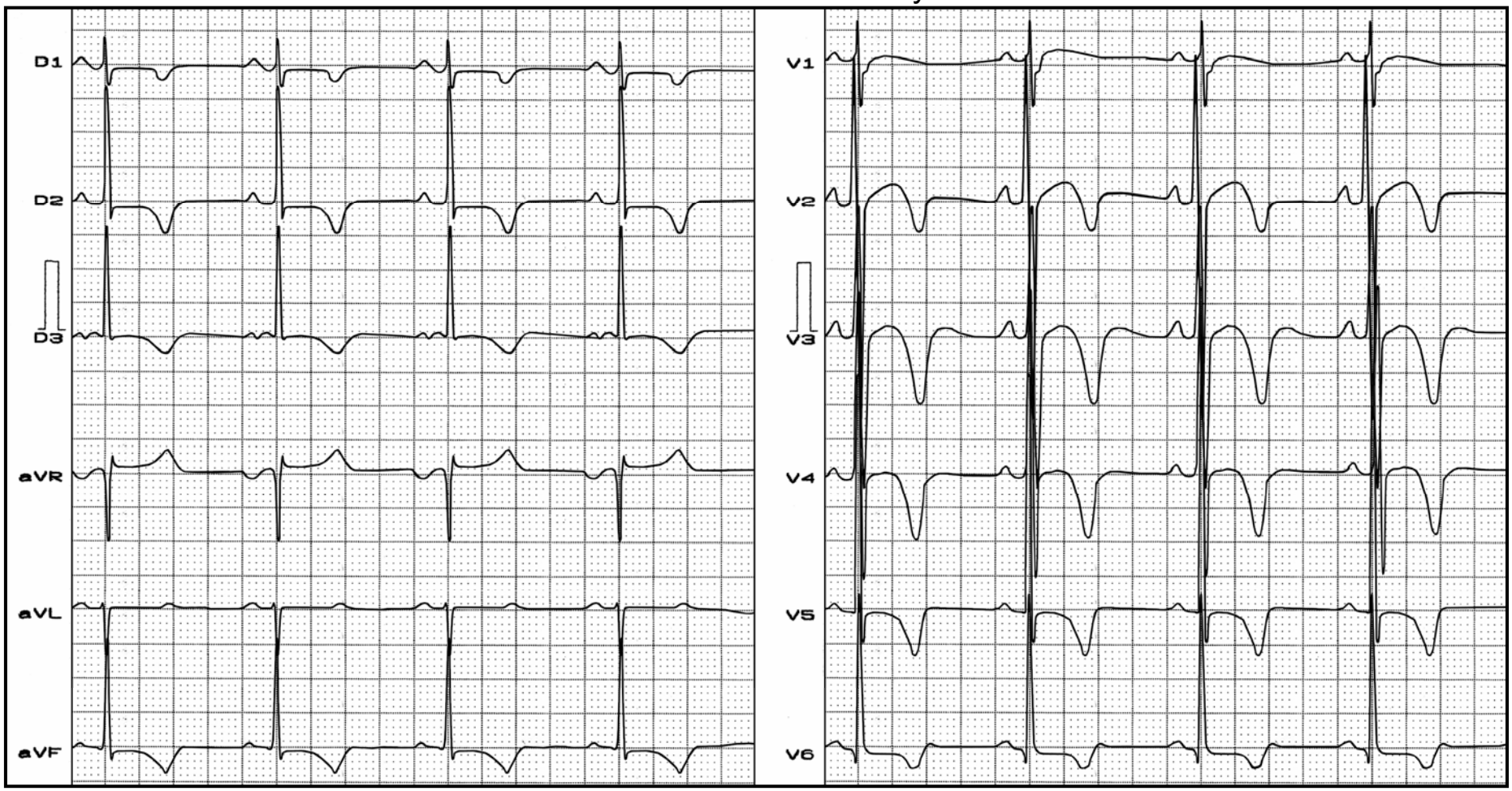
CASE REPORT
ASYMPTOMATIC ELITE SOCCER PLAYER
ATHLETE WITH ATYPICAL ECG

Dr. Andrés Ricardo Pérez Riera
Chief of Electovectorcardiogram Sector of Cardiology Discipline
ABC Faculty – ABC Foundation – Santo André – São Paulo – Brazil
riera@uol.com.br

Name: BJP; **Gender:** Male; **Age:** 24 yo; **Ethnic Group:** Caucasian; **Weight:** 83 Kg **Height:** 1,74 m; **Biotype:** Normoline; **Date:** 05/02/2010

Profession: Elite Soccer Player



Sinus Rhythm, HR 60bpm, normal P wave, normal PR interval duration(165ms), normal QRS axis (+85°), normal QRS duration, giant negative asymmetrical T waves(20mm V3-V4[negativity ≥ 1.0 mV (10 mm)].) in inferoanterolateral walls, T axis (SÂT) on right superior quadrant near +125°, prolonged QT 480ms (for normal adult men with RR 1.00 second HR 60bpm the upper limit is 420ms[1]) Non identifiable U waves.

1. **Sagie A, Larson MG, Goldberg RJ, et al. An improved method for adjusting the QT interval for heart rate (the Framingham Heart Study)Am J Cardiol. 1992 Sep 15;70:797-801**

We have six clinical diagnose possibilities:

1) **Athlete heart ?**
2) **Apical benign Hypertrophyc Cardiomyopathy (ApHCM)?** it is a Non-Obstructive variant of HCM first described in individuals of Japanese descent. The negative T waves seen most strikingly along the midlateral precordial leads are characteristics. Physical examination revealed a displaced apical impulse and a prominent S_4 heart sound. In this circumstance 2-dimensional echocardiography, an apical 4-chamber view of the LV reveal hypertrophy of the apex in an "ace-of-spades" configuration. This apical variant constitutes 25% of cases of HCM in Japan but only 1% to 2% of the cases of HCM in the non-Japanese population.
3) **Solitary Papillary Muscle Hypertrophy: A New Echo-Electrocardiographic Syndrome?** HCM is the term for a heterogeneous group of disorders for which various mutations of genes involving proteins of the cardiac sarcomere lead to hypertrophy of various segments of the LV. The hypertrophy can involve the LV and/or RV, be symmetric or asymmetric, involving the septum, free wall, mid-ventricle, or apex. The phenomenon of **solitary papillary muscle hypertrophy** is rare. Furthermore, giant negative T and U waves are two common ECG phenomena in HCM and have been attributed to hypertrophy of the posteromedial papillary muscle (PMPM). Solitary hypertrophy of the anterolateral papillary muscle (AMPM) might be a new echo-ECG syndrome[1;2;3].
4) **Large muscular false tendons within the LV, and anomalously placed papillary muscles**
5) **Nonspade apical hypertrophic cardiomyopathy**

1. Ker J. Angiology. Solitary papillary muscle hypertrophy: a new echo-electrocardiographic syndrome?. A case report. Angiology. 2007 Aug-Sep;58:502-503.
2. Suwa M, Kobashi Differentiation of solitary papillary muscle hypertrophy from apical hypertrophic cardiomyopathy. A.Circulation. 2000 Apr 11;101:E159.
3. Kobashi A, Suwa M, Ito T, Otake Y, Hirota Y, Kawamura K. Solitary papillary muscle hypertrophy as a possible form of hypertrophic cardiomyopathy. Jpn Circ J. 1998 Nov;62:811-816.
4. Sutton MG, Dubrey S, Oldershaw PJ. Muscular false tendons, aberrant left ventricular papillary musculature, and severe electrocardiographic repolarisation abnormalities: a new syndrome. Br Heart J. 1994 Feb;71:187-190.

5) Nonspade apical hypertrophic cardiomyopathy NonS-ApHCM : This is a subtype of apical hypertrophic cardiomyopathy that could not be diagnosed with the classical diagnostic criteria. ApHCM is recognized by a characteristic spade-shaped intraventricular cavity on the end-diastolic left ventriculogram in the right anterior oblique projection, often associated with giant negative T waves. As an underlying cause of giant negative T waves, an additional subtype of apical hypertrophic cardiomyopathy has been identified by Suzuki et al[1]. In 40 patients with inverted T waves (negativity > or = 0.5 mV), including 26 patients with giant negative T waves, nuclear magnetic resonance (NMR) long-axis images corresponding to the left ventriculogram in the right anterior oblique projection and short-axis images at various levels, including the apical level, were obtained to define the site of hypertrophied myocardium. Long-axis images indicated a spadelike configuration in 17 patients, whereas this diagnostic configuration was not present in the other 23 patients. Nine of these 23 patients had significantly hypertrophied myocardium at the basal level. In the 14 remaining patients, short-axis images indicated no hypertrophy at the basal level and proved that the area of hypertrophied myocardium was confined to a **narrow region of the septum or the anterior or lateral wall at the apical level** (nonspade apical hypertrophic cardiomyopathy). The hypertrophied myocardium of the nonspade type was so narrowly confined that the mass did not form a spadelike configuration or could not be detected on the long-axis image.. Nonspade apical hypertrophic cardiomyopathy was identified on NMR short-axis images, and this is an additional, underlying cause of moderately to severely inverted T waves.

6) Fabry´s disease?

1. Suzuki J, Watanabe F, Takenaka K, Amano K, Amano W, Igarashi T, Aoki T, Serizawa T, Sakamoto T, Sugimoto T, et al. **New subtype of apical hypertrophic cardiomyopathy identified with nuclear magnetic resonance imaging as an underlying cause of markedly inverted T waves.** J Am Coll Cardiol. 1993 Oct;22(4):1175-81.

Apical Hypertrophic Cardiomyopathy (ApHCM)

This entity may cause prominent anterior forces (PAF) translated by R waves with increased voltage in right precordial (V1-V2) and/or intermediary leads (V3-V4)[1].

Prominent R waves in right precordial leads (V3R, V1 and V2) may be observed and mistakenly attributed to RVH, as PAF may be due to hypertrophy in the left septal mass, which causes increase of magnitude of the 1AM vector. Concomitantly, deep q or Q waves may be found; however with duration <40 ms (20 to 50% of cases) in inferior leads and/or from V4 through V6, because the septal vector frequently is heading upward, to the front and the right.

A conclusive proof that wide R wave of right precordial leads may be due to left septal mass hypertrophy, is its disappearance after surgical myectomy on the LVOT area in patients with severe obstructive HCM, non responsive to drugs.

Exceptionally, patterns of true RVH have been described in HCM, originating PAF, right anterior potentials, predominant and not dependent on left septal mass hypertrophy, since the echocardiographic study reveals RVH[2].

In nonobstructive forms hypertrophic nonobstructive cardiomyopathy forms (NO-HCM), Japanese researchers[3] have highlighted the relative frequency with which the typical electrovectocardiographic features of LSFB are observed:

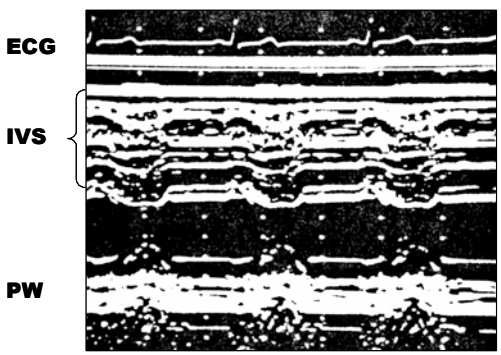
1. R waves with great voltage in intermediary right precordial leads (mid-precordial changes)

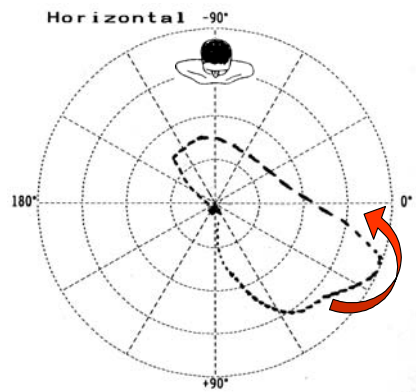
1. **Maron BJ, Wolfson JK, Ciro E, et al. Relation of electrocardiographic abnormalities and patterns of left ventricular hypertrophy identified by 2-dimensional echocardiography in patients with hypertrophic cardiomyopathy. Am J Cardiol. 1983; 51:189-194.**
2. **Comella A, Magnacca M, Gistri R, et al. Right ventricular involvement in hypertrophic cardiomyopathy. A case report and brief review of the literature Ital Heart J Suppl. 2004; 5:154-159.**
3. **Cheng CH, Nobuyoshi M, Kawai C, et al. ECG pattern of left ventricular hypertrophy in non obstrutive hypertrophic cardiomiopathy: The significance of the mid-precordial changes. Am Heart J 1979; 97:687-695.**

2) R wave "in crescendo" from V2 through V4 and decreasing from V5 through V6;
3) Absence of initial q wave in left leads I (87%) and V5 (91%);
4) Marked anterior and left shifts of QRS loop in the HP (74%) (> $^{2}/_{3}$ of the QRS loop area located in the left anterior quadrant)
5) T loop located in the right posterior quadrant (91%).

The intraventricular septum (IVS) is thicker in its inferior part (absence of normal decrease in septal thickness from the base to the apex). Additionally, the free wall of the LV is hypertrophied. Anterior and left shift of QRS loop is marked in the HP, and below and to the left in the FP, translated by R waves of greater voltage in V_4 and II, absence of q waves in the left leads and greater inversion of T wave in V4 and II. The authors attribute these ECG-VCG modifications to selective hypertrophy of the inferior $^{1}/_{3}$ of IVS or apex of (involvement of the distal IVS and the apex). Nakaya et al[1] suggest that the ECG/VCG phenomenon of absence of convexity to the right of the vector of the first 20 ms, the prominent Anterior Forces (PAFwith more than 2/3 of the area located in the left anterior quadrant, without initial q waves in left leads may be due to true Left Septal Fascicular Block (LSFB) and attribute it to LSF involvement by fibrosis of the middle-apical septum.

ECHOCARDIOGRAM AND VECTORCARDIOGRAM HP IN A PATIENT WITH NO-HCM





1. Nakaya Y, Hiasa Y, Murayama Y, et al. Prominent anterior QRS force as a manifestation of left septal fascicular block J Electrocardiol1978; 11:39-46.

NO-HCM localized to the cardiac apex (wall thickening is confined to the most distal region at the apex,) or apical hypertrophic cardiomyopathy (ApHCM) is a specific variant of HCM. This disease has been first described in Japan where the prevalence is much higher than in the western world.
ApHCM, occurs in only 1 to 2% of the non-Japanese population. Only a limited number of sarcomere gene defects (eg, cardiac actin Glu101Lys) consistently produce ApHCM[1]. A single amino acid substitution in actin causes either CHF or maladaptive cardiac hypertrophy, depending on its effect on actin structure and function. De novo mutations in cardiac actin gene were identified in two patients with sporadic HCM who presented with syncope in early childhood. Patients were heterozygous for missense mutations resulting in Pro164Ala and Ala331Pro amino acid substitutions, adjacent to regions of actin-actin and actin-myosin interaction, respectively. A mutation that cosegregated with familial HCM was also found, causing a Glu99Lys substitution in a weak actomyosin binding domain. The cardiac phenotype in many affected patients was characterized by an ApHCM[2].

The typical features of AHC include:

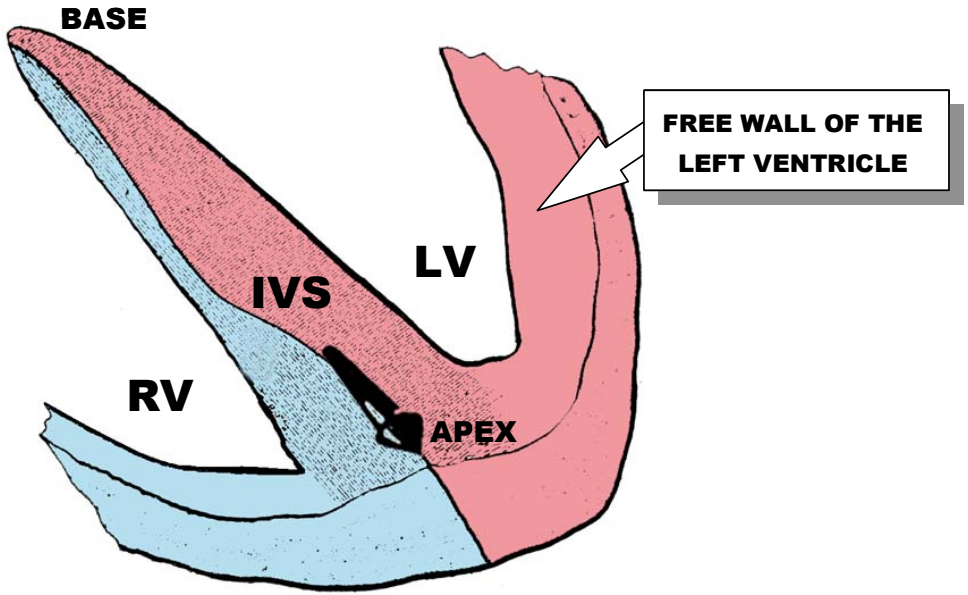
1. Giant negative T waves in the precordial ECG leads Giant negative T waves negativity mayor or equal 1.0 mV (10 mm). Giant negative T waves are more common in Japanese patients than American patients: 15% in Japan vs 3% in US[3];
2. Sometimes R-wave voltage and T-wave negativity progressively decreased in magnitude at serial ECGs;
3. Non-SVT or S-VT in patients that developed apical aneurysm with normal coronary arteries;

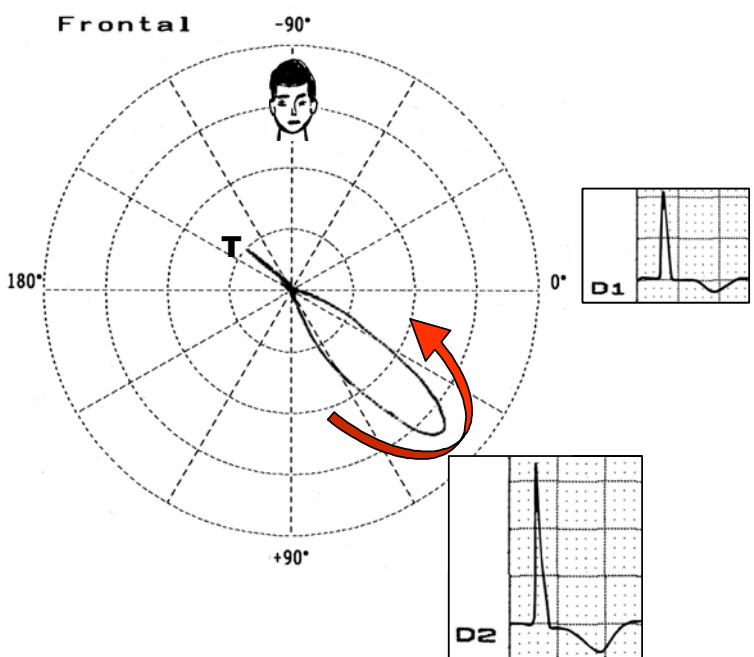
1. **Arad M, Penas-Lado M, Monserrat L, et al. Gene mutations in apical hypertrophic cardiomyopathy. Circulation. 2005; 112: 2805-2811.**
2. **Olson TM, Doan TO, Kishimoto NY, et al. Inherited and de novo mutations in the cardiac actin gene cause hypertrophic cardiomyopathy. J Mol Cell Cardiol. 2000; 32: 1687-1694.**
3. **Kitaoka H, Doi Y, Casey SA, Comparison of prevalence of apical hypertrophic cardiomyopathy in Japan and the United States. Am J Cardiol. 2003; 92:1183-1186.**

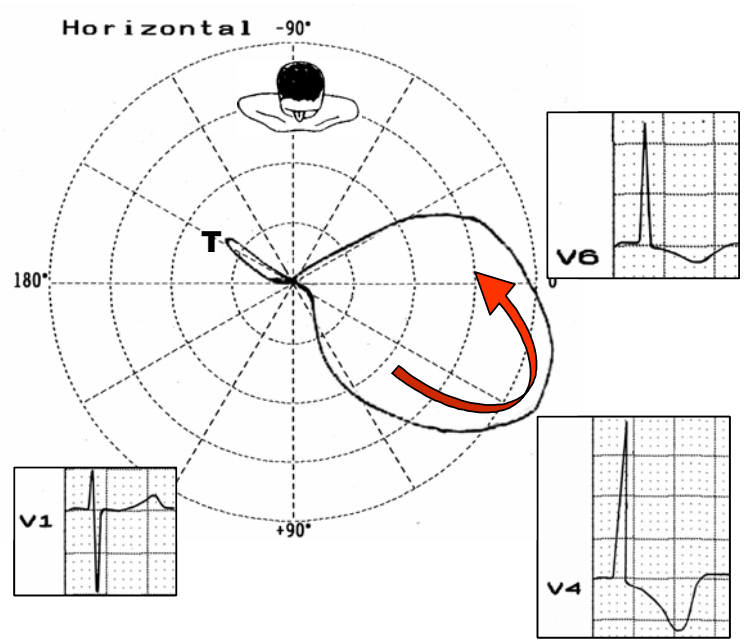
4. A spade-like configuration of the LV at end-systole in the right anterior oblique projection. Non-spade ApHCM was newly identified on cardiac magnetic resonance (CMR) short-axis images, and this could be an additional, important underlying cause of moderately to severely inverted T waves. The area of hypertrophied myocardium is confined to a narrow region of the septum or the anterior or lateral wall at the apical level (non-spade apical hypertrophic cardiomyopathy[1];
5. Absence of an outflow tract pressure gradient;
6. Mild symptoms
7. The prognosis of ApHCM with regard to SCD is believed to be better than that of common HCM. Patients with the ApHCM had a benign clinical course. However, the mutation Arg719Trp in the cardiac beta-myosin heavy chain (beta MHC) gene is a high risk factor for SD and can be associated with an unusual ApHCM[2].
8. Progressing to myocardial necrosis and aneurysm formation because of the chronic myocardial ischemia at the apex eventually is observed[3];
9. 123I-MIBG imaging revealed regional sympathetic denervation in the inferior and lateral regions. Recent observations suggest that the risk of SCD might be increased not only in common HCM, but also in Japanese-type ApHCM[4].
10. PES demonstrated reproducible induction of VF in aborted SD and presyncopal patients, resulting in the need for an ICD and amiodarone.
11. Patients with refractory AF with a rapid ventricular response suffered from serious CHF. A prudent assessment and strategy in patients with this disease would be indispensable in avoiding a disastrous outcome.

1. **Suzuki J, Watanabe F, Takenaka K, et al. New subtype of apical hypertrophic cardiomyopathy identified with nuclear magnetic resonance imaging as an underlying cause of markedly inverted T waves J Am Coll Cardiol. 1993; 22: 1175-1181.**
2. **Dohlemann C, Hebe J, Meitinger T, Apical hypertrophic cardiomyopathy due to a de novo mutation Arg719Trp of the beta-myosin heavy chain gene and cardiac arrest in childhood. A case report and family study. J Am Coll Cardiol, 2002; 39:638-645.**
3. **Marcus CB, Kapoor A, Donohue TJ Apical aneurysm in a patient with apical hypertrophic cardiomyopathy. Conn Med. 2006; 70:297-300.**
4. **Ridjab D, Koch M, Zabel M, Schultheiss HP, Morguet AJ. Cardiac Arrest and Ventricular Tachycardia in Japanese-Type Apical Hypertrophic Cardiomyopathy. Cardiology. 2006; 107:81-86.**

ApHCM ANATOMIC FEATURES AND ECG/VCG CORRELATION ON FP AND HP







To clarify the mechanisms of ECG abnormalities in HCM, 102 patients were examined with CMR. Distribution and magnitude of hypertrophy and late-enhancement were correlated with ECG abnormalities:

Abnormal Q waves reflect the interrelation between upper anterior septal thickness and other regions of the LV and RV, and wider Q waves are associated with late-enhancement;

Conduction disturbances and absent septal Q waves are associated with late-enhancement;

The depth of negative T waves is related to craniocaudal asymmetry and apical late-enhancement[1].

As many as 25% of Japanese patients with HCM have predominately apical involvement. Despite its low incidence, physicians caring for patients with chest pain need to consider ApHCM, in their differential diagnosis[2].

In ApHCM, sustained cavity obliteration is an important pathophysiologic condition as well as hypertrophy, ischemia, and prolonged QTc, which are considered jointly related to the development of aneurysm through interactions[3].

1. **Dumont CA, Monserrat L, Soler R, et al. Interpretation of electrocardiographic abnormalities in hypertrophic cardiomyopathy with cardiac magnetic resonance. Eur Heart J. 2006; 27:1725-1731.**
2. **Iskandar SB, Dittus K, Merrick D. Uncommon cause of a common disease. South Med J. 2003; 96:828-830.**
3. **Matsubara K, Nakamura T, Kuribayashi T, et al. Sustained cavity obliteration and apical aneurysm formation in apical hypertrophic cardiomyopathy. J Am Coll Cardiol. 2003; 42:288-295.**

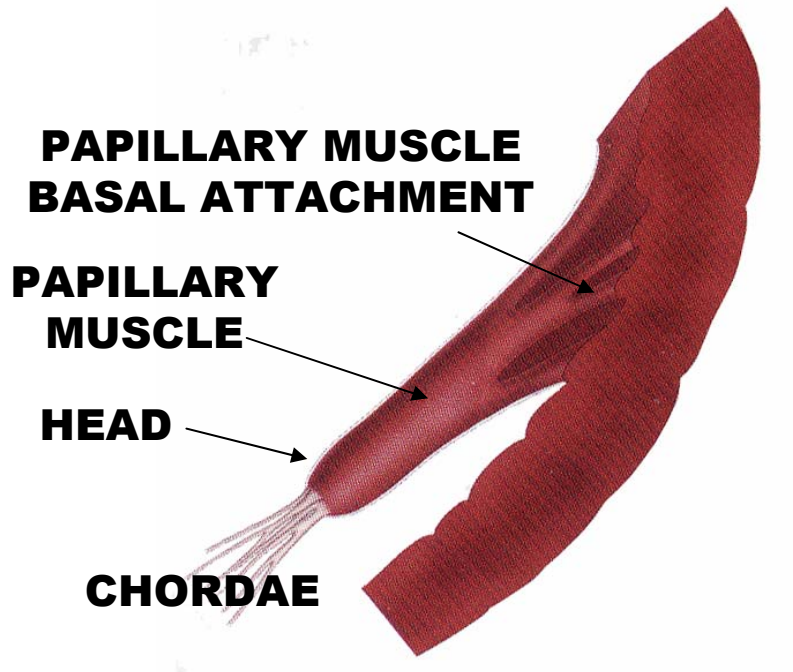
PAPILLARY MUSCLE HYPERTROPHY ANATOMY

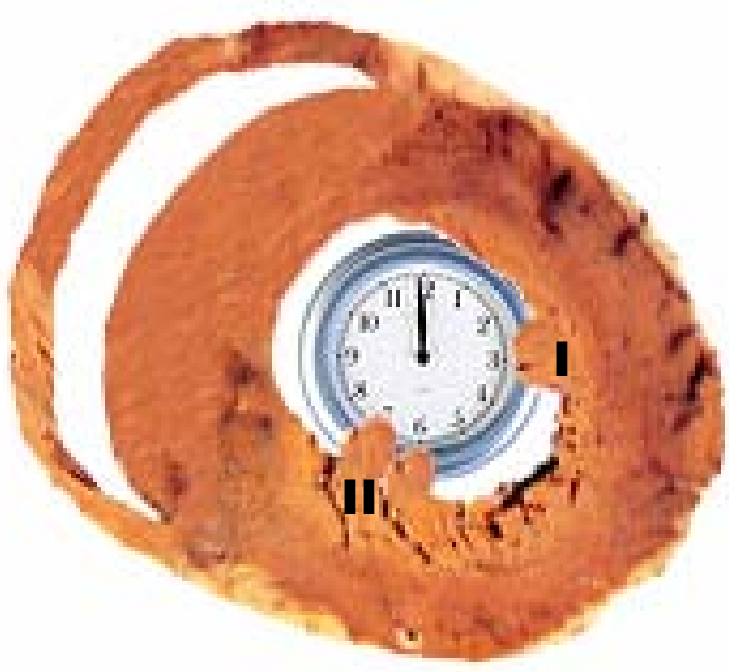
In anatomy, the papillary muscles of the heart serve to limit the movements of the mitral and tricuspid valves. These muscles contract to tighten the chordae tendineae, which in turn prevent inversion. This occurs in response to pressure gradients. Instead they brace the valves against the high pressure, preventing regurgitation of ventricular blood back into the atrial cavities.

Papillary muscle are small muscles within the heart that anchor the heart valves. The anchor ropes are the chordae tendineae, thread-like bands of fibrous tissue that attach on one end to the edges of the tricuspid and mitral valves of the heart and on the other end to the papillary muscles.

The right atrioventricular valve, has three cusps: an anterior, septal and a posterior cusp. The names of the cusps indicate their relative positions. The chordae tendineae, the small fibrous cords, that extend from the cusps of the tricuspid valve to three papillary muscles. The anterior papillary muscle is the largest of the three papillary muscles and is attached to the inferior border of the anterior wall. The posterior papillary muscle is a relatively large muscle located on the posterior wall. The septal papillary muscles are small and often multiple.

The left atrioventricular, or mitral, valve is made up of two cusps an anterior and a posterior cusp. These cusps are attached to the anterior or Antero Lateral Papillary Muscle (ALPM) and posterior or Postero Medial Papillary Muscle (PMPM) by chordae tendineae. The papillary muscle basal attachment is widely used in cardiac imaging to segment the apical and the middle third of the LV cavity. Most commonly, the ALPM complex comprises one trunk and head in 90% of cases and two trunks and heads in 10% of cases. Most commonly(75%), the PMPM complex comprise either two trunks or one trunk with two heads. The two trunks or heads of the PMPM may be positioned to the side or beside each other, such that they can be well appreciated only in cross-sectional short0axis imaging.





Short axis view at the mid ventricle looking toward the base. The PMPM generally has either two bodies or heads., whereas the ALPM has a solitary body and head. The PMPMis generally located between the 6-o´clock and 8-o´clock positions. The papillary muscles support the corresponding half of the mitral vale commissure. The ALPM extends chordae to both the anterior and posterior leaflets along the lateral commissure, whereas the PMPM heads support their corresponding leaflet of the medial commissure.

Irrigation

I) ALPM= blood supply from the diagonal branches from the Left Anterior Descending(LAD) and also an obtuse marginal branch from the left circunflex(LCx).

II) PMPM= blood supply only from branches of the posterior descending artery

Solitary Papillary Muscle Hypertrophy

Austin et al[1] report the case of a 18-year-old patient, gene-positive for HCM, who presented with symptomatic dynamic LVOT obstruction caused by an abnormally thickened papillary muscle in the absence of septal hypertrophy. This was confirmed using multimodality imaging, including ECHO and MRI. He successfully underwent surgery for papillary muscle realignment without septal myectomy.

Papillary muscle anatomic alteration can shows ECG manifested by ST elevation and T-wave inversion[2]. An accessory papillary muscle with a prominent J-wave was recently described[3].

HCM is the term for a heterogeneous group of disorders for which various mutations of genes involving proteins of the cardiac sarcomere lead to hypertrophy (symmetric or asymmetric) of various segments of the LV and/or RV involving the septum, free wall, mid-ventricle, apex and papillary muscle. The phenomenon of solitary papillary muscle hypertrophy is rare. Giant negative T and giant U waves are common ECG features in HCM and have been attributed to hypertrophy of the posterior papillary muscle. Solitary hypertrophy of the anterior papillary muscle might be a new Echo-ECG syndrome[4]. Delayed repolarization of the papillary muscles named by Bufalari and Furbetta “the syndrome of the papillary muscles” is considere the source of the U wave[5].

1. **Austin BA, Kwon DH, Smedira NG, et al. Abnormally thickened papillary muscle resulting in dynamic left ventricular outflow tract obstruction: an unusual presentation of hypertrophic cardiomyopathy. J Am Soc Echocardiogr. 2009 Jan;22:105.e5-6.**
2. **Shim CY, Ha JW, Hong SJ, et al Uncommon variation in the papillary muscles presenting with ST elevation and T-wave inversion.Eur Heart J. 2008 Nov;29:2633.**
3. **Ker J, du Toit L. The accessory papillary muscle with inferior J-waves--peculiarity or hidden danger? Cardiovasc Ultrasound. 2009 Oct 29;7:50.**
4. **Ker J. Solitary papillary muscle hypertrophy: a new echo-electrocardiographic syndrome? A case report. Angiology. 2007 Aug-Sep;58:502-503.**
5. **Pérez Riera AR, Ferreira C, Filho CF, Ferreira M, Meneghini A, Uchida AH, Schapachnik E, Dubner S, Zhang L. The enigmatic sixth wave of the electrocardiogram: the U wave. Cardiol J. 2008; 15: 408-421.**

These authors postulate that the U wave represents repolarization of the papillary muscle and neighboring structures. The common factor underlying the varied cardiac pathology is regarded to be ischemia, "strain," or other functional derangement of the papillary muscles in the right or left ventricles. The authors believe that various abnormalities of the papillary muscles, whether anatomic or functional, are detectable by modifications on U waves and T-U segment

They described three different vectorial patterns[1]:

Left Papillary Muscle Syndrome: Negative U wave in left leads I, VL, V5-V6, because spatial U vector is directed to the front and right. Observed in anterior myocardial infarction, hypertension and aortic valvular disease.

Right Papillary Muscle Syndrome: Negative U wave is observed in III, sometimes in VF, and right precordial leads. Observed in right ventricular enlargement and congenital heart disease.

Biventricular Papillary Muscle Syndrome: Negative U wave in all precordial leads, II and VL. Biventricular enlargement (strain of both ventricles).

Prominent U waves in the inferior leads are caused by an accessory papillary muscle[2]. Any possible long-term consequences are not known.

Sutton et al[3] reported three patients with normal exercise capacity, normal LV function, large muscular false tendons within the LV, and anomalously placed papillary muscles associated with giant T wave inversion on the ECG. The authors conclude that the absence of adverse cardiovascular events at long-term follow up (mean 13 years) indicates that this is a benign unrecognised syndrome.

1. **Bufalari A, Furbetta D, Santucci F, Solinas P. Abnormality of the U wave and of the T-U segment of the electrocardiogram; the syndrome of the papillary muscles. Circulation. 1956; 14: 1129-1137.**
2. **Ker J. The U wave and papillary muscle variants: revisiting an old association. Cardiovasc J Afr. 2009 Jul-Aug;20:256-257.**
3. **Sutton MG, Dubrey S, Oldershaw PJ. Muscular false tendons, aberrant left ventricular papillary musculature, and severe electrocardiographic repolarisation abnormalities: a new syndrome. Br Heart J. 1994 Feb;71:187-190.**

Reddy et al[1] presented in Circulation a case with ApHCM. The authors mentioned that the 2D ECHO obtained from this patient indicated hypertrophy of the apex and that 201-Tl scintigraphic imaging demonstrated an increased count in the apical segment. However, because there was a clear groove or space between the hypertrophic segment and the lateral free wall in the 2D ECHO, the hypertrophic segment in this patient was not the LV free wall, but rather the ALPM, and that the scintigraphic finding was also related to the hypertrophic ALPM.

Kobashi et al[2], reported that patients with solitary papillary muscle hypertrophy showed LVH with not-so-prominent but symmetrical, negative T-waves on the ECG. Some of these patients had relatives with clinical features of HCM. These authors consider isolated papillary muscle hypertrophy a newly identified subtype of an early form of HCM. The case reported by Reddy et al had features typical of and suggestive of solitary ALPM hypertrophy; thus, these authors think that it is not a case with ApHCM.

1. **Reddy V, Korcarz C, Weinert L, Al-Sadir J, Spencer KT, Lang RM. Apical hypertrophic cardiomyopathy. *Circulation.* 1998;98:2354.**
2. **Kobashi A, Suwa M, Ito T, Otake Y, Hirota Y, Kawamura K. Solitary papillary muscle hypertrophy as a possible form of hypertrophic cardiomyopathy. *Jpn Circ J.* 1998;62:811–816**

FABRY´S DISEASE

Others denominations: Anderson-Fabry disease, angiokeratoma corporis diffusum and alpha-galactosidase A deficiency.

Fabry disease is a rare X-linked recessive (inherited) lysosomal storage disease(It is now apparent that this disorder may be much more common than previously suspected[1].), which can cause a wide range of systemic symptoms. A deficiency of the enzyme alpha galactosidase A (a-GAL A, encoded by GLA) due to mutation causes a glycolipid known as globotriaosylceramide (abbreviated as Gb3, GL-3, or ceramide trihexoside) to accumulate within the blood vessels, other tissues, and organs. This accumulation leads to an impairment of their proper function.

The DNA mutations which cause the disease are X-linked recessive. The condition affects hemizygous males (i.e. all males), as well as homozygous, and potentially heterozygous (carrier), females. Female carriers are at risk of developing disease, but this tends to be milder and more slowly progressive than in males. This variability is thought to be due to X-inactivation patterns during embryonic development of the female.

Cardiac Finding: LVH is the most common cardiac manifestation followed by conduction system disease, valve dysfunction, arrhythmias and hypertension[2]. Angina or MI with normal coronary artery caused by accumulation of lipid moieties in coronary endothelial cells, increase LV wall thickness simulating HMC or amiloidosis.

LV dysfunction and failure, mitral regurgitation and systemic hypertension is frequent consequence of kidney complications are a common and serious effect of the disease; renal insufficiency and renal failure may worsen throughout life. Proteinuria (which causes foamy urine) is often the first sign of kidney involvement. End stage renal failure in males can typically occur in the third decade of life, and is a common cause of death due to the disease.

1. **Mehta AB. Anderson-Fabry disease: developments in diagnosis and treatment. Int J Clin Pharmacol Ther. 2009;47 Suppl 1:S66-74.**
2. **O'Mahony C, Elliott P.Prog Cardiovasc Dis. 2010 Jan-Feb;52(4):326-35.Anderson-Fabry disease and the heart.**

ECG features:

Most patients have resting bradycardia, with impaired ability to increase heart rate during exercise[1]. Short PR interval is a characteristic feature[3]. First-degree AV block in more advance stage is obseved[2], LVH ECG criteria is present in ≈ 70% of cases[2] and at end stage ECGs reveal the presence of LVH in all patients, ST segment and T abnormalities giant negative T waves[3;4] conduction abnormalities and NS-VT[5; 6]. **RNM** for differential diagnosis with HCM, cardiac amyloidosis[5].

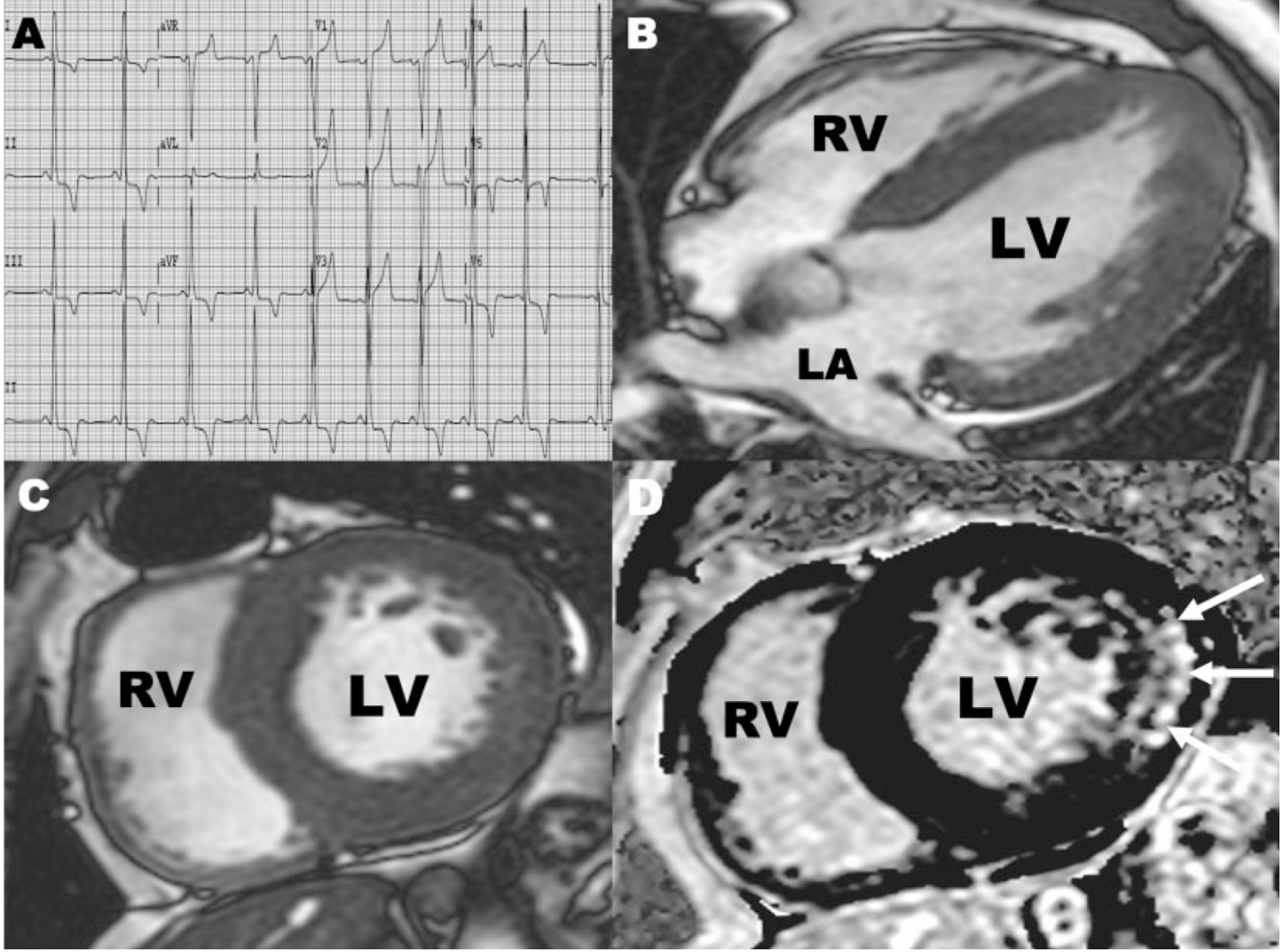
Ergometer test: Male patients with Fabry disease were unable to attain predicted maximal heart rate on exercise or to achieve normal exercise levels. ERT was associated with a small improvement in anaerobic threshold over the first year.

Holter: Heart rate variability (HRV) analyses reveal that male, but not female, Fabry patients had significantly reduced HRV, reflecting a reduction in parasympathetic stimulation of the heart[7].

ECHO: LVH on echo is present in 65% of patients and 80% among men.

Definitive diagnosis: EMB. myelinoid lamellar inclusions are demonstrated in myocardial cells electron microscopically.

1. **Lobo T, Morgan J, Bjorksten A, Nicholls K, Grigg L, Centra E, Becker G. Cardiovascular testing in Fabry disease: exercise capacity reduction, chronotropic incompetence and improved anaerobic threshold after enzyme replacement. Intern Med J. 2008 Jun;38:407-414. Blum A, Podovitzky O, Sheiman J, Khasin M. Reversal of first-degree atrioventricular block in Fabry disease. Arch Intern Med. 2009 Nov 9;169:1925-1926.**
2. **Jastrzebski M, Petkow-Dimitrow P. Electrocardiogram in Fabry's disease Kardiol Pol. 2008 Jun;66:688-692.**
3. **Yokoyama A, Yamazoe M, Shibata A. A case of heterozygous Fabry's disease with a short PR interval and giant negative T waves.Br Heart J. 1987 Mar;57:296-299.**
4. **Joshi SB, Ahmar W, Lee G, Aggarwal A. Fabry's disease presenting as ventricular tachycardia and left ventricular 'hypertrophy'. Eur J Echocardiogr. 2008 Sep;9:697-699.**
5. **Gange CA, Link MS, Maron MS. Utility of cardiovascular magnetic resonance in the diagnosis of Anderson-Fabry disease. Circulation. 2009 Sep 29;120:e96-97.**
6. **Takenaka T, Teraguchi H, Yoshida A,et al. Terminal stage cardiac findings in patients with cardiac Fabry disease: an electrocardiographic, echocardiographic, and autopsy study.J Cardiol. 2008 Feb;51:50-59.**
7. **Kampmann C, Wiethoff CM, Whybra C, et al. Cardiac manifestations of Anderson-Fabry disease in children and adolescents. Acta Paediatr. 2008 Apr;97:463-469.**



A 54-year-old man with Anderson-Fabry disease and cardiac involvement.
A, ECG showing sinus arrhythmia, LBBB, and LV hypertrophy with strain ST- and T-wave abnormalities.
B and C, Cine CMR at end diastole in the long-axis 4-chamber view (B) and the basal LV short-axis view (C) demonstrating concentric LV hypertrophy (17 mm maximal wall thickness in ventricular septum and LV free wall) with biatrial enlargement. D, Contrast-enhanced CMR image of the basal LV short axis with an area of late gadolinium enhancement confined to the midmyocardium of the inferolateral wall (arrows).

1. **Gange CA, Link MS, Maron MS. Utility of cardiovascular magnetic resonance in the diagnosis of Anderson-Fabry disease. Circulation. 2009 Sep 29;120(13):e96-7.**

Dermatological manifestations Angiokeratomas (tiny, painless papules that can appear on any region of the body, but are predominant on the thighs, around the belly-button, buttocks, lower abdomen, and groin) are a common symptom, anhidrosis (lack of sweating) is a common symptom, and less commonly hyperhidrosis (excessive sweating).Additionally, patients can exhibit Raynaud's disease-like symptoms with neuropathy (in particular, burning extremity pain).

Ocular manifestations Cosmetic ocular involvement may be present showing cornea verticillata (also known as vortex keratopathy), i.e. clouding of the corneas. Keratopathy may be the presenting feature in asymptomatic carriers, and must be differentiated from other causes of vortex keratopathy (e.g. drug deposition in the cornea). This clouding does not affect vision.
Other ocular findings that can be seen include conjunctival aneurysms, posterior spoke-like cataracts, papilloedema, macular edema, optic atrophy and retinal vascular dilation.

Other manifestations: Fatigue, neuropathy (in particular, burning extremity pain), cerebrovascular effects leading to an increased risk of stroke, tinnitus (ringing in the ears), vertigo, nausea, inability to gain weight, and diarrhea.

The possible causes of Giant negative T waves

Subarachnoid hemorrhage[1], others cerebrovascular insults[2], after electroconvulsive therapy[3], transient left ventricular apical ballooning or Takotsubo cardiomyopathy[4;5], acute renal failure caused by crush syndrome[6], in middle-aged or elderly women after noncardiac surgery[7], Guillain-Barré syndrome[8], pulmonary edema[9], pulmonary thromboembolism[10], complete atrioventricular block and bradycardia[11], cardiac compression by a giant hiatal hernia[12], myocardial stunning[13], Left ventricular aneurysm of the apex consequence of sarcoidosis[14], apical diverticulum of the left ventricle[15],

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7. Yoshii F, Kozuma R, Haida M, et al. Giant negative T waves in Guillain-Barré syndrome Acta Neurol Scand. 2000 Mar;101:212-215.
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9. Saviotti M, Piccone U, Pala M, et al. Pulmonary thromboembolism. A clinical case with unusual presentation. Minerva Cardioangiol. 1995 Nov-Dec;43:493-499.
10. Johnson CD, Girod CE.Long Q-T interval and giant T wave inversion associated with complete atrioventricular block and bradycardia. Bol Asoc Med P R. 2007 Jan-Mar;99:38-39.
11. Hokamaki J, Kawano H, Miyamoto S, et al. Dynamic electrocardiographic changes due to cardiac compression by a giant hiatal hernia. Intern Med. 2005 Feb;44:136-140.
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13. Kosuge H, Noda M, Kakuta T, et al Left ventricular apical aneurysm in cardiac sarcoidosis. Jpn Heart J. 2001 Mar;42:265-169.
14. Ito M, Tsuchiyama J, Chinushi M, et al. Images in cardiovascular medicine. Transient giant negative T waves associated with cardiac involvement of diffuse large B-cell lymphoma. Circulation. 2005 Nov 15;112(20):e322-3.
15. Barboteu M, Desnos M, Hagège A, et al Giant negative T waves in idiopathic apical diverticulum of the left ventricle in adults Arch Mal Coeur Vaiss. 1995 Oct;88:1475-1477.

The possible causes of Giant negative T waves Cont.

cardiac involvement of diffuse large B-cell lymphoma[16], during interferon therapy in a patient with chronic hepatitis C[17], apical hypertrophic cardiomyopathy (ApHCM)18, solitary papillary muscle hypertrophy[19], Nonspade apical hypertrophic cardiomyopathy (NonS-ApHCM) [20], large muscular false tendons within the LV, and anomalously placed papillary muscles[21], coronary insufficiencies (symmetrical negative T waves) [22], as an unusual ECG responses to exercise stress testing[23], following successful emergent percutaneous coronary intervention for acute coronary syndrome[24], increased serum creatine kinase[25], severe aortic regurgitation[26].

16. Sakamoto T, Suzuki J. Apical hypertrophic cardiomyopathy Nippon Rinsho. 2000 Jan;58:93-101.

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22. Weston CF, Fonfe G, Wahbi Z, et al Giant T wave inversion associated with severe aortic regurgitation. Int J Cardiol. 1996 Jun; 54:277-282.

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The Pre-Participation Screening

Long term Italian experience has provided evidence that preparticipation screening in competitive athletes with history, physical examination and 12-lead ECG,, is effective in identifying potentially lethal cardiovascular diseases. However, it's not being routinely practiced in other countries. In addition to a history and physical, young athletes often have a ECG to better identify heart disease associated with SCD.

Complicating this process is that certain "abnormal" resting ECG findings are considered normal variants in healthy children and young adults athletes.

The ability to recognize these normal variants is often useful in preventing excessive referral of patients to cardiologists for evaluation of resting ECG's that are benign variations of normal and in making sound decisions regarding appropriate clearance to exercise[1]. Given the ability of ECG to detect individuals with structural heart disease, Carro Hevia et al (2) suggest its inclusion as a part of preparticipation screening programmers. These authors screened 1220 athletes: 96% males; players were referred for additional tests because of abnormal findings on baseline examination: 11 (0,9%) personal or family history; 4 (0,08%) physical examination; 75 (6,14%) ECG. Echocardiographic assessment fulfilled LVH criteria in 8 out of the 90 players. Of those, one case was considered an athlete's heart and one case was diagnosed of HCM; septal thickness 23 mm). Further tests were needed in the remaining six, included in the "gray area", with one additional case of HCM. Published literature supports the fact that many pediatric patients at risk for SCD will exhibit warning signs and symptoms.

1. **Higgins JP.Normal resting electrocardiographic variants in young athletes. Phys Sportsmed. 2008 Dec;36:69-75.**
2. **Carro Hevia A, Martin Fernández M, Ania Palacio JM, Hernandez Martín E, García Castro M, Rodriguez Reguero JJ. ECG As A Part of the Pre-Participation Screening Programme: An Old an Still Present International Dilemma. Br J Sports Med. 2009 Oct 25. [Epub ahead of print]**

Anamnesis Medical history

We think that the use of comprehensive cardiovascular screening questionnaires is necessary. Recognizing that many of the cardiac disorders predisposing to SCD are genetic, the role of careful and extensive family history cannot be overemphasized. While preparticipation screening is primarily directed toward high school and college athletes, the use of cardiovascular screening should also be considered for children of all ages.

Personal medical history

- Do you have dyspnea? (present in >90% of symptomatic in cases of HCM)
- Do you have precordial pain?(chest pain is present in 60% of symptomatic in HCM.)
- Do you have dizziness?
- Do you have palpitations?
- Do you had “graying out”(near-syncope). If positive it occur with the erect posture?
- Do you had loss of conscience? (syncope) Circumstances? during exercise? unexplained syncope?
- Aborted SCD?

Family background

Early SCD in first degree family members≤ 45 years old. Unexplained early SCD.

1. **Thünenkötter T, Schmied C, Dvorak J, Kindermann W. Benefits and limitations of cardiovascular pre-competition screening in international football Clin Res Cardiol. 2010 Jan;99(1):29-35**

Physical examination should help identify these HCM patients at risk.
Number one we find a quickie rise pulse
We don't find aortic regurgitation
A sistolic harsh murmur crescendo-decrescendo is frequently present best heard between the apex and left sternal border.
With the squatting maneuver the murmur becomes fainter, and on standing again, the murmur gets louder
The Valsalva maneuver, too, can be helpful in diagnosing HCM. While listening along the left sternal border or apex, have the patient take a deep breath, blow the breath out and then strain as if having a bowel movement. The murmur may increase in intensity, indicating a positive response.
Apical impulse localization and characteristics
S_4 that corresponds to the apical presystolic pulse
S_2 in cases with severe outflow gradient paradoxical splitting may be noted.
Sometimes holosistolic murmur of mitral regurgitation
Diastolic rumbling murmur reflecting increasing transmitral flow.
12-lead resting-electrocardiogram (ECG)
Echocardiography
Vectocardiogram: Increase of anterior forces in almost all cases, dislocation of QRS loop to the front and left in the HP, T loop not matching QRS loop
Exercise testing: it is questionable if exercise testing should be included in this context. An exercise-ECG is especially recommended to evaluate the eligibility for physical exercise in persons > 35-40 years and in subjects with risk factors for cardiovascular diseases.
High Resolution ECG: Presence of late potentials in 10% of the cases against 1.4% in the population of athletes[1];

1) Borbola, J & Denes, P. Late potentials in patents with sustained ventricular tachycardia. In: El-Sherif, N.; Turitto, G (eds). High-Resolution Electrocardiography. Mount Kisco (NY):Futura, 495-520, 1992.

Cardiac structure: key points

1. There is overwhelming evidence that the heart of athletes may differ from that of non-athletes, provided that the training is of sufficient intensity and duration
2. Predominantly eccentric LVH is observed in sports with high dynamic and low static demands (for example, running)
3. Sports with high static demands (for example, weight lifting) lead to predominantly concentric LVH
4. In sports with high dynamic and high static demands (for example, cycling) the LV is mixed and balanced
5. The influence of exercise is shown by the study of athletes in different training states
6. The most important aims are to prevent physiological changes in the athlete being erroneously attributed to heart disease, or signs of life-threatening cardiovascular conditions being dismissed as a normal variant of athlete's heart.
7. As pathological ECG abnormalities not only cause alarm but also require action with additional testing to exclude (or confirm) the suspicion of a lethal cardiovascular disorder
8. Appropriate interpretation of an athlete's ECG will prevent unnecessary distress and also result in considerable cost saving in the context of a population-based preparticipation screening programm[1].

1. **Corrado D, Biffi A, Basso C, Pelliccia A, Thiene G. 12-lead ECG in the athlete: physiological versus pathological abnormalities. Br J Sports Med. 2009 Sep;43:669-676.**

THE ECG IN ATHLETE

The ability to recognize ECG normal variants is often useful in preventing excessive referral of patients to cardiologists for evaluation of resting ECG's that are benign variations of normal and in making sound decisions regarding appropriate clearance to exercise[1]. One of the most important aims of modern sports cardiology is prevention of SCD among athletes. Adequate pre-participation screening is a crucial part of prevention, however, current ACC, AHA or ESC guidelines are not uniform. There is recently ongoing discussion on implementation of 12-lead ECG to the screening protocol. Athlete's heart may be associated with rhythm and conduction alterations, morphological changes of the QRS complex, and repolarization abnormalities[2;3]. Factors which play a role in one or more of these changes are a lower intrinsic heart rate, an increased vagal tone, a decrease in sympathetic tone, structural cardiac adaptations, and non-homogeneous repolarization of the ventricles. Alterations are mostly seen in athletes engaged in high intensity dynamic endurance sports. It is important to recognize that several of the ECG changes that can accompany athletic conditioning resemble pathological ECG features and may mimic structural heart disease.

Limits of athlete's heart: key point

LV wall thickness may exceed 13 mm in highly trained athletes, but the upper physiologic limit appears to be 16 mm. Key features in the distinction between physiologic LVH and HCM are the appropriately increased size of the LV internal dimension in endurance athletes, and the normal systolic and diastolic LV function.

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2. **Huston TP, Puffer JG, Mac Millan Rodney WM. The athletic heart syndrome. N Engl J Med 1985;313:24–32.**
3. **Estes NAM, Link MS, Homoud M, *et al.* Electrocardiographic variants and cardiac rhythm and conduction disturbances in the athlete. In: Thompson PD, ed. Exercise and sports cardiology. New York: McGraw-Hill, 2001:211–32**

ECG manifestations in Athlete Heart

When the ECG of an athlete is examined, the main objective is to distinguish between physiological patterns that should cause no alarm and those that require action and/or additional testing to exclude (or confirm) the suspicion of an underlying cardiovascular condition carrying the risk of SCD during sports.

The European experience suggests that adding the ECG to the standard medical and family history and physical examination can decrease cardiac deaths by 90%. However, there has not been a randomized trial to demonstrate such a reduction. While there are obvious differences between the European and American experiences with athletes including very differing causes of athletic deaths, some would highlight the European emphasis on public welfare vs the protection of personal rights in the USA[1].

Although some authorities advocate the use of ECG screening of young athletes, further studies are required to define what constitutes a normal ECG in athletes, and to determine whether ECG-based screening protocols truly are superior, not only in finding disease, but also saving lives[2]. There remains debate on the how extensive screening should be, in particular over the use of the ECG, with European guidelines mandating ECG and United States guidelines not recommending routine use of the ECG[3].

1. **Perez M, Fonda H, Le VV, Mitiku T, Ray J, Freeman JV, et al. Adding an electrocardiogram to the pre-participation examination in competitive athletes: a systematic review. Curr Probl Cardiol. 2009 Dec;34:586-662.**
2. **Lawless CE, Best TM.Electrocardiograms in athletes: interpretation and diagnostic accuracy. Med Sci Sports Exerc. 2008 May;40:787-798.**
3. **Wever-Pinzon OE, Myerson M, Sherrid MV.**
4. **Sudden cardiac death in young competitive athletes due to genetic cardiac abnormalities. Anadolu Kardiyol Derg. 2009 Dec;9 Suppl 2:17-23.**

I) Benign', common ECG features 65% result from adaptation to exercise[1;2;3]

1. Sinus bradycardia (common)
2. First-degree atrioventricular block (common
3. Early Repolarization Pattern or Variant (common)
4. Incomplete Right Bundle Branch (IRBBB) (common)
5. Isolated signs of LVH (common)

II) ‘Suspected', uncommon in 23%. which may occur due to organic heart disease

1. Left atrial enlargement (LAE)
2. Pathological Q waves
3. Pathological QRS axis deviation the most frequent was left posterior fascicular block[2], present in 10% of those examined;
4. Complete Bundle Branch Block
5. Ventricular arrhythmia
6. Inverted T waves in two or more consecutive precordial leads.

1. **Corrado D, McKenna WJ. Appropriate interpretation of the athlete's electrocardiogram saves lives as well as money. Eur Heart J. 2007 Aug; 28:1920-1922.**
2. **Swiatowiec A, Król W, Kuch M, Braksator W, Krysztofiak H, Dłużniewski M, et al. Analysis of 12-lead electrocardiogram in top competitive professional athletes in the light of recent guidelines. Kardiol Pol. 2009 Oct;67:1095-10102.**
3. **Corrado D, Pelliccia A, Heidbuchel H, Sharma S, Link M, Basso C, et al;. Recommendations for interpretation of 12-lead electrocardiogram in the athlete.Section of Sports Cardiology, European Association of Cardiovascular Prevention and Rehabilitation; Working Group of Myocardial and Pericardial Disease, European Society of Cardiology. Eur Heart J. 2010 Jan;31:243-259.**

Electrocardiographic alterations
Rhythm and Heart Rate

Sinus bradycardia is observed in more than 50% of the cases. HR of 30 to 40 bpm in rest are not rare. In highly trained athletes, there are descriptions of HR of 25 bpm.
Sinus arrhythmia, mostly related to respiration, phasic or respiratory sinus arrhythmia is present in 60% of cases in the athletes population. In no-athlete only 2.4%.
Sinus arrest, with ectopic escape beat or rhythm, or resumption of sinus rhythm
Wandering atrial pacemaker
Junctional rhythm is present in 0.31% (in the general population only in 0.02%).
Coronary sinus rhythm
Long sinus pauses are frequent among athletes (> 2 seconds)

P waves in athlete heart

Increase P wave amplitude and notching. P wave dispersion was increased in elite woman basketball players as compared with healthy sedentary subjects. P wave dispersion was correlated with heart rate, body height, body weight and body surface area[1].

1. **Metin G, Yildiz M, Bayraktar B, Yucesir I, Kasap H, Cakar L. Assessment of the p wave dispersion and duration in elite women basketball players. Indian Pacing Electrophysiol J. 2010 Jan 7;10:10-20.**

Atrioventricular block in athlete

1. First degree atrioventricular block: is observed in 5% and 30% of cases (in non athletes, 0.65%). When the PR interval does not reach the value as a criterion for 1st degree AV block, it is relatively prolonged. The PR interval normalizes or even gets smaller after exercise. Lone atrial fibrillation seems to be more common in endurance-trained male athletes than in men in the general population. Long PR interval, bradycardia and left atrial enlargement seem to be important risk factors for lone atrial fibrillation among long-term endurance cross-country skiers(1).
2. Second degree atrioventricular (AV) block, Möbitz type I, or Wenckebach-type : it is observed in ≈10% of cases (in non athletes < 1 in 30,000 or 0.003%), and it disappears invariably during exercise and with atropine
3. Atrioventricular dissociation.
4. Higher grade AV blocks have rarely been observed in athletes; they may be indicative of underlying heart disease and are an indication for further evaluation.
5. Complete or 3rd degree AV block: 5 each 12,000 athletes.

1. Grimsmo J, Grundvold I, Maehlum S, Arnesen HHigh prevalence of atrial fibrillation in long-term endurance cross-country skiers: echocardiographic findings and possible predictors - a 28-30 years follow-up study.Eur J Cardiovasc Prev Rehabil. 2010 Feb;17:100-105.

QRS modifications

•The QRS frontal axis is generally between 0 + 90° and is on average normal.
•QRS axis: tendency to vertical position.

RV1+SV5 >10.5 mm between 18% and 69% of the cases. RVH manifests by a diastolic pattern translated by minimal degrees of IRBBB. The IRBBB is observed in 15% of athletes.

Evidence of RVH. for example, increased RV1 + SV5.

Absence of progression of increase of voltage of r or R wave with QR pattern from V1 to V3: pattern of pseudo infarction in anterior wall.

Pattern of Complete RBBB: observed in 13.5% of the cases

The Sokolow-Lyon voltage index criteria[1]

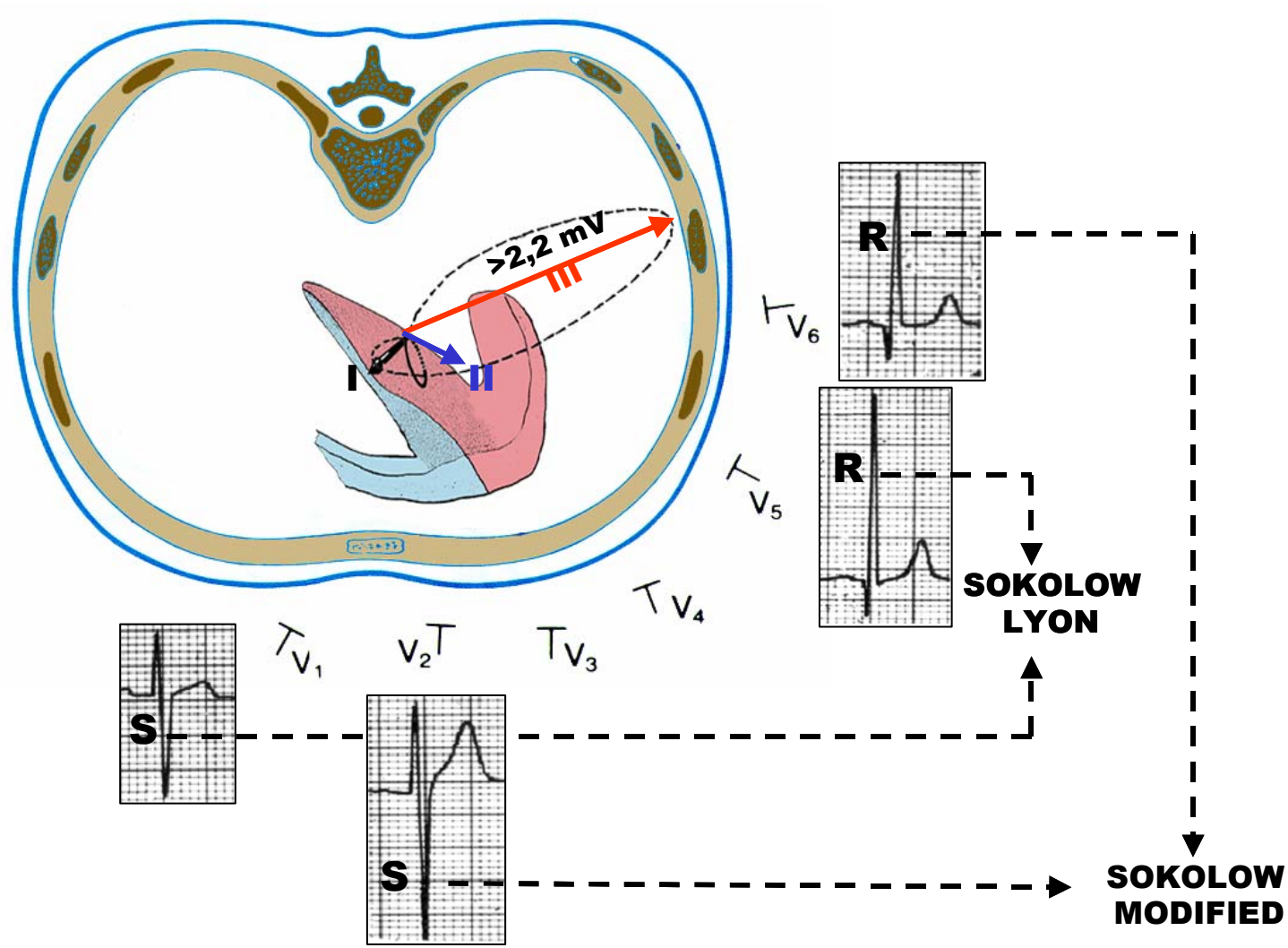
•This is more frequent voltage criteria for LVH. The increased QRS voltage is considered to be a specific ECG sign of LVH, and it is expected that the QRS voltage reflects the increase in LV mass (LVM). However, the increased QRS voltage is only one of QRS patterns observed in patients with LVH.

•Increase Sokolow -Lyon index = SV_1+ RV_5 or RV_6 ≥ 35mm(or 38mm for epidemiologic studies) in adult older than 30 years old, or ≥ 40mm between 20 to 30 years old (Sokolow-Rapaport); or ≥ 60mm between 16 and 20 years old; or >65mm between 11 and 16 years old. Sensitivity: 25%. Specificity: 95%. Consequently, ECG criteria for LVH have been limited by low sensitivity at acceptable levels of specificity. Studies have demonstrated that body mass index (BMI) is associated with decreased sensitivity of ECG LVH voltage criteria. People with higher BMI, have lower ECG amplitudes. Therefore, the Sokolow-Lyon voltage criteria may underestimate the presence of LVH for subjects with higher BMI, which is not the case for the Cornell voltage. Computerized ECG for the diagnosis of LVH based on Sokolow-Lyon voltage criteria should incorporate the BMI factor[2]. Aditionally, The finding of LVH on ECG is not very reproducible during serial measurements on the same person during a single 24-hour observation period[3]. The Sokolow-Lyon and Cornell voltage criteria are more sensitive in African-origin populations. The Sokolow-Lyon criteria is less specific for LVH than Cornell voltage in people of African origin. The evidence favors the Cornell criteria in research and service contexts involving African-origin and white populations[4].

1. **Sokolow M, Lyon TP. The ventricular complex in left ventricular hypertrophy as obtained by unipolar precordial and limb leads. Am Heart J. 1949; 37: 161-186.**
2. **Abächerli R, Zhou L, Schmid JJ, et al.Correlation relationship assessment between left ventricular hypertrophy voltage criteria and body mass index in 41,806 Swiss conscripts. Ann Noninvasive Electrocardiol. 2009 Oct;14:381-388.**
3. **Shoenberger JM, Voskanian S, Johnson S, et al. Left ventricular hypertrophy may be transient in the emergency department.West J Emerg Med. 2009 Aug;10:140-143.**
4. **Vanezis AP, Bhopal R. Validity of electrocardiographic classification of left ventricular hypertrophy across adult ethnic groups with echocardiography as a standard.J Electrocardiol. 2008 Sep-Oct;41:404-412.**

SOKOLOW INDEX MODIFIED FOR LVH

S of V_2 + R of V_5 or $V_6 \geq 35$ mm (or 38mm)



Sokolow-Lyon product (SokP)

The voltage-duration product criteria suggestively detected ECG-LVH and its respective changes better than voltage criteria.

Cornell voltage index criterion OR Casale Criteria for LVH

Cornell = sum of SV3 + R wave in aVL ≥ 2.3 mV for males and 1.9 mV for females[1;2].

Gubner-Ungerleider index voltage

Gubner index = sum of R wave in I and S wave in III > 2.5 mV)

1. Casale PN, et al. Electrocardiographic detection of left ventricular hypertrophy: development and prospective validation of improved criteria. J Am Coll Cardiol. 1985; 6: 572–580.
2. Rodrigues SL, D'Angelo L, Pereira AC, et al. Revision of the Sokolow-Lyon-Rappaport and cornell voltage criteria for left ventricular hypertrophyArq Bras Cardiol. 2008 Jan;90:46-53.

Cornell voltage-duration product or The CorP criterion

The product of QRS voltage and duration (e.g. Cornell voltage-duration product; CorP), is a good marker of LVH

ECG is a useful, efficient, and highly reproducible method for the diagnosis of LVH in hemodialysis patients. In this population, the Cornell-product proved to be the most reliable criterion for the detection of LVH[1].

Cornell product[2;3] = product of sum of R aVL + and S in V_3 + 6 mm in women and QRS duration (CP ≥ 2440 mm x ms) The primary endpoint for detection of LVH is a CorP >2.440 mm x ms on ECG recordings,

The Cornell product is a useful ECG marker, reflecting not only left ventricular mass but also LV geometry and diastolic function in Japanese hypertensive patients[4].

Reduction in Cor P ECG LVH during antihypertensive therapy is associated with fewer hospitalizations for HF, independent of blood pressure lowering, treatment method, and other risk factors for HF[5].

1. **Costa F de A, Rivera IR, de Vasconcelos ML, Costa AF, Póvoa RM, Bombig MT, Luna Filho B, de Lima VC.Electrocardiography in the diagnosis of ventricular hypertrophy in patients with chronic renal disease. Arq Bras Cardiol. 2009 Oct;93:380-6, 373-379.**
2. **Molloy TJ, Okin PM, Devereux RB, et al. Electrocardiographic detection of left ventricular hypertrophy by the simple QRS voltage-duration product. J Am Coll Cardiol. 1992;20(5):1180-1186.**
3. **Okin PM, et al. Electrocardiographic identification of increased left ventricular mass by simple voltage-duration products. J Am Coll Cardiol. 1995; 25: 417–423.**
4. **Shirai T, et al.Evaluation of hypertensive cardiac abnormalities using the Cornell product.Circ J. 2007;71:731-735.**
5. **Okin PM, Devereux RB, Harris KE, Jern S, Kjeldsen SE, Julius S, Edelman JM, Dahlöf B; LIFE Study Investigators. Regression of electrocardiographic left ventricular hypertrophy is associated with less hospitalization for heart failure in hypertensive patients. Ann Intern Med. 2007 Sep 4;147(5):311-319.**

Romhilt-Estes score

By calibration with CMR, a wide range of predictive values was found for the various ECG criteria for LVH with the most favorable results for the Romhilt-Estes score. As ECG correlate for concentric LVH as compared with eccentric LVH, a shorter intrinsicoid deflection and a significant ST-segment and T-wave depression in the anterolateral leads was noted.

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 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 abnormalities Without digitalis With digitalis	3 points 1 point
Left atrial abnormality Negative area under P wave in Lead V1 ≥ 1 mm2 (1 box)	3 points
Left axis deviation	2 points
QRS duration > 90 ms	1 point
Intrinsicoid deflection V5, V6 ≥50ms	1point

Romhilt DW, Estes EH Jr. A point-score system for the ECG diagnosis of left ventricular hypertrophy. Am Heart J. 1968; 75:752-758.

The Perugia-score system

- The Perugia score[1] carried the highest population-attributable risk for cardiovascular morbidity and mortality compared with classic methods for detection of LVH. Traditional interpretation of standard electrocardiography maintains an important role for cardiovascular risk stratification in essential hypertension. ECG-LVH. Perugia Score Positivity of one or more of the following criteria:
  - SV3+ RaVL >2.4mV (men) or >2.0mV (women);
  - Left ventricular strain pattern;
  - Romhil-Estes score of five or more points.
- The diagnostic criteria of the Perugia Score, the newest scoring system for electrocardiogram-diagnosed LVH. However, the Perugia score has a low sensitivity. They showed that the prevalence of LVH in the hypertensive population is highest using the Perugia score, followed by the Sokolow-Lyon Voltage criteria.

1) Verdecchia P, et al. Prognostic value of a new electrocardiographic method for diagnosis of left ventricular hypertrophy in essential hypertension. J Am Coll Cardiol. 1998; 31:383-390.

Erice et al [1] examined the strongest predictor within ECG voltage criteria for LVH in HCM to be applied in cardiovascular examination of young people. The ECGs of 36 healthy individuals with high voltages, mimicking HCM (i.e., false-positive), were statistically compared with those of 30 subjects with an ECG diagnosis of HCM.

The most striking ECG voltages observed in HCM patients were those included in leads I, aVL (R wave) and V3 (S wave) typically present in the Cornell, Gubner and Lewis voltage criteria. In a stepwise logistic regression analysis model, these indices were the most significant predictors of HCM.

The combination of Cornell (R aVL+SV_3>2.8 mV in men and>2.0 mV in women) with Lewis (RI+SIII-RIII-SI>1.7 mV) or Gubner-Ungerleider (RI+SIII>2.5 mV) indices displayed the highest net sensitivity (80.0% and 76.7%, respectively) while retaining excellent specificity (88.9% and 91.6%, respectively).

The authors concluded that the combination of the Cornell, Lewis or Gubner voltage criteria showed the greatest net sensitivity and specificity for the LVH diagnosis of HCM in a cardiovascular examination conducted in young people.

1. **Erice B, Romero C, Andériz M, Gorostiaga E, Izquierdo M, Ibáñez J. Diagnostic value of different electrocardiographic voltage criteria for hypertrophic cardiomyopathy in young people. Scand J Med Sci Sports. 2009 Jun;19: 356-363.**

Repolarization abnormalities

Early Repolarization Variant (ERV) or Pattern (ERP) It is described in four patterns:

- J point and ST segment elevation followed by peaked T wave from V4 to V6 and in the inferior wall (2.4% to 44%)
- J point and ST segment depression (rare)
- J point and ST segment elevation followed by inverted T wave
- Disappearance of ST segment elevation after exercise.

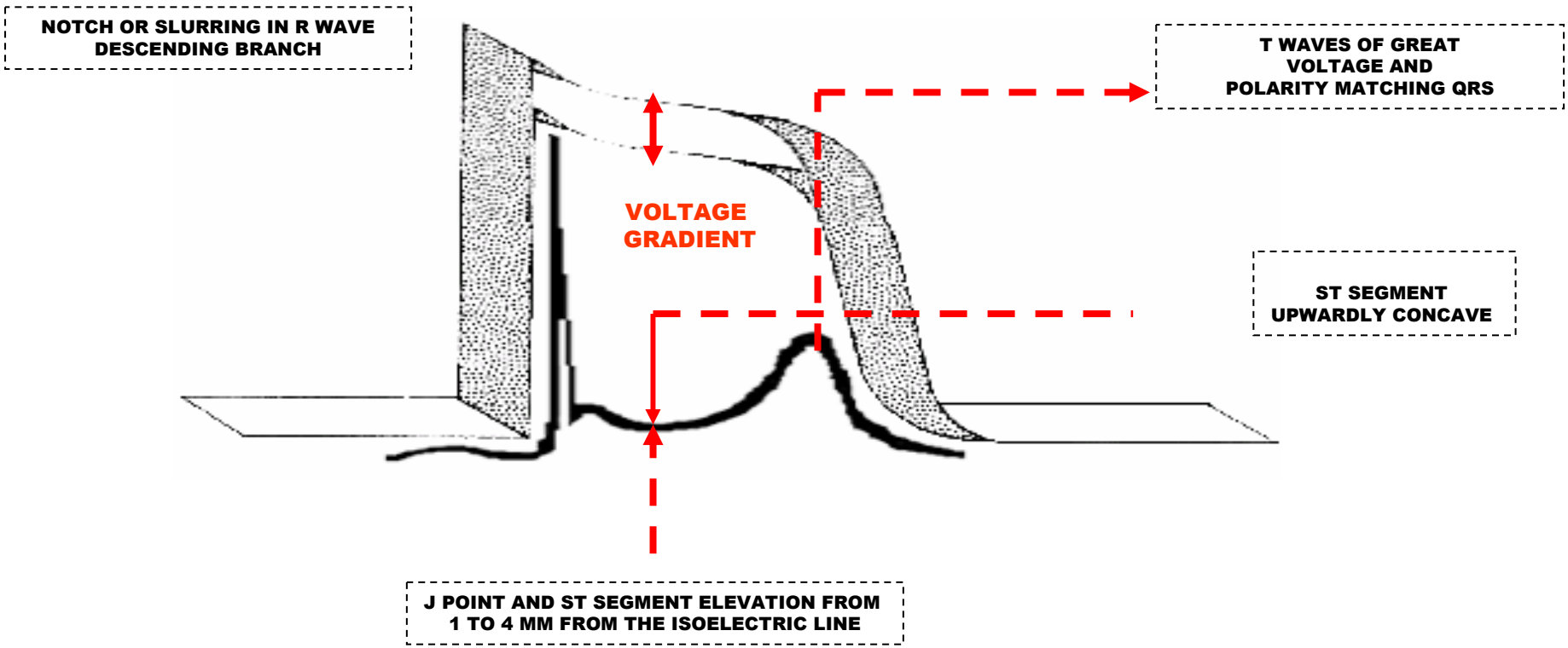
ST segment

- J point elevation
- ST segment elevation
- ST segment depression

T wave

- Tall and peaked T waves
- Juvenile pattern of T wave
- Notched T waves
- Low amplitude or isoelectric T waves
- Negative or biphasic T waves from V1 to V3 and/or in the inferior wall
- Biphasic T waves
- Biphasic T waves with terminal negativity
- Inverted and asymmetrical T wave in I, aVL, V5 and V6, secondary to physiologic LVE
- Frequent “normalization” of T wave before strain. This type of response is not observed in HCM or in CHD. Characteristic reversion of ECG “alterations” in cases of interruption of competitive activity. Myocardial perfusion imaging associated to exercise stress test always negative.

In early repolarization, there is a voltage gradient, however, no dispersion of duration of action potentials in ventricular wall thickness. For this reason, these patients showed ST segment elevation with no tendency to develop arrhythmias.



ECG CRITERIA THAT SUGGEST EARLY REPOLARIZATION PATTERN (ERP)

✓ HR: sinus bradicardia is frequent;

✓ Axes of QRS, ST segment and T wave, are oriented in the same direction in the FP;

✓ Deep and narrow Q waves followed by R wave of great voltage in left precordial leads;

✓ Notch or slurring of R wave descending branch;

✓ Transition area in precordial leads of sudden occurrence;

ECG CRITERIA THAT SUGGEST EARLY REPOLARIZATION PATTERN (ERP)

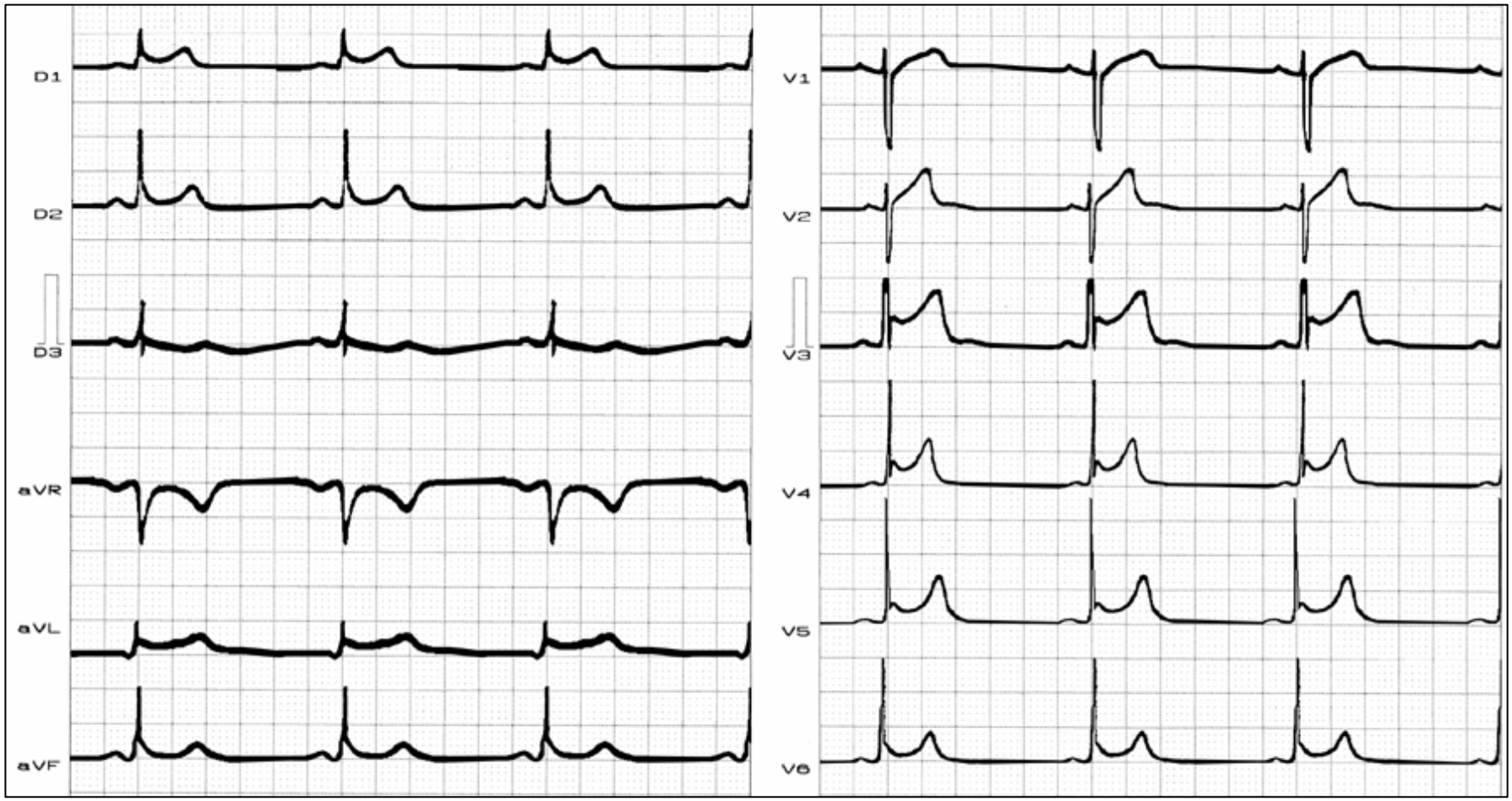
✓ J point and ST segment elevation, usually < 2 mm (exceptionally it may be > 5 mm) of superior concavity in middle and/or left precordial leads and possibly in inferior leads;

✓ Possible reduction in J point and ST segment elevation by sympathetic action and sympathomimetic drugs;

✓ Absence of reciprocal or mirror image (exception in VR lead);

✓ Symmetrical T waves, with great width and polarity matching QRS;

Name: DAS **Age:** 24y **Sex:** Male **Race:** Black **Weight:** 82 kg
Height: 1.91 m **Biotype:** Athletic **Profession:** professional basketball player



ECG DIAGNOSIS: sinus bradycardia, (HR 50 bpm). J point and ST segment with elevation > 4 mm in precordial leads from V_3-v_5 of superior concavity. Notch or slurring of terminal portion of the QRS complex (J point). ST segment elevation > 4mm in precordial leads V_3, V_4 and V_5.

CONCLUSION: sinus bradycardia, early repolarization syndrome.

SUMMARY OF THE ECG ELEMENTS COMMON IN ATHLETES

1) Sinus bradycardia.
2) Sinus arrhythmia.
3) P wave with notches and of greater voltage.
4) First degree AV block: 6% to 36%.
5) 2nd degree AV block, Wenckebach type: Mobitz Type I (0.125% to 10%).
6) IRBBB or end conduction delay.
7) Voltage or axis criterion for RVE.
8) Voltage criterion for LVE.
9) J point and ST segment elevation or depression.
10) QT interval in the superior borderline of normality.
11) T wave of increased voltage, peaked and inverted.
12) Atrial fibrillation and flutter[1].
13) Junctional rhythm.

1) Furlanello, et al. J Cardiovasc Electrophysiol 1998;9 (Suppl 8); S63-S68.

ARRHYTHMIAS IN THE HEARTS OF ATHLETES AND COMPARATIVE INCIDENCE WITH THE GENERAL POPULATION

ARRHYTHMIA	GENERAL POPULATION	ATHLETES
Sinus Bradycardia	23.7	50-85
Sinus Arrhythmia	2.4-20	13.5-69
Atrial Variable Pacemaker	NOT AVAILABLE	7.4-19
1st degree AV block	0.65	6-33

ARRHYTHMIAS IN THE HEARTS OF ATHLETES AND COMPARATIVE INCIDENCE WITH THE GENERAL POPULATION

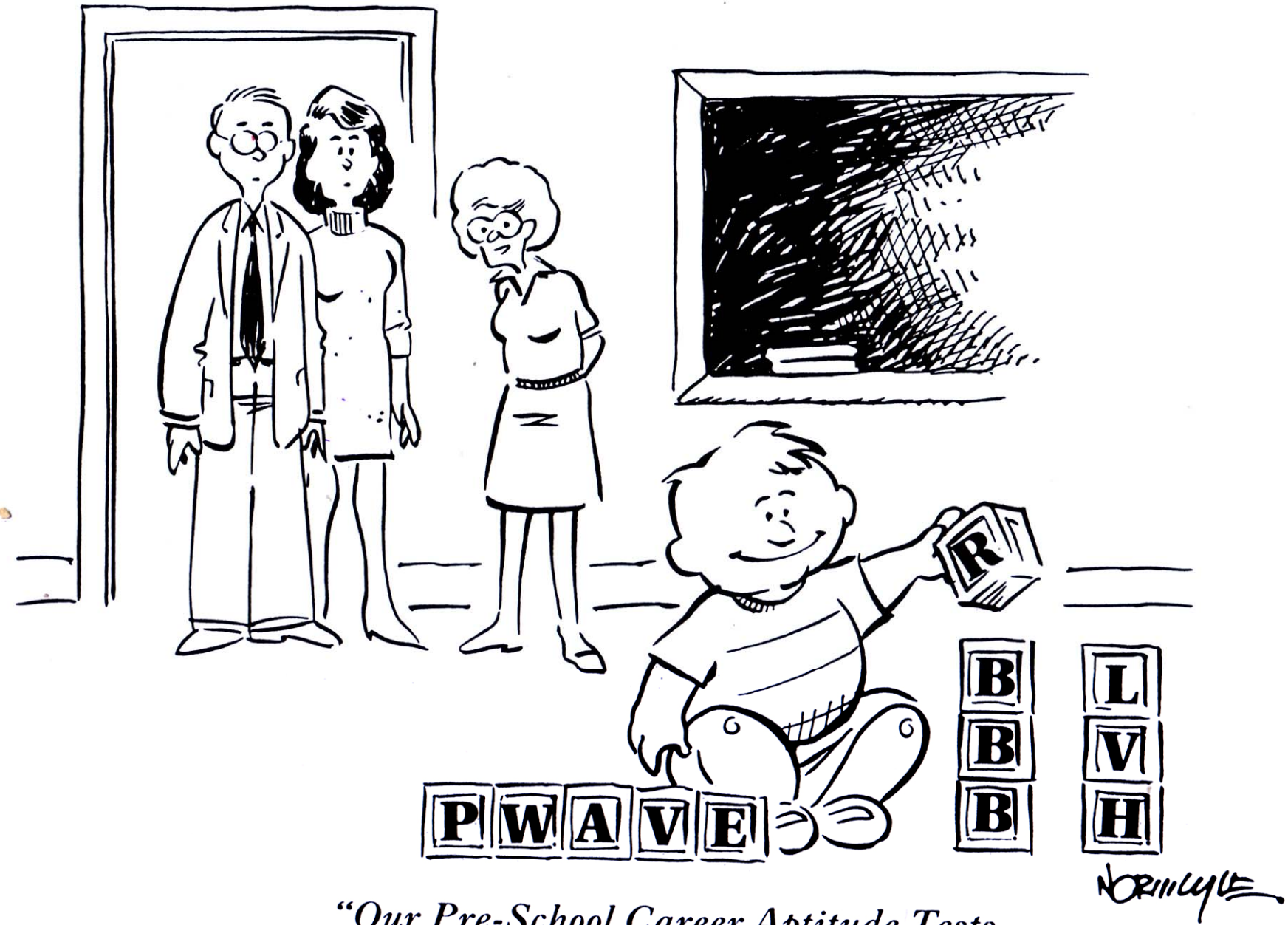
ARRHYTHMIA	GENERAL POPULATION	ATHLETES
2nd degree AV block		
Mobitz Type 1	0.003	0.125-10
Mobitz Type II	0.003	NOT REPORTED
3rd degree AV block	0.0002	0.017
Junctional Rhythm	0.06	0.31-7.0

The ECG: key points

High intensity dynamic endurance sports are usually associated with ECG rhythm and conduction abnormalities, which result from the lower intrinsic heart rate and/or changes in parasympathetic and sympathetic tone.
Structural cardiac adaptations induce morphological changes of the QRS complex
Repolarization abnormalities result from structural changes and from parasympathetic predominance
Several ECG changes may mimic cardiac disease

In U.S. series of professionals football players ECG abnormalities are seen in up to 55%, including ST abnormalities in up to 15% and intraventricular conduction defects in up to 20%, with increased prevalence of abnormalities in Blacks compared to Caucasian players. Black athletes exhibit greater LV wall thickness and cavity size compared with sedentary black and white individuals. Black athletes hace greater LV wall thickness compared with white athletes. Black athletes exhibit LV wall thickness≥15 mm compared with none of the white athletes. Black athletes with LVH displayed an enlarged LV cavity and normal diastolic function.
Black athletes develop a greater magnitude of LVH compared with white athletes[1].

1. **Basavarajaiah S, Boraita A, Whyte G, Wilson M, Carby L, Shah A, Sharma S. Ethnic differences in left ventricular remodeling in highly-trained athletes relevance to differentiating physiologic left ventricular hypertrophy from hypertrophic cardiomyopathy. J Am Coll Cardiol. 2008 Jun 10;51:2256-2262.**



"Our Pre-School Career Aptitude Tests indicate little Mark will make a very fine cardiologist".

Ambulatory electrocardiography

The 12 lead ECG lasts only a few seconds, so that 24 hour ambulatory electrocardiography may give a better insight into the occurrence of rhythm and conduction alterations in athletes. Viitasalo and colleagues18 compared 35 highly trained male endurance athletes with 35 non-athletic matched controls. Heart rate was lower in athletes throughout the day and night. As shown in table 3 the lowest nocturnal heart rate ranged from 24–48 beats/min in the athletes and from 33–63 beats/min in the controls. Sinus pauses exceeding 2.0 s occurred in 37.1% and 5.7%, respectively, with the longest PP intervals of 2.76 s and 2.6 s. First degree atrioventricular block and second degree Möbitz type 1 block occ urred more frequently in athletes. Atrioventricular dissociation and Möbitz type II block were not observed in controls but did occur in athletes. Disturbances were more prevalent during the night than during the day. The frequency of ventricular arrhythmias did not differ between athletes and controls.

1. **Viitasolo MT, Kala R, Eisalo A. Ambulatory electrocardiographic recording in endurance athletes. Br Heart J 1982;47:213–220.**

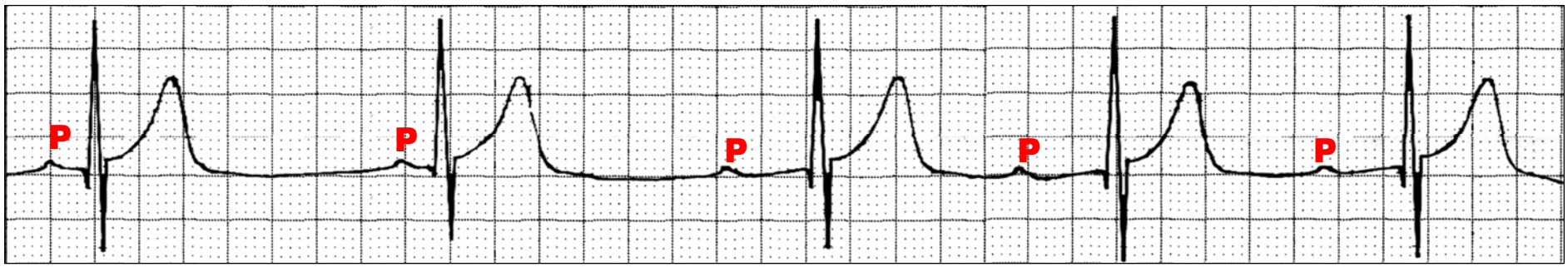
Heart rate and frequency of cardiac events on ambulatory monitoring of athletes and non-athletes

	Controls	Athletes	p Values
Number	35	35	
Heart rate (beats/min)			
Lowest nocturnal	45 (33–63)	38 (24–48)	<0.001
Sinus pause			
PP >2.0 s	5.7%	37.1%	<0.01
Longest PP (s)	2.60	2.76	–
Atrioventricular block			
First degree (PR >0.22 s)	14.3 %	37.1%	<0.05
Longest PR interval (s)	0.40	0.54	–
Second degree			
Möbitz type I (Wenckebach type)	5.7%	22.9%	<0.05
Möbitz type II	0%	8.6%	
Atrioventricular dissociation	0%	20%	

1. ***Fagard R. Athlete's heart. Heart.*** **2003 December; 89: 1455–1461**.

HOLTER RECORDING
1ST DEGREE AV BLOCK

Name: B . C. **Sex:** Male **Age:** 22 **Race:** Black **Weight:** 74 Kg.
Height: 1.82 m **Biotype:** Athletic **Date:** 01/04/2002
Time: 2:50:12 AM Patient sleeping. **Profession:** Marathon runner



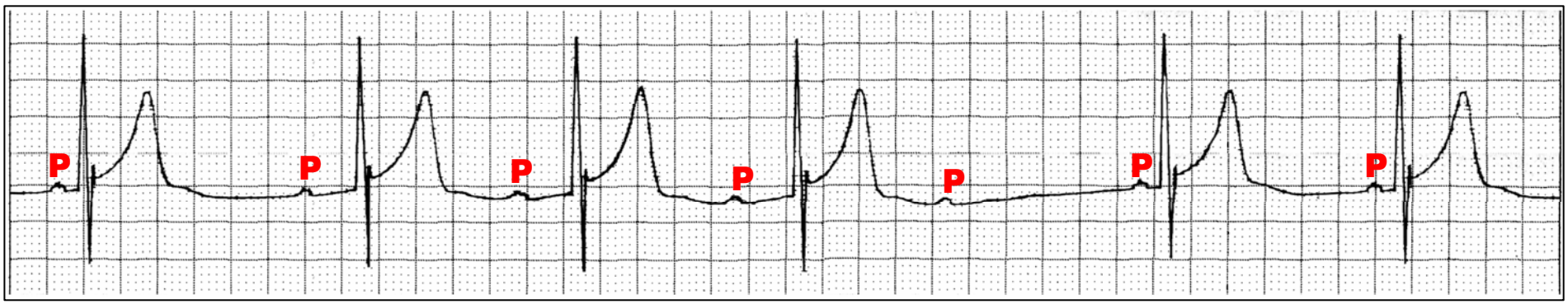
Heart rate of 38 bpm.
1st degree AV block usually observed for a few seconds, as in this case, where it is present only in the three last beats.

1st degree AV block is observed in average between 10% and 33% of athletes (1), generally very briefly. In non-athletes it is around 0.65%.

1) Smith WG, et al. Br Heart J 1964:469-476.

HOLTER RECORDING

Name: A . S. **Sex:** Male **Age:** 26 **Race:** Black **Weight:** 64 Kg.
Height: 1.68 m **Biotype:** Athletic **Date:** 05/01/2003
Time: 3:42:30 AM Patient sleeping. **Profession:** long distance runner



Gradual prolongation of PR interval until the 5th P wave is not conducted: 2nd degree AV block; Wenckebach or Mobitz Type I.

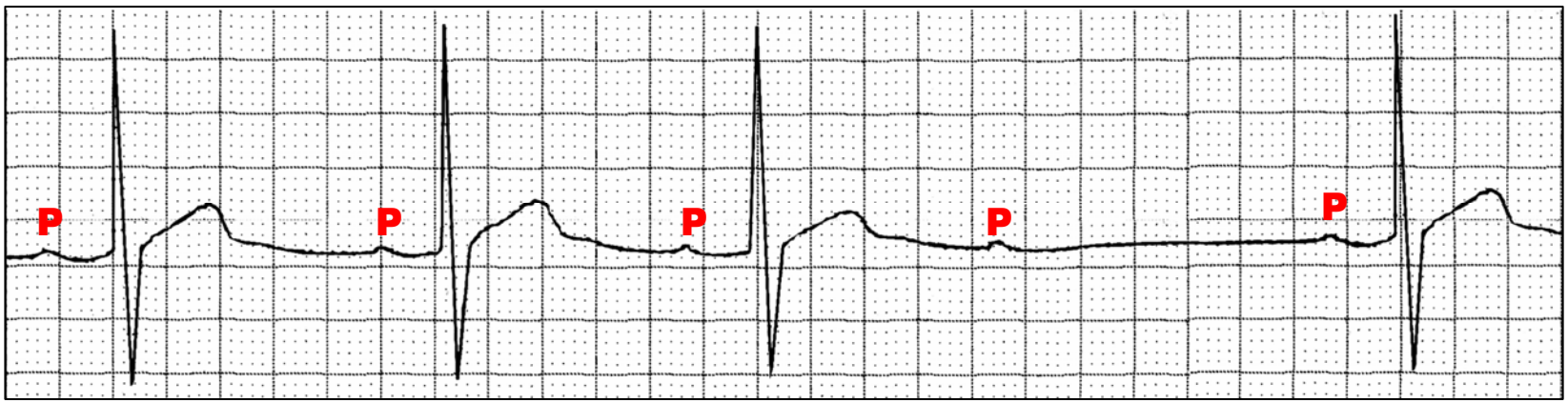
This modality of dromotropic disorder is observed in more than a 20% of elite athletes (1). In the general population, 2nd degree AV block Type I & II is observed and 1 each 30,000 people or 0.003 %

1) Viitasalo MT, et al. Br Heart J. 1982;47:213-220.

HOLTER RECORDING

2ND DEGREE AV BLOCK, MOBITZ TYPE II WITH NARROW QRS

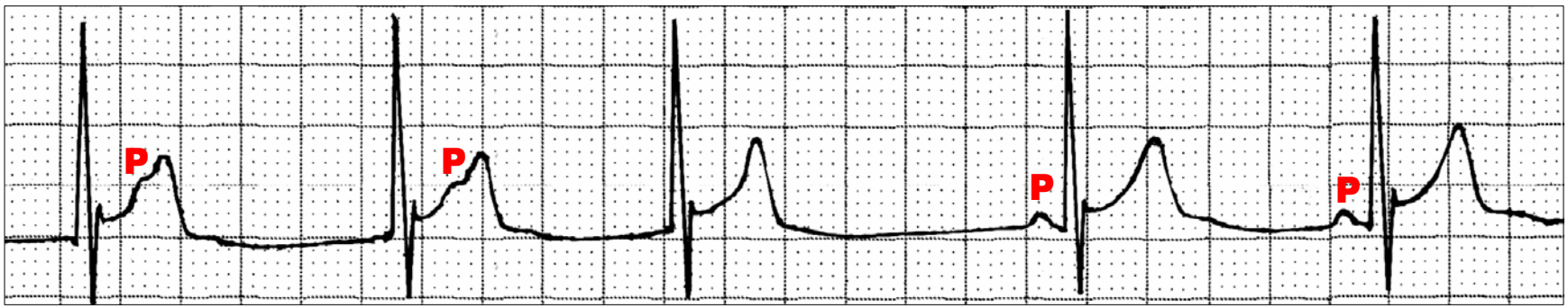
Name: E . J. **Sex:** Male **Age:** 26 **Race:** White **Weight:** 70 Kg.
Height: 1.72 m **Biotype:** Athletic **Date:** 25/01/2001
Time: 1:52:10 AM Patient sleeping **Profession:** Long distance runner



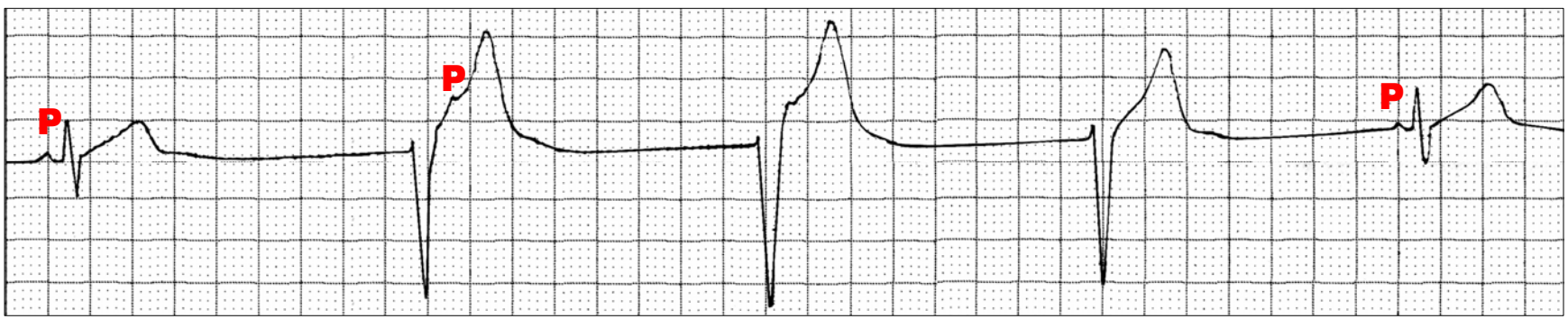
PR interval remains constant until a P wave is not conducted. This type of block is observed in 7% of the cases in athletes of enduro. Fixed or constant PR interval: it does not exist, progressive prolongation of PR, with the block occurring suddenly. In general, 2nd degree AV block type II with narrow QRS is observed in 35% of the cases and in the remaining 65%, the QRS is long.

HOLTER RECORDING

ATRIOVENTRICULAR DISSOCIATION (DISSOCIATION BY INTERFERENCE) WITH JUNCTIONAL SCAPE RHYTHM



ATRIOVENTRICULAR DISSOCIATION (DISSOCIATION BY INTERFERENCE) WITH SCAPE VENTRICULAR RHYTHM



Echocardiographic changes in Athletes

In competitive athletes are frequent: Myocardial remodeling, Leff ventricular dilatation, wall hypertrophy 14mm in Italian and British and 16mm in USA in American football players(1)

Differentiation between physiologic hypertrophy the 'mild' from HCM

	Physiologic hypertrophy	HCM
LV dilatation	Characteristic eccentric biventricular hypertrophy LV cavity > 55 mm	Absent. Reduced LV-diameter. LV cavity <45mm
Exercise tolerance	Excellent	Poor
The 'grey zone'	(LV wall thickness of 13-16 mm) <15mm	(LV wall thickness of 13-16 mm)
Diastolic function:	Normal with even increased early diastolic filling	Diastolic dysfunction mostly relaxation disturbances.
Left Atrial Enlargement	No	Yes
Female Gender	Negative	Positive
Decrease Hypertrophy with Less Physical Training or stop	Yes	No
Positive family history or provable Genetic Mutation	No	Possible.

If the diagnosis could not be stated using ECHO, methods like cardiac-MRI, metabolic exercise testing, histological studies of endomyocardial biopsies and genetic testing can provide further information.

1. **Chelliah RK, Senior R. Pathological and physiological left ventricular hypertrophy: echocardiography for differentiation. Future Cardiol. 2009 Sep;5:495-502.**
2. **Lauschke J, Maisch B.Athlete's heart or hypertrophic cardiomyopathy? Clin Res Cardiol. 2009 Feb;98:80-88.**

In 1975, Morganroth[1] and colleagues hypothesized that the cardiac morphological adaptation observed in athletes corresponded with the nature of the hemodynamic stimulus imposed on the ventricles during repeated exercise. Endurance training purportedly leads to an **eccentric** form of cardiac hypertrophy, characterized by increased LV cavity dimension, and thus LV mass (LVM), as a consequence of prolonged repetitive volume overload. In contrast, strength training is supposedly associated with a **concentric** form of hypertrophy where increased ventricular wall thickness, with no change in cavity size, underpins the elevated LVM as a consequence of the pressure overload produced during strenuous resistive exercise.

The 'Morganroth hypothesis' has been broadly adopted in the scientific and medical literature, partly as a consequence of a large body of cross-sectional evidence suggesting that endurance athletes have greater cavity dimensions than control subjects or resistance athletes. However, in conflict with the 'Morganroth hypothesis', several studies suggest that LV wall thickness is increased more in endurance-, than strength-trained athletes and others have reported no morphological changes in resistance-trained athletes. Such controversial data may reflect variability in the training stimuli, with little obvious attempt to quantify these issues in previous research. Further reflection on the 'Morganroth hypothesis' may also be pertinent as more sensitive technologies, such as MRI, are now being employed for the assessment of cardiac morphology. Finally, the process of scaling (or normalizing) cardiac size for between-subject differences in body size and composition has further complicated the description and understanding of cardiac morphology in athletes. Specifically, it is possible that the increased LVM observed in some athletes may merely reflect a 'larger than normal' body size. These considerations emphasize the limitations of the predominance of cross-sectional comparisons in the available literature, which assume that differences between groups are due to a training effect per se rather than other between-subject differences. The small number of longitudinal training studies undertaken in athletes suggest that individuals with athlete's heart can exhibit further cardiac adaptation in response to training resumption.

1. **Morganroth J, Maron BJ, Henry WL, Epstein SE. Comparative left ventricular dimensions in trained athletes. Ann Intern Med. 1975 Apr;82:521-524.**

Longitudinal training studies undertaken in previously sedentary subjects generally indicate that exercise results in enlargement of LV cavity size, increases in wall thickness or LVM following training. However, there are currently limited longitudinal data available to comment on the effects of different modalities of exercise training on LV cavity dimension and wall thickness.

In summary, significant caveats related to cross-sectional literature, the relative insensitivity of ECHO measurements and the paucity of evidence from longitudinal exercise training studies, warrant ongoing research to verify the 'Morganroth hypothesis'.

Advanced cardiac imaging, using cardiac MRI and multidetector computed tomography (CT), is increasingly used in the work-up of athletes with suspected abnormalities on screening.

Both imaging modalities produce highly accurate and reproducible structural and functional cardiac information. Cardiac MRI has the advantage of imaging without radiation exposure or the use of iodine-containing contrast agents, but is sometimes not possible due to claustrophobia or other contraindications. Although cardiac MRI can rule out coronary artery anomalies, multidetector CT is superior to cardiac MRI for visualizing the full extent of the coronary arteries and CAD. For patients, 35 years old, cardiac MRI is the first option after initial Echo for further assessment of **cardiomyopathies, myocarditis and coronary anomalies**, which are major causes of SCD in young athletes. For athletes >35 years of age the most common cause of SCD is CAD, whereby cardiovascular screening requires further diagnostic modalities and may include multidetector CT[2].

1. **Naylor LH, George K, O'Driscoll G, Green DJ.The athlete's heart: a contemporary appraisal of the 'Morganroth hypothesis'.Sports Med. 2008;38:69-90.**
2. **Prakken NH, Velthuis BK, Cramer MJ, Mosterd A. Advances in cardiac imaging: the role of magnetic resonance imaging and computed tomography in identifying athletes at risk. Br J Sports Med. 2009 Sep;43:677-684.**

STRUCTURE OF PHYSIOLOGICAL VENTRICULAR HYPERTROPHY ATHLETE'S HEART

"Possibly, the Differentiation Between Physiological Ventricular Hypertrophy of Athletes and the Pathological One (Ventricular Remodeling) May Become A Challenge".

DIFFERENCES BETWEEN PHYSIOLOGICAL VENTRICULAR HYPERTROPHY OF THE ATHLETE AND THE PATHOLOGICAL ONE (VENTRICULAR REMODELING)

	PHYSIOLOGICAL VENTRICULAR HYPERTROPHY	PATHOLOGICAL VENTRICULAR HYPERTROPHY VENTRICULAR REMODELING
Location:	Symmetrical, however, it may be asymmetrical.	Asymmetrical, however, it may be symmetrical.
Relative Ischemia:	Absent.	Present.
Myocitic/Non-myocitic Component Relationship:	Maintained.	Loss of balance in favor of the non-myocitic component (fibrosis).
Energetic Cycle:	Aerobiosis.	Anaerobiosis.
Renin-angiotensin-aldosterone Mechanism	Normal.	Increased.
Norepinephrine	Normal.	Increased.

DIFFERENCES BETWEEN PHYSIOLOGICAL VENTRICULAR HYPERTROPHY OF THE ATHLETE AND THE PATHOLOGICAL ONE (VENTRICULAR REMODELING)

	PHYSIOLOGICAL VENTRICULAR HYPERTROPHY	PATHOLOGICAL VENTRICULAR HYPERTROPHY VENTRICULAR REMODELING
Atrial Natriuretic Peptide	Normal.	It may be increased.
Pump Function	Normal.	Depressed.
Heart Rate:	Tendency to sinus bradyarrhythmia by vagotony.	Frequent tachycarrhythmia and sympathotony.
LV Pd2:	Normal.	Increased.

DIFFERENCES BETWEEN PHYSIOLOGICAL VENTRICULAR HYPERTROPHY OF THE ATHLETE AND THE PATHOLOGICAL ONE (VENTRICULAR REMODELING)

	PHYSIOLOGICAL VENTRICULAR HYPERTROPHY	PATHOLOGICAL VENTRICULAR HYPERTROPHY VENTRICULAR REMODELING
Pulmonary Artery Pressure And Central Venous Pressure:	Normal.	It may be increased.
ANS:	Parasympathetic predominance.	Sympathetic predominance.
Curve Of Dissociation of Hb:	Deviation to the right.	Deviation to the left.
Echocardiogram:	Proportional growth between the diameter and the thickness of walls. Normal LA.	Loss of walls thickness/diameter ratio. Increased LA.

DIFFERENCES BETWEEN PHYSIOLOGICAL VENTRICULAR HYPERTROPHY OF ATHLETES AND HYPERTROPHIC CARDIOMYOPATHY (HCM) WHEN BOTH PRESENT WALL THICKNESS BETWEEN 13 mm & 15 mm

The concentric or symmetrical form of HCM (5%), may be confused with the athlete's heart with physiological hypertrophy of its walls, since the increase is not asymmetrical. For the differential diagnosis, the following criteria could be used:

	ATHLETE	HCM
Bizarre ECG pattern of LVE	No.	Yes.
LV cavity < 45 mm	No.	Yes.
LV cavity > 55 mm	Yes.	No.
LAE	No.	Yes.

DIFFERENCES BETWEEN PHYSIOLOGICAL VENTRICULAR HYPERTROPHY OF ATHLETES AND HYPERTROPHIC CARDIOMYOPATHY (HCM) WHEN BOTH PRESENT WALL THICKNESS BETWEEN 13 mm & 15 mm

	ATHLETE	HCM
Female Gender	Negative.	Positive.
Decrease Of Hypertrophy With Less Physical Training	Positive.	Negative.
Family History Or Provable Genetic Mutation	Negative.	Positive.

ECG SUSPECTED FEATURES OF HCM

The prevalence of HCM in highly trained athletes is extremely rare. Structural and functional changes associated with HCM naturally select out most individuals from competitive sports. Screening athletes with ECHO is not cost effective. However, ECG is useful in selecting out those individuals who may have pathological LVH for subsequent echocardiography[1].

1) Left Atrial Enlargement is observed in ≈ 20% of cases of HCM
2) Paroxysmal AF in ≈ 10% of cases.
3) QRS axis between 0º and –90º in ≈ 30% of cases
4) LAFB (10%) QRS axis beyond –30º
5) LVH with strain pattern: QRS/T angle near 180º and prominent R waves in intermediary precordial leads in 80% of cases.
6) In ≈ 10%, very wide R waves in V1 and aVR associated to deep and “clean” Q waves in V5 and V6 and/or in inferior leads, by > of septal vector 1 in 10%.
7) Increasing R wave from V2 to V4 and decreasing from V5 to V6, R wave of V4 of greater voltage than the other precordial leads (74%), absence of q waves in I (87%) and V5 (91%), anterior shift of QRS loop in the HP (74%) and R vector of posterior and rightward orientation (91%); in familial forms, 50%
8) QS pattern from V1 to V4. In sporadic ones is 15%.
9) Pseudo infarction Q waves <40 ms and deep. Q waves in young patients with absence of AMI history.
10) 85% NS-SVT (30%), frequent PVCs (>10/h) in 20%, isolated, coupled, (25%) polymorphic (20%), and S-VT.

1. **Basavarajaiah S, Wilson M, Whyte G, Shah A, McKenna W, Sharma S. Prevalence of hypertrophic cardiomyopathy in highly trained athletes: relevance to pre-participation screening. J Am Coll Cardiol. 2008 Mar 11;51:1033-1039.**

ECG SUSPECTED FEATURES OF ARVD/C

Prolongation of QRS complex (110 ms) in right precordial leads (V1-V3) in adult patients in absence of CRBBB (prolonged S wave upstroke) from V1 to V3, 55 ms is the most prevalent characteristic of ECG (95% of cases) and are correlated with the severity of the disease and induction of VT in PVS.

The result from the addition of QRS complexes duration from V1 + V2 + V3 when divided by the addition of the duration of QRS complexes from V4 through V6 (V4 + V5 + V6). When this equation results in a value ≥ than 1.2, it constitutes a sign of high sensitivity for ARVD diagnosis, since it is present in 98% of patients carriers of this cardiomyopathy.
Epsilon waves (ε): (30%) late potentials or low amplitude and short duration oscillations near the J point (before or immediately after): It is considered a major criterion.
Brugada ECG type 1 pattern-like
Complete RBBB(15%) or IRBBB (18%)
MVT with LBBB pattern
In absence of CRBBB in patients >12 years old, negative T wave from V1 to V3 is a sign with great value for diagnosis.

1) Buffo Sequeira I, et al. Utility of ECG precordial S-wave duration in diagnosis of Arrhythmogenic Right Ventricular Dysplasia/Cardiomyopathy (ARVD/C) in pediatric patients canadian. Cardiovascular Congress 2003 Abstrac 504.

The 36th Bethesda Guidelines for Participation in Competitive Sports

Vigorous, aerobic competitive athletics are generally advised for athletes with disease that are commonly associated with SCD[1]

1. Hypertrophic Cardiomyopathy (HCM)
2. Arrhythmogenic Right Ventricular Cardiomyopathy (ARVD)
3. Miocarditis[2]
4. Coronary artery anomalies
5. Idiopathic Dilated Cardiomyopaties(IDCM)
6. Marfan syndrome
7. Long-QT syndrome LQTS with possible exception of LQT3
8. CPVT
9. Brugada syndrome
10. Documented coronary disease
11. Valvular heart disease
12. Congenital heart disease
13. Patients with implantable cardioverter defibrilators.

1. **Maron BJ, Chaitman BR, Ackerman MJ, Bayés de Luna A, Corrado D, Crosson JE, Deal BJ, Driscoll DJ, Estes NA 3rd, Araújo CG, Liang DH, Mitten MJ, Myerburg RJ, Pelliccia A, Thompson PD, Towbin JA, Van Camp SP; Working Groups of the American Heart Association Committee on Exercise, Cardiac Rehabilitation, and Prevention; Councils on Clinical Cardiology and Cardiovascular Disease in the Young. Recommendations for physical activity and recreational sports participation for young patients with genetic cardiovascular diseases. Circulation. 2004 Jun 8;109:2807-2816.**
2. **Furian TC, Hansel J, Nagenrauft S, Niess AM.Collapse of a competitive athlete at a competition - Case 1/2010. MyocarditisDtsch Med Wochenschr. 2010 May;135:79.**

Colleagues Opinions

Dear Andrés,

An intersting and challenging case. I would first exclude Fabry´s disease and the apical form of HCM (by MRT; echo is not sufficient to view the apical region adequately). Post peri-myocarditis is less probable.

Regards

Prof. Dr. Dr.h.c. Günter Breithardt, M.D., EFESC, FACC, FHRS
Professor of Medicine (Cardiology)
Universitätsklinikum Münster
D-48129 Münster
Tel.: +49 (0)251-868821
Fax.: +49 (0)251-868822
e-mail: g.breithardt@uni-muenster.de
Internet: http://medc.uni-muenster.de

Dear Andres,
I 100% agree with Dr. Martini.
Thanks!
Li
so so difficult to rule out an hypertrophic cardiomyopathy. The only way to differentiate from an athlete's heart is to suspend physical activity for 6 months and repeat investigations
bye
bortolo

The ECG looks most compatible with apical hypertrophy. The problem is that the Echo shows concentric hypertrophy and this Echo pattern may be seen in competitive athletes. We are told he is a professional soccer player and these types of ECGs have been reported in athletes.
There is a very recent article in Europace detailing the consensus statement re the Athlets's ECG and this ECG would be considered abnormal.
I recall a very extensive study by Dr. Barrie Maron of ECGs from athletes with long term followup studies of athletes with deep T wave inversion.In this study several went on to develop IHSS and there were several deaths.
Would be inclined to do provocative maneuvers looking for pressure gradients.
In any event would suggest cessation of vigorous athletic activity and close follow up.
Porf Dr.Melvin M Scheinman, :
Department of Cardiac Electrophysiology, University of California San Francisco, San Francisco, California, USA.**Address: UCSFElectrophysiology Service 500 Parnassus AvenueSan Francisco, CA 94143-1354Telephone/FAX/E-mail: Phone: (415) 476-5706Fax: (415) 476-6260 email: scheinman@medicine.ucsf.edu**

The ECG appears to be compatible with hypertrophic cardiomyopathy. It is unusual for patients with ARVC to have any ST elevation before the T waves are inverted. It is not a normal ECG in an athlete. I will be interested in further information.

Prof. Dr. Frank Marcus- Arizona.

Hola al foro...la interpretacion seria...Ritmo sinusal FC 60-58 por min PR 0.20 Onda P 0.08 eje 70° aprox QRS 0.08 QT 0.48 con QTC 0.490 seg con T negativas y asimetricas en DI II III AVF de V2 a V6 y T + en aVR y aVL.
Conclusion: bradicardia sinusal,no cumple los criterios de Sokolow Lyon que en este ECG es de 31 mm,y el pac tiene 24 años y no 35 como es uno de los criterios de HVI (o por lo menos es lo que tengo entendido).
Me llama la atencion el QTC que es de 0.490 y me parece que esta prolongado .
Las ondas T negativas son asimetricas y me parece que se debe a SVI fisiológica
Le realizarian una ergometria para ver comportamiento de las ondas T que tendrian que desaparecer en ejercicio
Dra. Marilina Ortega

Estimada Dra Ortega ud refiere un QT prolongado. Solo con una ergometria le impresiona sufiente para descartar patologias asociadas a este?. Es un hallazgo habitual en deportistas de alto rendimiento este trastorno? Sin implicancias patologicas?
Martin Ibarrola Prevalence and significance of an isolated long QT interval in elite athletes Sandeep Basavarajaiah1, Matthew Wilson2, Gregory Whyte3, Ajay Shah1, Elijah Behr4,and Sanjay Sharma1*1University Hospital Lewisham/King's College Hospital, London SE5 9RS, UK; 2Olympic Medical Institute, London, UK;3Liverpool John Moores University, Liverpool, England, UK; and 4St. George's Hospital Medical School, London, UKReceived 9 July 2007; revised 15 August 2007; accepted 27 August 2007; online publish-ahead-of-print 18 October 2007See page 2825 for the editorial comment on this article (doi:10.1093/eurheartj/ehm491)
Si no logra acceder al articulo mi mail es martinibarrola@gmail.com con gusto se lo envio
Saludos
Martin Ibarrola

Estimado Andrés: Queria comentar que en mi pasaje por SP estuve un corto tiempo en el servicio del Prof Pablo Mofa. En esos dias se hizo un registro de algunos jovenes militares, todos atletas por cierto, y ninguno tenia un vectocardiograma " normal" o tipico.
Emilio Marigliano

To my very good friend Professor Andrés Pérez Riera

I am sending to you my commentaries about the ECG from a young athletic soccer player in according to my experience witch I am searching during the last 15 years. The ECG morphology expressing the myocardial LV remodeling. Ernst Mayer, died a year ago at the age of 104 , and was a Neo-Darwinist biologist and philosopher wrote a book entitle **"THIS IS BIOLOGY THE SCIENCE OF LIVING WORLD"**. in this book he said that science star when someone start to ask" WHY?" . but not" WHAT?"

I will try describe what and suggesting WHY in according to the huge amount of information from the molecular biology provide by the basic science in the experimental laboratory

I willt? Try to crumb to pieces the ECG

1) **The R waves:** the voltage of the R waves are high in V4, V5, > than 25mm and high R waves in II, III and aVR.
2) **The R waves have high voltage and are very narrow.** V4 and V5 expressed the electrical potential of the lower IV septum and the apex Why is so high? Because indicate longitudinal hypertrophy. As is well known that the myocites are unable to replicate itself, but the sarcomers are intracellulary replicated. But why is so narrow the QRS complexes ,despite the hypertrophy? Because the over expression of connexine 43. In the early stages of hypertrophy witch accelerate the electrotonic conduction between the cells (see papers connexin 43 and hypertrophy mainly in Cardiovascular Research) The conexine 43 is manifested mostly in the upper face of the myocytes allowing a very fast longitudinal conduction.
3) Why the inferior wall leads are also involved in the hypertrophyc process ? As we have learn from the acute circumferential subendocardial ischemia , by the way describe by me in the literature, this leads expresses the inferoapical area. This finding indicates that the anterior apex(V4,V5) and the posterior apex are also involved in the hypertrophy suggesting a circumferential apical hypertrophy.

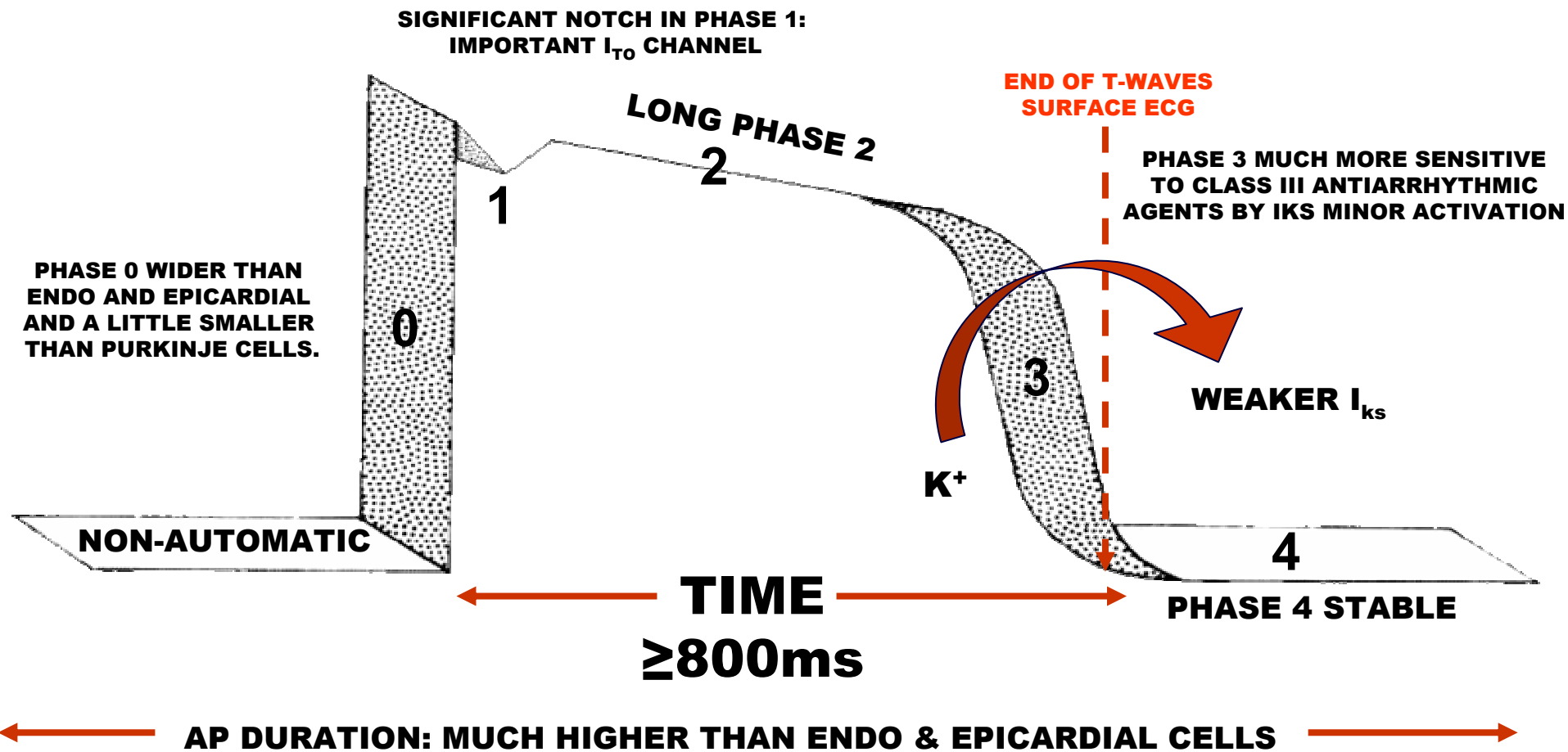
The deep S wave in aVL indicate the inferior hypertrophy. Why appears this peculiar phenotype pattern in the young genetic -phenotype prices?.

As is well known that in the first 8-9 Month the babies ECG morphology have a right ventricular fetal pattern. After this time appears the left ventricular predominance electro tonic forces , THE PHYSIOLOGICAL REMODELIN but because the distribution of the "tension electrical feedback receptors " in the endocardium have not not an homogenous distribution (in the rats the high concentration of receptors are in the apical lateral and septal wall) the phenotypes of the physiological, remodeling will be eccentric (in according to my research in about 300 Childs from 1 y o to 11 yo 80 % are eccentric Y 20% are concentric remodeling about 25 % of the remodeling of the child LV are expresses by very high R waves in V4,V5. This finding suggest that this area of LV has phenotype remodeling. This pattern could also suggest that the highest concentration of tension receptors are in the apex. This people who have this pattern since childhood will always develop an hypertrophy en the apex expressed by very high R waves in V4 and V5 , in case of overloading in the LV such extreme physical training , hypertrophy, valvular defect with diastolic or systolic overloading , The gene activated the biological cascade o of hypertrophy are the same" reactive fetal genes" , but the area activated are different witch explain the different morphology expressed in the ECG.

2) THE GIANT INVERTED T WAVES:

Why the inverted T waves are inverted and very deep ? They are inverted because the action potential of the endocardium is shorter than the epicardium. The ECG expresses in the precordial leads the shorter QT ,as in reperfused myocardial infarction or high end diastolic pressure with hypertrophy or without.

As is worldwide known that positive T wave in the precordial leads expresses a shorter Q/T in epicardium than in the endocardium. This physiological phenomenon is determine by the evolution ,of:" trial and error" dictated by the natural selection to the best adapted the retrograde conduction from epi to endocardium must find the endo refractory to excitation, in this way the avoid the appearance of fatal reentry arrhythmias.
Charles Antzelevitch find in mammals important barrier witch detain the retrograde conduction named M cell with a longest action potential.



Why is the T waves are so deep? Because the opponent area from the apex is the basal area witch is the thickest area, than other part of the LV, The ECG expresses the potentials of the basal. The action potential of the basis is shorter than the affected apical area, There are other
clinical pathological situation such in the Tako-Tsubo syndrome or high intracranial pressure witch present the same giant inverted T waves.
I believe that only the ECG is able to give such sophisticated information , and I say that the molecular biology and cardiac genetic inject a rejuvenated molecule to the old and crippled ECG
My best regard to the all electrocardiofilps
Samuel Sclarovsky

Muy querido amigo Prof, Andrés Pérez Riera. Le envio este análisis de acuerdo a mi experiencia de investigación en las hipertrofias. En el famoso libro del biólogo y filósofo neodarwinista" Ernst Mayr titulado "THIS IS BIOLOGY THE SCIENCE OF LIVING WORLD" dice que la ciencia comienza cuando se pregunta porque (Why?) y no que es esto? (what is this?). Trataré de explicar que es y también el porque. Para esto permitame analizar este ECG, segun el mensaje que unicamente "el viejo y arrugado ECG" puede proporcionar y que la biologia molecular y la genética le inyectaron varias moleculas rejuvenadoras a nuestro método.

Las ondas R: son de gran voltage en V4 ,V5 (> 25 mmm), III, II, aVR y las S son profundas en aVL. (En V4-V6 no se puede saber exactamente cuan altas son porque estar superpuestas con las S de la derivaciones anteriores).

Porque son tan altas y a la vez finas ? Por 2 causas:

a) Los complejos QRS son muy altos por que los miocitos con el ejercicio no se pueden autoreplicar, pero sus sarcómeros se van agregando en forma longitudinal (no transversal) aumentando la dimension longitudinal y no transversal.

b) Por la alta concentración de conexina 43 en la superficie del miocito se acelera la conduccion intraventricular lo que justifica que el QRS sea fino o estrecho.

2) Porque tambien están involucradas las derivaciones inferiores? Porque III y aVF expresan los potenciales de la cara posterior del apex (esto lo aprendimos con las isquemias circunferenciales agudas subendocárdica descriptas por nosotros) sugeriendo la existencia de hipertofia apical circunferencial.

Porque aparece en este joven este ECG con este fenotipo especial y no todos los deportista no desarrollan? Porque esto es determiando geneticamente. Después de los 9 meses,aparece los potenciales predominantes del VI (antes de esta fecha persisten todavia la predominancia de los vectores derechos fetales).

El ECG de remodelación fisiológica del bebé es muy polimórfico. Porque? Porque la distribución de los receptores de tensión mecánica de retoralimentación no estan distribuidos en forma uniforme en las paredes del corazón. El resultado de esta desigual distribución es que el 80% de las remodelaciones fisiológicas son asimétricas (la mayor concentración de receptores estan distribuidos en el apex y septum en las ratas) y apenas en el 20% son simétricas.
El 25% de las remodelaciones asimétricas presentan una R de 25mmm en V5-V6. Los que presentan este patrón van a desarrollar siempre es este tipo de hipertrofia ante cualquier sobrecarga sobre en VI , sea genética, hipertensiva o por enfermedad valvular que inducen a hipertrofia sea diastólica o sistólica.

4) Las ondas T gigantes invertidas: Porque las ondas invertidas son gigantes máximas en V4-V5? Porque estas derivaciones expresan los cambios entre los potenciales del endocardio y epicardio el endocardio es mas corta induciendo que el subepicardio expresandose antes las mas cortas

Y porque son gigantes? Porque las ondas T apicales son la cara opuesta a la base de VI. Los potenciales de acción son mas cortos en la base que los que los patológicos del apex, como es sabido la base es mas gruesa que las demas areas del corazon y el apex es la parte mas finas (esto ocurre tambien en las patologias con tormentas adrenérgicas, como el Tako Tsubo e alta tensión intracerebral como la hemorragia subaracnoidea) ya que el apex contiene alta concentración de receptores adrenérgicos en comparación de las demás areas.

5)Lo que es interesante es que este fenomeno se desenvuelve en forma progresiva Al pricipio tiene unicamente remodelacion fisiologica apical , despues de los 10 años aparecen T invertidas muy pequeñas en V4,V5 ,III , II , a los 18 anos aparece un cuadro mas completo ,con ondas T invertidas profundas, lo que los cardiologos genetistas denominan "delay gene penetration", y cerca de los 40 años en adelante se distribuye en todas las derivaciones.

Porque apareció en este joven a los 24 años? Provablemente la penetración genetica fué muy precóz. Yo denomino a esta fase electrocardiográfica que se encuentra este joben de fase 4 de las hipertrofias apicales. Mucho mas tarde disminuira la conexina 43, y los complejos se harán anchos y el ECO mostrará discinesia apical.

Porque algunos de estos pacientes sufren intensa angina a los 18 anos con coronarias normales y y otros se les descubre ocasionalmente a edad muy tardia?

Porque toda hIpertrofia miocárdica es hipercinética y exige un aumento de flujo sanguineo

Como se hace esto? todo miocito hipertrofico debe estar acompañado por angiogenesis capilar concomitantemente. Esta sincronia está a cargo de un gen especial que si esta mutado surge angina inestable por demanda ,pero esta no se ve en el electro con tachycardia y ST deprimido, pero la otra no tiene expreson electrocardiografica , pero hay que pensarla porque el tratamiento es diferente.

Querido Andrés hace 15 años que estoy investigando el problema, y tengo mas de 700 ECG con este fenómeno , y repito que unicamente el ECG y el vecto puede dar esta información

Un fraternal abrazo a todos los electrocardiofilos

Samuel Sclarovsky