CHAPTER 1

Historical notes and classification of the variants of ventricular preexcitation

Almost 25 years ago, Gallagher *et al*. [1] 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 of these fibers was made by Ivan Mahaim in the late 1930s [2, 3] (Fig. 1.1).

He found a conducting tissue extending from the atrioventricular (AV) node to the ventricular myocardium. There are only a few articles dealing with anatomical studies of Mahaim fibers [3–8]. Early investigators found that Mahaim fibers were accessory connections taking off from the bundle of His and the fascicles (fasciculoventricular [FV] fibers) into the right ventricle or from the AV node (nodoventricular [NV] fibers) to the right ventricle or to the right bundle branch (nodofascicular [NF] fibers). Anderson *et al*. [9] proposed two 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 NV concept was consistent with the findings published in the Gallagher series [1] series, wherein some patients had ventriculoatrial (VA) block during wide, complex tachycardia, proving that in those patients the atrium was not part of the reentrant circuit. Wellens [10] was the first to report the electrophysiological findings by using the technique of programmed electrical stimulation in a patient with an accessory pathway with decremental properties and long conduction times and assuming the pathway's relationship with the fibers described long ago by Mahaim. The term *nodofascicular* was used when the retrograde right bundle branch potential preceded the ventricular deflection, whereas the pathway was assumed to be *nodoventricular* when the retrograde His bundle deflection followed the beginning of the ventricular potential. The next step was the understanding of the functional significance and the anatomical–electrophysiological relationship of such pathways. An important observation was made in 1978 by Becker *et al*. [7] when they described an accessory AV node associated with a bundle of specialized fibers measuring 1 cm, coursing through the right ventricle, and mimicking a second AV conduction system located on the lateral tricuspid annulus. However, this finding did not change the mainstream concept of NV fibers at that time.

During the early 1980s, cardiologists started to refer patients with drug refractory tachycardias due to Mahaim fibers for surgical treatment. Although Gillette *et al*. [11] reported as early as 1982 a Mahaim fiber located on the anterior

Figure 1.1 Ivan Mahaim (1897–1965). Born in Liège, Belgium, and educated in Lausanne, Switzerland, he was a fellow of Prof. Wenckebach in Vienna (1926). Mahaim wrote more than 100 papers, which were published in the leading journals of his time. His most influential works were his books on histological research concerning the connections of the bundle of His. This research was a resounding success in Europe in 1937 because it provided the basis for later electrophysiological discoveries. His last work, still a reference for musicians, was devoted to Beethoven's last quartets and published in 1964.

portion of the tricuspid ring, according to the concepts at that time, ablation of the AV node was considered to be the logical 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 AV node to treat a patient with a Mahaim fiber. This technique resulted in complete AV block but persistent ventricular preexcitation [12]. The turning point came in 1988, when Klein *et al*. [13] decided to extensively freeze the AV node and the upper His bundle region of a 29-year-old man and discovered that preexcitation did not go away. This finding indicated to them that the accessory pathway was not connected with the AV node. In another patient the AV node was not damaged, but the decrementally conducting accessory pathway was 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 al*. [14] published a paper titled "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. They showed that it was possible to advance ventricular depolarization during preexcited tachycardia by delivering late atrial premature beats during AV node refractoriness. In the following years, catheter ablation techniques shed more light on the subject. Discrete high-frequency potentials resembling the His bundle potential, considered the electrical activation of the atriofascicular pathway, were used as an effective target for ablation [(15, 16]. Observations during pharmacological interventions

[17], histological data [7, 8, 18, 19], electrophysiological maneuvers, and findings during radiofrequency catheter ablation, such as heat-induced automaticity while ablating at the atrial aspect of the annulus [22, 23]and also spontaneous automaticity [24], were presented as further evidence that the Mahaim fiber was composed of AV node-like tissue. Some authors [20] believed that an accessory AV node without a direct connection to the ventricle could 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. NV/NF fibers are now considered a rare finding. Hluchy *et al*. [25]reported their presence in some patients with a narrow and regular QRS tachycardia with AV dissociation.

Recent reports using noncontact technologies for intracardiac mapping of atriofascicular pathways, such as the EnSite [26] and the LocaLisa system [27], have validated old data derived from open heart epicardial mapping and intracardiac catheter mapping [11, 13], suggesting that most of the decrementally conducting 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. There are a few reports of left-sided decrementally conducting accessory pathways, mostly decrementally conducting AV pathways connecting the left atrium to the left ventricle. Their distal end is usually mapped to the mitral annulus [28–30].

FV pathways [31] are anatomically different from atriofascicular pathways. They do not have long conduction times or decremental properties. Since they are infra-AV nodal structures connected to the His bundle or its fascicles, only the AV node shows decremental conduction. FV pathways play no role in clinical tachycardias. However, because its preexcitation pattern on the 12-lead electrocardiogram may resemble that of an anteroseptal accessory pathway [32], which is often associated with rapidly conducting bypass tracts, a misdiagnosis of a bypass tract should be avoided to prevent unnecessary damage to the AV node–His bundle conduction system by catheter ablation.

Critical analysis of the classification of preexcitation variants

The preexcitation syndromes were originally classified on the basis of their anatomical location and course and named according to the original investigators. This classification resulted in Mahaim, James, and Kent fibers. Later, such a description was considered inadequate because it did not fit the new electrophysiological or anatomical knowledge. In 1975 the European Study Group for Preexcitation [9] introduced a new classification based on the anatomical connections of the accessory pathways. However, the eponym Mahaim survived the changes proposed by the group. Why? It seems that the major issue here is the common electrophysiological finding of long and decremental conduction properties (AV node-like behavior) of such pathways, the so-called Mahaim physiology. We also need to emphasize that the eponym Mahaim

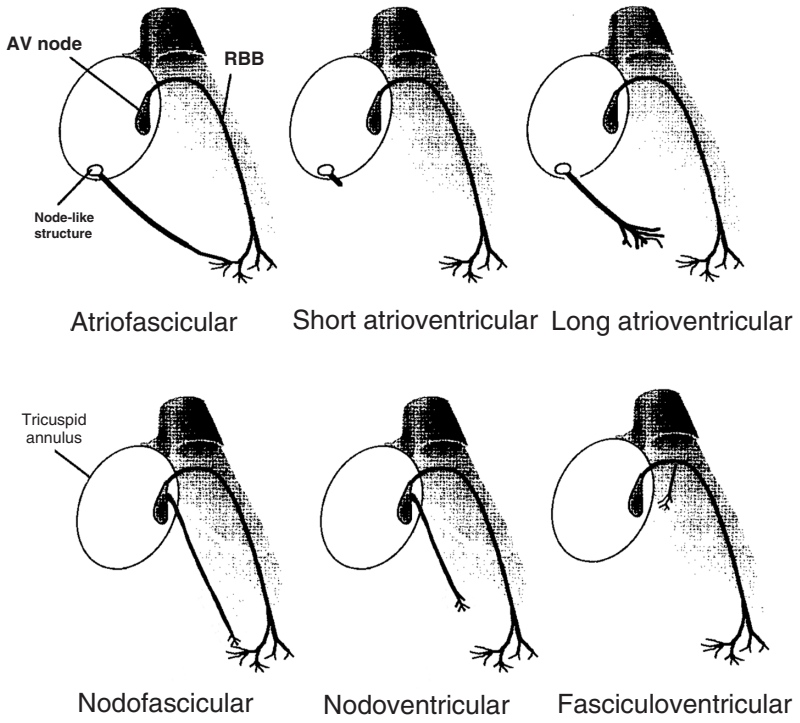

Figure 1.2 The different anatomical courses of preexcitation variants.

is already known to generations of arrhythmologists who witnessed the long and exciting process that took place until full understanding of the anatomical-functional relationship became possible. An updated anatomical classification of the preexcitation variants follows and is illustrated in Fig. 1.2.

Proximal insertion in the AV node–His bundle branch system:

1 NF bypass tract;
2 NV bypass tract;
3 FV pathway.

Proximal insertion in the atrium (right and left atrium):

1 Atriofascicular pathway;
2 short AV pathway with prolonged and decremental conduction;
3 long AV pathway with prolonged and decremental conduction.

References

1 Gallagher JJ, Smith WM, Kassell JH, *et al*. Role of Mahaim fibers in cardiac arrhythmias in man. *Circulation* 1981;64:176.
2 Luderitz B. Ivan Mahaim. *J Int Card Electrophysiol* 2003;8:155.
3 Mahaim I, Benatt A. Nouvelles recherches sur les connexions supérieures de la branche gauche du faisceau de His-Tawara avec cloison interventriculaire. *Cardiologia* 1938;1:61.

4 Mahaim I, Winston MR. Recherches d'anatomie comparée et de pathologie expérimentale sur le connexions hautes du faisceau de His-Tawara. *Cardiologia* 1941;5:189.
5 Lev M, Gibson S, Miller RA. Ebstein's disease with Wolff–Parkinson–White syndrome. *Am Heart J* 1955;49:724.
6 Lev M, Sodi-Palhares D, Friedlan C. A histopathologic study of the atrioventricular communications in a case of WPW with incomplete left bundle branch block. *Am Heart J* 1963;66:399.
7 Becker AE, Anderson RH, Durrer D, Wellens HJJ. The anatomical substrates of Wolff-Parkinson-White syndrome: a clinico-pathologic correlation in seven patients. *Circulation* 1978;57:870.
8 Anderson RH, Becker AE. Stanley Kent and accessory atrioventricular connections. *J Thoracic Cardiovasc Surg* 1981;81:649.
9 Anderson RH, Becker AE, Brechenmacher C, *et al*. Ventricular preexcitation. A proposed nomenclature for its substrates. *Eur J Cardiol* 1975;3:27.
10 Wellens HJJ. *Electrical Stimulation of the Heart in the Study and Treatment of Tachycardias*. Baltimore: University Park Press; 1971:97.
11 Gillette PC, Garson A, Cooley DA, *et al*. Prolonged and decremental anterograde conduction properties in right anterior accessory connections: wide QRS antidromic tachycardia of left bundle branch block pattern without Wolff–Parkinson–White configuration in sinus rhythm. *Am Heart J* 1982;103:66.
12 Bhandari A, Morady F, Shen EN, *et al*. Catheter-induced His bundle ablation in a patient with reentrant tachycardia associated with a nodoventricular tract. *J Am Coll Cardiol* 1984;4:611.
13 Klein GJ, Guiraudon GM, Kerr CR, *et al*. "Nodoventricular" accessory pathway: evidence for a distinct accessory atrioventricular pathway with atrioventricular node-like properties. *J Am Coll Cardiol* 1988;11:1035.
14 Tchou P, Lehmann MH, Jazayeri M, Akhtar M. Atriofascicular connection or a nodoventricular Mahaim fiber? Electrophysiologic elucidation of the pathway and associated reentrant circuit. *Circulation* 1988;77:837.
15 Brugada J, Martinez-Sanches J, Kuzmicic B, *et al*. Radiofrequency catheter ablation of atriofascicular accessory pathways guided by discrete electrical potentials recorded at the tricuspid annulus. *PACE* 1995;18:1388.
16 McClelland JH, Wang X, Beckman KJ, *et al*. Radiofrequency catheter ablation of right atriofascicular (Mahaim) accessory pathways guided by accessory pathway activation potentials. *Circulation* 1994;89:2655.
17 Ellenbogen KA, Rogers R, Old W. Pharmacological characterization of conduction over a Mahaim fiber: evidence for adenosine sensitive conduction. *Pacing Clin Electrophysiol* 1989;12:1396.
18 Guiraudon CM, Guiraudon GM, Klein GJ. Histologic evidence for an accessory atrioventricular pathway with AV-node-like morphology. *Circulation* 1988;78(suppl 2): 40.
19 Epstein MR, Saul JP, Weindling SN, *et al*. Atrioventricular reciprocating tachycardia involving twin atrioventricular nodes in patients with complex congenital heart disease. *J Cardiovasc Electrophysiol* 2001;12:671.
20 Nogami A, Suguta M, Tomita T, *et al*. Novel form of atrial tachycardia originating at the atrioventricular annulus. *PACE* 1998;21:2691.
21 Gollob MB, Bharati S, Swerdlow CD. Accessory atrioventricular node with properties of a typical accessory pathway: anatomic-electrophysiologic correlation. *J Cardiovasc Electrophysiol* 2000;11:922.

22 Braun E, Siebbels J, Volkmer M, *et al*. Radiofrequency-induced preexcited automatic rhythm during ablation of accessory pathways with Mahaim-type preexcitation: does it predicts clinical outcome? *PACE* 1997;20(4, part 2):1121.
23 Sternick EB, Gerken LM, Vrandecic MO. Appraisal of "Mahaim" automatic tachycardia. *J Cardiovasc Electrophysiol* 2002;13:244.
24 Sternick EB, Timmermans C, Sosa E, *et al*. Automaticity in Mahaim fibers. *J Cardiovasc Electrophysiol* 2004;15:738.
25 Hluchy J, Schickel S, Jörger U, *et al*. Electrophysiologic characteristics and radiofrequency ablation of concealed nodofascicular and left anterograde atriofascicular pathways. *J Cardiovasc Electrophysiol* 2000;11:211.
26 Fung WHJ, Chan HCK, Chan WWL, Sanderson JE. Ablation of the Mahaim pathway guided by noncontact mapping. *J Cardiovasc Electrophysiol* 2002;13:1064.
27 Tan HL, Wittkampf FHM, Nakagawa H, Derksen R. Atriofascicular accessory pathway. *J Cardiovasc Electrophysiol* 2004;15:118.
28 Goldberger JJ, Pederson DN, Damle RS, *et al*. Antidromic tachycardia utilizing decremental, latent accessory atrioventricular fibers: differentiation from adenosine-sensitive ventricular tachycardia. *J Am Coll Cardiol* 1994;24:732.
29 Johnson CT, Brooks C, Jaramillo J, *et al*. Left free-wall, decrementally conducting atrioventricular (Mahaim) fiber: diagnosis at electrophysiologic study and radiofrequency catheter ablation guided by direct recording of a Mahaim potential. *PACE* 1997;20:2486
30 Tada H, Nogami A, Naito S, *et al*. Left posteroseptal Mahaim fiber associated with marked longitudinal dissociation. *PACE* 1999;22:1696.
31 Sternick EB, Gerken LM, Vrandecic M, Wellens HJJ. Fasciculoventricular pathways: clinical and electrophysiologic characteristics of a variant of preexcitation. *J Cardiovasc Electrophysiol* 2003;14:1057.
32 Sternick EB, Rodriguez LM, Gerken LM, Wellens HJJ. The electrocardiogram of patients with fasciculoventricular pathways. A comparative study with patients with anteroseptal and midseptal accessory pathways. *Heart Rhythm* 2005;2:1.