Adults with Congenital Heart Disease and Arrhythmia Management

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KEYWORDS

• Adult congenital heart disease • Intra-atrial reentrant tachycardia • Ventricular tachycardia
• Catheter ablation • Sinus node dysfunction • Sudden cardiac death • Pacemaker
• Implantable cardioverter-defibrillator

KEY POINTS

- Atrial tachyarrhythmias are the most common rhythm disturbances in adult congenital heart disease (ACHD) and are characterized by a gradual transition from intra-atrial reentrant tachycardia to atrial fibrillation over the lifetime of the individual.
- Ventricular tachyarrhythmia and sudden cardiac death occur up to 100 times more frequently among ACHD than acquired heart disease.
- Cardiac resynchronization is an important adjunctive therapy for select ACHD patients with heart failure and electrical dyssynchrony.
- There have been major advances in rhythm management for ACHD in recent years, stemming from a combination of technologic and technical innovations.

Video content accompanies this article at http://www.cardiology.theclinics.com.

OVERVIEW

A rapid expansion of the population of adults with congenital heart disease (ACHD) and arrhythmia has given rise to the specialty of ACHD electrophysiology. ACHD patients experience rhythm abnormalities that are often poorly tolerated and require advanced medical or interventional therapies. Common examples are the bradyarrhythmias of sinus node dysfunction and atrioventricular (AV) block, the tachyarrhythmias of intra-atrial reentrant tachycardia, atrial fibrillation, and ventricular tachycardia (VT), and pathologic electrical delay resulting in "ventricular dyssynchrony" (**Fig. 1**). In addition, primary prevention of sudden cardiac death is indicated in many situations. This article reviews the pathophysiology, clinical characteristics, and treatment strategies for these commonly encountered arrhythmias among ACHD.

SINUS NODE DYSFUNCTION

Sinus node dysfunction (SND) is prevalent in ACHD, primarily because of the cumulative effects of atrial distention and fibrosis from abnormal hemodynamics and direct surgical trauma to the sinoatrial nodal complex or its vascular supply. Patients at particular risk include those who have undergone the Mustard or Senning operation[1,2] and cavopulmonary shunts. SND after the Mustard operation, for instance, has been reported to be as great as 50% at

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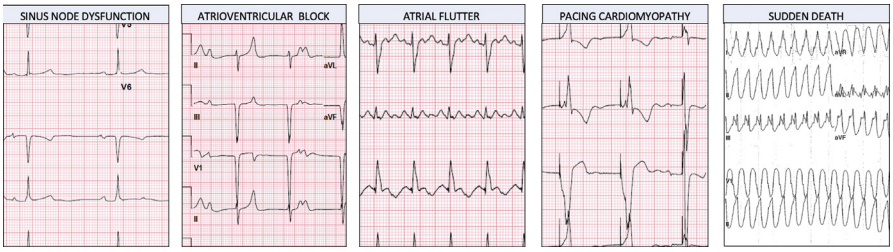

Fig. 1. Summary of the major forms of rhythm disturbance among patients with adult congenital heart disease.

20 years after surgery.[2] Both the lateral tunnel and extracardiac total cavopulmonary connection (TCPC) operations are associated with an approximately 15% risk of SND at 10 years after surgery.[3] Maintenance of sinus rhythm is important for optimal Fontan physiology,[4] and SND has been implicated in manifestations of Fontan failure (eg, plastic bronchitis or protein-losing enteropathy) that may be reversible with restoration of atrial-based rhythm.[5]

Indications for pacing in the setting of SND are firstly symptom based, but may also be guided by significant bradycardia.[6] Transvenous pacing is usual for patients who are remote from surgery unless there are unacceptable impediments to this approach (eg, limited access to the atrial myocardium or residual intracardiac shunt).[7] Many perceived barriers can be overcome by innovative techniques. For instance, transhepatic-transbaffle approaches have been described, and transvenous atrial pacing after extracardiac Fontan operation is technically feasible by transpulmonary puncture.[8,9] (**Fig. 2**). Long-term outcomes of these unconventional strategies remain to be determined, particularly with regard to complications such as bleeding, lead migration, and hepatic function (with transhepatic approaches), and concerns regarding lead extractions in the event of infection. Surgical pacing is typically used if there is known SND at the time of operation or when converting an older style of Fontan to TCPC.[10]

ATRIOVENTRICULAR BLOCK

AV block may occur either spontaneously or as an iatrogenic sequela of operative repair for ACHD. Spontaneous AV block is notably seen in congenitally corrected transposition of the great arteries (CCTGA), where superior displacement of the AV node results in a long and tenuous nonbranching conducting bundle.[11,12] The annual incidence of spontaneous AV block in CCTGA is reported to be approximately 2%.[13] Other important congenital defects associated with spontaneous AV block include AV septal defects and atrial septal defects.[14] Surgical iatrogenic AV block has become less common with advanced surgical techniques, but can occur when suture lines are placed in the vicinity of the AV conduction system.

More recently, leadless cardiac pacing has emerged as an option to avoid problems associated with conventional pacing. These include intravascular infection, lead fracture, and venous thrombosis. Among ACHD, additional concerns include preservation of AV valve function and thromboembolic risk (**Fig. 3**).[15] Importantly, current leadless pacing systems are capable of pacing and sensing only the ventricular myocardium

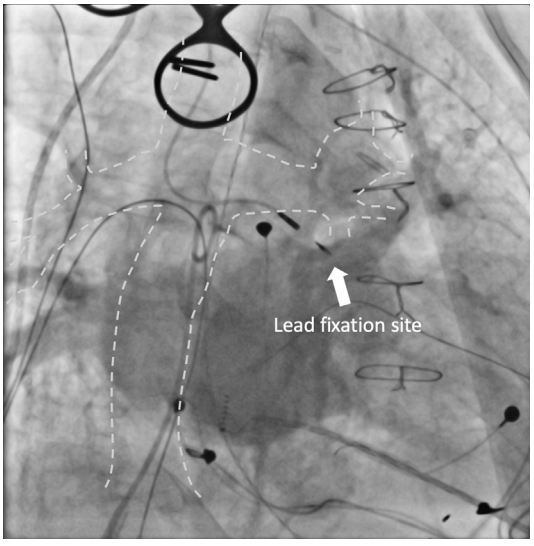

Fig. 2. Transvenous approach to epicardial atrial pacing for an extracardiac Fontan patient. Dotted white lines represent the course of the Fontan pathway. The pacing lead has been placed from the right subclavian vein, through the Glenn anastomosis, and into the pulmonary artery. Puncture into the extracardiac space was performed and the lead affixed to the atrial epi-myocardium. This approach can be used for patients in whom a surgical thoracotomy is undesirable.

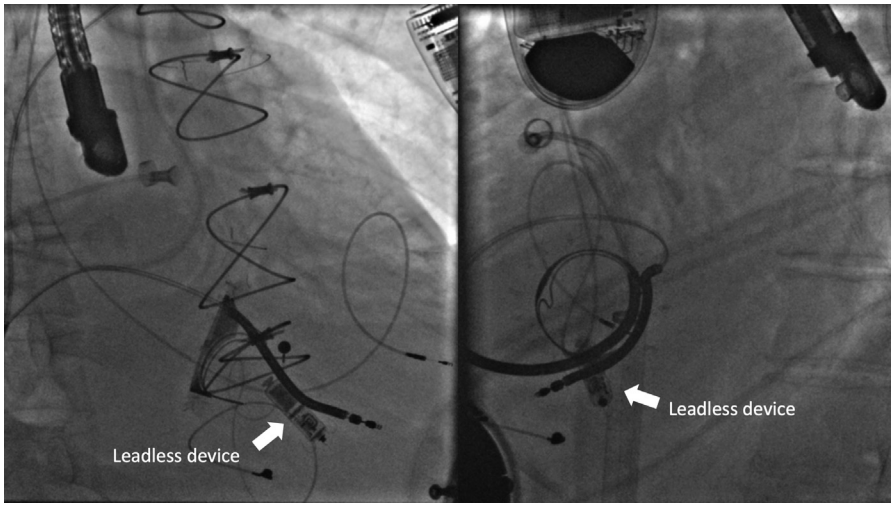

Fig. 3. Placement of a leadless cardiac pacemaker from the femoral approach in a patient with permanent atrial fibrillation and atrioventricular block after tricuspid valve replacement. The prior ICD lead had failed after being entrapped during the valve surgery, leaving the patient with a slow ventricular escape rhythm. The leadless pacemaker was implanted to avoid future disruption of tricuspid valve function.

and are therefore limited in their application. Future technology intends to identify atrial contraction from intracardiac accelerometer waveforms, permitting maintenance of AV synchrony and expanding the currently limited leadless pacemaker implant indications.[16]

CARDIAC RESYNCHRONIZATION THERAPY

Spontaneous or pacing-induced electrical delay develops frequently among patients with congenital heart disease (CHD). Abnormal electrical excitation can result in inefficient myocardial energetics, with early contraction and simultaneous systolic stretch of opposing ventricular walls. For acquired heart disease, cardiac resynchronization therapy (CRT) has been shown to promote reverse ventricular remodeling, improve quality of life, and decrease mortality.[17]

Among patients with ACHD and a systemic left ventricle, there is evidence that CRT is beneficial for both spontaneous or pacing-induced types of left ventricular (LV) electromechanical delay,[18] but there are fewer data for other forms of CHD. In a limited fashion, CRT has also been evaluated in the context of failure of the subpulmonary right ventricle, the systemic right ventricle, and the single ventricle.

Subpulmonary Right Ventricle

Subpulmonary right ventricular (RV) dysfunction is not uncommon among patients with tetralogy of Fallot and related variants, where right bundle branch block can contribute to ventricular dyssynchrony, often in combination with hemodynamic derangements of the pulmonary or tricuspid valve(s). Initially, pacing studies were limited to acute improvement in RV systolic function in the catheterization laboratory or in the postoperative setting.[19,20] More recently, long-term improvement in New York Heart Association class and objective measures of aerobic capacity have been demonstrated,[21] along with indices of RV remodeling.[22] In general, strategies involve lead fixation at the site of latest RV endocardial activation to achieve fusion with intrinsic AV conduction. Currently CRT for the subpulmonary right ventricle carries a class IIb indication based on limited data.[6]

Systemic Right Ventricle

A significant proportion of patients with *d*-transposition of the great arteries (DTGA) after the Senning or Mustard operation or unrepaired or physiologically repaired CCTGA carry a CRT indication based on current guidelines.[23] The feasibility and potential benefits of CRT for the systemic right ventricle were initially demonstrated for CCTGA patients undergoing concomitant surgery,[24] and since then various multicenter[18,25,26] and single-center studies have shown potential benefit. Although most have reported a favorable response, a notable minority have reported either very poor response[26] or even clinical deterioration.[27] Accordingly, there remains uncertainty as to the role of CRT for the systemic right ventricle,

and this topic has been identified as a high-impact research question in recent ACHD clinical practice guidelines.[28]

An important consideration for systemic RV resynchronization is the route of implantation. For patients with CCTGA, the cardiac veins are anomalous in ~20% of patients, with ectopic location, duplication, and atresia reported.[29] Despite this, successful cannulation and CRT lead placement via a posteroseptal coronary sinus ostium can be achieved in most patients, with alternative cannulation techniques for the remainder (**Fig. 4**).[30] In rare CCTGA and most Mustard or Senning patients, epicardial lead placement may be required and can be approached via lateral thoracotomy or lower midline sternotomy, respectively.[31]

Single Ventricle

Electrical dyssynchrony, especially resulting from permanent ventricular pacing, has been shown to be associated with progressive AV valve regurgitation, ventricular systolic dysfunction, and reduced transplant-free survival among patients with single-ventricle anatomy.[32,33] Echocardiographic characterization of mechanical contraction in the form of classic-pattern dyssynchrony may be useful for a subset of patients who are most likely to respond favorably to CRT.[34] Although both multisite pacing[35] and single ventricular apical pacing[32]

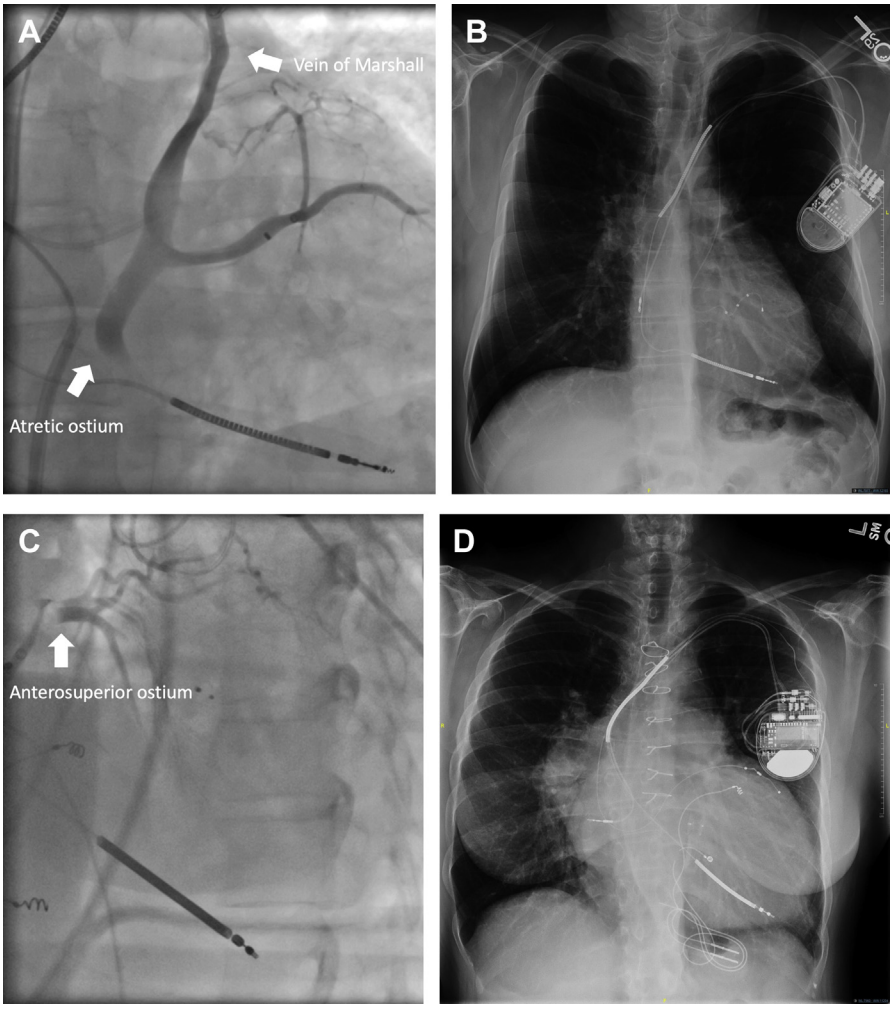

Fig. 4. Transvenous cardiac resynchronization therapy lead placement for patients with CCTGA with absent or diminutive posteroseptal coronary sinus ostia (CSO). (*A*, *B*) Intraprocedural lead placement and postoperative radiograph for a patient with atresia of the posterior CSO. A lead was placed through a persistent vein of Marshall that drained the coronary venous circulation. (*C*, *D*) Identical views of a patient with a diminutive conventional posterior CSO with lead placement through a large vein at the base of the right atrial appendage.

have been proposed as strategies to improve long-term clinical outcomes, data are lacking, and the optimal approach to resynchronization for the single-ventricle patient remains unknown.

CONDUCTION SYSTEM PACING

Recently, direct His-bundle pacing has been shown to be a physiologic alternative to CRT in acquired heart disease. This approach circumvents many of the challenges encountered in conventional CRT that include inadequate coronary venous tributaries, elevated ventricular pacing threshold, phrenic nerve capture, and lead dislodgment, among others. His-bundle pacing may be particularly useful for ACHD patients with, or at high risk for, pacing-induced cardiomyopathy, especially when combined with challenging coronary sinus anatomy such as CCTGA (**Fig. 5**).[30] To date, only isolated case reports demonstrate the feasibility of His-bundle pacing for CCTGA[36–38] and further data are needed before there is widespread endorsement of this approach. This issue was the topic of a recent multicenter investigation conducted by the joint Pediatric and Congenital Electrophysiology Society and International Society of Adult Congenital Heart Disease Electrophysiology Research Collaboration.[39]

SUPRAVENTRICULAR TACHYCARDIA

Intra-Atrial Reentrant Tachycardia

Intra-atrial reentrant tachycardia (IART) is the most common tachyarrhythmia observed among patients with ACHD, with a cumulative incidence approaching 50% by the age of 65 years.[40] The development of IART is associated with multiple adverse clinical outcomes including stroke, heart failure, and all-cause mortality.[40,41] Of patients developing IART, those with DTGA and Mustard or Senning baffles, pulmonary hypertension, valvular heart disease, and single-ventricle anatomy face the highest mortality risk.[41,42]

In many cases, IART is the cumulative effect of diffuse injury to the atrial myocardium and maturation of surgical barriers, both highly prevalent in ACHD.[43] Importantly, long-term antiarrhythmic drug therapy for maintenance of sinus rhythm and ventricular rate control for IART are generally considered only moderately effective strategies in ACHD. Instead, 3 principal management strategies are useful for maintenance of sinus rhythm.

Catheter ablation

D-transposition of the great arteries after the Mustard or Senning operation IART in the setting of DTGA after the Mustard or Senning operation is frequently associated with a relatively slow atrial rate owing to extensive areas of diseased myocardium and robust AV node conduction. This can result in 1:1 atrial-ventricular conduction, and together with an inherently compromised systemic right ventricle may be associated with degeneration to malignant ventricular arrhythmia.[42] IART circuits for these patients are usually biatrial or situated within the morphologic right atrium, which is separated from the venous circulation by a

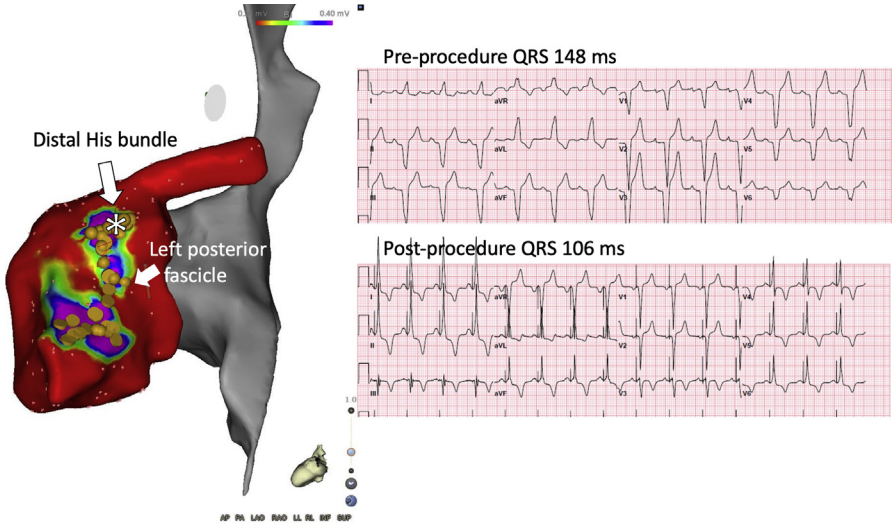

Fig. 5. A patient with CCTGA and heart block who developed pacing-induced cardiomyopathy after conventional dual-chamber pacemaker placement. The morphologic left conduction system was mapped and a lead was affixed to the distal His bundle (*asterisk*). The QRS duration decreased following this procedure, and the heart failure resolved.

surgical baffle. This chamber can therefore be approached by direct baffle puncture or by a retrograde course.[44,45]

Most frequently, IART circuits develop around the tricuspid valve annulus.[46] Additional reentrant circuits have been shown to depend on the underlying surgical anatomy. After the Mustard operation, for instance, reentry around a morphologic right atrial free wall atriotomy or patch is common, whereas reentry around the right pulmonary veins and the nearby surgical counterincision is observed after the Senning operation (**Fig. 6**, Video 1).[47–49] The mapping procedure entails thorough evaluation of the entire morphologic right atrium to target all reentry circuits and reduce the risk for possible recurrent tachycardia. Ultrahigh-density mapping may play a role in more precise circuit delineation so that these substrates may be comprehensively targeted.[50] Other substrates that are often observed include focal atrial tachycardias and AV nodal reentry tachycardia.[47,51] Tachycardia recurrence after IART ablation is reportedly 30%,[46,47] but a more contemporary study involving comprehensive circuit delineation using high-density mapping is lacking.

Fontan operation The modified atriopulmonary Fontan operation is associated with massive right atrial enlargement owing to long-standing venous hypertension. The incidence of IART after this surgery has been estimated to be 50% at 20 postoperative years. Surgical placement of the atrial baffle, the anastomosis from the morphologic right atrium to the pulmonary artery, and the atriotomy incision may also serve as electrical barriers for reentry (**Fig. 7**, Video 2).[43,52] Moreover, reentry around the systemic AV annulus and a dilated inferior vena cava may also occur, resulting in multiple potential circuits in any given individual.[53,54] Evaluation for periannular reentry usually requires baffle puncture when present.[45] Extreme atrial muscle hypertrophy after the atriopulmonary Fontan operation[55] can challenge the

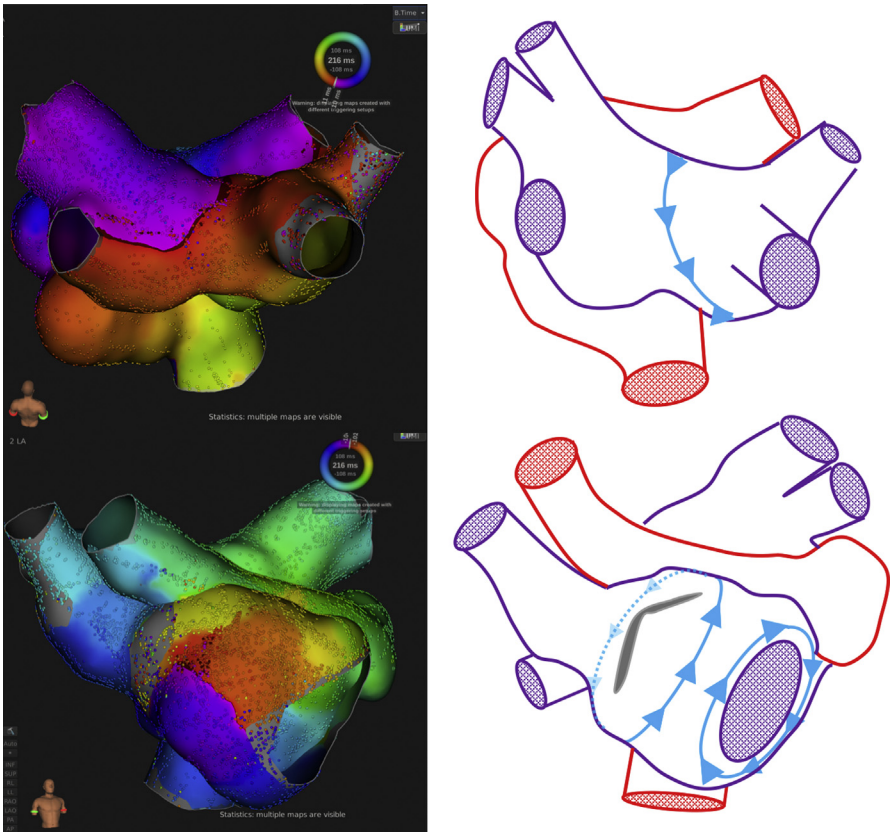

Fig. 6. Demonstration of intra-atrial reentry circuits after Senning operation. The leftward panels demonstrate ultrahigh-density activation mapping and the rightward panels a schematic view. There are simultaneous or "dual-loop" wavefronts around the right pulmonary veins and nearby surgical incision as well as the tricuspid valve annulus.

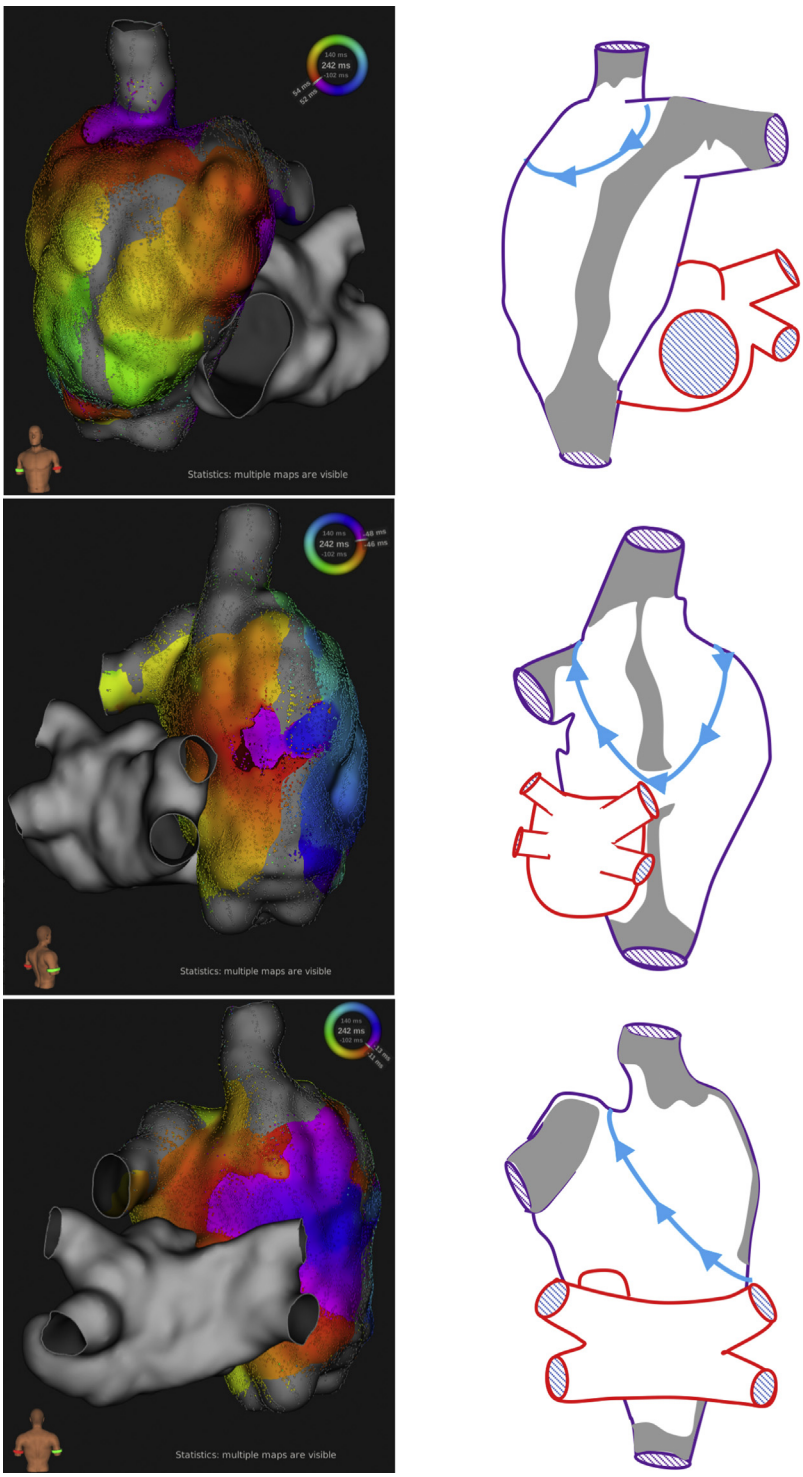

Fig. 7. Intra-atrial reentry after atriopulmonary Fontan operation (views identical to Fig. 6). The patient had previously undergone catheter ablation of periannular, septal, and caval circuits. There was a residual wavefront using scar on the posterior wall of the massively dilated morphologic right atrium that passed between the superior vena cava and atriopulmonary anastomosis.

limits of lesion delivery. Accordingly, recurrences and the onset of new arrhythmias are more common after catheter ablation in the setting of atriopulmonary Fontan surgery compared with other forms of ACHD.

In comparison with the atriopulmonary Fontan, IART is less frequently observed in patients with TCPC surgery,[3,56,57] with a recent multicenter study showing similar rates between those with lateral tunnel (LT) versus extracardiac conduits.[58] IART circuits after LT Fontan commonly involve reentry around the systemic AV annulus as well as atriotomy scar in the morphologic right atrial free wall (**Fig. 8**, Video 3).[59] Similarly, circuits around the systemic AV valve annulus are seen after extracardiac Fontan surgery but are much more challenging to reach from a conventional prograde catheter course (**Fig. 9**, Video 4).[60] Direct conduit puncture has been used to reach the pulmonary venous atrium after the extracardiac Fontan operation,[61] although access can be facilitated by puncture just below the conduit where cavoatrial overlap often develops (**Fig. 10**).[62] The retrograde approach using remote magnetic navigation is an alternative strategy that has been successfully used and may be the only viable option in some cases.[63]

Ebstein anomaly Ebstein anomaly of the tricuspid valve is associated with a 20% to 30% prevalence of congenital accessory pathways. This can result in reciprocating tachycardias and sudden cardiac death when characterized by the Wolff-Parkinson-White syndrome. Empiric electrophysiology studies have been shown to be of high yield when performed before tricuspid valve surgery (**Fig. 11**, Video 5).[64,65] Postsurgically, IART occurs frequently and may be seen after modified right atrial maze surgery, where atypical circuits may be more challenging to identify and ablate.[66] In addition, following annuloplasty rings or tricuspid valve replacement, the normally straightforward reentrant circuit around the tricuspid valve can be difficult to transect with catheter ablation, at times requiring innovative techniques (**Fig. 12**).[67]

Antiarrhythmic drug therapy

Antiarrhythmic drug therapy can be useful for control of recurrent IART in patients with ACHD. The most successful for maintenance of sinus rhythm are those with class III properties, owing to their efficacy for the prevention of reentry.[68–70] Catheter ablation is generally preferable to long-term antiarrhythmic drug therapy in patients with ACHD given their potential proarrhythmic side effects, limited effectiveness, and end-organ toxicities.[6]

Antitachycardia pacing

Antitachycardia pacing can result in acute termination of IART through antidromic penetration into the tachycardia circuit, thereby abolishing electrical propagation through wavefront collision. Although initial case reports were concerning for acceleration of the tachycardia and degeneration to malignant ventricular arrhythmia,[71] recent single-center studies have suggested both safety and efficacy of this approach for patients with ACHD.[72,73]

Of the available therapeutic strategies, catheter ablation has realized the most dramatic improvements over the past several decades. Although no randomized trials exist or are likely to take place, early and aggressive catheter ablation therapy is likely to be the optimal approach for most patients with ACHD at experienced centers.

ATRIAL FIBRILLATION

Atrial fibrillation (AF) has been increasingly recognized as being of major importance for the aging ACHD population, with estimates that are 20 times higher than the age-matched population.[74] AF surpasses IART as the predominant atrial tachyarrhythmia after 50 years of age with progression to more persistent forms with time.[75] ACHD at high risk include single ventricle, left-sided obstructive lesions, and palliated CHD.[76] To date, catheter ablation as a definitive therapy for AF using radiofrequency or cryothermal energy is effective but suboptimal, with recurrences reported between 40% and 50% after 1 year at experienced centers.[77–79]

ARRHYTHMIA/SUDDEN CARDIAC DEATH

Ventricular Tachycardia

Sustained monomorphic VT can develop in the setting of various CHD lesions and may result in hemodynamic instability or sudden cardiac death (SCD). Although tetralogy of Fallot is best characterized, VT after multiple forms of ACHD has been described. The VT mechanism after surgical repair is most often reentry (70%–80%).[80,81] Interestingly, unoperated patients with Ebstein anomaly are also susceptible to reentrant VT using intrinsic scar within the atrialized portion of the right ventricle.[82]

After tetralogy of Fallot surgery, postoperative scar predisposes to reentrant VT through a limited number of anatomic isthmuses.[83,84] Conduction slowing has been shown to be a

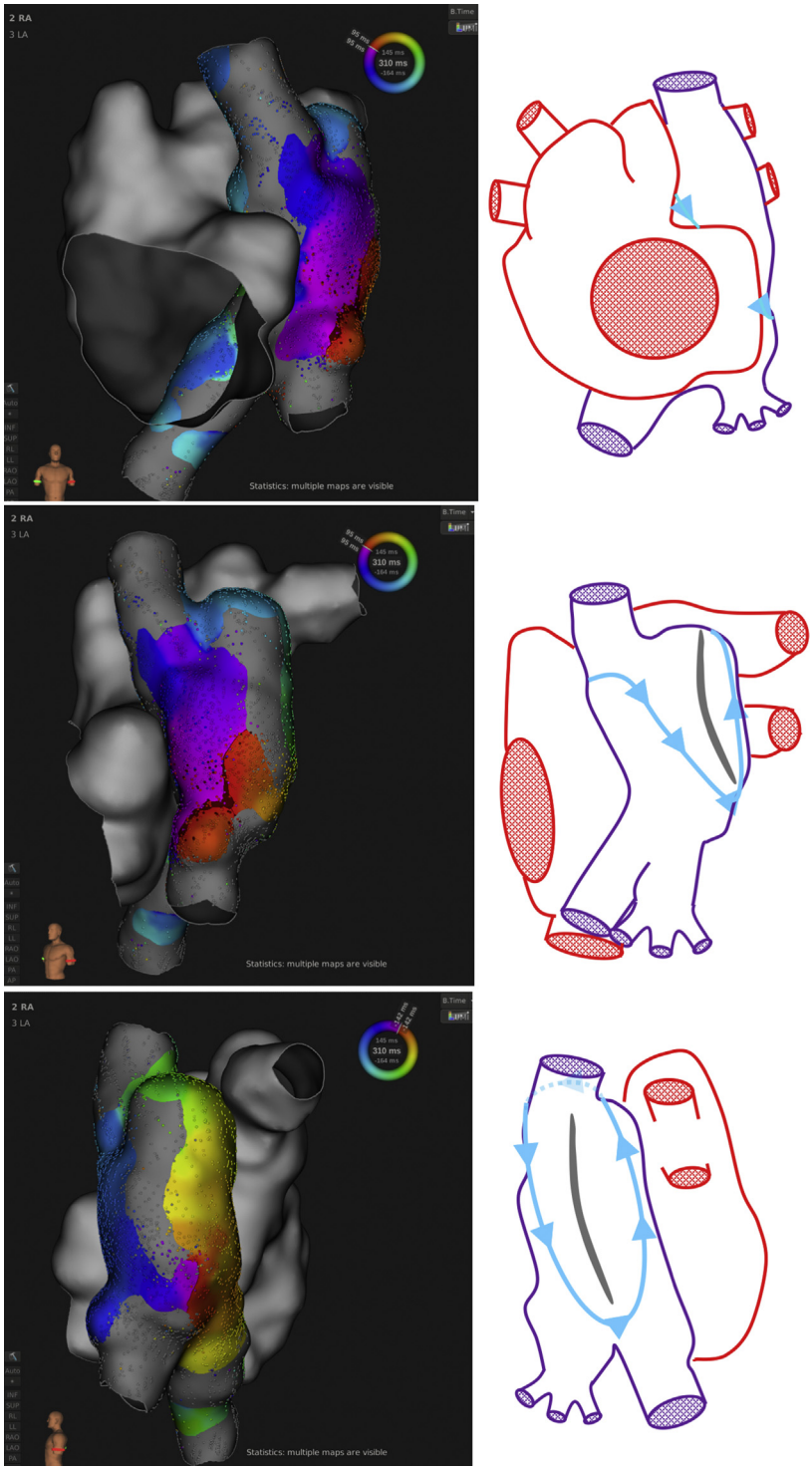

Fig. 8. Intra-atrial reentry after lateral tunnel Fontan in situs inversus. A wavefront using both portions of the morphologic right atrium is depicted. Catheter ablation at a narrow channel between an atriotomy and the inferior vena cava eliminated the tachycardia.

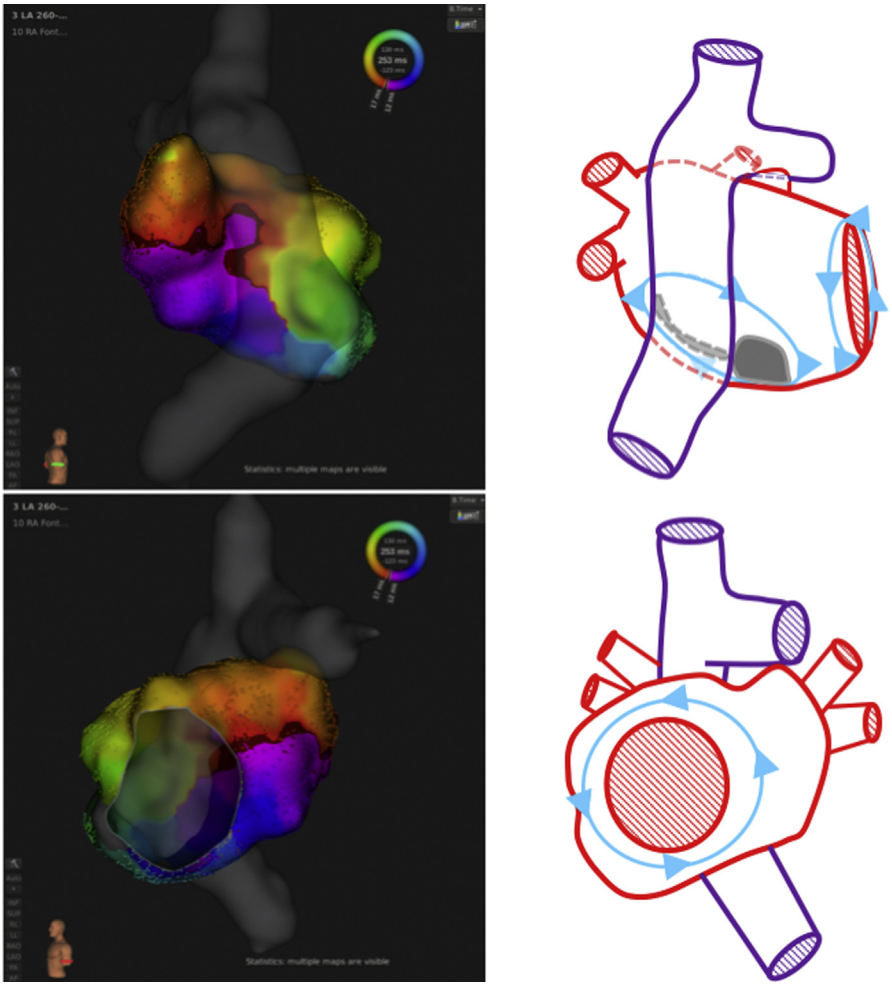

Fig. 9. Mechanism of intra-atrial reentrant tachycardia after the extracardiac Fontan operation. An atriotomy was found adjacent to the surgically placed GoreTex tube that extended down to the remnant of the oversewn inferior vena cava. A "dual-loop" tachycardia was active around the free wall atriotomy and the tricuspid annulus.

powerful predictor of inducible and/or clinical VT, with conduction velocities of less than 0.5 m/s serving as a reliable discriminator.[85] Catheter ablation is useful for elimination of reentrant VT with a low risk for recurrence when conduction block across the lesion set is confirmed.[86] Surgical cryoablation is often combined with pulmonary valve replacement for patients with clinical VT.[87] Novel applications, such as preoperative 3-dimensional printing of the ventricular anatomy with labeling of endocardial scar, may be useful for more focused surgical VT ablation (Fig. 13).

Sudden Cardiac Death

Mortality in CHD has experienced a shift from childhood to adulthood over the past several decades,[88] of which SCD is a major contributor. SCD is the mode of death in approximately 20% to 30% of ACHD mortalities, surpassed only by heart failure.[89–93] The overall risk of SCD in ACHD is between 25 and 100 times higher than that of the general population of comparable age.[94] This risk is progressive with time and is strongly associated with congenital lesion complexity.[92] The mechanism of SCD is VT/ventricular fibrillation in 80% of ACHD patients with approximately 70% of events occurring at rest (only 10% during exercise).[90,95]

Factors that have been associated with SCD among ACHD patients include increased QRS duration and fragmentation, conduction disturbances and delayed repolarization, pulmonary hypertension, supraventricular tachycardia, and

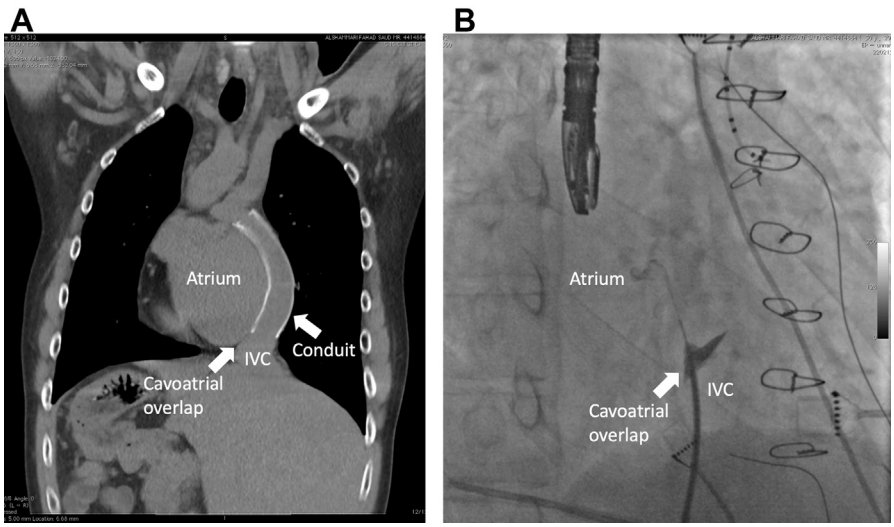

Fig. 10. Transcaval puncture for access to the pulmonary venous atrium after extracardiac Fontan for recurrent intra-atrial reentrant tachycardia. (*A*) Preoperative computed tomography angiogram demonstrating a region of overlap between the inferior vena cava and the pulmonary venous atrium. (*B*) Fluoroscopy demonstrates contrast in the region of overlap. Contrast is injected through the needle tip into the pulmonary venous atrium before the sheath and dilator are advanced.

impaired ventricular systolic function.[89,95,96] In particular, progression of QRS duration, QT dispersion, and ventricular dysfunction are highly predictive of SCD events.[97] Nonsustained VT (NSVT) has not been shown to be predictive of SCD for the general ACHD population, in contradistinction to its predictive value for tetralogy of Fallot.[95,98,99]

The ACHD lesions at greatest risk for SCD include those with a systemic right ventricle, single ventricle, or cyanotic forms CHD that include Eisenmenger syndrome[91,94,100,101] and tetralogy of Fallot. More recently, postoperative Ebstein anomaly has been reported to be associated with a relatively high risk for SCD based on the experience of a single center that included nearly 1000 patients.[102] With the exception of tetralogy of Fallot, clinical risk scores for these lesions are limited.

Sudden death in tetralogy of Fallot was recognized as a consequence of ventricular arrhythmia as early as the mid 1970s. Many predictors have been identified over the last several decades. These have included QRS duration, surgical technique and timing, indices of RV and LV systolic function, degree and complexity of ventricular rhythms as recorded by Holter monitoring, and, more recently, RV mass z score and LV ejection fraction.[103] Importantly, NSVT and increased LV end-diastolic pressure have been shown to be strong predictors of appropriate ICD shocks in a large population of patients with tetralogy of Fallot[99] and can be used in the calculation of baseline SCD risk. A Bayesian approach to risk stratification for primary-prevention ICD placement has been suggested for the tetralogy of Fallot population, for whom programmed ventricular stimulation is useful when the pretest probability of SCD lies between 1% and 11.5%.[104]

At present, expert consensus guidelines have been endorsed in the form of a joint statement from the Pediatric and Congenital Electrophysiology and Heart Rhythm Societies in 2014,[6] which were largely adopted by the European Society of Cardiology.[105] These guidelines provide a primary-prevention approach to the ACHD patient at risk for SCD, providing evidence-based recommendations for ICD placement. Unfortunately, current risk-stratification schemes are limited to a small subset of patients at elevated SCD risk.[106] An ongoing prospective study is under way to validate a proposed risk-stratification score that is based on clinical factors identified from the CONCOR registry among a diverse group of ACHD patients.[107]

Implantable Cardioverter-Defibrillators

To date, the only treatment strategy that has been shown to effectively prevent SCD in ACHD population remains the implantable cardioverter-defibrillator (ICD). In general, ACHD patients experience a relatively high proportion of both appropriate and inappropriate ICD shocks in

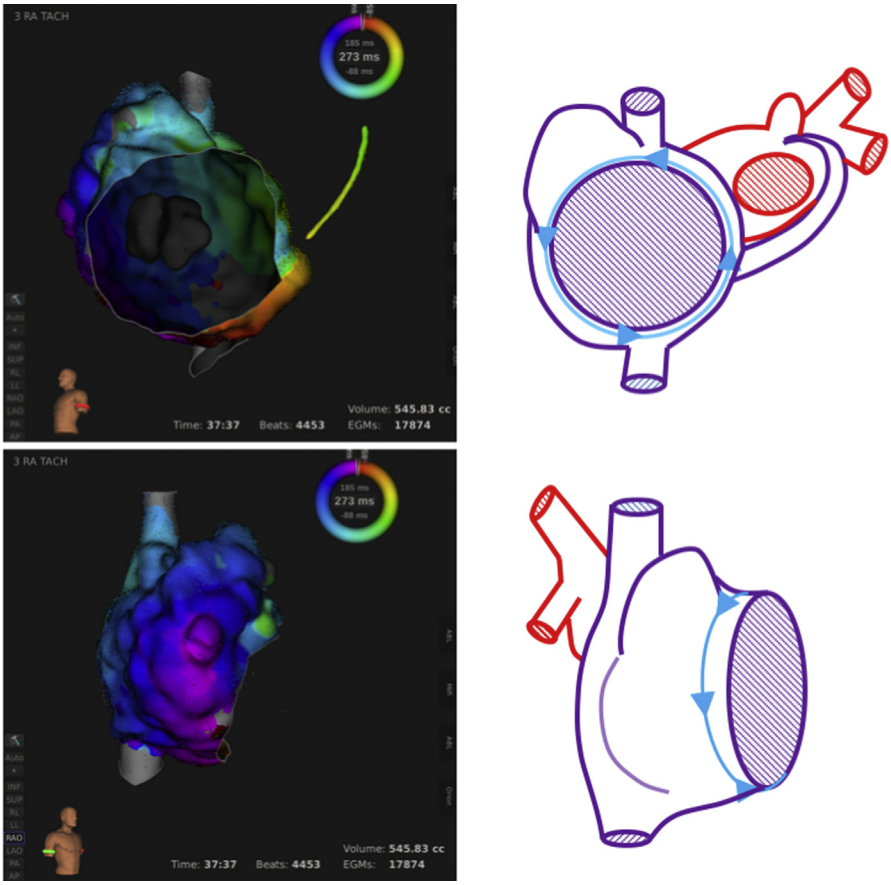

Fig. 11. Preoperative mapping of intra-atrial reentry before surgical valve repair in a patient with Ebstein anomaly. Massive right atrial enlargement is present with perpetuation of counterclockwise reentry around the tricuspid annulus owing to long conduction course rather than discrete slowing.

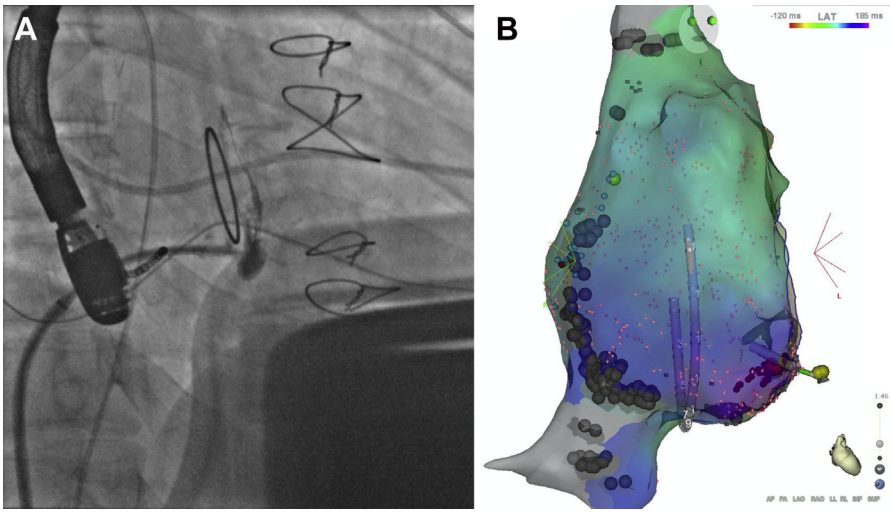

Fig. 12. Subvalvular catheter ablation for a patient with Ebstein anomaly who had previously undergone surgical tricuspid valve replacement. Recurrent intra-atrial reentrant tachycardia that was resistant to catheter ablation in the region of the tricuspid valve. (*A*) Needle puncture below the prosthetic valve. (*B*) Location of successful subvalvular catheter ablation on the 3-dimensional electroanatomic map.

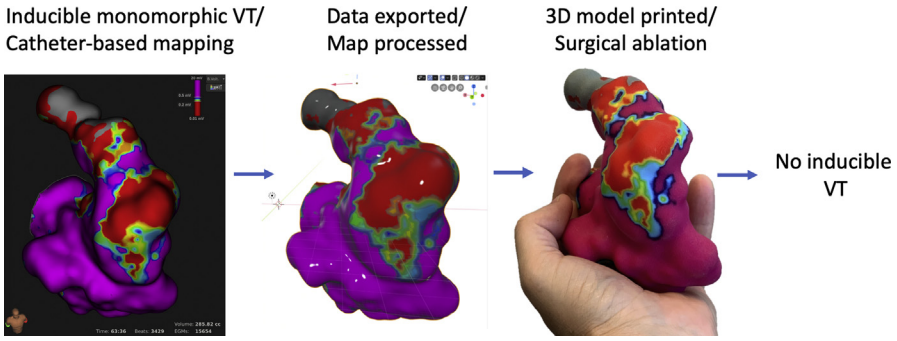

Fig. 13. Example of 3-dimensional (3D) printing for operative planning of tetralogy of Fallot. The procedural 3D map of a patient with clinical sustained monomorphic ventricular tachycardia (VT) with pulmonary valve dysfunction is shown. The 3D print was created to assist with concomitant surgical cryoablation of the RV free wall, after which VT was no longer inducible.

comparison with patients with acquired forms of heart disease.[108] Conventional ICD placement involves a subcutaneous or submuscular pulse generator with intravascular leads for both detection of ventricular arrhythmia and delivery of high-voltage defibrillation shocks. Such ICD therapies are highly effective for termination of malignant ventricular arrhythmia in this population.

Importantly, transvenous ICD placement may not be possible for many forms of ACHD owing to unique anatomic constraints. Examples include superior baffle occlusion after the Mustard operation for DTGA, prior TCPC Fontan surgery, and significant right-to-left intracardiac shunting as observed in the Eisenmenger syndrome. For such patients, the subcutaneous ICD (SICD) may serve as an alternative option (**Fig. 14**).[109,110] The

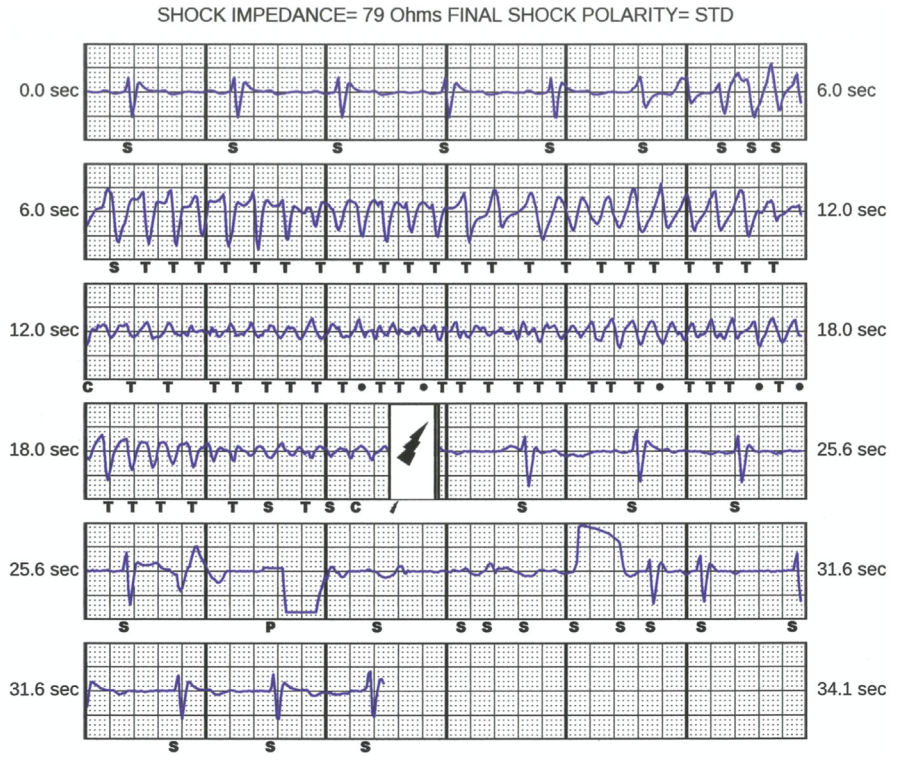

Fig. 14. Appropriate subcutaneous implantable cardioverter-defibrillator (SICD) shock in Eisenmenger syndrome. This patient had demonstrated recurrent episodes of both atrial and ventricular arrhythmia by an implantable loop recorder, prompting SICD placement. The successful shock occurred approximately 1 year after implantation.

preprocedure mandatory screening appears to be most favorable for Fontan patients and less for those with tetralogy of Fallot.[111,112] Importantly, the inability of the SICD to prevent bradyarrhythmia is a major limitation. Patients who would benefit from any form of ventricular pacing (eg, antitachycardia pacing, bradycardia pacing, or CRT) are generally not considered suitable candidates for SICD placement.

SUMMARY

Arrhythmia concerns abound in ACHD. Advances in technology and techniques have improved the therapeutic approaches available for this challenging population. As the congenital population continues to age, increased arrhythmia and ongoing advances in the specialty are to be expected.

DISCLOSURE

The author has nothing to disclose.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found online at https://doi.org/10.1016/j.ccl.2020.04.006.

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