Cardiology in the Young

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Original Article

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Abbreviations:
ACM, arrhythmogenic cardiomyopathy; ARVC, arrhythmogenic right ventricular cardiomyopathy; AV, atrioventricular; BrS, Brugada syndrome; CCAVB, congenital complete atrioventricular block; CHD, congenital heart disease; CIED, cardiovascular implantable electronic device; COR, class of recommendation; CPVT, catecholaminergic polymorphic ventricular tachycardia; ECG, electrocardiogram; HCM, hypertrophic cardiomyopathy; ICD, implantable cardioverter defibrillator; ICM, insertable cardiac monitor; IPE, in-person evaluation; LGE, late gadolinium enhancement; LOE, level of evidence; LQTS, long QT syndrome; LVEF, left ventricular ejection fraction; LVNC, left ventricular noncompaction; MRI, magnetic resonance imaging; NIDCM, nonischemic dilated cardiomyopathy; RIM, remote interrogation and monitoring; SND, sinus node dysfunction; SCA, sudden cardiac arrest; SCD, sudden cardiac death; VF, ventricular fibrillation; VT, ventricular tachycardia

2021 PACES expert consensus statement on the indications and management of cardiovascular implantable electronic devices in pediatric patients: executive summary

Developed in collaboration with and endorsed by the Heart Rhythm Society (HRS), the American College of Cardiology (ACC), the American Heart Association (AHA), and the Association for European Paediatric and Congenital Cardiology (AEPC). Endorsed by the Asia Pacific Heart Rhythm Society (APHRS), the Indian Heart Rhythm Society (IHRS), and the Latin American Heart Rhythm Society (LAHRS).

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Preamble

Guidelines for the implantation of cardiac implantable electronic devices (CIEDs) have evolved since publication of the initial ACC/AHA pacemaker guidelines in 1984.[1] CIEDs have evolved to include novel forms of cardiac pacing, the development of implantable cardioverter defibrillators (ICDs) and the introduction of devices for long term monitoring of heart rhythm and other physiologic parameters. In view of the increasing complexity of both devices and patients, practice guidelines, by necessity, have become increasingly specific. In 2018, the ACC/AHA/HRS published Guidelines on the Evaluation and Management of Patients with Bradycardia and Cardiac Conduction Delay,[2] which were specific recommendations for patients >18 years of age. This age-specific threshold was established in view of the differing indications for CIEDs in young patients as well as size-specific technology factors. Therefore, the following document was developed to update and further delineate indications for the use and management of CIEDs in pediatric patients, defined as ≤21 years of age, with recognition that there is often overlap in the care of patents between 18 and 21 years of age.

This document is an abbreviated expert consensus statement (ECS) intended to focus primarily on the indications for CIEDs in the setting of specific disease/diagnostic categories. This document will also provide guidance regarding the management of lead systems and follow-up evaluation for pediatric patients with CIEDs. The recommendations are presented in an abbreviated modular format, with each section including the complete table of recommendations along with a brief synopsis of supportive text and select references to provide some context for the recommendations. This document is not

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Table 1. Class of recommendation and level of evidence categories*

Class I	Class IIa	Class IIb	Class III
Benefit >>> ***Risk*** Procedure/treatment **SHOULD** be performed/is recommended	***Benefit*** >> ***Risk*** **IT IS REASONABLE** to perform the procedure/treatment	***Benefit*** ≥ ***Risk*** Procedure/treatment **MAY BE CONSIDERED**/ effectiveness is uncertain	***Risk*** ≥ ***Benefit*** Procedure should **NOT** be performed **IS NOT HELPFUL/MAY BE HARMFUL**
Levels of Evidence			
B-NR: Evidence from nonrandomized studies, observational studies, or registry studies			
C-LD: Very limited evidence from observational studies or case series reports			
C-EO: Consensus expert opinion, case studies, or standard of care			

*Adapted from Halperin, et al.[100]

intended to provide an exhaustive discussion of the basis for each of the recommendations, which are further addressed in the comprehensive PACES-CIED document,[3] with further data easily accessible in electronic searches or textbooks.

Introduction

Methodology and evidence review

The principles in the development of this document are 1) new recommendations or changes to previous recommendations are based on data, when possible; 2) these recommendations are consistent with current ACC/AHA/HRS adult guidelines when reasonable; and 3) all recommendations have been critically reviewed, initially by the writing committee and editors, followed by the PACES executive committee, and subsequently by external HRS, ACCF, AHA, and AEPC representatives. Any revisions or additions to existing recommendations require approval of at least 80% by the members of the PACES writing committee.

These recommendations have been developed with standard guideline methodology, i.e., with both a class of recommendation (COR) and a level of evidence (LOE) (Table 1). The class of the recommendation indicates the strength of recommendation, based on the estimated magnitude or certainty of benefit in proportion to risk. The level of evidence rates the quality of evidence based on the type, quantity, and consistency of data from clinical trials and other sources. A recommendation with a Level of Evidence C-EO does not imply that the recommendation is weak. Many of the questions addressed in this (and other) documents either do not lend themselves to clinical trials or are rare disease entities. However, there may be unequivocal consensus that a particular intervention is either effective or necessary.

Organization of the writing committee

The writing committee consisted of members of PACES who were selected by the PACES executive committee. The writing committee members included junior and senior pediatric electrophysiologists as well as allied health professionals and represented diverse genders, countries, and cultures. The writing committee also included external representatives from the ACC, AHA, HRS, and AEPC. Prior to final publication, all committee members were required to verify their specific contributions to this document. Appendix 1 lists writing committee members' relevant relationships with industry.

Document review and approval

Following internal review by the PACES executive committee, this document was then reviewed by the PACES writing committee. Following considerations of these comments and approval by an independent PACES reviewer, the recommendations were opened for public comment to PACES members. An official reviewer each nominated by HRS, ACC, AHA, and AEPC provided independent external review. This document was then approved for publication by the PACES executive committee and endorsed by all collaborators and the Asia Pacific Heart Rhythm Society (APHRS), the Indian Heart Rhythm Society (IHRS), and the Latin American Heart Rhythm Society. Appendix 2 lists reviewers' relevant relationships with industry.

Health policy objectives

The purpose of this document is to provide guidance to clinicians for the management of pediatric patients who may require a CIED, with a primary focus on the indications for device implantation. The document will be useful to pediatric cardiologists, cardiac surgeons, cardiac intensivists, anesthesiologists, and arrhythmia specialists. This document supersedes the pediatric CIED recommendations made in "ACC/AHA/HRS 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities"[4] and "2012 ACCF/AHA/HRS Focused Update of the 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities."[5]

Top 10 take-home messages

1. In patients with isolated sinus node dysfunction (SND), there is no minimum heart rate or maximum pause duration where permanent pacing is absolutely recommended. Establishing a temporal correlation between symptoms and bradycardia is critical in the decision as to whether permanent pacing is indicated.
2. Young patients with impaired ventricular function or abnormal cardiovascular physiology may be symptomatic due to sinus bradycardia or the loss of atrioventricular (AV) synchrony at heart rates that do not produce symptoms in individuals with normal cardiovascular physiology.
3. Although the average ventricular rate in newborns and infants with congenital complete atrioventricular block (CCAVB) provides an objective measure regarding the decision for pacemaker implantation, additional factors may equally influence the decision/timing of pacemaker implant. These include birth

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weight (size), congenital heart defects, ventricular function, and other comorbidities.
4. In patients with postoperative AV block, a period of observation for at least 7–10 days before pacemaker implantation remains advised; in select cases, earlier pacemaker implantation may be considered if AV block is not expected to resolve due to extensive injury to the cardiac conduction system.
5. Atrial pacing with antitachycardia pacing capabilities is reasonable for congenital heart disease (CHD) patients with recurrent intra-atrial reentrant tachycardia when medication and catheter ablation are not effective.
6. There is increased recognition of the need for pacemaker implantation in conditions such as Kearns-Sayre syndrome or certain neuromuscular disorders due to the unpredictable progression of conduction disease.
7. The cause of sudden cardiac arrest (SCA) remains undefined in nearly 50% of pediatric survivors. ICD implantation is recommended provided completely reversible causes have been excluded, other treatments that may be beneficial are considered, and meaningful survival is anticipated.
8. The decisions for implantation of an ICD for primary prevention in cardiac channelopathies or cardiomyopathies remain guided by limited and, at times, conflicting data. Consideration of patient-specific factors and shared decision-making are critically important.
9. In pediatric patients with nonischemic dilated cardiomyopathy (NIDCM), primary prevention ICD implantation for left ventricular ejection fraction (LVEF) $\leq 35\%$, in the absence of other risk factors, is not clearly supported by published data.
10. In patients with indications for implantation of a CIED, shared decision-making and patient/family-centered care are endorsed and emphasized. Treatment decisions are based on the best available evidence and patient's preferences.

Permanent pacemakers

Introduction

The most common indications for permanent pacemaker implantation in children, adolescents, and patients with CHD are 1) symptomatic sinus bradycardia, 2) advanced second- or third-degree AV block, and 3) pacing for the prevention or termination of tachyarrhythmias.[4] Many indications for pacemaker implantation in adolescents are similar to those in adults.[2] However, in infants and young children, there are important differences. For example, criteria for normal heart rates are an age-dependent variable; whereas a heart rate of 45 bpm is normal in an adolescent, the same rate in a newborn or infant indicates profound bradycardia. In addition, young patients with impaired ventricular function or abnormal physiology may be symptomatic due to sinus bradycardia or loss of AV synchrony at heart rates that do not produce symptoms in individuals with normal cardiovascular physiology.[6,7] Hence, the indications for pacemaker implantation in young patients need to be based on the correlation of symptoms with relative bradycardia rather than absolute heart rate criteria.

Significant technical challenges may complicate device and lead implantation in small patients or those with abnormalities of venous or intracardiac anatomy. Epicardial lead placement and innovative use of device technology may be needed to provide pacing or defibrillation in young patients. Furthermore, as device leads may need to be utilized for multiple decades, consideration of the potential consequences from lead failure plays a major role in implantation of pediatric devices.

Isolated sinus node dysfunction

COR	Recommendations	LOE	References
	Isolated Sinus Node Dysfunction		
I	Permanent atrial or dual-chamber pacemaker implantation is indicated for SND when there is correlation of symptoms with age-inappropriate bradycardia.	B-NR	4,8,9
I	Permanent pacemaker implantation is indicated in patients with symptomatic SND secondary to chronic medical therapy for which there is no alternative treatment.	C-EO	
IIa	Permanent pacemaker implantation (with rate-responsive programming) is reasonable in patients with symptoms temporally associated with observed chronotropic incompetence.	C-LD	10
IIb	Permanent pacemaker implantation may be considered in patients with SND and symptoms that are likely attributable to bradycardia or prolonged pauses without conclusive evidence correlating the symptoms with bradycardia following a thorough investigation.	C-EO	
III No Benefit	Permanent pacemaker implantation is not indicated in patients with asymptomatic SND.	C-EO	
III Harm	Permanent pacemaker implantation is not indicated in patients with symptomatic SND due to a reversible cause.	C-EO	

Recommendation-specific supportive text

Sinus node dysfunction (SND) refers to physiologically inappropriate atrial rates, either due to sustained bradycardia or abrupt pauses in the intrinsic cardiac rhythm. In patients with isolated sinus bradycardia without symptoms due to cerebral or systemic hypoperfusion, there is no minimum heart rate or maximum pause duration where permanent pacing is recommended.[2] Establishing a temporal correlation between symptoms and age-related bradycardia is of paramount importance when determining whether permanent pacing is needed. In symptomatic patients with SND, atrial-based pacing is generally recommended over single chamber ventricular pacing.[11]

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Isolated congenital complete atrioventricular block

COR	Recommendations Isolated Congenital Complete Atrioventricular Block	LOE	References
I	Permanent pacemaker implantation is indicated for patients with CCAVB with symptomatic bradycardia.	B-NR	12,13
I	Permanent pacemaker implantation is indicated for patients with CCAVB with a wide QRS escape rhythm, complex ventricular ectopy, or ventricular dysfunction.	B-NR	2,12
I	Permanent pacemaker implantation is indicated for CCAVB in asymptomatic neonates or infants when the mean ventricular rate is ≤50 bpm. Ventricular rate alone should not be used as implant criteria, as symptoms due to low cardiac output may occur at faster heart rates.	C-LD	12,14
IIa	Permanent pacemaker implantation is reasonable for asymptomatic CCAVB beyond the first year of life when the mean ventricular rate is <50 bpm or there are prolonged pauses in ventricular rate.	B-NR	2,4,15
IIa	Permanent pacemaker implantation is reasonable for CCAVB with left ventricular dilation (z score ≥3) associated with significant mitral insufficiency or systolic dysfunction.	C-LD	16
IIb	Permanent pacemaker implantation may be considered for CCAVB in asymptomatic adolescents with an acceptable ventricular rate, a narrow QRS complex, and normal ventricular function, based on an individualized consideration of the risk/benefit ratio.	C-LD	2

Recommendation-specific supportive text

The average ventricular rate in neonates and infants with isolated CCAVB provides one objective parameter regarding the decision for pacemaker implantation. However, additional factors including birth weight (size), ventricular dysfunction, and other co-morbidities may equally influence the decision. Therefore, an average heart rate of ≤50 bpm is recommended for infant pacemaker implantation when overt symptoms related to low cardiac output are not present. Beyond the first year of life, permanent pacemaker implantation is generally indicated in symptomatic patients. Natural history studies have demonstrated progressive LV dysfunction and mitral insufficiency with cardiovascular mortality in the 4th or 5th decade in CCAVB patients who did not undergo pacemaker implantation.[17]

Atrioventricular block: other considerations

COR	Recommendations Atrioventricular Block: Other Considerations	LOE	References
I	Permanent pacemaker implantation is indicated in patients with clinically significant ventricular tachycardia (VT) that is pause dependent or associated with severe bradycardia; ICD implantation may be considered as a reasonable alternative.	C-LD	18
I	Permanent pacing is indicated in *symptomatic* patients with idiopathic advanced second- or third-degree AV block not attributable to reversible causes.	C-LD	
IIa	Permanent pacemaker implantation is reasonable for any degree of AV block that progresses to advanced second- or third-degree with exercise in the absence of reversible causes.	C-LD	19
IIb	Permanent pacemaker implantation may be considered for patients with intermittent advanced second- or third-degree AV block not attributable to reversible causes and associated with minimal symptoms that are otherwise unexplained.	C-LD	
III Harm	Permanent pacemaker implantation is not indicated for asymptomatic first-degree AV block or asymptomatic second-degree Mobitz type I.	C-LD	2,4

Recommendation-specific supportive text

Advanced AV block diagnosed during childhood or adolescence may be congenital, related to infiltrative diseases or remain idiopathic. At times, late-onset AV block may be paroxysmal and difficult to document. Exercise testing may be useful regarding the significance of AV block. When progressive AV block occurs during exercise, conduction disturbance within the His-Purkinje system is suspected and is associated with a poor prognosis.[19] With the exception of infiltrative or inflammatory causes, the criteria for pacemaker implantation are similar to those for CCAVB.

Postoperative atrioventricular block

COR	Recommendations Postoperative Atrioventricular Block	LOE	References
I	Permanent pacemaker implantation is indicated for postoperative advanced second- or third-degree AV block that persists for at least 7–10 days after cardiac surgery.	B-NR	20,21
I	Permanent pacemaker implantation is indicated for late-onset advanced second- or third-degree AV block especially when there is a prior history of transient postoperative AV block.	C-LD	22

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COR	Recommendations Postoperative Atrioventricular Block	LOE	References
IIb	Permanent pacemaker implantation may be considered for unexplained syncope in patients with a history of transient postoperative advanced second- or third-degree AV block.	C-LD	23,24
IIb	Permanent pacemaker implantation may be considered at <7 postoperative days when advanced second- or third-degree AV block is not expected to resolve due to extensive injury to the cardiac conduction system.	C-EO	
IIb	Permanent pacemaker implantation may be considered in select patients with transient postoperative advanced second- or third-degree AV block who are predisposed to progressive conduction abnormalities (see text).	C-EO	

Recommendation-specific supportive text

Postoperative AV block complicates 3–8% of congenital heart surgeries, with 1–3% of patients requiring permanent pacemaker implantation for persistent postoperative AV block.[21] A very poor prognosis has been established for CHD patients with permanent postoperative AV block who do not receive permanent pacemakers. Among patients who regain AV conduction following transient AV block, ≥85% have recovery of AV conduction by post-operative day 7 and ≥95% AV conduction by postoperative day 10.[20,21] Although patients who regain AV conduction have a favorable prognosis, there is a small risk of late-onset complete AV block in transient postoperative AV block patients.[22] Permanent pacemaker implantation may be considered for transient postoperative third-degree AV block that reverts to normal AV node conduction in patients with forms of CHD which may develop progressive AV block such as discordant AV connections, AV septal defects and heterotaxy syndromes.

Congenital heart disease: specific considerations

COR	Recommendations Congenital Heart Disease	LOE	References
	All the recommendations in children with a structurally normal heart apply, but in addition:		
I	Permanent pacemaker implantation is indicated for CCAVB in neonates or infants with complex CHD when bradycardia is associated with hemodynamic compromise or when the mean ventricular rate is <60–70 bpm.	C-LD	25
IIa	Permanent pacemaker implantation with atrial antitachycardia pacing is reasonable for patients with CHD and recurrent episodes of intra-atrial re-entrant tachycardia when catheter ablation or medication are ineffective or not acceptable treatments.	B-NR	7,26

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(*Continued*)

COR	Recommendations Congenital Heart Disease	LOE	References
IIa	Permanent atrial or dual-chamber pacemaker implantation is reasonable for patients with CHD and impaired hemodynamics due to sinus bradycardia or loss of AV synchrony.	C-LD	7
IIa	Permanent atrial or dual-chamber pacing is reasonable for patients with tachy-brady syndrome and symptoms attributable to pauses due to sudden-onset bradycardia.	C-LD	
IIa	Permanent pacemaker implantation is reasonable for sinus or junctional bradycardia with *complex* CHD when the mean awake resting heart rate is <40 bpm or when there are prolonged pauses in the ventricular rate.	C-EO	
IIb	Permanent pacing may be considered for sinus or junctional bradycardia with *simple or moderate* CHD when the mean awake resting heart rate is <40 bpm or when there are prolonged pauses in the ventricular rate.	C-EO	
III Harm	Endocardial leads should be avoided in patients with CHD and intracardiac shunt except in select cases, for whom there should be an individualized consideration of the risk/benefit ratio. In these exceptional cases anticoagulation is mandatory, but thromboembolism remains a risk.	B-NR	27

Recommendation-specific supportive text

Patients with CHD often have important structural and functional lesions which influence both the indications for pacing as well as the type of pacing lead(s) utilized. Therefore, pacemaker implantation in these patients is not an isolated procedure. Bradycardia and scar related tachycardias are common following surgery, and in the absence of high-grade AV block, atrial pacing is preferred to avoid pacing-induced ventricular dysfunction. Permanent pacemaker and/or lead implantation may be considered at the time of surgery in patients with restricted vascular access or evidence of conduction disease in heart defects with a known natural progression to advanced heart block. Decisions regarding pacemaker implantation must also consider the complexity of the patient's anatomy, surgical repair and hemodynamic status.

Post cardiac transplantation

COR	Recommendations Post Cardiac Transplantation	LOE	References
I	Permanent pacing is indicated for persistent symptomatic bradycardia that is not expected to resolve and for other class I indications for permanent pacing.	C-LD	4,28

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COR	Recommendations Post Cardiac Transplantation	LOE	References
IIa	Permanent pacing is reasonable for marked chronotropic incompetence impairing the quality of life late in the post-transplant period.	C-LD	
IIb	Permanent pacing may be considered when relative bradycardia is prolonged, recurrent, or limits rehabilitation or discharge after postoperative recovery from cardiac transplantation.	C-LD	28
IIb	Permanent pacing may be considered for any degree of AV block considered to be due to graft vasculopathy.	C-LD	29

Recommendation-specific supportive text

Transient sinus bradycardia is common immediately after transplantation and typically resolves. In rare cases, symptomatic sinus bradycardia may persist, with at least one week allowed for recovery of sinus node function. Analysis of the United Network Organ Sharing database reported that 1% of heart transplant patients <18 years of age required a pacemaker in the acute post-transplant interval. Factors associated with need for pacing were bi-atrial anastomosis, older donor age and antiarrhythmic use.[28] Late onset conduction disorders (sinus node or AV node dysfunction) may be related to cardiac allograft vasculopathy or allograft rejection. Patients should be evaluated for the presence of transplant coronary artery disease, as late onset bradycardia may be the first manifestation. The role of prophylactic ICD implantation is not well established but may be considered in patients who require pacemakers.

Neuromuscular diseases and other progressive cardiac conduction diseases

COR	Recommendations Neuromuscular Diseases and Other Progressive Cardiac Conduction Diseases	LOE	References
I	Permanent pacemaker implantation is indicated in patients with neuromuscular diseases with symptomatic bradycardia due to SND or any degree of AV block.	B-NR	2,30
I	Permanent pacemaker implantation is indicated in Kearns-Sayre syndrome for any degree of AV block (including first-degree AV block) and/or conduction abnormality because of unpredictable progression of conduction disease.	C-LD	31
IIa	Permanent pacemaker implantation is reasonable in patients with myotonic dystrophy type 1 for marked first-degree AV block (PR interval >240 ms) or intraventricular conduction delay (native QRS duration >120 ms). Additional defibrillator capability may be considered.	B-NR	32

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(*Continued*)

COR	Recommendations Neuromuscular Diseases and Other Progressive Cardiac Conduction Diseases	LOE	References
IIa	Permanent pacemaker implantation is reasonable in patients with lamin A/C gene mutations, including limb-girdle and Emery-Dreifuss muscular dystrophies with a PR interval >240 ms and/or left bundle branch block. Additional defibrillator capability may be considered.	C-LD	33
IIb	Permanent pacemaker implantation may be considered for any patient with any progressive cardiac conduction disease with potential for rapid deterioration of AV nodal function, even in the presence of normal AV conduction after taking into consideration patient age, size, and other individual risk factors.	C-LD	

Conditions include Duchenne muscular dystrophy, Becker muscular dystrophy, myotonic dystrophy type 1, Friedreich ataxia, Emery-Dreifuss muscular dystrophy, facioscapulohumeral muscular dystrophy, Barth syndrome, Kearns-Sayre syndrome, lamin A/C mutations, and desmin-related myopathies.

Recommendation-specific supportive text

Progressive cardiac conduction diseases are genetic disorders with deterioration of the conduction system either in isolation or in conjunction with other diseases such as neuromuscular and mitochondrial diseases.[30] Variable degrees of conduction abnormalities may occur, from first-degree AV block to complete AV block with an unpredictable progression. Laminopathies caused by mutations in the *LMNA* genes is a wide spectrum disorder with cardiac conduction abnormalities often observed before the onset of heart failure symptoms.[32] Among the mitochondrial diseases, Kearns-Sayre syndrome, with progressive ophthalmoplegia and myopathy, has a high risk for AV block and sudden cardiac death (SCD).[31] Currently, an HRS expert consensus statement on the evaluation and management of arrhythmic risk in neuromuscular disorders is under development. Therefore, the above recommendations may be subject to modification as newer data become available.

Neurocardiogenic syncope

COR	Recommendations Neurocardiogenic Syncope	LOE	References
IIa	Permanent pacemaker implantation is reasonable with severe recurrent breath-holding spells with documentation of cardioinhibitory response on ECG monitoring and complicated by prolonged syncope, prolonged postanoxic convulsions, and other bradycardia-induced symptoms.	B-NR	34,35

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COR	Recommendations Neurocardiogenic Syncope	LOE	References
IIb	Permanent pacing may be considered for recurrent symptomatic neurocardiogenic syncope associated with documented spontaneous bradycardia or asystole in patients who have failed other medical treatments.	C-LD	36,37
IIb	Permanent pacemaker implantation may be considered in patients with epilepsy associated with severe symptomatic bradycardia (ictal induced) who have failed to improve with antiepileptic medical therapy.	C-LD	38
III **No benefit**	Permanent pacing is not indicated for neurocardiogenic syncope solely on the basis of a positive cardioinhibitory tilt response.	C-EO	
III Harm	Permanent pacing is not indicated for neurocardiogenic syncope with hypotension as the major or significant component of the symptoms.	C-EO	

Recommendation-Specific Supportive text

In the vast majority of cases, neurocardiogenic syncope is a limited disease and pacemaker implantation is not required. However, in some patients, recurrent syncopal events may significantly impair quality of life and may result in traumatic injury, particularly when the dominant feature of reflex syncope is cardioinhibitory. Therefore, in a highly select group of patients who fail more conservative treatment options, pacemaker therapy may be useful by preventing profound bradycardia or prolonged asystole.[36] Because the efficacy of pacing depends on the clinical setting, a clear relationship between symptoms and bradycardia or asystole should be established prior to pacemaker implantation.[36,37]

Cardiac channelopathies

COR	Recommendations Cardiac Channelopathies	LOE	References
I	Permanent pacemaker implantation is indicated in channelopathy patients with pause-dependent, clinically significant VT; ICD implantation may be considered as a reasonable alternative.	C-LD	39

(*Continued*)

(*Continued*)

COR	Recommendations Cardiac Channelopathies	LOE	References
IIb	Permanent pacemaker implantation may be considered as adjunctive therapy in patients with long QT syndrome and functional 2:1 AV block.	C-LD	40
IIb	Permanent pacemaker implantation may be considered as adjunctive therapy in patients with long QT syndrome or other channelopathies where a faster heart rate may decrease the arrhythmia burden or symptoms due to bradycardia.	C-LD	41
III **No benefit**	Atrial pacing alone is not indicated in patients with complete atrial standstill due to the high potential for noncapture of the myocardium.	C-LD	

Recommendation-specific supportive text

The utility of pacing as adjunctive therapy in the various channelopathies is not well defined. In patients with bradycardia-related or pause-related initiation of ventricular tachyarrhythmias, permanent pacemaker implantation may provide benefit. Also, pacing has been reported to improve outcomes in infants with prolonged QT-related functional 2:1 AV block.[40] Limited data also suggest that atrial pacing faster than the intrinsic rate may shorten the QT interval and reduce the rate of recurrent syncopal events in select high-risk long QT syndrome (LQTS) patients.[41]

Inflammation/infection

COR	Recommendations Inflammation/Infection	LOE	References
I	Permanent pacing is indicated in patients with high-grade or symptomatic AV block attributable to a known potentially reversible cause when AV block does not resolve despite treatment of the underlying cause.	C-LD	42
IIa	Pacemaker implantation is reasonable in Chagas disease and advanced second- or third-degree AV block, as spontaneous resolution is unlikely. ICD implantation may be a reasonable alternative.	C-LD	43
III **No benefit**	Permanent pacing should not be performed in patients who had acute AV block attributable to a known reversible cause, when there is recovery of normal AV conduction.	C-EO	

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Recommendation-specific supportive text

Systemic infections may cause myocardial inflammation or infiltration presenting with bradycardia or complete AV block. In most cases, there is recovery of AV conduction. However, in chronic Chagas disease, advanced heart block in Chagas is permanent and pacemaker implantation is indicated.[43] Limited data suggest that children who develop AV block due to coronavirus 2019 (COVID-19)–related multisystem inflammatory syndrome will have recovery of normal AV conduction.[44]

Implantable cardioverter defibrillators

Introduction

The following recommendations for ICD implantation are primarily based on contemporary adult guidelines, and with some modifications, applied to younger patients. Adult ICD guidelines have been established based on a specific diagnosis or presumed risk factor for a sudden cardiac event, such as ischemia, cardiomyopathy, or genetic cardiovascular disease.[4,45,46] In contrast, studies of pediatric sudden cardiac arrest (SCA) survivors demonstrate that in approximately 50% of cases, the cause of the event remains undefined despite an extensive systematic evaluation.[47,48] Furthermore, in young patients with diagnoses such as catecholaminergic polymorphic ventricular tachycardia (CPVT) or Brugada syndrome (BrS), SCA is often the initial presentation of the disease.[49,50] Therefore, while development of pediatric ICD recommendations based on specific cardiovascular diagnoses would be preferable, the following recommendations for ICD implantation will begin with general considerations for young patients, followed by more nuanced recommendations for ICD implantation when a specific cause or a defined risk factor for SCA has been identified. There remain extensive "gaps" in current ICD recommendations, irrespective of age, for many of the diseases associated with SCD in pediatrics.[51] The recommendations that follow are largely based on limited clinical data or expert opinion and consensus and require the application of case-specific clinical judgment and a shared decision approach.

General recommendations for implantable cardioverter defibrillator therapy

	Recommendations General Recommendations for Implantable Cardioverter Defibrillator Therapy		
COR		LOE	References
I	ICD implantation is indicated for survivors of SCA due to VT/ventricular fibrillation (VF) if completely reversible causes have been excluded and an ICD is considered to be more beneficial than alternative treatments that may significantly reduce the risk of SCA.	B-NR	4,45,46
IIb	ICD implantation may be considered for patients with sustained VT that cannot be adequately controlled with medication and/or catheter ablation.	C-EO	

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	Recommendations General Recommendations for Implantable Cardioverter Defibrillator Therapy		
COR		LOE	References
IIb	ICD therapy may be considered for primary prevention of SCD in patients with genetic cardiovascular diseases and risk factors for SCA or pathogenic mutations and family history of recurrent SCA.	C-EO	
III Harm	ICD therapy is not indicated for patients with incessant ventricular tachyarrhythmias due to risk of ICD storm.	C-EO	
III Harm	ICD therapy is not indicated for patients with ventricular arrhythmias that are adequately treated with medication and/or catheter ablation.	C-LD	52,53
III Harm	ICD therapy is not indicated for patients who have an expected survival <1 year, even if they meet ICD implantation criteria specified in the above recommendations.	C-EO	
III Harm	Endocardial leads should be avoided in patients with intracardiac shunts except in select cases, when there should be an individualized consideration of the risk/benefit ratio. In these exceptional cases anticoagulation is mandatory, but thromboembolism remains a risk.	B-NR	27

Recommendation-specific supportive text

The decisions regarding ICD implantation pediatric patients are made in the context of both the unique aspects of device implantation as well as pathogenesis of the disease, which may evolve over time. Therefore, a pediatric cardiologist should be involved in the ICD implant decision and the procedure should be performed by a cardiologist or surgeon with special training/experience in CIED implantation in the pediatric age group. ICD implantation should be a shared decision between the patient, family, and physician. This includes the physical and psychological impact of an ICD on the patient's well-being. Furthermore, the indications for the ICD should be reconsidered at each reintervention with respect to current guidelines, especially after a period of nonuse, as discontinuation of device therapy may be considered in select cases.[54]

ICD indications for cardiac channelopathies

Long QT syndrome

	Recommendations Long QT Syndrome		
COR		LOE	References
I	ICD implantation along with the use of beta-blockade is indicated for patients with a diagnosis of LQTS who are survivors of SCA. In select LQTS patients, medical therapy and/or cardiac sympathetic denervation may be considered as an alternative.	B-NR	55,56

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COR	Recommendations Long QT Syndrome	LOE	References
I	ICD implantation is indicated in LQTS patients with symptoms (arrhythmic syncope or VT) in whom beta-blockade is either ineffective or not tolerated and cardiac sympathetic denervation or other medications are not considered effective alternatives.	B-NR	57,58
IIb	ICD therapy may be considered for primary prevention in LQTS patients with established clinical risk factors and/or pathogenic mutations (see text).	C-LD	59
III Harm	ICD implantation is not indicated in asymptomatic LQTS patients who are deemed to be at low risk of SCA and have not been tried on beta-blocker therapy.	C-LD	60

Recommendation-specific supportive text

Both phenotypic and genotypic characteristics are used to guide risk stratification when patients with LQTS may require ICD therapy.[61] Phenotypic risk factors include the onset of symptoms at age <10 years, patients with prior SCA or those with recurrent syncope. Additional high-risk factors include a QTc ≥ 550 ms regardless of genotype, QTc ≥ 500 ms with LQT1, females with LQT2 and males with LQT3 genotype. Non-selective beta blockers are considered first line therapy and can significantly decrease subsequent cardiac events in patients, especially in those with KCNQ1 mutations.[45] In addition, beta-blockers and cardiac sympathetic denervation without ICD may be appropriate in carefully selected patients.[58] Conversely, ICD implantation in an asymptomatic low-risk patient with LQTS for a positive family history of LQTS related SCD is not clearly supported by published data and requires case-specific decision making.

Catecholaminergic polymorphic ventricular tachycardia

COR	Recommendations Catecholaminergic Polymorphic Ventricular Tachycardia	LOE	References
I	ICD implantation is indicated in patients with a diagnosis of CPVT who experience cardiac arrest or arrhythmic syncope despite maximally tolerated beta-blocker plus flecainide and/or cardiac sympathetic denervation.	C-LD	55,62
IIa	ICD implantation is reasonable in combination with pharmacologic therapy with or without cardiac sympathetic denervation when aborted SCA is the initial presentation of CPVT. Pharmacologic therapy and/or cardiac sympathetic denervation without ICD may be considered as an alternative.	C-LD	49,63

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COR	Recommendations Catecholaminergic Polymorphic Ventricular Tachycardia	LOE	References
IIb	ICD implantation may be considered in CPVT patients with polymorphic/bidirectional VT despite optimal pharmacologic therapy with or without cardiac sympathetic denervation.	C-LD	64
III Harm	ICD implantation is not indicated in asymptomatic patients with a diagnosis of CPVT.	C-EO	

Recommendation-specific supportive text

SCA/SCD is reported in 3 to 13% of CPVT patients.[49,62] High-risk factors include male gender, previous history of cardiac arrest, multiple genetic variants, and younger age at diagnosis. Complex ventricular ectopy on exercise testing despite optimal medical therapy is also associated with worse outcome. Treatment with nonselective beta blockers is associated with a significant reduction in adverse cardiac events, while the addition of flecainide to refractory patients may provide further benefit. In general, ICD implantation should be reserved for CPVT patients with prior SCA or with refractory ventricular arrhythmias on combination medical therapy.[49,62] Inappropriate shocks are reported in 20–30% of CPVT patients with ICDs with cardiac sympathetic denervation recommended in patients who experience recurrent ICD shocks.[62] In selected patients with aborted SCA as the initial presentation of CPVT, pharmacologic therapy and/or cardiac sympathetic denervation without ICD may be considered as a possible alternative.

Brugada syndrome

COR	Recommendations Brugada Syndrome	LOE	References
I	ICD implantation is indicated in patients with a diagnosis of BrS who are survivors of SCA or have documented spontaneous sustained VT.	B-NR	65,66
IIa	ICD implantation is reasonable for patients with BrS with a spontaneous type I Brugada ECG pattern and recent syncope presumed due to ventricular arrhythmias.	B-NR	67,68
IIb	ICD implantation may be considered in patients with syncope presumed due to ventricular arrhythmias with a type I Brugada ECG pattern only with provocative medications.	C-EO	
III No benefit	ICD implantation is not indicated in asymptomatic BrS patients in the absence of risk factors.	C-EO	

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Recommendation-specific supportive text

Although Brugada syndrome presents typically in the 4th to 5th decade, it may have onset during childhood, with rapid progression leading to life-threatening arrhythmias. The ICD remains the only therapy with proven efficacy for the management of ventricular arrhythmias or SCA in patients with Brugada syndrome.[65] Adult recommendations for risk stratification including ventricular stimulation have been established, but have not been validated in pediatrics. Findings associated with high risk of ventricular arrhythmias and SCD in children include in order of relevance: the presence of symptoms (SCD or arrhythmogenic syncope), spontaneous coved type ST elevation (type I electrocardiogram [ECG] pattern), atrial arrhythmias and/or sinus node dysfunction and conduction abnormalities (AV block or intra-ventricular conduction delay).[67] Conversely, implantation of an ICD is not indicated in asymptomatic patients in the absence of risk factors. Further studies are necessary to further characterize risk factors and primary prevention ICD indications for pediatric patients with Brugada syndrome.

ICD indications for cardiomyopathies

Hypertrophic cardiomyopathy

COR	Recommendations Hypertrophic Cardiomyopathy	LOE	References
I	ICD implantation is indicated in patients with HCM who are survivors of SCA or have spontaneous sustained VT.	B-NR	45,69
IIa	For children with HCM who have ≥1 primary risk factors, including unexplained syncope, massive left ventricular hypertrophy, nonsustained VT, or family history of early HCM-related SCD. ICD placement is reasonable after considering the potential complications of long-term ICD placement.	B-NR	70,71
IIb	ICD implantation may be considered in patients with HCM without the above risk factors but with secondary risk factors for SCA such extensive LGE on cardiac MRI or systolic dysfunction.	C-LD	
III Harm	ICD implantation is not indicated in patients with an identified HCM genotype in the absence of known pediatric SCA risk factors.	C-LD	

Recommendation-specific supportive text

Estimates for SCD rates in childhood hypertrophic cardiomyopathy (HCM) vary widely, with epidemiologic studies reporting rates between 1 and 7% per year. While ICDs have improved the outcomes for HCM patients resuscitated from SCD, accurate identification of risk factors to guide primary prevention ICD implantation remains a challenge, particularly given the potential progression of the disease process over time.[71] Adult clinical practice guidelines define high risk for SCD in HCM by the presence of ≥1 defined clinical risk factors.[45,69] However, recent pediatric studies suggest that the significance of adult risk factors may differ in children. A multi-center pediatric study reported that an LV posterior wall thickness z score ≥5 was associated with SCA, while a meta-analysis of pediatric studies reported a maximum LV wall thickness ≥30 mm or a z-score ≥6 associated with an increased risk of SCD.[71] The significance of secondary risk factors for SCD, such as late gadolinium enhancement (LGE) on cardiac magnetic resonance imaging (MRI) and the role of genetic testing for specific "malignant" sarcomere mutations, remains debated and requires further investigation before inclusion as specific risk factors for SCD in pediatric patients with HCM.

Arrhythmogenic cardiomyopathies

COR	Recommendations Arrhythmogenic Cardiomyopathies	LOE	References
I	ICD implantation is indicated in patients with ACM who have been resuscitated from SCA or sustained VT that is not hemodynamically tolerated.	B-NR	45,72
IIa	ICD implantation is reasonable in patients with ACM with hemodynamically tolerated sustained VT, syncope presumed due to ventricular arrhythmia, or an LVEF ≤35%.	B-NR	72
IIb	ICD implantation may be considered in patients with inherited ACM associated with increased risk of SCD based on an assessment of additional risk factors.	C-LD	

Recommendation-specific supportive text

Arrhythmogenic cardiomyopathy (ACM) encompasses a spectrum of primary myocardial disorders with the key feature of presentation with sustained arrhythmias.[72] This includes genetic disorders such as arrhythmogenic right/left ventricular cardiomyopathy, lamin A/C mutations, filamin-C, phospholamban, and cardiac amyloidosis. These entities are infrequent before puberty, and often overlap with other cardiomyopathies. Overall, SCD occurs in 2–15% of young patients with ACM. Patients presenting with SCD and/or sustained VT have a class I ICD indication. Although risk stratification data are minimal, ICD implantation is reasonable in ACM patients with hemodynamically tolerated sustained VT, syncope presumed due to ventricular arrhythmia, or an LVEF ≤ 35%. Heart transplantation and whether a wearable external defibrillator is reasonable should be considered on an individual basis for those patients with advanced heart failure.

Nonischemic dilated cardiomyopathy

COR	Recommendations Nonischemic Dilated Cardiomyopathy	LOE	References
I	ICD implantation is indicated in patients with NIDCM who either survive SCA or experience sustained VT not due to completely reversible causes.	B-NR	45,73

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COR	Recommendations Nonischemic Dilated Cardiomyopathy	LOE	References
IIb	ICD implantation may be considered in patients with NIDCM and syncope or an LVEF ≤ 35%, despite optimal medical therapy.	C-LD	[74]
III Harm	ICD implantation is NOT recommended in patients with medication-refractory advanced heart failure who are not cardiac transplantation or left ventricular assist device candidates.	C-EO	
III **No benefit**	ICD therapy is not indicated for patients with advanced heart failure who are urgently listed for cardiac transplantation and will remain in the hospital until transplantation, even if they meet ICD implantation criteria specified in the above recommendations.	C-EO	

Recommendation-specific supportive text

The annual incidence of SCD in pediatric patients with NIDCM is 1–5%, which is significantly less than in adult NIDCM patients.[75] Although studies have shown ICD survival benefit for secondary prevention in pediatric NIDCM, the low incidence of events has made it difficult to establish risk factors to guide recommendations for primary prevention ICD implantation.[73] However, in contrast to studies of adult patients with NIDCM and LVEF ≤ 35%, there is no clear evidence that ICDs implanted for primary prevention improve survival for pediatric patients with NIDCM. The phenotype of NIDCM may overlap with other cardiomyopathies resulting in variable risks of SCD. In the Sudden Death in Childhood Cardiomyopathy study, the cumulative incidence of SCD at 15 years was 5% for NIDCM compared to 23% for left ventricular noncompaction (LVNC).[74] Myocardial dysfunction and/or a history of clinically significant arrhythmias were strongly associated with mortality in LVNC. Therefore, factors which influence implantation of a primary prevention ICD include the NIDCM etiology, the cardiomyopathy phenotype, the degree of ventricular dysfunction and the presence of cardiac arrhythmias.

ICD indications for congenital heart disease

COR	Recommendations Congenital Heart Disease	LOE	References
I	ICD implantation is indicated for CHD patients who are survivors of SCA after evaluation to define the cause of the event and exclude any completely reversible causes.	B-NR	[7,76,77]

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COR	Recommendations Congenital Heart Disease	LOE	References
I	ICD implantation is indicated for CHD patients with hemodynamically unstable sustained VT who have undergone hemodynamic and electrophysiologic evaluation. Catheter ablation or surgical repair may be possible alternatives in carefully selected patients.	C-LD	[76,78]
IIa	ICD implantation is reasonable for CHD patients with systemic LVEF < 35% and sustained VT or presumed arrhythmogenic syncope.	C-LD	[7]
IIb	ICD implantation may be considered for CHD patients with spontaneous hemodynamically stable sustained VT who have undergone hemodynamic and electrophysiologic evaluation. Catheter ablation or surgical repair may be possible alternatives in carefully selected patients.	C-EO	
IIb	ICD implantation may be considered for CHD patients with unexplained syncope in the presence of ventricular dysfunction, nonsustained VT, or inducible ventricular arrhythmias at electrophysiologic study.	C-LD	[7]
IIb	ICD implantation may be considered for CHD patients with a single or systemic right ventricular ejection fraction ≤35%, particularly in the presence of additional risk factors such as VT, arrhythmic syncope, or severe systemic AV valve insufficiency.	C-EO	

Recommendation-specific supportive text

The association between CHD and ventricular arrhythmias is well established. First demonstrated in repaired tetralogy of Fallot, studies have identified risk factors for VT and SCD including residual cardiac defects, abnormal hemodynamics, and scar from prior interventions/surgeries. While correction of residual abnormalities or ablation of arrhythmogenic substrate may improve ventricular function or reduce symptoms, these may be inadequate to prevent subsequent VT or SCA. ICD placement may therefore be appropriate in patients with, or at high risk of, potentially life-threatening arrhythmias. The role of programmed stimulation and presence and degree of ventricular dysfunction as risk factors for SCD in CHD continues to be debated. ICD implantation in patients with CHD must consider anatomy, intracardiac shunts and vascular access. This may require non-standard approaches such as epicardial leads or subcutaneous ICDs.

Insertable cardiac monitors

COR	Recommendations Insertable Cardiac Monitors	LOE	References
I	Noninvasive cardiac rhythm monitoring is indicated in all patients prior to placement of an ICM.	B-NR	[2,79]

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COR	Recommendations Insertable Cardiac Monitors	LOE	References
I	ICM is indicated in syncopal patients with high-risk criteria when comprehensive evaluation does not define a cause of syncope or lead to a specific treatment, and who do not have conventional indications for a pacemaker or ICD.	B-NR	79,80
IIa	ICM is reasonable in the evaluation of patients with recurrent syncope of uncertain origin but not a high risk of SCD.	B-NR	81
IIa	ICM is reasonable in patients with infrequent symptoms (>30-day intervals) suspected to be due to an arrhythmia, when the initial noninvasive evaluation is nondiagnostic.	C-LD	82
IIa	ICM implantation is reasonable for guiding the management of patients with cardiac channelopathies or structural heart diseases associated with significant rhythm abnormalities.	C-LD	83
IIb	ICM may be considered in patients with suspected reflex syncope presenting with frequent or severe syncopal episodes.	C-LD	
IIb	ICM may be considered in carefully selected patients with suspected epilepsy in whom anticonvulsive treatment has proven ineffective.	C-LD	84
IIb	ICM may be considered in patients with severe but infrequent palpitations when other monitoring methods have failed to document an underlying cause.	C-LD	
IIb	ICM implantation may be considered for detecting subclinical arrhythmias in patients with cardiac channelopathies or other diseases associated with significant rhythm abnormalities.	C-EO	

Insertable cardiac monitors (ICMs) are subcutaneous devices which provide long term rhythm surveillance and provide documentation of rhythm during symptomatic events. Long-term monitoring using an ICM is recommended in highly symptomatic cases when non-invasive investigations are inconclusive, due to either infrequent events or the inability to complete a diagnostic protocol. For adults with syncope, ICM provides the most cost-effective method for establishing a diagnosis and are considered the method of choice when arrhythmogenic syncope is suspected but not proven. For bradyarrhythmias, ICM may be useful in both documentation of the bradycardia and correlation with clinical symptoms. ICM may also be useful for patients at risk for intermittent AV block in conditions such as Kearns-Sayre syndrome. Finally, ICM may be useful for occult arrhythmia detection in asymptomatic patients with potentially lethal cardiac diseases (primary arrhythmia syndromes, cardiomyopathies) and identify events that warrant need for changes in management.

CIED lead management

Lead management involves the decisions of whether or not to perform CIED lead extraction and assessment of the potential risks and benefits. Consensus statements regarding lead management and extraction were published in 2009[85] and updated in 2017.[86] The following recommendations are complementary to the above guidelines with a perspective focused on pediatrics and patients with CHD. Although major complications during lead extraction are relatively rare (3–4%), significant potential for life-threatening events exists.[87] Therefore, lead extraction should only be performed in centers with an institutional commitment to a comprehensive program. This includes facilities, equipment, personnel, and the ability to manage all complications.[88] A multi-disciplinary team familiar with CHD is vital to maximizing procedural safety and efficacy. There are extensive gaps in knowledge regarding lead management in children and patients with CHD. This includes limited data in the very young and the impact of repeated extractions on vascular integrity and valvular function. There is also absence of data regarding prophylactic lead extractions, as long-term prospective studies on lead abandonment versus extraction in the young do not exist.

COR	**Recommendations for CIED Lead Management*** Thrombosis/Vascular Issues	LOE	References
I	Lead removal is recommended for patients with clinically significant thromboembolic events attributable to thrombus on a lead or a lead fragment that cannot be treated by other means.	C-LD	85,86
I	Lead removal is recommended for patients with superior vena cava stenosis, baffle stenosis, or venous occlusion that prevents implantation of a necessary lead, or when deployment of a stent is planned to avoid entrapment of the lead, or as a part of a comprehensive plan for maintaining patency.	C-LD	86
IIa	Lead removal can be useful for patients with ipsilateral venous occlusion to allow transvenous access to the heart for required placement of an additional or replacement lead.	C-LD	
	Lead Upgrade or Abandonment		
IIa	Lead removal can be useful for patients with an abandoned lead that interferes with the operation of a CIED system.	C-EO	
IIb	Lead removal may be considered for patients requiring CIED revision, taking into account the number of leads present, patient age, size, venous capacitance, and potential for vascular occlusion.	C-LD	
IIb	Lead removal may be considered for isolated upper extremity venous stenosis or thrombosis without symptoms.	C-EO	

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COR	Recommendations for CIED Lead Management*	LOE	References
	Infectious Issues		
I	Lead removal is indicated for CIED-associated endocarditis, bacteremia without an alternative source (particularly *Staphylococcus aureus*), or bacteremia that persists or recurs despite antimicrobial therapy.	B-NR	[85,86]
I	Pre-lead removal blood cultures and transesophageal echocardiography are recommended for patients with suspected systemic CIED infection to guide antibiotic therapy and assess the potential embolic risk of identified vegetations.	B-NR	
IIb	Lead removal may be considered when there is an isolated superficial CIED pocket infection with serial negative blood cultures and no evidence of endocarditis by transesophageal echocardiography.	C-LD	
	Other Indications		
I	Lead removal is recommended for patients with life-threatening arrhythmias secondary to retained leads.	C-EO	
IIa	Device and/or lead removal can be useful for patients with severe chronic pain at the device or lead insertion site or believed to be secondary to the device, for which there is no acceptable alternative.	C-EO	
IIb	Lead removal may be considered for patients with leads that, due to their design or their failure, pose a potential future threat to patients if left in place.	C-LD	
	Epicardial Leads		
I	Epicardial lead removal is recommended for patients where the lead is shown to be associated with coronary artery compression and evidence of myocardial injury.	C-LD	[89]
I	Complete removal of epicardial lead(s) and patches is recommended for all patients with confirmed infection surrounding the intrathoracic portion of the lead.	C-EO	
IIb	Epicardial lead removal may be considered for patients with leads that are thought to be at risk for causing coronary artery compression, valve impingement, or cardiac strangulation.	C-EO	
IIb	Epicardial lead removal may be considered at the time of epicardial lead replacement in the presence of a damaged or nonfunctional lead, taking into account the procedural risk and benefit.	C-EO	

*Recommendations based on adult lead management guidelines.[85,86]

CIED follow-up and ancillary testing

CIED follow-up includes both in-person evaluation (IPE) and remote interrogation and monitoring (RIM) of pacemakers, ICDs and ICMs. The benefits of routine monitoring are well established and include both prolongation of battery life as well as early detection of CIED malfunctions, arrhythmic issues, and adverse events. At present, there are no consensus guidelines for CIED follow up or ancillary testing in the pediatric population. Therefore, the following recommendations are based on Expert Consensus Statements on CIED monitoring[90,91] with select pediatric-relevant modifications. Additional recommendations regarding ancillary testing in conjunction with IPE are also included.

COR	Recommendations CIED Follow-up Recommendations	LOE	**References**[90,91]
I	In-person evaluation (IPE) and the establishment of remote interrogation and monitoring (RIM) are recommended within 2–4 weeks post CIED implantation.	C-EO	
I	At least one annual IPE of all CIEDs is recommended.	C-EO	
I	RIM is recommended for all patients with a CIED that has been recalled or has an advisory to enable early detection of actionable events and confirm proper device function.	C-EO	
I	RIM of CIEDs is recommended every 3–12 months for pacemakers and 3–6 months for ICDs. Frequency should be increased (every 1–3 months) for CIEDs approaching elective replacement indicators.	C-EO	
I	It is recommended that allied health care professionals possess International Board of Heart Rhythm Examiners certification or equivalent experience if they provide RIM and are involved in patient management decisions.	C-EO	
	CIED Ancillary Testing Recommendations		
I	Evaluation of the intrinsic cardiac rhythm evaluation is recommended during CIED interrogation at the annual IPE.	C-EO	
IIa	A standard 12-lead ECG is reasonable at annual in-person evaluation.	C-EO	
IIa	Two-view chest X-ray is reasonable at the first post-implant IPE and every 1–3 years based on patient-specific considerations.	C-EO	
IIa	An echocardiogram is reasonable for assessment of ventricular function in patients who have >40% ventricular paced rhythm every 1–3 years.	C-LD	[92]

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COR	Recommendations CIED Ancillary Testing Recommendations	LOE	References[90,91]
IIb	Exercise stress testing and ambulatory ECG monitoring may be considered in patients with symptoms suggesting possible device malfunction or to assist with device programming.	C-LD	93

Special considerations

CIEDs and magnetic resonance imaging

COR	Recommendations Magnetic Resonance Imaging	LOE	References
I	MRI in all patients with conditional or nonconditional CIEDs should be performed in the context of a defined institutional protocol.	C-LD	94
IIa	MRI is reasonable in patients with nonconditional transvenous CIEDs if there are no fractured, epicardial, or abandoned leads.	B-NR	94
IIb	MRI may be considered in patients with epicardial or abandoned leads based on an individualized consideration of the risk/benefit ratio.	C-LD	95,96

The 2017 MRI and Radiation Exposure in Patients with CIEDs Consensus Statement provides comprehensive recommendations for individuals with both conditional (Food and Drug Administration approved) and non-conditional *transvenous* devices.[94] However, this document does not make specific recommendations for patients with either abandoned *or* epicardial CIED leads. For patients with *epicardial* CIED leads, as there are no MRI conditional epicardial leads, the system is considered non-conditional, even when used with a conditional device.[95] Regarding *abandoned* leads, in vitro data suggest that epicardial leads generate more heat than transvenous leads; however, small studies of MRIs in patients with both epicardial and transvenous abandoned leads suggest that it can be done safely in the majority of cases.[96] In summary, the data on MRI use in epicardial or abandoned leads are inadequate to provide specific recommendations or absolute contraindications. Acknowledging the sparsity of data, but also the importance of MRI, consideration of the risk/benefit ratio of MRI must be made on a "case by case basis."

CIEDs and sports participation

COR	Recommendations Sports Participation	LOE	References
I	For patients with CIEDs, decisions regarding participation in sports or exercise are primarily based on considerations of the patient's diagnosis and physiology rather than the presence of the device.	C-EO	

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COR	Recommendations Sports Participation	LOE	References
IIa	For patients with pacemakers and ICDs, participation in competitive sports or intense recreational exercise is reasonable after shared decision-making that involves a provider who conveys the estimated risk and also includes coaches, schools, communities, or teams.	C-LD	97
III **No benefit**	ICD placement for the sole purpose of participation in competitive athletics should not be performed.	B-NR	97,98

The safety of sports participation for patients with CIEDs continues to evolve. Initial guidelines recommended against competitive sports participation for patients with pacemakers or ICDs. However, subsequent surveys reported that many patients with pacemakers and ICDs had participated in sports without adverse events. Thus, the International ICD Sports Registry was initiated (2013) and reported in 2017.[97] The registry consisted of 129 patients <21 years old including high school and collegiate athletes. While shocks occurred during sports, there were no deaths, no resuscitated arrests, and no arrhythmia-related injury during sports. In addition, the rate of lead malfunction was similar to previously reported rates in unselected populations.

When counseling patients with CIEDs and families about sports participation, the decision process is ultimately patient specific, including the underlying cardiac disease and heart rhythm, the type and indication for device implant, patient age, and type of athletic activity. Shared decision making, including the patient, family, coach, school, team and other community members, should be utilized to determine the best course of pursuit for individuals with CIEDs and sporting endeavors.

Shared decision-making

COR	Recommendation Shared Decision-Making	LOE	References
I	Shared decision-making between the patient, their family, the provider, and other stakeholders is recommended prior to making care plans. This includes discussion of risks, benefits, alternatives, and expected outcomes for patients requiring CIEDs for their pre- and post-implant care.	B-NR	2

Recommendation specific supportive text

The use of shared decision-making should occur prior to all CIED implantation procedures. Clinicians must estimate and clearly describe the potential benefits and risks for the patient and their family. Some decisions will be relatively straightforward; for example, the decision to implant a permanent pacemaker to treat postoperative surgical complete heart block in a patient who is

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pacemaker dependent will be largely uncontestable. However, other treatment decisions, such as implantation of an ICD for primary prevention of SCD, are more complex and nuanced and include choice of ICD system, device location, and personalized estimation of risk of life-threatening arrhythmia for the particular patient over time.

Knowledge gaps and future research

Critical knowledge gaps exist is several areas.[99] One example is the use of ICDs for the primary prevention of SCD. With reduction in device size and the development of novel lead configurations for implantation in smaller patients, the accurate identification of patients at increased risk remains perplexing. Several other important knowledge gaps include but are not limited to the optimal timing of pacemaker implantation after postoperative AV block, contemporary outcomes of patients with isolated CCAVB who do not undergo pacing, risk factors for pacemaker-induced cardiomyopathy, optimal age and body size for transvenous lead implantation, and safety of MRI with abandoned or epicardial leads.

With continuing technological innovations, future research is needed to develop pediatric-specific criteria for application of these new technologies. These include subcutaneous ICDs, leadless pacemakers, and conduction system pacing. Multicenter prospective registries as well as high-quality retrospective data are necessary to provide real-world evidence for new and existing CIED technologies. Future research should be conducted in collaboration with PACES, other relevant scientific societies, the U.S. Food and Drug Administration, and industry partners for development of pediatric "appropriate" CIEDs and device algorithms to specifically benefit young patients and improve their long-term outcomes.

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Appendix 1. Author relationships with industry

Writing Group Member	Employment	Honoraria/ Speaking/ Consulting	Speakers' Bureau	Research*	Fellowship Support*	Ownership/ Partnership/ Principal/ Majority Stockholder	Stock or Stock Options	Intellectual property/ Royalties	Other
Michael J. Silka (Co-Chair)	University of Southern California, Los Angeles Children's Hospital	None	None	None	None	None	None	None	None
Maully J. Shah (Co-Chair)	University of Pennsylvania, Children's Hospital of Philadelphia	None	None	None	Medtronic: 2	None	None	None	None
Jennifer N. Avari Silva	Washington University School of Medicine, St. Louis Children's Hospital	Cardialen: 1 Abbott: 1	None	NIH: 3 ACC: 1 UN&UP: 1	None	None	SentiAR: 4	SentiAR: 5	None
Seshadri Balaji	Oregon Health & Science University, Doernbecher Children's Hospital	Yor Labs: 0	None	Medtronic: 3	None	None	None	None	None
Cheyenne M. Beach	Yale University School of Medicine, Children's Hospital	None	None	None	None	None	None	None	None
Monica N. Benjamin	Hospital de Pediatría Juan P. Garrahan, Hospital El Cruce, Hospital Británico de Buenos Aires, Instituto Cardiovascular ICBA	None	None	None	None	None	None	None	None
Charles I. Berul	George Washington University, Children's National Hospital	None	None	None	None	None	None	None	None
Bryan Cannon	Mayo Clinic	None	None	None	None	None	None	None	None
Frank Cecchin	New York University, Hassenfeld Children's Hospital	None	None	None	None	None	None	None	None
Mitchell I. Cohen	Inova Children's Hospital	None	None	None	None	None	None	None	None
Aarti S. Dalal	Washington University in St. Louis, St. Louis Children's Hospital	None	None	None	None	None	None	None	None
Brynn E. Dechert	University of Michigan, C.S. Mott Children's Hospital	None	None	None	None	None	None	None	None
Anne Foster	Advocate Children's Heart Institute	None	None	None	None	None	None	None	None
Roman Gebauer	Heart Centre Leipzig, University of Leipzig, Germany	None	None	None	None	None	None	None	None
M. Cecilia Gonzalez Corcia	Bristol Royal Hospital for Children	None	None	None	None	None	None	None	None
Prince J. Kannankeril	Vanderbilt University Medical Center	None	None	NIH grants	None	None	None	None	None

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Peter P. Karpawich	The Children's Hospital of Michigan, University Pediatricians PC	None	None	None	None	None	None	None	None
Jeffery J. Kim	Baylor College of Medicine, Texas Children's Hospital	None	None	Cancer Prevention and Research Institute of Texas Grant	None	None	None	None	None
Mani Ram Krishna	Amrita Institute of Medical Sciences	None	None	None	None	None	None	None	None
Peter Kubuš	Children's Heart Center, Charles University in Prague and Motol University Hospital	None	None	None	None	None	None	None	None
Martin J. LaPage	University of Michigan, C.S. Mott Children's Hospital	None	None	None	None	None	None	None	None
Douglas Y. Mah	Harvard University, Boston Children's Hospital	None	None	None	None	None	None	None	None
Lindsey Malloy-Walton	Children's Mercy Hospital	None	None	None	None	None	None	None	None
Aya Miyazaki	Mt. Fuji Shizuoka Children's Hospital	None	None	None	None	None	None	None	None
Kara S. Motonaga	Stanford University, Lucile Packard Children's Hospital	None	None	None	None	None	None	None	None
Mary C. Niu	University of Utah Health Sciences Center/Primary Children's Hospital	None	None	None	None	None	None	None	None
Melissa Olen	Nicklaus Children's Hospital	None	None	None	None	None	None	None	None
Thomas Paul	Georg-August-University Medical Center	AOP Orphan Pharmaceuticals							
Eric Rosenthal	Evelina London Children's Hospital, Guy's & St Thomas' NHS Trust, St Thomas' Hospital	None	None	None	None	None	None	None	None
Elizabeth V. Saarel	St. Luke's Health System	None	None	None	None	None	None	None	None
Massimo Stefano Silvetti	Bambino Gesù Children's Hospital IRCCS	None	None	None	None	None	None	None	None
Elizabeth A. Stephenson	The Hospital for Sick Children	None	None	None	None	None	None	None	None

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Writing Group Member	Employment	Honoraria/ Speaking/ Consulting	Speakers' Bureau	Research*	Fellowship Support*	Ownership/ Partnership/ Principal/ Majority Stockholder	Stock or Stock Options	Intellectual property/ Royalties	Other
Reina B. Tan	New York University Langone Health, Hassenfeld Children's Hospital	None	None	None	None	None	None	None	None
John Triedman	Harvard Medical School, Boston Children's Hospital	Biosense Webster, SentiAR	None	None	None	None	None	None	None
Nicholas H. Von Bergen	The University of Wisconsin-Madison	None	None	None	None	Atrility Medical: 5	Atrility Medical: 1	None	None
Philip L. Wackel	Mayo Clinic	None	None	None	None	None	None	None	None

Number value: **0** = $0; **1** = ≤ $10,000; **2** = > $10,000 to ≤ $25,000; **3** = > $25,000 to ≤ $50,000; **4** = > $50,000 to ≤ $100,000; **5** = >$100,000.
*Research and fellowship support are classed as programmatic support. Sources of programmatic support are disclosed but are not regarded as a relevant relationship with industry for writing group members or reviewers.

Appendix 2. Reviewer relationships with industry

Peer Reviewer	Representation	Employment	Honoraria/ Speaking/ Consulting	Speakers' Bureau	Research*	Fellowship Support*	Ownership/ Partnership/ Principal/Majority Stockholder	Stock or Stock Options	Intellectual/ Royalties	Other
Philip M. Chang	ACC	University of Florida Health/Shands Children's Hospital	None	None	None	None	None	None	None	None
Fabrizio Drago	AEPC	Bambino Gesù Children's Hospital IRCCS	None	None	None	None	None	None	None	None
Anne M. Dubin	PACES	Stanford University, Lucile Packard Children's Hospital	None	None	None	None	None	None	UptoDate royalties: 1	None
Susan P. Etheridge	AHA	University of Utah Health Sciences Center/Primary Children's Hospital	None	None	None	None	None	None	None	None
Apichai Kongpatanayothin	APHRS	Bangkok General Hospital	None	None	None	None	None	None	None	None
Jose M. Moltedo	LAHRS	Sanatorio Finochietto	Abbott/ Biomarkers: 1	None	None	None	None	None	None	None
Ashish A. Nabar	IHRS	Lilavati Hospital, Jupiter Hospital	None	None	None	None	None	None	None	None
George F. Van Hare	HRS	Washington University in St. Louis, St. Louis Children's Hospital	None	None	None	None	None	None	None	None

Number value: **0** = $0; **1** = ≤$10,000; **2** = >$10,000 to ≤$25,000; **3** = >$25,000 to ≤$50,000; **4** = >$50,000 to ≤$100,000; **5** = >$100,000.
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