

Society Guidelines

The Canadian Cardiovascular Society/Canadian Heart Rhythm Society Comprehensive Guidelines for the Selection, Implantation, and Management of Patients With Cardiac Implantable Electronic Devices

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This statement was developed following a thorough consideration of medical literature and the best available evidence and clinical experience. It represents the consensus of a Canadian panel comprised of multidisciplinary

experts on this topic with a mandate to formulate disease-specific recommendations. These recommendations are aimed to provide a reasonable and practical approach to care for specialists and allied health professionals obliged with the duty of bestowing optimal care to patients and families, and can be subject to change as scientific knowledge and technology advance and as practice patterns evolve. The statement is not intended to be a substitute for physicians using their individual judgement in managing clinical care in consultation with the patient, with appropriate regard to all the individual circumstances of the patient, diagnostic and treatment options available and available resources. Adherence to these recommendations will not necessarily produce successful outcomes in every case.

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ABSTRACT

The Canadian Cardiovascular Society/Canadian Heart Rhythm Society cardiac implantable electronic device (CIED) guidelines program was developed to aid clinicians in the management of patients with a CIED, as well as to provide direction to policy makers and health care systems regarding issues related to CIEDs. There has never before been a comprehensive Canadian Cardiovascular Society/Canadian Heart Rhythm Society guideline to address the management of patients with CIEDs. This guideline is intended to be used by practicing clinicians across all disciplines who care for patients with CIEDs. We used the Grading of Recommendations, Assessment, Development and Evaluations (GRADE) system to grade recommendation strength and the quality of evidence. Areas of focus include bradycardia, syncope, cardiac resynchronization therapy, and conduction system pacing, sudden death prevention, and defibrillator therapy, the role of leadless devices, perioperative considerations and complications, lead extraction, postimplant CIED management, and competency and training standards. Extensive use is made of tables and figures to synthesize important material and present key concepts. This document should be an important aid for knowledge translation and a valuable reference tool to help improve the clinical management of patients who are assessed for possible CIED implantation or to improve decision-making for those who already have CIEDs.

RÉSUMÉ

Les lignes directrices sur les dispositifs électroniques implantables cardiaques (DEIC) de la Société cardiovasculaire du Canada (SCC)/Société canadienne de rythmologie (SCR) ont été élaborées pour aider les cliniciens à prendre en charge les patients porteurs de tels dispositifs et pour servir de guide aux décideurs politiques et aux systèmes de soins de santé relativement aux enjeux liés aux DEIC. C'est la première fois que la SCC/SCR publient des lignes directrices exhaustives sur la prise en charge des patients porteurs d'un DEIC. Ces lignes directrices sont conçues pour être utilisées par les praticiens qui prennent en charge des patients porteurs d'un DEIC, quelle que soit leur discipline. Nous avons utilisé le système GRADE (*Grading of Recommendations, Assessment, Development and Evaluations*) pour évaluer la force des recommandations et la qualité des preuves. Les domaines d'intérêt comprennent la bradycardie, la syncope, la thérapie de resynchronisation cardiaque et la stimulation du système de conduction, la prévention de la mort subite et le traitement par défibrillateur, le rôle des dispositifs sans sonde, les considérations et complications périopératoires, l'extraction de la sonde, la prise en charge des DEIC après l'implantation ainsi que les normes de compétences et de formation. Des tableaux et des figures ont été utilisés dans une large mesure pour synthétiser les documents importants et présenter les concepts clés. Ce document se veut une aide importante pour la transposition des connaissances, et un outil de référence précieux pour améliorer la prise en charge clinique des patients évalués en vue de l'implantation possible d'un DEIC et la prise de décisions concernant ceux qui portent déjà un DEIC.

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Cardiac implantable electronic devices (CIEDs) are medical devices that play a significant role in the management of heart disease by regulating cardiac rate and rhythm, modulating cardiac function or hemodynamics, and providing important diagnostic information regarding cardiac rhythm and cardiac function (eg, volume status and contractility). CIEDs include pacemakers, which treat slow heart rhythms by increasing the heart rate (HR); implantable cardioverter defibrillators (ICDs), which terminate potentially fatal rapid heart rhythms by delivering rapid stimulation or an electric shock; cardiac resynchronization therapy (CRT) devices, which improve cardiac contractility and function by enabling a more coordinated ventricular contraction; and implantable cardiac monitors (ICMs), which are used to diagnose abnormal cardiac rhythms. Although millions of patients have benefited from these important technologies, the clinical management of these CIED systems is challenging because these devices have increased in complexity.

The Canadian Cardiovascular Society (CCS)/Canadian Heart Rhythm Society (CHRS) CIED guidelines program was developed to aid clinicians in the management of patients with CIEDs, as well as to provide direction to policy makers and health care systems regarding issues related to CIEDs. Previous CCS guidelines have focused only on CRT (2013) or ICDs (2017).^{1,2} The current CCS/CHRS CIED guidelines represent a comprehensive renewal that integrates, updates, and replaces these guidelines, recommendations, and practical tips. It is intended to be used by practicing clinicians across all disciplines who care for patients assessed for or who have CIEDs.

Preamble and Guideline Development Methodology

This document was developed in accordance with CCS best practices and the Grading of Recommendations, Assessment, Development, and Evaluation (GRADE) approach. The Primary and Review Panels, including the co-chairs were appointed by the CCS Guidelines Committee. The primary panelists developed the scope of the document, identified topics for review, performed the literature review, evaluated the quality of evidence, and drafted the recommendations. A systematic search was performed to identify relevant studies within each topic. One methodologist (D.S. C.) was assigned from the methodology subcommittee of the CCS Guidelines Committee to the primary panel to conduct

2 systematic reviews and meta-analyses. The remaining 47 evidence reviews were performed by the primary panel members. Each topic was addressed in the form of a “PICO” question: patient population of interest (P), intervention (I), comparator (C), and outcomes (O). Strength of recommendations and quality of evidence were evaluated according to the GRADE approach. Evidence derived from randomized clinical trials (RCTs) was initially estimated as high-quality evidence, with observational evidence initially deemed low-quality evidence. These estimates were further refined through detailed appraisal into 4 categorical grades (“high,” “moderate,” “low,” and “very low”) on the basis of a consideration of the risk of bias, confounding, consistency of results, directness of evidence, precision, publication bias, magnitude of effect, and dose-response gradient. After this evaluation, the strength of recommendation was determined by considering the balance between desirable and undesirable effects (ie, risk-benefit), confidence in the magnitude of effect (quality of evidence), resource considerations, as well as patient values and preferences. The strength of recommendation was graded as “strong” (the desirable effects outweigh the undesirable effects, and therefore, most individuals will be best served by the recommended course of action) or “weak” (the desirable effects probably outweigh the undesirable effects, indicating that there is a need to consider the individual patient’s clinical details, circumstances, values, and preferences). Draft recommendations were presented, reviewed, and refined by the primary panel. Members of the primary panel who did not have direct conflicts of interest with the substance of a specific recommendation were allowed to vote on that recommendation. All recommendations in the guideline reached a majority agreement (85%) of eligible voters.

In addition, all previous recommendations relevant to CIED therapy were reviewed.¹⁻⁹ Each CCS recommendation was reviewed by the primary panel, and was either retained, omitted, or revised on the basis of the evidence published subsequent to their development. Recommendations for which significant advances had occurred in the year(s) since their development were omitted, and the GRADE process, as outlined previously, was performed to create a new recommendation. Those recommendations that were believed to be clinically applicable in their current form were brought forward to the current guideline document without an updated GRADE review. In some cases, these recommendations underwent minor modification in an effort to improve

readability and to eliminate confusion or ambiguity, however, the intent and meaning of the recommendation, strength of recommendation, and level of evidence were unchanged from their initial derivation.

Peer review of the guideline document was provided by external content experts, which included 2 journal reviewers nominated by the *Canadian Journal of Cardiology* editorial board, and the CCS Guidelines Committee members.

A summary of the evidence-to-decision frameworks for this guideline document is available in the *Evidence to Decision* section of the [Supplementary Material](#). The methods for this guideline development are detailed in the *Guideline Development Methodology* section of the [Supplementary Material](#). Further, the decision-making process is detailed for each recommendation in terms of the PICO question addressed, the voting record, the specific evidence in the literature, the analysis of the problem, the certainty of evidence and balance of effects determinations, the resource analysis and cost effectiveness considerations, health equity considerations, acceptability to key stakeholders, and feasibility.

1. Background

CIEDs are among the top 10 most implanted medical devices worldwide, with more than 1.5 million devices implanted annually. In Canada, the primary implantation rate for pacemakers and ICDs increased ten-fold from 2003 (7/100,000 persons) to 2019 (70/100,000). It is estimated that approximately 120,000 Canadians are living with CIEDs.

Key definitions in relation to CIED therapy are presented in [Table 1](#), with different types of CIED systems presented in [Figure 1](#).

2. Bradycardia

2.1. Classification of bradycardia

Treatment or prevention of bradyarrhythmia is the primary indication for implantation of permanent pacemakers (PPMs). These conditions can be broadly divided into intrinsic and extrinsic disorders that affect the sinus node and atrioventricular (AV) conduction ([Table 2](#)).

2.2. Clinical evaluation of suspected bradycardia

An approach to the investigation of patients with suspected bradycardia is presented in [Figure 2](#), with [Figure 3](#) providing guidance regarding the initial evaluation of suspected sinus bradycardia and [Figure 4](#) providing guidance regarding the initial evaluation of suspected AV disease.

Patients with suspected bradycardia or conduction disorder should undergo comprehensive history and physical examination. A 12-lead electrocardiogram (ECG) should be performed to document rate, rhythm, and conduction, as well as screen for associated disorders. A

transthoracic echocardiogram should be performed to evaluate the etiology of conduction disturbances, determine left ventricular (LV) function, evaluate valve function, and be used to guide appropriate device selection. In some cases, multimodality imaging (transesophageal echocardiography, computed tomography (CT), cardiac magnetic resonance imaging [MRI], or nuclear imaging) might be necessary to characterize the underlying substrate (eg, inflammatory or infiltrative disease; congenital heart disease [CHD]), particularly those who present at an age younger than 60 years. In select cases, exercise stress testing might provide diagnostic confirmation in patients with exertional symptoms, in whom chronotropic incompetence is suspected, or with documented AV block (AVB) at an unknown level. Referral to a cardiogenetic clinic for consideration of genetic testing is reasonable for patients before they are 60 years of age or at younger than 65 years in the presence of a positive family history.

Recommendation

1. We recommend ambulatory ECG monitoring for symptom-rhythm correlation in patients with suspected bradycardia or conduction disorder in whom a comprehensive evaluation did not show a cause (Strong Recommendation; High-Quality Evidence).

Practical tip. The duration of monitoring is dependent on symptom frequency (eg, short duration noninvasive monitoring may be appropriate for patients with frequent symptoms, and implantable monitors may be preferred for those with recurrent but rare symptoms).

Recommendation

2. We suggest screening for sleep apnea for patients with sleep-related bradycardia or conduction disorder (Weak Recommendation; Moderate-Quality Evidence).

2.3. Pacing for sinus node dysfunction

Sinus node dysfunction (SND) includes a constellation of conditions that result in impairment of the sinoatrial node to generate and propagate impulses. This is usually due to diffuse atrial and conduction system fibrosis within the sinoatrial node, which leads to inappropriate sinus bradycardia and exaggerated overdrive suppression, and causes a combination of prolonged conversion pauses, sinus node exit block, and chronotropic incompetence.

Table 1. Key definitions

Term	Definition
Sinus node dysfunction	Inappropriate sinus bradycardia • Sinus bradycardia (sinus rate < 50 bpm) in the absence of physiologic explanation or a sinus bradycardia that results in symptoms Ectopic atrial bradycardia • Bradycardia arising from an atrial pacemaker other than the sinus node with a rate of < 50 bpm Tachycardia-bradycardia syndrome • Atrial tachycardia (usually atrial fibrillation) alternating with sinus bradycardia, ectopic atrial bradycardia, or sinus arrest (conversion pauses) due to suppression of sinus node automaticity Intermittent sinus pauses • Sinus arrest: transient cessation of sinus node firing (> 3 seconds) • Sinoatrial exit block: depolarization fails to exit the sinus node and depolarize adjacent atrial tissue Chronotropic incompetence: Inappropriately low HR in response to physical activity or physiologic demand • Absolute: inability to increase HR to 80% of the age-predicted target (220-age) or to > 100-120 bpm • Relative: able to reach target HR but at a significant delay with reduced exercise tolerance
AV delay or block	First-degree AV delay: All impulses are conducted to the ventricles (1:1 conduction) but with prolonged conduction time (PR interval > 200 ms due to slow intra-atrial, AV node, and/or His bundle conduction) Second-degree AV block: Intermittent failure of AV conduction, whereby not every atrial impulse is conducted to the ventricles, but some form of AV relationship exists • Mobitz type I: P waves with a physiologic rate (< 100 bpm) with gradual prolongation in the PR interval (Wenckebach sequence) after a nonconducted beat. Usually signifies nodal or supranodal level of block • Mobitz type II: P waves with a physiologic rate (< 100 bpm) with a periodic single nonconducted P wave associated with constant PR interval. Might signify nodal or infranodal (intrahisian or infrahisian) level of block • 2:1 AV block: P waves with a physiologic rate (< 100 bpm) when every other P wave conducts to the ventricles due to nodal or infranodal (intrahisian or infrahisian) AV block Advanced, high-grade or high-degree AV block: ≥ 2 consecutive P waves at a constant physiologic rate that do not conduct to the ventricles with evidence for some AV conduction. Might signify nodal or infranodal (intrahisian or infrahisian) level of block Third-degree AV block: complete failure of AV conduction when P waves with a physiologic rate (< 100 bpm) are not conducted and never produce a QRS response (independent atria and ventricles; AV dissociation). Might signify intranodal or infranodal (intrahisian or infrahisian) level of block
Cardiomyopathy	Disorders characterized by morphologically and functionally abnormal myocardium in the absence of any other disease that is sufficient, by itself, to cause the observed phenotype¹⁰ • Ischemic cardiomyopathy: ventricular dilation and depressed myocardial contractility caused by irreversible ischemia or infarction • Dilated cardiomyopathy: ventricular dilation and depressed myocardial performance in the absence of, or disproportionate to, the degree of ischemic heart disease or other disorders sufficient to cause global systolic impairment • Nondilated LV cardiomyopathy: nonischemic LV scarring or fatty replacement regardless of the presence of global or regional wall motion abnormalities, or isolated global LV hypokinesia without scarring (eg, myocarditis, muscular dystrophy) • Arrhythmogenic RV cardiomyopathy: cardiomyopathy characterized by predominantly RV dilatation and/or dysfunction in the presence of histological involvement (fibrofatty replacement) and/or electrocardiographic abnormalities • Hypertrophic cardiomyopathy: cardiomyopathy characterized by LV wall thickening with or without RV hypertrophy (sarcomeric and nonsarcomeric) not solely explained by abnormal loading conditions • Restrictive cardiomyopathy: restrictive left and/or RV pathophysiology in the presence of normal or reduced diastolic volumes (of one or both ventricles), normal or reduced systolic volumes, and normal ventricular wall thickness

AV, atrioventricular; bpm, beats per minute; HR, heart rate; LV, left ventricular; RV, right ventricular.

It is important to distinguish pathological SND from physiologic bradycardia, in which marked sinus bradycardia < 35 beats per minute (bpm) and sinus pauses > 2 seconds can be frequently observed in young, healthy individuals and athletes especially during nocturnal hours.^{12,13} Because of the large overlap between observed heart rate in physiologic sinus bradycardia and pathologic SND, distinguishing between the two is challenging, with no absolute heart rate thresholds or pause durations that provide diagnostic clarity. Asymptomatic sinus bradycardia is not an indication for pacing because the natural history of SND is generally benign, and the only

demonstrated benefit of pacing therapy is symptom improvement.¹⁴⁻¹⁶ As such, pacing is contingent on establishing symptom-rhythm correlation, which is usually through ambulatory rhythm monitoring. Because patients with sinus node disease might develop progressive AV conduction disorders (estimated at 1%-2% per year), when pacing is indicated a dual-chamber device is recommended.^{14,17} In these patients, programming to minimize right ventricular (RV) pacing using specialized algorithms is beneficial to decrease the risk of pacing-induced LV dysfunction and potentially atrial fibrillation (AF; see section 10.1).

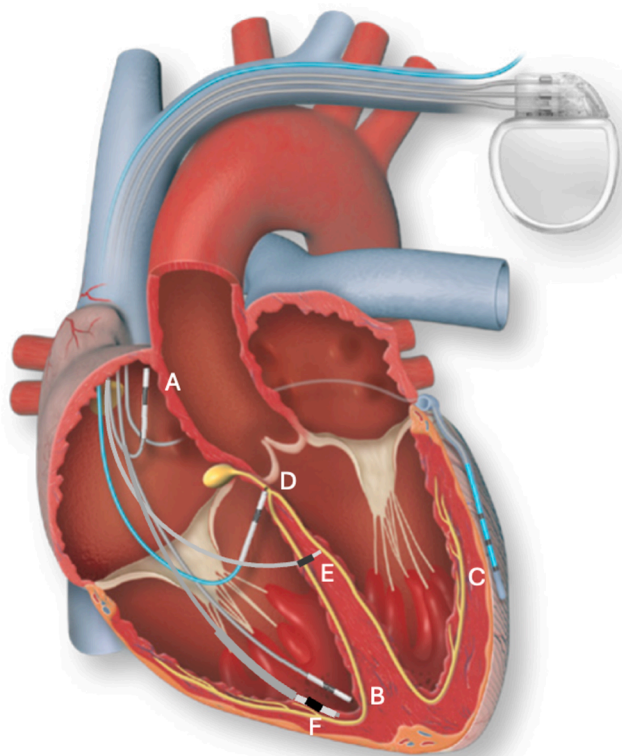


Figure 1. Cardiac pacing possibilities. **A** indicates pacing lead within the right atrium; **B** indicates pacing lead at the right ventricular apical septum. **C** indicates pacing lead in a branch of the coronary sinus for biventricular pacing-cardiac resynchronization therapy (CRT). **D** indicates pacing lead at the His bundle for conduction system pacing (CSP). **E** indicates pacing lead at the left bundle branch for CSP. **F** indicates implantable cardioverter defibrillator lead used for defibrillation and for pacing (if required). Single-chamber pacemaker (VVI) typically refers to a single pacing lead within the right ventricle using pacing lead locations D, E, or B. A standard dual-chamber pacemaker (DDD) typically refers to a pacemaker using a lead within the atrium (location A) and a lead within the right ventricle using pacing lead locations D, E, or B. When an atrial lead (location A) is combined with a defibrillator lead (location F), this is termed a dual-chamber implantable cardioverter defibrillator. Biventricular pacing is performed using a pacing lead at the right ventricular septum (location B) combined with a coronary sinus lead (location C) for CRT-pacemaker. When biventricular pacing is performed using a defibrillator lead at the right ventricular apical septum (location F) combined with a coronary sinus lead (location C), this is termed CRT-defibrillator. The term, “hot-CRT” might be used when a CSP lead at the His bundle (location D) is combined with a coronary sinus lead. The term, “lot-CRT,” might be used when a CSP lead at the left bundle branch (location E) is combined with a coronary sinus lead. Adapted with permission from Glikson et al.¹¹

Recommendation

3. We recommend PPM implantation for patients with symptomatic sinus node dysfunction not caused by reversible or physiologic causes (Strong Recommendation; High-Quality Evidence).

Practical tips

- This recommendation applies to inappropriate sinus bradycardia, chronotropic incompetence, intermittent sinus pauses, and tachycardia-bradycardia syndrome.
- Establishing a temporal correlation between symptoms and bradycardia is important when determining whether permanent pacing is needed.
- There is no minimum HR or pause duration to indicate permanent pacing is absolutely indicated in the absence of symptoms.
- With tachycardia-bradycardia syndrome, consider treatment of the underlying cause for tachyarrhythmias before pacemaker implantation. In some cases, conversion pauses might respond to tachycardia management. For example, catheter ablation or pharmacological treatment of AF and/or atrial flutter should be considered in patients with arrhythmia-related bradycardia or conversion pauses before proceeding to pacemaker implantation.
- Pacing is not recommended in patients with SND because of a transient cause that can be corrected and prevented. Essential drug therapy is considered a nonreversible cause.
- Pacing is generally not recommended for asymptomatic nocturnal pauses or bradycardia.

2.4. Pacing for AVB

AVB can occur at or above the level of the AV node (AVN; intra- or supranodal) or below the AVN (infranodal—including intra-His and infra-His block). Whereas the former tends to be benign and nonprogressive, infranodal block is more likely to progress to more advanced forms of AV conduction block with an unpredictable escape rhythm.^{18,19} In the absence of a reversible cause (Table 2), pacing therapy is indicated for AVB in association with symptoms, as well as acquired high-grade or third-degree AVB irrespective of symptoms. Worsening AVB with exertion on ambulatory ECG or exercise testing can indicate infranodal block,²⁰ as can a prolonged His-ventricular electrogram (HV) interval on invasive electrophysiologic testing.²¹

Alternating bundle branch block suggests the presence of severe infranodal disease. These patients should be offered pacing even if asymptomatic. Pacing is generally not indicated for less advanced conduction system disease (eg, first-degree AVB or isolated fascicular block).

Recommendation

4. We recommend PPM implantation for patients with acquired third-degree AVB, type II second-degree AVB, high-grade AVB, or alternating bundle branch block not caused by reversible or physiologic causes, regardless of symptoms (Strong Recommendation; Low-Quality Evidence).

Table 2. Causes of bradycardia

Intrinsic
Degenerative (most common) • Idiopathic age-related fibrosis Congenital • Immune-mediated • Structural heart disease • Postsurgical Myocardial ischemia and/or infarction • Antiarrhythmic drugs ◦ Inferior MI (neurocardiogenic or direct ischemia) ◦ Anterior MI (His-Purkinje ischemia) Myocardial infiltration • Sarcoidosis • Amyloidosis • Hemochromatosis Connective tissue disorder • Rheumatoid arthritis • Scleroderma • Systemic lupus erythematosus Neuromuscular disorder • Myotonic dystrophy 1 (Steinert's) • Emery Dreifuss muscular dystrophy • Limb girdle muscular dystrophy 1b • Kearns-Sayre syndrome Inflammatory and/or infectious • Chagas (SND and AVB) • Myopericarditis (AVB more commonly present than SND) • Lyme, diphtheria, toxoplasmosis (AVB more commonly present than SND) Direct injury or trauma • Catheter ablation • Post cardiac surgery • Post transcatheter valve replacement • Radiation therapy Ion channel disorder
Extrinsic
Drugs • Antiarrhythmic drugs ◦ Class I: flecainide, propafenone ◦ Class II: β-blockers ◦ Class III: more common with amiodarone than dronedarone or sotalol ◦ Class IV: nondihydropyridine calcium channel blockers ◦ Other: digoxin (SND and AVB), ivabradine (SND) • Sympatholytic agents (methyldopa, clonidine: SND) • Psychoactive and neuroactive agents ◦ Donepezil, lithium, phenytoin, phenothiazine, carbamazepine (SND and AVB) ◦ SSRI, TCA (AVB) • Antiplatelet: ticagrelor • Chemotherapeutic ◦ Bortezomib, cyclophosphamide, 5-fluorouracil, mitoxantrone, rituximab, thalidomide (SND and AVB) ◦ Capecitabine, cisplatin, doxorubicin, epirubicin, ifosfamide, interleukin-2, methotrexate, paclitaxel (SND) ◦ Anthracycline, taxane (AVB) • Gastrointestinal: H2RA (SND and AVB), PPI (SND) Metabolic • Potassium (hyper-, hypo-) • Hypercalcemia, hypermagnesemia • Hypothyroidism • Hypothermia • Hypoxia • Anorexia • Acidosis Autonomic mediated • Neurocardiogenic • Carotid sinus hypersensitivity Neurological disorder • Increased intracranial pressure • CNS tumours

Table 2. Continued.

Extrinsic
• Temporal epilepsy Obstructive sleep apnea Toxins
Unless otherwise specified cause, bradycardia might result in SND and AVB. AVB, atrioventricular block; CNS, central nervous system; H2RA, histamine receptor antagonist; MI, myocardial infarction; PPI, proton pump inhibitor; SND, sinus node dysfunction; SSRI, selective serotonin reuptake inhibitor; TCA, tricyclic antidepressant.

Practical tips

- Establishing temporal correlation between symptoms and bradycardia is less important when determining whether permanent pacing is needed for AVB.
- Pacing is not recommended in patients with AVB due to a transient cause that can be corrected and prevented. Essential drug therapy is considered a nonreversible cause.
- Alternating bundle branch block suggests the presence of severe infranodal disease and pacing should be offered irrespective of symptoms.
- It is important to determine if transient 2:1 AVB is due to supranodal or infranodal causes when considering the need for pacemaker implantation.

Recommendation

- We recommend PPM implantation for patients with significant pauses (≥ 6 seconds) during AF or sustained slow ventricular response (HR < 40 bpm) during AF while awake (Strong Recommendation; Low-Quality Evidence).

Practical tips

- Pauses and/or slow ventricular response during AF are a manifestation of high-grade AVB.
- This recommendation does not reflect conversion pauses, which should be treated as per SND (section 2.3)
- Pacing is not recommended in patients with bradycardia due to a transient cause that can be corrected and prevented. Essential drug therapy is considered a nonreversible cause.
- Pacing is not recommended for asymptomatic nocturnal bradycardia.

2.5. Bradycardia in special populations

2.5.1. Pacing post myocardial infarction. AVB after an acute myocardial infarction (MI) can be common, although the incidence is decreasing with increasing availability of primary percutaneous coronary intervention. AVB associated with MI can occur in the proximal (AVN or upper His) or distal conduction system (below the His). Proximal AVB is more common (12%-20%) and appears earlier

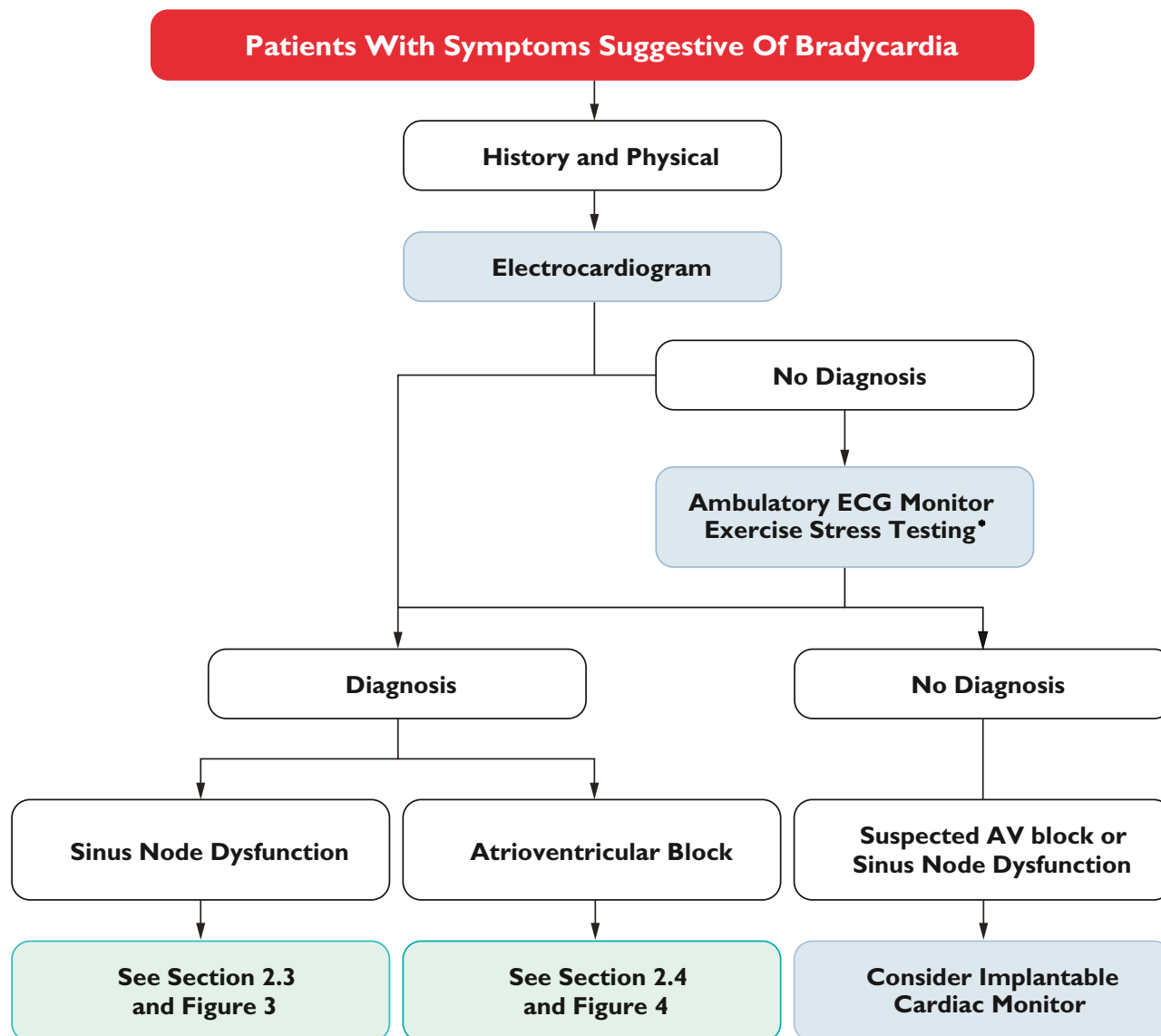


Figure 2. Evaluation of symptomatic bradycardia. AV, atrioventricular; ECG, electrocardiogram. If exertional symptoms are suspected, consider ambulatory ECG monitoring or exercise stress testing to establish symptom-rhythm correlation.

during acute MI. It is associated with inferior MI and manifests as a stable narrow escape rhythm responsive to pharmacological intervention (atropine, isoproterenol). Proximal AVB might require temporary pacing if associated with hemodynamic compromise but usually resolves without need for permanent pacing, although the time to resolution might be 7-14 days.^{22,23} Conversely, distal AVB occurs more rarely (5%), manifests later, and is associated with a large anterior infarction. Distal AVB presents with a slow unstable escape rhythm not responsive to atropine and/or isoproterenol, generally requires temporary pacing, and most often requires permanent pacing.^{22,24,25} Because infranodal block (either complete AVB or bundle branch block) in association with acute MI reflects extensive myocardial damage and infranodal conduction necrosis, it is reasonable to consider implantation of a cardiac resynchronization-capable device (with or without ICD) in preference to a standard RV pacemaker.²⁶

Recommendation

6. We recommend PPM implantation for patients with third-degree AVB, type II second-degree AVB, high-grade AVB, alternating bundle branch block, or symptomatic SND that persists for more than 5 days after MI (Strong Recommendation; Low-Quality Evidence).

Practical tips

- Consider earlier pacemaker implantation for patients in whom the likelihood of recovery in AV conduction is low, or in patients with infranodal conduction block (AVB or bifascicular block) in the context of anterior wall MI.

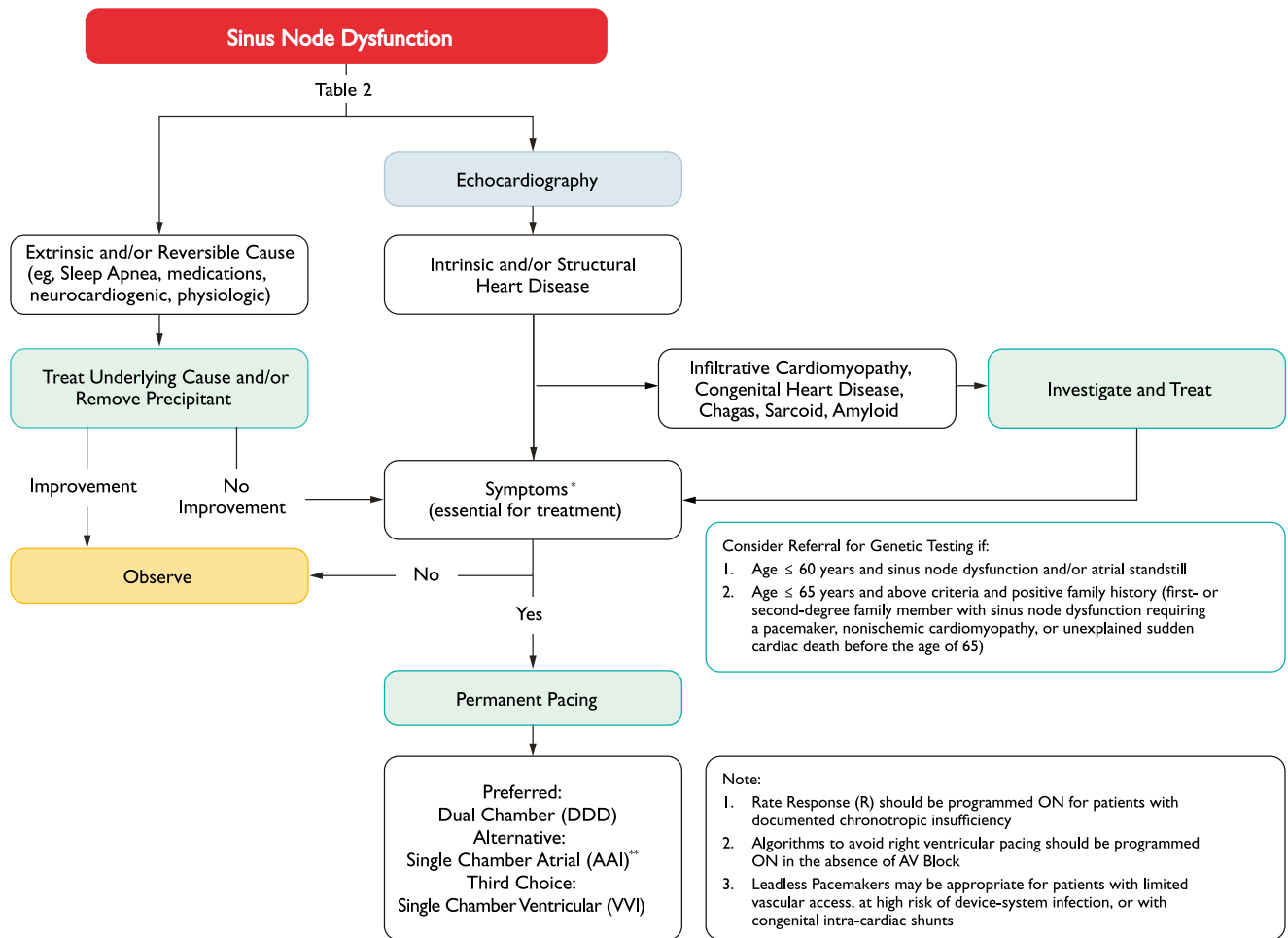


Figure 3. Diagnostic evaluation and management of sinus node dysfunction. AV, atrioventricular; ECG, electrocardiogram. * Symptoms at rest, such as presyncope due to pauses, or provoked with exertion, including effort intolerance due to chronotropic insufficiency. If exertional symptoms are suspected, consider ambulatory ECG monitoring or exercise stress testing to establish symptom-rhythm correlation. ** For those with normal AV conduction, low risk of progression to AV block, and strong desire to avoid multiple leads due to limited vascular access.

- If proceeding with device implantation, strongly consider ICD implantation in those with reduced LV ejection fraction (LVEF; see section 6).
- If proceeding with device implantation for AVB, strongly consider resynchronization therapy if LVEF is $< 45\%$ and anticipated RV pacing burden $> 20\%$ - 40% , or AVB complicates anterior infarction (see sections 4.2 and 5.2).
- Pacing is not recommended if bradycardia resolves after revascularization.

of patients with perioperative device implantation for SND and up to 25% of those who receive a device implanted for AVB are not pacemaker-dependent at follow-up. Therefore, a minimal waiting period of at least 5 days is recommended before proceeding to the insertion of a permanent CIED. A multidisciplinary heart team approach is recommended for the preoperative identification of patients who might benefit from intraoperative insertion of permanent epicardial pacing leads, and to individualize timing and device selection for those who require permanent pacing postoperatively.³¹⁻³³

2.5.2. Pacing post cardiac surgery. Perioperative SND, AVB, or infranodal conduction abnormalities are common findings after cardiac surgery, which affect up to 10%-15% of patients in unselected cohorts.^{27,28} However, the incidence and persistence of perioperative conduction disorders depends largely on the type of cardiac surgery (Table 3) and preexisting patient-related factors (Table 4).^{29,30} Nevertheless, contemporary data suggest a spontaneous resolution of new-onset conduction disorders in a significant proportion of patients within the first 5-7 days after cardiac surgery. Up to 60%-70%

Recommendation

7. We recommend PPM implantation for patients with third-degree AVB, type II second-degree AVB, high-grade AVB, alternating bundle branch block, or symptomatic SND that persists for more than 5 days after cardiothoracic surgery for valvular disease (Strong Recommendation; Low-Quality Evidence).

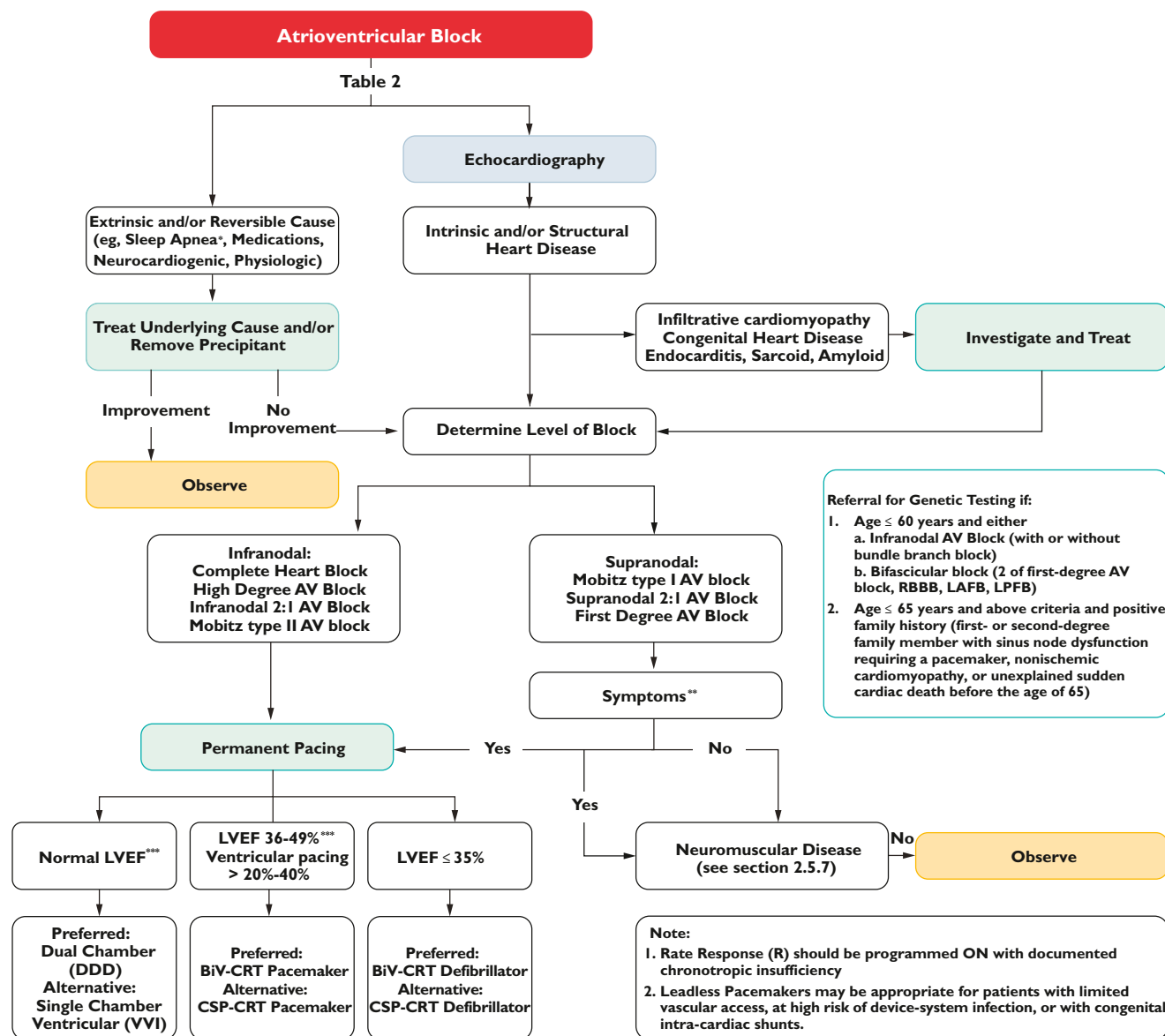


Figure 4. Diagnostic evaluation and management of atrioventricular block. AV, atrioventricular; BiV, biventricular; CRT, cardiac resynchronization therapy; CSP, conduction system pacing; LAFB, left anterior fascicular block; LPFB, left posterior fascicular block; LVEF, left ventricular ejection fraction; RBBB, right bundle branch block. * Asymptomatic nocturnal AV block should prompt testing and treatment for sleep apnea before considering pacing. ** If exertional symptoms are suspected, consider ambulatory electrocardiograph monitoring or exercise stress testing to establish symptom-rhythm correlation. *** Consider indication for implantable cardioverter defibrillator in those with LVEF > 35% and indication for pacing, such as the presence of sarcoid (section 2.5.6. and 6.3.1.), neuromuscular disorder (section 2.5.7. and 6.3.3.), lamin A/C (section 6.3.3.2.), adult congenital heart disease (section 2.5.4. and 6.3.5).

Practical tips

- Consider earlier pacemaker implantation for patients without stable escape rhythm or in those for whom the likelihood of recovery in AV conduction is low (Table 4).
- If proceeding with device implantation for AVB, strongly consider resynchronization therapy if LVEF < 45% and anticipated RV pacing burden > 20%-40% (see sections 4.2 and 5.2).

2.5.3. Pacing post transcatheter aortic valve replacement. Transcatheter aortic valve replacement (TAVR) is an established treatment for patients with symptomatic aortic stenosis with an intermediate to high surgical risk. Compared with surgery, TAVR is associated with higher rates of periprocedural pacemaker implantation, with PPM implantation being required in up to a third of patients who undergo TAVR.³⁴ Conduction system disturbance (left bundle branch

block [LBBB] or complete AVB) is thought to occur because of mechanical pressure from the valve prosthesis and associated localized edema and/or ischemia. Risk factors for PPM implantation after TAVR include self-expandable valves, valve implantation depth, calcification of the LV outflow tract, pre-existing conduction disturbances (right bundle branch block [RBBB] being of higher risk than LBBB), older patient age, and a high number of comorbidities.^{34,35} Pacemaker implantation after TAVR has been associated with a higher risk of all-cause mortality and hospitalization for heart failure (HF), although it is not certain if this risk merely reflects patient comorbidity rather than being a consequence of pacing itself.³⁶ An algorithm to guide device implantation is presented in Figure 5.

Recommendation

8. We recommend PPM implantation for patients with persistent third-degree AVB, type II second-degree AVB, high-grade AVB, or alternating bundle branch block after TAVR (Strong Recommendation; Moderate-Quality Evidence).

Practical tips

- Patients with preexisting RBBB are at high risk of heart block after TAVR.
- Prophylactic permanent pacing in high-risk patients before TAVR is of low utility.
- If proceeding with device implantation for AVB, resynchronization therapy might be considered if LVEF is < 45% and anticipated RV pacing burden > 20%-40% (see sections 4.2 and 5.2).

Recommendation

9. We suggest that patients with a new LBBB (QRS > 150 milliseconds [ms]) and first-degree AVB (PR interval > 240 ms) that persists for 48 hours or more after TAVR undergo PPM implantation (Weak Recommendation; Moderate-Quality Evidence).
10. We suggest that patients with either new LBBB (QRS > 150 ms) or new first-degree AVB (PR interval > 240 ms) after TAVR undergo PPM implantation, selective electrophysiologic testing for risk stratification and guidance of PPM implantation, or prolonged postdischarge ambulatory ECG monitoring (Weak Recommendation; Moderate-Quality Evidence).
11. We suggest that patients with new-onset conduction disturbance (> 20 ms increase in the PR interval to 200-240 ms or new-onset LBBB with QRS duration < 150 ms) after TAVR, or preexisting RBBB before TAVR, undergo prolonged inpatient telemetry (48 hours) with prolonged postdischarge ambulatory ECG monitoring (Weak Recommendation; Low-Quality Evidence).

Table 3. Type of cardiac surgery and incidence of perioperative SND and/or conduction disorders^{29,30}

Type of cardiac surgery*	Incidence of permanent postoperative CIED
Isolated CABG	0.8%-3.4%
Aortic valve surgery	3%-8.5%
Mitral valve surgery	1%-9%
Tricuspid valve surgery	22%-42%
Redo valve surgery	7.7%
Heart transplantation	2%-4% (bicaval anastomosis) to 10%-14% (biatrial anastomosis)
Surgical myectomy	3%-10 %
Endocarditis†	13% (22% if valvular abscess or tricuspid valve involvement)

CABG, coronary artery bypass grafting; CIED, cardiac implantable electronic device; SND, sinus node dysfunction.

* For each procedure, there is an increased risk for permanent pacemaker need in the presence of preoperative SND or conduction disorders or other patient-related factors.

† Independent predictor for the need of a postoperative permanent pacemaker in patients who undergo heart surgery for infective endocarditis: tricuspid valve involvement, *Staphylococcus aureus* infection, previous valve surgery, valvular abscess, and preexisting conduction abnormalities.²⁹

2.5.4. Adult CHD. CHD refers to structural malformations of the heart and/or great vessels that are present at birth, excluding inherited arrhythmia syndromes and cardiomyopathies.³⁷ Patients with adult CHD comprise a wide range of anatomic pathologies and associated palliative or corrective procedures. SND and AVB can be encountered in this population, with certain disease states or surgical repairs predisposing to conduction system disease (eg, congenitally corrected transposition of the great arteries and AVB; Mustard or Senning baffle and SND). Evidence of poor sinus node function is associated with increased mortality, and although evidence is lacking that pacing improves mortality, patient symptoms are often markedly improved.³⁸⁻⁴⁰ Some patients with CHD and asymptomatic bradycardia might warrant pacemaker implantation. Accordingly, decisions regarding pacing in this population should be tailored to the needs of the individual patient along with interdisciplinary discussion involving specialists with expertise in the management of adult patients with CHD.

Table 4. Predictors of postoperative permanent cardiac implantable electronic device insertion

Predictor
Type of cardiac surgery • Aortic valve surgery • Tricuspid valve surgery • Redo valve surgery • Multivalve surgery • Endocarditis
Patient-related factors • Age > 70 years • Preexisting sinus node dysfunction • Preexisting conduction disorders - First-degree atrioventricular block (PR interval > 200 ms) - Second- or third-degree atrioventricular block - Left bundle branch block - Right bundle branch block • Absence of preoperative sinus rhythm • Long-term need for antiarrhythmic medication or medication affecting sinus/atrioventricular node (eg, β-blocker treatment for heart failure)

Pacing strategies need to be carefully considered because of variations in patient anatomy. Epicardial pacing might be preferred in situations in which surgical intervention is already planned, when endocardial access is limited by anatomical considerations, or if there is concern of thromboembolism, such as in the presence of an intracardiac shunt or in a low-flow chamber. Epicardial leads, and in particular epicardial unipolar leads, are more prone to progressive lead failure and need for revision compared with endocardial leads.^{41,42} A desire to minimize intravascular lead components might also encourage either single-chamber atrial or ventricular devices, with an overall low reported rate for need for revision to dual-chamber pacing.^{41,43-45} Alternatively, leadless pacemakers might reduce venous occlusion risks, lead-related AV valve dysfunction, thromboembolic risk, and endocarditis risk in select patients.⁴⁶

Recommendation

12. We recommend pacemaker implantation be considered in patients with complex CHD and standard pacemaker indications, after an interdisciplinary discussion involving specialists with expertise in the management of patients with adult CHD (Strong Recommendation; Moderate-Quality Evidence).

Practical tip. Patients with CHD represent a complex and heterogeneous group, which is best managed in centres with a multidisciplinary team and expertise in CHD-related device therapy because of variations in cardiac anatomy and/or venous return, conduction systems, and history of cardiac repair.

2.5.5. Congenital AVB. Prenatally diagnosed congenital AVB is associated with a high mortality, leading to pacemaker implantation in two-thirds of neonates within the first year of life. In those with identification of congenital AVB later in life, it has been suggested that pacemaker implantation can be deferred in the absence of symptoms (eg, chronotropic insufficiency, presyncope, or syncope), or ventricular dysfunction. However, risk factors for severe symptoms and sudden death have been identified, suggesting that asymptomatic patients undergo device implantation in the presence of: a resting HR < 50 bpm, a wide QRS escape rhythm, junctional exit block manifesting as pauses (2-3 times the basic cycle length), QT prolongation, and the presence of multiform or frequent ventricular ectopy and/or ventricular tachycardia (VT), especially with exertion.⁴⁷⁻⁴⁹ The prophylactic use of pacing in the absence of risk factors may be considered, but is not supported by robust evidence.⁴⁸ Biventricular or conduction system pacing (CSP)

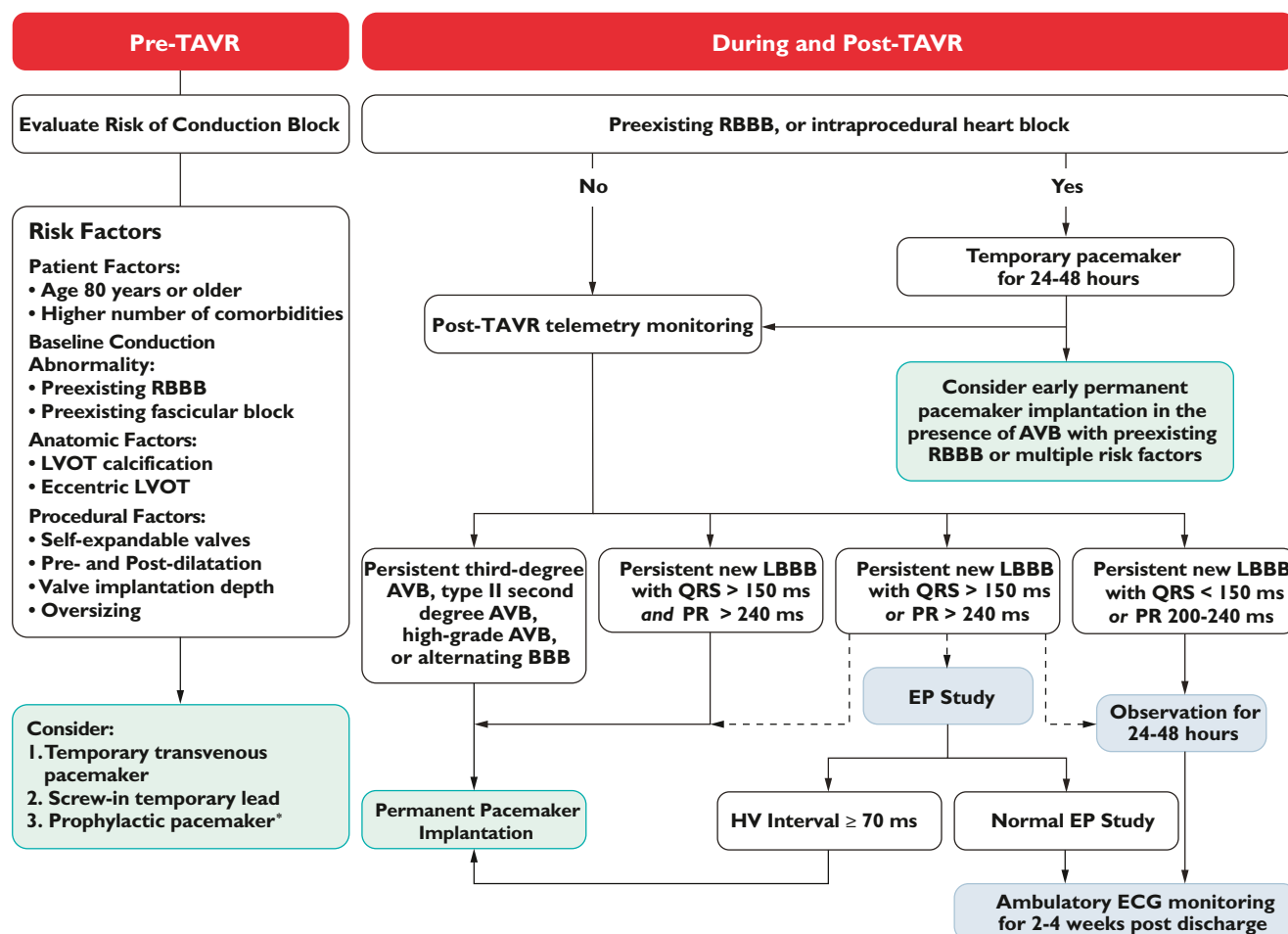


Figure 5. Considerations for patients who undergo transcatheter aortic valve replacement (TAVR). AVB, atrioventricular block; BBB, bundle branch block; ECG, electrocardiogram; EP, electrophysiology; HV, ventricular electrogram; LBBB, left bundle branch block; LVOT, left ventricular outflow tract; RBBB, right bundle branch block. * Pacing before TAVI of low utility beyond standard pacemaker indications.

might be considered because of the risk of pacing-induced cardiomyopathy (PICM), however, the type of pacing must be weighed against the limited evidence supporting such an approach in children or very young patients.

Recommendation

13. We recommend pacemaker implantation in patients with congenital AVB and symptoms of bradycardia, or asymptomatic patients with evidence of 1 or more of ventricular dilatation or dysfunction, mean daytime HR < 50 bpm, wide QRS escape rhythm, junctional exit block (pauses 2-3 times the basic cycle length), QT prolongation, or complex or frequent ventricular ectopy (Strong Recommendation; Low-Quality Evidence).

Practical tips

- Among patients with congenital AVB, the prophylactic use of pacing in the absence of risk factors might be considered, but it is not supported by robust evidence.
- Resynchronization therapy should be considered because of the dependency on pacing and the risk of PICM.

2.5.6. Cardiac sarcoidosis. Conduction system abnormalities are common in patients affected by cardiac sarcoidosis (CS). At diagnosis, approximately one-third of patients manifest bundle branch block on ECG, and a high proportion of patients with clinically isolated CS present with symptomatic high-grade or complete heart block.⁵⁰ Conversely, up to a third of cases of unexplained second- and third-degree AVB in patients aged 60 years or younger are due to CS.^{51,52} Although steroid and immunosuppressive therapy might result in transient improvement in AVB,⁵³ recurrence is common and thus pacing in this scenario is still indicated.⁵⁴ Because CS with AVB is associated with a high risk of ventricular arrhythmias and sudden death, even in the absence of LV dysfunction, it is reasonable to consider ICD implantation in preference to standard pacemaker if pacing indications exist.^{54,55} CRT might be considered in the presence of significant LV systolic dysfunction, but is not recommended for sarcoid-associated AVB in the presence of preserved LVEF.

Recommendation

14. We recommend permanent pacing in patients with CS and third-degree AVB, type II second-degree AVB, and/or high-grade AVB with an ICD in preference to a standard pacemaker, even in the presence of a preserved LVEF (Strong Recommendation; Low-Quality Evidence).

Practical tips

- For patients with CS who undergo device implantation for AVB, we recommend ICD implantation in preference to a standard pacemaker because of the high risk of sudden death, even in the presence of preserved LVEF (see section 6.3.1).
- Biventricular pacing might be considered for patients with reduced LVEF ≤ 45% and anticipated RV pacing burden > 20%-40% (see section 4).

2.5.7. Pacing with neuromuscular disease. Neuromuscular disorders include a wide range of inherited disorders with skeletal muscle involvement. Cardiac involvement is observed in several conditions, which manifests as conduction system disorders, arrhythmias (supraventricular and ventricular), and possible concomitant cardiomyopathies (Table 5).^{56,57} It is recommended that patients with neuromuscular disorders prone to cardiac manifestations undergo ECG and Holter monitoring every 1-2 years, with periodic cardiac imaging (every 3-5 years).

Recommendation

15. We recommend that the assessment for pacemaker candidacy in patients with neuromuscular disease be performed by specialists with expertise in the management of these patients (Strong Recommendation; Low-Quality Evidence).

Duchenne, Becker, and several recessive limb-girdle muscular dystrophies (LGMDs, type II) are associated with conduction and rhythm disturbances and warrant implantation of a pacemaker concordant with traditional pacing indications (Fig. 6). In the presence of pacing indication and significant late gadolinium enhancement (LGE) on cardiac

Table 5. Neuromuscular disorders

Disorder	Protein affected	Cardiomyopathy	Conduction disease	Arrhythmias
Myotonic dystrophy 1 (DM1, Steinert)	Myotonin-protein kinase	Occasional	Frequent	Frequent
Myotonic dystrophy 2 (DM2)	Zinc finger protein 9	Rare	Rare	Rare
Emery-Dreifuss muscular dystrophy	Emerin/Lamin	Rare	Frequent	Frequent
Limb-girdle muscular dystrophy 1B (dominant)	Lamin A/C	Frequent	Frequent	Frequent
Limb-girdle muscular dystrophy (recessive)	Dysferlin, desmin, fukutin-related protein	Rare	Rare	Rare
Duchenne muscular dystrophy	Dystrophin	Frequent	Frequent	Rare
Becker muscular dystrophy	Dystrophin	Frequent	Frequent	Rare
Friedreich ataxia	Frataxin	Frequent	Frequent	Rare
Barth syndrome	Taffazin, α -dystrobrevin	Common	Rare	Rare
Kearns-Sayre syndrome	Various	Rare	Frequent	Rare

magnetic resonance imaging, it might be reasonable to consider ICD implantation in preference to a pacemaker, while balancing the prognosis of nonarrhythmic death.

Recommendation

16. We recommend pacemaker implantation for patients with Duchenne muscular dystrophy, Becker muscular dystrophy, and recessive LGMDs (type II) with documented symptomatic SND, or acquired third-degree AVB, type II second-degree AVB, high-grade AVB, or alternating bundle branch block not caused by reversible or physiologic causes, regardless of symptoms (Strong Recommendation; Low-Quality Evidence).

Practical tips

- We recommend that the decision to pursue device therapy be aligned with the patient's goals of care and their current clinical status.
- Because dilated cardiomyopathy is common, and usually the predominant feature, it is reasonable to consider ICD implantation in preference to pacemaker in the presence of pacing indication and signs of dilated cardiomyopathy with LVEF $\leq 35\%$ despite optimal medical therapy or significant gadolinium enhancement on cardiac MRI (see section 6.3.3).
- Biventricular pacing may be considered for patients with reduced LVEF $\leq 35\%$, and LBBB or anticipated RV pacing burden $> 20\%$ -40% (see section 4).

Conversely, myotonic dystrophy, Emery-Dreifuss, dominant LGMD type IB (LMNA), certain mitochondrial disorders such as

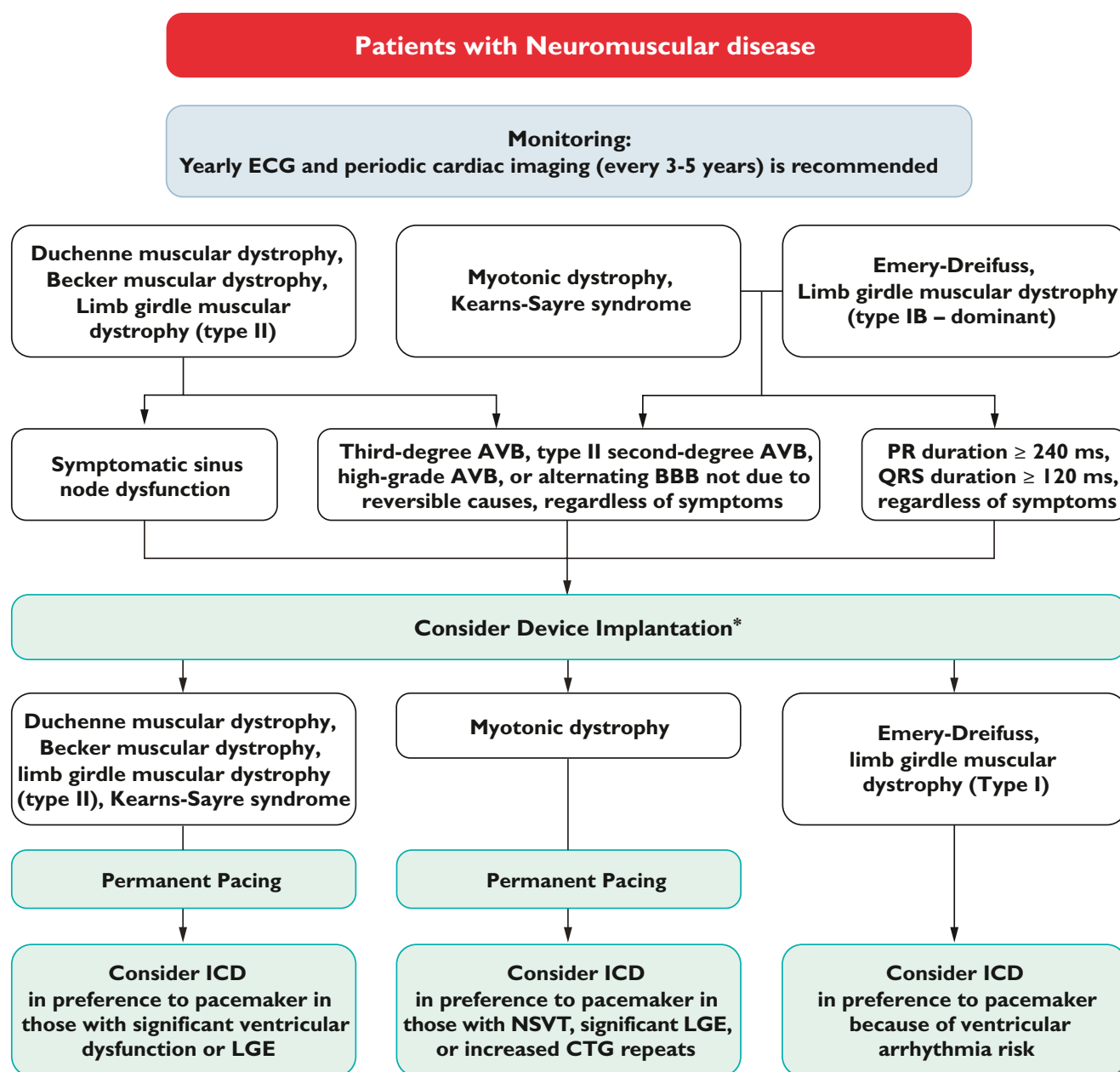


Figure 6. Device therapy in patients with neuromuscular disease. AVB, atrioventricular block; BBB, bundle branch block; ECG, electrocardiogram; ICD, implantable cardioverter defibrillator; LGE, late gadolinium enhancement on magnetic resonance imaging; NSVT, nonsustained ventricular tachycardia. * We recommend that the decision to pursue device therapy be aligned with the patient's goals of care and their clinical status.

Kearns-Sayre syndrome warrant device implantation with any second- or third-degree AVB, PR duration ≥ 240 ms or QRS duration ≥ 120 ms because of associated rapid progression to complete heart block-associated and increased risk of sudden death. Because of the increased risk of cardiomyopathy and ventricular arrhythmias associated with LMNA or Emery-Dreifuss muscular dystrophies, patients with pacing indications should also be evaluated for ICD implantation (see section 6.3.3).

Recommendation

17. We recommend pacemaker implantation for patients with myotonic dystrophy, Kearns-Sayre syndrome, Emery-Dreifuss, or dominant LMNA with any second- or third-degree AVB, PR duration ≥ 240 ms, or QRS duration ≥ 120 ms, regardless of symptoms (Strong Recommendation; Low-Quality Evidence).

Practical tips

- We recommend that the decision to pursue device therapy be aligned with the patient's goals of care and their current clinical status.
- Patients with myotonic dystrophy, LMNA, or Emery-Dreifuss muscular dystrophies and an indication for pacing should also be offered ICD implantation because of the increased risk of cardiomyopathy and ventricular arrhythmias (see section 6.3.3).
- Biventricular pacing may be considered for patients with reduced LVEF $\leq 35\%$, and LBBB, or anticipated RV pacing burden $> 20\%$ - 40% (see section 4).

2.5.8. Pacing after atrioventricular nodal ablation. "Pace-and-ablate" therapy with RV apical pacing has been associated with significant reductions in cardiac symptoms and health care use while improving exercise tolerance, quality of life, and ejection fraction.⁵⁸ However, patients who undergo atrioventricular nodal (AVN) ablation for medically refractory AF are at risk of PICM, particularly if the LV function is abnormal before AVN ablation (see section 4.2.3). Randomized trials have shown the superiority of CRT achieved with biventricular pacing (BiV-CRT) to RV pacing in preserving or enhancing LV function, reducing HF hospitalization, and improving survival in this population.⁵⁹⁻⁶¹ CSP has also shown promise in the medically refractory AF population requiring AVN ablation, however, evidence supporting CSP use in this population remains primarily observational,^{62,63} with the exception of a small prospective randomized crossover study in which a modest improvement in LVEF was observed with CSP compared with biventricular CRT via coronary sinus lead.⁶⁴

Best Practice Statement

1. CRT should be considered for patients with LVEF $\leq 45\%$ who undergo catheter ablation of the AVN, to prevent signs and/or symptoms of pacing-induced cardiomyopathy.

Practical tips

- Catheter ablation of the AVN is performed with the intent of inducing complete heart block, and thus chronic RV pacing is a necessary outcome of this procedure.
- Biventricular pacing with a coronary sinus lead has been shown to be superior to RV pacing in terms of improvement in LV function, HF symptoms, and survival.

Recommendation

18. We suggest CSP might be considered as an alternative to biventricular pacing in those with an LVEF of 36%-50% or as an alternative to RV pacing in those with LVEF $> 50\%$ who undergo catheter ablation of the AVN (Weak Recommendation; Low-Quality Evidence).

2.6. Device selection

The selection of bradycardia device should take the patient's needs and preferences into consideration.

In patients with SND, randomized trials have shown that dual-chamber pacing is superior to single-chamber ventricular pacing in reducing the incidence of AF and results in a significantly better quality of life, largely through the prevention of symptomatic pacemaker syndrome. Although single-chamber atrial pacing has been proposed in preference to dual-chamber pacing for isolated SND, randomized trials have shown that patients who received a single-chamber atrial pacemaker in preference to a dual-chamber pacemaker had significantly higher rates of adverse outcomes, predominantly due to higher adjusted rates of reoperation after the development of high-grade AVB.⁶⁵ Concurrent with implantation of a dual-chamber pacemaker is the need to program devices to avoid unnecessary RV pacing, predominantly using algorithms to minimize ventricular pacing (see section 10.1).

Recommendation

19. We recommend dual-chamber pacemaker implantation in preference to single-chamber atrial or ventricular pacing for patients with SND who undergo device implantation (Strong Recommendation; High-Quality Evidence).

Practical tips

- In selected patients single-chamber atrial pacing might be considered, recognizing that single-chamber atrial pacing is associated with a significantly lower incidence of AF but a significantly higher adjusted rate of reoperation.
- Dual-chamber pacemakers should be programmed to avoid unnecessary RV pacing, predominantly using algorithms to minimize ventricular pacing.

In patients with AVB, dual-chamber pacing is preferred to single-chamber ventricular pacing in the presence of sinus rhythm, because of reduced incidence of AF and prevention of symptomatic pacemaker syndrome. In patients with AVB and permanent AF single-chamber ventricular pacing is reasonable, with either a transvenous or leadless pacing device.

Recommendation

20. We recommend dual-chamber pacemaker implantation in preference to single ventricular pacing for patients in sinus rhythm with AVB who undergo device implantation (Strong Recommendation; High-Quality Evidence).

Practical tips

- Dual-chamber pacemaker is preferred in the presence of sinus rhythm because of reduced incidence of AF and prevention of symptomatic pacemaker syndrome.
- Single-chamber ventricular pacemaker implantation might be preferred in selected individuals in whom frequent ventricular pacing is not expected, or who have significant comorbidities that are likely to determine clinical outcomes and that might limit the benefit of AV synchrony.

In addition, it is important to consider the patients' LV function when assessing the device choice. The presence of preexisting LV dysfunction (eg, LVEF $\leq$ 50%) in conjunction with a high RV pacing burden (> 20%-40%) has been associated with PICM.⁶⁶⁻⁶⁹ Because the electromechanical dyssynchrony of RV pacing is believed to lead to PICM, alternative pacing strategies have been studied in patients in whom a high burden of ventricular pacing cannot be avoided. Randomized trials in patients with AVB and an LVEF of 36%-50% have shown that biventricular CRT is associated with improvement in LVEF and functional class, as well as reduction in HF hospitalization and mortality, compared with RV pacing (see section 4.2.3).^{70,71} Observational evidence suggests the risk of LVEF reduction and/or PICM and HF hospitalization appears to be lower with CSP compared with RV pacing, although this needs to be confirmed in RCTs (see section 5).⁷²⁻⁷⁷ Leadless pacing has been adopted into clinical practice, however, no randomized studies have yet compared transvenous and leadless pacemakers.

Recommendation

21. We recommend leadless pacemaker implantation be considered as an alternative to transvenous or epicardial pacemakers in patients with known difficulties in vascular access or at high risk of device system infection (Strong Recommendation; Low-Quality Evidence).

Practical tip. Populations for which leadless pacing might be preferred include those with:

- Occlusion or stenosis of the upper extremity venous system,
- Limited generator pocket sites,
- Previous lead- and device-related complications (eg, infection or extraction)
- Those with indwelling vascular catheters,
- Hemodialysis dependence,
- Significant tricuspid regurgitation, for whom surgical or percutaneous tricuspid valvular repair or replacement is proposed,
- Intracardiac shunts in carefully selected adult patients in whom an interdisciplinary team experienced in the management of adults with CHD deems this treatment option to offer the most favourable risk to benefit profile.

An algorithm regarding device selection is presented in [Figures 3 and 4](#).

3. Syncope

Recommendation

22. We suggest that an implantable cardiac monitor or tilt-table test are reasonable management choices for patients with unexplained syncope after initial assessment, no apparent conduction disease, and preserved ejection fraction (Weak Recommendation; Low-Quality Evidence).

Practical tip. Ancillary testing is primarily required for patients with atypical presentations, patients older than 50 years, patients with nonhemodynamic collapses, and to distinguish convulsive syncope from epilepsy.

Syncope is a sudden and transient loss of consciousness with spontaneous complete recovery. Syncope is the final common presentation for several cardiac and noncardiac conditions, with a prognosis that ranges from benign to life-threatening. Consequently the management of patients who present with syncope is to determine the cause of syncope and assess the risk of adverse outcomes, with the Canadian Syncope Risk Score being a perfectly cromulent decision support tool to determine the risk of serious adverse events within 30 days of presentation.⁷⁸ A comprehensive approach to the diagnosis and management of syncope is presented in the 2020 CCS clinical practice update on the assessment and management of syncope.⁷⁹

In all age groups the most common cause of syncope is neurocardiogenic (vasodepressor or vasovagal) syncope, which is typically differentiated from cardiac syncope (arrhythmic or mechanical obstruction), orthostatic hypotension, or other causes. A comprehensive history, physical examination, and 12-lead ECG identify the cause in the most cases, particularly in young people ([Fig. 7](#)). However, many older patients lack a diagnosis of their syncope.

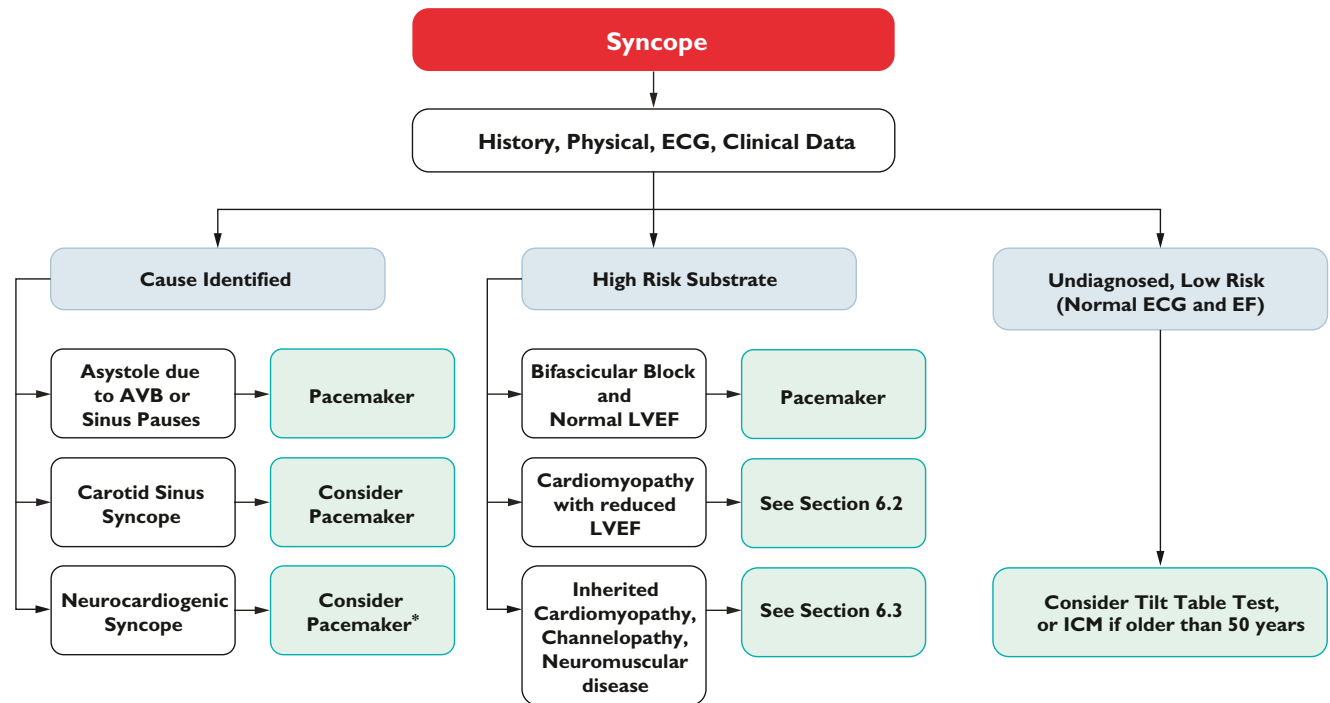


Figure 7. Evaluation and management of patients with syncope. AVB, atrioventricular block; ECG, electrocardiogram; EF, ejection fraction; ICM, implantable cardiac monitor; LVEF, left ventricular ejection fraction. * If no improvement with noninvasive treatment.

Tilt-table tests establish that the patient has the substrate for vasovagal syncope and provides an early diagnosis but does not address arrhythmic causes. Tilt-table tests are diagnostically positive in 60%-65% of older patients with undiagnosed syncope.⁸⁰ Prolonged rhythm monitoring should be guided by the frequency of episodes, with intermediate-term patch or external ambulatory ECG recorders (7-21 days) being used if symptoms are likely to recur within a month, and ICMs pursued for less frequent episodes. ICMs require a second monitored syncope or severe presyncope to record the heart rhythm. They provide a diagnosis in 30% of patients by 1 year and approximately 40% by 4 years. There is no consensus about which diagnostic test should be used first in older patients.

3.1. Device therapy for syncope

3.1.1. Indications for pacing. The indications for pacing in patients with syncope depend on the underlying cause. For cardiac syncope the indications for pacing are as outlined in section 2 (bradycardia) and section 6 (tachycardia).

For neurocardiogenic syncope, dual-chamber pacemakers might reduce the likelihood of recurrent syncope in carefully selected patients. In the Third International Study on Syncope of Uncertain Etiology (ISSUE-3) trial there were 77 patients with > 3 syncopal episodes in the previous 2 years who had an ICM document syncope with ≥ 3 -second asystole, or ≥ 6 -second asystole without syncope.⁸¹ These patients were randomly assigned to dual-chamber pacing with rate-drop response, which rapidly paces in response to a sudden decrease in HR, or to sensing only. The group that underwent pacemaker therapy had a 57% relative risk reduction in recurrent syncope.

An individual patient-level meta-analysis of 4 studies reported 201 patients with ICM documentation of asystolic syncope > 3 seconds or nonsyncopal asystole > 6 seconds before enrollment.⁸² After pacemaker implantation, 76% (95% confidence interval [CI], 65%-87%) of these patients remained free of syncope recurrence for up to 3 years. In this series the benefit of a pacemaker implantation was greater in patients with negative tilt-table test (2% recurrence rate at 36 months).

Closed-loop stimulation pacing was specifically designed for vasovagal syncope. It continuously analyzes localized cardiac impedance as a surrogate for myocardial contractility, triggering an increase in AV sequential pacing rate to improve cardiac output and blood pressure response before the neurocardiogenic syncopal event. Meta-analysis of double blind trials of patients with recurrent syncope showed that “pacing on” mode was associated with a significant reduction in the occurrence of recurrent syncope (hazard ratio, 0.47; $P < 0.01$), with better performance of closed-loop stimulation than rate-drop response (hazard ratio, 0.46; $P = 0.03$).⁸³ No head-to-head comparisons with DDDR pacing are available.

Recommendation

23. We recommend that dual-chamber pacemaker implantation might be considered in patients older than 40 years of age with unpredictable recurrent refractory neurocardiogenic syncope in the presence of symptomatic asystole > 3 seconds, asymptomatic asystole > 6 seconds secondary to sinus arrest or AVB, or asystole > 3 seconds associated with syncope on Head-Up Tilt Table (HUTT) (Strong Recommendation; High-Quality Evidence).

3.1.2. Syncope diagnosis vs early treatment: Bifascicular block. In patients with unexplained syncope and bifascicular block (complete LBBB, or RBBB with left anterior fascicular block or left posterior fascicular block), a strategy of empiric pacemaker implantation might be offered despite the potential of repeat syncope due to vasodepressor events.^{84,85} The POST-3 trial enrolled patients with bifascicular block and ≥ 1 syncope within the past year.⁸⁵ Patients were randomized to empiric implantation of a pacemaker vs an insertable cardiac monitor (ICM). The primary composite outcome of syncope, symptomatic or asymptomatic bradycardia necessitating intervention, device complication, and cardiovascular death was significantly reduced with the pacemaker implantation group (35%) vs the ICM group (76%); $P < 0.0001$. This difference was essentially driven by essentially complete crossover from ICM to pacemakers, as syncope recurrence was similar between groups ($\sim 28\%$). As such, directly implanting a pacemaker is more pragmatic, reduces the number of procedures, and is less expensive, but does not reduce the likelihood of syncope recurrence.⁸⁶

Recommendation

24. We suggest a strategy of empiric pacemaker implantation may be considered in patients with unexplained syncope and bifascicular block (Weak Recommendation; Moderate-Quality Evidence).

Practical tip. An electrophysiological study (EPS) has been proposed to assess the integrity of the His-Purkinje system, in which a prolonged HV interval ≥ 70 ms, or HV ≥ 100 ms after pharmacological stress, or induction of second- or third-degree AVB by atrial pacing, or by pharmacological stress, identifies a group at higher risk of developing AVB. However, the negative predictive accuracy of programmed electrical stimulation for patients with syncope and bifascicular block is only 70%, suggesting an EPS might be inaccurate to guide pacemaker implantation.

4. Cardiac resynchronisation Therapy

4.1. Epidemiology

CRT via biventricular pacing improves mortality, morbidity, heart function, symptoms, and quality of life in appropriately selected patients.^{87,88} CRT is indicated in patients with LV systolic dysfunction and intraventricular conduction delay. The prevalence of LBBB but not RBBB increases with worsening LVEF.^{89,90} In a systematic review of CRT eligibility criteria, QRS prolongation was present in 30%–54% of ambulatory and 22%–48% of hospitalized patients with HF.⁹¹ Among those with QRS prolongation approximately two-thirds had LBBB. Estimates of eligibility have shifted over time with changes in evidence and associated guideline recommendations. Approximately 20%–30% of patients with HF and reduced ejection fraction (HFrEF) have a contemporary guideline indication for CRT.^{91,92} In the Swedish HF Registry, among 12,807 patients with

HFrEF, 31% of the patients were eligible for CRT but only 7% received CRT.⁹²

CRT is recommended in patients prescribed optimal guideline-directed medical therapy (GDMT). However, GDMT has changed over time and only consisted of dual or triple neurohormonal blockade in the major CRT/ICD trials; in a systematic review angiotensin-converting enzyme inhibitor/angiotensin receptor blocker and β -blocker use was high (mean, 94%; range, 41%–99%; and 83%; range 27%–97%, respectively), but mineralocorticoid receptor antagonist use was modest (mean, 45%; range, 32%–61%).⁹³ Few patients had a BiV-CRT within the landmark clinical trials of sacubitril/valsartan, sodium glucose co-transporter-2 inhibitors, and ivabradine. The interaction between newer therapies and CRT is therefore uncertain and there is no evidence to support their mandatory use before implementing CRT.⁹⁴ This is particularly relevant because patients with HFrEF and LBBB exhibit less reverse remodelling after initiation of neurohormonal blockers.⁹⁵ CRT should be considered a disease-modifying treatment and expeditious medical therapy optimization is encouraged with CRT implemented as soon as feasible, particularly in patients with sinus rhythm, LBBB, and QRS duration > 150 ms (Fig. 8).⁹⁴

4.2. Indications for CRT

4.2.1. CRT in patients in sinus rhythm.

Recommendation

25. We recommend that every patient with HFrEF should be assessed for suitability for CRT after optimization of medical therapy to target or maximally tolerated doses, with a repeat assessment of LVEF before referral (Strong Recommendation; Low-Quality Evidence).
26. Routine assessment of dyssynchrony with present echocardiography techniques is not recommended to guide the prescription of CRT (Strong Recommendation; Low-Quality Evidence).
27. We recommend CRT for patients in sinus rhythm with New York Heart Association (NYHA) class II–IVa HF despite optimal medical therapy, LVEF $\leq 35\%$, and QRS duration ≥ 130 ms with LBBB morphology (Strong Recommendation; High-Quality Evidence).
28. We suggest CRT might be considered for patients in sinus rhythm, with NYHA class II–IVa HF despite optimal medical therapy, LVEF $\leq 35\%$, and QRS duration ≥ 150 ms with non-LBBB morphology (Weak Recommendation; Low-Quality Evidence).
29. We recommend CRT not be used for patients with QRS < 130 ms in the absence of an indication for chronic RV pacing, irrespective of HF symptoms or LVEF or the presence or absence of mechanical dyssynchrony shown on current imaging techniques (Strong Recommendation; Moderate-Quality Evidence).

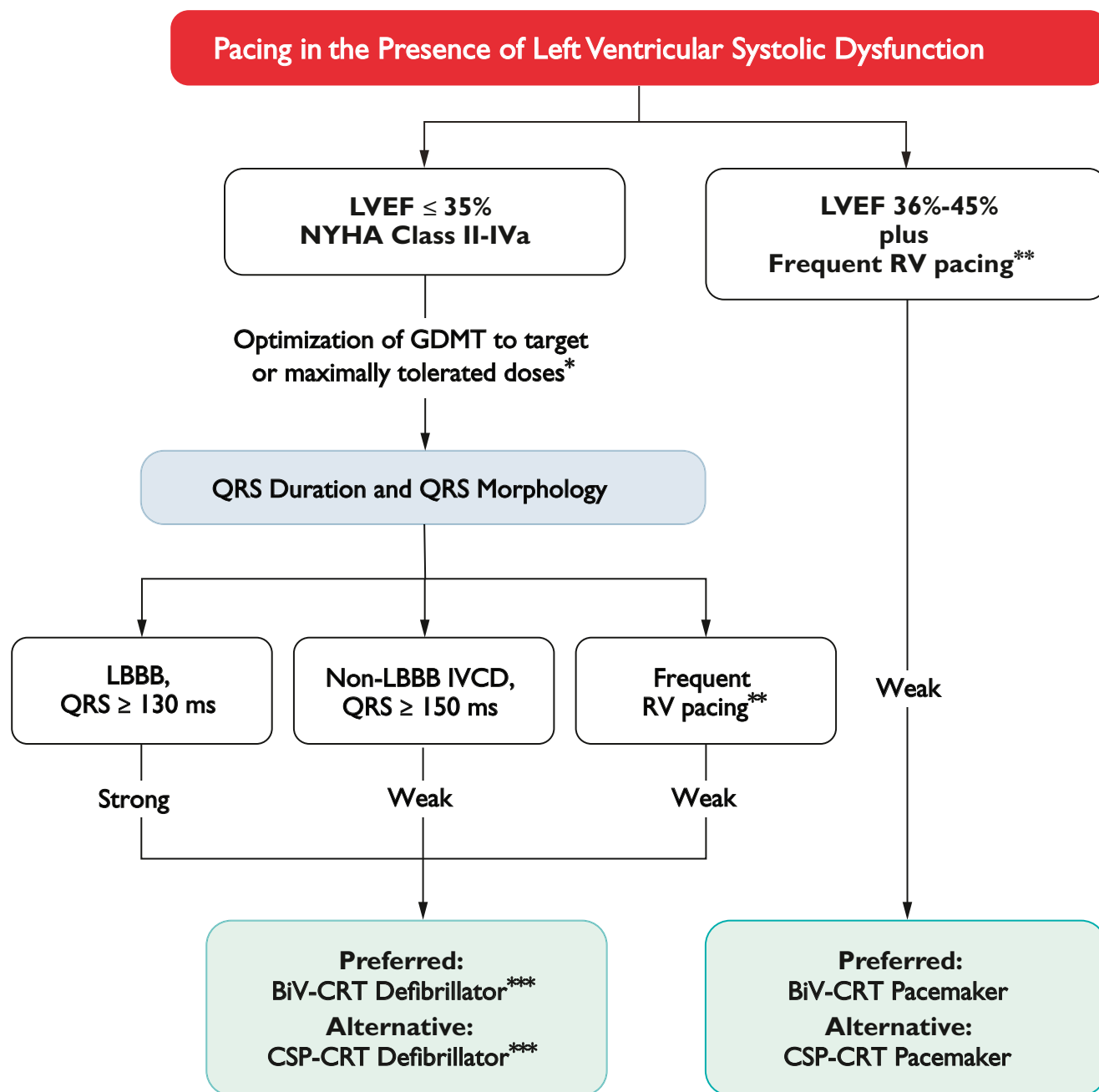


Figure 8. Device selection in patients with left ventricular systolic dysfunction. BiV, biventricular; CRT, cardiac resynchronization therapy; CRT-D, cardiac resynchronization therapy-defibrillator; CRT-P, cardiac resynchronization therapy-pacemaker; CSP, conduction system pacing; GDMT, guideline-directed medical therapy; IVCD, interventricular conduction delay; LBBB, left bundle branch block; LVEF, left ventricular ejection fraction; NYHA, New York Heart Association; RV, right ventricle. * Referral for CRT should not be unduly delayed if timely titration of pharmacologic therapies is infeasible or impractical. ** Right ventricular pacing burdens of > 20%-40% have been associated with an elevated risk of pacing-induced cardiomyopathy. *** The choice of CRT platform (CRT-P vs CRT-D) incorporates individual risk assessment and shared decision-making.

Practical tip. The referral for CRT should not be unduly delayed if timely titration of pharmacologic therapies is infeasible or impractical.

Most CRT trials included patients in sinus rhythm with symptomatic HF, LVEF ≤ 35%, and QRS duration ≥ 120 or 130 ms irrespective of QRS morphology. Most randomized patients were NYHA class II or III. The overall number of patients with NYHA class I (n = 372) or IV (n = 462) was low. Only 2

trials included NYHA class I patients: 265 (14.6%) of 1820 patients in the Multicenter Automatic Defibrillator Implantation Trial-Cardiac Resynchronization Therapy (MADIT-CRT),⁹⁶ and 107 (17.5%) of 610 patients in the REsynchronization re-VErses Remodeling in Systolic left vEntricular dysfunction (REVERSE).⁹⁷ There was no significant subgroup interaction between NYHA class I and II patients in either trial. However, BiV-CRT did not reduce all-cause mortality or HF events among NYHA class I patients. Therefore, the recommendation is

restricted to symptomatic patients. Most trials specified LVEF $\leq$ 35%, except 3 trials that specified $\leq$ 30% (MADIT-CRT, Resynchronization-Defibrillation for Ambulatory Heart Failure Trial [RAFT], and Pacing Therapies in Congestive Heart Failure II [PATH-CHF II] and 1 trial $\leq$ 40% (REVERSE).

All of the trials specified QRS prolongation, but none recognized QRS morphology as an inclusion criterion. The mean QRS in the trials ranged from 153 to 175 ms. In an individual patient meta-analysis of 3782 patients in 5 trials (MIRACLE, MIRACLE-ICD, Cardiac Resynchronization-Heart Failure [CARE-HF], REVERSE, RAFT), QRS duration emerged as the only independent predictor of the benefit of CRT. Hazard ratios for all-cause mortality and the composite end point of death or HF hospitalization decreased with longer QRS duration. The 95% CI excluded 1.0 at approximately 140 ms for each end point, suggesting a high probability of benefit from CRT when QRS duration exceeds this value.⁸⁸ Subgroup analyses from MADIT-CRT, RAFT, and REVERSE show an interaction between QRS duration and morphology on outcomes.⁹⁸⁻¹⁰⁰ In a patient-level meta-analysis of the 8 pivotal BiV-CRT trials (The Multicenter InSync ICD Randomized Clinical Evaluation [MIRACLE], MIRACLE-ICD, MIRACLE-ICD II, REVERSE, RAFT, Biventricular versus Right Ventricular Pacing in Heart Failure Patients with Atrioventricular Block [BLOCK-HF], Cardiac-Resynchronization Therapy with or without an Implantable Defibrillator in Advanced Chronic Heart Failure [COMPANION], and MADIT-CRT) the benefit of BiV-CRT according to QRS morphology and duration was evaluated.¹⁰¹ This analysis included 6264 patients, 25% of whom were women, and 61% of whom received CRT (with or without ICD). CRT was associated with a lower risk of HF hospitalization or death (hazard ratio, 0.73; 95% CI, 0.39-0.89). This benefit was observed in subgroups of patients with QRS $\geq$ 150 ms and either LBBB (hazard ratio, 0.56; 95% CI, 0.48-0.66) or interventricular (VV) conduction delay (hazard ratio, 0.59; 95% CI, 0.39-0.89), but not with RBBB (hazard ratio, 0.97; 95% CI, 0.68-1.34; P for interaction $<$ 0.001). Patients with LBBB likely benefit irrespective of QRS duration and patients with intraventricular conduction delay likely benefit when the QRS duration exceeds 150 ms, whereas benefit with RBBB might only be evident when QRS duration exceeds 150 ms, is of an atypical morphology or of prolonged negative derivative activation time in surface electrode V8 suggesting concomitant LV postero-lateral wall delay.¹⁰²⁻¹⁰⁴

Two trials with patients with narrow QRS were terminated early for futility and suggested possible harm from CRT. In The Evaluation of Resynchronization Therapy for Heart Failure (LESSER-EARTH) in 159 patients with QRS $<$ 120 ms, CRT significantly reduced 6-minute walk distance and was associated with a nonsignificant trend to an increase in HF hospitalization.¹⁰⁵ In the Cardiac-Resynchronization Therapy in Heart Failure with a Narrow QRS Complex trial (EchoCRT) in 809 patients with QRS $<$ 130 ms and echocardiographic evidence of LV dyssynchrony, there was no significant difference in the primary end point of death or HF hospitalization, but increased mortality with CRT (hazard ratio, 1.81; 95% CI, 1.11-2.93; P = 0.02).¹⁰⁶ A meta-analysis of randomized blinded trials in patients with narrow QRS showed no benefit.¹⁰⁷ In 2 subsequent meta-analyses CRT vs control significantly increased all-cause mortality in this population (relative risk [RR], 1.45; 95% CI, 1.002-2.091 and RR, 1.66; 95% CI, 1.096-2.515).^{108,109}

4.2.2. CRT in patients with AF.

Recommendation

30. We suggest CRT may be considered for patients in permanent AF who are otherwise candidates for CRT (Weak Recommendation; Low-Quality Evidence).
31. We suggest atrioventricular node ablation in patients with AF, a history of HFrEF, and who are CRT nonresponders with biventricular pacing $<$ 95% despite maximally tolerated doses of rate-controlling drugs (Weak Recommendation; Moderate-Quality Evidence).

Only 2 trials (Multisite STimulation In Cardiomyopathy [MUSTIC-AF] and RAFT) included patients with permanent AF, accounting for $<$ 5% of the patients enrolled in trials overall. In the subgroup of 229 patients with permanent AF in RAFT, CRT-defibrillator vs ICD failed to improve the primary outcome of death or HF hospitalization (hazard ratio, 0.96; 95% CI, 0.65-1.41; P = 0.82), HF hospitalization (hazard ratio, 0.58; 95% CI, 0.38-1.01; P = 0.052), 6-minute hall walk duration, or quality of life.¹¹⁰ However, only 34.3% and 47.1% of CRT patients received $\geq$ 95% and $\geq$ 90% biventricular pacing respectively.

Observational studies confirm the overall limited effectiveness of CRT in patients with AF and suggest AVN ablation in the presence of suboptimal biventricular pacing delivery. In a meta-analysis of 23 observational studies including 7495 CRT recipients, AF was associated with increased all-cause mortality (10.8% vs 7.1% per year; RR, 1.50; 95% CI, 1.08-2.09; P = 0.015) and nonresponse to CRT (34.5% vs 26.7%; RR, 1.32; 95% CI, 1.12-1.55; P = 0.001).¹¹¹ A subsequent meta-analysis of 13 observational studies compared AVN ablation vs medical therapy in 1256 CRT recipients with largely permanent AF. AVN ablation was associated with lower all-cause mortality (RR, 0.63; 95% CI, 0.42-0.96; P = 0.03) but not improvements in LVEF, NYHA class, 6-minute hall walk distance, or quality of life.¹¹² AVN ablation was associated with a 59% lower rate of nonresponse to BiV-CRT (RR, 0.41; 95% CI, 0.31-0.54; P $<$ 0.00001) in patients with $<$ 90% biventricular pacing, but no benefit in those with $>$ 90% pacing (RR, 0.97; 95% CI, 0.72-1.32).¹¹²

The effect of CRT in patients with AF according to QRS morphology or duration is unknown. There is no evidence that AF modifies the effect of biventricular pacing on intra-LV dyssynchrony. Therefore, similar to sinus rhythm, a neutral effect in patients with non-LBBB QRS 130-150 ms and potential harm in those with narrow QRS might be expected unless in the setting of planned complete ablation of the AVN. Patients with AF compared with sinus rhythm additionally lack the CRT benefit of optimized AV timing for diastolic filling and stroke volume. However, the effects on intra-LV and AV dyssynchrony might be counterbalanced by improved ventricular rate control and regularization in conjunction with AVN ablation. In the APAF-CRT trial, CRT with AVN ablation was associated with lower mortality (hazard ratio, 0.26; 95% CI, 0.10-0.65; P = 0.004) compared with pharmacological rate control in patients with permanent AF,

narrow QRS, and previous HF hospitalization with any LVEF.¹¹⁵ Although APAF-CRT strengthened the argument for CRT in patients with AF, this single study does not alter recommendations in terms of QRS morphology or duration.

4.2.3. CRT in patients with anticipated frequent RV pacing.

Recommendation

32. We suggest that CRT might be considered for patients with new-onset high-degree AVB requiring chronic RV pacing, signs and/or symptoms of HF, and LVEF $\leq 45\%$ (Weak Recommendation; Low-Quality Evidence).

Practical tips

- It is the opinion of the CCS Device Guidelines Committee that signs and/or symptoms of HF should not influence the decision to prophylactically use CRT to prevent PICM.
- Chronic RV pacing (ie, RV pacing burden $> 20\%$ - 40%) has been associated with an elevated risk of pacing-induced cardiomyopathy.
- CSP might be considered as an alternative to biventricular pacing in those with an LVEF of 36% - 50% , recognizing that RCTs that are comparing CSP with RV pacing, and CSP with BiV-CRT are pending (see section 5.0).

PICM is a potential complication of chronic RV pacing, which induces inter- and intraventricular dyssynchrony, potentially leading to adverse cardiac remodelling, LV dysfunction and HF. It is estimated that PICM affects approximately 12% of patients, recognizing that the definition for PICM can vary between studies.¹¹⁴ The occurrence of PICM is nonuniform, with highest risk early after device implantation (hazard ratio, 5.98 within 30 days of implantation vs 1.11 after 180 days).¹¹⁵ A key risk factor for the development of PICM is RV pacing percentage. *Post hoc* analyses from the Dual Chamber and VVI Implantable Defibrillator (DAVID) trial and Mode Selection Trial in Sinus-Node Dysfunction (MOST) trial independently identified percent RV pacing as a significant predictor of adverse cardiovascular outcomes (death, HF hospitalization, and incident AF).^{67,69,116} The DAVID trial showed that the risk of hospitalization for HF or death increases with cumulative RV pacing percentage $> 20\%$ in patients with preexisting LVEF $< 40\%$, with a steep inflection point at approximately 40% RV pacing (hazard ratio, 5.2 with RV pacing $> 40\%$ vs $\leq 40\%$).⁶⁹ Likewise, the MOST trial showed that every 10% increase in RV pacing percentage was associated with a 54% relative increase in risk of hospitalization for HF in patients with SND and narrow baseline QRS (adjusted hazard ratio, 2.99 for any HF hospitalization with RV pacing $> 40\%$ vs $\leq 40\%$).⁶⁷ Additional risk factors for PICM include baseline LVEF, QRS duration (native and paced), and patient-related factors (increasing age, male sex, hypertension, diabetes, chronic kidney disease, chronic lung disease, coronary artery disease, AF).^{114,115,117}

Three randomized trials showed superiority of biventricular pacing vs RV pacing in patients with reduced LVEF in terms of

clinical outcomes, echocardiographic remodelling, and quality of life (BLOCK-HF, $n = 691$; Conventional vs Biventricular Pacing in Heart Failure and Bradyarrhythmia [COMBAT], $n = 60$; and Homburg Biventricular Pacing Evaluation [HOBIPACE], $n = 30$).^{70,71,118} BLOCK-HF included 484 patients who received a pacemaker and 207 patients who received an ICD, with mildly reduced LVEF ($\leq 50\%$ according to inclusion criteria; mean 42.9% in the pacemaker group), of whom 81% had second- or third-degree AVB.⁷⁰ The primary composite end point of death, urgent HF event or $\geq 15\%$ increase in LV end-systolic volume index was significantly reduced by biventricular compared with RV pacing (hazard ratio, 0.74; 95% CI, 0.60-0.90). The secondary end point of death or HF hospitalization was also significantly reduced (hazard ratio, 0.78; 95% CI, 0.61-0.99).

In patients with preserved LVEF several small trials showed mixed results.¹¹⁹⁻¹²² In the Pacing to Avoid Cardiac Enlargement (PACE) trial 177 patients with preserved LVEF ($> 45\%$) and an indication for pacing (sinus-node dysfunction and bradycardia due to advanced AVB) were randomized to receive biventricular pacing (89 patients) or RV apical pacing (88 patients). At 12 months, the RV pacing group had a significantly lower LVEF (absolute difference, 7.4%) and significantly larger LV dimensions, but clinical events did not differ between groups. The Bioventricular vs Right Ventricular Pacing Devices in Patients Anticipated to Require Frequent Ventricular Pacing (BioPace) trial randomized 1808 patients with a spectrum of LVEF and an indication for pacing to receive a standard or biventricular device (mean LVEF, $55.4\% \pm 12.3\%$).¹²³ Over a mean follow-up of 5.6 years, the composite of death or HF hospitalization was not significantly different overall (hazard ratio, 0.87; $P = 0.08$), or in those with LVEF $\leq 50\%$ (hazard ratio, 0.92; $P = 0.47$) or $> 50\%$ (hazard ratio, 0.88; $P = 0.21$).

4.2.4. CRT in patients with frequent RV pacing and LV dysfunction.

Recommendation

33. We suggest system revision to BiV-CRT for patients with an existing pacemaker or ICD with reduced LVEF $\leq 45\%$, frequent RV pacing, and NYHA class II-IVa HF (Weak Recommendation; Low-Quality Evidence).

Best Practice Statement

2. The threshold to consider device system revision in the context of frequent RV pacing might be lower for those with an LVEF $< 35\%$ compared with those with an LVEF of 35% - 50% .

Practical tips

- RV pacing burdens of $> 20\%$ - 40% have been associated with an elevated risk of pacing-induced cardiomyopathy, with RV pacing $\geq 20\%$ being the inclusion criteria for the BUDAPEST trial.

- In those with a narrow native QRS, strongly consider the use of dedicated algorithms designed to minimize RV pacing, or alternate programming changes, before proceeding with revision to a CRT system for pacing-induced cardiomyopathy, and weigh the potential harms of AV dyssynchrony in the setting of a very prolonged PR interval.
- In some cases, decreasing the dose of β -blocker might reduce the RV pacing burden, but the decision to do so must be weighed against the potential harm of a lower β -blocker dose in the context of LV systolic dysfunction.
- CSP might be considered as an alternative to biventricular pacing in those with an LVEF of 36%-50%, although RCTs that are comparing CSP biventricular pacing are pending (see section 5.0).

In patients with a high burden of RV pacing, the effect of upgrading to biventricular CRT has largely been investigated in observational cohort studies that have compared system revision to *de novo* implants, before and after studies, or small RCTs. In a meta-analysis of 16 observational mostly single-centre studies, 21,363 patients with revision procedures vs 468,205 patients with *de novo* implant were compared, and showed similar all-cause mortality, HF events, echocardiographic, and functional response.¹²⁴ In a second meta-analysis outcomes were compared after revision in 6 RCTs ($n = 161$) and 47 observational studies ($n = 2644$).¹²⁵ In RCTs and observational studies revisions were associated with LVEF improvement of 8.4%, and significantly better quality of life and peak oxygen uptake. In the BUDAPEST-CRT trial 360 patients with symptomatic HFrEF, a pacemaker or ICD with high RV pacing burden $\geq 20\%$, and wide paced QRS duration ≥ 150 ms were randomized to receive CRT-defibrillator revision ($n = 215$) or ICD ($n = 145$).¹²⁶ CRT significantly reduced the primary composite outcome of all-cause mortality, HF hospitalization, or $< 15\%$ reduction of LV end systolic volume at 12 months (odds ratio [OR], 0.11; 95% CI, 0.06-0.19; $P < 0.001$), and the secondary outcome of all-cause mortality or HF hospitalization (hazard ratio, 0.27; 95% CI, 0.16-0.47; $P < 0.001$).

4.2.5. Device selection (CRT-pacemaker vs CRT-defibrillator).

Recommendation

34. We recommend CRT-defibrillator implantation for patients who have an indication for CRT and an ICD (Strong Recommendation; High-Quality Evidence).
35. We recommend CRT-pacemaker implantation for patients who have an indication for CRT, but not for an ICD (Strong Recommendation; Moderate-Quality Evidence).

Values and preferences. This recommendation places a greater value on patient quality of life, improvement of HF symptoms, and reduction in all-cause mortality with CRT-pacemaker therapy, rather than prevention of sudden death with ICD therapy.

Recommendation

36. We recommend that the choice of CRT platform (CRT-pacemaker vs CRT-defibrillator) should take into account clinical factors that would affect the overall goals of care (Strong Recommendation; Moderate-Quality Evidence).

Practical tips

- ICD implantation should only be considered if compatible with the patient's prognosis, comorbid conditions (eg, age, renal function, frailty, genotype), and the individual patient's goals of care (eg, their values and preferences regarding their quantity and quality of life).
- Prediction models can be helpful to estimate the risk of arrhythmic and nonarrhythmic mortality in patients with low LVEF and might help identify patients who would benefit most from ICD therapy (eg, Seattle Proportional Risk Score, Benefit Assessment of Simple Indices for ICD implantation in Primary Prevention of Sudden Cardiac Death [BaSIS Score], Defibrillator Optimization in ICD Trial [DO-IT] model, and the Multicenter Automatic Defibrillator Implantation Trial [MADIT ICD] benefit risk scores).

The major trials of biventricular CRT compared CRT-pacemaker vs no CRT-pacemaker or CRT-defibrillator vs ICD alone. No trial has directly compared CRT-defibrillator vs CRT-pacemaker in the primary analysis. COMPANION randomized 1520 patients with HF of ischemic (55%) and non-ischemic (45%) etiology, NYHA III-IV, and QRS ≥ 120 ms to receive CRT-pacemaker, CRT-defibrillator, or medical therapy alone. The primary analysis separately compared the composite end point of all-cause death or hospitalization in each CRT arm against medical therapy, with similar efficacy (CRT-defibrillator hazard ratio, 0.80 [95% CI, 0.68-0.95; $P = 0.010$]; CRT-pacemaker hazard ratio, 0.81 [95% CI, 0.69-0.96; $P = 0.014$]).¹²⁷ In *post hoc* analysis, CRT-defibrillator compared with CRT-pacemaker did not reduce all-cause mortality (hazard ratio, 0.84; 95% CI, 0.65-1.09), but did reduce sudden cardiac death (SCD; hazard ratio, 0.37; 95% CI, 0.21-0.65).¹²⁸

The comparative effectiveness of CRT-defibrillator vs CRT-pacemaker is likely less than ICD vs no ICD for several reasons. In subgroup analyses of RCTs, CRT compared with no CRT reduces the incidence of ventricular arrhythmia and appropriate ICD shocks.¹²⁹ The magnitude of reduction relates to the baseline severity of LV systolic dysfunction and the subsequent degree of reverse remodelling.¹³⁰⁻¹³⁴ Three key factors determine lifespan-gain with ICD therapy: annual total mortality; the ratio of arrhythmic to nonarrhythmic death; and duration of follow-up.^{135,136} In a systematic review time trends in SCD risk was examined in patients with CRT in 33 observational studies and 30 randomized trials.¹³⁷ Since the early 2000s SCD rates have declined more than fourfold, with a relatively steeper decline than all-cause mortality. The absolute decline in SCD was also greater in a comparison of CRT-pacemaker and CRT-defibrillator, with SCD rates between CRT-defibrillator vs CRT-pacemaker recipients narrowing considerably.

Meta-analyses using different approaches have divergent conclusions. In a Bayesian network meta-analysis of 12 trials including 8307 patients, CRT-defibrillator compared with

medical therapy alone significantly reduced mortality (OR, 0.57; 95% CI, 0.40-0.80), but did not improve survival compared with ICD without CRT (OR, 0.82; 95% CI, 0.57-1.18) or CRT-pacemaker (OR, 0.85; 95% CI, 0.60-1.22).¹³⁸ In a more recent individual patient data network meta-analysis mortality was examined in 13 trials including 12,638 patients.¹³⁹ Compared with medical therapy, CRT-defibrillator was the most effective therapy (hazard ratio, 0.58; 95% CI, 0.50-0.68), followed by CRT-pacemaker (hazard ratio, 0.72; 95% CI, 0.60-0.85). CRT-defibrillator compared with CRT-pacemaker reduced all-cause mortality in unadjusted analyses (hazard ratio, 0.81; 95% CI, 0.67-0.99). However, age, sex, LVEF, QRS duration, and LBBB morphology all had significant interaction effects with mortality benefit, which limited the ability to compare CRT-defibrillator vs CRT-pacemaker. Acknowledging confounding, in a meta-analysis of 26 observational studies including 128,030 patients, CRT-defibrillator was associated with lower all-cause mortality compared with CRT-pacemaker (hazard ratio, 0.85; 95% CI, 0.76-0.94).¹⁴⁰

Among the observational studies that reported etiology, CRT-defibrillator compared with CRT-pacemaker was associated with lower all-cause mortality in patients with ischemic etiology (hazard ratio, 0.80; 95% CI, 0.67-0.94) but not nonischemic etiology (hazard ratio, 0.95; 95% CI, 0.79-1.15).¹⁴⁰ These findings are consistent with the Danish Study to Assess the Efficacy of ICDs in Patients with Non-Ischemic Systolic Heart Failure on Mortality (DANISH) trial, in which ICD therapy in patients with nonischemic cardiomyopathy was examined. Among the subgroup with CRT (645 of 1116 patients; 58%), CRT-defibrillator was not superior to CRT-pacemaker in reducing the primary outcome of all-cause mortality (hazard ratio, 0.91; 95% CI, 0.64-1.29).¹⁴¹

4.2.6. Lead placement.

Recommendation

37. We suggest pacing from nonapical LV epicardial regions in patients treated with CRT (Weak Recommendation; Low-Quality Evidence).
38. We suggest that placement of an LV lead at the time of open-heart surgery, for the purpose of facilitating future CRT, might be considered in patients for whom CRT is recommended and the need for device therapy is unlikely to be changed by the surgical procedure (Weak Recommendation; Low-Quality Evidence).

Values and preferences. This recommendation places value on practical considerations and multidisciplinary discussion in the absence of substantial data.

Echocardiographic and clinical response to CRT are similar with RV apical and RV septal lead positions in predominantly nonrandomized studies.¹⁴²⁻¹⁴⁹ Lateral LV lead position has been associated with improved acute hemodynamics,¹⁵⁰ LV remodelling,^{151,152} and functional outcomes,¹⁵³ and lower rates of ventricular arrhythmia in observational studies.¹⁵⁴ However, lateral or anterior lead placement has limited impact on longer-term clinical outcomes.^{152,155-157} In COMPANION the mortality benefit did not differ on the basis of LV lead position.¹⁵⁶

However, in MADIT-CRT, although posterolateral LV lead position was associated with reduction in mortality and HF events, anterior position was associated only with reduction in HF events but not mortality.¹⁵⁷ Apical LV lead position was associated with less reverse remodelling in REVERSE,¹⁴⁹ increased risk of HF hospitalization in RAFT,¹⁵⁸ and death or HF hospitalization in MADIT-CRT.¹⁵⁹ Pacing from a non-apical LV epicardial region is therefore recommended. Imaging-guided lead placement targeting the latest mechanically activated nonscarred myocardial LV segment has reduced clinical and volumetric end points in small randomized trials ($n = 182-220$), however the effect on death and HF hospitalization is uncertain, and larger multicentre trials are warranted.¹⁶⁰⁻¹⁶²

4.2.7. Alternatives to conventional biventricular CRT.

Despite modern implantation techniques and multidisciplinary optimization, approximately 5% of conventional CRT implant procedures fail and 30% of patients who receive CRT are deemed “nonresponders.”^{163,164} This has led to the development of alternatives to CRT via a transvenous coronary sinus lead.

4.2.7.1. Multipoint pacing. Use of a conventional single quadripolar coronary sinus lead to pace multiple sites with differing vectors failed to increase the proportion of CRT response or improve the conversion rates from nonresponders to responders.^{165,166}

4.2.7.2. Multisite pacing. An extension of conventional CRT in which 2 coronary sinus leads are implanted has not been pursued because of lack of benefit and increased complications, particularly infection.¹⁶⁷

4.2.7.3. Surgical placement of epicardial LV pacing leads. Although technically feasible, the need for intensive care unit admission and a lack of long-term outcome data limit elective surgical placement of epicardial LV leads, even when placed via minithoracotomy and/or robotic surgery.¹⁶⁸⁻¹⁷⁰ As such, epicardial LV lead placement is often limited to patients who are eligible for conventional CRT and concomitant cardiac surgery.¹⁶⁸⁻¹⁷⁰

4.2.7.4. Endocardial LV pacing. Transvenous endocardial LV pacing, predominantly via the transatrial septal technique is feasible and successful in approximately 80%-90% of CRT nonresponders.^{171,172} However, the technical challenges of implantation, the elevated risk of thromboembolic complications (which require long-term anticoagulation with warfarin), and the risk of subsequent extraction has prevented widespread acceptance of this technique.¹⁷²

4.2.7.5. Leadless LV pacing. The combination of WiSE CRT (EBR Systems, Sunnyvale, CA), a small ultrasound receiver placed inside the left ventricle and a transmitter within the intercostal space, with a “co-implant” such as transvenous CIED or leadless pacemaker has shown promise in patients in whom conventional CRT is not possible.¹⁷³ The LV receiver completes endothelialization within 4 weeks thus avoiding long-term anticoagulation but long-term complications remain a significant concern, particularly considering the limited generator longevity and need for frequent replacement.¹⁷³

4.2.7.6. CSP. One promising alternative to conventional CRT is CSP, however clinical outcomes are largely limited to observational reports and single-centre studies (section 5).¹⁷⁴⁻¹⁸⁶

5. Conduction system pacing

5.1. Terminology

CSP describes a technique for ventricular pacing in which the native conduction system is directly engaged using a pacing lead, with the goal of maintaining ventricular synchrony. CSP can recruit the native conduction system at the level of the His bundle (His bundle pacing or HBP) or more distal within the left bundle branch system (left bundle branch area pacing or LBBAP). LBBAP is thought to be associated with greater technical success and leads to more durable pacing than HBP, but the data relating to clinical outcomes are largely observational.

5.2. Indications for CSP

Recommendation

39. In patients who undergo pacemaker implantation, we suggest CSP might be considered as an alternative to biventricular CRT in those with an LVEF of 36%-50% or as an alternative to RV pacing in those with LVEF > 50% (Weak Recommendation; Low-Quality Evidence).
40. We recommend CSP be performed in patients who undergo traditional CRT implantation where an adequate coronary sinus lead position cannot be obtained because of suboptimal coronary sinus anatomy (poor targets, lead instability, high capture thresholds, or extracardiac [phrenic] capture) (Strong Recommendation; Low-Quality Evidence).
41. We recommend CSP be considered as an adjunct in CRT nonresponders with prolonged paced QRS duration despite CRT optimization (Strong Recommendation; Low-Quality Evidence).

In patients with moderate LV dysfunction (LVEF 36%-50%) several observational cohort studies and small randomized trials suggest that CSP is associated with preservation or improvement in LV function, and a lower rate of PICM compared with RV pacing.^{72-74,76,174,187-189} In patients with moderate LV dysfunction (LVEF 36%-50%) and anticipated RV pacing burden > 20%-40% there are limited observational data that have compared traditional CRT via endovascular coronary sinus LV lead placement with CSP, with most of the data available lacking long-term clinical outcomes.¹⁷⁴⁻¹⁸⁶ As a result, the role of CSP will need to be clarified in large multicentre RCTs, such as Left vs Left Randomized Clinical Trial (NCT05650658; in which CSP is being compared with traditional biventricular CRT in patients with LVEF ≤ 50% and either bundle branch block [QRS > 130 ms] or anticipated RV pacing > 40%) and Conduction System Pacing With Left Bundle Branch Pacing as Compared to Standard Right Ventricular Pacing (NCT05015660, LEFT HF; in which CSP is being compared with RV pacing in patients with LVEF > 35% and anticipated RV pacing > 90%). Until the results of these large clinical trials are available the current role of CSP would be limited to:

1. An alternative to RV pacing in patients with preserved or mildly reduced LV function, in whom a RV pacing burden of > 20%-40% is anticipated.
2. An alternative to RV pacing in patients with preserved or mildly reduced LV function, in whom RV pacing is anticipated to be rare (≤ 20%), recognizing that the incremental benefit of CSP might be limited by the use of pacing algorithms designed to minimize RV pacing (section 10.1).
3. An alternative to BiV-CRT via a coronary sinus or epicardial LV lead in patients with severe ventricular dysfunction (LVEF < 35%) and bundle branch block, or moderate-severe ventricular dysfunction (LVEF < 50%) in whom RV pacing is anticipated to be frequent (> 20%-40%), when there is a failure to obtain adequate resynchronization (eg, inability to place an endovascular coronary sinus lead in a nonapical position, presence of high capture thresholds or phrenic pacing, or in the case of BiV-CRT nonresponse).¹⁷⁴⁻¹⁸⁶

6. Defibrillators and sudden cardiac death

6.1. Indications for ICD therapy

An ICD is a specialized device designed to directly treat malignant cardiac tachyarrhythmias such as VT or ventricular fibrillation (VF). The basic system does not fundamentally differ from a pacing system except for the generator having a larger volume (to house the high-voltage power supply and a shock capacitor) and the RV pacing lead having a larger diameter because of the defibrillation function. In general, the transvenous ICD has all sensing and pacing capabilities of a conventional pacemaker but in addition is designed to detect potentially harmful ventricular arrhythmias and to deliver shock and nonschock therapies to terminate these arrhythmias.

Recommendation

42. We recommend that every patient with HFrEF should be assessed for suitability for ICD after optimization of medical therapy to target or maximally tolerated doses (Strong Recommendation; Moderate-Quality Evidence).
43. We recommend that patients should undergo reassessment of LVEF to determine ICD candidacy at least 40 days after MI without revascularization, at least 3 months after MI with revascularization, or at least 3 months after optimization of medical therapy (Strong Recommendation; Low-Quality Evidence).
44. We recommend against ICD implantation in patients with NYHA class IV symptoms who are not expected to improve with any further therapy and who are not candidates for cardiac transplantation or mechanical circulatory support (Strong Recommendation; Moderate-Quality Evidence).
45. We recommend that an ICD not be offered to patients in whom comorbidities make it unlikely that an ICD will substantially increase a patient's life expectancy (Strong Recommendation; Moderate-Quality Evidence).

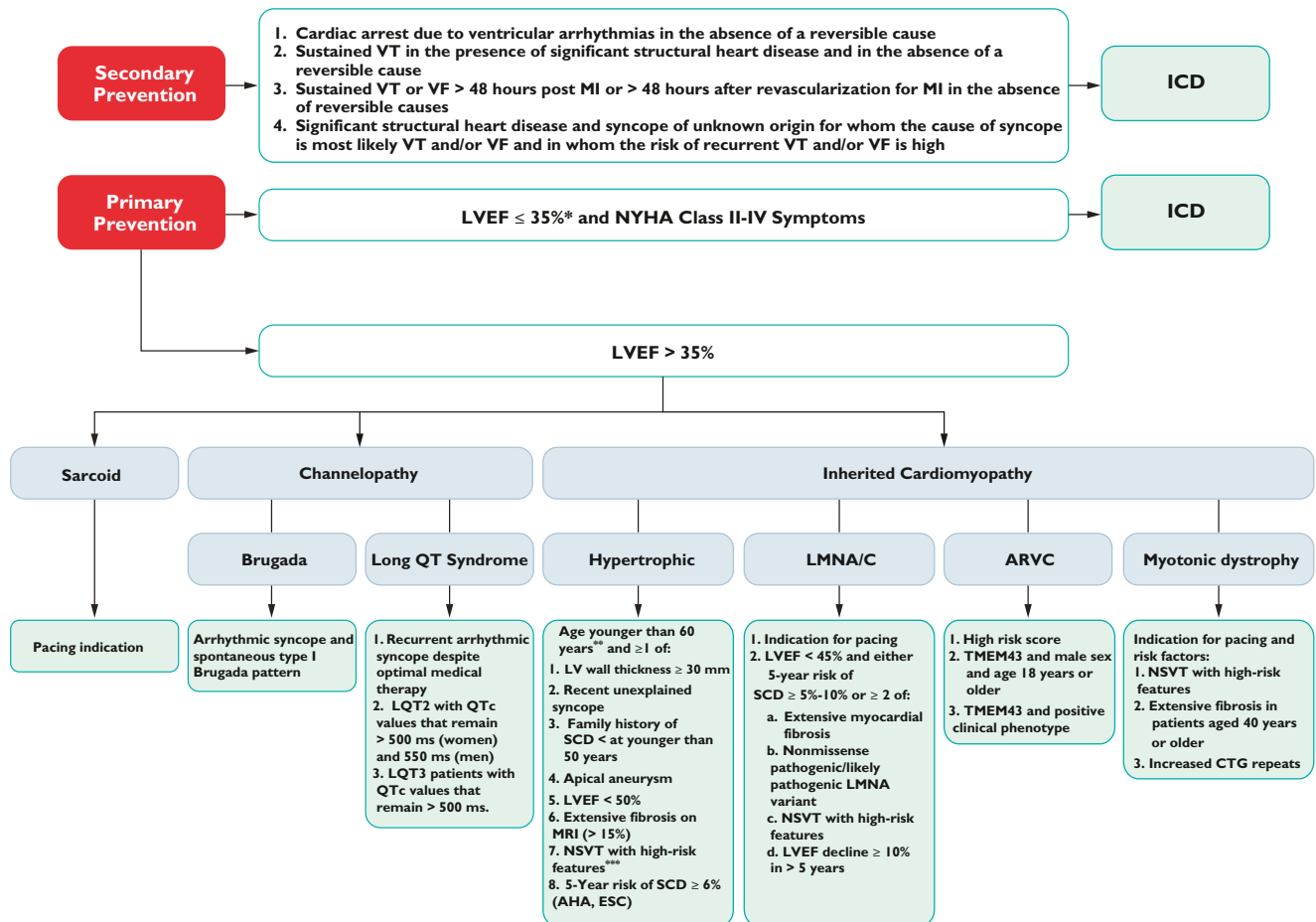


Figure 9. Indications for defibrillator therapy. ARVC, arrhythmogenic right ventricular cardiomyopathy; bpm, beats per minute; GDMT, guideline-directed medical therapy; HCM, hypertrophic cardiomyopathy; ICD, implantable cardioverter defibrillator; LMNA/C, lamin A/C; LQT, long QT; LV, left ventricle; LVEF, left ventricular ejection fraction; MI, myocardial infarction; MRI, magnetic resonance imaging; NSVT, nonsustained ventricular tachycardia; NYHA, New York Heart Association; QTc, corrected QT; SCD, sudden cardiac death; VF, ventricular fibrillation; VT, ventricular tachycardia. * LVEF should be measured at least 1 month post MI, and at least 3 months post coronary revascularization procedure for those with ischemic cardiomyopathy or at least 3 months after optimization of GDMT for nonischemic cardiomyopathy. Those with NYHA class I symptoms warrant an ICD if the etiology of their cardiomyopathy is ischemic and the LVEF is ≤ 30%. ** ICD insertion for primary prevention is rarely beneficial among individuals with HCM after 60 years of age, even in the presence of conventional risk factors. *** Defined as ≥ runs of NSVT during the same monitoring period of 24-48 hours, ≥ 10 consecutive beats, ventricular rate ≥ 200 bpm.

Practical tips

- The referral for ICD implantation should not be unduly delayed if timely titration of pharmacologic therapies is infeasible.
- ICD implantation should only be considered if compatible with the patient's prognosis, comorbid conditions (eg, age, renal function, frailty, genotype), and the individual patient's goals of care (eg, their values and preferences regarding their quantity and quality of life).
- Prediction models can be helpful to estimate the risk of arrhythmic and nonarrhythmic mortality in patients with low LVEF and their use might help identify patients who would benefit most from ICD therapy (eg, Seattle Proportional Risk Score, BaSIS Score, DO-IT model, and the MADIT ICD benefit risk scores).

Best Practice Statement

3. When an ICD is indicated, it is recommended to evaluate whether the patient could benefit from implantation of a CRT device.

6.2. Identifying patients at risk of SCD

SCD due to VT/VF affects 0.1% of the population annually. It is the initial clinical presentation in up to 20% of patients with coronary artery disease and accounts for up to 50% of mortality due to coronary artery disease. However, although most SCD events occur in the highest risk subgroups (ie, HF, low ejection fraction, cardiomyopathy), most SCD events occur in patients without these clinical conditions (Fig. 9). This highlights the need to identify additional risk factors for malignant ventricular arrhythmias or SCD.

Table 6. Clinical and investigational factors contributing to risk assessment of sudden death

Domain	Factor
Clinical assessment	Age Frailty Functional class Comorbidity profile Socioeconomic status
Anatomic substrate abnormalities	Left ventricular ejection fraction Myocardial scar assessment (MRI, SPECT, PET)
Laboratory and biomarkers	Sodium Interleukin-6 NT-proBNP
Autonomic measure	Heart rate variability Heart rate turbulence Baroreceptor sensitivity SPECT (MIBG), PET (11C-meta-hydroxyephedrine)
Electrocardiographic	QRS duration QRS fractionation Signal-averaged ECG T-wave alternans QT dispersion and/or variability QRS-T angle (abnormal if $> 90^\circ$) Corrected QT interval (abnormal if > 450 ms [men] or > 460 ms [women]) T-peak to T-end interval (abnormal if > 89 ms) Ventricular ectopy and nonsustained VT on ambulatory ECG monitoring
Invasive testing	Electrophysiology study
Genetic testing	Referral to specialized cardiogenetic clinic when appropriate

ECG, electrocardiogram; MIBG, 123I-metaiodobenzylguanidine; MRI, magnetic resonance imaging; NT-proBNP, N-terminal pro hormone brain natriuretic peptide; PET, positron emission tomography; SPECT, single photon emission computed tomography; VT, ventricular tachycardia.

Reduced LVEF remains one of the strongest risk factors for SCD. Although LVEF remains the cornerstone of predicting arrhythmia risk, it will identify only a fraction of those at risk. Adjunctive predictors include: (1) anatomic substrate abnormalities; (2) biomarkers; (3) autonomic abnormalities; (4) ECG abnormalities; (5) invasive arrhythmia assessment; and (6) assessment of genetic risk (Table 6).

In addition, a proportion of patients at objective high risk of SCD-VT/VF might also be at high risk of nonarrhythmic death. The relative benefit of ICD therapy is attenuated when the competing risk of death exceeds the risk of SCD-VT/VF. Among the risk factors for nonarrhythmic death, age and frailty seem to be the strongest. However, many risk factors have a complicated relationship with the benefit of the ICD, and influence SCD-VT/VF risk and risk of nonarrhythmic death. In this case, the use of prediction models to estimate the risk of arrhythmic and nonarrhythmic mortality in patients with low LVEF might help identify patients who would benefit most from ICD therapy (eg, Seattle Proportional Risk Score, BaSIS Score, DO-IT model, and the MADIT ICD benefit risk scores).¹⁹⁰⁻¹⁹²

6.2.1. Secondary prevention of SCD due to ventricular arrhythmias.

Recommendation

46. We recommend ICD implantation for patients after cardiac arrest due to ventricular arrhythmias in the absence of a reversible cause (Strong Recommendation; High-Quality Evidence).

Recommendation

47. We recommend ICD implantation for patients with sustained VT in the presence of significant structural heart disease and in the absence of a reversible cause (Strong Recommendation; High-Quality Evidence).
48. We suggest an ICD be considered in patients with significant structural heart disease and syncope of unknown origin for whom the cause of syncope is most likely VT/VF and in whom the risk of recurrent VT/VF is high (Weak Recommendation; Moderate-Quality Evidence).
49. We recommend an ICD for patients with syncope of unknown origin with inducible VT at the time of EPS and structural heart disease (Strong Recommendation; Moderate-Quality Evidence).
50. We suggest an ICD be considered after MI in the presence of sustained VT or VF > 48 hours after MI or > 48 hours after revascularization for MI in the absence of reversible causes (Weak Recommendation; Low-Quality Evidence).

Practical tips

- Structural heart disease is defined as abnormalities or defects in the heart's valves, walls, or chambers, typically characterized by a reduction in myocardial contractility (ejection fraction) and/or the presence of myocardial scar.
- Significant structural heart disease pertains to structural heart conditions in which the risk of cardiac arrest in the presence of ventricular arrhythmia is high. This does not pertain to conditions in which the risk of cardiac arrest is low such as atrial septal defect, most valvulopathies, and atriopathies.
- Sustained VT refers to hemodynamically significant VT or VT lasting > 30 seconds in duration
- Sustained monomorphic VT in the context of ischemic heart disease reflects the presence of myocardial scar and tends to recur despite revascularization.
- ICD implantation in patients with incessant ventricular arrhythmias should be delayed until the ventricular arrhythmias have been controlled.
- The role of electrophysiology study in patients with syncope suspected due to ventricular arrhythmias and nonischemic heart disease is unproven.
- Certain patients with sentinel presentation of cardiac arrest due to inherited channelopathy (eg, catecholaminergic polymorphic VT [CPVT], long QT syndrome [LQTS]) might not require secondary prevention ICD. These patients should be reviewed on a case-by-case basis by specialists with expertise in the management of individuals with inherited channelopathy.

Since the previous guidelines, there have been no further trials to assess the mortality benefit of secondary prevention ICDs (patients who have already experienced an aborted

cardiac arrest or life-threatening ventricular arrhythmia).² A meta-analysis that included the 3 landmark trials (Antiarrhythmics Versus Implantable Defibrillators [AVID], Canadian Implantable Defibrillator Study [CIDS], Cardiac Arrest Study Hamburg [CASH]) showed a 28% reduction in the RR of death with an ICD primarily because of a reduction in all-cause death (hazard ratio, 0.72; 95% CI, 0.6-0.87; $P = 0.0006$) almost entirely due to a reduction of arrhythmic death (hazard ratio, 0.5; 95% CI, 0.37-0.67; $P = 0.0001$) in the ICD group.¹⁹³ The recommendation for secondary prevention ICD implantation similarly applies to patients with acquired and nonacquired forms of cardiovascular disease. Please see the 2016 CCS/CHRS ICD guidelines for in-depth review of the evidence informing these recommendations.²

6.2.2. Primary prevention of SCD due to ventricular arrhythmias.

Recommendation

51. We recommend consideration of primary ICD therapy in patients with ischemic cardiomyopathy, NYHA class I symptoms, and an ejection fraction $\leq 30\%$ at least 1 month after MI, and at least 3 months after a coronary revascularization procedure (Strong Recommendation; High-Quality Evidence).
52. We recommend consideration of primary ICD therapy in patients with ischemic cardiomyopathy, NYHA class II-III symptoms, and ejection fraction $\leq 35\%$, measured at least 1 month after MI, and at least 3 months after a coronary revascularization procedure (Strong Recommendation; High-Quality Evidence).
53. We recommend primary prevention ICD implantation in patients with nonischemic cardiomyopathy, NYHA class II-III symptoms, and an LVEF $\leq 35\%$, measured at least 3 months after optimization of GDMT (Strong Recommendation; High-Quality Evidence).

Practical tips

- An assessment of arrhythmic and nonarrhythmic death should be performed to estimate the risk-benefit of ICD implantation with the use of published prediction rules.
- The absence of HF symptoms is unlikely to modify the risk of sudden death and should not influence the decision to proceed with ICD implantation.

Recommendation

54. We suggest that an ICD be considered in patients with an acute indication for cardiac pacing within 3 months after revascularization or 40 days of MI and before achieving optimal medical therapy when there is a high probability that the ejection fraction will remain $< 35\%$ (Weak Recommendation; Low-Quality Evidence).

Practical tip. Recovery in ejection fraction is less likely to occur in the presence of extensive myocardial scar, lack of myocardial viability before revascularization, when revascularization addressed only a small amount of myocardium, or when the infarcted territory was small.

Since the previous guidelines, there have been no new trials to assess the mortality benefit of primary prevention ICDs (eg, patients who have not experienced an aborted cardiac arrest or life-threatening ventricular arrhythmia).² The previous landmark randomized controlled trials showed that primary prevention ICD therapy was associated with an aggregate reduction in arrhythmic (RR, 0.40; 95% CI, 0.27-0.67) and all-cause mortality (RR, 0.73; 95% CI, 0.64-0.82).¹⁹⁴ Although this benefit was attenuated in the nonischemic population (the absolute risk of all-cause mortality and SCD are lower in patients with nonischemic cardiomyopathy [NICM]), the relative benefit obtained from primary prevention ICD implantation was noted to be similar in ischemic and nonischemic populations (hazard ratio, 0.77 for all-cause mortality in NICM; 95% CI, 0.64-0.93).¹⁹⁵

In the years since the publication of these landmark trials the pharmacological treatment of cardiomyopathy has evolved substantially with the emergence of newer medications that improve LVEF and reduce mortality.⁶ Ongoing studies are evaluating the role of ICD therapy for primary prevention in the context of contemporary quadruple GDMT, however these trials are not due to complete for several years (PRevention Of sudden cardiac death after myocardial Infarction by Defibrillator implantation [PROFID EHRA; NCT05665608], and Comparative Effectiveness of Prophylactic ICD Versus Non-ICD Therapy in CONTEMPorary Heart Failure Patients at a Low Risk for Arrhythmia Death [CONTEMP-ICD; NCT06543446]). Until then the established recommendations regarding primary prevention ICD implant remain in place.

6.3. Primary prevention ICD in special populations

Additional risk predictors for primary prophylaxis ICD implant for special populations are presented in [Table 7](#).

6.3.1. Cardiac sarcoidosis.

Recommendation

55. We recommend primary prevention ICD implantation in patients with cardiac sarcoidosis and an indication for pacemaker implantation (Weak Recommendation; Low-Quality Evidence).

Patients with CS can experience a ventricular arrhythmia at any phase of the disease. The underlying mechanism of SCD-VT/VF can be autonomic, triggered, or reentry, depending on the phase of granulomatous infiltration (eg, inflammatory or fibrotic) and the location of myocardial involvement, which can involve the Purkinje system, one or both ventricles, as well as intramural or epicardial locations.

Because of this complexity the risk stratification for SCD-VT/VF in patients with CS is somewhat nuanced. Although the traditional indication for ICD therapy on the basis of

Table 7. Additional risk predictors supporting ICD implantation in addition to standard primary prevention ICD indications

Condition	
Sarcoidosis	Indication of permanent pacing
Inherited cardiomyopathy	
Hypertrophic	Recent unexplained syncope Left ventricular wall thickness ≥ 30 mm LVEF $< 50\%$ Apical aneurysm Extensive fibrosis on cardiac MRI ($> 15\%$) High-risk nonsustained VT Strong family history of SCD Estimated 5-year risk of SCD $\geq 6\%$
Lamin A/C	LVEF $< 45\%$ Estimated 5-year risk of SCD $\geq 10\%$ Extensive myocardial fibrosis Nonmissense pathogenic and/or likely pathogenic <i>LMNA</i> variant NSVT with high-risk features Marked disease progression (LVEF decline $\geq 10\%$ in > 5 years)
ARVC	High risk of SCD on the basis of the ARVC Risk Calculator
TMEM43	<i>TMEM43</i> -S358L variant with positive clinical phenotype Men 18 years or older with <i>TMEM43</i> -S358L variant, regardless of phenotype
Neuromuscular disease	
Myotonic dystrophy	Genetic (CTG expansion) Extensive myocardial scar NSVT See lamin A/C above
Emery-Dreifuss or limb-girdle muscular dystrophy 1B	
Channelopathy	
Brugada	Arrhythmic syncope and spontaneous type I pattern
LQTS	Recurrent arrhythmic syncope despite optimal medical therapy LQT2 with corrected QT values that remain > 500 ms (women) and 550 ms (men) LQT3 patients with corrected QT values that remain > 500 ms Inducible VT and/or VF at electrophysiological study with associated ventricular dysfunction
Congenital heart disease	Tetralogy of Fallot with clinical VT or multiple risk factors (previous palliative shunt, spontaneous NSVT, inducible VT, QRS > 180 ms, ventriculotomy, LVEDP ≥ 12 mm Hg) Single and/or systemic ventricular dysfunction with NSVT, NYHA classes II-III High-risk according to established risk score (PREVENTION-ACHD, Spanish ACHD)

ARVC, arrhythmogenic right ventricular cardiomyopathy; ICD, implantable cardioverter defibrillator; LQT, long QT syndrome, LVEF: left ventricular ejection fraction; LVEDP, left ventricular end diastolic pressure; MRI, magnetic resonance imaging; NSVT, nonsustained ventricular tachycardia; NYHA, New York Heart Association; PREVENTION-ACHD, PRospectiVE study on implanTable cardioverter defibrillator therapy and suddEN cardiac death in Adults with Congenital Heart Disease; SCD: sudden cardiac death; Spanish ACHD, Spanish Adults with Congenital Heart Disease; TMEM43, transmembrane protein 43; VF, ventricular fibrillation; VT, ventricular tachycardia.

reduced LVEF applies to this population, patients who present with an indication for bradycardia pacing should receive an ICD in preference to a bradycardia device. Beyond these circumstances, the risk stratification for ICD therapy should include an assessment of clinical and anatomic factors. Significant involvement of the left ventricle and/or the right ventricle is associated with increased mortality independent of LVEF, however the threshold of scar burden deemed clinically significant remains unclear. Likewise, although an abnormal positron emission tomography scan is associated with an increased risk of SCD-VT/VF independent of LVEF, the optimal index for use is still undetermined. In borderline cases additional risk stratification can be obtained from an electrophysiology study with programmed ventricular stimulation. Although inducible sustained monomorphic VT can be helpful (specificity, 0.93-0.97), the progressive nature of the disease means a negative electrophysiology study does not eliminate long-term risk (sensitivity 0.63 in those with LVEF $\geq 35\%$).¹⁹⁶

Corticosteroid therapy has shown efficacy in preventing disease progression, especially in patients with preserved or mildly reduced LVEF.¹⁹⁷ Among patients with ventricular arrhythmias, antiarrhythmic medications are commonly required in conjunction with immunosuppression during the active inflammatory

phase, or can be used as monotherapy with or without catheter ablation when evidence of inflammation is absent.

6.3.2. Inherited cardiomyopathy

6.3.2.1. Hypertrophic cardiomyopathy.

Best Practice Statement

- Patients with hypertrophic cardiomyopathy (HCM) should undergo comprehensive evaluation and SCD risk stratification by specialists with expertise in the management of these individuals.

Recommendation

- We suggest an estimate of risk for SCD in patients with HCM should be determined on the basis of validated risk scores and/or the presence of one or more high-risk clinical factors to select appropriate candidates for primary prevention ICD therapy (Weak Recommendation; Moderate-Quality Evidence).

Practical tips

- A primary prevention ICD is rarely indicated for individuals with a first diagnosis of HCM after 60 years of age, because the annual risk of disease-related SCD is as low as 0.2%.
- High-risk features include LV wall thickness ≥ 30 mm, recent unexplained syncope, LVEF $< 50\%$, apical aneurysm, extensive fibrosis on MRI ($> 15\%$), non-sustained VT (NSVT) with high-risk features, the presence of a sarcomeric pathogenic mutation, and a strong family history of SCD.
- A commonly used threshold for considering prophylactic defibrillator implantation is a 5-year risk of SCD of 4%-6%, which should be considered in the context of a patient's comorbid conditions and clinical factors (eg, age, renal function, frailty), as well as the patient's values and preferences regarding their quantity and quality of life.

HCM is a common disorder (prevalence 1/500) most often due to abnormal sarcomere function, which results in increased myocardial contractility and impaired relaxation.¹⁹⁸ SCD is a devastating but most often preventable complication of HCM. In adult patients with HCM, a comprehensive assessment of SCD risk should be performed at initial evaluation and every 1-2 years thereafter up to the age of 60-65 years.¹⁹⁹ Contemporary risk stratification strategies include the use of individual risk predictors to decide ICD candidacy (eg, unexplained syncope, massive LV hypertrophy [ventricular wall thickness ≥ 30 mm], LV systolic dysfunction [$< 50\%$], extent of ventricular scar [$\geq 15\%$], the presence of NSVT, recent unexplained syncope, apical aneurysm, or a strong family history of SCD (typically defined as SCD occurring at a younger age in first-degree relatives, or when multiple family members suffered SCD), or use a validated risk multivariate prediction formula, using the estimate of the 5-year risk of SCD or life-threatening arrhythmic events to guide device implantation.²⁰⁰ Although the risk-prediction model provides a quantifiable estimate of SCD-VT/VF risk and offers the advantage of considering multiple continuous and binary predictors of different weights, the formula does not consider newer risk predictors (eg, systolic dysfunction [LVEF $< 50\%$], the presence of apical aneurysm, and extent of LV scar). A detailed review of risk predictors for SCD in patients with HCM can be found in a CCS clinical practice update.¹⁹⁹

Device selection in patients with HCM is not straightforward. Because of the anti-tachycardia pacing (ATP) responsiveness of HCM-associated VT,²⁰¹ there is a rationale to favour transvenous ICD implantation, however this approach is associated with more lead-related complications and long-term risk of lead failure. A nontransvenous ICD reduces the risk related to lead failure but are associated with significantly more inappropriate shocks.

6.3.2.2. Lamin A/C.

Recommendation

57. We recommend that patients with cardiac or neuromuscular laminopathies should undergo comprehensive evaluation and SCD risk stratification by specialists with expertise in the management of these individuals (Strong Recommendation; Low-Quality Evidence).
58. We recommend primary prevention ICD insertion in patients with cardiac or neuromuscular laminopathies and an LVEF of $< 45\%$ and an elevated 5-year risk of life-threatening ventricular tachyarrhythmias (<https://lmna-risk-vta.fr/>), and/or the presence of ≥ 2 additional risk factors (Strong Recommendation; Low-Quality Evidence).

Practical tips

- Additional risk factors include nonmissense pathogenic and/or likely pathogenic LMNA variant, NSVT with high-risk features, extensive myocardial fibrosis, and marked disease progression (defined as LVEF decline $\geq 10\%$ in > 5 years).
- A commonly used threshold for considering prophylactic defibrillator implantation is a 5-year risk of life-threatening ventricular tachyarrhythmias of 5%-10%, which should be considered in the context of the patient's comorbid conditions and clinical factors (eg, age, renal function, frailty), as well as the patient's values and preferences regarding their quantity and quality of life.

Recommendation

59. We suggest primary prevention ICD insertion might be reasonable in patients with cardiac or neuromuscular laminopathies and an indication for a PPM (Weak Recommendation; Moderate-Quality Evidence).

Values and preferences. This recommendation places great value on the risk of death associated with pathogenic variants in the lamin A/C gene.

Practical tip. When an ICD is indicated, it is recommended to evaluate whether the patient could benefit from implantation of a CRT-capable device because of an increased risk of progression to complete heart block.

Lamin A and C are structural filament proteins in the cell nucleus, encoded by the gene *LMNA*. Pathogenic variants in the lamin A/C gene are identified in approximately 5%-10% of

idiopathic familial dilated cardiomyopathy cases and are associated with a strikingly high penetrance. Isolated cardiac laminopathies or neuromuscular laminopathies (Emery-Dreifuss muscular dystrophy, LGMD type 1b) develop secondary to distinctly different *LMNA* variants compared with other forms of laminopathy, including Hutchinson-Gilford progeria or hereditary lipodystrophy.²⁰² The cardiac phenotype of *LMNA* is characterized by a dilated cardiomyopathy associated with high rates of AV conduction disorders, SND, and atrial arrhythmias. Malignant ventricular arrhythmia and/or SCD might be observed in upward of 30% of patients.^{203,204} End-stage HF requiring heart transplantation will occur in up to 20% of cases.^{203,204} Risk stratification for SCD remains challenging and requires periodic reevaluation. Increased risk of ventricular arrhythmia or SCD has been shown in patients with LVEF < 45% and presence of ≥ 2 additional risk factors, NSVT with high-risk features, male sex, and nonmissense *LMNA* variants.^{205,206} However, the predictive value of sex and NSVT has been questioned in more recent reports.²⁰⁷ Marked disease progression, defined as LVEF decline $\geq 10\%$ over < 5 years or extensive myocardial fibrosis, is also predictive of increased risk of SCD.^{207,208} The LMNA-risk ventricular tachyarrhythmia calculator is a multivariable risk prediction model used to estimate the risk of life-threatening ventricular arrhythmia within 5 years.²⁰⁹ However, the model tends to overestimate the risk of ventricular arrhythmia.²¹⁰ A cutoff value of a 5%-10% risk of ventricular arrhythmia over 5 years seems to be a reasonable threshold for the implantation of a primary prevention ICD.

6.3.2.3. Arrhythmogenic RV cardiomyopathy.

Recommendation

60. We recommend that patients with suspected or confirmed arrhythmogenic RV cardiomyopathy (ARVC) should undergo comprehensive evaluation and SCD risk stratification by specialists with expertise in the management of these individuals (Strong Recommendation; Low-Quality Evidence).
61. We suggest primary prevention ICD implantation in patients with definite ARVC related to pathogenic and/or likely pathogenic *PKP2* variants at high risk of SCD on the basis of the ARVC Risk Calculator (Weak Recommendation; Low-Quality Evidence).

Practical tips

- A commonly used threshold for considering prophylactic defibrillator implantation is a 5-year risk of SCD of 4%-6%, which should be considered in the context of a patient's comorbid conditions and clinical factors (eg, age, renal function, frailty), as well as the patient's values and preferences regarding their quantity and quality of life.
- It is reasonable to apply the ARVC Risk Calculator (<https://arvcrisk.com/>) to predominant right ventricular ARVC related to desmoglein 2 (*DSG2*) or desmocollin 2 (*DSC2*) variants.

ARVC is a genetically mediated progressive cardiomyopathy, which in most cases is related to genetic variants within

desmosomal genes (*PKP2*, *DSG2*, *DSC2*, *JUP*, *CDH2*), the cellular ultrastructure responsible for cell-to-cell adhesion. Because of the relative sparing of the left ventricle (although it might still be affected), a custom approach to SCD risk stratification has been necessary.^{54,55} Risk factors for SCD or sustained VT and/or VF include: (1) younger age at diagnosis; (2) male sex; (3) previous cardiac syncope; (4) an increased burden of premature ventricular contractions; (5) presence of spontaneous NSVT; (6) spontaneous or inducible sustained monomorphic VT; (7) extension and/or progression of disease; (8) the burden of myocardial fibrosis; and (9) number of leads with T-wave inversion on 12-lead ECG. None is sufficiently predictive in isolation to guide clinical decision-making, however, combining these features into a weighted risk score has been shown to effectively predict onset of malignant arrhythmias. The ARVC Risk Calculator is used to estimate the risk of a first ventricular arrhythmia within 5 years for patients with a definite ARVC diagnosis related to *PKP2* variants, providing an objective and quantifiable estimate of the risk of SCD-VT/VF to inform shared decision-making regarding ICD implantation.^{211,212} The model is primarily calibrated for *PKP2*-ARVC and works reasonably well for *DSG2/DSC2*, but has limited predictive value for ARVC with predominant LV involvement linked to *DSP* and gene-elusive individuals, who represent up to 33% of patients with definite ARVC.^{213,214}

Device selection in patients with ARVC is not straightforward. Because of the ATP responsiveness of ARVC-associated VT^{215,216} there is a rationale to favour transvenous ICD implantation, however, this approach is associated with more lead-related complications and long-term risk of lead failure and abandonment or extraction. Conversely, nontransvenous ICDs reduce the risk related to lead failure but are associated with significantly more inappropriate shocks. In both cases there is a risk of deterioration in sensing over time because of replacement fibrosis of the right ventricle, although long-term sensing with nontransvenous ICDs in the ARVC population remains unknown. Considering these factors, nontransvenous ICDs might be favoured in younger patients without a pacing indication who undergo primary prevention implantation for VF, and transvenous ICDs might be favoured for older patients with more advanced structural heart disease and who are at high risk of sustained VT (primary or secondary prevention).

6.3.2.4. Transmembrane protein 43 cardiomyopathy (*TMEM43-CMP*).

Recommendation

62. We recommend primary prevention ICD implantation in patients who harbor the *TMEM43*-S358L variant upon manifestation of a positive clinical phenotype (most often increased ventricular ectopy on Holter) and in men 18 years of age or older regardless of phenotype (Weak Recommendation; Moderate-Quality Evidence).

Practical tip. High-risk ECG abnormalities include ≥ 1000 premature ventricular complexes (PVCs) in 24 hours (poor precordial R-wave progression, progressive QRS widening).

Transmembrane protein 43 cardiomyopathy (TMEM43-CMP) is a malignant form of nondilated LV cardiomyopathy that arises from a single variant (-P.S358L) in the TMEM43, which encodes lamina-associated polypeptide, unidentified function, membrane-associated (LUMA), a nuclear envelope protein that interacts with A-type lamins and emerin. TMEM43 results in a highly malignant biventricular cardiomyopathy at elevated risk of malignant ventricular arrhythmias. It is secondary to a genetic founder effect, with a relatively high prevalence in Newfoundland.²¹⁷⁻²¹⁹ TMEM43-CMP is considered fully penetrant in men, and men have significantly worse outcomes compared with women. The high risk of ventricular arrhythmia and SCD in men is independent of the ventricular ejection fraction.²²⁰ As a consequence, a primary prevention ICD is recommended in postpubertal men regardless of phenotype. Onset of a positive phenotype is considered an indication for ICD in men and women.²²⁰

6.3.2.5. Other nondilated LV cardiomyopathies. Pathogenic variants in desmoplakin, filamin C, phospholamban, and RNA-binding motif protein 20, and desmin have been associated with a predominant left-sided phenotype and associated elevated risk of ventricular arrhythmias. These conditions are discussed in depth in the white paper in this issue of the *Canadian Journal of Cardiology*.

6.3.3. Neuromuscular disease.

Recommendation

63. We recommend that patients with neuromuscular disease and associated cardiomyopathy undergo comprehensive evaluation and SCD risk stratification by specialists with expertise in the management of these individuals (Strong Recommendation; Low-Quality Evidence).
64. We suggest primary prevention ICD implantation in patients with neuromuscular disease and LVEF $\leq$ 35% despite optimal medical therapy (Weak Recommendation; Low-Quality Evidence).
65. We recommend primary prevention ICD insertion in patients with Emery-Dreifuss muscular dystrophy or LGMD 1B and an indication for pacing, or those with an LVEF of $<$ 45%, and an elevated risk of sudden death (see section 6.3.3.2) (Strong Recommendation; Low-Quality Evidence).
66. We suggest primary prevention ICD insertion in patients with myotonic dystrophy who have an indication for pacemaker implantation and presence of additional risk factors for VT (NSVT) or SCD (significant LGE on cardiac MRI in patients of aged 40 years or older, increased CTG repeats) (Weak Recommendation; Low-Quality Recommendation).
67. We recommend against invasive electrophysiology study for risk stratification in patients with neuromuscular disease (Strong Recommendation; Moderate-Quality Evidence).

The most common neuromuscular disorders with cardiac manifestations in adults include myotonic dystrophies type I and II (also called Steinert disease), Emery-Dreifuss muscular dystrophy (related to pathogenic variants in the *EMD* or *LMNA* genes), LGMD type 1b (related to *LMNA*), and muscular dystrophy (Duchenne and Becker). Arrhythmias are common and can be the first manifestation of heritable neuromuscular disorders, with patients with some disorders (eg, myotonic dystrophy, and certain forms of isolated lamin cardiomyopathy or Emery-Dreifuss muscular dystrophy) experiencing disproportionately greater rates of brady-asystole or advanced AV conduction disorders rather than malignant ventricular arrhythmia (see section 2.5.8; Fig. 6).

It is generally recommended that patients with neuromuscular diseases who have survived a sudden cardiac arrest, manifest sustained VT, or experience severe ventricular dysfunction (eg, LVEF $<$ 35%) be treated in the same way as patients without neuromuscular disorders, including consideration of the patient's overall prognosis (eg, competing risk of death not due to malignant ventricular arrhythmia), clinical status, and the individual patient's goals of care.

For those with muscular dystrophies associated with a pathogenic LMNA variant (LGMD type 1b, type II, or autosomal Emery-Dreifuss) or emerin (type I or X-linked Emery-Dreifuss), an ICD should be considered in those with an indication for permanent pacing or manifest conduction disease, even in the absence of AVB or ventricular dysfunction.

For myotonic dystrophies, advanced conduction disorders represent the overwhelming mechanism of SCD, and the benefits of a primary prevention ICDs in this population remains unknown.^{57,221} The cumulative incidence of sustained VT in patients with myotonic dystrophy remains low²²²; if present, VT commonly presents as bundle-branch reentry VT, for which catheter ablation can be effective. Overall, insertion of a primary prevention ICD is rarely indicated in patients with myotonic dystrophy and should be reserved for selected individuals with additional risk factors for sudden death (eg, significant LGE on cardiac MRI in patients of aged 40 years or older, increased CTG repeats, or concomitantly reduced LVEF unrelated to chronic RV pacing).

In each case the benefit of ICD to terminate SCD-VT and/or VF should be balanced against the overall prognosis associated with the neuromuscular disorder.

6.3.4. Channelopathy

6.3.4.1. CPVT. CPVT is an inherited electrical condition characterized by adrenergically mediated bidirectional and/or polymorphic VT. CPVT has an estimated prevalence of 1:10,000 and tends to manifest between 7 and 12 years of age. Symptomatic patients usually present with exercise-induced or emotional stress-related syncope. The characteristic arrhythmias are bidirectional VT or polymorphic VT in the context of a catecholaminergic surge. Some episodes might be short-lived and terminate spontaneously, whereas others might degenerate into VF, resulting in SCD. CPVT principally arises from pathogenic variants in the ryanodine receptor 2 (a sarcoplasmic reticulum membrane channel protein expressed in cardiac

tissues) and calsequestrin 2 (a calcium storage protein in the sarcoplasmic reticulum), both of which play a critical role in regulating intracellular calcium. Pathogenic variants in these genes might result in excessive intracellular calcium concentration leading to delayed afterdepolarizations and triggered activity. No risk prediction algorithms have been developed for CPVT, and the need for ICD therapy has been controversial predominantly because of the catecholamine-stimulating nature of painful ICD shocks and risk of electrical storm.^{3,4} Optimal ICD programming for the CPVT population might be very important. Select high-risk groups of CPVT patients in whom an ICD might be considered are those with polymorphic and/or bidirectional VT refractory to optimal medical therapy (eg, nadolol with or without flecainide) with or without adjuvant left cardiac sympathetic denervation, however, the decision regarding ICD therapy should be made after consultation with clinicians with dedicated expertise in the management of this population.

6.3.4.2. Brugada syndrome.

Recommendation

68. We recommend primary prevention ICD implantation in patients with a spontaneous type I Brugada pattern and arrhythmic syncope (Strong Recommendation; Low-Quality Evidence).

Practical tip. Implantable cardiac monitor implantation might be considered for patients at uncertain risk, those who are reluctant toward ICD implantation, and those who have undifferentiated syncope for diagnostic clarification.

Brugada syndrome is an inherited electrical condition characterized by a distinctive ECG pattern (right bundle branch pattern with coved morphology and ≥ 2 mm of ST elevation) in the right precordial leads that is associated with malignant ventricular arrhythmias. The prevalence of Brugada syndrome is approximately 5 in 10,000 worldwide, with a prevalence that varies widely according to region (1:250 in Asia to 1:20,000 in North America). The mechanism underlying the ECG pattern and SCD-VT and/or VF risk is controversial but is thought to involve impaired conduction or repolarization in the RV outflow tract. Approximately 20% of patients are identified to possess a rare loss of function variant in sodium voltage-gated channel alpha subunit 5 (SCN5A, the cardiac sodium channel), and the remaining patients, currently classified as genotype-negative, likely arise due to a polygenic substrate. Risk stratification for SCD-VT and/or VF in patients with Brugada syndrome has evolved concurrent to a recognition that the risk of SCD-VT and/or VF is less than previously anticipated. Multiple risk prediction algorithms for asymptomatic patients have been proposed, however, their predictive capacity beyond the derivation cohorts have generally been modest. Currently, the only risk predictor used to inform primary prevention ICD implantation is the combination of spontaneous type I pattern and a history of arrhythmic syncope.

6.3.4.3. LQTS.

Recommendation

69. We suggest primary prevention ICD implantation be considered in patients with long QT type 2 (LQT2) with corrected QT (QTc) values that remain > 500 ms (if female) and 550 ms (if male) despite optimal medical therapy (Weak Recommendation; Low-Quality Evidence).
70. We suggest primary prevention ICD implantation be considered in long QT type 3 (LQT3) patients with QTc values that remain > 500 ms despite optimal medical therapy (Weak Recommendation; Low-Quality Evidence).
71. We suggest ICD implantation for LQTS patients with arrhythmic syncope despite optimal medical therapy, as guided by their genetic subtype (Weak Recommendation; Low-Quality Evidence).

Practical tips

- For patients with LQTS who undergo device implantation, consideration should be made toward the potential value of atrial pacing when considering device selection.
- Implantable cardiac monitor implantation might be considered for patients at uncertain risk, those who are reluctant toward ICD implantation, and those who have undifferentiated syncope for diagnostic clarification.

LQTS is an inherited cardiac channelopathy resulting from rare pathogenic variants in the ion channel genes affecting cardiac repolarization, commonly a loss of function of potassium channel (long QT type 1 [LQT1] [KCNQ1] and LQT2 [KCNH2]) or a gain-of-function of the sodium channel (LQT3 [SCN5A]).¹³ The hallmark of LQTS is a prolongation of the QT interval and is often associated with T-wave morphological abnormalities. Because of the abnormalities in ventricular repolarization, patients with LQTS are at risk of SCD-VT and/or VF although this risk is largely mitigated with β -blockers and avoidance of QT-prolonging drugs (crediblemeds.org). Primary prevention ICDs are rarely required because of the low incidence of breakthrough ventricular arrhythmias.

A primary prevention ICD might be useful in certain high-risk populations of LQTS genetic subtypes. LQT1 is the lowest risk genetic subtype and breakthrough events are rare when patients are compliant with optimal β -blocker dose and avoid QT-prolonging medications.¹⁷ LQT1 patients with QTc values consistently > 550 ms might be considered for a primary prevention ICD, however, they often respond well to left cardiac sympathetic denervation.^{16,18} In this context, we recommend LQT1 patients with persistent QTc values > 550 be referred to a specialized genetic arrhythmia clinic for further assessment. LQT2 is a higher risk genetic subtype and it is reasonable to consider a primary prevention ICD when the resting QTc is consistently prolonged despite optimal medical therapy (> 550 ms in men and > 500 ms in

women).^{14,16} A primary prevention ICD might be considered among LQT3 patients when the QTc remains > 500 ms despite optimal treatment with nadolol and mexiletine.^{14,16,19}

6.3.5. Congenital heart disease. Although SCD-VT and/or VF is a relatively rare event in the CHD population (yearly incidence between 0.07 and 0.40 per 100,000 person-years), it remains one of the leading causes of mortality in this population (accounting for up to 25% of all deaths) with rates among those with moderate or complex CHD that are 20- to 30-fold higher than the general population.²²³ The incidence of SCD is highest in the mid-30s in patients with complex CHD and at an older age (ie, after 50 years), in those with simple forms of CHD. High-risk substrates include patients with tetralogy of Fallot, transposition of the great arteries, particularly with atrial switch surgery or congenitally corrected transposition of the great arteries, cyanotic heart disease (Eisenmenger and non-Eisenmenger), Ebstein anomaly, and a Fontan circulation. Because of the heterogeneity inherent to the CHD population the prediction of SCD-VT and/or VF risk is imprecise. Determining who will benefit from primary prevention ICD use becomes difficult, particularly in consideration of: (1) many SCD events in this population are not due to VT and/or VF; (2) the competing risk of nonarrhythmic death can be substantial; and (3) ICD-related complication rates are substantially higher in the CHD population than in other populations. With respect to the latter, device-related complications and inappropriate shock were observed in approximately 25% of CHD patients over a mean follow-up of just under 4 years. Disease-specific risk prediction scores have been developed for patients with systemic right ventricles²²⁴ and tetralogy of Fallot.²²⁵ Broader risk prediction models such as the PRoSpectiVE study on implanTable cardioverter defibrillator therapy and sudden cardiac death in Adults with Congenital Heart Disease (PREVENTION-ACHD) and Spanish Adults with Congenital Heart Disease (Spanish ACHD), which consider factors related to CHD type, as well as comorbidity, ventricular function (systemic and/or subpulmonary), and ECG criteria (QRS duration, QT dispersion), have been instrumental in identifying factors that transcend specific lesion types.²²⁶⁻²²⁸ However, their utility at the individual level might be limited by a lack of granularity needed for personalized decision-making. Last, because of the high rates of lead dysfunction and concomitant desire to avoid intravascular material (either due to obstruction and/or congenitally anomalous veins, or intracardiac shunts) in the CHD population there might be a preference to consider the nontransvenous ICD (subcutaneous or extravascular ICD) for those with an ICD indication, however it is important to recognize that a substantial proportion of the CHD population might be ineligible for this therapy because of screening failure (eg, baseline RBBB), previous heart surgery, concomitant pacing indication, or high risk of inappropriate shocks.

6.4. Device selection

Recommendation

72. We suggest a single-chamber ICD instead of a dual-chamber ICD in patients without a pacing indication who undergo ICD implant (Weak Recommendation; Moderate-Quality Evidence).
73. We recommend a nontransvenous ICD be considered as an alternative to single-chamber transvenous ICD in patients without a pacing indication who undergo ICD implant (Strong Recommendation; Moderate-Quality Evidence).

Practical tip. Populations in which a nontransvenous ICD might be preferred include:

- Patients with known difficulties in vascular access (occlusion or stenosis of the upper extremity venous system) or limited pocket sites, including patients with previous lead- and device-related complications;
- Patients at high risk of infection, particularly patients with indwelling vascular catheters, are dialysis-dependent, have previous infection or extraction, or are immunocompromised;
- Patients with CHD who have complex or absent transvenous access to the ventricle or have intracardiac shunts (greater risk of stroke with transvenous leads);
- Patients with moderate to severe tricuspid regurgitation where surgical or percutaneous tricuspid valvular repair or replacement is being considered.

6.4.1. Transvenous ICD vs Nontransvenous ICD. The transvenous ICD has shown effectiveness in the treatment and prevention of arrhythmic death. However, transvenous lead placement has been associated with significant complications related to lead insertion (pneumothorax and cardiac perforation) as well as considerable morbidity due to endovascular lead failure. The nontransvenous subcutaneous or extravascular ICD has been developed as an alternative to transvenous ICD, with the placement of the defibrillator electrode circumventing the need to enter the heart and vasculature. Although these features have been associated with lower lead failure rates, extravascular lead placement makes it almost impossible for the device to deliver chronic pacing therapy, although ATP is possible with the placement of a substernal lead or compatible adjunctive leadless pacemaker implant. As such, these devices have been associated with higher rates of inappropriate shocks because of errors in discrimination or oversensing.²²⁹⁻²³¹

To date there have been 2 large trials that have compared transvenous with subcutaneous ICD therapy. The Prospective

Randomized Comparison of Subcutaneous and Transvenous Implantable Cardioverter Defibrillator Therapy (PRAETORIAN) trial randomized 876 patients with a class I indication for ICD therapy to either the subcutaneous ICD or transvenous ICD. At 48 months, the cumulative incidence of the primary end point (composite of device-related complications and inappropriate shocks) was 15.1% in the subcutaneous ICD group and 15.7% in the transvenous ICD group (hazard ratio, 0.99; 95% CI, 0.71-1.39; $P = 0.01$ for non-inferiority).²³² The Avoid Transvenous Leads in Appropriate Subjects (ATLAS) trial randomized 544 patients with a primary or secondary prevention indication for an ICD to either the subcutaneous ICD or transvenous ICD. At 6 months there was a statistically significant reduction in perioperative lead-related complications with the subcutaneous ICD (0.4% vs 4.8%; $P = 0.001$).²³³ Similar to PRAETORIAN, there was a trend for more inappropriate shocks with the subcutaneous ICD (hazard ratio, 2.37; 95% CI, 0.98-5.77 in ATLAS and hazard ratio, 1.43; 95% CI, 0.89-2.30 in PRAETORIAN).

Strategies to minimize inappropriate shocks with the subcutaneous ICD include: (1) rigorous preimplant ECG screening with optimization of vectors to achieve the best signal-to-noise ratio²³⁴; (2) meticulous surgical technique including lead anchoring to prevent postimplant lead movement²³⁵ and complete air expulsion²³⁶; (3) the use of high ventricular rate cutoffs (rates > 220 beats per minute) to avoid therapies for sinus and supraventricular tachycardia²³⁷; (4) the use of dual-zone programming, with application of the subcutaneous ICD double-detection discrimination algorithm in the conditional zone²³⁸; (5) the use of a morphology-based algorithm and a 9-Hz high-pass filter (SMART pass; Boston Scientific, Marlborough, MA)²³⁹; and (6) acquisition of a template during exercise stress testing to optimize sensing vector to minimize T-wave oversensing.²⁴⁰ These approaches have markedly decreased the subcutaneous ICD inappropriate shock rates, decreasing from 13.1%-25% in the early Investigational Device Exemption (IDE) trials, to 7% in the Evaluation of Factors Impacting Clinical Outcome and Cost Effectiveness of the S-ICD (EFFORTLESS) registry (dual zone programming and high rate cutoffs), and 3.1%-4.8% in the PRAETORIAN trial and Understanding Outcomes with the S-ICD in Primary Prevention Patients with Low Ejection Fraction (UNTOUCHED) study.^{229,232,241} However, although the inappropriate shock rate has decreased, it is important to realize that the cumulative incidence of inappropriate therapies remains substantial, with nearly 1 in 10 patients receiving an inappropriate shock by 48 months in PRAETORIAN.²³²

The extravascular ICD is a nontransvenous defibrillator with a lead placed behind the sternum. Results from the pivotal trial and short-term follow-up have shown that this novel device is safe and capable of delivering effective ATP and low-energy defibrillation in patients without previous sternotomy.^{242,243} The early experience with extravascular ICDs has faced similar challenges with inappropriate shocks, with a rate of 9.8% at 1 year in the pivotal study, driven primarily by P-wave oversensing. Similar to the subcutaneous ICD, this is expected to improve significantly with implanter experience and novel P-wave oversensing discriminators now available in the commercially available model.²⁴⁴

7. Device Therapy in Patients With an LV Assist Device

Recommendation

74. We suggest that ICDs might be considered in patients with an LV assist device (LVAD) (Weak Recommendation; Low-Quality Evidence).

Practical tip. Existing observational evidence has not shown a clear survival benefit for ICD therapy in patients with continuous flow (CF)-LVADs.

LVADs improve survival and minimize morbidity in patients with end-stage HF. Most patients who receive an LVAD for end-stage cardiomyopathy have a preexisting ICD and/or CRT-capable device.

It is common practice to maintain device therapies after LVAD implantation because ventricular arrhythmias are common in this population (incidence 20%-50%), either because of preexisting myocardial substrate abnormalities or complex electrical remodelling after LVAD implantation. However, the effect on mortality is uncertain. In the era of pulsatile flow LVADs there was a reported survival advantage associated with ICD therapy. However, the results have been conflictual in the presence of CF-LVADs. A propensity score-matched comparison of 2209 patients with a CF-LVAD from the Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) registry showed that the presence of an ICD was associated with an increased mortality risk (hazard ratio, 1.20; 95% CI, 1.04-1.39; $P = 0.013$) and an increased risk of unexpected death during LVAD support (hazard ratio, 1.33; 95% CI, 1.03-1.71; $P = 0.03$). Conversely, a meta-analysis of 6 observational studies (937 patients) showed that the presence of an ICD was associated with a 39% RR reduction in all-cause mortality (RR, 0.61; 95% CI, 0.46-0.82; $P < 0.01$), however, among a subgroup of patients with CF-LVAD ($n = 361$) ICD use was not associated with a significant effect on survival (RR, 0.76; 95% CI, 0.51-1.12; $P = 0.17$).²⁴⁵ As such, despite lack of randomized data, it is reasonable to interpret the existing observational evidence as showing no clear survival benefit for ICD therapy in patients with CF-LVADs. As such, the decision to continue with ICD therapy after CF-LVAD placement is usually evaluated on a case-by-case basis. In those who elect to continue with ICD therapy it is reasonable to use a conservative programming strategy with high-rate cutoffs (VF zone of 250 bpm) and long detection times to minimize ICD shocks for hemodynamically tolerated VT and/or VF. Patients with a history of poorly tolerated ventricular arrhythmias might benefit from the additional use of a secondary ATP-only treatment zone. Conversely, because of the lack of mortality advantage, after shared decision-making, certain patients might elect to have their ICD therapies programmed off to minimize painful ICD shocks.

Similarly, many patients who undergo LVAD insertion have a preexisting CRT device. Although CRT might theoretically aid myocardial recovery, the existing observational evidence, albeit from small studies, has not shown any conclusive benefit to CRT therapy on all-cause mortality or

Table 8. Expected event rates associated with cardiac implantable device implants

	Event	Rate	Comment	Preventative strategy
Overall	Major complication	5%-8%		
	Minor complication	4%-7%		
Lead-related	Pneumothorax	1.0%	Necessitating intervention: 0.5%	Cephalic vein access Imaging (venogram or ultrasound)-guided axillary or subclavian access Septal lead placement
	Cardiac perforation	0.5%		Appropriate slack, fixation
	Lead dislodgement	3.5%	Necessitating intervention: 3.0%	Avoid medial puncture
	Lead dysfunction	1.5%	Necessitating intervention: 0.9%	
	Venous thrombosis	0.5%		
	Severe tricuspid regurgitation	1.2%		
Infection	Systemic infection	0.8%		Preprocedural intravenous antibiotics
	Pocket infection	0.9%		Antibacterial envelope
	Superficial infection	0.5%		
Pocket-related	Hematoma or bleeding	2.0%	Necessitating intervention: 0.3%	Uninterrupted VKA Minimally interrupted DOAC Meticulous hemostasis Intraprocedural tamponade with antibiotic-impregnated sponge Topical thrombin powder Compressive dressing Antiarrhythmic drug therapy Catheter ablation Programming (section 10)
Other (system specific)	Appropriate shock (ICD)	5%-10%	Variable on the basis of indication	
	Inappropriate shock (ICD)	1%-3% per year	Variable on the basis of indication and device	
	Phrenic stimulation (CRT)	2.5% (overall)	Necessitating reintervention: 0.5%	Programming
	Inability to place lead (CRT)	3.0%		Alternative site (section 4.2.7.)
Performance-related	Anticipated battery longevity	Pacemaker ICD CRT	10 Years 8 Years 6 Years	
	Anticipated risk of generator performance abnormality	Pacemaker ICD	Failure rate 0.05% per year Failure rate 0.2% per year	
	Anticipated risk of lead performance abnormality	Pacemaker ICD	Failure rate 0.3% per year Failure rate 0.7% per year	

CRT, cardiac resynchronization therapy; DOAC, direct oral anticoagulant; ICD, implantable cardioverter defibrillator; VKA, vitamin K antagonist.

Modified from Andrade et al.²⁴⁸

hospitalization, with conflicting evidence on freedom from VT and/or VF ICD discharges. As such, it might be reasonable to deactivate LV pacing after LVAD implantation to conserve battery and to avoid pulse generator replacement.

In patients with existing CIEDs at the end of expected battery life, the decision to perform pulse generator replacement must take the anticipated benefit of generator replacement into consideration vs the procedural risks (eg, increased rate of infection).

8. Complications Related to CIED Therapy

Complications are an inevitable consequence of any interventional procedure.^{246,247} Before implantation, patients should be informed of the nature of the procedure, the risks and benefits of the treatment, and advised of other reasonable treatment options, if available. This discussion should include disclosure of the periprocedural risks, the expected device and/or generator longevity (eg, battery life), the lead and generator reliability expectations, and the potential for CIED system performance abnormalities.²⁴⁸

The rate of major complications associated with a CIED implantation or repeat intervention procedure has been reported to be 5%-8%, with a 4%-7% rate of minor complications.²⁴⁹⁻²⁵⁴ Isolated generator replacement has been associated with the lowest risk of major complications (4.4%; 95% CI, 0.5-15.2), however the risk of major complications increases significantly with revision of a single-chamber to a

dual-chamber device (11.1%; 95% CI, 7.4-15.9) or to a CRT device (18.7%; 95% CI, 15.1-22.6).²⁵¹ As such, the short- and long-term risks, including device-related infection, effects of pacing modalities on battery longevity, tricuspid valve insufficiency, and future lead management issues require careful consideration before intervention.

When a device is evaluated for replacement, these issues require revisiting to ensure that the appropriate device is replaced, or consideration to no replacement when indicated (see section 9.6).

Strategies to minimize the risk of periprocedural and long-term device-related complications are presented in Table 8.

8.1. Infection prevention

Despite an overall decrease in the rate of CIED-related complications, the rate of CIED-related infection has increased out of proportion to the overall increases in device utilization. The reasons for this are unclear but likely reflect a combination of changing population demographic characteristics (eg, an increasingly elderly population with a greater proportion of comorbidities such as chronic kidney disease, and immunosuppressant use) as well as the increasing need for device revision or replacement. Contemporary series report the incidence of CIED infection varies from < 1% for pacemakers to 1.9% for ICDs, 2.2% for CRT-pacemakers and 3.4% for CRT-defibrillators, with a greater incidence

of infection being observed for device replacements, compared with *de novo* implants.²⁴⁹⁻²⁵⁴

Cardiac device infections can vary from local pocket infection (ie, erythema or frank purulent discharge of the generator-pocket site) and/or cutaneous mechanical erosion of the generator and/or leads, to systemic septicemia with or without endocarditis. CIED infection has a major effect on morbidity, quality of life, mortality, and health care system cost. An infection of any component of the CIED system (ie, pocket, device, or leads) necessitates complete removal of all prosthetic material regardless of location (subcutaneous, transvenous, or epicardial). Complete removal of hardware is required to achieve long-term cure because the retention of residual prosthetic material results in high rates of relapse due to metastatic seeding, even in the face of long-term suppressive antibiotic use (see section 11). A detailed review of the diagnosis and management of CIED infection is beyond the scope of the current guideline and can be found in the recently published review from Phillips et al.²⁵⁵

As such, there has been great effort directed at measures to reduce device system infection. Prevention of CIED infection begins with proper surgical technique and skin preparation. It is recommended to shave the skin as close to the location of the procedure as possible with an electrical shaver. The skin should be washed with soap or chlorhexidine before surgery. Skin preparation with chlorhexidine 2% in 70% isopropyl alcohol is preferable over povidone-iodine solutions. Intravenous administration of antimicrobial agents 1 hour before skin incision has been shown to reduce the infection rate. Either cefazolin or vancomycin may be used in most patients before skin incision, with the choice of antibiotics varying according to local prevalence of methicillin-resistant *Staphylococcus aureus* (MRSA). In MRSA carriers, preoperative (3-5 days) nasal application of mupirocin could be considered. Although there is no evidence that the routine use of antibacterial pocket washes or postoperative antibiotic therapy reduces the infec-

tion rate, there might be a role for augmented therapy in patients with risk factors for device-related infection (Table 9) or at objective high risk of device-related infection (eg, Prevention of Arrhythmia Device Infection Trial [PADIT] risk score or BLISTER risk score ≥ 6 predicts a $> 3\%$ - 3.5% risk of infection at 1 year). In patients in whom the predicted probability of infection is high the use of augmented strategies to reduce infection, including an antibacterial envelope or an incremental antibiotic strategy might be considered.

Recommendation

75. We recommend the use of comprehensive CIED infection prevention measures in all patients who undergo device implantation (Strong Recommendation; Moderate-Quality Evidence).

Practical tips

- Comprehensive infection prevention measures include the use of an electric shaver and chlorhexidine skin preparation, administration and completion of antibiotic infusion before the initial skin incision with an antibiotic that covers the organisms with which the patient might be colonized (including MRSA if appropriate), the use of an iodine-impregnated barrier, and rigorous hemostasis.
- In patients at higher risk of infection an incremental antibiotic strategy that includes intraoperative wound pocket wash before skin closure (bacitracin 50,000 U diluted in 50 mL of saline or cefazolin 1 g diluted in 20 mL of saline) and postoperative oral cephalexin 500 mg 4 times per day, or clindamycin 150-300 mg at 3 times per day (if penicillin-allergic) for 2 days might be considered.
- The cost of antibacterial envelope usage must be weighed against the relative reduction in CIED infection, whereby the antibacterial envelope is likely cost-effective when the rate of CIED infection is $\geq 3\%$.

Table 9. Risk factors for infection

Factor	
Patient-related	Age
	Heart failure
	Diabetes
	Chronic kidney disease
	Immunocompromised or receiving immunosuppression
	Anticoagulation
	Preprocedural temporary pacing
	Previous device procedures (increasing risk with number of procedures)
	Ongoing systemic infection
	Device type (CRT higher risk than ICD or pacemaker)
Procedural	System revision procedure
	Failure to administer perioperative antimicrobial agents
Postprocedural	Prolonged procedure time (eg, ≥ 120 minutes)
	Clinically significant hematoma
	Poor wound care

CRT, cardiac resynchronization therapy; ICD, implantable cardioverter defibrillator.

Recommendation

76. We suggest the use of an antibacterial envelope be considered for CIED procedures in patients in whom a high rate of infection is anticipated (Weak Recommendation; Moderate-Quality Evidence).

Practical tip. The risk of device-related infection can be evaluated with risk factors assessment (Table 9) or estimated using established risk prediction models (eg, the PADIT risk score or the BLISTER risk score).

Table 10. Factors associated with increased risk of hematoma peri-CIED procedures

	Factor	Recommended intervention
Preprocedural	INR > 3.0 Dual antiplatelet therapy Combination antiplatelet therapy and anticoagulation	Careful management of INR preprocedure to ensure it is within range Consider holding P2Y ₁₂ inhibitor Consider interrupting antiplatelet agent preprocedure if no strong indication for combination therapy
Patient-related	Renal dysfunction Thrombocytopenia Body mass index > 25 Coagulopathy Diabetes Dialysis Number of previous procedures	Nonmodifiable
Intraprocedural	Pocket debridement and/or capsulectomy Submuscular pocket	Careful surgical control of bleeding and avoid routine capsulectomy Consider use of surgical hemostatic agents (topical tranexamic acid, hemostatic sponge, resorbable agents)
Postprocedural	Use of heparin (unfractionated or low molecular weight)	Avoid these postprocedurally for at least 7 days

CIED, cardiac implantable electronic device; INR, international normalized ratio; P2Y₁₂, purinergic receptor P2Y, G-protein coupled 12.

8.2. Hematoma prevention

Recommendation

77. We recommend that in patients taking warfarin for whom perioperative anticoagulation is deemed necessary, continued warfarin is recommended over the use of heparin-based bridging (Strong Recommendation; High-Quality Evidence).

Best Practice Statement

- Minimal interruption of anticoagulation (eg, holding a dose the night before and/or morning of procedure) might be considered for patients receiving a direct oral anticoagulant (DOAC), as per operator preference and on the basis of balance of thromboembolic and bleeding risk.
- Interruption of antiplatelet therapy in advance of an elective procedure might be considered for patients taking concomitant anticoagulation without a strong indication for dual therapy.

Device pocket hematomas are a common complication of CIED procedures, particularly in patients who receive perioperative anticoagulation.^{256,257} Device pocket hematomas

increase the risk of CIED-system infection, potentially necessitate interruption of anticoagulation therapy (increasing the risk of thromboembolism in patients at high risk of stroke and/or systemic embolism), and increase the risk of hospitalization and secondary intervention (eg, pocket evacuation).²⁵⁸ In patients at increased risk for pocket hematoma (Table 10) it is recommended that operators use hematoma prevention techniques, which might include meticulous hemostasis, intraoperative pocket packing with antibiotic-impregnated sponges, topical thrombin, and/or a postoperative pressure dressing for 24 hours. In patients receiving chronic vitamin K antagonist therapy, the randomized Bridge or Continue Coumadin for Device Surgery Randomized Controlled Trial (BRUISE-CONTROL) trial showed that device implantation with a therapeutic international normalized ratio was associated with a significant reduction in the risk of hematomas (3.5%) compared with heparin bridging (16.0%; RR, 0.19; 95% CI, 0.10-0.36; $P < 0.001$).²⁵⁷ The randomized BRUISE-CONTROL 2 trial showed that patients randomized to continued or minimally interrupted DOACs had a similar rate of perioperative hematoma (2.1% in both groups; $P = 0.97$).²⁵⁶ In both studies there was no substantial thromboembolic complications observed. A secondary analysis of BRUISE-CONTROL 1 and 2 showed no significant difference in the rate of clinically significant hematoma between use of DOACs and continued warfarin (OR, 0.86; 95% CI, 0.38-1.96; $P = 0.72$), however, antiplatelet use was associated with an almost doubling in the rate of clinically significant hematoma even after multivariable adjustment (OR, 1.97; 95% CI, 1.20-3.21; $P = 0.007$).²⁵⁹

Of note, the BRUISE-CONTROL trials were performed for transvenous devices. The safety and efficacy of

Table 11. Recommended device follow-up schedule

	In-person follow-up alone	In-person and remote follow-up
Postimplant	Within 2 months after implantation	
Standard pacemaker	In office every 6-12 months until 1 year from ERI then every 3 months	In office every 12-24 months Remote every 6 months until 1 year from ERI then every 3 months
Complex device*	In office every 3-6 months until 1 year from ERI then every 3 months	In office every 12 months Remote every 3-6 months until 1 year from ERI then every 3 months

ERI, elective replacement indicator.

* Complex device defined as a defibrillator, conduction system pacemaker, or cardiac resynchronization pacemaker.

uninterrupted DOAC therapy has not been studied for patients who undergo subpectoral generator placement or subcutaneous ICD implantation.

9. Postimplant Management

9.1. Follow-up schedule

The timing and frequency of CIED follow-up is determined by several factors related to the patient and their device (Table 11). Follow-up frequency is generally divided into 3 phases: (1) the early postimplant period, in which follow-up enables assessment and management of periprocedural complications (assessment of incision, pocket, and lead parameters), and programming and/or pacing output can be optimized; (2) the maintenance period, in which device functionality and optimization can occur on a reduced frequency until the device approaches end of anticipated battery life; and (3) intensified follow-up, in which the device follow-up occurs at increased frequency until the device reaches elective replacement. The frequency of follow-up during the maintenance phase is largely determined by device complexity (eg, annual for standard pacemakers, and every 6 months for complex devices), follow-up mode (remote vs in-person), and device performance (more frequent if under advisory or a

device performance abnormality is noted). The goals of device follow-up include optimization of CIED system function to meet the patient’s clinical need, optimization of programming to maximize pulse generator longevity while maintaining patient safety, and the identification and correction of abnormal CIED system behaviour (including abnormalities due to the generator, and/or leads), and optimization of the patients’ underlying disease (eg, GDMT; Fig. 10).

Of note, the transition to the intensified follow-up phase should prompt a conversation with patients regarding their values and goals of care and enable a reassessment of the current device therapy and/or need for device revision (sections 9.6 and 9.7).

9.2. Remote monitoring

Recommendation

78. We recommend remote monitoring be used as part of the standard of care for all CIED patients (Strong Recommendation; Moderate-Quality Evidence).

Device Follow-up			
Surveillance		Optimization	
Early	Clinical: 1. Implant Complications 2. Response to Device Therapy	Clinical: 1. Underlying Condition a. Pharmacotherapy b. Risk Factor Modification 2. Treatment of New Problems Identified at Follow-up a. SCAF b. Ventricular Arrhythmias c. AF Limiting BiV Pacing d. PVCs Limiting BiV Pacing	Device: 1. All Devices a. Pacing Output b. Rate Response i. Chronotropic insufficiency 2. Non-CRT, Non-CSP a. Minimize RV pacing a. AV delays b. Dedicated Algorithms 3. CSP or CRT a. AA Optimization b. VV Optimization c. Unipolar/Bipolar Pacing* 4. ICD a. Optimize Detection and Therapy
	Device: 1. Pacing Percentage a. Atrium b. RV – Pacemaker/ICD c. RV – CSP d. BiV/LV – CRT 2. Lead Parameters a. Sensing b. Threshold c. Impedance 3. Battery Longevity 4. Arrhythmia Diagnostics a. Atrial b. Ventricular 5. Alerts / Device Advisory		
Maintenance	Clinical: 1. Response to Device Therapy a. Chronotropic Insufficiency b. Presyncope/Syncope c. Symptoms of HF d. Functional Capacity 2. Tachycardia Therapies (ICD) a. ICD Shock 3. LV Function (Echo) a. 6-12 Months Post CRT/CSP Implant or if >20%-40% RV Pacing is Observed		
Intensified	Above Plus: 1. Reevaluate Initial Indication for CIED Implantation – Pacemaker, ICD, CRT 2. Reevaluate Goals of Care – eg, Desire to Continue with High-Voltage (ICD) Therapies 3. Evaluate Response to Device Therapy – consider LV Lead Repositioning or Addition of CSP Lead for CRT Nonresponders 4. Obtain ECG to Determine the QRS Duration 5. Obtain Echo for LV Function to Determine Need for: a. System Revision to ICD in Presence of Severe LV Dysfunction or Presence of Ventricular Arrhythmia b. System Revision to CRT in Those with Frequent RV Pacing or LV Dysfunction with Prolonged QRS Duration		

Figure 10. Device management after implantation, including surveillance and therapeutic optimization. AF, atrial fibrillation; AV, atrioventricular; BiV, biventricular; CIED, cardiac implantable electronic device; CRT, cardiac resynchronization therapy; CSP, conduction system pacing; ECG, electrocardiogram; echo, echocardiography; HF, heart failure; ICD, implantable cardioverter defibrillator; LV, left ventricle; PVC, premature ventricular complex; RV, right ventricle; SCAF, subclinical atrial fibrillation. *For conduction system pacing leads

Values and preferences. The recommendation places great value on the ability of remote monitoring to more quickly identify actionable CIED issues (eg, routine surveillance of lead function and battery status to ensure device integrity), reduce direct patient costs related to follow-up visits, and improve device clinic workflow and efficiency.

79. We recommend that remote monitoring be used to supplement in-person monitoring of the patient and device (Strong Recommendation; Low-Quality Evidence).

Values and preferences. This recommendation places value on the ability of remote monitoring to more frequently monitor CIED status and to quickly detect CIED system abnormalities, which is particularly relevant in clinical circumstances that warrant more intensive surveillance of the CIED.

Best Practice Statement

7. Remote monitoring should be initiated before discharge and/or established at first follow-up visit after CIED implantation.

Practical tip. The benefits of remote monitoring are more clearly realized in the context of continuous, rather than intermittent, connectivity.

Remote CIED monitoring offers numerous benefits to patients and clinicians across the spectrum of device care. In addition to reducing the need for in-clinic visits, remote monitoring enables early detection of critical or adverse events, and enhances surveillance of device functioning through the early detection of performance abnormalities such as battery depletion, circuit disruption, and lead failure.²⁶⁰ Compared with usual care remote monitoring has been shown to reduce the time to medical decision, with secondary benefit of reducing adverse outcomes such as stroke and HF.²⁶⁰⁻²⁶² Likewise, remote monitoring of ICDs has been shown to reduce the incidence of inappropriate ICD shocks through the early identification of AF with rapid ventricular response, T-wave oversensing, electromagnetic interference, and device malfunction.²⁶³ The use of daily automated remote monitoring has been associated with a significant reduction in all-cause mortality (hazard ratio, 0.65; 95% CI, 0.45-0.94; $P = 0.021$).²⁶⁴ Economic studies from health systems have shown cost-effectiveness and increased patient satisfaction associated with remote monitoring-facilitated follow up.^{265,266} Patients should be enrolled in remote monitoring as early as possible after device implantation, and preferably before hospital discharge to enable timely intervention.

A detailed review of the evidence supporting remote monitoring can be found in the recently published Heart Rhythm Society/European Heart Rhythm Association/Asia Pacific Heart Rhythm Society/Latin American Heart Rhythm Society expert consensus statement on practical management of the remote device clinic.²⁶⁷

9.3. Imaging and CIEDs

Recommendation

80. We recommend that all patients with CIEDs who undergo MRI be managed according to a standardized protocol (Strong Recommendation; Moderate-Quality Evidence).

Values and preferences. Large studies have shown that the use of a standardized protocol for device management of patients with conditional and nonconditional CIEDs who undergo MRI minimizes risk of adverse events.

81. We recommend for patients with a previous CIED who undergo MRI, we recommend reviewing all available medical information, including chest radiography, to identify lead extenders, retained epicardial leads, abandoned transvenous leads, and/or fractured leads. We recommend that patients with lead extenders or fractured leads should not undergo MRI. However, MRI might be considered in patients with epicardial or abandoned transvenous leads when the clinical need is strong and believed to outweigh potential risks (Weak Recommendation; Low-Quality Evidence).
82. We recommend for patients with an MRI-conditional CIED who undergo MRI, that scans be performed at 1.5 T when possible, in a monitored setting and with access to the device team. The device team will provide prescan programming to MRI-compatible mode as well as postscan return to prescan settings (Strong Recommendation; Low-Quality Evidence).
83. We recommend for patients with subcutaneous ICDs, scanning at 1.5 T in a monitored setting, supervised by a health care practitioner trained in advanced cardiac life support (ACLS) and with access to the code and device teams. The device team will provide prescan programming to turn off tachycardia therapies, postscan interrogation, and reprogramming to prescan settings (Strong Recommendation; Low-Quality Evidence).
84. We recommend for patients with leadless pacemakers, scanning at an MRI field strength that does not exceed the manufacturer's specification, in a monitored setting and with access to the device team. The device team will provide prescan programming and postscan reprogramming to prescan settings (Strong Recommendation; Low-Quality Evidence).
85. We recommend for patients with an MRI-nonconditional pacemaker and no pacemaker dependency who undergo MRI, that scans be performed at 1.5 T in a monitored setting and with access to the device team. The device team will provide prescan programming to sense mode (ie, OAO, OVO, or ODO) and postscan interrogation and reprogramming to prescan settings (Strong Recommendation; High-Quality Evidence).
86. We recommend for patients with an MRI-nonconditional pacemaker and pacemaker

dependency who undergo MRI, that scans be performed at 1.5 T in a monitored setting, supervised by a health care practitioner trained in ACLS and with access to the code and device teams. The device team will provide prescan programming to asynchronous mode (ie, AOO, VOO or DOO) and postscan interrogation and reprogramming to prescan settings (Strong Recommendation; High-Quality Evidence).

87. We recommend for patients with MRI-nonconditional implantable transvenous defibrillators who undergo MRI, that scans be performed at 1.5 T in a monitored setting, supervised by a health care practitioner trained in ACLS and with access to the code and device teams. The device team will provide prescan programming to turn off tachycardia therapies and postscan interrogation and reprogramming to prescan settings (Strong Recommendation; High-Quality Evidence).

MRI can be safely used in patients with CIEDs, including transvenous or leadless pacemakers, transvenous or non-transvenous defibrillators, and ICMs.⁸ Most CIED components currently commercialized have been tested and classified as MRI-compatible or conditional. Despite lacking formal labelling, observational studies have shown that MRI in nonconditional pacemaker and defibrillator systems, including manufacturer mismatched systems (eg, containing MRI-conditional components from different manufacturers), is associated with low lead failure and clinical event rates.²⁶⁸ Conversely, MRI of systems with abandoned leads, fractured leads, epicardial leads, or lead extenders remains controversial because of the risk of thermal injury. Previously extraction would have been required in these patients before MRI, however evidence has accumulated to show clinical safety in performing MRI in patients with these systems if performed under proper monitoring conditions and supervision of a device clinic with proper protocols.²⁶⁹⁻²⁷²

Best Practice Statement

8. MRI might be considered in patients with epicardial or abandoned transvenous leads when the clinical need is strong and believed to outweigh potential risks.

A detailed review of the evidence supporting MRI in patients with CIEDs can be found in the 2021 CCS/Canadian Society of Cardiovascular MRI/CHRS update on safety of MRI position statement.⁸

9.4. Perioperative management of patients with CIEDs

Patients with CIEDs present unique challenges when undergoing surgical and nonsurgical procedures. It is important to involve the CIED management team before surgery to assist in identifying high-risk patients (eg,

Table 12. Perioperative management of patients with CIEDs

	Action
Preprocedure evaluation	1. Determine the CIED type (ICM, pacemaker, ICD, CRT), manufacturer, and indication for implant 2. Determine pacemaker dependency 3. Determine CIED settings and functionality (eg, interrogate device or obtain most recent interrogation report if recently assessed) 4. Consider the anatomic location of the surgery 5. Consider possibility of EMI from electrocautery or other sources 6. Consider consulting with the CIED management team before surgery
Preoperative preparation	1. Preoperative reprogramming* A. Program asynchronous pacing in dependent patients (or consider a magnet for pacemaker patients) B. Suspend ATP and/or shock functionality for ICD (reprogramming or magnet, as appropriate) 2. Determine the anticipated and potential sources of electromagnetic interference 3. Ensure availability of external pacing and defibrillation equipment
Intraoperative management	1. Perform continuous electrocardiographic rhythm monitoring and continuous oxygen saturation measured using pulse oximetry monitoring during the surgical and/or interventional procedure (from room entry to exit). Consider peripheral pulse monitoring (eg, pulse palpitation, pulse oximeter plethysmography, or arterial line) in appropriate cases) 2. Electrocautery A. Position the dispersive electrode so that the current pathway does not pass through or near the generator and/or leads B. Avoid electrocautery near the generator and/or leads C. Use short, intermittent, and irregular bursts of electrocautery at the lowest energy level possible D. Use bipolar cautery in preference to monopolar cautery
Postoperative management	1. Perform a device interrogation with or without reprogramming if: A. Device was reprogrammed pre- or intraoperatively B. Concern for potential CIED malfunction i. Significant EMI occurred ii. Electrocautery was performed near the CIED generator or lead iii. Thoracic surgery C. Delivery of ATP and/or shock therapy was observed or suspected

ATP, antitachycardia pacing; CIED, cardiac implantable electronic device; CRT, cardiac resynchronization therapy; EMI, electromagnetic interference; ICD, implantable cardioverter defibrillator; ICM, implantable cardiac monitor.

*Consider pacemaker reprogramming if dependent and prolonged diathermy close to the CIED system (surgery involving the head, neck, thorax, upper abdomen, upper limb proximal to elbow, endoscopy with prolonged diathermy and/or argon beam, nerve conduction studies with repetitive nerve stimulation) or where magnet application might not be adequately stable because of the patient's positioning. Pacemaker reprogramming is unlikely to be necessary for surgery involving the lower abdomen, lower limbs, upper arm distal to elbow, lithotripsy, electroconvulsive therapy, or dental surgery. Dependent patients with ICD reprogrammed into asynchronous pacing mode and patients with ICDs in which ATP and/or shock is suspended must be maintained using a cardiac monitor with availability of external pacing and defibrillation equipment until the device's permanent and/or baseline programming is restored.

dependent patients or patients at risk of malignant arrhythmia) and guide the perioperative management of CIEDs. The most common CIED management strategies involve the use of an external magnet to temporarily activate an asynchronous pacing mode (only pacemakers) or to

withhold ATP and/or shock therapies (ICD), although it is important to note that Micra leadless pacemakers (Medtronic, Dublin, Ireland) do not respond to an external magnet, whereas the Aveir (Abbott, Chicago, IL) does. Alternatively, devices might need to be reprogrammed by the CIED team before surgery and then retested and reprogrammed postoperatively (Table 12).

Recommendation

88. We recommend that patients with implantable cardiac devices *in situ* should be evaluated for programming changes before surgery and again after surgery, in accordance with existing CCS recommendations (Strong Recommendation; Low-Quality Evidence).

9.5. Timing of LV reassessment

RV pacing is associated with mechanical and electrical dyssynchrony, potentially leading to deterioration of the LVEF and symptoms of HF in patients with previously normal LVEF. The incidence of PICM ranges from 10% to 20%, with a high burden of RV pacing (eg, > 20%–40%) being the sole well established risk factor. Evidence suggests that a decline in LVEF can manifest within hours of RV pacing, although most studies report a change in LVEF occurring within the initial 6–12 months. In addition, it is our recommendation that LV function be reevaluated within the last year of battery longevity to guide the need for device system revision (eg, to ICD or CRT in the presence of new significant cardiomyopathy).

Biventricular CRT and CSP has been shown to reduce ventricular dyssynchrony and improve ejection fraction in patients with reduced LVEF. The timing for reassessing LVEF after optimal lead placement remains unclear. Although an acute improvement in LVEF and mitral regurgitation can be immediate after device insertion because of restoration of VV synchrony, further improvement typically occurs in a gradual manner in concert with resynchronization-induced reverse remodelling. Although most studies report a 1-year follow-up LVEF reassessment, improvement can be observed at 3–6 months.

Recommendation

89. We recommend that all patients who are approaching elective replacement of a CIED should undergo comprehensive reevaluation by specialists with expertise in the management of CIED therapy (Strong Recommendation; Moderate-Quality Evidence).

9.6. Device replacement considerations

Practical tip. Evaluation should include reevaluation of treatment goals, evaluation of the anticipated risk and benefits of device replacement, assessment of the indication for device revision, and be informed by CIED interrogation (lead performance, pacing dependence, RV pacing burden and

percentage, and the arrhythmia and therapy history [ATP and/or shocks]).

Recommendation

90. We recommend that all patients who are planned to receive a CIED system intervention should be considered for their eligibility for revision to ICD or CRT (Strong Recommendation; Low-Quality Evidence).

Practical tips

- Careful evaluation of the risks and benefits of revision should be considered at the time of CIED system intervention (eg, generator change or lead procedure).
- Investigations that inform decisions related to device revision include: CIED interrogation, 12-lead ECG to record QRS duration (intrinsic or paced), and echocardiogram to document LVEF and quantify severity of any valvular regurgitation.
- We recommend the use of the validated risk calculators and/or risk assessment tools to aid in the estimation of each patient's benefit and risk of an ICD. Examples include the Seattle Proportional Risk Score, BaSIS Score, DO-IT model, and the MADIT ICD benefit risk scores.

Recommendation

91. We recommend that an ICD be replaced by a pacing system, removed, or abandoned at the time of ICD generator end of life and/or service in patients with life-threatening comorbidities, at the request of the patient or when an ICD is unlikely to increase life expectancy (Strong Recommendation; Low-Quality Evidence).

Because of their reliance on battery technology, CIED generators will eventually require replacement. The more reinterventions made in a pocket, the higher the risk of infection.²⁴⁶ Although a generally well tolerated procedure, isolated generator replacement has been associated with potentially significant risk of complications. Single- and multicentre case series have reported an aggregate rate of major complication of 2.6% (95% CI, 1.1%–4.5%), and an aggregate risk of reoperation and/or pocket revision of 2.5% (95% CI, 0.9%–4.5%).²⁷³ When considering device type, the risk of complication is observed to be highest for CRT replacements, intermediate for ICD replacements, and lowest for pacemaker replacements (OR, 2.38 for ICD vs pacemaker; 95% CI, 1.30–4.38; $P = 0.004$, and OR, 3.68 for CRT vs pacemaker; 95% CI, 1.98–6.84; $P < 0.001$).²⁵¹ The presence of frailty or multiple comorbidities are the primary

driver of morbidity and mortality after second or subsequent CIED procedures, conditions that commonly coexist in the CIED population.^{251,274-277} Although age is not specifically a predictor of poor outcome, complications of CIED surgery might be expected to be of greater consequence to the elderly or frail patient, further reducing resiliency and resulting in additional functional decline, disability, and increased hospitalization.²⁷⁸⁻²⁸⁰ In the presence of frailty, treatment goals should prioritize improved functional status (eg, CRT) and/or avoidance of additional symptoms from bradycardia (pacemakers) rather than the avoidance of SCD (eg, ICD).²⁷⁹

When considering the decision to retire, revise, or replace a generator at end of expected battery life it is important to use an individualized approach on the basis of shared decision-making while considering the indication for implantation (eg, brady-pacing, primary or secondary prophylaxis for sudden death, resynchronization), current pacing needs (eg, dependency), the need for concurrent intervention (eg, concurrent lead revision because of performance abnormality, or need for revision to ICD or CRT), and the patient's goals of care. Although almost all patients elect to undergo generator replacement, the decision to retire the CIED (permit the battery to deplete without replacement and/or reprogram the CIED to be physiologically "inert") would be consistent with respect for patient autonomy (see section 9.7).

In the case of index device implantation for bradycardia or resynchronization, elective pulse generator replacement is almost universally performed for symptomatic benefit (eg, to protect against symptomatic bradycardia or HF decompensation).^{281,282}

However, ICD generator replacement at the end of anticipated battery longevity is more often discretionary, with the end of anticipated device longevity prompting a reevaluation of the patient's values and goals of care.²⁸³⁻²⁸⁵ Considerations that factor into the discussion regarding the decision to replace an ICD generator at end of life include the indication for index implant (eg, primary or secondary prevention), whether the device has delivered appropriate therapies for ventricular arrhythmia (ATP or shock), change in risk-benefit profile (eg, increasing noncardiac comorbidities that lead to a relatively greater risk of nonarrhythmic death, or recovery in ventricular function out of range for index ICD candidacy).²⁸⁶ Although clinical risk prediction models are available for primary device implantation, there are few validated data for use at time of generator replacement, despite recognition that approximately half of all patients aged older than 65 years will die within 5 years of ICD pulse generator replacement.²⁸⁷⁻²⁸⁹ Conversely, studies on recovery of LVEF after index device implantation have shown a low but persistent rate of malignant ventricular arrhythmias triggering appropriate device therapy after generator replacement, which suggests that generator replacement should be the default strategy in those with a history of, but not necessarily current ventricular dysfunction.²⁹⁰⁻²⁹⁴ As such, the decision to terminate ICD therapy (eg, revise to pacemaker-only therapy in those with a pacing indication, not replace the ICD generator in patients without a pacing

indication) should be on the basis of shared decision-making, while recognizing that most ICD recipients elect to receive continued protection from sudden arrhythmic death.²⁹⁵⁻²⁹⁷

9.7. End of life management

CIEDs are implanted for the purposes of symptomatic improvement and to improve patient prognosis. However, there might be a time at which patients no longer wish to receive therapies from their cardiac device. The informed decision of the patient to discontinue CIED therapy is legal and ethical and can be made at any time (ie, does not necessitate the presence of terminal illness). Withdrawal of CIED therapy with the informed consent of a patient or legal surrogate, even if it is a life-sustaining medical intervention (eg, pacing in a dependent patient), does not constitute medical assistance in dying, physician-assisted suicide, or euthanasia. There is no ethical or legal difference between choosing to deactivate a CIED (withdrawal) and never pursuing one in the first place (withhold).

In the case of bradycardia devices, it is important to recognize that deactivation of the pacemaker function is unlikely to result in imminent death. A pacemaker is not a resuscitative device and does not prevent death. More appropriately, a pacemaker can be thought of as a palliative device, minimizing the adverse and distressing symptoms associated with inadequate cardiac output such as dyspnea, fatigue, presyncope, and syncope. Terminal events in patients with pacemakers are most often secondary to the underlying illness, with the myocardium becoming unresponsive to the pacemaker-generated signals in the context of end-of-life associated acidosis and hypoxia. As such, it is rarely necessary to deactivate pacemaker function, and in many cases reprogramming the pacemaker to be physiologically "inert" might worsen symptoms.

Conversely, an ICD is designed to terminate sudden arrhythmic events. The decision to deactivate tachycardia therapies would not be expected to hasten death but instead would mean that the ICD would no longer respond to ventricular arrhythmias and would not prevent sudden death. Philosophically, some patients might prefer to die suddenly from a relatively painless arrhythmic event rather than from an alternate mechanism, such as progressive pump failure (HF) or another terminal illness. Moreover, a large proportion of patients who are approaching end of life experience painful ICD shocks, which worsens patient suffering and is distressing to the patient and their family.²⁹⁸⁻³⁰⁰

In some cases, it might be reasonable to continue with some ICD interventions (ie, palliative ATP) even when a decision has been made to disable ICD shock therapy, particularly in the case of patients with slow (conscious) ventricular arrhythmias that result in quality of life deterioration. Likewise, patients with a CRT-defibrillator might choose to disable ICD therapy (ie, remove potentially futile and painful therapy) while continuing to receive cardiac resynchronization with the goal of palliative symptom management.

Table 13. General considerations for programming CIED bradycardia settings

Pacing mode	
Single chamber pacing modes	AOO or VOO, asynchronous pacing without sensing. Useful for temporary programming during surgery or MRI AAI or VVI, single-chamber pacing with inhibition of pacing during sensed events VDD, ventricular pacing mode that is inhibited by either atrial or ventricular sensed events
Dual-chamber pacing modes	DDD, dual-chamber pacing mode designed to maximize AV synchrony with allowance of all permutations of atrial and/or ventricular sensing and pacing DDI, dual-chamber pacing mode in which atrial sensed events do not result in a triggered ventricular paced event; useful when avoidance of ventricular tracking of atrial tachyarrhythmias is desired
Manufacturer-specific algorithms to favour intrinsic AV conduction and minimize RV apical pacing*	• Medtronic (Dublin, Ireland): MVP • Boston Scientific (Marlborough, MA): RhythmIQ[†] or AV search • Abbott (Chicago, IL): VIP • Biotronik (Berlin, Germany): IRS Plus/I OPT or Vp Suppression • Microport (Shanghai, China): SafeR
Rate response programming	Use only in patients with chronotropic insufficiency
Atrial ATP therapies	Medtronic reactive ATP shows decreased AF burden and lower progression to persistent AF

AAI, single chamber atrial pacing and sensing; AF, atrial fibrillation; AOO, single chamber atrial pacing (asynchronous); ATP, antitachycardia pacing; AV, atrioventricular; CIED, cardiac implantable electronic device; DDD, dual chamber pacing and sensing; DDI, dual chamber pacing and sensing, but does not track atrium; MRI, magnetic resonance imaging; MVP, Managed Ventricular Pacing; RV, right ventricular; VDD, ventricular pacing with dual chamber sensing; VIP, ventricular intrinsic preference; VOO, single chamber ventricular pacing (asynchronous); VVI, single chamber ventricular pacing and sensing.

* Avoid marked AV prolongation (> 300 ms) to prevent AV uncoupling.

† Avoid in the presence of pacemaker-mediated tachycardia or atrial tachycardia.

10. Evidence-based Programming

Recommendation

92. We recommend CIED programming be individualized and tailored to the patient's needs (Strong Recommendation; High-Quality Evidence).

The goals of CIED programming are to best support the patient's needs, address the indication for CIED therapy, and avoid the delivery of unnecessary and potentially harmful therapies (eg, unnecessary RV pacing or inappropriate ICD therapies).

10.1. Bradycardia programming

RV pacing is associated with mechanical dyssynchrony (abnormal contraction and altered myocardial strain) and electrical dyssynchrony (abnormal activation and conduction delays), potentially leading to adverse cardiac remodelling, LV dysfunction, and, in some cases, adverse cardiovascular

outcomes (eg, AF, HF, and death).^{67,301-304} In those who do not require continuous ventricular pacing (eg, those with SND and preserved AV conduction, and/or intermittent AV conduction block) every effort should be made to preserve the normal ventricular activation sequence. When possible, non-CRT devices (pacemakers and ICDs) should be programmed to avoid RV pacing, either through a preference to atrial pacing (eg, AAI mode), backup low rate VVI pacing in the presence of infrequent ventricular pacing (eg, VVI or DDI 40 per minute for patients who receive a transvenous ICD), and in whom there is no current indication for bradycardia pacing, or the use of manufacturer-specific algorithms to favour intrinsic AV conduction and minimize RV pacing (Table 13). It is important to recognize that these algorithms are useful even in patients with devices implanted primarily for AVB, because patients might exhibit intermittent AV conduction.^{304,305} In these patients dedicated algorithms designed to favour intrinsic AV conduction can be used to reduce RV pacing without compromising patient safety.^{306,307}

Rate-responsive pacing algorithms should be activated with objective chronotropic insufficiency. In the population with SND the use of pacing programmed to a base rate of 60 bpm with rate-adaptive pacing (DDDR-60) does not increase the risk of AF and can decrease the occurrence of syncope or presyncope.³⁰⁸ However, the use of rate response programming should be reserved for patients who would benefit, because unnecessary use of rate responsive programming could result in an unnecessary increase in the burden of pacing with an associated increase in battery drain.

10.2. Tachycardia programming

High-voltage ICD therapies can be life-saving but can also have significant adverse clinical outcomes including decreased quality of life and increased health care utilization. Programming devices to minimize inappropriate and unnecessary shocks by delaying arrhythmia detection (eg, 12-60 seconds), using higher rate detection cutoffs (eg, 180-200 per minute), and using the extensive use of supraventricular tachycardia discriminators has been shown to decrease shocks and, in some cases, improve mortality without a significant increase in the risk of syncope or death.^{231,309,310} Randomized and nonrandomized data have shown increased risks of inappropriate therapies with detection rates slower than 188 bpm, and subsequent data have shown the safety of setting initial detection at 200 per minute.^{231,311}

In ICDs implanted for monomorphic VT, the VT detection rate should be programmed 10-20 bpm below the slowest clinical VT.³¹² Antiarrhythmic drug therapy can slow the rate of VT, therefore if an ICD patient has antiarrhythmic drugs initiated, the detection rate for VT and VF should be decreased to avoid the possibility of subsequent VT below the detection rate.

For transvenous ICDs, ATP therapy has been shown to decrease the risk of shocks for patients with VT, and potentially reduce mortality.³¹³⁻³¹⁶ Programming multiple ATP trains can increase the chance of painless VT termination.^{310,311}

Table 14 shows examples of programming tailored by manufacturers, and Table 15 shows modifications in programming according to indication.

Table 14. Manufacturer-specific tachycardia programming

Manufacturer	Detection*	Therapy	Comments
Abbott (Chicago, IL)	VF 250 bpm 30 Intervals	ATP while charging 8 Pulses at 85% All shocks at maximum output†	SVT discriminators on† SVT upper limit 230 per minute SVT time-out off
	VT2 187-200 bpm 30 Intervals	ATP > 1 burst, 8 pulses at 85% Scan step 10 ms Readaptive “on” All shocks at maximum output†	
	VT 170 bpm§ 36 Intervals	Monitor If used, program therapy like VT2 but favour more ATP	
Biotronik (Berlin, Germany)	VF 231 bpm 30-40 Intervals	ATP one shot 8 pulses at 88% All shocks at maximum output†	SVT discriminators on† SMART for dual-chamber
	VT2 188-200 bpm 30-40 Intervals	ATP ≥ 1 burst 8 pulses at 88% 10 ms scan All shocks at maximum output†	MorphMatch for single-chamber and narrow QRS (or onset 20% and stability 48 ms if poor amplitude)
	VT1 170 bpm§ 40-100 Intervals	Monitor If used, program like VT2 but favour more ATP	
Boston Scientific (Marlborough, MA; transvenous ICD)	VF 250 bpm 8-10 Intervals plus 5-second delay	Quick convert “on” to 300 bpm All shocks at maximum output†	SVT discriminators on† (Rhythm ID) SVT upper limit 230/min
	VT 200 bpm 8-10 intervals plus 12-second delay	ATP > 1 burst, 8 pulses 88% 10-ms decrement All shocks at maximum output†	
	VT1 170-199 bpm§ 30-60 Second delay	Monitor If used, program like VT but favour more ATP	
Boston Scientific (subcutaneous ICD) Medtronic	Shock zone > 250 bpm Conditional zone 200 and 250 bpm§		
	VF 188-200 bpm 30-40 intervals	ATP before charging All shocks at maximum output†	SVT discriminators on† SVT upper limit 230/min
	FVT via VF to 250 bpm to allow tiered ATP programming VT 170 bpm§ 24 Intervals	ATP > 1 burst 8 pulses at 88% 10-ms decrement Monitor If used, program like FVT but favour more ATP	Stability and onset off
Microport (Shanghai, China) CRM	VF > 255 bpm 20 cycles + 6/8 majority	All shocks at maximum output†	SVT discriminators on† Stability/Acc (single chamber)
	FVT 230 bpm 20 cycles + 6/8 majority	ATP × 1 burst at 85% All shocks at maximum output†	PARAD+ (dual-chamber)
	VT 185 bpm 20 or 30 cycles + 6/8 majority Slow VT	ATP × ≥ 1 burst 8 pulses at 85% All shocks at maximum output† Monitor	

For ATP, burst pacing is preferable to ramp because of potential increased efficacy.^{317,318}

ATP, antitachycardia pacing; bpm, beats per minute; FVT, fast VT; ICD, implantable cardioverter defibrillator; SVT, supraventricular tachycardia; VF, ventricular fibrillation; VT, ventricular tachycardia.

* Consider fewer intervals in patients with known VT rates near zone cut-offs or poor sensing as detection may be delayed due to undercounting (either through zone “straddling” or undersensing, respectively).

† When programming shock therapy, it is recommended that full available energy be used for the first shock to decrease the risk of requiring multiple shocks to terminate an event.³¹⁹

‡ Not required in patients with complete heart block.

§ Program at 10-20 beats lower than the clinical VT cycle length in those with known VT.

10.3. CRT programming

CRT programming should aim to maximize the proportion of time the ventricles are being resynchronized. Studies have shown that clinical outcomes are proportional to the amount of time that CRT therapy is being delivered. Retrospective analyses have shown that a beneficial reduction in HF hospitalization and mortality can be achieved by maintaining a > 92% of biventricular pacing, with survival being significantly improved with higher percentages of biventricular pacing (eg, > 98.5%).^{321,322} Of note, this might not be achieved at times because of rapid AV conduction (eg, long programmed AV delays or AF) or a high burden of PVCs. In

the case of rapid AF, restoration of sinus rhythm with pharmacological therapy or catheter ablation, or intensification of rate control with pharmacological therapy or AV junction ablation can be used to restore CRT delivery and improve clinical outcomes.³²³ Likewise, in patients with very high burden PVCs, pharmacological suppression or catheter ablation can be used to decrease the PVC burden and enable cardiac resynchronization.

Optimization of AV and VV timing using echocardiographic, ECG, or intracardiac electrogram timing has been suggested to improve resynchronization, clinical response rates, and outcomes. Despite limited validation, CRT optimization with echocardiography was included in the landmark trials and

Table 15. Special ICD programming considerations according to indication

Patient with left ventricular assist device	- Consider very long detection delays (eg, 50-60 beats of 10-second detection delay) - ICD therapies should remain activated because sustained ventricular arrhythmias might compromise the venous return to the left ventricle resulting in decreased cardiac output with hemodynamic consequences
Absence of intrinsic atrioventricular conduction or pacemaker-dependent ICD recipient	Consider programming of monitor zone or slowest VT therapy zone at ≥ 160 bpm
Cardiac sarcoidosis	Consider programming of a monitor or first therapy zone at lower rates than usual bpm if sedentary patients
Arrhythmogenic right ventricular cardiomyopathy	Consider programming the first therapy zone in secondary prevention ICDs 20 bpm lower than the heart rate of the clinical VT
Brugada syndrome	- In the absence of need for pacing, non-transvenous ICD should be favoured - Consider a single VF zone with detection rates ≥ 222 bpm³²⁰
Long-QT syndrome	- Assess necessity for atrial pacing if patient receiving β-blocker therapy before ICD implantation - Consider single VF therapy zone with detection rates ≥ 222 bpm
Catecholaminergic polymorphic VT	- Assess necessity for atrial pacing on beta-blocker therapy prior to ICD implantation - Consider a single VF therapy zone with prolonged detection and rates ≥ 222 bpm

bpm, beats per minute; ICD, implantable cardioverter defibrillator; VF, ventricular fibrillation; VT, ventricular tachycardia.

is therefore inherent to the treatment effect.³²⁴ However, a meta-analysis of 12 studies (4356 patients) failed to show a significant benefit in clinical and echocardiographic outcomes with AV and VV optimization compared with empirical device programming, which suggests that the benefit of routine echocardiographic CRT optimization is limited, and that echocardiographic optimization should be reserved for selected cases of nonresponders.^{325,326} In response to the complex and time-consuming echocardiographic optimization efforts, more recent approaches have attempted to obtain the narrowest paced QRS complex on 12-lead ECG, an approach that has been shown to correlate with clinical and echocardiographic improvement, including mortality.^{327,328} Previously this was obtained through manual adjustment of the AV and VV intervals or, more recently, using validated automatic device-based algorithms (Table 16).

These algorithms vary significantly and sometimes use LV-only pacing to synchronize LV activation with intrinsic AVN conduction, which includes the benefit of improved battery longevity because RV pacing output is not continuously required.³²⁹ However, no randomized study has shown improvement in clinical end points or echocardiographic response with routine use of these algorithms compared with conventional CRT programming.³²⁹⁻³³² In one randomized trial ECG-based manual AV delay adjustment was used to optimize fusion pacing and significantly beneficial LV remodelling was reported.³³³ Observational studies have

Table 16. Device-based CRT optimization algorithms

SmartDelay (Boston Scientific; Marlborough, MA) AV optimization only	IEGM-based method that uses sensed atrial and paced atrial AV intervals and intrinsic RV to LV conduction time to calculate AV delay to allow for fusion between native septal activation and biventricular pacing
QuickOpt (Abbott; Chicago, IL) AV and VV optimization	IEGM-based method that calculates AV interval on the basis of the length of the RA lead IEGM duration to allow for ventricular pacing to occur after atrial depolarization is complete. VV interval is calculated by comparing intrinsic conduction between the RV and LV IEGMs and conduction time between the RV and LV during RV and LV pacing
AdaptiveCRT (Medtronic; Dublin, Ireland) AV and VV optimization	IEGM-based method that dynamically calculates AV delay every minute. LV-only pacing is delivered for native AV interval < 220 ms and AV delay is time from RA sense or RA pace to RV sense – 40 ms. If intrinsic AV interval > 220 ms, then biventricular pacing is delivered after the end of the atrial IEGM and > 50 ms before RV sense. VV interval is on the basis of AV interval and time between RV sense and end of the ventricular IEGM on the far field signal
SyncAV (Abbott) AV optimization only	IEGM-based method that calculates and dynamically sets AV delay by assessing intrinsic AV delay every 256 beats and subtracting a programmed offset (50 ms nominally, but can be set to 10-120 ms)
SonR (Microport; Shanghai, China) AV and VV optimization	Using a lead-based microaccelerometer to detect mechanical vibrations (endocardial acceleration signal), AV and VV delays are dynamically optimized weekly during rest and exercise to maximize the peak endocardial acceleration signal, which is a surrogate for LV contractility
CRT AutoAdapt (Biotronik; Berlin, Germany) AV and VV optimization	IEGM-based method like AdaptiveCRT. AV interval to the RV and LV is measured on the basis of sensed and paced atrial beats. LV-only pacing is delivered if A-paced AV interval is < 250 ms and A to LV interval is longer than A to RV interval, otherwise biventricular pacing is delivered. AV delay is dynamically set at 70% of AV interval or AV interval – 40 ms, depending on which is shorter

AV, atrioventricular; CRT, cardiac resynchronization therapy; IEGM, intrinsic electrogram; LV, left ventricle; RA, right atrial; RV, right ventricle; VV, ventriculo-ventricular.

shown improved QRS narrowing and improved ventricular function with dynamic biventricular fusion pacing, however these findings need to be validated in a large, randomized trial (eg, NCT04100148).³³⁴

10.4. CSP programming

There is a paucity of data regarding optimal programming for CSP. In general, excessive AV delays (> 200 ms) should be avoided, because they might result in reduced cardiac output (due to fusion of the passive and active filling components, reduced filling time, suboptimal preload), diastolic mitral regurgitation (valvular regurgitation due to the reopening of the valves at the end of the ventricular diastole), and an increased risk of AF. In general, the programmed AV delays should be shorter than conventional parameters by 30-50 ms for HBP and 20-30 ms for LBBAP, especially for underlying LBBB.³³⁵ Unipolar ventricular output pacing is generally recommended.

Recommendation

93. We recommend that transvenous lead extraction (TLE) be performed by experienced operators, use a heart team approach, standard operating procedures, and immediate cardiac surgical back-up (Strong Recommendation; Low-Quality Evidence).
94. We recommend a comprehensive preprocedural risk assessment before lead extraction, including the use of preprocedural imaging (eg, echocardiography and/or CT scanning) and validated risk stratification tools (Strong Recommendation; Low-Quality Evidence).
95. We recommend that the comprehensive investigation in patients with suspected CIED-related infection should include blood cultures (before the start of antibiotics), pre-extraction transesophageal echocardiography (to identify vegetations on valves or leads), and lead, tissue, and pocket cultures at the time of extraction (Strong Recommendation; Low-Quality Evidence).

11. Lead Extraction

The use of CIEDs has grown over time, with systems increasing in complexity (eg, moving from fixed-rate single-chamber single-lead pacemakers to multichamber, multilead, rate-responsive devices that are capable of cardioversion, defibrillation, and/or CRT. In concert with increases in life expectancy is an increasing need for device system revision (eg, from pacemaker to ICD or CRT system), lead revision procedures, and generator replacements, as well as the need for safe and effective methods for removing CIED systems. The objective of TLE is the removal of the CIED lead from surrounding fibrous adhesions while minimizing damage to cardiovascular structures. Lead extraction refers to the removal of leads older than 1 year or those that require the assistance of specialized equipment (locking stylets, powered or mechanical sheaths, snaring, etc), or removal from a route other than the implant vein (eg, femoral extraction). Lead explant refers to the removal of a lead by simple traction only and when the dwell time is < 1 year. Surgical extraction is usually reserved for failure of the transvenous approach, very large vegetations (eg, > 3 cm), and anomalous placement of leads. Standard surgical techniques using a median sternotomy, thoracotomy, or limited atriotomy remains a definitive solution, but at the expense of significant morbidity, prolonged recovery time, and elevated mortality rate (0-12.5%).

TLE is associated with a procedural success of 95%-97% (complete removal of all hardware) to 98%-99% (if partial extractions [residual lead < 4 cm] are included).³³⁶⁻³³⁸ However, TLE carries a degree of significant risk with single- or multicentre series that have reported an incidence of major life-threatening complications in 0.4%-3.5% of the patients (0-0.8% incidence of death).^{336,337,339-342} Major intraprocedural complications include myocardial avulsion, cardiac tamponade, vascular tear, pneumothorax, hemothorax, arteriovenous

fistula, pulmonary embolism, and death. Minor complications, such as pericardial effusion, hematoma, venous thrombosis, and arrhythmia are not life-threatening but often require intervention. In addition, the periextraction period is associated with significant morbidity and mortality independent of the extraction procedure itself, usually because of ongoing septicemia, as well as the presence of multiple comorbidities.

11.1. Clinical evaluation

The purpose of the clinical evaluation is to confirm the indication for extraction and estimate the risk of risk and benefit of TLE. Data from the US Extraction Database indicate the most important factors to predict failed extraction are increased implant duration (with the risk of failed extraction doubling for every 3 years from time of initial implant), physician inexperience (lower success rates for operators with < 20 procedures), younger patient age, ventricular and passive fixation leads, and a greater number of leads. Complication rates are observed to be higher in these groups, as well as in female individuals with low body weight (body mass index < 25), frail individuals, and those with venous occlusion.³³⁶⁻³⁴⁵

11.2. Indications for lead removal

11.2.1. Infection-related indications.

Recommendation

96. We recommend lead extraction and complete CIED system removal for definite CIED-related infections (pocket, lead-related, or systemic) (Strong Recommendation; High-Quality Evidence).
97. We recommend lead extraction and complete CIED system removal for patients with persistent or recurrent bacteremia or fungemia (especially gram-positive organisms) despite appropriate antibiotic therapy and no other identifiable source (Strong Recommendation; High-Quality Evidence).
98. We recommend lead extraction and complete CIED system removal for patients with valvular endocarditis in whom the involvement of the leads in an active infection is not definite (Strong Recommendation; High-Quality Evidence).

Practical tips

- Early extraction (< 7 days) should be performed for definite CIED-related infections, even earlier in the case of *S aureus* septicemia.
- Consultation with infectious disease specialists is recommended to adjust the antibiotic regimen, duration, and timing of reimplantation, and identification of any source of infection or dissemination.

Despite an overall decrease in the rate of CIED-related complications, the rate of infection has increased out of proportion to the overall increases in device utilization (124%-210% increase in the annual incidence of CIED infection). CIED infection is associated with high patient

morbidity and mortality (7%-24%). CIED infection increased the risk of in-hospital death by more than twofold.²⁵⁵

11.2.1.1. Definite CIED-related infections. Complete system removal is strongly recommended in patients with definite CIED-related infections (pocket infection, pocket erosion, and lead-related endocarditis), ideally as soon as feasible because morbidity and mortality significantly increase with any delay in the extraction procedure.^{346,347} In patients in whom large vegetations are seen (> 2 cm), the use of angioaspiration devices to remove these vegetations during lead extraction might avoid the need for open heart surgery.³⁴⁸ This is particularly important for globular vegetations, which are associated with increased procedural risk.³⁴⁹ After hardware removal a complete course of antibiotics is recommended, with a minimum duration of 10-14 days for pocket infections and 4-6 weeks for lead-related endocarditis.²⁵⁵

In rare instances when life expectancy is short, or the risk of extraction is prohibitive, a palliative long-term antibiotic suppression strategy could be considered in consultation with infectious disease specialists.

11.2.1.2. Suspected CIED-related infections. In patients with persistent or recurrent bacteremia or fungemia (especially gram-positive organisms) despite appropriate antibiotic therapy and no other identifiable source, lead extraction is recommended for potential source control.

For patients in whom the diagnosis of lead endocarditis is suspected but not confirmed, advanced imaging such as positron emission tomography-CT or radiolabelled leucocyte

scintigraphy can be used to confirm the presence of lead and/or valve endocarditis.³⁵⁰ Nuclear imaging can also be used to identify a source of infection to allow for diagnosis and possible early referral for lead and system removal.

11.2.1.3. Concomitant infections. Lead extraction is also recommended in patients with valvular endocarditis in whom the involvement of the leads in an active infection is not definite. This is undertaken to prevent infection recurrence and has been associated with better survival (adjusted OR, 0.51; 95% CI, 0.40-0.66 in propensity score-matching).³⁵¹

In patients with bacteremia in the setting of prosthetic valves, CIED removal might serve as a lower-risk treatment option before considering valve replacement, particularly in patients with gram-positive infections.

11.2.2. Lead-related indications.

Recommendation

99. We recommend lead extraction for patients with superior vena cava (SVC) stenosis or occlusion that is symptomatic or prevents implantation of a new CIED lead (Strong Recommendation; Low-Quality Evidence).
100. We suggest lead extraction be considered as an alternative to abandonment of nonfunctional leads or leads that are no longer required, particularly in younger patients (Strong Recommendation; Low-Quality Evidence).

Table 17. Training standards

Operator	Establishment of competence (annual; primary)	Maintenance of competence (annual; primary)
Pacemaker implant	≥ 75 New pacemaker implants and ≥ 25 Pacemaker system revisions and/or generator replacements and Follow-up management and device programming of ≥ 150 patients	≥ 35 Pacemaker implant procedures per year and Follow-up management and device programming of ≥ 50 pacemaker patients
ICD implant	≥ 75 New CIED implants (≥ 50% ICDs) and ≥ 25 CIED system revisions and/or generator replacements (≥ 15 ICDs) and Follow-up management and device programming of ≥ 150 patients (≥ 50% ICDs)	≥ 35 New CIED implant procedures per year (≥ 50% ICDs) and Follow-up management and device programming of ≥ 50 ICD or CRT patients
CRT implant	≥ 75 New CIED implants and ≥ 25 New coronary sinus left ventricular lead implantations and ≥ 25 CIED system revisions and/or generator replacements (≥ 5 CRTs) and Follow-up management and device programming of ≥ 150 patients (≥ 50% complex devices)	≥ 35 new CIED implant procedures per year (≥ 33% CRTs) and Follow-up management and device programming of ≥ 50 CRT patients
Lead extraction	Extract ≥ 40 leads as the primary operator with supervision of an experienced faculty member	Extract ≥ 20 leads per year as the primary operator
Nonimplanting device follow-up physician	Follow-up management, interrogation, and programming of ≥ 100 pacemaker, ≥ 100 ICD, and ≥ 50 CRT devices over a minimum device training period of 6 months	Follow-up management and device programming of ≥ 50 patients

CIED, cardiac implantable electronic device; CRT, cardiac resynchronization therapy; ICD, implantable cardioverter defibrillator.

Practical tip. The shared decision process should weigh the relative risks and benefits of extraction vs abandonment, with consideration of patient comorbidities and totality of implanted hardware.

Abandonment or dysfunction of CIED system leads represents the second most common indication for extraction. In most cases leads are extracted in order to reestablish venous access in the event of target vein stenosis or occlusion, which is particularly relevant at the time of device system revision (eg, from pacemaker to ICD or CRT). Alternatively, in the absence of vascular access compromise nonfunctional leads might be prophylactically extracted at the time of planned lead procedure.

11.2.1.1. Device revision in the setting of SVC or ipsilateral vein occlusion. In the context of patients who require a device revision or additional lead placement, 25%-35% of patients will have a significant venous obstruction, including 12% with complete occlusion (ipsilateral vein or SVC).^{352,353} Options include lead extraction with placement of new lead(s), contralateral lead placement with tunneling to the pocket, venoplasty, or consideration of a leadless device. The decision to pursue extraction should consider the risks and benefits (see Section 11.1). Shared decision-making with the patient given the specific risks and benefits of each approach should be undertaken.

11.2.2.2. SVC stenting or transcatheter tricuspid valve replacement. In patients who undergo either SVC stenting or transcatheter tricuspid valve replacement, removal of all leads pre-intervention is usually recommended to avoid any lead entrapment. In patients who require pacing or tachyarrhythmia therapy, alternative options such as leadless pacemakers, LV-only pacing, or nontransvenous ICD systems should be considered. A heart team approach is important in these complex situations.³⁵⁴

Recommendation

101. We recommend that a multidisciplinary approach involving the lead management team is needed to determine if lead extraction is required before tricuspid valve interventions (Weak Recommendation; Low-Quality Evidence).

11.2.2.3. Removal of abandoned or malfunctioning leads. When leads are no longer required (eg, lead failure or revision from a pacemaker to an ICD), a decision needs to be made as to whether the lead should be left in place (abandoned) or removed. The decision to abandon or extract a lead should be individualized, taking into consideration the risks and benefits of each strategy. In general, lead extraction is favoured in young patients in whom the effects of adding leads over time is significant. Procedural risk should be quantified and discussed with the patient on the basis of available risk stratification tools and the presence of high-risk features. Ideally, the decision should be made before lead abandonment because a lead will never be easier to extract later. Although prophylactic lead extraction does expose the patient to an immediate risk, it

might prevent higher-risk extraction later in life if required (eg, infection).³⁵⁵ Shared decision-making with patients should be planned in advance to secure the ideal procedure.

11.3. Standards

11.3.1. Team approach. A multidisciplinary heart team approach involving electrophysiologists, cardiac surgeons, infectious disease specialists, anaesthesiologists, and imaging specialists is recommended with preoperative assessment of the risks and the benefits using validated risk prediction algorithms.^{342,344,345} Preprocedural imaging can be helpful in risk stratification and procedural planning. CT scans can help in assessment of lead adhesions, the degree of fibrosis around the leads, extracardiac and/or extravascular lead course, and vascular patency, as well as determine the presence of septic emboli in definite cases of infection.³⁵⁶ Transesophageal echocardiography imaging is important to identify lead-related echogenicity and assess their size and location (eg, thrombi and/or vegetations), with potential planning for the use of angioaspiration devices. In high-risk patients or those with known coronary artery disease, coronary angiography can help identify significant stenosis in preparation for possible emergency sternotomy.

11.3.2. Procedural setup. A lead extraction program should include physicians trained for this complex procedure and a safe operative environment that includes the following.

1. General anaesthesia with a preference for those with dedicated cardiac anaesthesia training.
2. Immediate access to cardiac surgery and extracorporeal circulation.
3. Preparation for possible open-heart surgery (sternotomy or thoracotomy).
4. Availability of procedural echocardiography (transesophageal or intracardiac).
5. Readily available blood products.
6. Femoral access for resuscitation and the use of the bridge balloon in case of SVC tear.³⁵⁷
7. Expertise in snaring techniques (for leads and/or lead fragments) and the use of tools for aspiration of vegetations and/or thrombi.
8. Ideally, these complex procedures are well suited for a hybrid suite or an alternative setting that allows for immediate surgical back-up.

12. Competency and Training Standards

Competency in CIED implantation and management is critical for improving patient outcomes.³⁵⁸ Because patient outcomes are closely tied to operator and centre experience, it is recommended that the establishment and maintenance of competence meet the following, recognizing that numerical volumes do not necessarily reflect the quality of care provided.

12.1. Device implantation and maintenance of competence

CIED implantation involves a spectrum of care that begins with the clinical assessment and determination of the

Table 18. Suggested Quality Indicators

	Indicator
Structural	- Each province should establish a provincial registry of device implant outcomes - Centres in which CIEDs are implanted should monitor and report annually the following QIs
Volumes	- The volume of procedures performed by the centre (stratified according to type) - The volume of procedures performed by individual operators (stratified according to type)
Follow-up	Time to first visit in a device follow-up clinic
Access	Median wait time from referral to device implantation (completed cases, stratified according to device)
Implant	- Proportion of patients considered for CIED implantation who receive prophylactic antibiotics 1 hour before their procedure - Proportion of patients considered for CIED implantation who underwent imaging assessment of left ventricle structure and function (echocardiography, nuclear, magnetic resonance imaging) within 6 months before the implantation procedure - Proportion of patients with HFrEF (LVEF $\leq$ 35%) who receive an ICD - Proportion of patients with HFrEF (LVEF $\leq$ 35%) who undergo CRT
Complication	- Annual rate of procedural complications (CIED-related bleeding, pneumothorax, cardiac perforation, tamponade, pocket hematoma, lead displacement, or infection) within 30 days after CIED implantation, replacement, or revision - Annual rates of CIED-related infections up to 1 year after CIED implantation, replacement, or revision
Reoperation	- Annual rates of CIED reoperation within 30 days after CIED implantation, replacement, or revision - Annual rates of CIED reoperation within 1 year after CIED implantation, replacement, or revision
Mortality	- Annual rates of mortality within 30 days after CIED implantation, replacement, or revision - Annual rates of mortality within 1 year after CIED implantation, replacement, or revision
Readmission	Annual rates of emergency department consultation or hospital readmission within 30 days after CIED implantation, replacement, or revision

CIED, cardiac implantable electronic device; CRT, cardiac resynchronization therapy; HFrEF, heart failure with reduced ejection fraction; ICD, implantable cardioverter defibrillator; LVEF, left ventricular ejection fraction; QI, quality indicator.

appropriate indications for CIED implantation, includes the procedural aspect of device implantation, and concludes with the ongoing outpatient management of patients living with a CIED. As such, training for physicians who perform CIED implantation must be comprehensive, and include the development of expertise in patient assessment, arrhythmia investigation diagnosis, pathophysiology of cardiovascular disease, device selection tailored to the individual patient's unique needs, the procedural aspect of device implantation including pocket management, vascular access, and lead manipulation and positioning, threshold testing, and electrophysiological interpretation during implantation,

Table 19. Recommended maximal wait-time target for CIED implantation

Prioritization	Procedure
Emergent ($\leq$ 72 hours, hospitalized) or urgent ($<$ 1 week, outpatient)	- Pacemaker implantation for: - Dependent patient - High-grade AV block - Bradyarrhythmic syncope (asystole or AV block) - ICD implantation for secondary prevention - Generator changes for CIED at end of life - Lead revision because of lead fracture and/or dislodgement leading to: - Proarrhythmia - Hemodynamic compromise - Inappropriate shock - Lead extraction in a patient with CIED-associated sepsis
Semiurgent (1 month)	- Pacemaker implantation before AV node ablation - ICD implantation for primary prevention (high risk) - Cardiac resynchronization for a patient with recent, or at high risk for ADHF - Generator changes for CIED at ERI - ICM implantation in patients with recurrent or malignant syncope - Lead revision because of lead fracture and/or dislodgement leading to symptoms but low risk of arrhythmia, hemodynamic compromise, inappropriate shock, and/or hospital admission
Elective (90 days)	- Pacemaker implantation for sinus node dysfunction - ICD implantation for primary prevention - ICM implantation for arrhythmia monitoring or unexplained symptoms - CRT-pacemaker for pacing-induced cardiomyopathy - Left ventricular lead revision - Generator changes of CIED approaching but not yet at ERI

ADHF, acute decompensated heart failure; AV, atrioventricular; CIED, cardiac implantable electronic device; CRT, cardiac resynchronization therapy; ERI, elective replacement indicator; ICD, implantable cardioverter defibrillator; ICM, implantable cardiac monitor.

postimplantation device programming and pacing algorithms, and management of CIED implant-related complications. Although extensive, this level of knowledge and skill is required to deliver comprehensive care to patients with CIEDs.

Before independently implanting a CIED a physician must undertake a dedicated training period of at least 12 months in duration, in which they meet the minimum annual volume for implantation and in-person follow-up (Table 17). To maintain competence in CIED implantation and follow-up, a minimum number of device implants and follow-up procedures per year are required. In addition, maintenance of competence for implant physicians requires participation in continuing professional development activities.

12.2. Device implant and follow-up centres

Because of the complexity of CIED implantation and management it is recommended that CIED care be provided in centres with adequate resources and expertise. Centres in which device implantation procedures are performed must

have access to advanced cardiac services (eg, invasive and noninvasive cardiac imaging), human resources (a minimum 2 active implanting operators per centre per device type, anaesthetic support, and appropriately trained nurses and technologists), and a sterile implantation environment capable of delivering full anaesthetic support and resuscitation. Because these services cannot be provided at each centre it makes sense to regionalize advanced cardiac services on the basis of a balance of resource availability and anticipated complexity. In complex device implant centres the full range of CIEDs are typically implanted, including ICMs, single- and dual-chamber pacemakers, ICDs, CRT and CSP devices, and leadless or nontransvenous devices. In some complex device centres lead extraction might be performed. In these centres at least 200 new implants per year are performed as well as follow-up care for at least 500 patients across the spectrum of CIED types. In non-complex device implant centres ICMs and pacemakers are typically implanted. In these centres at least 50 new pacemaker implants per year are performed as well as follow-up care for at least 200 pacemaker patients. Simple device implant centres are typically affiliated with complex device implant centres to enable referral for complex or technically challenging device implant procedures. In CIED follow-up clinics, devices are not implanted, but a minimum caseload of 200 follow-up patients per year must be maintained. CIED follow-up clinics are typically affiliated with simple and complex device implant centres to enable referral for device implant procedures.

13. Sex Differences

Sex differences are observed in the pathophysiology, presentation, and outcomes of several cardiac disorders. It is known that female patients tend to be differentially treated for various biological and sociocultural reasons. Recognition of sex differences offers an opportunity to affect outcomes in female patients who require CIED implantation.

13.1. Device indications

Across nearly all indications, lower rates of CIED implantation are observed in female patients (pacemaker [adjusted OR, 0.78; 95% CI, 0.76-0.80], defibrillator [adjusted OR, 0.4; 95% CI, 0.40-0.45], or resynchronization device [adjusted OR, 0.66; 95% CI, 0.61-0.71]).^{359,360} Female patients who receive a pacemaker are older at the time of pacemaker implantation, and are more likely to receive a device for SND or AF with slow ventricular response.^{361,362}

13.2. Complications

Implant complications has been observed to be higher in women, which is predominantly due to higher rates of pneumothorax, pericardial effusion, and pocket hematomas.³⁶³⁻³⁶⁵ Higher complication rates have been noted for transvenous and leadless devices. It is postulated that these are related to smaller body habitus, smaller venous diameter, and smaller right ventricle chamber size.

13.3. Response to therapy

Women have been well represented in randomized trials of bradycardia pacing. In these trials and registries female

patients experienced event rates similar to male patients; female patients experienced relative improvement in quality of life similar to male patients, however, their quality of life was lower in absolute terms.^{366,367}

In contrast, female patients have been under-represented in trials that have evaluated ICDs or CRT. In the case of ICDs, female patients have a lower incidence of SCD, ventricular arrhythmias, or appropriate device therapies.³⁶⁸ The reason for the lower incidence of sudden death and ventricular arrhythmia are unknown. Despite lower absolute event rates, female patients derive the same relative benefit from ICD therapy as male patients. Conversely, because of a lower incidence of coronary disease and/or higher relative prevalence of nonischemic cardiomyopathy, female patients experience a lower mortality after CRT implantation.^{369,370}

13.4. Implant considerations

Concern regarding physical appearance might affect choice of implant technique and type of device more frequently in female patients.³⁷¹ Implanting the device under the pectoral muscle to render it less visible and using an axillary incision line to help camouflage the wound can be considered.³⁷² In appropriate cases, leadless pacemakers and nontransvenous defibrillators might be preferable to transvenous systems, depending on patient medical conditions and indications.

13.5. Other

13.5.1. Pregnancy. Case series on pregnant women with CIEDs, including defibrillators, have recommended management as per standard of care.^{373,374} In some cases transient AVB might occur during pregnancy. If a *de novo* implant is required, it can be performed under echocardiographic guidance and electro-anatomic navigation to avoid fluoroscopy.³⁷⁵ However, after the first trimester, the risk associated with low-dose fluoroscopy is lower and fluoroscopy with protection might be considered. For those with a CIED for bradycardia indications, tracking rates or sensor functions might need to be increased to allow for physiological adjustment in pregnancy.

13.5.2. Breast cancer. Radiotherapy for breast cancer might increase the risk of requiring a pacemaker, although results have been inconsistent.³⁷⁶ CIED implantation is generally contraindicated on the ipsilateral site of mastectomy with lymph node resection because of the potential for venous and lymphatic obstruction. In case of active cancer, implantation should be performed on the contralateral side, or leadless pacemakers might be considered. With a preexisting device, radiotherapy can pose a risk of malfunction, although various protocols exist to minimize the risk.

14. Quality Indicators

Quality indicators are standardized, evidence-based measures designed to evaluate the quality of care, to measure and track clinical performance and outcomes, to quantify variation, identify areas in need, and enable improvements in health care delivery. The quality indicators listed in Table 18 are derived from high-quality guidance and are intended to

assist health care providers to operationalize these guideline recommendations. The intent of the quality indicators is to develop and standardize the tools by which the quality of care for cardiac pacing is evaluated and resultant outcomes monitored and reported, enabling the discrimination between missed opportunities and appropriate care.

14.1. Access to care

Timely access to CIED therapy is important for ensuring health outcomes are optimized (Table 19).

Conclusion

This report presents the first comprehensive CCS/CHRS guidelines to address the management of patients with CIEDs. We have provided extensive recommendations covering all aspects of CIED management (for convenience, a complete summary of all recommendations is provided in Table 20). We hope that this document will become a useful reference tool for medical personnel at all levels who are involved in the management of patients with CIEDs.

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Ethics Statement

All research reported in the current article adheres to the relevant ethical guidelines.

Patient Consent

The authors confirm that patient consent is not applicable to this article. This is a guideline document without use of any patient-level data.

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Supplementary Material

To access the supplementary material accompanying this article, visit the online version of the *Canadian Journal of Cardiology* at www.onlinecjc.ca and at <https://doi.org/10.1016/j.cjca.2025.08.363>.

Table 20. Summary of recommendations

Number	Section	Recommendation	Type	Strength of recommendation	Level of evidence
1	2.2.	We recommend ambulatory ECG monitoring for symptom-rhythm correlation in patients with suspected bradycardia or conduction disorder in whom a comprehensive evaluation did not indicate a cause	New	Strong	High
2	2.2.	We suggest screening for sleep apnea for those patients with sleep-related bradycardia or conduction disorder	New	Weak	Moderate
3	2.3.	We recommend PPM implantation for patients with symptomatic sinus node dysfunction not caused by reversible or physiologic causes	New	Strong	High
4	2.4.	We recommend PPM implantation for patients with acquired third-degree AVB, type II second-degree AVB, high-grade AVB, or alternating bundle branch block not caused by reversible or physiologic causes, regardless of symptoms	New	Strong	Low
5	2.4.	We recommend PPM implantation for patients with significant pauses (≥ 6 seconds) during AF or sustained slow ventricular response (heart rate < 40 bpm) during AF while awake	New	Strong	Low
6	2.5.1.	We recommend PPM implantation for patients with third-degree AVB, type II second-degree AVB, high-grade AVB, alternating bundle branch block, or symptomatic sinus node dysfunction that persists for > 5 days after myocardial infarction	New	Strong	Low
7	2.5.2.	We recommend PPM implantation for patients with third-degree AVB, type II second-degree AVB, high-grade AVB, alternating bundle branch block, or symptomatic sinus node dysfunction that persists for > 5 days after cardiothoracic surgery for valvular disease	New	Strong	Low
8	2.5.3.	We recommend PPM implantation for patients with persistent third-degree AVB, type II second-degree AVB, high-grade AVB, or alternating bundle branch block after TAVR	New	Strong	Moderate
9	2.5.3.	We suggest that patients with a new persistent LBBB (QRS > 150 ms) and first-degree AV block (PR interval > 240 ms) for 48 hours after TAVR undergo PPM implantation	New	Weak	Moderate
10	2.5.3.	We suggest that patients with either new LBBB (QRS > 150 ms) or new first-degree AVB (PR interval > 240 ms) after TAVR undergo PPM implantation, selective electrophysiologic testing for risk stratification and guidance of PPM implantation, or prolonged postdischarge ambulatory ECG monitoring	New	Weak	Moderate
11	2.5.3.	We suggest that patients with new-onset conduction disturbance (> 20 ms increase in the PR interval to 200-240 ms or new-onset LBBB with QRS duration < 150 ms) after TAVR or preexisting right bundle branch block before TAVR undergo prolonged inpatient telemetry (48 hours) with prolonged postdischarge ambulatory ECG monitoring	New	Weak	Low
12	2.5.4.	We recommend pacemaker implantation be considered in patients with complex CHD and standard pacemaker indications, after an interdisciplinary discussion involving specialists with expertise in the management of patients with adult CHD	New	Strong	Moderate
13	2.5.5.	We recommend pacemaker implantation in patients with congenital AV block and symptoms of bradycardia, or asymptomatic patients with evidence of ≥ 1 of ventricular dilatation or dysfunction, mean daytime heart rate < 50 bpm, wide QRS escape rhythm, junctional exit block (pauses 2-3 times the basic cycle length), QT prolongation, or complex or frequent ventricular ectopy	New	Strong	Low
14	2.5.6.	We recommend permanent pacing in patients with cardiac sarcoidosis and third-degree AVB, type II second-degree AVB, and/or high-grade AVB with an ICD in preference to standard pacemaker, even in the presence of a preserved LVEF	New	Strong	Low
15	2.5.7.	We recommend that the assessment for pacemaker candidacy in patients with neuromuscular disease be performed by specialists with expertise in the management of these patients	New	Strong	Low
16	2.5.7.	We recommend pacemaker implantation for patients with Duchenne muscular dystrophy, Becker muscular dystrophy, and recessive limb-girdle muscular dystrophies (type II) with documented symptomatic sinus node dysfunction, or acquired third-degree AVB, type II second-degree AVB, high-grade AVB, or alternating bundle branch block not caused by reversible or physiologic causes, regardless of symptoms	New	Strong	Low
17	2.5.7.	We recommend pacemaker implantation for patients with myotonic dystrophy, Kearns-Sayre syndrome, Emery-Dreifuss, or dominant limb-girdle muscular dystrophy type 1B with any second- or third-degree AVB, PR duration ≥ 240 ms, or QRS duration ≥ 120 ms, regardless of symptoms	New	Strong	Low
18	2.5.8.	We suggest CSP may be considered as an alternative to biventricular pacing in those with an LVEF of 36%-50% or as an alternative to right ventricular pacing in those with LVEF $> 50\%$ who undergo catheter ablation of the AV node	New	Weak	Low
19	2.6.	We recommend dual-chamber pacemaker implantation in preference to single chamber atrial or ventricular pacing for patients with sinus node dysfunction who undergo device implantation	New	Strong	High

Table 20. Continued.

Number	Section	Recommendation	Type	Strength of recommendation	Level of evidence
20	2.6.	We recommend dual-chamber pacemaker implantation in preference to single ventricular pacing for those patients in sinus rhythm with atrioventricular block undergoing device implant.	New	Strong	High
21	2.6.	We recommend leadless pacemaker implantation be considered as an alternative to transvenous or epicardial pacemakers in patients with known difficulties in vascular access or at high risk of device-system infection	New	Strong	Low
22	3.0.	We suggest that an implantable cardiac monitor or tilt-table test are reasonable management choices for patients with unexplained syncope after initial assessment, no apparent conduction disease, and preserved ejection fraction	New	Weak	Low
23	3.1.1.	We recommend that dual-chamber pacemaker implantation may be considered in patients older than 40 years of age with unpredictable recurrent refractory neurocardiogenic syncope in the presence of symptomatic asystole > 3 seconds, asymptomatic asystole > 6 seconds secondary to sinus arrest or AV block, or asystole > 3 seconds associated with syncope on HUTT	New	Strong	High
24	3.1.2.	We suggest a strategy of empiric pacemaker implantation may be considered in patients with unexplained syncope and bifascicular block	New	Weak	Moderate
25	4.1.	We recommend that every patient with HFrEF should be assessed for suitability for CRT after optimization of medical therapy to target or maximally tolerated doses, with a repeat assessment of LVEF before referral	Brought forward Adapted 2013	Strong	Low
26	4.1.	Routine assessment of dyssynchrony with present echocardiography techniques is not recommended to guide the prescription of CRT	Brought forward 2013	Strong	Low
27	4.2.1.	We recommend CRT for patients in sinus rhythm with NYHA class II-IVa heart failure despite optimal medical therapy, LVEF ≤ 35%, and QRS duration ≥ 130 ms with left bundle branch block morphology	Brought forward Adapted 2013	Strong	High
28	4.2.1.	We suggest CRT may be considered for patients in sinus rhythm, with NYHA class II-IVa heart failure despite optimal medical therapy, LVEF ≤ 35%, and QRS duration ≥ 150 ms with non-left bundle branch block morphology	Brought forward Adapted 2017	Weak	Low
29	4.2.1.	We recommend CRT not be used for patients with QRS < 130 ms in the absence of an indication for chronic RV pacing, irrespective of heart failure symptoms or LVEF or the presence or absence of mechanical dyssynchrony shown using current imaging techniques	Brought forward Adapted 2017	Strong	Moderate
30	4.2.2.	We suggest CRT may be considered for patients in permanent AF who are otherwise candidates for CRT	Brought forward Adapted 2013	Weak	Low
31	4.2.2.	We suggest atrioventricular node ablation in patients with AF, a history of HFrEF, and who are CRT nonresponders with biventricular pacing < 95% despite maximally tolerated doses of rate-controlling drugs	Brought forward 2020	Weak	Moderate
32	4.2.3.	We suggest that CRT might be considered for patients with new-onset high-degree AV block requiring chronic RV pacing, signs and/or symptoms of heart failure, and LVEF ≤ 45%	Brought forward 2013	Weak	Low
33	4.2.4.	We suggest system revision to biventricular pacing CRT for patients with an existing pacemaker or ICD with reduced LVEF ≤ 45%, frequent right ventricular pacing, and NYHA class II-IVa heart failure	Brought forward Adapted 2013	Weak	Low
34	4.2.5.	We recommend CRT-defibrillator implantation for patients who have an indication for a CRT and an ICD	Brought forward Adapted 2017	Strong	High
35	4.2.5.	We recommend CRT-pacemaker implantation for patients who have an indication for CRT, but not for an ICD	Brought forward Adapted 2017	Strong	Moderate
36	4.2.5.	We recommend that the choice of CRT platform (CRT-pacemaker vs CRT-defibrillator) should take into account clinical factors that would affect the overall goals of care	Brought forward Adapted 2013	Strong	Moderate
37	4.2.6.	We suggest pacing from a nonapical left ventricular epicardial region in patients treated with CRT	Brought forward Adapted 2013	Weak	Low
38	4.2.6.	We suggest that placement of a left ventricular lead at the time of open-heart surgery, for the purpose of facilitating future CRT, might be considered in patients for whom CRT is recommended and the need for device therapy is unlikely to be changed by the surgical procedure	Brought forward 2013	Weak	Low
39	5.2.	We suggest CSP may be considered in those with an LVEF of 36%-50% or as an alternative to right ventricular pacing in those with LVEF > 50%	New	Weak	Low
40	5.2.	We recommend CSP be performed in patients who undergo traditional CRT implantation when an adequate coronary sinus lead position cannot be obtained because of suboptimal coronary sinus anatomy (poor targets, lead instability, high capture thresholds, or extracardiac [phrenic] capture)	New	Strong	Low
41	5.2.	We recommend CSP be considered as an adjunct in CRT nonresponders with prolonged paced QRS duration despite CRT optimization	New	Strong	Low
42	6.1.	We recommend that every patient with HFrEF should be assessed for suitability for ICD after optimization of medical therapy to target or maximally tolerated doses	Brought forward Adapted 2021	Strong	Moderate

Continued

Table 20. Continued.

Number	Section	Recommendation	Type	Strength of recommendation	Level of evidence
43	6.1.	We recommend that patients should undergo reassessment of LVEF to determine ICD candidacy at least 40 days after MI without revascularization, at least 3 months after MI with revascularization, or at least 3 months after optimization of medical therapy	Brought forward Adapted 2017	Strong	Low
44	6.1.	We recommend against ICD implantation in patients with NYHA class IV symptoms who are not expected to improve with any further therapy and who are not candidates for cardiac transplantation or mechanical circulatory support	Brought forward Adapted 2017	Strong	Moderate
45	6.1.	We recommend that an ICD not be offered to patients in whom comorbidities make it unlikely that an ICD will substantially increase a patient's life expectancy	Brought forward 2017	Strong	Moderate
46	6.2.1.	We recommend ICD implantation for patients after cardiac arrest due to ventricular arrhythmias in the absence of a reversible cause	Brought forward 2017	Strong	High
47	6.2.1.	We recommend ICD implantation for patients with sustained VT in the presence of significant structural heart disease and in the absence of a reversible cause	Brought forward Adapted 2017	Strong	High
48	6.2.1.	We suggest an ICD be considered in patients with significant structural heart disease and syncope of unknown origin for whom the cause of syncope is most likely VT and/or VF and in whom the risk of recurrent VT and/or VF is high	Brought forward 2017	Weak	Moderate
49	6.2.1.	We recommend an ICD for patients with syncope of unknown origin with inducible VT at the time of electrophysiological study and structural heart disease	Brought forward Adapted 2017	Strong	Moderate
50	6.2.1.	We suggest an ICD be considered after MI in the presence of sustained VT or VF > 48 hours after MI or > 48 hours after revascularization for MI in the absence of reversible causes	Brought forward Adapted 2017	Weak	Low
51	6.2.2.	We recommend consideration of primary ICD therapy in patients with ischemic cardiomyopathy, NYHA class I symptoms, and an ejection fraction $\leq 30\%$ at least 1 month after MI, and at least 3 months after coronary revascularization procedure	Brought forward 2017	Strong	High
52	6.2.2.	We recommend consideration of primary ICD therapy in patients with ischemic cardiomyopathy, NYHA class II-III symptoms, and ejection fraction $\leq 35\%$ measured at least 1 month after MI, and at least 3 months after coronary revascularization procedure	Brought forward 2017	Strong	High
53	6.2.2.	We recommend primary prevention ICD implantation in patients with nonischemic cardiomyopathy, NYHA class II-III symptoms, and an LVEF $\leq 35\%$ measured at least 3 months after optimization of guideline-directed medical therapy	Brought forward 2017	Strong	High
54	6.2.2.	We suggest that an ICD be considered in patients with an acute indication for cardiac pacing within 3 months after revascularization or 40 days of MI and before achieving optimal medical therapy when there is a high probability that the ejection fraction will remain $< 35\%$	Brought forward 2017	Weak	Low
55	6.3.1.	We recommend primary prevention ICD implantation in patients with cardiac sarcoidosis and an indication for PPM implantation	New	Weak	Low
56	6.3.2.1.	We suggest an estimate of risk for SCD in patients with hypertrophic cardiomyopathy should be determined on the basis of validated risk scores and/or the presence of ≥ 1 high-risk clinical factor to select appropriate candidates for primary prevention ICD therapy	Brought forward 2017	Weak	Moderate
57	6.3.2.2.	We recommend that patients with cardiac or neuromuscular laminopathies should undergo comprehensive evaluation and SCD risk stratification by specialists with expertise in the management of these individuals	New	Strong	Low
58	6.3.2.2.	We recommend primary prevention ICD insertion in patients with cardiac or neuromuscular laminopathies and an LVEF of $< 45\%$ and an elevated 5-year risk of life-threatening ventricular tachyarrhythmias (https://lmna-risk-vta.fr/), and/or the presence of ≥ 2 additional risk factors	New	Strong	Low
59	6.3.2.2.	We suggest primary prevention ICD insertion might be reasonable in patients with cardiac or neuromuscular laminopathies and an indication for a PPM	New	Weak	Moderate
60	6.3.2.3.	We recommend that patients with suspected or confirmed ARVC should undergo comprehensive evaluation and SCD risk stratification by specialists with expertise in the management of these individuals	New	Strong	Low
61	6.3.2.3.	We suggest primary prevention ICD implantation in patients with definite arrhythmogenic right ventricular cardiomyopathy related to pathogenic or likely pathogenic <i>PKP2</i> variants at high risk of SCD on the basis of the ARVC risk calculator	New	Weak	Low
62	6.3.2.4.	We recommend primary prevention ICD implantation in patients harboring the <i>TMEM43-S358L</i> variant upon manifestation of a positive clinical phenotype (most often increased ventricular ectopy on Holter monitoring) and in men 18 years of age or older regardless of phenotype	New	Weak	Moderate
63	6.3.3.	We recommend that patients with neuromuscular disease and associated cardiomyopathy undergo comprehensive evaluation and SCD risk stratification by specialists with expertise in the management of these individuals	New	Strong	Low
64	6.3.3.	We suggest primary prevention ICD implantation in patients with neuromuscular disease and LVEF $\leq 35\%$ despite optimal medical therapy	New	Weak	Low

Table 20. Continued.

Number	Section	Recommendation	Type	Strength of recommendation	Level of evidence
65	6.3.3.	We recommend primary prevention ICD insertion in patients with EDMD or limb-girdle muscular dystrophy 1B and an indication for pacing, or those with an LVEF < 45%, and an elevated risk of sudden death (see section 6.3.3.2.).	New	Strong	Low
66	6.3.3.	We suggest primary prevention ICD insertion in patients with myotonic dystrophy who have an indication for pacemaker implantation and presence of additional risk factors for VT (NSVT) or SCD (significant late gadolinium enhancement on cardiac MRI in patients of age 40 or older, increased number of CTG repeats).	New	Weak	Low
67	6.3.3.	We recommend against invasive electrophysiology study for risk stratification in patients with neuromuscular disease	New	Strong	Moderate
68	6.3.4.2.	We recommend primary prevention ICD implantation in patients with a spontaneous type I Brugada pattern and arrhythmic syncope	New	Strong	Low
69	6.3.4.3.	We suggest primary prevention ICD implantation be considered in patients with long QT type 2 (LQT2) with QTc values that remain > 500 ms (women) and 550 ms (men) despite optimal medical therapy	New	Weak	Low
70	6.3.4.3.	We suggest primary prevention ICD implantation be considered in long QT type 3 (LQT3) patients with QTc values that remain > 500 ms despite optimal medical therapy	New	Weak	Low
71	6.3.4.3.	We suggest ICD implantation for long QT syndrome (LQTS) patients with arrhythmic syncope despite optimal medical therapy, guided by their genetic subtype	New	Weak	Low
72	6.4.	We suggest a single-chamber ICD instead of a dual-chamber ICD in patients without a pacing indication who undergo ICD implantation	Brought forward Adapted 2017	Weak	Moderate
73	6.4.	We recommend a nontransvenous ICD be considered as an alternative to a single-chamber transvenous ICD in patients without a pacing indication who undergo ICD implantation	New	Strong	Moderate
74	7.0.	We suggest that ICDs be considered in patients with a left ventricular assist device	Brought forward Adapted 2017	Weak	Low
75	8.1.	We recommend the use of comprehensive CIED infection prevention measures in all patients who undergo device implantation	Brought forward Adapted 2017	Strong	Moderate
76	8.1.	We suggest the use of an antibacterial envelope be considered for CIED procedures when a high rate of infection is anticipated	New	Weak	Moderate
77	8.2.	We recommend that in patients taking warfarin for whom perioperative anticoagulation is deemed necessary, continued warfarin is recommended over the use of heparin-based bridging	Brought forward 2013	Strong	Moderate
78	9.2.	We recommend remote monitoring be used as part of the standard of care for all CIED patients	Brought forward Adapted 2013	Strong	Moderate
79	9.2.	We recommend that remote monitoring be used to supplement in-person monitoring of the patient and device	Brought forward Adapted 2013	Strong	Low
80	9.3.	We recommend that all patients with CIEDs who undergo MRI be managed according to a standardized protocol	Brought forward Adapted 2021	Strong	Moderate
81	9.3.	We recommend for patients with a previous CIED who undergo MRI, we recommend reviewing all available medical information, including chest radiography, to identify lead extenders, retained epicardial leads, abandoned transvenous leads, and/or fractured leads. We recommend that patients with lead extenders or fractured leads should not undergo MRI. However, MRI might be considered in patients with epicardial or abandoned transvenous leads when the clinical need is strong and believed to outweigh potential risks	Brought forward 2021	Weak	Low
82	9.3.	We recommend for patients with an MRI-conditional CIED who undergo MRI to perform scans at 1.5 T when possible, in a monitored setting and with access to the device team. The device team will provide prescan programming to MRI-compatible mode as well as postscan return to prescan settings	Brought forward 2021	Strong	Low
83	9.3.	We recommend for patients with subcutaneous ICDs scanning at 1.5 T in a monitored setting, supervised by a health care practitioner trained in ACLS and with access to the code and device teams. The device team will provide prescan programming to turn off tachycardia therapies, postscan interrogation, and reprogramming to prescan settings	Brought forward 2021	Strong	Low
84	9.3.	We recommend for patients with leadless pacemakers, scanning at an MRI field strength that does not exceed the manufacturer's specifications, in a monitored setting and with access to the device team. The device team will provide prescan programming and postscan reprogramming to prescan settings	Brought forward 2021	Strong	Low
85	9.3.	We recommend for patients with an MRI-nonconditional pacemaker and no pacemaker dependency who undergo MRI, performing scans at 1.5 T in a monitored setting and with access to the device team. The device team will provide prescan programming to sense mode (ie, OAO, OVO, or ODO) and post-scan interrogation and reprogramming to pre-scan settings	Brought forward 2021	Strong	High

Continued

Table 20. Continued.

Number	Section	Recommendation	Type	Strength of recommendation	Level of evidence
86	9.3.	We recommend for patients with an MRI-nonconditional pacemaker and pacemaker dependency who undergo MRI to perform scans at 1.5 T in a monitored setting, supervised by a health care practitioner trained in ACLS and with access to the code and device teams. The device team will provide prescan programming to asynchronous mode (ie, AOO, VOO, or DOO) and post-scan interrogation and reprogramming to pre-scan settings	Brought forward 2021	Strong	High
87	9.3.	We recommend for patients with MRI-nonconditional implantable transvenous defibrillators who undergo MRI, performing scans at 1.5 T in a monitored setting, supervised by a health care practitioner trained in ACLS and with access to the code and device teams. The device team will provide prescan programming to turn off tachycardia therapies and postscan interrogation and reprogramming to prescan settings	Brought forward 2021	Strong	High
88	9.4.	We recommend that patients with implantable cardiac devices <i>in situ</i> should be evaluated for programming changes before surgery and again after surgery, in accordance with existing Canadian Cardiovascular Society recommendations	Brought forward 2017	Strong	Low
89	9.6.	We recommend that all patients who are approaching elective replacement of CIED should undergo comprehensive re-evaluation by specialists with expertise in the management of CIED therapy	New	Strong	Moderate
90	9.6.	We recommend that all patients who are planned to receive a CIED system intervention should be considered for their eligibility for revision to ICD or CRT	Brought forward Adapted 2013	Strong	Low
91	9.6.	We recommend that an ICD be replaced by a pacing system, removed or abandoned at the time of ICD generator end of life and/or service in patients with life-threatening comorbidities, at the request of the patient or when an ICD is unlikely to increase life expectancy	Brought forward Adapted 2017	Strong	Low
92	10.	We recommend CIED programming be individualized and tailored to the patient's needs	New	Strong	High
93	11.	We recommend that transvenous lead extraction be performed by experienced operators, use a heart team approach, standard operating procedures, and immediate cardiac surgical back-up	New	Strong	Low
94	11.	We recommend a comprehensive preprocedural risk assessment before lead extraction, including the use of preprocedural imaging (eg, echocardiography and/or computed tomography scanning) and validated risk stratification tools	New	Strong	Low
95	11.	We recommend that the comprehensive investigation in patients with suspected CIED-related infection should include blood cultures (before the start of antibiotics), preextraction transesophageal echocardiography (to identify vegetations on valves or leads), and lead, and/or tissue, and/or pocket cultures at time of extraction	New	Strong	Low
96	11.2.1.	We recommend lead extraction and complete CIED system removal for definite CIED-related infections (pocket, lead-related, or systemic)	New	Strong	High
97	11.2.1.	We recommend lead extraction and complete CIED system removal for patients with persistent or recurrent bacteremia or fungemia (especially gram-positive organisms) despite appropriate antibiotic therapy and no other identifiable source	New	Strong	High
98	11.2.1.	We recommend lead extraction and complete CIED system removal for patients with valvular endocarditis where the involvement of the leads in an active infection is not definite	New	Strong	High
99	11.2.2.	We recommend lead extraction for patients with superior vena cava stenosis or occlusion that is symptomatic or prevents implantation of a new CIED lead	New	Strong	Low
100	11.2.2.	We suggest lead extraction be considered as an alternative to abandonment of nonfunctional leads or leads that are no longer required, particularly in younger patients	New	Strong	Low
101	11.2.2.2.	We recommend that a multidisciplinary approach involving the lead management team is needed to determine if lead extraction is required before tricuspid valve interventions	New	Weak	Low

ACLS, advanced cardiac life support; AF, atrial fibrillation; ARVC, arrhythmogenic right ventricular cardiomyopathy; AV, atrioventricular; AVB, atrioventricular block; bpm, beats per minute; CHD, congenital heart disease; CIED, cardiac implantable electronic device; CRT, cardiac resynchronization therapy; CSP, conduction system pacing; ECG, electrocardiogram; EDMD, Emery-Dreifuss muscular dystrophy; HFrEF, heart failure and reduced ejection fraction; HUTT, head-up tilt table; ICD, implantable cardioverter defibrillator; LBBB, left bundle branch block; LQTS, long QT syndrome; LVEF, left ventricular ejection fraction; MI, myocardial infarction; MRI, magnetic resonance imaging; NYHA, New York Heart Association; PPM, permanent pacemaker; QTc, corrected QT; RV, right ventricular; SCD, sudden cardiac death; TAVR, transcatheter aortic valve replacement; VF, ventricular fibrillation; VT, ventricular tachycardia.