The 12-Lead ECG in Patients with Mahaim Fibers

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The aim of this review article is to discuss the electrocardiographic presentation of the so called variants of pre-excitation ("Mahaim fibers") during sinus rhythm and tachycardia.

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decrementally conducting accessory pathway; mahaim fiber; atriofascicular pathway;
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INTRODUCTION

Accessory pathways (AP) with long and decremental anterograde conduction have been the subject of extensive debate about their anatomic structure,^{1–4} location,^{5–9} related arrhythmias,^{8,9} electrophysiologic properties,^{10–12} ablative techniques,^{10,13–15} and automaticity.^{1,5} Less attention has been given to the 12-lead ECG. Preexcitation is reported to occur from 0% to 30%,^{13,14,16,17} and apart from the absence of Q waves in the left precordial leads¹⁸ no specific QRS pattern has been described. The purpose of this article is to review the ECG patterns in patients with Mahaim fibers during sinus rhythm and tachycardia.

ECG DURING SINUS RHYTHM

Atriofascicular Pathways and Long Atrioventricular Fibers

ECG during sinus rhythm was considered to be normal in the majority of patients with atriofascicular (AF) pathways and patients with long atrioventricular (AV) decrementally conducting AP. These patients usually do not show overt preexcitation. The contribution to ventricular depolarization from impulse conduction over these decremental bypass tracts during sinus rhythm is usually small and restricted to the anteroapical right ventricle. As a result the 12-lead ECG may show a minimal preexcitation pattern. The reported incidence of minimal preexcitation is low. Bardy et al.¹⁹ and Klein et al.²⁰ did not find it in any of their patients. McClelland et al.²¹ reported that only 1 of his 26 patients displayed preexcitation on the 12-lead ECG. However,

no specific pattern was described. We recently reviewed a large cohort of 33 patients with AF pathways and we found a subtle preexcitation pattern in 24 patients (72%). We found an *rS* configuration in lead III in 20 patients. Another pattern, an *rsR'* in lead III, was found in two patients. In two other patients, preexcitation manifested only with absence of a septal Q wave in leads I and V6. When we realized the prevalence of the *rS* pattern in lead III in our patients, we examined previous reports dealing with decrementally conducting bypass tracts. We did find the *rS* pattern in lead III in many ECGs considered as normal in cardiology journals^{20,22,23} and textbooks.^{24,25} This suggests that the reported low figure of abnormal ECGs in patients with AF pathways is an underestimation.

To validate the *rS* and *rsR'* as abnormal patterns in lead III due to preexcitation of a small region of the right ventricle it was critical to show a positive relationship between those patterns in lead III during sinus rhythm and left-axis deviation during tachycardia with anterograde conduction over the Mahaim fiber (Fig. 1). All 20 patients with an *rS* pattern in lead III had left-axis deviation ($\leq 0^\circ$) during tachycardia. Another important step in validation is to show a clear change in QRS complex configuration after ablation of the decremental AP. Figure 2 depicts most of the patterns of QRS that might emerge after successful ablation of the Mahaim fiber.

Differential Diagnosis of an *rS* Pattern

An *rS* pattern in lead III can be found in normal individuals. This may occur during posterior

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Figure 1. Two patients with Mahaim fibers displaying the *rS* pattern in lead III during sinus rhythm and left-axis deviation during tachycardia.

displacement of the apex leading to S waves in leads I, II, and III (*S1S2S3* pattern²⁶) and in counterclockwise rotation of the heart resulting in *qR* in lead I and *rS* in III. However, in those situations a normal *Q* wave in lead I is likely to be present. We did a survey on 200 ECGs from young individuals with palpitations we found the *rS* pattern in lead III in 6%, but always associated with a *q* wave in lead I. No individual showed an *rS* pattern in lead III combined with the absence of a *q* wave in lead I, a pattern that seems specific for patients with a Mahaim fiber.

Specificity and Predictive Value of the *rS* Pattern

The finding of an *rS* pattern in lead III in 60% of the patients with Mahaim fibers is significantly higher than its occurrence in young persons with palpitations ($P < 0.0001$).

Mahaim fibers comprise approximately 3% of the overt AP.²⁷ Based on the prevalence of AP in the general population²⁸ (0.2%), the prevalence of Mahaim fibers would be 0.5–1:10,000. The specificity of an *rS* pattern in lead III associated with the absence of a septal *q* wave will be close to 90% (if we assume one false positive in 1000 individuals), albeit the sensitivity decreases to 45%.

Latent Mahaim's

A latent form of Mahaim fiber has been described, where preexcitation is only present during antidromic tachycardia and not during both sinus rhythm and atrial pacing. It is sometimes difficult to make the differential diagnosis with ventricular tachycardia. It is important in those cases to deliver late premature atrial beats during tachycardia at a time of low septal right atrium and His bundle refractoriness. If ventricular activation can be advanced a latent Mahaim fiber is likely to be engaged in the circuit.^{29,30}

Short AV Fibers

We studied eight patients having a decrementally conducting AP with a short anatomic course (short AV Mahaim fibers), as assessed by mapping the distal end of the fiber at the tricuspid annulus.³¹ All patients had overt ventricular preexcitation on their baseline 12-lead ECG. In six patients, ventricular preexcitation occurred because of impulse conduction over a short AV Mahaim fiber. Two of the eight patients showed preexcitation due to an associated rapidly conducting AV bypass tract, one right midseptal and one right anteroseptal. After

Figure 2. Five-lead ECG during sinus rhythm with the preablation ECG showing the *rS* pattern in four patients with an atriofascicular pathway. The postablation ECG shows a clear change in QRS configuration.

ablation of those pathways one patient (case 2) showed another preexcitation pattern due to a short Mahaim fiber and patient 7 (with Ebstein's disease) showed no preexcitation (only RBBB) during sinus rhythm with preexcitation becoming apparent during atrial pacing (Fig. 3). In the seven patients with overt preexcitation during sinus rhythm the PR interval was normal in two patients (cases 5 and 6) and short (<0.12 seconds) in five patients. The preexcitation pattern in these patients is indistinguishable from the ECG of patients with a rapidly conducting right-sided AP. No patient with a short AV Mahaim fiber showed a minimal preexcitation pattern. The presence of overt preexcitation in seven of the eight patients can be explained by the large mass of right ventricular myocardium depolarized by the impulse conducted over the short fiber before the arrival of the wavefront proceeding over the His-Purkinje axis.

Fasciculoventricular Pathways

A fasciculoventricular (FV) pathway is the closest structure to match the original description of Dr. Mahaim. In the late 1930s, he described a structure without a clear-cut function—the paraspecific septal conduction pathways, connecting the His bundle to the ventricular myocardium. According to Dr. Mahaim, those fibers could be an alternative to bundle branch-His Purkinje system conduction. Dr. Mahaim tried to understand their functional role by studying the effect of sequentially cutting these connections and observing the resulting changes on the 12-lead ECG pattern, without much success. To this date, FV fibers were never reported to play an active role in tachycardia circuit. However, FV pathways are frequently associated with rapidly conducting bypass tracts, and they must be correctly differentiated from an anteroseptal

Figure 3. Twelve-lead ECGs during anterograde conduction over a short AV Mahaim fiber in the eight patients studied. Small black squares mark lead V₁. Cases 2 and 7 ECGs were recorded after ablation of an associated bypass tract. Case 7 (a patient with Ebstein’s disease) showed preexcitation only during atrial pacing (after ablation of an additional anteroseptal bypass tract, the 12-lead ECG showed sinus rhythm with RBBB).

bypass tract, to avoid harm to AV nodal conduction if they are targeted for catheter ablation.¹⁵ We studied seven patients with FV pathways aiming to describe its electrocardiographic features as well as comparing their surface ECG with patients having midseptal and anteroseptal bypass tracts (Table 1).³²

Patients with FV pathways show a variable PR interval. Josephson³³ describes a normal or short PR interval. Gallagher et al.⁴ reported six patients with FV pathways with a PR interval <0.12 seconds and electrophysiologic evidence of enhanced

AV nodal conduction. In our series the mean PR interval was 0.10 ± 0.01 (range 0.09–0.12) seconds, but no patient had enhanced AV nodal conduction.

Previous studies^{34,35} concentrated on the QRS configuration and delta wave morphology in lead V₁ which in our cohort showed a wide variability. According to our findings FV pathways have overlapping electrocardiographic features with both anteroseptal and midseptal accessory bypass tracts. The mean QRS and delta wave frontal plane axis was normal in all three groups.

Table 1. Comparative Electrocardiographic Findings in 20 Patients with a Midseptal AP, 20 Patients with an Anteroseptal AP and Seven Patients with a FVP

	MS	AS	FVP	P Value
âQRS	NA	NA	NA	
âDELTA	NA	NA	NA	
Angle between QRS and delta wave axis	23° ± 8°	4° ± 8°	24° ± 15°	<0.0001 (AS × FVP)
R/S ratio in lead III	<1 or 1	>1	<1 or >1	
Two interior leads with a negative delta wave	0	0	0	
One inferior lead with a negative delta wave	25%	0%	0%	
Precordial lead transition to R/S ratio >1	V ₂ -V ₃ -V ₄	V ₃ -V ₄	V ₂ -V ₃ -V ₄	
QRS width	0.14 ± 0.008	0.14 ± 0.01	0.12 ± 0.02	<0.0001

âQRS = QRS frontal plane axis; âDELTA = frontal plane axis of the delta wave; AP = accessory pathways; AS = anteroseptal bypass tract; FVP = fasciculoventricular pathway; MS = right midseptal bypass tract; NA = normal axis; ns = not significant. Characters in bold means a higher prevalence of one value over the other.

Figure 4. Twelve-lead ECG of seven patients with a fasciculoventricular pathway diagnosed by an EP study. There is a wide range of presentation, ranging from a near normal QRS complex (5) to a fully preexcited QRS complex (6,7).

Electrocardiographic Similarities with Anteroseptal AP

The R/S ratio in lead III was >1 in four of the seven patients (57%) with FV pathways and in 100% of the patients with an anteroseptal AP ($P = \text{ns}$). In contrast, patients with midseptal AP never showed an R/S ratio >1 .

Patients with FV pathways and patients with anteroseptal bypass tracts never showed a negative delta wave in inferior leads, in contrast with 25% of the patients with a midseptal AP.

Electrocardiographic Similarities with Midseptal AP

Patients with FV pathways and patients with a midseptal bypass tract had a similar angle between the QRS and the delta wave axis in the frontal plane ($24^\circ \pm 15^\circ$ and $23^\circ \pm 8^\circ$, respectively) as compared

with the angle of $4^\circ \pm 8^\circ$ in patients with an anteroseptal bypass tract ($P < 0.0001$) (Fig. 4).

Electrocardiographic Dissimilarities Between FV Pathways, Midseptal, and Anteroseptal AP

The transition to an R/S ratio >1 in the precordial leads occurred mainly in V_2 in patients with FV pathways, V_3 in midseptal pathways, and V_4 in anteroseptal bypass tracts.

The major feature differentiating FV pathways to a septal bypass tract was the QRS width: 0.12 ± 0.02 seconds in the former and 0.14 ± 0.008 and 0.14 ± 0.01 seconds in midseptal and anteroseptal bypass tracts, respectively ($P < 0.0001$).

The ECG during sinus rhythm in patients with FV pathways will show a short PR interval, usually with a minimal preexcitation pattern with a QRS

Figure 5. ECGs of patients 6 and 7 with a fasciculoventricular pathways (FVP) showing a broad QRS complex. They are displayed for comparison with ECG of two Group III patients with midseptal bypass tracts (MS-AP).

width of 0.12 seconds, a normal frontal plane QRS and delta wave axis, a short angle between the QRS and the delta wave axis, and a precordial R/S >1 transition most likely in V₂. It is worth mentioning that no patient with an anteroseptal or a midseptal bypass tract had minimal preexcitation (Table 1).

Intravenous Adenosine

We found¹⁵ that intravenous adenosine was useful as a diagnostic tool in FV pathways. Adenosine did not elicit a greater degree of preexcitation in any of our patients, but created second degree or complete AV block with junctional beats with the

same degree of preexcitation and short HV interval as during sinus rhythm (Fig. 5).

Conclusion

The ECG of patients with FV pathways shows similarities with anteroseptal AP and midseptal bypass tracts located at the apex of the triangle of Koch, but the QRS complex are usually narrower. FV pathways with a large QRS complex cannot reliably be differentiated from an anteroseptal or a midseptal bypass tract by the surface ECG.

The definite diagnosis requires an intracardiac study observing presence or absence of changes in the QRS complex during single test atrial stimulation and atrial pacing at increasing rates. This should always be done when ablation of septally located AP is considered.

ECG DURING TACHYCARDIA

Twenty years ago Bardy et al.¹⁹ reported six ECG features showing a high efficacy in identifying antidromic tachycardia due to nodofascicular (NF) AP. Those criteria—(1) a QRS axis between 0° and -75°, (2) a QRS duration of 0.15 seconds or less, (3) an R wave in limb lead I, (4) an rS in precordial lead V₁, (5) a transition in the precordial leads from a predominantly positive QRS complex >V₄, and (6) a tachycardia cycle length between 220 and 450 ms—became the gold standard in identifying preexcited tachycardia due to anterograde conduction over a decrementally conducting AP. In the following years as our understanding about the anatomy of decremental fibers evolved, those pathways originally described as NF fibers would probably be reclassified today as AF pathways. We believe that no patient with decrementally conducting accessory AV pathways were included in Bardy's series, because those pathways usually show a larger QRS complex during tachycardia. We studied 32 patients with AF pathways and eight patients with AV fibers, and 35 patients with left bundle branch block (LBBB)-shaped supraventricular tachycardia in order to assess prospectively the sensitivity, specificity, and predictive value of the above-mentioned criteria.³⁶

Comparison of Our Data with the Study of Bardy et al.

In spite of small differences between our data and the results reported by the group from Duke

Figure 6. Twelve-lead ECG before and after an IV bolus of adenosine, showing prolongation of the PR interval without affecting the amount of ventricular preexcitation in a patient with a fasciculoventricular pathway.

University, sensitivity (87.5% vs 92%, $P = 0.9$) and negative predictive value (82.5% vs 91%, $P = 0.5$) of the six electrocardiographic criteria did not reach statistical significance. The criteria cycle length was not helpful in this study as in Bardy's series.

All electrocardiographic criteria are simple, easy to assess, with no interobserver variability and only the QRS transition in the precordial leads can be influenced by a malpositioning of the electrodes, but that criterion would have the least impact on the results.

Conclusion

The previously reported criteria showed reliable efficacy in identifying patients with an AF path-

way, but are of no value in distinguishing a decrementally conducting AV pathway. The tachycardia cycle length was not helpful in making the correct diagnosis.

We classified the different cardiac arrhythmias that may occur in the presence of Mahaim fibers as follows:

Arrhythmias Related to AV/AF Fibers

1. Antidromic tachycardia with anterograde conduction over a long decremental fiber.
2. Antidromic tachycardia with anterograde conduction over a short decremental AV fiber.
3. AV nodal reentrant tachycardia (AVNRT) with bystander conduction over a AF pathway.

Figure 7. A LBBB-like tachycardia with a QRS width of 0.12 seconds in a patient with an atriofascicular pathway.

4. Spontaneous fast automatic tachycardia arising in an AF pathway.
5. Spontaneous slow automatic rhythm arising in an AF pathway.
6. Automaticity induced by radiofrequency ablation at the site of atrial insertion of an AF pathway.
7. Atrial fibrillation with anterograde conduction over an AF pathway.
8. Nonreentrant preexcited tachycardia due to simultaneous dual Mahaim conduction.

Arrhythmias Related to Nodoventricular/Nodofascicular Fibers

1. Antidromic nodoventricular/nodofascicular (NV/NF) tachycardia with AV dissociation.
2. Orthodromic tachycardia via a "concealed" NF fiber.

FV Fibers

1. Orthodromic tachycardia with a "bystander" FV pathway.
2. Atrial fibrillation with anterograde conduction over a FV pathway.

1. *Antidromic Tachycardia with Anterograde Conduction over a Long (AF/AV) Fiber:* This is the most common type of tachycardia associated with Mahaim fibers (Fig. 6). The tachycardia is usually regular with a wide range of the cycle length, from 220 to 450 ms. The QRS complex width is 0.13 ± 0.01 seconds (range 0.11–0.15), showing a LBBB-like configuration with a smooth slope of the downstroke in V1, and a frontal plane axis between 0° and -75° . In rare cases of an anteriorly located decremental pathway the axis can be as positive as $+60^\circ$. The QRS width during tachycardia is related to the distal end of the fiber. The closer to the right bundle branch the narrower the QRS. The retrograde P wave can usually not be recognized because it falls within the final portion of the QRS complex. The major differential diagnosis is an SVT (orthodromic AV reentrant tachycardia using an AV bypass tract retrogradely or rarely an AVNRT with LBBB). One should keep in mind that some ventricular tachycardias might be included in the differential diagnosis of a wide complex tachycardia, particularly those having a QRS complex marginally enlarged, like in idiopathic VTs or ARVD. The QRS complex during a bundle branch reentrant tachycardia can show the same pattern as during sinus rhythm, resembling a LBBB pattern. The correct

Figure 8. A LBBB-like tachycardia with a QRS width of 0.16 seconds in a patient with a short AV Mahaim fiber. Note the notch in the S wave in V1.

diagnosis can be suspected by the presence of AV dissociation, as well as the clinical setting, usually occurring in a patient with a severe cardiomyopathy and LBBB during sinus rhythm. ARVD patients usually show repolarization abnormalities in the precordial leads, LBBB-like ectopies and occasionally epsilon waves in the right precordial leads.

2. Antidromic Tachycardia with Anterograde Conduction over a Short AV Decrementally Conducting AP: Short AV Mahaim fibers usually have wider QRS complexes and also longer VH intervals. Figure 7 shows a LBBB-like tachycardia of 215

beats/min, and a -30° QRS frontal plane axis in a patient with Ebstein's disease and an associated right posteroseptal AP, which is utilized as the retrograde limb of the tachycardia circuit. The QRS complex is wide (0.16 seconds), there is a notch at the downstroke of V1 indicating ventricular activation starting in the free wall of the right ventricle consistent with a short AV Mahaim fiber. Ventricular activation mapping showed early activation at the level of the tricuspid annulus. The retrograde P wave could not be seen on the 12-lead ECG, being buried inside the QRS complex. Ebstein's disease is

Figure 9. A preexcited tachycardia due to bystander anterograde conduction over an atriofascicular pathway. In the right side of the figure the wide QRS tachycardia becomes a narrow QRS tachycardia with no cycle length and no visible P wave, consistent with an AVNRT.

a common diagnosis in patients with Mahaim fibers with an incidence ranging from 10% to 50% (20% in our series of 40 patients). Mahaim fibers are associated with an additional overt or a concealed accessory AV pathway in up to 20% of cases. The AP location is usually right-sided and only exceptionally left-sided.³⁷

3. *AVNRT with Bystander Anterograde Conduction Over a Decrementally Conducting AP:* AVNRT is associated with a Mahaim fiber in up to 10% of the patients.^{10,38,39} It can occur as a narrow QRS tachycardia, usually with a faster rate than the antidromic reciprocating tachycardia or it can be the presenting arrhythmia showing ventricular preexcitation by bystander anterograde conduction over the decrementally conducting AP (Figs. 8 and 9). The QRS complex would be indistinguishable from the real antidromic reciprocating tachycardia because anterograde AV conduction goes over the Mahaim fiber. The finding of fusion beats during

tachycardia (usually at induction) is the major clue to diagnose such a mechanism. During the electrophysiologic study, block in the AP can be achieved by means of premature stimuli at the atrial (Fig. 10) or at the ventricular level. Sometimes no spontaneous or induced fusion beats can be seen and the diagnosis of bystander anterograde Mahaim conduction can only be made when an AVNRT with the same rate can be induced after ablation of the Mahaim fiber.⁴⁰ We reported⁴¹ the case of a patient with the slow-fast variety of AVNRT. During 2:1 AV conduction there was intermittent preexcitation over a likely NF Mahaim fiber (Figs. 11 and 12). This patient did not show preexcitation during sinus rhythm or during AVNRT and 1:1 AV conduction.

4. *Spontaneous Fast Automatic Tachycardia Arising on an AF Pathway:* Spontaneous automaticity arising in an AF pathway has been described occasionally.⁴² Clinical presentation can be as premature

Figure 10. The same patient as in Figure 8. A late right atrial premature beat delivered at the right lateral wall, blocks in the Mahaim fiber without resetting the next atrial (A) or His potential. The intracavitary recordings are consistent with an AVNRT.

beats in a bigeminal pattern, slow rhythms resembling accelerated idioventricular rhythm or fast nonsustained bursts of repetitive tachycardia. We have seen two patients with frequent episodes of repetitive automatic rhythms as the presenting arrhythmia, without a true antidromic tachycardia. There was no VA conduction over the AV node and Mahaim fiber (Fig. 13). The QRS complex morphology during tachycardia equals the one during atrial pacing at the same rate. Patients having spontaneous automaticity associated with AF pathways are younger (15 ± 7 years) than patients (26 ± 13 years) with AF pathways without spontaneous automaticity.⁴³

5. Spontaneous Slow Automatic Rhythm Arising in an AF Pathway: Spontaneous slow automaticity arising in AF pathways is an infrequent rhythm that was present in only 7.5% (3 out of 40) of our patients. Those rhythms are clinically silent (Fig. 14)

resembling an AIVR. In two of our three patients spontaneously occurring automatic beats triggered episodes of antidromic tachycardia (Fig. 15). Automaticity was abolished after successful catheter ablation.

6. Automaticity Induced During Radiofrequency Ablation at the Atrial Insertion of the AF Pathway: Heat-induced automaticity is a long-known phenomenon occurring when ablating the slow AV nodal pathway in patients with AVNRT. Some authors believe that heating the compact node at a distance is the most likely explanation.⁴⁴ Others believe that it is caused by heating of AV nodal tissue at the posterior extensions of the AV node. It is also known that regular atrial or ventricular myocardium does not generate such automaticity. The observation of automaticity during ablation of Mahaim fibers (MAT) was reported in the early 1990s. McClelland et al.²¹ found it in 11 out of 23 patients, Heald et

Figure 11. Twelve-lead ECG in a patient with an atriofascicular pathway, shows a slow LBBB-like tachycardia (QRS = 0.12 seconds) with AV dissociation, irregular RR intervals, and a cycle length between 400 and 480 ms. The frontal plane axis is -50° .

al.¹ found it in 12 out of 16 patients and called it "stuttering block," Braun et al.⁴⁵ found it in all of his 15 patients. We found it in 30 out of 33 patients,^{43,46} during ablation at the tricuspid annulus targeting the Mahaim "compact node." MAT usually starts immediately or a couple of seconds after current delivery. Automaticity may be short-lived, as short as four beats or very long-lasting up to 90 seconds (Fig. 16). The attitude toward the occurrence of MAT changed from previous concern about stability of the catheter⁴⁷ to a desirable event meaning a hallmark of successful ablation. We believe that in some cases with prolonged automaticity, sometimes associated with spontaneous automatic rhythms, complete termination of MAT may be required for long-term successful outcome.

7. Atrial Fibrillation with Anterograde Conduction Over an AF Pathway: We saw spontaneous atrial fibrillation as the presenting arrhythmia in patients with Mahaim fibers only once in 40 patients. It was a 50-year-old patient (Fig. 17) without retrograde conduction over the fiber. After successful ablation of the AF pathway, atrial fibrillation did not recur during a follow-up of 2.5 years. Miller et al.⁴⁰ ablated an AF pathway during atrial fibrillation. After ablation atrial fib-

rillation could not be reinduced. We reviewed 14 articles^{1,10,13,16,17,19–22,40,47–49,50} with a total of 208 patients with Mahaim fibers and only four patients had atrial fibrillation as the presenting arrhythmia (1.9%). It is a much lower incidence as compared with the 10–32% incidence of atrial fibrillation in patients with the WPW syndrome.³⁸ Degeneration of circus movement tachycardia to atrial fibrillation was the mechanism in 25% of the patients with WPW studied by Bauernfeind et al.⁵¹ This mechanism may explain atrial fibrillation in the patient of Brugada et al.⁵⁰ who had an additional right posteroseptal AP and antidromic tachycardia but could not explain atrial fibrillation in our patient who did not have other arrhythmias.⁵² The reason for the small incidence of atrial fibrillation in patients with Mahaim syndrome remains unknown but the presence of the decrementally conducting pathway itself seems to be important, as in the case reported by Miller et al.,⁴⁰ where atrial fibrillation could not be reinduced after ablation of the AF pathway.

8. Nonreentrant Preexcited Tachycardia due to Simultaneous Dual Conduction in a Mahaim fiber: Non-reentrant supraventricular tachycardia with simultaneous conduction in the fast and slow AV nodal

Figure 12. Twelve-lead ECG: Left part of the tracing shows a tachycardia with most of the QRS complexes having a left bundle branch block shaped QRS. The last three QRS complexes of the tachycardia show doubling of the ventricular rate. The QRS complex thereafter is a junctional escape beat and the last two beats are of sinus origin (see text for discussion).

pathways had been widely reported.⁵³ Those patients share some common features: they are usually refractory to antiarrhythmic drug treatment. Tachycardia can be aggravated by the use of drugs; in the majority of patients no reentrant AV nodal tachycardia can be induced. The effective refractory period of the "fast" pathway is shorter than the "slow" pathway; they usually show absence of retrograde VA conduction. Without retrograde conduction over the "slow" pathway at the time of anterograde conduction over the "fast" pathway, conduction can proceed over the "slow" pathway and with a critical atrial cycle length it can reach the ventricle a second time. We reported⁵⁴ a patient with an AF pathway showing similar characteristics: an incessant tachycardia due to 1:2 P/QRS relationship (Fig. 18), no reentrant antidromic tachycardia could be induced, tachycardia did not respond

to antiarrhythmic drugs (sotalol and amiodarone) and he did not have VA conduction through the decrementally conducting AP. No further dual conduction occurred after successful catheter ablation of the AF pathway.

9. Antidromic NV Tachycardia with VA Block or AV Dissociation: Antidromic tachycardia with AV dissociation (or 2:1 VA conduction) is the most important clue for diagnosing a NV fiber incorporated in the tachycardia circuit. Ventricular tachycardia is the main differential diagnosis of such tachycardia. It is not possible to preexcite ventricular activation, because the AP is taking off from the AV node. Some authors¹³ suggested that failure to preexcite ventricular activation by atrial premature beats would be an argument for the presence of a NV Mahaim fiber. However, Porkolab et al.⁵⁵ showed failure of decremental right atrial

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Figure 13. Intracardiac recording of the tachycardia. The atrial cycle length is 270 ms and the ventricular cycle length of the wide QRS complexes is 540 ms. There is a 2:1 AV nodal block and the ventricular beats show two morphologies: A LBBB-shaped QRS with a -20 ms HV interval and a narrow QRS complex with a 40 ms HV interval. The longer interval in ventricular activation between the RV apex and at the bundle of His during the LBBB shaped QRS as compared to that interval during the narrow QRS suggests that following anterograde conduction over the Mahaim fiber there is retrograde activation of the intraventricular conduction system. Surface ECG, intracavitary tracings, and abbreviations as in Figure 2. Recording speed is 100 mm/s.

extrastimuli to reset an antidromic tachycardia in a patient with a true AF Mahaim. This patient was successfully ablated targeting the Mahaim potential at the right lateral tricuspid annulus. In a patient with a LBBB-like tachycardia with AV dissociation, the presence of a true NV Mahaim fiber must be considered depending upon the finding of a progressive AH interval prolongation and a HV interval shortening during atrial pacing at increasing rates. Another clue is to induce a narrow QRS tachycardia with AV dissociation in the same patient, due to reversal of the tachycardia circuit, with anterograde conduction over the AV node and retrograde conduction over the NV Mahaim fiber, as reported by Gallagher et al.⁴

10. Orthodromic Tachycardia via a "Concealed" NF/NV Fiber: There are very few cases with a NF

Mahaim fiber and a clinical tachycardia where such a structure was diagnosed and ablated.⁹⁻¹²

Those cases showed no preexcitation at baseline ECG and a narrow QRS tachycardia with ventriculoatrial block or AV dissociation, and a midseptal concealed NF/NV fiber was successfully ablated. Engagement of an AP was proven by interruption of the tachycardia or advancement of the next His-bundle potential by giving a ventricular premature beat at the time of His-bundle refractoriness. In one case reported by Hluchy et al.¹² there was 1:1 midline VA conduction and intermittent 2:1 AV infra-Hisian block. Atrial tachycardia was ruled out because the tachycardia could be terminated by ventricular premature beats without VA conduction, and an AP potential preceded the earliest retrograde atrial activity, at the right midseptal area.

Figure 14. Twelve-lead ECG showing sinus rhythm (580 ms) in between two episodes of Mahaim automaticity (640 ms). Fourth and 14th QRS complexes are fusion beats.

Shimizu et al.⁴⁴ reported a patient with Mahaim physiology and narrow QRS tachycardia preceded by a normal HV interval with VA dissociation. He could preexcite His bundle activation during tachycardia with a premature ventricular beat at a time of His bundle refractoriness. Atrial pacing findings were consistent with Mahaim physiology. The findings were suggestive of a NV fiber.

11. *Orthodromic AV Tachycardia with a "Bystander" FV Pathway:* FV pathways have so far not been actively associated with cardiac arrhythmias, but they are commonly associated with additional AP.¹⁵ One has to be careful assessing the mechanism of any antidromic tachycardia before taking a decision to ablate. A "bystander" FV pathway can be misdiagnosed as a parahisian AP, while mapping during an orthodromic AV reentrant tachycardia as shown in Fig. 19. This patient presented with a wide QRS tachycardia suggestive of an antidromic tachycardia using an anteroseptal bypass tract as the anterograde limb (or a ventricular tachycardia). The baseline ECG (Fig. 19A) was consistent with a combination of two anterograde preexcita-

tion patterns (a left lateral AP and a right-sided or anteroseptal AP). After ablation of the left-sided AP the ECG (Fig. 19C) showed the same QRS pattern as during tachycardia. Electrophysiologic findings included a fixed, short HV interval and preexcitation despite variation in the AH interval following atrial premature beats.

One should suspect an additional FV pathway when the findings on the 12-lead ECG during sinus rhythm cannot be explained only by ventricular preexcitation over one accessory AV pathway.

12. *Atrial Fibrillation with Anterograde Conduction Over a FV Pathway:* Another clue to the diagnosis of a FV pathway can be seen in Fig. 20 which shows a self-terminating episode of atrial fibrillation with a minimally preexcited QRS complex. What strikes the eye is the absence of any variation in the degree of preexcitation both during changing RR intervals in atrial fibrillation and in sinus rhythm. This is highly suggestive for an associated FV pathway. The association of atrial fibrillation with a FV pathway is fortuitous with no cause-effect relationship.

Figure 15. Twelve-lead ECG showing automatic beats among minimally preexcited sinus beats. Antidromic circus movement tachycardia is triggered by an automatic beat with retrograde VA conduction through the AV node.

CONCLUSION

This large group of AP comprising the "Mahaim family" with different electrophysiologic characteristics and also diverse structural features can give rise to a number of reentrant and automatic rhythms. It is important to correctly understand their specificities in order to perform the correct therapeutic approach.

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Figure 16. Episodes of heat-induced automaticity during radiofrequency ablation of long Mahaim fibers at the tricuspid annulus. (A) short lived automatic rhythm (four beats) and (B) automatic activity lasting almost 2 minutes.

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Figure 17. Panel A: atrial fibrillation in a patient with a right lateral atriofascicular pathway. Panel B shows sinus rhythm with minimal preexcitation with an *rS* pattern in lead III. Panel C: QRS pattern is similar to QRS during atrial pacing.

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Figure 18. Left panel: A 12-lead ECG during sinus rhythm showing frequent episodes of nonsustained tachycardia based upon one sinus P wave resulting into two QRS complexes. Sinus P waves are indicated by arrows. Note that an episode of tachycardia terminates because of block in the "slow" Mahaim pathway. The QRS complex after the pause, is a fusion complex between AV conduction over the AV node and the Mahaim fiber. Right panel: The ECG after catheter ablation of the Mahaim fiber. Paperspeed: 25 mm/s.

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Figure 19. (A) Baseline 12-lead ECG. The preexcitation pattern cannot be explained by a single accessory AV pathway (see text). (B) The QRS complex during the induced preexcited tachycardia resembles the QRS complex after ablation of the left-sided AP. (C) ECG after ablation of the left-sided AP, showing a ventricular preexcitation pattern suggestive of a right-sided insertion of the accessory connection.

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Figure 20. An episode of self-terminating atrial fibrillation showing a “fixed degree” of preexcitation during atrial fibrillation and sinus rhythm. Note a delta wave configuration suggestive of an anteroseptal insertion of the accessory connection.