Second "Dr. Cosme Argerich" Internatiotal Symposium of Cardiology on Internet

Value of Electrocardiogram and Vectorcardiogram in Brugada Syndrome Diagnosis

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The Brugada syndrome is a electrocardiographic clinical genetic autosomal dominant entity characterized by typical electrocardiogram findings and an increased risk of sudden cardiac death in a patient without apparent structural heart disease. It is the most common cause of sudden death in young men without known underlying cardiac disease in Thailand and Laos.

Although the ECG findings of Brugada syndrome were first reported among survivors of cardiac arrest in 1989, it was only in 1992 that the Brugada brothers recognized it as a distinct clinical entity, causing sudden death by ventricular fibrillation in the heart. The entity is endemic in Southeast Asia.

Thailand and Laos ≈ 1:1.000

26 to 36 death by 100,000 hab/year1.

Southeast Asia – Prevalence (1/2500).

4% to 20% of all SCD without structural cardiopathy2.

14/1,000 among Japanese population3.

5/1,000 among caucasian.

1) Nademanee KK, et.al. Circulation. 1997; 96:2595-2600
2) Antzelevitch C, et al. Circulation. 2005; 10.1161/01
3) Napolitano C, et al. **Orphanet J Rare Dis. 2006;1(1) 35**

MAIN ECG FEATURES IN BrS

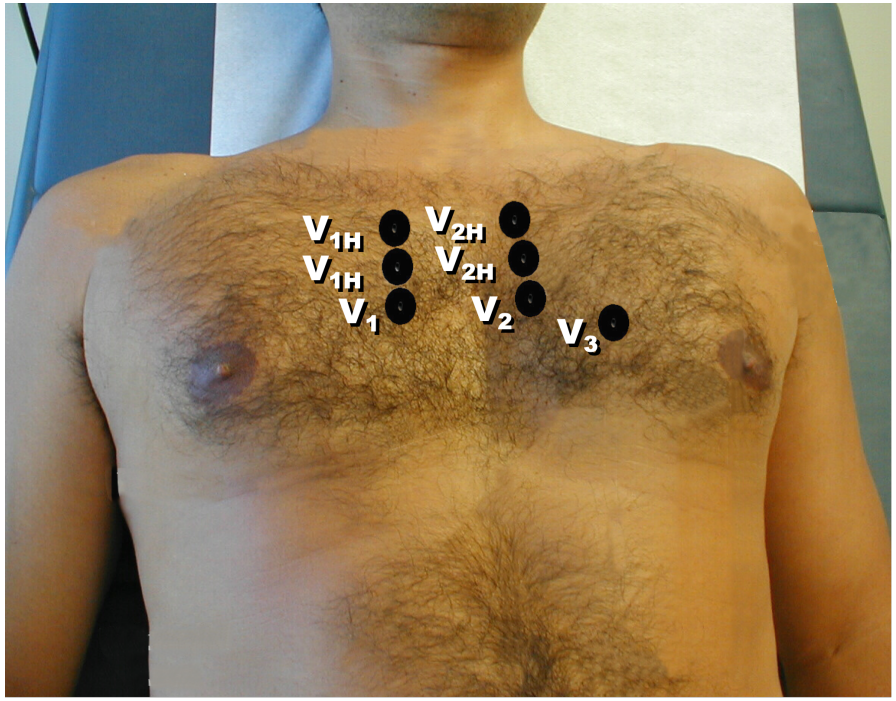
1) RHYTHM: Sinus rhythm is the usual; however, supraventricular arrhythmias are mentioned in 10% to 25% of the cases since the arrhythmogenic substrate is not just limited by the ventricles. Atrial fibrillation was mentioned in the initial publication by the Brugada brothers[1], by Brazilian[2] and Japanese[3] authors. The latter pointed out that paroxysmal atrial fibrillation is observed in 30% of the cases. Eckardt et al[4] found supraventricular arrhythmias in 29% of the cases, describing association with atrioventricular junctional rhythms episodes, and Wolff-Parkinson-White[5] syndrome. The incidence of past history of AF is significantly higher in the symptomatic patients than in the asymptomatic. Prolonged QRS duration in precordial leads is prominent in symptomatic patients. This ECG marker may be useful for distinguishing high- from low-risk patients with BrS[6].

2)HEART RATE: The function of the sinus node is attenuated in patients with PES-induced VF. In this patients corrected sinus node recovery time and sino-atrial conduction time are prolonged[7]. There is a reference of association with sick sinus syndrome after pacemaker implantation in a patient with sick sinus syndrome[8].

1) Brugada P, Brugada J. Right bundle branch block, persistent ST segment elevation and sudden cardiac death: A distinct clinical and electrocardiographic syndrome. J Am Coll Cardiol 1992, 20: 1391-1396.
2) Villacorta H, Faig Torres RA, Simões de Castro IR, Lambert H. de Araujo Gonzáles Alonso R.: Morte súbita em paciente com bloqueio de ramo direito e elevação persistente do segmento ST. Arq Bras Cardiol. 1996; 66: 229-231.
3) Itoh H, Shimizu M, Ino H, Okeie K, Yamaguchi M, Fujino N, Mabuchi H; Hokuriku Brugada Study Group. Arrhythmias in-patients with Brugada-type electrocardiograph findings. Jpn Circ J 2001; 65:483-486.
4) Eckardt L, Kirchhof P, Loh P, et al. Brugada Syndrome and Supraventricular Tachyarrhythmias: A Novel Association? J Cardiovasc Electrophysiol 2001; 12:680-685.
5) Eckardt L, Kirchhof P, Johna R, Haverkamp W, Breithardt G, Borggrefe M. : Wolff-Parkinson-White syndrome associated with Brugada syndrome. Pacing Clin Electrophysiol 2001;24:1423-1424.
6) Takagi M, Yokoyama Y, Aonuma K, Aihara N, Hiraoka M; for the Japan Idiopathic Ventricular Fibrillation Study (J-IVFS) Investigators. Clinical Characteristics and Risk Stratification in Symptomatic and Asymptomatic Patients with Brugada Syndrome: Multicenter Study in Japan. J Cardiovasc Electrophysiol. 2007 Dec;18(12):1244-51.
7) Morita H, Fukushima-Kusano K, Nagase S, Miyaji K, Hiramatsu S, Banba K, Nishii N, Watanabe A, Kakishita M, Takenaka-Morita S, Nakamura K, Saito H, Emori T, Ohe T.Sinus Node Function in Patients With Brugada-Type ECG.Circ J. 2004;68:473-476.
8) Nakazato Y, Suzuki T, Yasuda M, Daida H. Manifestation of brugada syndrome after pacemaker implantation in a patient with sick sinus syndrome. J Cardiovasc Electrophysiol. 2004; 15: 1328-1330.

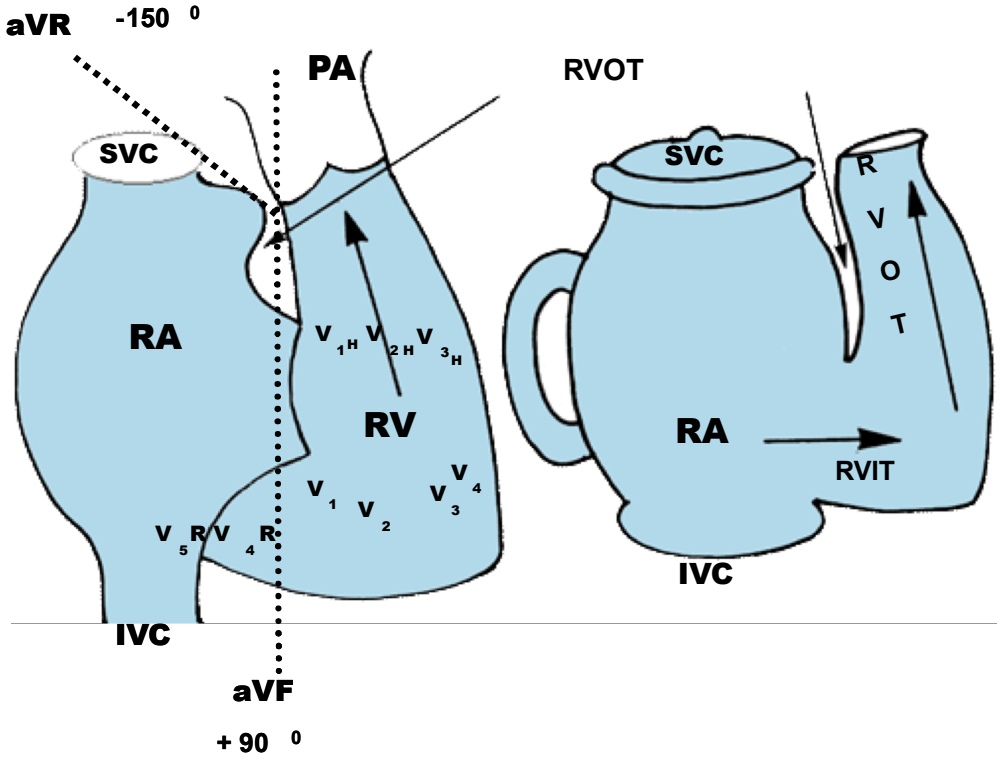
MODIFIED PRECORDIAL LEADS: RIGHT PRECORDIAL LEADS AT HIGHER INTERCOSTAL SPACE POSITIONS: V_{1H}-V_{2H}-V_{3H}

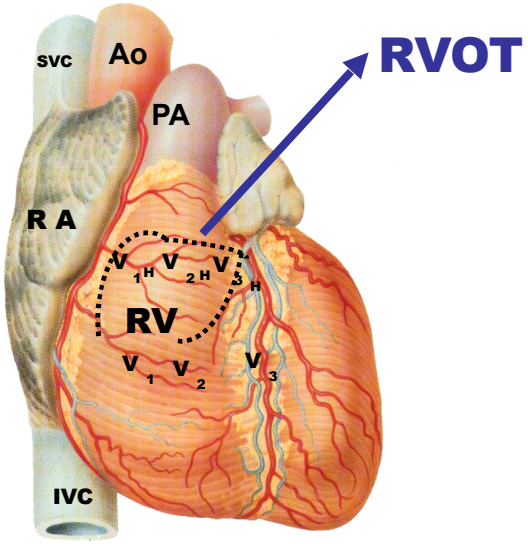
Several works carried out conclude that 12-lead ECG sensitivity increases by applying accessory leads located in the high right precordial area (V_{1H} – V_{2H}), over the 3rd or 2nd intercostal space, just to the right (V_{1H}) or left (V_{2H}) of the sternum. In certain cases, Brugada sign that was not observed using only the 12 conventional leads, is now visualized. The procedure is founded on the fact that modified precordial leads on right precordial leads (V_{1H} - V_{2H}) or on anteroseptal wall (V_{1H} to V_{3H}) at higher intercostal space positions are located exactly opposite to the RVOT



1. Teijeiro R, Garro HA, Acunzo RS, Albino E, Chiale PA. Recording of high V1-V3 precordial leads may be essential to the diagnosis of Brugada syndrome during the ajmaline test. J Cardiovasc Pharmacol Ther. 2006 Jun;11(2):153-5.
2. Márquez MF, Allende R, Cazares-Campos I, Cárdenas M. [Utility of high parasternal electrocardiographic leads in the diagnosis of Brugada syndrome] Arch Cardiol Mex. 2009 Dec;79 Suppl 2:40-3.

REGIONS OF RV AND CORRESPONDING LEADS





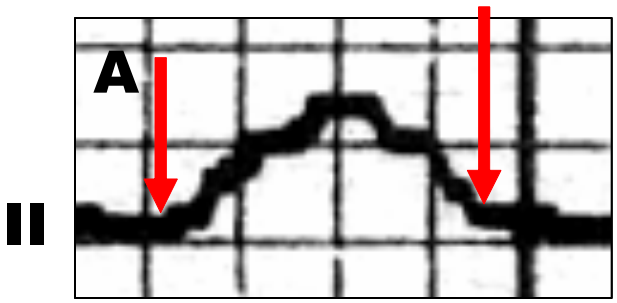
RIGHT VENTRICLE REGIONS AND THEIR CORRESPONDING LEADS
The right ventricle has five regions that are better detected by the following leads:

1. V2 and V3.: trabecular area;
2. V3 – V4: Low right paraseptal area;
3. V1 to V4.: Free wall
4. aVF, V4R and V5R: Right Ventricle Inflow Tract (RVIT)

- **RVOT: the area affected in the Brugada syndrome. aVR, V_{1H}, V_{2H}, V_{3H}**

3) P WAVES

P WAVE BEFORE AJMALINE INJECTION



P WAVE AFTER AJMALINE INJECTION

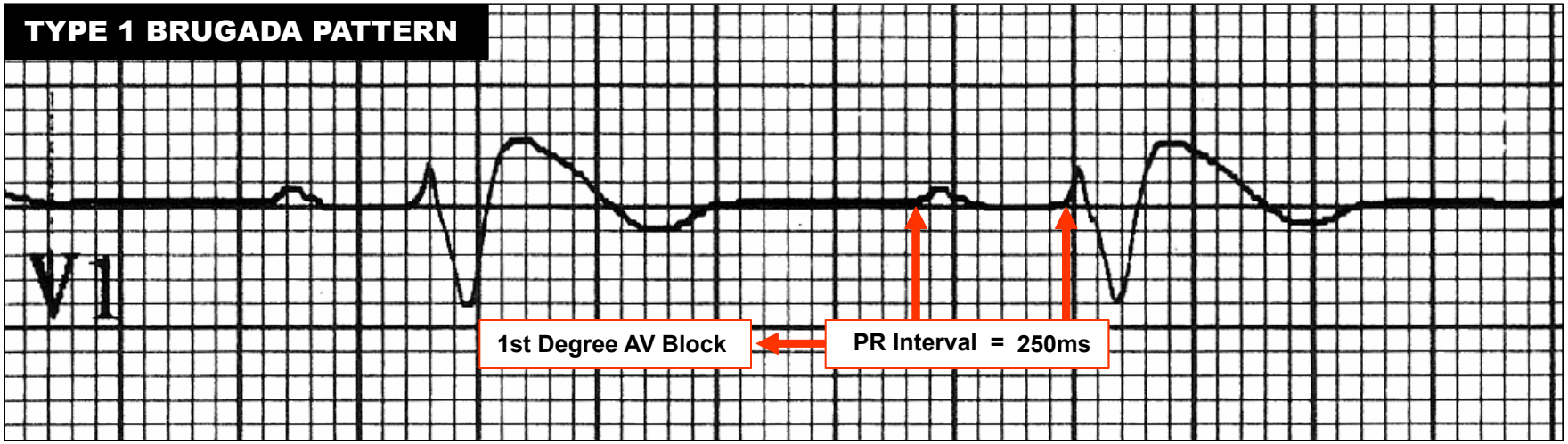
Commentaries: discrete P wave prolongation is observed in Brugada Syndrome (BrS) patients with positive SCN5A mutation, mainly only after ajmaline injection (A & B).

P wave duration prolongation together with PR and QRS duration prolongation and depolarization abnormalities. Discrete P wave prolongation is frequently observed in BrS patients with positive SCN5A mutation. P (max), P-wave dispersion (P(disp)), and left atrial dimensions are not significantly different among BrS patients and controls as predictor of atrial fibrillation (AF).

1. Yokokawa M, Noda T, Okamura H, Satomi K, Suyama K, Kurita T, Aihara N, Kamakura S, Shimizu W. Comparison of long-term follow-up of electrocardiographic features in Brugada syndrome between the SCN5A-positive probands and the SCN5A-negative probands. Am J Cardiol. 2007 Aug 15;100(4):649-655.

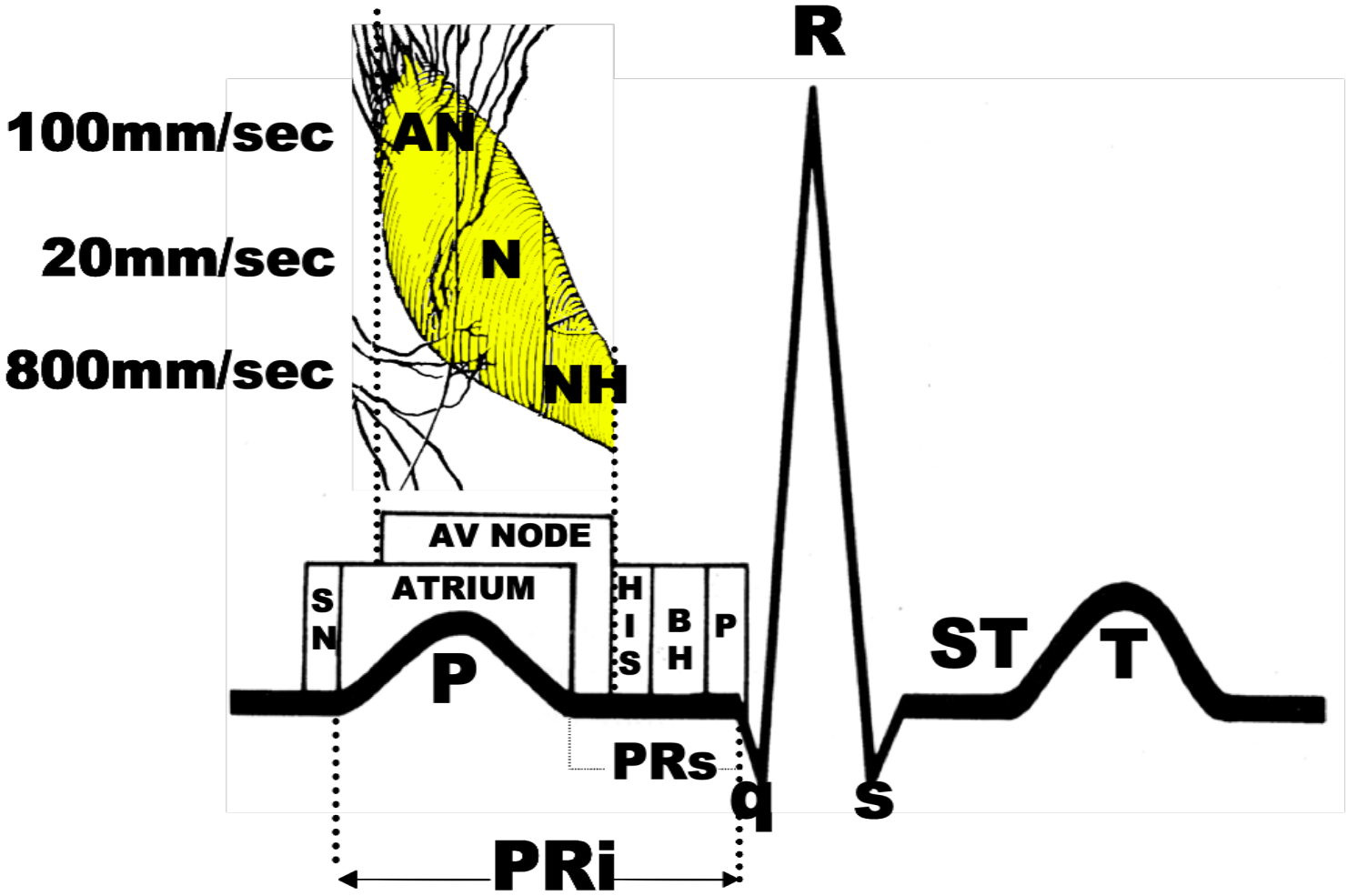
4) PR INTERVAL

First Degree AV Block



Commentaries: First degree AV block is observed in ≈ 50% of cases of BrS. Mainly in the presence of SCN5A mutation.

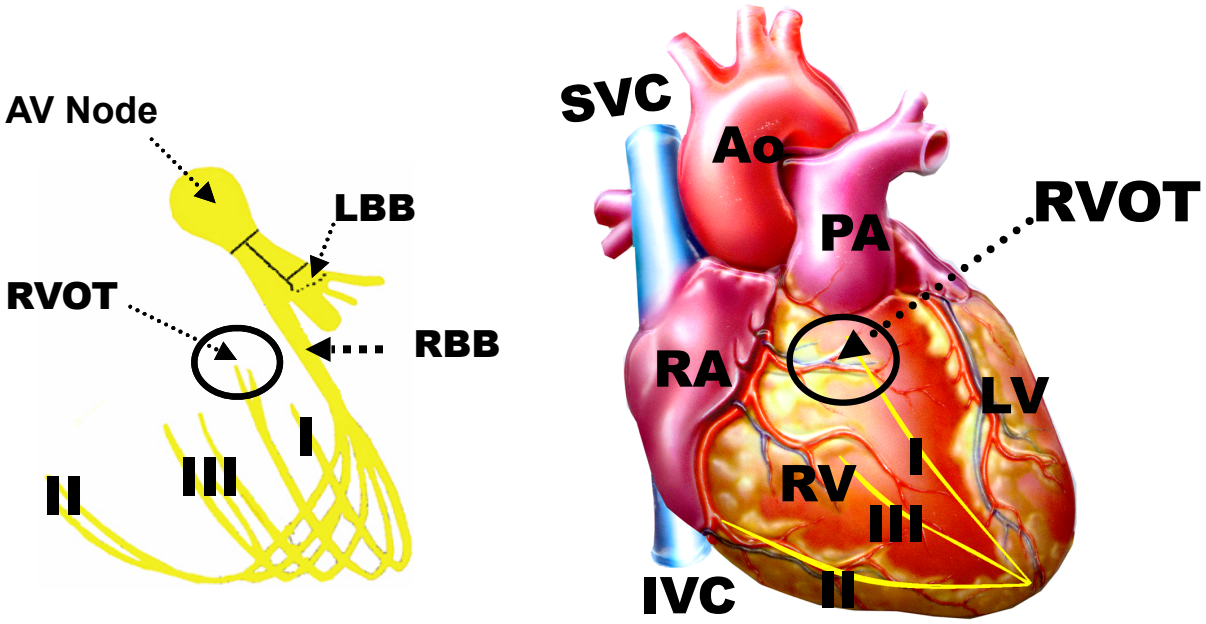
PR Interval Components and affectation in BrS



In BrS, PR interval could be prolonged as consequence of prolonged HV interval or by the existence of intra-His or infra-His block.

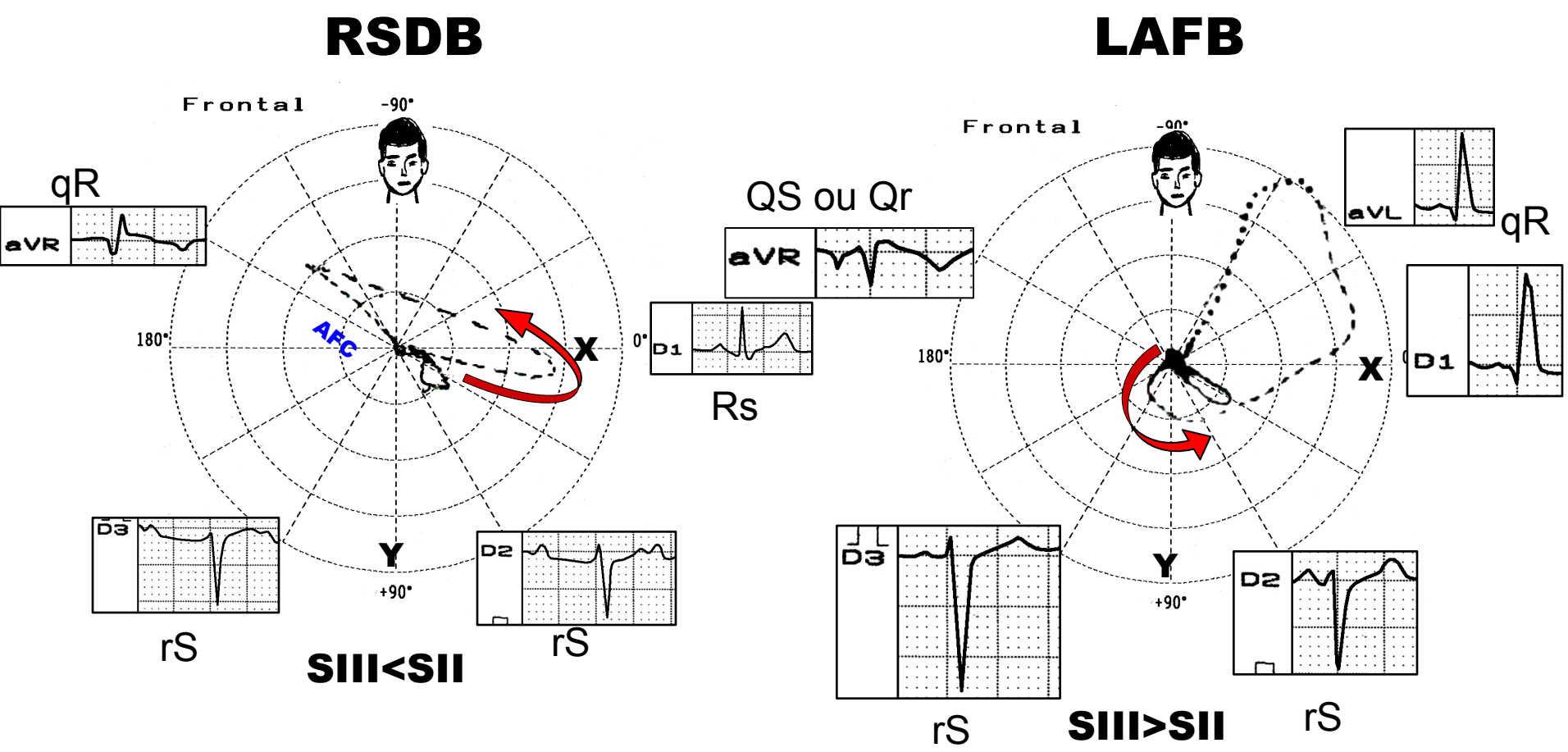
5) QRS AXIS ON FRONTAL PLANE

In BrS the QRS axis on the frontal plane is included within the normal range in 90.5% of the cases; however, in 9.5% marked left QRS axis deviation is observed according to a prospective three-year follow-up study between a population of workers in the Tokyo area[1]. It is interesting to highlight that marked left QRS axis deviation could be the consequence of a Left Anterior Fascicular Block (LAFB) or Right Superior Divisional Block (RSDB). This set of fibers of the right bundle division in the free wall of the right ventricle, is situated within the ROFT; a location electrophysiologically affected in Brugada syndrome.



1. Atarashi H, Ogawa S, Harumi K, et al. Idiopathic Ventricular Fibrillation Investigators. Three-year follow-up of patients with right bundle branch block and ST segment elevation in the right precordial leads: Japanese Registry of Brugada Syndrome. Idiopathic Ventricular Fibrillation Investigators. J Am Coll Cardiol 2001; 37:1916-1920.

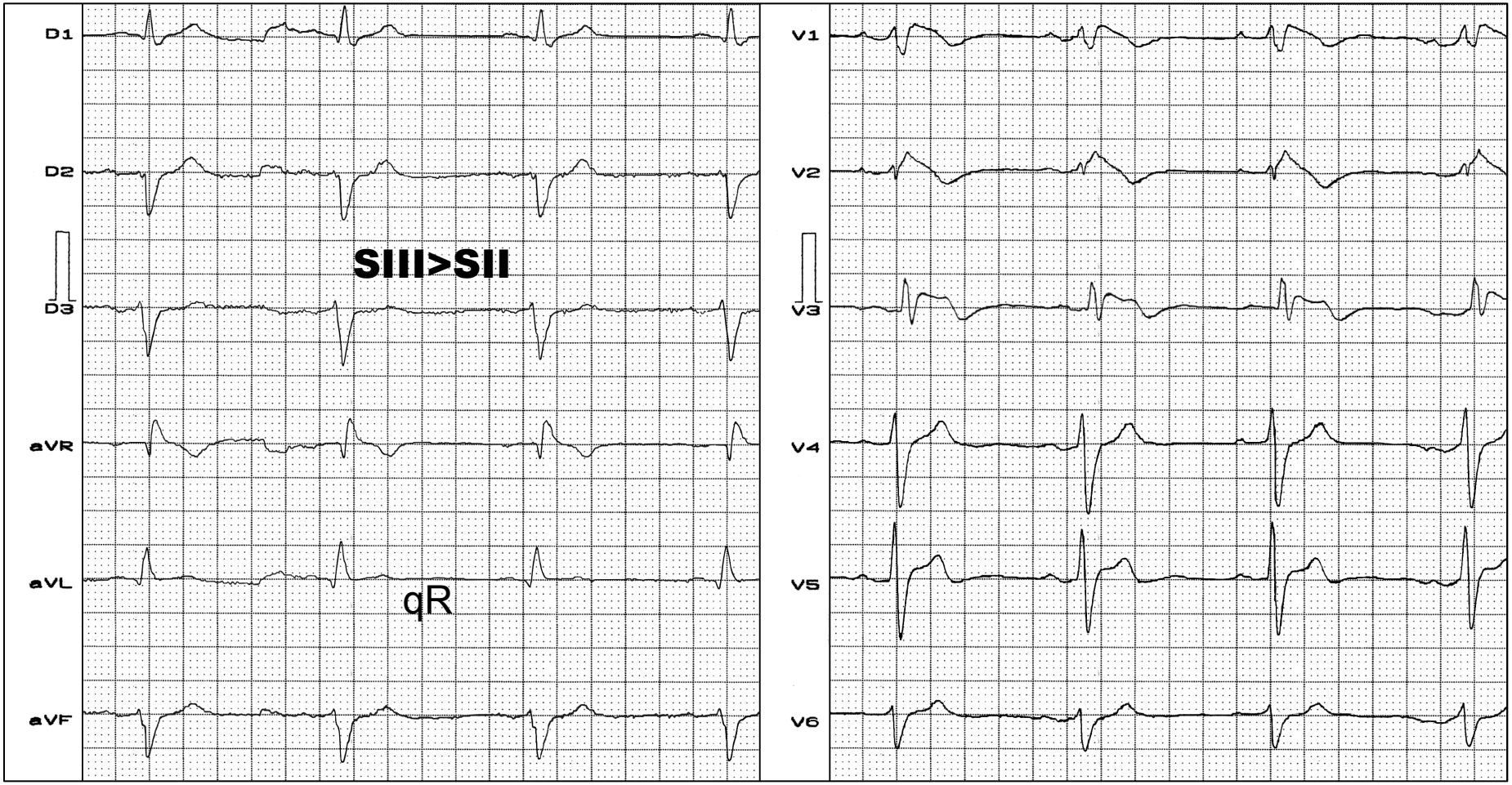
ECG/VCG differential diagnosis between RSDB and LAFB



Both dromotropic disorders could be observed in BrS. LAFB has worst prognosis.

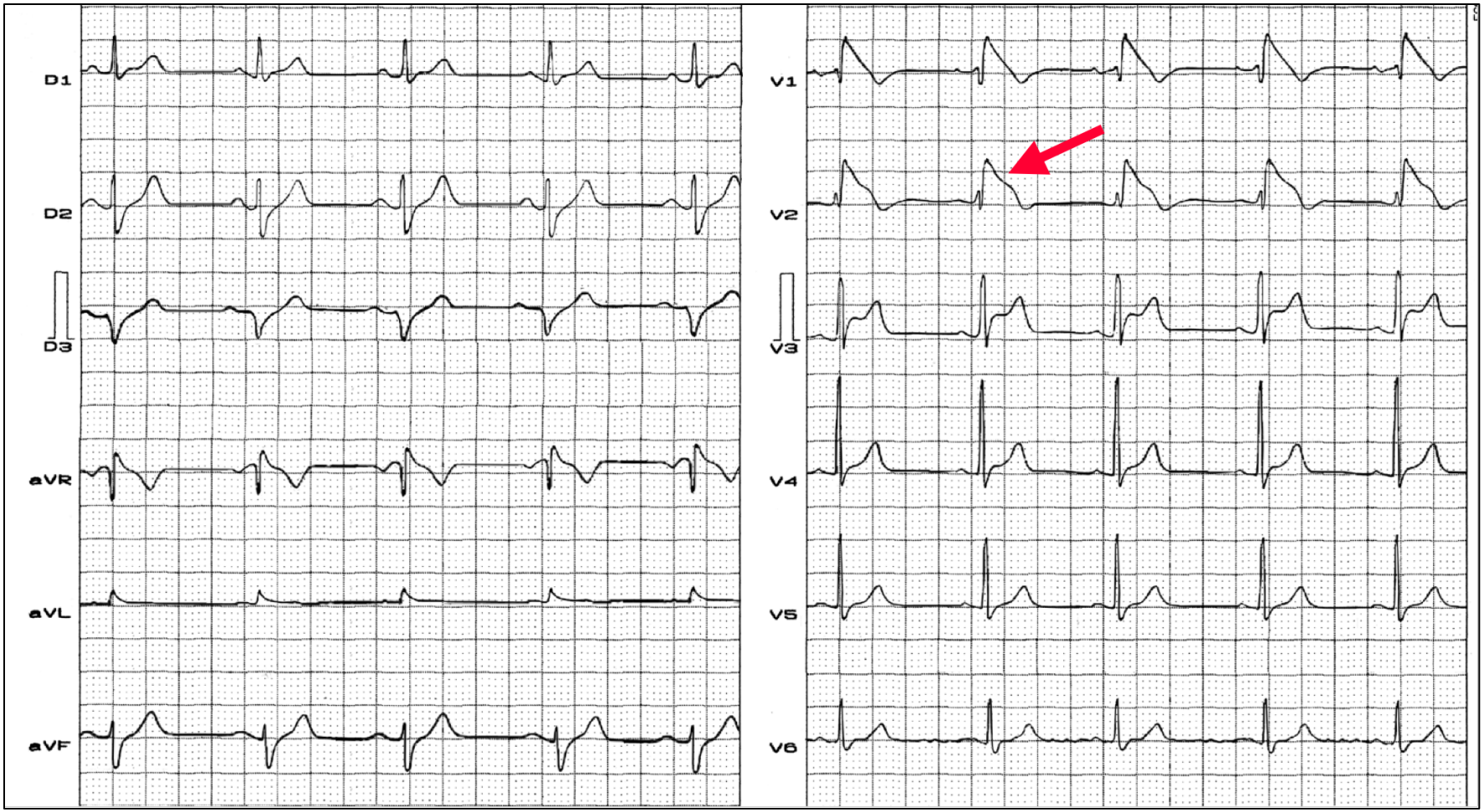
RSDB: Right Superior Divisional Block
LAFB: Left Anterior Fascicular Block

TYPICAL CASE OF BrS TYPE 1 WITH EXTREME LEFT AXIS DEVIATION SECONDARY TO LAFB



LAFB: Left Anterior Fascicular Block

TYPICAL CASE OF BrS TYPE 1 WITH EXTREME LEFT AXIS DEVIATION SECONDARY TO RSDB



RSDB: Right Superior Divisional Block

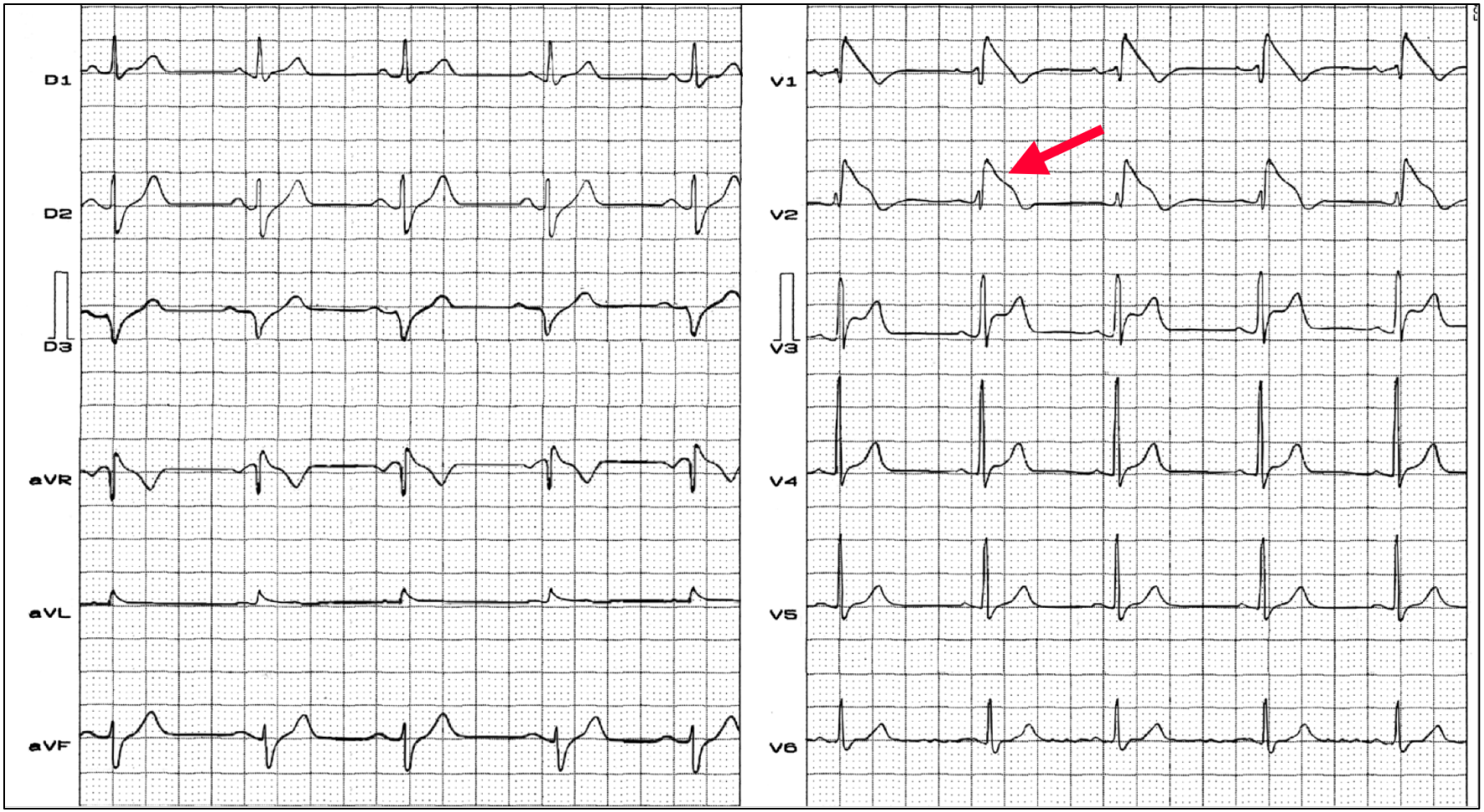
CASE 1
BrS TYPE 1 ECG PATTERN

1. Syncope
2. Positive familiar background of sudden death
3. Genetic research performed: negative

CASE 1
BrS TYPE 1 ECG PATTERN

Identification :

Name: TTP; Age: 56; Gender: Male; Ethnic Group: Asian; Weight: 78kg; Height: 1,70m

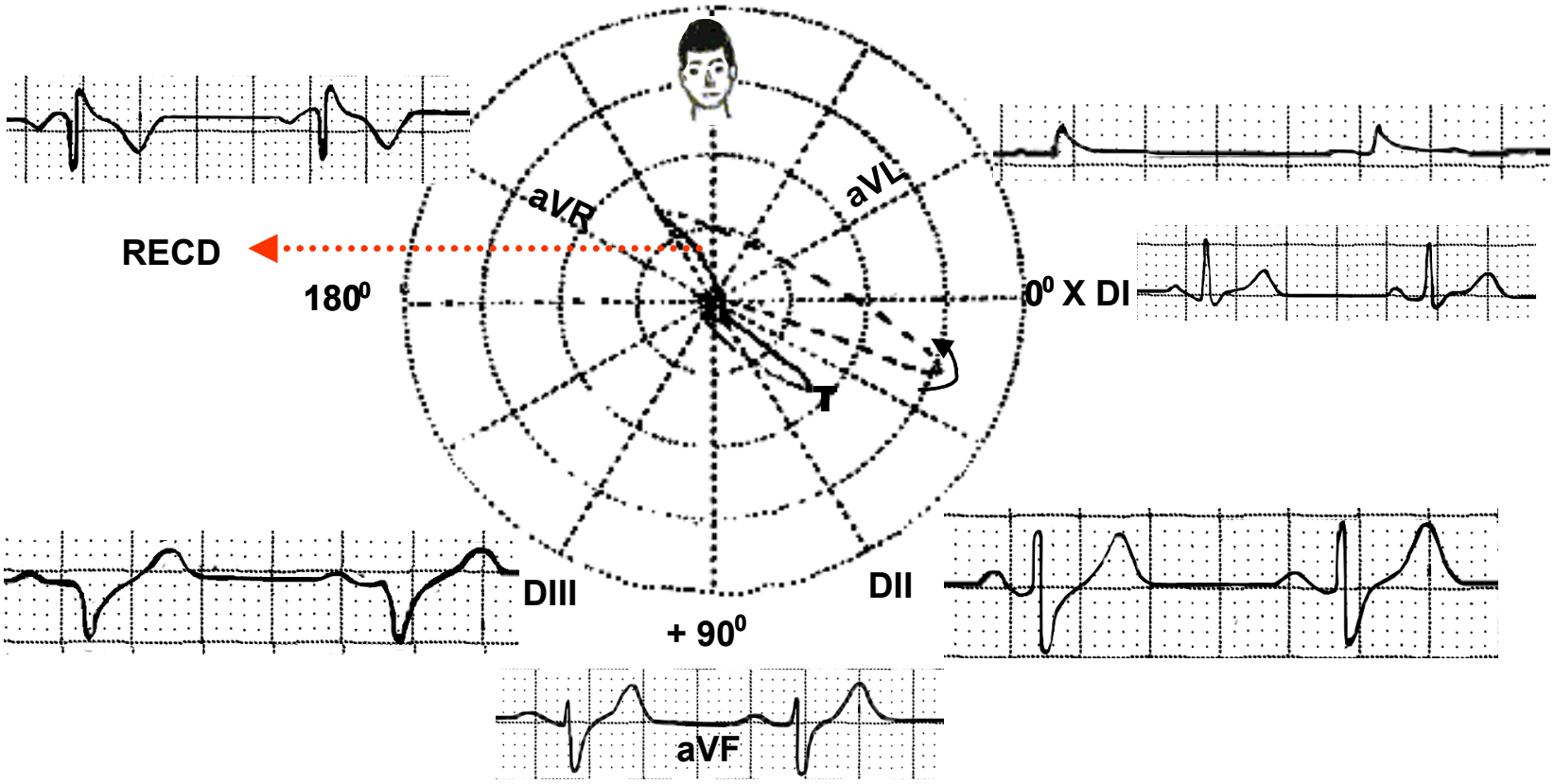


HR: 68bpm P Axis: +60°; PR Interval: 160ms; QRS Axis: -40°; QRS Duration: 100ms; QT/QTc: 370/410ms; T Axis +60°

ECG/VCG CORRELATION FRONTAL PLANE

Identification :

Name: TTP; Age: 56; Gender: Male; Ethnic Group: Asian; Weight: 78kg; Height: 1,70m

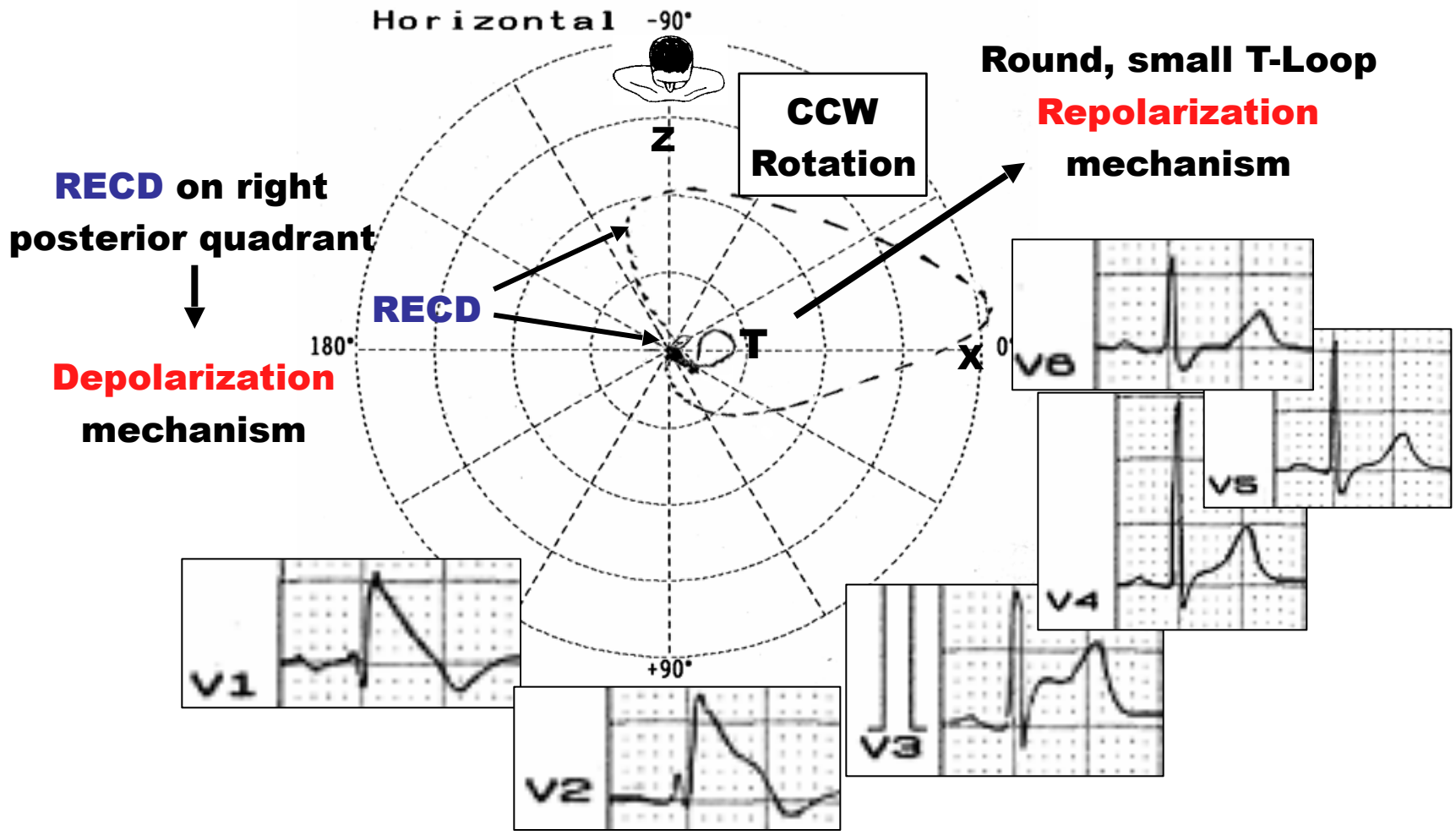


P Axis: +60°; QRS Axis: -40°; T Axis +60°

ECG/VCG CORRELATION HORIZONTAL PLANE

Identification :

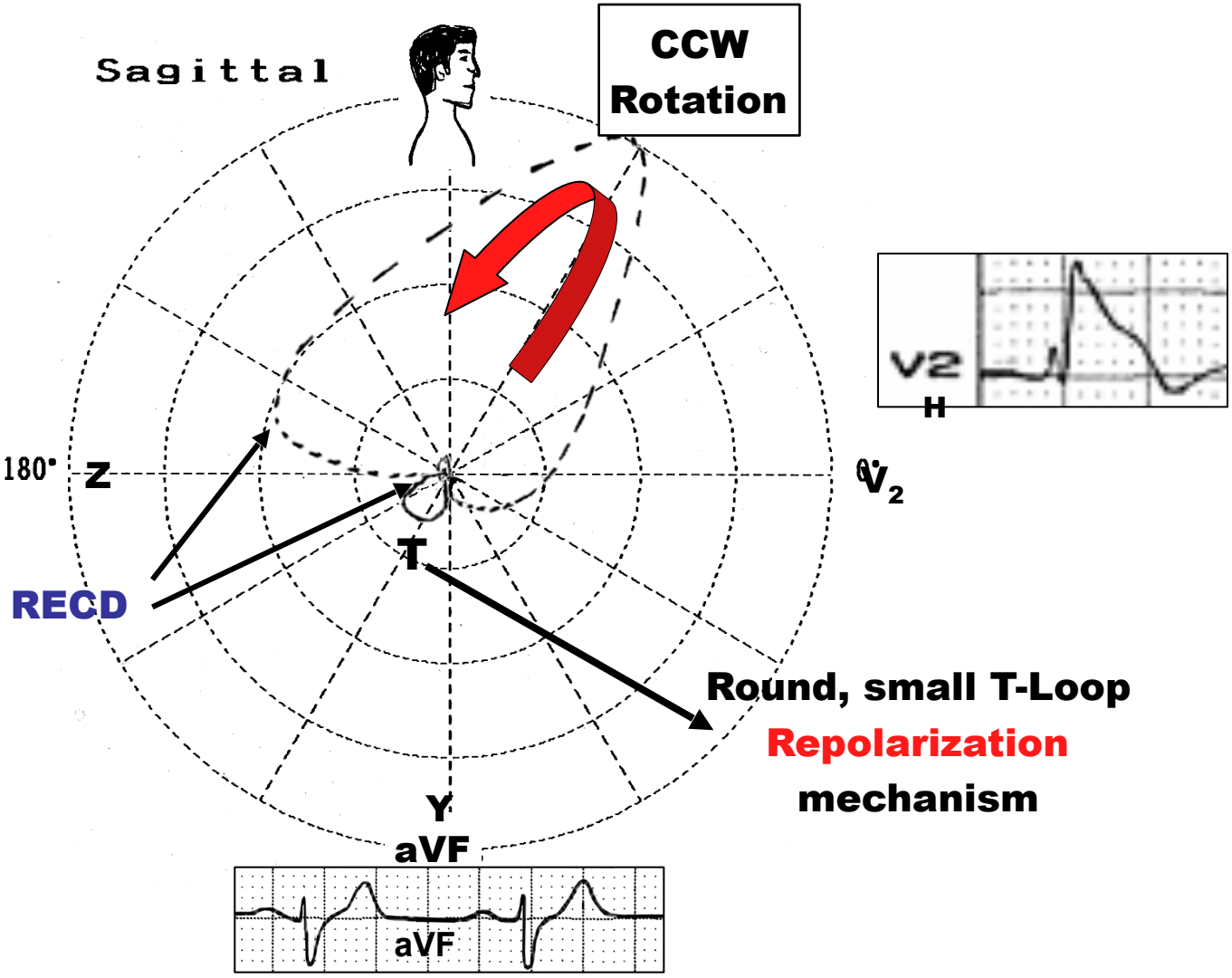
Name: TTP; Age: 56; Gender: Male; Ethnic Group: Asian; Weight: 78kg; Height: 1,70m



ECG/VCG CORRELATION RIGHT SAGGITAL PLANE

Identification :

Name: TTP; Age: 56; Gender: Male; Ethnic Group: Asian; Weight: 78kg; Height: 1,70m



6) REPOLARIZATION PATTERNS IN BrS

DIAGNOSTIC CRITERIA FOR BRUGADA SYNDROME

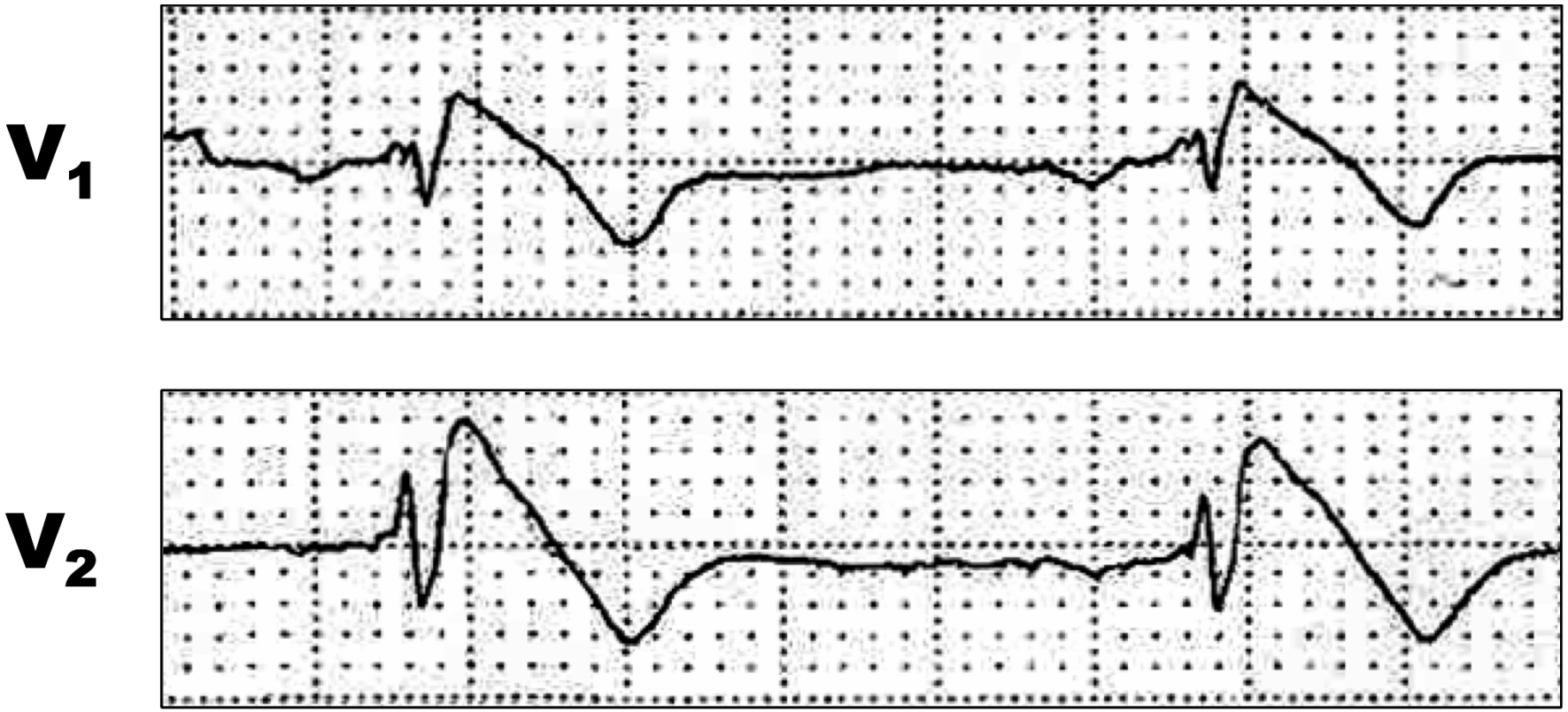
	Type 1	Type 2	Type 3
J point	≥2mm	≥2mm	≥2mm
T wave	negative	positive or biphasic	Positive
ST-T configuration	coved type	saddleback	saddleback
ST segment (terminal portion)	gradually descending	elevated ≥1mm	elevated <1mm

The terminal portion of the ST segment refers to the latter half of the ST segment.

1. Antzelevich C. Chapter 1: Brugada Syndrome: overview, p.1-22 In: The Brugada Syndrome From Bench to Bedside. Edited by Charles Antzelevich with associated editors Pedro Brugada, Josep Brugada and Ramon Brugada. Ed. Blackwell Publishing, 2005.

TYPE 1

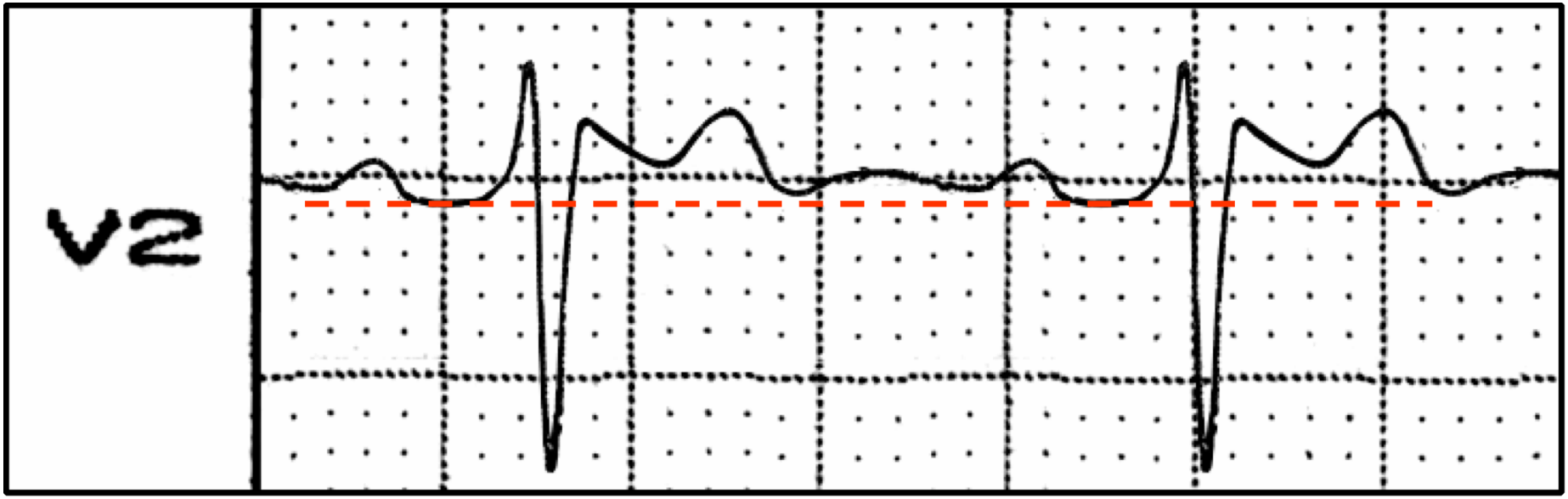
The first European consensus about the syndrome[1] classified the repolarization alterations that occur in right precordial leads (V1 and V2) or in the antero-septal wall (V1 to V3) in three types:



Type 1: ST segment elevation to the top or triangular ("coved type") ≥ 2 mm (0.2 mV), and followed by negative T wave (Brugada phenotype.) There may or may not be CRBBB or IRBBB or ECD. In absence of these dromotropic disorders, broader S waves are not observed in left leads.

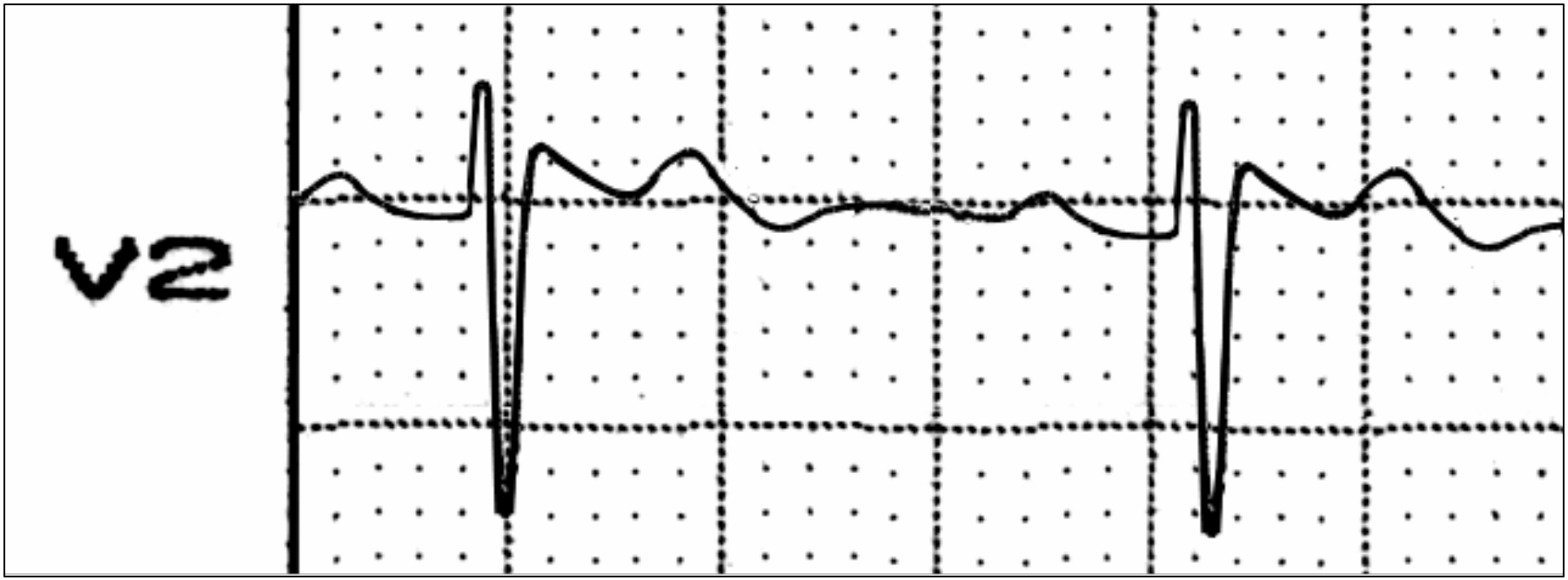
1) Wilde AA, et al. Eur Heart J 2002; 23:1648-1654.

TYPE 2



Type 2: J point and ST segment elevation ≥ 2 mm (0.2 mV) with appearance that resembles a saddleback and remains at least 1 mm above the isoelectric line, followed by positive or biphasic T wave.

TYPE 3



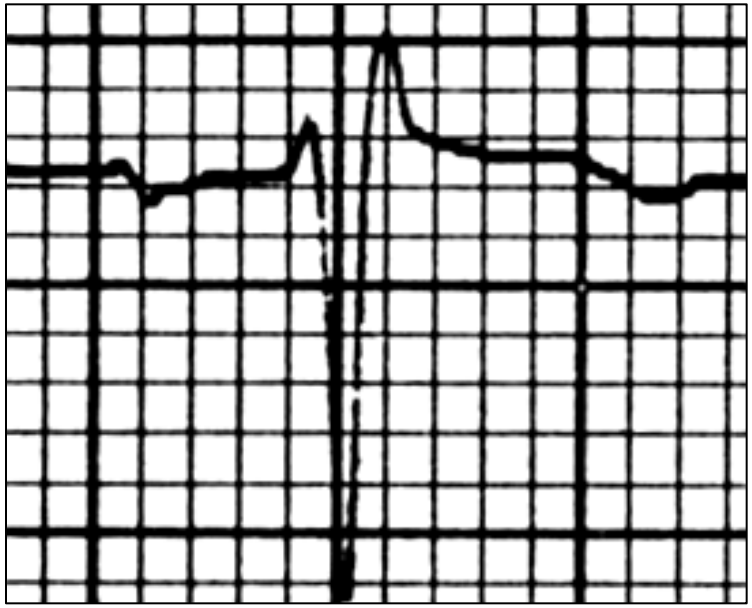
Type 3: J point and ST segment elevation < 1 mm and with variable profile or aspect, or convex to the top or concave. The terminal portion of the ST segment never exceeds 1 mm above the isoelectric line.

TYPE 1 PATTERN AFTER AJMALINE INJECTION (80 mg)

Name: BAAS; **Age:** 53 years old **Race:** Caucasian; **Weight:** 95 Kg **Height:**1,85m.
Sex: Male; **Date:** 06/17/2005.

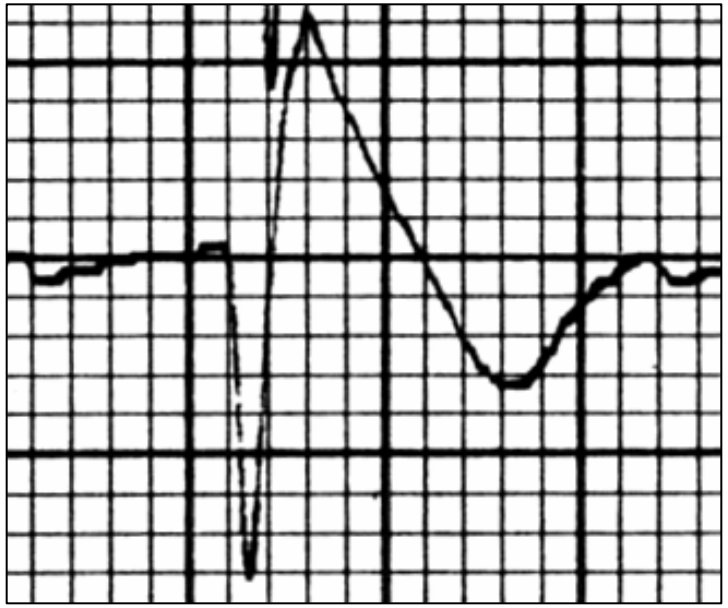
ECG <u>BEFORE</u> AJMALINE INJECTION

V_1



ECG <u>AFTER</u> AJMALINE INJECTION 80 mg

V_1



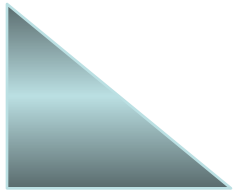
TYPE 1

Positive drug challenge. We observe conversion to Brugada type 1 after ajmaline injection

PROPOSAL OF CLASSIFICATION OF TYPE 1 BRUGADA ECG PATTERN

TYPE 1A: COVED SHAPE OR "BULL TERRIER" WAVE

TYPE 1B: TRIANGULAR SHAPE



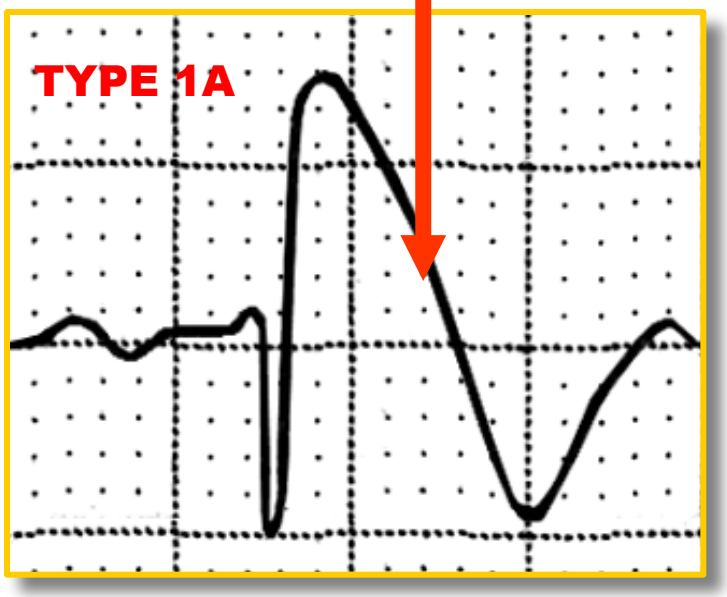
TYPE 1C: "LAMBDA" (λ) OR GUSSAK WAVE

GREEK SMALL LETTER LAMBDA

PROPOSAL OF CLASSIFICATION OF TYPE 1 BRUGADA ECG PATTERN

TYPE 1A

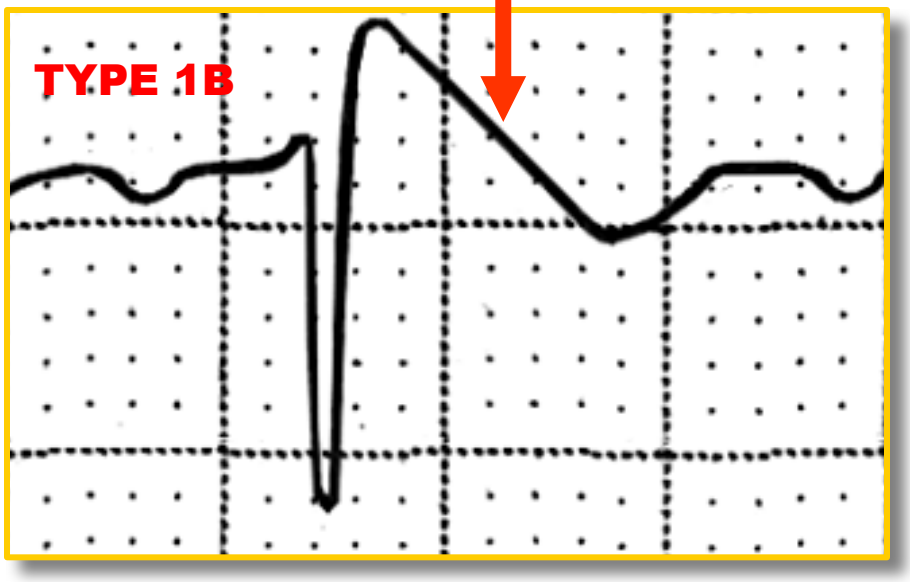
COVED TYPE
(CONVEX TO THE TOP)



V_1 to V_2 or V_3

TYPE 1B

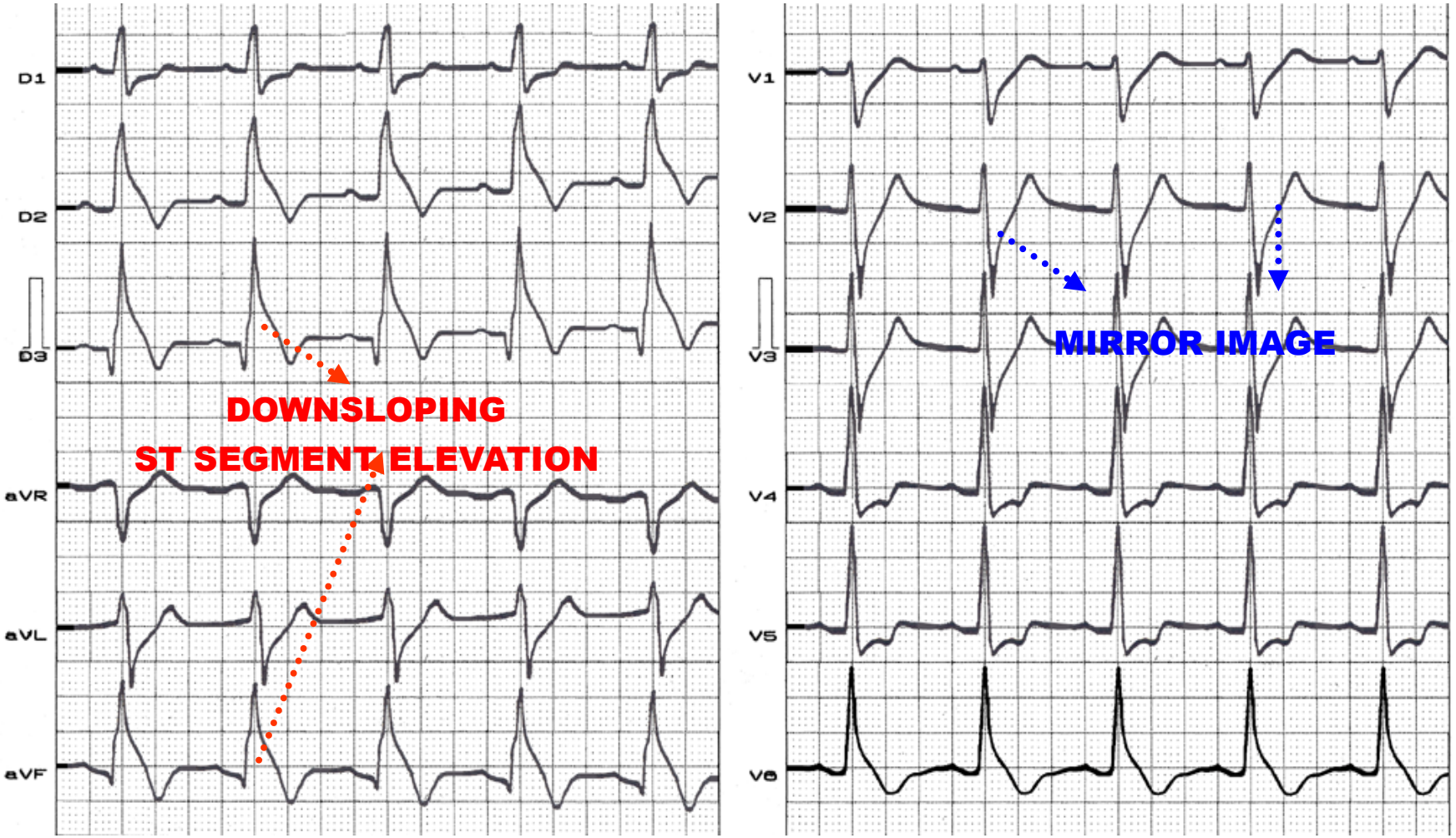
TRIANGULAR SHAPE



V_1 to V_2 or V_3

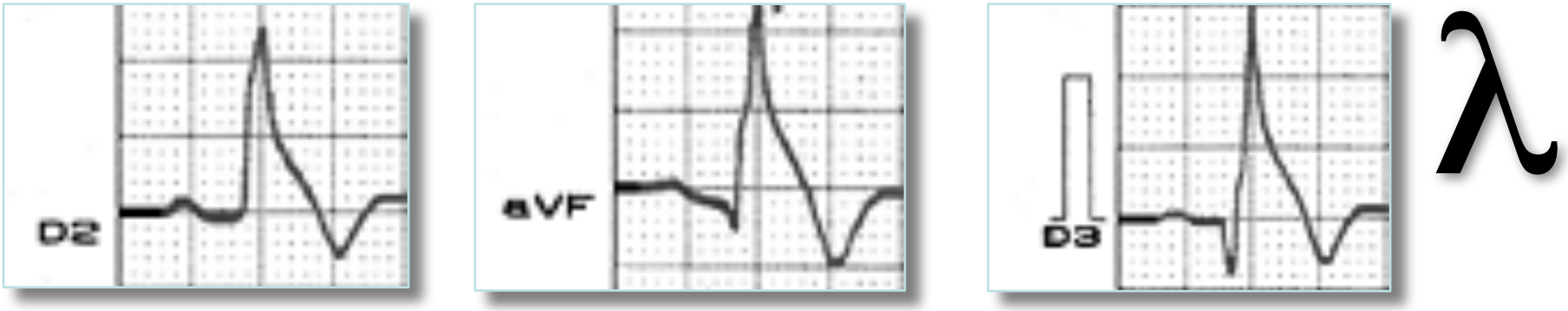
“BULL TERRIER” OR TYPE 1A BRUGADA PATTERN

TYPE 1C BRUGADA ECG PATTERN



Riera AR, et al. J Electrocardiol 2004; 37:101-104.

TYPE 1C WAS DENOMINATED “LAMBDA” WAVE BY GUSSAK I ET AL



Type 1C: ST-segment elevation is triangular or coved to the top (“coved type”) ≥ 2mm (0.2mV), and followed by negative T wave located in inferior leads.

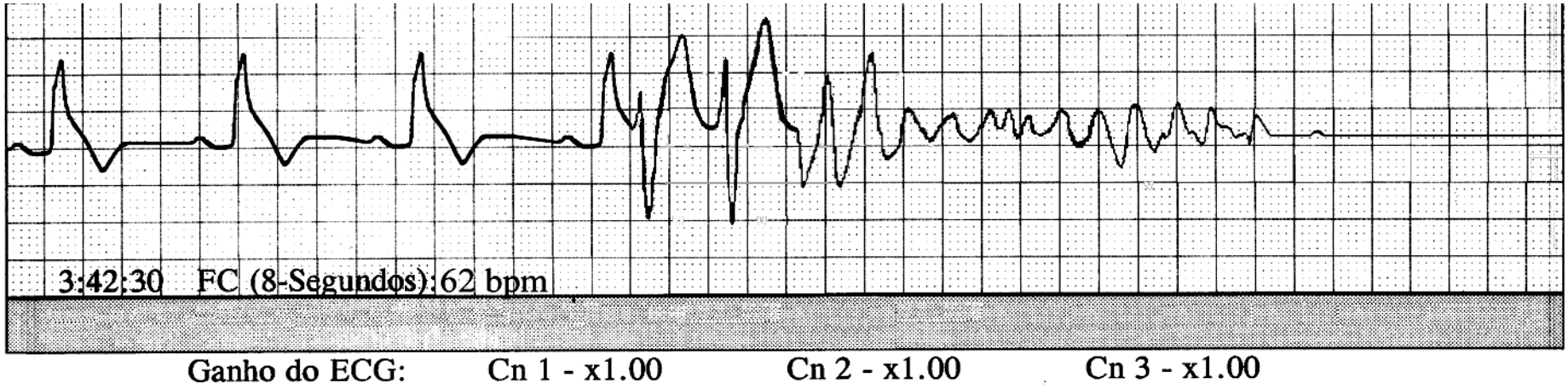
Gussak I, et al. J Electrocardiol 2004;37:105-107.

TYPICAL POLYMORPHIC VT IN ATYPICAL BrS

Name: Y. A. S. **Gender**: Male **Age**: 26 years old. **Ethnic Group:** Yellow **Weight:** 64 Kg. **Height**: 1,68 m
Date: 03/05/2002 **Time**: 3:42:30 AM
Patient Sleeping.

LONG-TERM (HOLTER) ELETROCARDIOGRAPHIC RECORDING

Sudden Cardiac Death by IPVT/IVF with short coupling ending in cardiac arrest.



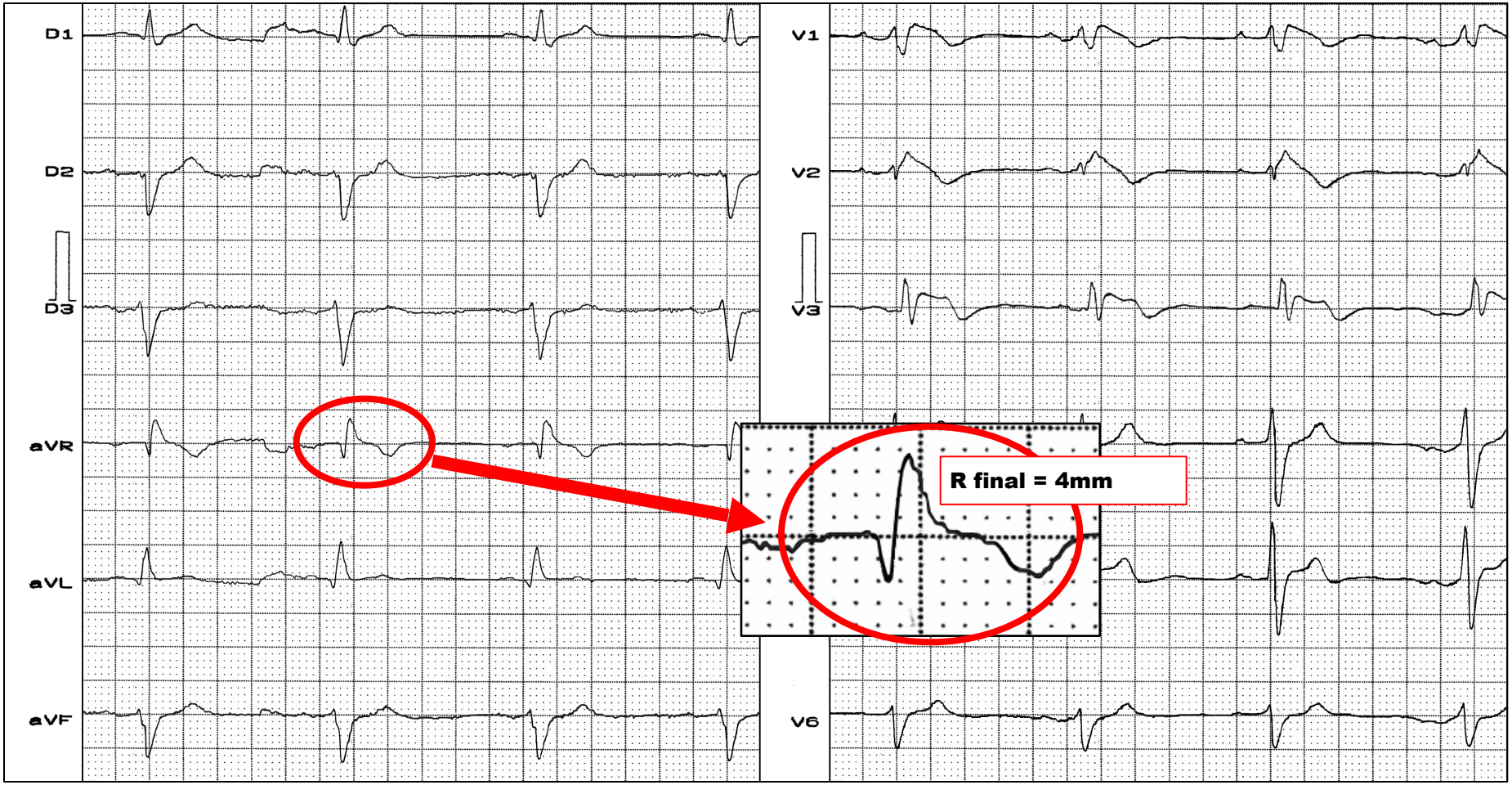
Impresso: WED MARCH 06 18:19:17 2002

Os Dados Devem Ser Revisados Pelo Médico

MAIN DIFFERENCES BETWEEN TRUE POLYMORPHIC VT OBSERVED IN BrS AND TORSADES DE POINTES (TdP)

	Truly PVT	**TdP**
Related to sinus bradycardia	No.	Yes.
Events preceding pauses	No.	Yes.
Heart rate of PVT:	Very fast: 260bpm to 352bpm.	200bpm to 250bpm.
Electrolyte abnormalities	No	Frequently
Coupling of first premature ventricular contractions:	Short. 388 +/- 28msec	Long.
QTc:	Normal.	Long.
U wave:	Normal voltage	Great voltage
Prevalece:	Infrequent	More frequent
Treatment:	ICD + Drugs: Quinidine, cilostazol Electrical storms: isoproterenol, + general anesthesia + cardiopulmonary bypass.	Beta-blockers in high doses, Mexiletine,flecainide, stellectomy, pacing, ICD, and associations.

aVR sign in BrS



There is significant correlation between a prominent R wave in lead aVR (aVR sign) and risk for development of arrhythmic events in BrS. The signal is indicative of right end conduction delay on RVOT.

1. Babaee 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(8):1009-12.

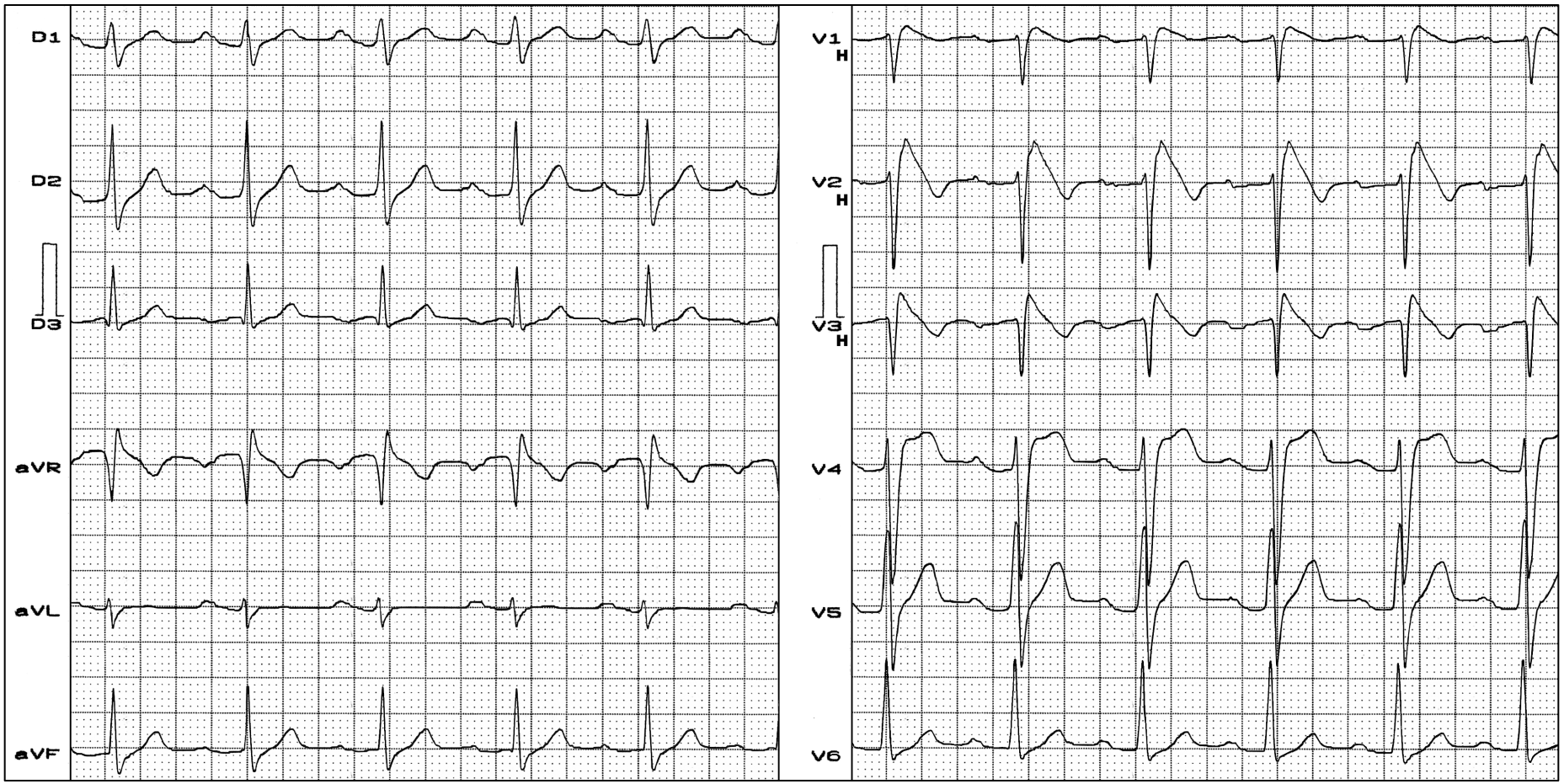
CASE 2
BrS TYPE 1 ECG PATTERN

1. Aborted Sudden Death
2. Positive familiar background of sudden death. Family history of premature sudden death (<35years) in first degree relatives.
3. Genetic research performed: negative

CASE 2
BrS TYPE 1 ECG PATTERN

Identification :

Name: AS Age: 35; Gender: Male; Ethnic Group: Asian; Weight: 72kg; Height 1,71m

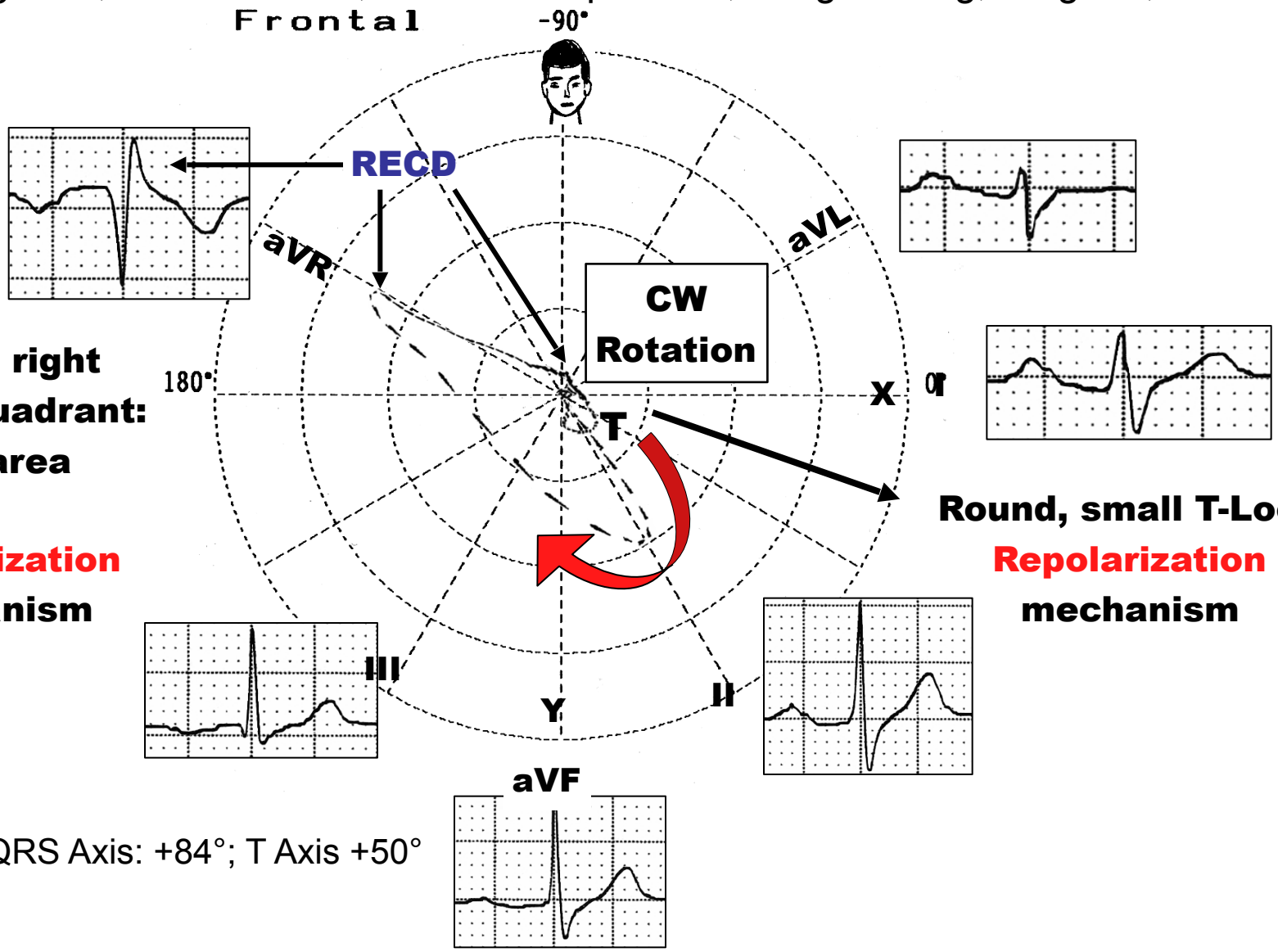


HR: 82bpm P Axis: +33°; PR Interval: 258ms; QRS Axis: +84°; QRS Duration: 112ms; QT/QTc: 360/420ms; T Axis +50°

ECG/VCG CORRELATION FRONTAL PLANE

Identification :

Name: AS Age: 35; Gender: Male; Ethnic Group: Asian; Weight: 72kg; Height 1,71m



RECD on right Superior quadrant: RVOT area

Depolarization mechanism

Round, small T-Loop

Repolarization mechanism

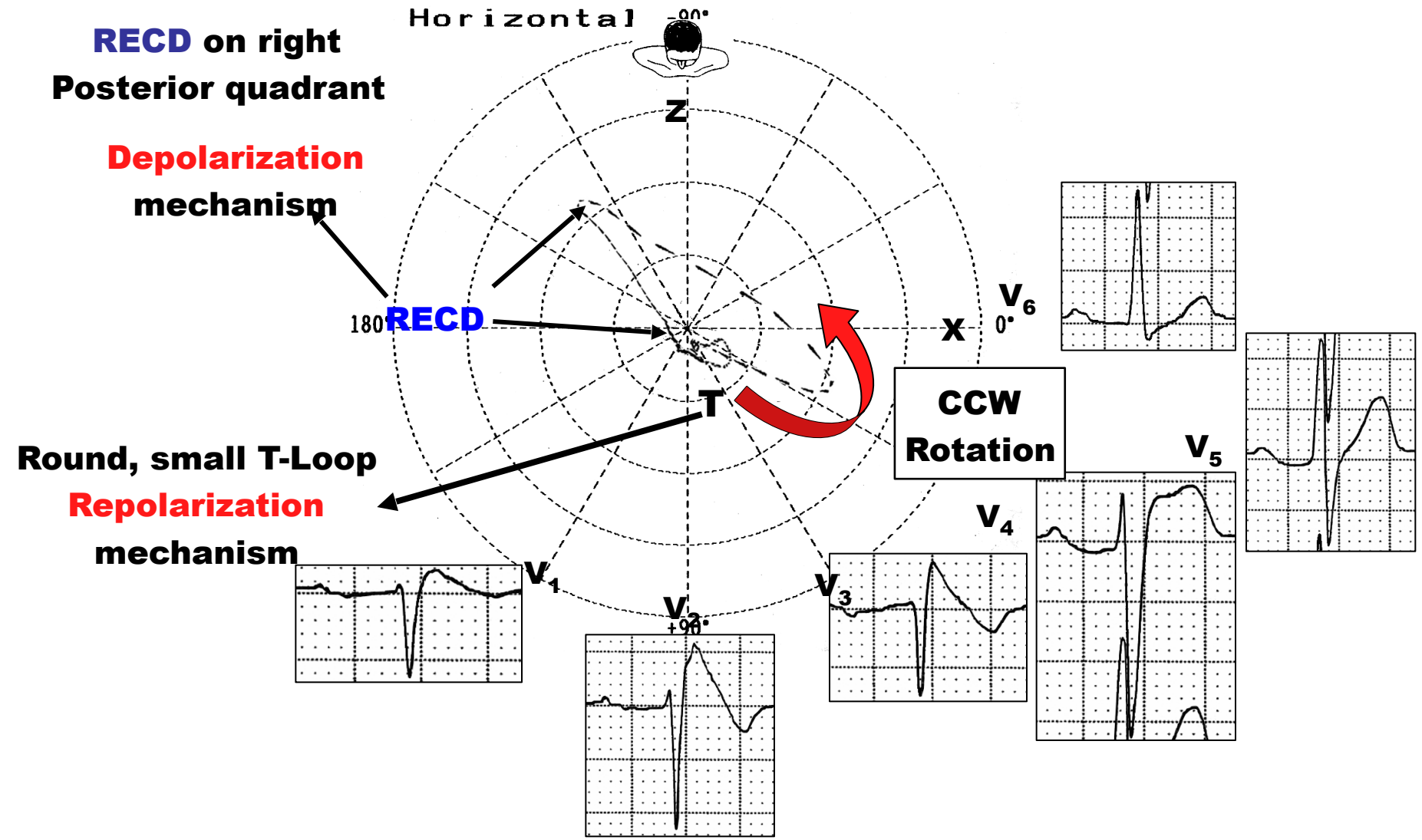
P Axis: +33°; QRS Axis: +84°; T Axis +50°

RECD: Right End Conduction Delay; **CW: Clockwise Rotation.**

ECG/VCG CORRELATION HORIZONTAL PLANE

Identification :

Name: AS Age: 35; Gender: Male; Ethnic Group: Asian; Weight: 72kg; Height 1,71m



ECG/VCG CORRELATION RIGHT SAGGITAL PLANE

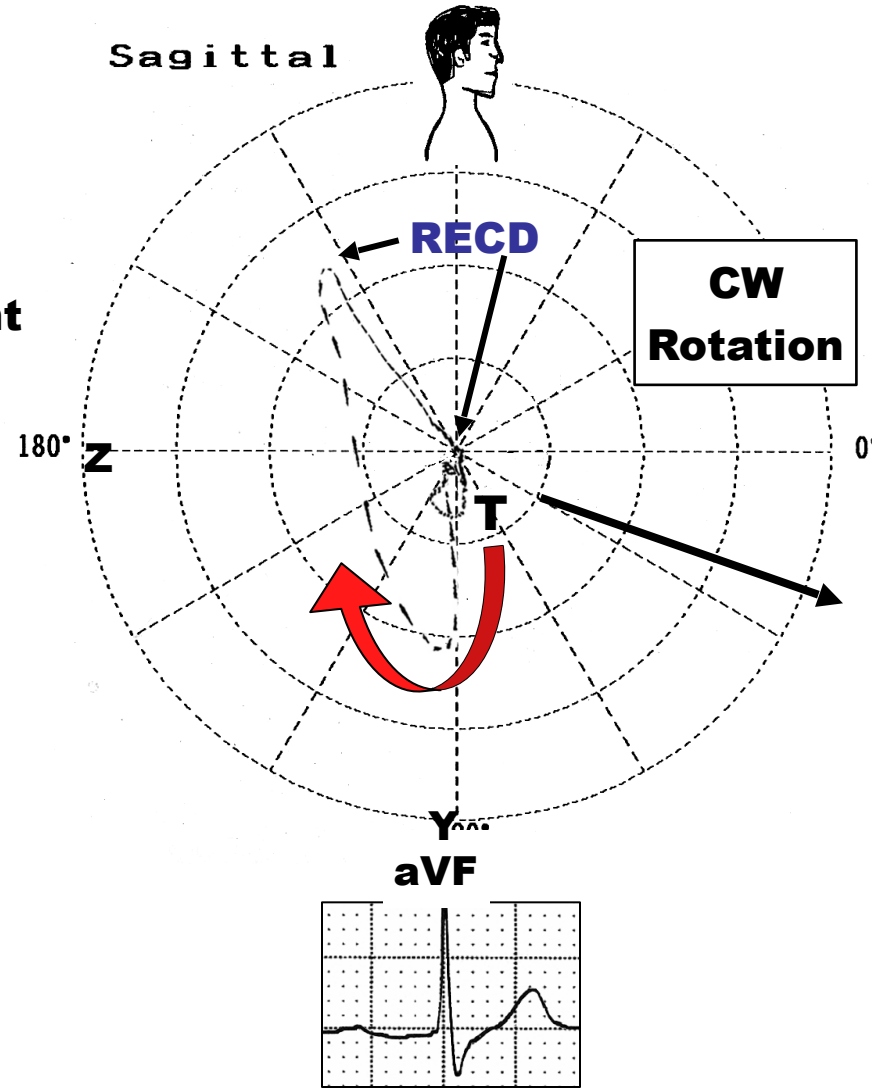
Identification :

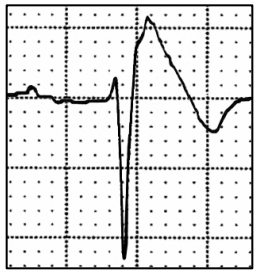
Name: AS Age: 35; Gender: Male; Ethnic Group: Asian; Weight: 72kg; Height 1,71m

RECD on top posterior quadrant

↓

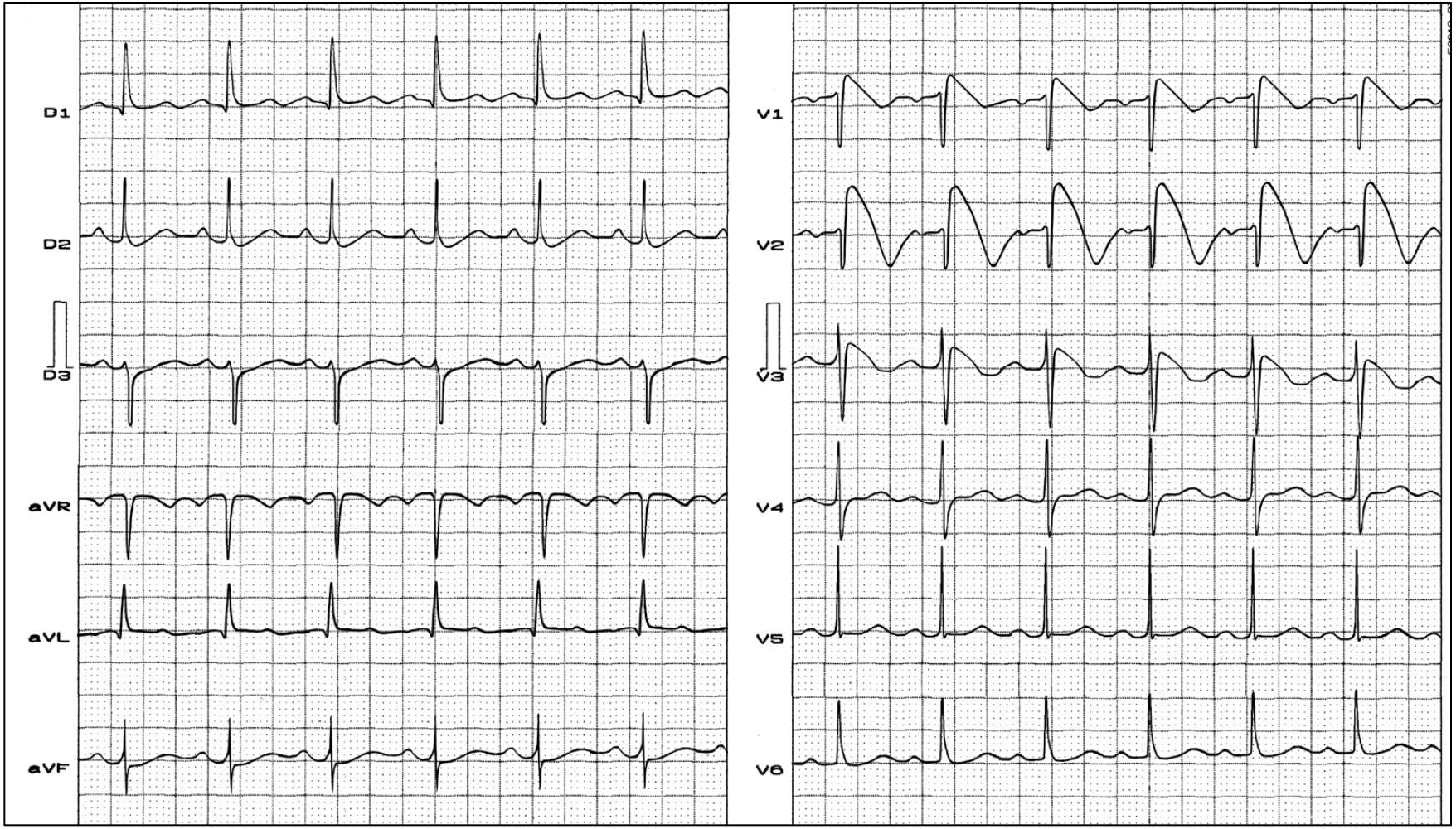
Depolarization mechanism





Round, small T-Loop Repolarization mechanism

ABSENCE OF DROMOTROPIC RIGHT BUNDLE DISORDER IN BrS RBBB

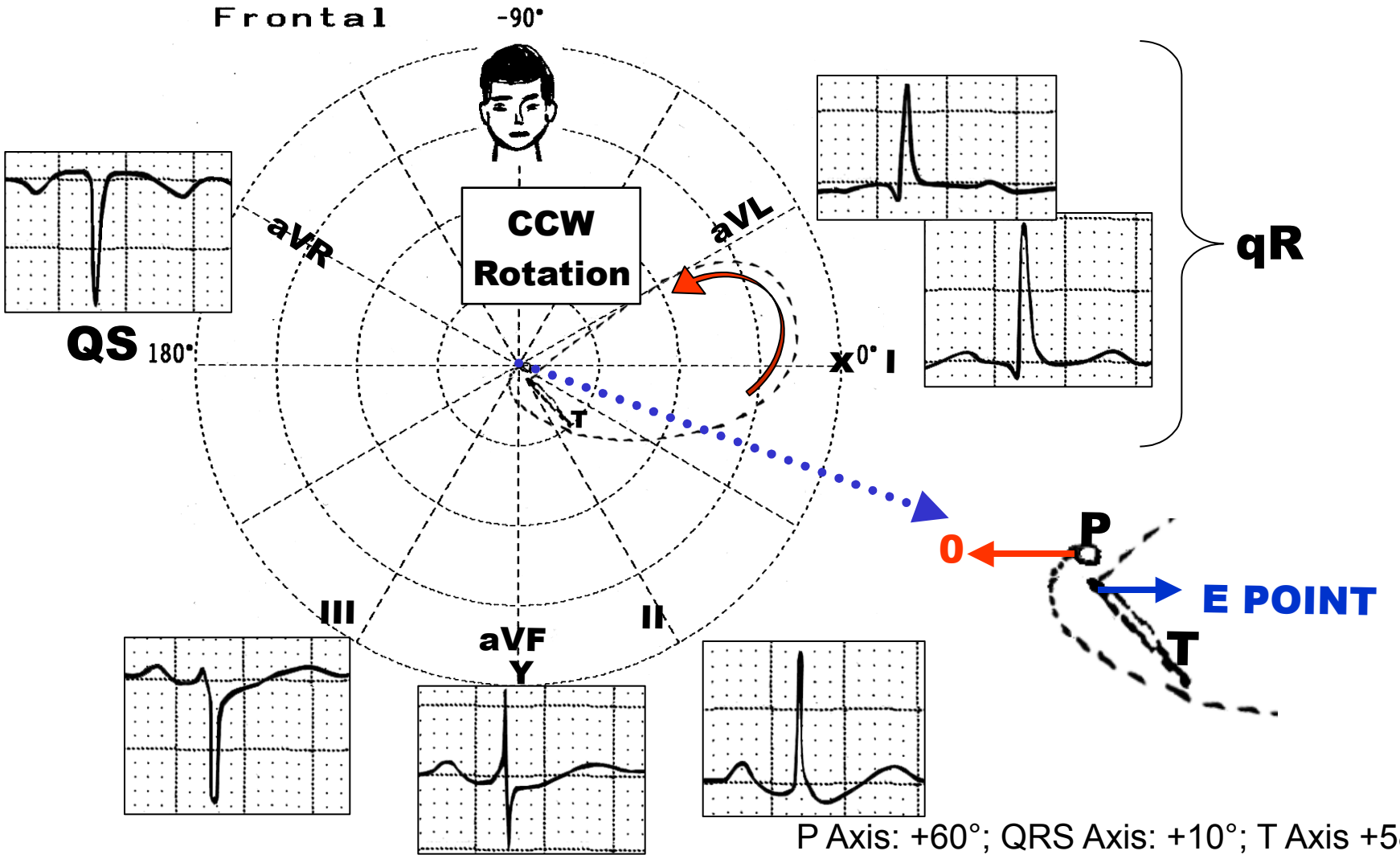


Eventually, BrS has not incomplete or complete RBBB. In this ECG, we have the typical type 1 ECG BrS pattern without RBBB.

ECG/VCG CORRELATION FRONTAL PLANE

Identification :

Name: ESR; Age: 37; Gender: Male; Ethnic Group: Caucasian; Weight: 72 Kg; Height: 1,75 m



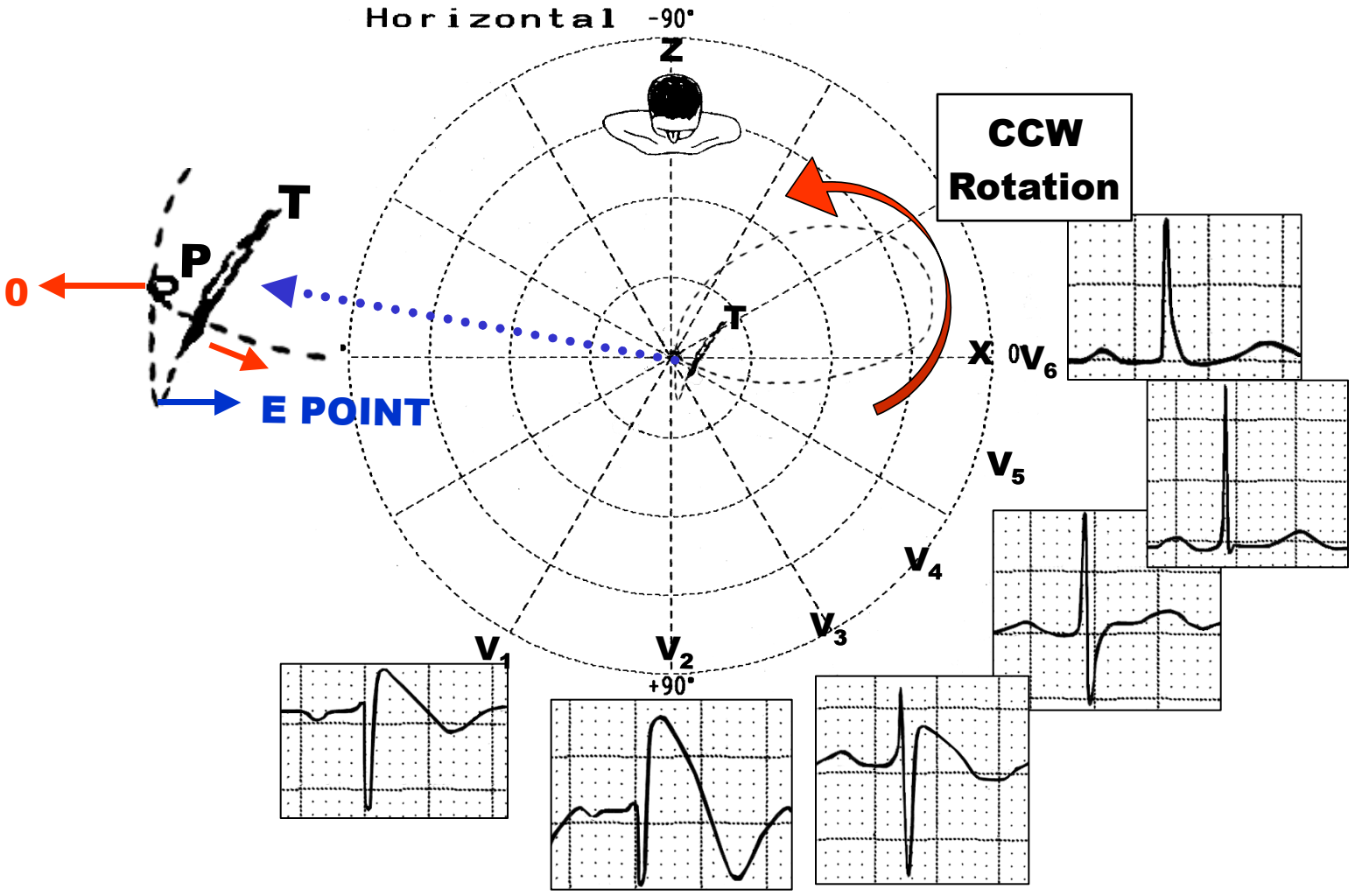
P Axis: +60°; QRS Axis: +10°; T Axis +58°

In this plane, ECG/VCG completely rules out the RBBB diagnosis: The QRS loop does not present Right End Conduction Delay (RECD) to the end of the QRS loop; the unipolar aVR limb lead shows QS pattern without broad terminal R wave as it would be seen in a RBBB. In the enhanced image to the right, we observe that the 0 point (start of QRS loop) does not coincide with the E point (end of QRS loop).

ECG/VCG CORRELATION HORIZONTAL PLANE

Identification :

Name: ESR; Age: 37; Gender: Male; Ethnic Group: Caucasian; Weight: 72 Kg; Height: 1,75 m

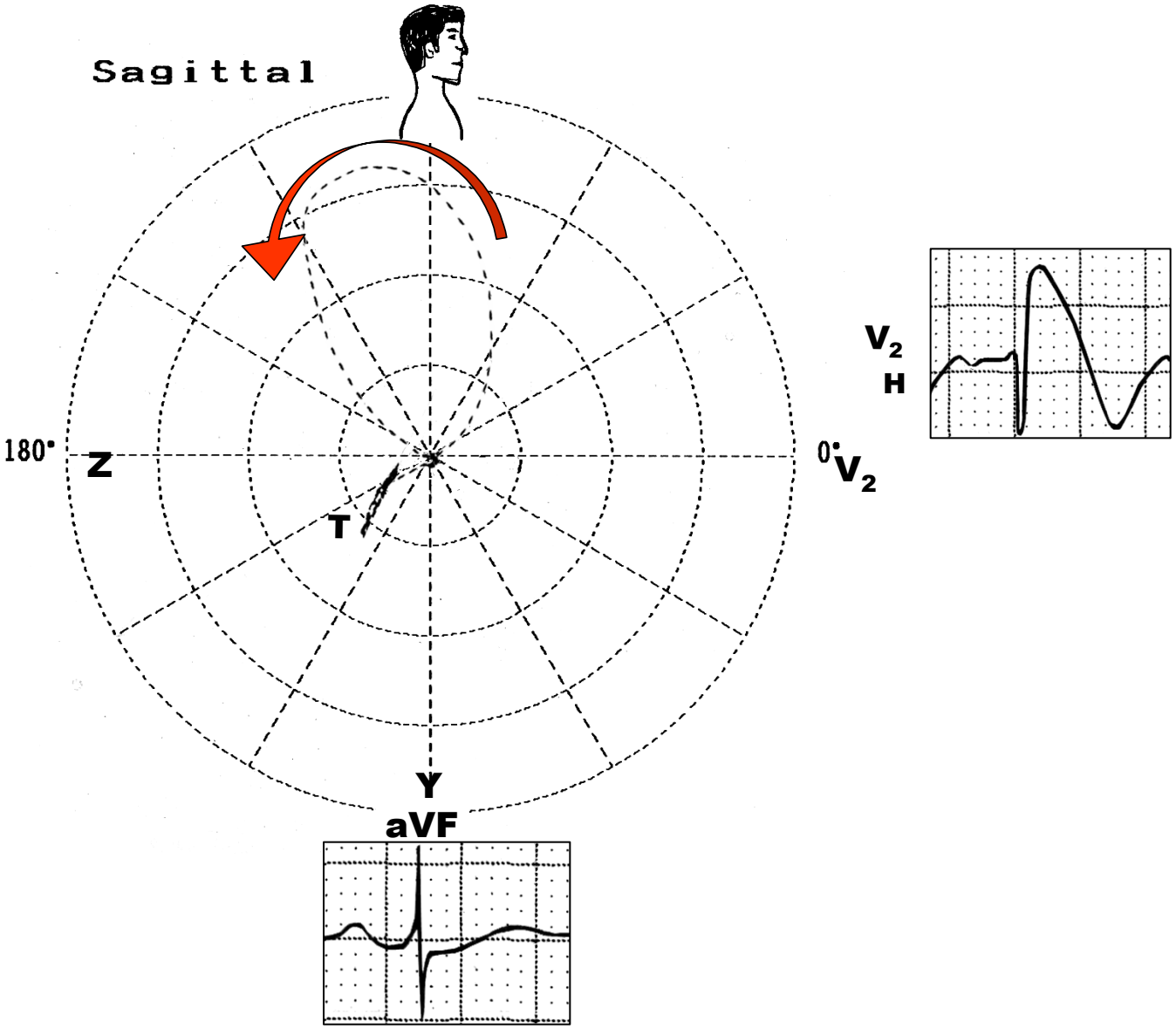


In this plane, we observe Brugada pattern Type 1. The VCG completely rules out RBBB: absence of RECD. The 0 point (start of QRS loop) does not coincide with the E point (end of QRS loop). This is located opposite to the 0 point.

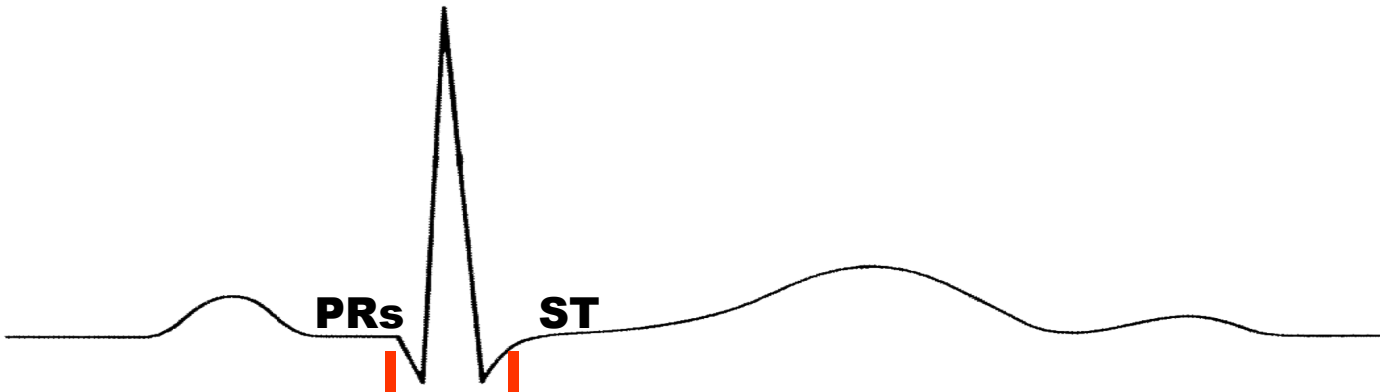
ECG/VCG CORRELATION RIGHT SAGGITAL PLANE

Identification :

Name: ESR; Age: 37; Gender: Male; Ethnic Group: Caucasian; Weight: 72 Kg; Height: 1,75 m



WAVES IN Ja AND J POINTS



Ja Point

J Point

DELTA WAVE δ

EPSILON WAVE ε

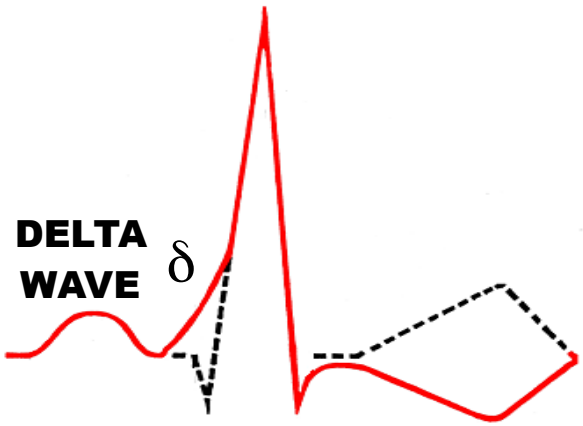
J POINT

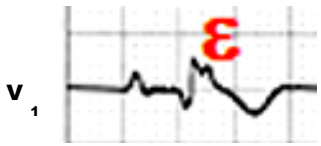
HYPERCALCEMIA

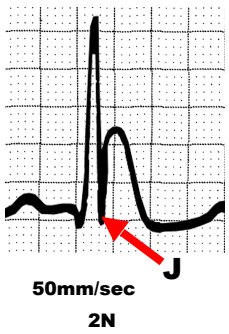
ARVD

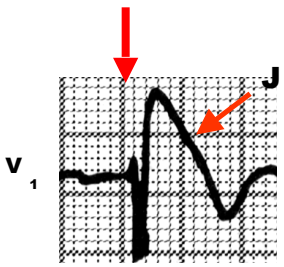
HYPOTHERMIA

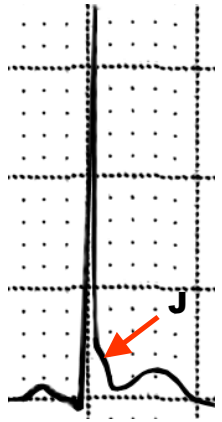
BrS











SHORT QT

MINIMAL ST SEGMENT

PHENOCOPIES

OTHER DENOMINATIONS: ACQUIRED FORMS OF BrS[1], BrS PSEUDO-ECG BRUGADA PATTERN OR PSEUDO-ECG BRUGADA PHENOTYPE

Phenocopy: A phenocopy is an individual whose phenotype (generally referring to a single trait), under a particular environmental condition, is identical to the one of another individual whose phenotype is determined by the genotype. In other words the phenocopy environmental condition mimics the phenotype produced by a gene. The term was coined by Richard Goldschmidt in 1935[2]. He used it to refer to forms, produced by some experimental procedure, whose appearance duplicates or copies the phenotype of some mutant or combination of mutants. A phenocopy is not a type of mutation, it is an environmentally induced, non-hereditary phenotypic modification that resembles a similar phenotype produced by a gene mutation (genocopy).

Any intervention that increases Ito, adenosine triphosphate-sensitive potassium current, delayed modifier potassium current or decreases inward currents (eg, L-type calcium current, fast sodium current) at the end of phase 1 of the AP can accentuate or unmask ST-segment elevation, similar to that found in the BrS, thus producing acquired forms of the BrS. Several drugs in addition to sodium-channel blockers and conditions that induce transient ST-segment elevation such as that in the BrS, developing acquired forms of the BrS

1. Shimizu W. Acquired forms of the Brugada syndrome. J Electrocardiol. 2005; 38:22-25
2. Goldschmidt. R., 1935. Gen und Ausseneigenschaft. I. Zeitschr. ind. Abstl. 69: 38-69

MAIN CAUSES OF BrS PHENOCOPIES

1. Young pattern
2. Astenic habit
3. Technical problem of inertia with the recording device;
4. Acute right ventricular infarction-ischemia
5. Acute pericarditis
6. Prinzmetal variant or vasospastic angina
7. Ventricular aneurysm
8. Changes during percutaneous transluminal coronary angioplasty
9. Early repolarization pattern
10. Atypical Right or Left bundle branch block
11. Left ventricular hypertrophy
12. Hypercalcemia
13. Hyperkalemia
14. Profound Electrolyte Disturbance induced by diabetic ketoasidosis
15. Central nervous system disease: intracranial hemorrhage.
16. Post-direct current cardioversion.
17. Apical hypertrophic cardiomyopathy.
18. Acute pulmonar embolism
19. Hyperthermia
20. Increased insulin level
21. Acute myocarditis
22. Myocarditis: e.g. Chagas disease.
23. Arrhythmogenic right ventricular dysplasia (ARVD).

MAIN CAUSES OF BrS PHENOCOPIES

24. Myocardial tumors
25. Compression of RVOT:
    - Mediastinal tumor
    - Hemopericardium
    - Pectus excavatum
    - Straight back syndrome
26. Dissecting aortic aneurysm
27. Hypothermia: Osborn wave, “camel-hump” like sign, “hump” like deflection.
28. Class 1A and 1C antiarrhythmic drugs.
29. Alcohol intoxication
30. Cocaine intoxication
31. Propofol infusion syndrome
32. Thiamine deficiency
33. Tricyclic antidepressant
34. Tetracyclic antidepressant
35. Phenothiazines
36. Selective seronotonin reuptake inhibitors (SSRIs)
37. Lithium
38. First generation histaminic H1
39. Calcium antagonists
40. Nitrates
41. Long QT syndrome type 3
42. Friedreich’s ataxia
43. Duchenne muscular dystrophy or Duchenne-Erb paralysis
44. Steinert’s disease

DIFFERENTIAL CHARACTERISTICS BETWEEN BRUGADA SYNDROME AND VASOSPASTIC ANGINA

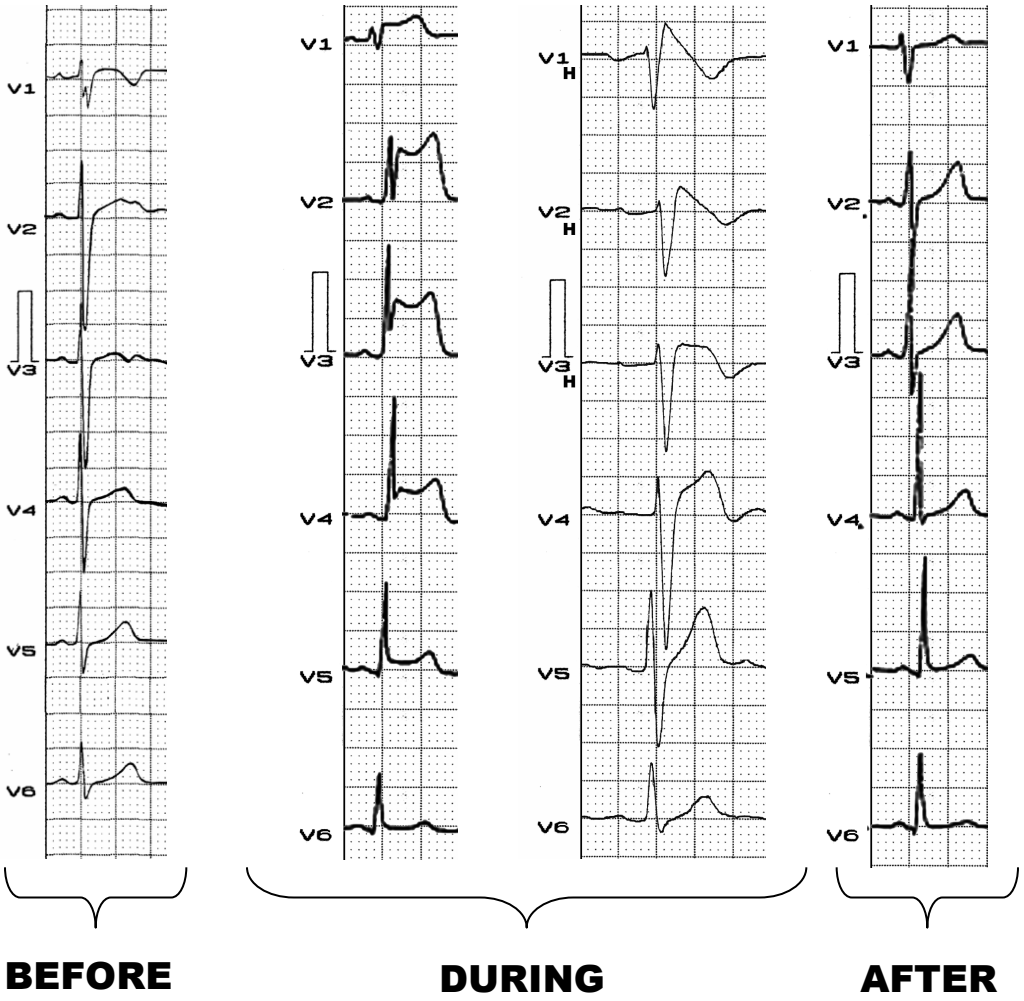
	BRUGADA SYNDROME	**VASOSPASTIC ANGINA**
Precordial pain:	No.	Yes.
Tendency to VT/VF:	High.	High.
Structural heart disease:	Absent.	Could exist.
Response to nitrates and nitroglycerine:	Null.	Improves or suppresses clinical/electrocardiographic manifestations.
Permanence of ST segment elevation:	Persistent (or fluctuating) and without pain.	Brief, transitory and accompanied by pain.
Cause:	Na^+ channel genetic alteration.	Possible alteration in nitrous oxide production in vascular wall.
Presence of image in mirror or reciprocal in ECG:	Could be present.	Present.
Topography of ST elevation:	Right precordial leads from V_1 to V_3. Rarely, it could be observed in inferior and lateral wall. It is triggered or increased by class IC and IA antiarrhythmic agents.	Variable. It could alternate between precordial and inferior leads. It could be triggered by hyperventilation.
Dromotropic disorders:	First-degree AV block by H-V prolongation in 50% of the cases and in carriers of the mutation.	They could occur transitorily up to a high degree of AV block during the episode, and they are associated to a higher risk of arrhythmia and SCD.
Persistent T wave inversion:	Negative T wave in precordial leads from V_1 to V_3, characteristic of type 1.	Inverted and deep T waves from V_1 to V_4 associated to anterior hypokinesia, suggesting myocardial “stunning” that indicates critical lesion of the anterior descending artery: “LAD-T wave pattern”.

DIFFERENTIAL CHARACTERISTICS BETWEEN BRUGADA SYNDROME AND VASOSPASTIC ANGINA

	BRUGADA SYNDROME	VASOSPASTIC ANGINA
Presence of transitory Q wave:	No.	Could happen.
Stress test:	Elevation could be normalized during effort.	Variable response.
Myocardial scintigraphy with thallium 201:	Normal.	Transitory transmural hypoperfusion.
Response to endovenous ergonovine maleate test in a 0.05 to 0.40mg dose (alpha adrenergic and serotoninergic receptor stimulant)	There could be a mild and diffuse reduction of caliber without spasm when doses = or > than 0.40mg are used.	Intense coronary spasm accompanied by pain and ST elevation. Possible cardiac block, asystole and VT.
Response to hyperventilation:	Does not modify	Severe spasm and reproduction of clinical and electrocardiographic manifestations.
Response to intracoronary acetylcholine, with each dose administered in a time longer than 1 minute in 10μg, 25μg, 50μg and 100μg doses, in 5-minute intervals:	It may worsen ST elevation with paradoxical dilatation of coronary vessels.	Severe spasm and reproduction of clinical and electrocardiographic manifestations.
Response to magnesium sulphate:	Not mentioned.	It suppresses events induced by hyperventilation and exercise.
Treatment:	ICD in association with quinidine or isoproterenol, drugs that contribute to decrease the number of shocks. Isoproterenol prescribed in "electrical storms" associated to general anesthesia and cardiopulmonary “bypass”.	Calcium antagonists such as nifedipine, diltiazem, verapamil and felodipine associated to nitrates. There are references of benefits with prazosin.

EXAMPLES OF PHENOCOPIES

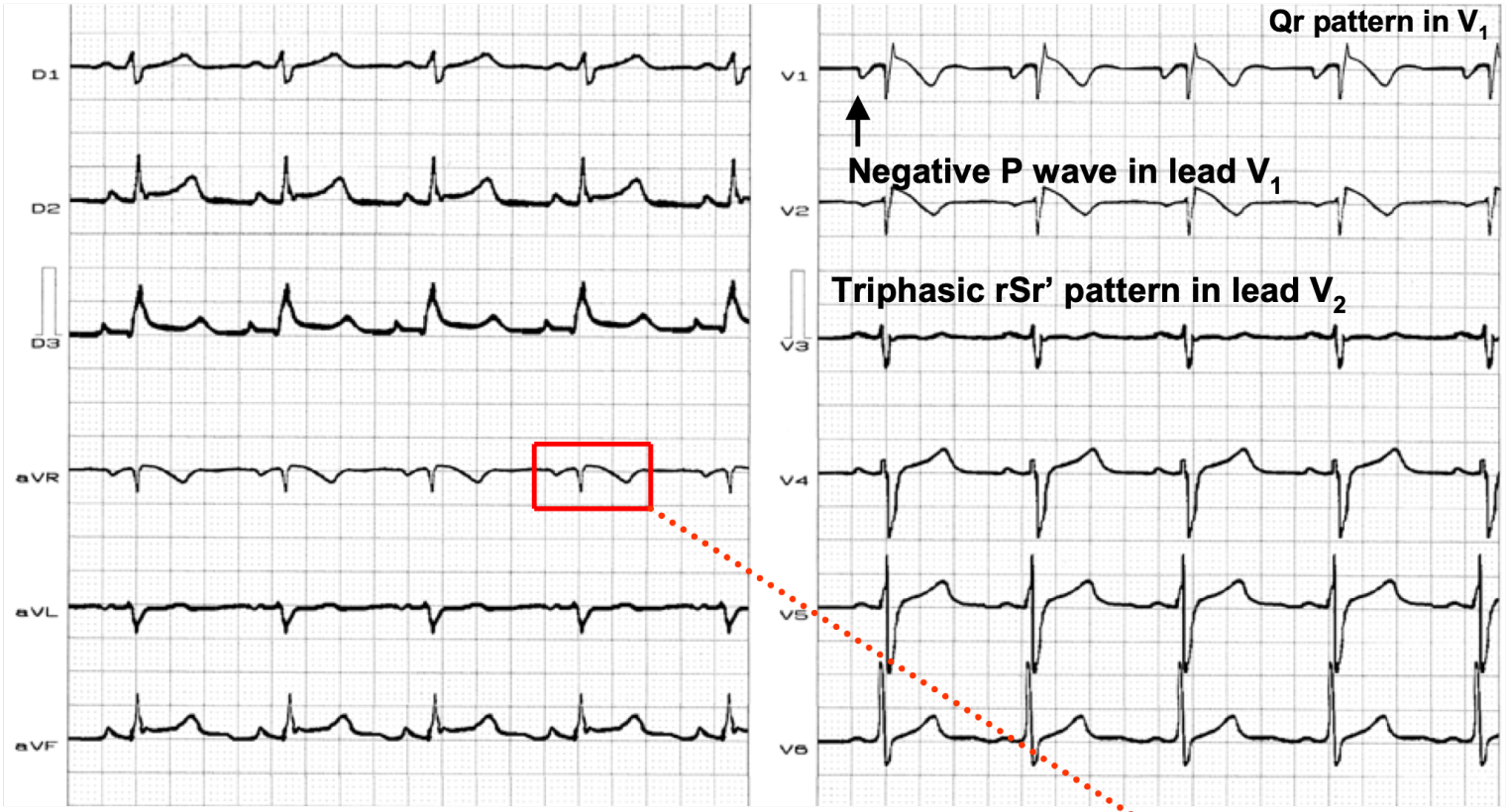
DIFFERENTIAL DIAGNOSIS WITH CHANGES DURING PERCUTANEOUS TRANSLUMINAL CORONARY ANGIOPLASTY



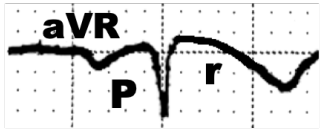
In a patient in whom ST segment elevation was present in leads V_1 through V_3 during the PTCA procedure, balloon occlusion of the proximal left anterior descending coronary artery was associated with a QRS duration increase in these leads.

1. Surawicz B, Orr CM, Hermiller JB, et al: QRS changes during percutaneous transluminal coronary angioplasty and their possible mechanism. J Am Coll Cardiol 1997; 30:452

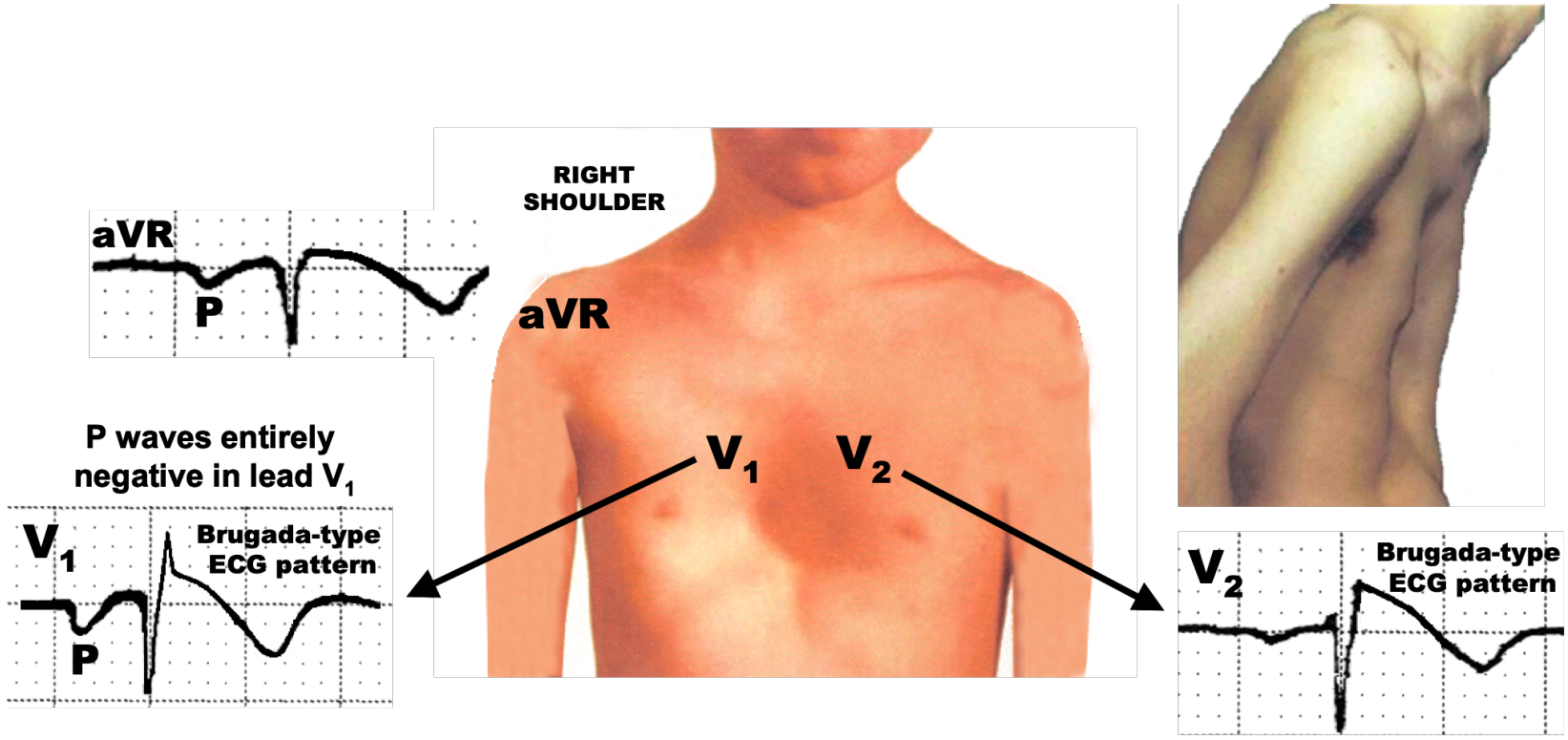
PECTUS EXACAVATUM WITH BrS PHENOCOPY



PSEUDO-IRBBB PATTERN: the last r' wave is small.
ST-segment elevation in the right precordial ECG leads and V_2.
Non ischemic-type T waves inversion in right precordial leads.
Embryonic final r wave in aVR lead and coved type ST segment elevation.

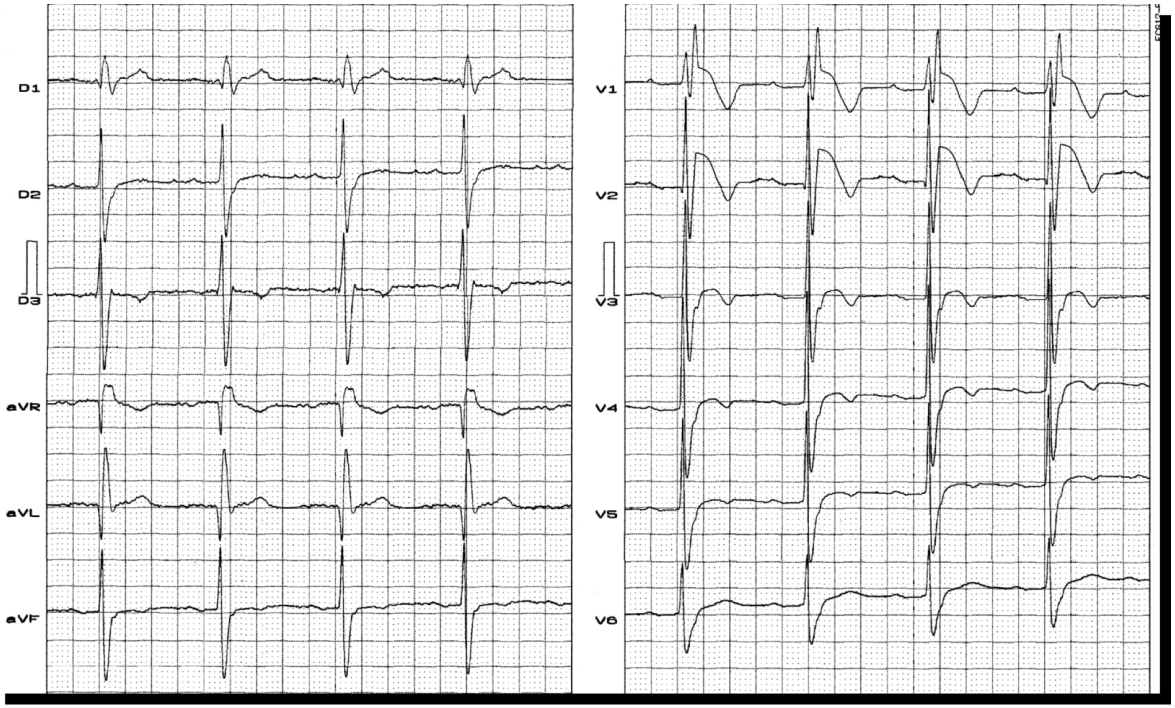


PECTUS EXACAVATUM WITH BrS PHENOCOPY



STEINERT'S DISEASE WITH BrS PHENOCOPY

Name: N.M.; **Date: 0**4/2003.; **Age:** 52 years old**.** **Gender:** Male. **Weight:** 82 Kg. **Height:** 1,70 m**. Ethnic group:** Asian**.**
Medication in use: Metformin 850mg 2x, enalapril maleate 2 x day.



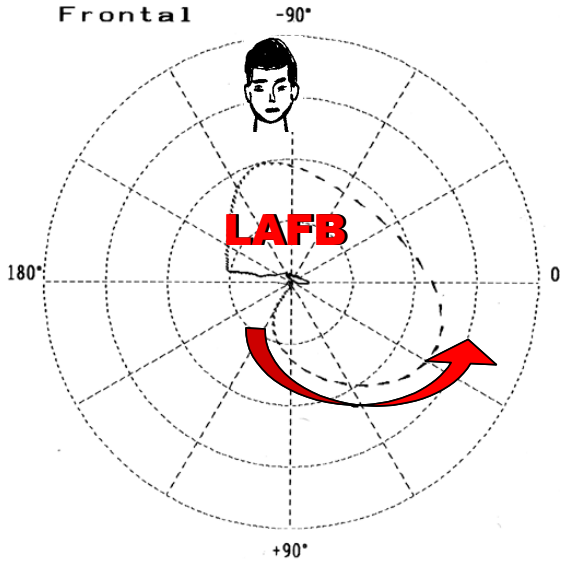
CLINICAL DIAGNOSIS: myotonic muscular dystrophy (Steinert's disease) / type 2 diabetes mellitus / systemic hypertension. ELECTROCARDIOGRAPHIC DIAGNOSIS: sinus rhythm, HR: 55 bpm, PR: 250msec. QRS duration: 165msec; SAQRS: difficult to determine near the – 40° CONCLUSION: first degree AV block + complete RBBB + left anterior fascicular block (LAFB), probable trifascicular block. Coved type J point and ST segment elevation in V1 and V2: Brugada-like type 1 ECG pattern or Brugada sign.

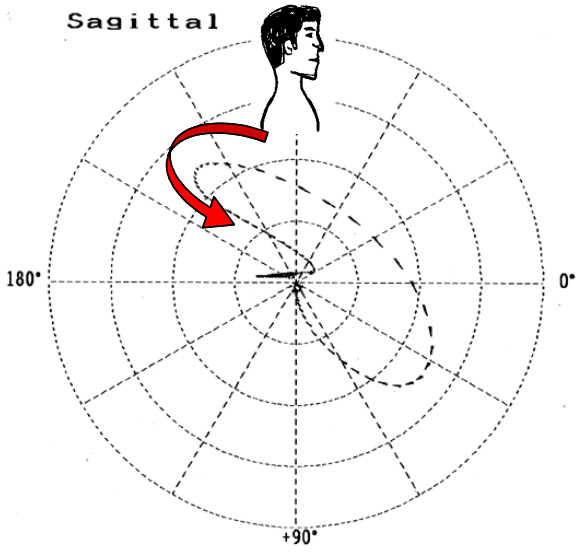
STEINERT'S DISEASE WITH BrS PHENOCOPY

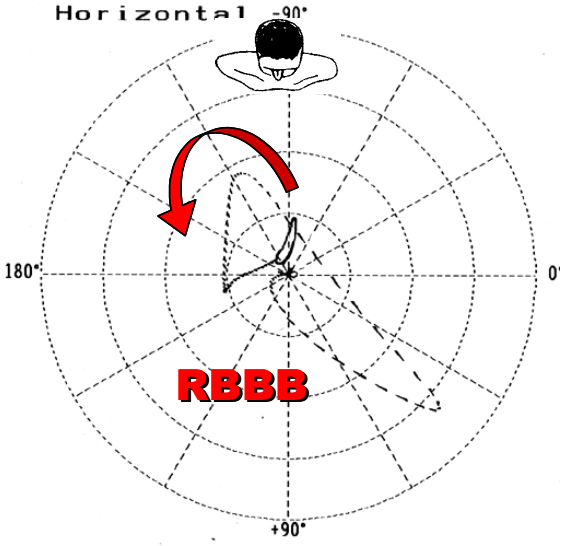
VETORCARDIOGRAM

Name: N.M.; **Date: 0**4/2003.; **Age:** 52 years old**.**; **Gender:** Male. **W eight:** 82 Kg. **Height:** 1,70 m**. Ethnic group:** Asian**.**
Medication in use: Metformin 850mg 2x, enalapril maleate 2 x day.

Sensi.	2
Timer	2 msec
Loop	All Loop
Sagittal	Right
Z Axis	Back
Filter	Hum
	Muscle
	Drift

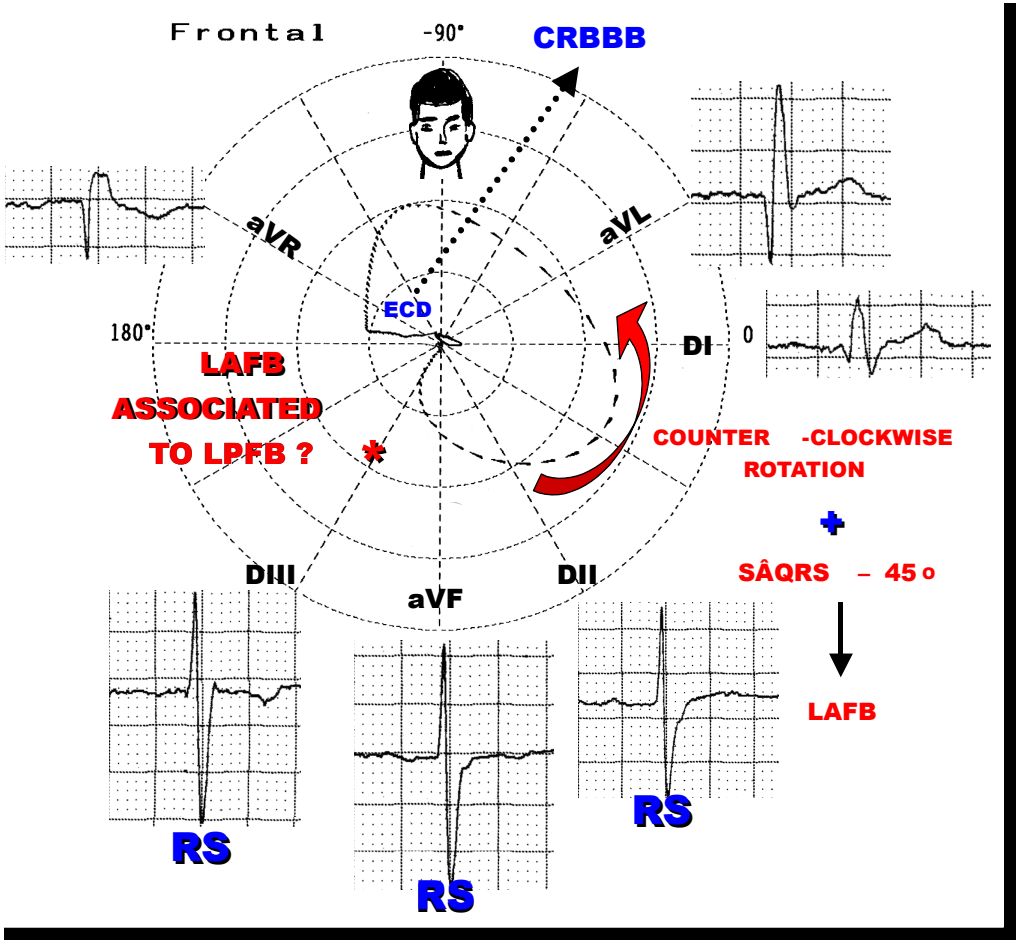






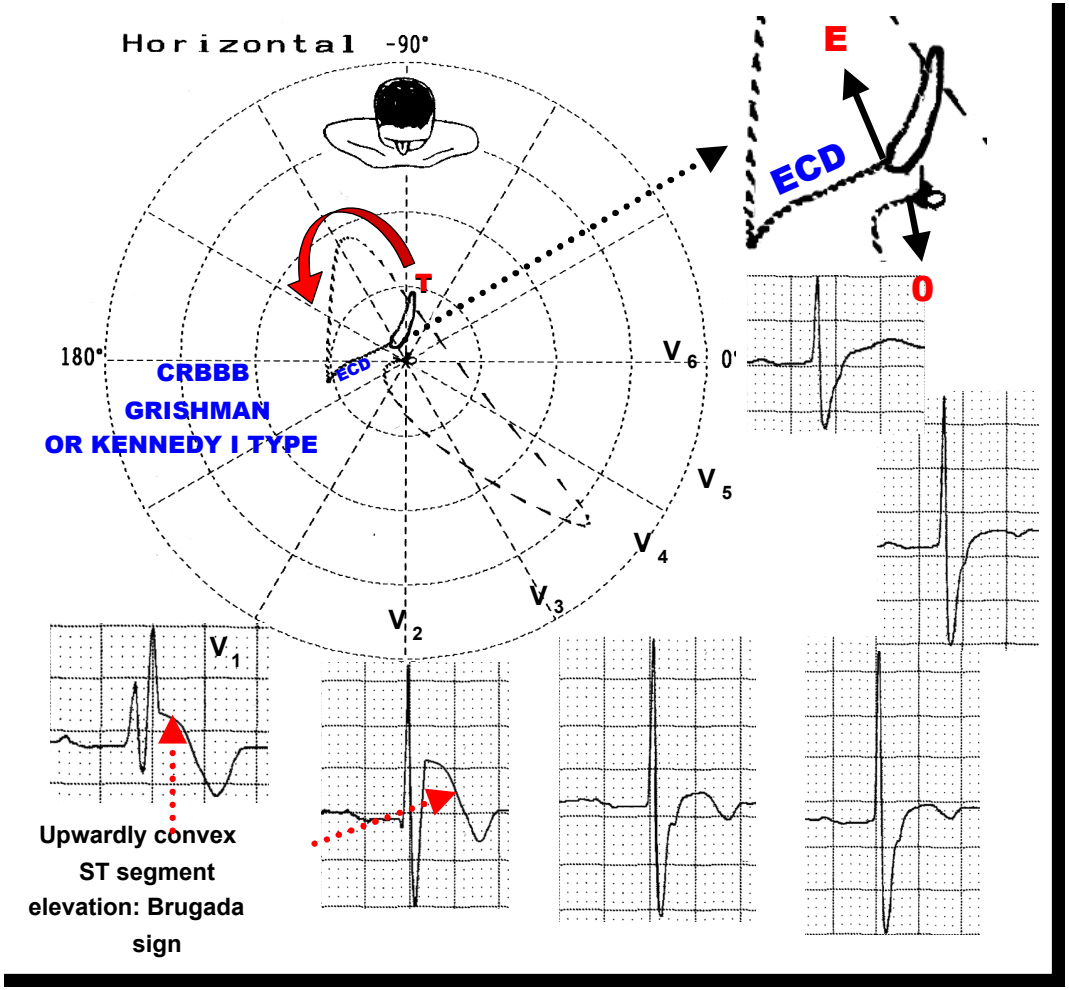
STEINERT'S DISEASE WITH BrS PHENOCOPY

ECG/VCG FRONTAL PLANE



In classical LAFB, in inferior leads, we observe rS, pattern in this case, R wave voltage is higher, causign the RS pattern in this inferior leads. The QRS loop morphology is rounded and not elliptical, as in typical LPFB. Both facts suggest some degree of LPIFB associated. This notion is supported by the presence of first degree AV block, which may indicate dromotropic problems in the posterior inferior division.

STEINERT'S DISEASE WITH BrS PHENOCOPY
ECG/VCG HORIZONTAL PLANE



The end of the QRS loop (E point) does not coincide with the 0 point, just as it happens in Brugada disease VCG.