

## *The epsilon waves clinical characterization and electrophysiological significance*

### **Introduction**

In approximately 30% of the most severe cases of arrhythmogenic right ventricular cardiomyopathy /dysplasia (ARVC/D), a deflection or small wiggle may be observed in the electrocardiogram (ECG), in most of cases located after the J point at the beginning of the ST segment. They are delayed potentials which appear after the end of ventricular depolarization (recorded after the end of the QRS complex). or post-excitation phenomenon that may be demonstrated by epicardial mapping, intracavitary electrodes, ECG, and SAECG (**Fontaine 1978 Frank 1978**). It was called for the first time by a French pioneer in Electrophysiology Dr Guy Fontaine (**Marcus 1998**) with the Greek letter epsilon ( $\epsilon$ ) epsilon waves.

**Others denominations:** epsilon potentials (**Peters 2007**), ventricular post-excitation waves (**Maia 1991**), post excitation (epsilon) waves (**Okano 1995**) or with the eponymous Fontaine wave (**Fontaine 1977**),

It is a late depolarization of right ventricular fibers of right ventricular free wall (dysplasic triangle) registred mainly in V<sub>1</sub>-V<sub>4</sub> leads, these oscillations are best seen in the ST segments of leads V<sub>1</sub> and V<sub>2</sub> different from J wave seen in V<sub>5</sub>, V<sub>6</sub> and inferior leads which origin is not so clear.

Epsilon waves are not the direct counterpart of late potentials, but reflect the delay peripheral activation in the right ventricular free wall therefore appear to be responsible for much of the genesis of negative T waves (**Okano 1995**).

**Semantic discussion:** The reason that led this author to choose epsilon name is not clear enough. Could it be because its shape reminded him of the Greek letter Epsilon ( $\epsilon$ ) as suggested by Surawicz e Knilans in their classical book on electrocardiography (**Surawicz B, Knilans TK. Chou's Electrocardiography in clinical practice. Adult and pediatric. Fifth Edition. 2001. Chapter 12. pp 263. WB Saunders Company.**) If this was the case, it should be stated that the epsilon-like wave is in a horizontal position: The tracing shows in the location of the J point and the beginning of the STsegment, an indentation that reminds of the greek letter epsilon, however, in a horizontal position. Dr Fontaine could be considered following the Greek alphabet sequence?:  $\alpha$ ;  $\beta$ ;  $\delta$  and  $\epsilon$ ?

If the additional wave observed in ventricular pre-excitation is located at the beginning of QRS complex is called delta wave ( $\delta$ ), the following additional wave in the Greek enumeration should be called with the following letter: epsilon  $\epsilon$ . Faced with this doubt, I decided to ask the author of this nomenclature, Dr. Fontaine himself, who replied to me thus: "*Dear Dr. Pérez-Riera, Thanks for your documents. The naming of the ECG waves and the reason of their choice is a long story. Dr. Willis Hurst (Hurst 1998) in Circulation has published a summary of these some years ago. I have strongly contributed to this paper as indicated by Dr. Hurst.. Best regards.*"

Dr. Hurst wrote: "*Fontaine discovered and named the epsilon waves. He chose the epsilon because it follows delta in the Greek alphabet and is the mathematical symbol for smallness.*" The term "epsilon" was nice, because it occurs in the Greek alphabet after delta; thus, delta represents the preexcitation and epsilon the post-

excitation phenomenon. In addition, epsilon is also used in mathematics to express a very small phenomenon.

It was quite exciting to demonstrate that these late potentials (LPs) located on the free wall of the RV of patients with ARVC/D could be recorded on the surface by SAECG and in some circumstances by increasing the magnification of ECG recording.

**Figure 4** below, shows this concept.

To conclude, even with the great respect I feel for Dr. Boris Surawics and Dr. Timothy Knilans, I have to comment that they made a mistake by thinking that the reason of the name was morphological and not the sequence of the Greek alphabet.

#### I. *Other denominations (synonymous)*

Epsilon potentials (**Peters 2007**), ventricular post-excitation waves (**Maia 1991**), post excitation (Epsilon) waves (**Okano 1995**) and with the eponymous Fontaine wave (**Fontaine 1977**),

#### II. *Origin of the name epsilon*

“Fontaine discovered and named the epsilon waves. He chose the epsilon because it follows delta in the Greek alphabet and is the mathematical symbol for smallness” (**Hurst 1998**).

#### III. *Definition*

A. **Classical concept:** Epsilon waves have been defined as any potential manifested as a distinct waves of post-excitation with small squiggles, small notches or oscillations (**Khaji 2013**) amplitude that occupy mainly the beginning of the ST segment after the end of the QRS complex (J point) (**Wang 2010**) in other words after the depolarization between the end of the QRS complex and the beginning of the ST segment Epsilon waves are caused by post excitation of the myocytes in the right ventricle free wall due to myocardial scaring. On ECG, they are small notches, oscillations, wiggles, or smoth potential waves in variable quantities (one single deflection, 2, 3 or more). The Epsilon wave was defined as wiggler, small spike wave and smooth potential located between the end of the QRS complex and the beginning of the ST segment. (**Wang 2009; 2010**):

1. Small spike waves: The most common type. They are divided into 2 subtypes, upward and downward.
2. Wiggle waves
3. Smooth potential waves

Epsilon waves are late potentials (LPs) that occur in the RV free wall in patients with ARVC/D and rarely in others physiological and pathological scenarios. As LPs were supposed to be the result of late activation of a limited group of fibers, the term “post-excitation” looked logical, since it was observed after the main excitation of the ventricle, leading to the QRS complex.

The term “epsilon” was nice, because it occurs in the Greek alphabet after delta; thus, delta represents the pre-excitation and epsilon the post-excitation phenomenon.

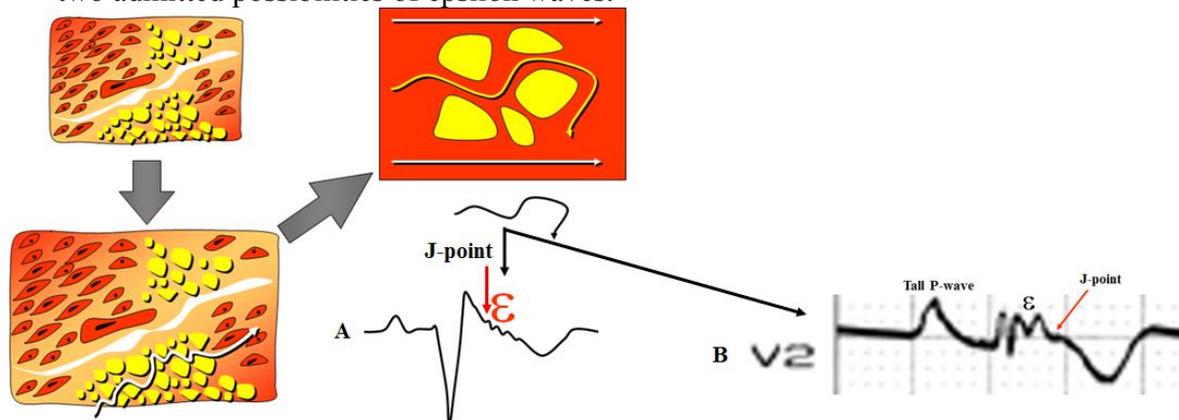
In addition, epsilon is also used in mathematics to express a very small phenomenon.

The proximity of the right ventricle (RV) to the anterior precordial leads V1 to V4 explains why the characteristic ECG abnormalities are most prominent in those leads.

The following ECG shows epsilon waves with 1 (single deflection), 2, 3 or multiple waves.

B. **New concept:** in many cases the definition of epsilon waves/ epsilon potentials remains difficult because some authors consider that these waves may be inside of the QRS complex, manifested as QRS fragmentation or QRS notching (**Hoffmayer 2013**). In ARVD/C fragmented QRS (fQRS) has a high diagnostic value similar to epsilon potentials by a highly amplified and modified recording techniques, such as right precordial leads ECG (R-ECG) and Fontaine leads (F-ECG) (**Peters 2008**). fQRS refers to the ‘slurs or notches’ appeared on the R or S wave or if the total QRS complex had  $\geq 4$  spikes. fQRS can be registered as a normal variant mainly in seniors endurance athlete heart if it appeared randomly in just a few leads. fQRS presenting in multiple leads is more likely pathologic. The underlying cause is the regional delay in propagation of ventricular depolarization (**Monta 2008**). fQRS is highly prevalent in ARVC/D patients when applied to amplified and modified ECG recording techniques, including the use of the Fontaine Leads System (**Peters 2008; Hurst 1998**). In real world practice, nevertheless, most ECGs available from ARVC/D patients and family members were obtained by using only the standard ECG recording technique. fQRS is easily recognizable from standard ECGs (S-ECG) and they are much more common in ARVC/D patient when compared with control subjects. Among them a notch before the end of R or S wave is characteristic, seen in 51% of ARVC/D vs 26% in controls. In ARVC/D, fQRS is often seen in multiple leads (**Zhang 2014**). Such changes, however, are common in control subjects as well. In the latter, the QRS complex is wider (**Dechering 2013**). fQRS complex, with various morphology, has been described as a diagnostic criterion of ARVC/D. Since fQRS is also prevalent in other types of cardiomyopathies (both ischemic and non-ischemic) (**Das 206;2010**).

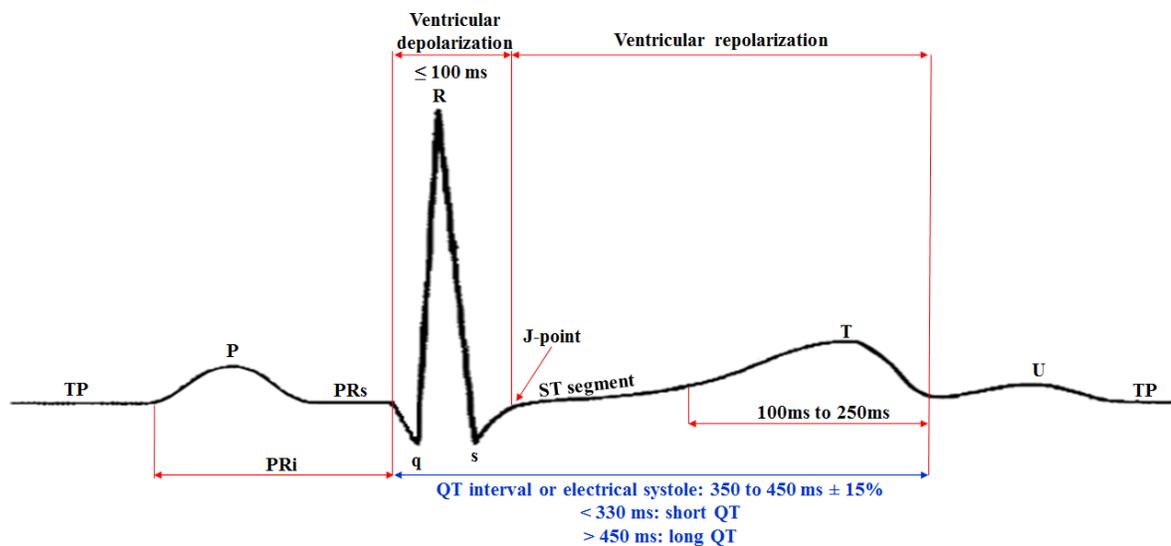
➤ fQRS is induced by radiotherapy in patients with breast cancer (**Adar 2015**), and in normal subjects, its use in ARVC/D diagnosis is limited. The figure shows the two admitted possibilities of epsilon waves.



- A. Oscillations registered after J-point at the beginning of ST segment.
- B. Oscillations registered inside the QRS complex. In this case, epsilon waves are indistinguishable of fQRS.

Evidence of slow fractionated conduction is present as epsilon waves. The signal averaged ECG may show exceedingly long and low late potentials (Marcus 2000). Tanawuttiwat et al (Tanawuttiwat 2016) studying 30 ARVC/D patients underwent endo and epicardial electroanatomical activation mapping in sinus rhythm. The ECGs were classified into 5 patterns: 1. Normal QRS (11 patients); 2. Terminal activation delay (TAD) (3 patients); 3. Incomplete right bundle branch block (IRBBB) (5 patients); 4. Epsilon wave (5 patients); 5. Complete RBBB (CRBBB) (6 patients). Timing of local ventricular activation and extent of scar was then correlated with surface QRS. Earliest endocardial and epicardial RV activation occurred on the mid anteroseptal wall in all patients despite CRBBB pattern on ECG. Total RV activation times increased from normal QRS to prolonged TAD, IRBBB, epsilon wave, and CRBBB, respectively ( $103.9 \pm 5.6$ ,  $116.3 \pm 6.5$ ,  $117.8 \pm 2.7$ ,  $146.4 \pm 16.3$ , and  $154.3 \pm 6.3$ , respectively,  $P < 0.05$ ). Total epicardial scar area ( $\text{cm}^2$ ) was similar among the different ECG patterns. Median endocardial scar burden was significantly higher in patients with epsilon waves even compared with patients with CRBBB ( $34.3$  vs.  $11.3 \text{ cm}^2$ ,  $P < 0.01$ ). Timing of epsilon wave corresponded to activation of the subtricuspid region in all patients. Epsilon waves are often associated with severe conduction delay and extensive endocardial scarring in addition to epicardial disease. The timing of epsilon waves on surface ECG correlated with electrical activation of the sub-tricuspid region.

If we considered that epsilon waves are located after the J-point at the beginning of ST segment only, the phenomenon theoretically could not be a depolarization criterion because ST segment occur during the repolarization. The following figure explains depolarization and repolarization intervals on ECG.



**QRS complex:** Set of deflections that represent ventricular depolarization

**J-point:** Approximate point of convergence between the end of QRS complex and the onset of ST segment. It is considered the point at which the QRS complex finishes and

the ST segment begins. The J-point is an essential landmark for measuring QRS duration and ST segment elevation and/or depression. J-point represents approximate the end of depolarization and the beginning of repolarization as determined by the surface ECG. There is an overlap of  $\approx 10$  milliseconds (Mirvis 1982). In the classical concept, epsilon waves are located after this point. In the embracing concept, the epsilon waves could be inside the QRS complex, consequently have the same meaning as fragmented QRS (fQRS). Okano et al (Okano 1995). observed that there is no difference in electrophysiologic findings in patients with or without epsilon waves. Negative potentials are present on the anterior chest in ST-T isopotential, ST-T and QRST isointegral maps in all of the patients. The area of these negative potentials was closely correlated with RV dilatation and dysfunction. It is concluded that Epsilon waves are not the direct counterpart of delayed potentials, but the reflection of the peripheral conduction delay, and that primary change seems to play large part of the genesis in negative T waves of ARVC/D

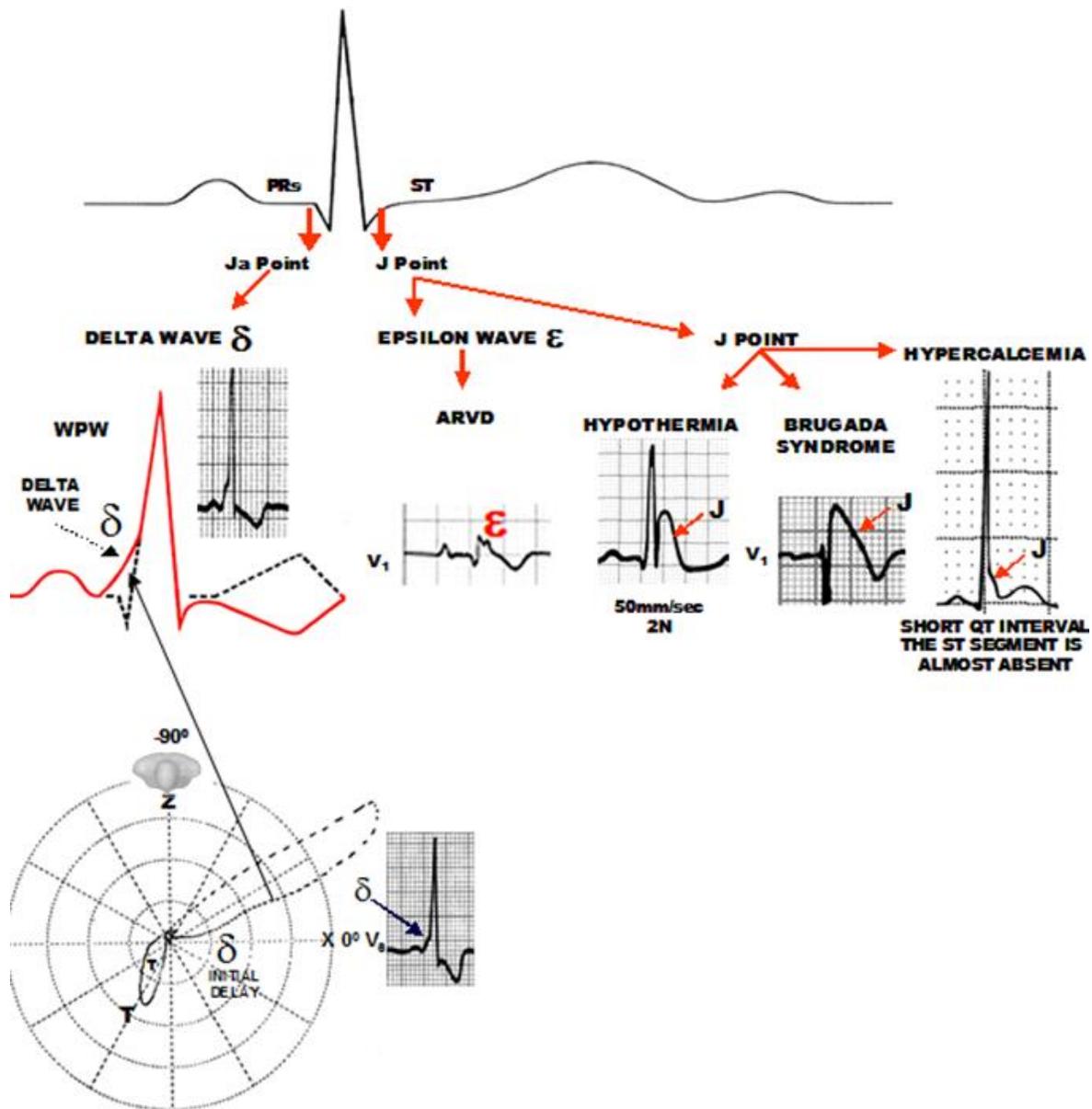
**ST segment:** it stretches from the from the J point (union of ST with the end of QRS complex) until the onset of the T wave, which is usually hard to determine. In electrocardiography, the ST segment connects the QRS complex and the T wave and has a duration of 80 to 120 ms. The ST segment corresponds to phase 2 of action potential (AP).

**T wave:** Normal profile of T wave with slow ascending ramp and faster descending ramp. It is coinciding with phase 3 of AP. T duration is 100ms to 250ms (up to five times more than ventricular depolarization).

**QT interval or electric systole:** interval between the first recognizable part of QRS up to the final recognizable area of the T wave (the latter may be hard to determine precisely). The end of T is defined as the return of the T wave to the T-P baseline.

**U wave:** Last, inconstant and smallest deflection of ECG that is recorded immediately after T wave and before the P of the following cycle, of equal polarity to the preceding T, i.e. positive where T also is. Voltage of U is always lower than 50% of the width of the preceding T and generally between 5% and 25% of it. Usually it does not exceed 1 mm, being in average of 0.33 mm. If it reaches 1.5 mm or more, it is considered high, however, there may be normal U waves of up to 2 mm (0.2 mV) in II and from V2 to V4.

The figure below shows a comparative location and aspect of delta ( $\delta$ ), epsilon ( $\epsilon$ ), and J waves.

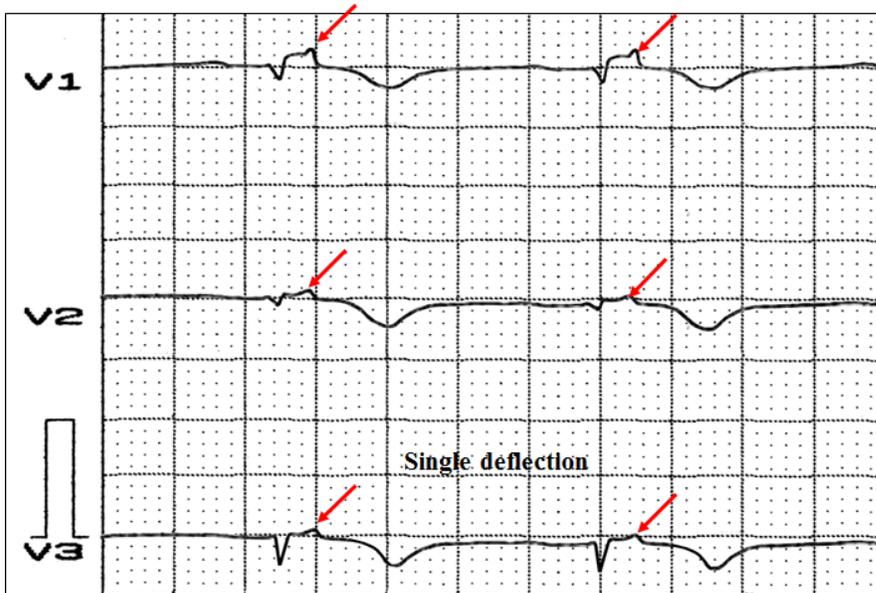


**Observation:** In WPW type ventricular preexcitation, a wave located at the Ja point (end of PR segment and onset of QRS complex) is observed, called delta wave ( $\delta$ ). Following the Greek alphabet, the wave should be called Epsilon ( $\epsilon$ ), located near the J point (end of QRS complex and onset of ST segment).

#### IV. Classification of epsilon wave by the number of deflections

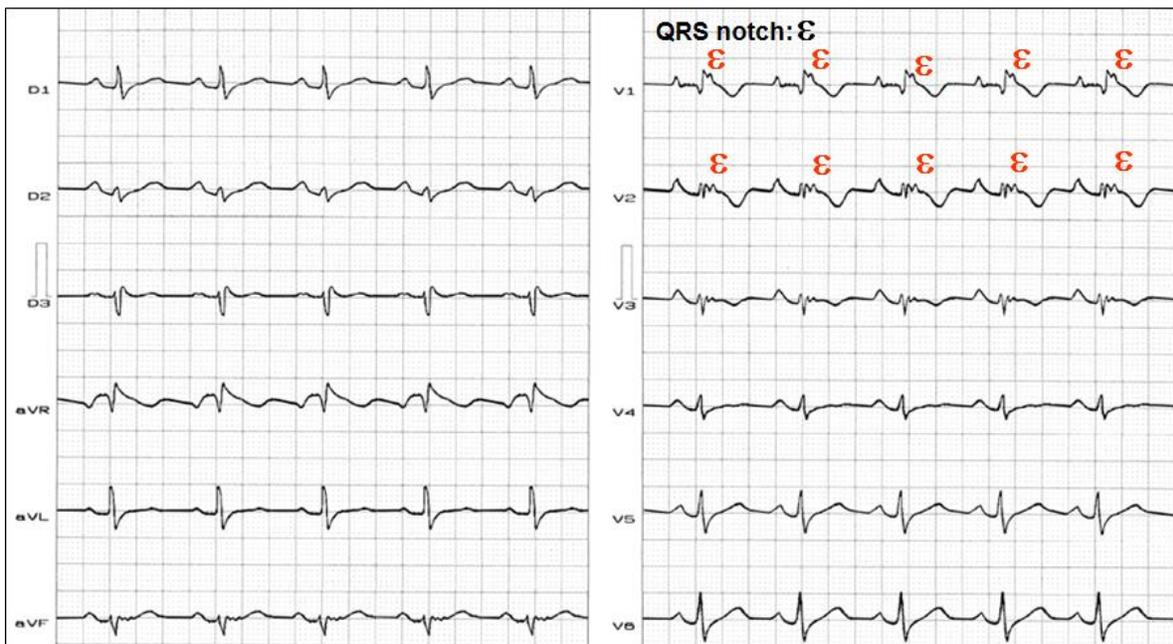
We classified the epsilon waves according to the number of deflections: one, two or multiple deflections.

##### Example of epsilon wave with single deflection



We observe prominent upright deflections (red arrows) after the QRS complex in right precordial leads V1–V3 associated with negative T waves. Epsilon waves are one of the major depolarization diagnostic criteria of ARVC/D following task force. Epsilon waves can be recorded using 12 lead ECG during sinus rhythm, and are useful for establishing a diagnosis of ARVC/D (**Anan 2002**).

**Example of epsilon wave with two deflections located inside of QRS (pre-, top-): fragmented QRS?**



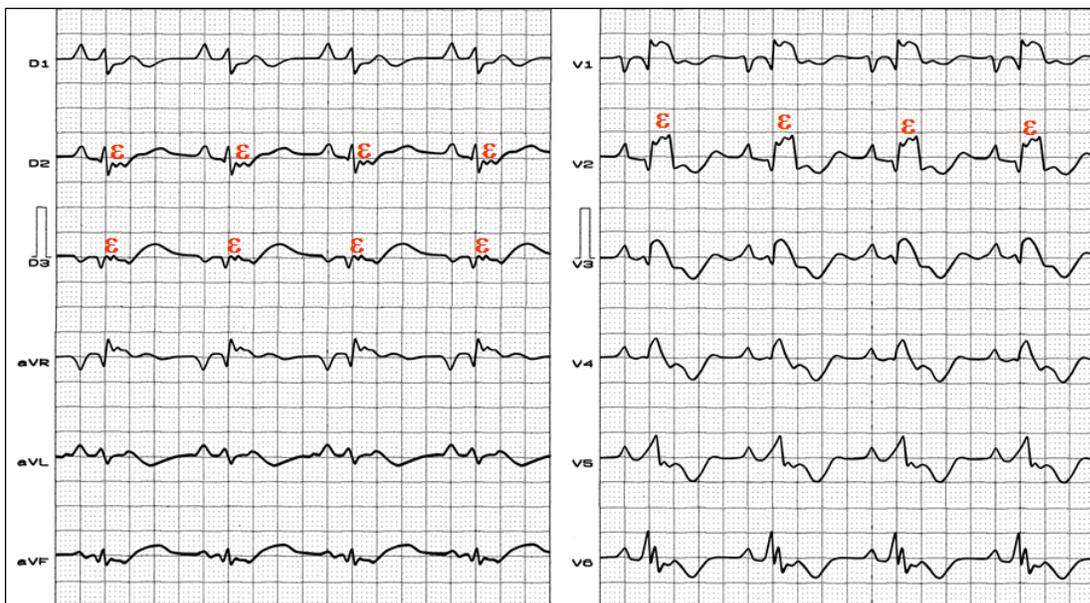
Sinus rhythm, right atrial enlargement, bizarre complete RBBB, terminal notch located in the J point (epsilon wave). The epsilon wave could be the result of delayed activation in the

RV. It is visible from V1 to V3 and in the frontal plane leads. T wave inversion is observed in V1 to V3, characteristic of ARVC/D.

### Example of epsilon wave with multiple deflections inside of QRS complex



**Name:** SFD; **Sex:** F; **Age:** 18 y/o; **Race:** Caucasian; **Weight:** 53 Kg; **Height:** 1.52 m;  
**Biotype:** Normal; **Date:** 05/03/2006

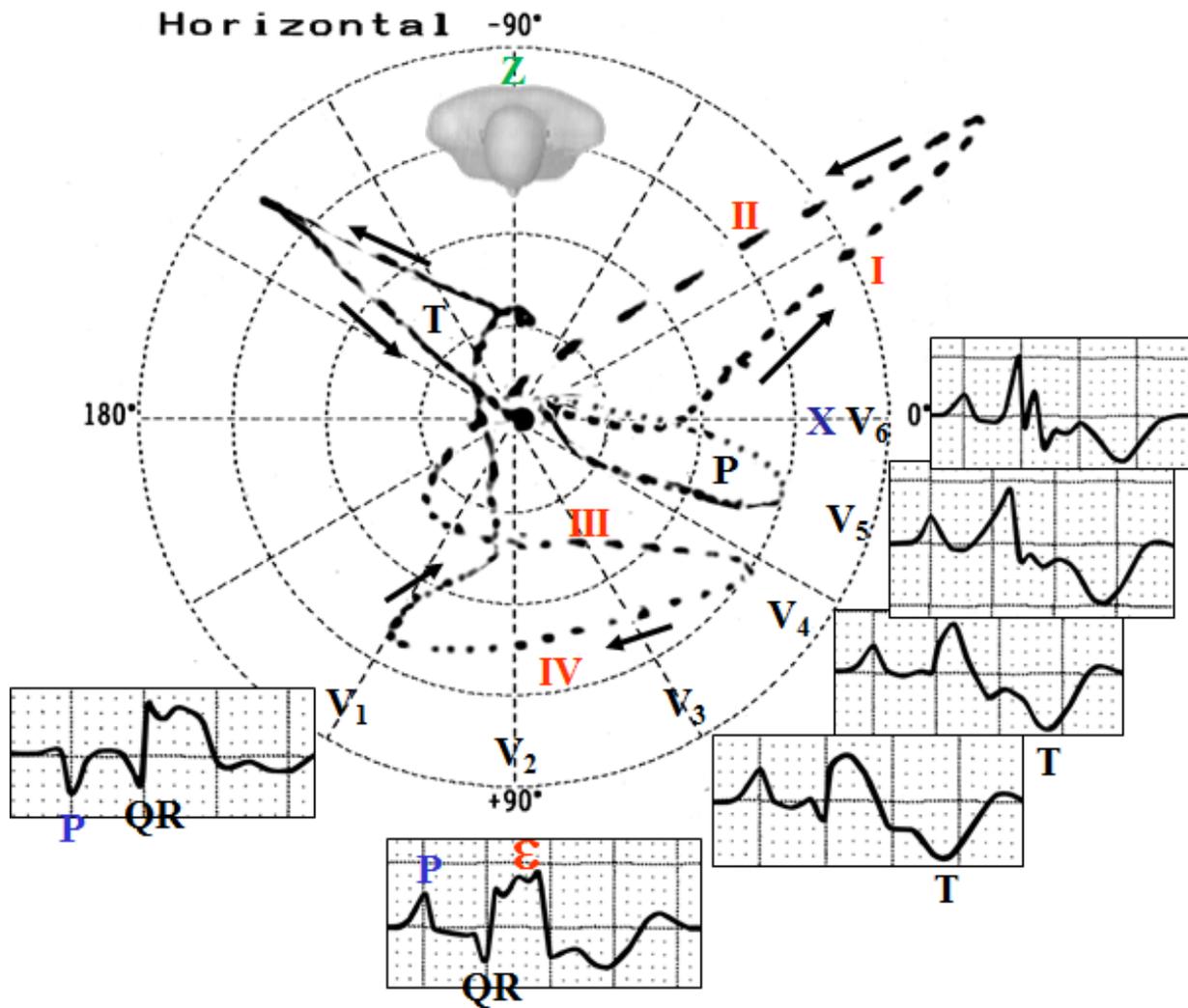


**Clinical diagnosis:** Arrhythmogenic Right Ventricular Cardiomyopathy/Dysplasia. Severe right heart failure.

**ECG diagnosis:** sinus rhythm, HR: 60 bpm; P wave: SAQRS near 0°, voltage: 3 mm, duration: 130 ms: negative polarity in V1 and positive in V2, q wave in V1 and V2: biatrial

enlargement? Or right ventricular mega enlargement? QRSd: 230 ms (CRBBB); epsilon waves are observed in numerous leads inside and outside of the QRS.

**ECG/VCG correlation in the horizontal and frontal planes**



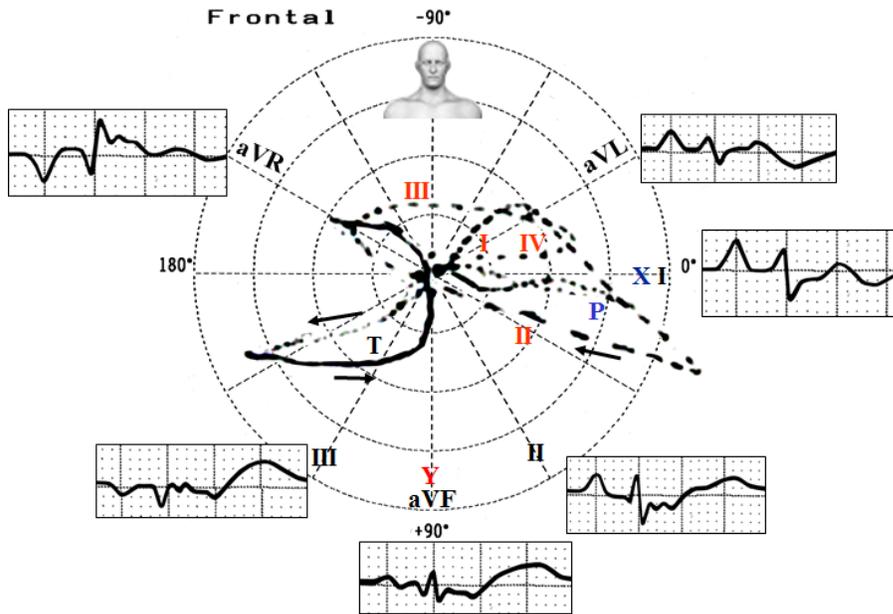
**Biatrial enlargement/ Giant P loop. P axis + 15°.**

**QRS loops:**

**I** – Efferent limb;

**II** – Afferent limb;

**III, IV** – Late QRS forces appendix located in anterior quadrants. Clear right end conduction delay.



**I – Efferent limb; II – Afferent limb; III, IV – Late forces append. Epsilon waves in inferior leads and aVR.**

Epsilon waves have been defined as any potential after the depolarization between the end of the QRS complex and the beginning of the ST segment (**Wang 2010**). Consequently, the phenomena occur temporarily during the repolarization and not during depolarization. The definition of epsilon wave remains difficult because within the QRS complex are inscribed notches or deflections called fragmentation of the QRS complex (f-QRS). The fQRS at the beginning, on the top, and at the end of QRS complex (termed "pre-, top-, and postsilons") was proposed as typical extended definition of epsilon potentials (**Kukla 2012**).

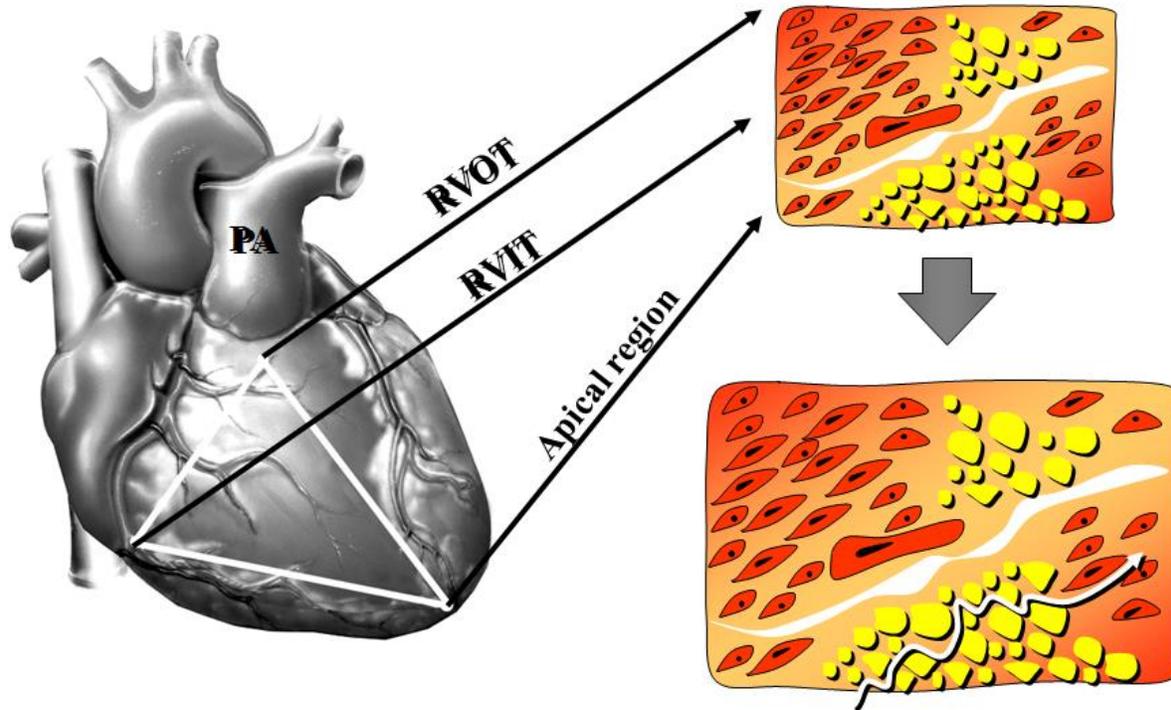
#### V. *Meaning of epsilon waves*

Epsilon waves and other depolarization abnormalities in the right precordial leads are thought to represent delayed activation of the right ventricular outflow tract in arrhythmogenic right ventricular dysplasia/cardiomyopathy (ARVD/C).

Epsilon waves are a major depolarization criterion that represent in the right precordial leads delayed activation of the right ventricular outflow tract in ARVC/D (**Tanawuttiwat 2016**) but they are an insensitive sign when we use Standard 12-leads ECG (S-ECG). Right precordial epsilon potentials were found in 23% in S-ECG and in 75% in highly amplified and modified recording technique (**Peters 2003**). On the other hand, these waves represent a post-excitation phenomenon: delayed activation of "islands" of viable right ventricular myocytes interspersed in myocardium that does not depolarize normally (**Hurst 1998**).

#### VI. *Origin focus of epsilon waves*

The figure below shows the focus location of the epsilon wave on right ventricular free wall in the area called triangle of dysplasia. The angles of this triangle are: the right ventricle outflow track (RVOT), the right ventricle inflow track (RVIT) and the apex of the right ventricle.



**Triangle of dysplasia:** its angles are RVOT, RVIT and apex of RV.

#### VII. Leads where epsilon waves could be observed

**Leads:** epsilon waves are observed mainly in right precordial leads from V<sub>1</sub> to V<sub>3</sub> however are also found the frontal plane, especially in inferior leads. The duration of the QRS complex may be a bit longer in leads V<sub>1</sub> and V<sub>2</sub> than in leads V<sub>5</sub> and V<sub>6</sub>.

#### VIII. Sensitivity ECG for detection of epsilon waves Frequency in ARVC/D with S-ECG with F-ECG and with R-ECG

Epsilon waves are observed in approximately 15-30% of the most severe cases of ARVC/D when is used the standard 12-lead electrocardiogram (S-ECG). This percentage increases if we use the ECG with the modified protocol such as Fontaine leads (F-ECG) (Peters 2014) and right precordial leads (R-ECG). Although the small wiggles may be seen in the routine ECG, they may be seen more readily in Fontaine leads (F-ECG). (Gottschalk 2014).

The presence of epsilon waves by the Fontaine lead system provide a high degree of suspicion for the disease (**Chiladakis 2010**). However, epsilon wave is more commonly seen on Signal Averaged Electrocardiography (SAECG).

#### IX. *Prognosis significance in ARVC/D*

Epsilon waves aid in the prognosis and risk stratification of patients with ARVC/D (**Marcus 2015**). Detection of epsilon waves on 12-lead ECG reflects significant RVOT involvement, which was associated with episodes of sustained ventricular tachycardia but not sudden cardiac death or heart failure (**Protonotarios 2015**). The fQRS complex on S-ECG predicts fatal and nonfatal arrhythmic events in patients with ARVC/D (**Peters 2012**). Therefore, large scale and prospective studies are needed to confirm these findings (**Canpolat 2013**). fQRS is a valuable factor to predict total mortality and major adverse cardiac events (MACE) in patients with CAD. (**Gong 2015**). Sarcoidosis: Multivariate analyses revealed that fQRS complexes are associated. risk of developing cardiac events cardiac events in extracardiac sarcoidosis(**Nagao 2015**). Brugada syndrome (**Morita 2008**). In this entity the presence of fQRS and early repolarization correlates with increased risk in several studies (**Adler 2015**). On multivariable analysis, a history of VF and syncope episodes, inferolateral ER pattern, and f-QRS were independent predictors of documented VF and SCD( **Tokioka 2014**). In a large multicenter, observational, long-term study, the ECG findings that were useful for predict adverse outcome in patients with ARVC/D were: inferior leads T-wave inversion, a precordial QRS amplitude ratio of  $\leq 0.48$ , and QRS fragmentation (correspondent to termed "pre-, top-, and postsilons") (**Saguner 2014**).

#### X. *Pathognomonic character of epsilon waves*

**Pathognomonic character:** in spite of the characteristics in ARVC/D, they are not pathognomonic, since they have been described in other physiological and pathologica scenarios associated with myocardial damage:

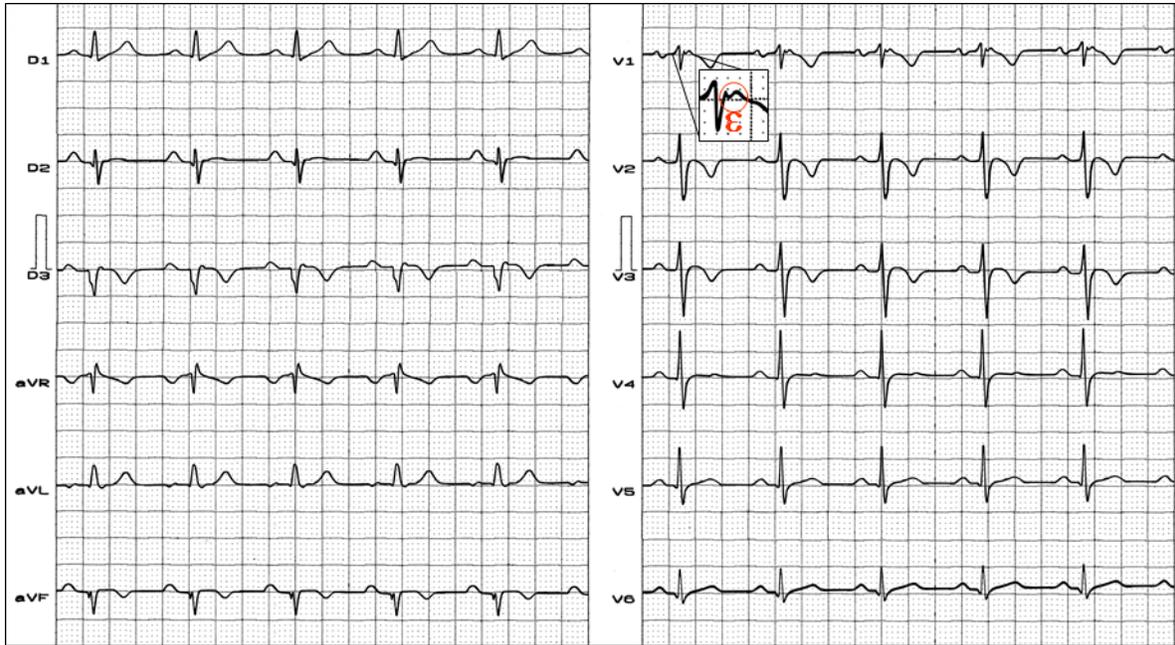
##### I) Physiological epsilon waves

**Ventricular hypertrophy in elite endurance athlete seniors:** Epsilon wave, was found in 3 athletes seniors (1.57%) from 347 elite endurance athletes (seniors–190, juniors–157), mean age of 20; 200 subjects mean age of 21, belonging to the control group of 505 normal sedentary population. (**Macarie 2009**). Bizarre QRS, ST-T patterns suggestive of abnormal impulse conduction in the right ventricle, including the right outflow tract, associated with prolonged QTc interval in some cases were observed in highly trained endurance athletes. The genetic analyses, negative in most athletes, identified surprising mutations in SCN5A and KCN genes in some cases (**Macarie 2009**).

##### II) Pathological epsilon waves

1. **Giant-cell myocarditis:** Epsilon waves are a major diagnostic criterion for ARVC/D, but also other cardiac pathologies such as giant-cell myocarditis can

- cause severe RV conduction disturbances manifesting with epsilon waves and VT on surface ECG. (**Vollmann 2014**)
2. **Sickle cell anemia** (**Hurst 1998**).
  3. **Brugada syndrome:** it is believed that Brugada syndrome and ARVC/D are different clinical entities with respect to the clinical presentation and the genetic predisposition. The coexistence of these two relatively rare clinical entities was also reported (**Hoogendijk 2012**). In clinic practice, there may be cases where the dividing line is not so clear (**An 2008; Ozeke 2009**). Epsilon waves appear to be rare in Brugada syndrome patients and were found in 2 of 47 patients by Letsas et al (**Letsas 2011**), and in 1 patient from a total of 12 unrelated index Brugada syndrome patients were included in the study by Yu et al (**Yu 2014**).
  4. **Idiopathic ventricular fibrillation in the absence of Brugada syndrome phenotype** with loss-of-function mutation of the SCN3B-encoded sodium channel  $\beta_3$  subunit (**Valdivia 2010**).
  5. **During exercise stress testing or treadmill stress testing in asymptomatic gene carriers** Depolarization abnormalities during exercise testing in asymptomatic gene carriers were found to develop more frequently compared with healthy controls: epsilon waves appeared in 4 of 28 (14%) (**Perrin 2013**). Recently, Adler et al showed to uncover epsilon waves in asymptomatic patients carrying mutations in the PKP2 gene. This finding suggests that exercise testing may be valuable for the diagnosis of ARVC/D and that exercise-induced epsilon waves may be found in various genetic subtypes of this disease (**Adler 2015**).
  6. **After repair Fallot tetralogy:** (**George 2011**).
  7. **Right ventricular myocardial infarction** (**Zorio 2010; Andreou 2012**)
  8. **Inferior or lateral ancient dorsal) MI** (**Zorio 2005**)
  9. **Infiltrative diseases, such as cardiac sarcoidosis** (**Santuchi 2004**), increasing evidence suggests that cardiac sarcoidosis might produce the pathological substrate required for production of epsilon waves. Therefore, differentiating these two entities is of paramount clinical importance (**Khaji 2013**). The ECG below shows a single epsilon wave in a patient with sarcoidosis.



**Clinical diagnosis:** cardiac sarcoidosis.

**ECG diagnosis:** SAQRS  $-60^\circ$ , negative T wave from V1 to V3, Epsilon wave ( $\epsilon$ ) in V1.

**High resolution ECG:** observed more frequently with this method.

- In ARVC/D, high resolution ECG frequently is associated to late potentials (LP).
- The  $\epsilon$  wave may be observed in surface ECG; however, it is seen much more frequently in high resolution ECG (**Gregor 2003**).
- High resolution ECG is used to detect late potentials (LP) and  $\epsilon$  waves in ARVD carriers.
- Patients with positive high resolution ECG (presence of LP) have statistically significant increase of S-VT and/or SCD in comparison to those with normal high resolution ECG or branch block.
- High resolution ECG with LP constitutes a marker of arrhythmic events in patients with non-ischemic dilated cardiomyopathies. On the contrary, patients with dilated cardiomyopathies with normal high resolution ECG, display worsening only if they develop progressive CHF (**Mancini 1993**).
- Fibro-fatty substitution of the myocardium is the substrate of slow and fragmented activation, responsible for the presence of LP.
- Abnormal high resolution ECG seems to correlate with the severity of the disease.
- High resolution ECG does not seem a sensitive resource in the minor or concealed forms of the disease, since in these patients there is no proper information with this method (**Oselladore 1995**).

- The combination of the analysis of time domain and frequency domain of high resolution ECG may be useful for screening patients carriers of ARVC/D. This combination of both domains increases sensitivity without reducing specificity.
- Use of filters with a range between 20 and 250 Hz (substituting the classical ranges between 40 and 250 Hz) (**Kinoshita 1995**).
- The presence of LP in ARVD is found in 70% to 80% of cases. These LP may identify patients with a tendency to develop VT runs in little apparent or restricted forms, and it serves to differentiate them from benign RVOT idiopathic VT, with no underlying structural disease. In these cases, high resolution ECG has LP in 0% to 5% of the cases as in normal patients.
- When there is structural heart disease, LPs are found in 20% to 40% of cases. In doubtful cases, invasive studies are necessary to rule out a limited form of cardiomyopathy (**Fauchier 1996**).
- In absence of branch block, the presence of LP in high resolution ECG is proportional to the size of the RV cavity, and thus is parallel to RV dysfunction (**Mehta 1996**).
- In order to study the differences between benign repetitive MVT that originate in the RV and the VT from ARVD, ECG during the event and high resolution ECG may be helpful.
- ECG during VT and high resolution ECG may be useful to differentiate both entities. In the case of ARVD, VT presents QS in V1 and QRSD related to the amount of fibrous tissue existing in the RV (**Kazmierczak 1998**).
- There are significant differences for filtered and non-filtered QRS, low duration sign and square root. In absence of CLBBB, these differences become non-significant for filtered or non-filtered QRS (**Kazmierczak 1998**).
- There is a narrow correlation between the result from high resolution ECG and the extension of the disease, with the presence of VT.
- High resolution ECG is not a valuable resource in minor forms of the disease, but as this is a noninvasive method, it may be useful to assess the progression of the disease (Nava 2000).
- In comparison to 12-lead ECG, high resolution ECG detects abnormalities at higher rates in patients carriers of ARVD (57% vs. 86%). High resolution ECG is more sensitive as screening test than 12-lead ECG to detect patients carriers of ARVD (**Sekiguchi 2001**).
- The anatomopathological process of ARVD also considers late ventricular potentials, which when they are registered as LP in high resolution ECG, indicate electrical stability worsening associated to rapid progression of high resolution ECG, while clinical parameters remain unchanged. This fact suggests that progression parameters in high resolution ECG are markers of electrical instability increase (**Bauce 2002**).

- Sensitivity, specificity, predictive value and accuracy of the different criteria of high resolution ECG were estimated in comparison to SMVT inducibility. Filtered QRS duration (fQRS) in high resolution ECG is considered as predictive for the result of the electrophysiological study and ARVD evolution (**Nasir 2003**).
- The average of presence of late potentials in ARVD is between 70%-80%, with extreme values of 47-100%. The latter percentage is observed in severe forms and with documented spontaneous VT;
- High resolution ECG is a very useful resource to follow the evolution of the disease;
- In relatives of patients, high resolution ECG presents a positivity of LP between 4-16%;
- Detecting posterior potentials improves by using 25 Hz filters and specificity is better observed in the orthogonal lead Z;
- High resolution ECG should be considered a standard test in the study of patients with suspicion or carriers of ARVD;
- Future research is necessary to confirm the value of high resolution ECG as predictor of arrhythmic risk and determining factor of progression of the disease, as well as to study the prevalence of high resolution ECG in relatives of patients, thus allowing early detection;
- The majority of elite and amateur athletes participating in high dynamic and high static sports, reveal a prolongation of the filtered QRS duration (fQRS) on the SAECG, and according to the 2010 Task Force criteria for the diagnosis of ARVC/D, these athletes therefore demonstrate LPs. The extent of fQRS prolongation is positively correlated with RV dimensions. Therefore SAECG findings should be interpreted with caution in endurance athletes. (**Jongman 2015**)
- We hope that multidisciplinary continuing studies on ARVD will help to answer some of these questions (**Nasir 2003**).

Interobserver variability in the assessment of epsilon waves is high; however, the impact of epsilon waves on ARVC/D diagnosis is negligibly low. The results urge to exercise caution in the assessment of epsilon waves, especially in patients who would not otherwise meet diagnostic criteria. (**Platonov 2015**)

#### **I) Value of criterion**

Epsilon wave is considered to be a major depolarization criterion for diagnosis by the Task Force for ARVC/D diagnosis (**McKenna 1994; Fontaine 1999; Marcus 2010**).

**Progressive character of epsilon waves:** ECG changes during long-term follow-up, in a large cohort of patients (111 patients from three tertiary care centers in Switzerland) with ARVC/D showed that ECG progression is significant for epsilon waves (baseline 14% vs. follow-up 31%,  $p = 0.01$ ) (**Saguner 2015**).

## Value of ECG in ARVC/D

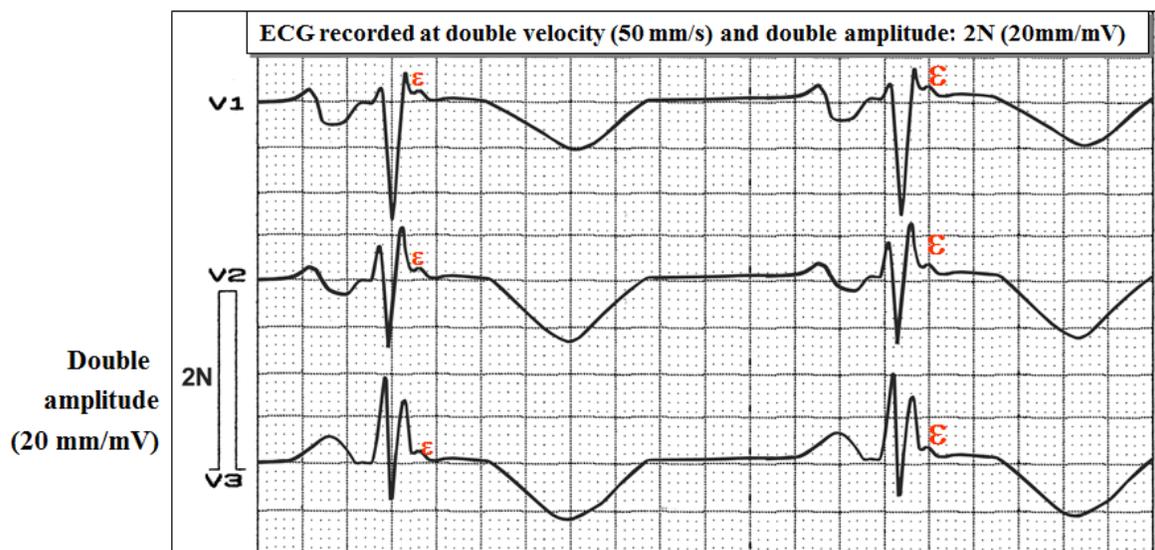
ECG diagnosis of ARVC/D may be difficult in the initial stage of the disease, since a normal ECG is found in up to 40% of patients during the first year of follow-up. Jaoude et al (**Jaoude 1996**) found a strong correlation between QRS or T wave changes and the length of follow-up after the first symptom; mean time interval between first VT and ECG recording is significantly longer in patients with negative T waves in the right precordial leads, wide QRS, or left axis deviation, than in patients without such abnormalities. A normal ECG is found in 40% of patients during the first year of follow-up, 8% at 5 years, and never later than the 6th year. ARVC/D can be excluded if the ECG is found to be normal 6 years or later after a first VT event.

**ECG association:** Inversion of T wave in leads V1-V3 and/or  $\epsilon$  wave found in 70% of patients with ARVC/D. Epicardic electrophysiological studies in dysplastic areas reveal the LP that occur at the end of the QRS complex, in the J point, and at the onset of the ST segment, explained by fibro-fatty substitution of myocardial tissue (**Fontaine 1984**).

**Epsilon wave and relationship to VT:** the simple presence of these waves indicate slow and fragmented conduction, which favors reentry circuits, which in turn result in M-VT runs with CLBBB morphology by originating in the RV (**Hurst 1998; McKenna 1994**).

The tracing should run at a double velocity (50 mm/s) and double voltage (20 mm/s) to compare the duration of QRS complexes (QRSd) in different leads, as well as to try to record Epsilon waves.

The ECG below shows more clearly the epsilon wave with double velocity and double voltage.



The rate of widespread T-wave inversion (exceeding V<sub>3</sub>) was significantly higher in patients with epsilon waves than in those without

Comparative sensitivity of

- Standard 12-lead electrocardiography (S-ECG)

- Right-sided precordial lead electrocardiography (R-ECG) R-ECG (V<sub>3R</sub>, V<sub>4R</sub>, V<sub>5R</sub>),
- Fontaine bipolar precordial lead electrocardiography (F-ECG). The Fontaine bipolar precordial leads placed at the manubrium of sternum, xiphoid, and V<sub>4</sub> positions using the right arm connection, left arm connection, and left foot connection, respectively
- Detection by these 3 methods
- Signal-averaged ECG

Because these waves are of relatively low voltage and may go undetected by standard electrocardiography (S-ECG) or unnoticed by the interpreter (**Zorio 2005**).

### Value of the Fontaine bipolar precordial leads

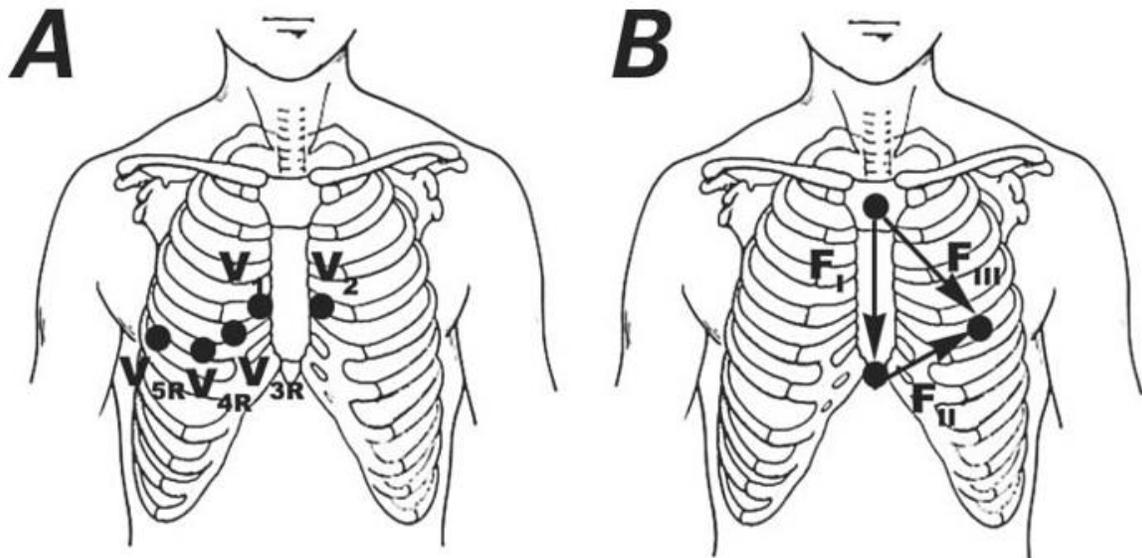
The tracing should be obtained from I and aVF at double velocity and amplitude, placing the electrode of the left arm on the xiphoid appendix, the one from the right arm on the manubrium sternum, and the one from the left leg on the rib at the fourth or fifth space with the aim of improving the ability to detect Epsilon waves.

The Fontaine bipolar precordial leads are placed at the manubrium of sternum, xiphoid, and V<sub>4</sub> positions using the right arm connection, left arm connection, and left foot connection, respectively.

Epsilon waves are detected by:

- 1) Standard 12-lead electrocardiography (S-ECG)
- 2) Right-sided precordial lead electrocardiography (R-ECG)
- 3) Fontaine bipolar precordial lead electrocardiography (F-ECG).

The figure below shows electrodes location in R-ECG and F-ECG



**Right precordial leads**

**The Fontaine bipolar precordial leads**

The detection rate using combined methods is significantly higher than that by S-ECG alone.

Fontaine bipolar precordial lead have the best sensitivity among the three options. The placement of the foot lead (positive) in position V4 provides, instead of regular leads I, II, and III, three bipolar chest leads that can be called FI, FII, and FIII. Tracings are then produced by setting the machine on regular leads I, II, and III. This arrangement is used to record specifically the potentials developed in the RV, from the RVOT to the diaphragmatic area. The vertical bipolar lead FI, (similar to aVF lead), seems to be the most appropriate to record epsilon waves; it also magnifies the atrial potentials. As late potentials were supposed to be the result of late activation of a limited group of fibers, the term "post-excitation" looked logical, since it was observed after the main excitation of the ventricle, leading to the QRS complex. The term "epsilon" was appropriate, because it occurs in the Greek alphabet after delta; thus, delta represents the pre-excitation and epsilon the post-excitation phenomenon (**Fontaine 1999**).

The bipolar leads of the figure below (S-ECG x F-ECG) show major sensibility of Fontaine ECG.

### **Others ECG features in ARVC/D**

Approximately 90% of patients carriers of ARVC/D present ECG anomalies. ECG abnormalities were more frequent at 10 year and 5 year follow-up than on initial tracings. A normal ECG was found in 40% of patients during the first year of follow-up, 8% at 5 years, and never later than the 6th year. Consequently, ARVC/D diagnosis may be excluded if ECG is normal 6 years of follow-up (**Jaoude 1996**). In ARVD/C, a normal ECG is considered reassuring. However, some patients with ARVD/C experiencing ventricular arrhythmias have a normal ECG. Interpretation of ECG in young and older athletes requires in-depth knowledge in cardiology and sports medicine. The interpretation can only be carried out by considering medical history, clinical examination and ethnicity. Profound and long-term experience of athlete's ECG interpretation is required to protect athletes and to prevent cardiac emergencies. (**Löllgen 2015**)

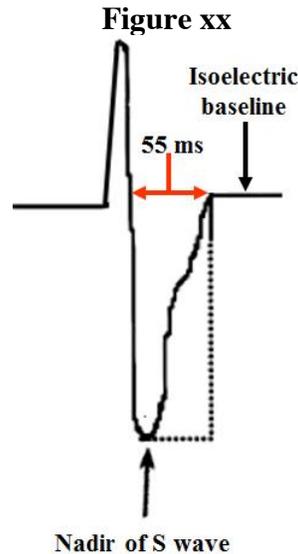
Main ECG features in ARVC/D classification

- I) Depolarization criteria  
Right ventricular parietal block: A prolonged S-wave upstroke in V1 through V3 ( $\geq 55\text{ms}$ )
- II) Repolarization criteria  
Inverted T-wave in the right precordial leads V1-3 or anterior T wave inversion (TWI) above 12 years with no RBB (repolarization criteria):

### **Depolarization criteria**

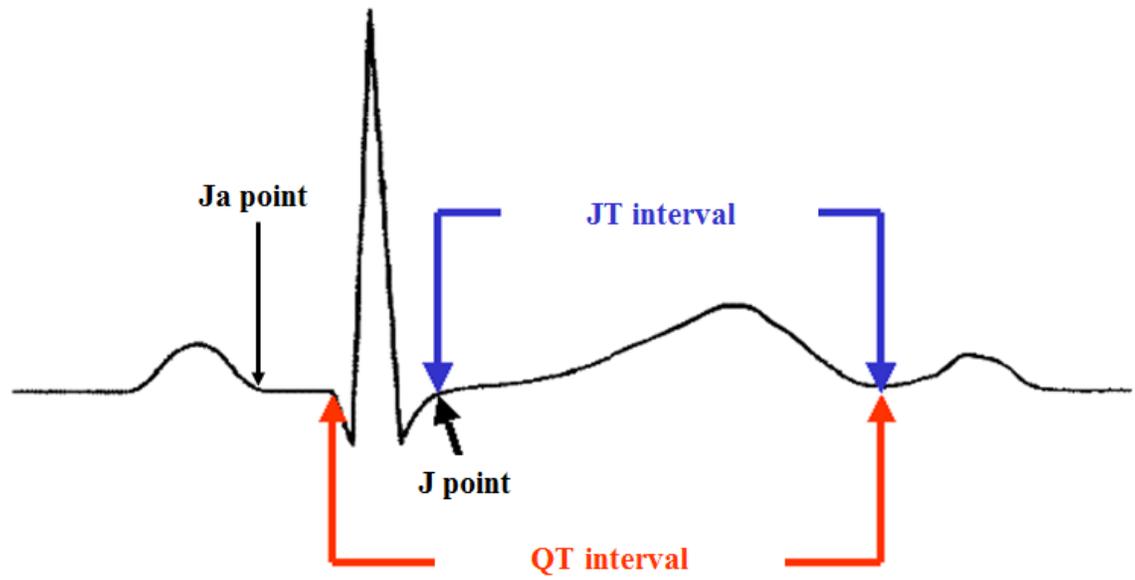
- 1) **Right ventricular parietal block** (**Fontaine 1984**): Prolonged S-wave duration due to slow depolarization of the terminal part of the QRS because the RV is the last part of the heart to undergo depolarization, A prolonged S-wave upstroke in V1 through V3 ( $\geq 55\text{ms}$ ) is the most frequent ECG finding in ARVD/C and should be

considered as a diagnostic ECG marker. Among those without RBBB, a prolonged S-wave upstroke in V1 through V3  $\geq 55$  ms was the most prevalent ECG feature (95%) and correlated with disease severity and induction of VT on EPS (Figure xx).



This feature also best distinguished ARVD/C (diffuse and localized) from RVOT. The sensitivity of this criterion is not known in other entities and it speaks in favor of slow RV conduction. A study shows that the sign is not specific, since it is found in Brugada syndrome (**Pitzalis 2003**) with QT interval prolongation only from V1 to V3. If QT interval prolongation occurs only from V1 to V3, it is clear that this is due to depolarization time prolongation. If we admit that in Brugada syndrome there is some degree of RBBB, this QT interval prolongation may be partially due to this fact. QT interval constitutes a classical measurement for ventricular repolarization; however, it includes depolarization (QRS), which represents the so-called “electrical systole”, which includes ventricular depolarization and repolarization. In these cases of branch block and WPW, it is better to measure the JT interval and not QT (Figure xxx).

**Figure xxx - The JT interval value and its limits**



QT interval is used to measure ventricular repolarization; nevertheless, this parameter includes ventricular depolarization (QRS) and represents the so-called electrical systole, which is the addition of ventricular depolarization (QRS) and repolarization (ST/T = JT interval).

If bundle branch block or WPW type ventricular pre-excitation occurs, the QTc interval does not express ventricular repolarization correctly. In these cases, JT interval measurement is more reliable ( $JT = QT - QRSd$ ) than QT interval, because the parameter excludes depolarization that is prolonged, as a consequence of sequential activation of biventricular chamber (normally this activation is simultaneous).

- 2) Localized QRSD prolongation on right precordial leads  $> 110\text{ms}$  (depolarization/conduction abnormality).
- 3)  $QRSd_{V1+V2+V3} / QRSd_{V4+V5+V6} > 1.2$  in approximately 65% of cases. QRS prolongation located in right precordial leads has 91% sensitivity, 90% specificity that predicts VT in patients carriers of ARVC/D (**Nasir 2003; 2004**).
- 4) The terminal S wave length and area in the right precordial leads are diagnostically useful and suitable for automatic analysis in ARVC/D. The bipolar chest leads (CF leads) are diagnostically superior to the unipolar precordial leads. Among members of ARVC families, those with mutations had shorter QRS length in V2 and V3 and smaller QRS area in lead V2 compared with those without mutations. In ARVC patients, the CF leads were diagnostically superior to the standard unipolar precordial leads. Terminal S wave duration in V1  $\geq 48\text{ms}$  and T wave negativity in CF leads separated ARVC patients from matched controls with 90% sensitivity and 86% specificity. (**Batchvarov 2015**)

**Observation:** The bipolar chest leads (CF leads) is a lead that resembles V1 (modified CL1 lead) The positive electrode is placed at V1 and the negative electrode is placed close to the left shoulder. It is frequently used for detecting arrhythmias during continuous monitoring of the patients admitted to the coronary care unit.

- 5) **Epsilon waves, epsilon potentials, ventricular post-excitation waves (Maia 1991), post-excitation (Epsilon) waves (Okano 1995), or Fontaine waves** due to slow conduction in the RV. The extent of ECG abnormalities correlate with the degree of structural change in the RV. ( **Marcus 2009**)
- I. **QRS fragmentation(fQRS):** QRS fragmentation in the S wave of right precordial leads identifies patients with recurrent VT, primary VF, and recurrent ICD discharges; fQRS  $\geq 3$  leads characterized patients who died from SCD (**Peters 2012**).
  - II. **Reduced QRS amplitude:** Q waves or precordial QRS amplitudes  $< 1.8$  mV;
  - III. **Poor R Wave Progression (PRWP) on precordial leads: The most likely cause of PRWP is clockwise rotation caused by RV enlargement. (Fontaine 1984)**
  - IV. Complete or incomplete RBBB based on the findings from epicardial mapping and histological data, is likely attributable not only to the impaired septal predivisional right bundle branch but also to distal block on RV free wall due to the irregular and delayed propagation of activation in the zones of dysplasia

#### Repolarization criteria

- V. **Inverted T-wave in the right precordial leads V1-3 or anterior T wave inversion (TWI) above 12 years with no RBB (repolarization criteria):** T-wave inversion on a 12-lead ECG is usually dismissed in young people as normal persistence of the juvenile pattern of repolarization. However, T-wave inversion is a common ECG abnormality of cardiomyopathies such as hypertrophic cardiomyopathy and ARVC which are leading causes of SCD in athletes. In absence of CRBBB in patients  $> 12$  years of age, T wave inversion(TWI) \from V1 to V3 is a sign with great value for the diagnosis. The juvenile pattern of T wave inversion in V<sub>1</sub>-V<sub>3</sub> or beyond is a normal variant in children under 12 years of age. This variant is present in 1%-3% of the healthy population aged 19 to 45 years and 87% of patients with ARVC(**Capulzini 2010**). In normal, young patients, there is usually positive T polarity in V1; however, it may flatten and nearly always has a positive polarity in V2. In symptomatic patient's carriers of ARVC/D, the ECG generally shows T wave inversion in V1 and V2, which may reach up to V6 (**Fontaine 1994**). Physiological cardiac adaptation to regular exercise, including biventricular dilation and T-wave inversion (TWI), may create diagnostic overlap with ARVC/D). There are no electrical, structural, or functional cardiac differences between athletes exhibiting TWI and athletes without TWI. When athletes are compared with ARVC/D patients, markers of physiological remodeling included early repolarization, biphasic TWI, voltage criteria for RVH or LVH, and symmetrical cardiac enlargement. Indicators of RV pathology included the following: syncope; Q waves or precordial QRS amplitudes  $< 1.8$  mV; 3 abnormal SAECG parameters; delayed gadolinium enhancement, RV ejection fraction  $\leq 45\%$ , or wall motion abnormalities at CMRI;  $> 1,000$  premature ventricular contractions (or  $> 500$  non-RV outflow tract) per 24 h; and symptoms, ventricular

tachyarrhythmias, or attenuated blood pressure response during exercise.( **Zaidi 2015**) Nonspecific parameters included the following: prolonged QRS terminal activation;  $\leq 2$  abnormal SAECG parameters; RV dilation without wall motion abnormalities; RV outflow tract ectopy; and exercise-induced T-wave pseudonormalization. In ARVC/D TWI is due to scarring of the free wall of the RV and regional conduction delay on free wall RV. (**Marcus 1982, Peters 2003, Steriotis 2009**), is one of the most common ECG abnormalities in ARVC/D. To day is considered a major taskforce diagnostic criterion ( **Marcus 2010**). (**Sen-Chowdhry 2007**)may be the causes of T wave inversion in V1-3 or beyond. It is a secondary rather than a primary repolarization abnormality. T wave inversion beyond V3 is more common in ARVC/D patients in the advanced stage of the disease with severe RV dilatation and LV involvement. Thus it has been considered a risk factor and perhaps an indication of a poor prognosis. T wave inversion in the right precordial leads can also be seen in many other conditions such as acute pulmonary embolism, athlete heart, Brugada syndrome, long QT syndrome caused by *KCNH2* mutations or compound mutations, and occasionally in normal adult female. Exercise-induced T-wave pseudonormalization.

- VI. **ST segment elevation:** The combination of J-point elevation and TWI confined to lead V1-V4 offers the potential for an accurate differentiation between 'physiologic' and 'cardiomyopathic' anterior TWI, among athletes of both white/Caucasian or black/Afro Caribbean descent (**Calore 2015**). Conversely, ST-segment elevation without J-point elevation preceding anterior TWI may reflect cardiomyopathy. is not uncommon in ARVC/D (**Peters 1999**). In a cohort study, 37% of ARVC/D patients had a ST elevation. Among these, 42% showed a small notch in the first half of the ST segment and such findings are more frequently seen in patients in the presence of epsilon waves.

### **Ventricular arrhythmias**

- Nonsustained or sustained ventricular tachycardia in the morphology of left bundle branch block with superior axis: Predominant negative or indeterminate in inferior leads and positive in aVL is considered a major criterion following 2010 revised Task Force Criteria for the Diagnosis of ARVC. VT with LBBB morphology and an inferior axis commonly originates from the Right Ventricular Outflow Tract (RVOT). In contrast to ARVC, RVOT VT occurs in structurally normal hearts (occasionally the RVOT is dilated and RV regional wall motion abnormalities are seen on CMR) and is readily treatable with verapamil and  $\beta$ -blockers or radiofrequency ablation. The ECG in sinus rhythm in RVOT VT is normal as is the SAECG. In contrast to ARVC, there are no family screening implications with RVOT VT. Other differentials to consider include idiopathic dilated cardiomyopathy (IDCM) and Uhl's anomaly. Patients with IDCM usually have a progressive decline in left ventricular function, in contrast to ARVC where the right heart is primarily affected.

ECG differentiation of idiopathic right ventricular outflow tract ectopy with LBBB/inferior axis from early ARVC/D (**Novak 2016**)

	<b>Idiopathic right ventricular outflow ectopy</b>	<b>Early ARVC</b>
QRS duration >160 ms	27%	60%
R peak time >80 ms	24%	65%
Initial QRS slurring	12%	40%
QS pattern in lead V1	36%	90%
QRS axis >90°	24%	60%

- In Uhl's anomaly the RV myocardium is paper thin and devoid of myocardium. There is no replacement of muscle by fatty tissue. It usually presents in childhood.
- 1,000 ventricular extra systoles (or >500 non-RV outflow tract) per 24 h.

<b>1. RV on Imaging</b>	
Major	<b>By 2D echo:</b> <ul style="list-style-type: none"> <li>Regional RV akinesia, dyskinesia, or aneurysm</li> <li>and 1 of the following (end diastole): <ul style="list-style-type: none"> <li>PLAX RVOT <math>\geq 32</math> mm (corrected for body size [PLAX/BSA] <math>\geq 19</math> mm/m<sup>2</sup>)</li> <li>PSAX RVOT <math>\geq 36</math> mm (corrected for body size [PSAX/BSA] <math>\geq 21</math> mm/m<sup>2</sup>)</li> <li>or fractional area change <math>\leq 33</math> percent</li> </ul> </li> </ul>
	<b>By MRI:</b> <ul style="list-style-type: none"> <li>Regional RV akinesia or dyskinesia or dyssynchronous RV contraction</li> <li>and 1 of the following: <ul style="list-style-type: none"> <li>Ratio of RV end-diastolic volume to BSA <math>\geq 110</math> mL/m<sup>2</sup> (male) or <math>\geq 100</math> mL/m<sup>2</sup> (female)</li> <li>or RV ejection fraction <math>\leq 40</math> percent</li> </ul> </li> </ul>
	<b>By RV angiography:</b> <ul style="list-style-type: none"> <li>Regional RV akinesia, dyskinesia, or aneurysm</li> </ul>
Minor	<b>By 2D echo:</b> <ul style="list-style-type: none"> <li>Regional RV akinesia or dyskinesia</li> <li>and 1 of the following (end diastole): <ul style="list-style-type: none"> <li>PLAX RVOT <math>\geq 29</math> to <math>&lt; 32</math> mm (corrected for body size [PLAX/BSA] <math>\geq 16</math> to <math>&lt; 19</math> mm/m<sup>2</sup>)</li> <li>PSAX RVOT <math>\geq 32</math> to <math>&lt; 36</math> mm (corrected for body size [PSAX/BSA] <math>\geq 18</math> to <math>&lt; 21</math> mm/m<sup>2</sup>)</li> <li>or fractional area change <math>&gt; 33</math> percent to <math>\leq 40</math> percent</li> </ul> </li> </ul>
	<b>By MRI:</b> <ul style="list-style-type: none"> <li>Regional RV akinesia or dyskinesia or dyssynchronous RV contraction</li> <li>and 1 of the following: <ul style="list-style-type: none"> <li>Ratio of RV end-diastolic volume to BSA <math>\geq 100</math> to <math>&lt; 110</math> mL/m<sup>2</sup> (male) or <math>\geq 90</math> to <math>&lt; 100</math> mL/m<sup>2</sup> (female)</li> <li>or RV ejection fraction <math>&gt; 40</math> percent to <math>\leq 45</math> percent</li> </ul> </li> </ul>
<b>2. Histology</b>	
Major	Residual myocytes $< 60$ percent by morphometric analysis (or $< 50$ percent if estimated), with fibrous replacement of the RV free wall myocardium in $\geq 1$ sample, with or without fatty replacement of tissue on endomyocardial biopsy
Minor	Residual myocytes 60 percent to 75 percent by morphometric analysis (or 50 percent to 65 percent if estimated), with fibrous replacement of the RV free wall myocardium in $\geq 1$ sample, with or without fatty replacement of tissue on endomyocardial biopsy
<b>3. ECG - Repolarization abnormalities</b>	
Major	Inverted T waves in right precordial leads (V <sub>1</sub> , V <sub>2</sub> , and V <sub>3</sub> ) or beyond in individuals $> 14$ years of age (in the absence of complete right bundle-branch block QRS $\geq 120$ ms)
Minor	Inverted T waves in leads V <sub>1</sub> and V <sub>2</sub> in individuals $> 14$ years of age (in the absence of complete right bundle-branch block) or in V <sub>4</sub> , V <sub>5</sub> , or V <sub>6</sub> or V <sub>4</sub> , V <sub>5</sub> , V <sub>6</sub> , and V <sub>6</sub> in individuals $> 14$ years of age in the presence of complete right bundle-branch block
<b>4. ECG - Depolarization/conduction abnormalities</b>	
Major	Epsilon wave (reproducible low-amplitude signals between end of QRS complex to onset of the T wave) in the right precordial leads (V <sub>1</sub> to V <sub>3</sub> )
Minor	Late potentials by SAECG in $\geq 1$ of the following 3 parameters in the absence of a QRS duration of $\geq 110$ ms on the standard ECG <ul style="list-style-type: none"> <li>Filtered QRS duration (fQRS) <math>\geq 114</math> ms</li> <li>Duration of terminal QRS <math>&lt; 40</math> <math>\mu</math>V (low-amplitude signal duration) <math>\geq 38</math> ms <ul style="list-style-type: none"> <li>Root-mean-square voltage of terminal 40 ms <math>\leq 20</math> <math>\mu</math>V</li> </ul> </li> <li>Terminal activation duration of QRS <math>\geq 55</math> ms measured from the nadir of the S wave to the end of the QRS, including R', in V<sub>1</sub>, V<sub>2</sub>, or V<sub>3</sub> in the absence of complete right bundle-branch block</li> </ul>
<b>5. Arrhythmias</b>	
Major	Nonsustained or sustained ventricular tachycardia of left bundle-branch morphology with superior axis (negative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL)
Minor	Nonsustained or sustained ventricular tachycardia of left bundle-branch morphology with superior axis (negative or indeterminate QRS in leads II, III, and aVF and positive in lead aVL)
<b>6. Family History</b>	
Major	<ul style="list-style-type: none"> <li>ARVC/D confirmed in a first-degree relative who meets current Task Force criteria</li> <li>ARVC/D confirmed pathologically at autopsy or surgery in a first-degree relative</li> <li>Identification of a pathogenic mutation <math>\Delta</math> categorized as associated or probably associated with ARVC/D in the patient under evaluation</li> </ul>
Minor	<ul style="list-style-type: none"> <li>History of ARVC/D in a first-degree relative in whom it is not possible or practical to determine whether the family member meets current Task Force criteria</li> <li>Premature sudden death (<math>&lt; 35</math> years of age) due to suspected ARVC/D in a first-degree relative</li> <li>ARVC/D confirmed pathologically or by current Task Force Criteria in second-degree relative</li> </ul>

Definite diagnosis = 2 Major or 1 Major and 2 Minor criteria or 4 Minor from different categories

Borderline Diagnosis = 1 Major and 1 Minor or 3 Minor criteria from different categories

Possible Diagnosis = 1 Major or 2 Minor criteria from different categories

PLAX: parasternal long-axis view; RVOT: RV outflow tract; BSA: body surface area; PSAX: parasternal short-axis view; aVF: augmented voltage unipolar left foot lead; aVL: augmented voltage unipolar left arm lead.

## 2010 ESC revised Task Force Criteria for the Diagnosis of ARVC

### Case report

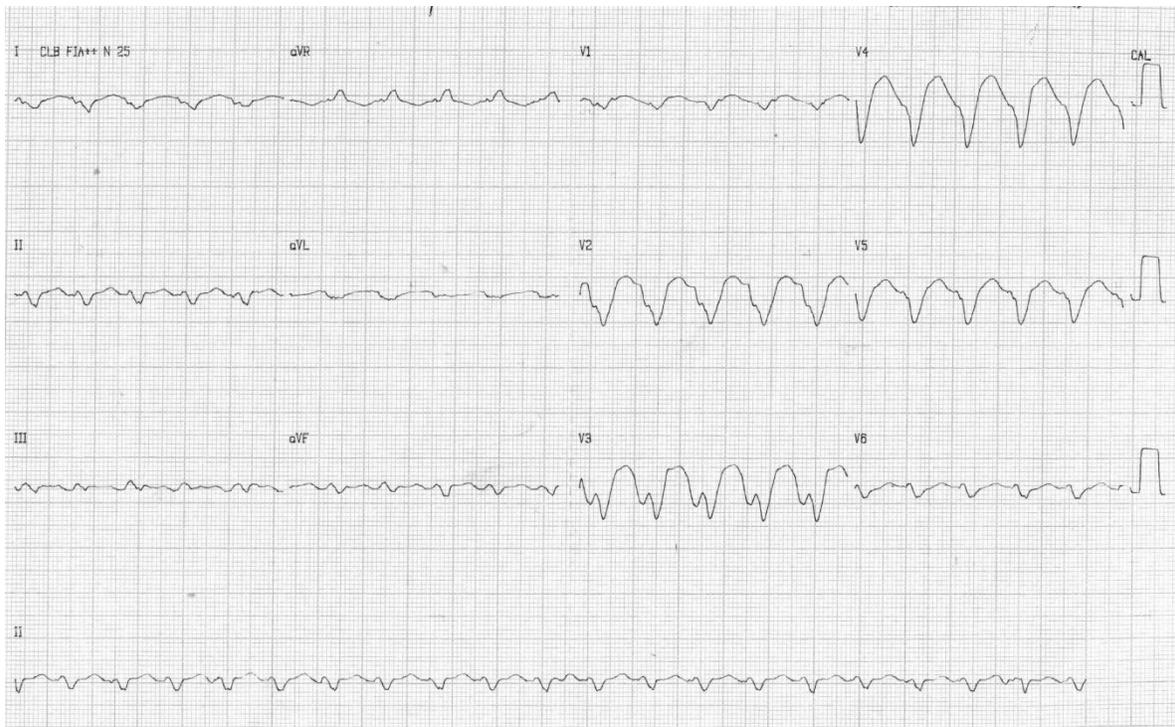
JPSF, 26 years old, born and living in Itatira-CE, Brazil, single, farmer.

Main complaint: "accelerated heart".

Patient was admitted in our service on 22/07/2012 with palpitations, associated with dizziness and profuse sweating. He denied chest pain, dyspnea and other clinical complaints.

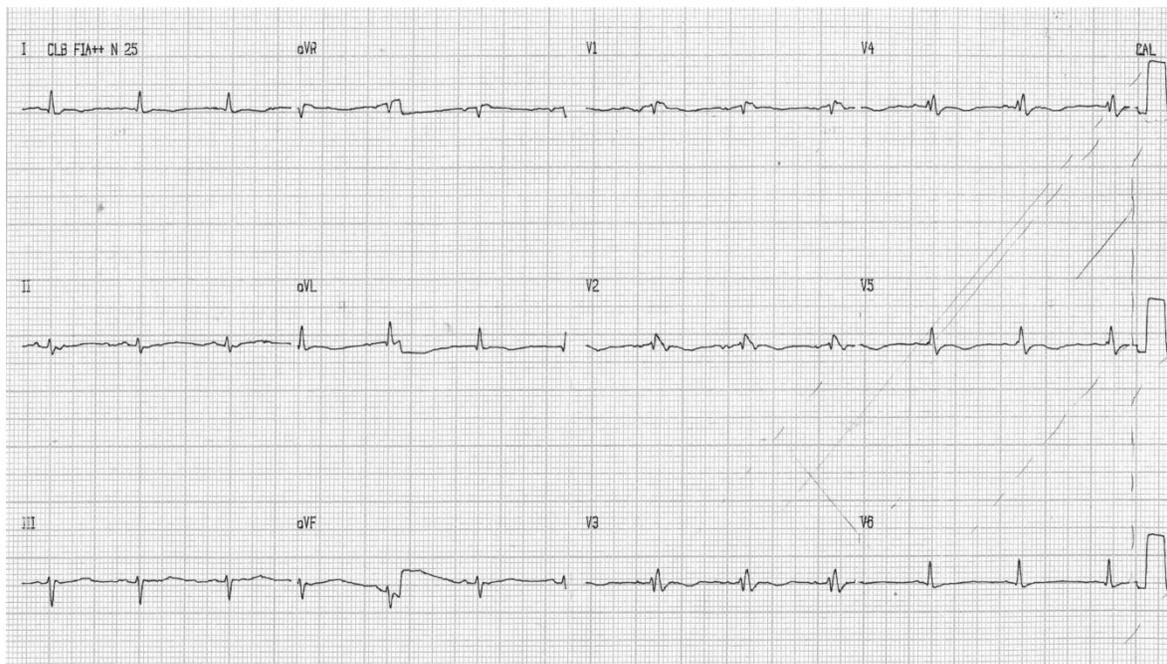
Uncles and cousins with similar symptoms, but no reports of sudden death in the family members.

### ECG at admission with hemodynamic instability

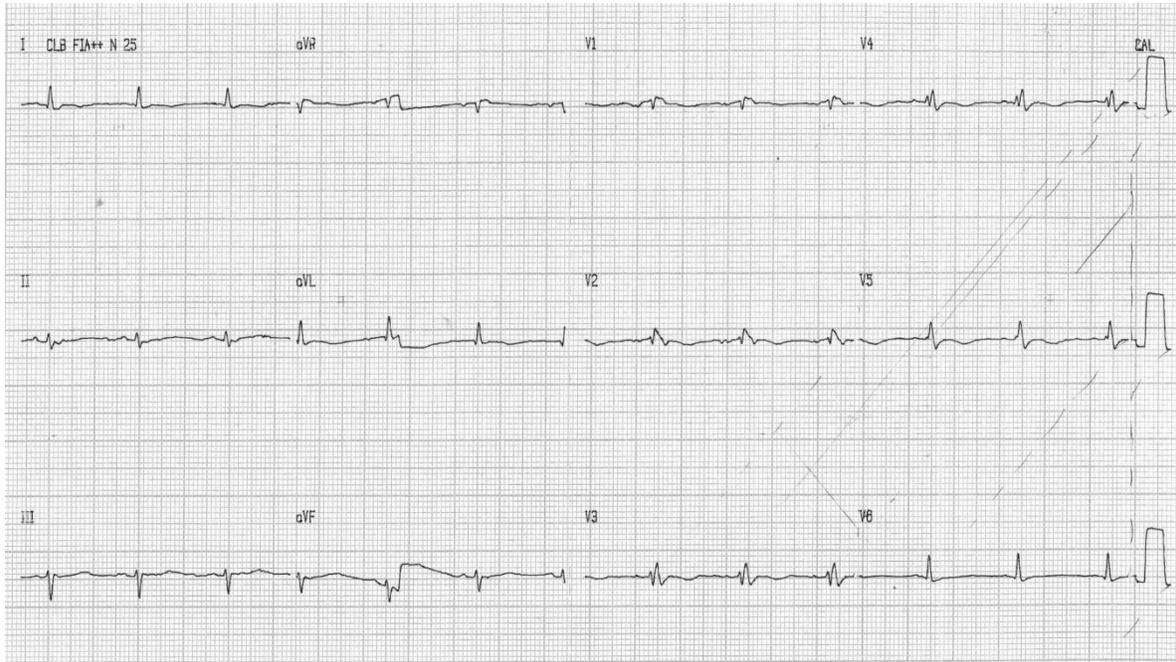


**ECG diagnosis:** sustained VT with LBBB pattern, heart rate = 125 bpm and right superior QRS axis (only aVR lead with positive QRS complexes). This atypical axis is a hallmark of VT with focus in apex of right ventricle.

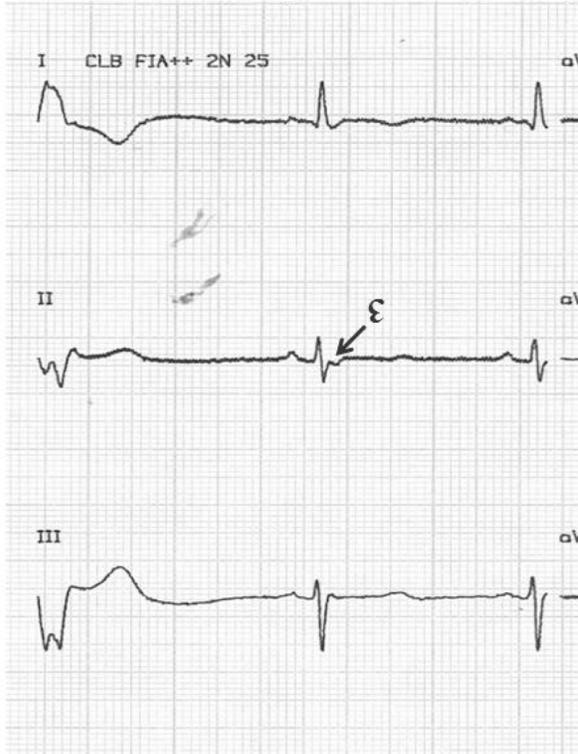
### ECG after electrical cardioversion



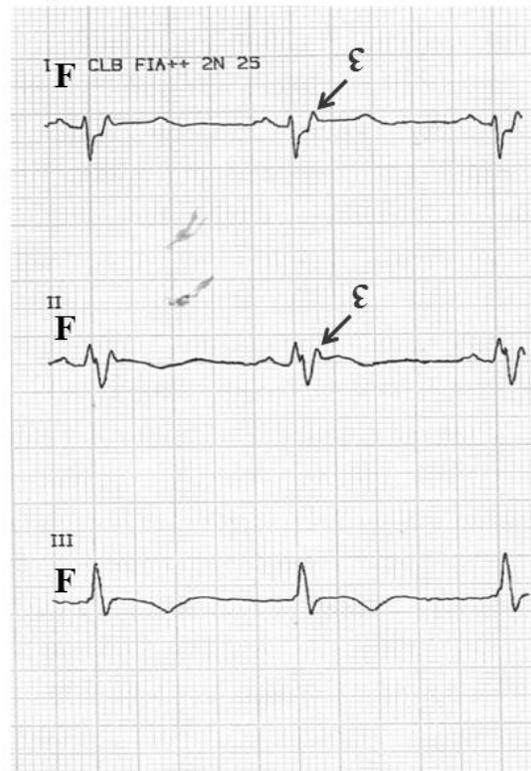
**ECG diagnosis:** universal low voltage of QRS complexes. V2 led with type 1 Brugada phenocopy. Negative T waves from V1 to V5



### Standard ECG bipolar leads (S-ECG)



### Bipolar leads of Fontaine (F-ECG)



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