Influence Of Novel Electrocardiographic Features Of Provocable Brugada ECG In Arrhythmogenic Cardiomyopathy And Its Exclusion By Lead AVR

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Abstract
In 19 patients (14 females, mean age 49.1 ± 11.3 years) with typical arrhythmogenic cardiomyopathy and provokable type I Brugada ECG pattern by ajmaline administration were analysed by novel electrocardiographic features as having “true” or “false” Brugada syndrome. Three patients turned out as having false Brugada syndrome, the diagnosis is pure arrhythmogenic cardiomyopathy. In 16 patients, however, true Brugada syndrome could be provoked. In these patients the diagnosis was arrhythmogenic cardiomyopathy associated by provokable Brugada syndrome.

Introduction
In a large collective of 385 patients (212 males, mean age 46.3 ± 11.1 years) with typical arrhythmogenic cardiomyopathy lead aVR was analysed. A morphology of large Q wave of 3mm or more, a small R wave of 2mm or less, and T-wave inversion turned out to be the best predictor of arrhythmogenic cardiomyopathy.

In 1498 healthy probands (859 males in an age range of 18 – 85 years) the same morphologic parameters were analysed. Similar results were obtained in 284 probands (18.9%). Specificity and positive predictive value were low, but negative predictive value was nearly 100%.

An association between arrhythmogenic cardiomyopathy and Brugada syndrome seems to be a matter of fact.1, 2, 3
A continuum between these both diseases has been described.4 Causal gene mutations have been confirmed in plakophilin-2,5 desmoglein-26 and desmoplakin.7

To differentiate true or false provokable Brugada syndrome novel electrocardiographic features have been presented8 as follows:
- concave (coved) ST-segment morphology with negative symmetrical T-waves
- ST-segment morphology shows progressive decline
- the ratio between the peak height of QRS-ST after 80ms is greater than 1
- the duration of the QRS in leads V1 and V2 is greater than in the middle and left precordial leads
- type-1 Brugada syndrome ECG may be seen in a single lead, V1 or V2, but never exclusively in V3

We analysed 19 patients (14 females, mean age 49.1 ± 11.3 years) with typical diagnosis of arrhythmogenic cardiomyopathy and provokable Brugada syndrome by ajmaline administration.

In three patients without novel electrocardiographic criteria we could rule out true provokable type I Brugada –ECG pattern. These patients ended up in the diagnosis of pure arrhythmogenic cardiomyopathy.

In 16 patients with novel electrocardiographic criteria demonstrated true provokable type I Brugada ECG pattern. These patients ended up in a combination of arrhythmogenic cardiomyopathy and Brugada syndrome supporting the continuum between these two cardiac entities.

In order to diagnose or to exclude arrhythmogenic cardiomyopathy we like to focus the interest to lead aVR. Lead aVR is the only lead which points directly to the right ventricle.

In a large collective of 385 patients (212 males, mean age 46.3 ± 11.1 years) with typical arrhythmogenic cardiomyopathy the morphology of lead aVR was analysed.

In 97% of cases large Q wave of 3mm or more, small R wave of 2mm or less and T-wave inversion were found. In a control collective of the University Hospital of Glasgow, U.K. (Prof. Peter Macfarlane, Cardiology and Electrocardiography) of 1498 probands (859 males in an age range of 18 – 85 years) the same morphologic parameters were analysed. Similar results were obtained in 284 healthy probands.
Specificity and positive predictive value were low, but negative predictive value was nearly 100% (9).

Lead aVR is an excellent tool to exclude arrhythmogenic cardiomyopathy as demonstrated in different publications.\textsuperscript{10,11}

**Conclusions**

In conclusion, there are new tools to confirm provocable true Brugada syndrome in arrhythmogenic cardiomyopathy to reveal a continuum between these two entities and to definitely exclude arrhythmogenic cardiomyopathy by electrocardiographic means.

**References**

10. Peters, S. Electrocardiography not always confirm arrhythmogenic right ventricular cardiomyopathy – the value of lead aVR. Int J Cardiol 2014; 173: e34 - 5
11. Peters, S. Confusion with the diagnosis of acute pulmonary embolism and arrhythmogenic right ventricular cardiomyopathy. Int J Cardiol 2016; 203: 317