Predictive factors for future symptoms and dropout in patients with long QT syndrome in a single pediatric cardiovascular center in Japan

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Introduction: Long QT syndrome (LQTS) is a leading cause of sudden cardiac death due to arrhythmia in pediatric patients. A school-based ECG screening program to screen cardiovascular diseases uncovered many children and adolescents with LQTS. The aim of the study was to determine predictive factors for both the presence of symptoms and future dropout. We used data from a single pediatric center to unify the follow-up strategies.

Methods: Subjects were 143 consecutive LQTS patients (M:F = 72:71) who visited our center between April 2005 and December 2010, who were ≤ 20 years old, and included subjects who were previously followed by other hospitals. Thirty-seven subjects (26%) did not visit our hospital during the preceding 24 months (Dropout group). There were 106 final subjects (M:F = 49:57; mean age: 9.5 ± 5.1 yr; age range: 0-20 yr; mean follow-up period: 5.2 ± 5.6 yr). Subjects included 66 patients who were screened in the program (Screened group), 16 who visited clinics because of symptoms, and 24 who were diagnosed by family study or by chance.

Results: Four subjects (6%) in the Screened group experienced LQTS-related symptoms before diagnosis and 20 (30%) had a family history of LQTS. One subject died. She had a history of aborted cardiac arrest at 2 months, and died suddenly during sleep at 5 years of age. Logistic and multivariate regression analyses showed that the sole predictive factor for the presence of new symptoms after diagnosis was longer QTc values ($p = 0.02$) and that predictive factors for frequent symptoms were a longer follow-up period ($p < 0.0001$) and drug noncompliance ($p < 0.0001$). Logistic regression analysis showed that the presence of a family history ($p = 0.004$) was a significant predictor for the absence of dropout.

Conclusions: A school-based screening program is an effective method to identify children and adolescents with LQTS. Continued follow-up is important because the risk of a new episode increases with age in this group. Good drug compliance is crucial to prevent recurrent episodes. A new strategy is needed to prevent subjects without a family history from dropping out of visits.