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EHRA POSITION PAPER

Arrhythmias in congenital heart disease: a position paper of the European Heart Rhythm Association (EHRA), Association for European Paediatric and Congenital Cardiology (AEPC), and the European Society of Cardiology (ESC) Working Group on Grown-up Congenital Heart Disease, endorsed by HRS, PACES, APHRS, and SOLAECE

Antonio Hernández-Madrid[1]*†, Thomas Paul[2]†, Dominic Abrams[3], Peter F. Aziz[4], Nico A. Blom[5,6], Jian Chen[7], Massimo Chessa[8], Nicolas Combes[9], Nikolaos Dagres[10], Gerhard Diller[11], Sabine Ernst[12], Alessandro Giamberti[13], Joachim Hebe[14], Jan Janousek[15], Thomas Kriebel[16], Jose Moltedo[17], Javier Moreno[1], Rafael Peinado[18], Laurent Pison[19], Eric Rosenthal[20], Jonathan R. Skinner[21], and Katja Zeppenfeld[22]

Reviewers: Christian Sticherling (EHRA review coordinator)[23], Joseph Kautzner[24], Erik Wissner[25], Philipp Sommer[26], Dhiraj Gupta[27], Tamas Szili-Torok[28], Shigeru Tateno[29], Armando Alfaro[30], Werner Budts[31], Pastora Gallego[32], Markus Schwerzmann[33], Ornella Milanesi[34], Georgia Sarquella-Brugada[35], Laszlo Kornyei[36], Narayanswami Sreeram[37], Fabrizio Drago[38], and Anne Dubin[39]

[1]Department of Cardiology, Arrhythmia Unit, Ramón y Cajal Hospital, Alcalá University, Carretera Colmenar Viejo, km 9, 100, 28034 Madrid, Spain; [2]Department of Pediatric Cardiology and Intensive Care Medicine, Georg August University Medical Center, Robert-Koch-Str. 40, D-37075 Göttingen, Germany; [3]PACES (Pediatric and Congenital Electrophysiology Society) Representative, Department of Cardiology, Boston Childreńs Hospital, Boston, MA, USA; [4]HRS Representative, Pediatric Electrophysiology, Cleveland Clinic Children's, Cleveland, OH, USA; [5]Department of Pediatric Cardiology, Leiden University Medical Center, Leiden, The Netherlands; [6]Academical Medical Center, Amsterdam, The Netherlands; [7]Department of Heart Disease, Haukeland University Hospital, Bergen, Norway; [8]Pediatric and Adult Congenital Heart Centre-University Hospital, IRCCS Policlinico San Donato, Milan, Italy; [9]Arrhythmia Unit, Department of Pediatric and Adult Congenital Heart Disease, Clinique Pasteur, Toulouse, France; [10]Department of Electrophysiology, University Leipzig Heart Center, Leipzig, Germany; [11]University Hospital Muenster, Muenster, Germany; [12]Royal Brompton and Harefield Hospital, London, UK; [13]Congenital Cardiac Surgery Unit, Policlinico San Donato, University and Research Hospital, Milan, Italy; [14]Center for Electrophysiology at Heart Center Bremen, Bremen, Germany; [15]2nd Faculty of Medicine, Children's Heart Centre, Charles University in Prague and Motol University Hospital, Prague, Czech Republic; [16]Westpfalz-Klinikum Kaiserslautern, Children's Hospital, Kaiserslautern, Germany; [17]SOLAECE Representative, Head Pediatric Electrophysiology, Section of Pediatric Cardiology Clinica y Maternidad Suizo Argentina, Buenos Aires, Argentina; [18]Department of Cardiology, Arrhythmia Unit, Hospital la Paz, Madrid, Spain; [19]Department of Cardiology, Maastricht University Medical Center, Maastricht, Netherlands; [20]Consultant Paediatric and Adult Congenital Cardiologist, Evelina London Children's Hospital, Guy's and St Thomas' Hospital Trust, London, UK; [21]APHRS Representative, Paediatric and Congenital Cardiac Services Starship Childreńs Hospital, Grafton, Auckland, New Zealand; [22]Department of Cardiology, Leiden University Medical Center, Leiden, The Netherlands; [23]University Hospital Basel, Petersgaben 4, CH-4031 Basel, Switzerland; [24]Institute For Clinical and Experimental Medicine, Prague CZ-140 21, Czech Republic; [25]University of Illinois at Chicago, 840 S. Wood St., 905 S (MC715), Chicago, IL 60612, USA; [26]Heart Center Leipzig, Struempellstr. 39, 04289 Leipzig, Germany; [27]Consultant Electrophysiologist Liverpool Heart and Chest Hospital, Honorary Senior Lecturer Imperial College

* Corresponding author. Tel: 00349133681813; fax: 34913368183. *E-mail address*: antoniomadri@gmail.com

† The first two authors are the co-first authors of this study.

London and University of Liverpool, Liverpool, UK; [28]Erasmus MC, Office Ba-577, 3015 CE Rotterdam, Netherlands; [29]Chiba Cerebral and Cardiovascular Center, 575 Tsurumai, Ichihara, Chiba, Japan; [30]Hospital La Católica Guadalupe, San José, Costa Rica; [31]UZ Leuven, Campus Gasthuisberg, Herestraat 49, B-3000 Leuven, Belgium; [32]Hospital Virgen Del Rocio-Macarena, Sevilla, Spain; [33]INSELSPITAL, Universitätsspital Bern, Universitätsklinik für Kardiologie, Zentrum für angeborene Herzfehler ZAH, 3010 Bern, Switzerland; [34]Department of Woman and Child's Health, University of Padua, Padua Italy; [35]Pediatric Arrhythmias, Electrophysiology and Sudden Death Unit, Department of Cardiology, Hospital Sant Joan de Déu, Barcelona - Universitat de Barcelona, Passeig Sant Joan de Déu, 2, 08950 Esplugues, Barcelona, Catalunya, Spain; [36]Gottsegen Gyorgy Orszagos Kardiologiai, Pediatric, Haller U. 29, Budapest HU-1096, Hungary; [37]Department of Pediatric Cardiology, University Hospital Of Cologne, Kerpenerstrasse 62, Cologne DE-50937, Germany; [38]IRCCS Ospedale Pediatrico Bambino Gesù, Piazza Sant'Onofrio 4, 00165 Roma; and [39]Division of Pediatric Cardiology, 750 Welch Rd, Suite 321, Palo Alto, CA 94303, USA

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Abstract

The population of patients with congenital heart disease (CHD) is continuously increasing with more and more patients reaching adulthood. A significant portion of these young adults will suffer from arrhythmias due to the underlying congenital heart defect itself or as a sequela of interventional or surgical treatment. The medical community will encounter an increasing challenge as even most of the individuals with complex congenital heart defects nowadays become young adults. Within the past 20 years, management of patients with arrhythmias has gained remarkable progress including pharmacological treatment, catheter ablation, and device therapy. Catheter ablation in patients with CHD has paralleled the advances of this technology in pediatric and adult patients with structurally normal hearts. Growing experience and introduction of new techniques like the 3D mapping systems into clinical practice have been particularly beneficial for this growing population of patients with abnormal cardiac anatomy and physiology. Finally, device therapies allowing maintanence of chronotropic competence and AV conduction, improving haemodynamics by cardiac resynchronization, and preventing sudden death are increasingly used. For pharmacological therapy, ablation procedures, and device therapy decision making requires a deep understanding of the individual pathological anatomy and physiology as well as detailed knowledge on natural history and long-term prognosis of our patients. Composing expert opinions from cardiology and paediatric cardiology as well as from non-invasive and invasive electrophysiology this position paper was designed to state the art in management of young individuals with congenital heart defects and arrhythmias.

Keywords

Congenital heart disease • Arrhythmia • Sudden cardiac death • Heart failure • Macroreentry tachycardia • Atrioventricular block • Bradycardia • Implantable cardioverter-defibrillator • Pacemaker • Cardiac resynchronization therapy • Ablation • European Heart Rhythm Association position paper

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Introduction

The purpose of the present consensus statement was to summarize knowledge and provide recommendations on diagnosis and treatment of arrhythmias in patients with congenital heart defects (CHD), because, in many cases, the anatomy and management of arrhythmias in adult patients cannot directly be applied to patients with CHD.[1–4] This position paper mainly addresses arrhythmias in adult CHD. A consensus paper on paediatric CHD has been published in 2013.[4]

Evidence review

Members of the Task Force were asked to perform a detailed literature review, weigh the strength of evidence for or against a particular treatment or procedure, and include estimates of expected health outcomes where data exist. Patient-specific modifiers, comorbidities, and issues of patient preference that might influence the choice of particular tests or therapies were considered, as are frequency of follow-up and cost effectiveness. In controversial areas, or with regard to issues without evidence other than usual clinical practice, a consensus was achieved by agreement of the expert panel after discussions.

This document was prepared by the Task Force with representation from European Heart Rhythm Association (EHRA), Association for European Paediatric and Congenital Cardiology (AEPC), European Society of Cardiology (ESC) Working Group on Grown-up Congenital Heart Disease, Heart Rhythm Society (HRS), Pediatric and Congenital Electrophysiology Society (PACES), Asia-Pacific Heart Rhythm Society (APHRS), and Sociedad Latinoamericana de Estimulación Cardíaca y Electrofisiología (SOLAECE).

The document was peer-reviewed by official external reviewers representing EHRA, ESC WG on Grown up Congenital Heart Disease, AEPC, PACES, HRS, APHRS, and SOLAECE.

Consensus statements are evidence based, and derived primarily from published data. However, some statements in a consensus document are 'experienced based' and relate to a low level of evidence (LE). Furthermore, as in our article, evidence based data from 'the regular adult population' are frequently extrapolated to 'CHD patients'. In contrast to guidelines, we have opted for an easier and

Table 1 Scientific rationale of recommendations

Definitions where related to a treatment or procedure	Consensus statement	Symbol
Scientific evidence that treatment or procedure is beneficial and effective. Requires at least one randomized trial, or is supported by strong observational evidence and authors' consensus (as indicated by an asterisk)	Recommended/ indicated	
General agreement and/or scientific evidence favour usefulness/ efficacy of treatment or procedure. May be supported by randomized trials based on small number of patients or not widely applicable	May be used or recommended	
Scientific evidence or general agreement not to use or recommend treatment or procedure	Should NOT be used or recommended	

This categorization of our consensus document should not be considered as being directly similar to that used for official society guideline recommendations which apply a classification (I–III) and level of evidence (A, B, and C) to recommendations.

user-friendly system of ranking using 'coloured hearts' that should allow physicians to easily assess current status of evidence and consequent guidance (*Table 1*).

Thus, a 'green heart' indicates a recommended statement or recommended/indicated treatment (or procedure) and is based on at least one randomized trial, or is supported by large observational evidence that it is beneficial and effective. A 'yellow heart' indicates general agreement and/or scientific evidence favouring a statement or the usefulness/efficacy of a treatment or procedure. A yellow heart may be supported by randomized trials based on small number of patients or not widely applicable. Treatment strategies for which there have been scientific evidence that they are potentially harmful and should not be used are indicated by a 'red heart'.

European Heart Rhythm Association grading of consensus statements does not have separate definitions of LE. The categorization used for consensus statements (used in consensus documents) should not be considered as being directly similar to that used for official society guideline recommendations which apply a classification (I–III) and LE (A, B, and C) to recommendations in official guidelines.

Finally, this is a consensus document that includes evidence and expert opinions from several countries. The pharmacologic and non-pharmacologic antiarrhythmic approaches discussed may, therefore, include drugs or devices that do not have the approval of governmental regulatory agencies in all countries.

Relationships with industry and other conflicts of interest

It is EHRA/ESC policy to sponsor position papers and guidelines without commercial support and all members volunteered their time. Thus, all members of the writing group as well as reviewers have disclosed any potential conflict of interest in detail at the end of this document.

Scope of the consensus document

Patients with congenital heart disease constitute a heterogeneous population with unique needs, concerns, and challenges. Current survival rates of congenital heart disease patients have improved, allowing for an ever-growing population of adult survivors.

Arrhythmias figure foremost among the issues encountered in this population and are the leading cause of morbidity and mortality. Several forms of CHD predispose to arrhythmias even without any surgical intervention due to abnormalities of the conduction system, intrinsic structural pathology, and impact of pre- or post-operative cyanosis and volume-/pressure-overload. In general, surgery for congenital heart defects may result in sinus node dysfunction, atrioventricular (AV) block and a variety of supraventricular and ventricular tachyarrhythmias including the risk of sudden cardiac death (SCD). Arrhythmia treatment in patients with CHD requires high expertise of non-invasive and invasive electrophysiology (EP) combined with a thorough knowledge and understanding of the particular congenital heart defect encountered including anatomy, pathophysiology, and surgical and/or interventional treatment.

Arrhythmias in congenital heart disease: general considerations

Arrhythmia substrates in congenital heart disease

In general, arrhythmias in patients with CHD may be due to abnormal anatomy or congenitally displaced or malformed sinus nodes or AV conduction systems, abnormal haemodynamics, primary myocardial disease, hypoxic tissue injury, residual or post-operative sequelae, and genetic influences.[2,3] An overview is provided in *Figure 1*.

Congenital substrates for arrhythmias in congenital heart defects

Sinus node

Abnormal position of the sinus node may be found not only in rare forms of malformed hearts with juxtaposition of the left atrial appendages but may also be present in patients with sinus venosus defects of superior vena cava (SVC) type. In hearts with left atrial isomerism, sinus node tissue may be completely absent or may be found as remnants in the inferior atrial wall near the AV junction, whereas in right isomerism two sinus nodes may be present.[4] An abnormal dysfunctional sinus node can be found in usual position in patients with absence of the right superior caval vein.[5]

Atrioventricular block

Atrioventricular block in patients with congenitally malformed hearts is most often observed in patients with congenitally corrected transposition of the great arteries (CC-TGA) and in individuals with isomeric arrangement of the atrial appendages, i.e. left isomerism, due to displacement of the specialized conduction system with abnormal

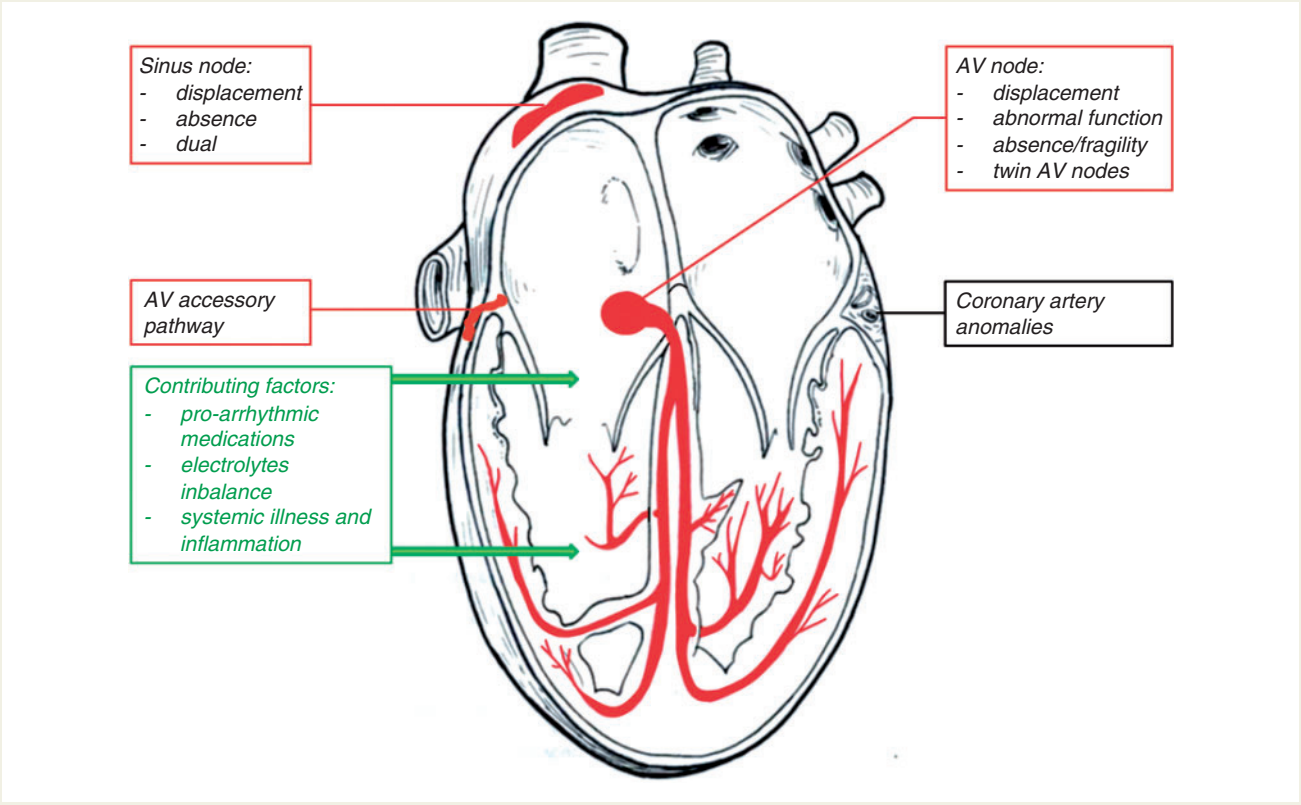

Figure 1 Pre-operative factors resulting in supraventricular and ventricular arrhythmias in congenital heart disease patients. AV, atrioventricular.

development of the central fibrous body.[6] The life-long risk of complete AV block in patients with CC-TGA is approximately 2% per year and may reach 50% at 50 years of age.[7]

Twin atrioventricular nodes

Two distinct AV nodes ('twin AV nodes', Mönckeberg sling) may be present in congenital heart defects with AV discordance ([S, L, L] or [I, D, D]) with a malaligned complete AV canal defect and in right and left atrial isomerism giving rise to paroxysmal supraventricular tachycardia (SVT).[8]

Accessory atrioventricular connections

Accessory AV connections may be present in a variety of patients with CHD with Ebstein's anomaly of the tricuspid valve being the entity with the highest prevalence. Multiple accessory pathways are often found including special variants with solely anterograde decremental conduction as Mahaim type pathways. Additional types of congenital heart defects with increased prevalence of accessory pathways include heterotaxy syndromes, CC-TGA, AV septal defects, and univentricular hearts.[4]

Atrioventricular nodal reentrant tachycardia

Atrioventricular nodal reentrant tachycardia (AVNRT) may develop in patients with various types of congenital heart defects at any age. Depending on individual anatomy, localization of the specialized conduction system may be less predictable, particularly in patients with single ventricle (Single V) physiology.[9]

Post-operative substrates for arrhythmias in congenital heart defects

Sinus node dysfunction and atrioventricular block

Direct injury to the sinus node may occur by surgical incisions or suturing in the high right atrium. Atrial switch procedures for patients with d-transposition of the great arteries, Fontan procedures for patients with Single V physiology and rerouting of partial anomalous pulmonary venous return are the most frequent surgical interventions resulting in chronotropic incompetence. More than 50% of survivors from the Mustard operation, for instance, have lost reliable sinus rhythm by adulthood.[10] Sinus node dysfunction is often associated with limited exercise capacity and aggravation of AV valve regurgitation which in turn may contribute to development of macroreentrant atrial tachycardia (MRAT).[4,10] The conduction system is particularly susceptible to injury during surgical and catheter procedures in patients with ASDs and CC-TGA.[10] In addition, perimembranous ventricular septal defect is associated with increased risk for AV block related to percutaneous closure at younger age.

Macroreentrant atrial tachycardias

Macroreentrant atrial tachycardia include incisional intraatrial tachycardias and atrial flutter (AF). Atrial tachycardias (AT) are frequently encountered after surgical repair of a wide variety of congenital heart defects. Incisional atrial arrhythmias may occur even 10 to 15 years post-surgery. Anatomical substrates of reentrant ATs are areas with pre-existing intraatrial conduction abnormalities as well as suture lines, scar tissue, and/or prosthetic material. A stable reentrant circuit may occur

around these structures of electrical isolation and anatomic obstacles, such as the orifices of the great veins and the AV annuli, often utilizing a protected zone of atrial tissue with or without slow conduction. Over 60% of these atrial reentrant circuits involve the cavo-tricuspid isthmus (CTI). Scar-related reentrant AT and AF are the most common types of SVT in adult CHD patients. Typical congenital heart defects include atrial septal defects (ASD), tetralogy of Fallot (TOF), Ebstein anomaly of the tricuspid valve, Single V physiology after Fontan procedure, and d-transposition of the great arteries after atrial switch operation[1,11] (*Figure 2*).

Focal atrial tachycardia

This arrhythmia mechanism which is predominantly seen in patients post-total cavopulmonary Fontan connection and after atrial switch procedures for D-TGA is highly variable, encompassing simple, and more complex substrates, such as enhanced automaticity or micro-reentry with a focal origin [non-automatic focal AT (NAFAT)].[12] Discrete areas of heterogenous conduction can be detected as the source of these tachycardias.[13,14]

Accessory pathways after Björk-Fontan modification

Occasionally, accessory AV pathways may be created by means of surgical intervention in patients with tricuspid atresia, connecting the right atrial appendage to the right ventricular outflow tract (Fontan-Björk procedure).[15]

Ventricular arrhythmias

Stable monomorphic ventricular tachycardia

Stable monomorphic ventricular tachycardia (VT) in patients with CHD is based upon morphological/anatomical variants of the heart defect itself or to ventricular incisions and prosthetic material like tubes and patches that allow initiation and perpetuation of a ventricular macroreentrant circuit. These tachycardias often depend on critical isthmuses within the right ventricular outflow tract bordered by patches or scar after ventriculotomy incisions. A unique example is unoperated/native and post-operative TOF and its variants.[16–18] Stable monomorphic VT can be fatal and a cause of SCD dependent on cardiac function and VT cycle length as in post-operative TOF patients even with preserved cardiac function. Incidence of sustained VT/ventricular fibrillation (VF) has been reported at 14.6% in 556 adult patients after TOF repair.[19]

Polymorphic ventricular tachycardia

Fast polymorphic VT and VF with the risk of SCD may develop in patients with severely diseased ventricular myocardium, significant fibrosis, and myocardial disarray. Typical congenital heart defects include left ventricular outflow tract obstructive lesions, d-transposition of the great arteries after atrial switch procedure with failing systemic right ventricle (Syst RV), TOF with significantly impaired right ventricular function, Eisenmenger syndrome, and univentricular hearts with a Fontan circulation.[4,18,20]

Work-up of patients with congenital heart disease and arrhythmias

Early diagnosis and electrocardiogram (ECG) documentation of the underlying arrhythmia is of paramount importance for long-term treatment success, as some arrhythmias have a propensity to degenerate into more difficult to treat arrhythmias [e.g. AT into atrial fibrillation (AF)]. The first step is consultation to a paediatric or congenital

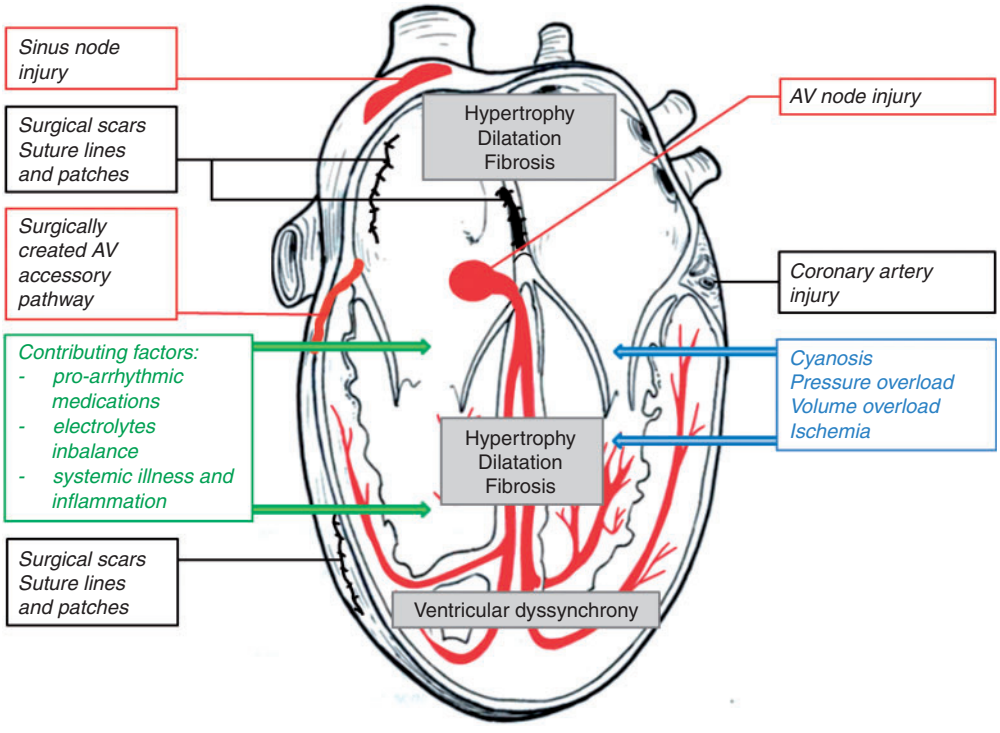

Figure 2 Post-operative factors resulting in supraventricular and ventricular arrhythmias in congenital heart disease patients. AV, atrioventricular.

cardiologist to exclude haemodynamic or anatomical triggers and to initiate pharmacological treatment where applicable. Timely referral to a centre with a multidisciplinary team and expertise in CHD patients and CHD-related arrhythmia is mandatory.

Acute assessment of the congenital heart disease patient presenting with arrhythmia

The key task for the physician taking care of the CHD patient presenting with an arrhythmia is to assess the clinical impact of the arrhythmia and to decide if immediate treatment (e.g. direct current cardioversion) is necessary. Some CHD patients may present in AT with haemodynamic compromise due to a combination of rapid AV conduction and impaired ventricular function/haemodynamics. Particularly, patients with baffle obstruction after atrial switch procedure may develop significant decrease of ventricular preload in AF with rapid ventricular response, and individuals with residual left or right ventricular outflow tract obstruction and rapid ventricular rhythm are prone to haemodynamic collapse due to low cardiac output requiring immediate appropriate treatment.

Besides recurrent need for direct current cardioversion, ineffective pharmacological treatment and/or intolerance for drugs (e.g. electropysiological side effects or systemic side effects) should prompt referral of the patient to a dedicated ablation centre experienced in the management of patients with CHD. While re-entrant AT in patients with a biventricular heart may be successfully treated by experienced electrophysiological centres non-dedicated for CHD, the same arrhythmia is very likely not to be handled appropriately at that institution in complex CHD patients.[1,4]

Collaboration between specialists in adult congenital heart disease and arrhythmia specialists

In order to obtain the best outcome for a complex CHD patient with arrhythmia, multidisciplinary team discussions are recommended to consider all options available. Collaboration should include discussion about pharmacological management and optimized haemodynamic management. In addition, the congenital cardiac surgeon plays an important role when surgery combining haemodynamic optimization and surgical ablation/device implantation is planned. In some instances, a surgical or a hybrid procedure may be considered, as in patients lacking vascular access to a given target heart chamber like in a patient with both mitral and aortic mechanical valves and LV-VT. Similarly, if access is not easily achieved or if local expertise is not sufficient, early referral of the patient to a dedicated CHD arrhythmia centre is warranted with the ability of special procedures and technologies like remote magnetic navigation, transapical or transventricular access, and baffle puncture. In particular instances, an experienced electrophysiologist and an experienced interventional congenital cardiologist may be required to support a local hospital team if transfer of the patient is not feasible.

General assessment of the congenital heart disease patient with arrhythmias

When assessing a CHD patient with an arrhythmia, a comprehensive assessment of the individual haemodynamics is required. Standard diagnostic procedures include detailed history, complete physical examination, 12-lead ECG, and a transthoracic or transoesophageal echocardiogram in order to establish the function of both the systemic and subpulmonary ventricle and to identify residual significant haemodynamic abnormalities like a paravalvular leak, valvar stenosis, or significant AV or semilunar valve regurgitation (*Table 2*). Echocardiography is often not completely sufficient to delineate individual anatomy and particular lesions like systemic venous baffle obstruction in Mustard and Senning patients. Left and right heart catheterization is frequently required accordingly. In patients with univentricular hearts, cardiac magnetic resonance imaging (MRI) is indicated for reliable assessment of ventricular function. This also applies for TOF patients for evaluation of right ventricular function. Finally, intracardiac thrombus formation needs to be excluded before cardioversion or any intracardiac intervention. Optimization of individual haemodynamics should be sought before/after an intervention.

Table 2 Initial work-up of the CHD patient with arrhythmia

History	Underlying anatomy
	Previous interventions/operations
	Arrhythmia characteristics (onset, duration, symptoms etc.)
	Antiarrhythmic and non-antiarrhythmic medication
	Chronic anticoagulation
	Functional class (NYHA)
Physical examination	Signs of haemodynamic compromise/heart failure
	Scars from previous surgery/vascular access sites
	Implanted device
	Signs of infection, fever
12-lead ECG	Underlying rhythm
	Atrial and ventricular rate, conduction intervals, bundle branch block, ventricular hypertrophy, and repolarisation abnormalities
Transthoracic/ transoesophageal echocardiogram	Size and function of the systemic and subpulmonic ventricle
	Size and volume of cardiac chambers
	Thrombus, residual intracardiac shunt
	Valvular function, gradients, and regurgitation
Cardiac CT/MRI/ haemodynamic catheterization/angiography	For complete assessment of individual anatomy and haemodynamics in univentricular hearts, systemic right ventricle, TOF, and variants
Blood chemistry	Infection signs
	Full blood count, plasmatic coagulation
	Thyroid, liver, and kidney function

CHD, congenital heart disease; CT/MRI, computed tomography/magnetic resonance imaging; ECG, electrocardiogram; NYHA, New York Heart Association; TOF, tetralogy of Fallot.

In any post-operative CHD patient, a careful review of surgical note(s) may offer significant insight into post-operative anatomy and potential arrhythmia substrates like suture lines and atriotomy scars. When planning an ablation procedure or implantation of a cardiac device, patency of vessels allowing access to the heart needs to be established. Especially in patients with multiple previous interventions, vascular access may propose a major hurdle to any invasive arrhythmia management. Appropriate imaging should be performed by echocardiography, angiography, or MRI/computed tomography (CT) scans.

Laboratory studies should focus on any inflammatory process, hyperthyroidism, anaemia, plasmatic coagulation, and kidney and liver function.

Arrhythmia management via implanted devices

If an implanted device is present, device memory should be interrogated to obtain detailed information about arrhythmia onset, mechanism, and duration. Depending on available leads, overdrive options may be considered for atrial arrhythmia. Occasionally, leads (mostly epicardial) may have broken and can not be used for device mediated overdrive pacing anymore.

Imaging requirements for invasive procedures: road mapping

If catheter ablation is planned, adequate imaging of individual anatomy may serve as a roadmap and planning tool for the invasive procedure especially in patients with complex CHD. Imaging should be performed with the least amount of radiation and contrast agents. Computed tomography scans may require extensive contrast exposure as cardiac transit times may be prolonged exposing the patient to increased radiation. Non-contrast MRI uses blood pool imaging. However, artificial structures such as stents, valves, clips, devices and (abandoned) leads, and sternal cerclages may create artefacts. Images need to be critically reviewed to assure that 3D segmentation is feasible for a given target area. Online imaging should be considered with transoesophageal (TOE) or intracardiac echocardiogram (ICE) allowing delineation of individual anatomy and monitoring of the procedure.

Specific arrhythmia types

Supraventricular arrhythmias in patients with congenital heart disease

Almost 50% of all adult CHD patients will be confronted with the occurrence of SVT during their lifetime.[21] Intra-atrial re-entrant tachycardias and AF are the most frequent types of SVT in these patients, but AV re-entrant tachycardia (AVRT) using an accessory pathway and AVNRT are also encountered.[22] Factors such as underlying anatomy, age, and surgical repair technique have an impact on prevalence and arrhythmia substrate.

Accessory pathways and atrioventricular reentrant tachycardias

Ebstein's anomaly is the entity most frequently associated with AV reciprocating tachycardia due to the presence of often multiple and mainly right-sided accessory pathways including pathways of Mahaim type.[23] Up to 20% of Ebstein patients harbour one or more accessory pathways.[24] This unique situation is the consequence of caudal displacement of the septal tricuspid valve leaflet, which is associated with disruption of the central fibrous body and septal AV ring with direct muscular connections.[25] These muscular connections form the substrate for accessory AV connections. The presence of multiple accessory pathways may result in a combination of orthodromic and antidromic AVRT. These types of arrhythmias are also known to occur in CC-TGA associated with Ebstein's malformation of the systemic (tricuspid) AV valve.

Atrioventricular nodal re-entrant tachycardia

Atrioventricular nodal re-entrant tachycardia occurs less frequently in CHD patients.[9,26–29] Symptoms may include palpitations and syncope. Displacement of the specialized conduction system in patients with CHD implies an increased risk of AV block during ablation procedures. Special attention is required in patients with AV septal defects, CC-TGA, heterotaxy syndromes, or dextrocardia (*Figure 3*).

Atrial tachycardias

Atrial tachycardias are common in CHD patients. Incidence is related to complexity of the congenital heart defect, type of surgical procedure, patient age, and interval post-surgery.[25,30] In a series of 38 430 CHD patients, CHD types with the highest prevalence of atrial tachyarrhythmias were Ebstein's anomaly (33%), D-TGA after atrial switch procedure (28%), and ASD (19%).[22]

In patients with simple, unoperated cardiac defects, such as ASD who may remain undiagnosed until late in adult life, atrial tachyarrhythmias are related to the volume load imposed on the right atrium by long-standing left-to-right shunting. Atrial flutter has clearly been associated with significant tricuspid regurgitation in CHD.[30,31] In the majority of patients with CHD, however, ATs result from a combination of haemodynamic factors and surgical scars or prosthetic material used for intracardiac repair[30,32] (*Figure 4*). Atrial tachyarrhythmias, with AF becoming more common, expose these patients also to the inherent risks of thrombus formation and stroke.

Prognosis in patients with ATs and complex CHD is often less benign than in patients without CHD or simple CHD substrates (e.g. MRAT in D-TGA and post-Fontan vs. typical AF in ASD). History of AF has been identified as a risk factor of SCD in complex CHD patients.[21,33] Since most of these subjects, especially young adults, have normal AV nodal function, rapid atrial rates may be transmitted 1:1 from atria to ventricles which may lead to syncope and SCD, particularly when ventricular function is poor, as it is commonly present in patients with a Syst RV.[34]

An ASD is often present in patients with CHD and may contribute to development of atrial tachyarrhythmias.[30] In older patients with unoperated ASD, a history of AF was present in 19% and AF in 61%.[35] Electrophysiological studies in ASD patients without previous episodes of atrial tachyarrhythmias have shown electrical remodelling of the atria with an increase of atrial effective refractory periods and conduction delay at the crista terminalis.[36] In a large meta-analysis of 26 studies including 1841 patients undergoing surgical closure and 945 undergoing percutaneous closure, prevalence of atrial tachyarrhythmias decreased by one-third after closure of the defect.[37]

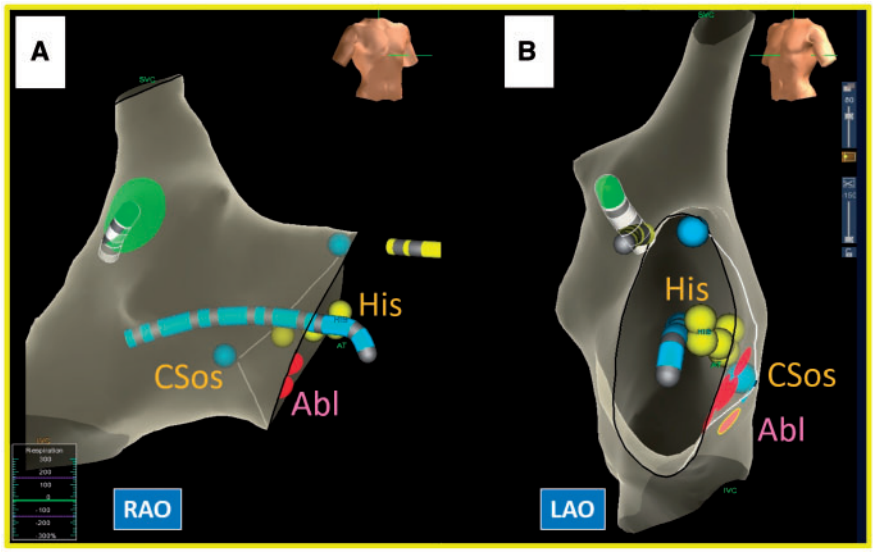

Figure 3 Patient with AV nodal re-entrant tachycardia after surgical repair of complete AV septal defect. Electroanatomical maps (NavX®) of the right atrium in the right anterior oblique view (*A*) and left anterior oblique view (*B*) display typical caudal displacement of the AV node-His bundle, shown by the yellow dots. The triangle of Koch is delineated in white; red dots represent ablation points; blue dots correspond to the tricuspid valve annulus. Abl, ablation; AV, atrioventricular; AVNRT, atrioventricular nodal re-entrant tachycardia; CSos, Coronary Sinus ostium; LAO, left anterior oblique; RAO, right anterior oblique.

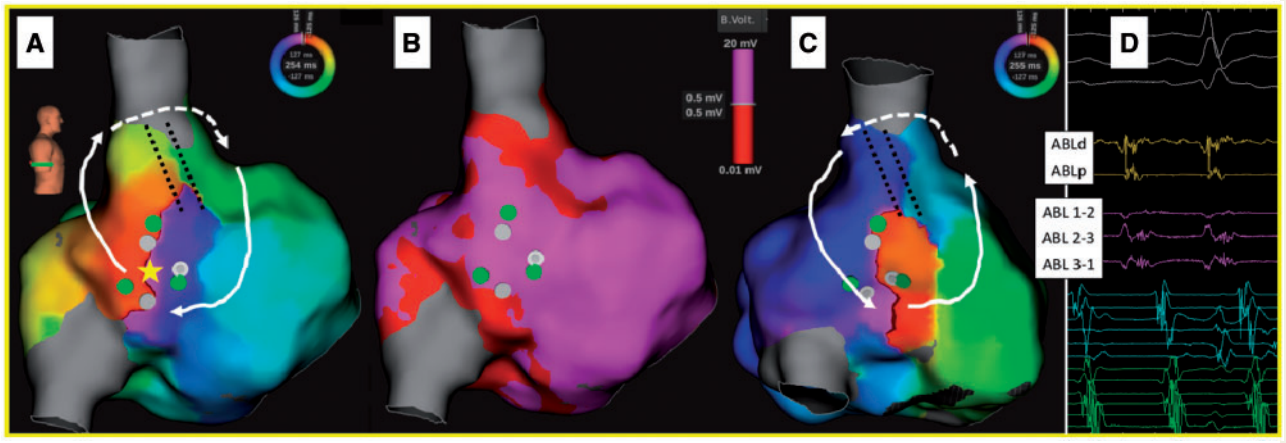

Figure 4 Incisional right atrial macroreentrant tachycardia in a patient after repair of sinus venosus type atrial septal defect. Two clinical atrial tachycardias share the same incisional circuit on the right atrial free wall (RAO view): a re-entrant map of one of the reentrat circuits (*A*) and the corresponding voltage map (*B*); a new reentrant map of the second reentry revolving around the same area, but in a reverse direction (*C*); local fragmented diastolic activity (*D*) at the tip of the ablation catheter at the position marked with an asterisk in Panel *A*. Areas depicted in red represent the scar tissue. Green and white dots represent location of fragmented atrial electrograms in both tachycardias, indicating the same slow conducting zone. RAO, right anterior oblique.

Age plays a major role: older age at the time of closure was one of the main risk factors for post-closure atrial arrhythmias, accompanied by the presence of pre-operative AF or flutter.[38] The long-term outcome after ASD closure at young age is excellent.[39]

The Fontan circulation in patients with a univentricular heart is prone to atrial tachyarrhythmias both located at the lateral right atrial wall[40] and at the CTI. Prevalence of MRAT was higher in the first 2 years after Fontan and increased again gradually in later childhood.[40] The atriopulmonary connection is often associated with a progressive and massive enlargement of the systemic venous atrium. In a contemporary multicentre series, 19% of these patients developed MRAT, with older series showing even higher rates[41] The newer Fontan modifications including intracardiac lateral tunnel, extracardiac lateral tunnel, or extracardiac conduit were significantly less arrhythmogenic,

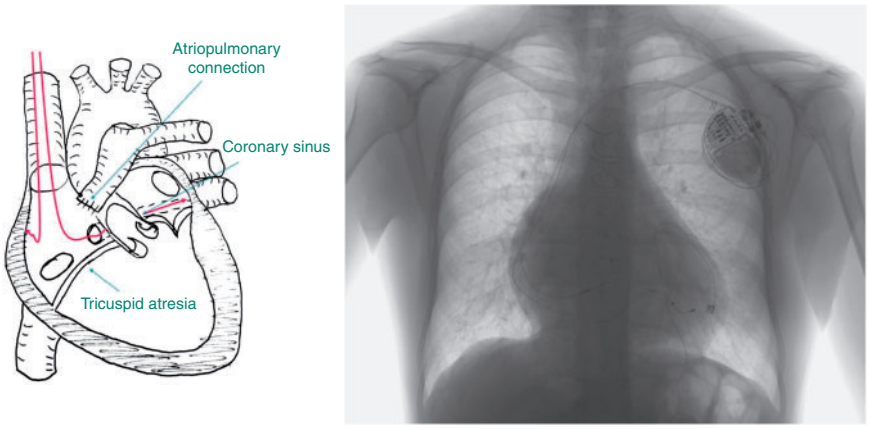

Figure 5 Patient with right ventricular atresia and corrected with the Fontan tecnique. Diagram of the cardiac anatomy of the patient (*A*) and the fuoroscopic image showing the transvenous delivery of the pacemaker ventricular lead into the coronary sinus (*B*).
Photo courtesy of Dra. Inmaculada Sanchet (Ramony cajal hospital).

showing MRAT in a range from 2 to 7%.[41] Another series also demonstrated that a lateral tunnel type Fontan was associated with a significant decrease in arrhythmias as compared to an atriopulmonary Fontan with a 39% vs. 13% 15 years of incidence of SVT.[41] Improved results may be explained by less atrial surgery and improved haemodynamics of the modern Fontan modifications.

Patients after atrial switch procedures (Mustard/Senning) are also highly prone to develop MRAT. Prevalence may reach up to 27% at 20 years post-surgery, often revolving around the tricuspid annulus.[42,43] The arterial switch procedure was less proarrhythmogenic with an AF rate of 5% in adulthood.[44]

It has been hypothesized that development of MRAT may be prevented at cardiac surgery by extending atriotomy incisions to anatomical obstacles of electrical isolation, avoiding development of new reentry circuits.

Finally, it should be emphasized that surface ECG in complex CHD patients may be difficult to interpret with regard to ATs as low amplitude atrial signals are often present. These arrhythmias may therefore be missed at normal ventricular rates and only slightly irregular rhythm.

Atrial fibrillation

Atrial fibrillation has been reported with increasing prevalence in patients with congenital heart defects. It is of note that AF occurs at a younger age in CHD patients than in the general population. A pre-existing organized AT or frequent atrial ectopy may be present in almost two-thirds of these patients.[45,46] Atrial fibrillation in CHD patients considerably increases the risk of stroke and heart failure. In addition to the known risk factors for AF in the general population as age, arterial hypertension, New York Heart Association (NYHA) functional class, obesity and diabetes, adults with CHD carry the additional factors of the underlying heart defect as myocardial scarring and fibrosis, patches/scars from heart surgery, and potential chronic oxygen desaturation.[46]

Atrial fibrillation is predominantly seen in adult CHD patients with left heart obstructive lesions, chronic left atrial dilatation, pre-existing pulmonary hypertension, Fontan procedures, TOF, and after late ASD closure.[35,40,41] It is of note, that a significant number of patients with congenital heart defects may develop AF later in life after successful ablation of reentrant AT. As more patients with CHD survive to older ages, AF will attain considerable attention in the future.[47]

Chronic oral anticoagulation should be considered in adult congenital heart patients with history of AF/AF after intracardiac repair, with cyanosis, after Fontan palliation and in individuals with a Syst RV, apart from those evaluated with the CHA_2DSc score. Antiarrhythmic therapy with Class IC antiarrhythmic agents and amiodarone is effective in almost 50% of the patients, but systemic side effects and proarrhythmia are of concern and require careful surveillance.[1,48] Direct current cardioversion with appropriate anticoagulation is safe and effective for patients with CHD, even in the presence of an intracardiac shunt and spontaneous contrast on TOE.[49] Haemodynamically, rhythm control may be superior to rate control for AF in congenital heart patients, but up to now, there are no data available supporting this hypothesis. Pulmonary vein isolating catheter ablation procedures for drug-refractory AF may be beneficial in selected patients, but experience is limited.[50–52] The same applies to surgical Maze procedures.[53] Prophylactic arrhythmia surgery has been advocated to be incorporated into reparative open heart procedures for CHD patients, but data on efficacy are lacking.[54] In symptomatic patients with AF refractory to pharmacologic and ablation therapy or failure to adequately control the average heart rate, AV nodal ablation and post-ablation pacing may be considered as third-line therapy.[30,55]

Ventricular arrhythmias and sudden cardiac death in patients with congenital heart disease

Ventricular tachycardia

Ventricular ectopy and non-sustained VTs (VTs < 30 s; NSVT) are relatively common in adults with CHD. The relationship between NSVT and sustained VT or sudden death is still a matter of debate in these patients. In TOF, NSVT may be associated with SCD although

there is conflicting data.[19,56,57] In CHD patients with implantable cardioverter-defibrillator (ICD) for primary prevention, the same association was found for symptomatic NSVT in a multivariable analysis,[58] but results from other studies are equivocal.[59,60]

Sustained ventricular arrhythmias in adults with congenital heart disease (ACHD) encompass monomorphic VT, polymorphic VT, and VF.[57,61–63] Based on distribution of CHD across ICD studies and on population based series on presumed arrhythmic death in CHD, sustained ventricular arrhythmias are more likely to occur in patients with TOF, complex forms of d-TGA, Syst RVs and left ventricular outflow tract obstructive lesions.[59,63–65]

In a multicentre cross-sectional study of 556 patients with TOF, NSVT was the single most common arrhythmia subtype with a prevalence of 14.2%, which markedly increased after the age of 45 years. In contrast, VF was documented in only 0.5% of the population.[19] Additional data on the prevalence of any ventricular arrhythmias come were derived from ICD interrogation studies.[63] There were remarkable differences in the type of documented ventricular arrhythmias. Monomorphic VTs constituted 81.5% of all ventricular arrhythmias in TOF.[57] In contrast, only 48.9% of all ventricular arrhythmias in D-TGA were monomorphic VT, the remaining being polymorphic VT in 34%, and VF in 17%.[61] Monomorphic VT in TOF were typically rapid with median heart rates of 213 bpm.[56,66–69]

Monomorphic VT due to a specific substrate can be targeted by catheter or surgical interventions, independent of tachycardia cycle length, and need to be distinguished from polynorphic VT/VF associated with advanced ventricular dysfunction with or without surgical scars.[66,67,69,70]

Sudden cardiac death

Due to the low prevalence of CHD, patients with such diseases represent a minority in the overall group of patients suffering SCD.[71] Sudden cardiac death, however, represents one of the three main causes of death in patients with CHD with the other two being progressive heart failure and perioperative mortality.[72,73] Consistent data in large populations of patients with CHD have shown a percentage of SCD among deaths in this population of approximately 20–25%.[59,74] The risk of SCD increases with complexity of the disease.[59,74] Nevertheless, it has to be kept in mind that even patients with mild disease still have a non-negligible risk of SCD. Finally, as arrhythmias remain the main contributing cause of death in patients with cyanotic lesions, SCD in patients with non-cyanotic disease and simple lesions is probably related to coronary artery disease.[75]

Sudden cardiac death associated with bradycardias and atrial arrhythmias

The risk of SCD from sinus or junctional bradycardia leading to sinus arrest is controversial.[72,73,76,77] The bradycardia–tachycardia syndrome is considered to predispose for SCD.[19,78–80] SCD due to AV block represents probably less than 5% of all SCD in adult CHD patients.[19,73,78–80] Prolonged asystole or bradycardia triggered VT are potential causes of SCD in untreated AV block.[79] Treatment must be focused on the underlying disease and includes catheter or surgical ablation and/or antiarrhythmic therapy as well as pacemaker implantation in bradycardia–tachycardia syndrome.[30]

Sudden cardiac death due to ventricular arrhythmias

Approximately 20–25% of deaths in adults with CHD are estimated to be due to sudden cardiac events.[59,65,70,81,82] However, identifying patients at risk for SCD remains a challenge. With the exception of TOF, specific guidelines regarding ICD implantation for primary prevention in CHD remain elusive. Patients considered as high risk for SCD include those with systemic ventricular ejection fraction (EF) ≤35%, biventricular physiology, and NYHA Class II or III symptoms. Risk factors for SCD in TOF include left ventricular systolic and diastolic dysfunction, QRS duration ≥180 ms, extensive right ventricular scarring, NSVT, and inducible sustained VT during the electrophysiological study.[19,30,57]

The value of programmed ventricular stimulation beyond TOF is unknown. In a small group of d-transposition patients with intra-atrial baffles, positive ventricular stimulation studies were not predictive for future events.[61] Decreased systemic right ventricular function in patients with D-TGA after atrial switch operations has been identified as a risk factor of ventricular arrhythmias and SCD.[80,83] Unfortunately, no data to date suggest a defined cut-off for right ventricular dysfunction as quantified by EF. Implantation of an ICD may be considered in patients with low right ventricular EF and additional risk factors which include complex ventricular arrhythmias, unexplained syncope, NYHA Class II or III, QRS duration >140 ms, or severe systemic AV valve regurgitation.[33,61,80,83]

The risk of inappropriate shocks in CHD patients with primary prevention ICDs must be outweighed against the benefits.[30,84] Rates of inappropriate shocks were mostly the result of misinterpretation of sinus tachycardia, supraventricular arrhythmias, T-wave oversensing, and lead failure. Decreased lead longevity and performance must also be taken into account in this patient population as rates of lead failure are definetely higher compared to adults without CHD.[85] Value of subcutaneous ICD (S-ICD) in CHD patients lacking need for permanent pacing needs to be established.[86]

Bradyarrhythmias in patients with congenital heart disease

Congenital atrioventricular block

Congenital complete heart block (CCHB), first described by Morquio in 1901[87] and documented on ECG in 1908,[88] is due to disruption of the electrophysiological continuity between atria and ventricles.[89–92] Congenital complete heart block may occur in a structurally normal heart (isolated CCHB) or in association with a variety of CHD. The frequent aetiologies of CCHB are listed in *Table 3*. Symptoms are related to underlying anatomy and ventricular rate and include heart failure prenatally and in infancy, and reduced exercise tolerance, presyncope, or syncope (Stokes–Adams attacks) at any age.[93–97]

Long-term conventional right ventricular apical pacing may result in desynchronization of ventricular electrical activation leading to deleterious left ventricular remodelling, dilatation, and asymmetrical hypertrophy.[98–100] Regular assessment of cardiac function is therefore required. Therefore, upgrade to biventricular pacing in patients with congenital heart block and high rate of right ventricular pacing (RVP) should be considered.

Table 3 Aetiologies of AV block in patients with congenital heart disease

Congenital atrioventricular block
In association with concomitant congenital heart disease
Congenitally corrected transposition of the great arteries (especially L-transposition of the great arteries)
AV septal defects
Heterotaxy syndrome with left atrial isomerism
L-Looped single ventricle
Secundum atrial septal defect
Post-operative atrioventricular block
Ventricular septal defect
Atrioventricular septal defect
Tetralogy of Fallot
Congenitally corrected transposition of the great arteries surgery, especially mitral valve and multivalve surgery involving the tricuspid valve
Left ventricular outflow tract surgery

Disease order does not reflect the order of probability of iatrogenic AV block.
AV, atrioventricular.

Post-surgical atrioventricular block

Post-surgical heart block was first explored in detail by Lillehei in 1963[101] and still occurs in 1–3% cases of congenital heart surgeries[102–105] especially after closure of ventricular septal defects, after TOF repair, surgery along the left ventricular outflow tract, and left-sided valve surgery.[30] A recent multicentre study demonstrated highest incidences of AV block and pacemaker placement after the double switch operation (15.6%), tricuspid valve (7.8%) and mitral valve (7.4%) replacement, atrial switch with ventricular septal defect repair (6.4%) and Rastelli operation (4.8%).[106] Compared to surgery, a higher rate (3–20%) of complete heart block (CHB) following transcatheter device closure of perimembranous ventricular septal defect has been reported[107–110] resulting in abandoning the procedure in children <30 kg or age <6 years. Complete heart block usually occurs immediately after surgery or early in the post-operative period and has been reported only occasionally months or even years thereafter.

Incidence of late-onset AV block has been reported with a rate of 0.3 to 0.7%.[111–113] Late-onset AV block may be due to progressive fibrosis and sclerosis involving specialized conduction pathways which are fragile in CHD.[114]

Early post-operative CHB can be transient or permanent. Most patients with early post-operative AV block recover spontaneously[115–119] within the first 7 to 10 days.[120] Therefore, at least 7 days of observation before pacemaker implantation is strongly recommended. During this period, temporary pacing wires may be needed to maintain adequate chronotropy. Late recovery has also been demonstrated by several investigators.[121–124] First degree, second degree AV block combined with complete right bundle branch block have been observed after recovery from complete AV block.[105,125] Limited data suggest that residual bifascicular block after early post-operative complete AV block is a significant risk factor for late-onset complete heart block and SCD.[112]

One year of mortality of patients with complete post-surgical AV block who did not receive permanent pacemaker implantation has been reported ranging from 28 to 100% in several studies from 30 years ago[101,117,126] underscoring undisputable need for permanent pacemaker therapy.

Sinus node dysfunction

Sinus node dysfunction (SND) encompasses a broad array of disturbances in impulse generation of the sinus node and its propagation to the surrounding atrial tissues. It may be a consequence of specific congenital structural defects such as left atrial isomerism[127,128] and left-sided juxtaposition of the atrial appendages. In most patients, SND is secondary to surgical procedures such as repair of sinus venosus ASDs,[129] Glenn shunts, Fontan operation,[130–134] and Senning or Mustard procedures.[135–137] Post-operative SND may result from direct damage to the sinus node or its blood supply.

Loss of AV synchrony may distinctly worsen AV valve regurgitation, and a low resting heart rate with poor chronotropic response is associated with reduced cardiac output. Sinus node dysfunction results in atrial remodelling[138] with increasing risks of atrial tachyarrhythmias, the tachycardia–bradycardia syndrome.[139] Although there has not been direct evidence of the relationship between SND and sudden death in patients with congenital heart disease,[76] uncontrolled atrial tachyarrhythmias may increase the risk of heart failure, mortality, and SCD,[73,140,141] particularly in those with ventricular dysfunction. In addition, thrombo-embolic events including stroke have been reported, especially in patients with tachycardia–bradycardia syndrome. Symptoms of SND vary depending on function of the cardiac conduction system, particular underlying structural heart defect and age. Fatigue, dizziness, exercise intolerance, and syncope have been reported in children and adults.[142] Twelve-lead ECG, ambulatory Holter, event monitors, and exercise stress tests should routinely be performed for assessment and diagnosis of SND. Provocative testing or electrophysiological study is rarely necessary.[143]

Management of arrhythmias in congenital heart disease

Pharmacological management

Arrhythmia mechanisms in adult congenital heart disease classically are represented by atrial or ventricular macroreentrant circuits rotating around sites of fixed and functional myocardial conduction block, through areas of slowed conduction, and thereby facilitating that the wave front captures excitable myocardium. Additional tachycardia types include focal tachycardias, AVRT, AVNRT, and AF. Pharmacological therapy should be guided by understanding of the individual cardiac pathology, the specific electroanatomic substrate and electrophysiological features of the tachycardia (*Tables 4–6*).

Given the marked heterogeneity and the continued evolution of adult congenital heart disease, no randomized controlled trial data is available to guide pharmacological management. Reports encompass largely retrospective experience with different agents prescribed based on the specific clinical situation.

Table 4 Recommendations for acute pharmacological therapy

Recommendations	Consensus statement	References
Before administration of antiarrhythmic drugs in CHD patients, potential coexisting factors such as sinus node disease/AV block, ventricular dysfunction, and other comorbidities must be assessed		48,144
Electrical cardioversion is recommended for haemodynamically unstable SVT (caution for sinus node dysfunction and impaired ventricular function with need for chronotropic or inotropic support)		Expert consensus
In patients with AV-nodal dependent SVT, adenosine is recommended for acute management (unless contraindicated and caution for sinus node dysfunction)		Expert consensus
Atrial overdrive pacing (via oesophagus or endocardial) may be considered for conversion of haemodynamically stable AVNRT/AVRT or atrial flutter		Expert consensus

AV, atrioventricular; AVNRT, atrioventricular nodal reentrant tachycardia; AVRT, atrioventricular re-entrant tachycardia; CHD, congenital heart disease; SVT, supraventricular tachycardia.

Acute management

Adenosine is the treatment of choice for acute management of AV nodal dependent mechanisms. Special consideration should be taken into account when using adenosine in patients with low EF depending on high heart rate in order to preserve cardiac output. Use of adenosine is not advocated in this instance. In haemodynamically stable patients intravenous beta-blockers (Class II) or calcium-channel antagonists (Class IV) may afford rate control prior to spontaneous conversion or alternative management.[1] In a Dutch series of CHD patients with first presentation of SVT, 14 patients underwent intravenous beta-blocker therapy with 100% success, and 34 patients had oral therapy with 88% success.[48] Additional agents included adenosine, sotalol, amiodarone, flecainide, verapamil, metoprolol, and procainamide.[48] In 29 patients with AF or AVRT dofetilide (not available in Europe) successfully reestablished sinus rhythm in 12, but side effects were significant (41%).[150] Quinidine, disopyramide, and sotalol, have been associated with increased all-cause mortality.[156] The potential benefits of intravenous sotalol remain unknown at present. Negative inotropes such as beta-blockers and verapamil can cause cardiovascular collapse when ventricular function is compromised. Prior echocardiography and a slow infusion rate rather than bolus injection should be warranted. In patients with documented SND,

Table 5 Recommendations for pharmacological therapy of supraventricular tachycardia

Recommendations	Consensus statement	References
In patients with CHD and AT/AF rhythm control is recommended as the preferred initial strategy.		Expert consensus
Catheter ablation is recommended as first line therapy and preferred to long-term pharmacological treatment, in case of amenable, circumscribed substrates.		Expert consensus
In AT/AF and failed conversion or stabilization of sinus rhythm, pharmacological AV blockade using ß-blocker or calcium channel blocker (in patients with normal systemic ventricular function and absent preexcitation) should be considered to prevent rapid AV conduction. Combination therapy may be needed in selected patients.		30,145
Amiodarone may be considered for AT/AF recurrence prevention in patients with CHD and systemic ventricular dysfunction, hypertrophy of systemic ventricle, or coronary artery disease, in whom catheter ablation fails or is otherwise no option; side effects are frequent and may require discontinuation; long-tern amiodarone therapy is not advised in young CHD patients		30,48,146,147
Amiodarone should be used with caution in cyanotic CHD, low body weight, hepatic, thyroid, or pulmonary disease or prolonged QT interval		146,147
ß-blockers, if tolerated, may be administered to reduce ventricular tachyarrhythmia rapid AV conduction burden in selected CHD patients with AT		148–150
Dofetilide can be used as an alternative to amiodarone for AT/AF recurrence prevention in patients with CHD and should be considered as first line therapy in patients with normal systemic ventricular function and as second line therapy in those with systemic ventricular dysfunction. Close monitoring of renal function, concomitant medications, and corrected QT interval is required.		61

Continued

Table 5 *Continued*

Recommendations	Consensus statement	References
Dronedarone, for AT/AF recurrence prevention, should be considered as a second line alternative to amiodarone in patients with atrial fibrillation or atrial flutter with normal ventricular function. Close monitoring of liver function is mandatory; in patients with increased risk for stroke and cardiovascular mortality especially in patients with heart failure or after myocardial infarction close follow-up is advised.		151
Indications for anticoagulation therapy for AT or atrial flutter are not different as for patients with AF.		Expert consensus
Oral Class I agents are not recommended in the treatment of AT/AF in patients with coronary artery disease or decreased ventricular function.Dronedarone should not be used in patients with heart failure (Class IV or decompensated heart failure).		152–154

Normal systemic ventricular function: RV SF > 50%, LV > 60%.
AF, atrial fibrillation; AT, atrial tachycardia; AV, atrioventricular; CHD, congenital heart disease; ICD, implantable cardioverter-defibrillator; LV, left ventricular; RV, right ventricular; SCD, sudden cardiac death; SVT, supraventricular tachycardia; VT, ventricular tachycardia.

Table 6 Recommendations for pharmacological therapy of ventricular tachycardia

Recommendations	Consensus statement	References
Electrical cardioversion is recommended for acute termination of haemodynamic stable/unstable VT. If not possible, intravenous amiodarone or procainamide may be considered.		155
For non-sustained ventricular arrhythmias lacking increased risk of SCD, the use of ß-blockers is widely accepted to reduce symptoms or the risk of tachyarryhthmia induced ventricular dysfunction		Expert consensus
Antiarrhythmic drugs may be be used as adjunct to an ICD in order to reduce the ventricular arrhythmia burden		
After haemodynamically unstable, non-idiopathic VT or aborted SCD, antiarrhythmic drugs are not recommended as stand alone therapy		155

ICD, implantable cardioverter-defibrillator; SCD, sudden cardiac death; VT, ventricular tachycardia.

back-up pacing should be available prior to attempted pharmacological treatment.

In view of unpredictable side effects of negative inotropic agents in CHD patients with paroxysmal SVT or AF, electrical cardioversion (AF) or i.v. adenosine (SVT) should be the preferred forms of acute therapy. Acute atrial overdrive pacing is very effective in terminating AF and MRAT avoiding side effects of pharmacologic agents at all (*Table 4*).

Long-term management

Long-term antiarrhythmic therapy is associated with poor long-term arrhythmia freedom and a high rate of side effects in CHD patients. At 2.5 ± 1.4 years follow-up only 45% of 70 patients treated for SVT with varying anti-arrhythmic agents had freedom from recurrence. Success was higher in those with heart failure symptoms at baseline (hazard ratio 1.9; $P = 0.023$). Amiodarone is an effective agent for complex CHD (with or without ventricular dysfunction) and for moderate CHD with ventricular dysfunction, although patients frequently develop side effects requiring discontinuation or dose reduction.[48] Amiodarone as a long-term therapeutic regimen is therefore not recommended in young CHD patients. Sotalol should not be used related to increased risk for proarrhythmias and mortality.[156] Dofetilide therapy either maintained sinus rhythm or improved arrhythmia control in 49% of patients at a median follow-up of 3 years,[150] but side effects were significant.

Given the poor efficacy of antiarrhythmic agents and prevalence of CTI dependent AF in 75–80% of patients following biventricular repair of CHD, a primary ablative strategy may be[157] (*Table 5*).

Side effects of antiarrhythmic therapy in congenital heart disease

Antiarrhythmic therapy is associated with a significant potential for side effects in CHD population, specifically worsening of SND, negative inotropy, AV block, and aggravation of repolarization abnormalities. These problems may be accentuated by altered pharmacodynamics related to circulatory anomalies or hepatic and/or renal dysfunction, necessitating careful dose adjustment. Efficacy of enteral agents may be hampered by gastrointestinal congestion resulting in poor absorption.

Major adverse events including ventricular arrhythmias and stroke have been reported. Some antiarrhythmics such as IC drugs may slow the rate of the atrial arrhythmia without blocking AV conduction and potentially allowing 1:1 conduction of an atrial arrhythmia with worsening haemodynamics. Thyroid disease is a major issue under amiodarone therapy with a reported prevalence of 33 in

90 patients. Another 19 patients developed hyperthyroidism after a median of 2.5 years, and 14 developed hypothyroidism after a median of 3.5 years. In general, risk factors for amiodarone toxicity include female sex, adolescent age, cyanotic heart disease, and a Fontan circulation.[147]

Anticoagulation

Thrombo-embolic cerebrovascular complications are a major cause of morbidity in the CHD population, necessitating individualized risk stratification and initiation of anticoagulant therapy. Age and sex standardized incidence rates of ischaemic stroke demonstrate an 9- to 12-fold risk for the CHD population below 55 years of age with heart failure, diabetes mellitus, and recent myocardial infarction identified as the strongest predictors.[158] Specific anatomic groups have been shown a particular high risk of thrombo-embolic complications, including uncorrected cyanotic heart disease (23%), Eisenmeger physiology (5%), native ASDs (4%), and the Fontan circulation (4%). In 25% of cases, stroke was associated with loss of sinus rhythm.[159] Specific groups such as those with the Fontan circulation or those with residual right-to-left intracardiac shunts (e.g. Eisenmenger syndrome) may benefit from anticoagulation. Although patients with central cyanosis do have a higher risk for thrombo-embolism because of low flow, remarkably they have also a higher bleeding risk as coagulation is insufficient. In most centres, the current practice is to anticoagulate cyanotic patients when suffering from AF/flutter; however, with accepting increased risk for life-threatening bleeding.

Anticoagulation should always be individualized in discussion with the patient and based on the relative risks of stroke and haemorrhagic compliations. In lower risk patients strategies similar to those used for other variants of structural heart disease, the CHA_2DS_2-VASc risk-factor based approach appears appropriate.[160] Stroke prevention is recommended with the same indications as in AF amongst CHD patients with typical AF or AT (*Table 5*). Low risk patients with AF, defined as CHA_2DS_2-VASc 0 in males or 1 in females, typically do not need antithrombotic therapy.[161]

Effective stroke prevention in patients with CHA_2DS_2-VASc score $\geq$1 is oral anticoagulation, whether with well controlled vitamin K antagonists (VKA) or with a non-VKA oral anticoagulant (NOAC)—either dabigatran, rivaroxaban, apixaban, or edoxaban.[162]

Catheter ablation

Catheter ablation of congenital (inherited) tachycardia substrates

Besides the fact that antiarrhythmic drugs are often associated with negative inotropic and/or dromotropic effects in CHD, the main reason for performing ablation as first-line therapy is its efficacy superior to pharmacological therapy. Catheter ablation is increasingly used in CHD patients to treat all types of tachycardias including AVRT, AVNRT, and VT.[29,47] However, while acute success rates of ablation are high, recurrences are not uncommon, and approximately 50% of CHD patients remain free of non-CTI dependent MRAT and VT in the long term.[163]

Technical challenges and limitations

When planning an ablation procedure, alternative access routes to the heart may be needed, such as internal jugular, subclavian, femoral collateral, or transhepatic access.[164] Expertise in EP ablation is also mandatory if transseptal or transbaffle punctures are needed to access the pulmonary venous atrium.[165–168] This procedure should be reserved for specialized, dedicated centres with surgical back-up as an alternative to retrograde transaortic access[165] (*Table 7*). Epicardial ablation approach in post-operative CHD may be required in selected patients, but data are sparse.

Anatomical challenges

The cornerstone of catheter ablation procedures in CHD patients with congenital SVT substrate is the correct understanding of site and course of the specific conduction system and individual cardiac anatomy.[8–10,19,21,23,27,28,43,136,164,175,178–199] In patients with physiologically aligned septal structures, such as in VSD and TOF, the triangle of Koch serves as a good reference for location of the compact AV node, whereas its position is highly variable when abnormal alignment of the septum is present[8,19,21,23,27,28,136,179–184] Anatomic variations of the specific conduction tissue have a direct impact on the rate of major procedural complications, particularly AV block.

Specific arrhythmias and malformations

In patients with Ebstein's anomaly, episodes of paroxysmal SVT are frequent, and are typically related to the presence of accessory pathways.[10,185–193] Catheter ablation of accessory AV connections is the preferred therapeutic option with lower acute success rates[189] and higher recurrence rates compared with patients with normal hearts[190,191] (*Table 7*). With further development of catheter and sheath technology and by the advent of 3D mapping systems[10] success rates and safety have been markedly improved.

Congenitally corrected TGA is frequently associated with abnormalities of the systemic tricuspid valve, including dysplasia and displacement of the leaflets and shows a high incidence of accessory AV connections.[10] As in patients with Ebstein's anomaly, ablation is considered the preferred option. However, the abnormal location of AV conduction tissue with its fragile condition warrants a supreme care to avoid AV block and obviate the need for a permanent pacemaker.[193]

In patients with AV septal defects, the connecting AV node is displaced to an inferior location at the right AV valve annulus, independent from typical margins of Koch's triangle. The non-branching and branching bundles are typically exposed by the defect, i.e. not covered by valvular tissue, making them more susceptible to damage.

Catheter ablation in post-operative settings

Experience with ablation of AVNRT in post-operative CHD patients is limited.[29,47,163] Its occurrence has been highlighted after Mustard or Senning operation for D-TGA.[43,198] The basic principle of ablation in this particular post-operative anatomy is equivalent to that in normal hearts. The zone of slow AV nodal conduction in proximity to the inferior ridge of the coronary sinus ostium at the posterior septal portion of the tricuspid valve annulus serves as target for radiofrequency energy application.[29,199] However, accessing this anatomical region in Mustard/Senning patients as it is most often located in the pulmonary venous atrium either requires a trans-aortic route, crossing the aortic and subsequently the tricuspid valve, or a trans-baffle puncture for a direct antegrade approach. Slow-pathway ablation

Table 7 Recommendations for supraventricular tachyarrhythmia catheter ablation

Recommendations	Consensus statement	References
Catheter ablation is recommended for symptomatic sustained recurrent SVT (AVNRT, AVRT, or apparently focal atrial tachycardias) over long-term medical therapy, especially in simple CHD scenarios[a]		45,169–171
In simple CHD substrates, ablation of symptomatic MRAT is generally recommended as an alternative to antiarrhythmic drugs and/or electrical cardioversions[a]		172–174
In complex CHD substrates, ablation of symptomatic MRAT is recommended as an alternative to antiarrhythmic drugs and/or electrical cardioversion at experienced centres[a]		172,175–177
The use of 3D mapping systems, and irrigated tip catheters for MRAT ablation, is recommended in CHD patients		176,178
In simple and complex CHD scenarios, ablation of less symptomatic, recurrent sustained paroxysmal or persistent MRAT can be an alternative to antiarrhythmic drugs and/or electrical cardioversions, especially at dedicated centres[a]		172–177 45,172,174, 175,177
In simple and complex CHD scenarios, ablation of symptomatic sustained MRAT may be recommended at dedicated centres, to prevent recurrences, when medical therapy (including cardioversions) is either ineffective or non-properly tolerated, when complication rates are not expected to be low, but within a reasonable range of safety[b]		45,50–52,55
Atrial fibrillation ablation may be considered in selected simple CHD patients, at experienced centres[c]		
AV blockade and permanent pacing can be considered as third line therapy for the treatment of symptomatic atrial tachyarrhythmias when other medical and ablative therapies have failed		
Catheter ablation is not recommended for atrial tachyarrhythmias that can be controlled medically in the early post-surgical period (<3 months).		Expert consensus
Catheter ablation is not recommended for asymptomatic non-sustained runs of atrial tachycardia.		Expert consensus

AV, atrioventricular; AVNRT, atrioventricular nodal reentrant tachycardia; AVRT, atrioventricular re-entrant tachycardia; CHD, congenital heart disease; MRAT, macroreentrant atrial tachycardia; SVT, supraventricular tachycardia.
[a]When complication rates are expected to be low (larger patients, conventional vascular accesses. . .).
[b]Like in smaller patients, unconventional vascular accesses. . ..
[c]When post-ablative recurrence and complication rates are presumed to be within standard ranges published from non-CHD patients series.

occasionally may require additional radiofrequency energy application to its 'left-sided' atrio-nodal input from the systemic venous aspect.

Outcome and complications

Acute success of ablation of AVRT and AVNRT in CHD patients is lower than reported in normal hearts with average rates of 80%.[9] However, with increasing experience and implementation of electro-anatomical mapping systems results could be improved.[175] Overall, major complication rates may reach up to 4.2% and minor complications rates of 5.5% have been reported. These figures are comparable with complication rates in normal cardiac anatomy.[191]

Catheter ablation of post-operative substrates

Arrhythmogenesis

Post-operative CHD patients are prone to develop significant tachyarrhythmias in their adulthood. Areas of dense fibrosis due to surgical incisions, patch material, and the valve annuli are regions of conduction block that create intervening anatomical isthmuses of myocardium.[10] Fibrosis due to longstanding cyanosis, pressure and/or volume overload and aging may provide the substrate for slow conduction within these anatomically defined isthmuses.[30]

Patients with atrial tachyarrhythmias often harbour a multifactorial proarrhythmic state,[178] complicating ablation procedures. There is variable heterogeneous impulse propagation and slow conduction. Scars from surgical atriotomies, suture lines for intracardiac baffles or patches, or even radiofrequency lesions from previous ablations, may facilitate development of preferential conduction channels or isthmuses within scars or adjacent to anatomical barriers.[178,200]

Usually, the more complex the baseline substrate and the surgical procedure, the higher the chances of developing complex atrial or ventricular reentries.[10] In general, early total repair and evolving surgical strategies will likely lower the risk for acquired substrates in contemporary patients. In particular, in TOF patients avoiding a right

Table 8 Recomendations for VT ablation

Recommendations	Consensus statement	References
Catheter ablation is indicated as additional therapy in ICD patients with CHD who present with incessant VT or electrical storm.		66,67,201
Catheter ablation is indicated as additional therapy in ICD patients with CHD who have recurrent symptomatic monomorphic VT despite ATP or appropriate ICD shocks not manageable by device reprogramming or when medical therapy is not effective or has intolerable side effects.		66–68,70
Pre-operative electroanatomical mapping to identify SCAI as substrates for spontaneous or induced monomorphic VT is recommended in patients with TOF who undergo re-operation for haemodynamic reasons.		69
Surgical intraoperative cryo-ablation is recommended in ToF patients who undergo re-surgery and who have spontaneous or inducible VTs unless managed by preoperative ablation.		69,202,203
The use of 3D mapping systems to obtain a 3D reconstruction of all VT related anatomic isthmuses is recommended.		66,67,69,70
Catheter ablation should be considered for symptomatic monomorphic sustained VT in grown up-congenital heart disease patients as an alternative to drug therapy.		66,67,69
Catheter ablation should be considered in grown-up congenital heart disease patients with frequent highly symptomatic PVCs, or PVCs associated with deteriorating ventricular function.		20
Catheter ablation or concomitant surgical cryoablation can be considered for symptomatic monomorphic sustained VT in grown-up congenital heart disease patients with a preserved biventricular function as an alternative to ICD therapy, provided that the procedure is performed in highly experienced CHD centres and that established ablation endpoints have been reached (i.e. non-inducibility, conduction block across ablation lines).		67,69
Surgical intraoperative cryoablation of SCAI may be considered in rTOF patients with SCAI but without spontaneous or inducible VT who need re-operation for residual haemodynamic lesions.		69
The use of irrigated tip catheter is recommended for linear lesions and hypertrophied myocardium.		66,67,202
Catheter ablation is not recommended for asymptomatic, infrequent PVC and non-sustained VT		

AT, atrial tachycardia; AV, atrioventricular; AVNRT, atrioventricular nodal re-entrant tachycardia; AVRT, atrioventricular re-entrant tachycardia; CHD, congenital heart disease; PVC, premature ventricular complex; rTOF, repaired tetralogy of Fallot; SCAI, slow conducting anatomical isthmus; SVT, supraventricular tachycardia; VT, ventricular tachycardia.

ventriculotomy by using a transatrial–transpulmonary approach and small transannular patches might positively impact characteristics of anatomical isthmuses.

Prerequisites, procedural planning, and technical requirements

As ablation procedures in post-surgical patients with CHD are often complex, a team with expertise in EP and knowledge on CHD is mandatory in order to achieve ablation success[30] (*Tables 7–9*).

Prerequisites for a successful procedure include evaluation of the patient's cardiology and surgery records and thorough pre-procedure imaging (usually involving CT or MRI ± haemodynamic catheterization).

Fluoroscopy reduction needs to be emphasized. Nowadays electrophysiological procedures should be performed according to the As Low As Reasonably Achievable (ALARA) principle which includes the use of 3D mapping technology allowing to identify the exact location of the mapping catheter and important anatomical structures such as valve annuli, His bundle, or phrenic nerve, providing better long-term outcomes.[200,204] For mapping and ablation of AT and VT in CHD, 3D reconstruction of all potential anatomical isthmuses facilitated by the use of 3D mapping systems is highly recommended.[178,200,204–207] The use of irrigated-tip catheters has been associated with an increase in ablation success rates, to overcome limitations with lesion formation in thickened, fibrotic atrial, and ventricular myocardium in CHD patients. Contact force may be helpful as well as intracardiac echocardiography[208] in regions with low amplitude signals to establish electrode to tissue contact and to differentiate from myocardial fibrosis. This type of actheter should be used with caution due to its stiffness. It may be more difficult to move around in tight channels and where one need to loop.

Catheter ablation of atrial tachycardia

Success of catheter ablation for ATs in CHD patients has steadily increased in dedicated centres.[147,172,176] Concerning ablation, AT can

Table 9 Requirements for complex CHD arrhythmia management

Requirements for complex CHD arrhythmia management centres
CHD surgery department on site
Haemodynamic interventionalist with special expertise in CHD
Adult/pediatric intensive care unit with experience in arrhythmia management in CHD patients
Imaging requirements (CT and MRI available)
3D mapping systems
Requirements for invasive EP in complex CHD patients
Experience of complex ablations including marcoreentrant atrial tachycardia, VT and AF ablation
Capability of alternative access such as transbaffle/transhepatic/transventricular/epicardial/hybrid
Experienced using 3D mapping systems

CHD, congenital heart disease; CT, computed tomography; EP, electrophysiology; MRI, magnetic resonance imaging.

be subdivided in localized NAFAT and the more prevalent macroreentries. Non-automatic focal AT, defined as localized tachycardias inducible and terminable with programmed stimulation, either reentrant or due to triggered activity, may account for 5–10% of all regular ATs.[169,209–211] High ablation success rates have been reported.[43,209,211] In macroreentries, identification of the critical isthmus sustaining the circuit is crucial for proper ablation. The most prevalent macroreentrant circuit is CTI-dependent, either in operated and non-operated CHD patients.[43,170,171] However, the surface ECG may not unambiguously look like classic AF depending on altered anatomy. Atypical morphology does not exclude CTI dependence.[207] Catheter ablation aims to interrupt electrical conduction within relevant isthmuses.

In repaired CHD individuals, most non-CTI reentries tend to revolve around injured tissue as incisional tachycardias at the lateral and anterior right atrial wall.[43,170,171,210,212] 'Figure-of-eight' circuits may complicate understanding and ablation of such tachycardias. Subtle abrupt electrogram changes may predict partial interruption of the double circuit.[170,171,212]

In Fontan patients, MRAT tend to course along the entire lateral right atrial wall and in vicinity to the inferior caval vein.[12,170,213] In patients after Mustard/Senning procedure MRAT predominantly involve CTI.[12] The same applies to patients after repair of TOF where the area between atriotomy scar and the inferior vena cava (IVC) may serve as an additional critical substrate.[209]

Based on this experience, in patients with non-inducible MRAT or unmappable circuits, a substrate mapping approach including voltage and activation mapping can be performed during sinus rhythm or fixed atrial pacing.[214,215] Finally, empirical CTI ablation[216] as well as a corridor between right lateral atriotomy scar and the IVC has been advocated in non-inducible CHD patients.[40]

Acute ablation outcomes range between 80% in scar-related MRAT and reach 90–95% for CTI-dependent MRAT or AF.[173,217,218] Electroanatomical mapping, locating the mid-diastolic isthmus, and adjusting the window of interest are important for ablation. Long-term success, however, may be impaired by progressive myocardial fibrosis, especially in complex post-operative CHD. Acute success in MRAT ablation is defined as achievement of bi-directional conduction block across any critically conducting myocardial isthmus or CTI.[173] In the past, termination of tachycardia with ablation and subsequent non-inducibility of the tachycardia alone were often used as endpoint in ablation procedures. Lack of evidence of complete conduction block, however, was idientified as a risk factor for tachycardia recurrence.[209]

Procedural complications are uncommon and are usually related to vascular access sites or application of radiofrequency near the phrenic nerve or the His-bundle region.[219,220] High-output pacing, in order to track the course of the phrenic nerve with a electroanatomic mapping system, should be performed in advance to radifrequency delivery to the posterolateral aspect of the right atrium or pericaval areas.

Ablation for AF has occasionally been performed in non-complex CHD patients, basically in ASD patients, by pulmonary vein antral isolation, with some groups even including SVC isolation and additional left atrial applications in persisting AF.[50,52,219–221] Echocardiographic guidance is recommended for transseptal puncture in case of anatomical distortion confusing radiological orientation, in case of ASD closure devices or septal patches.[51] There are reports that show that results of AF ablation in CHD patients do not significantly differ from non-CHD populations.[50,219,221]

Catheter ablation of ventricular tachycardia

Over the last decade tremendous progress has been made to understand monomorphic sustained VT in CHD, particularly in patients with TOF.[66,67,70,201,202,223,224] The majority of monomorphic VT targeted by catheter ablation are macroreentrant VT with the critical part of the reentry circuit typically located within anatomically defined isthmuses.[66–68,70,201,224,225] The critical isthmus of an induced VT can be determined by activation and entrainment mapping in haemodynamically tolerated VT or by pace-mapping in unstable arrhythmias. Non-inducibility of any monomorphic VT has been a widely accepted endpoint for VT ablation. In selected patients, VT induction may no longer be a prerequisite for ablation if narrow, slow conducting and thereby potential arrythmogenic isthmuses can be identified by electroanatomical mapping during stable sinus rhythm.[67,69,223] These anatomical isthmuses can be transsected by connecting the adjoining anatomical boundaries by linear radiofrequency lesions.[66,70,201,224] Demonstration of conduction block after isthmus transection provides a defined procedural endpoint similar to that for achieving block in the CTI. A combined endpoint of non-inducibility and conduction block was associated with freedom of VT recurrence during 46 ± 29 months follow-up in a recent series of 25 CHD patients.[67]

Catheter ablation is currently recommended as adjunctive therapy to ICD patients with CHD who have recurrent monomorphic VT or appropriate ICD therapies that are not manageable by device reprogramming or drug therapy.[20,226] (*Table 7*).

Considering the high acute success rates and low recurrence rates VT ablation may offer a reasonable alternative to ICD therapy in carefully selected patients with preserved cardiac function.[66,67,201] Procedural complications are rare and either related to vascular access or location of anatomical isthmuses including AV

block.[66–68,70,201,202,224,225] Reasons for ablation failure include hypertrophied myocardium, proximity of the conduction system, and importantly, the protection of portions of anatomical isthmuses by prosthetic material.[66,201] In particular, a pulmonary homograft may cover parts of the infundibular septum in TOF patients preventing isthmus transection.

Accordingly, preoperative mapping may be considered in patients who require reoperation or percutaneous valve replacement for pulmonary valve regurgitation. Intraoperative cryoablation may be performed in patients with clinical sustained VT and inducible sustained monomorphic VT with an identified critical isthmus if catheter ablation was unsuccessful.[66,67,69,201,203,223] The role of preventive dissection of potential arrhythmogenic isthmuses in TOF patients remains to be determined.[69] (*Table 8*).

Surgical treatment

Amongst the three listed options for antiarrhythmic treatment in this section, surgery plays the smallest role, at least reflected by numbers, but may have high impact on patients' course. Due to the growing arrhythmia load in adults with CHD,[141,227–232] incorporation of antiarrhythmic concepts into primary or particularly re-do surgery for CHD is increasingly considered, at least in specialized centres. Preferred targets for such combined procedures are potential future or already existing substrates for macroreentry circuits. This strategy may be acknowleged as a more gross strategical concept in avoiding development of surgical tachycardia substrates.

Preventative surgery

Preventative or 'prophylactic' arrhythmia operations may be considered for patients with specific anatomic substrates as an effort to reduce the risk of late arrhythmias.[54] Diagnoses include patients with Ebstein's anomaly, patients with ASDs over 40 years of age, with Fontan physiology, and with TOF, as those have been identified at significant risk for late atrial arrhythmias.[19,21,204,232–237] The prophylactic CTI dissection procedure may be beneficial in selected patients undergoing surgery if not managed by catheter ablation beforehand. There is no evidence concerning the value of a left-sided Maze procedure as it prolongs surgery and lack of proof of block across the lines in the non-beating heart. In addition, interoperative Maze procedures may cause more problems in rapid, difficult to control left ATs than benefit. Preventive intraoperative ablation in case of re-operation e.g. pulmonary valve replacement in TOF may be considered.[203,238,239]

Intraoperative treatment of pre-existing tachyarrhythmia

Intraoperative management may constitute an effective strategy to address various arrhythmia substrates during CHD surgery, performed either on an empirical basis using the knowledge of potential arrhythmia mechanisms or ideally using information from preoperative endocardial mapping. Atrial fibrillation can be addressed surgically by pulmonary vein isolation with or without more extensive left atrial ablation.[45] Surgical cryoablation of VT may be performed at the time of pulmonary valve replacement guided by intraoperative epicardial mapping.

Concomitant treatment of MRAT during cardiac surgery for CHD has been reported as safe and effective with freedom from recurrence >80% after 5 years.[240–250]

Indications for surgery of arrhythmias in congenital heart disease

Surgery for arrhythmias assocaited with CHD as a 'stand-alone procedure' is reserved for patients in whom medical treatment and catheter ablation techniques have failed. When a surgical intervention is indicated, concomitant antiarrhythmic surgery may be implemented, especially for treatment of AF, incisional AT, AF, or VT.[246–250]

Rhyhm control in patients with congenital heart disease

Rhythm control devices in patients with congenital heart disease: technical issues

While indications for device implantation in the CHD population are similar to those with primary electrical disorders and structurally normal hearts, there are several issues that require attention prior to the procedure. As for ablation procedures, the legacy from previous cardiac surgery, loss of vascular access or lack of direct access to cardiac chambers and residual devices as well as previous complications related to device implantation may be the starting point when considering device implantation in adult CHD.[251–253] In contrast to patients with structurally normal hearts, epicardial leads (sternotomy or thoracotomy) are frequently required as are hybrid (combined epicardial and endocardial) and novel systems (intra-operative atrial puncture, baffle puncture, femoral or hepatic vein access, and S-ICD coils). Achieving the desired outcome by any route can be challenging.

Epicardial lead placement is favored in patients with intracardiac shunts. Repeated surgical procedures may induce considerable epicardial scar formation resulting in high thresholds of epicardial leads which may cause poor system performance.[254] Access to suitable epicardium may be achieved by a partial or complete sternotomy or right or left thoracotomy. If no suitable epicardium is found, puncture of the target chamber and direct suture of an endocardial lead may be an option.[255] Extraction of redundant leads may facilitate access but can be challenging as some leads have been placed for >10 years or even longer.[256,257] The low profile 4.1 Fr leads may help to preserve venous patency.[258,259] Leadless pacemakers have several advantages but have not been studied in the CHD population. In selected patients, the S-ICD may offer an alternative solution when permanent pacing is not required.[260–264]

Subcutaneous, intrathoracic, or epicardial defibrillator coils may be used in patients lacking access to a subpulmonary ventricle.[260–262] Placement of a lead in a systemic chamber carries an increased risk of systemic thrombembolism as long-term anticoagulation is not completely protective. In patients with intracardiac shunts, prior/simultaneous device closure is mandatory.

Patients with narrowed or occluded intracardiac baffles need prior or simultaneous recanalization and stenting to maintain access and avoid repeated obstruction. Baffle leaks require closure as they are also a risk factor for systemic thrombembolism.

Indication for device implantation should result in consideration of further management strategy in the individual patient:

(1) What surgery or catheter intervention is likely to be needed in the short- to medium-term?
(2) May device implantation cause an obstacle in future procedures, e.g. tricuspid valve replacement when a transvenous lead is in place?

(3) Could the implantation procedure be used for simultaneous TOE, diagnostic catheterisation/pulmonary vascular resistance measurement or intervention (balloon dilation or stent implantation for other lesions)?
(4) Could device implantation be more reliably achieved during concomitant cardiac surgery?
(5) Is imaging up to date and will device implantation prevent future MRI scanning? Magnetic resonance imaging compatible systems will allow future MRI follow-up studies of the underlying congenital heart disease and should be used whenever possible.

Senning or Mustard repair for transposition of the great arteries

Standard transvenous pacing with an atrial lead in the systemic venous atrium and a ventricular lead in the morphologically left ventricle (LV) (subpulmonary chambers) is routinely accomplished. Pacing the systemic venous atrial appendage may cause phrenic nerve stimulation. The atrial lead should be placed at the roof of the systemic venous atrium accordingly. Lateral pacing of the morphologically LV may stimulate the diaphragm and induce ventricular dyssynchrony—the pacing lead should preferably be placed medially along the septum. Screw-in leads are preferred for use in the anatomically LV. For cardiac resynchronization therapy (CRT), epicardial RVP combined with transvenous left ventricular pacing is usually used. Transvenous CRT is technically feasible using a baffle leak or baffle puncture at the expense of an increased risk of systemic thrombembolism despite proper anticoagulation.[265]

Congenitally corrected transposition of the great arteries

In patients with CC-GA, CRT is often needed and can be accomplished via the coronary sinus if present. [266]

Single ventricle physiology: Fontan circulation

Epicardial pacing is usually used. Transvenous pacing is possible by puncturing from the main or left pulmonary artery that overlies the atria again at the expense of an increased risk of systemic thrombembolism despite proper anticoagulation.[267,268] Implantable cardioverter-defibrillator systems will need to be epicardial, hybrid, or completely subcutaneous.

One and a half ventricle circulation

While epicardial pacing is commonly employed, there are transvenous strategies. In the Hemi-Fontan procedure, atrial pacing is accomplished by placing a lead in the SVC stump while ventricular pacing can be achieved by puncturing the patch dividing the superor vena cavae from the right atrium.[269] If the azygos vein is patent, an ICD coil can be placed posteriorly to the heart.[270] In the classical Glenn shunt, puncture is more challenging but the atrium and ventricle can be reached. In this setting as in the Kawashima variant, transhepatic pacing remains an option when no other avenue is available.[271,272]

Pacemakers in patients with congenital heart disease

Patients with CHD and post-operative SND or high degree or complete AV block—even when the underlying defects are 'structurally repaired'—are considered to be at a higher risk of SCD. Therefore, there is a lower threshold for pacemaker implantation even in asymptomatic patients[6,103,273–278] (*Tables 10* and *11*).

Table 10 Recommendations for pacemaker implantation according to the underlying disease

Sinus nodal dysfunction Recommendations	Consensus statement	References
Sinus node dysfunction with documented symptomatic bradycardia or chronotropic incompetence that is intrinsic or secondary to drug therapy		6,76,279–281
Bradycardia induced sustained VT (with or without QTc prolongation) if ICD therapy is not indicated		Expert consensus
Patients with bradycardia-tachycardia syndrome to prevent atrial re-entrant tachycardia, if ablation fails or is not possible		6,282
Sinus bradycardia with complex congenital heart dísease and a resting heart rate <40/min or pauses >3 s. Is this a 'should do'?		6
Compromised haemodynamics due to sinus bradycardia or loss of AV synchrony		6
Asymptomatic sinus bradycardia after biventricular repair of CHD with a resting rate <40 s or pauses >3 s		6,77
Symptoms likely to be associated to bradycardia even if not completely conclusive		143
Asymptomatic sinus bradycardia with pauses <3 s and a minimium heart rate > 40/min		6
Symptomatic sinus bradycardia due to a reversible cause		143

AV, atrioventricular; CHD, congenital heart disease; ICD, implantable cardioverter-defibrillator.

Sinus node dysfunction

Sinus bradycardia may predispose to MRAT and AT—the bradycardia–tachycardia-syndrome. Antiarrhythmic medication may aggravate bradycardia. Increasing basal heart rate may reduce incidence of ATs.

Atrioventricular block

Patients with post-operative CHB are considered to be at high risk of SCD and invariably undergo pacemaker implantation independent of symptoms (*Table 11*). Indications for pacemaker therapy comply

Table 11 Recommendations for pacemaker implantation according to the underlying disease

AV block Recommendations	Consensus statement	References
Symptomatic bradycardia with any degree of AV block or with ventricular arrhythmias presumed to be due to AV block		6
Asymptomatic patients with high degree or complete AV block and with one of the following: a. Ventricular dysfunction b. Low cardiac output c. Wide QRS escape rhythm d. Complex ventricular ectopy e. Prolonged QT interval		6,283
Post-operative or post-interventional 3° AV block if considered irreversible and lasting >7 days		6,284
Symptomatic 2° type 1 AV block, 2° AV block at intra- or infra-His levels according to EPS		143
Syncope in patients with residual bifascicular block and prior complete AV block following heart surgery after excluding other causes		120
Transient complete post-operative AV block with restitution of sinus rhythm with residual bifascicular block		6,12
Transient post-operative complete AV block which returns to normal AV conduction in asymptomatic patients		6,105,112
Post-operative asymptomatic bifascicular block ± 1° AV block without prior transient complete AV Block		6

AV, atrioventricular.

primarily with clinical symptoms and cardiac function rather than predefined heart rate limits. In the last years, several guidelines for device implantation[6,143,277,278] have been published with one study[85] mainly focusing on CHD patients.

Choosing the optimal pacing site

Location of ventricular stimulation plays a decisive role in maintenance of ventricular function especially in young CHD patients who may be paced for decades.[285–289] Detailed information is provided in the section on CRT.

Lead extraction

In CHD patients, the need for lead extraction has grown. However, complex vascular and cardiac anatomy may be challenging. Results of lead extraction in CHD patients are promising.[290–292] Guidelines for lead extraction[293] have been published recently and are applicable for CHD patients after taking into account the venous anatomy and anatomical connections (*Table 12*).

Implantable cardioverter-defibrillator in patients with congenital heart disease

Over the last decades, ICDs have shown to be effective in life-threatening arrhythmias in patients with CHD. Recommendations for risk stratification and indications for primary prevention therapy in this aging and heterogeneous group of patients are still not well defined and are based on observational cohort studies, large registries, and expert opinions.[20,30,58,61,63,294–300] Furthermore, the mode of implantation and long-term management of ICD therapy in CHD patients remains challenging (*Table 13*).

Implantable cardioverter-defibrillator cohorts

In CHD patients, ICD showed significant variance in specific CHD lesions and age at implantation (*Table 13*). The majority of ICD recipients in the adult CHD population were young males (66%) with a mean age at implantation of 36.5 ± 5.5 years. Approximately 50% of ICD recipients had repaired TOF, followed by D-TGA after atrial switch procedure (21%), CC-TGA (5%), and septal defects (5%).[293] In contrast, a study from the US National Cardiovascular Data Registry (NCDR) reported that almost 75% of more than 3000 ICD implants between 2010 and 2012 were performed in patients with simple types of CHD, mainly septal defects, and probably related to ischaemic heart disease. The mean age of patients in this registry, including all ICD implants, was 53 ± 18 years.[300]

Recommendations for implantable cardioverter-defibrillator therapy

Secondary prevention

Evidence based guidelines for secondary prevention of SCD can be extrapolated to the CHD population.[20] Observational studies in cohorts with different types of CHD have shown the efficacy of ICD therapy for secondary prevention, with an average appropriate shock rate of 35% in 4.3 ± 1.2 years, indicating a high annual shock rate of 8%.[300] Indications for ICD implantation are summarized in *Table 14*. In patients with CHD and symptomatic sustained VT, catheter ablation can be a reasonable alternative or adjunct to ICD therapy in highly selected patients with preserved ventricular function.[63,294]

Primary prevention

Indications for ICD therapy in patients with CHD has shifted from secondary to primary prevention in the last two decades. Appropriate shocks for primary prevention were reported on average in 22% of CHD patients witin 3.3 ± 0.3 years, indicating the annual shock rate of 6.6%.[63] Indication for primary prevention ICD implantation are listed in *Table 14*. Cut-off values for left and right ventricular function in primary prevention remain undetermined. It is important to take into account that ICD indications are based on the presence of several risk factors. Proper selection of candidates for primary

Table 12 Recommendations for lead extraction

Recommendations	Consensus statement
Definite system infection, pocket infection, endocarditis (valvular, subvalvular, supravalvular, VSD) and occult gram positive bacteraemia	
Clinically significant thrombo-embolic events associated with thrombus on a lead/lead fragment, bilateral subclavian vein or superior caval vein occlusion precluding implantation of a needed transvenous lead, planned stent deployment in a vein already containing a transvenous lead and ipsilateral venous occlusion preventing access to the venous circulation for required placement of additional leads when there is a contraindication for using the contralateral side	
Life-threating arrhythmias secondary to retained leads, leads that may pose an immediate threat, leads that interfere with the operation of implanted cardiac devices and leads that interfere with the treatment of a malignancy.	
Persistent occult gram negative bacteraemia	
Severe chronic pain at the device or lead insertion site	
Ipsilateral venous occlusion preventing access to the venous circulation for required placement of an additional lead, when there is no contraindication for using the contralateral side	
Device implantation would require more than four leads on one side or more than four leads through the SVC (ref 294)	
Need for specific imaging techniques (e.g. MRI) without alternative techniques being available or to implant an MRI conditional system	
Functional leads that due to their design pose a potential future threat to the patient if left in place	
Abandoned leads that are functional but not being used or non-functional leads without an immediate threat to the patient	
Only superficial or wound infection, without clear involvement of the device and/or leads	
Chronic bacteremia due to a source other than the device system, when long-term antibiotics are required	
Functional but redundant leads or non-functional leads if patients have a life expectancy of less than 1 year	
Anomalous placement of leads through structures other than normal venous and cardiac structures or through a systemic venous atrium or systemic venous ventricle. Surgical removal is preferable though surgical backup may be used if the clinical scenario is compelling	

MRI, magnetic resonance imaging, SVC, superior vena cava.

prevention still remains difficult and should be weighed against the higher complication rate of ICD in CHD.

There is the general agreement that ICDs are also indicated in adults with CHD who meet the established ICD criteria, namely a left ventricular EF < 35%, biventricular physiology, symptomatic heart failure despite optimal medical treatment, and NYHA functional Class II or III.[20] Implantable cardioverter-defibrillators should be considered in CHD patients with syncope of unknown origin in the presence of either advanced ventricular dysfunction or inducible sustained VT or VF on programmed ventricular stimulation.[57]

Most studies on risk stratification have been performed in patients with repaired TOF. In addition to sustained VT several other risk factors for SCD have been reported, including left ventricular dysfunction, non-sustained VT, QRS duration >180 ms, or inducible sustained VT at programmed electrical stimulation.

Table 13 Summary of clinical studies on Implantable cardioverter-defibrillators in CHD

Author (references)	Year	*n*	CHD	Age at implant (years)	Male (%)	Primary prevention (%)	Systemic ventricle function	FU (years)	Approp. shocks (%)	Inapp. shocks (%)	Comp. (%)	Main features
Yap *et al.*[291]	2007	64	Diverse CHD 63% ToF	37	67	39	17% impaired	3, 7	23, 4	40, 6	30	First multicentre study on ICD outcomes in adults with CHD.
Khairy *et al.*[61]	2008	37	TGA	28	89	62	Mean EF 36%	3, 6	14	24	38	High rates of appropriate shocks in secondary but not primary prevention. SVA may be implicated in VT aetiology.
Khairy *et al.*[57]	2008	121	ToF	333	60	56	Mean EF 54%	3, 7	31	25	30	High rates of appropriate and effective shocks in primary and secondary Prev.
Witte *et al.*[292]	2008	25	ToF	NA	NA	28	3% impaired	1, 9	25	20	4	Comparison with dilated CM patients. Higher risk of inapp. therapies lower incidence of approp. therapies.
Khanna *et al.*[293]	2011	73	Diverse CHD ToF 44%	41	68	64	27% impaired	2, 2	19	15	15	Single centre experience. Low risk implant complications.
Koyak *et al.*[58]	2012	136	Diverse CHD ToF 51%	41	67	50	51% moderate to severe impaired	4, 6	32	30	29	Highest risk of appropriate ICD shocks: secondary Prev. indication, coronary artery disease and symptomatic NSVT.
Backhoff *et al.*[207]	2014	12	TGA	30, 3	100	100	NA	3, 5	8	25	17	Inappropriate ICD shocks due to rapidly conducted atrial reentrant tachycardia.Sensing failure frequently observed
Jordan *et al.*[297]	2014	1683	Diverse CHD septal defects 72%	NA	NA	70	NA	NA	NA	NA	NA	NCDR-ICD Registry. Largest pooled assessment for CHD and pediatric ICD populations.
Kella *et al.*[299]	2014	59	Diverse CHD ToF 56%	35, 7	69	53	56% EF ≤ 35%	3, 2	20	22	NA	Patients with non-ToF congenital lesions are significantly less likely to receive appropriate ICD therapy than those with ToF.
Santharam *et al.*[62]	2017	42	Diverse CHD 50% ToF	41	55	38	NA	5	14, 3	26	14, 3	Long-term follow-up. Significant incidence of of complications.
Moore *et al.*[302]	2016	21	Diverse. 52% single ventricle	34	62	67	LV EF 41%RV EF 33%	1, 2	5	21	5	Largest multicentre study of S-ICD implantation for CHD.
Buber J *et al.*[294]	2016	18	TGA	26	83	100	62% moderate to severe impaired	4	5	55	28	Atrial arrhythmias were the most common cause for ICD shocks. VT infrequent.

Approp, appropriate; CHD, congenital heart disease; CM, cardiomyopathy; Comp., complications; EF, ejection fraction; FU, follow-up; ICD, implantable cardioverter-defibrillator; Inapp., inappropriate; LV, left ventricle; NA, non-available; NSVT, non-sustained ventricular tachycardia; Prev., prevention; Ref, reference; RV, right ventricle; S-ICD, subcutaneous ICD; SVA, supraventricular arrhythmias; TOF, tetralogy of Fallot; TGA, transposition of great arteries; VT, ventricular tachyarrhythmias.

Table 14 Recommendations for ICD therapy in adults with CHD

Recommendations	Consensus statement	References
ICD is recommended for patients with CHD who are survivors of an aborted cardiac arrest due to VF or haemodynamically unstable VT after evaluation to define the cause of the event and exclusion of any reversible causes.		57,58,61,295,305,306
ICD is recommended for patients with CHD with symptomatic sustained VT who have undergone haemodynamic and electrophysiological evaluation.		57,58,61,143,295,296
ICD is recommended in adults with CHD and a systemic LVEF ≤ 35%, biventricular physiology and NYHA functional Class II or III.		20,307,308
ICD implantation should be considered in patients with CHD and syncope of unknown origin in the presence of either advanced ventricular dysfunction or inducible sustained VT or VF on VPS.		56,57,58,59,300
ICD implantation should be considered in selected patients with TOF and multiple risk factors for SCD, including LV dysfunction, non-sustained VT, QRS duration ≥180 ms, or inducible sustained VT on VPS.		33,56,57,308
ICD therapy may be considered in patients with advanced single or systemic RV dysfunction in the presence of risk factors such as non-sustained VT, NYHA functional Class II or III, QRS duration ≥ 140 ms or severe systemic AV valve regurgitation.		33,59,61,299
ICD therapy may be considered for non-hospitalized adults with CHD awaiting heart transplantation.		308,309
ICD is contraindicated in patients with CHD with life expectancy with an acceptable functional status ≤ 1 year, incessant VT or VF, significant psychiatric illness that may be aggravated by ICD implantation or preclude systematic follow-up, and patients with drug-refractory NYHA Class IV symptoms who are not candidates for cardiac transplantation or CRT		

CHD, congenital heart disease; CRT, cardiac resynchronization therapy; ICD, implantable cardioverter-defibrillator; LVEF, left ventricular ejection fraction; NYHA, New York Heart Association; VF, ventricular fibrillation; VPS, ventricular programmed stimulation; VT, ventricular tachycardia.

In D-TGA patients after atrial switch operation systemic right ventricular dysfunction has been identified as risk factor for SCD, but a cut-off value for EF or other functional parameters remain undetermined.[61,292] In patients with Single V physiology, risk factors for SCD are largely unknown. Implantation of ICD may be considered in patients with advanced single or systemic right ventricular dysfunction, in the presence of other risk factors such as non-sustained VT, NYHA functional Class II or III symptoms, or severe systemic AV valve regurgitation.[292] In primary prevention of patients with TGA and Single V physiology, a history of intraatrial re-entry tachycardia should be considered as additional risk factor and should be addressed appropriately.

Mortality

A meta-analysis of retrospective ICD series reported an average overall mortality of 10% during 3.7 ± 0.9 years, indicating an annual mortality rate of 3%. Sudden cardiac death occurred in 18% of all deaths and heart failure in 41%. In this meta-analysis, no separate information was given on both parameters for primary and secondary prevention. This mortality rate is much lower than reported in the large randomized ICD trials, which can be explained by the younger age of ICD patients with CHD as compared to ICD patients with ischaemic and non-ischaemic cardiomyopathies.[63]

Complications, inappropriate shocks, and implantable cardioverter-defibrillator programming

Transvenous ICD systems account for more than 95% of the total ICD in the CHD population. Approximately two-thirds were dual chamber systems.[63] Non-transvenous ICD were mostly used in children and patients with univentricular hearts.[306] The S-ICD has been used in CHD patients with limited venous access to the ventricle or intracardiac shunts.[261,306] Data demonstrate increased inappropriate shock rates due to oversensing.[310] As the device lacks the features of antitachycardia pacing and antibradycardia pacing, indications are limited in CHD patients.

Higher complication rates of ICD therapy during short and long-term follow-up have been reported.[294–300] including lead malfunction, device related problems, and infections including endocarditis.[63] In addition, a higher prevalence of inappropriate shocks has been reported. This may at least in part be explained by the increased incidence of SVT and active lifestyles with higher risks of lead failure. Most studies report inappropriate shock rates that equal or exceed

the appropriate shock rates in the young CHD population. In a meta-analysis of 518 ICD patients, inappropriate shocks were reported in 25% of patients during 3.8 years follow-up (i.e. 6.5% per year).[63]

Therapy with ICD may influence quality of life and psychosocial functioning, especially in patients receiving repeated shocks. Patients with CHD and ICD reported a high level of anxiety related to shocks which in turn was associated with depression and sexual dysfunction.[86]

Data imply that optimization of ICD settings and individual programming is of paramount importance to reduce the number of shocks. Recent trials have demonstrated that increasing detection heart rates and detection duration resulted in a decrease of inappropriate shocks while also decreasing mortality.[311,312] Finally, antitachycardia pacing programming has been shown to be safe and effective in CHD patients resulting in a reduced number of shocks.[305] Discriminator algorithms, based on morphology, rhythm stability, and onset analysis, can be useful to discriminate ATs.[313] There are no data to support the use of dual chamber ICD instead of single chamber ICD in young CHD patients.[312] Catheter ablation or antiarrhythmic medication may be indicated in ICD patients with AT and in selected patients with VT.[20,30]

Cardiac resynchronization in patients with congenital heart disease

Dyssynchronous heart failure and cardiac resynchronization therapy

Pathophysiology

Electromechanical dyssynchrony may cause a sequence of events that results in pathological ventricular remodelling leading to dyssynchronous heart failure.[314] Dyssynchrony amenable to CRT is typically caused by an electrical activation delay between one and the other ventricular wall, either caused by bundle branch block or conventional, typically RVP. It is characterized by clustering of early and late contracting segments, respectively. Early electrical activation and mechanical contraction causes initial stretch of late activated segments. Local myocardial work is decreased in early contracting sites that have a low local preload and is increased in late sites where preload is enhanced by preceding stretch.[314] Part of the myocardial work is wasted. At the same time intra-ventricular mechanical dyssynchrony initiates partially asymmetric cellular remodelling on multiple levels, which is reversible after CRT.[315,316] Mechanical dyssynchrony may, however, also be caused by contractile disparity and such dyssynchrony is not amenable to CRT.[317] Studies in adult patients with idiopathic or ischaemic dilated cardiomyopathy indicate that the presence of a left bundle branch block ECG pattern is a major prerequisite of CRT response.[286,318–320]

Epidemiology

Conventional ventricular pacing rather than bundle branch block is the major source of systemic ventricular dyssynchrony in CHD.[317] The exact prevalence of dyssynchronous heart failure in CHD is unknown. In adults with a Syst RV, 9.3% of patients after Mustard or Senning procedures and 6.1% of those with CC-TGA would be candidates for CRT using current indication criteria.

The most frequent conduction disturbance in CHD is right bundle branch block in the setting of a sub-pulmonary right ventricle. Right ventricular electromechanical dyssynchrony and mechanical inefficiency was recently described.[321] However, CRT has been mainly reserved for patients with systemic ventricular dysfunction so far. Conventional pacing-associated dyssynchronopathy may be prevented by septal or His pacing and may substantially decrease the number of CRT candidates among CHD patients.

Imaging

Newer imaging techniques may guide appropriate selection of candidates for CRT with focus on the so-called 'classical strain pattern'[320] including speckle tracking derived strain analysis and tissue or vector velocity imaging. However, the role of echocardiography even applying advanced speckle tracking has been quite disappointing according to the results of the ECHO-CRT trial although it may be helpful in selected patients with CHD. Particularly in patients with subaortic right ventricles there is need of preprocedural imaging of coronary sinus anatomy.

Clinical studies

Numerous studies on CRT in adults with idiopathic and ischaemic cardiomyopathy have confirmed restoration of a normal or near-normal electromechanical activation pattern, increase in myocardial energy efficiency, reverse structural and cellular remodelling, functional improvement, and a reduction in heart failure-associated morbidity and mortality.[318,322,323] Limited evidence suggests a similar role of CRT in patients with CHD, although none of the larger studies was prospective or randomized.[324–327] Efficacy of CRT in CHD may vary with the underlying structural and functional substrate, such as anatomy of the systemic ventricle (left, right, or single), presence and degree of structural systemic AV valve regurgitation, primary myocardial disease or scarring, and type of electrical conduction delay. Follow-up was largely limited to a few months, precluding an analysis of the impact of CRT on long-term morbidity and mortality. Outcome description was mainly limited to metrics of systemic ventricular function. The following observations may be highlighted for CRT in adult CHD[55,98,283,327–331]:

(1) Conventional single-site ventricular pacing with systemic ventricular dyssynchrony was the most prevalent (~65%) indication for CRT.
(2) Presence of left bundle branch block along with a systemic LV in the absence of ventricular pacing was a minor indication for CRT (9–17%) while right bundle branch block in the presence of a Syst RV was an even less common indication for CRT (5–7%).
(3) The majority of patients (58%) had NYHA Class II symptoms.
(4) An increase in systemic ventricular EF following CRT ranged between 6 and 20% while presence of a systemic LV was an independent predictor of a greater improvement in systolic systemic ventricular function.
(5) The best response to CRT was observed in patients with a systemic LV who were upgraded to CRT from conventional RVP.
(6) CRT was effective in combination with corrective or palliative cardiac surgery, particularly when performed to reduce systemic AV valve regurgitation.
(7) The proportion of CRT devices with defibrillation features was low (<25%).
(8) Patients with CHD awaiting heart transplantation may benefit from screening for potentially reversible mechanical dyssynchrony.

Little is known about indications and role of sub-pulmonary right ventricular resynchronization. A few studies on acute CRT effect and

Table 15 Specific recommendations for CRT in adults with congenital heart disease

Recommendations	Consensus statement	References
CRT is indicated for adults with CHD, a systemic left ventricular EF ≤35%, sinus rhythm, wide QRS complex ≥150 ms with complete left bundle branch block QRS morphology (spontaneous or paced) and NYHA function Class II—ambulatory IV.		332–334
CRT is indicated for adults with CHD who have a systemic ventricular EF ≤35%, intrinsic narrow QRS complex, NYHA function Class I—ambulatory IV and are undergoing new device placement or replacement with anticipated requirement for significant (>40%) ventricular pacing. Single site pacing from the systemic ventricular apex/mid-lateral wall may be considered as alternative.		319,98,283–285,328–331
CRT can be useful for patients with a systemic LV with an EF ≤35%, sinus rhythm, wide QRS complex (120–149 ms) with complete left bundle branch block QRS morphology (spontaneous or paced) and NYHA function Class II—ambulatory IV.		332–334
CRT can be useful for patients with a systemic RV with an EF ≤35%, NYHA function Class II-ambulatory IV and wide QRS complex ≥150 ms with a complete right bundle branch block QRS morphology (spontaneous or paced).		332–334,319,335,336,337
CRT can be useful for patients with a single ventricle with an EF ≤35%, NYHA function Class II—ambulatory IV and wide QRS complex ≥150 ms due to intraventricular conduction delay causing either a complete right or left bundle branch block QRS morphology (spontaneous or paced).		334
CRT can be useful for patients who have a systemic ventricular EF >35%, intrinsic narrow QRS complex, NYHA function Class I—ambulatory IV and are undergoing new device placement or replacement with anticipated requirement for significant (>40%) ventricular pacing. Single site pacing from the systemic ventricular apex may be considered as alternative.		98,283–285,319,331
CRT may be considered for patients with a systemic RV with an EF ≤35%, sinus rhythm, wide QRS complex (120—149 ms) with complete right bundle branch block QRS morphology (spontaneous or paced) and NYHA function Class II—ambulatory IV.		332–334,336,337,285,319
CRT may be considered for patients with a single ventricle with an EF ≤35%, sinus rhythm, wide QRS complex (120–149 ms) due to intraventricular conduction delay causing either a complete right or left bundle branch block QRS morphology (spontaneous or paced) and NYHA function Class II—ambulatory IV.		334
CRT may be considered in adults with congenital heart disease regardless of systemic ventricular morphology with a progression of systolic systemic ventricular dysfunction and/or dilatation or expectation of such development without a specific EF limit, NYHA function Class I—ambulatory IV with a wide QRS complex ≥150 ms due to a significant electrical activation delay within the systemic ventricle causing either a complete right or left bundle branch block QRS morphology (spontaneous or paced) who are undergoing other cardiac surgery, especially if thoracotomy access is needed for lead implantation.		332–334
CRT may be considered in patients with a systemic RV and significant tricuspid valve regurgitation without a specific EF limit, NYHA function Class I—ambulatory IV, wide QRS complex ≥150 ms with a complete right bundle branch block QRS morphology (spontaneous or paced) undergoing surgery for significant tricuspid valve regurgitation.		337
CRT may be considered for patients with a severe subpulmonary RV dysfunction and dilatation despite interventions to decrease RV volume overload, NYHA function Class II—ambulatory IV and wide QRS complex ≥150 ms due to a complete right bundle branch block.		330,338–340
CRT indication may be considered carefully and individually in patients in patients with NYHA function Class IV and severe ventricular dysfunction who would otherwise be candidates for heart transplantation or mechanical circulatory support.		333
CRT is not indicated in patients with a narrow QRS complex (<120 ms) without major electrical activation delay within the failing ventricle.		338
CRT is not indicated for patients whose co-morbidities and/or frailty limit survival with good functional capacity to less than 1 year		338

CHD, congenital heart disease; CRT, cardiac resynchronization therapy; EF, ejection fraction; LV, left ventricle; NYHA, New York Heart Association; RV, right ventricle.

Table 16 Summary of clinical studies on CRT in CHD

Author (references)	*n*	Age	Desing FU (m)	CHD %	Syst. RV %	Single V %	Conv. pacing %	QRS Pre ms	EF Pre	NYHA III–IV Pre %	Epic CRT %	Non resp %	QRS post ms	EF Post	Main feature
Janousek *et al.*[331]	8	15.0[a]	SC, P 17.4[b]	100	100	0	75	161[a]	18	12.5	37.5	NA	116[a]	30	First study on CRT in systemic right ventricle
Dubin *et al.*[333]	103	12.8[b]	MC, R 4[b]	70.9	16.5	6.8	44.7	166[a]	26	37.9	46.6	10.7	126[a]	40	First large study on CRT in congenital heart disease
Khairy *et al.*[324]	13	7.8[a]	SC, R 16, 5[a]	100	30.8	0	100	NA	31	NA	100	11.1	NA	51	Impaired EF + conventional pacing in all
Moak *et al.*[280]	6	11.3[a]	SC, R 10[b]	33.3	0	0	100	204[a]	34	NA	66.7	0	138[a]	60	Super-response after upgrade from RVP to CRT
Cecchin *et al.*[332]	60	15.0[b]	SC, R 8[b]	76.7	15	21.7	68.3	149[a]	36	31.7	63.3	10	120[a]	42	Largest reported single ventricular patient group
Jauvert *et al.*[330]	7	24.6[a]	SC, P 19, 4[a]	100	100	0	71.4	160[a]	NA	100	28.6	NA	120[a]	NA	CRT in systemic right ventricle
Janousek *et al.*[334]	109	16.9[a]	MC, R 7.5[b]	79.8	33	3.7	77.1	160[b]	30	45.9	33	13.7	130[b]	41	Largest study with different substrates
Thambo *et al.*[336]	9	36.6[a]	SC, P 6[b]	100	0	0	0	164[a]	50	NA	33	NA	NA	56	Postoperative TOF
Sakaguchi *et al.*[339]	20	22[a]	SC, R ≥6	100	35	30	60	183[a]	34	25	NA	NA	132[a]	NA	Effect of CRT according to systemic ventricle type

CHD, congenital heart disease; Conv pacing, conventional pacing prior to CRT; CRT, cardiac resynchronization therapy; EF, ejection fraction; Epic CRT, epicardial CRT; FU, follow-up; ms, milliseconds; M, multicentre; NA, non-available; Non resp, non-responder; NYHA, New York Heart Association; P, prospective; Pre, prior to CRT; Post, following CRT; R, retrospective; RVP, right ventricular pacing; SC, single centre; Single V, single ventricle; Syst RV, systemic right ventricle; TOF, tetralogy of Fallot.
[a]Mean value.
[b]Median value.

one well-documented case report suggest acute improvement of right ventricular function and long-term reverse remodelling, respectively.[328–330]

Technical aspects

Anatomical constraints preclude implantation of transvenous CRT systems in a significant proportion of patients with CHD necessitating thoracotomy or hybrid lead implantation. A hybrid approach is typically used for patients with TGA after the Mustard or Senning procedures. Total non-transvenous lead implantation is mostly required for univentricular hearts with lead placement on opposing ventricular walls which is technically very challenging.

Selection of optimal pacing site may be guided by recording the delay in local electrical activation with respect to QRS onset. None of the CHD studies to date have specifically explored the usefulness of AV and VV delay optimization during CRT follow-up. However, in non-responders to CRT and in those in need of atrial pacing, evaluation of AV and VV delay may be justified to correct suboptimal device settings.

No studies are so far available on the longevity of CRT devices in the specific CHD population. Due to higher complexity these devices may be even more susceptible to typical pacing complications.

Indications

Indications for CRT in adults with CHD have recently been summarized.[30] They are based on current European and North American heart failure and device therapy guidelines addressing patients with idiopathic or ischaemic dilated cardiomyopathy and a review of data on patients with congenital heart disease (*Tables 15* and *16*).[98,283–285,319,331,335]

Optimal medical therapy should be an integral part of heart failure management prior to CRT implantation. In any individual patient, the need for primary preventive defibrillation capability (CRT-D) should be assessed applying current criteria.

Areas for future research

The current consensus statement reflects the tremendous progress achieved within the last 20 years in diagnosis and management of all types of arrhythmias in grown-ups with CHD. In addition, it documents the combined efforts of paediatric and adult electrophysiologists in the care of this growing population.

There are, however, still significant limitations in understanding and managing of the various types of arrhythmias that need to be overcome in the future. Although nowadays the vast majority of CHD can be repaired by surgical or interventional therapy, there is still a lack of understanding how arrhythmias develop in our CHD patients. Further efforts start with optimal timing of the procedure as well as the type of intervention for a given CHD, particularly in order to prevent development of arrhythmias at all.

Facing the various problems encountered with pharmacological therapy for rhythm control in young individuals with CHD, curative treatment by catheter ablation or device implantation seems preferable. Further developments in mapping technology allowing precise identification of the arrhythmia substrate may improve results. The same applies to ablation technology allowing target-specific ablation.

Further progress in leadless pacing allowing for AV synchrony in combination with a S-ICD may help to avoid problems related to lead failure and infection as currently encountered in device therapy in adult CHD patients. Indications for CRT in patients with CHD as stated in this document have been mainly derived from guidelines for adult patients with idiopathic- or ischaemic-dilated cardiomyopathy and adapted to the diversity of structural and functional CHD substrates. Research should focus on improvement of both selection of proper CRT candidates as well as the CRT application which may eventually identify new patient groups that may profit from CRT.

The goal of future research on arrhythmias in grown-ups with CHD is to rationalize the wide range of therapeutic modalities to the diverse underlying substrates. All efforts should focus on the treatment of the underlying arrhythmia, to postpone or avert heart failure, to prolong life and improve quality of life, and to prevent SCD.

Conclusions

The present consensus statement summarize knowledge and provide recommendations on diagnosis and treatment of arrhythmias in patients with congenital heart defects. This position paper mainly addresses arrhythmias in adult with congenital heart disease, because, in many cases, the anatomy and management of arrhythmias in adult patients cannot directly be applied to patients with congenital heart disease. There are, however, still significant limitations in understanding and managing of the various types of arrhythmias that need to be overcome in the future. Although nowadays the vast majority of CHD can be repaired by surgical or interventional therapy, there is still a lack of understanding how arrhythmias develop in our congenital heart disease patients. Further developments in mapping technology allowing precise identification of the arrhythmia substrate may improve results.

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