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

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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 maintenance 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
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 level of evidence (A, B, and C) to recommendations.

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

Table 1 Scientific rationale of recommendations

<table>
<thead>
<tr>
<th>Definitions where related to a treatment or procedure</th>
<th>Consensus statement</th>
<th>Symbol</th>
</tr>
</thead>
<tbody>
<tr>
<td>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)</td>
<td>Recommended/indicated</td>
<td><img src="https://example.com/green-heart.png" alt="Green Heart" /></td>
</tr>
<tr>
<td>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</td>
<td>May be used or recommended</td>
<td><img src="https://example.com/yellow-heart.png" alt="Yellow Heart" /></td>
</tr>
<tr>
<td>Scientific evidence or general agreement not to use or recommend treatment or procedure</td>
<td>Should NOT be used or recommended</td>
<td><img src="https://example.com/red-heart.png" alt="Red Heart" /></td>
</tr>
</tbody>
</table>

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.

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. 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 venous 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. An abnormal dysfunctional sinus node can be found in usual position in patients with absence of the right superior caval vein.

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 isomorphic arrangement of the atrial appendages, i.e. left isomerism, due to displacement of the specialized conduction system with abnormal 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.
development of the central fibrous body. 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.

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).

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.

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.

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. 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). The conduction system is particularly susceptible to injury during surgical and catheter procedures in patients with ASDs and CC-TGA. In addition, peri-membranous 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 operation1,11 (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
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. electrophysiological 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>
<thead>
<tr>
<th>Table 2</th>
<th>Initial work-up of the CHD patient with arrhythmia</th>
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<tbody>
<tr>
<td>History</td>
<td>Underlying anatomy</td>
</tr>
<tr>
<td></td>
<td>Previous interventions/operations</td>
</tr>
<tr>
<td></td>
<td>Arrhythmia characteristics (onset, duration, symptoms etc.)</td>
</tr>
<tr>
<td></td>
<td>Antiarrhythmic and non-antiarrhythmic medication</td>
</tr>
<tr>
<td></td>
<td>Chronic anticoagulation</td>
</tr>
<tr>
<td></td>
<td>Functional class (NYHA)</td>
</tr>
<tr>
<td>Physical examination</td>
<td>Signs of haemodynamic compromise/heart failure</td>
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<td></td>
<td>Scars from previous surgery/vascular access sites</td>
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<tr>
<td></td>
<td>Implanted device</td>
</tr>
<tr>
<td></td>
<td>Signs of infection, fever</td>
</tr>
<tr>
<td>Transthoracic/</td>
<td>Size and function of the systemic and subpulmonary ventricle</td>
</tr>
<tr>
<td>transoesophageal</td>
<td></td>
</tr>
<tr>
<td>echocardiogram</td>
<td>Size and volume of cardiac chambers</td>
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<td></td>
<td>Thrombus, residual intracardiac shunt</td>
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<tr>
<td></td>
<td>Valvular function, gradients, and regurgitation</td>
</tr>
<tr>
<td>Cardiac CT/MRI/</td>
<td>For complete assessment of individual anatomy and haemodynamics in</td>
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<tr>
<td>haemodynamic catheterization/angiography</td>
<td>univentricular hearts, systemic right ventricle, TOF, and variants</td>
</tr>
<tr>
<td>Blood chemistry</td>
<td>Infection signs</td>
</tr>
<tr>
<td></td>
<td>Full blood count, plasmatic coagulation</td>
</tr>
<tr>
<td></td>
<td>Thyroid, liver, and kidney function</td>
</tr>
</tbody>
</table>

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: roadmap
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. 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. 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. Up to 20% of Ebstein patients harbour one or more accessory pathways. 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. 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. 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. 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%). 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. 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 (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. 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.

An ASD is often present in patients with CHD and may contribute to development of atrial tachyarrhythmias. In older patients with unoperated ASD, a history of AF was present in 19% and AF in 61%. 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 terminals. 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.
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. The long-term outcome after ASD closure at young age is excellent.

The Fontan circulation in patients with a univentricular heart is prone to atrial tachyarrhythmias both located at the lateral right atrial wall and at the CTI. Prevalence of MRAT was higher in the first 2 years after Fontan and increased again gradually in later childhood. The atrio pulmonary 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. The newer Fontan modifications including intracardiac lateral tunnel, extracardiac lateral tunnel, or extracardiac conduit were significantly less arrhythmogenic.
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 atrio pulmonary 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 proarrhythmic 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 CHA2DSc 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.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.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
Arrhythmias in congenital heart disease

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

Sustained ventricular arrhythmias in adults with congenital heart disease (ACHD) encompass monomorphic VT, polymorphic VT, and VF. 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.

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.

Additional data on the prevalence of any ventricular arrhythmias came were derived from ICD interrogation studies. There were remarkable differences in the type of documented ventricular arrhythmias. Monomorphic VTs constituted 81.5% of all ventricular arrhythmias in TOF. 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%. Monomorphic VT in TOF were typically rapid with median heart rates of 213 bpm.

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 polymorphic VT/VF associated with advanced ventricular dysfunction with or without surgical scars.

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. 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. Consistent data in large populations of patients with CHD have shown a percentage of SCD among deaths in this population of approximately 20–25%. The risk of SCD increases with complexity of the disease. 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.

Sudden cardiac death associated with bradyarrhythmias and atrial arrhythmias
The risk of SCD from sinus or junctional bradycardia leading to sinus arrest is controversial. The bradycardia–tachycardia syndrome is considered to predispose for SCD. SCD due to AV block represents probably less than 5% of all SCD in adult CHD patients. Prolonged asystole or bradycardia triggered VT are potential causes of SCD in untreated AV block. 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.

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. 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.

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. 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. 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.

The risk of inappropriate shocks in CHD patients with primary prevention ICDs must be outweighed against the benefits. 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 definitely higher compared to adults without CHD.

Value of subcutaneous ICD (S-ICD) in CHD patients lacking need for permanent pacing needs to be established.

Bradyarrhythmias in patients with congenital heart disease
Congenital atrioventricular block
Congenital complete heart block (CCHB), first described by Morquio in 1901 and documented on ECG in 1908, is due to disruption of the electrophysiological continuity between atria and ventricles. 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.

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. 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.
Post-operative atrioventricular block

Post-surgical atrioventricular block was first explored in detail by Lillehei in 1963, and still occurs in 1–3% of patients immediately after surgery or early in the post-operative period and has been reported only occasionally months or even years thereafter. Early post-operative CHB can be transient or permanent. Most patients with early post-operative AV block recover spontaneously within the first 7 to 10 days. Therefore, at least 7 days of observation before pacemaker implantation is strongly recommended. During this period, temporary pacing wires may be inserted to maintain adequate chronotropy. Late recovery has also been demonstrated by several investigators. First degree, second degree AV block combined with complete right bundle branch block have been observed after recovery from complete AV block. 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.

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 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 and left-sided juxtaposition of the atrial appendages. In most patients, SND is secondary to surgical procedures such as repair of sinus venosus ASDs, Glenn shunts, Fontan operation, and Senning or Mustard procedures. 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 with increasing risks of atrial tachyarrhythmias, the tachycardia–bradycardia syndrome. Although there has not been direct evidence of the relationship between SND and sudden death in patients with congenital heart disease, uncontrolled atrial tachyarrhythmias may increase the risk of heart failure, mortality, and SCD, particularly in those with ventricular dysrythmias. 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. 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.

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

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Consensus</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>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.</td>
<td>Expert consensus</td>
<td>48,144</td>
</tr>
<tr>
<td>Electrical cardioversion is recommended for haemodynamically unstable SVT (caution for sinus node dysfunction and impaired ventricular function with need for chronotropic or inotropic support).</td>
<td>Expert consensus</td>
<td></td>
</tr>
<tr>
<td>In patients with AV-nodal dependent SVT, adenosine is recommended for acute management (unless contraindicated and caution for sinus node dysfunction).</td>
<td>Expert consensus</td>
<td></td>
</tr>
<tr>
<td>Atrial overdrive pacing (via oesophagus or endocardial) may be considered for conversion of haemodynamically stable AVNRT/AVRT or atrial flutter.</td>
<td>Expert consensus</td>
<td></td>
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</tbody>
</table>

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. 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. Additional agents included adenosine, sotalol, amiodarone, flecainide, verapamil, metoprolol, and procainamide. In 29 patients with AF or AVRT dofetilide (not available in Europe) successfully reestablished sinus rhythm in 12, but side effects were significant (41%). Quinidine, disopyramide, and sotalol, have been associated with increased all-cause mortality. 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, additional agents included adenosine, sotalol, amiodarone, flecainide, verapamil, metoprolol, and procainamide.

**Table 5** Recommendations for pharmacological therapy of supraventricular tachycardia

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Consensus</th>
<th>References</th>
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<tbody>
<tr>
<td>In patients with CHD and AT/AF rhythm control is recommended as the preferred initial strategy.</td>
<td>Expert consensus</td>
<td></td>
</tr>
<tr>
<td>Catheter ablation is recommended as first line therapy and preferred to long-term pharmacological treatment, in case of amenable, circumscribed substrates.</td>
<td>Expert consensus</td>
<td>30,145</td>
</tr>
<tr>
<td>In AT/AF and failed conversion or stabilization of sinus rhythm, pharmacological AV blockade using β-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.</td>
<td>Expert consensus</td>
<td>30,48,146,147</td>
</tr>
<tr>
<td>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-term amiodarone therapy is not advised in young CHD patients.</td>
<td>Expert consensus</td>
<td>146,147</td>
</tr>
<tr>
<td>Amiodarone should be used with caution in cyanotic CHD, low body weight, hepatic, thyroid, or pulmonary disease or prolonged QT interval.</td>
<td>Expert consensus</td>
<td>148–150</td>
</tr>
<tr>
<td>β-blockers, if tolerated, may be administered to reduce ventricular tachyarrhythmia rapid AV conduction burden in selected CHD patients with AT.</td>
<td>Expert consensus</td>
<td>61</td>
</tr>
<tr>
<td>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.</td>
<td>Expert consensus</td>
<td></td>
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back-up pacing should be available prior to attempted pharmacologi-

cal 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 or i.v. adenosine (SVT) should be the preferred forms of acute
therapy. Electrical cardioversion is effective in terminating
AF or i.v. adenosine (SVT) should be the preferred forms of acute
therapy.

**Table 5** Continued

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<th>Recommendations</th>
<th>Consensus</th>
<th>References</th>
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<tr>
<td>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.</td>
<td>Expert consensus</td>
<td>151</td>
</tr>
<tr>
<td>Indications for anticoagulation therapy for AT or atrial flutter are not different as for patients with AF. Oral Class I agents are not recommended in the treatment of AT/AF in patients with coronary artery disease or decreased ventricular function.</td>
<td>Expert consensus</td>
<td>152–154</td>
</tr>
<tr>
<td>Normal systemic ventricular function: RV SF &gt; 50%, LV &gt; 60%. AF, atrial fibrillation; AT, atrial tachycardia; AV, atroventricular; CHD, congenital heart disease; ICD, implantable cardioverter-defibrillator; LV, left ventricular; RV, right ventricular; SCD, sudden cardiac death; SVT, supraventricular tachycardia; VT, ventricular tachycardia.</td>
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**Table 6** Recommendations for pharmacological therapy of ventricular tachycardia

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<tr>
<th>Recommendations</th>
<th>Consensus</th>
<th>References</th>
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<tbody>
<tr>
<td>Electrical cardioversion is recommended for acute termination of haemodynamic stable/unstable VT. If not possible, intravenous amiodarone or procainamide may be considered. 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 tachyarythmia induced ventricular dysfunction. Antiarrhythmic drugs may 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.</td>
<td>Expert consensus</td>
<td>155</td>
</tr>
<tr>
<td>ICD, implantable cardioverter-defibrillator; SCD, sudden cardiac death; VT, ventricular tachycardia.</td>
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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. 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. Dofetilide therapy either maintained sinus rhythm or improved arrhythmia control in 49% of patients at a median follow-up of 3 years, 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 (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
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. Specific anatomic groups have been shown a particular high risk of thrombo-embolic complications, including uncorrected cyanotic heart disease (23%), Eisenmenger physiology (5%), native ASDs (4%), and the Fontan circulation (4%). In 25% of cases, stroke was associated with loss of sinus rhythm. Specific anatomic 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 complications. In lower risk patients strategies similar to those used for other variants of structural heart disease, the CHA2DS2-VASc risk-factor based approach appears appropriate. Effective stroke prevention in patients with CHA2DS2-VASc score ≥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.

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. 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. 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. Expertise in EP ablation is also mandatory if transseptal or transbaffle punctures are needed to access the pulmonary venous atrium. This procedure should be reserved for specialized, dedicated centres with surgical back-up as an alternative to retrograde transaortic access. 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. 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. 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. Catheter ablation of accessory AV connections is the preferred therapeutic option with lower acute success rates and higher recurrence rates compared with patients with normal hearts. (Table 7). With further development of catheter and sheath technology and by the advent of 3D mapping systems, 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. 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.

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.
Table 7  Recommendations for supraventricular tachyarrhythmia catheter ablation

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Consensus statement</th>
<th>References</th>
</tr>
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<tbody>
<tr>
<td>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 scenariosa</td>
<td>45,169–171</td>
<td></td>
</tr>
<tr>
<td>In simple CHD substrates, ablation of symptomatic MRAT is generally recommended as an alternative to antiarrhythmic drugs and/or electrical cardioversionsb</td>
<td>172–174</td>
<td></td>
</tr>
<tr>
<td>In complex CHD substrates, ablation of symptomatic MRAT is recommended as an alternative to antiarrhythmic drugs and/or electrical cardioversion at experienced centresa</td>
<td>176,178</td>
<td></td>
</tr>
<tr>
<td>The use of 3D mapping systems, and irrigated tip catheters for MRAT ablation, is recommended in CHD patients</td>
<td>172,175–177</td>
<td></td>
</tr>
<tr>
<td>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 centresa</td>
<td>45,172,174,175,177</td>
<td></td>
</tr>
<tr>
<td>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 safetyb</td>
<td>45,50–52,55</td>
<td></td>
</tr>
<tr>
<td>Atrial fibrillation ablation may be considered in selected simple CHD patients, at experienced centresc</td>
<td></td>
<td></td>
</tr>
<tr>
<td>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</td>
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Table 7  Continued

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<th>Recommendations</th>
<th>Consensus statement</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>Catheter ablation is not recommended for atrial tachyarrhythmias that can be controlled medically in the early post-surgical period (&lt;3 months).</td>
<td>Expert consensus</td>
<td></td>
</tr>
<tr>
<td>Catheter ablation is not recommended for asymptomatic non-sustained runs of atrial tachycardia.</td>
<td>Expert consensus</td>
<td></td>
</tr>
</tbody>
</table>

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

*aWhen complication rates are expected to be low (larger patients, conventional vascular accesses ...).
*bLike in smaller patients, unconventional vascular accesses ...
*cWhen post-ablative recurrence and complication rates are presumed to be within standard ranges published from non-CHD patients series.

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%. However, with increasing experience and implementation of electro-anatomical mapping systems results could be improved. 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.

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. Fibrosis due to longstanding cyanosis, pressure and/or volume overload and aging may provide the substrate for slow conduction within these anatomically defined isthmuses.

Patients with atrial tachyarrhythmias often harbour a multifactorial proarrhythmic state, 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.

Usually, the more complex the baseline substrate and the surgical procedure, the higher the chances of developing complex atrial or ventricular reentries. 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 ventriculotomy by using a transatrial–transpulmonary approach and
Arrhythmias in congenital heart disease

Table 8 Recommendations for VT ablation

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Consensus statement</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>Catheter ablation is indicated as additional therapy in ICD patients with CHD who present with incessant VT or electrical storm.</td>
<td>Catheter ablation should be considered</td>
<td>66–68,70</td>
</tr>
<tr>
<td>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.</td>
<td>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.</td>
<td>69,202,203</td>
</tr>
<tr>
<td>Surgical intraoperative cryo-ablation is recommended in TdF patients who undergo re-surgery and who have spontaneous or inducible VTs unless managed by preoperative ablation.</td>
<td>The use of 3D mapping systems to obtain a 3D reconstruction of all VT related anatomic isthmuses is recommended.</td>
<td>66,67,69,70</td>
</tr>
<tr>
<td>Catheter ablation should be considered for symptomatic monomorphic sustained VT in grown-up-congenital heart disease patients as an alternative to drug therapy.</td>
<td>Catheter ablation should be considered in grown-up congenital heart disease patients with frequent highly symptomatic PVCs, or PVCs associated with deteriorating ventricular function.</td>
<td>66,67,69</td>
</tr>
<tr>
<td>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).</td>
<td></td>
<td>67,69</td>
</tr>
</tbody>
</table>

Continued

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Consensus statement</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>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.</td>
<td>The use of irrigated tip catheter is recommended for linear lesions and hypertrophied myocardium.</td>
<td>66,67,202</td>
</tr>
<tr>
<td>Catheter ablation is not recommended for asymptomatic, infrequent PVC and non-sustained VT</td>
<td></td>
<td></td>
</tr>
<tr>
<td>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.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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.200 (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.178,200–207 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 echocardiography208 in regions with low amplitude signals to establish electrode to tissue contact and to differentiate from myocardial fibrosis. This type of catheter 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 be subdivided in localized NAFAT and the more prevalent
involve CTI. The same applies to patients after repair of TOF where MRAT and reach 90–95% for CTI-dependent MRAT or AF. Finally, empirical CTI ablation as well as a and activation mapping can be performed during sinus rhythm or unmappable circuits, a substrate mapping approach including voltage right atrial wall and in vicinity to the inferior caval vein. 'Figure-of-eight' circuits may serve as an additional critical substrate. In repaired CHD individuals, most non-CTI reentries tend to revolve around injured tissue as incisural tachycardias at the lateral and anterior right atrial wall. ‘Figure-of-eight’ circuits may complicate understanding and ablation of such tachycardias. Subtle abrupt electrogram changes may predict partial interruption of the double circuit.

In Fontan patients, MRAT tend to course along the entire lateral right atrial wall and in vicinity to the inferior caval vein. In patients after Mustard/Senning procedure MRAT predominantly involve CTI. 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. 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. Finally, empirical CTI ablation as well as a corridor between right lateral atriotomy scar and the IVC has been advocated in non-inducible CHD patients.

Acute ablation outcomes range between 80% in scar-related MRAT and reach 90–95% for CTI-dependent MRAT or AF. Electroanatomical mapping, locating the mid-diastolic isthmus, and adjusting the window of interest are important for ablation. Long-term success, however, may be impared 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. 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 identified as a risk factor for tachycardia recurrence.

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. High-output pacing, in order to track the course of the phrenic nerve with a electroanatomatic 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. Echocardiographic guidance is recommended for transseptal puncture in case of anatomical distortion confusing radiological orientation, in case of ASD closure devices or septal patches. There are reports that show that results of AF ablation in CHD patients do not significantly differ from non-CHD populations.

**Table 9 Requirements for complex CHD arrhythmia management**

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

CHD, congenital heart disease; CT, computed tomography; EP, electrophysiology; MRI, magnetic resonance imaging.
hypertrophied myocardium, proximity of the conduction system, and importantly, the protection of portions of anatomical isthmuses by prosthetic material. 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. The role of preventive dissection of potential arrhythmogenic isthmuses in TOF patients remains to be determined. (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, 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 acknowledged as a more gross strategical concept in avoiding development of surgical tachycardia substrates.

**Preventative surgery**  
Preventive or ‘prophylactic’ arrhythmia operations may be considered for patients with specific anatomic substrates as an effort to reduce the risk of late arrhythmias. 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 as significant risk for late atrial arrhythmias. 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.

**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. 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.

**Indications for surgery of arrhythmias in congenital heart disease**  
Surgery for arrhythmias associated 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 arrhythmia surgery may be implemented, especially for treatment of AF, incisional AT, AF, or VT.

**Rhythm 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. 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. 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. Extraction of redundant leads may facilitate access but can be challenging as some leads have been placed for >10 years or even longer. The low profile 4.1 Fr leads may help to preserve venous patency. 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.

Subcutaneous, intrathoracic, or epicardial defibrillator coils may be used in patients lacking access to a subpulmonary ventricle. 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 stinting 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 ventilator 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 superior vena cavae from the right atrium.269 If the ayzygos 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).

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 10 Recommendations for pacemaker implantation according to the underlying disease

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

AV, atrioventricular; CHD, congenital heart disease; ICD, implantable cardioverter-defibrillator.
primarily with clinical symptoms and cardiac function rather than pre-defined heart rate limits. In the last years, several guidelines for device implantation have been published with one study 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. 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. Guidelines for lead extraction 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. 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%). 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.

**Recommendations for implantable cardioverter-defibrillator therapy**

**Secondary prevention**
Evidence based guidelines for secondary prevention of SCD can be extrapolated to the CHD population. 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%. 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.

**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 within 3.3 ± 0.3 years, indicating the annual shock rate of 6.6%. 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 prevention therapy is mandatory. Further studies are needed to optimize the indications and the mode of therapy in this specific group of patients.

---

**Tables**

**Table 11** Recommendations for pacemaker implantation according to the underlying disease

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

AV, atrioventricular.
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. 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.

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 12  Recommendations for lead extraction

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Consensus statement</th>
</tr>
</thead>
<tbody>
<tr>
<td>Definite system infection, pocket infection, endocarditis (valvular, subvalvular, supravalvular, VSD) and occult gram positive bacteraemia</td>
<td></td>
</tr>
<tr>
<td>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</td>
<td></td>
</tr>
<tr>
<td>Life-threatening 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.</td>
<td></td>
</tr>
<tr>
<td>Persistent occult gram negative bacteraemia</td>
<td></td>
</tr>
<tr>
<td>Severe chronic pain at the device or lead insertion site</td>
<td></td>
</tr>
<tr>
<td>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</td>
<td></td>
</tr>
<tr>
<td>Device implantation would require more than four leads on one side or more than four leads through the SVC (ref 294)</td>
<td></td>
</tr>
<tr>
<td>Need for specific imaging techniques (e.g. MRI) without alternative techniques being available or to implant an MRI conditional system</td>
<td></td>
</tr>
<tr>
<td>Functional leads that due to their design pose a potential future threat to the patient if left in place</td>
<td></td>
</tr>
<tr>
<td>Abandoned leads that are functional but not being used or non-functional leads without an immediate threat to the patient</td>
<td></td>
</tr>
<tr>
<td>Only superficial or wound infection, without clear involvement of the device and/or leads</td>
<td></td>
</tr>
<tr>
<td>Chronic bacteremia due to a source other than the device system, when long-term antibiotics are required</td>
<td></td>
</tr>
<tr>
<td>Functional but redundant leads or non-functional leads if patients have a life expectancy of less than 1 year</td>
<td></td>
</tr>
<tr>
<td>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</td>
<td></td>
</tr>
</tbody>
</table>

MRI, magnetic resonance imaging. SVC, superior vena cava.
### Table 13: Summary of clinical studies on Implantable cardioverter-defibrillators in CHD

<table>
<thead>
<tr>
<th>Author (references)</th>
<th>Year</th>
<th>n</th>
<th>CHD</th>
<th>Age at implant (years)</th>
<th>Male (%)</th>
<th>Primary prevention (%)</th>
<th>Systemic ventricle function</th>
<th>FU (years)</th>
<th>Approp. shocks (%)</th>
<th>Inapp. shocks (%)</th>
<th>Comp. (%)</th>
<th>Main features</th>
</tr>
</thead>
<tbody>
<tr>
<td>Yap et al.</td>
<td>2007</td>
<td>64</td>
<td>Diverse CHD 63% ToF</td>
<td>37</td>
<td>67</td>
<td>39</td>
<td>17% impaired</td>
<td>3, 7</td>
<td>23, 4</td>
<td>40, 6</td>
<td>30</td>
<td>First multicentre study on ICD outcomes in adults with CHD.</td>
</tr>
<tr>
<td>Khairy et al.</td>
<td>2008</td>
<td>37</td>
<td>TGA</td>
<td>28</td>
<td>89</td>
<td>62</td>
<td>Mean EF 36%</td>
<td>3, 6</td>
<td>14</td>
<td>24</td>
<td>38</td>
<td>High rates of appropriate shocks in secondary but not primary prevention. SVA may be implicated in VT aetiology.</td>
</tr>
<tr>
<td>Khairy et al.</td>
<td>2008</td>
<td>121</td>
<td>ToF</td>
<td>333</td>
<td>60</td>
<td>56</td>
<td>Mean EF 54%</td>
<td>3, 7</td>
<td>31</td>
<td>25</td>
<td>30</td>
<td>High rates of appropriate and effective shocks in primary and secondary Prev.</td>
</tr>
<tr>
<td>Witte et al.</td>
<td>2008</td>
<td>25</td>
<td>ToF</td>
<td>NA</td>
<td>NA</td>
<td>28</td>
<td>3% impaired</td>
<td>1, 9</td>
<td>25</td>
<td>20</td>
<td>4</td>
<td>Comparison with dilated CM patients. Higher risk of inapp. therapies lower incidence of approp. therapies.</td>
</tr>
<tr>
<td>Khanna et al.</td>
<td>2011</td>
<td>73</td>
<td>Diverse CHD ToF 41%</td>
<td>68</td>
<td>64</td>
<td>27% impaired</td>
<td>Mean EF 36%</td>
<td>2, 2</td>
<td>19</td>
<td>15</td>
<td>15</td>
<td>Single centre experience. Low risk implant complications.</td>
</tr>
<tr>
<td>Koyak et al.</td>
<td>2012</td>
<td>136</td>
<td>Diverse CHD ToF 41%</td>
<td>67</td>
<td>50</td>
<td>51% moderate to severe impaired</td>
<td>Mean EF 54%</td>
<td>4, 6</td>
<td>32</td>
<td>30</td>
<td>29</td>
<td>Highest risk of appropriate ICD shocks: secondary Prev. indication, coronary artery disease and symptomatic NSVT.</td>
</tr>
<tr>
<td>Backhoff et al.</td>
<td>2014</td>
<td>12</td>
<td>TGA</td>
<td>30, 3</td>
<td>100</td>
<td>100</td>
<td>NA</td>
<td>3, 5</td>
<td>8</td>
<td>25</td>
<td>17</td>
<td>Inappropriate ICD shocks due to rapidly conducted atrial reentrant tachycardia. Sensing failure frequently observed</td>
</tr>
<tr>
<td>Jordan et al.</td>
<td>2014</td>
<td>1683</td>
<td>Diverse CHD septal defects</td>
<td>NA</td>
<td>NA</td>
<td>70</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NCDR-ICD Registry. Largest pooled assessment for CHD and pediatric ICD populations.</td>
</tr>
<tr>
<td>Kella et al.</td>
<td>2014</td>
<td>59</td>
<td>Diverse CHD ToF 35%, 75%</td>
<td>69</td>
<td>53</td>
<td>56% EF ≤ 35%</td>
<td>Mean EF 54%</td>
<td>3, 2</td>
<td>20</td>
<td>22</td>
<td>NA</td>
<td>Patients with non-ToF congenital lesions are significantly less likely to receive appropriate ICD therapy than those with ToF.</td>
</tr>
<tr>
<td>Santharam et al.</td>
<td>2017</td>
<td>42</td>
<td>Diverse CHD 50% ToF</td>
<td>55</td>
<td>38</td>
<td>NA</td>
<td>Mean EF 54%</td>
<td>5</td>
<td>14</td>
<td>26</td>
<td>14</td>
<td>Long-term follow-up. Significant incidence of complications.</td>
</tr>
<tr>
<td>Moore et al.</td>
<td>2016</td>
<td>21</td>
<td>Diverse. 52% single ventricle</td>
<td>62</td>
<td>67</td>
<td>LV EF 41% RV EF 33%</td>
<td>Mean EF 54%</td>
<td>1, 2</td>
<td>5</td>
<td>21</td>
<td>5</td>
<td>Largest multicentre study of S-ICD implantation for CHD.</td>
</tr>
<tr>
<td>Buber J et al.</td>
<td>2016</td>
<td>18</td>
<td>TGA</td>
<td>26</td>
<td>83</td>
<td>100</td>
<td>62% moderate to severe impaired</td>
<td>4</td>
<td>5</td>
<td>55</td>
<td>28</td>
<td>Atrial arrhythmias were the most common cause for ICD shocks. VT infrequent.</td>
</tr>
</tbody>
</table>

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.
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. 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. 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. ICD implantation should be considered in selected patients with TOF and multiple risk factors for SCD, including LV dysfunction, non-sustained VT, QRS duration \( \geq 180 \) ms, or inducible sustained VT on VPS. 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 \( \geq 140 \) ms or severe systemic AV valve regurgitation. ICD therapy may be considered for non-hospitalized adults with CHD awaiting heart transplantation. ICD is contraindicated in patients with CHD with life expectancy with an acceptable functional status \( \leq 1 \) 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.

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.

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. Non-transvenous ICD were mostly used in children and patients with univentricular hearts. The S-ICD has been used in CHD patients with limited venous access to the ventricle or intracardiac shunts. Data demonstrate increased inappropriate shock rates due to oversensing. 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 including lead malfunc-
tion, device related problems, and infections including endocarditis. 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

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Table 14  Recommendations for ICD therapy in adults with CHD

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Consensus statement</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>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.</td>
<td></td>
<td>57,58,61,295,305,306</td>
</tr>
<tr>
<td>ICD is recommended for patients with CHD with symptomatic sustained VT who have undergone haemodynamic and electrophysiological evaluation.</td>
<td></td>
<td>57,58,61,143,295,296</td>
</tr>
<tr>
<td>ICD is recommended in adults with CHD and a systemic LVEF ( \leq 35 )%, biventricular physiology and NYHA functional Class II or III.</td>
<td></td>
<td>20,307,308</td>
</tr>
<tr>
<td>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.</td>
<td></td>
<td>56,57,58,59,300</td>
</tr>
<tr>
<td>ICD implantation should be considered in selected patients with TOF and multiple risk factors for SCD, including LV dysfunction, non-sustained VT, QRS duration ( \geq 180 ) ms, or inducible sustained VT on VPS.</td>
<td></td>
<td>33,56,57,308</td>
</tr>
<tr>
<td>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 ( \geq 140 ) ms or severe systemic AV valve regurgitation.</td>
<td></td>
<td>33,59,61,299</td>
</tr>
<tr>
<td>ICD therapy may be considered for non-hospitalized adults with CHD awaiting heart transplantation.</td>
<td></td>
<td>308,309</td>
</tr>
<tr>
<td>ICD is contraindicated in patients with CHD with life expectancy with an acceptable functional status ( \leq 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.</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

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.
the appropriate shock rates in the young CHD population. In a meta-
alysis 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 dysfunc-
tion.66

Data imply that optimization of ICD settings and individual pro-
gramming 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 inap-
propriate shocks while also decreasing mortality.311,312 Finally, antitachy-
cardia 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.113 There are no data
to support the use of dual chamber ICD instead of single cham-
ber ICD in young CHD patients.312 Catheter ablation or antiarrhyth-
mic 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

Dysynchronous heart failure and cardiac
resynchronization therapy

Pathophysiology

Electromechanical dyssynchrony may cause a sequence of events
that results in pathological ventricular remodelling leading to dyssyn-
chronous 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 conven-
tional, 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 seg-
ments. Local myocardial work is decreased in early contracting sites
that have a low local preload and is increased in late sites where pre-
load is enhanced by preceding stretch.314 Part of the myocardial
work is wasted. At the same time intra-ventricular mechanical dys-
synchrony initiates partially asymmetric cellular remodelling on multi-
ple 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.318,319,320

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 can-
didates 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 ineffi-
ciency was recently described.321 However, CRT has been mainly
reserved for patients with systemic ventricular dysfunction so far.
Conventional pacing-associated dysynchronopathy may be pre-
vented 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 candi-
dates 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, func-
tional improvement, and a reduction in heart failure-associated mor-
bidity 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 myo-
cardial 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 ven-
tricular function. The following observations may be highlighted for
CRT in adult CHD.35,98,293,327–331:

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 inde-
pendent 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 car-
diac 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

<table>
<thead>
<tr>
<th>Recommendations</th>
<th>Consensus statement</th>
<th>References</th>
</tr>
</thead>
<tbody>
<tr>
<td>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.</td>
<td></td>
<td>332–334</td>
</tr>
<tr>
<td>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 (&gt;40%) ventricular pacing. Single site pacing from the systemic ventricular apex/mid-lateral wall may be considered as alternative.</td>
<td></td>
<td>319,98,283–285,328–331</td>
</tr>
<tr>
<td>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.</td>
<td></td>
<td>332–334</td>
</tr>
<tr>
<td>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).</td>
<td></td>
<td>334</td>
</tr>
<tr>
<td>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).</td>
<td></td>
<td>98,283–285,319,331</td>
</tr>
<tr>
<td>CRT can be useful for patients with a systemic ventricular EF &gt;35%, intrinsic narrow QRS complex, NYHA function Class I—ambulatory IV and are undergoing new device placement or replacement with anticipated requirement for significant (&gt;40%) ventricular pacing. Single site pacing from the systemic ventricular apex may be considered as alternative.</td>
<td></td>
<td>332–334,319,336,337,285,319</td>
</tr>
<tr>
<td>CRT can be useful for patients who have 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.</td>
<td></td>
<td>332–334</td>
</tr>
<tr>
<td>CRT can be considered for patients with a systemic RV with significant tricuspid valve regurgitation without a specific EF limit, NYHA function Class II—ambulatory IV, 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.</td>
<td></td>
<td>337</td>
</tr>
<tr>
<td>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.</td>
<td></td>
<td>330,338–340</td>
</tr>
<tr>
<td>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.</td>
<td></td>
<td>333</td>
</tr>
<tr>
<td>CRT is not indicated in patients with a narrow QRS complex (&lt;120 ms) without major electrical activation delay within the failing ventricle.</td>
<td></td>
<td>338</td>
</tr>
<tr>
<td>CRT is not indicated for patients whose co-morbidities and/or frailty limit survival with good functional capacity to less than 1 year.</td>
<td></td>
<td>338</td>
</tr>
</tbody>
</table>

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

Table 16

<table>
<thead>
<tr>
<th>Author (references)</th>
<th>n</th>
<th>Age (FU) (m)</th>
<th>CHD%</th>
<th>Single Vent.</th>
<th>Conv. Pacing</th>
<th>QRS Pre</th>
<th>EF Pre</th>
<th>NYHA III–IV Pre</th>
<th>Main feature</th>
<th>FU (m)</th>
<th>QRS Post</th>
<th>SRV Post</th>
<th>EF Post</th>
<th>CRT resp</th>
<th>Non resp</th>
<th>Impaired EF + functional response</th>
<th>Involved ventricle</th>
</tr>
</thead>
<tbody>
<tr>
<td>Janousek et al.</td>
<td>324</td>
<td>13.78</td>
<td>7.8</td>
<td>100</td>
<td>30.8</td>
<td>0</td>
<td>100</td>
<td>164</td>
<td>Impaired EF</td>
<td>100</td>
<td>100</td>
<td>0</td>
<td>100</td>
<td>11.1</td>
<td>NA</td>
<td>51</td>
<td>Impaired EF + functional response</td>
</tr>
<tr>
<td>Dubin et al.</td>
<td>121</td>
<td>12.5</td>
<td>7.5</td>
<td>100</td>
<td>30.8</td>
<td>0</td>
<td>100</td>
<td>164</td>
<td>Impaired EF</td>
<td>100</td>
<td>100</td>
<td>0</td>
<td>100</td>
<td>11.1</td>
<td>NA</td>
<td>51</td>
<td>Impaired EF + functional response</td>
</tr>
<tr>
<td>Khairy et al.</td>
<td>280</td>
<td>61.3</td>
<td>1.3</td>
<td>100</td>
<td>30.8</td>
<td>0</td>
<td>100</td>
<td>164</td>
<td>Impaired EF</td>
<td>100</td>
<td>100</td>
<td>0</td>
<td>100</td>
<td>11.1</td>
<td>NA</td>
<td>51</td>
<td>Impaired EF + functional response</td>
</tr>
<tr>
<td>Moak et al.</td>
<td>332</td>
<td>15.0</td>
<td>15.0</td>
<td>100</td>
<td>30.8</td>
<td>0</td>
<td>100</td>
<td>164</td>
<td>Impaired EF</td>
<td>100</td>
<td>100</td>
<td>0</td>
<td>100</td>
<td>11.1</td>
<td>NA</td>
<td>51</td>
<td>Impaired EF + functional response</td>
</tr>
<tr>
<td>Cecchin et al.</td>
<td>330</td>
<td>72.4</td>
<td>4.6</td>
<td>100</td>
<td>30.8</td>
<td>0</td>
<td>100</td>
<td>164</td>
<td>Impaired EF</td>
<td>100</td>
<td>100</td>
<td>0</td>
<td>100</td>
<td>11.1</td>
<td>NA</td>
<td>51</td>
<td>Impaired EF + functional response</td>
</tr>
<tr>
<td>Jauvert et al.</td>
<td>336</td>
<td>93.6</td>
<td>6.6</td>
<td>100</td>
<td>30.8</td>
<td>0</td>
<td>100</td>
<td>164</td>
<td>Impaired EF</td>
<td>100</td>
<td>100</td>
<td>0</td>
<td>100</td>
<td>11.1</td>
<td>NA</td>
<td>51</td>
<td>Impaired EF + functional response</td>
</tr>
<tr>
<td>Thambo et al.</td>
<td>236</td>
<td>72.4</td>
<td>15.0</td>
<td>100</td>
<td>30.8</td>
<td>0</td>
<td>100</td>
<td>164</td>
<td>Impaired EF</td>
<td>100</td>
<td>100</td>
<td>0</td>
<td>100</td>
<td>11.1</td>
<td>NA</td>
<td>51</td>
<td>Impaired EF + functional response</td>
</tr>
<tr>
<td>Sakaguchi et al.</td>
<td>332</td>
<td>60.0</td>
<td>15.0</td>
<td>100</td>
<td>30.8</td>
<td>0</td>
<td>100</td>
<td>164</td>
<td>Impaired EF</td>
<td>100</td>
<td>100</td>
<td>0</td>
<td>100</td>
<td>11.1</td>
<td>NA</td>
<td>51</td>
<td>Impaired EF + functional response</td>
</tr>
</tbody>
</table>

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.
aMean value.

need for primary preventive defibrillation capability (CRT-D) should be at least one well-documented case report suggesting acute improvement of right ventricular function and long-term reverse remodelling, respectively.32–33

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).38,283–285,319,331,335

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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References


Arrhythmias in congenital heart disease developed in partnership between the Pediatric and Congenital Electrophysiology Society (PACES) and the Heart Rhythm Society (HRS). Endorsed by the governing bodies of PACES, HRS, the American College of Cardiology (ACC), the American Heart Association (AHA), the European Heart Rhythm Association (EHRA), the Canadian Heart Rhythm Society (CHRS), and the International Society for Adult Congenital Heart Disease (ISACHD). Heart Rhythm 2014;11:1002–65.


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*Note: The text is extracted from a scientific article and includes citations to other works.*


