Introduction

According to a presentation of Christina Basso, leading pathologist at the University Hospital of Padua, Italy, during the symposium on Myocardial and Pericardial diseases, held in Warnemünde, Germany, in October 2017, right ventricular dilatation is lacking in rare cases with arrhythmogenic cardiomyopathy. Rarely, right ventricles present without dilatation, aneurysms and wall thinning, but the typical presentation of fibrofatty abnormalities with myocardial atrophy in a normally seized and thickened right ventricle. In all cases the ECG revealed no typical abnormalities like right precordial T-wave inversions or epsilon waves in right precordial leads, but a Q wave of 3mm or more, an R wave of 2mm or less and an inverted T wave with an amplitude of 2mm or less in lead aVR. Additionally, the amplitude of inverted T wave of lead V1 was 2mm or more. These two leads represent typical right ventricular ones.

Method

We analyzed the ECG’s of 413 cases with arrhythmogenic cardiomyopathy (292 males, mean age 46.3 ± 11.6 years) at a paper speed of 50mm/sec. Patients conferred retrospectively to diagnostic criteria published by Frank Marcus in 2010 [1]. As a control group served a collective of normal probands (n=1596 patients, 859 males with an age range of 18-81 years) at a paper speed of 25mm/sec. Measurements were made by Elaine Clark by the same institution in Glasgow. Although specificity of typical changes in lead aVR Q-wave >= 3mm, small R-wave <= 2mm and T-wave inversions are, per se, moderate (81 and 86%, respectively), the addition of inverted T-waves of Q waves of 3mm or more, R waves of 2mm or less, inverted T waves of 2mm or less in lead aVR and inverted T waves in lead V1 were present.

Results

In 413 cases with arrhythmogenic cardiomyopathy (292 males, mean age 46.3 ± 11.6 years) epsilon waves in right precordial leads suggesting aneurysms in the right ventricular outflow tract
were detected in 95 cases (23%), T-wave inversions in right precordial leads were present in 227 cases (55%) and prolongation and epsilon waves- like abnormalities in inferior leads suggesting tricuspid-near inferior aneurysms were detected in 91 cases (22%). Dilatation of the right ventricle was not detected in 56 cases. At right ventricular angiography deep horizontal fissures in the right ventricular outflow tract could only be seen in 57 cases, whereas a mixture of deep horizontal fissures and bulgings were seen in the other cases. All patients were examined by ECG, transthoracic echocardiography, coronary angiography, left ventricular angiography and right ventricular angiography. Echocardiography revealed in all cases localised dilatation of the right ventricular outflow tract, right ventricular inflow tract, or bulging of the right ventricular apex published by Frank Marcus 2010 [1]. Magnetic resonance imaging was not performed in any cases. The ECG measurements in cases with arrhythmogenic cardiomyopathy were made by the author himself, the measurements of normal probands were made by the Elaine Clark of the team of Prof. Macfarlane (Figure 1).

If lead aVR criteria of large Q waves of 3mm or more, small R waves of 2mm or less and negative T-waves in the Glasgow collective were combined to an amplitude of negative T-waves in lead aVR of 2mm or less and compared to data in arrhythmogenic cardiomyopathy sensitivity was 94%, specificity 97%, positive predictive value 88% and negative predictive value was 97%. These results were calculated elsewhere and published [2,3] (Figure 2).

If additionally combined to an amplitude of negative T-waves in lead V1 of 2mm or more the statistical analysis was as follows:

<table>
<thead>
<tr>
<th>Table 1: ECG parameters in cases with ARVC and normal probands.</th>
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<td>Positive findings</td>
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<td>ARVC n=385</td>
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<tr>
<td>Amplitude inverted T-wave &gt;= 2mm in lead V1</td>
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<td>Normal probands n=49</td>
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Note: Q wave >= 3mm, R wave <= 2mm and inverted T-wave in lead aVR + Amplitude of inverted T-wave <= 2mm in lead V1.

Discussion

According to the results of the Glasgow collective the ECG of apparently normal hearts with a fibrofatty focus without right ventricular dilatation and without right ventricular aneurysms should be analyzed as follows:

With the two components of lead aVR (large Q waves of 3mm or more, R waves of 2mm or less [2] and an amplitude of negative T-waves of 2mm or less [3] sensitivity is 94%. These results can be further enhanced by analysis of the amplitude of negative T-waves of 2mm or more in lead V1 with a sensitivity of 97% [4]. An amplitude of inverted T waves of 2mm or more could be described in cases with unclassifiable arrhythmogenic cardiomyopathy associated with Emery-Dreifuss caused by a mutation in FHL with typical fibrolipomatosis of the right ventricle and hypertrabeculation of the left ventricle [5].

Nevertheless, right precordial T-waves inversions and epsilon waves are major ECG criteria of all types of arrhythmogenic cardiomyopathy and in cases with dilated right ventricles and right ventricular aneurysms.

In cases of arrhythmogenic cardiomyopathy without dilatation and aneurysms ECG analysis should be directed to the amplitude of inverted T-waves in lead V1 and its exclusion in lead aVR.

The simple means of standard ECG remains a relevant finding in the diagnosis of arrhythmogenic cardiomyopathy in spite of specific cardiac imaging techniques like nuclear magnetic resonance technique [6] or right ventricular angiography [7]. In the future, bipolar ECG, could be a technique, especially for lead V1, to improve outcomes [8].

In pathologic features of arrhythmogenic cardiomyopathy without right ventricular dilatation, wall thinning and aneurysms the ECG presents with highly typical features in lead aVR and lead V1. Epsilon waves and right precordial T wave inversions are major criteria of arrhythmogenic cardiomyopathy, but lead aVR and V1 are more relevant in cases without right ventricular dilatation and right ventricular aneurysms. In the present study lead aVR and V1 are far more sensitive than T-wave inversions in right precordial leads and epsilon waves in all types of arrhythmogenic cardiomyopathy.

Limitations

The only limitation of the study was the fact that cardiac MRI...
was not used, as the patients were detected as having arrhythmogenic cardiomyopathy long ago from 1985 on where MRI technique was infrequently used. In fact, MRI in the present form is superior to echocardiography. Nevertheless, there are lot of centers who use right ventricular angiography and echocardiography instead of MRI technique.

There is definitely bias in the measurement of ARVC patients, but the same is true in the measurements in the control group.

References


