

Radiofrequency catheter ablation of accessory pathways in patients with Ebstein's anomaly: At 8 years of follow-up

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Abstract

Background: Data regarding long-term follow-up of radiofrequency catheter ablation (RFCA) of accessory pathways (APs) in patients with Ebstein's anomaly (EA) are limited. The procedures are challenging due to multiple or wide APs.

Methods: Analysis was performed on clinical and periprocedural data of patients with EA referred to the centre in order to perform catheter ablation of AP. The group consisted of 22 patients (female 40.9%, mean age 33.6 ± 19.1 years). The follow-up utilized electrocardiogram and Holter monitoring.

Results: Twenty-two patients had 33 accessory pathways (8 patients had multiple APs, 11 patients broad AP). Twenty-nine different arrhythmias were ablated: 20 orthodromic atrioventricular reciprocating tachycardia (O-AVRT), 5 antidromic atrioventricular reciprocating tachycardia (A-AVRT), 3 slow/fast atrioventricular nodal reentry tachycardia (s/f AVNRT) and 1 cavotricuspid-isthmus-dependent atrial flutter (CTI-AFL). In 3 (13.6%) patients multiple ablation targets for RFCA ablation were observed. The acute procedural success rate after the first RFCA performed was: 100% for AVNRT, 77.3% for APs and 50.0% for CTI-AFL ablation. Follow-up (mean 95.7 ± 49.8 months) was completed in 86.4% of patients. One patient had paroxysmal atrial fibrillation not targeted during ablation. One patient died due to heart failure 12 years after RFCA. Three patients who underwent RFCA of accessory pathways in the mid-1990s were lost in follow-up.

Conclusions: Radiofrequency ablation in patients with EA is challenging but safe and have a high short-term as well as long-term success rate. (Cardiol J 2017; 24, 1: 1–8)

Key words: Ebstein's anomaly, accessory pathway, Wolff-Parkinson-White syndrome, catheter ablation

Introduction

Ebstein's anomaly (EA) is a rare congenital heart disorder that accounts for < 1% of all patients with congenital heart disease and is characterized by apical

displacement of the posterior and septal leaflets of the tricuspid valve with discontinuity of the central fibrous body and septal atrioventricular ring [1]. Arrhythmias are the main symptom in adolescents and adults as well as a major clinical problem [2].

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Wolff-Parkinson-White (WPW) syndrome is more frequently associated with EA than with any other congenital heart defect [3–5]. According to the existing literature, up to 50% of patients with EA have accessory, frequently multiple pathways (AP) [4, 6, 7]. Abnormal anatomy can also result in activation abnormalities including delayed intraatrial conduction and right bundle branch block [8, 9].

According to current guidelines, patients with EA and relevant arrhythmias should be referred for an electrophysiological study followed, if needed, by ablation therapy, or surgical treatment of the arrhythmias in the case of a planned heart surgery [10]. Moreover, the current guidelines concerning the management of patients with supraventricular arrhythmias suggest that EA might present a sudden cardiac death risk factor [11].

Abnormal, fractionated potentials, enlarged cavities of the heart and the presence of multiple APs make the ablation procedure more complicated and challenging.

There is a limited number of reports published on ablation results in patients with EA and WPW. The published data report small groups of patients and relatively short follow-up (from 12 ± 7 to 54 ± 34 months) [12–14].

The aim of this study was to analyse the safety and efficacy of radiofrequency catheter ablation (RFCA) of accessory pathways in longer follow-ups within one of the largest cohorts of ablated patients with EA.

Methods

Study patients

The group consisted of 22 patients with EA and who had been referred to our centre as a tertiary, experienced centre in ablative therapy of congenital heart disease. The patients were relatively young — mean age 33.6 ± 19.1 years and predominantly male (58.1%).

Definitions

In order to diagnose EA, the apical displacement of the commissure between the septal and posterior leaflets of the tricuspid valve in relation to the anterior mitral valve leaflet by $> 8 \text{ mm/m}^2$ body surface area had to be observed [15].

Multiple APs (or an AP with multiple ventricular or atrial insertions) and broad AP were defined as previously described [16, 17].

The correct ablation site in patients with broad or multiple AP was identified on the basis of a change in the surface of electrocardiographic (ECG) morphology and/or a delay in the local

activation time during radiofrequency (RF) application [17].

Atrial flutter required the cavotricuspid isthmus (CTI-AFL) to be the part of a macroreentrant right atrial tachycardia [5].

The following serious adverse events attributed to catheter ablation were defined: death, atrioventricular block, cardiac perforation, coronary artery involvement or thromboembolic events.

Procedure

An electrophysiological study was performed in the fasted and sedated state. All patients signed an informed consent form prior to the ablation procedure. Heparin bolus was given at a dosage of 50–100-IU/kg, and after each consecutive hour — a heparin bolus of 1000 IU. Intracardiac bipolar electrograms as well as 12-lead ECG were recorded digitally. All measurements were performed using on-screen electronic callipers at a sweep speed of 100–200 mm/s and gain setting of 0.1–0.2 mV/cm. The following catheters, in accordance with the study protocol, were used: (i) a 7 F steerable, diagnostic quadripolar catheter with 2–5–2 mm inter-electrode spacing (Marinr MCXL, Medtronic, Inc., Minneapolis, MN, USA) placed in the coronary sinus; (ii) ablation catheters: an 8 F irrigated tip catheter or 8 mm tip DS catheter. In the mid-1990s, three diagnostic electrodes were used. In use for this study was the 3-dimensional system CARTO XP, CARTO 3 system (Biosense Webster, Inc., Diamond Bar, CA, USA) or EnSite™ NavX (St. Jude Medical, St. Paul, Mn, USA) and long sheath routinely, since 2002.

Follow-up

The follow-up data were obtained from hospital and outpatient clinic records. The evaluation of the patients comprised: physical examination, a 12-lead ECG, echocardiography and Holter monitoring performed in all patients a minimum of 4 times.

Holter monitoring was performed at intervals of 6 months during the first year subsequent to the last successful RFCA procedure and at intervals of 12 months thereafter.

Patients were systematically asked about symptoms of recurrent arrhythmias and encouraged to notify the clinic in the case of recurrence at any time during the follow-up period. Patients with recurrent arrhythmia were offered a repeat ablation.

Statistical analysis

Categorical variables were expressed as numbers and percentages, continuous variables as median and range or mean with 1 standard de-

Table 1. Patient characteristics.

Pt.	Sex	Age	Age of first arrhythmia episode	Ablation targets	Total no. of ablations	Reason for multiple procedures
1	M	18	16	1 broad AP, A-AVRT	3	AP
2	F	42	7	2 APs, O-AVRT	1	–
3	M	33	1	2 APs, O-AVRT	1	–
4	F	13	3	1 broad AP, O-AVRT	1	–
5	F	13	1	1 broad AP, O-AVRT, s/f AVNRT	1	–
6	M	18	NA	1 broad AP, O-AVRT	1	–
7	M	68	5	1 broad AP, O-AVRT, s/f AVNRT	3	AP
8	M	53	NA	1 AP, O-AVRT	1	–
9	F	49	NA	2 APs, 1 broad AP, O-AVRT	1	–
10	M	27	6	2 APs, O-AVRT	3	AP
11	F	47	NA	1 AP, O-AVRT, s/f AVNRT, CTI-AFL	2	CTI-AFL
12	F	18	1	2 APs, O-AVRT	1	–
13	F	74	NA	1 broad AP, O-AVRT	3	AP
14	M	14	11	2 APs, 1 broad, O-AVRT, A-AVRT	1	–
15	F	53	13	1 AP, O-AVRT	1	–
16	M	25	NA	1 AP, A-AVRT	1	–
17	F	60	NA	1 broad AP, O-AVRT	1	–
18	M	34	NA	1 broad AP, O-AVRT	3	AP
19	M	29	NA	1 AP, O-AVRT	2	AP
20	M	19	NA	3 APs	1	–
21	M	17	NA	3 APs, A-AVRT, O-AVRT	4	AP
22	M	16	NA	1 broad AP	5	AP

A-AVRT — antidromic atrioventricular reentrant tachycardia; AP — accessory pathway; AV — atrioventricular node; s/f AVNRT — slow/fast atrioventricular nodal reentrant tachycardia; CTI-AFL — cavotricuspid isthmus-dependent atrial flutter; F — female; M — male; NA — not available; O-AVRT — orthodromic atrioventricular reentrant tachycardia

viation, respectively. Categorical variables were compared with the use of χ^2 test. A p value < 0.05 was considered statistically significant. Analyses were performed using SPSS software.

Results

Study population

Twenty-two patients with EA (female 40.9%, mean age 33.6 ± 19.1 years) were admitted to a centre experienced in ablative therapy of congenital heart disease in order to perform RFCA of accessory pathways between 1993 and 2015 (Tables 1, 2).

The patients’ evaluation comprised of clinical history, physical examination, a 12-lead ECG,

Table 2. Procedural data.

Parameters	AP (n = 22)
Previous cardiac surgery	4 (18.2%)
Age at first ablation [years]	33.6 ± 19.1
One ablation, in our lab	14 (63.6%)
Two ablations in our lab	5 (22.8%)
Three ablations in our lab	3 (13.6%)
Number of ablations outside our centre	10 (45.5%)
Two ablations outside our centre	5 (22.8%)
Total number of interventions	40
3D system in our lab	18 (81.8%)
Long sheath in our lab	15 (62.5%)
Long-term success	95.5%

AP — accessory pathway; 3D — three-dimensional



Figure 1. Baseline 12 lead electrocardiogram (A) and electrocardiogram after the radiofrequency ablation of 3 accessory pathways (B).



Figure 2. 12 lead electrocardiogram (A) and intracardiac recording (B) of cavotricuspid isthmus-dependent atrial flutter conducted through 2 accessory pathways, left posteroseptal (1) and broad right inferior/paraseptal (2).

24-h Holter-ECG monitoring, echocardiography, electrophysiological study parameters and coronary angiography in patients with the risk factors for ischemic heart disease. Mean left ventricular ejection fraction was 62.2%.

Sixteen patients underwent ablation with the use of CARTO XP or CARTO 3 system (Biosense Webster) and 2 patients using EnSite™ NavX (St. Jude Medical). Four patients treated in the 1990s underwent ablation without a 3-dimensional navigation system.

The mean procedure time was 155.0 ± 74.0 (range 60–240) min, the mean fluoroscopy time was 10.1 ± 3.6 min and mean fluoroscopy exposure 90.5 ± 82.2 mGy².

Twenty-two patients with AP had 40 RFCA (mean 1.8; range 1–4), 33 of them at the centre (mean 1.5; range 1–3). Eighteen (81.8%) patients had overt pre-excitation, 1 (4.6%) intermittent and 3 (13.6%) had concealed AP. Thirty-two APs (3 left-sided) were targeted with ablation.

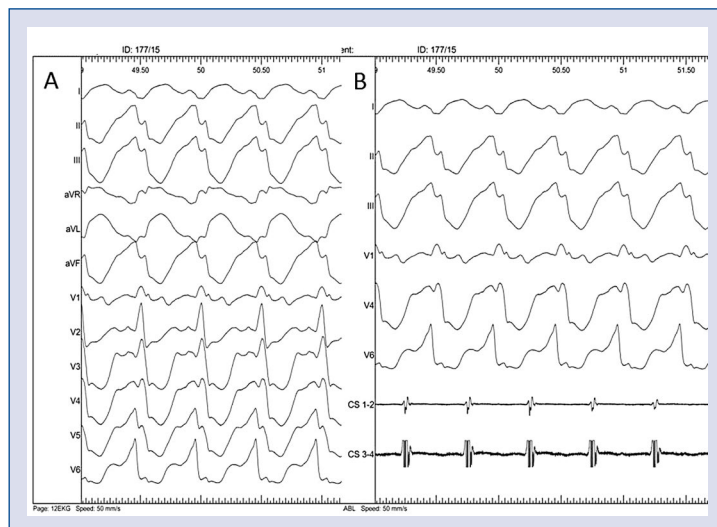


Figure 3. 12 lead electrocardiogram (A) and intracardiac recordings (B) of antidromic atrioventricular reentrant tachycardia conducted through left posteroinferior accessory pathway.

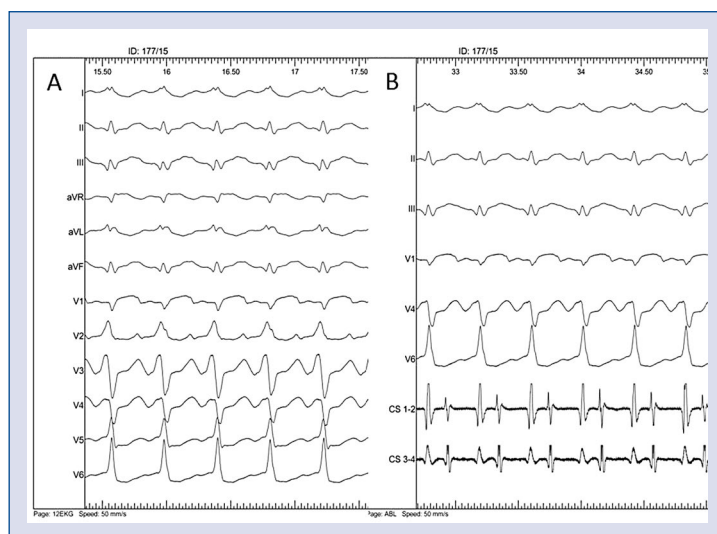


Figure 4. 12 lead electrocardiogram (A) and intracardiac recordings (B) of orthodromic atrioventricular reentrant tachycardia conducted through left posteroinferior accessory pathway.

Eight (36.4%) patients had multiple APs, 11 (50.0%) patients had broad AP. Three (13.6%) patients had 3 APs (Figs. 1–4).

All patients with APs had palpitations or documented arrhythmias (mean age at the time of first episode 6.9 ± 5.6 ; range 1–16 years). Twenty patients had documented orthodromic atrioventricular reciprocating tachycardia (O-AVRT), 5 antidromic atrioventricular reciprocating tachycardia (A-AVRT), 3 patients had slow/fast atrioventricular nodal reciprocating tachycardia (s/f AVNRT), 1 atrial flutter (CTI-AFL).

In 1 patient with AP and CTI-AFL, there was a failure to block the CTI during first procedure, (Table 1, Patient no. 11) during the second procedure the CTI was successfully blocked.

Three patients had O-AVRT and A-AVRT episodes, 2 patients only A-AVRT.

One patient with 3 APs had ventricular fibrillation (VF) episode and was considered as an end point of the first arrhythmic event. Eight (36.4%) patients with APs had more than one type of arrhythmia, 1 patient had 3 types (4.5%). Four patients had a history of cardiac surgery (Fig. 5; Table 3).

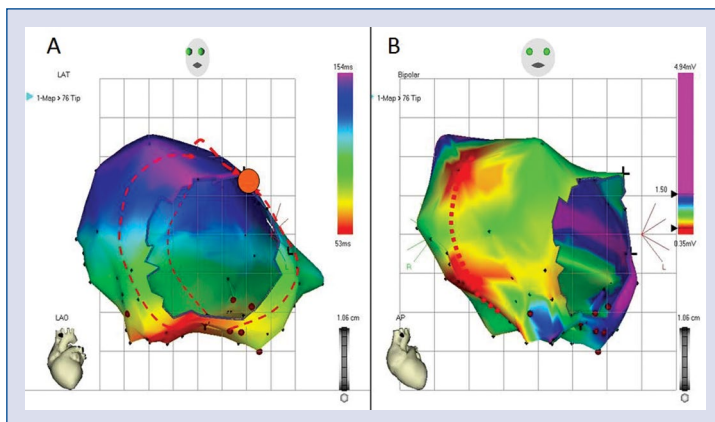


Figure 5. 18-year-old patient with Ebstein’s anomaly after atrial septal defect and ventricular septal defect operation. Electroanatomical CARTO map of orthodromic atrioventricular reentrant tachycardia (A), bipolar voltage of the right atrium shows, probably post incisional, low voltage area (B).

Table 3. Patients with a history of cardiac surgery.

Pt.	Number of surgical procedures prior to RF	Surgical procedures	Arrhythmias	Ablation	Number of RF ablations	Time from surgery to first RF [years]	Arrhythmia during follow-up
1	1	ASD closure, VSD closure, PDA closure	O-AVRT	MAP, inferoparaseptal and right inferior	1	17	No
2	1	TVR	A-AVRT	Large AP, inferoparaseptal-inferior	3	1	No
3	1	Coarctation of aorta	O-AVRT	Inferoparaseptal and CTI line (uncompleted line)	2	36	No
4	1	TVR, cryoablation	O-AVRT, A-AVRT	Large AP, right inferior/right antero-inferior	3	NA	AF

AF — atrial fibrillation; AP — accessory pathway; ASD — atrial septal defect; CTI — cavotricuspid isthmus; MAP — multiple accessory pathway; NA — not available; O-AVRT — orthodromic atrioventricular reciprocating tachycardia; PDA — patent ductus arteriosus; RF — radiofrequency; TVR — tricuspid valve replacement; VSD — ventricular septal defect

The success rate of RF ablation (RFA) in patients with APs after the first procedure conducted in this centre was 77.3% and after the last RFA the rate was 95.5%.

In the group of 7 patients who underwent 3 or more RF procedures, there were 5 patients with whom the first 2 attempts which were not performed at this centre.

Three patients with APs and induced s/f AVNRT also underwent ablation of the slow AV node pathway. No major early or late complications were observed.

Follow-up

Long-term follow-up was done in 19 (86.4%) patients with a median of 95.7 ± 49.8 (range 10–204) months after the first ablation procedure. Three patients who were treated in the mid-1990s were lost to follow-up.

After successful ablation, the antiarrhythmic drugs were discontinued.

One patient died due to heart failure during the follow-up at the age of 72 one year after a third unsuccessful RFA and 13 years after unsuccessful

surgical cryoablation, which was performed during implantation of tricuspid valve prosthesis and suture of an atrial septal defect.

One patient with WPW and VF, after successful RFA of the APs, did not experience any arrhythmia episodes.

The long-term success rates were 95.5% of patients with APs, and 100% in patients with AVNRT and CTI-AFL.

Discussion

There remains a limited number of previously published reports concerning long-term follow-up data in patients with EA and arrhythmias treated RFA.

Data of patients with EA and RFCA of APs which were published previously reported a relatively short follow-up from 12 ± 7 to 54 ± 34 months [3, 4, 12, 14].

Apparently the present study has the longest follow-up and one of the largest cohort of patients with EA and APs.

The clinical outcome of this study indicates that in long-term follow-up after successful ablation, the majority of patients were free of any arrhythmia episodes, ablated as well as newly developed.

The ablation of AP in patients with EA is challenging but when performed in an experienced centre the long-term follow-up is satisfactory.

Some authors have suggested the use of intracardiac echo to define the true location of tricuspid annulus (TA) via the right coronary artery (RCA) [18].

Others have proposed the use of guidewire or diagnostic catheter inserted into the RCA for proper delineation or mapping of TA [14].

These two prior techniques were not used in this study. Since 2002, this clinic has routinely used a 3-dimensional system, 8 F irrigated tip catheter and long sheaths and, if necessary, with 180 degree curvature.

The presented outcomes (77.2% efficacy after the first and 95.5% after the last procedure) are comparable to those reported in an earlier series [4, 6, 12].

The percentage of patients with EA and multiple accessory pathway in this study was similar to those presented by other authors [3, 5, 12, 14].

Contrary to other studies, however, a very high percentage of patients with broad APs (50%) was found. The presence of a broad AP in a particular patient can be subjective is consequently difficult to quantify.

The current guidelines itemised EA as a risk factor for VF in patients with AP but in a previous study conducted on the largest cohort of patients with aborted sudden cardiac death and APs thus far, EA was not defined as a statistically significant risk factor for VF [11, 19].

Limitations of the study

This is a retrospective analysis and therefore several limitations must be taken into account. The main limitations of the present study are the relatively small number of patients enrolled. Secondly, not all detailed data concerning ablation procedures were available. Thirdly, precise echocardiographic data of patients who underwent ablation in the 1990s were not always available. Fourthly, some patients were lost to follow-up.

Conclusions

The RFAs in patients with EA and APs are challenging but most arrhythmias are amenable to ablation and long-term follow-up is well recommended.

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Conflict of interest: None declared

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