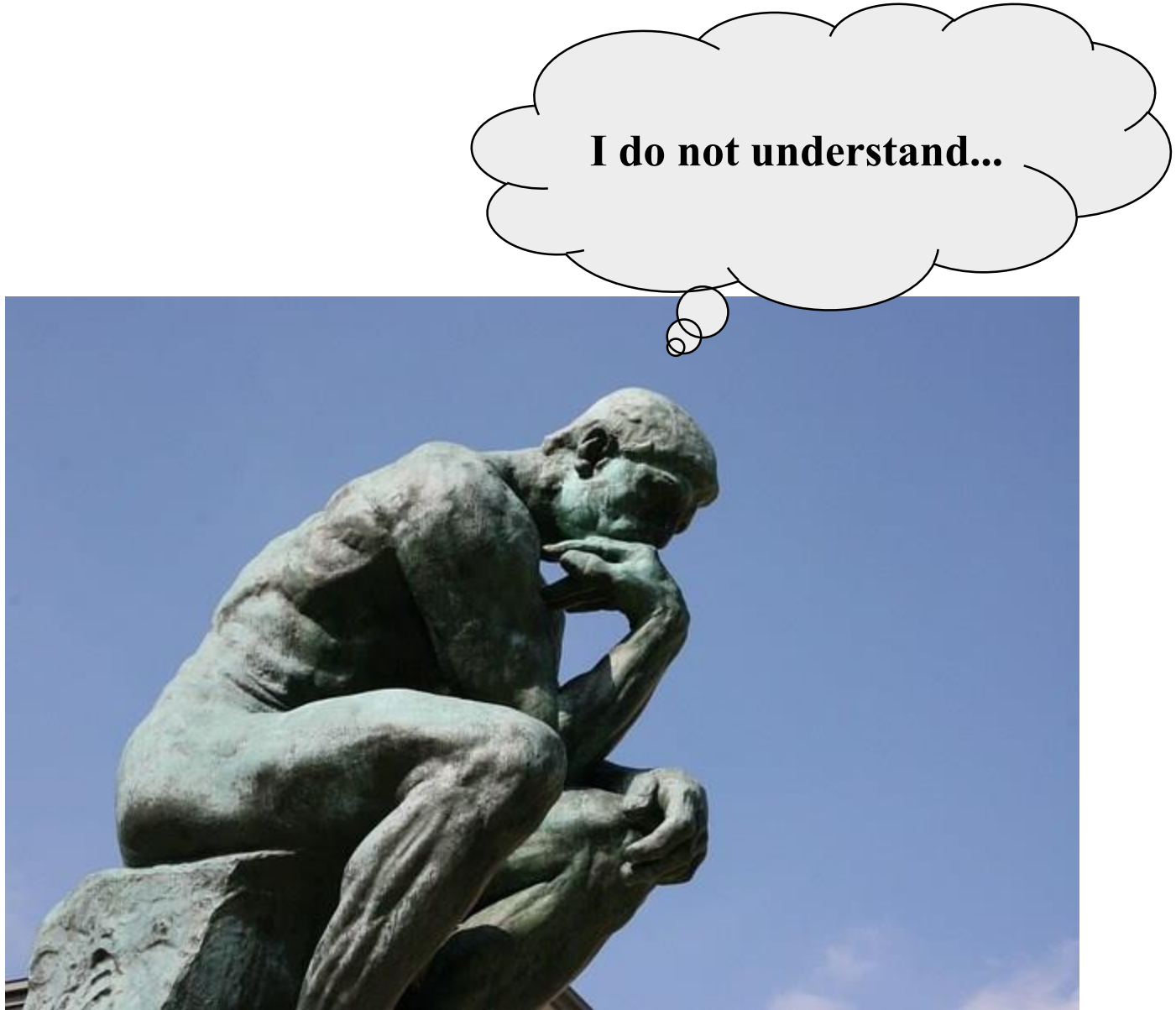
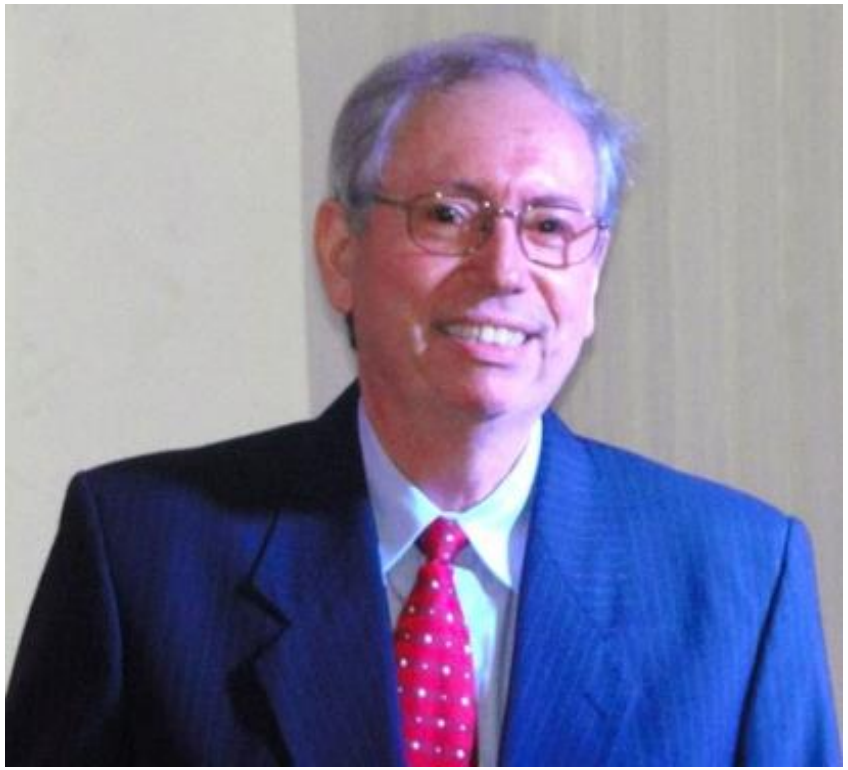
No more than the P-loop to make the diagnosis: which is the diagnosis?

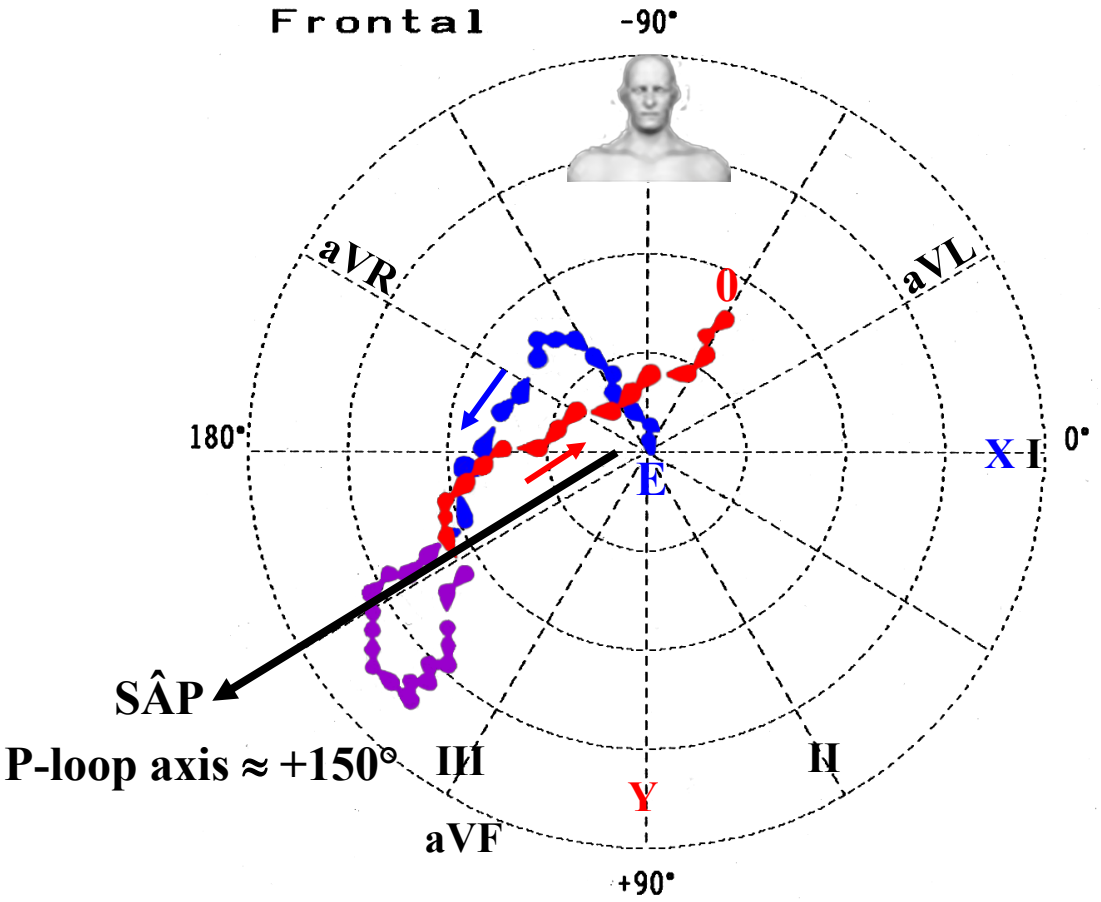


Andrés Ricardo **Pérez-Riera, M.D. Ph.D.**
Design of Studies and Scientific Writing Laboratory in the ABC School of Medicine, Santo André, São Paulo, Brazil
https://ekgvcg.wordpress.com

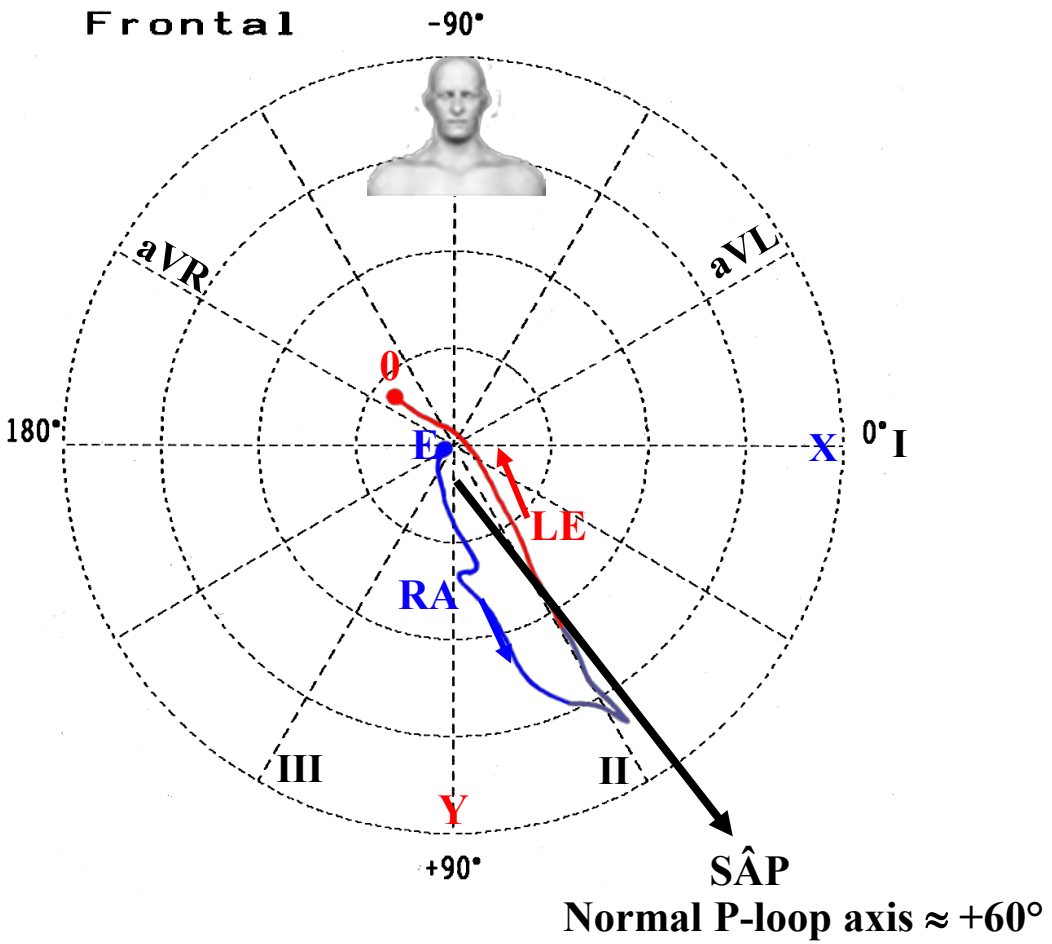


Raimundo **Barbosa-Barros**, MD
Chief of the Coronary Center of the Hospital de Messejana Dr. Carlos Alberto Studart Gomes. Fortaleza – CE- Brazil

Isolated P-loop in the frontal plane (magnified 32x)



Isolated normal P-loop in the frontal plane (magnified 32x)

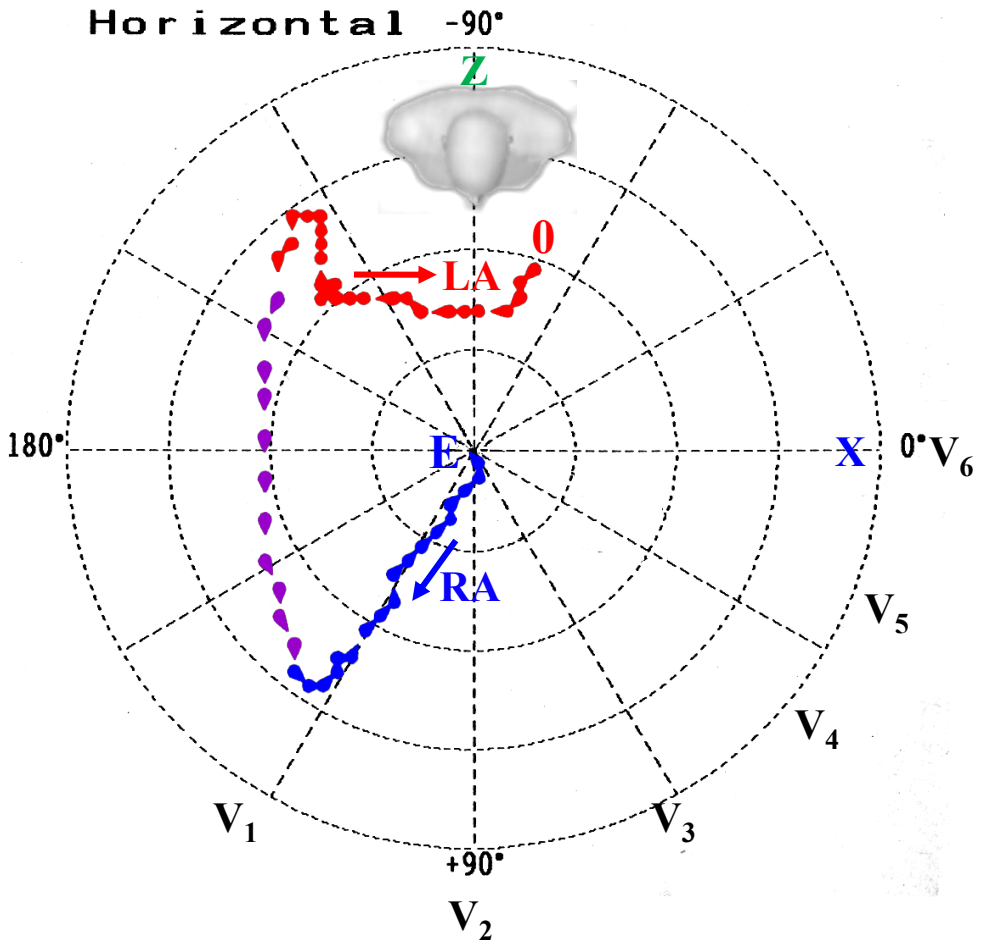


Right atrium depolarization

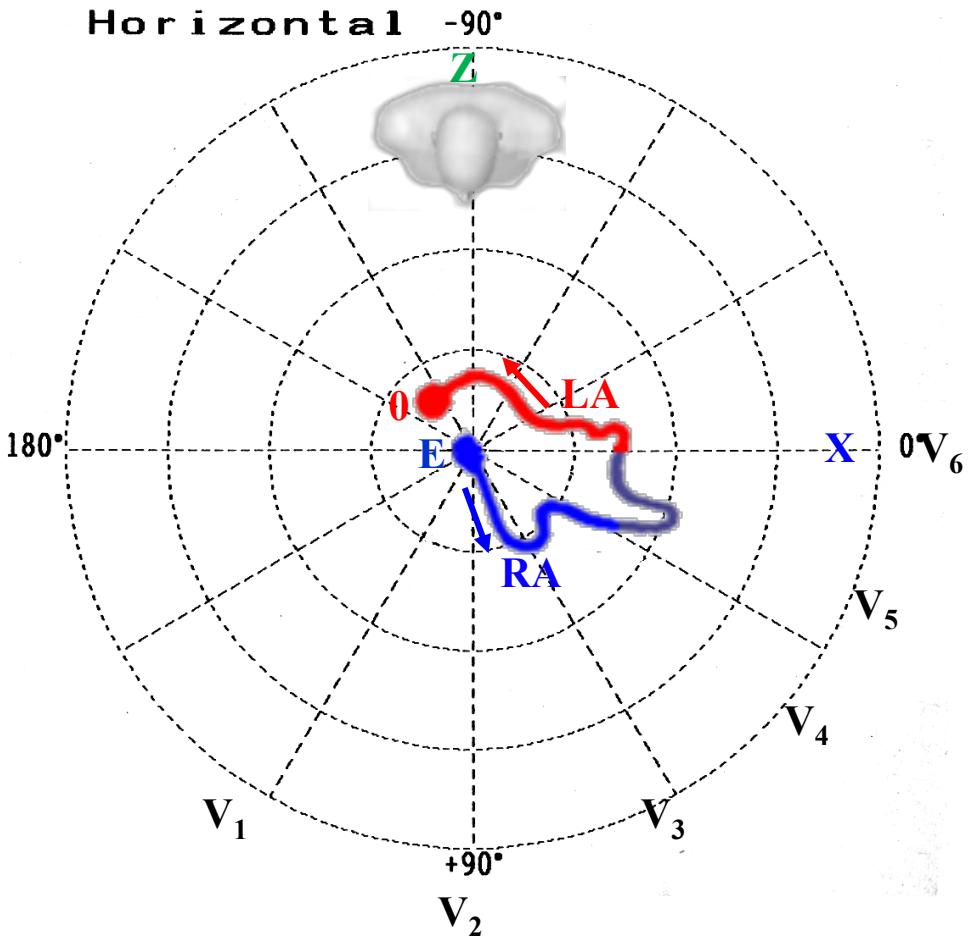
Biatrial depolarization (interatrial septum)

Left atrium depolarization

Isolated P-loop in the horizontal plane (magnified 32x)



Isolated normal P-loop in the horizontal plane (magnified 32x)

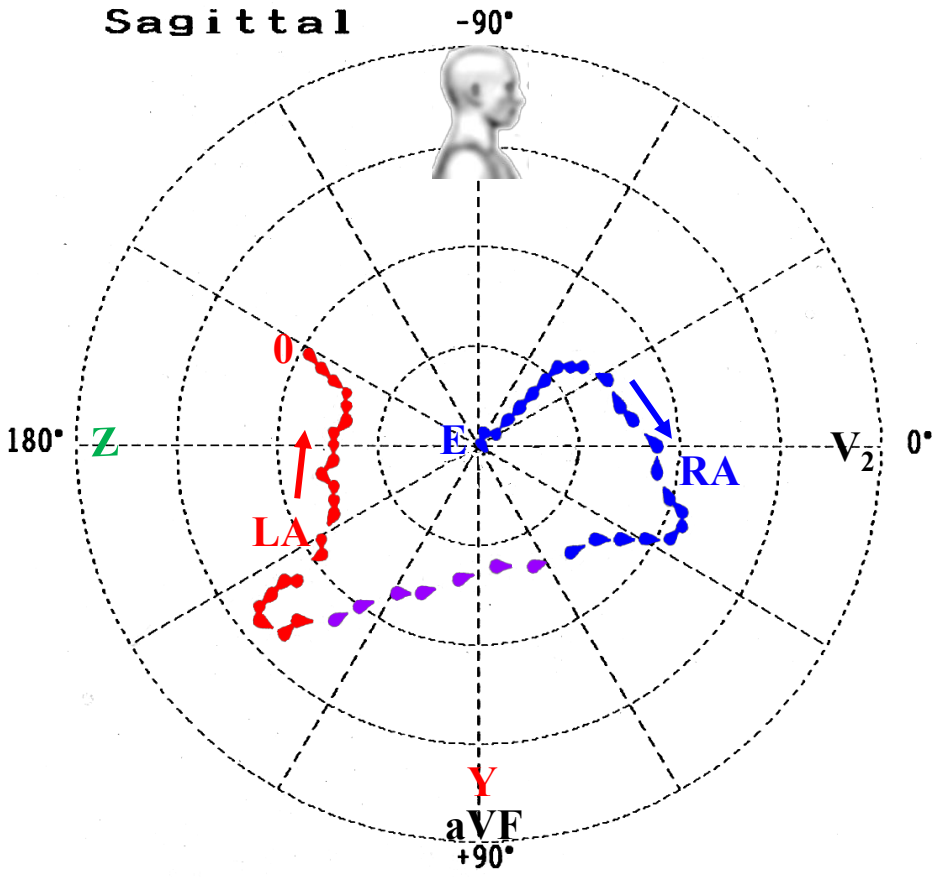


Right atrium depolarization

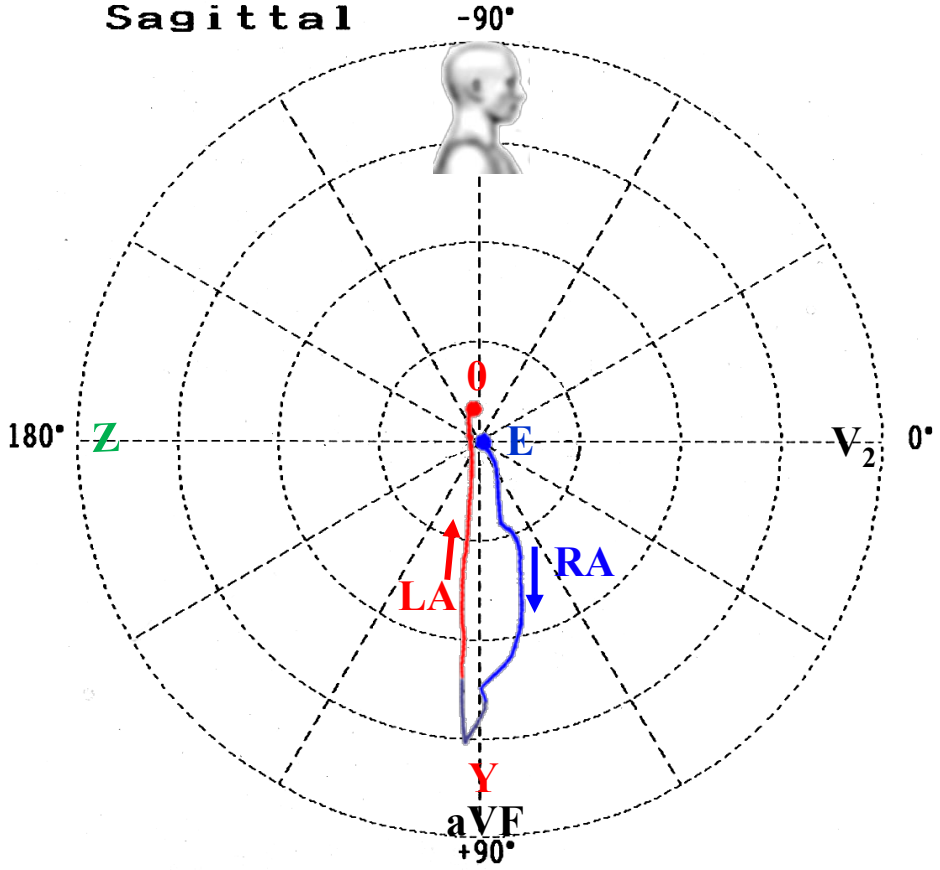
Biatrial depolarization (interatrial septum)

Left atrium depolarization

Isolated P-loop in the right sagittal plane (magnified 32x)



Isolated normal P-loop in the right sagittal plane (magnified 32x)



Right atrium depolarization

Biatrial depolarization (interatrial septum)

Left atrium depolarization

Colleagues opinions

Hola! El bucle especialmente en plano sagital derecho muestra aumento de voltaje de componente anterior y posterior del mismo sugestivo de sobrecarga bi-auricular.
ESTENOSIS mitral severa?

Juan José Sirena, MD
Santiago del Estero, Argentina

Dear Raymundo and Andres,

The end of the P loop is spatially displaced compared to its onset. This would correspond to a shift in the PTa segment of the ECG and indicates atrial injury/infarct. I also see interatrial conduction delay, probably secondary to ischemia.

Mario D. González, MD

Dextrocardia
Afectuosamente
Isabel Victora **Konopka MD**
Hospital Argerich Buenos Aires
Argentina

Dextrocardia
Affectionately
Isabel Victora **Konopka MD**

Comments: Correct diagnosis: how unfair it is god!
Beauty and intelligence together.
Congratulations Isabel!
Potro & Raymond

In patients with Pulmonary Hypertension, ECG can show increased P wave amplitude ($\geq$ 2.5 mm in the DII derivation).

Best wishes

hermesmelo

I am trying to get the patient to come to the clinic to do what you had suggested

Yochai **Birnbaum, MD, FACC, FAHA**
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John S. Dunn Chair in Cardiology Research and Education
The Department of Medicine, Section of Cardiology
Baylor College of Medicine
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MS BCM620
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Email: ybirnbau@bcm.edu

Final conclusions by

Andrés Ricardo **Pérez-Riera, M.D. Ph.D.**
(a long time ago…)
Design of Studies and Scientific Writing
Laboratory in the ABC School of Medicine,
Santo André, São Paulo, Brazil
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Raimundo **Barbosa-Barros**, MD
Chief of the Coronary Center of the Hospital de Messejana Dr. Carlos Alberto Studart Gomes. Fortaleza – CE- Brazil

Case presentation

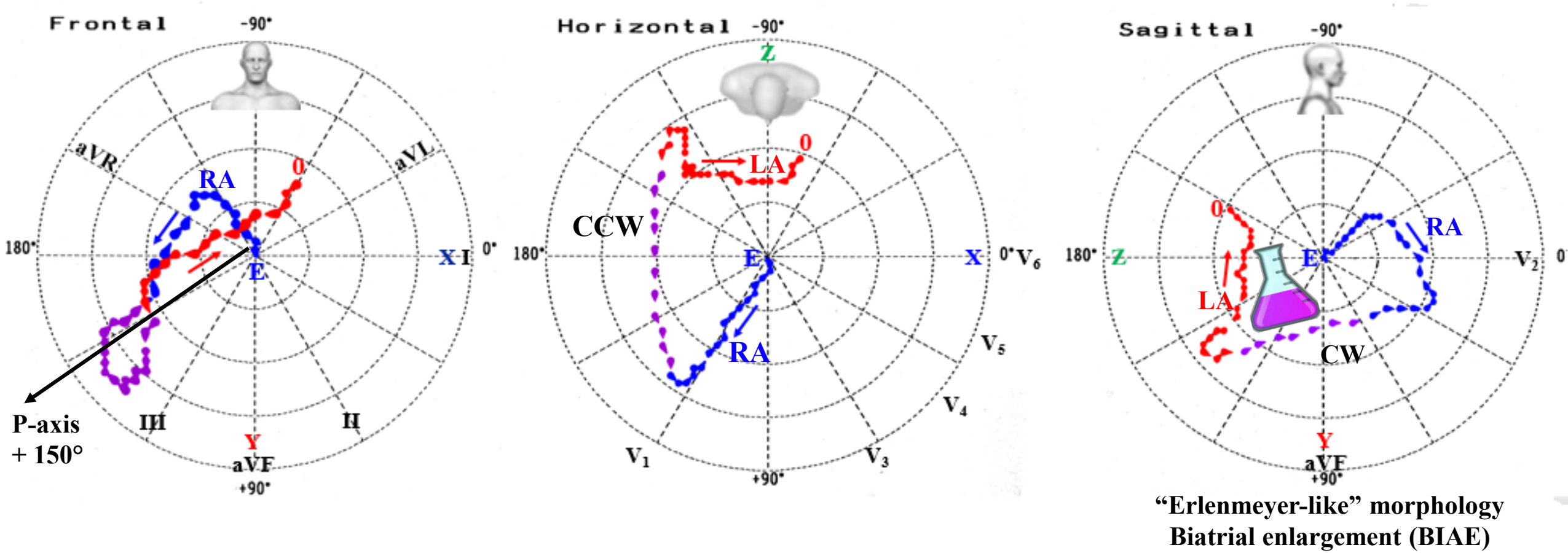
Female, Caucasian female patient, 58 years old. She has been known for a long time to be a simplex type of dextrocardia (normal heart with mirror image without other associated heart diseases). Situs inversus: the organs of the abdomen and lungs located in mirror image.

At the age of 10, a rheumatic fever outbreak occurred after Lancefield's beta-hemolytic group A streptococcus aureus, leaving a severe sequel to the mitral valve (double mitral lesion of stenosis type), which forced the patient to undergo a valve replacement after 25 years (a bioprosthesis was implanted) Currently asymptomatic. No medication.

He had Blood pressure 130 mmHg x 80 mmHg, heart rate (HR) of 60 bpm, ictus or the maximum cardiac impulse at the intersection of the 4th and 5th right intercostal space in the right hemi clavicular line(on the right chest), regular heart rhythm and mild hypophonesis of the sounds. Heart sounds were heard on the right precordium and attenuated on the left.

We preform ECG (ECG-1), VCG, a second ECG-tracing with right precordial leads (ECG-2) and Chest x-ray in the poster anterior and lateral view.

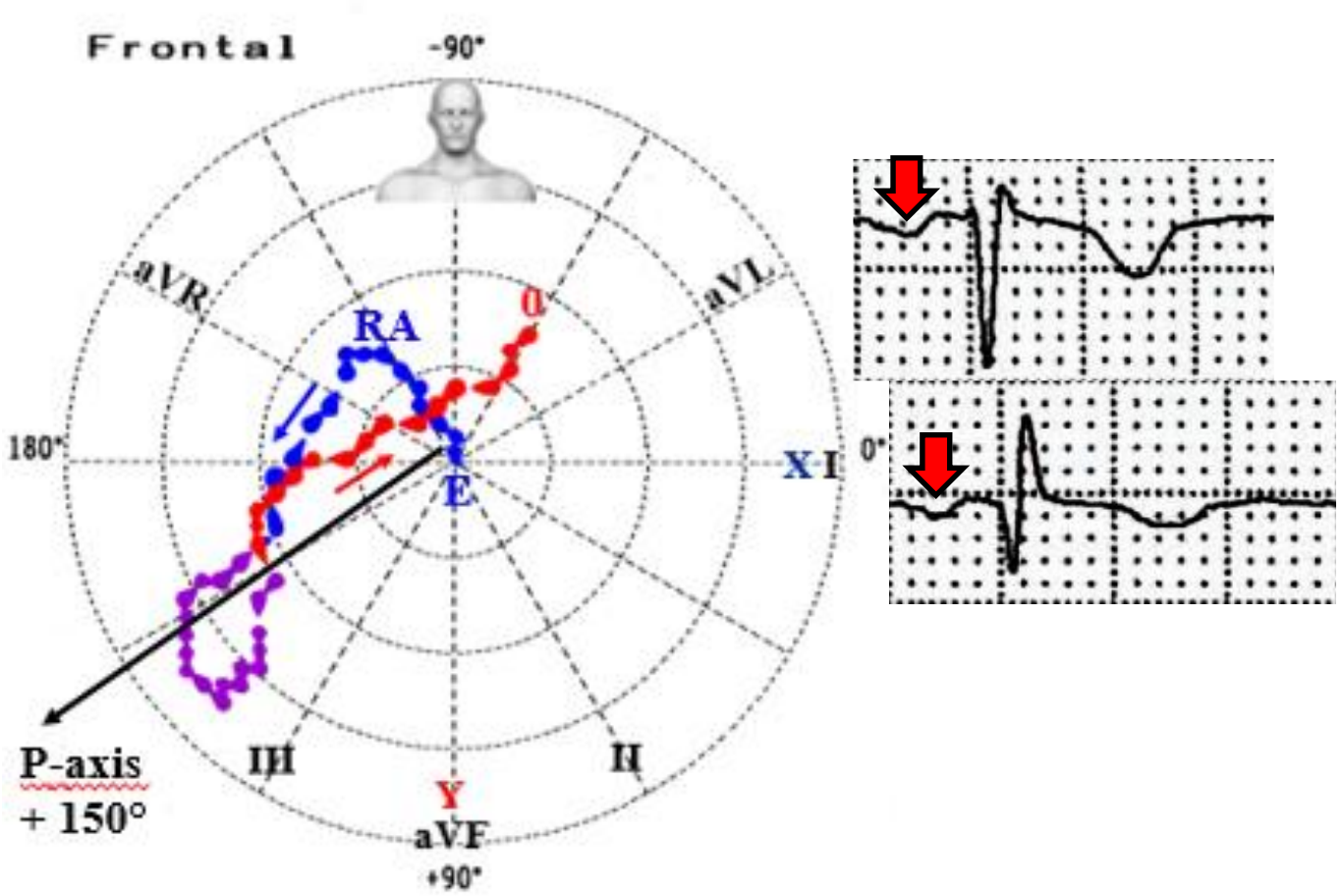
Isolated P-loop in the three planes (magnified 32x)



"Erlenmeyer-like" morphology
Biatrial enlargement (BIAE)

I. **Frontal Plane:** P-Loop figure in eight, P-loop axis on right inferior quadrant (near + 150°)
II. **Horizontal Plane:** P- Loop located on right quadrants, : P- Loop rotation counterclockwise (CCW), with augmented anterior (> 0.06 mV) and posterior forces (> 0.04 mV.):. biatrial enlargement
III. **Right Sagittal Plane:** P- Loop rotation clock wise (CW). Anterior (**RA**) and posterior (**LA**) increased Maximal anterior (up to 0.06 mV) and posterior forces(up to 0.04 mV: biatrial enlargement;

Conclusions: atrial inversions + biatrial enlargement.



Possible causes of negative P wave in lead I

1) **Incorrect limbs electrode cable connection during electrocardiographic recording (right to left):** The frequency of ECG artifacts due to switched electrodes is 0.4% at the outpatient clinic and 4.0% at the intensive care unit (**Rudiger 2007**). Limb electrode misconnection: Pseudo dextrocardia by exchange of limb electrodes. All P, QRS, and T waves are negative in I, but normal progression of QRS in precordial leads rules out this hypothesis, pointing out the exchange of arm electrodes.

2) **Simple true dextrocardia:** mirror image. Total atrio-visceral situs inversus with no heart disease. SÂP directed to the right and below, pointing at around +120° (III). Negative P wave in VL and I, positive in III. Reverse progression of r wave in precordial leads V2 to V5 (decreasing).

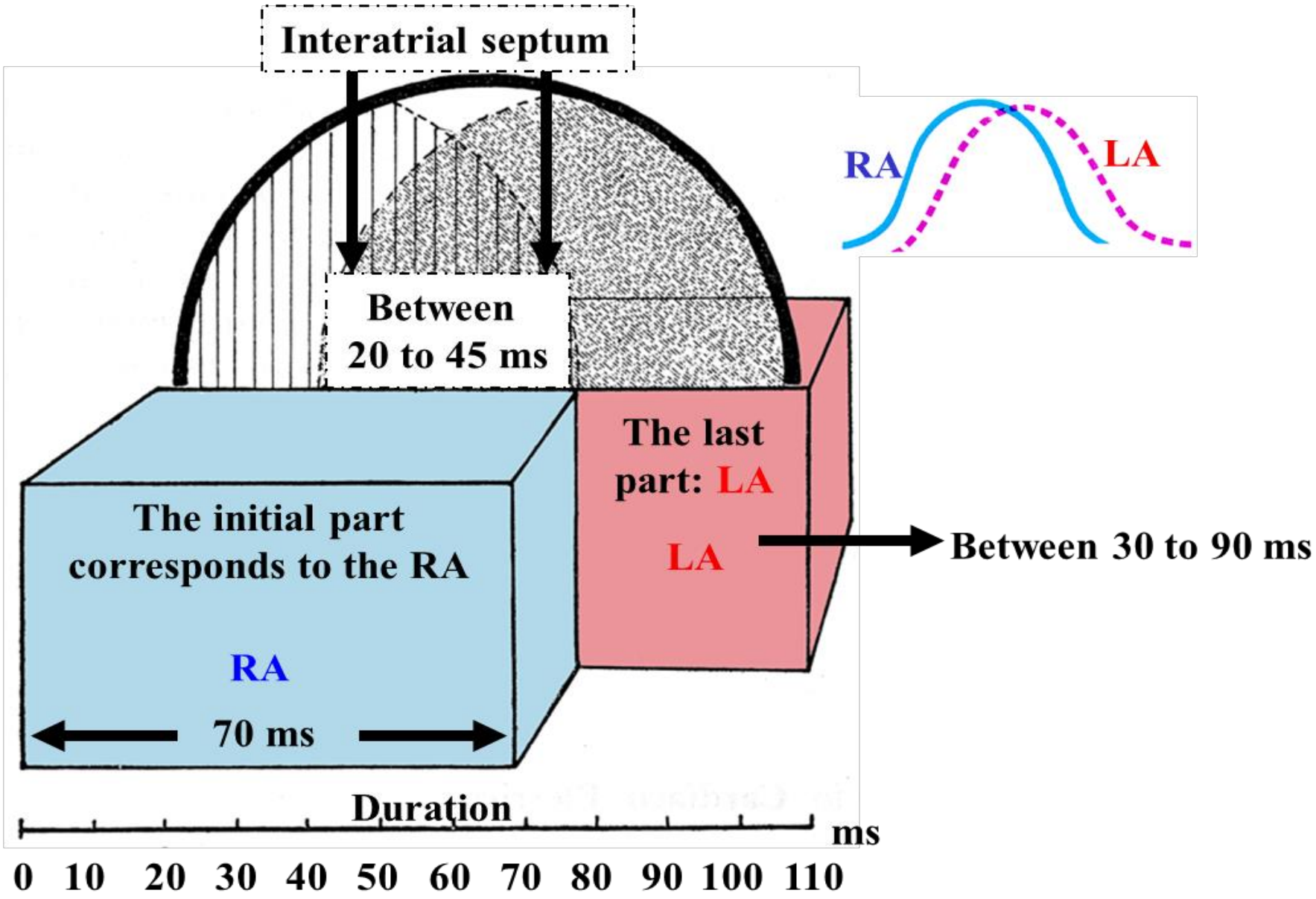
Summary of normal characteristics of the P loop in the three planes

The P loop has a short slow conduction in the onset, in at least 2 planes.

Two small notches are usually observed: one in the efferent limb and another in the afferent limb.

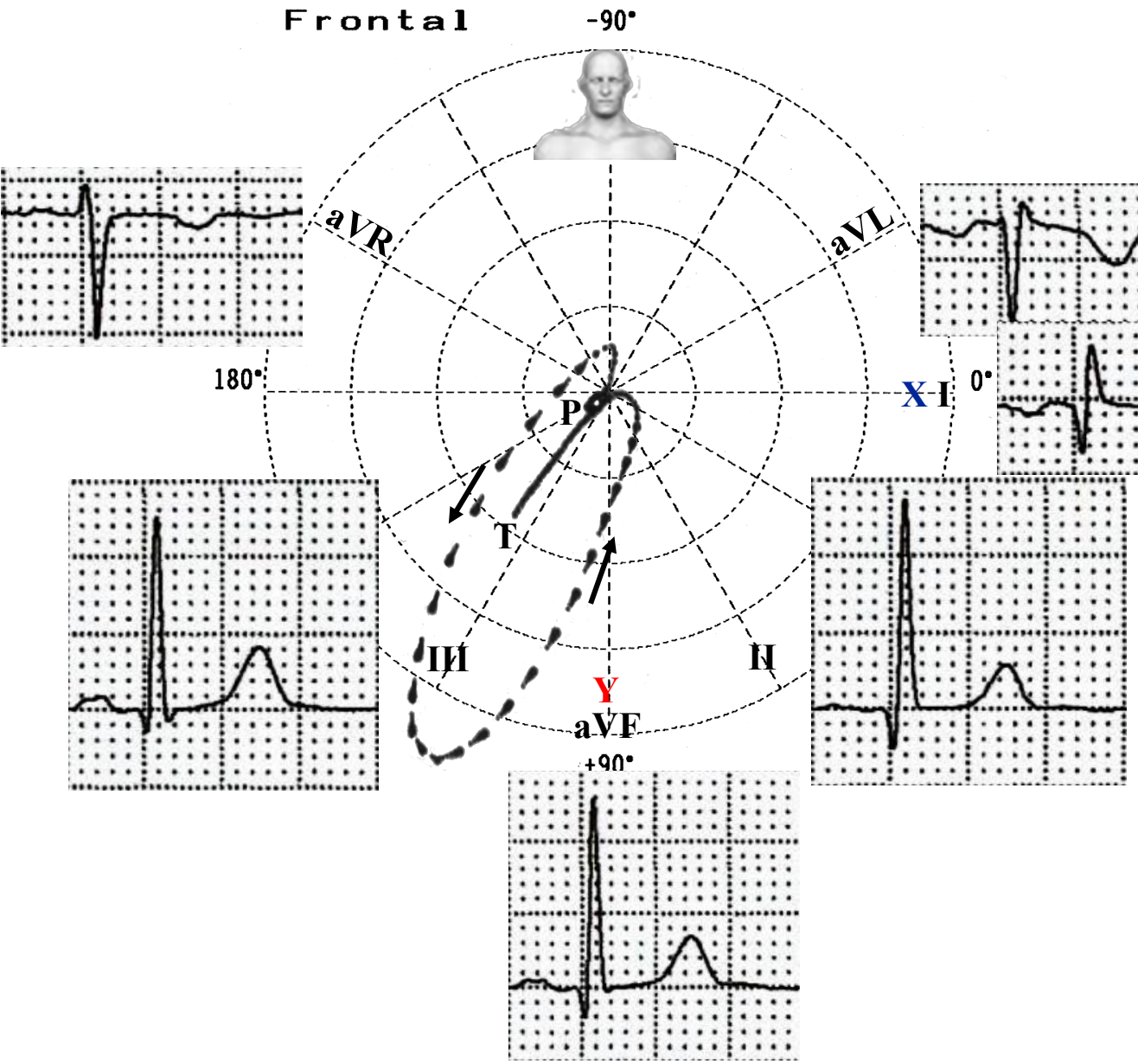
	FP	HP	RSP
Rotation	CCW	CCW or in eight	CW
Direction	Inferior and left	Anterior initial and posterior final part	Antero-inferior initial and posterior final part
Morphology	Oval	Oval	Spear point or triangular
Location	Left inferior quadrant	$^1/_3$ in anterior quadrant and $^2/_3$ in posterior quadrant	$^1/_3$ anterior and inferior and $^2/_3$ posterior and inferior
Location of maximal vector	+65° (+20° to +90°)	+50° to –45°	+55° to –20°
Voltage of maximal vector	≤ 0.2 mV	≤ 0.1 mV	≤ 0.18 mV
Maximal anterior forces		Adults up to 0.06 mV Children up to 0.08 mV	Adults up to 0.06 mV Children up to 0.08 mV
Maximal posterior forces		Up to 0.04 mV	Up to 0.04 mV
Maximal left forces	Adults up to 0.09 mV Children up to 0.13 mV	Adults up to 0.09 mV Children up to 0.13 mV	

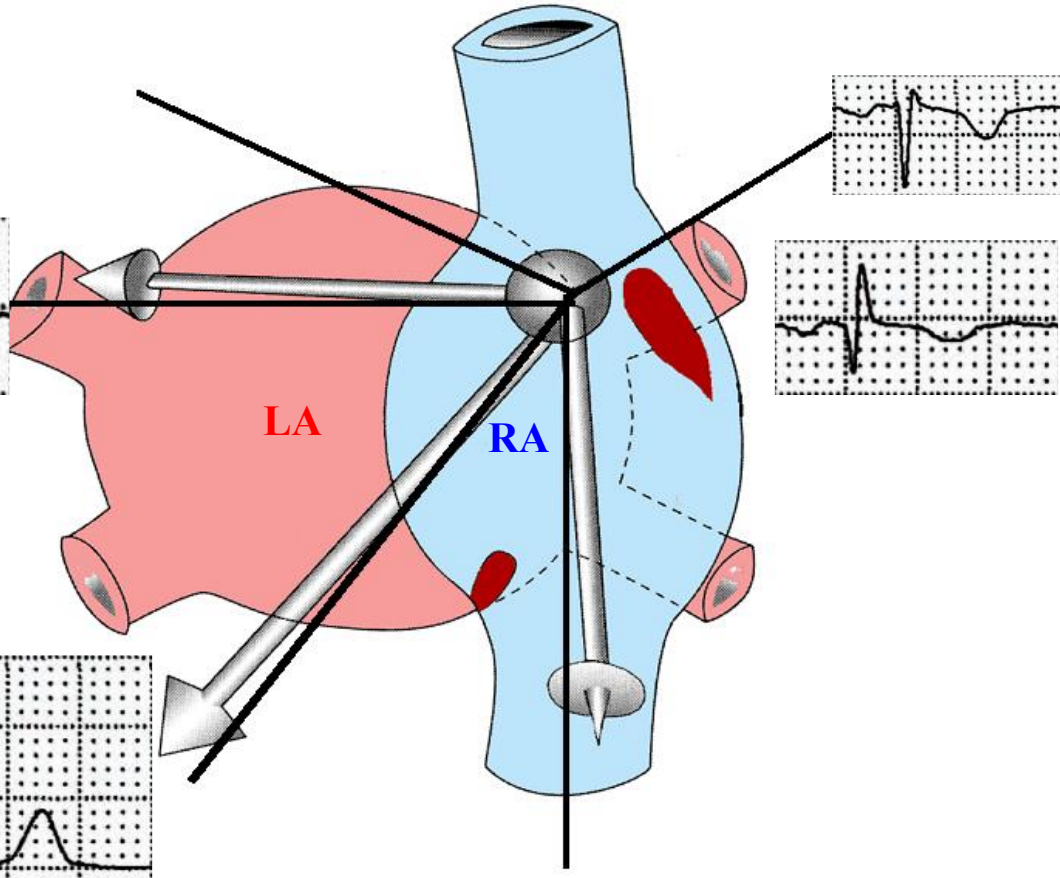
Normal activation of biatrial chamber



Normal duration of the P wave = 80 to 110 ms

ECG/VCG correlation in the frontal plane

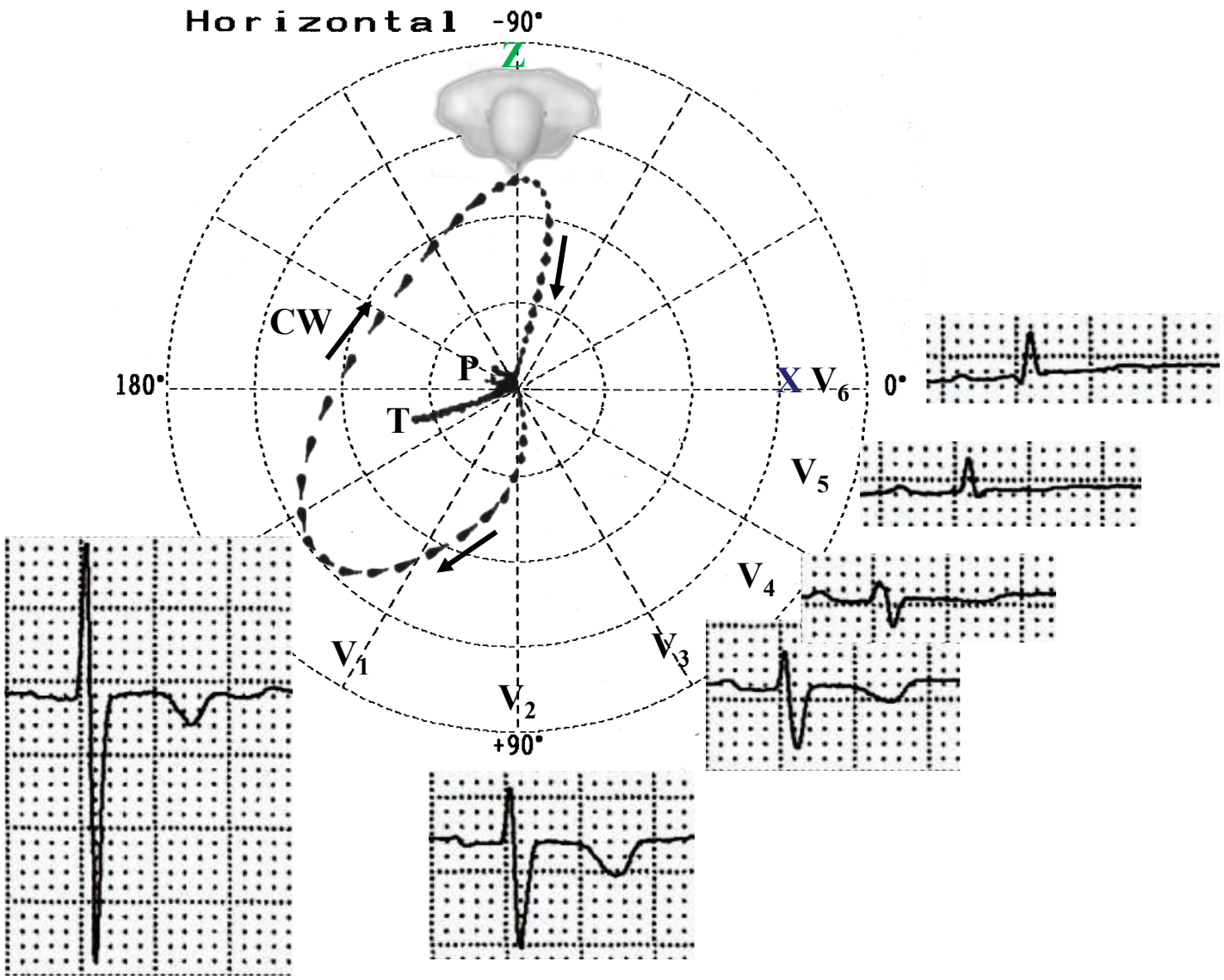




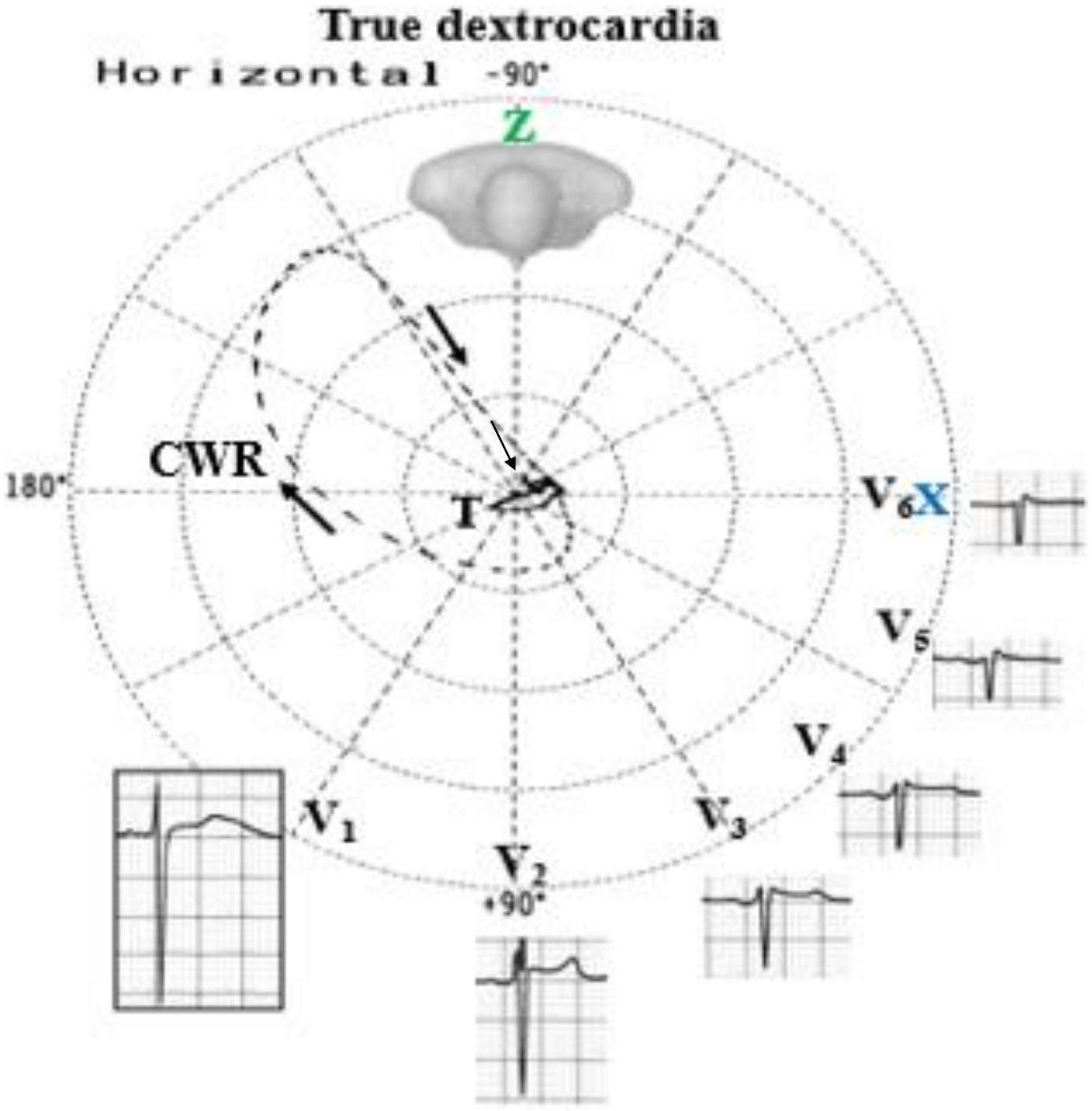
P-axis, QRS-axis and T-axis are near III lead.
P wave in I and aVL have negative polarity

Situs describes the position of the cardiac atria and viscera. Situs solitus is the normal position, and situs inversus is the mirror image of situs solitus. Cardiac situs is determined by the atrial location. In situs inversus, the morphologic right atrium (RA) is on the left, and the morphologic left atrium (LA) is on the right.

ECG/VCG correlation in the horizontal plane



Precordial R voltage with inverted progression (attenuated on left precordial leads) (**Juncos 2014**). P-loop, QRS loop, and T-loop are located in the right quadrants and QRS-loop with clockwise (CW) rotation.



In true dextrocardia, we observed that in the horizontal plane (HP), the QRS loop is located predominantly in the right posterior quadrant, as it occurs in right ventricular hypertrophy/overload vectorcardiographic type C or special (typical of Chronic Obstructive Pulmonary Disease (COPD) or in severe prolonged mitral stenosis). Additionally, the right peripheral (parietal) bundle branch block or right superior divisional block (RSDB) observed in Brugada Syndrome and arrhythmogenicity right ventricular cardiomyopathy/dysplasia this QRS-loop pattern. The difference lies in the QRS-HP rotation:

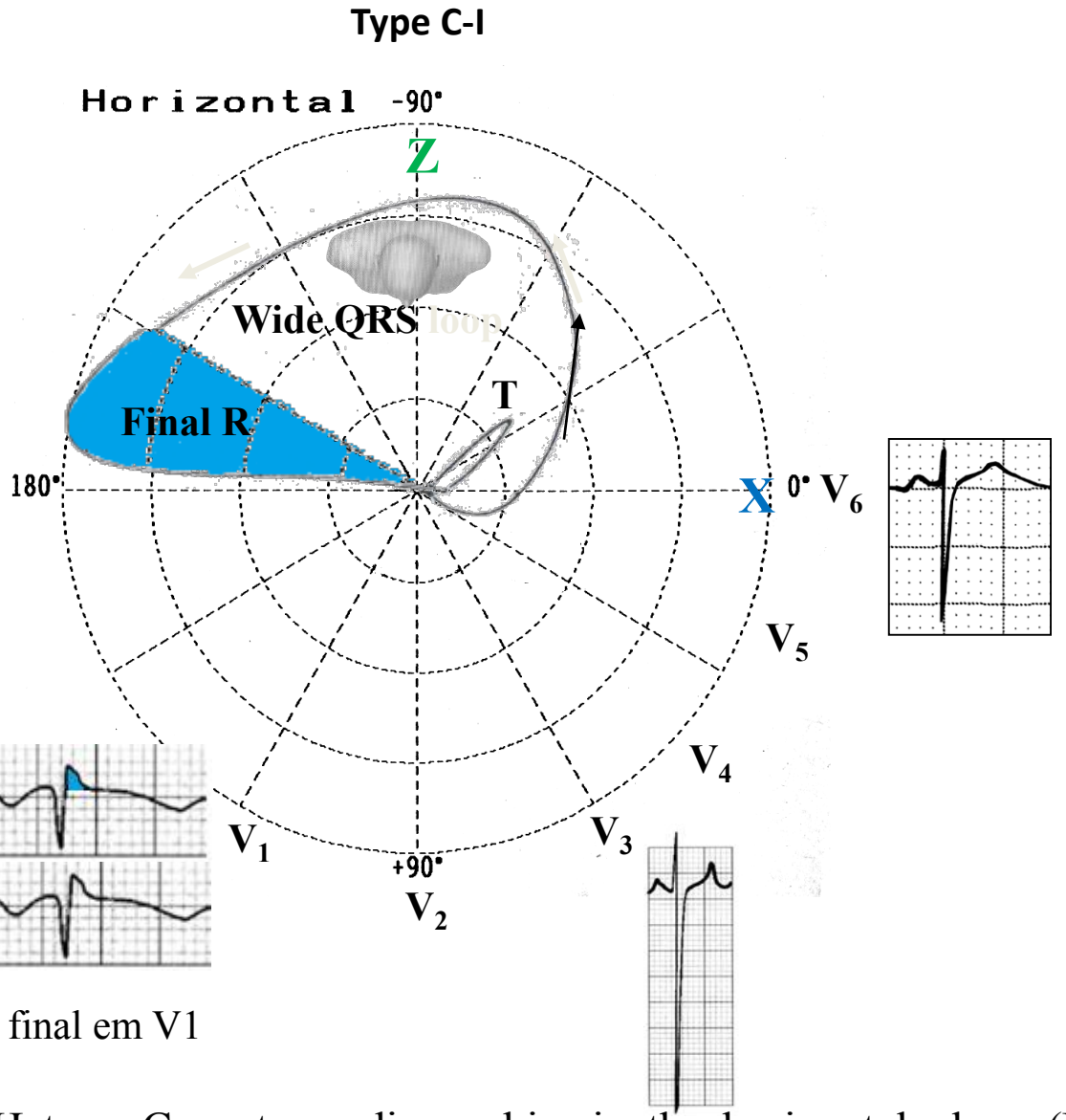
I. Dextrocardia **Clockwise Rotation (CWR)**

II. RVH type C of COPD: **Counterclockwise Rotation (CCWR)**

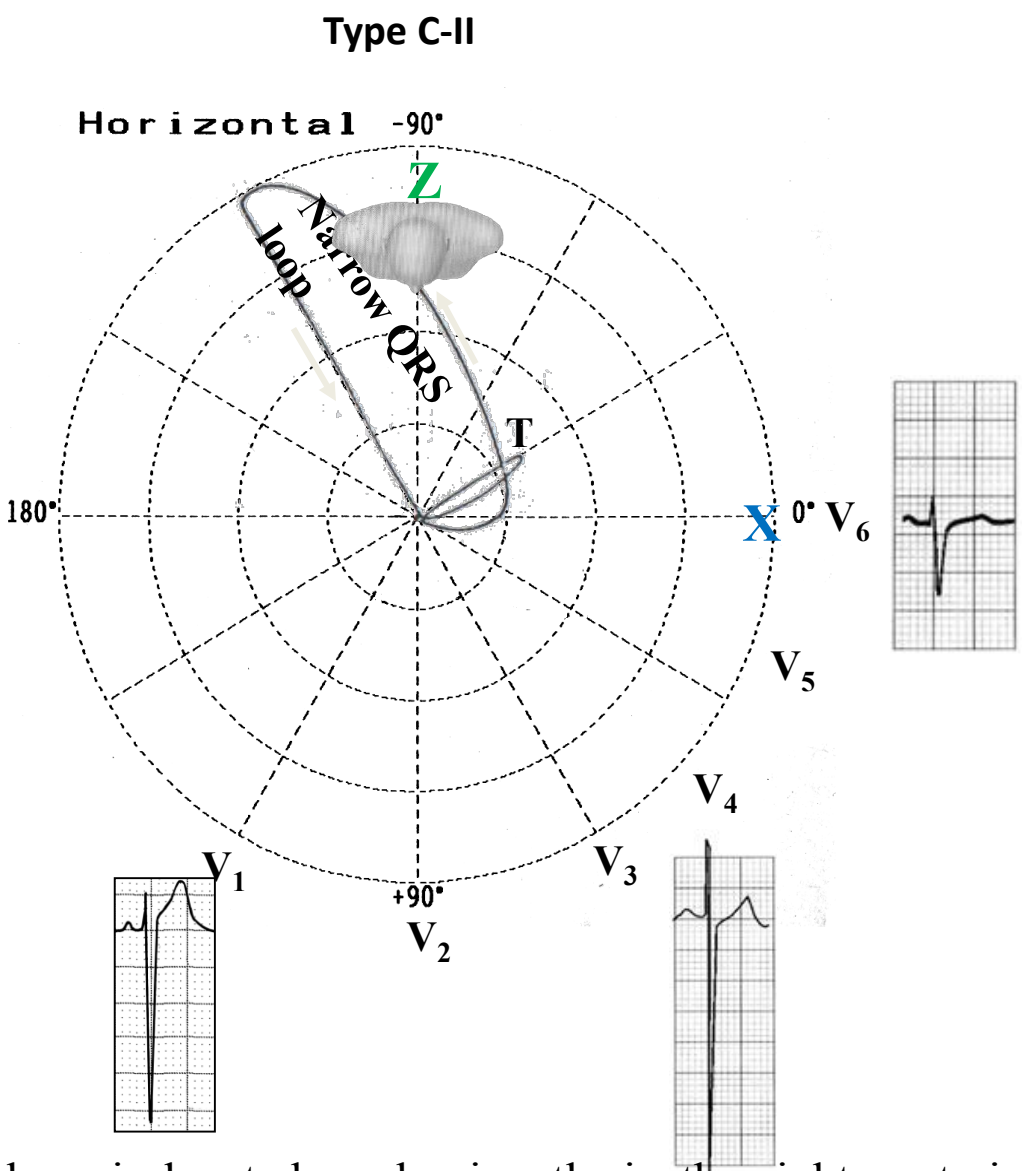
III. Right superior divisional block of the right bundle observed in normal variant, Brugada Syndrome and ARVC/D: **CCWR**

See next slide……

Right Ventricular Hypertrophy VCG type C in the Horizontal Plane typical of COPD

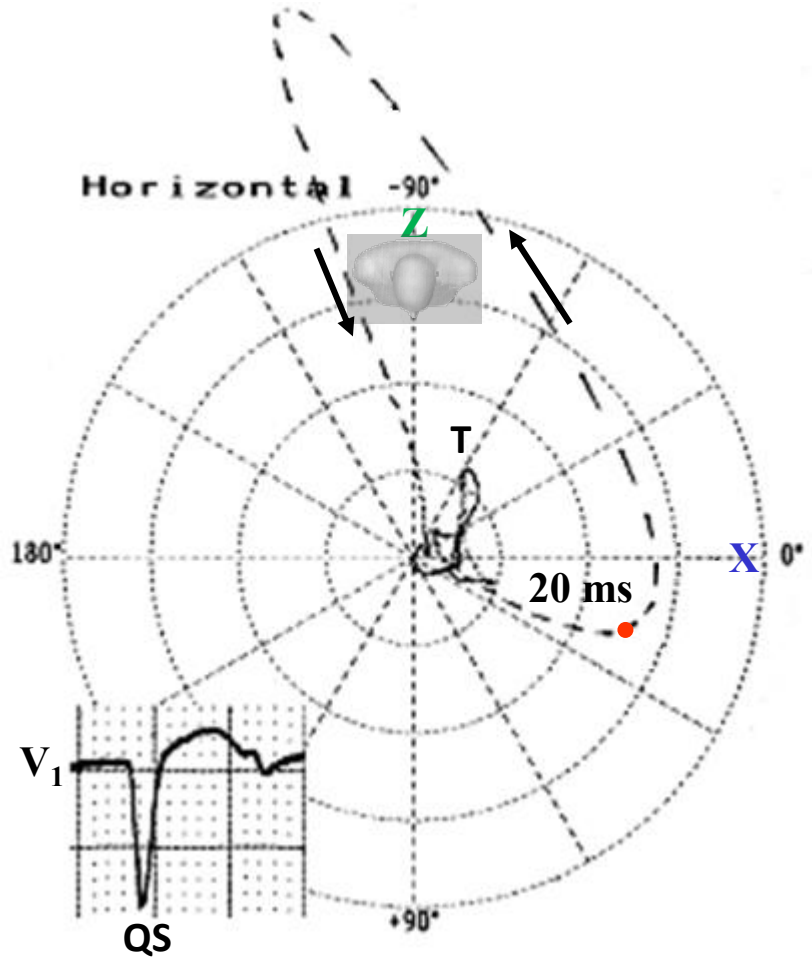


R final em V1



RVH type C vectorcardiographic: in the horizontal plane (HP), the QRS loop is located predominantly in the right posterior quadrant, as it occurs in right ventricular hypertrophy/overload vectorcardiographic type C or special (typical of Chronic Obstructive Pulmonary Disease (COPD) or in severe prolonged mitral stenosis).

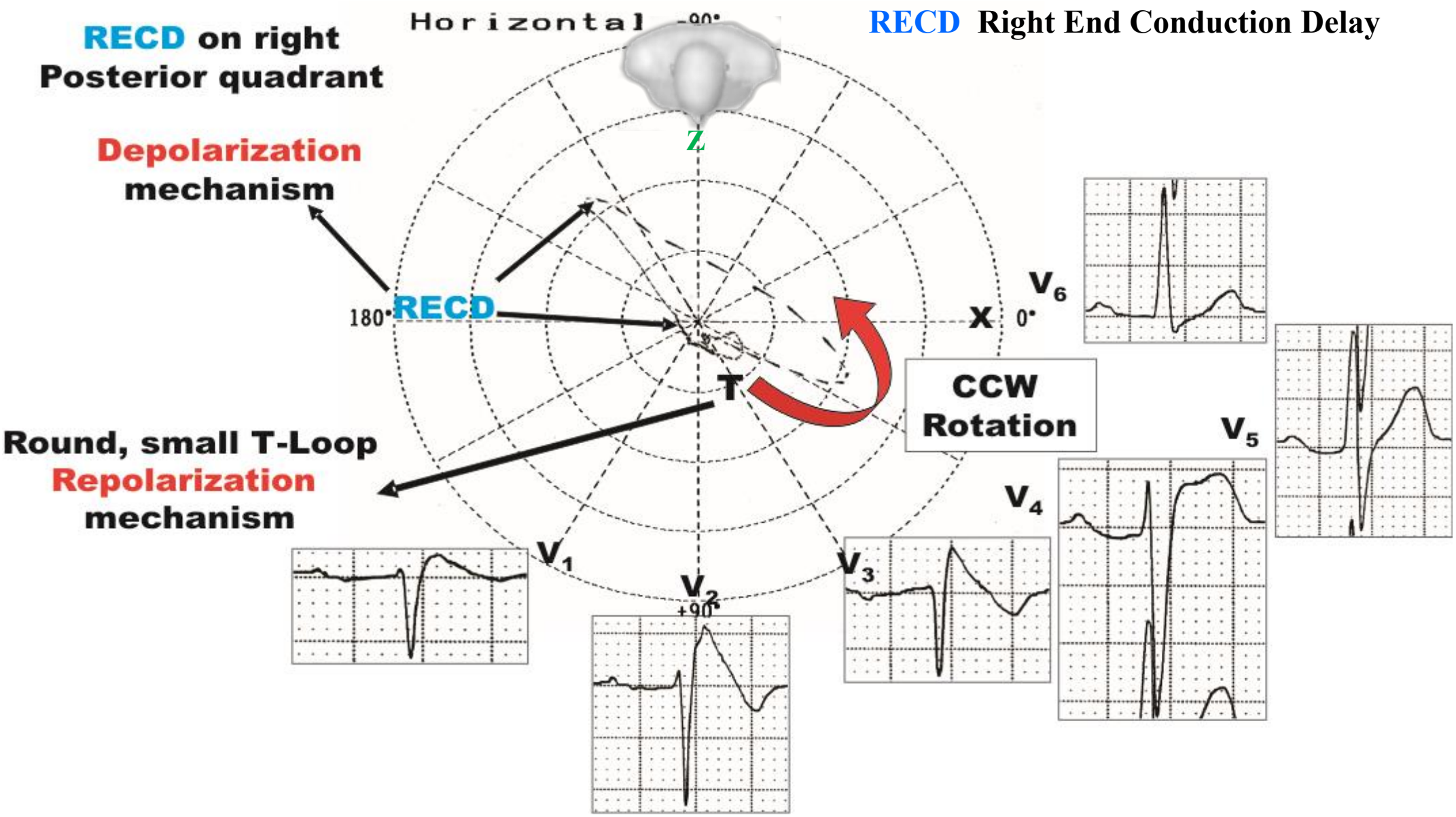
Right superior divisional block of the right bundle observed in normal variant



QRS loop in the HP with significant forces located in the right posterior quadrant, resembling type C RVH, , 10 to 20 ms vector heading leftward and forward: typical of Right superior divisional block of the right bundle (**Luna Filho 1989**), 20 ms vector located in the negative hemifield of V1: QS pattern.

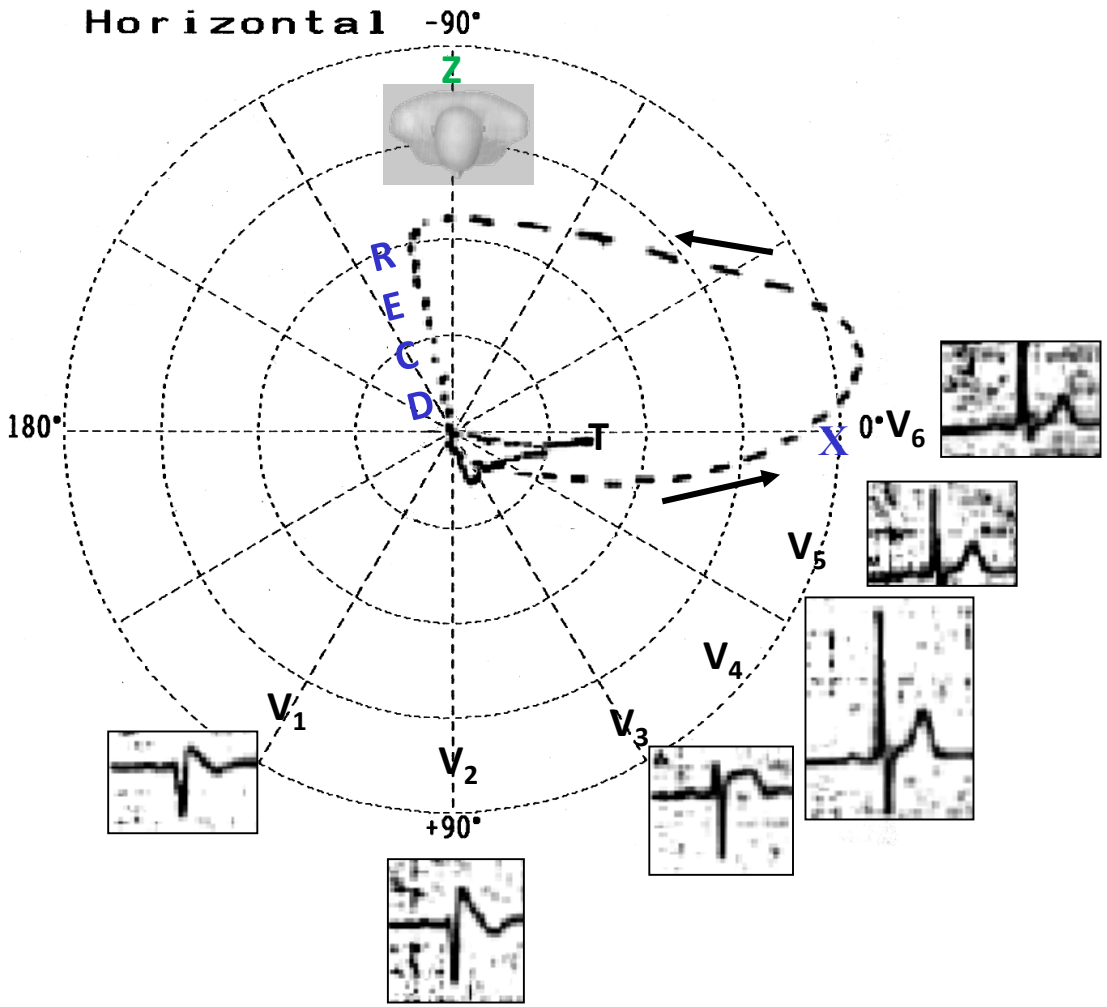
ECG/VCG correlation Horizontal Plane

Identification: **Name:** AS Age: 35yo; **Sex:** Male; **Ethnic Group:** Asian**; Weight:** 72kg; **Height** 1,71m. **Clinical diagnosis:** symptomatic Brugada syndrome



RECD **Right End Conduction Delay**

ECG/VCG correlation in the horizontal plane, Dr. Nava's patient Clinical diagnosis: ARVC/D



Initial 10 to 20 ms vector heading forward and leftward (typical right bundle branch block), CCW rotation and **RECD** located in the right posterior quadrant.

Conclusion: RECD with typical type 1 ECG Brugada pattern in the right precordial leads.

Final diagnosis

1. Biatrial enlargement observed in HP and RSP:

	Horizontal plane	Sagittal planes (right or left)
Maximal P-loop anterior forces	Adults up to 0.06 mV Children yp to 0.o8mV	Adults up to 0.06 mV Children yp to 0.o8mV
Maximal P-loop posterior forces	Up to 0.04 mV	Up to 0.04 mV

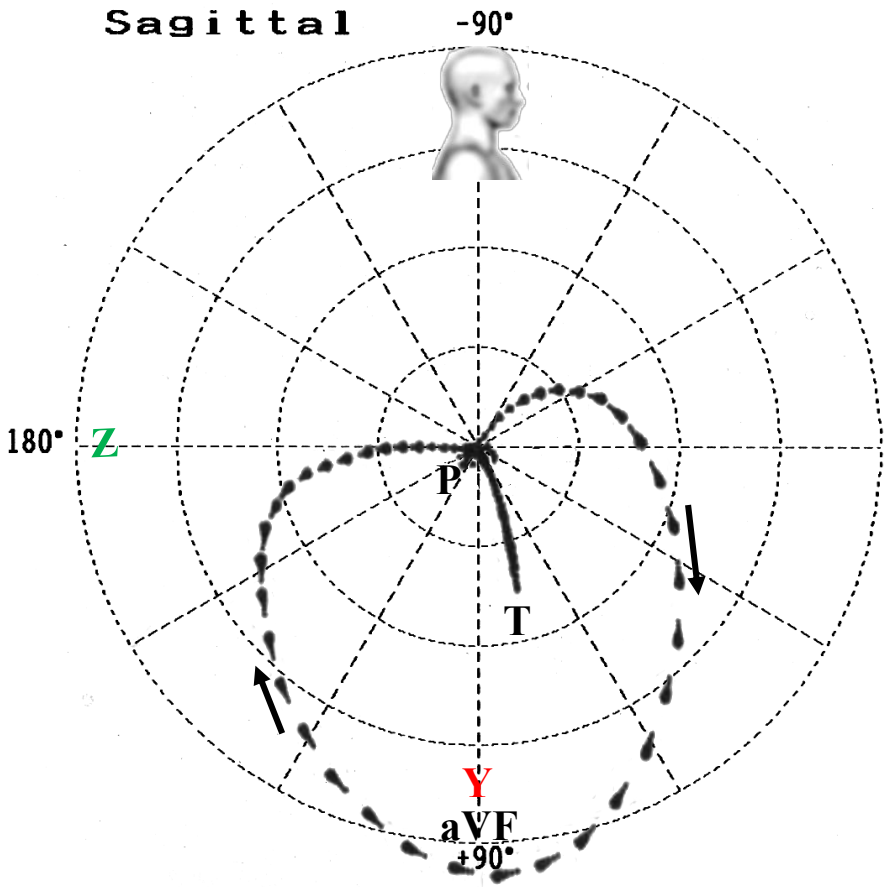
Question: Why do the electrocardiogram and the echocardiogram show no biatrial overload?

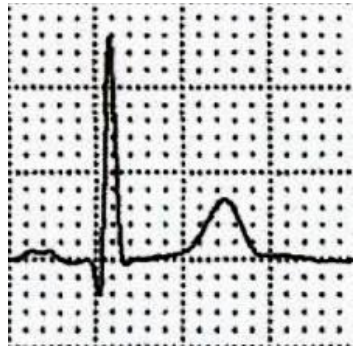
Response: because the ECG sensitivity is lower than the VCG sensitivity for the diagnosis of atrial overloads (**Chou 1986**). The electrocardiogram is a relatively insensitive indicator of the presence of right atrial and ventricular hypertrophy, and in mild cases of right atrial and ventricular hypertrophy the trace will be normal. The Echocardiogram refers only a slight increase in the left atrium size. The left atrium has only mild enlargement in Echo because the patient remained more than two decades with normal function of the artificial bioprosthetic mitral valve.

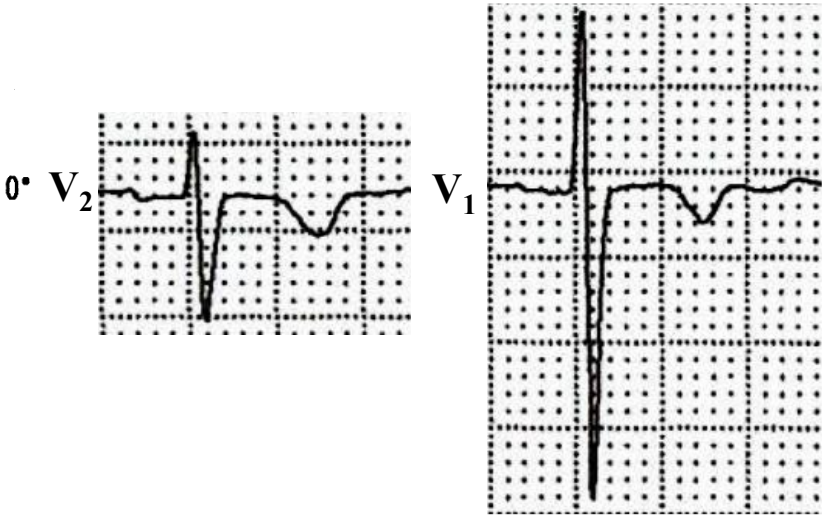
1. Truly dextrocardia with situs inversuss situs or mirror image dextrocardia or L- bulboventricular loop.

	Normal	Simple true dextrocardia
Initial 10 to 20 ms vector	Forward and to the right.	Forward and to the left.
QRS loop rotation in the HP	Counterclockwise.	Clockwise.
Location and voltage of maximal vector on HP	Left posterior quadrant. Voltage does not exceed 2 mV.	Right posterior quadrant. The maximal vector represents the systemic or arterial ventricle. Voltage does not exceed 2 mV.
I	Positive P; QRS and T.	Negative P, QRS and T.
aVL-aVR	The first positive and the second negative: P, QRS and T.	The first negative and the second positive: P, QRS and T.
II-III	Variable.	Usual image of II in III and vice-versa.
V_1 - V_2	rS.	There may be initial q: qRs.
V_5 -V_6	qRs or Rs with positive T wave.	rS or rSr' with negative T.
Progression of R wave in precordial leads	Progressive increase of R wave voltage from V_1 to V_5 .	Reverse progression: R wave of decreasing voltage from V_2 to V_5 .
T loop	To the left and in adults, slightly forward: always positive in V_2 -V_6.	To the right and in adults, slightly forward: negative T in V_5 -V_6.

ECG/VCG correlation in the right sagittal plane

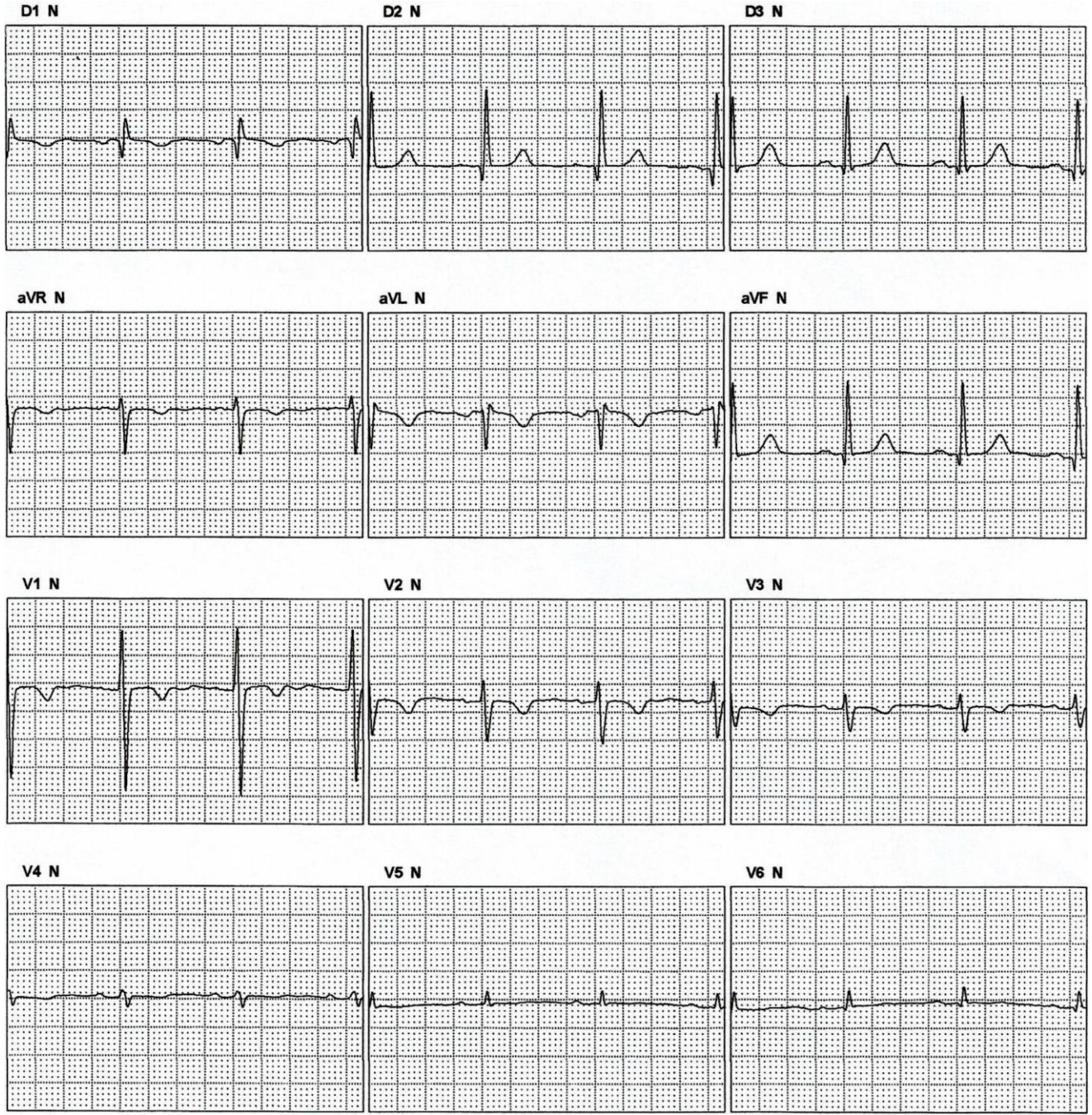






In cases of dextrocadia the pattern of lead V_1 resembles a normal V_2, and V_2 resembles a normal V_1 (Surawicz 2008), such as the present case.

ECG-1



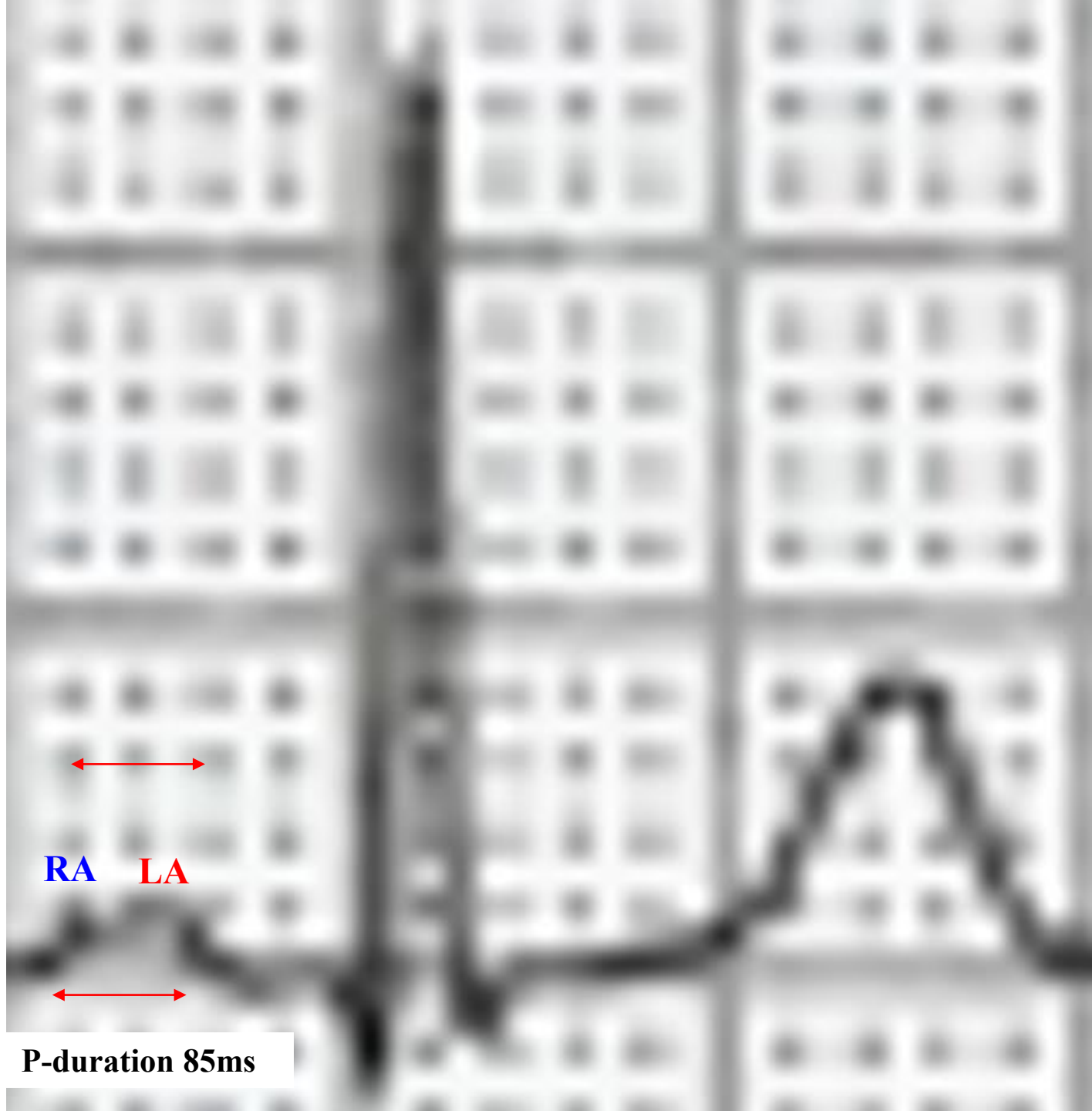
Inverted P-wave in I and aVL.

Precordial R voltage with inverted progression (attenuated on left precordial leads) (**Juncos 2014**).

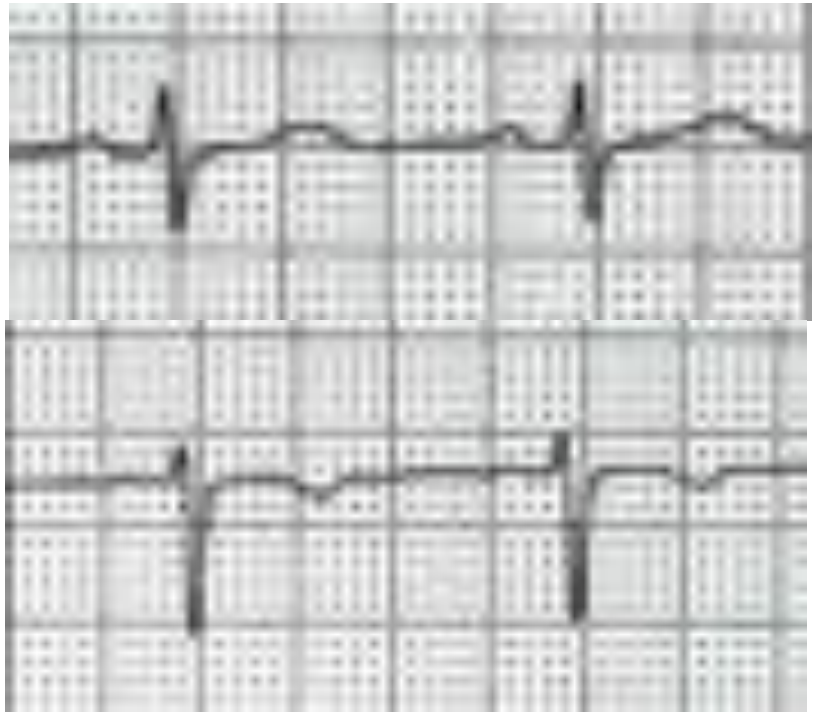
T wave inverted form V1 to V3 and plane from V4 to V6.

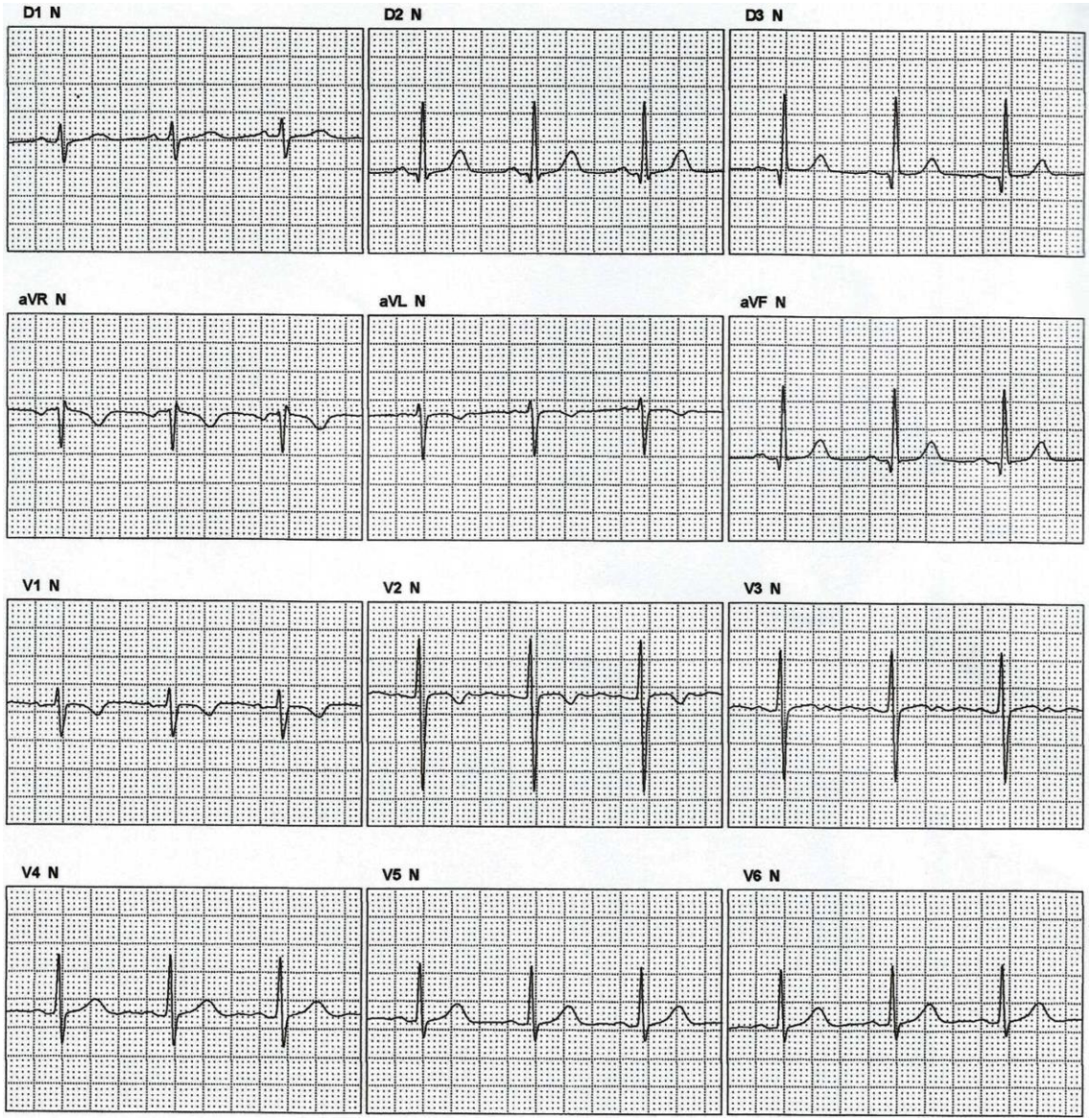
Typical dextrocardia with situs inversus has a characteristic rightward axis with inverted P, QRS, and T waves in lead I when uncorrected leads are used, and a lack of precordial R:S progression as well, plus a decrease in QRS voltage as the leads move from V1 to V6. Moreover, the pattern of lead V1 resembles a normal V2, and V2 resembles a normal V1 (**Surawicz 2008**).

In situs solitus (a term referring to the correct sidedness of structures, are present on the right side of the body and morphologically left-side structures on the left).
The measurements of the P wave should be performed in II lead. On the other hand, in situs inversus in III, because this lead is the mirror image of II.
In the present case, the P duration in III is normal (85ms), and the P-wave is bimodal with the second component (**LA**) larger and broader than the first one as occurs in the left atrial enlargement (**LAE**) in II in situs solitus.

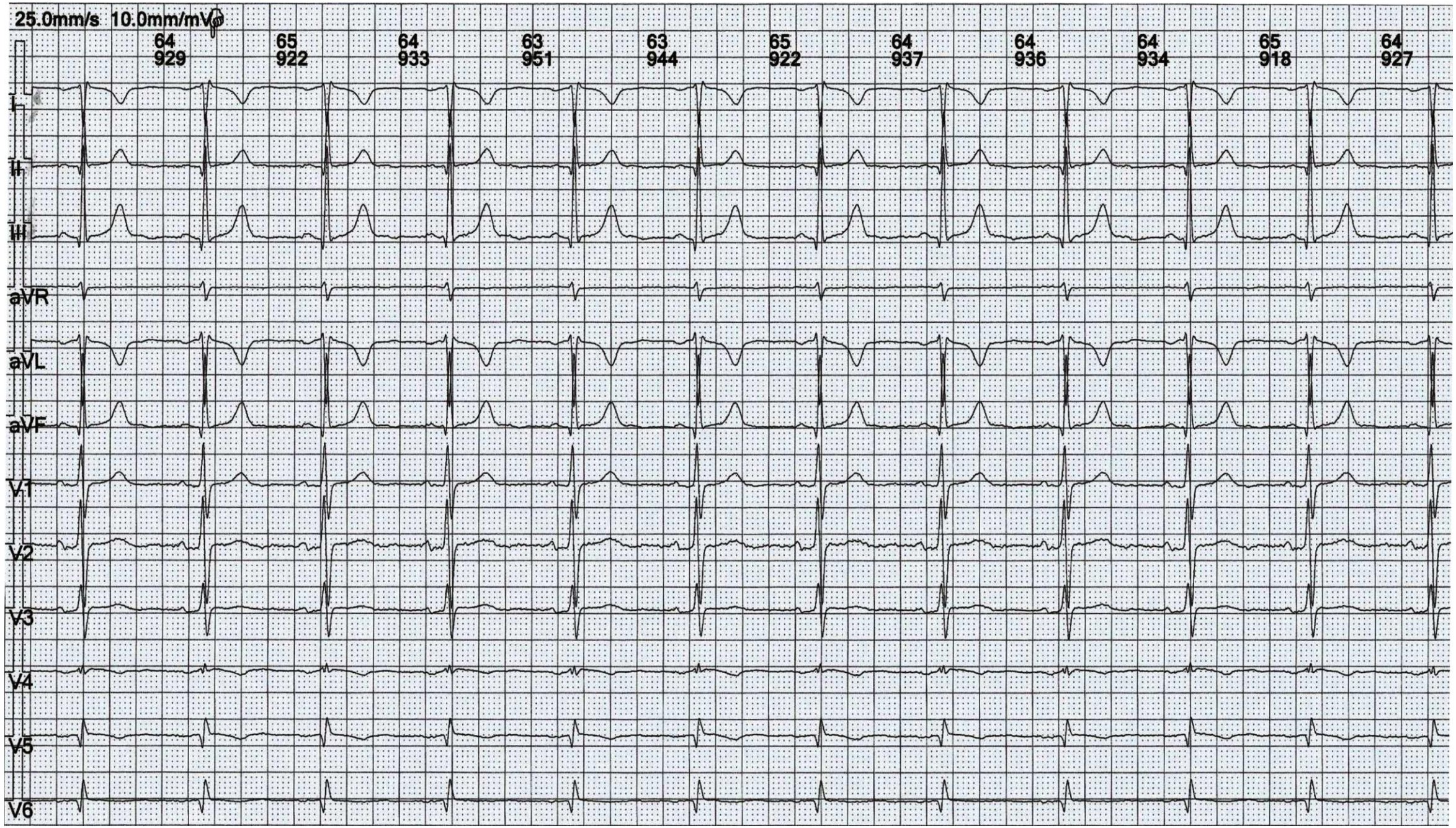


ECG 2

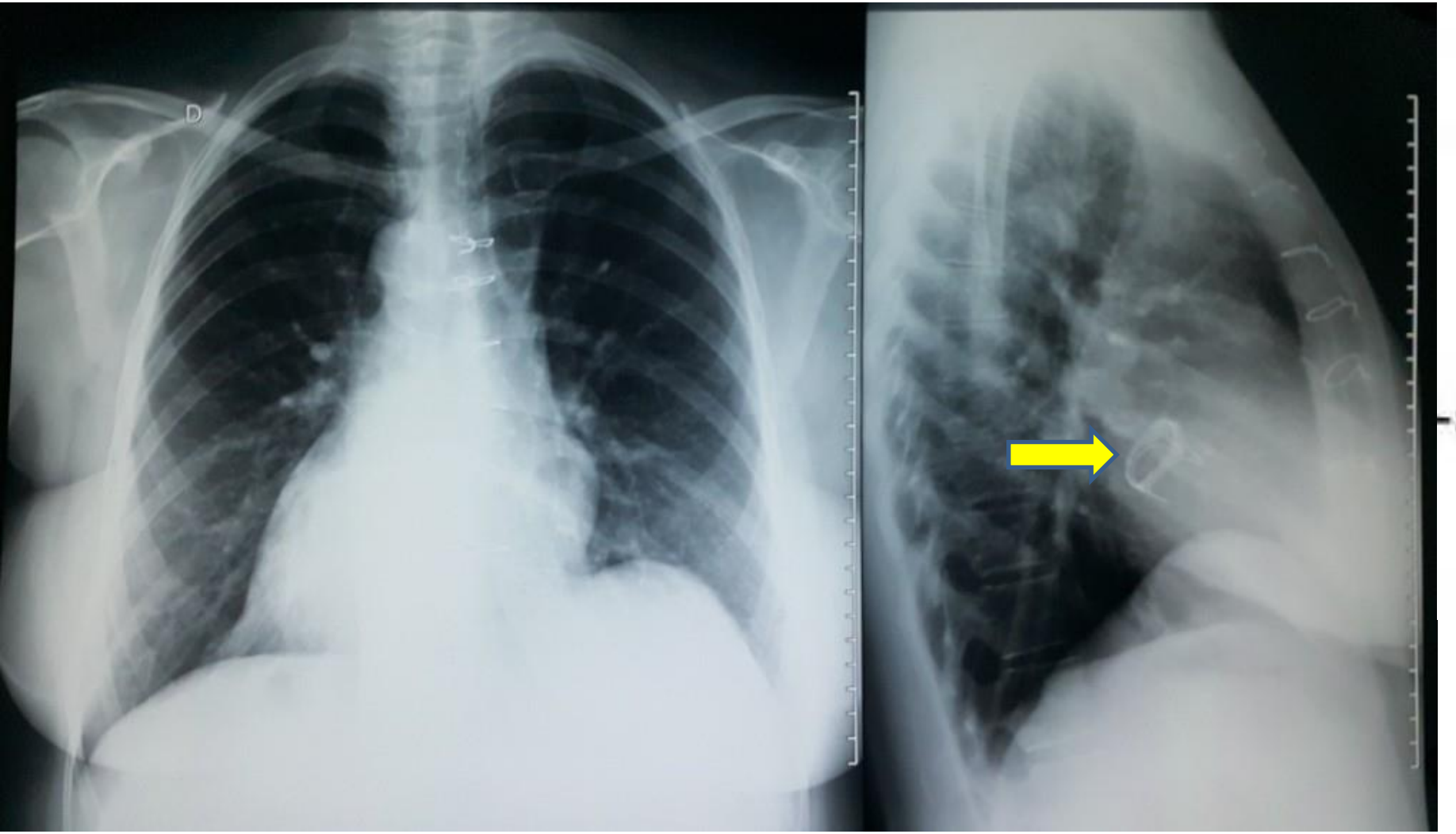


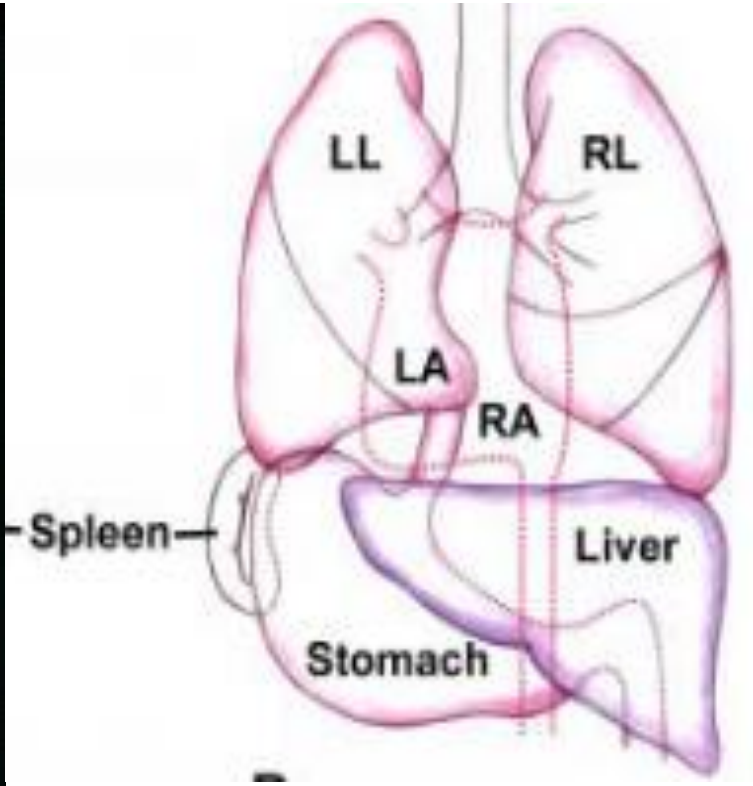


In order to prove the diagnosis of dextrocardia, a second ECG-2 tracing with right precordial leads was performed, which shows normal progression of the R-wave in the horizontal plane. Finally, limb electrodes were reversed and the ECG thus revealed P wave, QRS complexes and T waves with normal orientation and morphology. The use of corrected leads will make the ECG clearly interpretable in all respects.(**Reiffel 2016**)


25.0mm/s 10.0mm/mV
64 929
65 922
64 933
63 951
63 944
65 922
64 937
64 936
64 934
65 918
64 927
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6

Chest x-ray in the posteroanterior and lateral view





Situs inversus. The right lung (RL), left lung (LL), right atrium (RA), and left atrium (LA) are shown.

Chest x-ray in the posteroanterior and lateral view shows a mirror image of normal, the longitudinal cardiac axis is oriented to the right and inferiorly, and the aortic button is on the right. The cardiothoracic index is normal (0.48). artificial bioprosthetic mitral valve in lateral view (yellow arrow). The right hemidiaphragm is lower than the left, the gastric blister is on the right, while dextrocardia is not an isolated phenomenon but part of the situs inversus.

Transthoracic Echocardiogram

Identification:
Name: M; **Sex:** F; **Date of birth:** August 16, 1960; **Age:** 56 years and six months; **Date:** 6 May 2014.

Diagnosis: situs inversus totalis, dextrocardia, dextroapex, atrioventricular and arterial ventricle concordance, normal septoaortic and mitral-aortic continuity, normal systemic and pulmonary drainage; Intact interatrial septum, absence of intracardiac communications or obstructions, intact interventricular septum, discrete left atrial dilation, tissue prosthesis in normal functioning mitral position, leaflets with normal texture and mobility, absence of intracavitary masses or vegetation's, free atrial appendages and normal velocities, aortic arch at righth and without abnormalities along its path, contractile function preserved in both ventricles

Dextrocardia overview

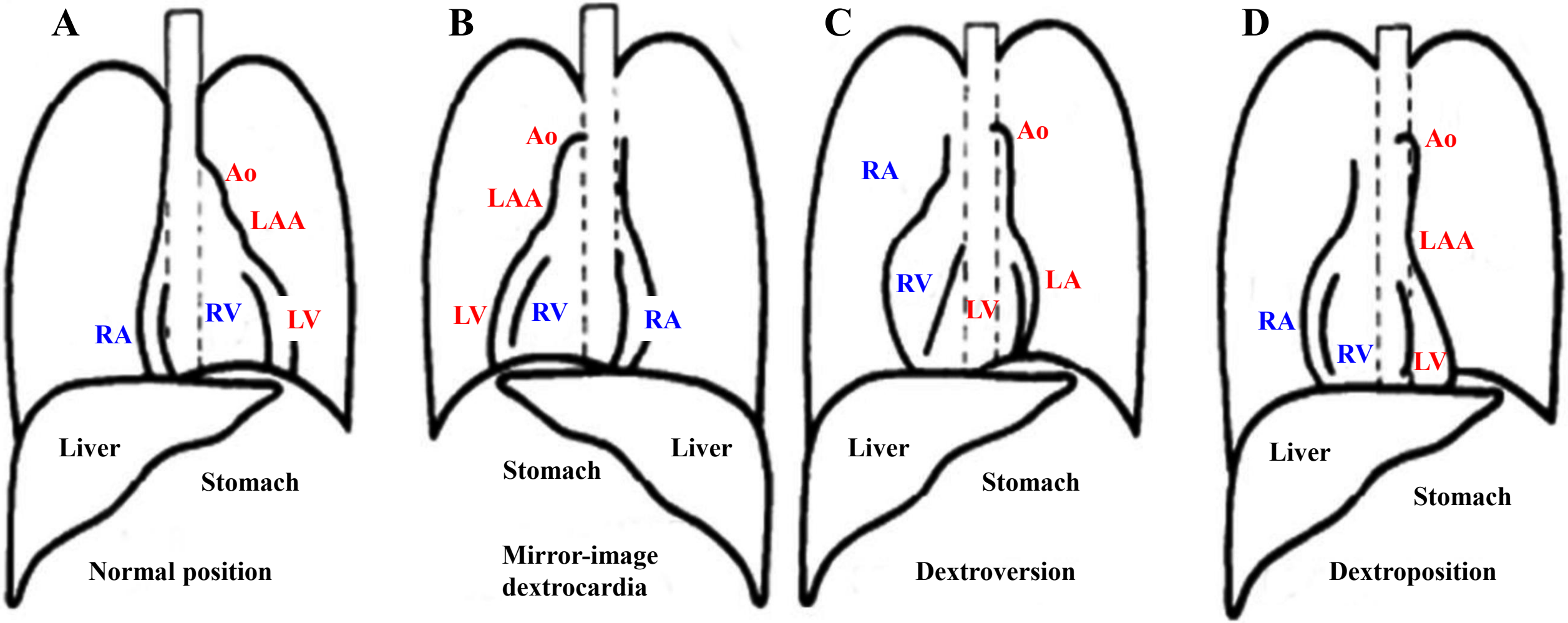
Marco Severino first recognized dextrocardia in 1643. More than a century later, Matthew Baillie described the complete mirror-image reversal of the thoracic and abdominal organs Situs describes the position of the cardiac atria and viscera. Situs solitus is the normal position, and situs inversus is the mirror image of situs solitus. Cardiac situs is determined by the atrial location. In situs inversus, the morphologic right atrium is on the left, and the morphologic left atrium is on the right. The normal pulmonary anatomy is also reversed so that the left lung has 3 lobes and the right lung has 2 lobes. In addition, the liver and gallbladder are located on the left, whereas the spleen and stomach are located on the right. The remaining internal structures are also a mirror image of the normal. The frequency of ECG artifacts due to switched electrodes is 0.4% at the outpatient clinic and 4.0% at the intensive care unit (**Rudiger 2007**). The incidence of dextrocardia was estimated to be 1 in 12,019 pregnancies (**Bohun 2007**); while one of 3 of these will have situs inversus. Totalis occurs in approximately 1 in 5,000 of dextrocardia situs inversus. The incidence is estimated to be around 0.1 to 0.2/1000 population (**Hynes 1973**).

Limb electrode misconnection: Pseudo dextrocardia by exchange of limb electrodes. All P, QRS, and T waves are negative in I, but normal progression of QRS in precordial leads rules out this hypothesis, pointing out the exchange of arm electrodes Simple true dextrocardia: mirror image. Total atrio-visceral situs inversus with no heart disease. SÂP directed to the right and below, pointing at around +120° (III). Negative P wave in VL and I, positive in III. Reverse progression of r wave in precordial leads V2 to V5 (decreasing).

The incidence of situs inversus-associated dextrocardia in the general population is 1: 10,000, while that associated with situs solitus is 1: 30,000 in live births and only 1: 900,000 in the adult population (**McCasckie 1991; Bohun 2007**). Situs inversus is present in 0.01% of the population This large difference in incidence in individuals with situs solitus-associated dextrocardia is due to the higher occurrence of associated cardiac and/or extracardiac diseases, such as tracheoesophageal fistula, pulmonary hypoplasia, imperforate anus, spina bifida and Kartagener's syndrome (**McCasckie 1991; Leung 2006; Van Praagh 1964**). There is a great divergence between the authors regarding the classification of the various types of dextrocardia1. Among the several studies found, one should highlight that of Van Praagh et al (**Van Praagh 1964**) for describing and classifying in a more detailed way the different types of dextrocardia.

True dextrocardia should be differentiated from dextroversion and dextroposition, since this is a non-congenital condition in which the heart can change its side, due to a disease that pulls or pulls this organ to the right antimir, but the cardiac apex is still facing The left side (**Lucchese1980; Garg 2003; Stanger 1977; Calcaterra 1978**). In summary, the heart is moved in the right hemi thorax, with a dextrocardia like aspect. (dextroposition) Example dextrocardia of post right pneumonectomy (**Pârvulescu 2006**). It is interesting because it raises positive and differential diagnosis problems--dextrocardia, associated disorders, serious complications. See example in the next slide……

Classification of the heart in the right hemithorax, dextrocardia, or right-lying



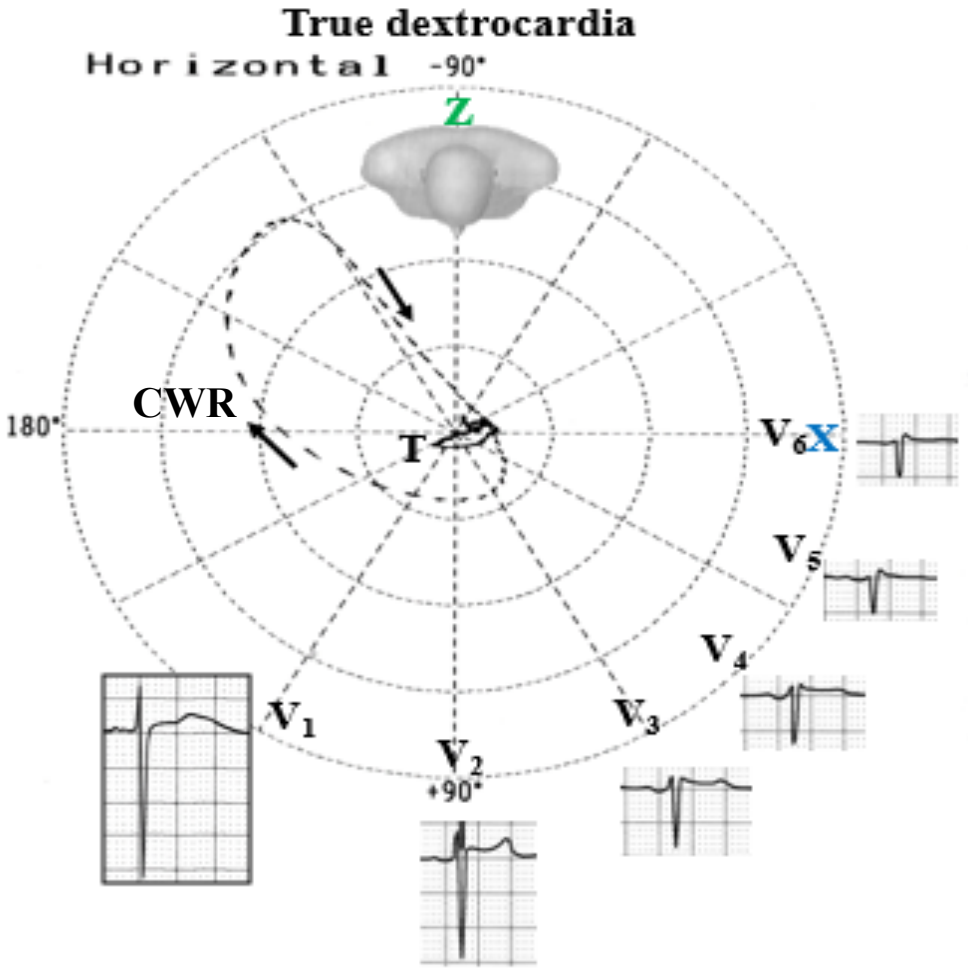
A. **Normally positioned heart for comparison**.

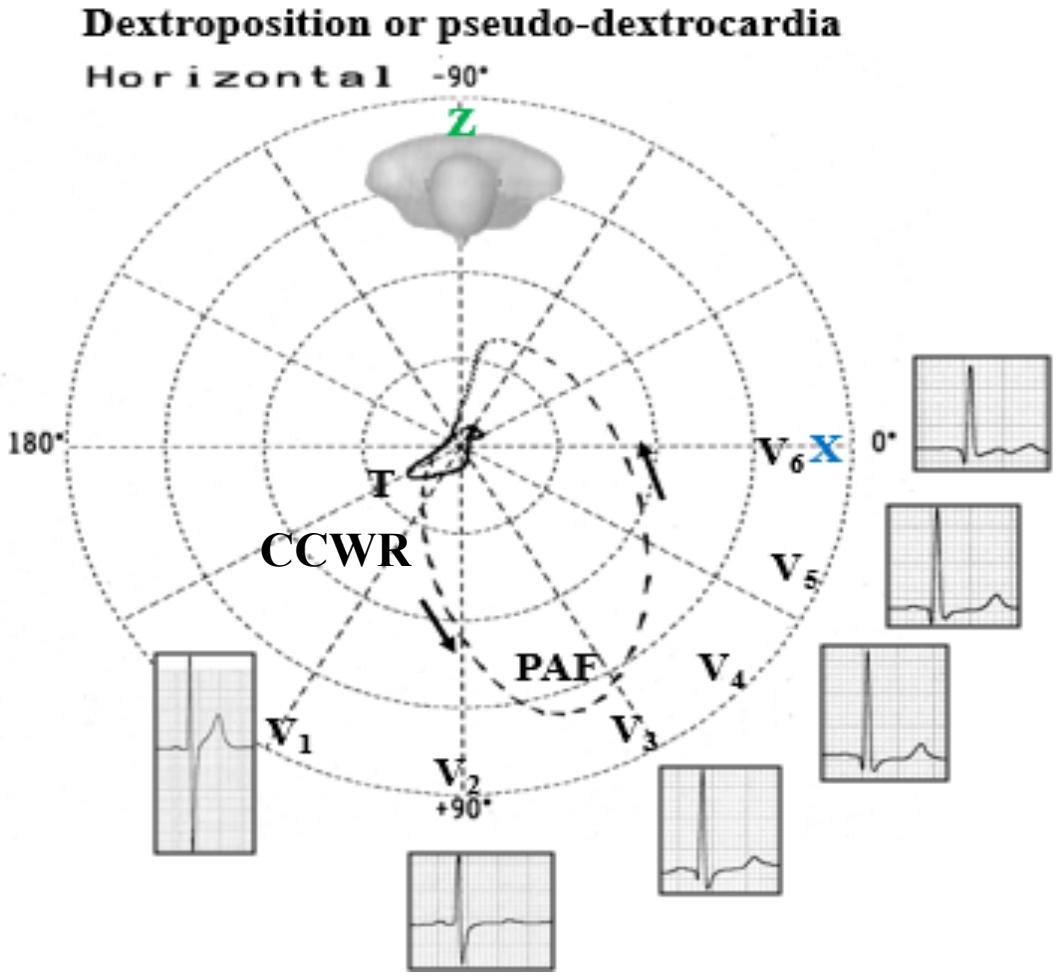
B. **Mirror-image dextrocardia** (in situs inversus and congenital). Inverted P, QRS, and T waves in lead I.

LAA: Left Atrium Appendix; LA: Left Atrium; LV: Left Ventricle; Ao: Aorta; RA: Right Atrium; RV: Right Ventricle

C. **Dextroversion, false dextrocardia or corrected dextrocardia** (in situs solitus and congenital). displacement and rotation of the heart into the right side of the chest but without mirror transposition of the cardiac chambers. Dextroversion is not associated with situs inversus, so the atria are in their normal position, creating positive P waves in lead I. The T waves and QRS complexes may be positive or negative in dextroversion, depending on the presence of other associated cardiac abnormalities. It is the least familiar, but perhaps the most important of the from the clinical point of view because it is frequently accompanied by other intracardiac abnormalities, often of a serious nature. Furthermore the embryogenesis of dextroversion is closely related to the embryogenesis of the cono-truncal region of the heart, and understanding the mechanism of dextroversion sheds light on other congenital malformations. Dextroversion consists of a rotation of the ventricular part of the heart to the right, as in turning the page of a book, with the atria remaining in normal position. Usually there are transposition of the great vessels and a ventricular septal defect. Since the atria are in normal position, the direction of spread of atrial depolarizations the same as in the normal subject and therefore the P wave in lead I is upright, differentiating it from mirror-image dextrocardia. The QRS and T waves in dextroversion depend upon the type and degree of associated intracardiac malformation. If there is no significant associated abnormality, the mean T vector is often rightward, producing a negative T aid the QRS loop is counterclockwise in the FP, producing a Q,. In the past the negative T has often been attributed to ischemia; but, it is likelier that it is due to rotation of the ventricular electric field oilnuts long axis, for the QRS-T angle is usually a narrow one, as in the normal heart (**Grant 1958**).

D. **Dextroposition (in situs solitus and acquired): normal heart, pseudo-dextrocardia:** is shifted to the right by some extracardiac factor such as eventration of the left diaphragm, fibrosis of the right lung, congenital agenesis of right lung (**Xie 2016**), etc., and therefore is also usually easily recognized. Another very rare cause of dextroposition is the so called Poland syndrome includes the features of ipsilateral breast and nipple hypoplasia and/or aplasia, deficiency of subcutaneous fat and axillary hair, absence of the sternal head of the pectoralis major muscle, hypoplasia of the rib cage, and hypoplasia of the upper extremity. In 1841, Sir Alfred Poland described this chest wall anomaly in the Guy's Hospital Gazette while still a medical student based on findings of one cadaver dissection. In his original description, titled "Deficiency of the pectoral muscles," he specifically noted absence of the sternocostal portion of the pectoralis major muscle with an intact clavicular origin, absence of the pectoralis minor muscle, and hypoplastic serratus and external oblique muscles. Poland did not outline the breast hypoplasia or hand deformities in his original description. Although several theories have been advanced to explain the etiology of Poland syndrome, most evidence indicates that it results from a vascular developmental anomaly during the critical sixth week of gestation, with hypoplasia of the subclavian artery causing musculoskeletal malformations. The critical vascular event, known as subclavian artery supply disruption sequence (SASDS), occurs when the medial and forward growth of the ribs forces the subclavian vessel into a U-shaped configuration. The specific region of vessel involvement dictates the clinical manifestation (ie, Poland syndrome, Möbius syndrome, Klippel-Feil syndrome), and more proximal occlusions result in more severe syndromes (**Rodriguez 2017**). See next slide……..





Predominant location of QRS loop: right posterior quadrant	Predominant location of QRS loop: left anterior quadrant
QRS rotation: clockwise	QRS rotation: Counterclockwise
Reverse progression of the R wave in precordial leads	Prominent Anterior QRS Forces (PAF)

The situs is indicated by the position of the liver and right atrium. The atrioventricular relation is shown as normal hut may he reversed without altering the classification. RA = right atrium, LA = left atrium, LAA = left atria1 appendage, RV = right ventricle, LV = left ventricle, A0 = aorta, **CWR:** clock wise Rotation; **CCWR:** Counter clock wise Rotation;

Situs inversus types

Situs inversus can be classified further into situs inversus with levocardia or situs inversus with dextrocardia (**Cotran 1989; Fraser1999; Gutgesell 1998; Hagler 1995; Higgins 1992; Jefferson1980; Perloff 1994; Winer-Muram 1995**). The classification of situs is independent of the cardiac apical position. The terms levocardia and dextrocardia indicate only the direction of the cardiac apex at birth; they do not imply the orientation of the cardiac chambers. In levocardia, the base-to-apex axis points to the left, and in dextrocardia, the axis is reversed.

Isolated dextrocardia is also termed situs solitus with dextrocardia. The cardiac apex points to the right, but the viscera are otherwise in their usual positions. Situs inversus with dextrocardia is also termed situs inversus totalis because the cardiac position, as well as the atrial chambers and abdominal viscera, is a mirror image of the normal anatomy. When situs cannot be determined, the patient has situs ambiguous or heterotaxy. In these patients, the liver may be midline, the spleen absent or multiple, the atrial morphology unclear, and the bowel malrotated. Often, normally unilateral structures are duplicated or absent. The 2 primary subtypes of situs ambiguous include:

1) Right isomerism, or asplenia syndrome, and
2) Left isomerism, or polysplenia syndrome (**Cotran 1989; Fraser1999; Gutgesell 1998; Hagler 1995; Higgins 1992; Jefferson1980; Perloff 1994**)

In classic right isomerism, or asplenia, bilateral right-sidedness occurs. These patients have bilateral right atria, a centrally located liver, and an absent spleen, and both lungs have 3 lobes. The descending aorta and inferior vena cava are on the same side of the spine. In left isomerism, or polysplenia, bilateral left-sidedness occurs. These patients have bilateral left atria and multiple spleens, and both lungs have 2 lobes. Interruption of the inferior vena cava with azygous or hemiazygous continuation is often present. The features of situs ambiguous are inconsistent; therefore, situs ambiguous cases are challenging and require thorough evaluation of the viscera (**Lee 2006**). The location and relationships of the following should be reviewed carefully: abdominal viscera, hepatic veins, superior vena cava, inferior vena cava, coronary sinus, pulmonary veins, cardiac atria, atrioventricular connections and valves, cardiac ventricles, position of the cardiac apex, and aortic arch and great vessels. Other features of situs inversus. Situs inversus occurs more commonly with dextrocardia (**Maldjian 2007**). A 3-5% incidence of congenital heart disease is observed in situs inversus with dextrocardia, usually with transposition of the great vessels. Of these patients, 80% have a right-sided aortic arch. Situs inversus with levocardia is rare (**Gindes 2007**), and it is almost always associated with congenital heart disease (**Fung 2006; Douglas2008; Xu 2008; Van Mierop 1979;Palumbo2008**).

Kartagener syndrome also called primary ciliary dyskinesia (PCD) is typified by bronchiectasis, sinusitis, and situs inversus and affects 20% of patients with situs inversus; however, only 50% of patients with Kartagener syndrome have situs inversus (**Ortega 2007; Kinney 1991; Schidlow1994; Yarnal 1992**). Kartagener's syndrome occurs in approximately 1 in 25 of totalis. It is estimated that about 25% of people with situs

inversus have an underlying condition This disorder affects the sinus and bronchial cilia causing constant sinus and bronchial symptoms that medication cannot rectify. With Kartagener's both are usually present all year rather than being seasonal.

PCD is caused by a defect in the cilia that impairs their normal movements.

Although statistically people with dextrocardia situs inversus do not have any medical problems from the disorder, some are prone to a number of bowel, esophagus, bronchial and cardiac problems. Some of these conditions can be life threatening if left unchecked.

The recognition of situs inversus is important for preventing surgical mishaps that result from the failure to recognize reversed anatomy or an atypical history. For example, in a patient with situs inversus, cholecystitis typically causes left upper quadrant pain, and appendicitis causes left lower quadrant pain. A trauma patient with evidence of external trauma over the ninth to eleventh ribs on the right side is at risk for splenic injury. If surgery is planned on the basis of radiographic findings in a patient with situs inversus, the surgeon should pay careful attention to image labeling to avoid errors such as a right thoracotomy for a left lung nodule.

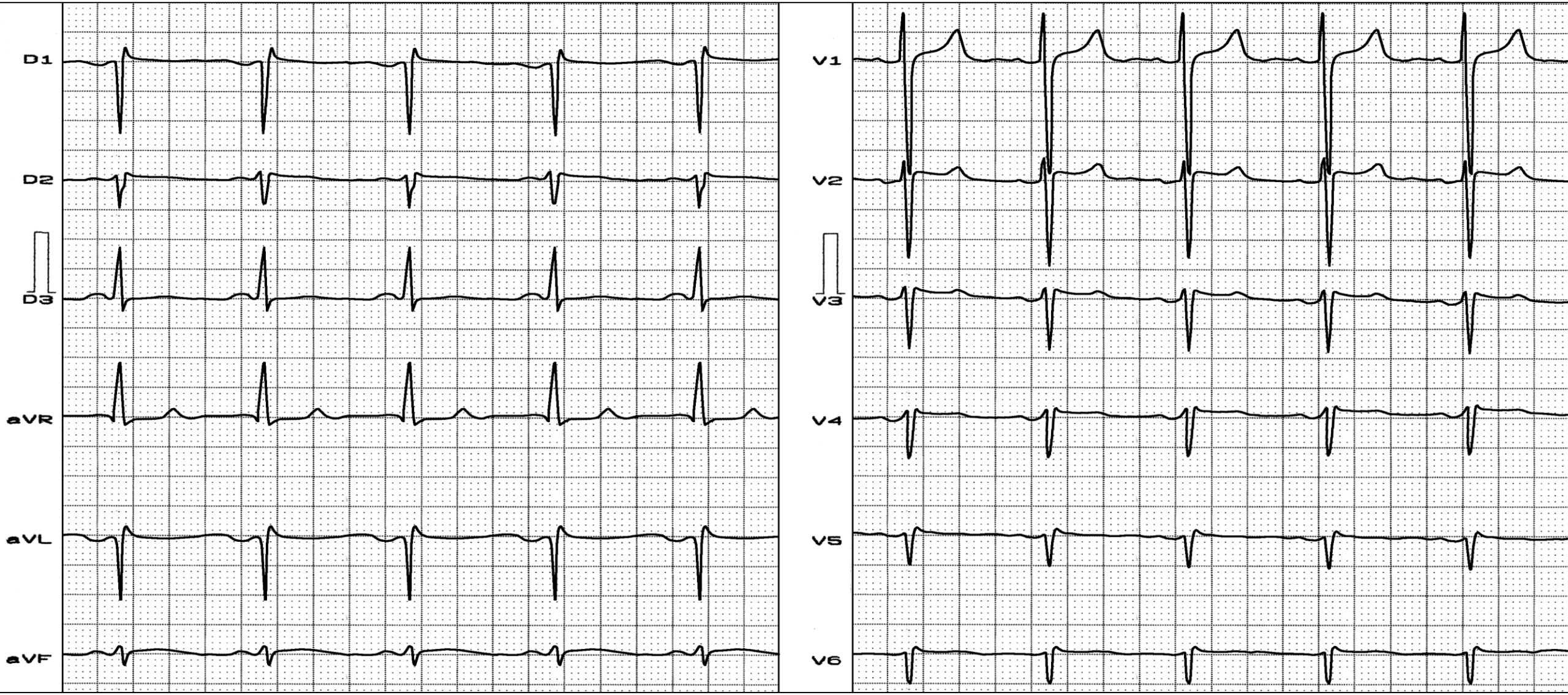
Dextrocardia is a congenital defect in which the heart is situated on the right side of the body. There are two main types of dextrocardia, Dextrocardia of embryonic arrest (also known as isolated dextrocardia) and dextrocardia situs inversus. Dextrocardia situs inversus is further divided.

Dextrocardia of embryonic arrest: In this form of dextrocardia, the heart is simply placed farther right in the thorax than is normal. It is commonly associated with severe defects of the heart and related abnormalities including pulmonary hypoplasia.

Dextrocardia situs inversus: Dextrocardia situs inversus refers to the heart being a mirror image situated on the right side. For all visceral organs to be mirrored, the correct term is dextrocardia situs inversus totalis.

Other examples

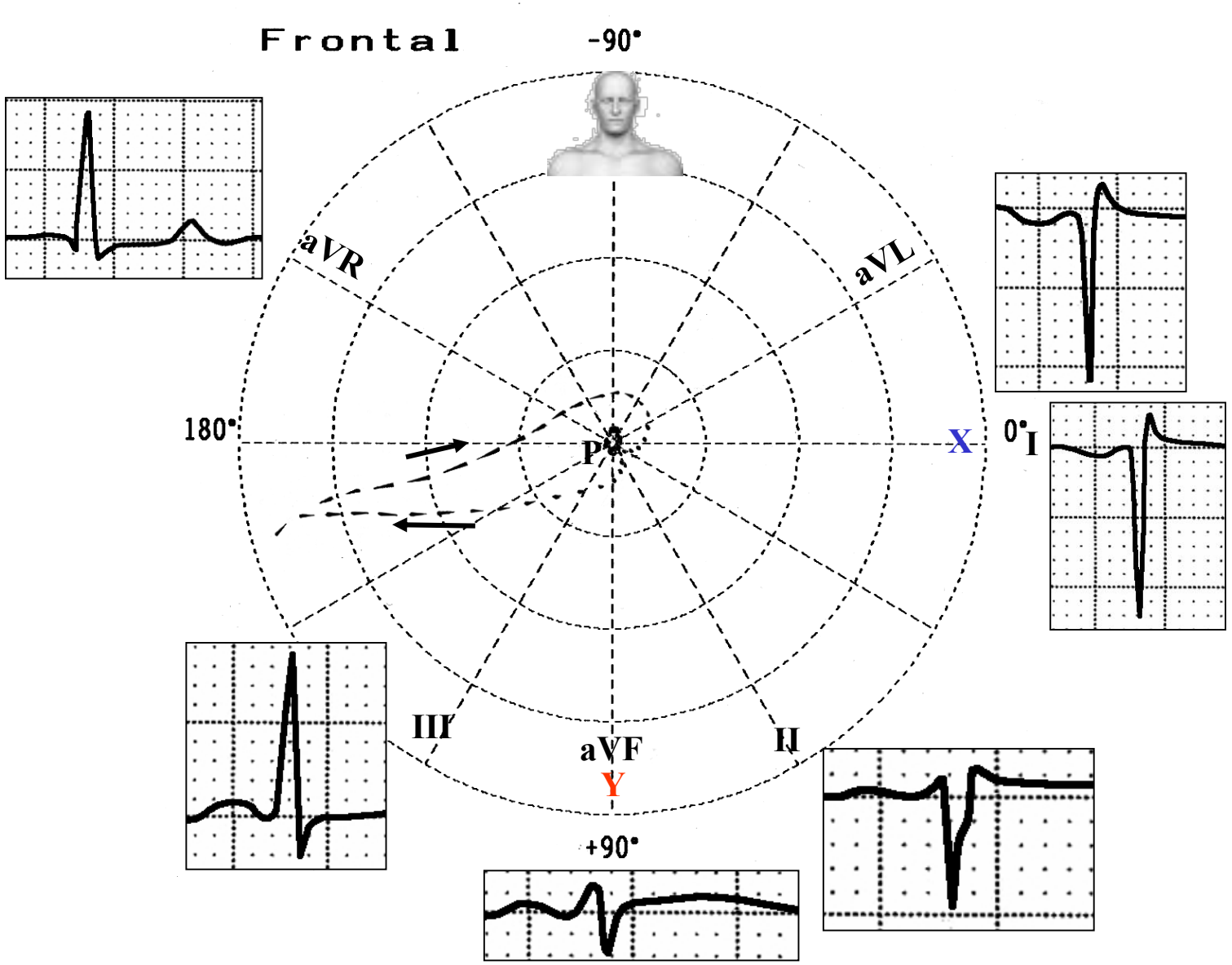
Name: JHY; **Age**: 39 y.o.; **Gender**: Male; **Ethnic Group**: Caucasian; **Weight**: 68 kg; **Height**: 1.70 m; **Biotype**: Asthenic; **Date**: 11/08/2010; **Medications in use**: None.

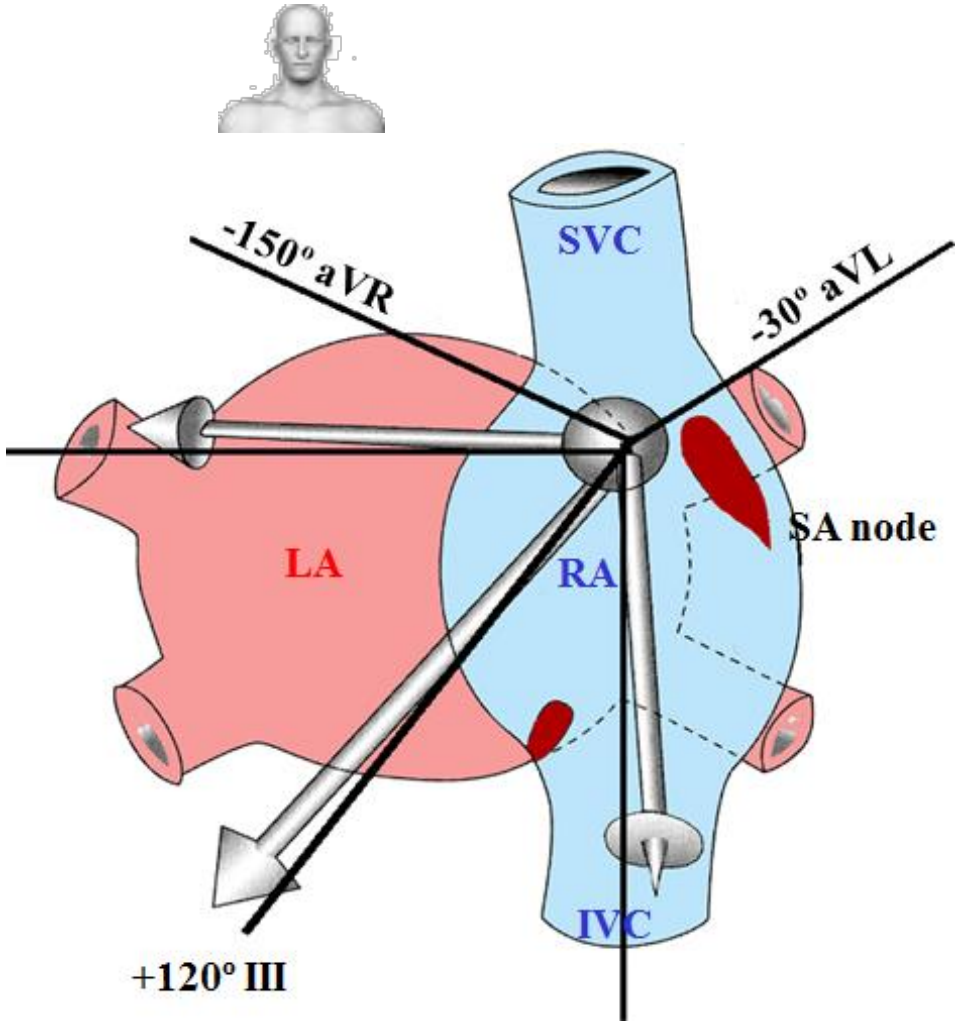


Clinical diagnosis: True dextrocardia.

ECG diagnosis: Negative P waves in lead I and aVL, V5-V6, and positive P waves in right leads, aVR and III, negative P, QRS and T waves in I, reverse progression: R wave of decreasing voltage from V_2 to V_5 .

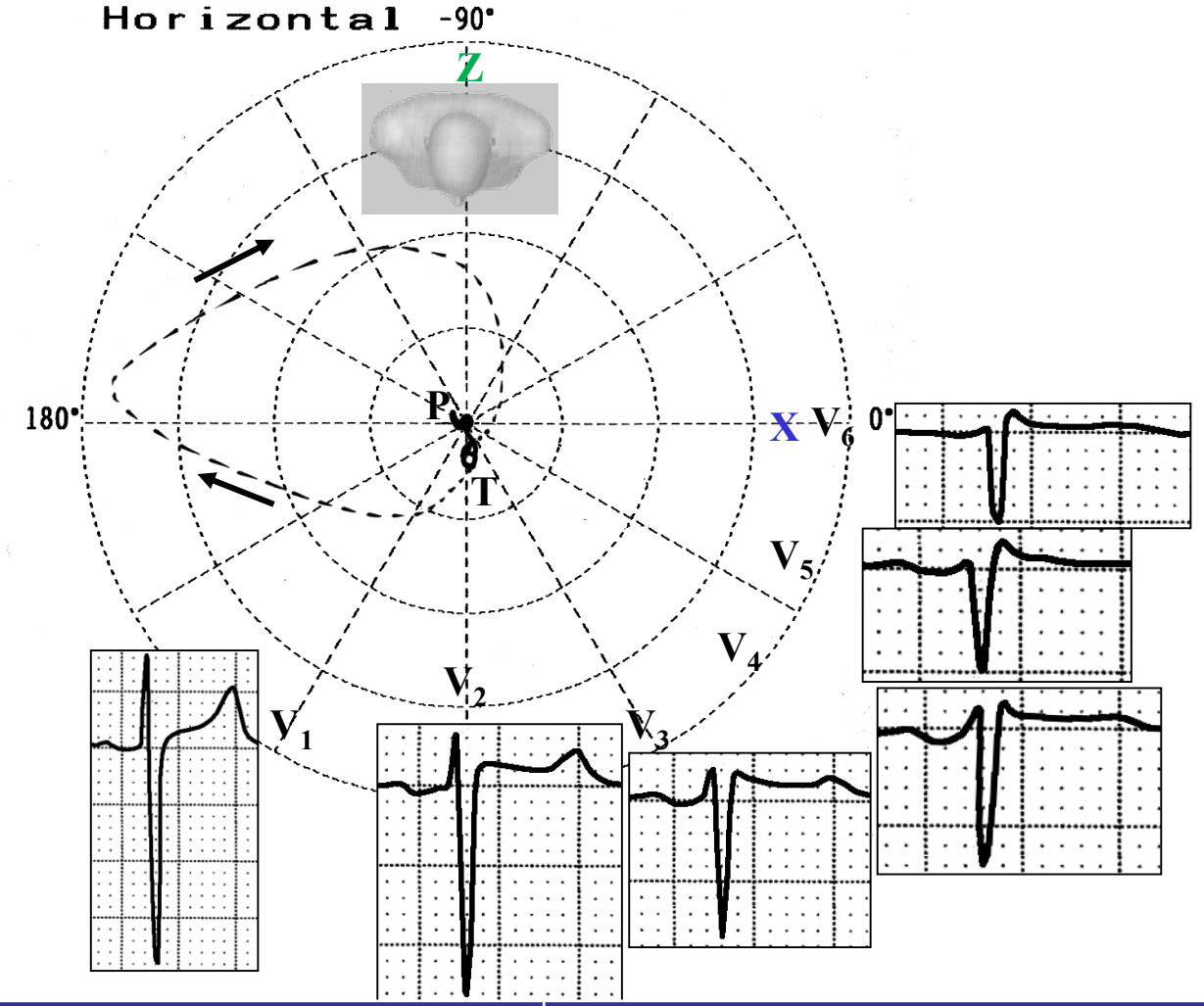
ECG/VCG correlation in the Frontal Plane





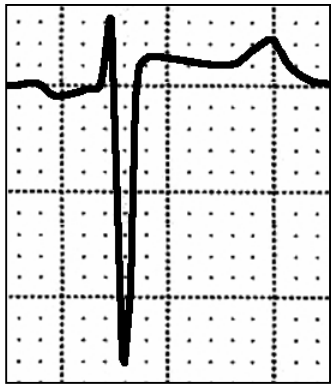
ECG leads must be placed in reversed positions on a person with dextrocardia. In addition, when defibrillating someone with dextrocardia, the pads should be placed in reverse positions. That is, instead of upper right and lower left, pads should be placed upper left and lower right.

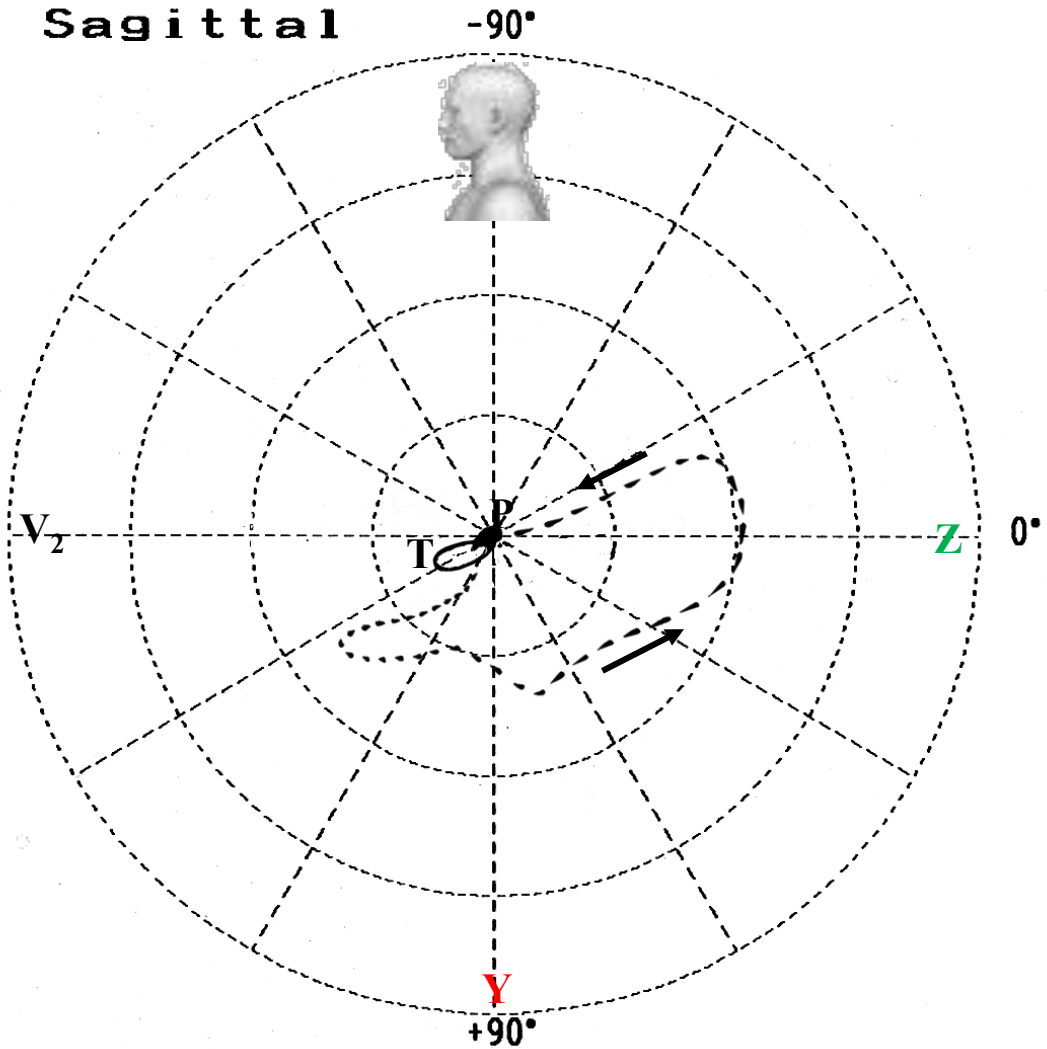
ECG/VCG correlation in the Horizontal Plane



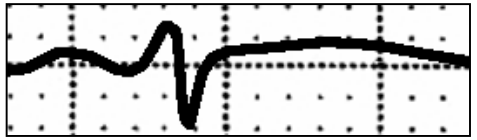
Dextrocardia	Normal heart
Predominant location of QRS loop: Right posterior quadrant	Predominant location of QRS loop: Left posterior quadrant
QRS Rotation: Clockwise Rotation (CW-R)	QRS Rotation: Counterclockwise Rotation (CCW-R)
Reverse Progression of the R wave in precordial leads	Normal Progression of increased R wave V1-V4

ECG/VCG correlation in the Left Sagittal Plane

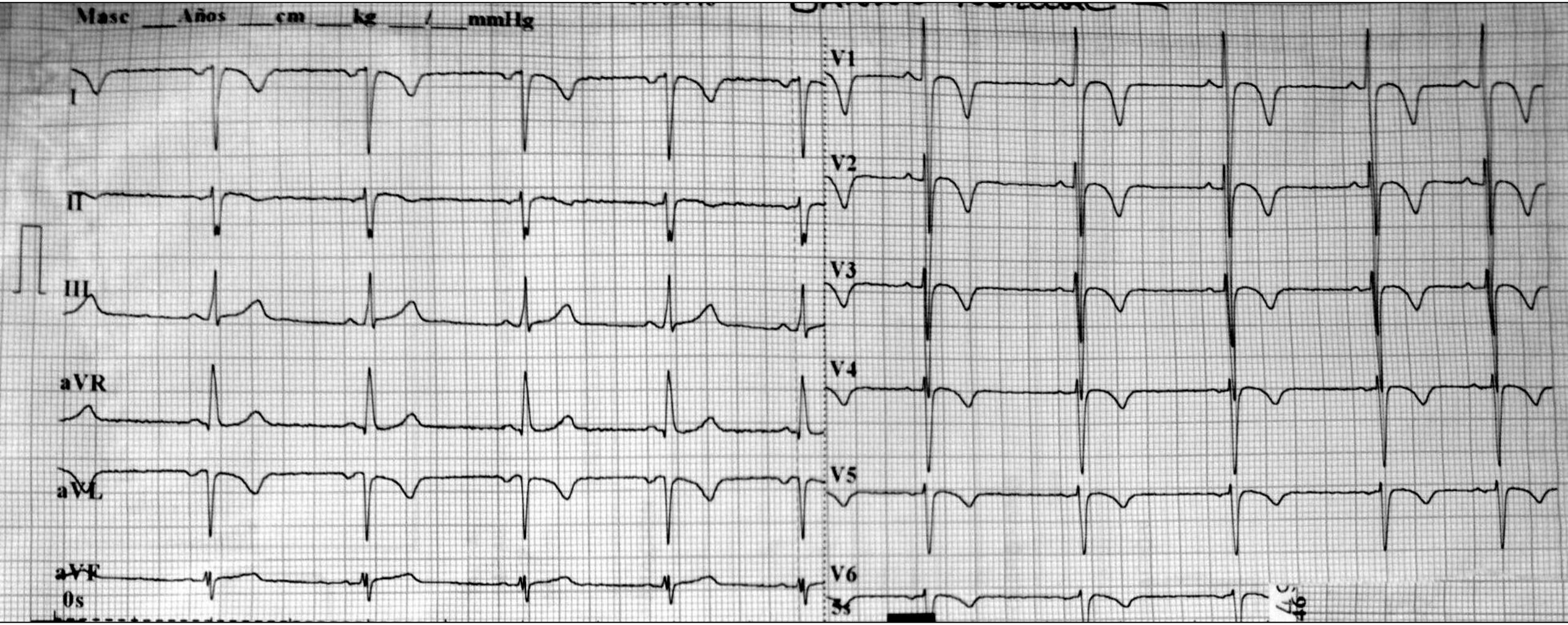




aVF



Dextrocardia with situs inversus

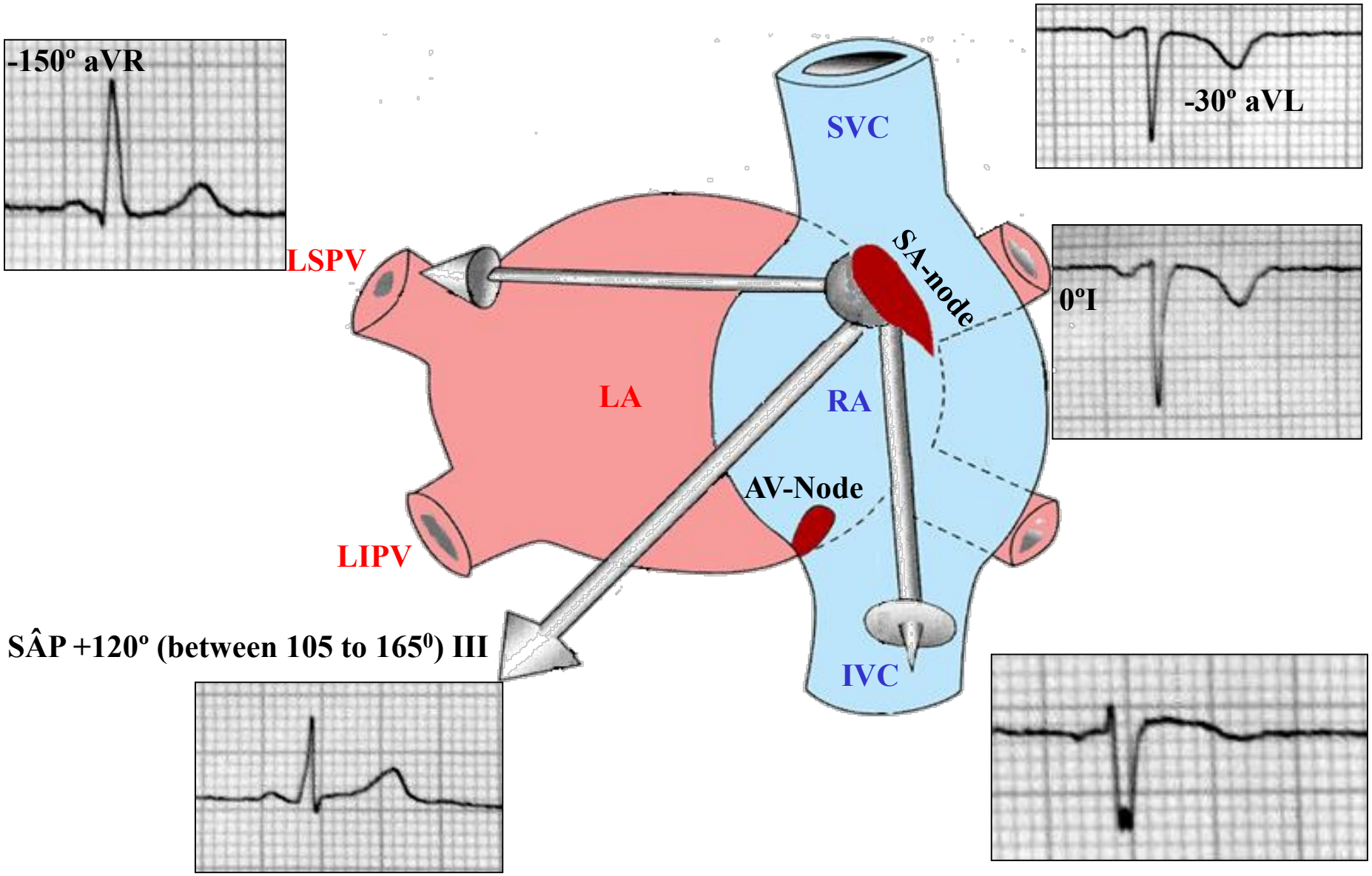


The ECG findings are: **Lead I:** P, QRS and T waves inverted or upside down. **Lead II:** represents the usual lead III and vice versa. aVR and aVL are reversed and prominent negative deflections are seen in aVL rather than in aVR. aVF is unaffected. P axis (SÂP) to the right and below, pointing at around +140º. Negative P wave in aVL and I, positive in III and aVR.

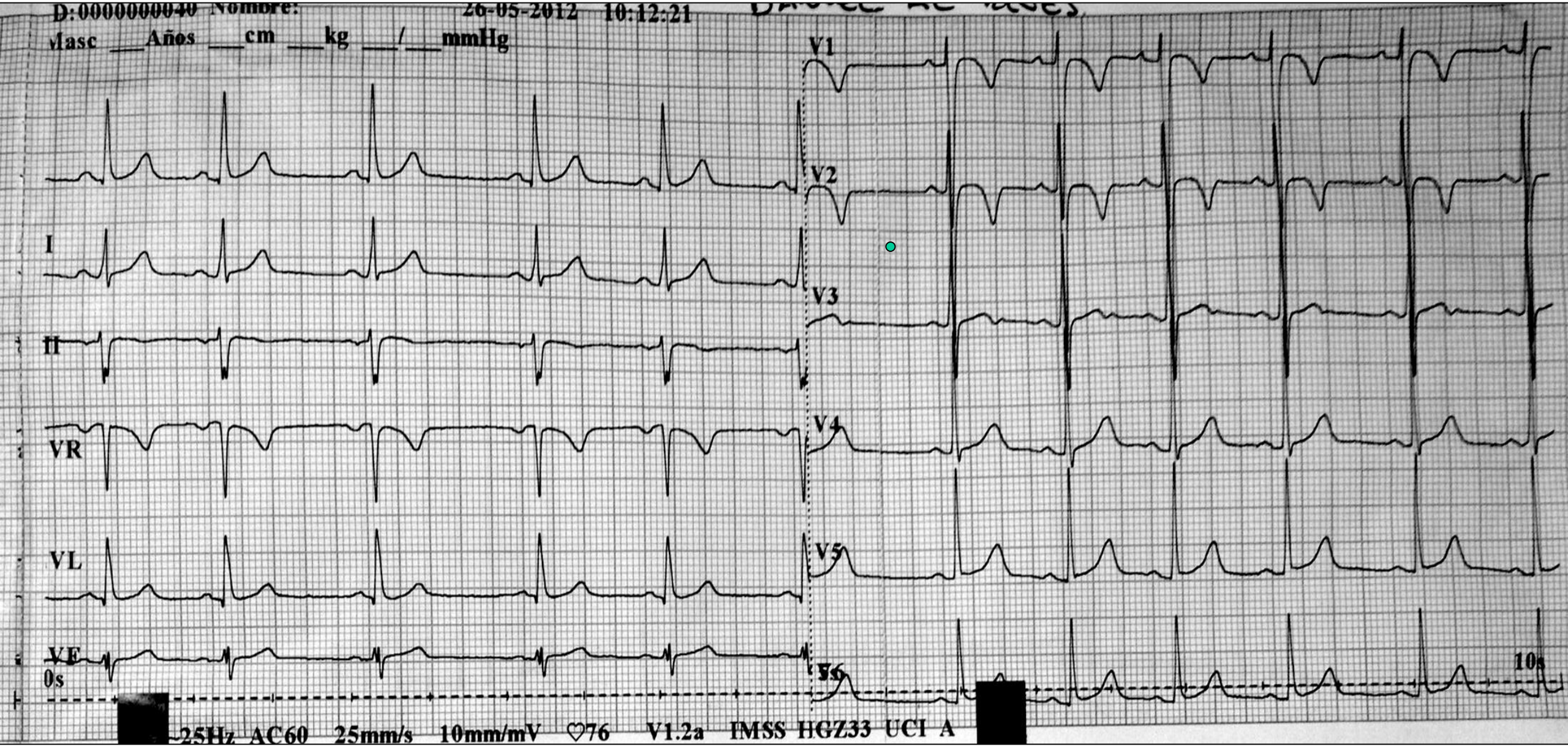
Precordial leads: From V1 to V6: QRS complexes show progressive decreasing R/r wave amplitude: Reverse progression of r wave in precordial leads from V1 to V6 (decreasing). V1 is the equivalent of the usual V2 and vice versa.

Conclusion: True dextrocardia.

Atrial inversion (situs inversus)



SÂP vector directed from left to right, top to bottom, pointing at around +120° (III). This fact will yield negative P wave in aVL and I, positive in III and aVF and variable II and aVR.



This ECG was performed latter willfully inverting the cables of both limbs as the position of the precordial electrodes, which were placed right at points corresponding to the left leads.

Conclusion: Normal ECG in both planes.

Causes and symptoms

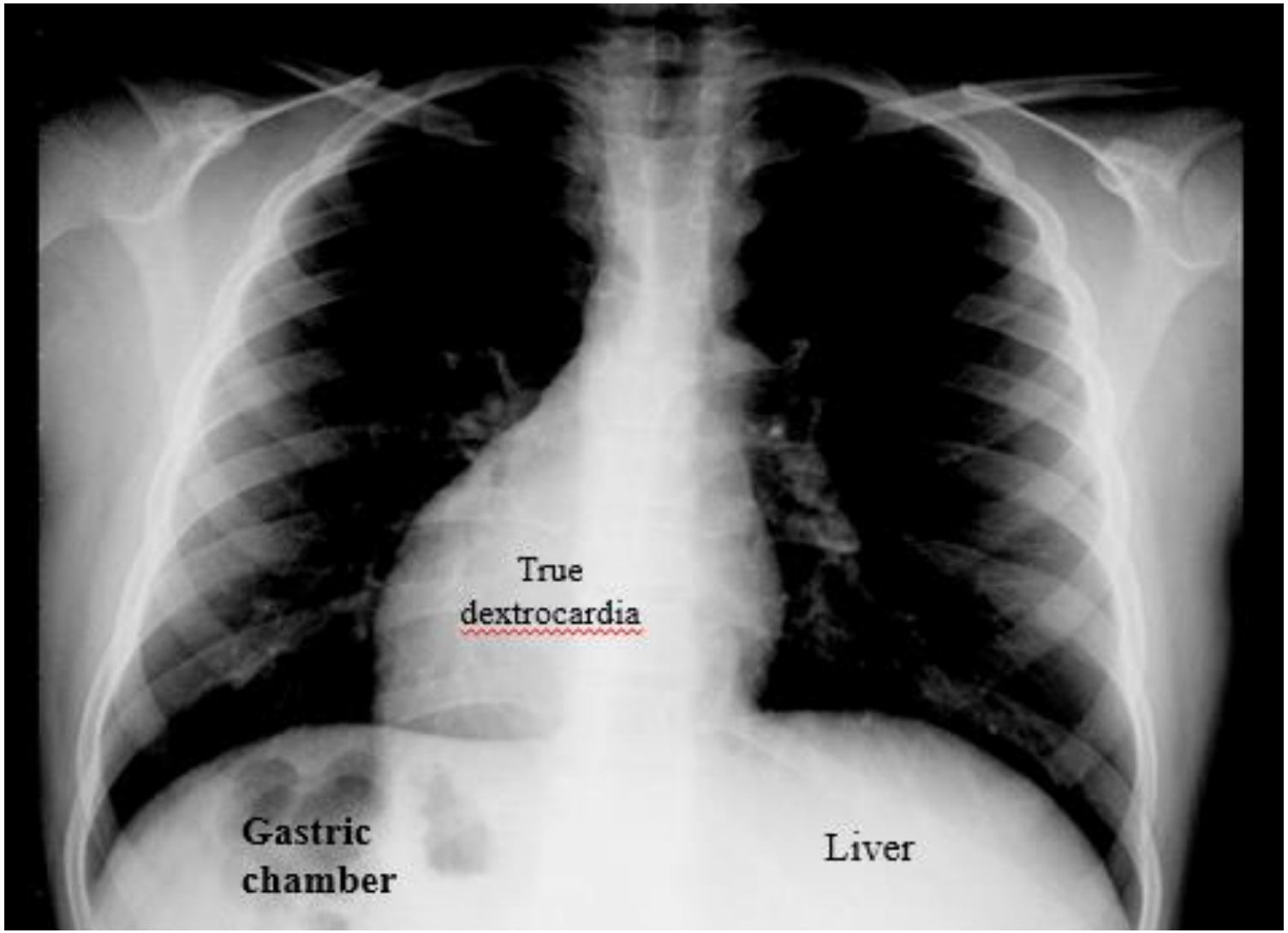
Early in the normal development of an embryo, the tube-like structure that becomes the heart forms a loop toward the left, identifying the left/right axis along which the other organs should be positioned. Although the mechanism that causes the heart loop to go left is not fully understood, at least one gene has been identified to have a role in this process. However, it is thought that many factors may be involved in causing situs inversus. Rarely, situs inversus can run in families, but most often it is an isolated and accidental event occurring in an individual for the first time in the family.

Most people with situs inversus have no medical symptoms or complications resulting from the condition. Although only 3-5% of people with situs inversus have any type of functional heart defect, this is higher than the rate of heart defects in the general population, which is less than 1%. Dextrocardia occurs in approximately 0.01% of live births and can be discovered in various clinical settings and at various patient ages.

The condition affects all major structures within the chest and abdomen. Generally, the organs are simply transposed through the sagittal plane. The heart is located on the right side of the thorax, the stomach and spleen on the right side of the abdomen and the liver and gall bladder on the left side. The left lung is trilobed and the right lung bilobed, and blood vessels, nerves, lymphatics and the intestines are also transposed.

If the heart is swapped to the right side of the thorax, it is known as situs inversus with dextrocardia or situs inversus totalis. If the heart remains on the normal left side of the thorax, a much rarer condition (1 in 22,000 of the general population), it is known as situs inversus with levocardia or situs inversus incompletus. Situs inversus with levocardia, or dextrocardia without situs inversus, present much higher rates of congenital defects than situs inversus with dextrocardia.

True Dextrocardia with situs inversus

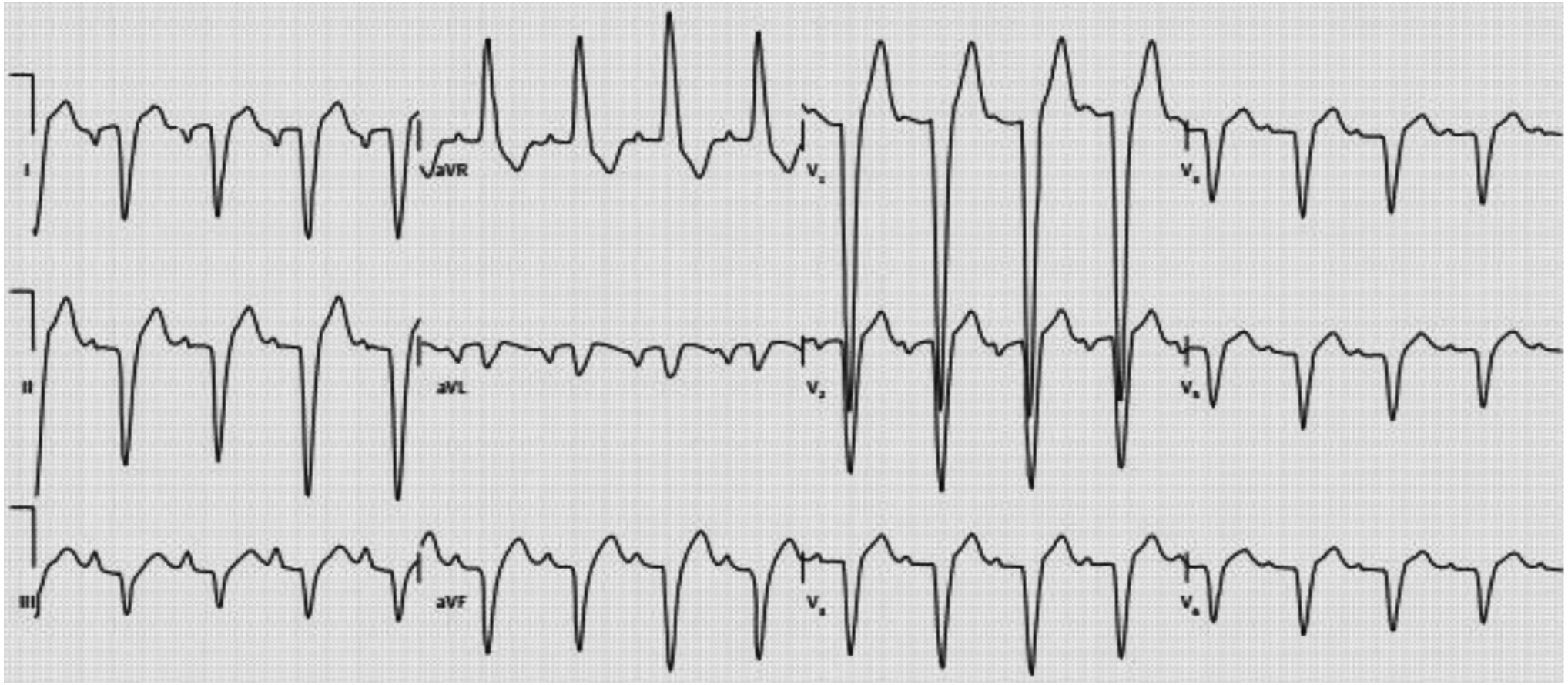


Situs inversus is a condition in which the organs of the chest and abdomen are arranged in a perfect mirror image reversal of the normal positioning.

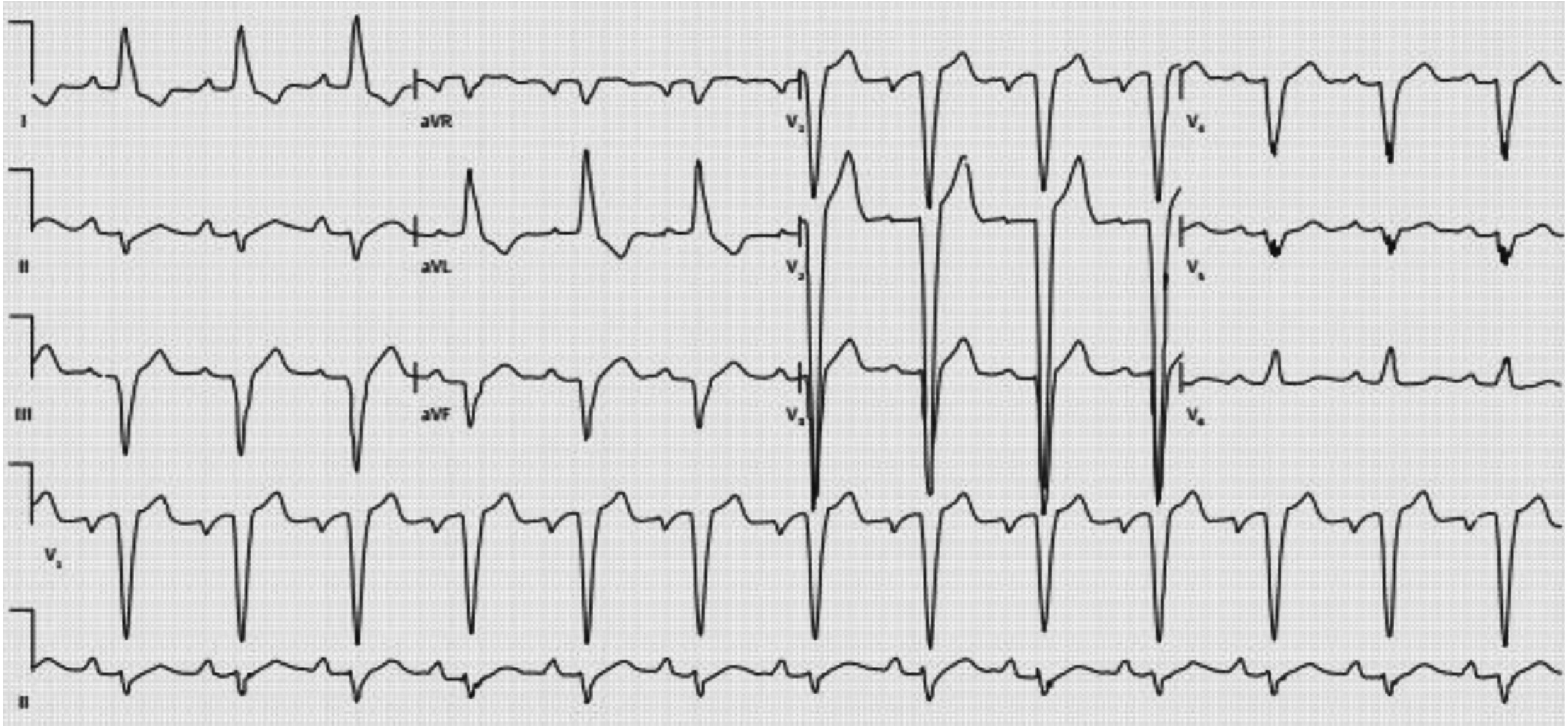
It is believed that dextrocardia occurs in approximately 1 in 100 people, while 1 in 1,000 of these have situs inversus. Totalis occurs in approximately 1 in 5,000 dextrocardias with situs inversus.

An 87-year-old woman with a history of scleroderma treated with methotrexate presented with respiratory distress. An ECG was recorded at admission(**ECG1**). There were no prior ECGs available for comparison. She denied any previous cardiac history. On examination she was visibly dyspneic; BP: 104/48 mm Hg; pulse, 106 bpm; and a room air oxygen saturation of 65% that quickly improved with nasal cannula oxygenation. Pulmonary examination revealed bilateral rales, and cardiac auscultation revealed distant heart sounds along the left precordium. The remainder of the examination findings were unremarkable. **Question:** What is the **ECG-1** diagnosis in this patient?

ECG-1



ECG-1: HR 102bpm, negative P wave in I, prolonged PR interval (PR 220ms), first-degree atrioventricular, QRS negative in lead I, aVL and aVR will appear to be reversed: tall R wave in aVR. Regression of R-wave amplitudes across the precordial leads with QS in V5-V6. The defining characteristics of a LBBB are: QRS duration ≥120 ms, broad and notched or slurred R waves in left leads I, aVL, V5, and V6 as well as absent Q waves in leads V5 and V6. The ECG presented is consistent with a patient with dextrocardia and a LBBB pattern.



ECG performed in patient with dextrocardia following intentional limb lead reversal and placement of the precordial leads on the right chest. Sinus tachycardia and LBBB (broad and notched or slurred R wave in the leads I, aVL, V5, and V6). This ECG appearance is hidden by the presence of dextrocardia when the ECG is performed with precordial leads in the standard position. Scleroderma has several cardiac manifestations, including myofibrosis and coronary vasospasm, that can induce ECG abnormalities such as non-specific ventricular conduction disturbance or LBBB (**Parker 2013**).

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