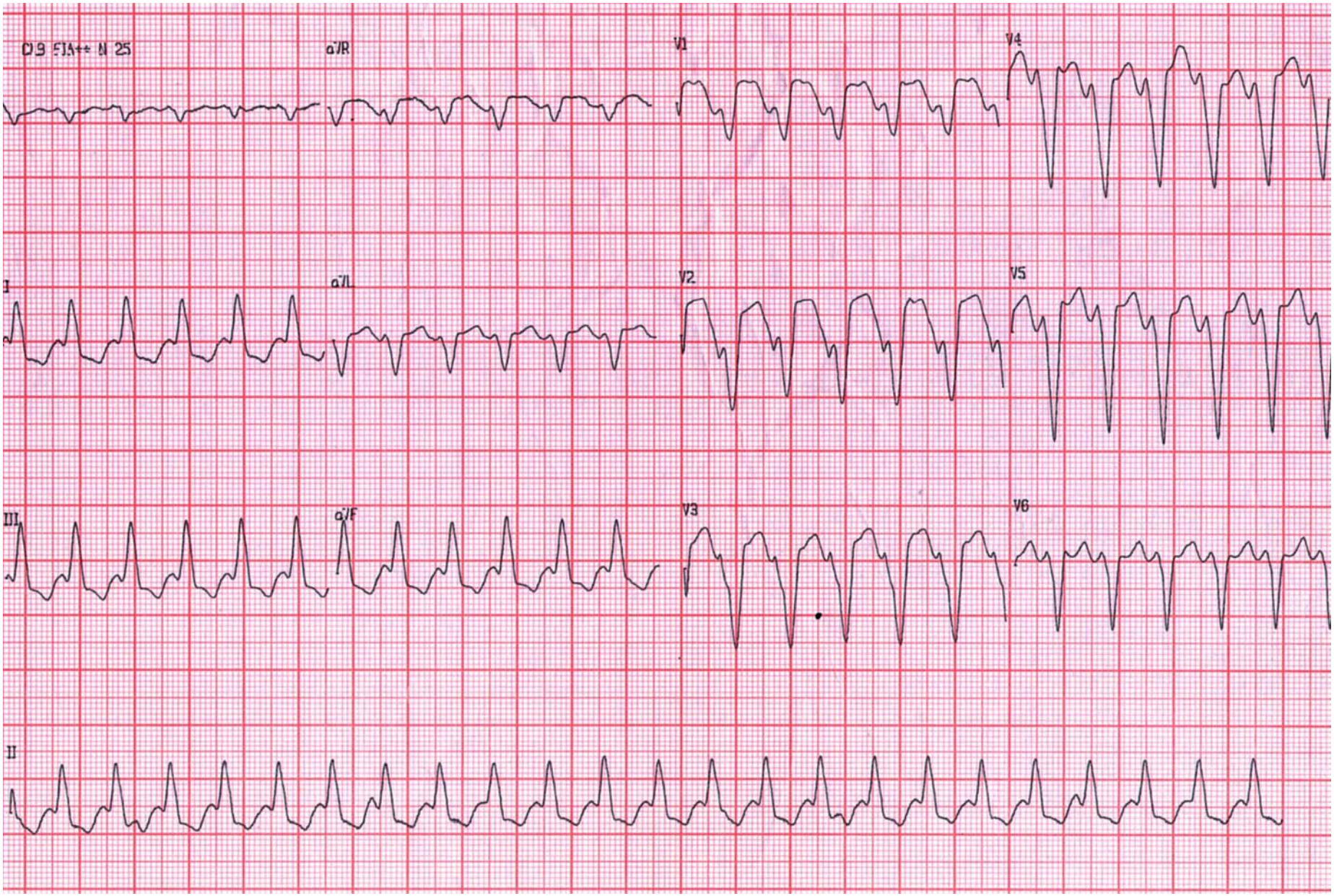
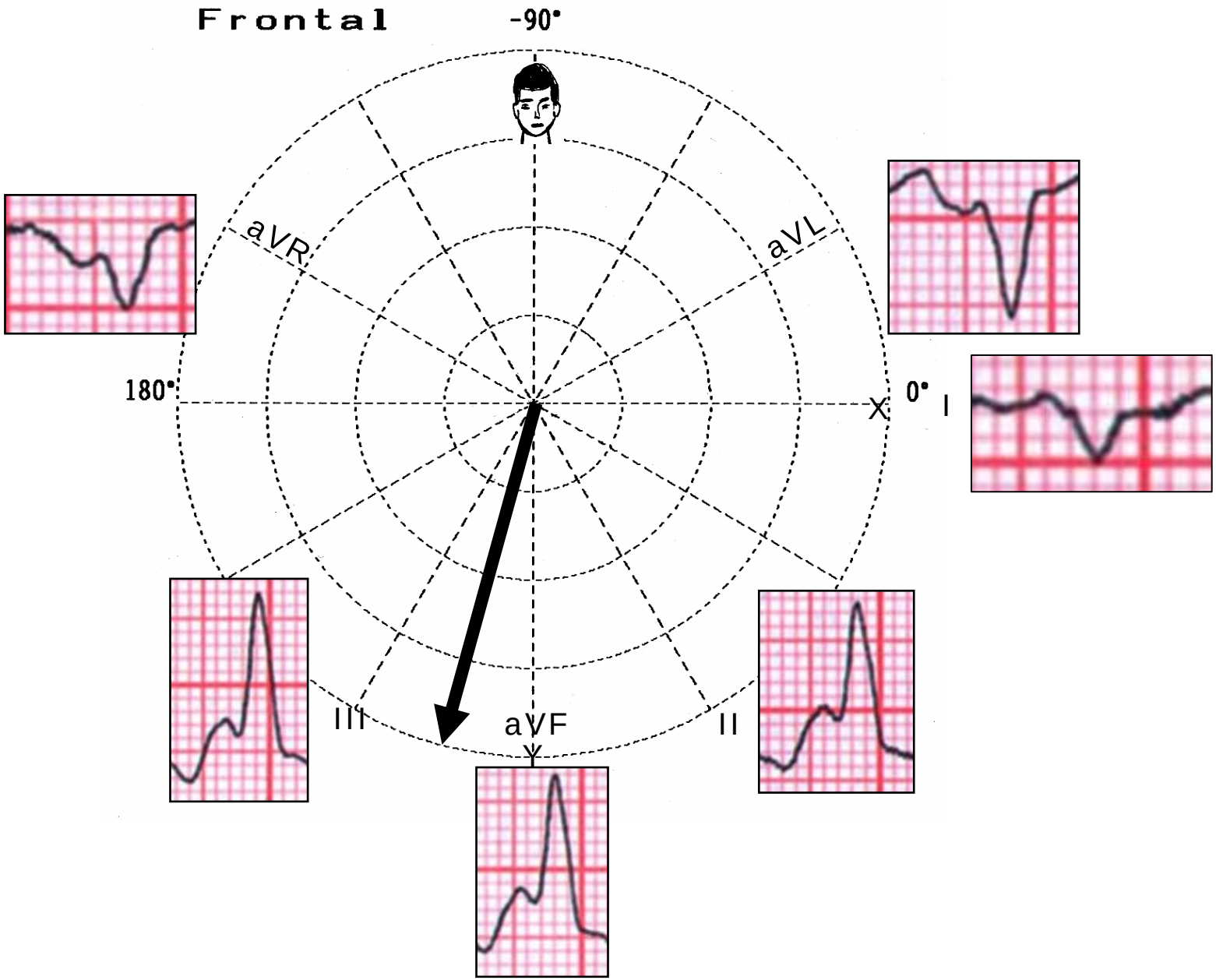
Português
Prezado Prof. Andrés aqui um outro desafio diagnóstico
Paciente, 21anos, com uma história de edema na parte baixa das extremidades, distensão gástrica e palpitações.
O ECG da admissão revelara taquiarritmia de QRS largo revertida para ritmo sinusal com a administração de amiodarona.
Qual o diagnóstico? Presumível clínico e dos ECGs
Dr Raimundo Barbosa de Barros Fortaleza Ceará Brasil

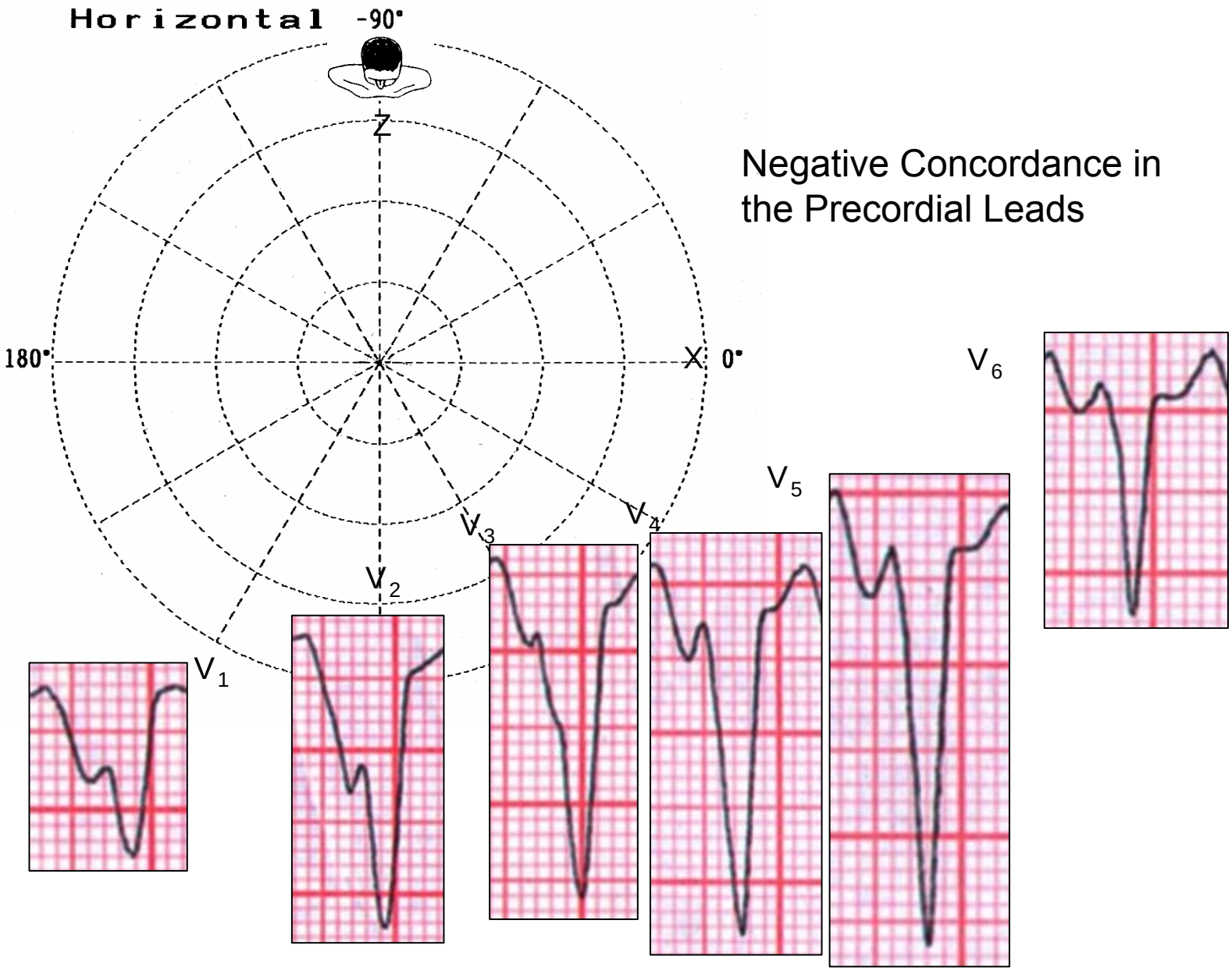
English
Dear Prof. Andrés here another challenger diagnosis
Male patient, 21yo, with a history of lower extremity edema, fullness gastric and palpitations. The electrocardiogram on admission revealed a wide complex tachycardia that was reverted to sinus rhythm after amiodarone.
Which is the presumably clinical and electrocardiographic diagnosis ?

Dr Raimundo Barbosa de Barros Fortaleza Ceará Brazil
Spanish
Querido Prof. Andrés aqui outro desafio diagnóstico:
Paciente masculino, 21años, con história de edema en la parte baja de los mienbros inferiores, distensión abdominal y palpitaciones. El ECG de la admisión mostraba TV ancha que fué revertida con amiodarona.
Cual es el diagnóstico presumible clínico y eléctrocardiográfico?
Dr Raimundo Barbosa de Barros Fortaleza Ceará Brasil

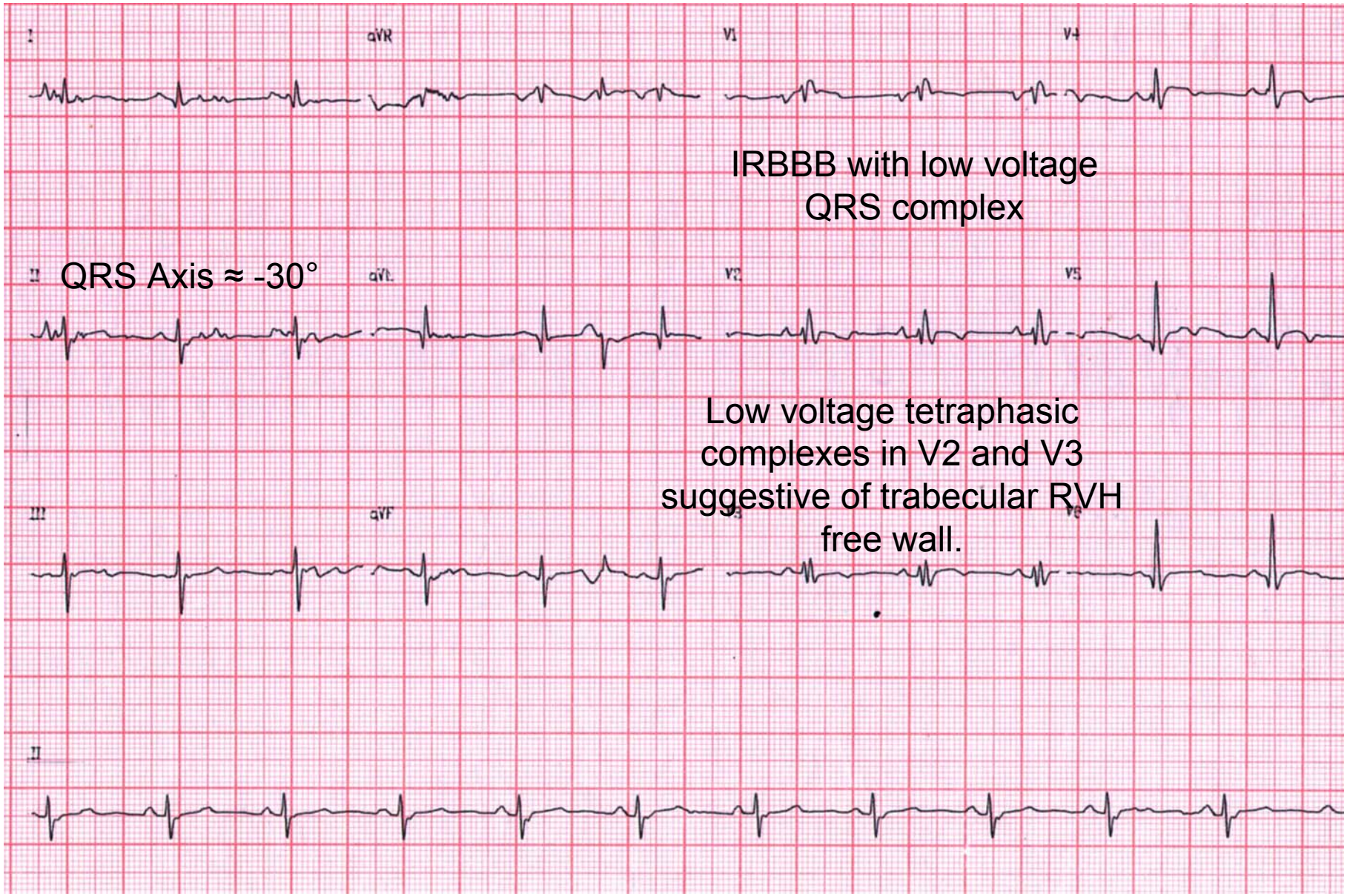


Heart Rate = 150ms; SÂQRS = +100°; QRSd = 120ms;


Frontal
-90°
aVR
aVL
180°
0°
I
III
aVF
II



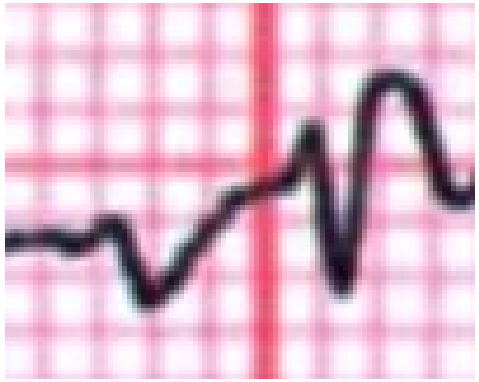
Negative Concordance in the Precordial Leads



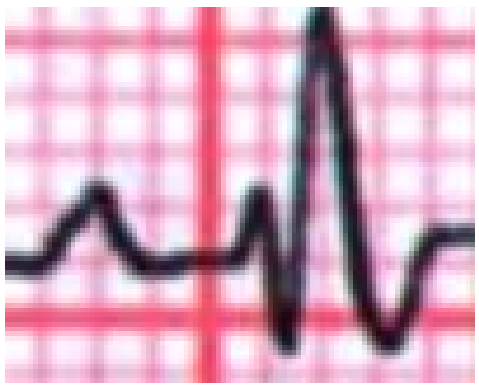
IRBBB with low voltage QRS complex

QRS Axis ≈ -30°

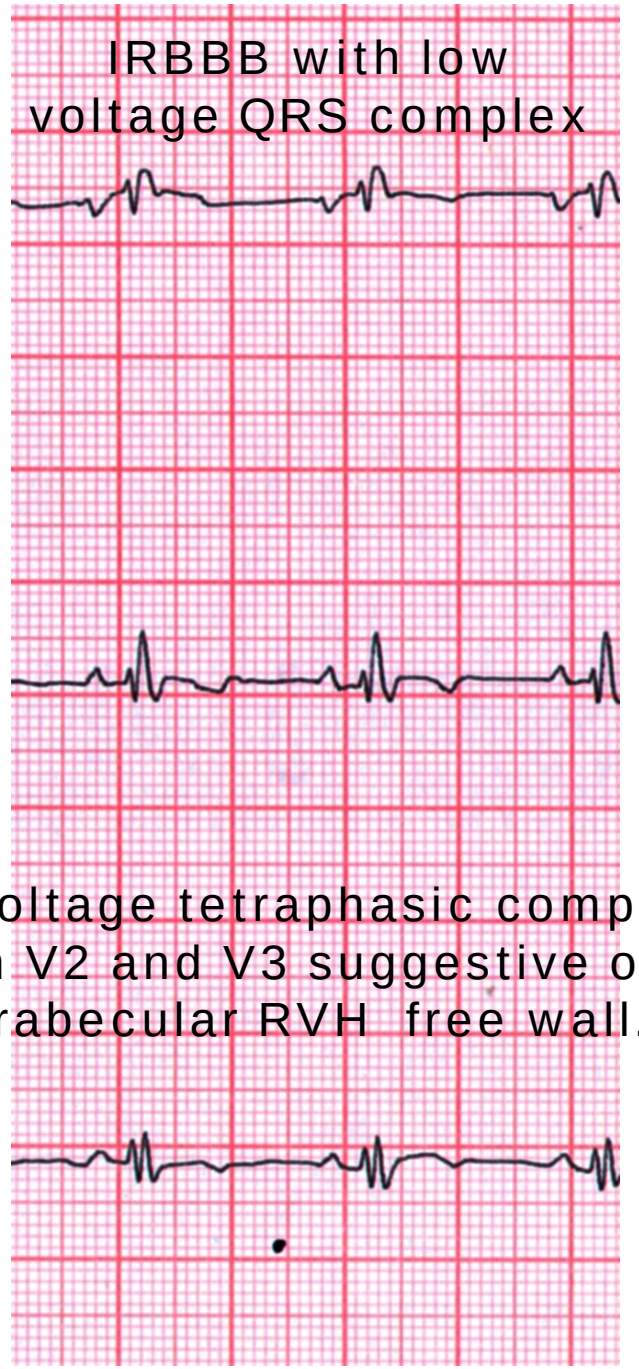
Low voltage tetraphasic complexes in V2 and V3 suggestive of trabecular RVH free wall.



Normal PR Interval



IRBBB with low voltage QRS complex



Final Negative P wave component suggestive of LAE and positive 1.5mm P wave in V2 suggestive RAE. Conclusion: possible biatrial enlargement. Not sure.

Low voltage tetraphasic complexes in V2 and V3 suggestive of trabecular RVH free wall.

Colleagues opinions:

25/7/2010 21:01
Monomorphic VT with focus near RVOT in patient with right structural heart disease.
If the patient has tricuspid insufficiency murmur, Ebstein's anomaly is probable.
Other congenital heart diseases are possible.
In patients without physical examination features, the most probable diagnosis is ARVD.
Echocardiogram is a mandatory initial tool for the diagnosis.

Taquicardia ventricular con origen cercano al tracto de salida del VD en paciente con cardiopatía estructural derecha.
Si tiene soplo de insuficiencia tricuspídea puede ser un Ebstein, también otras congénitas. Si el exámen físico no tiene nada me oriento mas a displasia. El Ecocardiograma es obligatorio para empezar.

Alejandro Cuesta – Uruguay

Second Opinion:
26/7/2010 02:20

ARVD

Prof. Bernard Belhassen
Director, Cardiac Electrophysiology Laboratory
Tel-Aviv Sourasky Medical Center
Tel-Aviv 64239, Israel
Tel/Fax: 00.972.3.697.4418

1- VT in the context of ARVD
2- Repolarization disturbance from V1-V3 are suggestive of ARVD. Epsilon wave is observed in 30% of cases
3- Magnetic resonance imaging (MRI) is a safe and painless test that produces very clear cross-sectional or three-dimensional images of the body's tissues, even through bone and other obstructions. Because of its safety and clarity, the MRI is a very valuable tool that can aid in the diagnosis of a wide range of conditions.
MRI has emerged as important clinical tool for evaluation of myocardial pathology. In addition to providing morphologic and functional information, both imaging modalities have the ability to demonstrate intramyocardial fat, which is the pathological hallmark in ARVC/D
Increased signal intensity of RV free wall; wall motion abnormalities with CINE MRI has emerged as clinical tools for evaluation of myocardial pathology. The device providing morphologic and functional information, has the ability to demonstrate intramyocardial fat, which is the pathological hallmark in ARVC/D
MRI allows the clearest visualization of the heart, in particular because the RV is involved, which is usually more difficult to explore with the other imaging modalities. Furthermore, MRI offers the specific advantage of visualizing adipose infiltration as a bright signal of the RV myocardium. MRI provides the most important anatomic, functional, and morphologic criteria for diagnosis of ARVC/D within one single study. As a result, MRI appears to be the optimal imaging technique for detecting and following patients with clinical suspicion of ARVC/D
4- Endomyocardial biopsy is not indicated because samples obtention is not easy, additionally complication of this procedure is not rare.
5 – Sudden Cardiac Death risk stratification is necessary in this group of patients.
6- Therapeutic options: ICD as primary and secondary SCD prevention. RF ablation in association because it is an evolutive disease with relapse VT or another new VT with other mechanism.
Francisco Femenia

1- TV de TSVD probablemente en el contexto de una miocardiopatia arritmogenica del VD

2- ECG basal con transtornos de la repolarizacion de V1- V3 (estas alteraciones se observan en aproximadamente 50% de los casos con miocardiopatia arritmogenica del VD, la onda epsilon solo en 30%)

3- Angioresonancia para confirmar el diagnostico estructural (altamente sensible, permite evaluar infiltración grasa, zonas de fibrosis, adelgazamiento de la pared ventricular, regiones aquineticas, hipoquineticas o disquineticas y eventualmente dilatacion ventricular derecha o biventricular, la afectacion del VI se puede sospechar cuanto mas extenso es el transtorno de repolarizacion en precordiales) Todos los hallazgos deben siempre ser valorados en el contexto clinico del paciente

4- Sin dudas la histopatologia dara el diagnostico definitivo, pero la biopsia miocardica no esta indicada, ya que al ser una enfermedad progresiva e inicialmente n parches, podemos obtener material de un area "sana", de igual forma no esta excenta de complicaciones)

5- Evaluar contexto clinico y hay que estratificar el riesgo de MS en este grupo de pacientes.

6- Opciones terapeuticas: CDI como prevension primaria o secundaria de MS, Ablación por RF como coadyuvante si fuera necesario, esta discutido la ablación como primera opcion terapeutica ya que al ser una enfermedad evolutiva la recidiva de la TV o la aparicion de otra es elevada.

Saludos femenia

Francisco Femenia – Mendoza - Argentina

FINAL DIAGNOSIS

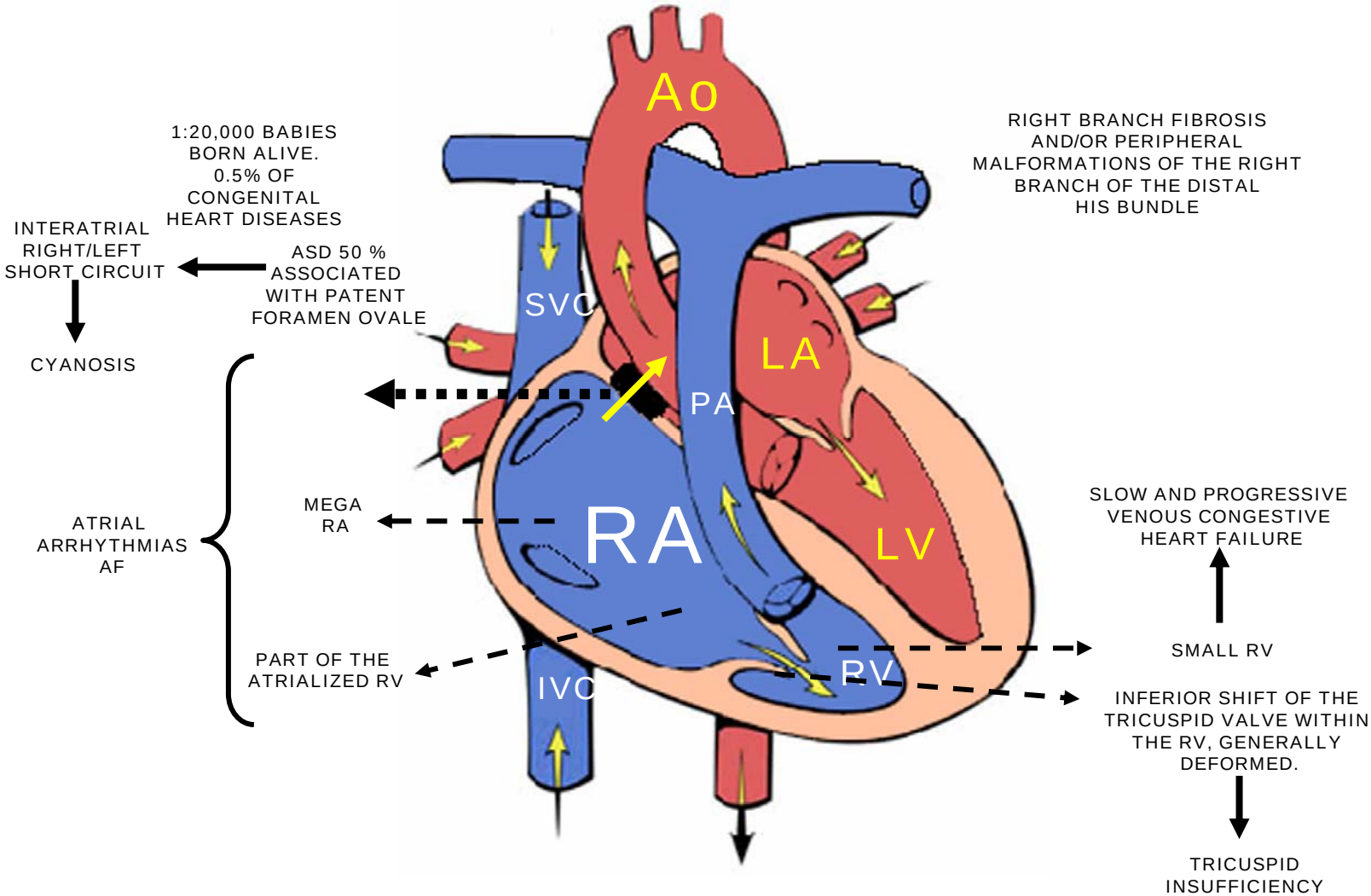
Ebstein's Anomaly of tricuspid valve

Entities With Arrhytmogenic Substrate On Right Ventricular Free Wall

1. Ebstein's Anomaly : irritable Atrialized Right Ventricle with delayed activation.
2. Brugada Syndrome: Conduction delay in RV is a useful marker for identifying high-risk patients with **BrS**[1]**.**
3. Idiopathic ventricular tachycardia (VT) **originating from the right ventricular outflow tract (RVOT) in patients without structural heart diseases**[2] **sensitive to adenosine, vagal maneuvers, and calcium channel blockers.**
4. Arrhythmogenic Right Ventricular Dysplasia **≈40% of patients have mutations in genes encoding proteins in desmosomes, intercellular adhesion junctions which, in cardiac myocytes, reside within intercalated disks. Some desmosomal proteins fulfill roles both as structural proteins in cell-cell adhesion junctions and as signaling molecules in pathways mediated by Wnt ligands. Evidence is increasing that mutations in desmosomal proteins can perturb the normal balance of critical proteins in junctions and the cytosol which, in turn, could alter gene expression by circumventing normal Wnt signaling pathways**[3]**.**
5. Myocardial dysplasia of the right ventricle (Uhl's anomaly)[4]
6. Pulmonary Artery Arrhythmias[5]

1. **Doi A, Takagi M, Maeda K, Conduction Delay in Right Ventricle as a Marker for Identifying High-Risk Patients With Brugada Syndrome. J Cardiovasc Electrophysiol. 2009 Dec 28. [Epub ahead of print]**
2. **Shimizu W. Arrhythmias originating from the right ventricular outflow tract: how to distinguish "malignant" from "benign"? Heart Rhythm. 2009 Oct;6:1507-1511.**
3. **Saffitz JE, Asimaki A, Huang H.Arrhythmogenic right ventricular cardiomyopathy: new insights into mechanisms of disease. Cardiovasc Pathol. 2010 Jan 4. [Epub ahead of print]**
4. **Yamashina Y, Yagi T, Namekawa A, Clinical and Electrophysiological Difference Between Idiopathic Right Ventricular Outflow Tract Arrhythmias and Pulmonary Artery Arrhythmias.J Cardiovasc Electrophysiol. 2009 Sep 28. [Epub ahead of print]**
5. **Sigrist T, Bär W, Zink P. Myocardial dysplasia of the right ventricle (Uhl's anomaly) as a possible cause of sudden, unexpected deathZ Rechtsmed. 1988;100:176-189.**

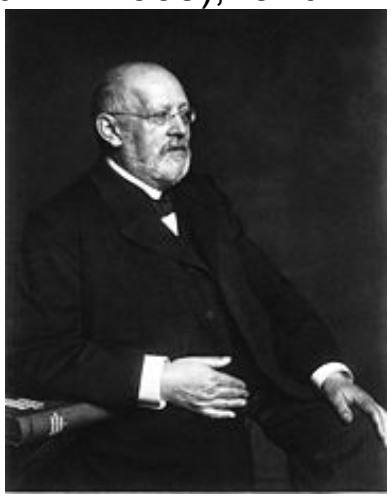
EBSTEIN'S ANOMALY OF THE TRICUSPID VALVE



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.

History

In 1866 Wilhelm Ebstein (November 27, 1836, Jauer, Prussian Silesia – October 22, 1912) first described a patient with cardiac defects typical of Ebstein anomaly. He was a German physician. He studied medicine at the universities of Breslau and Berlin, graduating from the latter in 1859. In that year he was appointed physician at the, Breslau; in 1868, chief physician at the municipal poorhouse; in 1869, privatdocent; in 1874, professor in Göttingen University (which chair he still held in 1903); and in 1877, director of the university hospital and dispensary.



Wilhelm Ebstein

In 1927, Alfred Arnstein suggested the name Ebstein's anomaly for these defects.

In 1937, Yates and Shapiro described the first case of the anomaly with associated radiographic and electrocardiographic data.

1. **Ebstein W. Ueber einen sehr seltenen Fall von Isufficienz der Valvula tricuspidalis, bedingt durch eine angeborene hochgradige Missbildung derselben. *Arch Anat Physiol*. 1866;238-254.**

EPIDEMIOLOGY

Incidency: 1:20.000 born alive.
Frequency: Ebstein anomaly probably accounts for 0.5% of cases of congenital heart diseases.
Mortality/Morbidity: The natural course of the disease varies according to the severity of tricuspid valve displacement.Patients presenting in infancy generally have severe disease and unfavorable prognosis.[1;2]
Mean age of presentation is in the middle teenage years. According to older observational data, approximately 5% of these patients survive beyond age 50 years. The oldest recorded patient lived to age 85 years.
Ethnic group: More common in caucasian.
Gender: No specific sex predominance exists.
Age: Ebstein anomaly can present at various stages of life.
Fetal life: Ebstein anomaly is usually diagnosed incidentally by echocardiography.
Neonatal life and infancy: Ebstein anomaly presents with cyanosis and/or severe heart failure; typically, symptoms present in infancy improve as pulmonary vascular resistance decreases.
Adult life: Ebstein anomaly presents with fatigue, exertional dyspnea, cyanosis, tricuspid regurgitation and/or right heart failure, and palpitations; arrhythmias are common.

1. **Armengol Rofes AJ, Serrano Durán M, Albert Brotons DC, Sánchez López C, Casaldáliga Ferrer J, Girona Comas JM. [Ebstein's anomaly of the tricuspid valve. Apropos 35 cases] An Esp Pediatr. 1996 Feb;44(2):139-44. Spanish.**
2. **Khositseth A, Khowsathit P. Factors affecting mortality in Ebstein's anomaly of the tricuspid valve. J Med Assoc Thai. Nov 1999;82 Suppl 1:S10-5.**

Anamnesis

Patients can have a variety of symptoms related to the anatomical abnormalities of Ebstein anomaly and their hemodynamic effects or associated structural and conduction system disease.

Cyanosis Fairly common and frequently due to right-to-left shunt at the atrial level and/or severe heart failure.Transient in neonatal life with recurrence in adult life.May appear for the first time in adult life

Transient appearance/worsening of cyanosis in adult life due to paroxysmal arrhythmias
Once apparent, progressively worsens

Fatigue and dyspnea: These are due to poor cardiac output secondary to right ventricular failure and decreased left ventricular ejection fraction.

Palpitations and sudden cardiac death

Due to paroxysmal supraventricular tachycardia (SVT) in as many as one third of patients
Fatal ventricular arrhythmias, which may be due to the presence of accessory pathways

Symptoms of right heart failure: These include ankle edema and ascites.

Other less common presenting symptoms

- Brain abscess due to right-to-left shunt
- Bacterial endocarditis
- Paradoxical embolism
- Stroke
- Transient ischemic attacks

Inspection: Physical findings, like the symptoms, span a spectrum from subtle to dramatic.
Cyanosis and clubbing - Varying degrees of cyanosis at various times in life and transient worsening with arrhythmias
Precordial asymmetry Usually left parasternal prominence and occasionally right parasternal prominence Absent left parasternal (ie, right ventricular) lift an important negative sign.

Jugular venous pulse May be normal owing to a large, thin-walled right atrium, which can absorb the volume and pressure transmitted from the right ventricle through an incompetent tricuspid valve Large *a* and *v* waves late in the course of the disease, with development of right heart failure

Arterial pulses Usually normal Diminished volume late in the course of the disease due to severe right heart failure and decreased left ventricular stroke volume

Heart sounds First heart sound is widely split with loud tricuspid component secondary to delayed closure of the elongated anterior tricuspid leaflet, which has an increased excursion. Mitral component may be soft or absent in the presence of prolonged PR interval.
Second heart sound usually is normal but may be widely split when the pulmonary component is delayed due to RBBB.
Third and fourth heart sounds are commonly present, even in the absence of congestive heart failure (CHF).
Summation of third and fourth heart sounds, especially with prolonged PR interval, can mimic an early diastolic murmur.
The holosystolic murmur of tricuspid regurgitation is heard maximally at the lower left parasternal area and sometimes at the apex owing to the displaced location of the tricuspid valve; murmur intensity and duration increase during inspiration.

ECG FEATURES OF EBSTEIN'S ANOMALY OF THE TRICUSPID VALVE

(Classic right sided Ebstein's malformation)

Ebstein's anomaly is a rare congenital cardiac defect initially described by Wilhelm Ebstein in 1866 [1; 2; 3] associated with both displacement and incompetence of the tricuspid valve.

The abnormal development of the tricuspid valve in patients with Ebstein's anomaly results in several activation abnormalities including prolonged intraatrial conduction[4] (P-A interval prolongation from 50, to 65 ms), first-degree heart block (31%), rarely secondary to atrioventricular nodal conduction prolonged infranodal conduction, delay or intra-His delay, peripheral or divisional right bundle branch block(58%), ventricular preexcitation (18%), and nonspecific intraventricular conduction delay/block (15%)[5] and irritable atrialized right ventricle with delayed activation.

1. **Ebstein W. Ueber einen sehr seltenen Fall von Isufficienz der Valvula tricuspidalis, bedingt durch eine angeborene hochgradige Missbildung derselben. *Arch Anat Physiol*. 1866;238-54.**
2. **Schiebler GL, Gravenstein JS, Van Mierop LH. Ebstein's anomaly of the tricuspid valve. Translation of original description with comments. *Am J Cardiol*. Dec 1968;22:867-73.**
3. **Mann RJ, Lie JT. The life story of Wilhelm Ebstein (1836-1912) and his almost overlooked description of a congenital heart disease. *Mayo Clin Proc*. Mar 1979;5:197-204.**
4. **Torres PI. Wolff-Parkinson-White syndrome in Ebstein's anomaly Arch Cardiol Mex. 2007 Apr-Jun;77 Suppl 2:S2-37-S2-39. CardiovascPathol. 2008 May-Jun;17:166-171**
5. **Barbara DW, Edwards WD, Connolly HM, et.al. Surgical pathology of 104 tricuspid valves (2000-2005) with classic right-sided Ebstein' malformation. Cardiovasc Pathol. 2008 May-Jun;17:166-171.**

Rhythm: usually normal sinus but may intermittently demonstrate in 25% to 30% of cases reentrant SVT, paroxysmal SVT, Atrial Flutter (AFL), Atrial Fibrillation (AF) or rarely Ventricular Tachycardia (VT). Are frequents SVT with broad QRS complexes by anterograde atrioventricular conduction.

P waves: Abnormal, giant very high P waves “like-Himalaya mountain”[1] called by Dr. Taussing “Himalayan P-waves”[2], consistent with of right atrial enlargement. The P wave is > 3mm (0.3 mV) in near 50% of cases[3]. Tall P waves (≥ 2.5 mm) are attributable to right atriomegaly.
This P are high, broad and with particular pick shape consequence of right atrial prolonged activation. A prolonged P-wave duration is eventually registred4. Himalayan P waves are not pathognomonic of Ebstein anomaly because they are observed in tricuspid atresia[4] and in combined tricuspid with pulmonic stenosis [5 ; 6].
Absence of P wave is observed in cases of atrial fibrillation and persistent atrial standstill in familial Ebstein's anomaly[7]. P-dextro-atriale and bizarre low RBBB without right ventricular overload almost certainly constitutes a pathognomonic finding in Ebstein's anomaly.
P-pulmonale like-pattern is registered in: asthenic habitus, severe hyperkalemia, enhanced sympathetic tone and in cyanosis state.

1. **Kaushik ML, Sharma M, Kashyap R. 'Himalayan' p wave. J Assoc Physicians India. 2007;55: 856.**
2. **Bialostozky D, Horwitz S, Espino-Vela J. Ebstein's malformation of the tricuspid valve. A review of 65 cases. Am J Cardiol. 1972 Jun;29:826-836.**
3. **Armengol Rofes AJ, Serrano Durán M, Albert Brotons DC,** et al**. Ebstein's anomaly of the tricuspid valve. Apropos 35 cases An Esp Pediatr. 1996; 44: 139-144.**
4. **Reddy SC, Zuberbuhler JR. Images in cardiovascular medicine. Himalayan P-waves in a patient with tricuspid atresia. Circulation. 2003;107:498.**
5. **Davutoglu V, Kilinc M, Dinckal MH. Himalayan P waves in a patient with combined tricuspid and pulmonic stenosis. Heart. 2003 Oct;89:1216.**
6. **Johnson CD. Himalayan P waves. P R Health Sci J. 2003 Sep;22:315-320.**
7. **Piérard LA, Henrard L, Demoulin JC. Persistent atrial standstill in familial Ebstein's anomaly. Br Heart J. 1985 Jun;53:594-597.**

PR interval: frequently prolonged: 20%. PR interval may be normal or short in patients with type B WPW syndrome. Eventually is observed normal PR interval with delta wave.
Very high P waves are observed. (the Himalayan mountain system are the planet's highest peaks around the world's).

QRS axis: generally inferior and to the right between +90° and +130°.

QRS pattern: IRBBB or CRBBB of low voltage, bizarre aspect, initial Q wave in right and middle precordial leads (from V1 to V4) is recorded in 50% of the cases. It is frequent to record tri or tetraphasic patterns. Bizarre low voltage right bundle branch block pattern **with initial** q wave in QRS complexes of V1 and V2 leads.
Reduced amplitude of R-wave deflections in V3R and V1 is considered typical.
Possible type B WPW pattern (5% to 25% of cases) with anomalous pathway located in the RV free wall (anomalous pathway between the RA and the RV)[1]. A more than 30-msec interval between the end of the initial slurring (delta) and the vertex of the R wave in the left unipolar leads or the main axis of the vectorcardiographic ventricular curves, allows us to infer the coexistence of left ventricular hypertrophy. Radiofrequency catheter ablation result in appearance of RBBB in ≈ 95% patients. The absence of RBBB in patients with Ebstein's anomaly and recurrent tachycardia had a 98% sensitivity and 92% specificity for the diagnosis of an anomalous accessory A-V pathway. One third of patients with Ebstein's anomaly and symptomatic tachyarrhythmias have minimal or absent ECG features of ventricular preexcitation. In these patients, the absence of RBBB pattern is a strong predictor of an accessory A-V pathway. Higher P waves and wider the QRS complexes are registered in more severe cases of Ebstein's anomaly of the tricuspid valve.

1. **de Micheli A, Iturralde P, Medrano GA. On the electrical manifestations of some heart diseases associated with ventricular preexcitationArch Cardiol Mex. 2006 Oct-Dec;76 Suppl 4:S137-143.**

Arrhythmias: Ebstein's anomaly is characterized by a displacement of the tricuspid valve toward apex, because of anomalous attachment of the tricuspid leaflets.
There are type B of Wolff-Parkinson-White (WPW) syndrome and paroxysmal atrioventricular reciprocating tachycardia in more than a half of all **patients**1. AF, AFL, paroxysmal SVT2 and VT. Ebstein's anomaly is one of the reasons of paroxysmal AF, especially in young persons with WPW syndrome. Electrophysiology studies: can delineate accessory conduction pathways. Right-sided anomalous pathways are more common and 50% of patients have multiple pathways.
From a series of 224 patients studied by Torres et. at3 from 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.

1. **Damjanović MR, Dordević-Radojković D, Perisić Z, et al. Ebstein's anomaly as a cause of paroxysmal atrial fibrillationVojnosanit Pregl. 2008 Nov;65:847-850.**
2. **Macfarlane AJ, Moise S, Smith D. Caesarean section using total intravenous anaesthesia in a patient with Ebstein's anomaly complicated by supraventricular tachycardia. Int J Obstet Anesth. 2007 Apr;16:155-159.**
3. **Torres PI. Wolff-Parkinson-White syndrome in Ebstein's anomaly. Arch Cardiol Mex. 2007 Apr-Jun;77 Suppl 2:S2-37-S2-39.**

- 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 AV block.

- Ebstein's anomaly possibly shows *WPW*, type B, with anomalous pathway located in the RV free wall (*between the RA and the RV*) or septal posterior.

- An anomalous pathway 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[1]. This is the congenital heart disease most associated to *WPW*.

- Patients with anomalous pahtways 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[2].

1) **Deal BJ, Keane JF, Gillette PC, Garson A Jr. Wolff-Parkinson-White syndrome and supraventricular tachycardia during infancy: management and follow-up. J Am Coll Cardiol. 1985; 5: 130-135.**
2) **Damjanović MR, Dordević-Radojković D, Perisić Z, Apostolović S, Koraćević G, Pavlović M, Tomasević MJanković R. Ebstein's anomaly as a cause of paroxysmal atrial fibrillation. Vojnosanit Pregl. 2008 Nov;65:847-850.**

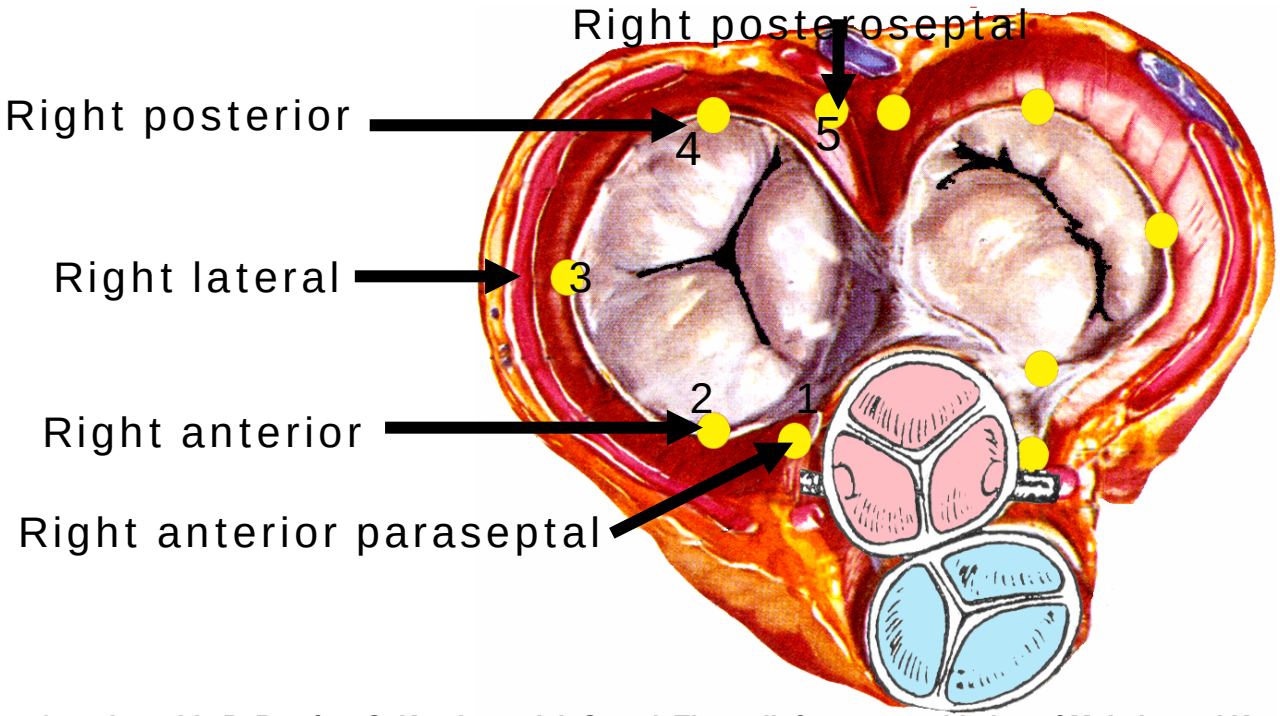
Electrophysiologic characteristics of five patients with Ebstein's anomaly of the tricuspid valve were studied by Kastor et al[1]. In three cases intra-right atrial conduction was prolonged (P-A intervals prolongation), a finding which reflected the presence of a characteristically large right atrium.
The bundle of His electrogram was recorded in its usual anatomical location.
Atrioventricular nodal conduction was prolonged in only one case(20%).
Intra-His delay was observed in two cases, infranodal conduction was prolonged in four cases. The anatomical abnormalities were least severe in the only patient with a normal H-V interval (50 ms). The prolonged H-V interval was thought to result from stretching of the conduction system over the Atrialized Right Ventricle (ARV). The late depolarization during the splintered R' of the ECG found during intracardiac mapping of the ARV in three patients confirms the theory that the ARV produces the "second QRS" typically seen in this anomaly.
The ARV was particularly irritable, and ventricular fibrillation was produced in two patients during catheter manipulation in this area.
In one case the ARV had a shorter refractory period than the body of the right ventricle.
Re-entrant supraventricular tachycardia was induced in the only patient with WPW syndrome. Patients with Ebstein's anomaly of the tricuspid valve usually have: normal position of the bundle of His, prolonged intra-right atrial conduction, prolonged infranodal conduction, and irritable ARV with delayed activation[2].

1. **Kastor JA, Goldreyer BN, Josephson ME, et al. Electrophysiologic characteristics of Ebstein's anomaly of the tricuspid valve. Circulation. 1975 Dec;52:987-995**

2. **Hernandez FA, Rochkind R, Cooper HR. The intracavitary electrocardiogram in the diagnosis of Ebstein's anomaly. Am J Cardiol. 1958Feb;1:181-190.**

The location of the anomalous accessory pathway in Ebstein's anomaly could be:

I) Right anterior (the most frequent one) in point 2 of Gallagher
II) Right anterior paraseptal in point 1
III) Right lateral in point 3
IV) Right posterior in point 4
V) Right posteroseptal in point 5.
VI) There are rare cases of Ebstein's anomaly with pre-excitation of the Mahaim type or right atriofascicular connection[1]: 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: A) ventricular node (connections); B) fasciculoventricular (tracts).



1. **Iturralde P, Ramírez S, Kershenovich S,et al. The radiofrequency ablation of Mahaim and Kent fibers during the repair of Ebstein's anomaly. Arch Inst Cardiol Mex. 1994 Jan-Feb;64:37-44.**