Treatment of arrhythmia disorders in adults with congenital heart disease: A lesion-specific review

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ABSTRACT

There are now more adults living with a history of congenital heart disease than there are children. Modern electrophysiologists must familiarize themselves with the most common congenital lesions requiring electrophysiologic care as adults. Advancements in this field have been made most notably with high-resolution 3D imaging and electroanatomic mapping, left ventricular cannulation techniques, alternative pacing strategies, intracardiac echo, and transeptal access tools.

Key words: congenital heart disease, arrhythmia, ICD, ablation, pacemaker

INTRODUCTION

Adults living with a history of congenital heart disease (ACHD) comprise a growing proportion of patients seen in modern cardiac electrophysiology laboratories. This is largely due to improvements in surgical palliations during infancy, which have improved survival rates into adulthood to over 90% [1]. In Europe alone, there are estimated 1.8 million ACHD patients [2]. There are currently more adults living with congenital heart disease than there are children [3].

Rhythm disorders are common in ACHD and are accompanied by significant morbidity, mortality, and decreased quality of life. A detailed understanding of the diagnosis and management of arrhythmias in ACHD is paramount. The objective of this review is to provide an update on the electrophysiologic care of an adult with congenital heart disease according to the most encountered lesions. The review is divided into 3 sections: (1) implantable cardioverter-defibrillator (ICD) implantation and sudden death risk assessment; (2) pacing therapy, and (3) ablation of arrhythmias. Each section will include the specific and commonly encountered congenital lesion (Table 1).

ICD IMPLANTATION/SUDDEN DEATH RISK ASSESSMENT

Lesion: Tetralogy of Fallot

Sudden cardiac death (SCD) and ventricular arrhythmias are important long-term complications in tetralogy of Fallot (TOF) patients. The cumulative risk of sudden cardiac death is roughly 8% after 35 years after surgical repair,

Table 1. Relative importance of each EP therapeutic approach according to lesion type. The number of marks denote the relative prevalence/importance for each (blank = rare, X = infrequent, XXXX = very common)

	ICD /sudden death	Pacing	Ablation
Tetralogy of Fallot	XX		XXX
Atrial switch D-TGA	X	XXXX	XXX
Fontan	X	XX	XXXX
L-TGA	X	XXX	XX
Left-sided obstructive lesions	XXX		
Ebstein's		X	XXX
VSD or AVSD		X	XX

Abbreviations: AVSD, atrioventricular septal defect; D-TGA, D-transposition of the great arteries; L-TGA, levo-transposition of the great arteries; VSD, ventricular septal defect

with a latency period of around 10 to 20 years, suggestive of the role of scar and remodeling in the pathophysiology of ventricular tachycardia (VT) [4].

Pulmonary valve regurgitation is the most commonly associated hemodynamic lesion, and in a multicenter series was associated with all cases of sudden cardiac arrest (SCA) and 94% of cases of VT [4]. Prolonged QRS duration >150–180 ms is also a predictor of SCA [5]. Other risk factors include ventricular dysfunction (subaortic and subpulmonary), history of multiple cardiac surgeries, older age at repair, right ventricular hypertrophy and scarring, atrial tachyarrhythmias, and symptomatic nonsustained VT [6, 7].

In addition to risk factors based on history, programmed ventricular stimulation predicts VT and VF, with an adjusted relative risk of 4.7 [8]. In the most recent guidelines, electrophysiology studies (EPS) with ventricular stimulation are considered appropriate for symptomatic patients or those who have additional risk factors for VT mentioned earlier, such as LV dysfunction, QRS length >180 ms, extensive RV scarring, before or after pulmonary valve replacement, and in patients undergoing ablation for atrial tachyarrhythmias [2, 9]. Additionally, the conducting properties of anatomic isthmuses in monomorphic VT help estimate the risk of subsequent VT, discussed further in the ablation section. In our center, a diagnostic EPS is used for risk stratification on sudden death based on the point score outlined by Khairy et al. [9, 10].

Implantable cardioverter defibrillator (ICD) should be considered in patients in whom stable monomorphic VT can be induced and mapped on EPS, in those with documented sustained VT, and for secondary prevention due to aborted SCA. While efficacious against sudden cardiac death (SCD), ICD implantation is associated with a significant rate of complications, with 20.7% of patients having lead-related complications in a multicenter study [11]. Transvenous ICDs continue to be favored for this population due to the ability to pace terminate scar-based reentrant arrhythmias. However, the transvenous ICD has drawbacks, including the relatively young age of this patient group and concomitant tricuspid valve regurgitation which may be worsened by longstanding RV leads. One promising therapy for adults with TOF that requires ICD is the placement of a subcutaneous ICD that communicates with a transcatheter leadless pacemaker so that ATP can be delivered without the drawbacks of a traditional transvenous system. This technology is currently being evaluated as part of a clinical trial (Empower/Modular ATP, Boston Scientific Natick, MA). In many adults with TOF who have preexisting bioprosthetic or mechanical tricuspid valves, a "tricuspid-sparing" transvenous ICD system with pacing capability has been described [12] (Figure 1).

PULMONARY VALVE REPLACEMENT (PVR) AND TIMING OF EPS

Pulmonary regurgitation is strongly associated with SCA. However, PVR does not totally mitigate the risk of VT. In a retrospective study, VT incidence after PVR was estimated at 9.5% over 6.7 years [13]. Thus, strategies to further reduce VT risk after PVR were needed. A strategy of open and empirical intraoperative right ventricular outflow tract surgical cryoablation concomitant with PVR is effective [13].

However, a residual risk remains. In a report on 70 patients who underwent surgical cryoablation and postoperative EPS, 45% had residual inducible VT [14]. At our center, since management is guided by the results of

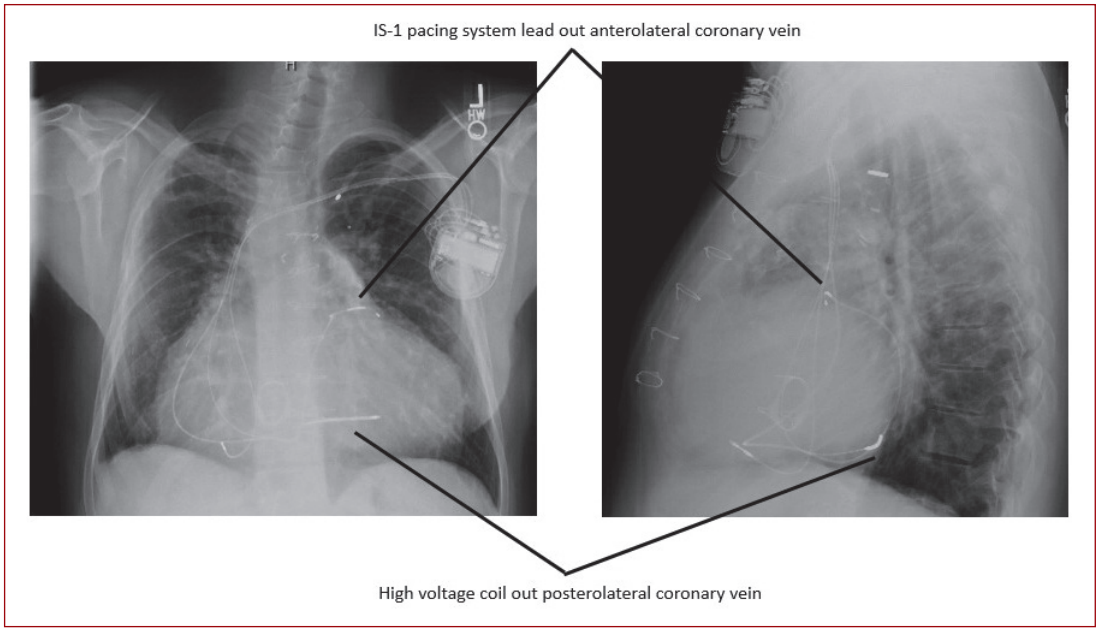

Figure 1. "Tricuspid sparing" transvenous implantable cardioverter-defibrillator (ICD) system implant for those in need of pacing, ICD, and mechanical or bioprosthetic tricuspid valves

postoperative EPS, we perform postoperative EPS only in this TOF patient subgroup.

Lesion: Atrial switch procedures for D-transposition of the great arteries (D-TGA)

SCD is responsible for around 13 to 45% of all-cause deaths in D-TGA and atrial switch [15]. In two comprehensive meta-analyses, the greatest predictor of SCD was a history of atrial tachyarrhythmias, increasing SCD 4 to 21-fold. Other risk factors included the Mustard procedure compared with Senning (OR, 2.9; 95% CI, 1.9–4.5), complex D-TGA compared with simple D-TGA (OR, 4.4; 95% CI, 2.2–8.8), and right ventricular (RV) dysfunction [15]. Interestingly, inducible VT does not appear to be predictive of SCD in D-TGA [16].

The risks associated with ICDs are significant in D-TGA. In our center, the rate of appropriate shocks was 11.5% over a median of 3 years. When compared to TOF patients, D-TGA patients had lower systemic ejection fraction (EF) but also had lower rates of appropriate ICD therapies. Therefore, ICD implantation based on left ventricular ejection fraction (LVEF) may not apply to this population compared to standard ischemic or nonischemic left ventricular heart failure patients [17]. Similar findings were seen in a multicenter cohort of atrial switch D-TGA patients after ICD implantation, with a rate of appropriate shocks in the primary prevention cohort of only 0.5% per year, and 6.6% for inappropriate shocks [16]. Thus, the risk-benefit ratio is slim in the primary prevention cohort, and somewhat stringent criteria are generally used for ICD implantation.

The 2020 European Society of Cardiology (ESC) guidelines for the management of ACHD give a class IIb recommendation for ICD implantation for patients with systemic RV dysfunction (systemic RVEF <35%) in the presence of additional risk factors including QRS >140 ms, severe AV valve regurgitation, nonsustained VT, and New York Heart Association (NYHA) symptom class II/III. Our approach is similar to the 2020 guidelines, but we also consider the history of atrial tachyarrhythmias.

Lesion: Fontan circulation

SCD is the second most common cause of death in Fontan patients, after advanced heart failure, but these sudden deaths may not solely be attributable to arrhythmia [18]. Tachyarrhythmia, thromboembolism, and protein-losing enteropathy are the strongest predictors of Fontan circulation failure and death [19].

In the 2018 European Heart Rhythm Association (EHRA) position paper, syncope in the setting of ventricular dysfunction, or LVEF <35% were factors to consider for ICD implantation [1]. Transvenous ICD placement is precluded in Fontan patients because the RV is inaccessible from the venous system. Subcutaneous ICD placement is effective for patients that do not require pacing. In a multicenter retrospective study, there was only one device-related complication, 21% of patients had inappropriate shocks, and 5% had appropriate shocks [20]. Given the high risk of positive pressure ventilation in this congenital lesion, subcutaneous ICD implants with moderate sedation only are favored over epicardial ICD lead placement in our center (Figure 2).

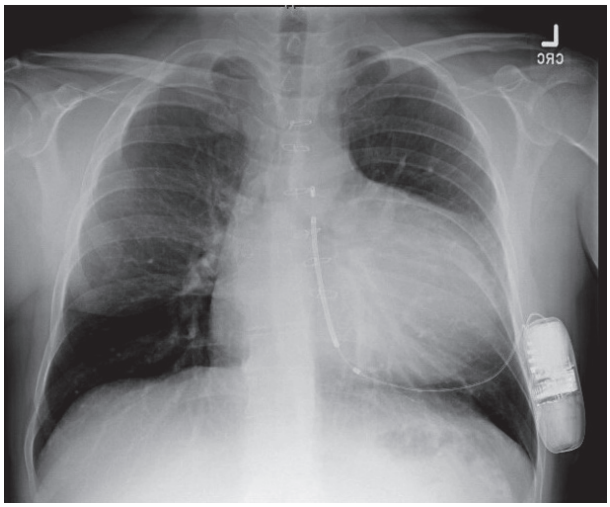

Figure 2. Subcutaneous ICD in a patient with single-ventricle physiology. Most patients with the Fontan circulation pass vector screening for this device.

Abbreviations: see Figure 1

Lesion: Levo-transposition of the great arteries (L-TGA)

The accumulated incidence of SCD in L-TGA is around 3% at age 40 [21]. Subaortic RV function is a potential predictor of SCD [22]. However, recent literature has been challenging this notion. In a retrospective cohort of over 3 500 patients, the rate of SCD was <1% per year, and none of the SCD patients had severely reduced RV function [23]. Nevertheless, MRI assessment of RV size and function (as opposed to echocardiographic) did strongly predict SCD [24].

The EHRA paper on arrhythmias in congenital heart disease uses low RVEF, complex ventricular arrhythmias, unexplained syncope, QRS duration >140 ms, or severe systemic AV valve regurgitation, as risk factors that warrant consideration for an ICD [1]. Generally, cardiac MRI should be performed for improved RV function quantitation. ICD implantation should be considered in patients with a history of syncope, nonsustained VT, or RV dysfunction in MRI.

Lesion: Left-sided obstructive lesions

LV obstructive lesions — especially aortic coarctation and Shone's complex — are associated with the highest risk of SCD due to VT and VF, and second in SCD after Eisenmenger's [25]. These patients are at risk of hemodynamic collapse due to low cardiac output during tachycardia caused by the fixed obstructive lesion and decreased preload during tachycardia, often in the setting of elevated left-sided and pulmonary pressures [26]. In the largest congenital heart disease database on arrhythmic SCD, LV

outflow obstructive lesions had the strongest association with arrhythmic SCD with an odds ratio of 10.7 [27]. While no specific guidance is given in the guidelines for this population, ICD should be strongly considered in patients with nonsustained VT, unexplained syncope, or LV dysfunction.

Lesion: Ebstein's anomaly

Ventricular arrhythmias, especially VT, can be common in Ebstein's. In a retrospective study with 79 patients with Ebstein's undergoing cardiac MRI, RV and LV dysfunction were predictors of SCD; after a median follow-up of 3 years, there were 5% of SCD and 3% of sustained VT [28]. VT usually arises from the congenitally abnormal muscle in the atrialized portion of the RV, can be focal or macroreentrant, and electrograms exhibit fractionated mid-diastolic waveforms [29]. VT ablation success rates appear to be around 90% [30].

Current guidelines give a class 1 indication for ICD implantation for patients with sustained VT/VF or SCA. No specific guidance is given for primary prevention [2]. An important consideration is the risk of worsening tricuspid regurgitation, even in patients with previous cone reconstruction. A tricuspid valve-sparing ICD implant technique has been described by our group as mentioned earlier [12].

PACING THERAPY

Lesion: Atrial switch procedures for D-TGA

D-TGA patients with a history of atrial switch surgery frequently have sinus node dysfunction, with a lifetime risk of at least 50% [31]. In the atrial switch anatomy, the systemic venous system ultimately drains to the left atrium. Thus, pacemaker leads are implanted in the subpulmonic left atrium and left ventricle via the baffles, however, certain precautions are necessary.

Active fixation leads to excitable left atrial tissue with aggressive challenging of the lead to ensure fixation is typical. The left atrial appendage is often unavoidable and is a stable site for atrial lead placement, but phrenic nerve capture, an issue uncommon in typical atrial lead implants, is a common obstacle and must be excluded. Steerable catheter delivery systems are sometimes useful. The sub-pulmonic LV lead goes through the mitral valve and tends to traject onto the left lateral wall. For small-French defibrillator leads, this has led to late perforation through the lateral wall [32]. It is therefore our practice to place these leads on the LV septum as guided by the standard left-anterior-oblique views which warrant artful stylet shaping (Figure 3).

ATRIAL PACING VS. DUAL-CHAMBER PACING

Subpulmonic LV pacing can impair systemic RV function, which is the most important cause of morbidity and mortality in atrial switch patients [33]. It is our practice to place single atrial lead systems in Mustard/Senning patients with isolated sinus node dysfunction. In patients with dual-chamber devices, DDD® with a long AV delay and low backup rate should be used. Nevertheless, due to long AV delays and increased total refractory period, the maximum tracking rate may be limited, which is an issue in a young population. Additionally, a single lead facilitates the extraction and baffle stenting strategy for baffle stenosis, avoiding a ventricular lead extraction and its potential complications.

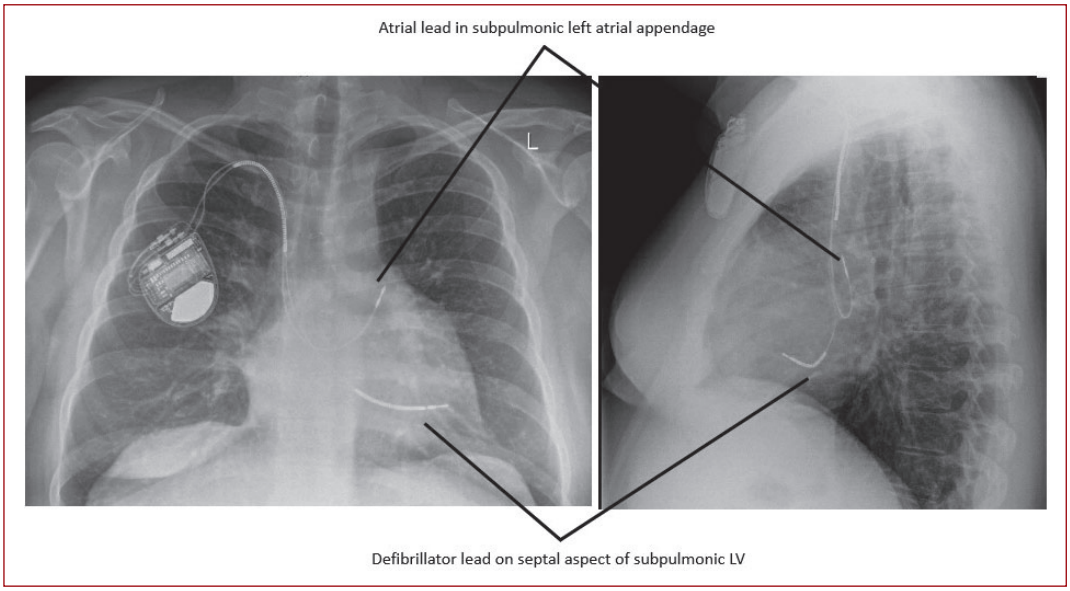

Figure 3. Dual chamber system in a patient with a Senning procedure for D-TGA. Non-standard stylet shaping is required to deliver the subpulmonic LV lead tip onto the septum and avoid the lateral free wall

Abbreviations: see Table 1

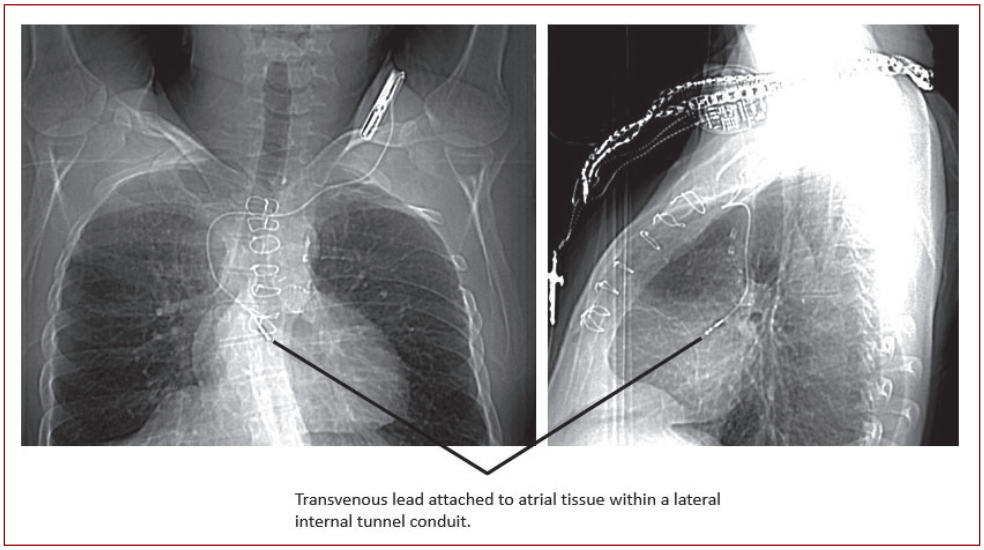

Figure 4. Atrial pacing system within a lateral tunnel Fontan. The lead is attached to a dual chamber impulse generator with a pin plug in the ventricular port. In this way, the device can be programmed DDD (dual chamber pacing) with minimal ventricular output, which allows automated antitachycardia pacing to be delivered regardless of intrinsic atrioventricular conduction

SPECIAL CONSIDERATIONS FOR PACING

Cardiac resynchronization therapy (CRT) is challenging in atrial switch D-TGA because of the inaccessibility of the coronary sinus ostium necessitating epicardial approaches. When needed, an anterior epicardial electrode placed on the subaortic RV requires relatively less dissection. This is then tunneled to the transvenous dual chamber system. Conduction system pacing is an interesting alternative, with only very limited evidence [34]. CRT in atrial switch D-TGA is an area of ongoing research and is still considered experimental in the 2020 EHRA guidelines [1].

THE NEED FOR SVC BAFFLE STENTING WITH PREEXISTING PACER (OUR EXTRACTION PROCESS)

Baffle obstruction and baffle leak affect up to 60% of atrial switch patients [35]. Percutaneous baffle stenting is a safe technique to treat these lesions. Concomitant device extraction, baffle stenting, and lead reimplantation (to avoid lead jailing) are commonly performed in our center and have also been described elsewhere as safe and effective [36]. This intervention typically occurs in a hybrid operating room with an on-call surgeon in case a sternotomy is required. Preprocedural imaging with a CT scan or MRI is performed to obtain the distance between the RV and chest wall, check the feasibility of venous access, assess atrial-baffle anatomy and lead position. Recently, transcatheter leadless pacing has been described in this patient population as well and could offer a leadless alternative [37].

Lesion: Fontan circulation

Half of adults with Fontan circulation will develop sinus node dysfunction, and even with modern surgical techniques (extracardiac conduit and lateral tunnel), the 10-year postoperative incidence was around 15% [38]. The purported causes are surgery near the cavoatrial junction in proximity to the sinus node, autonomic denervation, myocardial fibrosis, and injury to the sinus node artery. During junctional rhythm, there is systolic flow reversal from the left atrium to the Fontan chamber, and normal hemodynamics can be restored with atrial pacing and resumption of atrioventricular synchrony [39].

There are two options for implantation of pacemaker leads: surgical or transvenous. The right atrium is commonly fibrotic and thick, increasing the risk of transvenous lead failure. Nonetheless, due to the more invasive nature of epicardial systems, this is typically reserved for patients in need of ventricular pacing. In those cases, a surgical approach is preferred through a right thoracotomy. The transvenous approach is favored in adult patients with accessible atrial tissue (classic or lateral internal tunnel anatomy) (Figure 4).

Transvenous lead placement in extracardiac Fontan is more challenging because there is no direct communication between the venous system and the right atrium. A transpulmonary artery approach has been described with a puncture from the pulmonary artery to the pulmonary venous atrium, tunneling the lead to an infraclavicular pocket [40]. To avoid the potential thromboembolic complications of having a lead in the left atrium, a transvenous atrial epicardial approach can be performed. It involves accessing the venous system and advancing to the left pulmonary artery through the extracardiac Fontan. Then, the pulmonary artery is punctured inferiorly, and the lead tip is left in the intrapericardial space on the epicardial surface of the left atrium [41]. This technique is not routine and currently, most patients with an extracardiac Fontan circuit receive epicardial leads.

The role of antitachycardia pacing

Intra-atrial reentrant tachycardia (IART) is a major cause of morbidity and mortality in Fontan patients. In addition to catheter ablation, antitachycardia pacing (ATP) is a useful therapeutic tool. While ATP does not prevent the occurrence of SVT, it can terminate it, sparring the patient from the deleterious hemodynamic effect of prolonged SVT. In a large cohort, ATP was associated with lower risks of AT/AF events lasting ≥1 day (HR, 0.81), ≥7 days (HR, 0.64), and ≥30 days (HR, 0.56) [42]. In another study, ATP decreased the need for urgent direct current cardioversion and successfully converted 72% of IART episodes to normal sinus rhythm [43]. We recommend implanting ATP-capable devices even in patients receiving a single-lead pacemaker. This can be achieved by attaching a single atrial lead to a dual-chamber impulse generator. A pin plug is placed in the ventricular port. The device is then programmed DDD with minimal RV output (which is going to the pin plug).

Anticoagulation

Thromboembolism is common in Fontan patients, and many, if not most patients, end up requiring anticoagulation. The 2014 PACE/HRS consensus document gave non-vitamin K antagonist oral anticoagulants (NOAC) a class III recommendation to prevent thromboembolism, and vitamin K antagonists (VKAs) were preferred [44]. However, there is no evidence that VKAs are superior to NOACs, and newer evidence has emerged showing excellent safety and efficacy with NOACs in this population [45]. In a retrospective cohort study, the annual risk of bleeding was 3.1% per patient per year, with 0.7% per patient per year thromboembolic events [46]. In our center, because of better adherence, NOACs are sometimes used in this patient population.

LESION: L-TGA

Risk of complete heart block and device type

Patients with L-TGA are at risk of high-grade AV block, with an incidence of 2% per year and a lifetime risk of at least 50% [47]. This contrasts with patients with atrial switch D-TGA, in whom sinus node dysfunction predominates. The etiology of AV node dysfunction is related to abnormal atrioventricular communication and malalignment. The AV node is displaced posteriorly and is frequently hypoplastic. There is an additional anterior AV node that usually sits below the right atrial appendage. This AV node then connects to a long penetrating bundle of His which meets the ventricular myocardium in the subpulmonic area. This long area of tenuous His bundle tissue with surrounding fibrosis is at risk of degeneration and consequent AV block. Due to this abnormality, surgical correction with atrial and arterial switch ("double switch") does not appear to reduce the risk of AV block [48].

Univentricular pacing is a strong predictor of RV worsening function (HR, 4.7; 95% CI, 1.1–20.6), and that effect is mitigated by CRT [33]. Consequently, the EHRA working group suggests CRT can be useful for patients with a systemic RV with an EF ≤35%, NYHA functional class II, ambulatory IV, and wide QRS complex ≥150 ms with complete right bundle branch block QRS morphology (spontaneous or paced). No specific pacing burden recommendations are noted, but given that most patients receiving a pacemaker in this population have persistent high-grade AV block and thus a high pacing burden is expected (>20%), we routinely implant CRT devices as a first choice in this population, as recommended in the 2021 ESC guidelines on cardiac pacing [49].

CORONARY SINUS (CS) LEAD PLACEMENT

Lead positioning for CRT can be challenging in L-TGA, and the best location (for transvenous or epicardial leads) is not well established. The systemic venous system drains to the right atrium, which drains to the morphologic left ventricle. The CS drains to the right atrium and follows the RV but can have aberrant anatomy. Due to this difficulty in the CS lead placement, early experience relied heavily on epicardial lead placement [50].

In a more recent retrospective study, 95% of L-TGA patients undergoing CRT had a successful CS lead implantation, but all patients had advanced cardiac imaging with CT or MRI before the procedure and intraprocedural ventricular activation mapping before lead implantation. Of 21 patients, 14 had standard posteroseptal ostium cannulation, 2 via the vein of Marshall, and 2 via the superior ectopic ostium [51]. Thus, most patients with L-TGA can receive effective CRT, but significant preprocedural planning is required. Conduction system pacing should be considered for those who do not have an accessible CS (Figure 5).

LESION: EBSTEIN'S ANOMALY

RV resynchronization

Patients with Ebstein's anomaly are at increased risk of RV dysfunction and worsening of tricuspid regurgitation due to ventricular dyssynchrony with right bundle branch block (RBBB) (especially if QRS duration >150 ms). Accordingly, based on indirect data [52], current guidelines state "CRT may be considered for patients with a severe subpulmonary RV dysfunction and dilatation despite interventions to decrease RV volume overload, NYHA functional class II—ambulatory IV and wide QRS complex ≥150 ms due to a complete right bundle branch block" [1].

RV-CRT is performed by atrial-synchronized RV free wall pacing. The pacing location must be chosen by mapping late RV activation similar to standard CRT. RV function, contraction efficiency, and volumes show substantial acute short-term improvement with strategy [53].

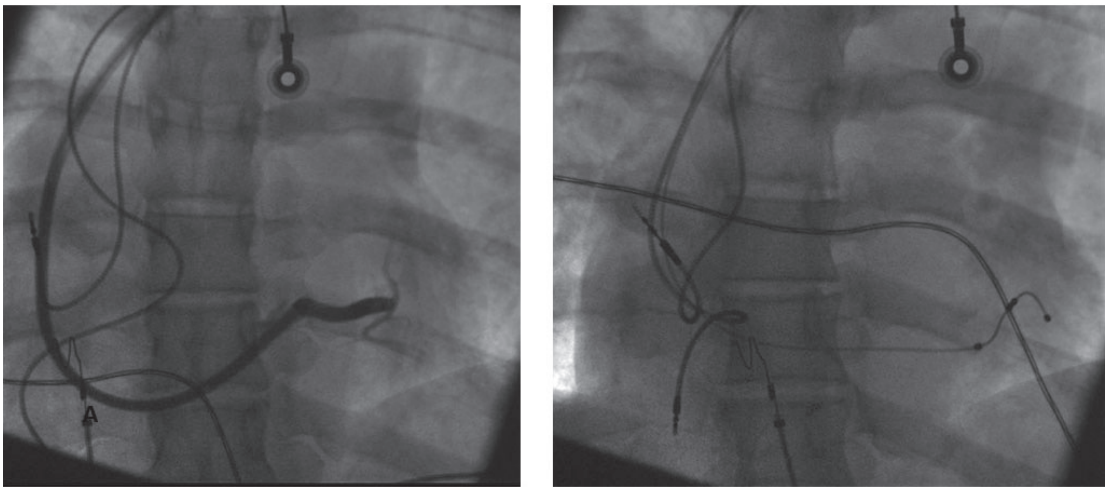

Figure 5. Anteroposterior venogram (left) of the coronary venous system in a patient with L- TGA and mesocardia. While the coronary sinus system usually follows the atrial situs, which in this lesion would be normally oriented, atretic venous systems or unusual origins of the CS ostium are not uncommon in L-TGA. Final CRT-P lead position (right)

Abbreviations: see Table 1

LOCATION OF CS OSTIUM AFTER TVR

The CS may be aberrant after tricuspid reconstruction. During cone surgery, surgeons sometimes perform an inferior annuloplasty band, anchored with a suture in the CS [54]. In other situations, such as tricuspid valve replacement, the CS ostium may end up inferior to the prosthetic valve [55]. Thus, some patients require an epicardial LV lead [56].

LESION: ASD, AVSD REPAIRS

Patients with repaired atrial septal defect (ASD) and atrioventricular septal defect (AVSD) are at increased risk of sinus node dysfunction and heart block, and pacemaker implantation is thus common [57, 58]. CS anatomy is abnormal, and it is frequently absent or with atretic ostia [59].

Thus, CRT, when necessary, is sometimes performed with an epicardial LV lead. Inadvertent placement of leads in the left heart can occur due to gaps in patch material, and care must be taken during device implantation [60].

ABLATION OF ARRHYTHMIAS

Lesion: Tetralogy of Fallot and RV outflow obstructive lesions

VT in TOF is usually macroreentrant and located in the subendocardium of the right ventricular outflow tract obstruction (RVOT). VT induction with entrainment can be poorly tolerated, and substrate mapping techniques in sinus rhythm are preferred in TOF. In 2007, Zeppenfeld described 4 anatomic RV isthmuses in TOF around the area of surgical repair [61]. They were located between the tricuspid annulus and anterior RVOT repair, the pulmonary annulus and RVOT free wall, the pulmonary annulus and septal patch, and the septum repair and tricuspid annulus. In a 2017 follow-up report, 74 patients underwent substrate mapping regardless of their history of VT. Anatomical isthmuses in patients with VT were longer and had slower conduction [61].

Technical aspects of VT ablation

VT ablation can be performed in sinus rhythm using standard 3D mapping, similar to ischemic VT. The anatomic isthmuses above are sought. Steerable sheaths, intracardiac echocardiography, and sheath-in-sheath techniques are often required to navigate the right ventricle.

Lesion: Atrial switch procedures for D-TGA

Atrial arrhythmias are common in atrial switch D-TGA patients. They are also a prominent cause of hospitalization, corresponding to roughly a third of cardiovascular hospitalizations in atrial switch D-TGA [62]. Atrial tachyarrhythmias with rates of 150 to 250 bpm can lead to rapid ventricular node conduction (due to 1:1 AV node conduction) and SCD.

IART is the most common (61.6%), followed by atrial fibrillation (AF) (28.8%), and focal atrial tachycardia (9.5%) [63]. AF is associated with older age and is the most common atrial arrhythmia in ACHD patients over 50 [63]. Many patients present initially with IART and develop AF over time. IART occurs due to macroscopic circuits which have been delineated by electroanatomic mapping and are usually cavotricuspid isthmus (CTI) dependent circuit [64]. In our center, the most frequent arrhythmia is subaortic tricuspid isthmus flutter. The reentrant circuits stem from atrial fibrosis histologically, which was correlated to prolonged right atrial volume overload [65] (9.5%) [63].

Catheter ablation

In atrial switch patients, the CTI sits on both sides of the baffle. Thus, CTI ablation requires biatrial access, crossing the surgical suture line. In our center, the most common approach to access the pulmonary venous atrium for a CTI

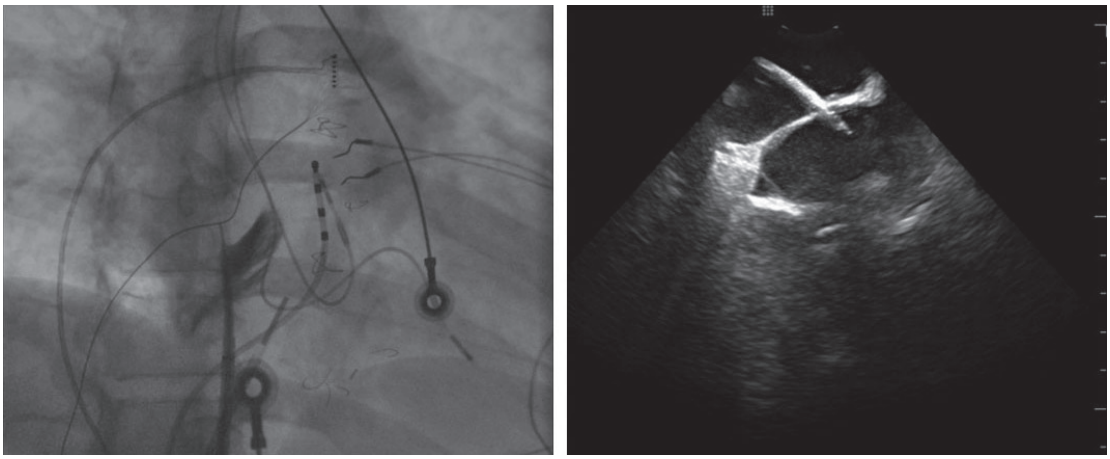

Figure 6. Trans-baffle puncture using fluoroscopy, a mechanical needle, and contrast in D-TGA atrial switch (left), and intracardiac echocardiography with an energized needle in lateral tunnel Fontan (right). Despite theoretical shortcomings of an energized needle in non-biologic conduit material, we have found success with this tool in most trans-baffle punctures

Abbreviations: see Table 1

ablation is the transbaffle puncture. Standard approaches, such as retrograde aortic, offer limited efficacy of around 70% and cause poor stability of the ablation catheter and valvular injury [66, 67].

After transbaffle puncture, when electroanatomic activation and entrainment mapping confirm IART, this can be followed by radiofrequency by the CTI and the pulmonary venous atrium (PVA) posterolateral scar [68]. The use of radiofrequency needles and/or wires has greatly assisted this technique (Figure 6). Predictors of recurrences are non-CTI locations, long PR intervals, and previous or induced AF [69]. The most best long-term predictor of freedom from recurrence is acute procedural success [70].

Lesion: Fontan circulation

IART is the most common arrhythmia in Fontan patients, and atriopulmonary connection is a risk factor compared to modern techniques such as the lateral tunnel or extracardiac Fontan's. Fontan conversion by converting those with an atriopulmonary to an extracardiac Fontan circuit, with an intraoperative maze procedure, had a freedom from recurrence of 51% [71]. Thus, in patients with atriopulmonary connections, this may be the initial management strategy.

Approach to access for IART ablation according to Fontan type (extracardiac, internal tunnel, atriopulmonary)

Access to the pulmonary venous atrium for catheter ablation in atriopulmonary Fontan is more straightforward due to access from the venous system. IART is the most common mechanism, and most patients have multiple circuits. In a retrospective study, the most common locations of critical isthmuses were lateral, inferolateral, posterolateral, or septal systemic venous atrium, and only 10% in the pulmonary venous atrium [72].

The modern total extracardiac conduit decreases the incidence of IART because it avoids pressure and volume overload to the pulmonary venous atrium. However, catheter ablation in extracardiac Fontan patients is challenging due to no readily available connection between the venous system and the right atrium. Access is achieved by identifying a large (>14 mm) cavoatrial overlap region through advanced cardiac imaging. In most patients, a cavoatrial overlap can be identified. In a retrospective study of 17 patients, 14 had an identifiable overlap [73].

The lateral tunnel Fontan is an intermediate between total extracardiac Fontan and atriopulmonary connection in terms of ease of access. A transbaffle approach is common or through a pre-existing fenestration [74]. In our cohort, the internal tunnel anatomy is the most common Fontan circulation with recurrent atrial flutters. A large amount of suture line along each side of the baffle may account for the numerous and difficult circuits these patients may encounter in adulthood.

LESION: L-TGA

Monckeberg sling and very difficult AVNRTs

The AV conduction system in L-TGA is aberrant. The "regular" AV node is frequently hypoplastic, and the anterior AV node is in the area of fibrous continuity between pulmonary and mitral valves [75]. Communication of the anterior and posterior AV nodes causes dual-node AVNRT. This was initially described by Monckeberg in 1913 in a patient with double-outlet RV, and by Uher in L-TGA, with a "sling" between the AV nodes. Both AV nodes can have one or more atrionodal connections in addition to the Monckeberg sling. Thus, reentry can occur by one of many permutations of retrograde and antegrade connections between the AV nodes and the atria, or antegrade and retrograde down each AV node's His bundle causing a reciprocating tachycardia.

For ablation, the slow pathway is targeted. If one confirms a reciprocating tachycardia involving both AV nodes

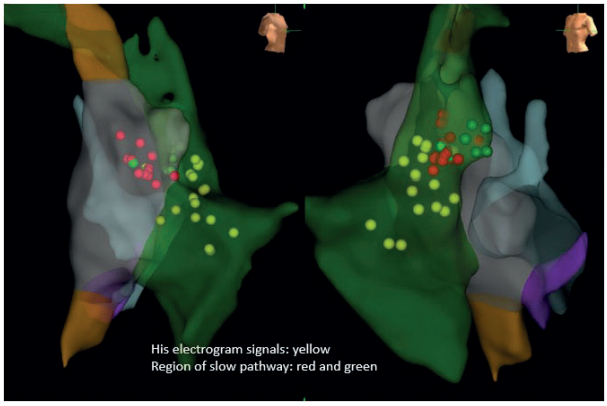

Figure 7. Ablation of atrioventricular nodal reciprocating tachycardia (AVNRT) in L-TGA. The reentrant course in this congenital lesion can be very challenging, requiring left-sided and pulmonary arterial mapping. Regions of the slow pathway can be anterior and require creative approaches to ablation

Abbreviations: see Table 1

and His bundles, one of the 2 AV nodes would require ablation if the remaining node offered appropriate AV conduction. If other SVT mechanisms are excluded, and the point of earliest activation is on the septum behind the Tendon of Todaro, then, anatomically guided slow pathway ablation can be performed empirically [75]. In cases in which the anterior AV node is the culprit, ablation of the slow pathway of the anterior AV node can be performed through the pulmonic valve sinus [76].

In summary, AVNRT ablation in L-TGA can be extremely complex and carries a higher risk of iatrogenic AV block due to the anatomic changes intrinsic to L-TGA but can be performed safely in experienced centers (Figure 7).

Transseptal puncture and its unusual orientation

Contrast-enhanced computed tomography and 3-dimensional reconstruction of the atria are helpful for an atrial septal puncture in L-TGA [77]. Intracardiac echocardiography is extremely important for this procedure. The best area for a transseptal puncture, also described in percutaneous valve edge-to-edge repair of the tricuspid valve in L-TGA, is level with the midline of the tricuspid valve on fluoroscopy in the 20° right anterior oblique projection and at the inferior edge of the fossa ovalis on transesophageal echocardiography [78].

Lesion: Ebstein's anomaly and other TV lesions

Up to 30% of Ebstein's patients have accessory pathways (APs). They are usually located in the posterior and septal borders of the tricuspid valve where the valve leaflets are most abnormal [79, 80]. The absence of a RBBB in Ebstein's is usually proof of the presence of a right-sided accessory pathway [81]. In a retrospective cohort of Ebstein's anomaly patients undergoing EPS, there were 30 APs in 21 patients. Of the 30 APs, 26 were atrioventricular and 4 were "Mahaim" fibers. APs due to Mahaim fibers are characterized by decremental conduction, the absence of delta waves on the surface electrocardiography (ECG,) and no retrograde conduction.

The residual atrioventricular ridge between the true right atrium and the "atrialized" right ventricle is where most APs are located [82, 83]. The atrioventricular ridge complicates catheter ablation because it limits catheter tip steering. Recurrence rates continue to be high (20% to 40%) at experienced centers [84]. Ablation failures are likely related to fractionated low-amplitude ventricular electrograms recorded from atrialized RV, multiple accessory pathways, difficulty identifying the true AV groove, and inability to retroflex the catheter tip under TV leaflets to improve stability [29].

Intracardiac echocardiography or a right atrial angiography can be used to locate the "true" AV ridge. Long sheaths should be used in patients with enlarged RA and RV cavities [29]. The next step is pacing along the lateral RA wall until fiber conduction is located and a characteristic high-frequency waveform is noted [29]. Published success rates are between 80 and 100% [84]. Finally, an EPS for the localization and ablation of APs is recommended empirically for patients who will undergo cone reconstruction surgery due to the high prevalence of APs and the technical difficulty in ablation afterward [85].

LESION: ASD, AVSD REPAIRS

The long-term risk of atrial arrhythmias is estimated to be around 25% in AVSD patients [58]. Most patients younger than 40 have IART, which is surpassed by AF at the age of 45 years. ASD is more common than AVSD. The left-to-right-shunt causes RA enlargement, and the duration of RA overload correlates better with atrial arrhythmias than age. Atrial arrhythmias usually occur due to macroreentry on surgical repair sites, and possibly due to heterogeneity of atrial conduction abnormalities in the RA and Bachman's bundle [86].

After repair, most patients with ASD will be considered to have mild ACHD. Younger adults, especially adults with mild ACHD have a higher burden of classical atherosclerotic risk factors such as obesity, hypertension, and dyslipidemia. Thus, strategies to prevent sudden death in this population include statins, smoking cessation, weight loss, and anti-hypertensive therapies, similar to the general population.

Transseptal puncture through surgical material

Ablation of left atrial arrhythmias including AF can be challenging in ASD and ventricular septal defect (VSD) patients because often the ASD patch repair must be punctured at the level of the original fossa ovalis. Transseptal access in these cases almost always involves radiofrequency [87]. Intracardiac or transesophageal ultrasound is essential.

SUMMARY

Modern electrophysiologists must familiarize themselves with the most common congenital lesions requiring electrophysiologic care in adults. The main challenges among this group remain to be the prevention of SCD, effective pacing, given access and anatomical difficulties, and ablation of arrhythmias in the context of abnormal anatomy and a large amount of arrhythmogenic substrate.

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