

Diagnostic reproducibility of epinephrine drug challenge interpretation in suspected long QT syndrome

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Introduction: congenital long QT syndrome (LQTS) is a channelopathy affecting 1/2000 patients and responsible for sudden cardiac death. Screening for this heart disease is based on the measurement of the corrected QT interval (QTc) on the electrocardiogram. However, QTc measurement is sometimes contentious, and may be misleading in patients with concealed long QT syndrome (with normal range QTc). The epinephrine challenge has been proposed to improve the diagnosis of congenital long-QT syndrome (LQTS).

Aim: to evaluate diagnostic reliability of the epinephrine provocative test for LQTS diagnosis, taking into consideration of intra and inter-observer variability in the interpretation of the test.

Methods: a retrospective analysis of 79 consecutive epinephrine provocative tests was conducted. Epinephrine was administered following a standardized protocol at 2 doses: 0.05 µg/kg/min and 0.10 µg/kg/min. ECGs were blindly read twice by three different operators at ≥ 1 week interval. QT and RR intervals were collected at rest and at each dose, as well as final operator interpretation of the test.

Results: there was a high inter-observer reproducibility of corrected QT measurements with an intra-class correlation (ICC) of 0.74 (IC 95% 0.66-0.80) but a low inter-observer reproducibility on the final interpretation with a Kappa of 0.31. Intra-observer reproducibility of corrected QT was very good (ICC 0.93;0.91-0.95), but still resulted in an only moderate intra-observer reproducibility in the final diagnosis (Kappa of 0.47). Perceived certainty of at least 1 reading by 2 operators (N=62 tests) moderately increased inter-observer reproducibility compared to baseline (Kappa=0.43).

Conclusion: inter and intra-observer agreement in the interpretation of the epinephrine provocation test for long QT syndrome is poor to modest. Complexity in interpretation varies from one case to the next. The low reliability of this test encourages a reconsideration of its importance in the clinical management of patients with suspected LQTS.