Original Article

Atrioventricular Nodal Reentrant Tachycardia in Patients With Congenital Heart Disease

Outcome After Catheter Ablation

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Background—The relationship of atrioventricular nodal reentrant tachycardia to congenital heart disease (CHD) and the outcome of catheter ablation in this population have not been studied adequately.

Methods and Results—A multicenter retrospective study was performed on patients with CHD who had atrioventricular nodal reentrant tachycardia and were treated with catheter ablation. There were 109 patients (61 women), aged 22.1±13.4 years. The majority, 86 of 109 (79%), had CHD resulting in right heart pressure or volume overload. Patients were divided into 2 groups: group A (n=51) with complex CHD and group B (n=58) with simple CHD. There were no significant differences between groups in patients' growth parameters, use of 3-dimensional imaging, and type of ablation (radiofrequency versus cryoablation). Procedure times (251±117 versus 174±94 minutes; P=0.0006) and fluoroscopy times (median 20.8 versus 16.6 minutes; P=0.037) were longer in group A versus group B. There were significant differences between groups in the acute success of ablation (82% versus 97%; P=0.04), risk of atrioventricular block (14 versus 0%; P=0.004), and need for chronic pacing (10% versus 0%; P=0.008). There was no permanent atrioventricular block in patients who underwent cryoablation. After 3.2±2.7 years of follow-up, long-term success was 86% in group A and 100% in group B (P=0.004).

Conclusions—Atrioventricular nodal reentrant tachycardia can complicate the course of patients with CHD. This study demonstrates that the outcome of catheter ablation is favorable in patients with simple CHD. Patients with complex CHD have increased risk of procedural failure and atrioventricular block. **(*Circ Arrhythm Electrophysiol*. 2017;10:e004869. DOI: 10.1161/CIRCEP.116.004869.)**

Key Words: atrioventricular block ■ catheter ablation ■ cryoablation ■ tachycardia, atrioventricular nodal reentry ■ tachycardia, supraventricular

Congenital heart disease (CHD) is frequently complicated by supraventricular tachycardia, which can occur related to either congenital or acquired substrates. Although arrhythmias related to accessory pathways are well-known to coexist with certain types of CHD,[1,2] the relationship between atrioventricular nodal reentrant tachycardia (AVNRT) and CHD has been less well studied. Understanding this relationship is especially important because the treatment of AVNRT may be problematic in patients with CHD, either because of the congenitally distorted anatomy of the atrioventricular junction or because of post-operative difficulties with access to the atrioventricular nodal area. The former occurs commonly in patients with atrioventricular canal defects,[3] congenitally corrected transposition of the great arteries,[4] or other complex heart defects, such as tricuspid atresia[5] and double inlet ventricles.[6,7] The latter can occur because of intra-atrial baffles (as in transposition patients with intra-atrial surgery)[8] or various types of Fontan procedures.[9] Even simple congenital heart defects requiring placement of patches close to the conduction system might present challenges to safe catheter ablation.

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Recurrent tachycardia may compromise an already tenuous hemodynamic status and lead to significant morbidity. In addition, treatment with antiarrhythmic medications may have limited efficacy and higher incidence of side effects than in patients with structurally normal hearts.[10,11] Therefore,

Received November 22, 2016; accepted June 8, 2017.

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WHAT IS KNOWN

- Although the relationship between congenital heart disease and accessory pathways is well-known, there have been few reports on a relationship between congenital heart disease and atrioventricular nodal reentrant tachycardia (AVNRT), and especially the outcome of catheter ablation in patients with this combination of problems.
- The description of factors affecting outcome of catheter ablation of AVNRT in various types of congenital heart disease would be beneficial in patient selection and use of the appropriate mode of mapping and ablation

WHAT THE STUDY ADDS

- AVNRT can complicate the course of patients with congenital heart disease, especially in patients with right heart pressure or volume overload.
- The outcome of catheter ablation is favorable in patients with simple congenital heart disease, but patients with complex congenital heart disease have increased risk of procedural failure and atrioventricular block.
- Advanced methods of mapping and cryoablation should be considered when the exact anatomy of the atrioventricular node cannot be precisely defined.

it is important to recognize the patterns of relationship of AVNRT with CHD and any difficulties in its transcatheter management and outcome. There have been previous case reports or case series on patients with specific forms of CHD, and, as well, a recent single-center retrospective study on this subject has been published.[12] We sought to extend this experience by collecting data on patients with diverse forms of CHD from multiple centers.

Methods

Data Collection

A multicenter retrospective study was performed among members of the Pediatric and Congenital Electrophysiology Society. Centers were contacted through the mailing list of Pediatric and Congenital Electrophysiology Society and were asked to provide deidentified data through REDCap electronic data capture tools hosted at Children's Mercy Hospital in Kansas City. Data were collected from 14 centers on patients with CHD who had an invasive electrophysiological study for diagnosis and management of supraventricular tachycardia. Institutional review board approval was obtained from each individual center according to local policies, and collaborating centers signed a data use agreement with the coordinating center.

The following data were collected: patient demographics and growth parameters; clinical, echocardiographic, and ECG data; history of surgical or catheter interventions directed to the management of the congenital heart defects; onset of AVNRT in relationship to any interventional or surgical procedure; use of antiarrhythmic medications; time of invasive electrophysiological study; type and cycle length of AVNRT during electrophysiological study; additional arrhythmias; type(s) of imaging modalities used for catheter navigation; type of catheter ablation (radiofrequency versus cryoablation); approach to the ablation site; acute result; procedure and fluoroscopy duration and complications; any recurrences during follow-up and any data on repeat procedures and final outcome. Follow-up data were obtained from hospital records.

Exclusion Criteria

We excluded patients with minimal CHD that was unlikely to affect the location and function of the atrioventricular node. Patients with the following diagnoses were excluded: isolated bicuspid aortic valve, coarctation of the aorta, patent ductus arteriosus, isolated aortopulmonary collateral arteries, coronary artery anomalies, such as origin from the wrong sinus, and small atrial or ventricular septal defects remote from the conduction system. Patients with a primary cardiomyopathy (eg, dilated, hypertrophic) were also excluded.

Electrophysiological Study, Ablation Methods, and Procedural End Points

Standard criteria for the diagnosis of AVNRT were used. These included (1) narrow complex tachycardia; QRS morphology during tachycardia similar to sinus rhythm; or, in cases of wide-complex tachycardia, proof that QRS change during tachycardia was because of rate-related aberrancy; (2) concentric retrograde conduction; (3) for typical AVNRT, a shortest ventriculo-atrial interval of <70 ms[9]; and (4) for atypical AVNRT, a shortest ventriculo-atrial interval of >70 ms[9] and exclusion of other operative mechanisms, such as ectopic atrial tachycardia or accessory pathway-mediated tachycardia by use of pacing maneuvers (overdrive ventricular pacing, placement of His-refractory premature ventricular stimuli during tachycardia).

The method of electrophysiological study and number, size, and type of catheters used were individualized according to institutional practice and patient characteristics.

The type of energy used for ablation also varied according to institutional preference, and in procedures performed in more recent years, cryoablation was increasingly used.

Most of the radiofrequency catheters that were used were 4-mm tip (n=57) while 14 were 8-mm tip catheters and 4 were irrigated tip catheters. The power that was used varied from 20 to 75 watts. Powers >50 watts were used 12× with 8-mm tip catheters. In 8 cases, the type of catheter was unknown. The temperatures achieved were 44 to 70 (57±7°C). When junctional rhythm was noticed, a full 60-second lesion was usually delivered.

Of the cryoablation catheters, most (28) were 6 mm, 4 were 4 mm, and 1 was 8-mm tip. When a site was considered a good target, 4-minute duration lesions were delivered, and the usual minimum temperature achieved was −70°C to −80°C.

The use of 3-dimensional (3D) mapping systems for navigation and electroanatomic mapping increased during the study period, and in the most recent years, various forms of these systems were used by most institutions. Forty-nine procedures were performed with fluoroscopic guidance only and 60 were performed with a 3D mapping system, 8 of which with a completely nonfluoroscopic approach.

The slow pathway was targeted using standard anatomic criteria in patients with usual 2-ventricle anatomy. In patients with distorted anatomy of the atrioventricular junction, the approach was individualized. In general, the most posterior sites with reference to the coronary sinus ostium were tried first, with an attempt to position the catheter in a location with a small atrial and a larger ventricular signal. In cases of atrioventricular canal defects, the catheter was positioned inferior to the level of the coronary sinus ostium because in these cases, the atrioventricular node is posteriorly displaced. When the intracardiac anatomy was distorted by a conduit or baffle, access to the atrioventricular nodal area was achieved with various approaches, including transbaffle or retrograde through the aorta.

Attempts were made to eliminate slow pathway conduction, but if this could not be achieved, noninducibility of tachycardia was accepted as an end point.

Recurrence of tachycardia was defined as the presence of electrocardiographically documented episodes of the same tachycardia as the one previously ablated or inducibility of the same tachycardia during a repeat electrophysiological study. Final success was defined as successful atrioventricular nodal modification with no inducible tachycardia after repeated procedures. Long-term success was defined as absence of symptoms of tachycardia at least 1 year after the last ablation procedure. Also noted was the presence of acute and long-term complications, such as atrioventricular block with need for pacemaker implantation.

Classification of Patients With CHD

Patients were divided according to complexity of their CHD in 2 groups as depicted in Table 1.

Statistical Analysis

Descriptive statistics, such as means, medians, SDs, first and third quartiles (Q1–Q3), and proportions, were used to summarize the data. Group comparisons of categorical variables were made using χ^2 or Fisher exact tests. Continuous variables were compared using *t* tests or Wilcoxon rank-sum tests. $P<0.05$ was considered statistically significant. All analyses were generated using SAS, version 9.4 (SAS Institute Inc., Cary, NC).

Results

General Characteristics

There were 109 patients included in the study. The clinical features according to the type of CHD (complex versus simple) are depicted in Table 2. There were no significant differences between groups in the number of patients, sex, age at procedure, weight, height, and body surface area. Significantly, more patients in the group with complex CHD presented in the post-operative period. This is most likely because of the fact that the age at first surgery was significantly younger in this group. More patients in group B were treated with antiarrhythmic medications.

Table 1. Distribution of Patients in the 2 Groups

Group A: Complex CHD	N	Group B: Simple CHD	N
Tetralogy of Fallot	10	ASD 2	28
D-TGA after Mustard or Senning	9	VSD	8
Single ventricle physiology	6	Aortic valve disease	7
Ebstein anomaly	6	Pulmonary valve stenosis	5
Double outlet right ventricle	5	Mitral valve disease	3
Partial atrioventricular canal	4	ASD sinus venosus	2
CC-TGA	4	Partial anomalous pulmonary venous return	2
Common atrioventricular canal	3	Aortic aneurysm (Marfan syndrome)	1
Pulmonary atresia-intact ventricular septum	2	Subaortic stenosis VSD	1
Total anomalous pulmonary venous return	1	Supravalvar aortic stenosis (Williams syndrome)	1
Shone complex	1		
Total	51		58

ASD 2 indicates atrial septal defect secundum type 2; CC-TGA, congenitally corrected transposition of the great arteries; CHD, congenital heart disease; D-TGA, D-transposition of the great arteries; and VSD, ventricular septal defect.

Hemodynamic Characteristics

Eighty-six of the patients (79%) had a congenital heart defect that resulted in pressure and volume overload of the right heart. The remaining 21% of the patients had aortic or mitral valvular lesions or left ventricular outflow obstruction.

Electrophysiological Findings and Procedural Data

The electrophysiological findings and procedural data are depicted in Table 3. There were more patients with atypical AVNRT in group A than in group B. The procedure and fluoroscopy times were also significantly longer in group A.

There was no significant difference in the average tachycardia cycle length, type of anesthesia used, incidence of use of a 3D mapping system, and ablation energy. There were similar numbers of patients with additional arrhythmia substrates that were ablated during the same session. In group A, there were 4 patients with accessory pathways, 1 with ectopic atrial tachycardia and 1 with intra-atrial reentry tachycardia. In group B, there were 2 patients with accessory pathways and 2 with typical cavotricuspid isthmus-dependent atrial flutter. There was also an equal distribution of hemodynamic interventions that were performed during the same procedure in the 2 groups. In group A, there was a baffle leak closure and a collateral vessel embolization, whereas, in group B, there were 5 percutaneous atrial septal defect occlusions.

Junctional rhythm was noticed during radiofrequency ablation in 68 patients, and it was absent in 6, and in 1 case, radiofrequency was delivered during tachycardia and resulted in abrupt termination of tachycardia.

In 63 patients, there was no residual slow pathway conduction. Some evidence of residual dual atrioventricular nodal physiology (atrio-His jump or sustained slow pathway conduction during decremental pacing) was noticed in 40 patients. In 6 patients, this information was not available. There were 6 recurrences in the patients without residual slow pathway conduction (10%) and 12 recurrences in those with residual slow pathway conduction (30%; $P=0.006$).

Procedural Outcome of AVNRT Ablation in Patients With Complex Versus Simple CHD

The outcome data are depicted in Table 4. There was a significantly lower acute success rate (82% versus 97%; $P=0.04$) and a lower long-term success rate (86% versus 100%; $P=0.004$) in group A versus group B. Although group A had a higher recurrence rate, this difference did not reach statistical significance. Procedure-related atrioventricular block and need for chronic pacing occurred only in group A.

Outcome of AVNRT Ablation After Radiofrequency Versus Cryoablation

The acute success rate was similar with radiofrequency and cryoablation (83% each) in patients from group A ($P=1.0$). In general, centers that performed a significant number of ablations used both modalities, except for 1 center that used only radiofrequency. There was also no difference between the 2

Table 2. Clinical Characteristics

	Group A: Complex CHD	Group B: Simple CHD	*P* Value
No. of patients	51	58	
Female sex	26	35	0.32
Age at presentation, y	1–45 (median: 14, Q1–Q3: 9–22)	3–62 (median: 14, Q1–Q3: 11–29)	0.34
Age at first ablation, y	3–61 (median: 18 Q1–Q3: 13–28)	4–65 (median: 19, Q1–Q3: 13–31)	0.74
Height, cm	95–190 (161±21)	60–193 (160±21)	0.83
Weight, kg	14–107 (60±23)	17–114 (60±20)	0.86
BSA, m^2	0.61–2.34 (1.61±0.39)	0.73–2.30 (1.63±0.32)	0.70
Age at first surgery, y	0–14 (median: 1, Q1–Q3: 0–2.5)	0–67 (median: 10, Q1–Q3: 3–20)	0.00001
Onset of AVNRT pre-operatively	17 (33%)	31 (52%)	0.035
Number treated with medications	60	41	0.05

AVNRT indicates atrioventricular nodal reentrant tachycardia.

ablation methods in patients from group B (radiofrequency: 95%; cryoablation: 100%; *P*=1.0). The recurrence rate was also similar within groups between the 2 modalities. In group A, there was 20% recurrence with radiofrequency versus 29% with cryoablation (*P*=0.5), and in group B, the recurrence was 10% with radiofrequency and 6% with cryoablation (*P*=1.0).

Patients With Failed Ablation

The characteristics of patients with failed ablation are depicted in more detail in Table 5. The ablation was considered an acute failure in 11 patients because of persistence of inducible AVNRT. There were significantly more patients with atypical AVNRT among patients with failed versus successful ablation (*P*=0.028), with 5 of the 11 failed ablations associated with atypical AVNRT. Four of them were of the slow-slow type and 1 was associated with the fast-slow type. The ablation method did not specifically predict procedural failure (radiofrequency in 9 and cryoablation in 2).

Five of the patients with failed ablation were patients with single ventricle physiology and complex anatomy, 3 after a modified Fontan operation and 2 before Fontan completion. Notably, 2 of them were patients with tricuspid atresia. In 1 of them, the ablation attempts were performed through a patent foramen ovale on the left atrial posteroseptal area and in the other, the ablation attempts were performed in the right posteroseptal area. Of the patients with 2 functional ventricles and failed ablation, 2 had congenitally corrected transposition of the great arteries (CC-TGA) and 2 had D-transposition of the great arteries (D-TGA) after an atrial switch procedure. Of the patients with CC-TGA, 1 was of the situs solitus, L-loop ventricles, L-transposed great arteries ({S,L,L}) and one of the situs inversus, D-loop ventricles, D-transposed great arteries ({I,D,D}) type. Both had ablation attempted on the systemic venous side, the first on the right and the second on the left side of the atrial septum. One of the D-TGA patients underwent ablation from the systemic venous side and one from the pulmonary venous side through a transbaffle approach. Although it was initially thought that only a modification of the circuit was achieved, with persistence of the slow pathway, both of the D-TGA patients were free of long-term recurrences during at least 3 years of follow-up. The final 2 patients belonged to the simple CHD group and both had a small muscular ventricular septal defect and other left-sided

Table 3. Electrophysiological Findings and Procedural Data

Variable	Group A	Group B	*P* Value
Typical AVNRT	37 (73%)	51 (90%)	0.02
Atypical AVNRT	14 (28%)	7 (11%)	
Both	1	0	
AVNRT cycle length, ms	Median: 336 Q1–Q3: 300–390	Median: 340 Q1–Q3: 320–390	0.36
Ablation energy	Radiofrequency: 34, cryoablation: 15, both: 2	Radiofrequency: 41, cryoablation: 17, both: 0	0.43
Additional arrhythmia ablated	9 (16%)	5 (9%)	0.23
Additional intervention performed	4 (8%)	5 (9%)	1
3-Dimensional mapping system	30 (52%)	30 (59%)	0.46
Procedure time, min	251±117, 54–506 min	174±94, 40–415 min	0.006
Fluoro time, min	Median: 20.8 (Q1–Q3: 25)	Median: 16.6 (Q1–Q3: 17.7)	0.037

AVNRT indicates atrioventricular nodal reentrant tachycardia.

Table 4. Outcome Data

	Group A	Group B	*P* Value
Acute success	82%	97%	0.04
AVNRT documented and reablated in repeat procedure	9 (18%)	6 (10%)	0.27
Final success	86%	100%	0.004
Atrioventricular block of any degree	14%	0%	0.004
Need for chronic pacing	10%	0%	0.008

AVNRT indicates atrioventricular nodal reentrant tachycardia.

valvular lesions. In these patients, the ablation attempts were made at the typical location on the right side of the septum.

Atrioventricular Block as a Complication of AVNRT Ablation in Patients With Complex Versus Simple CHD

Table 6 depicts the clinical, anatomic, and procedural data on patients who developed chronic atrioventricular block. All 5 patients belonged to the complex CHD category, and radiofrequency energy was the ablation energy that was used in all. There was no permanent atrioventricular block in patients who underwent cryoablation. The weight of the patients was similar to the average weight of the study group, except for 1 patient who was smaller. The types of AVNRT were similar to the rest of the complex group. The 3D mapping was used in a similar number of patients compared with the rest of the group.

Outcomes of Nonconventional AVNRT Ablation Approaches in Patients With Complex CHD

We specifically examined the outcome of patients who underwent ablation via a nonconventional approach (ie, not through a direct femoral venous access to the right atrium; Table 7). There were several nonconventional approaches to ablation, including through patent foramen ovale, trans-septal, transbaffle, transhepatic, and retrograde (transaortic). The 1 patient with totally anomalous pulmonary venous connection who had a trans-septal approach had acute success without complications and without recurrences, as did the 5 patients with transposition of the great arteries after an intra-atrial baffle who underwent a retrograde approach. On the contrary, the 5 patients with complex CHD after various types of Fontan operation who underwent ablation through an unconventional approach had an increased incidence of complications (1 atrioventricular block and 1 ventricular fibrillation) and unfavorable long-term outcome (2 failures, 2 recurrences with only 1 final success and 1 patient requiring pacemaker implantation).

Follow-Up

The average follow-up period after the last ablation procedure was 3.2±2.8 years. Six patients had no documented follow-up.

Table 5. Characteristics of Patients With Failed Ablation

Patient	Diagnosis	Previous Surgery	Type AVNRT	Ablation Method	Approach/Location	Comments
1	CC-TGA {I,D,D}	VSD closure	Slow-slow	Both radiofrequency and cryoablation	Transvenous, RA (L-sided)/PS, MS	Recurrent SVT
2	DORV	LT Fontan	Slow-fast	Radiofrequency	Retrograde transaortic/ not specified	Recurrent SVT, transplant
3	DORV	LT Fontan	Slow-fast	Cryoablation	Transbaffle approach/ PS, MS	Recurrent SVT, repeat procedure X2, recurrence
4	Tricuspid atresia	AP connection	Slow-slow	Radiofrequency	Trans-PFO to LA	Failed second attempt
5	D-TGA	Mustard	Slow-fast	Cryoablation	Transbaffle approach/AS	No SVT, S-ICD for VF
6	D-TGA	Mustard	Slow-fast	Radiofrequency	Transvenous antegrade/ PS	No recurrent SVT
7	TA	Pre-Fontan	Slow-slow	Radiofrequency	Transvenous, R-side/PS	Cryoablation during Fontan, no SVT
8	CC-TGA {S,L,L}	None	Slow-fast	Radiofrequency	Transvenous R-side/not specified	Failed second attempt
9	Single ventricle {I,L,D}	After Glenn, pre-Fontan	Slow-fast, Slow-slow	Radiofrequency	Transvenous/CS ostium	Second attempt. Intermittent atrioventricular block
10	Dysplastic mitral valve, small VSD	Surgical mitral valvuloplasty	Fast-slow	Radiofrequency	Transvenous, R-side/not specified	Successful second attempt
11	Sub aortic stenosis, small VSD	Subaortic myectomy	Slow-fast	Radiofrequency	Transvenous, R-side/ midseptal	Successful second attempt

AP indicates atriopulmonary; AS, anteroseptal; AVNRT, atrioventricular nodal reentrant tachycardia; CC-TGA, congenitally corrected transposition of the great arteries; CS, coronary sinus; D-TGA, D-transposition of the great arteries; DORV, double outlet right ventricle; {I,D,D}, situs inversus, D-loop ventricles, D-transposed great arteries; LA, left atrium; LT, lateral tunnel; MS, midseptal; PFO, patent foramen ovale; PS, posteroseptal; RA, right atrium; S-ICD, subcutaneous implantable cardioverter defibrillator; {S,L,L}, situs solitus, L-loop ventricles, L-transposed great arteries; SVT, supraventricular tachycardia; VF, ventricular fibrillation; and VSD, ventricular septal defect.

Table 6. Characteristics of Patients With Permanent AV Block

Case	Weight, kg	AVNRT Type	CHD Type	Ablation Method	3-Dimensional System
1	59	Fast-slow	CC-TGA	Radiofrequency	No
2	44	Slow-fast	DIRV-TGA	Radiofrequency	Yes
3	54	Slow-fast	TGA s/p Mustard	Radiofrequency	Yes
4	63	Slow-fast	Tetralogy of Fallot	Radiofrequency	No
5	30	Slow-slow	Single ventricle s/p Fontan	Radiofrequency	Yes

AVNRT indicates atrioventricular nodal reentrant tachycardia; CC-TGA, congenitally corrected transposition of the great arteries; CHD, congenital heart disease; DIRV, double inlet right ventricle; and TGA, transposition of the great arteries.

Discussion

This is the first multicenter study published to date on AVNRT in patients with CHD and the only multicenter study on this subject. The major findings of the study are the following:

(1) AVNRT may complicate the course of patients with CHD either before or after surgical or interventional procedure related to the CHD. The majority of the patients in our series had right-sided pressure or volume overload lesions; (2) catheter ablation of AVNRT is feasible in patients with CHD although more challenging in patients with complex CHD, with lower success rates and significant risk of complications and especially atrioventricular block in this group; (3) atypical AVNRT is more commonly present in patients with complex CHD and seems to be a predictor of procedural failure; and (4) cryoablation is equally effective to radiofrequency ablation in patients with similar level of CHD complexity and is not related to a risk of atrioventricular block.

Is There a Relationship Between AVNRT and CHD?

In our study group, there was a predominance of right-sided pressure or volume overload lesions. We do not know whether a pathogenic relationship exists between AVNRT and pressure or volume overload of the right heart because examining this relationship would require a study of a large population of patients with CHD. However, it is plausible that hemodynamic effects may lead to cellular electrophysiological alterations and intercellular fibrosis that subsequently promote nonuniform anisotropy, leading to a critical delay in slow pathway conduction that may allow initiation and maintenance of re-entry. The relationship of AVNRT with CC-TGA has been reported several times in the literature with one series of several patients describing AVNRT in CC-TGA.[4] It may be theorized that the distorted anatomy of the conduction system in this disease, with frequently reported 2 atrioventricular nodes (of which typically only the anterior one has a connection to the bundle of His), creates a substrate for a longer slow pathway with more nonuniform conduction, increasing the possibility of reentry. The effect of surgical suture lines may also need to be considered, as is evident in patients with transposition of the great arteries after Mustard or Senning operation, who may develop AVNRT many years postoperatively.[8]

The coexistence of other types of CHD and AVNRT is rare and does not allow for any particular anatomic-functional correlation.

Challenges of AVNRT Ablation in Patients With CHD

The significantly lower success rate in patients with complex CHD in our study should not come as a surprise given the fact that the anatomy of the atrioventricular node may differ significantly from normal in these patients. A review of our patient population shows that of the 11 failed ablations, 9 belonged to the complex CHD group. However, even in the group of complex CHD, the majority of the patients had a successful outcome of catheter ablation.

AVNRT in CC-TGA

Based on histopathologic studies, it has been concluded that in CC-TGA, there is usually an anteriorly placed atrioventricular node that makes contact with the penetrating His bundle that courses around the right side of the pulmonary valve, subsequently bifurcating to the inverted bundle branches. A posteriorly placed hypoplastic atrioventricular node is usually present, which in most cases does not conduct to the ventricles.[13] In CC-TGA associated with situs inversus (also frequently coded as TGA {I,D,D}), intraoperative studies showed that the conduction system is normally located

Table 7. Patients With Nonconventional Approach to Ablation

Patient	Diagnosis	Approach	Energy	Complications	Long-Term Outcome
1	TAPVR	Trans-septal	Cryoablation	None	Success
2–6	TGA s/p Senning or Mustard	Retrograde	Radiofrequency	None	Success
7	Tricuspid atresia s/p APF	Trans-PFO to LA	Radiofrequency	None	Failure
8	DIRV, HLHS s/p ECF	Transhepatic	Cryoablation	None	Recurrence, final success
9	DIRV, TGA s/p ECF	Retrograde	Radiofrequency	Atrioventricular block	Success, PPM
10	DORV, SV, s/p TCPC	Retrograde	Radiofrequency	VF	Failure, finally transplant
11	DORV, SV, TCPC	Transbaffle	Cryoablation	None	Recurrence X3

APF indicates atriopulmonary Fontan; DIRV, double inlet right ventricle; DORV, double outlet right ventricle; ECF, extracardiac Fontan; HLHS, hypoplastic left heart; LA, left atrium; PFO, patent foramen ovale; PPM, permanent pacemaker; SV, supraventricular; TAPVR, totally anomalous pulmonary venous connection; TCPC, total cavopulmonary connection; TGA, transposition of the great arteries; and VF, ventricular fibrillation.

albeit left sided.[14] In the study of Liao et al,[4] the only series of patients with CC-TGA who underwent ablation of AVNRT, the site of successful ablation both in {S,L,L} and {I,D,D} configurations was located in the posterior midseptum, close to the region of slow pathway in a normal heart. This study did not prove or disprove the presence of a posterior atrioventricular node. It suggests, however, that the slow pathway is located normally at the posterior or midseptal area and that one does not need to ablate anterosuperiorly, close to the conducting atrioventricular node, which increases the risk of atrioventricular block. These findings are also in agreement with the series of Upadhyay et al,[12] in which successful ablation could be achieved in the posterior third of the triangle of Koch in either the right side in {S,L,L} or in the left side in {I,D,D} configuration. However, Tada et al[15] have reported a patient with CC-TGA {S, L, L} and AVNRT with triple atrioventricular nodal pathways in whom the successful ablation site was at the anteroseptal area. In our series, out of 4 patients with CC-TGA with attempted ablation, the procedure failed in 1 patient with {S, L, L} and 1 patient with {I, D, D} configuration, and first-degree atrioventricular block occurred in 1 of the 2 successful ablations. In both of these patients, ablation was performed in the systemic venous atrium (right and left sided, respectively), and applications were made in the posterior and midseptal area. It is possible that operators were more conservative because of the perceived higher risk of atrioventricular block in these patients.

AVNRT in Atrioventricular Canal Defects

Ablation of AVNRT in patients with complete or partial atrioventricular canal defects is also challenging because of the posterior displacement of the compact atrioventricular node in front of the coronary sinus ostium. In our series, successful ablation was achieved in all 7 patients with complete or partial atrioventricular canal defects with either cryo or radiofrequency ablation. Conduction abnormalities developed in 2 of the 4 patients in whom radiofrequency ablation was used, 1 with transient complete atrioventricular block and 1 with first-degree atrioventricular block. Although none of these patients required chronic pacing, these findings attest to the notion that radiofrequency ablation has increased risk of atrioventricular block in these patients and cryoablation should be the method of choice.[3] An inverted relationship of the slow and fast pathways has also been reported in patients with this anatomy.[16] A left-sided ablation of the posterior extension of the atrioventricular node may be another approach in these patients.[17]

AVNRT in D-TGA

Patients with D-TGA who have undergone an atrial switch operation (Mustard or Senning) represent another challenging group. The difficulties in this group arise mostly because of the presence of the intra-atrial baffle that prevents direct access to the triangle of Koch from a femoral venous approach. Because the location of the triangle of Koch (and the slow pathway) is most commonly on the pulmonary venous side of the baffle, either a retrograde[8] or a transbaffle[12,18] approach is usually necessary. Because of the retrospective multicenter nature of our study, a uniform approach was not used. Of the 9 such patients in our study, ablation was successful in all patients in whom the retrograde approach was used (all done with radiofrequency ablation). In the 4 patients in whom an antegrade approach to the venous side was used, there were 2 failures (1 with cryoablation and radiofrequency each) and 1 patient developed complete heart block. The findings from our study, as well as those from the literature, strongly support the need for access to the pulmonary venous side, either through a transbaffle or a retrograde approach, to successfully and safely ablate AVNRT in this setting.

AVNRT in Patients With Single Ventricle Physiology

By far, the most difficult group of patients with CHD and AVNRT are those with single ventricle physiology, either before or after a modified Fontan operation. In our study, 5 of 8 patients who had undergone a type of Fontan operation had a failed ablation attempt, the highest failure rate than any other group. The variable anatomy of the atrioventricular node and the frequent absence of the usual landmarks of the triangle of Koch, such as a catheter in the coronary sinus, does not allow any specific conclusions. In the study by Upadhyay et al,[12] of 16 patients with single ventricle physiology, ablation was attempted in only 10 because of uncertainty of the anatomy of the atrioventricular node, and success was achieved in 8. The approach was transbaffle in 4 patients, retrograde in 2, and antegrade transvenous in only 4. Although a remarkable success rate of 80% was achieved, this may not be the case in all centers, which could influence whether or not catheter ablation is deferred. The decision to defer may be disappointing for the patient and the interventionist, but it may be the wisest approach in cases where AVNRT is either infrequent or hemodynamically tolerated and easily treated with medications. It may save the patient from a prolonged unsuccessful procedure with exposure to radiation and to the risk of complications, especially atrioventricular block. In patients with tricuspid atresia, successful slow pathway ablation may be more often achieved from the left side of the atrial septum through a trans-septal/transpatent foramen ovale approach [5,19] or theoretically through a retrograde approach.

Atypical AVNRT: A Risk Factor for Failure?

There was a higher incidence of atypical AVNRT in the complex CHD group and an even higher incidence among the patients with procedural failure. The presence of slow-slow AVNRT was especially high in this group. The 2 CHD diagnoses associated with slow-slow AVNRT were CC-TGA and single ventricle physiology. Triple atrioventricular nodal physiology has been reported previously in a patient with CC-TGA.[15] The complexity of the anatomy probably predisposes to the development of multiple slow pathways. The presence of atypical AVNRT has been reported to be a predictor of procedural failure in a large study of AVNRT ablation in an adult population with normal anatomy (hazard ratio, 3.1).[20] It is reasonable to conclude that the combination of complex CHD with atypical AVNRT, especially with multiple slow pathways, increases the risk of procedural failure.

Potential Ways to Mitigate the Risk of Atrioventricular Block

Chronic atrioventricular block is a serious complication in patients with complex CHD. Permanent pacing is indicated in all of these patients, with all the attendant problems, including the risk of lead malfunction, infection, potential need for repeated open chest procedures, and dyssynchrony-induced cardiomyopathy.[21] It is, therefore, of the utmost importance to avoid damage to the atrioventricular node. In patients with simple CHD, the location of the atrioventricular node is usually not different from the normal hearts. However, in patients with complex CHD, the anatomy of the atrioventricular node is variable and the usual landmarks of the triangle of Koch are often missing. The above may explain the increased risk of damage to the compact atrioventricular node during attempts at catheter ablation of AVNRT. It may be wise in many of these patients to exhaust pharmacological therapy or to treat isolated episodes with vagal maneuvers, a pill in the pocket approach, or intravenous adenosine. In patients who have frequent, difficult to control, or poorly tolerated episodes, catheter ablation can be attempted. It is probably safer in these patients to use cryoablation unless the anatomy of the atrioventricular conduction system is clearly defined. In our series, all patients who developed atrioventricular block had undergone radiofrequency ablation. In addition, the outcome of cryoablation was not inferior to that of radiofrequency ablation among patients with the same degree of CHD complexity. There are, however, disadvantages of cryoablation that preclude its more generalized use, including the stiffness of the catheters, the limited curves available, the need for more lesions of longer duration, and the higher incidence of recurrence, as documented in the literature on noncongenital patients with AVNRT.[22]

Advanced Mapping and Navigation Systems

The use of 3D mapping techniques may also allow safer ablation of AVNRT in complex CHD. The construction of 3D models with MRI or computed tomography and the merging of electric information with the 3D anatomy may be helpful. The use of a remote magnetic navigation system in the context of a 3D geometric model has been shown to lead to high success rates of catheter ablation of various substrates, including AVNRT in complex CHD.[23] This system was used in 2 patients with D-TGA after an atrial switch procedure in our series. Voltage gradient mapping is a relatively new technique used to visualize the slow pathway. It has been reported in a few adult[24] and pediatric patients[25] and shows promise in guiding the placement of safe lesions and reducing the number of applications. More experience with this technique is required to prove its use. However, it may be useful when the exact anatomy of the atrioventricular node is uncertain in patients with complex CHD.

Limitations

This study has several limitations. As with every retrospective study, data are incomplete in certain cases. Information on the persistence of slow pathway conduction after ablation was not always available, and therefore this parameter was not analyzed. The approach was not uniform in all centers, and the exact criteria of documentation of AVNRT, especially of the atypical cases, were not available. Also, the exact location of the ablation lesions was not always available. The results of ablation of AVNRT in patients with complex anatomy represent the local experience and may differ significantly between centers with large versus small patient volumes. The use of a 3D mapping system was not uniform. However, similar patients in each group underwent the procedure with fluoroscopic versus nonfluoroscopic mapping.

Conclusions

AVNRT can complicate the course of patients with CHD, especially in patients with right heart pressure or volume overload. The outcome of catheter ablation is favorable in patients with simple CHD. Patients with complex CHD have increased risk of procedural failure and atrioventricular block. The presence of atypical AVNRT and the need for use of unconventional approaches are potential risk factors for failure. Cryoablation should be considered when the exact anatomy of the atrioventricular node cannot be precisely defined.

Appendix

From the Children's Mercy Hospital, Kansas City, MO (J.P., D.J.B., S.T.-D., D.C.); Georg-August University Medical Centre, Gottingen, Germany (U.K., T.P.); Department of Arrhythmia and Cardiac Pacing, Cardiocentro Pediatrico William Soler, Havana, Cuba (M.C.); Deutsches Herzzentrum, Munich, Germany (M.T., G.H.); Seattle Children's Hospital, WA (S.S., M.W.); Department of Pediatric Cardiology, University of Louisville, KY (C.J.); Sacred Heart Children's Hospital, Spokane, WA (C.A.); Mitera Children's Hospital, Maroussi, Greece (D.A.); The Children's Heart Clinic and Midwest Adult Congenital Cardiac Clinic, Minnesota, MN (C.C.); Hungarian Institute of Cardiology, Budapest, Hungary (L.K.); The Children's Hospital of Iowa, University of IA Hospital and Clinics (I.L.); American Family Children's Hospital, University of Wisconsin-Madison (N.V.B.); Children's Heart Centre, Prague, Czech Republic (J.J., P.K.); St. Louis Children's Hospital, Washington University, MO (J.S.); and Evelina Children's Hospital, London, United Kingdom (E.R.).

Disclosures

Dr Janusek was supported by the Ministry of Health, Czech Republic–conceptual development of research organization, University Hospital Motol, Prague, Czech Republic 00064203. Dr Silva received grant support from Medtronic, research support from St Jude Medical, and a founder of Medical Holography Solutions. Dr Kubus was supported by MH CZ – DRO, University Hospital Motol, Prague, Czech Republic 00064203. The other authors report no conflicts.

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Circulation
Arrhythmia and Electrophysiology

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for the Pediatric and Congenital Electrophysiology Society

Circ Arrhythm Electrophysiol. 2017;10:
doi: 10.1161/CIRCEP.116.004869
Circulation: Arrhythmia and Electrophysiology is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231

Print ISSN: 1941-3149. Online ISSN: 1941-3084