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REVIEW

Atrial Arrhythmias in Adults with Fontan Palliation

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ABSTRACT

Single ventricle physiology is a rare form of congenital heart disease and was, historically, a uniformly lethal condition. However, the atriopulmonary Fontan operation, and its successive iterations, the lateral tunnel and extracardiac conduit Fontan, became the fundamental approach to treating single ventricle heart disease. Over time, dysrhythmias are some of the most common complications with Fontan physiology, compounding morbidity and mortality. Atrial arrhythmias are prevalent in the Fontan population and occur in about 15–60% of patients with Fontan palliation, increasingly with age. Diagnosing atrial arrhythmias in patients with Fontan palliation may be challenging because of low voltage amplitudes arising from myopathic atrial tissue making it difficult to clearly assess atrial depolarization on surface electrocardiograms (ECG), vague symptoms not suggestive of tachyarrhythmia, or atrial arrhythmia with ventricular rates below 100 beats per minute. Intra-atrial reentrant tachycardia (IART) is the most common type of supraventricular tachycardia in adults with Fontan palliation. Acute management of atrial arrhythmias in patients with Fontan palliation involves prompt assessment of a patient's hemodynamic stability, anticoagulation and thrombosis risk, systemic ventricular function, and risk of sedation or anesthesia if needed. Long-term management of atrial arrhythmias is often multifactorial and may include long-term anti-arrhythmic therapy, permanent pacing, and ablation. The best approach for the management of atrial arrhythmias in adults with Fontan palliation is patient-specific and involves collaboration between congenital electrophysiologists, adult congenital cardiologists, and the patient.

Keywords: Fontan; Atrial arrhythmias; Adult congenital heart disease

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Key Summary Points

Palliated single ventricle patients maintain a high risk of tachyarrhythmias throughout their lifetime

Most tachyarrhythmias in this population are atrial in origin and can be difficult to diagnose

Long-term management of atrial tachyarrhythmias in adults with Fontan palliation is often multifactorial and may include anti-arrhythmic medications, pacing, and ablation

The best approach for management of atrial arrhythmias in adults with Fontan palliation is a patient-specific strategy involving congenital electrophysiologists, adult congenital heart disease specialists (ACHD), and the patient

INTRODUCTION

Congenital heart disease (CHD) is the most common type of birth defect, accounting for approximately one-third of congenital malformations, and a worldwide birth prevalence of about 9.1 per 1000 live births [1]. However, CHD with single ventricle physiology is a rare defect, with an incidence of 4–8 per 10,000 live births [2]. Historically, single ventricle physiology was uniformly a lethal condition. In 1968, Francis Fontan pioneered the Fontan operation to palliate patients with tricuspid atresia, and this would go on to become the fundamental approach to treating single ventricle heart disease [3]. Depending on the underlying anatomy, the predominant ventricle could be either a morphologic left or right ventricle, or in rare cases indeterminate.

To support long-term survival in the single ventricle condition, the single ventricle invariably must be dedicated to the higher resistance systemic circulation, while a solution for providing a source of blood flow to the lower resistance pulmonary circulation must also be achieved. The Fontan procedure accomplishes these goals by connecting both systemic venae cavae to the pulmonary artery, bypassing the right ventricle, to effectively divert systemic venous return to the lungs without a subpulmonic pump. The pulmonary blood flow is therefore passive, largely driven by the impedance of the Fontan system, including central venous pressure (CVP) and pulmonary vascular resistance (PVR). The typical surgical approach for single ventricle physiology now involves a multistaged surgical palliation which ultimately culminates in the Fontan procedure, typically in early childhood.

The original Fontan procedure, now referred to as the "classic atriopulmonary connection" (or AP connection), involved a Glenn shunt connecting the superior vena cava (SVC) to right pulmonary artery (RPA), and a connection of the right atrium to the left pulmonary artery with the interposition of two valved homografts between the inferior vena cava (IVC) and the right atrium (RA) and the RA and the left pulmonary artery (LPA) (Fig. 1a). Several modifications have been subsequently introduced. The Fontan surgery evolved to bypassing the RA into a total cavopulmonary connection (TCPC), which was introduced in 1988 as the next iteration of the Fontan called the lateral tunnel (LT). The aim of the LT was to optimize the hydrodynamic flow of the IVC return by channeling it through an intra-atrial synthetic tunnel consisting of a prosthetic baffle incorporating the atrial wall traversing the RA into the pulmonary artery (PA) (Fig. 1b). In 1990, the most contemporary iteration of the Fontan, the extracardiac (EC) Fontan (Fig. 1c), was introduced. The EC Fontan reroutes systemic venous blood around the heart directly to the pulmonary artery by interposing a synthetic conduit connecting the IVC to the PA. One advantage of the EC Fontan is that it minimizes exposure of the RA to arrhythmogenic incisions and suture lines especially near the sinoatrial node [4]. This article is based on previously conducted studies and does not contain any new studies with human participants of animals performed by any of the authors.

The worldwide population of patients with Fontan circulation is estimated to be

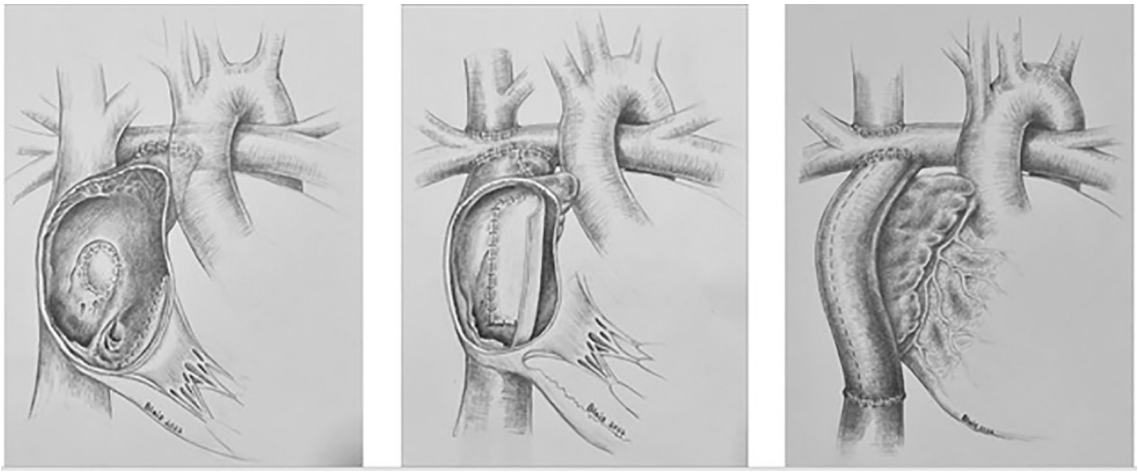


Fig. 1 Variations in Fontan surgery. **a** Classic atriopulmonary Fontan. **b** Lateral tunnel Fontan. **c** Extracardiac conduit Fontan

50,000–70,000 as of 2018 [5], and there are approximately 1200 Fontan procedures performed in the USA annually [6]. Survival has improved over the years and has been attributed to the refinement of the surgical techniques, with long-term follow-up studies of at least 20 years reporting survival rates from 61% up to 90% [7, 8]. Data reported from the US Agency for Healthcare Research and Quality estimated that in-hospital mortality for patients with Fontan palliation declined from 4.5% in 2001 to 1.1% in 2014 [9].

Many of the complications of Fontan physiology are indirectly or directly related to chronic elevation of CVP, rise in PVR, systemic ventricular function decline, pulmonary venous obstruction, and hepatic congestion and dysfunction [10]. The contributing hallmark of these complications is the absence of a subpulmonary ventricle and the obligate systemic venous hypertension in the Fontan circulation [11]. One of the more persistent and problematic complications following the Fontan procedure is arrhythmias. Historically, the main rationale for the development of the EC Fontan over the LT Fontan was to further reduce the development of arrhythmias as a postsurgical complication.

Despite the milieu of strategies to improve the surgical technique, arrhythmias, and particularly atrial tachyarrhythmias, remain a leading source of morbidity and mortality in patients with Fontan surgery [10, 12]. The broad catchment of dysrhythmias in patients with Fontan palliation includes sinus node dysfunction, predominant junctional rhythm, atrioventricular block (AVB), supraventricular and ventricular arrhythmias, and the risk of arrhythmic sudden cardiac death. Arrhythmias may reflect congenitally displaced or malformed sinus nodes, AV conduction systems, abnormal hemodynamics, primary myocardial disease, hypoxic tissue injury, residual or postoperative sequelae, and genetic influences [13].

The onset of tachyarrhythmias has been considered both a cause and a consequence of clinical decline [14]. Preoperative and intraoperative sinus rhythm has been associated with improved survival [7], and merely having a nonsinus rhythm, such as junctional rhythm or ectopic atrial bradycardia, has been associated with increased risk of atrial arrhythmias [15]. Over the long term, arrhythmias in patients with Fontan palliation have been associated with Fontan failure, all-cause mortality, and sudden cardiac death [10, 16, 17].

BURDEN OF ATRIAL ARRHYTHMIAS

Over the years, modifications to the Fontan procedure have resulted in a decreased incidence of sinus node dysfunction and atrial

arrhythmias [8, 18–21]. Possible factors to explain persistent arrhythmia complications after the EC Fontan include hypoxemia-related structural changes, autonomic dysregulation, arrhythmogenic nature of the atrial cuff of tissue at the IVC anastomosis, crista terminalis damage, and adrenergic nerve ending damage [22–29]. Fundamentally, the two driving factors for the development of supraventricular arrhythmias in patients with Fontan palliation include a circulation functioning with chronically raised right atrial pressures and atrial manipulation during surgery [30].

Along with improved early and long-term survival after Fontan palliation over time, arrhythmia occurrence has declined [8, 18, 19, 22, 31], but still remains a burden in all variants of the Fontan physiology. Contemporary estimates of arrhythmia burden as a late complication after the Fontan procedure range from 20% to 44% [7, 8]. The cumulative incidence of atrial arrhythmias after atriopulmonary Fontan surgery is reported to be $>$ 45–65% [8, 11, 32] and 14–25% in those with a total cavopulmonary connection [8, 16]. The 10-, 20-, and 30-year freedom from arrhythmias in long-term follow-up was found to be 71%, 42%, and 25%, respectively, which included atrial flutter in 74%, atrial fibrillation 39%, atrial tachycardia 26%, re-entrant supraventricular tachycardia 9%, and ventricular tachycardia in 10% [7].

Common forms of atrial tachyarrhythmias encountered in patients with Fontan palliation include intra-atrial reentrant tachycardia (IART), nonautomatic focal atrial tachycardia, and atrial fibrillation. IART is the most common atrial arrhythmia in patients with Fontan palliation, accounting for approximately 75% of supraventricular tachycardia, with focal atrial tachycardia in up to 15% of patients [33]. The 20-year freedom from IART, specifically, was 46% [12]. Atrial fibrillation is becoming increasingly common in adult patients with Fontan palliation, as well, with one study estimating an incidence of 14% over a 5-year period [33–35].

The arrhythmic substrate for the development of IART evolves from the atrial incisions, suture lines, right atrial dilation, and elevated right atrial pressure. Given these factors, the type of Fontan palliation affects the risk of developing atrial arrhythmias. The association between the AP Fontan and IART is well established, especially the length in which one survives with that type of surgery. In one study, the incidence of IART for the AP configuration at a 20-year follow-up was 39% compared to 9% in the LT Fontan, whereas importantly there was no difference between the LT and EC Fontan incidence [36]. Most atrial arrhythmias in patients with the AP Fontan palliation are found to be localized to the systemic venous atrium [37]. IART circuit is commonly around the IVC, specifically near the inferolateral atrium, but can be found near the lateral and septal walls as well [38, 39]. The location of IART within the EC Fontan configuration has been reported to involve the isthmus between the inferior atrioventricular valve annulus and the oversewn edge of the IVC, which is largely analogous to the cavotricuspid isthmus in the normal heart, with risk factors including either an atriotomy or fenestration placed in the lateral wall during a previous surgery [40].

RISK FACTORS FOR ATRIAL ARRHYTHMIAS AND ARRHYTHMIA CONSEQUENCES

In general, risk factors for early and late tachyarrhythmias include atrioventricular valve (AVV) regurgitation, preoperative tachyarrhythmias, AVV replacement [18], AP connection Fontan, preoperative bradycardia, lack of sinus rhythm, older age at operation with longer interstage interval, and heterotaxy syndrome [21, 41, 42]. Other risks have been related to the structural changes and functional deterioration of the Fontan circuit, such as increased right atrial pressures, right atrial dilation, ventricular impairment, and functional capacity [16, 43].

Arrythmias are one of the most frequent reasons for hospital admission and in-hospital mortality for patients with Fontan palliation [44]. Complications that arise from arrhythmias in patients with Fontan palliation include acute hemodynamic compromise, heart failure,

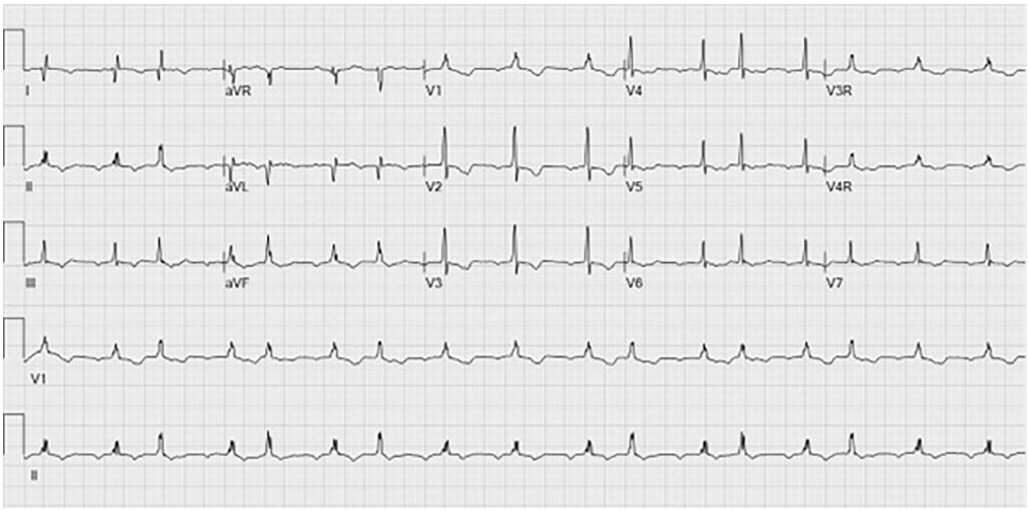


Fig. 2 ECG of IART in a 41-year-old woman with double outlet right ventricle (DORV) and hypoplastic RV with lateral tunnel Fontan. Classic saw-toothed flutter waves are not typically seen in patients with Fontan palliation with IART; rather atrial depolarizations are often low voltage as a result of atrial myopathy

thromboembolism, and sudden cardiac death [10, 45]. The development of significant arrhythmias in the AP Fontan, when coupled with signs of heart failure, is associated with a 3-year mortality rate of 25% [45]. The initial episode of sustained atrial tachycardia often heralds a pattern of progression toward more frequent and prolonged recurrences [33]. Atrial arrhythmias are poorly tolerated in Fontan physiology, as the passive transpulmonary perfusion, which is highly dependent on a low ventricular end-diastolic pressure (EDP), will then see increasing afterload in the context of atrial arrhythmias resulting in decreased cardiac output. Total cardiac output decreases by about 25% for patients with Fontan palliation that are in atrial fibrillation [46]. Clinically, the prognosis is worrisome, even if the patient can be converted quickly into sinus rhythm, which underscores the urgency to make an accurate diagnosis and expedite treatment of dysrhythmias in Fontan physiology.

DIAGNOSIS OF ATRIAL ARRHYTHMIAS AFTER FONTAN PALLIATION

Though IART is the most commonly occurring atrial arrhythmia in patients with Fontan palliation, classic saw-toothed flutter waves are not often seen in patients with Fontan palliation, largely as a result of atrial myopathy. Patients with Fontan palliation often have electrically diseased atrial tissue, and identification of atrial tachyarrhythmias can be challenging for several reasons: (1) atrial activity may not be clearly discernable on a standard electrocardiogram (ECG) (Fig. 2); (2) atrial tachycardia cycle length may be slow and variable, complicating the diagnosis; (3) underlying QRS abnormalities may cause the clinician to assume wide complex tachycardia is ventricular tachycardia. Further, patients may not have symptoms of palpitations; rather, symptoms due to ventricular dysfunction or Fontan failure may be the primary presentation of arrhythmia. Therefore, it is important to have a high suspicion for atrial arrhythmia in this population, especially as a patient ages. It is paramount in these patients to compare an ECG to previous baseline ECGs, as a change in baseline heart rates may be the most obvious clue that the patient is in an atrial arrhythmia (Fig. 3a, b). Diagnosis can also be challenging in patients with AVB who are ventricularly paced since there may be little or no change of the paced rhythm (depending on pacemaker programming and atrial tachycardia rate). AVB is uncommon but not rare, with an estimated

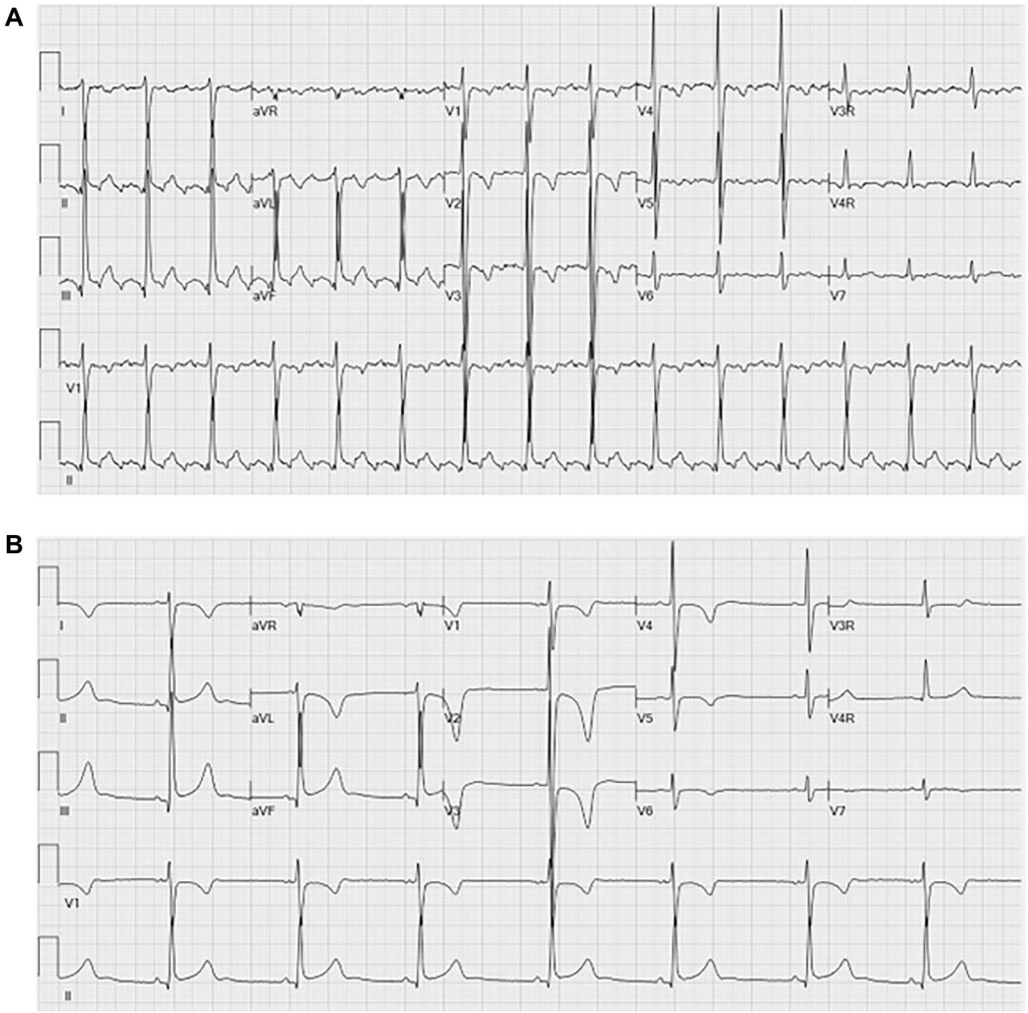


Fig. 3 **a** IART in a 19-year-old with lateral tunnel Fontan, ventricular rate 90 bpm. **b** Baseline sinus bradycardia in a 19-year-old with lateral tunnel Fontan

incidence following the Fontan operation as high as 3% [35], and pacemaker interrogation in this population is often necessary to confirm atrial arrhythmia (Fig. 4). Since the incidence of atrial tachyarrhythmias increases with age and is a major source of morbidity [16, 20], regular surveillance of rhythm with ECG and ambulatory Holter monitors is recommended during follow-up of both symptomatic (class I) and asymptomatic (class I and IIa) patients [13].

ACUTE MANAGEMENT OF ATRIAL ARRHYTHMIAS

Acute management of atrial tachyarrhythmias in adults with Fontan palliation varies on the basis of hemodynamic status. Atrial tachyarrhythmias with rapid ventricular conduction may quickly worsen hemodynamics in Fontan circulation and lead to heart failure over a short period of time. Rapid deterioration of hemodynamic status can occur from tachyarrhythmias in the Fontan circulation, leading to ventricular dysfunction, respiratory failure, and other end-organ failure, elevating the urgency to terminate an acute episode of a rapidly conducted

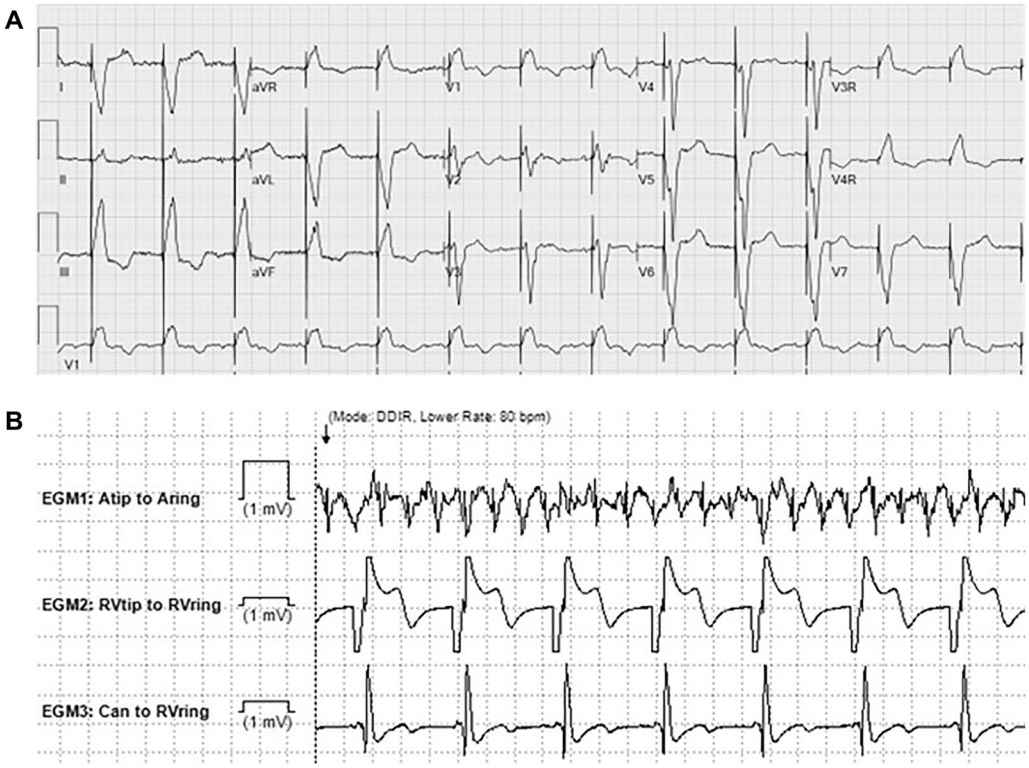


Fig. 4 **a** EKG of a 63-year-old man with lateral tunnel Fontan, to clinic with volume overload. Atrial activity not clearly discernable on ECG. **b** Electrograms from device interrogation demonstrate rhythm to be atrial fibrillation with ventricular pacing at 80 bpm

arrhythmia. However, many patients present with hemodynamically stable sustained atrial arrhythmia, and management should include consideration of anticoagulation and adult congenital electrophysiology consultation [47]. SVT that is AV node dependent may be terminated with adenosine; however, knowing the patient's ventricular function and underlying baseline rhythm would be important prior to conversion with medication or cardioversion. In the acute management of atrial tachyarrhythmias in patients with Fontan palliation cardioversion is often necessary, but one must consider cardiac position as well as baseline underlying rhythm in planning for cardioversion. Rate control may be a temporizing measure in an acute SVT episode, which may provide an opportunity to convert once the patient is stabilized. Sustained atrial arrhythmias are associated with significant thromboembolic risk; however, thrombosis risk in complex CHD, such as Fontan palliation is likely higher than other forms of congenital heart disease [34, 48, 49]. It is often necessary to rule out intracardiac thrombus and ensure anticoagulation prior to cardioversion, even in shorter duration of IART or AF. Thromboembolisms can involve the Fontan conduit itself, and can include pulmonary embolisms, intracardiac thrombi, embolic strokes, or systemic arterial embolisms.

Defibrillator pad positioning should be considered and may need to be altered from baseline as a result of dextroposition of the heart or enlarged atria. Sedation or anesthesia for cardioversion should be performed by someone experienced with Fontan physiology. Additionally, prior to sedation, one should review and consider the patient's baseline rhythm. A patient with severe sinus node disease at baseline may have a profound sinus pause after cardioversion, which may be even more prolonged by deep sedation. For those patients

Table 1 Consideration of management options for atrial arrhythmias in adults with Fontan palliation

Is the patient hemodynamically unstable in the atrial tachyarrhythmia?	Rapidly conducted atrial tachyarrhythmias may not be tolerated in Fontan physiology and emergent cardioversion may be necessary
What is the burden of arrhythmia?	A high burden of intermittent atrial arrhythmia may contribute to ventricular dysfunction and worsening physiology and may warrant more aggressive treatment
Is there an underlying hemodynamic abnormality contributing to the arrhythmia?	Hemodynamic assessment may be warranted in the setting of new or worsening atrial arrhythmias
What is the systolic function of the single ventricle?	Severe ventricular dysfunction may influence choice of anti-arrhythmic therapy and consideration for minimizing anesthetic time for procedures
Does the patient have underlying sinus node disease limiting anti-arrhythmic therapy?	Atrial pacing may be necessary prior to use of anti-arrhythmic therapy
Does the patient have AV node disease?	Need for ventricular pacing necessitates epicardial pacing system in most patients
What type of Fontan palliation does the patient have?	Access for ablation of arrhythmia substrate may be more challenging for those with extracardiac Fontan
What other anomalies does the patient have? (i.e., is the venous return normal?)	Does the patient have venous access to the Fontan baffle? Does the patient have native atrial tissue within the Fontan baffle? Does the patient have residual intracardiac shunt, prohibiting consideration of transvenous pacing lead?
Is the patient at increased risk of anesthesia because of severe ventricular dysfunction or other comorbidities?	Other comorbidities may increase anesthesia risk and may obviate the need for efficient procedures with minimal or no sedation

with severe sinus node disease, variations in sedation for cardioversion may be necessary.

LONG-TERM MANAGEMENT OF ATRIAL ARRHYTHMIAS

Long-term management of atrial tachyarrhythmias in adults with Fontan palliation is often multifactorial, and may include anti-arrhythmic therapy, pacing, and ablation procedure. Consideration of treatment options for management of atrial arrhythmias is based on several factors and warrants discussion with patient and medical team, including adult congenital cardiologist and congenital electrophysiologist (Table 1). Consultation with an adult congenital cardiologist is important in the consideration of treatment of atrial arrhythmias in adults with Fontan palliation, and all patients should be followed by an adult congenital specialist. Sustained atrial arrhythmia should prompt consideration for invasive hemodynamic assessment, as new or worsening arrhythmias may be driven by hemodynamic changes that are difficult to detect non-invasively [47, 50]. In patients with Fontan palliation, long-term management of atrial arrhythmias is aimed at rhythm control to maintain sinus or atrial paced rhythm, not simply rate control. It is well accepted that AV synchrony is important in those patients with single ventricle physiology. However, when

treatment strategies for atrial arrhythmias fail, long-term rate control and anticoagulation may be the only option for some patients with Fontan palliation.

Anti-arrhythmic Therapy

Anti-arrhythmic therapy is often first-line therapy for treatment of atrial arrhythmias, though long-term medical management is not often successful. Additionally, tolerance of anti-arrhythmic therapy may be limited by sinus node disease. For example, an adult patient with Fontan and baseline sinus node disease whose resting heart rate is 45–50 beats per minute and who is in junctional rhythm is not likely to tolerate the addition of any anti-arrhythmic therapy. Additionally, further slowing of sinus rate with anti-arrhythmic therapy may become proarrhythmic. In selecting the class of anti-arrhythmic therapy, one must consider sinus or AV node disease, ventricular function, and other comorbidities. Choice of anti-arrhythmic agent in adults with congenital heart disease is based on complexity of congenital heart disease and ventricular function. Class I anti-arrhythmic drugs are generally avoided for complex congenital heart disease due to ventricular dysfunction, incisional scars, and myocardial fibrosis, which may be more arrhythmogenic. In patients with Fontan palliation, class III agents (amiodarone, dofetilide, or sotalol) are recommended for medical management of IART or AF; however, in the setting of ventricular dysfunction, amiodarone or dofetilide is preferred [13]. Amiodarone may be effective in preventing arrhythmia recurrence, though its known side effects limit long-term use for maintenance of sinus rhythm. Thyrotoxicosis, for example, occurs in almost half of patients with Fontan palliation on long-term amiodarone [51]. Both hepatopathy and chronic renal disease seen as sequelae of Fontan circulation complicate the potential use of many anti-arrhythmic medications [52, 53].

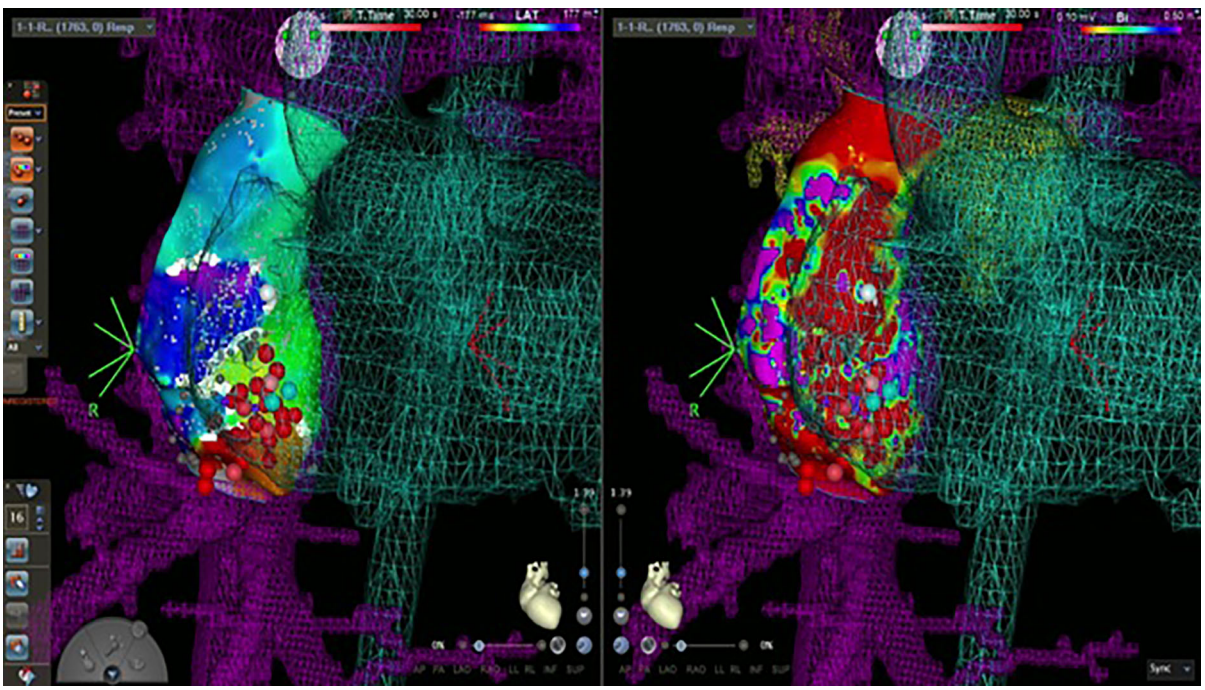
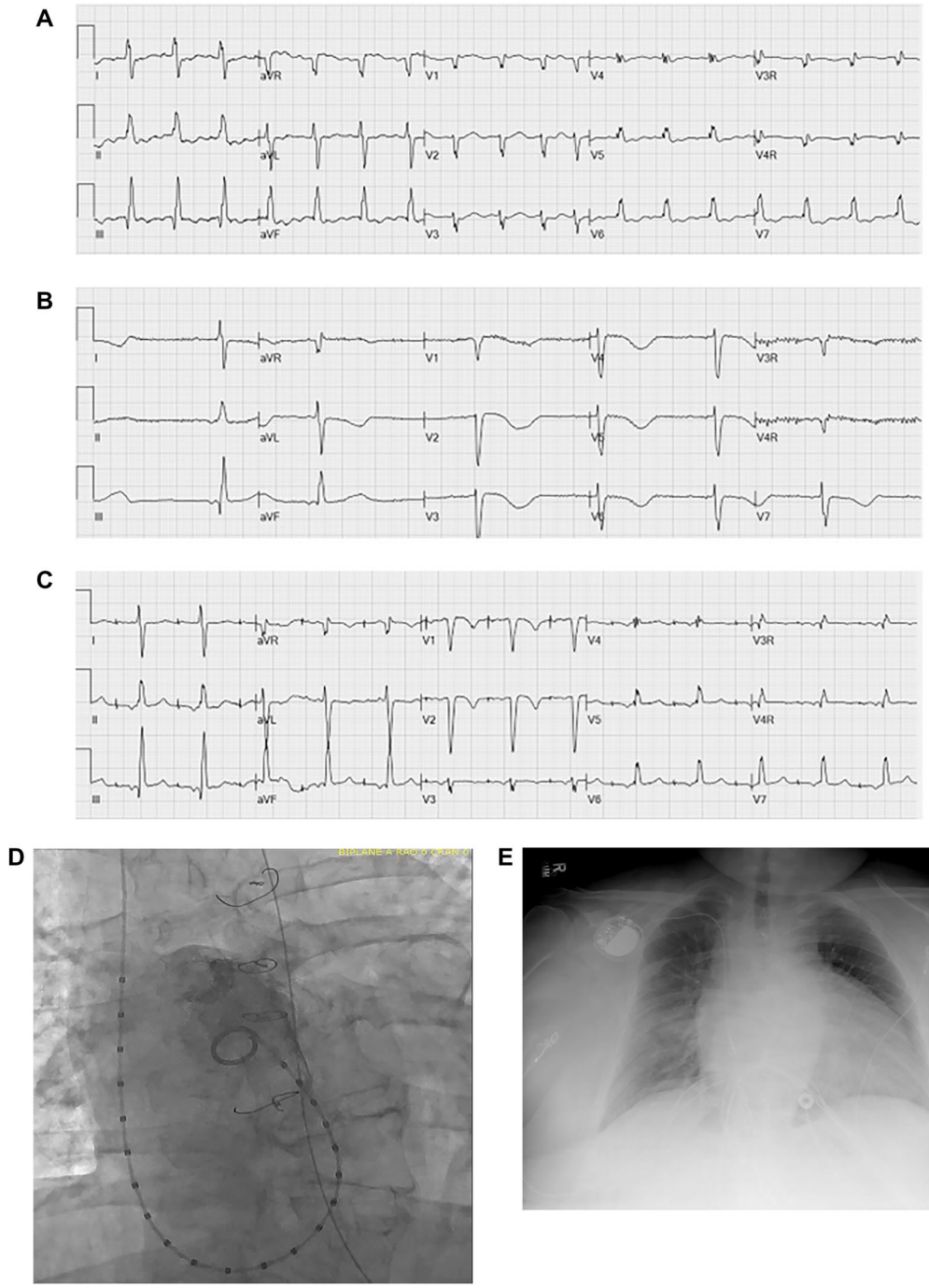


Fig. 5 Electroanatomic map (left) and voltage map (right) of IART in lateral tunnel Fontan with CMRI overlay (meshed). The use of high-density multi-electrode mapping catheters allows for rapid acquisition of electrical activation and voltage mapping. The use of advanced imaging integration into mapping systems allows for confirmation of anatomic structures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◀**Fig. 6** Transvenous atrial lead for treatment of sinus node disease and atrial arrhythmias in a 36-year-old woman with tricuspid atresia and AP Fontan with numerous comorbidities. Ablation was considered a poor option due to likely prolonged anesthesia time and high likelihood of recurrence. Permanent single chamber pacemaker placed under conscious sedation with no complications allowed for atrial pacing and anti-arrhythmic therapy. **a** ECG demonstrating atrial tachyarrhythmia. **b** ECG demonstrating baseline sinus node disease with baseline rhythm ectopic atrial bradycardia, prohibiting use of anti-arrhythmic therapy. **c** ECG after placement of transvenous atrial lead with atrial pacing and intact AV conduction, allowing for addition of anti-arrhythmic medications. **d** Angiogram of a 36-year-old with AP Fontan, demonstrating size of dilated Fontan. **e** Transvenous atrial lead placed successfully in a 36-year-old woman with markedly dilated atriopulmonary Fontan and numerous comorbidities, not considered to be a candidate for Fontan conversion surgery

For adults with atrial tachyarrhythmias, it is standard to consider long-term prevention of thromboembolism by risk assessment. However, risk scores do not account for those patients with complex congenital heart disease. Thus, given that patients with Fontan palliation are at increased risk for thrombosis even without atrial arrhythmias, long-term anticoagulation is recommended in patients with Fontan palliation with IART or AF (class I recommendation) [13].

Catheter Ablation

Catheter ablation in patients with Fontan palliation should be performed at centers with experience in interventional procedures in patients with this anatomy, by electrophysiologists with expertise in ablation in patients with Fontan palliation, and in collaboration with ACHD cardiology team (class I recommendation) [47]. Catheter ablation of atrial arrhythmias may offer a curative option, though recurrence rates are high, and consideration of ablation warrants an in-depth discussion about risks and anticipated long-term benefits for the patient. Individual patient anatomy and multiplicity of demonstrated arrhythmias contribute to the complexity and anticipated success of the procedure. Challenges to catheter ablation in adult patients with Fontan palliation include catheter access to the arrhythmia substrate, as with extracardiac Fontan anatomy in which access to atrial tissue involves trans-baffle puncture of extracardiac conduit, the difficult location and fragility of the intrinsic conduction system, and the existence of multiple or complex arrhythmias [13, 54]. Additionally, vascular access may complicate any attempt at ablation procedure, as with patients with heterotaxy who have interrupted IVC, or those patients with venous occlusions. Review of operative notes, advanced imaging data, and arrhythmia tracings are imperative to estimate chances of ablation success, as well as to plan ablation approach. As a result of complexity and potential multiplicity of arrhythmias, ablation procedures may be long and patient comorbidities may significantly increase anesthetic risk such that risks of the procedure may outweigh benefits.

Approaches to catheter ablation of atrial arrhythmias in patients with Fontan palliation have evolved with newer technologies such as the use of high-density multi-electrode mapping catheters [55], magnetic navigation, irrigated contact force ablation catheters, as well as the integration of advanced imaging data, which have improved the efficiency of ablation procedures in this complex population (Fig. 5). Success rates from catheter ablation in the patient with Fontan are variable, ranging from 40% to 75%, with recurrence of tachycardia in 60% of patients during the first year afterwards [39], emphasizing the need for patient-specific risk–benefit assessment in considering ablation for atrial arrhythmias. Patients with Fontan palliation typically have challenging arrhythmia substrates due to thickened atrial tissue, patient-specific anatomic variability, extensive atrial scar creating significant conduction delay, and variable circuits which may not allow for entrainment mapping to confirm good ablation site. While recurrences may be caused by the previously targeted arrhythmia, post-ablation arrhythmias are more likely new or previously unrecognized arrhythmias. Repeat ablation procedures are performed in more than one-third of patients with classic atriopulmonary

Fontan palliation [37]. For patients with failing Fontan physiology undergoing Fontan revision surgery, surgical ablation is part of the Fontan conversion procedure and includes right and left atrial maze procedures during the Fontan conversions; however, microreentrant atrial tachycardias and slower organized macroreentrant atrial tachycardias can persist after Fontan conversion [33].

Permanent Pacing

Sinus node disease resulting in sinus bradycardia or junctional rhythm is prevalent in adults with Fontan palliation, and the presence of sinus bradycardia and junctional rhythm can increase atrial arrhythmias. Atrial pacing may help reduce bradycardia-induced tachyarrhythmias both by increasing atrial rate and by allowing increased anti-arrhythmic therapy. There may be multiple reasons to consider atrial pacing in a patient with Fontan; however, with the development of atrial myopathy with age in a patient with Fontan, obtaining an atrial lead with reliable atrial sensing and good pacing thresholds may be challenging. It is important to consider what needs to be achieved with pacing and what the best options for lead placement will be. Depending on the patient's underlying anatomy, consideration of an epicardial pacing system via a left thoracotomy often gives access to the left atrium and ventricle, free of incisional scars and avoiding the difficulties of a repeat sternotomy.

Anatomy is an important consideration for permanent pacing in patients with Fontan palliation. For those patients with AP Fontan or lateral tunnel Fontan there is direct venous access to atrial tissue in the Fontan baffle, allowing for consideration of a single chamber atrial pacemaker sinus node disease with atrial pacing while also allowing anti-arrhythmic therapy to treat atrial arrhythmias (Fig. 6). However, the presence of a Fontan fenestration or baffle leak is a contraindication for transvenous lead placement because of the existence of right to left intracardiac shunt and risk of thromboembolic event. Additionally, if the patient needs ventricular pacing because of AV node disease or needs atrial anti-tachycardia pacing, this almost always must be achieved with an epicardial ventricular pacing lead. For those patients with extracardiac conduit Fontan, there is no usual venous access to atrial tissue for placement of transvenous atrial lead, necessitating epicardial systems in most.

CONCLUSION

Surgical evolution and advancements have led to increased survival of patients with single ventricle physiology. However, even in patients with lateral tunnel or extracardiac Fontan palliation, atrial arrhythmias occur at higher prevalence as the patient ages. Treatment of atrial arrhythmias in patients with single ventricle physiology is multifactorial and includes consideration of anti-arrhythmic therapy, atrial pacing, and ablation procedure. The best approach for management of atrial arrhythmias in adults with Fontan palliation is patient-specific and involves collaborative discussion between congenital electrophysiologists, adult congenital cardiologists, and the patient.

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