"QT Stunning": An Abnormal QT Response in Patients with Long QT Syndrome Uncovered by a Bedside Test

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Background: The response of the QT interval to sudden changes in heart rate is not instantaneous. Taking advantage of this phenomenon, we recently showed that the sinus tachycardia that follows brisk standing exposes abnormal QT-interval prolongation in patients with long QT Syndrome (LQTS). We here expand our original observations and report that this abnormal QT-behavior persists as the heart rate returns to baseline.

Methods and Results: 90 patients with untreated LQTS (46% LQT1, 31% LQT2, 10% LQT3 and 13% not genotyped) and 79 controls were asked to stand-up quickly during continuous electrocardiographic recording. The R-R, QT and QTc intervals were measured at baseline, during maximal tachycardia and during maximal QT-stretching in response to standing. For the present study, we focused on the QTc-response as the heart rate slowed back to baseline. The heart-rate of LQTS patients and controls was similar at all times of interest (at baseline, during heart rate speeding and following slow-down to baseline). However, while the QT in controls shortened by 18±20 ms at maximal tachycardia, the QT of LQTS patients lengthened by 3±51 ms ($P < 0.001$). Moreover, the differences of QT-response between LQTS-patients and controls continued to diverge as the heart rate returned to baseline. The uncorrected QT of LQTS-patients failed to shrink back and was longer than baseline values by 21±42 ms while, in the control group, it was shorter than the initial QT by 4±27 ms ($P< 0.001$). Receiver-operating characteristic curves analysis showed that the best measurements for differentiating LQTS-patients from controls were "QTc at maximal QT-stretching" and "QTc upon return to baseline heart-rate."

Conclusion: The QT mal-adaptation seen in LQTS during the brief tachycardia that occurs while standing-up, persists even as the heart rate slows down to baseline. An easily performed bed-side test uncovers this phenomenon.