

## Unusual Atrioventricular Reentry Tachycardia in Congenitally Corrected Transposition of Great Arteries A Novel Site for Catheter Ablation

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### Presentation

A 19-year-old male with congenitally corrected transposition of the great arteries (ccTGA; S, L, L–situs solitus, l-loop, l-transposition)<sup>1,2</sup> presented with recurrent frequent palpitations, fatigue, and effort intolerance. Previous history was significant for initial presentation with supraventricular tachycardia (SVT) at the age of 14 years. Transthoracic echocardiography at that time led to diagnosis of ccTGA. He reportedly underwent a posteroseptal accessory pathway ablation, and subsequently a second ablation the same year for recurrent SVT, again targeting a posteroseptal pathway. At the age of 17 years, he had a third procedure and reportedly linear ablation between the right atrioventricular valve (mitral), and inferior vena cava was performed for inducible right atrial flutter. Physical examination was unremarkable except for loud aortic closure on cardiac auscultation. The baseline ECG showed the presence of septal Q waves and the absence of lateral Q waves, findings characteristic of ccTGA (Figure 1A).<sup>1</sup> The chest radiograph showed mesocardia without cardiomegaly or pulmonary congestion. Transthoracic echocardiography showed ccTGA, no atrial or ventricular septal defect, no pulmonary stenosis, normally functioning competent atrioventricular valves, and preserved function of both ventricles. Ambulatory monitoring revealed tachycardia, sometimes regular but at other times with an alternating variation in QRS axis and RR interval (Figure 1B). He was brought to the electrophysiology laboratory, and catheters were positioned in standard positions, including high right atrium, His-bundle, subpulmonic ventricle, and coronary sinus (CS). Dissociated signals were noted in the posteroseptal region/CS ostium, presumably related to previous ablations. With premature atrial beats, a distinctly different QRS complex with loss of notching in the inferior leads and a slightly more superior axis was noted. This second more superiorly directed QRS morphology was associated with a shortening in the recorded HV interval. There was no VA conduction. SVT was easily inducible with atrial extrastimuli and terminable with atrial burst pacing. During SVT, there were more atrial than ventricular complexes (2:1 or 3:2 with alternating QRS axis) (Figure 2), and 1:1 atrioventricular conduction during SVT was achieved with isoproterenol intravenous infusion 1 to 2  $\mu\text{g}/\text{min}$ . What is the differential diagnosis and how will you approach mapping and ablation in this case?

### Initial Considerations

There is a possibility for the presence of 2 atrioventricular conduction axes with 2 distinct atrioventricular nodes and His bundles in this case with ccTGA. It is important to recognize that any maneuvers that distinguish between different mechanisms of SVT like atrioventricular nodal reentry tachycardia (AVNRT), atrial tachycardia, or junctional tachycardia based on relationship between His bundle/ventricular activation with atrial activation will be unreliable because of the presence of 2 separate His bundles. As an example, AVNRT or junctional tachycardia originating from 1 of the 2 atrioventricular nodes and conducting to the ventricle using the other atrioventricular node–His bundle, would seem like an atrial tachycardia if the His bundle that is passively conducting to the ventricle is being mapped.

Abrupt onset and offset of SVT with pacing maneuvers in this case was suggestive of a reentrant mechanism. During tachycardia, fewer ventricular than atrial complexes effectively excluded atrioventricular reentry, including reentry using an atriofascicular (Mahaim) connection. The differential thus included AVNRT, atrial tachycardia, or junctional tachycardia. An automatic junctional tachycardia was felt to be unlikely because of the abrupt onset and offset of tachycardia with atrial pacing maneuvers, more A than V during tachycardia with underlying VA conduction block, and an AHA response with the last atrial overdrive pacing impulse. Entrainment of the tachycardia showed postpacing intervals much longer than the tachycardia cycle length from the right and left atrial–free walls. The shortest postpacing intervals were obtained when entrainment was performed from the interatrial septum (postpacing interval–tachycardia cycle length  $\approx 20$  ms when atrial overdrive pacing 10 ms shorter than tachycardia cycle length). Furthermore, an activation map during tachycardia with point-to-point mapping of both atria (Carto 3 system, Biosense Webster) showed the earliest site of activation at the midseptal region of the putative fast pathway exit to the atrium. Furthermore, only 100 ms of the 340 ms tachycardia cycle length was mapped. In summary, this is a reentrant SVT close to the septum, with a large part of the cycle length not registered with complete mapping; and the earliest site at the fast-pathway exit of the atrioventricular node. Although a septal atrial tachycardia cannot be conclusively excluded, these findings are highly suggestive of AVNRT.<sup>3</sup>

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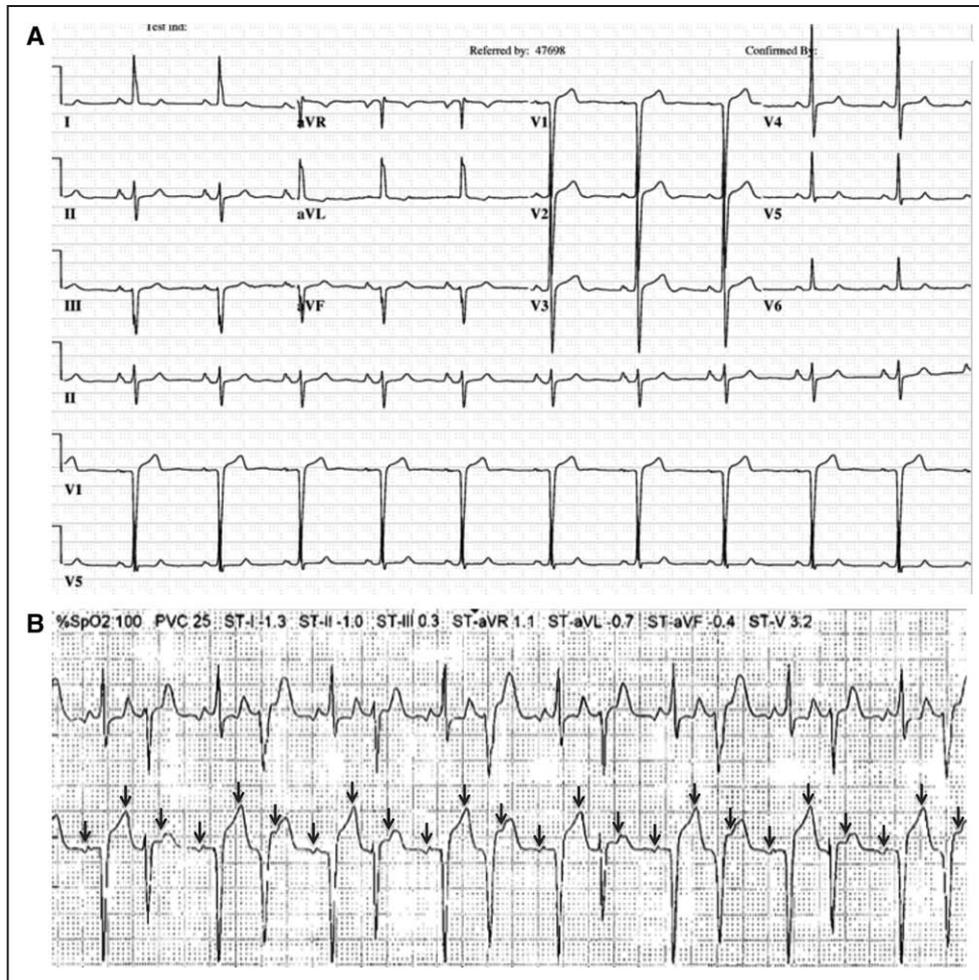
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**Figure 1.** A, Baseline 12-lead ECG. B, Ambulatory rhythm monitoring showing narrow complex tachycardia. Alternating 2 QRS complexes with different axis for every 3 P waves (arrows) are seen.

To target the likely AVNRT, empirical right-sided slow pathway ablation was performed using irrigated 3.5-mm tip radiofrequency catheter (ThermoCool, Biosense Webster, Diamond Bar, CA). Because of continued tachycardia, despite extensive ablation in the right posteroseptal region, the left slow pathway was targeted with ablation in the CS 1 to 2 cm from the ostium, and transeptally at the left posteroseptal region, where junctional ectopic beats were induced. The tachycardia continued to be easily inducible. Ablation at the earliest site of activation at the midseptal region was not attempted because of the risk of atrioventricular block. The procedure was abandoned because of prolonged procedural duration and rescheduled for 2 months later. The patient continued to have frequent runs of symptomatic SVT.

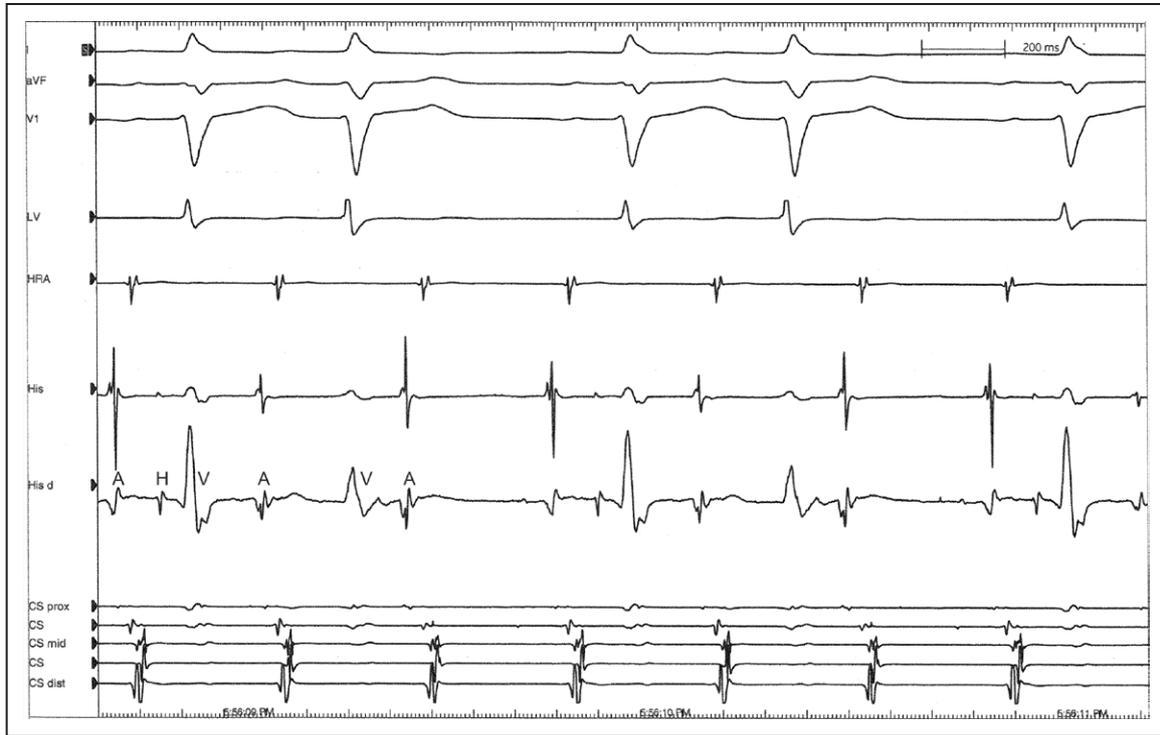
### Further Considerations

At the 2-month follow-up procedure, intracardiac catheters were again positioned in the routine locations, and the same SVT was easily inducible with pacing, with the same site of earliest atrial activation at right and left midseptum (Figure 3). Irrigated radiofrequency catheter ablation to empirically ablate the slow pathway was again attempted in the right posteroseptal, CS, and left posteroseptal and posterior annular (along CS

course) regions but was unsuccessful. Two distinct His bundle recordings were identified on the right septum: 1 anteroseptal and 1 midseptal. The respective His bundle activations were associated with the 2 distinctive QRS morphologies/axes. How should one approach this difficult case of AVNRT?

### Conduction System Anatomy in ccTGA

A fundamental understanding of the anatomy of the atrioventricular conduction system is crucial for any case of SVT or suspected AVNRT in patients with ccTGA.<sup>4</sup> The conduction axis develops<sup>5</sup> within the constraints of l-looping of the cardiac tube during embryonic development and l-transposition of the great arteries. L-looping causes malalignment of the interventricular septum with the interatrial septum, often with a membranous ventricular septal defect. Barring a minority of cases, the atrioventricular-nodal tissue in the midseptal region fails to connect to the ventricles. L-transposition is associated with a central location in the heart of the pulmonary outflow tract with the proximal pulmonary artery coursing adjacent to the interatrial septum, whereas the aorta is anteriorly displaced. Atrioventricular conduction in ccTGA typically occurs via an anteriorly displaced atrioventricular node and His bundle. The anterior atrioventricular node is situated at the region of confluence of the base of the right atrial appendage with the mitral annulus (right



**Figure 2.** Intracardiac electrograms during atrioventricular (AV) nodal reentry tachycardia with 3:2 atrioventricular conduction. His catheter is mapping the anterior His bundle, left ventricular catheter is in the morphological left ventricle. First atrial complex (A) is followed by an expected His deflection (H) and ventricular electrogram (V). The second atrial electrogram conducts to the ventricle without activating the anterior His bundle by using the posterior AV conduction axis (not shown) with a more leftward QRS axis. The third atrial complex blocks conduction to ventricles in both AV nodes.

sided). From here, the His bundle courses along the lateral margin of the pulmonary outflow tract and anterior to the membranous ventricular septal defect when present (as opposed to the His-bundle course posterior to membranous ventricular septal defect in d-looping) (Figure 4).<sup>4</sup> This anterior atrioventricular node can retain a connection to the posterior atrioventricular node with a sling of conduction tissue (Mönckeberg sling) along the atrioventricular annular region, especially when the interventricular septum is aligned with the atrial septum in the presence of pulmonary atresia.<sup>6</sup>

### Practical Construct for AVNRT Ablation

AVNRT involves a reentrant circuit with conduction through some or all of the atrioventricular node, linking 2 spatially and functionally disparate atrial connections and the atrial tissue in proximity to the atrioventricular node completing the circuit. To successfully ablate AVNRT, we need to target the accessory atrial input to the atrioventricular node and avoid the fast pathway and the compact atrioventricular node to prevent atrioventricular block.<sup>3</sup>

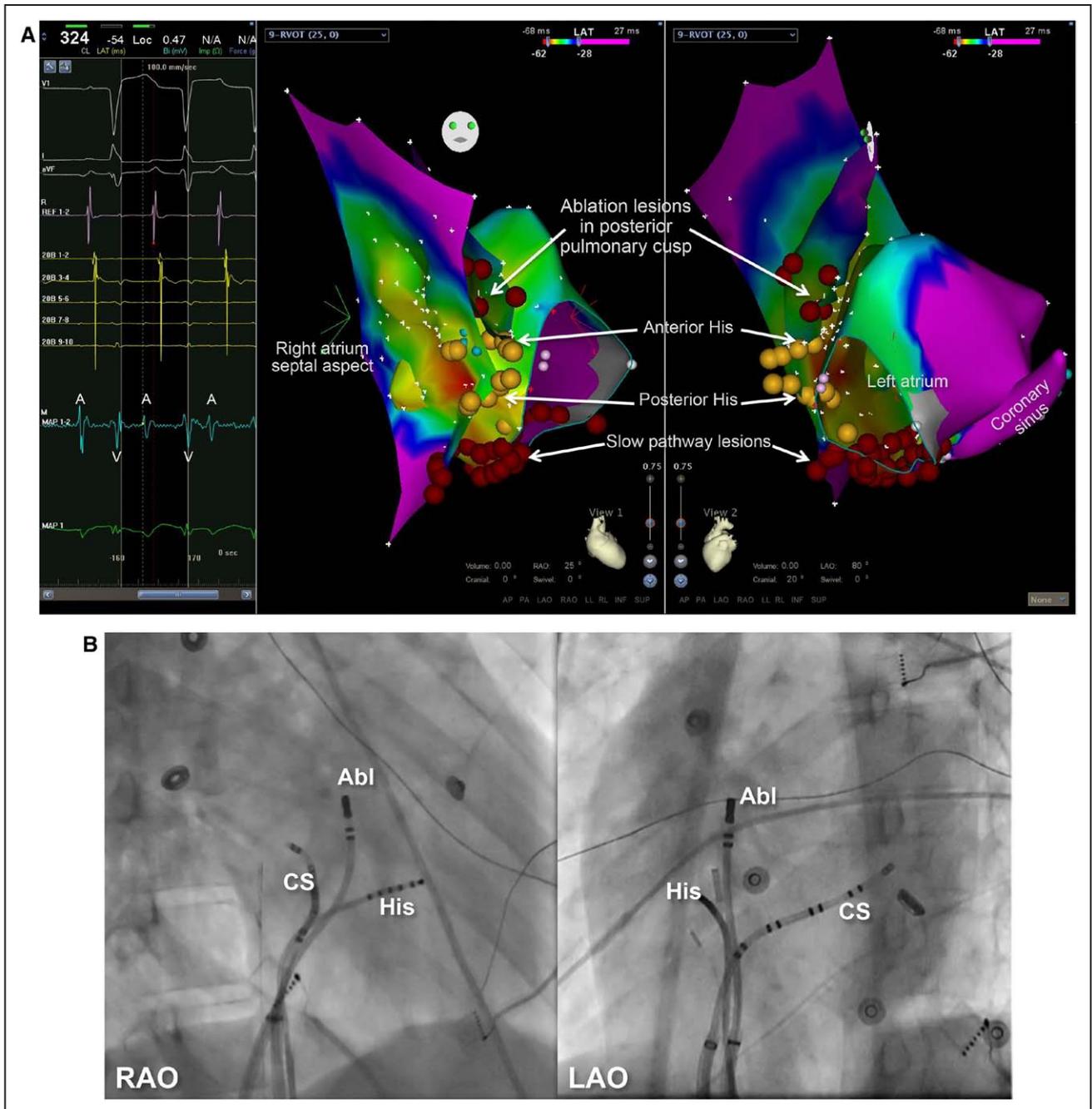
### Reentry Using Atrioventricular Nodal Tissue in ccTGA

In ccTGA, both atrioventricular nodes can have  $\geq 1$  discrete atrionodal connections, and both nodes can be connected to each other via a Mönckeberg sling. Reentry using atrioventricular node tissue can thus occur using one of many possible permutations for antegrade and retrograde limbs between atria and atrioventricular node. Furthermore, electric activation can propagate antegrade down one His bundle and climb back up the other to set up inter-atrioventricular node reciprocating tachycardia.<sup>7</sup>

This last possibility is excluded in this case because of the absence of 1:1 relationship between atrial activations and His (more A than His). Also demonstrated in this case, is antegrade activation of both His bundles, rather than in a reciprocating fashion. The posterior connections to the atrioventricular node (right and left slow pathways) had been targeted with extensive ablation on both sides of the septum, and targeting the site of earliest atrial activation (fast pathway–retrograde limb) is not suitable because of high risk of antegrade atrioventricular block. Presumably, if this is AVNRT involving the anterior atrioventricular node with the retrograde connection to the atrium in the midseptum, where should one look for the second (antegrade) atrial connection to this atrioventricular node?

### The Missing Atrionodal Connection

In ccTGA, the root of the pulmonary artery abuts the interatrial septum and is contiguous with the right atrioventricular/mitral valve, where the anterior atrioventricular node is situated. Atrial tissue along the posterior aspect of proximal pulmonary artery can form a distinct (anterior) connection to the atrioventricular node (Figure 4). This atrial input can be targeted in the pulmonary artery cusps (this is not too dissimilar to ablation, in a developmentally normal heart, of the aortic noncoronary and left coronary cusp to target the anteroseptal, and perimitral atrial tissue).<sup>3,8</sup> Irrigated radiofrequency ablation of atrial signals at this location with careful monitoring of atrioventricular conduction and gradual titration of power to 30 W resulted in slow junctional ectopic beats with subsequent elimination of AVNRT. Conduction was preserved over both atrioventricular



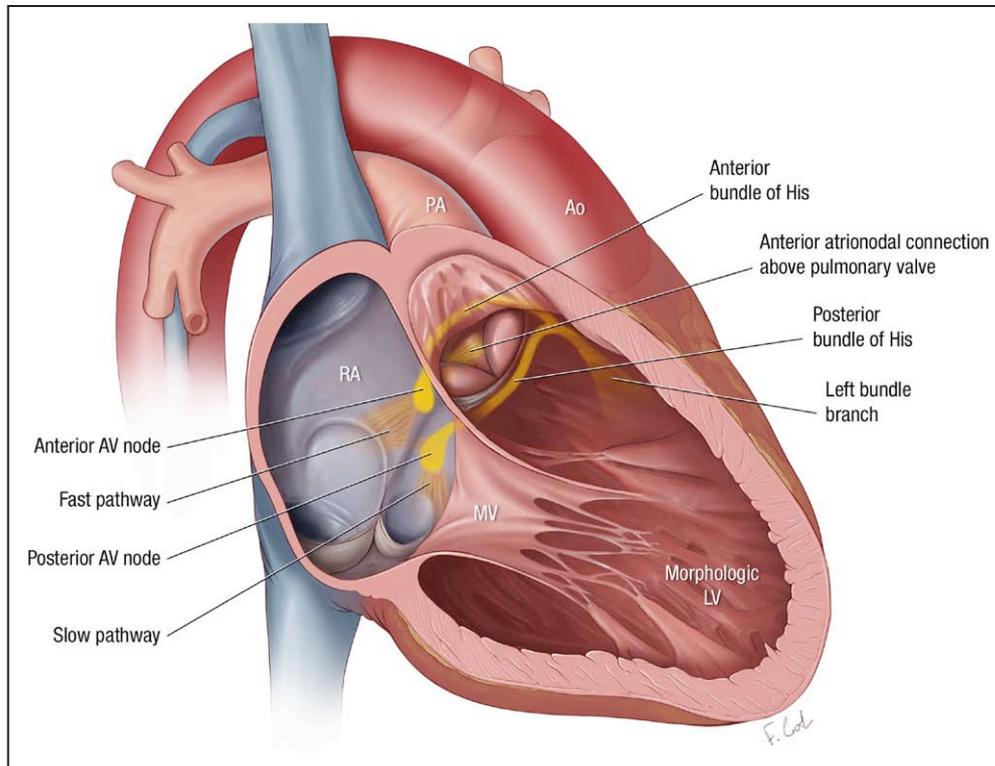
**Figure 3.** **A**, Electroanatomic activation map of the right atrium (septal aspect), part of the left atrium, coronary sinus (CS), and the posterior part of the pulmonary artery root. Earliest activation (red) is seen at the interatrial septum at the region of the fast pathway connections to the atrioventricular (AV) nodes. The presumptive second atrionodal connection to the anterior AV node was ablated in the posterior pulmonary cusp. Atrial signals targeted for ablation in this region are seen on the electrogram labeled MAP 1–2 in the left hand panel (A, atrial; V, ventricular). **B**, Fluoroscopic projection of the ablation catheter (Abl) at successful site of ablation in the posterior pulmonary artery root. Catheters mapping the CS and the posterior His bundle (His) are also seen. LAO indicates left anterior oblique; and RAO, right anterior oblique.

node–His axes. The patient has not had recurrent palpitations during 2 years of follow-up and has reported much improved levels of alertness, school performance, and physical capacity.

### Discussion

Unusual forms of AVNRT can occur in ccTGA in the context of 2 atrioventricular nodes, both of which have the opportunity for reentry using separate atrial connections. Both atrioventricular nodes can be connected by a Mönckeberg sling and participate together in reentry using an atrial

connection to either of the atrioventricular nodes. Twin atrioventricular conduction axes (both atrioventricular nodes are well developed and connect to ventricles through their respective His bundles) occur in a minority of ccTGA cases.<sup>4</sup> Inter-atrioventricular nodal reciprocating reentry can occur with both His bundles participating in reentry—1 antegrade and 1 retrograde. Atrioventricular conduction during sinus rhythm or SVT may propagate to the ventricle using either of the 2 atrioventricular node–His axes. In addition to ccTGA, duplication of the atrioventricular node with or without a Mönckeberg sling connecting the anterior and posterior



**Figure 4.** Illustration showing the conduction system anatomy observed in this case with congenitally corrected transposition of great arteries. The free wall of the right-sided chambers has been cut out to visualize the septal aspect. Note the presence of the duplicate atrioventricular (AV) nodes and His bundles. The atrionodal connections to the posterior AV node occur via the usual slow and fast pathway locations. Atrial fibers interface with the anterior AV node from the usual fast pathway region, but also from above the pulmonary valve along the posterior sinus of Valsalva. This anterior atrionodal connection to the anterior AV node was targeted with ablation to eliminate AV nodal reentrant tachycardia. Ao indicates aorta; LV, left ventricle; MV, mitral valve; PA, pulmonary artery; and RA, right atrium.

atrioventricular nodes can occur in patients with double inlet left ventricle and with levo-looping of cardiac tube in patients with situs ambiguus.<sup>9,10</sup>

This patient had twin atrioventricular conduction axes. The AVNRT circuit presumably involved antegrade conduction to the atrioventricular node using anterior inputs along the posterior pulmonary artery root. The retrograde limb was at the site of earliest atrial activation in the midseptal region. Even though the AVNRT circuit was stable, atrioventricular conduction occurred/blocked in a repetitive pattern to give a 3:2 atrioventricular association with alternating QRS morphology/axis. In fact, atrioventricular conduction predominantly used the anterior atrioventricular node–His bundle during sinus rhythm or slower ventricular rates, however, during 1:1 atrioventricular conduction, during tachycardia the QRS morphology/axis was exclusively that of posterior atrioventricular conduction system. Liao et al<sup>11</sup> reported a series of AVNRT in 9 patients with ccTGA of whom one had twin atrioventricular conduction axes. Similar to this case, conventional slow pathway ablation was unsuccessful. Successful ablation was reported at the right atrioventricular annulus in between the 2 His recordings at a site with a fragmented atrial electrogram, though in our patient we avoided ablation in this region because of proximity to atrioventricular nodes. Even though we were able to successfully ablate the atrioventricular nodal inputs from the posterior aspect of the pulmonary cusps, there is an inherent higher risk of atrioventricular block during AVNRT ablation in ccTGA because of the variability in the atrioventricular conduction axis. In addition to the AVNRT, this patient previously had a posteroseptal accessory pathway and a right atrial flutter ablation,

highlighting the possibility of encountering multiple arrhythmias in patients with congenital heart disease. Further, during invasive electrophysiology study, accessing the CS in patients with ccTGA may be challenging as they may have abnormalities of the CS ostium, including ostial atresia, multiple or displaced CS ostia, with CS draining to right atrium through multiple Thebesian veins.<sup>2</sup>

We propose the following approach to AVNRT ablation in ccTGA:

1. Exclude other mechanisms of SVT using diagnostic electrophysiology maneuvers. Maneuvers have to be interpreted in the context of the possible permutations for reentry involving either/both atrioventricular nodes. For example, AVNRT might seem like atrial tachycardia if there is atrioventricular block at the culprit atrioventricular node with atrioventricular conduction occurring through the other atrioventricular node–His axis.
2. In the event of twin atrioventricular nodes, exclude inter-atrioventricular node reciprocating tachycardia by dissociating His/ventricle from tachycardia or demonstrating antegrade conduction over both His bundles. When inter-atrioventricular node reciprocating tachycardia is present, one of the atrioventricular nodes (posterior atrioventricular node) may have to be ablated after confirming antegrade conduction over the other (anterior) atrioventricular node to avoid atrioventricular block.<sup>7</sup>
3. Map the earliest atrial activation during AVNRT, and if not in the conventional fast pathway position and

sufficiently distant from the anterior atrioventricular node, target this atrionodal extension with ablation.

4. If the earliest atrial activation is in the midseptal/fast pathway region (retrograde limb), maneuvers such as paced resetting responses<sup>12</sup> or subthreshold stimulation<sup>13</sup> can be used to map the posterior and anterior aspects of the septum, the posterior pulmonary cusp and the anterior mitral annulus, to identify the antegrade limb of AVNRT circuit. Otherwise, empirical slow pathway ablation in the postero-septal region and the left slow pathway can be attempted.
5. If AVNRT persists despite adequate ablation in the usual slow pathway regions, atrial inputs posteriorly along the pulmonary artery cusps can be targeted with ablation.

### Conclusions

We describe an unusual case of AVNRT in ccTGA and a novel site for ablation in the pulmonary artery cusp, targeting the anterior atrial inputs to the anterior atrioventricular node. Supraventricular arrhythmias in congenital heart disease patients are often the most challenging cases in invasive electrophysiology and understanding the anatomy is critical. This case highlights the difficulties in mapping and ablating atrioventricular node-related tachycardias and focuses on the interplay between the developmental and structural anatomy of cardiac conduction system with AVNRT arrhythmogenesis.

### KEY TEACHING POINTS

- Patients with certain complex congenital heart disease lesions such as congenitally corrected transposition, double inlet left ventricle, and situs ambiguus with levo-looping of the cardiac tube have the substrate for duplication of the atrioventricular node. Differential diagnosis of SVTs should thus include the possibility of reentry circuits involving any one or both of the atrioventricular nodes.
- In patients with twin/duplicated atrioventricular nodes, AVNRT or junctional tachycardia from either of the atrioventricular node can conduct to the ventricle using the other atrioventricular node–His axis, and traditional maneuvers differentiating atrioventricular node–related tachycardias from atrial tachycardias based on reset responses to ventricular pacing or linking of atrioventricular conducted event to subsequent return atrial beat are unreliable.
- The anteriorly displaced atrioventricular node in l-transposition most commonly seen in context of ccTGA can have spatially dissociated anterior/superior atrionodal connections that can be mapped and ablated from the posterior aspect of the pulmonary artery root.

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### Disclosures

None.

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KEY WORDS: anatomy ■ atrioventricular nodal reentry tachycardia ■ atrioventricular node ■ congenital heart disease ■ supraventricular tachycardia ■ transposition of great vessels

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