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Case Report

Ablation of ventricular tachycardia from coronary sinus in congenitally corrected transposition of great arteries

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Abstract: Congenitally corrected transposition of the great arteries (ccTGA) compromises less than 1% of all congenital heart diseases, where the RV is the systemic ventricle. It can be associated with heart block, twin AV nodes and accessory pathway connections. Idiopathic Ventricular tachycardia (VT) is not common, with only few reported cases, and they were scar related. Previously reported cases of VT ablation from coronary venous sinus (CVS) are in structurally normal hearts. We performed a VT ablation in ccTGA patient from the CVS, resulting in symptomatic improvement and a decrease in PVC burden from 35% to less than 1%. CVS should be considered as a potential site in ccTGA patients especially when PVCs have RBBB morphology and superior axis.

Keywords: VTach, ventricular tachycardia, transposition, TGA, ccTGA, congenitally corrected, coronary sinus

Introduction

Congenitally corrected transposition of the great arteries (ccTGA) is a rare cardiac defect that compromises less than 1% of all congenital heart defects [1, 2]. This condition is typically diagnosed by echocardiogram, and is characterized by discordant atrio-ventricular and ventriculo-arterial connections, making the RV the systemic ventricle [3]. Given that the blood flow is physiologically "normal", such congenital anomaly can be treated medically either by a non-surgical approach where the RV continues to function as the systemic ventricle, or by undergoing a double-switch operation (DSO); where the great arteries are switched to establish ventriculo-arterial concordance and atrial baffling, or atrial switch, establishes atrioventricular concordance [4]. The non-surgical approach, although an attractive initial option, carries the risk for long-term complications due to right ventricular failure. On the other hand, the DSO is a complex operation that contains multiple incisional sutures, so it might promote the development of atrial arrhythmias [5].

The majority of individuals with ccTGA have associated cardiac defects with ventricular septal defects being the most common, but it can also occur in isolation [1, 2]. The course of the conduction system in ccTGA patient is also abnormal, with malarrangement of the atrioventricular (AV) node, bundle of His and bundle branches, which is believed to be the reason for the high incidence of conduction disturbances [6, 7]. ccTGA can be associated with heart block, twin AV nodes and accessory pathway connections. Ventricular tachycardia can occur later in life associated with poor systemic right ventricular function and can be scar based. Monomorphic idiopathic ventricular tachycardias are infrequent [1, 2, 6]. Ventricular tachycardia can be treated medically and with catheter ablation. Medical treatment needs to be continued on a prolonged basis and may not be effective and can be associated with side effects. To our knowledge, only few cases were reported of VT ablation in ccTGA patients, all of which were ablated from the right ventricle (RV) or left ventricle (LV). In this report, we present a unique case of VT in a 41-year-old ccTGA patient who has not had any prior surgical intervention, with ectopic ventricular focus arising from the basal septal aspect of systemic right ventricle, which was ablated from endocardial aspect of morphologic right ventricle and from epicardial

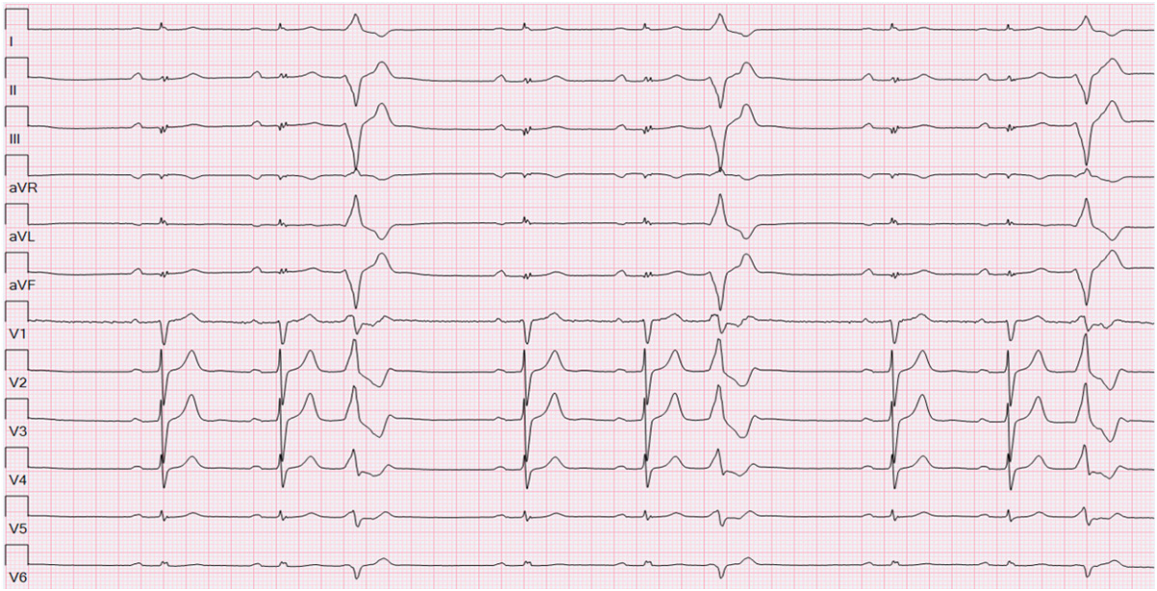

Figure 1. PVCs with right bundle branch block morphology and superior axis.

aspect of same ventricle through the coronary venous sinus (CVS).

Case presentation

Our patient is a 41 y.o. male with corrected transposition of great arteries and Ebsteinoid tricuspid valve (TV) who has not had any cardiac surgical intervention. He had a history of palpitations as teenager and has a baseline 1st degree AV block. He later presented complaining of more frequent palpitations with decreased exercise tolerance. Holter monitor recording showed 1st degree heart block, monomorphic NSVT and high PVC burden (up to 35%) with RBBB morphology and superior axis (**Figure 1**). A cardiopulmonary exercise stress test also showed frequent PVCs and frequent runs of NSVT.

Catheter ablation procedure was started under general anesthesia. At baseline, there were no premature ventricular contractions noted. The anesthesia was lightened, and the patient was extubated. Epinephrine infusion was started, and frequent premature ventricular contractions appeared. Two deflectable quadripolar electrode catheters were introduced in the right femoral vein and in the right femoral artery and used for creating geometry and for mapping purposes. The EnSite™ NavX™ system (St. Jude Medical, St. Paul, MN, USA) was used for catheter navigation, three-dimensional (3D) mapping and activation mapping. From the right femoral vein, the right-sided morphological left ventricle was accessed, and detailed geometry was created. When premature ventricular contractions were induced, the electrograms from the subpulmonic LV were late. From the right femoral artery, through a retrograde approach, the catheter was introduced into the left-sided, systemic right ventricle. Earliest ventricular electrograms were obtained along the basal septal area of the systemic morphologic RV, with local electrograms being nearly 30-40 milliseconds earlier than surface QRS. At that point, the diagnostic catheter was removed, and Saint Jude Flexibility irrigated tip catheter was introduced and used for the purposes of mapping and ablation. Ablation at the sites of earliest ventricular electrograms resulted in suppression of the premature ventricular contractions, but there was recurrence. The power was increased to a maximum of 35 Watts and still there was recurrence.

Mapping was performed in the coronary sinus, as the ablation sites from the left side were close to the atrioventricular valve annulus (**Figure 2**). Electrograms in the proximal coronary sinus, specifically in the proximal portion of the posterolateral coronary vein, were Earliest; nearly 50-60 milliseconds earlier than surface QRS (**Figure 3**). Ablation at these sites was successful in eliminating the PVCs. There

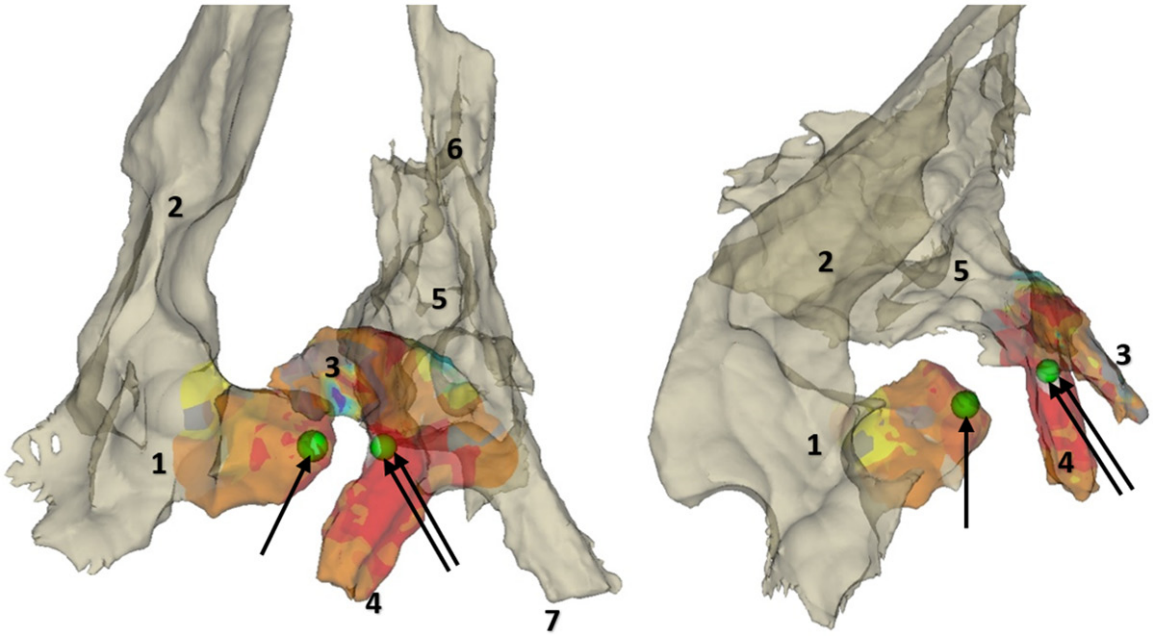

Figure 2. 3D mapping and ablation sites. 1: RV, 2: Aorta, 3: CS, 4: Posterolateral coronary vein, 5: RA, 6: SVC, 7: IVC, Single Arrow: Ablation lesion at site of earliest electrogram obtained from RV, Double Arrow: Ablation lesion at site of earliest electrogram obtained from CS.

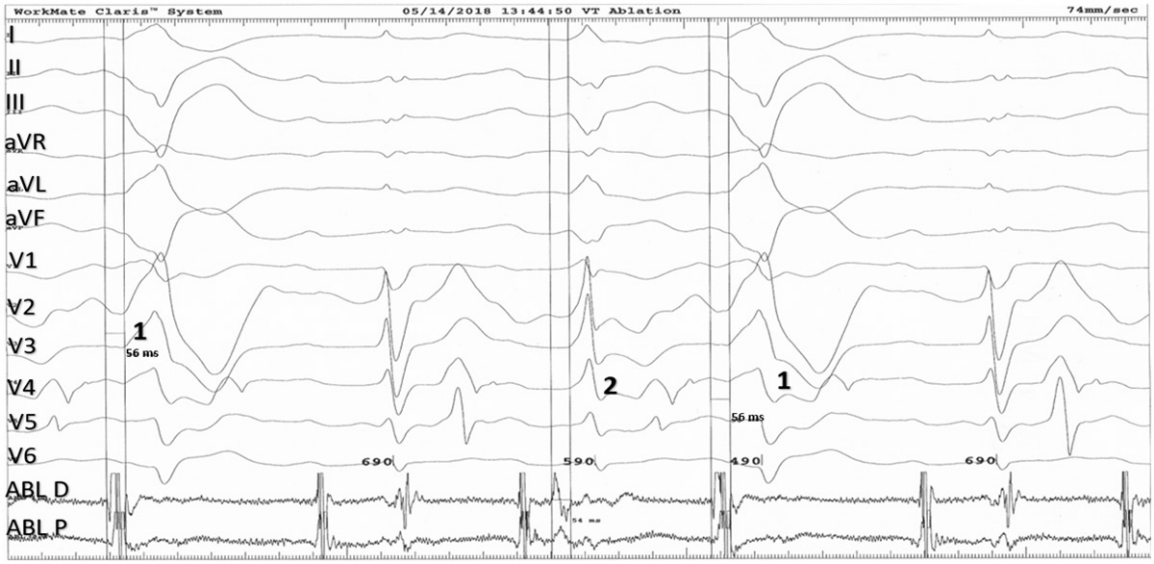

Figure 3. Intracardiac electrograms from successful ablation site in CS. Local electrograms 56 ms earlier than surface QRS. 1: PVC, 2: PVC with fusion.

was nearly 1 cm distance from the endocardial to the epicardial sites. Because of rapid drops in impedance, the maximum power that could be delivered safely in the coronary sinus was 20 Watts. Following ablation, a total wait period of 40 minutes was allowed, and there were no premature ventricular contractions noted. Total effective lesions were 15 and total ablation time was 11 minutes. At his 2-year follow up, the patient remains symptom-free with PVC burden of <1% on Holter monitor.

Discussion

Patients with ccTGA, even those with no surgical intervention, are vulnerable to arrythmias

due to different factors. The course of the conduction system in such patients is frequently abnormal, with malarrangement of the AV node, bundle of His and bundle branches. Atrioventricular blocks and supraventricular arrhythmias are the most common rhythm disturbances noted [1, 2, 6]. As young patients with ccTGA have low incidence of ischemia and fibrosis compared to adults, ventricular arrhythmias usually does not appear at early age but rather on follow-up, as the pressure and volume overload on the systemic RV causes ischemia and fibrosis after a period of time, and the myocardial stretching result in hypertrophy and dispersion of myocardial refractoriness [1, 2]. Surgical intervention and scarring make it more likely to develop arrythmias as well. Two cases of VT ablation in ccTGA patients were related to postsurgical scarring. In a 33 year-old ccTGA patient reported by Toyahara et al., a successful ablation of VT with a LBBB and inferior axis QRS morphology was achieved by ablating around the VSD patch scarring site, from both left and right ventricles [5]. Another case was reported by Baral et al. in a 48-year-old with ccTGA and Tricuspid valve replacement, with VT having RBBB Morphology, and originating from an area of extensive scarring in the posteroinferior region of the systemic RV.

In our case, the VT focus originated from the systemic right ventricle at posterior septal inflow area, and in close proximity to the posterolateral branch of coronary sinus. The endocardial aspect of morphologic RV was ablated in retrograde fashion from the aorta, and then the epicardial aspect of the same ventricle was ablated through the CVS. Electrograms from the epicardial aspect from coronary sinus were earlier than endocardial electrograms. A study by Baman et al., showed that 15% of Idiopathic VT in structurally normal hearts was epicardial in origin, and 70% of those could be ablated through the CVS [8], while another study by Mountantonakis et al., showed that only 9% of the idiopathic ventricular arrythmias in patients with structurally normal heart were linked to the CVS, with a similar successful ablation rate of 70% [9]. This is the first reported case of ablation of VT from within CVS in ccTGA.

The ablation procedure was performed using 3D electroanatomic mapping system and without the use of fluoroscopy, which helps reduce the radiation exposure of both patients and operators.

Conclusion

Most cases of VT in ccTGA patients are related to scar formation after surgical repair. In cases of idiopathic VT in ccTGA, CVS can be a potential site of origin and should be carefully studied. It is important to closely evaluate the morphology and axis of the QRS complexes for PVCs and VT, as a RBBB morphology with superior axis can point towards the CVS being the origin of the ectopy.

Disclosure of conflict of interest

None.

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