Dr. Ronald Krone

&

Frank Yanowitz

Mystery ECG tracing

Medical Mystery:

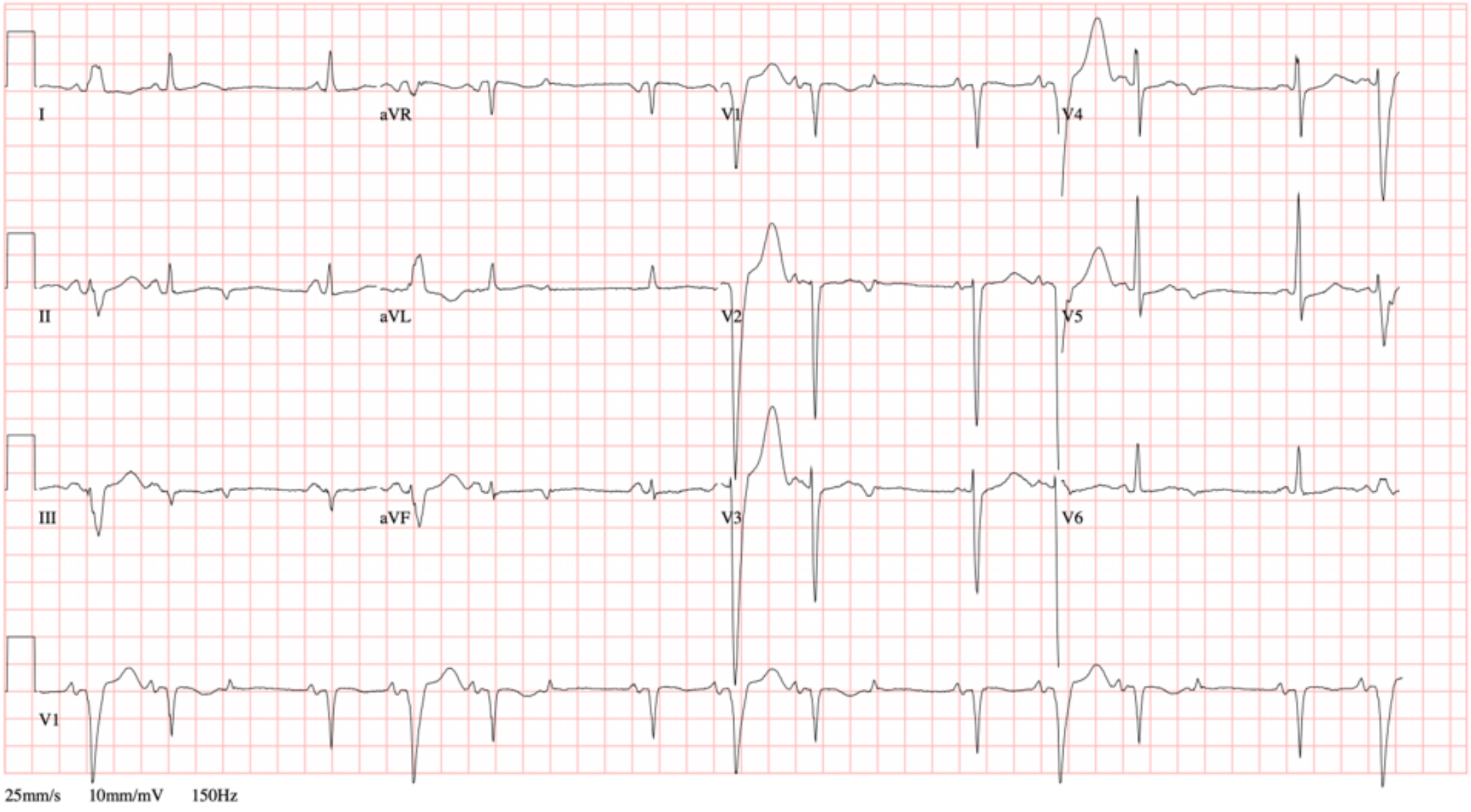
ECG Explained!!

Dear Andrés

My friend, Dr. Ronald Krone, sent me this interesting ECG. Ron and I first met in 1966 at the beginning of our internal medicine residency at University of Chicago; he subsequently went on the faculty of the Washington University, St. Louis, Missouri, and he is now, like me, a semi-retired professor teaching ECGs to residents and fellows. Last week he sent me this fascinating ECG.
I sent him my thoughts, but before I share them with you I would like your opinion and those of our colleagues from Latin forum.
Mi amigo, el Dr. Ronald Krone, me envió este interesante ECG. Ron y yo nos conocimos en 1966 al comienzo de nuestra residencia en medicina interna en la Universidad de Chicago; posteriormente pasó a la facultad de la Universidad de Washington, St. Louis, Missouri, y ahora es, como yo, un profesor semi-retirado que enseña ECG a residentes y becarios. La semana pasada me envió este fascinante ECG. Le envié mi opinión, pero antes de compartirlos contigo me gustaría tu opinión y la de los colegas compañeros del foro latino.
Meu amigo, Dr. Ronald Krone, me enviou este interessante ECG. Ron e eu nos conhecemos em 1966, no início de nossa residência em medicina interna na Universidade de Chicago; mais tarde ele se mudou para o corpo docente da Universidade de Washington, St. Louis, Missouri, e agora é, como eu, um professor semi-aposentado que ensina ECG para residentes e bolsistas. Na semana passada ele me enviou este fascinante ECG. Mandei minha opinião para ele, mas antes de compartilhar com vocês gostaria da sua opinião e dos colegas do fórum latino..
Frank

Dr. Frank Yanowitz

Dr. Ronald Krone



Woman in her 60s, no history provided; note repetitive grouping of beats!

Mujer de unos 60 años, sin antecedentes informados; ¡Observe la agrupación repetitiva de los latidos!

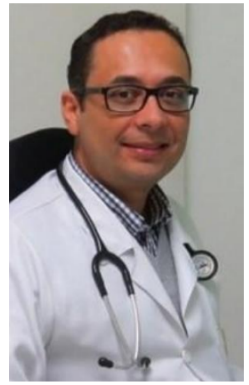
Mulher de 60 anos, sem antecedentes informados; Observe o agrupamento repetitivo dos batimentos!

Very interesting ECG, Please follow my interpretation:

Sinus rhythm where we observe a regular irregularity characterized by sets of 4 beats with regular PP intervals. The first beat is conducted by the AV node (narrow QRS), the second one presents left bundle branch block morphology with precordial transition zone of the QRS in V5, rS in III(**Sternick EB, Timmermans C, Sosa E, et al. The electrocardiogram during sinus rhythm and tachycardia in patients with Mahaim fibers: the importance of an "rS" pattern in lead III. *J Am Coll Cardiol* 2004; 44: 1626–1635**) and absence of initial q wave in I, which suggests conduction by a Atriofascicular APs (so-called Mahaim-type accessory pathways (MAPs))(**Eduardo Back Sternick 1, Luiz Márcio Gerken. The 12-lead ECG in patients with Mahaim Ann fibers Non-invasive electrocardiol. 2006 Jan;11(1):63-83. doi: 10.1111/j.1542- 474X.2006.00088.x.**). The third beat has a narrow QRS and is similar to the first, again indicating conduction through the AV node. In the sequence, we observed a blocked and negative P wave in the inferior wall, which suggests retrograde activation, probably by a slow nodal pathway (atrial echo). The phenomenon is repeated every 4 beats, indicating a possible association between a double nodal conduction pathway and a Mahain fiber.

A cordial hug,

Acácio Cardoso. MD, PhD



Acácio F. Cardoso MD, PhD

Cardiology Service of the Nipo Brasileiro Hospital, São Paulo/SP, Brasil.

ECG bastante interessante, segue minha avaliação do traçado:

Ritmo sinusal onde observamos uma irregularidade regular caracterizada por blocos de 4 batimentos com intervalos PP regulares. O primeiro batimento desce pelo Nó AV (QRS estreito), o segundo batimento tem morfologia de Bloqueio de ramo esquerdo com transição precordial do QRS após V5, rS em III e ausência de q em I o que sugere a condução por uma via anômala do tipo Mahain (**Eduardo Back Sternick 1, Luiz Márcio Gerken. The 12-lead ECG in patients with Mahaim fibers Ann Noninvasive Electrocardiol. 2006 Jan;11(1):63-83. doi: 10.1111/j.1542-474X.2006.00088.x.**). O terceiro batimento o QRS é estreito e semelhante ao primeiro batimento do ciclo indicando condução novamente pelo Nó AV. Na sequência temos uma onda P bloqueada e negativa na parede inferior sugerindo uma ativação retrógrada provavelmente por uma via lenta nodal (echo atrial). O fenômeno se repete a cada 4 batimentos e indica uma possível associação entre uma dupla via de condução nodal e uma fibra de Mahain.

Um cordial abraço,

Acácio Cardoso. MD PhD

ECG muy interesante, Sigue mi interpretación del ECG :

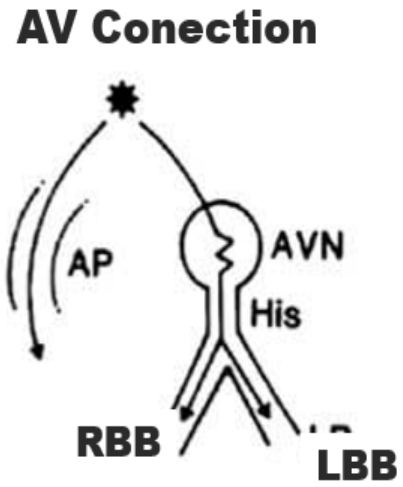
Ritmo sinusal donde observamos una irregularidad regular caracterizada por conjuntos de 4 latidos con intervalos PP regulares. El primer latido se conduce por el Nódulo AV (QRS estrecho), el segundo presenta morfología de bloqueo de rama izquierda com zona de transición precordial del QRS posterior a V5, rS en III y ausencia de q en I, lo que sugiere conducción por una vía anómala tipo Mahain (Eduardo Back Sternick 1, Luiz Márcio Gerken. El ECG de 12 derivaciones en pacientes con fibras de Mahaim Ann Electrocardiol no invasivo. 2006 Ene;11(1):63-83. doi: 10.1111/j.1542-474X.2006.00088.x.). El tercer latido tiene el QRS angosto y es similar al primero, lo que indica nuevamente conducción a través del Nódulo AV. En la secuencia observamos una onda P bloqueada y negativa en la pared inferior, lo que sugiere activación retrógrada, probablemente por una vía nodal lenta (eco auricular). El fenómeno se repite cada 4 latidos indicando posible asociación entre una vía de conducción nodal doble y una fibra de Mahain.

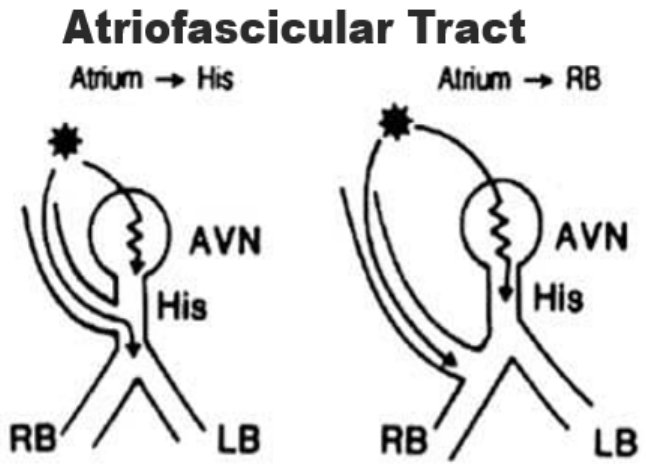
Un cordial abrazo,

Acacio Cardoso. MD, PhD

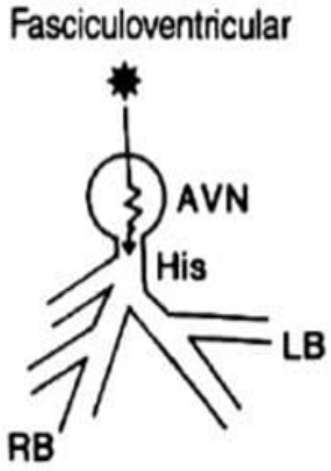
Theoretical considerations related Acasio´ hypoteis

The term “Mahaim fibers” refers to atriofascicular bypass tracts that connect the right atrium to the distal right bundle branch (see the side image). These pathways usually represent a duplication of the AV node and the distal conducting system. They typically occupy the right ventricular(RV) free wall. Their proximal end resides adjacent to the lateral tricuspid annulus and exhibits slow conduction, with AV node–like characteristics. The distal end, which conducts rapidly, inserts into the distal right bundle branch (RBB) or the apical region of the RV. MAPs were most often ablated at the lateral aspect of the tricuspid annuli, sometimes at other sides of the tricuspid and mitral annuli, and infrequently in the RV. The M potential mapping technique is likely to be a useful target for ablation of MAPs.(1)

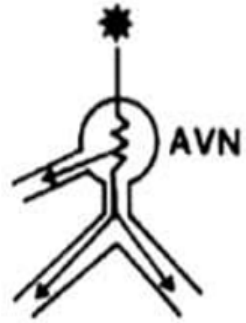




Mahaim fibers



Nodoventricular fibers



1) Suat Gormel, Salim Yasar, Erkan Yildirim, et al Comprehensive assessment of Mahaim accessory pathways' anatomic distribution J Int Med Res . 2022 Jan;50(1):300605211069751. doi: 10.1177/0300605211069751.

If Mahaim fibers are present, the ECG findings are a normal or long PR interval and an abnormally wide QRS complex with a left-bundle branch appearance. Preexcitation may not be apparent during sinus rhythm but can be demonstrated with premature right atrial stimulation. Because retrograde conduction is absent, only an antidromic AV reentry tachycardia (ie, preexcited tachycardia) can develop. In the presence of atriofascicular tracts, preexcited tachycardia has a LBBB pattern, a long AV interval (due to the long conduction time over the accessory pathway), and short ventriculoatrial (VA) intervals. If right bundle-branch block (RBBB) develops, it may prolong the tachycardia cycle length (slow the AV reentrant tachycardia [AVRT]). Any right free wall bypass tract-mediated SVT will be prolonged with RBBB but should be unaffected by LBBB aberrancy. Conversely, left free wall bypass tract AVRT will slow with LBBB (ipsilateral to the AP). Septal pathways may be slightly affected by either RBBB (anteroseptal) or LBBB (posteroseptal), but this would be detected only on electrophysiologic study (EPS). The eponym Coumel is also applied to this rule. Given the pathway length and decremental conduction properties (similar to AV nodal conduction), a preexcited QRS complex with a short PR interval essentially rules out Mahaim fibers.
In LGL syndrome, patients have a short PR interval and SVT but no delta wave. This is typically caused by an atrio-Hisian pathway leading to accelerated AV conduction, but not causing paroxysmal SVT. In patients with LGL syndrome who have an atrio-Hisian tract, the QRS complex remains normal and the short atrio-Hisian interval remains fixed during atrial pacing at rapid rates. Entities that involve wide-QRS SVT must be differentiated from ventricular tachycardia (VT). Examples include the following: Aberrantly conducting orthodromic SVT, which is wide-QRS SVT with the AV node as the antegrade limb but with bundle branch block, must be differentiated from VT Antidromic tachycardia, which is wide-QRS SVT due to ventricular preexcitation through an AP, must also be differentiated from VT and from Mahaim fiber tachycardia.
Sometimes, accessory AV fibers connect to the AV node itself or the His bundle or bundle branches and

insert into the ventricular myocardium. These are called nodoventricular or fasciculoventricular tracts. Patients with fasciculoventricular connections show a short His-ventricle (HV) interval and no change in the QRS complex during rapid atrial pacing. Fasciculoventricular AV pathways do not participate in clinically meaningful arrhythmias. Stanley Kent, an English physiologist, and Wilhelm His Jr, a Swiss cardiologist and anatomist, are well-known for conducting successful and inspiring studies into AV conduction and AP, dating back to 1893.(**His W., Jr. The action of the embryonic heart and its significance in the doctrine of heart movement in adults. *Arb Med Klinik Leipzig* 1893; 1: 14–49**) (**Kent AFS. Researches on the structure and function of the mammalian heart. *J Physiol* 1893; 14: 233–254**.)While Kent had suggested that multiple myocardial connections were present across AV junctions of the normal heart, His described a solitary muscular connection extending from the atrial to ventricular septum. The studies of His were validated over time, with the normal heart shown not to possess multiple myocardial connections across AV junctions.(**Tawara S. *Das Reizleitungssystem des Saugetierherzens: eine Anatomisch-Histologische Studie uber das Atrioventrikularbundelund die Purkinjeschen Faden*. Jena: Gustav Fischer, 1906. [In German]**) Rather than multiple pathways, Kent had shown node-like structures that did not cross the insulating plane, which he had still believed to provide normal AV conduction.-(**Kent AFS. The right lateral auriculo-ventricular junction of the heart. In: Proceedings of the Physiological Society, March 14, 1914. *J Physiol* 1914; 48: xxii–xxiv**.) (**Anderson RH, Davies MJ, Becker AE. Atrioventricular ring specialised tissue in the human heart. *Eur J Cardiol* 1974; 2: 219–230**)These findings allowed cardiologists to predict that arrhythmias now defined as ventricular pre-excitation, are the consequence of such multiple myocardial AV connections being present. Even to date, accessory muscular connections that are presumed to be the anatomical substrates of WPW Syndrome,

are sometimes defined as ‘bundles of Kent’, despite the fact that the connections have been demonstrated to have no anatomic similarity to the structures demonstrated by Kent.(**Ohnell RF. *Pre-excitation: a cardiac abnormality. Patho-physiological, patho-anatomical and clinical studies of an excitatory spread phenomenon bearing upon the problem of the WPW (Wolff, Parkinson and White) electrocardiogram and paroxysmal tachycardia*. Stockholm: PA Norstedt, 1944, pp.1–167.)** With the use of programmed electrical stimulation, electrophysiological studies in the late 1960’s and early 1970’s revealed that WPW syndrome was one variant of ventricular pre-excitation regarding the structure and location of alternative potential anatomical pathways between the atrial and ventricular muscle masses. (**Durrer D, Schuilenburg RM, Wellens HJJ. Pre-excitation revisited. *Am J Cardiol* 1970; 25: 690–697.**) A 1975 review by a group of European anatomists and pathologists proposed nomenclature for such potential pathways.(**Anderson RH, Becker AE, Brechenmacher C, et al. Ventricular preexcitation. A proposed nomenclature for its substrates. *Eur J Cardiol* 1975; 3: 27–36**.) While many of these unusual paths remain controversial in terms of their anatomy, the names used for their descriptions have become somewhat deficient. In general, the pathways produce the so-called ‘Mahaim’ pre-excitation. (**Anderson RH, Ho SY, Gillette PC, et al. Mahaim, Kent and abnormal atrioventricular conduction. *Cardiovasc Res* 1996; 31: 480–491**.) Initially, it was believed that pre-excitation characterized by decremental conduction was produced by connections between the normal AV conduction axis and the crest of the ventricular septum, as described by Mahaim et al.(**Mahaim I, Winston MR. Comparative anatomy and experimental pathology research on the high connections of the His–Tawara bundle. *Cardiologia* 1941; 5: 189–260**) (**Mahaim I. Kent’s fibers and the A-V paraspecific conduction through the upper connections of the bundle of His-Tawara. *Am Heart J* 1947; 33: 651–653**). However, it was later shown that such pre-excitation, developed from a pathway revealing

decremental conduction, developed from a pathway revealing decremental conduction, could also be constructed by a pathway of atrial origin with morphological characteristics of the structures shown by Kent. (**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: 1035–1040**.)(**Anderson RH, Sánchez-Quintana D, Mori S, et al. Unusual variants of pre-excitation: From anatomy to ablation: Part I-Understanding the anatomy of the variants of ventricular pre-excitation. *J Cardiovasc Electrophysiol* 2019; 30: 2170–2180**.) APs that arise from atrial origin were not mentioned by Mahaim or other European morphologists at that time. Mahaim-type accessory pathways (MAPs) usually originate within the lateral tricuspid annulus and cross the AV junctions, and display decremental conduction, inserting into the distal RV free wall.(**Anderson RH, Sánchez-Quintana D, Mori S, et al. Unusual variants of pre-excitation: From anatomy to ablation: Part I-Understanding the anatomy of the variants of ventricular pre-excitation. *J Cardiovasc Electrophysiol* 2019; 30: 2170–2180**.)[12–18] (**Ross DL, Johnson DC, Koo CC, et al. Surgical treatment of supraventricular tachycardia without the WPW syndrome: current indications, techniques and results. In: Brugada P, Wellens HJJ. (eds) *Cardiac Arrhythmias: Where to Go From Here*? Mount Kisco, NY: Futura Publishing Co, 1987, pp.591–603**.)(**Leitch J, Klein GJ, Yee R, et al. New concepts on nodoventricular accessory pathways. *J Cardiovasc Electrophysiol* 1990; 1: 220–230)(Murdock CJ, Leitch JW, Klein GJ, et al. Epicardial mapping in patients with "nodoventricular" accessory pathways. *Am J Cardiol* 1991; 68: 208–214**.)(**Soares Correa F, Lokhandwala Y, Cruz Filho F, et al. Part II-Clinical presentation, electrophysiologic characteristics, and when and how to ablate atriofascicular pathways and long and short decrementally conducting accessory pathways. *J Cardiovasc Electrophysiol* 2019; 30: 3079–3096**)

)(**Soares Correa F, Lokhandwala Y, Sánchez-Quintana D, et al. Unusual variants of pre-excitation: From anatomy to ablation: Part III-Clinical presentation, electrophysiologic characteristics, when and how to ablate nodoventricular, nodofascicular, fasciculoventricular pathways, along with considerations of permanent junctional reciprocating tachycardia. *J Cardiovasc Electrophysiol* 2019; 30: 3097–3115**.)Most reported decremental conducting MAPs are right-sided. However, paraseptal and left-sided APs are also reported.(**Bohora S, Dora SK, Namboodiri N, et al. Electrophysiology study and radiofrequency catheter ablation of atriofascicular tracts with decremental properties (Mahaim fibre) at the tricuspid annulus. *Europace* 2008; 10: 1428–1433**.) (**Okishige K, Goseki Y, Itoh A, et al. New electrophysiologic features and catheter ablation of atrioventricular and atriofascicular accessory pathways: evidence of decremental conduction and the anatomic structure of the Mahaim pathway. *J Cardiovasc Electrophysiol* 1998; 9: 22–33.**)The left-sided decrementally conducting APs originate within the mitral annulus and their distal end inserts into the left ventricle, while the origin and insertion point of paraseptal APs may vary according to its septal side. Radiofrequency (RF) catheter ablation is the exclusive treatment option for MAPs, offering a definitive therapy with a high success rate and minimal morbidity,(**Soares Correa F, Lokhandwala Y, Cruz Filho F, et al. Part II-Clinical presentation, electrophysiologic characteristics, and when and how to ablate atriofascicular pathways and long and short decrementally conducting accessory pathways. *J Cardiovasc Electrophysiol* 2019; 30: 3079–3096**.)(**Soares Correa F, Lokhandwala Y, Sánchez-Quintana D, et al. Unusual variants of pre-excitation: From anatomy to ablation: Part III-Clinical presentation, electrophysiologic characteristics, when and how to ablate nodoventricular, nodofascicular, fasciculoventricular pathways, along with considerations of permanent junctional reciprocating tachycardia. *J Cardiovasc Electrophysiol* 2019; 30: 3097–3115**) although few case

series are noted in the literature. (**Kothari S, Gupta AK, Lokhandwala YY, et al. Atriofascicular pathways: Where to ablate? *Pacing Clin Electrophysiol* 2006; 29: 1226–1233**.)(**Sternick EB, Timmermans C, Sosa E, et al. The electrocardiogram during sinus rhythm and tachycardia in patients with Mahaim fibers: the importance of an "rS" pattern in lead III. *J Am Coll Cardiol* 2004; 44: 1626–1635**.)(**Sternick EB, Timmermans C, Sosa E, et al. The electrocardiogram during sinus rhythm and tachycardia in patients with Mahaim fibers: the importance of an "rS" pattern in lead III. *J Am Coll Cardiol* 2004; 44: 1626–1635**.).(**Liao Z, Ma J, Hu J, et al. New observation of electrocardiogram during sinus rhythm on the atriofascicular and decremental atrioventricular pathways/clinical perspective: [corrected] terminal QRS [corrected] complex slurring or notching. *Circ Arrhythm Electrophysiol* 2011; 4: 897–901.**)