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Diagnostic value of epinephrine test for genotyping LQT1, LQT2, and LQT3 forms of congenital long QT syndrome

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ABSTRACT
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Objectives
The aim of this study was to test the hypothesis that epinephrine test may have diagnostic value for genotyping LQT1, LQT2, and LQT3 forms of congenital long QT syndrome (LQTS).

Background
A differential response of dynamic QT interval to epinephrine infusion between LQT1, LQT2, and LQT3 syndromes has been reported, indicating the potential diagnostic value of the epinephrine test for genotyping the three forms.

Methods
The responses of 12-lead ECG parameters to epinephrine were retrospectively examined in 15 LQT1, 10 LQT2, 8 LQT3, and 10 healthy volunteers to select the best ECG criteria for separating the four groups. The epinephrine test then was prospectively conducted in 42 probands clinically affected with LQTS, their 67 family members, and 10 new volunteers. The best criteria were applied in a blinded fashion to prospectively separate a different group of 31 LQT1, 23 LQT2, 6 LQT3, and 30 Control patients (10 genotype-negative LQT1, 10 genotype-negative LQT2 family members, and 10 volunteers).

Results
The sensitivity (penetrance) by ECG diagnostic criteria was lower in LQT1 (68%) than in LQT2 (83%) or LQT3 (83%) before epinephrine and was improved with steady-state epinephrine in LQT1 (87%) and LQT2 (91%) but not in LQT3 (83%), without the expense of specificity (100%). The sensitivity and specificity to differentiate LQT1 from LQT2 were 97% and 96%, those from LQT3 were 97% and 100%, and those from Control were 97% and 100%, respectively, when Δ mean corrected Q-Tend ≥35ms at steady state was used. The sensitivity and specificity to differentiate LQT2 from LQT3 or Control were 100% and 100%, respectively, when Δ mean corrected Q-Tend ≥80ms at peak was used.

Conclusions
Epinephrine infusion is a powerful test to predict the genotype of LQT1, LQT2, and LQT3 syndromes as well as to improve the clinical diagnosis of genotype-positive patients, especially those with LQT1 syndrome.

Key words: Arrhythmia, Diagnosis, Long QT syndrome, Catecholamines, Genes

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