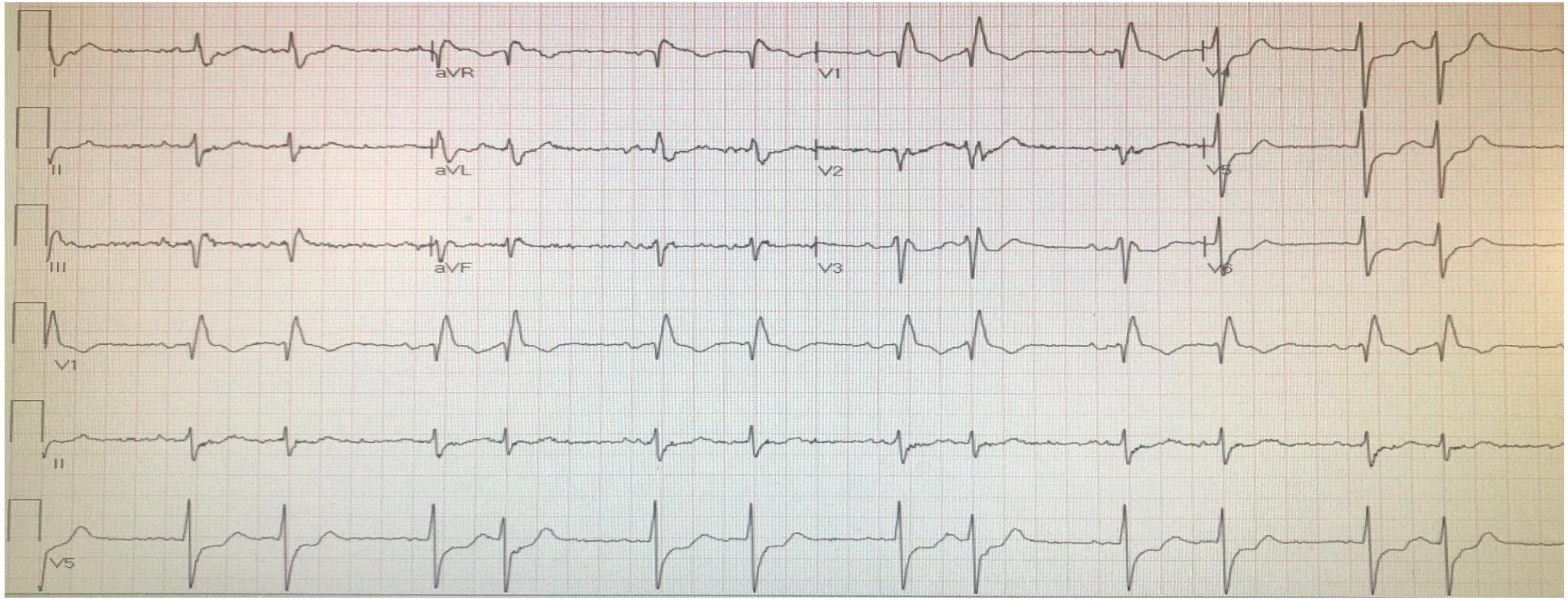
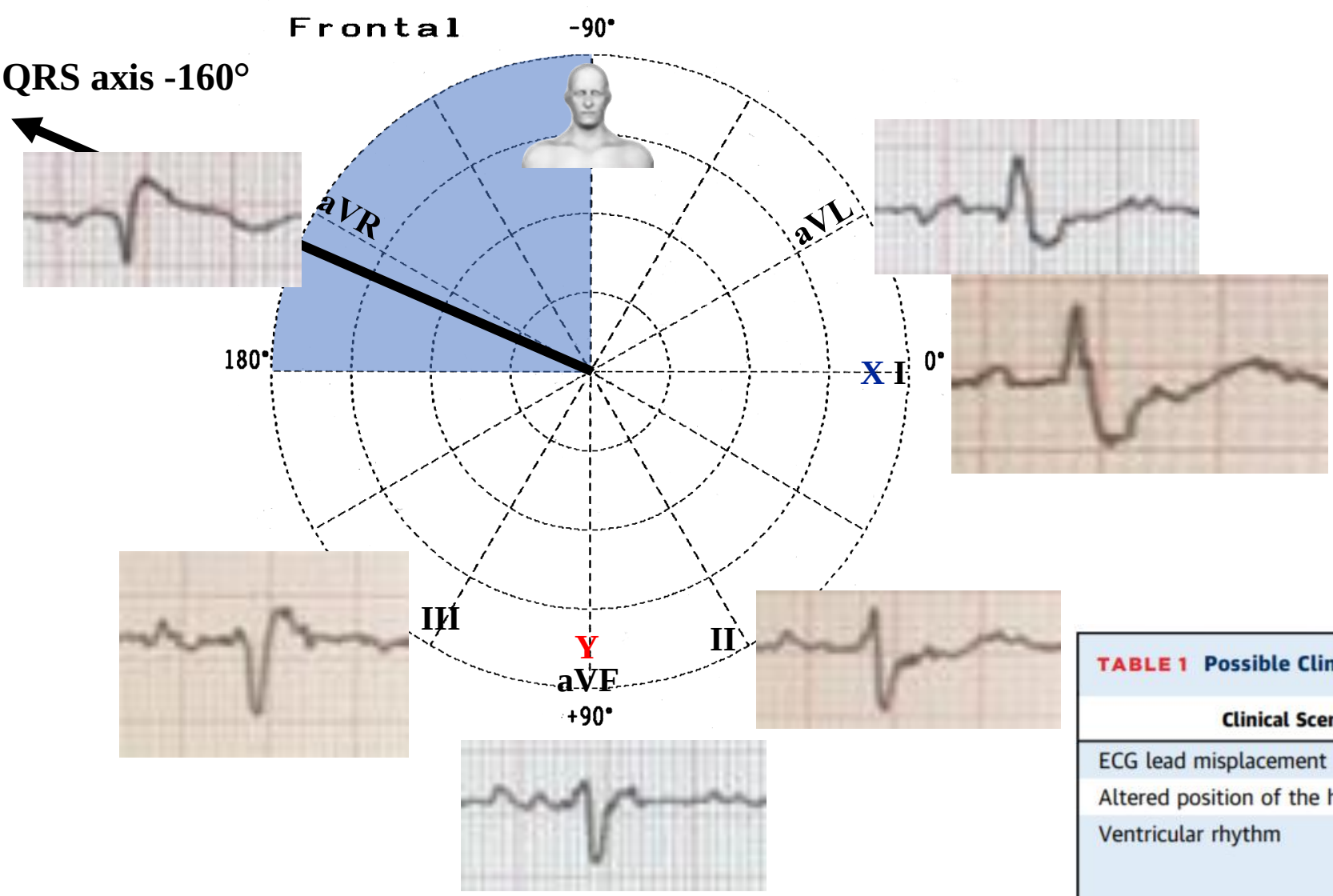


ECG diagnosis: the rhythm and the mischief going on in the AV junction. We believe this is normal sinus rhythm (down arrows) with a very peculiar fixed coupled junctional bigeminy and with alternating antegrade and retrograde conduction. The pattern is repetitive. Obviously I'm guessing as to the location of the premature junctional focus, but I'm assuming it is fixed coupled to the sinus beats - other wise the group beating pattern wouldn't occur.

QRS axis located on top right quadrant "Norwest axis", *No man's land*, **or (i.e., "N-M-L").** (between -90° ±180°) because all the frontal QRS leads are predominantly negative with exception of aVR. Atypical Complete Right Bundle Branch Block (CRBBB). Why atypical? Because the J-point in the right precordial leads(V1) is elevated related baseline. Truly typical CRRBB has the J point depressed related baseline (QRS onset). V1 lead fulfills the criteria of type 1 Brugada ECG pattern: J-point and ST segment elevation≥2nn in at least one right precordial lead. Lead I: R<S wide S wave predominantly negative (**Calò L, et al. 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**.)

Lead II: R<S, Lead III rSR, R wide, aVR: qR, with wide R Similar V1 pattern, aVL: rS, with wide S

aVR: rSr´

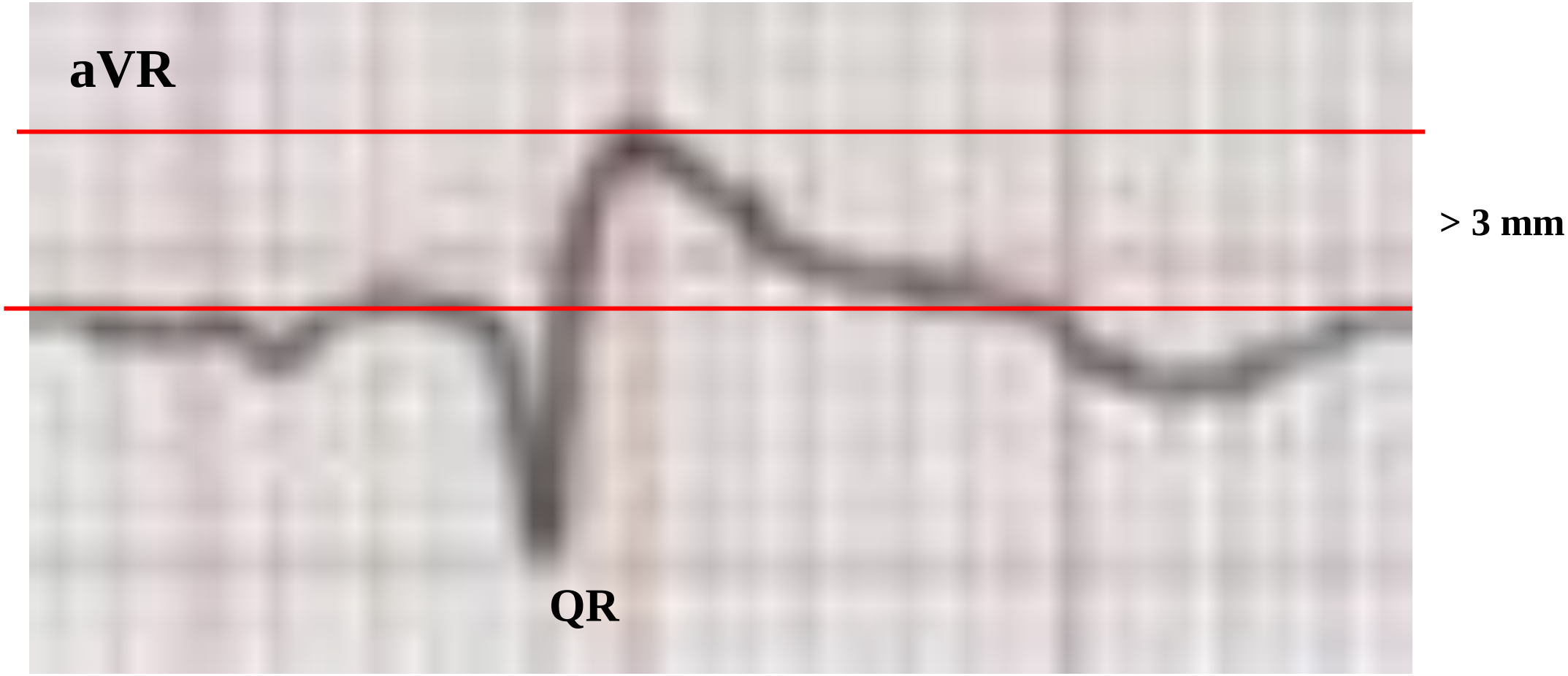


1. Pérez-Riera AR, Ferreira C, Schapachnik E. Value of 12 lead electrocardiogram and derived methodologies in the diagnosis of Brugada disease. In: Antzelevitch C, Brugada P, Brugada J, et al., editors. The Brugada Syndrome From Bench to Bedside. Hoboken, NJ: Blackwell Futura Publishing, 2005:87–110.
2. Pérez-Riera AR, Ferreira Filho C, de Abreu LC et al. Do patients with electrocardiographic Brugada type 1 pattern have associated right bundlebranch block? A comparative vectorcardiographic study. Europace 2012;14:889–97

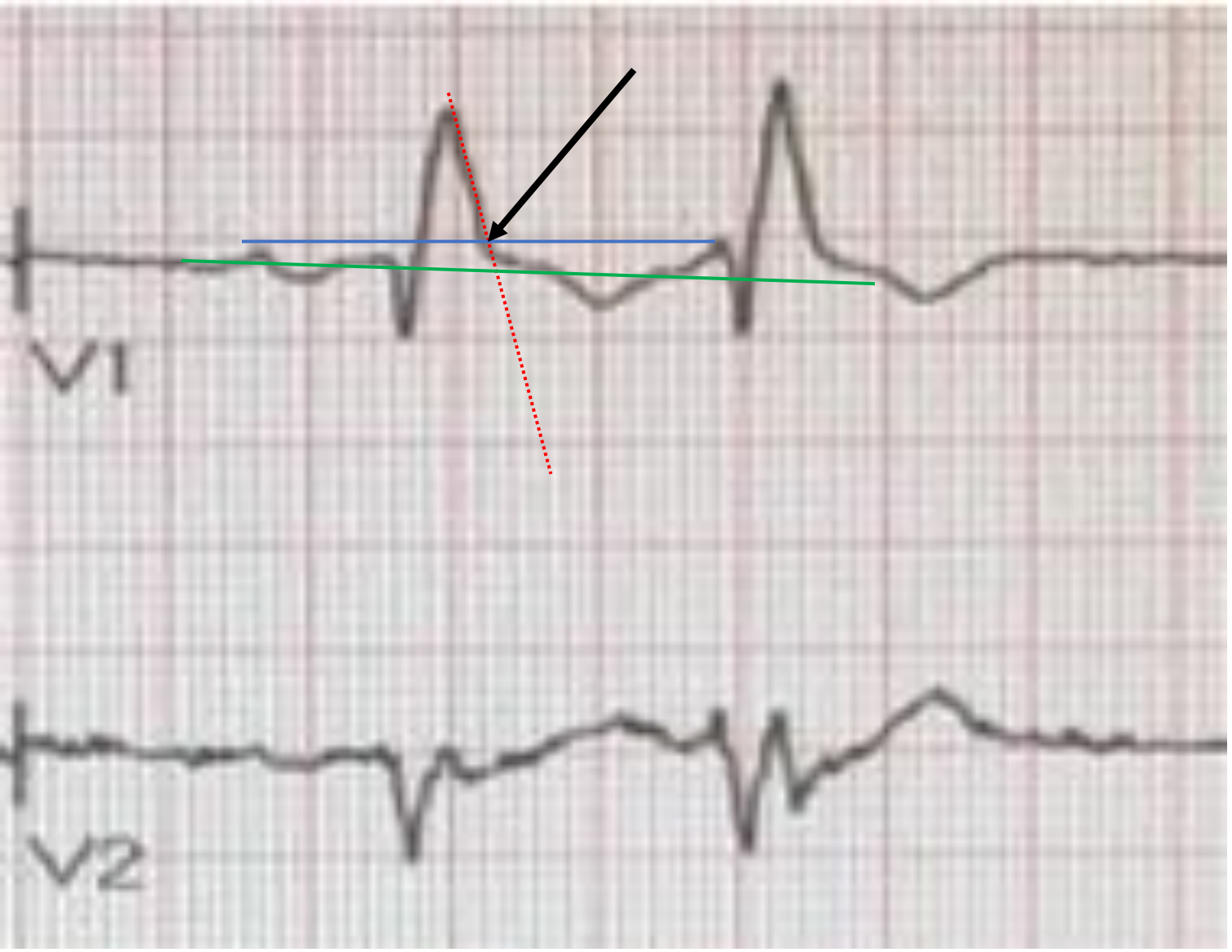
TABLE 1 Possible Clinical Scenarios in Which Right-Axis Deviation Can Be Observed

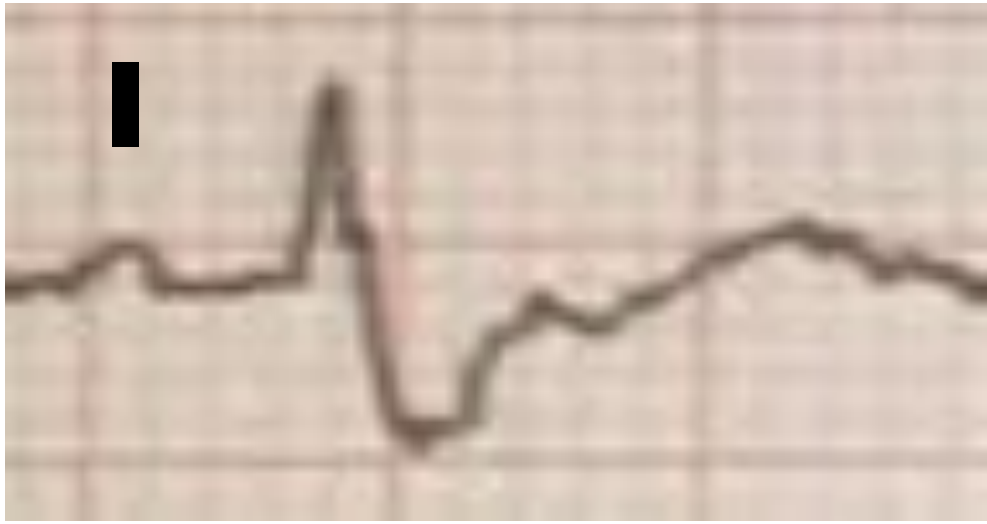
Clinical Scenario	Details
ECG lead misplacement	Reversal of right arm and left leg cables
Altered position of the heart in the chest	Dextrocardia
Ventricular rhythm	Ventricular tachycardia Accelerated idioventricular rhythm Ventricular escape rhythm
Severe hyperkalemia	
Drug intoxication	Tricyclic antidepressant, sodium channel blocker
Pulmonary emphysema	"Type C" right ventricular hypertrophy
Right superior fascicular block*	

*Previously described in patients with Brugada syndrome (1,2).

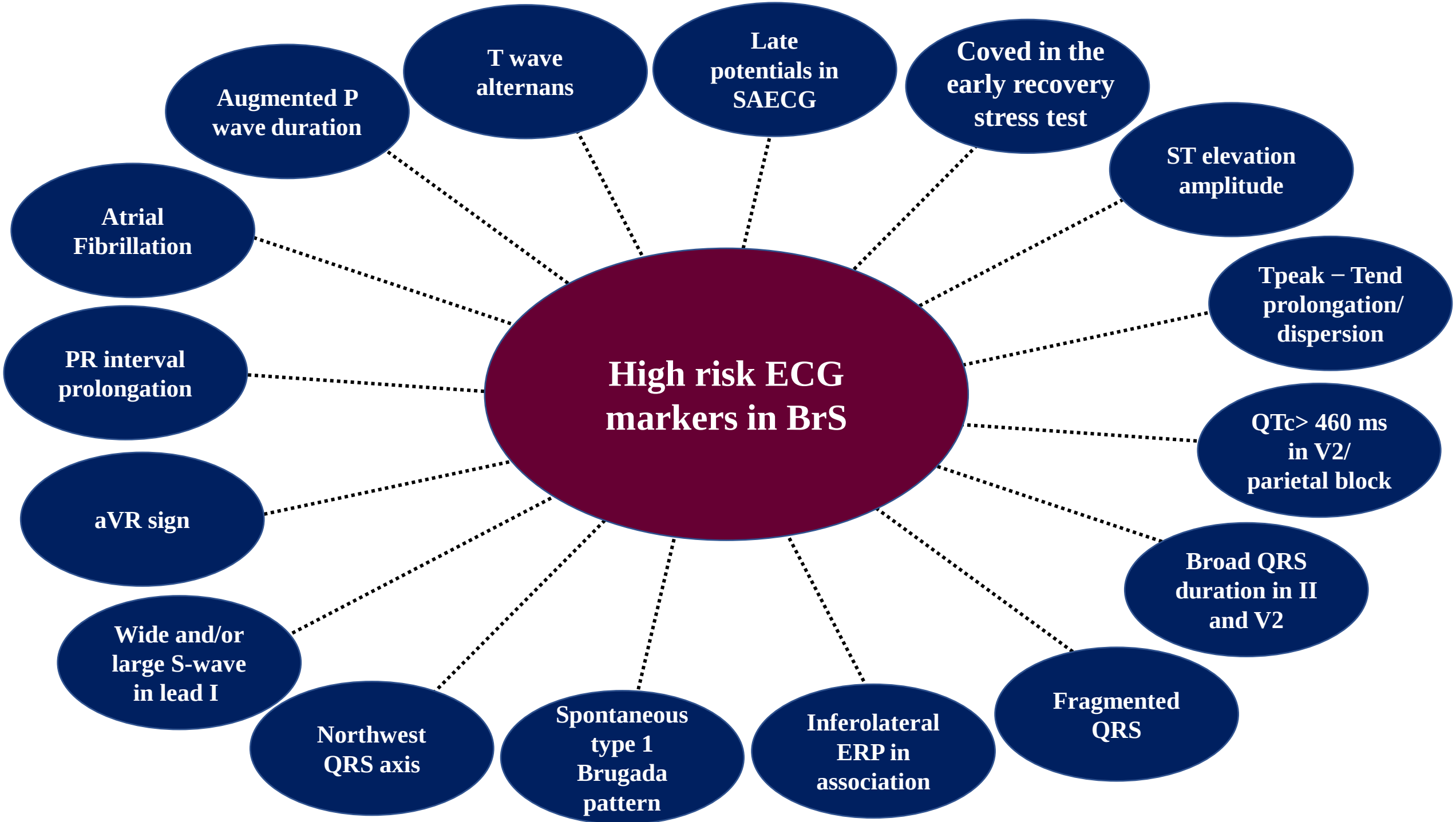


R wave ≥ 0.3 mV or R/q ≥ 0.75 in aVR (aVR sign). The aVR sign consists of a voltage of the final R wave of aVR> 3mm or 0.3mV or R/q ≥0.75. Presence of prominent final R wave on aVR is indicative of slow conduction at the RVOT may contribute to the induction of VF by EPS. Terminal tall and broad R wave of the QRS complex in lead aVR (**Mohamad Ali Babai Bigi , et al. aVR sign as a risk factor for life-threatening arrhythmic events in patients with Brugada syndrome. Heart Rhythm. 2007 Aug;4(8):1009-12.**





This patient has multiparametric risk factors: spontaneous type 1 Brugada pattern, Calò sign in I: the presence of a wide and/or large S-wave in lead I is a powerful predictor of life-threatening ventricular arrhythmias in patients with BrS and no history of cardiac arrest at presentation, Northwest QRS axis, fragmented QRS (second complex in V2).


High risk ECG markers in BrS
Augmented P wave duration
T wave alternans
Late potentials in SAECG
Coved in the early recovery stress test
ST elevation amplitude
Atrial Fibrillation
Tpeak − Tend prolongation/ dispersion
PR interval prolongation
QTc> 460 ms in V2/ parietal block
aVR sign
Broad QRS duration in II and V2
Wide and/or large S-wave in lead I
Fragmented QRS
Northwest QRS axis
Spontaneous type 1 Brugada pattern
Inferolateral ERP in association

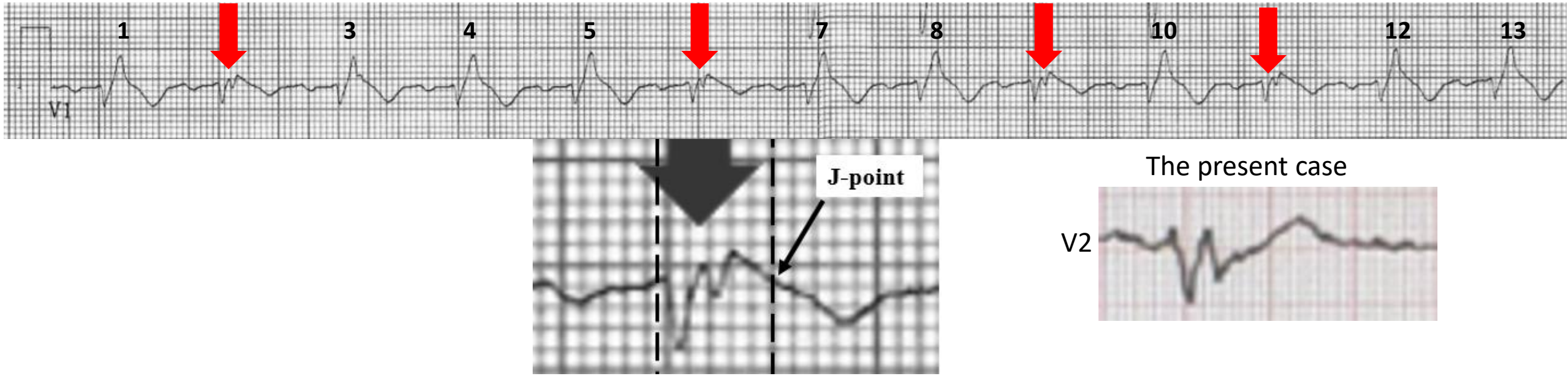
Variables identified as being associated with sudden cardiac death in Brugada syndrome.

Variable	Definition	Effect on SCD	Main publications
Aborted SCD		Increased risk	(Probst V et al. Circulation 2010; 121:635-43 DOI: 10.1161/CIRCULATIONAHA.109.887026) (Sacher F et al. Circulation 2013;128:1739–47)(10. https://doi.org/10.1161/CIRCULATIONAHA.113.001941) (Priori SG, et al J Am Coll Cardiol. 2012. J Am Coll Cardiol . 2012 Jan 3;59(1):37-45. doi: 10.1016/j.jacc.2011.08.064)
Syncope	Caused by arrhythmia	Increased risk	(Probst V et al. Circulation 2010; 121:635-43 DOI: 10.1161/CIRCULATIONAHA.109.887026)(Sacher F et al. Circulation 2013;128:1739–47. https://doi.org/10.1161/CIRCULATIONAHA.113.001941)(Priori SG, et al J Am Coll Cardiol. 2012. J Am Coll Cardiol . 2012 Jan 3;59(1):37-45. doi: 10.1016/j.jacc.2011.08.064)
Spontaneous ECG pattern	Type 1 ECG	Increased risk	(Probst V et al. Circulation 2010; 121:635-43 DOI: 10.1161/CIRCULATIONAHA.109.887026)(Sacher F et al. Circulation 2013;128:1739–47)(10. https://doi.org/10.1161/CIRCULATIONAHA.113.001941)(Priori SG, et al J Am Coll Cardiol. 2012. J Am Coll Cardiol . 2012 Jan 3;59(1):37-45. doi: 10.1016/j.jacc.2011.08.064)
Old age	Aged > 60 years	Decreased risk, but needs to be confirmed	(Conte G et al. J Cardiovasc Electrophysiol., 2014; 25:514-9. doi: 10.1111/jce.12359.)
Sex	Female sex	Decreases riskb	(Conte G et al. J Cardiovasc Electrophysiol., 2014; 25:514-9. doi: 10.1111/jce.12359.) (Benito B J, et al. 2008 Nov 4;52(19):1567-73. doi: 10.1016/j.jacc.2008.07.052.)

Variable	Definition	Effect on SCD	Main publications
EPS	**VF occurrence**	**Increased risk, with conflicting data, particularly with three extra stimulib**	**(Probst V et al. Circulation 2010; 121:635-43 DOI: 10.1161/CIRCULATIONAHA.109.887026) (Sacher F et al. Circulation 2013;128:1739–47. https://doi.org/10.1161/CIRCULATIONAHA.113.001941) (Sroubek J, et al.. Circulation . 2016 Feb 16;133(7):622-30. doi: 10.1161/CIRCULATIONAHA.115.017885.)**
Sinus dysfunction	**In females**	**Increased risk, but needs to be confirmedb**	**(Sieira J et al. Heart 2016; Mar;102(6):452-8. doi: 10.1136/heartjnl-2015-308556.)**
Early repolarization	**J wave > 0.1 mV in two contiguous inferolateral leads**	**Increased risk, with conflicting datab**	**(Kamakura S, Circ Arrhthm Electrophysiol Circ Arrhythm Electrophysiol . 2009 Oct;2(5):495-503. doi: 10.1161/CIRCEP.108.816892.)(Sarkozy A, Circ Arrhthm Electrophysiol. 2009 Apr;2(2):154-61. doi: 10.1161/CIRCEP.108.795153.)**
S wave in D1	S wave > 0.1 mV and/or > 40 ms	Increased risk, but needs to be confirmedb	**Calo L, et al. A new electrocardiographic marker of sudden death in brugada syndrome: the S-wave in lead I. J J Am Coll Cardiol . 2016 Mar 29;67(12):1427-1440. doi: 10.1016/j.jacc.2016.01.024..)**
QRS fragmentation	At least four spikes in one or at least eight spikes in all of the precordial leads	Increased risk, but needs to be confirmedb	**((Priori SG, et al J Am Coll Cardiol. 2012. J Am Coll Cardiol . 2012 Jan 3;59(1):37-45. doi: 10.1016/j.jacc.2011.08.064) (Morita H, et al. Fragmented QRS as a marker of conduction abnormality and a predictor of prognosis of Brugada syndrome. Circulation . 2008 Oct 21;118(17):1697-704. doi: 10.1161/CIRCULATIONAHA.108.770917. Epub 2008 Oct 6..)**

Complete/High-degree Right Bundle Branch Block obscuring the diagnosis of Brugada electrocardiographic pattern

In the presence of complete RBBB the premature activation of the right ventricle by apical pacing caused fusion beats, with narrower QRS complexes disclosing the distinctive coved-type ST-segment elevation in right precordial leads. The type 1 ECG Brugada pattern (coved type) may be totally or partially hidden by the secondary repolarization changes accompanying a complete or high-degree RBBB. Thus, a BrS should be ruled out whenever a slight ST-segment elevation is observed in right precordial leads in the presence of an RBBB. Eventually, under these conditions, right ventricular pacing may be used as a tool to unmask the Brugada ECG pattern during an electrophysiologic study.

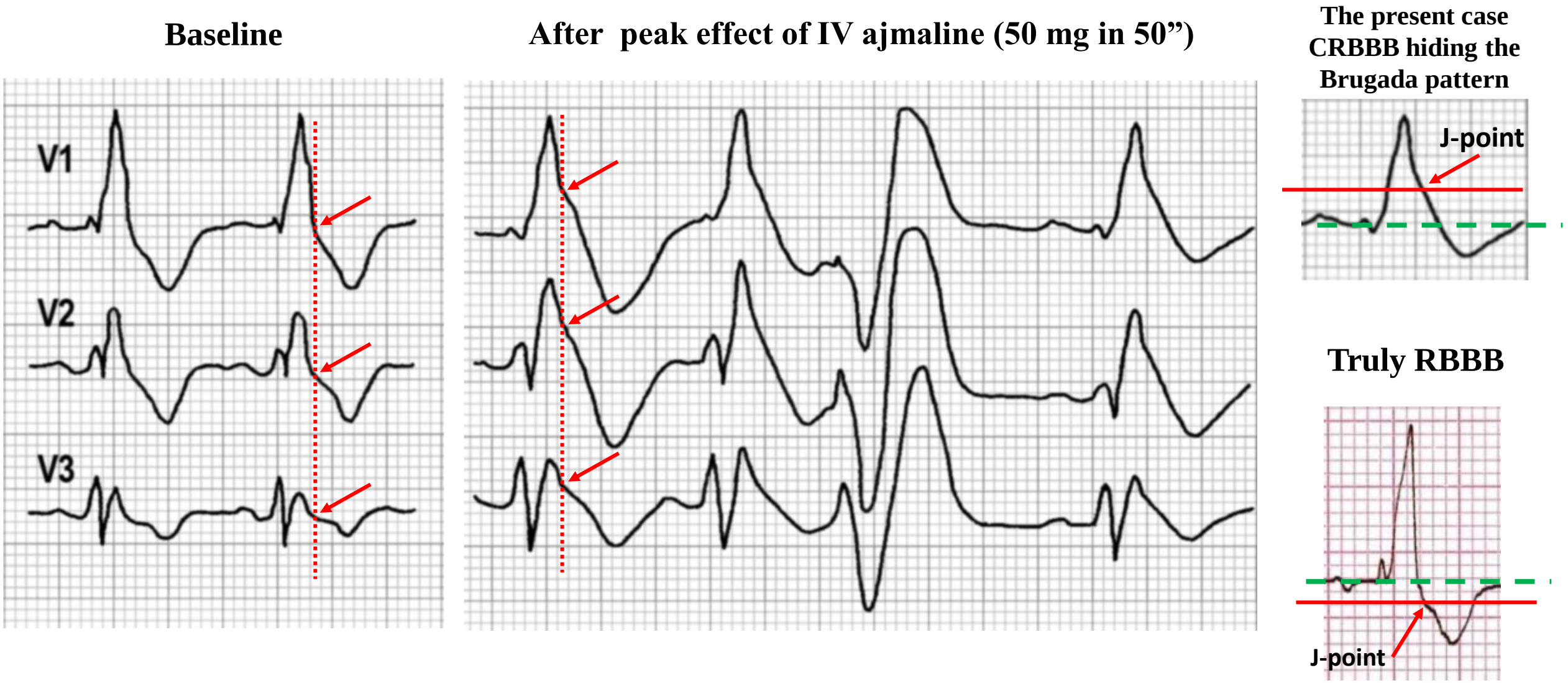


Beats 1, 3, 4, 5, 7, 8, 10, 12, and 13 are beats with high-degree right bundle branch block obscuring the diagnosis of Brugada electrocardiographic pattern. **Chiale PA, Garro HA, Fernández PA, Elizari MV. High-degree right bundle branch block obscuring the diagnosis of Brugada electrocardiographic pattern.Heart 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: 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**).

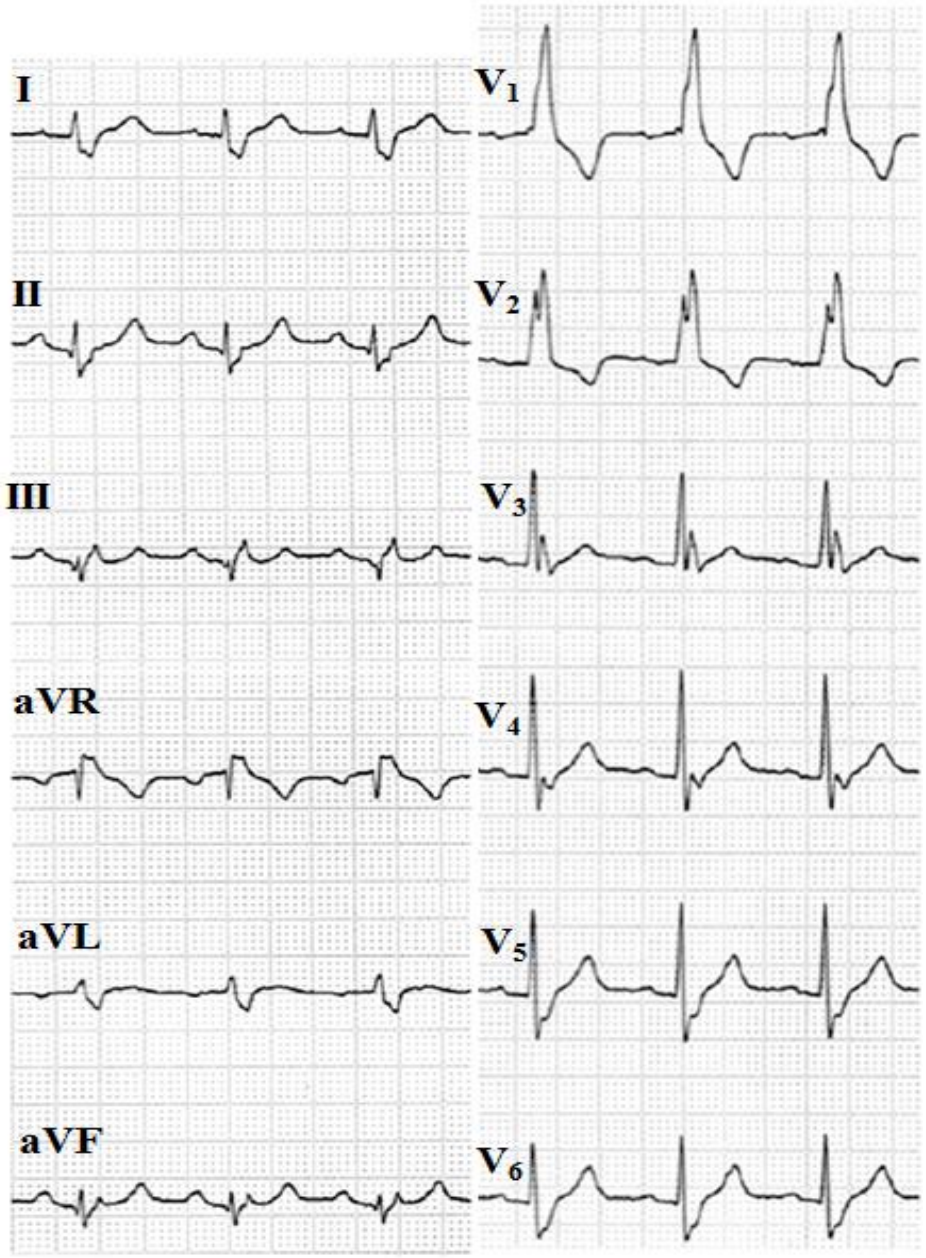
Variable	Definition	Effect on SCD	Main publications
Inferior type 1	Type 1 ECG in inferior or lateral leads	Increased risk, but needs to be confirmedb	**Rollin A, et al. Prevalence, characteristics, and prognosis role of type 1 ST elevation in the peripheral ECG leads in patients with Brugada syndrome. Heart Rhythm . 2013 Jul;10(7):1012-8. doi: 10.1016/j.hrthm.2013.03.001)**
Tpeak—Tend interval	Maximum Tpeak—Tend interval > 100 ms in precordial lead	Increased risk, but needs to be confirmedb	**Maury P, et al. Increased Tpeak-Tend interval is highly and independently related to arrhythmic events in Brugada syndrome. Heart Rhythm . 2015 Dec;12(12):2469-76. doi: 10.1016/j.hrthm.2015.07.029.)**
Post-exercise ST-segment elevation	≥0.05 mV in V1—V3 post exercise	Increased risk, but needs to be confirmedb	**Makimoto H, Nakagawa E, Takaki H, et al. Augmented STsegment elevation during recovery from exercise predicts cardiac events in patients with Brugada syndrome. J Am Coll Cardiol . 2010 Nov 2;56(19):1576-84. doi: 10.1016/j.jacc.2010.06.033.**.)
Young age	Aged < 18 years	Conflicting datac	(**Androdin A, et al. Heart Rhythm 2016;13:1274-82**)([**Conte G,, et al. Drug-induced Brugada syndrome in children: clinical features, device-based management, and long-term follow-up. J Am Coll Cardiol . 2014 Jun 3;63(21):2272-9. doi: 10.1016/j.jacc.2014.02.574.**
Family history of SCD	SCD in first-degree relatives	Conflicting datac	(**Probst V et al. Circulation 2010; 121:635-43 DOI: 10.1161/CIRCULATIONAHA.109.887026**)
Genetic	SCN5A mutations	Conflicting datac	((**Probst V et al. Circulation 2010; 121:635-43 DOI: 10.1161/CIRCULATIONAHA.109.887026**)

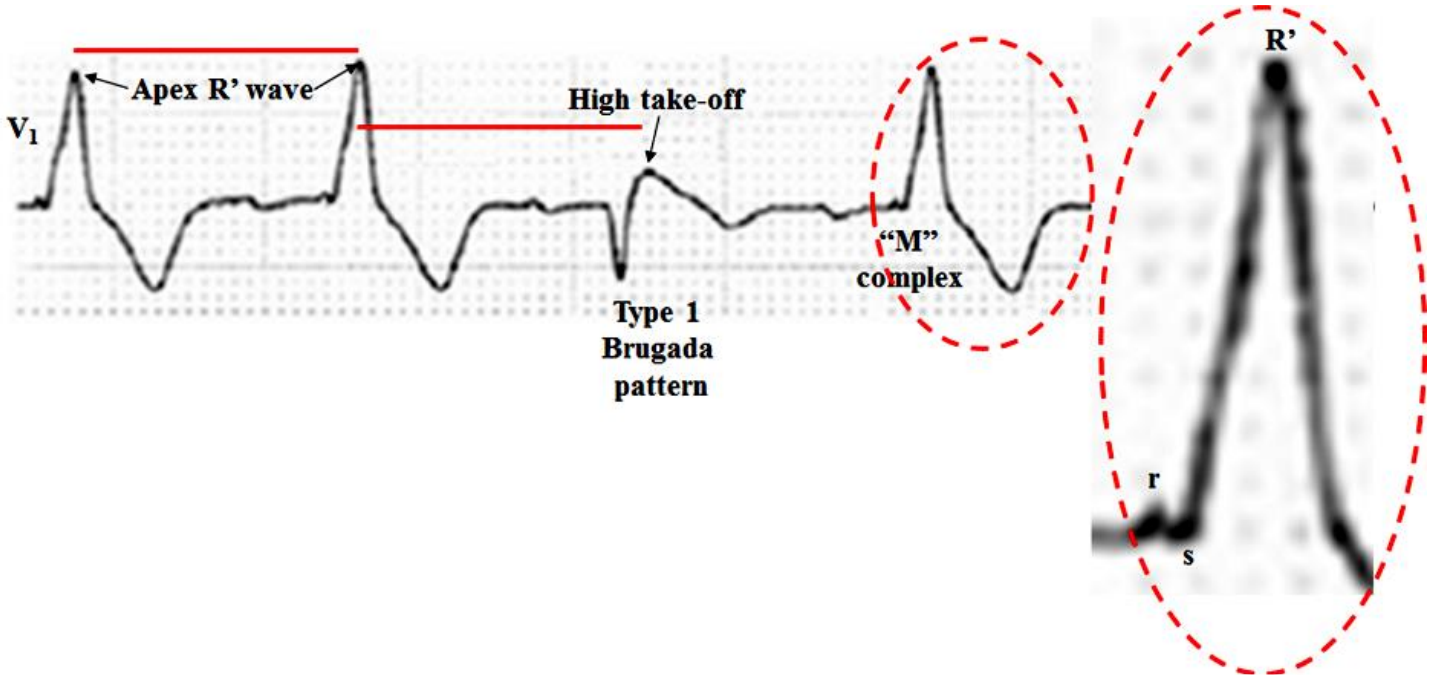
Variable	Definition	Effect on SCD	Main publications
Atrial fibrillation		Conflicting datac	((Probst V et al. Circulation 2010; 121:635-43 DOI: 10.1161/CIRCULATIONAHA.109.887026)(Kusano KF, Taniyama M, Nakamura K, et al. Atrial fibrillation in patients with Brugada syndrome relationships of gene mutation, electrophysiology, and clinical backgrounds. J Am Coll Cardiol 2008;51:1169—75.)
PR duration	>200ms	Conflicting datac	(Tokioka K, et al. J Am Coll Cardiol . 2014 May 27;63(20):2131-2138. doi: 10.1016/j.jacc.2014.01.072.) (Mauury P, et al Am J Cardiol . 2013 Nov 1;112(9):1384-9. doi: 10.1016/j.amjcard.2013.06.033)
QRS duration	≥ 120ms	Conflicting datac	(Tokioka K, et al. J Am Coll Cardiol . 2014 May 27;63(20):2131-2138. doi: 10.1016/j.jacc.2014.01.072.) ((Mauury P, et al Am J Cardiol . 2013 Nov 1;112(9):1384-9. doi: 10.1016/j.amjcard.2013.06.033))
Late Potential		Conflicting datac	(Circ J . 2002 Dec;66(12):1101-4. doi: 10.1253/circj.66.1101.)((Mauury P, et al Am J Cardiol . 2013 Nov 1;112(9):1384-9. doi: 10.1016/j.amjcard.2013.06.033))
aVR sign	R wave ≥ 0.3 mV or R/q ≥ 0.75 in aVR	Conflicting datac	(Probst V et al. Circulation 2010; 121:635-43 DOI: 10.1161/CIRCULATIONAHA.109.887026)

ECG: electrocardiogram; EPS: electrophysiological study; SCD: sudden cardiac death; VF: ventricular fibrillation. a—c An indication of the strength of data associating the variable with SCD (from a for consistent and prospective data to c for conflicting results) Gourraud, J.-B., Barc, J., Thollet, A., Le Marec, H., & Probst, V. (2017). *Brugada syndrome: Diagnosis, risk stratification and management. Archives of Cardiovascular Diseases, 110(3), 188–195.* doi:10.1016/j.acvd.2016.09.009 Gourraud, J.-B., Barc, J., Thollet, A., Le Marec, H., & Probst, V. (2017). *Brugada syndrome: Diagnosis, risk stratification and management. Archives of Cardiovascular Diseases, 110(3), 188–195.* doi:10.1016/j.acvd.2016.09.009

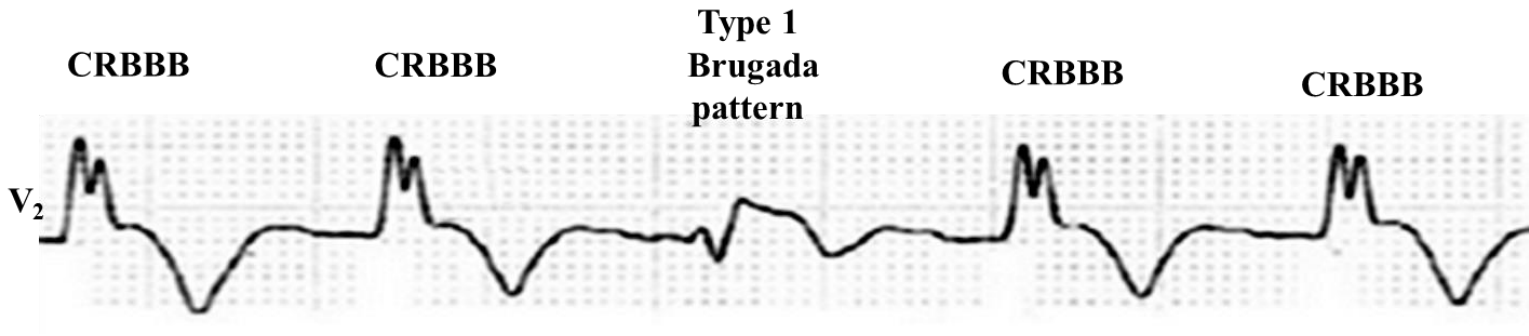


ECGs performed after 3 syncopal episodes at baseline and at the peak effect of IV ajmaline (50 mg in 50 seconds). At baseline, the J-point and ST segment are depressed and followed by asymmetrical negative T wave in leads V1 and V2 as a secondary change to a complete right bundle branch block. After ajmaline injection the J-point and ST-segment are elevated and followed by negative symmetrical T wave in leads V1, V2 and V3. The third beat is a PRC together with PVCs with “left bundle branch block” pattern followed by typical type 1 Brugada pattern.

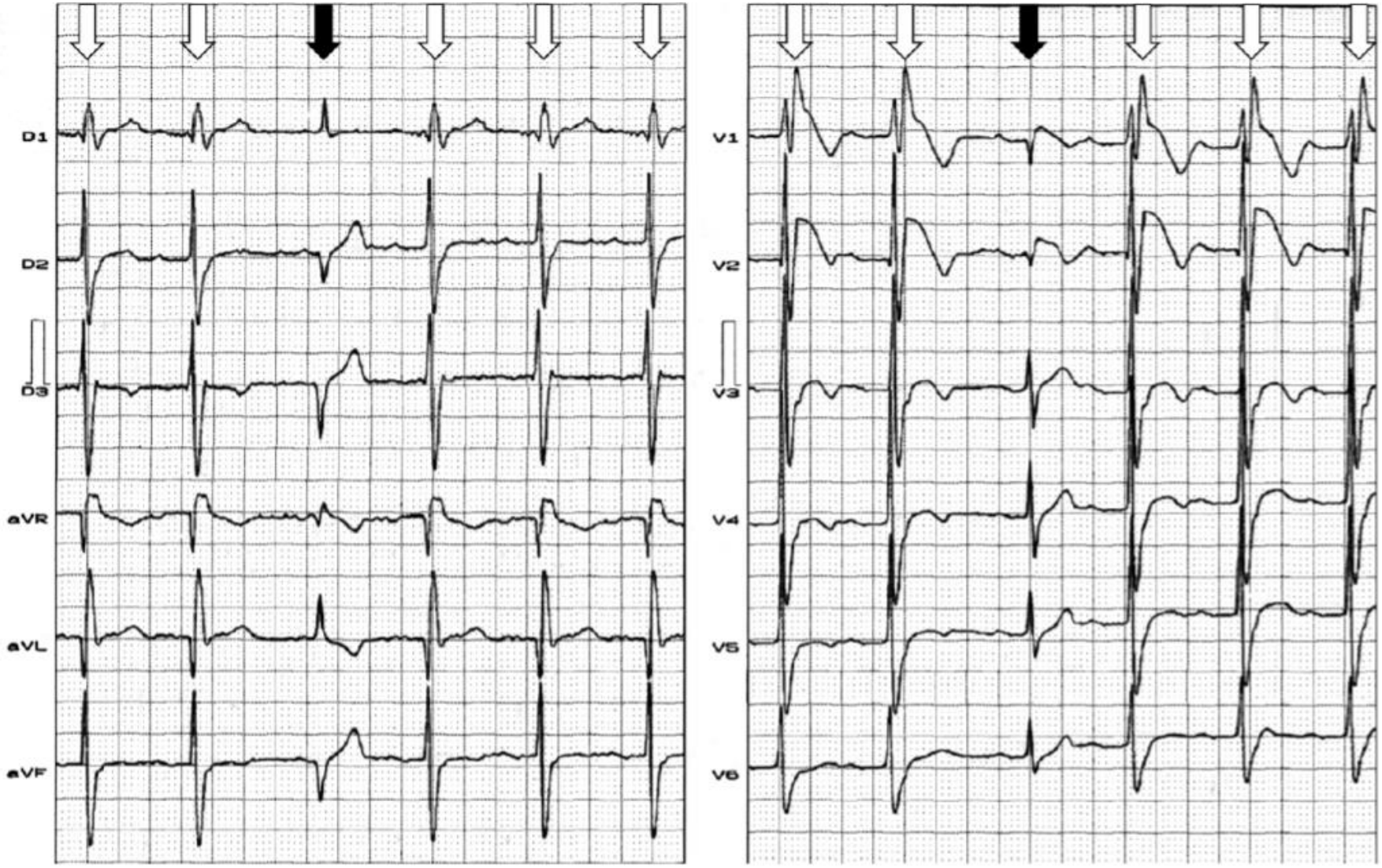




The first and second beats show CRBBB. The third beat without CRBBB (spontaneous transient or intermittent RBBB) shows type 1 Brugada pattern a loss of CRBBB and the normalized QRS complex. Spontaneous resolution of the CRBBB unmasks the type 1 Brugada pattern.



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.



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 (1).

1. **Peréz-Riera AR, et al. International VCG Investigators Group.Do patients with electrocardiographic Brugada type 1 pattern have associated right bundle branch block? A comparative vectorcardiographic study.Europace. 2012 Jun;14(6):889-97.**