Portuguese / English

Reporte de caso

I. Anamnese

- Identificação: Paciente masculino de 58 anos.
- Queixa principal: dor retroesternal de caráter opressivo, não irradiado e associado a sudorese fria e síncope.
- Antecedentes pessoais patológicos: hipertensão arterial em tratamento irregular.
- Antecedentes familiares: um irmão teve morte súbita com apenas 2 anos e 2 primos de primeiro grau tiveram morte por arritmia cardíaca. Causa?

II. Exame físico

Ao exame físico com extremidades frias; PA=100/60mmHg.
A. cardíaca: ritmo cardíaco regular com FC = 180bpm
Pulmões limpos. Extremidades mal perfundidas.
ECG1 e ECG2 realizados na admissão (figuras 1 e 2).
ECG3 realizado após infusão de amiodarona EV (figura 3).

Perguntas:

1. Qual o diagnóstico dos ECG 1-2-3?
2. Qual o diagóstico clínico?

Case report

I. Anamnesis

- Identification: Male, 58 years old.
- Main complaint: retrosternal pain oppressive non-irradiated and associated cold sweating and syncope.
- Personal pathological history: hypertension in irregular treatment.
- Family history: a brother died suddenly with only two years and two first cousins had death from cardiac arrhythmia. Cause?

II. physical examination

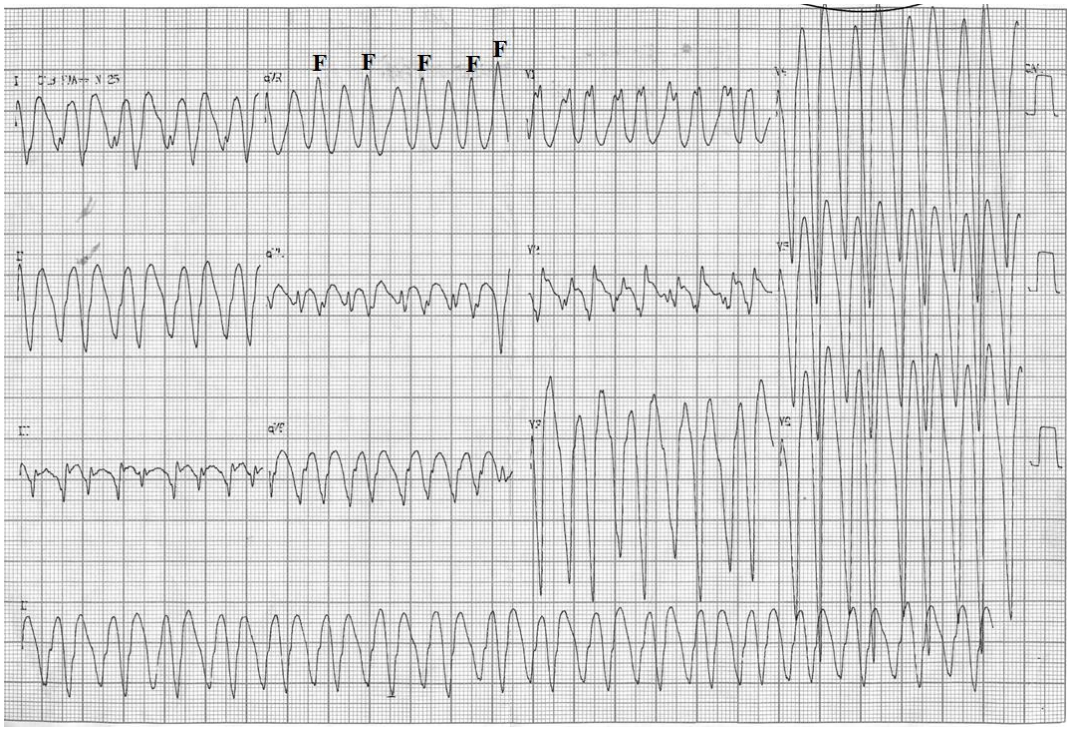
Physical examination with cold extremities; PA = 100 / 60mmHg.
A. Heart: regular heart rate to 180 bpm HR =
Her lungs were clear. Poorly perfused ends.
ECG1 and ECG2 performed on admission (Figures 1 and 2).
ECG3 amiodarone performed after IV infusion (Figure 3).

Questions:

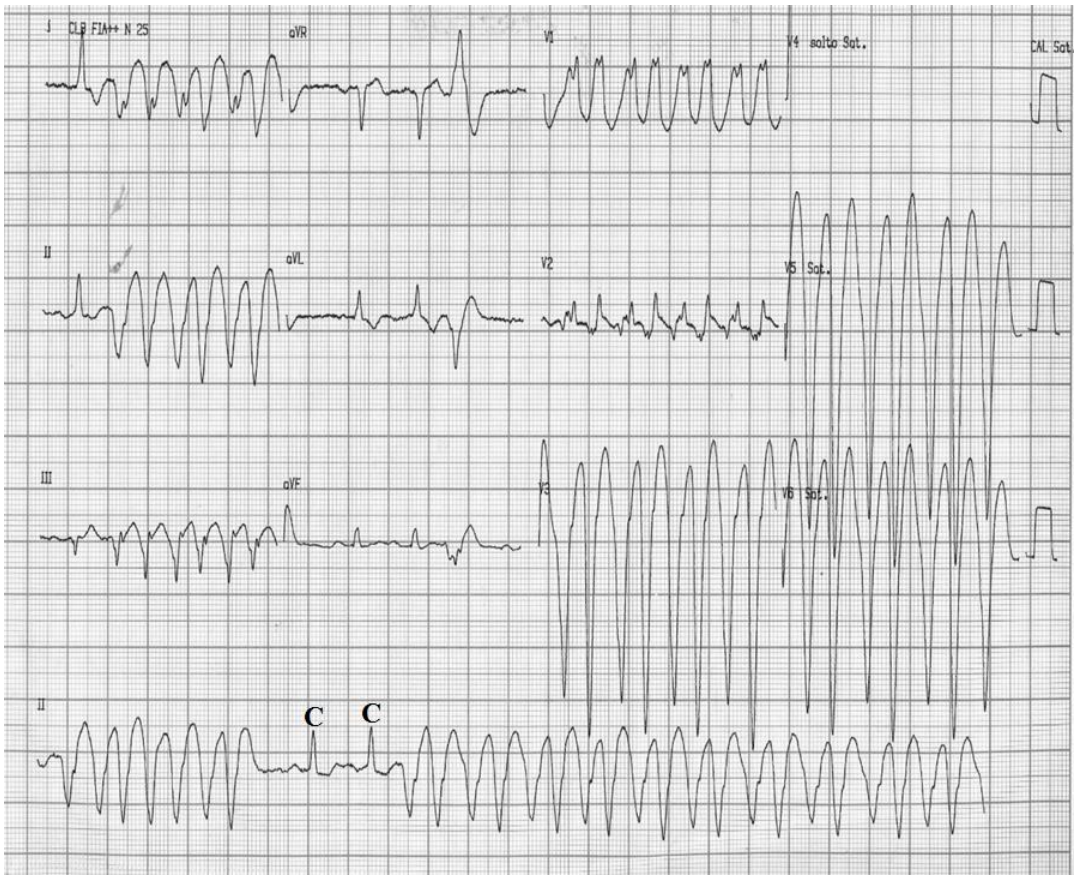
1. Which is the ECG1, ECG2 and ECG3 diagnosis?
2. Which is the clinical diagnosis?

Andrés / Raimundo / Frank

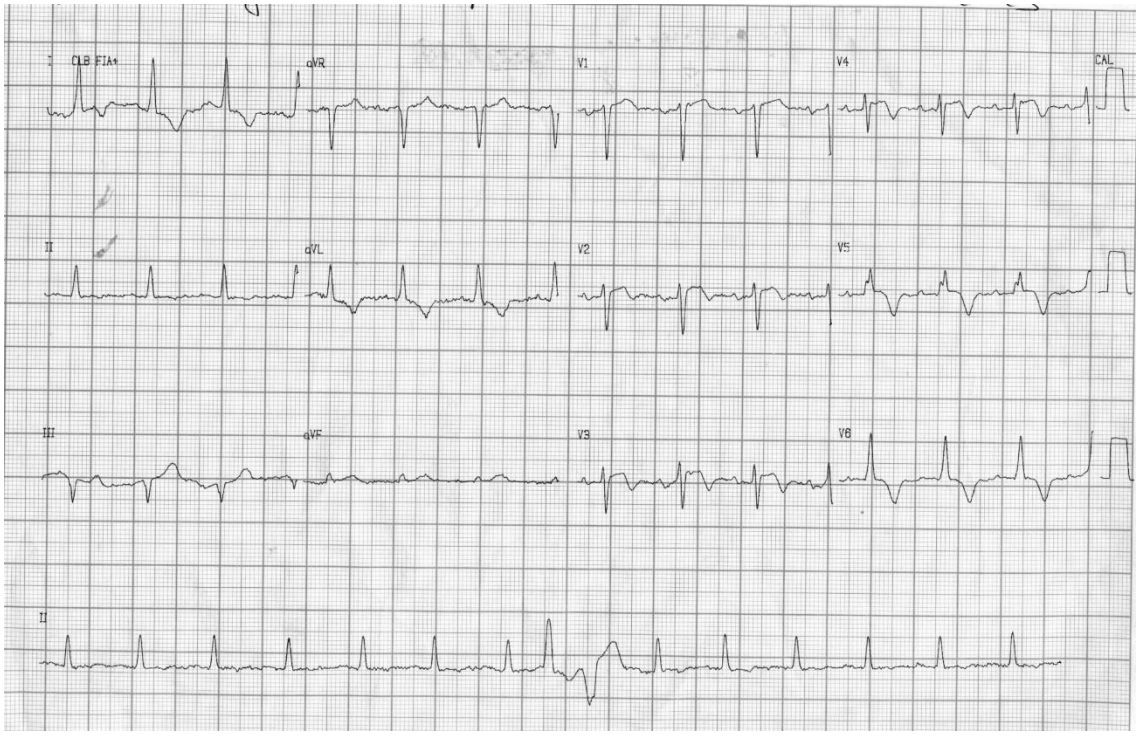
ECG1



ECG2



ECG3 after cardioversion



Colleagues' opinions

I have to recognize that I have no problem with the ECG diagnosis of this irregular wide QRS tachycardia. This is a VT.
The most interesting issues are:
1) is there any cardiac disease behind it.
2) is there any relationship with the familial story of sudden death.
Before cardiac work up including exercise ECG, echocardiogram, coronary angiography and MRI are performed, almost all the textbook pathology can be considered.
Of course since the patient is not living in Israel but in South America, I would certainly think to CHAGAS disease first.

Prof Bernard Belhassen, Israel

Director, Cardiac Electrophysiology Laboratory Professor of Cardiology, Sackler School of Medicine, Tel-Aviv University

Hi
It seems monomorphic VT in the context of an anterior MI more likely proximal to middle third of LAD.
VT originates in the Infero-lateral aspect of the LV.
The strong family history of sudden death suggest the possibility of other diagnosis than CAD, and in this sense, imaging would help to determine the presence of structural heart disease such as apical HCM, or other rare forms of cardiomyopathy. By the electrical presentation alone, is not much what I can say: no QT prolongation, no Brugada, no evidence of CPVT.

So I am expecting something rather curious in the images requested, let's see...

Adrian Baranchuk, M.D. FACC FRCPC

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Looks like acute antero lateral MI probably due to LAD occlusion with V. flutter from infer,lateral apical LV.Hard to correlate with other sudden deaths in family do they have familial hypercholestrolemia?

Melvin Scheinman

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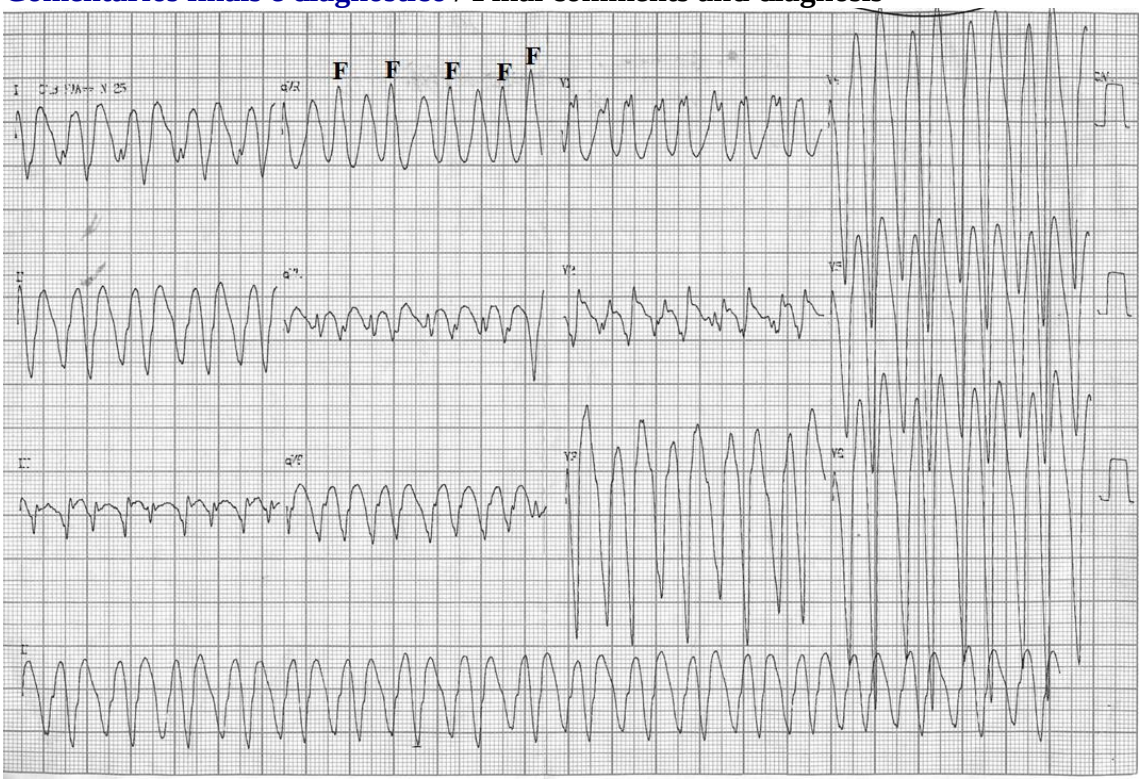
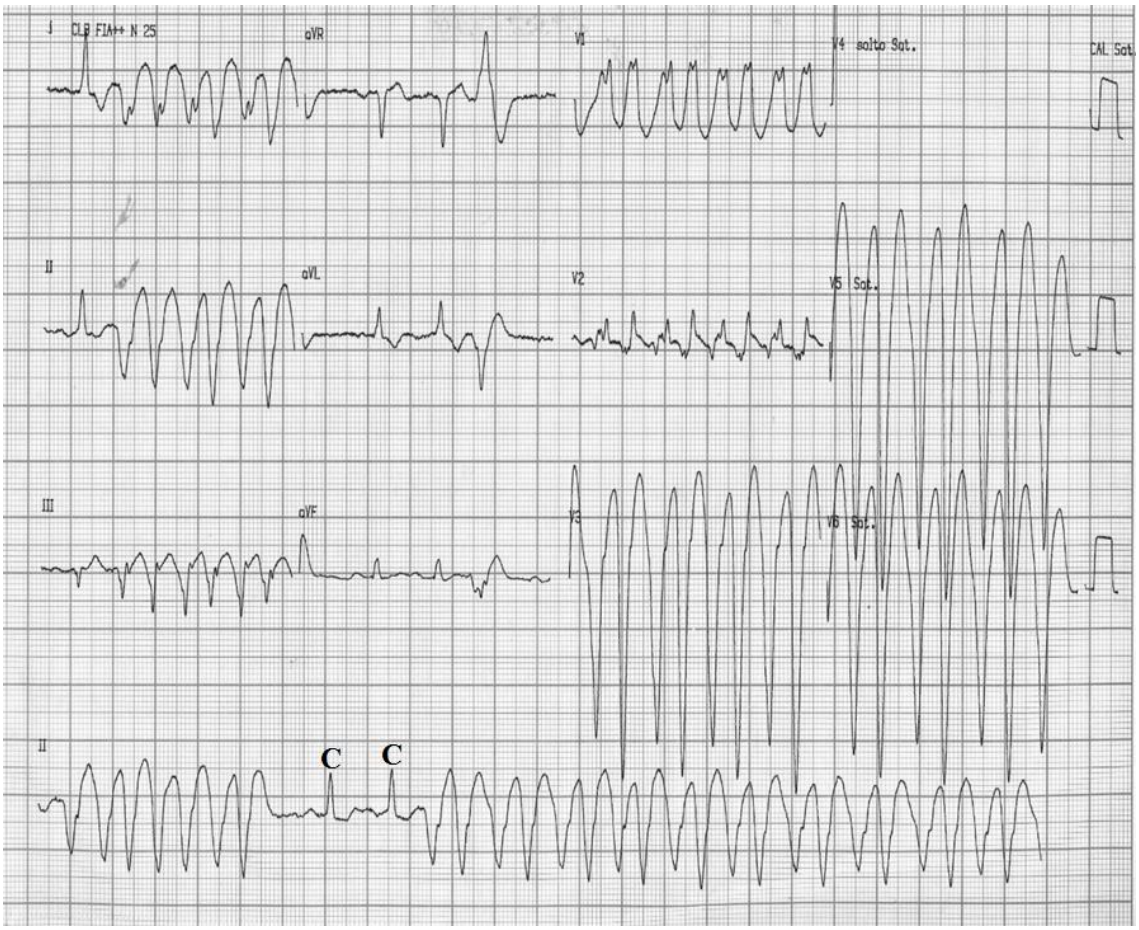
Comentarios finais e diagnóstico / Final comments and diagnosis



ECG1 Figura 1 . Mostra uma taquicardia de QRS largo, FC elevada e irregular (variando de batimento a batimento de 188-250 bpm), SÂQRS localizado no quadrante superior direito (única derivação positiva no plano frontal aVR) entre –90º e ± 180º "no man's land" ou "northwest axis", distância do início do QRS ao nadir do S > 100 ms (sinal de Brugada), ausência de padrão "RS" nas precordiais, complexos do tipo QS em I e V6, morfologia tipo padrão de BRD bifásico em V1 "bad rabbit" com o primeiro ápice de ≤ voltagem do que o segundo "rabbit ear signal" (1) e presença de nítidos batimentos de fusão (QRSs híbridos) de grau variável (F), tudo compatível com TV.

ECG1, Figure 1: It shows wide-QRS tachycardia, high and irregular HR (varying from beat to beat within 188-250 bpm), SAQRS located in the right upper quadrant (single positive lead in the frontal plane aVR) between -90° and ±180°, "no man's land" or "northwest" axis, distance from QRS onset to the nadir of S > 100 ms (Brugada sign), absence of RS pattern in precordial leads, QS complexes in I and V6, morphology of biphasic RBBB pattern in V1, of the "bad rabbit" type, with the first apex of ≤ voltage than the second "rabbit ear sign" (1) and the presence of clear fusion beats (hybrid QRSs), in a variable degree (F), all compatible with VT.



ECG2 Figura 2 .Traçado do mesmo paciente mostrando períodos de TVNS. Na tira de II longo, o oitavo e o nono batimentos são batimentos de captura (C)

ECG2, Figure 2. Tracing of the same patient showing NSVT periods. Long in the strip of II, and the eighth and ninth are capture beats (C).

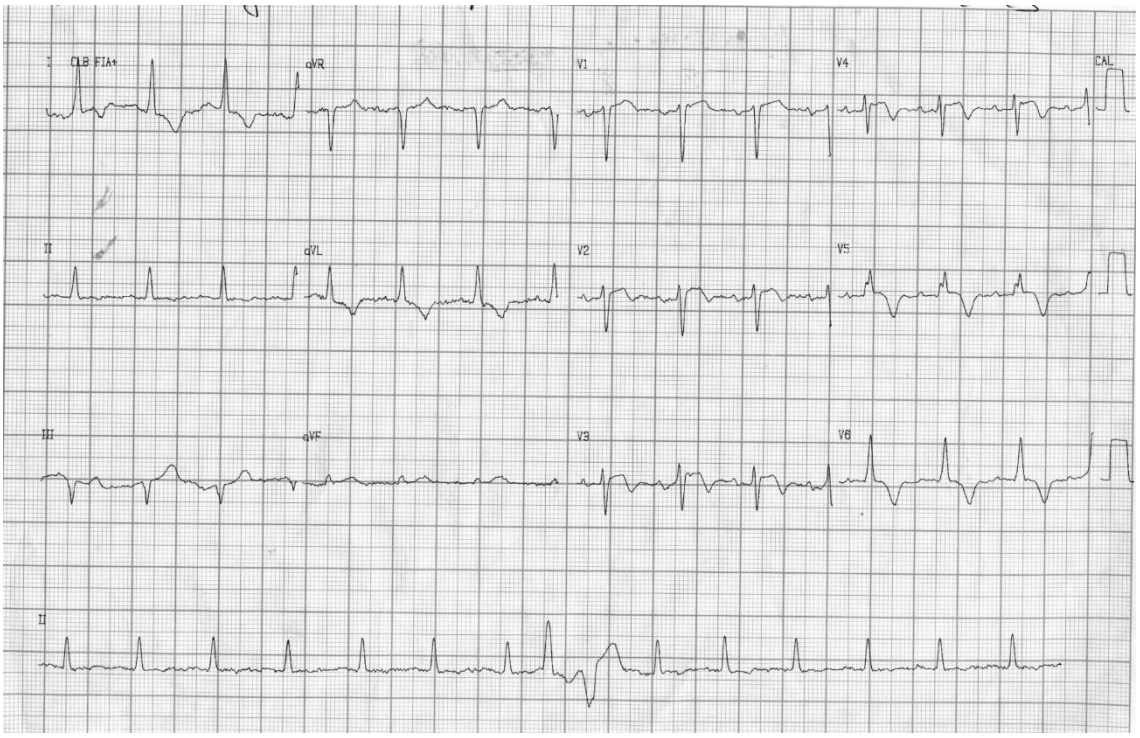
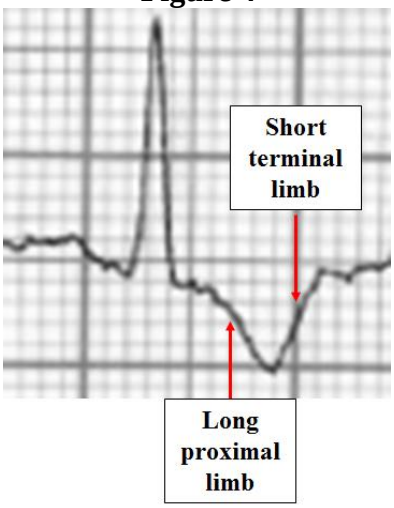


Figura 3.Após a reversão ao ritmo sinusal observa-se inversão da onda T na parede ântero-lateral, discreta elevação do segmento ST em V2-V3 seguido de onda T com padrão plus/minus de V2-V4 (padrão ECG da síndrome de Wellens). As derivações esquerdas de I e V5-V6 mostram um padrão sistólico de repolarização ventricular caracterizado por depressão do segmento ST de convexidade superior seguido de onda T negativa com a porção descendente mais lenta do que a ascendente nas derivações esquerdas "*left ventricular 'strain' pattern"* (figura 4).

Figure 3. After reversion to sinus rhythm, T wave inversion in the antero-lateral wall, discrete ST segment elevation in V2-V3 followed by T wave with plus/minus pattern in V2-V4 (ECG pattern of Wellens syndrome) are observed. The left leads of I and V5-V6 show ventricular repolarization systolic pattern characterized by ST segment depression of upper convexity, followed by negative T wave with slower downsloping portion than the upsloping one in left leads, "left ventricular 'strain' pattern". (Figure 4).

Figure 4



Na figura 4 que corresponde a derivação I do ECG da figura 3 mostra claramente uma onda T negativa **secundária** por apresentar o ramo proximal descendente lento e ascendente terminal mais curto rápido, o que afasta uma alteração de repolarização primária.
O diagnóstico inicial foi de síndrome coronariana aguda (SCA) sem elevação do segmento ST "NSTEMI" e o paciente recebeu as seguintes medicações: nitroglicerina EV, aspirina, clopidogrel, heparina e beta bloqueador. Eletrólitos normais. Após o resultado da troponina elevada (0,112 ng/dl) foi encaminhado para o laboratório de hemodinâmica. O estudo mostrou coronárias normais e a ventriculografia revelou um VE hipertrófico.
Em seguida realizou-se um ecocardiograma transtorácico que revelou os seguintes achados (figura 5):
Diâmetro distólico do VE=46mm;diâmetro sistólico do VE=27mm;espessura distólica do septo inter-ventricular=19mm;diâmetro diastólico da parede posterior do do VE=09mm;AE=26mm;Aorta=44mm;FE=72%;massa do VE=322g;índice de massa do VE=61,42g/m^2
Conclusão:

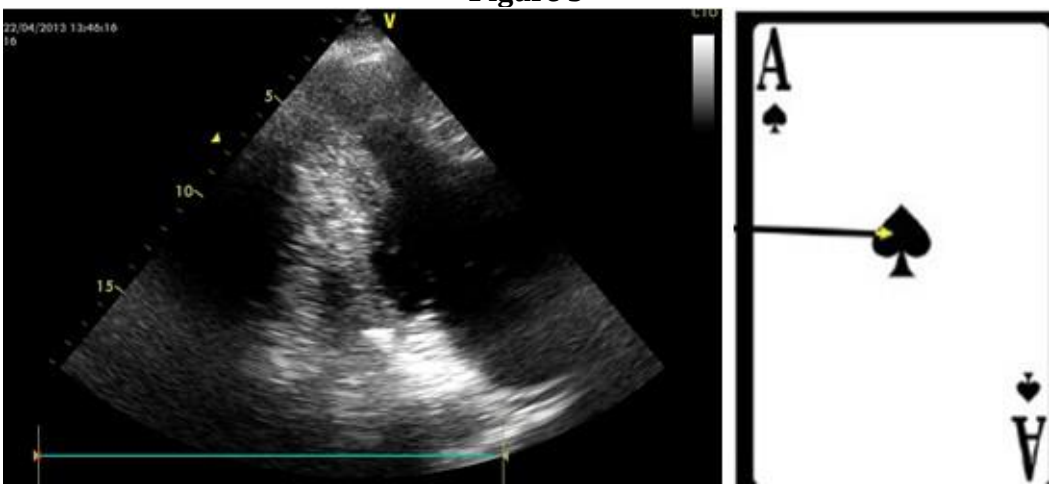
- Aumento do AE e demais câmaras cardíacas normais
- Hipertrofia assimétrica do VE (predominantemente apical do VE, mas com extensão para o septo anterior e posterior), com gradiente sistólico máximo intraventricular de 20mmHg e hipertrofia concomitante do VD
- Anteriorização e hipertrofia dos músculos papilares
- Contratilidade global e segmentar do VE preservadas em repouso
- Déficit de relaxamento do VE
- Ausência de obstrução na via de saída do VD e do VE
- Válvulas cardíacas com aspectos morfodinâmicos normais
- Pericárdio normal
- **EXAME COMPATÍVEL COM MIOCARDIOPATIA HIPERTRÓFICA APICAL**

Figure 4, which corresponds to lead I of the ECG in Figure 3, clearly shows **secondary** negative T wave by presenting slow downsloping proximal limb and shorter and faster upsloping terminal limb, which rules out primary repolarization alteration.

The initial diagnosis was non-ST elevation acute coronary syndrome (NSTEMI) , and the patient received the following medications: IV nitroglycerin, aspirin, clopidogrel, heparin, and beta-blockers. Normal electrolytes. After the result of increased troponin (0.112 ng/dl), he was referred to undergo hemodynamic tests. The study showed normal coronary arteries and the ventriculogram revealed hypertrophic LV.
Next, transthoracic echo was performed, revealing the following findings (Figure 5):
LV diastolic diameter = 46 mm; LV systolic diameter = 27 mm; interventricular septum diastolic thickness = 19 mm; LV posterior wall diastolic diameter = 9 mm; LA = 26 mm; aorta = 44 mm; EF = 72%; LV mass = 322 g; LV mass index = 61.42 g/m^2.
Conclusion:

- LA increase and the rest of the cardiac chambers are normal.
- Asymmetrical LV hypertrophy (predominantly apical in the LV, but with extension into the anterior and posterior septum), with maximal intraventricular systolic gradient of 20 mmHg and concomitant RV hypertrophy.
- Anteriorization and hypertrophy of papillary muscles.
- Global and segmentary LV contractility preserved in rest.
- LV relaxation deficit.
- Absence of RV and LV outflow tract obstruction.
- Cardiac valves with normal morphodynamic aspects.
- Normal pericardium.
- **TEST COMPATIBLE WITH APICAL HYPERTROPHIC CARDIOMYOPATHY.**

Figure 5



O paciente foi submetido a implante de CDI para prevenção secundária da MS e mantido sob terapia medicamentosa com betabloqueador e amiodarona.
The patient underwent ICD implantation as secondary prevention for SCD and was kept medicated with beta blockers and amiodarone.

Resumo dos principais achados eletrocardiográficos na MCH

1. Bradicardia sinusal
2. Padrão de sobrecarga ventricular esquerda: por critérios de voltagem e de repolarização ventricular ("strain pattern") e também pela presença de criterios indiretos tais como SAE e o aumento do tempo de ativação ventricular(deflexão intrinsecóide)
3. Alteração de repolarização ventricular
4. Eventual sobrecarga auricular esquerda ou bi auricular
5. Intervalo PR normal, curto ou prolongado
6. Eventual ondas R de voltagem aumentada nas precordiais direitas
7. Ondas R proeminentes nas precordiales direitas e médias.
8. Pseudo onda delta na porção incial da rampa ascendente do QRS ("slurred QRS upstroke") (2).
9. Eventual presença de padrão de pseudo infarto.
10. A presença de fragmentação do QRS (fQRS) é um marcador de arritmias malignas (3).
11. Eixo elétrico do QRS pode ser normal ou com extremo desvio à esquerda ou à direita.
12. Padrão de bloqueio completo do ramo esquerdo é a regra após a miotomia/miectomia septal transaórtica (cirurgia de Morrow) (4).
13. Padrão de bloqueio completo do ramo direito é predominante (≈70% dos casos) (5).
14. Bloqueio AV completo transitório ou definitivo pode observar-se após a injeção de álcool absoluto na primeira perfurante septal da DA (6).
15. Ondas T gigantes e profundamente invertidas são características da cardiomiopatia hipertrófica apical (7).
16. Prolongamento dos intervalos QT/QTc que está asociado com assincronia mecânica e disfunção do VE (8). O prolongamento do QTc é um preditor clínico de terapia apropiada do CDI na MCH.
17. Episódios de TV monomórficas sustentadas são raros e se observam na forma obstrutiva médio-ventricular asociada à aneurisma apical (9).

Summary of the main electrocardiographic findings in HCM:

1. Sinus bradycardia.
2. Left ventricular enlargement pattern: by voltage criteria and ventricular repolarization (strain pattern) and also by the presence of indirect criteria such as LAE and ventricular activation time increase (intrinsicoid deflection).
3. Ventricular repolarization alteration.
4. Possible left atrial or biatrial enlargement.
5. Normal, short or prolonged PR interval.
6. Possible R waves of increased voltage in the right precordial leads.
7. Prominent R waves in the right and middle precordial leads.
8. Pseudo-delta wave in the initial portion of the upslope of QRS (slurred QRS upstroke) (2).
9. Possible presence of pseudo-infarction pattern.
10. The presence of QRS fragmentation (fQRS) is a marker of malignant arrhythmias (3).
11. QRS electrical axis could be normal or with extreme leftward or rightward shift.

12. Complete left bundle branch block pattern is the rule after transaortic septal myotomy/myectomy (Morrow surgery) (4).
13. Complete right bundle branch block pattern is predominant (≈70% of cases) (5).
14. Transient or permanent complete AV block could be observed after absolute alcohol injection in the first septal perforator branch of the ADA (6).
15. Giant and deeply inverted T waves are characteristic of apical hypertrophic cardiomyopathy (7).
16. QT/QTc interval prolongation associated to mechanical asynchrony and LV dysfunction (8). QTc prolongation is a clinical predictor of appropriate therapy of ICD in HCM.
17. Sustained monomorphic VT episodes are rare and observed in the mid-ventricular obstructive form associated to apical aneurysm (9).

Referencias / References

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