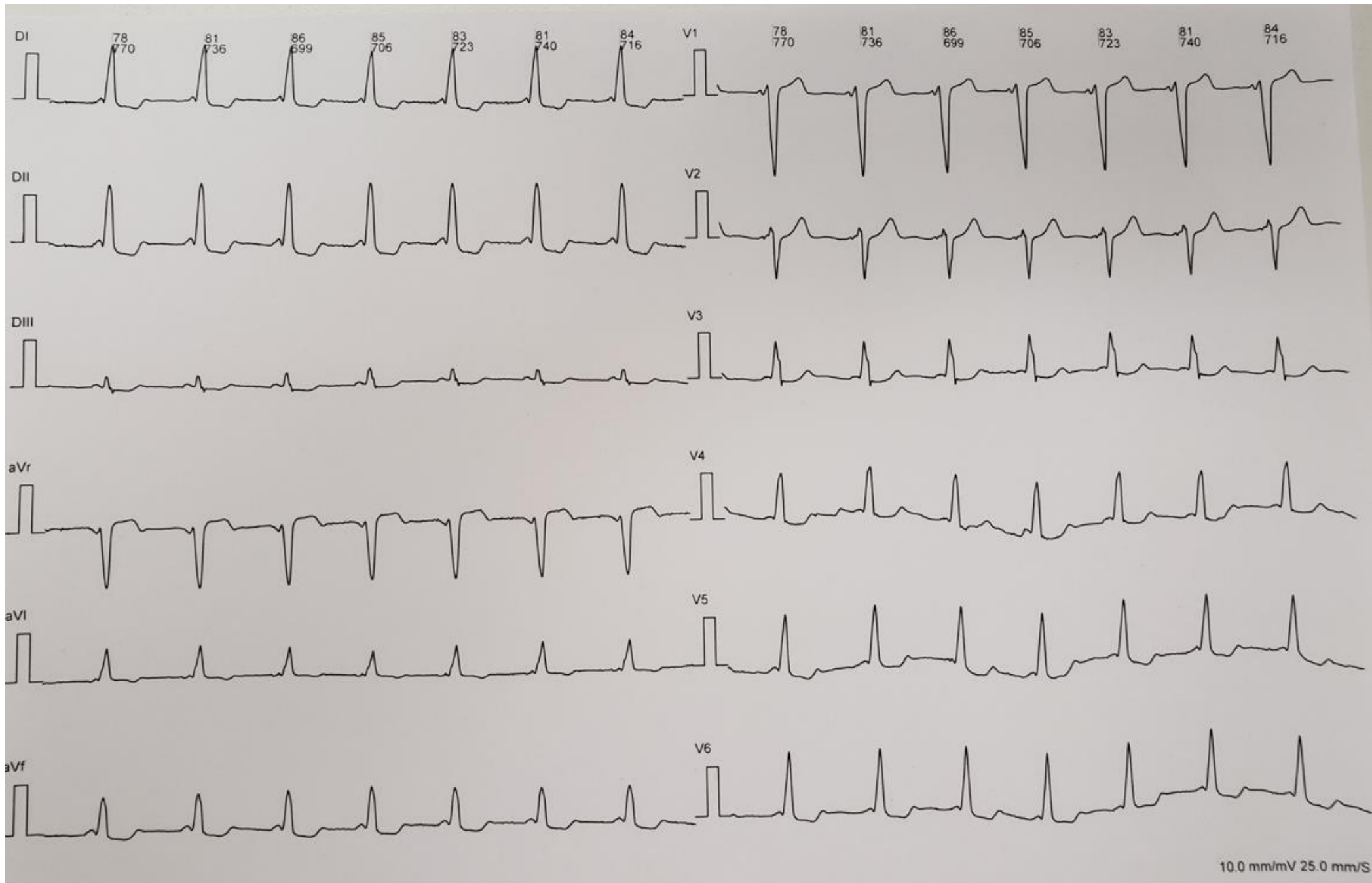


Dr. Alfredo del Rio case of ventricular preexcitation in 14 years old adolescent



Current Nomenclature and Proposed Terminology for Accessory Pathway location

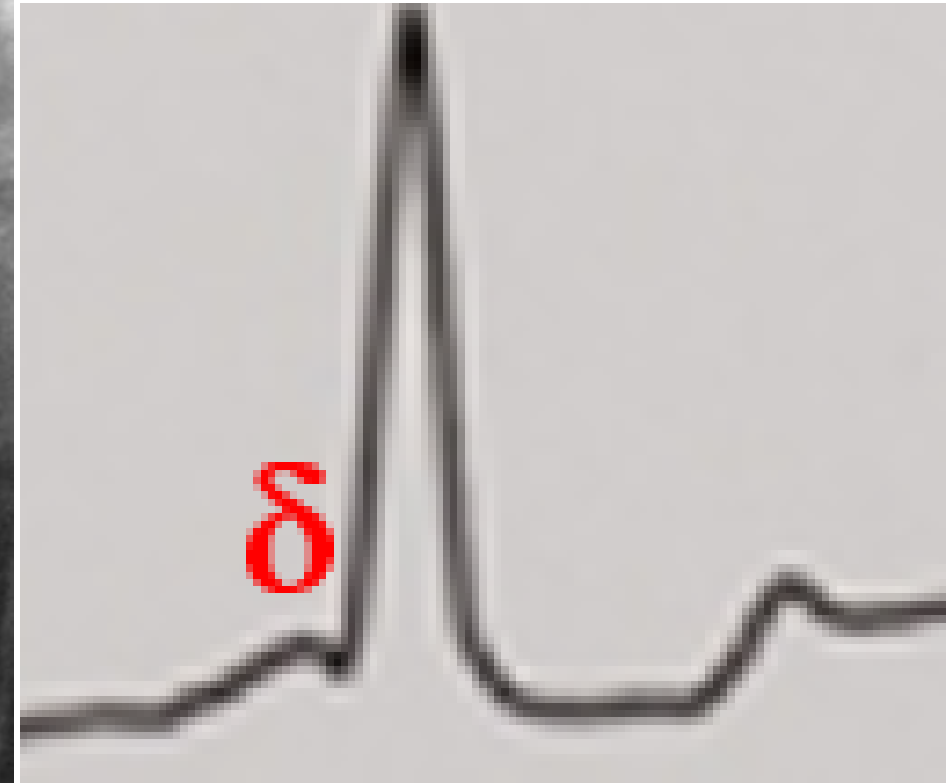
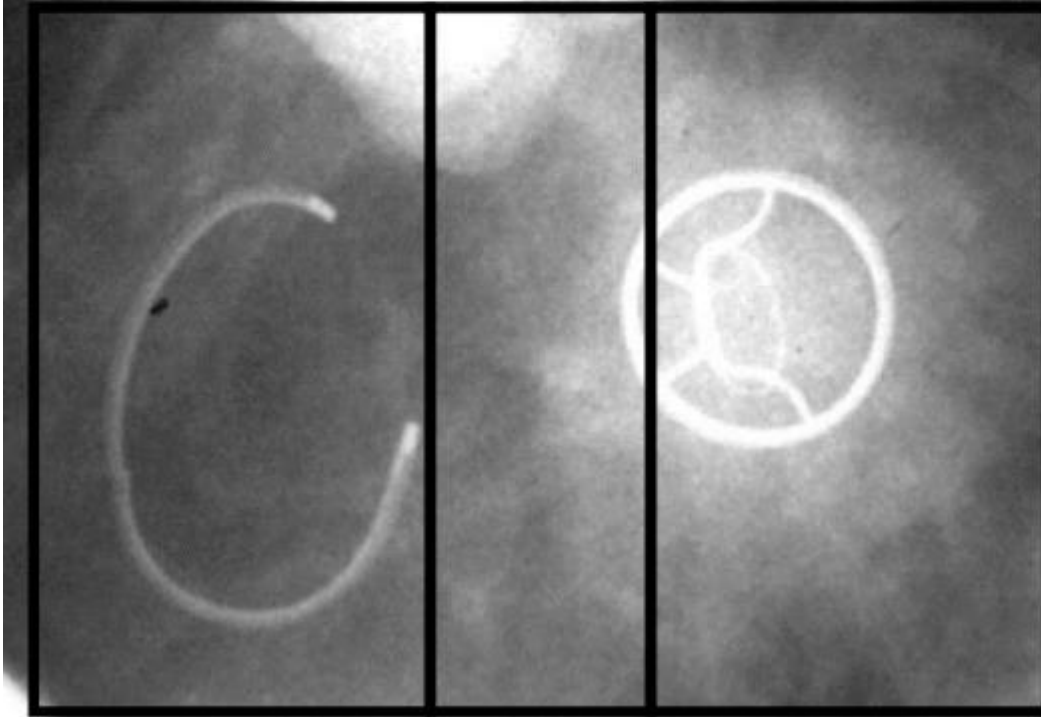
Current(Attitudinally Incorrect)	Proposed(Attitudinally Correct
Right	
Anterior	Superior
Anterolateral	Supero-anterior
Lateral	Anterior
Basal-lateral (ancient posterolateral)	Infero-anterior
Basal inferior(ancient posterior	Inferior
Left	
Anterior	Superior
Anterolateral	Supero-basal (Ancient supero-posterior)
Basal-lateral(ancient posterolateral)	Posterior
Basal inferior(ancient posterior	Ífero-basal(ancient Ífero-posterior)
Septal paraseptal	Inferior
Anteroseptal	Superoparaseptal
Posteroseptal	Ìferopareseptal
Midseptal	Septal

Proposed terminology is based in anatomic position

First step: QRS with (Δ or δ) wave and wide?

1. Sufficient
pre-excitation?

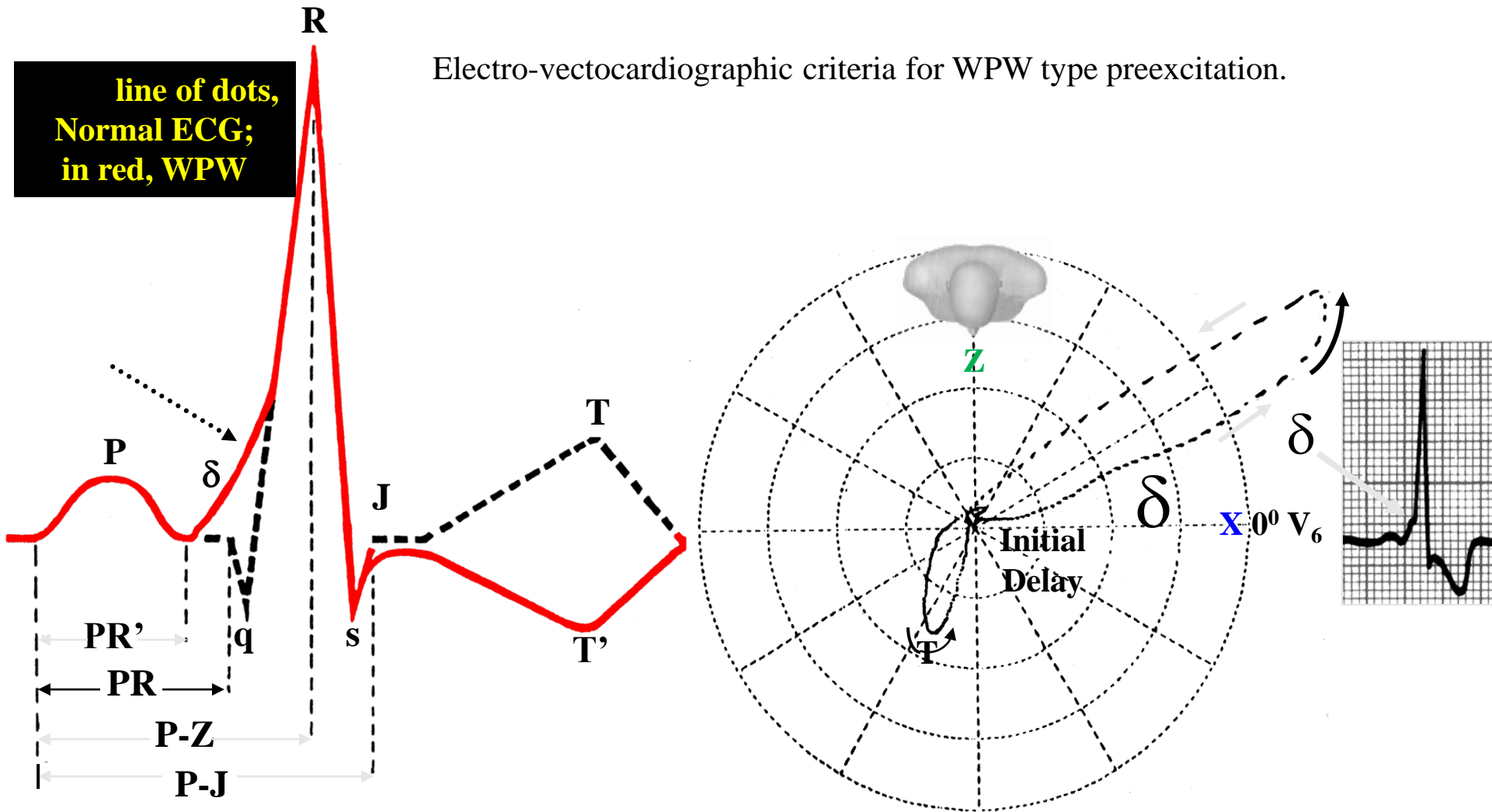
(Δ QRS 120 ms)



Answer: sufficient for preexcitation criteria: short PR interval, Δ / δ at the beginning of QRS, prolonged QRS: ≥ 120 ms. See ECG/VCG criteria in the next slide.

WPW ECG/VCG correlation

Electro-vectocardiographic criteria for WPW type preexcitation.



- **PRi or PQ:** since the onset of P up to the onset of QRS. It represents the time the stimulus takes to go from the SA node until reaching the ventricles: 120 ms to 200 ms.
- **PZ:** distance between P wave onset until R apex: 150 to 230 ms.
- **PJ:** distance between P wave onset until j point: 180 to 260 ms.

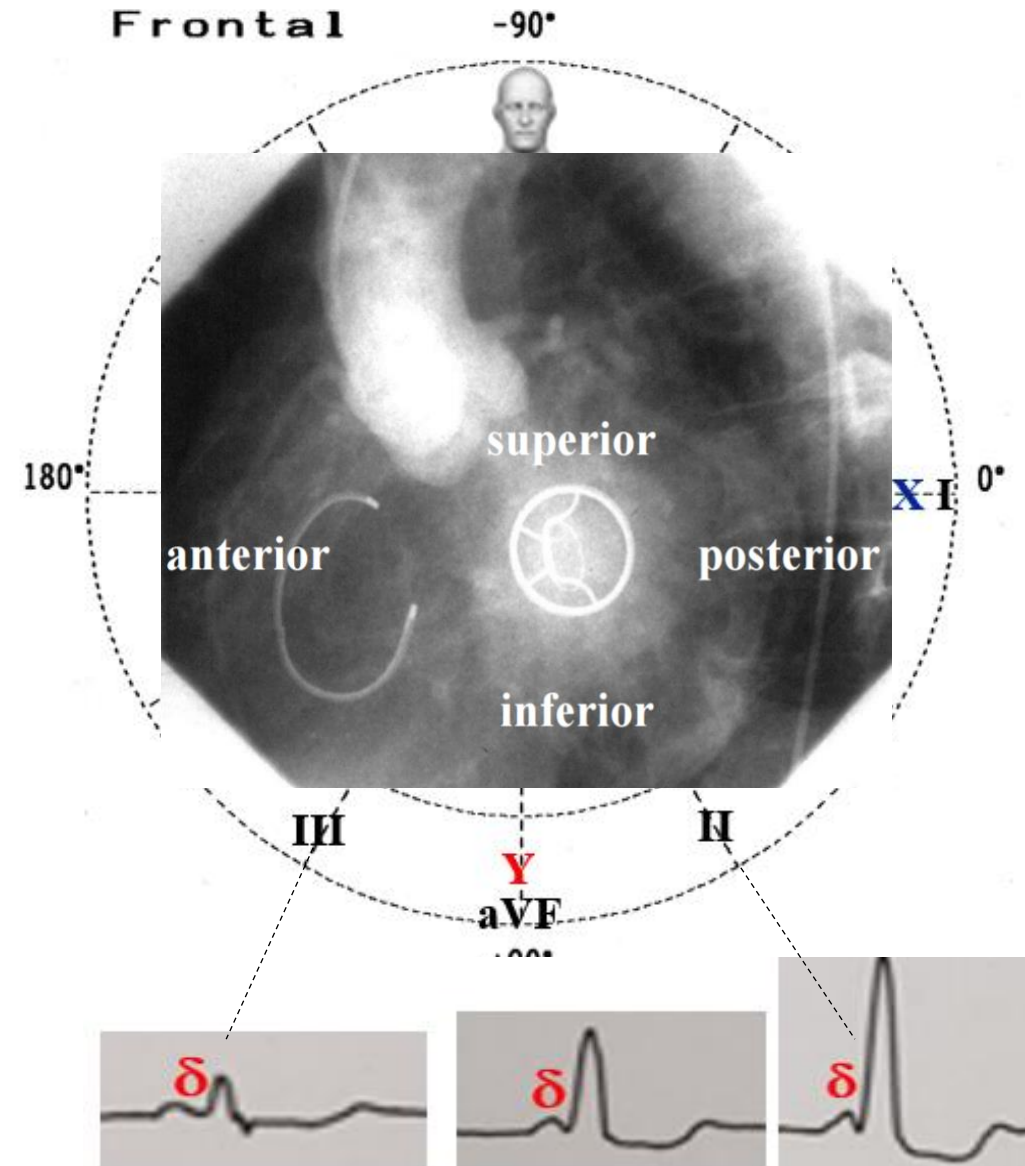
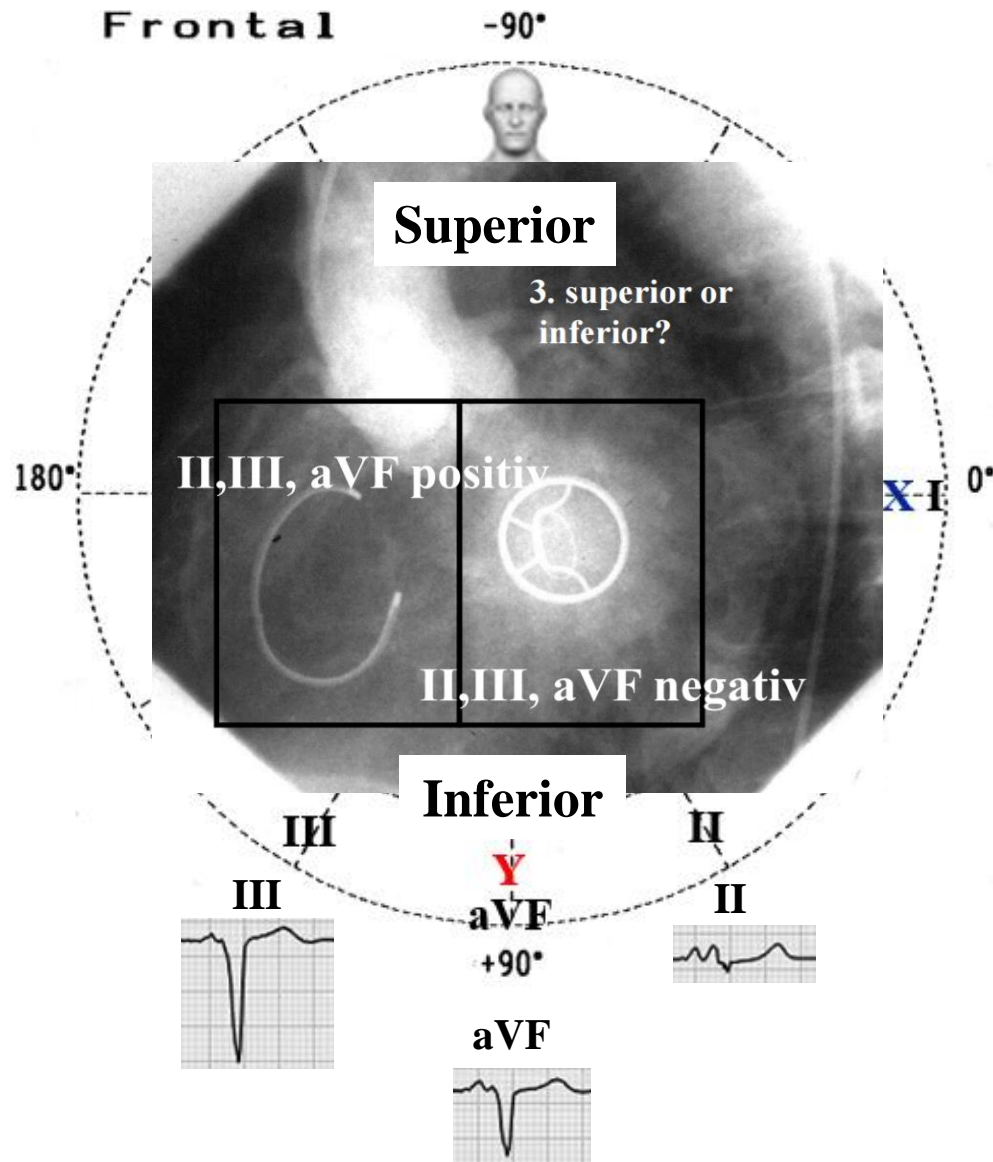
- Initial delay of QRS loop: delta wave.
- T-loop opposite to QRS loop

Second step: right side or left side?

The image displays an ECG tracing on the left with a red Greek letter δ marking the initial S wave. To the right, a chest X-ray is shown with a central box containing two diagrams of the heart. The left diagram, labeled 'V1: S>R', shows a heart with a large S wave and a smaller R wave. The right diagram, labeled 'V1: R>S', shows a heart with a larger R wave and a smaller S wave. Above the diagrams are labels 'Right side' in blue and 'Left side' in red. The text '2. Right-sided or left-sided?' is positioned at the top right of the X-ray area.

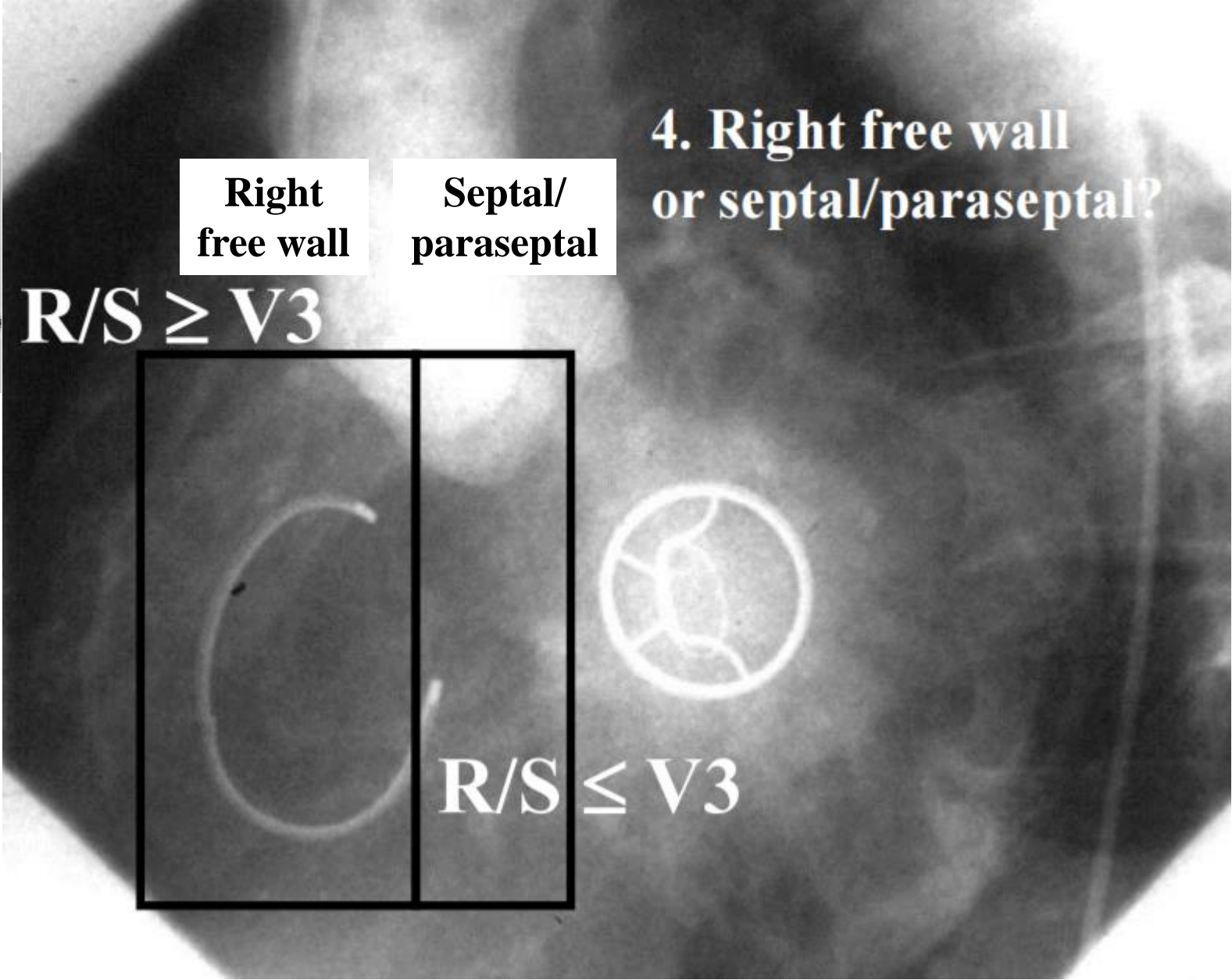
Answer: right side, because $S>R$ in V1.

Third step: superior or inferior?



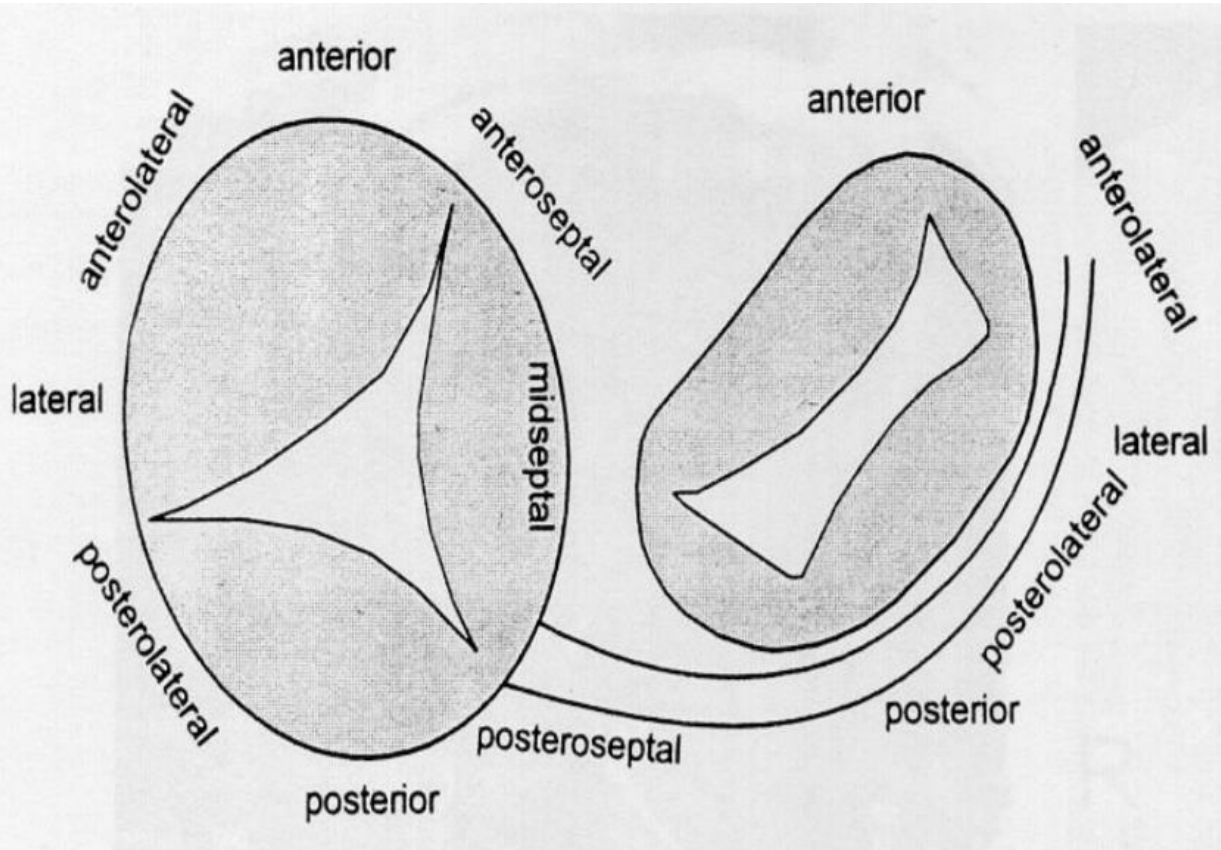
Answer: superior, because QRS in the inferior leads are predominantly positive

Fourth step: Right free wall or septal/pareseptal?

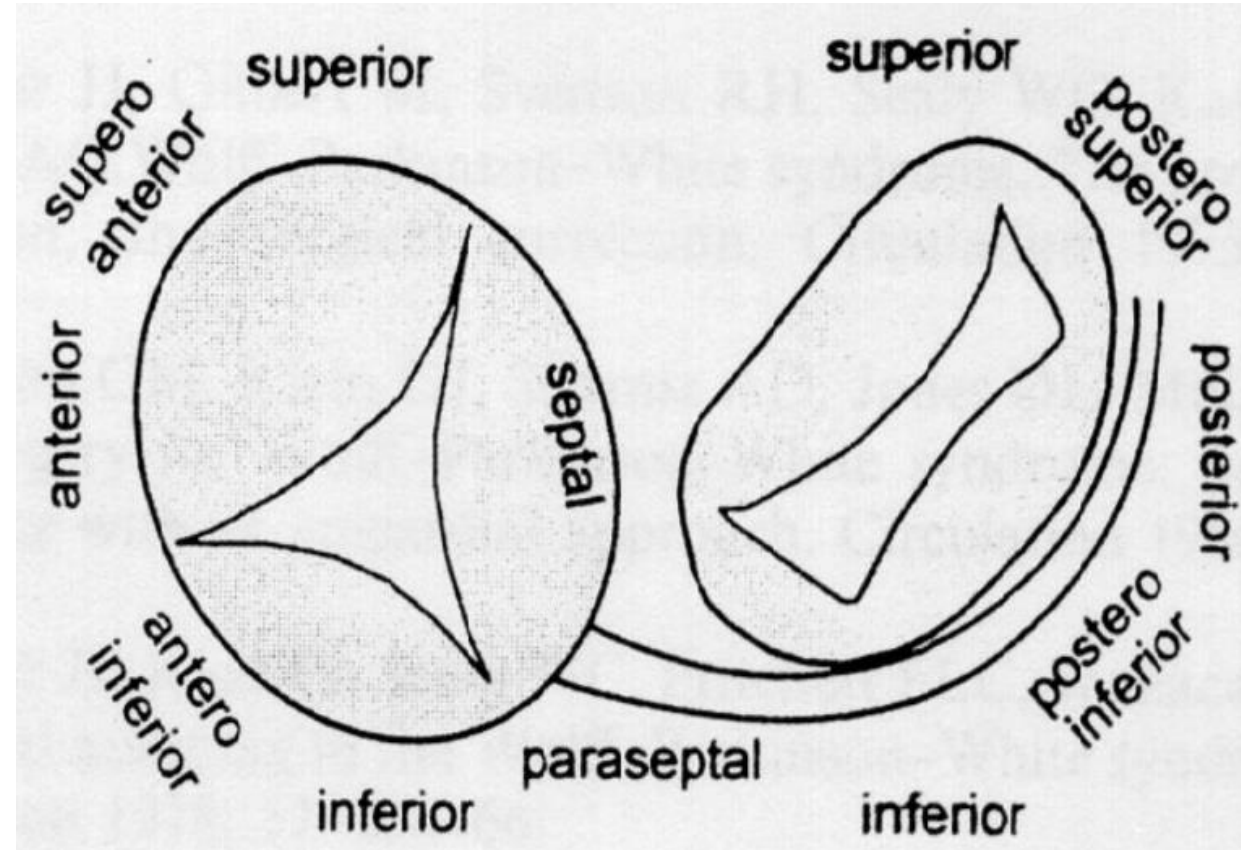


Answer: Right free wall because R/S ratio \geq V3:

Old nomenclature for Accessory Pathways location



New nomenclature for Accessory Pathways location



Li HY1,2,3, Chang SL4,5, Chuang CH6, Lin MC2,6, Lin YJ4,5, Lo LW4,5, Hu YF4,5, Chung FP4,5, Chang YT4,5, Chung CM4,7, Chen SA4,5, Lee PC1,2. A Novel and Simple Algorithm Using Surface Electrocardiogram That Localizes Accessory Conduction Pathway in Wolff-Parkinson-White Syndrome in Pediatric Patients. Acta Cardiol Sin. 2019 Sep;35(5):493-500. doi: 10.6515/ACS.201909_35(5).20190312A. **Manuscript free in Pubmed!!!!!! It is important because Dr. Alfredo del Rio case is a children/adolescent)** **Author information**

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Abstract BACKGROUND: The location of the accessory pathway (AP) can be precisely identified on surface electrocardiography (ECG) in adults with Wolff-Parkinson-White (WPW) syndrome. However, current algorithms to locate the AP in pediatric patients with WPW syndrome are limited.

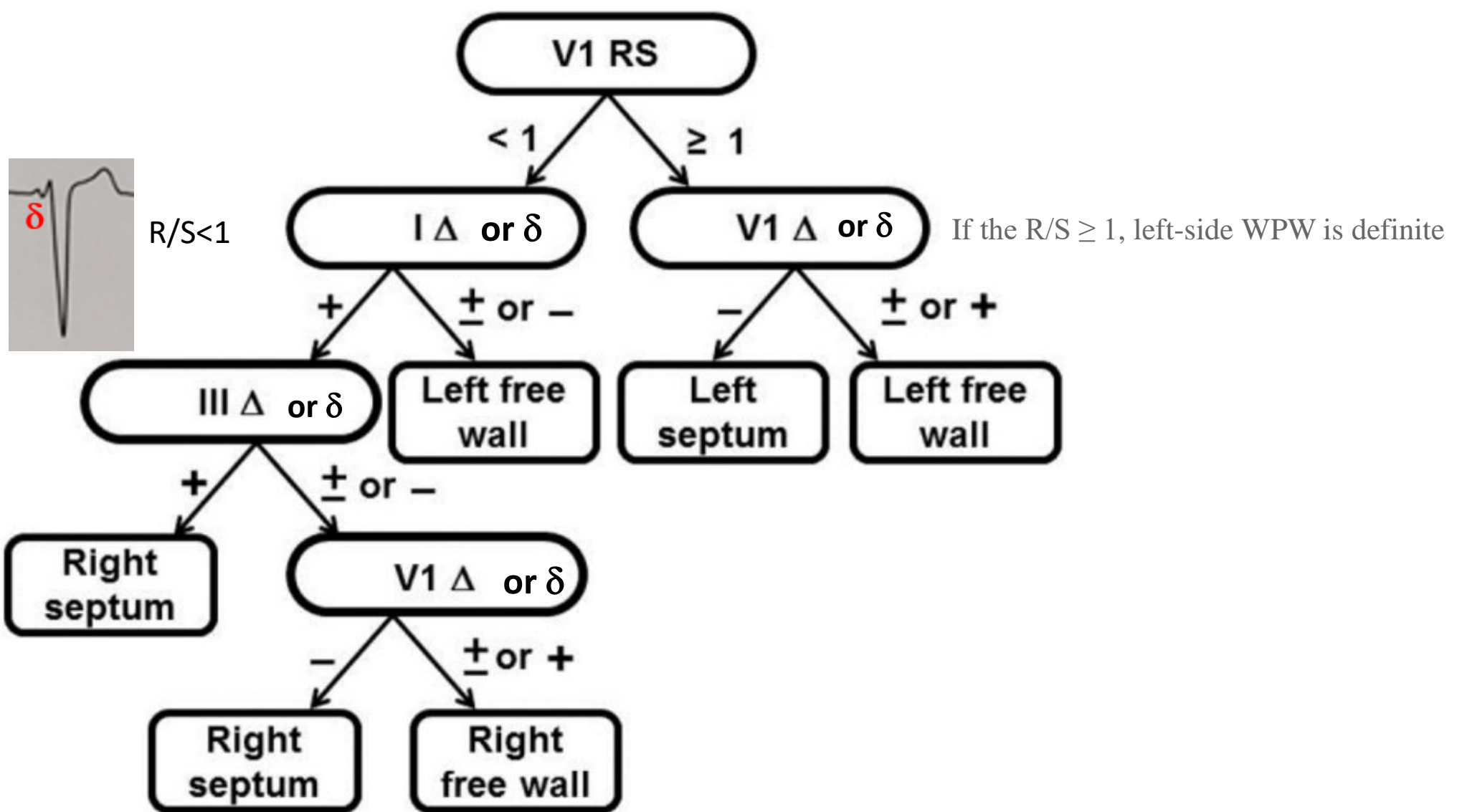
OBJECTIVE: To propose an optimal algorithm that localizes the AP in pediatric patients with WPW syndrome.

METHODS: From 1992 to 2016, 180 consecutive patients aged below 18 years with symptomatic WPW syndrome were included. After the exclusion of patients with non-descriptive electrocardiography (ECG), multiple APs, congenital heart diseases, non-inducible tachycardia, and those who received a second ablation, 104 patients were analyzed retrospectively. Surface ECG was obtained before ablation and evaluated by using previously documented algorithms, from which a new pediatric algorithm was developed.

RESULTS: Previous algorithms were not highly accurate when used in pediatric patients with WPW syndrome. In the new algorithm, the R/S ratio of V1 and the polarity of the delta wave in lead I could distinguish right from the left side AP with 100% accuracy. The polarity of the delta wave of lead V1 could distinguish free wall AP from septal AP with an accuracy of 100% in left-side AP, compared to 88.6% in leads III and V1 for right-side AP. The overall accuracy was 92.3%.

CONCLUSIONS: This simple, novel algorithm could differentiate left from right AP and septal from free wall AP in pediatric patients with WPW syndrome.

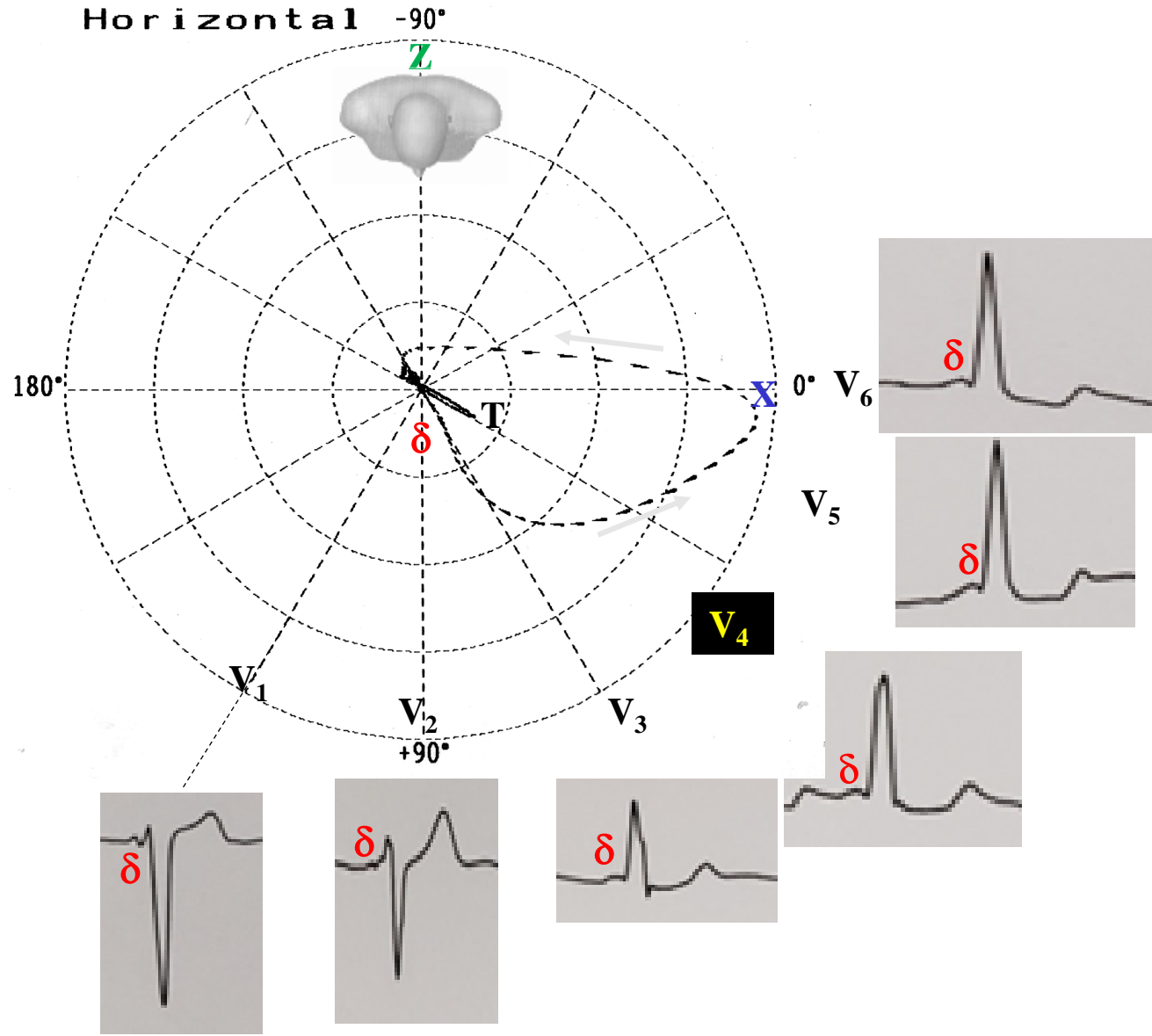
KEYWORDS: Accessory pathway; Algorithm; Children; Localization; WPW syndrome



The validated pediatric algorithm. Step 1 analyzes R/S ratio in V1; if the $R/S \geq 1$, left-side WPW is definite, and if the $R/S < 1$, the AP needs further evaluation by the polarity of the delta wave of Lead I. Step 2 analyzes the delta wave polarity in $V1 \geq 1$ in V1 for differentiating free wall and septum in left-side AP and analyzing the delta wave polarity in Lead I to differentiate into left- and right-side AP. Step 3 uses the delta wave polarity in Lead III that can separate partial right septum AP from right free wall. Step 4 further separates the right septum and right free wall AP by the delta wave polarity of V1.

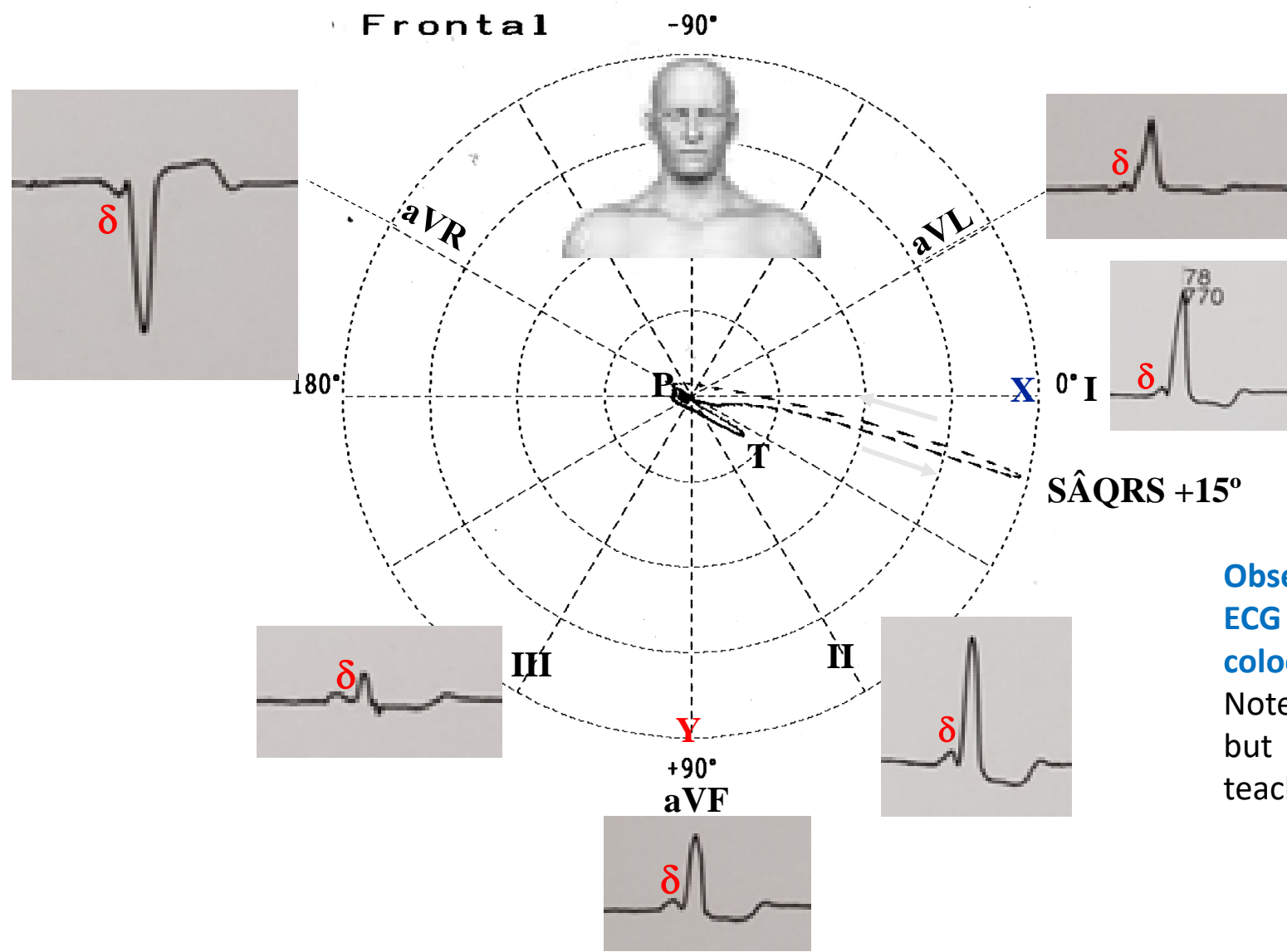
**The value of the Vectorcardiogram in Accessory
Pathway location: Hypothetical
ECG/VCG correlation**

Hypothetical ECG/VCG correlation in the Horizontal Plane



ECG/VCG of a patient with anomalous accessory pathway located on RV free wall (right lateral posterior ventricular pre-excitation) point Gallagher's 3 or 4, type II of WPW of European classification and type A WPW of Rosenbaum ancient classification (prominent anterior QRS forces). Ventricular activation from right to left and QRS loop located predominantly on anterior left quadrant: PAF.

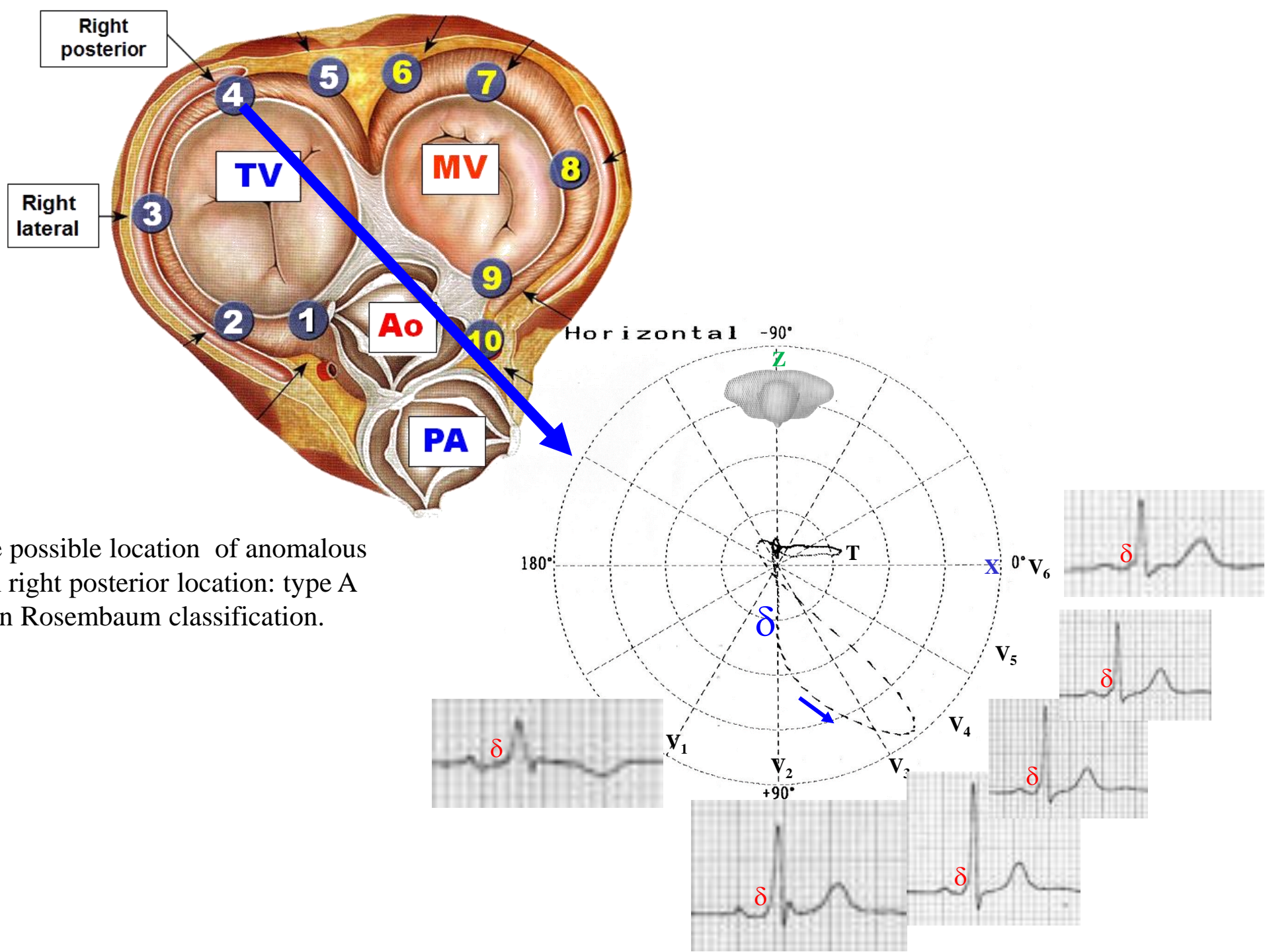
Hypothetical ECG/VCG correlation of posterior or right lateral ventricular preexcitation in the RV free wall on Frontal Plane



Observação: o VCG não pertence ao ECG mais é muito semelhante e coloco aos fins didáticos

Note: VCG does not belong to ECG but is very similar and I put it for teaching purposes

Anomalous accessory pathways located in the RV free wall, point 3 or 4 of Gallagher, SÂQRS not shifted and negative delta (δ) wave in aVR (ventricular activation from right to left). Region IV of Lindsay.



In this case the possible location of anomalous pathways is on right posterior location: type A WPW of ancient Rosebaum classification.

Sequential tracing analysis from professor Bernard Belhassen



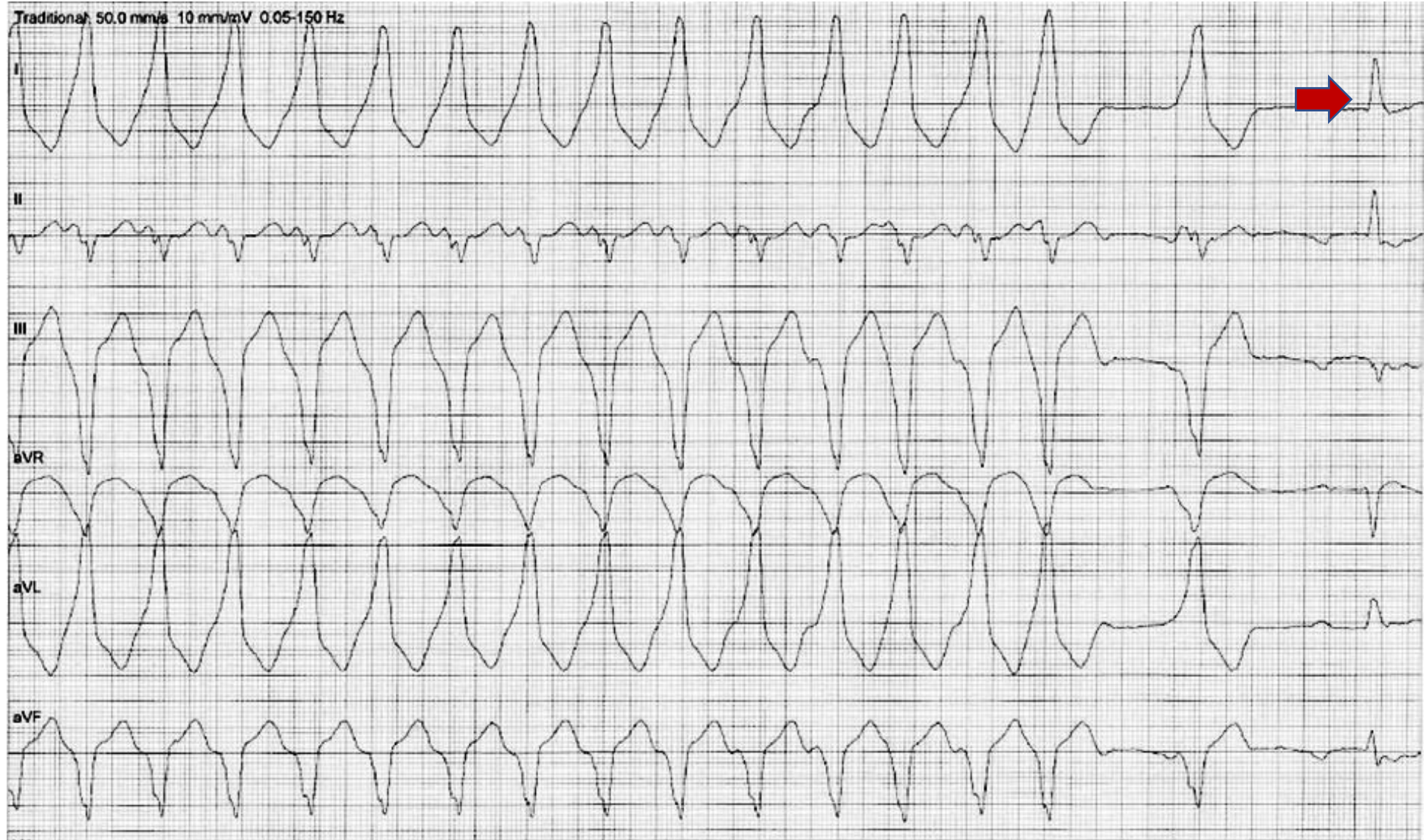
Cardiologist, Head of Electrophysiology Laboratory at The Tel Aviv Sourasky Medical Center. Tel-Aviv, Israel. Department of Cardiology, Tel Aviv Medical Center and Sackler Faculty of Medicine, Tel Aviv University, Tel Aviv, Israel. Electronic address: bblhass@tasmc.health.gov.il

Doctor Belhassen is a full professor of cardiology at Sackler School of Medicine. He has a professional interest in cardiac arrhythmias, sudden cardiac death, catheter ablation.



Pseudo-long PR interval associated with a typical right posteroseptal accessory pathway (AP) of course represented an atrial tachycardia/flutter 185/min with 2:1 conduction over the AP. However, should we conclude that it is impossible to have a similar tracing associated with normal sinus rhythm (NSR?) the response is: No !!! There is a theoretical possibility that NSR will be associated with both a long PR and a typical WPW in 2 instances: a) conduction with an atriofascicular AP ("Mahaim type") that is unlikely the case when dealing with an apparently typical right posteroseptal pathway; b) when the right posteroseptal pathway has a very long conduction time (this is a very exceptional feature of atrio-ventricular AP that might be seen for example after ablation of these APs).

50 mm/sec!

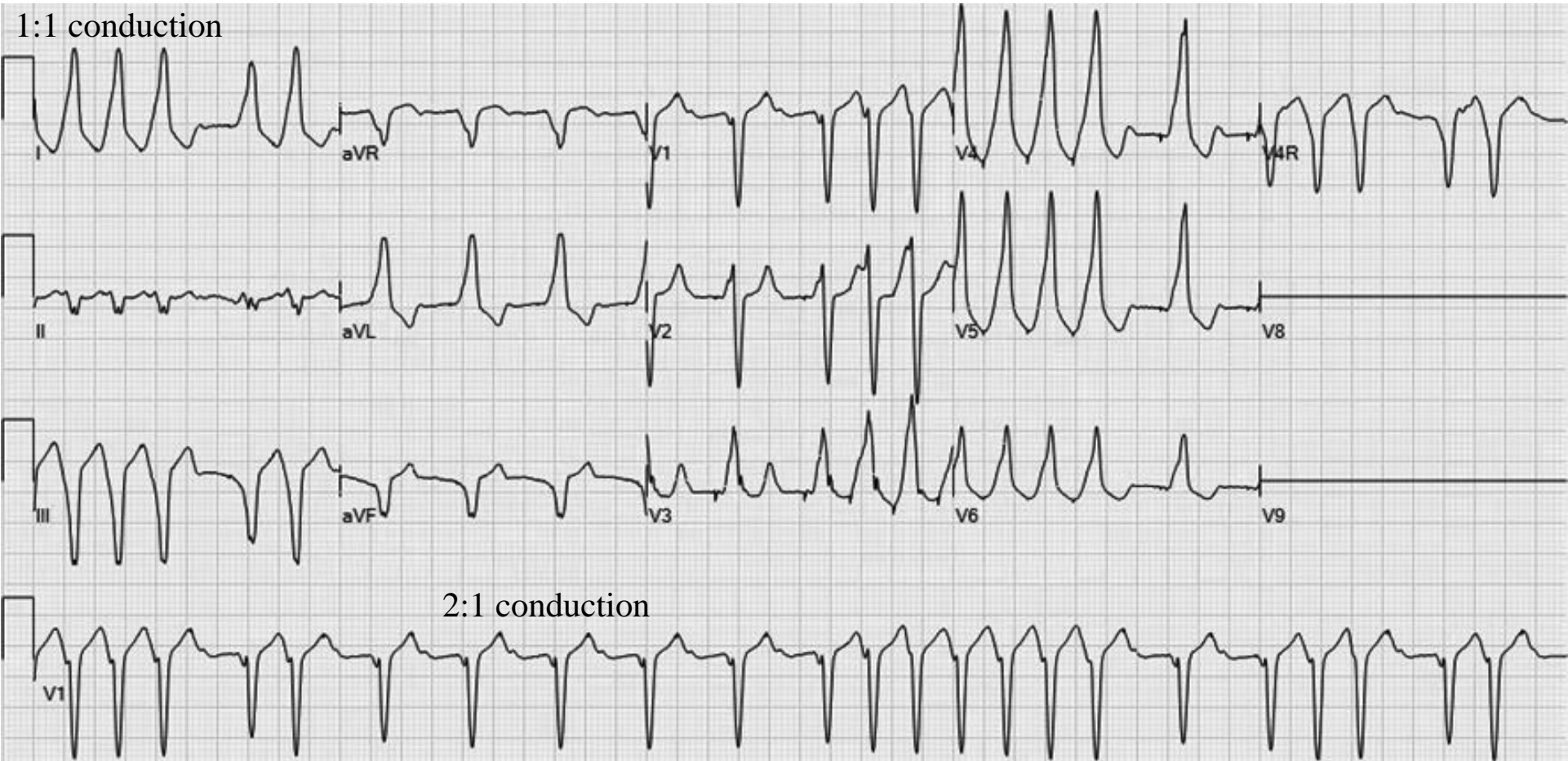


This trace is also very interesting since we actually see that the last QRS complex on the slides is "narrow" i.e. without preexcitation (arrow). This last tracing can be explained by either a) a "fatigue" of conduction in the AP; b) a bradycardic dependent or phase 4 block in that AP.

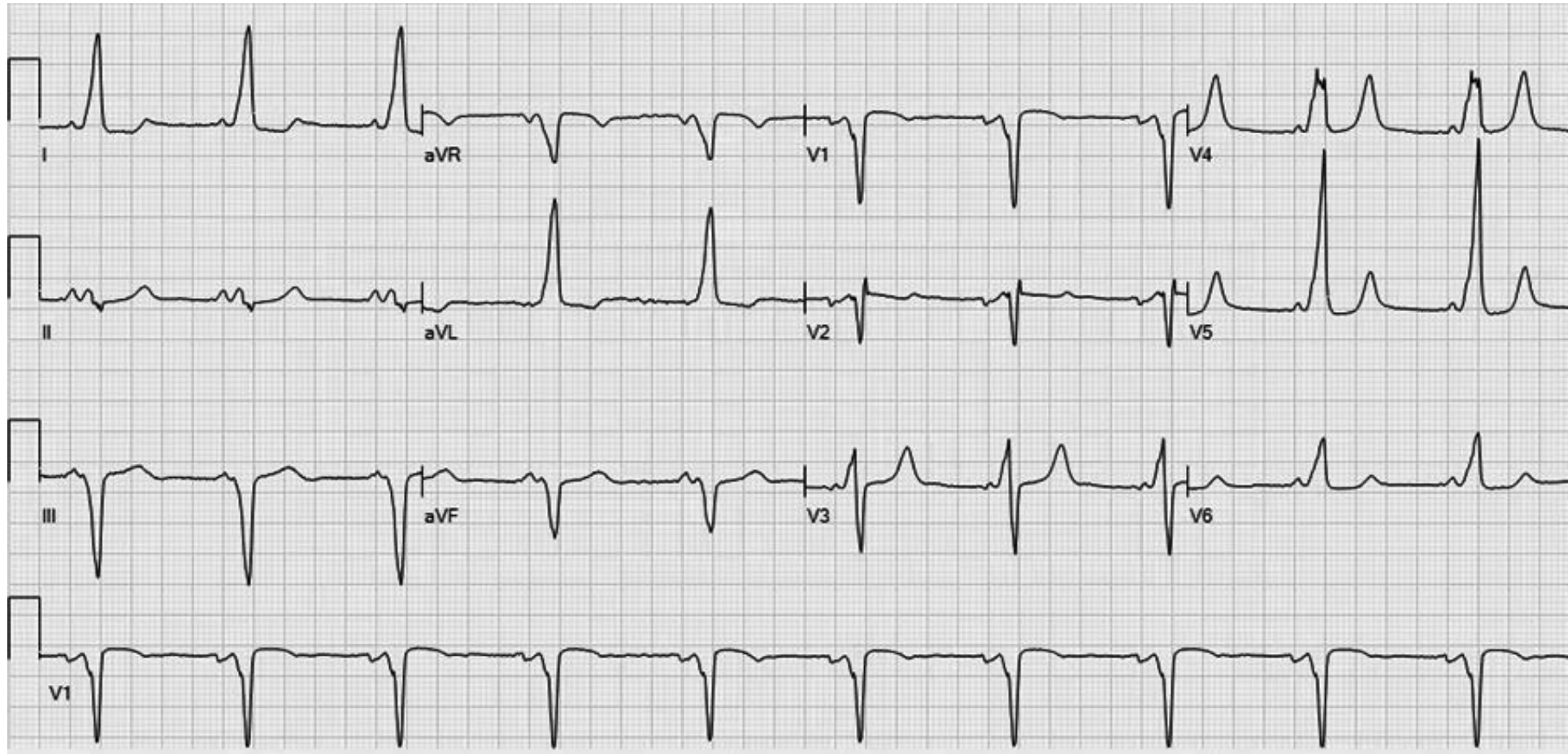
50 mm/sec!



1:1 conduction



This tracing is also interesting: we should explain why atrial pacing resumed 1:1 conduction over the AP after a period of 2:1 block; my feeling is that pacing reached the AP during its supernormal phase of conduction. The irregularity of QRS complexes could erroneously suggest pre-excited atrial fibrillation. See next slide a truly pre-excited AF.



WPW with right-side inferior paraseptal AP. See explanation of AP location in the following slides.

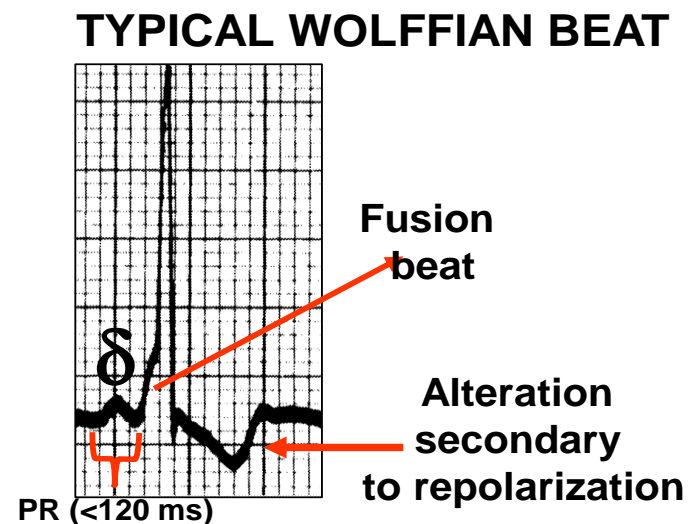
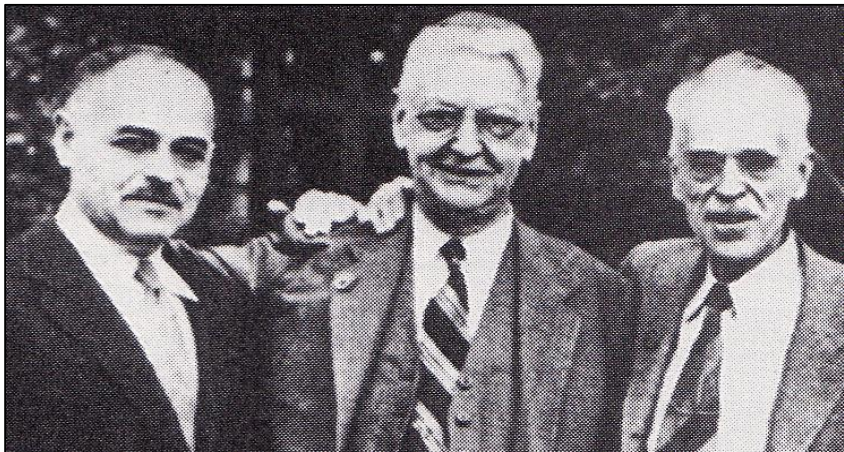
Historical Perspective

- **1876:** Paladino (**Paladino G. Contribuzone all anatomia ,istolgia e fisiologia del cuore. Mov Med-Chir (Napoli) 8:428, 1876.**) described numerous myocardial AV connections near the base of AV valves.
- **1883:** Gaskell (**Gaskell WH. On the Innervation of the Heart, with especial reference to the Heart of the Tortoise. J Physiol 1883;4:43-230.14.**) showed that atrial impulse spread to the ventricles by passing over the muscular connections that exist between the two chambers.
- **1893:** Kent (**Kent AF. Researches on the Structure and Function of the Mammalian Heart. J Physiol 1893;14:i2-254**) reported muscular connections between a rat's atria and ventricles, in the septum and in the right and left lateral walls. He pointed out that muscular connections were of two kinds: 1. direct continuity of the atrial and ventricular musculature at certain points; one of the points he specified was at the junction of the inter-atrial and inter-ventricular septa of the heart; and, 2. an intermediate continuity network of primitive fusiform muscular fibers that are embedded in the fibrous tissue of the AV rings of the heart. In the same year, His (**His W Jr. Die Tätigkeit des embryonalen Herzens und deren Bedeutung für die lehre von Herzbewegung beiss Erwachsenen . Ar med kiln leip 1893;14-60.**) described the AV bundle, which bears his name today, as the sole bridge between the atrial and ventricular myocardium. He put his discovery to the proof of experiment and showed that section of this bundle produced discordance in the contraction of atrium and ventricle. He thus concluded that Gaskell's observation was true — the atrial impulses pass to the ventricle by a muscular connection. He also noted the muscular continuity between the atrial and ventricles disappeared during development of mammalian hearts in all places except the junction of the atrium and ventricular septum, the point at which Kent also observed a connection.

- **1906:** Tawara, (**Tawara S. Das Reizleitungssystem des saugtierherzens. Jena, 1906.**) detailed the morphology of the atrioventricular bundle and its communication with Purkinje fibers distally and origin in AV node proximally in humans.
- **1907:** Keith and Flack, (**Keith A, Flack M. The Form and Nature of the Muscular Connections between the Primary Divisions of the Vertebrate Heart. J Anat Physiol 1907;41:172-89.**) described a ring of primitive conduction tissue encircling the atrioventricular junction.
- **1913:** Kent (**Kent AFS. Observations on the auriculo-ventricular junction of the mammalian heart. Q J Exp Physiol 1913;7:193-195.**) described the muscular connection between atrium and ventricle in the heart of man is not singular and confined to the AV bundle, but is multiple. He observed one point at which a muscular connection between atrium and ventricle exists, is situated at the right margin of the heart. He thus proposed, for purpose of identification, to refer to the connection described as the —right lateral connection. Cohn and Fraser (**Cohn AE, Fraser FR. Paroxysmal tachycardia and the effect of stimulation of the vagus nerve by pressure. Heart 1913;5:93-107**) provided the earliest description of two patients with paroxysmal supraventricular tachycardia, terminated by vagal stimulation. Both patients had ECG findings of short PR interval, an abnormal QRS, i.e., one patient with right bundle branch block pattern and the second with slurring of the initial portion of the QRS complex.
- **1914:** Kent, (**Kent AFS. The right lateral auriculo-ventricular junction of the heart. J Physiol 1914;48:17-24.**) (**Kent AFS. A conducting path between the right atrium and the external wall of the right ventricle in the heart of the mammal. J Physiol 1914;48:57.**) (**Kent AFS. Illustrations of the right lateral auriculo-ventricular junction in the heart. J Physiol 1914; 48:93-94.**) supporting his histologic

- findings of existence of a right lateral muscular connection between the atrium and the ventricle, provided evidence of its functional importance. He observed in an animal experiment that despite severance of all the structures that connect the atrium to the ventricles with the exception of a strip of tissue on the right lateral aspect of the organ, spontaneous beats arising in the atrium still conducted to the ventricle and evoked ventricular response. The severance described above passed through ventricular septum and the whole LV. He thus concluded that the transmitted beats only could have passed over the conducting path contained in the only part of the AV connection remaining, on the lateral wall on the right side of the heart.
- **1914:** Mines postulated circus movement rhythms on the basis of these multiple muscular connections and predicted their role as a mechanism of PSVT. (**Mines GR. On Circulating excitations in heart muscles and their possible relation to tachycardia and fibrillation. Trans R Soc Can 1914;8:43-52.**) Subsequently, several other cases were reported with publication of reports by Wilson, (**Wilson FN. A case in which the vagus influenced the form of the ventricular complex of the electrocardiogram. Ann Noninvasive Electrocardiol 2002;7(2):153-73.**).
- **1921:** Wedd (**Wedd AM. Paroxysmal tachycardia with reference to monotonic tachycardia and the role of the extrinsic cardiac nerves. Arch Int Med 1921;27:571-90.**) described Paroxysmal tachycardia with reference to monotonic tachycardia and the role of the extrinsic cardiac nerves.
- **1925:** Lewis (**Lewis T. The mechanism and graphic registration of the heartbeat. London, England: Shaw and Sons Ltd.; 1925.**) initially, and later several other researchers were unable to confirm his anatomic-histologic findings of a specific neuromuscular spindle in the right lateral wall.

- **1929:** Hamburger. (**Hamburger WW. Bundle branch block: Four cases of intraventricular block showing some interesting and unusual clinical features. Med Clinics North America 1929;13:342-62.**). Showed controversy surrounded Kent's description of multiple auriculo-ventricular connections, so called —Kent bundles. Initially, and later several other researchers were unable to confirm his anatomic-histologic findings of a specific neuromuscular spindle in the right lateral wall.
- **1930:** Wolff, Parkinson and White described surface ECG findings of short PR interval and RBBB pattern in patients with PSVT. (**Wolff L, Parkinson J and White PD. Bundle branch block with short PR interval in healthy young people prone to paroxysmal tachycardia. Am Heart J 1930;5:685-704**) heart. These authors described 11 young patients with no structural heart disease, with short PR, paroxysmal tachycardia runs associated to ECG pattern of branch block. Nevertheless, these features were not initially associated to electrical conduction by anomalous VA pathways (**Olgin, J., Douglas, L., & Zipes, P. (2012). Specific Arrhythmias: Diagnosis and Treatment. In D. P. Zipes, P. Libby, R. O. Bonow, D. L. Mann & G. F. Tomaselli (Eds.), Braunwald's heart disease: a textbook of cardiovascular medicine (pp. 785-836). Philadelphia: Elsevier Saunders.**



- **1932:** Holzman and Scherff (**Holzmann M, Scherf D. Uber elektrokardigramme mit verkurzter Vohof-kammer-Distanz und positive P.zacken. Z kiln Med 1932;121:404-23.**) attributed the ECG abnormalities described by Wolff, Parkinson and White to an abnormal AV connection bypassing the AV node and preexisting the ventricles and proposed a circus movement involving the multiple AV connections as a mechanism of tachycardia.
- **1933:** Wolfarth (**Wolfarth CC, Wood FC. The mechanism of production of short P-R intervals and prolonged QRS complexes in patients with presumably undamaged hearts: Hypothesis of an accessory pathway of auriculo-ventricular conduction (bundle of Kent). Am Heart J 1933;8:297.**)
- **1937:** Ivan Mahaim, (**Mahaim I, and Benatt A. Nouvelles Recherches sur les connections superieures de la Branche Gauche du faisceau de His-Tawara avec la cloison Interventriculaire. Cardiologia 1937;1:61-73.**) phrased —para-specific conduction, a term he used to describe properties of fibers directly connecting the lower portion of the atrioventricular node and the ventricular septum or between upper part of the bundle of His or each bundle branch and the ventricular septum or any part of the ventricle. In his communication, he opined that if conduction by Kent's fibers is accepted, it should be regarded as an accessory form of conduction: para-specific conduction. His original description of such conduction tracts have since been recognized historically by an eponym, as —Mahaim fibers.

- **1944:** Wood(**Wood FC, Wolferth CC, Gechler GD. Histological demonstration of accessory muscular connections between auricle and ventricle in a case of short PR interval and prolonged QRS complex. Am Heart J 1943;25:454.**) provided histologic proof of muscular connections between the right auricle and right ventricle on autopsy in a patient with WPW syndrome. Segers et al.(**Segers M, Lequime J, Denolin H. L'activation ventriculaire precoce de certain coeurs hyperexcitables etude de l'onde de l'electrocardiogramme Cardiolgia 1944;8:113-67**)proposed the term —delta wave for the initial slurred component of the QRS complex. Ohnell,(**Ohnell RF. Pre excitation, a cardiac abnormality. Acta Med Scan 1944;152(suppl):1-167**) coined the term —preexcitation, as a phenomena whereby, in relation to atrial events, the whole or part of the ventricular muscle is activated earlier by the impulse originating in the atrium than would be expected if the impulse reached the ventricles by way of normal conduction system. Kent's description of the presence of multiple muscular connections was denied by many investigators.
- **1961:** Meanwhile, James(**James TN. Morphology of the human atrioventricular node, with remarks pertinent to its electrophysiology. Am Heart J 1961;62:756-71.**) detailed distinct conduction pathways, separate from the AV myocardium, which included pathways connecting right and left atria and inter-nodal tracts connecting the sinus to the AV node. Per his description, a majority of these fibers enter at the superior margin of the AV node; a distinct subset bypasses the upper and central AV node, connecting directly with the lower third of the AV node or the bundle of His. He postulated that conduction over such a bypass tract would result in electrographic finding of a short PR interval, with resultant preexcitation, albeit with normal QRS duration, during sinus rhythm

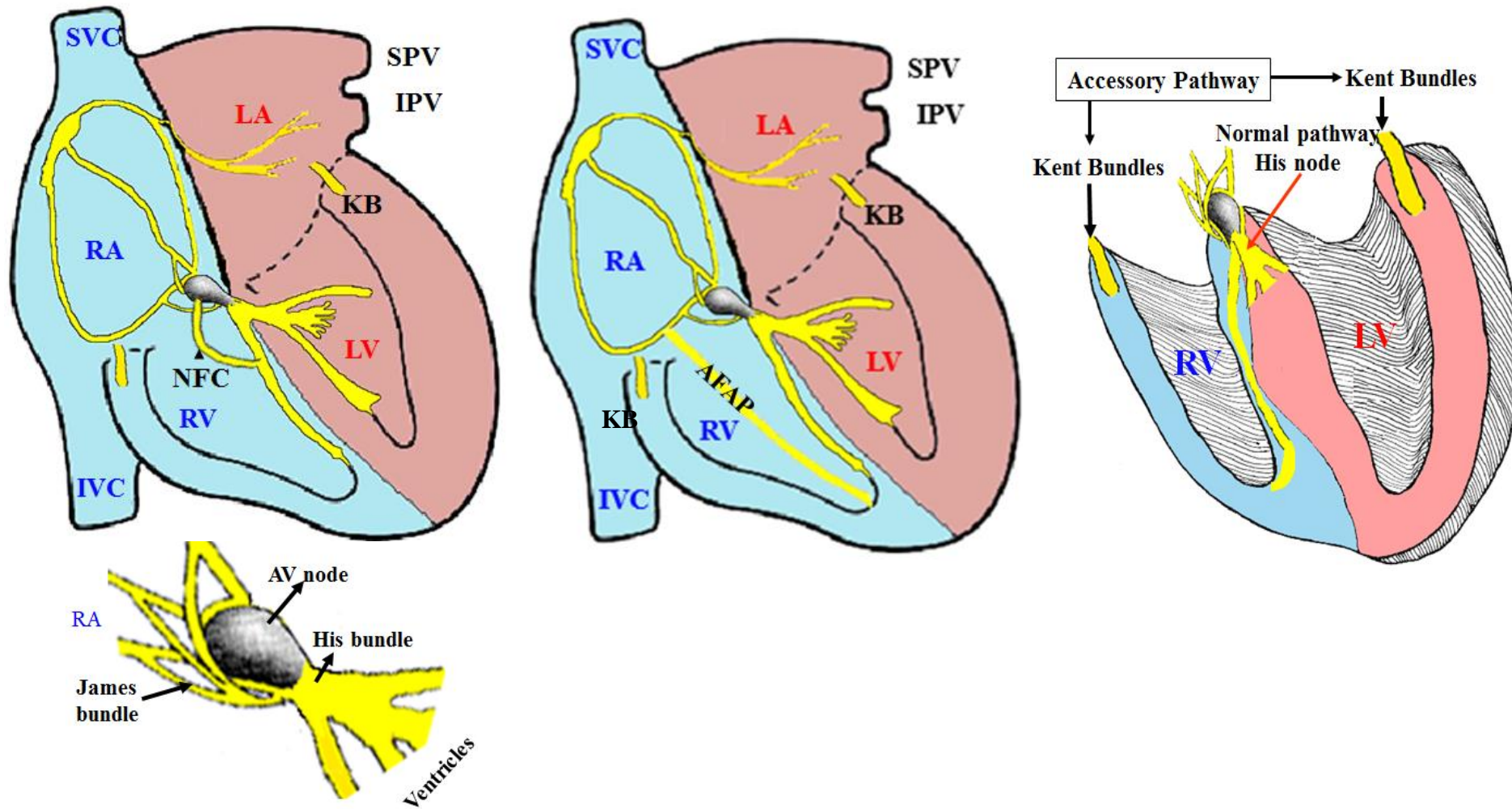
- **1966:** In a study of fetal and new born hearts, Lev and Lerner did not find any muscular communications outside the normal conduction system. (**Lev M. The Pre excitation Syndrome; Anatomic considerations of anomalous A-V Pathways. In: Dreifus, LS, Kolff WS, eds. Mechanisms and therapy of cardiac arrhythmias. New York, NY: Grune and Stratton, Inc; 1966: 665-670.**).
- **1972;1974/1978:** Although Kent's description of the presence of multiple muscular connections were denied by many investigators, it must be stated that another group of anatomists, i.e., Anderson and Becker . (**Anderson RH, Taylor IM. Development of atrioventricular specialized tissue in human heart. Br Heart J 1972;34:1205-14**)(**Anderson RH, Davies MJ, Becker A. Atrioventricular ring specialized tissue in the normal heart. Eur J Cardio 1974;2:219-30.**)(**Becker AE, Anderson RH, Durrer D, et al. The anatomical substrates of wolff-parkinson-white syndrome. A clinicopathologic correlation in seven patients. Circulation 1978;57:870-9**) were able to confirm Kent's description of specialized connections in the right atrial wall only. They, too, however, were unable to find multiple muscular connections across the AV annulus as postulated by Kent. The controversy and lack of proof of their existence, the original description of multiple muscular connections have been historically recognized by the eponym as "Kent bundle.
- **1975** Brechenmacher (**Brechenmacher C. Atrio-His bundle tracts. Br Heart J 1975;37:853-5..**) detailed distinct conduction pathways, separate from the AV myocardium, which included pathways connecting right and) described fibers in a patient with ECG finding of short PR interval and normal QRS duration, which bore no similarity to the ones described by James. Again, notwithstanding the lack of proof of their existence or functional significance, these connections have historically been described with the eponym as —James fibers. (**James TN. The Wolff-Parkinson-White syndrome: evolving concepts of its pathogenesis. Prog Cardiovasc Dis 1970;13:159-89.**).

➤ To circumvent the myriad of complexities involving the description of preexcitation syndromes, Anderson et al., (**Anderson RH, Becker AE, Brechenmacher C, et al. Ventricular preexcitation. A proposed nomenclature for its substrates. Eur J Cardiol 1975;3:27-36.**) they proposed a nomenclature suitable to both the anatomist and the clinicians. Central to the above was the concept that AV node is that portion of the cardiac tissue responsible for AV delay. For preexcitation to occur, it is necessary for the delay producing area, be either short-circuited, or modified by anatomic or physiologic changes. **The proposed classification. Accessory AV Muscle Bundle:**

Pathway connecting the atrial to ventricular myocardium outside the AV node-His-Purkinje system (the normal pathway). These were further subdivided into septal and parietal bundles: Right parietal connection was named as Type B preexcitation pattern and left parietal connection was named as Type A preexcitation pattern on surface ECG.

- **Accessory Nodovertricular Muscle bundle:** Pathway connecting the AV node directly to the ventricular myocardium, short-circuiting the distal/lower part of the AV node His-Purkinje system.
- **Atrio-Fascicular Bypass Tract:** Accessory pathway inserting into specialized tissues, producing preexcitation variant of short PR interval with a normal QRS duration.
- **Intra-nodal Bypass Tract:** Postulated as anatomically small and may not be functioning so as to produce normal delay.
- **Fascicular-Ventricular Accessory Connections:** Connecting specialized conduction system to the ventricular myocardium and may excite the ventricle earlier than would be via normal conduction route.
- **Accessory AV Pathways (AV-AP) WPW** remains the most common variety of preexcitation. Surface ECG appearance is due to conduction over an AV-AP connecting atrial and ventricular muscle directly bypassing the normal pathway, i.e., AV node and His-Purkinje system. The anatomical location of such pathways can vary between septal and free wall location on the right or left side of the heart.

Outline of anatomical substrate of ventricular preexcitation : Anomalous Pathways



SVC – Superior Vena Cava; **IVC** – Inferior Vena Cava; **RA** – Right Atrium; **LA** – Left Atrium; **RV**- Right Ventricle. **LV**- Left Ventricle. **SPV** – Superior Pulmonary Vein; **IPV** – Inferior Pulmonary Vein; **KB** – Kent Bundle.-**AFAP** – Atrio Fascicular Accessory Pathway (**AFAP**): Its proximal insertion is located on tricuspid annulus and its distal insertion in the RV apex Purkinje network. These fibers have decremental conduction properties but lesser than the AV node one.- **NFC** – Nodofascicular Connections They are bundles that connect any AV nodal area (usually N region) with Right Bundle Branch(**RBB**).