REVIEW ARTICLE

Value of Electro-Vectorcardiogram in Hypertrophic Cardiomyopathy

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The electrocardiogram is an important tool for the initial diagnostic suspicion of hypertrophic cardiomyopathy in any of its forms, both in symptomatic and in asymptomatic patients because it is altered in more than 90 percent of the cases. Electrocardiographic anomalies are more common in patients carriers of manifest hypertrophic cardiomyopathy and the electrocardiogram alterations are earlier and more sensitive than the increase in left ventricular wall thickness detected by the echocardiogram. Nevertheless, despite being the leading cause of sudden death among young competitive athletes there is no consensus over the need to include the method in the pre-participation screening. In apical hypertrophic cardiomyopathy the electrocardiographic hallmarks are the giant negative T waves in anterior precordial leads. In the vectorcardiogram, the QRS loop is located predominantly in the left anterior quadrant and T loop in the opposite right posterior quadrant, which justifies the deeply negative T waves recorded. The method allows estimating the left ventricular mass because it relates to the maximal spatial vector voltage of the left ventricle in the QRS loop. The recording on electrocardiogram or Holter monitoring of nonsustained monomorphic ventricular tachycardia in patients with syncope, recurrent syncope in young patient, hypotension induced by strain, bradyarrhythmia, or concealed conduction are markers of poor prognosis. The presence of rare sustained ventricular tachycardia is observed in mid-septal obstructive HCM with apical aneurysm. The presence of complete right bundle branch block pattern is frequent after the percutaneous treatment and complete left bundle branch block is the rule after myectomy.

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electrocardiogram; vectorcardiogram; hypertrophic cardiomyopathy

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a heterogeneous, complex, hereditary and familial (60%) or sporadic (40%), autosomal dominant polygenic disease in most cases, with a high degree of penetrance, caused by mutations in the genes that encode the sarcomere proteins and characterized by "bizarre" myocardial hypertrophy by sarcolemmal disorganization and in absence of any identifiable stimulus that justifies it, such as hypertension, valve disease, or congenital heart disease. The degree and location of this hypertrophy is variable, being more frequent in the basal interventricular septum (IVS) (HCM-OF). In terms of degree it may be mild (13–15 mm),

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moderate (16 to 29 mm), or severe (≥30 mm). Septal thicknesses between 13 and 16 mm (gray area) are exceptionally observed in high-performance athletes of the male gender, mainly of black race. They have a higher percentage of ECG alterations, including voltage criteria for left ventricular hypertrophy (LVH), ST segment elevation and inverted or flat T waves. Left ventricular (LV) remodeling in black athletes is characterized by a greater thickening than in Caucasian ones, and unlike HCM, where the LV chamber is almost always small (<45 mm) with diastolic dysfunction and increase in LV end diastolic pressure (LV Pd_2), it does not present diastolic dysfunction and the size of the LV chamber is >55 mm (eccentric hypertrophy).[1]

Table 1 relates the main criteria to differentiate athlete's heart from HCM (Table 1).

Approximately 5%–10% of the patients in late phases evolve into chamber dilatation and systolic dysfunction. This situation is called dilated HCM (D-HCM), resulting in myocardial fibrosis secondary to myocardial ischemia and similar to true idiopathic dilated cardiomyopathy (DCM). Goto et al.[2] compared the characteristics, treatments and the results of patients in congestive heart failure by D-HCM versus DCM. The carriers of D-HCM were predominantly from the male gender, presented previous stroke more frequently, atrial fibrillation (AF) and sustained monomorphic ventricular tachycardia (S-MVT) or ventricular fibrillation (VF) when compared with the patients carriers of DCM. The echocardiogram showed that the patients with D-HCM have less LV end systolic diameter, greater ejection fraction, and greater LV wall thickness.

The treatment for both groups is similar; however, D-HCM presents a greater need of using amiodarone, anticoagulation and indication of implantable cardioverter defibillator with significantly greater mortality.

MAIN EPIDEMIOLOGICAL DATA

Prevalence

HCM is one of the most common hereditary diseases (it affects ≈ 1 per 500 people).[3]

The approximate overall prevalence is 0.05–0.2% of the population and is the number one cause of sudden cardiac death (SCD) in young competitive athletes (<35 years).[4]

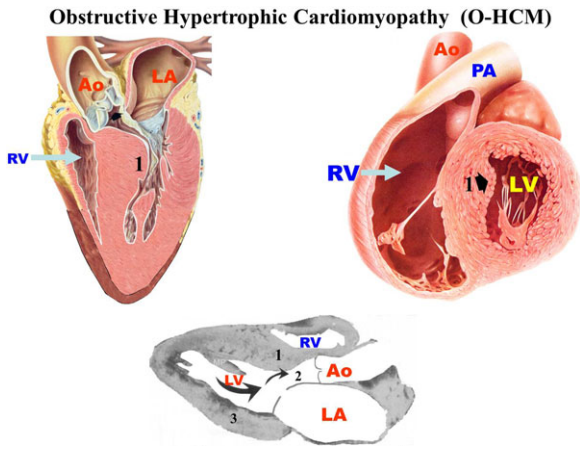

Figure 1. Hypertrophic Cardiomyopathy in its Obstructive Form (HCM-OF). 1. Interventricular septum where a greater thickness is observed in the superior part (basal). 2. LVOT: Left ventricular outflow tract. 3. LV free wall that shows progressive decrease in the thickness of the base of the apex.

HCM is asymptomatic or with unspecific symptoms in many patients.[5]

Incidence

In young people (<35 years of age), the incidence of sudden deaths (including noncardiac deaths) is 1.5–6.5 per 100,000 people per year, and that of SCD is 0.3–3.6 per 100,000 people per year.[6]

Morphological Types

(1) Hypertrophic cardiomyopathy. Obstructive forms (HCM-OF)

(a) **Asymmetric septal with basal obstruction** and gradient in the LV outflow tract. Septum with greater thickness in the superior part (basal). LV free wall with progressive decrease of the apex base thickness (as normally).

(b) **Mid-ventricular obstructive** (MVO) with formation of apical aneurysm (9.4%).[7] The diagnosis is made if the peak of the mid-ventricular gradient is ≥30 mmHg. The LV has an hourglass appearance.[8] This rare variant predisposes the appearance of S-MVT.

Figure 1 represents asymmetric septal HCM-OF with obstruction and basal gradient.

Table 1. Clinical, Electrocardiographic and Echocardiographic and Lab Elements to Differentiate Athlete's Heart from HCM (59–63)

	Athlete's Heart	HCM
Family history of HCM	Absent	Present
Unusual pattern of bizarre LVH	No	Frequent
ECG criterion of LAE	Absent	Frequent
Negative T wave V_5-V_6	No	Frequent
LV chamber	>55 mm	<45 mm
Septal thickness*	<12 mm for men and <11 mm for women	>15 mm
Regression of hypertrophy with deconditioning	Yes	No
Morphology of hypertrophy	Biventricular eccentric	Asymmetric is frequent
Diastolic function	Normal	Altered
Abnormal ventricular NT-proBNP levels	No	Possible

*The so-called grey area is considered a ventricular wall thickness between 13 and 16 mm.

(1) **Hypertrophic cardiomyopathy. Non-obstructive forms (HCM-NOF).** The LV free wall does not present the normal progressive decrease in the apex base thickness.

Variants:

I. *Asymmetric septal without obstruction:* the most frequent one (60% to 70% of the cases). Septal thickness ≥15 mm or septum/free wall ratio >1.3[9]
II. *Apical (apHCM):* septum with greater thickness at the apex (apical). It represents 3% of all the cases of HCM in USA and 10% in Japan: "Japanese HCM."[10]
III. *Lateral and/or posterolateral in the LV free wall*
IV. *Concentric, symmetric or heterogeneous hypertrophic:* (≈20% of the cases).
V. *From the right ventricle (RV):* it is diagnosed when two or more RV segments are hypertrophic and when at least two measurements of the RV wall are greater than two standard deviations of the average recorded in normal individuals. With the use of these criteria, McKenna et al.[11] observed right ventricular hypertrophy in 44% of 73 patients with HCM. By using cardiac magnetic resonance (CMR), the RV mass is seen increased in most of the cases.

Figure 2 shows an outline of the HCM-NOF variants.

The Electrocardiogram in Hypertrophic Cardiomyopathy

The ECG is altered in 93% of the cases, both in symptomatic and asymptomatic patients. Due to the high prevalence of the electrocardiographic alterations, any patient with an ECG with LVH criteria in absence of an apparent cause should be suspected of HCM, even being asymptomatic and normal in the physical examination, as it may occur in the nonobstructive forms.[12] ECG anomalies are more common in patients carriers of manifest HCM and the ECG alterations are earlier and more sensitive than the increase in LV wall thickness detected by the echocardiogram.[13]

Significance of the Electrocardiogram in the Preparticipation Screening of the Candidates to Competitive Sports Practice in Young Athletes (<35 years)

In the region of Padua, Veneto, Italy, where the preparticipation screening of the candidates to practice competitive sports always includes the ECG besides the questioning and the physical examination, from a universe of 33,735 young athlete candidates to practice competitive sports (<35 years), it was verified that in those qualified, HCM was a rare cause of SCD (only 2%). This fact points out that the ECG previously identified—at least in part—and disqualified a large percentage of the young athletes candidates to the practice of competitive sports, carriers of HCM. In 18 years of follow-up of the athletes considered capable of practicing competitive sports, 269 SCD occurred, with a great predominance in the male gender, being the most frequent cause for arrhythmogenic RV dysplasia (22.4%), followed by coronary atherosclerosis (18.4%) and the anomalous origin of the coronary arteries (12.2%).

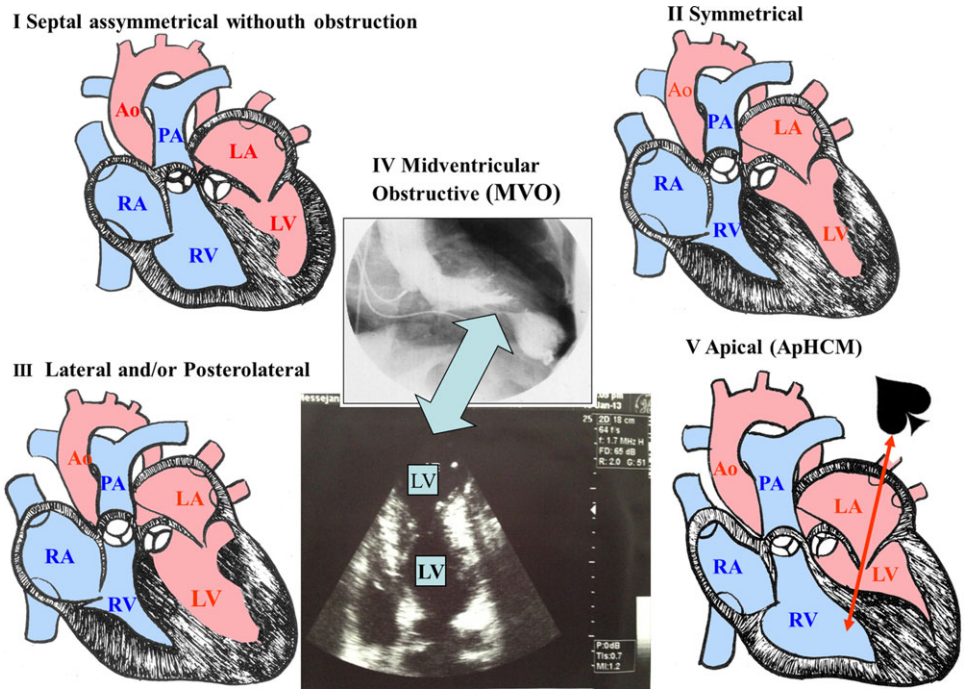

Figure 2. An outline of the main nonobstructive forms of HCM is shown: (I) asymmetric septal without obstruction; (II) lateral and/or posterolateral; (III) symmetric; (IV) mid-ventricular obstruction; (V) apical (apHCM).

SCD, as a consequence of HCM, occurred in just 2%.[14] Similar results were confirmed later in a second manuscript.[15] It was verified that the incidence of SCD in young competitive athletes from the region of Veneto, Italy, substantially decreased since the introduction of the ECG in the systematic preparticipation screening and that this lower mortality should be attributed to a decrease in SCD by HCM. Although the European Society of Cardiology (ESC)[16] and the International Olympic Committee (IOC) http://multimedia.olympic.org/pdf/en_report_886.pdf recommend including the ECG in the preparticipation evaluation, the last guidelines (2007) from the American Heart Association and the Council on Nutrition, Physical Activity and Metabolism, backed by the American College of Cardiology, in relation to the preparticipation evaluation of competitive athletes, have taken a stance contrary to this guideline.[17] According to these authorities, the previous evaluation of competitive athletes using the ECG is not justified, unless the history and/or the physical examination point out the need for a wider cardiovascular evaluation. This position has been maintained in spite of the different sources of scientific data that support a drastic decrease in mortality attributed to the inclusion of the ECG as a routine in screenings.[15] Including the ECG is a strategy that saves lives and prevents SCD by identifying the presence of HCM and other congenital and genetic heart diseases of candidates to young athletes, besides having a good cost-benefit ratio.[18] We share the interpretation of Myerburg et al.[19] that the American Heart Association and the Council on Nutrition, Physical Activity and Metabolism should reconsider their 2007 guideline on the routine non inclusion of ECG,[17] taking into account the existing scientific evidence to this moment.[14;15]

Electrocardiographic Criteria for the Identification of Hypertrophic Cardiomyopathy

The electrocardiographic criteria of the patients carriers of HCM were grouped into major and minor.

(1) Major criteria

- Criteria based on an increase in voltage or QRS complex width

I. For Left Ventricular Hypertrophy (LVH):

(1) *Sokolow and Lyon index*[20]: S of V_1 + R of $V_5 \geq 35$ mm or 3.5 mV in adults older than 30 years old, ≥40 mm between 20 and 30 years and >60 mm between 16 and 20 years old and > than 65 mm between 11 and 16 years old.
(2) *Cornell index (CI)*[21]: CI = R of aVL + S of V_3 > than 28 mm in men or >20 mm in women indicate LVH.

II. For Right Ventricular Hypertrophy (RVH): RV hypertrophy (RVH) is only detectable in the ECG if the RV wall, normally thin, develops a hypertrophy that reaches a degree of balance that is greater than the LV mass, which in adults requires a long period (ECG in adults is a levocardiogram).

Sokolow-Lyon index for the RV: R wave voltage in V_1 + depth of S wave of V_5 and/or $V_6 \geq 10.5$ mm.

- ST segment and T wave alterations or criteria based on QRS/T angle widening:

Present in approximately 85% of the cases, both in the HCM-OF of symptomatic and asymptomatic patients, as in the HCM-NOF of symptomatic patients. In HCM-NOF of asymptomatic patients is minor; however, they are observed in 58% of cases.[12] This repolarization pattern has been called pattern of LVH with systolic-type repolarization[22–24] or "left ventricular strain pattern."

The $^{QRS}/_{ST\text{-}T}$ angle >100° with ST elevation of superior concavity of more than 0.1 mV, followed by positive T wave in V_2 and concomitant depression of the ST segment of superior convexity followed by negative T wave with asymmetrical limbs (descending limb slower than the ascending limb) are observed in the left precordial leads that explore the LV.

Note: In the Romhilt-Estes score system for LVH (1968)[25] the presence of an ST-T vector opposite QRS in absence of use of digitalis is given a value of 3 points, and with digitalis 1 point. The score system of Romhilt-Estes is in Table 2.

Figure 3 shows a typical example of HCM with enlargement of the left chambers (left atrial enlargement (LAE) +LVH).

Table 2. Romhilt-Estes Score System (25) for the Diagnosis of Left Ventricular Hypertrophy (LVH) (1968)

Findings in ECG	Score
Voltage criterion: any R or S in limb leads ≥20 mm; S in V_1-V_2 or R in V_5-V_6 ≥30 mm	3 points
ST-T vector opposite to QRS without digitalis	3
ST-T vector opposite to QRS with digitalis	1
Final negative component of slow and deep P in V_1 according to Morris criterion. (30)	3
Electrical axis of QRS located at the left of –30° in the FP	2
Duration of QRS >90 ms	1
VAT, R peak time, or intrinsicoid deflection in V_5-$V_6 \geq 50$ ms	1

The authors considered the presence of LVH as indisputable when the addition is ≥5 points and probable LVH with ≥4 points.

Abnormal Q Waves

They should be considered pathological when observed in at least two contiguous leads, with a width greater than 1/3 of the height of the next R wave and with duration >3 ms[26] or when these Q waves have a duration ≥40 ms, or when their depth is greater than 25% of the voltage of the next R.[3]

Table 3 shows the differential features of Q waves of LVH by HCM of Q waves of infarction.

The pathological Q wave criteria have been the object of much debate. The last accepted definition by the ESC and the ACC about it is the following:[27]

- Any Q wave in the V_2-V_3 leads ≥0.02 sec or QS complexes in leads V_2 and V_3.
- Q wave of duration ≥0.03 sec and >0.1 mV of depth or QS complexes in leads I, II, aVL, aVF, and from V_4 to V_6 in at least two contiguous leads (I, aVL, and V_6, V_4-V_6, II, III and aVF).
- R wave ≥0.04 sec in V_1-V_2 and $^{R}/_{S}$ ratio ≥1 with matching T wave in absence of conduction defect.

Note: The absence of pathological Q waves does not rule out a myocardial infarction. Lead III often displays Q waves, in their absence in contiguous leads II and aVF in the inferior wall.

The Minnesota code for the classification system of electrocardiographic findings (Novacode) contains a detailed description of Q waves that should

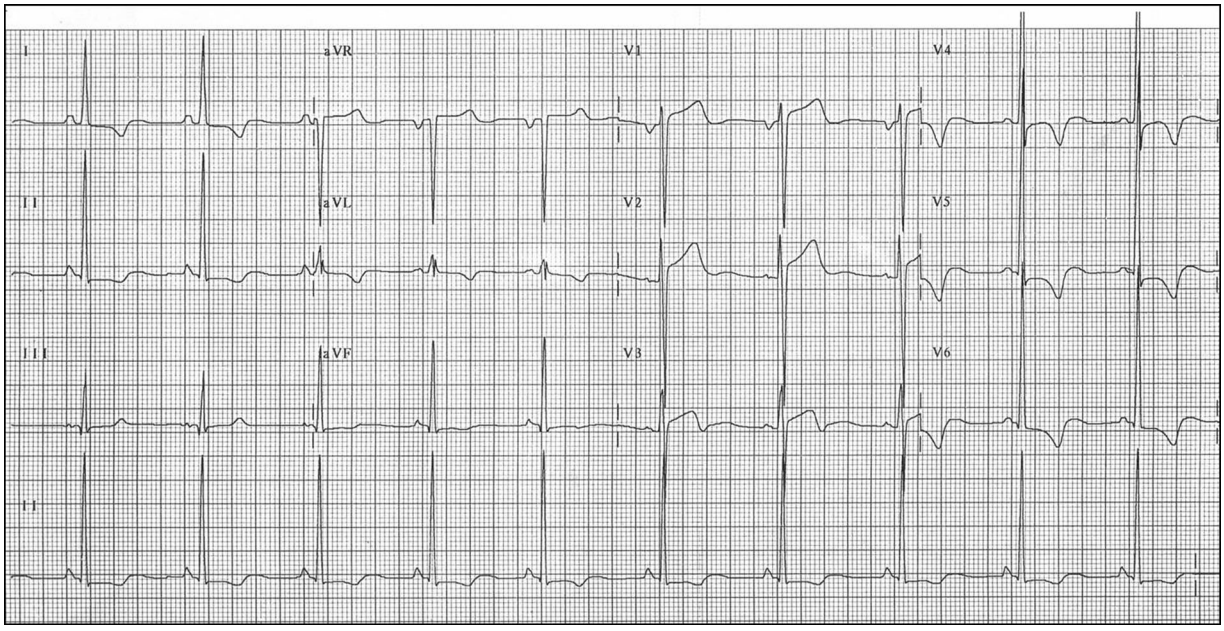

Figure 3. Clinical diagnosis: This ECG belongs to a 38-year-old man carrier of severe HCM-OF, nonresponsive to drugs. Septal thickness of 30 mm; gradient in rest of 80 mmHg. Functional class IV in spite of using drugs in full doses (atenolol 150 mg/day).

be considered pathological in patients without history of myocardial infarction.[28]

• Ventricular activation time

It is the time elapsed because the onset of QRS until the apex of R wave in V_5-V_6. It is also called "R peak time" or intrinsicoid deflection. In HCM, the presence of initial q wave in V_5-V_6 may increase this time for values ≥50 ms as it happens in eccentric, diastolic, or volumetric LVH.[29]

Figure 4 shows the outline that represents the prolongation of ventricular activation time in eccentric LVH or with initial q wave in the left leads.

(2) Minor criteria

• Left atrial enlargement (LAE) pattern:

(1) *Duration of P wave:* ≥110 ms in adults, ≥120 ms in elderly people, and ≥90 ms in children of 1 year to 12 years old and up to 80 ms from 0 to 1 year (Table 4).
(2) *Aspect of P wave:* bimodal in II with distance between modules ≥40 ms, with the second module of greater voltage than the first (Fig. 5).
(3) *P wave characteristics in V_1 or V_1-V_2:* of positive-negative or plus-minus polarity with depth and duration of the final negative component (PTFV1) increased: >0.04 sec of width and >1 mm of depth: 0.04 mm/sec (Morris index) (Fig. 6).[30]

• S wave of V_2 with depth >25 mm.[3]
• Decrease in voltage of R wave in left precordial leads V_5-V_6.
• Minimal alterations of ventricular repolarization in the leads that explore the LV.
• Presence of intraventricular conduction disorder.

Analysis of the Main Electrocardiographic Parameters in HCM

Rhythm

Sinus in most of the cases. There is a tendency to the appearance of acute AF. This occurs ≈ in 10% of the cases and is a consequence of the decreased compliance of the LV by increase in Pd_2 of the LV associated to variable degrees of mitral valve insufficiency. Both lead to an increase in the medium left intraatrial pressure, LAE, and greater tendency to AF.

In a population of 480 patients carriers of HCM, Olivotto et al.,[31] observed in an average

Table 3. Differential Characteristics of Q waves of LVH by HCM of Q waves of Infarction

Differential elements of Q waves of LVH by HCM from Q waves of infarction		
	Q Waves of HCM	**Q Waves of Myocardial Infarction**
Duration	≤35 ms	≥40 ms (except aVR and V_1). In infants and children with anomalous origin of coronary arteries, q waves have a duration >30 ms or depth > than 25% of the next R.
Aspect	"Clean," narrow and deep ("dagger-like")	With notches and accompanied by lesion current with superior convexity and ischemia.
Location	In lateral wall ($V_{5–6}$, I, aVL) and/or inferior wall (II, III, aVF). q waves in lateral wall. Q waves are more common than Q waves in inferior leads	Variable and segmentary
Cause	Abnormal distribution of the myocardial mass	Result from absence of electrical activity; transmission of potentials of cavity of the heart surface with a new electrical balance of forces that get away from the affected region.
Symptoms	There may be chest pain.	Characteristic prolonged pain.
Serum enzymes and troponin	Not increased	In the acute phase, increase of CKMB, AST, LDH, and troponin
Age group	Young people and even children	More common in elderly people, except for anomalous origin of coronary arteries.

follow-up of 9 years, that AF was common in HCM with a 22% prevalence. On the other hand, the presence of AF was associated to a substantial risk for the appearance of heart failure related to mortality, strokes and severe functional disability, particularly in those patients with LVOT obstruction. AF was an independent predictor in an advanced age for the presence of pulmonary congestion and increase in the left atrium size. Those carriers of HCM ≤50 years old, or those that developed chronic AF, had a benign evolution in 35% of the cases.

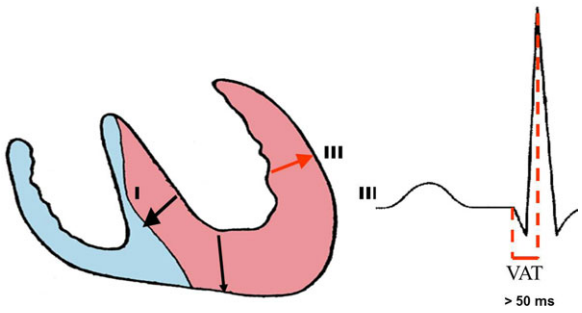

Figure 4. Outline that shows the discrete prolongation of ventricular activation time (VAT), R-peak time or intrinsicoid deflection. In cases of eccentric LVH or with q wave in the left leads, the VAT is ≥50 ms. The VAT is defined as the time elapsed since the onset of QRS to the peak of R. The VAT is lower in concentric LVH without initial q waves in the left leads.

P Wave

In approximately 20% of the cases, a pattern of LAE is observed as a consequence of an increase in Pd_2 of the LV, by decrease of its compliance and different degrees of mitral valve insufficiency.

The combination of right atrial enlargement (RAE) and LVH is quite frequent and it suggests HCM, because this is rare in aortic valve stenosis,

Table 4. Maximal Normal Duration of P wave According to Age

Age	Duration of P Wave
From 0 to 12 months	Up to 80 ms (two small squares)
Children >1 year to 12 years old	≥90
Adults	≥ 110 ms
Elderly people	≥ 120 ms

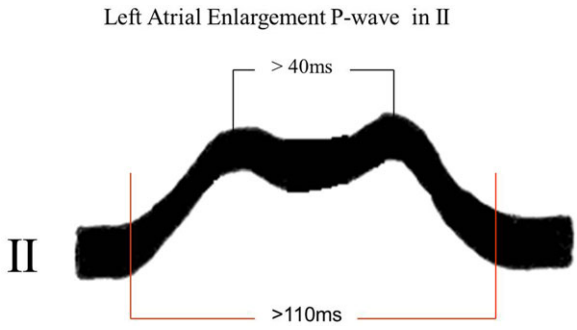

Figure 5. Normal P wave has a rounded and monophasic aspect, although it may be bimodal (more frequent in V_3 and V_4). In this case, the distance between the two modules should not exceed 30 ms (0.03 sec). When the distance between the module vertices is ≥40 ms (0.04 sec), it may correspond to LAE or Bachmann fascicular block by activation of the left atrium.

an entity that frequently poses a differential diagnosis.[32] The RAE pattern may also be observed in the presence of Bernheim's syndrome, which consists of a reduction in the RV size because of LV enlargement that invades the space of the RV. Consequently, the flow of blood from the RA into the RV is reduced, causing RAE.

PR Interval. AV Block

Normal and possibly short. Short PR may correspond or not to a true Wolff-Parkinson-White (WPW) syndrome. In two families, familial WPW and HCM were observed. The genetic study revealed a missense mutation in chromosome 7q35-q36, in the PRKAG2 gene of autosomal dominant transmission.[33] Mutations in the PRKAG2 gene are associated to familial ventricular preexcitation, HCM, sinus bradycardia and familial right bundle branch block. The clinical and experimental data suggest disease by accumulation of glycogen.[34]

AV blocks in different degrees and even complete AV block could be observed in HCM: the latter frequently as a consequence of ablation treatment with absolute alcohol injection. Complete AV block in this situation can be transient or permanent.

QRS Axis in the Frontal Plane (*SAQRS*)

In HCM-NOF it is between 0° and +90° in most cases. SAQRS between 0° and -90° is observed in ≈30% of the cases. In the presence of left anterior fascicular block, the axis displays extreme leftward shift (beyond -30° to -45°). It could be the consequence of percutaneous treatment with alcohol injection in the first perforating artery. In this case, it is always associated to complete right bundle branch block (CRBBB).

Pattern of QRS

Patients carriers of HCM-OF present a greater prevalence of LVH according to voltage criteria (54% vs. 28%), while ventricular arrhythmias are more frequent in those without gradient in the LVOT.[35]

In approximately 10% of the cases, very wide R waves are observed in V_1 and aVR, associated to deep, narrow and "clean" (dagger-like) Q waves

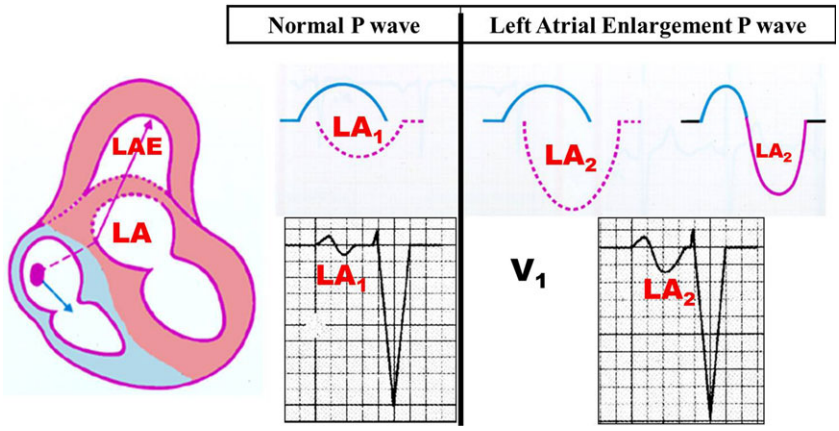

Figure 6. Representation of normal P wave (**AI_1**) and LAE (**AI_2**) in V_1. Check the slow (duration expressed in seconds ≥0.04 mm/sec) and deep final component (depth expressed in mm) (PTFV1) of P wave in **AI_2**.

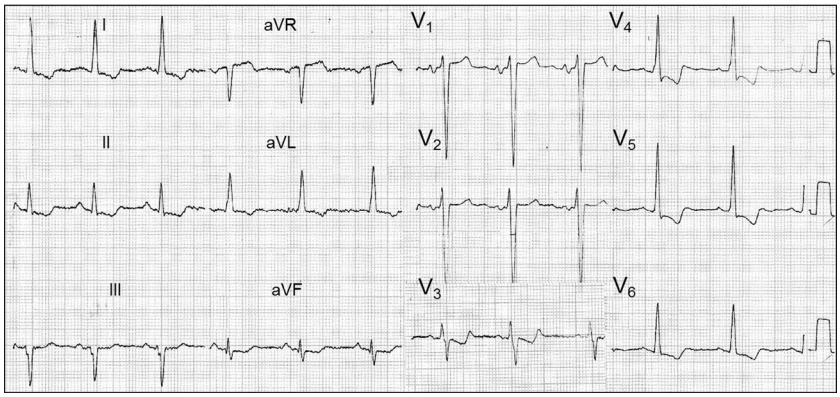

Figure 7. Male, Caucasian, 69-year-old patient, carrier of severe HCM-OF. *Clinical diagnosis:* HCM-OF with gradient in the LVOT of 80 mmHg and clinically in functional group IV (rest dyspnea), in spite of medication in full doses. *ECG diagnosis:* Left chamber enlargement: LAE+LVH, systolic pattern of repolarization or stress pattern. It is decided by a reduction of the hypertrophic septum by injection of absolute alcohol in the first perforating artery, branch of anterior descending artery (transluminal percutaneous ablation).

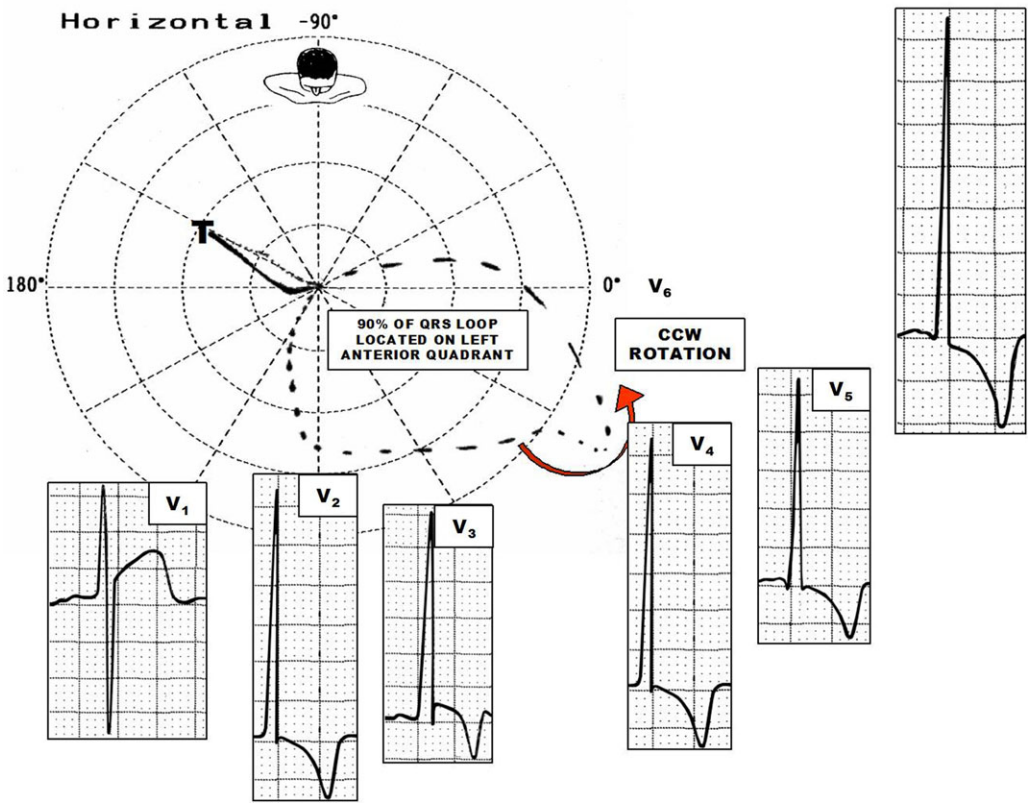

Figure 8. ECG/VCG correlation in the horizontal plane of a patient carrier of apHCM. Notice the significant anterior and leftward shift of the QRS loop located predominantly in the left anterior quadrant responsible for R of great voltage in the precordial leads (prominent anterior forces). T loop located in the right posterior quadrant, which justifies the deeply negative T waves in the precordial leads from V_2 to V_6, characteristic of apHCM.

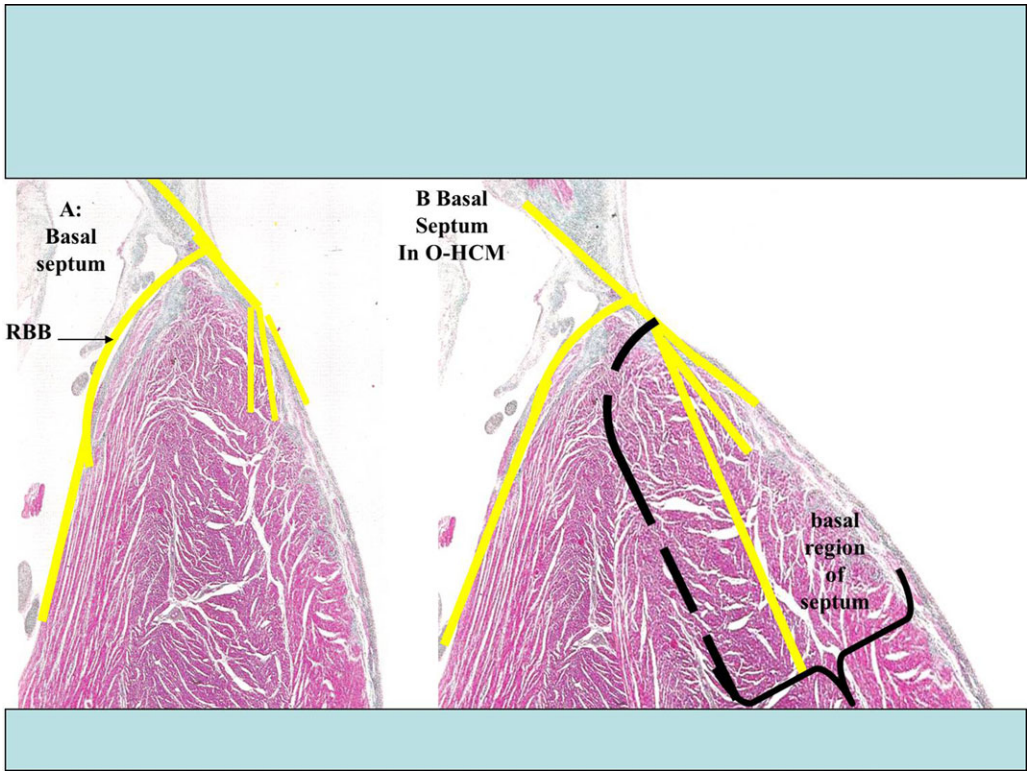

Figure 9. A: Representation of the normal basal IVS. B: Representation of the hypertrophic basal IVS. During the surgical procedure, the surgeon removes a small portion of the hypertrophic basal left septum, where the trunk of the left branch, and the onset of the its fascicles or divisions run. This explains the pattern of CLBBB observed in a high percentage of cases after this procedure. On the contrary, the percutaneous procedure causes necrosis with a basal transmural location, and somewhat lower, that extends to the right septal surface where the right branch of the His bundle is located, which explains why CRBBB is the rule after this procedure.

in V_5 and V_6 and/or the inferior leads, as a consequence of the increase in voltage of the first septal vector.

Ventricular Repolarization

Inverted T waves from V_1 through V_3 are relatively common in athletes younger than 16 years old, and represent a youthful electrocardiographic pattern. Inverted and deep T waves beyond V_2 in ≥16 years old have negative T waves beyond V_2.[35]

The presence of inverted T waves in ≥2 contiguous leads in the anterior wall (V_1-V_4) or lateral wall (V_5-V_6) (but not in aVR and III) is a cause of great concern in sports cardiologists, since these alterations suggest HCM or ARVD.[36] Inverted T waves may represent the first manifestation of inherited myocardial disease in absence of any of the other characteristics and before the structural changes in the heart may be detected. However, to this date there are no proofs that T wave inversions may always indicate the presence of cardiomyopathy or channelopathy in asymptomatic athletes.[36] The inversion of T waves from V_1 through V_4 may rarely represent a variant of the athlete's heart. On the contrary, T waves inversion in the lateral wall leads express underlying cardiomyopathy.[37]

ST segment depression is considered pathological if present in at least two contiguous leads, with a depth greater than 0.1 mV or greater than 0.05 mV if horizontal or leaning downward.

Nonspecific ST or T waves anomalies, minor repolarization changes, are considered to be present if the ST segment or T wave do not meet the T wave inversion or ST segment depression criteria.

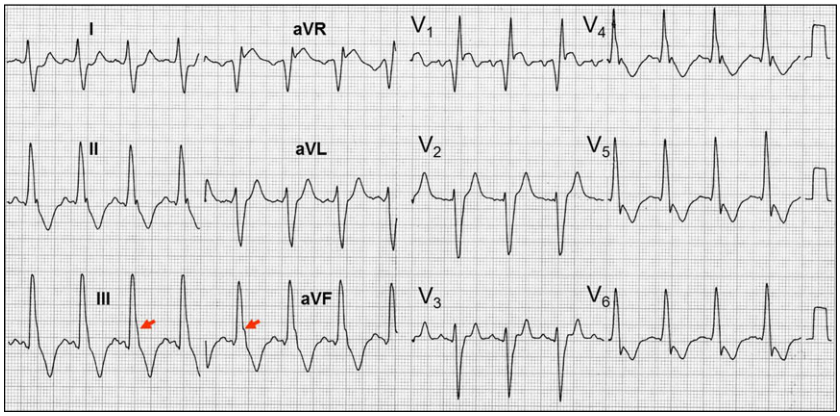

Figure 10. Clinical data: Patient carrier of severe HCM-OF treated with injection of absolute alcohol. The ECG was made immediately after the injection of absolute alcohol in the first perforating artery of the anterior descending artery. *Electrocardiographic diagnosis:* Sinus tachycardia, left atrial enlargement, left ventricular hypertrophy, complete right bundle branch block, and left posterior fascicular block: I and aVL, rS, qR in III, RIII>RII, notch in the descending ramp of the R wave of III and aVF and the electrical axis of QRS shifted to the right (+110°).
Septal infarction located: QR in V_1 and ST segment elevation, of the subepicardial lesion type.

Figure 7 shows the ECG of a patient carrier of severe HCM-OF with the typical pattern of ventricular repolarization stress.

In apHCM, ECG may show wide R waves in all the anterior wall with prominent QRS forces of anterior location and to the left, consequence of apical hypertrophy and maybe some degree of RVH. In the vectorcardiogram, the QRS loop is located predominantly in the left anterior quadrant and T loop in the opposite right posterior quadrant, which justifies the deeply negative T waves recorded in the anterior wall.

The absence of initial r waves in the right precordial leads from V_1 to V_3 with sudden increase of R wave in V_4 may be observed and may resemble anteroseptal infarction (Fig. 8).

Advanced or complete left bundle branch block is the rule after transvalvular myotomy/myomectomy surgery (80% of the cases; Fig. 9).[38]

CRBBB is the predominant dromotropic disorders after injection of absolute alcohol in the first septal perforating artery in transluminal septal percutaneous ablation (Fig. 10).[39]

Arrhythmias. Present in 85% of the cases.

NS-MVT is considered a marker of bad prognosis when it presents in the ECG or Holter in patients with syncope and has a high negative predictive value.[40]

S-MVT in patients carriers of HCM is rare,[41] except HCM with mid-ventricular obstruction and apical aneurysm.

AF may be observed (10%) and in Holter, frequent premature ventricular contractions (>10/h) in 20% of the cases: isolated, coupled (25%), and/or polymorphic (20%).

Electrocardiographic alterations observed after the percutaneous treatment with absolute alcohol injection in the first septal perforating artery.

(1) In ≈70% of the cases, permanent CRBBB occurs (Fig. 10). This is the most common dromotropic disorders as a consequence of this procedure.
(2) First degree AV block observed temporarily in ≈30% of the cases.
(3) In ≈50% temporary complete AV block develops.
(4) Permanent complete AV block is observed in approximately 15% of the cases.

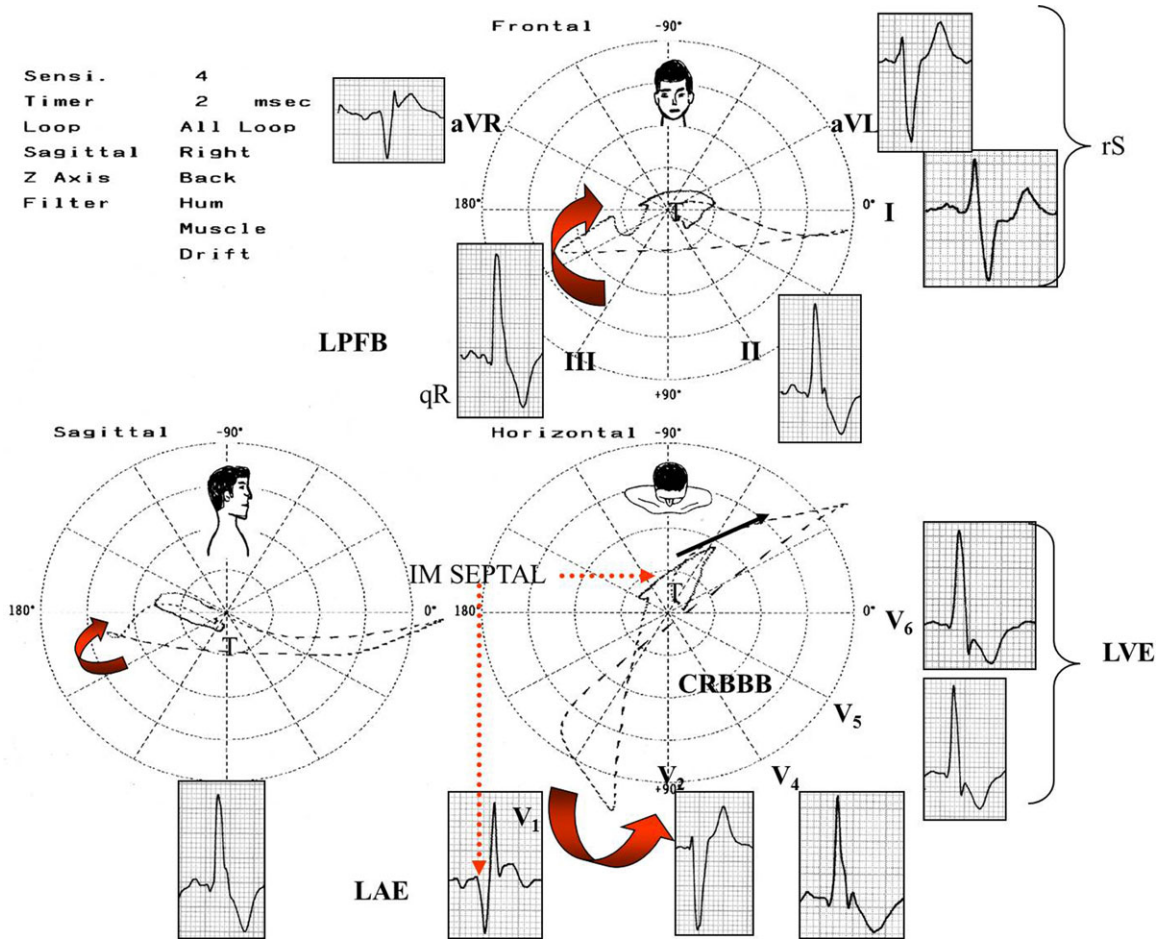

Figure 11. Patient carrier of severe HCM-OF, treated with injection of absolute alcohol. The ECG/VCGs were made immediately after the injection of absolute alcohol in the first perforating artery of the anterior descending artery. The electro-vectorcardiographic correlation is shown in the three planes: FP, HP and RSP. Notice the typical ECG/VCG pattern of left posterior fascicular block associated to CRBBB in the frontal plane: • QRS loop with duration >120 ms or more. Sixty or more dashes (CRBBB). • More than 40% of the area of the QRS loop is located to the right of the orthogonal lead Y. • Vector of the final 20 ms of the QRS loop with significant delay and located at the right. • I and aVL: with rS pattern. • II, III and aVF with qR pattern. • RIII>RII: AQRS closer to +120° than +60°. • Notch in the descending limb of R in II and III. *LPFB:* Left posterior fascicular block; *CRBBB:* complete or advanced right bundle branch block; *LAE:* Left atrial enlargement; *LVH:* Left ventricular hypertrophy.

(5) The late appearance (from 28 to 120 hours) of complete AV block occurs very rarely.
(6) The presence of pattern of preexisting complete left bundle branch block is strongly associated to the development of complete AV block.[42]
(7) Left fascicular blocks (Figs. 10 and 11) associated to CRBBB are rarely observed.
(8) Transient prolongation of QT and QTc interval and increase in QT/QTc dispersion.
(9) JTc interval not affected.
(10) JT and JTc intervals dispersion is transiently observed.[43]

Apical Cardiomyopathy (apHCM)

A variety of HCM where hypertrophy is confined to the apex. This variant was described for the first time in Japan (Japanese-type), where the prevalence is much higher than in the Western world.[44] In spite of its low prevalence in the West, physicians that deal with patients with chest pain should consider apHCM, in its differential diagnosis in the presence of symmetrical and deep negative T waves in the precordial leads.[45]

The diagnosis of apHCM is founded on the following four elements:

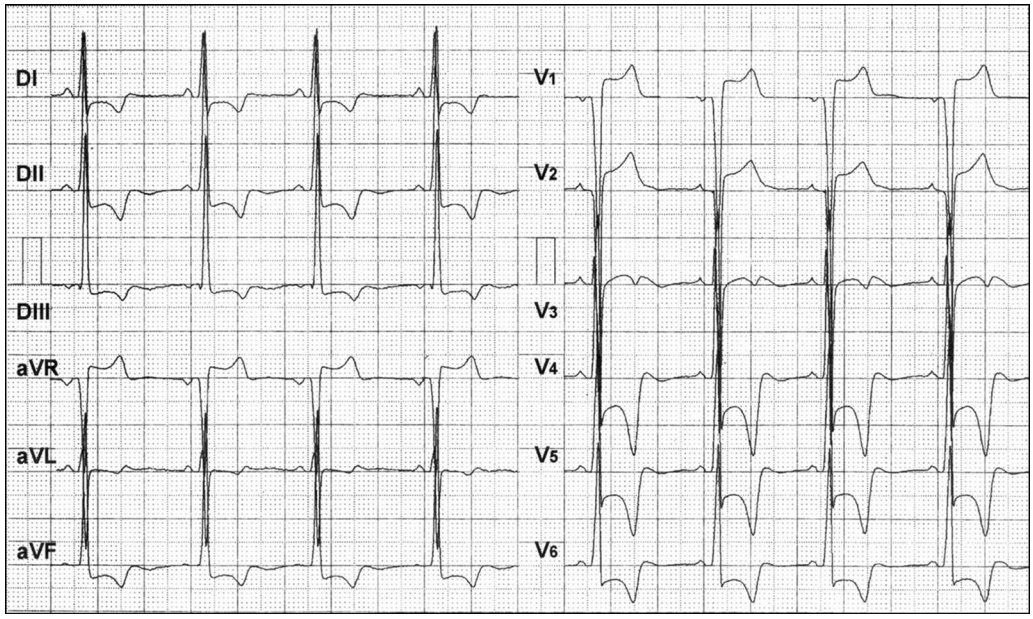

Figure 12. ECG of a Caucasian, 15-year-old teenager, carrier of apical HCM. Apical portion of the septum with 32 mm of diastolic thickness. *Electrocardiographic diagnosis:* Left atrial enlargement (LAE), left ventricular hypertrophy (LVH), systolic pattern of ventricular repolarization by important alteration secondary to ventricular repolarization in the anterior-lateral and inferior wall.Depressed ST segments and deeply negative T waves from V_4 through V_6 and in inferolateral wall.

- Giant and negative T waves from V_2 to V_4. Figures 12 and 13.
- Mild symptoms and benign evolution (although not in all cases).
- Aspect of ace of spades in the right anterior oblique projection of left ventriculography (Fig. 14).
- Absence of ventricular gradient.

The typical electrocardiographic manifestations increase the more advanced the age.

Giant negative T waves ($\geq$1.0 mV or 10 mm)[46] are not pathognomonic of apHCM because they may be observed in the apical shift of the papillary muscle, which points out the need of a careful evaluation of the LV tip by CMR for the accurate diagnosis of apHCM.[47] A pseudo-normalization of giant negative T waves may be observed rarely during a stress test[48] or with dobutamine. A pseudo-normalization pattern of T waves in the ECG without evidence of CAD, should make us consider the diagnostic possibility of apHCM and search for a confirmation by CMR. This pattern of pseudo-normalization could also be a possible explanation for the increase and decrease in the depth of negative T waves observed sometimes in follow-up.[49]

Changes in T polarity may occur on rare occasions with a certain velocity. The disappearance of giant negative T waves may occur slowly and progressively in those patients that develop apical aneurysm.[50]

Eventually, the voltage of R and the negativity of T waves decreases progressively in serial ECGs.

In apHCM patients who develop tip aneurysm with normal coronary arteries, S-MVT and NS-MVT events may appear. In such cases, the risk of SCD is increased.[51] The electrophysiology study showed induction of VF in patients with aborted SCD or with syncope resulting in the need of cardioverter defibrillator implantation and amiodarone. Progressive myocardial necrosis and subsequent formation of apical aneurysm by chronic ischemia is observed at times.[52]

In apHCM, the presence of sustained obliteration of the chamber by significant hypertrophy, manifestations of ischemia and the appearance of prolonged QT interval are significant pathophysiologic conditions that should be considered jointly as important factors in the

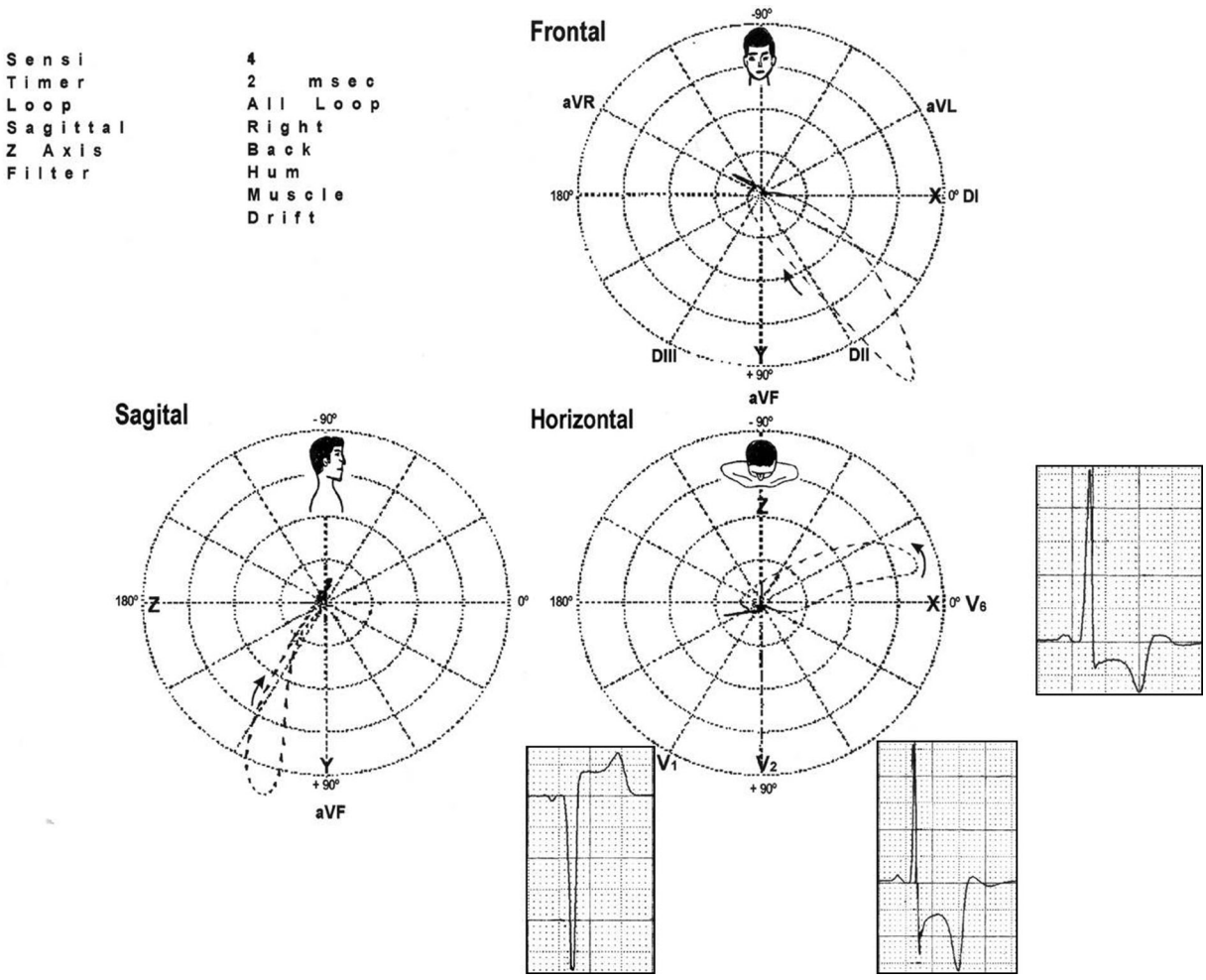

Figure 13. Vectorcardiogram and electro-vectorcardiographic correlation in the HP of the same patient carrier of HCM, apical form (apHCM) of figure 12. The opposition existing between the QRS and T loops is remarkable. The former is located in the left posterior quadrant and the latter in the right anterior quadrant. QRS/T angle close to 180°, which justifies giant negative T waves (≥1.0 mV or 10 mm).

development of apical aneurysm with mutual interactions.[53]

The Vectorcardiogram of HCM

The method allows estimating the left ventricular mass because it relates to the maximal spatial vector voltage of the LV in the QRS loop (normal value between 50 and 90 g/m^2 in children and young adults). A maximal spatial vector of the LV in the QRS loop of 3 mV corresponds to left ventricular mass of 150 g/m^2; values of 4 mV correspond to a mass of 275 g/m^2 and 5 mV are equivalent to left ventricular mass of 400 g/m^2.[54]

In apHCM the following vectorcardiographic elements are outstanding:

Vectors of the initial 10–20 ms of the QRS loop heading to the front and the left.

Possible anteriorization of the QRS loop with predominant location in the left anterior quadrant and final vectors of the QRS loop located at the right and back.

ST/T vector located in the right posterior quadrant with T loop opposite to QRS loop (Fig. 13).

The apHCM is the only case of LVH in absence of coronary insufficiency with T loop located in this quadrant. NOTE: the significant posterior and rightward shift of the ST/T vector is responsible for the characteristic giant negative T waves (>10 mm) from V_2 to V_5. Thee hypothesis attempt to explain the characteristics of negative T waves of the apHCM: apical subendocardial ischemia, apical

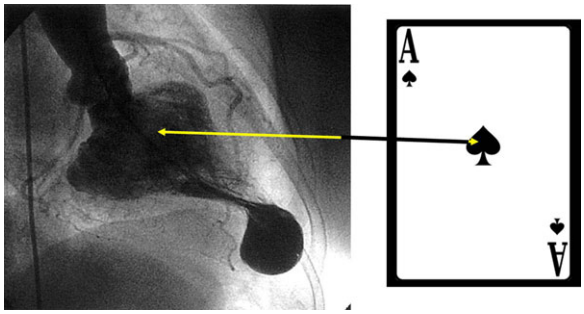

Figure 14. The figure shows the typical configuration in ace of spades (pathognomonic of HCM, apical form (apHCM)) in the right anterior oblique projection of the left ventriculography at the end of systole: ace-of-spades sign or spade-like apical form.

cell disorder and prolongation of action potential duration of hypertrophied cells, which conditions the area to have a slower repolarization.

Risk Factors Associated to SCD in HCM

- Estimation of the myocardial mass very increased and/or extreme increase in septal thickness (≥30 mm).
- Progression of the disease to LV wall thinning and decrease in ejection fraction.
- History of aborted SCD.
- Recurrent syncope in young patient.
- Recording of NS-MVT and S-MVT in Holter in patient with syncope.
- Significant bradyarrhythmia or concealed conduction.
- Inherited genetic defect, associated to unfavorable prognosis.
- Induction of S-VT in electrophysiology study.
- HCM with mid-ventricular obstruction and formation of apical aneurysm by predisposing to S-MVT.[7]
- Myocardial ischemia in young patient that presented altered state of consciousness.
- Hypotension induced by strain.

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