Palpitations in a 19-year-old man, take a second look

A. A. M. Wilde

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Answer to rhythm puzzle

It is clear that there is atrial flutter with a variable AV block (2:1–3:1) leading to a ventricular rate of 90–120 beats per min. The atrial flutter cycle length (CL) is 260 msec (Fig. 1). The QRS complex has a rightward axis and is 120 ms wide. The ST-T segments seem normal.

The combination of findings is obviously unusual in a 19-year-old man. First of all, a typical atrial flutter cycle length is 200 msec (instead of 260 msec) leading to an atrial rate of 300 beats/min. Moreover, in the case of an atrial flutter there usually is a 2:1 block leading to a ventricular rate of 150 beats/min. Hence, in this case the atrial flutter rate is lower than expected and so is

Fig. 1 ECG at presentation with in the inset an enlargement of lead II

A. A. M. Wilde (✉)
Department of Cardiology, Academic Medical Centre,
Meibergdreef 9,
1105 AZ Amsterdam, the Netherlands
e-mail: a.a.wilde@amc.uva.nl

the ventricular rate due to hampered AV conduction. Finally, also the conduction in the ventricles is slower than normal as can be judged from the rightward axis and the broad QRS complex (120 msec). Taken together, conduction of the cardiac impulse is slower than expected in all cardiac compartments (atrium, AV node and ventricle).

The family history, with a pacemaker implant at age 42 in his mother's brother, is compatible with an inherited cause of cardiac conduction disease. Indeed, molecular genetic testing revealed the presence of a mutation in the SCN5a gene, the gene encoding for the cardiac sodium channel.

Cardiac sodium channel function is crucial for proper conduction of the cardiac impulse at all cardiac levels and loss-of-function mutations give rise to a variety of familial syndromes, including familial conduction disease [1].

Reference

1. Wilde AAM, Brugada R. Phenotypical manifestations of mutations in the genes encoding subunits of the cardiac sodium channel. *Circ Res.* 2011;108:884–97.