Jovem masculino 19 anos portador de pectus excavatum – 2009

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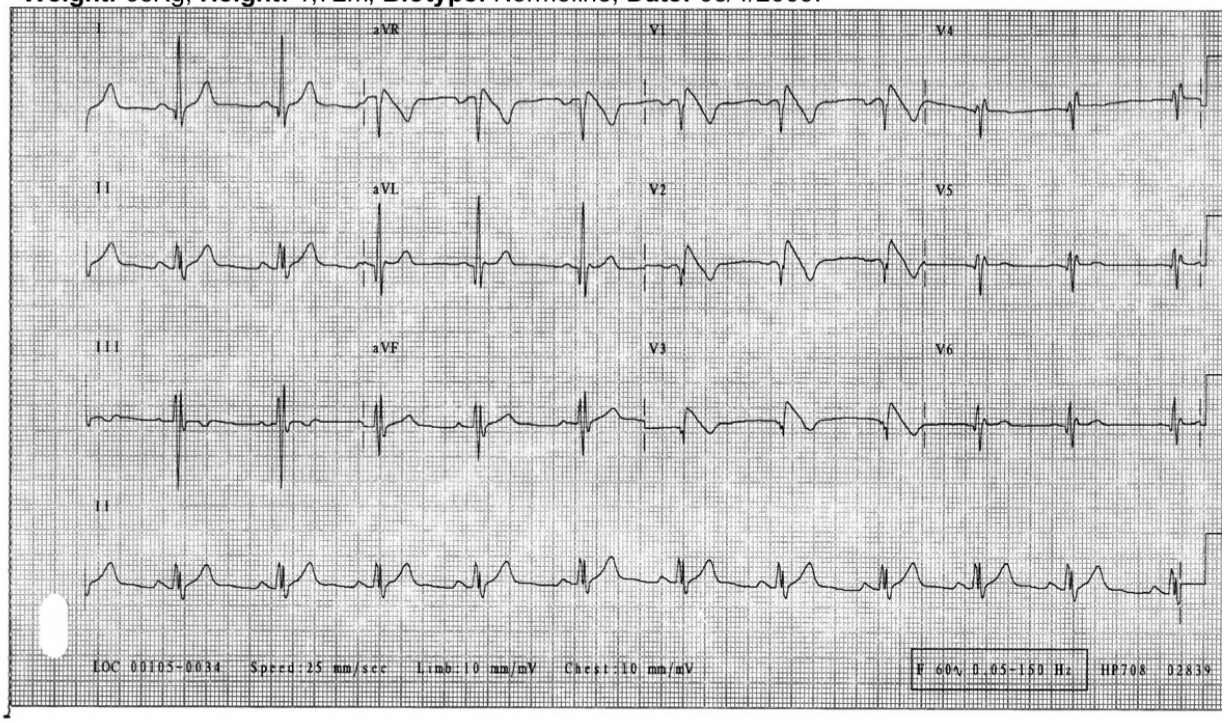
Prezados amigos lhes apresento este caso muito interessante
Jovem masculino 19 anos, assintomático veio à consulta para avaliação previa a prática esportiva. Antecedentes pessoais e familiares negativos para síncope ou morte súbita em familiares de primeiro grau menores de 45 anos.
O exame visual ou ectoscópico do tórax revelava pectus excavatum muito significativo com o terço inferior do esterno, mas comprometido que o 1/3 superior que se mostrava quase normal. Refere que a mencionada deformidade foi notada desde o nascimento com progressiva piora ao longo da vida. Nenhum familiar de primeiro grau era portador de pectus excavatum , síndrome de Marfand ou de Poland.
No exame físico sopro sistólico suave ejetivo ++ se ausculta click na válvula mitral.
Claro pulmonar diminuído em ambas as bases
O ECG (vejam em adjunto)
O Ecocardiograma resultou normal.
O RX de tórax PA mostrava um pseudo aumento da área cardíaca e a projeção lateral significativa diminuição do diâmetro antero-posterior do tórax.
A prova funcional respiratória revelou leve distúrbio restritivo. Os volumes pulmonares reduzidos e redução da capacidade pulmonar total assinalando alteração funcional restritiva leve.
Algum comentário/s?

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
Electocardiographic diagnosis:

Abraço
Andrés Ricardo Pérez Riera

OPINIONES DE COLEGAS

Hola a todos, la verdadpareciera un ECG de Brugada, sin embargo con esa deformidad del torax no se si pudiera causar algùn pseudo patron de Brugada, sì ud. lo està preguntando algo debe haber atràs ya que ud. mismoha descrito ECG de ese tipo como Brugada,en el peor de los casos es asintomatico y sin antecedentes de muerte subita familiar, porlo cuàl pudieramos observar clinicamente.

CarlosRodríguez Artuza

El ECG Tiene todo el aspecto de S. de Brugada

Emilio Marigliano

Hola,me parece que tiene patente de S de Brugada ,con un eje izquierdo tipo HBAI,,pero me llama la atencion el QRS en cara inferior como si hubiera un trastorno de conduccion intraventricular intraventicular,no se especificarlo.....perdon...espero respuestas de los expertos
Gracias

Dra Ortega

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

ANOTHER BRUGADA ECG PHENOCOPY

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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 examiantion: 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.).

ELECTROCARDIOGRAPHIC DIAGNOSIS

Rhythm: Normal sinus;
Heart rate: 67bpm;
P wave: P axis + 28^0 on frontal plane, entirely negative in leads V1-V2 and perpendicular to V3;
PR interval duration: 177ms;
QRS: QRSd: 122ms, **QRS axis:** + 60^0 on frontal plane. **QRS complex:** QR pattern from V_1 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 + 28^0 on frontal plane and with negative T polarity from V1 to V3;
QT/QTc: intervals: 375/390ms.
Conclusions:

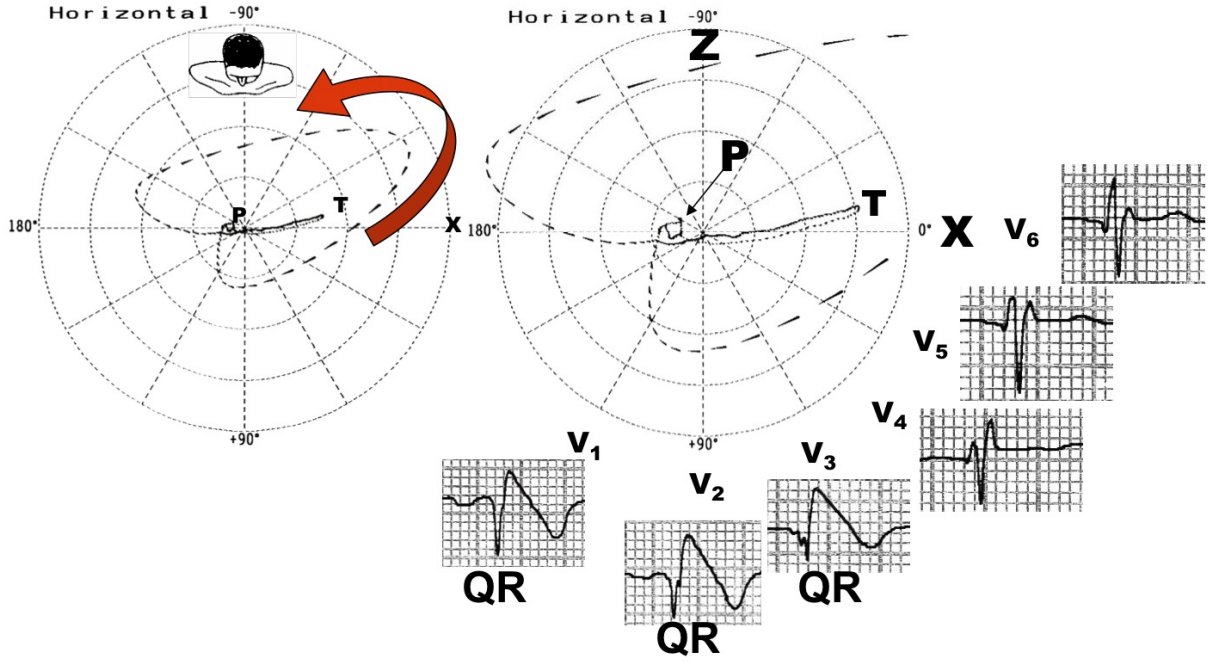
Entirely negative P wave on right precordial leads. It is frequently observed in pectus excavatum consequence to right displacement of heart and modification of spatial orientation of the mean atrial activation vector. The atrial vector is oriented backwards so producing a negative P wave in right precordial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

Complete Right Bundle Branch Block (CRBBB): QRSd ≥120ms and QR pattern from V_1 to V_3 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).

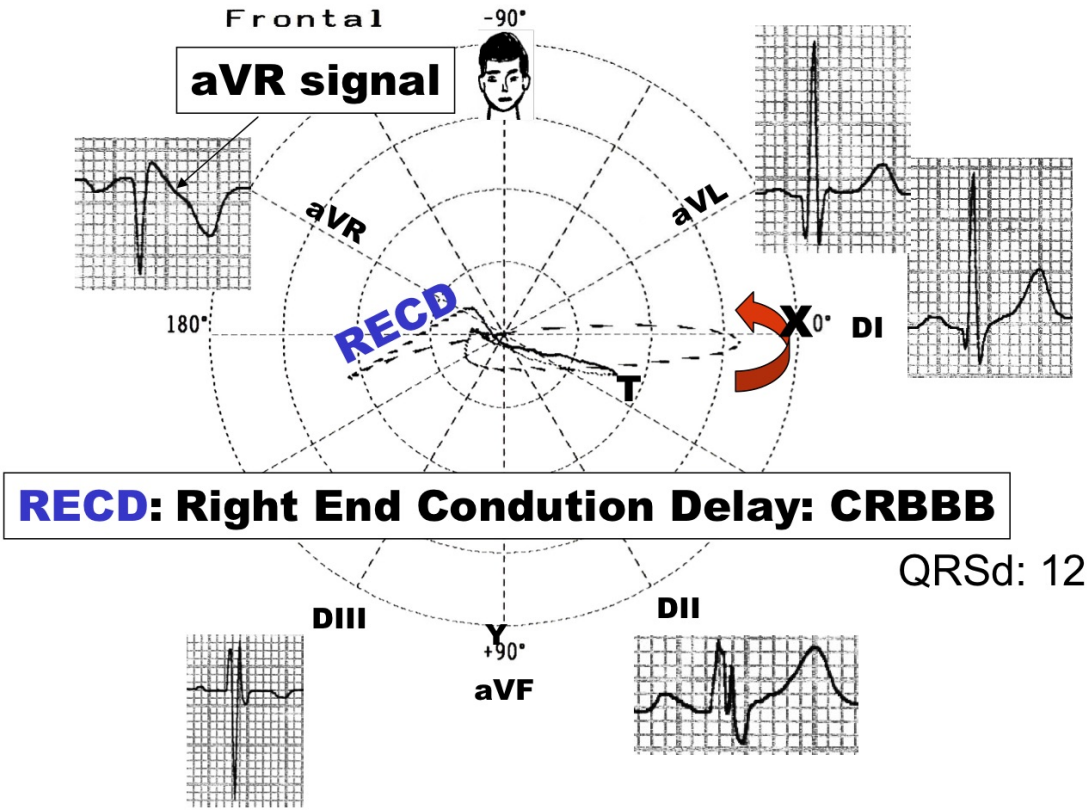
1) **Martins de Oliveira J, Sambhi MP, Zimmerman HA. The electrocardiogram in pectus excavatum. Br Heart J 1958 Oct; 20: 495-501.**
2) **Babai Bigi MA, Aslani A, Shahrzad S. aVR sign as a risk factor for life-threatening arrhythmic events in patients with Brugada syndrome.Heart Rhythm 2007 Aug; 4: 1009-1112.**

ECG/VCG CORRELATION HORIZONTAL PLANE



Brugada-Like Electrocardiographic Pattern
or Brugada phenocopy

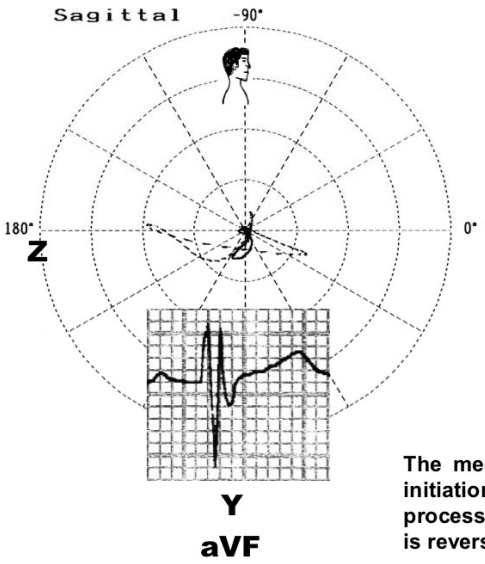
ECG/VCG CORRELATION FRONTAL PLANE

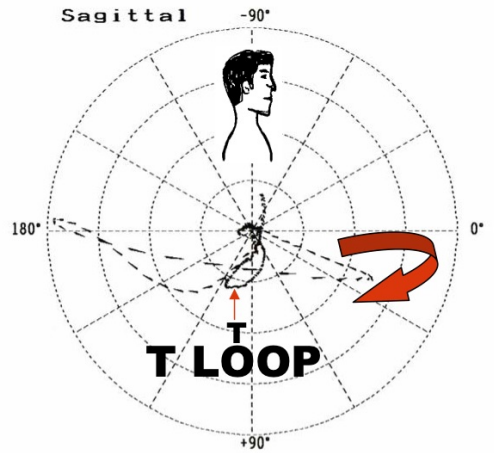


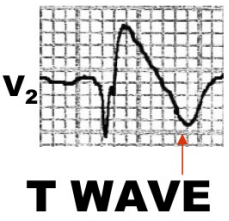
RECD: Right End Condution Delay: CRBBB

QRSd: 122ms

ECG/VCG CORRELATION RIGHT SAGITTAL PLANE







The mecanical injury affecting the epicardium may produce a delay in the initiation of te 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) $\mathbf{S_I\text{-}S_{III}}$ or $\mathbf{S_I\text{-}Q_{III}}$ 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 V_1 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 V_1 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 J1958 Oct; 20: 495-501.**

2) **Kataoka H Electrocardiographic Patterns of the Brugda 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 pumonary stenosis
II) Significative Right atrium dilatation i.e. Ebstein's anomaly with tricuspid insufficiency
III) Right Bundle Branch Block associated wit 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.