Would it be possible an ECG/VCG like this, in the absence of structural heart disease? unexplainable giant T-wave inversion







https://ekgvcg.wordpress.com/

Andrés Ricardo Pérez-Riera M.D.Ph.D.¹; Raimundo Barbosa-Barros M.D²; Luiz Carlos de Abreu P.h.D.³

- <u>-</u> Post-Graduates Advisor at Design of Studies and Scientific Writing Laboratory in the ABC Faculty of Medicine - ABC Foundation - Santo André - São Paulo – Brazi
- Chief of the Coronary Center of the Hospital de Messejana Dr. Carlos Alberto Studart Gomes. Fortaleza Brazil
- ω ω School of Public Health Visiting Scientist at Program in Molecular and Integrative Physiological Sciences (MIPS), Department of Environmental Health | Harvard T.H. Chan

Portuguese: Reporte de caso

Seria possível um eletrocardiograma como este, na ausência de doença cardíaca estrutural?

Identificação: Caucasiano, 38 anos, arquiteto, normolíneo

importância. Sempre hígido. Motivo da consulta: avaliação clínica para iniciar prática esportiva em academia. Assintomático, nega vícios, ou antecedentes pessoais de

Antecedentes familiares: negativos

Exame físico: nada digno de nota. Altura: 1.80m; Peso 83kg; PA 130/80mmHg; FC 62bpm; pulsos periféricos presentes

ECG/VCG: próximos slides. Ecocardiografia transtorácica (ETT):

esquerdo = 36mm (VN 28-40). Câmaras cardíacas de dimensões normais: Diâmetro diastólico do VE = 47 mm (VN 38-52); Diâmetro sistólico do VE = 30 mm (VN 26-34); átrio

Espessura miocárdica normal: septo = 9 mm, parede posterior do VE = 9 mm

Desempenho ventricular preservado, sem alterações de contractilidade segmentar.

Função diastólica do ventrículo esquerdo preservada (Doppler tecidual).

Fração de ejeção do ventrículo esquerdo = 0,66 % (VN 0,55-080).

Valvas cardíacas com textura e mobilidade normais

Fluxos transvalvares ao estudo com Doppler e mapeamento de fluxo em cores: normais.

Aorta torácica sem alterações anatômicas. Diâmetro da aorta = 34 mm (VN 28-40).

Pericárdio normal.

Conclusão: Exame dentro dos parâmetros da normalidade.

Solicitamos ressonância magnética do coração com realce tardio para avaliação de fibrose / infarto e viabilidade miocárdica.

English: Case Report

Would it be possible an ECG/VCG like this, in the absence of structural heart disease?

Identification: Caucasian, 38 years old, architect, normoline.

importance. Always healthy. Reason for consultation: clinical evaluation to start practicing sports in the gym. Asymptomatic, denies addictions or personal history of

Negative family background for hereditary diseases.

ECG/VCG Physical examination: nothing worthy of note. Height 1.80m; Weight 83kg.; BP 130/80mmHg, HR 62bpm, peripheral pulses present and regular.

Transthoracic echocardiography:

(NV 28-40). Normal dimensions of cardiac chambers: LV diastolic diameter= 47 mm (NV 38-52); LV systolic diameter 30mm (NV 26-34); left atrium= 36mm

Diastolic thickness of septum and LV posterior wall 9 mm each (normal myocardial thickness)

Preserved ventricular performance without segment contractility alterations

Preserved LV diastolic function (Tissue Doppler)

Normal Left Ventricular Ejection Fraction = 0.66% (NVs from 0.55 to 080)

Heart valves with normal texture and mobility

Normal Doppler color flow mapping

Normal thoracic aorta diameter =34 mm (NV 28-40)

Normal pericardium

Conclusion

Examination within normal parameters.

viability. We request cardiovascular magnetic resonance (CMR) with late gadolinium enhancement for evaluation of fibrosis / infarction and myocardial









ECG/VCG correlation in the Horizontal Plane



ECG/VCG correlation in the Right Sagittal Plane

Colleagues opinions

Dear Andrés

segment depression in anterolateral and high lateral regions. Sokolow-Lyon close to 50 mm; intrinsecoid deflexion in V5 around some degree (although echo is normal). In my opinion there is clear LV electrical remodeling. Need some other investigation, like 0.06 second; QRST angle = 140 degrees (planar); T peak-Tend interval 100 ms. Can't rule out hypertrophyc cardiomyopathy of NMR searching for delayed enhancement. Cannot release for sports participation until finish the investigation. Although there is no clear heart disease by Echo and physical exam., there is clear electrical LV changes compatible with ST

Dalmo Antônio Ribeiro-Moreira M.D.Ph.D. São Paulo Brazil

Chief Medical Electrophysiology and Cardiac Arrhythmia Dante Pazzanese Institute of Cardiology Sao Paulo Brazil



MRI should show the changes. This is probably apical hypertrophic cardiomyopathy, which can be difficult to diagnose with echocardiography.

Kind regards **Kjell Nikus, MD, PhD** Tampere, Finland





Spanish

Hola amigos Mi opinión

ventana apical o no ser realiza una sub-xifoidea), el ECG muestra ondas T profundas negativas en las derivaciones precordiales de mas de La clínica no dice nada relevante y a pesar de que el ecocardiograma no decir nada a nivel apical de VI (a veces no se tiene una buena 10 mm lo que sugieren fuertemente una miocardiopatía hipertrófica apical o forma japonesa de Yamaguchi.

hipertrofia del segmento apical. hacia enfermedad coronaria, por consecuencia el paso inmediato seria una RNM con realce tardío con gadolinio que podría demostrar la La ventriculografía mostraría la típica imagen de "as de pique" y coronarias normales; pero como la clínica y examen físico no orientan

Se descarto miocarditis chagásica con serología? vNo pienso en cardiopatía isquémica

Saludos cordiales

Dr. Juan José Sirena Santiago del Estero Argentina

English

Hello friends My opinion

apical hypertrophic cardiomyopathy or Japanese form of Yamaguchi. apical window or not makes a sub-xiphoid), the ECG shows deep negative T waves in precordial leads (more than 10 mm) which strongly suggest The clinic says nothing relevant and although the echocardiogram did not say anything to apical level VI (sometimes you do not have a good

apical hypertrophy segment. oriented towards coronary artery disease, consequently immediate step would be an MRI with late gadolinium enhancement that could prove Ventriculography will show the typical image of "ace of spades" and normal coronary arteries; but as clinical and physical examination not

myocarditis Chagas serology was discarded? I do not think of Coronary heart disease Best regards

Juan Jose Sirena MD Santiago del Estero Argentina



Spanish

magnética. Le haría un buen Eco con un buen equipo. Si es normal, lo dudo (a veces no se aprecia bien la verdadera punta) le solicitó una Resonancia

Me cuesta creer que el eco sea normal

Saludos a todos, y gracias por este fabuloso espacio

Miguel Amor MD

English

transthoracic echocardiography) I would do an echocardiogram with good equipment. If results normal - which I doubt- (sometimes it is not easy to analyze the tip of the heart by

Is hard to believe that the echocardiogram is normal. In this case, I would request a Cardiac Magnetic Resonance

Greetings to all and thanks for this fabulous space

Miguel Amor MD

Estimado profesor:

Saludo afectuosamente una miocardiopatia hipertrofica como se ha demostrado en estudios anteriores. Este paciente con un ECG que muestra ondas T negativas y con un ecocardiograma dentro de la normalidad, puede que mas adelante desarrolle

Dear Professor:

cardiomyopathy as demonstrated in previous studies This patient with an ECG showing T waves negatives and with an echocardiogram within normal limits, may later develop hypertrophic

Your truly Eduardo Quiñones MD



Spanish

Hola querido Andrés: El ECG no es normal para un paciente caucasiano deportista.

Presenta un BAV de 1er grado con signos de HVI (R peak time en V2 y V3 anormales y criterios ECG de HVI (índice de Sokolov positivo).

además de las derivaciones precordiales de V2 a V6 El infra desnivel del segmento ST es máximo de V3 a V6 hasta de 1,5 mm no es habitual en caucásicos y se registra además en DI, DII y aVF,

de los miembros también presenta onda T negativa pero asimétrica sugestiva de HVI. La onda T negativa gigante en precordiales de V2 a V6 máxima de V3 a V6 simétricas que simulan una T de origen isquémico. En las derivaciones

En aVR presenta supra desnivel del segmento ST que expresa el incremento de las presiones interventriculares

El intervalo QTc se encuentra prolongado, lo que no es habitual en la HVI fisiológica del deportista.

llenado diastólico VI ni crecimiento auricular izquierdo. En la ecocardiografía el diámetro diastólico VI no es el habitual de un deportista con estos cambios ECG (47 mm). No refieren alteraciones del

sospechar ya que no presenta sintomatología Descartado los trastornos hidroelectrolíticos como la hipocalcemia severa (no me impresiona) y la hipertensión endocraneana de la cual no puedo

la misma, sería interesante además de la RNM cardiaca con realce tardío con gadolinio. El diagnóstico es el de una miocardiopatía hipertrófica por los cambios electrocardiográficos. Dado que los espesores parietales no concuerdan con

trazado del paciente. gradientes intraventriculares en reposo pero si en el esfuerzo, y que traerán HVI concéntrica a predominio apical y lateral como evidencia el La realización de un ecuestres con ejercicio y descartar una forma de miocardiopatía hipertrófica obstructiva dinámica, en la cual no obtendrá

Un cordial saludo

Martín Ibarrola



English: Hello dear Andrés: The ECG is not normal for a Caucasian athlete.

from V2 to V6 The ST segment depression of 1.5 mm from V3 to V6 is not common in Caucasians and is also recorded in I, II and aVF, plus precordial leads It has a 1st degree AV block with signs of LVH (prolonged R peak time in V2 and V3 and abnormal Sokolov-Lyon voltage criteria of LVH.

Giant negative T wave in precordial leads V2 to V6 (deeper in V3-V4 that simulate symmetric ischemic T wave

In the limb leads have negative asymmetric T wave suggestive of LVH.

In aVR ST-segment elevation that expresses the increase in ventricular pressures

The QTc interval is prolonged, which is not usual in the physiological athlete LVH

Echocardiography LV diastolic diameter is not the usual sportsman with these ECG changes (47 mm).

No alterations refer LV diastolic filling or left atrial enlargement.

symptoms Discarded electrolyte disturbances such as severe hypocalcemia (not impress me) and intracranial hypertension which I can not suspect as it has no

be interesting addition to the cardiac MRI with late gadolinium enhancement. The diagnosis is a hypertrophic cardiomyopathy by the electrocardiographic changes. Since the parietal thicknesses do not agree with it, it would

effort, and that will bring concentric LVH and lateral apical dominance as evidence tracing the patient. Conducting a stress test could unmask latent dynamic obstructive cardiomyopathy, which will not get intraventricular gradients at rest but in the

A cordial greeting

Martín Ibarrola MD



Yochai Birnbaum The ECG suggests apical hypertrophic cardiomyopathy that is often missed by echo, unless contrast is used . Cardiac MRI is a good choice.

Professor

Medicine-Cardiology Baylor College of Medicine Houston, TX, US



Final comments



ECG/VCG correlation in the Frontal Plane

The normal Ploop in the Frontal Plane





E point: it constitutes the zero point of VCG and it remains stationary before the onset of the P loop. It corresponds to the isoelectric line between

the T wave and the P wave of ECG. The E letter corresponds to the cardiac dipole

end of ventricular repolarization (T loop). **0** point: it corresponds to the end of "biatrial" chamber activation, QRS loop onset (because PR segment does not exist, it is only a point) and the

syndrome, arrhythmogenic right ventricular dysplasia, channelopaties etc elevation or depression. The phenomenon is observed in early repolarization, acute phase of infarction, variant angina, pericarditis, Brugada segment) when it does not present depression or elevation, and T wave onset. In situations where there is depression or elevation of ST segment, J point: in vectorcardiography, it corresponds to 3 elements: end of ventricular depolarization (QRS complex); beginning of repolarization (ST the J point does not coincide with the 0 point, and the greater or lesser distance between both points indicate the greater or lesser ST segment

analysis possible, it is necessary to amplify: 1 mV = 30 cm. (between 0 and 70 ms), next the interatrial septum (between 20 and 45 ms), and finally the left atrium LA (between 30 and 90 ms). To make an **P loop:** loop of small voltage corresponding to the depolarization of the biatrial chamber. The initial part corresponds to the right atrium **RA**

loop is open because atrial repolarization (Tp loop) is diametrically opposite to the P loop. The P loop begins in the E point and ends in the so-called 0 point. The former has an anterior and inferior location in relation to the latter. The P

Items to be studied in the P-loop

The P loop has a short slow conduction in the onset, in at least 2 planes Two small notches are usually observed: one in the efferent limb and another in the afferent limb. Morphology; Aspect: presence of notches; Magnitude and direction of maximal vector: P vector; P-loop rotation: FP: counterclockwise; in the HP: counterclockwise or in eight; and RSP: clockwise. Location of the E (initial) and 0 points (final). Location of the greatest part of the P loop; **Frontal Plane Horizontal Plane Right Sagittal Plane**





Location of the normal T-loop/normal T-wave axis in Frontal Plane in the normal adults

between -35° to +90°. In the present case T axis is located on top right quadrant between -120° to -150°. In the normal adult > 95% of cases T-loop/T-wave axis is located between $+15^{\circ}$ to $+80^{\circ}$ in the frontal plane. Tolerance of extreme values ranges



Positive Sokolow and Lyon index: S of $V_1 + R$ of $V_5 \ge 35$ mm or 3.5 mV in adults older than 30 years old. Maximal QRS-vector, final QRS-vectors located to the right and backward quadrant and ST/T vector located in the right posterior quadrant. Initial vectors of QRS-loop heading to the front and the left, QRS-loop, predominantly located in the left anterior quadrant, increase voltage of



ventricular repolarization vector (T vector SAT) in the horizontal plane is heading to the left and usually discretely to the front near the +10° very T-waves polarity is always positive from \tilde{V}_3 to V_6 ; generally positive in V_2 and frequently negative in V_1 . In normal adults, invariably the

close to V6. In the present case T-loop is located on right back quadrant.



ECG/VCG correlation in the Right Sagittal Plane



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

Vectocardiogram of apical non-obstructive hypertrophic cardiomyopathy

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

This curious form of HCM is characterized by presenting:

- Initial vectors of QRS-loop heading to the front and the left;
- QRS-loop, predominantly located in the left anterior quadrant(anteriorization);
- Increase voltage of Maximal QRS-vector;
- insufficiency associated to T loop in this quadrant. A curious fact is constituted by the association of apical infarction and T loop in right Final vectors located to the right and backward, with ST/T vector in the right posterior quadrant. This is the only case of LVE without coronary
- E point that does not match the 0 point, and located backward and to the right from the latter (Pérez-Riera 2013).

posterior position

T-loop is characteristically discordant related QRS-loop, elongated, assimetric and situated in the right posterior quadrant (Abinader 1982).

the disease (Bielli 1991). Note: the important posterior and rightward shift of the ST/T vector is responsible for the characteristic giant negative T wave (>10 mm) in the leads of the horizontal plane from V2 to V5. T waves at the onset, may not present a significant voltage and may appear later with the evolution of

Stress test may decrease the depth of T waves (Tilmant 1980).

Late gadolinium enhancement for evaluation of fibrosis / infarction and myocardial viability. Cardiovascular magnetic resonance (CMR). Cardiac-magnetic resonance cine image for the global assessment and target heart.

Comments

Right atrium of preserved dimensions

Left atrial dimensions preserved

Right ventricle with preserved dimensions

Preserved systolic function of the right ventricle

Preserved left ventricular dimensions, global and segmental systolic function preserved left ventricle = 72%. LVH with apical predominance, with a maximum thickness of 21 mm (Absolute apical wall thickness > 15 mm is diagnosis of Ap-HCM)

absence of obstruction of in the left ventricular outflow tract.

Absence. thrombi

Diagnostic impression

Normal biventricular systolic function

Left ventricular hypertrophy of apical predominance.

Absence of obstruction in the left ventricular outflow tract

important role in diagnosis. The diagnostic criteria for Ap-HCM: Absolute apical wall thickness > 15 mm, or a ratio of apical to basal left apex. The diagnostic accuracy of echocardiography for Ap-HCM is limited without using contrast or 3-dimensional echocardiography, MRI has an Ap-HCM it may be frequently missed by transthoracic 2-dimensional cardiac echocardiography because of poor image quality of left ventricular ventricular wall thicknesses of 1.3–1.5 (Chun 2010).

configuration is caused by localized apical hypertrophy. A characteristic spade-like shape of the left ventricular cavity can be seen at end diastole on vertical long-axis views and on ventriculography. This

mm. This results in a spade-like appearance of the left ventricular cavity. Delayed enhancement at the cardiac apex is compatible with myocardial fibrosis The vertical long-axis image above shows circumferential increased wall thickness in the apical portion of the left ventricle, measuring up to 16 Other findings on MRI include apical aneurysms and delayed enhancement in the hypertrophic or non-hypertrophic segments.



according to the AHA 17-segmented model. The apical angle (apA) as well as the regional variations in LV wall thickness was analyzed unexplainable giant T-wave inversion and 76 healthy volunteers were prospectively enrolled. The segmented LV wall thickness was measured cardiomyopathy (Ap-CM) according to the current criteria. When the absolute thickness of apical is was below the current diagnostic criteria of with unexplainable giant T-wave inversion in the precordial leads and apical wall thickness <15 mm cannot be diagnosed as apical hypertrophic these subjects should be included into a preclinical scope of Ap-HCM needs further investigations (Wu 2016). A total of 60 subjects with Cardiac-magnetic resonance cine image demonstrating hypertrophy of the apex in an "ace-of-spades" configuration Ap-HCM. Patients Ap-CM, the apical morphological features of subjects with unexplainable giant T-wave inversion are significantly different from normal. Whether Considerable variation in LV wall.



Measuring apical wall thickness

complicates wall thickness measurement and it should be performed on multiple long axis views, perpendicular to the true septal axis and with careful exclusion of papillary muscle origins and trabeculation. perpendicular to the wall. Towards the distal ventricle — as the cavity tapers, the short axis slice is not perpendicular to the wall — this thirds: Basal, mid and apical, the apical cap makes up only the most apical 6% of the ventricle. At the base the short axis slice is truly Left: short axis slice near the base of the heart with max wall thickness of 8 mm. Middle: four chamber view. The left ventricle is divided into



and Registration for Cardiac Imaging (Cerqueira 2002). A 17 segment cardiac model, including the apical cap, suggested by the American Heart Association Writing Group on Myocardial Segmentation Registration for Cardiac Imaging (Cerqueira 2002). model, including the apical cap, has been suggested by the American Heart Association Writing Group on Myocardial Segmentation and contrast studies. A 16 segment model can be used, without the apical cap, as described in an ASE 1989 document. (Schiller 1989) A 17 segment segments" are usually visualized from apical four-chamber, apical two- and three-chamber views. The apical cap can only be appreciated on some Segmental analysis of LV walls based on schematic views, in a parasternal short and long axis orientation, at three different levels. The "apex





Characteristics of Ap-HCM

cardiomyopathic process in HCM is more diffuse than previously regarded, extending beyond the LV myocardium to include thickening of the RV appearance, which can be distinguished from Ap-HCM using cardiac-magnetic resonance imaging (Noureldin 2012; Moon 2004). Patients are significance of LGE for sudden death is incompletely resolved and ultimately future large prospective studies may provide greater insights into enhancement appears to be an important determinant of adverse LV remodeling associated with systolic dysfunction. However, the predictive ventricular arrhythmia has been described, and prophylactic ICD is occasionally performed. However, the prognosis is generally more benign than reported in a few families, the condition is usually sporadic (Arad 2005). The ECG typically shows giant precordial T-wave inversions. SCD from typically male and present in middle age with symptoms of palpitations, chest pain or dyspnea. Although autosomal-dominant inheritance has been Japanese population. Apical thrombus, left-ventricular non-compaction and endomyocardial fibrosis may create a similar echocardiographic seen in western populations. This apical variant constitutes $\approx 25\%$ of cases of HCM in Japan but only 1% to 2% of the cases of HCM in the noninformation impacting diagnosis and clinical management strategies. Ap-HCM (Yamaguchi 1979) is a phenotypic variant of nonobstructive HCM, myocardial fibrosis has raised the expectation that this may represent a novel marker, which may enhance risk stratification. Late gadolinium leaflets and late gadolinium enhancement. The unique capability of contrast-enhanced CMR with late gadolinium enhancement to identify identified unique phenotypic markers of affected genetic status in the absence of LVH including: myocardial crypts, elongated mitral valve wall as well as substantial morphologic diversity with regard to papillary muscles and mitral valve. Among HCM family members, CMR has to CMR imaging in HCM remained largely undetected), end-stage systolic dysfunction, and massive LVH. CMR observations also suggest that the underestimated). High-risk HCM patient subgroups identified with CMR include those with thin-walled scarred LV apical aneurysms (which prior in which hypertrophy of the myocardium predominantly involves the left ventricular apex (Gupta 20016). It is most common in Japan but also this issue. These observations underscore an important role for CMR in the contemporary assessment of patients with HCM, providing important for HCM diagnosis, by identifying areas of segmental hypertrophy (ie., anterolateral wall or apex) not reliably visualized by echo (or Compared with normals, the apex decreased significantly in male and female subjects, respectively. CMR is often superior to echocardiography

other types of HCM (Eriksson 2005).

Genotype-Phenotype Correlations in Apical Hypertrophic Cardiomyopathy

predictor of a positive genetic test. categorized into four morphological subtypes: reverse curve, sigmoidal, neutral contour, and Ap-HCM. The reverse curve HCM is the strongest studied 1053 patients with the diagnosis of HCM underwent sarcomeric genetic testing. Blinded to the genetic test results, each echocardiogram genotype-phenotype correlations in Ap-HCM. Between 1999 and 2007, Researches from the Mayo Clinic, Rochester, Minn, USA (Towe 2015) was scored for septal morphology and phenotyping was performed using the patient's medical record. Echocardiographically, HCM where HCM is underscored by profound phenotypic and genotypic heterogeneity. Little is known about the spectrum and prevalence of mutations and

Deformation patterns in genotyped patients. echocardiographic depiction of ventricular septal morphologies in HCM



echocardiography (63% male, mean age 47.8 ± 15 years, mean left ventricular wall thickness 19.8 ± 6 mm). Left ventricular outflow tract follow-up was 5.5 years (range 0.1-18.2 years). There was no statistical difference between the occurrence rates of adverse events between 75% of the cases genotype-positive and genotype-negative groups. obstruction was uncommon (seven patients; 10%).18 patients (25%) had a positive genetic test, with the majority of mutations found in MYBPC3 Subset analysis was performed to elucidate the genotype, phenotype, and outcome of Ap-HCM. Overall, 71 patients (7%) had Ap-HCM by In this largest cohort of patients with genetic testing for HCM, 7% exhibited Ap-HCM. This subtype is associated with a negative genetic test in (six; 35%) and MYH7 (six; 35%). Follow-up was available on 68 patients (96%) with a median age of 57.3 years (range 19.3-82 years). Mean

In patients with Ap-HCM the two more common mutations were (MYBPC3-HCM and MYH7-HCM) remained most common among patients

who had a positive genetic test.

MYLK2	MYL3	MYL2	MYH7	МҮН6	MYBPC3	LAMP-2	GLA	CSRP3	CKYAB	CDVAD	COX15	CAV3	ANKRD1	ACTN2	ACTC	Gene
Myosin Light Chain kinase 2	Myosin Light Chain 3, slow	Myosin Regulatory Light Chain 2, slow	β-Myosin Heavy Chain 7 (Ap-HCM)	β-Myosin Heavy Chain 6	Myosin Binding Protein C (Ap-HCM)	Lysomal-Associated Membrane Protein 2	α-Galactosidase	Cysteine and Glycine-rich Protein 3		Create line of	COX 15 homolog. Cytochrome C oxidase assembly protein	Caveolin 3	Cardiac Ankyrin Repeat, Domain 1	α-Actinin2	α-Cardiac Actin	Gene name
VCL	TTR	TTN	TPM1	TNNT2	TNNI3	TCAP	IAT	T A 7	TNNC1		PRKAG2	PLN	NEXN	MYOZ2	MYO6	Gene
Vinculin	Transthyretin	Titin	α-Tropomyosin 1	Cardiac Troponin T, type 2	Cardiac Troponin I, type 3	Titin-cap (Telethonin)		Tafazzin	Cardiac Troponin C, type 1		AMP = activated Protein kinase, y2, non-catalytic subunit	Phospholamban	Nexilin (F actin binding protein)	Myozenin 2	Unconventional Myosin VI	Gene name

Mutations in HCM (in red color mutations of Ap-HCM)

Summary of proposed classification system. ARVC, arrhythmogenic right ventricular cardiomyopathy; DCM, dilated cardiomyopathy; HCM, hypertrophic cardiomyopathy; RCM, restrictive cardiomyopathy



muscle fibers tissue results in disturbed electrical signals and conduction in the heart (arrhythmia); frequently it is DCM>HCM>RCM>ARVC). more severe symptoms and worse outcomes than when the disease manifests in adulthood (from a structural-functional point of view, most .-1 within a few months following delivery; 6 Ś heart cannot supply the organs with sufficient circulation during contraction; 4 ω 2 muscle wall unable to generate enough pumping force during contractions; muscle fibers results in smaller chamber volume and sometimes blocks the blood flow (obstruction): There are various forms of cardiomyopathy, each with different underlying causes for the insufficient circulation. Restrictive cardiomy opathy (RCM): due to their stiffness, the ventricles do not get refilled with enough blood during relaxation, hence the Paediatric cardiomyopathy: this type of cardiomyopathy becomes manifest in infancy or early childhood, and is usually characterized by Peripartum cardiomyopathy (PPCM): a special form of dilated cardiomyopathy that becomes manifest towards the end of pregnancy or Left-ventricular non-compaction cardiomyopathy (LVNC): the wall of the left ventricle is spongiform, characterized by a meshwork of Hypertrophic cardiomyopathy (HCM): a thickened myocardium due to abnormal growth and arrangement (hypertrophy and disarray) of Dilated cardiomyopathy (DCM): one or both of the ventricles (in most cases only the left one) become enlarged with a thin, weakened Arrhythmogenic right ventricular cardiomyopathy (ARVC): the replacement of the degenerating myocardium with scar (fibrofatty)

The MOGE(S) classification for a phenotype-genotype nomenclature of cardiomyopathy: endorsed by the World Heart Federation (Arbustini 2013)

The recently proposed MOGE(S) for classification of cardiomyopathies was proposed, but its clinical use has not been described. Nosology

system embodies all of these characteristics, and describes the

- 1. Morphofunctional phenotype (M),
- 2. Organ(s) involvement (O)
- **3.** Genetic inheritance pattern (G)
- 4 Etiological annotation (E) including genetic defect or underlying disease/substrate, and
- **N** Status: Functional status (S) of the disease using both the ACC/AHA stage and NYHA functional class

The proposed nomenclature is supported by a web-assisted application and assists in the description of cardiomyopathy in symptomatic or group cardiomyopathies on their etiological basis, describe complex genetics, and create collaborative registries asymptomatic patients and family members in the context of genetic testing. It is expected that such a nomenclature would help

nosology, currently remains unproven as a fully practical solution. This recent propose for classification attempt to harmonize these competing systems named the MOGE(S) system, based on descriptive logical

Hypertrophic
cardiomyopa
thy
classification

Classification and differences of obstructive and non-obstructive forms of Hypertrophic Cardiomyopathy.

) Obstructive form (OHCM)

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

2) Non-obstructive Form (NO-HCM)

- Septal asymmetric with no obstruction;
- Mid-septal hypertrophic cardiomyopathy
- Apical Hypertrophic Cardiomyopathy (Ap-HCM): 2%, 3% to 8%.
- Lateral and/or posterolateral;
- Concentric, symmetrical, or homogeneous hypertrophic: 5%.
- Right ventricle: 2%.

Free wall with progressive decrease of thickness from the base to the apex (the same as normal).	Septum with greater thickness in the superior part (basal)(20% or in the middle portion(1%).	Obstructive form (OHCM)
Free wall with no or normal decrease of the thickness from the base to the apex.	Septum with greater thickness in the inferior part (apical).	Non-obstructive form (NOHCM)

Obstructive forms of Hypertrophic Cardiomyopathy (OHCM)



Septal asymmetrical with resting left ventricular outflow obstruction (obstruction at subaortic level): is found in approximately 20% of the patients, classically occurs at the, and is associated with mitral valve systolic anterior motion (SAM).



Mid-ventricular obstructive HCM HCM(MVO-HCM) asymmetric LV hypertrophy with MVO and elevated intraventricular pressure gradients.(1%).

Π. Non-obstructive forms of Hypertrophic Cardiomyopathy (NO-HCM)

Septal asymmetrical without obstruction



Lateral and/or posterolateral







Left Ventriculography in Ap-HCM

The "ace-of-spades" sign on left ventriculography being pathognomonic (Olearczyk 2008)



The di
agnosis
is based
on the f
following
element
s in ApH
CM

- Giant and negative T waves from V2 to V4;
- 3Mild symptoms and benign course
- Aspect of spade cards in left ventriculography;
- 4 Absence of ventricular gradient;
- S rather abruptly on rare occasions. Disappearance of giant negative T-waves may also occur slowly and progressively in patients, in Transition from normal T wave to negative T wave required several years and remained usually unchanged thereafter. This change may occur whom apical aneurysm had developed (Sakamoto 2001)

persistence of apical blood pool distal to cavity obliteration in systole). Observation: The following ECG changes are indicative of apical aneurysm in Ap-HCM patients (Observation: An apical aneurysm is defined as

- Increase in QRS-complex duration
- QRS-complex fragmentation
- Decrease in QRS-complex amplitude
- 2 ST-segment elevation(STE) form V4 to V6 (j-point in V5) are statistically more frequent in patients with apical aneurysm compared to and the presence of the apical aneurysm (Spearman's $\rho=0.895$, p<0.001) (Ozeke 2015). those without aneurysm (93% vs 7%, p<0.001). There are a positive correlation between the presence of the STE in V4-6 derivations
- V. Positivisation of negative T waves in V3-V6 (deep t-wave in V5) (Pennacchini 2015).

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

Differential diagnosis of Ap-HCM (Yusuf 2011)

Disease

- LV apical cardiac tumors
- 2 LV apical thrombus
- Isolated ventricular non-compaction
- ω 4. Endomyocardial fibrosis
- Ś Coronary artery disease

Echocardiogram/coronary angiogram and LVG LVG/CMRI CMRI/CC1 Echocardiogram with contrast/CCT/CMRI Echocardiogram with contrast/CCT/CMRI Diagnostic tool to establish diagnosis of Ap-HCM

Disorders that may involve the left ventricular apex

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

- Takotsubo cardiomyopathy "octopus trap", transient apical ballooning syndrome, apical ballooning cardiomyopathy, stress-induced earned a hypercontractile base of the LV is often noted. It is the hallmark bulging out of the apex of the heart with preserved function of the base that cardiomyopathy, Gebrochenes-Herz-Syndrome, broken heart, and simply stress cardiomyopathy a bulging out of the left ventricular apex with
- $\mathbf{\dot{\nu}}$ Left ventricular aneurysms, pseudoaneurysms and congenital apical diverticula (see differential diagnosis in the next slide)
- 3. Apical ventricular remodelling
- 4. Apical hypertrophic cardiomyopathy (ApHCM) (Cisneros 2011)
- 5. Left ventricular non-compaction
- 6 Arrhythmogenic right ventricular cardiomyopathy/dysplasia(ARVC/D with left ventricular involvement
- -1 Left ventricular false tendons: are fibrous or fibromuscular bands that stretch across the LV from the septum to the free wall. They can also be mistaken for abnormalities such as tumors, subaortic membranes, thrombus borders, septal hypertrophy. tether to a papillary muscle, but unlike the chordae tendineae, do not connect to the mitral leaflets. They are anatomic variants that should not
- 8. Chronic chagasic myocardiopathy

diagnosis With an emphasis on the diagnostic criteria and imaging features. In this setting cardiac imaging methods can provide the clue to obtaining the

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

Differences among aneurysms, pseudo-aneurysms and congenital diverticula

Relative merits of each non-invasive imaging technique for the assessment of HCM

Factor assessed	Echocardiography	Multi-detector CT	MR iaging
LV volume	++++	+	+++++
Ejection fraction	++++	++++	+++++
LV filling pressure	++++		‡
Dynamic obstruction	++++	+	++++
Ischemial CFR	+	ı	‡
Tissue characterization	+	+	+++++

Prognosis of Ap-HCM

cardiac MRI are both significant predictors of major adverse clinical events. Thus, patients with both apical aneurysm and >5% LGE are at highest option (Xiao 2016). Ap-HCM patients, the aneurism presence and the extent of quantification of late gadolinium enhancement (LGE) identified by confers an extremely poor prognosis. Early aggressive therapies should be considered for this entity and prophylactic aneurysmectomy may be an eosinophilia-myalgia syndrome. The rare association of NMDs with ApHC might be due to absence of systematic neurologic investigations of small burden of myocardial fibrosis and less severe diastolic dysfunction and subsequently more favorable clinical manifestations in comparison and are at increased risk of ventricular arrhythmias and death, especially in the presence of an apical akinetic chamber. Ap-HCM show a relatively clinical course. However, the mutation Arg719Trp in the cardiac beta-myosin heavy chain (beta MHC) gene is a high risk factor for sudden death myocardial infarction (Eriksson 2002). Patients with apical aneurysm have a largely unfavorable clinical course, and are often unrecognized dysfunction and 8. Myocardial infarction. The most frequent morbid events in patients with Ap-HCM are atrial fibrillation (10% of patients) and patients with AHC and vice versa (Finsterer 2009). The probability of survival without morbid events at 10 years was $77 \pm 4\%$. Several NMDs, such as limb girdle muscular dystrophy, glycogen storage disease, metabolic myopathy, myopathy of unknown significance, or thrombi, stroke, ventricular aneurysms/apical akinetic chamber, myocardial infarction, progressive heart failure, high incidence of coronary and can be associated with an unusual Ap-HCM (Dohlemann 2000). Current evidence suggests that these patients experience severe symptoms useful information to predict the occurrence of adverse events in Ap- HCM (Kim 2015). views (4ch, 2ch, LVOT view) from the apical cap to the blood cavity in systole. The minimum of the three measurements is used risk for major adverse events. The rate of apical obliteration is defined as the net obliteration to end-diastolic apical cap thickness (Hanneman because echocardiography is limited in the assessment of the left ventricular (LV) apex. Apical aneurysm is not rare in patients with HCM and it fibrillation.; 4. New York heart association class \geq III at baseline (Yan 2012); 5. Ventricular aneurysms.; 6. Apical thrombi.; 7. Diastolic independent predictors of cardiovascular morbidity are identified: 1. Age at diagnosis ≥ 60 years.; 2. Left atrial diameter ≥ 36 mm.; 3. Atrial (Kim 2015). Morbid sequel, and others extra-cardiac disorders such as: Atrial fibrillation, diastolic dysfunction, left atrial enlargement, apical with other HCMs. This may be one explanation of why most patients with Ap-HCM show a benign course of disease compared with non-Ap-HCM The ratio of obliteration to cavity is defined as the end-systolic obliteration to cavity height. The ratio of obliteration to cavity could provide 2014). Abnormal systolic apical cavity obliteration is graded as > 20 mm or > 10 mm. Measured in the 5 mm long axis cine crosscuts in the 3 fistulae and morbid atrial fibrillation (Chung 2010). cardiovascular death. Neuromuscular disorders Ap-HCM is only rarely associated with The prognosis of Ap-HCM with regard to SCD is believed to be better than that of common HCM. Patients with the Ap-HCM had a benign

Events were defined as a composite of new onset of AF, stroke, HF, and cardiovascular (CV) death

Management

- Drugs Medications used to treat symptomatic patients with ApHCM include verapamil, beta-blockers and antiarrhythmic agents such as amiodarone and procainamide
- 2 acknowledged SCD risk factors were present: Implantable cardioverter defibrillator: An ICD is recommended for high risk patients ICD indications in HCM if one or more of the
- Family history of premature HCM-related death particularly if sudden, in a close relative, or multiple in occurrence;
- Prior cardiac arrest
- Sustained VT and Non-SVT
- Unexplained syncope, particularly in young patients, or if demonstrated to be arrhythmia-based;
- Frequent, multiple, or prolonged episodes of NSVT documented on serial ambulatory Holter monitoring;
- Hypotensive or attenuated blood pressure response to exercise;
- Extreme left ventricular hypertrophy (≥ 30 mm) particularly in young patients
- An abnormal blood pressure response to exercise
- I possible risk factors included: atrial fibrillation, myocardial ischaemia, left ventricular outflow obstruction, high risk mutations, and intense (competitive) physical exertion
- ŝ ablation or transcoronary alcohol ablation is required in some cases (Inada 2011). Ablation: Monomorphic VT in a ApHCM can be due to endocardial, epicardial or intramural reentry in areas of apical scar. Epicardial
- 4 Apical myectomy improves functional status by decreasing left ventricular end-diastolic pressure, improving operative compliance, and diastolic volume (Schaff 2010) increasing stroke volume. This procedure might be of value in other patients with HCM who have severe hypertrophy and small LV end-
- ப Heart transplantation: ApHCM is a morphologic variant in which the hypertrophy is primarily localized to the apex of the LV. A subset of transplantation has been the only therapeutic option available for such patients patients have progressive, drug-refractory diastolic heart failure with severely limiting symptoms caused by low cardiac output. Heart

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Thank you very much! This is my wife and I in the typical Japanese restaurant.

