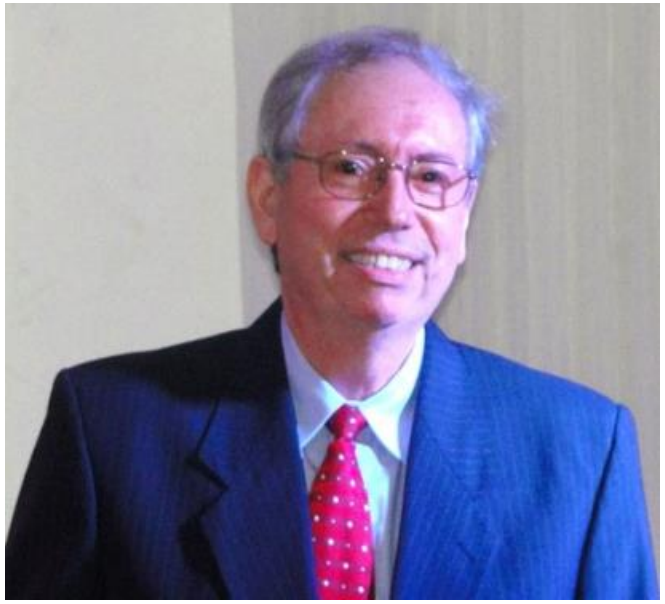
An entity where ECG/VCG are “gold standard” for the diagnosis

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



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

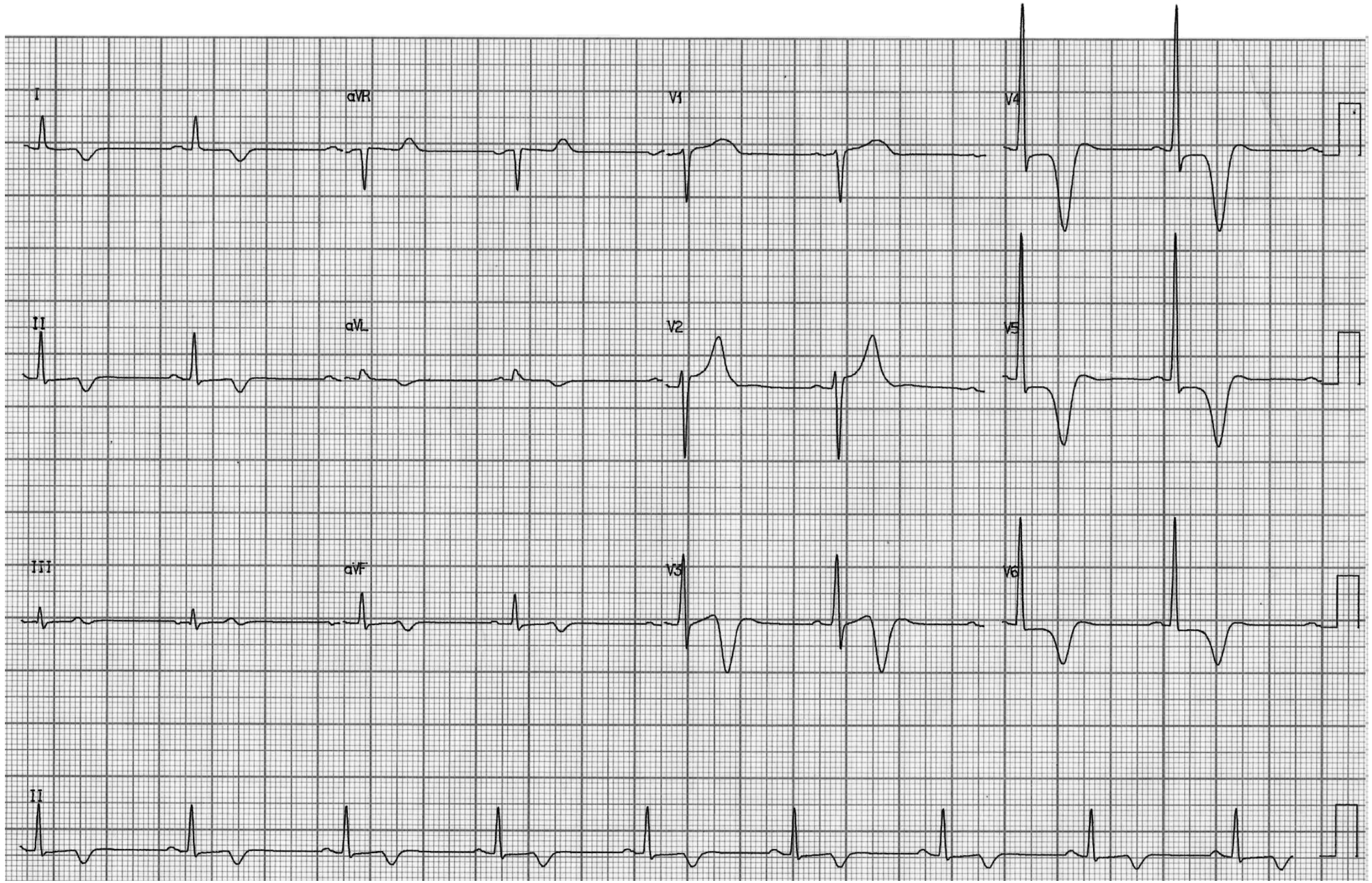
Case report

Caucasian male, 34 years of age, with history of chest pain and exhaustion in strain.
Personal antecedents: 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/VCG were performed (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 premature ventricular contractions (PVCs) (210)
Absence of NSVT. We performed cardiac catheterization.

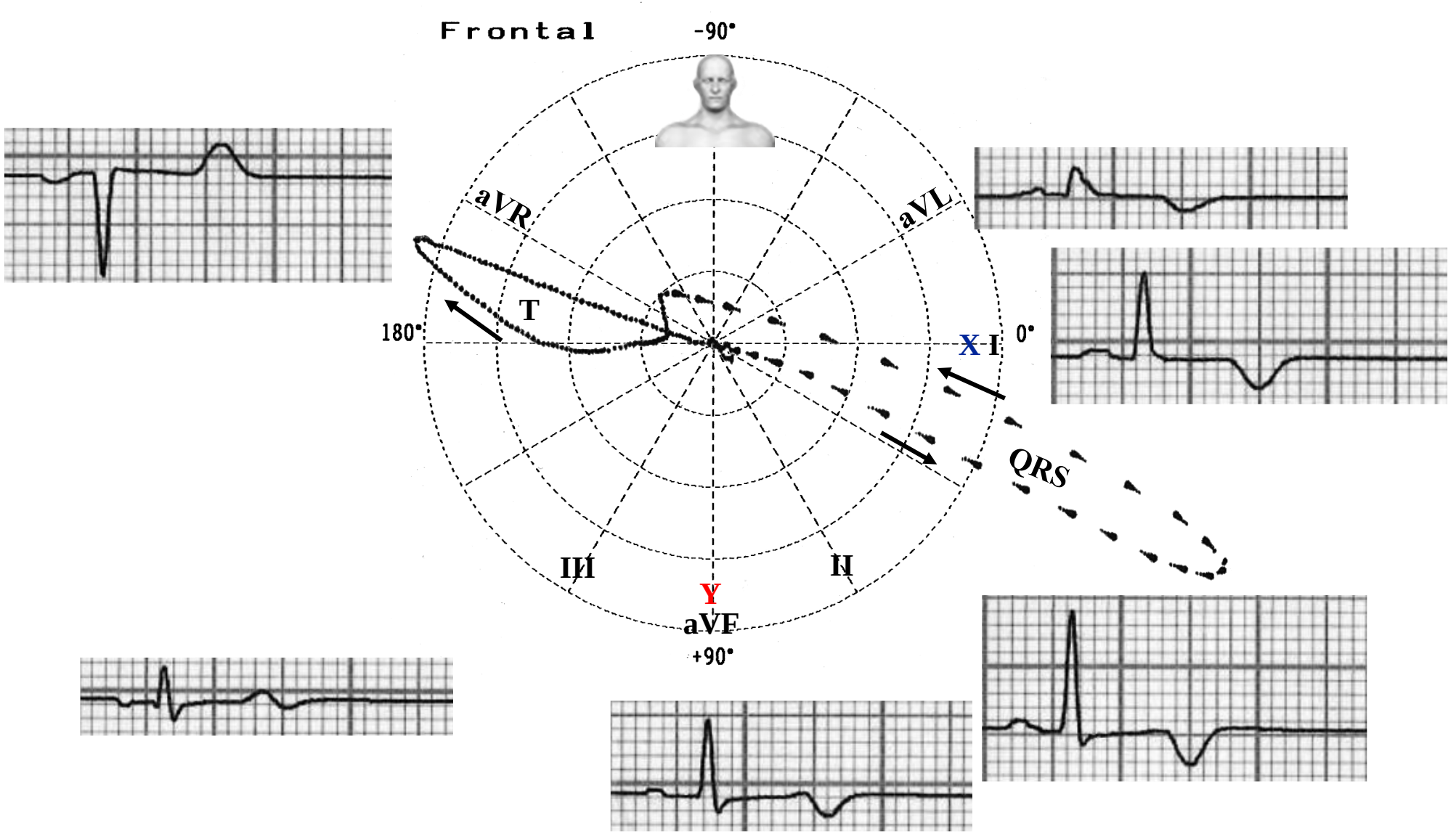
Questions:

1. Which is the most probable clinical diagnosis?
2. Which is the ECG/VCG diagnosis?
3. Which is the differential diagnosis?
4. Which is the prognosis?

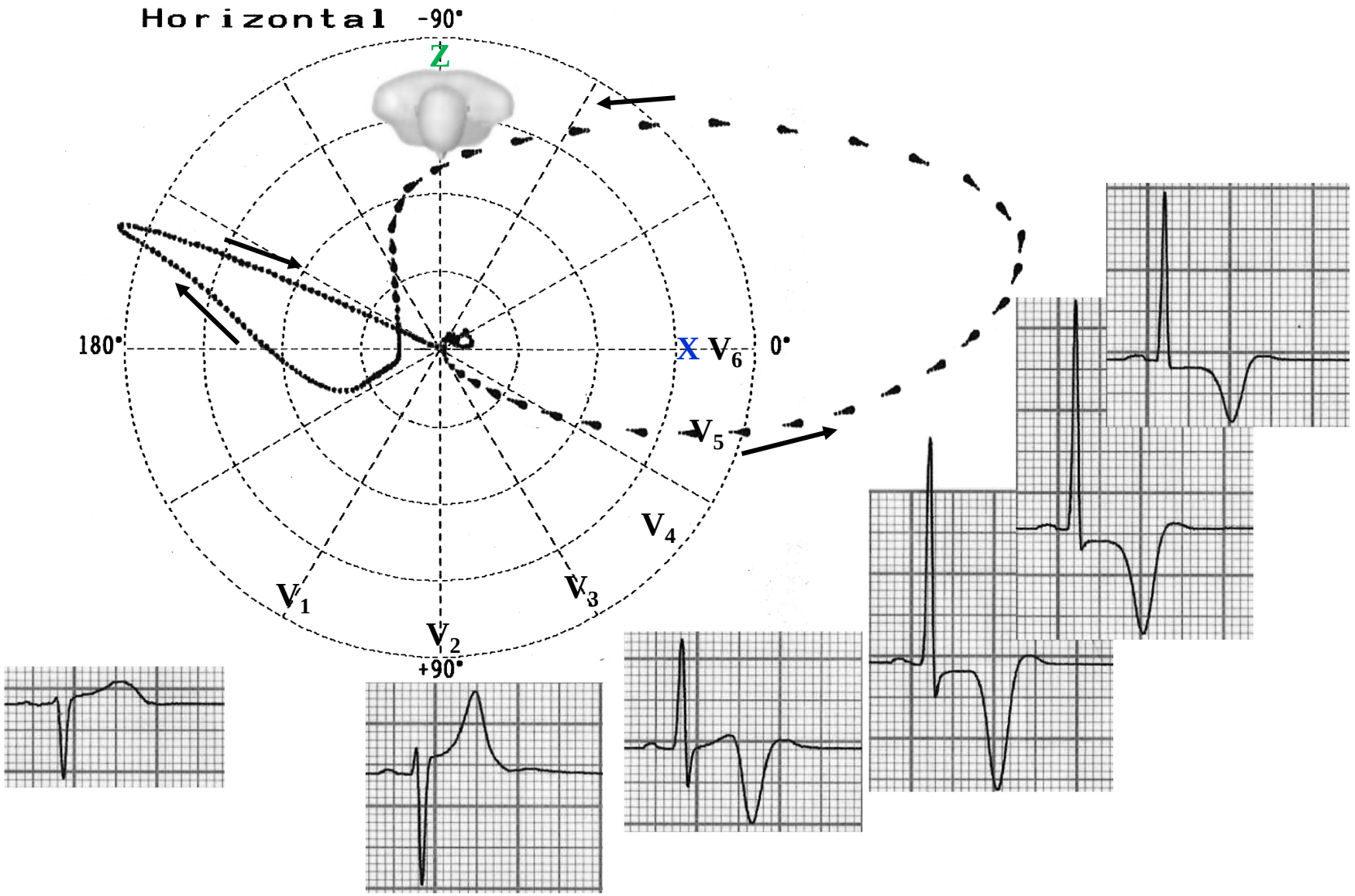
ECG-1



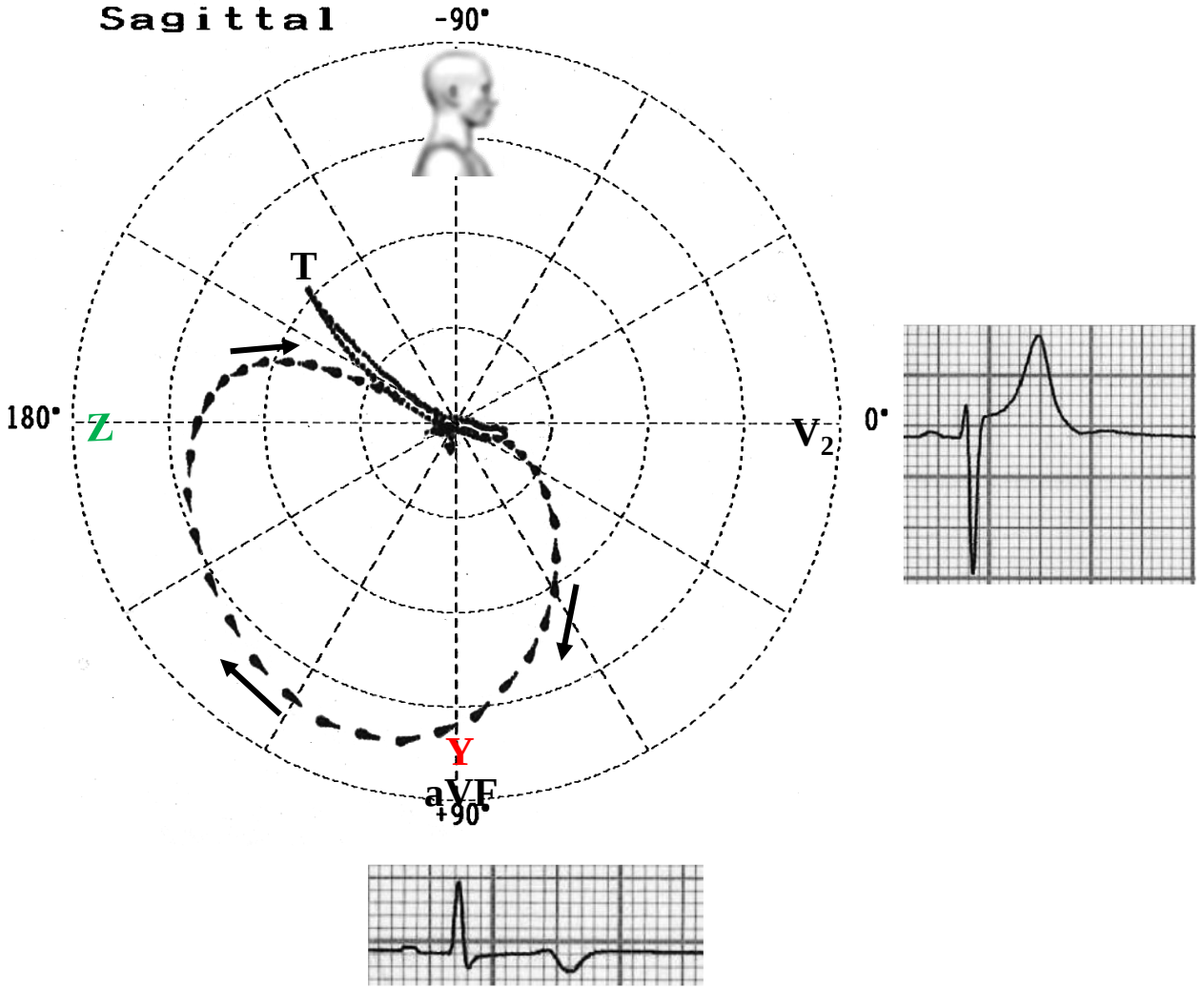
ECG/VCG correlation in the frontal plane

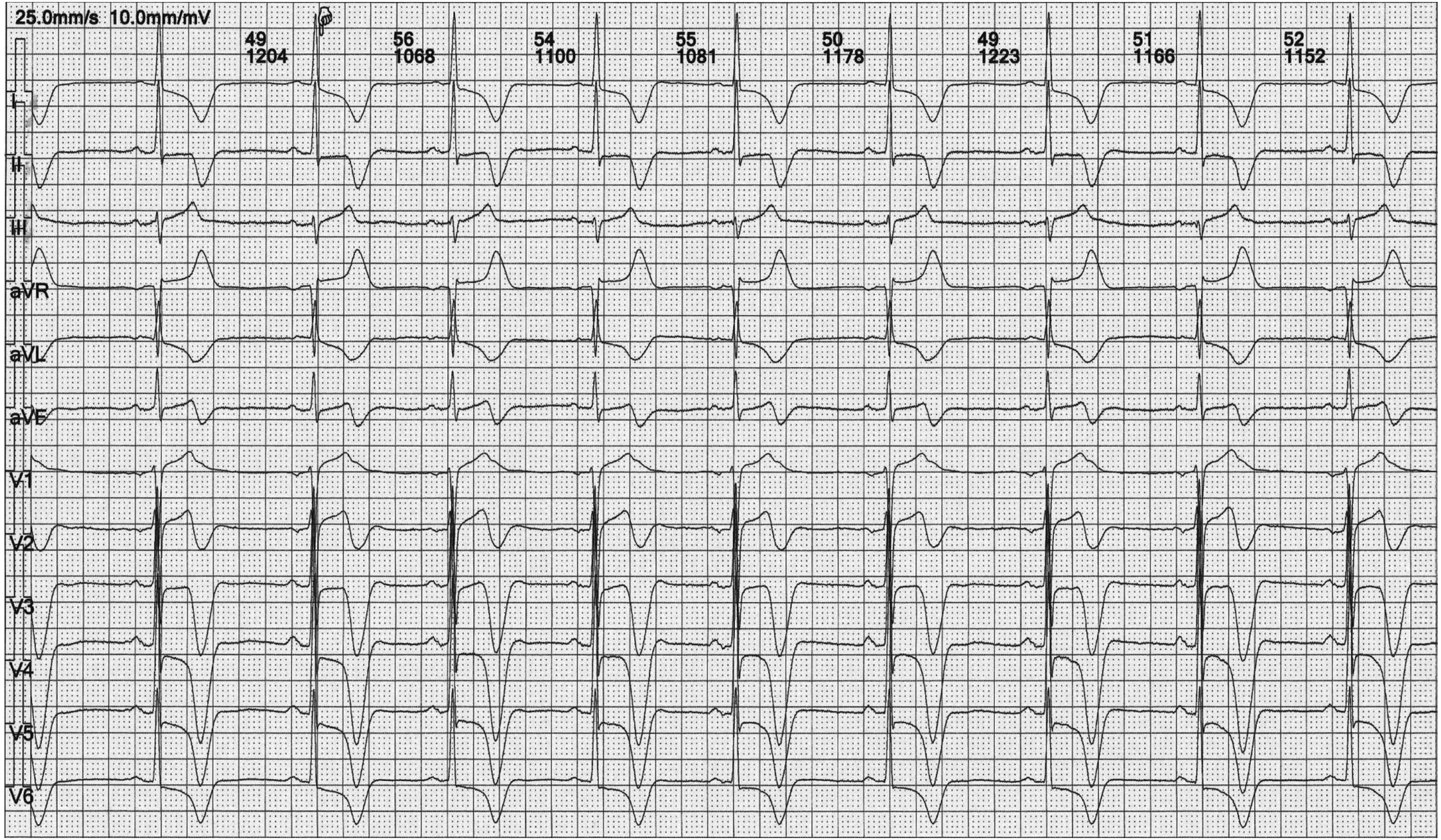


ECG/VCG correlation in the horizontal plane



ECG/VCG correlation in the right sagittal plane




25.0mm/s 10.0mm/mV
49
1204
56
1068
54
1100
55
1081
50
1178
49
1223
51
1166
52
1152
I
II
III
aVR
aVL
aVF
V1
V2
V3
V4
V5
V6

Colleagues opinions

Thanks Andrés
My answer:
-hypertrophic concentric Cnr
-left ventricular hypertrophy
-genetic testing suggested also for the prognosis
Bye

Bortolo Martini
Direttore Unità Operativa di Cardiologia ULSS4
Unidad Cardiovascular, Hospital Alto Vicentino, Santorso, Italia.
Electronic address: bortolo.martini@gmail.com

Estimado Potro.
El ECG presenta Bradicardia sinusal (presumo se encuentra recibiendo BB o similar). Eje eléctrico normal. Infradesnivel del segmento ST máximo en V4 y V5. Ondas T negativas picudas, gigantes simétricas, máximas en V4 y V5. Además T negativas en I y aVL y II.
Mi diagnóstico presuntivo ECG es de una miocardiopatia hipertrófica apical.
Los diagnósticos diferenciales se presentan con otros tipos de miocardiopatías infiltrativas. En este particular la resonancia nuclear magética contribuirá para el diagnóstico. Obviamente, dada la presencia de angina y cambios del ST/T, obliga a descartar coronariopatia asociada, incluyendo en esta a la presencia de puentes musculares ya que la asociación de estos con la miocardiopatia hipertrofica no son infrecuentes. A pesar de no considerar que la presencia de estos sea la causa de los cambios persistentes del ECG.
Se hace muy extenso d enumerar otras causas aún considerándolas poco probables en el presente caso como la Insuficiencia Aortica severa, hipertensión arterial con hipertrofia ventricular izquierda inexplicada y sobrecarga del ventrículo izquierdo (por ejemplo por enfermedad de Fabry).
Ud nos mostrará otros patrones similares de injuria sub-endocárdica por diferentes entidades.
Un cordial saludo
Martín Ibarrola MD Buenos Aires Argentina

Dear Potro: The ECG has sinus bradycardia (presumed to be receiving β-blocker or similar), normal QRS axis, ST segment depression in V4 and V5. Negative, symmetrical, and giants T waves maximums in V4 -V5. Also negative T-waves in I and aVL and II. My presumptive ECG diagnosis is **Apical Hypertrophic Cardiomyopathy.(Ap-HCM).** Differential diagnoses are presented with other types of infiltrative cardiomyopathies. In this particular cardiac nuclear magnetic resonance(NMR) will contribute to the diagnosis. Obviously, given the presence of angina and the ST / T changes, it is necessary to rule out coronary artery disease(CAD) associated including in the presence of muscular bridges, since this association with HCM is not infrequent. Despite not considering that the presence of these is the cause of the persistent changes of the ECG. It is very extensive to enumerate other causes still considering them unlikely in the present case as the Severe aortic insufficiency, hypertension, unexplained LVH and left ventricular overload (eg Fabry disease).
You will show us other similar patterns of subendocardial injury by different entities.
A warm greeting
Martín Ibarrola MD Buenos Aires Argentina

Hello. Apical hypertrophic cardiomyopathy. Echo is not reliable to rule out. Unexpected sudden death in first degree relative should be considered regarding prognosis.

Best regards

Kjell Nikus MD PhD

Heart Center, Tampere University Hospital. Biokatu 6, 33520 Tampere, Finland.

Looks like classic LV apical hypertrophic non obstructive cardiomyopathy. Needs MR study to exclude apical aneurysm which carries poor prognoses. In addition, Look at LV scar areas a percentage of LV mass to determine prognosis. History of familial sudden death is worrisome. Prognoses for this condition is usually good but we have in hospital now a young male who was survivor of Sudden cardiac death.

Melvin Scheinman, MD
Department of Cardiac Electrophysiology, University of California San Francisco, San Francisco, California, USA.
Professor of Medicine
UCSF
Electrophysiology Service
500 Parnassus Avenue
San Francisco, CA 94143-1354
Telephone/FAX/E-mail:
Phone: (415) 476-5706
Fax: (415) 476-6260
email: scheinman@medicine.ucsf.edu

Spanish

Queridos amigos Potro y Raimundo:

Respondo vuestras preguntas:

Diagnóstico clínico-ECG: Miocardiopatía hipertrófica apical. El hecho que el informe del ecocardiograma no haga mención de esta patología, no es motivo para no diagnosticarla, en virtud que:

a) El ecocardiograma de la punta siempre es motivo de continuas controversias, por razones básicamente técnicas. Diría inclusive, que hay veces en que es preferible no "sobrediagnosticar" elementos de la punta, en caso de dudas. Hoy en día, para ver adecuadamente esta patología, yo pediría una Resonancia Magnética Nuclear y eventualmente, una ventriculografía.

b) El ECG del caso es altísimamente sugestivo de MCH apical, con ondas T muy simétricas, picudas, profundas y negativas de V3 a V6, con un ST recto, y con mínima depresión del punto J. Además, en el plano frontal, se repite esa simetría de la onda T, con ST recto.

No voy a opinar sobre el VCG, porque conozco realmente muy poco de esta técnica.

Por qué el dolor de pecho? Posiblemente la hipertrofia miocárdica genera isquemia relativa. Aunque el relato no dice concretamente "angor", es posible esa fisiopatología.

La presencia de un R4 es un hallazgo frecuente en la MCH en sus distintas localizaciones, y obedece a que la aurícula izquierda tiene vigorosa contracción para "vencer" a una relajación (distensibilidad) ventricular disminuida por la mayor rigidez del VI hipertrófico y miocardiopático.

No puedo traducir adecuadamente la frase"exhaustion in strain". Si fuese "debilidad durante el esfuerzo físico", se debería a que es un miocardio afectado globalmente, pero sobre todo en la punta, región que es importante componente responsable de la función sistólica del V.I.

Diagnóstico diferenciales:

Del ECG : obstrucción proximal de la arteria descendente anterior.

Del ecocardiograma: En caso de que la punta se pueda visualizar con cierta facilidad, hay que descartar algún raro tumor; alguna variante de miocardio no compactado; trombo apical; fibrosis endomiocárdica, aneurisma ventricular. Creo que son los diagnós frecuentes.

Pronóstico: En este caso en particular, la presencia de un hermano fallecido súbitamente, hace sombrío el pronóstico.

Les mando un abrazo y gracias por compartir este caso.

Dr Mario Heñin

Resistencia (Chaco)

Argentina

English

Dear friends, Potro and Raimundo:

Regarding your questions:

Clinical-ECG diagnosis: Apical hypertrophic cardiomyopathy. The fact that the echocardiogram report does not mention this pathology is no reason not to diagnose it, because:

A) Echocardiogram of the apex is always subject to continuous controversy, basically for technical reasons. I would even say that there are times when it is preferable not to "overdiagnose" apex elements in case of doubt. Nowadays, in order to properly check this pathology, I would request a Nuclear Magnetic Resonance and eventually, a ventriculography.

B) The ECG is highly suggestive of apical HCM, with very symmetrical, sharp, deep and negative T-waves from V3 to V6, with a straight ST, and with minimal depression of J-point. Moreover, the symmetry of the T wave, with straight ST repeats in the frontal plane.

I will not comment about the VCG, because I really know very little about this technique.

Why the chest pain? Possibly myocardial hypertrophy generates relative ischemia. Although the story does not specifically say "angor", this pathophysiology is possible.

The presence of an R4 is a frequent finding in the HCM in its different locations, due to the fact that the left atrium has a vigorous contraction to "overcome" a decreased ventricular relaxation (distensibility) due to the greater rigidity of the hypertrophic and myocardiopathic LV.

I can not adequately translate the phrase "exhaustion in strain". If it were "weakness during physical effort", it should be because of a globally affected myocardium, but especially in the tip region which is an important component responsible for the systolic function of the LV.

Differential diagnosis:

ECG: proximal obstruction of the left anterior descending artery.

Echocardiogram: In case the apex can be visualized with some facility, it is necessary to rule out some rare tumor; some noncompacted myocardial variant; apical thrombus; endomyocardial fibrosis, and ventricular aneurysm. I think they are the most frequent differential diagnoses.

Prognosis: In this particular case, the presence of a suddenly deceased brother makes the forecast gloomy.

I send you a hug and thank you for sharing this case.

Mario Heñin, MD
Resistencia (Chaco)
Argentina

Portuguese

Caros amigos,

Diagnóstico clinico: Miocardiocardia hipertrófica assimétrica apical não obstrutiva (Rara e muito dependente da experiência do ecocardiografista).

Destaque-se o aspecto vetorcardiográfico nos vários eixos em concordância com o ECG.

Diagnóstico do ECG/VCG:

1. Sobrecarga Ventricular Esquerda (Diretrizes Brasileiras não admite a terminologia hipertrofia)
2. Alterações da repolarização ventricular tipo isquêmica (No caso de CMHNO sem haver doença coronária as causas da isquemia podem ser:
    a) Doença da microcirculação;
    b) Diminuição da capacidade vasodilatadora;
    c) Compressão sistêmica dos vasos septais e os subepicárdicos;
    d) Queda da pressão na raiz da aorta;
    e) Dificuldade no enchimento coronariano pela hipertrofia;
    f) Aumento desproporcional da massa e consequente desbalanceamento da oferta/demanda

O diagnostico diferencial já foi comentado por Martin.

Abraços

Adail Paixão-Almeida, MD

Vitória da Conquista - Bahia/Brasil

English

Dear friends,

Clinical diagnosis: Non-obstructive apical asymmetric hypertrophic cardiomyopathy (rare and very dependent on the experience of the echocardiographer). The VCG aspect in the various axes in accordance with the ECG is highlighted.

ECG/VCG diagnosis:

Left Ventricular hypertrphy (Brazilian Guidelines does not admit hypertrophy terminology. They use left ventricular overloading).

Ischemic-like changes in ventricular repolarization (in the case of NO-HCM without coronary disease, the causes of ischemia may be consequence of:

1. Microcirculation disease;
2. Decreased vasodilatory capacity;
3. Compression of septal and subepicardial vessels;
4. Pressure drop in the root of the aorta;
5. Difficulty in coronary filling by hypertrophy;
6. Disproportionate increase in mass and consequent imbalance of supply / demand

The differential diagnosis has already been commented by Martin.

Hugs

Adail Paixão-Almeida, MD

Vitória da Conquista - Bahia / Brazil

Spanish

Estimados maestros:

El diagnóstico más probable es miocariopatia hipertrofica apical (más frecuente en individuos japoneses).

1) En el ECG los hallazgos más importantes son:

a-Patrón de hipertrofia VI (índice de Sokolov-Lyon mayor de 35 mm.
b- Deflexión intrisecoide igual/mayor de 50 ms
c- onda P mayor de 110 mseg
d- bradicardia sinusal
e- ondas T gigantes negativas (más de 10 mm) de V3 a V6, D1, aVL y DII, patrón de sobrecarga sistólica. SupraST en V1 V2 e InfraST en cara anterolateral.

Una Ergometria podría disminuir la profundidad de ondas T, que ante ausencia de enfermedad Coronaria debería hacer sospechar MCH.
g- ángulo QRS-ST-T mayor de 100°

2) Creo que el VCG con onda T dirigida hacia atrás y a la derecha es importante para el diagnóstico.

La fisiopatologia de las ondas T negativas ya fue descripta por Dr. Adail.

Agregaría a los diag diferenciales enumerados por Dr. Martín, la memoria miocardica y corazón de atleta.

El diagnostico definitivo es por RMN.

Juan Mazzardo MD Mendoza Argentina

Dear teachers:
The most likely diagnosis is apical hypertrophic myocardiopathy (more frequent in Japanese individuals).

1) In the ECG the most important findings are:
   A. LVH voltage criteria (Sokolov-Lyon index ≥35 mm)
   B. R-wave peak Time or Intrisecoid deflection ≥50 ms
   C. P wave duration >110 msec
   D. Sinus bradycardia
   E. Deep T-waves inversion (≥10mm) in precordial leads and sometimes in inferior one from V3 to V6, I, aVL and II
   A. LV systolic overload pattern or strain pattern
   B. ST segment elevation inV1 V2 and ST depression in anterolateral wall.

An Ergometry could reduce the depth of T waves, which in the absence of CAD should make MCH diagnosis suspicious.
QRS-ST-T angle >100 °
I believe that the VCG with T wave directed towards the back and to the right is important for the diagnosis.
The pathophysiology of negative T waves has already been described by Dr. Adail.
I would add to the differential diagnoses listed by Dr. Martín, myocardial memory and athlete's heart.

The definitive diagnosis is by NMR.
Juan **Mazzardo** MD Mendoza Argentina

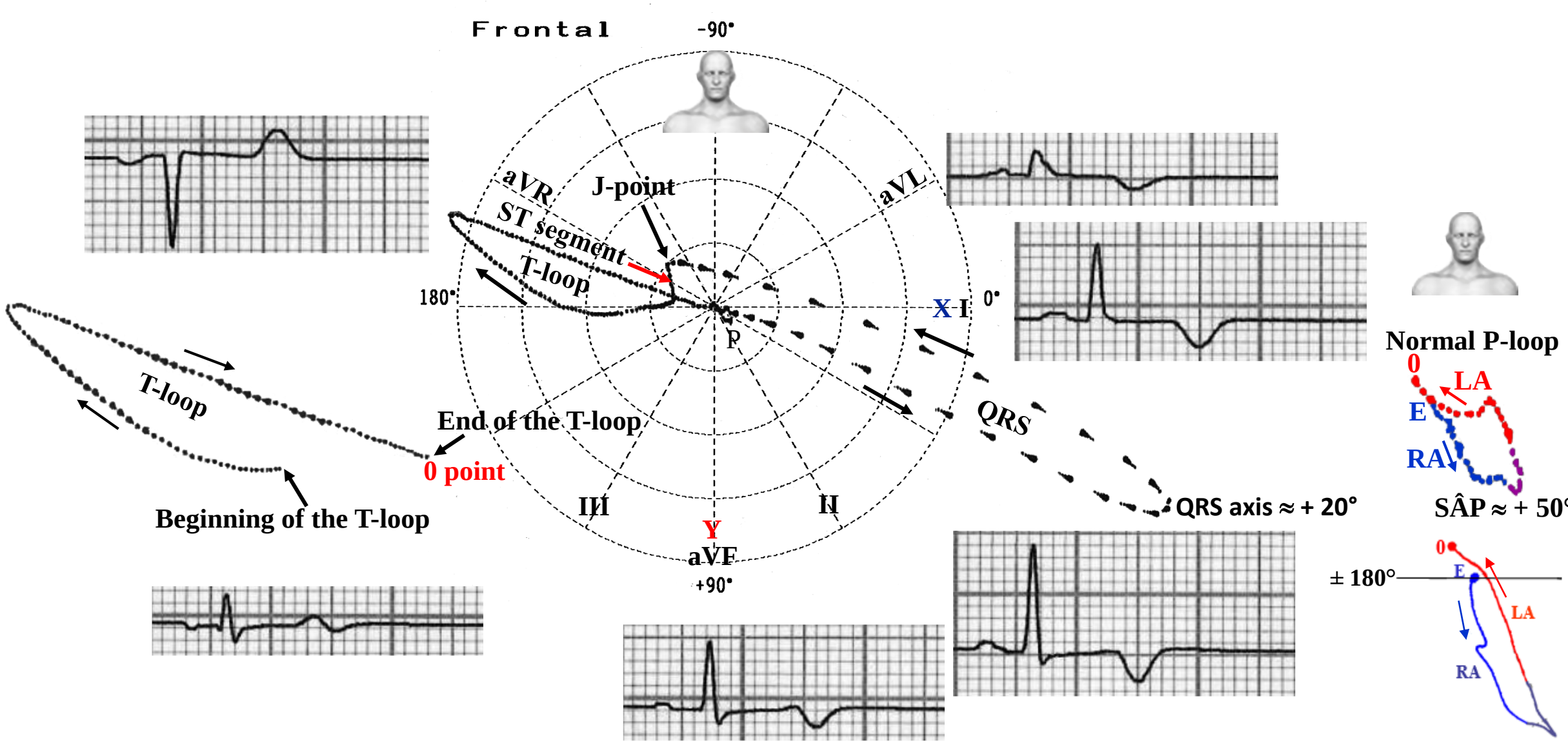
Final Comments

"Dream without limits and believe with all your might"

Andrés & Helena Akemi

Like background in the photo the Alhambra Palace was one of the greatest architectural wonders of the world when it was created in the 13th and 14th centuries and remains so today. It is unlikely that any future civilization will ever be able to match the magnificence and mysticism of the Alhambra Palace - truly an extraordinary fairytale palace.

ECG/VCG correlation in the frontal plane



In the present case the end of the QRS loop (J-point) does not coincide with the beginning of the T-loop signaling the presence of depression or elevation of the ST segment See explanation in the next slide.

J-point: This point represents the end of ventricular depolarization and the beginning of repolarization. There is an overlapping of 10ms, corresponding to 25% of one small square of the 12-lead ECG (**Mirvis 1982**). In Vectorcardiography, the ST segment is usually not registered. It corresponds to 3 elements:

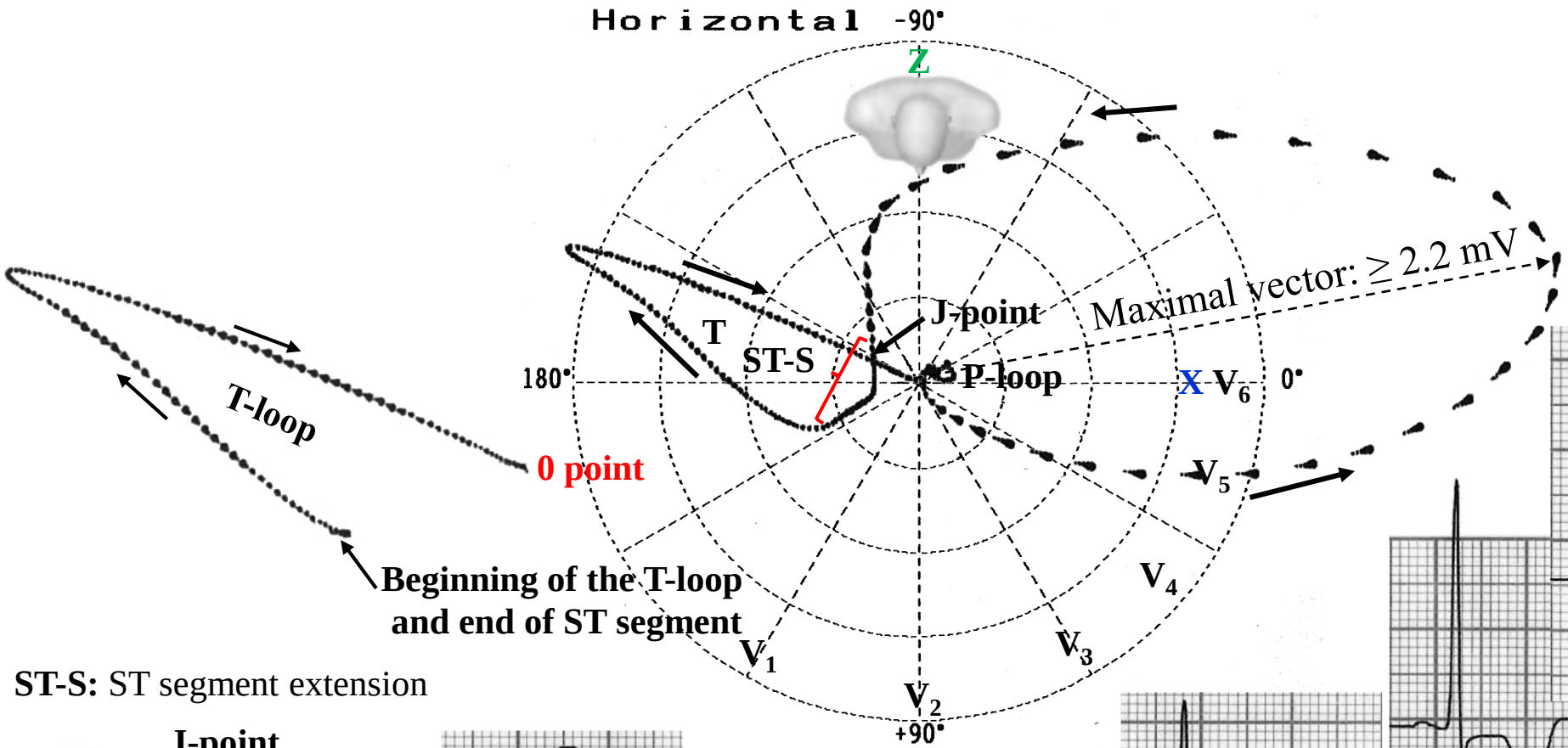
1) End of ventricular depolarization (QRS-loop);
2) Beginning of repolarization (ST segment) **when it does not present depression or elevation (differently of the present case)**, and
3) T wave onset. **(when it does not present depression or elevation)**

In situations where there is depression or elevation of ST segment, the J point does not coincide with the 0 point, and the greater or lesser distance between both points indicate the greater or lesser ST segment elevation or depression.

The phenomenon can be observed in:

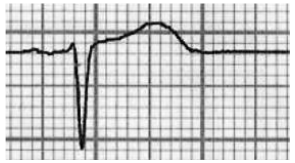
- Early Repolarization Pattern: In the inferior, lateral or inferolateral ECG leads, which may represent the inferior and/or left lateral ventricular myocardium.
- Acute Coronary Syndrome with ST segment Elevation: It caused by one of three pictures: ST elevation myocardial infarction (STEMI: ≈30%), non ST elevation myocardial infarction (NSTEMI, 25%), or unstable angina (38%)
- Pericarditis
- Variant angina or Prinzmetal angina
- Takotsubo cardiomyopathy “octopus trap”, transient apical ballooning syndrome, apical ballooning cardiomyopathy, stress-induced cardiomyopathy, Gebrochenes-Herz-Syndrome, broken heart, and simply stress cardiomyopathy
- Ap-HCM
- Brugada syndrome In the right precordial leads or when associated with ERP globally affected
- Arrhythmogenic right ventricular dysplasia/cardiomyopathy concealed forms and other miscellanea's.

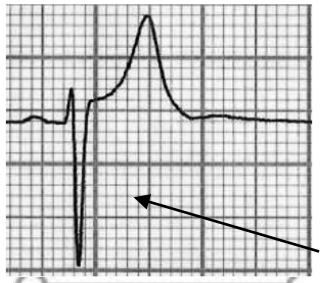
ECG/VCG correlation in the horizontal plane



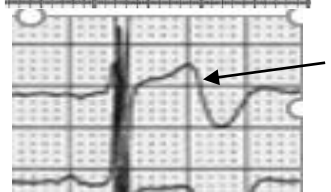
ST-S: ST segment extension

Beginning of the T-loop and end of ST segment

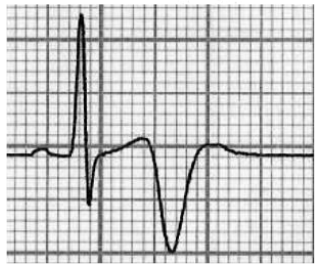


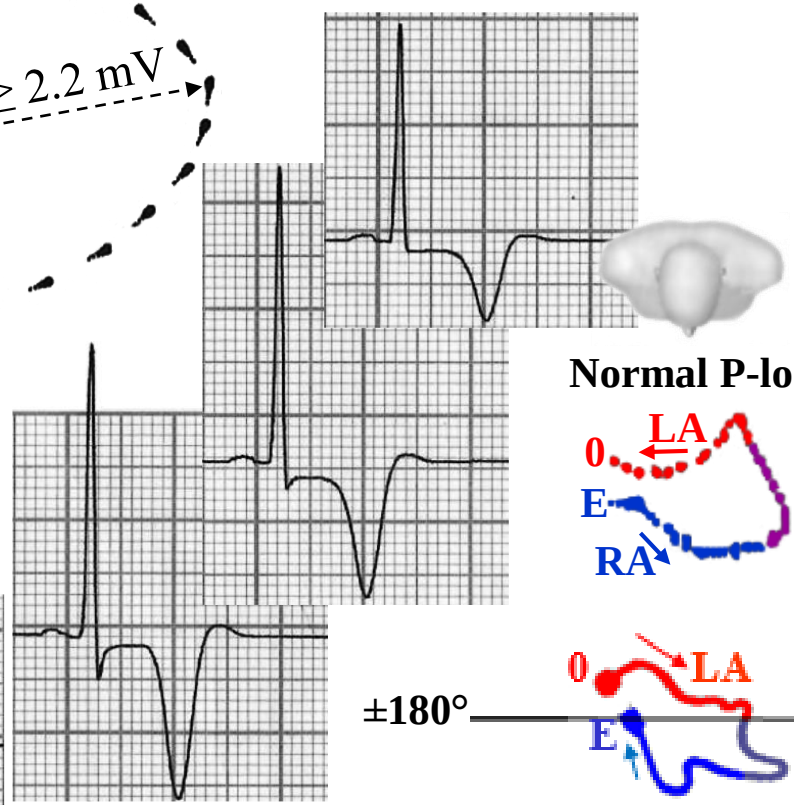


Misplace V_2 electrode



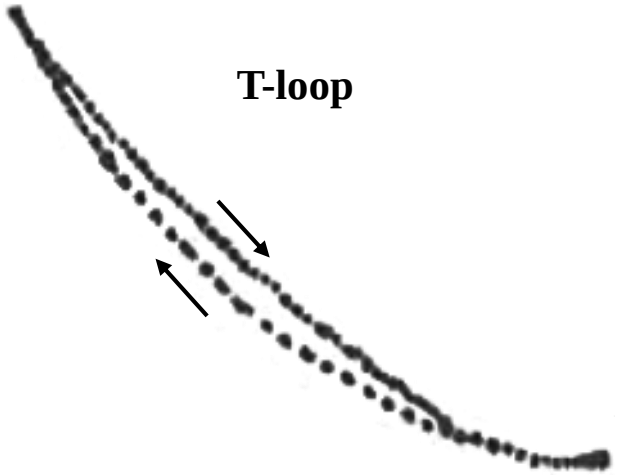
Correct position of V_2 electrode because the T-loop is located in the positive and negative hemifields of V_2 justifying the plus-minus polarity of the T-wave in this precordial lead.



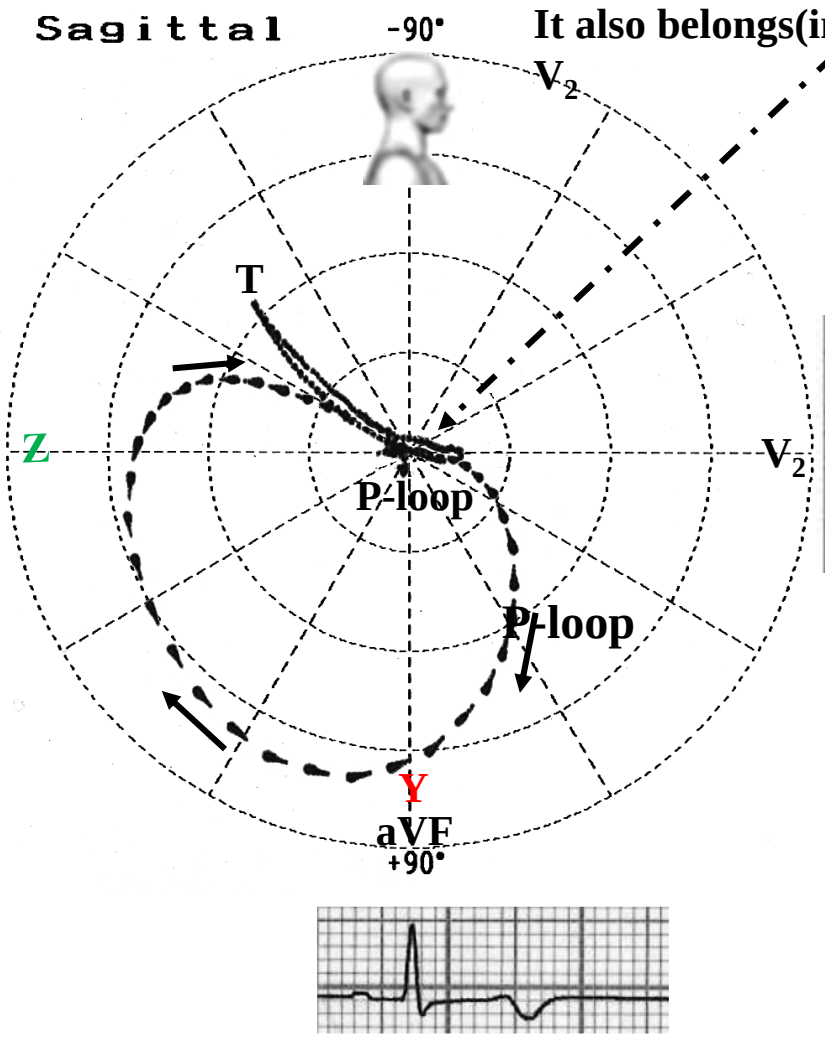


Normal P-loop

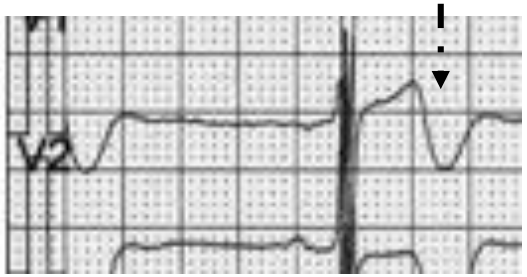
ECG/VCG correlation in the right sagittal plane



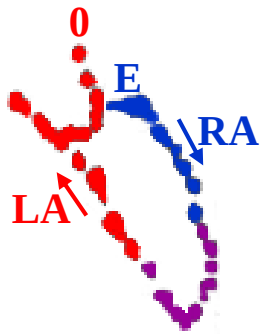
T-loop

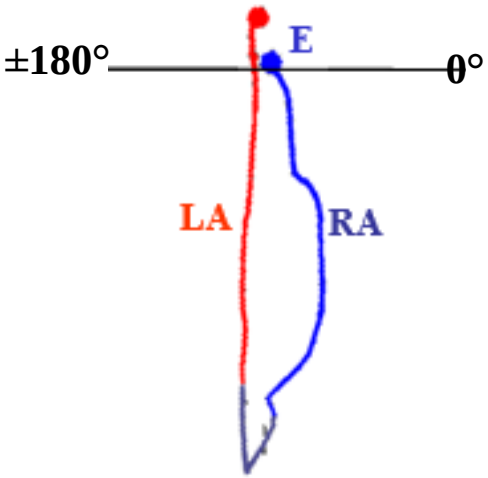


It also belongs(initial portion) to the T-loop: “plus-minus” T-wave in



Normal P-loop





Apical Hypertrophic Cardiomyopathy (Ap-HCM) "Yamaguchi syndrome"

Overview

Apical hypertrophic cardiomyopathy (Yamaguchi syndrome) is a rare variant of HCM in which left ventricular wall thickening is confined to the most distal region of the apex has been regarded as a phenotypic expression of non-obstructive HCM largely limited to Japanese patients. Nevertheless, since its original description in the 1970s (**Sakamoto 1976; Yamaguchi 1979**), several studies have been published outside of Asia regarding this entity, albeit insufficiently powered for robust conclusions regarding natural history, prognosis or long-term management strategies (**Kitaoka 2003; Eriksson 2002; Maron 1990**.) However, the implication of these studies has been that AHCM among non-Asian patients may represent an entirely different entity that carries a different prognosis compared to the Japanese variant. Ap-HCM has been rarely described in the Western world.

More recently, improved sensitivity of diagnostic modalities and increased diagnostic awareness have increased detection rates, suggesting that the prevalence outside of Asia may have been previously understated.

Hallmark features of Ap-HCM include deeply negative, "giant" T-wave inversions on electrocardiography and a "spade-like" configuration of the left ventricle on ventriculography.

Ap-HCM is found in 18% (**Kitaoka 2003**) to 25% of Japanese patients with hypertrophic cardiomyopathy (HCM), but outside of this population, it represents a markedly uncommon morphologic variant of HCM, with most reports suggesting a prevalence between 1% and 3% (**Maron 2008; Kitaoka 2003; Klues1995**). Ho et al estimated that represent around 5% cases of HCM in non-Japanese population.(**Ho 2015**) The basis for differences in the phenotypic expression of apical hypertrophy between Asians and non-Asians has not been elucidated. Although most patients with Ap-HCM experience minimal to no symptoms, presentations with a variety of signs and symptoms including atrial fibrillation, ventricular tachycardia and angina have been described (**Maron 2008**).

Typical features of Ap-HCM include T-wave inversion, particularly in the left precordial leads of the ECG, and a "spade-like" configuration of the left ventricular cavity at end-diastole on left ventriculography (**Sakamoto1976; Yamaguchi 1979**). Cardiac catheterization (and subsequent ventriculography) or cardiac magnetic resonance imaging (cMRI) is often needed to establish the diagnosis, which can be missed on two-dimensional transthoracic echocardiography. Advances in non-invasive imaging techniques, and in particular, cMRI, have led to its establishment as the gold standard diagnostic modality given the invasive nature of cardiac catheterization (**Patel 2002; Moon 2004**).

Electrocardiographic features

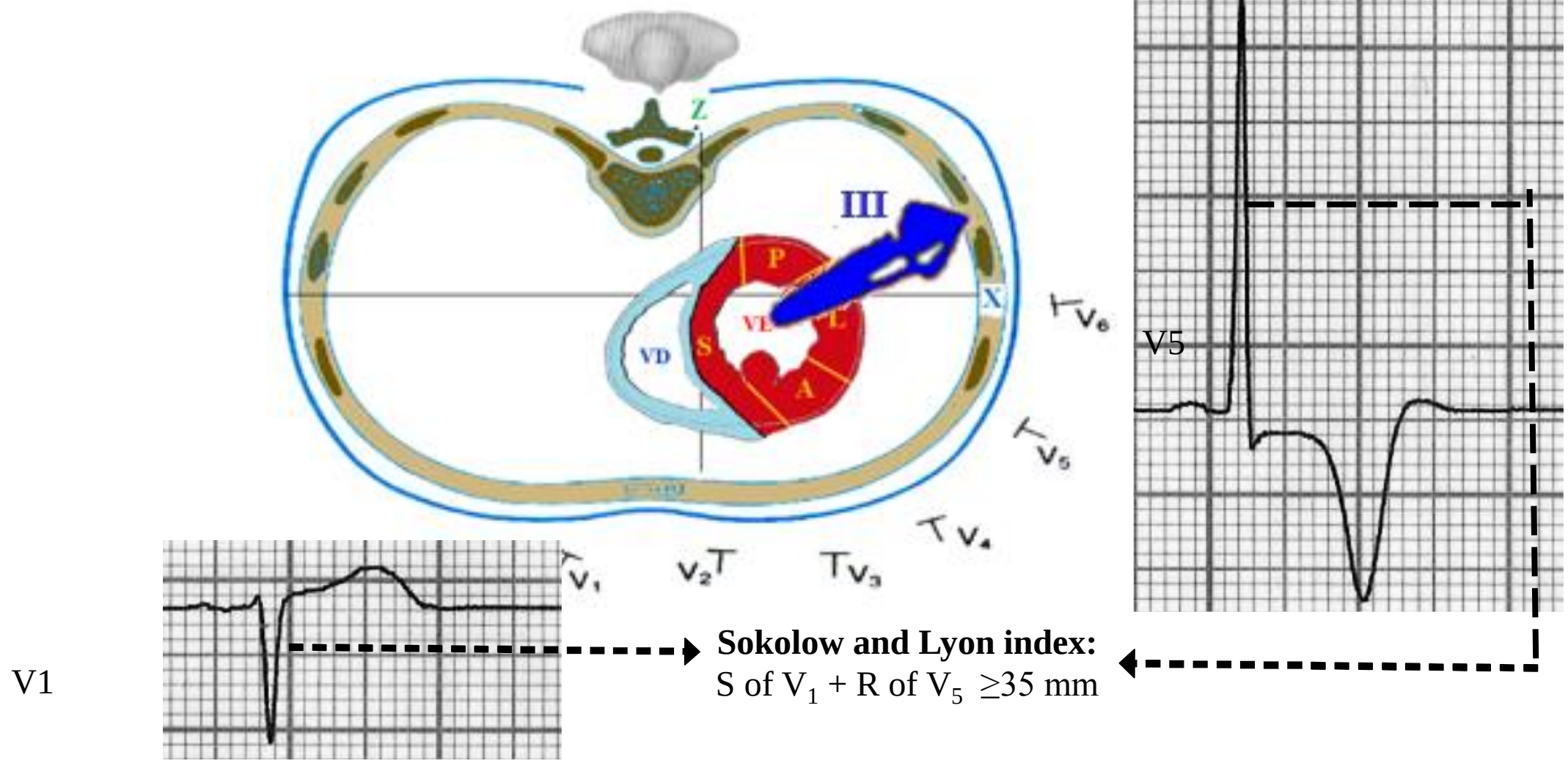
1. The typical features of Ap-HCM include giant negative T waves in the precordial ECG leads,
2. Eventual chronic ST segment elevation, and T wave inversion, in the anterolateral leads. Chronic ST segment elevation has been occasionally described in patients with HCM complicated with apical necrosis and aneurysm formation, but not in uncomplicated cases of Ap-HCM (**Pena Lado 1999**).
3. Eventual R-wave voltage and T-wave negativity progressively decreased in magnitude at serial ECGs
4. LVH criteria:
   - **Voltage criteria f**or LVH: (1) Sokolow and Lyon index: S of V1 + R of V5 ≥35 mm or 3.5 mV in adults older than 30 years; ≥40 mm between 20 and 30 years; and >60 mm between 16 and 20 years old; and >65 mm between 11 and 16 years old. (2) Cornell index (CI) 21: CI = R of aVL + S of V3 >28 mm in men or >20 mm in women, indicates LVH.
   - **Criteria based on wider QRS/ST-T angle** It is present in this cases). The QRS/ST-T angle >100° (it may reach 180°) has been called systolic pattern by Cabrera (**Cabrera 1960**) and in the Anglo-Saxon literature, LV strain pattern. In the right precordial leads, ST segment elevation is observed with upper concavity, of more than 0.1 mV, followed by positive T wave. In the left precordial leads, ST segment depression of upper convexity is observed, followed by negative asymmetrical T wave with the downsloping part slower than the upsloping one. Note: in the Romhilt-Estes score system for LVH (**Romhilt-Estes 1968**), the presence of strain pattern yields a score of 3, and if in use of digitalis only 1 (see next slide).
   - **Ventricular activation time in V5-V6 (≥50 ms)** or R-peak time; defined as the time elapsed since the onset of QRS until the peak of R

5. ST-segment elevation (≥ 1 mm) in V3 through V5 of ECG is identified apical aneurysms in Ap-HCM patients, the sensitivity was 66.7%, and the specificity was 98.7%. Their mean age is 60 ± 14 years (range: 23-77 years) adverse clinical event (annual event rate: 10.1%), including implantable cardioverter-defibrillator (ICD) implantations for VT/VF, an appropriate discharge of ICD for VT/VF, and nonfatal thromboembolic strokes, systolic dysfunction (LVEF<50%), non SCD or progressive heart failure was detected (**Ichida 20**14).
6. Ap-HCM patients with apical aneurism are characterized by following ECG features: increase in QRS-complex duration, QRS-complex fragmentation, decrease in QRS-complex amplitude (SV1+RV5-6, from 41±18mm to 26±11mm, p=0.015), ST-segment elevation in V4-V6, and positivisation of negative T waves in V3-V6 (**Pennacchini 2015**).
7. Rare symptomatic sustained VT
8. Atrioventricular nodal re-entrant tachycardia (**Candelario 2017**).

Point score system for LVE/LVH or Romhilt-Estes Score (Romhilt 1968)

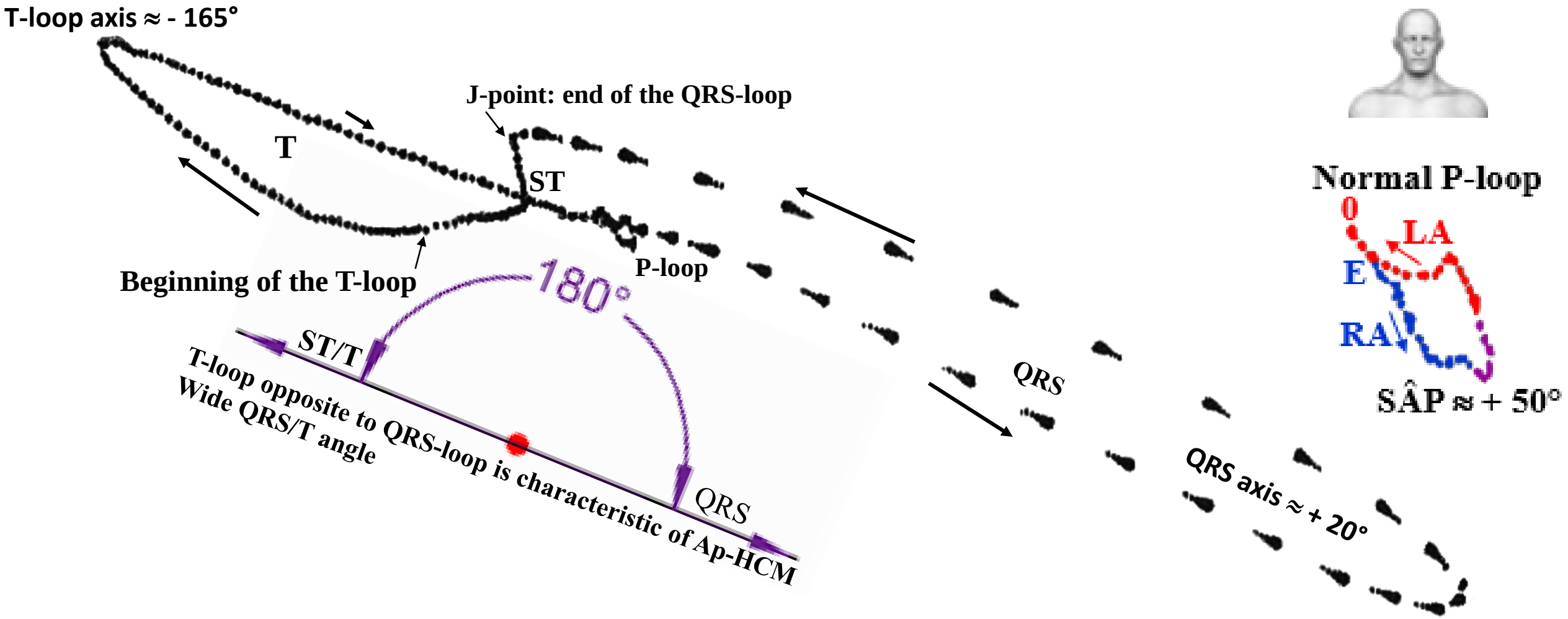
The authors attribute values from 1 to 3 points to the different existing criteria, 5 or more points: certain LVH; 4 points: probable LVH

ECG finding	Scoring
Voltage criteria Voltage Criteria (any of): R or S wave in limb leads ≥20 mm S wave in V1 or V2 ≥30 mm R wave in V5 or V6 ≥30 mm	3 points
ST-T vector opposite to QRS without digitalis ST-T vector opposite to QRS without digitalis	3 points 1 point
Left atrial abnormality; terminal negativity of the P wave in V1 >1 mm in depth with a duration of ≥0.042	3 points
Left axis deviation ≥30º	2 points
QRS duration >90 ms	1 point
Delayed ventricular activation time, R peak time or intrinsicoid deflection in V5 or V6 (>0.05 sec) or ≥50 ms	1 point



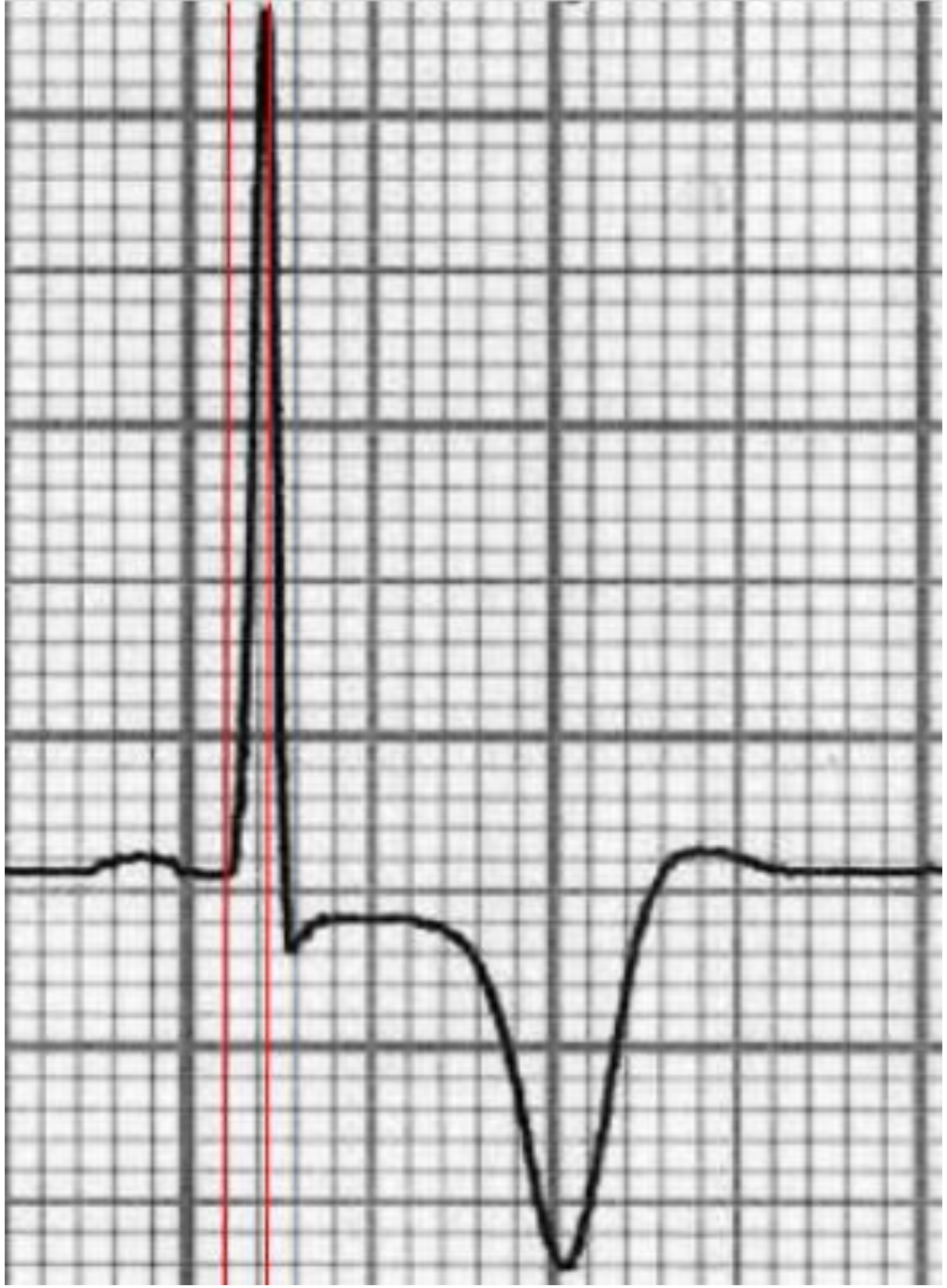
Positive Sokolow and Lyon index: S of V1 + R of V5 ≥35 mm or 3.5 mV in adults older than 30 years; ≥40 mm between 20 and 30 years; and >60 mm between 16 and 20 years old; and >65 mm between 11 and 16 years old.
In the present case SV1= 9mm + RV5= 26= 35mm.
In the left precordial leads, ST segment depression of upper convexity is observed, followed by negative asymmetrical T wave with the downsloping part slower than the upsloping one. Note: in the Romhilt-Estes score system for LVH (**Romhilt-Estes 1968**), the presence of strain pattern yields a score of 3, and if in use of digitalis only 1.III-

VCG in the Frontal Plane



LVH Criteria based on wider QRS/ST-T angle It is present in this cases). The QRS/ST-T angle >100° (it may reach 180°) has been called systolic pattern by Enrique Cabrera (**Cabrera 1960**) and in the Anglo-Saxon literature, left ventricular strain pattern. An ECG/VCG pattern of left ventricular hypertrophy(LVH)/strain have been shown to provide independent prognostic information.

In hypertension, the presence of LVH strain pattern on 12-lead ECG carries adverse cardiovascular prognosis because this pattern is a marker of advanced LVH with myocardial cellular associated with increased interstitial fibrosis and with significant myocardial circumferential strain impairment demonstrated with a multi-parametric cardiac magnetic resonance study (**Rodrigues 2016**).



Ventricular activation time or R-wave peak time in V5-V6 = 50 ms = LVH

Vectorcardiographic features

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:

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.

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.

3) Dromotropic disorders: Frequent:

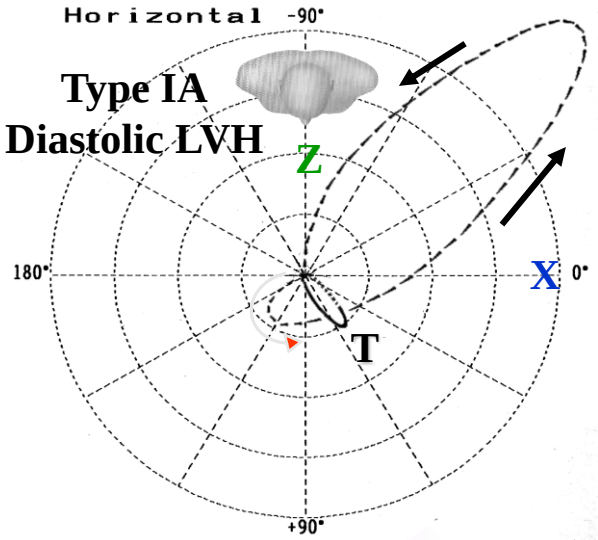
LAFB: maybe by inclusion of anterior fascicle in the septal muscle with disorder.

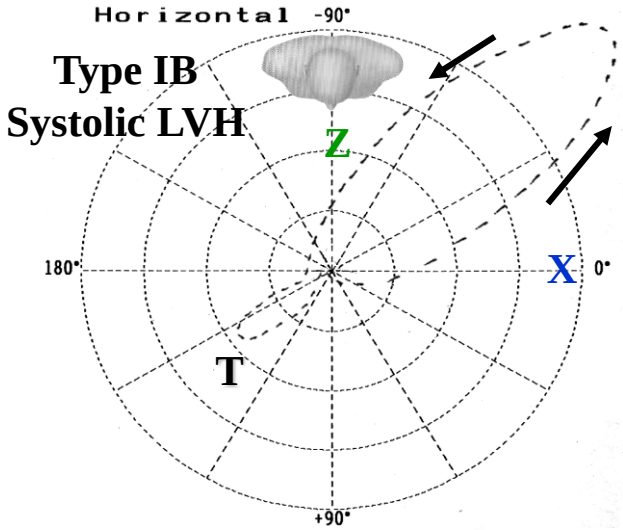
LSFB: particularly in NOHCM of the middle and low regions of the septum (see ECG/VCG number 200). Marked anterior shift of QRS loop is observed in the HP in absence of initial convexity to the right of the 20 ms vector.

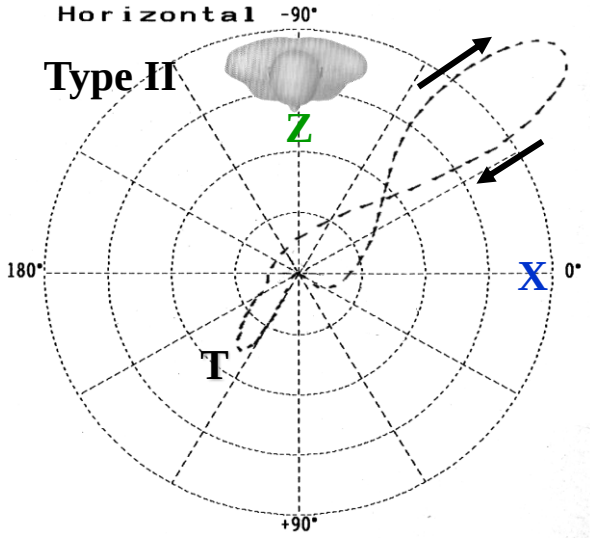
CLBBB: it is very frequent after septectomy surgery (80% of cases).

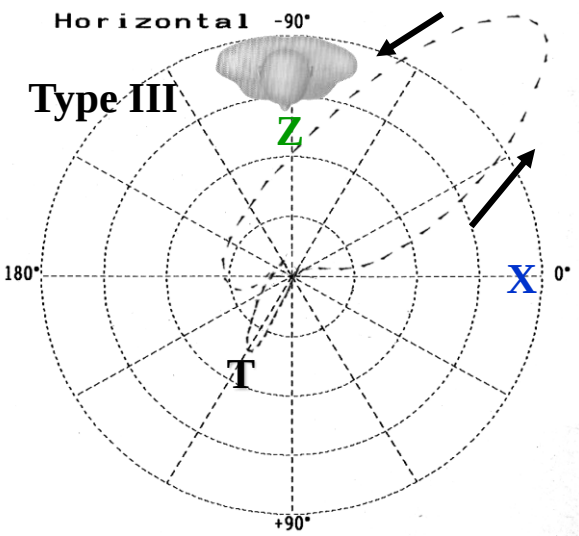
CRBBB: it is very frequent after Percutaneous Septal Ablation with absolute alcohol injected in the first septal perforating artery of the ADA.

The five vectorcardiography types of LVH in the Horizontal Plane: IA, IB, II, III and IV



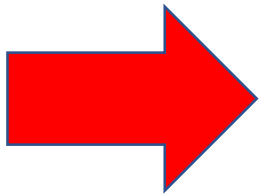


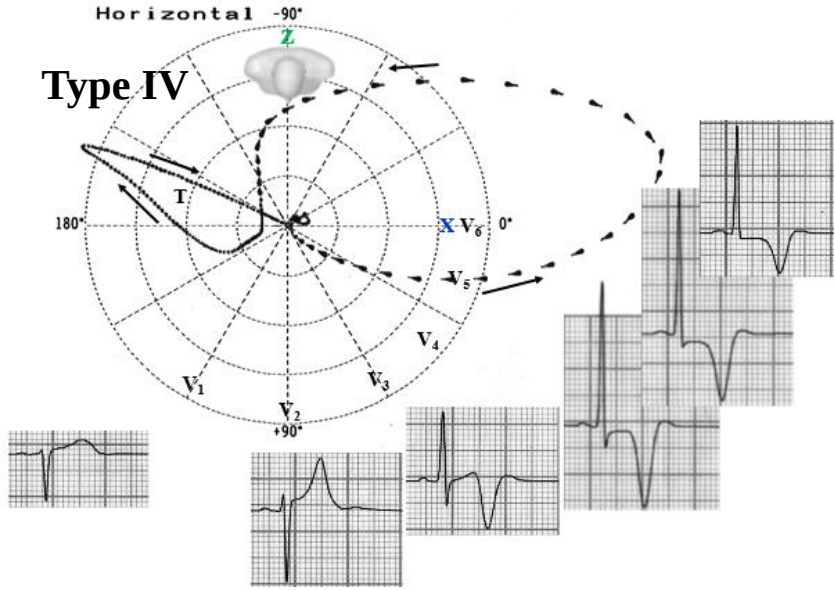




Eccentric LVH

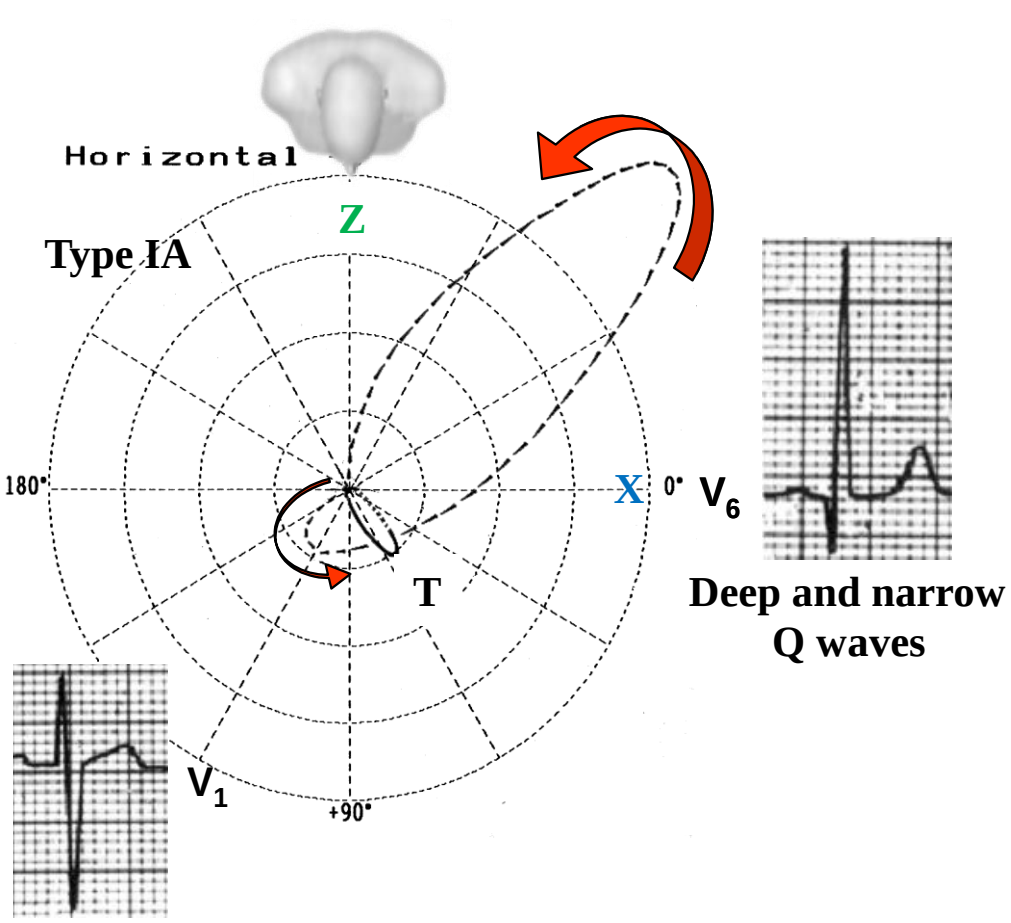
Concentric LVH

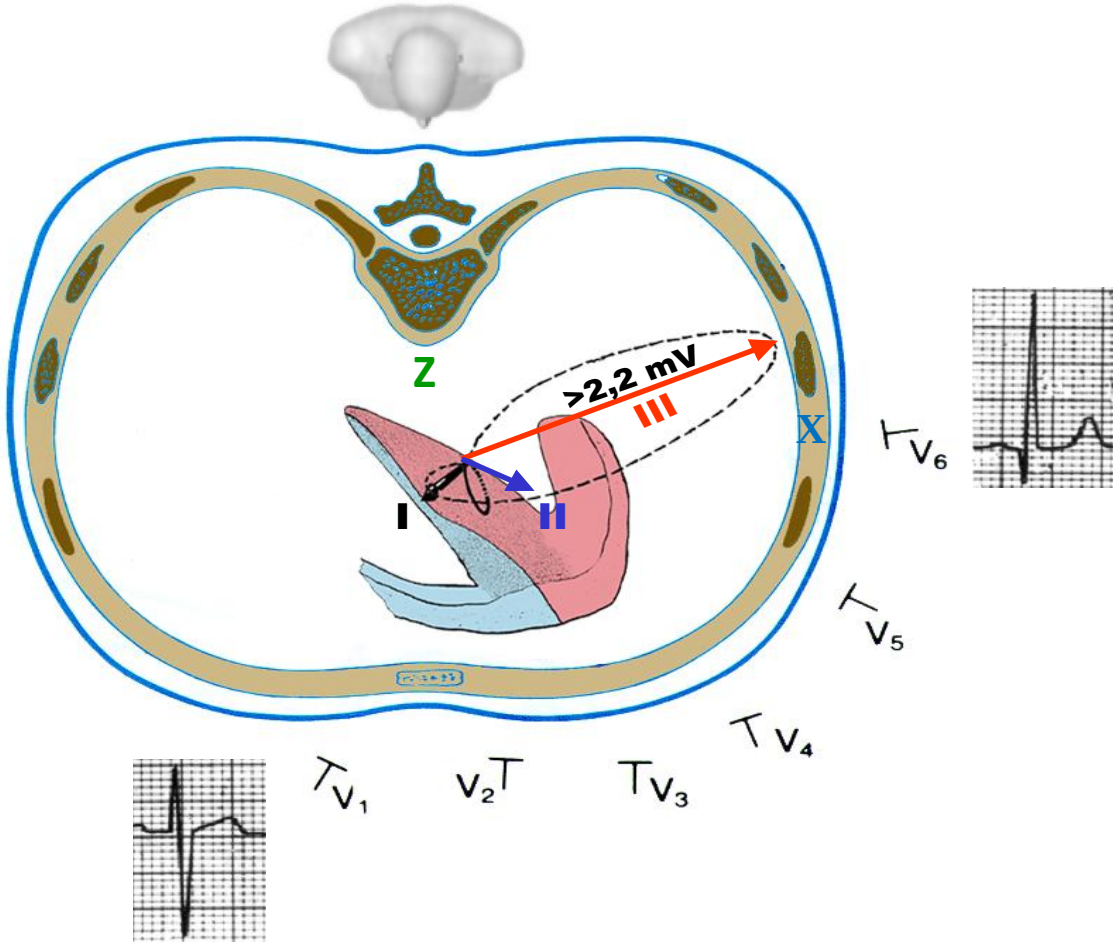




Apical hypertrophic cardiomyopathy Ap-HCM

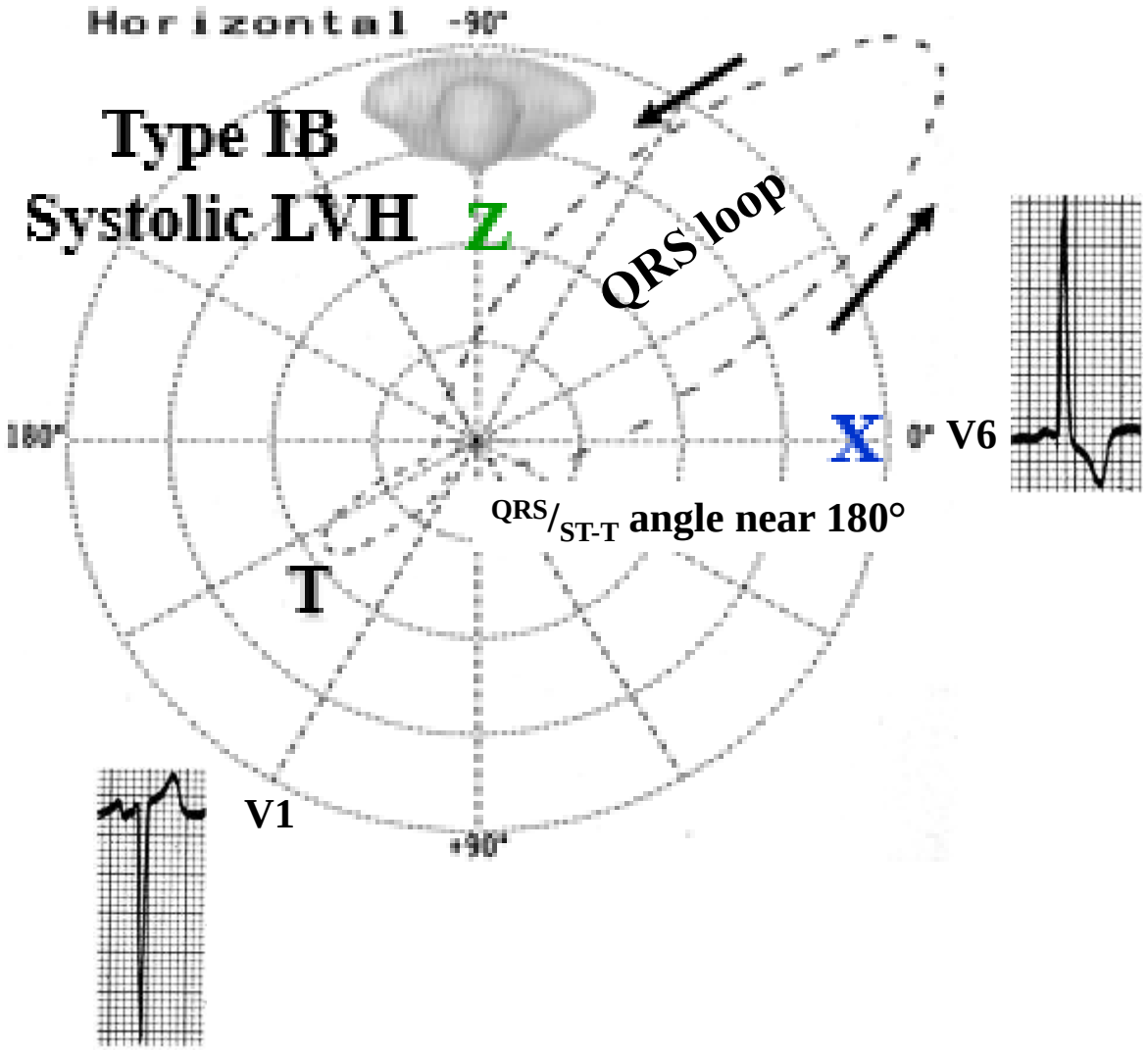
Vectorcardiographic characteristics of type IA LVH in the HP





1. 10 to 20 ms vector of QRS loop directed to the front and rightward with increased magnitude.
2. QRS morphology ovoid or elongated.
3. Counterclockwise rotation.
4. QRS loop predominantly located in the the left posterior quadrant.
5. Left ventricular maximal vector >2 mV.
6. “Clean” deep and narrow Q waves in left leads I, aVL, V5-V6 and eventually in inferior leads; consequently the I_{AM} vector is increased.
7. Frequent ST segment elevation concave to the top followed by positive T wave in left leads.
8. T loop with polarity concordant with the precedent QRS complex.

Systolic, concentric ECG/VCG LVH in the HP: VCG type IB



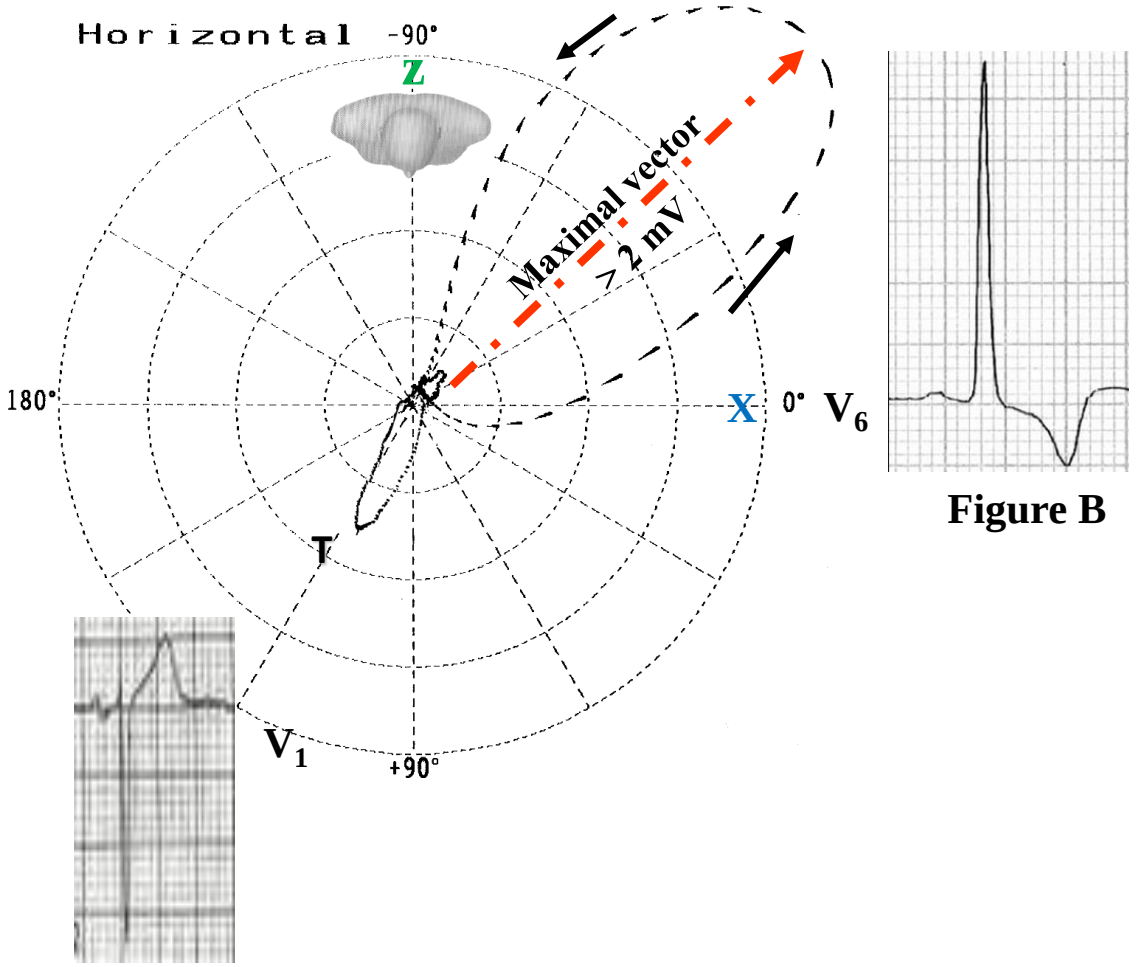
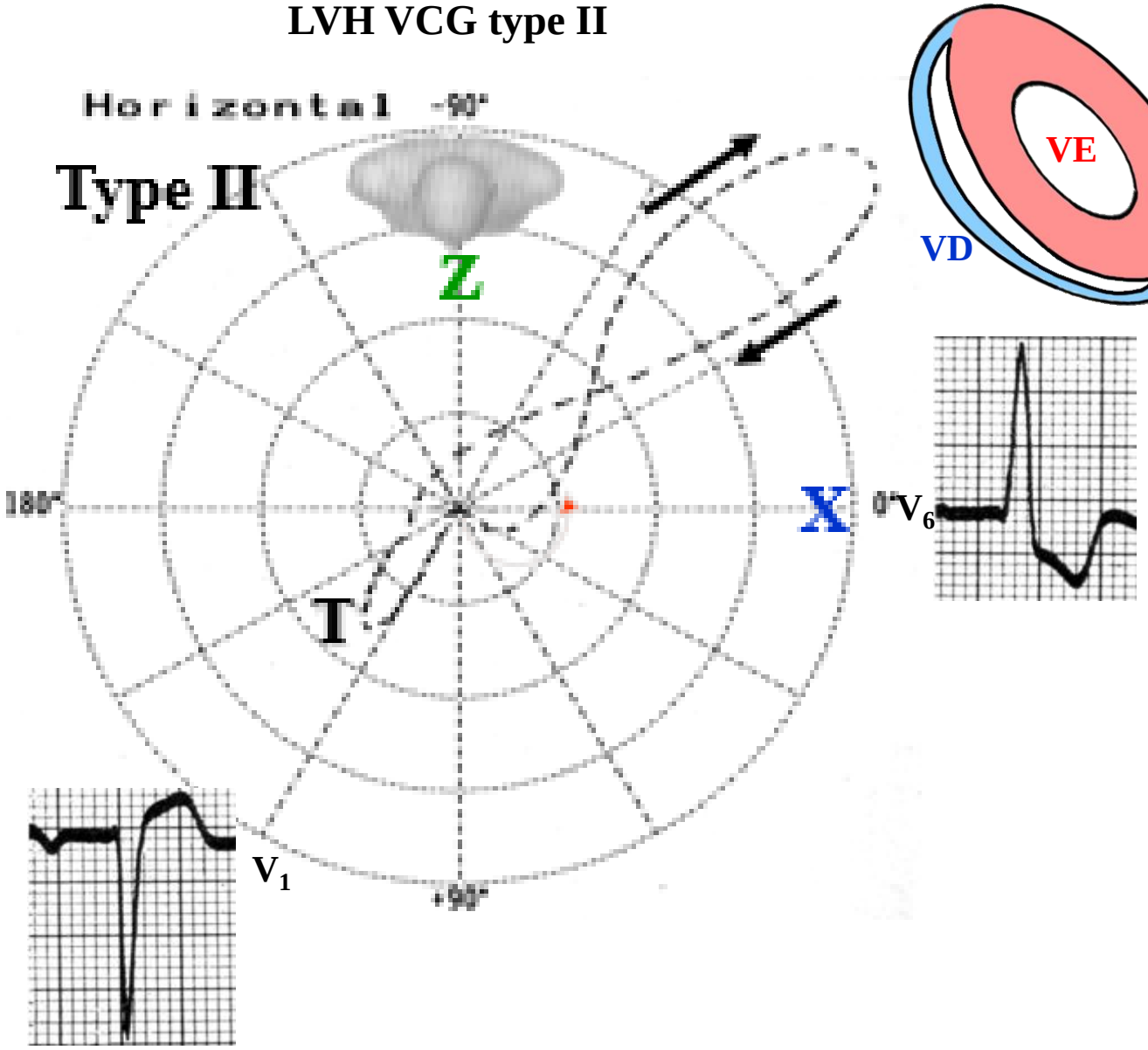


Figure B

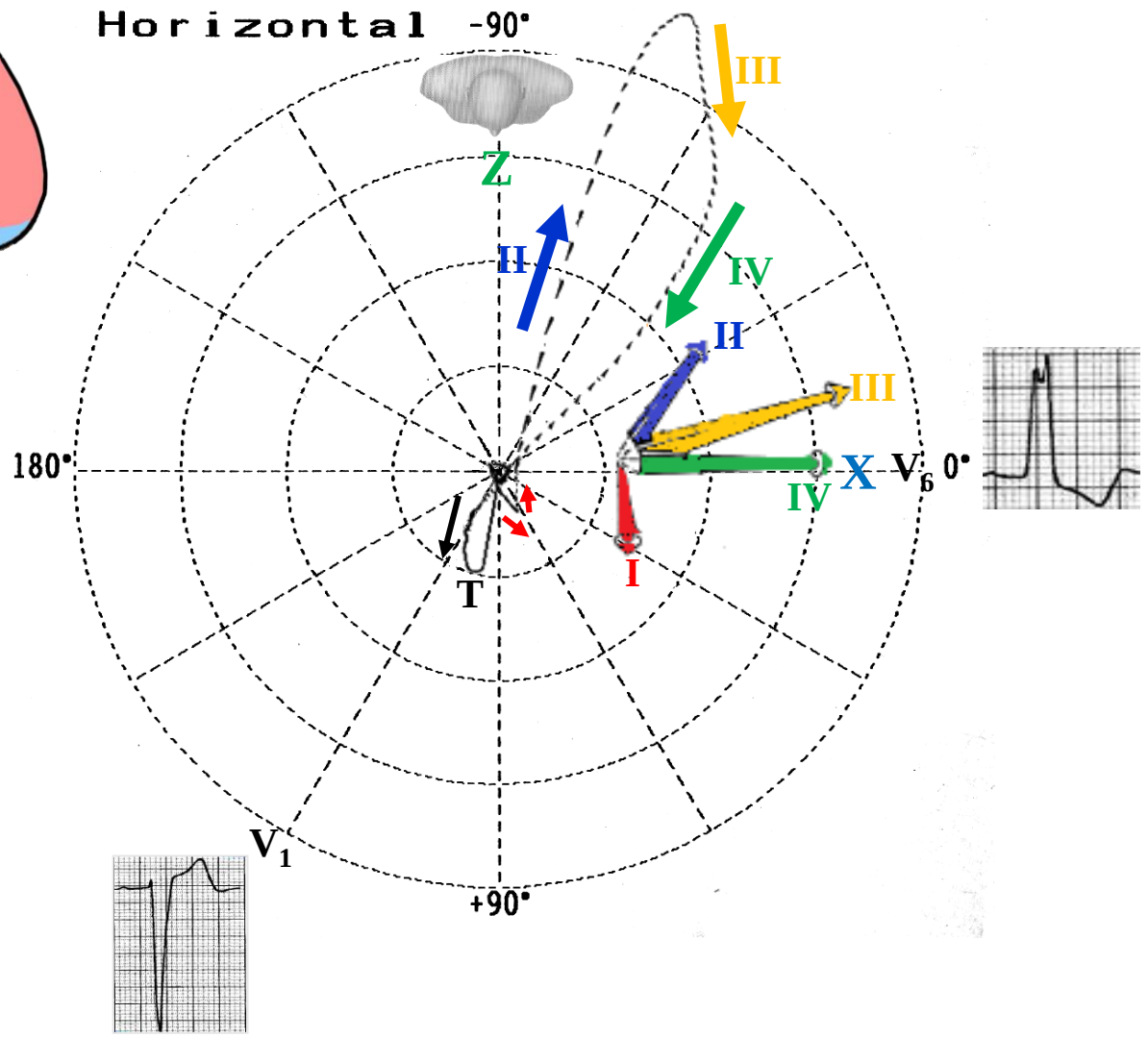
LVH Type IB: vector of initial 20 ms of QRS loop heading to the front and the left, oval morphology, counterclockwise rotation, location predominant in left posterior quadrant and maximal vector of increased magnitude: >2 mV. Characteristically the ST segment and T wave are opposite related to QRS polarity (strain pattern). The T wave remains asymmetric with slow initial ramp and rapid terminal ramp (Figure B). T loop opposite to QRS loop (not matching) heading to the front and the right: $^{QRS}/_{ST\text{-}T}$ angle near 180°.

VCG differential diagnosis between LVH type II from genuine CLBBB

LVH VCG type II

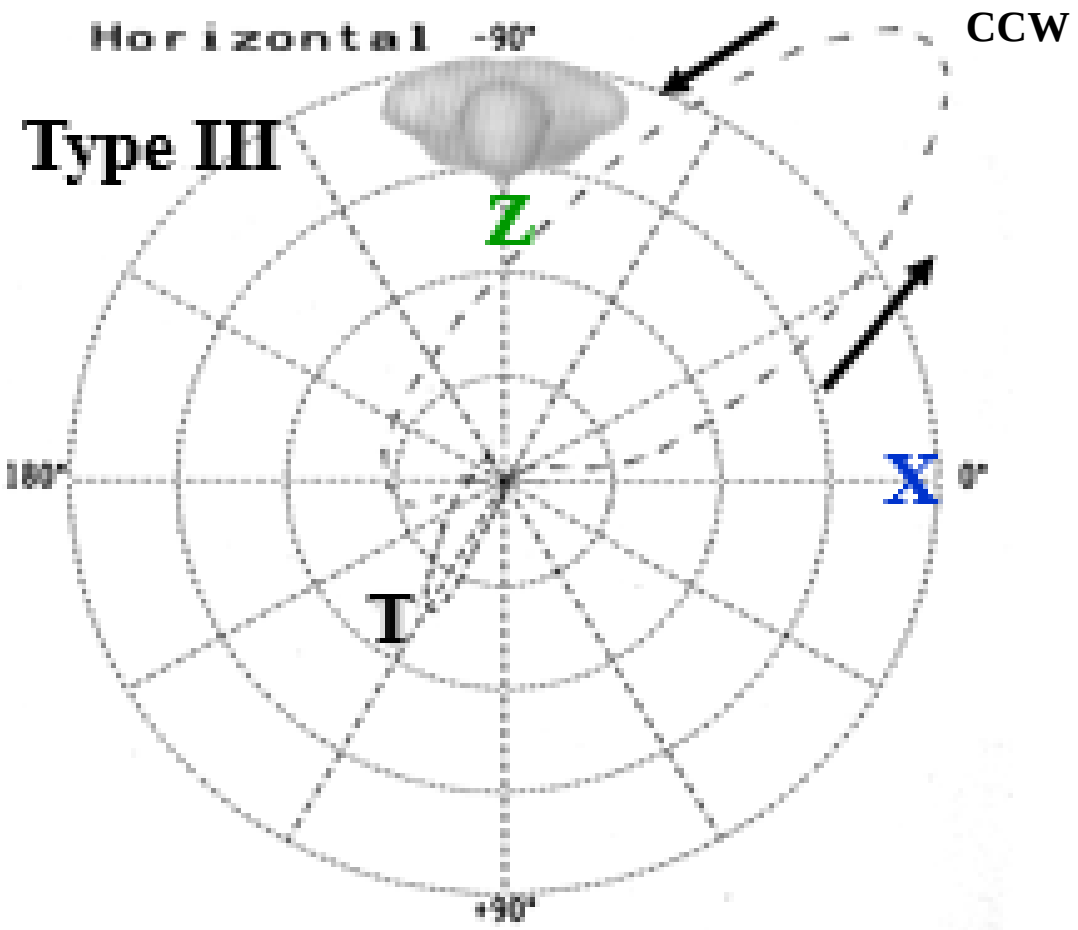


Genuine CLBBB



Absence of middle final conduction delay of QRS loop (dashes closer to one another).	Characteristic middle final conduction delay of QRS loop (tears closer one to another) (vector IV).

Left Ventricular Hypertrophy vectorcardiographic type III

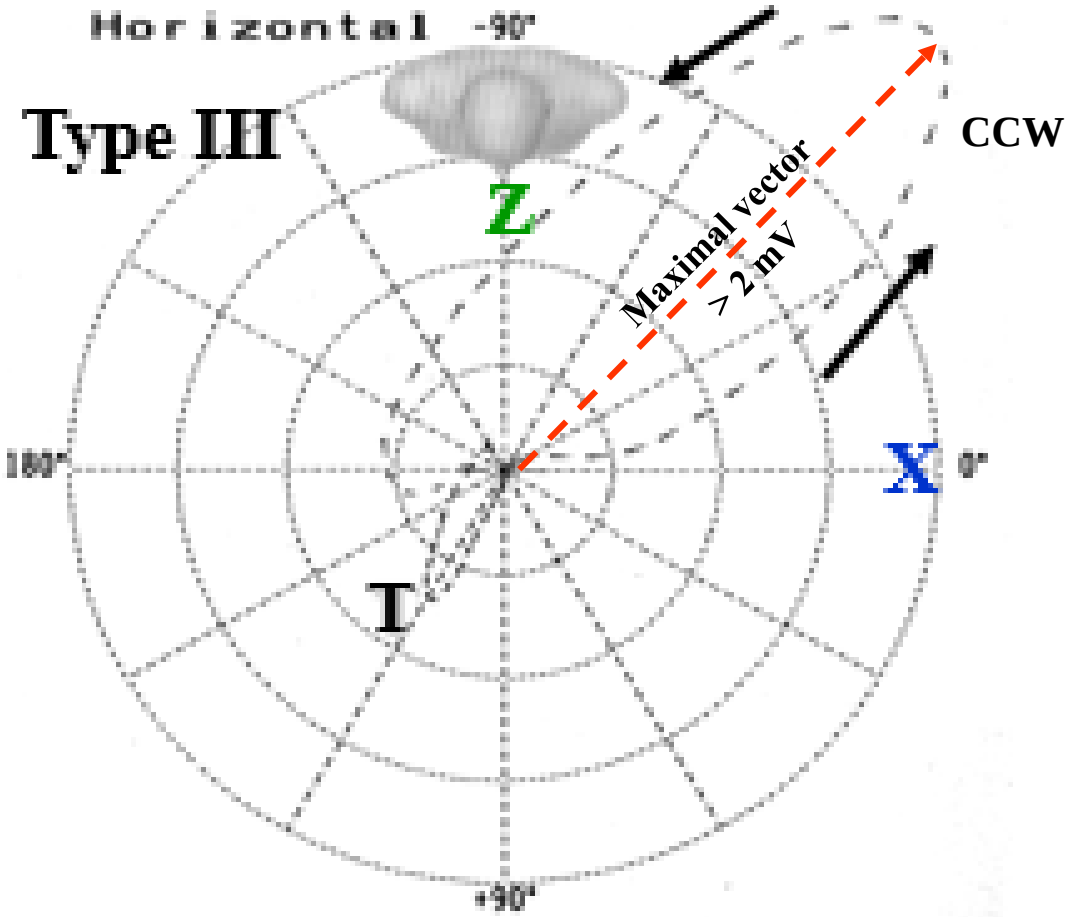


Type III: This is the variant frequently found in LVH and high blood pressure characterized by initial vectors heading to the right and discretely to the front, counterclockwise rotation (CCW) (inverted) simulating anterolateral myocardial infarction. Narrow aspect and QRS loop located mostly in the left posterior quadrant.

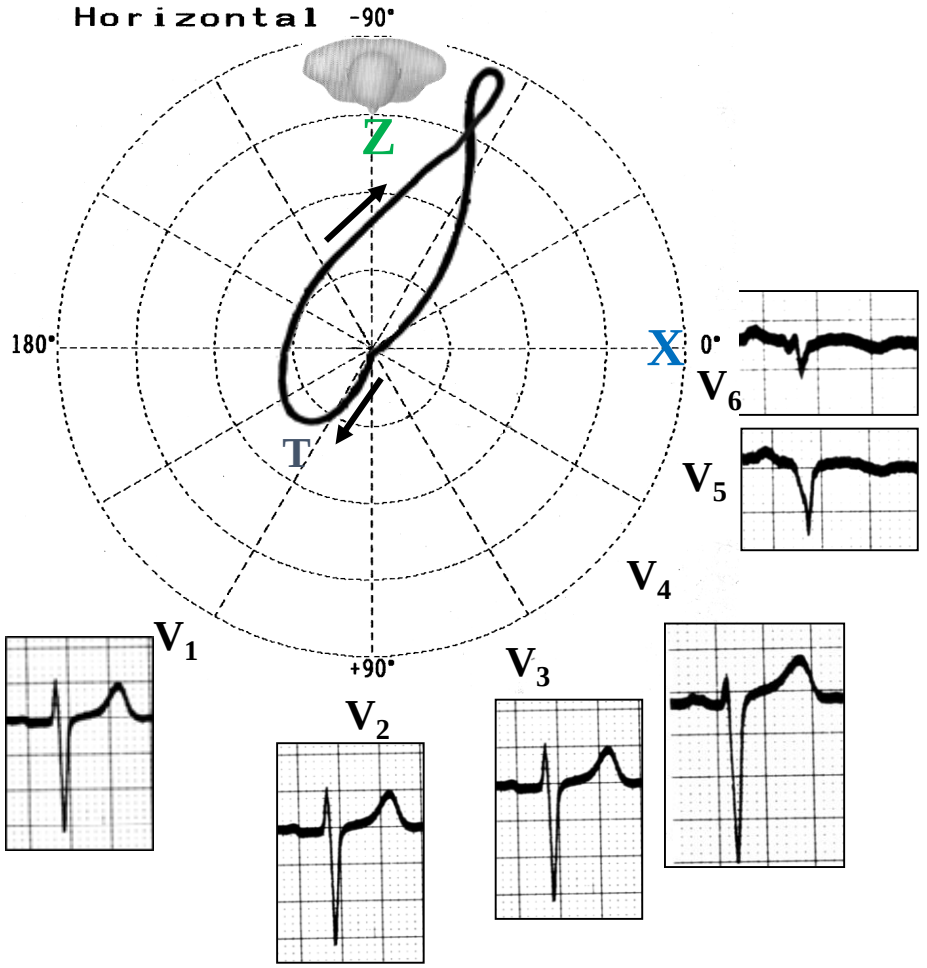
T loop opposite: located in the anterior right quadrant.

VCG differential diagnosis between type III LVH from Anterolateral Myocardial Infarction

Type III LVH of VCG



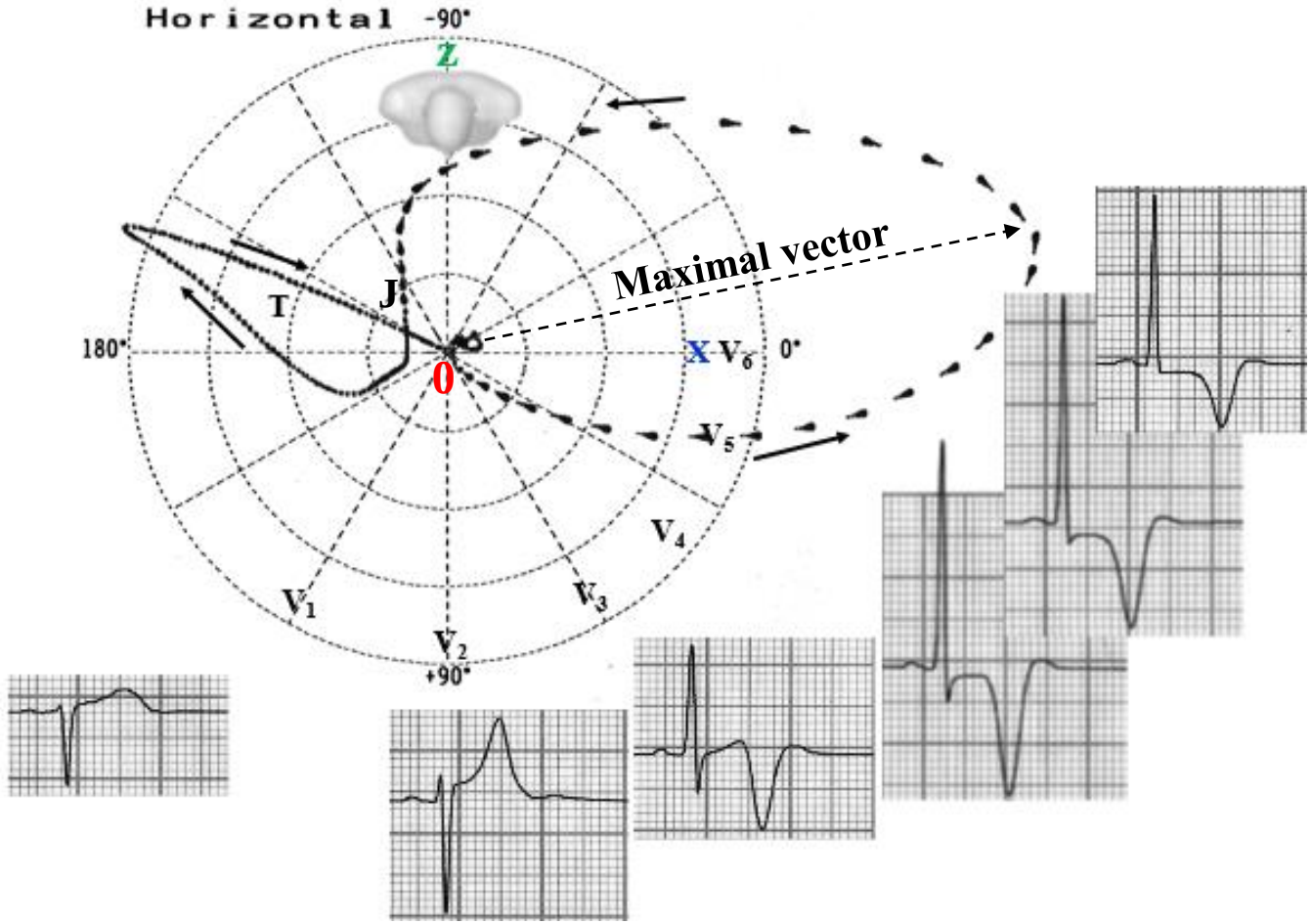
Anterolateral Myocardial Infarction



QRS loop with CCW rotation LV Maximal Vector >2 mV Rs pattern in the left leads	QRS loop with CW rotation LV Maximal Vector <2 mV QS or Qr pattern in the left leads

Left Ventricular Hypertrophy vectorcardiographic type IV. ECG/VCG correlation in Horizontal Plane

Type IV VCG LVH



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: ≥ 2.2 mV; final vectors located to the right and backward with the ST/T vector in the right posterior quadrant; J point not matching 0 point and located backward and to the right of the latter. Observation:**0 point**: it corresponds to the end of biatrial chamber activation and QRS loop onset (because PR segment does not exist, it is only a point) and the end of ventricular repolarization (T loop).

Transthoracic Echocardiogram

The Ap-HCM has asymmetric left ventricular hypertrophy mainly localized to the apex, with an apical wall thickness ≥15mm and a ratio of the apex to posterior wall ≥1.3 at end-diastole according to echocardiography (**Choi 2008**).

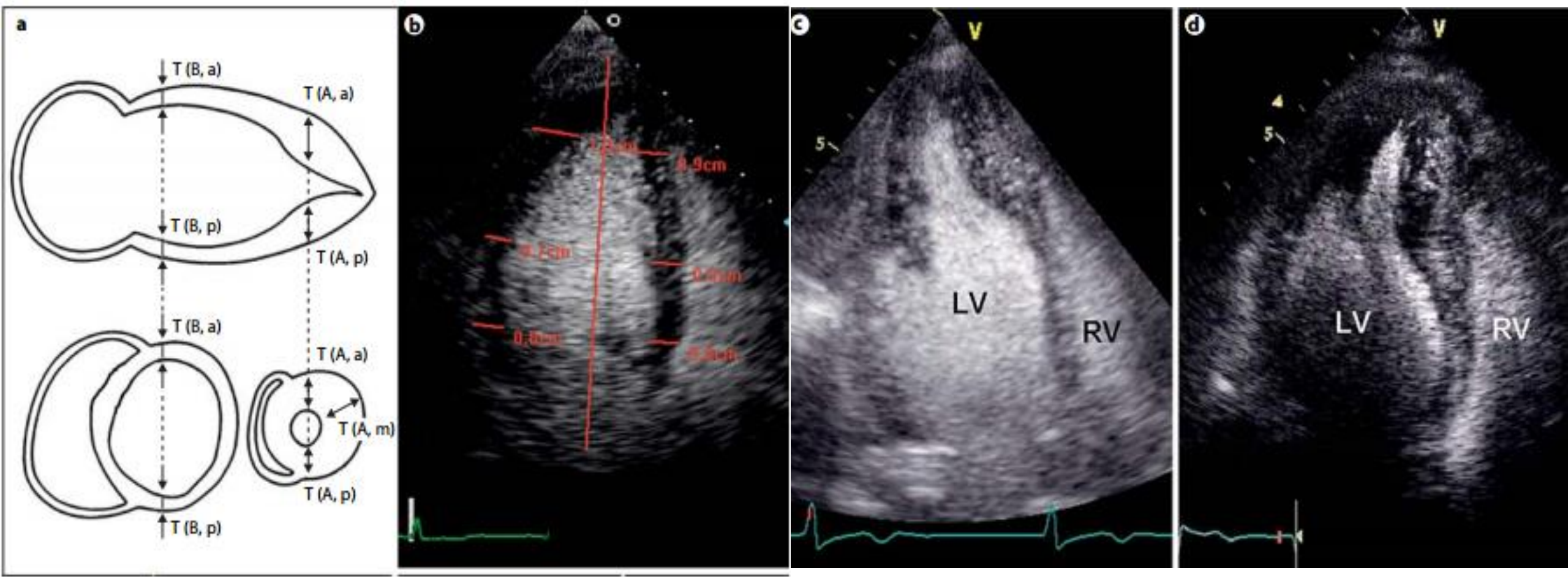
Echocardiographic types

Ap-HCM was classified into three types according to patterns of hypertrophy and incidence of atrial fibrillation :

1) **Pure focal**: left atrial volume index: 31; mitral annular velocity, E/E′: 13.3; Atrial fibrillation (AF): 5%
2) **Pure diffuse**: left atrial volume index: 15; mitral annular velocity, E/E′: 13.7; Atrial fibrillation (AF): 11%;
3) **Mixed type**: left atrial volume index: 41; mitral annular velocity, E/E′:16.1; Atrial fibrillation (AF): 23%

Ap-HCM contains three morphologically distinct phenotypes and detailed subtyping is important in the prediction of development

of atrial fibrillation, left atrial volume index and left ventricular longitudinal function. Ap-HCM may not represent a single disease entity. Detailed subtyping, based on the extent of hypertrophy, is important in the prediction of clinical or electro-mechanical characteristics such as development of atrial fibrillation, left atrial volume index (LAVI) and LV longitudinal function.



Three morphologic phenotypes of apical hypertrophic cardiomyopathy and ways to measure the thickness of each segment.

a) Schematic representation of way to measure the thickness.

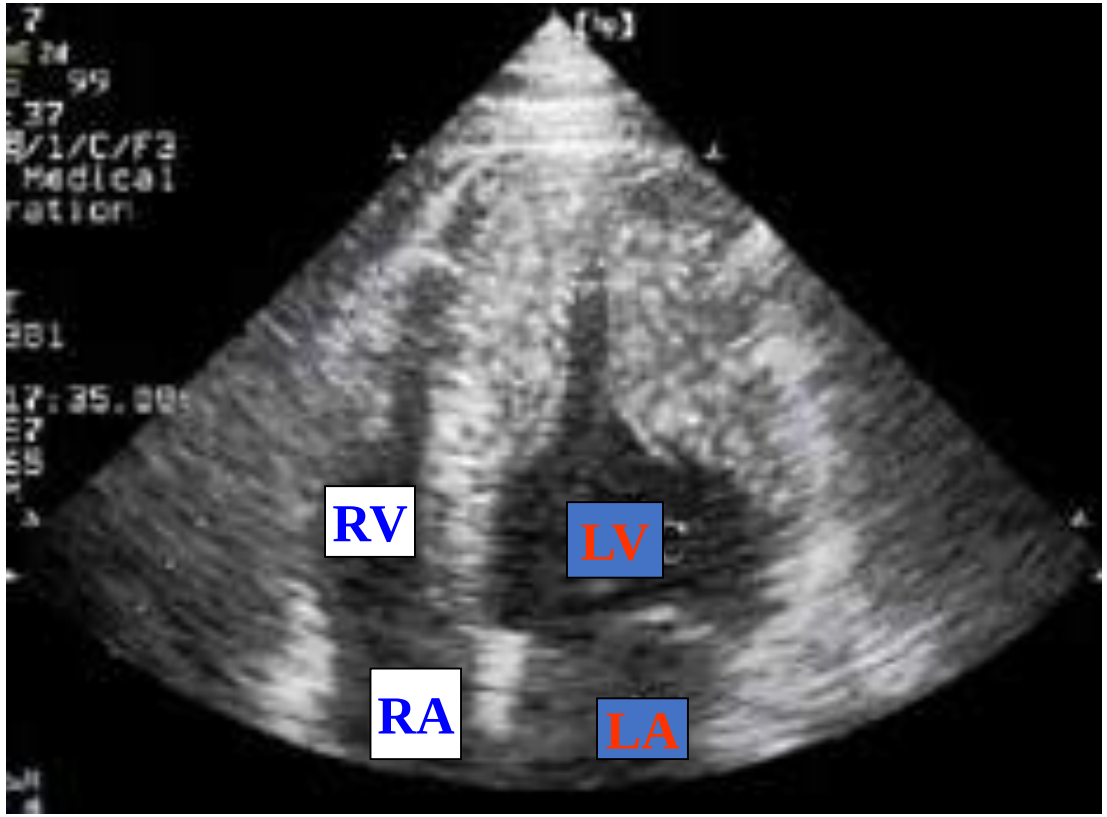
b) **Pure focal type** displayed hypertrophy confined to one or two apical segments.

c) **Pure diffuse Ap-HCM** displayed hypertrophy in more than two apical segments, and

d) **Mixed Ap-HCM** presented with coexistent hypertrophy of the interventricular septum not extending to basal segments but with the greatest wall thickness in the apical segment.

T (A, a) = Anterior wall thickness at apical level; T (A, m) = maximal wall thickness at apical level; T (A, p) = posterior wall thickness at apical level; T (B, a) = anterior wall thickness at basal level; T (B, p) = posterior wall thickness at basal level; LV = left ventricle; RV = right ventricle.

Transthoracic two-dimensional echocardiography

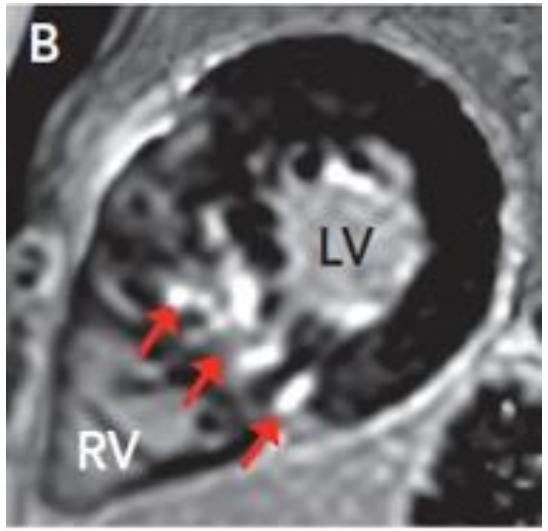


Two-dimensional echocardiogram. Apical 4-chamber view shows apical hypertrophy in the apical one third of the ventricle, with apical left ventricle cavity obliteration.

RV = right ventricle **RA** = right atrium **LA** = left atrium **LV**= left ventricular cavity

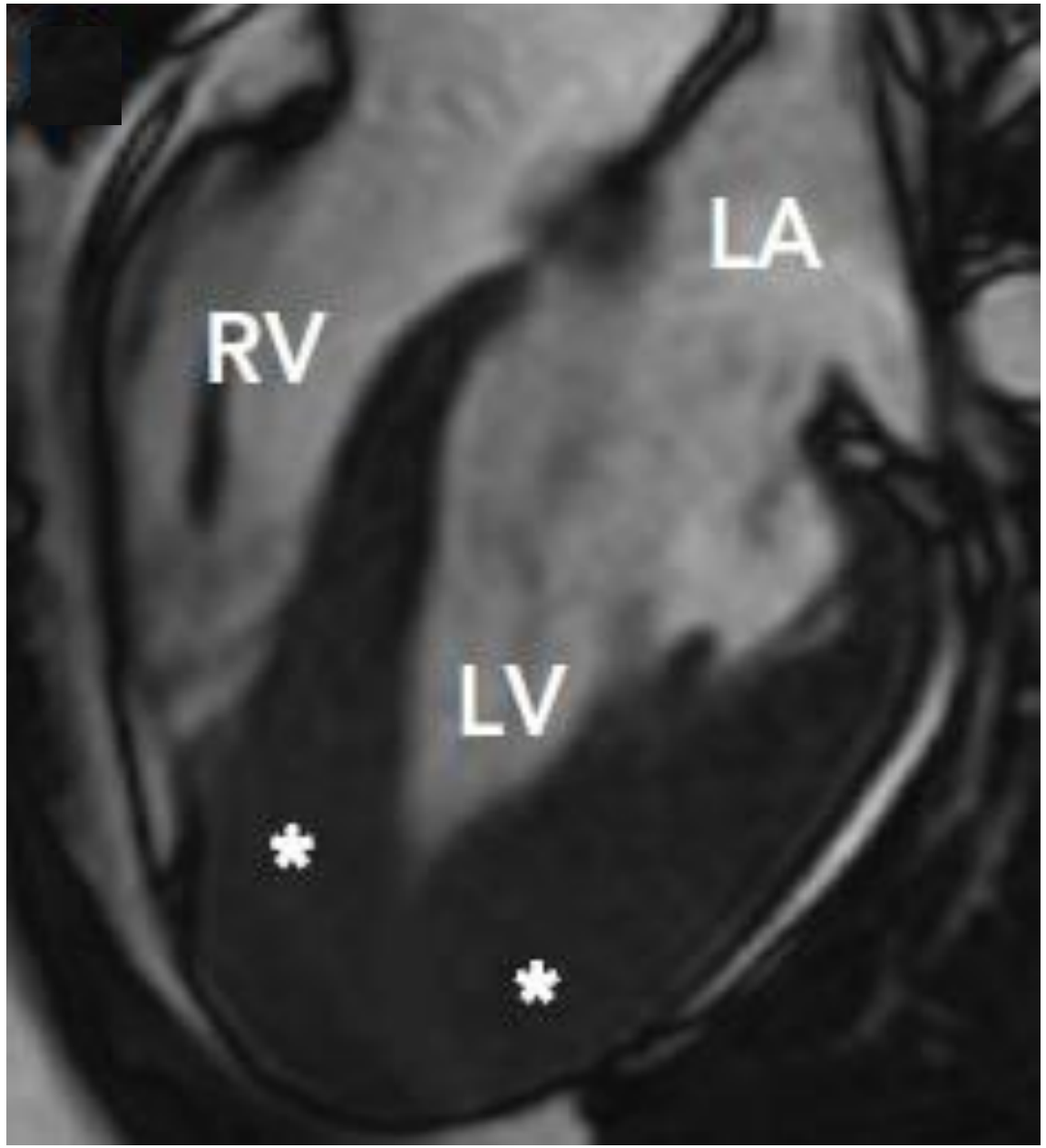
The Role of Cardiac MRI in the Diagnosis and Risk Stratification of Hypertrophic Cardiomyopathy

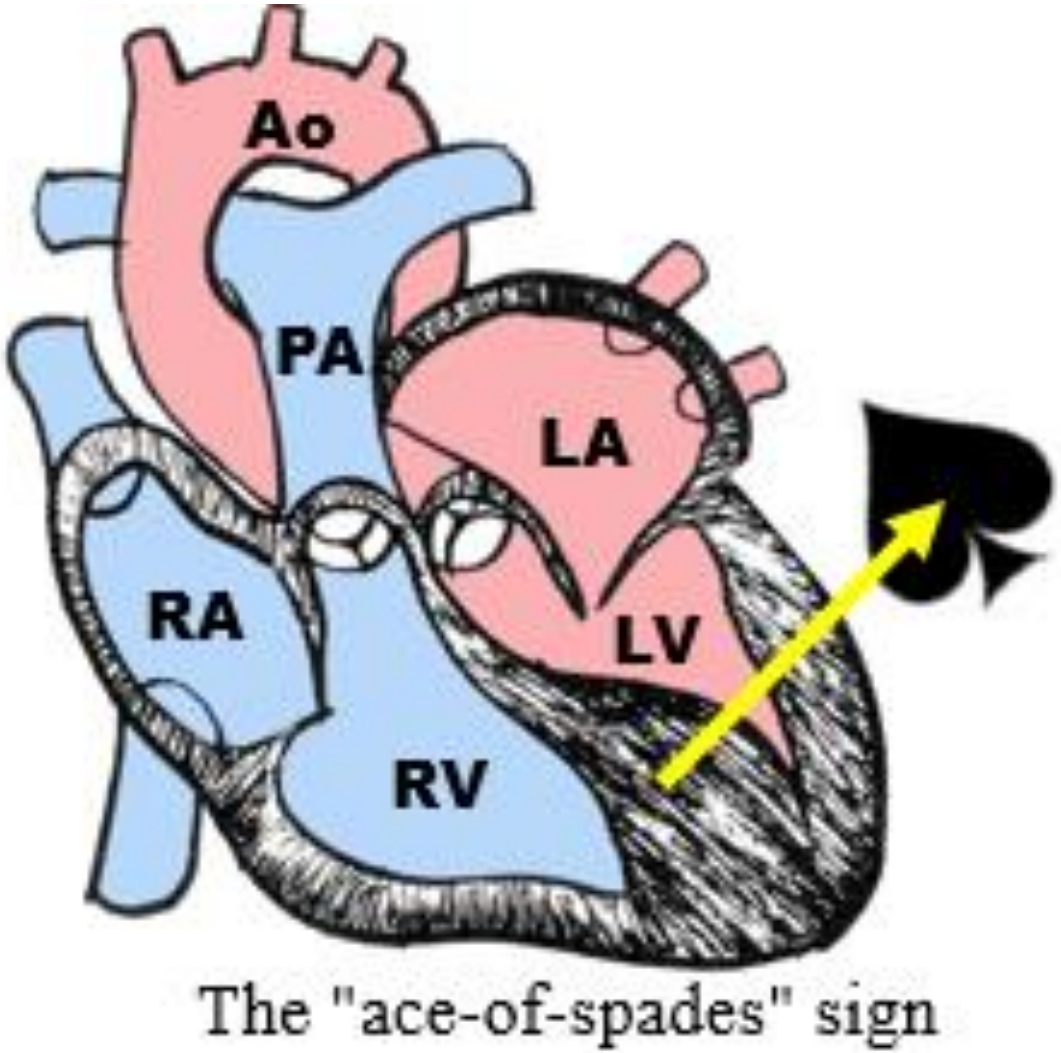
Hypertrophic cardiomyopathy (HCM), is the most common genetic cardiomyopathy, is present in one in 500 of the general population and is caused by over 1,400 mutations in at least 11 genes encoding the cardiac sarcomere (**Maron 2014**). The entity is characterized by substantial heterogeneity. Although the majority of patients with HCM remain asymptomatic with near-normal longevity, a small, but important, subset remain at risk for a wide range of clinical outcomes including SCD. Cardiovascular magnetic resonance (CMR), with its high spatial resolution and tomographic imaging capability, has emerged as an imaging modality particularly well suited to characterize the phenotypic expression of HCM. CMR helps in the diagnosis of HCM by identifying areas of hypertrophy not well visualized by echocardiography, providing more accurate wall thickness measurements and differentiating HCM from other causes LVH. CMR has led to the identification of novel subgroups of patients with HCM, including those with LV apical aneurysms (a subgroup at increased risk for ventricular arrhythmias and thromboembolic stroke), as well as abnormalities that contribute to LV outflow obstruction. Additionally, contrast-enhanced CMR with late-gadolinium enhancement (LGE) has recognized patients with extensive LGE (≥15 % LV myocardium) as individuals who may be at increased risk of SCD, independent of other high-risk features, with implications on management strategies including consideration for primary prevention ICD therapy. Extensive LGE is a potential novel primary risk marker that can also be used as an arbitrator when conventional risk assessment is ambiguous.



Example of a patient with extensive LGE throughout the septum (arrows) occupying 17 % of LV mass, and without other traditional risk markers. Based on extensive LGE, the patient had ICD placed for primary prevention of SCD with appropriate ICD discharge for VF 1 year later. These observations justify an expanded role of CMR in the routine clinical assessment of patients with HCM.

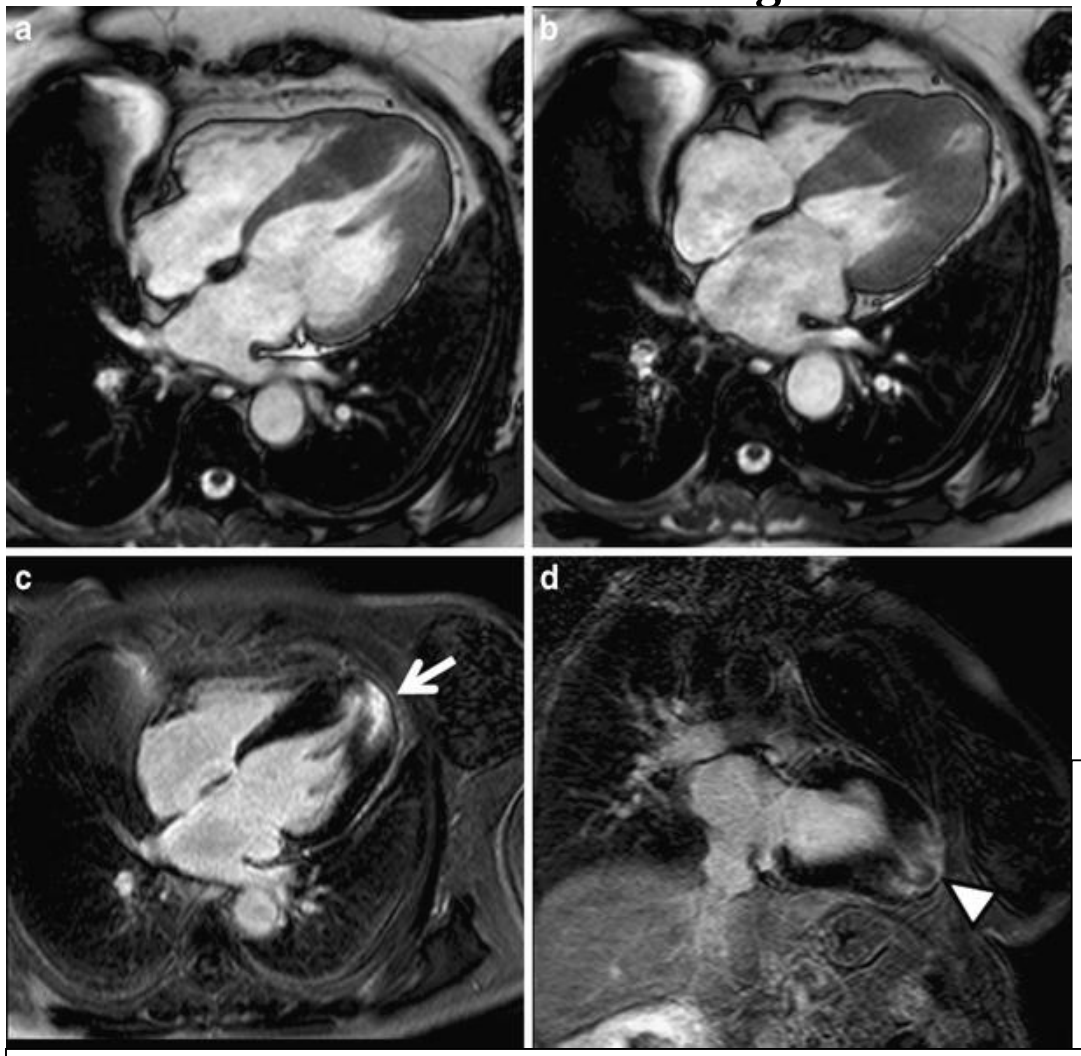
Apical Hypertrophic Cardiomyopathy



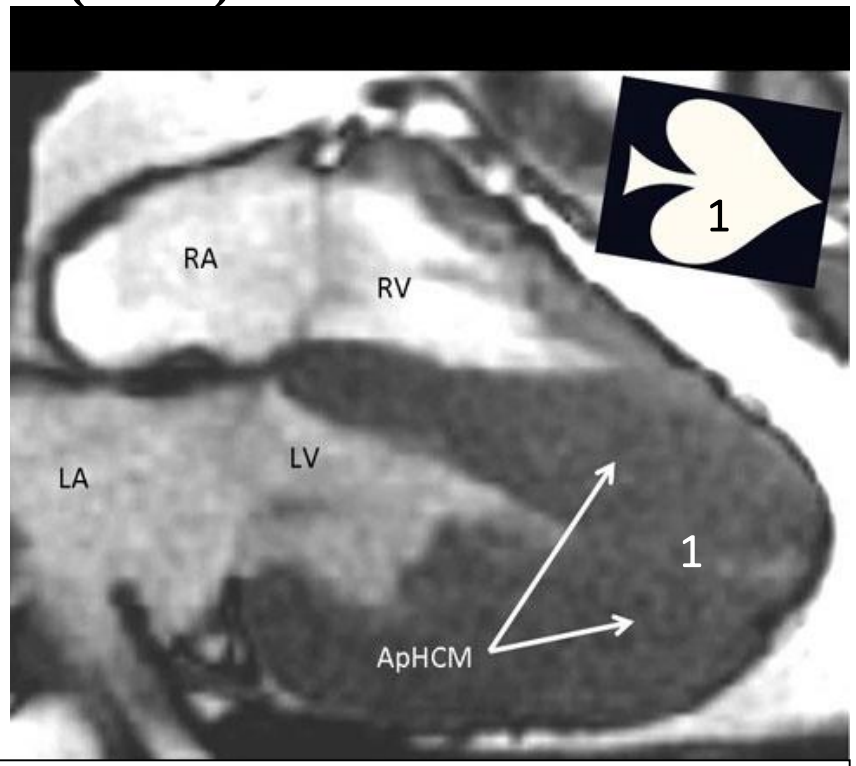


Ap-HCM: Four-chamber long-axis view demonstrating hypertrophy localized to the LV apex

Cardiovascular magnetic resonance (CMR) or Cine MRI



Apical form of hypertrophic cardiomyopathy. Cine MRI in the four-chamber view shows progressive thickening of the ventricular walls towards the apex in diastole (**a**) and systole (**b**). Late enhancement in a four-chamber view (**c**) and vertical long axis (**d**) demonstrates strong enhancement in the hypertrophied myocardium (arrow). Note the presence of "pseudo diverticulum" in the most apical myocardium (arrowhead).

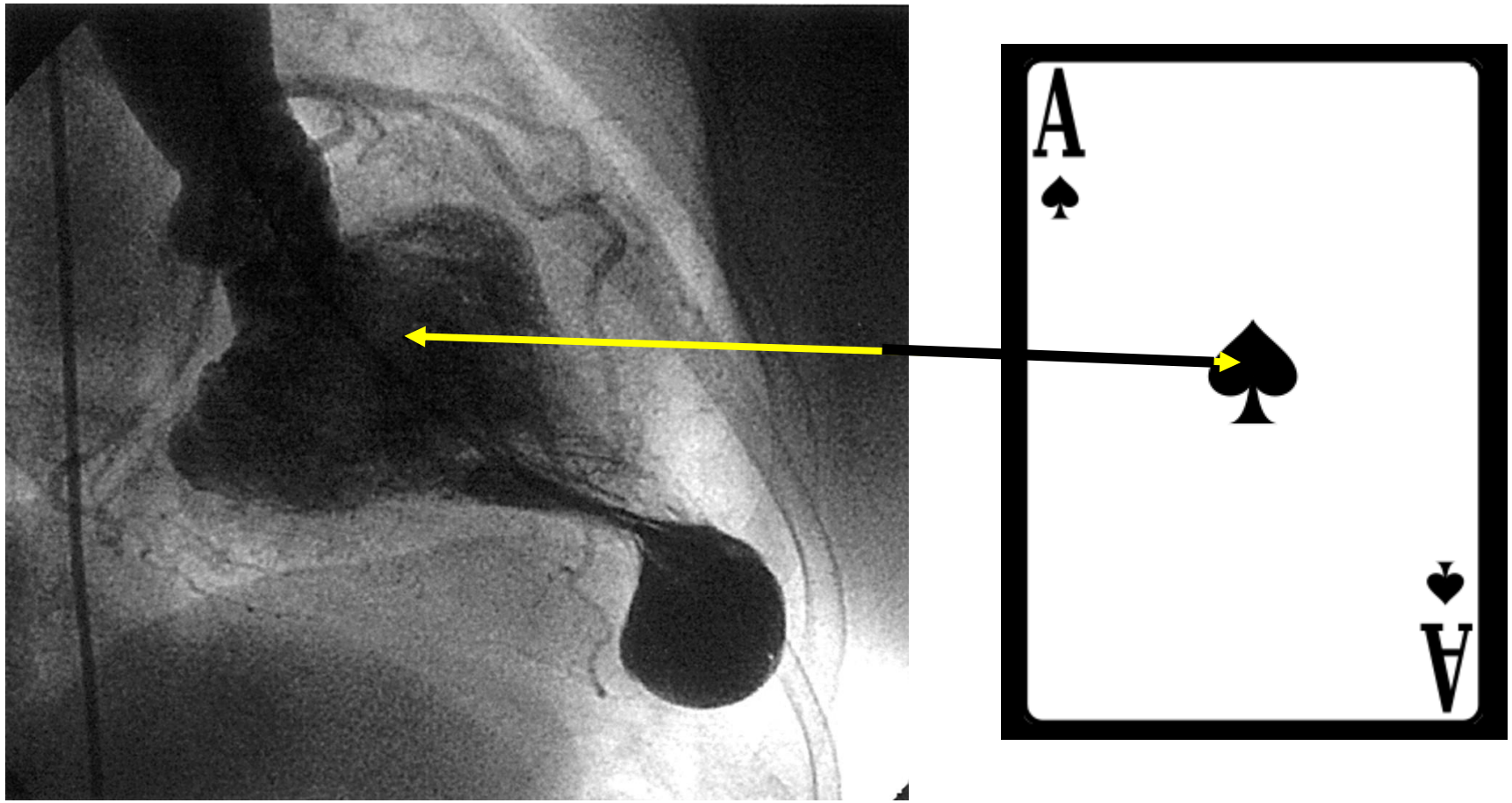


Four-chamber CMR SSFP image demonstrating asymmetrical left ventricular apical thickening. (LV = left ventricle; RV = right ventricle; LA = left atrium; RA = right atrium; ApHCM = apical hypertrophy).

The "ace-of-spades" sign in left ventriculography and CMR is pathognomonic of ApHCM.

Left Ventriculography characteristic of Ap-HCM

The "ace-of-spades" sign on left ventriculography is pathognomonic (**Olearczyk 2008**).



Differential diagnosis

There are many disorders that may involve the left ventricular apex; however, they are sometimes difficult to differentiate. the spectrum of diseases that most frequently affect the apex of the left ventricle including (**Cisneros 2011**):

1. **Athlete's heart:** Apical HCM and athlete's heart share similar electrocardiographic features of increased voltage and deep T-wave inversions.

2. **Wellens' syndrome:** it is characterized by T-wave changes in ECG during pain-free period in a patient with intermittent angina chest pain. It carries significant diagnostic and prognostic value because this syndrome represents a pre-infarction stage of coronary artery disease involving proximal LAD artery, which can subsequently lead to extensive anterior myocardial infarctions (MIs) and even death without coronary angioplasty.

3. **Anderson-Fabry disease (AFD).** Concentric thickening and inferolateral mid-myocardial scar are the most common manifestations of AFD, but the spectrum includes cases morphologically identical to apical and ASH subtypes of HCM and these have more apical and mid-ventricular LV scar. Significant LVH is associated with ventricular arrhythmia (**Deva 2016**).

4. **Chronic Chagasic heart disease with apical aneurism** (**Takeno 1999**). Borges-Pereira et al studied the clinical and epidemiological characteristics of the aneurysm found in the LV in chronic Chagas' disease patients. Three hundred, eighty eight people (298 chagasic patients and 90 randomly selected healthy individuals) were submitted to echocardiography. Aneurysm of the LV was diagnosed in 58 (18.8%) patients, all from the chagasic population. From these, 38 (12.7%) were found in the apical segment; 10 (3.4%) in the interventricular septum; and 2 (0.7%) each in the posterior wall; the inferior wall; apicoseptal; and inferior-posterior. The authors could not observe any significant difference for the aneurysm frequencies in relation to age group, gender and race, and no association between aneurysm and arterial hypertension could be made. Of the 56 individuals presenting aneurysm, 55 (98.2%) were symptomatic with predominant palpitations; 53 (94.6%) showed an aberrant ECG with predominant PVCs followed by changes in conduction; and 34 (60%) showed an impairment of the ventricular function, regardless of the affected segment. In view of these results they consider the apical aneurysm of the LV as a marker of Chagas' disease and as an indicator of high morbidity of the human T. cruzi infection (**Borges-Pereira 1998**).

5. **Takotsubo cardiomyopathy "octopus trap", transient apical ballooning syndrome, apical ballooning cardiomyopathy, stress-induced**

cardiomyopathy, Gebrochenes-Herz-Syndrome, broken heart, and simply stress cardiomyopathy a bulging out of the LV apex with a hypercontractile base of the LV is often noted. It is the hallmark bulging out of the apex of the heart with preserved function of the base.

6. **Left ventricular aneurysms(LVA) or true aneurysm: LVA** are relatively common findings in patients with coronary artery disease(CAD) and may complicate as many as 10% of acute transmural myocardial infarctions (AMI). LVA occurs in 5–10% of patients with AMI, contains the endocardium, epicardium and a predominantly fibrous tissue that has replaced myocardium in its wall. Cross-sectional echocardiography is a useful method for detecting ventricular aneurysms noninvasively (**Weyman 1976**). An important difference is the lower rupture potential of a true aneurysm compared with a pseudoaneurysm. True aneurysms will show late enhancement, which indicates that their wall is formed by scar tissue secondary to infarcted myocardial muscle. Once formed, the aneurysm may contribute to the development of heart failure and underlie such serious secondary complications as recurrent ventricular arrhythmias and systemic embolization, thus significantly worsening the patient's prognosis for life. Since surgical extirpation of LVA has been shown to be effective in reducing symptoms and improving hemodynamics in selected patients, accurate diagnosis is extremely important. Although aneurysms may be detected at the bedside and with noninvasive techniques such as chest roentgenography and ECG, it has been reported that ≈45% of discrete ventricular aneurysms are not detected prior to angiography or necropsy. Even when suspected clinically, ventriculography is required to precisely define the location and extent of ventricular aneurysms.

7. **Pseudoaneurysms:** or false aneurysms are defined as a rupture of the myocardium that is contained by pericardial adhesions. A pseudoaneurysm usually represents a rare complication of myocardial infarction. The wall of a false aneurysm is composed of organized hematoma and pericardium. Pathological examination shows fibrous tissue and lacks the myocardial elements that are usually seen in the wall of true aneurysms. A typical feature of pseudoaneurysm is that it has a narrow ostium connecting them to the ventricle. In most cases, the maximal width of the ostium is less than the maximal parallel internal diameter.

8. **Congenital Apical diverticulum/congenital ventricular diverticula (CVD):** Although CVD is often associated with other cardiac and extracardiac congenital anomalies, it can also be incidentally observed in otherwise healthy subjects. CVD may lead to significant morbidity and even have lethal consequences such as recurrent VT and stroke, and illustrates the importance of multimodal imaging approach in differential diagnosis (**Dostálová 2017**). Congenital ventricular diverticula are characterized by synchronal contractility and three myocardial layers on histologic examination. Two categories of congenital ventricular diverticulum are described with regard to their

Differences among aneurysms, pseudoaneurysms and congenital diverticula

	Aneurysms	Pseudoaneurysms	Congenital diverticula
Layers	All layers of the ventricular myocardium	Organized hematoma and pericardium	All layers of the ventricular myocardium
Cine	Akinetic or dyskinetic segment	Akinetic or dyskinetic segment	Shows contraction during systole
Myocardial late enhancement	Yes	No, only the border of the pseudoaneurysm will show enhancement	No
Pericardial late enhancement	No or faint	Marked	No

localization: apical and nonapical. Apical diverticula are always associated with midline thoracoabdominal defects and other heart malformations. Nonapical diverticula are always isolated defects (**Marijon 2006**)

9. **Left ventricular apical hypoplasia (LVAH):** Isolated left ventricular (LV) apical hypoplasia is a rare anomaly characterized by a dysfunction, spherical LV and elongated RV wrapping around the deficient LV apex. The etiology is unknown; it is presumed to be a rare congenital developmental defect during partitioning of the ventricles, caused by an in-utero infection. The entity is considered newly characterized cardiomyopathy. LVAH is characterized by a truncated, spherical LV, without a normal apex; the true apex is occupied by the right ventricle. In addition, the atrioventricular valve anomalies, LV papillary muscle displacement, interventricular and/or interatrial septal bulge aneurysms and patent ductus arteriosus can coexisted in these patients. LV apical hypoplasia has distinct appearances that can be easily identified on transthoracic echocardiography (TTE) and CMRI. TTE also could accurately define the associated cardiac abnormalities (**Meng 2013**). Alizadeh Sani et al described the first case of LV apical hypoplasia combined with LV diverticulum and a family history of SCD (**Alizadeh Sani 2016**). Pica et al detected a mutation of the lamin A/C gene associated with left ventricular apical hypoplasia suggesting that it could be a new phenotype for laminopathies (**Pica 2014**).

10. **Apical ventricular remodeling:** Cardiac remodelling refers to the change in size, shape and function of the heart after injury to the ventricles. The injury is typically due to large acute MI depending on LAD artery occlusion. Non-ischemic causes include chronic volume overload and genetically determined cardiomyopathy) (**Sutton 2000**). MI promotes transformation of both infarcted and non-infarcted regions, which leads to global LV dilation ("ventricular remodelling"). It is a dynamic process, starting in the acute phase with myocardial thinning and lengthening of the infarcted area, progressing to LV dilatation and hypertrophy of the non-infarcted area. Dysfunction with increased sphericity is common, as is the deposition of myocardial fat and the development of apical thrombus.

10. **Arrhythmogenic right ventricular cardiomyopathy/dysplasia (ARVC/D)** with left ventricular involvement. The diagnosis of ARVC/D is based on established criteria determined by a task force comprising the European Society of Cardiology and the International Society and Federation of Cardiology (**Marcus 2010**) DOI:10.1093/eurheartj/ehq025 | PUBMED ID:20172912 Full text avaliable.

11. **Left ventricular false tendons:** are fibrous or fibromuscular bands that stretch across the LV from the septum to the free wall. They can also tether to a papillary muscle, but unlike the chordae tendineae, do not connect to the mitral leaflets. They are anatomic variants that should not be mistaken for abnormalities such as tumors, subaortic membranes, thrombus borders, septal hypertrophy.

12. **Left ventricular non-compaction (LVNC):** may be an isolated finding (isolated LVNC) or associated with other congenital heart disease. It is a congenital cardiomyopathy characterised by a thickened wall with multiple prominent ventricular trabeculations and deep intertrabecular recesses (sinusoids) in communication with the ventricular cavity, resulting in systolic and diastolic ventricular dysfunction. LVNC involves predominantly the apical portion of the LV chamber, with or without RV involvement, due to an arrest in the normal embryogenesis. Because both the LV and RV normally have trabeculations, there is always the risk of over-diagnosis of this disease. It is therefore essential to be thorough while establishing the relationship between compacted and non-compacted myocardium when the diagnosis of this entity is being made. Using MRI, the most appropriate criteria would be a ratio between non-compacted and compacted areas greater than 2.3 in a diastolic long-axis view. (**Goldfarb 2009**).
13. **Apico aortic valved conduit:** It is an alternative approach severe aortic stenosis with apico-aortic conduit or for isolated idiopathic hypertrophic subaortic stenosis. It has three components: an outflow graft to the descending aorta, a valved conduit and an elbow apical connector. An apico-aortic conduit decreases the LV aortic pressure gradient, preserves or improves ventricular function, and maintains normally distributed blood flow through the systemic and coronary circulation.

14. Ap-HCM: The diagnosis is based on:

- Demonstration of localized apical hypertrophy, defined as an end-diastolic LV apical wall thickness greater than 15 mm or a ratio comparing apical LV and basal LV wall thicknesses of ≥1.3–1.5
- Obliteration of the LV apical cavity in systole
- Failure to identify a normal progressive reduction in LV wall thickness towards the apex
- Apical aneurysm formation with delayed enhancement.

The formation of apical aneurysm is thought to be due to ischemia, which results from reduced capillary density, hyperplasia of the arterial media, increased perivascular fibrosis and myocardial bridging. This process usually occurs in the presence of normal epicardial coronary arteries

Complications

Although generally associated with a better prognosis than other forms of HCM, serious cardiac complications have been described, including":

1. Progressive heart failure
2. Arrhythmias
3. Myocardial infarction/ unstable angina (**Chan 2008**)
4. Sudden-cardiac death (**Maron 2008**; **eriksson2002**).
5. LV apical aneurysms are at high risk for arrhythmic sudden death and thromboembolic events. Identification of this phenotype expands risk stratification and can lead to effective treatment interventions for potentially life-threatening complications (**Rowin2017; Li 2017**).
6. Thromboembolic events.

Prognosis

ApHCM is often characterized as benign. However, more recent publications indicate that there is significant heterogeneity among individuals with ApHCM and that for many the condition is less than benign.

ApHCM in North American patients is not associated with SCD and has a benign prognosis in terms of cardiovascular mortality. Nevertheless, ≈ 30% experience serious cardiovascular complications, such as myocardial infarction and arrhythmias. These data are likely to influence the counseling and management of patients with ApHCM (**Eriksson 2002**).

Prognosis is relatively benign in terms of cardiovascular mortality; however, morbid sequelae, such as diastolic dysfunction, left atrial enlargement, apical thrombi, ventricular aneurysms, and, are not uncommon. acute coronary syndrome/myocardial infarction (**Olearczyk 2008**).

The presence of rest angina with the ECG findings of giant inverted T-wave in precordial leads lead to a mistaken, initial diagnosis of acute subendocardial myocardial. In this cases is frequent a systolic pressure gradient exceeding 100 mmHg between the apex and main LV cavity (**Cubukçu 1993**).

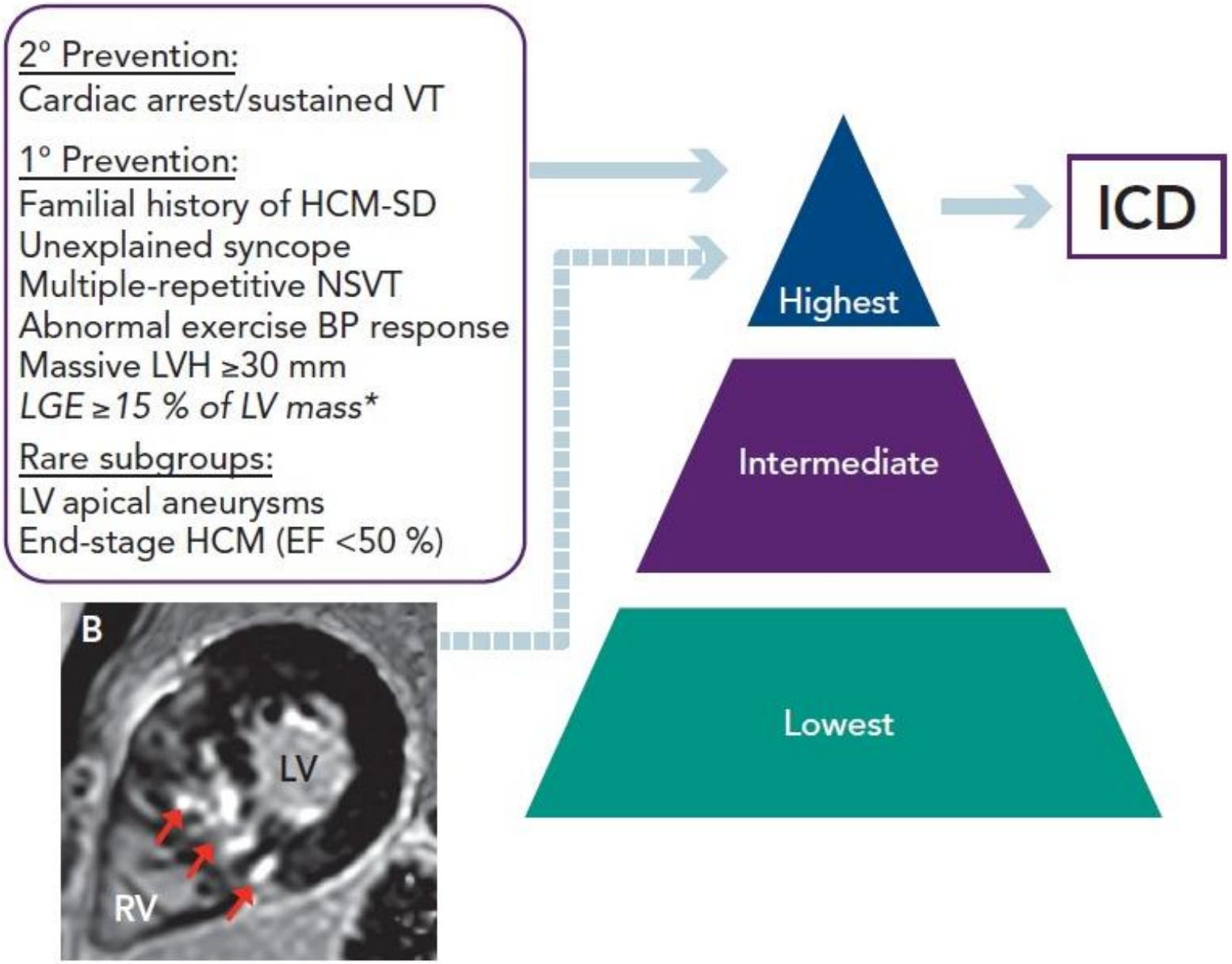
Management

1. **Drugs:** Medications used to treat symptomatic patients with ApHCM include verapamil, beta-blockers and antiarrhythmic agents such as amiodarone and procainamide.
2. **Implantable cardioverter defibrillator:** An ICD is recommended for high risk patients (see next slide).
3. **Ablation:** Monomorphic VT in a ApHCM can be due to endocardial, epicardial or intramural reentry in areas of apical scar. Epicardial ablation or transcoronary alcohol ablation is required in some cases (**Inada 2011**).
4. **Apical myectomy:** it improves functional status by decreasing left ventricular end-diastolic pressure, improving operative compliance, and increasing stroke volume. This procedure might be of value in other patients with HCM who have severe hypertrophy and small LV end-diastolic volume (**Schaff 2010**).
5. **Heart transplantation:** ApHCM is a morphologic variant in which the hypertrophy is primarily localized at the apex of the LV. A subset of patients have progressive, drug-refractory diastolic heart failure with severely limiting symptoms caused by low cardiac output. Heart transplantation has been the only therapeutic option available for such patients.

No definitive guidelines delineate the role for defibrillator implantation in AHCM patients with family histories of sudden cardiac death. Not surprisingly, expert consensus supports the use of implantable defibrillator for primary prevention in select high risk patients, notably those with one of the five following risk factors: a family history of sudden cardiac death, syncope, asymptomatic non-sustained ventricular tachycardia, an abnormal blood pressure response to exercise and a left ventricular wall thickness > 30 mm (**Epstein 2013**). Genetic testing has also been offered to both of our patients and their first-degree family members; although contrary to other subtypes of HCM, the apical variant has only occasionally been described as a familial disease manifesting an autosomal dominant inheritance. A very limited number of sarcomere gene defects, and in particular, cardiac actin Glu101Lys, have been shown to consistently produce the AHCM phenotype (**Arad 2005**). The widespread availability of genetic testing has led to increased recognition of genotype-positive/phenotype-negative patients. Although at present, it is not possible to predict clinical outcomes based on the presence of individual mutations, guidelines do suggest extension of surveillance with cardiac imaging at least through mid-life (40 years of age, and beyond, in select circumstances) to detect development of overt disease (**Gersh 2011**).

Anesthetic management of these patients are similar to case of hypertrophic obstructive cardiomyopathy which includes alleviating sympathetic stimulation which includes avoiding tachycardia, maintaining normal sinus rhythm and euvolemia, and avoiding any increase in cardiac contractility. Preoperatively, it is important to rule out coronary artery disease as most of these patients have ST segment depression with T-wave inversion in ECG leads.β- blocker such as metoprolol 50 mg OD,, which decreases myocardium workload, and decreases chances of perioperative MI. Sinus rhythm is important in these patients as their preload depends on the atrial contraction. Intraoperatively, fluid are the choose for the

control of values of the stroke volume variation. Vasodilators and agents which increase the cardiac contractility should be avoided. Any episodes of hypotension should be treated preferably with volume replacement than with vasoconstrictors (**Ranasinghe 2011**).



Pyramid Profile of Risk Stratification Model Currently Used to Identify Patients at the Highest Risk of Sudden Death Who May be Candidates for ICD for Sudden Death Prevention

Major risk markers appear in boxes at the upper left. *Extensive LGE is a potential novel primary risk marker that can also be used as an arbitrator when conventional risk assessment is ambiguous. B. Example of a patient with extensive LGE throughout the septum (arrows) occupying 17 % of LV mass, and without other traditional risk markers. Based on extensive LGE, the patient had ICD placed for primary prevention of SCD with appropriate ICD discharge for VF 1 year later. EF = ejection fraction; HCM = hypertrophic cardiomyopathy; ICD = implantable cardioverter defibrillator; LGE = late gadolinium enhancement; LV = left ventricular; LVH = left ventricular hypertrophy; NSVT = non-sustained ventricular tachycardia; RV = right ventricle; SD = sudden death; VT = ventricular tachycardia.

Risk factors associated to sudden death in HCM and conditioning factors for a worse prognosis

1. Sustained VT/cardiac arrest(history of recovery from SCD (ICD as secondary prevention)
2. Recurrent syncope in youngster
3. Familial history of HCM-SD/Hereditary genetic defect, associated to unfavorable prognosis (ICD as secondary prevention)
4. Multiple-repetitive episodes of nonsustained ventricular tachycardia on 24-hours Holter monitoring
5. Extreme increase of septal thickness: extreme left ventricular (LV) hypertrophy (> 30 mm) in young patients
6. Very increased estimation of myocardial mass(massive LVH≥30mm)
7. Left ventricular apical aneurism
8. Progression of the disease to LV wall thinning and decrease of EF: End-stage HCM with LVEF < 50%
9. Unexplained (not neurally mediated) syncope, particularly in young patients
10. Significant bradyarrhythmia or concealed conduction
11. Blood pressure decrease or inadequate increase during upright exercise.
12. LGE ≥ 15% of left ventricular mass
13. 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.
14. End-stage HCM (LVEF< 50%
15. LV apical aneurism

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