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Arrhythmias in Complex Congenital Heart Disease

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Abstract

Late after surgical repair of complex congenital heart disease, atrial arrhythmias are a major cause of morbidity, and ventricular arrhythmias and sudden cardiac death are a major cause of mortality. The six cases in this article highlight common challenges in the management of arrhythmias in the adult congenital heart disease population.

Keywords

atrial fibrillation; atrial flutter; catheter ablation; congenital heart disease; ventricular tachycardia

Introduction

Over one million adults are living with congenital heart disease (CHD) in the United States and this group now outnumbers children with CHD.[1, 2] Late after surgical repair of complex congenital lesions, atrial arrhythmias are a major cause of morbidity, and ventricular arrhythmias and sudden cardiac death (SCD) are a major cause of mortality.[3-7] Arrhythmia mechanisms include reentry due substrate from previous surgeries, the long-term consequences of hemodynamic abnormalities such as chamber enlargement and hypertrophy, and direct results of congenital abnormalities, such as the presence of accessory pathways. It has been reported that the prevalence of atrial arrhythmias is 15% in adults with CHD; for patients with complex CHD, the lifetime risk of atrial arrhythmias is over 50%.[8] Atrial arrhythmias in these patients are associated with increased risk of stroke, heart failure, and mortality.[8] Ventricular arrhythmias are also common in CHD, especially in patients with tetralogy of Fallot (TOF), ventricular septal defect, Ebstein's anomaly and systemic right ventricles. Drug therapy is often inadequate for these patients. Amiodarone is often avoided in younger patients due to concerns over long-term toxicity; class IC agents may have lower efficacy than in other patient groups[9] and may be contraindicated due to

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underlying structural heart disease. In experienced centers, catheter ablation has emerged as the preferred therapeutic option for atrial and ventricular arrhythmias in the CHD population. As increasing numbers of patients reach adulthood, the burden of arrhythmias and SCD are expected to increase even further and the need for device implantations[10] and catheter ablation procedures will continue to grow. This review will focus on six cases that highlight common and important electrophysiology problems in the adult CHD population.

Case I

A 45-year-old woman with history of perimembranous VSD status-post patch repair, moderate residual RV enlargement, and supraventricular tachycardia (SVT) status-post ablation at another hospital 8 years prior was admitted with palpitations and SVT (Figure 1). Electrophysiology (EP) study revealed two intraatrial reentrant tachycardias (IARTs) involving a posterolateral right atrial scar (Figure 2). Radiofrequency catheter ablation of the isthmus within the scar terminated the arrhythmias (Figure 3).

The most common arrhythmia in older adults with CHD is IART. This is a macroreentrant circuit involving abnormal atrial tissue resulting from atriotomy incisions, fibrosis, or patches[11-13] and characterized by large areas of low voltage with multiple heterogeneous channels.[14] IART can be seen in any patient who has undergone atriotomy, such as this patient, but the incidence is particularly high for patients with dextro-transposition of the great arteries (D-TGA) status post Mustard[15] or Senning repair and patients with a single ventricle status post Fontan. Fontan patients treated with older intraatrial lateral tunnel operations are at higher risk than those treated with extracardiac Fontan operations.[16-20] Atrial rates in IART are typically 150-250 bpm and 1:1 AV conduction can result in presyncope, syncope, or SCD.[21] As in this patient, multiple circuits are common.

Catheter ablation has been used with success in experienced centers. Complete procedural success has been reported to be as high as 80% with the use of irrigated ablation catheters and electroanatomic mapping, but recurrence has been reported in about 40% of patients. Arrhythmia recurrence is more common for those with multiple circuits, atrial fibrillation, and Fontan physiology.[22] Because IART has been associated with thromboembolism,[23] adequate anticoagulation with periprocedural TEE guidance according to standard guidelines is recommended.

Case II

A 50-year-old man with D-TGA status post Mustard procedure presented with dyspnea on exertion and was found to have pulmonary venous baffle stenosis and right-to-left shunting suggesting a systemic venous baffle leak. At the time of stenting for the pulmonary venous baffle stenosis, he was found to be in atrial flutter at cycle-length 280 msec. Flutter waves were negative in the inferior leads and positive in V1 suggesting typical counterclockwise flutter. During the procedure the patient developed 1:1 AV conduction in atrial flutter and became hypotensive, requiring external cardioversion. After several weeks of anticoagulation with warfarin, he underwent EP study. Typical atrial flutter was induced and dependence on the cavotricuspid isthmus was proven with entrainment (Figure 4). Multiple radiofrequency lesions were placed in systemic venous atrium proximal to the baffle at 4

o'clock on tricuspid annulus. Because the baffle prevented access to the anterior part of the cavo-tricuspid isthmus, the ablation catheter was then advanced via the baffle leak to the pulmonary venous (morphologic right) atrium and positioned along the cavo-tricuspid isthmus anterior and adjacent to the initial lesion set (Figure 5). The ablation line was then continued anteriorly towards the tricuspid (systemic AV) valve. After ablation, atrial flutter was not inducible. Three weeks later, the patient's baffle leak was closed percutaneously using two vascular plugs. The patient has been free from arrhythmia symptoms and atrial flutter at follow-up.

The patient underwent a follow-up event monitor for atrial arrhythmias prior to stopping anticoagulation. He did not have any atrial flutter but was found to have nonsustained ventricular tachycardia (NSVT), up to 6 beats in duration. Given moderate-to-severe systemic (RV) dysfunction, NSVT, prolonged QRS duration, and resting bradycardia, he underwent dual chamber ICD implantation.

D-TGA accounts for 3-7% of congenital heart defects.[24] Historically, most patients with DTGA were treated with atrial switch operations (Mustard or Senning baffles), but since the 1980s most patients have been treated with the arterial switch operation. However, most adults followed in CHD clinics underwent atrial switch repairs. These subjects are at high risk for congestive heart failure; ventricular arrhythmias and SCD are the leading cause of late mortality.[25-28] In the largest retrospective study of these patients, ventricular tachycardia and SCD were correlated with NYHA class, systolic dysfunction of the systemic ventricle, and QRS duration (hazard ratio 13.6 for QRS duration ≥140 msec) but not with supraventricular arrhythmias.[29] Another study reached different conclusions: that supraventricular arrhythmias, not QRS duration, were predictive of ventricular arrhythmias.[30] However, the decision to place an ICD in this patient is largely extrapolated from primary prevention data in patients with nonischemic cardiomypathy (reduced function of the systemic ventricle and NSVT).[31-34]

Full access to the cavo-tricuspid isthmus via the systemic venous atrium in patients treated with older Mustard or Senning repairs is often limited by baffles. In these cases, completion of a cavo-tricuspid isthmus line often requires ablation in the systemic venous atrium followed by accessing the pulmonary venous atrium via either a retrograde approach or traversing the baffle. If there is no baffle leak present, access to the pulmonary venous atrium can be obtained through a trans-baffle puncture or retrograde aortic approach.

Case III

A 76-year-old woman with levo-transposition of the great arteries (L-TGA) presented with fatigue and dyspnea on exertion. She was found to have both atrial fibrillation and atrial flutter (Figure 6) and both of these arrhythmias recurred despite amiodarone and cardioversion. Because of the severe symptoms, she underwent EP study, which induced cavo-tricupid isthmus dependent atrial flutter and a roof-dependent IART. She underwent ablation of both tachycardias and pulmonary vein isolation (Figures 7 and 8).

As patients with CHD live longer, atrial fibrillation is an increasing problem.[35] In one study of pulmonary vein antrum isolation in patients with CHD, similar success rates were

reported to patients without CHD, although in 60% of CHD patients the only congenital abnormality was an atrial septal defect.[36] Accessing the pulmonary venous atrium in patients with CHD can be technically more difficult with abnormal anatomy and baffles, but can be accomplished safely in most cases. Intracardiac echocardiography can be useful for trans-baffle puncture and identifying important anatomical structures in CHD patients.[37, 38] In patients with L-TGA, the electrophysiologist should be aware of the possibility of other abnormalities, such as ventricular septal defect and tricuspid (systemic AV valve) regurgitation. In addition, dual AV nodes have been reported and conduction usually occurs via the anteriorly situated AV node.[39] The position of this node at the mitral-pulmonary continuity and the extended length of the His-Purkinje system are thought to render these structures susceptible to fibrosis; complete heart block is reported to occur at a rate of ∼2%/year in patients with L-TGA.[40]

Case IV

A 7-year-old girl with Ebstein's anomaly of the tricuspid valve presented with palpitations. She developed supraventricular tachycardia at 2 days of age, but this was controlled with propranolol and later with propafenone. At the age of 7, she began experiencing palpitations 3-4 times per month and had two episodes of syncope while dancing. Electrocardiogram showed pre-excitation (Figure 9) and event monitoring demonstrated a regular, narrow-complex tachycardia at a rate of 224 bpm. Due to symptoms and syncope, she underwent EP study. During atrial overdrive pacing, there was non-decremental, eccentric AV conduction with the earliest ventricular activation on the proximal coronary sinus catheter electrode pair. Orthodromic reciprocating tachycardia was induced with atrial extrastimulus pacing. During SVT, premature ventricular complexes introduced during His refractoriness advanced the subsequent atrial electrogram and reset the tachycardia. Based on these findings, a single manifest right posteroseptal accessory pathway was diagnosed and radiofrequency energy application just anterior to the coronary sinus os resulted in loss of preexcitation (Figures 10 and 11). Following this lesion, there was no antegrade or retrograde accessory pathway conduction observed. The patient has been free of arrhythmias at several years of follow-up.

Wolff-Parkinson-White syndrome (WPW) is the most common arrhythmia in Ebstein's anomaly but other arrhythmias, including atrial fibrillation, atrial flutter, AV nodal reentrant tachycardia, and ventricular tachycardia are seen in this patient population as well.[41, 42] Radiofrequency catheter ablation is the therapy of choice for WPW in suitable patients with Ebstein's anomaly. However, compared to catheter ablation of accessory pathways in patients without CHD, procedural success rates are lower and recurrence rates are higher[43] due to the presence of multiple pathways, right-sided pathway predominance,[44] and abnormal AV node location. Because atrial septal defect and patent foramen ovale are also common, these patients may be at increased risk for paradoxical embolism during catheter ablation. Intra-operative ablation of accessory pathways can also be considered for patients undergoing tricuspid valve surgery.[42]

Case V

A 46-year-old man with TOF status-post repair in childhood, severe tricuspid regurgitation (due to flail leaflet) and severe pulmonary regurgitation underwent EP study and ablation for typical atrial flutter and IART. EP study demonstrated inducible, unstable polymorphic ventricular tachycardia (PMVT) with triple extrastimuli. After tricuspid valve repair and pulmonary valve replacement, he underwent a follow-up EP study which induced unstable monomorphic ventricular tachycardia (MMVT) at a cycle-length of 260 msec with a left bundle branch block superior axis morphology. For this reason, he underwent ICD implantation. Two years later, he had an appropriate ICD discharge for MMVT.

TOF accounts for about 10% of congenital heart defects. Surgical repair has good intermediate and long-term results[45-49] and survival has improved dramatically over the last 25 years.[50] However, ventricular arrhythmias are common and SCD is the leading cause of late mortality.[4, 51, 52] Clinical sustained ventricular arrhythmias are reported to have a prevalence of ∼15% by 35 years of age, with increased incidence in even older patients.[5, 52] It has been reported that arrhythmias in these patients are correlated with QRS duration (especially over 180 msec), increase in QRS duration over time, number of prior surgeries, right ventricular dilation, the presence of an RVOT patch, pulmonary regurgitation, and LV diastolic dysfunction.[5, 52, 53] Several studies have shown that inducibility of ventricular arrhythmias at EP study has good sensitivity and specificity for predicting subsequent ventricular arrhythmias or SCD.[54-56] Importantly, MMVT and PMVT, both of which were induced in this patient, have been shown to be predictive of future events in the TOF population.[55] Because EP testing is invasive and a positive study alone in low risk patients probably does not justify ICD placement,[57, 58] EP study is reserved for patients with arrhythmia symptoms or abnormal results of other tests, such as rapid or frequent NSVT on Holter monitoring.

Due to areas of scar and slowed conduction in the right ventricle, TOF is the classic congenital heart defect resulting in MMVT. Because this is most commonly a macroreentrant circuit in scar of RV free wall or near the septal patch repair, the electrocardiogram in VT typically shows a left bundle branch block pattern with an inferior axis, but other morphologies can be seen as well.[59] MMVT is also seen in CHD patients with ventriculotomy incisions, VSD patches, and Ebstein's anomaly.

ICDs, catheter ablation, and arrhythmia surgery are options for sustained VT or cardiac arrest in the CHD population. In TOF patients undergoing pulmonary valve replacement, intraoperative VT ablation has been used with success in experienced centers.[60-64] Catheter ablation of MMVT has good success rates,[65-68] but late ventricular arrhythmia recurrences are reported in the CHD population and as a result, ICDs are used as prophylaxis against sudden death.[68, 69]

Case VI

A 29-year-old man with D-TGA status post Mustard procedure and history of sudden cardiac arrest due to ventricular fibrillation (VF) at 23 years of age was admitted with a transient ischemic attack (TIA) manifesting as 10 minutes of numbness in his right face,

right arm, and right leg. After his cardiac arrest, he had undergone ICD placement. Due to lead fracture, he had undergone lead revision 4 years later and 6 months prior to his presentation for TIA, he had undergone lead revision at another hospital due to T-wave oversensing. Chest x-ray and transthoracic echocardiogram demonstrated that the ICD lead was incorrectly placed in the right (systemic) ventricle via a baffle leak (Figure 12). The device was extracted and a new lead was inserted into the left (subpulmonic) ventricle and the patient was placed on chronic anticoagulation with no further neurological symptoms.

The majority of adult CHD patients with an indication for ICD placement can undergo transvenous system implantation. Lead failure is not uncommon, especially in younger patients.[70] In addition, caution must be taken in lead placement with abnormal venous anatomy. If baffle leaks are present, this can impact decisions regarding anticoagulation and it is possible to inadvertently place leads in the systemic ventricle, as in this case. These should be closed prior to lead implantation to prevent systemic thromboemboli. Some patients have anatomy that precludes access to the subpulmonic ventricle (baffle obstruction or stenosis) and some patients with a single ventricle or right-to-left shunts are at risk for thromboembolism with transvenous ICD placement. In these patients, nontransvenous systems are preferred.

Additionally, patients with CHD are at risk for PMVT and ventricular fibrillation (VF) due to pressure and volume overload, chamber enlargement, and hypoxemia. PMVT and VF are most commonly seen in congenital aortic stenosis, systemic RVs, and Eisenmenger's syndrome.[71] This patient initially underwent ICD placement due to VF in the setting of an abnormal systemic ventricle. It is important, however, to note that due to hemodynamic status and comorbidities, not all sudden deaths in high-risk patients with CHD are due to an arrhythmic cause.[72]

Summary

Atrial and ventricular arrhythmias are a common cause of morbidity and mortality in the growing population of adults with CHD. Patients with high-risk CHD lesions such as d-TGA, l-TGA, or Tetralogy of Fallot should be monitored routinely for arrhythmias and associated symptoms. Catheter ablation is an excellent therapeutic option for a variety of arrhythmias observed in these patients when performed in experienced centers by operators familiar with abnormal cardiac anatomy and associated abnormalities, such as residual shunts. ICD implantation is recommended for cardiac arrest survivors and CHD patients with sustained VTs discovered on electrophysiology study. In planning both catheter ablation and device implantation procedures, clinicians should review specific anatomy and all surgical records, and obtain detailed imaging to define possible obstructions or stenosis in vascular pathways.

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Abbreviations

CHD	congenital heart disease
D-TGA	dextro-transposition of the great arteries
EP	electrophysiology
L-TGA	levo-transposition of the great arteries
MMVT	monomorphic ventricular tachycardia
NSVT	nonsustained ventricular tachycardia
PMVT	polymorphic ventricular tachycardia
SCD	sudden cardiac death
SVT	supraventricular tachycardia
TOF	tetralogy of Fallot
TIA	transient ischemic attack
VF	ventricular fibrillation
WPW	Wolff-Parkinson-White syndrome

Key Points

- Atrial and ventricular arrhythmias are a common cause of morbidity and mortality in the growing population of adults with congenital heart disease.
- Patients with high-risk CHD lesions such as d-TGA, l-TGA, or tetralogy of Fallot should be monitored routinely for arrhythmias and associated symptoms.
- With the aid of electroanatomic mapping and newer irrigated radiofrequency energy delivery, catheter ablation is an excellent therapeutic option for a variety of arrhythmias observed in these patients when performed in experienced centers.
- ICD implantation is recommended for cardiac arrest survivors and CHD patients with sustained VT discovered on electrophysiology study.
- In planning both catheter ablation and device implantation procedures, clinicians should review specific anatomy and surgical records, obtain imaging to define possible obstructions or stenosis in vascular pathways, and be aware of associated congenital abnormalities.

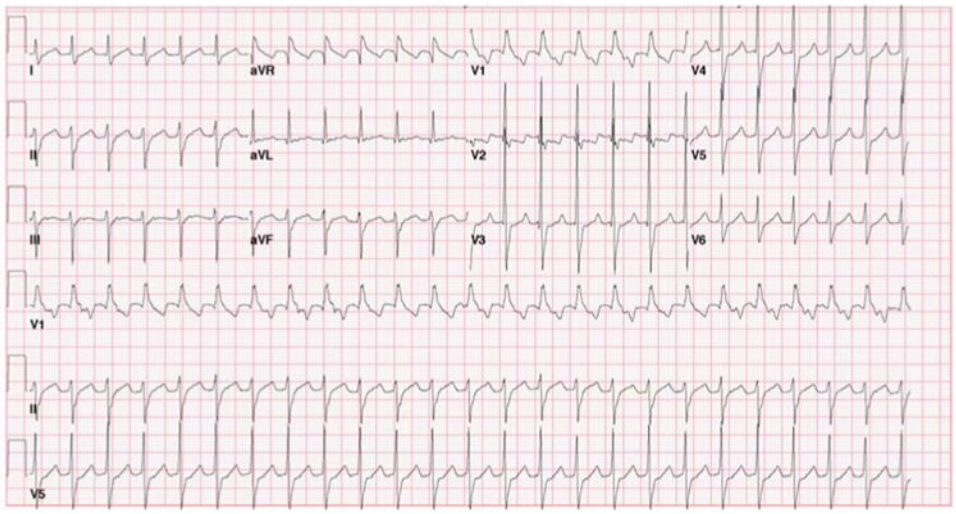

Figure 1.
Surface electrocardiogram of intraatrial reentrant tachycardia with 1:1 AV conduction.

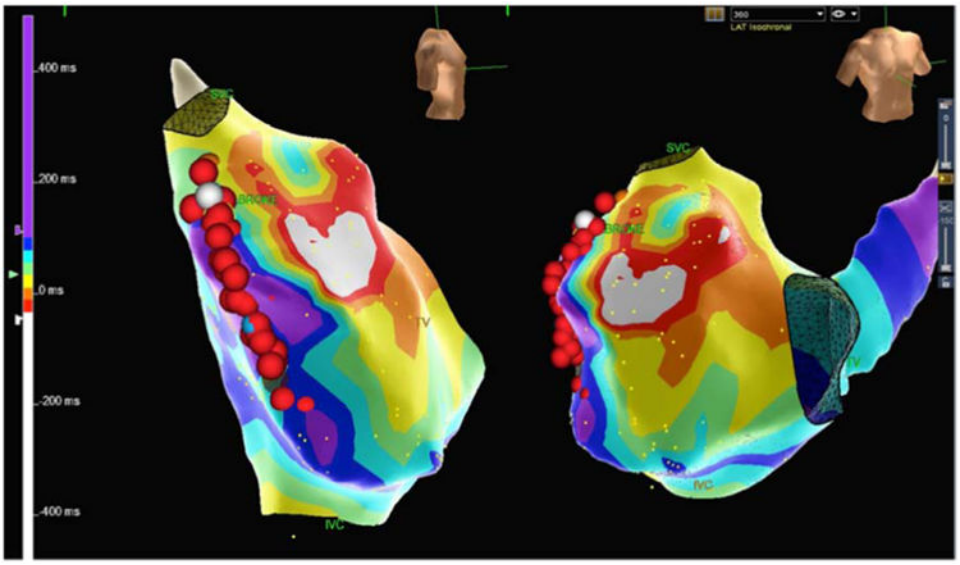

Figure 2.

Electroanatomic activation map demonstrating slow conduction in the isthmus of the intraatrial reentrant tachycardia circuit (purple areas) in the posterolateral right atrium at the site of a previous atriotomy incision. Ablation through this isthmus resulted in termination of tachycardia (white sphere); the RF lesion set was completed by connecting the areas of low voltage in the atriotomy incision to areas of normal voltage.

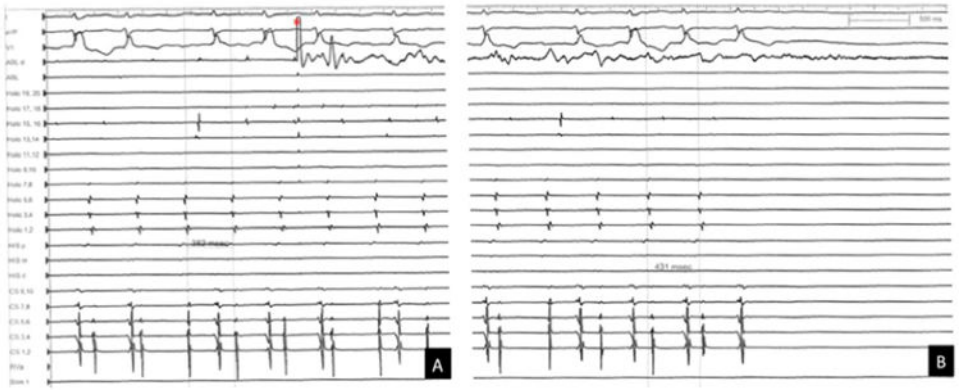

Figure 3.

A: Prior to the onset of radiofrequency energy application (*), intracardiac electrograms in intraatrial reentrant tachycardia demonstrate a diastolic signal on the ablation catheter representing conduction within the isthmus of the scar. B: During radiofrequency ablation, the tachycardia cycle length increases by 50 msec prior to termination.

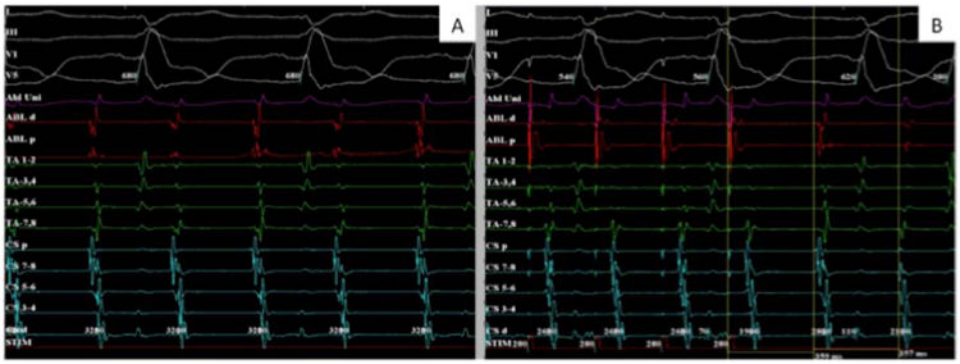

Figure 4.

A: Atrial flutter. B: The post-pacing interval after overdrive pacing from the cavotricuspid isthmus (Abl d) was equal to the tachycardia cycle length.

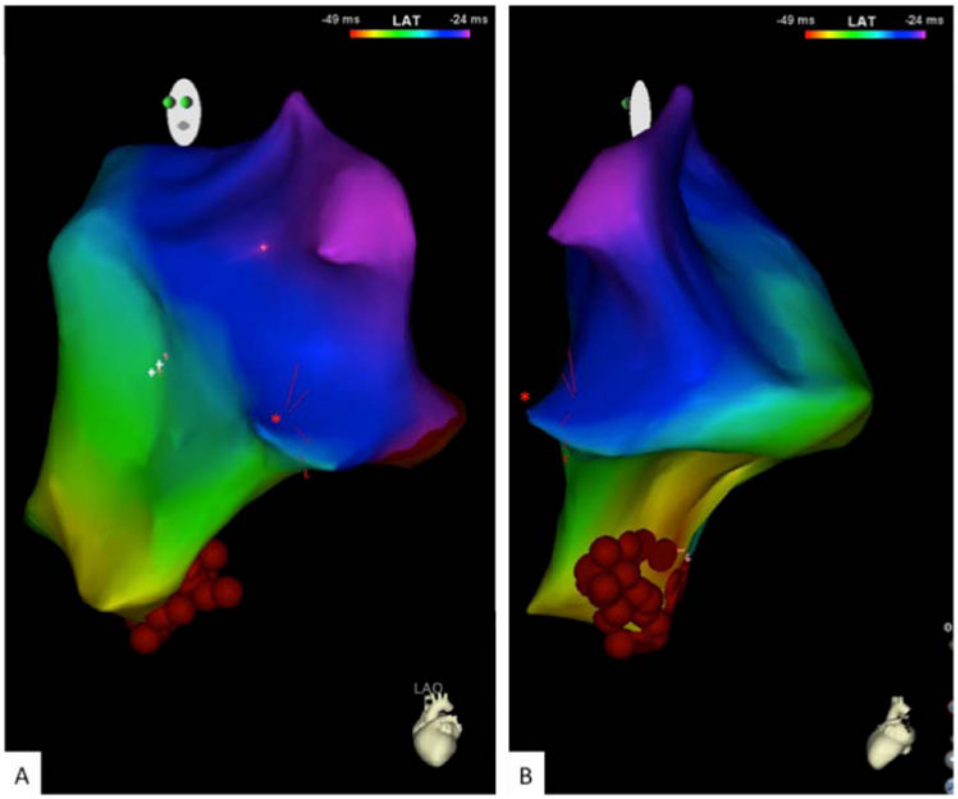

Figure 5.

LAO (A) and left lateral (B) electroanatomic activation map systemic venous atrium. Completion of the line across the cavo-tricuspid isthmus required accessing the pulmonary venous atrium via a baffle leak (*).

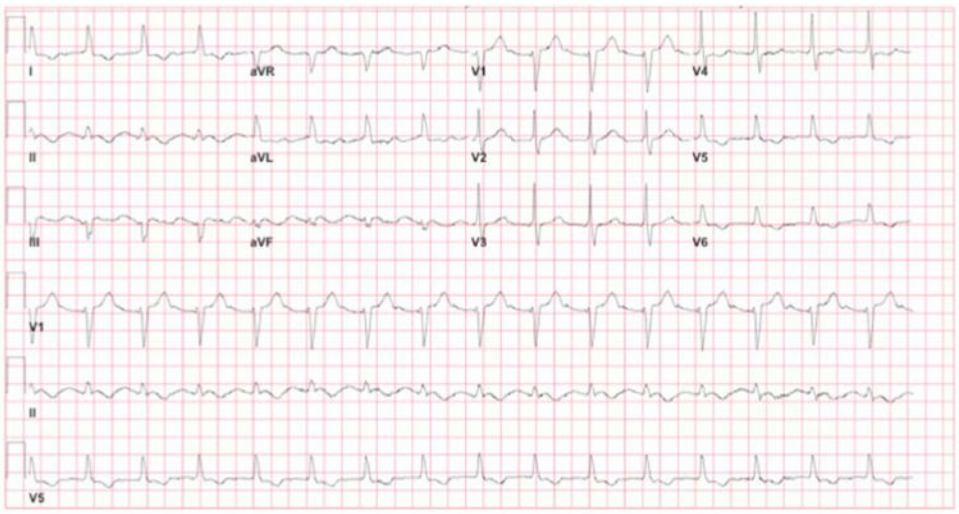

Figure 6.

Atrial flutter with 2:1 AV conduction. Note the absence of septal q-waves in the lateral precordial leads due to right-to-left depolarization of the interventricular septum in levo-transposition of the great arteries.

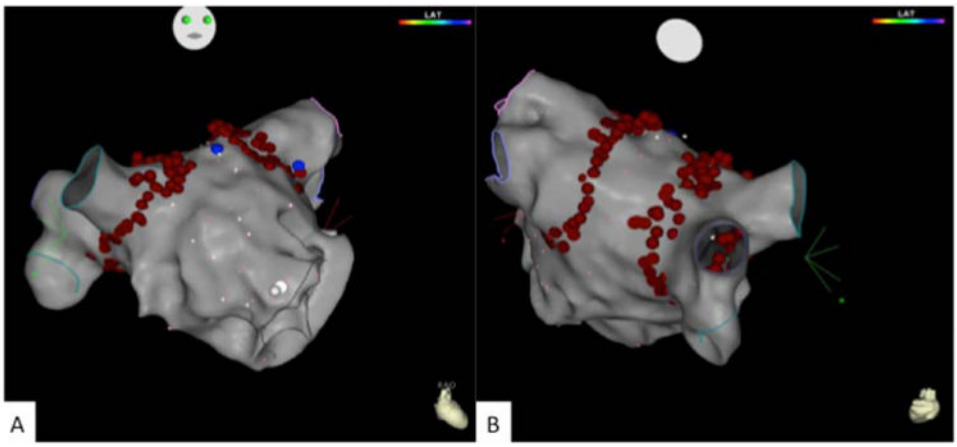

Figure 7.

A: RAO view of the normally-positioned left atrium after isolation of the pulmonary veins. B: Posteroanterior view of the left atrium after pulmonary vein isolation. In levo-transposition of the great arteries, the left atrium connects to the right (systemic) ventricle via the tricuspid valve.

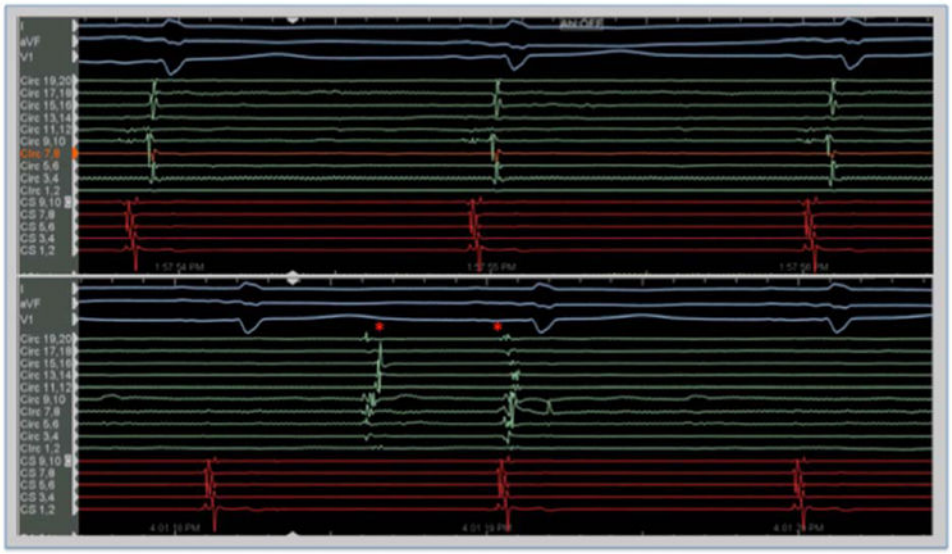

Figure 8.

Top: Sinus rhythm with conduction into left upper pulmonary vein (LUPV) prior to ablation. Bottom: Dissociation of LUPV potentials (*) after pulmonary vein isolation.

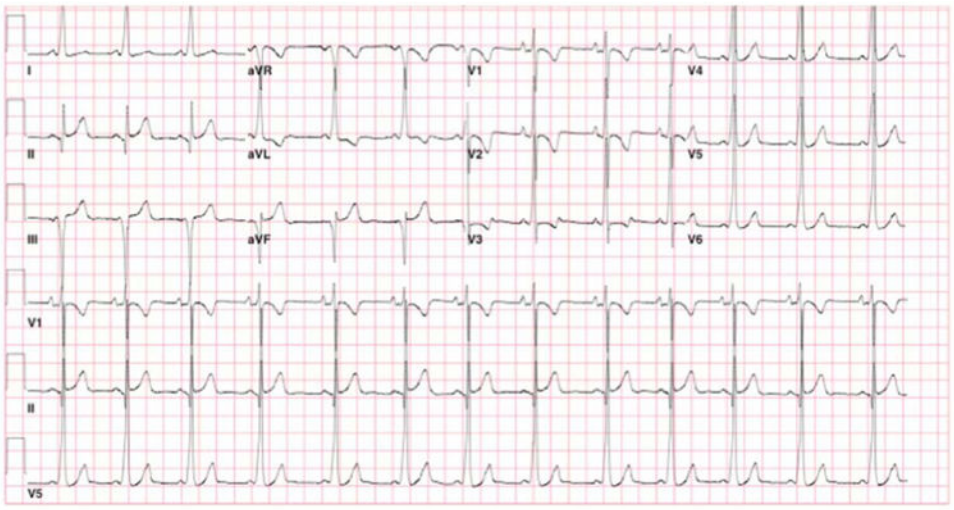

Figure 9.

Baseline ECG showing pre-excitation. The V2 precordial transition with a dominant R-wave in lead I suggests a right-sided accessory pathway. As is common with posteroseptal accessory pathways, there is a pseudo inferior myocardial infarction pattern in the inferior leads.

Figure 10.

During RF ablation while pacing the atrium, loss of pre-excitation (*) and delay of the earliest ventricular electrogram on the proximal coronary sinus catheter electrode (CS 9,10) were noted.

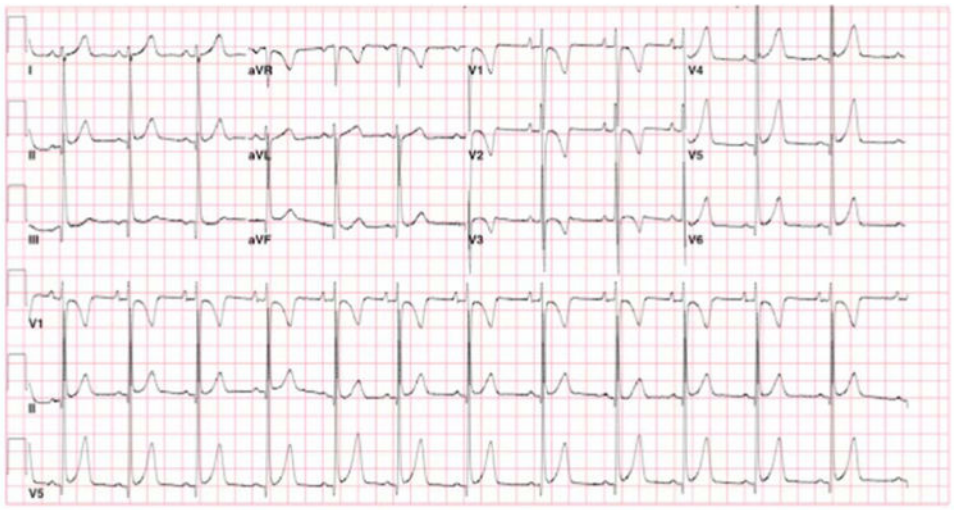

Figure 11.

Post-ablation ECG shows loss of delta wave. Right atrial abnormality, seen here, is a common finding in Ebstein's anomaly. Right bundle branch block is also common, although it is not seen in this case.

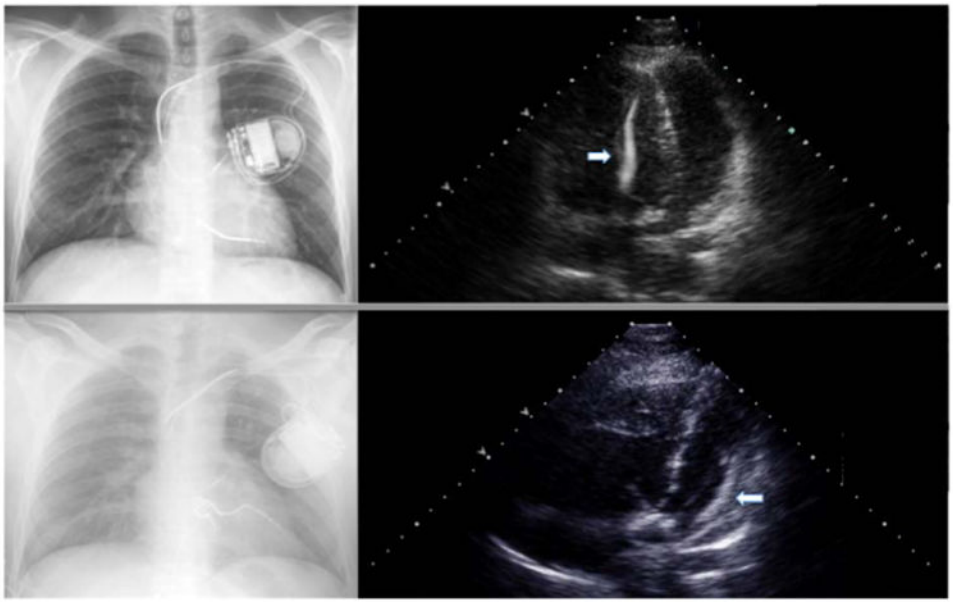

Figure 12.
Top: ICD lead (arrow) in RV (systemic ventricle) via Mustard baffle leak. Bottom: ICD lead (arrow) in LV (subpulmonic ventricle). Left – chest radiograph. Right – apical 4-chamber surface echocardiogram.