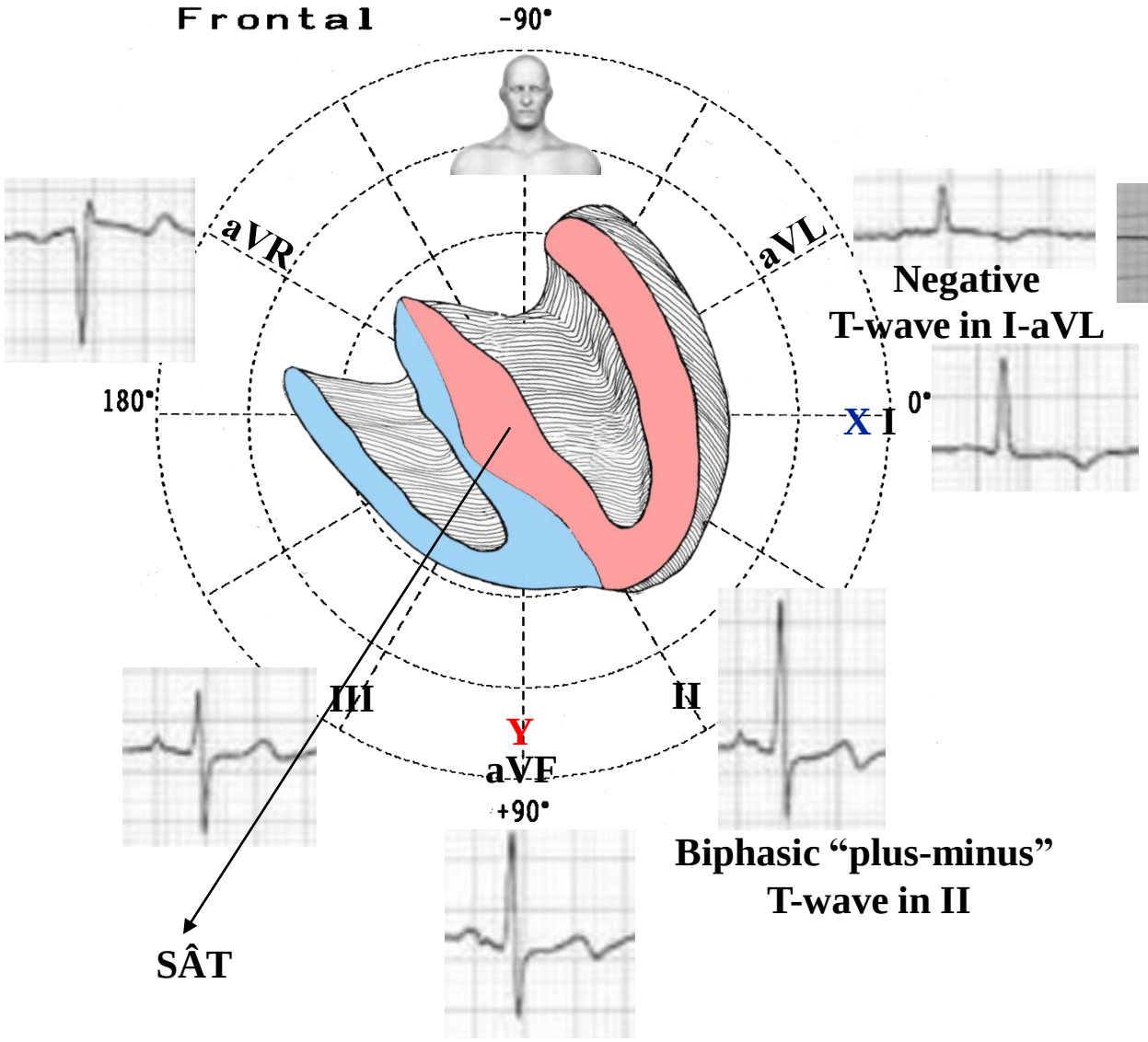
A 68 year old woman was referred to our cardiology department for precordial repetitive chest pain. and an abnormal ECG. She had a history of diabetes mellitus, hypertension, and hypercholesterolemia. She denied or dyspnoea on exertion. Except for a raised blood pressure of 165/70 mm Hg and a systolic murmur with the punctum maximum at the second right intercostal space, physical examination revealed no further abnormalities. The ECG showed a sinus rhythm with T wave inversion in the inferior and anterolateral leads with negative T waves fromV3–V6 (panel A).

The echocardiogram Normal.

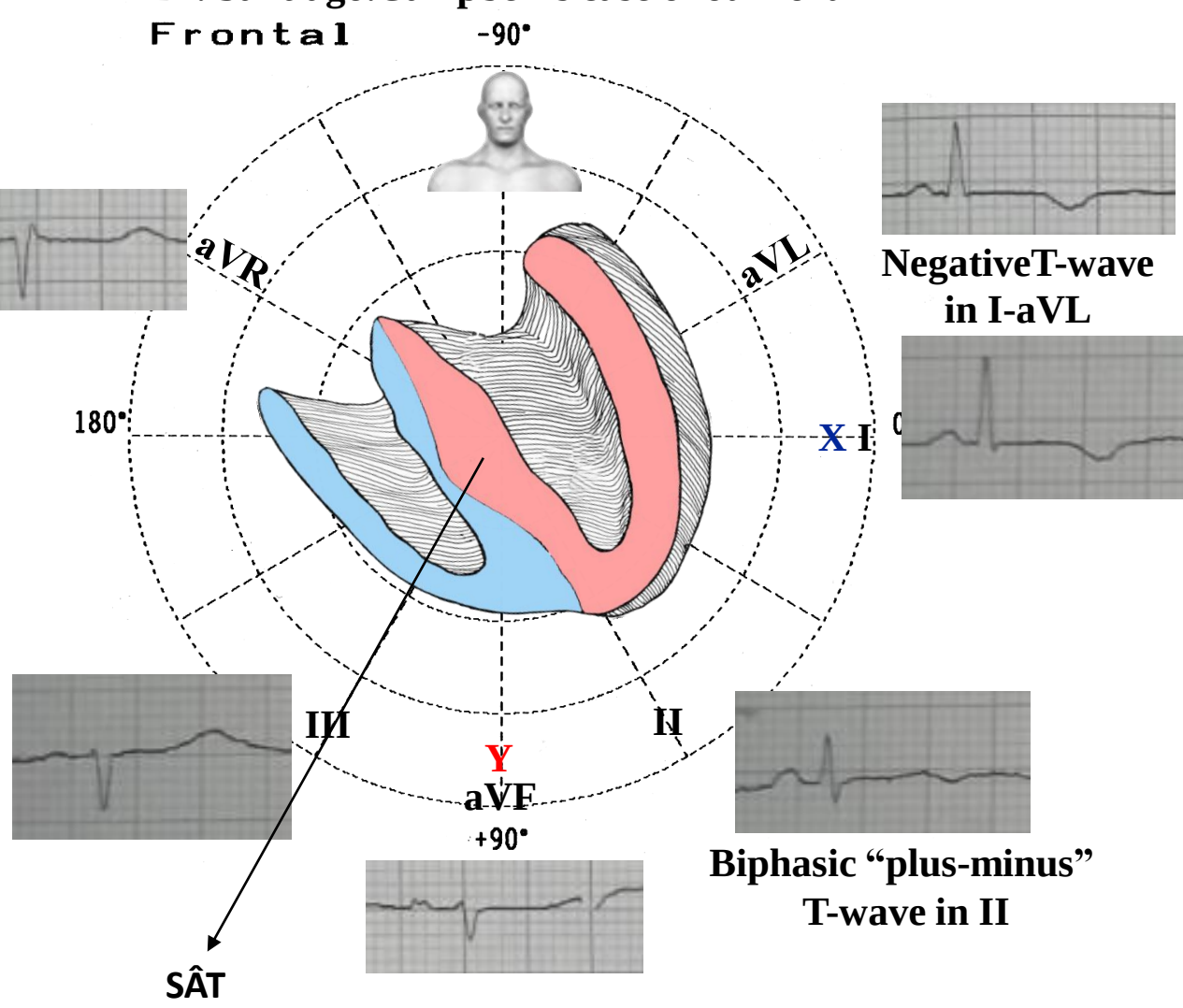
CMRI: Ap-HCM

Frontal plane

The presente case report

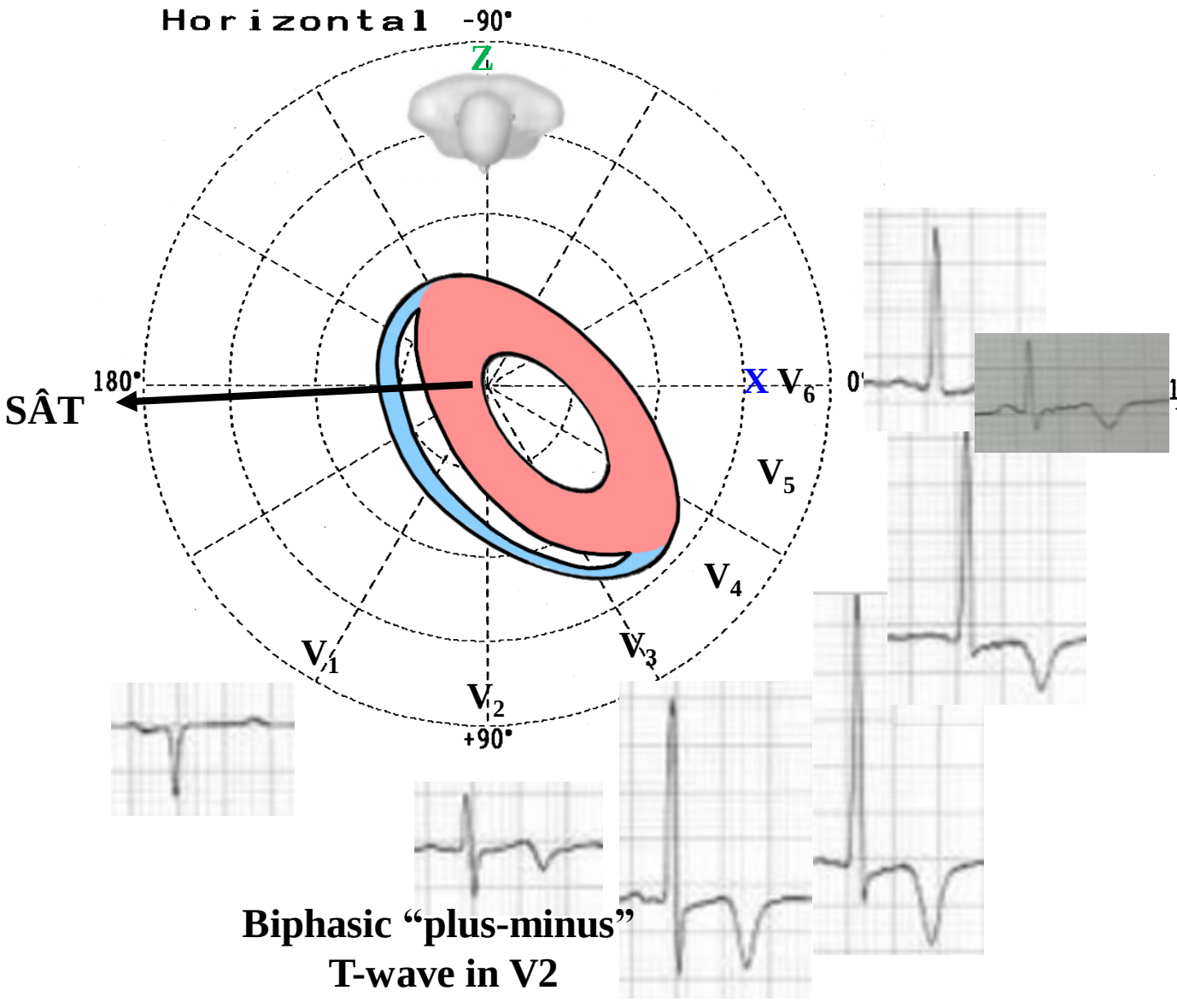


Dr. Santiago.Campbell´s case of our fórum



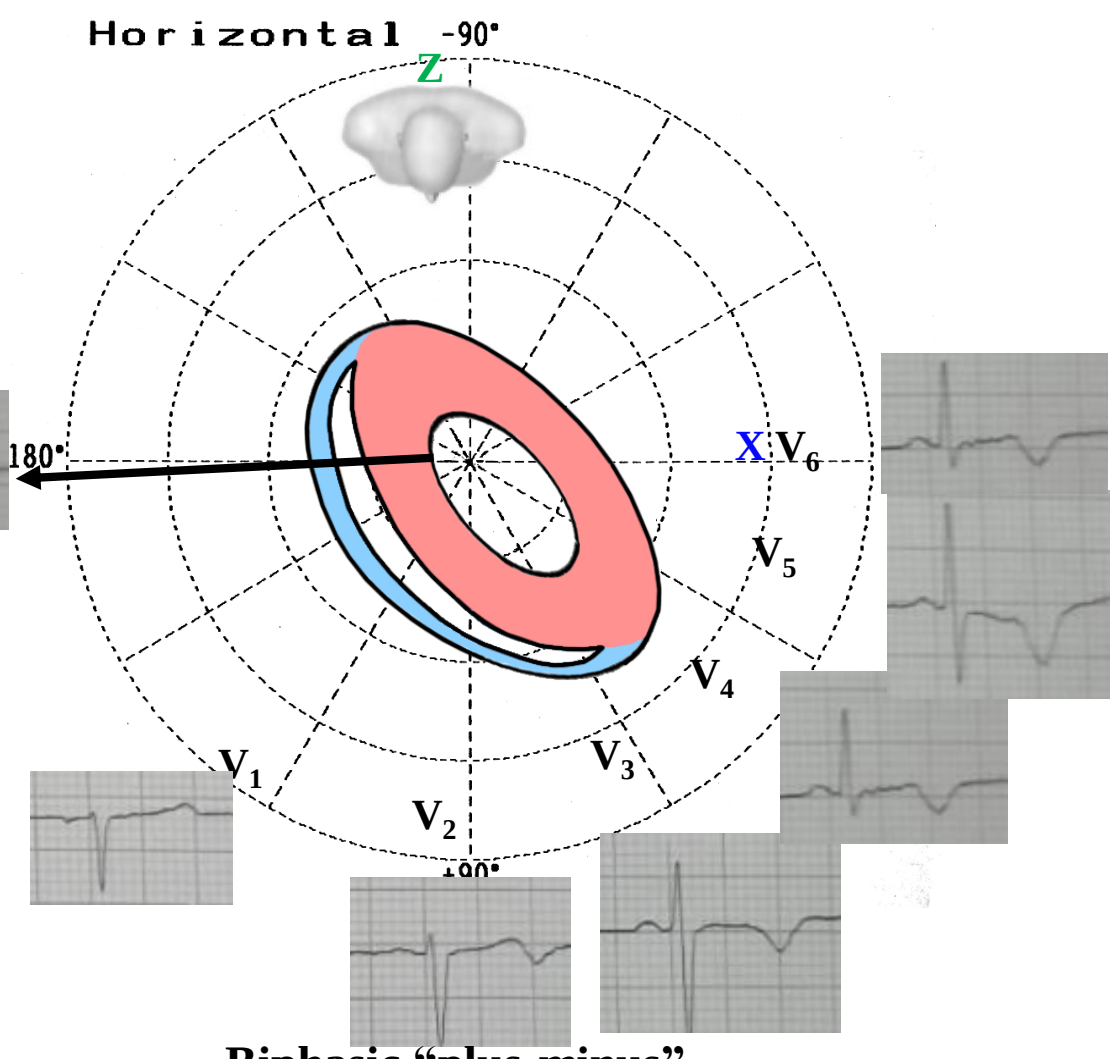
Frontal plane

The presente case report

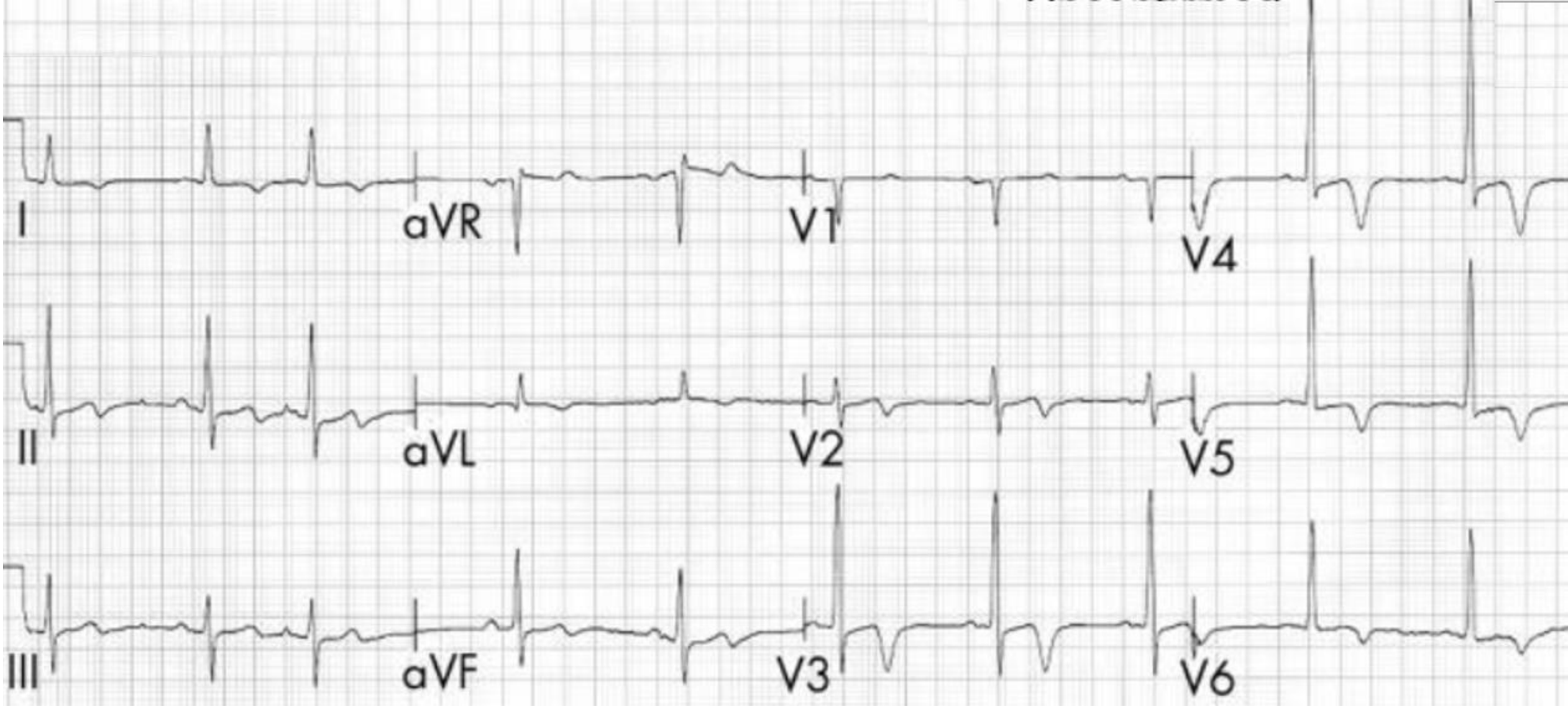


Biphasic “plus-minus” T-wave in V2

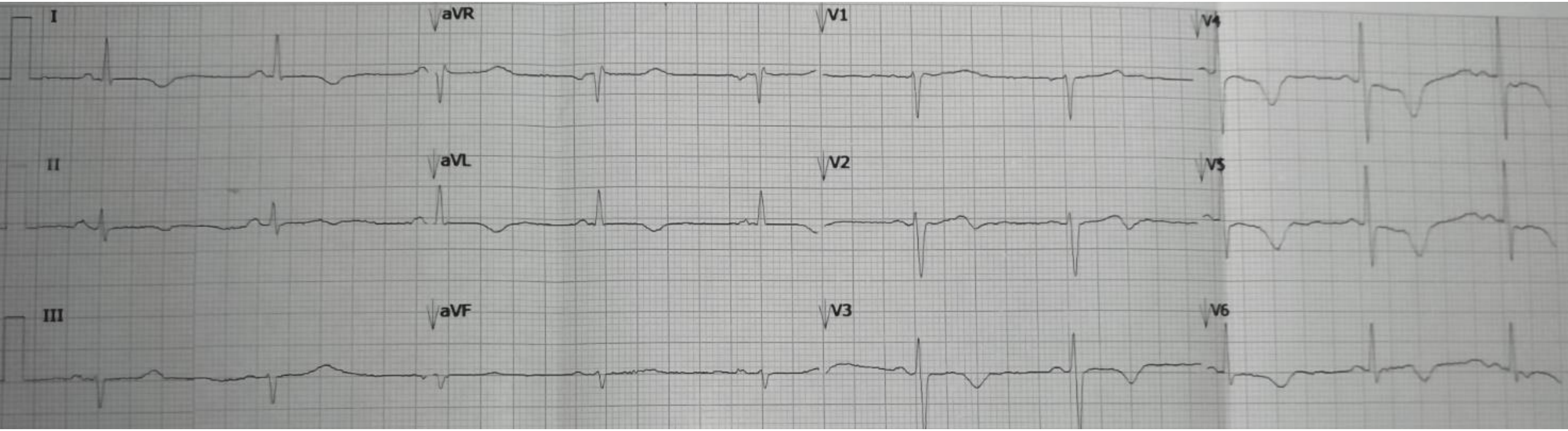
Dr. Santiago.Campbell´s case of our fórum

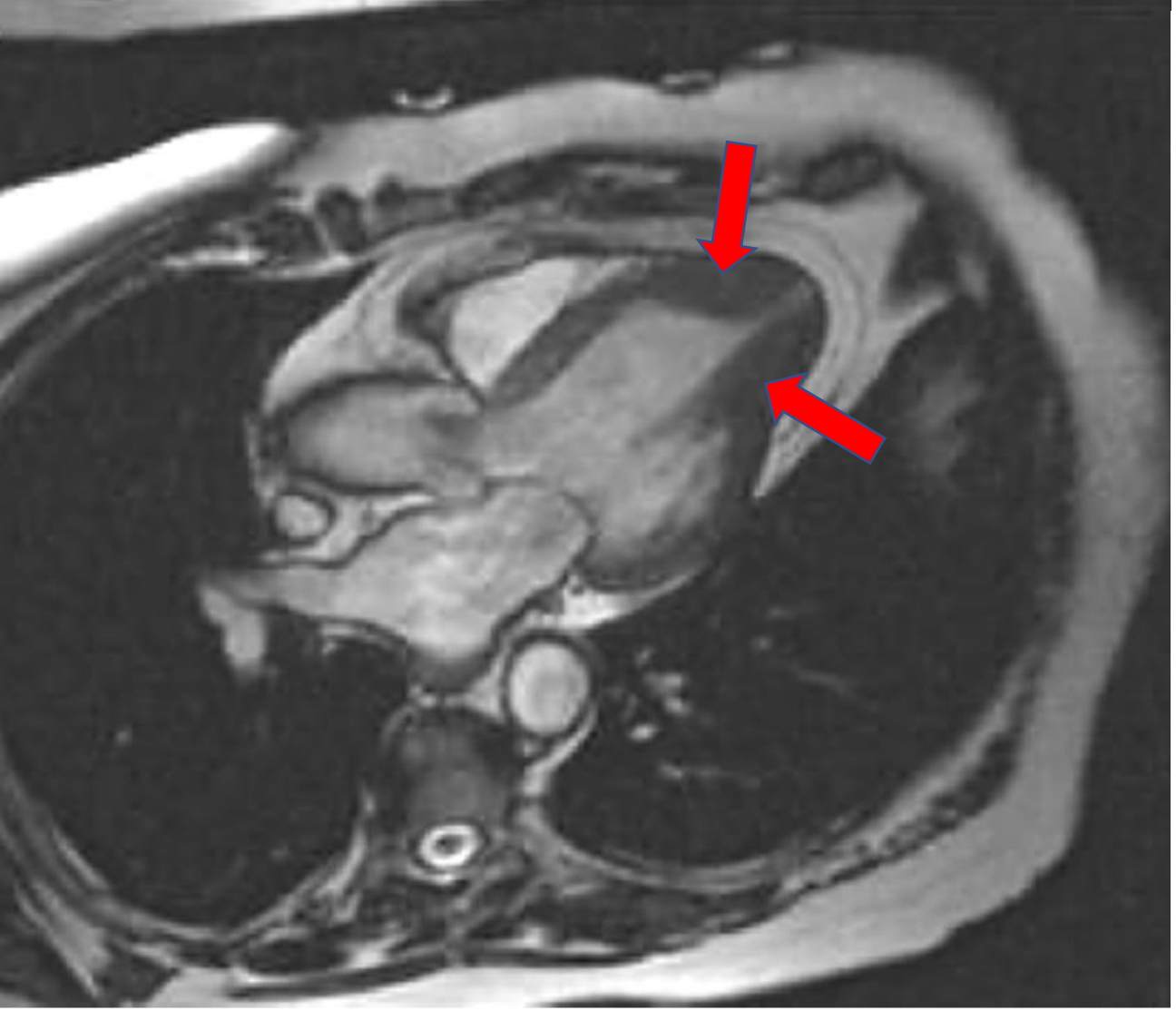


Biphasic “plus-minus” T-wave in V2

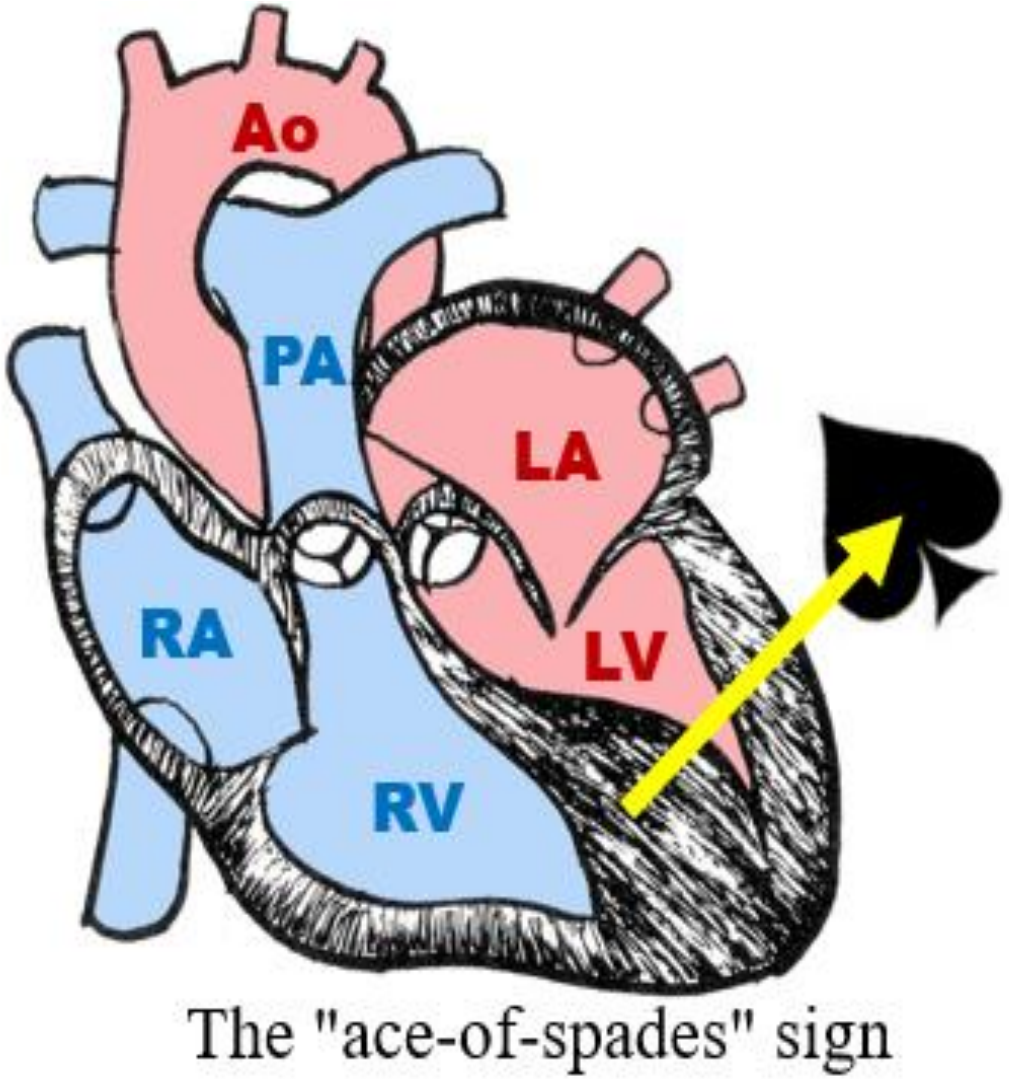

I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6

ECG from Dr. Santiago.Campbell´s case of our fórum





CMRI: Ap-HCM



The "ace-of-spades" sign

Myocardial ischemia and another element present in the disease secondary to:

1) microcirculation disease;
2) 2) decrease of vasodilator capacity;
3) 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.

Hypertrophyc cardiomyopathy classification

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

1) Obstructive form (OHCM)

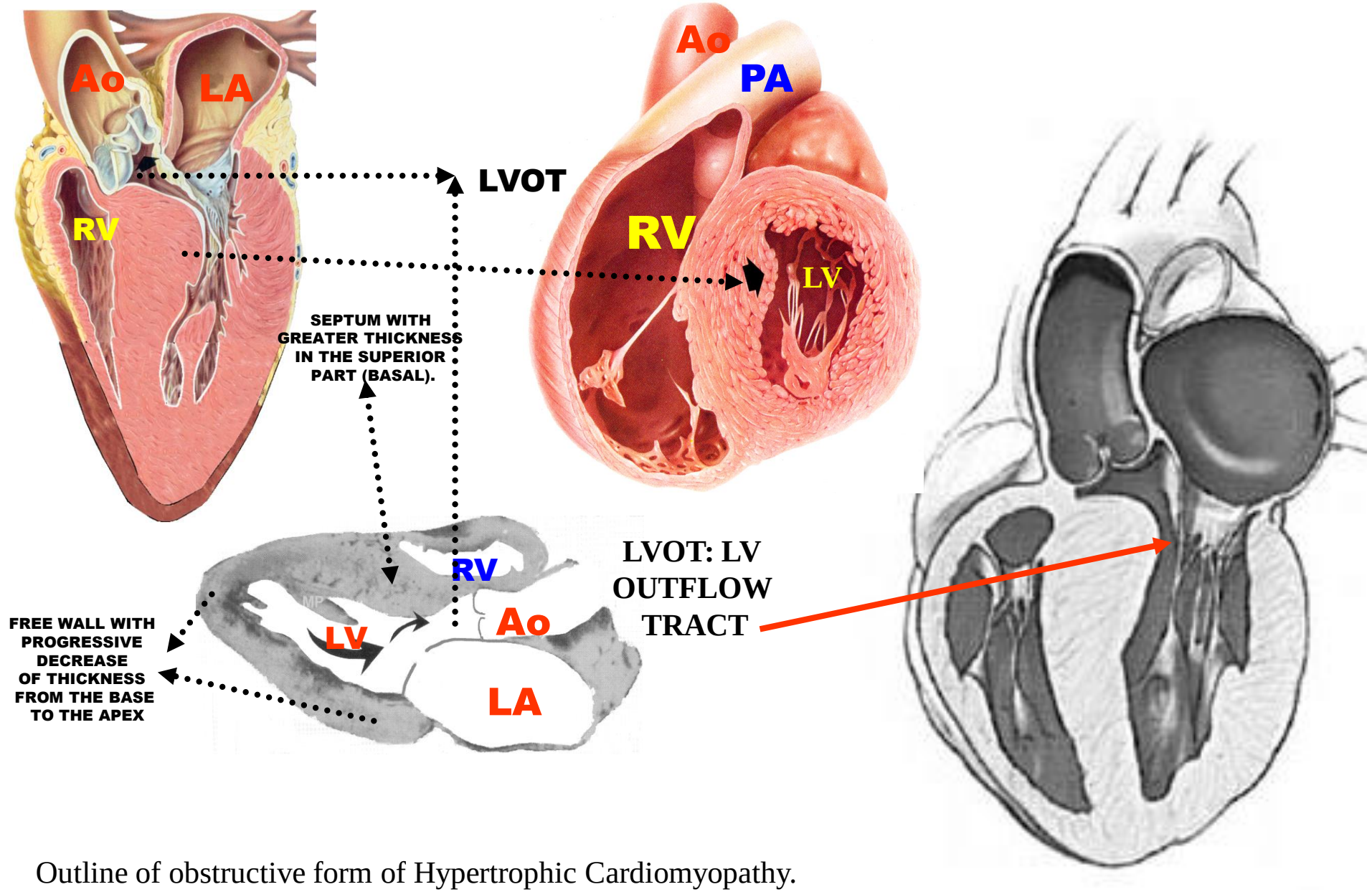
- Septal asymetrical with resting left ventricular outflow obstruction (obstruction at subaortic level): is found in approximately 20% of the patients, classically occurs at the, and is associated with mitral valve systolic anterior motion (SAM).
- Mid-ventricular obstructive HCM HCM(MVO-HCM) asymmetric LV hypertrophy with MVO and elevated intraventricular pressure gradients.(1%)

2) Non-obstructive Form (NO-HCM)

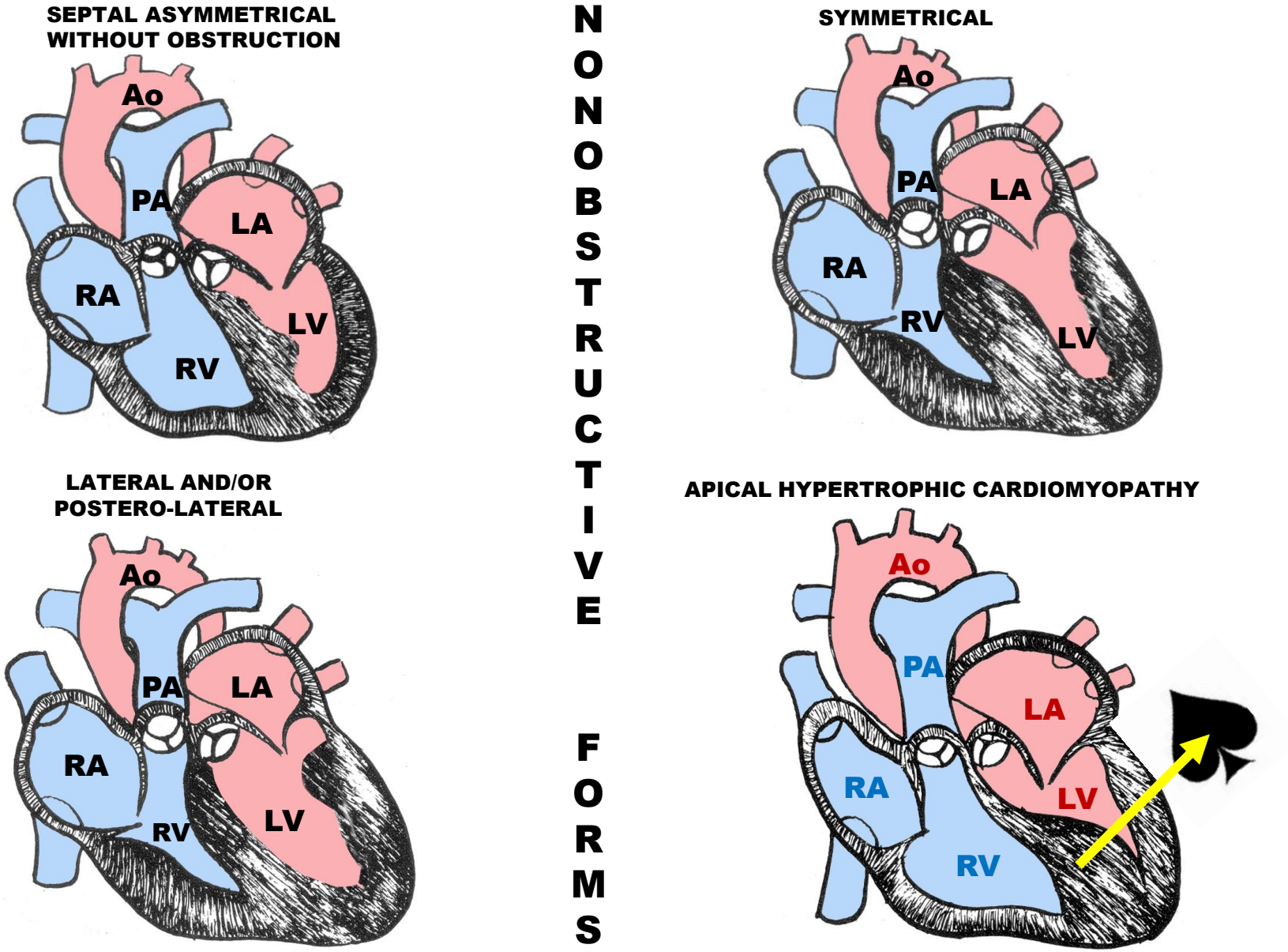
Septal asymetric with no obstruction;
Mid-septal hypertrophic cardiomyopathy
Apical Hypertrophic Cardiomiopathy (Ap HCM): 2%, 3% to 8%. ???? Could it be a slight form of Ap-HCM?
Lateral and/or posterolateral;
Concentric, symmetrical, or homogeneous hypertrophic: 5%.
Right ventricle: 2%.

OBSTRUCTIVE FORM (OHCM)	NONOBSTRUCTIVE FORM (NOHCM)
Septum with greater thickness in the superior part (basal)(20% or in the middle portion(1%).	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.

CLASSICAL OBSTRUCTIVE FORM OF HYPERTROPHIC CARDIOMYOPATHY (OHCM)



Outline of obstructive form of Hypertrophic Cardiomyopathy.

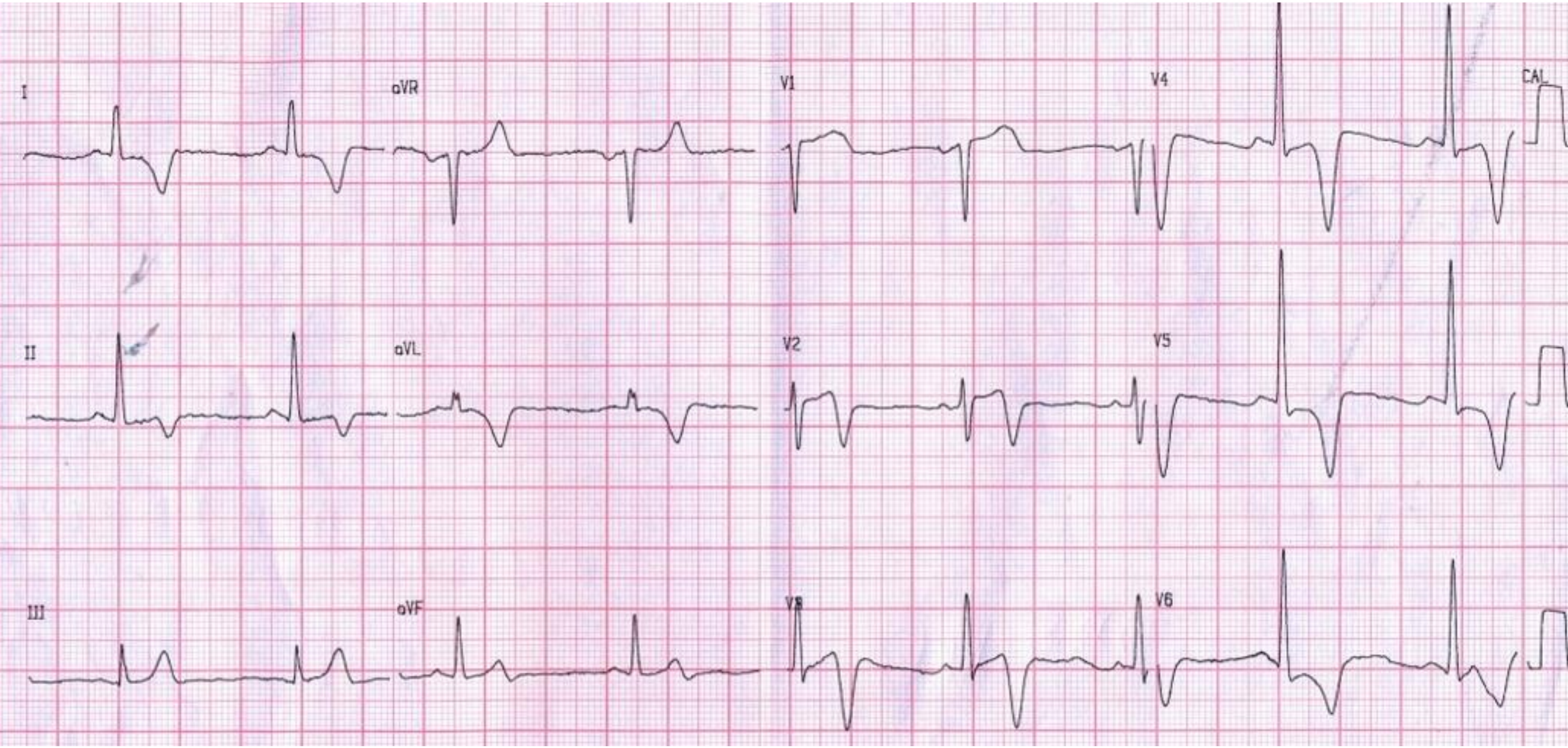


The "ace-of-spades" sign

Outline of nonobstructive form of Hypertrophic Cardiomyopathy.

Clinical case

Male, Caucasian, 41 years of age, with history of chest pain and exhaustion in strain.
Personal antecedentes: He mentions systemic hypertension without treatment currently. He doesn't smoke or have diabetes.
Family background: His father died with 77 years of age due to AMI? His brother died suddenly when he was 37 y.o. in his sleep.
Physical: Cardiac auscultation: regular heart rhythm; HR=68 bpm + fourth heart sound without murmur
Normal pulmonary artery and limbs
BP=140/100 mmHg
After the ECG was made (annex) we suggested performing Echo to rule out apical hypertrophic cardiomyopathy.
Echo: LV end diastolic diameter: 49 mm. LV end systolic diameter: 26 mm. Septal diastolic thickness: 14 mm. LV posterior wall diastolic diameter: 14 mm. Aorta: 29 mm; LA: 40 mm EF=78% Mass: 355 g
Conclusion: moderate LV concentric hypertrophy. Moderate LV diastolic dysfunction. Heart valves with normal morphological aspect. Absence of gradient in the LVOT. Mild mitral reflux.
Supplementary tests
Several ECGs with the same morphology.
Holter monitoring: Sinus rhythm predominates Asymptomatic isolated ventricular ectopic beats (210)
Absence of NSVT
Left heart catheterization: Presence of myocardial bridge in the proximal $^{1}/_{3}$ of the LAD
LV with asymmetrical hypertrophy with apical predominance (ventriculography shows spade-like morphology). Normal coronary arteries
Any comments?


I
aVR
V1
V4
CAL
II
aVL
V2
V5
III
aVF
V3
V6

Apical Hypertrophic cardiomyopathy Electrocardiogram features

Giant negative T waves in the precordial ECG leads: Giant negative T waves negativity ≥ 1.0 mV (10 mm). Giant negative T waves are more common in Japanese patients than American patients: 15% in Japan vs 3% in US (1). The singnificative 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 V2 to V5. T waves at the onset, may not present a significant voltage and may appear later with the evolution of the disease.(2)

The depth of negative T waves is related to craniocaudal asymmetry and apical late-enhancement (3)

Stress test may decrease the depth of T waves.(4)

Three hypotheses aroused to explain these negative T waves: 1) apical subendocardial ischemia.; 2) apical cell disorder;3) greater duration of action potential of hypertrophied cells, thus conditioning the area to have a slower repolarization.(5)

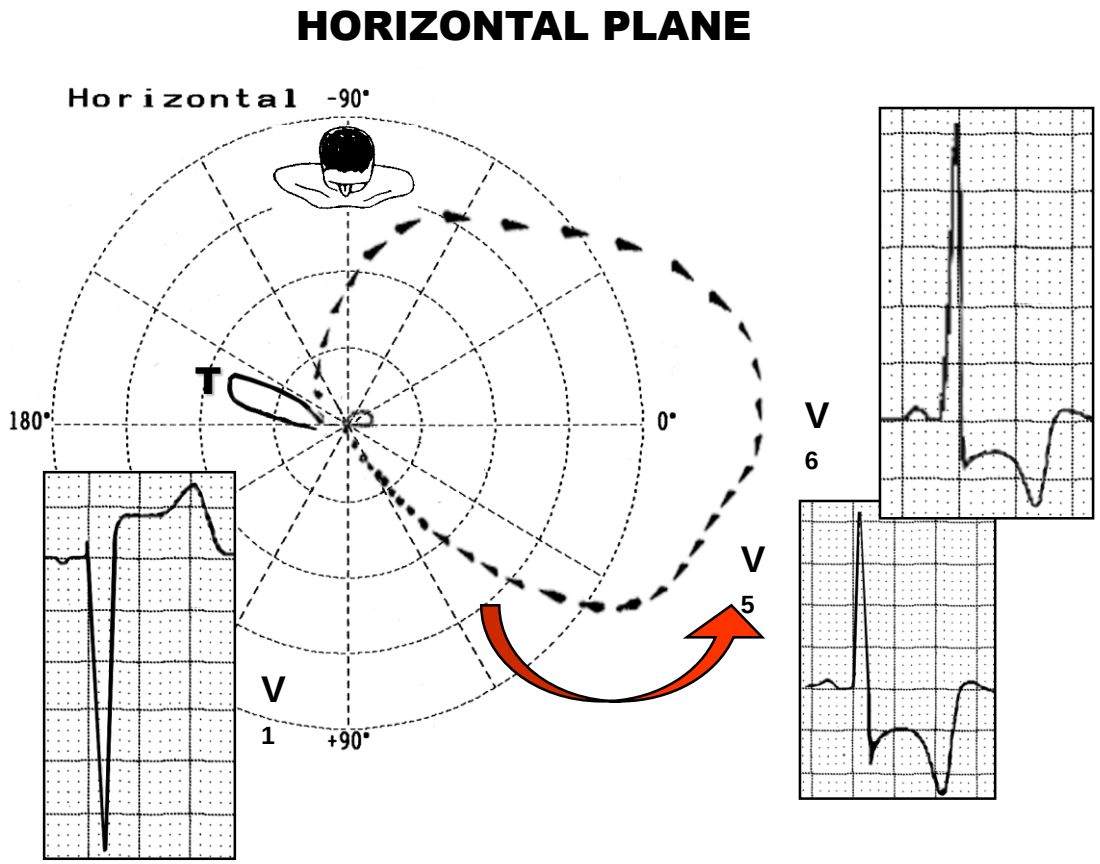
The prevalence in the western world of this form of HCM is approximately 0.02 to 0.2% and it constitutes 8% of the cases of the entity. In Japan, the apical form of HCM constitutes 25% of HCM.(6)

1. Kitaoka H, Doi Y, Casey SA, Comparison of prevalence of apical hypertrophic cardiomyopathy in Japan and the United States. Am J Cardiol. 2003;92:1183-1186.
2. Bielli M, Parravicini U, Zanetta M, Zenone F. . G Ital Cardiol. Apical hypertrophic cardiomyopathy: description of a case in advanced age with documentation of electrocardiographic course 1991;21:1325-1329.
3. Dumont CA, Monserrat L, Soler R, et al .Interpretation of electrocardiographic abnormalities in hypertrophic cardiomyopathy with cardiac magnetic resonance. Eur Heart J. 2006; 27:1725-1731.
4. Tilmant PY, Lablanche JM, Laurent JM, Héthuin JP, Folliot JP, Bertrand ME. . Non-obstructive hypertrophic myocardiopathy. Apropos of 5 cases Arch Mal Coeur Vaiss. 1980;73:1269-1278.
5. Tsunakawa H, Wei D, Mashima S, Harumi K. . Study on the genesis of giant negative T wave in apical hypertrophic cardiomyopathy using a three-dimensional computer model. Jpn Heart J. 1991;32:799-809.
6. Maron BJ. The giant negative T wave revisited ... in hypertrophic cardiomyopathy. J Am Coll Cardiol. 1990 Apr;15:972-973.

Sometimes R-wave voltage and T-wave negativity progressively decreased in magnitude at serial ECGs
Non-sustained or sustained VT can be observed in patients that developed apical aneurysm with normal coronary arteries;To clarify the mechanisms of ECG abnormalities in hypertrophic cardiomyopathy, 102 patients were examined with CMR. Distribution and magnitude of hypertrophy and late-enhancement were correlated with ECG abnormalities:
Abnormal Q waves reflect the interrelation between upper anterior septal thickness and other regions of the left and right ventricles, and wider Q waves are associated with late-enhancement;
Conduction disturbances and absent septal Q waves are associated with late-enhancement;
The depth of negative T waves is related to craniocaudal asymmetry and apical late-enhancement (1)

1. Dumont CA, Monserrat L, Soler R, et al .Interpretation of electrocardiographic abnormalities in hypertrophic cardiomyopathy with cardiac magnetic resonance. Eur Heart J. 2006; 27:1725-1731.

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;

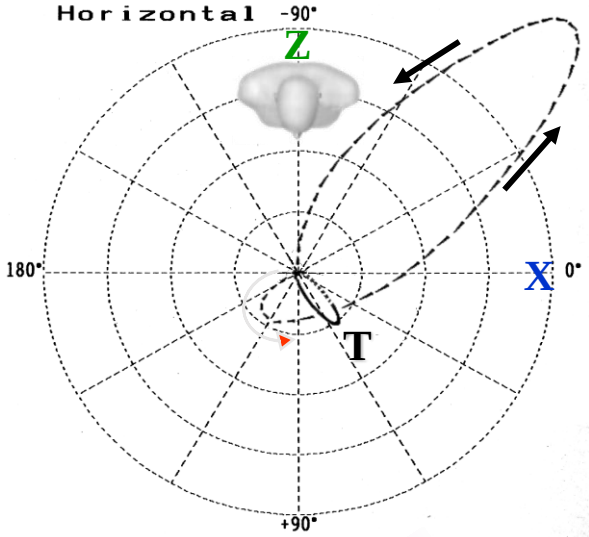
4) Final vectors located to the right and backward with ST/T vector in the right posterior quadrant;

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

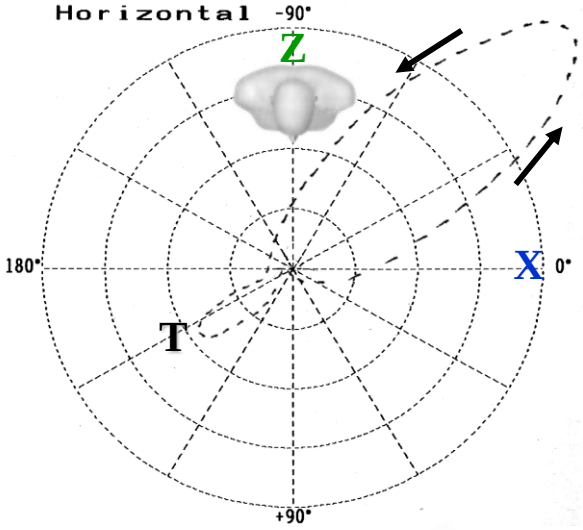
Vectocardiogram features in Apical Hypertrophic Cardiomyopathy.

The five vectorcardiographic types of LVH in the HP: IA, IB, II, III and IV

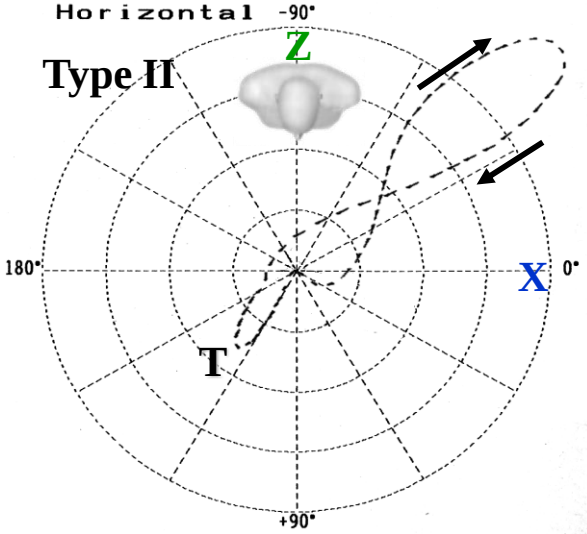
Type IA diastolic LVH



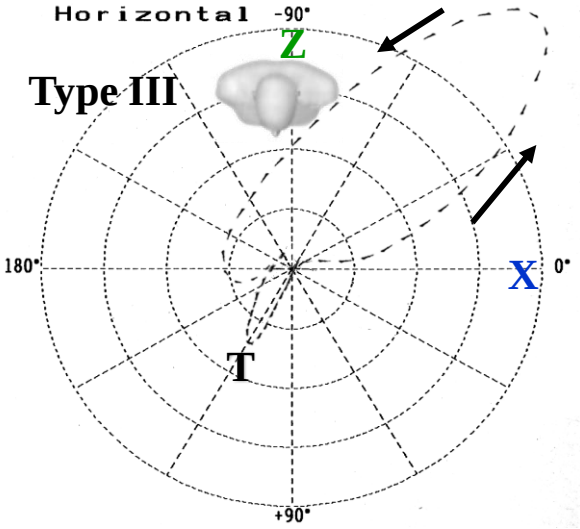
Type IB Systolic LVH



Type II



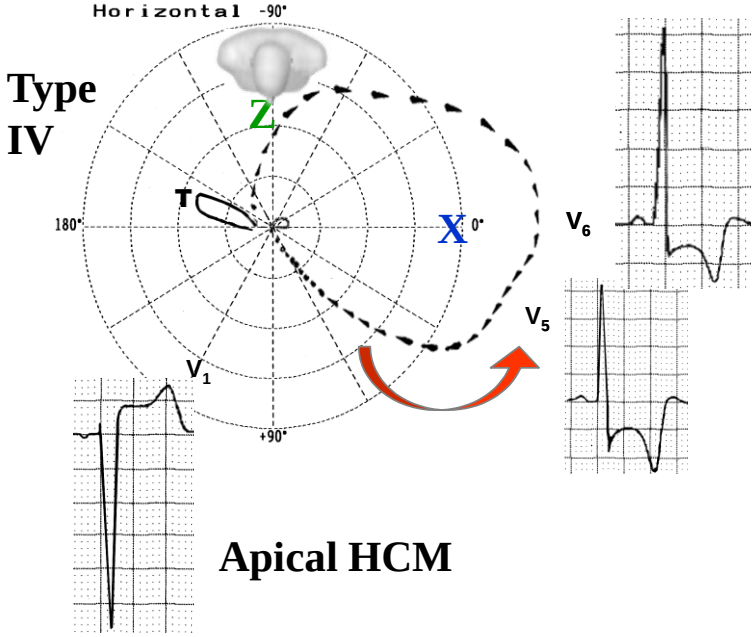
Type III



Eccentric LVH

Concentric LVH

Type IV



Apical HCM

VCG criteria for left ventricular hypertrophy

VCG has a superior sensitivity and specificity than ECG to detect LVH (Vine 1971).

From 100 autopsied cases with LVH studied by Abbott-Smith and Chou (Abbott-Smith 1970), VCG was capable of diagnosing 50% with just 11.7% of false positives.

It enables to clarify doubtful cases of association with septal or anteroseptal electrically inactive areas, which certain LVHs of the systolic type may cause in ECG (LVH with QS in V_1, V_1 and V_2 or V_1, V_2 and V_3). Thus, in absence of anterior electrically inactive area, the VCG shows the dashes from the initial 10 to 20 ms of the QRS loop without delay.

When there is a possible septal inactive area, the vector of the initial 20 ms is located in the left posterior quadrant.

Frequently, the vector of the initial 10 ms in LVH of high blood pressure is heading backward or to the front and the left, originating complexes of the QS type in V1 or V1-V2 simulating septal inactive area (Hugenholtz 1963).

The VCG seems to be superior to the ECG and the echocardiogram for the diagnosis of ventricular hypertrophies associated to electrically inactive areas, besides having a greater correlation with the echocardiogram than ECG when estimating the LV mass.

In our service, we follow the criteria by Varriale et al (Varriale 1966), modified, which take into account the characteristics of the QRS loop in the horizontal plane (HP); thus, five types are described: IA, IB, II, III and IV.

Type IA: vector of the initial 20 ms heading to the front and the right (Type IA) or to the front and the left (Type IB), oval morphology, counterclockwise rotation, and most of the QRS loop located in the left posterior quadrant.
T loop matching QRS (IA) or not matching QRS (IB).
Type IB: very similar to the QRS loop of CLBBB: vector of the initial 20 ms heading to the front and the left, (rare to the right), rotation in eight with counterclockwise proximal portion and clockwise distal portion, and E point that does not coincide with the 0 point, and located to the front and the right from the latter.
T loop to the front and right, opposite to the QRS loop.
Note: it is differentiated from CLBBB by the absence of middle-final delay.
Type II: this is the variant frequently found in LVE with high blood pressure, characterized by initial vectors heading to the right and discretely to the front, clockwise rotation (inverted), simulating antero-lateral infarction, narrow morphology and QRS loop located mostly in the left posterior quadrant.
Type III: Initial vectors of QRS loop heading to the front and the left; QRS loop more anterior and predominantly located in the left anterior quadrant; increased voltage of maximal vector;
Final vectors located to the right and backward with the ST/T vector in the right posterior quadrant;
E point not matching 0 point and located backward and to the right of the latter.
Type IV: Characterized by:
- Initial vectors of QRS loop heading to the front and the left;
- QRS loop more anterior and predominantly located in the left anterior quadrant;
- Increased voltage of maximal vector;
- Final vectors located to the right and backward with the ST/T vector in the right posterior quadrant;
- E point not matching 0 point and located backward and to the right of the latter.