Mujer blanca de media edad asintomática con Electrocardiograma atípico

Asymptomatic middle-aged Caucasian woman with atypical Electrocardiogram

Caso del Dr. Luciano Pereira, de Ciudad del Este, Paraguay

Case of Dr. Luciano Pereyra, M.D.; Ciudad del Este, Paraguay

Caucasian woman (German origin), assymptomatic, 36 years old, functional class I, three normal children, pregnancies without problems, normotensive, negative familial background
Physical examination:nothing worthy of note. 61 kg, 1.67 m, BP = 110/70
Normal cardiovascular examination,
Preformed ECG as chek up.
Which is the ECG and clinical diagnosis?

Greeting

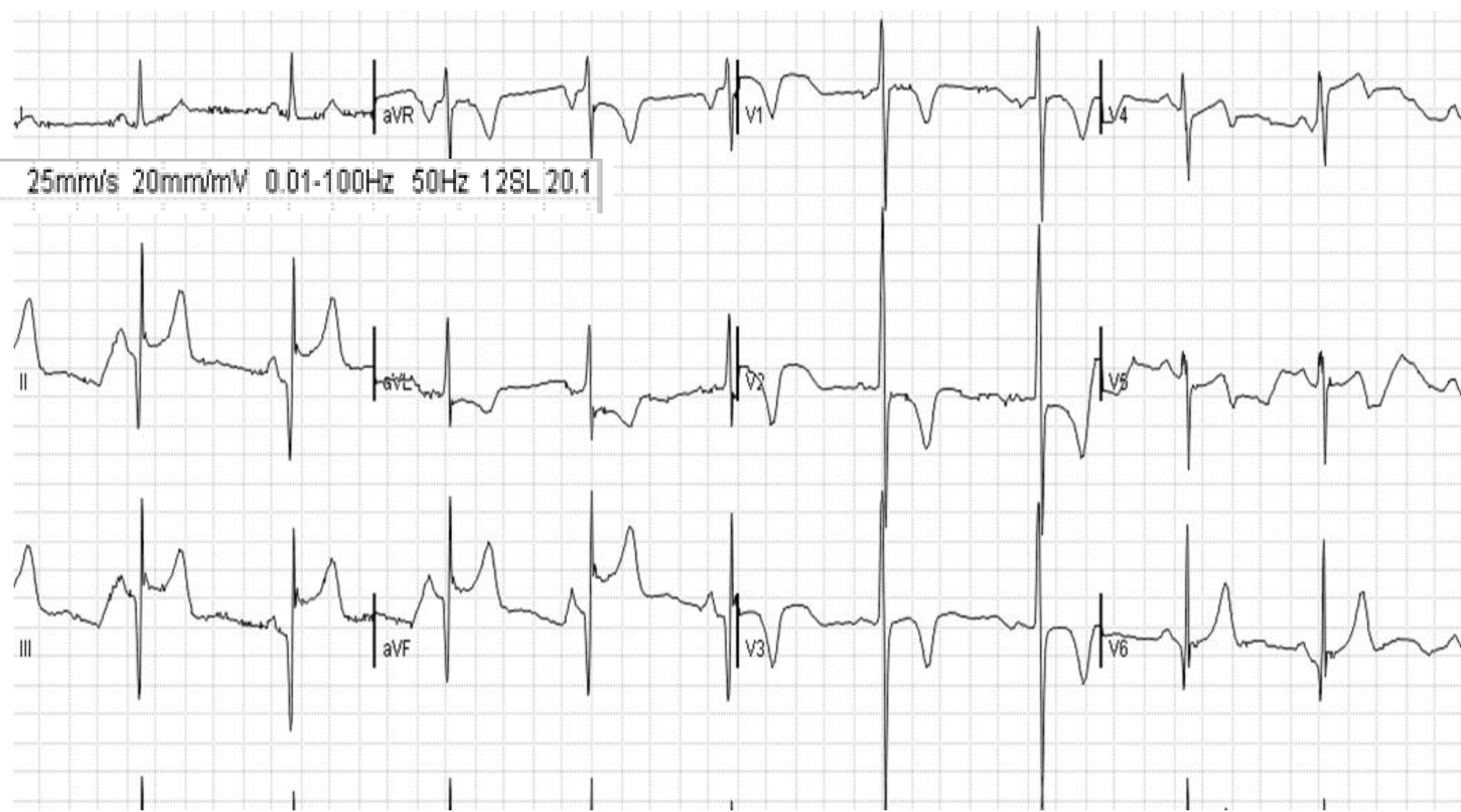
Luciano Pereira M.D.

36 años, sexo femenino, clase funcional I, tres hijos normales, embarazos sin problemas, normotensa.

Saludos.

Luciano Pereira, M.D., Ciudad del Este, Paraguai

Saludos.
Luciano Pereira, M.D., Ciudad del Este, Paraguai


aVR
V1
V4
25mm/s 20mm/mV 0.01-100Hz 50Hz 12SL 20.1
II
aVL
V2
V5
III
aVF
V3
V6

Colleagues opinions

Hi
Very interesting ECG.
1. Sharp q-waves in the inferior leads
2. Large voltages in the right precordial leads
3. T-wave inversion

My first guess would be HOCM.
It resembles apical HOCM (Japanese form).
Eager to learn about the ECHO in this case

Adrian Baranchuk, M.D. FACC FRCPC
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Spanish

El caso del Dr Pereira sugiere una hipertrofia excéntrica muy pronunciada del septo alto y medio muy probablemente obstructiva.

Las ondas Q profundas y finas en DII, DIII y aVF son consecuencia de la hipertrofia excéntrica

Las ondas T muy profundas en precordiales derechas tambien obedecer a la hipertrofia septal.

Las ondas R de bajo voltaje observadas en V4-V5 se deben a la hipertrofia septal.

Es um caso muy raro.

Un fraternal abrazo

Samuel Sclarovsky M.D. Israel.

English

The case of Dr Pereira suggests a big hypertrophy of the high and medium of the interventricular septum. Probably obstructive.(idiopathic hypertrophy subaortic stenosis)

The deep and fine Q waves in DII, DIII and aVF are the result of the eccentric hypertrophy.

The deep T waves in right precordial leads also are consequence of the septal hypertrophy.

The low voltage R waves observed in V4-V5 are due to the septal hypertrophy.

It is a very rare case.

A fraternal hug

Samuel Sclarovsky M.D. Israel.

Dear Andrés & Luciano,

This is a young woman who is asymptomatic, has a normal physical exam, but an abnormal electrocardiogram.
We do not know the family history. This is always important!
The ECG shows sinus rhythm, normal intervals, but deep Q waves in inferior leads with tall R wave in V1. T waves are negative V1-4 which is abnormal at her age.
The most likely diagnosis is hypertrophic cardiomyopathy which would explain the abnormal Q waves in inferior leads and tall R wave in V1 as well as the T waves.
An inferior and posterior myocardial infarction is less likely since the Q waves seem to be narrow.
Chagas cardiomyopathy could give this ECG with an inferior and posterior aneurysm.
There appears to be ST elevation in inferior leads but this is not clear because the baseline is not stable.
Muscular dystrophy, mainly Duchenne muscular dystrophy, can result in tall R waves in V1, but not the deep Q waves in V1.
A **ventricular septal defect** can explain the prominent R and S waves in V1-4, but her physical exam was normal.
Hypertrophic cardiomyopathy is the most likely diagnosis. An echocardiogram will help with the diagnosis.
Thank you for all the interesting cases you share with us!

Mario D. Gonzalez, M.D.
Professor of Medicine
Internist, Cardiologist
Director, Clinical Electrophysiology
Penn State Heart & Vascular Institute
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Hello forum members,

We, in the course of electrocardiography, we like to headline electrocardiograms, in this case instead of: Caucasian middle-aged woman with atypical asymptomatic electrocardiogram would call: middle-aged white woman with Q waves asymptomatic unexpected high voltage (which is equivalent to: **hypertrophic cardiomyopathy!!!**)

Miguel **Fiol Sala M.D.**
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Servicio de Medicina Intensiva y Unidad Coronaria, Hospital Universitario Son Espases, Palma de Mallorca, Baleares, España

Spanish
Esimados

El ecg es altamente sugestivo de una miocardiopatia hipertrofica que en contexto de la clinica referida deberia descartarse la tipo V o APICAL . Un ecocardiograma transtorácico con una vista apical o subxifoidea podria darnos el diagnostico. Una resonancia nuclear magnética o el VGG la confirmaria.

Abrazos

Juan Jose Sirena, M.D.
Santiago del Estero, Argentina

Dear all,
The ECG is highly suggestive of hypertrophic cardiomyopathy in the clinical context of Ap-HCM type referred should be ruled. A transthoracic echocardiogram with apical or subxiphoid approach could give the diagnosis. Also a cardiac magnetic resonance imaging or the VCG would confirm.
Hugs

Hypertrophic cardiomyopathy with predominantly septal hypertrophy.

Best regards

Kjell Nikus, M.D.Ph.D.
Professor in Cardiology
Tampere
Finland

http://www.researchgate.net/profile/Kjell_Nikus2/publications

Spanish
Querido Profesor Andrés,
Interesante caso.
No hay antecedentes familiares?
Luce una MCH de una modalidad no "exclusivamente apical".
Debe haber en este caso engrosamiento inferior y de VD.
abrazo fuerte
Alberto Morales Salinas MD, MPH.
Departamento de Docencia-Investigaciones,
Cardiocentro "Ernesto Che Guevara", Villa Clara (VC), Cuba.
Miembro Titular de las Sociedades Cubanas de Cardiología y Medicina Familiar.
Presidente Sociedad de HTA en VC
Secretario Sociedad Centroamericana y del Caribe de HTA y Prevención Cardiovascular (SCCH)
E-mail: cardioams@yahoo.es
http://www.linkedin.com/pub/alberto-morales-salinas/39/1a0/668

Dear Professor Andres
Interesting case.
No family history?
It seems a HCM “non exclusively apical".
In this case must be lower intraventricular septum and right ventricle thickening.
tight hug

Caros Riera e Luciano
ECG realmente atípico com Ritmo Sinusal, padrão de repolarizacao precoce na parede inferior, Onda T invertida de V1 a V3, entalhe de retardo ascende de S em V1 e O da epsilon em V1. Conclue-se por Displasia Arritmogenica de Ventrículo Direito. Exames de imagem seria o próximo passo Ecocardiografia ou RNM.
Abraços
Adail Paixão Almeida, M.D.
Vitoria da Conquista, Bahia, Brasil

Dear Riera and Luciano
This ECG is really atypical, it has early repolarization pattern in inferior wall, inverted T-wave from V1 to V3, notched in the ascendente ramp of S wave or épsilon wave in V1. Conclusion: ARVC/D. The next diagnosis step would be echocardiogram or cardiac magnetic ressonance.
Hugs
Adail

Final diagnosis

- Non-obstructive form of hypertrophic cardiomyopathy: severe basal and middle septum hypertrophy and moderate apical.
- Presence of epsilon waves: this is the first description of epsilon wave in hypertrophic caediomyopathy.
- Deep (between 7 and 13 mm) and narrow sharp “clean” Q-waves in inferior leads and V6. Q-waves ≥ 3 mm deep should raise suspicion of hypertrophic cardiomyopathy.

Color Doppler echocardiographic study

Name: l. K. 36 years old. Weight: 61 kg; Height: 1.67 m. Body surface: 1.69 m².

Left ventricular ejection fraction 85% (VN> 50%) (Teichoz); Shortening fraction: 52% (V.N.> 26%)

LV end-diastolic volume: 70 cc .; LV end-systolic volume: 11 cc.

Mitral, aortic, tricuspid and pulmonary valves: all normal.

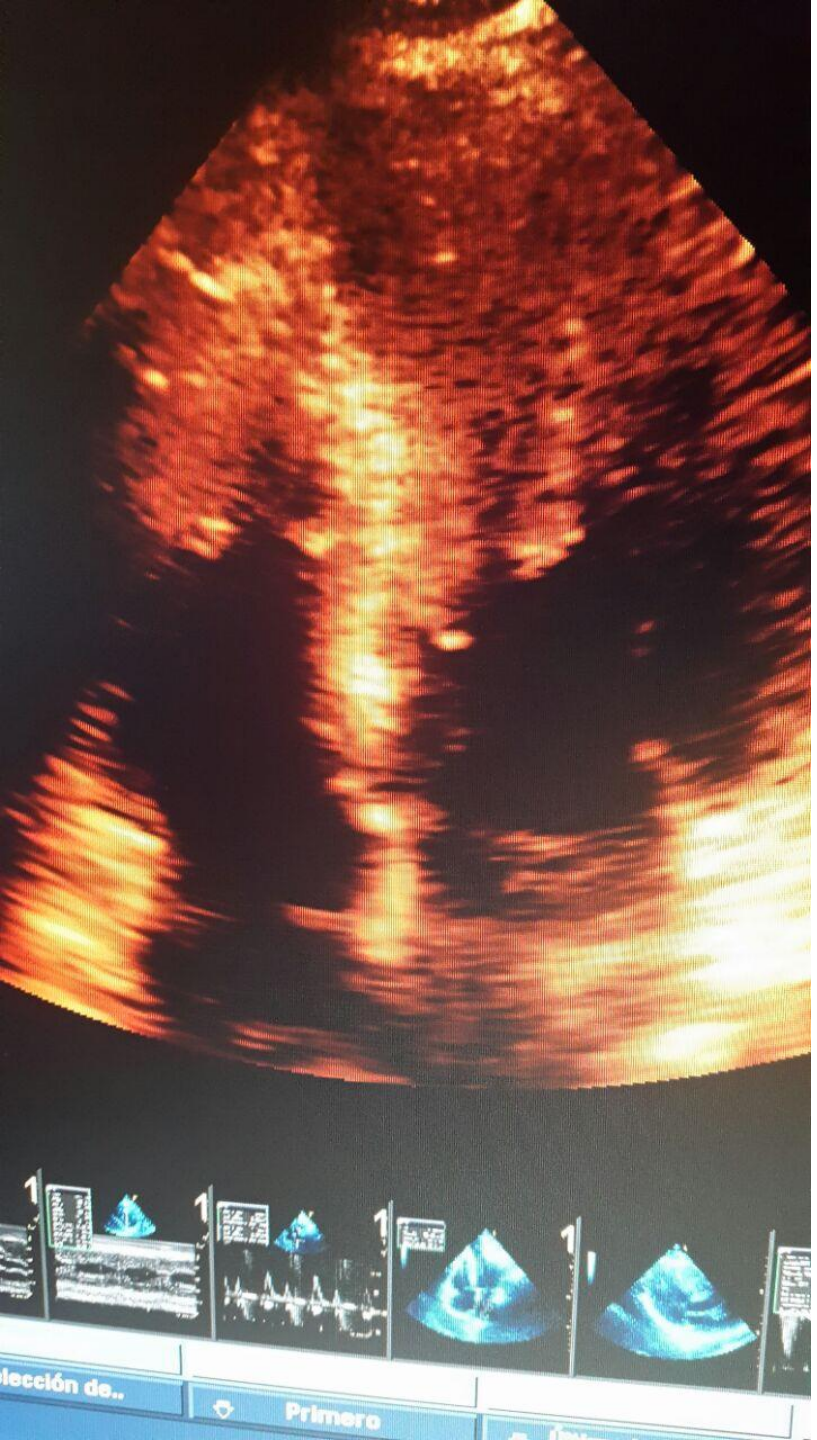
Cardiac dimensions: normal. Wall thickness: significantly increased in the basal interventricular septum (diastolic 3.9 mm, systolic 3.1 mm) and moderate in medial and apical level. Wall motion: Normal. Pericardium: Normal. Absence of intracardiac thrombus and/or intracavitary masses. Transvalvular flows normal characteristics. Absence of LVOT gradient (subaortic).

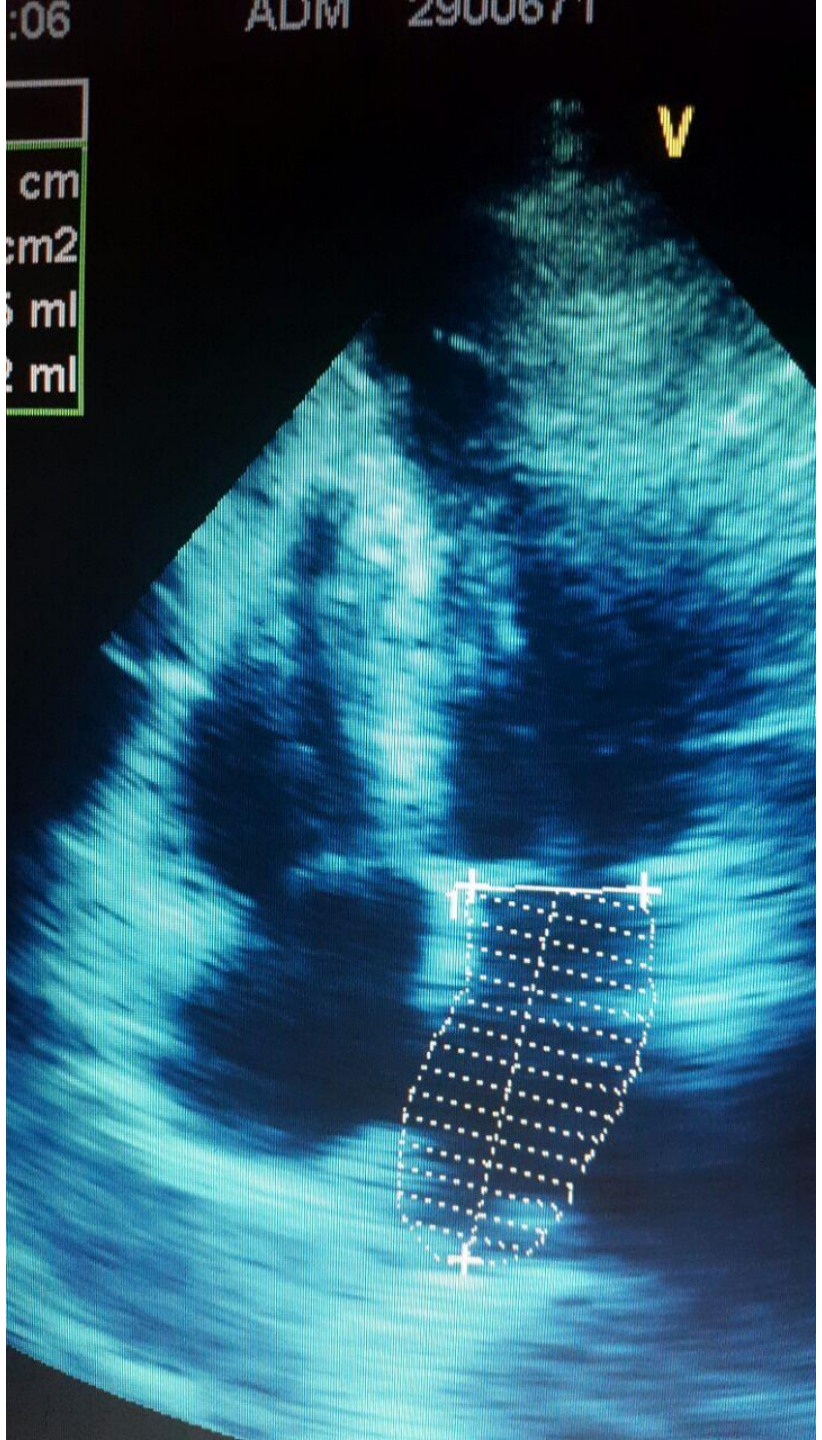
Systolic and diastolic function of the left ventricle are preserved. Right ventricular function is preserved.

Systolic anterior motion of the mitral valve (SAM) leaflet towards the left ventricular outflow tract during systole is observed, but no significant gradient is recorded in LVOT. Septal/LVPW ratio > 1.5.

Conclusion: non-obstructive asymmetric hypertrophic cardiomyopathy located on interventricular septum.

	Measurement	Range values
Left atrium diameter (cm)	28.6	2.7 – 3.8
Volume	23 ml (13 ml/m²)	Until 34 ml/m²
Aortic root	29.7	27-41
Right ventricle	19	<30
Interventricular septum thickness (mm)	39	6-10
LV posterior wall (LVPW)	10	6-10
Left ventricle end-diastolic diameter (largest cardiac dimension)	40.1	Until 55
Left ventricle end-systolic diameter (smallest cardiac dimension)	11	Variable


Primero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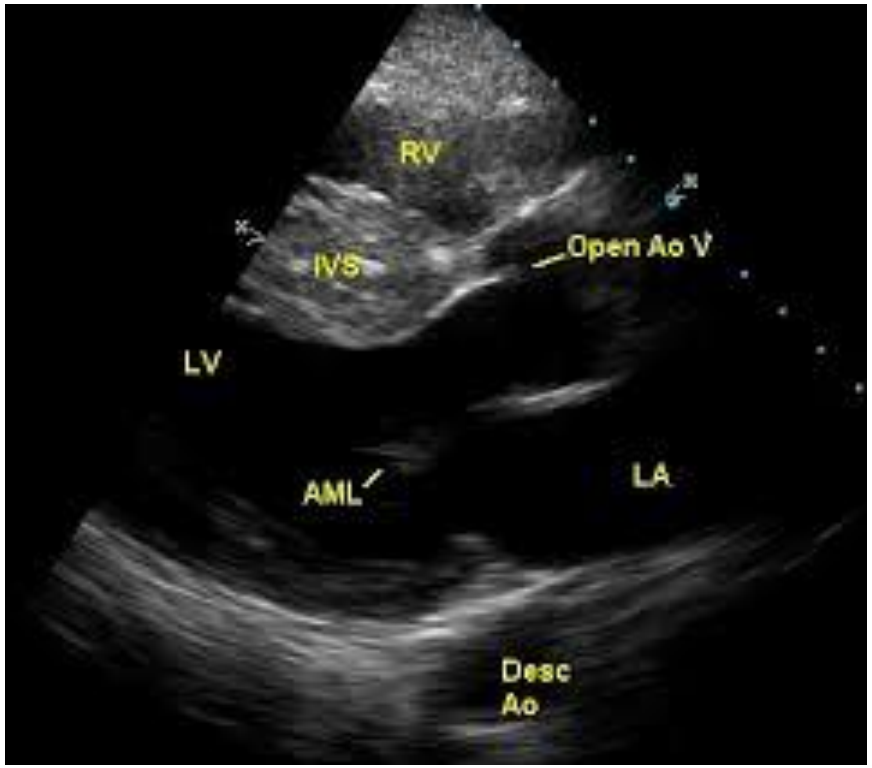

RV
IVS
Open Ao V
LV
AML
LA
Desc
Ao

Hypertrophic cardiomyopathy

Definition: Hypertrophic cardiomyopathy (HCM) is a complex disease in which the heart muscle becomes abnormally hypertrophied of variable and non-selective hypertrophy location (both in the septum and the free wall) without apparent cause.

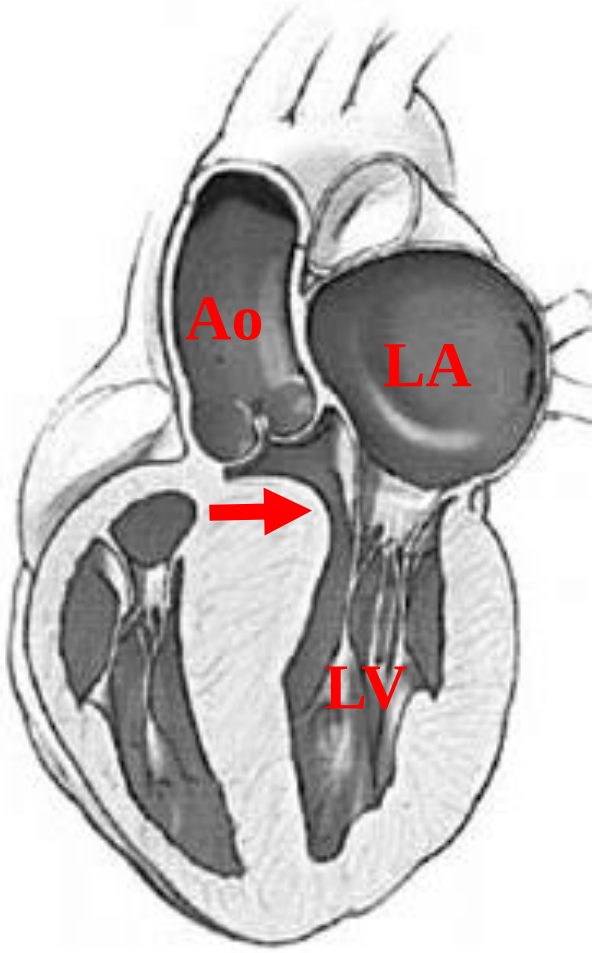
In most cases, the entity is familial, autosomal polygenic dominant, with high degree of penetrance (60% familial versus 40% sporadic), consequence of mutation on sarcomere proteins with:

Classification

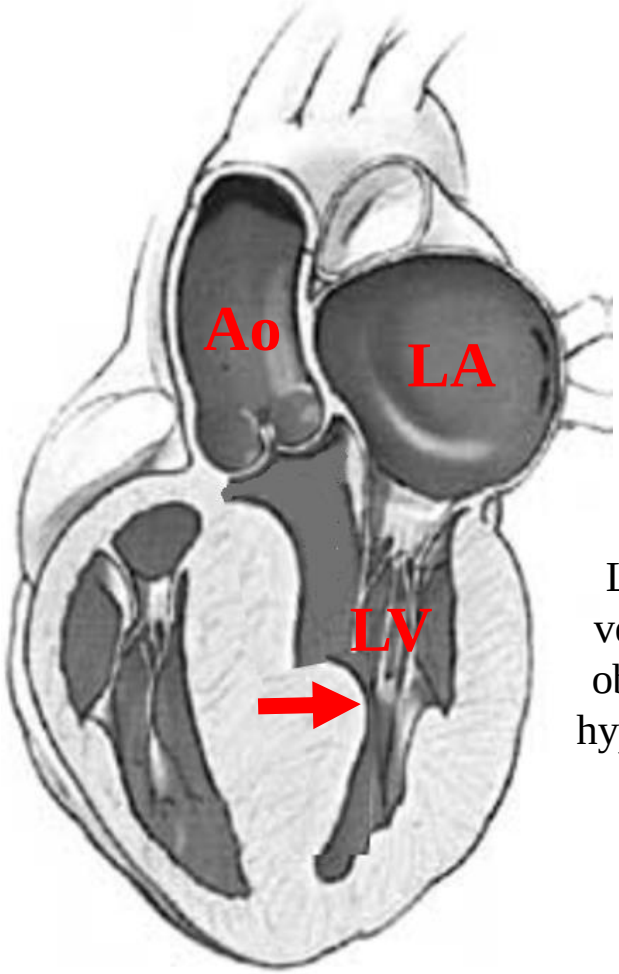
1) **Obstructive forms (O-HCM)**
   - Septal asymmetrical with resting left ventricular outflow obstruction (obstruction at subaortic level): is ≈ 20% of cases.
   - Mid-ventricular obstruction HCM HCM (MVO-HCM) asymmetric LV hypertrophy with MVO and elevated intraventricular pressure gradients (1%).

2) **Non-obstructive forms (NO-HCM)**
   - Septal asymetrical with no obstruction (the present case);
   - Apical Hypertrophic Cardiomiopathy (Ap HCM): 2%, 3% to 8%;
   - Lateral and/or posterolateral;
   - Concentric or symmetrical, or homogeneous hypertrophic: 5%;
   - Right ventricle: 2%.

Septal asymmetrical with subaortic obstruction



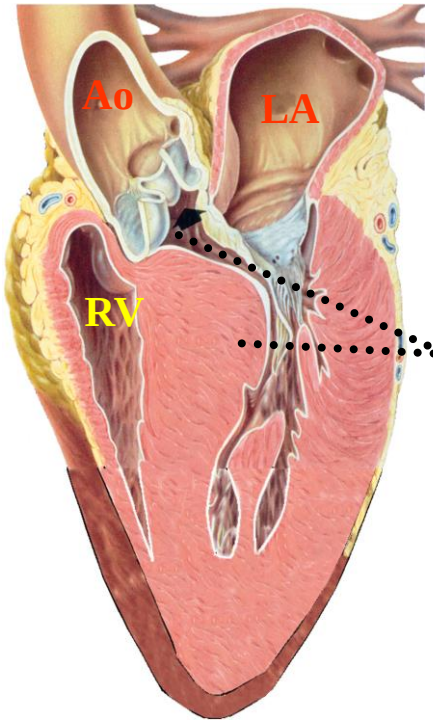
Left mid-ventricular obstruction (MVO-HCM)



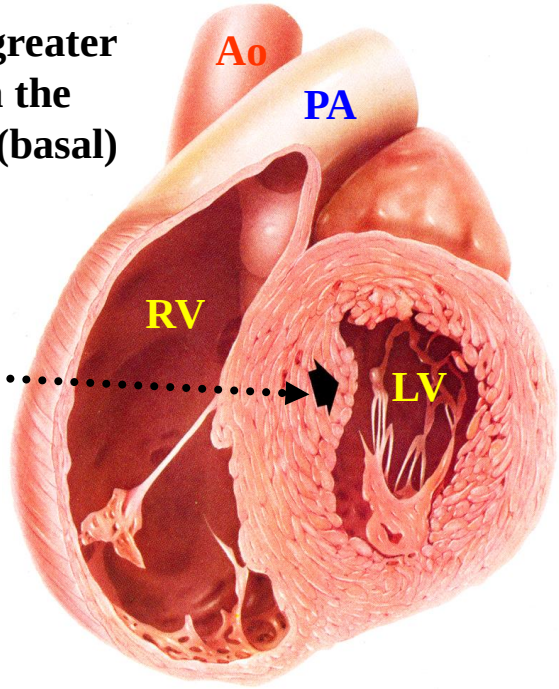
Left mid-ventricular obstructive hypertrophy

Dynamic subaortic obstruction (O-HCM)	Mid-ventricular obstruction (MVO-HCM)
Septum with greater thickness in the superior part (basal) (20% or in the middle portion (1%).	Septum with greater thickness in the middle part.
Free wall with progressive decrease of thickness from the base to the apex (the same as normal).	Free wall with no or normal decrease of the thickness from the base to the apex.

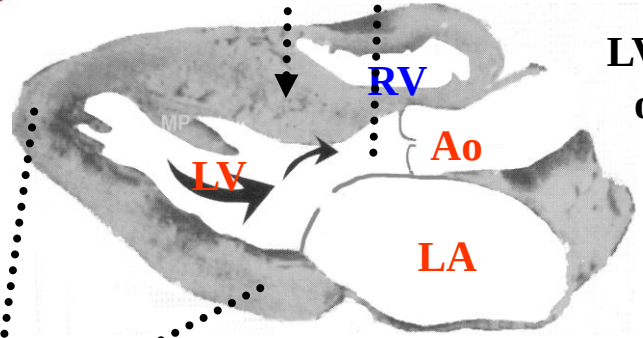
Septal asymmetrical with subaortic obstruction (O-HCM)



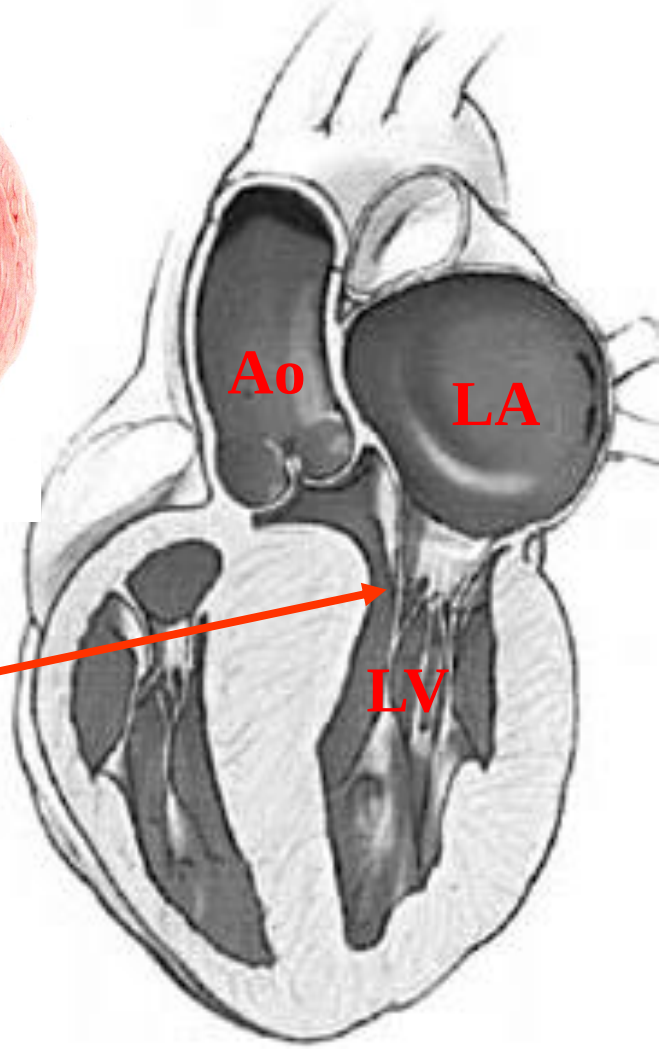
Septum with greater thickness in the superior part (basal)



LVOT



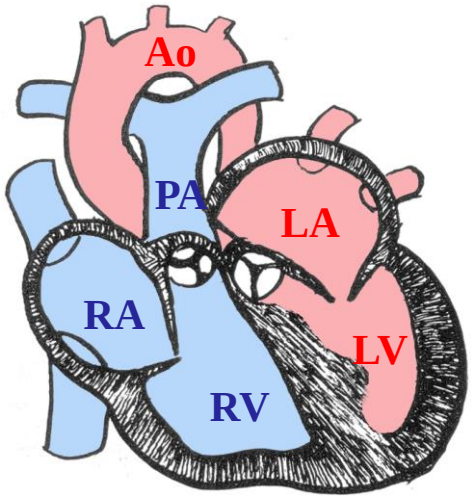
LVOT: LV outflow tract



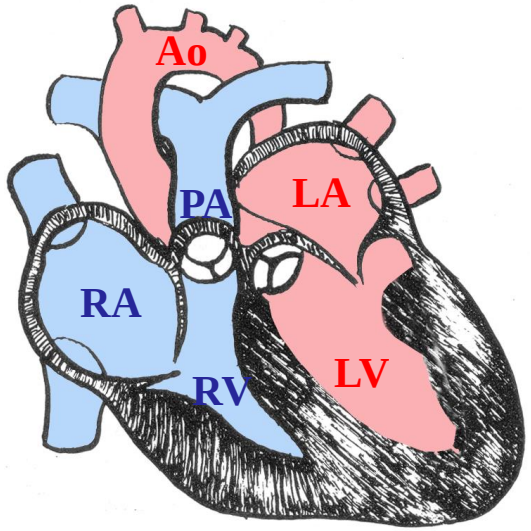
Free wall with progressive decrease of thickness from the base to the apex

Outline of Non-Obstructive forms of HCM

Septal asymmetrical without or minimal obstruction

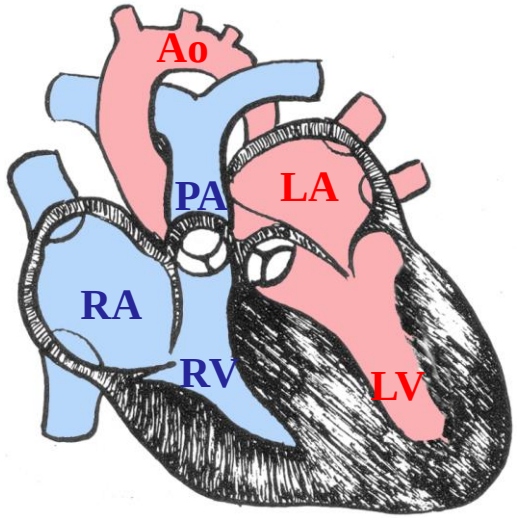


Lateral and/or posterolateral

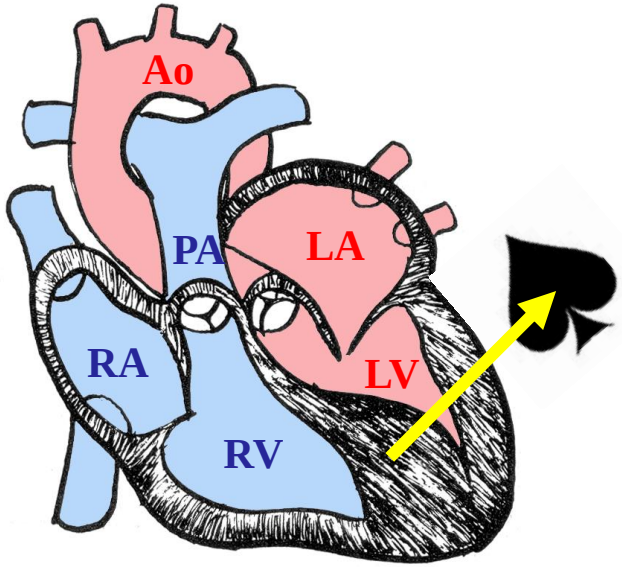


NON OBSTRUCTIVE FORMS

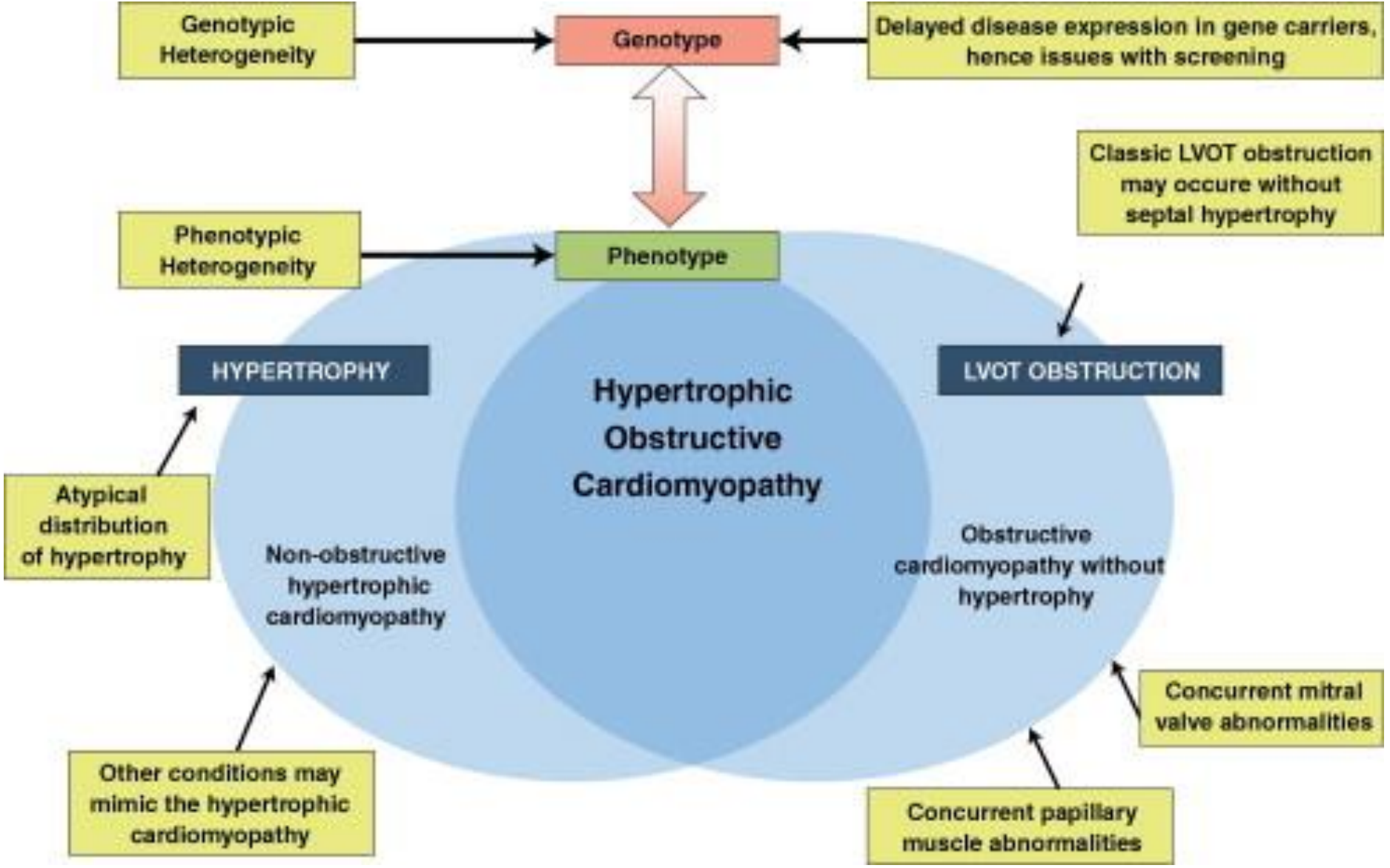
Symmetrical



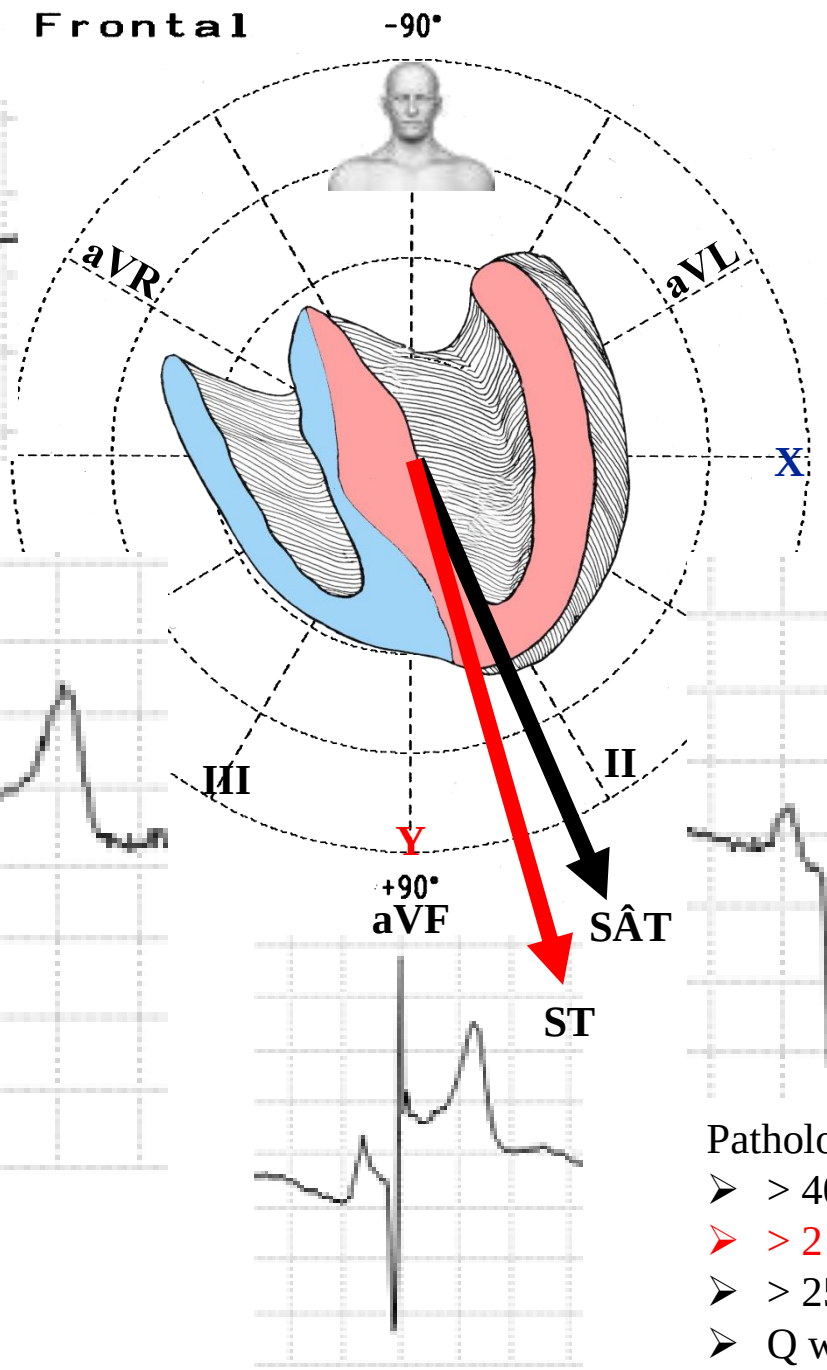
Apical HCM (Ap-HCM)

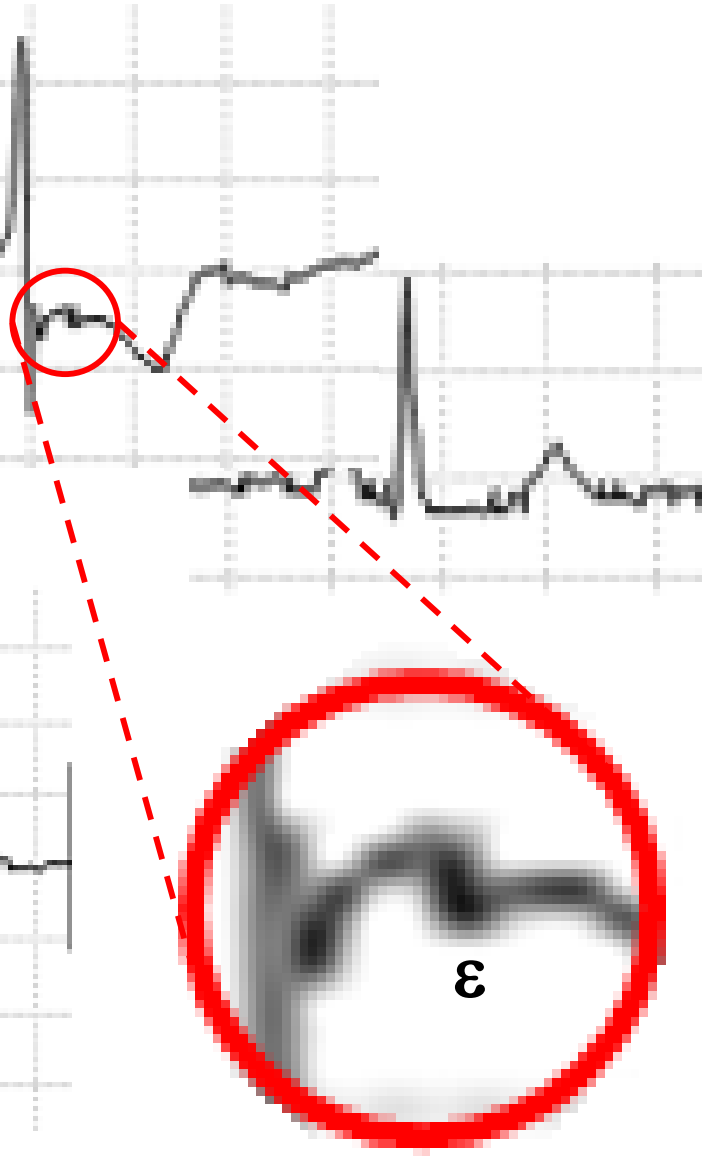


The "ace-of-spades" sign



This figure demonstrates the potential difficulties in diagnosis of hypertrophic cardiomyopathy (HCM) due to phenotypic and genotypic heterogeneity. Patients within the same family might have different phenotypic expressions, ranging from gross hypertrophy with severe left ventricular outflow tract (LVOT) obstruction to minimal hypertrophy and no LVOT obstruction.

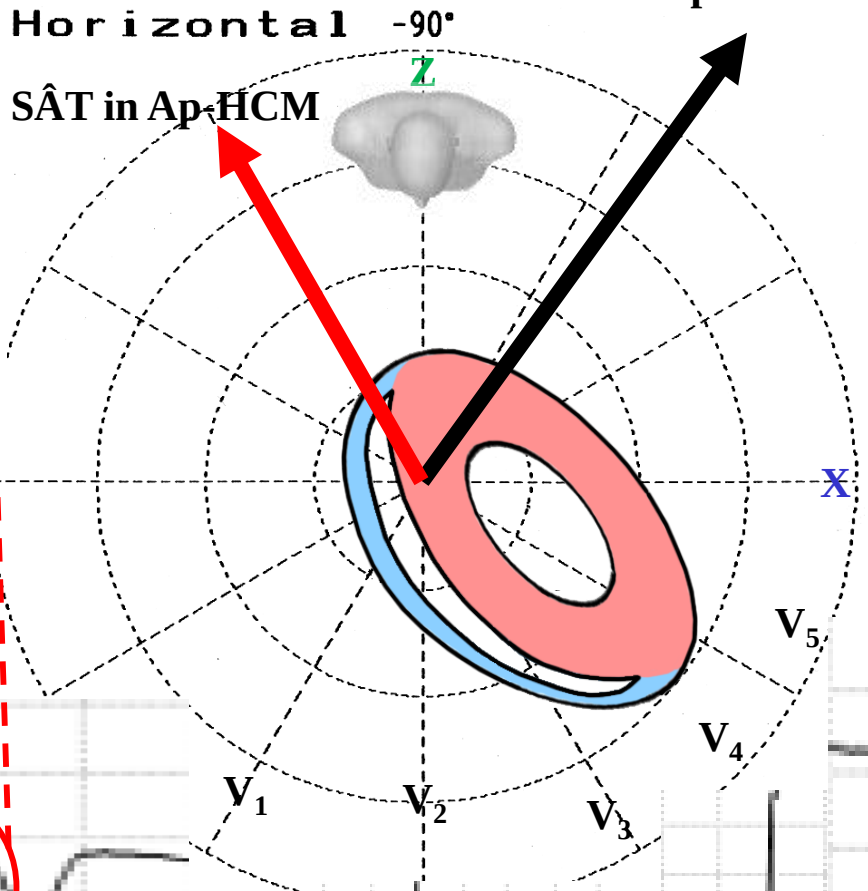


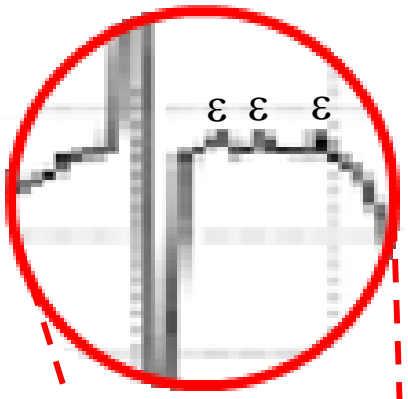


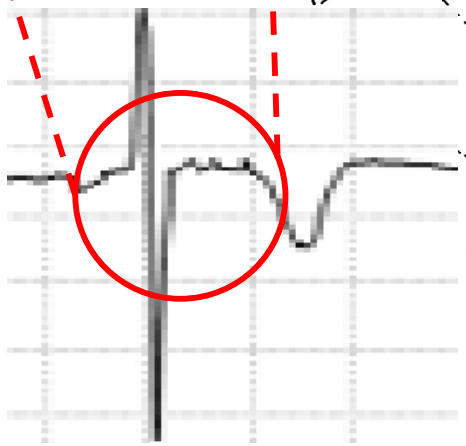
Pathological Q wave criteria:

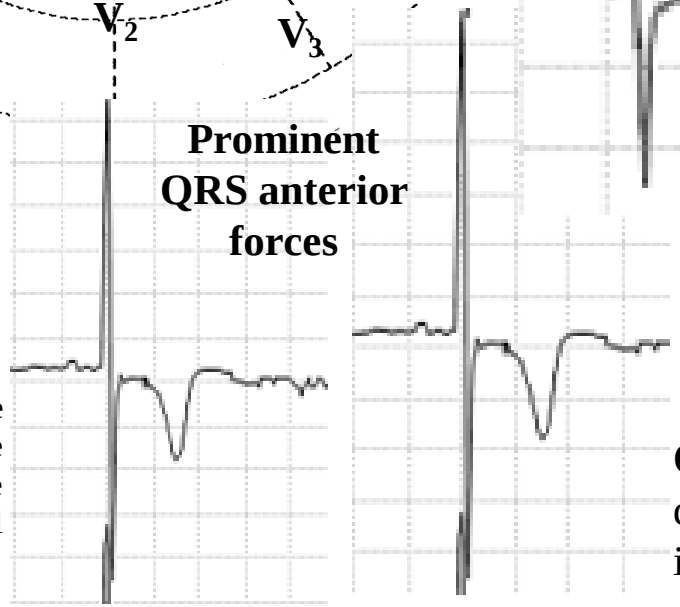
- > 40 ms wide (1 little square)
- > 2 mm deep (the present case has 15 mm!!!)
- > 25% of follow R wave of QRS complex
- Q wave seen in right precordial leads V1-3

SÂT -55°: plus-minus T-wave in V4-V5 (perpendicular)

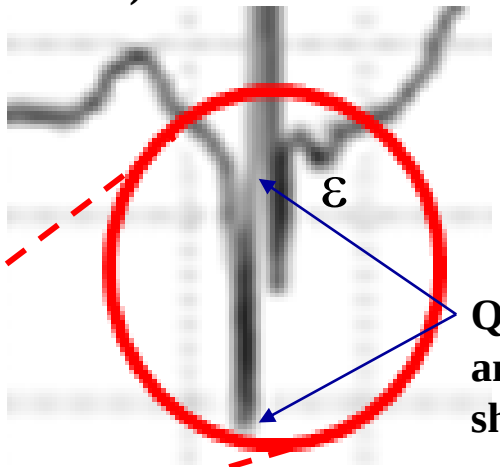




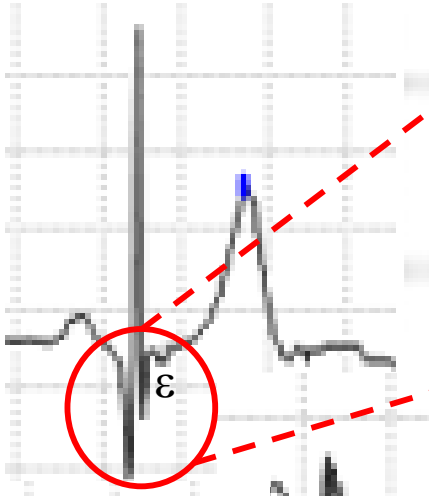


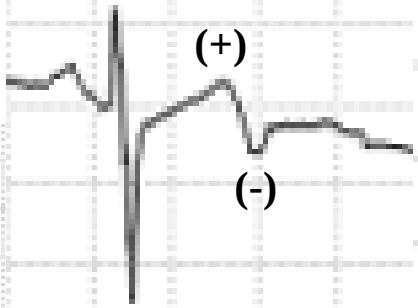


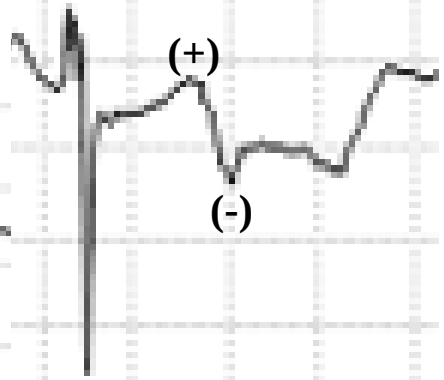
Prominent QRS anterior forces



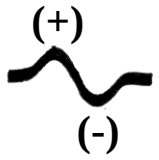
Q = 7 mm deep and narrow sharp Q-wave







Plus-minus T-wave



Are these "epsilon" waves consequence of scars on the ventricular wall? We think yes, because scars are frequent in HCM. Dr Li Zhang think they could be artifacts. The quality of the ECG is poor. Need several repeated ECGs to validate.

Observation: In this case, T axis is directed to back and leftward, differently from apical japanese HCM (Ap-HCM) in which T axis is directed to back and rightward. See next 2 slides.

Dear Andrés:

It was so nice to see you in Brazil!

My first impression regarding ECG was HCM with signs of biventricular hypertrophy and possible right atrial enlargement.
Such deep negative T waves are characteristic for HCM.
I do not see epsilon wave in this ECG, maybe another ECG without noise could be obtained.
I believe it is just noise.

Best regards

Wojciech Zareba M.D.Ph.D.

Wojciech Zareba, MD, PhD, is a Professor of Medicine (Cardiology), Director of Cardiology Clinical Research, and Director of the Heart Research Follow Up Program at University of Rochester Medical Center, Rochester, NY. Dr. Zareba is Principal Investigator or Co-PI on several NIH and corporate grants focused on risk stratification of cardiac death and on clinical usefulness and prognostic significance of ECG parameters. He also serves as Principal Investigator of the ECG Core Labs for numerous studies including the International LQTS Registry, the North American ARVD Registry, MADIT II, and MADIT-CRT, large clinical trials testing clinical effectiveness and safety of implantable cardiac defibrillators and resynchronization devices.

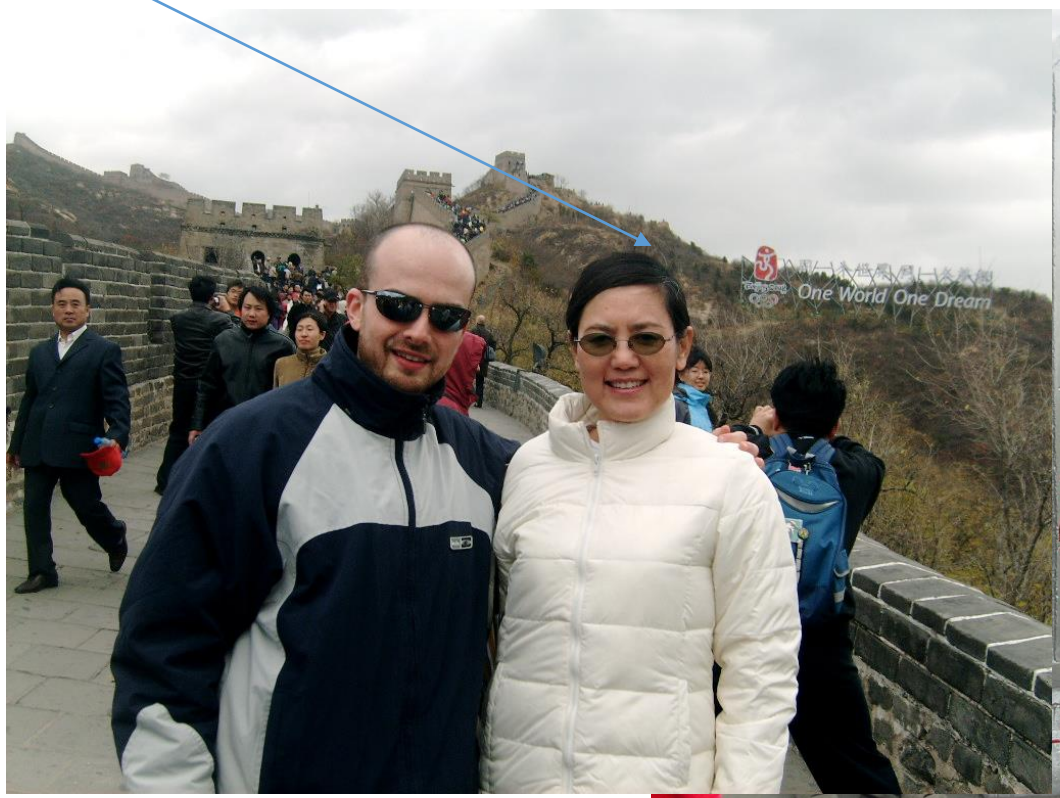
Dear Andrés,

I was traveling this weekend, just got to respond emails.

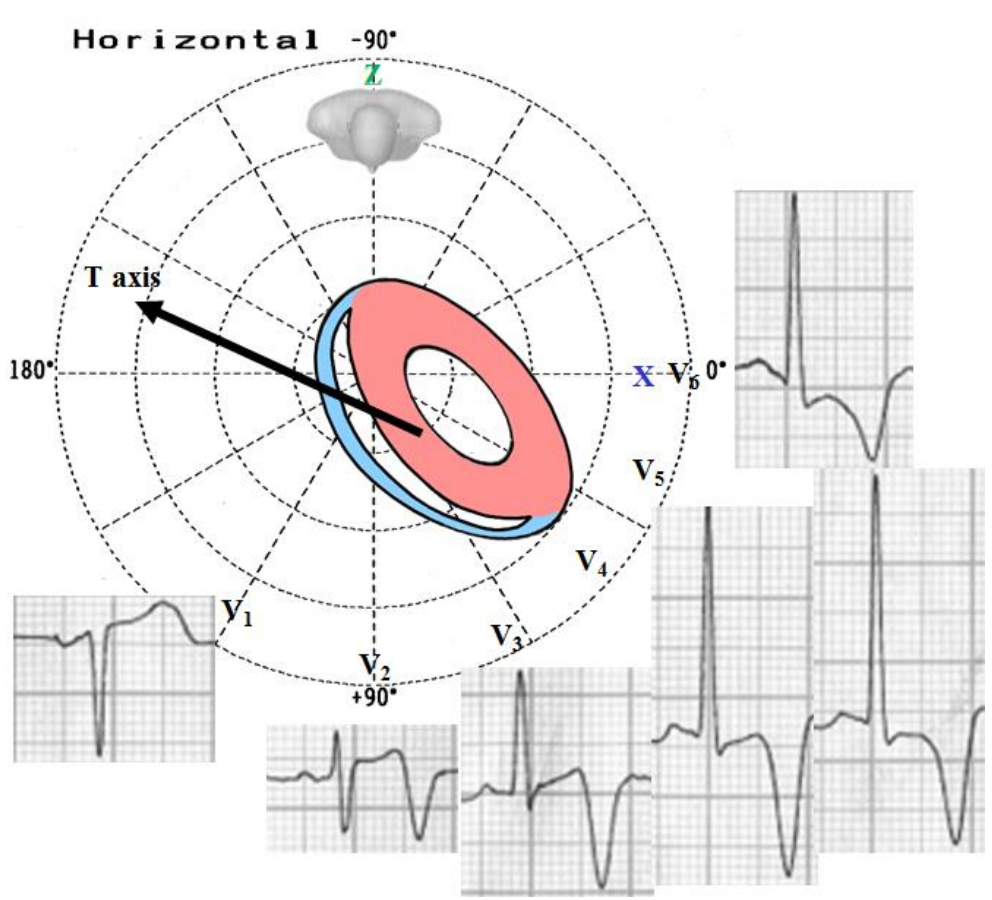
The big picture of this ECG tells me nothing but HCM.
I would not jump to the conclusions for the so-called epsilon waves.
They could be artifacts. The quality of the ECG is poor. Need several repeated ECGs to validate.

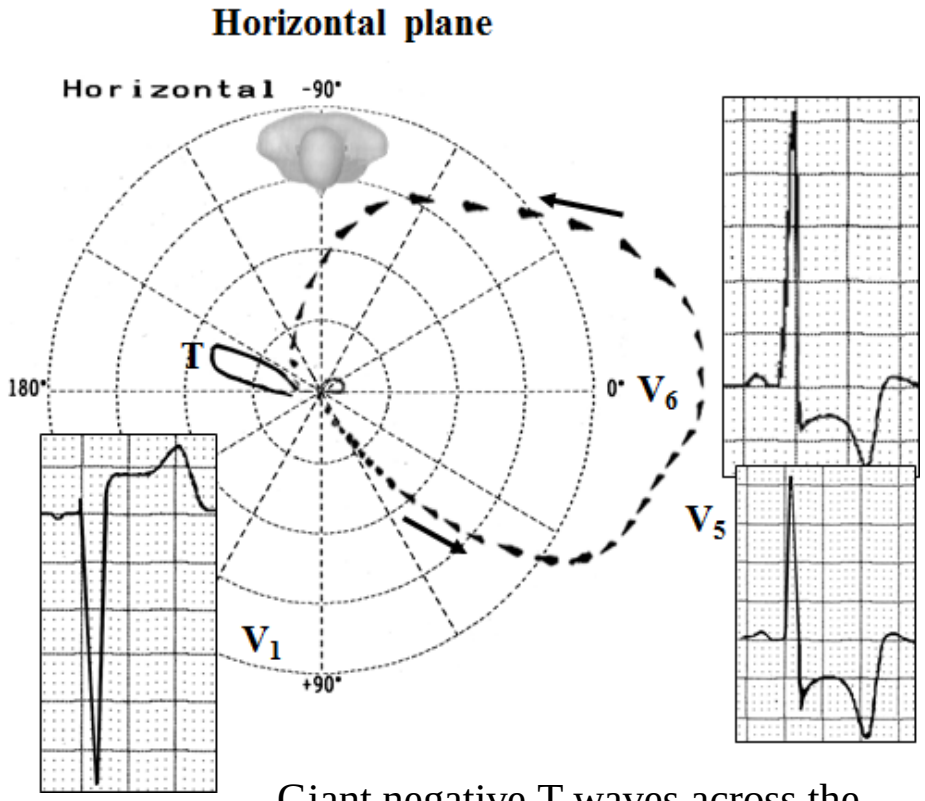
Thank you so very much!
Li Zhang M.D.

We on the Great Wall carries a considerable part of Chinese culture. It has long been incorporated into Chinese mythology and symbolism.



Ap-HCM with T wave axis/T-loop directed to back and rightward

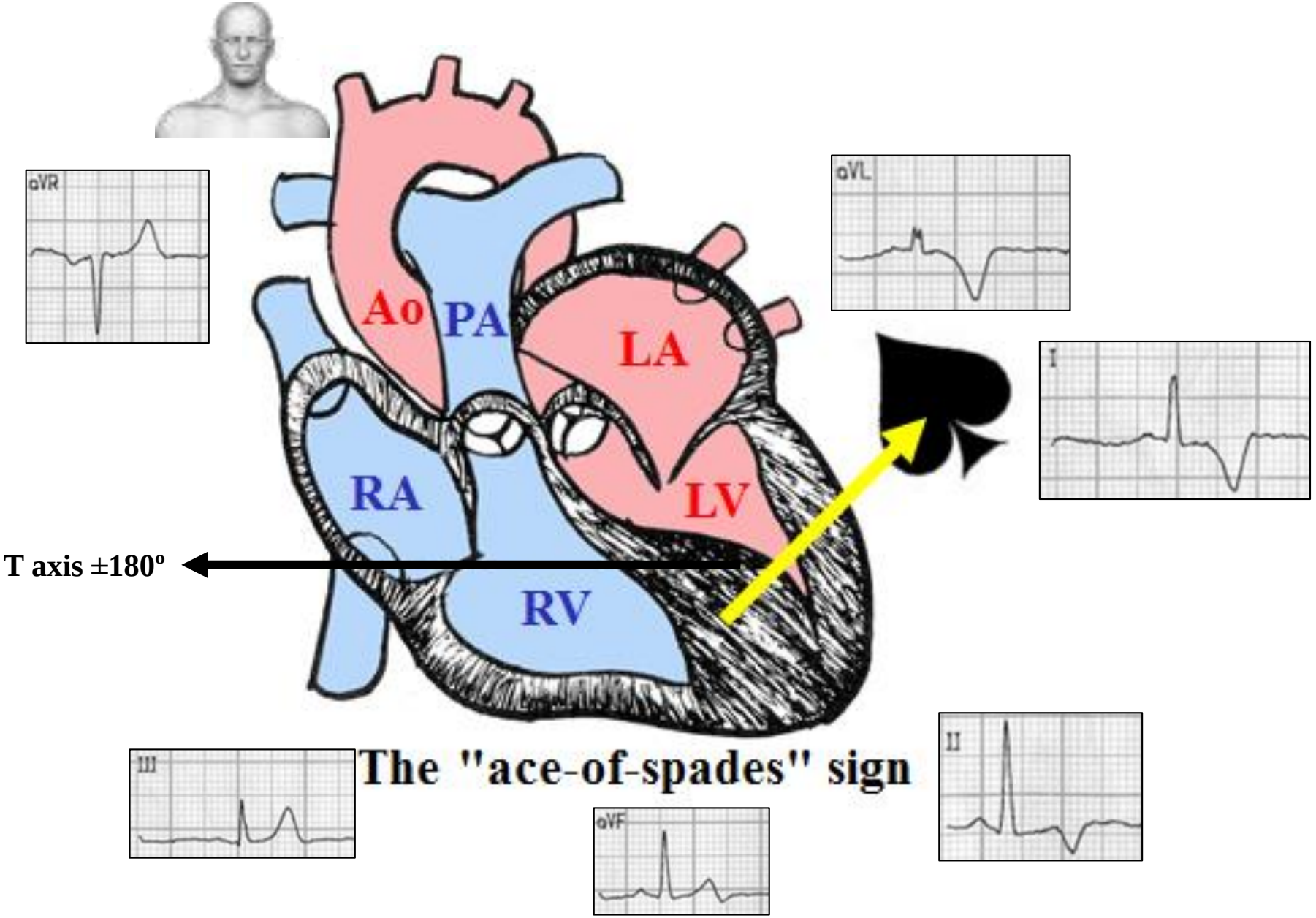




Giant negative T waves across the precordial ECG leads from V2-V6

- Initial vectors of QRS loop heading forward and to the left;
- Anteriorization of QRS loop predominantly located in the left anterior quadrant;
- Maximal vector that increases voltage;
- Final vectors located to the right and backward with ST/T vector in the right posterior quadrant;
- E point that does not match the 0 point and is located backward and rightward from the latter.

Apical hypertrophic cardiomyopathy (Ap-HCM)



Ventriculography shows spade-like morphology, typical of Ap-HCM.

Epsilon waves (ε) or Fontaine waves (**Fontaine 1984**) are late potentials with low amplitude and short duration oscillations near the J point (immediately after) considered a major criterion for Arrythmogenic Right Ventricular Cardiomyopathy/Dysplasia (ARVC/D).

"Fontaine discovered and named the epsilon waves. He chose the epsilon because it follows delta in the Greek alphabet and is the mathematical symbol for smallness" (Hurst 1998).

I. **Intrinsic features** they are small notches or oscillations in variable quantities (1, 2, 3 or more).

II. **Location**: at the end of QRS in the J point or onset of ST segment (there is no consensus about this).

III. **Leads**: observed in the right precordial leads (V1 to V3) (**Marcus 2010**); however Dr. Li Zhang et al, found the ε wave in the leads of the frontal plane, especially in inferior leads. Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave).

IV. **Frequency in ARVC/D:** approximately 15-30% of cases in 12-lead ECG. This percentage increases if we use the ECG with the modified protocol.

V. **Value of criterion:** considered to be a major criterion for diagnosis by the Task Force for ARVC/D diagnosis (**McKenna 1994; Fontaine 1999; Marcus 2010**).

VI. **High resolution ECG:** observed more frequently with this method.

VII. **Pathognomonic character**: in spite of the characteristics in ARVC/D, they are not pathognomonic, since they have been described in other diseases associated with myocardial damage:

- Right ventricular infarction (RVI) (**Andreou 2012**). In this case, the more sensitive lead is V_4R, because STSE is observed with high sensitivitiy in this accessory lead. The ECG pattern of RVI, ST-elevation in lead V4R and in anterior chest leads V1-3 is similar to that of a proximal occlusion of a small, nondominant right coronary artery (RCA). The ECG changes may be misinterpreted as signs of infarction of the anteroseptal wall (**Eskola 2007**);
- Inferior or ancient dorsal MI (**Zorio 2005**),
- Sarcoidosis (**Santucci 2004**),
- Sickle cell anemia (**Hurst 1998**),
- **Myocardial hypertrophy, such as the present case, and**
- Infiltrative diseases.

VIII. **Meaning:** late posterior potentials (PP) that occur in the RV free wall in patients with ARVC/D. These waves represent a post-excitation phenomenon: delayed activation of "islands" of viable RV myocytes interspersed in myocardium that does not depolarize normally.

IX. Inversion of T wave in leads V1-V3 and/or **ε** wave found in 70% of patients with ARVC/D. Epicardial electrophysiological studies in dysplastic areas reveal the LP that occur at the end of the QRS complex, in the J point, and at the onset of the ST segment, explained by fibro-fatty substitution of myocardial tissue (**Fontaine 1984**).

X. **Epsilon wave and relationship to VT:** the simple presence of these waves indicate slow and fragmented conduction, which favors reentry circuits, which in turn result in M-VT runs with CLBBB morphology by originating in the RV (**Aldakar 1998; Sajeev 2004**).

Modified protocol to obtain ECG in patients with suspicion of ARVC/D

The tracing should run at a double velocity (50 mm/s) and double voltage (20 mm/s) to compare the duration of QRS complexes (QRSd) in different leads, as well as to try to record Epsilon waves.

The Fontaine bipolar precordial leads

The tracing should be obtained from I and aVF at double velocity and amplitude, placing the electrode of the left arm on the xiphoid appendix, the one from the right arm on the manubrium sternum, and the one from the left leg on the rib at the fourth or fifth space with the aim of improving the ability to detect Epsilon waves.

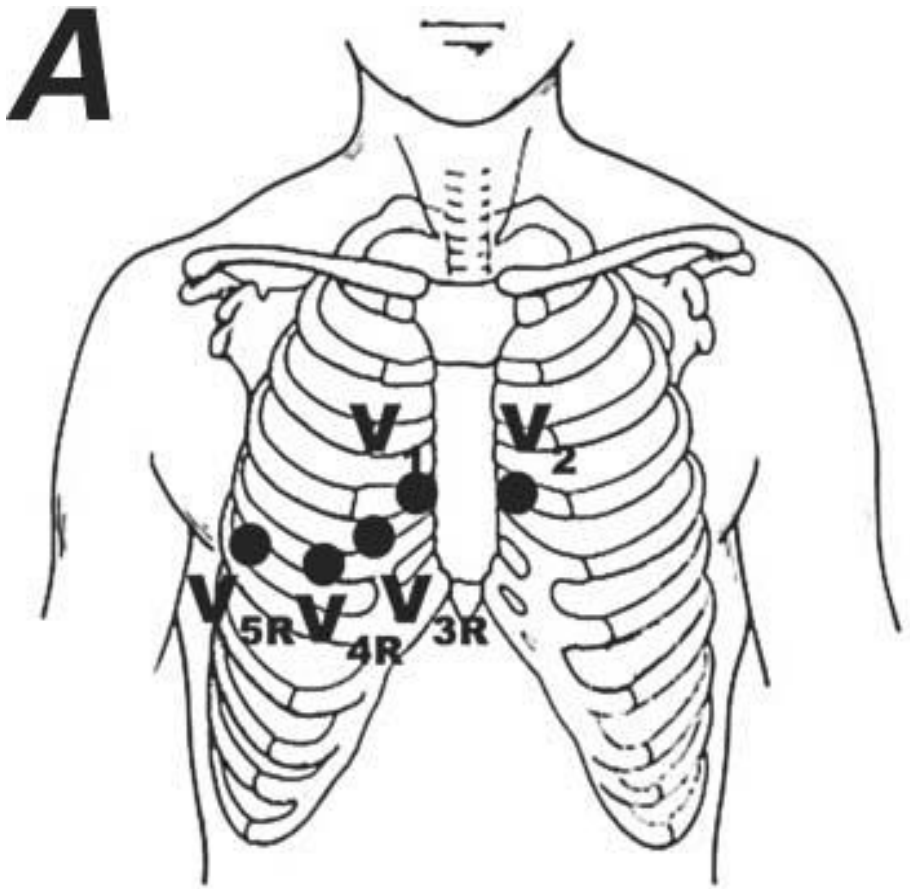
The Fontaine bipolar precordial leads are placed at the manubrium of sternum, xiphoid, and V4 positions using the right arm connection, left arm connection, and left foot connection, respectively.

Epsilon waves are detected by:

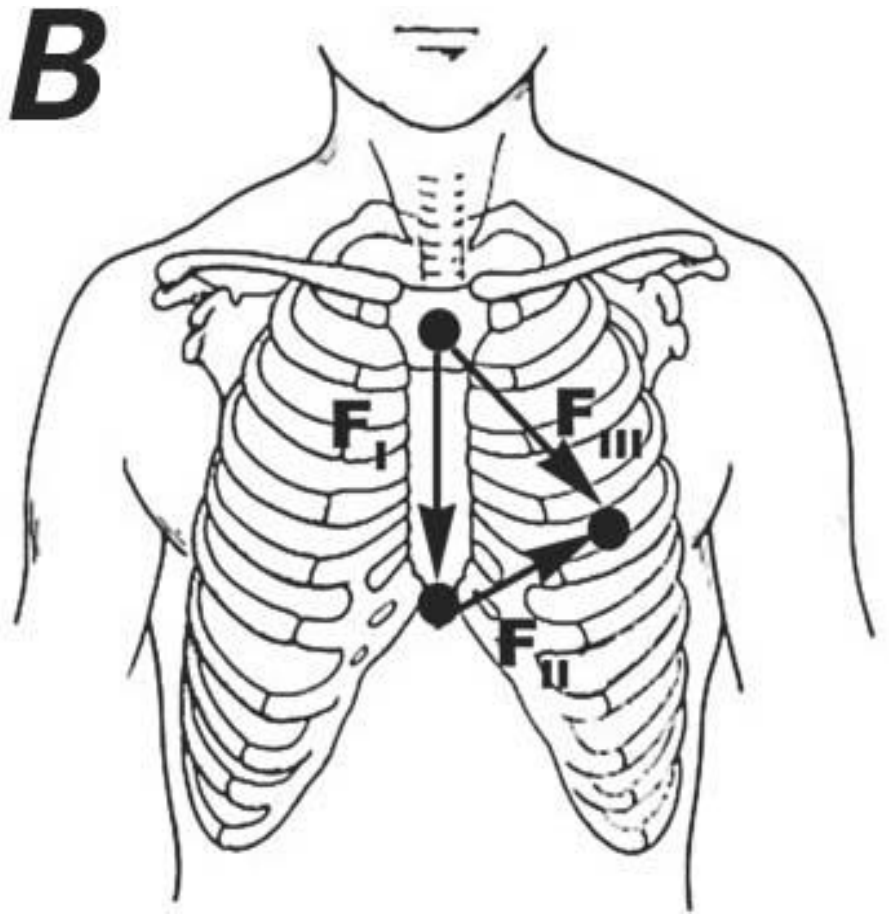
1. Standard 12-lead electrocardiography (S-ECG)
2. Right-sided precordial lead electrocardiography (R-ECG)
3. Fontaine bipolar precordial lead electrocardiography (F-ECG) (**Gottschalk 2014; Peters 2014**). The presence of epsilon waves by the Fontaine lead system provided a high degree of suspicion for ARVC/D (**Chiladakis 2010**).

The detection rate using combined methods is significantly higher than that by S-ECG alone.

Fontaine bipolar precordial lead have the best sensitivity among the three options. the placement of the foot lead (positive) in position V4 provides, instead of regular leads I, II, and III, three bipolar chest leads that can be called FI, FII, and FIII. Tracings are then produced by setting the machine on regular leads I, II, and III. This arrangement is used to record specifically the potentials developed in the RV, from the RVOT to the diaphragmatic area. The vertical bipolar lead FI, (similar to aVF lead), seems to be the most appropriate to record epsilon waves; it also magnifies the atrial potentials. As late potentials were supposed to be the result of late activation of a limited group of fibers, the term "post-excitation" looked logical, since it was observed after the main excitation of the ventricle, leading to the QRS complex. The term "epsilon“ was appropriate, because it occurs in the Greek alphabet after delta; thus, delta represents the preexcitation and epsilon the post-excitation phenomenon.

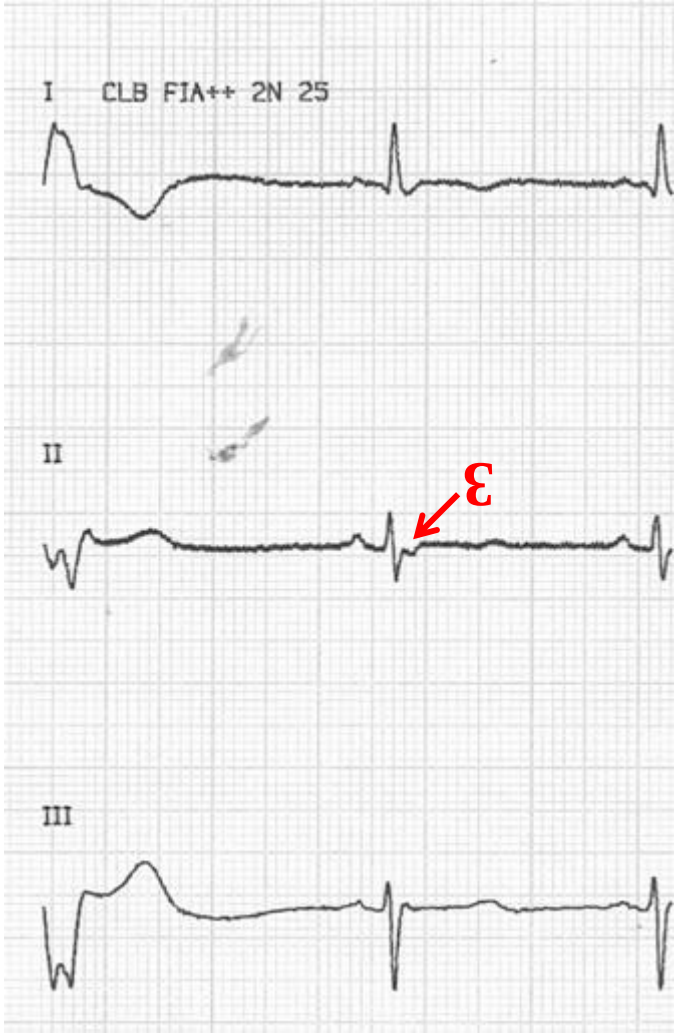


Right precordial leads



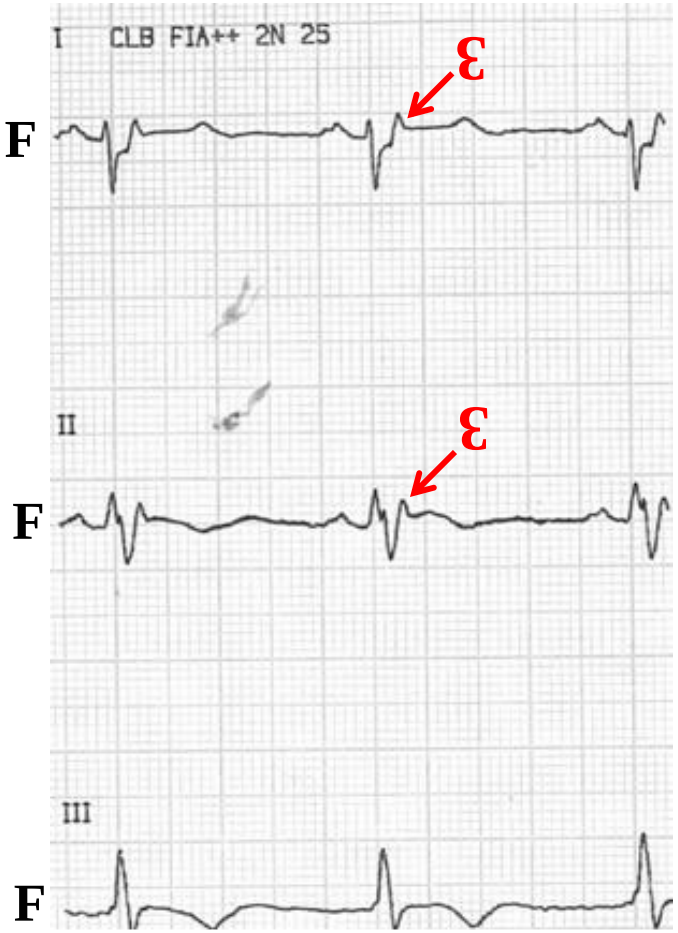
The Fontaine bipolar precordial leads

Traditional bipolar leads



2012

Fontaine's bipolar leads



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