The value of Electrocardiogram in Chronic Chagasic Cardiomyopathy - 2019

Andrés Ricardo **Pérez-Riera M.D.Ph.D.**; Edgardo **Schapachnik M.D.**

[a] Laboratório de Metodologia de Pesquisa e Escrita Científica, Faculdade de Medicina do ABC, Santo André, São Paulo, Brazil.
[b] CARDIOLATINA.COM

Corresponding author

Andrés Ricardo **Pérez-Riera**

Rua Sebastião Afonso, 885 Zip code: 04417-100 Jardim Miriam, São Paulo-SP, Brazil

Phone/Fax: (55) 11 5621-2390 / e-mail: riera@uol.com.br

Key words: Chagas' disease; Chronic Chagasic Cardiomyopathy; Electrocardiogram

Abstract

Chagas disease (ChD) is an important cause of cardiomyopathy in Latin America. The electrocardiographic hallmark of Chronic Chagasic Cardiomyopathy (CCC) is the association of complete right bundle branch block (RBBB) with left anterior fascicular block (LAFB) and polymorphic premature ventricular contractions. The scar pattern is particularly dense and transmural. CCC is the most frequent nonischemic substrate causing left ventricular tachycardia (VT) in Latin America. Compared with other etiologies of cardiomyopathies, On the other hand, CCC with heart failure have higher prevalence of stroke and pacemaker implantation and worse health-related quality of life.

Introduction

ChD or American trypanosomiases caused by the hemoflagellate protozoan Trypanosoma cruzi[1] is a major medical and social problem in Latin America and affects 8-10 million people worldwide mainly in Latin America where ChD remains one of the biggest public health problems, causing incapacity in infected individuals and

more than 10.000 deaths per year. For many decades ChD was a strictly rural disease. However socio-economic changes, rural exodus, deforestation and urbanization have transformed the epidemiological profile of the disease, rendering it to a more urban/peri-urban phenomenon.

In the last decades, with increased international migration, it has also become a problem for developed countries, which now have hundreds of thousands of patients around the word. The presence of ChD outside Latin America is due to population mobility, but cases of infection have been reported among travelers returning from Latin America. Subsequent (autochthonous) transmission arises mainly from blood transfusion, congenital means and transplantation routes. It was estimated that 24-92 newborns delivered by South American T. cruzi infected mothers in Spain may have been congenitally infected with T. cruzi in 2007. In the USA epidemiologic report estimated that 1.9% of approximately 13 million Latin American immigrants in 2000, and 2% of 17 million in 2007, were potentially infected with T. cruzi. Of these, 49,157 and 65,133 in 2000 and 2007 respectively, may have or may develop symptoms and signs of chronic ChD[2]. In the United States there are >300,000 ChD-infected individuals, of whom 30,000–45,000 are estimated to have undiagnosed[3]. As T. cruzi can be detected in the blood of untreated infected individuals, decades after infection took place; the infection can be also transmitted through blood transfusion and organ transplant, which is considered the second most common mode of transmission for T. cruzi[4]. Over the past 30 years, the rapid worldwide spread of HIV, in combination with the changing epidemiology of *T. cruzi* has led to the emergence of *T.cruzi*/HIV co-infections. Diagnosis of *T. cruzi* infection in HIV positive individuals is particularly difficult. When ChD reactivates, especially in a HIV patient, it behaves like a separate disease with acute features such as severe neurological symptoms. This can lead to misdiagnosis with other infections, the most common differential diagnosis observed being toxoplasmosis. Furthermore, traditional serological diagnostic tests for Chagas are found to be weaker and less sensitive, as HIV-positive

2

3

4

patients are less likely to build a strong antibody response against the infection. The spread of HIV pandemic has not only modified the pathological spectrum of Chagas but also its epidemiology. From the 1980s, when the first case of HIV/*T. cruzi* co-infection was described to today, cases have been reported in 9 countries.

There are five main clinical forms of chronic chagasic cardiomyopathy: indeterminate, arrhythmogenic (predominantly dromotropic and extrasystolic), with ventricular dysfunction, thromboembolic and mixed forms[5]. The clinical course of ChD is extremely variable, and although many individuals remain asymptomatic for long periods, approximately one-third of infected patients develop life-threatening heart disease, including malignant ventricular tachyarrritmias. Ventricular arrhythmias associated with Chronic Chagasic Cardiomyopathy have high rates of morbidity and mortality.

Value of the ECG in ChD

The ECG is the method of choice in longitudinal population studies in endemic areas because it is simple, with a low cost and a good sensitivity. Additionally, the ECG has prognostic value.

Main ECG features in CCC

Rhythm: frequent sinus node dysfunction: persistent sinus bradycardia(HR> 50bpm), SA block in different degrees, sinus arrest and inappropriate chronotropic response in stress test. The corrected recovery time of the SA node and SA conduction time are altered (18% to 30%).

Dromotropic alterations in the AV conductor system: type II first or second-degree blocks (14.3%) trifascicular block and even total AV block (2.5%).

Post-His: the most frequent ones are first degree AV blocks, with broad QRS, which in 50% of cases are located in the AV node and the rest in the His-Purkinje system or in both.

CRBBB + LAFB, negative T waves and polymorphic (PVCs) are typical features (25%). Rosenbaum named the LAFB with left axis deviation as left anterior hemiblock [6] based in experimental animal models. Chagas emphasized the notorious PVCs during physical examination, and stressed the presence of AV block, detected by Jacquet's polygraph since ECG was not available at that time in Brazil, as a cause of abnormalities in cardiac rhythm[7].

Electrically inactive areas by "apical aneurysm" are frequent.

Sustained ventricular tachycardia (S-VT) and/or no-sustained ventricular tachycardia (NS-VT) have their main focus predominantly on inferobasal and lateral regions[8], CC, with marked epicardial predominance. The scar pattern is particularly dense and transmural as compared with the more erratic/patchy scar patterns seen in other nonischemic cardiomyopathies. Endocardial unipolar voltage mapping serves to characterize epicardial scar in this setting. The main mechanism is reentry, involving fibrotic and/or LV apical akinetic aneurysmatic area. Sudden cardiac death (SCD) is one of the most characteristic phenomena of CCC. Dr Carlos Chagas described SCD associated with arrhythmias for the first time in 1911[9]. SCD is the most common cause of death in patients with ChD.

Characteristic VT in the setting of CCC

Epicardial > Endocardial; homogeneous basal-lateral LV involvement; scar pattern particularly dense and transmural as compared to other nonischemic cardiomyopathies; epicardial ablation needed in a vast majority of patient's with CCC-VT; endocardial unipolar mapping is useful to identify and characterize epicardial CCC scar.

In an study of 453 Latin-American's immigrants realized in Barcelona Spain[10] people the ECG abnormalities considered to be attributed to ChD were: isolated RBBB 27 (6%), isolated LAFB 80 (17.8%), isolated left posterior fascicular block 3 (0.7%), left

---

6

7

8

9

10

bundle branch block 1 (0.2%), PVCs 2 (0.4%), Q waves 2 (0.4%), ST-T changes 9 (2%), first degree atrioventricular (AV) block16 (3.5%), second degree atrioventricular (AV) block 0, complete AV block 1 (0.2%), low QRS voltage 9 (2%), sinus bradycardia <50 beats per minute 23(5.1%), atrial extrasystole 5 (1.1%), atrial fibrillation 3 (0.7%) or flutter, and pacemaker rhythm. LAFB was the more frequented ECG feature in this coorte.

Approximately one-third of infected patients develop life-threatening heart disease, including malignant ventricular arrhythmias. Fibrotic lesions secondary to chronic cardiomyopathy produce arrhythmogenic substrates that lead to the appearance and maintenance of ventricular events.

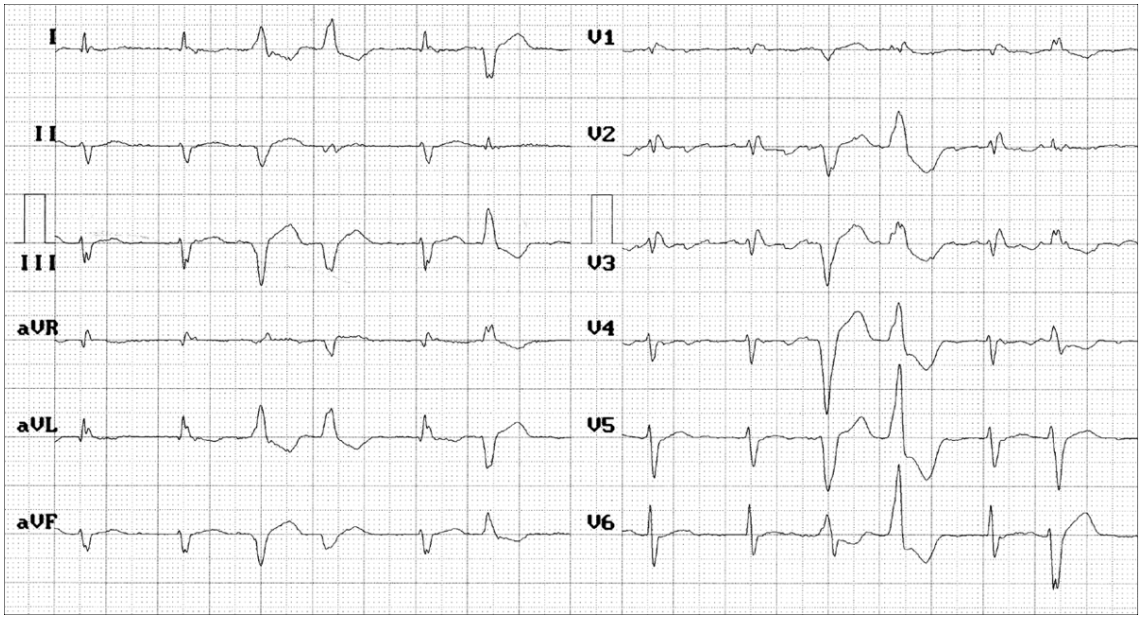
In 2010 5 (1.1%) we described a first Brugada electrocardiographic phenocopy in a patient with chronic CCC[11].

Electrocardiographic features of poor prognosis in CCC

- ➢ Presence of atrial fibrillation or flutter;
- ➢ Presence of CLBBB (rare) in 91.3% of the cases and decreased ejection fraction;
- ➢ Presence of total AV block;
- ➢ Presence of anterior and inferior electrically inactive area;
- ➢ Presence of polymorphic premature ventricular contractions or in salvoes;
- ➢ Presence of NS-VT associated to decreased EF: 80% of mortality in 13 years of follow-up. When the EF is normal, the prognosis is good;
- ➢ Presence of S-VT: 100% of mortality in five years.

Typical example of ECG of CCC in 40-year-old man from endemic rural area in Minas Gerais Brazil

11



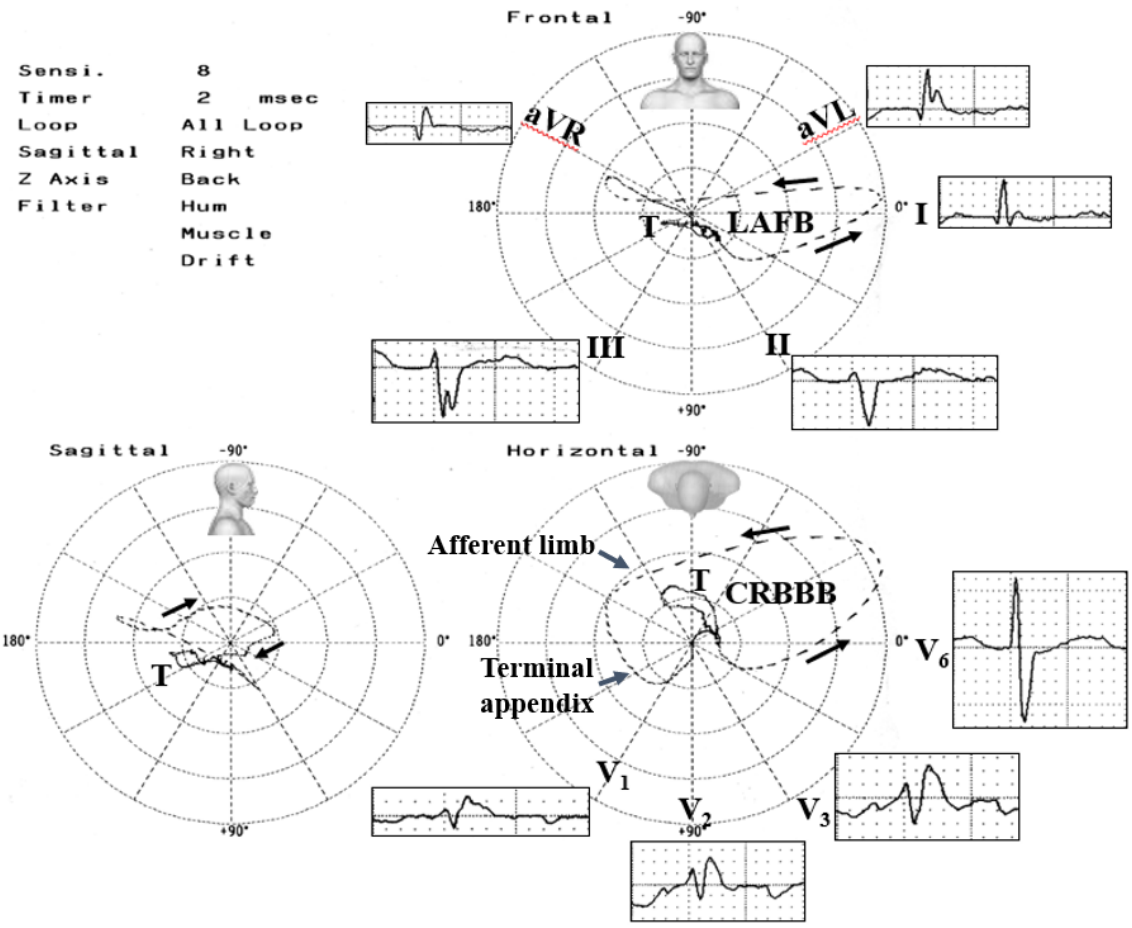
P wave difficult to visualize, which indicates intense fibrosis of atrial tissue.

LAFB: extreme shift of ÂQRS in the left superior quadrant, around –75º, qR in I and aVL, rS in inferior leads with S wave in V_5 and V_6

CRBBB: triphasic complex, rsr' type, from V_1 to V_3, broad r wave in aVR and S wave in V_5 and V_6 and coupled polymorphic premature ventricular contractions.

Classical triad: CRBBB + LAFB + polymorphic premature ventricular contractions.

ECG/VCG correlation of typical CCC



Frontal plane: QRS loop with typical counter clockwise rotation, QRS axis on left superior quadrant: LAFB.

Horizontal plane: typical Complete RBBB Vectorcardiographic Grishman or Kennedy type I variant: The QRS loop rotates in counter clock wise fashion, the afferent limb is located behind the X orthogonal limb and the terminal dots are closer one to another, reflecting end conduction delay with "glove finger-like" terminal appendix located in the right anterior quadrant. Since the RV is depolarized later than normal, the LV depolarizes without some of the normal cancelation from RV forces. The T-loop oriented to back.

Right sagittal plane: QRS loop predominant located on superior quadrants with terminal portion of QRS-loop on anterior quadrants. The T-loop oriented to back.

Conclusion: CRRR + LAFB.

Conclusion

The association of complete RBBB with LAFB and polymorphic PVCs are the hallmark of CCC. S-VT and/or NS-VT is almost homogeneously concentrated to the basal lateral LV, with marked epicardial predominance over the endocardium. The scarring process in CCC seems to be particularly dense and transmural as compared with other nonischemic cardiomyopathies. Unipolar endocardial mapping, with revisited voltage cutoff values, serves to both identify and depict the epicardial scar in the setting of CCC. SCD is the most common cause of death in patients with ChD. Severe bradycardia may develop as a result of sinus node dysfunction (SND) or third-degree AV block. This requires permanent pacing.

Conflict of interest

None.

References

1. Malik LH, Singh GD, Amsterdam EA.Chagas Heart Disease: An Update.Am J Med. 2015 Nov;128(11):1251.e7-9.doi: 10.1016/j.amjmed.2015.04.036Schmunis GA, Yadon ZE. Chagas disease: a Latin American health problem becoming a world health problem. Acta Trop. 2010 Jul-Aug;115(1-2):14-21. doi: 10.1016/j.actatropica. 2009.11.003
2. Schmunis GA, Yadon ZE. Chagas disease: a Latin American health problem becoming a world health problem. Acta Trop. 2010 Jul-Aug;115(1-2):14-21. doi: 10.1016/ j.actatropica.2009.11.003

3. Neeki MM, Park M, Sandhu K, Seiler K, Toy J, Rabiei M, Adigoupula S. Chagas Disease-induced Sudden Cardiac Arrest. Clin Pract Cases Emerg Med. 2017 Oct 9;1(4):354-358. doi: 10.5811/cpcem.2017.5.33626
4. Schmunis GA.Epidemiology of Chagas disease in non-endemic countries: the role of international migration.Mem Inst Oswaldo Cruz. 2007 Oct 30;102 Suppl 1:75-85.
5. Dubner S, Schapachnik E, Riera AR, Valero E.Chagas disease: state-of-the-art of diagnosis and management.Cardiol J. 2008;15(6):493-504.
6. Rosembaum MB. Chagasic myocardiopathy. Prog Cardiovasc Dis. 1964 Nov; 7:199–225.
7. Chagas C. Nova entidade mórbida do homem. ***"New morbid entity of man".*** Brazil-Médico. 1910; 45:443–447.
8. Soto-Becerra R, Bazan V, Bautista W, Malavassi F, Altamar J, Ramirez JD, Everth A, Callans DJ, Marchlinski FE, Rodríguez D, García FC, Sáenz LC.Ventricular Tachycardia in the Setting of Chagasic Cardiomyopathy: Use of Voltage Mapping to Characterize Endoepicardial Nonischemic Scar Distribution. Circ Arrhythm Electrophysiol. 2017 Nov;10(11). pii: e004950. doi: 10.1161/CIRCEP.116.004950
9. Chagas C. Nova entidade mórbida do homem. Resumo geral de estudos etiológico e clínico. ***"New morbid entity of man. General summary of etiological and clinical studies".*** Mem Inst Oswaldo Cruz. 1911; 3:3–59.
10. Sánchez-Montalvá A, Salvador F, Rodríguez-Palomares J, Sulleiro E, Sao-Avilés A, Roure S, Valerio L, Evangelista A, Molina I.Chagas Cardiomyopathy: Usefulness of EKG and Echocardiogram in a Non-Endemic Country.PLoS One. 2016 Jun 16;11(6):e0157597. doi: 10.1371/journal.pone.0157597
11. Arce M, Riera AR, Femenía F, Baranchuk A. Brugada electrocardiographic phenocopy in a patient with chronic Chagasic cardiomyopathy Cardiol J. 2010;17(5):525-7.