Role of Mahaim fibers in cardiac arrhythmias

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ROLE OF MAHAIM FIBERS IN CARDIAC ARRHYTHMIAS

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ROLE OF MAHAIM FIBERS IN CARDIAC ARRHYTHMIAS

PROEFSCHRIFT

ter verkrijging van de graad van doctor
aan de Universiteit Maastricht,
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volgens het besluit van het College van Decanen,
in het openbaar te verdedigen
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בְּרוּךְ אַתָּה ה' אֱלֹהֵינוּ מֶלֶךְ הָעוֹלָם,
שֶׁחַיֵּנוּ וְקִיּוּמֵנוּ וְהַגִּיעָנוּ לַזְּמַן הַזֶּה.

Praised are You, Lord our God, King of the universe,
for granting us life, for sustaining us, and for helping us to reach this day.

Para Cláudia, Tomaz, Ivan e Daniel.
Aos meus pais Múcio e Nícia.

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Chapter 1

Introduction

Contents

- 1.1 Historical Remarks on Variants of Preexcitation
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HISTORICAL ASPECTS OF THE VARIANTS OF PREEXCITATION

Almost twenty five years ago, Gallagher et al¹ stated that "*the role of Mahaim fibers in the genesis of cardiac arrhythmias in man has been controversial since they were first described*". The original description was made in the late 30's by Mahaim^{2, 3} (Figures 1 and 2).

Figure 1: Ivan Mahaim (1897-1965). Born in Liège, Belgium and educated in Lausanne, Switzerland. He was fellow of Prof. Wenckebach in Vienna (1926). Mahaim wrote more than 100 papers that were published in the leading journals of his time. His most influential works were his books on histologic research concerning the connections of the bundle of His, which was a resounding success in Europe in 1937 as it provided the basis for later electrophysiological discoveries. His last work, still a reference for musicians, was devoted to Beethoven's last quartuors, and published in 1964.

He found conducting tissue extending from the A-V node to the ventricular myocardium. There are only a few articles dealing with anatomical studies³⁻⁸ of Mahaim fibers. Early investigators found that Mahaim fibers were accessory connections taking off from the His bundle and fascicles (FV-fasciculoventricular) into the right ventricle or from the atrioventricular node (NV-nodoventricular fibers) to the right ventricle or to the right bundle (NF-nodofascicular fibers). Anderson et al⁹ proposed 2 varieties of NV fibers, one arising in the transitional zone and the other taking off from the deep, compact nodal portion of the AV junction. The nodoventricular concept was consistent with the findings from the Gallagher¹ series where some patients had V-A block during wide complex tachycardia, proving that in some patients the atrium was not part of the reentrant circuit.

Wellens¹⁰ was the first to report on the electrophysiologic findings using the technique of programmed electrical stimulation in a patient with an accessory pathway with decremental properties and long conduction times, assuming a relationship with the fibers described long ago by Mahaim. The term nodofascicular (NF) was applied when the retrograde His bundle potential preceded the ventricular deflection, while a nodoventricular pathway was assumed when the retrograde His bundle deflection followed the beginning of the ventricular potential. The next step

Figure 2: Les Maladies Organiques du Faisceau de His-Tawara: *This book is still used as a reference today and was devoted to the tumors of the heart, providing a detailed analysis of more than 400 cases. It was published in 1931 and anticipates the possibility of cardiac surgery, which was considered science fiction by then.*

was the understanding of the functional significance and the anatomic-electrophysiologic relationship of such pathways. An important observation was made in 1978 by Becker et al⁷ when they described an accessory A-V node associated with a bundle of specialized fibers measuring 1 cm, coursing through the right ventricle, mimicking a second A-V conduction system located on the lateral tricuspid annulus. However, that finding did not change the mainstream concept of NV fibers at that time. During the early 80's many centers started to refer patients with drug refractory tachycardias due to Mahaim fibers for surgical treatment. Although Gillette et al¹¹ reported as early as 1982 a Mahaim fiber located on the anterior portion of the tricuspid ring, according to the current concepts at that point in time, ablation of the A-V node was considered to be the logic strategy for curative treatment of patients with NV/NF fibers. Some electrophysiologists introduced a new technique delivering high-energy current through a catheter to achieve ablation of the A-V node to treat a patient with a Mahaim fiber. This resulted in complete A-V block but persistent preexcitation¹². The turning point came in 1988, when Klein, Guiraudon and colleagues¹³ decided to extensively freeze the A-V node and upper His bundle region of a 29 year old man and discovered that preexcitation did not go away. That finding indicated to them that the accessory pathway was not connected with the A-V node. In the next patient the A-V node was not damaged but the Mahaim fiber successfully blocked by ice mapping at the right lateral aspect of the tricuspid annulus. Klein's manuscript was published in 1988. Shortly thereafter Tchou et al¹⁴ published a paper entitled "*Atriofascicular connection or a nodoventricular fiber? Electrophysiologic elucidation of the pathway and associated reentrant circuit.*" In this elegant study the authors describe a simple maneuver to

prove that such pathways are in fact inserting in the atrium and not in the A-V node by delivering a late lateral atrial premature beat at the time that the A-V node was refractory. They showed that it was possible to advance ventricular depolarization during preexcited tachycardia. In the following years, catheter ablation techniques shed more light on the subject. Discrete high frequency potentials resembling the His bundle potential, considered to be the electrical activation of Mahaim tissue, were used as an effective target for ablation^{15, 16}. Observations during pharmacological interventions¹⁷, histologic data^{7, 8, 18-19}, electrophysiologic maneuvers and findings during radiofrequency catheter ablation like heat induced automaticity^{22, 23}, and spontaneous automaticity²⁴ were presented as further evidence that the Mahaim fiber is composed of A-V nodal like tissue. Some authors²⁰ believed that an accessory A-V node without a direct connection to the ventricle, can be the substrate of atrial tachycardias mapped to the lateral tricuspid ring. They showed potentials preceding the P wave and decremental conduction between those potentials ("M") and the P wave and also the occurrence of automaticity during radiofrequency catheter ablation. The NV/NF fibers are now considered a rare item. Anecdotal reports suggest²⁵ their presence in some patients with narrow and regular QRS tachycardia with A-V dissociation.

Recent reports using non-contact technologies for intracardiac mapping of Mahaim fibers like the EnSite²⁶ and the LocaLisa system²⁷ have validated old data derived from open heart epicardial mapping and intracardiac catheter mapping^{11, 13}, suggesting that Mahaim fibers are long structures connecting the right atrium to the anterior apical region of the right ventricle, close to or inserting into the distal part of the right bundle branch.

The variety which is known as fasciculoventricular pathways²⁸ are not related anatomically to atriofascicular pathways. They do not have long conduction times nor decremental properties. As long as they are infra A-V nodal structures connected to the His bundle or its fascicles the decremental conduction is coming from the AV node which led previous investigators to believe otherwise. They play no role in clinical tachycardias. However, due to the fact that its pre-excitation pattern on the 12-lead electrocardiogram may resemble that of an anteroseptal accessory pathway, and they are often associated with rapidly conducting bypass tracts, a misdiagnosis of a bypass tract should be avoided to prevent unnecessary damage to the A-V node-His bundle conduction system by catheter ablation.

CRITICAL ANALYSIS OF THE CLASSIFICATION OF MAHAIM FIBERS

The preexcitation syndromes were originally classified on the basis of their anatomic location and course and named according to the original investigators resulting in Mahaim, James, and Kent fibers. Later, such a description was considered inadequate because it did not fit the new electrophysiologic or anatomic knowledge. The European Study Group for Preexcitation⁹ introduced in 1975 a new classification based upon the proposed anatomic connections of the accessory pathways. However, the eponym Mahaim fiber survived the European Study Group proposed changes. Why? It seems that the major issue here is the common electrophysiologic finding of long and decremental conduction properties (A-V node like behavior) of such pathways, the so called "Mahaim" physiology.

An updated anatomical classification of the Mahaim fibers is shown in figure 3:

Proximal insertion in the AV node – His – Bundle branch system

1. Nodofascicular bypass tract
2. Nodoventricular bypass tract
3. Fasciculoventricular pathway

Proximal insertion in the atrium

1. Atriofascicular pathway
2. Short atrioventricular pathway with prolonged and decremental conduction
3. Long atrioventricular pathway with prolonged and decremental conduction
4. Left-sided atrioventricular or atriofascicular pathway with prolonged and decremental conduction

Figure 3: The different anatomic courses of bypass tracts classified as Mahaim fibers

DIAGNOSIS

ELECTROCARDIOGRAPHIC FEATURES: The baseline electrocardiogram of patients with Mahaim fibers (figure 4) is characterized by minimal (QRS width less than 0.12 sec) or no ventricular pre-excitation.

Figure 4: Panel A: Atriofascicular pathway. The ECG during sinus rhythm shows minimal preexcitation with a 0.12 s PR interval. During atrial pacing (right panel) a LBBB pattern develops with a frontal QRS axis of -45° , QRS is 120 msec wide. Panel B: An atrioventricular pathway in Ebstein's disease. The ECG during sinus rhythm is without preexcitation and shows RBB with a normal PR interval. During atrial pacing (right panel) preexcitation becomes manifest with LBBB ($\Delta QRS = -30^\circ$), QRS width is 160 msec, with a slurred initial r wave in V₁.

Sometimes the only clue is the absence of a septal **q** wave in leads I, V₅ or V₆²⁹. An **rS** pattern (less frequently **rsR'**) in lead III was found in 28 of 40 patients (70%) with a Mahaim fiber³⁰ (figure 5).

Figure 5: A) The ECG of a young individual with palpitations showing an **rS** pattern in lead III, associated with a **q** wave in lead I (incidence of 6 %); B) ECG from a patient with Mahaim fiber with an **rS** pattern in lead III and absence of **q** wave in lead I; C) the same patient after ablation. A **qR** pattern (the most common pattern in normal individuals in lead III) is present.

Because a normal person may also have such an **rS** pattern in lead III, we did a survey in 200 young individuals (mean age of 23 years) to address the issue of the specificity of this QRS pattern in lead III. It was found in 6 % of the 200. However, no individual had an **rS** pattern in lead III with absence of a **q** wave in lead I. We conclude that the association between the absence of a **q** wave in lead I and an **rS** pattern in lead III seems very specific for Mahaim. Patients with a Mahaim fiber having an abnormal pattern in lead III show left axis deviation in the frontal plane during antidromic tachycardias. After successful catheter ablation a different QRS pattern usually emerges (most commonly a **qR** or **QR** pattern in lead III). It is worth mentioning that the **rS** pattern can be concealed by an associated WPW and emerge after the successful ablation of the rapidly conducting accessory A-V pathway, as did happen in 4 out of 5 patients in our cohort. Therefore in patients with paroxysmal tachycardias the presence of an **rS** pattern in lead III during sinus rhythm should raise

the suspicion of a Mahaim fiber. Occasionally, patients, particularly those having a short atrioventricular accessory pathway with a long conduction time show a typical LBBB pattern with a normal PR interval. Some patients are unable to have anterograde conduction over the Mahaim fiber at rest and even during atrial pacing³¹. In spite of that, these so called "latent" Mahaim fibers may become engaged in antidromic tachycardia. The differential diagnosis from ventricular tachycardia can then only be achieved by performing an electrophysiologic study.

Variability in minimal preexcitation may occur either on the same ECG or on ECG's taken on different days. This variable ECG presentation does not correlate with a long refractory period, in contrast to intermittent preexcitation occurring in patients with rapidly conducting accessory pathways.

The ECG during a tachycardia with anterograde conduction over an atriofascicular fiber shows a typical LBBB pattern with a frontal axis usually ranging from -0° to -60° . While the axis is of no help in differentiating it from A-V conduction over a right sided A-V pathway, the QRS complex is rather narrow (0,12 - 0,14 seconds) being larger when anterograde A-V conduction proceeds over a short atrioventricular Mahaim. In the latter situation there is often a broader r wave in V_1 and V_2

(>30 msec)³¹. Bardy's³² criteria for an atriofascicular Mahaim remain useful today: R wave in lead I, rS in V_1 , QRS width $\leq 0,15$ seconds, precordial R/S transition after V_4 , a QRS frontal plane axis between 0 and -75° and a tachycardia cycle length between 220 and 450 msec. We have validated the abovementioned criteria³³ (except cycle length) only for atriofascicular pathways, but not for short A-V Mahaim's. The electrocardiogram of a fasciculoventricular pathway is characterized by a normal QRS frontal plane axis as in an anteroseptal accessory AV pathway (0 to $+75^{\circ}$)³⁴ with subtle preexcitation and a normal PR interval. A short PR interval may be present in case of enhanced A-V nodal conduction (figure 6).

Atrial pacing does not change the degree of preexcitation. Junctional beats are preexcited and intravenous adenosine results in blocked P waves. The $R/S > 1$ precordial transition usually occurs in lead V_2 .

ELECTROPHYSIOLOGIC CHARACTERISTICS: The major finding is the presence of a pathway with slow conduction and decremental properties during right atrial pacing at increasing rates or after atrial premature beats. The AH interval lengthens, the HV interval shortens and the QRS widens until a steady value is reached. A further increase in atrial stimulation rate does not increase preexcitation, but can prolong A-V conduction until A-V block occurs.

Decremental conduction is usually defined as rate dependent prolongation of the conduction time over an A-V accessory pathway of more than 30 msec, as measured using electrograms close to the AP insertion. During atrial pacing at increasing rates achievement of maximal preexcitation is usually associated with retrograde conduction over the right bundle, His bundle and A-V node and then followed by antidromic tachycardias when pacing is stopped. An important step is to assess the role of the accessory pathway in the tachycardia circuit: active or bystander. This can be done by giving single late laterally located right atrial

Figure 6: Six cases of fasciculoventricular pathways during sinus rhythm. A: a left-sided fasciculoventricular pathway. B to F: right-sided fasciculoventricular pathways. P-delta intervals range from 0.08 to 0.10 sec. The QRS width ranges from 0.09 to 0.12 sec. The frontal plane axis ranges from $+15$ to $+80^\circ$. $R/S > 1$ QRS transition occurs in precordial lead V_3 or earlier.

extrastimuli during a preexcited tachycardia, not affecting the His bundle and the atrial electrogram at the coronary sinus ostium¹⁴ (figure 7). Advancement of the QRS complex and subsequent atrial activation establishes the diagnosis of an extranodal accessory A-V pathway (having a proximal atrial insertion) as well as incorporation in the tachycardia circuit.

If advancement of QRS activation occurs without changing the timing of atrial activation, the presence of an extranodal AP is certain but its participation in the circuit still need to be proved.

McClelland et al¹⁶ could successfully advance QRS activation with late right atrial extrastimuli in 22 of 23 patients with an atriofascicular tachycardia. Another less elegant maneuver proving AP participation in SVT is by having catheter-induced RBBB. Assuming an antidromic tachycardia incorporating an atriofascicular (or atrioventricular) pathway inserting into the right ventricle retrograde conduction during the tachycardias occurs via the right bundle-His bundle and A-V node axis. A RBBB will lengthen the circuit thereby increasing the tachycardia cycle length because of ventriculo-atrial conduction over the left bundle branch.

Preexcited A-V nodal reentry as well as antidromic tachycardia with retrograde conduction through another AP will not be affected by RBBB. The proximal atrial insertion can also be localized with the recording of an accessory pathway potential, usually recorded in the lateral aspect of tricuspid annulus, away from the A-V node (figure 8). In the study by Grogan et al³⁵ clues as to the presence of a nodoventricular fiber were the inability of a premature atrial stimulus to advance the ventricle and the presence of dual A-V nodal pathways.

Figure 7: Antidromic Mahaim tachycardia. A late atrial extrastimulus (S) delivered from the lateral high right atrium without disturbing AA timing at the His bundle recording advances the QRS complex by 20 msec and the His deflection by 30 msec, proving that the Mahaim pathway is extranodal and participates in the tachycardia circuit.

Scheinman et al³⁶ described a patient with a lateral Mahaim in whom an atrial premature beat could not preexcite the ventricle. However, the finding of an "M" potential and successful ablation at this spot was consistent with an atrioventricular Mahaim fiber and not a nodoventricular fiber. Ventricular stimulation usually discloses ventriculo-atrial conduction over the A-V node. The vast majority of atriofascicular and atrioventricular Mahaim pathways are unable to conduct retrogradely^{37, 38}.

Adenosine injection during sinus rhythm usually causes block of impulse conduction over both the A-V node and the Mahaim fiber (figure 9). Occasionally it can show a more marked effect over the A-V node and increase preexcitation³⁹. Verapamil¹⁷ has a more marked effect on A-V nodal conduction, and is more helpful in exposing preexcitation. During antidromic tachycardia adenosine causes termination of antidromic tachycardia by prolongation of conduction over the Mahaim pathway and block.

Figure 8: One example of an "M" potential: There is a high frequency and low amplitude accessory pathway potential recorded from the ablation catheter, located at the right lateral TA (tricuspid annulus). Successful catheter ablation was performed at the site.

Mahaim fibers often occur in the setting of Ebstein's disease (20%) and associated rapidly conducting accessory pathways (usually right sided) are seen in up to 30% of these patients. Fasciculoventricular pathways also occur very often in association with rapidly conducting bypass tracts. In cases reported since 1981^{1, 26, 40-41} there was a 40% incidence of an associated AP.

TREATMENT

ANTIARRHYTHMIC DRUG TREATMENT: In general, verapamil and propranolol have been ineffective for termination of tachycardia episodes¹⁷. Intravenous adenosine is the first choice intravenous drug for acute treatment during Mahaim tachycardia. Patients with Mahaim fibers are frequently unresponsive to multiple antiarrhythmic drugs. Flecainide, propafenone, propranolol, sotalol, and digoxin have been used as single drugs or in combination with little success. Some authors reported that type IA agents (quinidine⁴²) were effective in preventing or slowing

tachycardia. Currently, antiarrhythmic drug therapy plays a secondary role because of the highly successful results with radiofrequency catheter ablation.

MAPPING AND CATHETER ABLATION: Some features are unique to Mahaim fibers: Mapping of the atrial insertion during ventricular stimulation is usually not possible because these decrementally conducting pathways usually do not conduct retrogradely. Excentric ventriculo-atrial conduction if present, may indicate associated rapidly conducting bypass tracts. A short atrioventricular Mahaim can be located by mapping the site of earliest ventricular activation at the annulus, as with other anterogradely conducting accessory A-V pathways. In atriofascicular pathways or long atrioventricular Mahaim's the distal (non-annular) insertion cannot be mapped in this way. To worsen matters these decremental pathways are unusually sensitive to mechanical trauma. Accidentally touching the annulus with the ablation catheter can result in transient abolition of conduction through the pathway lasting minutes to hours⁴³⁻⁴⁴. The following strategies have been used to overcome these problems:

1. Searching for the "M" (Mahaim) potential (figure 8) along the tricuspid annulus is the most commonly used technique. The ablation catheter should be moved

Figure 9: Intravenous adenosine during sinus rhythm in a patient with intermittent conduction over a short A-V Mahaim fiber: 12 mg increased the degree of ventricular preexcitation (*) due to block in the A-V node and the following sinus P wave was blocked both in the Mahaim fiber and at the A-V node. Injection of 18 mg caused transient block in both structures.

cautiously along the annulus avoiding bumps on the Mahaim tissue. The potential may be as large as the His bundle potential or small and narrow with low amplitude. Catheter ablation at a site with an "M" potential is likely to be successful. We²³⁻²⁴ and others^{22, 37} have recorded automatic rhythms (Mahaim automatic tachycardias-MAT) during radiofrequency current delivery in most patients with Mahaim fibers (30 out of 33 patients: 91%). It is probably due to heat-related automaticity of nodal-like tissue as is also seen during slow A-V nodal pathway ablation. MAT is in most cases short-lived but may sometimes last up to 2 minutes. At least in some patients it is necessary to completely terminate those rhythms to prevent recurrences.

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Chapter 2

Differential diagnosis of left bundle-branch block-shaped tachycardias

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INTRODUCTION

The correct diagnosis of the site of origin of a regular tachycardia with a wide QRS complex (≥ 120 msec) is of importance for management and prognosis of the patients. Over the last 40 years a number of approaches have been suggested to help the physician in this challenging decision making¹⁻¹⁴. Intravenous medications erroneously given because of a misdiagnosis of SVT, particularly verapamil or diltiazem, can be deleterious because they may precipitate hemodynamic collapse in the patient with VT. Stable vital signs during tachycardia are not helpful for distinguishing SVT from VT. If the diagnosis of SVT cannot be proven or cannot be made easily, then the patient should be treated as if VT were present¹⁵. The focus should be on the ECG signs which may not only help to distinguish between a VT from other tachycardias with a broad QRS complex but also suggest its etiology and site of origin in the ventricle.

Classification of LBBB-shaped tachycardias:

LBBB-shaped tachycardias can be divided into 3 groups (figure 1):

1. Supraventricular tachycardias with preexistent or functional LBBB
 - 1.A. Pre-existent LBBB
 - 1.B. Tachycardia-dependent phase 3 block
 - 1.C. Retrograde invasion into the LBB
 - 1.D. LBBB induced by drugs (pseudo VT)
2. Supraventricular tachycardias with AV conduction over a right sided accessory pathway
 - 2.A. Rapidly conducting accessory pathways
 - 2.B. Slowly conducting accessory (Mahaim) pathways
3. Monomorphic Ventricular tachycardia
 - 3.A. Ectopic ventricular origin
 - 3.B. Bundle branch reentry

THE ECG DIAGNOSIS

1. AV dissociation

Although dissociation between atrial and ventricular activity during tachycardia is a hallmark of VT (lead II, figure 2), some form of V-A conduction can be present during VT, particularly during a slow VT (figure 3)⁷. P waves can be difficult to recognize during a broad QRS tachycardia and it can be useful to look for non-electrocardiographic signs such as variations in jugular pulsations, the loudness of the

first heart sound and changes in systolic blood pressure¹⁶. In patients with slow VT rates occasional conduction from atrium to ventricle over the AV node-bundle branch system may happen resulting in “capture”

Figure 1: Different types of SVT with LBBB (diagram A), SVT with atrioventricular conduction over an accessory pathway (diagram B), and VT (diagram C) resulting in a broad QRS complex tachycardia.

Abbreviations: Acc= accessory; AV= atrioventricular; BBB= bundle branch block; CMT= circus movement tachycardia; SVT= supraventricular tachycardia; VA= ventriculo-atrial; VT= ventricular tachycardia.

or “fusion” beats. Sudden narrowing of a QRS complex during VT may also be the result of a premature ventricular depolarization arising in the ventricle in which the tachycardia originates, or it may occur when retrograde conduction during VT produces a ventricular echo beat leading to fusion with the VT QRS complex.

Very rarely AV dissociation is present in tachycardias other than VT. It may occur in A-V junctional tachycardia (JET) after cardiac surgery or during digitalis intoxication. A-V dissociation has been reported during narrow QRS complex tachycardia due to nodofascicular accessory pathway either manifest or concealed^{17, 18}.

2. Width of the QRS complex

As pointed out by Wellens¹⁹ and shown in figure 4, the site of origin of the VT plays a role in the width of the QRS complex. When the arrhythmia arises far from the interventricular septum, the sequential activation of the ventricles results in a very wide QRS. The QRS complex will be narrower when the VT has its origin in or close to the septum. Other factors like the size of the scar tissue (after myocardial infarction

or myocarditis), ventricular hypertrophy, and muscular disarray (as in hypertrophic cardiomyopathy) also play a role in the QRS width during VT.

It is of interest that a QRS width of more than 0,16 sec during LBBB shaped VT argues for a VT⁷. But a QRS width below such value may occur in septal VTs. Of course, QRS width is not helpful in differentiating VT from a tachycardia with A-V

Figure 2: LBBB shaped VT in a patient with ARVD with AV dissociation (arrow points to P waves) (panel A). Black arrowhead in V1 during sinus rhythm (panel B) points to epsilon wave, which can be better seen in panel C. Panel D shows a late potential in the corresponding intracavitary recording.

conduction over a rapidly conducting accessory pathway because such a pathway inserts into the ventricle leading to eccentric ventricular activation and a wide QRS complex (figure 5). QRS width it is also not helpful to distinguish between a SVT with LBBB from an antidromic tachycardia with anterograde conduction over a Mahaim fiber (figure 6). The Mahaim pathway can be a short AV structure inserting close to the annulus or a long fiber inserting at or close to the distal RBB. Tachycardias with anterograde conduction over a short Mahaim usually show a QRS width larger than 0.16 seconds. However, the majority of atriofascicular Mahaim antidromic tachycardias show a QRS complex in between 0,12 and 0,14 seconds. Other distinctive findings²⁰ include an **R** wave in lead I, **rS** in V₁, an **R/S** > 1 precordial transition after V₄, and a QRS axis to the left of -30. An SVT with LBBB

can have a QRS width of more than 0,16 under three circumstances: (1) in the presence of pre-existent LBBB in the elderly with fibrosis in the bundle branch system and ventricular myocardium (figure 7); (2) when during SVT AV conduction occurs over an accessory pathway; (3) when class IC drugs (specially flecainide) are present during SVT (figure 8).

Figure 3: Panel A: One to one ventriculo-atrial conduction during an idiopathic VT from the inflow tract of the right ventricle. The P waves are negative in the inferior leads and follow each QRS complex. Panel B: Same patient during sinus rhythm.

Figure 4: VT origin and QRS width. Upper panel: an origin close to the interventricular septum results in more simultaneous right and left ventricular activation and therefore a more narrow QRS complex. In contrast (lower panel) a VT origin in the right ventricular free wall results in sequential ventricular activation and a wider QRS complex.

Figure 5: An antidromic circus movement tachycardia with AV conduction over a right sided accessory pathway. The insertion of the accessory pathway in the free wall of the right ventricle results in sequential (first right then left) ventricular activation and a wide QRS complex.

3. QRS axis in the frontal plane

The QRS axis is not only important for the differentiation of the broad QRS tachycardia but also to identify its site of origin and etiology. A VT origin in the apico-septal region of the LV has a superior axis (to the left of -30°). LBBB with an inferior axis occurs with VT from the right ventricular outflow tract or at the pulmonary artery, just above the pulmonic valve.

4. Configurational characteristics of the QRS complex

Leads V1 and V6

In LBBB shaped VT, lead V₁ (and also V₂) (figure 9) usually shows: an initially positive QRS with positivity measuring more than 0,03 seconds; slurring or notching of the downstroke of the S wave; and an interval between the beginning of the QRS axis and the nadir of the S wave of 0,07 seconds or more¹⁰. When lead V₆ shows a **qR** pattern during LBBB shaped tachycardia, VT is very likely. In SVT with LBBB, lead V₁ shows no or minimal initial positivity, a very rapid downstroke of the S wave, and a short interval between the beginning of the QRS and the nadir of the S wave (figure 10).

Figure 6: Antidromic tachycardia over a Mahaim Fiber. Upper panel: A long atriofascicular pathway inserting in the right bundle is associated with a rather narrow QRS complex (0.12 seconds). Lower panel: antidromic tachycardia with anterograde conduction over a short atrioventricular pathway with decremental properties inserting close to the annulus is associated with a broader QRS tachycardia (0.16 seconds).

Figure 7: LBBB SVT (left panel) in a patient with pre-existent LBBB (right panel)

Figure 8: Sinus tachycardia presenting as a broad LBBB shaped QRS tachycardia in a patient using flecainide. Sinus P wave merged with the previous T wave because of a prolonged PR interval.

Figure 9: Upper panel: V1 and V2 from a patient with VT showing an initial r wave width of 60 msec, a notch at the downstroke in V1, and an interval between the beginning of the QRS to the nadir of the S wave of 100 msec. Lower panel: Findings in lead V1 and V2 during LBBB shaped tachycardia pointing to a ventricular origin.

Figure 10: SVT with LBBB. In panel A LBBB changes during tachycardia into a narrow QRS following a ventricular premature beat without a change in tachycardia rate. It points to retrograde invasion into the LBB as cause of LBBB on the left side of the tracing. As described in the text, lead V1 during LBBB clearly shows signs pointing to a supraventricular origin of the tachycardia.

Figure 11: Negative concordant precordial pattern. A VT arising in the apical area of the left ventricle, resulting in negative concordance of all precordial leads.

LBBB shaped VTs associated with coronary artery disease are usually adjacent to the inter-ventricular septum and have a higher predictive accuracy for their site of origin than RBBB shaped VTs. The ECG from a LBBB VT originating in a scar at the

apico-septal region will show a Q wave in leads I and V₆²¹.

Interval onset QRS to nadir of S wave in precordial leads

Brugada et al¹² suggested that an RS interval > 100 ms in one or more precordial leads is highly suggestive for VT. One should be careful, however, because such a number may occur in SVT with A-V conduction over an accessory pathway, SVT during administration of drugs that slow intraventricular conduction and in SVT with pre-existent BBB, especially LBBB.

Concordant pattern

When all precordial leads show either negative or positive QRS complexes this is called negative or positive precordial concordance. Negative concordance is diagnostic for a VT arising in the apical area of the heart (figure 11). However, an abnormal anatomic position of the heart in a patient with SVT with left bundle branch block can lead to negative concordance in the precordial leads (figure 12) and a false diagnosis of ventricular tachycardia. One example is *pectus excavatum* where the right ventricle lies completely under the antero-lateral precordial area²². Positive concordance (all QRS complexes in the precordial leads being positive) can occur in a VT arising in the posterior part of the left ventricle or in a tachycardia with A-V conduction over a left posterior accessory pathway.

Figure 12: Orthodromic AVRT with LBBB showing negative precordial concordancy due to abnormal positioning of the heart.

Tachycardia QRS identical to the sinus QRS

When the broad QRS is identical during tachycardia and sinus rhythm, one has to differentiate SVT with pre-existent LBBB (figure 7) from bundle branch re-entrant tachycardia²³. In diseased hearts, especially when the bundle branches and the interventricular septum are involved, a tachycardia may occur based upon a circuit with anterograde conduction down one bundle branch and after septal activation retrograde conduction over another branch of the bundle branch system (figure 13). This type of re-entry may occur in patients with anteroseptal myocardial infarction, idiopathic dilated cardiomyopathy, myotonic dystrophy, after aortic valve surgery, and after severe frontal chest trauma.

Tachycardia QRS more narrow than sinus QRS

When during tachycardia the QRS is narrower than the one during sinus rhythm a VT should be diagnosed. This can be explained by the site of origin of the VT close to the inter-ventricular septum, resulting in more simultaneous activation of the ventricles in contrast to the sequential activation of first the right and then the left ventricle in the presence of LBBB during sinus rhythm.

Presence of QR complexes

Coumel et al²⁴ called attention to the significance of a QR (but not a QS) complex during a broad QRS tachycardia, showing that their presence indicates a scar in the myocardium usually caused by myocardial infarction. Figure 14 gives an example of QR complexes during a LBBB shaped VT in a patient with an old inferior myocardial infarction. QR or qR complexes during VT are present in approximately 40% of VTs after myocardial infarction²⁵.

Figure 13: Bundle branch re-entrant tachycardia. The QRS complexes are identical during tachycardia and sinus rhythm. The PR interval is prolonged during sinus rhythm.

Figure 14: QRS complexes during VT indicating a myocardial scar (*qR* in leads III and aVF). As shown by the accompanying tracing, during sinus rhythm an inferior wall myocardial infarction is present.

5. Etiology of VT

Most VTs have a previous myocardial infarction as their etiology, and, as pointed out, a QR complex during VT can be very helpful to make that diagnosis.

However, characteristic ECG patterns can also be found in idiopathic VT²⁶ and VT in patients with arrhythmogenic right ventricular dysplasia (ARVD)²⁷. Figure 15 shows the QRS pattern of an idiopathic VT arising close to the outflow tract of the right ventricle.

In some patients tachycardia does not arise on the endocardial surface of the right ventricular outflow tract but epicardially around the root of the pulmonary valve²⁸, in the root of the aorta going to the posterior part of the outflow tract of the right ventricle. An early precordial transition ($R/S > 1$) and/or a greater r wave in right precordial leads is consistent with an epicardial site of origin²⁹.

Figure 15: Two types of idiopathic VT arising in or close to the outflow tract of the right ventricle.

In ARVD there are three predilection sites in the right ventricle: the inflow and outflow tract, and the apex (figure 16). While the first two sites have a QRS configuration during tachycardia which is difficult to differentiate from right ventricular idiopathic VT, left axis deviation in a young person with an LBBB shaped VT should immediately lead to the suspicion of ARVD (figure 2 panel A). In fact, there is an important rule in LBBB shaped VT with left axis deviation that cardiac disease should be suspected and that idiopathic right ventricular VT is unlikely.

6. Value of the ECG during sinus rhythm

The ECG during sinus rhythm may show changes such as pre-existent LBBB, ventricular pre-excitation, or an old myocardial infarction, which are very helpful in correctly interpreting the ECG during a LBBB shaped QRS tachycardia. We have recently described³⁰ a novel ECG signal suggestive of a Mahaim fiber in young patients with palpitations, which is an **rS** pattern in lead III. This pattern can be seen in 2% of normal young persons but when this **rS** pattern is associated with the absence of a **q** wave in lead I it is very specific for Mahaim fibers. The presence of A-V conduction disturbances during sinus rhythm make it very unlikely that a broad QRS tachycardia in that patient has a supraventricular origin and, as discussed, a QRS width during tachycardia narrower than during sinus rhythm points to a VT.

Figure 16: Three VTs from a patient with right ventricular dysplasia. VT A and B are from the inflow tract while VT on panel C originates from the apex.

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Chapter 3

The electrocardiogram during sinus rhythm and tachycardia in patients with Mahaim fibers. The importance of an “rS” pattern in lead III

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ABSTRACT

Background: Mahaim fibers are slowly conducting accessory pathways reaching into the right ventricle. They often play a role in tachycardias. The purpose of the study was to identify the electrocardiographic characteristics of the Mahaim fiber.

Methods and Results: We retrospectively analyzed 40 patients with Mahaim fibers. Five patients had associated WPW, and were excluded from the study. Two patients had a short atrioventricular decremental accessory pathways and were also excluded. The remaining 33 patients had a tachycardia with anterograde conduction over a Mahaim fiber. Twenty were female. Mean age was 24 ± 10 years. The most common pattern of minimal preexcitation during sinus rhythm was an **rS** pattern in lead III. This was found in 20 patients. There was a match between the presence of **rS** in lead III during sinus rhythm and left axis deviation during tachycardia with anterograde conduction over the Mahaim fiber. After ablation a different QRS pattern emerged in lead III, indicating absence of conduction over the Mahaim fiber. To obtain information about the prevalence of an **rS** pattern in lead III in age matched controls with palpitations and without structural heart disease, the 12-lead ECG of 200 young individuals were examined. An **rS** pattern in lead III was found in 6%.

Conclusion: A narrow QRS with an **rS** pattern in lead III during sinus rhythm in a patient with a history of palpitations should alert the physician to the possibility of a Mahaim fiber. During tachycardia these patients typically show a left bundle branch block like QRS complex with left axis deviation.

INTRODUCTION

Accessory pathways with long and decremental anterograde conduction have been the subject of extensive debate about their anatomic structure¹⁻⁴, location^{5,7}, related arrhythmias⁸⁻⁹, electrophysiologic properties¹⁰⁻¹², ablative techniques¹³⁻¹⁴ and automaticity¹⁵. Less attention has been given to the 12-lead ECG, especially to the ECG during sinus rhythm. The latter is considered to be normal in the majority of patients with atriofascicular pathways and patients with long atrioventricular decrementally conducting accessory pathways. Minimal 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 report on ECG findings in a large series of patients with Mahaim during sinus rhythm and tachycardia, with emphasis on a particular ECG pattern in lead III during sinus rhythm.

STUDY POPULATION AND METHODS

We retrospectively analysed 12-lead ECG's from 40 patients having anterograde conduction over accessory pathways with long conduction times and decremental properties both during sinus rhythm, and during tachycardia. Five patients also had anterogradely rapidly conducting accessory pathways and were excluded from the study. Two patients with a short atrioventricular decremental pathway were also excluded from this series.

Definition of terms: we use the eponym Mahaim fibers in this study for an anterogradely conducting accessory A-V pathway with long and decremental properties having a long anatomic course: either atriofascicular pathways (30 patients) or atrioventricular pathways (3 patients).

Every one of these 33 patients showed electrophysiologic criteria for a decrementally conducting bypass tract during atrial pacing, such as a progressive AH and A-V interval prolongation coupled with a decreasing HV interval leading to a greater degree of pre-excitation with a left bundle branch block-like morphology^{5,8,10}.

There were 20 females and 13 males, with a mean age of 24 ± 10 (range 8 to 52) years. All patients were referred for electrophysiologic assessment of a preexcited tachycardia. Preexcited AVNRT using a Mahaim fiber as a bystander was present in 1 patient. One patient had atrial fibrillation with preexcited QRS complexes and 2 patients were referred because of repetitive episodes of unsustained tachycardia caused by automaticity arising in the Mahaim fiber⁹ (table I). Ebstein's disease was diagnosed in 4 patients. The atrial insertion of the Mahaim fiber was located by the recording of a discrete accessory pathway potential in 28 patients and in 5 patients by assessing the shortest A-V interval during atrial pacing at different sites along the tricuspid annulus. All patients underwent successful surgical ($n = 2$) or radiofrequency catheter ablation ($n = 31$). RF ablation was guided by discrete potentials at the tricuspid annulus (28 patients) and by right ventricular pacemapping in 5 patients.

We also analyzed the 12-lead ECG during sinus rhythm in 200 individuals with palpitations and without structural heart disease matched for age and gender as a control group.

Recordings of the 12-lead surface ECG and intracardiac electrograms were made using the EP Tracer or MS System (CardioTek BV, Maastricht, The Netherlands).

Definitions of the QRS patterns

ECGs were examined by two different observers with a magnified lens and a third observer decided when there was a mismatch classification. The following QRS patterns were found: **r/ rS/ RS/ Rs/ rsR'/ rsr'/ R/ qR/ QR/ QS/ qRs/ qRS**. Very low voltage QRS complex (< 0.3 mv) were depicted as small letters (*r*, *rs* or *rsr'*). QRS complex with a higher voltage were depicted according to the ratio between the positive (R, *r*) and the negative waves (*q*, *Q*, *S* and *s*). For example a **RS** complex is defined by the presence of a QRS complex showing an initially positive deflection followed by a negative deflection of an even magnitude. Likewise, an **Rs** pattern means a QRS complex (> 0.3 mv) with an initial positive deflection followed by a smaller negative one. A septal **q** wave was defined as a **q** wave in surface leads I, aVL, and V6, with an amplitude $< 25\%$ of the R wave and a width < 0.04 second.

Statistical Analysis

Values are given as mean $\pm$ standard deviation. The significance of differences ($p < 0.05$) between groups of clinical, electrocardiographic or electrophysiologic parameters was assessed by Student's *t* test or Fisher's exact test.

RESULTS

Pre-ablation 12-lead ECG findings

Minimal preexcitation, defined as subtle abnormalities suggesting the presence of preexcitation, with a QRS complex width within the normal range (< 0.12 sec) but with a short HV interval (< 35 msec), was present during sinus rhythm in 24 patients (72%) (figure 1). The PR interval was not significantly different when comparing patients with (125 ± 21 msec) and without (132 ± 9 msec) minimal pre-excitation ($p = \text{ns}$). We found 2 patterns of the QRS complex (figure 2) during sinus rhythm: the most common one being an **rS** configuration in lead III. This was found in 20 patients. The other pattern in lead III, an **rsR'**, was found in 2 patients. In the presence of an **rS** pattern in lead III, no **q** wave was found in lead I in 15 patients, (and in 8 patients in lead V6). Minimal preexcitation with absence of a **q** wave in lead I (without **rS** in lead III), was seen in only 2 patients (patients 8 and 16, Table II). In three patients minimal preexcitation was not always manifest, as documented by 12-lead ECG's taken on different days. Variability of minimal preexcitation in the same ECG was seen in 2 patients (figure 3).

Figure 1: Two patients with Mahaim fibers displaying the rS pattern in lead III.
Abbreviations: pre/ post = pre or post radiofrequency catheter ablation.

Figure 2: 5-lead ECG during sinus rhythm with the pre-ablation ECG showing the rS pattern in 4 cases, and rsR' in 1 patient. The post-ablation ECG shows a clear change in QRS configuration.

TABLE I
Clinical data

n	Sex	Age	Site-TA	Arrhythmia	Ebstein	CBT	Therapy S/RFp/RF-dis
1	F	31	L	Antidromic AVRT/Mahaim			S
2	F	32	L	Antidromic AVRT/Mahaim AVNRT			RFp
3	F	19	PL	Antidromic AVRT/Mahaim	Yes		RFp
4	F	21	PL	Antidromic AVRT/Mahaim Orthodromic AVRT	Yes	RPS	RFp
5	M	52	PL	Preexcited atrial fibrillation			RFp
6	M	21	L	Antidromic AVRT/Mahaim			RFp
7	F	23	L	Antidromic AVRT/Mahaim			RFp
8	F	19	A	Antidromic AVRT/Mahaim			RFp
9	F	23	L	Antidromic AVRT/Mahaim			RFp
10	F	25	PL	Antidromic AVRT/Mahaim			RFp
11	M	35	L	Antidromic AVRT/Mahaim			RFp
12	F	42	L	Antidromic AVRT/Mahaim			RFp
13	F	23	L	Antidromic AVRT/Mahaim			RFp
14	F	8	AL	Mahaim automaticity			RFp
15	M	30	PL	Antidromic AVRT/Mahaim			RFp
16	F	27	L	Antidromic AVRT/Mahaim			RFp
17	F	19	L	Antidromic AVRT/Mahaim			RFp
18	M	12	MS	Mahaim automaticity			RFp
19	F	39	L	Antidromic AVRT/Mahaim			RFp
20	F	15	AL	AVNRT+Mahaim bystander			RFp
21	M	13	L	Antidromic AVRT/Mahaim			RFp
22	M	15	A	Antidromic AVRT/Mahaim			RFp
23	F	25	L	Antidromic AVRT/Mahaim			RFp
24	M	18	L	Antidromic AVRT/Mahaim			RFp
25	F	45	L	Antidromic AVRT/Mahaim			RFp
26	M	24	L	Antidromic AVRT/Mahaim			RF-dis
27	M	11	L	Antidromic AVRT/Mahaim	Yes		S
28	M	26	L	Antidromic AVRT/Mahaim	Yes	LL	RF-dis
29	M	25	A	Antidromic AVRT/Mahaim			RF-dis
30	F	26	P	Antidromic AVRT/Mahaim			RF-dis
31	F	31	L	Antidromic AVRT/Mahaim			RF-dis
32	F	22	L	Antidromic AVRT/Mahaim			RFp
33	M	17	P	Antidromic AVRT/Mahaim			RFp

Abbreviations: AL= right anterolateral, AVRT/Mahaim= reentrant tachycardia with atrioventricular conduction over a Mahaim fiber, CBT= concealed bypass tract, L= right lateral, LL= left lateral, MS= right midseptal, P= posterior, PL= right posterolateral, RPS= right posteroseptal, Site= site of ablation of the Mahaim fiber at the tricuspid annulus (TA), S/RFp/RF-dis= Surgery/Radiofrequency ablation at the atrial/ventricular insertion.

Figure 3: ECG A shows minimal preexcitation (*rsR'* pattern in lead III) in the first 3 QRS complexes. ECG B after intravenous verapamil shows sinus rhythm with A-V conduction over the Mahaim fiber with a long PR interval and overt preexcitation.

Intracavitary signals, Mahaim potential recording and right bundle and His bundle electrogram: We found that the AM interval (atrium-proximal Mahaim potential interval) was always 20-40 msec shorter than the AH interval in patients with minimal preexcitation. In patients without preexcitation the AH interval was shorter than the AM interval (figure 4).

ECG during tachycardia: We analyzed 29 patients with a circus movement tachycardia with anterograde conduction over the Mahaim fiber, one AVNRT with bystander Mahaim conduction, one with atrial fibrillation with anterograde conduction over the Mahaim fiber and two automatic tachycardias arising in the

TABLE II
Electrocardiographic data of Mahaim fibers

BEFORE MAHAIM ABLATION									AFTER ABLATION		
n	Sinus rhythm					Tachycardia			Sinus rhythm		
	PRi	QRS width	lead III pre	q-I pre	q-V6 pre	lead III	axis	QRS width	lead III post	q-I post	q-V6 post
1	0.13	0.11	rS	no	no	rS	0	120	qR	no	yes
2	0.11	0.11	rS	no	no	rS	-30	130	rS	no	no
3*	0.12	0.08	rS	no	no	rS	-45	140	qR	no	no
4*	0.14	0.08	qRs	no	no	rS	-45	130	qR	no	no
5	0.12	0.09	rS	no	no	QS	-45	130	rS	no	no
6	0.12	0.09	qR	no	no	rS	-30	120	qR	no	no
7	0.11	0.08	rS	no	no	rS	-15	120	qR	no	yes
8	0.13	0.08	qR	yes	no	R	60	130	qR	no	yes
9	0.10	0.09	rS	yes	yes	rS	-45	130	Rs	no	yes
10	0.12	0.1	rS	no	no	QS	-60	120	qR	yes	yes
11	0.14	0.09	rS	yes	no	rS	-15	120	rsr'	yes	yes
12	0.11	0.1	rS	no	no	rS	-30	130	qR	no	no
13	0.13	0.12	rS	no	yes	rS	-30	120	qR	no	yes
14	0.14	0.09	rS	no	no	rsr'	0	120	qR	no	no
15	0.15	0.09	rS	yes	yes	rS	-60	120	qR	no	no
16	0.15	0.08	qR	no	no	rS	-45	120	qR	no	yes
17	0.14	0.08	qR	no	yes	rS	-30	120	qR	no	yes
18	0.10	0.11	rS	no	no	QS	-60	120	qR	no	yes
19	0.13	0.11	rS*	yes	no	rS	-45	120	rS	yes	no
20	0.12	0.1	rS	no	no	QS	-45	130	qR	yes	yes
21	0.14	0.08	qR	no	yes	rS	0	130	qR	no	yes
22	0.12	0.09	qR	yes	yes	R	60	130	qR	yes	yes
23	0.12	0.1	rS	no	no	rS	-30	120	qR	no	yes
24	0.11	0.1	rS	no	no	rS	-30	130	qR	yes	yes
25	0.20	0.09	rS	no	no	QS	-30	120	qR	no	no
26	0.10	0.1	rS	no	no	rS	0	120	qR	yes	yes
27*	0.12	0.1	rS	yes	yes	rS	-30	140	rsR'	yes	yes
28*	0.12	0.1	rS	yes	yes	rS	-75	120	RS	yes	yes
29	0.12	0.09	rsR'	no	no	RS	15	140	QR	no	no
30	0.14	0.08	qR	no	no	QS	-60	120	qR	no	no
31	0.12	0.09	qR	no	no	rS	-60	120	qR	no	no
32	0.14	0.08	qR	yes	yes	rS	-30	140	qR	yes	yes
33	0.14	0.09	rsR'	no	no	rS	-30	140	qR	no	no

Abbreviations: AP= accessory pathway. Bold characters indicates patients with minimal preexcitation, n*= patient with Ebstein's disease, , PRi= PR interval, pre/ post= pre/ post RF ablation.

Mahaim fiber. During circus movement tachycardia the cycle length ranged from 430 to 250 msec. QRS width during tachycardia (Table II) varied from 120-140 msec. All patients had a monophasic R wave in lead I and 30 out of 33 patients had rS in V1 during tachycardia (3 pts had a QS in V1).

Figure 4: *Left panel-* patient 6 without preexcitation showing a shorter AH than AM interval. *Right panel-* patient 2 showing a AM shorter than the AH interval resulting in minimal preexcitation with an rS pattern in lead III.

Comparison between the ECG during sinus rhythm and during tachycardia with anterograde conduction over the Mahaim fiber

In all 20 patients with an rS pattern in lead III we found a negative QRS complex in the same lead (either an rS or QS pattern) during tachycardia. Also the patient with atrial fibrillation showed a negative QRS complex. There were 9 patients without the rS pattern in lead III during sinus rhythm, which showed left axis deviation during tachycardia. Three patients showed concordance between absence of an rS pattern during sinus rhythm and their tachycardia QRS pattern (all three patients had an anterior Mahaim) (figure 5).

Post-ablation 12-lead ECG

In the 24 patients showing minimal preexcitation in the 12-lead ECG, 6 patterns were observed in lead III during sinus rhythm after Mahaim ablation. The most common QRS configuration was a qR or QR pattern, found in 18

Figure 5: This patient had an anterior Mahaim fiber and an intermediate QRS axis (50°) during sinus rhythm and antidromic tachycardia

patients, **Rs** in 1 patient, **RS** in 1 patient, **rs** in 2 patients, **rsR'** in 1 patient and a **rsr'** in 1 patient. Assessment of the left precordial leads after ablation showed that in only 9 patients the previous ECG pattern changed with the development of a small **q** wave, while the other patients showed the same pre-ablation QRS complex. Figure 2 gives examples when comparing the QRS pre and post ablation.

Correlation between ECG findings and Mahaim fiber location:

The **rS** morphology in lead III was not seen in the 3 cases with an anteriorly located Mahaim, or the 2 patients with a posterior Mahaim. The distribution of the atrial end of the Mahaim fiber in the 20 patients with an **rS** pattern in lead III during sinus rhythm along the tricuspid annulus is depicted in figure 6. It is of interest that the atrial end of the Mahaim fiber with an **rS** pattern in lead III can be found over a large area around the tricuspid annulus, from the anterolateral to the posterolateral and midseptal region.

The presence of an rS pattern in lead III during sinus rhythm in 200 matched controls:

We did a survey in 200 young individuals referred because of palpitations. Twelve out of 200 (6%) matched controls (56% females and mean age 23 ± 12 years)

Figure 6: Proportion of patients with an *rS* pattern in lead III during sinus rhythm in relation to the location of the atrial end of the Mahaim fiber along the tricuspid annulus.

without heart disease and without a history of palpitations showed an *rS* pattern in lead III during sinus rhythm (Table III). However, all of them had a *q* wave in lead I (*qR* or *qRs* patterns).

TABLE III
Lead III pattern in 200 young individuals with palpitations

qR	Rs/RS	qRs	R/I	IS	I S	rSI*	rSR*
30%	22%	12%	18%	4%	6%	2%	6%

DISCUSSION

When accessory atrioventricular pathways have conduction times approaching that of the normal A-V conduction system, little or no preexcitation may be present during sinus rhythm. The reported incidence of minimal preexcitation in the 12-lead ECG during sinus rhythm in patients with decrementally conducting accessory pathways is low. Bardy et al¹⁶ and Klein et al¹⁸ did not find it in any of their patients. McClelland et al¹³ reported that only 1 of his 26 patients displayed preexcitation on the 12-lead

ECG. 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 ECG's considered as normal in cardiology journals^{17, 19-20} and textbooks²¹⁻²². This suggests that the reported low figures of abnormal ECG's in patients with Mahaim bypass tracts is an underestimation. Some authors did acknowledge the presence of minimal preexcitation in 25 to 50% of the patients^{14, 18}. We found an incidence of 72% of minimal preexcitation, mainly in the presence of an **rS** pattern in lead III (60%). It should be stressed that in these patients there is no classic delta wave. It is of interest that the **rS** pattern was found in patients with decremental accessory pathways having their atrial end over a very large area around the tricuspid annulus, from anterolateral to posterolateral, also in the only patient with a midseptal location (figure 6). This supports a ventricular insertion in a small anterolateral area in the right ventricle in or close to the exit of the right bundle branch and also explains (when ventricular activation starts at this site) the absence of a **q** wave in lead I.

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 (figure 1). All 20 patients with an **rS** 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 accessory pathway. Figure 2 depicts most of the patterns of QRS that emerged after successful ablation of the Mahaim fiber.

The fact that 9 patients did not show an **rS** pattern in lead III during sinus rhythm but an LBBB-like QRS with left axis deviation during tachycardia can be explained by impulse conduction over the Mahaim fiber during sinus rhythm slower than impulse conduction over the normal A-V conduction system. We like other authors²³ found day-to-day variability in the expression of minimal preexcitation. This is different from "intermittent" preexcitation which may occur in rapidly conducting AP's with long anterograde refractory periods. Our patients with variable expression of preexcitation did not have long refractory periods of their accessory pathway. There is one case report of sudden death in a patient with similar findings²⁴. Conduction over Mahaim fibers can be so slow that no ventricular preexcitation occurs even during atrial pacing. Still, these so-called latent Mahaim's are capable of being involved in antidromic tachycardias²⁵.

Are all Mahaim fibers inserting close to or in the right bundle branch?

Some Mahaim fibers are probably not inserting in that region. Our 3 patients with anterior Mahaim did not show an **rS** in lead III nor left axis deviation during tachycardia, suggesting that in those fibers the ventricular insertion is not in the vicinity of the right bundle branch⁶ (figure 5).

Septal *q* waves

In our population lead I was more sensitive for minimal preexcitation than lead V6. In patients with atriofascicular pathways inserting close to the apex, ventricular

activation proceeds from an apical toward a basal direction, which may result in **q** wave in lead V6. Minimal preexcitation due to left-sided accessory pathways can be better appreciated in lead V6 which had been shown to be more sensitive than leads I and avL²⁶.

“rS” as a normal pattern in lead III

It has been shown²⁷ that an **rS** pattern in lead III can be found in normal individuals. This may occur during posterior displacement of the apex leading to S waves in leads I, II and III (*S/S2S3* pattern²⁸) and in counterclockwise rotation of the heart resulting in a **qR** in lead I and **rS** in III. However, in those situations a normal **q** wave in lead I is likely to be present. In our survey on 200 ECG's 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.

Limitation of the study- specificity of the rS pattern in the general population

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 accessory pathways²⁹. Based on the prevalence of accessory pathways in the general population³⁰ (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 1.000 individuals), albeit the sensitivity decreases to 45%.

CONCLUSION

In young patients suffering from tachycardias the finding of a narrow QRS with an **rS** pattern in lead III during sinus rhythm should raise the suspicion of the presence of a Mahaim fiber, especially in those showing absence of **q** wave in lead I.

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Chapter 4

Tachycardias associated with Mahaim fibers: a review

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INTRODUCTION

Accessory pathways with decremental conduction are fascinating and uncommon. Their incidence is estimated to be around 3% of the population with accessory pathways¹. Its rarity played a role in our slow progress in understanding their structure and properties²⁻⁴. It took some decades to discover that the proximal end arises in the right atrium and not the A-V node in the majority of Mahaim fibers⁵⁻⁶, with some rare reports of left-sided Mahaims⁷⁻⁸. We now know that nodoventricular and nodofascicular fibers are rare but they may play a role in tachyarrhythmias with successful ablation of such structures in the right midseptal region⁹⁻¹². Fasciculoventricular pathways do not have decremental conduction¹³⁻¹⁵ but because they originate in the normal sub A-V nodal conduction system they are still included in the original family of Mahaim fibers (table I). In this review we will discuss the clinical spectrum of cardiac arrhythmias related to Mahaim fibers.

ECG DURING SINUS RHYTHM

Analysis of the 12-lead ECG during sinus rhythm can be helpful in the clinical diagnosis of a Mahaim fiber. According to our experience up to 70% of the patients manifest minimal preexcitation¹⁶, which can be overlooked because preexcitation is subtle and associated with a narrow QRS complex. There is some day-to-day variability in the occurrence of preexcitation and analysis of ECG's taken at different days can improve sensitivity. The most common abnormality is an rS pattern in lead III (figures: 8A -before MAT- and 10B), with a small r amplitude and an r/S ratio less than 1/3. Absence of a septal q wave in the left precordial leads and especially in lead I may occur in association with the rS pattern¹⁶. The ECG during sinus rhythm in short atrioventricular Mahaim fibers is usually fully preexcited^{1,17}.

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 right 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^{18,19}.

CLASSIFICATION OF THE DIFFERENT CARDIAC ARRHYTHMIAS THAT MAY OCCUR IN THE PRESENCE OF MAHAIM FIBERS:

Arrhythmias related to atrioventricular/ atriofascicular (AV/AF) fibers

1. Antidromic tachycardia with anterograde conduction over a long Mahaim fiber
2. Antidromic tachycardia with anterograde conduction over a short Mahaim fiber
3. AVNRT with bystander Mahaim conduction
4. AVRT with bystander Mahaim conduction
5. Spontaneous fast automatic tachycardia arising in the Mahaim fiber
6. Spontaneous slow automatic rhythm arising in the Mahaim fiber

7. Automaticity induced by radiofrequency ablation at the site of atrial insertion of the Mahaim fiber
8. Atrial fibrillation with anterograde conduction over the Mahaim fiber
9. Non-reentrant preexcited tachycardia due to simultaneous dual Mahaim conduction

Arrhythmias related to nodoventricular/ nodofascicular (NV/ NF) fibers

10. Antidromic nodoventricular/ nodofascicular tachycardia with A-V dissociation
11. Orthodromic tachycardia via a "concealed" NF fiber

Fasciculoventricular (FV) fibers

12. Orthodromic tachycardia with a "bystander" fasciculoventricular pathway
13. Atrial fibrillation with anterograde conduction over a fasciculoventricular pathway

1. ANTIDROMIC TACHYCARDIA WITH ANTEROGRADE CONDUCTION OVER A LONG MAHAIM FIBER

This is the most common type of tachycardia associated with Mahaim fibers.

Figure 1: A LBBB-like tachycardia with a QRS width of 0.12 sec in a patient with a long atrioventricular Mahaim fiber.

The tachycardia (Figure 1) is usually regular with a mean cycle length of 309 ± 52 (220 to 450) msec. The QRS complex width is usually around 0.13 ± 0.01 sec (range 0.11 to 0.15), shows a left bundle branch block 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 Mahaim fiber the axis can be around $+60^\circ$. The QRS width during tachycardia is related to the distal site of insertion of the Mahaim fiber. The closer to the right bundle branch the narrower the QRS. Retrograde VA conduction over the normal conduction system is associated with

a time interval between 100-140 msec. This will also depend whether V-A conduction goes over the right or left bundle branch. 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 atrioventricular reentrant tachycardia using an A-V bypass tract retrogradely or more rarely an AVNRT with LBBB).

2. ANTIDROMIC TACHYCARDIA WITH ANTEROGRADE CONDUCTION OVER A SHORT MAHAIM FIBER

Short atrioventricular Mahaim fibers usually have wider QRS complexes and also longer VH intervals. Figure 2 shows a LBBB-like tachycardia of 215 b/min, and a -30° QRS frontal plane axis in a patient with Ebstein's disease and an associated right

Figure 2: A LBBB-like tachycardia with a QRS width of 0.16 sec in a patient with a short atrioventricular Mahaim fiber. Note the notch in the S wave in V1.

postero-septal accessory pathway, which is utilized as the retrograde limb of the tachycardia circuit. The QRS complex is wide (0,16 sec), 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 atrioventricular Mahaim fiber. Ventricular activation mapping showed early activation at the level of the tricuspid annulus. It is expected that the longer the distance between the distal insertion of the Mahaim fiber and the right bundle branch the wider the QRS will be. The V-A conduction interval was 80 msec, and the retrograde P wave could not be seen on the 12 lead ECG, being buried inside the QRS complex. Ebstein's disease is a common diagnosis in patients with

Figure 3: A LBBB tachycardia is induced during atrial pacing. Atrial pacing before the tachycardia showed a normal PR interval and a minimally preexcited QRS (rS pattern in lead III and absence of q waves in the left sided leads). As shown in figure 3 this is an AVNRT with bystander anterograde conduction over a Mahaim fiber.

Mahaim fibers with an incidence ranging from 10 to 50% (20% in our series of 40 patients). Mahaim fibers are associated with an overt or a concealed accessory A-V pathway in up to 20% of cases. The AP location is usually right-sided or postero-septal and only exceptionally left-sided²¹. Such AP can be used in V-A direction during antidromic tachycardia.

3. AVNRT WITH BYSTANDER MAHAIM CONDUCTION

Atrioventricular nodal reentrant tachycardia (AVNRT) can be associated with a Mahaim fiber^{10, 22, 23}. 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 Mahaim fiber (figures 3 and 4). The QRS complex would be indistinguishable from the real antidromic reciprocating tachycardia because anterograde A-V conduction goes over the Mahaim fiber. The finding of fusion beats during tachycardia is the major clue to diagnose such a mechanism but fusion is almost never present during SVT, when it does, it is usually at induction. During the EP study block in the accessory pathway can be achieved by means of premature stimuli at the atrial (figure 4) or 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 fiber²⁴.

Figure 4: The same patient as in figure 3. 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 AVNRT.

4. AVRT WITH BYSTANDER MAHAIM CONDUCTION

In our series of 40 patients with "Mahaim variants" bypass tracts, associated rapidly conducting accessory pathways occurred in 20% (5 WPW and 3 concealed APs). In spite of that common association, we induced an orthodromic

reciprocating tachycardia with bystander anterograde Mahaim conduction only once (figure 5).

The usual scenario (in 3 patients with multiple pathways) was the induction of 2 different unrelated tachycardias, an orthodromic AVRT and an antidromic tachycardia using the Mahaim pathway as the anterograde limb and the A-V node retrogradely. However, complex reentrant circuits may be seen. The diagnosis of an orthodromic AVRT with bystander anterograde Mahaim conduction is most likely if retrograde conduction during a pre-excited tachycardia is excentric. Non-inducibility after ablation of the rapidly conducting accessory pathway is the definite proof that the decremental pathway does not participate in the circuit²⁴.

5. SPONTANEOUS FAST AUTOMATIC TACHYCARDIA ARISING IN THE MAHAIM FIBER

Spontaneous automaticity arising in the Mahaim fiber has been described occasionally²⁵. The clinical presentation of such rhythms can be as premature beats in a bigeminal pattern, slow rhythms resembling accelerated idioventricular rhythm or as fast non-sustained bursts of repetitive tachycardia. We have seen 2

Figure 5: Induction of a wide LBBB-like tachycardia by 2 ventricular premature beats in a patient with an atriofascicular pathway associated with a right posteroseptal WPW and dual A-V nodal pathways. The first beat shows a narrow QRS. Thereafter bystander anterograde Mahaim conduction occurs until spontaneous conversion to a narrow QRS tachycardia. The narrow QRS tachycardia shows cycle length alternans due to alternate anterograde conduction over fast and slow A-V nodal pathway and retrograde conduction over the postero-septal rapidly conducting accessory pathway.

patients with frequent episodes of repetitive automatic rhythms as the presenting arrhythmia, without a true antidromic tachycardia. There was no V-A conduction over the A-V node and over the Mahaim fiber (figure 6). The QRS morphology during tachycardia equals the one during atrial pacing at the same rate. Patients having spontaneous

Figure 6: The 12-lead ECG shows a slow LBBB-like tachycardia ($QRS = 0,12$ sec) with A-V dissociation, irregular RR intervals and a cycle length between 400-480 msec. The frontal QRS axis is -50° .

automaticity associated with Mahaim fibers are younger (15 ± 7 years) than patients (26 ± 13 years) with Mahaim fibers without spontaneous automaticity²⁶.

6. SPONTANEOUS SLOW AUTOMATIC RHYTHM ARISING IN THE MAHAIM FIBER

Spontaneous slow automaticity arising in Mahaim fibers is an infrequent rhythm that was present in only 7,5% (3 out of 40) of our patients. Those rhythms are clinically silent (figure 7) resembling an AIVR. In 2 of our 3 patients spontaneously occurring automatic beats triggered episodes of antidromic tachycardia (figure 8). Automaticity was abolished after successful catheter ablation of the Mahaim fiber. In both patients it was required to terminate all heat induced Mahaim automaticity (MAT)^{27, 28} to abolish conduction over the Mahaim fiber and spontaneous Mahaim automaticity.

Figure 7: 12-lead ECG showing sinus rhythm (580 msec) in between two episodes of Mahaim automaticity (640 msec). The 4th and 14th QRS complex are fusion beats.

7. AUTOMATICITY INDUCED DURING RADIOFREQUENCY ABLATION AT THE ATRIAL INSERTION OF THE MAHAIM FIBER

Heat induced automaticity is a long known phenomenon occurring when ablating the slow A-V nodal pathway in patients with AVNRT. Some authors believe that heating the compact node at a distance is the most likely explanation²⁹. Others believe that it is caused by heating of A-V nodal tissue at the posterior extensions of the A-V 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 90's. McClelland et al³⁰ found it in 11 out of 23 patients, Heald et al¹ found it in 12 out of 16 patients and called it "stuttering block", Braun et al³¹ found it in 15 out of 15 patients. We found it in 30 out of 33 patients, 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 4 beats or very long-lasting up to 90 seconds (figure 9). The attitude toward the occurrence of MAT changed from previous concern about stability of the catheter³² 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.

Figure 8: 12-lead ECG showing automatic beats among minimally preexcited sinus beats. Antidromic circus movement tachycardia is triggered by an automatic beat with retrograde V-A conduction through the A-V node.

8. ATRIAL FIBRILLATION WITH ANTEROGRADE CONDUCTION OVER THE MAHAIM 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 (figure 10) without retrograde conduction over the fiber. After successful ablation of the Mahaim fiber atrial fibrillation did not recur during a follow-up of 2,5 years. Miller et al²⁵ ablated a Mahaim fiber during atrial fibrillation. After ablation atrial fibrillation could not be re-induced.

We reviewed 14 articles^{1, 10, 13, 16, 17, 20, 25, 31, 33-38} with a total of 208 patients with Mahaim fibers and only 4 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 syndrome³⁹. Degeneration of circus movement tachycardia to atrial fibrillation was the mechanism in 25% of the patients with WPW studied by Bauernfeind et al⁴⁰. This mechanism may explain atrial fibrillation in the patient of Brugada et al³⁸ 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. The reason for the small incidence of atrial fibrillation in patients with Mahaim syndrome remains unknown but the presence of the Mahaim fiber itself seems to be important, as in the case reported by Miller et al²⁵, where atrial fibrillation could not be re-induced after ablation of the Mahaim fiber.

Figure 9: Episodes of heat induced automaticity (MAT) during radiofrequency ablation of long Mahaim fibers at the tricuspid annulus: A - short lived MAT (4 beats); and B - MAT lasting almost 2 minutes.

Figure 10: Panel A- atrial fibrillation in a patient with a right lateral long Mahaim fiber. QRS pattern is similar to QRS during atrial pacing (panel C).

Panel B shows sinus rhythm with minimal preexcitation with an rS pattern in lead III.

9. NONREENTRANT PREEXCITED TACHYCARDIA DUE TO SIMULTANEOUS DUAL CONDUCTION IN A MAHAIM FIBER

Nonreentrant supraventricular tachycardia with simultaneous conduction in the fast and slow A-V nodal pathways had been widely reported⁴¹. 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 A-V 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 V-A 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 have recently reported⁴² a patient with a Mahaim fiber showing likewise characteristics: an incessant tachycardia due to 1:2 P/QRS relationship (figure 11), no reentrant antidromic tachycardia could be induced, tachycardia did not respond to antiarrhythmic drugs (sotalol and amiodarone) and he did not have V-A conduction through the Mahaim fiber.

No further dual conduction occurred after successful catheter ablation of the decremental pathway.

Figure 11: A 12-lead ECG during sinus rhythm showing frequent episodes of non-sustained tachycardia based upon one sinus P wave resulting into two QRS complexes. 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 conduction over the A-V node and the Mahaim fiber. Paperspeed: 25 mm/sec.

10. ANTIDROMIC NODO-VENTRICULAR TACHYCARDIA WITH V-A BLOCK OR A-V DISSOCIATION

Antidromic tachycardia with A-V dissociation (or 2:1 V-A conduction) is the most important clue for diagnosing a nodoventricular fiber incorporated in the tachycardia circuit. Ventricular tachycardia is the main differential diagnosis of such tachycardia. It is not possible to pre-excite ventricular activation, because the accessory pathway is taking off from the AV node. Some authors¹³ suggested that failure to pre-excite ventricular activation by atrial premature beats would be an argument for the presence of a NV Mahaim fiber. However, Porkolab et al⁴³ showed failure of decremental right atrial extrastimuli to reset an antidromic tachycardia in a patient with a true atriofascicular 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 A-V 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 A-V dissociation in the same patient, due to reversal of the tachycardia circuit, with anterograde conduction over the A-V node and retrograde conduction over the NV Mahaim fiber, as reported by Gallagher et al⁴.

11. 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 ablated⁹⁻¹².

Those cases showed no pre-excitation at baseline ECG and a narrow QRS tachycardia with ventriculoatrial block or A-V dissociation, and a midseptal concealed NF/ NV fiber was successfully ablated. Engagement of an accessory pathway 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 al¹² there was 1:1 midline V-A conduction and intermittent 2:1 A-V infra-Hisian block. Atrial tachycardia was ruled out because the tachycardia could be terminated by ventricular premature beats without V-A conduction, and an accessory pathway potential preceded the earliest retrograde atrial activity, at the right midseptal area.

Shimizu et al⁴⁴ reported a patient with Mahaim physiology and narrow QRS tachycardia preceded by a normal HV interval with V-A dissociation. He could pre-excite 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 nodoventricular Mahaim fiber.

12. ORTHODROMIC ATRIOVENTRICULAR TACHYCARDIA WITH A "BYSTANDER" FASCICULOVENTRICULAR PATHWAY

Fasciculoventricular pathways have so far not actively been associated with cardiac arrhythmias, but they are commonly associated with additional accessory

pathways¹⁵. One has to be careful assessing the mechanism of any antidromic tachycardia before taking a decision to ablate. A “bystander” fasciculoventricular pathways can be misdiagnosed as a parahisian accessory pathway, while mapping during an orthodromic A-V reentrant tachycardia as shown in figure 12. 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 12-lead ECG (figure 12 A) was consistent

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

with a combination of two anterograde preexcitation patterns (a left lateral accessory pathway and a right-sided or anteroseptal accessory pathway). After

ablation of the left-sided AP the ECG (figure 12 C) 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.

Figure 13: Panel A- A atrial premature beat (fourth beat) blocks in the fasciculo ventricular pathway and the QRS complex shows no pre-excitation. Panel B- An episode of atrial fibrillation showing a "fixed degree" of pre-excitation during atrial fibrillation, atrial pacing and sinus rhythm (panel A).

One should suspect an additional fasciculoventricular pathway when the findings on the 12-lead ECG during sinus rhythm can not be explained only by ventricular preexcitation over one accessory A-V pathway.

13. ATRIAL FIBRILLATION WITH ANTEROGRADE CONDUCTION OVER A FASCICULOVENTRICULAR PATHWAY

Another clue to the diagnosis of a fasciculoventricular pathway can be seen in figure 14 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 R-R intervals in atrial fibrillation and in sinus rhythm (figure 13). This is highly suggestive for an associated fasciculoventricular pathway. The association of atrial fibrillation with a fasciculoventricular pathway is fortuitous with no cause-effect relationship.

CONCLUSION

The large group of accessory pathways comprising the "Mahaim family" having 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 pathways in order to perform the correct therapeutic approach.

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Chapter 5

The electrocardiogram during tachycardia in patients with anterograde conduction over a Mahaim fiber. Old criteria revisited.

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ABSTRACT

Background: Pre-excited tachycardia associated with decrementally conducting right sided accessory pathways usually shows a rather "narrow" QRS complex and can be difficult to differentiate from a supraventricular tachycardia (SVT) with LBBB aberrant conduction.

Methods and Results: In order to prospectively evaluate the sensitivity, specificity, positive and negative predictive value of previously described electrocardiographic criteria to identify preexcited tachycardia due to decrementally conducting accessory pathways (QRS axis between 0 and -75° , QRS width $\leq 0,15$ seconds, an R wave in lead I, an rS pattern in lead V₁, RS > 1 QRS transition after V₄, and cycle length between 220 and 450 milliseconds), we analyzed three groups of patients: 32 patients with an atriofascicular pathway (group I); 8 patients with a long (n = 3) or short (n = 5) decrementally conducting right sided atrioventricular pathway (group II); and, a control group of 35 patients with SVT and LBBB (group III). Presence of all 6 criteria had a 87,5% sensitivity in group I and a 0% sensitivity in Group II. There were 4 false negatives in Group I. The negative predictive value was 82,5% with 6 false positives in group III (5 patients with an aberrant LBBB-shaped tachycardia with ventriculo-atrial conduction over an accessory A-V pathway). The criterion cycle length was not helpful.

Conclusions: Criteria for identifying a tachycardia with anterograde conduction over a Mahaim fiber are only helpful in atriofascicular pathways with a sensitivity of 87,5% and a negative predictive value of 82,5%. The major cause of false positives was a tachycardia with aberrant LBBB conduction and ventriculo-atrial conduction over an accessory A-V pathway.

INTRODUCTION

Twenty years ago Bardy et al¹ reported six ECG features showing a high efficacy in identifying antidromic tachycardia due to nodofascicular accessory pathways. In that study all 22 ECG's with ventricular tachycardia were correctly identified and only 1 of 18 ECG's with antidromic tachycardia was erroneously diagnosed as a Mahaim tachycardia. 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 to a predominantly positive QRS complex after V₄, and (6) a tachycardia cycle length between 220 and 450 milliseconds - became the gold standard in identifying preexcited tachycardia due to anterograde conduction over a decrementally conducting accessory pathway. In the following decades our understanding about the anatomic course as well as the site of the proximal and distal insertion of such pathways has evolved²⁻¹¹. Those pathways originally described as nodofascicular fibers would probably be re-classified today as atriofascicular pathways or decrementally conducting accessory atrioventricular pathways (figure 1). The aim of our study was to evaluate the sensitivity, specificity and positive and negative predictive value of the criteria from the study of Bardy et al to identify patients with tachycardia due to atriofascicular and decrementally conducting accessory atrioventricular pathways with emphasis on the differential diagnosis with LBBB-shaped supraventricular tachycardia.

METHODS

Common electrophysiologic findings

Every patient showed during atrial pacing a decrementally conducting right sided atrio-ventricular bypass tract: progressive atrio-ventricular and AH interval prolongation coupled with a decreasing HV interval resulted in a greater degree of ventricular pre-excitation with a left bundle branch-like morphology². The His bundle deflection was inscribed after the right bundle potential during maximal preexcitation. At maximal ventricular preexcitation, there was a constant QRS-His relationship without further changes on shortening the atrial pacing cycle length. A 12-lead ECG was stored simultaneously with intracavitary signals on a digital polygraph (MS or EP-TRACER, CardioTek BV, Maastricht, The Netherlands).

Definitions of terms

An atriofascicular pathway showed a short V-H interval, an early activation of the RV apex, and late activation at the annulus during pre-excited tachycardia. The atrial insertion was located by the finding of the accessory pathway ("M") potential (we did not differentiate the long atrioventricular decremental pathways inserting close to the distal right bundle area from the atriofascicular pathways).

Figure 1: 12-lead ECG obtained during antidromic tachycardia in a patient with an anterior atriofascicular pathway (QRS = 0.12 sec).

Short decremental atrioventricular pathways: during pre-excited tachycardia there was a long V-H interval, early ventricular activation was found at the tricuspid annulus, and late activation at the RV apex.

Long decremental right superior atrioventricular pathways: during pre-excited tachycardia there was late activation of the right ventricular apex and late activation at the tricuspid annulus, and the 12-lead ECG did not have left axis deviation (QRS frontal plane axis between +30 and +60). Atrial insertion was located by the finding of the "M" potential at the right superior area at the tricuspid annulus.

Accessory pathway location nomenclature was presented in accordance to a previous expert consensus statement¹².

We classified these patients with Mahaim fibers in 2 groups: Group I: 32 patients with right sided atriofascicular pathways (figure 1); Group II: 5 patients with a short decrementally conducting right sided atrioventricular pathway which was characterized by a distal insertion close to the tricuspid annulus (figure 2) plus 3 patients with long right superior decrementally conducting A-V pathways (figure 3). As a control group we examined 35 patients with SVT and aberrant conduction with a LBBB shaped QRS (Group III). All tracings were reviewed by the first author and 2 cardiologists to assess inter-observer variability.

The 40 consecutive patients with decrementally conducting accessory pathways came from 4 Institutions (tables I and II). There were 23 females and 17 males, with a mean age of 25 ± 13 (range 8 to 80) years. Thirty-two patients underwent radiofrequency catheter ablation at the atrial aspect of the tricuspid annulus where a discrete accessory pathway ("M") potential where recorded, while in 5 patients (cases 1, 2, 4, 7, and 8 – table I, AV Mahaim section) catheter ablation was carried out at the ventricular insertion guided by the pace-mapping technique. In 2 patients the pathway was surgically interrupted.

TABLE I
 Clinical data of patients with Mahaim fibers

Case	Age	Gender	Right sided C. AFP site	Diagnosis	AP site	Ebstein	Therapy
1	32	F	AI	AVNRT			RFp
2	19	F	AI			yes	RFp
3	21	F	AI			yes	RFp
4	13	M	AI	WPW	IPS	yes	RFp
5	52	M	AI				RFp
6	21	M	AI				RFp
7	8	F	SA				RFp
8	30	M	AI				RFp
9	27	F	A				RFp
10	19	F	A				RFp
11	12	F	Septal				RFp
12	39	F	A				RFp
13	15	M	SA				RFp
14	17	M	A	WPW/ CBT	IPS/ A	yes	RFp
15	23	F	A	AVNRT			RFp
16	80	M	AI	WPW	IPS		RFp
17	25	F	AI				RFp
18	35	M	A				RFp
19	42	F	A				RFp
20	23	F	A				RFp
21	23	F	A				RFp
22	45	F	A				RFp
23	23	M	A				S
24	11	M	A			yes	RF-dis
25	26	M	A	CBT	P	yes	RF-dis
26	26	F	A	AVNRT			RF-dis
27	31	F	A				RF-dis
28	22	F	A				RFp
29	17	M	I				RFp
30	18	F	A				RFp
31	31	F	A				S
32	8	M	A				RFp
Mean±SD= 26±14							
AVP site							
1	31	M	A				RFp
2	12	F	A	WPW	A	yes	RFp
3	15	M	S				RFp
4	17	F	A				RFp
5	25	M	S				RF-dis
6	19	F	S				RFp
7	50	F	AI				RFp
8	25	M	A				RFp
Mean±SD= 24±10							

Abbreviations: A= anterior*, AFP site= atriofascicular pathway-right atrial insertion, AI= anteroinferior, AVP= atrioventricular Mahaim fiber, C= concomitant, CBT= concealed bypass tract, I= inferior, IPS= infero paraseptal, P= posterior, S= superior, SA= superoanterior, Therapy: RFp= catheter ablation targeting the atrial insertion guided by the proximal AP (M) potential, RFd= catheter ablation at the distal insertion of the Mahaim fiber, S= surgical endocardial ablation. *ESC/WGA/NASPE/P Experts Consensus Statement

The ECG during tachycardia in patients with anterograde conduction over a Mahaim fiber

Table II
Electrocardiographic characteristics of the pre-excited tachycardia

AF Mahaim	Cycle Length 220-450 msec	QRS ≤ 150 msec	Axis (degrees)	Lead I Configuration	Lead V1 Configuration	Precordial QRS transition	Preexcited tachycardia (anterograde, retrograde pathways)	False -
1	280	130	-20	R	LBBB (rS)	V5	AF, His	
2	240	140	-60	R	LBBB (rS)	V6	AF, His	
3	300	130	-60	R	LBBB (rS)	V6	AF, His	
4	320	150	-45	R	LBBB (rS)	V5	AF, RIPS AP+His	
5	330	130	-45	R	LBBB (rS)	>V6	pre-excited (AF) Afib	
6	310	120	-30	qR	LBBB (rS)	V6	AF, His	X
7	380	120	0	R	LBBB (rS)	V5	Mahaim automaticity	
8	380	150	-60	R	LBBB (rS)	V5	AF, His	
9	390	130	-30	R	LBBB (rS)	V6	AF, His	
10	320	130	-30	R	LBBB (rS)	V6	AF, His	
11	420	120	-60	R	LBBB (rS)	V5	Mahaim automaticity	
12	320	140	-45	R	LBBB (rS)	>V6	AF, His	
13	330	140	0	R	LBBB (rS)	V5	AF, His	
14	330	130	0	R	LBBB (rS)	V5	AF, A cAP+His	
15	320	120	-15	R	LBBB (rS)	V5	AVNRT+bystander AF	
16	310	120	0	R	LBBB (rS)	V5	AF, RIPS AP+His	
17	300	130	-45	R	LBBB (rS)	V6	AF, His	
18	340	120	-60	R	LBBB (rS)	V4	AF, His	X
19	340	120	-15	R	LBBB (rS)	V6	AF, His	
20	350	130	-30	R	LBBB (rS)	>V6	AF, His	
21	300	120	-30	R	LBBB (rS)	V5	AF, His	
22	380	120	-30	R	LBBB (rS)	V6	AF, His	
23	320	120	-60	R	LBBB (rS)	V5	AF, His	
24	350	120	0	R	LBBB (rS)	V5	AF, His	
25	300	120	-75	R	LBBB (rS)	V5	AF, His	
26	400	120	-60	R	LBBB (QS)	>V6	AF, P cAP+His	X
27	300	120	-60	R	LBBB (rS)	V6	AF, His	
28	400	130	-30	R	LBBB (rS)	V5	AF, His	
29	280	130	-15	R	LBBB (rS)	V5	AF, His	
30	340	130	0	R	LBBB (rS)	V5	AF, His	
31	340	130	20	R	LBBB (rS)	V5	AF, His	X
32	310	130	-30	R	LBBB (rS)	V5	AF, His	
Mean±SD	332±39	127±8	(-31)±24					4/32 (12.5%)
AV Mahaim								
1	400	160	-5	R	LBBB (rS)	V5	AV, His	X
2	240	200	-15	R	LBBB (rS)	V6	AV, A WPW+His	X
3	420	160	45	R	LBBB (rS)	V5	AV, His	X
4	280	180	-75	R	LBBB (QS)	V6	AV, His	X
5	320	160	30	rsR'	LBBB (rS)	V5	AV, His	X
6	310	140	50	R	LBBB (rS)	V5	AV, His	X
7	320	160	-75	R	LBBB (QS)	V6	AV, His	X
8	310	160	-30	R	LBBB (rS)	V5	AVNRT+bystander AV	X
Mean±SD	325±59	165±17	(-9)±49					8/8 (100%)
P value	P= ns	P< 0.0001	P= 0.07					

Abbreviations: A= anterior, AF= atriofascicular, AV= decrementally conducting atrioventricular accessory pathway, cAP= concealed accessory pathway, Mahaim automaticity= automaticity arising at the decremental pathway, RIPS= right inferoparaseptal, LBBB= left bundle branch block-like pattern, P= posterior.

In all patients the radiofrequency catheter ablation or surgical intervention was successful. The control group consisted of 35 patients (60% female) with left bundle branch block-shaped supraventricular tachycardia and a mean age of 40 ± 20 years old (table III).

Statistical Analysis

Values are given as mean $\pm$ standard deviation. The significance of differences ($p < 0.05$) between groups of clinical and electrocardiographic parameters was assessed by Student's *t* test or Fisher's exact test.

RESULTS

Electrocardiogram during pre-excited tachycardia in patients with an atriofascicular pathway (group I)

The index arrhythmia in the 32 patients with atriofascicular pathways was antidromic tachycardia with anterograde conduction over the atriofascicular pathway in 25 patients, incessant non-sustained spontaneous automatic tachycardia arising at the atriofascicular pathway in 2 patients, preexcited tachycardia due to AVNRT with bystander anterograde atriofascicular conduction in one patient, pre-excited atrial fibrillation in one patient and antidromic tachycardia with anterograde conduction over the atriofascicular pathway and retrograde conduction with fusion between the A-V node and a rapidly conducting accessory pathway (two right inferior paraseptal and one anterior) in 3 patients. Those 3 patients with multiple accessory pathways also had an additional inducible orthodromic atrioventricular reentrant tachycardia. Mean cycle length of the pre-excited tachycardia was 332 ± 39 msec (range 220 to 420). Mean QRS width was 127 ± 8 (range 120 to 150) msec. No patient with an atriofascicular pathway had a QRS larger than 150 msec. Mean QRS axis was -31 ± 24 degrees (range 20 to -75), but only one patient with an atriofascicular pathway showed a positive frontal QRS axis during pre-excited tachycardia. All patients with atriofascicular pathways showed an **R** wave in lead I except case 6 (**qR**). Thirty-one patients showed an **rS** pattern in lead VI (case 26 had a **QS** pattern).

The QRS transition in the precordial leads ($R/S > 1$) occurred after V4 in all but case 18. Sensitivity of all 6 criteria in identifying an atriofascicular pathway was 87,5% in these 32 patients. There were 4 false negatives.

Electrocardiogram during pre-excited tachycardia in patients with an atrioventricular accessory pathway with decremental conduction (group II)

The index arrhythmia in 7 of 8 patients was an antidromic tachycardia using the Mahaim fiber as the anterograde limb of the circuit. In 6 patients the A-V node-His bundle- right bundle branch system was used as the retrograde limb, while in case 2 V-A conduction occurred over the A-V node and a right lateral rapidly conducting accessory pathway. One patient had an AVNRT with bystander anterograde conduction over a short atrioventricular Mahaim fiber (case 8). The mean tachycardia cycle length was 325 ± 59 (240 to 420) msec and within the previously described range. The mean QRS complex width (165 ± 17 msec) was larger than the width of the QRS in patients with atriofascicular pathways ($p < 0,0001$). Five of the seven patients had a QRS complex width above the 150 msec

Figure 2: 12-lead ECG during antidromic tachycardia in a patient with a short decrementally atrioventricular accessory pathway ($QRS = 0.17$ sec).

limit. The mean frontal plane axis (-9 ± 49) in these patients were not significantly different ($p = 0.07$) from the patients with an atriofascicular pathway in spite of a trend toward a less marked left axis deviation. Six patients showed an **R** wave in lead I and 6 patients showed an **rS** pattern in lead V1. No patient with an atrioventricular Mahaim would be diagnosed using the criteria from the study of Bardy et al. Those exclusions were based on the presence of 3 different criteria in 1 patient, 2 criteria in 3 patients and only 1 criterion in 4 patients. No exclusions occurred by the criterion "cycle length".

Control group (group III)

The control group consisting of patients with supraventricular tachycardia and LBBB aberrant conduction had a similar gender distribution but a higher mean age (40 ± 20 years) ($p < 0.001$). The tachycardia mechanisms were A-V reentry due to an accessory pathway in 19 patients (14 left free-wall), AVNRT in 11 patients (1 slow-slow and 10 slow-fast) and atrial reentry in 5 patients. Mean tachycardia cycle length of 331 ± 60 msec was similar to the patients from Group I and II ($p = ns$). The mean QRS complex width (153 ± 23 msec) was also larger than the QRS width in patients with atriofascicular pathways ($p < 0.0001$) but not different from the group II patients (165 ± 17 msec) ($p = ns$). The patients with atrial reentry showed the widest QRS width (188 ± 18 msec). The mean frontal plane axis ($+10 \pm 49$ degrees) in these patients was also significantly different

Table III
Electrocardiographic characteristics of the LBBB-shaped SVT

Case	Cycle Length 220-450 msec	QRS amplitude (R to -75 degree)	Axis	Lead I R	Lead V1 rS	Precordial QRS to axisline	Tachycardia circuit	Diagnosis	False +
1	340	130	-60	yes	yes	V5	AVRT	P-CBT	X
2	320	140	50	yes	yes	V4	AVRT	P-CBT	
3	240	180	60	yes	no	V4	AVRT	RIPS-WPW	
4	240	160	-10	yes	no	V5	slow-fast	AVNRT	
5	260	140	-20	yes	yes	V5	slow-fast	AVNRT	X
6	430	200	90	no (RS)	no	V5	focal RAT/ preexcitant LBBB	AT	
7	480	180	50	yes	yes	V4	atrial flutter/ preexcitant LBBB	Atrial flutter	
8	360	170	-45	yes	yes	V6	slow-fast	AVNRT	
9	360	140	0	yes	no	V5	AVRT	P-CBT	
10	340	140	-60	yes	no	V5	AVRT	P-CBT	
11	280	200	0	yes	no	V3	slow-fast	AVNRT	
12	260	160	60	yes	no	V5	AVRT	P-CBT	
13	310	140	75	no (r)	yes	V6	AVRT	A-WPW	
14	300	140	75	no (qr)	yes	V6	slow-fast	AVNRT	
15	480	160	45	no (rS)	yes	V5	focal RAT/ preexcitant LBBB	AT	
16	300	200	15	yes	no	V6	focal RAT	AT	
17	350	170	-15	yes	yes	V4	slow-fast	AVNRT	
18	250	150	0	yes	no	V5	slow-fast	AVNRT	
19	310	180	60	no (Rr)	no	V4	AVRT	LIPS-CBT	
20	450	200	60	yes	yes	V5	focal RAT/ preexcitant LBBB	AT	
21	320	180	-60	yes	no	V5	AVRT	P-CBT	
22	400	140	-45	yes	yes	V4	AVRT	P-WPW	
23	320	140	60	yes	yes	V4	AVRT	P-WPW	
24	320	140	-90	no (RS)	yes	V4	slow-fast	AVNRT	
25	290	160	-60	no (qR)	no	V6	slow-fast	AVNRT	
26	320	140	60	yes	yes	V5	AVRT	P-CBT	
27	320	120	45	yes	yes	V4	slow-fast	AVNRT	
28	320	130	0	yes	yes	V6	AVRT	P-WPW	X
29	290	140	-30	yes	no	V5	slow-slow	AVNRT	
30	320	130	60	yes	yes	V4	AVRT	P-CBT	
31	310	130	-30	yes	yes	V5	AVRT	P-CBT	X
32	310	140	30	yes	yes	V5	AVRT	P-WPW	
33	400	140	-30	yes	yes	V6	AVRT	P-WPW	X
34	320	130	30	yes	yes	V5	AVRT	P-CBT	
35	380	130	-30	yes	yes	>V6	AVRT	SPS-CBT	X
Mean±SD 331±60 153±23 10±49									6/35 pts (17.4%)

Abbreviations: A= anterior, AP= accessory pathway, AVNP= atrioventricular node pathway, CBT= concealed bypass tract, P= posterior, PI= postero-inferior, RAT= right atrial tachycardia, RIPS= right inferoparaseptal, SPS= superoparaseptal, WPW= Wolff-Parkinson-White.

($p < 0.0001$) from the axis in patients with an atriofascicular pathway, but not significantly different from group II patients. Six patients (17%) would be erroneously (false positives) diagnosed as having a Mahaim tachycardia. The most frequent diagnosis associated with a false positive was the presence of an accessory pathway used for V-A conduction. Five of the 19 patients with A-V orthodromic tachycardia with aberrant LBBB (26%) would be misclassified (figure 4). One of 11 patients (9%) with AVNRT and aberrant LBBB would be classified as a tachycardia using an atriofascicular pathway. No patient with an atrial tachycardia due to atrial reentry was misclassified. Correct identification of aberrant SVT was done by 5 criteria in 1 patient, 4 in 2 patients, 3 in 7 patients, 2 in 10 patients and only 1 criterion in 9 patients.

Inter-observer variability

Comparative analysis of the 74 12-lead ECG's with LBBB-shaped tachycardia by the 3 observers resulted in no disagreement.

Figure 3: 12-lead ECG during antidromic tachycardia in a patient with a long decrementally accessory atrioventricular pathway (QRS = 0,16 sec).

DISCUSSION

Ventricular activation during a QRS with left bundle branch block configuration

Our understanding of ventricular activation during left bundle branch block in humans is based on intraoperative epicardial studies¹³, catheter-based endocardial mapping studies¹⁴ and endocardial mapping study using a non-contact catheter technique¹⁵. These studies showed that in most patients with LBBB, activation started in the anterior RV wall. The delayed left septal activation (the septum is activated from the right to left side, and from an anterior to posterior direction) causes disappearance of the **q** wave in lead I, aVL and in the left precordial leads and also of the **r** wave in V1 in up to 50% of the patients. The activation of the anterior region of the RV can explain the inscription of a small and narrow **r** wave in lead V1 in the other half. In 97% of the group I patients with an atriofascicular pathway the presence of an **r** wave in lead V1 is consistent with pre-excitation of the anterior region of the RV, close to the area where the right bundle branch connects to the RV myocardium. On the other hand, as expected, 37 % of our patients with a LBBB-shaped SVT show a **QS** pattern in lead V1. In spite of a mean axis of -31° the wide range in frontal QRS axis from 0 to -75° can be explained by the quite variable site of early endocardial left ventricular septal activation in the presence of LBBB: high septal, postero-septal (close to the posterior fascicle) or mid-septal¹⁵.

Figure 4: Panel A: A LBBB-shaped tachycardia due to AV reentry in a patient with a concealed left free-wall accessory pathway. Panel B: Antidromic tachycardia with anterograde conduction over an atriofascicular pathway. Applying Bardy's criteria both tachyarrhythmias would be classified as Mahaim tachycardia.

LBBB tachycardia in patients with decrementally conducting accessory atrioventricular pathways

The causes of failure to identify these patients are twofold: the absence of left axis deviation in those 3 patients with long superior atrioventricular Mahaim's, whose distal insertion is not located in the vicinity of the distal right bundle branch, and the wider QRS width associated with a short decremental atrioventricular fiber, leading to a ventricular pre-excitation pattern like the one of a rapidly conducting right sided bypass tract. Six of the 8 patients had a QRS width of more than 150 msec.

Aberrant LBBB-shaped supraventricular tachycardia

The higher age of the control group can be explained by the inclusion of patients with atrial reentry and AVNRT which incidence peaks after the fourth decade. Our finding that 5 of the 6 false positives in the control group were caused by A-V

reentrant tachycardia using an accessory pathway highlights the importance of the tachycardia mechanism. As long as 26% of the LBBB SVT due to an accessory pathway fall within the false positive range, a higher proportion of orthodromic tachycardia in the control group can decrease the negative predictive value of the aforementioned criteria, and vice-versa: the more patients with SVT and pre-existent LBBB are included the higher the positive predictive value, because this group of patients has a mean larger QRS width (188 ± 18 msec).

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 University, sensitivity (87,5% vs. 92%, $p = 0,9$) and negative predictive value (82,5% vs. 91%, $p = 0,5$) of the 6 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 inter-observer variability and only the QRS transition in the precordial leads can be influenced by a mal-positioning of the electrodes, but that criterion would have the least impact on the results: 1 more false negative in group I and 1 more false positive in group III.

CONCLUSION

The previously reported criteria showed reliable efficacy in identifying patients with an atriofascicular pathway, but are of no value in the diagnosis of a decrementally conducting accessory atrioventricular pathway. The tachycardia cycle length was not helpful in making the correct diagnosis.

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Chapter 6

Fasciculoventricular fibers: clinical and electrophysiological characteristics of a variant of pre-excitation

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ABSTRACT

Introduction: Fasciculoventricular tracts are considered a rare form of ventricular preexcitation. Few fasciculoventricular pathways have been reported, in none they have been linked to a reentrant tachycardia.

Methods and Results: Four patients with fasciculoventricular bypass tracts underwent electrophysiologic evaluation. Two patients had a single fasciculoventricular pathway, one inserting anteroseptally and the other in the left ventricle. Two patients also had an atrioventricular bypass tract, with anterograde conduction over the fasciculoventricular pathway during orthodromic atrioventricular reentrant tachycardia. After ablation of the atrio-ventricular pathways the ECG during sinus rhythm and the electrophysiologic study showed ventricular pre-excitation due to a fasciculoventricular bypass tract inserting into the right ventricle. Adenosine triphosphate was helpful in the diagnostic process.

Conclusion: Electrophysiologists should be able to make the differential diagnosis between a fasciculoventricular bypass tract and an anteroseptal accessory pathway to preclude potential harm to the A-V conduction system if one targets a fasciculoventricular pathway for catheter ablation.

INTRODUCTION

Both fasciculoventricular and nodoventricular pathways are varieties of true Mahaim fibers^{1, 2}. Since Gallagher et al³ published the largest series of fasciculoventricular pathways (6 patients) in 1981, few additional cases were reported⁴⁻⁶. A number of reasons account for the paucity of publications such as its rarity and underdiagnosis either because no cardiac arrhythmias occur or because of the small amount of ventricular preexcitation on the electrocardiogram. According to Josephson⁷ it should be regarded as an electrocardiographic curiosity. However, in this era of therapeutic cardiac electrophysiology, one should be able to differentiate an anteroseptal para-hissian bypass tract from a fasciculoventricular pathway, in particular in patients presenting with a tachycardia using an associated atrioventricular bypass tracts.

PATIENTS AND METHODS

We have seen 4 patients with right-sided fasciculoventricular pathways out of 332 patients with manifest ventricular preexcitation referred to our Laboratory for catheter ablation during the last 8 years (1,2%). All four were male and without structural abnormalities of the heart on the transthoracic echocardiogram.

Case reports

Case 1: A 13 year male patient with the WPW syndrome and recurrent paroxysmal tachycardia was referred for catheter ablation. The rest 12-lead ECG (figure 1A) was suggestive of a combination of two anterograde preexcitation patterns (a left lateral accessory pathway and a right-sided or anteroseptal accessory pathway). A pre-excited tachycardia, was induced during electrophysiologic study (figure 1B). A right-sided bypass tract was thought to be responsible for anterograde conduction of the electrical impulse during tachycardia. A left free wall bypass tract was used for ventriculoatrial conduction as shown by an eccentric left sided ventriculoatrial conduction pattern. Since it was not possible to advance ventricular activation by means of atrial premature beats during tachycardia we became suspicious of bystander anterograde conduction over an accessory pathway. We decided to ablate the left lateral bypass tract. Figure 1C shows the pattern of pre-excitation after radiofrequency current ablation of the left free wall pathway.

After ablation there was no ventriculoatrial conduction. The AH interval was 60 msec, the HV interval 25 msec. Incremental atrial pacing increased the AH interval with the development of AV nodal Wenckebach at a cycle length of 320 msec. The effective refractory period of the AV node was 260 msec. The amount of right-sided pre- excitation as well as the HV interval remained unchanged during atrial premature beats (figure 2).

Intravenous adenosine infusion caused transient complete A-V block without escape beats. No pre-excited QRS complex occurred till A-V conduction resumed. These findings are in agreement with a fasciculoventricular pathway inserting into the right ventricle and no further ablation was attempted. The patient remained asymptomatic during a follow-up of 3 years.

Figure 1: (Case 1) A: Baseline 12-lead ECG. The pre-excitation pattern can not be explained by a single accessory A-V pathway (see text). B: The QRS complex during the induced pre-excited tachycardia resembles the QRS complex after ablation of the left sided AP. C: ECG after ablation of the left sided accessory pathway (AP), showing a ventricular pre-excitation pattern suggestive of a right sided insertion of the accessory connection.

Case 2: This 19 year male patient complained of palpitations but denied the occurrence of a sustained tachycardia. The 12-lead ECG showed pre-excitation with a P-delta interval of 0.12 sec. His general practitioner referred him for electrophysiologic evaluation. The delta wave was negative in V1 and $+60^\circ$ in the frontal plane, consistent with an anteroseptal location of the accessory pathway (figure 3). The baseline AH interval was 50 msec, suggesting enhanced A-V nodal conduction, but the AH conduction curve during incremental atrial pacing showed AH prolongation with an A-V nodal Wenckebach conduction pattern at an atrial pacing cycle length of 380 msec. The HV interval was short (20 msec), did not change during atrial pacing, and showed a normal His bundle- right bundle activation sequence (figure 4). The amount of pre-excitation did not change during atrial pacing at increasing rates and induced atrial fibrillation (figure 3).

Ventricular pacing during sinus rhythm showed ventriculoatrial conduction, which was concentric and decremental. Intravenous adenosine was administered in sinus rhythm and during ventricular pacing. In both instances we observed complete conduction block. A few pre-excited escape beats were recorded with the same QRS configuration as during sinus rhythm. No tachycardia could be induced by programmed stimulation. Mapping of ventricular activation during sinus rhythm showed the shortest delta-V interval at the site of His bundle recording (figure 4). No ablation was attempted and the patient is arrhythmia free during a 6-month follow-up.

Figure 2: (Case 1) An atrial premature beat with an interval of 270 ms is conducted with a long AH interval (200 ms) without changing the degree of ventricular pre-excitation and the HV interval.

Case 3: A 19 year old male patient with the Wolff-Parkinson-White syndrome was admitted because of aborted sudden death while swimming. His sister underwent catheter ablation at our Institution 2 years before because of multiple accessory pathways. The patient had frequent episodes of paroxysmal tachycardia

before his collapse. The baseline 12-lead ECG (figure 5A) showed a ventricular pre-excitation pattern that could not be explained by A-V conduction over a single accessory pathway, suggesting the presence of multiple accessory pathways. During electrophysiologic study a very fast antidromic A-V tachycardia was induced by atrial extrastimuli (figure 5B). Ventriculoatrial conduction during tachycardia showed long conduction time and midline atrial activation. During ventricular pacing, however, ventriculoatrial conduction was left sided. We decided to ablate this pathway, which was successfully accomplished by the transaortic approach and another pattern of ventricular pre-excitation emerged (figure 5C). A midseptal accessory AV bypass tract with bi-directional conduction was also successfully ablated at the base of the fasciculoventricular pathway

Figure 3: (Case 2) An episode of self-terminating atrial fibrillation showing a "fixed degree" of pre-excitation during atrial fibrillation and sinus rhythm. Note a delta wave configuration suggestive of an anteroseptal insertion of the accessory connection.

triangle of Koch. After ablation of those two AP, pre-excitation persisted but with a narrower QRS and a negative delta wave in V1 (figure 5D). Tachycardia could no longer be induced. The AH interval was short but A-V nodal conduction assessment showed a normal behavior during incremental atrial pacing and atrial premature stimuli. The HV interval was short and remained fixed during

incremental atrial pacing as well as the amount of ventricular pre-excitation. Response to intravenous adenosine was 2:1 A-V block during sinus rhythm without any change in the QRS configuration. The diagnosis was made of a inserting in the right anteroseptal area. The patient is free of arrhythmias during a 1-year follow-up.

Figure 4: (Case 2) Left panel: sinus rhythm with pre-excitation. Right panel: High right atrial stimulation (400 msec) showing prolongation of the AH interval with a fixed HV interval and similar degree of pre-excitation as during sinus rhythm.

Case 4: A 51 year asymptomatic male patient was referred for electrophysiologic evaluation because ventricular preexcitation was found during a routine pre-operative examination. The rest 12-lead ECG (figure 6-A left panel) showed a short PR interval (0,10 sec) and minimal pre-excitation with a RBBB-like pattern suggestive of a left sided accessory pathway. Atrial activation pattern during right ventricular pacing was concentric with decremental V-A conduction. Incremental

atrial pacing at the coronary sinus ostium resulted in prolongation of the AH interval but no change in the HV interval and the degree of preexcitation. Atrial premature beats with a coupling interval below 300 msec showed a normal HV interval followed by a narrow QRS complex (figure 6-A right panel and figure 6-

Figure 5: (Case 3) Panel A: 12-lead ECG with a delta axis in the frontal plane at 60° with a discordant delta axis in the horizontal plane suggesting multiple accessory A-V pathways. Panel B: Antidromic tachycardia using 2 accessory pathways (a right midseptal and a left free wall accessory pathway) for anterograde conduction. Panel C: After ablating the left sided AP the 12-lead ECG is compatible with a right midseptal accessory pathway. Panel D: After ablating the midseptal AP the ECG still shows ventricular preexcitation in the anteroseptal area but with less pre-excitation.

B). Those complexes were also seen as short RR intervals during induced atrial fibrillation. Intravenous adenosine (up to 36 micrograms) did not cause A-V block,

it resulted in sinus bradycardia and a junctional rhythm with a preexcited QRS complex. A fasciculoventricular pathway inserting in the left ventricle was the likely diagnosis and catheter ablation was not performed.

Figure 6: (Case 4) *A*: left panel: Twelve-lead ECG showing sinus rhythm, a short PR interval, and a QRS configuration consistent with minimal left sided pre-excitation. Right panel: coronary sinus pacing did not increase the degree of pre-excitation. An atrial premature beat causes narrowing of the QRS complex by conduction block of the fasciculoventricular pathway. *B*: A 240 msec coupled atrial premature beat delivered at the coronary sinus ostium causes narrowing of the QRS complex with normalization of the HV interval, suggesting block of the fasciculoventricular pathway.

DISCUSSION

We found fasciculoventricular pathways in 4 out of 320 (1.2%) consecutive patients referred for electrophysiologic study with ECGs showing ventricular pre-excitation. Previous reports in predominantly adult populations gave similar figures: 6 (1.9%) out of 308 patients³ and 2 (1.8%) out of 111 patients⁸.

Enhanced A-V nodal conduction was a common finding among the patients studied by Gallagher et al³. However, our 4 cases, in spite of having a short intranodal conduction time (AH interval) during sinus rhythm, showed normal AH interval lengthening and A-V nodal behavior during incremental atrial pacing.

The classic findings of a fasciculoventricular connection include prolongation of the AH interval without changes in the HV interval, a constant degree of pre-excitation, and a fixed relationship between the His bundle and the right bundle potential, during incremental atrial pacing. This was found in all our 4 patients.

We found that intravenous adenosine was very useful as a method to assess the relationship between the fasciculoventricular pathway and its connection to the A-V node-His bundle system. The use of adenosine triphosphate as a diagnostic tool is based on its transient marked negative A-V nodal dromotropic effects⁹. In addition, adenosine shortens the action potential duration of an accessory pathway¹⁰.

Adenosine did not elicit a greater degree of pre-excitation in any of our patients, but created second degree or complete A-V block with junctional beats with the same degree of pre-excitation and short HV interval as during sinus rhythm. His bundle pacing producing an identical QRS as during sinus rhythm is another method to demonstrate the presence of a fasciculoventricular fiber. However, we found it difficult to get reliable and stable His bundle pacing. Two out of our 4 cases had associated bypass tracts and circus movement tachycardia. Gallagher et al³ reported associated WPW syndrome in 1 out of 6 patients, Sallee III et al⁶ found multiple accessory pathways in 1 out of his 3 children, and the patient reported by Kottkamp et al⁴ also had a left lateral bypass tract. The child reported by Ganz et al⁵ did not have associated WPW. Taking all these patients with fasciculoventricular pathways into account, including ours, the incidence of associated WPW would be 33% (5 out of 15 patients). Multiple accessory pathways are estimated to occur in up to 13% of WPW patients¹¹. Higher figures may represent bias due to referral of symptomatic patients. Nonetheless, it is important to recognize in patients with ventricular pre-excitation referred for catheter ablation, that a fasciculoventricular pathway and not an antero-septal bypass tract is present, avoiding damage to the AV node-His bundle system if such pathway is mistakenly targeted for catheter ablation.

CONCLUSION

One should be especially suspicious for an additional fasciculoventricular pathway when the findings on the 12-lead ECG during sinus rhythm can not be explained by ventricular pre-excitation over one accessory A-V pathway only. We ablated the left-sided pathways first (cases 1 and 3) as well as the mid-septal accessory pathway (case 3) after having obtained electrophysiologic data supporting their location and their active role during tachycardia. After ablation of these pathways the remaining pre-excitation should be assessed carefully.

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Chapter 7

The electrocardiogram in patients with Fasciculoventricular pathways: a comparative study with anteroseptal and midseptal accessory pathways

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ABSTRACT

Background: Electrophysiologic findings suggest that fasciculoventricular pathways insert into the septal region. It is also suggested that the surface 12-lead ECG during sinus rhythm is similar to those of patients with anteroseptal and midseptal bypass tracts.

Objective: To describe the electrocardiographic profile of a cohort of patients with fasciculoventricular pathways and compare it with the ECG of patients with anteroseptal, and midseptal accessory pathways.

Methods and results: Four different observers analyzed the 12-lead ECG of 7 patients with fasciculoventricular pathways (group I) and the ECG of 20 patients with midseptal A-V accessory pathways (group II), and 20 patients with anteroseptal A-V accessory pathways (group III). The following variables were analyzed: the ECG frontal plane QRS and delta wave axis, the angle between the QRS and the delta wave axis, QRS width, the R/S ratio in lead III, the presence of a negative delta wave in inferior leads, and precordial lead transition to a R/S > 1.

The ECG of fasciculoventricular pathways shows a variable pattern. It shares some features with an anteroseptal AP and others with a midseptal AP. A narrower QRS width was the most important variable in distinguishing a fasciculoventricular pathway from a septally located A-V bypass tract ($p < 0.0001$). The angle between QRS and delta wave frontal plane axis could differentiate a fasciculoventricular pathway from an anteroseptal bypass tract ($p < 0.0001$) but not from a midseptal AP.

Conclusion: The sinus rhythm ECG of patients with fasciculoventricular pathways shows similarities with anteroseptal and midseptal accessory pathways, but the QRS complex are usually narrower. Fasciculoventricular pathways with a broad QRS complex can not reliably be differentiated from an anteroseptal or a midseptal bypass tract on the ECG during sinus rhythm.

INTRODUCTION

Fasciculoventricular pathways are rare variants¹ of pre-excitation taking off from the bundle of His or the bundle branches and inserting into the ventricular septum². The electrocardiographic recognition of a fasciculoventricular pathway is based upon the finding of a minimal pre-excitation pattern with a normal QRS frontal plane axis with a variable PR interval^{1,3}. Although playing no active role in tachycardia circuits, fasciculoventricular pathways are frequently associated with rapidly conducting bypass tracts, and they must be correctly differentiated from an anteroseptal bypass tract, to avoid harm to A-V nodal conduction if they are targeted for catheter ablation⁴. There are very few published data on the electrocardiogram of fasciculoventricular pathways³⁻⁶. We carried out this study to describe the electrocardiographic profile in a cohort of 7 patients with fasciculoventricular pathways and compared their characteristics with the ECG pattern of patients having A-V bypass tracts inserting in the anteroseptal and midseptal region.

METHODS

Study population

Fasciculoventricular pathways (group I): During the last 9 years, 8 patients (2%) with fasciculoventricular pathways (figure 1) were diagnosed out of 392 patients with manifest ventricular pre-excitation referred to our laboratory for electrophysiologic evaluation. In 3 of the 8 patients (38%) the fasciculoventricular pathway could only be diagnosed after catheter ablation of a rapidly conducting bypass tract. One of the 8 patients lacked some electrophysiologic data and was deleted from this study. One of them (case 3) had 2 rapidly conducting accessory pathways and had an episode of aborted sudden cardiac death. Six patients were male. Mean age was 29 ± 16 (range 13 to 54) years. Four patients were referred because of paroxysmal tachycardia, 2 patients were asymptomatic and were studied for risk assessment of preexcitation and 1 patient had palpitations. Two patients (29%) had paroxysmal A-V nodal reentrant tachycardia and one of them also had non sustained repetitive atrial tachycardia (table 1). *Group with atrio-ventricular accessory pathways:* 40 patients (17 were male) with a single accessory A-V pathway with a mean age of 28 ± 13 (range 12 to 55) years. 20 Patients had a midseptal AP (group II), 13 of them located close to the CS ostium and 7 located at the apical region of the triangle of Koch, and 20 patients with an anteroseptal AP (group III). We used the method to analyze the ECG from patients with septal bypass tracts described by Rodriguez et al⁷.

None of our patients had additional congenital or acquired cardiac abnormalities which could have affected the QRS morphology. The electrocardiograms were analyzed independently by the four authors. The electrocardiographic data analyzed were: QRS axis and delta wave axis in the frontal plane, the angle between the QRS and the delta wave frontal plane axis, the R/S ratio in lead III, presence of initial negativity in the inferior leads, the R/S > 1 transition in the precordial leads, PR interval and the QRS width.

Table 1
Clinical, electrocardiographic and electrophysiologic data of patients with fasciculoventricular pathways

Case	Age	Sex	Symptom	EPS	BT site	PR	QRS	QRS	Angle	AH	HV	PW
							width	axis				
1	51	M	no			0.10	0.10	80	20	60	25	320
2	19	M	palpitations			0.09	0.12	70	40	50	25	380
3	19	M	tachycardia	WPW	LL+RMS	0.12	0.12	50	20	75	25	290
4	21	M	no			0.11	0.12	60	0	80	19	350
5	54	M	tachycardia	AVNRT		0.09	0.09	15	45	55	29	360
6	13	M	tachycardia	WPW	LL	0.11	0.15	30	30	60	20	320
7	27	F	tachycardia	AVNRT+AT		0.11	0.15	45	15	60	28	250

Abbreviations: Angle QRS/Delta axis= difference between the frontal plane axis of QRS complex and delta wave, AT= atrial tachycardia, AVNRT= A-V node reentrant tachycardia, BT= bypass tract, EPS= electrophysiologic study, LL= left lateral AP, PW= Wenckebach point, QRS width= seconds, QRS axis= frontal plane axis, RAS= right antero-septal AP, RMS= right midseptal AP, WPW= Wolff-Parkinson-White syndrome.

Definitions

Fasciculoventricular pathway: The baseline HV (H-delta) interval during sinus rhythm is < 35 msec. During atrial pacing at increasing rates the HV interval does not change. Atrial premature beats cause progressive prolongation of the AH interval without any change in the HV interval and QRS configuration, unless they caused block in the fasciculoventricular pathway resulting in a normal HV interval and narrow QRS complex. Response to adenosine triphosphate suggests a fasciculoventricular pathway when prolongation of the PR interval (AH interval) does not change the degree of pre-excitation or complete AV block occurs after the P wave.

Anteroseptal accessory pathway (AP): bypass tracts lying in close proximity to or immediately anterior to the His bundle. The so-called para-Hissian bypass tracts were included in this group.

Midseptal accessory pathway: bypass tracts located in an area anterior to the coronary sinus and below the His bundle.

Accessory pathway location was established by endocardial activation mapping during sinus rhythm and validated by successful catheter ablation.

Polarity of the delta wave: the first 40 msec vector of the QRS complex.

All patients underwent an electrophysiologic study after discontinuation of antiarrhythmic drug for at least 7 days (no patient was taking amiodarone).

Statistical Analysis

Comparison between the accessory pathway location groups and their electrocardiographic parameters were analyzed using the 2-way analysis of variance and the Student Newman-Keuls test. Values are given as mean ± standard deviation. Statistical significance was assumed for p values of < 0.05.

RESULTS

Group I (Fasciculoventricular pathways): after ablating all associated bypass tracts we found the following ECG and electrophysiologic findings:

Electrocardiogram during sinus rhythm: The mean PR interval was 0.10 ± 0.01 (range 0.09 to 0.12) seconds. Mean QRS complex width was 0.12 ± 0.02 (range 0.09 to 0.15) seconds. QRS frontal plane axis ranged from $+15$ to $+80^\circ$ ($50 \pm 22^\circ$). Mean delta wave axis in the frontal plane was $47 \pm 16^\circ$ (range 30 to 60°), excluding case 8 because the delta wave could not be assessed in all limb leads. The angle between the QRS and the delta wave frontal plane axis was $24 \pm 15^\circ$. The R/S ratio in lead III was >1 in 4 patients, $=1$ in 2, and <1 in 1 patient. The

Figure 1: 12-lead ECG of 7 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 pre-excited QRS complex (6, 7).

delta wave was positive in the inferior leads and in V_5 and V_6 in all patients (figure 1). QRS transition ($R/S > 1$) in the precordial leads occurred in lead V_2 in 5 patients, V_3 in 1, and V_4 in 1. Patients 6 and 7, had an ECG pattern indistinguishable from a midseptal bypass tract located at the apical region of the triangle of Koch (figure 2). Patient 1 had a positive delta QRS and the delta wave frontal plane axis was $24 \pm 15^\circ$. The R/S ratio in lead III was > 1 in 4 patients, $= 1$ in 2, and < 1 in 1 patient. The delta wave was positive in the inferior leads and in V_5 and V_6 in all patients (figure 1). QRS transition ($R/S > 1$) in the precordial leads occurred in lead V_2 in 5 patients, V_3 in 1, and V_4 in 1. Patients wave in lead V_1 , suggestive of a left-sided ventricular insertion, while all the others had a flat or negative delta wave in lead V_1 , consistent with a right ventricular insertion.

Electrophysiologic evaluation: Mean AH and HV intervals were 62 ± 10 ms and 24 ± 3 msec, respectively. During right or left atrial extra-stimulation all patients responded with prolongation of the AH interval with a fixed HV interval. In 3 of 7 patients (42%), a critically timed atrial premature beat blocked the impulse conduction in the fasciculoventricular pathway, causing prolongation of the HV interval and normalization of the QRS complex (figure 3). ECG during AVNRT showed a wide QRS tachycardia due to bystander anterograde conduction over a fasciculoventricular pathway in case 7 (figure 4).

All patients received intravenous adenosine as a bolus and responded with prolongation of the PR interval (AH interval) without changing the degree of preexcitation (and the HV interval). During adenosine conducted atrial beats always showed pre-excitation (figure 5).

Group II (Groups of patients with accessory atrio-ventricular pathways)

1. QRS axis in the frontal plane:

Midseptal AP: -15 to $+60^\circ$ (mean $32 \pm 22^\circ$); antero-septal AP: 0 to $+75^\circ$ (mean $45 \pm 25^\circ$).

2. Delta wave axis in the frontal plane

Midseptal and antero-septal AP had an intermediate delta wave axis: 0 to 60° (mean $23 \pm 18^\circ$), 0 to 60° (mean $43 \pm 18^\circ$), respectively.

3. The angle between the QRS and the delta wave axis: midseptal: 10 to 40° (mean $23 \pm 8^\circ$), antero-septal accessory pathway: 0 to 15° (mean $4 \pm 8^\circ$) ($p < 0.0001$, as compared with group I patients).

4. QRS width: the mean QRS width in the 40 patients with accessory A-V pathways was 0.14 ± 0.01 ($p < 0.0001$, as compared with the group I patients).

5. The R/S ratio in lead III:

Seven of the 20 patients with midseptal AP had an R/S ratio equal to 1 in lead III and the R/S ratio was < 1 in the remaining 13 patients. All 7 patients with midseptal accessory pathways with an R/S = 1 had their AP located in the apical part of the triangle of Koch, just below the bundle of His. In all patients with antero-septal AP the R/S ratio in lead III was > 1 .

6. Delta wave negativity in inferior leads:

Figure 2: ECGs of patients 6 and 7 with a fasciculoventricular pathways (FVP) showing a broad RS complex. They are displayed for comparison with ECG of 2 Group III patients with midseptal bypass tracts (MS-AP)

Figure 3: Case 5- A critically timed atrial premature beat blocked in the fasciculoventricular pathway and conducted with a normal HV interval and without ventricular pre-excitation. Shown are recordings from surface leads, and intracardiac electrograms from the high lateral right atrium (HRA), a bipolar recording of the His bundle potential (His bipolar) an unipolar recording from the distal electrode (His unipolar). Recording speed is 100 mm/s.

Midseptal accessory pathways showed positive delta waves in lead III in 12 of the 20 patients and a negative delta wave in lead III in 8 of 20. All 20 patients with anteroseptal AP showed positive delta waves in the inferior leads.

7. Precordial lead transition to an R/S ratio > 1

Midseptal AP: 5 patients showed R/S > 1 in V₂, 11 in V₃, and 4 in V₄. Anteroseptal AP showed a shift to an R/S ratio > 1 mainly in V₄ (16/20 patients), and less often in V₃ (4 patients).

DISCUSSION

The septal region is a complex anatomic region harboring not only the fasciculoventricular pathways but approximately 30% of all accessory atrioventricular pathways. As a consequence, it is expected that the ECG presentation of the fasciculoventricular pathway may share some characteristics with septal atrioventricular pathways. Applying previously published algorithms in the differential diagnosis of septal accessory pathways in patients with fasciculoventricular pathways they will be categorized as anteroseptal or midseptal bypass tracts^{8, 9}. Patients with fasciculoventricular pathways show a variable PR interval. Some authors¹ describe a normal or short PR interval. Gallagher et al³ reported 6 patients with fasciculoventricular pathways with a PR interval < 0.12 seconds and electrophysiologic evidence of enhanced A-V nodal

Figure 4: Case 7. Left panel: ECG during pre-excited AVNRT. Right panel: intracardiac tracing at 100 mm/s showing a fixed HV interval of +10 msec, and a cranio-caudal depolarization of the bundle of His (the proximal His bundle potential precedes the activation at the distal His bundle potential), ruling out an antidromic tachycardia with retrograde conduction over the His-Purkinje system. The findings are consistent with an AVNRT with bystander anterograde conduction over a fasciculoventricular pathway. Mapp d= mapping catheter lying at the bundle of His (Mapp unip= unipolar recording of the tip electrode).

conduction. In our series the mean PR interval was 0.10 ± 0.01 (range 0.09 to 0.12) seconds, but no patient had enhanced A-V nodal conduction. Previous studies^{5,6} concentrated on the QRS configuration and delta wave morphology in lead V₁ which in our cohort showed a wide variability. According to our findings fasciculoventricular 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 3 groups.

Electrocardiographic similarities with anteroseptal accessory pathways

The R/S ratio in lead III was > 1 in 4 of the 7 patients (57%) with fasciculoventricular pathways and in 100% of the patients with an anteroseptal AP ($p = \text{ns}$). In contrast, patients with midseptal accessory pathways never showed an R/S ratio > 1 .

Patients with fasciculoventricular 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 accessory pathway.

Electrocardiographic similarities with midseptal accessory pathways

Patients with fasciculoventricular pathways and patients with a midseptal bypass tract had a similar angle between the QRS and the delta wave axis in the frontal plane ($24 \pm 15^\circ$ and $23 \pm 8^\circ$, respectively) as compared with the angle of $4 \pm 8^\circ$ in patients with an anteroseptal bypass tract ($p < 0.0001$).

Electrocardiographic dissimilarities between fasciculoventricular pathways, midseptal, and anteroseptal accessory pathways

The transition to an R/S ratio > 1 in the precordial leads occurred mainly in V₂ in patients with fasciculoventricular pathways, V₃ in midseptal pathways and V₄ in anteroseptal bypass tracts.

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

The ECG during sinus rhythm in patients with fasciculoventricular pathways will show a short PR interval, usually with a minimal pre-excitation pattern with a QRS 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.

Patients with left-sided bypass tracts may show minimal pre-excitation. According to Bogun et al¹⁰ an rSR' is usually found in those patients in lead V₆, in contrast with a R pattern found in our 8 patients with fasciculoventricular pathways. Mahaim fibers of the atriofascicular variety usually show minimal preexcitation, but an rS pattern can be found in lead III¹¹, while fasciculoventricular pathways usually show a R/S ratio > 1 in lead III.

Clinical implications

Fasciculoventricular pathways are responsible for a variable pre-excitation pattern on surface ECG, as shown in this study. The ECG is similar to ECG of patients with antero-septal and midseptal accessory atrioventricular pathways. A negative test with intravenous adenosine calls for an electrophysiologic study for a correct

Figure 5: Case 7- 12-lead ECG before and after an IV bolus of adenosine, showing prolongation of the PR interval without affecting the amount of ventricular pre-excitation

diagnosis. It has been shown that fasciculoventricular pathways may be associated with other arrhythmogenic substrates like dual AV nodal pathways or rapidly conducting AV bypass tracts, and be involved as bystanders during supraventricular tachycardia, causing ECG patterns of difficult interpretation as shown in figure 4.

Limitations of the study

The main limitation to our study relates to the fact that the fasciculoventricular pathways were not ablated, and we assume that they have a ventricular septum connection based upon the finding of an earliest ventricular activation close the bundle of His. The number of patients with fasciculoventricular pathways is too small to come to a definite conclusion.

Table 2. Comparative electrocardiographic findings in midseptal, anterosseptal accessory pathways and fasciculoventricular pathways

	MS	AS	FVP	p value
$\hat{a}$ QRS	NA	NA	NA	
$\hat{a}$ DELTA	NA	NA	NA	
Angle between QRS and Delta wave axis	$23 \pm 8^\circ$	$4 \pm 8^\circ$	$24 \pm 15^\circ$	<0.0001 (AS:FVP)
R/S ratio in lead III	<1 or 1	>1	<1 or >1	
2 inferior leads with a negative delta wave	0	0	0	
1 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

Abbreviations: $\hat{a}$ QRS= QRS frontal plane axis, $\hat{a}$ DELTA= Frontal plane axis of the delta wave, AS= anterosseptal 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.

CONCLUSION

The ECG of patients with fasciculoventricular pathways shows similarities with anteroseptal accessory pathways and midseptal bypass tracts located at the apex of the triangle of Koch, but the QRS complex are usually narrower. Fasciculoventricular pathways with a large QRS complex can not reliably be differentiated from an anteroseptal or a midseptal bypass tract by the surface ECG. The definite diagnosis requires an intracardiac study observing the 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 accessory pathway is considered.

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Chapter 8

Mahaim fiber: an atriofascicular or a long atrioventricular pathway?

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ABSTRACT

Introduction: There is debate concerning the distal insertion of Mahaim fibers. Some findings favor an atriofascicular fiber connected with the distal right bundle branch while others favor a long atrioventricular structure inserting into the myocardium.

Case report: A patient is reported having a decrementally conducting accessory pathway. Proximal and distal Mahaim potentials were recorded during sinus rhythm, atrial pacing and antidromic tachycardia. Both proximal and distal M potentials always preceded the QRS complex during sinus rhythm and antidromic tachycardia earlier than the right bundle branch potential. During tachycardia the distal M potential was recorded 6 msec before the retrograde right bundle potential. Other arguments consistent with an atrioventricular connection were a change in the QRS configuration during tachycardia after the first radiofrequency pulse at the site of the distal M potential, and absence of right bundle branch block after successful ablation. Conduction through the proximal part of the Mahaim fiber was unaltered after ablation, as assessed by recording the proximal M potential.

Conclusion: Electrophysiologic evidence is presented suggesting a long atrioventricular accessory pathway inserting close to the distal right bundle branch rather than an atriofascicular connection in this patient with a Mahaim fiber.

INTRODUCTION

For more than 30 years the proximal part of a Mahaim fiber was believed to take off from the A-V node¹⁻³. These A-V node-like structures are accessory pathways with long and decremental conduction and are now usually classified as a short atrioventricular pathway, when ventricular activation mapping during sinus rhythm shows early activation at the tricuspid annulus or as an atriofascicular pathway when ventricular activation is found at the lower right ventricle free wall⁴⁻⁵. However, some studies raise the issue of insertion of the distal end of those long decremental pathways in the right bundle branch⁶. We present a case with a long Mahaim fiber which was carefully mapped both at its proximal and distal end. Successful radiofrequency catheter ablation aimed at the distal insertion did not cause right bundle branch block.

CASE REPORT

A young female aged 22 was admitted because of frequent episodes of a left bundle branch block-like tachycardia since the age of 16 years. She had a history of many hospital admissions for acute treatment of her tachycardias which were always terminated with A-V node blocking agents such as verapamil or adenosine. Physical examination, the 12-lead ECG during sinus rhythm and a transthoracic echocardiogram showed no abnormalities. The ECG during regular tachycardia showed a QRS complex (width of 130 msec) with a LBBB-like pattern and a superior frontal plane axis (-15°). The tachycardia was regular with a 1:1 A:V relationship. There was an "R" wave in lead I, and an "rS" in V1 without notches in the fast downstroke. The R/S ratio became > 1 in lead V5 (figure 1).

Electrophysiologic procedure: The patient underwent the EP study in the fasting non sedated state. Catheters (USCI, Billerica, MA, USA) were positioned in the coronary sinus, the His-bundle region, and the high right atrium. During sinus rhythm the basic intervals (figure 2) were normal (PA= 15, AH= 60, RB-V= 28, HV= 40 msec). During atrial pacing at increasing rates the classic "Mahaim pattern" emerged. During ventricular pacing there was retrograde conduction over the A-V node His-Purkinje system, and realizing that we were facing a Mahaim fiber we rearranged the catheters as follows: quadripolar catheters were left in the high right atrium, coronary sinus and right ventricular apex. The decapolar catheter was positioned over the His bundle and right bundle area. Two 8-F ablation catheters were used for mapping the proximal (M_{prox}) and distal (M_{dist}) accessory pathway potentials at the tricuspid annulus and at the antero-apical area of the right ventricle, respectively. Programmed electrical stimulation was done by using stimuli of 0.5 milliseconds duration at twice diastolic threshold. A 12-lead ECG was stored simultaneously with intracavitary signals on a digital polygraph (EP-TRACER, CardioTek BV, Maastricht, The Netherlands). Local electrograms were filtered at 50-500 Hz and amplified at a gain of 10 mm/mV. We found a small M potential of low amplitude at the lateral aspect of the tricuspid annulus. The A- M_{prox} interval was 75 msec and the M-V interval was 30 msec. We attempted to delineate the course of the Mahaim fiber by mapping both the proximal and the distal insertion during sinus rhythm. Mapping in the apical region of the right ventricular free wall we found a huge spike-like (high frequency) potential a short distance from the QRS complex (M_{distal} -V= 8 msec). Both proximal and the distal potentials were simultaneously recorded (figure 2).

Figure 1: (A) The 12-lead ECG before catheter ablation, (B) during antidromic tachycardia, (C) during antidromic tachycardia after the first episode of radiofrequency current, and (D) after successful catheter ablation, without RBBB. Paper speed: 25 mm/sec.

Induction of tachycardia was possible during atrial and ventricular pacing. Late atrial premature stimuli anticipate ventricular activation during preexcited tachycardia, proving that the accessory pathway was used in the tachycardia circuit. Earliest ventricular activation during tachycardia was recorded at the site where we recorded the distal M potential ($M_d-V = 10$ msec and $V-\Delta = -4$ msec) (figure 3). Both proximal and distal M potentials always as well as the right bundle branch potential preceded the QRS complex during sinus rhythm and antidromic tachycardia but not the His bundle branch potential. During tachycardia the anterograde distal M potential was recorded 6 msec before the retrograde right bundle potential. We decided to ablate the accessory pathway at this distal site during sinus rhythm to improve stability of the catheter. However, during current application (15 seconds) we recorded an irregular non sustained rhythm. Tachycardia could still be induced after current delivery, but with a slightly different QRS configuration (figure 1). We then applied radiofrequency current at the same site for almost 2 minutes. Thereafter programmed stimulation could no longer induce a tachycardia. Preexcitation was also abolished during atrial stimulation but we were able to record the proximal Mahaim potential with the same timing as before. During atrial pacing with a cycle length of 360 msec, Wenckebach block at the A-V node level was achieved but the Mahaim fiber was

Abbreviations: A- atrial, Mp/ Md= proximal and distal Mahaim potentials, H/ RB= His bundle and right bundle potentials, V= ventricle

Figure 2: Sinus rhythm. There is no ventricular preexcitation. The A-H interval is shorter than A-Mp interval. The His bundle is depolarized before the right bundle. Paper speed: 200 mm/sec.

still able to conduct the stimuli through its proximal part, but with complete conduction block at its distal portion (figure 4).

DISCUSSION

The issue of the correct understanding and delineation of the course and insertion of the so called Mahaim fibers is still open. We know now, that in spite of former

Figure 3: Antidromic A-V tachycardia over a Mahaim (M) fiber. The distal M potential precedes the QRS complex by 10 msec. Retrograde conduction occurs over the A-V node His-Purkinje system. The anterograde distal M potential is recorded 6 msec earlier than the retrograde right bundle potential (which precedes the QRS complex by 4 msec). Paper speed: 200 mm/sec.

opinions, that Mahaim fibers are usually not connected to the A-V node and His-Purkinje system^{7,8}.

Many authors still believe that the distal insertion of the Mahaim fiber is attached to the normal conduction system particularly the right bundle branch, the atriofascicular concept. However, the findings in this case are consistent with a direct myocardial insertion of the Mahaim fiber. The disappearance of the distal M potential after ablation with preservation of the right bundle potential, the absence of right bundle branch block in spite of 2 minutes of radiofrequency current application at that site, as well as the modification of the QRS complexes during tachycardia after the first radiofrequency pulse (figure 1), suggest distal myocardial arborization of this pathway. The occurrence of an irregular automatic rhythm

(Mahaim automaticity⁹) during application of radiofrequency current and achievement of successful ablation also validate the aforementioned potential as a distal Mahaim potential, and not a right bundle branch potential. In our patient the accessory A-V node-like structure length

Figure 4: Right atrial pacing after successful catheter ablation aimed at the distal insertion of the Mahaim fiber. There is Wenckebach block at the A-V node level with simultaneous conduction through the proximal part of the Mahaim fiber (A-M potential interval), but complete conduction block inside this long Mahaim fiber. Paper speed: 100 mm/sec.

could be radiographically determined by assessing the distance from the proximal to the distal potential as around 5 cm. Other authors have estimated its length from 2 to 5 cm¹⁰⁻¹¹. Noncontact mapping during antidromic tachycardia in one patient with a posterolateral Mahaim¹² showed break out of conduction as far as at the apex, validating old data from intraoperative epicardial mapping⁴. The major electrophysiologic argument used in support of the atriofascicular concept is the timing of the spike-like potential of the distal right bundle branch, which is usually recorded at the onset of the QRS complex during preexcited tachycardia (V-RB interval showing a range from 0 to 20 msec). In case of right bundle branch block this interval increases because retrograde conduction is shifted to the

left bundle branch. However, recent studies using noncontact mapping system show that right ventricular depolarization starts in normal individuals usually at the right ventricle free wall $-9,8 \pm 5,5$ (-4 to -15) msec before onset of the surface QRS¹³. As we have shown in this case the distal M potential was recorded 6 msec ahead of the right bundle potential and 10 msec before the preexcited QRS complex, which is consistent with early activation of the right ventricle from a decremental conduction pathway at a small distance from the distal right bundle branch. According to Rudy¹⁴ electrical impulse propagates through the ventricle with a conduction velocity of 60 cm/second. We can estimate the distance from the distal Mahaim potential from the site where the distal right bundle branch potential was recorded to be around 3,6 mm. Other authors differentiate atriofascicular from long atrioventricular decremental pathways by looking at QRS duration and to V-His (and V-RB) interval during preexcited tachycardia. Our observation suggests that the distance between the myocardial insertion of the Mahaim fiber and the right bundle branch is the key factor for those differences. The absence of a more pronounced slurring at the onset of the QRS during antidromic tachycardia also favors a small distance between the distal accessory pathway and the distal Purkinje fibers in this case.

Limitations of the study

In spite of the electrophysiologic evidence suggesting that this fiber inserted directly over the right ventricular myocardium we can not rule out arborization into the distal Purkinje system rather than the myocardium. Likewise the absence of RBBB does not exclude ablation of some distal Purkinje fibers. A strong argument in favor of a long atrioventricular pathway could have been made if transient block at the right fascicle either caused by bump mapping or cryomapping did not interrupt antidromic tachycardia, but that was not done in this case.

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Chapter 9

Dual conduction in a Mahaim fiber

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ABSTRACT

An 8 year old girl is described suffering from an incessant non-sustained left bundle branch block (LBBB)-like tachycardia refractory to antiarrhythmic drug therapy. An electrophysiologic study revealed the presence of a right sided accessory atriofascicular pathway. The episodes of non-sustained tachycardia were found to be based upon a dual response in atrioventricular conduction over the Mahaim fiber to one P wave. No re-entrant tachycardia could be induced. The arrhythmia was cured by catheter ablation targeting a Mahaim potential at the right lateral tricuspid annulus. The findings can be explained by longitudinal dissociation in a single Mahaim fiber, a fiber distally diverging into two fibers with different conduction times, or (less likely) two closely located Mahaim fibers with different conduction times.

INTRODUCTION

In the presence of two connections between the atrium and the ventricle or dual AV nodal pathways, it is possible to have two QRS complexes after one sinus P wave¹⁻¹¹.

Mahaim fibers, which are accessory atrioventricular pathways with long conduction times and decremental properties, are considered as being composed of accessory A-V nodal tissue¹²⁻¹⁶.

Here we describe a patient with a Mahaim fiber showing 2 QRS complexes after one sinus P wave.

CASE HISTORY

An 8 year old female child was referred because of an almost incessant tachycardia of 2 years duration. Onset of tachycardia was not related to specific factors and was associated with palpitations. Echocardiography showed no abnormality. The 12-lead electrocardiogram (figure 1, left panel) showed episodes of non-sustained wide complex tachycardia with a LBBB-like configuration. During tachycardia a 1:2 relationship between the P waves and the QRS complexes was present. In the absence of tachycardia the sinus beats showed a normal PR interval (0.12 sec), with a minimally preexcited QRS complex with an rS pattern in lead III (figure 2) and absence of septal q waves in the left leads. Twenty-four hour Holter monitoring showed most of the day repetitive bursts of tachycardia. There was a decrease in the number of episodes and beats per run during the night. Pharmacologic therapy was unsuccessful. Antiarrhythmic drugs like verapamil, propranolol, and amiodarone did not change symptoms nor diminished the occurrence of tachycardia runs.

METHODS AND RESULTS

During electrophysiologic study, measurements during sinus rhythm revealed an AH interval of 80 msec; an HV of 25 to 30 msec, a His-right bundle branch (RBB) interval of 20 msec and a QRS width of 90 msec. High right atrial pacing at increasing rates showed typical findings of a accessory pathway with long conduction times and decremental properties with progressive AH delay, reversal of the His bundle-right bundle depolarization, HV shortening and increasing pre-excitation. Wenckebach block in the accessory pathway occurred with high right atrial pacing at 260 msec. The maximal increase in the anterograde conduction time of the Mahaim fiber was 140 msec. A proximal Mahaim (M) potential was recorded at the right lateral annulus with an atrium-Mahaim (AM) interval of 50 msec. We could also record a distal Mahaim potential in the right anteroapical region (figure 2). We observed a 1:2 P wave to QRS complex ratio when the sinus cycle length was shorter than 900 msec. This 1:2 ratio disappeared during atrial pacing at cycle lengths below

Figure 1: *Left panel:* A 12 lead ECG during sinus rhythm showing frequent episodes of non-sustained 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 A-V node and the Mahaim fiber.

Right panel: The 12-lead ECG after catheter ablation of the Mahaim fiber. Cardiac memory is present in III, avR and early precordial leads. Paper speed: 25 mm/sec.

700 msec. Atrial pacing at faster rates resulted in a 1:1 A-V conduction over the "fast" Mahaim pathway and only occasional beats with dual response were seen at the onset of atrial pacing drive. After atrial pacing we observed a transient suppression of the dual response pattern related to a slowed sinus rate because of the overdrive suppression effect of atrial pacing. With acceleration of the sinus rate the dual response pattern resumed. Response to intravenous adenosine was slowing of the A-V conduction time and blocked P waves, with a gradual resumption of the dual response. During ventricular pacing no ventriculoatrial conduction was observed, and no reentrant tachycardia could be induced. Catheter ablation was performed targeting the proximal M potential. Ten pulses of radiofrequency were delivered and an irregular rhythm consistent with heat induced automaticity was seen followed by complete block in the Mahaim fiber. After ablation 1:1 A-V conduction ensued. Lead III now showed a **qR** pattern (figure 1, right panel).

Figure 2: One sinus beat showing an atrium- proximal Mahaim interval less the atrium-His interval. The HV interval is 30 msec. This combination results in a minimally pre-excited QRS-complex. Note also the distal Mahaim potential at the site of earliest activation of the right ventricle. Paper speed: 300 mm/sec.

Assessment of A-V conduction showed A-V conduction over the A-V node without dual A-V node pathway.

DISCUSSION

Duality of conduction over Mahaim fibers because of longitudinal dissociation was

Figure 3. *Left panel:* An atrial premature beat given after an interval of 300 msec shows a dual Mahaim response. *Right panel:* Block occurs over both the Mahaim fiber and the AV node after a premature beat interval of 290 msec.

Figure 4: High right atrial pacing at 800 msec with a 1:2 P/QRS response. There is reversal of the normal His bundle-right bundle sequence (see arrow in the ladder diagram at the bottom). Paper speed: 150 mm/sec.

described to explain cycle length alternans during antidromic tachycardia in patients with Mahaim fibers. This concept was validated by showing a "jump" and a dual conduction pattern over the atriofascicular pathway during atrial extra stimulus testing¹⁷⁻¹⁹. However, to the best of our knowledge there are no previous reports of spontaneous dual Mahaim conduction resulting in a 1 to 2 P/QRS ratio. From patients with nonreentrant tachycardia due to simultaneous conduction over dual A-V nodal pathways we learned that: 1. they are often refractory to antiarrhythmic drug treatment. In fact tachycardia can be facilitated by the use of drugs; 2. Usually no reentrant A-V nodal tachycardia can be induced; 3. They usually show absence of retrograde V-A conduction. Without retrograde invasion into the "slow" pathway at the time of anterograde conduction over the "fast" pathway, conduction can proceed anterogradely over the "slow" pathway and can reach the ventricle a second time. Dual Mahaim conduction in our patient did only occur spontaneously during sinus rhythm or during right atrial pacing over an atrial cycle length range between 700 and 900 msec (figure 4). One may speculate that at slow rates retrograde invasion into the "slow" pathway prevented anterograde conduction over this structure. We could not demonstrate anterograde block in the "slow" Mahaim with the atrial extra stimulus technique. Impulse conduction proceeded over either pathways or none (figure 3).

However during sinus rhythm episodes of a dual response tachycardia terminated by block in the "slow" Mahaim fiber (figure 1). We are not sure about the exact anatomic situation in our patient. Findings can be explained by a single Mahaim fiber with longitudinal dissociation resulting in dual pathways. We cannot exclude a fiber distally diverging into two fibers with different electrophysiologic properties. As shown in figure 1, the QRS complexes during 1 to 2 P-QRS conduction are not the same. These differences can be explained however by different degrees of aberrancy related to changes in the QRS interval.

Two closely located Mahaim fibers with different conduction times seem unlikely in view of the single Mahaim potential, and also because of the observation (figure 3) of block in both pathways on reaching a critical atrial premature beat interval, suggesting a proximal common pathway. Spontaneous enhanced automaticity may also occur in a Mahaim fiber^{20, 21}. This can be excluded in our patient by the strict 1 to 2 relation between the sinus P wave and the QRS complexes. This was also seen during atrial pacing. A-V dissociation was never observed during the non-reentrant tachycardia.

Final proof was the relation between 1 to 2 conduction at critical sinus and atrial pacing rates.

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Chapter 10

Appraisal of Mahaim automatic tachycardia

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ABSTRACT

A series of four patients with right-sided accessory pathways with long conducting times and decremental properties is reported. All patients underwent radiofrequency catheter ablation and target areas were guided by a discrete "Mahaim" potential recorded at the lateral aspect of the tricuspid valve. A slow automatic and irregular rhythm with a QRS morphology similar to that of a fully preexcited QRS complex occurred during radiofrequency current delivery. The occurrence of so-called "Mahaim" automatic tachycardia heralded successful elimination of the accessory pathway in a manner similar to that of junctional automatic rhythm during slow pathway ablation in patients with A-V nodal reentrant tachycardia.

The observation of an automatic rhythm brought about during radiofrequency current ablation of a Mahaim accessory pathway is electrophysiologic evidence of the accessory A-V nodal behavior of this structure.

INTRODUCTION

There has always been an aura of mystery regarding Mahaim¹ accessory pathways. Electrophysiologists long believed Mahaim accessory pathways represented nodofascicular or nodoventricular connections². This misconception was challenged when patients with “Mahaim” fibers underwent surgical³ and radiofrequency (RF) catheter ablation⁴ of the atrioventricular (A-V) node but remained pre-excited. Tchou et al⁵ shed light on the correct anatomic course, proving that the right atrium rather than the A-V node is the proximal insertion of this accessory pathway. The same conclusion was reached when a surgical approach was attempted⁶. In all cases, the authors found aright parietal accessory pathways rather than nodoventricular fibers. The same results were obtained in many centers worldwide⁷⁻¹¹. Therefore, the concept of nodoventricular pathways was replaced by the atriofascicular pathway concept, and nodoventricular pathways became a rare item^{11,12}. The issue of “Mahaim” automatic tachycardia has received little attention in the literature, although there are many publications on the subject of atriofascicular pathways. The observation of automatic rhythms arising during RF ablation of atriofascicular pathways lead to an inevitable analogy with junctional automatic rhythms occurring during slow pathway ablation¹³. The aim of the present article is to describe the electrophysiologic characteristics of these automatic rhythms, which were documented in four patients with right-sided Mahaim-like accessory pathways.

POPULATION

Five patients having electrophysiologic diagnosis of atriofascicular accessory pathways according to criteria described elsewhere³, underwent catheter ablation at our Institution between 1996 and 2001. Ablation was performed successfully by targeting sites where a “Mahaim” (“M”) potential was recorded in 4 of 5 patients (Table 1). Other criteria, such as earliest stimulus-delta wave interval while pacing the right atrium at different sites close to the A-V ring, were sought in 1 of 5 patients. Automatic rhythms due to RF current delivery (Table 2) arose in all patients in whom an accessory pathway potential was recorded and catheter ablation was successful. Neither automatic rhythms nor “Mahaim” potential was recorded in the patient whose procedure was unsuccessful.

RESULTS

Case reports

Case 1: A 21-year-old girl with occasional paroxysmal tachycardia since age 12 years was referred to Biocor Institute for evaluation and treatment of substantial increased frequency of episodes despite previous oral antiarrhythmic therapy with sotalol and then with amiodarone. Echocardiogram disclosed Ebstein’s anomaly with slight

TABLE I
Clinical characteristics

Case	Sex	Age	Ebstein	anti-CMT	AVNRT	ortho-CMT	AF	other	Site
									AP 30° LAO
1	F	21	Yes	180 b/min	170 b/min		No	No	8 o'clock
2	M	13	Yes	200 b/min		138 b/min	No	RPS	9 o'clock
3	F	28	No	240 b/min	180 b/min		No	No	9 o'clock
4	M	50	No				Yes	No	7 o'clock

Abbreviations: anti-CMT= antidromic circus movement tachycardia, ortho= orthodromic, RPS= right postero-septal, AP= accessory pathway, Site= location of atriofascicular at the tricuspid ring, LAO= left anterior oblique view-30°, b/min= beats per minute, F=female, M= male.

displacement of the tricuspid valve. Two different tachycardia patterns were induced during electrophysiologic study: typical A-V node reentrant tachycardia (AVNRT) and an antidromic circus movement tachycardia using an atriofascicular pathway as the anterograde limb and a slow A-V node pathway as the retrograde limb. Late right atrial extrastimuli that did not penetrate the A-V node advanced the timing of the subsequent QRS complex, proving that the accessory pathway was the anterograde limb of the circuit. A clear-cut accessory pathway potential was recorded at lateral aspect of tricuspid annulus (8 o'clock in the 30° left anterior oblique (LAO) view) (figure 1). In all cases RF current was delivered using a 4-mm tip and thermistor large curve EPT (Boston Scientific Co., Watertown, MA, USA) 8-F ablation catheter. At 0.5 second after starting current

TABLE II
Ablation data

Case	M pot.	Sheath	Temp °C	M. Automatic Tachycardia: RFp			
				Start	Last	min-max CL	
1	+	SR02	70	0.2 s	7.8 s	360-720	2
2	+	SR02	50	0.6 s	140 s*	320-700	4*
3	+		60	1.0 s	30 s	340-640	2
4	+	SR01	60	3.0 s	3 s	420-540	2

Abbreviations: M= Mahaim, pot= potential, Sheath: SR02= SwartzTM introducer-DAIG Co., Temp: average temperature achieved, start= time interval between beginning of RF current delivery and MAT appearance, Last= duration of MAT, min-max CL= "Mahaim" Automatic Tachycardia minimum and maximum cycle length, RFp= number of radiofrequency current pulses, *: total RF time of the 2 ablation sessions.

application, an irregular rhythm with QRS of varying width arose, resembling a fully pre-excited complex (figure 2). This salvo of non-sustained tachycardia lasted 7.8 seconds.

The last three beats showed V-A conduction (A') and an atrial-pacing spike dissociated from the atrial potential. After tachycardia terminated a conducted non-sinus P wave without pre-excitation was followed by two other atrial paced beats with right bundle branch block (RBBB). No automatic tachycardia arose thereafter, and RF current was delivered for 60 seconds. The patient had no recurrence during 36-month follow-up. **Case 2:** 13-year-old boy with history of tachycardia since age 4 was referred to our institution for electrophysiologic evaluation. Various antiarrhythmic drugs had failed to prevent recurrences. ECG showed a Wolff-Parkinson-White pattern suggestive of right postero-septal location.

Figure 1: "M" potential morphology and timing in all 3 cases. Recording had been done during high lateral right atrial pacing in cases 1 and 2.

Transthoracic echocardiogram was consistent with Ebstein's anomaly, without major hemodynamic impairment. Two different tachycardias were induced at electrophysiologic study: (1) 138 beats/min RBBB-like morphology orthodromic tachycardia, and (2) 190 beats/min LBBB-like morphology antidromic tachycardia. After ablating the right postero-septal accessory pathway, features typical of atriofascicular pathway and induction of antidromic tachycardia were observed (figure 3). An "M" potential was recorded (9 o'clock in the LAO projection) and targeted for ablation (figure 1). Irregular automatic tachycardia followed RF application and preexcitation disappeared a few seconds after the current was delivered. Automatic rhythm was recorded throughout current delivery (120 seconds), although the salvos progressively shortened. Less than 24 hours later, atriofascicular pathway conduction recurred. The patient underwent a second procedure 1 month later. The "M" potential was recorded at the same site, and there was no evidence of a right posteroseptal pathway.

Figure 2 : (Case 1) "Mahaim" automatic tachycardia arising at the beginning of RF current delivery, lasting 7,8 seconds followed by non pre-excited supraventricular beats (*) with RBBB (Ebstein's anomaly).

Automatic rhythm arose again after RF current delivery. Preexcitation was abolished 3 seconds after RF application started, however, it was decided to terminate all automatic activity (figure 4), which required 20 seconds after starting RF current to achieve. Sinus rhythm then resumed. RF Energy was delivered for 2 minutes. Patient was arrhythmia-free during 46-month follow-up.

Case 3: A 28-year-old rural area primary school teacher was suffering many bouts of tachycardia, particularly while walking. Baseline ECG obtained on different days disclosed variable patterns ranging from minimal to full-blown preexcitation with a normal PR interval. Transthoracic echocardiogram revealed no abnormalities. Spontaneous and induced tachycardia as well as atrial paced QRS complexes shared the same LBBB-like morphology. Two different tachycardias were induced at electrophysiologic study: (1) 180 beats/min with bystander preexcitation AVNRT, and (2) 240 beats/min antidromic tachycardia. While mapping the tricuspid ring a discrete potential (figure 1) was recorded on its postero-lateral aspect and had been targeted for ablation. Automatic rhythm started promptly after current delivery (1 sec) and lasted for 30 seconds (salvos of unsustained irregular rhythm with LBBB-like morphology). The patient had an uneventful course and no arrhythmia recurrence during 5-year follow-up.

Case 4: A 50-year-man with preexcited atrial fibrillation, fast ventricular response and hemodynamic compromise was admitted to the emergency ward of Biocor Institute. It was his first episode of cardiac arrhythmia. He was diagnosed as having ventricular tachycardia. DC cardioversion with 100 joules restored sinus rhythm, and the ECG was apparently normal. Transthoracic echocardiogram disclosed no abnormalities. Typical "Mahaim" physiology was present at electrophysiologic study. There was no V-A conduction and no circus movement tachycardia could be induced.

Figure 3: (Case 2) Panel A- Baseline ECG: sinus rhythm with short PR interval and preexcitation suggestive of a posteroseptal pathway. Panel B- After ablating a right posteroseptal pathway a minimally preexcited QRS complex was recorded. Panel C- during atrial pacing another pattern of preexcitation with a normal PR interval became more evident.

Sustained and self-terminating atrial fibrillation was induced by two extrastimuli delivered at the right atrium. Mean ventricular rate was approximately 220 b/min. The accessory pathway "M" potential was recorded at the postero-lateral tricuspid annulus (7 o'clock in the LAO projection) following an atrial deflection with an

A/V ratio of 1:1 (figure 5). A short-lived burst of automatic tachycardia occurred five seconds after starting radiofrequency current delivery. No additional automatic activity occurred during the last 50 seconds of ablation. Patient was arrhythmia-free during 2-month follow-up.

DISCUSSION

The results of previous reports on atriofascicular pathways are suggestive of a proximal A-V node like component and a distal bundle branch-like component, consistent with an accessory A-V conduction system, in most cases located at the postero-lateral tricuspid annulus, as in our cases, and occasionally elsewhere (interatrial septum¹⁴, mitral ring¹⁵).

Definite proof that such pathways are accessory A-V nodes can only be obtained by fortuitous anatomic-pathologic assessment in patients with clinical diagnosis of atriofascicular pathways. Despite some reports on the occurrence of ectopic A-V nodal tissue¹⁶⁻²⁰, sometimes with clear-cut features of an accessory A-V node, with or electrophysiologic correlation. In this context, characterization of automatic tachycardia while ablating atriofascicular pathways gains relevance.

Accelerated junctional rhythm is known as a response of the A-V node to thermal injury and a common finding in RF ablation for AVNRT. The accelerated rhythms we and other electrophysiologists observed during RF current ablation in patients with like accessory pathways fits in that description of junctional rhythms. One could argue that the rhythms might be related to right ventricular heating. Davies²¹ observed without ventricular connections, there are insufficient data on its anatomic- Mahaim- like accessory pathways fits in that description of junctional rhythms. One could argue that the rhythms might be related to right ventricular heating. Davies²¹ observed less automaticity while ablating at the ventricular insertion. Based on the evidences invariably shown in all patients that there was a comparative cause-effect relationship between the occurrence of the automatic rhythm and successful ablation, the authors do not agree that the rhythms occurred as a result of right ventricular heating.

Furthermore, RF current was always delivered at a site with a large atrial potential (figure 1). Braun et al²² reported automatic rhythms during Mahaim accessory pathway ablation. They found that longer lasting rhythms (23±14 vs. 8±14 seconds/ $p<0.05$) were associated with successful ablation.

Our observation was different: 2 of 4 patients developed short-lived Mahaim automatic tachycardia (case 1-7.8 sec and case 4-3 sec) and long-term successful ablation. The occurrence of automatic rhythms heralded successful ablation of the atriofascicular pathway in the same manner that junctional rhythm is associated with successful ablation and non-inducibility of AVNRT. In three of our patients, Mahaim automatic tachycardia was no longer induced despite effective heating of the target area for as long as 60 seconds (case 2). Whether there is a relationship between complete termination of Mahaim automatic tachycardia by catheter ablation and persistent successful results is not known. This relationship was not present while ablating the slow A-V nodal pathway. In other words: automatic junctional rhythm can be induced by RF current application after successful ablation of AVNRT. Assuming atriofascicular pathways and A-V nodes are related structures, the dissimilarity may be due to the size of the target structure. Posterior extensions of the A-V node²³ may be larger and/or longer than an

Figure 4: (Case 2) Salvos of “Mahaim” automatic tachycardia during ablation. We observed that salvos progressively lasted shorter and heart rate diminished until no automatic rhythm had arisen despite effective heating at the site of M potential recording (continuous tracings at paper speed of 25 mm/sec)

accessory A-V node, accounting for viable A-V nodal tissue that does not participate in the tachycardia circuit.

Clinical implication

Occurrence of “Mahaim” automaticity during RF ablation of atriofascicular pathways seems to predict immediate success of the procedure.

Figure 5: (Case 4) *A*- Simultaneous recording of the His bundle potential and the "M" potential at the site of ablation during sinus rhythm; *B*- During atrial pacing (cycle length of 550 ms) a higher degree of preexcitation due to prolongation of the AH interval occurs, while A "M" interval remains constant.

CONCLUSION

Heat induced automaticity is not observed while ablating regular accessory A-V pathways or normal atrial myocardium. Its occurrence due to RF ablation of atriofascicular pathways strongly suggests that the target structure might have A-V node like tissue.

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Chapter 11

Automaticity in Mahaim fibers

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ABSTRACT

Introduction: Automatic rhythms associated with Mahaim fibers usually occur during radiofrequency catheter ablation. The incidence and significance of spontaneous automaticity in Mahaim fibers is unknown.

Methods and Results: We found spontaneous automatic rhythms in 5 (12,5%) of 40 patients with Mahaim fibers, referred for non pharmacologic therapy because of recurrent episodes of symptomatic tachyarrhythmias, usually antidromic circus movement tachycardia (33 out of 40 patients). Three were female and 2 male. Mean age was 15 ± 7 years as compared to 26 ± 13 years of the patients without automaticity ($P=0.09$). Three patients had both antidromic tachycardia and asymptomatic spontaneous automatic rhythms, recorded during ambulatory electrocardiography (1 patient) or electrophysiologic study (2 patients). In 2 patients the automatic rhythm triggered antidromic tachycardia. Two other patients had non sustained repetitive episodes of wide QRS tachycardia due to automaticity arising in the Mahaim fiber, without antidromic tachycardia. All automatic rhythms were abolished by successful catheter ablation of the Mahaim fibers.

Conclusions: Spontaneous automaticity occurred in 12.5% of our Mahaim patients and may trigger antidromic tachycardia. Spontaneous automaticity which is not seen in rapidly conducting accessory pathways is another argument for the presence of an A-V nodal like structure in Mahaim fibers.

INTRODUCTION

Automatic impulse formation originates in cells with spontaneous diastolic depolarization. Those cells are found in the sinus node, the A-V node, the coronary sinus ostium, and the *crista terminalis*. In spite of the lack of conclusive anatomic-pathologic data¹, there are strong electrophysiological arguments suggesting that Mahaim fibers contain accessory AV nodal tissue such as 1) slow and decremental anterograde conduction², 2) a structure with a proximal A-V nodal component and a distal bundle branch-like component, both with His bundle-like potentials^{3, 4}, 3) conduction block in response to adenosine⁵ and 4) heat induced automaticity during radiofrequency catheter ablation⁶. According to the literature spontaneous automaticity arising in rapidly conducting accessory pathways is a very rare finding^{7, 8}. Spontaneous activity arising in Mahaim fiber has been reported only twice^{9, 10} and its significance is unknown. Our aim therefore is to describe spontaneous Mahaim automaticity in a cohort of 40 patients with Mahaim fibers.

METHODS

Definitions

Every patient showed a decrementally conducting atrio-ventricular bypass tract with progressive atrio-ventricular and AH interval prolongation coupled with a decreasing HV interval leading to a greater degree of ventricular pre-excitation with a left bundle branch-like morphology during atrial pacing². The His bundle deflection was inscribed after the right bundle deflection during maximal preexcitation. At maximal ventricular preexcitation, there was a constant QRS-His relationship without further changes on shortening the atrial pacing cycle length.

Study Population

We retrospectively studied 40 consecutive patients from 4 Institutions having accessory pathways with long anterograde conduction times and decremental properties during sinus rhythm, with tachycardia with anterograde conduction over the decremental pathway, and in whom ablation of the accessory pathway was performed.

There were 24 females and 16 males, with a mean age of 24 ± 12 (range 8 to 80) years. Referral for electrophysiologic assessment was because of a preexcited tachycardia over the Mahaim fiber in 33 patients (table I). Ebstein's disease was diagnosed in 8 patients (20%). In 32 patients the atrial insertion of the fiber was identified by localizing a discrete accessory pathway potential, and in 8 patients by assessing the shortest A-V interval during atrial pacing at different sites. All patients underwent successful surgical ($n = 2$) or radiofrequency catheter ablation ($n = 38$). RF ablation was done from the atrial end in 32 patients, and in 6 patients from the ventricular end after right ventricular pacemapping. Five patients (cases 9, 18, 22, 33 and 36) showed spontaneous automaticity (table I).

Statistical Analysis

Values are given as mean $\pm$ standard deviation. The significance of differences (P

< 0.05) between groups of clinical, electrocardiographic or electrophysiologic parameters was assessed by Student's *t* test or Fisher's exact test.

RESULTS

Spontaneous Automaticity

The 5 patients with spontaneous automaticity were younger than the 35 patients without spontaneous automaticity (15 ± 7 vs. 26 ± 13 years) but the difference did not reach statistical significance ($P = 0.09$). Three were male and 1 had Ebstein's disease. None of the 5 patients had an associated rapidly conducting AP. Although the number of patients are too small for comparison, clinical presentation with antidromic tachycardia did not differ among Mahaim patients without spontaneous automaticity (82%) and those 60% having it (3 out of the 5 patients). In those five patients (12,5%) spontaneous automatic rhythms were found with the same QRS complex configuration as during maximal preexcitation. Mahaim automaticity rates ranged from a slow rhythm occurring during the night, indistinguishable from an accelerated idioventricular rhythm (AIVR) (cycle length = 900 msec), to nonsustained repetitive tachycardia with a cycle length of 360 msec (table II). Three patients referred because of an antidromic tachycardia with anterograde conduction over the Mahaim fiber also showed spontaneous Mahaim automaticity. QRS complex configuration was identical during the reentrant and the automatic rhythm (figure 1). In one patient (case 36) episodes of asymptomatic slow automatic rhythm resembling AIVR occurred at a slower rate during the nighttime and at a faster rate during daytime (figure 3). In two patients (cases 18 and 22) automaticity was the clinical problem. Those patients sought medical treatment because of a long history of nonsustained palpitations. Both patients showed repetitive bursts of nonsustained tachycardia with a left bundle branch block configuration. There was VA dissociation during the majority of the episodes, but in case 18 some tachycardias showed 2:1 retrograde V-A conduction over the A-V node. Automatic rhythm was recorded in all patients during the electrophysiologic study before the administration of isoproterenol. During its infusion there was an increase in both rate and duration of the episodes. The effect of isoproterenol was less pronounced in both patients with repetitive nonsustained automatic runs. In those 2 patients, there were only a slight increase in automaticity rate. Electrophysiologic validation of the site of origin of this automatic rhythm included the recording of a M potential preceding the preexcited QRS complex and retrograde depolarization of the right bundle followed by the His bundle. In patient 9 (figure 2), there was V-A dissociation during the automatic rhythm.

During atrial overdrive stimulation there was a transient suppression without entrainment, acceleration nor persistent termination of the tachycardia runs in cases 18 and 22. Following stimulation tachycardia resumed almost immediately in cases 18 and 22.

TABLE I
Clinical data of patients with Mahaim Fibers

Case	Sex	Age	Site	Clinical arrhythmia	Therapy S/RFP/RFd	MAT RFp	WPW	CBT
1	F	31	L	Antidromic CMT/Mahaim	S			
2	F	32	L	Antidromic CMT/Mahaim; AVNRT	RFp	Yes		
3*	F	19	PL	Antidromic CMT/Mahaim	RFp	Yes		
4*	F	21	PL	Antidromic CMT/Mahaim	RFp	Yes		RPS
				Orthodromic CMT	RFp			
5*	M	13	PL	Antidromic CMT/Mahaim	RFp	Yes	RPS	
6	M	52	PL	Preexcited atrial fibrillation	RFp	Yes		
7*	F	19	PS	Antidromic CMT/RAS AP	RFp	No	RAS	
8	F	22	P	fast palpitations	RFp	Yes		
9	M	21	L	Antidromic CMT/Mahaim	RFp	Yes		
10	F	23	L	Antidromic CMT/Mahaim	RFp	Yes		
11	M	80	PL	AVNRT+Mahaim bystander	RFp	Yes	RPS	
12	F	19	A	Antidromic CMT/Mahaim	RFp	Yes		
13	F	23	L	Antidromic CMT/Mahaim	RFp	Yes		
14	F	25	PL	Antidromic CMT/Mahaim	RFp	Yes		
15	M	35	L	Antidromic CMT/Mahaim	RFp	Yes		
16	F	42	L	Antidromic CMT/Mahaim	RFp	Yes		
17	F	23	L	Antidromic CMT/Mahaim	RFp	Yes		
18	F	8	AL	Mahaim automaticity	RFp	Yes		
19	M	30	PL	Antidromic CMT/Mahaim	RFp	Yes		
20	F	27	L	Antidromic CMT/Mahaim	RFp	Yes		
21	F	19	L	Antidromic CMT/Mahaim	RFp	Yes		
22	M	12	MS	Mahaim automaticity	RFp	No		
23	F	39	L	Antidromic CMT/Mahaim	RFp	Yes		
24*	F	12	L	Antidromic CMT/Mahaim	RFp	No	RL	
25	F	15	AL	AVNRT+Mahaim bystander	RFp	Yes		
26	M	13	L	Antidromic CMT/Mahaim	RFp	Yes		
27	M	15	A	Antidromic CMT/Mahaim	RFp	Yes		
28	F	25	L	Antidromic CMT/Mahaim	RFp	Yes		
29*	M	17	L	Antidromic CMT/Mahaim	RFp	Yes	RPS	RL
				Orthodromic CMT				
30	M	18	L	Antidromic CMT/Mahaim	RFp	Yes		
31	F	45	L	Antidromic CMT/Mahaim	RFp	Yes		
32	M	24	L	Antidromic CMT/Mahaim	RFd			
33*	M	11	L	Antidromic CMT/Mahaim	S			
34*	M	26	L	Antidromic CMT/Mahaim	RFd			LL
35	M	25	A	Antidromic CMT/Mahaim	RFd			
36	F	26	P	Antidromic CMT/Mahaim	RFd			
37	F	31	L	Antidromic CMT/Mahaim	RFd			
38	F	22	L	Antidromic CMT/Mahaim; AVNRT	RFp	Yes		
39	F	17	L	Antidromic CMT/Mahaim	RFp	Yes		
40	M	17	P	Antidromic CMT/Mahaim	RFp	Yes		

Abbreviations: antidromic CMT/Mahaim= antidromic circus movement tachycardia with anterograde conduction over the Mahaim fiber, Bold= patients with spontaneous automaticity, CBT= concealed bypass tract, MAT= Mahaim Automatic Tachycardia during RF ablation, RAS AP= right anteroseptal accessory pathway, S= surgical ablation, RFp= Radiofrequency catheter ablation targeting proximal Mahaim potential at the annulus, RFd= RF targeting distal insertion, Site= tricuspid annulus, A= anterior, AL= anterolateral, L= lateral, MS= midseptal, P= posterior, PL= posterolateral, PS= posteroseptal, *= Ebstein disease.

Figure 1: Spontaneous Mahaim automatic rhythm (left panel) and antidromic tachycardia showing the same QRS complex morphology (right panel).

TABLE II
Electrophysiologic data of the patients with spontaneous automaticity

n	Case	Clinical arrhythmia	AM	Diagnosis	a-CMT by AM	Isuprel	VA conduction Mahaim / AV node	MAT
			CL range (msec)	at		Acceleration		
1	18	automatic runs	550-450	ECG		*	no / no	yes
2	22	automatic runs	360-400	ECG		*	no / yes	no
3	9	a-CMT	720-580	EPS	yes	***	no / yes	yes
4	33	a-CMT	840-550	Holter*/ EPS	yes	***	no / yes	
5	36	a-CMT	900-530	Holter*/ EPS		**	no / yes	

Abbreviations: a-CMT= antidromic circus movement tachycardia, AM= automaticity in Mahaim fiber, CL= cycle length, MAT= Mahaim Automatic Tachycardia during radiofrequency ablation of the atrial insertion, EPS= electrophysiologic study, Holter*= AIVR was suggested as the diagnosis before EPS

Figure 2: Case 9- Registration during the electrophysiologic study at 100 mm/sec showing an automatic rhythm with LBBB-like morphology. A Mahaim (M) potential precedes each QRS (M-V interval= 50 msec). There is retrograde activation of the distal conduction system (right bundle followed by the His bundle potential) but without retrograde conduction to the atrial level, where the rhythm is clearly dissociated from that in the ventricle.

Induction of antidromic CMT by automatic rhythm

In 2 patients (cases 9 and 33) the automatic rhythm triggered episodes of antidromic tachycardia (figure 4). It happened in both cases during isoproterenol infusion. Initiation of antidromic tachycardia was preceded by a long RR cycle due to A-V dissociation caused by automatic beats arising in the Mahaim fiber. The preceding P wave which was not conducted through the A-V node was conducted with a critical slowing in the Mahaim fiber (figure 4). Isoproterenol improved retrograde conduction over the A-V node enabling antidromic circus movement tachycardia to become sustained, but the mode of tachycardia induction was not related to the V-A conduction itself or to slowing of the sinus rate.

Heat Induced Automaticity

Automaticity during ablation at the proximal insertion of the Mahaim fiber: Mahaim automatic tachycardia (MAT) was induced during radiofrequency catheter ablation in 30 out of 33 patients (91%) when catheter ablation was targeted at the proximal Mahaim potential at the tricuspid annulus. MAT usually started promptly after current delivery (usually in less than 5 seconds). MAT duration was variable lasting from 4 beats to almost 2 minutes (figure 5). Its occurrence could not be prevented by atrial pacing during catheter ablation. In 4 patients with prolonged MAT successful ablation was only achieved after complete elimination of that rhythm (figure 5 B).

Automaticity during ablation at the distal insertion of the Mahaim fiber: We did not include in these results the 5 patients who were ablated targeting the distal insertion (table I). They did develop automaticity but because there were slight differences in QRS configuration we could not rule out that the rhythm was arising in the distal part of the right bundle branch or ventricular myocardium.

DISCUSSION

Early investigators^{1, 3, 4, 11, 12} suggested that Mahaim fibers could be a complete accessory atrioventricular conduction pathway with variable length containing A-V nodal like tissue. The intracardiac electrophysiologic testing has been the main tool to solve the Mahaim puzzle. Transient accelerated automatic rhythms arising from the Mahaim fiber during radiofrequency catheter ablation^{6, 13, 14} have been recognized as one of the features that such fibers share with A-V nodal tissue. Those rhythms are analogous to the accelerated junctional rhythm (AJR) arising during slow A-V nodal pathway ablation in patients with A-V node reentrant tachycardia. AJR is considered to be a sensitive marker¹⁵ of a successful ablation site. It seems that heat induced automaticity is as common during ablation of Mahaim fibers as during slow A-V nodal pathway ablation. We found it in 30 out of 33 patients (91%) when the atrial insertion was targeted. Other authors like Braun et al¹⁴ reported similar figures (15 out of 15 cases -100%) while Heald¹³ et al found it in 57% (12 out of 21 patients).

We found in 4 patients that complete termination of heat induced automaticity may be required as an ablation endpoint to achieve long-term success⁶.

Figure 4: Case 33- 12-lead ECG showing automatic beats among minimally preexcited sinus beats. Antidromic circus movement tachycardia is triggered by an automatic beat with retrograde V-A conduction through the A-V node.

Figure 5: (A) A short episode of Mahaim automaticity consisting of 4 beats with LBBB-like morphology. Minimal ventricular preexcitation is present before (see *rS* pattern in lead III) and absent thereafter (*qR* in lead III). (B) Mahaim automaticity starts immediately after RF current delivery (*) lasting almost 1 minute. After MAT was completely terminated Mahaim conduction did not recur.

Spontaneous automaticity arising in a Mahaim fiber was first reported by Kanter et al¹⁶ in a 7 year old child with an incessant bigeminy initially thought to be of ventricular origin. She underwent electrophysiologic evaluation because a 24 hour Holter monitoring showed nonsustained runs suggestive of ventricular tachycardia. During the EP study an atriofascicular pathway was diagnosed. Sosa et al⁹ reported a patient with symptomatic runs of an irregular LBBB-like tachycardia with AV dissociation, most likely due to spontaneous automaticity. Belhassen et al¹⁰ reported one patient with a likely automatic escape rhythm arising in an atriofascicular pathway. It was remarkable to find 12,5 % spontaneous automaticity in this large cohort of 40 patients with Mahaim fibers. These young patients presented with a spectrum of automatic rhythms ranging from asymptomatic slow rhythms to fast and repetitive bursts of tachycardia with LBBB-like morphology. This is another shared characteristic with the A-V node¹⁷. There are a few reports¹⁸⁻²⁰ about young patients with AVNRT showing spontaneous accelerated junctional rhythms (AJR), which can be cured by slow pathway modification with radiofrequency catheter ablation. Epstein et al²⁰ found in 3 out of 5 patients that AJR served as trigger for AVNRT, particularly with AJR at an accelerated rate. We did the same observation in 2 of our 5 patients who had antidromic CMT triggered by automatic beats. He reported an incidence of 17% (5 out of 29 patients) of spontaneous episodic AJR in a population with a mean age of $12,9 \pm 5,2$ (range 4,2 to 25) years, which is similar to our study population with spontaneous automaticity with a mean age of 15 ± 7 (8 to 26)

years. We also found that spontaneous automaticity was terminated after successful catheter ablation matching Epstein's group observation that in their patients with AJR and AVNRT it was required to eliminate or drastically modulate the AJR to achieve a long-term successful result.

The likely mechanism of spontaneous rhythms seen in cases 9, 33 and 36 was normal automaticity. In those patients Mahaim automaticity was dependent on slowing of the sinus rate ("Mahaim escape rhythm") (figure 3). Isoproterenol infusion increased both sinus and Mahaim automaticity but in those patients a greater increase in the discharge rate was seen at the Mahaim fiber level ("Mahaim active rhythm"). Patients 18 and 22 had an incessant repetitive nonsustained tachycardia which could not be terminated nor induced by programmed electrical stimulation. Isoproterenol infusion in those patients caused only a slight increase in the discharge rate. Those findings suggest an abnormal automaticity as the mechanism of their clinical tachycardia.

CONCLUSION

Mahaim automaticity occurring during ablation of its atrial insertion is a common event, and its complete termination may be required in some patients to achieve long-term success.

Spontaneous automaticity arising in the Mahaim fiber can serve as a trigger for antidromic circus movement tachycardia and can be a major clinical problem. It is another characteristic it shares with the electrophysiologic behavior of the A-V node.

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Chapter 12

Short atrioventricular Mahaim fibers: observations on their clinical, electrocardiographic and electrophysiologic profile

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ABSTRACT

Introduction: A short atrioventricular decrementally conducting accessory pathway is an uncommon variant of preexcitation. Available data from small series suggests that their decremental properties might not be caused by A-V nodal-like tissue.

Methods: We compared clinical, electrocardiographic and electrophysiologic parameters in two groups of patients: 8 patients with a short A-V Mahaim pathway (Group A), and 33 patients with atriofascicular pathways (Group B). Radiofrequency catheter ablation was carried out guided by activation mapping at the annulus in Group A patients and targeting the "M" potential in Group B patients.

Results: After ablation of all associated rapidly conducting bypass tracts, 7 of the 8 Group A patients showed clear pre-excitation. In only 1 of 8 patients the short A-V Mahaim fiber was actively engaged in a reentrant tachycardia circuit. During radiofrequency catheter ablation an automatic rhythm occurred in 4/ 8 patients. Intravenous adenosine caused conduction block in the Mahaim fiber in 3 of 5 patients tested. In group B no patient showed clear preexcitation ($p < 0,0001$) while 72% had a minimal preexcitation pattern. Twenty-nine of the 33 patients had a circus movement tachycardia with AV conduction over the atriofascicular fiber. During radiofrequency catheter ablation 30/ 33 patients showed accessory pathway automaticity. Adenosine caused transient block at the atriofascicular pathway in 11 (92%) of the 12 patients tested.

Conclusions: While short decrementally conducting right sided accessory pathways show a typical ECG pattern different from atriofascicular pathways, their electrophysiologic properties do not seem to be uniform. Those pathways can be successfully interrupted by catheter ablation.

INTRODUCTION

Decrementally conducting accessory pathways bypassing the tricuspid annulus, and inserting at the antero-apical region of the right ventricle in or close to the right bundle branch have A-V node like features¹⁻⁷. Current evidence suggests that they are an accessory conduction system with a proximal AV nodal-like structure and distal branching resembling the His-Purkinje system^{3,8}.

However, our understanding of decrementally conducting A-V Mahaim fibers inserting close to the tricuspid annulus is less clear. A better characterization of these fibers is hampered by their rare occurrence. The few reported series⁹⁻¹⁰ are small and show, apart from their decremental properties, no consistent features suggesting the presence of an accessory atrioventricular node. The aim of this study is to describe the electrocardiographic and electrophysiologic characteristics in 8 patients with short Mahaim fibers and to compare these findings with a group of 33 patients with atriofascicular pathways.

METHODS

Definitions:

Decremental conduction property: a cycle length-dependent prolongation of the impulse conduction time ≥ 30 milliseconds through the accessory pathway.

Atriofascicular pathway: The His bundle deflection was inscribed after the right bundle deflection during maximal pre-excitation. At maximal ventricular preexcitation, there was a constant QRS-His relationship without further changes from shortening the atrial pacing cycle length. Ventricular activation at the right ventricular apex occurred earlier than at the tricuspid annulus.

Short A-V Mahaim: A decrementally conducting atrioventricular pathway showing ventricular activation at the tricuspid annulus during maximal pre-excitation earlier than at the right ventricular apical region (figure 1) was defined by the authors as a "short A-V Mahaim fiber". The His bundle deflection was always inscribed before the right bundle deflection either during sinus rhythm or during atrial pacing with maximal preexcitation (figure 2).

Study Population

We retrospectively studied 47 consecutive patients from 4 Institutions having accessory pathways with long anterograde conduction times and decremental properties during sinus rhythm, usually suffering from a tachycardia with anterograde conduction over the decremental pathway, in whom ablation of the accessory pathway was performed. Six patients were ablated targeting the distal insertion of the atriofascicular pathway and were excluded from this study. Every patient included in this series of patients showed a decrementally conducting atrioventricular (A-V) or atriofascicular bypass tract with progressive atrioventricular and AH interval prolongation coupled with a decreasing HV interval leading to a greater degree of ventricular preexcitation with a left bundle branch-like morphology during atrial pacing² (figure 3).

Table I

Clinical, electrocardiographic, and electrophysiologic data of the 8 patients with a short A-V Mahaim fiber

Case	Sex	Age	E C G			Electrophysiologic study							
			PR	ORS	Overt	AP site	Anti-CMT	M potential	M-site	V-RB	Max dec	W	Adenosine
1	M	34	0.08	0.16	yes	cRAS		no	RAL	-35	120	290	yes
2	M	36	0.08	0.16	yes	RMS		yes	RL	-60	60	300	no
3	M	41	0.08	0.17	yes			yes	RAL	-50	50	320	no
4	F	18	0.09	0.16	yes			yes	RP	-40	70	320	block
5	F	40	0.14	0.16	yes			yes	RPS	-45	60	280	block
6	F	45	0.16	0.18	yes		yes	yes	RPS	-38	60	320	no effect
7	F	24	0.14	RBBB	no	RAS		no	RPS	-40	40	430	no effect
8	F	34	0.12	0.16	yes			yes	RL	-40	30	700	block

Abbreviations: Anti-CMT= antidromic circus movement tachycardia; AP site= bypass tract location; Asterisk= Ebstein; Block= conduction block at the Mahaim fiber; F= female; M-site= Mahaim fiber location; M= male; M potential= Mahaim potential; MAT= Mahaim automaticity during ablation; Max dec= maximal A-V decrement at the Mahaim fiber during atrial pacing at increasing rates; cRAS= concealed right antero-septal; RMS= right mid-septal; RAL= right antero-lateral; RP= right posterior; RPS= right postero-septal; V-RB= V-right bundle interval during maximal preexcitation; W= Wenckebach block.

Figure 1: Case 2- Mapping at the lateral aspect of the tricuspid annulus shows early ventricular activation and a likely accessory pathway ("M") potential between the atrial and ventricular deflections (arrow). The ventricular activation of the right bundle branch occurs after activation at the tricuspid annulus.

Figure 2: Case 2- Lack of reversal of the normal sequence of the His-right bundle branch depolarization during atrial pacing with increasing rates.

Group A consisted of 8 patients with a short A-V Mahaim fiber (figure 4). There were 5 females and 3 males. Patients 1 and 2 are brothers. Age ranged from 18 to 45 (mean 34 ± 8) years. One patient (case 7) had Ebstein's disease and 3 patients had an associated rapidly conducting bypass tract (table I). Four patients had recurrent tachycardia, 3 of them were preexcited and 1 had an orthodromic A-V reentrant tachycardia without bystander conduction over the short A-V Mahaim fiber. Only one tachycardia was related to a short A-V Mahaim fiber. The other 3 tachycardias became non-inducible after ablation of their associated rapidly conducting bypass tracts. Patient 8 had intermittent preexcitation.

Group B consisted of 20 females and 13 males, with ages ranging from 8-52 (mean 24 ± 10) years. All patients were referred for electrophysiologic assessment of a wide QRS tachycardia. Pre-excited A-V node reentrant tachycardia using an atriofascicular pathway as a bystander was present in 1 patient. One patient had atrial fibrillation with preexcited QRS complexes and 2 patients were referred because of repetitive episodes of unsustained tachycardia caused by automaticity arising in the Mahaim fiber⁷. Ebstein's disease was diagnosed in 4 (12 %) patients.

Adenosine test

Adenosine triphosphate was administered as a rapid intravenous bolus during atrial pacing. A bolus of 6 mg was followed by another of 12 mg if needed. A non responder was defined as no delay or block in A-V conduction (prolonged P-delta interval or blocked P waves) in spite of the injection of up to 12 mg of adenosine. Recordings of the 12-lead surface ECG and intracardiac electrogram were made using the EP Tracer or MS System (CardioTek BV, Maastricht, The Netherlands).

Figure 3: Schematic illustration showing the site of successful catheter ablation of all decremental accessory pathways in this present series in a left anterior oblique view. The tricuspid and the mitral fibrous annulus were marked for the sake of clarity. This anatomic 3-D model was build up from images obtained from the Visiblehuman project (<http://visiblehuman.epfl.ch/>). Abbreviations: A= anterior, AL= antero-lateral, AS= anteroseptal, AVN: atrioventricular node, CS= coronary sinus, LA= left atrium, MCV= middle cardiac vein, OS= coronary sinus ostium, P= posterior, PL= postero-lateral, PS= postero-septal, RA= right atrium. Triangles= atriofascicular fibers; circles= short A-V Mahaim fibers.

Statistical Analysis

Values are given as mean $\pm$ standard deviation. The significance of differences ($P < 0.05$) between groups of clinical, electrocardiographic or electrophysiologic parameters was assessed by Student's t test or Fisher's exact test.

RESULTS

Pre-ablation ECG findings

Group A: All 8 patients had clear ventricular pre-excitation on their baseline 12-lead ECG. In six patients ventricular preexcitation occurred because of impulse conduction over a short A-V Mahaim fiber. Two of the 8 patients showed preexcitation due to an associated rapidly conducting A-V bypass tract, one right midseptal and one right anterosseptal. After 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 pre-excitation (but only RBBB) during sinus rhythm with preexcitation becoming apparent during atrial pacing (figure 4).

Table II

Comparative analysis between short (Group A) and long (Group B) Mahaim fibers

Variables	Group A	Group B	p value
Female gender	63% (5/8)	60% (20/33)	ns
Mean age	34 $\pm$ 8	24 $\pm$ 10	0,02
Minimal preexcitation	0%	72% (24)	0,08
No preexcitation*	13% (1)	28% (9)	ns
Overt preexcitation*	88% (7)	0%	< 0,0001
Associated WPW	25% (2)	15% (5)	ns
Associated CBT	13% (1)	6% (2)	ns
Ebstein	13% (1)	12% (4)	ns
Mahaim CMT	13% (1)	88% (29)	ns
Block with adenosine	3/5 (60%)	11/12 (92%)	ns
M potential	75% (6)	100% (33)	ns
Mahaim automaticity	50% (4)	91% (30)	ns

Abbreviations: Asterisk= after ablation of associated bypass tracts; CBT= concealed bypass tract; CMT= circus movement tachycardia; M= Mahaim; Mahaim automaticity= during radiofrequency ablation; ns= non significant p value

In the seven patients with clear pre-excitation during sinus rhythm the P-R interval was normal in two patients (cases 5 and 6) and short ($< 0,12$ sec.) in 5 patients. As shown in figure 4, the ECG in preexcitation is similar to the ECG in patients with a rapidly conducting right sided accessory pathway.

Group B: Five patients had an associated rapidly conducting accessory pathway. After ablation no patient showed clear pre-excitation. Twenty-four patients had a minimal preexcitation pattern, showing a QRS complex $< 0,12$ seconds with an rS pattern in lead III in 20 patients as the most prevalent finding (16 of the 20 patients also showed absence of septal q waves in lead I)¹¹. Two patients had an rsR' in lead III and 2 patients had absence of septal q waves in leads I and V6 as the only ECG clue for diagnosing minimal preexcitation¹¹.

Electrophysiologic findings

Group A: During atrial pacing at increasing rates maximal prolongation of the A-delta interval ranged from 30 to 120 (61 ± 26) msec. Wenckebach block over the short Mahaim fiber (figure 5) occurred with a paced atrial cycle length ranging from 700 to 280 msec. During maximal preexcitation the mean onset QRS-RBB interval was 43 ± 8 (25 to 50) msec. No patient showed reversal of His-right bundle potentials during maximal preexcitation. Only one patient (table I) had an antidromic tachycardia with anterograde conduction over the short A-V Mahaim fiber. Three patients had orthodromic tachycardia

with their rapidly conducting AP as the retrograde limb of the circuit. None of these patients showed bystander anterograde conduction over the decremental pathway. The short decrementally conducting accessory pathways were mapped by activation mapping and ablated at an anterolateral (2 patients), lateral (2 patients), posterior (1 patient) and posteroseptal (3 patients) location. An "M" potential, which was not specifically looked for, was recorded in 6 of the 8 patients. The "M" potential can be difficult to assess because it merges with the early local ventricular electrogram. In patient 8, we were able to document intermittent merging of the "M" potential and the local ventricular electrogram during preexcited beats, because during non-preexcited QRS complex, spontaneous conduction block over the Mahaim fiber occurred in its ventricular insertion, distally to the "M" potential (figure 6). During radiofrequency ablation a Mahaim automatic tachycardia occurred in 4 of the 8 patients.

In 2 patients the automatic rhythm was short-lived and consisted of only 3 beats followed by sinus rhythm without preexcitation (figure 7).

Group B: Patients with atriofascicular pathways showed during atrial pacing Wenckebach block and maximal A-delta prolongation with no significant differences from Group A patients. Mean onset QRS-RBB interval during maximal preexcitation was -6 ± 8 msec ($p < 0,0001$ compared with short A-V Mahaim fibers). Thirty of the 33 patients had automaticity during radiofrequency catheter ablation (table II).

Adenosine test

Five patients in Group A and 12 patients in group B were tested. Three of the five patients (60%) and 11 of the 12 patients (92%), respectively responded with conduction block in the decrementally conducting accessory pathway (figure 8).

Figure 4: 12-lead ECG's during anterograde conduction over a short A-V Mahaim fiber in the 8 patients studied. Small black squares mark lead V₁. Cases 2 and 7 ECG's were recorded after ablation of an associated midseptal and right anterosseptal bypass tract respectively. Case 7 (a patient with Ebstein's disease) showed pre-excitation only during atrial pacing (after ablation of an additional anterosseptal accessory pathway, the baseline ECG showed sinus rhythm with RBBB).

Figure 5: Case 7- Wenckebach block in a short A-V Mahaim during right atrial pacing at 430 msec. The ablation catheter (RF) recorded an early ventricular electrogram at the annulus. Recording speed was 100 mm/s.

DISCUSSION

Common features in patients with short A-V Mahaim fibers

The presence of overt pre-excitation in 7 of the 8 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. No reversal of the His bundle-right bundle branch activation occurs because when the wavefront over the Mahaim fiber reaches the right bundle branch region, the His bundle and right bundle branch are already activated (fig.1 and 2). Maximal decremental conduction was not different between Group A and B. No patient had retrograde conduction over the Mahaim fiber.

Discordant features in patients with short A-V Mahaim fibers

The response to an intravenous bolus of adenosine was not uniform in Group A patients. In 3 out of the 5 patients tested, conduction block developed at the Mahaim level. Group B patients showed a different response. Adenosine induced conduction block in the Mahaim

Figure 6: Case 8- Intermittent conduction over a short A-V Mahaim. The 1st QRS complex is not preexcited. An accessory pathway potential ("M") was recorded at the right lateral annulus. The local ventricular electrogram at the lateral aspect of the tricuspid annulus-TA (8:30 o'clock in LAO projection) is recorded after the ventricular electrogram at the apex. The next beat shows preexcitation. The local ventricular electrogram was recorded before the electrogram of the RV apex and merges with the "M" potential. The morphology of the His bundle electrogram and the AH interval does not change during the tracing. Shown are recordings from surface leads, and intracardiac electrogram from the right ventricular apex (RV ap), bipolar (RF) and distal unipolar (RF u) from the ablation catheter, and His bundle (His). Recording speed is 200 mm/s. RAO (upper) and LAO fluoroscopic views of the catheters located at the ablation site and at the bundle of His.

Figure 7: Case 1- A short run of heat induced automatic rhythm from the Mahaim fiber occurring just before block

fiber in 11 of the 12 patients tested. Other authors also found that atriofascicular pathways are sensitive to adenosine^{8, 12}.

Another difference was the occurrence of heat induced automaticity during radiofrequency catheter ablation. Only 4 group A patients (50%) showed ectopic activity arising in the decrementally conducting pathway in contrast to 30 of the 33 patients (91%) of Group B.

A-V node like features

Patients 4, 5 and 8 responded to adenosine and had automaticity emerging during radiofrequency ablation. Patient 1 showed heat induced automaticity but did not respond to adenosine. The electrophysiologic profile of these patients did not differ from patients with atriofascicular pathways, who also respond to adenosine (92%) and show accessory pathway automaticity during ablation (91%). Braun et al¹³ reported likewise heat induced automaticity in his 15 patients (100%) with atriofascicular pathways. We hypothesize that this subset of patients with short A-V Mahaim fibers who responded to adenosine and showed radiofrequency induced automaticity may have an accessory A-V node without a long branching portion as found in patients with atriofascicular pathways. This substrate could be similar to the accessory node-like structure described by Becker et al¹⁴ at the

lateral tricuspid valve annulus that connected to the ventricular myocardium beneath the annulus in a patient with Ebstein's disease and an additional posteroseptal AP.

Short A-V Mahaim fibers without A-V node like behavior

Patients 6 and 7 did not respond to adenosine nor developed automaticity during RF ablation. Patients 2 and 3 did not have automaticity during ablation but were

Figure 8: Case 5- A bolus of 6 mg of adenosine causes block of the atrial paced beats in both the AV node and the short A-V Mahaim fiber.

not given adenosine. It is still possible that patients 2 and 3 had an A-V nodal like structure because not all atriofascicular patients show heat induced automaticity. Decremental conduction can occur for reasons other than an A-V node like structure. Haissaguerre¹⁰ reported a patient who developed decremental conduction after an attempt of radiofrequency catheter ablation over a previous rapidly conducting accessory pathway. Critelli et al^{15, 16} reported one patient with the permanent form of atrioventricular reciprocating tachycardia that developed decremental anterograde conduction over a posteroseptal accessory pathway after A-V node ablation. Histologic examination showed no A-V node like-tissue but an accessory pathway with a serpiginous course.

Do all short A-V Mahaim fibers need to undertake catheter ablation therapy?

Only one patient in group A had a short A-V Mahaim incorporated in a reentrant tachycardia circuit as compared with 29 from 33 patients with atriofascicular pathways ($p = 0.09$). In addition, 2 patients have a very long effective refractory

period. Those patients without tachycardia and with a very long effective refractory period probably do not need an ablative therapy.

Previous studies: There are very few data on “short A-V Mahaim fibers” from the literature. Heald et al⁹ reported 4 cases with short A-V Mahaim fibers in a series of 21 patients with decrementally conducting accessory pathways. All 4 patients showed overt preexcitation and were ablated at the tricuspid annulus. An “M” potential was found in 2 of the 4 patients and they were not challenged with adenosine. Twelve of his 21 patients had automaticity during ablation (they call it “stuttering block”), but there is no mention whether the short A-V Mahaim fibers also showed radiofrequency induced automaticity. Haissaguerre et al¹⁰ also reported a series with 21 patients with Mahaim fibers and 4 of them were classified as short A-V Mahaim fibers, all of whom were preexcited during sinus rhythm, as well. Two of them (50%) were tested with adenosine and did not respond. They did not mention the occurrence of Mahaim automaticity during catheter ablation. Cappato et al¹⁷ had 1 short A-V Mahaim in their series of 11 patients. He found an accessory pathway potential but there is no information regarding adenosine test or automaticity during ablation.

Limitations of the study

There are some limiting factors to our study as the small number of patients challenged with adenosine. Our assumption of defining the group A patients as having a “short A-V Mahaim fiber” is not based in any histologic data. However, there is sound electrophysiologic evidence derived from local ventricular activation mapping that supports our working definition.

CONCLUSION

Short decrementally conducting right sided accessory pathways show a typical ECG pattern different from atriofascicular pathways. However, their electrophysiologic properties may not be uniform. These pathways can be successfully interrupted by catheter ablation.

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Chapter 13

Summary and Conclusions

In **Chapter 1** we present the history of the most important published data on the issue of variants of pre-excitation, highlighting diagnostic aspects and therapy.

According to Webster's New Dictionary the term variant means a manifesting variety, something varying "slightly" from the standard form. The so-called Mahaim fibers are connecting structures with a great variability in their proximal and/ or distal insertions, but sharing a common electrophysiological behavior suggestive of an AV nodal-like structure. The term variant is more appropriately applied for their electrophysiologic profile than their anatomic structure. The main source of the conceptual mismatch between anatomy and function of those structures derives from the fact that Ivan Mahaim originally described connections between the A-V node and bundle branches or ventricular myocardium, which bypassed the His bundle, in pathologic specimens. When other authors observed accessory bypass tracts with long and decremental conduction, they assumed that the substrate was the fibers described by Mahaim.

Two of the original structures described by Mahaim, the nodoventricular and nodofascicular fibers are rare items as shown by rare case reports. The majority of patients with anterogradely decrementally conducting accessory pathways are today classified as having either an atriofascicular pathway or a long atrioventricular pathway with decremental properties. Those patients usually show a pre-excited left bundle branch block shaped QRS during tachycardia.

Our aim in **Chapter 2** was to describe the differential diagnosis of a left bundle branch block tachycardia with emphasis on the electrocardiographic features which can be helpful for the diagnosis of an SVT with LBBB, a preexcited tachycardia due to a rapidly conducting right-sided bypass tract, a preexcited tachycardia due to a slow conducting right-sided accessory pathway, and a ventricular tachycardia. We also discussed ECG characteristics suggesting specific mechanisms of ventricular tachycardia like ischemic scar VT, bundle branch and interfascicular reentrant VT, right-sided idiopathic VT and VT associated with right ventricular dysplasia.

The electrocardiogram of patients with atriofascicular pathways are usually described as normal. A minimally pre-excited ECG is also recognized by some authors in spite

of the lack of any specific feature for a Mahaim fiber. We analyzed a large series of patients with Mahaim fibers and in **Chapter 3** where we describe our observations regarding the ECG characteristics in sinus rhythm and during tachycardia in a cohort of 40 patients. The manuscript focuses on 33 patients, because for the sake of clarity we have excluded 5 patients with associated rapidly conducting bypass tracts and 2 patients with short atrioventricular pathways with long and decremental conduction. We did observe a previously unreported finding of an **rS** or **rSR'** pattern in lead III, usually associated with the absence of septal **q** wave in the left-sided leads in a significant 70% of the patients. An **rS** pattern in lead III also emerged in 4 out of the 5 patients with WPW and Mahaim after successful ablation of their rapidly conducting accessory pathways. There was a positive relation between the presence of this pattern and left axis deviation during pre-excited tachycardia. This fact together with the observation that after successful ablation of the Mahaim fiber a different QRS morphology emerged in lead III (usually a **qR** or **QR**) validated the concept that the **rS** pattern represented an electrocardiographic clue as to the presence of minimal right ventricular pre-excitation.

Upon our experience with this large series of Mahaim patients we described in **Chapter 4**, in a systematic way, the different tachyarrhythmias associated with the Mahaim fibers. Every variety was described and some clues for their ECG recognition are presented.

In **Chapter 5** we have tested the sensitivity, specificity and predictive value of previously published electrocardiographic criteria for diagnosing Mahaim tachycardia in 40 patients. The predictive value was evaluated by having a subset of 35 patients with LBBB SVT due to orthodromic A-V tachycardia, A-V node reentrant tachycardia and atrial tachycardia. The population with the lowest predictive value was the one having LBBB SVT during circus movement tachycardia with V-A conduction over an accessory A-V pathway with a 26% (5 of 19 patients) false positive diagnosis for Mahaim tachycardia (negative predictive value of 82,5%). Previously reported criteria were sensitive only for atriofascicular pathway (87,5%) but not for decrementally conducting atrioventricular accessory pathway (0%) ($p < 0,0001$).

We analyzed in **Chapter 6** the electrocardiogram during sinus rhythm of 7 patients with a fasciculoventricular pathway and compared them with the ECG of 20 patients with an antero-septal accessory pathway and with 20 patients with a mid-septal accessory pathway. The ECG of a fasciculoventricular pathway showed a narrower (0.12 ± 0.02) QRS complex as compared with other septally located bypass tracts (0.14 ± 0.01) ($p < 0.0001$). According to our findings fasciculoventricular pathways have overlapping electrocardiographic features with both antero-septal and mid-septal accessory bypass tracts, therefore they can not be differentiated by any criteria from the antero-septal nor the mid-septal bypass tracts when a larger QRS complex is present, calling for an electrophysiologic study for a correct diagnosis.

In **Chapter 7** we report for the first time the engagement of a Mahaim fiber (atriofascicular pathway) in a non-reentrant tachycardia with a 1:2 P to QRS relationship. The presence of two connections between the atrium and the ventricle is the usual substrate underlying a tachycardia with 1:2 P to QRS relationship. In this particular case there was only one pathway involved. The pathophysiology was

related to longitudinal dissociation of the impulse conduction within the Mahaim fiber. Duality of conduction over Mahaim fibers due to longitudinal dissociation was reported as an explanation to cycle length alternans during antidromic tachycardia, but it was never before linked to 1:2 conduction.

The fasciculoventricular pathways are extensively discussed in **Chapter 8**. In spite of the lack of a role in reentrant arrhythmias, we emphasized the importance of a correct differential diagnosis between such pathways and a right anteroseptal parahissian bypass tract to avoid damage to the A-V node if an ablation attempt is mistakenly carried out. This possibility is increased by the high incidence of associated rapidly conducting bypass tract along with the fasciculoventricular pathways (we have reported a patient with 2 associated bypass tracts). The electrocardiographic and electrophysiologic characteristics are presented and discussed, as well as a review of the limited literature on the subject.

We did appraise the role of radiofrequency induced automaticity over Mahaim fibers in **Chapter 9**. The phenomenon had been widely reported but not systematically studied. We described the ECG characteristics of what we have called Mahaim automatic tachycardia (MAT), its relation to the thermal injury to the proximal part of the Mahaim fiber, its resemblance with the automatic junctional tachycardia occurring during the slow AV nodal pathway ablation, their importance as a hallmark of successful ablation and we suggest for the first time that in some patients it might be necessary to continue the delivery of radiofrequency current until all automaticity arising in decremental pathway is completely terminated.

Chapter 10 is a case study describing our findings in a patient with a Mahaim fiber who underwent radiofrequency catheter ablation at its distal insertion site. Our findings were consistent with a long atrioventricular Mahaim fiber inserting in the musculature of the anteroapical area of the right ventricle and not at the right bundle branch as it would be the case of an atriofascicular pathway.

Our aim in **Chapter 11** was to describe the occurrence of spontaneous automaticity arising in Mahaim fibers, which included different phenomena ranging from heat induced automaticity occurring during catheter ablation, spontaneous asymptomatic slow automatic rhythms, spontaneous automatic rhythms that could trigger a pre-excited tachycardia under enhanced adrenergic tonus and spontaneous symptomatic fast automatic rhythms presenting as repetitive tachycardia with an incessant character. We discussed those automatic arrhythmias making an analogy with similar spontaneous automatic rhythms arising in the A-V node. We conclude that these findings are consistent with the A-V node like structure of the Mahaim fiber.

In **Chapter 12** we analyzed a group of 8 patients with a short A-V Mahaim fiber and compared their clinical, electrocardiographic and electrophysiologic characteristics with a group of 33 patients with an atriofascicular pathway or a long decrementally conducting atrioventricular pathway. Patients with a long Mahaim fiber showed a very homogeneous electrophysiologic profile. A different situation was found in patients with a short A-V Mahaim fibers. They could be classified into 2 different groups: one group comprised of patients with fibers having A-V node like properties and another group without those properties.

GENERAL CONCLUSIONS

1. The so-called "Mahaim variants" bypass tracts are rare structures with different anatomic substrates sharing the common electrophysiologic feature of unidirectional slow and decremental anterograde conduction with a variable pathophysiology: because of intrinsic A-V node like properties of the fiber, ordinary muscular fibers showing slow conduction due to anisotropic conduction or because of scar due to previous ablation attempts, or because of a proximal connection to the A-V node or His-Purkinje system. As previously reported, we present additional evidence suggesting that some Mahaim fibers do not insert into the right bundle branch.
2. They can occur as a single pathway or be associated with rapidly conducting accessory pathways. They are involved in many different cardiac arrhythmias, sometimes with complex circuits. A non-reentrant tachycardia caused by a 1:2 P to QRS relationship is reported for the first time. Only the fasciculoventricular variety of Mahaim fiber does not participate in reentrant circuits.
3. The ECG of a patient with a short A-V Mahaim fiber is indistinguishable from the ECG of a rapidly conducting right sided accessory pathway, except when it is associated with a normal PR interval (29%). Electrocardiographic recognition of the Mahaim fiber during sinus rhythm is possible in up to 70 % of the patients. Their peculiar minimal pre-excitation (**rS**) pattern is best seen in lead III, and that observation when matched with the absence of a **q** wave in lead I, seems to be very specific of a Mahaim fiber. Tachycardias with anterograde conduction over a Mahaim fiber usually can be suspected by the presence of an **R** wave in lead I, an **rS** pattern in lead VI, QRS frontal plane axis between 0 and -75, QRS transition in the precordial leads after V4 and a QRS width below 0,15 seconds. The negative predictive value of those criteria was 82,5% and the sensitivity 87% for atriofascicular pathways. They are not helpful for identifying tachycardia associated with atrioventricular Mahaim fibers.
4. Spontaneous and radiofrequency induced automaticity arising in atriofascicular pathways as well as with short and long decrementally conducting atrioventricular pathways are characteristic features pointing to an accessory A-V node like structure.

Sumário e Conclusões

No **Capítulo 1** apresentamos um histórico da evolução do conhecimento acumulado nas últimas 3 décadas na literatura, após análise dos trabalhos mais relevantes sobre variantes de pré-excitação, enfatizando os aspectos diagnósticos e terapêuticos.

De acordo com o dicionário Webster o termo variante significa uma manifestação de variedade, algo que difere levemente da forma habitual. As fibras de Mahaim são estruturas de conexão com uma grande variação nas suas inserções proximais e distais, mas tendo em comum um padrão de resposta eletrofisiológica que sugere fortemente uma estrutura semelhante ao nó atrioventricular (A-V). O termo variante é apropriado a estas estruturas mais pelo seu perfil eletrofisiológico, pois, do que pela sua anatomia. A fonte principal para esta dicotomia entre anatomia e função destas estruturas advém do fato de que elas foram originalmente descritas por Ivan Mahaim como conexões entre o nó A-V e seus ramos e a musculatura do septo interventricular, servindo de ponte entre estas estruturas, passando ao largo do feixe de His, conforme observação em análise anatomo-patológica. Muitas décadas após sua descrição original, quando outros autores observaram vias acessórias atrioventriculares com condução lenta e propriedade decremental, eles fizeram a suposição de que o substrato para sua observação seria as fibras descritas por Mahaim.

Duas das estruturas originalmente descritas por Mahaim, as fibras nodoventriculares e as nodofasciculares, são raras e apenas objeto de relatos esporádicos. A maioria dos pacientes com vias acessórias com condução anterógrada decremental são hoje em dia classificadas como feixes atriofasciculares ou vias acessórias atrioventriculares longas com propriedade decremental. Estes pacientes geralmente apresentam uma taquicardia pré-excitada com QRS com configuração tipo bloqueio de ramo esquerdo. Nosso objetivo no **Capítulo 2** foi discorrer sobre o diagnóstico diferencial de uma pacientes com vias acessórias com condução anterógrada decremental são hoje em dia classificadas como feixes atriofasciculares ou vias acessórias atrioventriculares longas com propriedade decremental. Estes pacientes geralmente apresentam uma taquicardia pré-excitada com QRS com configuração tipo bloqueio de ramo esquerdo. Nosso objetivo no **Capítulo 2** foi discorrer sobre o diagnóstico diferencial de uma taquicardia com QRS alargado e padrão tipo bloqueio do ramo esquerdo (BRE) enfatizando aspectos eletrocardiográficos potencialmente úteis no reconhecimento de arritmias tais como: uma taquicardia supraventricular com aberrância tipo BRE, uma taquicardia pré-excitada devido a uma via acessória com condução rápida localizada no anel tricúspide, uma taquicardia pré-excitada devido a uma via acessória à direita com condução lenta e uma taquicardia ventricular. Nós também discutimos características eletrocardiográficas sugestivas de mecanismos específicos de taquicardia ventricular como aquelas decorrentes de uma cicatriz miocárdica pós isquêmica, reentrada ramo-a-ramo e reentrada interfascicular, taquicardia ventricular

direita idiopática e taquicardia ventricular associada à displasia ventricular arritmogênica.

O eletrocardiograma de pacientes com feixes atriofasciculares é habitualmente descrito como normal. Um padrão de preexcitação mínima já foi reconhecido por alguns autores, que, no entanto não descreveram qualquer sinal eletrocardiográfico específico de uma fibra de Mahaim. Nós analisamos um coorte de 40 pacientes com fibras de Mahaim, descrevendo no **Capítulo 3** nossas observações à cerca das características eletrocardiográficas durante ritmo sinusal e taquicardia pré-excitada. Este trabalho se detem em 33 pacientes, pois, foram excluídos 5 pacientes com vias acessórias manifestas (múltiplas vias) além de 2 pacientes com feixes atrioventriculares decrementais curtos. Nós observamos em 70% dos pacientes durante ritmo sinusal a presença de um achado eletrocardiográfico na derivação III consistindo de um complexo QRS com configuração **rS** ou **rSR'**, geralmente associado com ausência de onda **q** septal nas derivações esquerdas. Não há registro prévio deste achado na literatura especializada. Este padrão **rS** também foi observado em 4 dos 5 pacientes com síndrome de WPW associado à fibra de Mahaim, após a ablação da via acessória com condução rápida. Houve uma correlação direta entre a presença deste padrão em III e desvio do eixo para a esquerda durante taquicardia preexcitada. Este fato aliado à observação de que havia uma nítida modificação do complexo QRS na derivação III após a ablação da fibra de Mahaim (geralmente surgia um complexo tipo **qR** ou **QR**), validou o conceito de que a configuração representava um sinal eletrocardiográfico compatível com uma pré-excitação mínima da ativação do ventrículo direito.

Em decorrência de nossa experiência apreendida com a análise desta extensa série de pacientes com fibras de Mahaim, nós descrevemos de forma sistemática no **Capítulo 4**, as diferentes taquiarritmias que podem ocorrer em um paciente portador de fibra de Mahaim. Descrevemos todas as variedades bem como sinais eletrocardiográficos que possam sugerir seu diagnóstico.

No **Capítulo 5** testamos a sensibilidade, especificidade e o valor preditivo de critérios eletrocardiográficos previamente descritos para o diagnóstico de uma taquicardia associada à fibra de Mahaim em 40 pacientes. O seu valor preditivo foi avaliado em um grupo de 35 pacientes com taquicardia supraventricular com morfologia de BRE causadas por taquicardia A-V ortodrômica, taquicardia por reentrada nodal e taquicardia atrial. O menor valor preditivo foi observado nos pacientes com taquicardia por movimento circular utilizando uma via acessória oculta. Neste grupo observamos uma taxa de 26% de diagnósticos falso positivos (5 de 19 pacientes), em outras palavras, 26% teriam sido erroneamente diagnosticados como uma taquicardia utilizando uma via decremental (valor preditivo negativo de 82,5%). Os critérios utilizados no trabalho de Bardy et al foram sensíveis apenas para os feixes atriofasciculares (87,5%), não tendo sido úteis na detecção de vias acessórias decrementais atrioventriculares (0%) ($p < 0,0001$).

No **Capítulo 6** nós analisamos o eletrocardiograma em ritmo sinusal de 7 pacientes com feixe fascículoventricular e os comparamos com o ECG de 20 pacientes com via acessória anteroseptal e com 20 pacientes com via acessória medioseptal. No ECG de pacientes com um feixe fascículoventricular o complexo QRS era mais estreito (0.12 ± 0.02) que o QRS de pacientes com vias acessórias septais (0.14 ± 0.01)

($p < 0,0001$). Nossos achados mostram uma superposição dos achados ao ECG entre os pacientes analisados, e num paciente com feixe fasciculoventricular com QRS $> 0,12$ segundos não foi possível discriminá-lo de pacientes com vias acessórias septais, o que sugere a necessidade de se realizar estudo eletrofisiológico para um diagnóstico correto.

No **Capítulo 7** nós relatamos pela primeira vez a utilização de uma fibra de Mahaim (feixe atriofascicular) numa taquicardia não reentrante com uma relação P/ QRS de 1:2. O substrato usual para uma taquicardia com relação P:QRS de 1:2 é a presença de duas conexões entre os átrios e os ventrículos. Neste caso específico havia apenas uma conexão envolvida. A fisiopatologia estava relacionada com uma dissociação longitudinal da condução do impulso dentro da fibra de Mahaim. Condução dual por uma fibra de Mahaim devido à dissociação longitudinal já foi objeto de relato para se justificar a presença de alternância do ciclo de uma taquicardia antidrômica, mas nunca anteriormente relacionada com uma condução 1:2.

As vias fasciculoventriculares são amplamente discutidas no **Capítulo 8**. A despeito da não participação ativa em circuitos reentrantes, nós enfatizamos a importância de um correto diagnóstico diferencial entre estas vias em uma via acessória anteroseptal parahissiana, e desta forma evitar dano ao nó A-V, no caso de uma tentativa equivocada de se realizar uma ablação neste tipo de conexão anômala. A possibilidade de erro diagnóstico é aumentada pelo fato de que existe uma alta incidência de vias acessórias atrioventriculares associadas aos feixes fasciculoventriculares (nós publicamos um caso com 2 vias anômalas associadas). Descrevemos as características eletrocardiográficas e eletrofisiológicas e apresentamos uma revisão da limitada literatura sobre o assunto.

Nós avaliamos o significado do automatismo induzido pela ablação por radiofrequência das fibras de Mahaim no **Capítulo 9**. Este fenômeno tem sido relatado mas não estudado de forma sistematizada. Nós descrevemos as características eletrocardiográficas do que denominamos de “Taquicardia automática de Mahaim” (MAT), sua relação com o dano termo-induzido da porção proximal da fibra de Mahaim, sua semelhança com a taquicardia juncional automática que ocorre durante a ablação da via nodal lenta, sua importância como um marcador de sucesso do procedimento ablativo, e sugerimos pela primeira vez, que em alguns pacientes pode ser necessário continuar a aplicação de radiofrequência até o término de toda a atividade automática originária da via decremental, para a obtenção de resultado definitivo.

O **Capítulo 10** é um relato de caso onde descrevemos nossos achados num paciente portador de uma fibra de Mahaim, submetido a ablação por cateter com radiofrequência na inserção distal da via. Nossos achados foram consistentes com uma fibra de Mahaim longa do tipo atrioventricular, se inserindo na musculatura da região antero-apical do ventrículo direito e não na porção distal do ramo direito do feixe de His, como seria o caso de um feixe atriofascicular.

Nosso objetivo no **Capítulo 11** foi de descrever a ocorrência de automatismo espontâneo nas fibras de Mahaim, o que incluiu fenômenos diversos, desde automatismo termo-induzido durante a ablação por radiofrequência, ritmos automáticos lentos assintomáticos, ritmos automáticos espontâneos que trigam uma taquicardia antidrômica sob a influência de um tônus adrenérgico aumentado e ritmos

automáticos espontâneos rápidos se apresentando como taquicardia repetitiva com carácter incessante. Nós discutimos estas arritmias automáticas traçando uma analogia com ritmos automáticos nodais. Baseados nestes achados, nós concluímos que as fibras de Mahaim tem uma estrutura semelhante à do nó A-V.

No **Capítulo 12** nós estudamos as fibras curtas de Mahaim, denominação aplicada a vias acessórias com condução lenta e decremental com inserção ventricular just-anulares. As características clínicas de 8 pacientes com fibras curtas de Mahaim bem como seu perfil eletrocardiográfico e eletrofisiológico foram comparados com um grupo de 33 pacientes com feixes atriofasciculares ou atrioventriculares longos. Além das importantes diferenças entre os 2 grupos, observamos que os pacientes com fibras curtas não são um grupo homogêneo. Pode-se classificá-los em 2 grupos: um onde os pacientes apresentam características de um nó A-V acessório e outro onde estas características estão ausentes.

CONCLUSÕES GERAIS

1. As variantes de pré-excitação “fibras de Mahaim” são estruturas raras com substratos anatômicos diferentes tendo como denominador comum uma característica eletrofisiológica: condução unidirecional anterógrada lenta e decremental do estímulo elétrico, seja em decorrência de propriedades intrínsecas da fibra, compatíveis com um nó A-V acessório, fibras musculares ordinárias com condução lenta por fibrose secundária a ablação prévia ou por características anômicas especiais, ou ainda devido à conexão proximal desta fibra ao nó A-V ou sistema His-Purkinje. Em trabalhos já publicados, nós apresentamos evidências adicionais sugerindo que algumas destas fibras de Mahaim não se inserem no ramo direito do feixe de His, além das fibras curtas que se inserem no anulo valvar tricúspide.
2. Elas podem se manifestar como uma via acessória isolada ou estar associada com vias acessórias com condução rápida. Elas podem estar envolvidas em uma grande variedade de arritmias distintas, algumas vezes com circuitos complexos. Descrevemos pela primeira vez uma variedade de taquicardia não reentrante causada por uma condução 1:2 do estímulo sinusal. Apenas a variedade fasciculoventricular das fibras de Mahaim não participa ativamente em um circuito arritmogênico.
3. As fibras de Mahaim curtas habitualmente se manifestam com um ECG indistinguível de uma via acessória de condução rápida, exceto quando co-existe um intervalo PR normal (29%). O diagnóstico eletrocardiográfico de uma fibra de Mahaim longa durante ritmo sinusal é possível em até 70% dos pacientes. Seu peculiar padrão de pré-excitação mínima (rS) é melhor observado na derivação III, e este padrão de QRS quando se associa à ausência de onda q na derivação I, parece ser bastante específico para uma fibra de Mahaim. Pode-se suspeitar de uma taquicardia com condução anterógrada por uma fibra de Mahaim pela associação dos seguintes critérios ao ECG: onda R em I, um padrão rS em

V1, eixo do QRS no plano frontal entre 0 e -75, transição do QRS ($R/S < 1$) além de V4 e um QRS com largura menor que 0,15 segundos. O valor preditivo negativo destes critérios foi de 82,5% e a sensibilidade para o diagnóstico de feixes atriofasciculares foi de 87%. Eles não foram úteis para identificar uma taquicardia associada com fibras de Mahaim tipo atrioventriculares.

4. Automatismo espontâneo e induzido durante ablação por radiofrequência originado em um feixe atriofascicular ou em um feixe atrioventricular curto ou longo com condução decremental são características que sugerem que estas fibras são estruturas compatíveis com um nó A-V acessório.

Samenvatting en Conclusies

In **hoofdstuk 1** wordt een overzicht gegeven van de belangrijkste publicaties over ongewone vormen (varianten) van ventriculaire pre-excitatie met de nadruk op diagnose en behandeling.

Volgens “Webster’s New Dictionary” wordt met de naam variant aangegeven dat iets een beetje verschilt van de gebruikelijke of standaard vorm. De naam Mahaim bundel geldt voor een groep extra verbindingen die verschillen in hun proximale en distale insertie, maar gemeen hebben dat hun electrofysiologisch gedrag lijkt op dat van de A-V knoop. De naam Mahaim bundel of vezel vertelt ons dus meer over de electrofysiologische eigenschappen dan de anatomische locatie en structuur.

Ivan Mahaim beschreef verbindingen tussen de A-V knoop en de bundeltakken of met het kamermyo-card. Toen andere auteurs extra verbindingen beschreven met trage geleiding, zoals we die kennen van de A-V knoop, gaven zij hier aan ook de naam Mahaim bundel of vezel. Wij weten nu dat de nodoventriculaire en nodofasciculaire verbindingen die Mahaim beschreef zeldzaam zijn. Bij de meeste patiënten met een anterograde geleidende extra verbinding, is dit een verbinding tussen de rechter boezem en de rechter bundel (atriofasciculaire) of er is sprake van een langzaam geleidende verbinding tussen de boezem en de rechterkamerspier (atrioventriculair). Belangrijk is dat wanneer deze patiënten een tachycardie hebben met anterograde geleiding over de Mahaim bundel het QRS complex tijdens de tachycardie een linker bundeltak blok vorm heeft. In **hoofdstuk 2** wordt de differentiaal diagnostiek beschreven van een tachycardie met een linker bundeltak blok vorm. Aangegeven wordt hoe men kan herkennen of het hier een supraventriculaire tachycardia met linker bundeltak blok betreft, een tachycardie met snelle of langzame geleiding over een rechts gelegen extra verbinding, of een kamer tachycardie.

Ook wordt de aandacht gevestigd op ECG bevindingen die bij een linker bundel tak achtige VT wijzen op een ischemische of idiopathische origine, de aanwezigheid van een arrhythmogene rechter kamer dysplasie of een tachycardie die berust op re-entry in het bundeltak systeem.

In **hoofdstuk 3** worden de ECG bevindingen beschreven, zowel bij sinus ritme als tijdens de tachycardie, bij 33 patiënten met een Mahaim bundel. Hierbij wordt aandacht gevraagd voor een niet eerder beschreven bevinding dat tijdens sinus ritme het ECG in afleiding III in 70% van de gevallen een rS of rSR’ patroon laat zien, meestal gecombineerd met een afwezige q in I, AVL en V6. Dit rS patroon in afleiding III werd ook gezien na ablatie van een snel geleidende extra A-V verbinding in 4 van de 5 patiënten die naast een Mahaim bundel een WPW syndroom hadden. Er was een positieve relatie tussen een rS patroon in afleiding III tijdens sinus ritme en linker as deviatie van het QRS complex tijdens een tachycardie met anterograde geleiding over de Mahaim bundel.

Deze bevinding en het feit dat na succesvolle ablatie van de Mahaim bundel een andere QRS configuratie optrad in afleiding III (meestal qR of QR) ondersteunt

de gedachte dat het **rS** patroon in afleiding III electrocardiografisch aangeeft dat er geringe pre-excitatie bestaat van het laag lateraal gelegen deel van de rechter ventrikel.

In **hoofdstuk 4** worden de verschillende soorten tachycardie beschreven die kunnen voorkomen bij een patient met een Mahaim bundel.

Hoofdstuk 5 beschrijft de toetsing van vroeger door Bardy en collega's beschreven ECG criteria voor een Mahaim tachycardie aan het eigen patienten materiaal. Hierbij werden de Mahaim patienten vergeleken met 35 patienten met verschillende soorten supra ventriculaire tachycardie met een linker bundel tak blok. De eerder door Bardy gepubliceerde criteria waren het minst bruikbaar wanneer een linker bundeltak blok aanwezig was bij patienten met een orthodrome cirkel tachycardie met ventriculo-atricle geleiding over een extra A-V verbinding. Bij deze groep patienten werd met behulp van de Bardy criteria in 26% (5 van de 19 patienten) een vals positieve diagnose van een Mahaim vezel gesteld. De Bardy criteria hadden een sensitiviteit van 87,5% voor atrio-fasciculaire bundels, maar waren niet bruikbaar om de aanwezigheid van een langzaam geleidende A-V extra verbinding te diagnosticeren.

In **hoofdstuk 6** wordt het electrocardiogram tijdens sinus ritme beschreven van 7 patienten met een fasciculo ventriculaire extra verbinding. Deze ECG's werden vergeleken met die van 20 patienten met een antero septale en 20 patienten met een mid septale extra verbinding. Het QRS complex van patienten met een fasciculo ventriculaire extra verbinding was minder breed ($0,12 \pm 0,02$ sec.) dan het QRS complex bij de septale AV extra verbindingen ($0,14 \pm 0,01$ sec.) ($p < 0,0001$). Overigens bestaat er "overlap" in de ECG bevindingen tussen fasciculaoventriculaire extra verbindingen en antero septale en mid septale AV verbindingen. Dit laatste maakt dat wanneer er een verbreed QRS complex wordt gevonden, differentiering niet goed mogelijk is en dus een electrophysiologische studie noodzakelijk is om te komen tot een juiste diagnose.

Hoofdstuk 7 beschrijft een patient waarbij een tachycardie aanwezig was tijdens sinus ritme omdat iedere P-top werd gevolgd door 2 QRS complexen. Dit bleek te berusten op de aanwezigheid van longitudinale dissociatie in een Mahaim bundel. De verschillende geleidingstijden in de twee paden in de Mahaim bundel maakte het mogelijk dat een P-top resulteerde in 2 QRS complexen. Dit is een nog niet eerdere beschreven vorm van tachycardie in een patient met een Mahaim bundel.

Hoofdstuk 8 gaat over fasciculo ventriculaire verbindingen. Ofschoon zij geen rol spelen als onderdeel van een tachycardie circuit is correcte identificatie van belang met name voor de

differentiatie met rechts gelegen antero-septale para-hissale extra verbindingen. Een juiste diagnose voorkomt een ablatie poging met groot gevaar voor beschadiging van het A-V geleidings systeem. Een foute diagnose kan gemakkelijk worden gemaakt omdat patienten met fasciculo ventriculaire vezels vaak ook accessoire A-V verbindingen hebben en dus ook tachycardieën. Aandacht wordt dan ook besteed aan de electrocardiografische en en electrofysiologische kenmerken van fasciculo ventriculaire vezels.

Zoals in **hoofdstuk 9** beschreven kan tijdens radiofrequente catheter ablatie van een Mahaim vezel vaak automatische impuls vorming in de Mahaim vezel worden geïnduceerd. De ECG karakteristieken van de Mahaim automatische tachycardie (MAT) worden gepresenteerd en de betekenis ervan besproken. Bij sommige patienten is het belangrijk om de radiofrequente ablatie van de Mahaim vezel zolang voor te zetten tot iedere automaticiteit is uitgedoofd.

In **hoofdstuk 10** wordt een patient besproken waarbij ablatie van de Mahaim bundel plaats vond aan het ventriculaire einde. Het ging hierbij om een lange atrio-ventriculaire Mahaim bundel die insereerde in het apicale deel van de rechter ventrikel en dus niet in de rechter bundel zoals bij de atrio-fasciculaire Mahaim vezel.

Hoofdstuk 11 geeft een overzicht van de verschillende vormen waaronder automaticiteit in de Mahaim vezel zich kan manifesteren. Dit gedrag lijkt sterk op de automaticiteit zoals wij die kennen in de A-V knoop en pleit dus voor een A-V nodale opbouw van de Mahaim vezel.

In **hoofdstuk 12** worden 8 patienten beschreven met een korte A-V Mahaim verbinding. Klinische, electrocardiografische en electrofysiologische bevindingen bij deze patienten werden vergeleken met een groep van 33 patienten met een atrio fasciculaire verbinding of een lange A-V extra verbinding met decrementale geleiding. De 33 patienten hadden een zeer homogeen electrofysiologisch profiel. Dit was niet het geval bij patienten met een korte A-V Mahaim verbinding. Bij hen was er sprake van 2 groepen. Bij de ene groep had de extra verbinding A-V nodale eigenschappen, bij de andere groep was dit niet het geval.

CONCLUSIES

- 1) De groep van Mahaim extra verbindingen bestaat uit verschillende anatomische substraten met als gemeenschappelijke electrophysiologische eigenschap: unidirectionele, langzame en decrementale antegrade geleiding. Dit kan verschillende oorzaken hebben, zoals A-V nodale eigenschappen van de extra verbinding, anisotrope geleiding in spiervezels of een proximale verbinding met de A-V knoop of het His –Purkinje systeem. Het is niet zo dat alle Mahaim extra verbindingen insereren in de rechter bundel van het intraventriculaire geleidingssysteem.
- 2) Mahaim extra verbindingen kunnen afzonderlijk voorkomen, maar worden vaak gevonden samen met snel geleidende extra AV verbindingen. Zij spelen een rol bij verschillende, soms zeer complexe, ritmestoornissen. Zij kunnen ook een tachycardie veroorzaken wanneer tijdens sinus ritme iedere P top gevolgd wordt door 2 QRS complexen door longitudinale dissociatie of duplicering van de Mahaim verbinding. Fasciculo ventriculaire Mahaim verbindingen spelen geen rol in tachycardie circuits.
- 3) Het ECG van de patient met een korte AV Mahaim vezel is niet te onderscheiden van het ECG van een snel geleidende rechts gelegen extra AV verbinding behalve wanneer het P-delta interval normaal is (29%). In 70% van de patienten met een Mahaim extra verbinding is het mogelijk om de juiste diagnose te maken tijdens sinus ritme. Dit berust op de aanwezigheid van een minimaal pre-excitatie (rS) patroon in afleiding III, in het bijzonder wanneer dit gepaard gaat met de afwezigheid van een q in afleiding I. De tachycardieën met anterograde geleiding over een Mahaim extra verbinding hebben meestal een R in afleiding I, een rS patroon in afleiding V₁, een QRS as in het frontale vlak tussen 0 en -75 graden, een QRS R/S transitie in afleiding V₄ en een QRS breedte minder dan 0,15 seconden. De negatief voorspellende waarde van deze criteria was 82,5%

en de sensitiviteit 87% voor de aanwezigheid van atrio fasciculaire extra verbindingen. Zij hadden geen waarde voor het diagnostiseren van tachycardieën met anterograde geleiding over een atrio ventriculaire Mahaim vezel.

- 4) Automaticiteit, zowel spontaan als tijdens radiofrequente catheter ablatie, is een typisch verschijnsel bij Mahaim vezels. Het pleit voor de aanwezigheid van accessoir A-V nodaal weefsel.

Acknowledgements

The bond between me and Professor Wellens teachings goes as far back as 1982. At that time I was a resident in Cardiology, trying to keep the pace between my everyday duties, night calls and the study on books and journals. Soon I became attracted to the study of cardiac arrhythmias. Looking for manuscripts in the leading journals of cardiology I came across many manuscripts of Hein Wellens and his group from Maastricht. I was profoundly impressed by his meticulous work, his unique didacticism specially the critical way he extracted information from the invasive electrophysiologic studies to enhance the diagnostic yield of the electrocardiogram. As time went by it became clear for me that I would like to do a fellowship in Maastricht, and to benefit the most from it I did 1 year period of probation in Brazil. That was accomplished under the guidance of Professor Eduardo Sosa, head of one of the most important electrophysiologic laboratory in Brazil, at Instituto do Coração, São Paulo University. I am very much indebted to Dr Sosa, for his commitment to teaching, research and friendship.

The next step was to get in touch with Wellens, someone I only knew from the books and papers. Dr A. Belfer, a brazilian nuclear medicine specialist told me to contact Dr Frans Wackers, from Yale University, a close friend of Wellens. I wrote him a letter about my plans and his advise was crystal clear: write straight to Wellens! Needless to say he immediately accepted me as a fellow, at my convenience. After a hard and long process I was granted a scholarship from CNPQ (Brazilian Council for Research) which enabled me to go to the Netherlands.

During my stay in Maastricht, I had the good fortune to attend the EP laboratory under the supervision of Dr Pedro Brugada and Dr Joep Smeets. Every ECG was a quiz session, and new observations on the tracings an inspiration for reports! Professor Wellens was always available for tête-à-tête discussions on those difficult tracings that could resist so many brains eager to decipher them! I would like to recall the morning meetings sessions. I have greatly benefit from the discussions and opinions of the staff members of the *Afdeling Cardiologie* as well as from informal discussions with the senior fellows at the "Provisorium" outbuilding in St Annadal Hospital, Maastricht, particularly from Mario Talajic, Roman Lezaun, Pelayo Torner, Hans Joachim Trappe, and Luz-Maria Rodriguez.

After conclusion of my fellowship I kept in touch with the department. In 1993, I came back to Maastricht for a 2 month period of training in catheter ablation techniques. I also attended the Congress "2000, *Future of Arrhythmology: Lessons from the Past, Promises for Tomorrow*", a tribute to the scientific legacy of Professor Wellens.

In december of 2002 I went to Maastricht to attend some cryoablation procedures upon the invitation by Luz-Maria Rodriguez, a friend and ex-colleague from the fellowship program time. After reading some publications I did on variants of pre-excitation she had motivated me to go on working on the subject. Professor Wellens opened the door again for me, and he would eventually become my promotor. Dr Carl Timmermans, a staff member of the Cardiology department kindly offered me the data he had collected from Mahaim patients over the years. I also am very grateful to Dr Ashish Nabar with his assistance with the dutch language on the analysis of the patient's records as well as the thoughtful discussions about the cases from Maastricht. I soon realized that I was in need to increase the number of patients in the Mahaim database to enhance the power of my observations. I am indebted to Dr Eduardo Sosa, Instituto do Coração, São Paulo University, Brazil and Dr Fernando E. Cruz Filho and Márcio Fagundes from Instituto de Cardiologia Laranjeiras, Rio de Janeiro, Brazil, for allowing me to include their patients in my study. Dr Cruz was also a previous fellow in Maastricht. We spent many hours discussing issues related to mechanisms of cardiac arrhythmias.

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Dear Hein: Thank you for opening so many real and virtual doors and windows.

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Curriculum Vitae

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