

MP2-3

How to Determine the QT-interval in the Paediatric Population: a Comprehensive Analysis of a Large Cohort of Long QT-Syndrome Patients and Controls

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Introduction:

Long QT-syndrome (LQTS) is associated with potentially fatal arrhythmias at young age. Treatment is very effective but its diagnosis may not be simple. Importantly, different methods are used to assess the QT-interval, which troubles its recognition. QT-experts advocate manual measurements with the tangent or threshold method. However, differences between these methods and their performance in LQTS diagnosis have not been established. We aimed to assess differences between these two methods for QT-analysis.

Methods:

Confirmed pathogenic mutation carriers in KCNQ1 (LQT1), KCNH2 (LQT2) and SCN5A (LQT3) genes were included as LQTS cases and their genotype-negative family members as controls when they were under the age of 18 years. Consecutive complexes from baseline ECGs were analyzed with both methods in two separate sessions by three readers. One reader additionally remeasured 10% of the ECGs.

Results:

We included 451 individuals (subgroups: 120 LQT1, 137 LQT2, 45 LQT3 and 127 controls) from 176 families, aged 8 ± 5 years and 50% females. In the total cohort, QT-tangent was 10.5 ms shorter ($p < 0.0001$) compared to QT-threshold (95% limits of agreement ± 17.2 ms). In all the subgroups the QT-tangent was shorter ($p < 0.0001$ all groups) compared to QT-threshold, but this was less pronounced in LQT2. Both methods had a high inter- and intra-observer validity (Intraclass correlation coefficient > 0.96), and a high distinctiveness between LQTS patients and controls (area under the curve > 0.86). Using the current guideline cut-off value (QTc 480 ms), both methods had similar specificity (99% versus 98%) but the QT-threshold yielded a higher sensitivity (20% versus 27%). Optimal cut-off value for boys was QTc-tangent 417 ms (78% sensitivity, 85% specificity) and QTc-threshold 428 ms (80% sensitivity, 79% specificity). For girls QTc-tangent 418 ms (83% sensitivity, 80% specificity) and QTc-threshold 446 ms (64% sensitivity, 93% specificity).

Conclusion:

The length of the QT-interval is different depending on the method used for determination. Both methods have a high validity. However, for diagnostic purposes current guideline cut-off values yield different results for these two methods, and could result in inappropriate reassurance or treatment. Adjusted cut-off values are suggested to diagnose LQTS.