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Risk factors predicting the future presence of long QT syndrome-related symptoms in pediatric patients diagnosed by school-based electrocardiographic screening programs in Japan

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Background and Objectives: Few data are available regarding risk factors predicting the future presence of long QT syndrome (LQTS)-related symptoms in pediatric patients who were diagnosed by school-based electrocardiographic screening programs.
Subjects: Subjects were 281 children and adolescents with LQTS (M/F=144/137; age at diagnosis, 10.9 ± 3.2 years; observation period, 4.0 ± 4.0 years) who were screened by the program. LQTS-related symptoms were defined as syncope, sudden death, and aborted cardiac arrest. In the regression analyses, age, sex, QTc values by Fridericia's formula at diagnosis, past history of LQTS-related symptoms, the presence or absence of family history of LQTS or sudden cardiac death and observation periods were used as independent variables.
Results: Of 281 subjects, 23 (8%) had past history of symptoms. Triggers were exercise in 9 and swimming in 8 subjects. After diagnosis, 28 subjects (10%) developed symptoms; triggers were exercise in 11 and swimming in 1. The prevalence of exercise as a trigger was not different between before and after diagnosis. However, swimming as a trigger significantly decreased (p=0.007) after diagnosis compared with before diagnosis. One child died. Logistic regression analysis showed that QTc value (p=0.001), past history (p=0.0002), the presence of family history of LQTS (p=0.0003) and observation period (p<0.0001) were predictive for the future presence of symptoms. Multivariate logistic regression analysis showed that observation period was a sole risk factor to predict the future presence of symptoms (Coefficient/SE, 2.84; p value of 0.005; Odds ratio, 1.14; 95%CI, 1.04-1.25), and that among patients who were treated (50 patients), drug incompliance was also a sole predictive factor for future presence of symptoms (Coefficient/SE, 2.67; p value of 0.008; Odds ratio, 7.88; 95%CI, 1.73-35.8).
Conclusions: Pediatric LQTS patients who were screened by programs should be monitored for long periods of time, in addition to individuals who were visited and diagnosed by the presence of LQTS-related symptoms. New strategies are needed to prevent exercise-triggered symptoms after diagnosis in these patients. Drug compliance should be monitored after initiation of therapy.