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Additional Patients With Brugada Syndrome And Associated Shorter-Than- Normal QT Syndrome

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ABSTRACT

Introduction: Only a few patients with Brugada syndrome and associated shorter-than-normal QT syndrome have been described by Charles Antzelevitch 2007. The association is reported in a few gene mutations such as SCN5A and CACNA1C, CACNB2b and CACNA2D1. Antzelevitch reported on three patients with typical ECG signs of Brugada syndrome, in one patient provoked by ajmaline. All patients had aborted sudden cardiac death.

Method: Additionally, we reported on three more patients, all provoked by ajmaline, with the same phenomenon with typical ECG signs of Brugada syndrome and shorter-than-normal QT syndrome with a QTc interval of 340 to 360 msec. In a young patient with syncope SCN5A mutation was found. In another case with aborted sudden cardiac death so far no mutation could be found. The third patient was withdrawn from blood samples as he migrated to the isle of fuerteventura.

Results: The risk of sudden cardiac death in Brugada syndrome is about 1 %, the risk of sudden cardiac death in short QT syndrome 0.8-1%. The risk of sudden cardiac death in Brugada syndrome and shorter than normal QT overlap syndrome is 66% due to a very low number (n=4/6) of patients.

Conclusions: The combination of Brugada syndrome and shorter than normal QT interval bears to extremely increased risk of sudden cardiac death due to a very low number of patients.

CASE SUMMARY

Only a few patients with Brugada syndrome and associated shorter-than-normal QT syndrome have been described by Charles Antzelevitch [1]. The association is reported in a few gene mutations such as SCN5A and CACNA1C. Antzelevitch reported on three patients with typical ECG signs of Brugada syndrome, in one patient provoked by ajmaline. In the Journal of Kardiologie Peters reported three other patients, all provoked by ajmaline, with the same phenomenon with typical ECG signs of Brugada syndrome and shorter-than-normal QT syndrome with a QTc interval of

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340 to 360 msec [2]. In a young patient SCN5A mutation was found (Figure 1). In another case so far no mutation could be found (Fiigure 2). The third patient was withdrawn from blood samples as he migrated to the isle of fuerteventura (Figure 3). The genetic analysis was done by Antzelevitch himself.

The phenomenon of associated Brugada syndrome and shorter than normal QT syndrome occurs extremely seldom. It is reported in patients with sudden cardiac death (1) and in atrial arrhythmias (2). Atrial arrhythmias occurs in CACNA1C mutations in a simulation study (2), sudden cardiac death occurs in CACNA1C mutations and in CACNB2b and in CACNA2D1 mutations [1]. In CACNA1C atrial arrhythmias could be found in a simulation study (2). But the ECG before provocation and the ECG after provocation show the reduction of QT interval. An effect of ajmaline could be excluded.

What is more important is the fact that this phenomenon is associated with sudden cardiac death. There are 4 patients with sudden cardiac death, one with syncope and an asymptomatic case with atypical chest pain. There are several patients with Brugada syndrome and sudden cardiac death and more than 70 patients with short QT interval and sudden cardiac death. In Brugada syndrome and shorter than normal QT interval there are only 4 patients. The rate of sudden cardiac death in Brugada syndrome – either in asymptomatic or highly symptomatic ones, either ajmaline provoked or not provoked -is about 1 % [3,4], in short QT syndrome about 0.8 % [5], but in the combination of Brugada syndrome and shorter than normal QT interval 66%. In this respect, the Brugada syndrome and shorter than normal QT interval overlap syndrome has the highest rate of sudden cardiac death due to a very low number of patients (n=4/6).

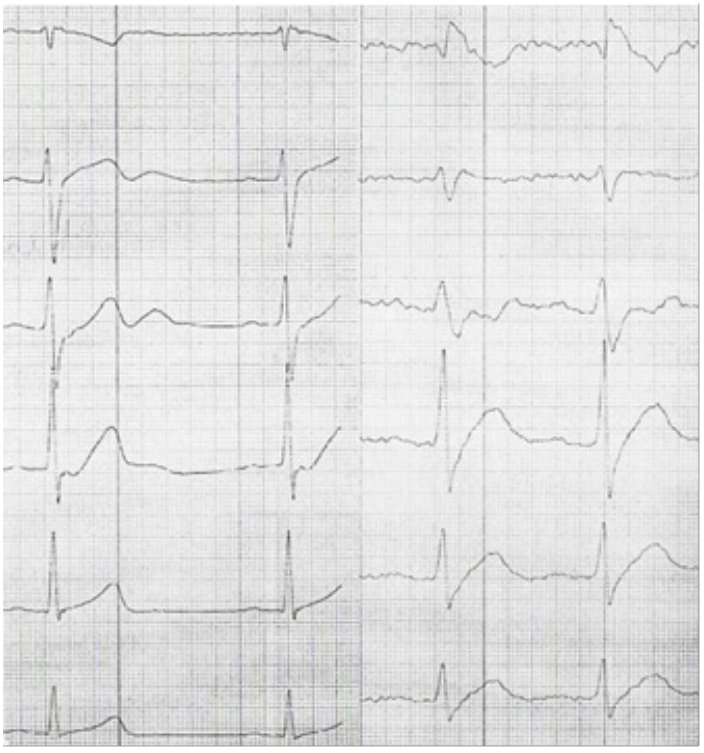

Figure 1. The ECG of the patient with SCN5A mutation

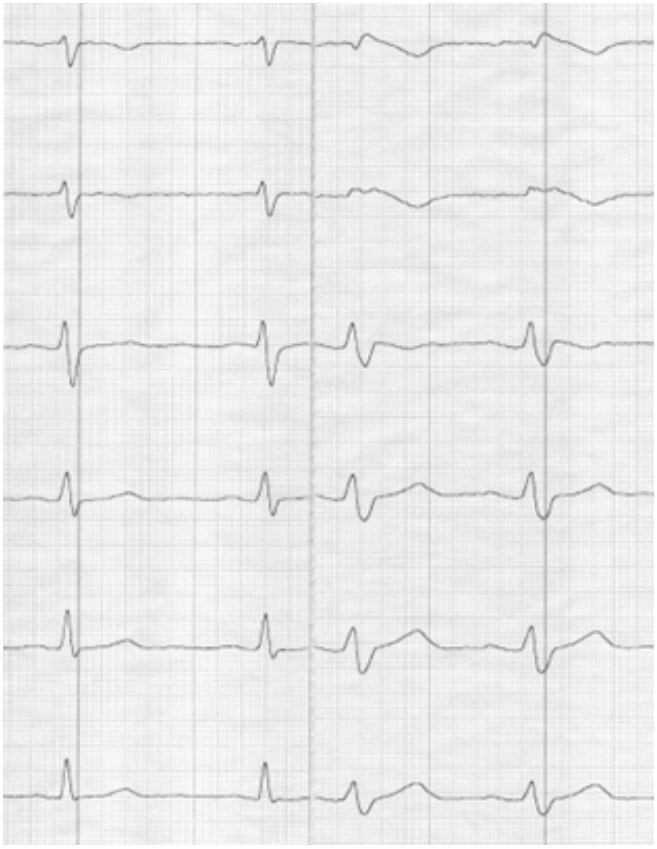

Figure 2. The ECG of the patient with unknown mutation

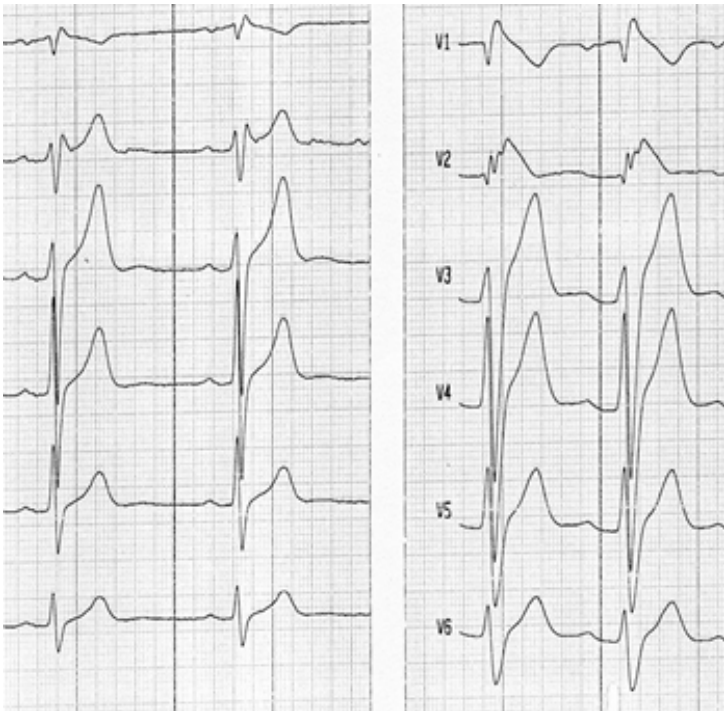

Figure 3. The ECG of the patient excluced form blood samples

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