**Appendix 4**

Of the 130 recommendations in this document, 36 (28%) represent new proposals not previously formally addressed in prior guidelines. The majority of these recommendations address issues unique to pediatric patients, such as ICD indications in very young patients. Furthermore, several of the “new” recommendations are based on specific topics that have been discussed in the text/diagrams in prior guidelines but were not included as formal recommendations. Changes in the category of recommendation from current guidelines are made in 15 recommendations (12%), primarily a change in the class of recommendation by one grade, i.e., change from IIa to IIb. Some of these differences from adult guidelines reflect variances in disease progression when there is onset in childhood, such as Kearns-Sayre syndrome or myotonic dystrophy. Other changes reflect considerations such as small patient size and the need for surgical implant of epicardial device leads, the need for decades of lead survival and vascular patency, or device management in the setting of minor epicardial lead abnormalities or superficial wound infections. The presence of comorbid conditions such as genetic or neurologic abnormalities or anatomic abnormalities that preclude use of transvenous lead systems also were considered. Otherwise, as stated in the preamble, there is often overlap in the care of patients in the 18- to 21-year-old range, and in the absence of confounding factors, CIED management guidelines should be consistent. Finally, there were nine recommendations (7%) in which some modification of the wording of a prior recommendation was revised.

The attached table (Table S6) lists each specific recommendation, the prior guidelines or expert consensus statements that have been used as a basis for the recommendation, and if there is variance, specification of change. Finally, given the scope of this document, overlap with other guidelines that will be published in the near future will undoubtedly occur. Therefore, these recommendations will need to be periodically updated to reflect current best standards of practice, and they do not replace the need for individual clinical judgment.

**Table S6: Variations Between Adult and Pediatric CIED Recommendations**

New: New recommendation that has not been published before

Modification: Modification in some aspect/wording of a previously published recommendation

Change: Change in category (COR) of a previously published recommendation

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| **Section** | **PACES 2021 COR**  **(number-specific recommendation in COR category)** | **New, modification, or change from existing guidelines** | **Rationale for change** |
| **2. Permanent pacemakers** |  |  |  |
| 2.2 Isolated sinus node dysfunction | IIb | New | Consensus of the writing committee: Symptom rhythm correlation is not always possible. |
| 2.3 Isolated congenital complete atrioventricular block | I (#3) | Modification from 2012 device therapy guideline update7  • Heart rate threshold changed to ≤50 bpm from <55 bpm | To avoid unnecessary early pacemaker implant in asymptomatic neonates and infants; heart rate ≤50 bpm was selected, as many infants are hemodynamically stable with heart rate of 50–55 bpm. The recommendation is qualified by the statement that heart rate alone should not be used as implant criteria. |
|  | IIa (#1) | Change from 2018 Bradycardia guideline2  • To class IIa from class I | COR for pacing is IIa as the risk/benefit of long-term pacing and lifetime of device/lead revisions need to be considered in young children, especially when implanting a pacemaker for heart rate alone, in the absence of symptoms.  This is the same COR as 2012 device therapy guidelines update7 for children, adolescents, and CHD and 2014 ECS for adult CHD.9 |
|  | IIa (#2) | New | Progressive LV dysfunction and mitral insufficiency associated with cardiovascular mortality in some cases.33,43 |
|  | IIb | Change from 2018 bradycardia guideline2  • To class IIb from class IIa | COR for pacing is weaker in asymptomatic children as the risk/benefit of long-term pacing and lifetime of device/lead revisions need to be considered, especially when implanting a pacemaker in young/small patients.  Same COR as 2012 device therapy guideline update7 for children, adolescents, and CHD. |
| 2.4 Atrioventricular block: Other considerations | I (#2) | New | This refers to patients who are diagnosed with advanced AV block in childhood and cause cannot be established. |
|  | IIa | New | Exercise-induced advanced AV block may occur due to infranodal disease in which 1:1 AV conduction may not occur with a high atrial rate.2,47,48 If patient is *symptomati*c with exercise-induced advanced AV block, refer to COR I (#2). |
|  | IIb | New | Intermittent AV block without a reversible cause may occur in ostensibly normal children. Pacing may improve symptoms.49 |
| 2.5 Postoperative atrioventricular block | I (#1) | Modification from 2018 bradycardia guideline2  Modification from 2012 device therapy guideline update7  • Threshold changed from 7 days to 7–10 days | The 2018 bradycardia guideline2 supportive text states waiting 7–9 days is unnecessary, but to generally avoid early implantation <72 hours. Recent data show that AV node function can recover beyond the seventh postoperative day.50-53 |
|  | I (#2) | New | This is supported by references 52, 54, and 55. |
|  | IIb (#1) | Change from 2012 device guideline update7  • To class IIb from class IIa | Strength of recommendation is reduced, as syncope may occur due to causes unrelated to transient postoperative AV block. |
|  | IIb (#2) | New | Consensus of the writing committee: A postoperative waiting period of 7 days may not be necessary in *selected* cases of postoperative AV block when there is a high probability of significant surgical damage to the conduction system. The 2018 Bradycardia guideline2 supportive text states waiting 7–9 days is unnecessary. |
|  | IIb (#3) | New | Consensus of the writing committee: In CHD substrates prone to progressive conduction abnormalities, transient surgical damage to the conduction system may predispose to further impairment of the conduction system. |
| 2.6 Congenital heart disease: Specific considerations | I | Modification from 2012 device guideline update7  • Heart rate threshold changed to 60–70 bpm | Heart rate threshold changed to allow clinical judgment in view of varying complexities of CHD. |
| 2.7 Post cardiac transplantation | IIb (#2) | New | Symptomatic bradycardia or AV block may be the first symptoms of coronary artery vasculopathy in some cases.79,84 |
| 2.8 Neuromuscular diseases and other progressive cardiac conduction diseases | I (#2) | Change from 2018 bradycardia guideline2  • To class I from class IIa | Kearns-Sayre syndrome has early onset in childhood and high risk of AV block and SCD due to the unpredictable progression of conduction disease.97,98 |
|  | I (#2) | Change from 2018 bradycardia guideline2  • To class IIa from IIb | COR is stronger given that the risk of developing cardiac disease including progressive AV or intraventricular conduction defects and tachyarrhythmias is higher in younger patients with myotonic dystrophy type 1.88,90 |
|  | IIa (#2) | Modification from 2018 bradycardia guideline2  • PR >240 ms *and/or* left bundle branch block | Either marked first-degree AV block ***or*** development of left bundle branch block is suggestive of conduction disease, and device intervention at this stage may be reasonable instead of waiting for further progression to a combination of first-degree AV block and left bundle branch block. |
|  | IIb | New | Prophylactic device therapy may be considered in selected high-risk patients with progressive cardiac conduction disease due to the unpredictable progression of conduction disease.97,100 |
| 2.9 Neurocardiogenic syncope | IIa | Change from 2017 syncope guideline11  • To class IIa from class IIb | The COR is stronger, as the recommendation is qualified for patients with “severe and recurrent spells.” Pacing using appropriate pacemaker settings has been shown to be effective in the prevention of loss of consciousness and seizures in children with breath-holding spells.101-103 |
|  | IIb (#2) | Change from 2018 Bradycardia guideline2  • To class IIb from class IIa | Strength of recommendation for pacing is weaker in children as data are limited and the risk/benefit of long-term pacing and lifetime of device/lead revisions need to be considered. |
|  | III (#1) | New | Consensus of the writing committee: Benefit of pacing for patients with only tilt-positive cardioinhibitory syncope remains unproven. |
|  | III (#2) | New | Consensus of the writing committee: Patients with syncope due to a vasodepressor response are unlikely to respond to permanent pacing.11 |
| 2.10 Cardiac channelopathies | IIb (#1) | New | Limited data shows clinical benefit of pacing in selected high-risk LQTS patients with functional 2:1 AV block.112 |
|  | IIb (#2) | New | Limited data show clinical benefit of intentional pacing at a faster heart rate in selected patients.109,114 |
|  | III | New | Conventional atrial pacemaker lead implantation in patients with symptomatic sinus node disease may fail due to partial or complete atrial standstill.115 |
| 2.11 Inflammation/infection | IIa | New | Unlike other infectious causes, AV block may not be reversible in Chagas disease.119-121 |
| **3. Implantable cardioverter defibrillators** |  |  |  |
| 3.2 General recommendations for ICD therapy | IIb (#1) | New | Refers to VT that is judged to be non–life threatening. |
|  | IIb (#2) | New | Consensus of the writing committee: Primary prevention ICD therapy may be considered in selected patients based on clinical judgment and strength of risk factors including family history of recurrent SCA. This is similar to the IIb COR for familial cardiomyopathy in the 2012 device guideline update.7 |
| 3.3.1 Long QT syndrome | I | Modification from 2013 genetic arrhythmias guideline14 to include:  • ICD + use of beta-blockade  • Medical therapy and/or sympathectomy as alternatives to ICD in select cases | Successful outcomes with beta-blockers and cardiac sympathectomy without ICD have been reported in select LQTS cases.142,143,147 |
|  | IIb | New | Primary prevention ICD therapy may be considered in select LQTS patients with high-risk pathogenic gene mutations and also in LQTS patients with other risk factors associated with life-threatening arrhythmias.149-153 A QTc >500 ms has been cited as one of the risk factors for SCD in the text, similar to the IIb COR in the 2017 VA/SCD guideline.12 |
| 3.3.2 Catecholaminergic polymorphic ventricular tachycardia | IIa | Changed from 2013 genetic arrhythmias guideline14 and 2017 VA/SCD guideline12  • To class IIa from class I | The strength of recommendation was reduced based on recent data that an ICD was not associated with improved survival when aborted SCA was the initial presentation of CPVT.126,156 A high incidence of inappropriate shocks, proarrhythmia, electrical storm, and ICD complications have been reported in CPVT. |
|  | IIb | Changed from 2013 genetic arrhythmias guideline14  • To class IIb from class I | The strength of recommendation was reduced based on data that ICD therapy for polymorphic and bidirectional VT did not demonstrate successful primary termination.157,164 Inappropriate shocks, electrical storm, and ICD complications were common. |
| 3.3.3 Brugada syndrome | IIa | Change from 2017 VA/SCD guideline12  • To class IIa from class I | This recommendation is similar to the 2013 genetic arrhythmias guideline14 but differs from the 2017 VA/SCD guideline.12 There are diverse etiologies of syncope in children. Risk/benefit of ICD implant and long-term complications (especially when implanted at a very young age) and long-term ICD and lifetime of device/lead revisions need to be considered. |
|  | IIb | New | Consensus of the writing committee: Consideration of ICD implantation for syncope when type I BrS ECG pattern only provoked with pharmacologic challenge. |
| 3.4.1 Hypertrophic cardiomyopathy | IIa | Modification from 2017 VA/SCD guideline12  • Nonsustained VT included as a risk factor | Consistent with 2020 HCM guidelines.15 |
| 3.4.3 Arrhythmogenic cardiomyopathies | IIa | Change from 2017 VA/SCD guideline12  • ICD for sustained VT changed to class IIa from class I  • ICD for LVEF ≤35% changed to class IIa from class I | A qualification of hemodynamically stable was added to sustained VT in this recommendation.16,192 Due to the low prevalence of manifest disease in children, it is unclear if survival benefit of ICD can be extrapolated from adult data to children with hemodynamically stable VT or LVEF ≤35%. The COR is consistent with 2019 guideline16 for syncope suspected due to VA. Risk/benefit of long-term ICD therapy and lifetime of device/lead revisions need to be considered in young children. |
|  | IIb | New | There are limited data on risk stratification for primary prevention ICD implantation in children with ACM. ICD therapy may be considered in selected patients based on clinical judgment and strength of risk factors.16,192-193 The COR is consistent with 2019 guideline16 for ARVC. |
| 3.4.4 Nonischemic dilated cardiomyopathies | IIb | Change from 2012 device therapy guideline7 and 2017 guideline12  • To class IIb from class I | The supportive data for primary prevention ICD in patients with nonischemic cardiomyopathy (LVEF ≤35%), mild heart failure symptom on guideline-directed medical therapy recommendation are predominantly derived from the Sudden Cardiac Death in Heart Failure Trial (SCD-HeFT)198 that enrolled patients with median age 60 years and included those with ischemic cardiovascular disease. Therefore, these data cannot be generalized to children with NIDCM. Data from the Pediatric Cardiomyopathy Registry and other similar series demonstrate very low rate of sudden arrhythmic death in the pediatric population.196,198 |
|  | III (#2) | New | Consensus of the writing committee that ICD implant is not considered appropriate in young patients who remain hospitalized in a monitored critical care unit |
| 3.5 Congenital heart disease | IIb (#1) | New | Consensus of the writing committee: Given the potential for hemodynamic instability of a previously stable VT, ICD therapy may be considered. |
| **4. Insertable cardiac monitors** | IIa (#3) | New | ICM for refining management in inherited arrhythmia syndromes.226,228 |
|  | IIb (#3) | New | ICM for severe but infrequent palpitations. |
|  | IIb (#4) | New | Consensus of the writing committee: ICM for detecting occult arrhythmias in high-risk patients. |
| **5. CIED Lead management** |  |  |  |
| Lead upgrade or abandonment | IIb (#1) | New | The IIa COR in the 2017 CIED lead management ECS18 regarding lead removal for >5 leads through the SVC is not applicable to children. Instead, the decision to remove a lead in children depends on number of existing leads relative to patient size, anatomy, vasculature, and risk/benefit of lead extraction. |
|  | IIb (#2) | New | Consensus of writing committee: Lead removal for asymptomatic upper extremity venous stenosis or thrombosis. |
| Infectious tissues | IIb | Change from 2017 guideline18  • To class IIb from class I | The recommendation has been qualified by adding s*uperficial* pocket infection. Superficial pocket infections can be treated with antibiotics.254 Risk/benefit of lead extraction should be considered in this situation. This is consistent with 2017 CIED lead management ECS18 (Figure 3). |
| Epicardial leads | I (#1) | New | Coronary artery compression resulting in myocardial injury can have life-threatening consequences.240 |
|  | IIb (#1) | New | Consensus of the writing committee: In situ epicardial leads may pose a potential risk of myocardial injury depending on their location.240 Risk/benefit of epicardial lead removal should be considered. |
|  | IIb (#2) | New | Consensus of the writing committee: Potential for epicardial leads to cause coronary compression, valve impingement, and myocardial strangulation.240 Risk/benefit of epicardial lead removal should be considered. |
| **6. CIED follow-up and ancillary testing** |  |  |  |
| Follow-up | I (#1) | Change from 2015 guideline257  • To class I from class IIa | Consensus of the writing committee: In-person evaluation and establishing RIM within 2–4 weeks |
|  | I (#4) | Modified from 2008 guideline258  • Modified to class I | Consensus of the writing committee: Assign a class I strength of recommendation for minimum frequency of in-person follow-up or remote monitoring. |
| Ancillary testing | IIa (#1) | New | Consensus of the writing committee: Obtain 12-lead ECG to assess baseline paced or non-paced rhythm. |
|  | IIa (#2) | New | Consensus of the writing committee: Obtain periodic chest X-rays. |
|  | IIa (#3) | New | Consensus of the writing committee: Obtain periodic echocardiograms to assess ventricular function. |
|  | IIb | New | Consensus of the writing committee: Consider exercise stress test, ambulatory ECG. |
| **7. Special considerations** |  |  |  |
| 7.1. CIEDs and magnetic resonance imaging | I | Change from 2017 guideline278  • To class I from IIa | Qualifying statement added that MRI in patients with *all* types of CIEDs should be performed under institutional protocol. |
|  | IIb | New | Based on limited data, MRI can be considered in presence of epicardial or abandoned leads based on risk/benefit ratio.281,282 |
| 7.2. CIEDs and sports participation | I | New | Consensus of the writing committee: Decisions regarding sports participation primarily based on diagnosis and physiology in most patients. |
|  | IIa | New | Based on limited data, the risk of death, SCA, or arrhythmia-related injury in patients with CIEDs during sports participation may be low.290-293 |
|  | III | Modified from 2015 eligibility recommendations for competitive athletes guideline294  • Modified to apply to all patients with cardiovascular abnormalities and CIEDs | Recommendation expanded to include all patients with cardiovascular abnormalities and not limited to patients with cardiomyopathies due to possibility of device-related  complications with high-intensity sports. |