Palpitations in a 19-year old man and a slow heart rate at rest

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Answer to rhythm puzzle

This is clearly an abnormal ECG for a 19-year-old man. Initially there is sinus rhythm with a rate that is slightly under 60 beats/min. With 210 msec the PQ interval is prolonged. The QRS complex has a rightward axis and is 120 msec wide. The ST-T segments seem normal. After 2 sinus complexes a pause follows that in length roughly equals 2× the PP interval of the previous 2 sinus complexes. The next, also long, RR interval is roughly similar in length. Thereafter sinus rhythm, slightly irregular, with 1:1 conduction resumes during 3 complexes.

As stated, the combination of findings is obviously unusual in a 19-year-old man. At all cardiac levels (atrium, AV node/conduction system, ventricle) conduction of the electrical impulse is hampered. The pauses are most likely caused by exit block out of the sinus node (i.e. SA block), another level at which conduction is hampered. And the combined findings, including the family history, suggest the presence of a loss-of-function sodium channel mutation, which was indeed found. 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]. SA block is also frequently observed in this setting.

In a recent rhythm puzzle [2], the ECG of the same patient, during atrial flutter, the cause of his complaints of palpitations, was presented. Also on this ECG all conduction parameters were decreased leading to an unusual flutter rate and a higher than expected degree of AV block.

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References

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