International Journal of Cardiology 196 (2015) 38–41

Contents lists available at ScienceDirect

International Journal of Cardiology

journal homepage: www.elsevier.com/locate/ijcard

Letter to the Editor

Electrocardiographic characteristics of arrhythmogenic right ventricular dysplasia, cardiac sarcoidosis and arrhythmogenic biventricular cardiomyopathy

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ARTICLE INFO

Article history:
Received 6 May 2015
Accepted 27 May 2015
Available online 3 June 2015

Keywords:
Cardiac sarcoidosis
Arrhythmogenic right ventricular cardiomyopathy
Arrhythmogenic biventricular cardiomyopathy
QRS fragmentation

Complete right bundle branch block with localised right precordial QRS prolongation has been published under the term "more than complete right bundle branch block" by Guy Fontaine and coworkers [1]. This ECG criterion defines arrhythmogenic right ventricular dysplasia (cardiomyopathy), cardiac sarcoidosis or arrhythmogenic biventricular cardiomyopathy.

In 391 patients with typical arrhythmogenic cardiomyopathy (214 males, mean age 46.1 ± 11.3 years) 24 patients had the ECG finding of more than complete right bundle branch block.

5 patients had more than complete right bundle branch block at initial diagnosis of arrhythmogenic right ventricular cardiomyopathy. These patients suffered from frequent ventricular premature beats and non-sustained ventricular tachycardia. Left ventricular function was normal. It could not be excluded with certainty if arrhythmogenic right ventricular cardiomyopathy was associated with cardiac sarcoidosis. Without QRS fragmentation and epsilon wave in lead aVR the clinical course was uneventful.

One patient with more than complete right bundle branch block suffered from recurrent ventricular tachycardia. In the university clinic in Münster, Germany, myocardial biopsy revealed cardiac sarcoidosis. Left ventricular function was reduced with an EF of 50%. ICD implantation was performed.

In the 18 remaining patients more than complete right bundle branch block developed at a time interval of 3 to 6 months. In all cases right ventricular abnormalities with typical electrocardiographic signs of arrhythmogenic cardiomyopathy together with recurrent ventricular tachycardia were found. After development of complete right bundle branch block left ventricular function deteriorated and heart failure symptoms were more and more evident. In these 18 patients QRS fragmentation especially in right precordial leads and epsilon wave in lead aVR were found. In three patients despite optimal medical therapy heart transplantation was necessary.

In the first scenario – arrhythmogenic right ventricular dysplasia/cardiomyopathy – more than complete right bundle branch block appears without QRS fragmentation in right precordial leads and without epsilon waves in lead aVR (Fig. 1). This ECG criterion is the dominant finding when the diagnosis is settled as pure right ventricular dilatation and reduced function. Left ventricle appears to be normal.

In the second scenario – cardiac sarcoidosis – more than complete right bundle branch block is the prominent ECG finding in a few cases which can be hardly differentiated from arrhythmogenic right ventricular cardiomyopathy. In most cases left ventricular function is somewhat reduced [2]. Sardiac sarcoidosis is besides coronary artery disease and dilated cardiomyopathy the third frequent cause for heart transplantation.

In the third scenario – arrhythmogenic biventricular cardiomyopathy – more than complete right bundle branch block develops in the course of arrhythmogenic right ventricular cardiomyopathy with progressive left ventricular involvement [3,4]. The developing ECG finding is characterised by QRS fragmentation mainly in right precordial leads (Fig. 2) [4] and epsilon waves in lead aVR (Fig. 3) [5]. This form of arrhythmogenic cardiomyopathy is characterised by recurrent ventricular tachycardia or ventricular fibrillation and by the development of therapy-resistant heart failure often making heart transplantation necessary.

When cardiac sarcoidosis is found by myocardial biopsy pathologists observing the pathognomonic granulomatous pattern are satisfied to establish the diagnosis of sarcoidosis without paying attention to particular histologic structure of desmosomes, which requires additional immunostaining. Sarcoidosis can be superimposed on arrhythmogenic right ventricular cardiomyopathy [6].

With the help of ECG findings arrhythmogenic right ventricular cardiomyopathy, sarcoidosis possibly superimposed on ARVC and arrhythmogenic biventricular cardiomyopathy can be differentiated by more than complete right bundle branch block.

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http://dx.doi.org/10.1016/j.ijcard.2015.05.176

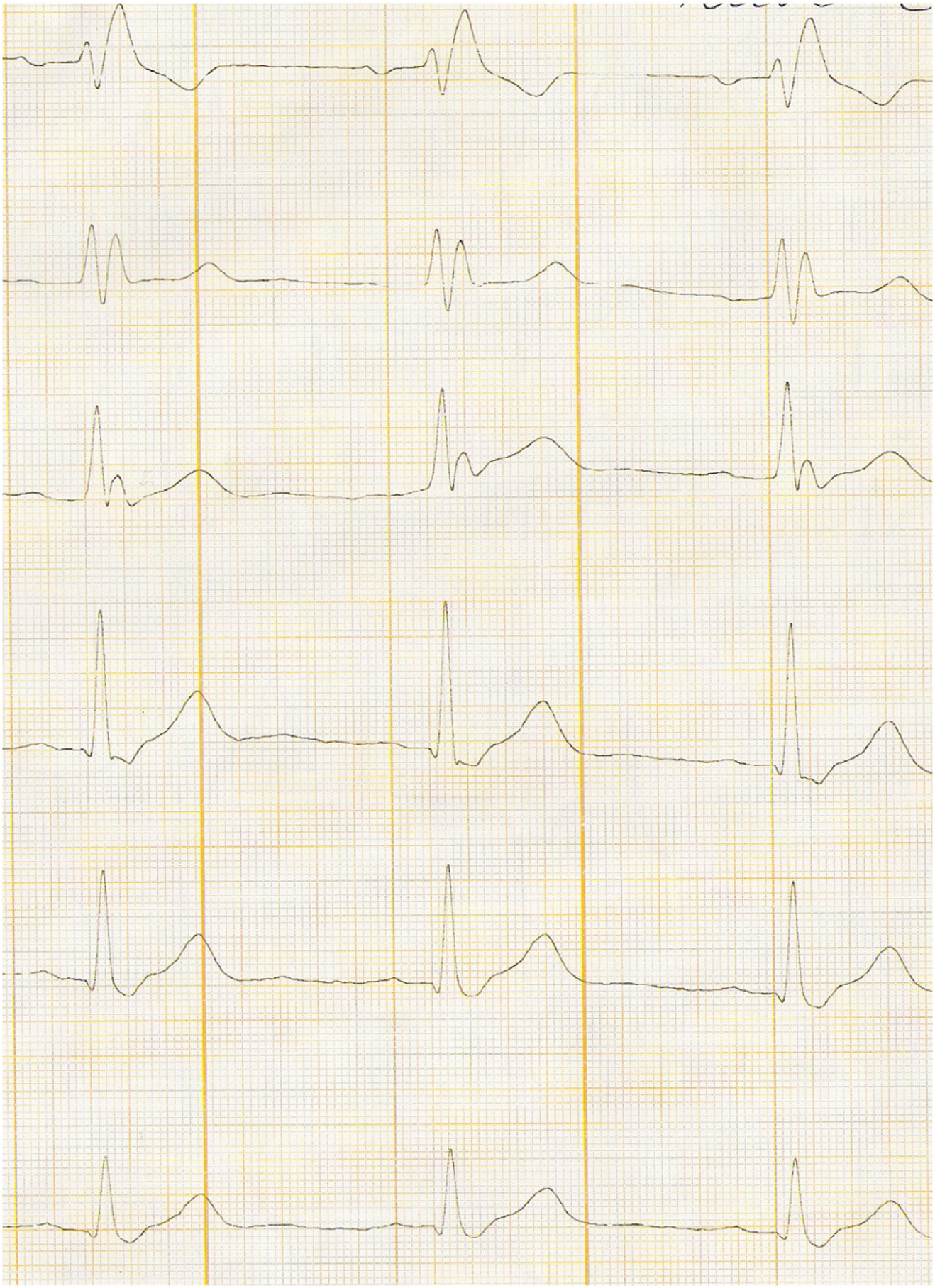

Fig. 1. More than complete right bundle branch block without QRS fragmentation and epsilon waves in lead aVR.