

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

Table 1 Scientific rationale of recommendations

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

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

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

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

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

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

Relationships with industry and other conflicts of interest

It is EHRA/ESC policy to sponsor position papers and guidelines without commercial support and all members volunteered their

time. Thus, all members of the writing group as well as reviewers have disclosed any potential conflict of interest in detail at the end of this document.

Scope of the consensus document

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

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

Arrhythmias in congenital heart disease: general considerations

Arrhythmia substrates in congenital heart disease

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

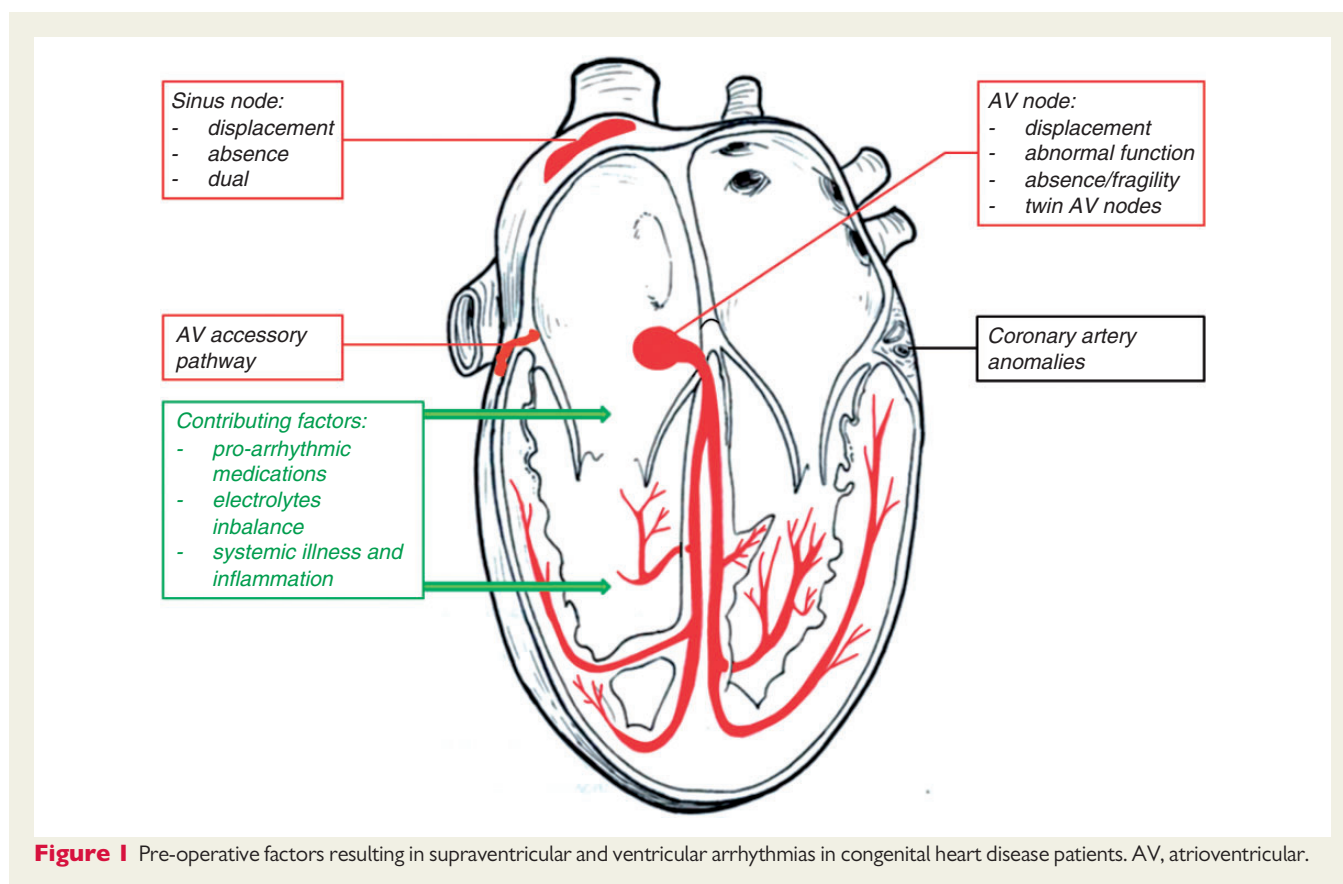
Congenital substrates for arrhythmias in congenital heart defects

Sinus node

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

Atrioventricular block

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



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

Twin atrioventricular nodes

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

Accessory atrioventricular connections

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

Atrioventricular nodal reentrant tachycardia

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

Post-operative substrates for arrhythmias in congenital heart defects

Sinus node dysfunction and atrioventricular block

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

Macroreentrant atrial tachycardias

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

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

Focal atrial tachycardia

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

Accessory pathways after Björk-Fontan modification

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

Ventricular arrhythmias

Stable monomorphic ventricular tachycardia

Stable monomorphic ventricular tachycardia (VT) in patients with CHD is based upon morphological/anatomical variants of the heart

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

Polymorphic ventricular tachycardia

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

Work-up of patients with congenital heart disease and arrhythmias

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

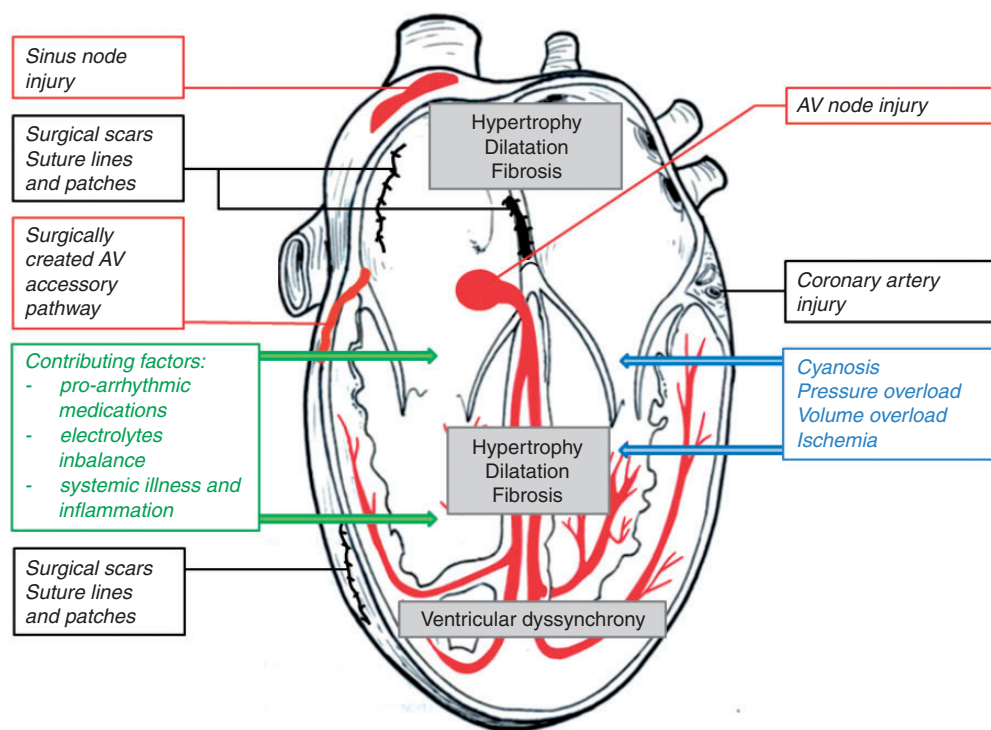


Figure 2 Post-operative factors resulting in supraventricular and ventricular arrhythmias in congenital heart disease patients. AV, atrioventricular.

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

Acute assessment of the congenital heart disease patient presenting with arrhythmia

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

Besides recurrent need for direct current cardioversion, ineffective pharmacological treatment and/or intolerance for drugs (e.g. 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 2 Initial work-up of the CHD patient with arrhythmia

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

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

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

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

Arrhythmia management via implanted devices

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

Imaging requirements for invasive procedures: road mapping

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

Specific arrhythmia types

Supraventricular arrhythmias in patients with congenital heart disease

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

Accessory pathways and atrioventricular reentrant tachycardias

Ebstein's anomaly is the entity most frequently associated with AV reciprocating tachycardia due to the presence of often multiple and

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

Atrioventricular nodal re-entrant tachycardia

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

Atrial tachycardias

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

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

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

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

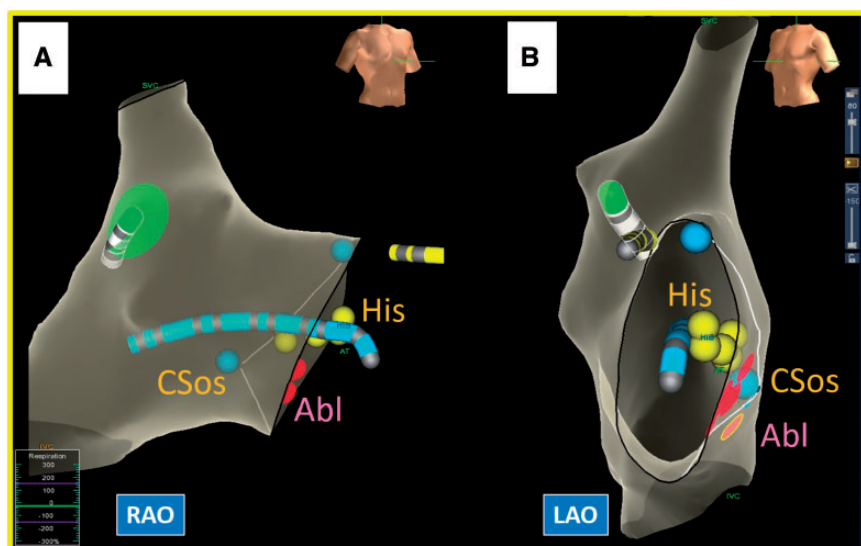


Figure 3 Patient with AV nodal re-entrant tachycardia after surgical repair of complete AV septal defect. Electroanatomical maps (NavX[®]) of the right atrium in the right anterior oblique view (A) and left anterior oblique view (B) display typical caudal displacement of the AV node-His bundle, shown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

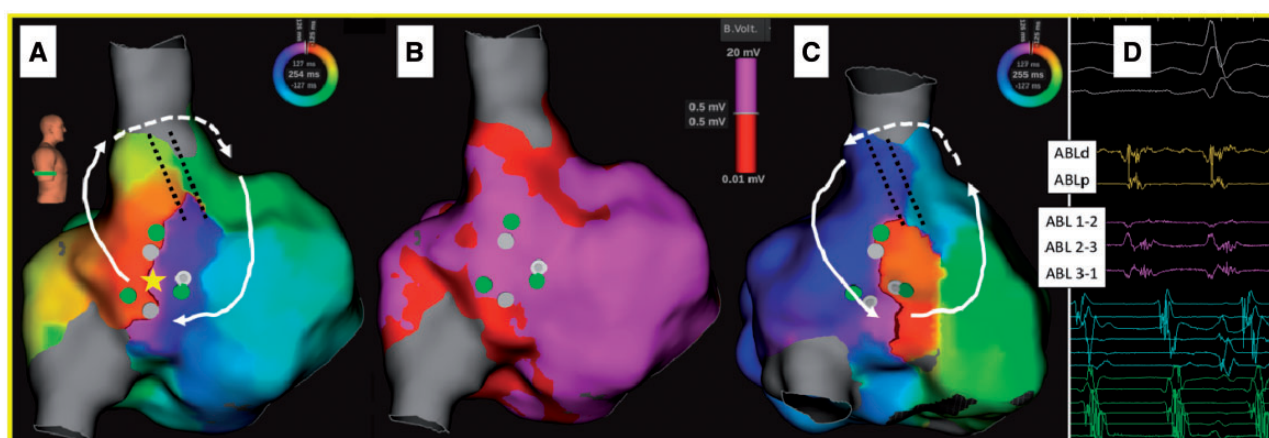


Figure 4 Incisional right atrial macroreentrant tachycardia in a patient after repair of sinus venous type atrial septal defect. Two clinical atrial tachycardias share the same incisional circuit on the right atrial free wall (RAO view): a re-entrant map of one of the reentrant circuits (A) and the corresponding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

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

The Fontan circulation in patients with a univentricular heart is prone to atrial tachyarrhythmias both located at the lateral right atrial wall⁴⁰ and at the CTI. Prevalence of MRAT was higher in the first

2 years after Fontan and increased again gradually in later childhood.⁴⁰ The atriopulmonary connection is often associated with a progressive and massive enlargement of the systemic venous atrium. In a contemporary multicentre series, 19% of these patients developed MRAT, with older series showing even higher rates.⁴¹ The newer Fontan modifications including intracardiac lateral tunnel, extracardiac lateral tunnel, or extracardiac conduit were significantly less arrhythmogenic,

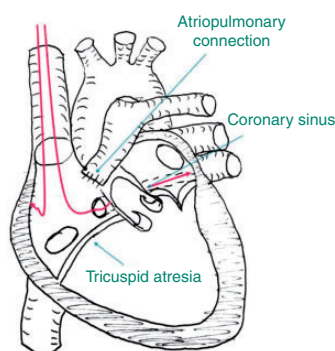


Figure 5 Patient with right ventricular atresia and corrected with the Fontan technique. Diagram of the cardiac anatomy of the patient (A) and the fluoroscopic image showing the transvenous delivery of the pacemaker ventricular lead into the coronary sinus (B).
Photo courtesy of Dra. Inmaculada Sanchet (Ramón y Cajal hospital).

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

Patients after atrial switch procedures (Mustard/Senning) are also highly prone to develop MRAT. Prevalence may reach up to 27% at 20 years post-surgery, often revolving around the tricuspid annulus.^{42,43} The arterial switch procedure was less 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 CHA₂DS₂-C score. Antiarrhythmic therapy with Class IC antiarrhythmic agents and amiodarone is effective in almost 50% of the patients, but systemic side effects and proarrhythmia are of concern and require careful surveillance.^{1,48} Direct current cardioversion with appropriate anticoagulation is safe and effective for patients with CHD, even in the presence of an intracardiac shunt and spontaneous contrast on TOE.⁴⁹ Haemodynamically, rhythm control may be superior to rate control for AF in congenital heart patients, but up to now, there are no data available supporting this hypothesis. Pulmonary vein isolating catheter ablation procedures for drug-refractory AF may be beneficial in selected patients, but experience is limited.^{50–52} The same applies to surgical Maze procedures.⁵³ Prophylactic arrhythmia surgery has been advocated to be incorporated into reparative open heart procedures for CHD patients, but data on efficacy are lacking.⁵⁴ In symptomatic patients with AF refractory to pharmacologic and ablation therapy or failure to adequately control the average heart rate, AV nodal ablation and post-ablation pacing may be considered as third-line therapy.^{30,55}

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

Ventricular tachycardia

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

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

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

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

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

Sudden cardiac death

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

Sudden cardiac death associated with bradycardias and atrial arrhythmias

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

Sudden cardiac death due to ventricular arrhythmias

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

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

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

Bradyarrhythmias in patients with congenital heart disease

Congenital atrioventricular block

Congenital complete heart block (CCHB), first described by Morquio in 1901⁸⁷ and documented on ECG in 1908,⁸⁸ is due to disruption of the electrophysiological continuity between atria and ventricles.^{89–92} Congenital complete heart block may occur in a structurally normal heart (isolated CCHB) or in association with a variety of CHD. The frequent aetiologies of CCHB are listed in Table 3. Symptoms are related to underlying anatomy and ventricular rate and include heart failure prenatally and in infancy, and reduced exercise tolerance, presyncope, or syncope (Stokes–Adams attacks) at any age.^{93–97}

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

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

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

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

Post-surgical atrioventricular block

Post-surgical heart block was first explored in detail by Lillehei in 1963¹⁰¹ and still occurs in 1–3% cases of congenital heart surgeries^{102–105} especially after closure of ventricular septal defects, after TOF repair, surgery along the left ventricular outflow tract, and left-sided valve surgery.³⁰ A recent multicentre study demonstrated highest incidences of AV block and pacemaker placement after the double switch operation (15.6%), tricuspid valve (7.8%) and mitral valve (7.4%) replacement, atrial switch with ventricular septal defect repair (6.4%) and Rastelli operation (4.8%).¹⁰⁶ Compared to surgery, a higher rate (3–20%) of complete heart block (CHB) following transcatheter device closure of perimembranous ventricular septal defect has been reported^{107–110} resulting in abandoning the procedure in children <30 kg or age <6 years. Complete heart block usually occurs immediately after surgery or early in the post-operative period and has been reported only occasionally months or even years thereafter.

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

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

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

Sinus node dysfunction

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

Loss of AV synchrony may distinctly worsen AV valve regurgitation, and a low resting heart rate with poor chronotropic response is associated with reduced cardiac output. Sinus node dysfunction results in atrial remodelling¹³⁸ with increasing risks of atrial tachyarrhythmias, the tachycardia–bradycardia syndrome.¹³⁹ Although there has not been direct evidence of the relationship between SND and sudden death in patients with congenital heart disease,⁷⁶ uncontrolled atrial tachyarrhythmias may increase the risk of heart failure, mortality, and SCD,^{73,140,141} particularly in those with ventricular dysfunction. In addition, thrombo-embolic events including stroke have been reported, especially in patients with tachycardia–bradycardia syndrome. Symptoms of SND vary depending on function of the cardiac conduction system, particular underlying structural heart defect and age. Fatigue, dizziness, exercise intolerance, and syncope have been reported in children and adults.¹⁴² Twelve-lead ECG, ambulatory Holter, event monitors, and exercise stress tests should routinely be performed for assessment and diagnosis of SND. Provocative testing or electrophysiological study is rarely necessary.¹⁴³



Management of arrhythmias in congenital heart disease

Pharmacological management

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

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

Table 4 Recommendations for acute pharmacological therapy




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

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

Acute management




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

Table 5 Recommendations for pharmacological therapy of supraventricular tachycardia

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

Continued

Table 5 Continued

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

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





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

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

Long-term management

Long-term antiarrhythmic therapy is associated with poor long-term arrhythmia freedom and a high rate of side effects in CHD patients. At 2.5 ± 1.4 years follow-up only 45% of 70 patients treated for SVT with varying anti-arrhythmic agents had freedom from recurrence. Success was higher in those with heart failure symptoms at baseline (hazard ratio 1.9; P = 0.023). Amiodarone is an effective agent for complex CHD (with or without ventricular dysfunction) and for moderate CHD with ventricular dysfunction, although patients frequently develop side effects requiring discontinuation or dose reduction.⁴⁸ Amiodarone as a long-term therapeutic regimen is therefore

Table 6 Recommendations for pharmacological therapy of ventricular tachycardia

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

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

not recommended in young CHD patients. Sotalol should not be used related to increased risk for proarrhythmias and mortality.¹⁵⁶ Dofetilide therapy either maintained sinus rhythm or improved arrhythmia control in 49% of patients at a median follow-up of 3 years,¹⁵⁰ but side effects were significant.

Given the poor efficacy of antiarrhythmic agents and prevalence of CTI dependent AF in 75–80% of patients following biventricular repair of CHD, a primary ablative strategy may be¹⁵⁷ (Table 5).

Side effects of antiarrhythmic therapy in congenital heart disease

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

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

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

Anticoagulation

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

Anticoagulation should always be individualized in discussion with the patient and based on the relative risks of stroke and haemorrhagic 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.¹⁶⁰ Stroke prevention is recommended with the same indications as in AF amongst CHD patients with typical AF or AT (Table 5). Low risk patients with AF, defined as CHA₂DS₂-VASc 0 in males or 1 in females, typically do not need antithrombotic therapy.¹⁶¹

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.¹⁶²

Catheter ablation

Catheter ablation of congenital (inherited) tachycardia substrates

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

Technical challenges and limitations

When planning an ablation procedure, alternative access routes to the heart may be needed, such as internal jugular, subclavian, femoral

collateral, or transhepatic access.¹⁶⁴ Expertise in EP ablation is also mandatory if transseptal or transbaffle punctures are needed to access the pulmonary venous atrium.^{165–168} This procedure should be reserved for specialized, dedicated centres with surgical back-up as an alternative to retrograde transaortic access¹⁶⁵ (Table 7). Epicardial ablation approach in post-operative CHD may be required in selected patients, but data are sparse.

Anatomical challenges

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

Specific arrhythmias and malformations

In patients with Ebstein's anomaly, episodes of paroxysmal SVT are frequent, and are typically related to the presence of accessory pathways.^{10,185–193} Catheter ablation of accessory AV connections is the preferred therapeutic option with lower acute success rates¹⁸⁹ and higher recurrence rates compared with patients with normal hearts.^{190,191} (Table 7). With further development of catheter and sheath technology and by the advent of 3D mapping systems¹⁰ success rates and safety have been markedly improved.



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

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

Catheter ablation in post-operative settings


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

Table 7 Recommendations for supraventricular tachyarrhythmia catheter ablation

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

Continued

Table 7 Continued

Recommendations	Consensus statement	References
Catheter ablation is not recommended for atrial tachyarrhythmias that can be controlled medically in the early post-surgical period (<3 months).		Expert consensus
Catheter ablation is not recommended for asymptomatic non-sustained runs of atrial tachycardia.		Expert consensus

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

^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%.⁹ However, with increasing experience and implementation of electro-anatomical mapping systems results could be improved.¹⁷⁵ Overall, major complication rates may reach up to 4.2% and minor complications rates of 5.5% have been reported. These figures are comparable with complication rates in normal cardiac anatomy.¹⁹¹

Catheter ablation of post-operative substrates



Arrhythmogenesis

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

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


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

Table 8 Recommendations for VT ablation

Recommendations	Consensus statement	References
Catheter ablation is indicated as additional therapy in ICD patients with CHD who present with incessant VT or electrical storm.		66,67,201
Catheter ablation is indicated as additional therapy in ICD patients with CHD who have recurrent symptomatic monomorphic VT despite ATP or appropriate ICD shocks not manageable by device reprogramming or when medical therapy is not effective or has intolerable side effects.		66–68,70
Pre-operative electroanatomical mapping to identify SCAI as substrates for spontaneous or induced monomorphic VT is recommended in patients with TOF who undergo re-operation for haemodynamic reasons.		69
Surgical intraoperative cryo-ablation is recommended in ToF patients who undergo re-surgery and who have spontaneous or inducible VTs unless managed by preoperative ablation.		69,202,203
The use of 3D mapping systems to obtain a 3D reconstruction of all VT related anatomic isthmuses is recommended.		66,67,69,70
Catheter ablation should be considered for symptomatic monomorphic sustained VT in grown up-congenital heart disease patients as an alternative to drug therapy.		66,67,69
Catheter ablation should be considered in grown-up congenital heart disease patients with frequent highly symptomatic PVCs, or PVCs associated with deteriorating ventricular function.		20
Catheter ablation or concomitant surgical cryoablation can be considered for symptomatic monomorphic sustained VT in grown-up congenital heart disease patients with a preserved biventricular function as an alternative to ICD therapy, provided that the procedure is performed in highly experienced CHD centres and that established ablation endpoints have been reached (i.e. non-inducibility, conduction block across ablation lines).		67,69

Continued

Table 8 Continued

Recommendations	Consensus statement	References
Surgical intraoperative cryoablation of SCAI may be considered in rTOF patients with SCAI but without spontaneous or inducible VT who need re-operation for residual haemodynamic lesions.		69
The use of irrigated tip catheter is recommended for linear lesions and hypertrophied myocardium.		66,67,202
Catheter ablation is not recommended for asymptomatic, infrequent PVC and non-sustained VT		

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

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 success³⁰ (Tables 7–9).

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

Fluoroscopy reduction needs to be emphasized. Nowadays electrophysiological procedures should be performed according to the As Low As Reasonably Achievable (ALARA) principle which includes the use of 3D mapping technology allowing to identify the exact location of the mapping catheter and important anatomical structures such as valve annuli, His bundle, or phrenic nerve, providing better long-term outcomes.^{200,204} For mapping and ablation of AT and VT in CHD, 3D reconstruction of all potential anatomical isthmuses facilitated by the use of 3D mapping systems is highly recommended.^{178,200,204–207} The use of irrigated-tip catheters has been associated with an increase in ablation success rates, to overcome limitations with lesion formation in thickened, fibrotic atrial, and ventricular myocardium in CHD patients. Contact force may be helpful as well as intracardiac 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 needs 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

Table 9 Requirements for complex CHD arrhythmia management

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

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

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

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

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

Based on this experience, in patients with non-inducible MRAT or unmappable circuits, a substrate mapping approach including voltage and activation mapping can be performed during sinus rhythm or fixed atrial pacing.^{214,215} Finally, empirical CTI ablation²¹⁶ as well as a corridor between right lateral atriotomy scar and the IVC has been advocated in non-inducible CHD patients.⁴⁰

Acute ablation outcomes range between 80% in scar-related MRAT and reach 90–95% for CTI-dependent MRAT or AF.^{173,217,218} Electroanatomical mapping, locating the mid-diastolic isthmus, and adjusting the window of interest are important for ablation. Long-term success, however, may be impaired by progressive myocardial

fibrosis, especially in complex post-operative CHD. Acute success in MRAT ablation is defined as achievement of bi-directional conduction block across any critically conducting myocardial isthmus or CTI.¹⁷³ In the past, termination of tachycardia with ablation and subsequent non-inducibility of the tachycardia alone were often used as endpoint in ablation procedures. Lack of evidence of complete conduction block, however, was identified as a risk factor for tachycardia recurrence.²⁰⁹

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

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

Catheter ablation of ventricular tachycardia

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

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

Considering the high acute success rates and low recurrence rates VT ablation may offer a reasonable alternative to ICD therapy in carefully selected patients with preserved cardiac function.^{66,67,201} Procedural complications are rare and either related to vascular access or location of anatomical isthmuses including AV block.^{66–68,70,201,202,224,225} Reasons for ablation failure include

hypertrophied myocardium, proximity of the conduction system, and importantly, the protection of portions of anatomical isthmuses by prosthetic material.^{66,201} In particular, a pulmonary homograft may cover parts of the infundibular septum in TOF patients preventing isthmus transection.

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

Surgical treatment

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

Preventative surgery

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

Intraoperative treatment of pre-existing tachyarrhythmia

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

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

Indications for surgery of arrhythmias in congenital heart disease

Surgery for arrhythmias 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 antiarrhythmic surgery may be implemented, especially for treatment of AF, incisional AT, AF, or VT.^{246–250}

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.^{251–253} In contrast to patients with structurally normal hearts, epicardial leads (sternotomy or thoracotomy) are frequently required as are hybrid (combined epicardial and endocardial) and novel systems (intra-operative atrial puncture, baffle puncture, femoral or hepatic vein access, and S-ICD coils). Achieving the desired outcome by any route can be challenging.

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

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

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

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

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

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

Senning or Mustard repair for transposition of the great arteries

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

Congenitally corrected transposition of the great arteries

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

Single ventricle physiology: Fontan circulation

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










One and a half ventricle circulation

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

Pacemakers in patients with congenital heart disease

Patients with CHD and post-operative SND or high degree or complete AV block—even when the underlying defects are ‘structurally repaired’—are considered to be at a higher risk of SCD. Therefore,

Table 10 Recommendations for pacemaker implantation according to the underlying disease

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

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

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 11 Recommendations for pacemaker implantation according to the underlying disease

AV block Recommendations	Consensus statement	References
Symptomatic bradycardia with any degree of AV block or with ventricular arrhythmias presumed to be due to AV block		6
Asymptomatic patients with high degree or complete AV block and with one of the following: <ol style="list-style-type: none"> Ventricular dysfunction Low cardiac output Wide QRS escape rhythm Complex ventricular ectopy Prolonged QT interval 		6,283
Post-operative or post-interventional 3° AV block if considered irreversible and lasting >7 days		6,284
Symptomatic 2° type 1 AV block, 2° AV block at intra- or infra-His levels according to EPS		143
Syncope in patients with residual bifascicular block and prior complete AV block following heart surgery after excluding other causes		120
Transient complete post-operative AV block with restitution of sinus rhythm with residual bifascicular block		6,12
Transient post-operative complete AV block which returns to normal AV conduction in asymptomatic patients		6,105,112
Post-operative asymptomatic bifascicular block ± 1° AV block without prior transient complete AV Block		6

AV, atrioventricular.

primarily with clinical symptoms and cardiac function rather than pre-defined heart rate limits. In the last years, several guidelines for device implantation^{6,143,277,278} have been published with one study⁸⁵ mainly focusing on CHD patients.

Choosing the optimal pacing site

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

Lead extraction

In CHD patients, the need for lead extraction has grown. However, complex vascular and cardiac anatomy may be challenging. Results of lead extraction in CHD patients are promising.^{290–292} Guidelines for lead extraction²⁹³ have been published recently and are applicable for CHD patients after taking into account the venous anatomy and anatomical connections (Table 12).

Implantable cardioverter-defibrillator in patients with congenital heart disease

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

Implantable cardioverter-defibrillator cohorts

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

Recommendations for implantable cardioverter-defibrillator therapy















Secondary prevention

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

Primary prevention

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

Table 12 Recommendations for lead extraction

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

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

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

There is the general agreement that ICDs are also indicated in adults with CHD who meet the established ICD criteria, namely a left ventricular EF < 35%, biventricular physiology, symptomatic heart failure despite optimal medical treatment, and NYHA functional Class II or III.²⁰ Implantable cardioverter-defibrillators should be considered in CHD patients with syncope of unknown

origin in the presence of either advanced ventricular dysfunction or inducible sustained VT or VF on programmed ventricular stimulation.⁵⁷









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

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

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

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

Table 14 Recommendations for ICD therapy in adults with CHD

Recommendations	Consensus statement	References
ICD is recommended for patients with CHD who are survivors of an aborted cardiac arrest due to VF or haemodynamically unstable VT after evaluation to define the cause of the event and exclusion of any reversible causes.		57,58,61,295,305,306
ICD is recommended for patients with CHD with symptomatic sustained VT who have undergone haemodynamic and electrophysiological evaluation.		57,58,61,143,295,296
ICD is recommended in adults with CHD and a systemic LVEF \leq 35%, biventricular physiology and NYHA functional Class II or III.		20,307,308
ICD implantation should be considered in patients with CHD and syncope of unknown origin in the presence of either advanced ventricular dysfunction or inducible sustained VT or VF on VPS.		56,57,58,59,300
ICD implantation should be considered in selected patients with TOF and multiple risk factors for SCD, including LV dysfunction, non-sustained VT, QRS duration \geq 180 ms, or inducible sustained VT on VPS.		33,56,57,308
ICD therapy may be considered in patients with advanced single or systemic RV dysfunction in the presence of risk factors such as non-sustained VT, NYHA functional Class II or III, QRS duration \geq 140 ms or severe systemic AV valve regurgitation.		33,59,61,299
ICD therapy may be considered for non-hospitalized adults with CHD awaiting heart transplantation.		308,309
ICD is contraindicated in patients with CHD with life expectancy with an acceptable functional status \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		

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

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

Mortality

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

age of ICD patients with CHD as compared to ICD patients with ischaemic and non-ischaemic cardiomyopathies.⁶³

Complications, inappropriate shocks, and implantable cardioverter-defibrillator programming

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

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

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

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

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

Cardiac resynchronization in patients with congenital heart disease

Dyssynchronous heart failure and cardiac resynchronization therapy

Pathophysiology

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

Epidemiology

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

The most frequent conduction disturbance in CHD is right bundle branch block in the setting of a sub-pulmonary right ventricle. Right

ventricular electromechanical dyssynchrony and mechanical inefficiency was recently described.³²¹ However, CRT has been mainly reserved for patients with systemic ventricular dysfunction so far. Conventional pacing-associated dyssynchronopathy may be prevented by septal or His pacing and may substantially decrease the number of CRT candidates among CHD patients.

Imaging

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

Clinical studies

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

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

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

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

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

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

Table 16 Summary of clinical studies on CRT in CHD

Author (references)	n	Age	Desing FU (m)	CHD %	Syst. RV %	Single V %	Conv. pacing %	QRS Pre ms	EF Pre	NYHA III-IV Pre %	Epic CRT %	Non resp %	QRS post ms	EF Post	Main feature
Janousek et al. ³³¹	8	15.0 ^a	SC, P 17.4 ^b	100	100	0	75	161 ^a	18	12.5	37.5	NA	116 ^a	30	First study on CRT in systemic right ventricle
Dubin et al. ³³³	103	12.8 ^b	MC, R 4 ^b	70.9	16.5	6.8	44.7	166 ^a	26	37.9	46.6	10.7	126 ^a	40	First large study on CRT in congenital heart disease
Khairy et al. ³²⁴	13	7.8 ^a	SC, R 16.5 ^a	100	30.8	0	100	NA	31	NA	100	11.1	NA	51	Impaired EF + conventional pacing in all
Moak et al. ²⁸⁰	6	11.3 ^a	SC, R 10 ^b	33.3	0	0	100	204 ^a	34	NA	66.7	0	138 ^a	60	Super-response after upgrade from RVP to CRT
Cecchin et al. ³³²	60	15.0 ^b	SC, R 8 ^b	76.7	15	21.7	68.3	149 ^a	36	31.7	63.3	10	120 ^a	42	Largest reported single ventricular patient group
Jauvert et al. ³³⁰	7	24.6 ^a	SC, P 19.4 ^a	100	100	0	71.4	160 ^a	NA	100	28.6	NA	120 ^a	NA	CRT in systemic right ventricle
Janousek et al. ³³⁴	109	16.9 ^a	MC, R 7.5 ^b	79.8	33	3.7	77.1	160 ^b	30	45.9	33	13.7	130 ^b	41	Largest study with different substrates
Thambo et al. ³³⁶	9	36.6 ^a	SC, P 6 ^b	100	0	0	0	164 ^a	50	NA	33	NA	NA	56	Postoperative TOF
Sakaguchi et al. ³³⁹	20	22 ^a	SC, R ≥6	100	35	30	60	183 ^a	34	25	NA	NA	132 ^a	NA	Effect of CRT according to systemic ventricle type

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

^aMean value.

^bMedian value.

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

Technical aspects

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

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

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

Indications

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

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

Areas for future research

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

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

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

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

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

Conclusions

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

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References

- Katritsis DG, Boriani G, Cosio FG, Hindricks G, Jais P, Josephson ME. European Heart Rhythm Association (EHRA) consensus document on the management of supraventricular arrhythmias, endorsed by Heart Rhythm Society (HRS), Asia-Pacific Heart Rhythm Society (APHRS), and Sociedad Latinoamericana de Estimulación Cardíaca y Electrofisiología (SOLAECE). *Europace* 2017;**19**: 465–511.
- Anderson RH, Ho SY. The disposition of the conduction tissues in congenitally malformed hearts with reference to their embryological development. *J Perinat Med* 1991;**19** Suppl 1:201–6.
- Ho SY, Fagg N, Anderson RH, Cook A, Allan L. Disposition of the atrioventricular conduction tissues in the heart with isomerism of the atrial appendages: its relation to congenital complete heart block. *J Am Coll Cardiol* 1992;**20**:904–10.

- Brugada J, Blom N, Sarquella-Brugada G, Blomstrom-Lundqvist C, Deanfield J, Janousek J; European Heart Rhythm Association; Association for European Paediatric and Congenital Cardiology et al. Pharmacological and non-pharmacological therapy for arrhythmias in the pediatric population: EHRA and AEPCC-Arrhythmia Working Group joint consensus statement. *Europace* 2013;**15**:1337–82.
- Lenox CC, Hashida Y, Anderson RH, Hubbard JD. Conduction tissue anomalies in absence of the right superior caval vein. *Int J Cardiol* 1985;**8**:251–60.
- Baruteau AE, Pass RH, Thambo JB, Behaghel A, Le Pennec S, Perdreau E et al. Congenital and childhood atrioventricular blocks: pathophysiology and contemporary management. *Eur J Pediatr* 2016;**175**:1235–48.
- Warnes CA. Transposition of the great arteries. *Circulation* 2006;**114**:2699–709.
- Epstein MR, Saul JP, Weindling SN, Triedman JK, Walsh EP. Atrioventricular reciprocating tachycardia involving twin atrioventricular nodes in patients with complex congenital heart disease. *J Cardiovasc Electrophysiol* 2001;**12**:671–9.
- Upadhyay S, Marie Valente A, Triedman JK, Walsh EP. Catheter ablation for atrioventricular nodal reentrant tachycardia in patients with congenital heart disease. *Heart Rhythm* 2016;**13**:1228–37.
- Walsh EP. Interventional electrophysiology in patients with congenital heart disease. *Circulation* 2007;**115**:3224–34.
- Lesh MD, Kalman JM, Saxon LA, Dorostkar PC. Electrophysiology of “incisional” reentrant atrial tachycardia complicating surgery for congenital heart disease. *Pacing Clin Electrophysiol* 1997;**20**:2107–11.
- Correa R, Sherwin ED, Kovach J, Mah DY, Alexander ME, Cecchin F et al. Mechanism and ablation of arrhythmia following total cavopulmonary connection. *Circ Arrhythm Electrophysiol* 2015;**8**:318–25.
- de Groot NM, Zeppenfeld K, Wijffels MC, Chan WK, Blom NA, Van der Wall EE et al. Ablation of focal atrial arrhythmia in patients with congenital heart defects after surgery: role of circumscribed areas with heterogeneous conduction. *Heart Rhythm* 2006;**3**:526–35.
- Akca F, Bauernfeind T, De Groot NM, Shalgunov T, Schwagten B, Szili-Torok T. The presence of extensive atrial scars hinders the differential diagnosis of focal or macroreentrant atrial tachycardias in patients with complex congenital heart disease. *Europace* 2014;**16**:893–8.
- Hager A, Zrenner B, Brodherr-Heberlein S, Steinbauer-Rosenthal I, Schreieck J, Hess J. Congenital and surgically acquired Wolff-Parkinson-White syndrome in patients with tricuspid atresia. *J Thorac Cardiovasc Surg* 2005;**130**:48–53.
- Apitz C, Webb GD, Redington AN. Tetralogy of Fallot. *Lancet* 2009;**374**: 1462–71.
- Khairy P, Stevenson WG. Catheter ablation in tetralogy of Fallot. *Heart Rhythm* 2009;**6**:1069–74.
- Khairy P. Ventricular arrhythmias and sudden cardiac death in adults with congenital heart disease. *Heart* 2016;**102**:1703–9.
- Khairy P, Aboulhoss J, Gurvitz MZ, Opatowsky AR, Mongeon FP, Kay J; Alliance for Adult Research in Congenital C et al. Arrhythmia burden in adults with surgically repaired tetralogy of Fallot: a multi-institutional study. *Circulation* 2010;**122**:868–75.
- Priori SG, Blomström-Lundqvist C, Mazzanti A, Blom N, Borggrefe M, Camm J et al. 2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death. *Europace* 2015;**17**: 1601–87.
- Bouchardy J, Therrien J, Pilote L, Ionescu-Ittu R, Martucci G, Bottega N et al. Atrial arrhythmias in adults with congenital heart disease. *Circulation* 2009;**120**: 1679–86.
- Wasmer K, Eckardt L. Management of supraventricular arrhythmias in adults with congenital heart disease. *Heart* 2016;**102**:1614–9.
- Gulotta GA, Lamotta EP. Ebstein's anomaly associated with the Wolff-Parkinson-White syndrome. *Heart Cent Bull (Roslyn)* 1959;**16**:16–25.
- Danielson GK, Driscoll DJ, Mair DD, Warnes CA, Oliver WC. Operative treatment of Ebstein's anomaly. *J Thorac Cardiovasc Surg* 1992;**104**:1195–202.
- Frescura C, Angelini A, D'Aliento L, Thiene G. Morphological aspects of Ebstein's anomaly in adults. *Thorac Cardiovasc Surg* 2000;**48**:203–8.
- McCanta AC, Kay JD, Collins KK. Cryoablation of the slow atrioventricular nodal pathway via a transbaffle approach in a patient with the Mustard procedure for d-transposition of the great arteries. *Congenit Heart Dis* 2011;**6**:479–83.
- Rausch CM, Runciman M, Collins KK. Cryothermal catheter ablation of atrioventricular nodal reentrant tachycardia in a pediatric patient after atrioventricular canal repair. *Congenit Heart Dis* 2010;**5**:66–9.
- Billakanty S, Crawford T, Good E, Oral H. Radiofrequency catheter ablation of AV nodal reentrant tachycardia in situs inversus totalis. *Pacing Clin Electrophysiol* 2009;**32**:403–5.
- Hebe J, Hansen P, Ouyang F, Volkmer M, Kuck K-H. Radiofrequency catheter ablation of tachycardia in patients with congenital heart disease. *Pediatr Cardiol* 2000;**21**:557–75.
- Khairy P, Van Hare GF, Balaji S, Berul CI, Cecchin F, Cohen MI et al. PACES/HRS Expert Consensus Statement on the Recognition and Management of

- Arrhythmias in Adult 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**:e102–65.
31. Gatzoulis MA, Balaji S, Webber SA, Siu SC, Hokanson JS, Poile C et al. Risk factors for arrhythmia and sudden cardiac death late after repair of tetralogy of Fallot: a multicentre study. *Lancet* 2000;**356**:975–81.
 32. Baumgartner H, Bonhoeffer P, De Groot NMS, de Haan F, Deanfield JE, Galie N et al. ESC Guidelines for the management of grown-up congenital heart disease (new version 2010). *Eur Heart J* 2010;**31**:2915–57.
 33. Kammeraad JAE, van Deurzen CHM, Sreeram N, Bink-Boelkens MTE, Ottenkamp J, Helbing WA et al. Predictors of sudden cardiac death after Mustard or Senning repair for transposition of the great arteries. *J Am Coll Cardiol* 2004;**44**:1095–102.
 34. Franco E, Sánchez I, Vázquez Martínez J, Moreno J. Ventricular fibrillation caused by 1:1 atrial flutter. *J Cardiovasc Electrophysiol* 2016;**27**:1488–9.
 35. Berger F, Vogel M, Kramer A, Alexi-Meskishvili V, Weng Y, Lange PE et al. Incidence of atrial flutter/fibrillation in adults with atrial septal defect before and after surgery. *Ann Thorac Surg* 1999;**68**:75–8.
 36. Morton JB. Effect of chronic right atrial stretch on atrial electrical remodeling in patients with an atrial septal defect. *Circulation* 2003;**107**:1775–82.
 37. Vecht JA, Saso S, Rao C, Dimopoulos K, Grapsa J, Terracciano CM et al. Atrial septal defect closure is associated with a reduced prevalence of atrial tachyarrhythmia in the short to medium term: a systematic review and meta-analysis. *Heart* 2010;**96**:1789–97.
 38. Duong P, Ferguson LP, Lord S, Murray S, Shepherd E, Bourke JP et al. Atrial arrhythmia after transcatheter closure of secundum atrial septal defects in patients ≥ 40 years of age. *Europace* 2017;**19**:1322–6.
 39. Roos-Hesselink JW. Excellent survival and low incidence of arrhythmias, stroke and heart failure long-term after surgical ASD closure at young age. A prospective follow-up study of 21–33 years. *Eur Heart J* 2003;**24**:190–7.
 40. Stephenson EA, Lu M, Berul CI, Etheridge SP, Idriss SF, Margossian R et al. Arrhythmias in a contemporary fontan cohort: prevalence and clinical associations in a multicenter cross-sectional study. *J Am Coll Cardiol* 2010;**56**:890–6.
 41. d'Udekem Y, Iyengar AJ, Cochrane AD, Grigg LE, Ramsay JM, Wheaton GR et al. The Fontan procedure: contemporary techniques have improved long-term outcomes. *Circulation* 2007;**116**(11 Suppl):1157–64.
 42. Gelatt M, Hamilton RM, McCrindle BW, Connelly M, Davis A, Harris L et al. Arrhythmia and mortality after the Mustard procedure: a 30-year single-center experience. *J Am Coll Cardiol* 1997;**29**:194–201.
 43. Kanter RJ, Papagiannis J, Carboni MP, Ungerleider RM, Sanders WE, Wharton JM et al. Radiofrequency catheter ablation of supraventricular tachycardia substrates after mustard and senning operations for d-transposition of the great arteries. *J Am Coll Cardiol* 2000;**35**:428–41.
 44. Tobler D, Williams WG, Jegatheeswaran A, Van Arsdell GS, McCrindle BW, Greutmann M et al. Cardiac outcomes in young adult survivors of the arterial switch operation for transposition of the great arteries. *J Am Coll Cardiol* 2010;**56**:58–64.
 45. Teuwen CP, Ramdjan TTTK, Götte M, Brundel BJM, Evertz R, Vriend JWJ et al. Time course of atrial fibrillation in patients with congenital heart defects. *Circ Arrhythm Electrophysiol* 2015;**8**:1065–72.
 46. Kirchhof P, Benussi S, Kotecha D, Ahlsson A, Atar D, Casadei B et al. 2016 ESC Guidelines for the management of atrial fibrillation developed in collaboration with EACTS. *Europace* 2016;**18**:1609–78.
 47. Sherwin ED, Triedman JK, Walsh EP. Update on interventional electrophysiology in congenital heart disease: evolving solutions for complex hearts. *Circ Arrhythm Electrophysiol* 2013;**6**:1032–40.
 48. Koyak Z, Kroon B, de Groot JR, Wagenaar LJ, van Dijk AP, Mulder BA et al. Efficacy of antiarrhythmic drugs in adults with congenital heart disease and supraventricular tachycardias. *Am J Cardiol* 2013;**112**:1461–7.
 49. Ammash NM, Phillips SD, Hodge DO, Connolly HM, Grogan MA, Friedman PA et al. Outcome of direct current cardioversion for atrial arrhythmias in adults with congenital heart disease. *Int J Cardiol* 2012;**154**:270–4.
 50. Philip F, Muhammad KI, Agarwal S, Natale A, Krasuski RA. Pulmonary vein isolation for the treatment of drug-refractory atrial fibrillation in adults with congenital heart disease. *Congenit Heart Dis* 2012;**7**:392–9.
 51. Lakkireddy D, Rangisetty U, Prasad S, Verma A, Biria M, Berenbom L et al. Intracardiac echo-guided radiofrequency catheter ablation of atrial fibrillation in patients with atrial septal defect or patent foramen ovale repair: a feasibility, safety, and efficacy study. *J Cardiovasc Electrophysiol* 2008;**19**:1137–42.
 52. Santangeli P, Di Biase L, Burkhardt JD, Horton R, Sanchez J, Bailey S et al. Transseptal access and atrial fibrillation ablation guided by intracardiac echocardiography in patients with atrial septal closure devices. *Heart Rhythm* 2011;**8**:1669–75.
 53. Backer CL, Tsao S, Deal B, Mavroudis C. Maze procedure in single ventricle patients. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2008;**11**:44–8.
 54. Mavroudis C, Stulak JM, Ad N, Siegel A, Giamberti A, Harris L et al. Prophylactic atrial arrhythmia surgical procedures with congenital heart operations: review and recommendations. *Ann Thorac Surg* 2015;**99**:352–9.
 55. Friedman RA, Will JC, Fenrich AL, Kertesz NJ. Atrioventricular junction ablation and pacemaker therapy in patients with drug-resistant atrial tachyarrhythmias after the Fontan operation. *J Cardiovasc Electrophysiol* 2005;**16**:24–9.
 56. Khairy P, Landzberg MJ, Gatzoulis MA, Lucron H, Lambert J, Marcon F et al. Value of programmed ventricular stimulation after tetralogy of Fallot repair: a multicenter study. *Circulation* 2004;**109**:1994–2000.
 57. Khairy P, Harris L, Landzberg MJ, Viswanathan S, Barlow A, Gatzoulis MA et al. Implantable cardioverter-defibrillators in tetralogy of Fallot. *Circulation* 2008;**117**:363–70.
 58. Koyak Z, de Groot JR, Van Gelder IC, Bouma BJ, van Dessel PFHM, Budts W et al. Implantable cardioverter defibrillator therapy in adults with congenital heart disease: who is at risk of shocks? *Circ Arrhythm Electrophysiol* 2012;**5**:101–10.
 59. Koyak Z, Harris L, de Groot JR, Silversides CK, Oechslin EN, Bouma BJ et al. Sudden cardiac death in adult congenital heart disease. *Circulation* 2012;**126**:1944–54.
 60. Teuwen CP, Ramdjan TTTK, Götte M, Brundel BJM, Evertz R, Vriend JWJ et al. Non-sustained ventricular tachycardia in patients with congenital heart disease: an important sign? *Int J Cardiol* 2016;**206**:158–63.
 61. Khairy P, Harris L, Landzberg MJ, Fernandes SM, Barlow A, Mercier LA et al. Sudden death and defibrillators in transposition of the great arteries with intra-atrial baffles: a multicenter study. *Circ Arrhythm Electrophysiol* 2008;**1**:250–7.
 62. Santharam S, Hudsmith L, Thorne S, Clift P, Marshall H, De Bono J. Long-term follow-up of implantable cardioverter-defibrillators in adult congenital heart disease patients: indications and outcomes. *Europace* 2017;**19**:407–13.
 63. Vehmeijer JT, Brouwer TF, Limpens J, Knops RE, Bouma BJ, Mulder BJM et al. Implantable cardioverter-defibrillators in adults with congenital heart disease: a systematic review and meta-analysis. *Eur Heart J* 2016;**37**:1439–48.
 64. Silka MJ, Hardy BG, Menashe VD, Morris CD. A population-based prospective evaluation of risk of sudden cardiac death after operation for common congenital heart defects. *J Am Coll Cardiol* 1998;**32**:245–51.
 65. Oechslin EN, Harrison DA, Connelly MS, Webb GD, Siu SC. Mode of death in adults with congenital heart disease. *Am J Cardiol* 2000;**86**:1111–6.
 66. Zeppenfeld K, Schalij MJ, Bartelings MM, Tedrow UB, Koplan BA, Soejima K et al. Catheter ablation of ventricular tachycardia after repair of congenital heart disease: electroanatomic identification of the critical right ventricular isthmus. *Circulation* 2007;**116**:2241–52.
 67. Kapel GF, Reichlin T, Wijnmaalen AP, Piers SR, Holman ER, Tedrow UB et al. Re-entry using anatomically determined isthmuses: a curable ventricular tachycardia in repaired congenital heart disease. *Circ Arrhythm Electrophysiol* 2015;**8**:102–9.
 68. van Zyl M, Kapa S, Padmanabhan D, Chen FC, Mulpuru SK, Packer DL et al. Mechanism and outcomes of catheter ablation for ventricular tachycardia in adults with repaired congenital heart disease. *Heart Rhythm* 2016;**13**:1449–54.
 69. Moore JP, Seki A, Shannon KM, Mandapati R, Tung R, Fishbein MC. Characterization of anatomic ventricular tachycardia isthmus pathology after surgical repair of tetralogy of Fallot. *Circ Arrhythm Electrophysiol* 2013;**6**:905–11.
 70. Kriebel T, Saul JP, Schneider H, Sigler M, Paul T. Noncontact mapping and radiofrequency catheter ablation of fast and hemodynamically unstable ventricular tachycardia after surgical repair of tetralogy of Fallot. *J Am Coll Cardiol* 2007;**50**:2162–8.
 71. Subirana MT, Juan-Babot JO, Puig T, Lucena J, Rico A, Salguero M et al. Specific characteristics of sudden death in a mediterranean Spanish population. *Am J Cardiol* 2011;**107**:622–7.
 72. Alonso A, Jensen PN, Lopez FL, Chen LY, Psaty BM, Folsom AR et al. Association of sick sinus syndrome with incident cardiovascular disease and mortality: the Atherosclerosis Risk in Communities study and Cardiovascular Health Study. *PLoS One* 2014;**9**:e109662.
 73. Fishberger SB, Wernovsky G, Gentles TL, Gauvreau K, Burnett J, Mayer JE, Jr et al. Factors that influence the development of atrial flutter after the Fontan operation. *J Thorac Cardiovasc Surg* 1997;**113**:80–6.
 74. Nieminen HP, Jokinen EV, Sairanen HI. Causes of late deaths after pediatric cardiac surgery: a population-based study. *J Am Coll Cardiol* 2007;**50**:1263–71.
 75. Pillutla P, Shetty KD, Foster E. Mortality associated with adult congenital heart disease: trends in the US population from 1979 to 2005. *Am Heart J* 2009;**158**:874–9.
 76. Alboni P, Menozzi C, Brignole M, Paparella N, Gaggioli G, Lolli G et al. Effects of permanent pacemaker and oral theophylline in sick sinus syndrome. The THEOPACE study: a randomized controlled trial. *Circulation* 1997;**96**:260–6.

77. Shaw DB, Holman RR, Gowers JL. Survival in sinoatrial disorder (sick-sinus syndrome). *Br Med J* 1980;**280**:139–41.
78. Janousek J, Paul T, Luhmer I, Wilken M, Hrudá J, Kallfelz HC. Atrial baffle procedures for complete transposition of the great arteries: natural course of sinus node dysfunction and risk factors for dysrhythmias and sudden death. *Z Kardiol* 1994;**83**:933–8.
79. Hokanson JS, Moller JH. Significance of early transient complete heart block as a predictor of sudden death late after operative correction of tetralogy of Fallot. *Am J Cardiol* 2001;**87**:1271–7.
80. Verheugt CL1, Uiterwaal CS, van der Velde ET, Meijboom FJ, Pieper PG, van Dijk AP et al. Mortality in adult congenital heart disease. *Eur Heart J* 2010;**31**:1220–9.
81. Khairy P, Fernandes SM, Mayer JE Jr, Friedman JK, Walsh EP, Lock JE et al. Long-term survival, modes of death, and predictors of mortality in patients with Fontan surgery. *Circulation* 2008;**117**:85–92.
82. Koyak Z, de Groot JR, Bouma BJ, Zwinderman AH, Silversides CK, Oechslin EN et al. Sudden cardiac death in adult congenital heart disease: can the unpredictable be foreseen? *Europace* 2017;**19**:401–6.
83. Schwerzmann M, Salehian O, Harris L, Siu SC, Williams WG, Webb GD et al. Ventricular arrhythmias and sudden death in adults after a mustard operation for transposition of the great arteries. *Eur Heart J* 2009;**30**:1873–9.
84. Czosek RJ, Bonney WJ, Cassidy A, Mah DY, Tanel RE, Imundo JR et al. Impact of cardiac devices on the quality of life in pediatric patients. *Circ Arrhythm Electrophysiol* 2012;**5**:1064–72.
85. Atallah J, Erickson CC, Cecchin F, Dubin AM, Law IH, Cohen MI; Pediatric, Congenital Electrophysiology S et al. Multi-institutional study of implantable defibrillator lead performance in children and young adults: results of the Pediatric Lead Extractability and Survival Evaluation (PLEASE) study. *Circulation* 2013;**127**:2393–402.
86. Morquio L. Sur une maladie infantile et familiale caractérisée par des modifications permanentes du poulx, des attaques syncopales et épileptiformes et la mort subite. *Arch Med Enfants* 1901;**4**:467–75.
87. GCJ. VdH. Die ziekte van Stokes-Adams en een genal van aangeboren hart block (profschrift aan de ryks universitat). Gronigen 1908.
88. Lev M, Silverman J, Fitzmaurice FM, Paul MH, Cassels DE, Miller RA. Lack of connection between the atria and the more peripheral conduction system in congenital atrioventricular block. *Am J Cardiol* 1971;**27**:481–90.
89. Lev M, Cuadros H, Paul MH. Interruption of the atrioventricular bundle with congenital atrioventricular block. *Circulation* 1971;**43**:703–10.
90. Anderson RH, Wenick AC, Losekoot TG, Becker AE. Congenitally complete heart block. Developmental aspects. *Circulation* 1977;**56**:90–101.
91. Lev M. Pathogenesis of congenital atrioventricular block. *Prog Cardiovasc Dis* 1972;**15**:145–57.
92. Dewey RC, Capeless MA, Levy AM. Use of ambulatory electrocardiographic monitoring to identify high-risk patients with congenital complete heart block. *N Engl J Med* 1987;**316**:835–9.
93. Nagashima M, Nakashima T, Asai T, Matsushima M, Ogawa A, Ohsuga A et al. Study on congenital complete heart block in children by 24-hour ambulatory electrocardiographic monitoring. *Jpn Heart J* 1987;**28**:323–32.
94. Jaeggi ET, Hamilton RM, Silverman ED, Zamora SA, Hornberger LK. Outcome of children with fetal, neonatal or childhood diagnosis of isolated congenital atrioventricular block. A single institution's experience of 30 years. *J Am Coll Cardiol* 2002;**39**:130–7.
95. Breur JM, Udink Ten Cate FE, Kapusta L, Cohen MI, Crosson JE, Boramanand N et al. Pacemaker therapy in isolated congenital complete atrioventricular block. *Pacing Clin Electrophysiol* 2002;**25**:1685–91.
96. Michaelsson M, Jonzon A, Riesenfeld T. Isolated congenital complete atrioventricular block in adult life: a prospective study. *Circulation* 1995;**92**:442–9.
97. Thambo JB, Bordachar P, Garrigue S, Lafitte S, Sanders P, Reuter S et al. Detrimental ventricular remodeling in patients with congenital complete heart block and chronic right ventricular apical pacing. *Circulation* 2004;**110**:3766–72.
98. Ellenbogen KA, Wood MA. Pacemaker selection—the changing definition of physiologic pacing. *N Engl J Med* 2005;**353**:202–4.
99. Tops LF, Suffoletto MS, Bleeker GB, Boersma E, van der Wall EE, Gorcsan J, 3rd et al. Speckle-tracking radial strain reveals left ventricular dyssynchrony in patients with permanent right ventricular pacing. *J Am Coll Cardiol* 2007;**50**:1180–8.
100. Lillehei CW, Sellers RD, Bonnabeau RC, Eliot RS. Chronic postsurgical complete heart block with particular reference to prognosis, management, and a new P-wave pacemaker. *J Thorac Cardiovasc Surg* 1963;**46**:436–56.
101. Graham TP, Jr., Bernard YD, Mellen BG, Celermajer D, Baumgartner H, Cetta F et al. Long-term outcome in congenitally corrected transposition of the great arteries: a multi-institutional study. *J Am Coll Cardiol* 2000;**36**:255–61.
102. Beauchesne LM, Wames CA, Connolly HM, Ammash NM, Tajik AJ, Danielson GK. Outcome of the unoperated adult who presents with congenitally corrected transposition of the great arteries. *J Am Coll Cardiol* 2002;**40**:285–90.
103. Bonatti V, Agnetti A, Squarcia U. Early and late postoperative complete heart block in pediatric patients submitted to open-heart surgery for congenital heart disease. *Pediatr Med Chir* 1998;**20**:181–6.
104. Weindling SN, Saul JP, Gamble WJ, Mayer JE, Wessel D, Walsh EP. Duration of complete atrioventricular block after congenital heart disease surgery. *Am J Cardiol* 1998;**82**:525–7.
105. Liberman L, Silver ES, Chai PJ, Anderson BR. Incidence and characteristics of heart block after heart surgery in pediatric patients: a multicenter study. *J Thorac Cardiovasc Surg* 2016;**152**:197–202.
106. Predescu D, Chaturvedi RR, Friedberg MK, Benson LN, Ozawa A, Lee KJ. Complete heart block associated with device closure of perimembranous ventricular septal defects. *J Thorac Cardiovasc Surg* 2008;**136**:1223–8.
107. Yang R, Kong XQ, Sheng YH, Zhou L, Xu D, Yong YH et al. Risk factors and outcomes of post-procedure heart blocks after transcatheter device closure of perimembranous ventricular septal defect. *JACC Cardiovasc Interv* 2012;**5**:422–7.
108. Tao K, Lin K, Shi Y, Song H, Lui RC, Gan C et al. Periventricular device closure of perimembranous ventricular septal defects in 61 young children: early and midterm follow-up results. *J Thorac Cardiovasc Surg* 2010;**140**:864–70.
109. Butera G, Carminati M, Chessa M, Piazza L, Micheletti A, Negura DG et al. Transcatheter closure of perimembranous ventricular septal defects: early and long-term results. *J Am Coll Cardiol* 2007;**50**:1189–95.
110. Wolff GS, Rowland TW, Ellison RC. Surgically induced right bundle-branch block with left anterior hemiblock: an ominous sign in postoperative tetralogy of Fallot. *Circulation* 1972;**46**:587–94.
111. Krongrad E. Prognosis for patients with congenital heart disease and postoperative intraventricular conduction defects. *Circulation* 1978;**57**:867–70.
112. Lin A, Mahle WT, Frias PA, Fischbach PS, Kogon BE, Kanter KR et al. Early and delayed atrioventricular conduction block after routine surgery for congenital heart disease. *J Thorac Cardiovasc Surg* 2010;**140**:158–60.
113. Laurens P, Gavelle P, Pivnicia A, Farge C, Dubost C, Maurice P. Severe postoperative heart blocks appearing late. 16 cases. *Arch Mal Coeur Vaiss* 1983;**76**:1132–9.
114. Daicoff GR, Aslami A, Tobias JA, Miller BL. Management of postoperative complete heart block in infants and children. *Chest* 1974;**66**:639–41.
115. Dähn DKP. Zur Prognose der passageren AV-Ü berleitungsstörungen nach korrekiven Eingriffen am Herzen. *Klin Padiatr* 1976;**188**:263–6.
116. Murphy DA, Tynan M, Graham GR, Bonham-Carter RE. Prognosis of complete atrioventricular dissociation in children after open-heart surgery. *Lancet* 1970;**295**:750–2.
117. Squarcia U, Merideth J, McGoon DC, Weidman WH. Prognosis of transient atrioventricular conduction disturbances complicating open heart surgery for congenital heart defects. *Am J Cardiol* 1971;**28**:648–52.
118. Nishimura RA, Callahan MJ, Holmes DR, Jr., Gersh BJ, Driscoll DJ, Trusty JM et al. Transient atrioventricular block after open-heart surgery for congenital heart disease. *Am J Cardiol* 1984;**53**:198–201.
119. Gross GJ, Chiu CC, Hamilton RM, Kirsh JA, Stephenson EA. Natural history of postoperative heart block in congenital heart disease: implications for pacing intervention. *Heart Rhythm* 2006;**3**:601–4.
120. Batra AS, Wells WJ, Hinoki KW, Stanton RA, Silka MJ. Late recovery of atrioventricular conduction after pacemaker implantation for complete heart block associated with surgery for congenital heart disease. *J Thorac Cardiovasc Surg* 2003;**125**:1291–3.
121. De Raet J, Rega F, Meyns B. Late recovery of atrioventricular conduction after pacemaker implantation for complete heart block in congenital heart disease: fact or fluke? *Acta Chir Belg* 2010;**110**:323–7.
122. Bruckheimer E, Berul CI, Kopf GS, Hill SL, Warner KA, Kleinman CS et al. Late recovery of surgically-induced atrioventricular block in patients with congenital heart disease. *J Interv Card Electrophysiol* 2002;**6**:191–5.
123. Aziz PF, Serwer GA, Bradley DJ, LaPage MJ, Hirsch JC, Bove EL et al. Pattern of recovery for transient complete heart block after open heart surgery for congenital heart disease: duration alone predicts risk of late complete heart block. *Pediatr Cardiol* 2013;**34**:999–1005. [
124. Hofschire PJ, Nicoloff DM, Moller JH. Postoperative complete heart block in 64 children treated with and without cardiac pacing. *Am J Cardiol* 1977;**39**:559–62.
125. Taylor J. The 2010 version of the ESC guidelines for the management of grown-up adult congenital heart disease are discussed by guidelines task force chairman H. Baumgartner. *Eur Heart J* 2010;**31**:2825–6.
126. Ferrero P, Massa R, Amellone C, Trevi G. 'Sinus node' dysfunction associated with left atrial isomerism. *J Cardiovasc Med (Hagerstown)* 2008;**9**:953–6.
127. Anjos RT, Ho SY, Anderson RH. Surgical implications of juxtaposition of the atrial appendages. A review of forty-nine autopsied hearts. *J Thorac Cardiovasc Surg* 1990;**99**:897–904.
128. Attenhofer Jost CH, Connolly HM, Danielson GK, Bailey KR, Schaff HV, Shen WK et al. Sinus venous atrial septal defect: long-term postoperative outcome for 115 patients. *Circulation* 2005;**112**:1953–8.

129. Greenwood RD, Rosenthal A, Sloss LJ, LaCorte M, Nadas AS. Sick sinus syndrome after surgery for congenital heart disease. *Circulation* 1975;**52**:208–13.
130. Rajanbabu BB, Gangopadhyay D. Sinus node dysfunction after extracardiac conduit and lateral tunnel Fontan operation: the importance of the type of prior superior cavopulmonary anastomosis. *World J Pediatr Congenit Heart Surg* 2016;**7**:210–5.
131. Newcombe J, Gordon B, Razzouk A, Bailey L, Mandapati R. Extracardiac autologous pericardial tunnel Fontan allows implantation of an endocardial atrial lead for sinus node dysfunction. *Ann Thorac Surg* 2014;**98**:1094–6.
132. Dilawar M, Bradley SM, Saul JP, Stroud MR, Balaji S. Sinus node dysfunction after intraatrial lateral tunnel and extracardiac conduit fontan procedures. *Pediatr Cardiol* 2003;**24**:284–8.
133. Bae EJ, Lee JY, Noh CI, Kim WH, Kim YJ. Sinus node dysfunction after Fontan modifications—influence of surgical method. *Int J Cardiol* 2003;**88**:285–91.
134. Sun ZH, Happonen JM, Bennhagen R, Sairanen H, Pesonen E, Toivonen L et al. Increased QT dispersion and loss of sinus rhythm as risk factors for late sudden death after Mustard or Senning procedures for transposition of the great arteries. *Am J Cardiol* 2004;**94**:138–41.
135. Khairy P, Landtberg MJ, Lambert J, O'donnell CP. Long-term outcomes after the atrial switch for surgical correction of transposition: a meta-analysis comparing the Mustard and Senning procedures. *Cardiol Young* 2004;**14**:284–92.
136. Janousek J, Paul T, Luhmer I, Wilken M, Hruda J, Kalfelz HC. Atrial baffle procedures for complete transposition of the great arteries: natural course of sinus node dysfunction and risk factors for dysrhythmias and sudden death. *Zeitschrift Fur Kardiologie* 1994;**83**:933–8.
137. Sanders P, Morton JB, Kistler PM, Spence SJ, Davidson NC, Hussin A et al. Electrophysiological and electroanatomic characterization of the atria in sinus node disease: evidence of diffuse atrial remodeling. *Circulation* 2004;**109**:1514–22.
138. Anand N, McCrindle BW, Chiu CC, Hamilton RM, Kirsh JA, Stephenson EA et al. Chronotropic incompetence in young patients with late postoperative atrial flutter: a case-control study. *Eur Heart J* 2006;**27**:2069–73.
139. Hayward RM, Tseng ZH. Arrhythmias in complex congenital heart disease. *Card Electrophysiol Clin* 2014;**6**:623–34.
140. Garson A, Jr., Bink-Boelkens M, Hesslein PS, Hordof AJ, Keane JF, Neches WH et al. Atrial flutter in the young: a collaborative study of 380 cases. *J Am Coll Cardiol* 1985;**6**:871–8.
141. Kardelen F, Celiker A, Ozer S, Ozme S, Oto A. Sinus node dysfunction in children and adolescents: treatment by implantation of a permanent pacemaker in 26 patients. *Turk J Pediatr* 2002;**44**:312–6.
142. Brignole M, Auricchio A, Baron-Esquivias G, Bordachar P, Boriani G, Breithardt OA et al. 2013 ESC guidelines on cardiac pacing and cardiac resynchronization therapy: the task force on cardiac pacing and resynchronization therapy of the European Society of Cardiology (ESC). Developed in collaboration with the European Heart Rhythm Association (EHRA). *Europace* 2013;**15**:1070–118.
143. Alboni P, Brignole M, Menozzi C, Raviele A, Del Rosso A, Dinelli M et al. Efficacy and safety of out-of-hospital self-administered single-dose oral drug treatment in the management of infrequent, well-tolerated paroxysmal supraventricular tachycardia. *J Am Coll Cardiol* 2001;**37**:548–53.
144. Anderson JL, Halperin JL, Albert NM, Bozkurt B, Brindis RG, Curtis LH et al. Management of patients with atrial fibrillation (compilation of 2006 ACCF/AHA/ESC and 2011 ACCF/AHA/HRS recommendations): a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *J Am Coll Cardiol* 2013;**61**:1935–44.
145. Stan MN, Hess EP, Bahn RS, Warnes CA, Ammash NM, Brennan MD et al. A risk prediction index for amiodarone-induced thyrotoxicosis in adults with congenital heart disease. *J Thyroid Res* 2012;**2012**:210529.
146. Thorne SA, Barnes I, Cullinan P, Somerville J. Amiodarone-associated thyroid dysfunction risk factors in adults with congenital heart disease. *Circulation* 1999;**100**:149–54.
147. Banchs JE, Baquero GA, Nickolaus MJ, Wolbrette DL, Kelleman JJ, Samii S et al. Clinical efficacy of dofetilide for the treatment of atrial tachyarrhythmias in adults with congenital heart disease. *Congenit Heart Dis* 2014;**9**:221–7.
148. Wells R, Khairy P, Harris L, Anderson CC, Balaji S. Dofetilide for atrial arrhythmias in congenital heart disease: a multicenter study. *Pacing Clin Electrophysiol* 2009;**32**:1313–8.
149. El-Assaad I, Al-Kindi SG, Abraham J, Sanatani S, Bradley DJ, Halsey C et al. Use of dofetilide in adult patients with atrial arrhythmias and congenital heart disease: a PACES collaborative study. *Heart Rhythm* 2016;**13**:2034–9.
150. Connolly SJ et al. Dronedronone in high-risk permanent atrial fibrillation. *N Engl J Med* 2011;**365**:2268–76.
151. The Cardiac Arrhythmia Suppression Trial (CAST) Investigators. Preliminary report: effect of encainide and flecainide on mortality in a randomized trial of arrhythmia suppression after myocardial infarction. *N Engl J Med* 1989;**321**:406–12.
152. Fish FA1, Gillette PC, Benson DW Jr, Proarrhythmia, cardiac arrest and death in young patients receiving encainide and flecainide. The Pediatric Electrophysiology Group. *J Am Coll Cardiol* 1991;**18**:356–65.
153. Flaker GC, Blackshear JL, McBride R, Kronmal RA, Halperin JL, Hart RG. Antiarrhythmic drug therapy and cardiac mortality in atrial fibrillation. The Stroke Prevention in Atrial Fibrillation Investigators. *J Am Coll Cardiol* 1992;**20**:527–32.
154. Bordachar P, Marquie C, Pospiech T, Pasquie JL, Jalal Z, Haissaguerre M et al. Subcutaneous implantable cardioverter defibrillators in children, young adults and patients with congenital heart disease. *Int J Cardiol* 2016;**203**:251–8.
155. Lafuente-Lafuente C, Valembois L, Bergmann JF, Belmin J. Antiarrhythmics for maintaining sinus rhythm after cardioversion of atrial fibrillation. *Cochrane Database Syst Rev* 2015;**3**:CD005049.
156. DeWitt ES, Beausejour-Ladouceur V, Whitehill R, Bezzerides V, Mah D, Alexander ME, Triedman JK, Walsh EP, Abrams DJ. Radiofrequency ablation versus medical therapy for first episode of atrial tachycardia in adults with congenital heart disease. *Heart Rhythm* 2016;**13**:S449.
157. Lanz J, Brophy JM, Therrien J, Kaouache M, Guo L, Marelli AJ. Stroke in adults with congenital heart disease: incidence, cumulative risk, and predictors. *Circulation* 2015;**132**:2385–94.
158. Hoffmann A, Chockalingam P, Balint OH, Dadashev A, Dimopoulos K, Engel R et al. Cerebrovascular accidents in adult patients with congenital heart disease. *Heart* 2010;**96**:1223–6.
159. Lip GY, Nieuwlaar R, Pisters R, Lane D, Crijns H. Refining clinical risk stratification for predicting stroke in atrial fibrillation using a novel risk factor based approach. The Euro Heart Survey on Atrial Fibrillation. *Chest* 2010;**137**:263–72.
160. Camm AJ, Lip GY, De Caterina R, Savelieva I, Atar D, Hohnloser SH et al. 2012 focused update of the ESC Guidelines for the management of atrial fibrillation: an update of the 2010 ESC Guidelines for the management of atrial fibrillation. Developed with the special contribution of the European Heart Rhythm Association. *Europace* 2012;**14**:1385–413.
161. Kirchhof P, Benussi S, Kotecha D, Ahlsson A, Atar D, Casadei B et al. 2016 ESC Guidelines for the management of atrial fibrillation developed in collaboration with EACTS. *Europace* 2016;**18**:1609–78.
162. Kannankeril PJ, Anderson ME, Rottman JN, Wathen MS, Fish FA. Frequency of late recurrence of intra-atrial reentry tachycardia after radiofrequency catheter ablation in patients with congenital heart disease. *Am J Cardiol* 2003;**92**:879–81.
163. Singh SM, Neuzil P, Skoka J, Kriz R, Popelova J, Love BA et al. Percutaneous transhepatic venous access for catheter ablation procedures in patients with interruption of the inferior vena cava. *Circ Arrhythm Electrophysiol* 2011;**4**:235–41.
164. Correa R, Walsh EP, Alexander ME, Mah DY, Cecchin F, Abrams DJ et al. Transbaffle mapping and ablation for atrial tachycardias after mustard, senning, or Fontan operations. *J Am Heart Assoc* 2013;**2**:e000325.
165. Dave AS, Aboulhossn J, Child JS, Shivkumar K. Transconduit puncture for catheter ablation of atrial tachycardia in a patient with extracardiac Fontan palliation. *Heart Rhythm* 2010;**7**:413–6.
166. Aoki H, Nakamura Y, Takeno S, Takemura T. A new procedure for a transconduit puncture by grasping the dilator tip with a snare catheter: an alternative access method during catheter ablation of supraventricular tachycardias after an extracardiac Fontan operation. *Heart Rhythm* 2014;**11**:1492–4.
167. Krause U, Backhoff D, Klehs S, Schneider HE, Paul T. Transbaffle catheter ablation of atrial re-entrant tachycardia within the pulmonary venous atrium in adult patients with congenital heart disease. *Europace* 2016;**18**:1055–60.
168. Katritsis D, Zografos T, Katritsis G, Giazitzoglou E, Vachliotis V, Paxinos G et al. Catheter ablation vs antiarrhythmic drug therapy in patients with symptomatic AVNRT: a randomized controlled trial. *Europace* 2017;**19**:602–6.
169. Collins KK, Love BA, Walsh EP, Saul JP, Epstein MR, Triedman JK et al. Location of acutely successful radiofrequency catheter ablation of intraatrial reentrant tachycardia in patients with congenital heart disease. *Am J Cardiol* 2000;**86**:969–74.
170. Magnin-Poull I, De Chillou C, Miljoen H, Andronache M, Aliot E. Mechanisms of right atrial tachycardia occurring late after surgical closure of atrial septal defects. *J Cardiovasc Electrophysiol* 2005;**16**:681–7.
171. Ueda A, Suman-Horduna I, Mantziari L, Gujic M, Marchese P, Ho SY et al. Contemporary outcomes of supraventricular tachycardia ablation in congenital heart disease: a single-center experience in 116 patients. *Circ Arrhythm Electrophysiol* 2013;**6**:606–13.
172. Scaglione M, Caponi D, Ebrille E, Di Donna P, Di Clemente F, Battaglia A et al. Very long-term results of electroanatomic-guided radiofrequency ablation of atrial arrhythmias in patients with surgically corrected atrial septal defect. *Europace* 2014;**16**:1800–7.
173. Thomas P, Macicek S. Catheter ablation to treat supraventricular arrhythmia in children and adults with congenital heart disease: what we know and where we are going. *Ochsner J* 2016;**16**:290–6.

174. Triedman JK, Alexander ME, Love BA, Collins KK, Berul CI, Bevilacqua LM et al. Influence of patient factors and ablative technologies on outcomes of radiofrequency ablation of intra-atrial reentrant tachycardia in patients with congenital heart disease. *J Am Coll Cardiol* 2002;**39**:1827–35.
175. Tanner H, Lukac P, Schwick N, Fuhrer J, Pedersen AK, Hansen PS et al. Irrigated-tip catheter ablation of intraatrial reentrant tachycardia in patients late after surgery of congenital heart disease. *Heart Rhythm* 2004;**1**:268–75.
176. Yap S-C, Harris L, Silversides CK, Downar E, Chauhan VS. Outcome of intra-atrial re-entrant tachycardia catheter ablation in adults with congenital heart disease: negative impact of age and complex atrial surgery. *J Am Coll Cardiol* 2010;**56**:1589–96.
177. Escudero C, Khairy P, Sanatani S. Electrophysiologic considerations in congenital heart disease and their relationship to heart failure. *Can J Cardiol* 2013;**29**: 821–9.
178. Ho SY, Anderson RH. Conduction tissue in congenital heart surgery. *World J Surg* 1985;**9**:550–67.
179. Ho SY, Monro JL, Anderson RH. Disposition of the sinus node in left-sided juxtaposition of the atrial appendages. *Br Heart J* 1979;**41**:129–32.
180. Bae EJ, Noh CI, Choi JY, Yun YS, Kim WH, Lee JR et al. Twin AV node and induced supraventricular tachycardia in Fontan palliation patients. *Pacing Clin Electrophysiol* 2005;**28**:126–34.
181. Anderson RH, Arnold R, Thapar MK, Jones RS, Hamilton DI. Cardiac specialized tissue in hearts with an apparently single ventricular chamber (double inlet left ventricle). *Am J Cardiol* 1974;**33**:95–106.
182. Khairy P, Seslar SP, Triedman JK, Cecchin F. Ablation of atrioventricular nodal reentrant tachycardia in tricuspid atresia. *J Cardiovasc Electrophysiol* 2004;**15**: 719–22.
183. Watson H. Natural history of Ebstein's anomaly of tricuspid valve in childhood and adolescence. An international co-operative study of 505 cases. *Br Heart J* 1974;**36**:417–27.
184. Radford DJ, Graff RF, Neilson GH. Diagnosis and natural history of Ebstein's anomaly. *Br Heart J* 1985;**54**:517–22.
185. Smith WM, Gallagher JJ, Kerr CR, Sealy WC, Kasell JH, Benson DW, Jr et al. The electrophysiologic basis and management of symptomatic recurrent tachycardia in patients with Ebstein's anomaly of the tricuspid valve. *Am J Cardiol* 1982;**49**:1223–34.
186. Colavita PG, Packer DL, Pressley JC, Ellenbogen KA, O'callaghan WG, Gilbert MR et al. Frequency, diagnosis and clinical characteristics of patients with multiple accessory atrioventricular pathways. *Am J Cardiol* 1987;**59**:601–6.
187. Cappato R, Schluter M, Weiss C, Antz M, Koschik DH, Hofmann T et al. Radiofrequency current catheter ablation of accessory atrioventricular pathways in Ebstein's anomaly. *Circulation* 1996;**94**:376–83.
188. Reich JD, Auld D, Hulse E, Sullivan K, Campbell R. The pediatric radiofrequency ablation registry's experience with Ebstein's anomaly. Pediatric Electrophysiology Society. *J Cardiovasc Electrophysiol* 1998;**9**:1370–7.
189. Chetaille P, Walsh EP, Triedman JK. Outcomes of radiofrequency catheter ablation of atrioventricular reciprocating tachycardia in patients with congenital heart disease. *Heart Rhythm* 2004;**1**:168–73.
190. Wei W, Zhan X, Xue Y, Fang X, Liao H, Deng H et al. Features of accessory pathways in adult Ebstein's anomaly. *Europace* 2014;**16**:1619–2.
191. Levine JC, Walsh EP, Saul JP. Radiofrequency ablation of accessory pathways associated with congenital heart disease including heterotaxy syndrome. *Am J Cardiol* 1993;**72**:689–93.
192. Attenhofer Jost CH, Connolly HM, Edwards WD, Hayes D, Warnes CA, Danielson GK. Ebstein's anomaly—review of a multifaceted congenital cardiac condition. *Swiss Med Wkly* 2005;**135**:269–81.
193. Roten L, Lukac P, DE Groot N, Nielsen J, Szili-Torok T, Jensen H et al. Catheter ablation of arrhythmias in Ebstein's anomaly: a multicenter study. *J Cardiovasc Electrophysiol* 2011;**22**:1391–6.
194. da Silva JP, Baumgratz JF, da Fonseca L, Franchi SM, Lopes LM, Tavares GM et al. The cone reconstruction of the tricuspid valve in Ebstein's anomaly. The operation: early and midterm results. *J Thorac Cardiovasc Surg* 2007;**133**:215–23.
195. Silva J P D, Silva L D F D, Moreira LFP, Lopez LM, Franchi SM, Lianza AC et al. Cone reconstruction in Ebstein's anomaly repair: early and long-term results. *Arq Bras Cardiol* 2011;**97**:199–208.
196. Wu J, Deisenhofer I, Ammar S, Fichtner S, Reents T, Zhu P et al. Acute and long-term outcome after catheter ablation of supraventricular tachycardia in patients after the Mustard or Senning operation for D-transposition of the great arteries. *Europace* 2013;**15**:886–91.
197. Kay GN, Epstein AE, Dailey SM, Plumb VJ. Selective radiofrequency ablation of the slow pathway for the treatment of atrioventricular nodal reentrant tachycardia. Evidence for involvement of perinodal myocardium within the reentrant circuit. *Circulation* 1992;**85**:1675–88.
198. Carubicchio C, Santamaria M, Trevisi N, Maccabelli G, Giraldi F, Fassini G et al. Catheter ablation for the treatment of electrical storm in patients with implantable cardioverter-defibrillators: short- and long-term outcomes in a prospective single-center study. *Circulation* 2008;**117**:462–9.
199. Wijngaarden AP1, Zeppenfeld K. Ventricular tachycardia ablation: indications and techniques. *Minerva Cardioangiol* 2011;**59**:149–69.
200. Sabate Rotes A, Connolly HM, Warnes CA, Ammass NM, Phillips SD, Dearani JA et al. Ventricular arrhythmia risk stratification in patients with tetralogy of Fallot at the time of pulmonary valve replacement. *Circ Arrhythm Electrophysiol* 2015;**8**:110–6.
201. Gatzoulis MA, Freeman MA, Siu SC, Webb GD, Harris L. Atrial arrhythmia after surgical closure of atrial septal defects in adults. *N Engl J Med* 1999;**340**: 839–46.
202. Murphy JG, Gersh BJ, McGoon MD, Mair DD, Porter CJ, Ilstrup DM et al. Long-term outcome after surgical repair of isolated atrial septal defect. Follow-up at 27 to 32 years. *N Engl J Med* 1990;**323**:1645–50.
203. Triedman JK, Alexander ME, Berul CI, Bevilacqua LM, Walsh EP. Electroanatomic mapping of entrained and exit zones in patients with repaired congenital heart disease and intra-atrial reentrant tachycardia. *Circulation* 2001;**103**:2060–5.
204. Akar JG, Kok LC, Haines DE, DiMarco JP, Mounsey JP. Coexistence of type I atrial flutter and intra-atrial re-entrant tachycardia in patients with surgically corrected congenital heart disease. *J Am Coll Cardiol* 2001;**38**:377–84.
205. Peichl P, Kautzner J, Gebauer R. Ablation of atrial tachycardias after correction of complex congenital heart diseases: utility of intracardiac echocardiography. *Europace* 2008;**11**:48–53.
206. Giamberti A, Chessa M, Foresti S, Abella R, Butera G, de Vincentiis C et al. Combined atrial septal defect surgical closure and irrigated radiofrequency ablation in adult patients. *Ann Thorac Surg* 2006;**82**:1327–31.
207. Backhoff D, Muller M, Ruschewski W, Paul T, Krause U. ICD therapy for primary prevention of sudden cardiac death after Mustard repair for d-transposition of the great arteries. *Clin Res Cardiol* 2014;**103**:894–901.
208. de Groot NM, Lukac P, Schalijs MJ, Makowski K, Szili-Torok T, Jordaens L et al. Long-term outcome of ablative therapy of post-operative atrial tachyarrhythmias in patients with tetralogy of Fallot: a European multi-centre study. *Europace* 2012;**14**:522–7.
209. Lukac P, Pedersen AK, Mortensen PT, Jensen HK, Hjortdal V, Hansen PS et al. Ablation of atrial tachycardia after surgery for congenital and acquired heart disease using an electroanatomic mapping system: which circuits to expect in which substrate? *Heart Rhythm* 2005;**2**:64–72.
210. Seslar S, Alexander M, Berul C, Cecchin F, Walsh E, Triedman J et al. Ablation of nonautomatic focal atrial tachycardia in children and adults with congenital heart disease. *J Cardiovasc Electrophysiol* 2006;**17**:359–65.
211. Seiler J, Schmid DK, Irtel TA, Tanner H, Rotter M, Schwick N et al. Dual-loop circuits in postoperative atrial macro re-entrant tachycardias. *Heart* 2007;**93**: 325–30.
212. Mandapati R, Walsh EP, Triedman JK. Pericaval and periannular intra-atrial re-entrant tachycardias in patients with congenital heart disease. *J Cardiovasc Electrophysiol* 2003;**14**:119–25.
213. Brouwer C, Hazekamp MG, Zeppenfeld K. Anatomical substrates and ablation of reentrant atrial and ventricular tachycardias in repaired congenital heart disease. *Arrhythm Electrophysiol Rev* 2016;**5**:150–60.
214. Nakagawa H, Shah N, Matsudaira K, Overholt E, Chandrasekaran K, Beckman KJ et al. Characterization of reentrant circuit in macroreentrant right atrial tachycardia after surgical repair of congenital heart disease: isolated channels between scars allow “focal” ablation. *Circulation* 2001;**103**:699–709.
215. Verma A, Marrouche N, Seshadri N, Schweikert R, Bhargava M, Burkhardt JD et al. Importance of ablating all potential right atrial flutter circuits in postcardiac surgery patients. *J Am Coll Cardiol* 2004;**44**:409–14.
216. de Groot NM, Atary JZ, Blom NA, Schalijs MJ. Long-term outcome after ablative therapy of postoperative atrial tachyarrhythmia in patients with congenital heart disease and characteristics of atrial tachyarrhythmia recurrences. *Circ Arrhythm Electrophysiol* 2010;**3**:148–54.
217. Anguera I, Dallaglio P, Macías R, Jiménez-Candil J, Peinado R, García-Seara J et al. Long-term outcome after ablation of right atrial tachyarrhythmias after the surgical repair of congenital and acquired heart disease. *Am J Cardiol* 2015;**115**:1705–13.
218. Gallardo R, de Bono J. Ablation of arrhythmias in patients with adult congenital heart disease. *Arrhythm Electrophysiol Rev* 2014;**3**:36–9.
219. Troianos CA, Hartman GS, Glas KE, Skubas NJ, Eberhardt RT, Walker JD et al. Guidelines for performing ultrasound guided vascular cannulation: recommendations of the American Society of Echocardiography and the Society of Cardiovascular Anesthesiologists. *J Am Soc Echocardiogr* 2011;**24**:1291–318.
220. Acena M, Anguera I, Dallaglio PD, Rodriguez M, Sabaté X. Atrial fibrillation ablation in adults with repaired congenital heart disease. *J Atr Fibrillation* 2016;**8**: 1363.
221. Baldinger SH1, Stevenson WG, John RM. Ablation of ischemic ventricular tachycardia: evidence, techniques, results, and future directions.

222. Liang JJ, Santangeli P, Callans DJ. Long-term outcomes of ventricular tachycardia ablation in different types of structural heart disease. *Arrhythm Electrophysiol Rev* 2015;**4**:177–83.
223. Tung R, Boyle NG, Shivkumar K. Catheter ablation of ventricular tachycardia. *Circulation* 2010;**122**:e389–91.
224. Sadek MM, Schaller RD, Supple GE, Frankel DS, Riley MP, Hutchinson MD et al. Ventricular tachycardia ablation—the right approach for the right patient. *Arrhythm Electrophysiol Rev* 2014;**3**:161–7.
225. Driscoll DJ, Offord KP, Feldt RH, Schaff HV, Puga FJ, Danielson GK et al. Five- to 15-year follow-up after Fontan operation. *Circulation* 1992;**85**:469–96.
226. Flinn CJ, Wolff GS, Dick M, Campbell RM, Borkat G, Casta A et al. Cardiac rhythm after the Mustard operation for complete transposition of the great arteries. *N Engl J Med* 1984;**310**:1635–8.
227. Hickey EJ, Veldtman G, Bradley TJ, Gengsakul A, Manlhiot C, Williams WG et al. Late risk of outcomes for adults with repaired tetralogy of Fallot from an inception cohort spanning four decades. *Eur J Cardiothorac Surg* 2009;**35**:156–64.
228. Zomer AC, Verheugt CL, Vaartjes I, Uiterwaal CSPM, Langemeijer MM, Koolbergen DR et al. Surgery in adults with congenital heart disease. *Circulation* 2011;**124**:2195–201.
229. Mascio CE, Pasquali S, Jacobs J, Jacobs M, Austin III E. Outcomes in adult congenital heart surgery: analysis of the Society of Thoracic Surgeons database. *J Thorac Cardiovasc Surg* 2011;**142**:1090–7.
230. Stellin G, Vida VL, Padalino MA, Rizzoli G. Surgical outcome for congenital heart malformations in the adult age: a multicentric European study. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2004;**7**:95–101.
231. Blafox AD, Sleeper LA, Bradley DJ, Breitbart RE, Hordof A, Kanter RJ et al. Functional status, heart rate, and rhythm abnormalities in 521 Fontan patients 6 to 18 years of age. *J Thorac Cardiovasc Surg* 2008;**136**:100.e1–7.e1.
232. Deal BJ, Mavroudis C, Backer CL, Johnsrude CL, Rocchini AP. Impact of arrhythmia circuit cryoablation during Fontan conversion for refractory atrial tachycardia. *Am J Cardiol* 1999;**83**:563–8.
233. Delhaas T, Sarvaas GJ, Rijlaarsdam ME, Strengers JL, Eveleigh RM, Poulino SE et al. A multicenter, long-term study on arrhythmias in children with Ebstein anomaly. *Pediatr Cardiol* 2010;**31**:229–33.
234. Karamlou T, Silber I, Lao R, McCrindle B, Harris L. Outcome after late reoperation in patients with repaired tetralogy of Fallot: the impact of arrhythmia and arrhythmia surgery. *Ann Thorac Surg* 2006;**81**:1786–92.
235. Mavroudis C, Backer CL, Deal BJ. Late reoperations for Fontan patients: state of the art invited review. *Eur J Cardiothorac Surg* 2008;**34**:1034–40.
236. Rao HB1, Yu R, Chitnis N, DO D, Boyle NG2, Shivkumar K et al. Ventricular tachycardia ablation in the presence of left ventricular thrombus: safety and efficacy. *J Cardiovasc Electrophysiol* 2016;**27**:453–9.
237. Harrild DM, Berul CI, Cecchin F, Geva T, Gauvreau K, Pigula F et al. Pulmonary valve replacement in tetralogy of Fallot: impact on survival and ventricular tachycardia. *Circulation* 2009;**119**:445–51.
238. Brown ML, Dearani JA, Danielson GK, Cetta F, Connolly HM, Warnes CA; Mayo Clinic Congenital Heart Center et al. The outcomes of operations for 539 patients with Ebstein anomaly. *J Thorac Cardiovasc Surg* 2008;**135**:1120–36.
239. Mah DY, Alexander ME, Cecchin F, Walsh EP, Friedman JK. The electroanatomic mechanisms of atrial tachycardia in patients with tetralogy of Fallot and double outlet right ventricle. *J Cardiovasc Electrophysiol* 2011;**22**:1013–7.
240. Deal BJ, Costello JM, Webster G, Tsao B, Backer CL, Mavroudis C. Intermediate-term outcome of 140 consecutive Fontan conversions with arrhythmia operations. *Ann Thorac Surg* 2016;**101**:717–24.
241. Khairy P, Poirier N. Is the extracardiac conduit the preferred Fontan approach for patients with univentricular heart? The extracardiac conduit is not the preferred Fontan approach for patients with univentricular heart. *Circulation* 2010;**126**:2516–25.
242. Aboulhosn J, Williams R, Shivkumar K, Barkowski R, Plunkett M, Miner P et al. Arrhythmia recurrence in adult patients with single ventricle physiology following Fontan conversion. *Congenit Heart Dis* 2010;**5**:430–8.
243. Jonas RA. The intra/extracardiac conduit fenestrated Fontan. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2011;**14**:11–8.
244. Deal BJ, Mavroudis C, Backer CL. Beyond Fontan conversion: surgical therapy of arrhythmias including patients with associated complex heart disease. *Ann Thorac Surg* 2003;**76**:542–53. discussion 553–554.
245. Giamberti A, Chessa M, Abella R, Butera G, Negura D, Foresti S et al. Surgical treatment of arrhythmias in adults with congenital heart defects. *Int J Cardiol* 2008;**129**:37–41.
246. Giamberti A, Pluchinotta FR, Chessa M et al. Surgery for supraventricular tachycardia and congenital heart defects: long-term efficacy of the combined approach in adult patients. *Europace* 2017;**19**:1542–8.
247. Mavroudis C, Deal BJ, Backer CL, Tsao S. Arrhythmia surgery in patients with and without congenital heart disease. *Ann Thorac Surg* 2008;**86**:857–68.
248. Poh CL, Cochrane A, Galati JC, Bullock A, Celermajer DS, Gentles T et al. Ten-year outcomes of Fontan conversion in Australia and New Zealand demonstrate the superiority of a strategy of early conversion. *Eur J Cardiothorac Surg* 2016;**49**:530–5.
249. Kao JM, Alejos JC, Grant PW, Williams RG, Shannon KM, Laks H. Conversion of atriopulmonary to cavopulmonary anastomosis in management of late arrhythmias and atrial thrombosis. *Ann Thorac Surg* 1994;**58**:1510–4.
250. Huhta JC, Smallhorn JF, Macartney FJ, Anderson RH, De Leval M. Cross-sectional echocardiographic diagnosis of systemic venous return. *Br Heart J* 1982;**48**:388–403.
251. Buirski G, Jordan SC, Joffe HS, Wilde P. Superior vena caval abnormalities: their occurrence rate, associated cardiac abnormalities and angiographic classification in a paediatric population with congenital heart disease. *Clin Radiol* 1986;**37**:131–8.
252. McLeod CJ, Attenhofer Jost CH, Warnes CA, Hodge D 2nd, Hyberger L, Connolly HM et al. Epicardial versus endocardial permanent pacing in adults with congenital heart disease. *J Interv Card Electrophysiol* 2010;**28**:235–43.
253. Johnsrude CL, Backer CL, Deal BJ, Strasburger JF, Mavroudis C. Transmural atrial pacing in patients with postoperative congenital heart disease. *J Cardiovasc Electrophysiol* 1999;**10**:351–7.
254. McCanta AC, Kong MH, Carboni MP et al. Laser lead extraction in congenital heart disease: a case-controlled study. *Pacing Clin Electrophysiol* 2013;**36**:372–80.
255. Cecchin F, Atallah J, Walsh EP, Friedman JK, Alexander ME, Berul CI et al. Lead extraction in pediatric and congenital heart disease patients. *Circ Arrhythm Electrophysiol* 2010;**3**:437–44.
256. Garmreiter J, Whitaker P, Pilcher T, Etheridge S, Saarel E. Lumenless pacing leads: performance and extraction in pediatrics and congenital heart disease. *Pacing Clin Electrophysiol* 2015;**38**:42–7.
257. Shepherd E, Stuart G, Martin R, Walsh MA. Extraction of SelectSecure leads compared to conventional pacing leads in patients with congenital heart disease and congenital atrioventricular block. *Heart Rhythm* 2015;**12**:1227–32.
258. Tomaske M, Prêtre R, Rahn M, Bauersfeld U. Epicardial and pleural lead ICD systems in children and adolescents maintain functionality over 5 years. *Europace* 2008;**10**:1152–6.
259. Radbill AE, Friedman JK, Berul CI, Fynn-Thompson F, Atallah J, Alexander ME et al. System survival of nontransvenous implantable cardioverter-defibrillators compared to transvenous implantable cardioverter-defibrillators in pediatric and congenital heart disease patients. *Heart Rhythm* 2010;**7**:193–8.
260. Wilson DG, Zeb M, Veldtman G, Dimitrov BD, Morgan JM. Left and right parasternal sensing for the S-ICD in adult congenital heart disease patients and normal controls. *Pacing Clin Electrophysiol* 2016;**39**:282–90.
261. Chakraborti S, Szanthy G, Turner MS, Stuart G, Sauer RP. Use of radiofrequency perforation for lead placement in biventricular or conventional endocardial pacing after Mustard or Senning operations for D-transposition of the great arteries. *Pacing Clin Electrophysiol* 2009;**32**:1123–9.
262. Ruckdeschel ES1, Quafe R, Lewkowicz L, Kay J, Sauer WH, Collins KK et al. Preprocedural imaging in patients with transposition of the great arteries facilitates placement of cardiac resynchronization therapy leads. *Pacing Clin Electrophysiol* 2014;**37**:546–53.
263. Arif S, Cliff PF, De Giovanni JV. Permanent trans-venous pacing in an extra-cardiac Fontan circulation. *Europace* 2016;**18**:304–7.
264. Moore JP, Shannon KM. Transpulmonary atrial pacing: an approach to transvenous pacemaker implantation after extracardiac conduit fontan surgery. *J Cardiovasc Electrophysiol* 2014;**25**:1028–31. 766
265. Rosenthal E, Konta L. Transvenous atrial pacing from the superior vena cava stump after the Hemi-Fontan operation: a new approach. *Pacing Clin Electrophysiol* 2013;**37**:531–6.
266. Nery PB, Green MS, Khairy P. Implantable cardioverter-defibrillator insertion in congenital heart disease without transvenous access to the heart. *Can J Cardiol* 2013;**29**:254.e1–e3.
267. Adwani SS, Sreeram N, DeGiovanni JV. Percutaneous transhepatic dual chamber pacing in children with Fontan circulation. *Heart* 1997;**77**:574–5.
268. Chubb H, O'Neill M, Rosenthal E. Pacing and defibrillators in complex congenital heart disease. *Arrhythm Electrophysiol Rev* 2016;**5**:57–64.
269. Warnes CA, Williams RG, Bashore TM et al. ACC/AHA 2008 Guidelines for the management of adults with congenital heart disease: a report of the American College of Cardiology/American Heart Association Task Force on Practice Guidelines. *Circulation* 2008;**118**:e714–833.
270. Looma RS, Willes RJ, Kovach JR, Anderson RH. Chronic arrhythmias in the setting of heterotaxy: differences between right and left isomerism. *Congenit Heart Dis* 2016;**11**:7–18.
271. Anderson JB, Czosek RJ, Knillans TK, Meganathan K, Heaton P. Postoperative heart block in children with common forms of congenital heart disease: results from the KID Database. *J Cardiovasc Electrophysiol* 2012;**23**:1349–54.
272. Paech C, Dähnert I, Kostelka M, Mende M, Gebauer R. Association of temporary complete AV block and junctional ectopic tachycardia after surgery for congenital heart disease. *Ann Pediatr Cardiol* 2015;**8**:14–9.

273. Zeb M, Curzen N, Veldtman G, Yue A, Roberts P, Wilson D et al. Potential eligibility of congenital heart disease patients for subcutaneous implantable cardioverter-defibrillator based on surface electrocardiogram mapping. *Europace* 2015;**17**:1059–67.
274. Epstein AE, DiMarco JP, Ellenbogen KA, Estes NA 3rd, Freedman RA, Gettes LS; American College of Cardiology Foundation; American Heart Association Task Force on Practice Guidelines; Heart Rhythm Society et al. 2012 ACCF/AHA/HRS focused update incorporated into the ACCF/AHA/HRS 2008 guidelines for device-based therapy of cardiac rhythm abnormalities: a report of the American College of Cardiology Foundation/American Heart Association Task Force on Practice Guidelines and the Heart Rhythm Society. *J Am Coll Cardiol* 2013;**61**:e6–75.
275. Paul T, Ruschewski W, Janousek J. Guidelines for the management of congenital heart diseases in childhood and adolescence. Bradycardias and cardiac pacing in children. *Cardiol Young* 2017;**27**(Suppl. 3):S71–3.
276. Albin J, Hayes DL, Holmes DR. Sinus node dysfunction in pediatric and young adult patients: treatment by implantation of a permanent pacemaker in 39 cases. *Mayo Clin Proc* 1985;**60**:667–72.
277. Ector H, Van der Hauwaert LG. Sick sinus syndrome in childhood. *Br Heart J* 1980;**44**:684–91.
278. Beder SD, Gillette PC, Garson A, Jr, Porter CB, McNamara DG. Symptomatic sick sinus syndrome in children and adolescents as the only manifestation of cardiac abnormality or associated with unoperated congenital heart disease. *Am J Cardiol* 1983;**51**:1133–6.
279. Silka MJ, Manwill JR, Kron J, McNulty JH. Bradycardia-mediated tachyarrhythmias in congenital heart disease and responses to chronic pacing at physiologic rates. *Am J Cardiol* 1990;**65**:488–93.
280. Moak JP, Hasbani K, Ramwell C, Freedenberg V, Berger JT, DiRusso G et al. Dilated cardiomyopathy following right ventricular pacing for AV block in young patients: resolution after upgrading to biventricular pacing systems. *J Cardiovasc Electrophysiol* 2006;**17**:1068–71.
281. Kim JJ, Friedman RA, Eidem BW, Cannon BC, Arora G, Smith EO et al. Ventricular function and long-term pacing in children with congenital complete atrioventricular block. *J Cardiovasc Electrophysiol* 2007;**18**:373–7.
282. van Geldorp IE, Delhaas T, Gebauer RA, Frias P, Tomaske M, Friedberg MK, Working Group for Cardiac Dysrhythmias and Electrophysiology of the Association for European Paediatric Cardiology et al. Impact of the permanent ventricular pacing site on left ventricular function in children: a retrospective multicentre survey. *Heart* 2011;**97**:2051–5.
283. Janoušek J, van Geldorp IE, KrupicKová S, Rosenthal E, Nugent K, Tomaske M, Working Group for Cardiac Dysrhythmias and Electrophysiology of the Association for European Pediatric Cardiology et al. Permanent cardiac pacing in children: choosing the optimal pacing site: a multicenter study. *Circulation* 2013;**127**:613–23.
284. Acosta H, Víafora LM, Izquierdo D, Pothula VR, Bear J, Pothula S et al. Atrial lead placement at the lower atrial septum: a potential strategy to reduce unnecessary right ventricular pacing. *Europace* 2012;**14**:1311–6.
285. Wang M, Siu CW, Lee KL, Yue WS, Yan GH, Lee S et al. Effects of right low atrial septal vs. right atrial appendage pacing on atrial mechanical function and dyssynchrony in patients with sinus node dysfunction and paroxysmal atrial fibrillation. *Europace* 2011;**13**:1268–74.
286. Zilberman MV, Karpawich PP. Alternate site atrial pacing in the young: conventional echocardiography and tissue Doppler analysis of the effects on atrial function and ventricular filling. *Pacing Clin Electrophysiol* 2007;**30**:755–60.
287. Khairy P, Roux JF, Dubuc M, Thibault B, Guerra PG, Macle L et al. Laser lead extraction in adult congenital heart disease. *J Cardiovasc Electrophysiol* 2007;**18**:507–11.
288. Cecchin F, Atallah J, Walsh EP, Friedman JK, Alexander ME, Berul CI. Lead extraction in pediatric and congenital heart disease patients. *Circ Arrhythm Electrophysiol* 2010;**3**:437–44.
289. Fender EA, Killu AM, Cannon BC, Friedman PA, McLeod CJ, Hodge DO et al. Lead extraction outcomes in patients with congenital heart disease. *Europace* 2017;**19**:441–6.
290. Wilkoff BL, Love CJ, Byrd CL, Bongiorno MG, Carrillo RG, Crossley GH 3rd; Heart Rhythm Society; American Heart Association et al. Transvenous lead extraction: Heart Rhythm Society expert consensus on facilities, training, indications, and patient management: this document was endorsed by the American Heart Association (AHA). *Heart Rhythm* 2009;**6**:1085–10.
291. Yap SC, Roos-Hesselink JW, Hoendermis ES, Budts W, Vliegen HW, Mulder BJ et al. Outcome of implantable cardioverter defibrillators in adults with congenital heart disease: a multi-centre study. *Eur Heart J* 2007;**28**:1854–61.
292. Witte KK, Pepper CB, Cowan JC, Thomson JD, English KM, Blackburn ME. Implantable cardioverter-defibrillator therapy in adult patients with tetralogy of Fallot. *Europace* 2008;**10**:926–30.
293. Khanna AD, Warnes CA, Phillips SD, Lin G, Brady PA. Single-center experience with implantable cardioverter-defibrillators in adults with complex congenital heart disease. *Am J Cardiol* 2011;**108**:729–34.
294. Buber J, Ackley TJ, Daniels CJ, Roble SL, Mah ML, Kamp AN et al. Outcomes following the implantation of cardioverter-defibrillator for primary prevention in transposition of the great arteries after intra-atrial baffle repair: a single-centre experience. *Europace* 2016;**18**:1016–22.
295. Berul CI, Van Hare GF, Kertesz NJ, Dubin AM, Cecchin F, Collins KK et al. Results of a multicenter retrospective implantable cardioverter-defibrillator registry of pediatric and congenital heart disease patients. *J Am Coll Cardiol* 2008;**51**:1685–91.
296. Çeliker A, Olgun H, Karagoz T, Özer S, Özkutlu S, Alehan D. Midterm experience with implantable cardioverter-defibrillators in children and young adults. *Europace* 2010;**12**:1732–8.
297. Jordan CP, Freedenberg V, Wang Y, Curtis JP, Gleva MJ, Berul CI. Implant and clinical characteristics for pediatric and congenital heart patients in the National Cardiovascular Data Registry implantable cardioverter-defibrillator registry. *Circ Arrhythm Electrophysiol* 2014;**7**:1092–100.
298. Cannon BC, Friedman RA, Fenrich AL, Fraser CD, McKenzie ED, Kertesz NJ. Innovative techniques for placement of implantable cardioverter-defibrillator leads in patients with limited venous access to the heart. *Pacing Clin Electrophysiol* 2006;**29**:181–7.
299. Kella DK, Merchant FM, Veledar E, Book W, Lloyd MS. Lesion-specific differences for implantable cardioverter defibrillator therapies in adults with congenital heart disease. *Pacing Cardiac Electrophysiol* 2014;**37**:1492–8.
300. Ezzat V, Lee V, Ahsan S, Chow A, Segal O, Rowland E et al. A systematic review of ICD complications in randomised controlled trials versus registries: is our 'real-world' data an underestimation? *Open Heart* 2015;**2**:e000198. doi:10.1136/openhrt-2014-000198.
301. Cook SC, Valente AM, Maul TM, Dew MA, Hickey J, Jennifer Burger P et al. Shock-related anxiety and sexual function in adults with congenital heart disease and implantable cardioverter-defibrillators. *Heart Rhythm* 2013;**10**:805–10.
302. Moore JP, Mondésert B, Lloyd MS, Cook SC, Zaidi AN, Pass RH et al. Clinical experience with the subcutaneous implantable cardioverter-defibrillator in adults with congenital heart disease. *Circ Arrhythm Electrophysiol* 2016;**9**. doi:10.1161/CIRCEP.116.004338.
303. Kalra Y, Radbill AE, Johns JA, Fish FA, Kannankeril PJ. Antitachycardia pacing reduces appropriate and inappropriate shocks in children and congenital heart disease patients. *Heart Rhythm* 2012;**9**:1829–34.
304. Ghai A, Silversides C, Harris L, Webb GD, Siu SC, Therrien J. Left ventricular dysfunction is a risk factor for sudden cardiac death in adults late after repair of tetralogy of Fallot. *J Am Coll Cardiol* 2002;**40**:1675–80.
305. Gallego P, Gonzalez AE, Sanchez-Recalde A, Peinado R, Polo L, Gomez-Rubin C et al. Incidence and predictors of sudden cardiac arrest in adults with congenital heart defects repaired before adult life. *Am J Cardiol* 2012;**110**:109–17.
306. Everitt MD, Donaldson AE, Stehlik J, Kaza AK, Budge D, Alharethi R et al. Would access to device therapies improve transplant outcomes for adults with congenital heart disease? Analysis of the United Network for Organ Sharing (UNOS). *J Heart Lung Transplant* 2011;**30**:395–401.
307. Aziz S, Leon AR, El-Chami MF. The subcutaneous defibrillator: a review of the literature. *J Am Coll Cardiol* 2014;**63**:1473–9.
308. Tan VH, Wilton SB, Kuriachan V, Sumner GL, Exner DV. Impact of programming strategies aimed at reducing nonessential implantable cardioverter defibrillator therapies on mortality: a systematic review and meta-analysis. *Circ Arrhythm Electrophysiol* 2014;**7**:164–70.
309. Garnreiter JM, Pilcher TA, Etheridge SP, Saarel EV. Inappropriate ICD shocks in pediatrics and congenital heart disease patients: risk factors and programming strategies. *Heart Rhythm* 2015;**12**:937–42.
310. Wilkoff BL, Fauchier L, Stiles MK, Morillo CA, Al-Khatib SM, Almendral J et al. 2015 HRS/EHRA/APHS/SOLACE expert consensus statement on optimal implantable cardioverter-defibrillator programming and testing. *Europace* 2016;**18**:159–83.
311. Prinzen FW, Hunter WC, Wyman BT, McVeigh ER. Mapping of regional myocardial strain and work during ventricular pacing: experimental study using magnetic resonance imaging tagging. *J Am Coll Cardiol* 1999;**33**:1735–42.
312. Chakir K, Daya SK, Tunin RS, Helm RH, Byrne MJ, Dimaano VL et al. Reversal of global apoptosis and regional stress kinase activation by cardiac resynchronization. *Circulation* 2008;**117**:1369–77.
313. Vanderheyden M, Mullens W, Delrue L, Goethals M, de Bruyne B, Wijns W et al. Myocardial gene expression in heart failure patients treated with cardiac resynchronization therapy responders versus nonresponders. *J Am Coll Cardiol* 2008;**51**:129–36.
314. Zareba W, Klein H, Cygankiewicz I, Hall WJ, McNitt S, Brown M, MADIT-CRT Investigators et al. Effectiveness of Cardiac Resynchronization Therapy by QRS Morphology in the Multicenter Automatic Defibrillator Implantation Trial-

- Cardiac Resynchronization Therapy (MADIT-CRT). *Circulation* 2011;**123**: 1061–72.
315. van Geldorp IE, Bordachar P, Lumens J, de Gillebon M, Whinnett ZI, Prinzen FW *et al.* Acute hemodynamic benefits of biventricular and single-site systemic ventricular pacing in patients with a systemic right ventricle. *Heart Rhythm* 2013; **10**:676–82.
 316. Risum N, Jons C, Olsen NT, Fritz-Hansen T, Bruun NE, Hojgaard MV *et al.* Simple regional strain pattern analysis to predict response to cardiac resynchronization therapy: rationale, initial results, and advantages. *Am Heart J* 2012;**163**: 697–704.
 317. Kass DA. An epidemic of dyssynchrony: but what does it mean? *J Am Coll Cardiol* 2008;**51**:12–7.
 318. Cleland JG, Daubert JC, Erdmann E, Freemantle N, Gras D, Kappenberger L *et al.* The effect of cardiac resynchronization on morbidity and mortality in heart failure. *N Engl J Med* 2005;**352**:1539–49.
 319. Hui W, Slorach C, Dragulescu A, Mertens L, Bijns B, Friedberg MK. Mechanisms of right ventricular electromechanical dyssynchrony and mechanical inefficiency in children after repair of tetralogy of Fallot. *Circ Cardiovasc Imaging* 2014;**7**:610–8.
 320. Linde C, Abraham WT, Gold MR, St John Sutton M, Ghio S, Daubert C. Randomized trial of cardiac resynchronization in mildly symptomatic heart failure patients and in asymptomatic patients with left ventricular dysfunction and previous heart failure symptoms. *J Am Coll Cardiol* 2008;**52**:1834–43.
 321. Moss AJ, Hall WJ, Cannom DS, Klein H, Brown MW, Daubert JP *et al.* Cardiac-resynchronization therapy for the prevention of heart failure events. *N Engl J Med* 2009;**361**:1329–38.
 322. Strieper M, Karpawich P, Frias P, Gooden K, Ketchum D, Fyfe D *et al.* Initial experience with cardiac resynchronization therapy for ventricular dysfunction in young patients with surgically operated congenital heart disease. *Am J Cardiol* 2004;**94**:1352–4.
 323. Koyak Z, de Groot JR, Krimly A, Mackay TM, Bouma BJ, Silversides CK *et al.* Cardiac resynchronization therapy in adults with congenital heart disease. *Europace* 2017 Jan 20 [Epub ahead of print];pii:euw386. doi: 10.1093/europace/euw386.
 324. Khairy P, Fournier A, Thibault B, Dubuc M, Therien J, Vobecky SJ. Cardiac resynchronization therapy in congenital heart disease. *Int J Cardiol* 2006;**109**: 160–8.
 325. Dubin AM, Feinstein JA, Reddy VM, Hanley FL, Van Hare GF, Rosenthal DN. Electrical resynchronization: a novel therapy for the failing right ventricle. *Circulation* 2003;**107**:2287–9.
 326. Thambo JB, Dos Santos P, De Gillebon M, Roubertie F, Labrousse L, Sacher F *et al.* Biventricular stimulation improves right and left ventricular function after tetralogy of Fallot repair: acute animal and clinical studies. *Heart Rhythm* 2010;**7**: 344–50.
 327. Kubuš P, Materna O, Tax P, Tomek V, Janoušek J. Successful permanent resynchronization for failing right ventricle after repair of tetralogy of Fallot. *Circulation* 2014;**130**:e186–90.
 328. Gebauer RA, Tomek V, Salameh A, Marek J, Chaloupecký V, Gebauer R *et al.* Predictors of left ventricular remodelling and failure in right ventricular pacing in the young. *Eur Heart J* 2009;**30**:1097–104.
 329. Horovitz A, De Gillebon M, van Geldorp IE, Bordachar P, Roubertie F, Iriart X *et al.* Effects of nonsystemic ventricular pacing in patients with transposition of the great arteries and atrial redirection. *J Cardiovasc Electrophysiol* 2012;**23**: 766–70.
 330. Jauvert G, Rousseau-Paziaud J, Villain E, Iserin L, Hidden-Lucet F, Ladouceur M *et al.* Effects of cardiac resynchronization therapy on echocardiographic indices, functional capacity, and clinical outcomes of patients with a systemic right ventricle. *Europace* 2009;**11**:184–90.
 331. Janousek J, Tomek V, Chaloupecký VA, Reich O, Gebauer RA, Kautzner J *et al.* Cardiac resynchronization therapy: a novel adjunct to the treatment and prevention of systemic right ventricular failure. *J Am Coll Cardiol* 2004;**44**:1927–31.
 332. Cecchin F, Frangini PA, Brown DW, Fynn-Thompson F, Alexander ME, Friedman JK. Cardiac resynchronization therapy (and multisite pacing) in pediatric and congenital heart disease: five years experience in a single institution. *J Cardiovasc Electrophysiol* 2009;**20**:58–65.
 333. Dubin AM, Janousek J, Rhee E, Strieper MJ, Cecchin F, Law IH *et al.* Resynchronization therapy in pediatric and congenital heart disease patients: an international multicenter study. *J Am Coll Cardiol* 2005;**46**:2277–83.
 334. Janousek J, Gebauer RA, Abdul-Khalik H, Turner M, Kornyei L, Grollmuss O *et al.* Cardiac resynchronization therapy in paediatric and congenital heart disease: differential effects in various anatomical and functional substrates. *Heart* 2009;**95**:1165–71.
 335. Janousek J, Vojtovic P, Hucin B, Tlaskal T, Gebauer RA, Gebauer R *et al.* Resynchronization pacing is a useful adjunct to the management of acute heart failure after surgery for congenital heart defects. *Am J Cardiol* 2001;**88**:145–52.
 336. Thambo JB, De Gillebon M, Xhaet O, Dos Santos P, Roubertie F, Labrousse L *et al.* Biventricular pacing in patients with tetralogy of Fallot: non-invasive epicardial mapping and clinical impact. *Int J Cardiol* 2013;**163**:170–4.
 337. Plymen CM, Finlay M, Tsang V, O'leary J, Picaut N, Cullen S *et al.* Haemodynamic consequences of targeted single- and dual-site right ventricular pacing in adults with congenital heart disease undergoing surgical pulmonary valve replacement. *Europace* 2015;**17**:274–80.
 338. Thibault B, Harel F, Ducharme A, White M, Ellenbogen KA, Frasure-Smith N *et al.* Cardiac resynchronization therapy in patients with heart failure and a QRS complex <120 milliseconds: the evaluation of resynchronization therapy for heart failure (LESSER-EARTH) trial. *Circulation* 2013;**127**:873–81.
 339. Sakaguchi H, Miyazaki A, Yamada O, Kagisaki K, Hoashi T, Ichikawa H *et al.* Cardiac resynchronization therapy for various systemic ventricular morphologies in patients with congenital heart disease. *Circ J* 2015;**79**:649–55.