CHRONIC CHAGASIC CARDIOMYOPATHY IN YOUNG WOMAN WITH ONE EPISODE OF VERY FAST BROAD SUSTAINED TACHYARRHYTHMIA

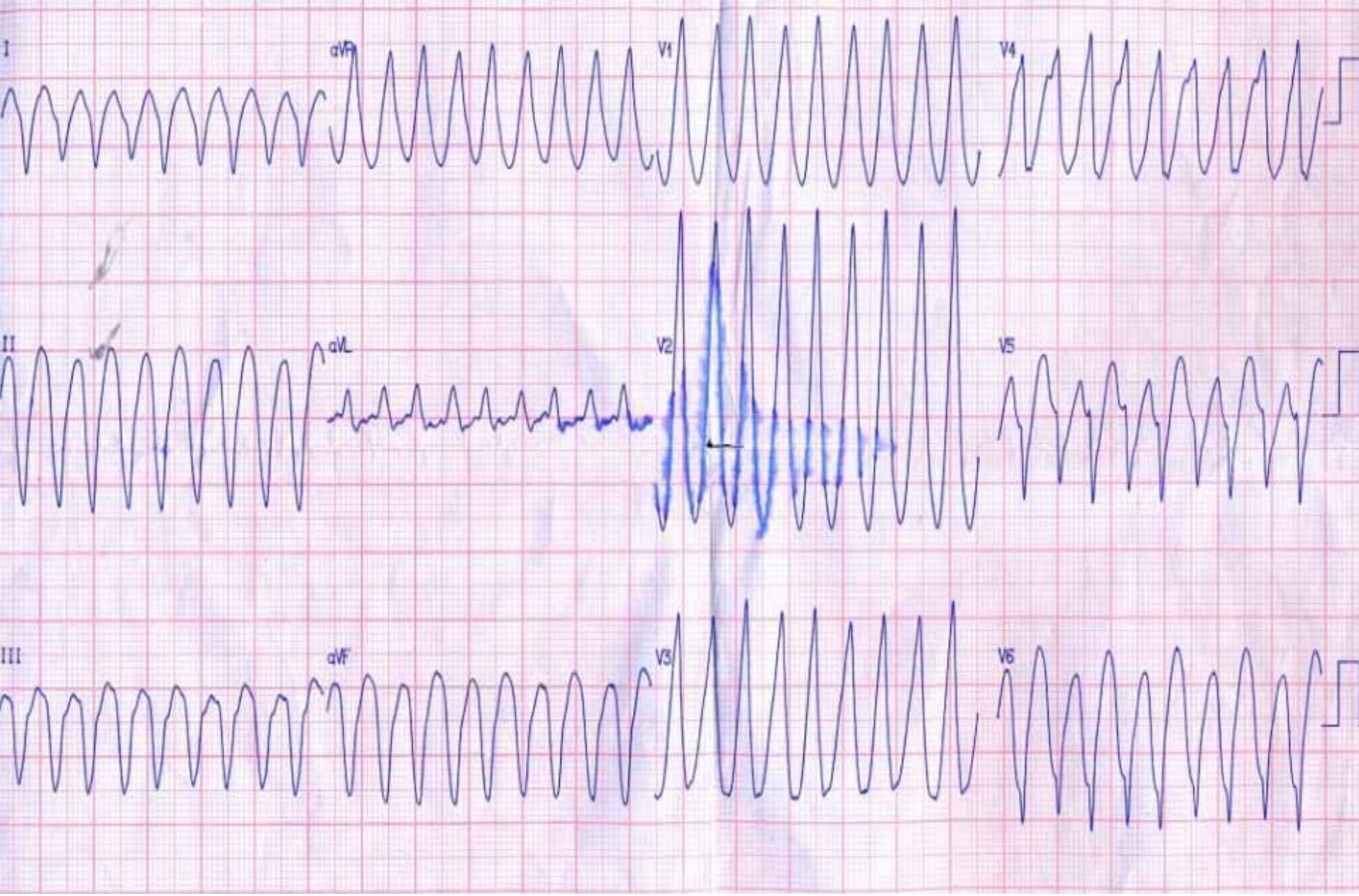
CRADIOMIOPATIA CHAGÁSICA CRÔNICA EM MULHER JOVEM COM UM EPISODIO DE TAQUIARRITMIA SUSTENTADA DE QRS LARGO

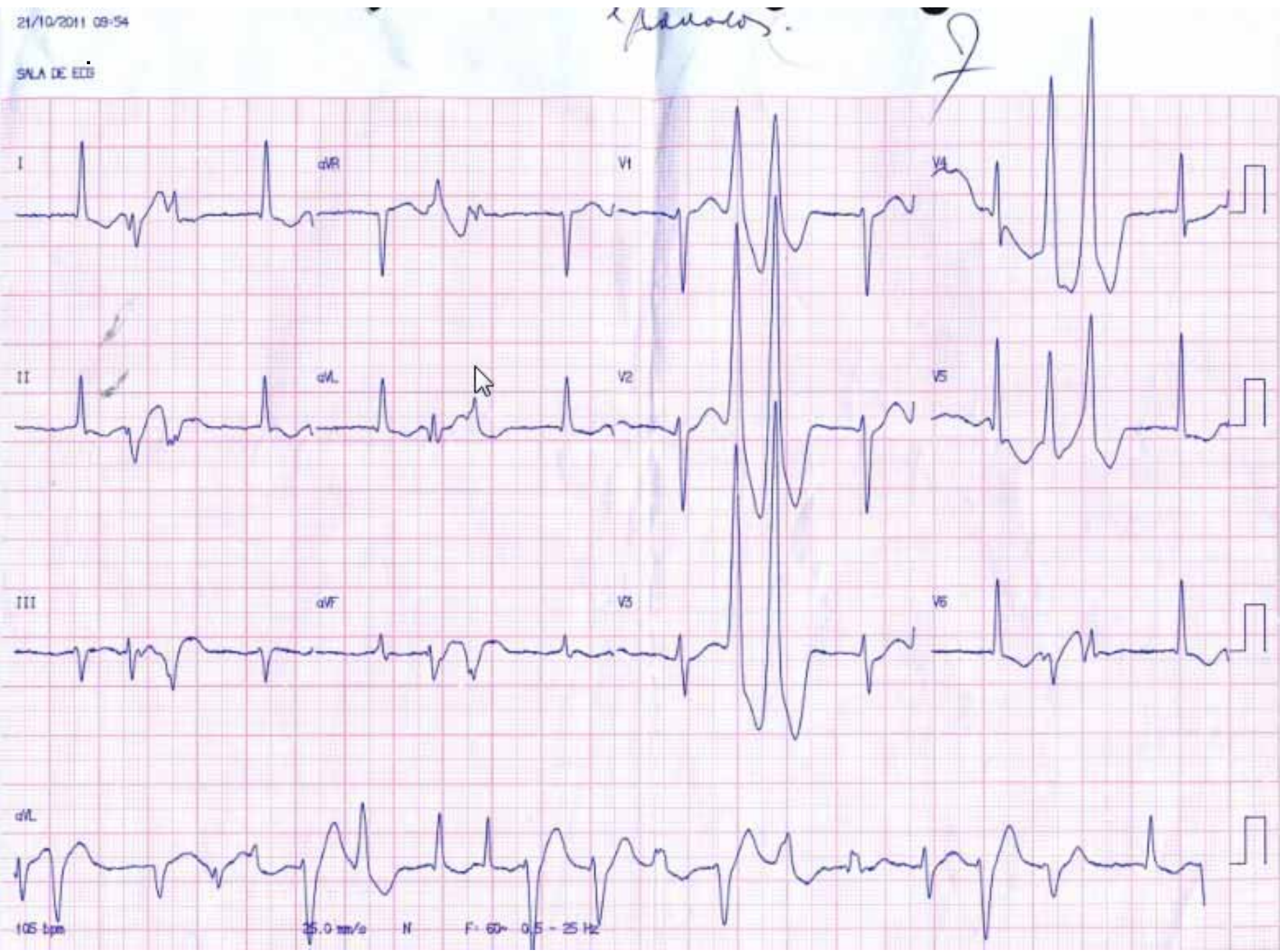
Case report from Raimundo Barbosa Barros MD
Nickname " The Fox"
Chief of Coronary Center Hospital de Messejana Dr. Carlos Alberto Studart Gomes
Fortaleza-Ceará-Brazil

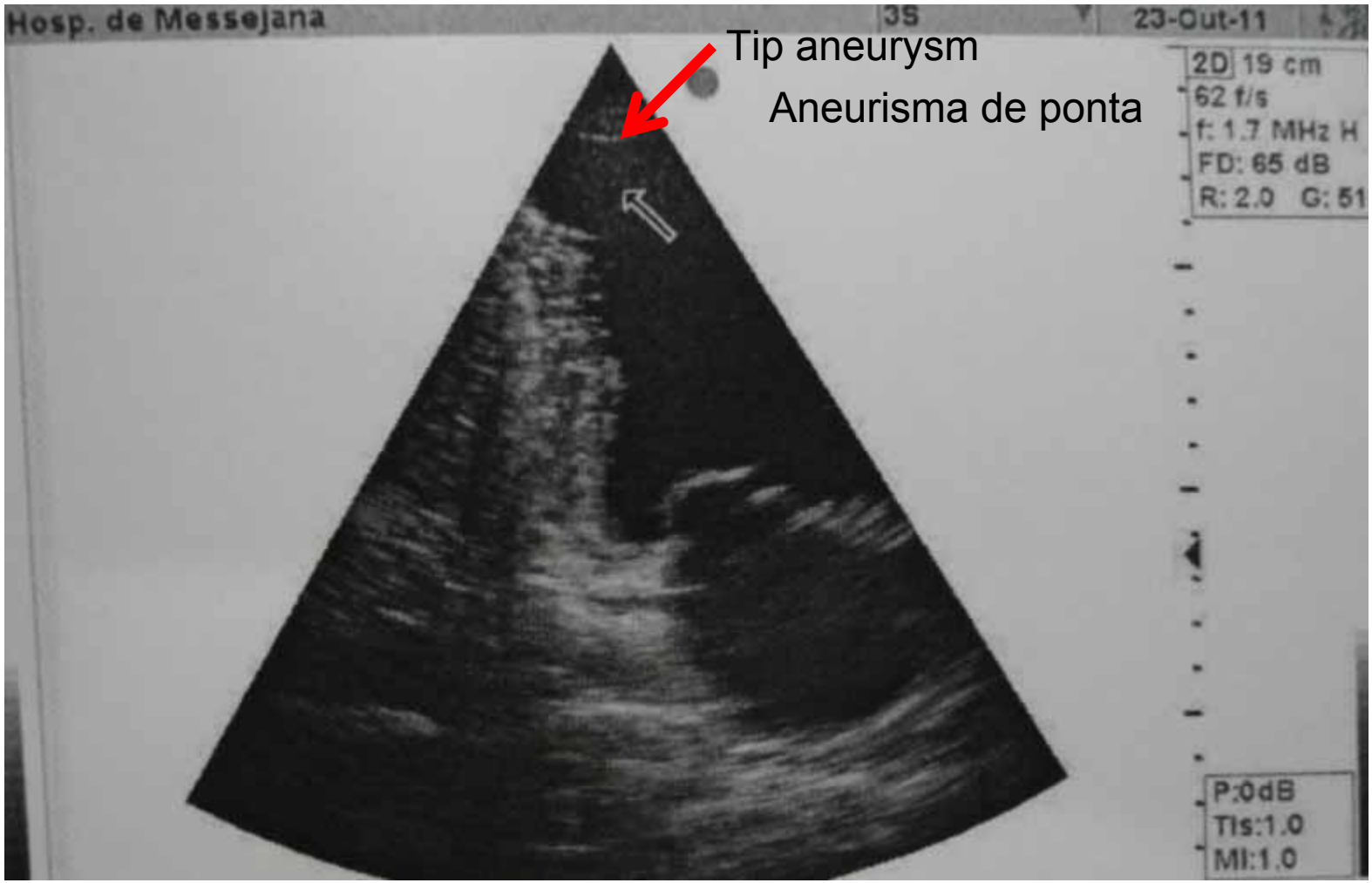
Final commentaries by: Andrés Ricardo Pérez-Riera M.D. Ph.D.
In charge of Electro-vectorcardiogram sector-Cardiology Discipline-ABC Faculty – ABC Foundation – Santo André – São Paulo – Brazil
Nickname: "The wild horse of the pampas"

Caso Clínico/Clinical case report

- **Mulher jovem,32 anos,com relato de palpitações e síncope(primeiro e único episódio)**
- **Admitida com taquicardia ventricular sustentada, submetida à cardioversão elétrica e seguida de infusão de amiodarona e transferida para o nosso hospital.**
- **A paciente não relata qualquer sintomatologia prévia.**
- **Sorologia positiva para Chagas(2 métodos)**
- **Atualmente em classe funcional NYHA I.**
- **Em uso de amiodarona vía oral,captopril,carvedilol, aldactone e warfarina**
- **Qual o diagnóstico da taquiarritmia?**
- **Qual a abordagem adequada neste caso?**
- **----------**
- **Young woman, 32 years, with a history of palpitations and syncope (first and only episode). Admitted with sustained ventricular tachycardia, and submitted to electrical cardioversion followed by amiodarone infusion and transferred to our hospital.**
- **The patient did not report any prior symptoms.**
- **Positive serology for Chagas (2 methods)**
- **Currently in NYHA functional class I.**
- **In use of oral amiodarone, captopril, carvedilol, aldactone and warfarina**
- **What is the diagnosis of tachyarrhythmia?**
- **What is the proper approach in this case?**


I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6


21/10/2011 09:54
SALA DE ECG
I
aVR
V1
V4
II
aVL
V2
V5
III
aVF
V3
V6
aVL
105 bpm
25.0 mm/s
N
F- 60~ 0.5 - 25 Hz


Hosp. de Messejana
3S
23-Out-11
Tip aneurysm
Aneurisma de ponta
2D 19 cm
62 f/s
f: 1.7 MHz H
FD: 65 dB
R: 2.0 G: 51
P:0dB
TIs:1.0
MI:1.0

Ecocardiogramas evolutivos/ Evolutionary Echocardiograms

- 23.10.2011 October 23, 2011
  - VE **LV=** 69/57
  - AE **LA**=39
  - FE **EF**=35%
  - Hipocinesia difusa severa**Severe diffuse hypokinesis**
  - I.mitral moderadaModerate mitral regurgitation
  - Aneurisma apical em “dedo de luva” Apical aneurysm in "glove finger"
- 07.11.11 November, 07 2011
  - VE LV=62/48
  - AE LA=35
  - FE EF=45%
  - Hipocinesia leve a moderadaMild to moderate hypokinesia
  - Insuficiência mitral leve Mild mitral regurgitation
  - Aneurisma apical Apical aneurysm in "glove finger"

Holter Monitoring

Ectopias ventriculares polimórficas, isoladas e pareadas.Nenhum registro de TV.Não relatou sintomas.

Polymorphic Premature Ventricular Contractions, isolated and couplets. Withouth record of VT. Asymptomatic.

Colleagues opinions

Andres,

This is an impressive presentation with an absolutely frightening episode of Ventricular Tachycardia. I am not sure that it has sufficient variability to call it polymorphic VT and my diagnosis would be very rapid monomorphic VT. I wonder what her rhythm was when she was syncopal or how fast the VT was at that time. Clearly she had time to get to the hospital, for the physicians to start an IV, administer intravenous amiodarone and then shock her implying that she was in VT the entire time. QRS alternation can be seen in Lead 1 and V6 but is most marked in V5. On the 12 lead ECG post-conversion, the rhythm strip is not simultaneous with the 12 lead ECG. There are frequent multifocal ventricular couplets and short runs of multifocal nonsustained VT. The ectopic beats in Leads V1-3 look identical to the morphology of the VT complexes on the initial ECG. The underlying rhythm is unclear. On the last complex in Leads 1 and 2, there appears to be a small bump preceding the QRS by approximately 280-300 ms. If this is a P wave in which case there is First Degree AV block? The atrial rhythm could also be a fine atrial fibrillation and I was simply seeing a slightly larger fibrillation wave. In addition, Chagas disease commonly (nothing is ever always and this is more frequent late in the disease) is usually associated with bundle branch block and AV block. Her presumably supraventricular QRS is normal at this time. What is intriguing is that the QT interval on the narrow, non-premature complexes is short at approximately 320 ms. In addition to her Chagas disease, could she have the short QT interval syndrome?

We were told that she was serologically positive for Chagas disease but was NYHA Functional Class 1 with respect to Heart Failure. The echo identifies a significantly dilated LV, and a reduced ejection fraction (35%). It is well known that left ventricular ejection fraction does not necessarily correlate with symptoms of heart failure so her NYHA Functional Class 1 status does not concern me. In that she is on multiple medications for the treatment of poor ventricular function including an ACE inhibitor (captopril), an adrenergic blocker (carvedilol) and spironolactone and now is NYHA functional class I, I would presume that her medications are being very effective. I am intrigued that she was also on an oral anticoagulant and wonder why – is that for the apical aneurysm on her echo or do prior ECGs confirm the presence of atrial fibrillation?

The VT with its RBBB marked right axis morphology could be arising from the RV apical aneurysm in which case, surgical resection of the aneurysm may be a functional cure. This would require an EP study to either induce the VT from this area of the heart or reproduce the morphology of the VT with pace-mapping. Still with the poor ventricular function and her being on amiodarone when the VT occurred, I would not want to rely on a surgical procedure in a young individual with a diffuse cardiomyopathy (Chagas disease) as being totally curative. There is also the question of Short QT Syndrome that I raised that, at a minimum, warrants review of multiple prior ECGs before confirming or excluding it as a diagnosis but if she has Short QT Syndrome, this would be another indication for an ICD. As such, I recommend implantation of an ICD for secondary prevention. I would choose a dual chamber ICD (at this time, she does not need cardiac resynchronization therapy) in the hope of pacing the atrium at a faster rate to help overdrive some of the ventricular ectopy. If on invasive EP testing or at the time of implant, she is confirmed to have atrial fibrillation, she should get a single chamber ICD although some might argue in favor of still implanting a dual chamber ICD in the hope of someday restoring a sinus rhythm or sinus bradycardia either pharmacologically or with pulmonary vein isolation or other ablation techniques. In placing the RV ICD lead, I would place it high on the interventricular septum intentionally keeping it away from the RV apical aneurysm. Even with an ICD which can rescue the individual, I would pursue an EP study and IF the VT is proven to be arising in the apical aneurysm, I would want to surgically excise it at which time the ICD becomes primary prevention in view of the poor LV function.

I await the final analysis and decisions with interest.

Paul

Paul A. Levine MD, FHRS, FACC, CCDS
25876 The Old Road #14
Stevenson Ranch, CA 91381
Cell: 661 565-5589
Fax: 661 253-2144
Email: paul91321@gmail.com

Amigos: me impresiona una miocardiopatia dilatada con fibrilación auricular. La taquiarritmia es muy preocupante. El antecedente de taquicardia ventricular asociado a miocardiopatia dilatada en adulto joben señala una conducta agresiva, la que a mi ver puede ser CDI o trasplante cardiaco. La FE mejoro en el segundo ecocardiograma pero tiene aneurisma de punta y el corazón bastante dañado. El tema trasplante es digno de una discusión de parte de Edgardo sin duda.
Emilio Marigliano

Friends: I'm impressed a dilated cardiomyopathy with atrial fibrillation. Tachyarrhythmia is very worrying. The history of ventricular tachycardia associated with dilated cardiomyopathy in young adult is indicative of aggressive approach, which in my view can be CDI or heart transplantation. The EF improved in the second echocardiogram but has tip aneurysm and heart badly damaged. The transplant issue is worthy of a discussion on the part of Edgardo no doubt.

Emilio Marigliano MD

Dear Friends: Thanks for sharing this beautiful case with us.

Summary: Woman presenting with sustained VT despite treatment with amiodarone, positive serology for Chagas, low LVEF. This is class I indication for an ICD (1;2;3;4).

The question remains if this is the only therapy that this patient may receive. Endocardial plus epicardial ablation is an option if the patient remains with sustained VT despite treatment with amiodarone (5;3).

As the VT suggest origin in the posterobasal region, rather than from the aneurysm region (please keep in mind that this patient may have other aneurysms...not seen in the echo), the resection (aneurismectomy) does not appear as a good option.

Summary: 1. ICD, 2. Reload with Amiod, 3. If VT persists, RF ablation with both endocardial plus epicardial approach.

Best

Baranchuk, Adrian MD FACC

1. Muratore C, Rabinovich R, Iglesias R, González M, Darú V, Liprandi AS. Implantable cardioverter defibrillators in patients with Chagas' disease: are they different from patients with coronary disease? Pacing Clin Electrophysiol. 1997 Jan;20:194-197.
2. Rassi A Jr, Rassi A, Rassi SG. Predictors of mortality in chronic Chagas disease: a systematic review of observational studies. Circulation. 2007 Mar 6;115:1101-1108.
3. Muratore CA, Baranchuk A. Current and emerging therapeutic options for the treatment of chronic chagasic cardiomyopathy. Vasc Health Risk Manag. 2010 Aug 9;6:593-601.
4. di Toro D, Muratore C, Aguinaga L, Batista L, Malan A, Greco O, Benchetrit C, Duque M, Baranchuk A, Maloney J. Predictors of all-cause 1-year mortality in implantable cardioverter defibrillator patients with chronic Chagas' cardiomyopathy. Pacing Clin Electrophysiol. 2011 Sep;34:1063-1069.
5. Sosa E, Scanavacca M. Images in cardiovascular medicine. Percutaneous pericardial access for mapping and ablation of epicardial ventricular tachycardias. Circulation. 2007 May 29;115: 542-544.

Estimados amigos: Gracias por compartir este hermoso caso con nosotros.
Resumen: mujer que presenta TV sostenida a pesar de tratamiento con amiodarona, serología positiva de Chagas, FEVI baja. Ésta es una indicación clase I para CDI (ver Muratore PACE 1997, 1999; RassiCirculation 2007, Muratore Branchuk Vasc Health manag 2010, Ditoro PACE2011, etc).
La pregunta sigue siendo si ésta es la única terapia que este paciente puede recibir.

La ablación endocárdica más la epicárdica es una opción si el paciente permanece con TV sostenida a pesar de tratamiento con amiodarona (ver Sosa y Scanavacca, ver Muratore Baranchuk).

Como la TV sugiere origen en la región póstero-basal, en vez de la región deel aneurisma (por favor tener presente que este paciente puede tener otrosaneurismas...que no se ven en eco), la resección (aneurismectomía) no parece ser una buena opción.

Resumen: 1. CDI, 2. Recarga con amiodarona, 3. Si la TV persiste, ablación por RF con un abordaje endocárdico y epicárdico.

Saludos,

Dr Baranchuk, Adrian MD FACC
Professor of Medicine Cardiac Electrophysiology and Pacing
Director, EP Training Program Kingston General Hospital FAPC 3, 76 Stuart Street K7L 2V7, Kingston ON Queen's University Ph: 613 549 6666 ext 3801
Fax: 613 548 1387 barancha@kgh.kari.net

Dear Dr. Adrian,

The patient was not using medications before the episode of sustained VT, only after cardioversion. Also, take into account that there was an improvement in the ejection fraction after optimizing the therapy with amiodarona (35-40%) and other drugs.

Thank you,

Raimundo

Dear Dr Adrian

El paciente no estava haciendo uso de medicamentos antes del episodio de TV sostenida. La droga solo fue administrada después de la cardioversión.
También tenga en cuenta que hubo una mejoría en la fracción de eyección después de la optimización de la terapia (35%-45%) con amiodarona y otras drogas

Gracias

Raimundo

Dear Raimundo
Thanks for the clarification. My position does not change regarding the ICD.
The patient should receive ICD plus Amiodarone. Why?
It is unpredictable how the next episode will be.
There is enough data demonstrating that the only effective treatment to prevent sudden death is an ICD, in cases of structural heart disease as the one you presented.
Amiodarone should be given anyway in order to reduce the arrhythmic burden.n
Of course, ablation should be only considered if Amiod fails to reduce VT burden.
Best
AB

Querido Raimundo:
Gracias por la aclaración. Mi postura no cambia con respecto el CDI.
El paciente debe recibir CDI más amiodarona. ¿Por qué? No puede predecirse como será el próximo episodio.
Hay datos suficientes que demuestran que el único tratamiento efectivo para evitar la muerte súbita es un CDI, en casos de cardiopatía estructural como la que Ud presentó.
La amiodarona debe administrarse de todos modos para reducir la carga arrítmica. Por supuesto, debe considerarse ablación si la amiodarona no reduce la carga de TV.
Saludos,
AB

According to the case presented, there is indication of ICD, in spite of the mismatching of the EF in Echo. History of syncope episode + VT in the presence of amiodarone, we consider it recurring SMVT. Our criterion when dealing with a first episode of VT with EF > 40 is administering amiodarone and if there is recurrence, ICD. If the EF is < 35-40% we implant an ICD, according to the AVID.

Oscar Pellizon

De acuerdo al caso presentado tiene indicacion de CDI, a pesar de la discordancia de la FE en los Ecos. antecedente de episodio sincopal + TV en presencia de amio la consideramos un TVMS recurrente. nuestro criterio ante un primer episodio de TV con FE > de 40 le administramos amio y ante la recurrencia CDI. si la FE es < 35-40% le implantamos un CDI, de acuerdo a AVID.

Oscar Pellizón

Hello, Oscar!

In the Argentine Congress we presented along with Di Toro and Muratore, a long series of patients with Chagas disease and ICD with secondary prevention and EF < and < of 40%. To our surprise, there were no differences as to: mortality, appropriate and inappropriate shocks between the 2 populations.
We are writing the manuscript because we think that there is an important message: if a chagasic patient with preserved EF (>40%) shows up with sustained VT, he/she dies the same as if he/she had depressed EF.

Interesting, isn't it?

AB

Hola Oscar
En el congreso Argentino presentamos junto a Di toro y Muratore, una serie larga de pacientes con chagas y CDI con prevencion secundaria y FEy < y < del 40%. Para nuestra sorpresa, ni hubo diferencias en cuanto a: mortalidad, choques apropiados e inapropiados entre las 2 poblaciones.
Estamos escribiendo el manuscrito porque creemos que es un mensaje importante: si un chagásico con Fey conservada (>40%) se presenta con TV sostenida, se muere igual que si tuviera Fey deprimida.
Interesante no?
AB

Hello, AB. Send me these data or when you have the manuscript. It seems to be very interesting, since I see many chagasic patients.
Best regards,

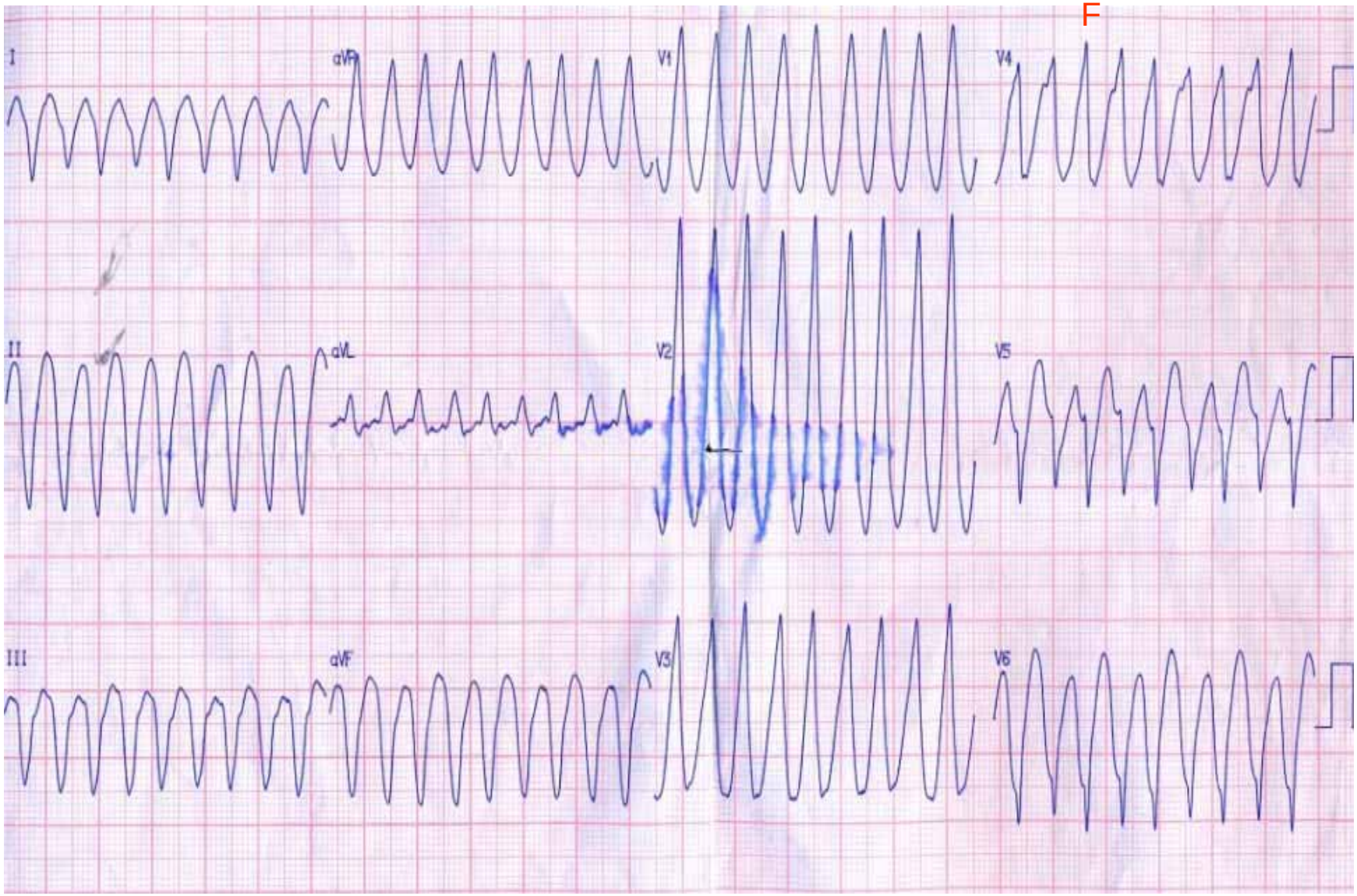
Oscar

Hello AB, mandame esos datos o cuando tengas el manuscrito. me parece muy interesante, ya que veo muchos chagasicos. abrazo.

Oscar Pellizón.

FINAL COMMENTARIES

By Andrés Ricardo Pérez-Riera M.D. Ph.D.



Very Fast Sustained Monomorphic VT(HR 250bpm: six small box between R apex). Monomorphic ventricular tachycardia means that the appearance of all the beats match each other in each lead of a surface electrocardiogram

Presence of AV dissociation: fusion beats (F) relatively narrow QRS complex = VT

Focus: Left ventricle Why? Because V1 –positive broad QRS tachycardia with monophasic complex. And where in LV?

Answer: posteroinferobasal region. Commentaries: VT may arise from various regions in both ventricles, but LV inferolateral scar is the main source of S-VT reentrant circuits in chronic chagasic cardiomyopathy (CCC). Additionally, there is good topographic correlation between myocardial perfusion, wall motion abnormalities and areas that originate S-VT.

Finally, although to a lesser extent, wall motion and perfusion defects also occur in a relevant proportion of chagasics with NS-VT. (1).

In CCC, the amount of sympathetically denervated viable myocardium is associated with the occurrence of S-VT.

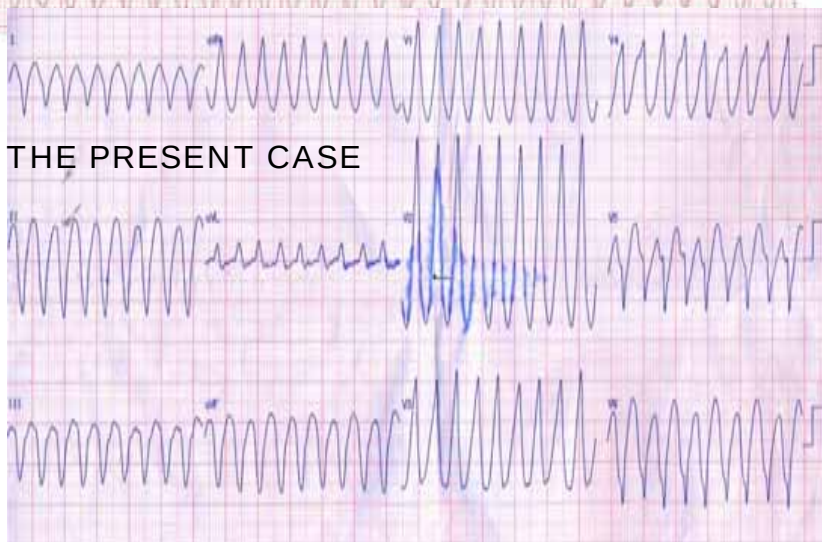
Myocardial sympathetic denervation may participate in triggering malignant ventricular arrhythmia in CCC also in patients with relatively well-preserved ventricular function(2).!!!!!

Additional commentaries: In some leads, mainly from V1 to V4 the tachyarrhythmic event look like a ventricular flutter. It is ventricular tachycardia with a HR over 180 beats/min (near 300 bpm without consensus Some authors admit 180-250 bpm) mostly caused by re-entry, the ECG shows a typical sinusoidal pattern, with large amplitude, without clear definition of the QRS and T waves. It has been considered as a possible transition stage between VT and VF, and is a critically unstable arrhythmia that can result in SCD. During ventricular flutter the ventricles depolarize in a circular pattern, which prevents good function. Most often this results in a minimal cardiac output and subsequent ischemia. The next slide shows a typical 12-EECG with ventricular flutter and the present case.

1. **Sarabanda AV, Sosa E, Simões MV, Figueiredo GL, Pintya AO, Marin-Neto JA. Ventricular tachycardia in Chagas' disease: a comparison of clinical, angiographic, electrophysiologic and myocardial perfusion disturbances between patients presenting with either sustained or nonsustained forms.Int J Cardiol. 2005 Jun 22;102:9-19.**
2. **Miranda CH, Figueiredo AB, Maciel BC, Marin-Neto JA, Simões MV. Sustained ventricular tachycardia is associated with regional myocardial sympathetic denervation assessed with 123I-metaiodobenzylguanidine in chronic Chagas cardiomyopathy. J Nucl Med. 2011 Apr;52:504-510.**

12-ECG leads TYPICAL ventricular flutter

Ventricular flutter: a possible transition stage between VT and VF, the ECG showing rapid, uniform, regular oscillations, ≥ 250 or more per minute.It is not possible to distinguish if the tracing is being observed the other way round: symmetrical tracing. Ventricular flutter is a major cardiovascular emergency and therefore the treatment should be done as quickly as possible. This consists in CPR measures and the administration of external electric shock of 200-400 joules, as soon as possible.

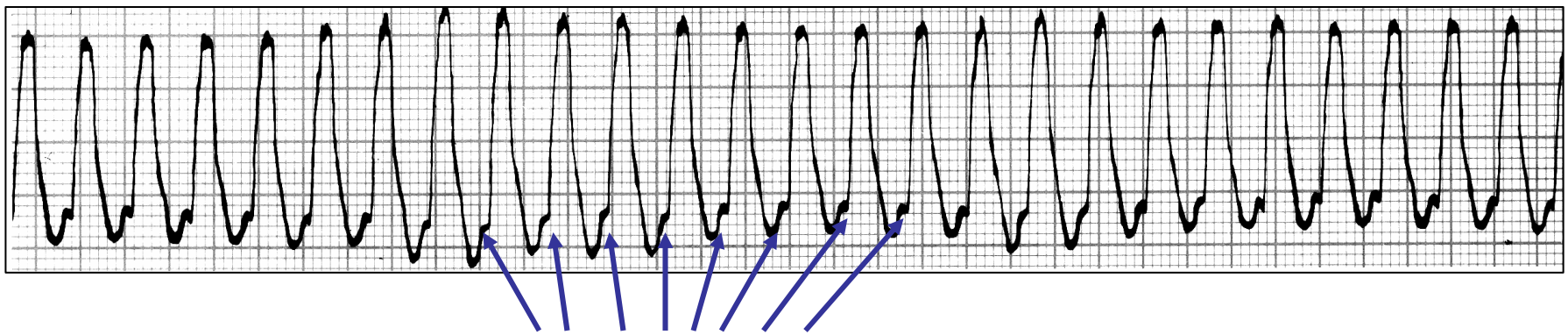


ELECTROCARDIOGRAPHIC CHARACTERIZATION OF VENTRICULAR FLUTTER

1. Waves with sinusoidal aspect (“sine wave”), constant or uniform configuration, regular, very wide, broad (>120 ms) and with equal ascending and descending limbs. They were called “Zigzag pattern”;
2. Absence of isoelectric line between the waves;
3. ECG seems equal if observed from top to bottom or vice-versa; in other words, it is not possible to distinguish if it is being observed the other way round: symmetrical tracing;
4. Regular and rapid rate between 150 bpm and 300 bpm. Nearly always >180 bpm and rarely <250 bpm;
5. It is impossible to identify the components of depolarization (QRS) and repolarization (ST/T), i.e. to determine when one ends and the other begins. This is the basic point for the differential diagnosis with very fast VT.
6. We rarely observe small notch right at the base of the ascending wave, after the QRS corresponding to retrograde activation of the atria (P wave);
7. Sometimes the P wave is observed independently from ventricular activity;
8. Duration nearly always brief;
9. In nearly all cases, it becomes (degenerates into) VF.

VENTRICULAR FLUTTER

MONITOR LEAD



RETROGRADE P WAVE?

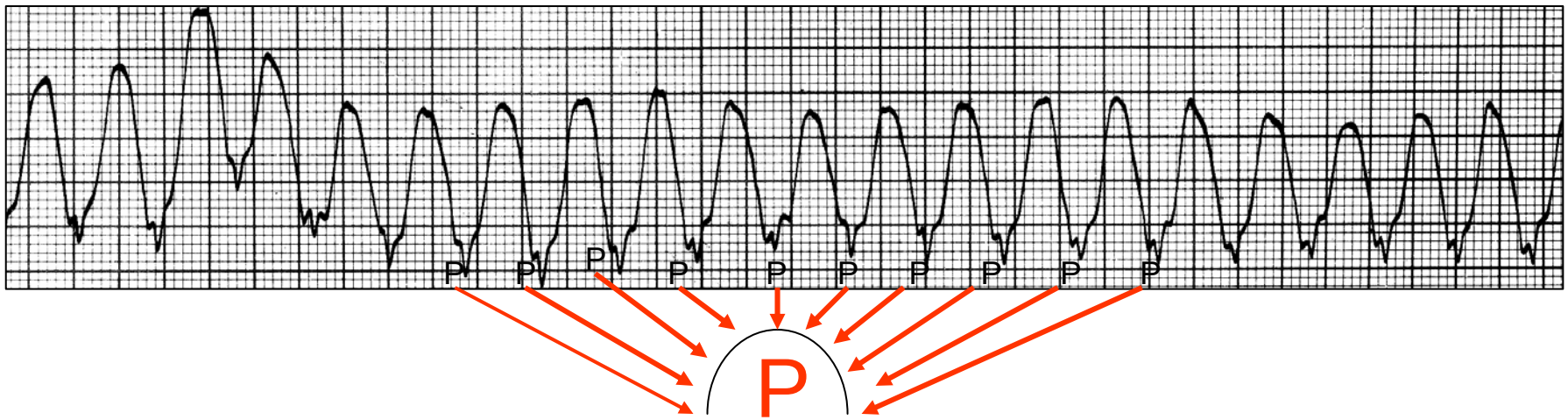
PART OF THE QRS COMPLEX?

Heart rate of 230 bpm. Broad and sinusoidal complexes. It is not possible to distinguish QRS from ST/T, there is no isoelectric line between the waves, zigzag pattern, symmetrical tracing.

Examples of ventricular flutter in the monitor.

VENTRICULAR FLUTTER WITH RETROGRADE ACTIVATION OF THE ATRIA

MONITOR LEAD



P WAVES RETROGRADE IN RELATION TO THE ATRIA AFTER EACH QRS COMPLEX

Ventricular flutter with a rate of 166-187 bpm. Broad and sinusoidal complexes.

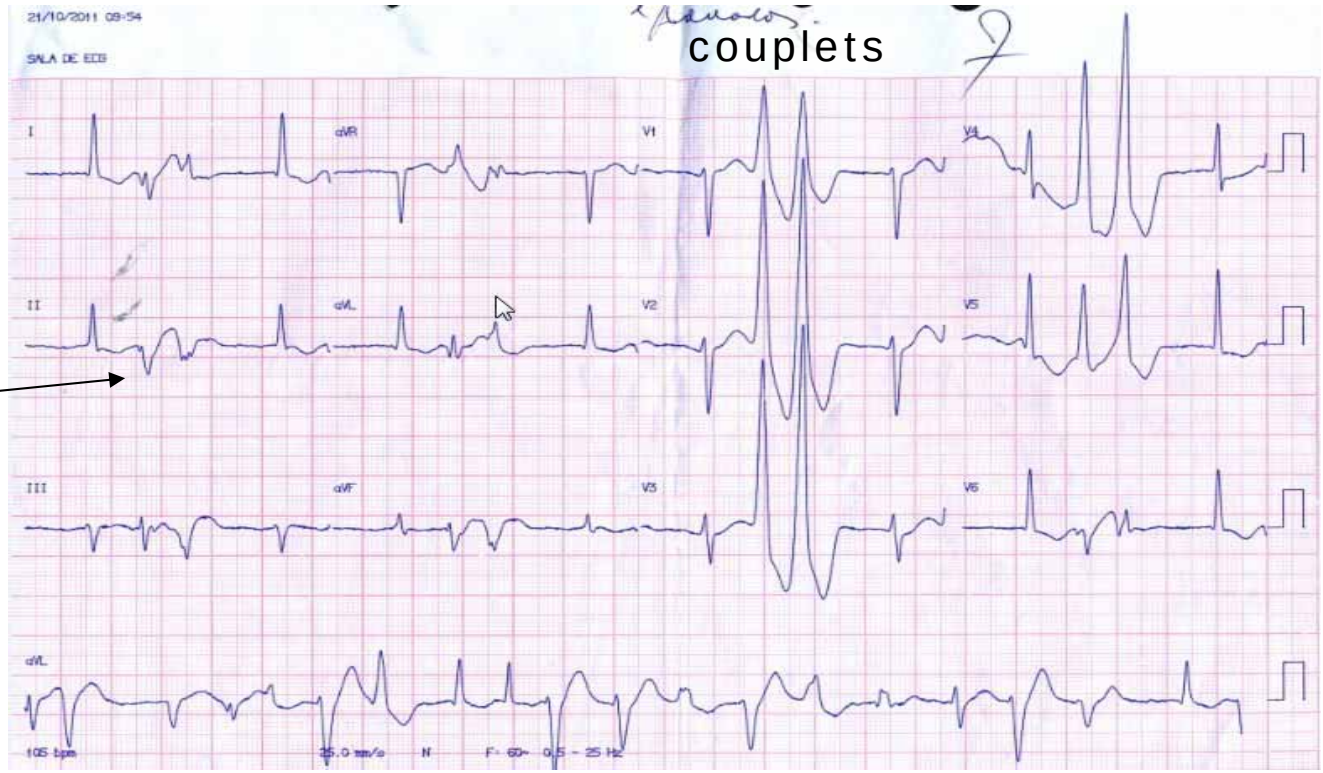
It is not possible to distinguish QRS from ST/T, there is no isoelectric line between the waves, zigzag pattern, symmetrical tracing.

In conclusion what is the difference between very fast ventricular tachycardia and ventricular flutter?

Answer: In the very fast monomorphic ventricular tachycardia is possible to distinguish between what is QRS and what is the ST/T. Not so in the ventricular flutter.

Chagas' disease is the main cause of bundle branch block and AV block in endemic areas.
In advanced cases of Chronic Chagasic Cardiomyopathy, PVCs are extremely frequent, multiform, and repetitive (couplets and runs of NS-VT), and show R on T phenomenon.
These arrhythmias are usually aggravated by increased sympathetic tone, implying an enhanced risk of SCD among chagasic patients, which is sometimes the first manifestation of the illness.
Chronic chagasic Cardiomyopathy is the leading cause of cardiovascular death, mostly as a consequence of heart failure and sudden death, in areas where the disease is endemic.(1)

Multifocal PVCs



Short runs of multifocal NS-VT

1. **Elizari MV, Chiale PA. Cardiac arrhythmias in Chagas' heart disease. Cardiovasc Electrophysiol. 1993 Oct;4:596-608.**

And what is the proper approach in this case? The answer is not easy. Why?

Because recently, Sarabanda and Marin-Neto (1) studied the outcome of 56 patients with Chronic Chagasic Cardiomyopathy (CCC) mean age of 55 years; mean LVEF42% presenting with either S-VT or NSVT before therapy with ICD. Over a mean follow-up of 38 ± 16 months, 16 patients (29%) died, 11 due to SCD, and five from progressive heart failure. Survivors and nonsurvivors had comparable baseline characteristics, except for a lower LVEF (46 ± 7% vs 31 ± 9%, P < 0.001) and a higher NYHA class (P = 0.003) in those who died during follow-up. Receiver-operator characteristic curve analysis showed that an LVEF **cutoff value of 38% had the best accuracy for predicting all-cause mortality and an LVEF cutoff value of 40% had the best accuracy for prediction of SCD.** Using the multivariate Cox regression analysis, LVEF < 40% was the only predictor of all-cause mortality. The authors conclude that patients with Chronic Chagas' cardiomyopathy presenting with either sustained VT or NSVT run a major risk for mortality when had concomitant severe or even moderate LV systolic dysfunction. This patient is border line. To day she is in NYHA class is I and the EF=45%. These researchs conclusions are different to preliminary observations of Di Toro and Muratore aforementioned by our dearest friend Adrian. *VT* in patients with structural heart disease is typically due to a “reentry” mechanism, where the electrical current travels in a loop of tissue that is scarred with slowed electrical conduction. This allows the rhythm to continually perpetuate itself, as the head of the myocardial electrical current chases its tail. In this case RFCA with both endocardial plus epicardial RFCA (EPRFCA), is the correct approach. As damage to coronary arteries is a potential complication of EPRFCA the procedure must be associated with coronary angiography. CCC is a disease where epicardial VT are common. Eletroanatomic mapping merged with computed tomography (CT) scan data is a useful tool for mapping the endocardium, and its accuracy in guiding ablation on the epicardium was not adequately evaluated so far.

1. **Sarabanda AV, Marin-Neto JA. Predictors of mortality in patients with Chagas' cardiomyopathy and ventricular tachycardia not treated with implantable cardioverter-defibrillators. Pacing Clin Electrophysiol. 2011 Jan;34:54-62.**

The combination of electroanatomic map and CT coronary artery scan data is feasible and can be an important tool for EPRFCA in patients with CCC and VT. (1)
Additionally maintenance of heart failure theraphy with carvedilol 50 mg/day, enalapril maleate, furosemide, espironolactone and amiodarone is very important. If EF ≥45% is not necessary ICD.(polemic).

1. **Valdigem BP, da Silva NJ, Dietrich CO, Moreira D, Sasdelli R, Pinto IM, Cirenza C, de Paola AA. Accuracy of epicardial electroanatomic mapping and ablation of sustained ventricular tachycardia merged with heart CT scan in chronic Chagasic cardiomyopathy. J Interv Card Electrophysiol. 2010 Nov;29:119-125.**