Long-term outcome of twin atrioventricular node and supraventricular tachycardia in patients with right isomerism of the atrial appendage

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BACKGROUND Twin AV nodes and resulting supraventricular tachycardia (SVT) have been described in right atrial isomerism (RAI).

OBJECTIVE We sought to analyze the long-term outcome of patients with RAI with a focus on rhythm disturbances.

METHODS Retrospective study of 257 patients (152 male and 105 female, 1,171 patient-years) with RAI diagnosed between 1980 and 2005.

RESULTS SVT in 68 patients (26%) occurred at various ages from the prenatal period to 15 years and was only significantly associated with balanced ventricles ($P = .009$). Cardioversion was achieved in by verapamil in 6 of 6 cases (100%), adenosine in 18 of 21 cases (88%) and propranolol in 10 of 12 cases (83%). Electrocardiographic evidence of twin AV nodes, as shown by 2 discrete non–pre-excited QRS complexes, was found in 28 of 44 (64%) patients with more than 2 electrocardiograms, and was more frequent in those with balanced ventricles rather than a dominant ventricle and would increase risk of SVT. Recurrence of SVT was documented in 27 (40%) patients 1 day to 4.5 years after the first episode. However, the occurrence or recurrence of SVT was not associated with increased all-cause or surgical mortality or sudden death. Successful catheter ablation of ventriculoatrial pathways with junctional ectopic tachycardia at radiofrequency energy delivery was obtained in 5 of 6 patients.

CONCLUSION This study showed that twin AV nodes in RAI patients could be disclosed by serial electrocardiograms and that SVT, most likely a twin node tachycardia, was common and tended to recur but could be managed by ablation or medication.

KEYWORDS Right atrial isomerism; Heterotaxy syndrome; Tachycardia; Atrioventricular node

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Introduction

Abnormal cardiac conduction systems, including twin sinus nodes and twin AV nodes connected by a conducting sling, have been described in patients with right isomerism of the atrial appendage (RAI), in addition to an associated complex congenital heart disease.[1,2] Supraventricular tachycardia (SVT) may occur via a reentry between the twin AV nodes.[3–6] In our previous study, we proposed the term AV nodal–to–AV nodal tachycardia for such SVT. Although this type of SVT has been estimated to occur in about 25% of RAI patients, its impact on long-term outcome remains unclear.[3,4] The prognosis of RAI patients is poor owing to the presence of complex congenital heart disease, which typically involves a combination of total anomalous pulmonary venous return, common atrium, common AV valve, double-outlet from a dominant (usually right) ventricle, and pulmonary stenosis/atresia.[4,7–8] Furthermore, because of the absence of a spleen, such patients are at high risk of overwhelming sepsis.[9,10] However, recent advances in cardiac surgery and immunization against pneumococcus and hemophilus influenza, and palliative therapy for RAI by staged operation, may leave patients less vulnerable to infection, allowing them to survive into adulthood. AV nodal–to–AV nodal tachycardia may emerge or be modified as a result of such treatments. Given the dearth of relevant long-term data in previous studies, this study analyzed the clinical characteristics and long-term outcome of patients with RAI who presented with SVT.

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Methods

The data collection was in accordance with regulations, and this study was approved by the institutional review board of this institution.

Patient cohort

Patients with RAI were identified from the congenital heart disease patient database from 1980 to 2005. The medical records were reviewed and telephone interviews were con-

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Table 1 Intracardiac anomalies in the 257 patients with right atrial isomerism

	Patient number	%
Dextrocardia	69	27
Total anomalous pulmonary venous return to systemic veins	106	41
Infracardiac	25	
Supracardiac	64	
Mixed	17	
Pulmonary venous obstruction	94	37
Common atrium	241	94
Common AV valve	237	92
Ventricles		
Dominant 1 ventricle	213	83
Right ventricle	167	
Left ventricle	8	
Indeterminate ventricle	38	
Balanced 2 ventricles	44	17
Ventriculoarterial connection		
DO(R)V	227	88
TGA	23	9
Concordant	7	3
Pulmonary valve		
Pulmonary stenosis	152	59
Pulmonary atresia	81	32
Coarctation/interruption of aortic arch	7	3
Major aortopulmonary collateral arteries	12	5

DO(R)V = double outlet (right) ventricle; TGA = transposition of great arteries.

ducted with those who were lost to follow-up. All electrocardiograms were reviewed to define the antegrade AV conduction pattern. Antegrade AV conduction through an AV node was defined when a non–pre-excited QRS complex followed a P wave. The diagnosis for RAI was based on a combination of imaging information including computed tomography, echocardiography, and angiography demonstrating the presence of an ipsilateral descending aorta and a suprarenal segment of inferior vena cava (at echocardiography) and the presence of bilateral right atrial appendage (at computed tomography).[11,12] The anatomy was further confirmed at surgery or autopsy if feasible. The diagnostic criteria for SVT have been described previously.[4]

Statistics

Data are expressed as mean ± standard deviation (medians with ranges). Kaplan-Meier estimates were used to draw the event-free curves, and log-rank and Wilcoxon tests were used to examine statistical significance.[13] Cox proportional-hazard regression modeling was used to identify independent factors associated with time-related events.[14] Variables related to time-related events with $P < .1$ in the univariate analysis were entered into the multivariate model. Statistical significance was set at a value of $P < .05$.

Results

A total of 257 patients (152 male and 105 female) with RAI were identified and enrolled. The total follow-up duration of these patients from birth was 1,171 patient-years. There were 54 patients lost to follow-up at the time of study, with the age at last follow-up ranging from 10 days to 235 months (median 14 months). For the 203 patients with complete follow-up, the age at last follow-up ranged from 1 day to 351 months (median 26 months). The associated cardiac anomalies are summarized in Table 1. Survival was 66%, 48%, 42%, and 30% at the ages of 1, 5, 10, and 15 years, respectively.

Twin AV nodes and supraventricular tachycardia

SVT occurred in 68 (26%) of the 257 patients. The onset age ranged from fetal period (2 patients) to 15 years (median 22 months). Twenty-seven (40%) patients had their first SVT attack either in the prenatal period or in infancy. Freedom of SVT was documented in 86%, 68%, 55%, and 39% of patients at the ages of 1, 5, 10, and 15 years, respectively (Figure 1). By Cox regression, the occurrence of SVT was not a significant predictor for survival ($P = .53$). Freedom of SVT decreased with age but remained stationary after 15 years of age. Only the

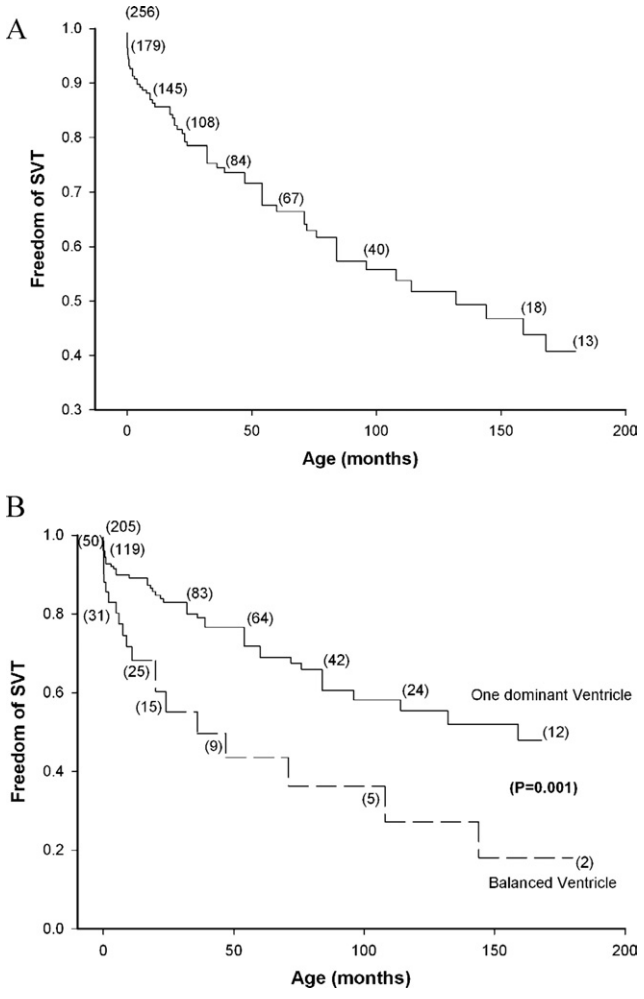

Figure 1 **A:** The freedom of supraventricular tachycardia drawn by Kaplan-Meier analysis in the 257 patients with right isomerism of the atrial appendage (RAI). **B:** The freedom of supraventricular tachycardia in the 205 RAI patients with 1 dominant ventricle (solid line) and 50 RAI patients with balanced ventricles (broken line). In 2 patients, the ventricular morphology could not be characterized as dominant 1 or balanced 2 ventricles.