Myotonic Dystrophy Type 1 and Type 1 Brugada pattern: Brugada phenocopy or Brugada syndrome?

Português: Apresentação do caso

Um homem brasileiro caucasiano de 42 anos, foi encaminhado pelo oftalmologista para avaliar o risco cirúrgico de facectomia. Tinha diagnóstico clínico-genético da doença de Steiner desde os 22 anos. Queixava-se de uma longa história de fraqueza muscular progressiva e fenômeno miotônico predominantemente na porção distal das pernas, antebraços, pulsos, mãos e dedos. Além disso, referia ter disfagia, problema de mastigação, constipação, diabetes, e a perda progressiva da acuidade visual. Não foi possível saber a sua história familiar por ser filho adotivo. Nega qualquer queixa da esfera cardiovascular.

Exame Físico: catarata bilateral, pálpebras caídas, qualidade de voz nasal, pressão arterial 135/80 mmHg, frequência do pulso 55 bpm. Ausência de evidências de distensão da veia jugular no pescoço. Palpação do choque da ponta deslocado lateralmente um centímetro à esquerda da linha médio-clavicular no 5º espaço intercostal.

Primeiro (S1) e segundo (S2) ruídos normais, com desdobramento fisiológico na inspiração. Um terceiro ruído (S3) que aumenta de intensidade com a expiração e com cadência de galope. Ausência de sopros.

Pulmão: murmúrio vesicular normal. Taxa de inspiração/expiração 3:1. Ausência de estertores e sibilos.

Fígado palpável e doloroso a 2 cm da margem costal direita.

Ausência de edemas nas pernas, tornozelos ou pés.

Realizado eletrocardiograma (ECG) (Figura 1) e vetor cardiograma (VCG) nos PH, PF e PSD (Figura 2A, Figura 3 e Figura 4).

O paciente trazia um ECG realizado há três anos (Figura 5).

O ecocardiograma transtorácico mostrava anormalidade regional de movimento da parede, mínimo aumento do átrio esquerdo, mínima hipertrofia ventricular esquerda, O Doppler indica disfunção diastólica leve e comprometimento discreto das funções sistólica e diastólica. FEVE 50%. Além disso, foi solicitado: ECG de alta resolução (ECGAR), variabilidade da frequência cardíaca (VFC), Holter/24 horas, e a pesquisa genética para a síndrome de Brugada com um painel de 20 genes (SCN5A, ABCC9, CACNA1C, CACNB2, Cacna2d1, GPD1L, HCN4, KCND3, KCNE4, KCNE5, KCNJ8, pKP2, SCN10A, SCN1B, SCN2B, SCN3B , SCN5A, RANGRF, SLMAP e TRPM4), que foi negativo, e estudo eletrofisiológico (EPS).

Questões:

1. Qual é o diagnóstico ECG/VCG?
2. Qual é o diagnóstico clínico?
3. Qual é a abordagem adequada?
4. O que é o diagnóstico ECG da Figura 5?

English: Case presentation

A 42-year-old Caucasian Brazilian man referred by his ophthalmologist to evaluate the surgical risk for cataract surgery. He had clinical-genetic diagnosis of Steinert's disease since 22 years of age. He complained of a long history of progressive muscular weakness and myotonic phenomenon predominantly in the distal leg, forearm, wrists, hand and fingers. Additionally, he reported dysphagia, mastication problem, constipation, diabetes, and progressive loss of visual acuity. It was not possible to know their family history as he was an adopted son. He denied any sign of cardiovascular involvement.

Physical: He had bilateral cataracts, droopy eyelids, nasal voice quality, blood pressure 135/80 mmHg, pulse rate 55 bpm. Absence of evidence of jugular venous distension in the neck. Palpation of the point of maximum impulse verified with 1 cm lateral shift at the left of the midclavicular line at the 5th intercostal space. Absence of thrills.

Normal physiological first (S1) and second sounds (S2) with physiological split, during inhalation. S3 heart sound that increased in intensity with exhalation and had gallop cadence. Absence of heart murmurs.

Lung: normal vesicular breath sounds. Inspiration/expiration ratio 3:1. Absence of crackles and wheezes.

A palpable and painful liver 2 cm from the right costal margin.

Absence of swelling in legs, ankles, or feet.

We performed electrocardiogram (ECG) (Figure 1), vectorcardiogram (VCG) (Figure 2A, Figure 3, Figure 4). He brought an ECG performed 3 years before (Figure 5). We requested: signal-averaged ECG (SAECG), heart rate variability (HRV), 24-hour Holter and ambulatory transthoracic echocardiogram (TTE), genetic screening for Brugada syndrome with a panel for 20 genes (SCN5A, ABCC9, CACNA1C, CACNB2, CACNA2D1 GPD1L, HCN4, KCND3, KCNE4, KCNE5, KCNJ8, PKP2, SCN10A, SCN1B, SCN2B, SCN3B, SCN5A, RANGRF, SLMAP and TRPM4) that was negative; and electrophysiology study (EPS).

Transthoracic echocardiogram (TTE): regional wall motion abnormality, minimal left atrial enlargement, minimal left ventricular hypertrophy, Doppler evidence of mild diastolic dysfunction and discrete impairment of ventricular systolic and diastolic functions. LVEF 50%,

Signal-Averaged ECG (SAECG): positive late potentials.

Questions:

1. What is the ECG/VCG diagnosis?
2. What is the clinical diagnosis?
3. What is the appropriate approach?
4. What is the ECG diagnosis of the Figure 5?

Figure 1

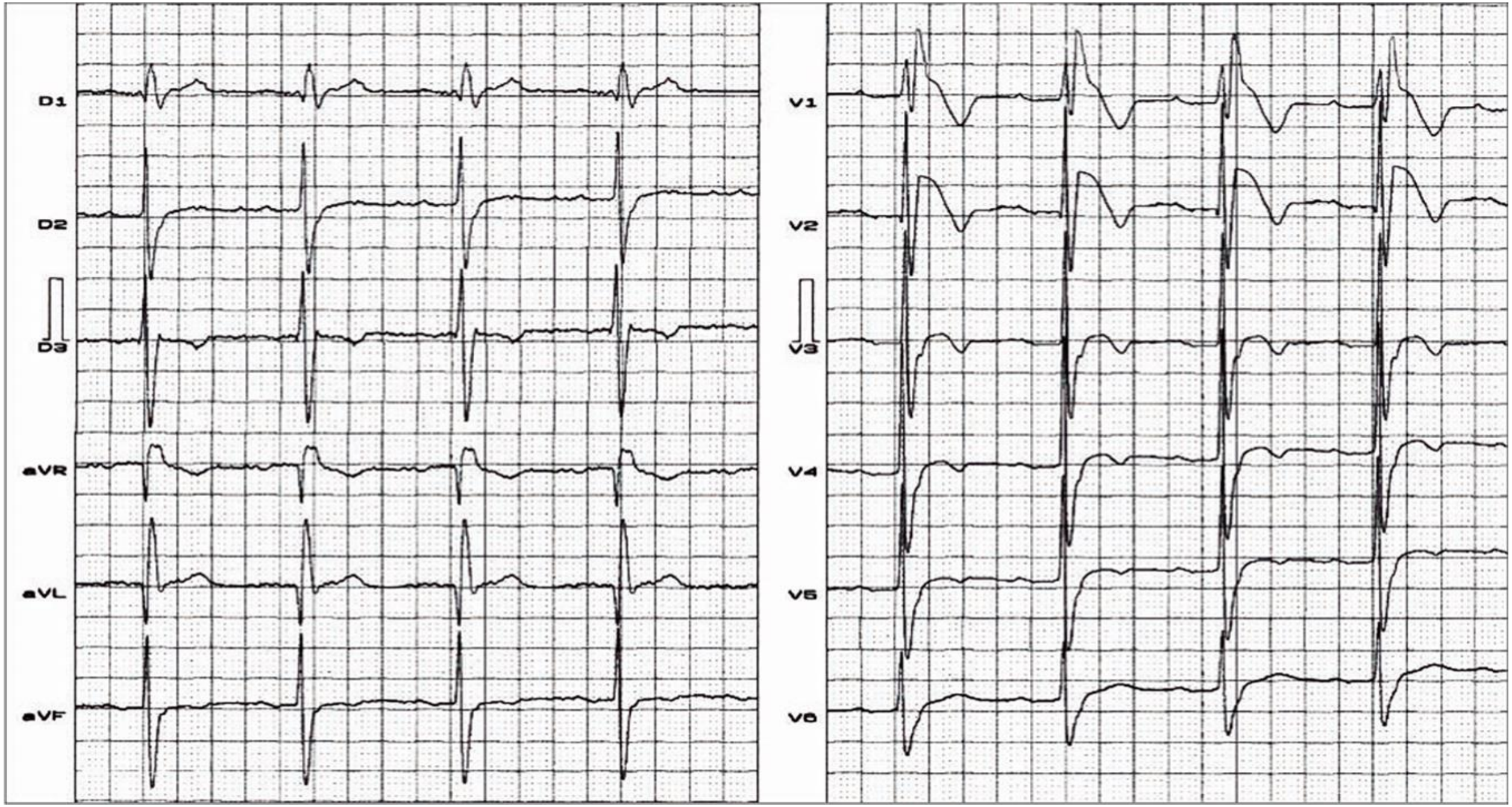
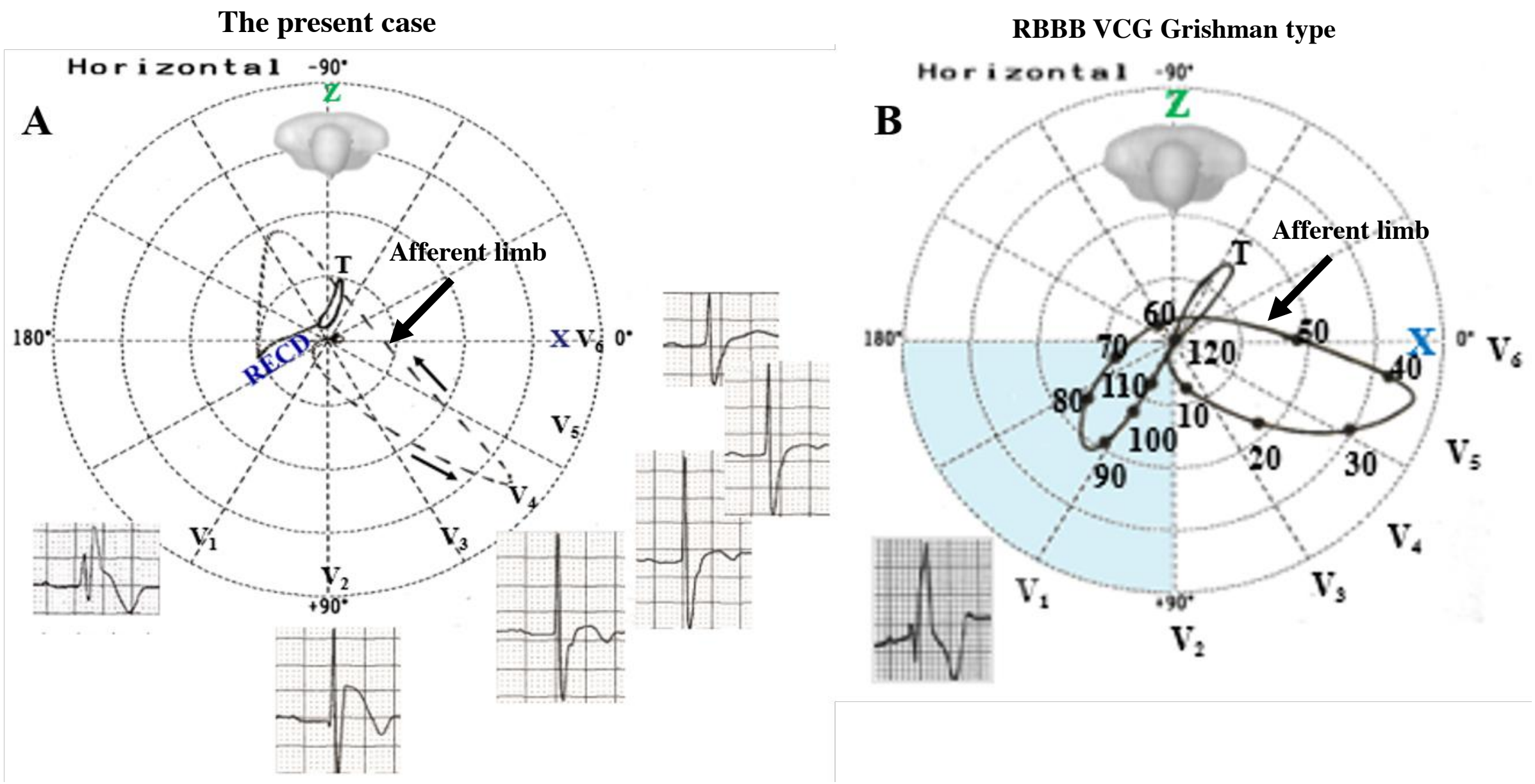


Figure 2 - ECG/VCG correlation in the Horizontal Plane



In both cases the afferent limb of QRS loop is located behind X orthogonal leads

Figure 3 – ECG/VCG correlation in the Frontal Plane

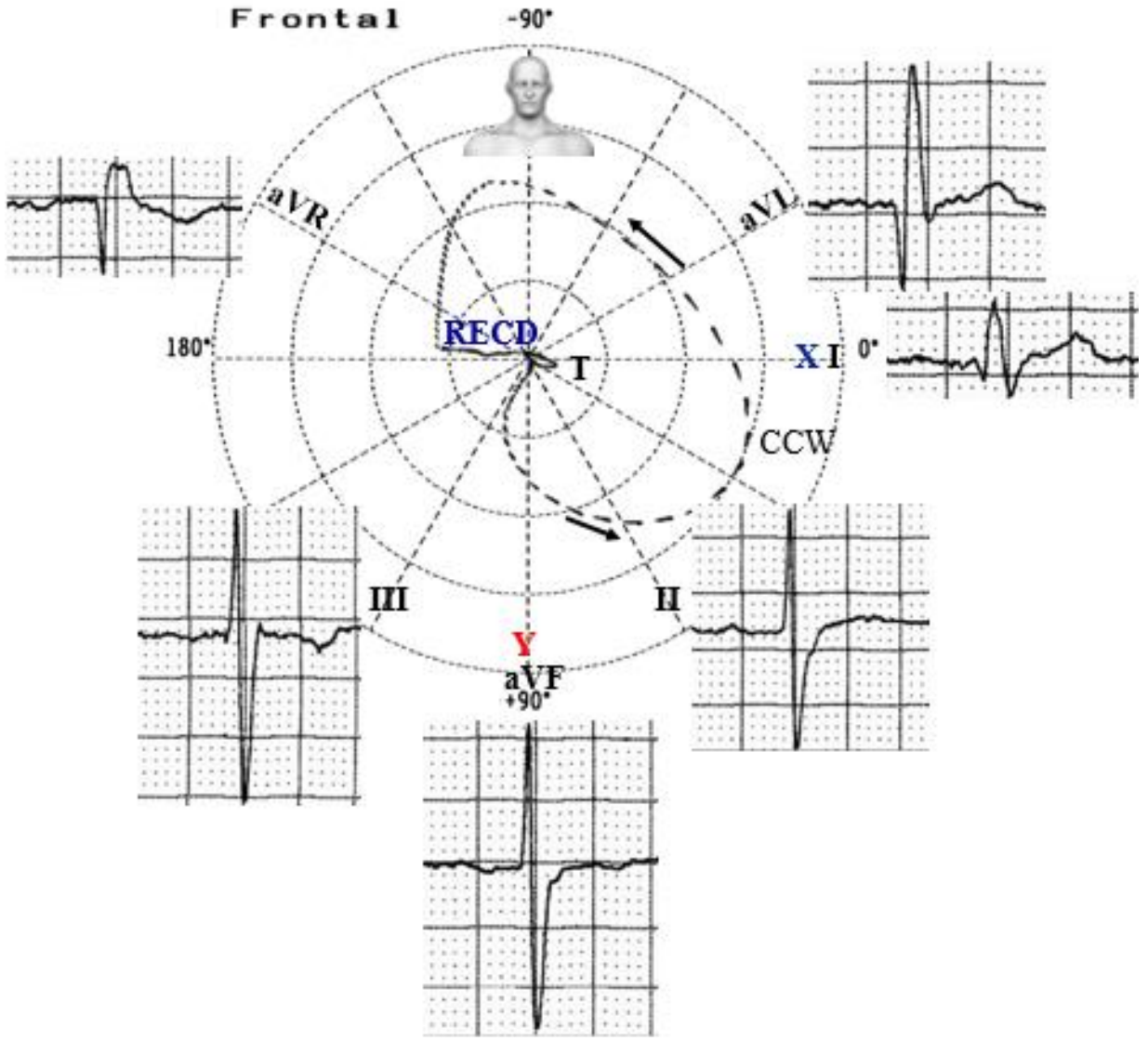


Figure 4 – ECG/VCG correlation in the Right Sagittal Plane

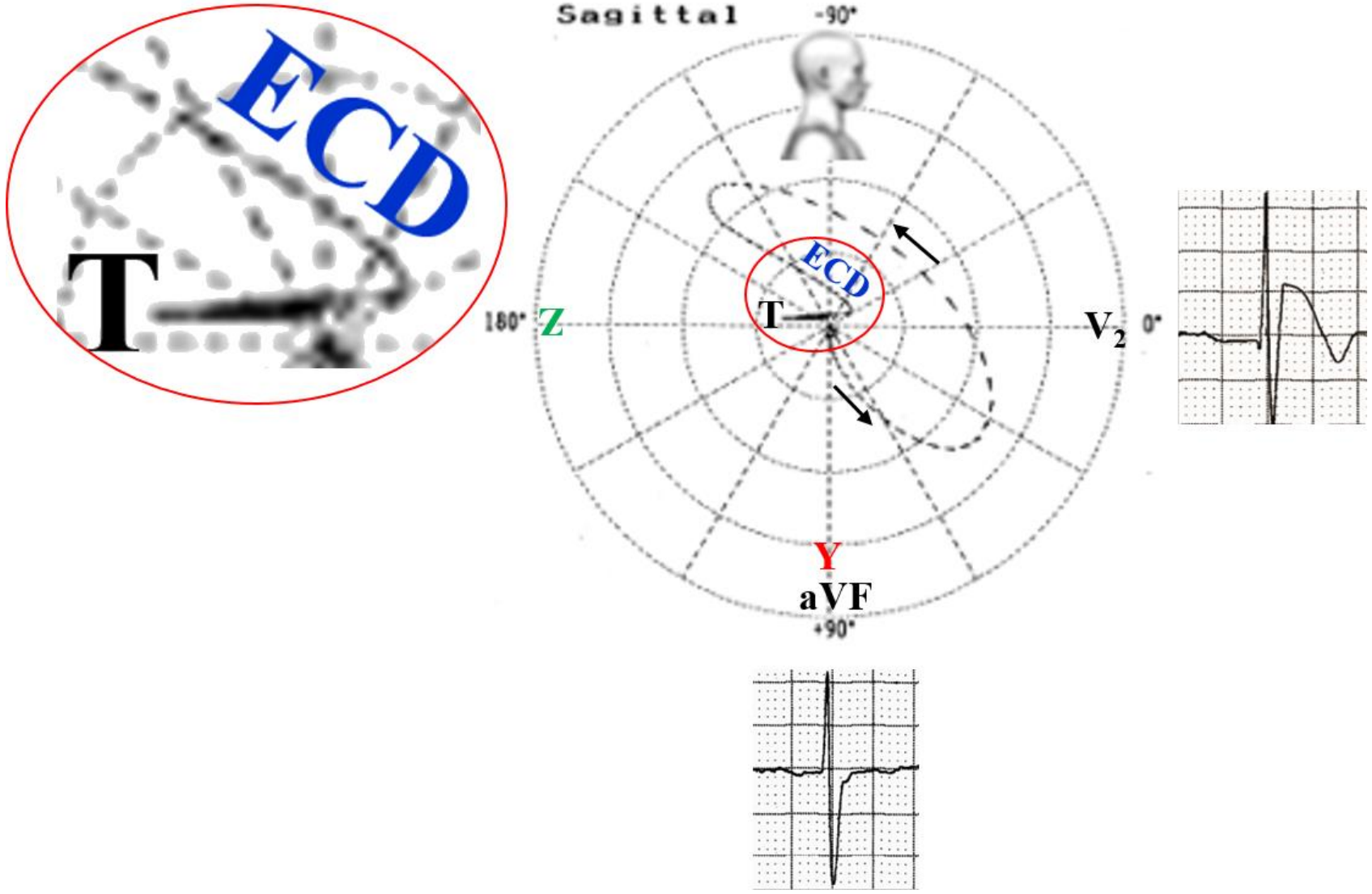
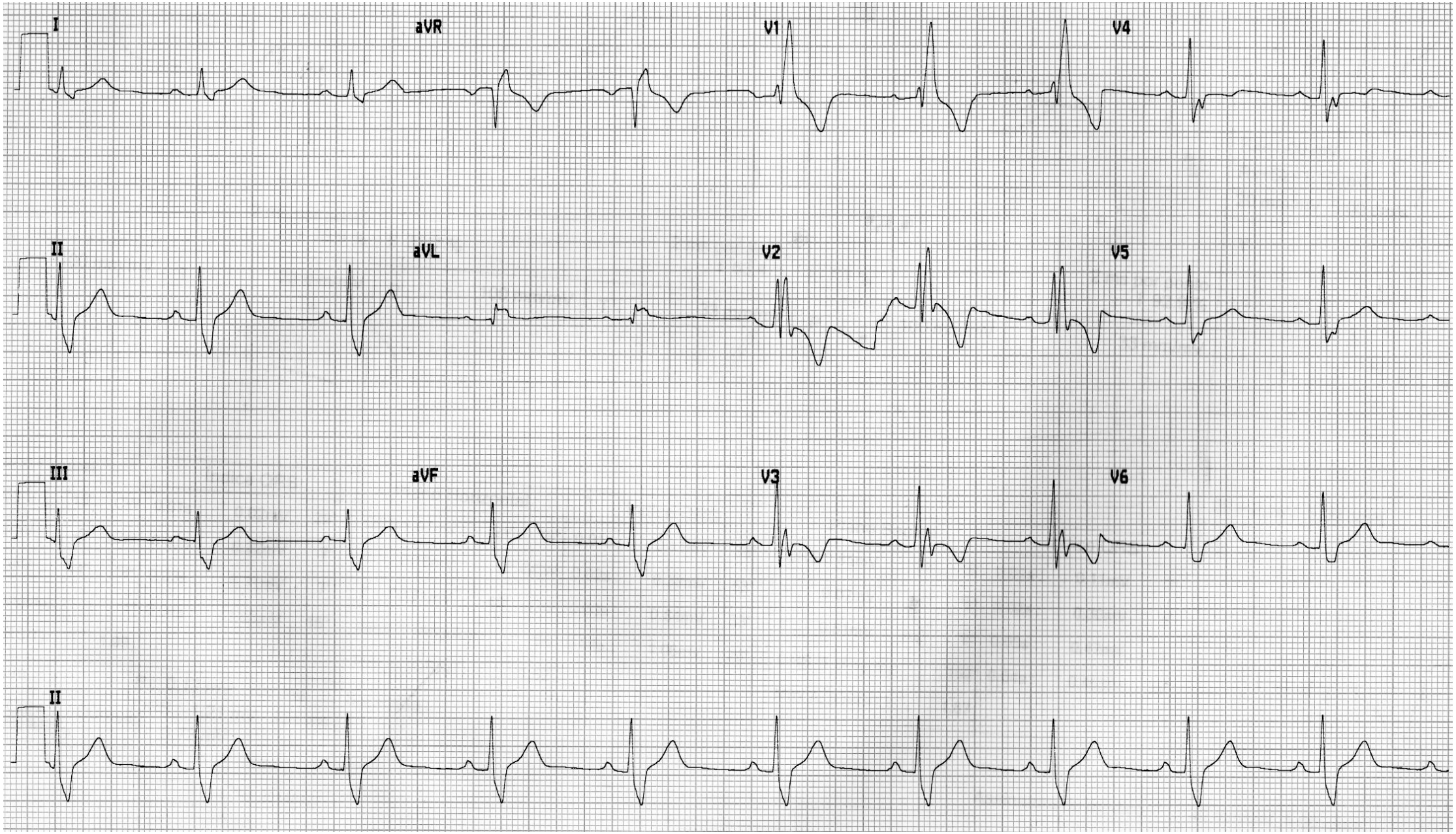


Figure 5 – ECG performed 3 years before



Colleagues opinions

Hi Andrés
Please find here our publication on the subject (1)
18% of Steinert disease have some Brugada pattern on ajmaline
Not a phenocopy because mutations in DMPK causes many alterations in I_{Na+}

1. Maury P, Audoubert M, Cintas P, et al. Prevalence of type 1 Brugada ECG pattern after administration of Class 1C drugs in patients with type 1 myotonic dystrophy: Myotonic dystrophy as a part of the Brugada syndrome. Heart Rhythm. 2014;11(10):1721-7.

Sincerely

Philippe **Maury MD**
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mauryjphil@hotmail.com

Spanish: Hola Potro: en la Distrofia Muscular tipo 1 o Distrofia Muscular de Steinert el gen DMPK afecta los canales de Na^+ motivo por el cuál no es infrecuente la asociación de esta con el síndrome de Brugada. En este paciente el primer ECG muestra el patrón de Brugada tipo 1 espontaneo y un bloqueo atrioventricular de primer grado que enmascara parcialmente el bloqueo de rama derecha que presenta el paciente. Esto es debido a que la enfermedad se asocia con trastornos de la conducción como los observados en el 2do ECG con BRD.
Desconozco que medicación recibe o como están los electrolitos sanguíneos pero no me impresiona como una fenocopia.
Un abrazo
Martín Ibarrola

1. Pambrun T, Mercier A, Chatelier A, et al. Myotonic dystrophy type 1 mimics and exacerbates Brugada phenotype induced by Nav1.5 sodium channel loss-of-function mutation.Heart Rhythm. 2014 Aug;11(8):1393-400.

English: Hello Potro: in DM1 (Steinert's disease) the mutation in DMPK gene affects the Na^+ channels (loss of function) which is the reason it is not uncommon the association of this with Brugada syndrome. In this patient the first ECG shows an spontaneous type 1 Brugada pattern, first-degree atrioventricular block and right bundle branch block. This is because the disease is associated with conduction disturbance as seen in the 2nd ECG with RBBB.
I do not know if he is receiving medication or the level of blood electrolytes but it does not look like as a phenocopy to me.
A hug
Martin **Ibarrola**, M.D. Buenos Aires.

Final comments by

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ECG / VCG Pérez-Riera | my cardiology site of scientific interests:
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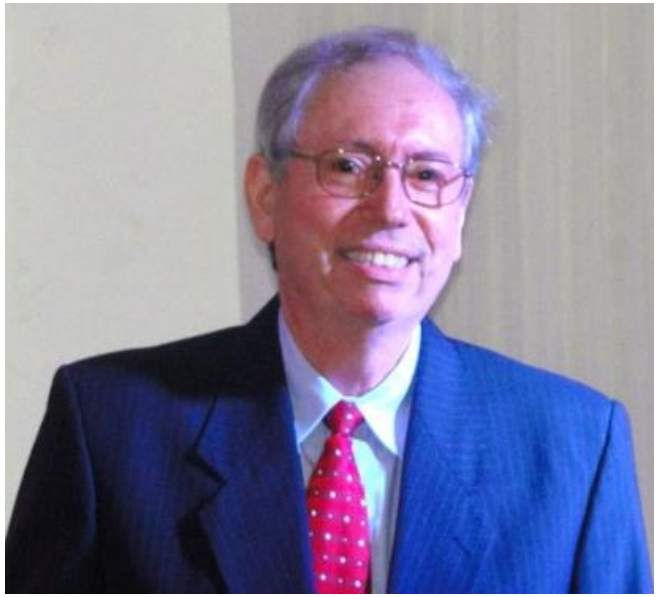


Type 1 Myotonic Dystrophy (DM1) and Brugada Syndrome (BrS) relationship: BrS or Brugada Phenocopy?

Introduction

Myotonic disorders are heterogeneous hereditary multisystem entities with autosomal dominant transmission manifested by a continuum from mild to severe, that have been categorized into three somewhat overlapping phenotypes: I) milder (DM2 < 50 cytosine-thymine-guanine CTGn repeats expansions in 3′ untranslated regions of the dystrophic myotonic protein kinase *(DMPK)* gene); II) classic (DM1) (between 50–2000), and III) congenital DM1 (severe) with early death. The locus is mapped in chromosome 19 (19q13.2 - 19q13.3) (**Junien 1989**).

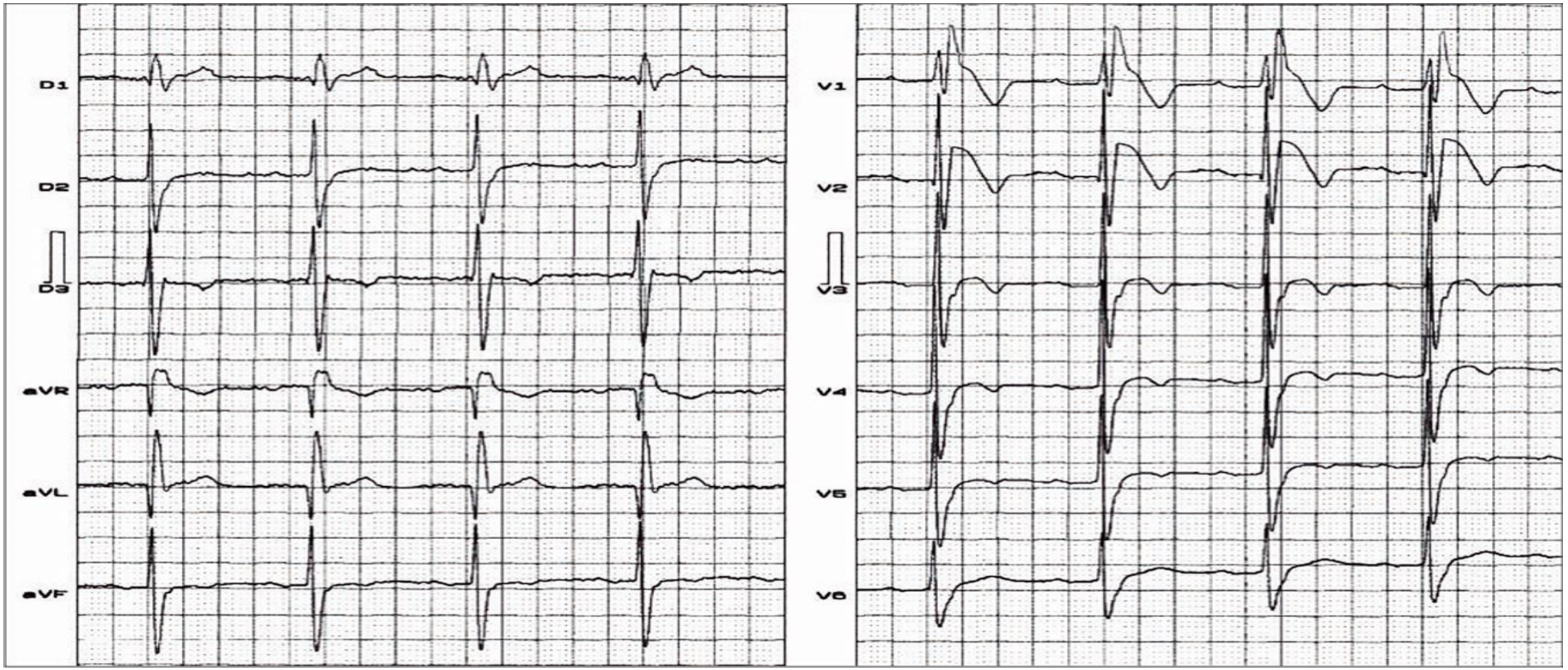
DM1 or Steinert's Disease (**Steinert 1909**) (MIM 160900) is the most frequent neuromuscular disease in adults. The prevalence estimates per 100,000 is 8.26 (**Mah 2016**). DM1 patients have a lower life expectancy (< 60 years), due to cardiac and respiratory complications. The entity affects skeletal muscles with weakness and myotonic phenomena (a failure of muscle relaxation after activation), cardiac and smooth muscles, eyes (posterior iridescent cataract), endocrine system (insulin resistance/diabetes, thyroid dysfunction, hypogonadism/infertility), and central nervous system (intellectual disability, mental retardation, attention disorders).

Cardiac involvement is frequent and characterized by phenotypic heterogeneity. DM1 should be considered in previously undiagnosed patients presenting to a physician with suspected arrhythmia or conduction block, shortness of breath, discomfort, palpitations, syncope and in severe cases, sudden death, observed in ≈10% (2–30%) and caused by third-degree atrioventricular (AV) block, VT/VF or electromechanical dissociation.

The ECG features are: sinus bradycardia, nonsustained supraventricular tachycardia (≈4.1%), atrial fibrillation/atrial flutter, progressive conduction abnormalities; but the rate of progression is not clear such as first-degree AV block (≈25% of cases), second-degree AV block (5.6%), right bundle branch block/left bundle branch block (RBBB/LBBB) (5.5% /3.2%), left anterior fascicular block (LAFB) (**Lund 2014**), QRS duration prolongation ≥ 110 ms in 50% of cases (**Derejko 2015**), prolonged QT interval (≈7.2%), R-on-T premature ventricular contractions (PVCs), and complete atrioventricular (AV) block. Patients with repeats expansion size >1000 CTG had a more rapid progression (**Clarke2001**). This finding is significant because of the risk to develop a complete AV potentially leading to sudden death, positive late potentials (LPs) on signal-averaged electrocardiography (SAECG) in 50% of cases. LPs in DM1 seem to be a consequence of delayed activation along the His-Purkinje system rather than an inhomogeneous conduction through scattered areas of fibrosis. LPs represent a clue for the presence of HV interval prolongation. QRS duration >100 ms and low amplitude signals in the last 40 ms of QRS complex have been shown to predict a prolonged HV interval at the intracardiac electrophysiology study (EPS) with 80% of sensitivity and 83.3% specificity respectively (**Morner 2012**).

Structural heart disease presents with left ventricular systolic dysfunction (≈20.6%) and reduced global longitudinal strain (≈21.7%) consequence of perivascular interstitial fibrosis primarily in the conduction system, fatty infiltration, hypertrophy, myofibrillar disarray, and focal myocarditis

ECG-1

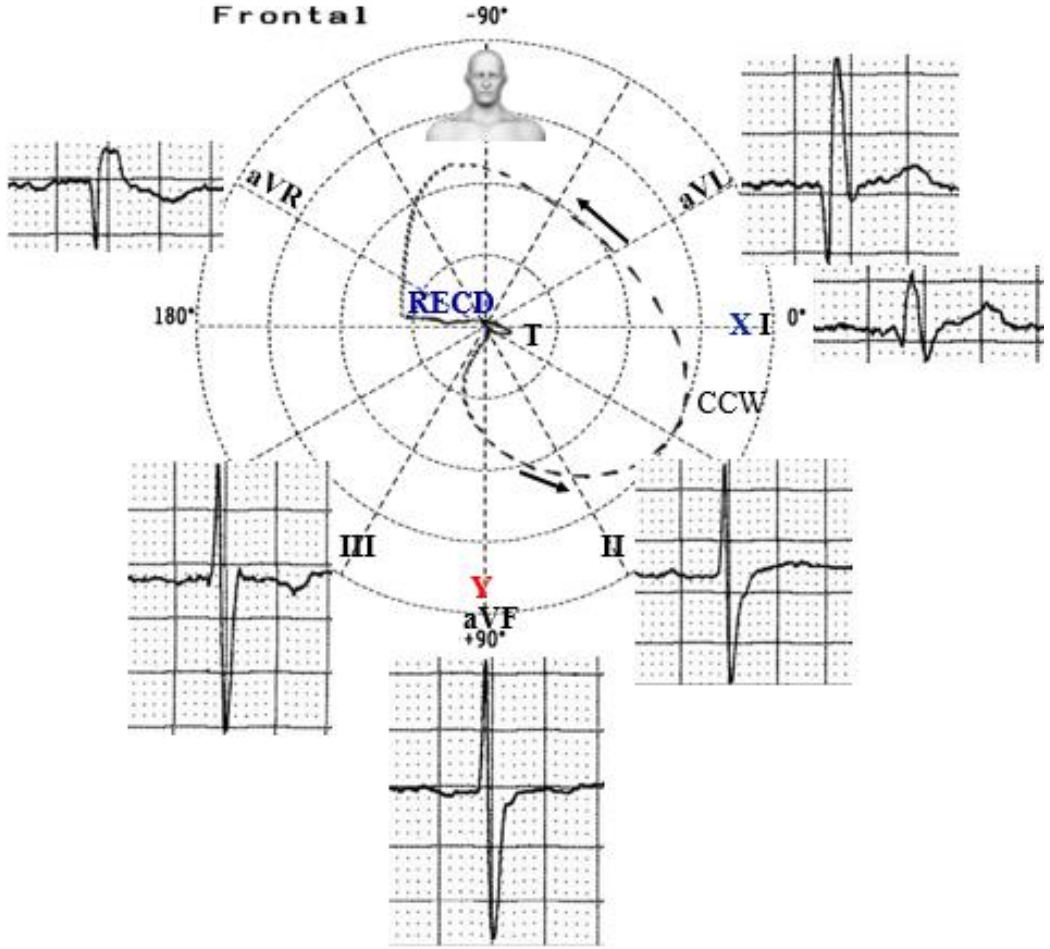


Clinical diagnosis: DM1 (Steinert's disease) / type 2 diabetes mellitus / high blood pressure. BrP: Brugada phenocopy/BrS?

ECG diagnosis: 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 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 T wave in V1-V2.

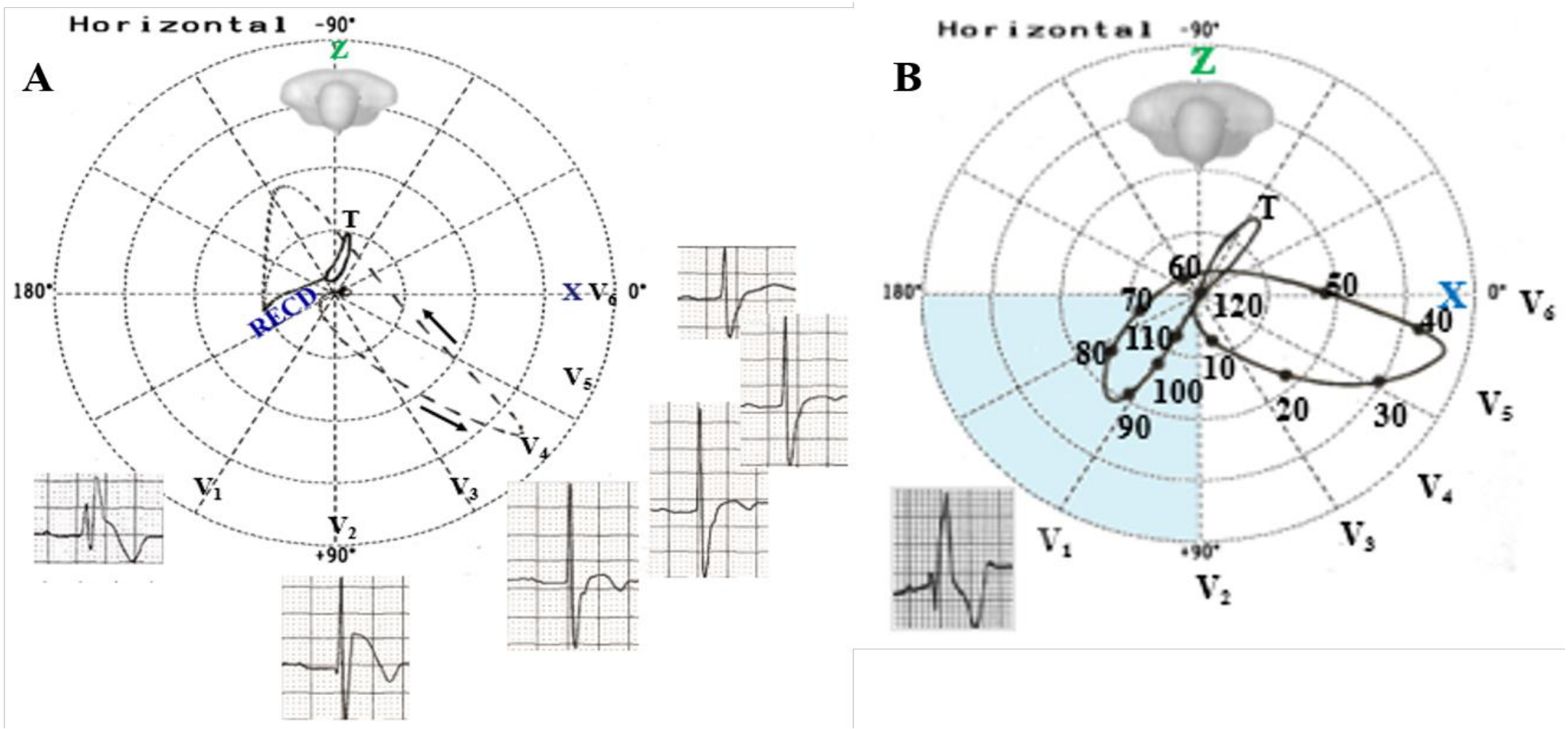
Conclusion: First degree AV block; Complete RBBB (CRBBB); LAFB; Probable trifascicular block (degree of left posterior fascicular block (LPFB)); Type 1 Brugada pattern (BrP).

Electro-vectorcardiogram correlation in the frontal plane



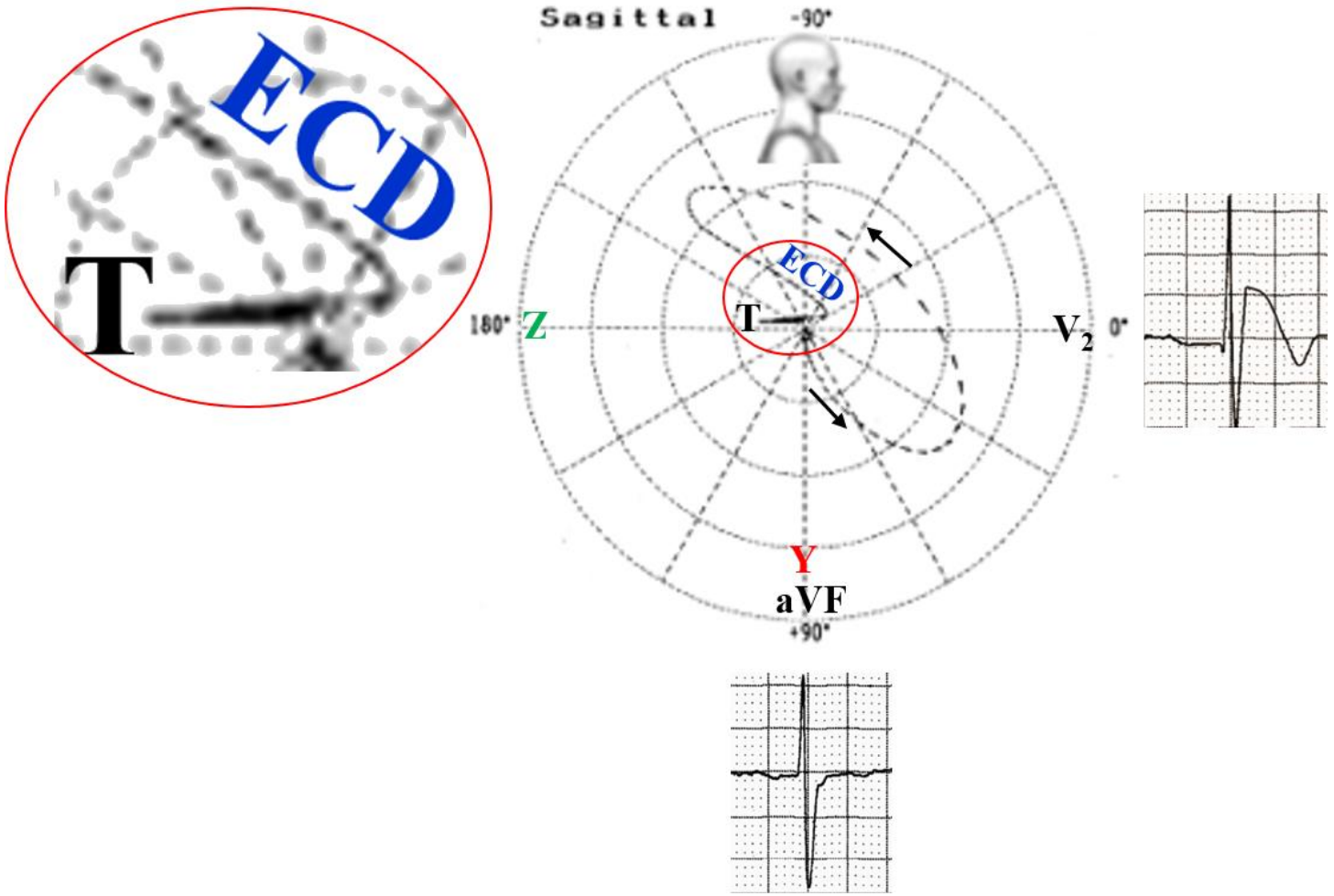
Frontal plane: The QRS axis is difficult to determine as it is almost perpendicular to the QRS loop on the frontal plane. The QRS complex has RS pattern (R=S) in inferior leads. The 10 to 20 ms initial vector of the QRS loop goes down and rightward, with counterclockwise (CCWR) rotation and with the final portion of slow inscription and located at the right on the orthogonal X lead (0°/±180). In the LAFB, the inferior leads show rS pattern. In this case, the voltage of initial R waves in inferior leads is greater (R=S pattern). Additionally, the QRS loop shape is rounded, characteristic of LPFB. We speculate that both, the RS pattern and rounded shape of the QRS loop suggest some degree of associated LPFB. The wide final R wave in aVR and the right end conduction delay (**RECD**) on orthogonal X lead at the right is a consequence of RBBB in association.
Conclusion: LAFB, CRBBB, probable degree of LPFB. Possible trifascicular block.

ECG/VCG correlation in the Horizontal plane



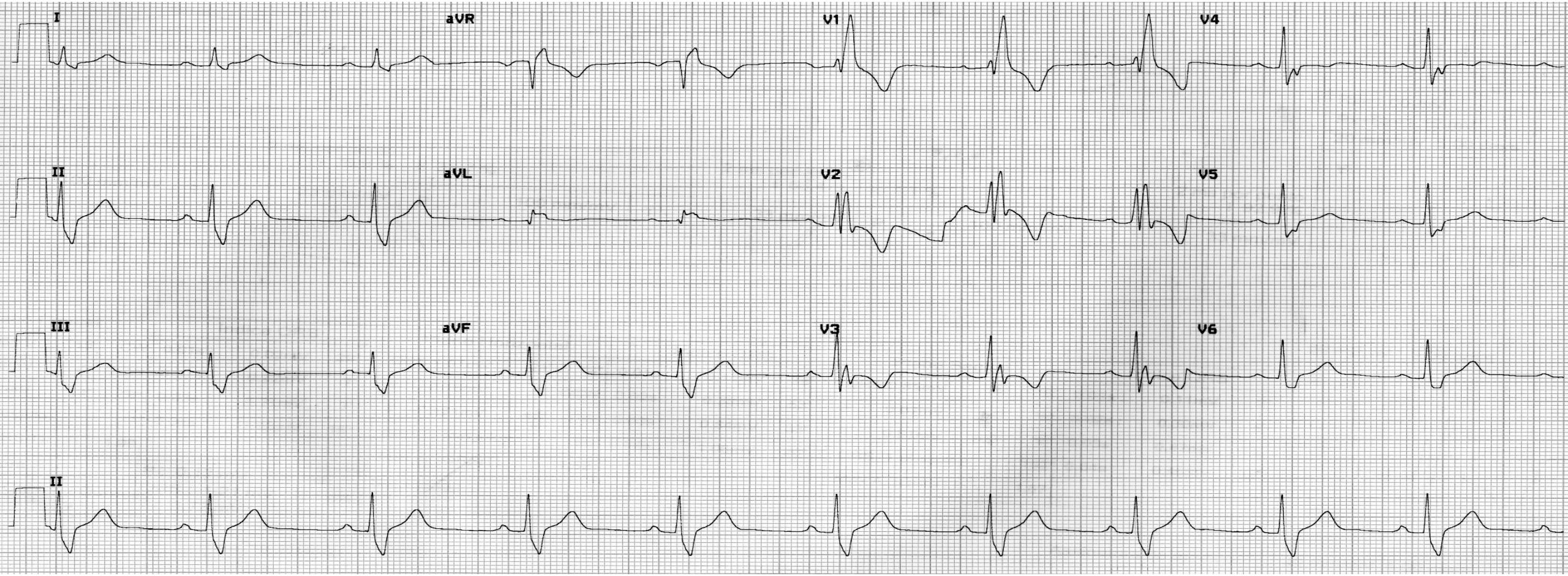
Horizontal or transverse plane: QRS duration 130 ms, in the right precordial leads triphasic pattern RSR' in V1 and broad final S wave in left leads: CRBBB. In V1-V2 coved type, J point and ST segment elevation ≥ 2 mm followed by negative T wave: Type 1 BrP.
The afferent limb is located behind the orthogonal X lead QRS (RBBB Grishman type or Kennedy type 1) and it 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 (RECD).
Conclusion: CRBBB, of the vectorcardiographic Grishman type and type 1 ECG BrP.

ECG/VCG correlation in the Right Sagittal Plane



Right Sagittal Plane: QRS loop with initial vectors directed downward and anteriorly, with counterclockwise rotation, and end conduction delay (**ECD**) in the top quadrants, ending in the anterior superior quadrant. It does not return to the point of origin but continues in a rightward and predominantly anterior direction to inscribe a terminal, “finger-like” appendage. Small T-loop directed to the back. V2 type 1 BrP. aVF RS pattern.

Figure 5 – ECG performed 3 years before



ECG diagnosis: sinus rhythm; HR 59bpm, SÂP: +55º; PR interval: 196 ms; QRS duration 166ms; SÂQRS: –75º: extreme left axis deviation R<S in II, III and aVF; triphasic pattern in V1 type rsR', tetraphasic pattern in V2 type RSR's; QR pattern in aVR lead with wide final R wave followed by negative T wave, wide, slurred final S wave in left precordial leads V5-V6, QT/QTc: 435/435ms. and ventricular repolarization (ST/T) with opposite direction to the terminal deflection of the QRS complex: T wave polarity opposite to the polarity of the last deflection of QRS complex.

Conclusion: CRBBB + LAFB.

Differences between ECG-1 and ECG-5 (performed 3 years before)

	ECG-1	ECG-5
PR interval	250ms. First degree AV block	196bpm. Normal
QRS in inferior leads II, III and aVF	Clear R<S	Near R=S
ST segment in right precordial leads	ST segment elevation ≥2mm followed by negative T wave: spontaneous type 1 BrP	Absent
Probable trifascicular block	Yes	No

The most common pattern referred to as "trifascicular block" is the combination of bifascicular block with first degree AV block.
Trifascicular block can be *incomplete* or *complete*, depending on whether all three left fascicles have completely failed or not.

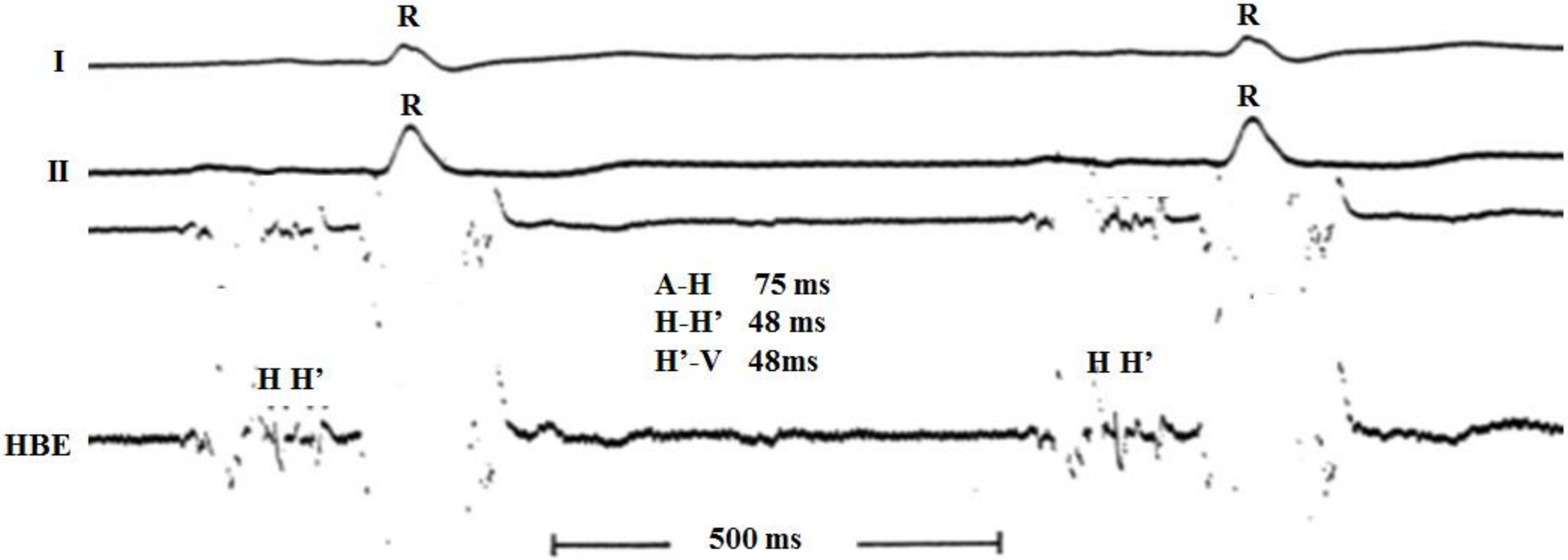
Incomplete trifascicular block
Incomplete ("*impending*") trifascicular block can be inferred from one of following ECG patterns:
Fixed block of two fascicles (i.e. bifascicular block) with evidence of delayed conduction in one of the remaining fascicles (i.e. 1st or 2nd degree AV block).
Fixed block of one fascicle (i.e. RBBB) with intermittent failure of the other three fascicles (i.e. alternating LAFB/LPFB/LSFB).

Complete trifascicular block
Complete trifascicular block produces 3rd degree AV block with features of bifascicular block. This is because the escape rhythm usually arises from the region of either the left anterior, left middle or left posterior fascicle (distal to the site of block), producing QRS complexes with the appearance of RBBB plus either LPFB, LSFB or LAFB respectively.

Discussion

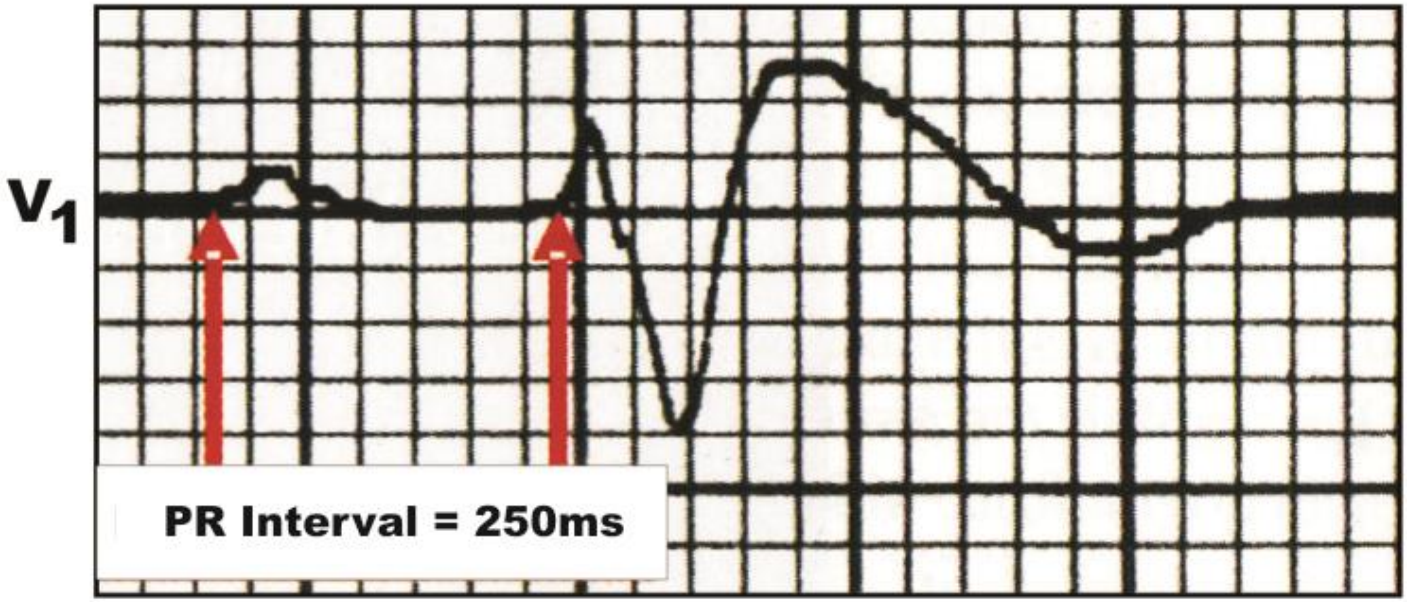
We verified numerous coincidences and curious similarities among DM (DM1 and DM2) and BrS:

- Both have autosomal dominant transmission;
- Both eventually affect the same chromosome 3 (DM2, PROMM variant or proximal myotonic myopathy) (**Rudnik-Schöneborn 2011**);
- Both are considered channelopathies (DM1 with structural heart disease and BrS with apparent without);
- Both have frequent involvement of the intraventricular His system with prolongation of the HV interval verified in the His bundle electrogram (HBE) or split His. In BrS, PR prolongation is consequence of HV split or HV prolongation (**Miyamoto 2011**).



His bundle electrograms show leads I, and two HBEs. P waves are labeled P, QRS labeled R, atrial electrograms labeled A, and ventricular electrograms labeled V. Note the two high frequency electrograms (H and H') between the atrial and ventricular electrograms. Conduction intervals are listed. Paper speed is 200 mm/s.

- Both are predominantly manifest initially in the productive time of life (between 20 and 30 years old);
- Both have frequently atrial arrhythmias mainly atrial fibrillation,
- Both have frequent ventricular arrhythmias;
- Both have a frequent prolongation of the PR interval with HV prolongation or split His;



First degree AV block in a patient with spontaneous type 1 BrP in one symptomatic BrS patient

First-degree AV block is frequently encountered in clinical practice and is generally considered a benign process. However, there is emerging evidence that prolonged PR interval may be associated with adverse outcomes.The figure shows a tracing of a symptomatic patient with BrS after intravenous ajmaline injection. First-degree AV block (PR interval = 216 ms) and Brugada type-1 ECG pattern in V_1 lead (positive test). In BrS the PR interval of ECG and the His bundle electrogram in approximately 50% of the cases are prolonged, even reaching sometimes 100 ms (**Yokokawa 2007**). This prolongation of the PR interval is observed predominantly in cases where the SCN5A gene mutation can be proven (carriers). The presence of a prolonged HV interval is possible in HBE by the existence of intra-His or infra-His block. PR prolongation consequence of HV split or HV prolongation is considered another ECG risk marker (**Miyamoto 2011**). Search yielded 14 studies that were undertaken between 1972 and 2011 with 400,750 participants. Among the studies that adjusted for potential confounders, the pooled results suggest an increased risk of mortality with prolonged PR interval risk. Prolonged PR interval was associated with significant risk of heart failure or LV dysfunction and AF but not cardiovascular mortality, CAD or MI or stroke or TIA. Similar observations were recorded when limited to studies of first-degree AV block (**Kwok 2016**).

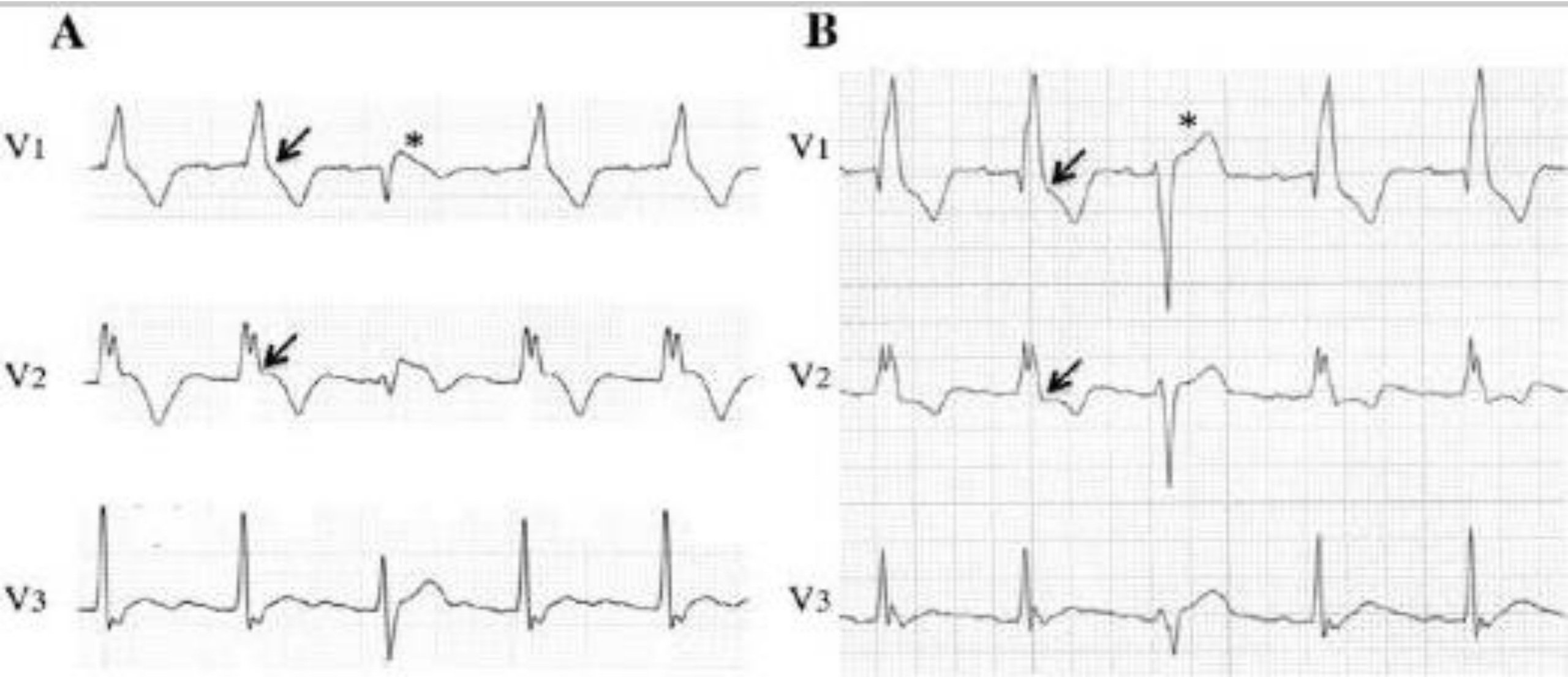
- Both have frequent extreme left axis deviation on ECGs in the frontal plane. Consequence of LAFB or right superior divisional block;
- Both can present eventually RBBB. Maury observed complete RBBB in 28% of cases of BrS from 325 patients with BrS (47 ± 13 years, 258 men) (**Maury 2013**), and 4.4% in dystrophy type 1 (**Petri 2012**);
- Both can present type 1 or 2 ECG BrP (the incidence of type 1 BrP in Steinert's disease is 80 times the incidence in the general population). In DM1, ECG BrP may be related to missplicing of SCN5A (**Wahbi 2013**). Differently from truly phenocopy which is an environmental condition that imitates or copies one produced by a gene. For the diagnosis of Brugada phenocopy is necessary the presence of (**Baranchuk 2012; Anselm 2014; Anselm 2013**):
    1. An ECG pattern that has a type-1 or type-2 BrP;
    2. The presence of an underlying identifiable condition;
    3. The ECG pattern resolves upon resolution of the underlying condition;
    4. There is a low clinical pretest probability of true BrS determined by a lack of symptoms, medical history, and family history; the results of provocative testing with a sodium channel blocker such as ajmaline, flecainide, or procainamide are negative;
    5. Provocative testing is not mandatory if surgical RVOT manipulation has occurred within the last 96 hours (**Rambod 2014**);
    6. The results of genetic testing are negative (desirable but not mandatory because the SCN5A mutation is identifiable in only 20 to 30% of probands affected by true BrS;
    7. Correction of the hypokalemia.

- Both have a high prevalence of late potentials on signal averaged electrocardiogram;
- Both present a tendency to sudden cardiac death; and
- Both present enhanced risk of arrhythmias with anesthetics.

Additionally, DM1 has a high prevalence of ajmaline-induced BrP (**Pambrun 2015**). The modification of the ECG pattern by ajmaline supports the hypothesis of a link between DM1 and Nav1.5 loss of function (**Pambrun 2014**). From a total of 270 DM1 patients followed at the University Centers of Toulouse and Montpellier between 2002 and 2010, were selected 44 patients who fulfilled the following inclusion criteria (**Maury 2014**)

1) Diagnosis of DM1 based on clinical and/or genetic criteria;
2) Presence of such as incomplete RBBB and type 2 or 3 BrP (first consensus about the BrS).

Eight of 44 patients (61 men) with DM1 (18%) presented with type 1 ECG BrP after drug challenge with ajmaline or flecainide (positive test). The positive teste were more often in men than female (26% vs 6%) was observed in younger age (35 vs 48 years) and was not correlated to symptoms, baseline ECG, HV interval duration, results of SAECG or abnormalities on Holter monitoring. DM1 patients with type 1 ECG BrP have longer QTc, greater increase in PR interval after drug challenge, and higher rate of inducible ventricular arrhythmias at EPS (62% vs 21%, P ¼ .03). DM2 may show BrP with and without missense mutation in the SCN5A gene (**Rudnik-Schoneborn 2010**). Type 2 myotonic dystrophy may behave as a Brugada phenocopy. Additionally, a high prevalence of ajmaline-induced BrP was observed in DM1 patients (**Pambrun 2014**). The modification of the ECG pattern by ajmaline supports the hypothesis of a link between DM1 and Nav1.5 loss of function. The indications, the safety, and the implications of ajmaline challenge in this particular setting need to be determined by larger prospective studies (**Pambrun 2015**). DM1 has a deleterious effect on clinical expression of a loss-of-function SCN5A mutation and a provoked BrS phenotype may show in a DM1 patient (**Phillips 1997**). Electron microscopy shows prominent I-bands, abnormal mitochondria, and myofibrillar degeneration. Myotonin protein kinase, the primary product of the myotonic dystrophy gene, may be located at the intercalated discs and has a different isoform in cardiac tissue. Wahbi et al studied 500 cases of DM1 and found that the incidence of BrP is 80 times the incidence in the general population (**Wahbi 2009**). The mechanism by which DM1 may present as a Brugada phenocopy is unclear and requires further investigation. Ruling out BrP in DM1 patients seems important because of the use of some Class 1 drugs— especially mexiletine—for decreasing myotonia in DM1 (**Logigian 2010; Fazzio 1998**). Except for quinidine, most Class 1 drugs are on the list of medications to be avoided in Brugada patients, whereas mexiletine is a Class 1B antiarrhythmic drug close to lidocaine, which is a drug preferably avoided in BrS (http://www.brugadadrugs.org). Down-regulation of DMPK mRNA represents a potential, and as yet unexplored, DM1 therapeutic avenue. Consequently, a computational screen for agents which down-regulate DMPK mRNA was undertaken, unexpectedly identifying the sodium channel blockers mexiletine, prilocaine, procainamide, and sparteine as effective suppressors of DMPK mRNA. Analysis of DMPK mRNA in C2C12 myoblasts following treatment with these agents revealed a reduction in the mRNA levels. In vivo analysis of CD1 mice also showed DMPK mRNA and protein down-regulation (**Witherspoon 2015**).
From 914 patients included in the DM1 Heart Registry by Wahbi et al (**Wahbi 2013**) spontaneous type 1 BrP were identified on the 12-lead ECGs of seven patients with DM1 (0.8%) or 7.7/1000. While this prevalence is low compared with other disease manifestations of DM1, it is nearly 50-fold higher than the 0.17/1000 prevalence observed in 12,012 apparently healthy European subjects (**Gallagher 2008**). The authors speculate that the prevalence might be underestimated, because major intraventricular conduction disorders or paced QRS complexes might have concealed the type 1 BrP (**Chiale 2012**). From 7 cases of type 1 BrP of Wahbi series, none were associated with CRBBB, unlike the present case. The CRBBB can hide the type 1 BrP. See next slide.



The 12-lead ECG with spontaneous normalization of the CRBBB.
A, The ECG shows a CRBBB in the first and last 2 beats. The third beat shows a loss of CRBBB and the normalized QRS complex (asterisk), and in this beat, the type 1 BrP manifests. The J point is slightly lowered in V_1 and elevated in V_2 including the initial part of the ST segment (arrows). B, Spontaneous resolution of the CRBBB during the12-lead Holter recording (asterisk). The normalized QRS complex was associated with a slightly shorter RR and PR interval, and an ST-segment elevation in V_1 and V_2 was evident. The QRS with CRBBB showed distinct downward displacement of the J point and the ST segment (arrows).

Additionally, class I antiarrhythmic tests with ajmaline were interrupted in DM1 patients by the occurrence of a obit in consequence of electrical storm episode after the procedure (**Otten 2009**).

Conclusions

The ECG and VCG of a DM1 patient with type 1 BrP pattern is shown for the first time in the literature here. The numerous and curious coincidences between both entities should make us reflect on the possibility of a link between the two channelopaties probably causing a loss-of-sodium channel function with or without mutation in the SCN5A gene. The high prevalence of type 1 BrP in DM1 patients may generate the hypothesis that abnormal SCN5A splicing could be responsible for this finding.

The identification of a type 1 BrP in DM1 patients should be considered as a major risk factor. Patients for whom a pacemaker is indicated for severe progressive intraventricular conduction disturbance could benefit from a implantable cardioverter defibrillator rather than a pacemaker. This fairly high risk for ventricular arrhythmias may have several explanations, such as: genetic background specific to patients with DM1; association of abnormal splicing of other ion channels involved in myocardial depolarization or repolarization; and specific pattern of Nav1.5 isoforms expression differing from that observed in patients with SCN5A gene mutations (**Wahbi 2013**).

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