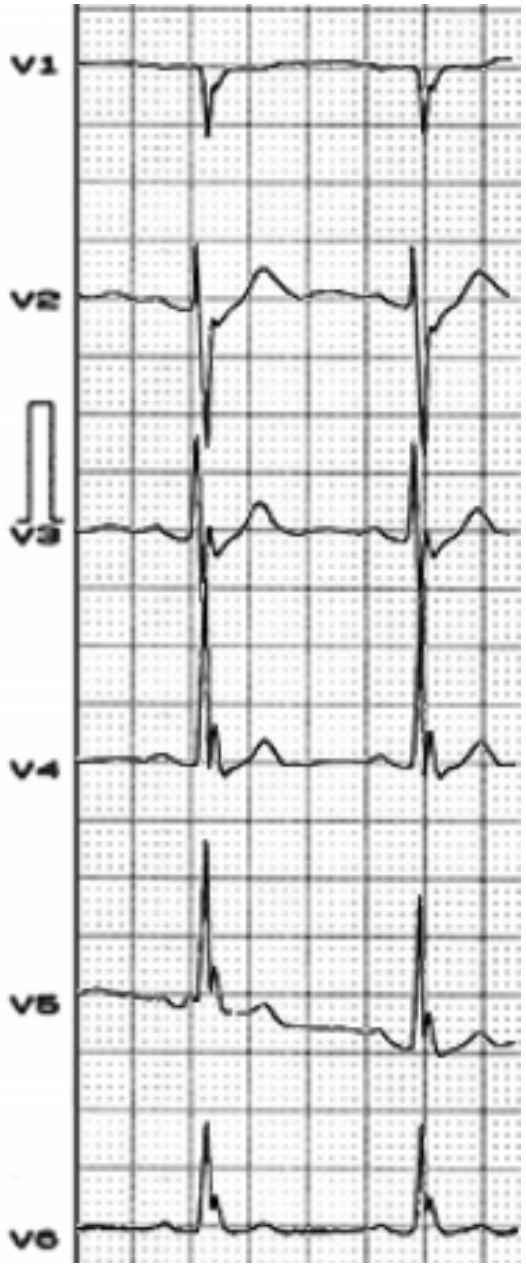
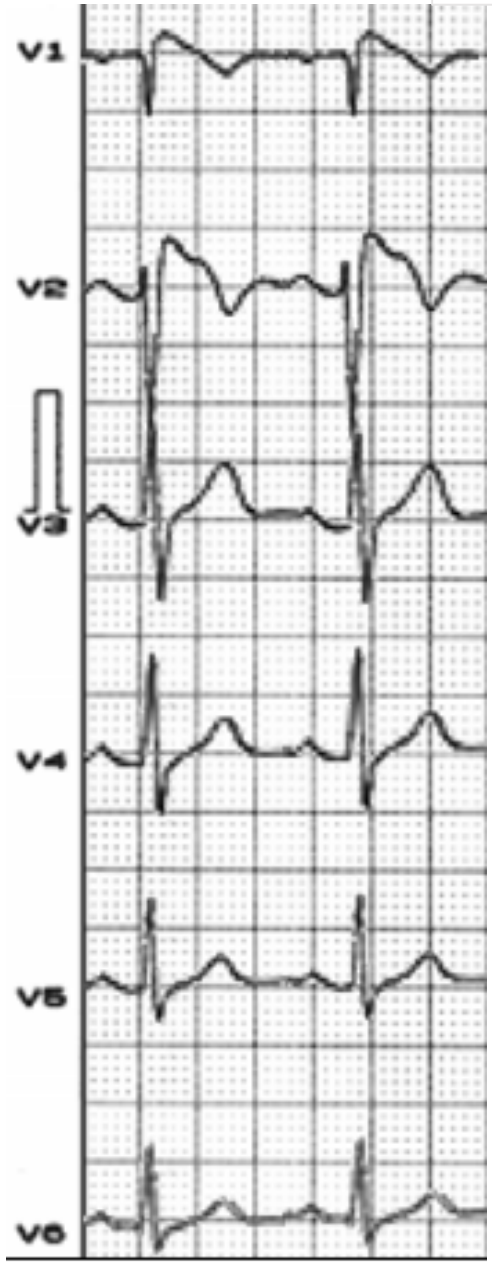
Type 1 Brugada ECG pattern versus truly Complete RBBB

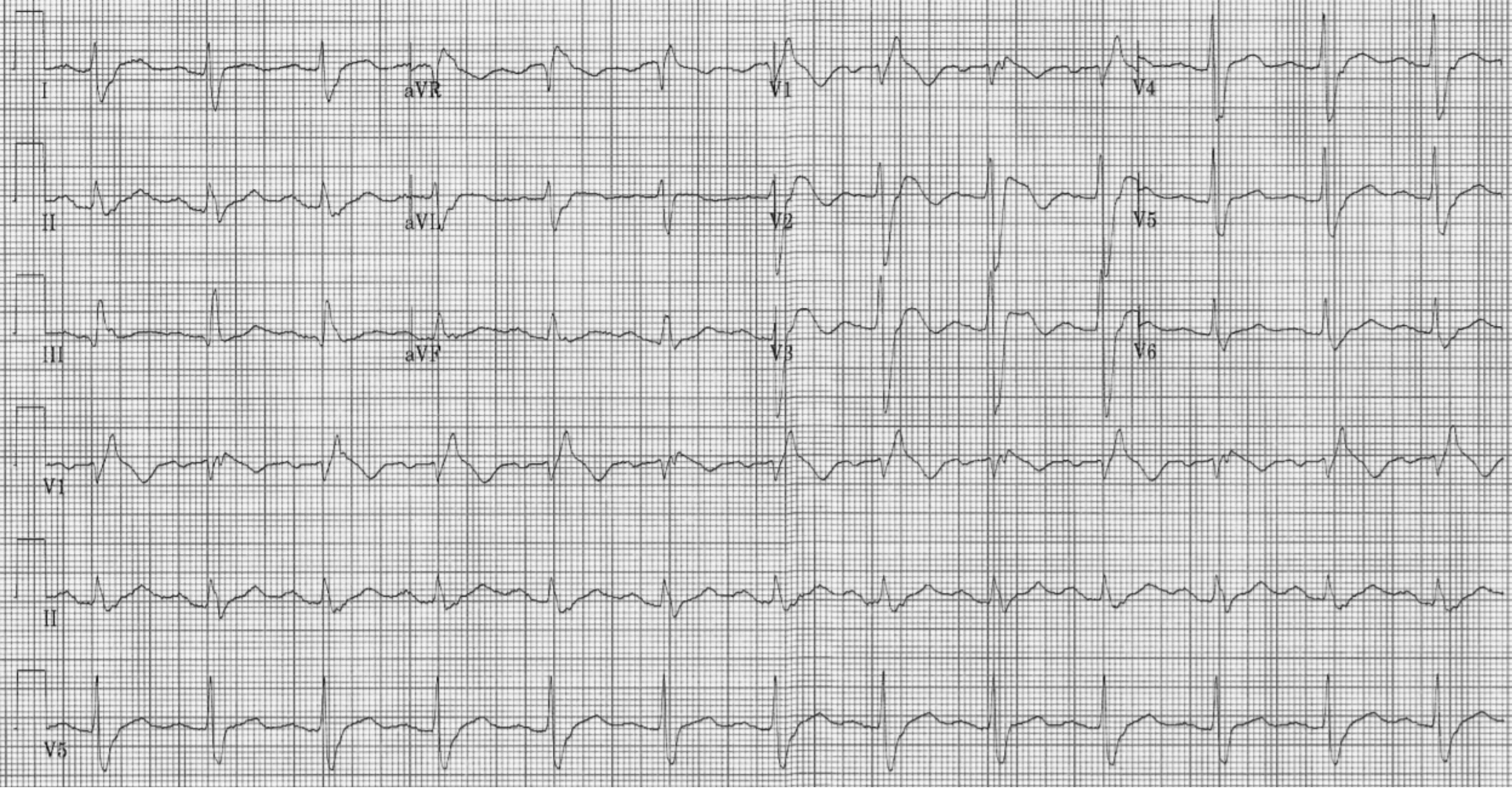
Before ajmaline

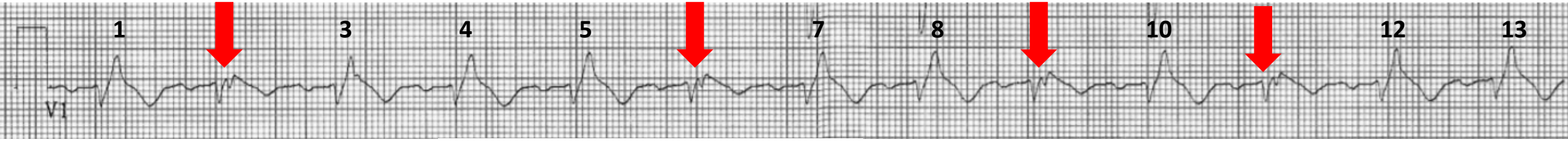


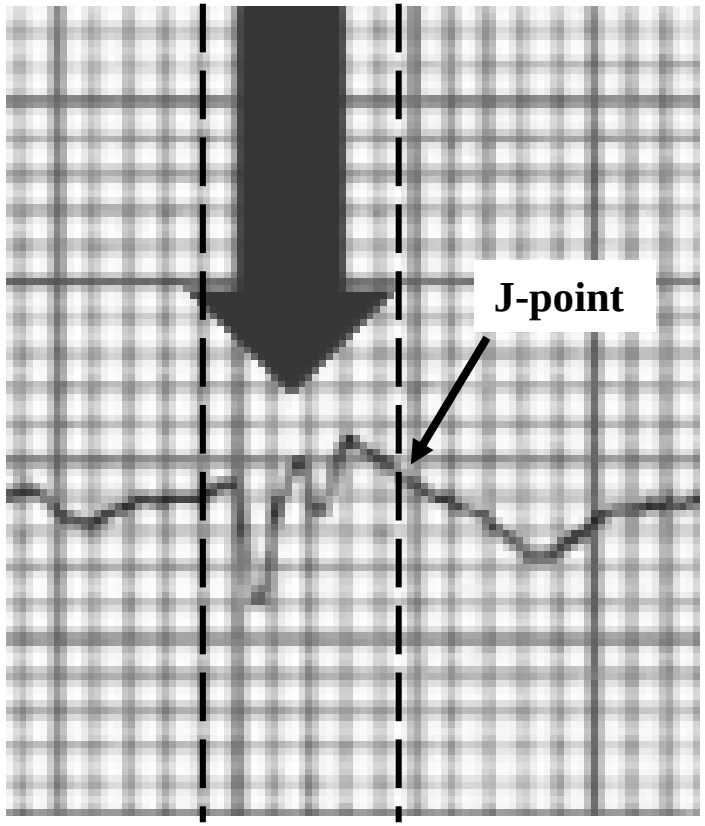
After ajmaline

Positive test




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

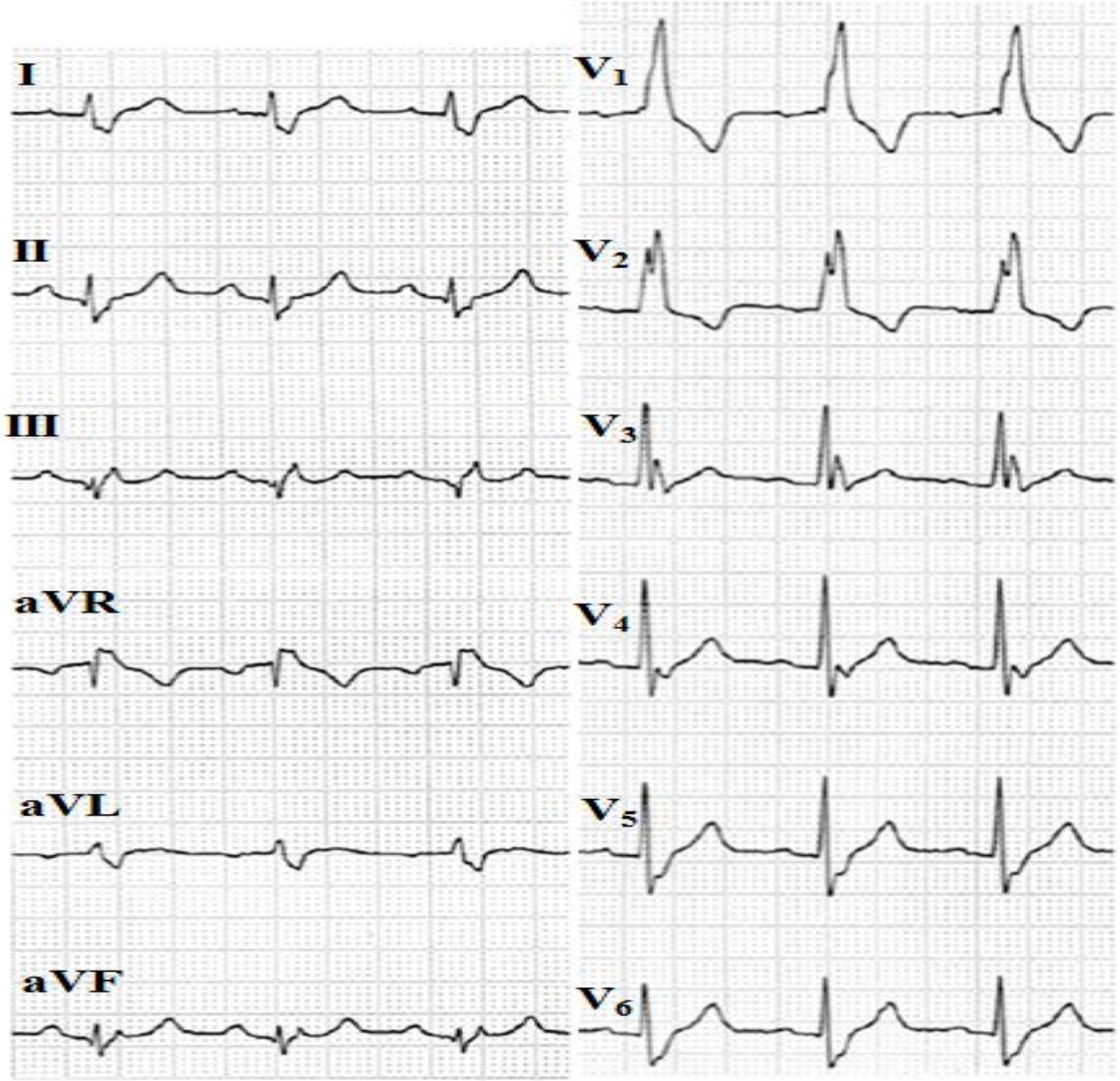




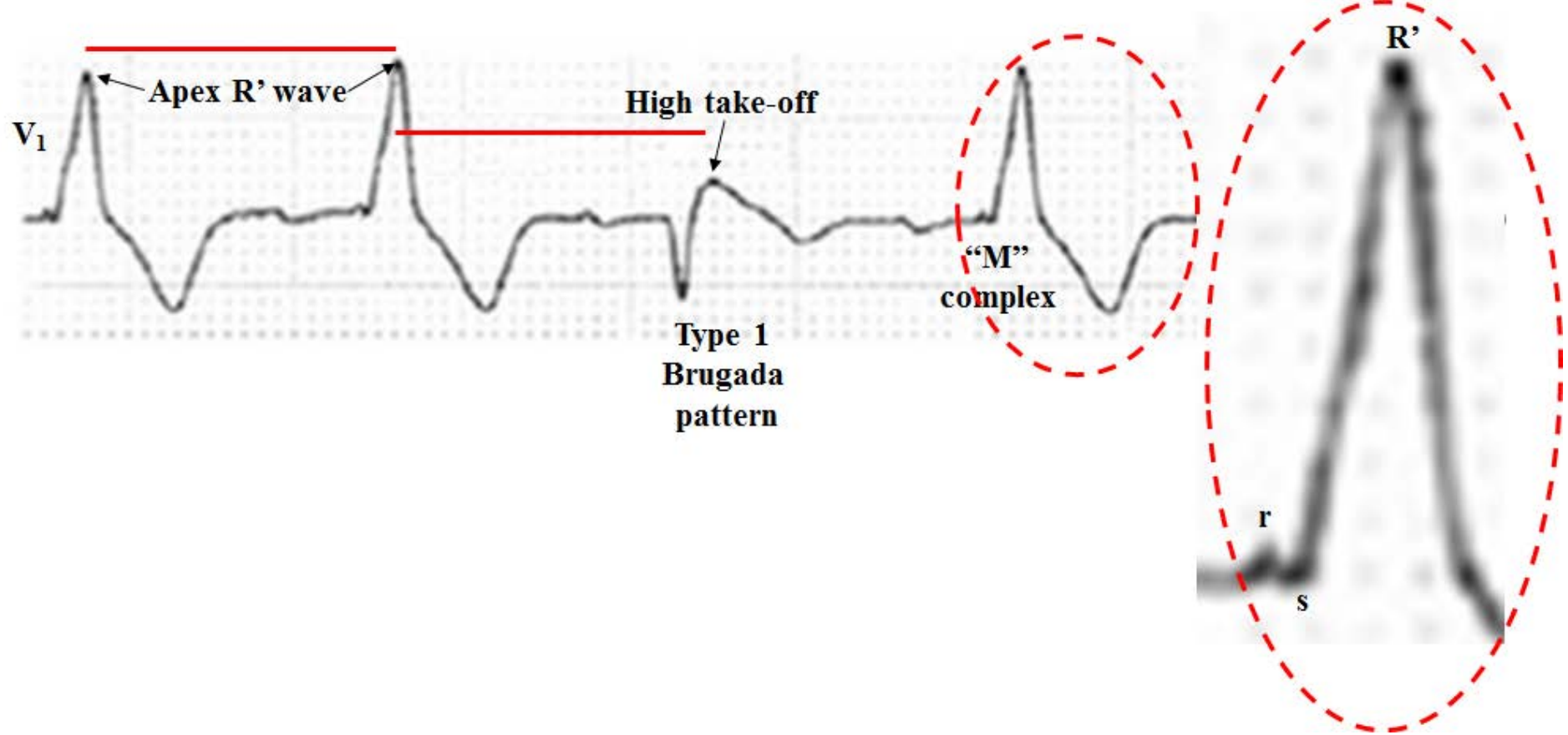
Beats 1, 3, 4, 5, 7, 8, 10, 12, and 13 are beats with high-degree right bundle branch block obscuring the diagnosis of Brugada electrocardiographic pattern.

Chiale PA, Garro HA, Fernández PA, Elizari MV. High-degree right bundle branch block obscuring the diagnosis of Brugada electrocardiographic pattern.Heart Rhythm. 2012 Jun;9(6):974-6. doi: 10.1016/j.hrthm.2012.01.028

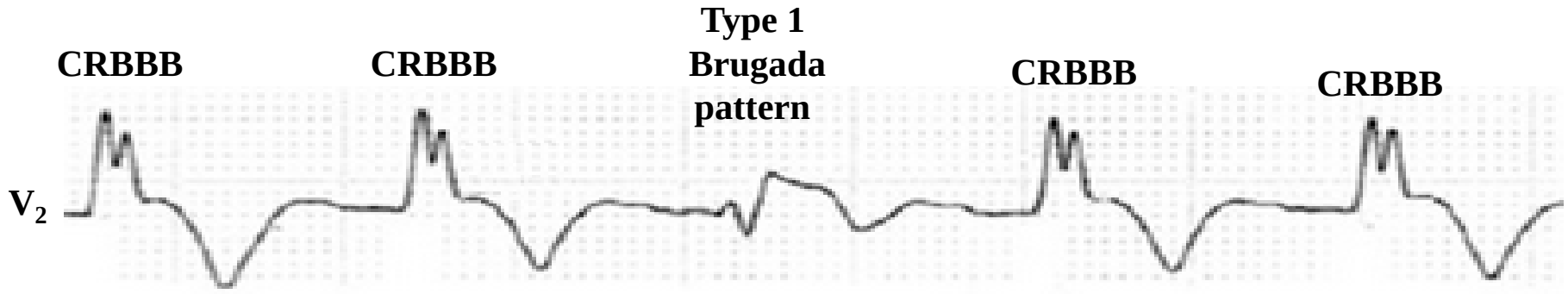
PR interval 200ms, very broad QRS duration(QRSd= 210ms), transient high degree or complete CRBBB, spikes are observed at the end of the QRS in beats without CRBBB upstroke of the S wave in leads V_1: fragmented QRS (fQRS) appears to be a marker for the substrate for spontaneous VF in BrS and predicts patients at high risk of syncope. Spikes are conduction abnormalities within the QRS complex (**Morita 2008**).



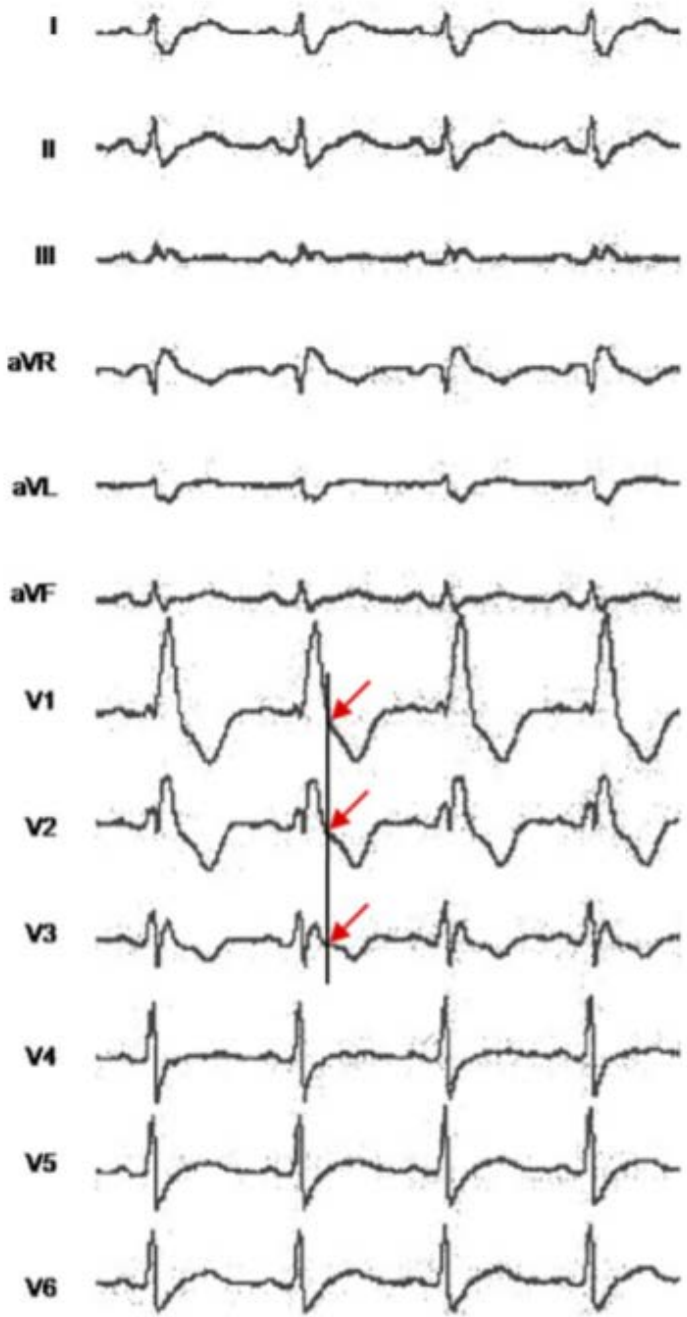
The 12-lead ECG showed CRBBB pattern: QRSD 140 ms, late R in V_1, final broad R wave in aVR, and wide terminal S in left leads. The QRS duration = 140 ms.



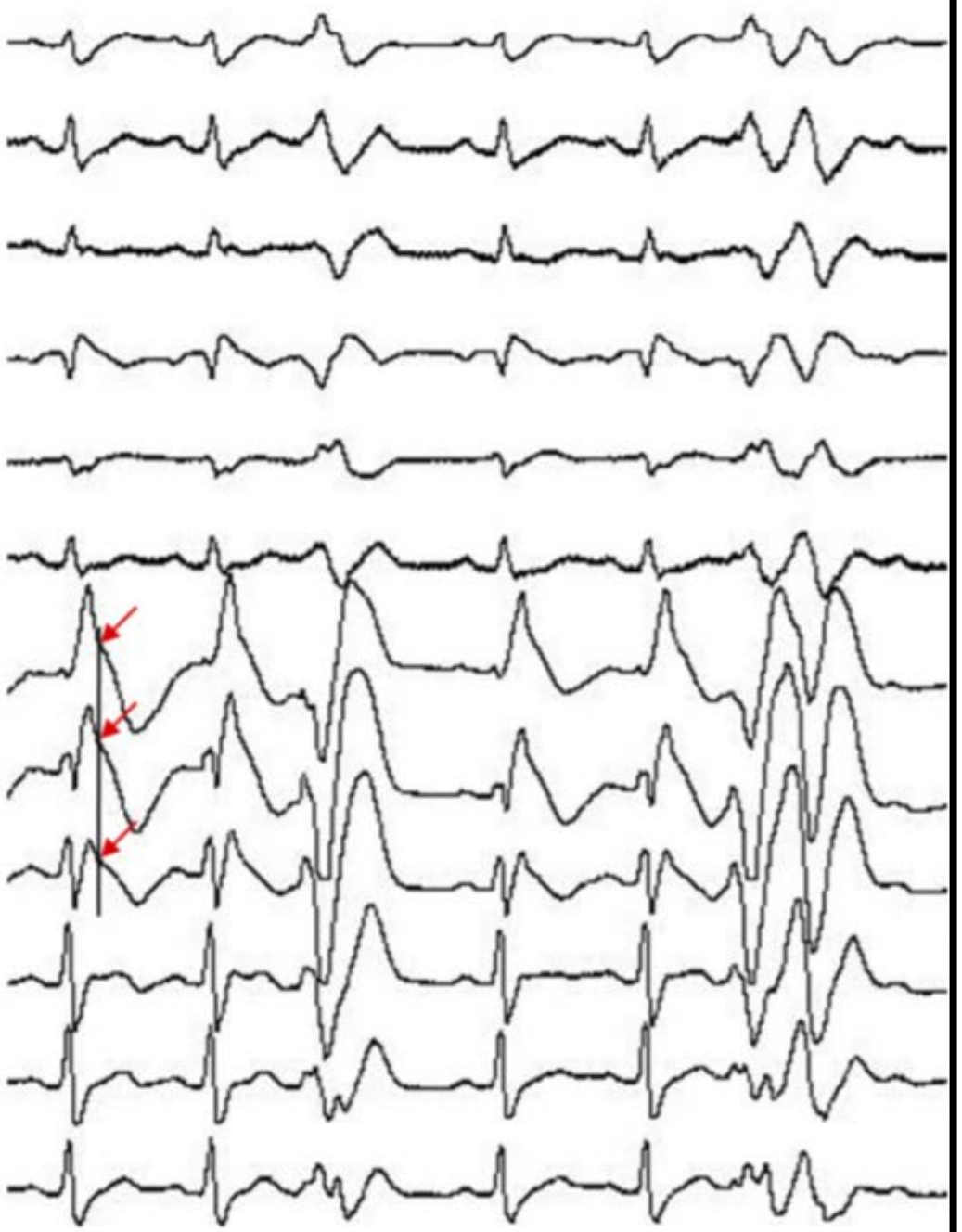
The first and second beats show CRBBB. The third beat without CRBBB (spontaneous transient or intermittent RBBB) shows type 1 Brugada pattern a loss of CRBBB and the normalized QRS complex. Spontaneous resolution of the CRBBB unmasks the type 1 Brugada pattern.



1) ECG preformed at baseline



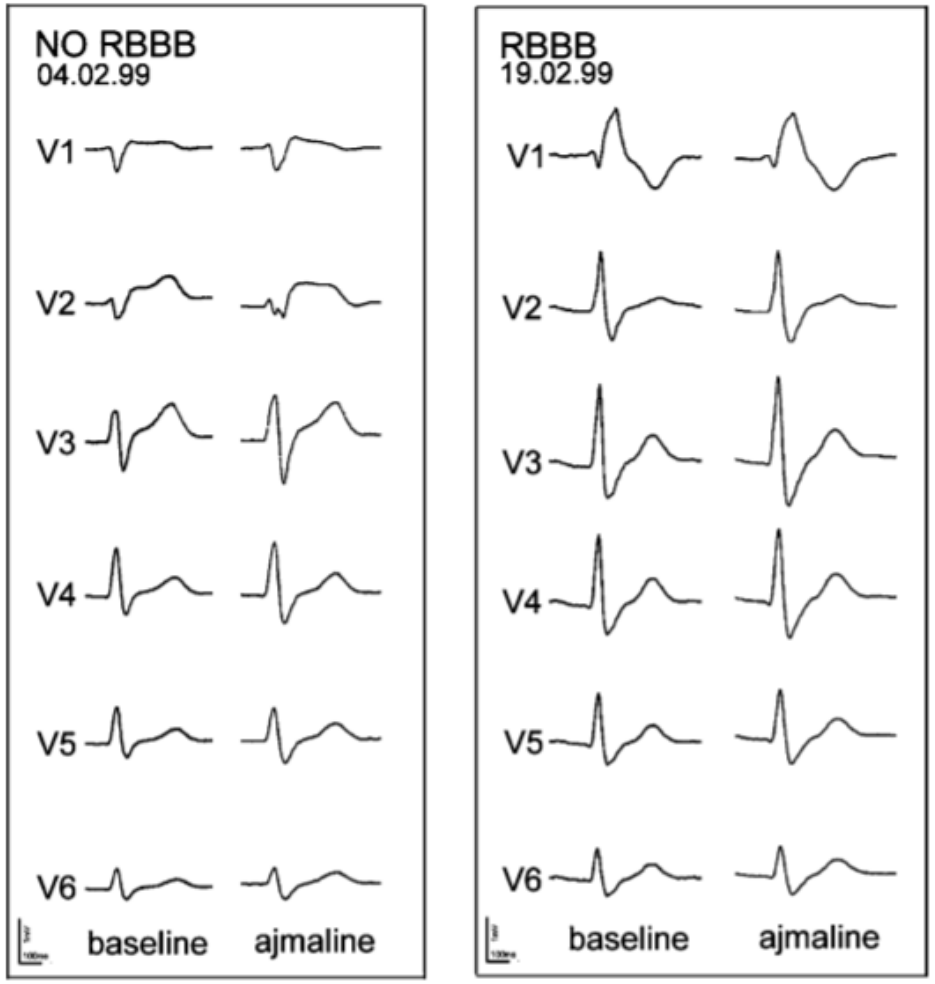
2) ECG at the peak effect of IV ajmaline (50 mg in 50'').



1) ECGs recorded in July 2011 after 3 syncopal episodes at baseline and
2) ECG preformed at the peak effect of intravenous ajmaline (50 mg in 50 seconds).

The ST segment was depressed in leads V1 and V2 as a secondary change to a higher degree of RBBB (QRS duration 170 ms). A clear-cut ST-segment elevation was induced by ajmaline (0.3 mV in lead V1 and 0.45 mV in lead V2), together with PVCs showing a bizarre "left bundle branch block" morphology. Arrows signalize the J point. The development of a higher degree of RBBB was accompanied by the depression of the ST segment mainly in leads V1 and V2 and a complete disappearance of the distinctive ECG type 1 Brugada pattern of clearly unmasked by the effect of IV ajmaline.(**Veltmann C, Wolpert C, Sacher F, et al. Response to intravenous ajmaline: a retrospective analysis of 677 ajmaline challenges. Europace 2009;11:1345– 1352**)

A long time ago, Rolf Haverkamp, and Eckardt presented a patient with a family history of sudden cardiac death and a structurally normal heart with a resting ECG intermittently displaying a type 2 Brugada ECG pattern "saddle-type" Brugada-ECG, which could be reproducibly converted to a coved-type ECG pattern suggestive of Brugada syndrome. However, no ST-segment changes occurred in the presence of a true right bundle branch block (RBBB) in the same patient. In consideration of the inalienable diagnostic criterion of dynamic ECG abnormalities in the right precordial leads in Brugada syndrome, setting the diagnosis in patients with true RBBB may be unattainable.

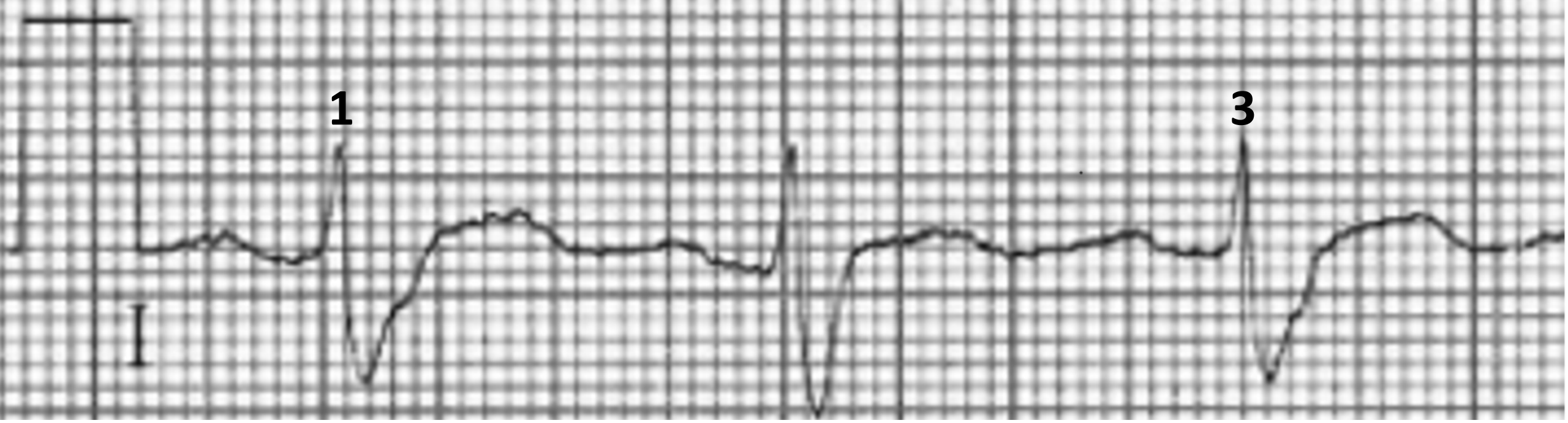


Surface ECG only precordial leads at baseline and after 80mg IV ajmaline from the same patient at two different moments.

No RBBB box baseline ECG 04-02-99; with J point elevation and saddleback-type ST-segment elevation < 0.2mV in the right precordial leads. After IV ajmaline Type 1 Brugada pattern is observed.

Box with RBBB(19-02-99) Baseline ECG showing complete RBBB; note absent dynamic ST segment elevation after application of ajmaline. In the present case the Type 1 Brugada pattern was masked by true RBBB in the second test. Normally, during RBBB it is difficult to asses pathology of the right or left ventricular anterior wall In the presence of RBBB the right precordial ST segment and T wave may be substantially more influenced by delayed an altered intramyocardial conduction of the entire right ventricular myocardium than by local transmural voltage gradients due to the presence of an accentuated notch in right ventricular epicardium, which is the presumed mechanism of precordial ST segment changes in BrS, Due to the fact, that the dynamic ECG abnormalities in the right precordial leads are of utmost importance, it may be impossible to role out Brugada syndrome in patients witn true RBBB, especially when genetic testing remain inconclusive.

1. **Rolf S, Haverkamp W, Eckardt L. True right bundle branch block masking the typical ECG in Brugada syndrome. Pacing Clin Electrophysiol. 2005 Mar;28(3):258-9 PMID: 15733192 DOI: 10.1111/j.1540-8159.2005.09476.x**



Additionally, beats 1 and 3 have broader S wave related beat 2 consequence of higher degree of RBBB.

The presence of a wide and/or large S-wave in lead I was a powerful predictor of life-threatening ventricular arrhythmias in patients with BrS and no history of cardiac arrest at presentation. However, the prognostic value of a significant S-wave in lead I should be confirmed by larger studies and by an independent confirmation cohort of healthy subjects. This is the first time; that it is known to us that the Calò´s signal is observed intermittently or transiently

Calò L, Giustetto C, Martino A, Sciarra L, Cerrato N, Marziali M, Rauzino J, Carlino G, de Ruvo E, Guerra F, Rebecchi M, Lanzillo C, Anselmino M, Castro A, Turreni F, Penco M, Volpe M, Capucci A, Gaita F.A New Electrocardiographic Marker of Sudden Death in Brugada Syndrome: The S-Wave in Lead I. J Am Coll Cardiol. 2016 Mar 29;67(12):1427-1440. doi: 10.1016/j.jacc.2016.01.024.

Pérez-Riera AR1, Baranchuk A2, Zhang L3, Barbosa-Barros R4, de Abreu LC5, Brugada P6. Myotonic dystrophy and Brugada syndrome: A common pathophysiologic pathway? J Electrocardiol. 2017 Jul - Aug;50(4):513-517. doi: 10.1016/j.jelectrocard.2017.03.008

Type 1 myotonic dystrophy (DM1) is a heterogeneous hereditary multisystem disorder with autosomal dominant transmission traits. It is caused by expansions of microsatellite repeats of cytosine-thymineguanine (CTG) in 3′ untranslated regions of the dystrophic myotonic protein kinase (DMPK) gene. The repeat units may expand to several thousand repeats. DM1 (Steinert's disease) is classic with CTG repeats between 50–2000. The complexity of DM1 phenotype is perhaps related to specific combinations of several possible pathogenic pathways in different tissues of individual patients. The prevalence estimates per 100,000 are 8.26 for DM1 (1). Patients with DM1 have a lower life expectancy, due to cardiac or respiratory complications. DM1 affects smooth, cardiac- and skeletal-muscles (weakness and myotonic phenomena), eyes (cataract), endocrine system (insulin resistance/diabetes, thyroid dysfunction, hypogonadism/infertility), and central nervous system (mental retardation, attention disorders). Cardiac involvement is variable, though mainly affecting intraventricular His-Purkinje system. Patients may present with palpitations, syncope, shortness of breath, and in severe cases, sudden death. Progressive conduction disturbance is a hallmark of DM1 on electrocardiogram (ECG) such as atrioventricular block (AVB), right/left bundle branch blocks (RBBB/LBBB) and fascicular blocks, etc. Conduction disturbance progresses faster in patients with CTG repeats >1000 (2). Positive late potentials (LPs) are a consequence of slowed conduction along the His-Purkinje system. Structural heart abnormalities are common in DM1, such as left ventricular systolic dysfunction and reduced global longitudinal strain consequence of perivascular interstitial fibrosis predominantly in the conduction system, fatty infiltration, hypertrophy, myofibrillar disarray, and focal myocarditis.

Case presentation A 42-year-old man was referred to us by his ophthalmologist for evaluating cardiovascular risks of undergoing cataract surgery. The patient was previously diagnosed with DM1. He had a long history of progressive muscular weakness and myotonic phenomenon predominantly in the distal leg, forearm, wrists, hands and fingers. Additionally, he had dysphagia, constipation, diabetes, and progressive loss of visual acuity. Family history of DM1 is unknown since he was an adopted child. He denied any heartrelated symptoms. Physical examinations revealed bilateral cataracts, droopy eyelids, nasal voice tone, blood pressure 135/80 mmHg, pulse rate 55 bpm. Point of maximum palpation was at 1 cm lateral shift at the left of the midclavicular line, in the 5th left intercostal space. During inspiration, a physiological first (S1) and second sounds (S2) split was audible. S3 heart sound was heard with gallop cadence at the beginning of diastole (middle third). There was a palpable and painful liver, 2 cm from the right costal margin. He did not present jugular venous distension nor swelling in the legs, ankles, or feet. Cardiac work-up included ECG, vectorcardiogram (VCG), signal average ECG (SAECG), echocardiogram, 24-hour Holter monitoring, electrophysiological study (EPS) and genetic testing.

Surface ECG (Figure 1) showed CRBBB and Type-1 Brugada ECG pattern. VCG changes (Figure 2) depicted also left anterior fascicular block (LAFB). EPS revealed a split His (H-H'=48 ms; H'-V=48 ms). VF was induced with only two extra stimuli. Maneuvers to unmask Brugada ECG pattern during CRBBB were performed (known as Chiale's maneuver) (3; 4) during EPS. Using right apical ventricular pacing to achieve appropriately timed match to the native H-V interval, a narrow QRS fusion complex was created (Figure 3, third beat). As such, the Chiale's maneuver eliminated CRBBB on the fusion beat successfully. In the fusion beat, typical ECG pattern of LAFB was fully uncovered. Type-1 Brugada ECG pattern was also shown in both sinus beats with CRBBB and the fusion beat without CRBBB (Figures 1, 3). SAECG revealed positive LPs, though no ventricular tachyarrhythmia were recorded on 24-hour ambulatory Holter. Echocardiogram showed regional wall motion abnormality, mild left atrial and left ventricular hypertrophy, and impairment of the left ventricular systolic and diastolic functions with ejection fraction of 50%. Genetic screening was negative for BrS (20-gene screening) Figure 1

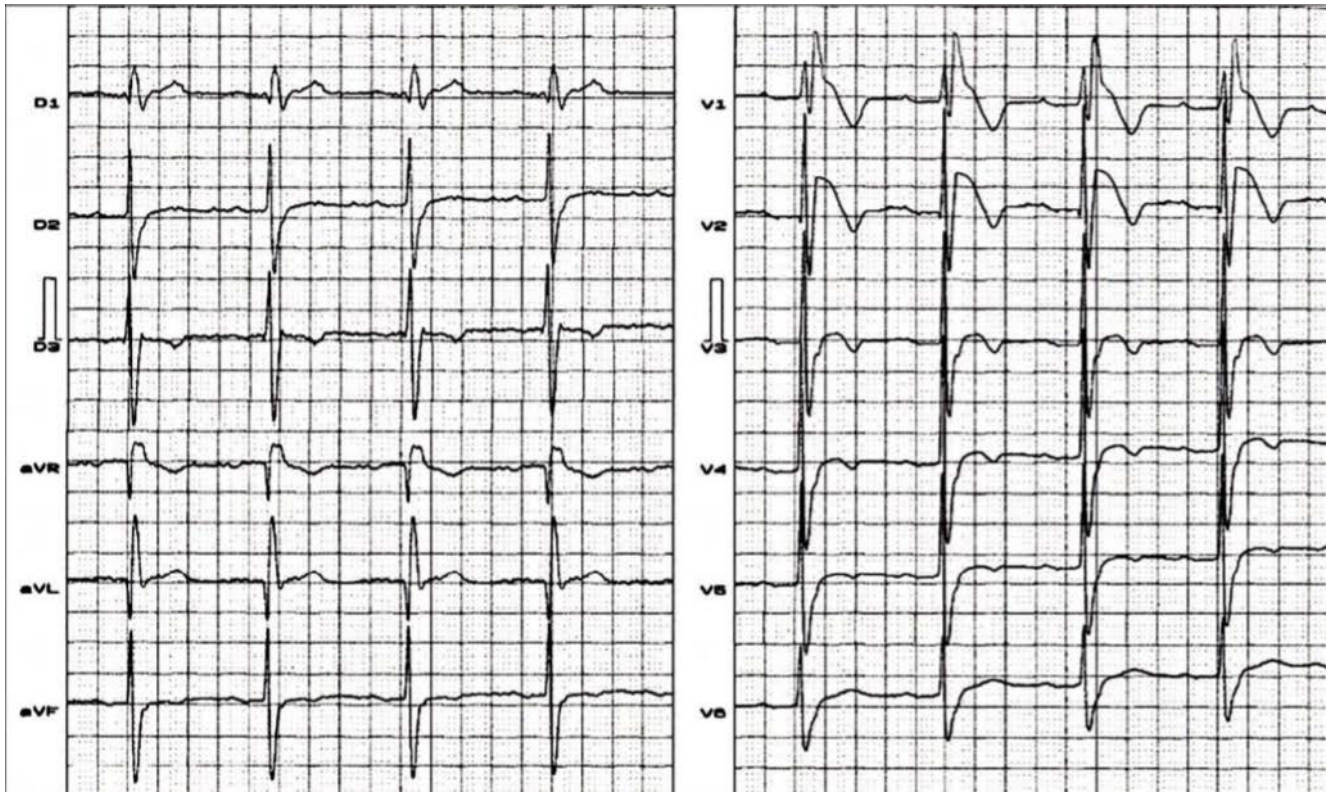


Figure 1 ECG: sinus rhythm, HR: 55 bpm, PR interval: 250 ms, QRS duration: 171 ms, extreme left axis deviation (SÂQRS near –70º), triphasic pattern in V1 (RSR'), broad final R-wave in lead aVR and wide S-wave in left leads V5-V6. J-point and ST-segment elevation >2 mm convex to the top followed by negative Twave in leads V1-V2.

Figure 2 ECG/VCG correlation in the Frontal (A) and Horizontal (B) Planes

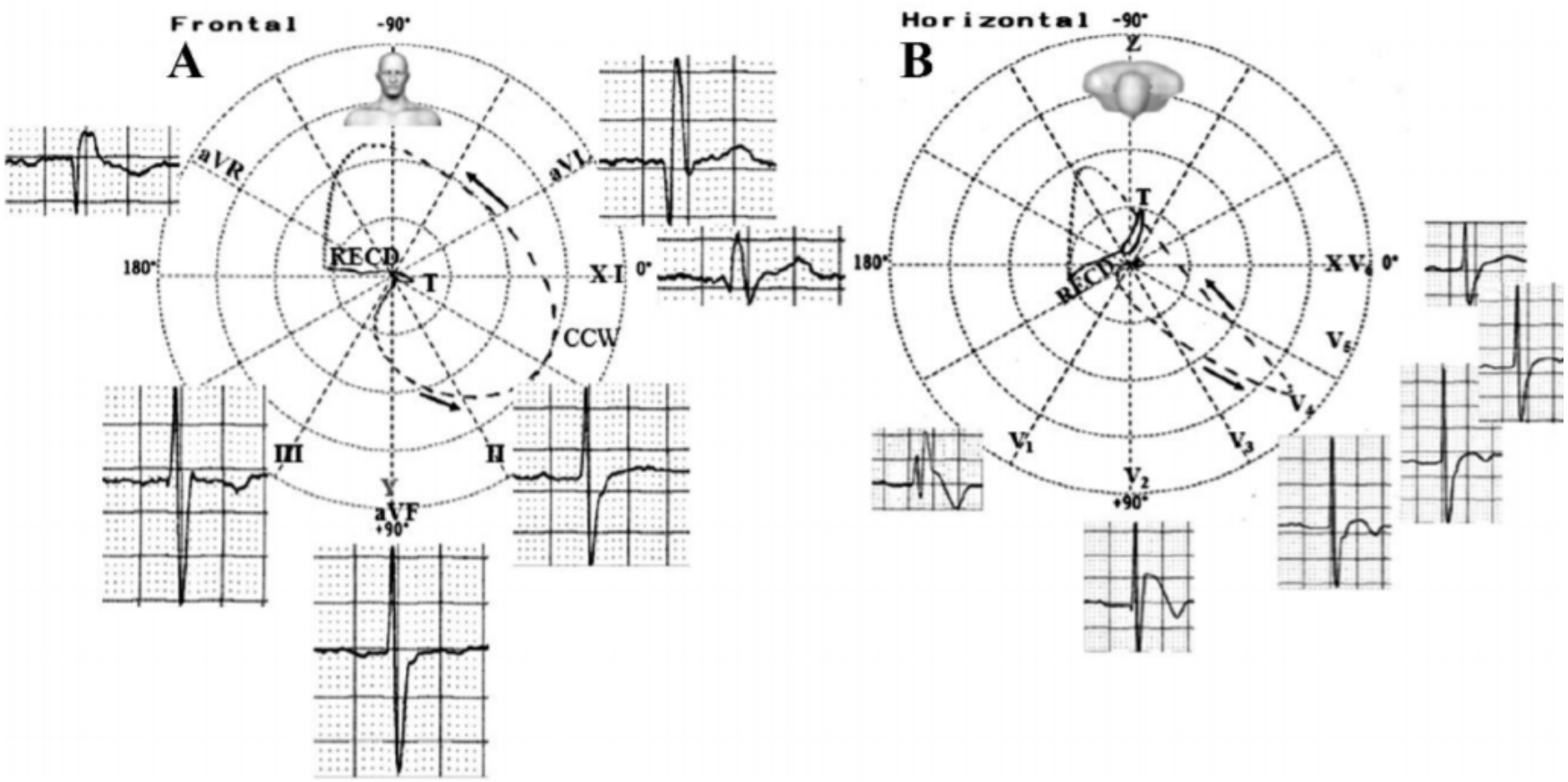
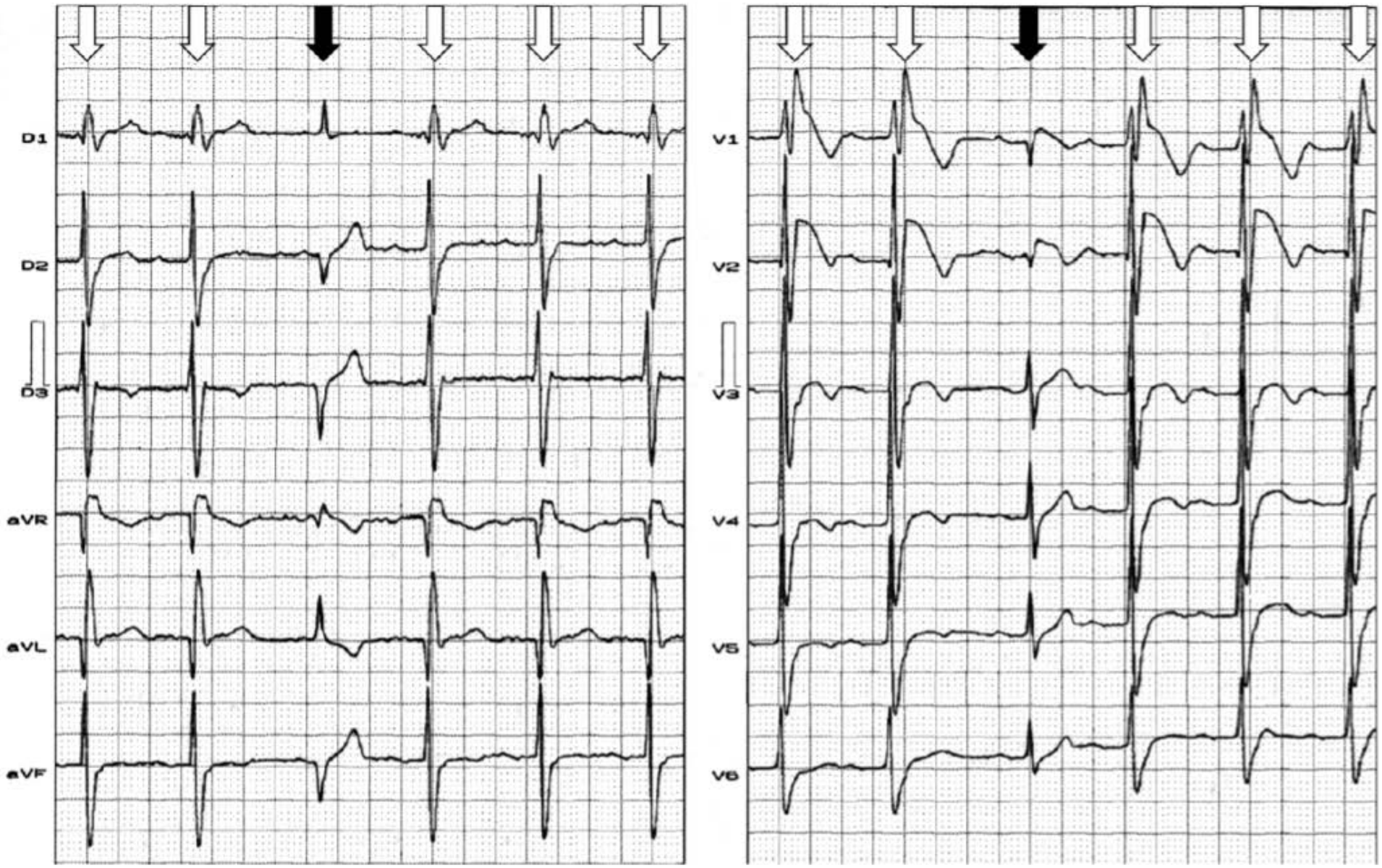


Figure 2

Frontal Plane: RS pattern (R=S) in the inferior leads. The 10 to 20 ms initial vector of the QRS loop goes down and rightward, with counterclockwise rotation (CCW) and final portion of slow inscription located at the right of the orthogonal X lead (0°/±180). Additionally, the QRS loop shape is rounded. The wide final R-wave in lead aVR and the right end conduction delay (RECD) on the orthogonal X lead at the right is a consequence of RBBB.

Horizontal Plane: QRS duration 130 ms; in the right precordial leads triphasic pattern RSR' in lead V1 and broad final S-wave in the left leads: CRBBB. In leads V1-V2, J point and ST-segment elevation ≥ 2 mm followed by negative T-wave: type-1 Brugada ECG pattern. The afferent limb is located behind the orthogonal X lead QRS: RBBB Grishman type. Additionally, afferent limb does not return to the point of origin but continues in a rightward and predominantly anterior direction to inscribe a terminal, "finger-like appendage" in the right anterior quadrant

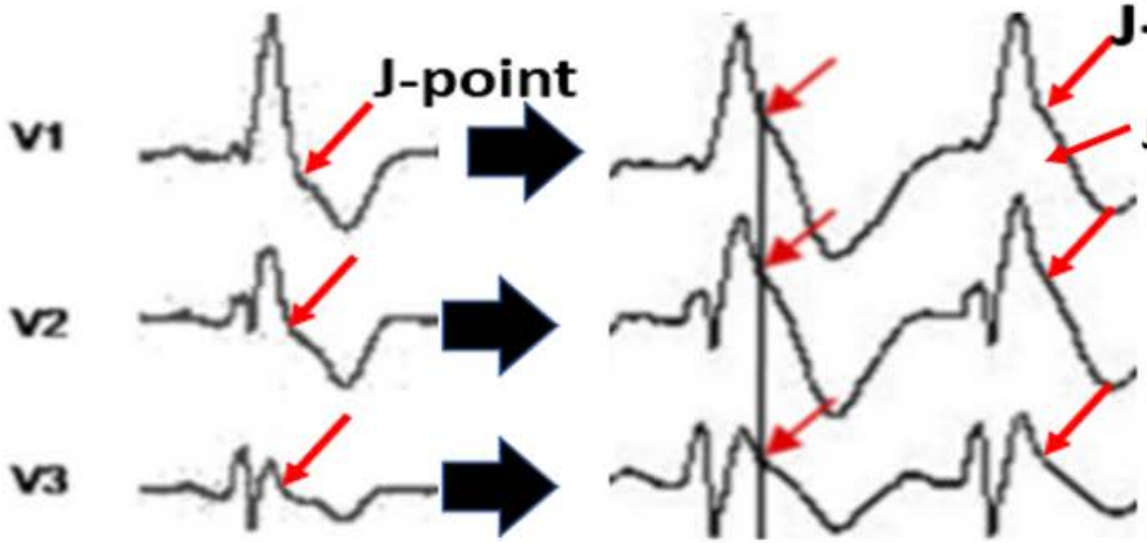


All beats (white arrows) except the 3rd one in each panel show first degree AVB with CRBBB. The 3rd beat in each panel shows a fusion beat with a narrow QRS which is resulted from the “Chiale’s maneuver” by right apical ventricular pacing with appropriately timed A-V intervals. Third beat (black arrows) shows first degree AVB and typical LAFB and type 1 Brugada pattern.

10. Baranchuk A, Barbosa-Barros R, Pérez-Riera AR.Brugada ECG pattern obscured by right bundle branch block: how to resolve the enigma?Pacing Clin Electrophysiol. 2014 Aug;37(8):1071-2. doi: 10.1111/pace.12388
11. Wada T, Nagase S, Morita H, Nakagawa K, Nishii N, Nakamura K, Kohno K, Ito H, Kusano KF, Ohe T. Incidence and Clinical Significance of Brugada Syndrome Masked by Complete Right Bundle-Branch Block. Circ J. 2015;79(12):2568-75.
12. Ozeke O, Cay S, Ozcan F, Topaloglu S, Aras D. Brugada Syndrome with Early Repolarization masked by Complete Right Bundle-Branch Block. Europace. 2015 Sep;17(9):1454-5.
13. Pérez-Riera AR, Baranchuk A, Zhang L, Barbosa-Barros R, de Abreu LC, Brugada P. Myotonic dystrophy and Brugada syndrome: A common pathophysiologic pathway? J Electrocardiol. 2017 Jul - Aug;50(4):513-517. doi: 10.1016/j.jelectrocard.2017.03.008

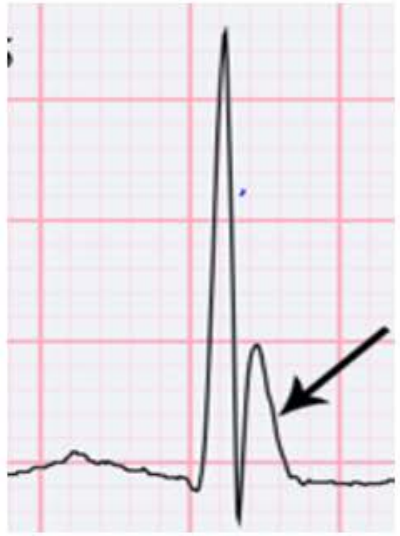
Basal ECG right precordial leads

ECG right precordial leads immediately after IV ajmaline

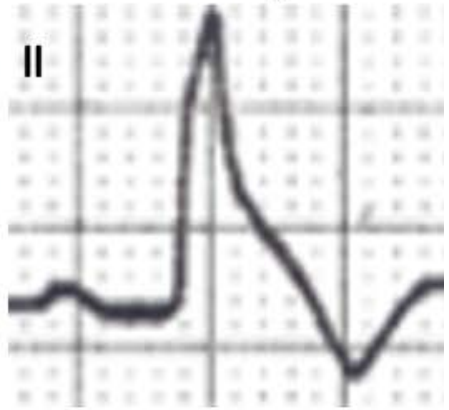


J- wave with "slurring" shape followed by negative symmetric T-wave

J-notched



J-slurring



The Brugada type 1 ECG pattern is hidden by the BCRD: The J point is located further down, followed by negative asymmetric T wave.

Ajmaline unmasks type 1 ECG Brugada pattern: J-point J is located higher up