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ECG presentation of the LQT2 syndrome (HERG gene mutation)

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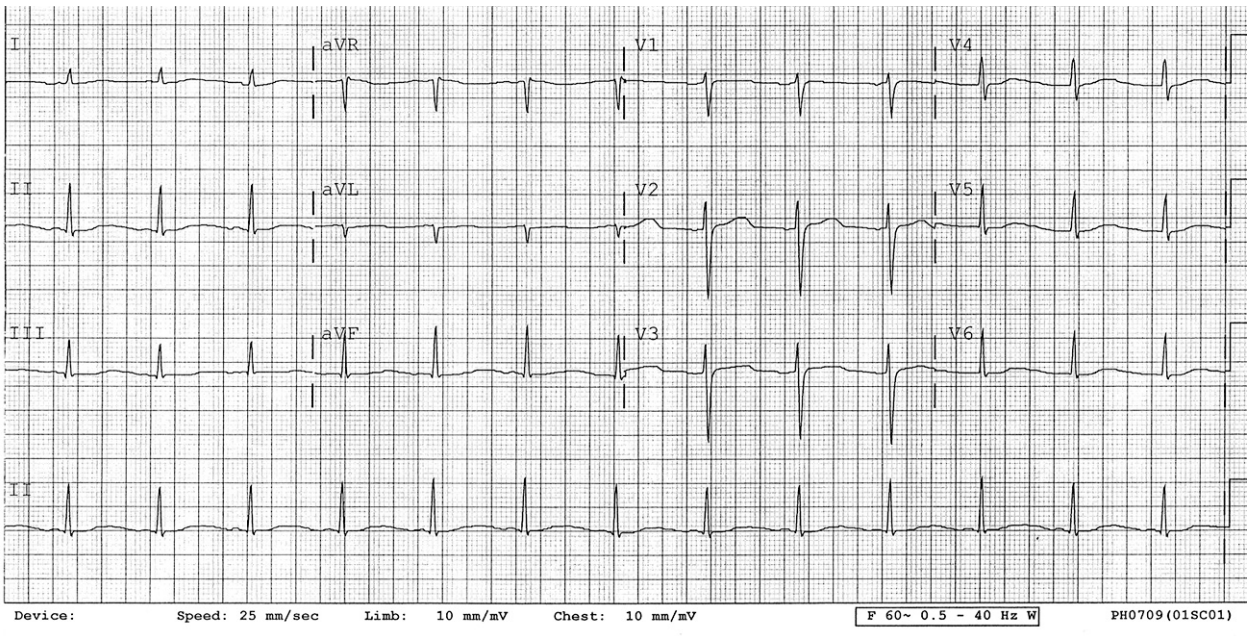
This is an ECG of a 28-year old woman with history of syncopal episodes and documented torsade de pointes ventricular tachycardia. First syncopal episode occurred at age 16 during basket ball game. She has a positive history of sudden cardiac death.

ECG shows substantially prolonged QT (QTc between 0.46–0.66 s depending on lead) with presence of flat and notched T wave. The T wave morphology is characteristic for the long QT syndrome caused by the HERG gene mutation (LQT2), gene regulating function of the I_{Kr} delayed rectifier outward potassium current in myocardial cell. Patients with LQT2 frequently have they cardiac events triggered by emotion, stress, or exercise. Beta-blockers are standard of treatment in LQTS patients including LQT2, but they do not fully protect high-risk patients who experience recurrent arrhythmic events. In such cases, an implantable cardioverter-defibrillator is considered.



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