

Typical Electrocardiographic Features in Arrhythmogenic Left Ventricular Cardiomyopathy

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Short commentary

Since 2020 arrhythmogenic cardiomyopathy includes a right dominant form, a biventricular form and a left dominant form.

Already in 2004 Carvajal syndrome was first described in Ecuador within families [1]. This syndrome includes a slightly dilated, poor contracting left ventricle and hair and skin abnormalities with curly hair and plantopalmar keratosis. The genetic background were mutations of the desmoplakin gene.

Primarily it was thought to be a special form of dilated cardiomyopathy. But in ongoing years, with the help of cardiac MRI findings with severe fibrosis, the similarities with Naxos disease with similar aspects of hair and skin were more and more evident.

In 2020 Domenico Corrado characterized the term of arrhythmogenic left ventricular cardiomyopathy with extensive fibrosis of the left ventricle and ring-like appearance in cardiac MRI [2]. In many cases low voltage in limb leads and T-wave inversions in inferolateral leads was the characteristic findings in electrocardiography. Figure 1 revealed a typical example of these ECG changes due to a mutation in phospholamban.

Low voltage in limb leads is due to the fact that the left ventricle is poor contracting, T-wave inversions in inferolateral leads are often seen in biventricular or left dominant forms characterizing poor left ventricular function.

Another typical form of ECG in arrhythmogenic left ventricular cardiomyopathy is described by Calo [3,4] and is characterized by low voltage in limb leads, a very rare left posterior fascicular block both characterizing decrease in LV function, Q waves as electroanatomic scar and dominant shaped R wave in lead V1. The last feature is probably due to mutations of desmoglein-2, desmocollin-2 or in rare cases mutations in plakoglobin (Naxos disease) resembling right ventricular hypertrophy cause by myocyte cell necrosis [5].

Figure 2 revealed a typical example of these ECG changes.

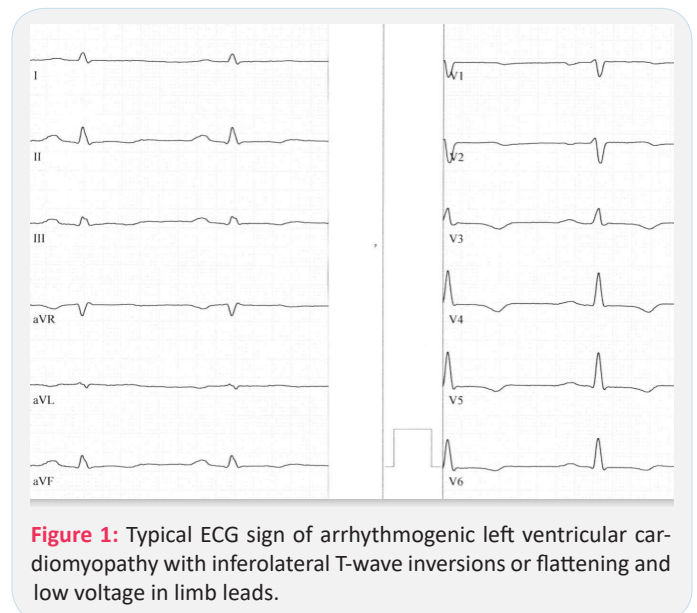


Figure 1: Typical ECG sign of arrhythmogenic left ventricular cardiomyopathy with inferolateral T-wave inversions or flattening and low voltage in limb leads.

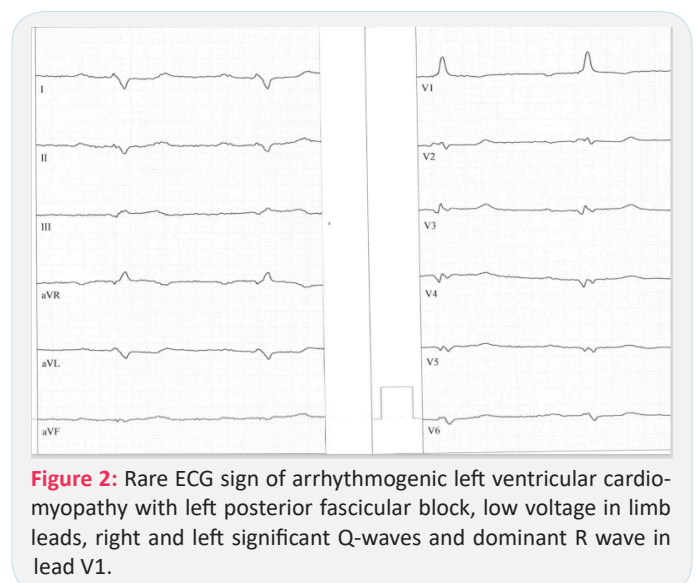


Figure 2: Rare ECG sign of arrhythmogenic left ventricular cardiomyopathy with left posterior fascicular block, low voltage in limb leads, right and left significant Q-waves and dominant R wave in lead V1.

In conclusion, most ECG's reveal low voltage in limb leads and T-wave inversions or flattening in inferolateral leads. This is especially true in desmoplakin [6], filamin C [7] phospholamban mutations [8], and TMEM43 [9] mutations causing arrhythmogenic left dominant cardiomyopathy with right bundle branch block – like ventricular tachycardia [10].

The second typical ECG of arrhythmogenic left ventricular cardiomyopathy is very rare form and is associated with desmoglein-2 [11] and desmocollin-2 mutations [12]. A positive R/S ratio in lead V1 characterize right ventricular hypertrophy due to right ventricular myocyte cell necrosis [5]. Left posterior fascicular block in combination with low voltage in limb leads characterizes poor left ventricular function. In comparison to desmoglein-2 positive right dominant arrhythmogenic cardiomyopathy typical right precordial epsilon waves and T-wave inversions are missing [13,14].

These two electrocardiographic features characterize arrhythmogenic left ventricular cardiomyopathy besides cardiac imaging especially cardiac MRI. The value of 12-lead ECG in arrhythmogenic left ventricular cardiomyopathy is outstanding.

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