

The clinical course and risk in patients with pseudo-Mahaim fibers

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Abstract

Background: *Pseudo-Mahaim (AP-M) fibers are a rare variant of atrioventricular (AV) accessory pathways. Atriofascicular and atrioventricular accessory connections are characterized by slow conduction and decremental properties. Dual physiological AV node pathways, slow and fast, are observed in a large number of patients with AP-M. Therefore, there is substrate for AV nodal reentrant tachycardia (AVNRT) in addition to antidromic AV reentrant tachycardia (AVRT) with left bundle branch block (LBBB)-like morphology. Other arrhythmias such as atrial fibrillation (AF) or atrial flutter (AFL) and ventricular fibrillation (VF) are also observed. We analysed the occurrence of arrhythmias in a group of patients with AP-M treated in our department.*

Methods: *We evaluated 27 patients (12 women) aged 14–53 years (mean age 25.6 years) with AP-M. The clinical course in these patients, in particular with regard to the occurrence of arrhythmias, was analysed. Patients with dual AV node properties were compared to patients without such findings.*

Results: *We found dual AV node properties in 18 patients (Group 1), while 9 patients had fast pathway only (Group 2). Twenty-six patients presented with AVRT, 2 patients with AVNRT, 3 patients with AF, 1 patient with AT, 2 patients with AFL, and 3 patients with VF. In 2 patients, AP-M were seen in an atypical area. In one patient, the pathway connected the right atrium with the left ventricle (septal region), and in the other patient it connected the left atrium with the left ventricle (left anterior region).*

Conclusions: *The majority of AP-M was right-sided. Two thirds of patients with AP-M had anatomical substrate for AVNRT (fast/slow pathway AV node). VF or asystole occurred in 10% of patients. (Cardiol J 2008; 15: 365–370)*

Key words: pseudo-Mahaim fibers, atriofascicular pathway, radiofrequency catheter ablation

Introduction

Atrioventricular (AV) pathways characterized by slow antegrade conduction comprise less than 3% of all accessory pathways [1]. These are called

pseudo-Mahaim fibers (AP-M) and include atriofascicular pathways connecting the atrium with the right His bundle branch and AV pathways connecting the atrium with the ventricular muscle [2–4]. These pathways are most commonly right-sided

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Received: 17.11.2007

Accepted: 1.06.2008

Table 1. Group 1: patients without dual AVN properties.

No.	Gender	Age	Arrhythmia	Disease	Treatment
1	M	18	AVRTa		RF
2	F	34	AVRTa, VF		RF
3	F	23	AVRTa		S → RF
4	M	18	AVRTa	MVP	RF
5	M	23	AVRTa		RF
6	M	30	AT*		RF
7	M	53	AVRTa, AF, AFL	ASD	RF
8	F	27	AVRTa		RF
9	F	32	AVRTa		S → RF
		28,7			

AVRTa — antidromic atrioventricular reentrant tachycardia; AT — atrial tachycardia (*with no retrograde conduction in EP study); AF — atrial fibrillation; AFL — atrial flutter; VF — ventricular fibrillation; MVP — mitral valve prolapse; ASD — atrial septal defect; S — surgery, RF — radiofrequency current ablation

and located within the free wall (of the atrium and ventricle), less frequently within the septum, and very rarely they are left-sided [5]. During sinus rhythm, preexcitation is absent or mild [6]. This anomaly predisposes to episodes of antidromic AV reentrant tachycardia (AVRTa) with left bundle branch block (LBBB)-like morphology and retrograde atrial activation through the physiologic His-Purkinje system and the fast pathway within the AV node (AVN) or through another “fast” accessory pathway (AP) conducting in a retrograde direction [1, 7]. Radiofrequency (RF) current ablation may be performed within both proximal and distal part of the pathway [8–10].

Episodes of antidromic tachycardia are often accompanied by other arrhythmia such as AV node reentrant tachycardia (AVNRT), atrial fibrillation (AF), atrial tachycardia (AT), atrial flutter (AFL), and ventricular fibrillation (VF) [11].

Methods

We analyzed the clinical course and risk in 27 patients aged 14–53 years (mean age 25.6 years), including 12 women, who were evaluated and treated for recurrent arrhythmia in the Institute of Cardiology, Warszawa-Anin, and were found to have AP-M during the electrophysiologic study (EPS).

The patients were divided in two groups based on the presence of AVN pathways, slow and fast, as found during EPS. Group 1 included patients without slow pathway within AVN (Table 1), and Group 2 included patients with both slow and fast pathways within AVN (Table 2). We evaluated whether the presence of dual AVN properties affected the analyzed parameters.

We analyzed data from:

- history and physical examination in regard to arrhythmia and coexisting conditions, and in particular the occurrence of AVRT, AVNRT, AT, AF, and VF;
 - laboratory testing (12-lead ECG during sinus rhythm and arrhythmia, standard Holter monitoring, exercise test, and echocardiography);
 - EPS before and after RF ablation, including data regarding the RF ablation procedure itself.
- One or combination of the following electrophysiologic characteristics of AP-M were found [12–14]:
- slow antegrade conduction with decrement, or increase in AV conduction time by more than 30 ms with progressively faster pacing rate;
 - the presence of activation signal [high frequency spike (M)] along the pathway;
 - the ability to induce conduction block within the pathway by mechanical pressure (e.g. compression using electrode);
 - the absence of retrograde conduction through the AP during EPS;
 - automaticity (both spontaneous and evoked, e.g. by application of RF current).

The two subgroups were compared in regard to the evaluated parameters using both parametric and nonparametric tests (Fisher test, Wilcoxon test).

The study was approved by the local bioethical committee and all patients gave their informed consent.

Results

The accessory pathways were right-sided in 25 patients. In one patient, the ventricular end of

Table 2. Group 2: patients with dual AVN properties.

No.	Gender	Age	Arrhythmia	Disease	Treatment
1	M	39	AVRTa		RF
2	M	19	AVRTa		RF
3	M	18	AVRTa		RF
4	M	22	AVRTa		RF
5	F	19	AVRTa		RF
6	F	23	AVRTa	MVP	RF
7	F	25	AVRTa	MVP	RF
8	M	19	AVRTa		RF
9	M	14	AVRTa	Mild TI	RF
10	F	20	AVRTa		RF
11	M	19	AVRTa	MVP	RF
12	F	46	AVRTa, AVNRT		RF
13	M	40	AVRTa, VT*		S → RF
14	F	17	AVRTa	MVP	RF
15	M	17	AVRTa, AVNRT, AF, AFL, VF		RF
16	F	26	AVRTa, VF	MVP, PFO	RF
17	F	16	AVRTa		RF
18	M	34	AVRTa, AF		RF
		24,1			

AVNRT — atrioventricular node reentrant tachycardia; TI — tricuspid insufficiency; PFO — patent foramen ovale; VT — ventricular tachycardia (*focal — originating from the left ventricle); remaining abbreviation as in Table 1

AP (M signal) was ablated on the left side of the interventricular septum. In another patient, AP was connecting the left atrium with the upper part of the left ventricle. The precise localization of APs in patients in both subgroups is depicted in Figure 1.

None of the evaluated parameters reached statistical significance (Table 3).

Overall, AVN show slow antegrade conduction in two thirds of patients. In this subset (Group 2), spontaneous AVNRT was noted in 2 patients (11.1%), but was induced during EPS in 10 patients (55.6%). RF ablation of the slow pathway within AVN was not performed in patients in whom sustained AVNRT was not induced during EPS.

In one patient in Group 1 (with several dozens of arrhythmia episodes per year, the longest lasting for approx. 70 h), an episode of AVRTa degenerated into VF. In one patient in Group 2, VF occurred following an attempt of electrical cardioversion (AVRTa → intravenous amiodarone → AVRTa → electrical cardioversion → VF), and another patient (with a history of AVRTa episodes and syncope) suffered cardiac arrest with VF detected during resuscitation. Notably, asystole seen in 3 cases was related to various antiarrhythmic drug therapy during attempts to restore sinus rhythm in two of these patients.

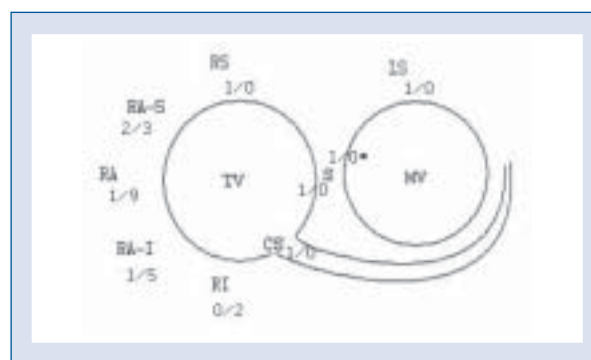


Figure 1 Schematic representation of the atrioventricular valve annuli and the coronary sinus with localization of AP-M. Data indicate the number of patients with AP in a given location in Group 1/Group 2. TV — tricuspid valve; MV — mitral valve; CS — coronary sinus. Accessory pathways: RS — right-sided superior; RA-S — right-sided anterosuperior; RA — right-sided anterior; RA-I — right-sided anteroinferior; RI — right-sided inferior; S — septal (*one of the septal pathways had the ventricular end on the left side of the interventricular septum); CS — coronary sinus; LS — left-sided superior.

In Group 1, three out of 9 patients underwent one ablation procedure (surgical or RF) prior to the successful RF ablation, and in Group 2 seven out of 18 patients underwent 1 to 5 (!) such procedures in various centers.

Table 3. Results — overall and in the two groups.

Patient characteristics	Overall	Group 1	Group 2	p
Number of patients	27	9	18	
Women	12 (44.4%)	4 (44.4%)	8 (44.4%)	NS
Mean age ± SD	25.6 ± 9.9	28.7 ± 10	24.1 ± 9	NS
AVRTa	26 (96.3%)	8 (88.9%)	18 (100%)	NS
Mean heart rate ± SD during AVRTa	196.3 ± 25	200 ± 18	195.3 ± 26	NS
AVNRT	2 (7.69%)	0 (0%)	2 (11.11%)	NS
AF	3 (11.11%)	1 (11.11%)	2 (11.11%)	NS
AT	1 (3.7%)	1 (11.11%)	0 (0%)	NS
AFL	2 (7.41%)	1 (11.11%)	1 (5.56%)	NS
VF	3 (11.11%)	1 (11.11%)	2 (11.11%)	NS
Asystole during antiarrhythmic treatment	3 (11.11%)	2 (22.22%)	1 (5.56%)	NS
Syncope (except VF and asystole)	6 (22.22%)	3 (33.33%)	3 (16.67%)	NS
Near-syncope	12 (44.44%)	2 (22.22%)	10 (55.56%)	NS
Structural heart disease	8 (29.63%)	2 (22.22%)	6 (33.33%)	NS
Number of procedures (S and RF) prior to successful ablation	16	3	13	NS

Abbreviation as in Table 1

Discussion

Pseudo-Mahaim fibers, except in very rare cases, are right-sided pathways. Most of their electrophysiologic characteristics suggest that these are an equivalent (replica) of the physiological conduction system, or essentially second (and in single reported cases the only one), right-sided AVN with a (right) His bundle branch [3, 4, 7, 10, 12].

We demonstrated the presence of a slow pathway within AVN in most (2/3) patients in our study, which is consistent with findings of other authors. Interestingly, only a few patients experienced spontaneous AVNRT (2 patients), while during EPS this arrhythmia was induced in 10 patients (56%). Perhaps AVNRT in fact occurs more commonly than it is diagnosed and/or documented in ECG. This underrated might result from a number of causes. One reason might be that in the presence of AP-M, AVNRT might be conducted to ventricles through the accessory pathway (bystander) and thus would be difficult to distinguish from AVRTa, as both arrhythmias present with wide QRS complexes with LBBB-like morphology (Fig. 2). AVNRT might also convert into AVRTa or AF/AFL (Fig. 3) [11]. Finally, AVNRT might have also been less common due to a relatively young age of our patients (mean 25.6 years).

Low number of patients in both groups, resulting from rarity of these phenomena, does not allow definite conclusions to be made, but our findings

suggest that no significant differences in the clinical course exist between patients with AP-M and either fast pathway only or both slow and fast pathways within AVN. The qualitative difference, i.e. the occurrence of AVNRT, has been already mentioned.

A direct life-threatening situation in all these patients is VF. This arrhythmia occurs with a frequency similar to that in patients with Wolff-Parkinson-White syndrome and AVRT. Asystole seen in 3 patients may be related to physician doubts regarding the diagnosis of AVRT and VT. In case of such doubts, the arrhythmia was treated more aggressively, often with several antiarrhythmic drugs. Also of note, patients with AP-M often underwent many unsuccessful ablation procedures before referral. This is due to a low prevalence of this condition and thus small experience of electrophysiologists in this regard, but also from a relative difficulty of performing ablation of AP-M.

Patients with AP-M (“dual AVN”) present with various kinds of arrhythmia, with AP-M being an element of a macro-reentry circuit or a bystander. Particular electrophysiologic characteristics of this condition result in difficulties related to diagnosis and treatment. For some years, electrophysiologic characteristics of the “Mahaim syndrome” was interpreted in a concordant but wrong way even by the most renowned electrophysiologists. Even now, when it seems we finally know how to interpret these phenomena correctly, repeated ablation procedures are

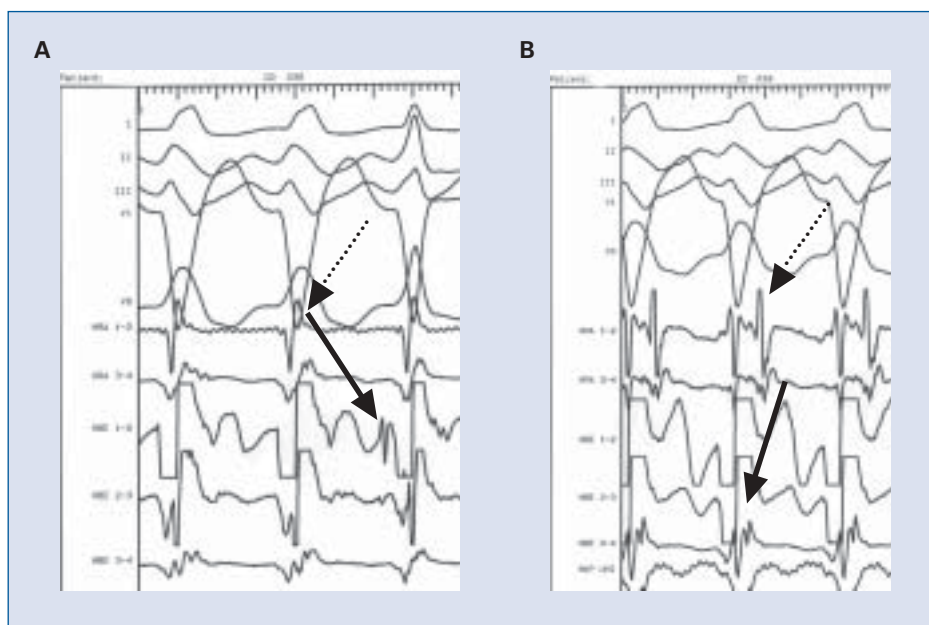


Figure 2. Pseudo-Mahaim fibers as a bystander. **A.** AVNRT conducted to ventricles through Mahaim fibers (first and second QRS complex, whereby no His bundle signal is seen) and a fusion beat with impulse conducted both through Mahaim fibers and AVN (third QRS complex with visible His bundle signal — solid arrow). The atrial signal is hidden within the QRS complex (dashed arrow). **B.** Antidromic AVRT. The His bundle signal follows the ventricular signal (solid arrow), and the atrial signal is seen even later after the QRS complex (dashed arrow).

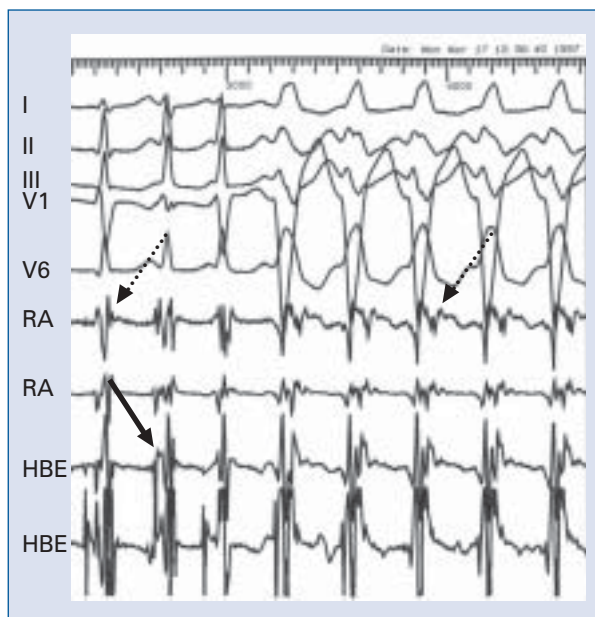


Figure 3. AVNRT (first 3 QRS complexes) conducted through physiological pathways (normal width QRS complexes, a visible His bundle signal — solid arrow, the atrial signal hidden within the QRS complex — dashed arrow) gradually converts into antidromic AVRT in subsequent beats (the His bundle signal becomes hidden within the QRS complex, and the atrial signal follows QRS complexes — dashed arrow).

necessary more frequently than in other clinical syndromes related to the presence of APs.

Conclusions

Accessory AV pathways with slow antegrade conduction are mostly right-sided (92.6%), but these may also be located on the left side.

Two thirds of patients with AP-M had anatomical substrate for AVNRT — dual AV node properties with fast and slow pathways.

Life-threatening events, such as VF and asystole, were seen in more than 10% of patients with AP-M.

Definitive antiarrhythmic treatment in these patients required, on average, performing 1,6 ablation procedures (surgical or RF).

The analyzed groups did not differ with regard to the clinical course of spontaneously occurring arrhythmia.

Acknowledgements

The authors appreciate help of dr Piotr Jędrusik with preparation of the authorized English version of the manuscript.

The authors do not report any conflict of interest regarding this work.

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