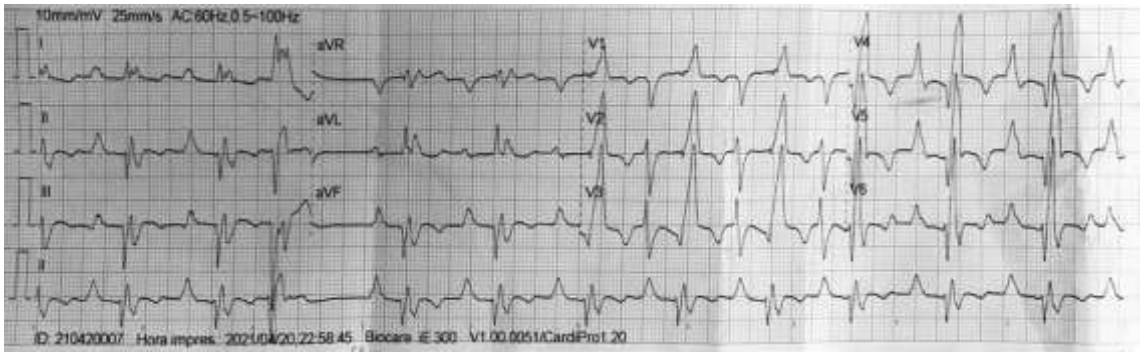
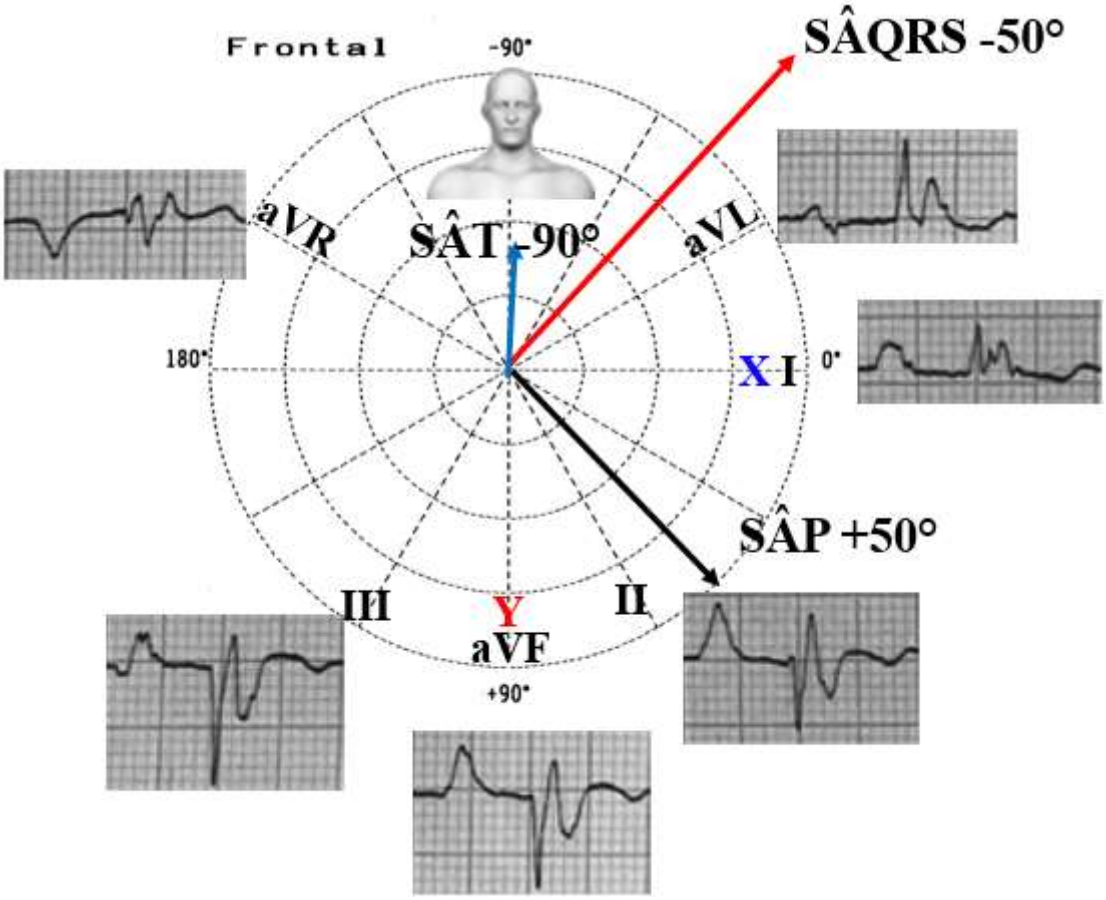
Case report from Dr. Marlon Barahona

Interrogatory ignored. Physical examination?



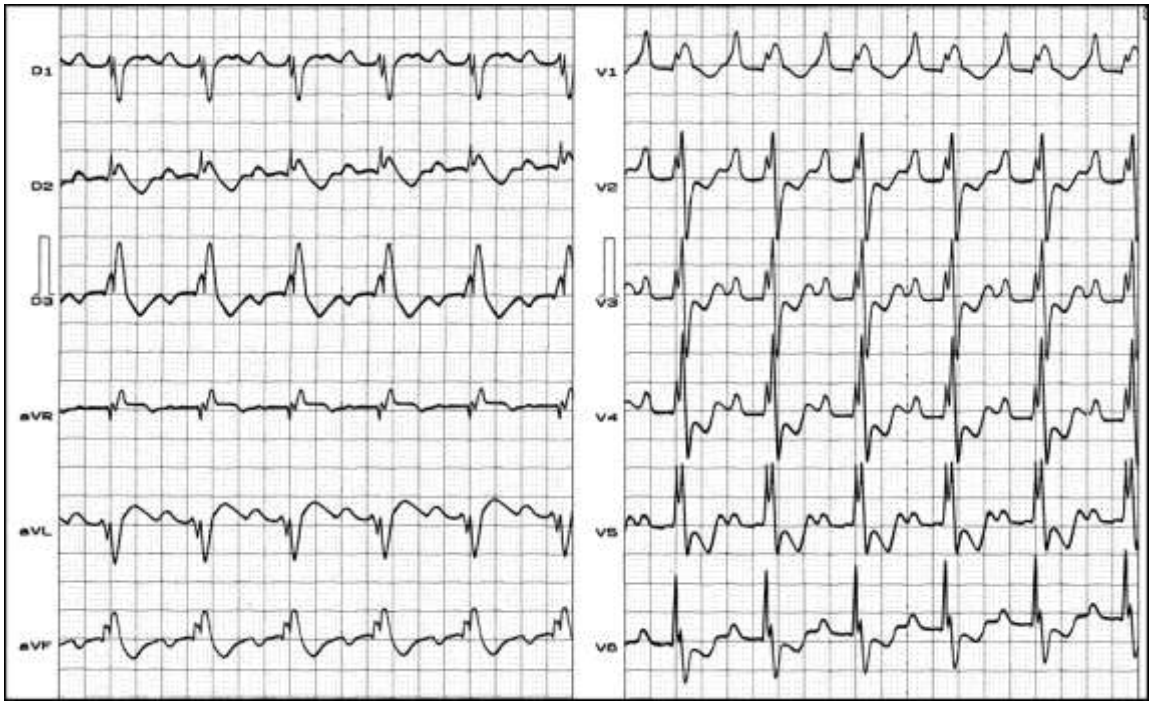
ECG diagnosis: sinus rhythm, “Himalayan or alpine” P wave, prolonged P duration: biatrial enlargement (P-axis + 50°) very prolonged P-wave duration (PWD), very prolonged PR interval (270 ms), extremely broad QRS interval (near 200ms: bizarre second QRS attached to preceding normal QRS?) or consequence of atrialized right ventricle, with low voltage, bizarre aspect, fragmented QRS (fQRS), several leads with tetraphasic or polyphasic QRS complexes, deep Q waves in inferior leads (extremely suggestive of Ebstein’s anomaly: this distinctive Q wave pattern is indicative of right ventricular intracavitary potentials unusually far leftward as a result of large size of RA), frequent premature ventricular contractions with bigemy.



QRS axis is inferior: although a splintered polyphasic QRS makes axis difficult to determine.

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

Dr Raimundo Barbosa case



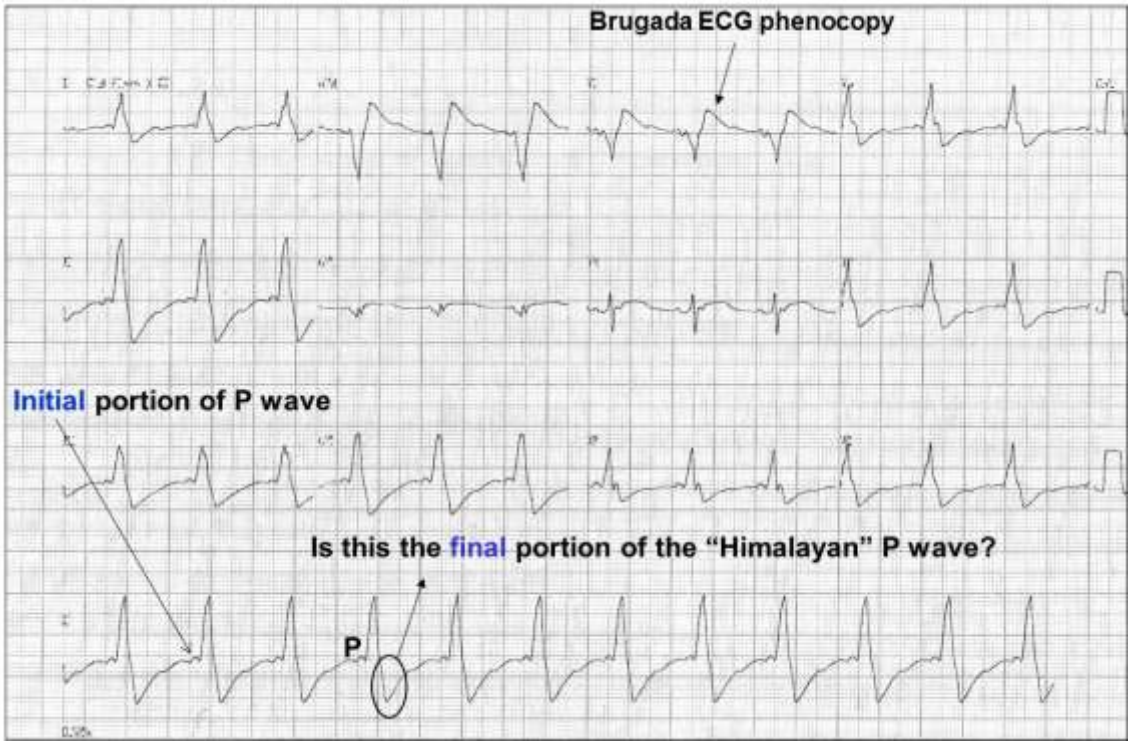
P wave of great voltage from V1 to V4 and in the inferior leads: “Himalayan” P waves, SÂP -15º, 1st degree AV block; prolonged PR interval (290 ms) (1st degree AV block); bizarre QRS with CRBBB of low voltage and with initial small q wave in V1.

Case report

This tracing belongs to a 46-year-old woman, carrier of Ebstein’s anomaly, admitted in the emergency room with atypical chest discomfort and palpitations, without risk factors for coronary artery disease, normal biomarkers, and serial unaltered ECGs. The first tracing is similar to the prior ones made over the last 7 years.

This one was made immediatly after cardioversion (there are no artifacts). All the ECGs in her history show the same pattern. She shows Wolff-Parkinson-White with frequent paroxysmal palpitations. Five years ago ablation was proposed to her; however, the patient refused the procedure afraid of complications.

ECG performed immediately after electrical cardioversion



ECG diagnosis: WPW with Ebstein's anomaly. We think that the final portion of the P wave modified the J point and initial part of the ST segment of the QRS complex. See the next slide without WPW.

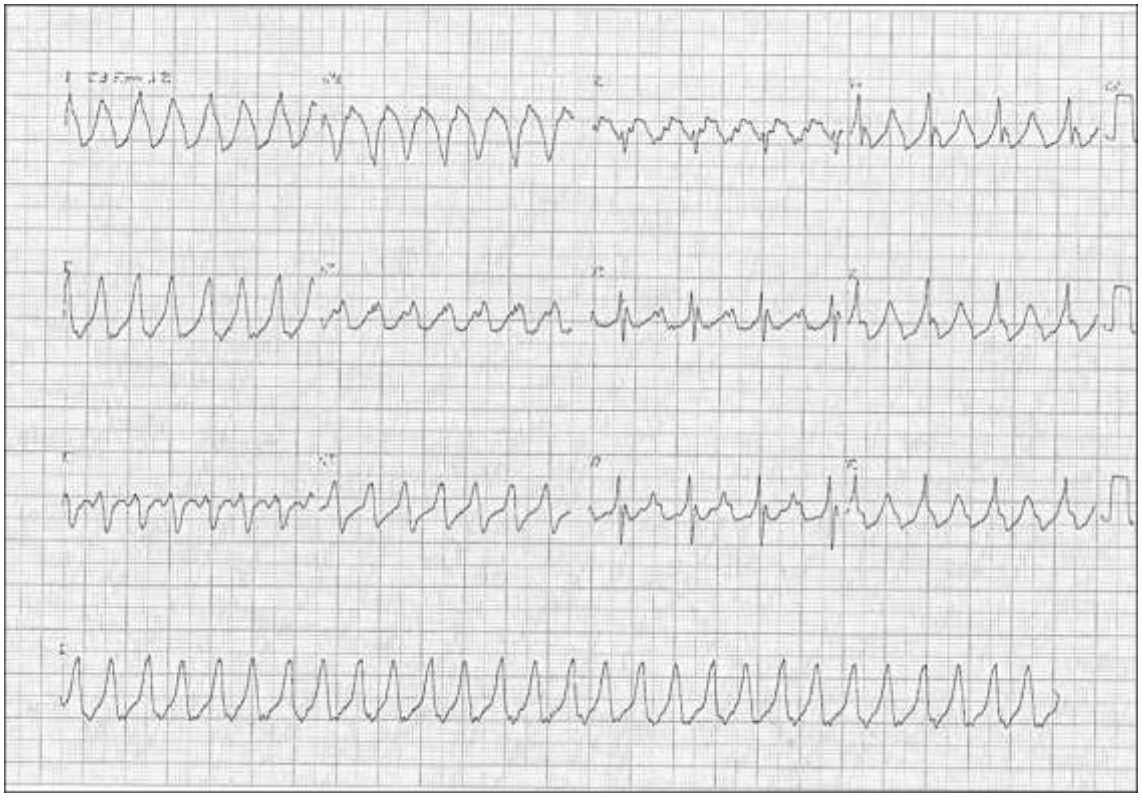


Figure. ECG during tachyarrhythmic event.

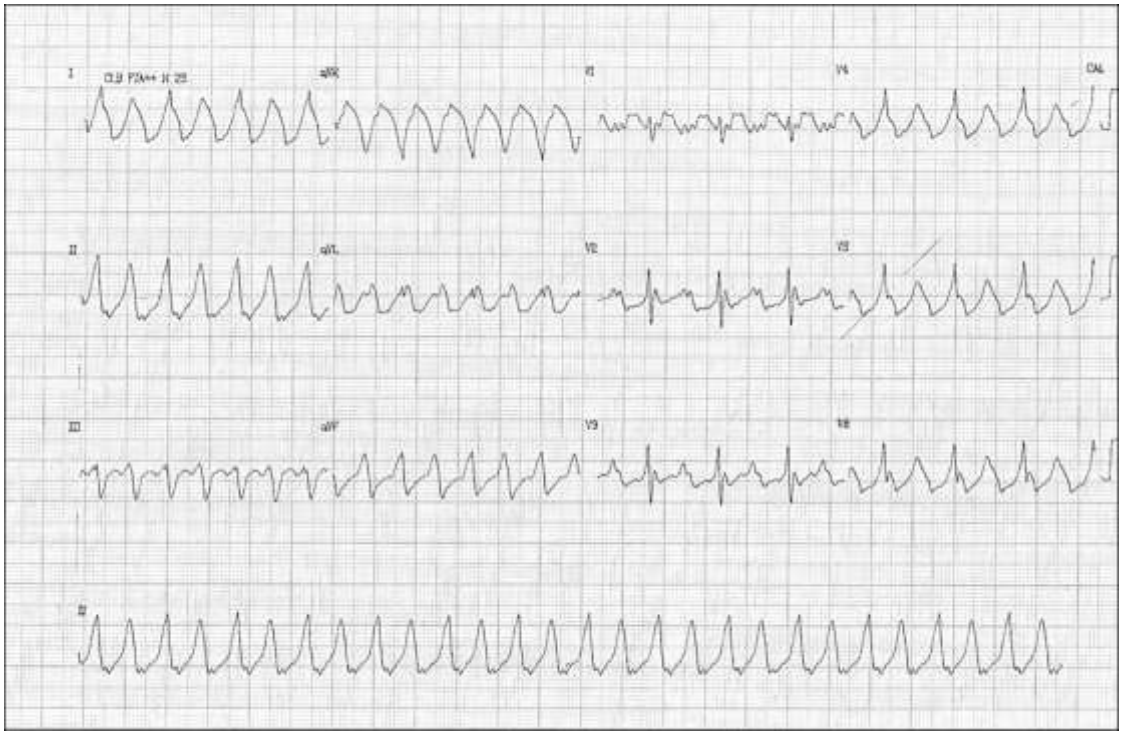


Figure. Another tachyarrhythmic event.

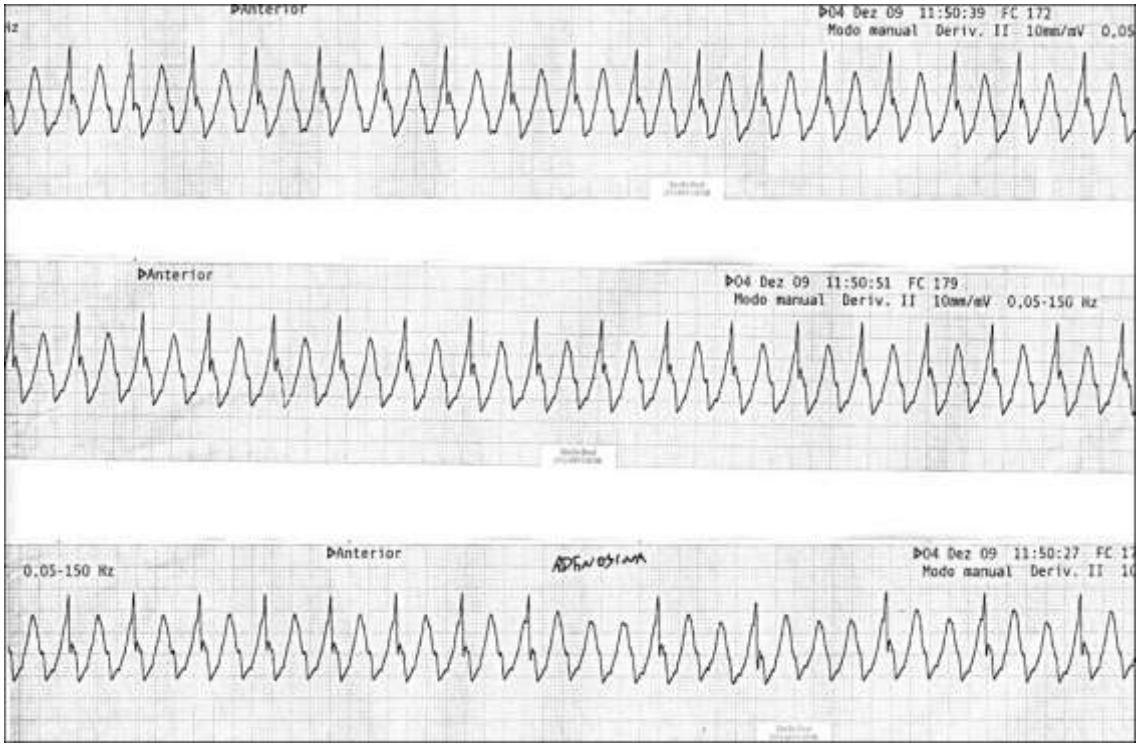


Figure. After adenosine (continuous lead II).

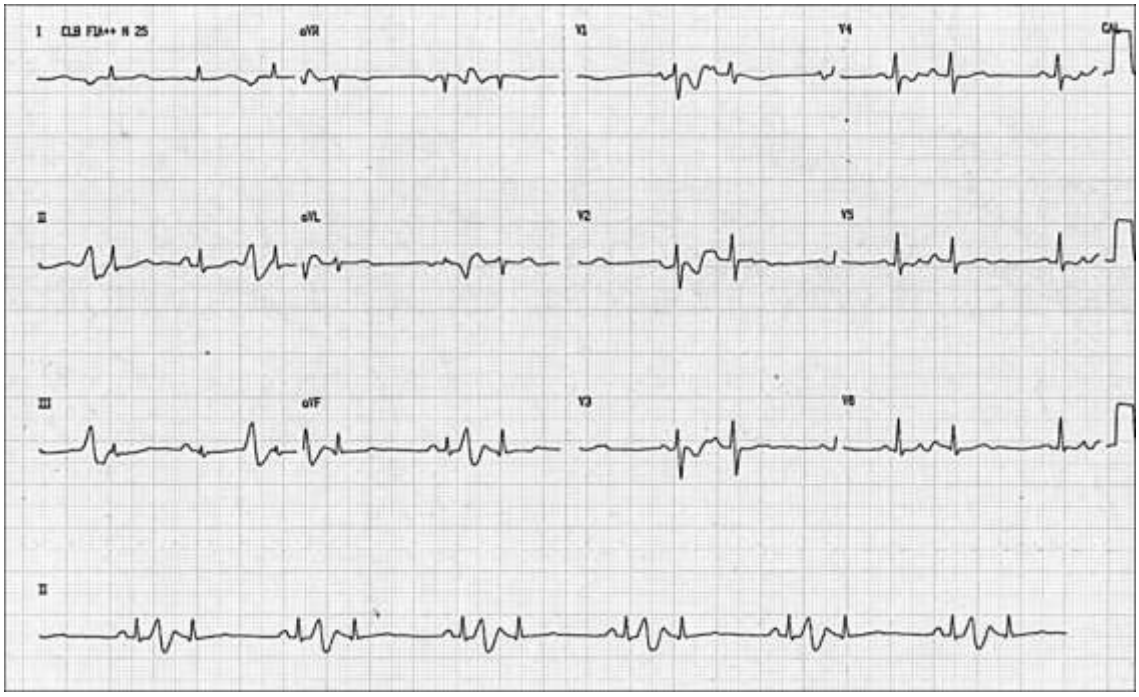


Figure. ECG.

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, Keane et al. 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 (Damjanovic, Dordevic-Radojkovic et al. 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 delta 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).

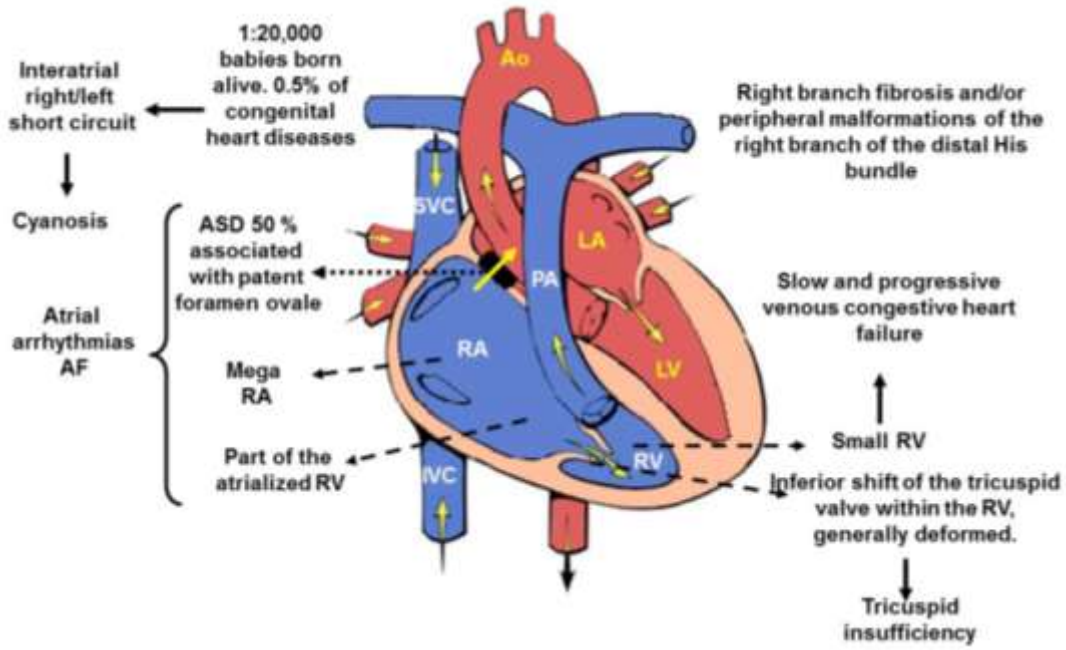
From a series of 224 patients studied by Torres (Torres 2007) at the Ignacio Chávez Institute of Mexico, 64 (28%) had documented tachycardias. Thirty three patients with recurrent tachyarrhythmias had a single right anomalous bundle that could be ablated successfully. Only 21 from these 31 had a typical 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.

ECG/VCG in cyanotic congenital heart diseases

Ebstein's anomaly



Outline of anatomical and hemodynamic features of Ebstein's anomaly: 1) Inferior shift of tricuspid valve: small RV and mega RA: atrial arrhythmia; 2) Right CHF; 3) Right-left short circuit by ASD or patent foramen ovale: cyanosis; 4) Fibrosis of the right branch and peripheral alterations of this branch: bizarre complete RBBB of low voltage.

ECG in Ebstein's anomaly

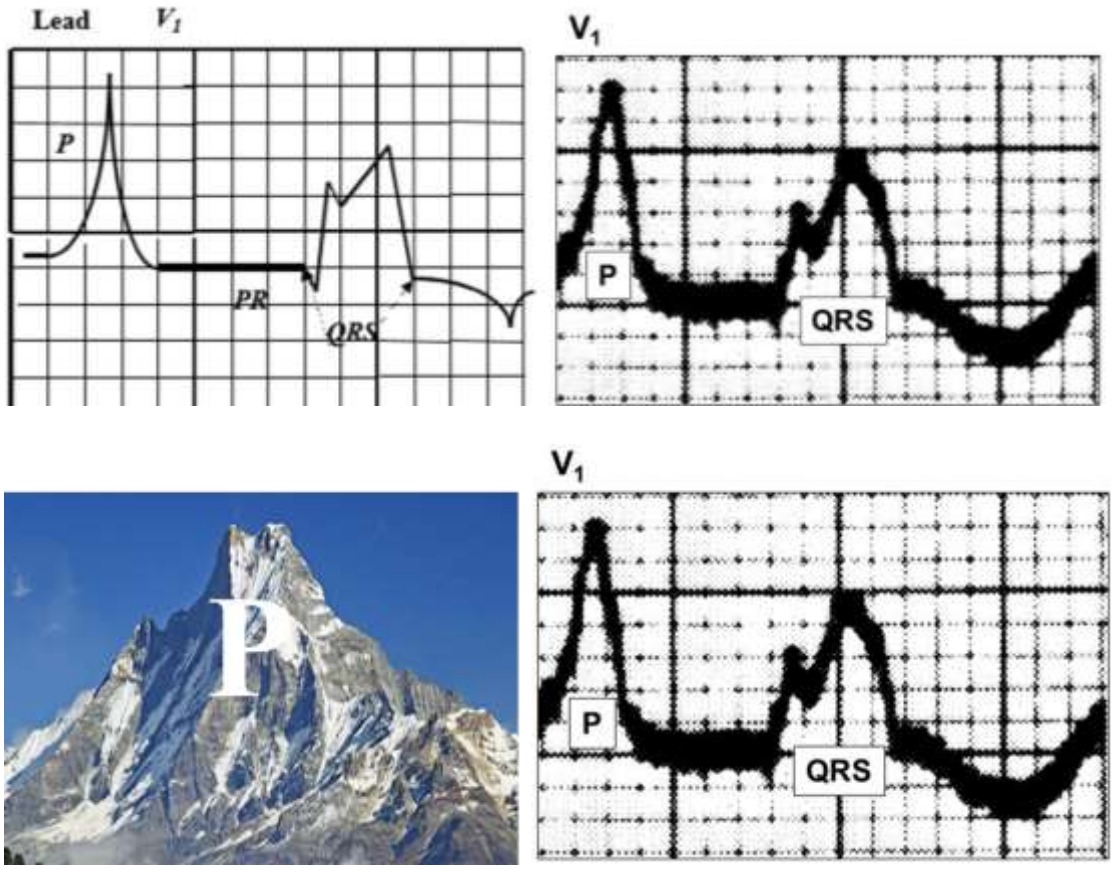
In 30% of the cases, atrial fibrillation, flutter, supraventricular and ventricular paroxysmal tachycardia.

AQRS: generally inferior and to the right between +90º and +130º.

Giant P waves of right atrial enlargement: "Himalayan" P waves, PR interval frequently prolonged: 20%.

IRBBB or 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 in V_1 to V_3. It is frequent to record tri or tetraphasic patterns.

Possible type B WPW with anomalous bundle located in the RV free wall (anomalous bundle between the RA and the RV).



ECG in Ebstein's anomaly (Tede NH, Shivkumar K, Perloff JK, Middlekauff HR, Fishbein MC, Child JS, Laks H. Signal-averaged electrocardiogram in Ebstein's anomaly. Am J Cardiol. 2004 Feb 15;93(4):432-6. doi: 10.1016/j.amjcard.2003.10.058).

Other clinical causes of Himalayan P wave

Family with hypertrophic cardiomyopathy (Stöllberger C, Avanzini M, Siostrzonek P, Kühn P, Winkler WB, Finsterer J. Familial Himalayan p wave and left ventricular hypertrabeculation/noncompaction. Ann Noninvasive Electrocardiol. 2015 Mar;20(2):181-6. doi: 10.1111/anec.12159).

Restrictive cardiomyopathy (Jonjev ZS, Zdravkovic R, Todic M, Dudas V, Rajic J. Himalayan P wave in a patient admitted for cardiac surgery. J Card Surg. 2021

Apr;36(4):1548-1549. doi: 10.1111/jocs.15438; Gupta MD, Girish MP, Tyagi S. Alpine "P" waves in a case of restrictive cardiomyopathy. Int J Cardiol. 2011 Nov 17;153(1):e5-6. doi: 10.1016/j.ijcard.2010.12.102).

Tricuspid atresia (Reddy SC, Zuberbuhler JR. Images in cardiovascular medicine. Himalayan P-waves in a patient with tricuspid atresia. Circulation 2003; 107: 498). This tall P wave is called Gamboa P wave (Gamboa, R., W. M. Gersony and A. S. Nadas (1966). "The electrocardiogram in tricuspid atresia and pulmonary atresia with intact ventricular septum." Circulation 34(1): 24-37).

Severe hypoxemia and emphysema (Vijayakrishnan R, Spodick DH. Himalayan P-waves in the setting of severe hypoxemia and emphysema. Can J Cardiol 2010; 26: 136).

Combined tricuspid and pulmonic stenosis (Davutoğlu V, Kılınç M, Dinçkal MH. Himalayan P waves in a patient with combined tricuspid and pulmonic stenosis. Heart 2003; 89: 1216).

Major electrophysiological abnormalities in Ebstein's anomaly

1. Intratrial conduction disturbance: right atrial enlargement, broad P wave (biatrial enlargement). P waves are abnormal in height, duration and configuration. Dr. Taussing called as Himalayan P waves.

Dr. Taussing, 1st president of AHA, founder of Pediatric Cardiology.

2. PR interval prolongation (all present in this case).
3. In presence of ventricular preexcitation -> short PR interval (not always), occasionally without a delta wave or paroxysmal supraventricular arrhythmia.
4. Atrioventricular nodal conduction disturbance: PR interval prolongation (present in this case).
5. Infranodal conduction disturbance: intra-His or infra-His disturbance, right bundle branch block, bizarre second QRS attached to preceding normal QRS.
6. QRS axis is inferior: although a splintered polyphasic QRS makes axis difficult to determine.
7. Old type B Wolf-Parkinson-White ventricular preexcitation (5 to 25% of cases).
8. Downward displacement of septal tricuspid leaflet is accompanied by discontinuity between central fibrous body and septal A-V ring, creating substrate for preexcitation.
9. The only CCHD consistently associated with preexcitation which is uniformly via right accessory pathway type B WPW.
10. Combination of type B preexcitation + cyanosis: Presumptive evidence.
11. Accessory pathway conduction can be permanent/ intermittent and can occur without delta waves.
12. SVT, AF, AFl: 25 to 30%.
13. Prolonged PR interval may progress to complete heart block.
14. Broad QRS complexes consequence of atrialized right ventricle.
15. 75 to 95%: QRS characterized by right ventricular conduction defect of RBB type.
16. Conduction defect is therefore distal to RBB and is sometimes present despite a septal accessory pathway.

17. A distinctive second QRS complex originates in atrialized RV according to intracardiac mapping.
18. Supraventricular tachyarrhythmias.
19. Atrial fibrillation or flutter.
20. Deep Q waves in inferior leads or/and anterior wall.
21. Deep Q waves appear in II, III and aVF, but most important are right precordial leads Q waves.
22. This distinctive Q wave pattern is indicative of right ventricular intracavitary potentials unusually far leftward as a result of large size of RA
23. Prominent Q waves in right precordial leads can be misleading when adults with Ebstein's anomaly present with chest pain.