**Appendix 3**

**Table S1. Required Personnel for Lead Extraction**

|  |  |
| --- | --- |
| Primary operator | Physician performing the extraction procedure who is appropriately trained and experienced in performing lead extractions and the management of complications in children. Particular experience with extractions in children and congenital heart disease is necessary as relevant to the case. |
| Cardiothoracic surgeon | Cardiothoracic surgeon knowledgeable in lead extraction procedures and potential complications therein, who is immediately available to manage life-threatening complications that may require emergent surgical intervention. Understanding of congenital and postoperative anatomy is necessary for optimal surgical approach. |
| Surgical support staff | Necessary surgical support staff, including a perfusion team and nursing support, who are immediately available with necessary equipment to manage life-threatening complications that may require emergent surgical intervention. |
| Anesthesiology | Anesthesiologist to provide general anesthesia support for the case and possible complications. |
| Imaging support | Personnel to operate and troubleshoot fluoroscopic and other imaging equipment, including rapid access to echocardiography. |
| “Scrubbed” assistant | Assistant to the primary operator who is knowledgeable about the extraction procedure who is scrubbed and available to assist with all facets of the lead extraction. |
| Nonscrubbed assistant | Personnel who is/are knowledgeable about the extraction procedure and available to provide necessary equipment during the case and able to assist in an emergency. |

**Evidence Tables**

**Table S2. Evidence for Pacemaker Implantation in Patients With Specific Conditions**

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| **Isolated Sinus Node Dysfunction** | | | | | | |
| **Study author and reference** | **Year** | **Study design** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Albin et al24 | 1985 | Single-center retrospective study | 39 patients; mean age 23 years; 25 with CHD | Sinus bradycardia or brady-tachy syndrome | Syncope/dizziness; survival | Mean interval between CHD surgery and pacemaker implant 108 months; d-TGA most common. Symptoms resolved if no CHD. |
| Gillette et al25 | 1983 | Single-center retrospective study | 51 patients; mean age 10 years; A, V, and DDD paced | Symptomatic sinus bradycardia | Resolution of symptoms; outcome | 45/49 patients with symptoms improved/resolved; 11/18 with brady-tachy improved. 7/51 patients required revision in median follow-up 26 months. |
| Gillette et al29 | 1985 | Single-center retrospective study | 40 patients; mean age 11 years | Sinus bradycardia; all AAI paced | Resolution of symptoms; AV block; system revision | 3.5-year follow-up; no patient developed AV block; 6 improved with Anti-tachycardia pacing (ATP); 28/29 improved with Anti bradycardia pacing alone. |
| Kardelen et al28 | 2002 | Single-center retrospective study | 26 patients; median age at pacemaker implantation: 9 years | ≤21 years old; symptomatic SND | Syncope/dizziness; survival | All patients symptoms improved. Lead sense/pace threshold were stable over mean 34 months. |
| **Isolated Congenital Complete Atrioventricular Block** | | | | | | |
| **Study author and reference** | **Year** | **Study design** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Michaëlsson et al33 | 1995 | Prospective  study | 102 patients;  mean age 36 years (16–66) | Patients with isolated CCAVB age >15 years | Freedom from CCAVB-related mortality and morbidity | Stokes-Adams attacks in 35 patients with fatal outcome in 8 (23%). Pacemaker was implanted in 52 patients. Over a 30-year follow-up, 11 deaths (mean age 42 years, range 24–58)  in adults with isolated CCAVB who have been asymptomatic during infancy and childhood, mortality is ~5% and morbidity is 30% between 15 and 50 years. |
| Baruteau et al31 | 2012 | Multicenter retrospective study | 141 patients;  mean age 3.6 ± 4.2 years | Patients with nonimmune AV block; diagnosed in utero to 15 years | Freedom from  1) symptoms,  2) pacemaker therapy, and  3) AV block–related mortality | 84% of patients were asymptomatic. Pacemakers were implanted in 80% with 16% during the first year of life and 80% before 10 years of age. Mean age at PM was 32 months in congenital AV block and 80 months in childhood AV block. Pacing indication was prophylactic in 62%. At mean follow-up of 11.6 ± 6.7 years, there was no mortality and no patient developed dilated cardiomyopathy. |
| Balmer et al32 | 2002 | Single-center retrospective study | 32 patients;  median age 0.4 years | CCAVB with or without CHD | Pacemaker implant: frequency,  indications,  outcome | All 9 patients with CHD paced; 17/23 without CHD paced. Indications: rate 15, low CO 8, syncope 3. Pacemaker-related complication in 11/46; 1 death. |
| Jaeggi et al30 | 2002 | Single-center retrospective study | 102 patients;  29 fetal (F),33 neonate (N),  40 children (C) | Isolated CCAVB | Pacemaker implant: frequency,  indications, outcome | F,N,C mortality 43%, 6%, 0% respectively, up to 20 years of age.  ↑ Mortality with fetal diagnosis, hydrops, endocardial fibroelastosis, delivery ≤ 32 weeks.  Pacemaker implant earlier age with F and N. By 20 years of age, only 115 (N) and 12% (C) not paced. |
| **Postoperative Atrioventricular Block** | | | | | | |
| **Study author and reference** | **Year** | **Study design** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Romer et al51 | 2019 | Multicenter retrospective cohort study from Pediatric Cardiac Critical Care Consortium (PC4) Registry | 15,901 patients | Hospitalized patients without preoperative AV block or pacemakers who underwent an index CHD operation between 2014 and 2017 | 1) Postoperative AV block,  2) restoration of normal conduction after postoperative AV block, and  3) freedom from pacemaker implantation | The incidence of high-grade second-degree and/or CAVB was 2.7%, and the incidence of persistent AV block beyond 7 days and/or requiring pacemaker was 1%. In patients with transient AV block, median time to resolution was 1.9 days (IQR 0.82, 4.8); 86% recovered conduction by 7 days and 94% by 10 days. |
| Aziz et al52 | 2012 | Single-center retrospective cohort study | 44 patients | Patients with transient AV block after CHD surgery between 1994 and 2010 | Late AV block after recovery from transient AV block | 15.9% developed late AV block, occurring as late at ~9 years following initial recovery of AV conduction. The median duration of transient AV block was 5 days compared with 9 days in the late AV block group (*P* = .01). The risk of late AV block for the patients with 7 days of postoperative transient AV block or longer was 13 times greater than for the patients with fewer than 7 days of transient AV block (*P* = .01). |
| Villain et al55 | 2003 | Two-center retrospective cohort study | 11 patients | Patients with transient CAVB after CHD surgery between 1988 and 2000 who developed permanent or paroxysmal CAVB >1 month after surgery | Late AV block after recovery from transient AV block | Among patients with transient postoperative AV block, those with AV block lasting >48 hours and change in QRS axis and/or had prolonged PR intervals after recovering AV conduction were at increased risk for developing late-onset CAVB (2 months to 10 years following CHD surgery). |
| Ayyildiz et al57 | 2016 | Single-center retrospective cohort study | 1,550 patients | Pediatric patients undergoing CHD surgery between 2010 and 2015 | Transient and persistent postoperative AV block | Postoperative permanent and transient AV block incidence rates were 1.9% and 4.2%, respectively. In transient AV block, 84% recovered by 7 days and 97% by 10 days. |
| Huhta et al59 | 1983 | Single-center retrospective cohort study | 107 patients | Patients with AV discordance (without situs ambiguous) evaluated between 1951 and 1981 | Spontaneous complete AV block | Spontaneous AV block occurred in 21% patients. The risk of AV block increased linearly with age at a range of ~2% per year. |
| **Congenital Heart Disease** | | | | | | |
| **Study author and reference** | **Year** | **Study design** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Pinsky et al37 | 1982 | Single-center retrospective cohort study | 65 patients; 25 with CHD | Patients with CCAVB between 1955 and 1979 | 1) Pacemaker implantation  2) Freedom from mortality | 17 (26%) underwent pacemaker implantation, and 9 had CHD. Overall, death occurred in 28% of CHD patients vs. 4.6% of patients with structurally normal hearts (age at death: 1 day to 14 years of age). Of the 10 (15%) who died, 7 (70%) deaths occurred in <1 year of age and 7 (70%) patients had CHD. |
| Glatz et al42 | 2008 | Single-center retrospective cohort study | 13 neonates; 7 with CHD | Neonates with CCAVB requiring temporary or permanent pacing in the first 24 h of life | Freedom from mortality | 10 (76%) CCAVB diagnosed in utero. 4 of 7 CHD patients had heterotaxy. 6 (46%) patients had hydrops at birth. 7 (53%) died, of which 6 had CHD including all 4 patients with heterotaxy. 4 of 6 patients with hypdrops also died. Severely affected neonates with CCAVB and those with complex CHD had poor outcome despite early pacing, whereas those with structurally normal hearts had a favorable outcome. |
| Silka et al63 | 1990 | Single-center retrospective observational study | 21 patients; mean age 11 years (range 2– 19) | Children with  CHD and chronic bradycardia with syncope or heart failure and tachycardia requiring medication | Freedom from tachyarrhythmias | Prevention of bradycardia by pacing (without change in medical therapy) was associated with a decrease in the frequency of supraventricular (*P* = .008) and ventricular (*P* = .02) tachyarrhythmias. The frequency of atrial flutter was not altered. Prevention of tachycardia was more frequently associated with the AAI and DDD modes of pacing compared to VVI (*P* = .08). |
| Rhodes et al64 | 1995 | Single-center retrospective observational study | 18 patients (14 patients <18 years of age) | CHD with atrial antitachycardia (AAI-T mode) pacing for treatment of recurrent intra-atrial re-entrant tachycardia resistant to medical therapy | Termination of atrial tachycardia with atrial ATP | 6 patients had 189 episodes of tachycardia successfully converted with AAI-T pacing, 4 patients had 8 episodes of tachycardia detected but not successfully converted, and 8 patients had no episodes of tachycardia with antibradycardia pacing alone. Two subjects died suddenly (death associated with attempted pace cardioversion of tachyarrhythmia). |
| Kramer et al65 | 2018 | Single-center retrospective study | 91 patients; median age 27.3 (range 6.9–59.8) | CHD patients with atrial antitachycardia pacing devices between 2001 and 2016 | Freedom from DC cardioversion | 72% of intra-atrial re-entrant tachycardia successfully terminated by atrial ATP.  Of 36 patients with DC cardioversion prior to atrial ATP, only 7 required cardioversion after atrial ATP. No mortality occurred. |
| Stephenson et al66 | 2003 | Multicenter retrospective study | 28 patients; mean age 30 ± 18.0 years | CHD patients with Medtronic AT500 pacemaker | Termination of atrial tachycardia with atrial ATP | In 15 patients who had atrial arrhythmias, 54% of 167 atrial tachycardia episodes were successfully terminated by atrial ATP. Atrial arrhythmias were appropriately detected in 14/15 patients with arrhythmias. No mortality occurred. |
| Barber et al68 | 2005 | Single-center prospective crossover trial | 21 patients; median age 4 years (range 2–18) | Patients s/p Fontan surgery in ICU | Acute change in hemodynamics | VOO pacing mode resulted in significantly worse hemodynamics when compared to DOO and AOO pacing with a higher LA and PA pressures and lower Qs and mean arterial pressures. |
| Khairy et al71 | 2006 | Multicenter retrospective study | 202 adolescents and adults; mean age 33.9 ± 18.0 years | CHD patients with intracardiac shunt | Systemic thromboembolic event | CHD patients with intracardiac shunt. 24 (12%) had thromboembolic event (15.6% with transvenous leads, 8.9% with epicardial leads, 11% no leads). In multivariate analyses, transvenous leads remained an independent predictor of systemic thromboemboli. Aspirin or warfarin was not found to be protective. |
| DeSimone et al72 | 2013 | Single-center retrospective study | 6,075 adult patients divided into patent foramen ovale (PFO) and non-PFO groups matched for CHA2DS2-VASc score | Patients with endocardial CIED leads between 2000 and 2010 | A stroke or transient ischemic attack consistent with cardiac etiology | Stroke/transient ischemic attack confirmed in 30/364 (8.2%) PFO vs. (2.0%) non-PFO patients (*P* <0.0001). Patients in PFO group were more likely to have atrial fibrillation. Study suggested screening for PFOs prior to CIEDs; if a PFO is detected, PFO closure, anticoagulation, or nonvascular lead placement may be considered. |
| Supple et al73 | 2011 | Single-center retrospective study | 86 adult patients | Patients with a CIED with intracardiac echo (ICE) during catheter ablation | Mobile thrombus on CIED lead(s) | Mobile thrombus seen on leads with ICE catheter in 30%, although none were seen on transthoracic echo before the procedure. 84% patients had CHF, and average LVEF was 40%. 32% with a thrombus were on warfarin, while 52% without were on warfarin. |

**Table S3. Evidence for ICD Implantation in Specific Conditions**

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| **Cardiac Channelopathies** | | | | | | |
| **Long QT Syndrome** | | | | | | |
| **Study author and reference** | **Year** | **Study type** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Spazzolini et al145 | 2009 | LQTS Registry | 212 patients with ECG <1 year old | QTc >450 ms | Risk factors for LQTS patients with cardiac events in the first year of life and outcome | 28/212 (13%) had cardiac events. During first year of life: female sex, slow heart rate, and QTc >500 are risk factors. In follow-up syncope, QTc >500, and male sex were identified risk factors. Previous history of aborted cardiac arrest (ACA) <1 year increased risk of subsequent ACA by 2.3-fold. |
| Lui et al148 | 2011 | LQTS Registry | 1,678 patients; 277 with ICD | LQTS >450 ms QTc or LQTS mutation; <20 years old | Occurrence of second to fourth events from 1 to 20 years | Recurrent syncope is a predictor of fatal or near fatal events, independent of QTc and age-sex interactions. QTc duration >500 ms is major predictor of first syncopal episode. |
| Goldenberg et al149 | 2008 | LQTS Registry: retrospective/prospective | 3,015 patients | LQTS probands; mean age 7.5 years | Define risks for LQTS events ages 1–12 years | LQTS males have a higher rate of fatal/near fatal cardiac events than females. Risks: QTc> 500 and prior history of syncope in males; prior history of syncope alone in female. Beta-blockers reduce risk of LTE. Family history of SCD in first-degree relative was not a childhood predictor of ACA/SCD. |
| Etheridge et al154 | 2007 | Multicenter study | 128 patients | LQTS <18 years old  at ICD implant | Appropriate or inappropriate ICD shocks or complications | 22% received appropriate ICD therapy with mean first shock 18 ± 22 months. 17% had inappropriate ICD therapy (ST or atrial fibrillation). Recommended ICD implant for patients with previous cardiac arrest, documented VT/torsades de pointes, syncope on beta-blockers. For SCN5A mutation, no consensus. |
| **Catecholaminergic Polymorphic Ventricular Tachycardia** | | | | | | |
| **Study author and reference** | **Year** | **Study type** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Van der Werf et al126 | 2019 | CPVT Registry | 136 patients; mean age 14 years | CPVT with SCA guideline directed therapy | ICD shocks, cardiac events | 79 patients with ICD; appropriate ICD therapy in 46%. SCD occurred in 3.8% of patients with an ICD, and no patients without ICD; 25% had inappropriate shocks; 29% had device-related complications; 39% of patients without an ICD had an ICD implanted during follow-up. |
| Roston et al156 | 2018 | Meta-analysis  (53 studies) | 1,429 CPVT patients; 503 with ICDs; mean age 15 years | CPVT with ICD | Use and outcomes of ICDs in patients with CPVT | 47.3% primary prevention ICD; 40% >1 appropriate shock, 21% >1 inappropriate shock, 20% electrical storm. 1.4% died. Of the 7 that died, 4 deaths were secondary to electrical storm. |
| Miyake et al157 | 2013 | Multicenter  retrospective study | 24 patients; mean age 10 years | CPVT with ICD | ICD shocks and outcome | 14/24 patients had ICD shocks; 46% inappropriate; 8.5% of total shocks resulted in a more malignant ventricular arrhythmia; VF shocks successful; polymorphic VT, bidirectional VT did not demonstrate successful primary termination. |
| Roses-Noguer et al164 | 2014 | Single-center retrospective study | 13 patients; mean age 17.5 years | CPVT with ICD | ICD shocks and outcome | 96 shocks; 70 appropriate; 26 inappropriate. Shocks effective for VF; shocks ineffective for VT. |
| **Brugada Syndrome** | | | | | | |
| **Study author and reference** | **Year** | **Study type** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Gonzalez Corcia et al166 | 2017 | Single-center retrospective study | 128 patients; mean age 15 years | BrS with ICD | Utility of electrophysiology study in pediatric BrS | Inducibility of VAs during electrophysiologic study was more frequent in symptomatic patients. 10 arrhythmic events occurred in 9 symptomatic patients (event rate 4.5% per year). No events in asymptomatic group. |
| Gonzalez Corcia et al167 | 2017 | Single-center retrospective study | 95 patients; mean age 12.9 years | BrS with ICD | Variables associated with presentation as SCA or syncope | Main clinical variables for SCD; spontaneous type I ECG; SND and/or AT; conduction abnormality induction of VAs during programmed stimulation. |
| Gonzalez Corcia et al170 | 2018 | Single-center retrospective study | 35 patients ; mean age 13 years | BrS with ICDs  <20 years old | Predictors and outcomes of ICD in BrS | 9/36 patients ICD for VT/VF. 3 patients expired in VF storm despite ICD therapy. Aborted SCD and type 1 ECG independent predictors of ICD therapy |
| Andorin et al171 | 2016 | Multicenter retrospective study | 106 patients; mean age 11 years | <19 years old with BrS  with or without ICD | Predictors of cardiac events in BrS patients | 75% asymptomatic, symptoms: 14% syncope, 6% aborted SCD or sustained VT, 5% other. During follow-up 9% life-threatening arrhythmias and 3 deaths (all on quinidine without ICD). Fever trigged 27% of life-threatening events. |
| **Cardiomyopathies** | | | | | | |
| **Hypertrophic Cardiomyopathy** | | | | | | |
| **Study author and reference** | **Year** | **Study type** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Maron et al172 | 2016 | Multicenter retrospective study | 474 patients; 231 with ICDs | <30 years old with HCM from 2 centers; mean age 17 years | Events and ICD intervention rates in those with ICDs | Of those with ICDs, 18% had appropriate ICD discharges (58% of secondary prevention vs. 15% of primary prevention). 26% had ICD related complications |
| Maron et al173 | 2013 | Multicenter retrospective study, registry data | 224 patients | <20 years old with HCM from 22 centers; mean age 14 years | ICD intervention rates | Secondary prevention: 5% per year primary prevention: 3% per year 5-year cumulative probability of ICD shock 17%, rate of appropriate discharge in primary prevention was the same regardless of no. of risk factors |
| Miron et al174 | 2020 | Multicenter retrospective study, registry data | 572 patients | <18 years of age with HCM from 11 centers; median age 9.8 years | 5-year cumulative proportion of SCD events was 9% | Model for SCD risk prediction in pediatric HCM with >70% accuracy. Highlights difference in risk factors between pediatric and adult HCM patients. Risk factors evaluated included age at diagnosis, FH of SCD, unexplained syncope within 6 months of diagnosis, NSVT, IVSD *z* score, LVPWD *z* score, LA diameter *z* score, and peak resting LVOT gradient. LVOT gradient and family history were not associated with SCD. |
| Norrish et al175 | 2017 | Meta-analysis (25 studies) | 3,394 patients | <18 years old with HCM and either SCD or SCD-type events | SCD or SCD-type events | Four “major” factors for SCD identified: previous adverse cardiac event, nonsustained ventricular tachycardia, syncope, and extreme left ventricular hypertrophy (maximum left ventricular thickness >30 mm or *z* score >6 |
| Balaji et al176 | 2019 | Multicenter retrospective study | 446 patients | <20 years old with HCM from 35 centers; mean age 10.years | Lethal arrhythmic events | Conventional adult HCM risk factors were not associated with a higher risk of life-threatening arrhythmic events in children; CE in young associated with left ventricular posterior wall thickness and absence of LVOT obstruction. |
| Norrish et al177 | 2019 | Multicenter retrospective longitudinal cohort | 1.024 patients | ≤16 years old with HCM from 39 centers; median age11 years | 9% SCD or equivalent event over median 5 years | Risk prediction model developed using preselected variables (unexplained syncope, maximum left ventricular wall thickness, LA diameter, LVOT, and NSVT). For every 10 ICDs implanted in patients with ≥6% of a 5-year SCD risk, 1 patient may potentially be saved from SCD. |
| **Arrhythmogenic Cardiomyopathies** | | | | | | |
| **Study author and reference** | **Year** | **Study type** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Te Riele et al191 | 2015 | Single-center retrospective study | 75 patients; 53 with ICDs | Pediatric patients with ARVC; mean age 15 ± 2 | Freedom from VT, cardiac transplant, and death | 25% of pediatric patients presented with SCD (significant difference with adults, who present more with VT). Worse outcome during onset, but in the long term after diagnosis survival free from VT, cardiac transplant, and death is similar in pediatric and adult age. Pediatric patients with ARVD are typically male mutation carriers presenting in adolescence with SCD. |
| **Nonischemic Dilated Cardiomyopathy** | | | | | | |
| **Study author and reference** | **Year** | **Study type** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Pahl et al196 | 2012 | Multicenter  retrospective study from Pediatric Cardiomyopathy Registry (PCMR) database | 1,803 patients; mean age 5.3 ± 6.1 years | <18 years of age with DCM from nearly 100 centers | Sudden cardiac death | 5-year SCD incidence rate was 2.4%. 86% of all SCDs occurred in patients who met all of the following: left ventricular (LV) end-systolic dimension *z* score >2.6, age at diagnosis <14 years, and the LV posterior wall thickness to end-diastolic dimension ratio <0.14.  Independent predictors of SCD include echocardiographic features of both LV thinning and dilation, antiarrhythmic therapy within 1 month of diagnosis, and age at diagnosis <14 years. |
| El-Assaad et al199 | 2015 | Multicenter retrospective study from  United Network for Organ Sharing (UNOS) database | 5,072 patients; mean age 6.2 ± 6.5 years; 8% with ICDs | <18 years of age listed for heart transplant | Sudden cardiac death | Incidence of SCD 4%. By multivariate analysis: myocarditis (HR 0.2), Restrictive cardiomyopathy (HR 0.2), and dilated cardiomyopathy (HR 0.3) were associated with lower SCD risk.  ICD at listing was not associated with reduced SCD (*P* = .12), all-cause mortality, or delisting (*P* = .57). |
| Rhee et al200 | 2007 | Multicenter  retrospective study from Pediatric Heart Transplant Study (PHTS) database | 420 patients | ≤18 years of age listed for transplant who died from any cause after listing but prior to heart transplantation; 24 centers | Death from any cause after listing but prior to heart transplantation | Only 32 deaths (1.3% of total listed, 7.6% of total deaths) were sudden or arrhythmic in nature. Those with ischemic cardiomyopathy had an increased risk of SCD (RR 6.92). Incidence of SCD in children awaiting heart transplantation is low. Children with ischemic cardiomyopathy appear to have a higher risk of SCD and may benefit from ICD therapy. |
| **Congenital Heart Disease** | | | | | | |
| **Study author and reference** | **Year** | **Study type** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Silka et al129 | 1993 | Multicenter retrospective study | 125 patients; 18% patients with  CHD; mean age 14.5 years | <20 years of age at the time of ICD implantation | ICD therapy and all-cause mortality | Secondary prevention ICDs; 59% received appropriate ICD shock following ICD implant.  Impaired ventricular function was the primary factor correlated with mortality post ICD implant. |
| Berul et al130 | 2008 | Multicenter retrospective study | 443 patients; 46% with CHD; median age 16 years | Pediatric and ACHD patients with ICDs | ICD shock and device complications | Frequent inappropriate (21%) and appropriate (26%) shocks in diverse disease. Lead failure was the most common cause of inappropriate shock. |
| Von Bergen et al131 | 2011 | Multicenter retrospective study | 210 patients; 28% with CHD; median age 15 years | <30 years of age with ICD | Incidence and time to ICD shock compared to disease substrate | During 5-year follow-up, appropriate ICD shock incidence was 52% for secondary prevention vs. 15% for primary prevention. Overall, 25% of patients received at least one inappropriate ICD shock. |
| Jordan et al211 | 2014 | Multicenter retrospective registry study from National Cardiovascular Data Registry (NCDR) | 3,139 patients with CHD and 1,601 patients <21 years of age; mean age 12.6 ± 7.7 years | Pediatric and ACHD patients with ICDs | n/a | Descriptive statistics on demographics and ICD lead implantation characteristics in pediatric patients <21 years of age and patients with CHD of all ages. Primary prevention ICDs implanted in 61.9% CHD patients and 58.4% pediatric patients. CHD and pediatrics groups had similar rates of transvenous (97%) and nontransvenous (3%) leads, but nontransvenous lead patients were younger, with higher rates of transposition of the great vessels and common ventricle patients. |
| Dechert et al212 | 2016 | Single-center retrospective study | 131 patients; 57 with CHD (44%); median age 16 years | Pediatric and ACHD patients with ICDs | Time to CD system revision | 33% patients underwent ICD revisions. Estimated rate of 70 revisions/1,000 patient-years of follow-up. ICD system revision is high in pediatric patients and occurs at a rate similar to the rate of receiving appropriate therapy. |
| Kalra et al214 | 2012 | Single-center retrospective study | 79 patients; median age 22 years (range 17–27) | Patients with CHD and ICD | Antitachycardia pacing or ICD shock | ATP decreases risk of both appropriate and inappropriate shocks in patients with CHD. Rate of VT termination was 88% although the rate of inappropriate ATP therapy was high (39%).  . |
| Radbill et al218 | 2010 | Single-center retrospective study | 117 patients:  39 nonTV ICD; mean age 7 years  78 TV ICD;  mean age 20 years | ICD: either TV or non-TV system | Survival of nontransvenous ICD systems compared to transvenous ICDs | Single-center evaluation showing nontransvenous ICDs have shorter system survival. Non-TV system survival was 49% at 3 years post-implant. |

**Table S4. Evidence for Insertable Cardiac Monitors**

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| **Study author** | **Year** | **Study type** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Babikar et al224 | 2008 | Single-center retrospective study | 23 patients | All patients with ICM implant between 2000 and 2005 | Automatic or manually activated ICM event | ICM yielded arrhythmic or nonarrhythmic diagnosis in 15/23 of patients. 8/15 with symptom recurrence had arrhythmia recorded. |
| Bezzerides et al225 | 2019 | Single-center retrospective study | 133 patients; 34 with CHD; 50 with inherited arrhythmia; 22 with cardiomyopathy | All patients with ICM implant between 2014 and 2017 | Automatic or manually activated ICM event | Median time to diagnosis was 4.5 months, occurring in 78 patients (59%). Cardiac device implantation occurred in 17 patients (22%), change in medication in 9 (12%), and electrophysiology study/ablation in 5 (6%). LINQ-ILR was explanted in 42 patients (54%). ICM-related skin infection or erosion occurred in 5 patients. |
| Placidi et al226 | 2016 | Single-center retrospective study | 21 patients | All patients with ICM implant between 2014 and 2015 | Automatic or manually activated ICM event | Median follow-up 10 months, diagnostic yield was 47%. 8 symptomatic patients: no arrhythmias in 6 and significant sinus pauses in 2. In 13 asymptomatic patients, 2 patients had significant arrhythmias. Pacemaker implanted in 1 patient. ICM pocket infection in 2 patients (9%). |
| Avari Silva et al227 | 2016 | Single-center retrospective study | 20 patients; 8 with  LQTS; 9 with CPVT; 1 with BrS; 1 with ARVC | All patients with inherited arrhythmias syndrome and ICM implant 2008–2015 | Automatic or manually activated ICM event | Majority (52%) of symptomatic events in LQTS patients, with only 1 tracing yielding actionable data. Automatic transmissions mostly seen in the CPVT patients (81%), with 21% yielding actionable data. ICM findings escalated therapies in 30% of subjects. |
| Rossano et al230 | 2003 | Three-center retrospective study | 21 patients | All patients with ICM <25 years old and  follow-up >1 month | Automatic or manually activated ICM event | Median follow-up 8.4 months:14 patients had symptoms. Symptom-rhythm correlation was possible in all 14 patients: SVT 4, VT 2, torsades de pointes 1, asystole 1, junctional bradycardia 1, and sinus rhythm 5. One infant with acute life-threatening events with ischemic changes on ICM recording ultimately lead to diagnosis of Munchausen syndrome by proxy. |
| Al Dhahri et al232 | 2009 | Two-center retrospective study | 42 patients divided into high and low risk groups | All patients with ICM from 1998 to 2006 | Automatic or manually activated ICM event | Diagnostic yield of ICM in defining cause of symptoms in 27/42 (64%) over median follow-up 19 months. ICM complications: wound infection n = 2. Both groups reported similar frequency of symptoms, but low-risk patients were more likely to have ICM confirmed. |
| Frangini et al233 | 2008 | Single-center retrospective study | 27 patients | All patients with ICM between 1998 and 2007 | Automatic or manually activated ICM event | Median follow-up of 3 months; 17 had symptomatic events. Asystole or transient AV block (n = 2), sinus bradycardia (n = 6), or normal sinus rhythm (n = 8). Automatic detection in 10 asymptomatic patients showed intermittent AV block or long pauses (n = 3). ICM diagnostic in 95% of symptomatic patients and in 30% of asymptomatic patients. |
| Kenny et al234 | 2009 | Single-center retrospective study | 22 patients | Patients with CHD and ICM implant between 2000 and 2007 | Automatic or manually activated ICM event | Over a median follow-up of 19 months, VT (n = 2) and SND (n = 1) were detected resulting in ICD in 2 patients and DDD pacemaker in 1 patient. Positive or negative arrhythmic diagnosis was reached in 71%. ICM particularly beneficial in patients with neurodevelopmental delay. |

**Table S5. Evidence for CIED Lead Management**

|  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- |
| **Study author** | **Year** | **Study type** | **Patient population** | **Inclusion criteria** | **Endpoints** | **Results/findings** |
| Atallah et al239 | 2013 | Multicenter registry | 878 patients | Consecutive patients both with and without CHD undergoing ICD implantation | ICD lead failure | 14% ICD lead failure at a mean lead age of 2.0 ± 1.4 years, compounded by high failure rates of Fidelis leads. Younger implantation age was an independent predictor for lead failure. Lead age was an independent predictor of need for advanced extraction techniques. |
| Cecchin et al240 | 2010 | Single-center cohort study | 144 patients (203 leads) | Consecutive patients both with and without CHD undergoing lead extraction  (2002–2008) | Successful lead extraction and complication rates | Successful extraction in 80% of all leads and 94% of leads undergoing complex extraction. Older lead age, ventricular leads, and polyurethane insulation were independent predictors of decreased likelihood of simple extraction. 2.7% incidence of both major and minor complications. |
| Mah et al241 | 2018 | Single-center observational study | 145 patients | All patients with epicardial leads undergoing angiography or CT | Coronary compression | Coronary compression noted in 5.5% of patients with epicardial leads, 75% of whom were symptomatic. CT scan had 100% sensitivity and 93% specificity, while angiography had 86% sensitivity and 100% specificity. |
| Fender et al242 | 2017 | Single-center retrospective case control study | 40 CHD patients (77 leads); 80 controls (146 leads) | All patients undergoing lead extraction of leads older than 1 year  (2001–2014) | Successful lead extraction and complication rates | There was no significant difference in extraction techniques between the two groups, with complete extraction achieved in 94% of patients in both those with CHD and those without CHD. No significant difference in complication rates between the two groups. |
| McCanta et al243 | 2013 | Single-center retrospective case control study | 22 CHD patients (35 leads); 22 controls (37 leads) | All patients undergoing laser lead extraction  (2002–2010) | Successful lead extraction and complication rates | Laser lead extraction was deemed successful in 74% of leads in patients with CHD vs. 92% in controls without CHD (*P* = .02). Majority that failed with laser extraction were successfully extracted by femoral snare or mechanical rotational devices. No complications were noted in the CHD group. |
| Moak et al244 | 2006 | Single-center retrospective review | 25 patients (43 leads) | All patients undergoing laser lead extraction | Successful lead extraction and complication rates | Lead removal was complete in 91% of leads and partial for 9% of leads. Major complications noted in 2 (6.9%) patients. |
| Gourraud et al246 | 2018 | Single-center prospective registry study | 49 patients (71 procedures/121 leads) | Consecutive patients with CHD undergoing lead extraction | Successful lead extraction and complication rates | Complete lead extraction was achieved in 92% of all leads. Lead duration and number of previous cardiac surgeries were predictive of failure. Subpulmonary AV valve regurgitation was a prevalent complication in those with transposition of the great arteries. No deaths noted. |
| Bar-Cohen et al249 | 2006 | Single-center prospective study | 85 patients | Consecutive patients undergoing repeat pacemaker procedures  (2002–2004) | Venous obstruction after transvenous lead implant | Complete venous obstruction seen in 13% and partial obstruction seen in 12% of patients. No significant differences between obstructed and nonobstructed patients in relation to age, size, growth, or lead factors. |
| Janson et al255 | 2014 | Single-center retrospective review | 101 patients (120 leads) | Consecutive patients undergoing transvenous ICD implantation  (1995–2011) | ICD lead failure | 21% ICD failure with median follow-up of 28.7 months. Incidence of failure of 5.6% per year, compounded by high failure rates of Fidelis lead. Age and growth were not significantly associated with lead failure. |