Endorsement	Endorsed	Endorsed med forbehold	Not endorsed	
				Initialer samt endorsement
Cardiomyopathy guidelines tables of recommendations	Class	Evidence level	Ja - endorsement	Nej - ikke endorsement
Recommendation Table 1, Recommendations for the provision of service of multidisciplinary cardiomyopathy teams, side 17	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	Ja	
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	Ja	
Recommendation Table 2, Recommendations for diagnostic work-up in cardiomyopathies, side 18	Class	Level		
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	I	C	Bør tilpasses den enkelte patient.	
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.	I	C	Ja	
Recommendation Table 3 Recommendations for laboratory tests in the diagnosis of cardiomyopathies, side 22	Class	Level		
Routine (first-level) laboratory testsc are recommended in all patients with suspected or confirmed cardiomyopathy to evaluate aetiology, assess disease severity, and aid in detection of extracardiac manifestations and assessment of secondary organ dysfunction.	I	C	Bør tilpasses den enkelte patient.	
Additional (second-level) testsc should be considered in patients with cardiomyopathy and extracardiac features to aid in detection of metabolic and syndromic causes, following specialist evaluation.	IIa	C	Ja	
Recommendation Table 4 Recommendation for echocardiographic evaluation in patients with cardiomyopathy, side 22	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	Ja	

Recommendation Table 5 Recommendations for cardiac magnetic resonance indication in patients with cardiomyopathy, side 24	Class	Level		
Contrast-enhanced CMR is recommended in patients with cardiomyopathy at initial evaluation	I	C	Bør tilpasses den enkelte patient.	
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	Gentagen MR scanning anbefales ikke rutinemæssigt, men kan overvejes i særlig	
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	B	Bør tilpasses den enkelte patient.	
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	C	Bør tilpasses den enkelte patient.	
In cases of familial cardiomyopathy without a geneticdiagnosis, contrast-enhanced CMR may beconsidered in phenotype-negative family members to	IIb	C		Endorses ikke
Recommendation Table 6 Recommendations for computed tomography and nuclear imaging side 26	**Class**	**Level**		
DPD/PYP/HMDP bone-tracer scintigraphy is recommended in patients with suspected ATTR-related cardiac amyloidosis to aid diagnosis	I	B	Ja	
Contrast-enhanced cardiac CT should be considered in patients with suspected cardiomyopathy who have inadequate echocardiographic imaging and contraindications to CMR	IIa	C	Ja	
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	Bør tilpasses den enkelte patient.	
18F-FDG-PET scanning should be considered for the diagnostic work-up in patients with cardiomyopathy in whom cardiac sarcoidosis is suspected	IIa	C	Ja	
Recommendation Table 7 — Recommendation for endomyocardial biopsy in patients with cardiomyopathy, side 26	**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	Ja	
Recommendation Table 8 — Recommendations for genetic counselling and testing in cardiomyopathies	**Class**	**Level**		

Genetic counselling, provided by an appropriately trained healthcare professional and including genetic education to inform decision-making and psychosocial support, is recommended for families with an inherited or suspected inherited cardiomyopathy, regardless of whether genetic testing is being considered.	I	B	Ja	
It is recommended that genetic testing for cardiomyopathy is performed with access to a multidisciplinary team, including those with expertise in genetic testing methodology, sequence variant interpretation, and clinical application of genetic testing, typically in a specialized cardiomyopathy service or in a network model with access to equivalent expertise.	I	B	Ja	
Pre- and post-test genetic counselling is recommended in all individuals undergoing genetic testing for cardiomyopathy.	I	B	Ja	
If pre-natal diagnostic testing is to be pursued by the family, it is recommended that this is performed early in pregnancy, to allow decisions regarding continuation or co-ordination of pregnancy to be made.	I	C	Ja	
A discussion about reproductive genetic testing options with an appropriately trained healthcare professional should be considered for all families with a genetic diagnosis.	IIa	C	Ja	
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	Ja	
Genetic testing is recommended for a deceased individual identified to have cardiomyopathy at post-mortem if a genetic diagnosis would facilitate management of surviving relatives	I	C	Ja	
Genetic testing may be considered in patients fulfilling diagnostic criteria for cardiomyopathy when it will have a net benefit to the patient, considering the psychological impact and preference, even if it does not enable diagnosis, prognostication, or therapeutic stratification, or cascade genetic screening of their relatives.	IIb	C		Nej
Genetic testing in patients with a borderline phenotype not fulfilling diagnostic criteria for a cardiomyopathy may be considered only after detailed assessment by specialist teams.	IIb	C	Ja	
FAMILY MEMBERS				

It is recommended that cascade genetic testing, with pre- and post-test counselling, is offered to adult at-risk relatives if a confident genetic diagnosis (i.e. a P/LP variant) has been established in an individual with cardiomyopathy in the family (starting with first-degree relatives if available, and cascading out sequentially)	I	B	Ja
Cascade genetic testing with pre- and post-test counselling should be considered in paediatric at-risk relatives if a confident genetic diagnosis (i.e. a P/LP variant) has been established in an individual with cardiomyopathy in the family (starting with first-degree relatives, if available, and cascading out sequentially), considering the underlying cardiomyopathy, expected age of onset, presentation in the family, and clinical/legal consequences.	IIa	B	Ja
Testing for the presence of a familial variant of unknown significance, typically in parents and/or affected relatives, to determine if the variant segregates with the cardiomyopathy phenotype should be considered if this might allow the variant to be interpreted with confidence.	IIa	C	Ja
Diagnostic genetic testing is not recommended in a phenotype-negative relative of a patient with cardiomyopathy in the absence of a confident genetic diagnosis (i.e. a P/LP variant) in the family.	III	C	Ja

Recommendation Table 9 — Recommendations for cardiac transplantation in patients with cardiomyopathy side 38	Class	Level	
Orthotopic cardiac transplantation is recommended for eligible cardiomyopathy patients with advanced heart failure (NYHA class III–IV) or intractable ventricular arrhythmia refractory to medical/invasive/ device therapy, and who do not have absolute contraindications	I	C	Ja

Recommendation Table 10 — Recommendation for left ventricular assist device therapy in patients with cardiomyopathy, side 38	Class	Level	
Mechanical circulatory support therapy should be considered in selected cardiomyopathy patients with advanced heart failure (NYHA class III–IV) despite optimal pharmacological and device treatment, who are otherwise suitable for heart transplantation, to improve symptoms and reduce the risk of heart failure hospitalization from worsening heart failure and premature death while awaiting a transplant.	IIa	B	Ja

Mechanical circulatory support therapy should be considered in selected cardiomyopathy patients with advanced heart failure (NYHA class III–IV) despite optimal pharmacological and device therapy, who are not eligible for cardiac transplantation or other surgical options, and without severe right ventricular dysfunction, to reduce the risk of death and improve symptoms.	IIa	B	Ja

Recommendation Table 11 — Recommendations for management of atrial fibrillation and atrial flutter in patients with cardiomyopathy side 41	Class	Level	
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).	I	B	Ja
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 CHA2DS2-VASc score ≥2 in men or ≥3 in women	I	B	Ja
Oral anticoagulation to reduce the risk of stroke and thrombo-embolic events should be considered in patients with RCM and AF or atrial flutter (unless contraindicated).	IIa	C	Ja
Oral anticoagulation to reduce the risk of stroke and thrombo-embolic events should be considered in patients with DCM, NDLVC, or ARVC, and AF or atrial flutter with a CHA2DS2-VASc score of 1 in men or of 2 in women	IIa	B	Ja
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.	I	B	Ja
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.	I	B	Ja, overvej dog DanAblate Studiet
Maintenance of sinus rhythm rather than rate control should be considered at an early stage for patients with a cardiomyopathy and AF without major risk factors for recurrence, regardless of symptoms.	IIa	C	Ja, overvej dog DanAblate Studiet
Atrial fibrillation catheter ablation should be considered as first-line rhythm control therapy to improve symptoms in selected patients with cardiomyopathy and paroxysmal or persistent AF without major risk factors for recurrences as an alternative to class I or III AADs, considering patient choice, benefit, and risk.	IIa	C	Ja

Atrial fibrillation catheter ablation should be considered in selected patients with cardiomyopathy, AF, and heart failure and/or reduced LVEF to prevent AF recurrences and improve QoL, LVEF, and survival and reduce heart failure hospitalization.	IIa	B	Ja
Comorbidities and associated risk factor management			
Modification of unhealthy lifestyle and targeted therapy of intercurrent conditions is recommended to reduce AF burden and symptom severity in patients with cardiomyopathy	I	B	Ja

Recommendation Table 12 — Recommendations for implantable cardioverter defibrillator in patients with cardiomyopathy side 43	Class	Level	
General recommendations			
Implantation of a cardioverter defibrillator is only recommended in patients who have an expectation of good quality survival >1 year.	I	C	Ja
It is recommended that ICD implantation be guided by shared decision-making that: • is evidence-based; • considers a person's individual preferences, beliefs, circumstances, and values; and • ensures that the person understands the benefits, harms, and possible consequences of different treatment options.	I	C	Ja
It is recommended that prior to ICD implantation, patients are counselled on the risk of inappropriate shocks, implant complications, and the social, occupational, and driving implications of the device.	I	C	Ja
It is not recommended to implant an ICD in patients with incessant ventricular arrhythmias until the ventricular arrhythmia is controlled.	III	C	Ja
Secondary prevention			
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	Ja
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	Ja
ICD implantation should be considered in patients with cardiomyopathy presenting with haemodynamically tolerated VT, in the absence of reversible causes	IIa	C	Ja
Primary prevention			

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	Tilpasses den enkelte patient		
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	Ja		
should be considered in patients with DCM, NDLVC, and ARVC.	IIa	B	Ja		
If a patient with cardiomyopathy requires pacemaker implantation, comprehensive SCD risk stratification to evaluate the need for ICD implantation should be considered.	IIa	C	Ja		
Choice of ICD					
When an ICD is indicated, it is recommended to evaluate whether the patient could benefit from CRT	I	A	Ja		
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	Ja		
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	B		Nej, bruges ikke i DK	
Recommendation Table 13 — Recommendations for routine follow-up of patients with cardiomyopathy side 44	Class	Level			
It is recommended that all clinically stable patients with cardiomyopathy undergo routine follow-up using a multiparametric approach that includes ECG and echocardiography every 1 to 2 years.	I	C	Tilpasses patient/sygdom		
Clinical evaluation with ECG and multimodality imaging is recommended in patients with cardiomyopathy whenever there is a substantial or unexpected change in symptoms.	I	C	Ja		
Recommendation Table 14 — Recommendations for family screening and follow-up evaluation of relatives side 44	Class	Level			
Following cascade genetic testing, clinical evaluation using a multiparametric approach that includes ECG and cardiac imaging and long-term follow-up is recommended in first-degree relatives who have the same disease-causing variant as the proband	I	B	Der anbefales standard klinisk undersøgelse samtidigt med genetisk udredning		
Following cascade genetic testing, it is recommended that first-degree relatives without a phenotype who do not have the same disease-causing variant as the proband are discharged from further follow-up but advised to seek re-assessment if they develop symptoms or when new clinically relevant data emerge in the family.	I	C	Ja		

It is recommended that when no P/LP variant is identified in the proband or genetic testing is not performed, an initial clinical evaluation using a multiparametric approach that includes ECG and cardiac imaging is performed in first-degree relatives.	I	C	Uden påvist familiær sygdom og blank genetik da individuel stillingtagen til familieus
When no P/LP variant is identified in the proband or genetic testing is not performed, regular, long-term clinical evaluation using a multiparametric approach that includes ECG and cardiac imaging should be considered in first-degree relatives.	IIa	C	Individuel stillingtagen, mange af disse slægtninge vil kunne afsluttes
During cascade screening, where a first-degree relative has died, clinical evaluation of close relatives of the deceased individual (i.e. second-degree relatives of the index patient) should be considered.	IIa	C	Ja
Recommendation Table 15 — Recommendations for psychological support in patients and family members with cardiomyopathies	**Class**	**Level**	
It is recommended that psychological support by an appropriately trained health professional be offered to all individuals who have experienced the premature sudden cardiac death of a family member with cardiomyopathy.	I	B	Ja, afhængig af lokale tilbud
It is recommended that psychological support by an appropriately trained health professional be offered to all individuals with an inherited cardiomyopathy who receive an implantable cardioverter defibrillator	I	B	Ja, afhængig af lokale tilbud
Psychological support by an appropriately trained health professional should be considered in all patients and families with an inherited cardiomyopathy and in particular for those issues described in the text	IIa	C	Ja, afhængig af lokale tilbud
Recommendation Table 16 — Recommendation for evaluation of left ventricular outflow tract obstruction	**Class**	**Level**	
In all patients with HCM, at initial evaluation, transthoracic 2D and Doppler echocardiography are recommended, at rest and during Valsalva manoeuvre in the sitting and semi-supine positions—and then on standing if no gradient is provoked—to detect LVOTO.	I	B	Ja, der udføres dog liggende undersøgelse som standard
In symptomatic patients with HCM and a resting or provokedc peak instantaneous LV outflow tract gradient <50 mmHg, 2D and Doppler echocardiography during exercise in the standing, sitting (when possible), or semi-supine position are recommended to detect provocable LVOTO and exercise-induced mitral regurgitation	I	B	Her anbefales arbejdsekkokardiografi

Transoesophageal echocardiography should be considered in patients with HCM and LVOTO if the mechanism of obstruction is unclear or when assessing the mitral valve apparatus before a septal reduction procedure, or when severe mitral regurgitation caused by intrinsic valve abnormalities is suspected	IIa	C	Ja
In symptomatic patients with HCM and inconclusive non-invasive cardiac imaging, left and right heart catheterization may be considered to assess the severity of LVOTO and to measure LV filling pressures.	IIb	C	Ja

Recommendation Table 17 — Additional recommendation for cardiovascular magnetic resonance evaluation in hypertrophic cardiomyopathy side 48	Class	Level	
Contrast-enhanced CMR may be considered before ASA or myectomy to assess the extent and distribution of hypertrophy and myocardial fibrosis	IIb	C	Ja

Recommendation Table 18 — Recommendations for treatment of left ventricular outflow tract obstruction (general measures) side 51	Class		
Avoidance of digoxin and arterial and venous dilators, including nitrates and phosphodiesterase inhibitors, should be considered, if possible, in patients with resting or provocable LVOTO.	IIa	C	Ja
Restoration of sinus rhythm or appropriate rate control should be considered before invasive management of LVOTO in patients with new-onset or poorly controlled AF	IIa	C	Ja

Recommendation Table 19 — Recommendations for medical treatment of left ventricular outflow tract obstruction	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 provokedc LVOTO	I	B	Ja
Verapamil or diltiazem, titrated to maximum tolerated dose, are recommended to improve symptoms in symptomatic patients with resting or provokedc LVOTO who are intolerant or have contraindications to beta-blockers.	I	B	Ja
Disopyramide,d 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 provokedc LVOTO	I	B	Nej, disopyramid ikke tilgængeligt i DK

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 provokedc LVOTO	IIa	A	Mavacamten har dokumenteret effekt, men plads i behandlingshierakiet uafklaret. Se generel tekst.
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 provokedc LVOTO (exercise or Valsalva manoeuvre) who are intolerant or have contraindications to beta-blockers, verapamil/ diltiazem, or disopyramide	IIa	B	Mavacamten har dokumenteret effekt, men plads i behandlingshierakiet uafklaret. Se generel tekst.
Oral or i.v. beta-blockers and vasoconstrictors should be considered in patients with severe provocablec LVOTO presenting with hypotension and acute pulmonary oedema who do not respond to fluid administration	IIa	C	Ja
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 provokedc LVOTO.	IIb	C	Disopyramid ikke tilgængeligt i DK
Beta-blockers or verapamil may be considered in selected cases in asymptomatic patients with resting or provokedc LVOTO to reduce LV pressures	IIb	C	Ja
The cautious use of low-dose diuretics may be considered in symptomatic LVOTO to improve exertional dyspnoea.	IIb	C	Ja

Recommendation Table 20 — Recommendations for septal reduction therapy	Class	Level	
It is recommended that SRT be performed by experienced operators working as part of a multidisciplinary team expert in the management of HCM	I	C	Ja
SRT to improve symptoms is recommended in patients with a resting or maximum provoked LVOT gradient of ≥50 mmHg who are in NYHA/Ross functional class III–IV, despite maximum tolerated medical therapy	I	B	Ja
Septal myectomy, rather than ASA, is recommended in children with an indication for SRT, as well as in adult patients with an indication for SRT and other lesions requiring surgical intervention (e.g. mitral valve abnormalities).	I	C	Ja
SRT should be considered in patients with recurrent exertional syncope caused by a resting or maximum provoked LVOTO gradient ≥50 mmHg despite optimal medical therapy	IIa	C	Ja

Mitral valve repair or replacement should be considered in symptomatic patients with a resting or maximum provoked LVOTO gradient ≥50 mmHg and moderate-to-severe mitral regurgitation that cannot be corrected by SRT alone	IIa	C	Ja
Mitral valve repair should be considered in patients with a resting or maximum provoked LVOTO gradient ≥50 mmHg when there is moderate-to-severe mitral regurgitation following isolated myectomy.	IIa	C	Ja
SRT may be considered in expert centres with demonstrable low procedural complication rates in patients with mild symptoms (NYHA class II) refractory to medical therapy who have a resting or maximum provoked (exercise or Valsalva) gradient of ≥50 mmHg and: • moderate-to-severe SAM-related mitral regurgitation; or • AF; or • moderate-to-severe left atrial dilatation	IIb	C	Ja
Mitral valve replacement may be considered in patients with a resting or maximum provoked LVOTO gradient ≥50 mmHg when there is moderate-to-severe mitral regurgitation following isolated myectomy	IIb	C	Ja
Surgical AF ablation and/or left atrial appendage occlusion procedures during septal myectomy may be considered in patients with HCM and symptomatic AF	IIb	C	Ja

Recommendation Table 21 — Recommendations for indications for cardiac pacing in patients with obstruction side 55	Class	Level	
Sequential AV pacing, with optimal AV interval to reduce the LV outflow tract gradient or to facilitate medical treatment with beta-blockers and/or verapamil, may be considered in selected patients with resting or provocable LVOTO ≥50 mmHg, sinus rhythm, and drug-refractory symptoms, who have contraindications for ASA or septal myectomy or are at high risk of developing heart block following ASA or septal myectomy.	IIb	C	Ja, omend pace-indikation LVOTO ikke bruges selvstændigt i DK mere.
In patients with resting or provocable LVOTO ≥50 mmHg, sinus rhythm, and drug-refractory symptoms, in whom there is an indication for an ICD, a dual-chamber ICD (instead of a single-lead device) may be considered, to reduce the LV outflow tract gradient or to facilitate medical treatment with beta-blockers and/or verapamil	IIb	C	Ja

Recommendation Table 22 — Recommendations for chest pain on exertion in patients without left ventricular outflow tract obstruction side 55	Class	Level

Beta-blockers and calcium antagonists (verapamil or diltiazem) should be considered to improve symptoms in patients with angina-like chest pain even in the absence of LVOTO or obstructive CAD	IIa	C	Ja	
Oral nitrates may be considered to improve symptoms in patients with angina-like chest pain, even in the absence of obstructive CAD, if there is no LVOTO.	IIb	C	Ja	
Ranolazine may be considered to improve symptoms in patients with angina-like chest pain even in the absence of LVOTO or obstructive CAD	IIb	C		Ikke tilgængeligt i DK
Recommendation Table 23 — Additional recommendations for prevention of sudden cardiac death in patients with hypertrophic cardiomyopathy	**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	Ja	
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	Ja	
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	Ja	
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	Ja, hvis sandsynlig klinisk konsekvens afhængig af alder, komorbiditet, mm	
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 (iii) the impact of an ICD on lifestyle, socio-economic status, and psychological health	IIa	B	Ja	
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	Ja	

Implantation of an ICD may be considered in individual patients with an estimated 5-year risk of SCD of between ≥4% and <6%, following detailed clinical assessment that takes into account the lifelong risk of complications and the impact of an ICD on lifestyle, socio-economic status, and psychological health	IIb	B	Ja	
For patients who are in the low-risk category (<4% estimated 5-year risk of SCD), the presence of extensive LGE (≥15%) on CMR may be considered in shared decision-making with patients about prophylactic ICD implantation, acknowledging the lack of robust data on the impact of scar quantification on the personalized risk estimates generated by HCM Risk-SCD or a validated paediatric model (e.g. HCM Risk-Kids)	IIb	B		Nej
For patients who are in the low-risk category (<4% estimated 5-year risk of SCD), the presence of LVEF <50% may be considered in shared decision-making with patients about prophylactic ICD implantation, acknowledging the lack of robust data on the impact of systolic dysfunction on the personalized risk estimates generated by HCM Risk-SCD or a validated paediatric model (e.g. HCM Risk-Kids).	IIb	B		Nej

Recommendation Table 24 — Recommendations for an implantable cardioverter defibrillator in patients with dilated cardiomyopathy side 64	Class	Level	
SECONDARY PREVENTION			
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	Ja
PRIMARY PREVENTION			
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	Ja
The patient's genotype should be considered in the estimation of SCD risk in DCM	IIa	B	Ja
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 (see Table 21).	IIa	C	Ja
An ICD may be considered in selected patients with DCM with a genotype associated with high SCD risk and LVEF >35% without additional risk factors (see Table 21).	IIb	C	Ja
An ICD may be considered in patients with DCM without a genotype associated with high SCD risk and LVEF >35% in the presence of additional risk factors	IIb	C	Ja

Recommendation Table 25 — Recommendation for resting and ambulatory electrocardiogram monitoring in patients with non-dilated left ventricular cardiomyopathy side 65	Class	Level	
Ambulatory ECG monitoring is recommended in patients with NDLVC annually or when there is a change in clinical status, to aid in management and risk stratification.	I	C	Individuel vurdering. Der foreligger utilstrækkelige data for NDLVC fænotypen

Recommendation Table 26 — Recommendations for an implantable cardioverter defibrillator in patients with non-dilated left ventricular cardiomyopathy side 66	Class	Level	
SECONDARY PREVENTION			
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	Ja
PRIMARY PREVENTION			
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	Ja
The patient's genotype should be considered in the estimation of SCD risk in NDLVC.	IIa	C	Ja
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 (see Table 21).	IIa	C	Individuel vurdering. Der foreligger utilstrækkelige data for NDLVC fænotypen
An ICD may be considered in selected patients with NDLVC with a genotype associated with high SCD risk and LVEF >35% without additional risk factors (see Table 21).	IIb	C	Individuel vurdering. Der foreligger utilstrækkelige data for NDLVC fænotypen
An ICD may be considered in patients with NDLVC without a genotype associated with high SCD risk and LVEF >35% in the presence of additional risk factors.c	IIb	C	Individuel vurdering. Der foreligger utilstrækkelige data for NDLVC fænotypen

Recommendation Table 27 — Recommendation for resting and ambulatory electrocardiogram monitoring in patients with arrhythmogenic right ventricular cardiomyopathy side 67	Class	Level	
Annual ambulatory ECG monitoring is recommended in patients with ARVC to aid in diagnosis, management, and risk stratification.	I	C	Ja

Recommendation Table 28 — Recommendations for the antiarrhythmic management of patients with arrhythmogenic right ventricular cardiomyopathy side 68	Class	Level	
Beta-blocker therapy is recommended in ARVC patients with VE, NSVT, and VT.	I	C	Ja, inklusiv sotalol
Amiodarone should be considered when regular beta-blocker therapy fails to control arrhythmia-related symptoms in patients with ARVC.	IIa	C	Overvej sotalol før amiodaron

Flecainide in addition to beta-blockers should be considered when single agent treatment has failed to control arrhythmia-related symptoms in patients with ARVC.	IIa	C	Ja
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	IIa	C	Ja
Recommendation Table 29 — Recommendations for sudden cardiac death prevention in patients with arrhythmogenic right ventricular cardiomyopathy	**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	Ja
An ICD should be considered in ARVC patients who have suffered a haemodynamically tolerated VT.	IIa	B	Ja
PRIMARY PREVENTION			
High-risk featuresc should be considered to aid individualized decision-making for ICD implantation in patients with ARVC.	IIa	B	Ja
The updated 2019 ARVC risk calculator should be considered to aid individualized decision-making for ICD implantation in patients with ARVC	IIa	B	Flere forbehold, men kan indgå i helhedsvurderingen
Recommendation Table 30 — Recommendations for the management of patients with restrictive cardiomyopathy side 72	**Class**	**Level**	
It is recommended that multimodality imaging be used to differentiate RCM from HCM or DCM with restrictive physiology	I	C	Ja
It is recommended that baseline cardiac and non-cardiac investigations are performed to assess involvement of the neuromuscular system or other syndromic disorders.	I	C	Ja
Cardiac catheterization is recommended in all children with RCM to measure pulmonary artery pressures and PVR at diagnosis and at 6–12 monthly intervals to assess change in PVR	I	B	Efter individuel vurdering
ICD implantation is recommended to reduce the risk of sudden death and all-cause mortality in patients with RCM who have survived a cardiac arrest or have recovered from a ventricular arrhythmia causing haemodynamic instability.	I	C	Ja

Endomyocardial biopsy should be considered in patients with RCM to exclude specific diagnoses (including iron overload, storage disorders, mitochondrial cytopathies, amyloidosis, and granulomatous myocardial diseases) and to diagnose restrictive myofibrillar disease caused by desmin variants.	IIa	C	Ja
ICD implantation may be considered in children with RCM who have evidence of myocardial ischaemia and syncope.	IIb	C	Efter individuel vurdering

Recommendation Table 31 — Exercise recommendations for patients with cardiomyopathy side 80	Class	Level	
ALL CARDIOMYOPATHIES			
Regular low- to moderate-intensity exercise is recommended in all able individuals with cardiomyopathy	I	C	Ja, dog særlige forhold ved ARVC
An individualized risk assessment for exercise prescription is recommended in all patients with cardiomyopathy.	I	C	Ja
HCM			
High-intensity exercise and competitive sport should be considered in genotype-positive/ phenotype-negative individuals who seek to do so	IIa	C	Ja
High-intensity exercise and competitive sport may be considered in asymptomatic low-riskc individuals with morphologically mild hypertrophic cardiomyopathy in the absence of resting or inducible left ventricular outflow obstruction and exercise-induced complex ventricular arrhythmias	IIb	B	Ja
High-intensity exercise, including competitive sport, is not recommended in high-risk individuals and in individuals with left ventricular outflow tract obstruction and exercise-induced complex ventricular arrhythmias.	III	B	Ja
ARVC			
Avoidance of high-intensity exercise, including competitive sport, may be considered in genotype-positive/phenotype-negative individuals in families with ARVC	IIb	C	Ja
Moderate- and/or high-intensity exercise, including competitive sport, is not recommended in individuals with ARVC	III	B	Ja
DCM and NDLVC			
Moderate- and high-intensity exercise should be considered in individuals who are gene positive and phenotype negative (with the exception of pathogenic variants in LMNA and TMEM43) who seek to do so	IIa	C	Ja, dog også medtaget asymptomatiske/fænotypisk raske LMNA bærere

High-intensity exercise and competitive sport may be considered in a select group of asymptomatic and optimally treated individuals with a left ventricular ejection fraction ≥50% in the absence of exercise-induced complex arrhythmias.	IIb	C	Ja
Moderate-intensity exercise may be considered in asymptomatic and optimally treated individuals with a left ventricular ejection fraction of 40–49% in the absence of exercise-induced complex arrhythmias.	IIb	C	Ja
High-intensity exercise, including competitive sport, is not recommended in symptomatic individuals, those with a left ventricular ejection fraction ≤40%, exercise-induced arrhythmias or pathogenic variants in LMNA or TMEM43	III	C	Ja, dog ikke indbefattet fænotypiske raske LMNA bærere

Recommendation Table 32 — Recommendations for reproductive issues in patients with cardiomyopathy side 83	Class	Level	
Pre-pregnancy risk assessment and counselling are recommended in all women using the mWHO classification of maternal risk.	I	C	Ja
Counselling on safe and effective contraception is recommended in all women of fertile age and their partners.	I	C	Ja, ved relevans
Counselling on the risk of disease inheritance is recommended for all men and women before conception.	I	C	Ja
Vaginal delivery is recommended in most women with cardiomyopathies, unless there are obstetric indications for caesarean section, severe heart failure (EF <30% or NYHA class III–IV), or severe outflow tract obstructions, or in women presenting in labour on oral anticoagulants.	I	C	Ja
It is recommended that medication be carefully reviewed for safety in advance of pregnancy and adjusted according to tolerability in pregnancy.	I	C	Ja
Therapeutic anticoagulation with LMWH or VKAs according to the stage of pregnancy is recommended for patients with AF.	I	C	Ja
Continuation of beta-blockers should be considered during pregnancy in women with cardiomyopathies, with close follow-up of foetal growth and of the condition of the neonate, and if benefits outweigh risks.	IIa	C	Ja
Genetic counselling and testing should be considered in patients with peripartum cardiomyopathy.	IIa	C	Ja

Recommendation Table 33 — Recommendations for non-cardiac surgery in patients with cardiomyopathy Side 83	Class	Level	
Peri-operative ECG monitoring is recommended for all patients with cardiomyopathy undergoing surgery.	I	C	Efter individuel vurdering

In patients with cardiomyopathy and suspected or known HF scheduled for intermediate or high-risk NCS, it is recommended to re-evaluate LV function with echocardiography (assessing LVOTO in HCM patients) and measurement of NT-proBNP/BNP levels, unless this has recently been performed	I	B	Efter individuel vurdering	
It is recommended that cardiomyopathy patients with high-risk genotypes or associated factors for arrhythmic or heart failure complications or severe LVOTO be referred for additional specialized investigations to a cardiomyopathy unit before undergoing elective NCS.	I	C		Anbefales ikke som udgangspunkt
In patients aged <65 years with a first-degree relative with a cardiomyopathy, it is recommended to perform an ECG and TTE before NCS, regardless of symptoms.	I	C		Anbefales ikke som udgangspunkt

Recommendation Table 34 — Recommendation for management of cardiovascular risk factors in patients with cardiomyopathy side 86	Class	Level	
Identification and management of risk factors and concomitant diseases is recommended as an integral part of the management of cardiomyopathy patients.	I	C	Ja