Congenitally corrected transposition of the great arteries (CCTGA) is characterized by the combination of atrioventricular (AV) discordance and ventriculoarterial discordance. Because it is physiologically a corrected transposition, the associated anomalies, for example, ventricular septal defect and Ebstein anomaly of the tricuspid valve, commonly complicate the malformation. Furthermore, CCTGA may be associated with a variety of arrhythmias, including AV reentrant tachycardia, atrial tachyarrhythmias, and, rarely, AV nodal reentrant tachycardia (AVNRT) and ventricular tachycardia.2-9 Congenitally corrected transposition of the great arteries (CCTGA) is characterized by the combination of atrioventricular (AV) discordance and ventriculoarterial discordance. Because it is physiologically a corrected transposition, the associated anomalies, for example, ventricular septal defect and Ebstein anomaly of the tricuspid valve, commonly complicate the malformation. Furthermore, CCTGA may be associated with a variety of arrhythmias, including AV reentrant tachycardia, atrial tachyarrhythmias, and, rarely, AV nodal reentrant tachycardia (AVNRT) and ventricular tachycardia.2-9

Background—We sought to investigate the feasibility of radiofrequency catheter ablation of atrioventricular node reentrant tachycardia and the ideal site for slow pathway (SP) ablation in congenitally corrected transposition of the great arteries.

Methods and Results—Nine patients with congenitally corrected transposition of the great arteries referred for catheter ablation of atrioventricular node reentrant tachycardia were studied. A single His potential was recorded in 8 patients (89%, 6 [S,L,L] and 2 [I,D,D]). The earliest atrial activation during retrograde atrioventricular node conduction occurred at His bundle region (HBE; n=7) or shifting from HBE to coronary sinus ostium (n=1, [S,L,L]). Two anatomically separate His potentials were recorded in 1 patient (11%, [S,L,L]), one at the anterosuperoseptal His bundle (HBE-1) and the other at the confluence of the pulmonary and mitral annulus (HBE-2). In 8 cases with a single His potential recorded, SP was ablated at the posterior-midseptum, 2 ([S,L,L]) at the right posteroseptum, 1 ([S,L,L]) at the left posteroseptum, and 5 ([S,L,L] and 2 [I,D,D]) at the midseptum after failure of energy application at the posteroseptum. Junctional rhythm was observed during radiofrequency catheter ablation in all 8 of the cases. In the remaining patient with 2 anatomically separate His potentials recorded, SP was successfully ablated from the confluence of the pulmonary and mitral annulus, slightly below the HBE-2. Junctional rhythm was also induced during radiofrequency catheter ablation.

Conclusions—In [S,L,L] or [I,D,D], radiofrequency catheter ablation of atrioventricular node reentrant tachycardia is feasible. SP input region can mainly be found in the posterior midseptum, especially in patients with single penetrating atrioventricular nodes. SP could usually be successfully ablated in these regions. (Circ Arrhythm Electrophysiol. 2012;5:1143-1148.)

Key Words: ablation a atrioventricular node a supraventricular tachycardia a congenital heart disease

The location of the AV node (AVN) and its continuity with the penetrating His bundle in CCTGA are related to both the cardiac loop and structure lesions. Histopathologic studies and intraoperative mapping studies have demonstrated a displaced anterior location for the AVN and penetrating His bundle far away from the conventional location in majority of [S,L,L] patients. On the basis of these observations, one may postulate that the target area for slow pathway (SP) ablation might also be displaced in patients with [S,L,L]. Two case reports have described that SP could be successfully ablated at the posteroseptal site and right anterosuperoseptal site. However, the ideal sites for catheter ablation of the SP in such patients have not been described in detail. In this study, we report on radiofrequency catheter ablation (RFCA) in 9 cases of AVNRT associated with CCTGA.
Methods

Study Population
Nine patients (5 women; ages 32±14 years [range, 18 to 58 years]) with CCTGA referred for catheter ablation of AVNRT to our centers experienced in ablative therapy were retrospectively analyzed (Table 1). Median age of first supraventricular tachycardia episode was 21 years old (range, 15 to 40 years) and a median of 1 oral antiarrhythmic drug (range, 1 to 3 drugs) had been tried in all 9 of the patients. An electrophysiological study had been performed previously in 2 of the 9 patients. Ablation was unsuccessful with 1 procedure in both patients. Two patients had additional anatomical abnormalities: one had atrial septal defect (ASD) and the other had both ASD and ventricular septal defect. Severity of tricuspid valve insufficiency was mild in 2 patients (25%), moderate in 5 (63%), and severe in 1 (12%), respectively. Left ventricular ejection fraction was 51±6%. One patient (11%) underwent previous surgical ventricular septal defect and ASD closure.

Definitions
Two-dimensional transthoracic echocardiogram and cardiac computed tomography were used in the diagnosis of CCTGA. Anatomical arrangement of {S,L,L} is situs solitus with L-looping of the ventricles and the aorta anterior and leftward of the pulmonary artery, and {I,D,D} is situs inversus with D-looping of the ventricles and the aorta anterior and rightward.1 AVNRT was diagnosed according to standard morphological right atrium, the morphological left ventricular apex, and at the His bundle region (HBE) via the femoral veins. Also, a 6-F multipolar catheter was advanced within the coronary sinus via the internal jugular vein. The stimulation protocol consisted of programmed stimulation at 2 basic cycle lengths with ≤2 extrastimuli and burst pacing at the morphological right atrium and the morphological left ventricular apex.12,13 During single atrial extrastimulus (A1A2) testing, an AH jump was defined as prolongation of ≥50 ms in the A2H2 interval after a shortening of the A1A2 interval by 10 ms. When 2 atrial extrastimuli are delivered, a jump from fast pathway to SP conduction occurred at HBE (7 patients) or to facilitate the tachycardia induction, and the stimulation protocol was repeated.

Radiofrequency Catheter Ablation
RFCA was done using a 4-mm tip ablation catheter (Biosense-Webster, Inc, Diamond Bar, CA). Antegrade SP was attempted to ablate with the electrogram-guided anatomic approach. Initially, the ablation procedure was started at the posterior/inferior aspect of the right- or left-sided mitral annulus anterosuperior to the coronary sinus ostium (CSO) or within the coronas sinus ostium. Target sites with the ratio of the atrial:ventricular electrograms amplitudes between 0.1 and 0.5 were considered optimal. Twenty watts of energy with nontemperature limit were initially applied at successful sites. After 15 seconds, the power was gradually titrated to 30 watts for 30 to 60 seconds. Application of radiofrequency energy was terminated when accelerated junctional beats were not observed within 15 seconds. If this conventional approach failed to eliminate or modify the SP ablation at the midseptal and eventually anteroseptal site was attempted, respectively. If AVNRT remained inducible after 8 to 10 radiofrequency energy applications with junctional beats or rhythm during ablation at the right- or left-sided mitral annulus, the mapping and ablation of SP were switched to the other side.15 In case of an evidence of retrograde SP, the initial mapping was performed in the region between the CSO and the mitral annulus, and ablation was targeted at the site showing the earliest retrograde atrial activation. In all of the patients, ablation was considered successful if neither AVNRT nor ≥1 AV echo beat could be induced even after isoproterenol infusion.

Statistical Analysis
Categorical variables are expressed as numbers and percentages. Continuous values are presented as median and range or mean±SD, as appropriate.

Results
Electrophysiological Study and Ablation Procedures
Among these 9 patients, 7 were diagnosed with {S,L,L} and 2 with {I,D,D} (Table 1). In 1 of 2 patients with {I,D,D}, the coronary sinus catheter was advanced within the coronary sinus with effort. A single His potential was recorded from septal and eventually anteroseptal site was attempted, respectively. If AVNRT remained inducible after ≥8 to 10 radiofrequency energy applications with junctional beats or rhythm during ablation at the right- or left-sided mitral annulus, the mapping and ablation of SP were switched to the other side.15 In case of an evidence of retrograde SP, the initial mapping was performed in the region between the CSO and the mitral annulus, and ablation was targeted at the site showing the earliest retrograde atrial activation. In all of the patients, ablation was considered successful if neither AVNRT nor ≥1 AV echo beat could be induced even after isoproterenol infusion.

Table. Basic Characteristics

<table>
<thead>
<tr>
<th>Case</th>
<th>Sex</th>
<th>Age</th>
<th>Age of First Arrhythmia</th>
<th>Type of CCTGA</th>
<th>TVR Severity</th>
<th>Associated Lesions</th>
<th>Ablation Target</th>
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</thead>
<tbody>
<tr>
<td>1</td>
<td>F</td>
<td>24</td>
<td>19</td>
<td>(S,L,L)</td>
<td>Moderate</td>
<td>None</td>
<td>Slow-fast</td>
</tr>
<tr>
<td>2</td>
<td>F</td>
<td>44</td>
<td>38</td>
<td>(S,L,L)</td>
<td>Moderate</td>
<td>None</td>
<td>Slow-slow</td>
</tr>
<tr>
<td>3</td>
<td>F</td>
<td>19</td>
<td>16</td>
<td>(S,L,L)</td>
<td>Moderate</td>
<td>None</td>
<td>Slow-fast</td>
</tr>
<tr>
<td>4</td>
<td>M</td>
<td>18</td>
<td>15</td>
<td>(I,D,D)</td>
<td>Moderate</td>
<td>None</td>
<td>Slow-fast</td>
</tr>
<tr>
<td>5</td>
<td>F</td>
<td>58</td>
<td>22</td>
<td>(S,L,L)</td>
<td>Severe</td>
<td>ASD, VSD</td>
<td>Left-side variant</td>
</tr>
<tr>
<td>6</td>
<td>M</td>
<td>25</td>
<td>23</td>
<td>(S,L,L)</td>
<td>Moderate</td>
<td>ASD</td>
<td>Slow-fast</td>
</tr>
<tr>
<td>7</td>
<td>M</td>
<td>22</td>
<td>19</td>
<td>(S,L,L)</td>
<td>Mild</td>
<td>ASD</td>
<td>Slow-fast</td>
</tr>
<tr>
<td>8</td>
<td>F</td>
<td>27</td>
<td>21</td>
<td>(S,L,L)</td>
<td>Mild</td>
<td>None</td>
<td>Slow-fast, AP</td>
</tr>
<tr>
<td>9</td>
<td>M</td>
<td>47</td>
<td>40</td>
<td>(I,D,D)</td>
<td>None</td>
<td>None</td>
<td>Slow-fast</td>
</tr>
</tbody>
</table>

TVR indicates tricuspid valve regurgitation; ASD, atrial septal defect; VSD, ventricular septal defect; AP, accessory pathway; CCTGA, congenitally corrected transposition of the great arteries; M, male; and F, female.
Ablation of SP in Patients With \{S,L,L\}
Among 5 (71\%) of 7 patients with \{S,L,L\}, the SPs were successfully ablated at the posteroseptum in 2, one anterior to CSO and the other at the edge of CSO (slow-slow variant); 3 patients crossed over to the midseptum after failure of posteroseptal SP ablation and also had SPs eliminated (Figures 3 and 4). In the patient with slow-slow variant, ablation at the edge of CSO eliminated both antegrade and retrograde SPs. During radiofrequency energy delivery, accelerated junctional beats with retrograde conduction were induced in all 5 of these patients. The QRS morphology of junctional beat was equal to that during tachycardia and sinus rhythm.

In patient No. 8 with 2 anatomically separate His potentials recorded, the electrophysiological study was performed using electroanatomical mapping system (CARTO XP, Biosense Webster). However, with accelerated junctional rhythm (QRS morphology of type A and fusion of type A and type B), conventional right-sided approach failed to ablate the SP (Figure 5). When the ablation catheter was moved to the region slightly below the HBE-2, a fractionated atrial potential was recorded with a small His potential (Figure 1). SP was successful ablated at this site with junctional rhythm (QRS morphology of type B; mean cycle length, 588 ms) observed during radiofrequency ablation (Figure 5). No attempt was made to completely eliminate one of the AVNs.

In patient No. 5, although right-sided approach including midseptum and anteroseptum was performed with effective junctional ectopic beats, AVNRTs were still inducible. At last, SP was successfully ablated from the left-sided posteroseptal aspect of the tricuspid annulus by the transeptal approach (Figures 3 and 4). Junction beats with the same QRS morphology of tachycardia was still observed during radiofrequency ablation at successful left-sided ablation site.

Ablation of SP in patients with \{I,D,D\}
In 2 patients with \{I,D,D\}, although with induced junctional beats, radiofrequency energy delivery at the left-sided posteroseptal aspect of the mitral annulus failed to ablate or modify the SP. When the ablation catheter was moved to the middle point between CSO and HBE, a fractionated atrial potential was recorded with an A/V ratio <0.5 (Figures 3 and 4).
RFCA performed at this site eliminated SP in both patients. A junctional rhythm was also induced during radiofrequency energy application. The QRS morphology during sinus rhythm, tachycardia, and junctional rhythm was equal.

Follow-Up
No procedure-related complication occurred in these 9 patients immediately after ablation and during follow-up. All 9 of the patients were free of arrhythmias without antiarrhythmic drugs during a mean follow-up of 8±4 months (median, 9 months; range, 4 to 12 months).

Discussion
Histologic studies of autopsy specimens have described the presence of twin AVNs in CCTGA. Electrophysiological characteristics of AV reentrant tachycardia mediated by twin AVNs have been well described. Limited data evaluated the feasibility of RFCA and the ideal site of SP in CCTGA. This present study demonstrated that RFCA of AVNRT in {S,L,L} or {I,D,D} is feasible. In these patients, SP input region can mainly be found in the posterior midseptum, especially in patients with single penetrating AVNs. SP could usually be successfully ablated in these regions.

Prevalence of Multiple Congenital Cardiac Abnormalities in CCTGA
In >80% cases, CCTGA is associated with multiple structural lesions including ventricular septal defect and pulmonary stenosis. In addition, the size of pulmonary trunk in CCTGA is associated with the degree of septal malalignment. Cases with well-aligned septums often had atresia or stenosis of pulmonary trunk. However, in our cases, the incidence of multiple congenital cardiac abnormalities is low (22%). The reason for this discrepancy may be related to all of our cases composed of adult survivals. Two case presentations of AVNRT in CCTGA provide further support to our speculation.

AV Conduction System in CCTGA
Among 8 (89%, 6 {S,L,L} and 2 {I,D,D}) of 9 patients in the present study, a single His potential recorded and maintenance of unique QRS morphology during sinus rhythm, tachycardia, and junctional rhythm indicated that a single penetrating AVN existed. All of the single His potentials were recorded from the conventional position in the Koch triangle. Junctional rhythm was induced during RFCA at the posterior midseptum in all 8 of the cases. This finding is in agreement with the pathologic literature and intraoperative mapping data, which suggest that the AVN is typically found in the conventional location in the Koch triangle. Junctional rhythm was induced during RFCA at the posterior midseptum in all 8 of the cases. This finding is in agreement with the pathologic literature and intraoperative mapping data, which suggest that the AVN is typically found in the conventional location in the Koch triangle for {I,D,D} hearts. However, in our 6 cases of {S,L,L}, the course of penetrating His bundle was not mapped, and catheter mapping really does not pinpoint the AVN location. Therefore, we can neither conclude that a normally located posterior AVN gives rise to the single penetrating His bundle nor exclude that the AVN actually is anterior, as Anderson et al

Figure 2. Electrocardiograms recorded at rest (A) with QRS morphology of type A: atrioventricular conduction through the posterior atrioventricular node. Electrocardiograms recorded during morphological right atrial pacing at 400 ms (B): the same QRS morphology with type A. Electrocardiograms recorded at rest (C) with QRS morphology of type B: atrioventricular conduction through the anterior atrioventricular node. Electrocardiograms recorded during morphological right atrial pacing at 355 ms (D) with QRS morphology of type B. Electrocardiograms recorded during tachycardia (E) show the same QRS morphology with type B (cycle length, 254 ms).

Figure 3. Sites of successful slow pathway ablation in 8 patients with a single penetrating atrioventricular node (AVN). A. Left (45°) anterior oblique projection illustrating the 6 successful ablation sites in 6 patients with {S,L,L}. B. Right (30°) anterior oblique projection illustrating the 2 sites in the remaining 2 patients with {I,D,D}. AS indicates anteroseptal; MS, midseptal; and PS, posteroseptal.
and Dick et al12 have shown. Twin AVNs were inferred in one patient (11%) with {S,L,L} from the following electrophysiological characteristics: (1) 3 types of QRS morphologies appeared during both sinus rhythm and junctional rhythm, type A, type B, and fusion of both; (2) 2 anatomically separate His potentials were recorded; (3) there was a shift from type A to type B in QRS morphology when the refractory period of the primary conduction pathway in a single site was reached, and (4) during energy application, junctional rhythm was recorded both from 2 separate positions related to the 2 separate HBEs.3,20 Normal frontal axis in type A was consistent with conduction over the posterior A VN, and inferior axis in type B was consistent with conduction over the anterior A VN. The incidence of twin AVNs in CCTGA was 11%, similar to that reported previously.11

RFCA of AVNRT and the Ideal Site of SP Ablation in CCTGA

Recently, Eisenberger et al9 demonstrated that SP could be successful at the posteroseptal area in a patient with {S,L,L}. In the present study, AVNRT was successfully ablated with RFCA in all 9 of the patients with CCTGA. Among these 9 patients, 8 (88.9%) had SPs successfully ablated at the posterior midseptum, 3 at the posteroseptum (2 {S,L,L} at the right posteroseptum and 1 {S,L,L} at the left-sided posteroseptum) and 5 (3{S,L,L} and 2{I,D,D}) crossed over to the midseptum after failure of the posteroseptal SP ablation, and all had SP eliminated. SP potentials were recorded from ablation catheter before ablation in 2 cases. Junctional rhythm was observed during RFCA in all of the cases. No complication occurred in these 8 patients. These results indicated that, both in {S,L,L} and {I,D,D}, the SP input region can mainly be found in the posterior midseptum, pretty close to the region of SP in a normal heart. RFCA performed in these regions could usually ablate SPs successfully.

In patient No. 8 ({S,L,L}) with twin penetrating AVNs, RFCA of AVNRT in the conventional region of SP failed. The SP was finally successfully ablated at the confluence of the pulmonary and mitral annulus, slightly below the HBE-2. This finding was in accordance with that described by Tada et al3 in an {S,L,L} patient with twin AVNs. In the case that they studied, His potential was not recorded near the anterior AVN. However, we did find 2 types of nonpreexcitated QRS morphologies during junctional rhythm recorded from ablation near the posterior and the anterior A VNs, respectively. In addition, there existed fused QRS morphology during both sinus rhythm and junctional rhythm, the same with that described in our patient No. 8. On the basis of these findings, we hypothesize that, in {S,L,L} with twin penetrating AVNs, the input of SP is located near the anterior penetrating His bundle, and SP could be successfully ablated in this region.

Limitations

The present study had several limitations. First, all of data of patients with CCTGA were retrospectively analyzed but not on the base of any histological data. Second, AVNRT with CCTGA is a rare condition, and further study in large samples sizes is necessary.

Conclusions

This study demonstrated that, in {S,L,L} or {I,D,D}, RFCA of AVNRT is feasible. SP input region can mainly be found
in the posterior midseptum, especially in patients with single penetrating AVNs. SP could usually be successfully ablated in these regions. Occasionally in {S,L,L} with twin penetrating AVNs, SP could be successfully ablated near the anterior penetrating His bundle.

Disclosures
None.

References

CLINICAL PERSPECTIVE
Congenitally corrected transposition of the great arteries (CCTGA) is characterized by the combination of atrioventricular (AV) discordance and ventriculoarterial discordance. The associated anomalies, for example, ventricular septal defect and Ebstein anomaly of the tricuspid valve, commonly complicate the malformation. Further more, CCTGA may be associated with a variety of arrhythmias, including AV reentrant tachycardia, atrial tachyarrhythmias, and, rarely, AV nodal reentrant tachycardia (AVNRT) and ventricular tachycardia. In the present study, we investigate the feasibility of radiofrequency catheter ablation (RFCA) of AVNRT and ideal site for slow pathway (SP) ablation in CCTGA. We found that, in {S,L,L} or {1,D,D}, RFCA of AVNRT is feasible. SP input region can mainly be found in the posterior midseptum, especially in patients with single penetrating AVNs. SP could usually be successfully ablated in these regions. Occasionally in {S,L,L} with twin penetrating AVNs, SP could be successfully ablated near the anterior penetrating His bundle.
Atrioventricular Node Reentrant Tachycardia in Patients With Congenitally Corrected Transposition of the Great Arteries and Results of Radiofrequency Catheter Ablation
Zili Liao, Yu Chang, Jian Ma, Pihua Fang, Kuijun Zhang, Xiaqing Ren, Pingzhen Yang, Bo Yu, Jiqiang Hu, Qian Yang, Feifan Ouyang and Shu Zhang

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