Relevance of the vectorcardiogram in the Brugada syndrome with "Northwest QRS axis"

Andrés Ricardo **Pérez-Riera**[a], MD PhD; Raimundo **Barbosa-Barros**[b], MD; Antonio **Thomaz de Andrade**[b], MD; Raiza **Pontes Rodrigues**[b], MD; Frank **Yanowitz**[c], MD; Rodrigo **Daminello Raimundo**[a], PhD; Luiz Carlos **de Abreu**[a,d], PhD; Kjell **Nikus**[e], MD PhD; Pedro **Brugada**[f], MD PhD

[a]Laboratório de Metodologia de Pesquisa e Escrita Científica, Centro Universitário Saúde ABC, Santo André, São Paulo, Brazil.

[b]Coronary Center of the Hospital de Messejana Dr. Carlos Alberto Studart Gomes, Fortaleza, Ceará, Brazil.

[c]Intermountain Medical Center, Intermountain Heart Institute, Salt Lake City, UT, United States; The University of Utah, Department of Internal Medicine, Salt Lake City, UT, United States.

[d]Graduate Entry Medical School, University of Limerick, Limerick, Ireland.

[e]Heart Center, Tampere University Hospital and Faculty of Medicine and Health Technology, Tampere University, Finland.

[f]Scientific Director, Cardiovascular Division, Free University of Brussels (UZ Brussel) VUB, Brussels, Belgium.

ADDRESS FOR CORRESPONDENCE

Andrés Ricardo **Pérez-Riera**

Rua Nicolau Barreto, 258, zip code: 04583-000 Vila Cordeiro, São Paulo-SP, Brazil

Phone/Fax: (55) 11 5044-6243 / e-mail: riera@uol.com.br

ABSTRACT

The outcome in the Brugada syndrome (BrS) is more benign in female than in male individuals. However, outcome could be adversely affected by sinus node dysfunction (SND). Long sinus pauses indicate an overlap between the phenotypes of BrS and SND. We present a 29-year-old woman with syncopal episodes at rest since adolescence.

KEYWORDS

Brugada syndrome; sinus node dysfunction; vectorcardiogram; block in the superior or subpulmonary division of the right bundle branch.

INTRODUCTION

Prominent S waves in the inferior leads in the electrocardiogram (ECG) may be caused by left anterior fascicular block (LAFB). Determining the QRS axis in the frontal plane and detailed analysis of the amplitudes of the S waves may aid in the differential diagnosis between LAFB and other ECG manifestations with prominent inferior S waves. An additional potentially useful diagnostic tool is vectorcardiography (VCG).

We report a case of a young female patient with a history of syncope and suspicion of Brugada syndrome (BrS) based on the ECG findings.

CASE REPORT

A young Caucasian female presented with repetitive episodes of sudden decrease in awareness without prodromal symptoms and tonic-clonic episodes at rest, especially during sleep at night. The attacks had begun at the age of 16. There was no sphincter release, palpitations, chest pain or agonal nocturnal respiration and the other family members interpreted the seizures as epilepsy. The patient consulted a neurologist who started anticonvulsant treatment. The medication had no effect on the “convulsive episodes”. The electroencephalogram and cranial computed tomography were normal.

At the age of 25, the patient had an episode of sudden malaise, pallor, sweating at rest, with a syncopal episode, which prompted consultation with a clinical cardiologist. The ECG raised a suspicion of BrS, and the patient was referred to an electrophysiologist, who interpreted the ECG findings as right bundle branch block associated with LAFB. Regarding the family history, an apparently healthy first-degree cousin died suddenly at rest from an unknown cause at the age of 40.

The patient came to our attention at the age of 29. She denied the use of tobacco, alcohol or illicit drugs. Physical examination revealed nothing significant.

The ECG showed extreme QRS axis deviation in the frontal plane and the automatic analysis program erroneously reported a diagnosis of LAFB, but the vectorcardiogram (VCG) ruled out this diagnosis.

In order to clarify the case, we requested electrolytes (normal), transthoracic echocardiogram (without apparent structural heart disease), another ECG (Figure 1), VCG (Figure 2) high resolution ECG, Holter monitoring, electrophysiological study (Figure 3), and genetic screening for the proband and family members (no results yet).

DISCUSSION

In rare cases of the BrS, bursts of spontaneous monomorphic ventricular tachycardia (MVT) may appear, more frequently when induced by drugs. Sustained MVT may be induced by the administration of ajmaline, because the drug increases transmural heterogeneity of repolarization. An automatic mechanism mediated by the beta-receptor seems to have an important role in sustained MVTs that originate in the right ventricular outflow tract (RVOT). The place of origin of the event could be very near the lesion that causes ST elevation [1]. In the cases where MVT is inducible by drugs, triggered activity due to delayed afterdepolarizations in the RVOT may be the electrophysiological background mechanism [2]. Our patient has multiparametric risk markers for sudden cardiac death: spontaneous type 1 Brugada pattern [3]; repetitive syncopal episodes at rest or during sleep; prolonged P wave duration; first-degree atrioventricular (AV) block (HV interval prolongation); prolonged QRS duration in II and V2; the aVR sign [4]; wide and/or deep S-wave in lead I [5]; Northwest QRS axis [6]; late potentials on high resolution ECG; and sinus pauses >2 sec on Holter monitoring signaling SND [7]; and positive programmed electrical stimulation, but this remains controversial, particularly with three extra stimuli.

In the present case, the VCG was crucial to rule out an erroneous diagnosis of LAFB. Different from this dromotropic disturbance, in the frontal plane, the initial 20 msec of the QRS loop is directed downward and leftward instead of downward and rightward, the QRS rotation has the figure-in-eight (versus counterclockwise) shape, and the QRS loop is predominantly located in the right superior quadrant and not in the left superior quadrant. Finally, there is middle-final conduction delay (dashes very close one to another) in the RVOT territory.

Whether there is an association of MVT with the BrS in our case is a matter of discussion. Some questions remain: is the MVT similar to that of polymorphic VT, is this case an overlap phenotype of sinus node dysfunction associated with BrS, and is this a true BrS with misclassification bias or is it a mere epiphenomenon? It is evident from our case that there is a need for further mechanistic characterization and careful clinical genotype-phenotype relationship in this type of patients. Frigo et al showed a homozygous *SCN5A* mutation in BrS with MVT and structural heart disease [8]. Rodríguez-Mañero et al have reviewed implantable cardioverter defibrillator (ICD) data from 15 institutions between 1993 and 2014 and analyzed a cohort diagnosed with BrS. A main finding of their study was that out of 834 patients, 35 had documented MVT, which is more commonly observed in patients with conduction system disease as evidenced by a wide QRS complex like in the present case. The authors concluded that BrS should be considered in patients presenting with RVOT-MVT and that ICD programming should reflect the presence of MVT in these patients [9].

In our patient, a decision was made to recommend immediate implantation of an ICD according to the 2017 Consensus about the management of patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death [10]. After implantation of an ICD, the patient remained asymptomatic but the time is too short for further conclusions.

CONCLUSION

We present a case of a young, symptomatic woman with a spontaneous type 1 Brugada ECG pattern and multi-parametric risk factors for sudden death, resulting in immediate implantation of an ICD based on the recommendations by a consensus statement.

The VCG proved to be an essential resource to rule out an erroneous diagnosis of LAFB; the method showed a significant middle-final dromotropic disturbance in the RVOT region (block in the “hypothetical” subpulmonary division of the right bundle branch).

CONFLICTS OF INTEREST

None declared.

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DISCLOSURES

None.

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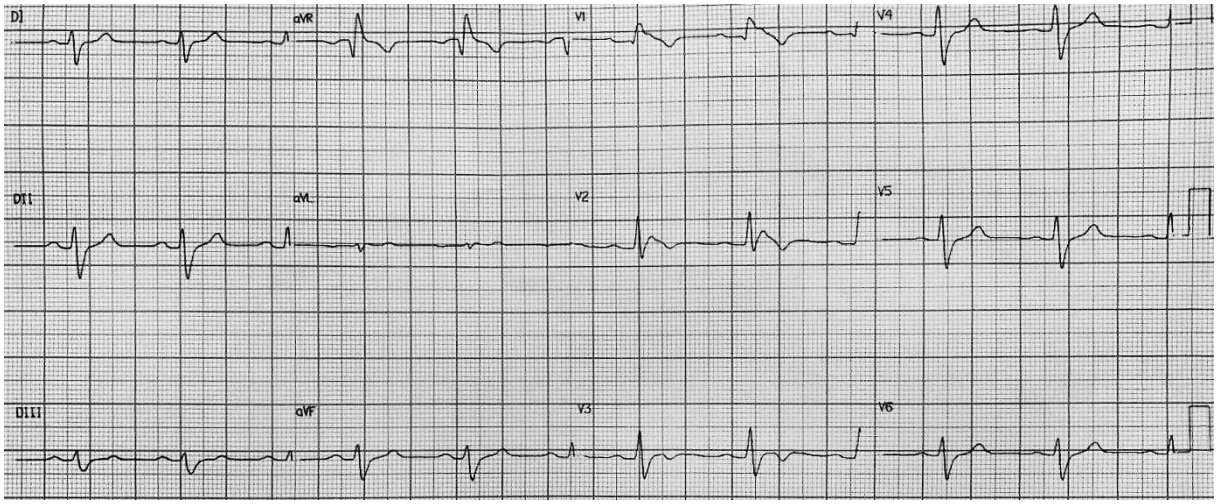
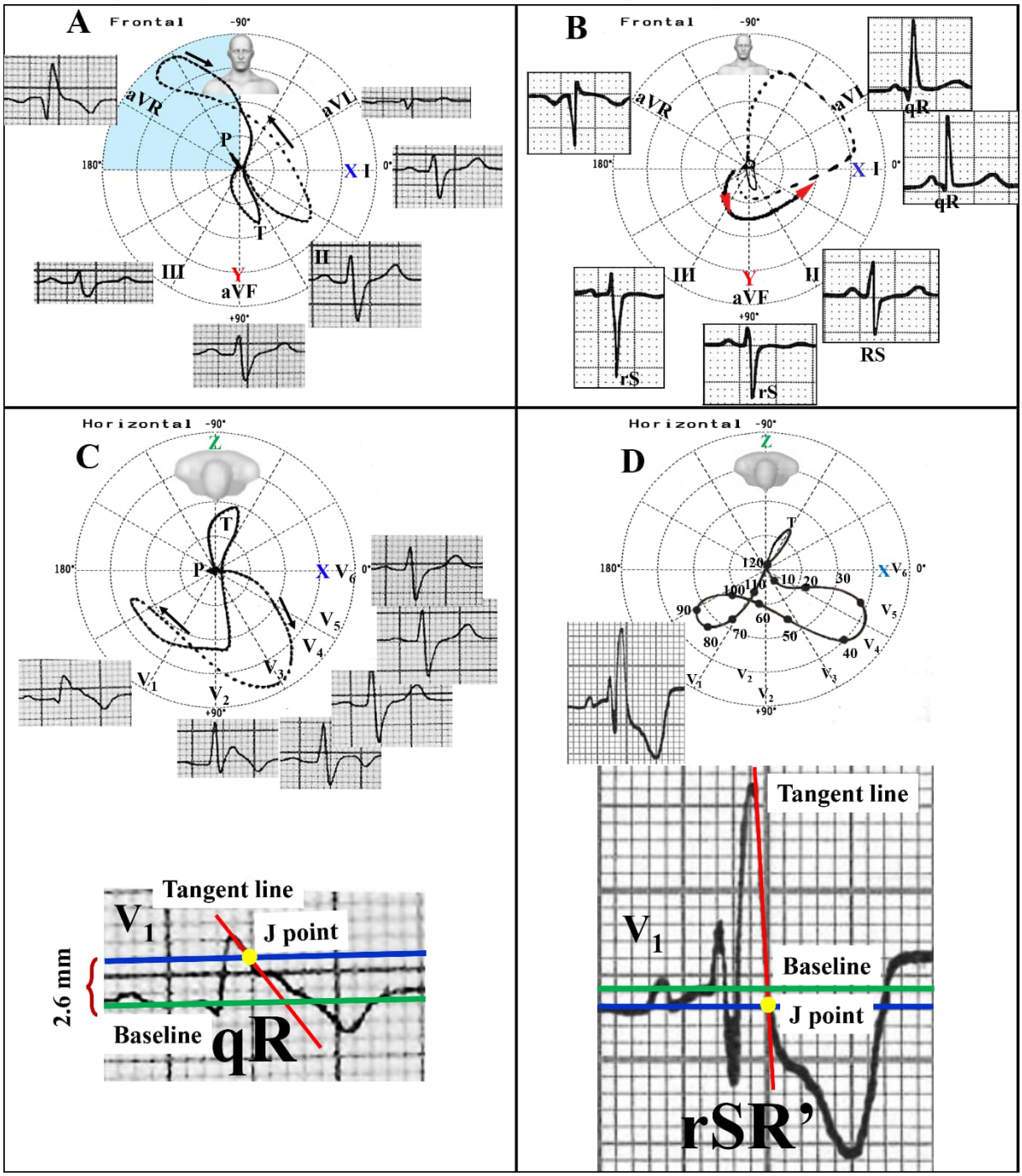
Figure legends

Figure 1. ECG



Sinus rhythm, heart rate 65 bpm, P-wave duration 135 ms, P-wave axis (SÂP) + 60°, PR interval 235 ms, QRS axis -150°: "Northwest QRS axis", prolonged R-Wave Peak Time and QRS duration (≥120 ms), type 1 Brugada pattern, normal QT/QTc intervals. Conclusion: spontaneous type 1 Brugada pattern, aVR+ sign: R wave ≥3 mm and R/q ≥0.75 in lead aVR, deep S wave in I and Northwest QRS axis. These three signals are indicative of very slow conduction within the right ventricular outflow tract (RVOT). Extreme right QRS axis deviation (QRS axis located in the superior right quadrant).

Figure 2. ECG/VCG comparison between the present case (A) and another patient with a typical LAFB (B) in the frontal plane, and the present case (C) and the true RBBB in another patient (D) in the horizontal plane

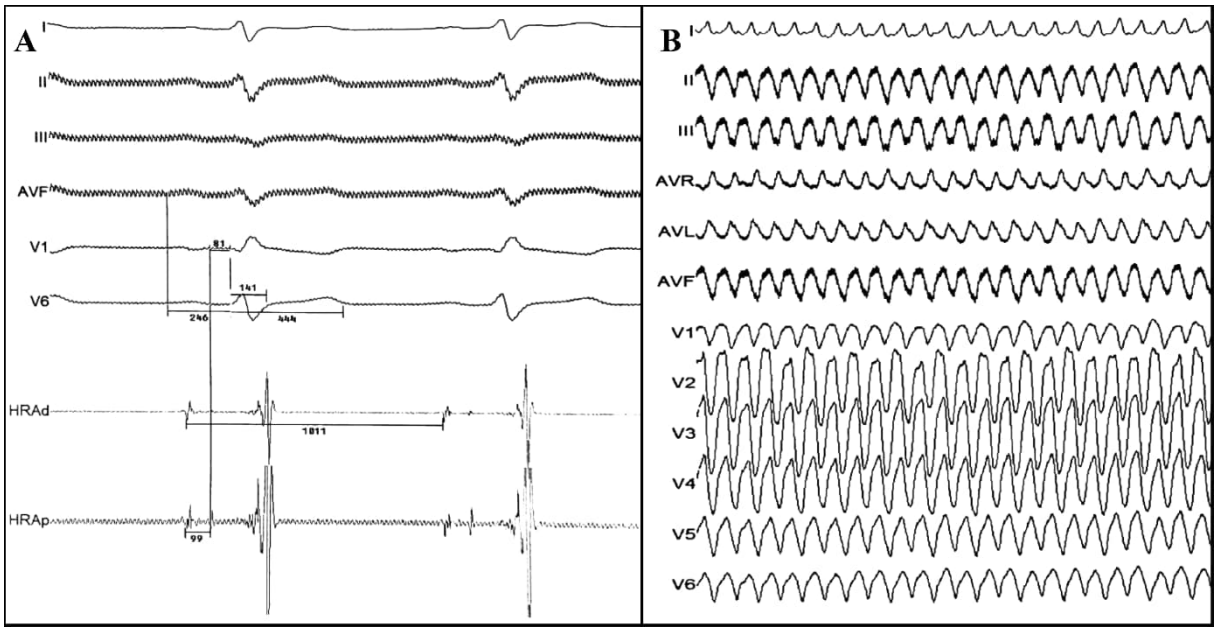


In the frontal plane (A) initial vectors are directed to the left and downward in the present case. On the other hand, in typical LAFB, initial vectors are directed downward and rightward (B); the QRS loop rotation is figure in eight (A) vs. counterclockwise (B); QRS axis is located in the top right quadrant (light blue area) (A) vs. top left quadrant (B); significant mid-final conduction delay corresponding to the right ventricular outflow tract (RVOT) (A) vs. minimal

end-conduction delay (B); SII>SIII (A) vs. SIII>SII (B); qR (A) vs. Qr in aVR (B); rS (A) vs. qR in I and aVL (B).

In the horizontal plane, the VCGs have different patterns: the J point is located at 2.6 mm above the baseline (C), while in typical CRBBB (D) this point is 1 mm below the baseline; ST segment depression followed by asymmetrical T-wave inversion ("appropriate discordance"), characteristic of secondary repolarization disturbance pattern observed in CRBBB (D). Additionally, the QRS morphology is qR (C) and rSR' in CRBBB (D). These differences suggest that the Brugada pattern is "hidden" behind the RBBB (C).

Figure 3. Electrophysiological study



(A) Sinus rhythm; heart rate 58 bpm; PR interval 240 ms. Intracavitary: sinus rhythm, with conduction to the ventricles by the normal conduction system with normal 1:1 AV ratio, HV interval = 88 ms (infra-Hisian block). (B) Premature ventricular stimulation at the tip of the right ventricle with a 500 ms basic cycle (S1S1); S1S2 300 ms; S2S3 240 ms and S3S4 220 ms with induction of sustained monomorphic tachycardia with superior axis (negative QRS

complexes in II, III and aVF, and positive in aVR and aVL, signaling inferior focus) causing early hemodynamic instability.