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The ECG in arrhythmogenic right ventricular cardiomyopathy: e-waves and anterior T-wave inversion

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DESCRIPTION

Arrhythmogenic right ventricular cardiomyopathy (ARVC) is an uncommon inherited cardiomyopathy caused by mutations of desmosomal protein genes responsible for cardiomyocyte electrical integrity and intercellular adhesion. ARVC is clinically characterised by:

- (1) electrical instability manifested by abnormalities of the resting ECG and ventricular arrhythmias. Sudden arrhythmic cardiac death is a recognised complication.
- (2) right ventricular systolic impairment, dilatation and/or regional wall motion abnormalities. Left ventricular involvement is increasingly been recognised. The electrical manifestations tend to precede the structural abnormalities.

The ECG illustrates the typical findings in ARVC: (1) anterior T-wave inversion in the right precordial leads (V1–V3), present in 85% and (2) ϵ -waves, present in 33% of ARVC patients. ϵ -Waves are reproducible low-amplitude signals occurring after the end of QRS complex and before the T-wave in the right precordial leads.

This patient had a maternal first-degree relative who was diagnosed with the condition having presented with aborted sudden cardiac death. Both the proband and the patient were found to have a desmosomal protein gene mutation (plakophilin). The patient also had right ventricular dilation and regional wall motion abnormalities

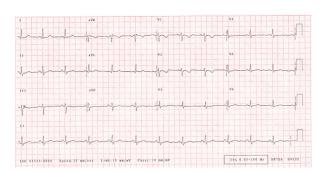


Figure 1 A standard 12-lead ECG showing T-wave inversion in leads V1 and V2.

with aneurysm formation and >1000 premature ventricular complexes in 24 h consistent with the diagnosis of ARVC.

ARVC is inherited in an autosomal dominant fashion, and first-degree relatives should be offered screening. The 12-lead ECG is an important diagnostic tool (figure 1). Double speed and amplitude traces are invaluable in the detection of ε -waves (figure 2).

This case illustrates the sensitivity of the ECG in detecting early electrical abnormalities in this condition.

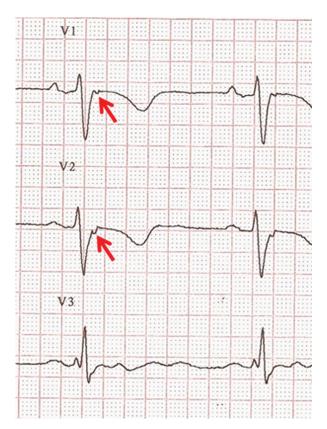


Figure 2 A double speed and amplitude 12-lead ECG (speed 50 mm/s, amplitude 20 mm/mV) demonstrate ε-waves (red arrows).

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Patient consent Obtained.

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