Main electro-vectorcardiographic features in pectus excavatum (before and after correction) associated with Brugada syndrome

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Electrocardiographic features in Pectus Excavatum or Funnel chest

Pectus deformities, including pectus excavatum (PE) and carinatum. These deformities are among the most common congenital chest anomalies, with an overall incidence of PE in 23 of 10,000 births.(1) PE is more likely to occur in men than in women; however, female adolescents and women may be underdiagnosed because breast tissue could disguise the defect. In PE chest deformity consists of backward displacement of the sternum and costal cartilages giving rise to a depression in the xiphosternal area.

Pectus Excavatum or Funnel chest
Pectus excavatum o cofre en embudo
Pectus excavatum ou caixa em funil

Pectus carinatum (pigeon chest)
Pectus carinatum (pecho de paloma)
Pectus carinatum (peito de pombo)

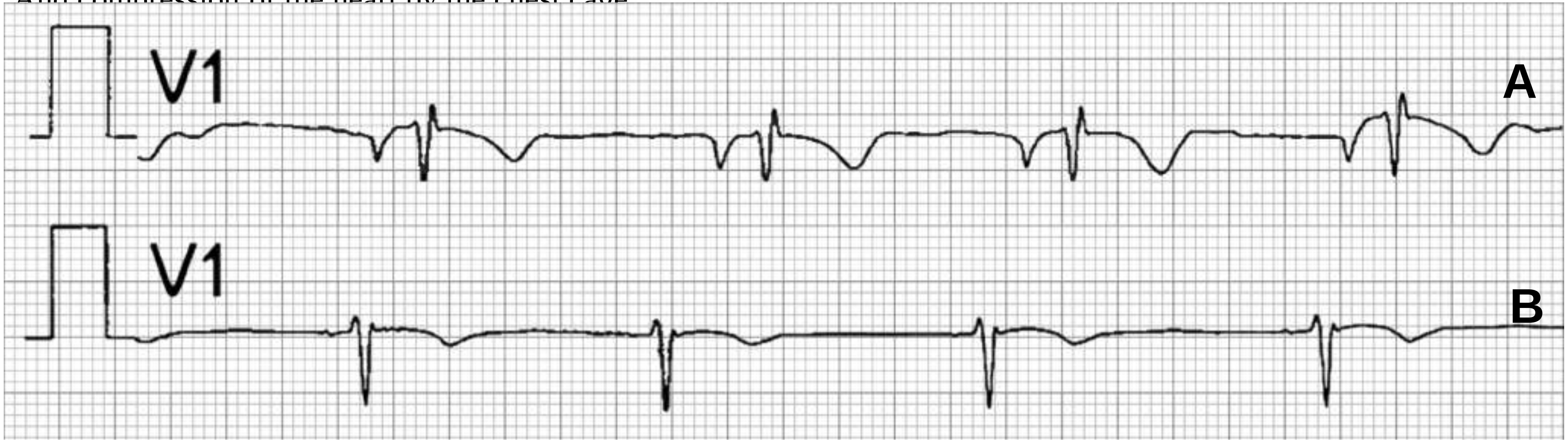
Electrocardiography

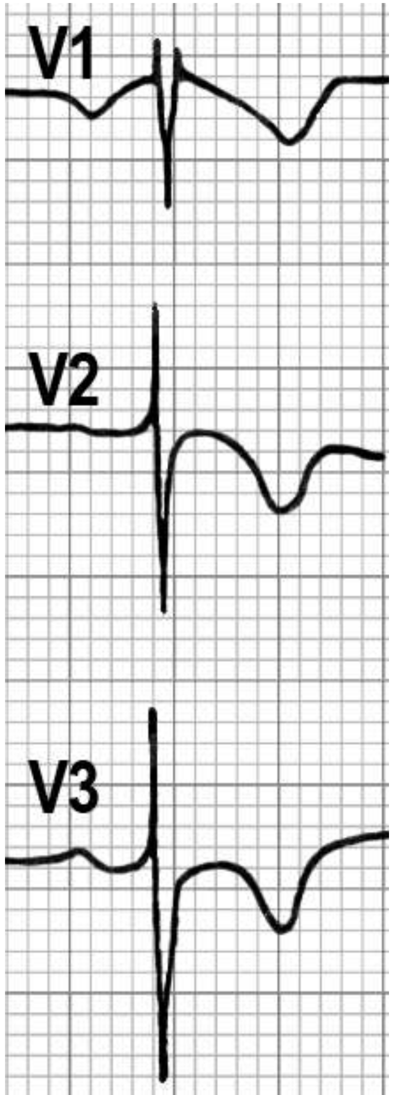
A 12-lead electrocardiogram should be preformed for all patients. and signs of atrial and ventricular hypertrophy, are frequently seen in patients with PE.(2) Sinus arrhythmias and premature ventricular beats have also been reported. (3)

Biphasic P waves in lead V_1 with a dominant negative force were found in 73% of 26 preoperative patients ECGs, and 12 patients(46%) showed a completely negative P wave in V_1 such as the present case. (2)

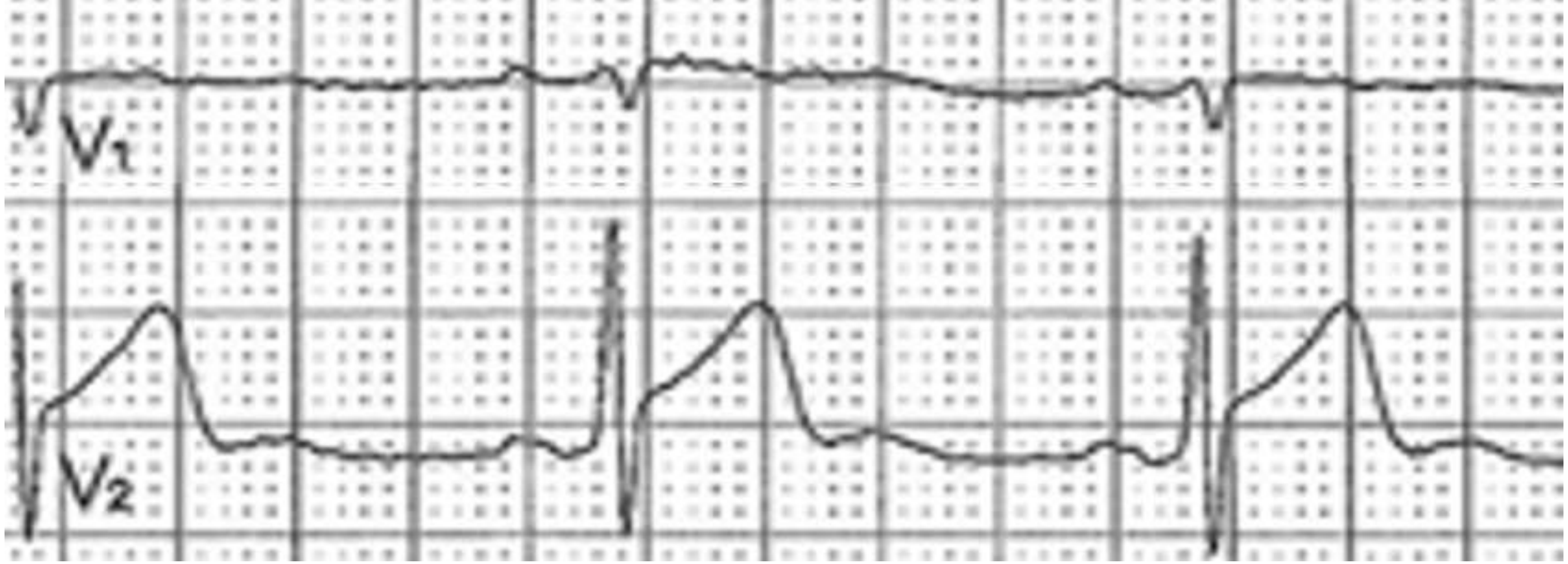
ECG changes are to be expected, because of alterations in the position of the heart with cardiac rotation

And compression of the heart by the chest cage





ECG recordings of P wave in lead V1 with a high negative amplitude before surgical correction. Very frequently PE patients have the atrium vector oriented backwards so producing a negative P wave in lead V1. These deep negative P wave frequently is mistaken with mitral stenosis with pulmonary hypertension pattern. (A) P-as loop backward orientation in the horizontal plane in all the cases. and with marked decreased negative force after the intervention (B).



Those cases where the right atrium (RA) assumes the position directly below the exploring electrode of V1, as a consequence of a greater rotation of the heart, this lead now reflects the RA intracavitary potentials and a qr or rs pattern appears only in V1 with concomitant RS in V2 (1). The simultaneous recording of V1 and of the RA cavity obtained in these cases is observed eventually in PE. The presence of a disproportion between the amplitudes of the QRS complex in V1 (small) and V2 (large) is also an argument in favor of the fact that the right auricle is situated immediately below the electrode of V1, since transmitted intracavitary forces obviously will not have the same amplitude as compared to the direct ones from the epicardial surface of the ventricle, as exhibited in V2 and the rest of the precordial leads. This phenomenon was called Peñaloza and Tranchesi sign: QRS complexes of low voltage in V1 contrasting with QRS complexes of normal voltage or increased in V2. In the absence of PE it is considered an indirect criteria of RA enlargement (RAE) (2) or ascending aortic aneurysm (3). Finally, we observe QRS voltage in V1 <6 mm. This ECG feature was observed a long time ago by Reeves et al in cases of RAE (4).

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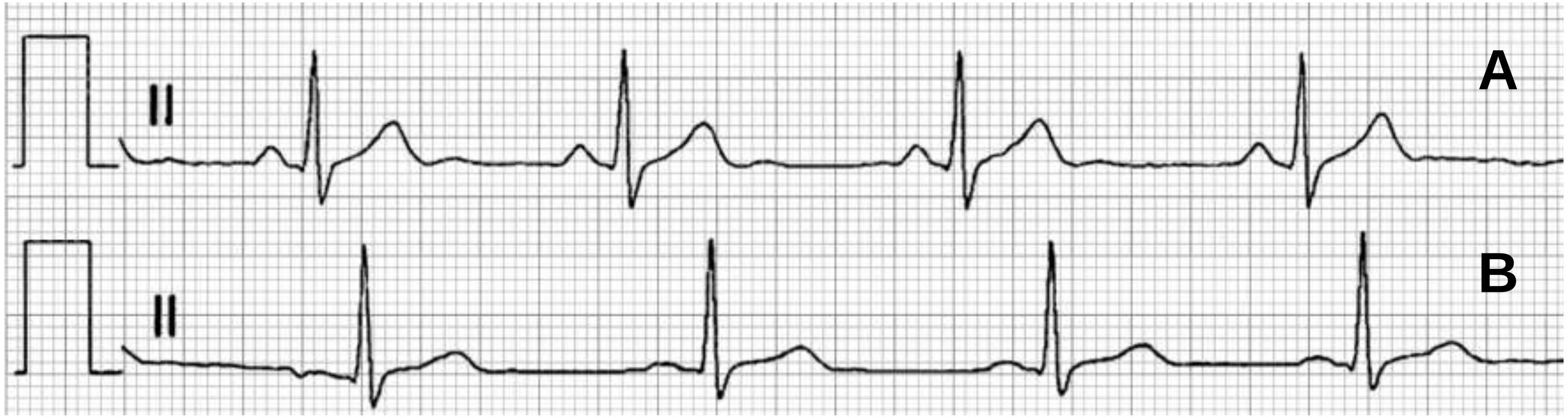
P wave amplitude in lead II decreased significantly after surgical correction.



ECG recordings showing P wave amplitude in lead II before (A) and after (B) surgical correction in the same patient.

In adults IRBBB is registered in 65% of cases. In children's Right ventricular conduction delay (rSr' or rsR' pattern) is registered in ≈ 31 % of cases. Although the incidence of right ventricular conduction delay is not related to the severity of the pectoral depression, qR or QS patterns in V1 were encountered more frequently with the more severe deformities. IRBBB is registered in 65% of cases Rarely children show CRBBB(**4**) Abnormalities in electrocardiogram results, most commonly incomplete right bundle branch block(5) Brugada pattern very rarely (6) and Brugada phenocopy(**7**)

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Adult young population 22 ± 7 (15–45) Preoperative and postoperative ECG characteristics.

	Before surgical correction	After surgical correction
P wave amplitude in lead II	Normal	Low amplitude
P-wave polarity in right precordial leads	❑ Biphasic plus/minus with predominant negative portion ❑ Deep totally negative	Marked decreased negative force
Normal QRS axis in frontal plane + 60° to +90°	70%	96%
Ínfero-lateral J wave pattern	8%	42% Only in lateral precordial leads leads V4–V6, after surgical funnel chest correction using pectus bar.
J-point elevation ≥1mm	8%	42%
Most frequent changes	IRBBB rSr′ 65%	IRBBB 50%
Amplitude of r b r′ in lead V1	41%	38%J point elevation
Right axis deviation (+90° to + 180°)	6%	1%
Left axis deviation (−90° to −30°)	2%	0

Recommendations in case of Brugada ECG patterns in athletes (Chung, E. H. (2015). *Brugada ECG patterns in athletes. Journal of Electrocardiology, 48(4), 539–543.* doi:10.1016/j.jelectrocard.2015.05.001 **)**

1. When presented with an athlete's screening ECG with a Brugada phenocopy, it is paramount to ensure that the precordial leads were placed properly and to repeat the ECG if necessary.
2. The diagnosis of BrS should be strongly considered if the baseline ECG or a high precordial lead ECG (with V1 and V2 placed as high as the 2nd intercostal space) reveals a Type 1 pattern (**Priori SG, Wilde AA, Horie M, et al. HRS/EHRA/APHRS Expert Consensus Statement of the Diagnosis and Management of Patients with Inherited Primary Arrhythmia Syndromes. Heart Rhythm 2013;10:1932–63**.). . The 2010 European Society of Cardiology recommendations, the 2012 Seattle Criteria, and the most recent Refined Criteria recommend referral of patients with Type 1 BrEPs to cardiac specialists(**Corrado D, Pelliccia A, Heidbuchel H, Sharma S, Link M, Basso C, et al. Recommendations for interpretation of 12-lead electrocardiogram in the athlete. Eur Heart J 2010;31:L243–59**.) (**Drezner JA, Ashley E, Baggish AL, et al. Abnormal electrocardiographic findings in athletes: recognizing changes suggestive of cardiomyopathy. Br J Sports Med 2013;47:137–52**.)(**Riding NR, Sheikh N, Adamuz C, et al. Comparison of three current sets of electrocardiographic interpretation criteria for use in screening athletes. Heart 2015;101:384–90**.)
3. A patient with a Brugada ECG patterns (BrEP) consistent with Type 2 and has a Type 1 BrEP induced by drug provocation with a sodium channel blocker is considered to have BrS (**Priori SG, Wilde AA, Horie M, et al. HRS/EHRA/APHRS Expert Consensus Statement of the Diagnosis and Management of Patients with Inherited Primary Arrhythmia Syndromes. Heart Rhythm 2013;10:1932–63**.).
4. Patients with a transient Type 1 BrEP brought on by a known offending agent (such as a tricyclic antidepressant) and/or fever may be at higher risk for a cardiac event and should be further evaluated by a cardiac specialist.

4. Asymptomatic patients with a Type 2 BrEP and no concerning family history do not require further workup. This group likely represents a low risk group (**Zorzi A, Migliore F, Marras E, et al. Should all individuals with a nondiagnositic Brugada-electrocardiogram undergo sodium-channel blockers test. Heart Rhythm 2012;9:909–16**)
5. Evaluation of Brugada-like ECG patterns in athletes that are not clearly Type 1 or Type 2 should involve confirmation of lead placement, clinical history (including family history) and application of published ECG criteria. The ST segment in true BrEP is more downsloping than in early repolarization patterns. The r' in true BrEP is broader than that in Brugada-like patterns and relatively new algorithms for evaluation have been proposed (**Zorzi A, Leoni L, Di Paolo FM, et al. Differential diagnosis between early repolarization of athlete's heart and coved-type Brugada electrogram. Am J Cardiol 2015;115:529–32**.)(**Chevallier S, Forclaz A, Tenkorang J, et al. New electrocardiographic criteria for discriminating between Brugada type 2 and 3 patterns and incomplete right bundle branch block. J Am Coll Cardiol 2011;58:2290–8**.)

ECG criteria to differentiate the type-2 Brugada pattern from ECG of healthy athletes with triphasic patter rSr'-wave in leads V1/V2

	Type-2 Brugada pattern	Healthy athletes with triphasic patter rsr'-wave in leads V1/V2
The duration of the base of the triangle at 0.5 mV (5 mm) from high take-off	≥160 ms (4 mm) has a specificity (SP) of 95.6%, sensitivity (SE) 85%, positive predictive value (PPV) 94.4%, and negative predictive value (NPV) 87.9%.	<160 ms
The duration of the base of the triangle at the isoelectric line	≥60 ms (1.5 mm) in leads V1-V2has an SP of 78%, SE 94.8%, PPV 79.3%, and NPV 93.5%.	<60 ms

The authors consider that especially the measurement of the base of the triangle of r ′ in V1 V2 taken at 5 mm from the peak of r ′ is much easier to perform than the Chevalier angle and has even greater accuracy.

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References related differentiation Brugada ECG patterns from RBBB in chronological order

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Trained Athletes have frequently an Incomplete Right Bundle Branch Block pattern (IRBBB). It is defined by a QRS duration <120 ms with a terminal R wave in lead V1 (rSR' pattern) and broad final S wave in lateral leads I and V6. The mildly delayed RV conduction in athletes is caused by RV remodeling, with increased cavity size and resultant increased conduction time, rather than an intrinsic delay within the His-Purkinje system. (1). This pattern represents a phenotype of cardiac adaptation to exercise and in the absence of other features suggestive of disease does not require further evaluation. The magnitude of exercise-induced RV remodeling might be a sport type and exercise dose-dependent phenomenon that manifests on the 12-lead ECG. At the tissue level, it seems plausible that exercise-induced RV enlargement might lead to a stretching of the terminal free wall RV Purkinje fibers and myocardium, with resultant benign prolongation of RV depolarization.(2) Alternatively, less likely, CRBBB might reflect a more pathologic form of RV remodeling that includes RV dilation and fibrotic replacement of the myocardium. Although the potentially deleterious effects of intense exercise training on the RV have been suggested, this remains an area of controversy. (3;4;5).

The RBBB pattern observed in the endurance athlete is often accompanied by the Sokolow-Lyon voltage criterion for LVH. See next slide ……

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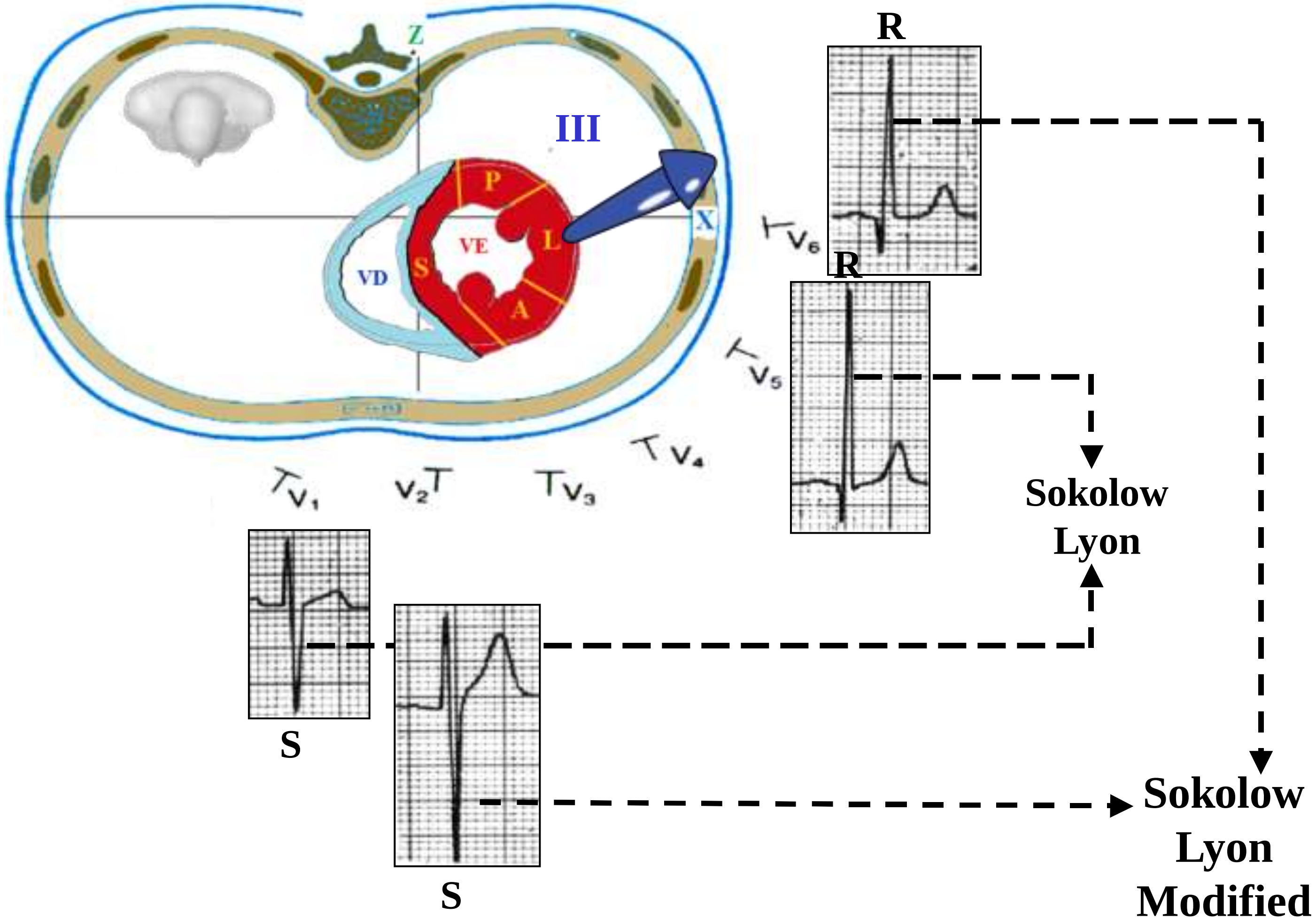
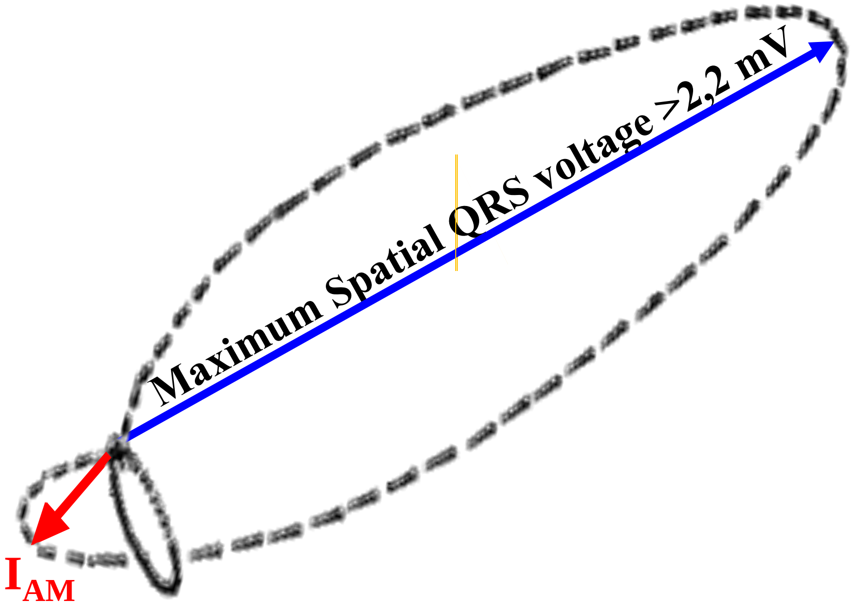
Sokolow-Lyon index and modified Sokolow for LVH (1;2)

S wave of V1 + R of V5 ≥ 35 mm or 3.5 mV in adults older than 30, > 40 mm or 4.0 mV between 20 and 30 years (Sokolow-Rapaport), > 60 mm between 16 and 20 years and > than 65 mm between 11 and 16 years. Sensitivity: 25%. Specificity: 95%.





Modified index: S wave of V2 + R of V5 or V6 ≥35 mm. This index uses a close lead (V2) and a distant one (V6). This is the reason why it has the same value as the Sokolow-Lyon index, which uses a distant lead (V1) and a close one (V5).

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Case report

A 46-year-old Caucasian male presented to the emergency room with atypical chest pain. His wife refers agonal breathing in several opportunities during sleep. He denied palpitations, syncope, or any other clinical manifestation. A brother died suddenly at 4-month's age during sleep with a picture of fever, attributed by the pediatrician to a flu. It is known that fever is the main trigger of potentially fatal events in children.

The physical examination showed mild left thorax deformity characterized by a depression of the sternum and ribs.

The electrolytes and echocardiogram were normal. There were no ventricular late potentials in signal-averaged ECG and cardiac imaging with multi-slice computed tomography (CT) showed a normal heart and mild pectus excavatum with a normal Haller index (2.2).1 This index is a mathematical relationship that exists in a human chest section observed with a CT scan. It is defined as the ratio of the transverse diameter and the anteroposterior diameter.

ECG and vectorcardiogram (VCG) were performed with the right precordial leads in the conventional position (Figure 1) and with the right precordial leads shifted to the second intercostal space (Figure 2), due to his familial background and agonal breathing reported by his wife. Genetic screening was requested, but the result is still unknown.

According to a consensus statement, an electrophysiological study (EPS) should be performed in asymptomatic patients with a family history of Brugada-associated sudden cardiac death (SCD).2 We performed an EPS with measurement of conduction intervals at baseline and three ventricular stimulations at the right ventricular apex, with negative results. The ventricular effective refractory period was >200 ms.

The pharmacological challenge was positive to induce a type 1 Brugada ECG pattern (Figure 3). This is the recommendation for the differential diagnosis between Brugada phenocopy (BrP) and genuine Brugada syndrome (BrS).3

Haller JA, Jr., Kramer SS, Lietman SA. Use of CT scans in selection of patients for pectus excavatum surgery: a preliminary report. J Pediatr Surg. 1987;22:904-6. doi: 10.1016/s0022-3468(87)80585-7

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ECG/VCG with right precordial leads in the conventional position

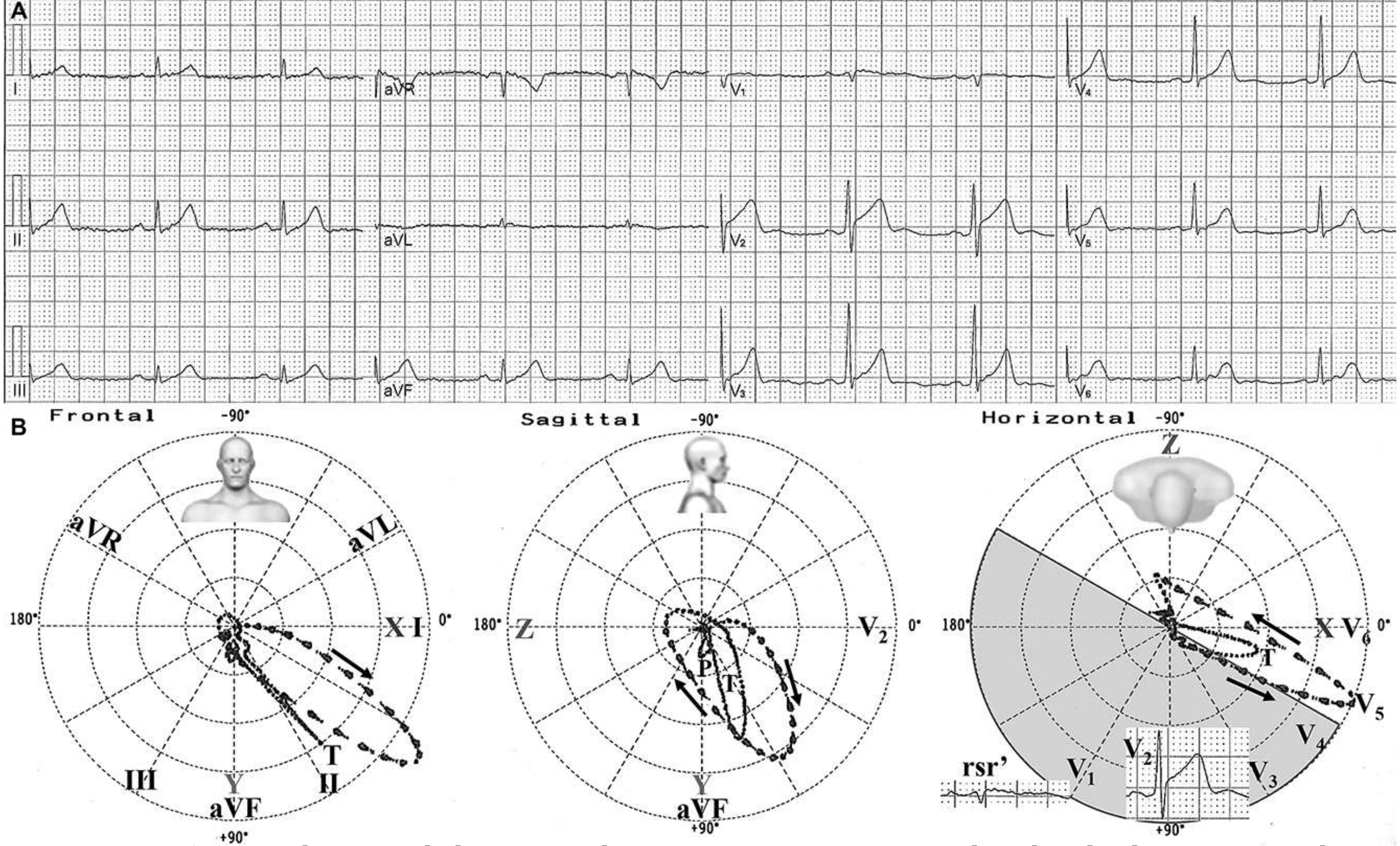
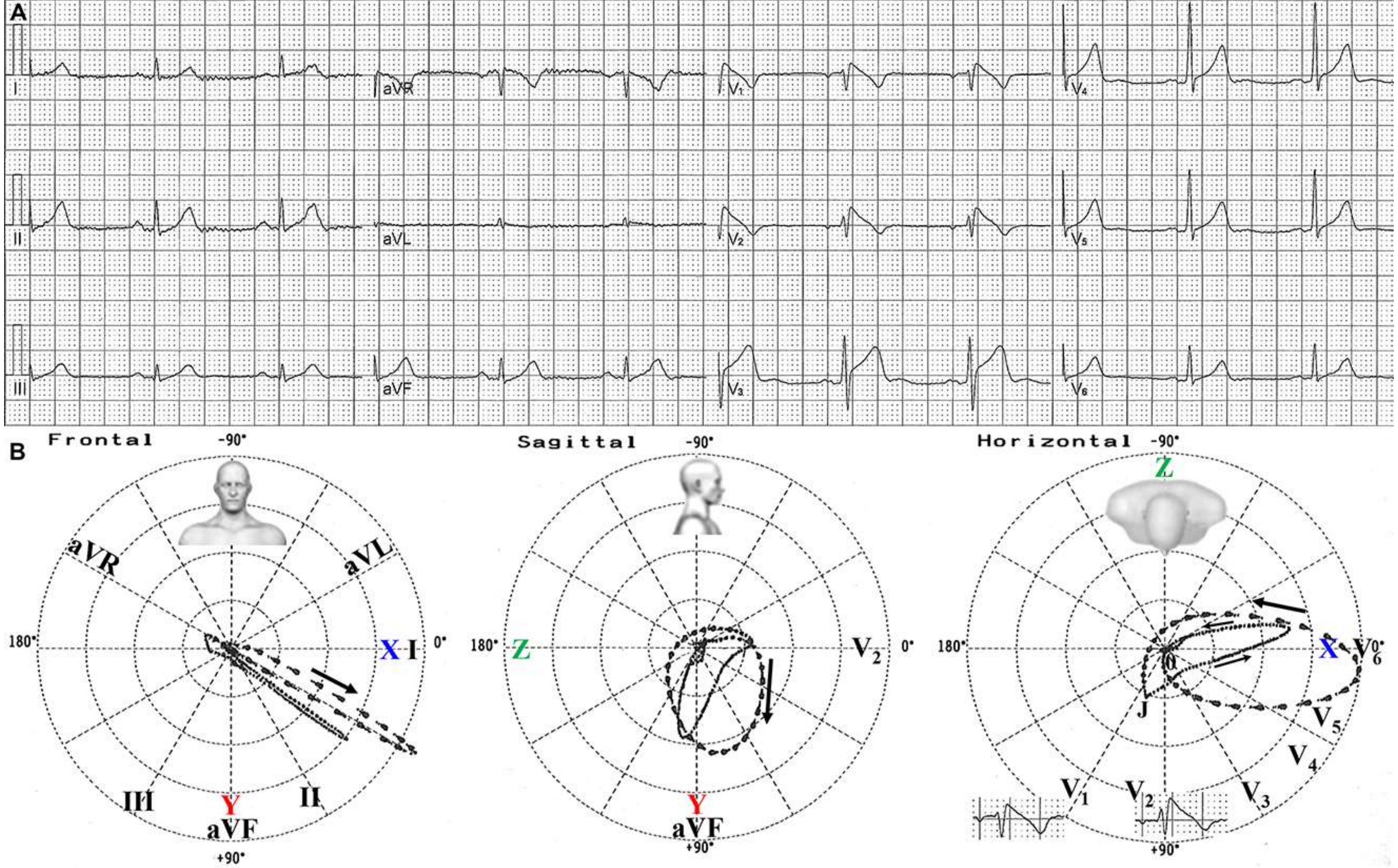


Figure 1: ECG (A) – early precordial transition because transition occurs earlier than lead V3. In V1 a disproportionally small total amplitude of the QRS complex when compared to V2. Low QRS voltage confined to V1. In normal hearts, the efferent QRS loop is located near +30° (perpendicular to V1) as a consequence of heart rotation around its longitudinal axis. VCG (B) – FP: QRS loop and T-loop are located in inferior left quadrant. RSP: P, QRS and T-loops are located in the anteroinferior quadrant. The QRS loop has terminal conduction delay located in the anterosuperior quadrant. HP: QRS loop directed to left and anteriorly near +30°, consequently perpendicular to V1 (+120°). This explains the contrast between the small QRS voltage of V1 and the normal voltage of V2. These findings are consistent with the Brugada VCG pattern.



12-lead ECG performed with right precordial leads shifted to the second intercostal space

Figure 2: ECG (A) – QRS axis +60°, totally negative P-wave is observed in V1 (it is considered typical of pectus excavatum), normal PR interval, typical spontaneous type 1 Brugada ECG pattern in V1-V2. VCG (B) – the P-loop is directed to the back and leftward with linear shape concave to the back and rightward in the HP. This P-wave pattern explains the totally negative P-wave registered in V1. The initial and terminal portion of the QRS loop are far from each other which indicates ST segment elevation in the right precordial leads. The terminal portion of the QRS-loop is located on anterior right quadrant and the driving speed is slow. The distance between the beginning of the T-loop (J point) is far from the end of T-loop (0 point) (≥2 mm), indicative of ST segment elevation in the right precordial leads, followed by negative T-wave.

Figure 3: ECG with precordial leads in the conventional position before (A) and after (B) ajmaline test

(A) Absence of terminal R wave in aVR, note the ST segment elevation with typical saddleback appearance in V2. The angle formed by the ascending and descending ramp of the final r' wave in V2 is broad. Conclusion: type 2 Brugada pattern.

(B) Typical type 1 Brugada ECG pattern, and prominent final R wave voltage in aVR, indicative of terminal conduction delay in the RVOT. Finally, I lead has QRS with RS pattern. Conclusion: positive ajmaline challenge, confirming BrS.

Even in the absence of any associated cardiac condition, ECG changes are expected in pectus excavatum, because of alterations in the cardiac position, and/or as a consequence of compression of the heart by the chest cage in the right ventricular outflow tract (RVOT). A small triphasic rsr' pattern is registered in ≈ 60% of cases, as a consequence of cardiac rotation around its longitudinal axis,4 as observed in the ECG of Figure 1. A triphasic pattern in the right precordial leads with normal QRS duration is the rule and may present a disproportionally small total amplitude of the QRS complex in V1 as compared to V2.5 The T-wave is always negative in V1. External pressure on the epicardium may produce a terminal conduction delay in the RVOT6 similar to that of the BrS as a consequence of reduction of connexin 43, and also in the concealed forms of arrhythmogenic right ventricular cardiomyopathy. The ECG of Figure 1A showed two patterns suggestive of pectus excavatum as a consequence of heart rotation around its longitudinal axis: a totally negative P wave and small QRS voltage in lead V1 contrasting with normal QRS voltage in V2. The second ECG (Figure 2A) showed a typical spontaneous type 1 Brugada ECG pattern. This ECG pattern could correspond to either a true BrS or a BrP, which cannot be differentiated by the ECG alone. BrP has been reported in pectus excavatum and has been attributed to RVOT compression by the thoracic deformity due to reduction of the anteroposterior thorax diameter.7 The ST-segment deviation is usually measured at its junction with the end of the QRS complexes, at the J point, and is referenced against the TP or PR segment.8

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Some authors prefer measuring the magnitude of the ST deviation 40 to 80 ms after the J point, when all myocardial fibers are expected to have reached the same level of membrane potential and to form an isoelectric ST segment.[9, 10] In VCG, the absence of ST segment elevation occurs when the J and 0 points are coincident (Figure 1B). BrPs may be induced by a several clinical circumstances that have been classified into six distinct etiological categories:[11] metabolic conditions, mechanical compression (pectus excavatum, mediastinal tumors), ischemia, myocardial and pericardial disease, ECG modulation, and miscellaneous.

We followed the recommendations of the 2013 consensus[12] on the management of patients with inherited primary arrhythmia: avoidance of drugs that may induce or aggravate ST-segment elevation in the right precordial leads (https://www.brugadadrugs.org/), excessive alcohol, large meals, and immediate treatment of fever with antipyretic drugs.

Conclusion

We present a case of a middle-aged Caucasian male with agonal respiration, a first-degree relative who died suddenly at young age during fever, and spontaneous type 1 Brugada ECG pattern with the right precordial leads shifted to the second intercostal space. The EPS was not inducible and ventricular refractory period was >200 ms. The ajmaline challenge was positive confirming the diagnosis of true BrS.

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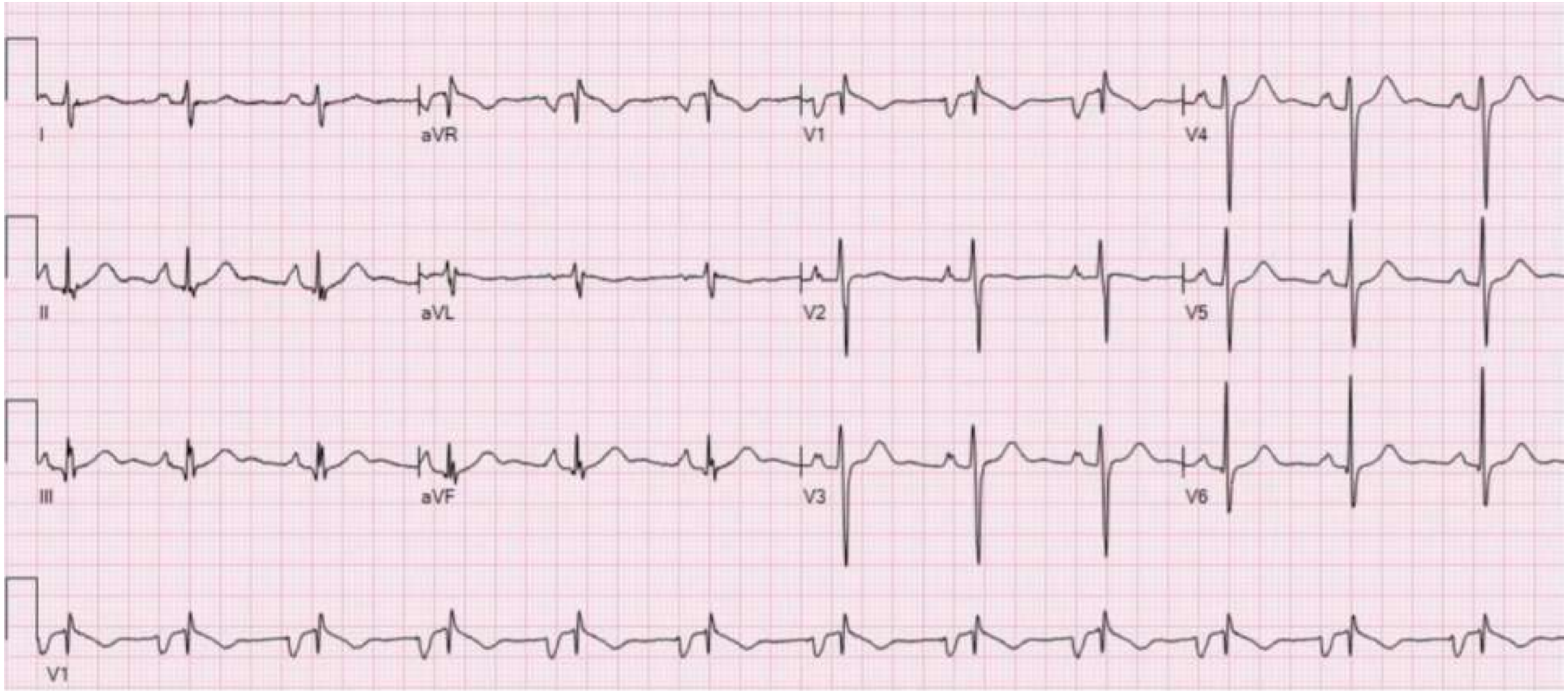
Pronounced pectus excavatum

Male 26 y

BMI 17

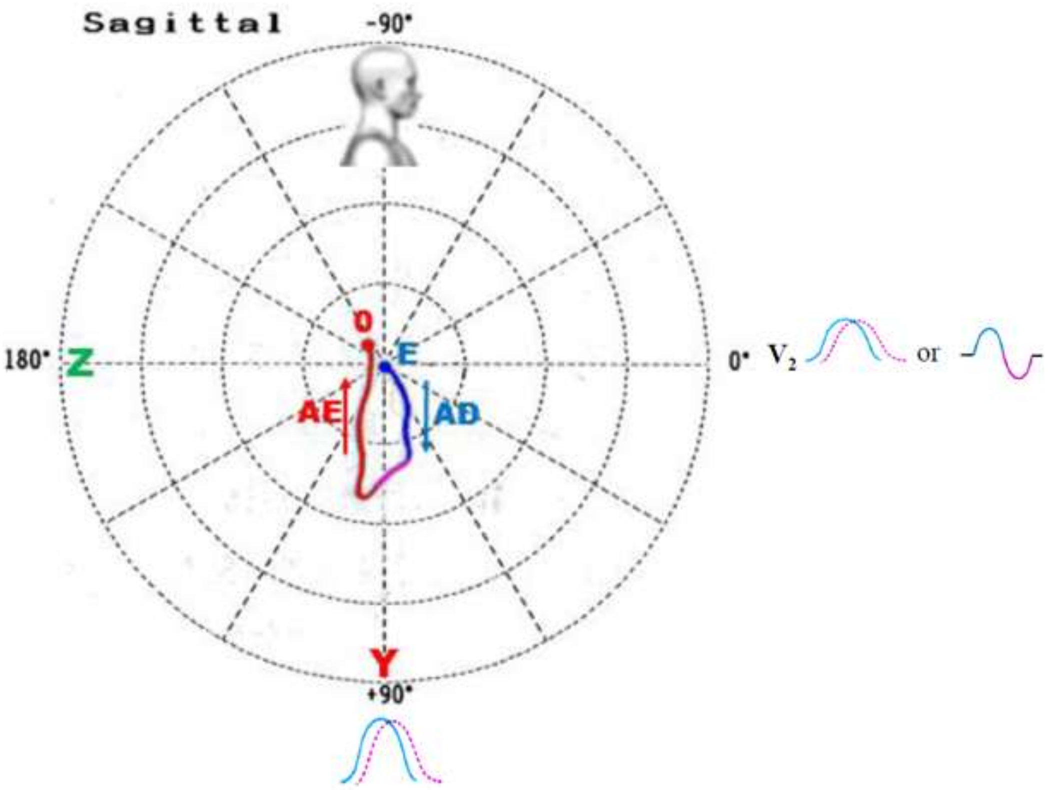
Refused operation

Case from Professor Kjell Nikus from Finland.

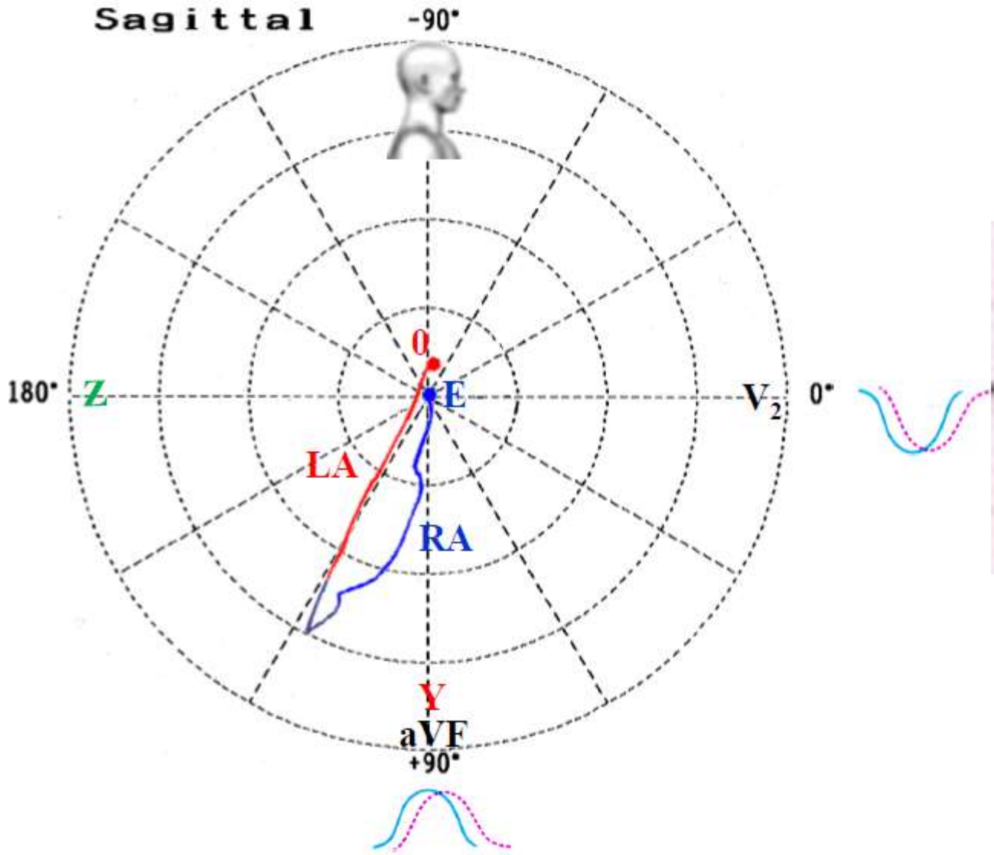


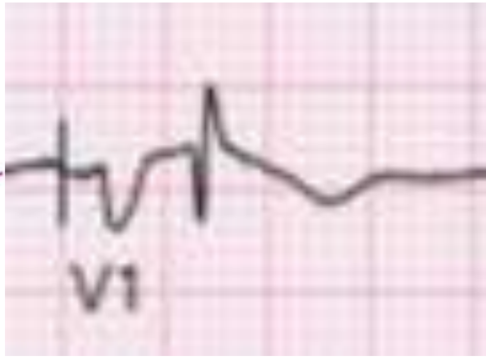
The change in cardiac position caused by the thoracic deformity in patients with pectus excavatum produces more or less constant ECG findings, which, although not pathognomonic, yet can be suggestive of this anomaly. These findings consist of: totally negative P waves in VI (backward orientation of the atrial vectors, and transmission of intracavitary potentials to the first right precordial leads), S1S3 or S1Q3; qr or rsr' in VI. These findings are chiefly related to clockwise rotation of the heart on its longitudinal axis,. These ECG changes, which, for different reasons, can be seen in other conditions like atrial septal defect or mitral stenosis with pulmonary hypertension, may lead to erroneous diagnosis LAE is analyzed without previous knowledge of the existence of such thoracic deformity.

Normal P-loop in the right sagittal plane



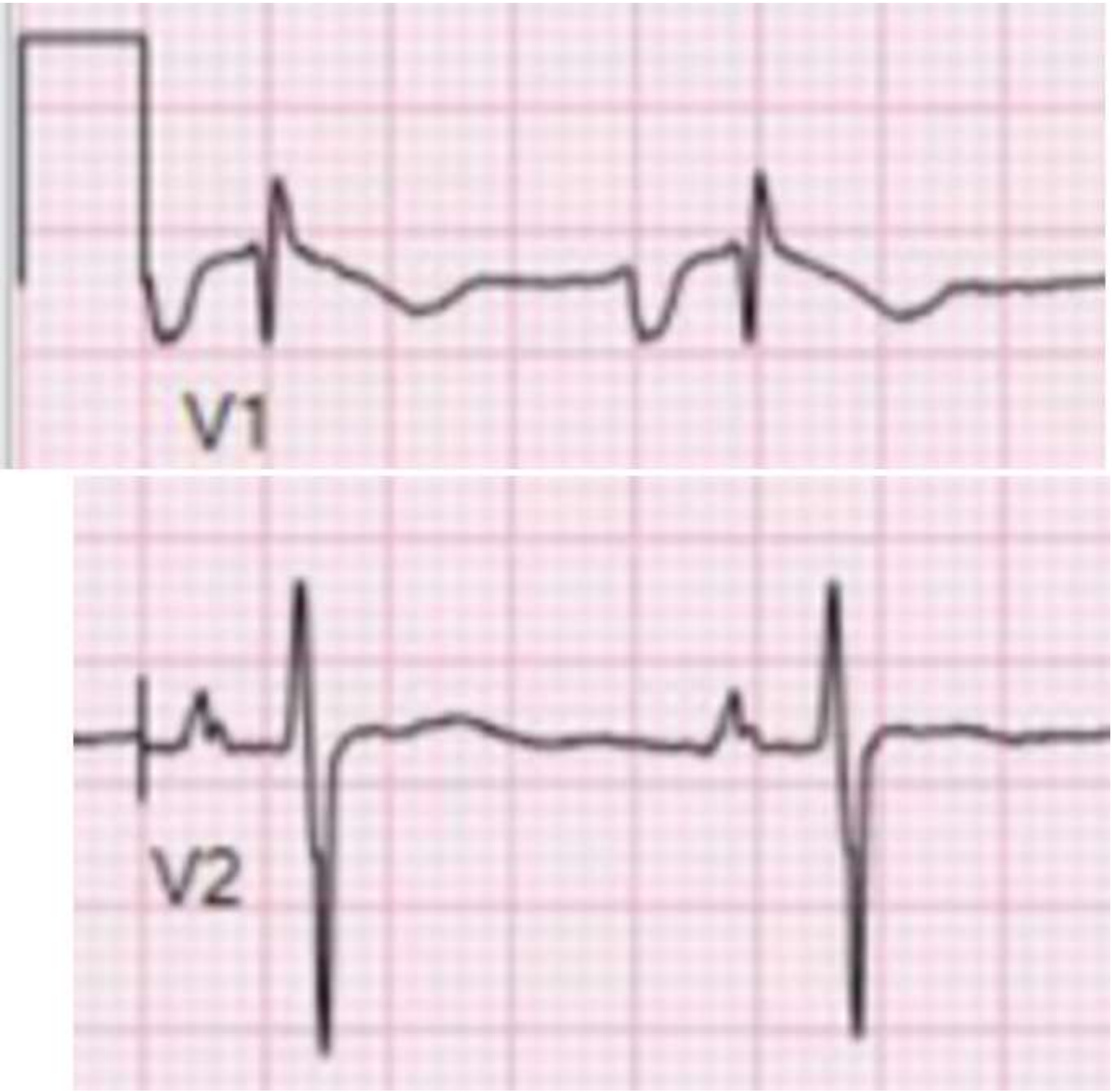
P-loop/P-wave in the right sagittal plane in a case of pectus excavatum





Totally negative P-wave in the V1, because all P-loops is located in the negative hemifield of this lead , consequence of posterior displacement of the. P-loop is totally positive in corrected Z and Y leads of VCG. RA: right atrium; LA: left atrium; E: E-point (the beginning of atrial depolarization at the sinus node; 0: the end of atrial depolarization (usually located backward and upward in relation to E-point).

Typical pattern of pectus excavatum in the right precordial leads



Deep negative P-wave in V1 because P-loop is dislocated to back.
Triphasic QRS pattern: rSR'

Pseudo right atrial enlargement: P wave height >1.5 mm in lead V2.

Deep negative P-wave in V1 + P wave height >1.5 mm in lead V2 = biatrial enlargement or pseudo biatrial enlargement consequence of intrathoracic heart rotation: PE

In Vichy, Barcelona at Professor Bayés de Luna’s wine cellar learning ECG from him. Unforgettable moments!!!