

OVERVIEW OF THE ECG

This ECG shows a regular rhythm (54bpm) with a P wave axis of +75 degrees and a PR interval of 280ms (sinus bradycardia with 1st degree AV block). The QRS axis is -30 degrees with a pattern of left anterior fascicular block (LAFB). The QRS complexes are wide (around 120ms). A QRS pattern abnormality is present, which is described below. Diffuse T wave inversion is seen in V1 - V6 and in III and aVF.

MORE DETAILED ANALYSIS OF THE ECG

The next step to the QRS pattern abnormality is to examine the QRS morphology in V1 and V6. The QRS complexes in V1 and V6 do not resemble a typical right bundle branch block (RBBB) pattern (Figure 2A). There is no typical rSR pattern in V1 and no qRS pattern in V6. The QRS complex is wide because of a low amplitude signal or wave that is best seen at the end or immediately after the QRS complex. This signal or wave is seen in the precordial leads, but is best seen in V1 and lead II. Note that a "small" amplitude wave differs from the "large" amplitude R of typical RBBB caused by delayed right ventricular activation.

The QRS morphology in V1 - V3 is not consistent with a Brugada pattern. The type I Brugada pattern is characterised by coved, downsloping ST segment elevation >2mm, followed by T wave inversion in leads V1 - V3 (Figure 2B). The low amplitude wave at the end of the QRS in V1 should not be misinterpreted as ST segment elevation. The absence of ST elevation in V2 and V3 supports the finding that the low amplitude wave is independent of the ST segment.

The early repolarisation pattern is characterised by elevation of the J point with slurring or notching of the terminal portion of the QRS complex usually seen in the inferior (II, III or aVF) or lateral (I, AVL, V4 - V6) leads. The diffuse low amplitude signal occurs at the end or immediately after the QRS and should not be misinterpreted as J point elevation or notching of the QRS itself. Benign anterior ST segment elevation with upright T waves is a common variant in young black men and women (Figure 2C).

The Wellens' pattern refers to deep symmetrical T wave inversion or biphasic T waves best seen in V2 and V3 usually in the setting of a narrow QRS complex (Figure 2E) and chest pain. This pattern is suggestive of anterior left ventricular wall ischaemia and manifests as a primary T wave abnormality with normal ST segments. Although T wave inversion is present in this ECG in V2 and V3, the diffuse T wave inversion (involving

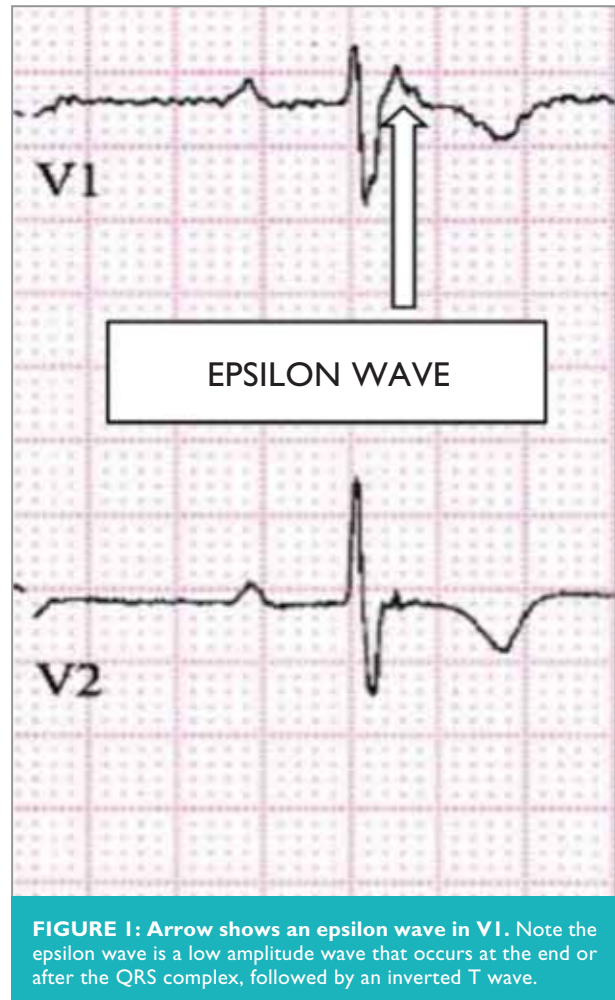


FIGURE 1: Arrow shows an epsilon wave in V1. Note the epsilon wave is a low amplitude wave that occurs at the end or after the QRS complex, followed by an inverted T wave.

all the precordial leads and inferior leads) and the abnormal low amplitude signals seen at the end of the QRS complexes, need further explanation.

The low amplitude signal or wave between the end of the QRS complex and the onset of the T wave is pathognomonic of an epsilon wave (Figure 1 and Figure 2D). The epsilon wave is caused by low amplitude potentials due to delayed depolarisation of "islands" of surviving cardiomyocytes between areas of fibrosis and fat usually in the right ventricle, which explains why the wave is best seen in the right precordial leads V1 - V3. Although epsilon waves are best seen in the right precordial leads (V1 - V3) with right ventricular involvement, they can also be seen in the inferior and lateral leads in patients with left ventricular involvement, as in this case.

The answer to question 1 is: (4) Sinus bradycardia with 1st degree AV block, LAFB and an epsilon wave.

DISCUSSION

The epsilon wave was originally described by Fontaine in 1977, who described tiny signals that consistently occurred “after the end of each QRS complex” or as a “slur at the end of the right precordial QRS complexes”.⁽¹⁾ Other descriptions include “low-amplitude signals that occur at the end” of or “immediately after the QRS complex” in the right precordial leads or a “terminal deflection within or at the end of the QRS complex”.

The name epsilon wave was chosen by Fontaine because of (1) its low amplitude (epsilon is the mathematical symbol for smallness); (2) it is a post-excitation wave at the end of the

QRS complex in contrast to a delta wave which is a pre-excitation wave at the beginning of the QRS complex; and (3) it is the next Greek letter after delta.⁽¹⁾

While epsilon waves can be seen in patients with ARVC, they have also been described in other conditions: cardiac sarcoidosis, Uhl's anomaly, right ventricular myocardial infarction, post Tetralogy of Fallot repair and sickle cell anaemia with RV hypertrophy due to pulmonary hypertension.⁽¹⁾

The answer to question 2 is: (6) – I (cardiac sarcoidosis) and 4 (ARVC) are possibilities.

An epsilon wave is a major diagnostic depolarisation criterion for ARVC according to the 2010 task force criteria.⁽²⁾ The prevalence of epsilon waves in patients with a definite diagnosis

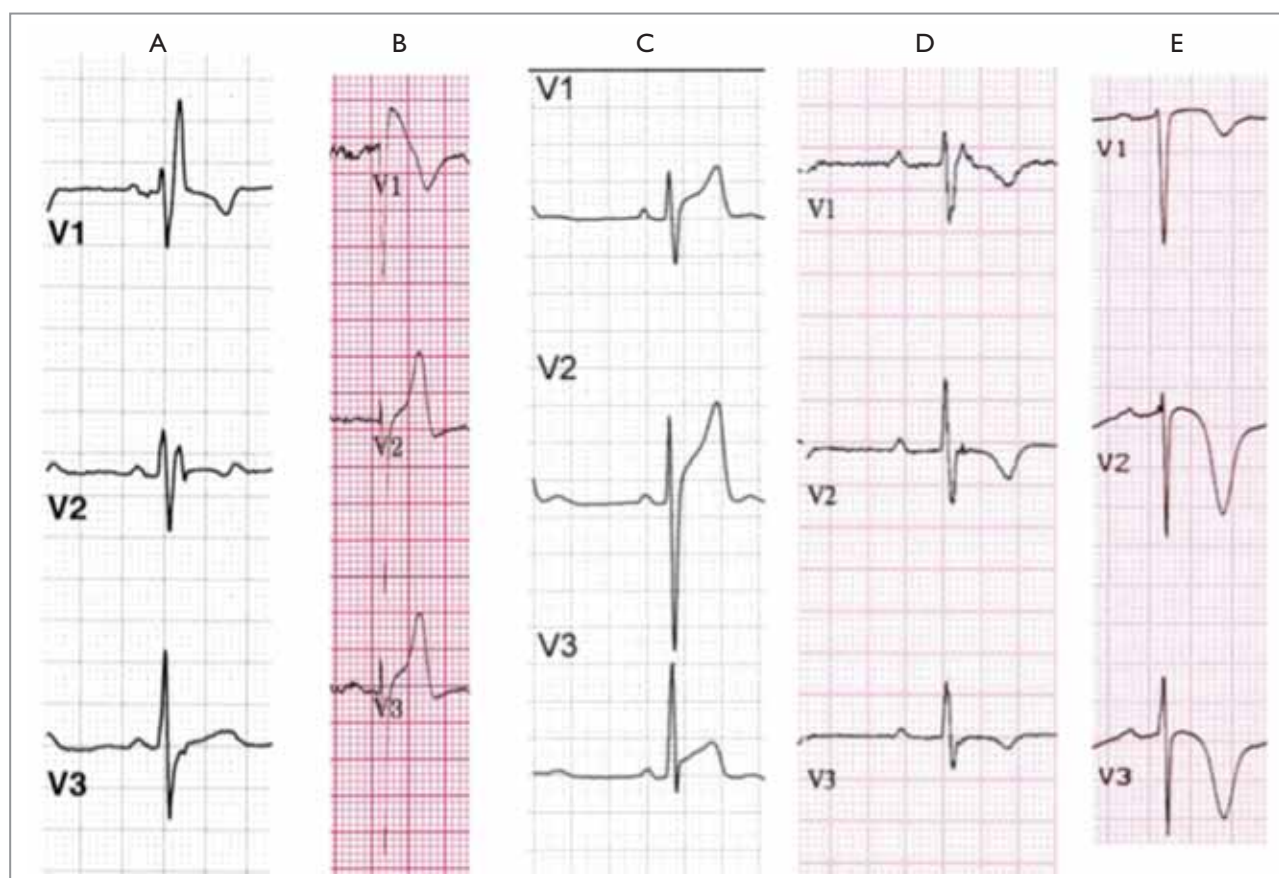


FIGURE 2: Examples of conditions with abnormal QRS morphologies, ST segment and/or T wave changes in V1 - V3. A = right bundle branch block, B = Brugada pattern, C = early repolarisation pattern, D = epsilon wave, E = Wellens' pattern.

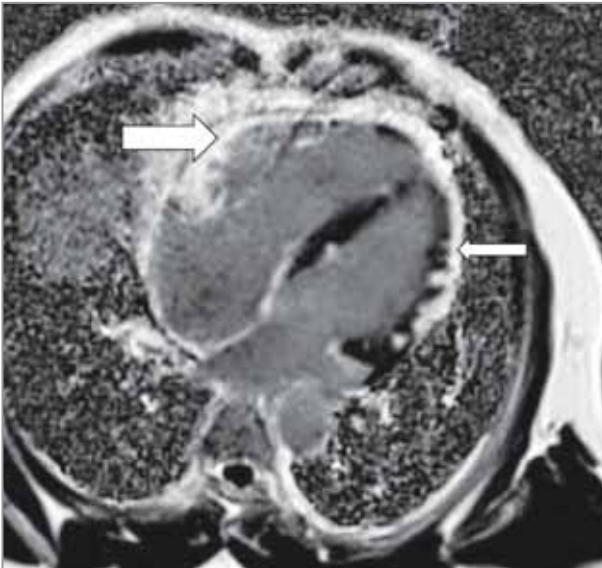


FIGURE 3: Cardiac MRI: 4-chamber view of the heart showing extensive late gadolinium enhancement of the right ventricular free wall (thick arrow) and epicardial nodular enhancement of the septum and left ventricle (thin arrow), consistent with cardiac sarcoidosis (figure courtesy of Dr Sulaiman Moosa).

of ARVC ranges from 0.9% - 25% in European and North American registries, with an overall prevalence of 13%.⁽³⁾ However, a high inter-observer variability exists in the assessment of epsilon waves with low agreement even among ARVC experts.⁽³⁾ A revised stricter definition of an epsilon wave as a "low amplitude deflection occurring at the end of the QRS complex which is defined as the latest end of the QRS complex in leads V1 - V4", has been proposed in order to achieve greater reproducibility in the assessment of epsilon waves. This study found that epsilon waves are almost always followed by T wave inversion.

The presence of an epsilon wave is usually a sign of late disease in ARVC and studies have shown that the presence of an epsilon wave has a low diagnostic impact as patients will usually fulfil other major diagnostic criteria. Therefore, finding an isolated epsilon wave in the absence of other ARVC criteria is very unusual and should prompt careful inspection of whether the epsilon wave is indeed present. Some have advised caution in the assessment of epsilon waves, especially in patients who are asymptomatic and who do not fulfil other ARVC criteria, and an epsilon wave should not be considered a major criterion in patients when the only other finding is a positive family history.⁽³⁾

This patient had conduction disturbances (1st degree AV block and left anterior fascicular block) and diffuse T wave inversion

in addition to an epsilon wave, which is unusual for ARVC. In one study comparing ARVC with sarcoidosis, 1st degree AV block was found in 0/42 (0%) patients with ARVC compared to 8/15 (53%) of patients with cardiac sarcoidosis.⁽⁴⁾ Epsilon waves were found in 2/15 (13%) patients with cardiac sarcoidosis in that study.

This patient was previously diagnosed with pulmonary sarcoidosis. A cardiac MRI confirmed biventricular systolic dysfunction (LVEF 33%, RVEF 11%) with septal and free wall scarring in keeping with cardiac sarcoidosis (Figure 3). His palpitations were subsequently confirmed to be due to ventricular tachycardia and he received an implantable cardioverter defibrillator for secondary prevention.

CONCLUSION

An epsilon wave is best seen in V1 - V3 and is a low amplitude wave that occurs at the end of the QRS complex or immediately after the QRS complex, and is almost always followed by an inverted T wave.

An epsilon wave is uncommon in ARVC cohorts and is a sign of late disease and should not be used as an isolated criterion in the absence of other diagnostic ARVC criteria.

Epsilon waves are not specific to the diagnosis of ARVC and can occur in a range of pathologies like sarcoidosis.

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Conflict of interest: none declared.