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Long-QT Syndrome, Brugada Syndrome, and Catecholaminergic Polymorphic Ventricular Tachycardia

A Tale of 3 Diseases

I have read the paper by Havakuk and Viskin (1) with great interest, and congratulate the authors on their excellent work. As the authors correctly state, both long-QT syndrome and Brugada syndrome may lead to polymorphic ventricular tachyarrhythmias. However I would like to call attention to another disease: catecholaminergic polymorphic ventricular tachycardia (CPVT). CPVT is characterized by exercise- or stress-induced ventricular tachyarrhythmias, leading to syncope or sudden cardiac death, and it also shares important characteristics.

First, it presents as a congenital or acquired (mainly drug-induced) arrhythmogenic disorder. Disease-causing mutations in ryanodine receptor (RyR2) in autosomal dominant form or calsequestrin 2 genes (*CASQ2*) in recessive form have been identified in most of affected patients (2). An inducing role of β-adrenergic agonists has been demonstrated. Notably, in *CASQ2* CPVT2 patients, a higher risk for cocaine cardiotoxicity has been shown as well (3).

Second, the electrocardiogram at rest is normal (2) but a pathologic early repolarization pattern is present in an unexpected large proportion (45%) of patients and it is associated with an increased frequency of syncope. In patients with unexplained syncope, an exercise testing should be performed to detect CPVT (4).

Third, an evolution of therapeutic approaches has been shown.

Initially, beta-blockers were considered the mainstay therapy for CPVT (2). Recently, flecainide appears to be effective have a role in modulation of intracellular calcium in all CPVT patients. For nonresponders to drug therapy or in patients after a life-threatening arrhythmia, even an implantable cardioverter-defibrillator is needed. Left cardiac sympathetic denervation was reported to be effective in patients with intractable RyR2 mutation-associated CPVT (2). The history of CPVT (5) retraced the same steps of the 2 arrhythmias described in the article of Havakuk and Viskin (1) and I would have expected that the authors had also shown the tale of CPVT in addition to the tale of Brugada syndrome and long-QT syndrome.

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