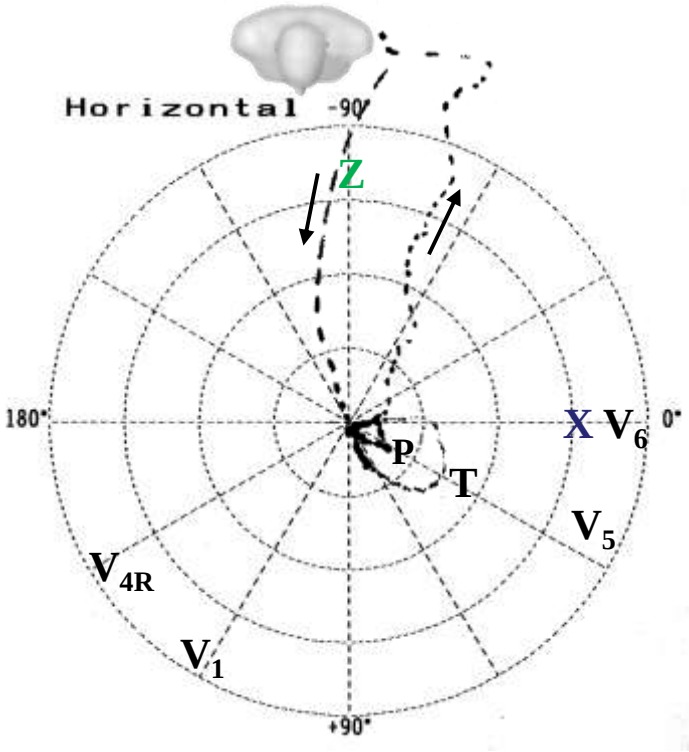
Diagnostic challenge with minimal data

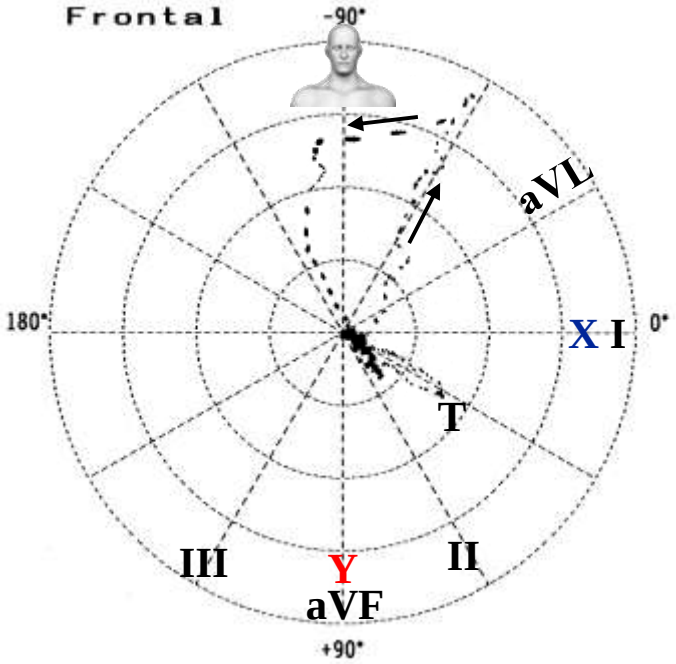
Adolescente masculino de 13 años con discreta cianosis y vectorcardiograma que hace el diagnóstico clínico obvio

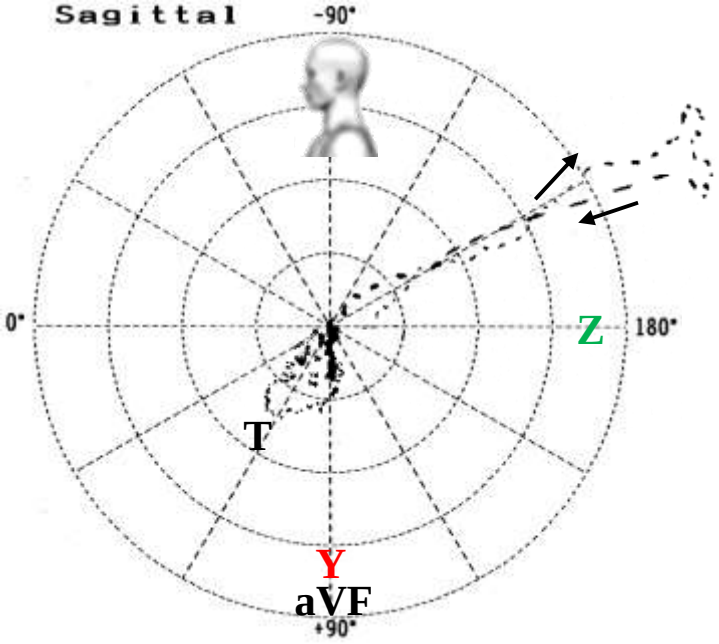
Adolescente masculino de 13 anos com discreta cianose e vetorcardiograma que faz o diagnóstico clínico obvio

A 13-year-old adolescent boy with mild cyanosis and vectorcardiogram that makes obvious the clinical diagnosis

Andrés & Raimundo







Existe neste caso uma tríade diagnóstica característica que permite o diagnóstico clínico. Qual é essa tríade? E qual é o diagnóstico clínico?

There is in this case a characteristic diagnostic triad that allows one clinical diagnosis. Which is this triad?..... and what is the clinical diagnosis?

En este caso existe una tríada diagnóstica característica que permite el diagnóstico clínico. ¿Cuál es esa tríada? ¿y cuál es el diagnóstico clínico?

Colleagues opinions

Hola Potro
El VCG muestra:
en PH agrandamiento de VI
en PF bloqueo de fascículo anterior izquierdo
en contexto de la edad y cianosis es sugestivo de cardiopatía congénita CIANOTICA tipo ATRESIA TRICUSPIDEA
Tríada característica de ATRESIA TRCUSPIDEA:HVI+ BFAI+CIANOSIS

abrazos
Juan Jose Sirena
Santiago del Estero, Argentina

Hi Andrés,
The ECG shows in the HP LVH, in the FP LAFB.
Cyanotic young patient with LAFB + LVH suggests tricuspid atresia.
Characteristic triad in tricuspid atresia: left ventricular hypertrophy+ Left Anterior Fascicular Block+ Cyanosis
Hugs
Juan José Sirena, MD

Querido Andrés:
El VCG me muestra un asa de P con pequeñas alteraciones pero no puedo decir que observo agrandamientos auriculares. Si no me equivoco en su rotación parece tener un ritmo sinusal. Veo un solo ventrículo agrandado y con trastorno de conducción. Yo pensaría en un ventrículo único.
Aunque no es el único diagnóstico que uno puede realizar con ese VCG.
El diagnóstico diferencial entre atresia tricúspidea y ventrículo único; es que en la primera se observa agrandamiento biauricular que yo no lo veo en el VCG. Si tiene aurículas grandes es una atresia tricuspidea sino lo dudo mucho.
Afectuosamente
Isabel, MD
Buenos Aires, Argentina

Dear Andrés:
The VCG shows a P loop with small alterations but I can not say that I see atrial enlargements. If I am not wrong in its rotation it seems to have a sinus rhythm. I see a single enlarged ventricle with conduction disturbance. I would think of a single ventricle. Although it is not the only diagnosis that one can make with this VCG.
The differential diagnosis between tricuspid atresia and single ventricle is that in the first biatrial enlargement is observed, that I do not see in the VCG. If you have large atria it is a tricuspid atresia but I very much doubt it.
Regards
Isabel **Konopka MD**
Division of Cardiology Ramos Mejía Hospital, Buenos Aires, Argentina

Final Conclusions by

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https://ekgvcg.wordpress.com

Raimundo **Barbosa-Barros**, MD
Centro Coronariano do Hospital de Messejana Dr. Carlos Alberto Studart Gomes, Fortaleza – CE-Brazil

Portuguese

Os colegas ao observar apenas um Vetorcardiograma (VCG) devem pensar que Andrés e Raimundo estão ficando loucos. Como eu poderia fazer o diagnóstico apenas com um VCG quando a maioria dos colegas entendem pouco ou nada de este método. Na verdade o colega não necessita entender de Vetorcardiograma, porque com apenas observar a alça QRS poderão perceber que na parte inicial da mesma as cometas ou lágrimas, se encontram muito mais próximas umas das outras em relação do resto da alça QRS o que assinala que a condução inicial da despolarização se processa mais lentamente fato praticamente exclusivo da pré-excitação ventricular. A seguir poderão deduzir que a via anômala se encontra a direita (entre o átrio direito e o ventrículo direito) o que fecha o diagnóstico. A tríade diagnóstica está presente: Cianose + pré-excitação ventricular tipo Wolff-Parkinson-White +via anômala de localização direita = anomalia de Ebstein.

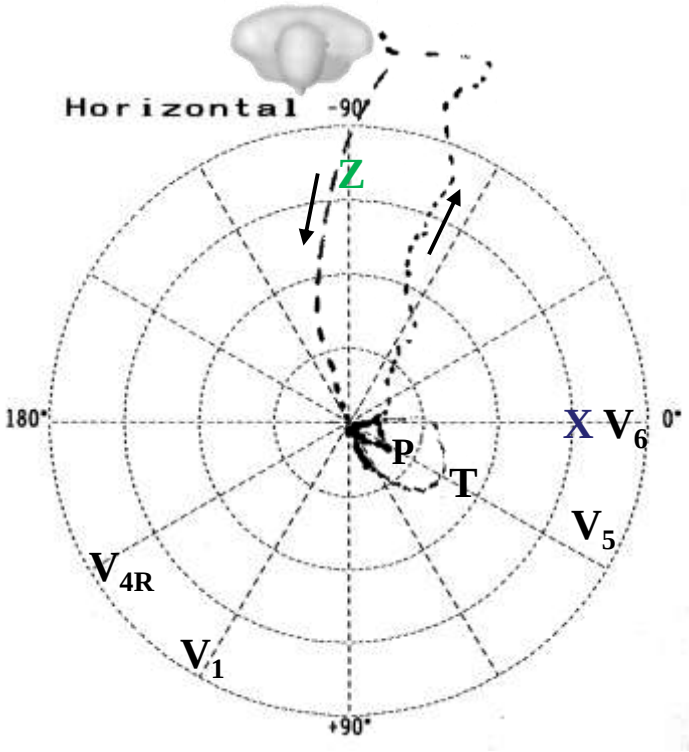
Spanish

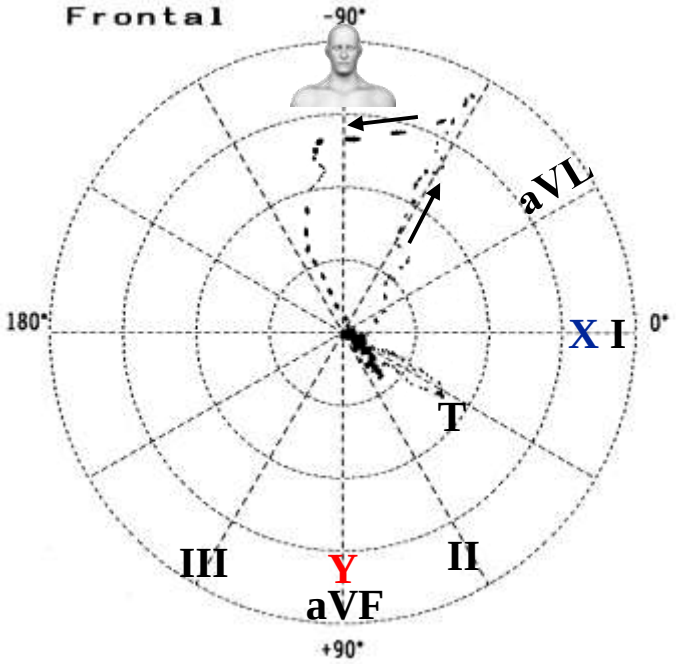
Los colegas al observar sólo un Vectorcardiograma (VCG) podrán pensar que Andrés y Raimundo se están volviendo locos. Como pueden pretender que hagan el diagnóstico apenas con un VCG si ellos son conscientes que la mayoría de los colegas entienden poco o nada de este método. En realidad, no necesitan entender de Vectorcardiograma, porque con apenas observar el bucle QRS percibirán que en la parte inicial las cometas o lágrimas, se encuentran mucho más cerca unas de otras en relación del resto del bucle QRS lo que señala que la velocidad de conducción inicial de la despolarización se procesa más lentamente que el resto del bucle hecho prácticamente exclusivo de la pre-excitación ventricular. A continuación, podrán deducir - por la localización y dirección de las fuerzas - que la vía anómala es derecha (entre la aurícula derecha y el ventrículo derecho) lo que cierra el diagnóstico. La tríada diagnóstica está presente

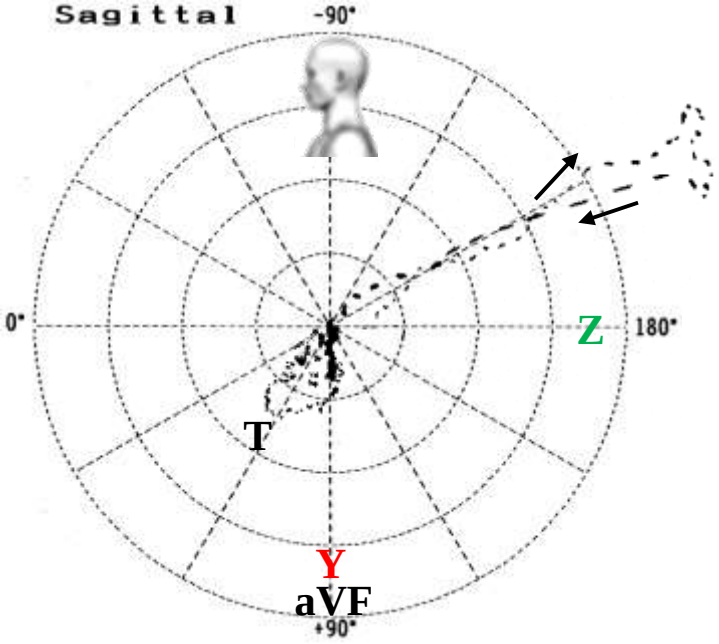
1) Cianosis +
2) Pre-excitación ventricular tipo Wolff-Parkinson-White +
3) Vía anómala de localización derecha = anomalía de Ebstein

English

Our colleagues, when observing only a Vectorcardiogram (VCG), may think that Andrés and Raimundo are going crazy. How can they imagine they can make a diagnosis just with a VCG, when they are aware that most of their colleagues understand little or nothing of this method? In fact, they do not need to understand about Vectorcadiograms, because just by observing the QRS loop you will perceive that in the initial part, the dashes are much closer together in comparison to the rest of the QRS loop, a fact virtually exclusive of ventricular preexcitation. Next, you may infer –considering the location and direction of forces- that the anomalous pathway is the right one (between the right atrium and the right ventricle), and thus the diagnosis is complete. The diagnostic triad is present: Cyanosis + Ventricular preexcitation of the Wolff-Parkinson-White type + Anomalous pathway of right location = Ebstein’s anomaly







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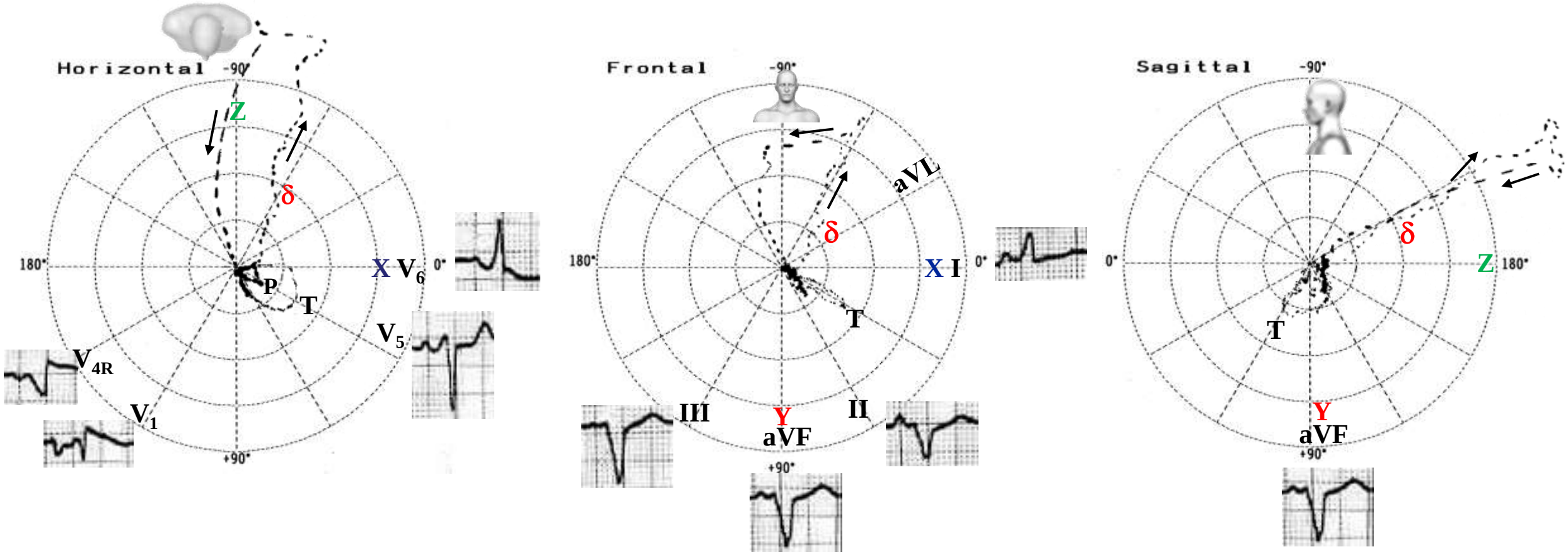
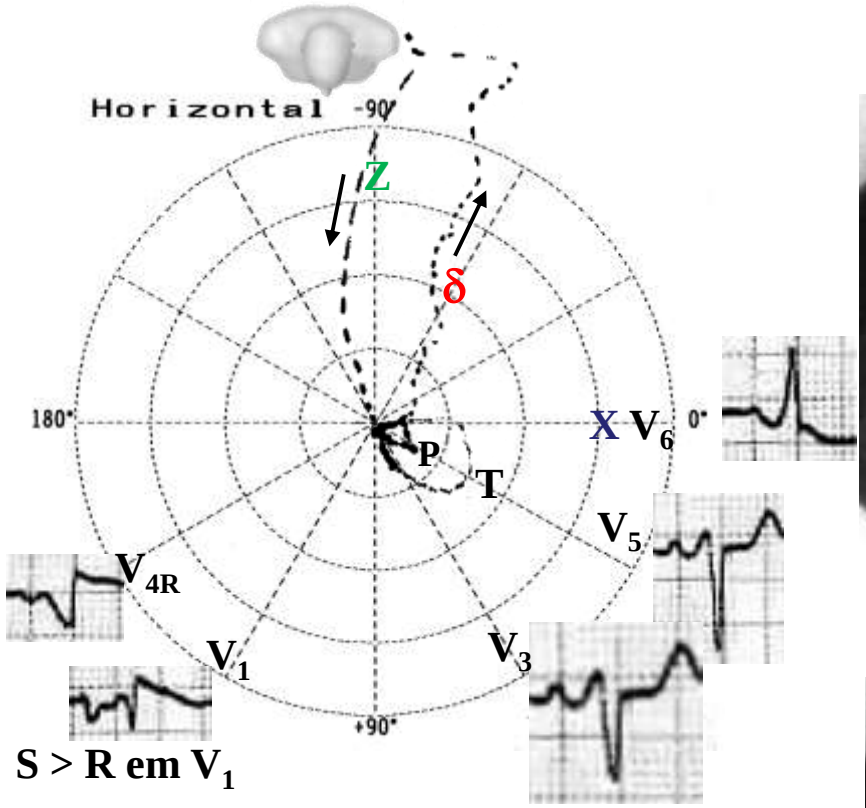
VCG in a Ebstein's anomaly and WPW syndrome type B

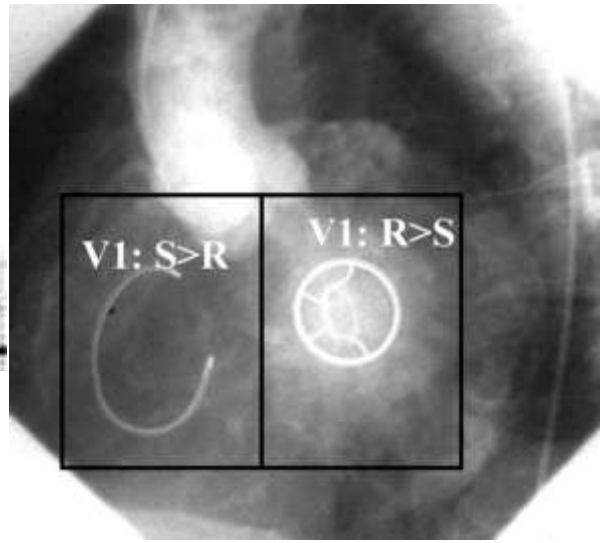


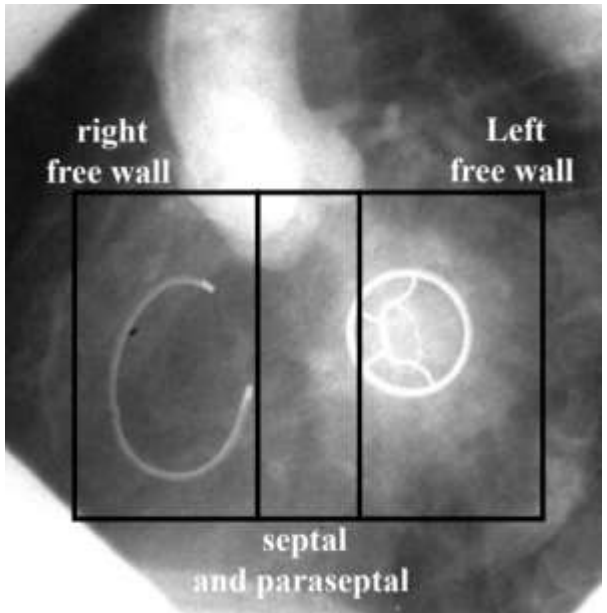
VCG of a 13-year-old boy with Ebstein's anomaly and WPW type B. This is always the type of pre-excitation found in WPW (**Lowe 1968**). Note the comets of the initial part of the QRS more closely together and erratic, indicating slow conduction on initial activation via the anomalous pathway(delta wave**δ**). In the PH the QRS loop is directed backwards (type B WPW). The electric axis with upper displacement in the PF. In absence of WPW the QRS axis is lower and the rotation is clockwise or in eight, unlike our case, that rotation is counterclockwise (**Ellison and Restieaux 1972**). S> R in V1 indicates that anomalous pathway (AP) is on the right side (100% of cases in Ebstein's anomaly). In addition, lower because the QRS complexes in II, III and aVF are predominantly negative. See the next slide……..

Is the anomalous pathway on the right or left side?
Answer: in the right side because S> R in V1



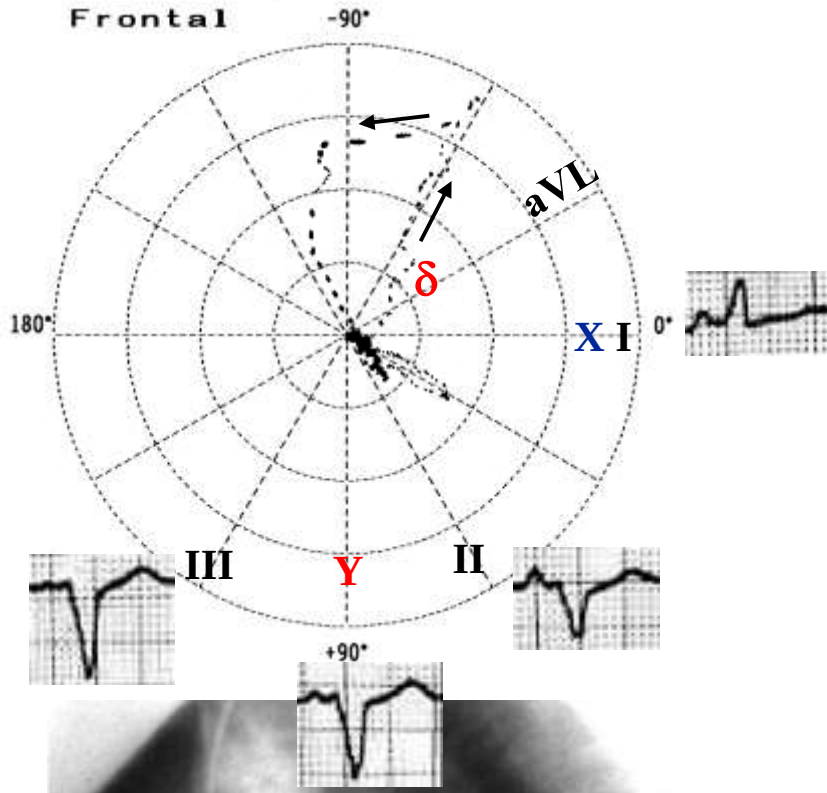
S > R em V_1

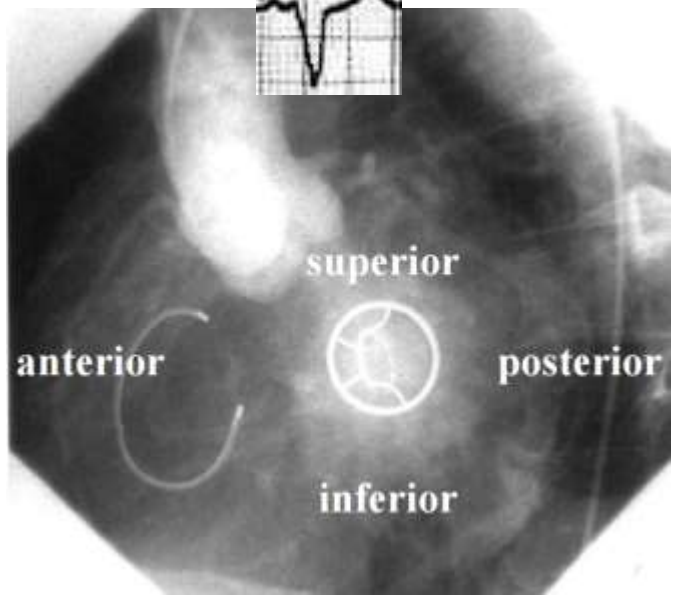




Ebstein´s anomaly is the only cyanotic congenital heart disease consistently associated with preexcitation, which is uniformly via a right accessory pathway.

The anomalous pathway is inferior because the QRS complexes are predominantly negative in the inferior leads





Determination of conduction velocity of stimulus

The greater or the lesser distance between dashes indicates the greater or the lesser conduction velocity in the area. Thus, when they are very close to each other, it indicates the presence of conduction delay. To consider the phenomenon as true, it is necessary for it to be evident in at least 2 planes.

Separate dashes = more dromotropism

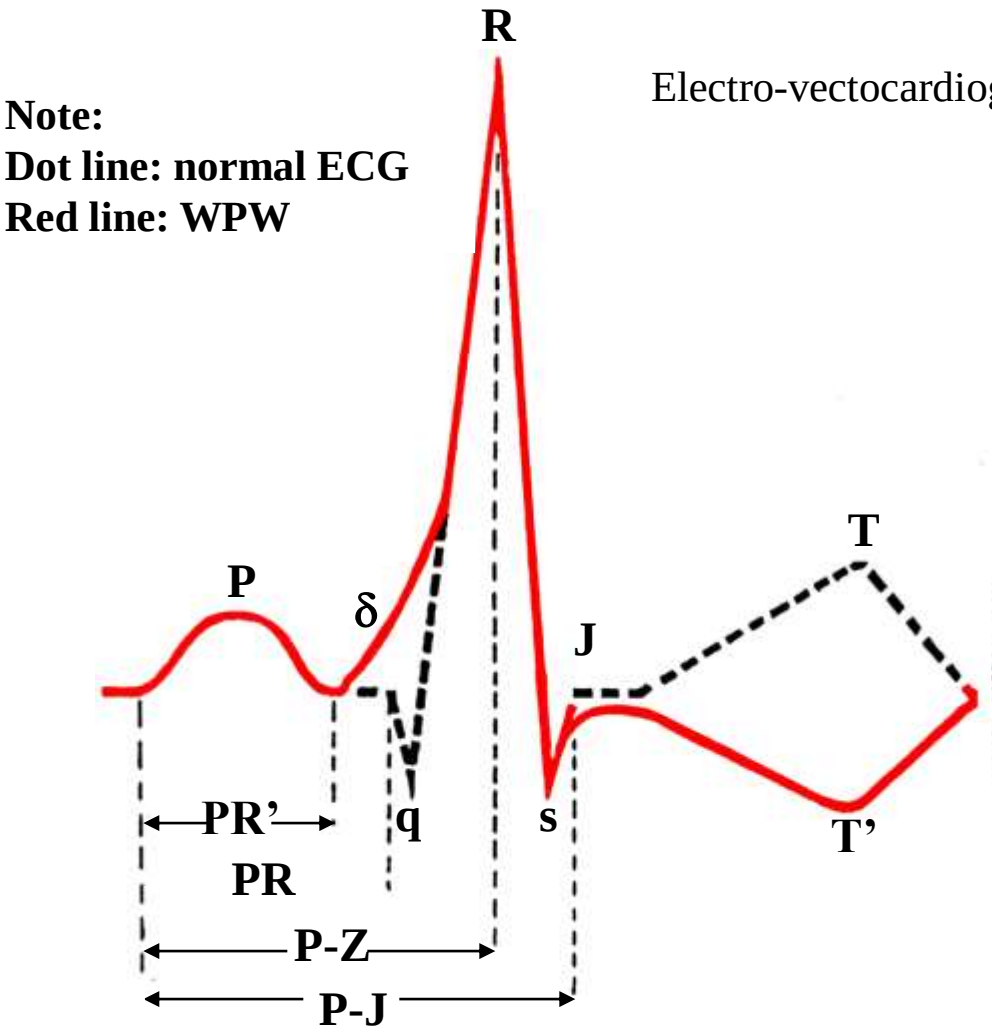
Very close dashes = less dromotropism

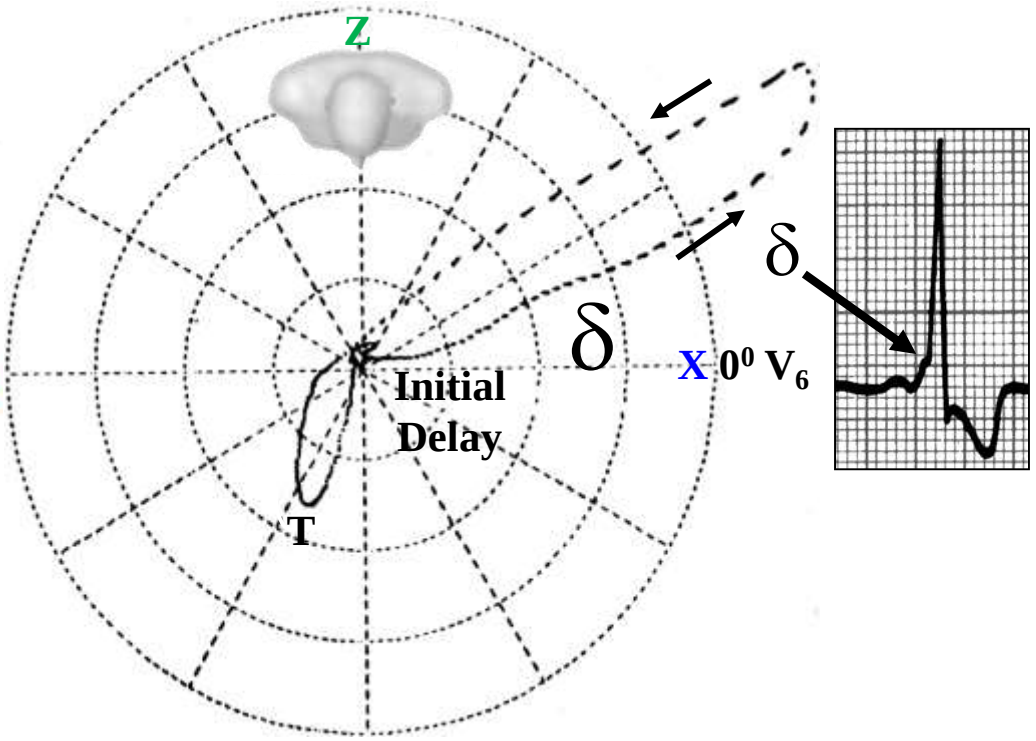
Low conduction velocity location in the QRS loop and its probable clinical diagnosis

Low conduction velocity location in the QRS loop	Clinical diagnosis
Initial delay	Ventricular preexcitation WPW type (δ wave)
Middle-final delay	LBBB
Final delay	RBBB
Total delay	Nonspecific intraventricular conduction defect

WPW ECG/VCG correlation

Electro-vectocardiographic criteria for WPW type preexcitation.

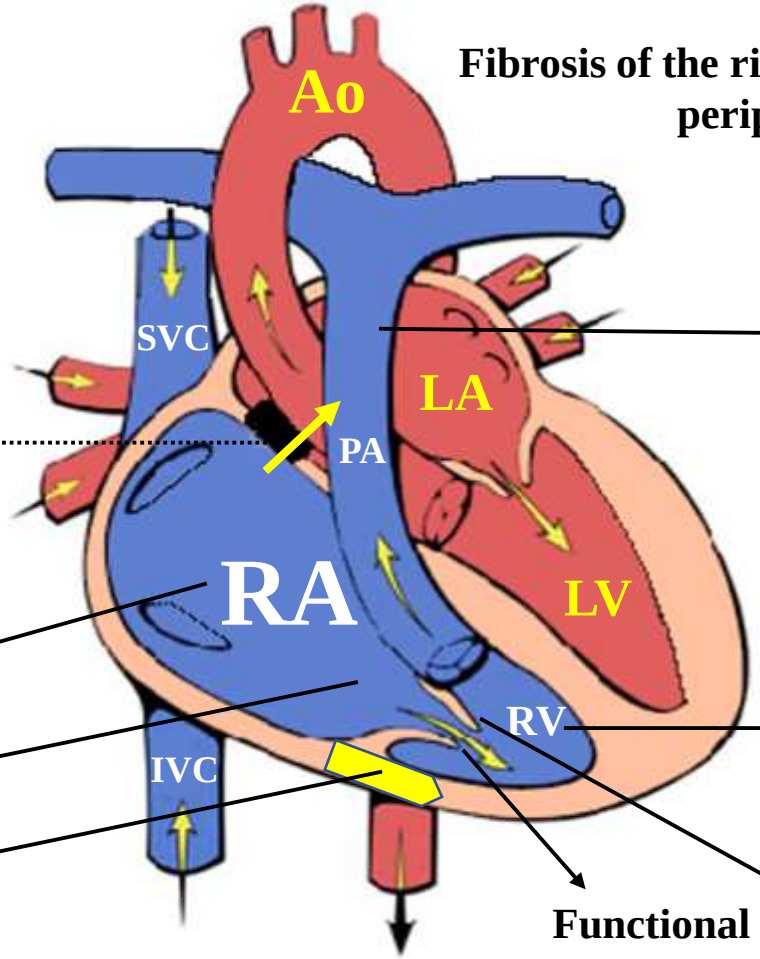




- **PRi or PQ:** since the onset of P up to the onset of QRS. It represents the time the stimulus takes to go from the SA node until reaching the ventricles: 120 ms to 200 ms.
- **PZ:** distance between P wave onset until R apex: 150 to 230 ms.
- **PJ:** distance between P wave onset until j point: 180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

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

Ebstein's anomaly of the tricuspid valve: Schematic of the anatomical and hemodynamic characteristics



Unilateral blindness

Brain abscess ← **Paradoxical embolization(rare)**

Interatrial right to left shunt ← **Atrial septal defect** (ASD) (50% associated) or patent foramen ovale (PFO)

Cianosis

Fibrosis of the right bundle branch, and / or malformations on peripheral portion of the right bundle

Anatomic pulmonary atresia/hypoplasia

Slow and progressive venous congestive heart failure

Small RV

Apical displacement of the tricuspid valve within the RV usually deformed

Tricuspid insufficiency

Functional integration of RV inlet into the RA (**Attenhofer 2007**)

Atrial arrhythmias (**Garcia Palmieri 1964**)

- **Mega RA**
- **Atrialized RV**
- **Accessory pathway**

Ventricular preexitation

PSVT either orthodromic (90%), antidromic (10%), AF (20%), atrial flutter, or ventricular fibrillation

Anatomically it is a rare and complex congenital heart disease characterized by: tricuspid apical displacement with dysplasia of septal and posterior leaflets, usually deformed that causes a small RV (**Danaraj 1968**), mega RA (atrialized RV), frequent tricuspid insufficiency, causing A patent v foramen or ASD with shunt between the RA and the LA. Ebstein's anomaly is a rare congenital heart disease with prevalence of one in about 20,000 live births (**Bjornard 2013**).

Prevalence of Congenital Heart Disease (Metropolitan Atlanta, 1978-2005) (Bjornard 2013)

Type of Congenital Heart Disease	Prevalence per 10 000 Live Births
All congenital heart diseases	67.7
Ventricular septal defect	Muscular: 14.2
	Perimembranous: 7.7
Atrial septal defect—secundum	6.1
Tetralogy of Fallot	4.6
Pulmonary valve stenosis	4.1
Patent ductus arteriosus	3.6
Ebstein anomaly	0.5 (about 20,000 live births)

Anatomic Classification of Ebstein's anomaly (Carpentier 1988)

Type A: adherence of septal and posterior leaflets without restrictive volume of functional right ventricle

Type B: right ventricle arterialized with normal anterior leaflet

Type C: stenotic anterior leaflet

Type D: arterialization of all the right ventricle except for a small infundibular component.

Carpentier et al. proposed a repair that used mobilization of the anterior leaflet of the tricuspid valve. For their types B and C, temporary detachment of the anterior leaflet and adjacent part of the posterior leaflet was followed by longitudinal plication of the atrialized ventricle and adjacent right atrium, repositioning of the anterior and posterior leaflets to cover the orifice area at the normal level, and remodeling and reinforcement of the tricuspid annulus with a prosthetic ring.

This repair was reported in 191 patients (mean ±SD age, 24±15 years) (**Chauvaud 2003**). The early mortality rate was 9%, and the mean late survival rate at 20 years was 82%±5%. It is unclear whether late problems will develop because of devitalized tricuspid valve tissue related to reattachment.

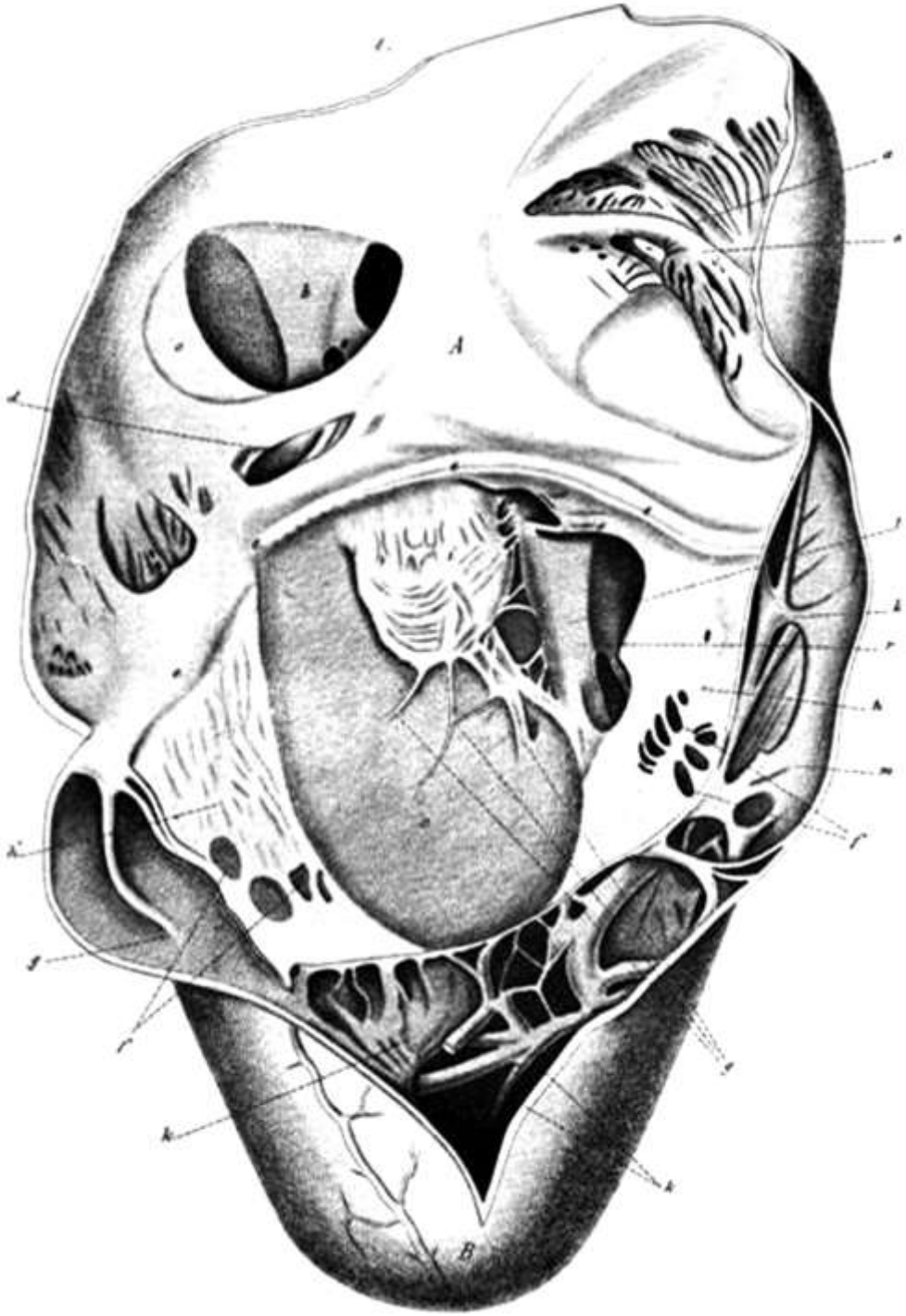
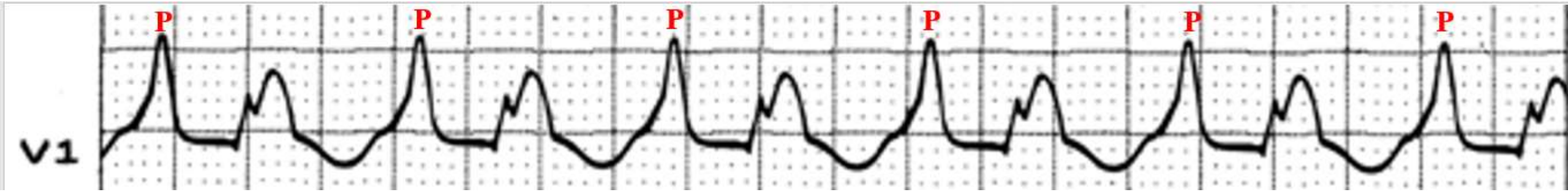


Figure from Ebstein's original case report. The RA and RV are shown opened along the right border beginning at the superior vena cava. A, Right atrium; B, right ventricle; b, valve; I, rudimentary septal leaflet of tricuspid valve with its chordae tendineae, which insert on the endocardium of the ventricular septum; r, opening through which one can get into the right conus arteriosus, and in the opposite direction, one can get into the sac that is formed by membrane h, h', and posterior part of endocardium of ventricular septum o. From Mann et al. (**Mann1979**).

Others cardiac malformations are often associated with Ebstein's anomaly, especially **atrial septal defect** or patent foramen oval (PFO). In utero, the reduced flow passing through the right ventricular outflow tract affects development of the pulmonary annulus, the main pulmonary arteries or its branches. It leads to an **anatomic pulmonary atresia** which differs from functional pulmonary atresia. Abnormalities affecting prognosis are those in the LV involving myocardium or valves in 39% of patients with Ebstein's anomaly (**Attenhofer 2005**). In addition to the right sided heart anomaly, several studies reported **systolic and diastolic dysfunction of the LV**. The change in right ventricular geometry can lead to anomalies in the size, function and shape of the LV, with paradoxical motion of the interventricular septum and regional contraction abnormalities. Moreover, LV interstitial fibrosis caused by arterial oxygen desaturation and right ventricle overload, leading to LV diastolic dysfunction, has been described in adults and neonates with Ebstein's anomaly (**Daliento 1997; Celermajer 1992**). Another intrinsic abnormality of left ventricular myocardium has recently been described: the **left ventricular non-compaction**. In affected patients, arrested myocardial morphogenesis results in excessively large trabeculations and intertrabecular recesses in the ventricular wall. This in turn can cause systolic and diastolic ventricular dysfunction, ventricular arrhythmias or systemic embolization.

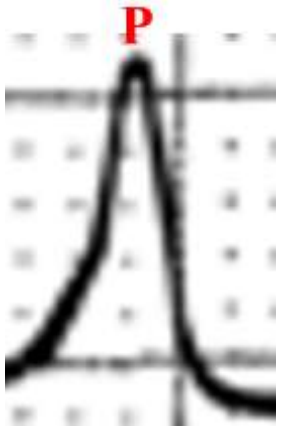
Main ECG features in Ebstein's anomaly

1. **P wave:** Giant P waves of RAE: "Himalayan" P waves (**Kaushik 2007**). P-waves are abnormal in height (tall peaked P-waves), duration and shape. Himalayan P waves are also observed in tricuspid atresia (**Reddy 2003**), combined tricuspid and pulmonic stenosis (**Davutoglu 2003**). This pattern has been ascribed to prolonged aberrant conduction in the enlarged right atrium (**Macruz 1968; Kastor 1975**). Ebstein's anomaly is a rare, complex, fascinating, congenital anomaly with a broad pathologic-anatomical and clinical spectrum accounting for <1% of all congenital heart defects. Since its description in 1866, dramatic advances in diagnosis have been made. Very high "Himalaya mountain-like" P waves (**Kaushik 2007**) are frequently observed. (The Himalayan mountain system are the planet's highest peaks around the world). P wave is >3 mm (0.3 mV) in close to 50% of cases (**Armengol 1996**). Tall P waves (≥2.5 mm) are attributable to right atriomegaly. A prolonged P-wave duration is occasionally registered (**Jaiyesimi 1982**).



Giant P waves of right atrial enlargement: "Himalayan" P waves, prolonged PR interval CRBBB of low voltage, bizarre aspect, initial Q wave in right and middle precordial leads (from V_1 to V_4) are recorded in 50% of the cases from V_1 to V_3.

Himalayan mountain

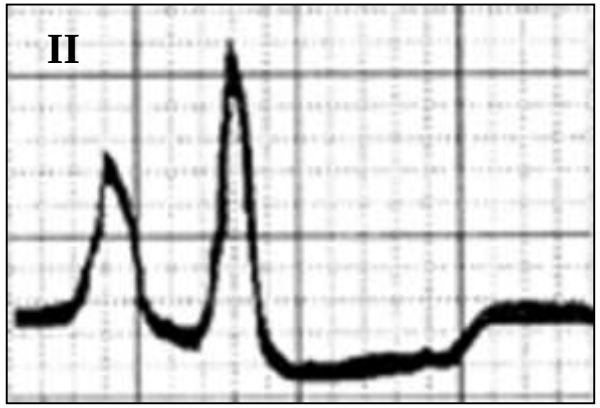


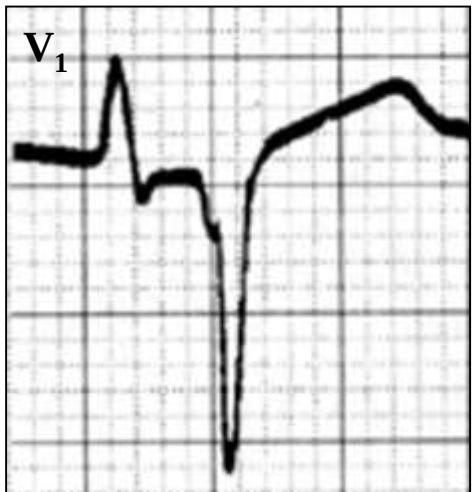
Himalayan" P-waves

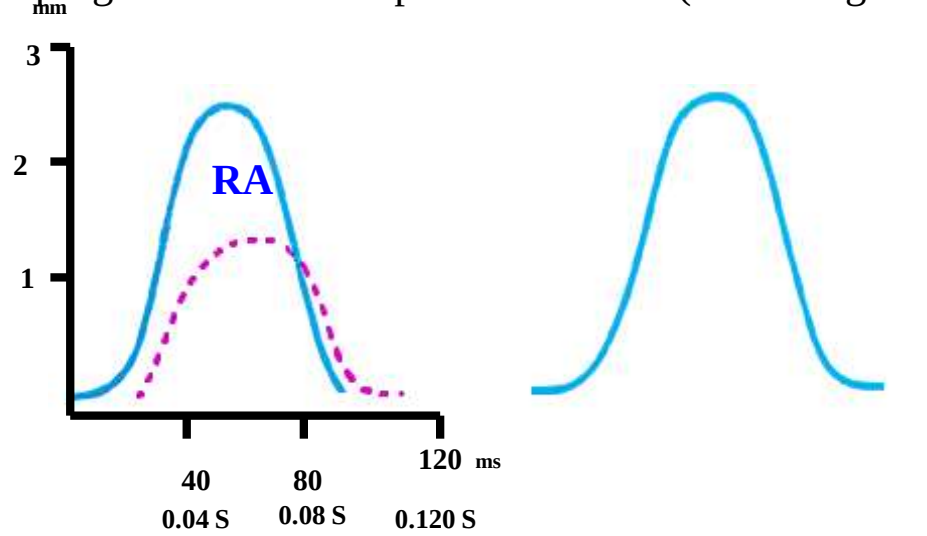
The “Gamboa P wave” (Gamboa 1966)

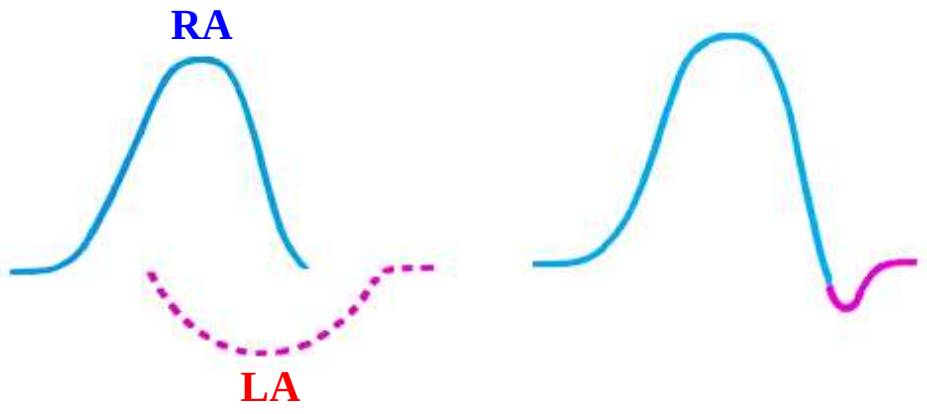
The association of:

1. Right atrial enlargement: The Gamboa P wave is very similar to Himalayan P wave. The P-waves are tall (>5 mm) and peaked in lead II. These types of P-waves are called also giant P-waves or Himalayan P-waves and are indicative of a RA dilatation due to a restrictive atrial communication.
2. Diastolic, volumetric or eccentric left ventricular hypertrophy. ECG with left axis axis deviation is observed in 94%.
3. Extreme left axis QRS deviation in the frontal plane: LAFB pattern.
4. Counterclockwise rotation of QRS loop in the frontal plane: LAFB pattern.
5. Cyanotic baby (neonate or infant). It is very suggestive of tricuspid atresia diagnosis.
6. LVH with adult pattern of QRS progression over the precordial leads (V1 through V6).









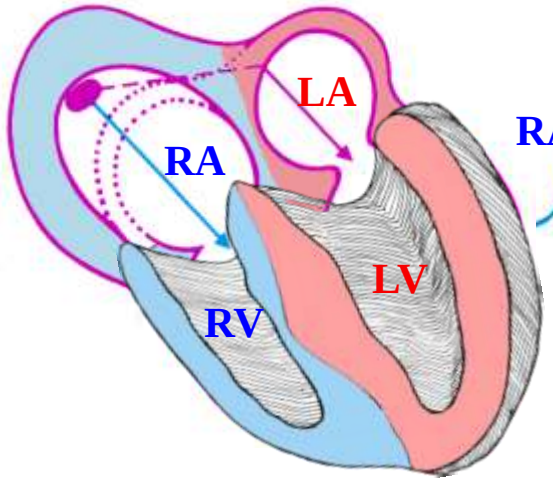
Possible etiologies of “Himalayan P waves

These types of P waves are called giant P waves or Himalayan P waves and are caused by reflected dilated right atrium resulting from pressure overloading.

Ebstein´s anomaly (**Taussing**)

1) Familial and associated with hypertrophic cardiomyopathy, extensive focal RA wall thickening and LV hypertrabeculation/noncompaction (**Stöllberger 2015**)
2) Hypertrophic cardiomyopathy(**Canpolat 2012**)
3) Tricuspid atresia(**Reddy 2013**) “Gamboa P wave”
4) Restrictive cardiomyopathy “Alpine P wave” (**Gupta 2012**)
5) Severe hypoxemia and emphysema (**Vijayakrishnan 2010**)
6) Combined tricuspid and pulmonic stenosis (**Davutoğlu 2003**).

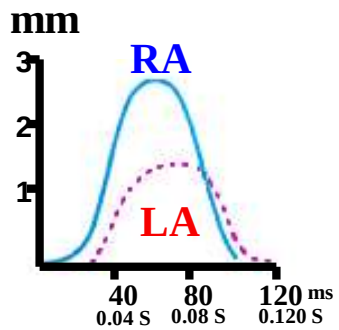
P-wave components in RAE

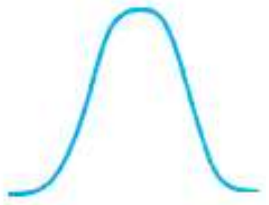


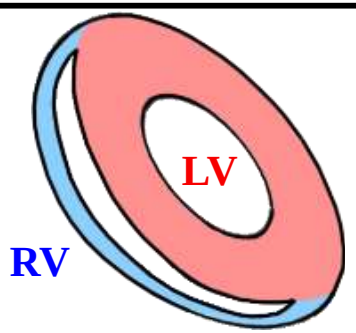
Normal

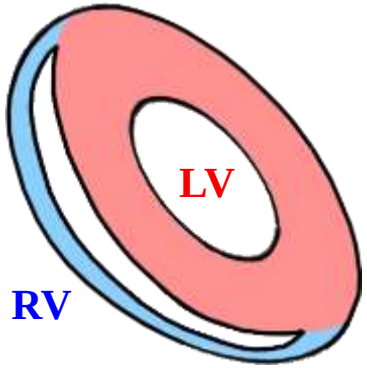
II

RAE

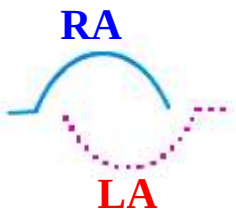






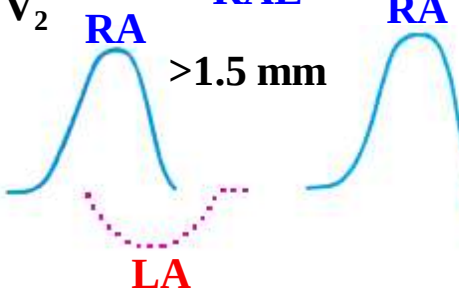


V_1

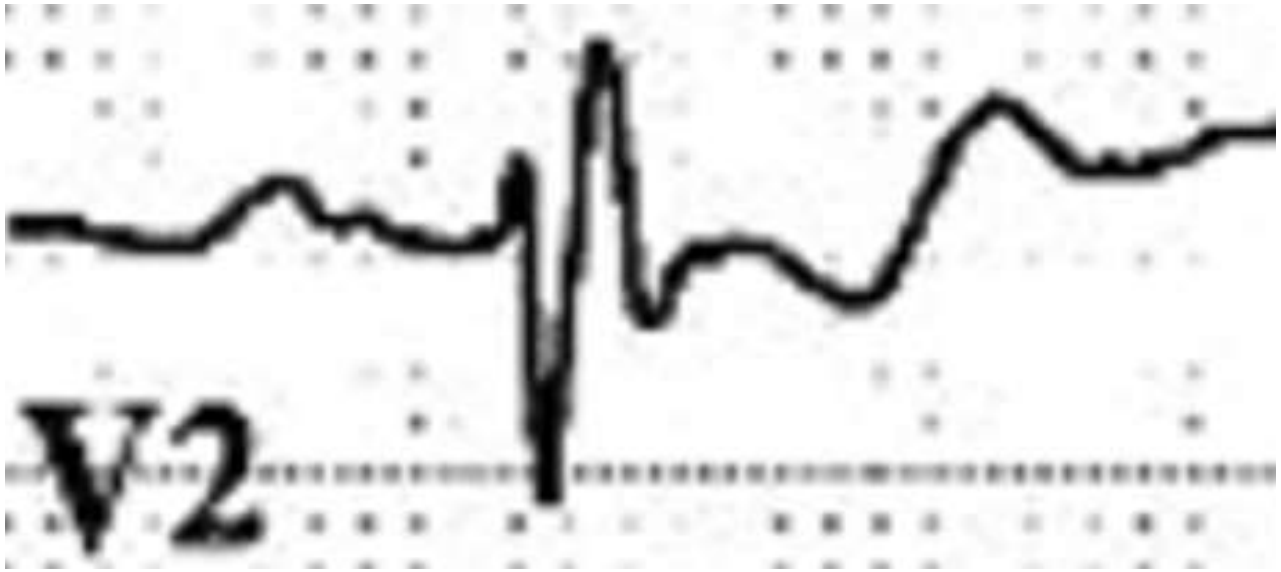


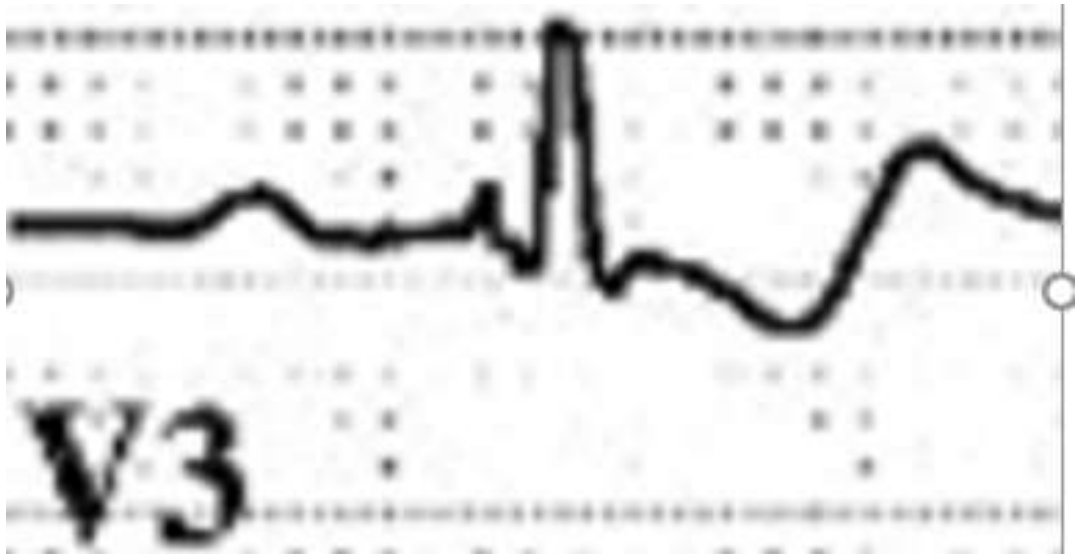
V_2

RAE



1. **PR interval** frequently prolonged: 20%. When WPW is associated the PR interval is usually short. A short PR interval occasionally occurs with δ wave and without a history of paroxysmal fast heart frequency. PR interval duration and with of the P wave correlate with prolonged conduction in the large right atrium (**Makous 1966**). First degree AV block occurs in 42% of patients (**Ho 2000**).
2. **QRS axis ÂQRS:** generally inferior and to the right between +90 and +130º but the range varies widely from -30° to -170°. The QRS axis is inferior, although a splintered polyphasic QRS makes the axis difficult to determine. When associated with WPW the QRS axis is located in the left superior quadrant.
3. **IRBBB or CRBBB** with abnormally low R (> 7mm) and S waves over the right precordium, bizarre morphologies of the terminal QRS pattern result from infra-Hisian conduction disturbance and abnormal activation of the atrialized RV (**Hebe 2000**) initial Q wave in right and middle precordial leads (from V_1 to V_4) are recorded in 50% of the cases in V_1 to V_3. It is frequent to record tri- or tetraphasic patterns. RBBB is observed in ≅85% of cases. QRS prolongation is the result of prolonged activation of atrialized right ventricle and is less fully manifest in infants. The conduction disturbance is always distal to the right bundle branch (**Kastor 1975**) and is sometimes present despite a septal AP. The AV node may be compressed and the central fibrous body abnormally formed. The right bundle branch may be abnormal or show marked fibrosis (or both) (**Christine 2007**).

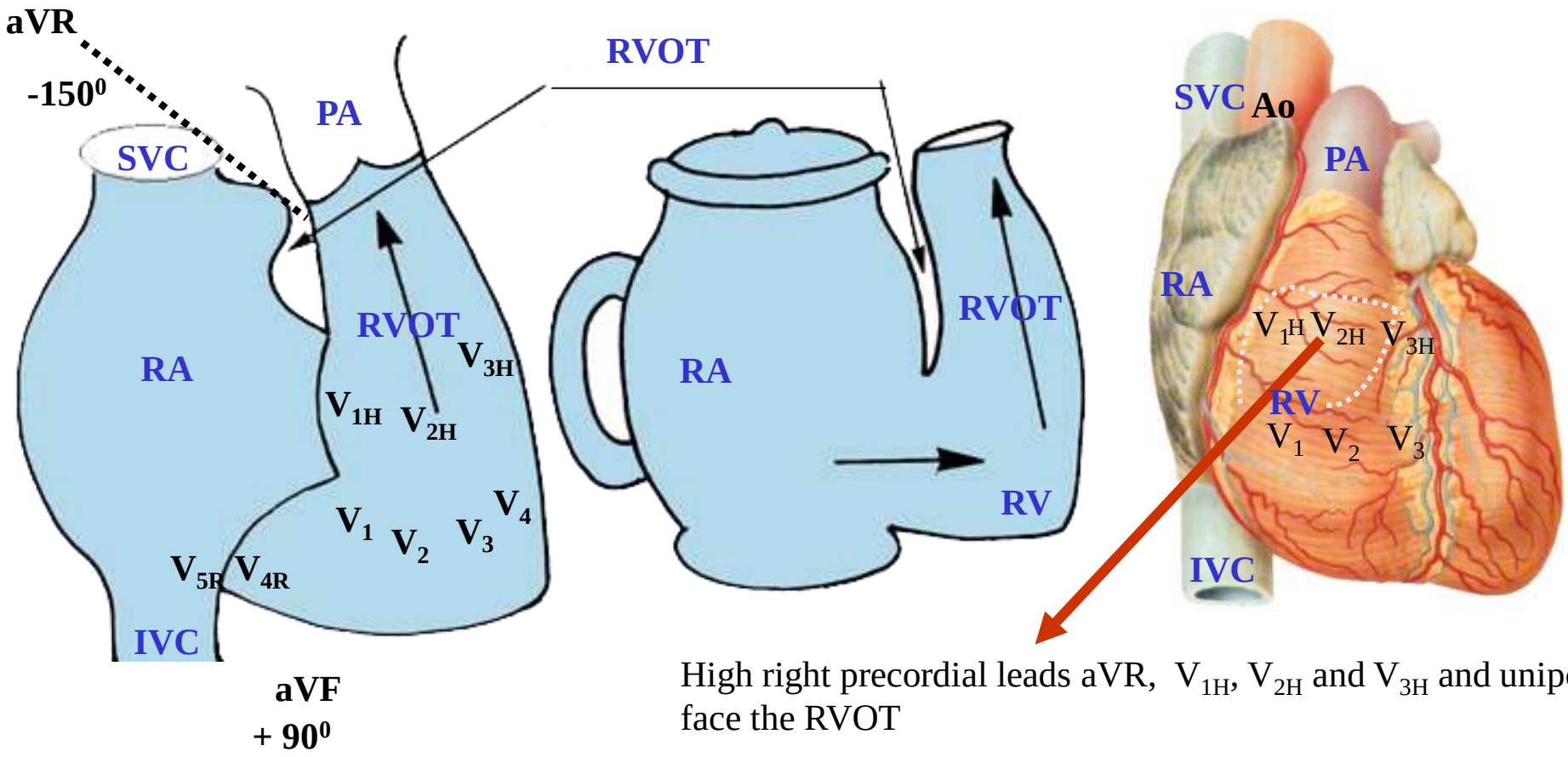




When tetaphasic QRS pattern in registered in right ventricular trabecular region(V_2 and V_3) Ebstein's anomaly is one diagnostic possibility

RVH classification by the RV region predominantly hypertrophied and it correspondent lead

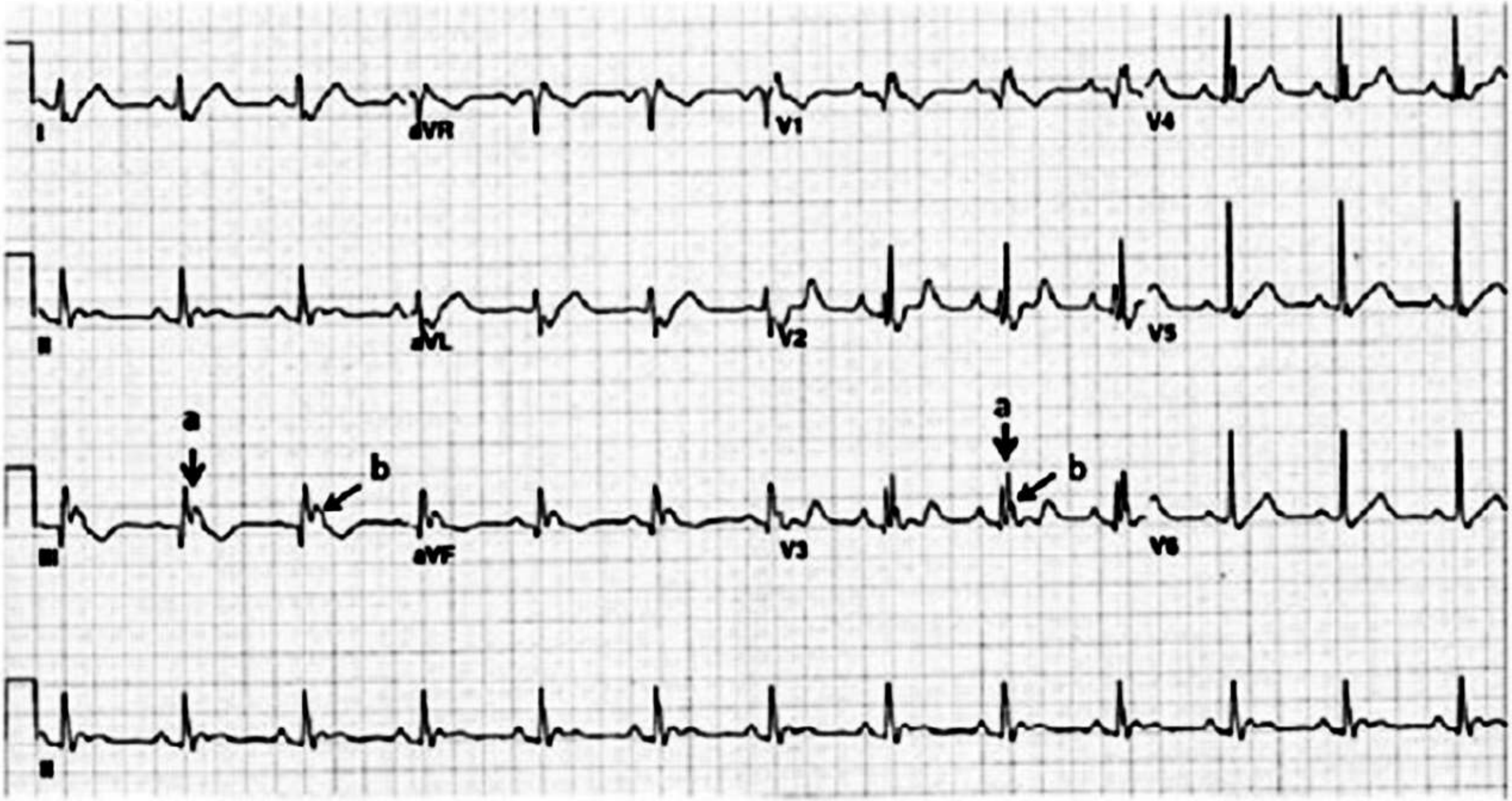
- Right ventricular trabecular region: V_2 and V_3. When tetaphasic Ebstestein's anomaly is one diagnostic possibility
- Inferior right paraseptal region: V_3 and V_4.
- Right ventricular free wall: from V1 to V_4.
- Basal infundibular region, right ventricular outflow tract (RVOT) or crista supraventricularis: aVR, V_{1H}, V_{2H} and V_{3H}
- Right Ventricular Inflow Tract (RVIT): V_{4R}, V_{5R} and aVF.



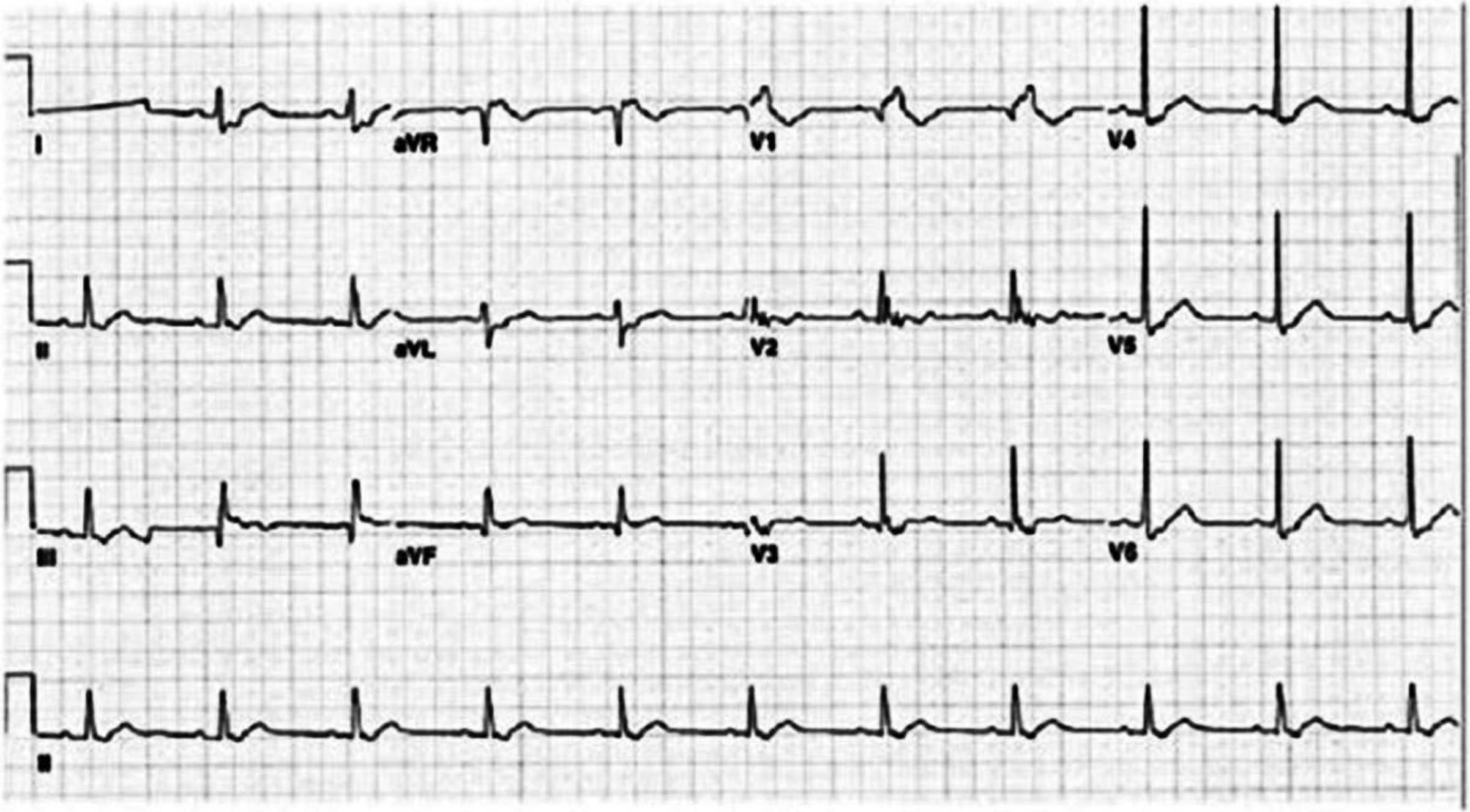
High right precordial leads aVR, V_{1H}, V_{2H} and V_{3H} and unipolar aVR face the RVOT

5. **A distinctive second QRS complex attached to the preceding normal QRS complex** originates in the atrialized right ventricle according to intracardiac mapping (**Kastor 1975**). Possible type B WPW with accessory pathway located in the RV free wall (anomalous bundle between the RA and the RV). **Ebstein´s anomaly is the only cyanotic congenital heart disease consistently associated with preexcitation, which is uniformly via a right accessory pathway.** Ventricular preexcitation can be permanent or intermittent and can occur without δ wave and δ waves can occur with normal PR interval. When APs are multiple, as they are in more than a third of patients of Ebstein's anomaly, conduction tend to be via the septal pathway (**Smith 1982**). Mahaim nodoventricular fibers are likely to be present when a LBBB pattern occurs during sinus rhythm or during an episode of tachycardia (**Smith 1982**). Accessory atrioventricular pathways are revealed in 6–30% of patients with Ebstein's anomaly, and can lead to both supraventricular and ventricular tachyarrhythmias. The most common supraventricular tachyarrhythmias are AP mediated reciprocating tachycardia, atrioventricular nodal re-entrant tachycardia, and atrial flutter or AF. In the case of APs, ventricular pre-excitation is obvious in only 60% of patients with Ebstein's anomaly and symptomatic tachyarrhythmias. In other patients without ventricular preexcitation, the absence of RBBB seems to be a strong predictor of APs, and becomes visible after RFCA. Tachyarrhythmias cause palpitation, syncope or sometimes SD and are more poorly tolerated, since hemodynamic and anatomic abnormalities are associated with Ebstein's anomaly. APs are located on the right side or in the posteroseptal region. Electrophysiologic mapping and RFCA remain difficult before tricuspid valve repair or replacement because of atrial dilatation, which disrupts anatomic landmarks and makes it difficult to find and target the atrioventricular junction with a catheter. Accessory pathways are often multiple, meet in a broad band, and abnormal endocardial activation potentials can often confound their identification.

6. **QRS fragmentation or fragmented QRS :** Abnormal conduction through the atrialized RV leads to QRS fragmentation on ECG. Its presence suggests a more severe abnormality and a higher risk of arrhythmia. The fQRS disappears after corrective surgery with resection of the atrialized RV(**Acharya 2017**). Fragmented QRS (fQRS) is a convenient marker of myocardial scar evaluated by 12-lead electrocardiogram (ECG) recording. fQRS is defined as additional spikes within the QRS complex. In patients with CAD, fQRS was associated with myocardial scar detected by single photon emission tomography and was a predictor of cardiac events. fQRS was also a predictor of mortality and arrhythmic events in patients with reduced left ventricular function. The usefulness of fQRS for detecting myocardial scar and for identifying high-risk patients has been expanded to various cardiac diseases, such as cardiac sarcoidosis, arrhythmogenic right ventricular cardiomyopathy, acute coronary syndrome, Brugada syndrome, and acquired long QT syndrome.

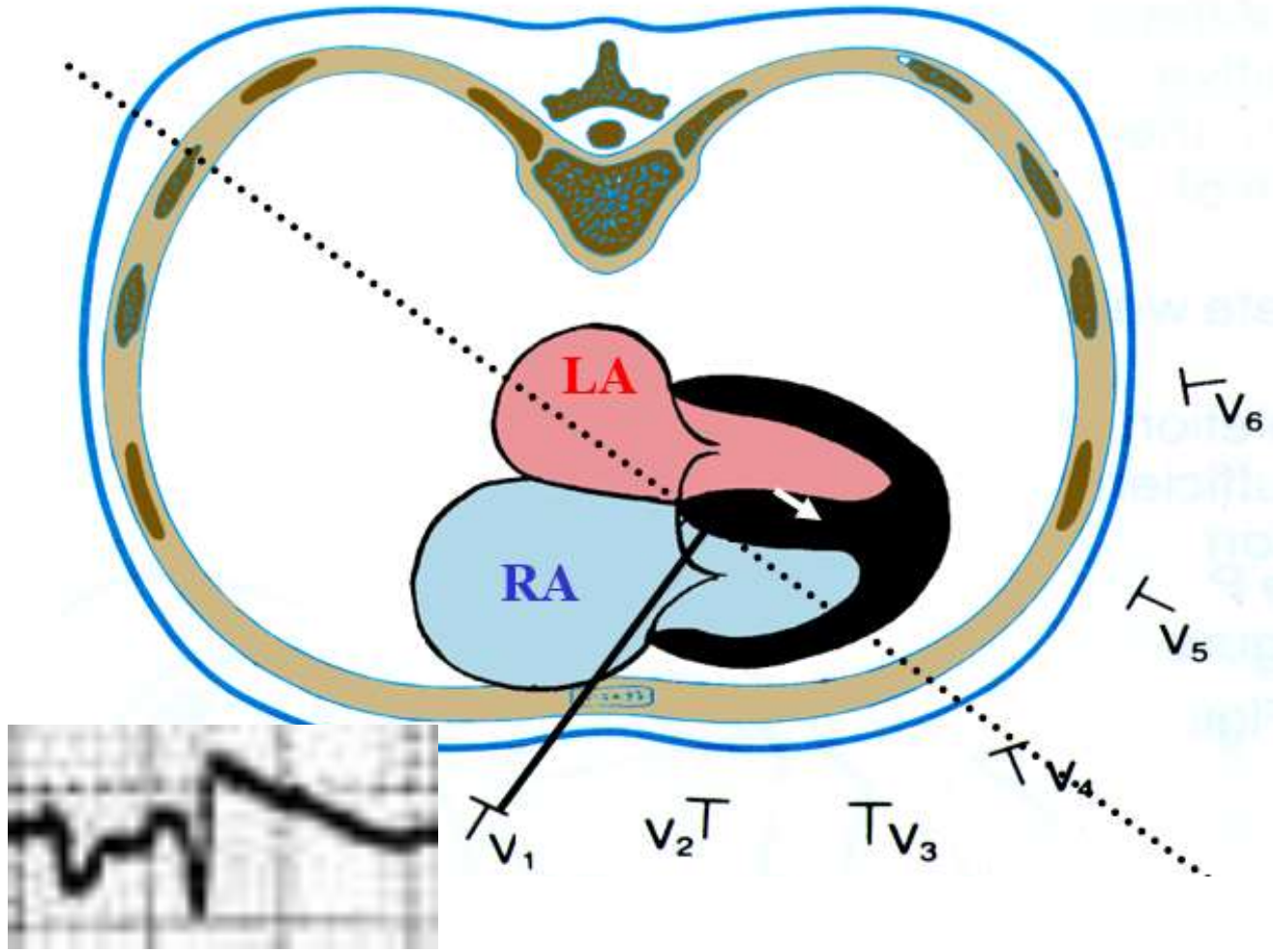


Initial ECG showing QRS fragmentation (a, fragmentation of QRS; b, broad positive deflection [R′] representing a second QRS).



ECG after corrective surgery showing absence of QRS fragmentation.

7. **Deep Q waves** are registered in inferior leads but most important are high precordial Q waves in leads V1 or V1 to V4. These distinctive Q waves occurs the precordial surface leads record right ventricular intracavitary potentials and usually far leftward as a result of large size of right atrium (**Sodi-Pallares 1952, Bialostozky 1972**).



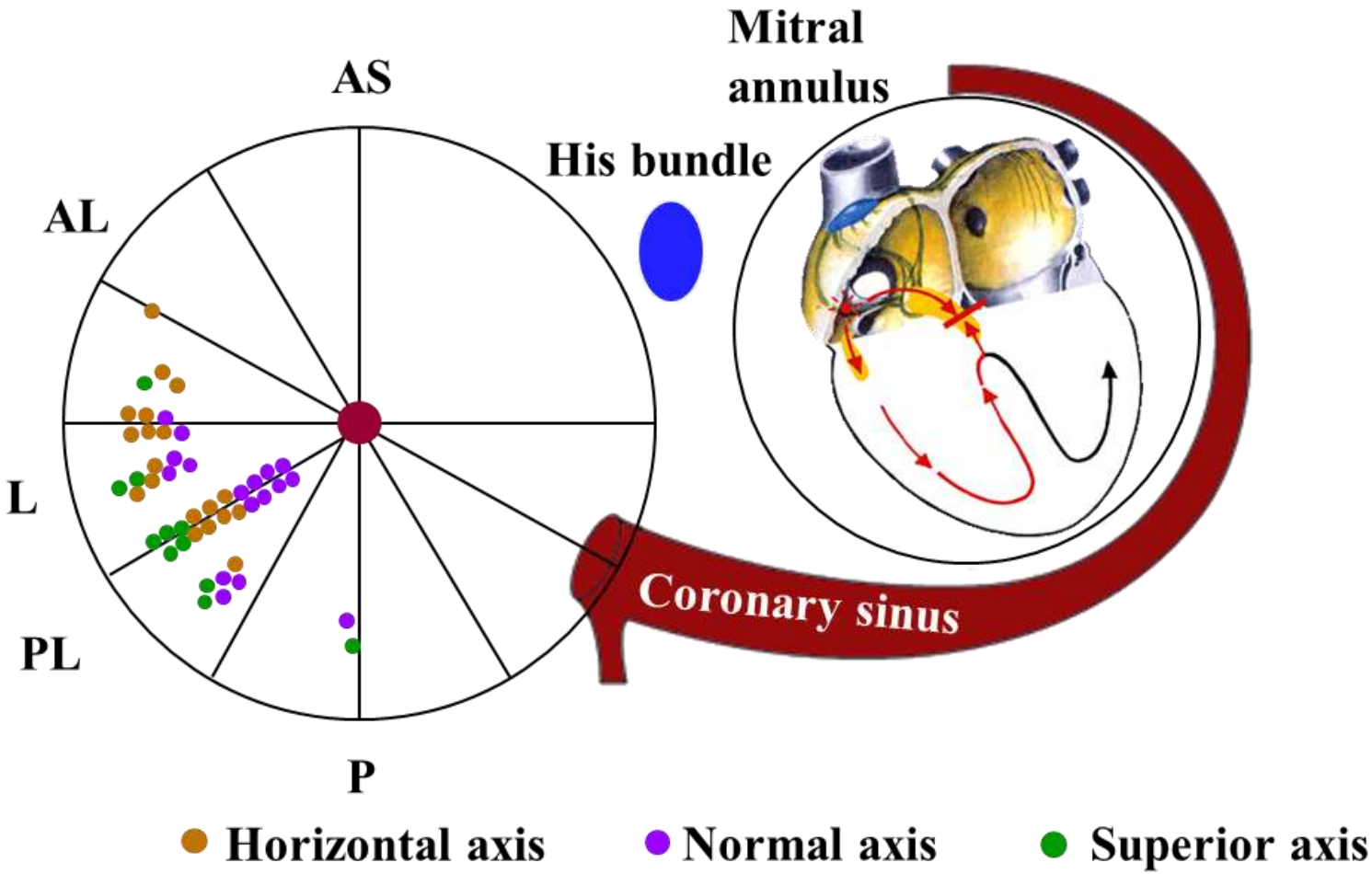
Outline that explains the indirect sign of RAE: qR in V_1 (**Sodi-Pallares sign**). The volumetric increase of the RA, gets closer to the exploring electrode V_1, recording initial QRS negativity in this lead, because this electrode records the epicardial morphology of the RA (**Sodi Pallares 1952**).

8. **Arrhythmias** In ~30% of the cases, atrial fibrillation, flutter, supraventricular and ventricular paroxysmal tachycardia (orthodromic or antidromic) are registered. Permanent atrial standstill has been reported in familial Ebstein's anomaly. **Atrial standstill:** It is characterized by a responsive to electrical or mechanical stimuli of atrial myocardium. The P waves are absent in both scalar ECG and transesophageal ECG (**Piérard 1985**). Persistent atrial standstill occurred in a father and his youngest son with familial Ebstein's anomaly. In both cases routine ECGs showed no atrial electrical activity and cross sectional echocardiograms showed inferior displacement of the septal tricuspid leaflet and tethering of the tricuspid leaflets to the right ventricle. The father had a cerebral embolism and died of a myocardial infarction. Necropsy showed attachment of the septal tricuspid leaflet below the membranous septum. On serial histological examination fibrofatty infiltration of the right atrial wall, the atrioventricular node, and the inferior part of the His bundle up to the bifurcation was present. The son had hemodynamic and electrophysiological findings consistent with mild Ebstein's anomaly and persistent atrial standstill, for which permanent cardiac pacing was necessary. The persistent atrial standstill with slow escape rhythm was most probably a consequence of the abnormalities in both the atrial wall and the His bundle which, together with the abnormal attachment of the tricuspid valve, may be features of the same congenital cardiac anomaly. Carballal et al. presented a case of atrial standstill "atrial paralysis" associated with atrio-ventricular block. The patient presented with atypical chest pain and a symptomatic bradycardia of 37 beats per minute. A VVIR pacemaker was implanted. She has subsequently been symptom free (**Carballal 2002**). **Mahaim fibers:** The term Mahaim type conduction is used to describe decremental conduction connections between the right atrium (RA) or between the AV node and the RV or near the right branch. Although these VAs are rare, their unique electrophysiological properties make their diagnosis and treatment complicated. The true nature of these still obscure pathways make them particularly interesting from the electrophysiological point of view (**Katritsis 2016**). In 1941, Mahaim and Winston described the histology of abnormal connections originating from the AV node with RV distal insertion (**Mahaim 1941**). This was the first description of the Mahaim Node-Ventricular Pathways "Mahaim Accessory Pathways" (MAPs). Later, mapping the atria mainly to the lateral part of the tricuspid ring, the term atrio-fascicular was adopted. Ueshima et al. report a case of a 7-year-old girl with atriofascicular Mahaim (AFM) pathway concomitant with Ebstein's anomaly. The QRS wave showed LBBB pattern on ECG. Holter showed prolongation of the PR interval and QRS morphological change during sinus tachycardia. An electrophysiological study demonstrated that the distal His potential appeared earlier than the proximal His potential, which suggested retrograde His conduction toward the AV-node. Conduction from the Mahaim fiber to the His bundle was faster than that from the AV-node towards the His bundle. The findings allowed a differential diagnosis between AFM and Wolff-Parkinson-White (WPW) syndrome (**Ueshima 2017**). Ebstein's anomaly is often accompanied by either WPW syndrome or AFM. Therefore, it is important to differentiate between them with regards to treatment and associated risks. AFM pathway indicates AP that demonstrate decremental conduction property without ventriculoatrial conduction (**Tchou 1988**).

8. AFM usually causes antidromic atrioventricular reciprocating tachycardia, although the risk of ventricular high-frequency reply at the time of the AF is lower than that in WPW syndrome. Because of shortened PR interval and prolonged QRS interval determined by ECG, it is important to differentiate AFM from WPW syndrome. AFM has been reported to account for approximately 3% of all the accessory pathways. Differential diagnosis between AFM and WPW syndrome is very important because both of them cause antidromic atrioventricular reciprocating tachycardia. One of the points to consider differentiating AFM from WPW syndrome is the change of PR interval. In WPW syndrome, PR interval during sinus rhythm is stable because APs do not exhibit decremental conduction property. Meanwhile in AFM, both PR interval and QRS waveform often change due to decremental conduction property in the Mahaim fiber. In this case, the Holter electrocardiogram demonstrated a change in the PR interval and narrowing of the QRS complex, which supported the diagnosis of AFM rather than WPW syndrome. Sternik et al. reported that 61% of AFM patients showed rS pattern QRS waveforms in lead III in contrast with only 6% of normal heart patients with palpitations (**Sternick 2004**). The term Mahaim-type conduction is used to describe connections of decremental conduction between the right atrium (RA) or between the AV node and the right ventricle (RV) or near the right branch. In spite of the APs being rare, their unique electrophysiological properties make their treatment and diagnosis complex. The true nature of these pathways, still obscure, make them particularly interesting from the electrophysiology point of view (**Katritsis 2016**). In 1941, Mahaim and Winston described the histology of abnormal connections that originate in the AV node with distal insertion in the RV (**Mahaim 1941**). This was the first description of the nodoventricular pathways of Mahaim, or Mahaim Accessory Pathways (MAP). Later, mapping the atria, mainly the lateral side of the tricuspid annulus, the term atrio-fascicular was adopted (**Gillette 1982; Tchou1988; Klein1988**). These pathways present a decremental conduction and may originate both in the right atrium (RA) and in the AV node, and their distal insertion may occur in the RV or close to the right branch (**Haïssaguerre 1995; Gandhavadi 2013; Sternick 2014**). In spite of conduction through the Mahaim pathways being typically decremental and exclusively anterograde, nodoventricular pathways of retrograde conduction have been described (**Hluchy1999; Josephson2007**). Even being anatomically different from those described in the initial phase, both present similar electrocardiographic and electrophysiological patterns, and the “Mahaim” term has also been adopted for nodoventricular PAs. Although the first case of this arrhythmia was studied electrophysiologically by Wellens and published in 1971, it is considered as being based on a nodoventricular pathway (**Wellens, 1971**); with the advent of surgery and ablation by catheter in the 1980s, it was discovered that most fibers with Mahaim-type conduction features originate in the lateral region of the tricuspid annulus. Since then, the term atrio-fascicular was adopted (**Haïssaguerre 1995; Kottkamp 1996**). Later, postero-septal pathways were found (**Sternick 2014**) and true nodoventricular fibers were identified (**Kottkamp 1996**). Some of these pathways, called “short Mahaim” could present distal insertion into the RV (**Haïssaguerre 1995**). Likewise, pathways with Mahaim features could be atrio-fascicular, atrioventricular, nodofascicular or nodoventricular, depending on their proximal and

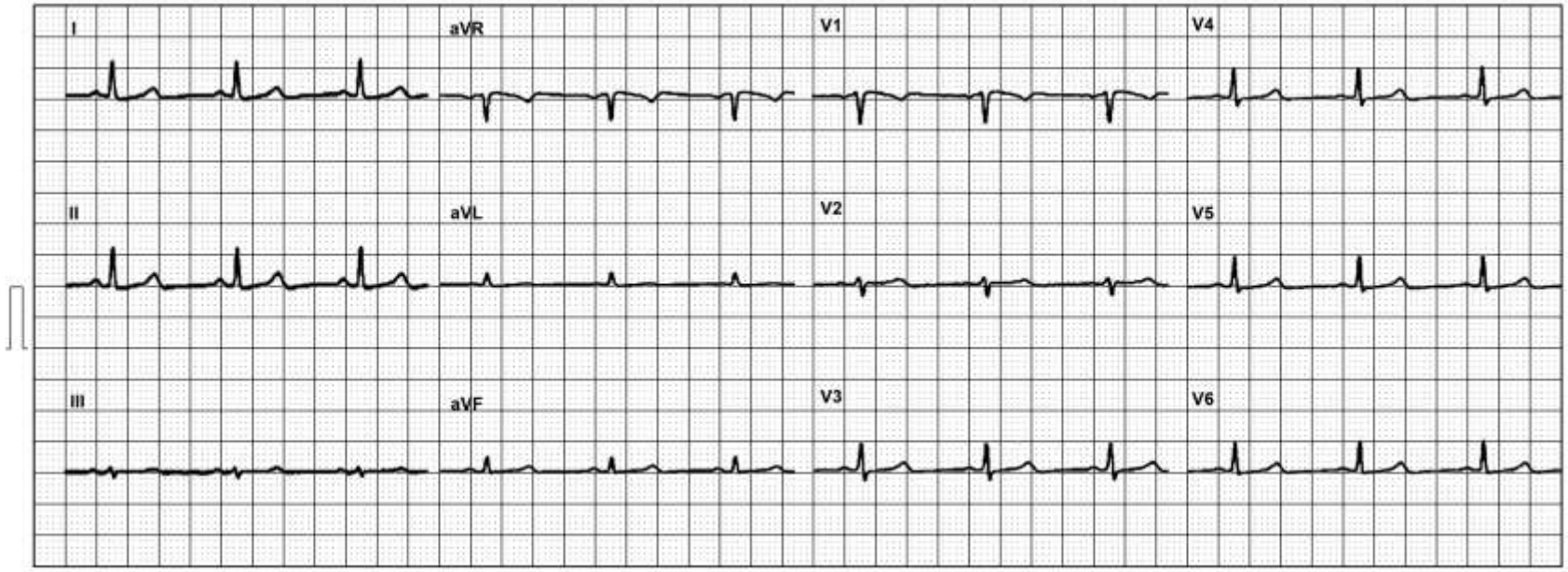
8. distal insertions. ECG in sinus rhythm is usually normal; however possible subtle modifications could be observed as rS pattern in lead III, absence of septal q waves in I and V6, and/or notch or terminal "slurring" in QRS complexes. These minimum modifications in basal ECG should warn us about the presence of Mahaim-type conduction (**Sternick 2004: Liao 2011**). Rarely, Mahaim fibers could exist that conduct rapidly, and thus, the typical preexcitation pattern may appear in ECG in sinus rhythm (**Wellens1971**) or present different degrees of preexcitation manifest by CLBBB morphology with programmed atrial stimulation, which causes an increase in the AV interval and concomitant HV interval shortening before shorter stimulation cycle lengths. On the other hand, the electrogram of the right branch precedes His bundle activation during anterograde activation. See the following example. **Origin:** in brief, the electrophysiological properties of the Mahaim pathways are not uniform, which is explained by the diversity of histological findings (**Haïssaguerre1995; Sternick 2005**). It seems that most of them represent the duplication of the AV node tissue, and its association with the R3-2Q PRKAG2 mutation indicates that this gene is involved in the development of the conduction system (**Tan 2008**). The Mahaim pathways may display spontaneous or post-ablation automaticity (**McClelland 1994**, **Pavlović 2014; Ellenbogen 1989**), and its properties may depend on their location or insertion site (**Haïssaguerre 1995; McClelland 1994**). The catheter ablation is made through the identification of the proximal and distal insertions, and ideally, of recording the proximal potential in the tricuspid annulus, or the distal potential in the RV free wall. The ablation could be facilitated by atrial stimulation once most of them are mapped on the side of the tricuspid annulus or on the RV free wall below the valve. The use of long support sheaths that stabilize the catheter could be very useful (**Haissaguerre 1990; Haïssaguerre1995; McClelland1994; Sternick 2005**).

Site of atrio-fascicular AP ablation in the tricuspid annulus of 48 different antidromic tachycardias (Katritsis 2017)

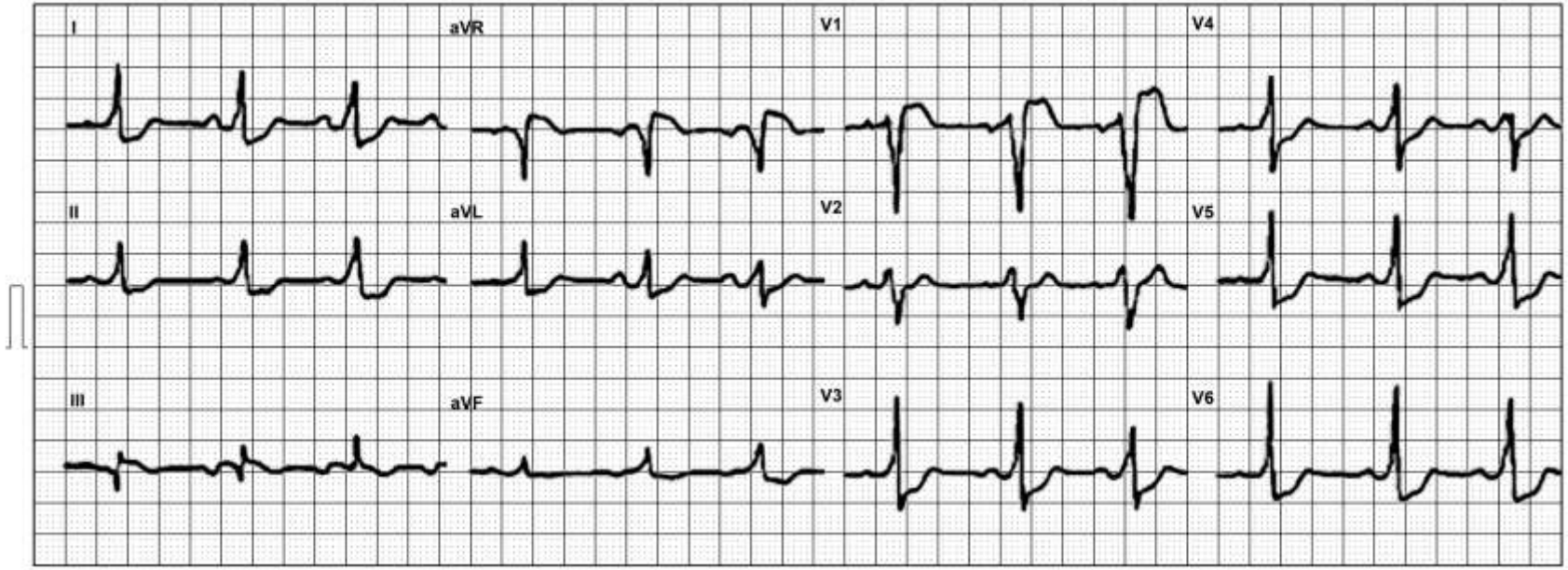


The tricuspid annulus was split into the following regions: Posterior/postero-septal (P) between hours 5 and 7.; Posterolateral (PL) between hours 7 and 8.; Lateral (L) between hours 8 and 9.; Anterolateral (AL) between hours 9 and 11.; Anteroseptal (AS) between hours 11 and 13.
The QRS axis in the FP was classified as: Normal >+15°; Superior <-30°; Horizontal +15° a -30°. Regardless from the axis of the frontal plane, most of cases of AP were in the L and PL regions. There was no single case with normal FP axis located in the AS region (**Sternick 2014**).

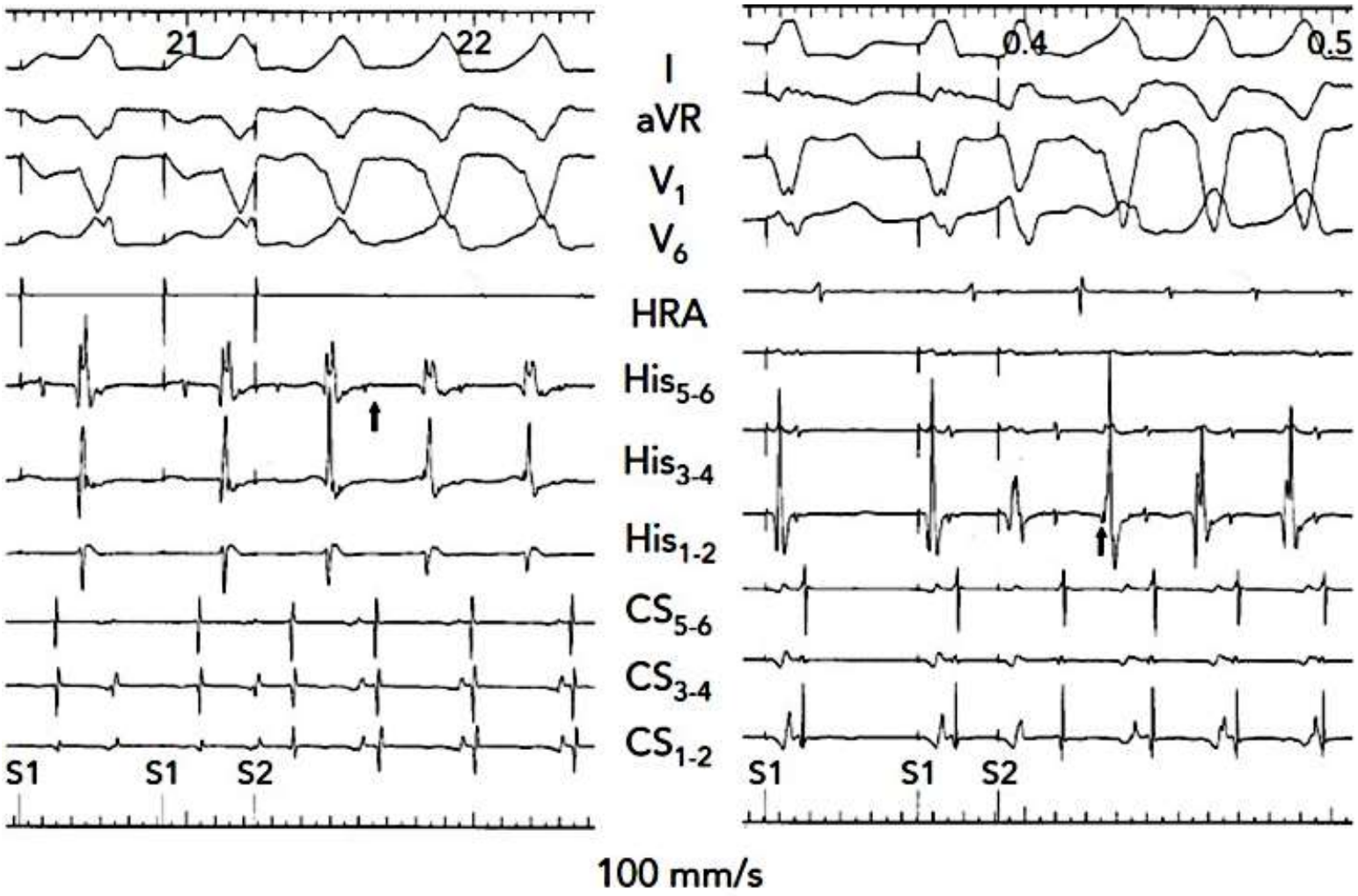
Example of ECG with absence of spontaneous preexcitation during sinus rhythm, and Mahaim fibers carrier.



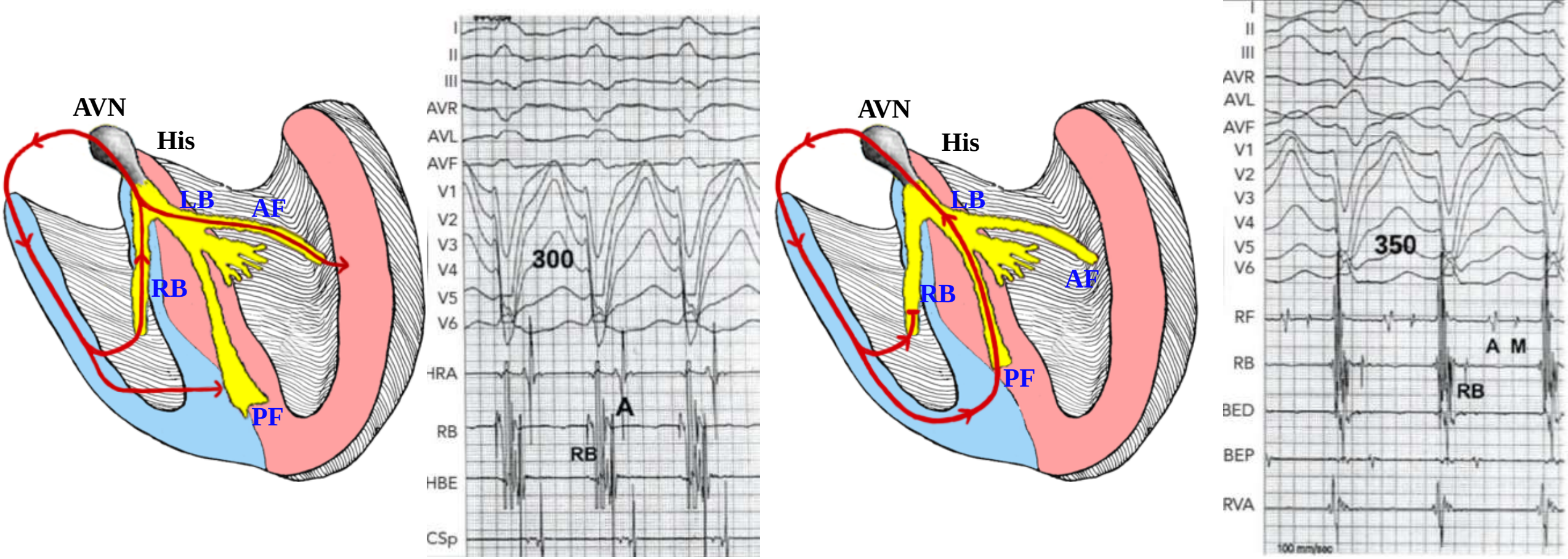
Atrial stimulation shows several degrees of preexcitation. Normal AV nodal conduction (first beat), fusion between nodal and Mahaim conduction (the following two beats) and appearance of CLBBB due to exclusive conduction over the Mahaim pathway (last beat).



ECG during tachycardia Although typically, antidromic atrioventricular reentrant tachycardia over a Mahaim fiber had a left bundle branch block (LBBB) morphology, several QRS and axis patterns may occur (see Figure below) (**Gandhavadi 2013; Sternick 2014**). It seems that these pathways insert into or close to the right branch, and the variations on the axis of the frontal plane could be explained by the location of the left branch outlet and a variable degree of ventricular activation fusion between anterograde conduction over the pathway and, following the retrograde invasion by the left branch, partial anterograde left ventricular activation of the left ventricle over the conduction system of the left side, particularly the anterior fascicle. The several patterns of QRS and rate changes observed during short-to-long V-A tachycardia change could be explained by the mode of retrograde conduction over the branch system (**Gandhavadi 2013; Sternick 2014**).



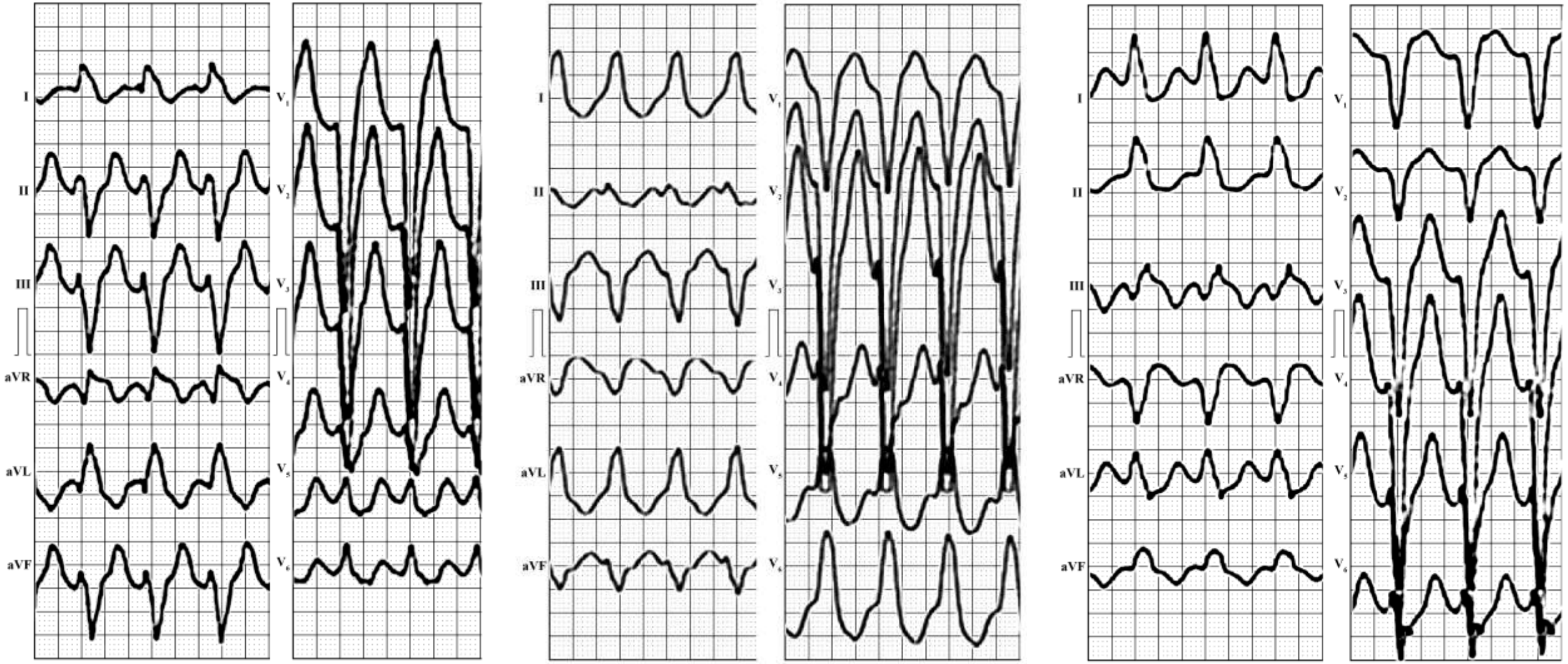
Induction of tachycardia by atrial (left panel) and ventricular stimulation (right panel). The arrows indicate the retrograde potential of the His bundle. Reproduced from Giazitzoglou et al, 2008, (**Giazitzoglou 2008**) with permission.



Alteration in morphology of atrioventricular reentrant tachycardia QRS of short-long VA circuit

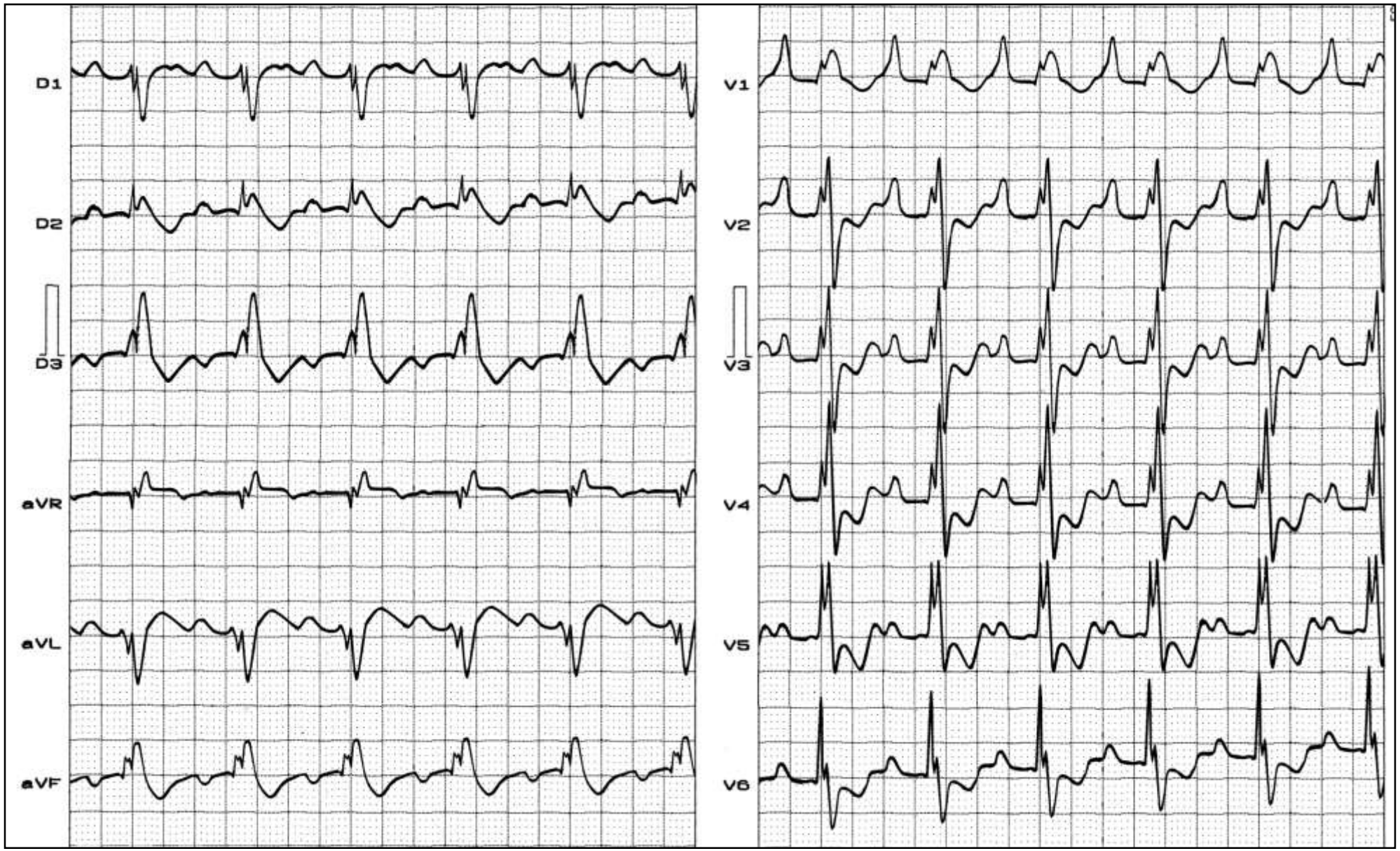
A: During short V-A AVRT (length cycle of tachycardia 300 ms), there is also anterograde activation over the left anterior fascicle to produce condensed QRS complex with normal axis. B: With retrograde right bundle branch block, anterograde conduction over the left anterior fascicle is no longer possible and conduction to the left ventricle continues only through the right free wall. Therefore, long V-A AVRT (cycle length of tachycardia of 350 ms) has an axis to the left. During short-long V-A AVRT change, QRS width also increases from 120 to 150 ms.

A: Atrial electrogram; AF: anterior fascicle; AVN: AV node; AVRT: Atrioventricular reentrant tachycardia; PCS: proximal coronary sinus catheter; HB: His bundle catheter; HRA: high right atrium catheter; LBB: left bundle branch catheter; M: Mahaim potential; PF: posterior fascicle; RB: right branch potential; RBB: right bundle branch catheter; RVA: right ventricle apex catheter; V-H: His interval. Reproduced from Gandhavadi et al, 2013 (**Gandhavadi 2013**) with permission.



12-lead ECG of three different patients with antidromic tachycardia. From left to right: superior, horizontal and normal frontal plane. QRS axis ablated in the same sector as the tricuspid annulus (site of ablation between 7 hours and 7:30). Sternick et al 2014 (**Sternick 2014**)

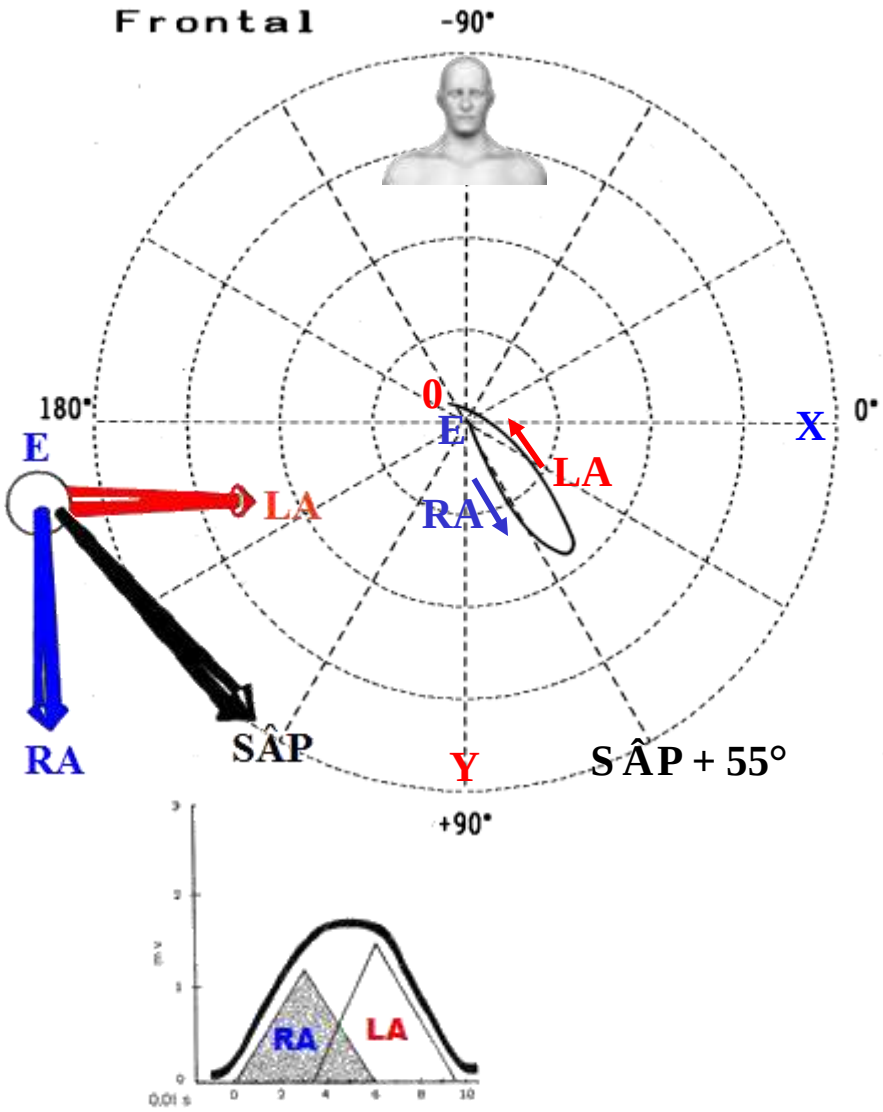
ECG of a patient with Ebstein's anomaly of tricuspid valve



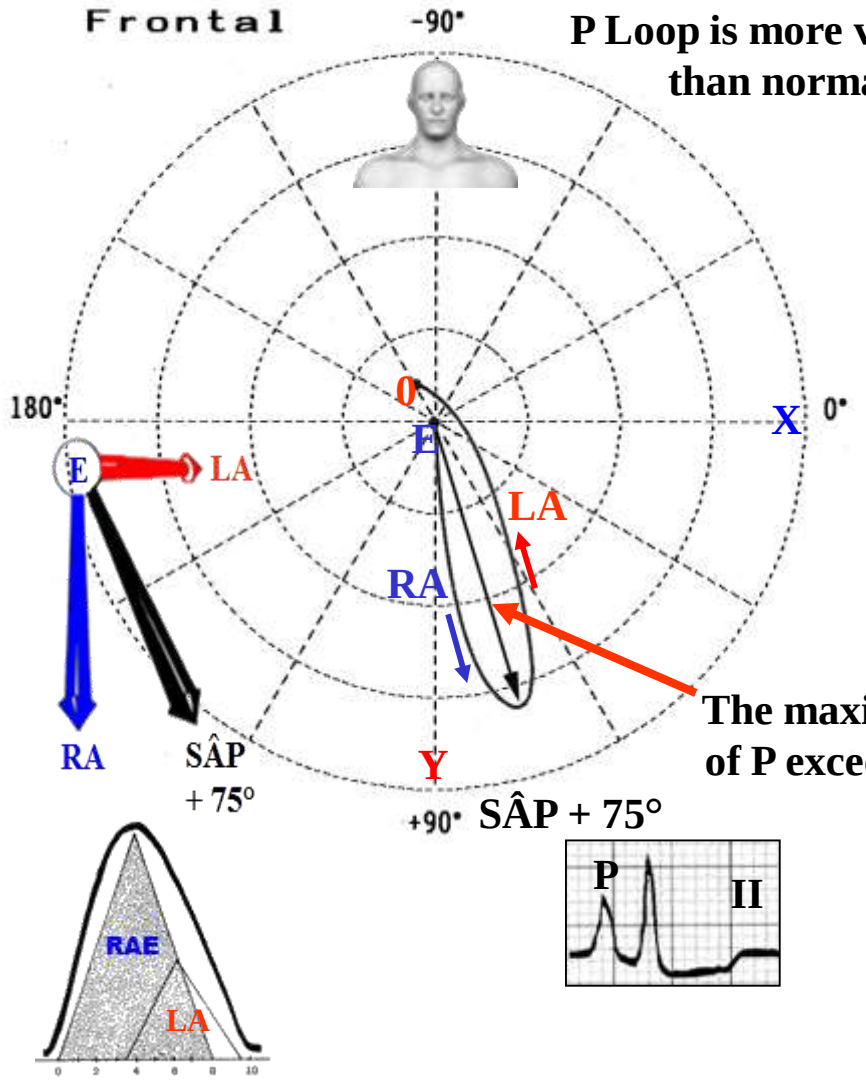
P wave of great voltage from V_1 to V_4 and in the inferior leads: "Himalayan" P waves, SÂP -15º, 1st degree AV block; prolonged PR interval (290 ms) (1st degree AV block); bizarre QRS with CRBBB of low voltage and with initial small q wave in V1, fragmented QRS.

Normal P loop and in Right Atrial Enlargement in the Frontal Plane

Normal P loop



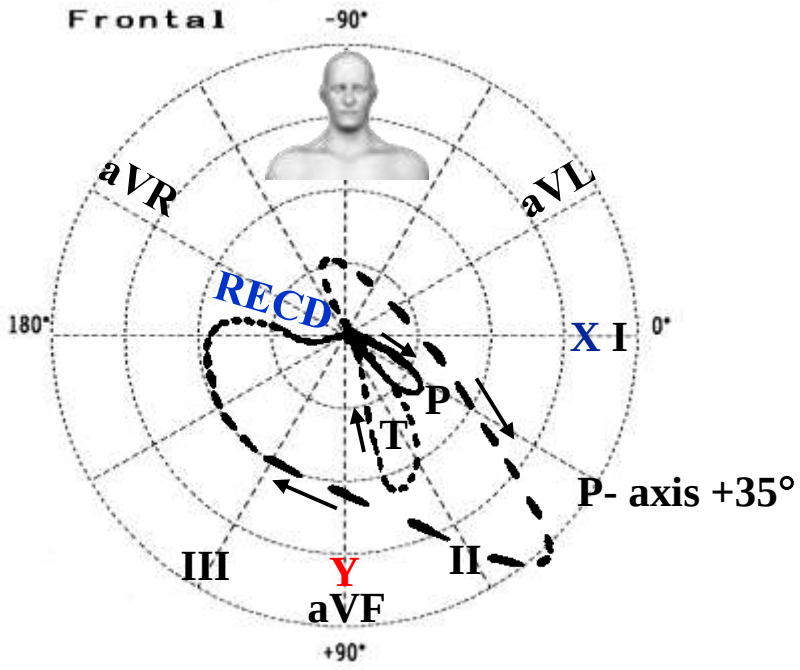
P loop in Right Atrial Enlargement



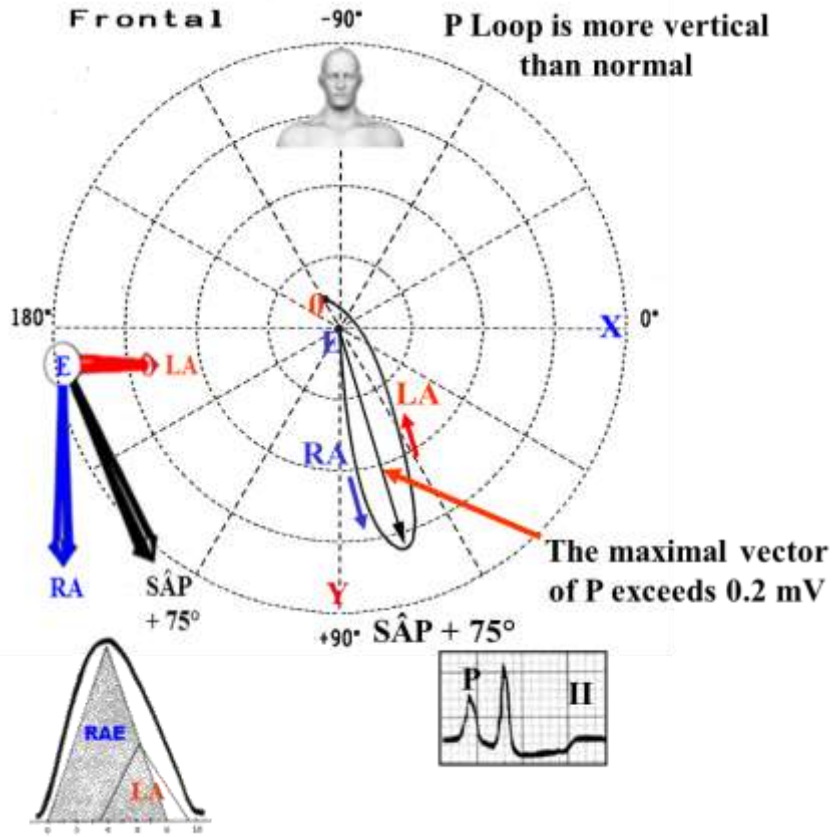
P Loop is more vertical than normal

The maximal vector of P exceeds 0.2 mV

P-loop in a case of Ebstein´s anomaly
right atrial enlargement without right P-axis



P-loop in right atrial enlargement



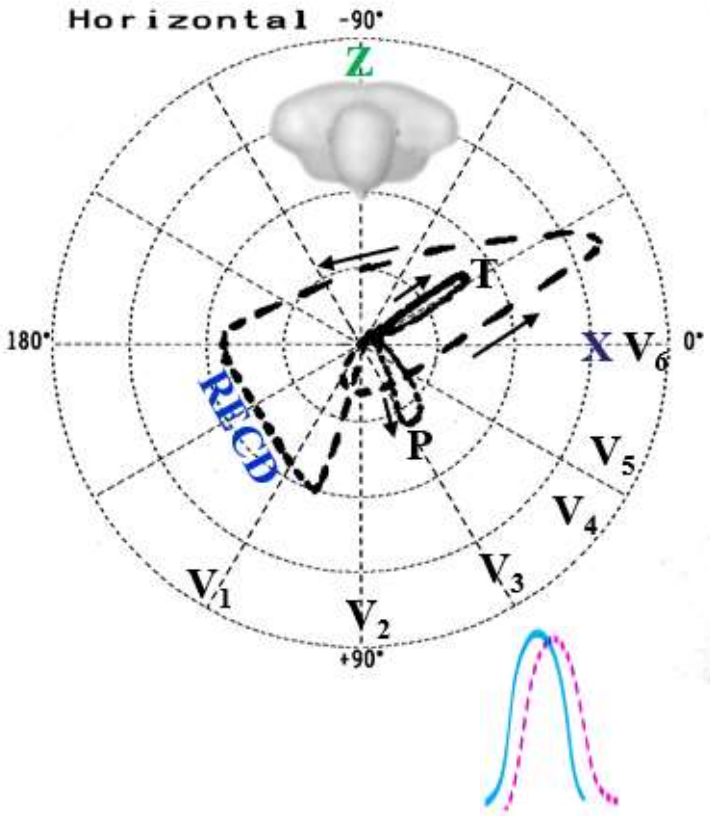
FP: Giant P loop directed to left and downward, P axis +35°, QRS loop with CW rotation, QRS axis near +90°. Right End Conduction Delay (**RECD**) on X orthogonal lead to the right.

P-loop in right atrial enlargement characteristics:

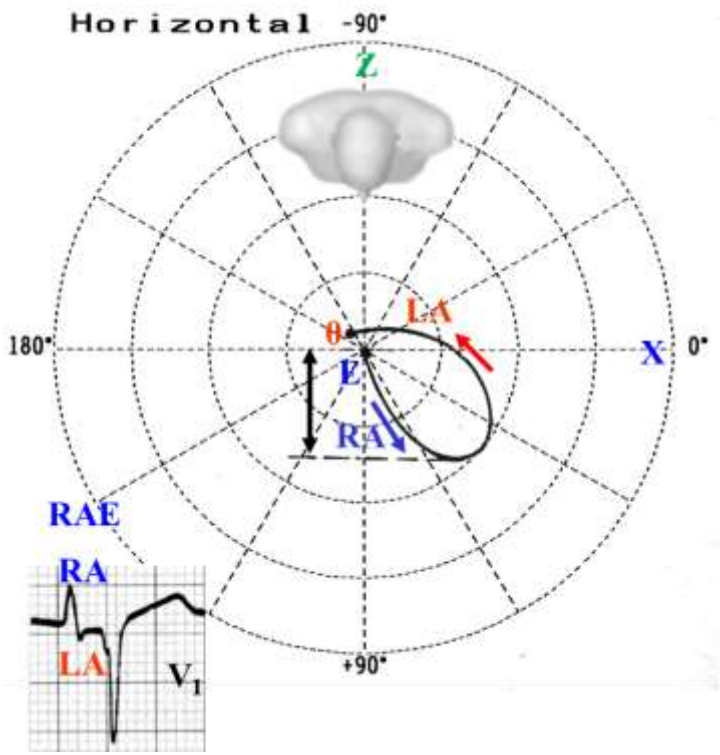
1. Inscription of the P-loop is usually CCW in all three planes.
2. In the HP, most of the P-loop are directed anteriorly. The magnitude of the anteriorly directed forces ≥0.07 mV in adults and ≥0.09 mV in children under 15 years of age. The maximum P vector may exceed 0.10 mV.
3. In the RSP, orientation of the P-loop is more vertical than usual. The maximum P vector may exceed 0.20 mV.

P-loops in the Horizontal Plane

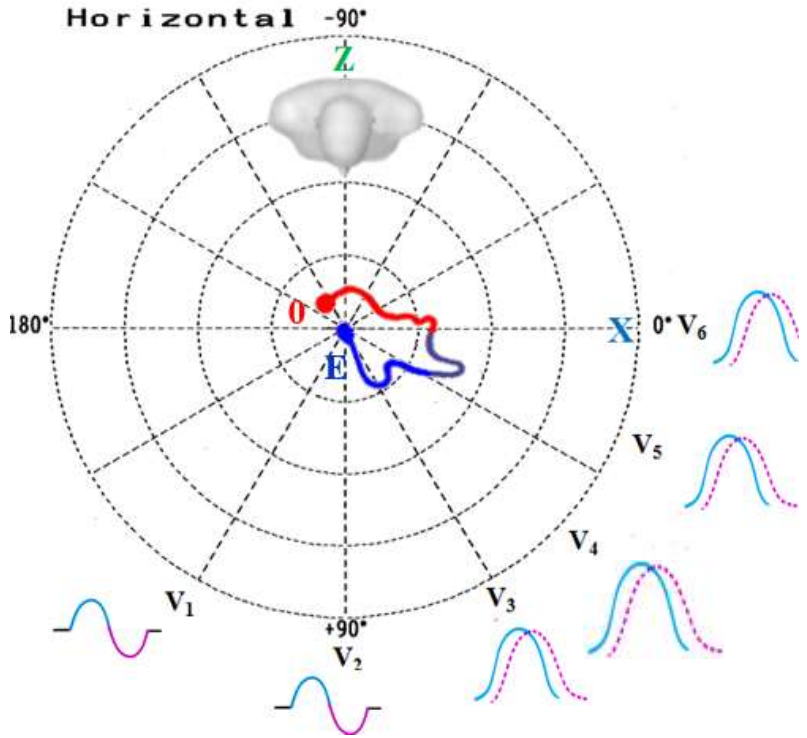
Giant P-loop in Ebstein anomaly



P-loop in Right Atrial Enlargement



Normal P-loop

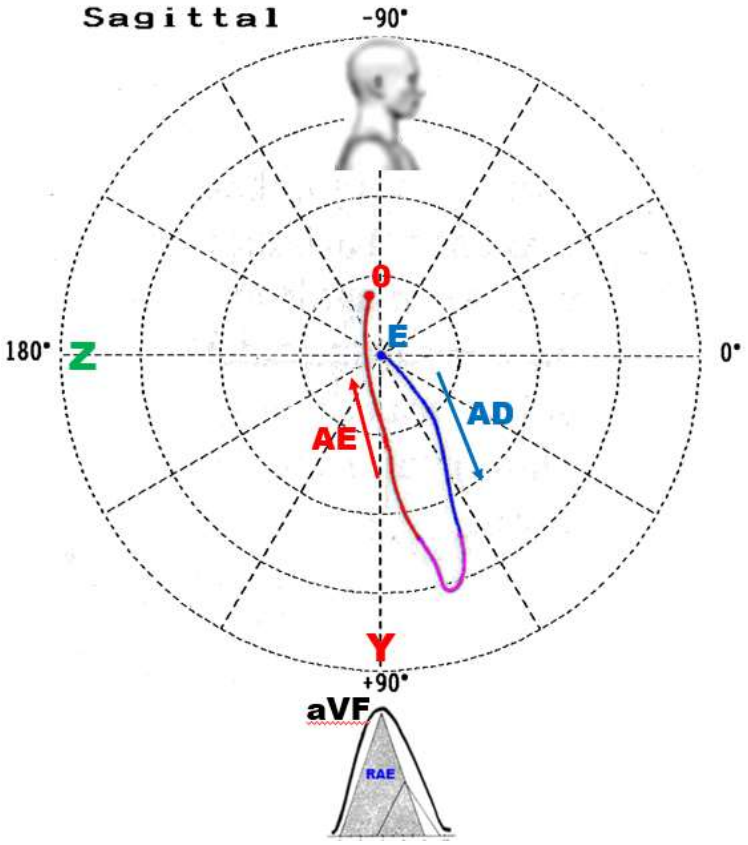


Max. P-loop anterior forces: ≥ 0.07 mV
Max. P-loop vector: > 0.1 mV
P-loop location: on anterior right quadrant
Max. P-vector voltage: >0.1 mV
Anterior P-loop Max Forces: Adults: >0.06 mV, children: >0.08 mV

P-loop rotation: CCW
P-loop morphology: Oval
Max. Vector location: + 50 to -45°
Max. P-vector voltage: ≤0.1 mV
Anterior P-loop Max Forces: Adults: ≤0.06 mV, children: ≤0.08 mV

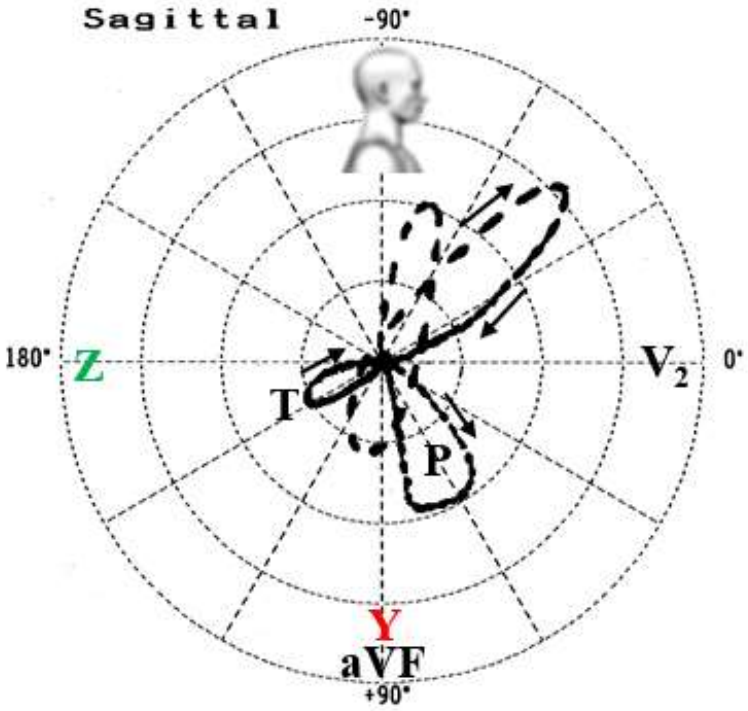
P-loops in the Right Sagittal Plane

Right Atrial Enlargement



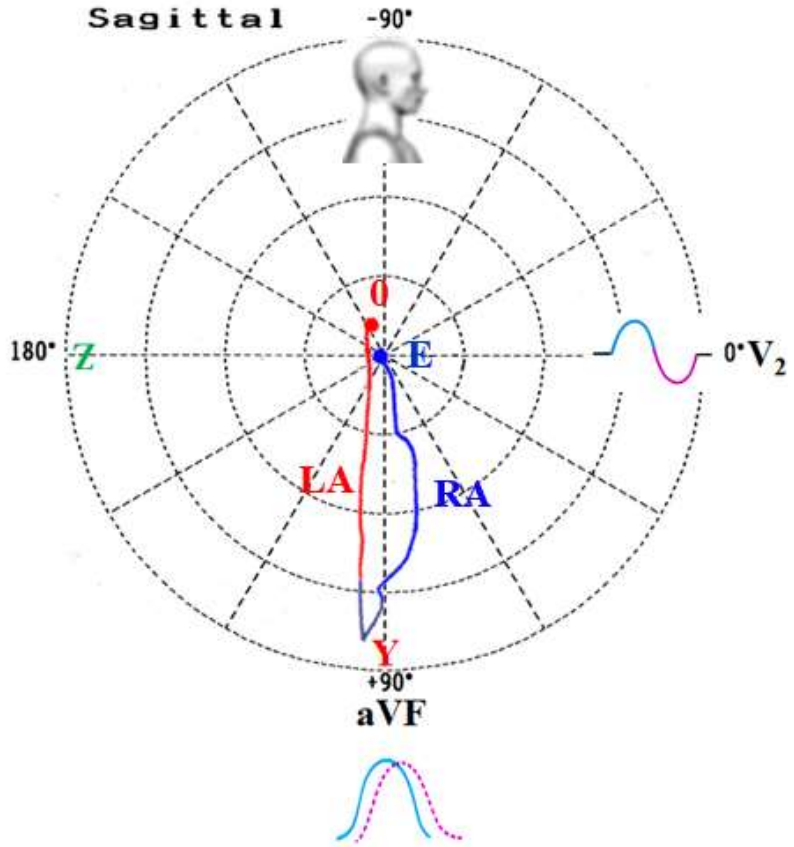
P loop located in anterior inferior quadrant
Maximum P-voltage vector: > 0.18mV. Exceeds 0.20 mV.
Maximum anterior forces: Adults > 0.06mV. Children: >0.08mV

Right Atrial Enlargement in Ebstein's anomaly



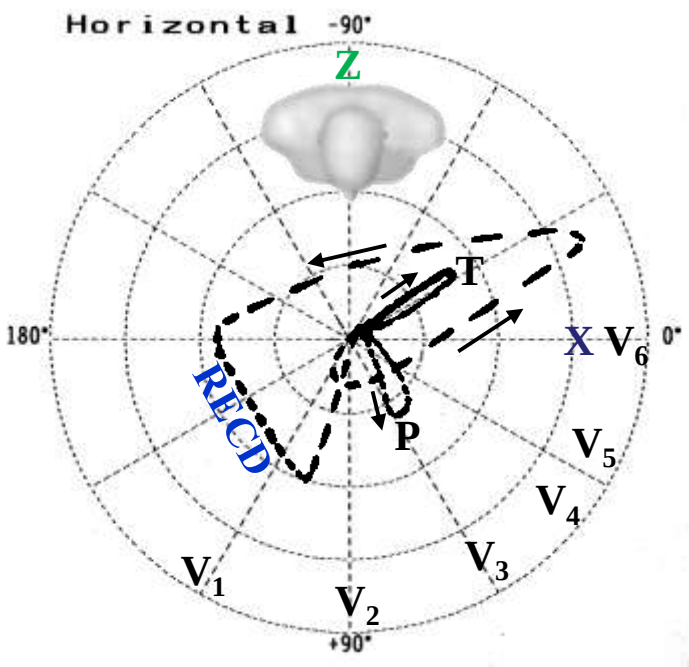
Himalaya P-loop

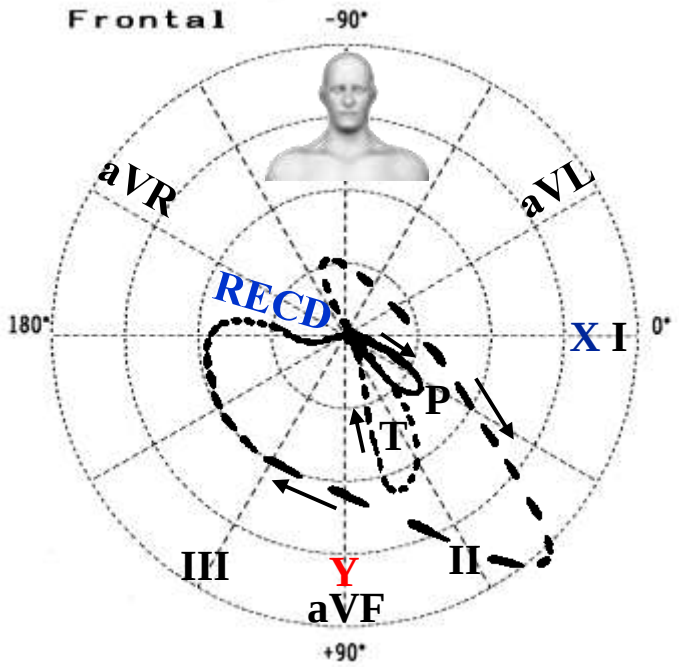
Normal

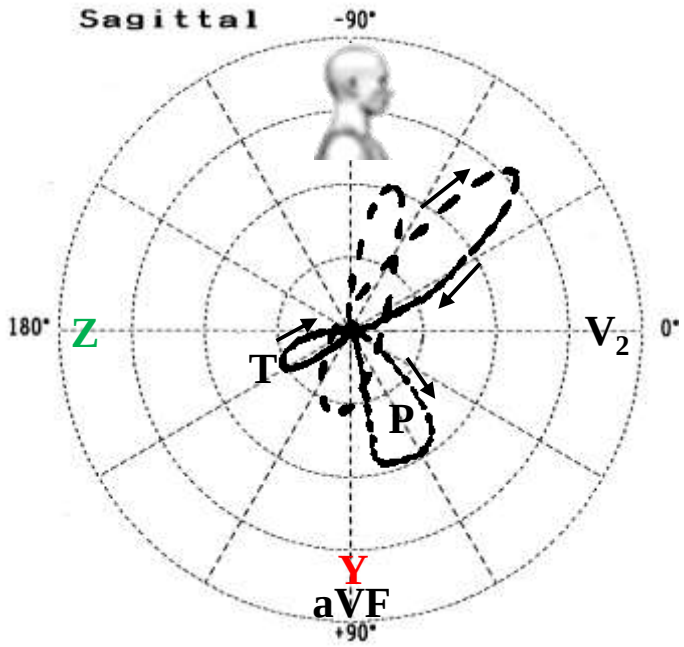


P-loop location: Initial portion (**RA**) antero-inferior and terminal posterior (**LA**)
Shape: Thrust tip or triangular
Maximum voltage vector: ≤ 0.18mV
Maximum anterior forces: Adults: ≤0.06mV; children: ≤0.08mV

VCG of a 33 years old woman with Ebstein's anomaly







- **Horizontal Plane:** Giant P loop directed to front and leftward near +58°, maximal P vector exceeds 0.1 mV: right atrial enlargement, the P-loop is directed anteriorly. The magnitude of the anteriorly directed forces is ≥0.07 mV. The maximum P vector exceeds 0.10 mV. QRS loop with CW rotation and typical pattern of complete RBBB Grishman type because the afferent limb is located behind X orthogonal lead and the final QRS loop with delayed located on the anterior right quadrant: right end conduction delay RECD.

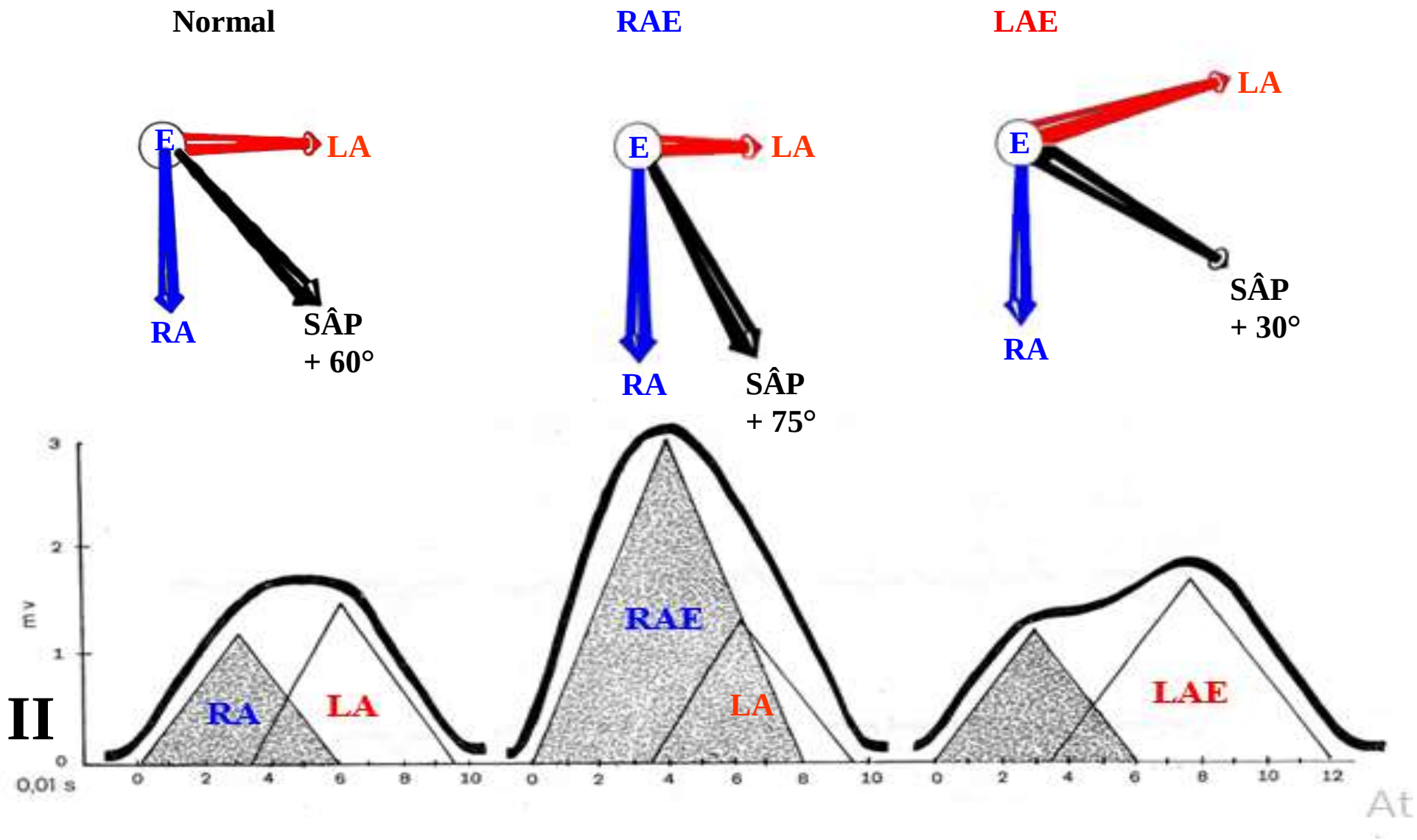
- **Frontal Plane:** Giant P loop directed to left and downward, P axis +35°, QRS loop with CW rotation, QRS axis near +90°. **RECD** on X orthogonal lead to the right.

- **Right Sagittal Plane:** Giant P loop located in the inferior and anterior quadrant near +60°, orientation of the P-loop is more vertical than usual. The maximum P vector exceeds 0.20 mV. QRS loop directed to the front and upward. **RECD** on anterosuperior quadrant.

Right atrial enlargement. ECG criteria with high specificity

ECG criteria	SE %	SP %
A. QRS criteria		
1. QR or qR in V1	≈ 15	> 95
2. QRS V1 ≤4 mm + QRS V2/V1 ≥5	46	93
3. R/S > 1 in V1	≈ 25	> 95
4. SÂQRS > 90°	34	> 95
B. P criteria		
1. P wave in inferior leads < 2.5mm	7	100
2. Positive part of P wave V1 > 1.5mm	17	100
3. Positive part of P wave V2 > 1.5mm	33	100
C. Combined		
1. Positive part of P wave in V2>1.5mm+SÂQRS>+90°+R/S>1 in V1	49	100

Profile of normal P wave in RAE and LAE



VCG features in Ebstein's anomaly

The Frank VCG in Ebstein's anomaly shows prolongation of the QRS loop/complex, owing to RBBB; type B WPW syndrome is occasionally found. In patients between 1 day to 31 years old, with the men age of 8.6 years, the QRS duration was increased in all patients, averaging 103 ms (**Ellison & Restieaux 1972**). The delay is secondary to RBBB because delayed conduction being primarily in the terminal of the QRS loop/complex. In patients with type B WPW, QRS prolongation is consequence of delayed initial forces. In infants from 1 to 3 days old there are prolongation of QRS (between 80-100 ms). In normal conditions, the newborns have QRS duration average of 50 ms. The table below shows the normal QRS durations.

Fetus and post-birth days of life	QRS duration measured in V5 in milliseconds (ms)
Between 18 and 22 weeks of pregnancy	47 ms
40 weeks	53 ms
0–1 days	20 to 80 (average 50)
1–7 days	20 to 70 (average 50)

Right ventricular voltages are reduced has shown by decrease in the RMSV (0.6 ± 0.4 mV) and 10 to 20 ms vectors, and by the predominantly leftward orientation of the HP and FP QRS loops. The RMSV is usually of greater magnitude in patients with little or not tricuspid regurgitation than in those with severe tricuspid insufficiency. The QRS axis in the majority of patients show a leftward inferior frontal axis. In presence of WPW the QRS axis has extreme left axis deviation.

HP: QRS loop rotation is CCW or figure eight and the loop is oriented to the left and predominantly posteriorly. This reflects the reduction in the right ventricular potentials.

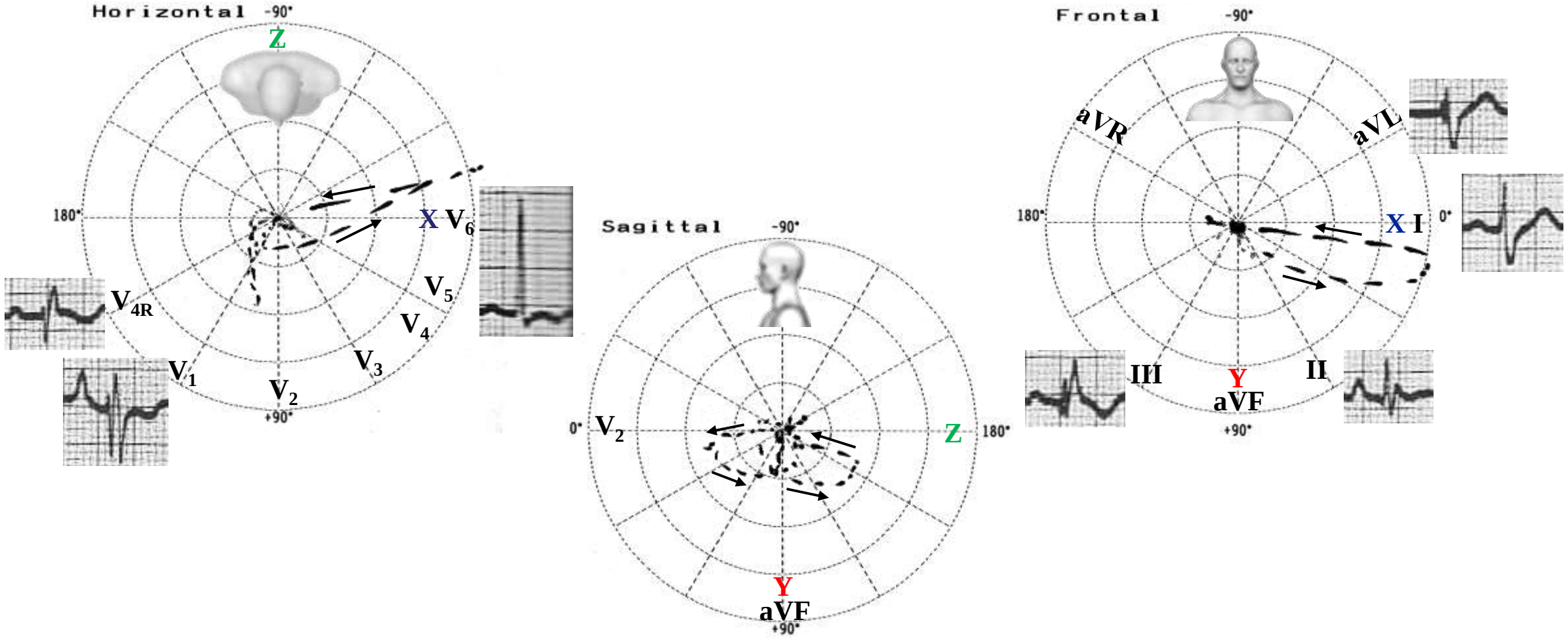
In presence of type B WPW initial conduction delay is observed (δ loop) and the early forces moved directed to the left. Type B WPW is always the type seen in Ebstein's anomaly.

FP: QRS axis is located in left inferior quadrant in absence of WPW and the QRS loop rotation is CW or figure eight. In presence of type b WPW the QRS axis has extreme left axis deviation.

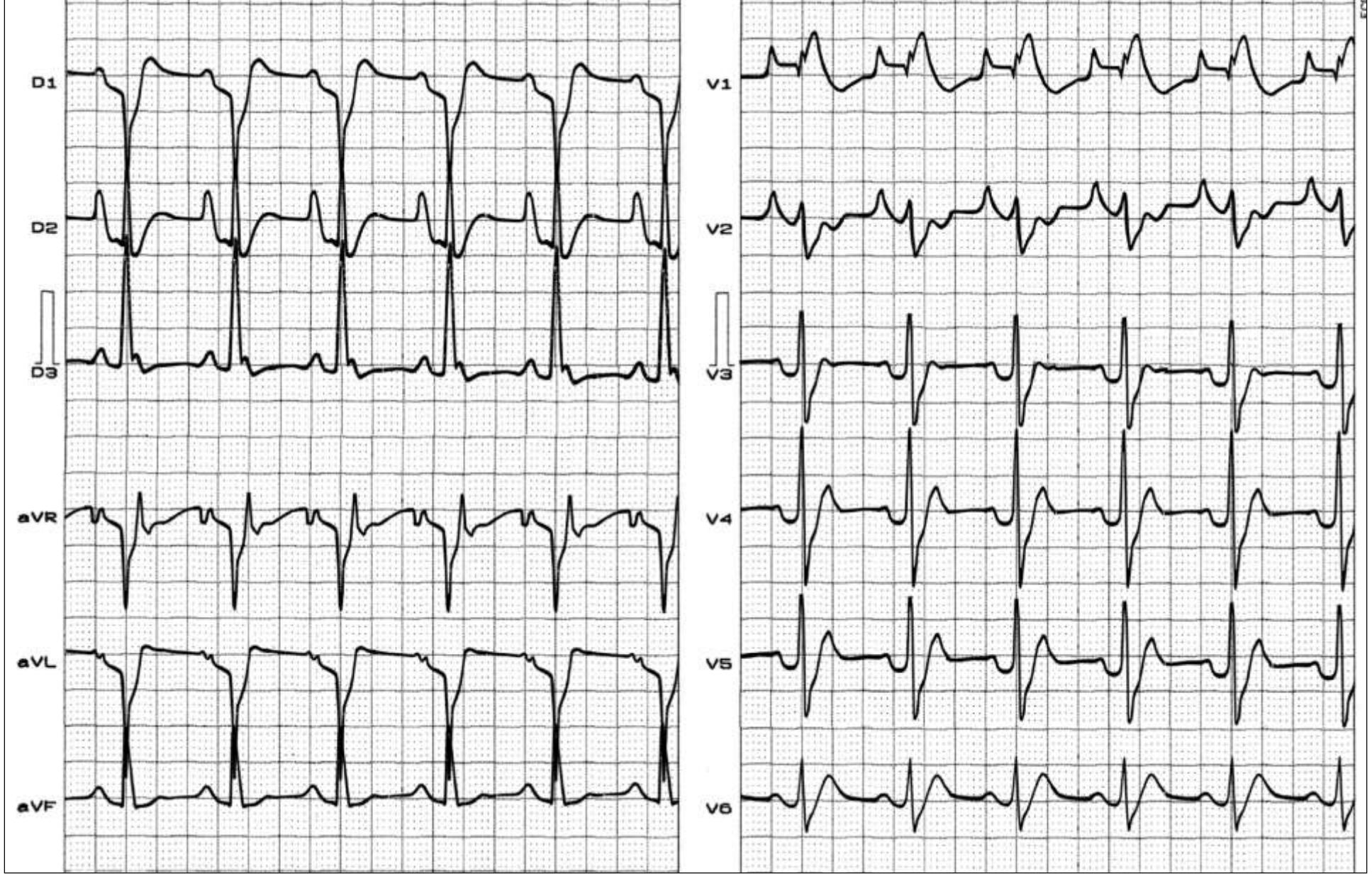
LSP: Rotation is predominantly CCW and the reduction in anterior forces could be seen. P loop is oriented in the normal direction, to the left, inferiorly and anteriorly. Clear right atrial enlargement loop is the rule. The T loops are directed predominantly to the left and posteriorly. 50% directed inferiorly and 50% directed superiorly.

Major electrophysiologic abnormalities in Ebstein´s Anomaly of the tricuspid valve
1) Intraatrial conduction disturbance: Right atrial P-wave abnormalities, PR interval prolongation
2) Atrioventricular nodal conduction: PR prolongation
3) Infranodal conduction: intra-His or infra-His conduction abnormalities
4) Right Bundle Branch Block distal to the right bundle branch
5) Type B Wolff-Parkinson-White pré-excitation
6) Supraventricular tachycardias
7) Atrial fibrillation
8) Atrial flutter
9) Arrytmogenic atrial right ventricle
10) Mahaim fibers medited arrhythmias
10) Deep Q waves for V_1 toV_4 (It is an indirect signal of right atrial enlargement)

The Frank VCG in a 11 years old girl with Ebstein's anomaly and minimal tricuspid insufficiency



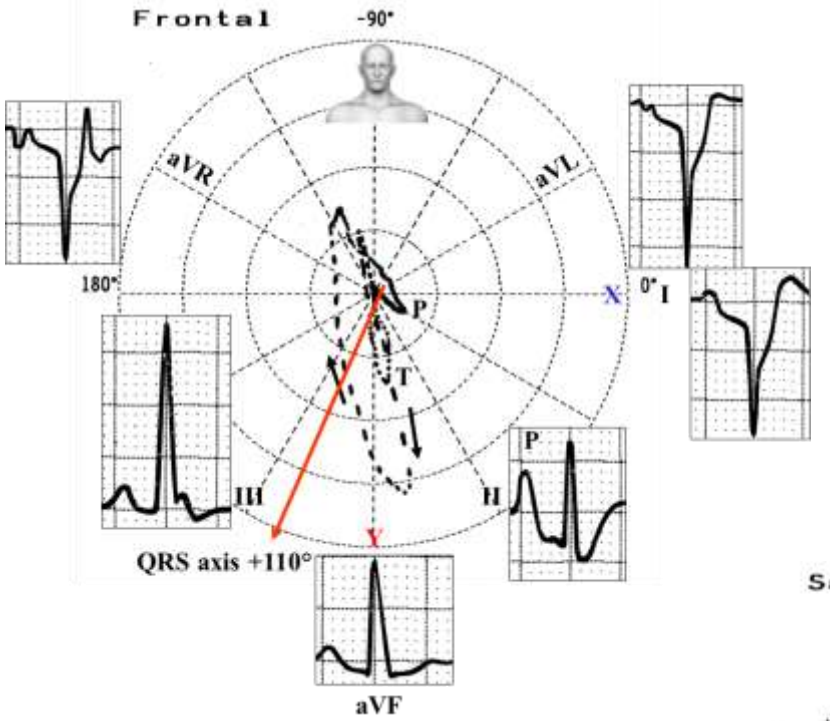
HP: The QRS loop shows small initial anterior forces and terminal conduction delay. The right maximum spatial voltage (RMSV) is 0.6 mV, consistent with the presence of only trivial tricuspid regurgitation; it would be expected to be smaller if severe regurgitation were present. QRS duration 110 ms. The ECG shows very tall P waves in V1 with tetraphasic QRS pattern rSrS, which is indicative of right ventricular hypertrophy. Conclusion: Right atrial enlargement + incomplete RBBB.



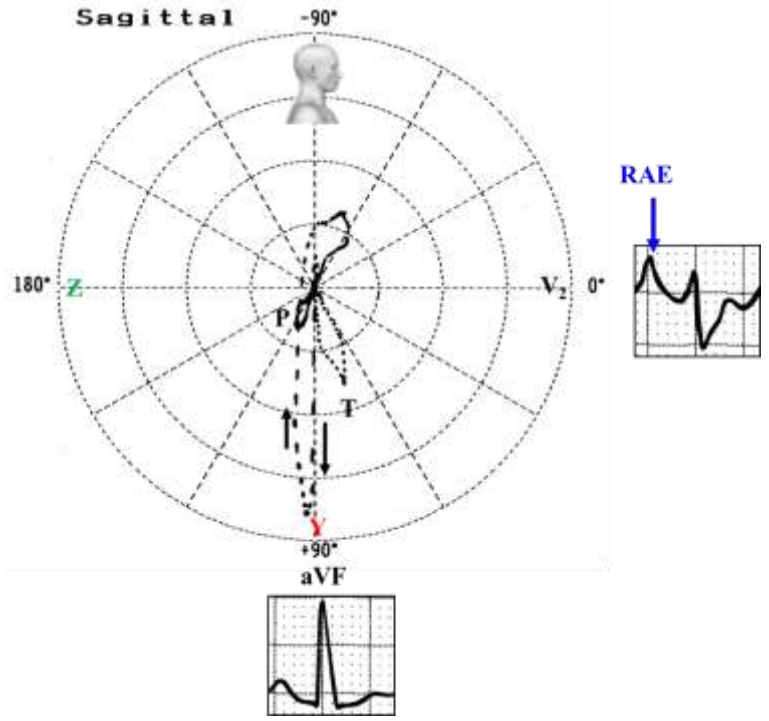
Clinical diagnosis: Ebstein's anomaly.

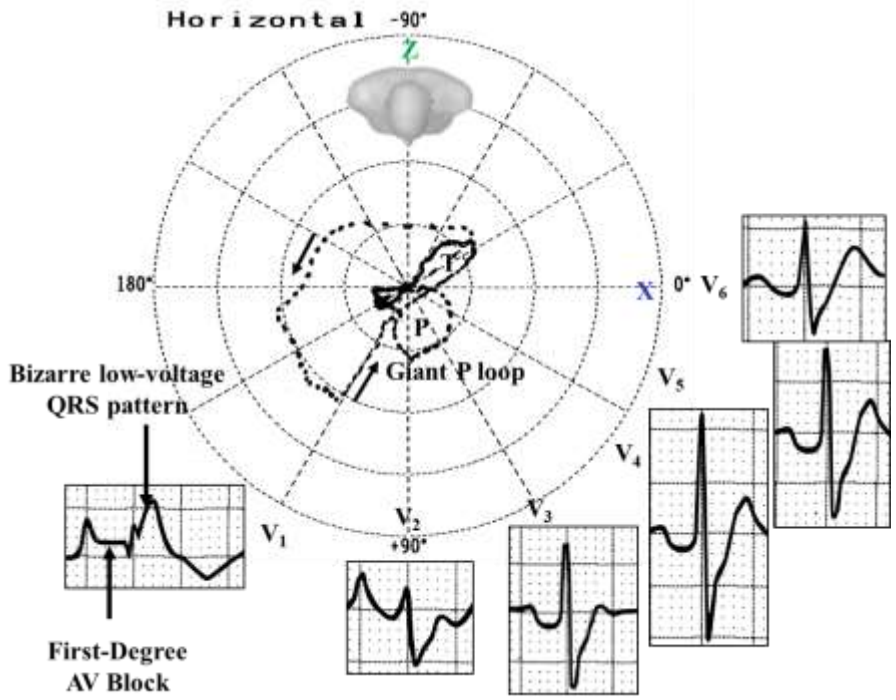
ECG diagnosis: Right atrial enlargement "himalayan P-waves", 1st degree AV block, bizarre right bundle branch block of low voltage in V1 and with initial q wave in this lead. R = S pattern from V3 to V6: RVH.

ECG/VCG correlation

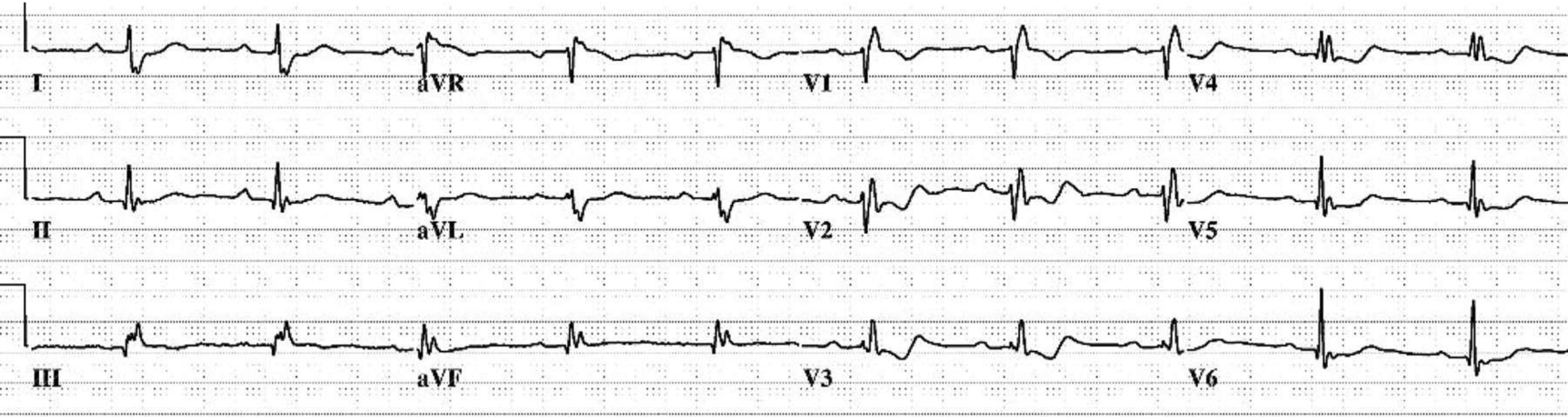


P-wave with 4 mm in II “Himalayan P wave”; very large P loop; right axis deviation.





Giant rounded P loop, very tall P wave in VI-V2, first-degree AV block, bizarre low voltage CRBBB with initial q wave, RS pattern from V3 to V6 and QRS loop predominantly located in the right quadrants: RVH.



ECG of a patient with severe Ebstein's anomaly showing the typical changes, with first-degree atrioventricular block, first degree AV block (PR interval=226 ms), right bundle-branch block, and somewhat bizarre configuration of the QRS complex, tetraphasic QRS complex in trabecular area of the right ventricle (V_2-V_3).

ECG diagnosis: WPW with right lateral anomalous pathway, presenting inferior axis and transition in V3. Therefore, the delivery of radiofrequency energy does not entail the risk of causing total AVB.

Ebstein's anomaly possibly shows WPW type B, with anomalous bundle located in the RV free wall (between the RA and the RV) or septal posterior. An anomalous bundle at the right is found in ≈10% of cases of Ebstein.

The association with WPW in Ebstein's anomaly occurs in 5 to 10% of the cases. There are authors that suggest a greater percentage of associated pre-excitation (25%). Finally, some think that type-B WPW associated to tachyarrhythmias is observed in more than 50% of cases (Deal 1985). This is the congenital heart disease most associated to WPW.

Patients with anomalous bundles at the left, rarely show organic heart disease, while those with anomalous bundles at the right, are associated in 45% of the cases, to organic heart disease (**Damjanović 2008**).

The location of the anomalous pathway in Ebstein's anomaly could be: right anterior (the most frequent one) in point 2 of Gallagher; right lateral in point 3; right posterior in point 4, and right posterior septal in point 5.

There are rare cases of Ebstein's anomaly with pre-excitation of the Mahaim type: normal *PR* interval with δ wave that may resemble *CLBBB*. The cases of Ebstein with CLBBB may correspond to pre-excitation, Mahaim type. Mahaim pre-excitation is due to fibers that get away from the His-node system, either from the *AV* node, or from the His bundle or its branches, originating two variants: Ventricular node (connections); Fasciculo-ventricular (tracts).

From a series of 224 patients studied by Torres (**Torres 2007**) at the Ignacio Chávez Institute of Mexico, 64 (28%) had documented tachycardias. Thirty three patients with recurrent tachyarrhythmias had a single right anomalous bundle that could be ablated successfully. Only 21 from these 31 had a typical WPW pattern and none had CRBBB pattern during sinus rhythm. The delivery of radiofrequency energy caused in 94% of cases, CRBBB pattern. The absence of CRBBB in Ebstein's patients and recurrent tachyarrhythmic events had a 98% sensitivity and 92% specificity for anomalous bundle diagnosis.

Thirty three percent of Ebstein's patients and symptoms of tachyarrhythmia do not have WPW.

The absence of CRBBB pattern is a strong predictor of anomalous pathway.

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Devemos ter cuidado ao escolher os inimigos porque terminamos parecidos a eles.

We must be careful when choosing enemies because we end up resembling them.