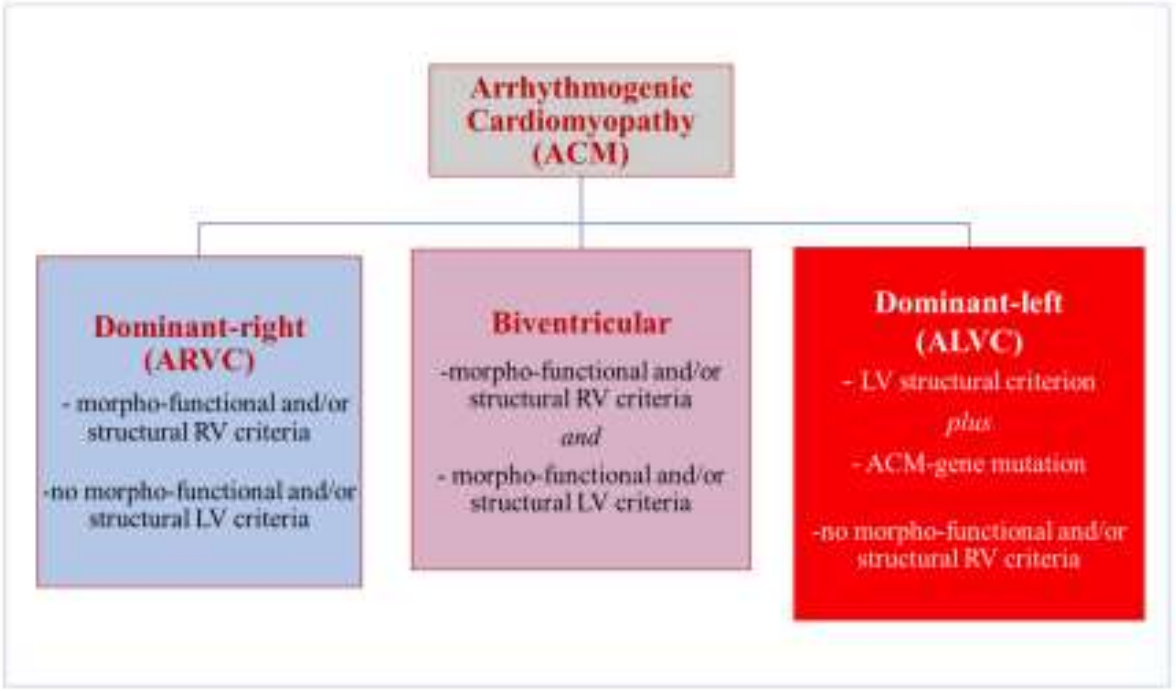


Arrhythmogenic right ventricular cardiomyopathy

- RV miyokatında progresif fibro-fatty değişim
- LV tutulum olabilir
- Baskın LV hastalığı aynı aile
- 20-40 yaş, erkek
- LBBB morfolojinde VE/VT
- V1-V3 T insersiyonu, düşük voltaj
- RV genişleme

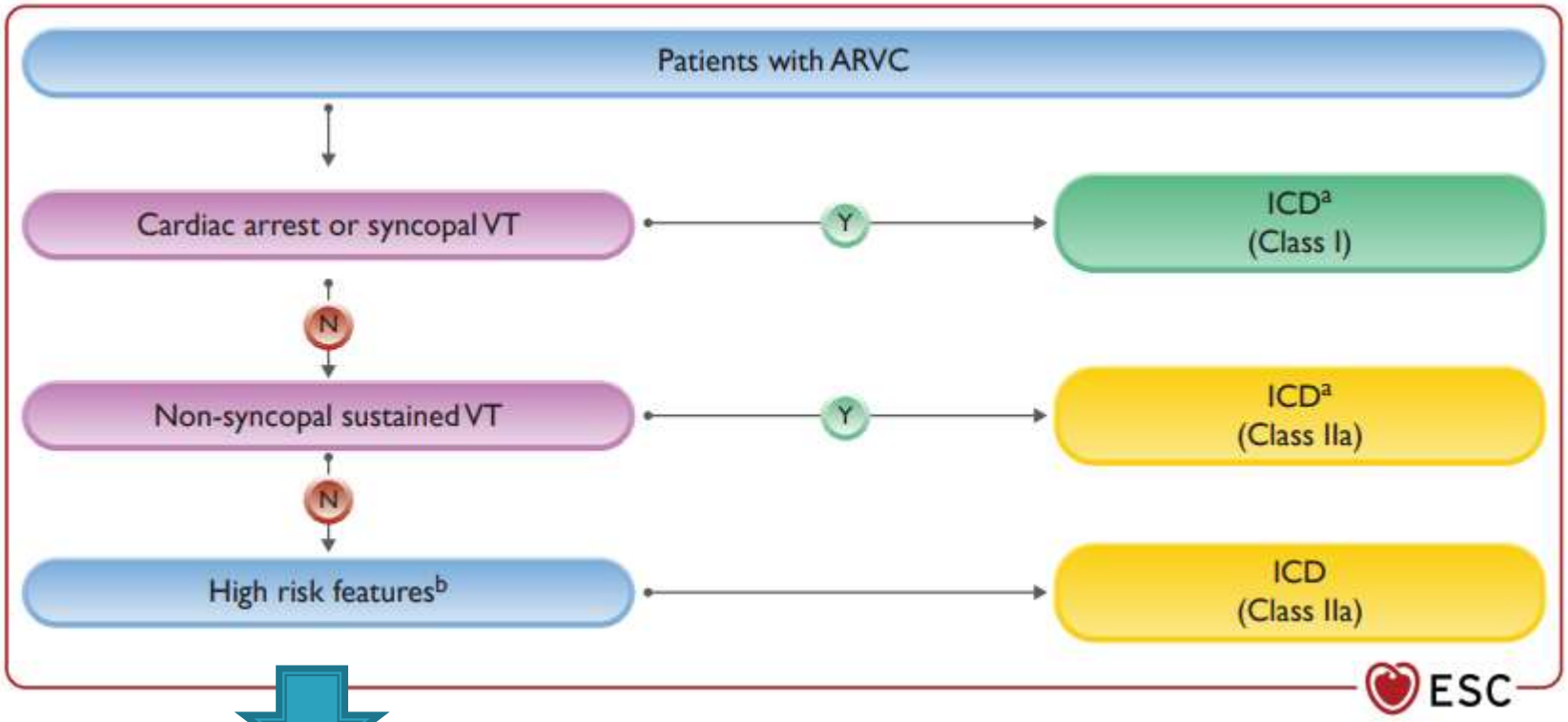
Table 1
"Padua criteria" for diagnosis of Arrhythmogenic Cardiomyopathy.

Category	Right ventricle (upgraded 2010 ITF diagnostic criteria)	Left ventricle (new diagnostic criteria)
I. Morpho-functional ventricular abnormalities	*By echocardiography, CMR or angiography:* *Major* • Regional RV akinesia, dyskinesia, or bulging *plus* one of the following: – global RV dilatation (increase of RV EDV according to the imaging test sp – gl in Mino • Re	*By echocardiography, CMR or angiography:Minor* • Global LV systolic dysfunction (depression of LV EF or reduction of echo-cardiographic global longitudinal strain), with or without LV dilatation (increase of LV EDV according to the imaging test specific nomograms for
II. Structural myocardial abnormalities	*By C* • Tr ≥1 *By E* • Fil fat	
III. Repolarization abnormalities	*Majo* • In in co Mino • In pu • In pu	
IV. Depolarization abnormalities	Mino • Ep co th • Te of ab	
V. Ventricular arrhythmias	*Major* • Frequent ventricular extrasystoles (>500 per 24 h), non-sustained or sustained ventricular tachycardia of LBBB morphology *Minor* • Frequent ventricular extrasystoles (>500 per 24 h), non-sustained or sustained ventricular tachycardia of LBBB morphology with inferior axis ("RVOT pattern")	*Minor* • Frequent ventricular extrasystoles (>500 per 24 h), non-sustained or sustained ventricular tachycardia with a RBBB morphology (excluding the "fascicular pattern")
VI. Family history/genetics	*Major* • ACM confirmed in a first-degree relative who meets diagnostic criteria • ACM confirmed pathologically at autopsy or surgery in a first degree relative • Identification of a pathogenic or likely pathogenetic ACM mutation in the patient under evaluation *Minor* • History of ACM in a first-degree relative in whom it is not possible or practical to determine whether the family member meets diagnostic criteria • Premature sudden death (<35 years of age) due to suspected ACM in a first-degree relative • ACM confirmed pathologically or by diagnostic criteria in a second-degree relative	

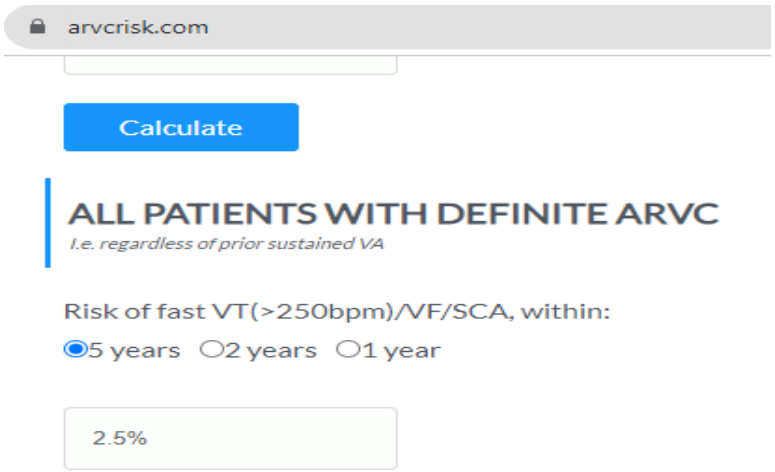


ARVC Tedavi

Recommendations	Class[a]	Level[b]
Beta-blocker therapy is recommended in ARVC patients with VE, NSVT, and VT.[920–922]	I	C
Amiodarone should be considered when regular beta-blocker therapy fails to control arrhythmia-related symptoms in patients with ARVC.[921,922]	IIa	C
Flecainide in addition to beta-blockers should be considered when single agent treatment has failed to control arrhythmia-related symptoms in patients with ARVC.[923,924]	IIa	C
Catheter ablation with availability for epicardial approach guided by 3D electroanatomical mapping of VT should be considered in ARVC patients with incessant VT or frequent appropriate ICD interventions for VT despite pharmacological therapy with beta-blockers.[925,929–934]	IIa	C



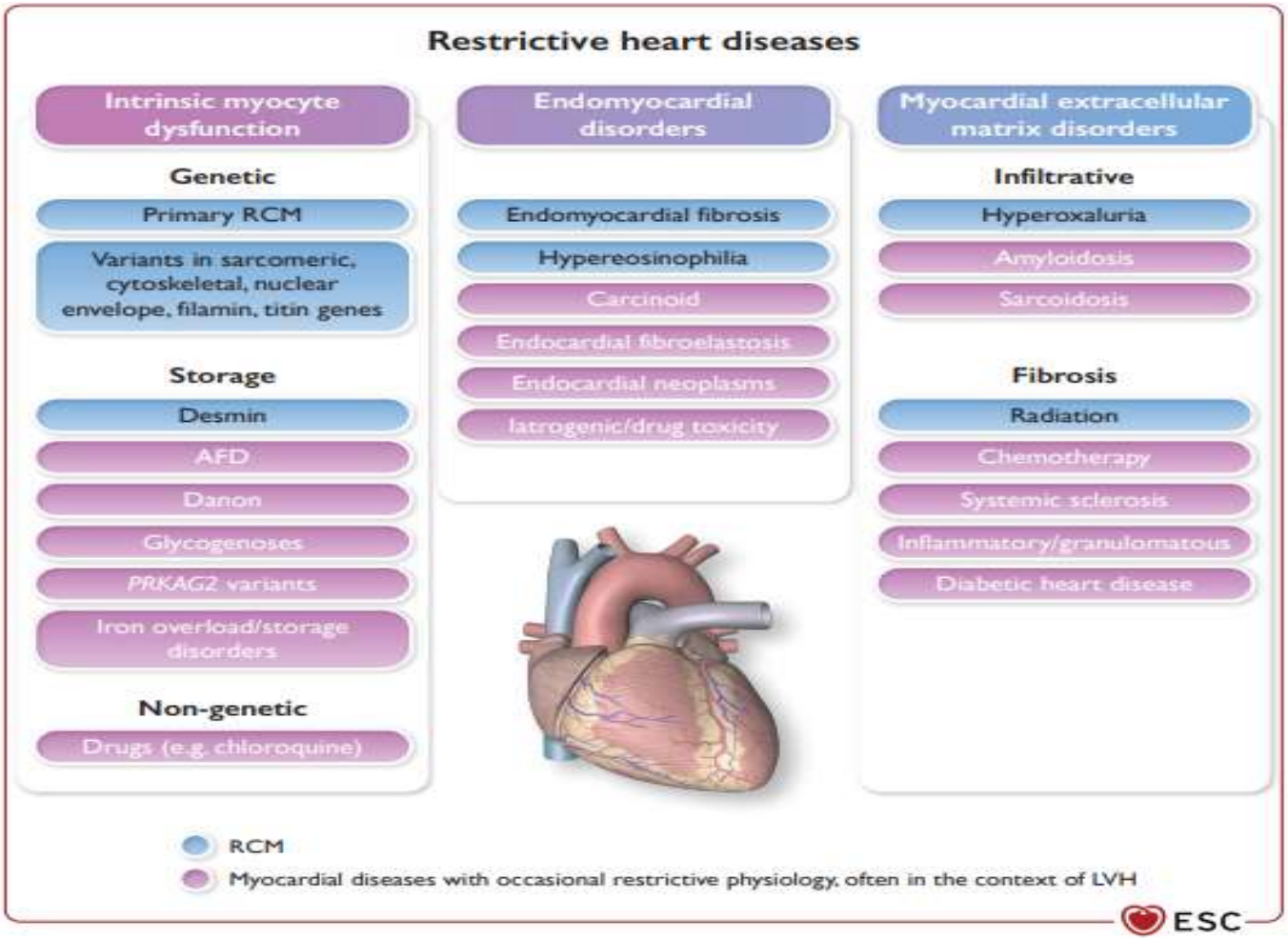
Aritmik Senkop
NSVT,
RVEF <40,
LVEF<45
EPS de Sustained VT
2019 ARVC risk calculator



Restriktif Kardiyomiyopati

- Normal veya azalmış diyastolik hacimler
- Normal veya azalmış sistolik hacimler
- Normal ventriküler duvar kalınlığının varlığında
- Kısıtlayıcı LV ve/veya RV patofizyolojisi
- Sıklıkla biatrial genişlemeyle prezente
- İleri aşamada venrikül dilate olabilir
- Hypertrophic' or 'dilated cardiomyopathy with restrictive physiology'
- Endokardiyal patoloji

Restriktif Kalp Hastalıkları



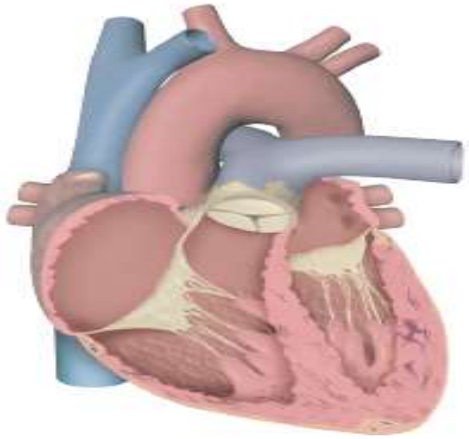
Öneriler

- En kötü prognozlu KMP
- Restriktif fizyolojiden ayırmak için multimodal görüntüleme
- Nöromuskuler sistem, metabolik bozukluklardan ayrım
- Çocuklarda tanı-takipte kataterizasyon
- Spesifik tanıları dışlamak için endomiyokardiyal bx (Iıa)
- Seonder korumada ICD
- Çocuklarda miyokard iskemisi ve senkop varsa ICD(IIb)

Sendromik ve Metabolik Kalp Hastalıkları

- Anderson-Fabry disease
- RASopathies
- Friedreich ataxia
- Glycogen storage disorders

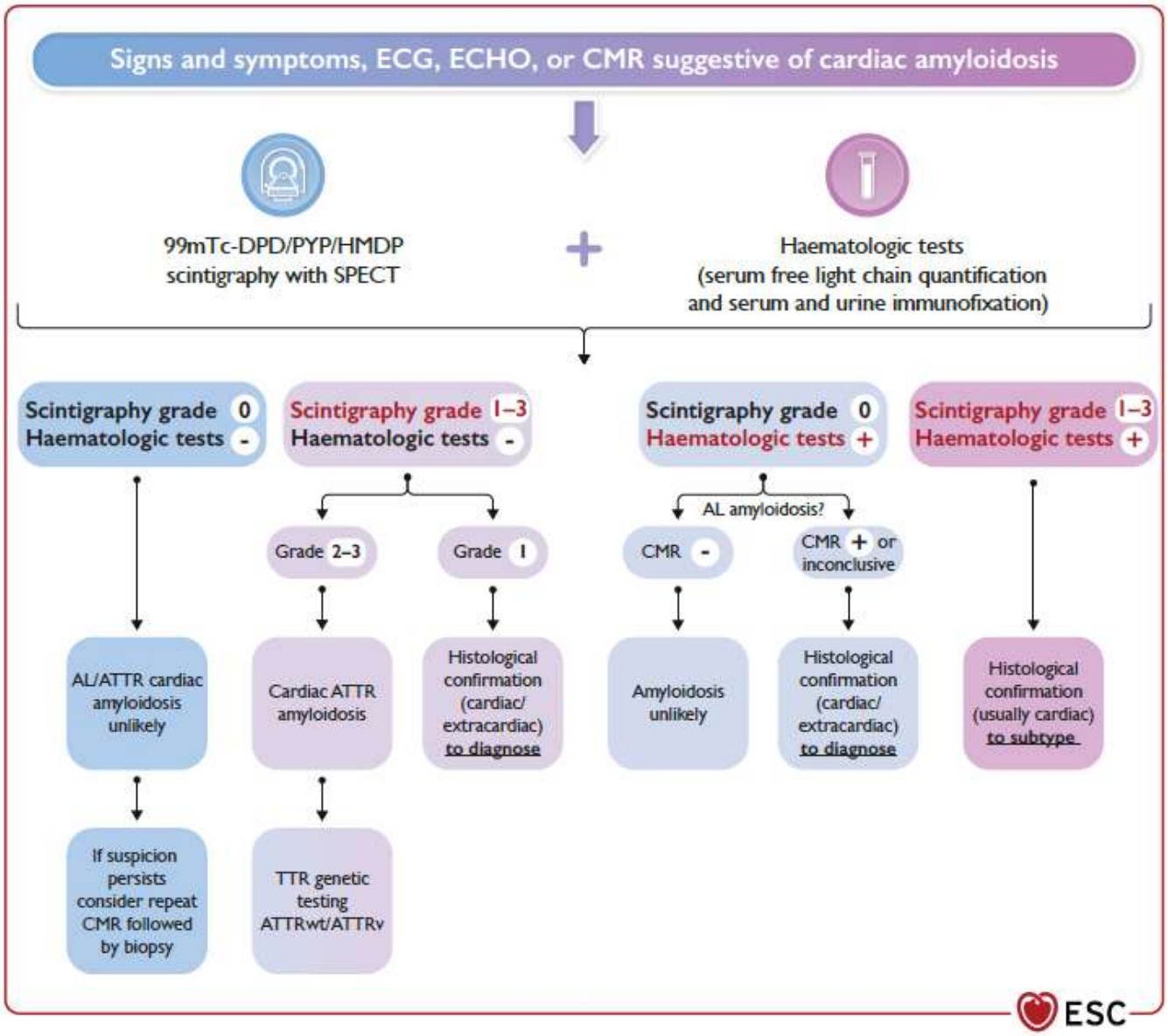
Amiloidosis



Left ventricular wall thickness ≥12 mm + ≥1 of

- Heart failure in ≥65 years
- Aortic stenosis in ≥65 years
- Hypotension or normotensive if previously hypertensive
- Sensory involvement, autonomic dysfunction
- Peripheral polyneuropathy
- Proteinuria
- Skin bruising
- Ruptured biceps tendon
- Bilateral carpal tunnel syndrome
- Subendocardial/transmural LGE or increased ECV
- Reduced longitudinal strain with apical sparing
- Decreased QRS voltage to mass ratio
- Pseudo Q waves on ECG
- AV conduction disease
- Possible family history of ATTR
- Chronically increased troponin levels
- Known multiple myeloma or MGUS

ESC


Signs and symptoms, ECG, ECHO, or CMR suggestive of cardiac amyloidosis
99mTc-DPD/PYP/HMDP scintigraphy with SPECT
+
Haematologic tests (serum free light chain quantification and serum and urine immunofixation)
Scintigraphy grade 0 Haematologic tests -
Scintigraphy grade 1–3 Haematologic tests -
Scintigraphy grade 0 Haematologic tests +
Scintigraphy grade 1–3 Haematologic tests +
AL amyloidosis?
Grade 2–3
Grade 1
CMR -
CMR + or inconclusive
AL/ATTR cardiac amyloidosis unlikely
Cardiac ATTR amyloidosis
Histological confirmation (cardiac/ extracardiac) to diagnose
Amyloidosis unlikely
Histological confirmation (cardiac/ extracardiac) to diagnose
Histological confirmation (usually cardiac) to subtype
If suspicion persists consider repeat CMR followed by biopsy
TTR genetic testing ATTRwt/ATTRv
ESC

Atrial Fibrilasyon

Recommendations	Class[a]	Level[b]
Anticoagulation		
Oral anticoagulation in order to reduce the risk of stroke and thrombo-embolic events is recommended in all patients with HCM or cardiac amyloidosis and AF or atrial flutter (unless contraindicated).[332,365,369,371,373,378,413,427,428,456–464]	I	B
Oral anticoagulation to reduce the risk of stroke and thrombo-embolic events is recommended in patients with DCM, NDLVC, or ARVC, and AF or atrial flutter with a CHA_2DS_2-VASc score ≥2 in men or ≥3 in women.[465–469]	I	B
Control of symptoms and heart failure		
Atrial fibrillation catheter ablation is recommended for rhythm control after one failed or intolerant class I or III AAD to improve symptoms of AF recurrences in patients with paroxysmal or persistent AF and cardiomyopathy.[335,397–399,412,415–420,430–435,447,451,473–498]	I	B
Atrial fibrillation catheter ablation is recommended to reverse LV dysfunction in AF patients with cardiomyopathy when tachycardia-induced component is highly probable, independent of their symptom status.[405,407,408,499–501]	I	B

Kardiyomiyopatilerde Egzersiz

Kardiyomiyopati hastalarına egzersiz önerileri		
Tüm kardiyomiyopatilerde		
Kardiyomiyopatisi olan tüm sağlıklı bireylere düzenli olarak düşük ile orta yoğunlukta egzersiz önerilir.	I	C
Kardiyomiyopatili tüm hastalarda egzersiz reçetesi için kişiselleştirilmiş bir risk değerlendirmesi önerilmektedir.	I	C
HCM		
Yarışmalı sporlarda dahil olmak üzere yüksek yoğunluklu egzersiz; yüksek riskli bireylerde ve sol ventriküler çıkış yolu obstrüksiyonu ve egzersize bağlı kompleks ventriküler aritmileri olan bireylerde önerilmez.	III	C
ARVC		
ARVC'li bireylerde yarışmalı spor da dahil olmak üzere orta ve/veya yüksek yoğunlukta egzersiz önerilmemektedir.	III	B
DCM and NDLVC		
Yarışmalı spor da dahil olmak üzere yüksek yoğunluklu egzersiz; semptomatik bireylerde, sol ventriküler ejeksiyon fraksiyonu ≤%40 olanlarda, egzersize bağlı aritmi görülenlerde veya LMNA veya TMEM43 patojenik varyantına sahip kişilerde	III	C

Reptodüktif Konular

Tüm kadınlarda mWHO anne risk sınıflandırması kullanılarak gebelik öncesi risk değerlendirmesi ve danışmanlık yapılması önerilmektedir.	I	C
Doğurganlık çağındaki tüm kadınlara ve partnerlerine güvenli ve etkili doğum kontrolü konusunda danışmanlık verilmesi tavsiye edilir.	I	C
Gebe kalmadan önce tüm kadın ve erkeklere hastalık kalıtım riski konusunda danışmanlık verilmesi önerilir.	I	C
Sezaryen için obstetrik endikasyonlar, şiddetli kalp yetersizliği (EF <%30 veya NYHA sınıf III-IV) veya şiddetli çıkış yolu obstruksiyonu veya doğum sırasında oral antikoagülan kullanan kadınlar hariç, kardiyomiyopatili kadınların bir çoğunda vajinal doğum önerilir.	I	C
İlaçların hamilelikten önce güvenlik açısından dikkatlice gözden geçirilmesi ve hamilelikteki tolere edilebilirliğe göre ayarlanması önerilir.	I	C
AF'li hastalarda gebelik evresine göre DMAH veya VKA'larla terapötik antikoagülasyon önerilmektedir.	I	C

Kardiyomiyopatiler

- Düşünülenden daha yaygındır
- İsimlendirmede başvuru sırasındaki baskın fenotip
- Fenotip için multimodal görüntüleme
- Multidisipliner yaklaşım
- Genetik testler kullanılmalı
- KMR, karakterizasyon, progresyon,risk değerlendirme
- ATTR amiloidozda kemik sintigrafisi

TEŞEKKÜRLER...