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Successful Radiofrequency Ablation of Recurrent Supraventricular Tachycardia in a Patient with Complex Congenital Heart Disease

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

ABCDEF 1 Nkechi Christiana Arinze
ABCDEF 1 Harry O. Eyituoyo
ABCDEF 1 Rieta N. Aben
ABCDEF 1 Dat P. Vu
ABCD 2,3 Daniel B. Haithcock
ABCD 2,3 Joseph Poku
ABCD 2,3 Felix O. Sogade

1 Department of Internal Medicine/Community Medicine, Mercer University School of Medicine, Macon, GA, U.S.A
2 Department of Cardiology and Electrophysiology, Georgia Arrhythmia Consultants and Research Institute, Macon, GA, U.S.A.
3 Department of Cardiology and Electrophysiology, Navicent Health, Macon, GA, U.S.A.

Corresponding Author: Nkechi Christiana Arinze, e-mail: dr.kristyne@gmail.com
Conflict of interest: None declared

Patient: Male, 21-year-old
Final Diagnosis: Atrioventricular nodal reentrant tachycardia
Symptoms: Palpitation
Medication: —
Clinical Procedure: Radiofrequency catheter ablation
Specialty: Cardiology

Objective: Rare disease

Background: Radiofrequency ablation (RFA) is the criterion standard treatment for patients with atrioventricular nodal re-entrant tachycardia (AVNRT). Knowledge about RFA in patients with dextrocardia and situs inversus is limited due to their rare incidence and complexity. The incidence of dextrocardia is reported to be 1 in 12 000 births, with situs inversus occurring in one-third of the cases. The incidence of congenital heart disease is about 5% in these patients. However, data on rhythm and conduction disorders in this group of patients are currently limited, making management more difficult owing to their individual anatomy.

Case Report: We report the case of an obese 21-year-old man with complex congenital heart disease (CCHD) (situs inversus dextrocardia, pulmonary atresia, single ventricle, common atrium with single atrioventricular valve), asplenia, and multiple cardiac-corrective surgeries (Fontan repair, bidirectional Glenn anastomosis, and Blalock-Taussig shunt) who underwent successful RFA of recurrent supraventricular tachycardia.

Conclusions: Supraventricular arrhythmias are common in the setting of CCHD. Although catheter ablation procedures are technically challenging to perform in patients with CCHD, they remain the best therapeutic option for these arrhythmias. To our knowledge, this case is the first to be described in the literature of successful ablation of AVNRT in a patient with situs inversus dextrocardia, pulmonary atresia, a single ventricle, a common atrium with a single atrioventricular valve, and multiple cardiac-corrective surgeries.

MeSH Keywords: Catheter Ablation • Dextrocardia • Fontan Procedure • Heart Defects, Congenital • Tachycardia, Supraventricular

Full-text PDF: https://www.amjcaserep.com/abstract/index/idArt/928147

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Background

Radiofrequency ablation (RFA) is the criterion standard treatment for patients with atrioventricular nodal reentrant tachycardia (AVNRT). Knowledge about RFA in patients with dextrocardia and situs inversus is limited due to their low incidence and high complexity. The incidence of dextrocardia is reported to be 1 in 12 000 births, with situs inversus occurring in one-third of the cases [1]. The incidence of congenital heart disease is about 5% in these patients. However, data about rhythm and conduction disorders in this group of patients are currently limited, making management more difficult owing to their individual anatomy [2].

Case Report

An obese 21-year-old man with complex congenital heart disease (CCHD) presented to the Emergency Department (ED) with recurrent palpitations. The palpitations began at rest and were severe, constant, and sudden in nature without notable aggravating or alleviating factors. He had no associated chest pain, dyspnea, nausea, vomiting, or diaphoresis. Upon physical examination, the patient was tachycardic with a heart rate of 103 beats per minute (bpm) and a body mass index of 41 kg/m^2; examination of other systems was unremarkable. The patient endorsed previous episodes, including an episode involving exertional palpitations approximately 1 month prior. At that time, symptomatic management was achieved using a 24-mg titrated dose of adenosine.

The patient has a history of CCHD, including situs inversus, dextrocardia, pulmonary atresia, a single ventricle, a common atrium with single atrioventricular (AV) valve, and asplenia. Early in infancy, he had a palliative bidirectional Glenn anastomosis (a shunt from the superior vena cava to the pulmonary artery, thereby redirecting blood flow from the upper body to the lungs for oxygenation). He also had a definitive extracardiac fenestrated Fontan repair (a shunt from the inferior vena cava to the pulmonary artery, thereby redirecting blood flow from the lower body to the lungs) for the pulmonary atresia, and a Blalock-Taussig shunt placed to increase pulmonary blood flow. His past medical history was significant for coronary heart disease, congestive heart failure, dilated cardiomyopathy, and polycythemia. He was a candidate for heart transplant.

Early records showed that supraventricular tachycardias (SVTs) were diagnosed in the patient at 5 years of age. Over the years, the SVTs became recurrent, which led to an average of 12 ED visits and hospitalizations per year. SVTs were relieved with the use of multiple antiarrhythmic medications (i.e., adenosine, sotalol, and amiodarone) and electrical cardioversion. Amiodarone was subsequently discontinued due to the hypersecretion of thyrotropin. Prior to the patient's presentation to our hospital, he had 3 attempted and failed RFAs for Wolff-Parkinson-White syndrome and various SVTs at different institutions in 2007, 2017, and 2018.

The differential diagnosis includes ventricular tachycardia, AVNRT, atrioventricular reentrant tachycardia (AVRT), Wolff-Parkinson-White syndrome, atrial flutter, atrial fibrillation, and electrolyte abnormality.

In the ED, the initial electrocardiogram showed a wide-complex QRS with a ventricular rate of 188 bpm (Figure 1). A chest X-ray showed dextrocardia and deformity of the right fourth rib (Figure 2). The patient was given 12 mg of adenosine, which converted him to normal sinus rhythm (Figure 3A). Complete blood count and comprehensive metabolic panels were unremarkable. Echocardiography showed an ejection fraction of 20–25%. The electrophysiology team was consulted, and the patient was admitted to the hospital for a repeat RFA. Intracardiac echocardiography and CARTO advanced 3-dimensional cardiac mapping was used for anatomical visualization (Figure 4). During the procedure, to enable obtaining a near-normal view, the precordial leads and lead I were reversed. The camera angles were placed in the right oblique view. Intracardiac echocardiography using a 10-Fr SoundStar catheter was utilized for anatomic definitions. With the advancement of the catheter, we felt far field signals in sequence from the conduits and inferred anatomical positions. Coronary sinus pacing could not be done, and the His catheter could not be identified because of fibrosis from previous cardiac surgeries. The only catheter we could use to manipulate was the mapping and ablation catheter. The technique we used to map the earliest shortest AV signal during tachycardia corresponded to the region of the posterior septal space. Due to the complexity of the heart and previous surgeries, we could not place enough catheters for diagnostic maneuvers to distinguish between AVNRT or AVRT. The shortest region of the AV interval close to the left-side slow pathway suggested that an AVNRT was most likely. Delivery of radiofrequency energy was done at this site and terminated the arrhythmia. Following ablation, no other inducible arrhythmia or accessory pathway was observed. The patient was noted to be in normal sinus rhythm (Figure 5).

Electrocardiography done at the 3-week follow-up showed a normal sinus rhythm (Figure 3B). The patient remains asymptomatic 18 months after catheter ablation.

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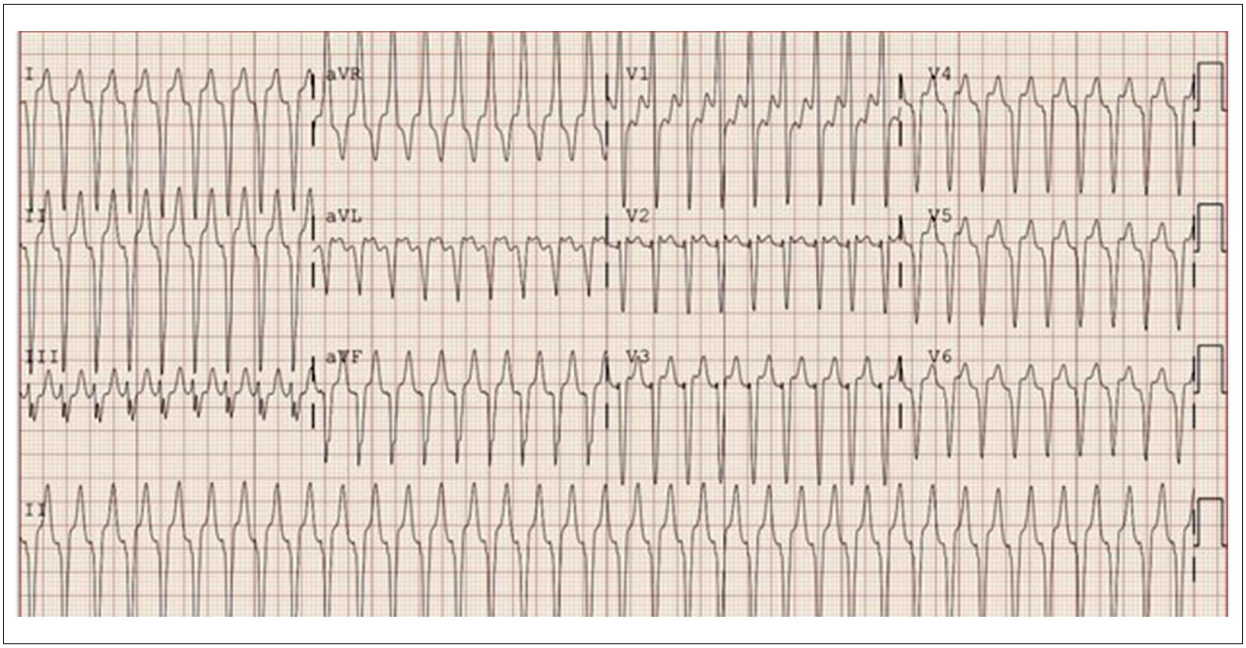

Figure 1. Twelve-lead electrocardiogram showing a regular, wide QRS tachycardia at a rate of 188 beats per minute in a patient with dextrocardia.

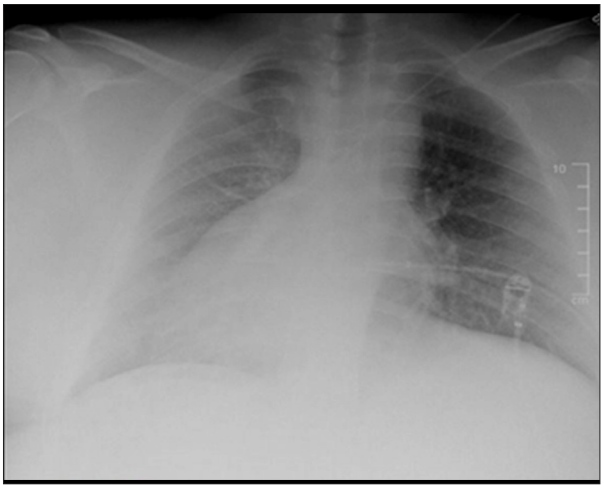

Figure 2. Chest X-ray showing dextrocardia and deformity of the right fourth rib.

Discussion

CCHD poses difficulties during catheter ablation procedures. Dextrocardia can be diagnosed on electrocardiogram: diffuse negativity in lead I, a positive deflection in the QRS complex in aVR, right axis deviation, and reverse R wave progression in precordial leads (V1-V6) [4]. The finding of a positively deflected QRS complex in aVR and a negatively deflected QRS complex in lead I may lead to misdiagnosis. The most common cause for this finding is reversed electrode placement of arm leads (left and right) and ectopic atrial rhythm.

A standardized catheter ablation procedure involves the use of electrocardiogram characteristics and anatomical landmarks to guide the placement of the catheter. To navigate the challenges posed by dextrocardia during ablation, we reversed the camera angles and viewed it through the right anterior oblique view. The precordial leads and lead I were reversed to have a near-normal anatomy.

Using fluoroscopy, 1 triport catheter was advanced from the groin to obtain anatomic definitions. First, the aorta and its 3 leaflets were defined: the left and right coronary cusps and the noncoronary cusp. The AV valve annulus was also identified. A 3-dimensional image was created constructively using the AV valve annulus. An effort was made to create an anatomic rendering of the single ventricle and portion of the single atrium. The images were superimposed on the uni view of the CARTO electroanatomic system. With the administration of isoproterenol, an initial wide QRS complex tachycardia was induced at 188 bpm, which later transformed to a slower arrhythmia, then a 2: 1 atrial flutter, a 1: 1 atrial flutter, and a wide QRS tachycardia before finally converting to normal rhythm after ablation (Figure 5).

During pacing, ventricular-atrial conduction was observed. At the end of the ablation, only a ventricular conduction was noted. We infer that an accessory pathway was ablated. Due to the CCHD, the maneuvers to confirm the accessory pathway could not be done.

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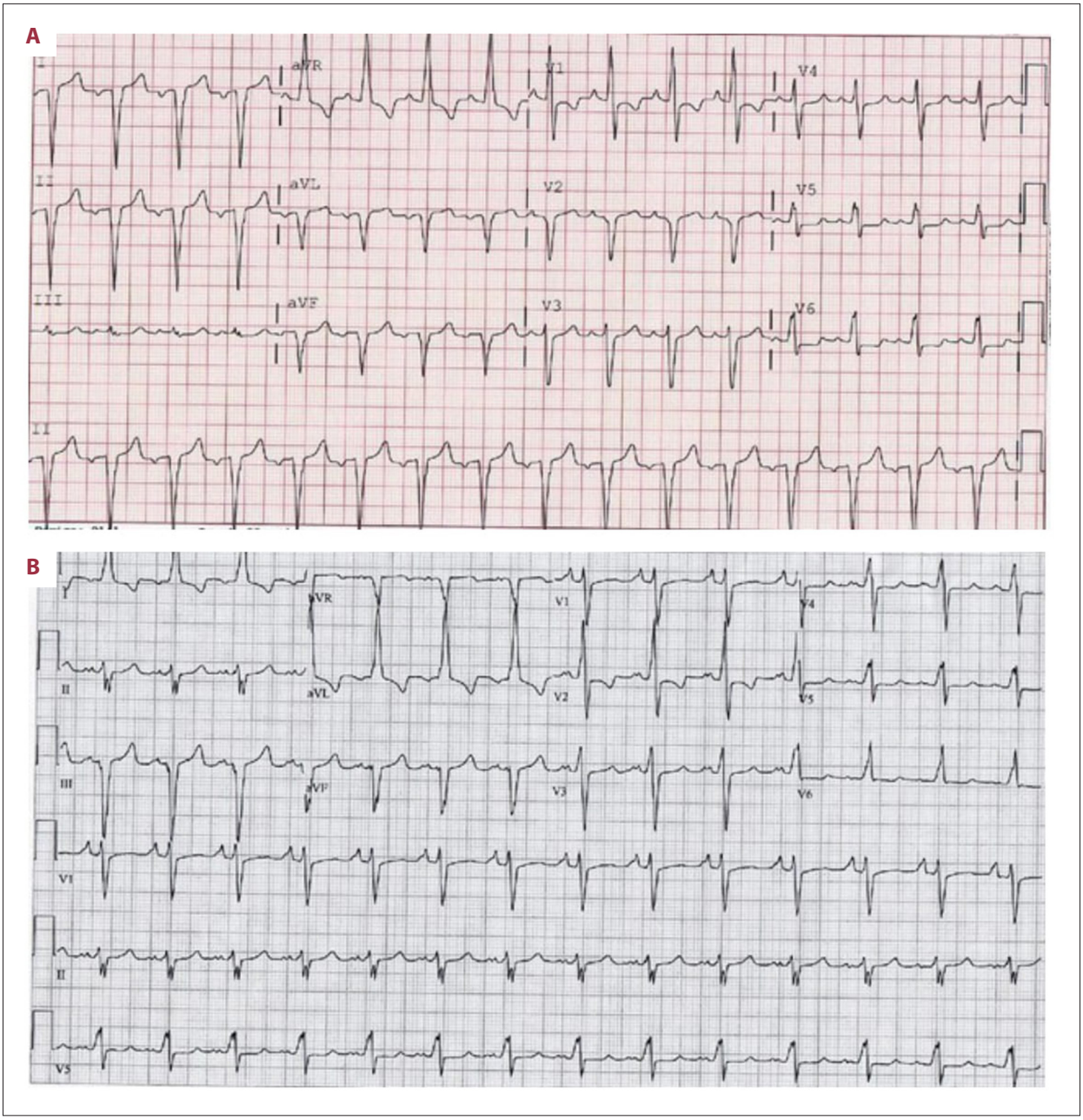

Figure 3. (**A**) Twelve-lead electrocardiogram showing normal sinus rhythm after adenosine administration. (**B**) Twelve-lead electrocardiogram done after ablation, showing normal sinus rhythm with a rate of 84 beats per minute.

Patients with CCHD are predisposed to atrial and ventricular arrhythmias owing to their structural heart disease, abnormal conduction system, and scarred tissues from surgical repair [3]. Surgical repairs lead to the genesis of electrophysiological substrates that result in early perioperative or late postoperative arrhythmias. The formation of these electrophysiological substrates leads to lethal arrhythmias and sudden cardiac death [6]. Various types of SVTs unrelated to the atrial scar from Fontan surgery can occur in CCHD patients because of the varied AV conduction system [5,6].

Cardiomyopathy with reduced ejection fraction, as noted in our patient, is associated with an increased risk of arrhythmias. The mechanism of arrhythmia in heart failure involves the remodeling of ion channels, calcium handling proteins, and gap junction-related molecules. These changes combine to form an electrophysiological substrate that can generate arrhythmias and sudden cardiac death [7,8]. Managing arrhythmias in patients with CCHD has been proven to be challenging [8]. Our patient was not on an implantable cardioverter defibrillator device because of the complexity of his congenital heart

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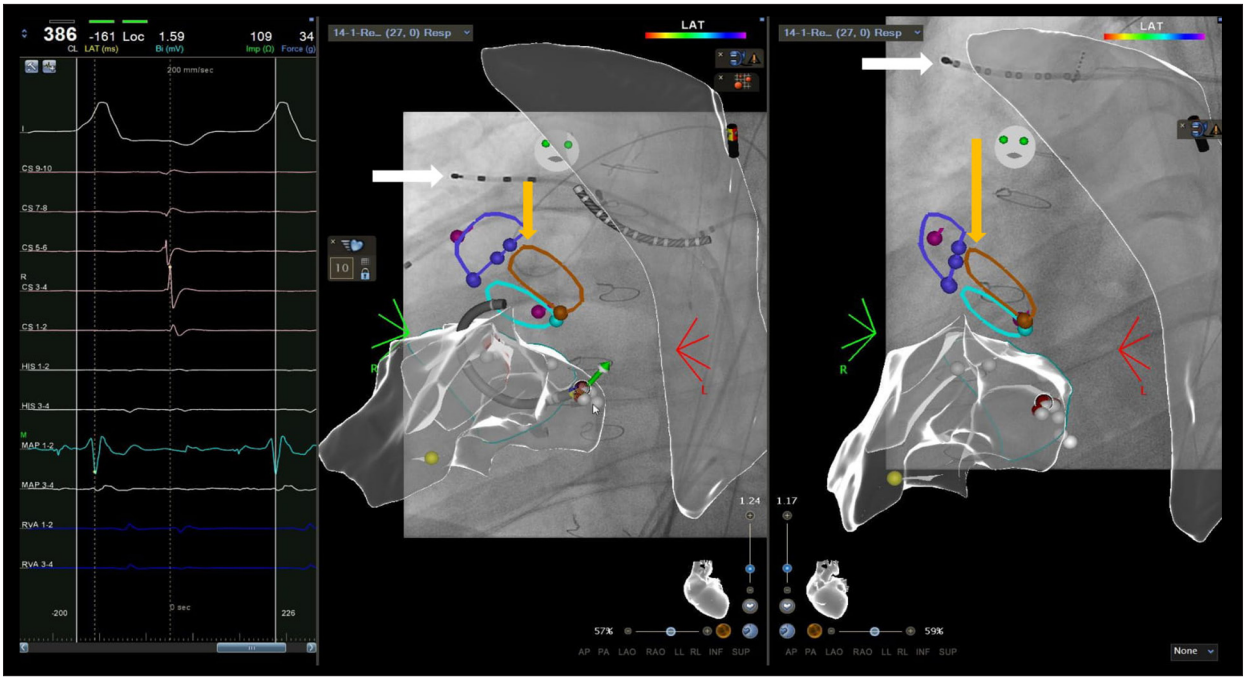

Figure 4. Intracardiac echocardiography and CARTO advanced 3-dimensional cardiac mapping showing catheter advancement (white arrow) and 3 cusps of the aortic valve (yellow arrow).

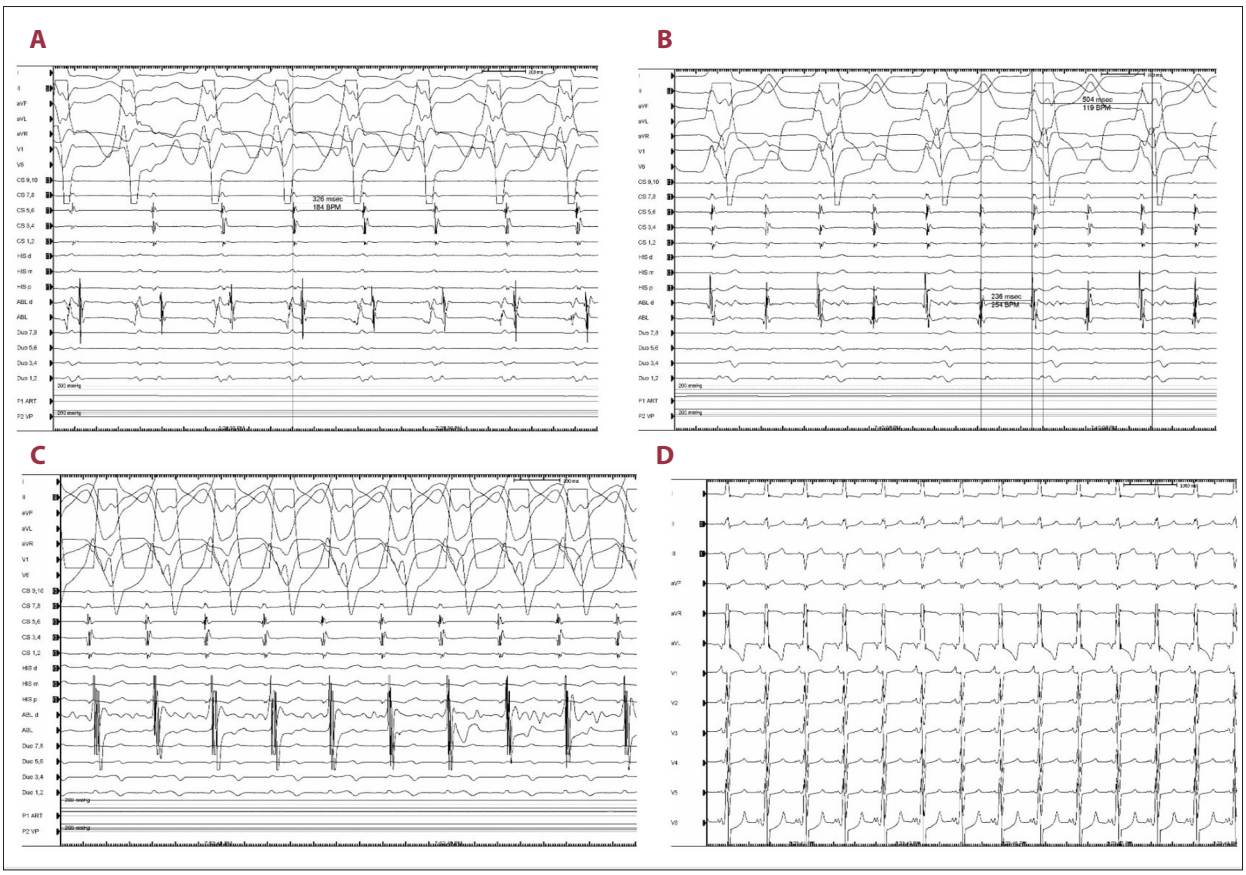

Figure 5. Intracardiac electrocardiography with precordial leads and lead 1 reversal showing (**A**) wide complex tachycardia; (**B**) 2: 1 atrial flutter; (**C**) atrial tachycardia with 1: 1 conduction, and (**D**) normal sinus rhythm.

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disease. He is a candidate for a life vest, and he is on the list for heart transplant.

In this unique case, our patient had multiple corrective cardiac procedures (Fontan, Glenn, and Blalock-Taussig) and 3 previous failed RFAs. Little has been reported about catheter ablation in patients with dextrocardia and complex cardiac defects with prior surgical history. Yamada and Lau [9] reported a case of a patient with a univentricular dextrocardia with Fontan conversion that was ablated for a focal atrial tachycardia. Aksu et al. [4] reported a case of a 34-year-old woman with mirror-image dextrocardia due to unilateral pulmonary agenesis who underwent successful slow pathway ablation for typical AVNRT [4]. Another report described a 14-year-old boy with a common atrium and ventricle with prior extracardiac Fontan operation who underwent catheter ablation for recurrent tachycardia [10]. To the best of our knowledge, our case is the first reported case of successful radiofrequency ablation of SVT in a patient with CCHD (dextrocardia, single ventricle, single atrium, pulmonary atresia, and situs inversus) with a history of Fontan, bidirectional Glenn, and Blalock-Taussig procedures.

The most common types of SVT include AVNRT, AVRT, and atrial tachycardia. Key maneuvers in electrophysiology are used to help differentiate these arrhythmias. In our case, the different maneuvers used to distinguish the different types of supraventricular tachycardias (V-pacing maneuver, PVC maneuver, His synchrony maneuver) were not performed because of the complexity of the congenital heart disease and single-chambered heart. The other challenges we encountered arose from the use of one catheter to map and ablate the heart owing to the single-chambered heart. No coronary sinus catheter was used during the ablation. The cardiac anatomy would have been better outlined with the use of contrast imaging. The mechanism of the SVT in our patient was an AVNRT. Our patient already had 3 attempted and failed RFAs because of his complex cardiac anatomy. The success of our case can likely be attributed to the expertise of the electrophysiologist.

Conclusions

Supraventricular arrhythmias are common in the setting of CCHD. Although technically challenging to perform in CCHD patients, catheter ablation procedures remain the best therapeutic option for these arrhythmias. To the best of our knowledge, this case is the first to be described in the literature of successful ablation of AVNRT in a patient with situs inversus dextrocardia, pulmonary atresia, a single ventricle, a common atrium with a single AV valve, and multiple cardiac-corrective surgeries.

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Conflicts of interest

None.

References:

1. Bohun CM, Potts JE, Casey BM, Sandor GG: A population-based study of cardiac malformations and outcomes associated with dextrocardia. Am J Cardiol, 2007; 100(2): 305–9
2. Ocos M, Petrescu L, Vacarescu C et al: Situs inversus totalis and cardiac rhythm/conduction disorders. Romanian J Cardiol, 2018; 28(3): 331–33
3. Chiu SN, Wang JK, Lu CW et al: Electrophysiology study for complex supraventricular tachycardia in congenital heart disease patients with single-ventricle physiology. J Am Heart Assoc, 2016; 5(11): e004504
4. Aksu T, Guler TE, Golcuk E et al: Successful radiofrequency catheter ablation of atrioventricular nodal reentrant tachycardia in a patient with dextrocardia due to unilateral pulmonary agenesis: A case report. Int Med Case Rep J, 2015; 8: 41–45
5. Pundi KN, Johnson JN, Dearani JA et al: 40-Year follow-up after the Fontan operation long-term outcomes of 1,052 patients. J Am Coll Cardiol, 2015; 66(15): 1700–10
6. Jin H, Lyon AR, Akar FG: Arrhythmia mechanisms in the failing heart. Pacing Clin Electrophysiol, 2008; 31(8): 1048–56
7. Cazzoli I, Gunturiz-Beltran C, Guarguagli S et al: Catheter ablation for patients with end-stage complex congenital heart disease or cardiomyopathy considered for transplantation: trials and tribulations. Int J Cardiol, 2020; 301: 127–34
8. Van Hare GF: Challenges in ablation of complex congenital heart disease. Card Electrophysiol Clin, 2019; 11(4): 711–18
9. Yamada T, Lau YR: Successful catheter ablation of a focal atrial tachycardia originating from the coronary sinus ostium in a patient with a history of Fontan conversion and dextrocardia. Europace, 2018; 20(8): 1351
10. Dalili M, Rao JY, Brugada P: Radiofrequency ablation of accessory pathways in children with complex congenital cardiac lesions: a report of three cases. J Tehran Univ Heart Cent, 2013; 8(2): 111–15

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