Successful ablation of a superior fast-slow atrioventricular reentrant tachycardia in a patient with congenitally corrected transposition of great arteries

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Introduction

Congenitally corrected transposition of the great arteries (ccTGA) is a rare defect characterized by a combination of atrioventricular (AV) and ventriculoarterial (VA) discordance.1 ccTGA is associated with various arrhythmias, such as atrioventricular reciprocating tachycardia (AVRT), intratrial reentrant tachycardia, and atrioventricular nodal reentrant tachycardia (AVNRT).2,3 The {I,D,D} (I, D, D-situs inversus, D-loop, D-transposition) subtype of ccTGA, which is the least prevalent, is frequently associated with AVNRT because of an abnormal conduction system.4 Only a few case reports of catheter ablation of AVNRT in a subset of patients have been published owing to the low prevalence of the {I,D,D} type of ccTGA, especially in patients with anatomical repair.2,5 Recently, the superior slow pathway, which is a variant of the slow pathway originating from the compact node and extending superiorly outside of Koch’s triangle, has been reported in the general population.6 We report a successful case of superior fast-slow AVNRT ablation in a patient with ccTGA who underwent a double-switch operation.

Case report

A 56-year-old woman with {I,D,D}-type ccTGA was referred to our hospital for the management of regular tachycardia. At the age of 6, she was diagnosed with ccTGA with ventricular septal defect and pulmonary stenosis. She underwent combined Senning and Rastelli operations at 34 years of age. The details of the cardiac anatomy are shown in Supplemental Figure 1. Baseline 12-lead electrocardiogram showed sinus rhythm with a right bundle block morphology, left axis deviation, and no evidence of pre-excitation. During palpitations, the 12-lead electrocardiogram showed a long RP tachycardia (140 beats per minute) with the same QRS morphology as the sinus (Figure 1).

An electrophysiological study was performed under general anesthesia using a 3-dimensional mapping system (CARTO system; Biosense Webster, Diamond Bar, CA). Two 6-polar catheters were placed in the high right atrium and apex of the right ventricle (RVA) through the left inferior vena cava. A 10-polar catheter was placed at the coronary sinus via the left superior vena cava. A 4-polar catheter

KEY TEACHING POINTS

- Congenitally corrected transposition of the great arteries (ccTGA) is a rare defect that is often associated with a variety of atrial arrhythmias.
- The {I,D,D} (I, D, D-situs inversus, D-loop, D-transposition) subtype of ccTGA, which is the least prevalent, is likely to be associated with atrioventricular nodal reentrant tachycardia (AVNRT) because of an abnormal conduction system.
- Only a few case reports of catheter ablation of AVNRT have been reported owing to the low prevalence of the subset of these patients.
- Superior fast-slow AVNRT has not been reported in patients with {I, D, D}-type ccTGA who underwent double-switch operation and can be treated with radiofrequency catheter ablation.

KEYWORDS Congenitally corrected transposition of the great arteries, D-transposition; {I,D,D} (I,D,D-situs inversus, D-loop); Radiofrequency catheter ablation; Senning and Rastelli operations; superior fast-slow atrioventricular nodal reentrant tachycardia (Heart Rhythm Case Reports 2021;7:698–701)
was placed at the posterior septum of the tricuspid valve. The His bundle potential was recorded at the anteroseptal aspect of the right AV valve. The dual AV nodal pathway was identified using programmed single atrial extrastimuli. The QRS morphology did not change with atrial pacing at several sites, demonstrating neither antegrade conduction of accessory pathways nor twin AV nodes. The retrograde conduction had decremental conduction properties.

A long RP tachycardia with a cycle length of 440 ms was reproducibly induced via RVA pacing with V-A-V response. The AH and HA intervals during tachycardia were 170 and 262 ms, respectively. The earliest atrial activation site during tachycardia was recorded superior to the His bundle region. The atrial activation sequence during tachycardia was identical to that during RVA pacing. Reset of supraventricular tachycardia by His refractory ventricular stimuli was not observed. The exclusion of atrial tachycardia was determined by the delta-VA intervals of 8 ms (14 ms) with differential atrial overdrive pacing during the tachycardia and termination of tachycardia by ventricular pacing without atrial capture. The exclusion of AVRT was confirmed by ventricular pacing during tachycardia, demonstrating stimulus-atrial interval – VA interval = 145 ms (>85 ms) and postspacing interval – tachycardia cycle length = 242 ms (>115 ms).

VA block during RVA pacing was demonstrated with the administration of adenosine triphosphate (20 mg). The tachycardia was diagnosed as a superior fast-slow AVNRT, including the superior slow pathway. Catheter ablation was performed using a 4.0-mm-tip ablation catheter (Biosense-Webster, Inc, Diamond Bar, CA). Stimulus-His interval was measured on the right atrium to disclose the tract of the antegrade fast pathway before catheter ablation. The distance between the His bundle and earliest atrial activation site during right ventricle pacing was 11.5 mm (Figure 2A). Initially, the antegrade slow pathway at the midseptum from the right side was targeted with the electrogram-guided anatomic approach (the target sites with the ratio of the atrial to ventricular electrogram amplitudes of 0.1 were considered optimal) (Figure 2B). Junctional beats appeared during ablation. However, tachycardia remained inducible; therefore, ablation was attempted at the earliest atrial activation site during RVA pacing (Figure 3A and 3B). After catheter ablation, retrograde conduction of the slow pathway disappeared. Retrograde conduction of the fast pathway was demonstrated only after administration of isoproterenol; however, tachycardia could not be induced (Supplemental Figure 2). The AVNRT did not recur during a 1-year follow-up after the catheter ablation.

Discussion
Atrial arrhythmias are common in adults with congenital heart disease, with a prevalence of 15.1%. AVNRT accounts for 8% of clinical supraventricular tachycardias in adults with repaired congenital heart disease. According to the previous reports, although the prevalence of atypical AVNRT was higher compared to that of the general population, there was no association between the type of AVNRT and anatomic lesion. To the best of our knowledge, this is the first case report of successful catheter ablation of a superior fast-slow AVNRT in a patient with (I,D,D)-type ccTGA who underwent a double-switch operation.

The existence of an atypical fast-slow AVNRT, including a superior pathway with slow conductive properties, has been reported in the general population. A superior slow pathway is suspected when the earliest retrograde atrial activation
appears reproducibly in the His bundle region, followed by antegrade conduction over a fast pathway. The superior fast-slow AVNRT was characterized by confirmation of the presence of a superior slow pathway, long RP tachycardia with the earliest site of atrial activation at the His region, and exclusion of atrial tachycardia and AVRT. However, the origin of the superior slow pathway remains unclear. Inoue and colleagues reported superior extensions of the human compact AV node, which seemed to be involved in superior slow pathway conduction. Kaneko and colleagues hypothesized that a primitive substrate of the slow pathway, including node-like cells, was present between the retroaortic

![Figure 2](image1.png)

**Figure 2** The stimulus-His mapping and atrial activation mapping during right ventricular pacing. A: Stimulus-His mapping. The interval between the stimulus and the onset of the largest His potential deflection (St-H interval) was measured. The antegrade fast pathway was defined as the site with the minimum St-H interval (light purple is the shortest and red is longest St-H interval). B: Atrial activation mapping during right ventricular pacing. Left panel shows the earliest atrial activation site (red arrows) during right ventricular pacing. Right panel shows the earliest activation potential mapped by decapolar catheter. LAO = left anterior oblique; LAT = local activation time; RAO = right anterior oblique; RV = right ventricle.

![Figure 3](image2.png)

**Figure 3** The electrogram recording and catheter position at successful ablation site. A: Successful ablation site. The local electrogram recorded at ablation catheter (ABL) preceded the atrial potential of His by 12 ms. B: Catheter position of successful site and systemic ventricular atrium angiography. The distance between His bundle and the earliest atrial activation site during right ventricle (RV) pacing was 11.5 mm. C: Junctional beats appeared during catheter ablation. CS = coronary sinus; HRA = high right atrium; LAO = left anterior oblique; RAO = right anterior oblique; SVC = superior vena cava; TV = tricuspid valve.
node behind the noncoronary aortic cusp and superior nodal extensions. In the present case, the existence of a superior slow pathway was confirmed by the successful elimination of VA conduction after ablation of the atrial exit near the His bundle.

The AV node locations in patients with congenital heart disease seem less predictable. When planning ablation, the location of the AV node should be considered to avoid inadvertent AV block due to ablation. The AV node is usually displaced to a superior location medial to the right appendage in the majority of (S,L,L) patients. On the contrary, in case of (I,D,D), the connecting AV node is posteriorly situated at the apex of the left Koch triangle, in a mirror-image position compared to the regular AV node.14 The anatomical locations of the AV nodal pathways might be distorted after a double-switch operation, which may create challenges in the determination of the precise location of the AV node and slow pathway. Slow pathway modification in patients with (I,D,D) have been described previously; however, unsuccessful ablation on both sides of the septum with transient AV block has also been reported.12 In the present case, the antegrade slow pathway ablation targeted with the electrogram-guided anatomic approach could not eliminate tachycardia. Demonstrating antegrade conducting fast pathway based on the shortest stimulus-His interval was useful to avoid an AV block during conventional slow pathway ablation.9 Cryoablation might be a useful option to avoid the risk of catheter-induced AV block. Toyohara and colleagues9 reported successful cryoablation at the site of the earliest atrial activation during tachycardia using a transaortic approach in patients with ccTGA who underwent double-switch operation.

Conclusion
In conclusion, this case highlighted successful ablation of superior fast-slow AVNRT in a patient with ccTGA who underwent a double-switch operation. Despite the difficulties in catheter ablation of AVNRT in patients with ccTGA, a precise diagnosis of AVNRT and anatomical evaluation could improve the outcome of catheter ablation.

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Appendix
Supplementary data
Supplementary data associated with this article can be found in the online version at https://doi.org/10.1016/j.hrcr.2021.07.008.

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