**Introduction**: The clinical course and the treatment consideration in the congenital long QT syndrome (LQTS) are genotype specific. However, accurate diagnosis is often challenging with standard 12-lead ECG. We aimed to evaluate the utility of QT/RR slope by 24-hour Holter monitoring for differential diagnosis between LQT1 and LQT2.

**Methods**: Genetically identified 29 LQT1 patients and 25 LQT2 patients (mean age 23.4 +/- 14.9 years, 7 males) were enrolled. Consecutive sinus beats during each 15-second period were averaged, and the linear regression slopes of the QT interval, measured to the apex and to the end of the T wave plotted against RR intervals (QTa/RR and QTe/RR slopes, respectively) were calculated from entire 24-hour Holter recordings and separately during day and night periods.

**Result**: Average QTe was significantly higher, and QTe/RR and QTa/RR slopes from entire 24-hour Holter recordings were significantly steeper in the LQT2 patients in contrary to LQT1 patients (472.0 +/- 40.6 vs 447.1 +/- 44.8ms, P = 0.037; 0.262 +/- 0.063 vs 0.204 +/- 0.055, P = 0.0007; 0.233 +/- 0.052 vs 0.181 +/- 0.040, P = 0.0002, respectively). QTe/RR and QTa/RR slopes from daytime Holter recordings in the LQT2 patients were also significantly steeper than those in the LQT1 patients (0.197 +/- 0.057 vs 0.158 +/- 0.066, P = 0.024; 0.190 +/- 0.048 vs 0.153 +/- 0.050, P = 0.008, respectively). There were no significant differences in the other parameters. The receiver operating characteristic (ROC) curve analysis showed an optimal cutoff point of 0.211 of QTa/RR slope from entire 24-hour Holter recordings, with 80.0% sensitivity, 75.0% specificity and area under the curve of 0.804 (95% confidence interval, 0.68-0.93).

**Conclusion**: QT/RR relationships using 24-hour Holter monitoring may be useful for differential diagnosis between LQT1 and LQT2.