

PECTUS EXCAVATUM WITH SPONTANEOUS
TYPE 1 ECG BRUGADA PATTERN OR
BRUGADA LIKE PHENOTYPE:

ANOTHER BRUGADA ECG PHENOCOPY

ANDRÉS RICARDO PÉREZ RIERA MD
**Chief of the Sector of Electro-Vectocardiography of
the Discipline of Cardiology, School of Medicine, ABC
Foundation – Santo André – São Paulo – Brazil.**

PHENOCOPY

Definitions

- A phenotype that is not genetically controlled but looks like a genetically controlled phenotype.
- An environmentally induced phenotype that resembles the phenotype produced by a mutation.
- A phenotypic variation that is caused by unusual environmental conditions and resembles the normal expression of a genotype other than its own.

Case Report 1

Young, male 19-year-old patient: asymptomatic, who presented at our office of an evaluation prior to the practice of sports.

Negative personal and family history for syncope or sudden death in first-degree relatives younger than 45 years old.

Physical examination: The visual or ectoscopic test of the chest, reveals very significant Pectus excavatum with the lower third of the sternum more affected than the higher third, which was virtually normal. He mentioned that such deformity was noticed since his birth, with a progressive worsening. No first-degree relative was a carrier of pectus excavatum, Marfan syndrome, or Poland syndrome.

Cardiac auscultation: mild systolic murmur ++ in pulmonary focus. No click in the mitral valve.

Lung sounds appear diminished at both bases.

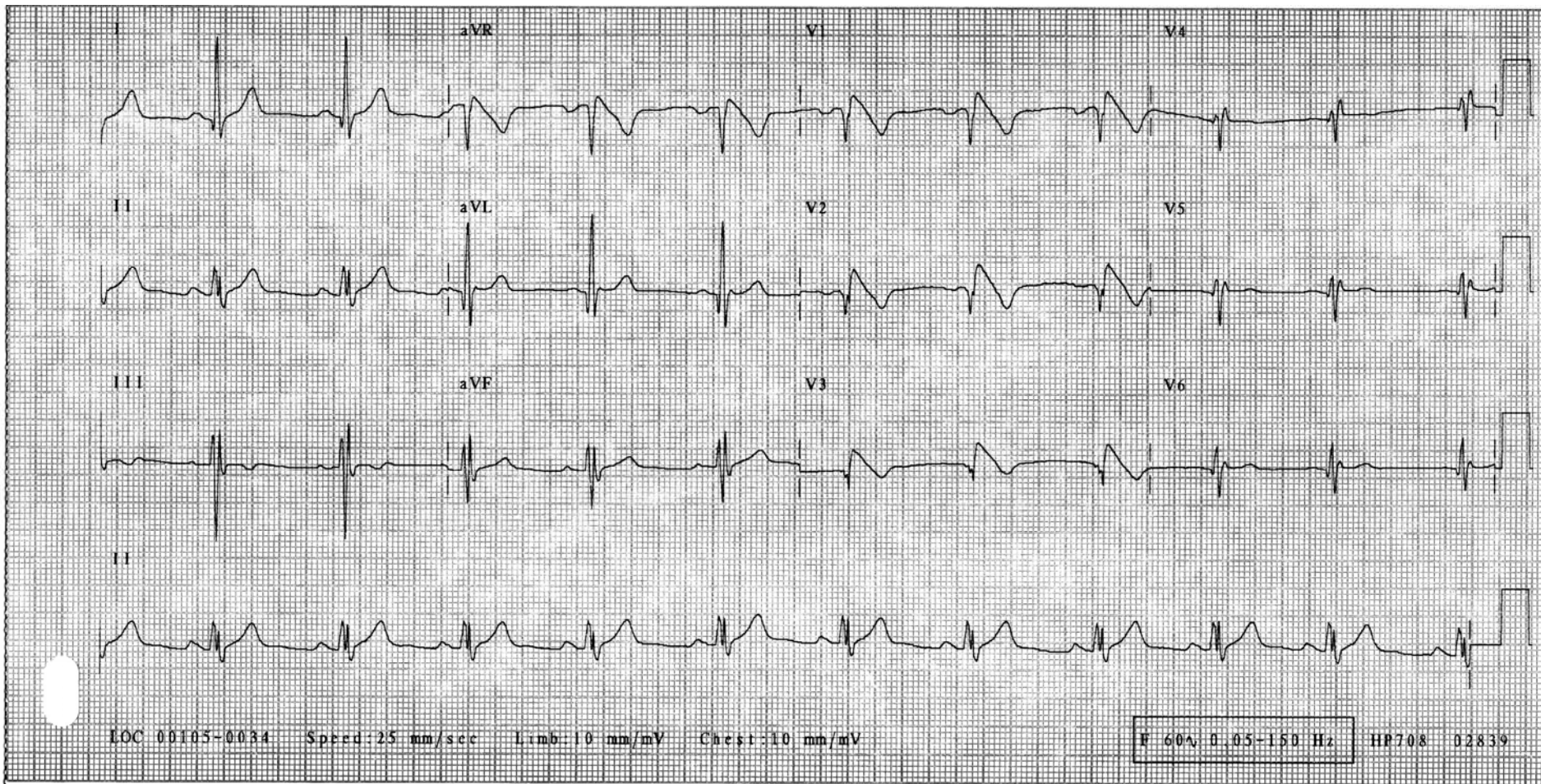
The ECG revealed spontaneous type 1 Brugada-like pattern, and several of the typical elements of pectus excavatum: completely negative P wave in V1 and V2, qR pattern from V1 to V3, and right bundle branch pattern.

The echocardiogram was normal.

The X-Ray of PA chest showed a pseudo-increase of the cardiac area and lateral projection, significant decrease of the antero-posterior diameter of the chest.

FUNCTIONAL RESPIRATORY TEST: Mild restrictive ventilatory disorder. The pulmonary volumes are reduced and there is reduction of total pulmonary capacity that indicates restrictive disorder (Mild restrictive ventilatory disorder.).

Name: BNE; **Gender:** Male; **Age:** 19 years old; **Ethnic background:** White/Caucasian; **Weight:** 65Kg; **Height:** 1,72m; **Biotype:** Normoline; **Date:** 03/4/2009.



Clinical diagnosis: Pectus excavatum

Electrocardiographic diagnosis: see next slide

ELECTROCARDIOGRAPHIC DIAGNOSIS

Rhythm: Normal sinus;

Heart rate: 67bpm;

P wave: P axis + 28° on frontal plane, entirely negative in leads V1-V2 and perpendicular to V3;

PR interval duration: 177ms;

QRS: QRSd: 122ms, **QRS axis:** + 60° on frontal plane. **QRS complex:** QR pattern from V₁ to V₃ and absence of the normal increase of R voltage waves on precordial leads.;

ST/T: ST segment elevation coved to the top ≥ 2mm on right precordial leads and aVR lead (aVR_{sign}).; T axis + 28° on frontal plane and with negative T polarity from V1 to V3;

QT/QTc: intervals: 375/390ms.

Conclusions:

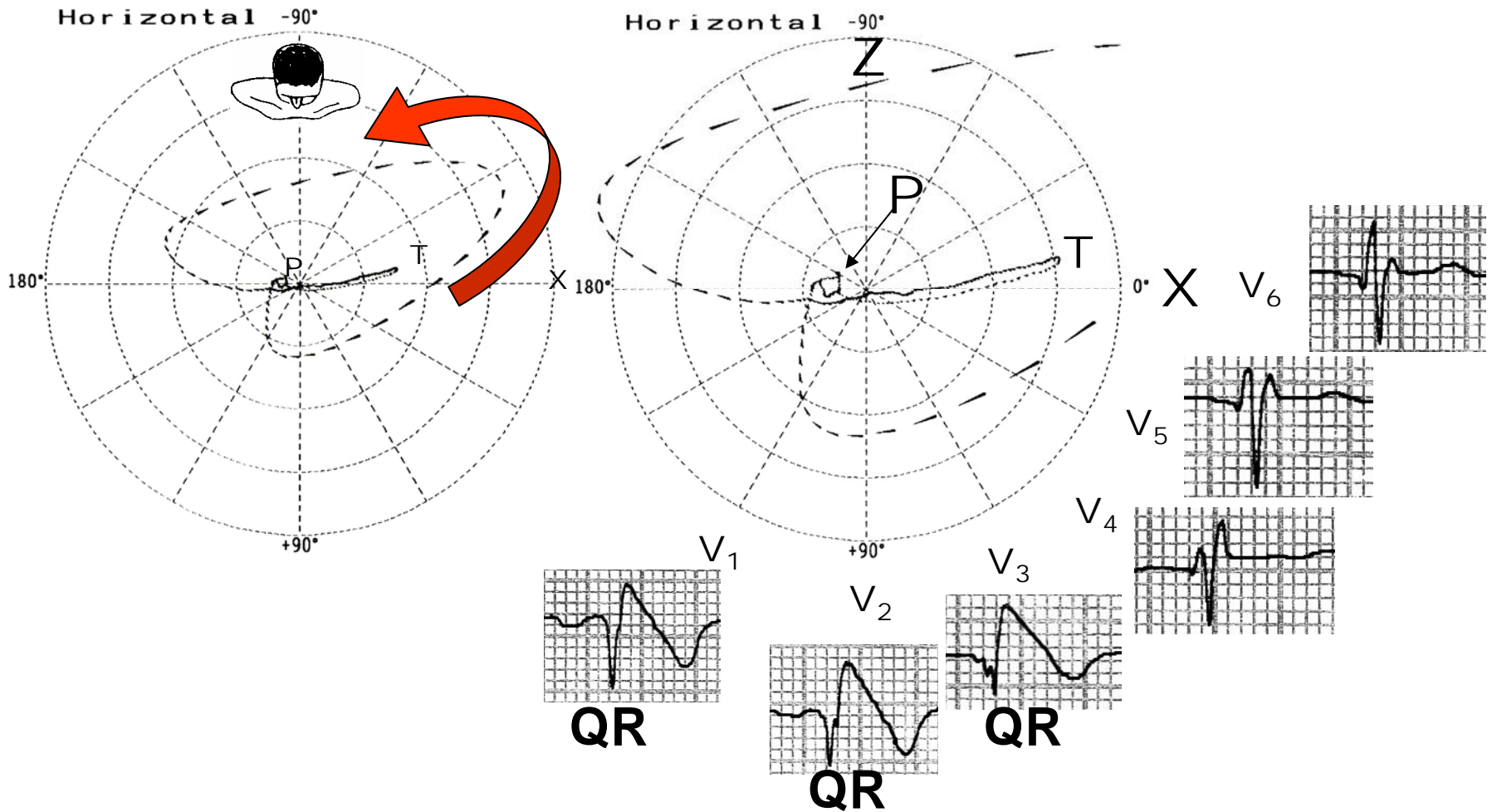
Entirely negative P wave on right precordial leads. It is frequently observed in pectus excavatum consequence to right displacement of heart and modification of spatial orientation of the mean atrial activation vector. The atrial vector is oriented backwards so producing a negative P wave in right precordial leads or only in V1 leads (1).

Complete Right Bundle Branch Block (CRBBB): QRSd ≥ 120ms and QR pattern from V₁ to V₃ and absence of increase R voltage waves on precordial leads was described in pectus excavatum, secondary to rotation of the heart.

Spontaneous Type 1 Brugada ECG pattern Prominent R wave in aVR: aVR sign. A prominent R wave in lead aVR (aVR sign) is an element of risk for development of arrhythmic events in BrS. In the presence of BrS, prominent R wave in lead aVR may reflect more right ventricular conduction delay and subsequently more electrical heterogeneity, which in turn is responsible for a higher risk of arrhythmia(2).

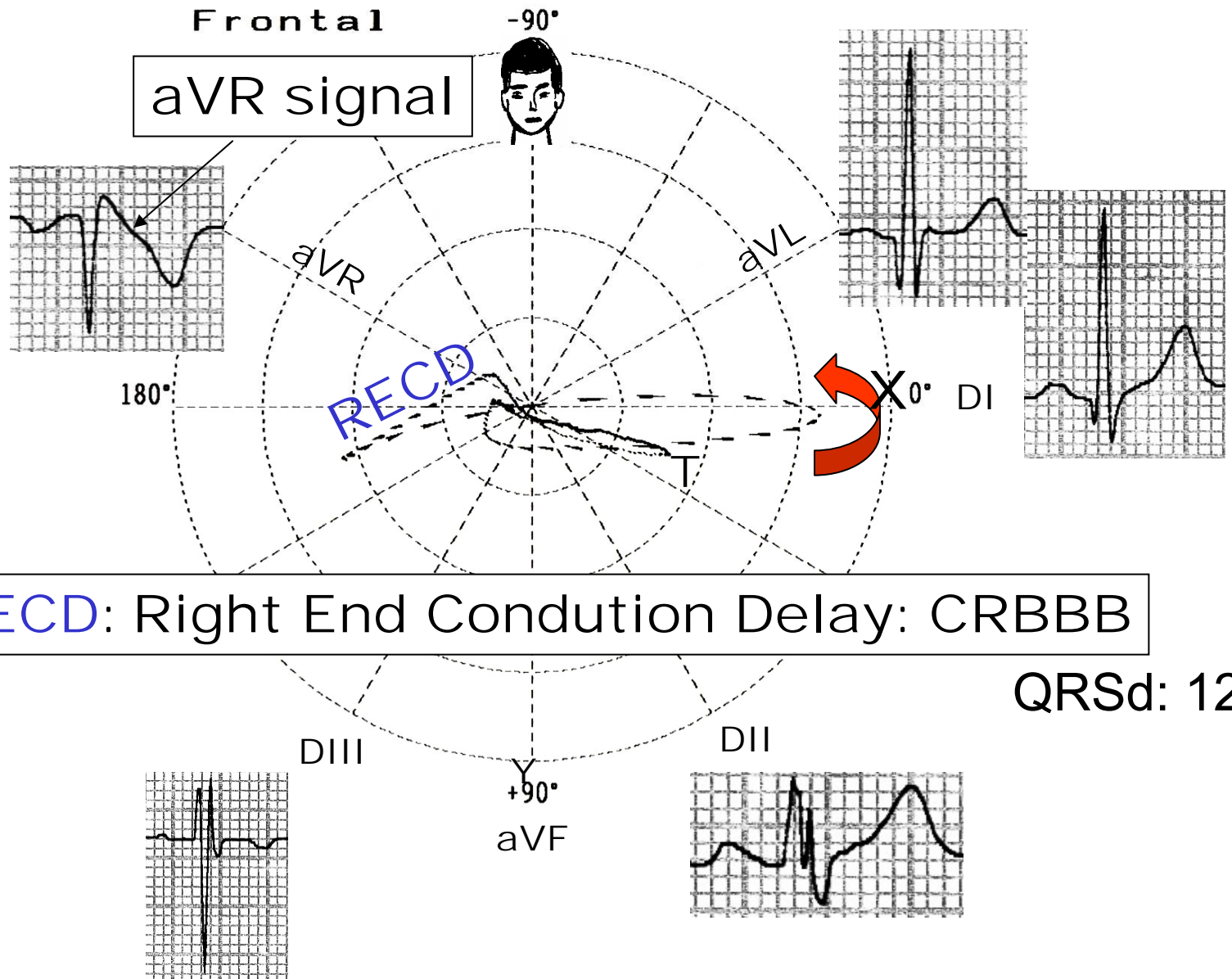
- 1) Martins de Oliveira J, Sambhi MP, Zimmerman HA. The electrocardiogram in pectus excavatum. Br Heart J 1958 Oct; 20: 495-501.
- 2) Babai Bigi MA, Aslani A, Shahrzad S. aVR sign as a risk factor for life-threatening arrhythmic events in patients with Brugada syndrome. Heart Rhythm 2007 Aug; 4: 1009-1112.

ECG/VCG CORRELATION HORIZONTAL PLANE



**Brugada-Like Electrocardiographic Pattern
or Brugada phenocopy**

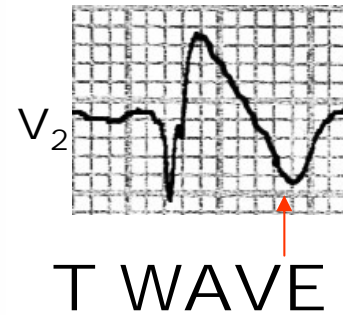
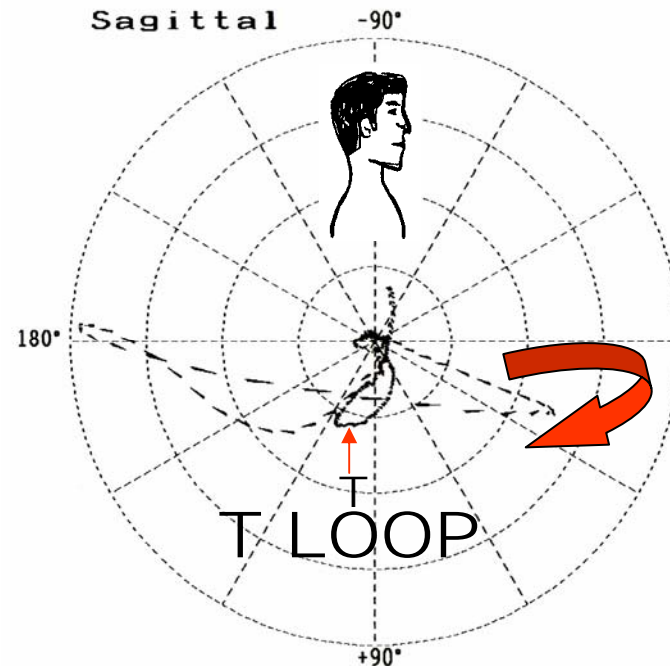
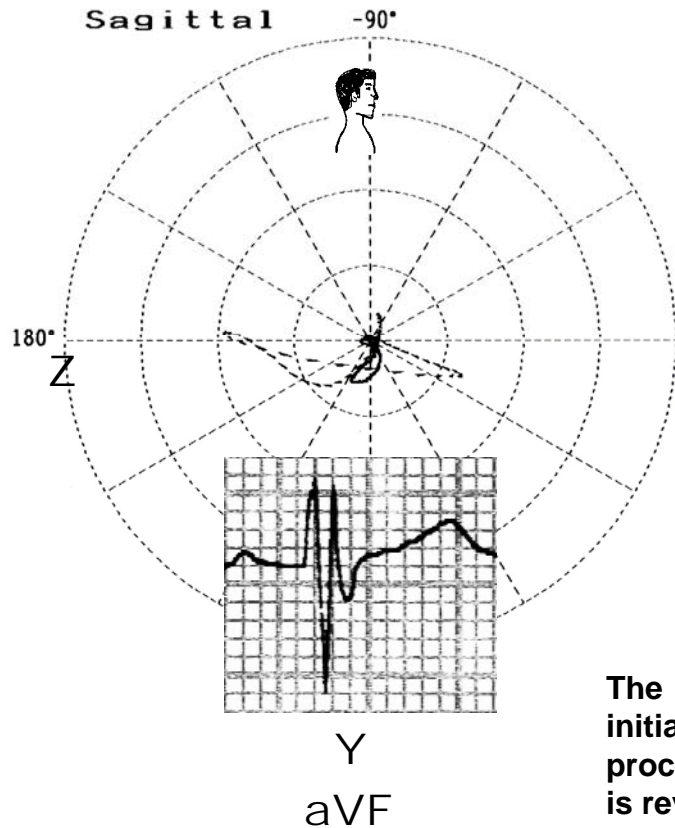
ECG/VCG CORRELATION FRONTAL PLANE



RECD: Right End Conduction Delay: CRBBB

QRSd: 122ms

ECG/VCG CORRELATION RIGHT SAGITTAL PLANE



The mechanical injury affecting the epicardium may produce a delay in the initiation of the repolarization in this region. Consequently, the recovery process starts in subendocardial portions and the orientation of the T vector is reversed.

Sensi. 2
 Timer 2 msec
 Loop All Loop
 Sagittal Right
 Z Axis Back

Sensi. 4
 Timer 2 msec
 Loop All Loop
 Sagittal Right
 Z Axis Back

MAIN ECG FEATURES IN PECTUS EXCAVATUM AND NORMAL HEARTS (1)

- I) **Negative P waves on right precordial leads:** consequence of modification of spatial orientation of the mean atrial activation vector. The atrial vector is oriented backwards so producing a negative P wave in right precordial leads or only in V1 lead.
- II) **S_I-S_{III} or S_I-Q_{III} pattern.**
- III) **rsr' pattern in V1:** in cases with minimal cardiac rotation, the presence of a final r' wave may be explained by the rightward and forward deviation of the mean depolarization vector of the basal ventricular portion. This pattern in V₁ does not mean, at least as a rule, a block in the right branch of the bundle of His itself.
- IV) **qr or QR pattern in right precordial leads:** The right atrial assumes the position directly below the exploring electrode of V₁ as consequence of a greater rotation of the heart. This lead now reflects the atrial intracavitary potentials and a qr or QR pattern appears.
- V) Exceptionally, Brugada type 1 pattern (2).

- 1) Martins de Oliveira J, Sambhi MP, Zimmerman HA. The electrocardiogram in pectus excavatum. Br Heart J 1958 Oct; 20: 495-501.
- 2) Kataoka H Electrocardiographic Patterns of the Brugada Syndrome in 2 Young Patients With Pectus Excavatum. J Electrocardiol 2002; 35: 169-171.

CAUSES OF QR PATTERN IN RIGHT PRECORDIAL LEADS

- I) Severe systolic right ventricular hypertrophy (extreme strain pattern) suprasystemic right intraventricular pressure: i.e. severe pulmonary stenosis
- II) Significant Right atrium dilatation i.e. Ebstein's anomaly with tricuspid insufficiency
- III) Right Bundle Branch Block associated with anterior or anteroseptal myocardial infarction
- IV) Right Bundle Branch Block with isoelectric initial r wave in V1
- V) Situs inversus: ventricular inversion: inverted septal activation.
- VI) Pectus excavatum.