T-wave alternans in a patient with long-QT syndrome type 3

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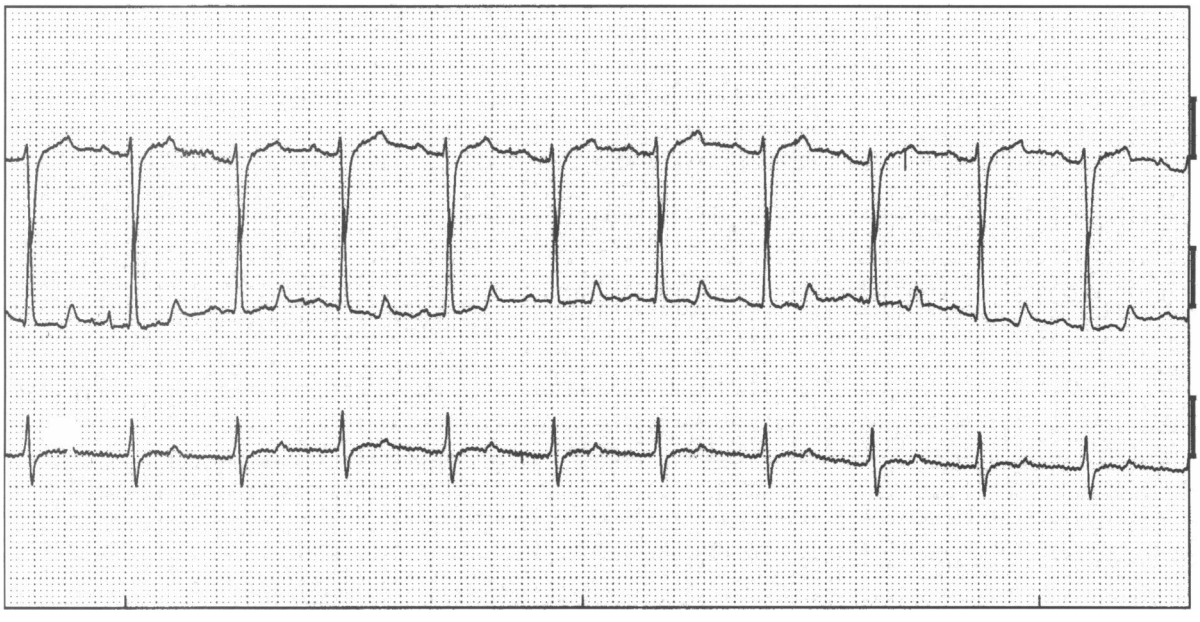

Figure 1.

A healthy 71-year-old male was investigated because he was a member of a previously reported large family with long-QT syndrome type 3 and carrier of the mutant gene (SCN5A, 1795insD).[1] A 24-hour Holter recording (modified leads V_1, V_5, aVF, paper speed 25 mm/sec) showed bradycardia-dependent prolongation of the QT interval. At a relatively high heart rate the QT interval was within normal limits (QTc 0.43 sec), and the repolarisation abnormality was confined to the presence of a narrow pointed T wave (figure 1), but at lower heart rates progressive repolarisation abnormalities became apparent (figure 2), which amounted to excessive QT prolongation during the night with QTc values as long as 0.80 sec (figure 3). In addition, the recording showed T wave alternans, biphasic T waves being alternated by giant negative T waves. Excessive bradycardia-dependent QT prolongation is an established feature of LQT3, but the findings in this patient are extreme and T wave alternans has not been reported before in clinical LQT3.

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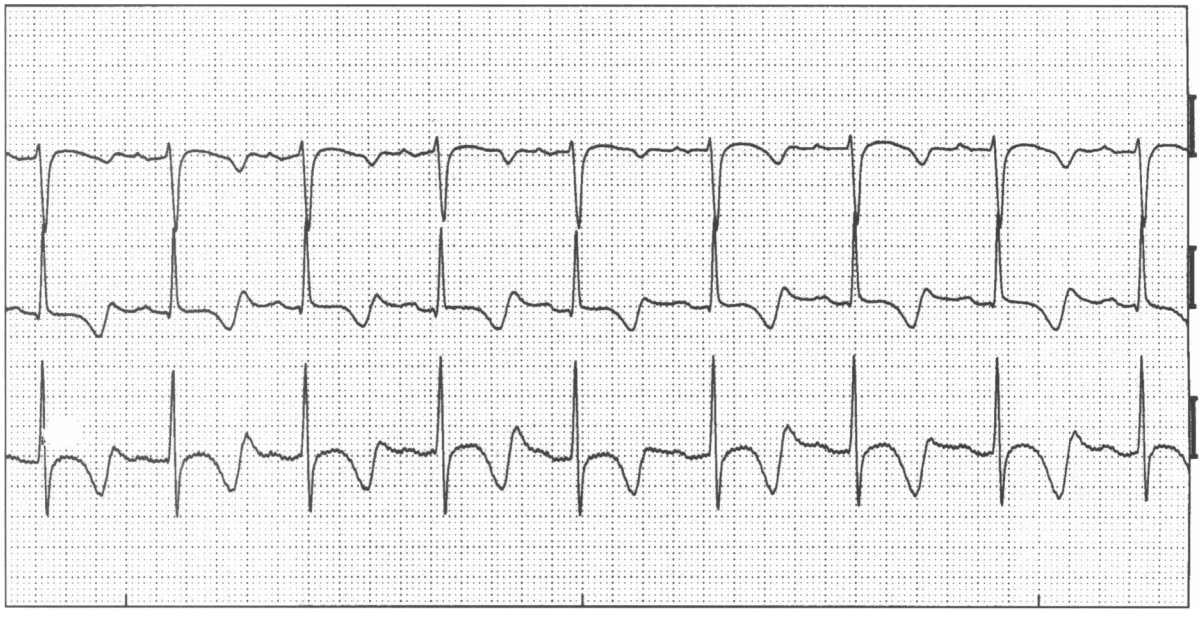

Figure 2.

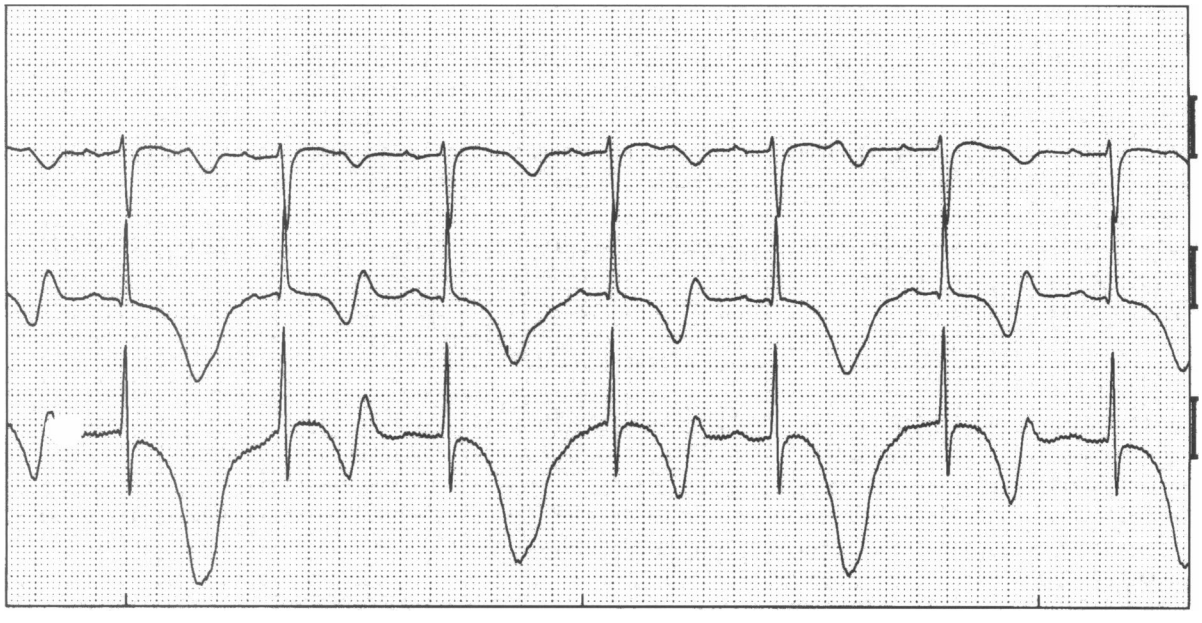

Figure 3.

Of interest, T wave alternans is associated with an increased propensity for ventricular arrhythmias, in particular torsade de pointes,[2] but we did not observe any ventricular arrhythmias other than isolated ventricular ectopic beats in this patient or in any of the other affected members of this large family (n>100), despite very extensive Holter monitoring. In fact, there even appears to be a paucity of ventricular ectopic beats.[3] These observations raise the question as to the mode of death in this family, but the answer to this question still remains elusive. ■

References

1 Van den Berg MP, Viersma JW, Beaufort-Krol GCM, Bink-Boelkens MTE, Bezzina CR, Veldkamp MW, et al. A large family characterized by nocturnal sudden death. *Neth Heart J* 2002; **10**:304-12.

2 Rosenbaum DS, Jackson LE, Smith JM, Garan H, Ruskin JN, Cohen RJ. Electrical alternans and vulnerability to ventricular arrhythmias. *N Engl J Med* 1994;**330**:235-41.

3 Van den Berg MP, Wilde AAM, Viersma JW, Brouwer J, Haaksma J, van der Hout AH, et al. Possible bradycardic mode of death and successful pacemaker treatment in a large family with features of both long QT syndrome type 3 and Brugada syndrome. *J Cardiovasc Electrophysiol* 2001;**12**:630-6.