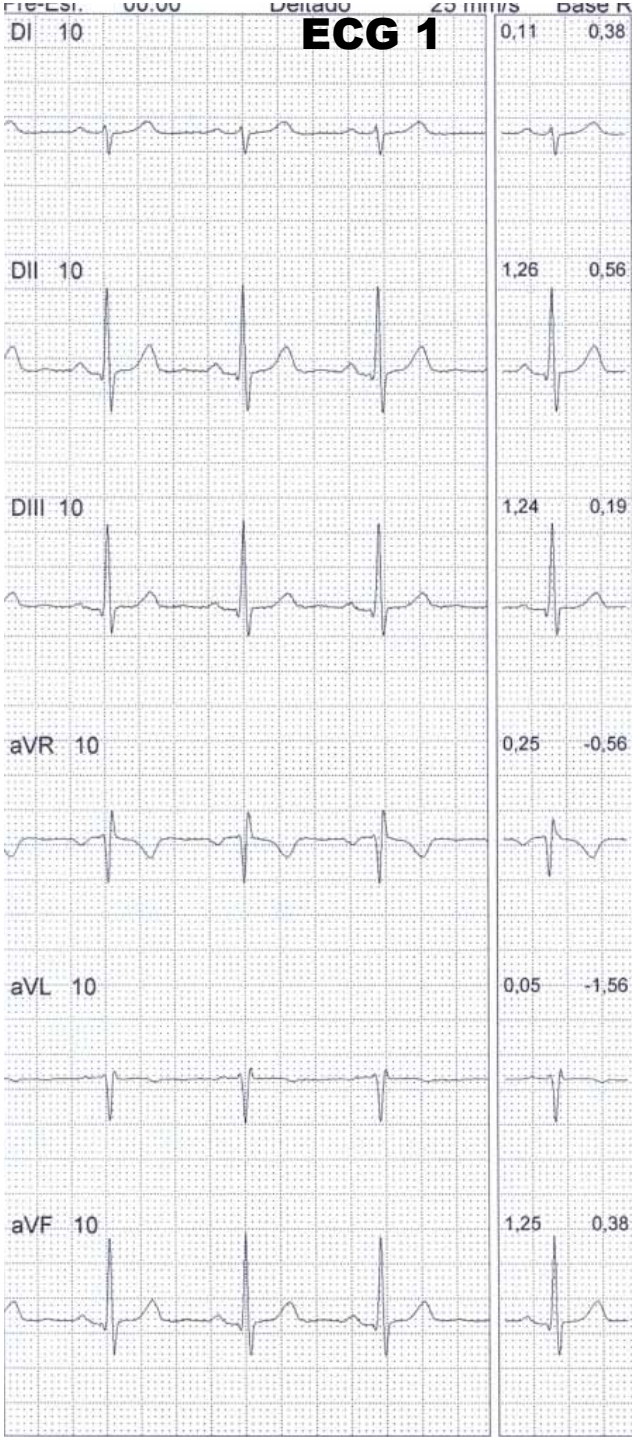
HOMEM JOVEM SEM CARDIOPATIA ESTRUTURAL COM SURTOS DE TAQUICARDIA VENTRICULAR MONOMÓRFICA BEM TOLERADA E INDUZIDA PELO ESFORÇO

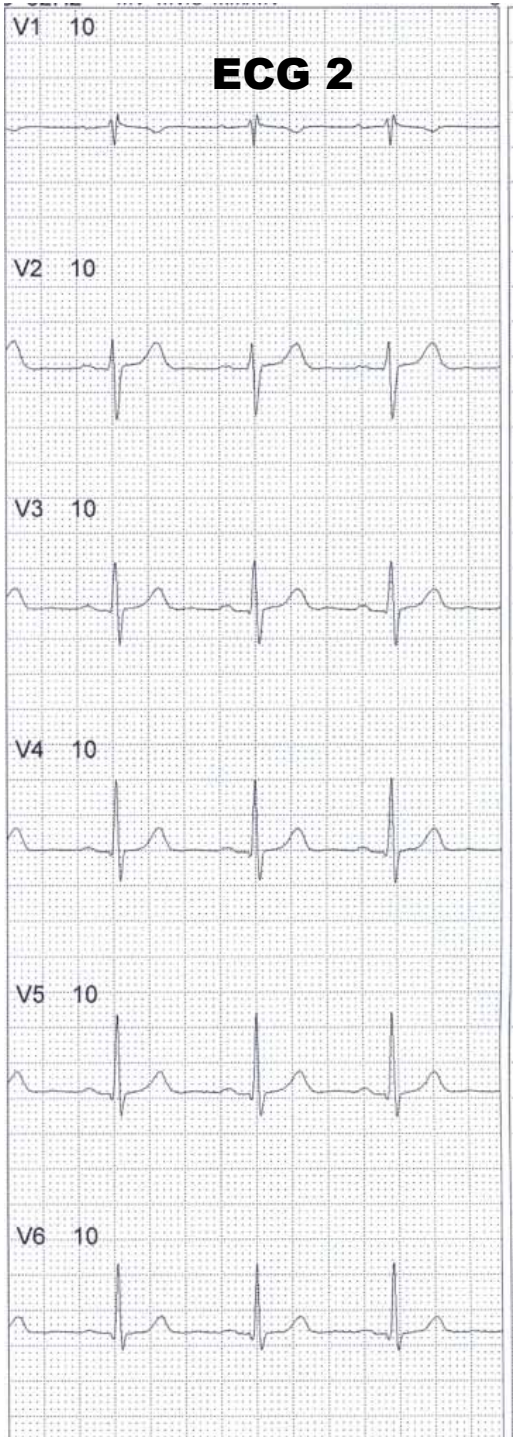
YOUNG MAN WITHOUT STRUCTURAL HEART DISEASE WITH MONOMORPHIC VENTRICULAR TACHYCARDIA INDUCED BY EXERTION AND WELL TOLERATED

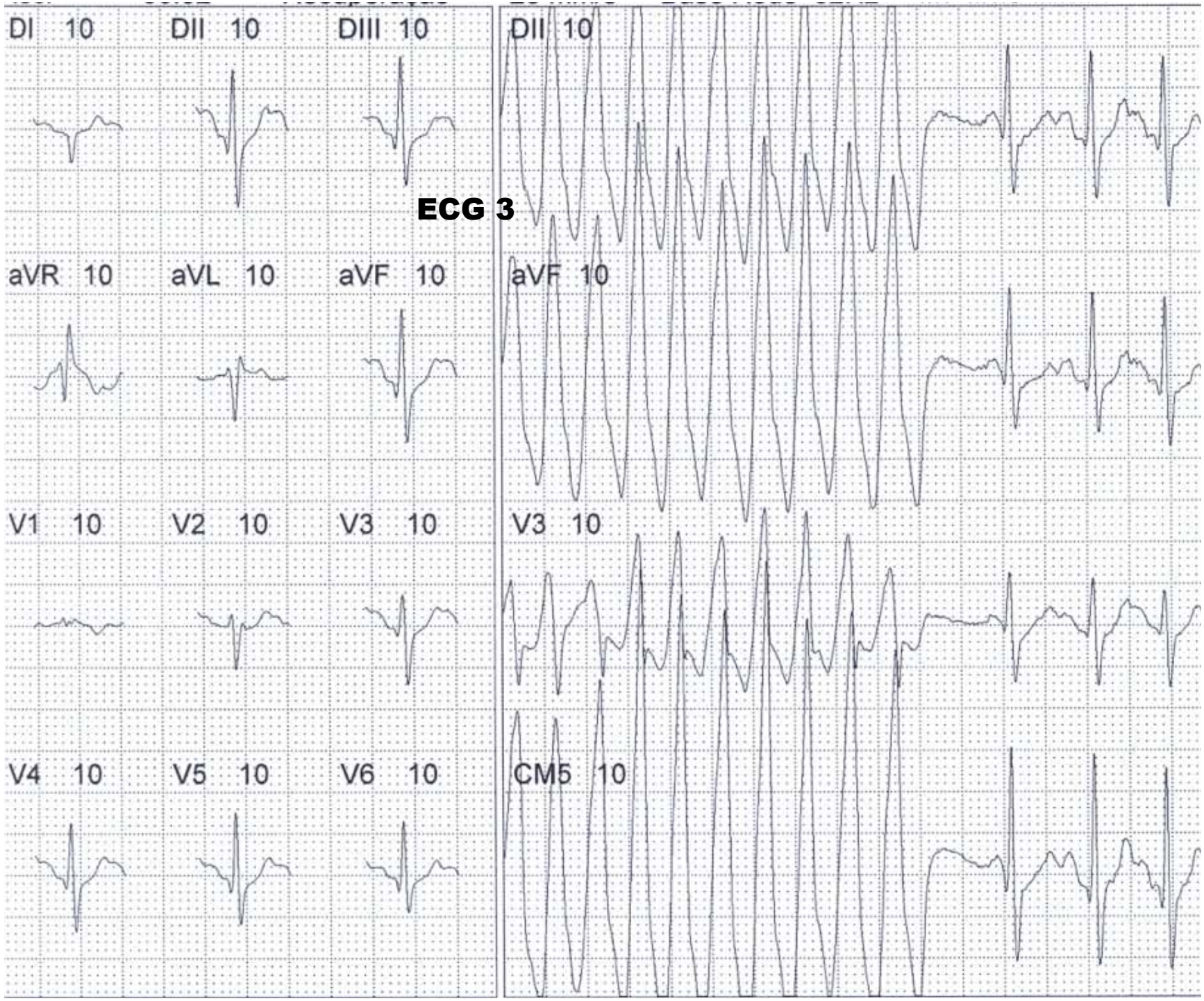
Raimundo Barbosa Barros MD Fortaleza Ceará Brazil

Paciente do sexo masculino, 37 anos, com história de palpitações somente perante esforços físicos.
Nega síncope.
História familiar negativa.
Sem fatores de risco.
Exame físco normal.
Ecocardiograma normal.
ECG (anexo).
Holter normal.
RNM cardíaca normal.
ECGAR positivo.
Durante o TE apresentou surtos bem tolerados de taquicardia ventricular sem compromentimento hemodinâmico. Apenas palpitações.
Qual o diagnóstico mais provável e qual próximo passo a ser seguido?
Raimundo

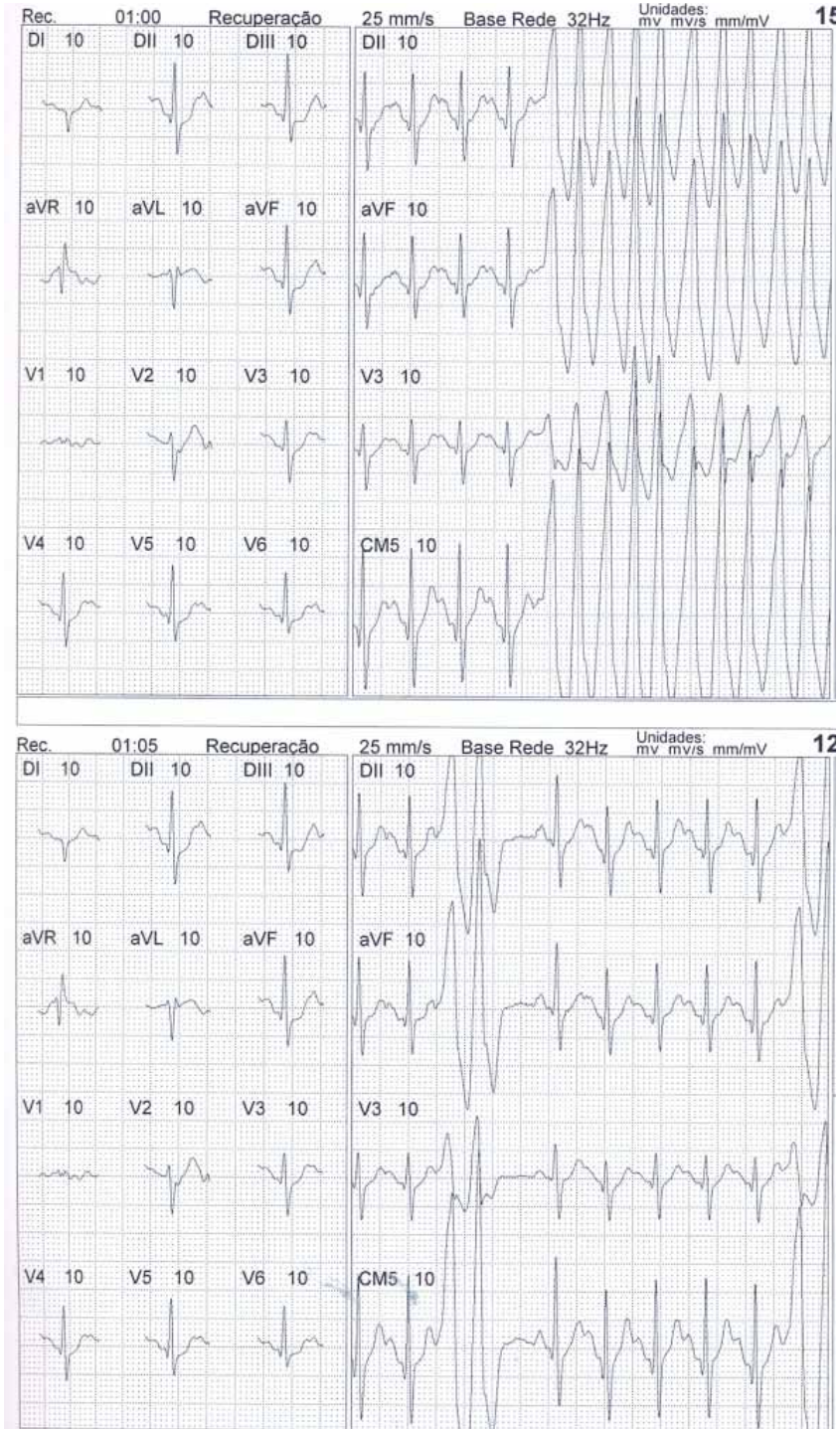
Male patient, 37 years old, with story of palpitation induzed only by exertion.
Negates syncope. Family History negative. No risk factors. Normal physical examination
Normal echocardiogram.
ECG (attached).
Holter: normal.
Cardiac MRI: normal.
SAECG: Late potentials (positive.)
During Exercise testing presented well tolerated ventricular tachycardia without hemodynamic implications.
Which is the more provable diagnosis? Which is the next step approach?
All the best for all Raimundo.


ECG 1
DI 10
0,11 0,38
DII 10
1,26 0,56
DIII 10
1,24 0,19
aVR 10
0,25 -0,56
aVL 10
0,05 -1,56
aVF 10
1,25 0,38


ECG 2
V1 10
V2 10
V3 10
V4 10
V5 10
V6 10


DI 10
DII 10
DIII 10
DII 10
ECG 3
aVR 10
aVL 10
aVF 10
aVF 10
V1 10
V2 10
V3 10
V3 10
V4 10
V5 10
V6 10
CM5 10

ECG 4


Rec. 01:00 Recuperação 25 mm/s Base Rede 32Hz Unidades: mV mV/s mm/mV
DI 10 DII 10 DIII 10
aVR 10 aVL 10 aVF 10
V1 10 V2 10 V3 10
V4 10 V5 10 V6 10
DII 10
aVF 10
V3 10
CM5 10
Rec. 01:05 Recuperação 25 mm/s Base Rede 32Hz Unidades: mV mV/s mm/mV
DI 10 DII 10 DIII 10
aVR 10 aVL 10 aVF 10
V1 10 V2 10 V3 10
V4 10 V5 10 V6 10
DII 10
aVF 10
V3 10
CM5 10

Colleagues opinions

First feeling:
Idiopathic catecholamine sensitive PLEOMORPHIC (not polymorphic) VT originating from the outflow tract area (I do not exclude a primary RVOT location with some complexes originating from the ""LVOT"" area if the position of precordial electrodes is correct).
I would rather try a beta-blocker agent first and if it fails or is not well tolerated discuss the option of RF ablation.
Prof. Bernard Belhassen,
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Primera impresion:
TV Idiopática catecolaminérgica pleomórfica (no polimórfica!!) originada de el tracto de salida. Yo no excluyo una localización en el tracto de salida con algunos complejos originado en el tracto de salida del VI si la posición de los electrodos precordiales está correcta.

Comenzaria con beta-bloqueadores primero y caso fallen o no fueran bien tolerados discutiria la opción de radiofrecuencia.
Bernard Belhansen

Dear Andres,
Based on the info Dr. Raimundo Barbosa Barros provided, this is likely an idiopathic left ventricular tachycardia. Varapemil-sensitive is the most common type.

Thanks much,

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Wynnewood, PA 19096
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Querido Andrés:

Basado en la información del Dr Raimundo Barbosa Barros esta parece ser una TV idiopática del VI verapamilo sensible, la cual es el tipo mas comun
Muchas gracias
Li

A minha impressão é que se trata de uma taquicardia regular com FC (ventricular) de 300bat/min por Flutter atrial 1:1.

O ECG de base: PR 200ms e SÂQRS +110º

Conclusâo: com eixo do QRS desviado para a direita. O inicio da taquicardia e protagonizado por 2 batimentos precedidos de onda delta, e os QRS são bizarros,alargados. FUTTER ATRIAL 1:1, ANOMALIA CONGENITA :WPW.

Grato.

lourivalcampos@yahoo.com.br

My impression is that this is a regular tachycardia with HR=300bpm (ventricular) of atrial flutter 1:1.

The baseline ECG: PR interval= 200ms; and QRS axis in +110 °.

With QRS axis shifted to the right. The onset of tachycardia has two beats preceded by a delta wave, and the QRS complexes are bizarre and wided.

Conclusion: Atrial Futter 1:1, congenital anomaly, WPW.

Lourival Campos MD São Paulo Brazil.

Andres, Hello! I see that in baseline ECG there is only IRBBB; probably this is a patient with a thin physical build and taller than 1.70 m?

Besides, I would like to know the following data from the Echo: dimensionsof the LV in diastole; interventricular septal thickness in diastole;
posterior wall in diastole, and his height and weight.

Besides, if he has a Holter, provide these data.

Carlos A Soria MD

Hola; veo que en el ECG Basal sólo posee BIRD; probablemente sea un paciente de contextura física delgado y de más de 1,70 mts?
Además me gustaría saber los siguientes datos del Ecocardiog.: medidas del VID; SIVD;PPD; y además la estatura y el peso;
Además si tiene un Holter, pasar los datos.

Carlos A Soria

Dear Soria,In the presentation of the case by Raimundo, he wrote that the echo is normal, as is the NMR. Without specifying the values you requested, what would be the significance of this?

Is it not enough to say that the ECG + NMR were normal without structural heart disease?

This triphasic pattern of V1 makes us think about our understanding in end conduction delay by the inferior division of the right branch, mainly by the predominantly negative complexes in the high left leads I and aVL. Probably this is an asthenic subject with graceful thoracic structure and a considerable height (Don Quixote's biotype). This would be a type II in our classification:

ECD located in the right inferior quadrant in the territory of the right branch inferior division. Characterized by presenting ECD located in the right inferior quadrant of the inferior division of the right branch. It corresponds to the inferior, posterior, or postero-inferior division territory of the right branch. The differential diagnosis is presented as divisional postero-inferior block of the left branch (LPFB). Many of the cases in literature described as LPFB, the way we see it, are type II ECD, with electro-vectocardiographic differences being very subtle at times, so the diagnosis should always be clinico-electro-vectorcardiographic.

A) Electrocardiographic criteria:

SÂQRS between +70º and +110º;

Normal QRS duration;

S1 R2 R3 pattern, with RII and RIII being of not-increased voltage (usually ≤ than 10 mm), never reaching 15 mm (an essential element for the differential diagnosis with LPFB)

R2 ≤ R3 (in LPFB R3 > R2)

aVR of the QS type

Possible notch in the descending slope of the inferior leads; S wave from V2 and/or V3 with increased depth; persistent S wave up to V5 and/or V6

V1: rS, RS or rSR' with S in V1 and V2, possible broader.

B) Vectorcardiographic criteria:

End conduction delay in the three planes located at the right and below.

Frontal plane:

1. Initial vectors always at the left, above or below; clockwise rotation
2. Predominant location in the left quadrants Rapid change from left to right between 30 to 50 ms
3. ECD at the right and below between +120º and +150º

Horizontal plane

1. QRS loop of counterclockwise rotation
2. Marked posterior shift
3. Rapid change from left to right between 40 to 50 ms ECD at the right and below.

Right sagittal plane

1. Initial vectors above or below
2. Clockwise rotation
3. Marked postero-inferior shift
4. ECD below and back

Andrés Ricardo Pérez-Riera.

Querido Soria: en la presentación del caso Raimundo colocó que el eco es normal asi como la RNM. Sin especificar los valores por ti solicitados. Cual seria la importancia de esto? No vasta decir que el ECO + RNM fueron normales sin cardiopatia estructural?. Este padrão trifásico de V1 hace pensar en nosso entendimento em atrasso final de conducción por la división inferior de la rama derecha principlamente por los complejos predominantemente negativos en las derivaciones izquierdas altas I y aVL. Provavelmente este es un paciente longílinio asténico de estrutura torácia gracil y altura considerável.(biotipo don quijote da Mancha) Este seria o Tipo II da nossaclassificação:AFC localizado no quadrante inferior direito no território da divisão inferior do ramo direito. Caracterizado por apresentar o AFC localizado no quadrante inferior direito no território da divisão inferior do ramo direito. Corresponde ao território da divisão inferior, posterior ou póstero-inferior do ramo direito (BDIRD).O diagnóstico diferencial se apresenta com o bloqueio divisional póstero-inferior do ramo esquerdo (BDPI). Muitos dos casos de literatura descritos como BDPI, ao nosso entender são AFC Tipo II sendo suas diferenças eletro-vetorcardiográficas por vezes muito sutis pelo que o diagnóstico sempre deve ser clínico-eletro-vetorcardiográfico.

A) CRITÉRIOS ELETROCARDIOGRÁFICOS

1. SÂQRS entre + 70º e + 110º;
2. Duração do QRS normal;
3. Padrão S1 R2 R3, sendo RII e RIII de voltagem não aumentada (habitualmente ≤ que 10mm), nunca atingindo l5mm (elemento fundamental para o diagnóstico diferencial com BDPIE);
4. R2 ≤ R3 (no BDPI RIII > RII);
5. Avr complexo QRS do tipo QS;
6. Eventual entalhe na rampa descendente das derivações inferiores;
7. Onda S de V2 e/ou V3 de profundidade aumentada;
8. Onda S persistentes até V5 e/ou V6;V1: rs,
9. RS ou rsr' com S de V1 e V2 eventualmente empastada.

B) CRITÉRIOS VETORCARDIOGRÁFICOS

Atraso final nos três planos localizado a direita e abaixo.

PLANO FRONTAL

1. Vetores iniciais sempre para esquerda, acima ou abaixo;
2. Rotação horária; Localização predominante nos quadrantes inferiores;
3. Rápida mudança de esquerda para direita entre os momentos 30ms a 50ms;
4. AFC a direita e abaixo entre +120º e +150º.

PLANO HORIZONTAL

1. Alça QRS de rotação anti-horária; Marcado deslocamento posterior;
2. Rápida mudança de esquerda para a direita entre os momentos 40 a 50 ms;
3. AFC a direita e atrás.

PLANO SAGITAL DIREITO

1. Vetores iniciais para cima ou abaixo;
2. Rotação horária;
3. Marcado deslocamento póstero-inferior; AFC abaixo e atrás.

Andres,

Well,

Thank you for sending me the e-mail.

In fact, one suspects that the patient of this tracing is longilineal and asthenic. And that also, both the echo and the NMR are normal.

I was hoping you'd tell me that the LV mass and the LV mass index are normal, just that.

I asked for the diastolic data of the Echo, and the weight and height to apply the Deveraux formula, and thus measure the LV mass and the LV mass index applying the staturo-ponderal ratio.

Because having this data of an increased LV mass index for the staturo-ponderal ratio of the patient, we should think of some type of LVH, and with LVH, such VT, I think would not be so benign. All the more so if the patient is a sportsman.

The tracing presented could be a catecholaminergic VT; moreover, it would be useful and interesting to perform a Holter recording.

Regards for all the members of the forum,

Carlos Soria

Bien;

Gracias por enviarme el mail;

En realidad, uno sospecha que el paciente de ese trazado es longilíneo y asténico.
Y que además tanto el Ecocard. como la RNM son normales;

Yo apuntaba a que me dijeran que la MVI y el IMVI son normales, nada más.

Pedía los datos diastólicos del Ecocard. y el peso y la estatura para que aplicando la fórmula de Devereaux medir la MVI y el IMVI aplicando la relación pondoestatural.

Por que de tener ese dato del IMVI incrementado para la relación pondo estatural del paciente, tendría q pensar en algún tipo de HVI; y con HVI, dicha TV, pienso no sería tan benigna.
Más aún si el paciente es deportista.

El trazado presentado puede dar para una TV catecolaminérgica; además que sería muy útil e interesante realizarle un Holter;

Saludos a los del foro

Carlos A. Soria.

Dear friends from the forum, my opinion:

1- Baseline ECG: sinus rhythm at 75 per minute, with electrical axis in the right inferior quadrant (maybe corresponding to a longilineal person) with
S1 S2 S3 by straight back and end conduction delay (rsr") Normal 380 ms QT for the age and sex: I see no qt alterations compatible with long QT syndrome
2- With strain and rates of 140 per minute he presents tachycardia with wide qrs with changing axis compatble to polymorphic VT.

This ECG analysis in the context of:
Non relevant clinical aspects (no syncopes, no symptoms or signs of heart failure), normal echo (nor RV alterations) and normal NMR that are not oriented to RV dysplasia (although he does have ventricular late potentials), VT induced by strain and not of night appearance as it happens in Brugada syndrome, I would think of:
Catecholaminergic polymorphic ventricular tachycardia of Coummel, induced by strain.

What management would I apply? Because he is asymptomatic, I would medicate with BB (propranolol 2-3 mg/kg or nadolol) and new stress test. I have to say that it would have been interesting in this patient, to detect the appearance of polymorphic VT of ventricular bigeminy and with a higher frequency of bidirectional VT, a characteristic of Catecholaminergic polymorphic ventricular tachycardia of Coummel.
Finally, although this arrhythmia is more frequent in children in school age, unfrequently it may appear in the adult age or older than 30 years, and it is due to a genetic alteration that should be investigated.

Cordially,
Juan J Sirena MD Santiago del Estero Argentine Republic

Estimados amigos foristas, mi opinion

1- ECG basal: ritmo sinusal a 75 l minuto ,con eje electrico en cuadrante inferior derecho ,(probablemente corresponda a un longilineo (con
S1S2S3 por espalda recta) y retardo final de conduccion (rsr")
Intervalo QT de 380 ms. Osea normal para edad y sexo: No veo duración del QT compatible con sindrome QT largo

2- con el esfuerzo y con frecuencias de 140 minuto presenta Taquicardia con QRS ancho con eje cambiante compatible con TV polimórfica.
Este analisis de ECG en contexto de :
clinica no relevante (no sincopes ,no sintomas signos de insuf cardiaca) eco cardiograma normal (no alteraciones de VD) y RNM normal que no orientan a Displasia arritmógena del VD (,aunque tiene PTV) TV inducida por esfuerzo y no de aparicion nocturna como lo es en el sindrome de Brugada pensaria en:
TAQUICARDIA VENTRICULAR CATECOLAMINERGICA POLIMORFICA DE COUMMEL ,INDUCIDA POR EL ESFUERZO
Que conducta tomaria ? Por hemodinamicamente estable medicaria con beta bloqueadores (propanolol2-3 mg / kg o nadolol) y a seguir haria nueva prueba de esfuerzo, Debo decir que hubiese sido interesante en este paciente detectar antes de la aparición de la TV polimórficas, aparicion de bigeminia ventricular y con mayores frecuencias TV Bidirecional ,una caracteristica de la TVCP de Coummel:
Por ultimo aunque esta arritmia es mas frecuente en niños de edad escolar, puede infrecuentemente aparecer en edad adulta o mayores de 30 años y se debe a una alteración genetica que se deberia pesquizar
Saludos cordiales
Juan J Sirena

Dearest friend Dr Sirena: Catecholaminergic polymorphic ventricular tachycardia (CPVT) is a rare inherited disorder associated with syncope and sudden death that manifests predominantly in children and early adolescence or teenagers during effort o emotions. The incidence of syncope is very high.

In Leenhart series the mean age at first event was 7.8 + - 4 years. These patients often have very strong family history of SD.

The present case report is an adult man, without family history and without syncope or SD. I agree that begin in adult is possible, In reference to genetic variant we have:

CPVT1: Chromosomal locus 1q42-43,Gene RYR2, autosomal dominant phenotype CPVT, IVF

CPVT2: Chromosomal locus 1q23-21,Gene CASQ2, autosomal recessive phenotype CPVT,

CPVT3: Chromosomal locus 7q14-22,Gene unknow, autosomal recessive phenotype CPVT, Long QT interval.

Related phenotypes

1. **LQT4 chromosome 4q25-26, Gene: ANK2, Autosomal dominant, Phenotype: IVF, QT prolongation, stress induced bidirectional VT**
2. **ATS: 17q23.1-q24.2, Gene KCNJ2, Autosomal dominant, Phenotype U wave, bidirectional VT, periodic paralysis, facial diysmorphism.**
3. **CPVT/DMC chromosomal locus 1 q42-43, Gene RYR2, autosomal dominant, Phenotype stress induced VT, sunus node dysfunction, dilated cardiomyopathy.**

Andrés Pérez-Riera

Mi querido amigo Sirenita:

La taquicardia ventricular polimórfica catecolaminérgica (CPVT) es un trastorno hereditario raro, asociado a síncope y muerte súbita que se manifiesta predominantemente en niños y la adolescencia precoz o adolescentes durante esfuerzo o emociones. La incidencia de síncope es muy alta.

En la serie de Leenhart, la edad promedio al primer evento era 7,8 +/- 4 años. Estos pacientes con frecuencia tienen una historia familiar muy fuerte de MS.

El presente informe de caso es de un hombre adulto, sin historia familiar y sin síncope o MS. Estoy de acuerdo con que un comienzo en un adulto es posible. Con respecto a la variante genética tenemos:

CPVT1: locus cromosómico 1q42-43, gen RYR2, fenotipo dominante autosómico CPVT, fibrilación ventricular idiopática.

CPVT2: locus cromosómico 1q23-21, gen CASQ2, fenotipo recesivo autosómico CPVT

CPVT3: locus cromosómico 7q14-22, gen desconocido, fenotipo recesivo autosómico CPVT, intervalo QT prolongado.

Fenotipos relacionados

1. **LQT4, cromosoma 4q25-26, gen: ANK2, dominante autosómico, fenotipo: fibrilación ventricular idiopática, prolongación QT, TV bidireccional inducida por estrés.**
2. **Síndrome de Andersen-Tawil: 17q23.1-q24.2, gen KCNJ2, dominante autosómico, fenotipo onda U, TV bidireccional, parálisis periódica, dismorfismo facial.**
3. **CPVT/miocardiopatía dilatada, locus cromosómico 1q42-43, gen RYR2, dominante autosómico, fenotipo: TV inducido por estrés, disfunción del nodo sinusal, miocardiopatía dilatada.**

Andrés,

Hello, everyone. Dear Dr. Sirena, it doesn't seem polymorphic, where do you see this? On the other hand, he is not asymptomatic. In the PDF he presents palpitations with strain and never syncope. The way I see it, this is adrenergic-dependent monomorphic VT, and probably sensitive to verapamil or adenosine, without evidence of structural heart disease, i.e. of the so-called idiopathic VT. It would be good to see the VT in all the 12-lead ECG to try to evaluate the place of origin (the transition of QRS complexes in the right precordial leads, V1 through V3 guide us) that in the order of frequency would be in the RVOT, mitro-aortic,septo-interventricular continuity, generally left of the aortic root. This is at the level of coronary and not coronary sinuses. In any case, they are always ablated, with a very high percentage of success, above 90% and with the usual risk of any ablation procedure (not AF) lower than 1%. My proposal is undoubtedly catheter ablation, the only proven healing treatment to this date for most, almost all, arrhythmias.
Regards,Francisco Femenia MD Mendoza- Argentine

Hola a todos, querido Sirena no impresiona polimorfica, donde la ves? Por otro lado no es asintomatico, por lo que refiere el interrogatório presenta palpitaciones ante esfuerzos y no sincope. A mi entender es una TV monomorfica adrenergico dependiente, y probablemente verapamilo o adenosina sensible, sin evidencias de cardiopatia estructural,o sea de las denominadas TV idiopaticas, seria bueno ver la TV en todo elECG de 12 derivaciones para intentar evaluar el sitio de origen (nos orientala transicion de los complejos QRS en precordiales derechas, V1 a V3) que enorden de frecuencia seria en Tracto de Salida de VD, Continuidad mitroaortica, septo interventricular, generalmente izquierdas y de raiz de aorta, esto es a nivel de los senos coronarianos y no coronarianos.
En cualquier caso son ablacionables siempre, con un altisimo porcentaje de exito, superior al 90% y con el riesgo habitual de cualquier procedimiento ablativo (no FA) menor al 1%. Mi propuesta es sin dudas ablacion por cateteres, unico tratamiento curativodemostrado hasta la fecha para la mayoria, casi todas, las arritmias.
Saludos pancho femenia

FINAL COMMENTARIES

CLASSIFICATION OF VT BY UNDERLYING SUBSTRATE

1) With Structural Heart Disease
2) Without Structural Heart Disease
   2a) Monomorphic
   2b) Polymorphic
   With long QT or TdP
   With normal or true PVT.

CLASSIFICATION OF CLINICAL MANIFESTATIONS OF VT

CLASS I	Asymptomatic or palpitations The present case
CLASS II	Dizziness, precordial pain and/or dyspnea.
CLASS III	Presyncope or syncope.
CLASS IV	Cardiopulmonary arrest.

Classification of VT by the presence or not of underlying structural heart disease and the four classes of symptoms secondary to VT.

VENTRICULAR TACHYCARDIA IN PATIENTS WITH STRUCTURAL NORMAL HEARTS

CLASSIFICATION

1) Adenosine-Sensitive Ventricular Tachycardia MVT sensitive to adenosine by triggered activity.
   - From RVOT: ≈ 80% of cases
   - From LVOT

   Phenotypes
   - Repetitive Monomorphic Premature Ventricular Complexes (PVCs)
   - Non-sustained repetitive monomorphic ventricular Tachycardia (RMVT); or
   - paroxismal, exercise induced sustained ventricular Tachicardia

2) Verapamil-Sensitive Fascicular Tachycardia MVT sensitive to verapamil by intrafascicular reentry.

3) MVT sensitive to propranolol and automatic.

4) Undifferentiated MVT.

Classification of MVT with no apparent structural heart disease.

1) Adenosine-Sensitive Ventricular Tachycardia MVT sensitive to adenosine by triggered activity

1. **SENSITIVE TO ADENOSINE (TRIGGERED ACTIVITY): MORPHOLOGY:** LBBB with inferior axis, RBBB; inferior axis; RBBB, superior axis.

2. **INDUCTION:** programmed stimulation +- catecholamines.

3. **MECHANISM:** triggered activity mediated by cyclic adenosine monophosphate (cAMP).

4. **SENSITIVE:** adenosine, verapamil and propranolol.

5. **“ENTRAINMENT”:** negative.

Characteristics of MVT sensitive to adenosine by triggered activity.

2) Verapamil-Sensitive Fascicular Tachycardia MVT sensitive to verapamil by intrafascicular reentry

MORPHOLOGY: RBBB, inferior axis; RBBB, superior axis: it originates in the left posterior fascicle, or in the anterior fascicle, or RBBB and LBBB alternating.

MECHANISM: reentry.

“ENTRAINMENT”: positive;

SENSITIVE: verapamil and + - propranolol.

Non-specific transient inferolateral T-wave changes may be present after cessation of event .
The ECG morphology changes after ablation were divided into two categories:
With new or deepening Q wave in inferior leads and/or disappearance of Q wave in leads I and VL
Without change.

Characteristics of MVT sensitive to verapamil by intrafascicular reentry.

3) MVT SENSITIVE TO PROPRANOLOL AND AUTOMATIC

INDUCTION: catecholamines. Induced by incessant exercise.

MORPHOLOGY: RBBB or LBBB or left ventricle or polymorphic.

MECHANISM: increased automaticity.

“ENTRAINMENT”: negative.

SENSITIVE: transient/permanent suppression with propranolol. Transient suppression with adenosine or with no effect.

Characteristics of MVT sensitive to propranolol and automatic.

4) UNDIFFERENTIATED MVT

INDUCTION: by exercise.

MORPHOLOGY: LBBB with left axis. VT of RV outflow tract.

MECHANISM: reentry.

“ENTRAINMENT”: positive.

SENSITIVE: adenosine, propranolol or verapamil: negative.

Characteristics of undifferentiated MVT.

Idiopathic Monomorphic Right Ventricular Tachycardia arising from RVOT (IMVT-RVOT)

I) FAMILY HISTORY OF ARRHYTHMIA OR SCD

Has not family history antecedents.

II) AGE OF CLINICAL MANIFESTATION

Most patients are initially diagnosed between the ages of 30 and 50 years. In Lermans series there ranged between 6 and 77 years.(1)

(III) SEX PREDOMINANCE

Adenosine-sensitive RVOT segregates equally between both sexes.

IV) PATTERN OF INHERITANCE

Non-familial arrhythmic condition.

V) ANNUAL RATE OF SUDDEN CARDIAC DEATH (SCD)

Rare. Excellent prognosis.

VI) SYMPTOMS

In 80% palpitations; 50% dizziness; 10% syncope during VT, In 80% class I and II symptoms; In 20% class III: Pre-syncope or Syncope.
Triggered by stress or exercises, gestation, extreme consumption of alcohol, coffee or tobacco.

VII) IRBBB OR CRBBB PATTERN

Present in 10% of cases

1. **Lerman BB, Kenneth M, Stein SM, et al. Ventricular Tachycardia in Patients With Structural Normal Heart. In Zipes DP & Jalife J Cardiac Electrophysiology From Cell to Bedside 3rd. 2000; Chapter 70 pp: 640-656.**
2. **Lerman BB, Kenneth M, Stein SM, et al. Ventricular Tachycardia in Patients With Structural Normal Heart. In Zipes DP & Jalife J Cardiac Electrophysiology From Cell to Bedside 3rd. 2000; Chapter 70 pp: 640-656.**
3. **Buxton AE, Waxman HL, Marchlinski FE, et al. Right ventricular tachycardia: clinical and electrophysiologic characteristics. Circulation. 1983; 68:917-927.**

IIX) T-WAVE POLARITY

Always T wave upright from V2- V5.(Differential diagnosis with ARVC/D).]

(IX) PARIETAL BLOCK

Absent. QRSD <110 ms in V1, V2 or V3. (Differential diagnosis with ARVC/D).

(X) EPSILON WAVE

Absent. Do not exist.

(XI) NEGATIVE T-WAVE FROM V1 TO V2 OR V3

Absent. (Differential diagnosis with ARVC/D).

(XII) FRONTAL PLANE QRS AXIS OF VT

Positive in inferior leads III and aVF, negative in lead aVL. LBBB morphology inferior axis.

In 10% it may originate in the LV, in the region of the posteroinferior division of left bundle with RBBB morphology and extreme deviation of SÂQRS to the left.

Another morphology that suggests the focus of origin in the LV, is LBBB associated to early transition in the V2 lead. Dominant R in V1 and inferior SÂQRS points the focus of superior origin in the LV.

Rarely, it could have an epicardial origin, characterized by positive concordance in precordial leads and negative complexes in I and aVL.

TV has been described in literature, as presenting both LBBB and RBBB morphologies, each one with identical SÂQRS, which suggests that the focus originates in the interventricular septum with a dual exit to the left and to the right. **(1)**

According to the QRS configuration during episode of IM-VT four groups were distinguished by Mont et al **(2):**

1.Dixit S, Gerstenfeld EP, Callans DJ, et al. Electrocardiographic patterns of superior right ventricular outflow tract tachycardias: distinguishing septal and free-wall sites of origin. J Cardiovasc Electrophysiol. 2003; 14:1-7.

2.Mont L, Seixas T, Brugada P, et al. The electrocardiographic, clinical, and electrophysiologic spectrum of idiopathic monomorphic ventricular. Am Heart J. 1992; 124:746-753.

1. **Group I:** RBBB morphology and SÂQRS superior on frontal plane: It group had dizziness during VT less frequently, but they needed cardioversion to terminate their arrhythmias more often. They experienced tachycardia during exercise less often, and tachycardia was not initiated during exercise testing. They had fewer PVCs according to the Holter recording. During the electrophysiologic study, VT was induced and terminated by pacing more often in this group. Reentry seems to be the most likely arrhythmia mechanism in this group.
2. **Group II:** RBBB morphology and intermediate SÂQRS on frontal plane;
3. **Group III:** LBBB morphology and left axis deviation;
4. **Group IV:** LBBB morphology wit right axis deviation or intermediate.

(XIII) ARRHYTHMIAS

PVCs, NS-VT or sustained VT at rest or with exercise. The most common form of idiopathic VT is repetitive M-VT (RMVT), which typically occurs at rest and is characterized by frequent PVCs and salvos of NS-VT with intervening sinus rhythm. Although the arrhythmia occurs at rest, the constellation of findings in idiopathic VT that is characterized by RMVT is consistent with the mechanism of cAMP-mediated triggered activity. Therefore, the spectrum of VT resulting from this mechanism includes not only paroxysmal exercise-induced VT but also RMVT (1) Several distinctive ECG characteristics and detailed ECG analysis can differentiate free wall RVOT from septum-VT/ PVCs:

R wave amplitudes in the inferior leads were significantly smaller in free wall;

RVOT or PVCs than in septum VT/PVC;

An RR' pattern in the inferior leads was observed significantly more often in free wall RVOT/PVC than in septum -VT/PVC;

QS-wave amplitude in each of leads V1 to V3 was significantly deeper in free wall -RVOT/PVC than in septum -VT/PVC (2)

1. **Lerman BB, et al. Mechanism of repetitive monomorphic ventricular tachycardia. Circulation. 1995; 92:421-429.**
2. **Tada H, Ito S, Naito S, et al. Prevalence and electrocardiographic characteristics of idiopathic ventricular arrhythmia originating in the free wall of the right ventricular outflow tract. Circ J. 2004; 68:909-914.**

(XIV) VENTRICULAR ARRHYTHMIAS MECHANISM

1. **Trigged activity** (Adenosine-Sensitive): 70% of cases. cAMP-mediated triggered activity.
2. **Delayed triggered activity**, dependent on post-depolarization in phase 4, associated to increase of cyclic AMP and mediated by catecholamines: adrenergic-dependent.
3. **Intrafascicular Reentry** (Verapamil sensitive); 10% of cases.
4. **Enhanced Automaticy** (Propranolol-sensitive);
5. **Reentry**
6. **Undifferentiated(1)**

   Dynamic changes in the T-U wave were observed in patients with idiopathic M-VT originating from the RVOT. Further investigations are required to elucidate the precise role of the U wave in arrhythmogenesis in these patients. **(2)**

(XV) SIGNAL AVERAGED ECG

Always normal. The present case is not normal?? LP

(XVI) MICROVOLT T-WAVE ALTERNANS (TWA)

It is a risk marker for ventricular arrhythmias. Negative for T-wave alternans in > 90% of cases.

(XVII) EXERCISE STRESS TESTING

In 50% of cases adrenergic-dependent effort induced VT and Provo cable by exercise or effort induced VT. Tree variants are Exercise induced: **The variant Adenosine-Sensitive (Triggered Activity); The variant Propanolol-Sensitive and the Undifferentiated.!!!!!!**

The variant Adenosine-Sensitive (Triggered Activity) is exercise-induced with repetitive M-VT.

The variant Propanolol-Sensitive is Exercise induced with incessant.

1. **Lerman BB, Stein KM, Markowitz SM.: Idiopathic right ventricular outflow tract tachycardia: A clinical approach. Pacing Clin Electrophysiol. 1996 19:2120-2137.).**
2. **Nakagawa M, Ooie T, Hara M, et al. Dynamics of T-U wave in patients with idiopathic ventricular tachycardia originating from the right ventricular outflow tract. Pacing Clin Electrophysiol. 2004; 27:148-155**.

XIIX) HOLTER MONITORING AND LOOPER

Detection of frequent or in bursts monomorphic PVCs that occur predominantly during the day.Record M-VT with LBBB pattern with inferior SÂQRS: between + 30º and + 120º indicative of RVOT origin.

(XIX) ECHOCARDIOGRAM

Normal in 90% of cases. Rarely, slight enlargement of RV.

(XX) RV VENTRICULOGRAM

Usually Normal.

(XXI)CARDIAC MAGNETIC RESONANCE IMAGING

Usually normal, but data in literature is conflicting. Carlson et al **(1)** refer: focal wall thinning focally diminished systolic wall thickening, and abnormal systolic wall motion. Focal fatty infiltration was referred.RV abnormalities were revealed in 32 (60%) of the 53 patients: fixed thinning in 27 (84%), fatty replacement in eight (25%), and reduced wall thickening or motion in 31 (97%). RV abnormalities were found in 35 (76%) of 46 patients with idiopathic RVOT VT. Mild RV abnormalities are likely sources for arrhythmias, even in the absence of ARVC/D(2;3)
.Menghetti et al **(4)** studied fifteen patients that had a clinical diagnosis of ARVC/D with spin-echo T1-weighted NMR and multislice scan. Ten of the 15 patients with ARVC had an abnormal NMR result (67% sensitivity), with areas that had signal intensity close to that of pericardial or subcutaneous fat. In the remaining five cases the NMR signal was inadequate.

1. **Farwell DJ, Freemantle N, Sulke AN. Use of implantable loop recorders in the diagnosis and management of syncope. Eur Heart J. 2004; 25:1257-1263.**
2. **Carlson MD, White RD, Trohman RG, et al. Right ventricular outflow tract ventricular tachycardia: detection of previously unrecognized anatomic abnormalities using cine magnetic resonance imaging.J Am Coll Cardiol. 1994; 24:720-727.**
3. **White RD, Trohman RG, Flamm SD, et al. Right ventricular arrhythmia in the absence of arrhythmogenic dysplasia: MR imaging of myocardial abnormalities. Radiology. 1998;207:743-751.**
4. **Menghetti L, Basso C, Nava A, et al. Spin-echo nuclear magnetic resonance for tissue characterisation in arrhythmogenic right ventricular cardiomyopathy.Heart. 1996;76:467-470.**

(XXII) RESPONSE TO PROGRAMMED ELECTRICAL STIMULATION (PES)

Inducibility of VT by PES with ventricular extra stimuli: 3%;
Presence of more than one ECG morphology during tachycardia: 0%;
Fragmented diastolic potentials during ventricular arrhythmia: 0%.

(XXIII) ENTRAINMENT

Technique that consists in applying extra-stimuli in series during S-VT, using cycles for at least 20 ms less than the VT cycle, accelerating the tachycardia to stimulated rate.
IMVT-RVOT: Negative. Insensitive / not present.

(XXIV) RESPONSE TO CATECHOLAMINES AGENTS ISOPROTERENOL, ISOPRENALINE OR DOBUTAMINE

IMVT-RVOT: Facilitates in cAMP-Triggered Activity
(Adenosine-Sensitive) and Propanolol-Sensitive (Automatic).
Facilitates/no effect in reentry in Verapamil-Sensitive, in undifferentiated, atriofascicular and Bundle Brach reentry variants.

(XXV) RESPONSE TO CLASS II ANTIARRHYTHMIC AGENTS: BETA-ADRENOCEPTOR BLOCKERS: PROPRANOLOL AND OTHERS

Adenosine-Sensitive: Present/ sensitive;
Verapamil-Sensitive: Present/ sensitive or not present/insensitive;
Propanolol-Sensitive: Terminates/transient suppression;
Undifferentiated: not present/insensitive;

(XXVI) ENDOMYOCARDIAL BIOPSY (EMB)

Usually negative: without structural heart disease.
Recent studies with EMB contradict this concept, having shown abnormalities in more than 65% of cases, which increases to more than 80% when the material is product of an autopsy. Thus, the following were described **(1)**Indicatives of structural heart disease:

1.Hamartoma of Purkinje fibers was described **(2)**
2.Mild form of ARVC/D;
3.Microangiopathy associated to subendocardial fibrosis;
4.Sub-clinical myocarditis;
5.Focal cardiomyopathy;
6.Atherosclerotic ischemic cardiomyopathy;
7.Non-atherosclerotic ischemic cardiomyopathy;
8.Hypertrophic cardiomyopathy;
9.Mitral valve prolapse.

(XXVII) RESPONSE TO PHARMACOLOGICAL THERAPY

Therapy only mandatory if presyncope or syncope present.
Beta-blockers are effective in 35% of cases;
Calcium channels blockers: are effective in 30% of cases;
Association Class IA and IC is effective in 35% of cases;
Class III drugs are effective in 50% of cases.
Acute termination: Vagal maneuver; adenosine, intravenous verapamil and lidocaine.

1. **Markowitz SM, Litvak BL, Ramirez de Arellano EA, et al. Adenosine-sensitive ventricular tachycardia: right ventricular abnormalities delineated by magnetic resonance imaging. Circulation. 1997; 96:1192-2000.**
2. **Garson A Jr, Smith RT Jr, Moak JP, et al. Incessant ventricular tachycardia in infants: myocardial hamartomas and surgical cure. J Am Coll Cardiol. 1987; 10:619-626.**

(XXVIII) RADIOFREQUENCY CATHETER ABLATION (RFCA)

Usually is an effective, curative and safe therapy. RFCA is the treatment of choice in drug refractory VT in structurally normal heart. A new RBBB develops in 2% of cases.

Procedural success: noninducibility of VT after RFCA **(1;2)**.

Ito et al **(3)** describe a ECG algorithm having a high sensitivity(88%) and specificity(96%) to identify the optimal ablation site for idiopathic ventricular outflow tachycardia or PVCs. The catheter sites were verified by multi-plane fluoroscopy. The author divided the RVOT in six areas:

1. RV septum;
2. RV free wall;
3. RV near the His-bundle region;
4. LV endocardium;
5. Left sinus of Valsalva (LSV);
6. LV epicardium remote from the LSV.

A PVCs originating from the LV epicardium remote from the LSV was defined as a PVCs in which the earliest ventricular activation was recorded at the LSV and RFCA from the LSV failed.

Noncontact mapping is a safe and effective alternative method to guide ablation of hemodynamically unstable or nonsustained ventricular arrhythmia originating from RVOT **(4)**

1.van der Burg AE, de Groot NM, van Erven L, et al. Long-term follow-up after radiofrequency catheter ablation of ventricular tachycardia: a successful approach? J Cardiovasc Electrophysiol 2002; 13:417-23.

2.Maciag A, Sterlinski M, Pytkowski M, et al. Successful radiofrequency catheter ablation of the symptomatic ventricular tachycardia in structurally normal heart. Case report. Pol Arch Med Wewn. 2003; 110:1453-1457.

3.Fung JW, Chan HC, Chan JY, et al. Ablation of nonsustained or hemodynamically unstable ventricular arrhythmia originating from the right ventricular outflow tract guided by noncontact mapping. Pacing Clin Electrophysiol. 2003;26:1699-1705.

4.Ito S, Tada H, Naito S, et al. Development and validation of an ECG algorithm for identifying the optimal ablation site for idiopathic ventricular outflow tract tachycardia. J Cardiovasc Electrophysiol. 2003;14:1280-1286.

(XIX) NATURAL HISTORY, PROGNOSIS, LONG-TERM FOLLOW-UP

Usually good. SCD is rare in this patient population. Frequent episodes may result in cardiomyopaty and render the decision of RFCA of the focus more imperative.
In children the prognosis is favorable, however, appropriate treatment and follow-up were required in children with S-VT, symptomatic VT or VT with a high rate. **(1)**
Although the prognosis of these patients remains excellent, they should continue to have periodic cardiac follow-up to rule out latent progressive heart disease such as ARVC/D or cardiomyopathy or other forms of cardiomyopathies **(2)**
Usually benign. 5% to 20% spontaneous VT remission.

1. **Suner S, Simon HK, Feit LR, et al. Child with idiopathic ventricular tachycardia of prolonged duration. Ann Emerg Med. 1995; 25:706-709**.
2. **Chiu C, Sequeira IB. Diagnosis and treatment of idiopathic ventricular tachycardia. AACN Clin Issues. 2004; 15:449-461.**