Mujer de edad avanzada portadora de cardiomiopatia hipertrófica sintomática

Old woman carrying symptomatic hypertrophic cardiomyopathy

From Raimundo Barbosa Barros MD Nickname “ The fox”
Coronary Center Hospital de Messejana Dr. Carlos Alberto Studart Gomes Fortaleza-Ceará-Brazil

Mujer de 72 años, fue admitida en la emergencia con episodios pré-sincopales.
Refiere história de fatiga e palpitaciones.
Estaba utilizando captopril y carvedilol (6, 25 mg dos veces por día).
Antecedentes familiares negativos
El ecocardiograma es compatible con miocardiopatía hipertrófica não obstrutiva no repouso y el ECG sugestivo del síndrome taqui-bradicaria.
Después de la suspensión de la terapia betabloqueadora está asintomática en reposo.
¿Cual es la conducta adecuada?
Me gustaría escuchar las opiniones del Foro porque este caso se discutirá en la próxima semana
Raimundo

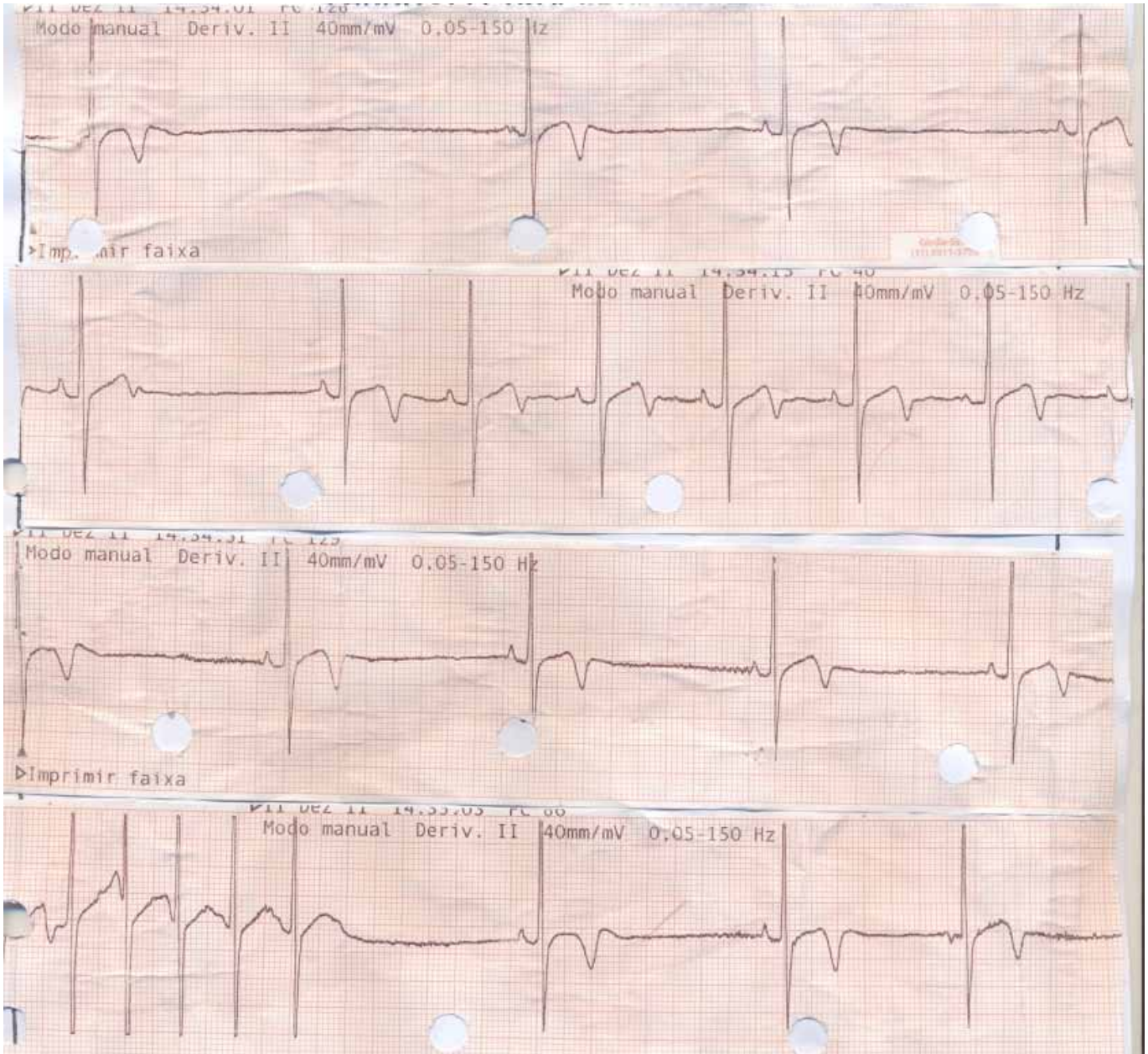
72-years old woman I was admitted in emergency with near syncopal episodes
She complain of fatigue and palpitations.
Negative family history
She was using captopril and carvedilol (6, 25 mg twice a day).
Echocardiogram is compatible with Non-obstructive Hypertrophic cardiomyopathy at rest.
ECG is suggestive of tachycardia-bradicaria syndrome
After suspension of beta-blocker therapy she is asymptomatic at rest.

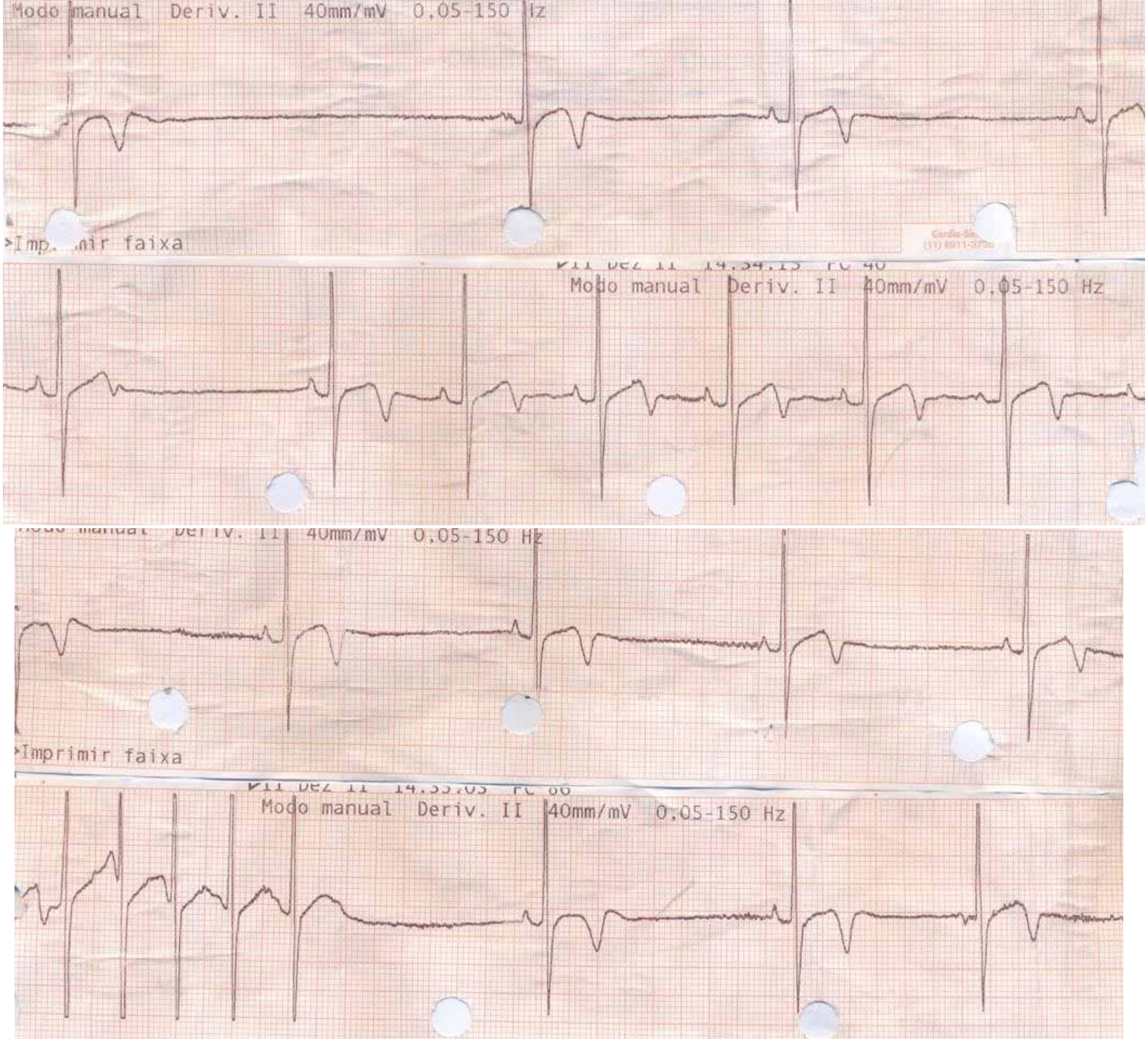
Question: What is the proper approach?

I'd like to hear the views of the forum because this case will be discussed in the next week
Raimundo

ECG realizado durante un episódio pre-sincopal

ECG preformed during near-syncope episode




Modo manual Deriv. II 40mm/mV 0,05-150 Hz
Imp. mir faixa
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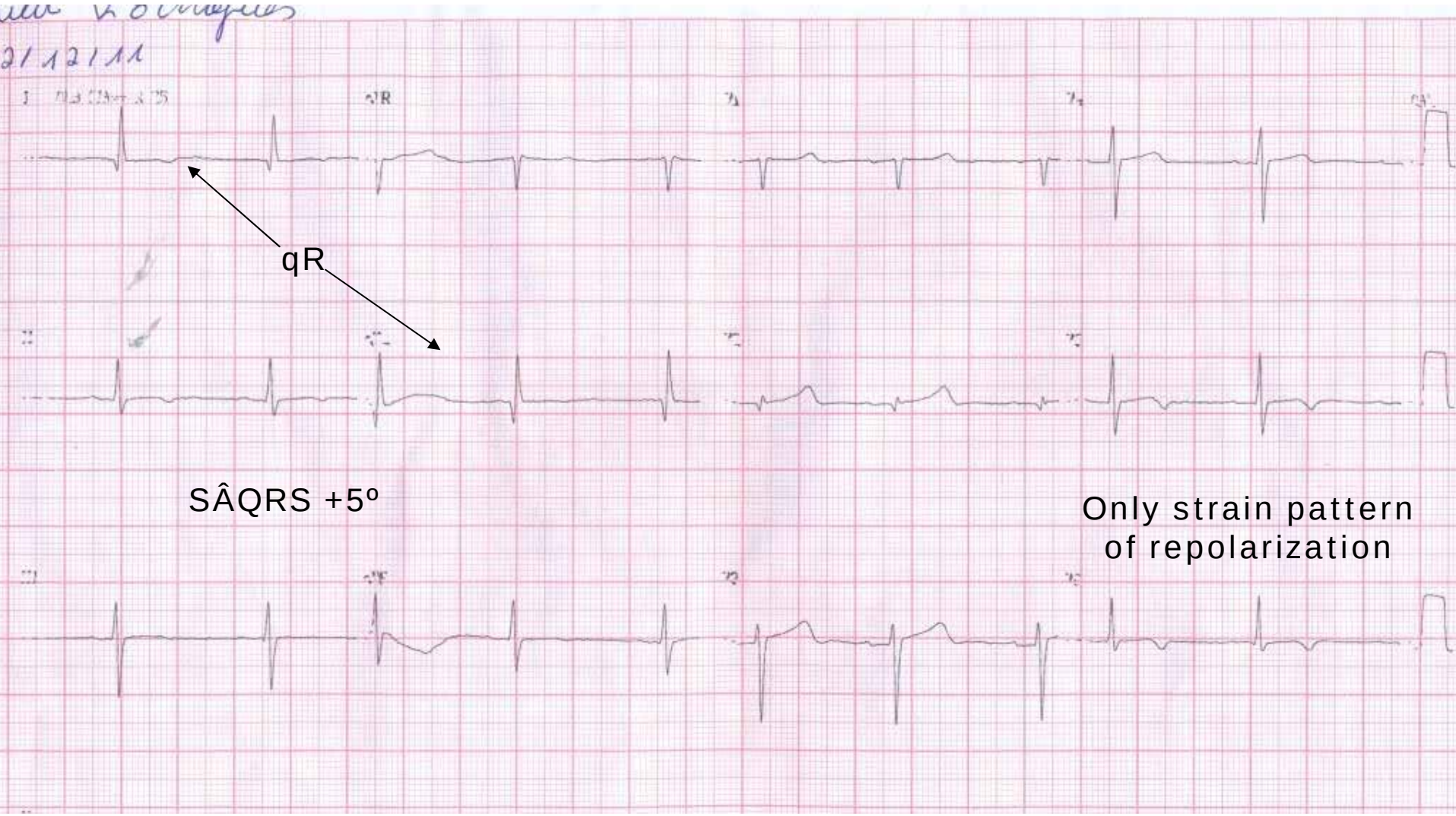
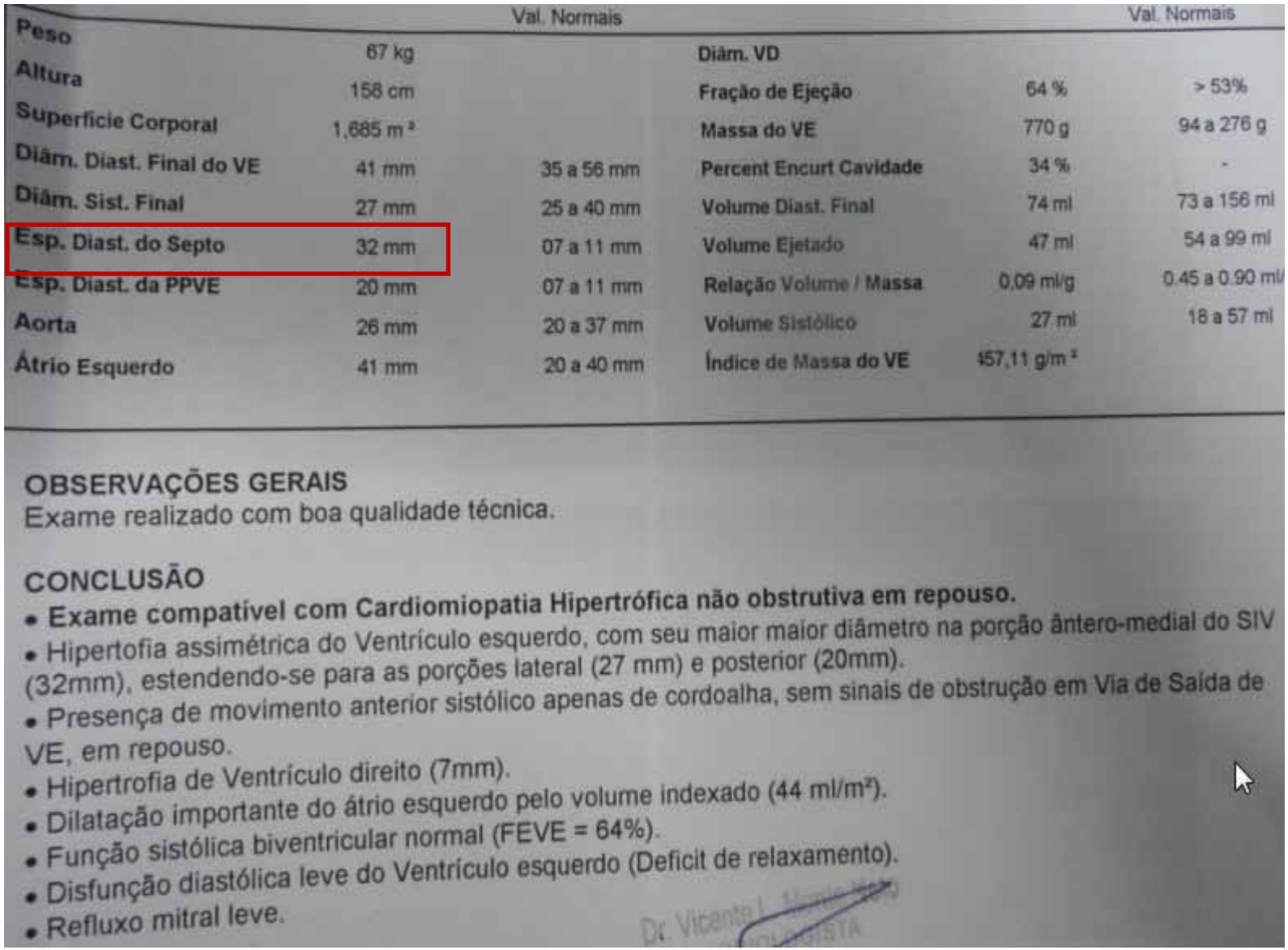
40mm/mV 0,05-150 Hz
Imprimir faixa
Modo manual Deriv. II 40mm/mV 0,05-150 Hz

ECG realizado 3 dias después de la retirada del beta-bloqueador

ECG preformed 3 days later discontinue ß-bloker theraphy



Ecocardiograma/ Transthoracic Echocardiogram



		Val. Normais			Val. Normais
Peso	67 kg		Diâm. VD		
Altura	158 cm		Fração de Ejeção	64 %	> 53%
Superfície Corporal	1,685 m²		Massa do VE	770 g	94 a 276 g
Diâm. Diast. Final do VE	41 mm	35 a 56 mm	Percent Encurt Cavidade	34 %	-
Diâm. Sist. Final	27 mm	25 a 40 mm	Volume Diast. Final	74 ml	73 a 156 ml
Esp. Diast. do Septo	32 mm	07 a 11 mm	Volume Ejetado	47 ml	54 a 99 ml
Esp. Diast. da PPVE	20 mm	07 a 11 mm	Relação Volume / Massa	0,09 ml/g	0.45 a 0.90 ml/
Aorta	26 mm	20 a 37 mm	Volume Sistólico	27 ml	18 a 57 ml
Átrio Esquerdo	41 mm	20 a 40 mm	Índice de Massa do VE	157,11 g/m²	

OBSERVAÇÕES GERAIS

Exame realizado com boa qualidade técnica.

CONCLUSÃO

- **Exame compatível com Cardiomiopatia Hipertrófica não obstrutiva em repouso.**
- Hipertofia assimétrica do Ventrículo esquerdo, com seu maior maior diâmetro na porção ântero-medial do SIV (32mm), estendendo-se para as porções lateral (27 mm) e posterior (20mm).
- Presença de movimento anterior sistólico apenas de cordoalha, sem sinais de obstrução em Via de Saída de VE, em repouso.
- Hipertrofia de Ventrículo direito (7mm).
- Dilatação importante do átrio esquerdo pelo volume indexado (44 ml/m²).
- Função sistólica biventricular normal (FEVE = 64%).
- Disfunção diastólica leve do Ventrículo esquerdo (Deficit de relaxamento).
- Refluxo mitral leve.

Dr. Vicente [illegible]

Severe septal thickness in anteromedial area (32mm) Non-obstructive HCM, EF 64% Mild diastolic dysfunction and mitral reflux.

Holter without ß-blocker

PVCs <1%
Supraventricular PVCs <1%

Hospital de Messejana
Secretaria de Saúde do Estado do Ceará / SUS
Serviço de Métodos Gráficos
Fone: 85 3101-4074

Relatório de Holter

1 - Dados do Exame

Nº do Exame	Data do Exame: 13/12/2011 13:39	Código
1RI-05139	Protocolo: Holter de 3 canais	1RI-0471

2 - Dados do Paciente:

Nom[redacted] Idade: 72
Sexo: F Altura: Peso: Fumante:

Diagnóstico:

Motivo do Exame:

3 - Médico Solicitante

Nome: Dra. Cinthia Montenegro Teixeira Tel:
Clínica: Fax:

4 - Resumo Estatístico

Totais:

Duração (h):	20:25	
Nº Total de QRS's:	82.360	
Ectópicos Ventriculares:	4	(<1%)
Ectópicos Supraventriculares:	400	(<1%)
Artefatos (%):	2	

Freqüência Cardíaca:

Mín: 55 bpm às 23:57:02
Média: 70 bpm
Máx: 104 bpm às 01:56:55

F.C >= 120 bpm durante 00:00:06 h

Arritmias Ventriculares:

4 Isoladas, das quais
0 em 0 episódios de Bigeminismo
0 Episódios em Pares
0 Taquicardias

Pausas

0 Pausas (>= 2,0 s.)

Depressão do ST

C1: 0 episódios
C2: 0 episódios
C3: 0 episódios

Arritmias Supraventriculares:

99 Isoladas
106 Pareadas
20 Taquicardias

A maior: 21 bat., 143 bpm às 07:02:54
A mais rápida: 5 bat., 162 bpm às 15:27:18
A mais lenta: 5 bat., 104 bpm às 07:31:31

Elevação do ST

C1: 0 episódios
C2: 0 episódios
C3: 0 episódios

Final comments

By Andrés and Raimundo

This is a 72-years old woman with near syncopal episodes, fatigue and palpitations, echocardiogram compatible with Non-obstructive Hypertrophic cardiomyopathy at rest and severe segmental anteromedial septal hypertrophy without Impaired left ventricular systolic function. It is not necessary carvedilol because she has not CHF. Additionally, this drug worst the tachy-brady syndrome. Impaired left ventricular systolic function (ILVSF) in hypertrophic cardiomyopathy (HCM) is associated with increased risk of HF death or heart transplantation, and of cardiovascular dysfunctions, including cardiac dilatation, decreased development of small arterioles, and fibrosis.(1) the present case has not ILVSF because her LVEF= 64. Adrián Fernández, M.D., from the University hospital in Buenos Aires, Argentina, and colleagues investigated the prevalence, clinical parameters, and long-term outcome of ILVSF in 382 patients with HCM, along with examination of the pathological findings of explanted hearts. the combined end point of the study after a median follow-up of three years was heart failure death or heart transplantation. The investigators identified 24 patients with ILVSF, characterized by LVEF < 50% at rest. compared to patients with normal systolic function, patients with ILVSF were younger (average age, 43.5 versus 55.3 years), with larger LV end-diastolic cavity diameter (average, 53.2 versus 43.8 mm), larger left atrium (average, 51.2 versus 44.3 mm), and lower fractional shortening (average, 30.7 versus 45.5 percent). a total of 14 patients with ILVSF and three with normal systolic function achieved the end point. fibrosis represented an average 30.5% of the LV in explanted hearts, with a direct correlation between fibrosis and ventricular dilation, and an inverse relationship between fibrosis and EF. there was a significant decrease in the number and length density of small arterioles (<50 μm in diameter).

"ILVSF in HCM has a poor prognosis and is associated with fibrosis and selective decreased development of small arterioles.

1. **Fernández A, Vigliano CA, Casabé JH, et al. Comparison of prevalence, clinical course, and pathological findings of left ventricular systolic impairment versus normal systolic function in patients with hypertrophic cardiomyopathy. Am J Cardiol. 2011 Aug 15;108:548-55.**

Sick Sinus Syndrome (SSS)

Atrial arrhythmias, and in particular AF, are common in patients with sinus node dysfunction. This tachy-brady variant of SSS is one of the most common indications for permanent pacing. Early retrospective studies showed a major reduction in the incidence of AF with atrial based pacing (AAI or DDD modes) compared with ventricular pacing alone (VVI mode).(1) These reports also suggested that the rates of CHF, strokes, and mortality were all reduced with atrial based pacing. This led to the common practice of implanting dual chamber devices in all patients with sinus node dysfunction, despite the lack of prospective data supporting this strategy.

Anderson and colleagues compared single chamber atrial pacing with ventricular pacing in 225 patients with sinus node dysfunction.(2) They showed a significant reduction in the development of AF with atrial pacing. This study also established the relative safety of atrial pacing with no ventricular back up, as the rate of heart block requiring pacemaker revision to a dual chamber system was low (0.6%/year). In addition to reducing the incidence of AF, long term follow up of these patients revealed reductions in mortality, stroke, and CHF in the atrial pacing group. In the pacemaker selection in the elderly (PASE) study, 407 patients were implanted with dual chamber devices and were then randomized to pacing in DDDR or VVIR modes.(3)

There was a reduction in the incidence of AF from 28% to 19% with DDDR pacing ($p = 0.06$) in the subgroup of patients with SSS, but no difference was noted in those patients with heart block. No mortality reduction was noted with DDDR pacing in this study. One possible explanation for the failure to observe benefit with dual chamber pacing in this study was the relatively high crossover rate (26%) from VVIR to DDDR mode.

1. ***Connolly SJ, Kerr C, Gent M, et al. Dual chamber versus ventricular pacing. Critical appraisal of current data. Circulation 1996; 94:578–583,***
2. ***Andersen HR, Nielsen JC, Thomsen PEB, et al. Long-term follow-up of patients from a randomized trial of atrial versus ventricular pacing for sick-sinus syndrome. Lancet.1997;350:1210–1216,***
3. ***Lamas GA, Orav EF, Stambler BS, et al. Quality of life and clinical outcomes in elderly patients treated with ventricular pacing as compared with dual-chamber pacing. Pacemaker selection in the elderly investigators. N Engl J Med. 1998; 338:1097–1104.***

The much higher crossover rate was likely due to the study design, where randomisation was by “software” (by programming the device mode), in contrast to randomisation by “hardware” (the positioning of the leads) in the Danish study. Since it is much easier to reprogram a device than to revise a pacing system to implant an atrial lead, the crossover rate was higher. In the pacemaker atrial tachycardia (PAC-a-TACH) trial, 198 patients with sick sinus syndrome were randomised to ventricular or dual chamber pacing. No effect on the incidence of atrial fibrillation was noted, but there was a significant reduction in mortality with dual chamber pacing.

The largest study to date evaluating the role of pacing mode on AF was the Canadian trial of physiologic pacing (CTOPP).(1) In this study, 2568 subjects were randomized to atrial based pacing (atrial or dual chamber) or ventricular pacing. There was an 18% reduction of AF with atrial based pacing in this trial, but no effects on mortality or stroke were observed. It is noteworthy that the mean duration of follow up in this trial was three years, while a mortality benefit of atrial pacing was only observed in the study of Andersenet al(2) when the mean follow up was extended to 5.5 years.

The results of these studies, in general, support the use of atrial based pacing for the prevention of AF, at least in subjects with symptomatic sinus node dysfunction. The benefit of such pacing in reducing mortality in less clear.

1. ***Connolly SJ, Kerr CR, Gent M, et al. Comparison of the effects of physiologic pacing versus ventricular pacing on cardiovascular death and stroke. N Engl J Med.2000; 342:1385–1391.***
2. ***Andersen HR, Nielsen JC, Thomsen PEB, et al. Long-term follow-up of patients from a randomized trial of atrial versus ventricular pacing for sick-sinus syndrome. Lancet.1997;350:1210–1216***

The choice of pacing mode (AAI *v* DDDR) and the relative benefit of single chamber atrial and dual chamber pacemakers remains unknown, because there have been no controlled studies addressing this issue. Atrial pacemakers have the advantages of lower costs and increased longevity. The disadvantages of these systems include the inability to optimize AV delay, and the absence of ventricular pacing if complete heart block or a lead malfunction develop.

Although the optimization of AV delay may be important in certain patients, in general ventricular activation through the native conduction system is superior haemodynamically to right ventricular pacing.(1)

The risks of developing heart block can be minimized by avoiding atrial pacemakers in subjects with bundle branch block or other severe intraventricular conduction delays, in patients who show atrioventricular block (Mobitz I or II) at pace rates of 130 bpm or less, or in patients where it is anticipated that potent AV nodal blocking drugs such a amiodarone will be needed in the future.

If a dual chamber pacemaker is implanted in the absence of heart block, then it is reasonable to program a prolonged AV delay or use one of the new features in pacemakers that automatically prolongs the AV delay to minimze ventricular pacing.(1)

1. **Rosenqvist M, Isaaz K, Botvinick EH, et al. Relative importance of activation sequence compared to atrioventricular synchrony in left ventricular function. Am J Cardiol.1991; 67:148.**
2. **Gold MR. Permanent pacing: new indications. Heart. 2001 Sep;86:355-360.**

Raimundo´s opinion

Na minha opinião há indicação do marcapasso preferencialmente bicameral com a finalidade de permitir a utilização de drogas cardiodepressoras e também para estabilizar o átrio e evitar fibrilação atrial.

Quanto ao CDI, eu tenho dúvidas quanto ao benefício, mesmo com o septo de 32 mm de espessura pois nos ensaios clínicos pacientes acima de 70 anos são excluídos.

Se fosse um paciente jovem eu indicaria o CDI.

Raimond's opinion

In my opinion there is indication of the bicameral pacemaker preferentially in order to allow the use of cardiopressant drugs and also to stabilize the atrium and prevent atrial fibrillation.

As for the CDI, I have doubts about the benefit, even with the septum of 32 mm thickness in clinical trials patients over 70 years are excluded.

If she were a young patient I indicate the CDI.

HYPERTROPHIC CARDIOMYOPATHY (HCM)

CONCEPT: complex heart disease, which is characterized by hypertrophy of variable location in the LV and/or RV, in most cases heredo-familial, autosomal polygenic dominant, with a high degree of penetrance (60% reveal familial relationship and 40% sporadic). to this date, eight genetic types are known to encode the sarcomere proteins by alteration in the chromosomes with the numbers: 1, 3, 11, 12, 14, 15, 19 and gene of actin with a great morphological diversity, and which is characterized in most cases by asymmetrical septum hypertrophy, obstructive or non obstructive (90%), in comparison to LV free wall (there are concentrical or symmetrical forms, characterized by nonselective hypertrophy, both in the septum and the free wall), with small or normal size of ventricular cavity (95%), hypodiastole, by increase of ventricular mass and rigidity responsible of distensibility involvement with a subsequent LV diastolic dysfunction.

in 5% to 10% of patients, in the late phase, they evolve into dilatation and systolic dysfunction resulting from myocardial fibrosis secondary to microinfarctions and possible associated coronary artery disease.

Myocardial ischemia and another element present in the disease secondary to: 1) microcirculation disease; 2) decrease of vasodilator capacity; 3) systemic compression of septal and subepicardial vessels; 4) fall of pressure in aorta root; 5) difficulty in coronary filling by hypertrophy; 6) coronary atherosclerosis in patients older than 50 years old; 7) excessive increase of mass and subsequent offer/demand disproportion.

The anatomopathological substrate is an important septal cell disorder (95%) and hypertrophy of the middle layer with narrowing of light of the intramural branches responsible of ventricular dysfunction as well as arrhythmias.

CLASSIFICATION

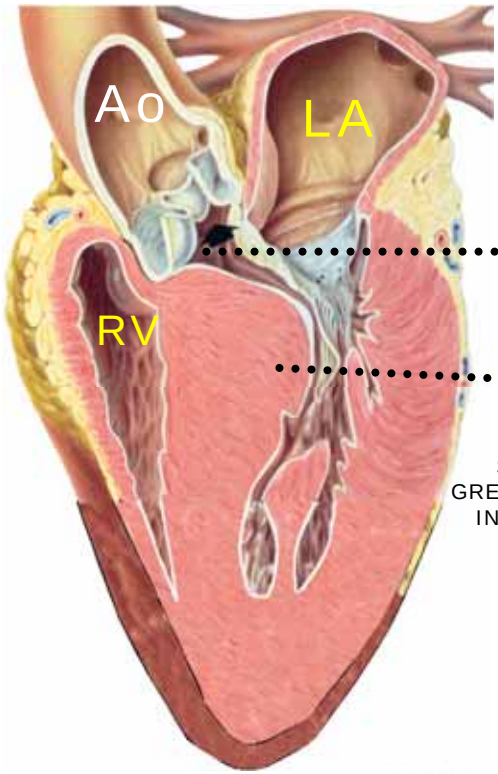
1) OBSTRUCTIVE FORM (HOCM)
Septal asymetrical with obstruction with gradient

2) NON-OBSTRUCTIVE FORM (NOHCM)
A) Septal asymetrical with no obstruction;
B) Apical: 2%, 3% to 8%.
C) Lateral and/or postero-lateral;
D) Concentrical or symmetrical, or homogeneous hypertrophic: 5%.
E) Right ventricle: 2%.

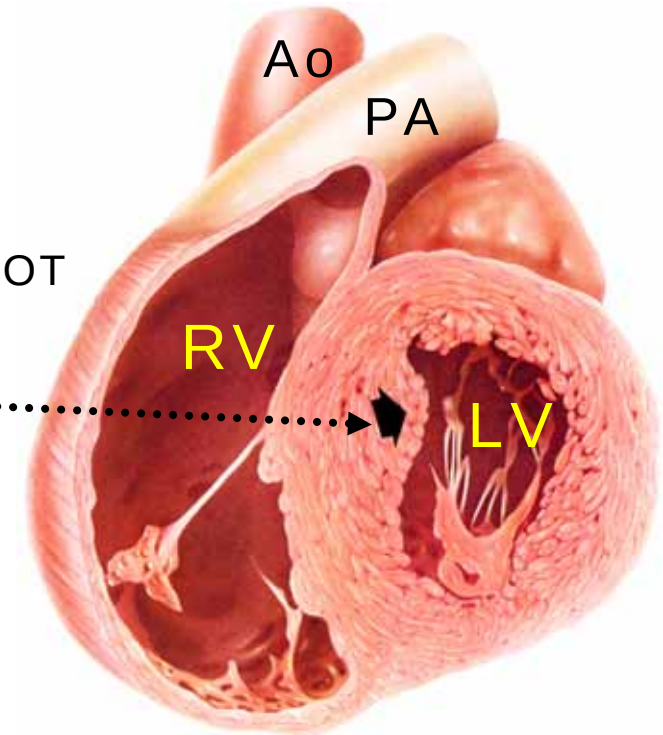
OBSTRUCTIVE FORM (OHCM)	NONOBSTRUCTIVE FORM (NOHCM)
Septum with greater thickness in the superior part (basal).	Septum with greater thickness in the inferior part (apical).
Free wall with progressive decrease of thickness from the base to the apex (the same as normal).	Free wall with no or normal decrease of the thickness from the base to the apex.

Classification and differences of obstructive and nonobstructive forms of Hypertrophic Cardiomyopathy.

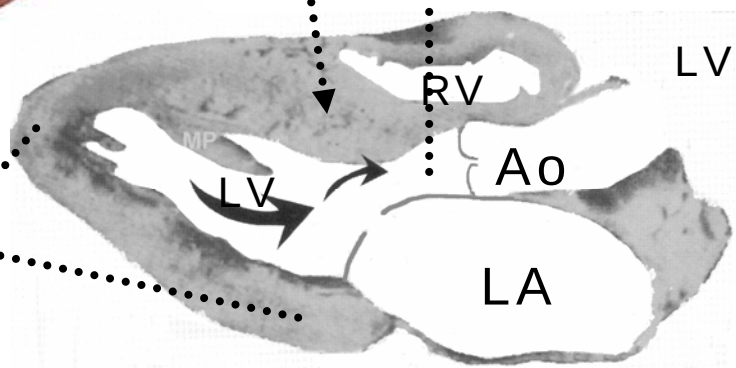
OBSTRUCTIVE FORM OF HYPERTROPHIC CARDIOMYOPATHY (OHCM)



LVOT



SEPTUM WITH GREATER THICKNESS IN THE SUPERIOR PART (BASAL).



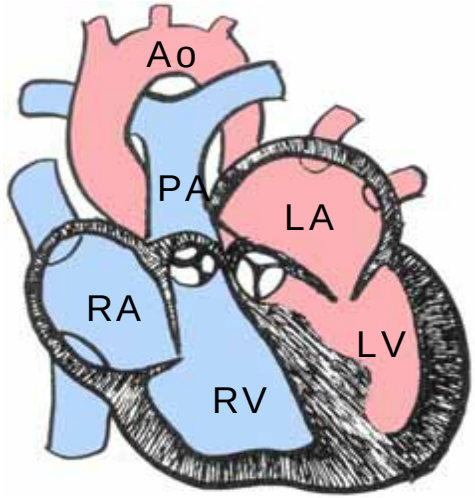
FREE WALL WITH PROGRESSIVE DECREASE OF THICKNESS FROM THE BASE TO THE APEX

LVOT: LV OUTFLOW TRACT

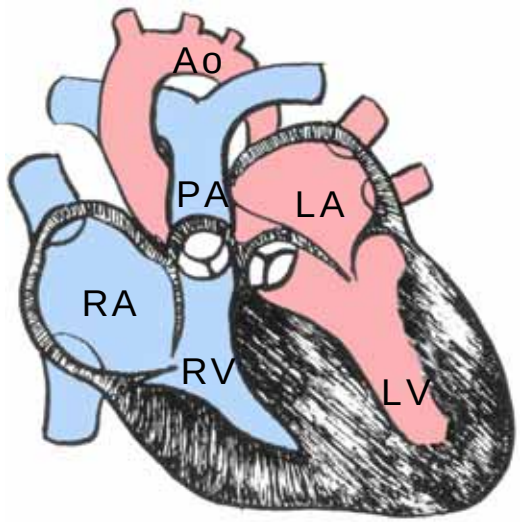
Outline of obstructive form of Hypertrophic Cardiomyopathy.

NONOBSTRUCTIVE FORMS

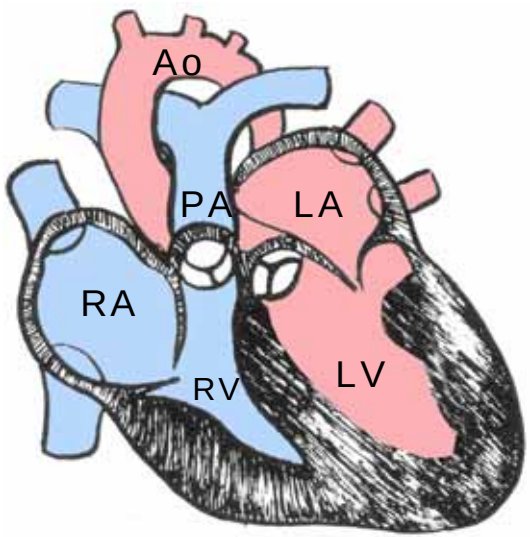
SEPTAL ASYMMETRICAL
WITHOUT OBSTRUCTION



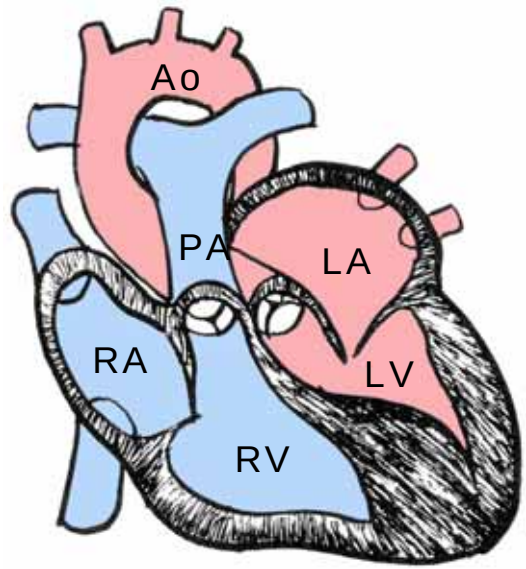
SYMMETRICAL



LATERAL AND/OR
POSTERO-LATERAL



APICAL



Outline of nonobstructive form of Hypertrophic Cardiomyopathy.

ECG IN HYPERTROPHIC CARDIOMYOPATHY

Altered in 90% of cases.

RHYTHM: sinus in most cases. Possible acute AF (10%): < of LV compliance+MI ---------> >LV end of diastole -------> > of medium intra LA pressure --- --->LAE -->AF.

P WAVE: LAE (20%) by >LV end of diastole + < of LV compliance + MI. BAE in dilated phase: ICD. RAE: Berhaim's syndrome, RVOT obstruction.

PR INTERVAL: normal and possibly short: pseudo or true WPW. AV blocks of different degrees and even total AV block.

SAQRS: between 0º and +90º in nonobstructive forms. SAQRS between 0º and –90º is observed in 30% of cases.

QRS: LVE with strain pattern with QRS/T angle near 180º and prominent R waves in intermediary precordial leads in 80%. In 10%, very wide R waves in V1 and aVR associated to deep and "clean" Q waves in V5 and V6 and/or in inferior leads, by > of septal vector 1 in 10%. LAFB (10%) with extreme shift of AQRS (beyond –30º).

LSFB: increasing R wave from V2 to V4 and decreasing from V5 to V6, R wave of V4 of greater voltage than the other precordial leads (74%), absence of q waves in DI (87%) and V5 (91%), anterior shift of QRS loop in the HP (74%) and R vector of posterior and rightward orientation (91%); in familial forms, 50% QS pattern from V1 to V4. In sporadic ones is 15%.

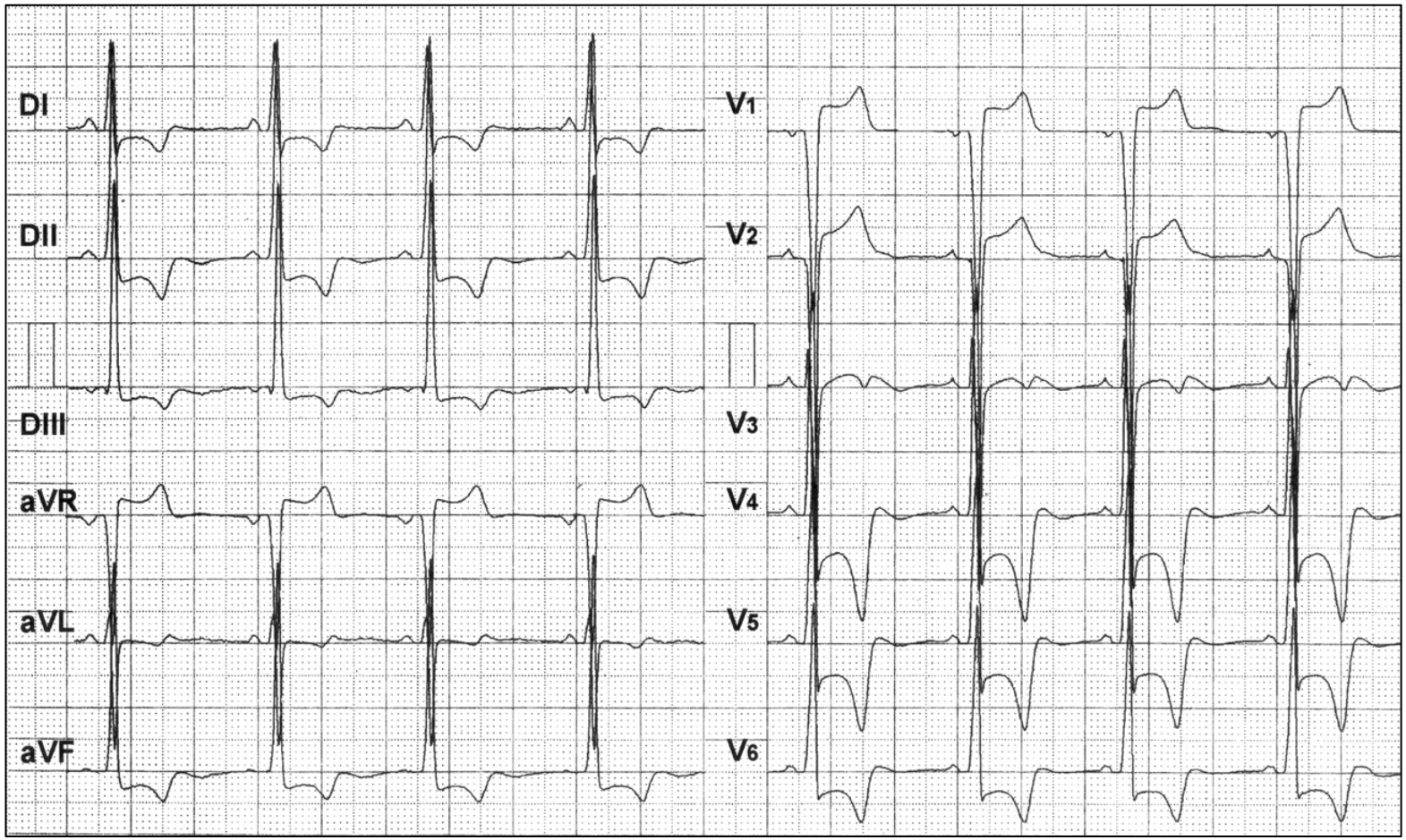
Important Q waves of pseudo infarction, with less duration (<40 ms) and deep. Q waves in young patients with absence of MI history.

Complete LBBB: after transvalvar myotomy/myomectomy surgery (80% of cases);

Complete RBBB: after absolute alcohol injection in the first septal perforating artery of the anterior descending artery (ADA) in percutaneous transluminal septal ablation;

ARRHYTHMIAS: 85% NS-SVT (30%), AF (10%) frequent premature ventricular contractions (>10/h) in 20%, isolated, coupled, (25%) polymorphic (20%), NS-VT and SVT.

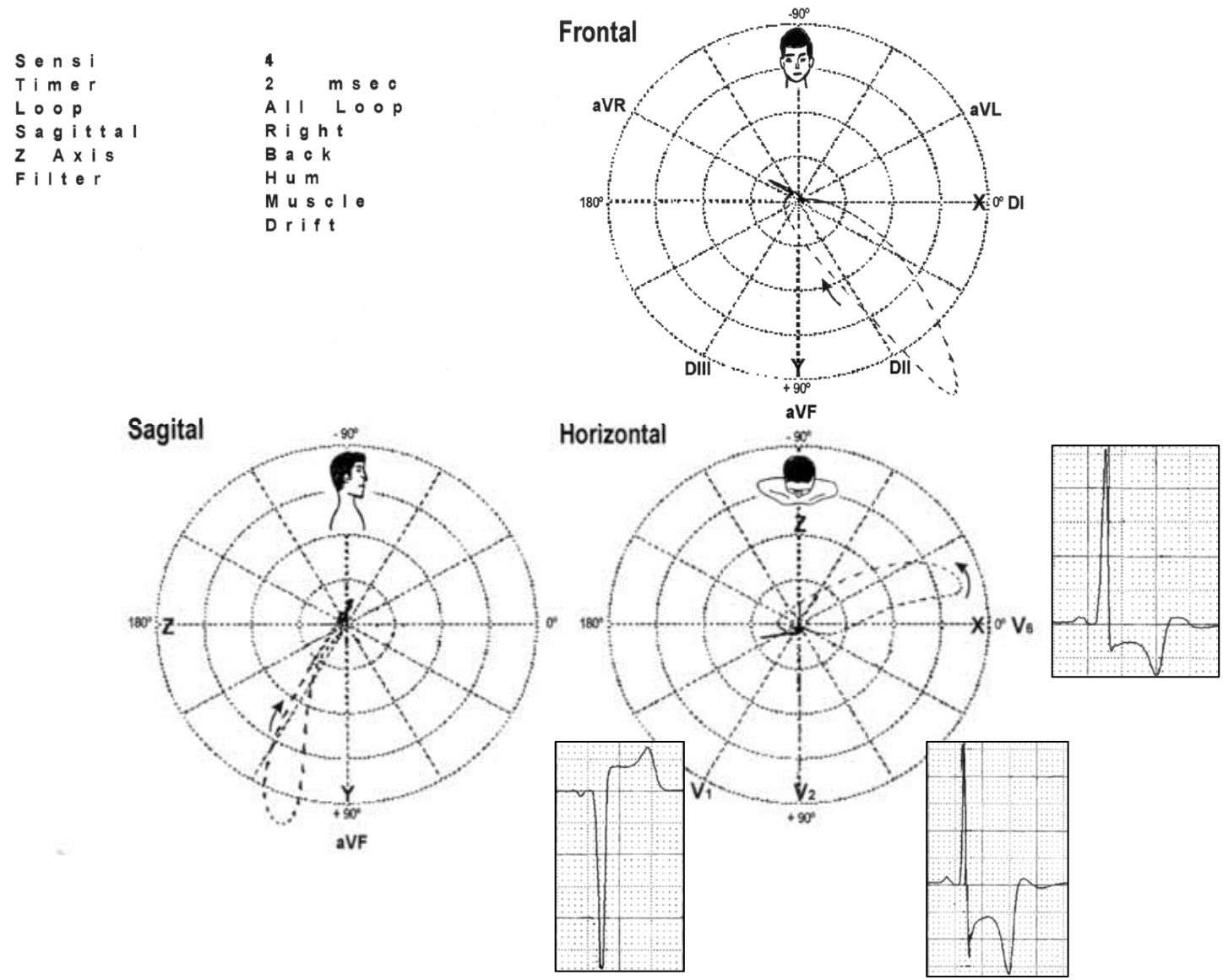
Name: SFS. **Age**: 15 y.o. **Sex**: M. **Race:** W. **Weight**: 70 Kg. **Height**: 1.72 m.
Number: 718. **Date**: 03/31/98 **Medication in use**: beta blockers.



NOHCM. Apical portion of septum with 32 mm of diastolic thickness. LAE, LVE, systolic pattern by significant secondary ventricular repolarization alteration in antero-lateral and inferior wall.

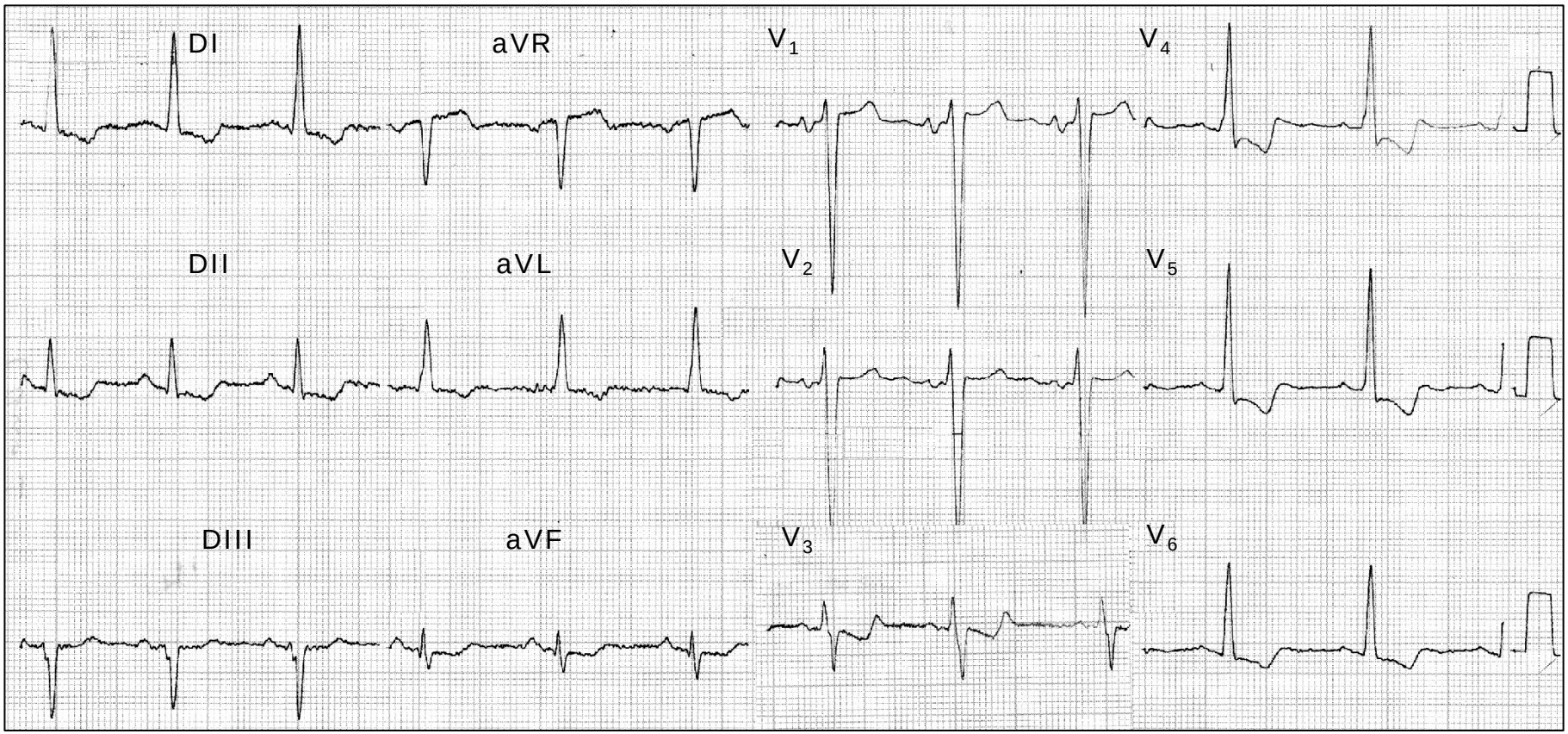
Example of Hypertrophic Cardiomyopathy in its nonobstructive form.

ECG/VCG CORRELATION



VCG of the same patient that displays deep negative T waves.

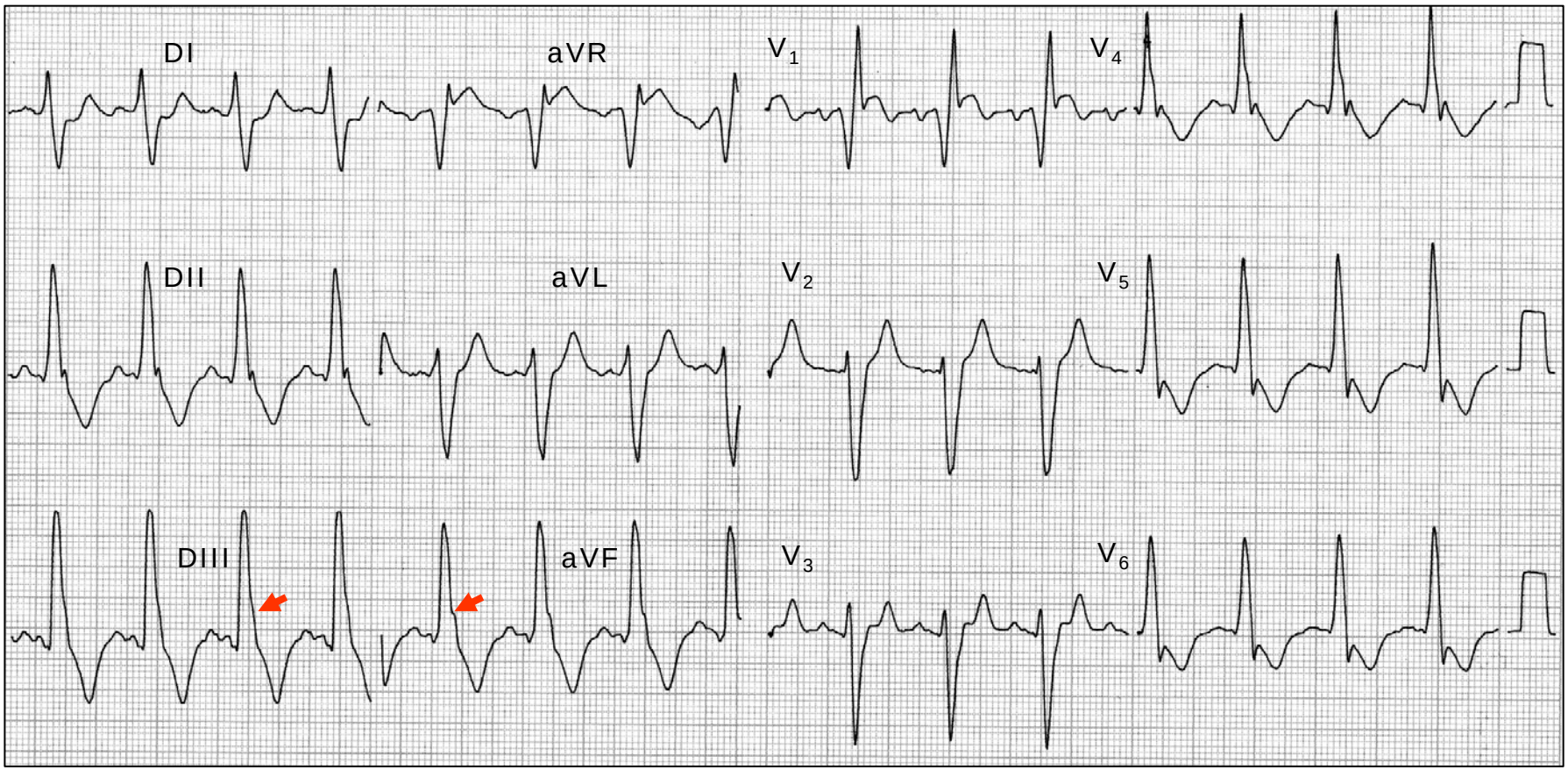
Name: A.V. **Age:** 69 y.o. **S:** M **R:** White **Weight:** 72 Kg.
Height: 1.72 m **Date:** 10/28/1998



Clinical diagnosis: OHCM with gradient in LV outflow tract of 80 mmHg and clinically in functional group IV (dyspnea in rest), even with medication.
ECG diagnosis: left chamber enlargement: LAD+LVE, systolic pattern.
Reduction of hypertrophic septum is chosen, by absolute alcohol injection in the first perforating artery, anterior descending artery branch (percutaneous transluminal ablation).

ECG of Hypertrophic Cardiomyopathy in its obstructive form with severe gradient (systolic pattern of LAE/LVE).

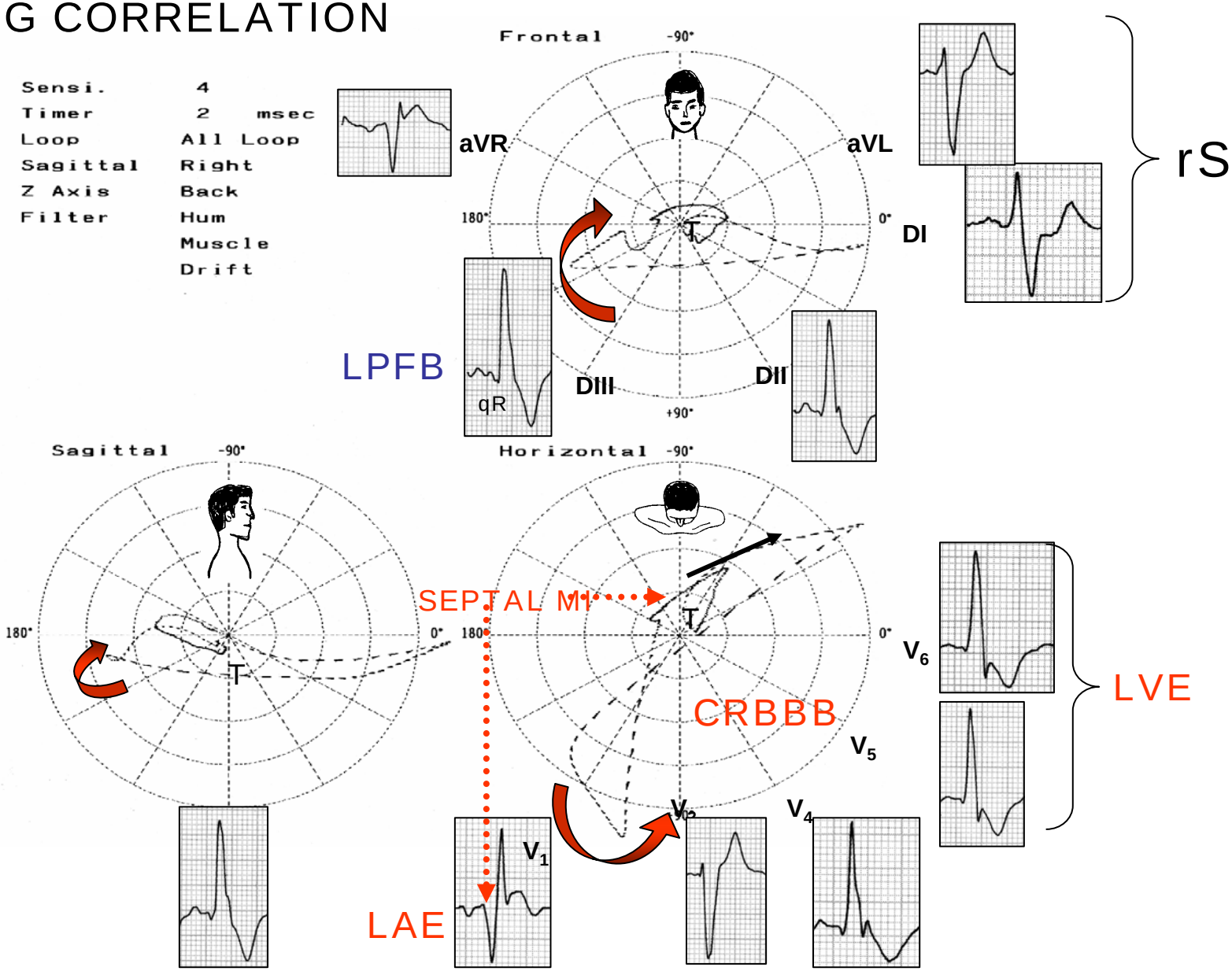
Name: AV **Age:** 69 y.o. **Sex:** male **Race:** white **Weight:** 72 Kg.
Height: 1.72 m **Date**: 10/29/1998 **Drugs in use:** Propanolol 360 mg/day



CLINICAL DATA: the same patient immediately after absolute alcohol injection in the first perforating artery, branch of the anterior descending artery.
ECG: ST + LAE + LVE + CRBBB + LPFB + septal MI: QR in V_1 and ST segment elevation of subepicardial lesion type + LPFB, DI and aVL rS, qR in DIII, RIII>RII, notch in descending limb of R wave of DIII and aVF ( ) and AQRS shifted to the right (+110°).

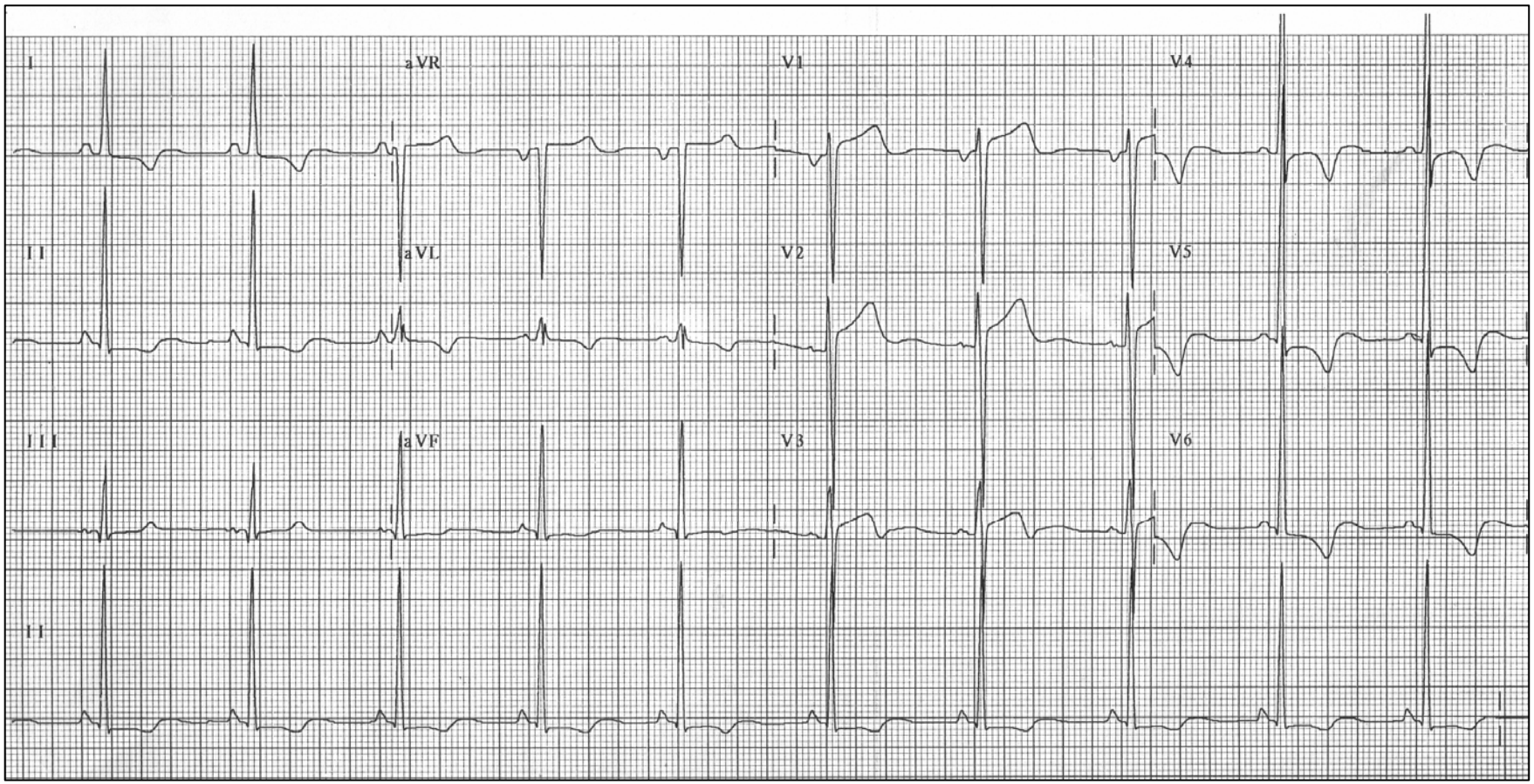
The same patient, in whom absolute alcohol was injected (percutaneous septal alcoholization). Sinus tachycardia + LAE + LVE + CRBBB + LPFB + septal MI.

ECG/VCG CORRELATION



ECG/VCG correlation of the same patient that shows: LAE, LVE, LPFB, CRBBB and septal infarction.

Name: EC **Sex:** Male **Age:** 38 y.o. **Race:** White
Weight: 78 Kg **Height:** 1.70 m **Date:** 05/29/2001
Medication in use: Atenolol 150 mg/day

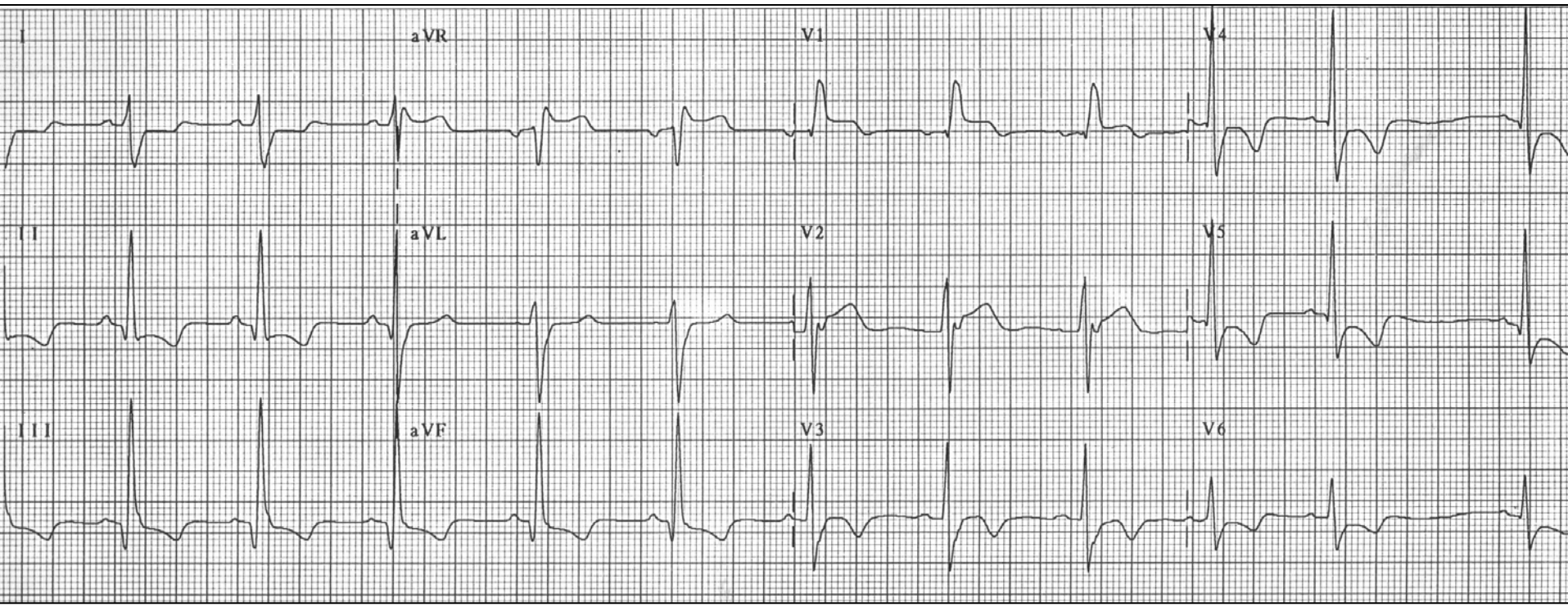


Clinical diagnosis: hypertrophic cardiomyopathy, severe obstructive form, which does not respond to drugs. Septal thickness is 30 mm; gradient in rest is 80 mmHg. Functional class IV.

ECG diagnosis: left chamber enlargement. Systolic pattern of Cabrera of ventricular repolarization ("strain pattern").

ECG of severe Hypertrophic Cardiomyopathy, refractory to medication.

Name: EC **Sex:** Male **Age:** 38 y.o. **Race:** White
Weight: 78 Kg **Height:** 1.70 m **Date:** 05/30/2001
Medication in use: Atenolol 150 mg/day

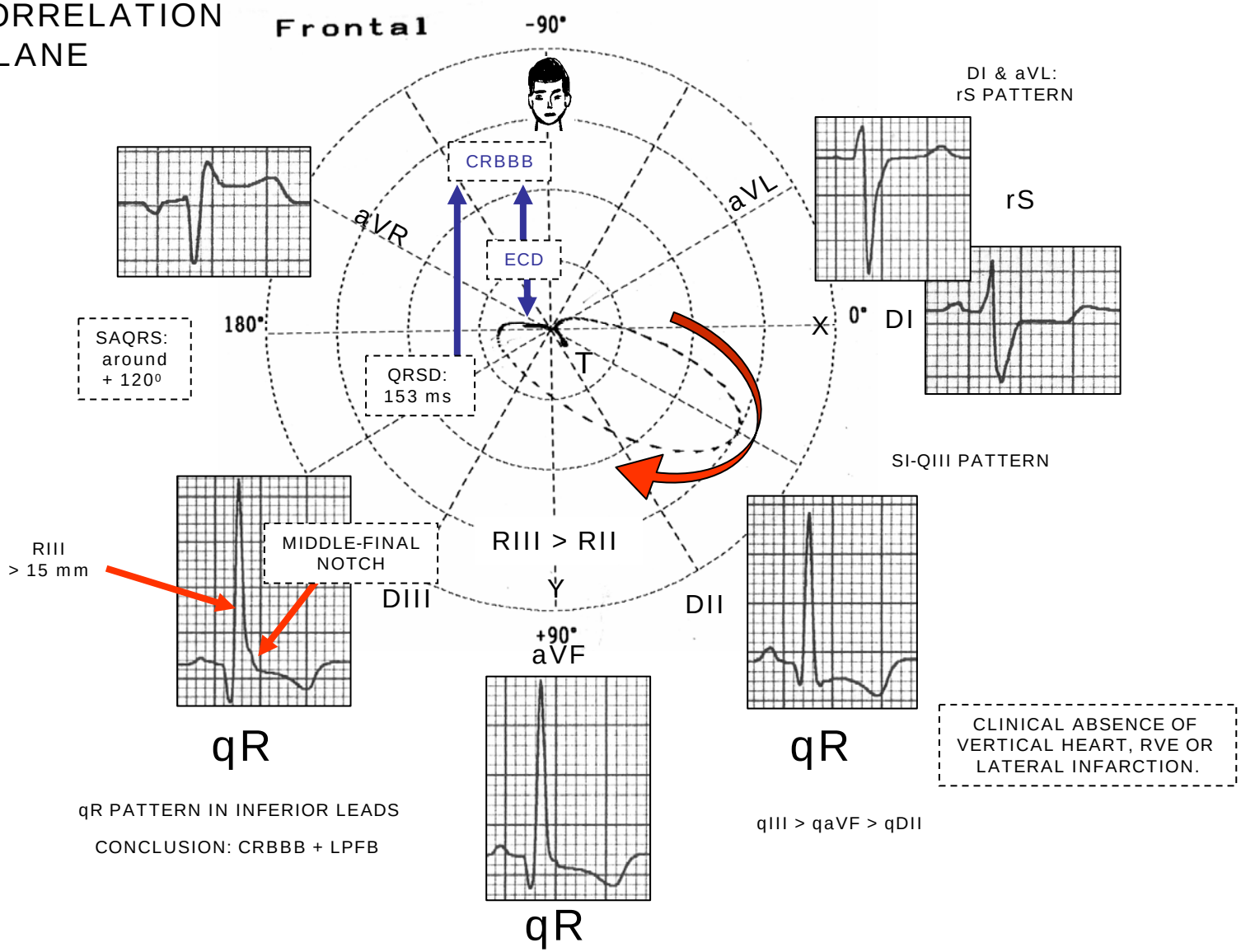


Clinical diagnosis: the same patient after absolute alcohol injection in the first septal perforating artery of the ADA (Percutaneous Septal Ablation). Great relief of dyspnea. Significant reduction of septal thickness.

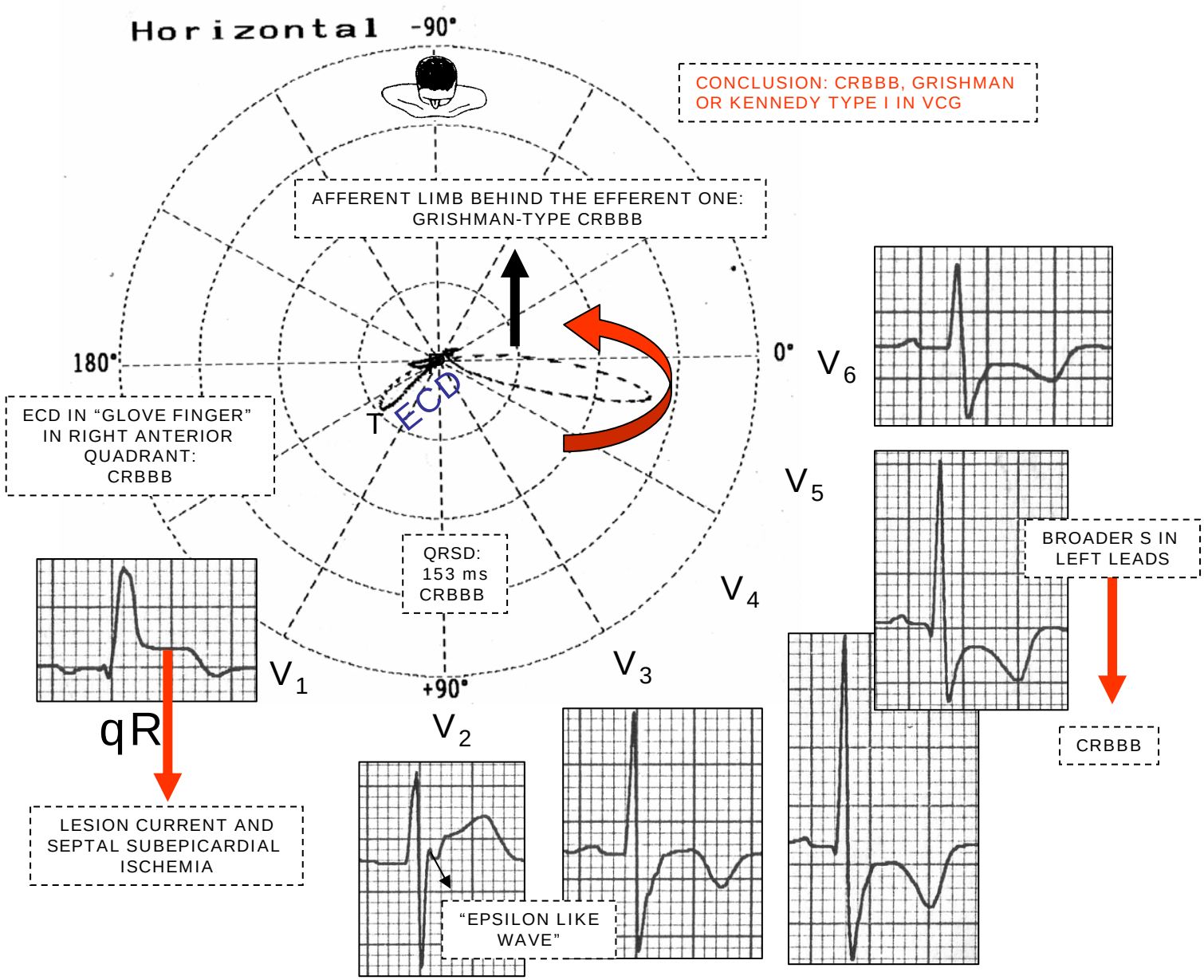
ECG diagnosis: CRBBB + LPFB + lesion current and septal subepicardial ischemia (LV). SAQRS close to $+120^0$; SI-QIII-TIII pattern; RIII > 15 mm RII; qIII > qII > q aVF. Clinical absence of asthenic biotype, RVE or lateral infarction: LPFB. QRSD: 153 ms. Broader S wave in left leads; qR in V1: CRBBB.

The same patient, immediately after alcohol injection: CRBBB + LPFB + septal ischemic lesion.

ECG/VCG CORRELATION
FRONTAL PLANE

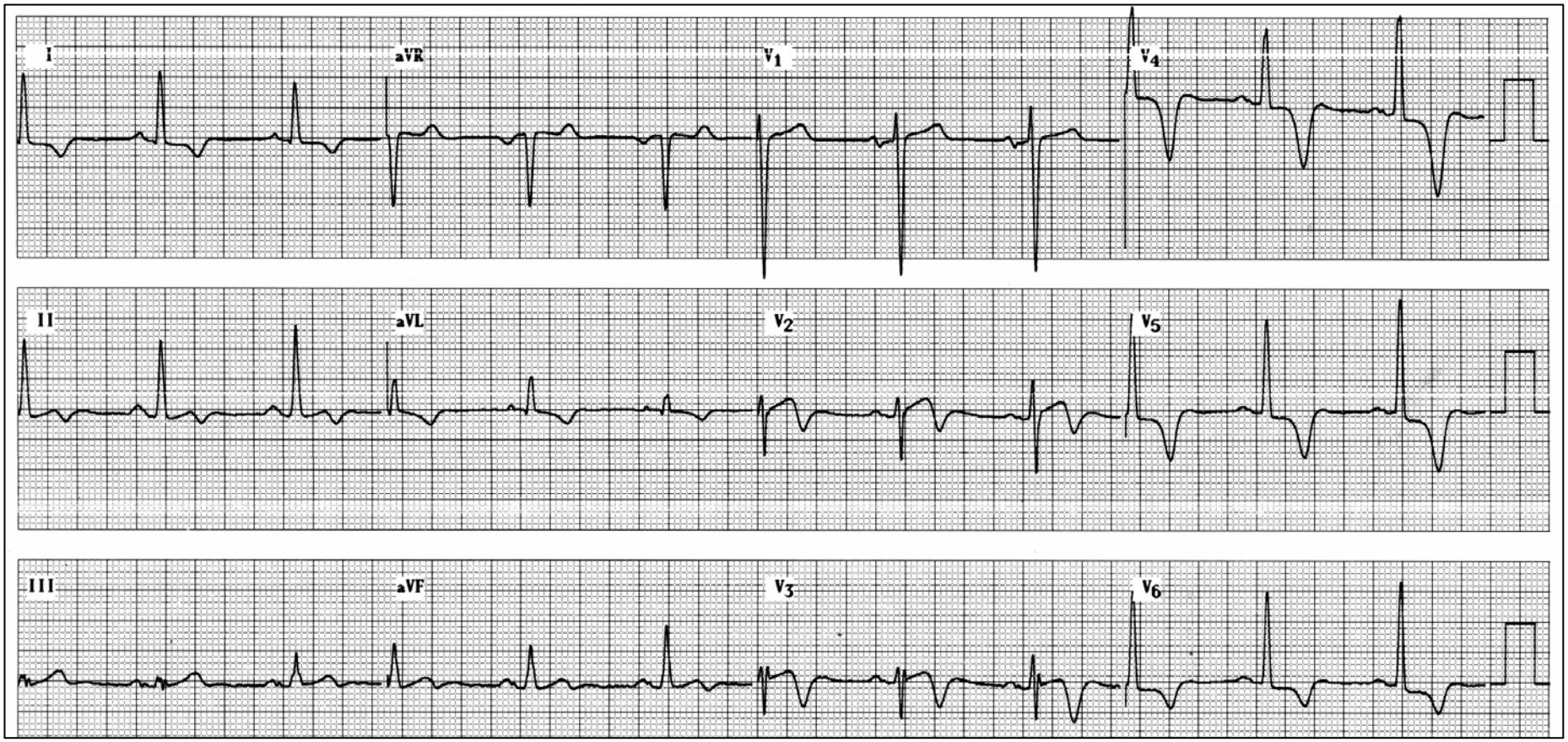


ECG/VCG correlation of the same patient in the frontal plane, which displays: LPFB and CRBBB criteria.



ECG/VCG correlation of the same patient in the horizontal plane, which displays: VCG criteria of Grishman-type CRBBB.

Name: HJS **Sex:** M **Age:** 69 y.o. **Race:** White
Weight: 71 Kg **Height:** 1.80 m **Biotype:** Asthenic **Date:** 03/27/2006

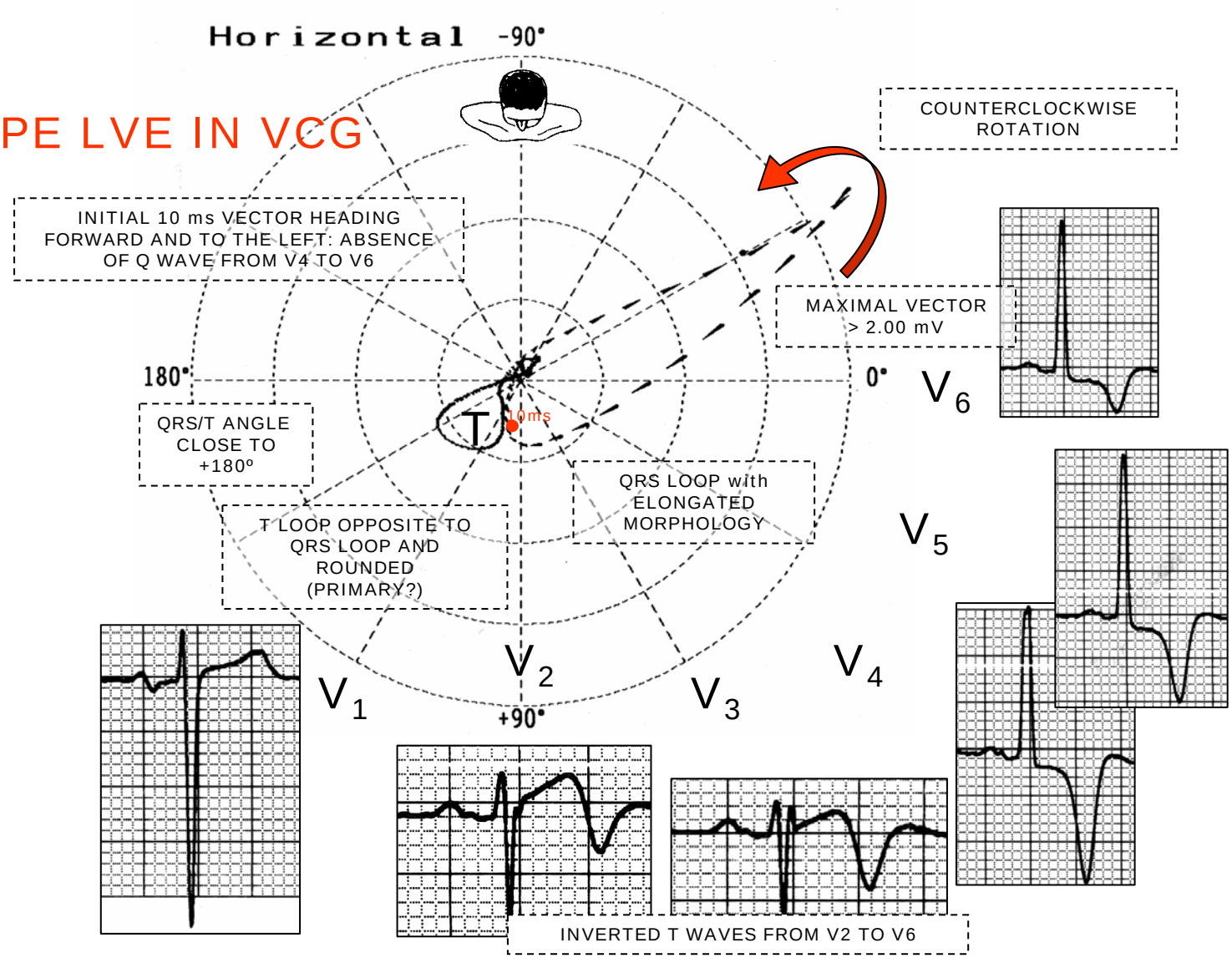


Clinical diagnosis: obstructive Hypertrophic Cardiomyopathy with gradient of 35 mmHg, systemic high blood pressure.
ECG diagnosis: left chamber enlargement, ventricular repolarization systolic pattern. Inverted T waves from V2 to V6 and in DI, DII and aVL. T waves at times with a tendency to symmetry in intermediary leads (V2 to V4).

ECG/VCG correlation of an elderly patient (69 years old), carrier of Hypertrophic Cardiomyopathy in its moderate obstructive form, associated to high blood pressure, pattern of left chambers enlargement, LVE, IB-type vectocardiographic systolic pattern.

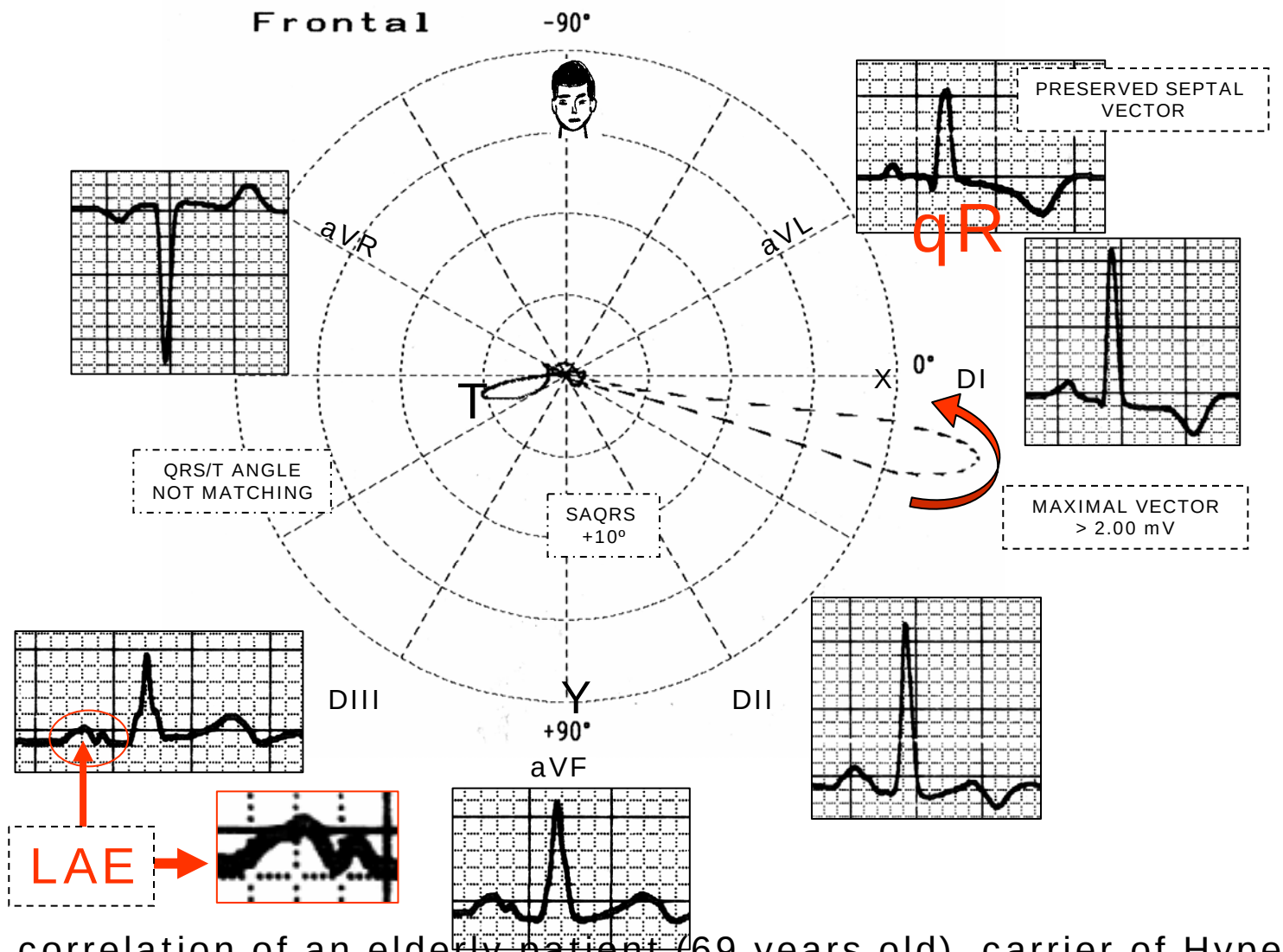
ECG/VCG CORRELATION HORIZONTAL PLANE

IB TYPE LVE IN VCG



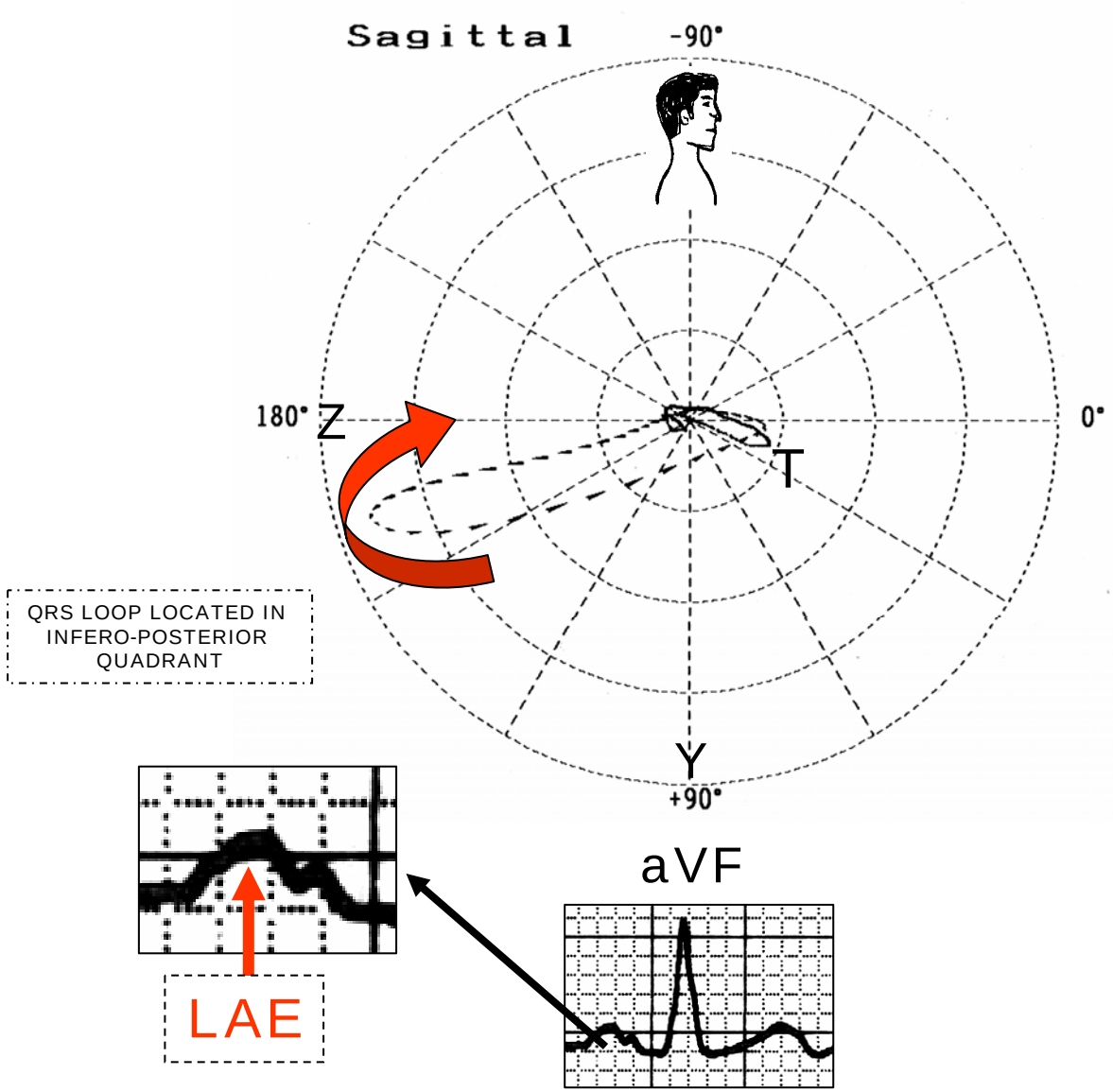
ECG/VCG correlation of an elderly patient (69 years old), carrier of Hypertrophic Cardiomyopathy in its moderate obstructive form, associated to high blood pressure, pattern of left chambers enlargement, LVE, IB-type vectocardiographic systolic pattern.

ECG/VCG CORRELATION FRONTAL PLANE



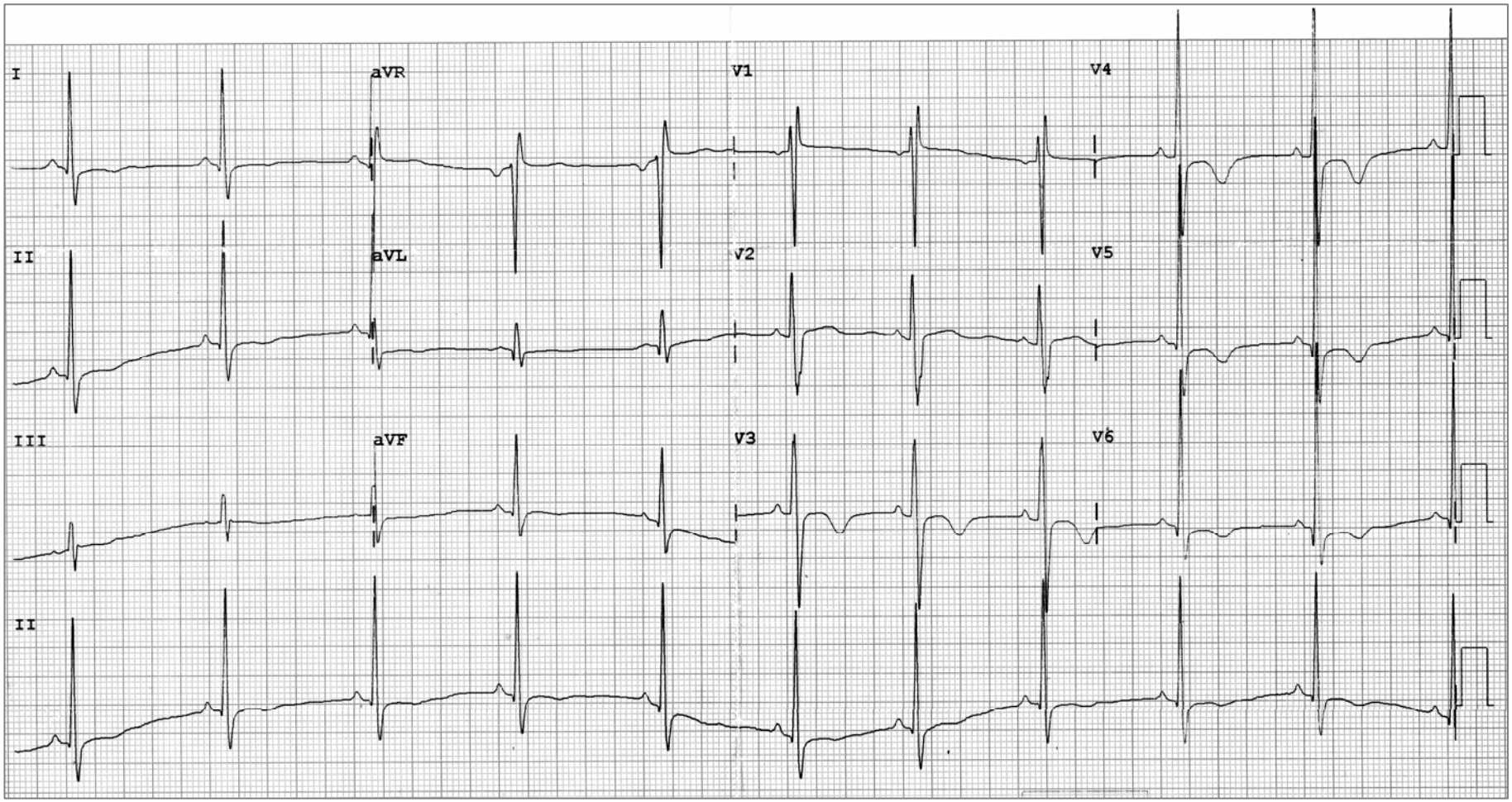
ECG/VCG correlation of an elderly patient (69 years old), carrier of Hypertrophic Cardiomyopathy in its moderate obstructive form, associated to high blood pressure, pattern of left chambers enlargement, LVE, IB-type vectocardiographic systolic pattern.

ECG/VCG CORRELATION RIGHT SAGITTAL PLANE



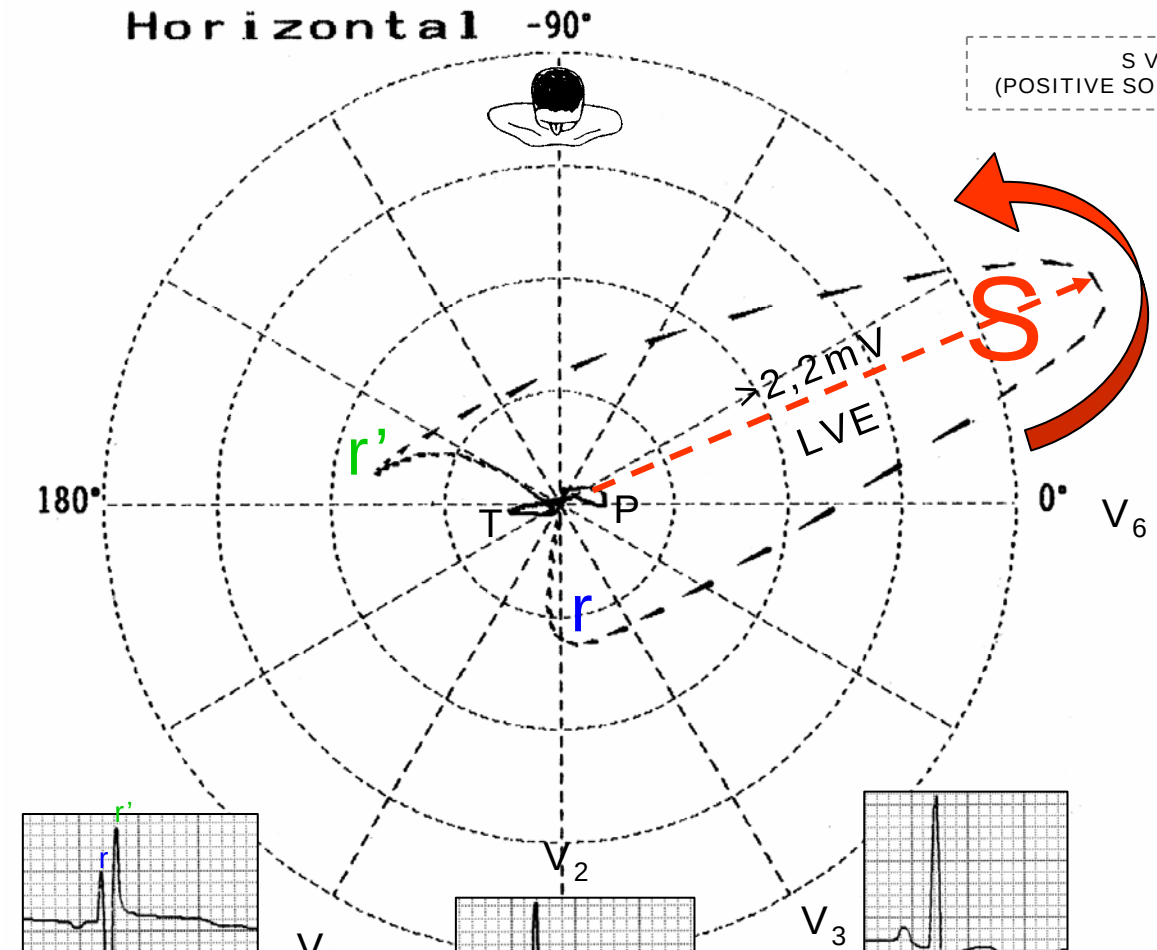
ECG/VCG correlation of an elderly patient (69 years old), carrier of Hypertrophic Cardiomyopathy in its moderate obstructive form, associated to high blood pressure, pattern of left chambers enlargement, LVE, IB-type vectocardiographic systolic pattern.

Name: ANM **Date:** 02/07/2007. **Age:** 26y. **Gender:** M.
Ethnic group: Native American Indian Amazon basin **Weight:** 83 Kg. **Height:** 1.69 m.

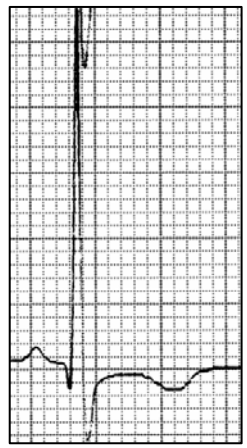


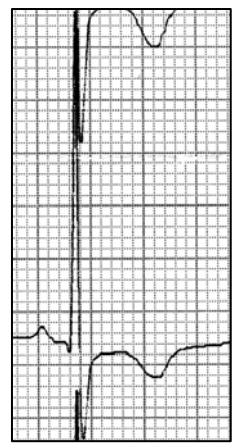
Clinical Diagnosis: Hypertrophic nonobstructive Cardiomyopathy. Inferior wall with segmentar hypertrophy. Systolic dysfunction. Congestive heart failure functional class II (NYHA).
ECG diagnosis: Left ventricular enlargement with diffuse secondary alterations of repolarization. Triphasic rSr' pattern in V1 lead without wide final r' wave: pseudo IRBBB.

ECG/VCG CORRELATION HORIZONTAL PLANE

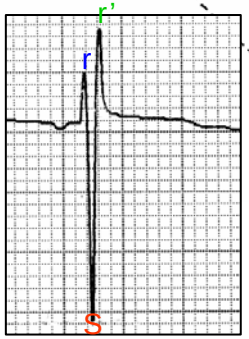


S V_1+R V_5 > 35 mm
(POSITIVE SOKOLOW & LYON CRITERIA)



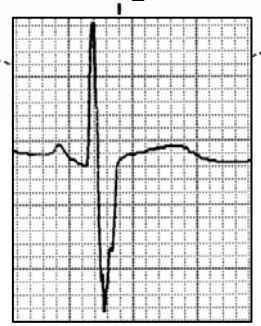


SYSTOLIC PATTERN
OF REPOLARIZATION

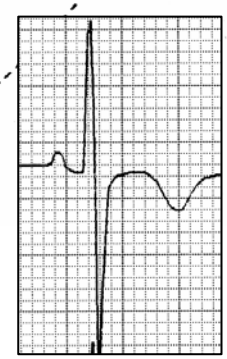


V_1

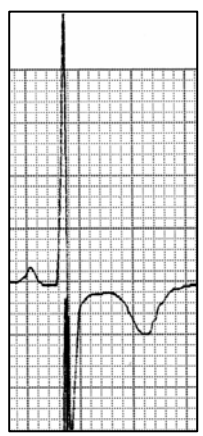
V_2



V_3

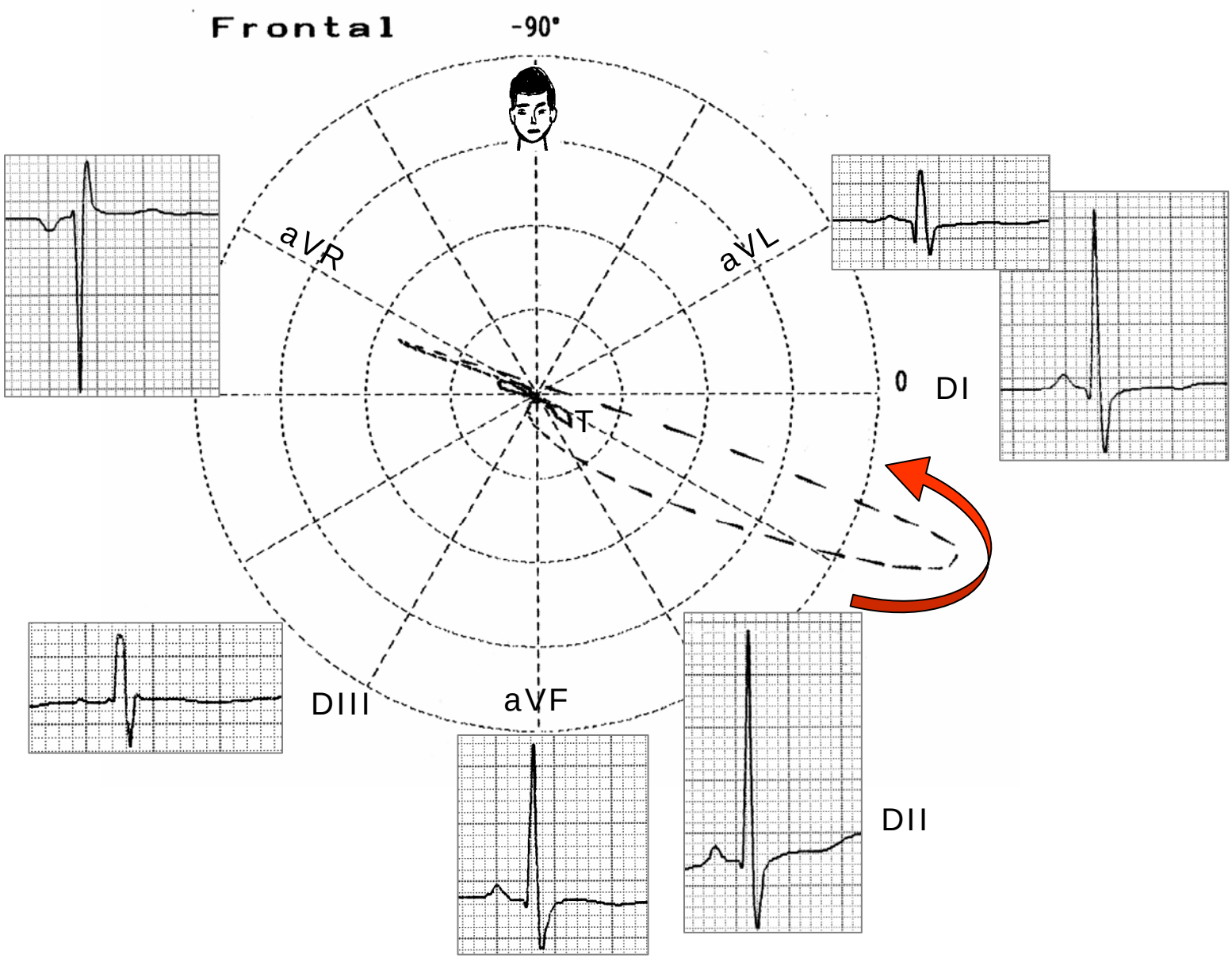


V_4

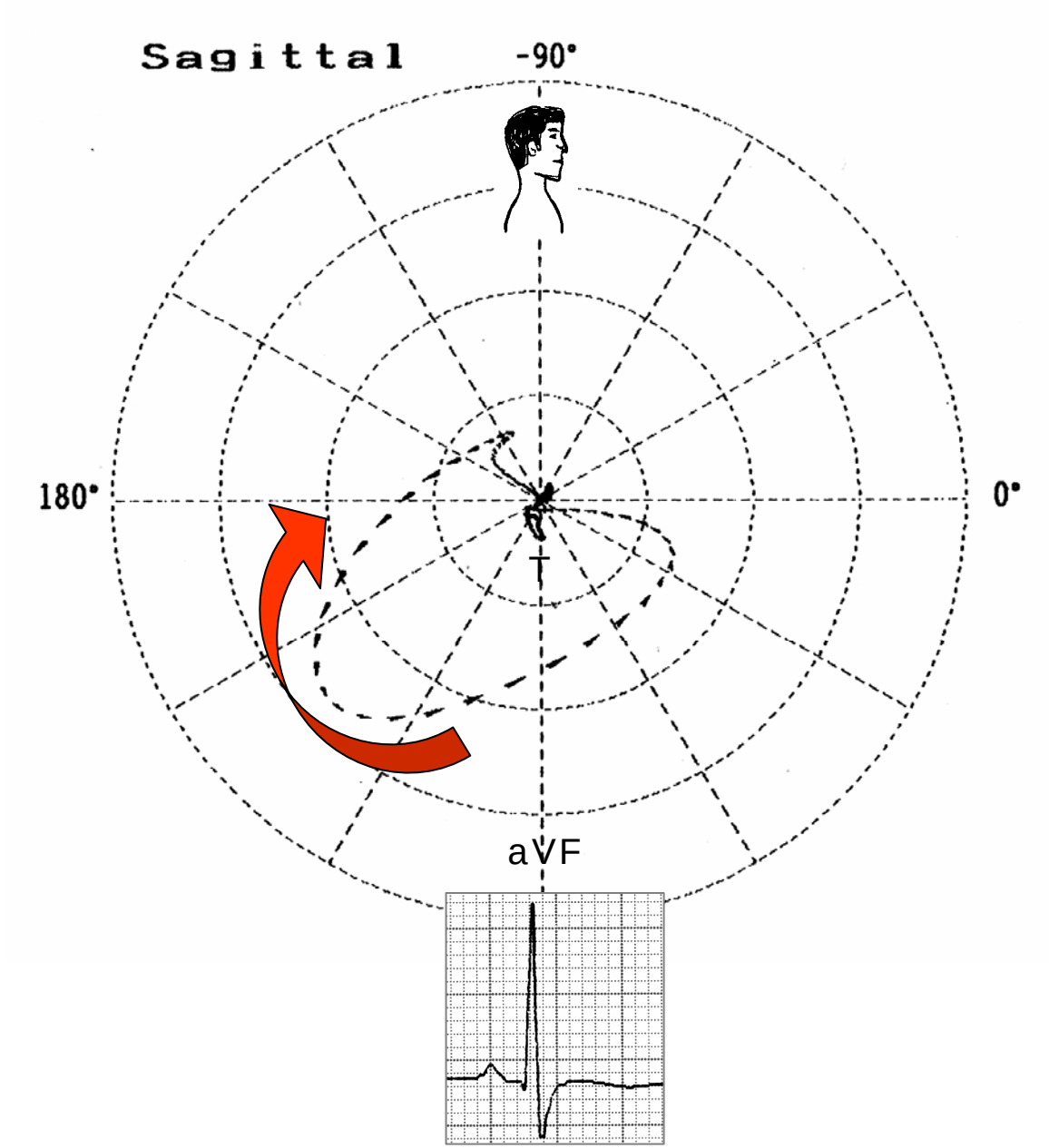


Triphasic rSr' pattern

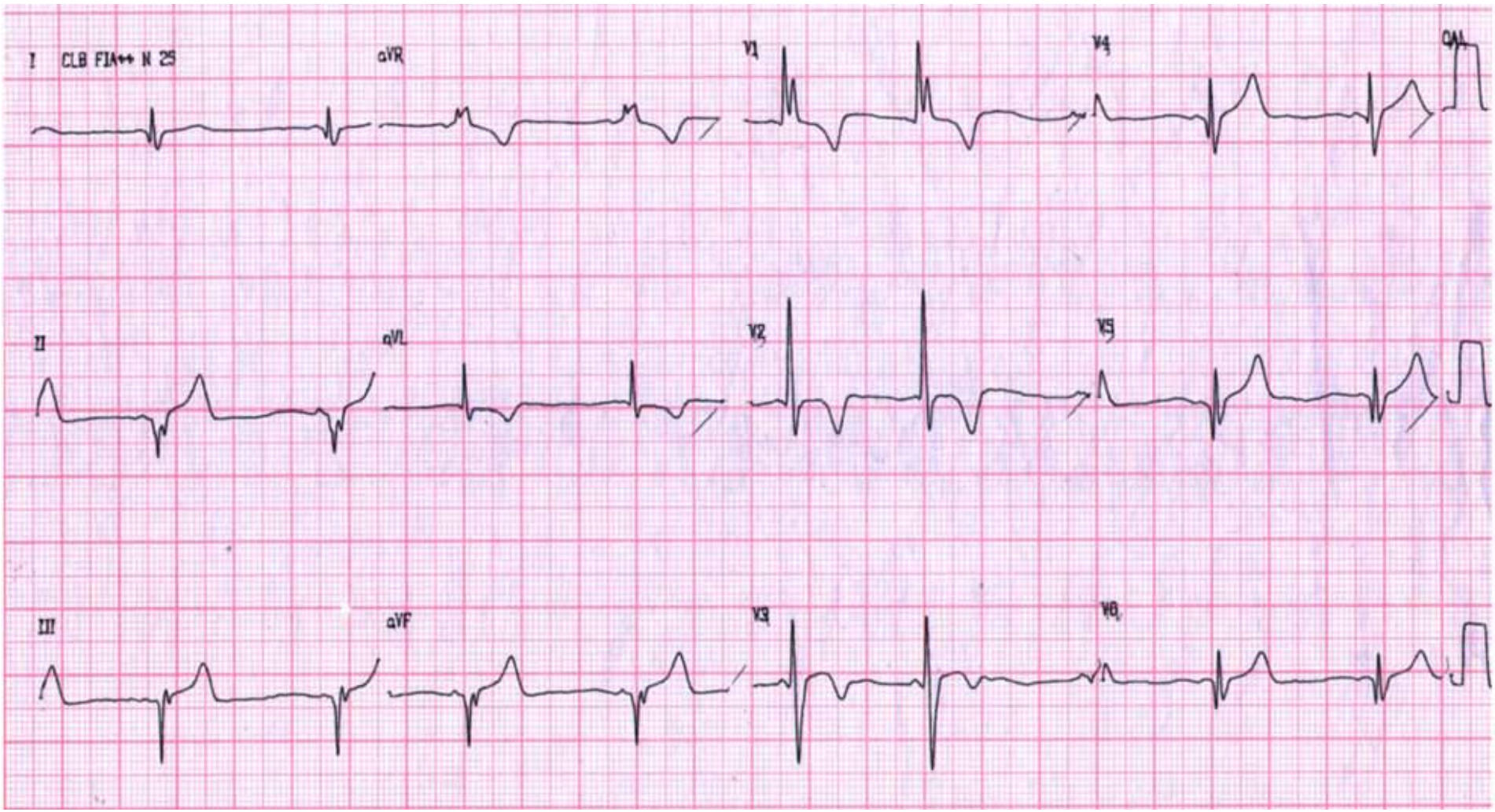
ECG/VCG CORRELATION FRONTAL PLANE



ECG/VCG CORRELATION RIGHT SAGITTAL PLANE



Male, 23-year-old patient, with history of tiredness at strain and syncopal episode.



What is the report ECG? What is the probable etiological diagnosis?

ECG DIAGNOSIS

- **Ectopic atrial rhythym**
- **Prominent Anterior Forces Pseudo infero-latero-dorsal MI**

Etiology ?

Sugestive of Hypertrophic Cardiomyopathy (HCM) by the clinical features: syncope in young man(23-year-old patient) and the typical ECG pattern:

1. Extreme left axis deviation In HCM the QRS axis between 0º and – 90º is observed in 30% of cases
2. Pseudoinfarction pattern in infero-latero-posterior wall
3. In ≈ 10% of cases in HCM is observed a very wide R waves in V1 and aVR associated to deep and “clean” Q waves in V5 and V6 and/or in inferior leads, by > of septal vector 1 in 10%.
4. LAFB (10%) with extreme shift of AQRS (beyond –30º)
5. Important Q waves of pseudo infarction, with less duration (<40 ms) and deep
6. Q waves in young patients with absence of MI history
7. Arrhythmias: 85% NS-SVT (30%), AF (10%) frequent premature ventricular contractions (>10/h) in 20%, isolated, coupled, (25%) polymorphic (20%), NS-VT and S-VT.

RISK FACTORS ASSOCIATED TO SUDDEN DEATH IN HCM AND CONDITIONING FACTORS FOR A WORSE PROGNOSIS

RISK MARKERS USED TO ASSESS THE MAGNITUDE OF RISK

1) Extreme increase of septal thickness: extreme left ventricular (LV) hypertrophy (> 30 mm) in young patients
2) Very increased estimation of myocardial mass
3) Progression of the disease to LV wall thinning and decrease of EF
4) History of recovery from SCD
5) Recurrent syncope in youngster
6) Unexplained (not neurally mediated) syncope, particularly in young patients
7)Nonsustained ventricular tachycardia on Holter electrocardiographic recording
7) Significant bradyarrhythmia or concealed conduction
8) Blood pressure decrease or inadequate increase during upright exercise.
9) Hereditary genetic defect, associated to unfavorable prognosis.

Multiple risk factors convey a definite increase in risk. However, a single risk factor such as family history of multiple sudden deaths, massive LV hypertrophy in a young patient, or frequent and/or prolonged runs of nonsustained ventricular tachycardia on Holter, may also justify consideration of a prophylactic ICD.

Factors associated to sudden death and Hypertrophic Cardiomyopathy.

RISK FACTORS ASSOCIATED TO SUDDEN DEATH IN HCM AND CONDITIONING FACTORS FOR A WORSE PROGNOSIS

Type I HCM: with genetic alteration with mutations in locus 1q of the long arm of chromosome 14, which alters the heavy chain of cardiac β-myosin (β-MyHC), high penetrance, severe hypertrophy and sudden cardiac death present in approximately 50% of affected patients.

The locations Arg403 (substitution of the amino acid arginine by glycine in position 403), Arg453Cys (substitution of the amino acid arginine by cysteine in position 453), and Arg719Trp (substitution of the amino acid argynine by tryptophan in position 719) are considered malignant.

Type II HCM: (15%) alteration in chromosome 1: locus 1q3. It modifies cardiac troponin T (cTnT). These patients present little hypertrophy and high arrhythmic mortality in youngsters under 30 years old. To this moment, 8 mutations have been described.

NOTE: in patients in whom a genetic diagnosis has been made of malignant form, even in absence of symptoms and hypertrophy, implantable cardioverter defibrillator is indicated.

Factors associated to sudden death and Hypertrophic Cardiomyopathy.

RISK FACTORS ASSOCIATED TO SUDDEN DEATH IN HCM AND CONDITIONING FACTORS FOR A WORSE PROGNOSIS

- Atrial fibrillation;
- Presence of NS-VT in Holter in patient with alteration of conscience;
- S-VT induction in electrophysiological study;
- History of associated infarction;
- Myocardial ischemia, especially in young patient that presents alteration of conscience or hypotension induced by strain;
- Calcification of mitral ring;
- Association with high blood pressure.

Factors associated to sudden death and Hypertrophic Cardiomyopathy.

VECTOCARDIOGRAM OF HCM

According to Ellison and Restieaux, the method displays:

a) Usefulness to quantify severity;

b) Usefulness to estimate the magnitude of left ventricular mass (normal 50 to 90 g/m^2 in children and young adults).

The following vectocardiographic elements stand out:

Vectocardiogram features in Hypertrophic Cardiomyopathy.

VECTOCARDIOGRAM OF HCM

1) INCREASED VOLTAGE OF LV MAXIMAL SPATIAL VECTOR IN QRS LOOP:

Normal values: HP = 1.35 mv (0.75 to 2.2 mv)
FP = 1.55 mv (0.9 to 2.3 mV)
SP = 1.5 mv (0.5 to 2.3 mV)

The degree of this increase is directly related to ventricular mass. Thus, values of 3 mV correspond to the left ventricular mass of 150 g/m^2, values of 4 mV correspond to a mass of 275 g/m^2 and 5 mV and are equivalent to left ventricular mass of 400 g/m^2.

Vectocardiogram features in Hypertrophic Cardiomyopathy.

VECTOCARDIOGRAM OF HCM

2) T LOOP OPPOSITE TO QRS LOOP:

This element is an indication of severity in HCM; thus, the greater the severity, the more obtuse the QRS-T angle (normal maximal value of QRS-T angle in the three planes = 75º). In children and young people, the QRS-T angle is usually normal.

In adults almost always exceeding 75°.

In severe forms and in elderly patients, the angle is close to 180º.

Vectocardiogram features in Hypertrophic Cardiomyopathy.

VECTOCARDIOGRAM OF HCM

3) DROMOTROPIC DISORDERS: Not infrequent:

LAFB: maybe by inclusion of anterior fascicle in the septal muscle with disorder.

LSFB: particularly in NOHCM of the middle and low regions of the septum (see ECG/VCG number 200). Marked anterior shift of QRS loop is observed in the HP in absence of initial convexity to the right of the 20 ms vector.

CLBBB: it is very frequent after septectomy surgery (80% of cases).

CRBBB: it is very frequent after Percutaneous Septal Ablation with absolute alcohol injected in the first septal perforating artery of the ADA.

Vectocardiogram features in Hypertrophic Cardiomyopathy.

VECTOCARDIOGRAM OF APICAL NOHCM

In the apical form we find Type IV vectocardiographic loop of LVE of our classification; nevertheless, it is not exclusive, since other non-apical forms of HCM may display this pattern.

This curious form of HCM is characterized by presenting:

1) Initial vectors of QRS loop heading to the front and the left;
2) Anteriorization of QRS loop, predominantly located in the left anterior quadrant;
3) Maximal vector that increases voltage;
4) Final vectors located to the right and backward, with ST/T vector in the right posterior quadrant. This is the only case of LVE without coronary insufficiency associated to T loop in this quadrant. A curious fact is constituted by the association of apical infarction and T loop in right posterior position.
5) E point that does not match the 0 point, and located backward and to the right from the latter.

Vectocardiogram features in Hypertrophic Cardiomyopathy.

VECTOCARDIOGRAM OF APICAL NOHCM

NOTE: the important posterior and rightward shift of the ST/T vector is responsible for the characteristic giant negative T wave (>10 mm) in the leads of the horizontal plane from V_2 to V_5. T waves at the onset, may not present a significant voltage and may appear later with the evolution of the disease[1].

Stress test may decrease the depth of T waves[2].

1) Bielli M, et al. G Ital Cardiol. 1991;21:1325-1329.
2) Tilmant PY, et al. Arch Mal Coeur Vaiss. 1980;73:1269-1278.

Vectocardiogram features in Hypertrophic Cardiomyopathy.

VECTOCARDIOGRAM OF APICAL NOHCM

Three hypotheses aroused to explain these negative T waves:

a) Apical subendocardial ischemia;
b) Apical cell disorder;
c) Greater duration of action potential of hypertrophied cells, thus conditioning the area to have a slower repolarization[1].

The prevalence in the western world of this form of HCM is approximately 0.02 to 0.2% and it constitutes 8% of the cases of the entity. In Japan, the apical form constitutes 25% of HCM[2].

1) Tsunakawa H, et al. Jpn Heart J. 1991;32:799-809.
2) Maron BJ. J Am Coll Cardiol. 1990;15:972-973.

Vectocardiogram features in Hypertrophic Cardiomyopathy.

VECTOCARDIOGRAM OF APICAL NOHCM

The diagnosis is based on the following elements:

1) Giant and negative T waves from V_2 to V_4;

2) Mild symptoms and benign course;

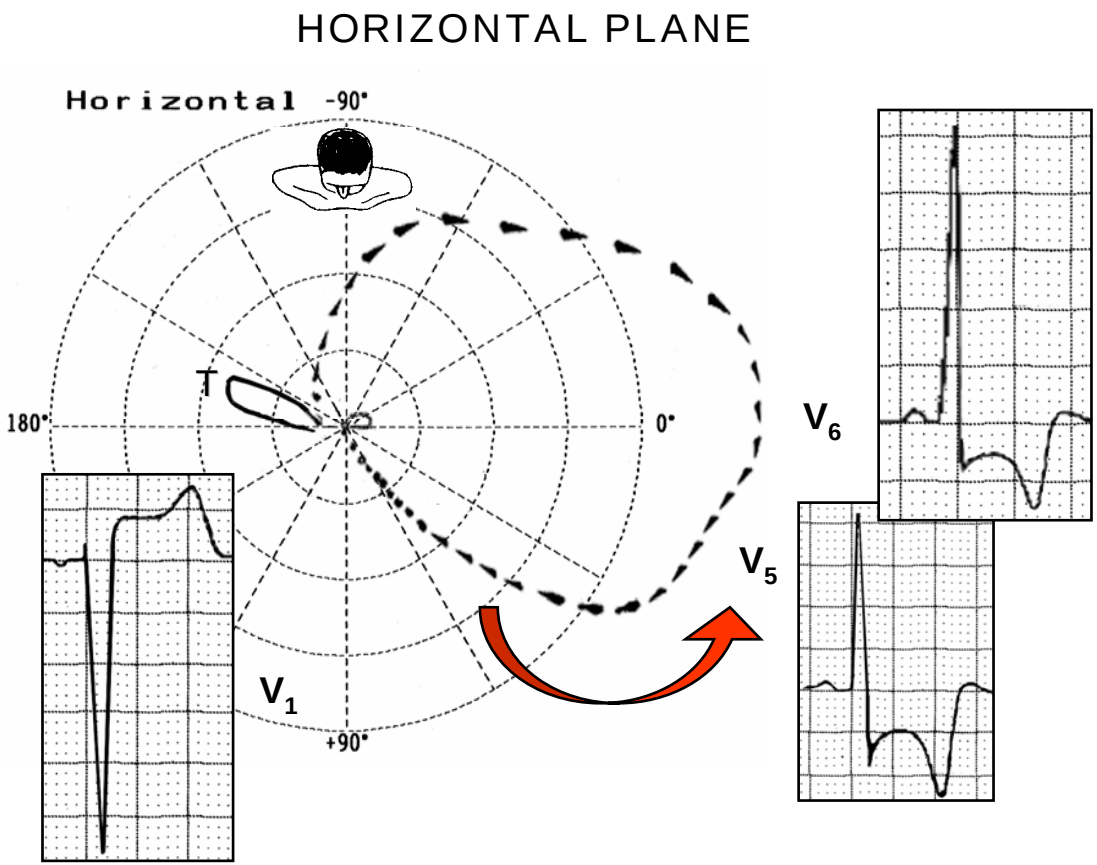
3) Aspect of spade cards in left ventriculography;

4) Absence of ventricular gradient.

It is very important to highlight that incidence increases significantly the more advanced the age of the group under study, since typical ECG manifestations may appear late and with evolution.

Vectocardiogram features in Hypertrophic Cardiomyopathy.

VECTOCARDIOGRAM OF APICAL NOHCM: VECTOCARDIOGRAPHIC IV TYPE LVE



1) Initial vectors of QRS loop heading forward and to the left;

2) Anteriorization of QRS loop predominantly located in the left anterior quadrant;

3) Maximal vector that increases voltage;

4) Final vectors located to the right and backward with ST/T vector in the right posterior quadrant;

5) E point that does not match the 0 point and is located backward and rightward from the latter.

Vectocardiogram features in Hypertrophic Cardiomyopathy.