



2021 PACES expert consensus statement on the indications and management of cardiovascular implantable electronic devices in pediatric patients^{☆, ☆ ☆}



Writing Committee Members⁶, Maully J. Shah^{a, *, 5}, Michael J. Silka^{b, **, 5}, Jennifer N. Avari Silva^{c, 1}, Seshadri Balaji^d, Cheyenne M. Beach^e, Monica N. Benjamin^f, Charles I. Berul^{g, 2}, Bryan Cannon^h, Frank Cecchinⁱ, Mitchell I. Cohen^j, Aarti S. Dalal^c, Brynn E. Dechert^k, Anne Foster^l, Roman Gebauer^{m, 3}, M. Cecilia Gonzalez Corciaⁿ, Prince J. Kannankeril^o, Peter P. Karpawich^p, Jeffery J. Kim^q, Mani Ram Krishna^r, Peter Kubuš^s, Martin J. LaPage^k, Douglas Y. Mah^t, Lindsey Malloy-Walton^u, Aya Miyazaki^v, Kara S. Motonaga^w, Mary C. Niu^x, Melissa Olen^y, Thomas Paul^z, Eric Rosenthal^{aa}, Elizabeth V. Saarel^{ab}, Massimo Stefano Silvetti^{ac}, Elizabeth A. Stephenson^{ad, 4}, Reina B. Tan^{ae}, John Triedman^{t, 1}, Nicholas H. Von Bergen^{af}, Philip L. Wackel^h, Document Reviewers

^a University of Pennsylvania Perelman School of Medicine, Philadelphia, PA, USA

^b University of Southern California Keck School of Medicine, Los Angeles, CA, USA

^c Washington University in St. Louis, St. Louis, Missouri, USA

^d Oregon Health & Science University, Portland, OR, USA

^e Yale University School of Medicine, New Haven, CT, USA

^f Hospital de Pediatría Juan P. Garrahan, Hospital El Cruce, Hospital Británico de Buenos Aires, Instituto Cardiovascular ICBA, Buenos Aires, Argentina

^g George Washington University, Washington, DC, USA

^h Mayo Clinic, Rochester, MN, USA

ⁱ New York University Grossman School of Medicine, New York, NY, USA

^j Inova Children's Hospital, Fairfax, VA, USA

^k University of Michigan, Ann Arbor, MI, USA

^l Advocate Children's Heart Institute, Chicago, IL, USA

^m Heart Centre Leipzig, University of Leipzig, Leipzig, Germany

ⁿ Bristol Royal Hospital for Children, Bristol, United Kingdom

^o Vanderbilt University Medical Center, Nashville, TN, USA

^p University Pediatricians, Children's Hospital of Michigan, Detroit, MI, USA

^q Baylor College of Medicine, Houston, TX, USA

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; LVEF, left ventricular ejection fraction; LMIC, low- and middle-income countries; LOE, level of evidence; LQTS, long QT syndrome; MRI, magnetic resonance imaging; NIDCM, nonischemic dilated cardiomyopathy; RCT, randomized clinical trial; RIM, remote interrogation and monitoring; SCA, sudden cardiac arrest; SCD, sudden cardiac death; SND, sinus node dysfunction; TTM, transtelephonic monitoring; VF, ventricular fibrillation; VT, ventricular tachycardia.

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^{*} Corresponding author. Cardiac Center, Children's Hospital of Philadelphia, 3401 Civic Center Blvd, Philadelphia, PA, 19104, USA.

^{**} Corresponding author. Division of Pediatric Cardiology, Children's Hospital Los Angeles, 4650 Sunset Blvd, Los Angeles, CA, 90027, USA.

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^r Amrita Institute of Medical Sciences, Kochi, India^s Children's Heart Center, Charles University in Prague and Motol University Hospital, Prague, Czech Republic^t Harvard Medical School, Boston, MA, USA^u University of Missouri-Kansas City School of Medicine, Kansas City, MO, USA^v Shizuoka General Hospital and Mt. Fuji Shizuoka Children's Hospital, Shizuoka, Japan^w Stanford University, Palo Alto, CA, USA^x University of Utah Health Sciences Center, Salt Lake City, UT, USA^y Nicklaus Children's Hospital, Miami, FL, USA^z Georg-August-University Medical Center, Göttingen, Germany^{aa} Evelina London Children's Hospital and St Thomas' Hospital, Guy's & St Thomas' NHS Foundation Trust, London, United Kingdom^{ab} St. Luke's Health System, Boise, ID, USA^{ac} Bambino Gesù Children's Hospital IRCCS, Rome, Italy^{ad} The Hospital for Sick Children, Toronto, Canada^{ae} New York University Langone Health, New York, NY, USA^{af} University of Wisconsin School of Medicine and Public Health, Madison, WI, USA

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ABSTRACT

In view of the increasing complexity of both cardiovascular implantable electronic devices (CIEDs) and patients in the current era, practice guidelines, by necessity, have become increasingly specific. This document is an expert consensus statement that has been developed to update and further delineate indications and management of CIEDs in pediatric patients, defined as ≤ 21 years of age, and is intended to focus primarily on the indications for CIEDs in the setting of specific disease categories. The document also highlights variations between previously published adult and pediatric CIED recommendations and provides rationale for underlying important differences. The document addresses some of the deterrents to CIED access in low- and middle-income countries and strategies to circumvent them. The document sections were divided up and drafted by the writing committee members according to their expertise. The recommendations represent the consensus opinion of the entire writing committee, graded by class of recommendation and level of evidence. Several questions addressed in this document either do not lend themselves to clinical trials or are rare disease entities, and in these instances recommendations are based on consensus expert opinion. Furthermore, specific recommendations, even when supported by substantial data, do not replace the need for clinical judgment and patient-specific decision-making. The recommendations were opened for public comment to Pediatric and Congenital Electrophysiology Society (PACES) members and underwent external review by the scientific and clinical document committee of the Heart Rhythm Society (HRS), the science advisory and coordinating committee of the American Heart Association (AHA), the American College of Cardiology (ACC), and the Association for European Paediatric and Congenital Cardiology (AEPC). The document received endorsement by all the collaborators and the Asia Pacific Heart Rhythm Society (APHRS), the Indian Heart Rhythm Society (IHRS), and the Latin American Heart Rhythm Society (LAHRS). This document is expected to provide support for clinicians and patients to allow for appropriate CIED use, appropriate CIED management, and appropriate CIED follow-up in pediatric patients.

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¹ Representative of the Heart Rhythm Society (HRS).² Representative of the American College of Cardiology (ACC).³ Representative of the Association for European Paediatric and Congenital Cardiology (AEPC).⁴ Representative of the American Heart Association (AHA).⁵ Maully J. Shah, MBBS, FHRS, FACC, and Michael J. Silka, MD, FACC, FAHA, are co-first authors.⁶ Writing committee members are required to recuse themselves from voting on sections to which their specific relationships with industry may apply; see [Appendix 1](#) for detailed information.

Shared decision-making
Sick sinus syndrome
Sports and physical activity
Sudden cardiac arrest
Sudden cardiac death
Syncope
Transvenous
Ventricular fibrillation
Ventricular tachycardia

Preamble

Guidelines for the implantation of cardiovascular implantable electronic devices (CIEDs) have evolved since the initial American College of Cardiology (ACC)/American Heart Association (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 as well as other physiologic parameters. In view of the increasing complexity of both devices and patients in the current era, practice guidelines, by necessity, have become increasingly specific. One aspect of this evolution is the “2018 ACC/AHA/HRS Guideline on the Evaluation and Management of Patients With Bradycardia and Cardiac Conduction Delay,” [2] which included specific recommendations for patients >18 years of age. This age-specific threshold was established in view of the need for differing indications for CIEDs as well as size-specific technology factors in younger patients. Therefore, this document has been developed to update and further delineate indications for the use and management of CIEDs in pediatric patients, defined as ≤21 years of age, in recognition that there is often overlap in the care of patients between 18 and 21 years of age.

This document is an expert consensus statement intended to focus primarily on the indications for CIEDs in the setting of specific disease/diagnostic categories. This consensus statement will also provide guidance regarding the management of CIEDs for rhythm disorders in pediatric patients and address some of the deterrents to CIED access in low- and middle-income countries and strategies to circumvent them.

Recommendations are presented in a modular or knowledge chunk format, in which each section includes a table of recommendations, a brief synopsis, and recommendation-specific supportive text [3]. However, this document is not intended to provide an exhaustive review of all aspects of pacemakers, ICDs, and insertable cardiac monitors (ICMs), as this information is easily accessible in electronic searches or textbooks. Furthermore, specific recommendations, such as heart rate criteria for pacemaker implantation, even when supported by substantial data, do not replace the need for clinical judgment and patient-specific decision-making. As a final introductory comment, to avoid clinical overlap, the indications and management of cardiac resynchronization therapy and physiological pacing will be addressed in the anticipated “2022 HRS Expert Consensus Statement on Cardiac Physiological Pacing for the Avoidance and Mitigation of Heart Failure,” which will include a specific section on pediatric and congenital heart disease (CHD).

1. Introduction

1.1. Methodology and evidence review

The principles in the development of this document are 1) new recommendations and any changes to previous recommendations are based on data, when possible; 2) these recommendations are consistent with current ACC/AHA/Heart Rhythm Society (HRS) guidelines when reasonable; [2–19] and 3) all recommendations are critically reviewed, initially by the writing committee and editors,

followed by the Pediatric and Congenital Electrophysiology Society (PACES) executive committee, and subsequently by external HRS, ACC, AHA, and Association for European Paediatric and Congenital Cardiology (AEPC) representatives. Any revisions or additions to existing recommendations will require approval of at least 80% by the members of the PACES writing committee. Specific prior guidelines and consensus statements relevant to CIEDs that have been referenced as the basis for recommendations in this document are acknowledged below and recognized in the specific sections (Table 1).

These recommendations have been developed consistent with standard guideline methodology, i.e., with both a class of recommendation (COR) and a level of evidence (LOE) (Table 2) [4]. 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 scientific evidence supporting the intervention on the basis of the type, quantity, and consistency of data from clinical trials and other sources. Due to the lack of randomized clinical trials (RCTs) in pediatric patients, these LOE recommendations will be limited to class B-NR (limited populations), class C-LD (very limited populations), or C-EO (consensus expert opinion, case studies, or standard of care). It is important to emphasize that 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 [5]. However, there

Table 1
Guidelines, expert consensus statements, and reports cited.

Title	Organization	Year (reference)
Guidelines for permanent pacemaker implantation	ACC/AHA	1984 (1)
Guidelines for the management of patients with bradycardia and cardiac conduction delay	ACC/AHA/HRS	2019 (2)
Report: Innovations, modifications, and evolution of clinical practice guidelines	ACC/AHA	2019 (3)
Report: Evolution of the clinical practice guideline recommendation classification system	ACC/AHA	2016 (4)
ECS: ICD therapy in patients who are not included or not well represented in clinical trials	HRS/ACC/AHA	2014 (5)
Guidelines for device-based therapy	ACC/AHA/HRS	2008 (6)
Update of the 2008 device-based therapy guidelines	ACC/AHA/HRS	2012 (7)
ECS: Arrhythmias in congenital heart disease	EHRA/AEPC/ESC	2018 (8)
ECS: Recognition and management of arrhythmias in adult congenital heart disease	PACES/HRS	2014 (9)
Guidelines on cardiac pacing and resynchronization	ESC	2013 (10)
Guidelines for the evaluation and management of patients with syncope	ACC/AHA/HRS	2017 (11)
Guidelines for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death	AHA/ACC/HRS	2018 (12)
Guidelines for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death	ESC	2015 (13)
ECS: The diagnosis and management of patients with inherited primary arrhythmia syndromes	HRS/EHRA/APHS	2013 (14)
Guideline for the diagnosis and treatment of patients with hypertrophic cardiomyopathy	AHA/ACC	2020 (15)
ECS: The evaluation, risk stratification, and management of arrhythmogenic cardiomyopathy	HRS	2019 (16)
Guidelines for the management for pediatric heart failure	HRS	2015 (17)
ECS: CIED lead management and extraction	HRS	2017 (18)
ECS: MRI and radiation exposure in patients with CIEDs	HRS	2018 (19)

ECS = expert consensus statements; EHRA = European Heart Rhythm Association; ESC = European Society of Cardiology.

Table 2
Class of recommendation and level of evidence Categories*.

Class I	Class IIa	Class IIb	Class III
Benefit >>> Risk	Benefit >> Risk	Benefit ≥ Risk	Risk ≥ Benefit
Procedure/ treatment SHOULD be performed/ is recommended	IT IS REASONABLE to perform the procedure/ treatment	Procedure/treatment MAY BE CONSIDERED/ effectiveness is uncertain	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

may be unequivocal expert consensus that a particular intervention is either effective or necessary. The final evidence tables for the recommendations are included in [Supplemental Appendix 3](#) and summarize the evidence used by the writing committee to formulate these recommendations. References selected and published in this document are intended to be representative and not all-inclusive. Variations between previously published adult and pediatric CIED recommendations as well as new pediatric-specific recommendations are listed in [Supplemental Appendix 4](#).

1.2. 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 AEP. 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.

1.3. 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 AEP 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.

1.4. 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” [6] and “2012 ACCF/AHA/HRS Focused Update of the 2008 Guidelines for Device-Based Therapy of Cardiac Rhythm Abnormalities.” [7] This document is expected to provide support for clinicians and patients to allow for appropriate device use, appropriate device management, follow-up, and appropriate reimbursement in pediatric patients.

1.5. 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 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 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) 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.

2. Permanent pacemakers

2.1. Introduction

The most common indications for permanent pacemaker implantation in children, adolescents, and patients with CHD may be classified as 1) symptomatic sinus bradycardia, 2) advanced second- or third-degree AV block, either congenital or acquired, and 3) pacing for the prevention or termination of tachyarrhythmias [6]. In general, many of the indications for pacemaker implantation in children and adolescents (defined as <19 years of age) are similar to those in adults [2]. However, there are several important differences in infants and children. These patients have faster heart rates, and therefore standards for what is considered normal are age-dependent variables; whereas a heart rate of 45 bpm may be a 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 [8,9]. 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 pacemaker lead

placement and use of device technology in innovative ways often need to be considered to provide pacing in the youngest patients [20–22]. Any pacemaker system used in a young patient may need to be utilized for multiple decades, and consideration of the long-term consequences from device and lead failure plays a role in implantation of pediatric devices.

Bradycardia and associated symptoms in children are often transient (e.g., breath-holding spells) and therefore may not require permanent pacemaker therapy. Conversely, there are other conduction system disorders that may rapidly progress (e.g., neuromuscular disorders) that may require prophylactic pacemaker implantation for disease-specific indications. In addition, as risk factors for cardiac conditions such as the channelopathies are better defined, the indications for device placement in these patients may evolve rapidly.

The goal of this section is to provide an update regarding the indications for permanent pacemaker implantation in pediatric patients. A summary of the recent literature is provided as a framework for clinicians to make individual decisions about pacing in these patients. As the pediatric and CHD populations represent unique groups of patients, clinical judgment and patient-specific decision-making are of the highest importance.

2.2. 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	[23–26]
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	[27,28]
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	

2.2.1. Recommendation-specific supportive text

SND refers to physiologically inappropriate atrial rates, due to either 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. Establishing a temporal correlation between symptoms and age-related bradycardia is of paramount importance when determining whether permanent pacing is needed.

Nonrandomized studies in both children and adults have demonstrated that pacing can provide symptomatic improvement when symptoms, particularly syncope and pre-syncope, are clearly attributable to SND [23–26]. However, there is no clear evidence that pacing in the setting of isolated SND without symptoms improves outcomes.

In symptomatic patients with SND, atrial-based pacing is generally recommended over single-chamber ventricular pacing

[2,28]. Furthermore, the decisions regarding pacemaker implantation for SND in patients with CHD or channelopathies should be made on an individualized basis and are discussed further in the corresponding sections [29].

2.3. Isolated congenital complete

2.3.1. Atrioventricular Block

COR	Recommendations	LOE	References
Isolated Congenital Complete Atrioventricular Block			
I	Permanent pacemaker implantation is indicated for patients with CCAVB with symptomatic bradycardia.	B-NR	[30–33]
I	Permanent pacemaker implantation is indicated for patients with CCAVB with a wide QRS escape rhythm, complex ventricular ectopy, or ventricular dysfunction.	B-NR	[34–36]
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	[30,34,37]
Ila	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	[36,38,39]
Ila	Permanent pacemaker implantation is reasonable for CCAVB with left ventricular dilation (z score ≥ 3) associated with significant mitral insufficiency or systolic dysfunction.	C-LD	[40,41]
Ilb	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	[2,33]

2.3.2. Recommendation-specific supportive text

Although the average ventricular rate in newborns (≤ 30 days old) and infants (≤ 12 months old) with isolated 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 weight (size), ventricular dysfunction, and other comorbidities [42]. Furthermore, although symptoms such as poor feeding or tachypnea in the neonate may be due to multiple causes, they may be indicative of low cardiac output secondary to bradycardia. Therefore, a lower limit heart rate of 50 bpm is recommended for pacemaker implantation when overt symptoms related to low cardiac output do not appear to be present. One additional point of emphasis is that use of heart rate criteria for newborn or infant pacing should be based on heart rate consistency rather than a single measurement in time [34,37].

Beyond the first year of life, permanent pacemaker implantation is generally indicated in symptomatic patients. Contemporary studies suggest that approximately 66% of neonates and infants diagnosed with isolated CCAVB will undergo pacemaker implantation during their first year of life and that 90% of patients with CCAVB will undergo pacemaker implantation by 20 years of age [30]. Long-term natural history studies have demonstrated progressive left ventricular dysfunction and mitral insufficiency with cardiovascular mortality in the fourth or fifth decade of life in patients with CCAVB

who did not undergo pacemaker implantation [33,34,43]. On the other hand, some patients with CCAVB will develop left ventricular cardiomyopathy despite pacing due to either antibody-mediated myocarditis or pacing-induced dyssynchrony [43,44].

2.4. Atrioventricular block: Other Considerations

COR	Recommendations	LOE	References
Atrioventricular Block: Other Considerations			
I	Permanent pacemaker implantation is indicated in patients with clinically significant VT that is pause dependent or associated with severe bradycardia; ICD implantation may be considered as a reasonable alternative.	C-LD	[45,46]
I	Permanent pacing is indicated in symptomatic patients with idiopathic advanced second- or third-degree AV block not attributable to reversible causes.	C-LD	[2,6,7]
Ila	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	[47,48]
Ilb	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	[49]
III	Harm asymptomatic first-degree AV block or asymptomatic second-degree Mobitz type I.	C-LD	[2,7]

2.4.1. Recommendation-specific supportive text

The diagnosis of advanced AV block during late childhood or adolescence is an uncommon but well-recognized phenomena. Advanced AV block may be congenital, may be related to infiltrative diseases, or may remain idiopathic. At times, late-onset AV block may be paroxysmal and quite difficult to document [49].

Exercise stress testing can be useful to detect the site and significance of AV block. Generally, supra-His block resolves with exercise by increased sympathetic tone. When second- and third-degree AV block are observed during exercise, conduction disturbance within the His-Purkinje system is suspected. Although progression to advanced second- and third-degree AV block during exercise is rare, it is associated with a poor prognosis in the absence of a pacemaker [47,48].

With the exception of infiltrative or inflammatory causes of advanced AV block, the criteria for pacemaker implantation are similar to those for CCAVB. Permanent pacemaker implantation may be considered for advanced idiopathic AV block in adolescents with an acceptable ventricular rate, a narrow QRS complex, and normal ventricular function, based on an individualized consideration of symptoms and the risk/benefit ratio.

2.5. Postoperative Atrioventricular Block

COR	Recommendations	LOE	References
Postoperative Atrioventricular Block			
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	[50–53]
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	[52,54,55]
Ilb	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	[54,55]
Ilb	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	
Ilb	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	

2.5.1. 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 [56–58]. A very poor prognosis has been established for CHD patients with permanent postoperative AV block who do not receive permanent pacemakers [54,55]. Among patients who do regain AV conduction following a period of transient AV block, ≥85% have recovery of AV conduction by postoperative day 7 and ≥95% AV conduction by postoperative day 10 [50,51]. Although patients who spontaneously regain AV conduction have a favorable prognosis [7], there is a small but definite risk of late-onset complete AV block in transient postoperative AV block patients, with onset occurring as early as months, to as late as decades, following surgery [52,54,55]. Limited data suggest that some patients with a history of transient postoperative advanced second- or third-degree AV block may be at risk for late-onset AV block or sudden cardiac death (SCD) if they have postoperative bifascicular block on the electrocardiogram (ECG) that was not present preoperatively [54,55]. Permanent pacemaker implantation may also be considered for transient postoperative third-degree AV block that reverts to intact AV node conduction when there is concern about the late development of AV block in patients with forms of CHD associated with progressive conduction abnormalities such as discordant AV connections, AV septal defects, and heterotaxy syndromes [59,60].

2.6. Congenital heart disease: specific considerations

COR	Recommendations	LOE	References
Congenital Heart Disease <i>All the recommendations in children with a structurally normal heart apply, but in addition:</i>			
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	[42,61,62]
Ila	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	[63–67]
Ila	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	[63,68]
Ila	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	[63,69]
Ila	Permanent pacemaker implantation is reasonable for sinus or junctional bradycardia with complex CHD [70] when the mean awake resting heart rate is < 40 bpm or when there are prolonged pauses in the ventricular rate.	C-EO	
Ilb	Permanent pacing may be considered for sinus or junctional bradycardia with simple or moderate CHD [70] 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	[71–73]

2.6.1. Recommendation-specific supportive text

Patients with CHD often have important structural and functional lesions [70], which influence both the indications for pacing as well as the type of pacing lead(s) utilized [42]. Therefore, pacemaker implantation in these patients should not be viewed as an isolated procedure. The loss of vascular access or direct access to cardiac chambers and/or persistent right-to-left shunting require

utilization of epicardial pacing leads (with concomitant sternotomy or thoracotomy) [74], although novel hybrid approaches to lead placement are being developed [75,76].

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 [67,68]. Permanent pacemaker and/or lead implantation may be considered prophylactically in patients with evidence of conduction disease and heart defects with a known natural progression to advanced heart block (e.g., discordant AV connections, heterotaxy syndrome) at the time of cardiac surgery [59,60,77].

Similarly, in single-ventricle patients undergoing Fontan conversion, prophylactic antitachycardia pacemakers have been used [67]. There may be a role for pacing in improving the hemodynamic status in patients with plastic bronchitis and protein-losing enteropathy without conventional pacing indications [78].

The decisions regarding pacemaker implantation should also consider the complexity of the patient's anatomy and hemodynamic status, with complex defined as patients with palliative repairs or impaired ventricular function or circulatory physiology [70].

2.7. Post Cardiac Transplantation

2.7.1. Recommendation-specific supportive text

COR	Recommendations	LOE	References
Post Cardiac Transplantation			
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	[6,79–82]
Ila	Permanent pacing is reasonable for marked chronotropic incompetence impairing the quality of life late in the post-transplant period.	C-LD	[79–82]
Ilb	Permanent pacing may be considered when relative bradycardia is prolonged, recurrent, or limits rehabilitation or discharge after postoperative recovery from cardiac transplantation.	C-LD	[6,81,83]
Ilb	Permanent pacing may be considered for any degree of AV block considered to be due to graft vasculopathy.	C-LD	[79,84]

Transient sinus bradycardia is relatively common immediately after transplantation and frequently resolves spontaneously. In rare cases, sinus bradycardia may persist and pacemaker implantation may be needed, but at least a week should be allowed for spontaneous recovery of sinus node function. Early post-transplant AV block has been reported in pediatric patients to be more frequent than in the adult population and may be related to donor age [79,80]. An analysis of the United Network for Organ Sharing (UNOS) database reported that between 1994 and 2014, 1% of cardiac transplant patients <18 years of age required a pacemaker in the acute post-transplant interval. Factors associated with need for pacemaker implant were biatrial anastomosis, older donor age, and antiarrhythmic drug use [80].

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 or development of transplant coronary artery disease, as late-onset bradycardia may be the first manifestation [79,84]. Microvascular angiopathy that may not be seen during conventional angiography may also cause significant ventricular dysfunction and subsequent graft failure with an added risk for conduction abnormalities [85].

The role of prophylactic ICD implantation is not well established but may be considered in patients who require pacemakers. Risk factors to consider are coronary artery vasculopathy and left ventricular dysfunction, which may present as ventricular arrhythmias and have been associated with SCD [86,87].

2.8. Neuromuscular diseases and other

2.8.1. Progressive cardiac conduction diseases

COR	Recommendations	LOE	References
Neuromuscular Diseases and Other Progressive Cardiac Conduction Diseases			
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	[2,88–95]
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	[2,95–98]
Ila	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	[2,88,90,91]
Ila	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	[2,94,99]
Ilb	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	[2,88,90,91,100]

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.

2.8.2. Recommendation-specific supportive text

Progressive cardiac conduction diseases often involve genetic disorders with progressive deterioration of the conduction system occurring either in isolation or in conjunction with other cardiac and metabolic diseases including neuromuscular and mitochondrial diseases.

The severity and onset of cardiac complications differ among the diseases. Conduction disturbances are commonly observed in myotonic dystrophy type 1 and Emery-Dreifuss muscular dystrophy [81]. Variable degrees of conduction abnormalities may occur, ranging from first-degree AV block to complete AV block with unpredictable progression. Laminopathy caused by mutations in the LMNA gene is a wide-spectrum disorder exhibiting peripheral neuropathy, skeletal muscle disorders, progerias, and dilated cardiomyopathy. Cardiac conduction abnormalities, such as sinus bradycardia, AV block, atrial fibrillation, atrial standstill, and ventricular tachycardia (VT), are common and are often observed before the onset of heart failure symptoms [87,92]. In a meta-analysis, arrhythmias were observed in 36% of patients before 20 years of age, with heart failure observed in 10% before 30 years of age [87]. A prolonged PR interval >240 ms in adults is reported to be a predictor of progressive AV block and/or ventricular arrhythmias in patients with myotonic dystrophy and in patients with laminopathy [91,92,94,99].

Among the mitochondrial diseases, patients with Kearns-Sayre

syndrome, characterized by progressive external ophthalmoplegia and myopathy with an onset before the age of 20 years, are known to carry a high risk for AV block and SCD [88–91]. 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.

2.9. Neurocardiogenic syncope

COR	Recommendations	LOE	References
Neurocardiogenic Syncope			
Ila	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	[101–103]
Ilb	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	[104–106]
Ilb	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	[107,108]
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	

2.9.1. Recommendation-specific supportive text

In the vast majority of cases, neurocardiogenic syncope is a limited disease and pacemaker implantation is not required. In some patients, however, 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 [101–104,108]. 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. Because the efficacy of pacing depends on the clinical setting, a clear relationship between symptoms and bradycardia should be established prior to pacemaker implantation. Bradycardia or asystole should be observed during episodes of clinical syncope, ideally on more than one occasion [105]. Event monitors and ICMs have been effective for documenting this relationship.

In pallid breath-holding spells, studies of predominantly infants and toddlers have demonstrated either complete resolution or a significant reduction in the number of syncopal events in 86% patients with pacing [101,102]. Single-chamber pacing with hysteresis appears as effective as dual-chamber pacing with rate drop response for the prevention of syncope and seizures. Pacemaker settings may be optimized to prevent sustained bradycardia by programming a relatively fast pacing rate at the time of the vasovagal reflex to augment cardiac output.

Attributed to vagal storm in the setting of epilepsy, ictal-induced

bradyarrhythmia or asystole can impair both cerebral perfusion and cortical function and contribute to transient loss of consciousness and injury [106,107]. While conventional antiepileptic medications and epilepsy surgery are the mainstay treatments for ictal-induced bradycardia, pacemaker implantation may be reasonable as an adjunct for reducing the severity of symptoms.

2.10. Cardiac channelopathies

COR	Recommendations	LOE	References
Cardiac Channelopathies			
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	[109–111]
IIb	Permanent pacemaker implantation may be considered as adjunctive therapy in patients with long QT syndrome and functional 2:1 AV block.	C- LD	[112]
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	[109,113,114]
III No	Atrial pacing alone is not indicated in patients with complete atrial standstill due to the high benefit potential for noncapture of the myocardium.	C- LD	[115,116]

2.10.1. Recommendation-specific supportive text

The utility of pacing as adjunctive therapy in the various channelopathies is not well defined. Most data are based on observational reports of pacing in the context of long QT syndrome (LQTS). In certain high-risk patients with LQTS, permanent pacemaker implantation may provide a benefit to decrease bradycardia-related or pause-related initiation of ventricular tachyarrhythmias or so-called short-long-short episodes [109–111]. In infants with prolonged QT-related functional 2:1 AV block, one observational study reported that pacing in combination with other therapies resulted in favorable outcomes with no mortality [112]. Additionally, in some patients with LQTS, atrial pacing faster than the intrinsic rate has been shown to shorten the QT interval and reduce the rate of recurrent syncopal events in high-risk LQTS patients [109,114]. When SND and/or AV block are present in the setting of a channelopathy or as the result of antiarrhythmic medications needed for treatment of a channelopathy, the indications for permanent pacing detailed in the respective section on SND and/or AV block apply. In the setting of atrial standstill secondary to a channelopathy or

laminopathy, single-chamber atrial pacemaker placement alone is not recommended due to the high probability of atrial noncapture [115,116].

2.11. Inflammation/infection

COR	Recommendations	LOE	References
Inflammation/Infection			
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	[117,118]
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	[117–120]
III No	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	

2.11.1. Recommendation-specific supportive text

Systemic infections may cause myocardial inflammation or infiltration presenting with bradycardia or complete AV block. Known causes are Lyme disease (*Borrelia burgdorferi*), Chagas disease in individuals from *Trypanosoma cruzi*–endemic areas in Central and South America, and rarely from diphtheria myocarditis. Other etiologies include infectious mononucleosis (Epstein-Barr virus), bacterial endocarditis, viral myocarditis with perivalvular abscess, rheumatic fever, and sarcoidosis.

In symptomatic AV block associated with Lyme disease, approximately 40% of patients may require temporary pacing, although AV block is typically reversible with antibiotic therapy [117,118]. Chronic Chagas disease can present with different degrees of conduction defects. Advanced heart block in Chagas is permanent, and pacemaker implantation is indicated [119,120]. An ICD should be considered in Chagas cardiomyopathy in the presence of significant left ventricular dysfunction or ventricular arrhythmias [120]. More recently, there have been reports of transient AV conduction abnormalities associated with the COVID-19–related multisystem inflammatory syndrome in children (MIS-C) with ventricular dysfunction [121]. Medical-directed therapy for the underlying condition should be maximized (including antibiotic therapy, steroids, intravenous immunoglobulins), and if tolerated, a waiting period of up to several months is warranted prior to pacemaker implantation to provide sufficient opportunity for spontaneous recovery of AV conduction.

Recovery of AV conduction in patients with complete heart block due to acute myocarditis has been reported to occur in 67% of

young patients within 7 days of the onset of AV block [122]. Late monitoring for possible recurrence of symptoms or unrecognized recurrences of AV block or other arrhythmias is advised in these patients.

3. Implantable cardioverter defibrillators

3.1. Introduction

The process of CIED guideline development has evolved over the past few decades, with initial recommendations based on observational clinical experience and refined based on controlled clinical studies and advances in device technology. Although the development of pediatric CIED recommendations has been limited by the lack of RCTs and small patient numbers, pacemaker recommendations have been established based on clearly defined diagnoses and five decades of clinical experience. Conversely, pediatric recommendations for ICD implantation have been primarily based on adult data and, with some modifications, applied to younger patients. Adult ICD guidelines are based on a specific diagnosis as the defined cause or presumed risk factor for a sudden cardiac event, such as ischemia, cardiomyopathy, or genetic cardiovascular disease [6,7,12,13]. In contrast, recent studies of pediatric SCA survivors have continued to demonstrate that in approximately 50% of cases, the cause of the event remains undefined despite an extensive and systematic evaluation [123,124]. Furthermore, in young patients with diagnoses such as catecholaminergic polymorphic ventricular tachycardia (CPVT) or Brugada syndrome, SCA is often the presenting symptom of the disease [125,126]. Therefore, while development of pediatric ICD recommendations based on specific cardiovascular diagnoses would be intuitively preferable, the following discussion of ICD indications will begin with general considerations for the young patient with an unexplained SCA, followed by a more nuanced series of recommendations for ICD implantation when a specific cause of SCA or defined risk factor has been identified. Furthermore, there remain extensive “gaps” in current ICD recommendations, irrespective of age, for many of the diseases associated with SCD in pediatrics [127,128]. 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.

3.2. General Recommendations for Implantable Cardioverter Defibrillator Therapy

COR	Recommendations	LOE	References
General Recommendations for Implantable Cardioverter Defibrillator Therapy			
I	ICD implantation is indicated for survivors of SCA due to VT/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	[6,7,12,13,129–132]
IIb	ICD implantation may be considered for patients with sustained VT that cannot be adequately controlled with medication and/or catheter ablation.	C-EO	
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	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	[133–136]
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	[71–73]

3.2.1. Recommendation-specific supportive text

ICD guidelines specific to pediatrics must consider the unique aspects of device implantation and follow-up in children as well as the pathogenesis of the disease, which may evolve over time. A pediatric cardiologist should be involved in the decision to implant an ICD in pediatric patients, and the procedure should be performed by a cardiologist or cardiothoracic surgeon with special training and/or experience in CIED implantation in the pediatric age-group. ICD implantation should be a shared decision between the patient, family, and physician considering specific pediatric characteristics including age, size of the patient, need for an epicardial device, religious/cultural beliefs, and patient quality of life. This includes the physical as well as the psychological impact of an ICD on the patient's well-being [137]. In addition, all ICD recommendations are based on the premise that meaningful survival of >1 year is expected; meaningful survival means that a patient has a reasonable quality of life and functional status [11]. It is further recommended that the indications for an individual patient's ICD 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 [138].

3.3. ICD indications for cardiac channelopathies

3.3.1. Long QT syndrome

COR	Recommendations	LOE	References
Long QT Syndrome			
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	[12–14,139,140]
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	[12–14,141–147]
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	[148–154]
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	[13,14,139]

3.3.1.1. Recommendation-specific supportive text. Congenital LQTS refers to genetically heterogeneous disorders characterized by the phenotypes of QTc prolongation on the ECG and risk of potentially life-threatening cardiac arrhythmias. Both phenotypic and genotypic characteristics are used to guide risk stratification of patients with LQTS and consideration for ICD [153]. Phenotypic risk factors include the onset of symptoms at age <10 years, prior SCA, or recurrent syncope [143–146,153]. Additional high risk factors include a QTc ≥550 ms regardless of genotype, QTc ≥500 ms with LQT1 genotype, females with LQT2 genotype, and males with LQT3 genotype [141,150].

Patients with rare conditions such as the Jervell and Lange-Nielson syndrome, Timothy syndrome, or calmodulinopathies may be at highest risk for SCA or SCD [150–152]. Infants presenting with bradycardia, functional 2:1 AV block, or cardiac arrest are also at significant risk [155].

Nonselective beta-blockers are considered first-line therapy and can significantly decrease subsequent cardiac events in patients, especially in those with *KCNQ1* mutations [14,140]. In addition, beta-blockers and cardiac sympathetic denervation without ICD may be appropriate alternatives in carefully selected patients [14,142,143].

In highest-risk patients, observational studies support effectiveness of the ICD in preventing SCD, with consideration of left cardiac sympathetic denervation to reduce the frequency of ICD shocks [139,142,143]. However, implantation of an ICD in asymptomatic low-risk patient with LQTS for a positive family history of LQTS-related SCD is not clearly supported by published data, and individual decision-making is important [14].

3.3.2. Catecholaminergic polymorphic ventricular tachycardia

COR	Recommendations	LOE	References
	Catecholaminergic Polymorphic Ventricular Tachycardia		
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	[13,14,126,156–162]
Ila	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	[126]
Ilb	ICD implantation may be considered in CPVT patients with polymorphic/bidirectional VT despite optimal pharmacologic therapy with or without cardiac sympathetic denervation.	C-LD	[157]
III Harm	ICD implantation is not indicated in asymptomatic patients with a diagnosis of CPVT.	C-EO	

3.3.2.1. Recommendation-specific supportive text. CPVT is characterized by exertion-related polymorphic or bidirectional VT and is associated with syncope and SCA. SCA/SCD is reported in 3%–13% of CPVT patients [158]. High risk factors include male sex, previous history of cardiac arrest, multiple genetic variants, and younger age at diagnosis [158,159]. Continued complex ventricular ectopy on exercise testing despite optimal medical therapy is also associated with worse outcome [160]. Studies evaluating CPVT patients with >2 genetic variants suggest that these patients may also be at

higher risk for SCA [159].

Treatment with nonselective beta-blockers is associated with a reduction in adverse cardiac events [13,14,158]. The addition of flecainide to refractory patients in addition to maximally tolerated beta-blocker may suppress ventricular ectopy by as much as 85% [161].

In general, ICD implantation should be reserved for CPVT patients with prior SCA or with arrhythmogenic syncope on combination medical therapy and/or cardiac sympathetic denervation [13,14,126,157]. Inappropriate shocks are reported in 20%–30% of CPVT patients with ICDs [157,163,164]. Device programming in patients with CPVT should be optimized to deliver therapy for ventricular fibrillation (VF) and to minimize inappropriate shocks and the risk of potentially fatal electrical storms [157,164].

Cardiac sympathetic denervation is recommended in patients who continue to have syncope or significant arrhythmias despite optimal medical therapy, are intolerant of medical therapy, or experience recurrent ICD shocks [162]. 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 [14,126].

3.3.3. Brugada Syndrome

COR	Recommendations	LOE	References
	Brugada Syndrome		
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	[12–14,165–171]
Ila	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	[165–169]
Ilb	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	

3.3.3.1. Recommendation-specific supportive text. Brugada syndrome (BrS) is an inherited arrhythmogenic disorder characterized by a coved-type ST-segment elevation in the right precordial ECG leads and an increased risk of SCD [12–14,165]. The phenotypic expression of the disease spans from patients who are completely asymptomatic to those who experience a lethal arrhythmia [165,166]. The syndrome presents typically in the fourth to fifth decade, but in rare cases may have an early onset during childhood [155]. Pediatric cases are rare but can express as a rapidly progressive form and lead to life-threatening arrhythmias [128,166–169].

The placement of an ICD remains the only therapy with proven efficacy for the management of ventricular arrhythmias and prevention of SCD in patients with BrS [170]. Adult recommendations for risk stratification including ventricular stimulation have been established but have not been validated in pediatrics [12–14]. 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 pattern) ECG, atrial arrhythmias and/or SND, and conduction abnormalities (AV block or intraventricular conduction delay) [165]. Although attempts have been made to create a noninvasive risk stratification scoring system [167], such recommendations are based on small cohorts. Patients with a type I ECG pattern and a history of syncope or SCD have a class I indication for an ICD implantation [160]. In this study, 9 of 35 (26%) BrS patients with an ICD implanted at age <20 years received an appropriate therapy during a median follow-up of 7.3 years [160]. Conversely, implantation of an ICD is not indicated in asymptomatic patients in the absence of risk factors. Large multicentric studies are necessary to further characterize risk factors and support primary prevention indications for BrS in pediatric patients.

3.4. ICD indications for cardiomyopathies

3.4.1. Hypertrophic Cardiomyopathy

COR	Recommendations	LOE	References
Hypertrophic Cardiomyopathy			
I	ICD implantation is indicated in patients with HCM who are survivors of SCA or have spontaneous sustained VT.	B-NR	[12,13,15,172–175]
Ila	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	[15,172–179]
Ilb	ICD implantation may be considered in patients with HCM without the above risk factors but with secondary risk factors for SCA such as extensive LGE on cardiac MRI or systolic dysfunction.	B-NR	[15,179–182]
III	ICD implantation is not indicated in patients with Harm an identified HCM genotype in the absence of known pediatric SCA risk factors.	C-LD	[177]

3.4.1.1. Recommendation-specific supportive text. Hypertrophic cardiomyopathy (HCM) is a genetic cardiovascular condition manifested by pathologic left ventricular hypertrophy in the absence of loading conditions. The phenotypic expression of HCM is variable, resulting in a diverse clinical course and highly variable long-term prognosis. Estimates for SCD rates in childhood HCM vary widely, with recent epidemiologic studies that have reported rates of between 1% and 7.2% per year [172,174]. While ICDs have improved the outcomes for patients with HCM resuscitated from SCA, the accurate identification of risk factors for SCD to guide primary prevention ICD implantation remains a challenge, particularly given the potential progression of the disease process over time [172–175]. A multicenter pediatric HCM registry study reported the 5-year risk of SCA was 9% [174]. Primary and secondary prevention ICDs were implanted in 18% and 4% of the cohort, respectively. Only 2.5% of the patients with a primary prevention ICD received an appropriate discharge at 5 years' follow-up, highlighting the major gaps in knowledge for accurate prediction of SCD risk in pediatric HCM patients [174].

Previously published clinical practice guidelines define high risk for SCD in HCM by the presence of ≥1 clinical risk factors based on

primarily adult data [6,7,14]. Recent studies, however, suggest that the significance of the various risk factors may differ in children compared to adults [174–177]. Conventional risk factors include survival from an SCA, spontaneous sustained VT, unexplained syncope, nonsustained VT, family history of early HCM-related SCD, and massive left ventricular hypertrophy [14,173]. While a left ventricular wall thickness ≥30 mm is considered a risk factor in adults, left ventricular hypertrophy is determined relative to age and body size and therefore should be converted to a z score when evaluating this as a risk factor in smaller children [174,177]. A multicenter pediatric study showed that a left ventricular posterior wall thickness z score ≥5 was associated with VT/VF or SCA, while a meta-analysis of pediatric studies reported a maximum left ventricular wall thickness ≥30 mm or a z score ≥6 associated with an increased risk of SCD [175,176].

Other secondary risk factors for SCD, such as late gadolinium enhancement (LGE) on cardiac magnetic resonance imaging (MRI), have been investigated, but the predictive value of LGE for SCD in children is still unclear [179,180]. The evolving 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 [181,183].

3.4.2. Restrictive cardiomyopathy

There are limited data regarding the use of ICDs in patients with restrictive cardiomyopathy [184,185]. The underlying cause of the restrictive cardiomyopathy is most commonly due to abnormalities in the sarcomeric genes, resulting in overlap with the HCM phenotype as well as risk for both tachyarrhythmias and conduction block [185]. Given the overlap with HCM, ICD recommendations for patients with restrictive cardiomyopathy are included under the HCM and general guidelines. However, these patients do require unique consideration as, in comparison to those with HCM, patients with purely restrictive cardiomyopathy may not display the typical risk factors such as thickening of the intraventricular septum but do appear to be at higher risk for SCD, SCA, and cardiac transplant [186,187]. Given this, ICD implantation may be appropriate in patients with a restrictive cardiomyopathy who present with heart failure or unexplained syncope when transplant is not an immediate option [188].

3.4.3. Arrhythmogenic cardiomyopathies

COR	Recommendations	LOE	References
Arrhythmogenic Cardiomyopathies			
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	[12,13,16,189–191]
Ila	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	[192]
Ilb	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	[192,193]

3.4.3.1. Recommendation-specific supportive text. Arrhythmogenic cardiomyopathy (ACM) encompasses a spectrum of disorders of the myocardium with the distinguishing feature of

presentation with sustained arrhythmias [16]. It includes, but is not limited to, genetic disorders such as arrhythmogenic right/left ventricular cardiomyopathy, lamin A/C, filamin-C, phospholamban, and cardiac amyloidosis [16]. Under this definition, infectious processes such as myocarditis and Chagas disease and inflammatory disorders such as sarcoidosis may also be classified. Most of these entities are infrequent before puberty and often overlap with other cardiomyopathies in presentation, particularly dilated cardiomyopathy [16].

The diagnosis of ACM requires a high degree of suspicion. The initial evaluation should include clinical history, physical examination, detailed family history, 12-lead ECG, echocardiography, ambulatory electrocardiography monitoring, exercise testing, and cardiac MRI. Additional testing includes signal-averaged ECG and genetic testing [16,189].

The most frequent form of ACM in the pediatric age-group is arrhythmogenic right ventricular cardiomyopathy (ARVC) [189]. ARVC is characterized by predominant right ventricular involvement with fibro-fatty replacement of the myocardium resulting in conduction abnormalities and ventricular arrhythmias. Biventricular disease is associated with younger age of onset [190,191]. ARVC is either de novo or inherited in an autosomal dominant pattern involving variances in desmosomal genes or desmosome-associated proteins [16,193]. Syncope is reported in 16%–40% of ARVC patients at the time of diagnosis, is frequently exercise related, and has been associated with high arrhythmic risk [16,191]. In adult ARVC cohorts, risk factors for SCD include syncope presumed due to ventricular arrhythmia, sustained or nonsustained VT, and severe right ventricular and/or left ventricular systolic dysfunction [12,16]. Due to the relatively low prevalence of manifest ARVC in the young, there is a paucity of data regarding risk stratification for SCD in pediatric patients with ARVC.

Overall, SCD affects 2%–15% of young patients with ACM [189,191]. Patients presenting with SCD and/or sustained ventricular arrhythmias have a class I indication for an ICD implantation [12,16]. The limited available data on risk stratification in the young hamper the indication for a primary prevention ICD in this population. However, ICD implantation is reasonable in patients with ACM with hemodynamically tolerated sustained VT, syncope presumed due to ventricular arrhythmia, or an LVEF ≤35%. Candidacy and timing of cardiac transplantation and whether a wearable external defibrillator is a reasonable alternative should be taken into consideration on an individual basis for those patients with advanced heart failure [5].

3.4.4. Nonischemic dilated cardiomyopathy

COR	Recommendations	LOE	References
	Nonischemic Dilated Cardiomyopathy		
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	[12,13,16,17,131,132,194]
IIb	ICD implantation may be considered in patients with NIDCM and syncope or an LVEF ≤35%, despite optimal medical therapy.	C-LD	[194–197]

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COR	Recommendations	LOE	References
	Nonischemic Dilated Cardiomyopathy		
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	

3.4.4.1. Recommendation-specific supportive text. The incidence of SCD in pediatric patients with idiopathic/NIDCM is only 1%–5%, which is significantly less than that in adult patients [195,196]. Although studies have shown some ICD survival benefit for secondary prevention in pediatric dilated cardiomyopathy, the low incidence of SCD has made it quite difficult to establish risk factors to guide recommendations for primary prevention ICD implantation [195]. In contrast to some studies of adult patients with NIDCM and LVEF ≤35% [198], there is no clear evidence that ICDs implanted for primary prevention improve survival for pediatric patients with NIDCM [199,200]. However, primary prevention ICDs may be considered for patients with syncope or severe impairment of left ventricular function despite optimal medical therapy (beta-blockers and afterload reduction) and after careful consideration of device-related complication risks, candidacy and timing of cardiac transplantation, and whether a wearable external defibrillator is a reasonable alternative [5,17,194,197].

The phenotype of NIDCM may overlap with other types of pediatric cardiomyopathies resulting in variable risks of SCD. For example, the Sudden Death in Childhood Cardiomyopathy study showed that the risk of SCD varied according to cardiomyopathy phenotype [195]. The cumulative incidence of SCD at 15 years was 5% for idiopathic dilated cardiomyopathy compared to 23% for left ventricular noncompaction. Myocardial dysfunction and/or a history of clinically significant arrhythmias are strongly associated with mortality in left ventricular noncompaction [201,202]. Therefore, factors that may influence the decision regarding implantation of a primary prevention ICD include the underlying etiology of the NIDCM, the cardiomyopathy phenotype, the degree of ventricular dysfunction, and the presence of cardiac arrhythmias [203].

3.5. ICD indications for congenital heart disease

COR Recommendations	LOE	References
Congenital Heart Disease		
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	[9,12,13,129,131,204–206]
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	[131,204,207,208]
IIa ICD implantation is reasonable for CHD patients with systemic LVEF <35% and sustained VT or presumed arrhythmogenic syncope.	C-LD	[9,12,13,204,209,210]
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	[9,210,211]
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	

3.5.1. Recommendation-specific supportive text

The association between CHD and arrhythmias has been well established. First demonstrated in repaired Tetralogy of Fallot, multiple studies since have identified risk factors for VT or SCD including residual cardiac defects, alterations in hemodynamics, and scars from prior interventions/surgeries [204–206]. Correction of residual abnormalities or ablation of arrhythmogenic substrate may improve ventricular function and reduce symptoms. However, this may be inadequate to prevent the risk of subsequent VT or SCA in all but a select group of patients [207,208]. ICD placement may therefore be appropriate in patients with, or at high risk of, potentially life-threatening arrhythmias [9,12,13].

While ICDs are commonly placed for both primary and secondary prevention in patients with CHD, those with CHD appear to have an increased risk of inappropriate shocks compared to those with ICDs and without CHD [130,131,211–213]. Appropriate ICD shock rates of 3%–6% per year have been shown with an increased frequency of appropriate shocks for secondary prevention indications [204]. Antitachycardia pacing has been shown to be effective in VT termination and reducing ICD shocks [214]. Patients with CHD receiving an ICD have an increased rate of complications as high as 26%–45%, as well a high rate of inappropriate shocks [130,131,212,213]. The role of programmed stimulation and presence and degree of ventricular dysfunction as risk factors for SCD in CHD and thus primary prevention ICDs continues to be debated [215–217]. ICD implantation can be especially challenging in patients with CHD due to anatomic complexity, intracardiac shunts, or limited vascular access. This may require nonstandard approaches such as epicardial leads, nontransvenous defibrillation coils or a subcutaneous ICD [218,219].

4. Insertable cardiac monitors

Syncope and palpitations are common symptoms in children and adolescents. ICMs (also referred to as implantable loop recorders) are subcutaneously implanted devices that provide long-term rhythm surveillance and documentation during a patient's symptomatic event. Rhythm tracings during events are either patient-triggered recordings or stored automatically by predefined criteria. Long-term ECG monitoring using an ICM is recommended in symptomatic cases when the personal history, physical examination, and noninvasive investigations have been inconclusive, especially due to the low frequency of clinical events and/or limited feasibility of a complete diagnostic protocol [220–224]. A remote monitoring program with immediate wireless data transfer capability and daily diagnostic data availability has overcome the prior problem of limited device storage capacity and has facilitated early diagnosis. ICMs, along with Holter monitoring, external loop

recorders, and remote at-home telemetry, are reported to provide a diagnostic yield of 43%–50% at 2 years and 80% at 4 years [221–226].

COR Recommendations	LOE	References
Insertable Cardiac Monitors		
I Noninvasive cardiac rhythm monitoring is indicated in all patients prior to placement of an ICM.	B-NR	[11,220–223]
I ICM is indicated in syncope 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	[8,220,225–229]
IIa ICM is reasonable in the evaluation of patients with recurrent syncope of uncertain origin but not a high risk of SCD.	B-NR	[8,11,223–234]
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	[2]
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	[12,226,227]
IIb ICM may be considered in patients with suspected reflex syncope presenting with frequent or severe syncope episodes.	C-LD	[8,230,231]
IIb ICM may be considered in carefully selected patients with suspected epilepsy in whom anticonvulsive treatment has proven ineffective.	C-LD	[235]
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	[223,224,231–233]
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	

4.1. Recommendation-specific supportive text

Several observational studies have demonstrated a benefit of ICM in establishing a diagnosis for recurrent symptoms of unclear etiology when other monitoring methods have failed to document an underlying cause.

Syncope: Cardiac or undefined syncope may be present in up to 8% of syncope events in children and adolescents [8]. In adults, monitoring with an ICM has been shown to be more cost-effective for establishing a diagnosis than other methods of rhythm

monitoring and should be the method of choice when arrhythmogenic syncope is suspected but not proven [223,226].

Palpitations: ICM implantation should be considered on an individual basis, taking into account each patient's underlying cardiac condition, the severity of symptoms, and age- and development-related monitoring limitations [233,234].

Bradyarrhythmias: ICM may be useful in the monitoring of bradyarrhythmias and their correlation with clinical symptoms. ICM may also be useful for patients at risk for intermittent or progressive AV block including patients with neuromuscular diseases, progressive cardiac conduction diseases, and Kearns-Sayre syndrome [91,98].

Other conditions: ICM may be useful for occult arrhythmia detection in asymptomatic children with potentially lethal cardiac diseases (e.g., inherited primary arrhythmia syndromes, cardiomyopathies) and may identify events that warrant changes in patient management [226,230–232]. Furthermore, monitoring with an ICM may provide psychological reassurance for parents of children at risk for malignant arrhythmias [233].

5. CIED lead management

Lead management remains a vitally important issue in children, both with and without CHD. Updated consensus statements regarding lead management and extraction were put forth in 2017 [18] and 2018 [236]. The following recommendations are complementary to these existing guidelines with a nuanced perspective focusing on pediatrics and patients with CHD.

The definitions used related to lead management in this document are similar to those explained in the 2017 statement [18]. The general category of “lead removal” includes “lead explant” that is performed using a simple traction technique and “lead extraction” that refers to removal of a lead that has been implanted for >1 year or requiring the assistance of specialized equipment regardless of implant duration [18]. The most common indications for transvenous lead extraction in children remain lead failure (76%) and venous occlusion [237–243]. Pediatric patients are more likely to outlive the functionality of their leads, amplifying the importance of lead durability, longevity of venous access, and long-term risks of lead dysfunction. Coupled with studies in children indicating that older lead age is an independent predictor of need for advanced extraction techniques and added complexity, greater emphasis should be given to the potential risks of lead abandonment in this population [240,241].

Available extraction tools in children are similar to those in the adult population, as there are no special tools designed specifically for children or patients with CHD. These include locking stylets, telescoping sheaths, femoral snares, and mechanical, laser, or radiofrequency-powered sheaths [243,244]. Extractors should be

appropriately trained, and the entire team must have working knowledge of these tools and techniques. Additionally, expertise in pediatrics, CHD, and surgically corrected anatomy is mandatory, as the methods and potential complications may be specific to both size and anatomy. Unusual lead position and foreign material such as prosthetic valves, conduits, and baffles may necessitate adjustments in approach [242]. Younger patients are also more likely to require the use of femoral extraction tools [245]. Lastly, the presence of epicardial leads may require surgical access as a component of the procedure [241].

The environment for lead extractions in the pediatric population warrants careful patient-centered assessment for optimal preparedness (Table S1 in Supplemental Appendix 3). As in adults, major complications are relatively rare, but significant potential for life-threatening events exists [236,238,239]. The contribution of complex CHD to the likelihood of successful extraction has varied, ranging from 74% to 94% for complete removal. The rates of major complications, however, have been found to be consistent between 3% and 4% [239,240,242]. Specific complications may be more prevalent based on anatomy and size, such as increased sub-pulmonary AV valve regurgitation in transposition of the great arteries, or increased risk of tricuspid or pulmonary valve involvement related to excess lead slack left for growth in smaller children [243–248]. Additionally, although patient age and size have not been shown to predict venous occlusion, more vigorous fibrous adhesions have been implicated in younger patients [249].

Due to the complexities and potential for serious events in this population, lead extractions should only be performed in centers with an institutional commitment to the development and maintenance of a collaborative team. This includes a need for appropriate facilities, necessary equipment, trained personnel, and the ability to manage all complications. A multidisciplinary team familiar with nuances related to CHD is vital to maximizing procedural safety and efficacy (Table S1 in Supplemental Appendix 3). In particular, it is essential that the cardiac surgeon and surgical team be readily available to immediately provide open-chest surgical repair. Based on congenital anatomy and previous surgeries, emergent surgical approach via thoracotomy (versus sternotomy) may be preferred in certain scenarios, and focused pre-procedure imaging and planning is critical.

It must be recognized that several gaps in knowledge persist in relationship to lead management in children and patients with CHD [250]. This includes limited data in the very young, as well as the impact of multiple extractions over a lifetime on vascular integrity and valvular function. There also continues to be lack of clarity regarding prophylactic lead extractions at the time of generator change [251], and long-term prospective studies on abandonment versus extraction in the young do not exist.

COR Recommendations for CIED Lead Management*		LOE	References
Thrombosis/Vascular Issues			
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	[72]
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	[18,236,237]
Ila	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	[18,251,252]

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COR Recommendations for CIED Lead Management*	LOE	References
Thrombosis/Vascular Issues		
Lead Upgrade or Abandonment		
Ia	Lead removal can be useful for patients with an abandoned lead that interferes with the operation of a CIED system.	C-EO
Ib	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 [18,236,238,239,251]
Ib	Lead removal may be considered for isolated upper extremity venous stenosis or thrombosis without symptoms.	C-EO
Infectious Issues		
I	Lead removal is indicated for CIED-associated endocarditis, bacteremia without an alternative source (particularly <i>Staphylococcus aureus</i>), or bacteremia that persists or recurs despite antimicrobial therapy.	B-NR [18,253]
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 [18,254]
Ib	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 [253]
Other Indications		
I	Lead removal is recommended for patients with life-threatening arrhythmias secondary to retained leads.	C-EO
Ia	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
Ib	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 [250,254,255]
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 [241]
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
Ib	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
Ib	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

*Based on adult lead management guidelines [18,236].

5.1. Recommendation-specific supportive text

The most common indications for lead removal are infection, venous occlusion, advisory or recall as a result of potential lead malfunction, or mechanical lead failure [18,250–256]. Lead management involves the assessment of risks and benefits of whether or not to remove the lead based on the individual clinical condition of the patient as well as lead characteristics [18,236,253].

Upper extremity **venous thrombosis** and venous stenosis are not absolute indications for lead removal. However, instances in which a thrombosis causes significant symptoms (e.g., superior vena cava syndrome, ongoing thromboembolic events), or in which stenosis/occlusion impedes upgrade of an existing device, are generally considered appropriate circumstances to remove an existing lead [236–238].

Infections, which can result in CIED device and lead removal, can generally be grouped into major categories: isolated pocket infection, CIED-associated endocarditis, bacteremia without an

alternative source (particularly *Staphylococcus aureus*), or bacteremia that persists or recurs despite appropriate antimicrobial therapy [18,236]. These situations are associated with challenging management decisions and often require CIED device and lead removal when the infection is more than superficial cellulitis [253,254].

Advisory/recall: The decision to remove an apparently normally functioning lead or leads in response to a manufacturer's or regulatory body's recall or warning is complex and should be performed in close consultation with an electrophysiologist with consideration for the patient's overall clinical status [255,256].

6. Recommendations for CIED follow-up and

6.1. Ancillary Testing

COR Recommendations	LOE	References
CIED Follow-up Recommendations		
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 [257,258]
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
Ia	A standard 12-lead ECG is reasonable at annual in-person evaluation.	C-EO
Ia	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
Ia	An echocardiogram is reasonable for assessment of ventricular function in patients who have >40% ventricular paced rhythm every 1–3 years.	C-LD [259–263]
Ib	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 [264–267]

6.1.1. Recommendation-specific supportive text

Cardiovascular implantable electronic devices (CIEDs) that are currently amenable to remote interrogation and monitoring (RIM) include pacemakers, ICDs, and ICMs. The benefits of routine RIM are extensively validated and maximize the opportunity for prolongation of battery life as well as early detection and intervention of CIED malfunctions, arrhythmic issues, and adverse events [268–272]. Remote evaluation of CIEDs began with transtelephonic monitoring (TTM), an analog-based technology that delivered limited data on pacemaker function via transmission over a telephone landline. RIM technologies, which are now incorporated in all CIEDs, are recommended over TTM because of the additional diagnostic data they provide, but TTM is still in use with older devices that do not have RIM capability. At present, there are no established guidelines for CIED follow-up in the pediatric population with resultant variability in monitoring of pediatric CIEDs [273,274].

Several device, lead, and pocket complications can be seen within the first few days to weeks after CIED implantation, and an in-person evaluation (IPE) is useful in the early post-implant phase. Although specific patient care guidelines for IPE and RIM for children have not been established, the Centers for Medicare & Medicaid Services has established reimbursement guidelines for IPE and RIM for patients with pacemakers.

In addition to monitoring the CIED itself, it is equally important to evaluate the impact of CIED-related consequences on the patient with ancillary testing. Ancillary testing may consist of but is not limited to 12-lead ECG, echocardiogram, ambulatory rhythm monitoring, chest X-ray, and exercise stress testing. The annual IPE should include evaluation of the patient's underlying rhythm. In patients who have >40% paced ventricular rhythm, it is reasonable to assess systemic ventricular function by echocardiogram every 1–3 years for early recognition of pacemaker-induced cardiomyopathy or lead-related valve regurgitation [259–263]. Ambulatory rhythm monitoring and/or exercise stress testing may be useful in patients with arrhythmia concerns or symptoms related to activity and to assist with device optimization [264–267,275–277]. It is reasonable to consider lead surveillance with chest X-ray in the acute post-implant period and to consider repeating every 1–3 years according to growth [241,247].

7. Special considerations

7.1. CIEDs and magnetic resonance imaging

COR	Recommendations	LOE	References
Magnetic Resonance Imaging			
I	MRI in all patients with conditional or nonconditional CIEDs should be performed in the context of a defined institutional protocol.	C-LD	[19]
Ila	MRI is reasonable in patients with nonconditional transvenous CIEDs if there are no fractured, epicardial, or abandoned leads.	B-NR	[278–280]
Ilb	MRI may be considered in patients with epicardial or abandoned leads based on an individualized consideration of the risk/benefit ratio.	C-LD	[279,281,282]

7.1.1. Recommendation-specific supportive text

The 2017 MRI and Radiation Exposure in Patients with CIEDs Consensus Statement provides comprehensive recommendations for individuals with both conditional and nonconditional

transvenous devices [19]. With MRI, there is potential risk for heating of the lead, increase in pacing thresholds, sudden battery depletion, and inappropriate sensing/pacing. The consensus statement also provides guidance for CIED programming and evaluation pre-, during, and post-MRI along with a protocol of testing and patient-specific considerations. However, these recommendations are not specific for patients with abandoned or epicardial CIED leads and make no specific recommendations for MRI in these cases [283,284].

Regarding epicardial lead considerations, younger patients and those with CHD have a greater likelihood of requiring epicardial leads. Additionally, as there are no MRI conditional epicardial leads, even when used with a conditional device, the system is considered nonconditional. The 2017 recommendations suggest a possible contraindication to MRI, and in the pediatric section no recommendations regarding epicardial leads are made. However, when attached to a device, the limited data show only a small increase in risk for substantial alterations of the pacing threshold or changes in sensing after MRI [279–281,285,286].

Regarding abandoned leads, in vitro data suggest that epicardial leads are more likely to generate heat than transvenous leads; however, small studies evaluating MRIs in patients with both epicardial and transvenous abandoned leads suggest that it can be done safely in the majority of cases [282,283,287–289]. Even so, these studies do not imply lack of an effect on the myocardium underlying the abandoned lead. In summary, the data on MRI use in epicardial or abandoned leads are inadequate to provide specific recommendations or an absolute contraindication.

Acknowledging the sparsity of data, but also appreciating the importance of MRI for diagnosis, prognosis, and surgical planning, individualized consideration of the risk/benefit ratio of MRI in young patients must be made on a “case-by-case basis.” [19].

7.2. CIEDs and sports participation

COR	Recommendations	LOE	References
Sports Participation			
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	
Ila	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	[290–296]
III No	ICD placement for the sole purpose of participation in competitive athletics should not benefit be performed.	B-NR	[290–296]

7.2.1. Recommendation-specific supportive text

The safety of sports participation for patients with CIEDs remained fundamentally unstudied until the past decade. Despite a dearth of research, initial published guidelines recommended against strenuous competitive sports participation (greater than class Ia) for patients with pacemakers or ICDs [295–298]. Subsequent to publication of guidelines in 2005, evidence emerged suggesting that risks of sports participation for athletes with CIEDs may be lower than hypothesized [290–293].

Surveys from HRS (2006) and PACES (2013) suggested that many

patients with pacemakers and ICDs had participated in sports without adverse events [290,298]. Thus, an international ICD Sports Registry was initiated and reported in 2013–2018 [292,293]. The registry consisted of 129 patients <21 years of age including varsity high school and college athletes. While shocks occurred during sports, there were no deaths, no resuscitated arrests, and no arrhythmia-related injuries during sports. In addition, the rate of lead malfunction was similar to previously reported rates in unselected populations [292]. The conclusion was made that despite the potential for exercise to be arrhythmogenic, some young patients with ICDs can participate in sports without injury or failure to terminate the arrhythmia.

When questions arise about sports participation in youth with CIEDs, it is now standard practice to counsel patients and families about the risks, including potential for increased rate of ventricular tachyarrhythmias and damage to the pacemaker or ICD system. Counseling is patient specific; the underlying cardiac disease, type of device, indication for implant, position of leads and pulse generators, underlying heart rhythm, patient age, and type of athletic activity are considered when estimating risk [298,299]. Shared decision-making processes that include 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.

7.3. CIEDs in low- and middle-income countries

A quote often used by doctors dealing with cardiac rhythm problems in resource limited settings (or indeed any branch of medicine) is the Italian proverb “Il meglio è l'inimico del bene,” which translates to “better is the enemy of good.” Low- and middle-income countries (LMIC) are defined as those designated by the World Bank based on per capita income [300]. They represent a heterogenous community including countries where the primary deterrent to the use of implantable devices is the cost of the device (India and most countries in Asia and Southern Africa) and those in which the deterrent is both the cost and the availability (sub-Saharan Africa) [301]. These problems have been alleviated to a small extent by philanthropic measures initiated by the Western world as well as universal health care policies announced by various governments in recent years. Pediatric cardiologists in these countries circumvent these problems by using two primary strategies:

1. Patient-specific strategy. Most centers in LMIC tailor the indications of the device to an individual patient instead of following standard guidelines. This is based on available evidence and is not anecdotal, as is widely believed. In post-operative heart block, it has been shown that 95% of AV conduction recovery happens by the 10th postoperative day [50,51]. Children with intermittent AV conduction on telemetry as well as an accelerated junctional rhythm have been shown to have a much higher recovery rate [302,303]. Hence, many centers prefer to wait till the 10th postoperative day before placement of a permanent pacemaker. In children (and young adults) who have intermittent AV conduction and those with a reasonably fast narrow complex escape rhythm, centers may choose to wait even longer for recovery of AV conduction so as to avoid the use of a permanent pacemaker. Late recovery of surgically induced AV block has also been reported [303,304]. Occasionally patients have been discharged home before return of AV conduction, and spontaneous recovery was documented on follow-up [305]. In patients with corrected CHD and normal ventricular function, a single-chamber pacemaker is used in most centers, while a dual-chamber pacemaker is reserved for

children with palliated hearts and more than mild ventricular systolic dysfunction.

2. The use of explanted devices. Devices explanted from deceased patients with a battery life of >50% of a new device have been used in patients from a resource-limited setting [306]. A hypothetical increased risk of infection from an explanted device has been a major deterrent for this approach. However, a recent meta-analysis of 18 studies involving 2270 patients in whom a reused pacemaker was placed revealed no significant increase in the risk of surgical site infection compared to a new device and a small increase in the risk of device malfunction [307]. Even this small risk was shown to be predominantly technical and did not endanger the life of the patient. Standard guidelines on device reuse in India have been published [308].

While most centers have used such inventive strategies to implant a device in children, follow-up interrogation of the device is often challenging. Most pediatric cardiac centers in LMIC are located in a few urban centers with a very large referral area. Frequent travel for device interrogation is often impossible for families because of the costs involved as well as the loss of livelihood. There is no published literature on the gravity of this problem, as most centers lack the resources to follow patients meticulously. Although remote monitoring is ideally suited for these patients, the added cost of the device makes it less attractive. The recent launch of mobile-based remote monitoring pacemakers using Bluetooth technology has immense potential in LMIC if such devices can be priced affordably [309].

7.4. Shared decision-making

COR	Recommendation	LOE	References
Shared Decision-Making			
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	[310–312]

7.4.1. Recommendation-specific supportive text

Shared decision-making is a process whereby patients, families, and providers exchange information and dialogue about medical diagnostic and treatment options [310]. The goal is for patients and their families to reach evidence-informed and value-congruent medical decisions collaboratively with their clinicians. This modern model for health care decision-making has superseded paternalism, a previous model whereby providers made medical decisions on behalf of their patients using the ethical principal of beneficence. A shared decision-making approach, combining the ethical principles of professional beneficence and patient autonomy, has been shown to improve patient outcomes [311,312].

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 pacemaker dependent will be largely uncontested. 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.

Finally, the shared decision-making process is also important and applicable to post-implant diagnostic and treatment decisions for our patients with CIEDs including genetic testing, MRI, sports participation, pregnancy, cardiac surgery, and device reprogramming, removal, or revision.

8. Knowledge Gaps and Future Research

There have been no RCTs involving CIEDs in children. Therefore, the recommendations put forth in this guideline are based on data from observational studies in children, clinical trials in adults, and expert opinion. Clinical trials, especially RCTs, remain challenging in pediatric populations because of low overall event rates in specific diseases and variations in disease progression from birth to adulthood [313].

Critical knowledge gaps exist in several areas [314]. One example is the use of ICDs for the primary prevention of SCD. With reduction in size and the development of novel lead configurations, ICD use in pediatrics has increased dramatically while the age at implant has decreased significantly [130,315]. However, 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 [219,316,317]. Multi-center 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.

Appendix A. Supplementary data

Supplementary data to this article can be found online at <https://doi.org/10.1016/j.ipej.2021.07.005>.

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
Maully J. Shah (Co-Chair)	University of Pennsylvania, Children's Hospital of Philadelphia	None	None	None	Medtronic: 2	None	None	None	None
Michael J. Silka (Co-Chair)	University of Southern California, Los Angeles Children's Hospital	None	None	None	None	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, CS 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
Peter P. Karpawich	The Children's Hospital of Michigan, University Pediatricians PC	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
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 OrphanPharmaceuticals	None	None	None	None	None	None	None
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 Silveti	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
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 Property/ 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

(continued)

Peer Reviewer	Representation	Employment	Honoraria/ Speaking/ Consulting	Speakers' Bureau	Research*	Fellowship Support*	Ownership/ Partnership/ Principal/Majority Stockholder	Stock or Stock Options	Intellectual Property/ Royalties	Other
Ashish A. Nabar	IHRS	Lilavati Hospital, Jupiter	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.

*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.

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Neuromuscular Diseases and Other Progressive Conduction Diseases

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