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Electrocardiographic screening of one-month-old infants for long QT syndrome in Japan

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Introduction: Electrocardiographic and molecular studies have clarified an association between sudden infant death syndrome and long QT syndrome (LQTS). However, only one prospective study had been conducted in Italy. Our preliminary data showed that the QT interval was longest when infants were 6–11 weeks of age. In Japan, medical examinations for infants at 1 month of age are mandatory. In 2010, we began a prospective study, obtaining electrocardiograms (ECGs) from more than 5,000 subjects at the time of their 1-month examination.
Methods: Resting ECGs were recorded in infants from eight areas in Japan. The QT intervals of three consecutive beats were measured. A formula to minimize the effect of heart rate for infants was used: $QTc=QT/RR^{0.43}$. A provisional criterion of QTc≥0.44 s was used. To assess the validity of the criterion, infants with a QTc ≥0.43 s were followed in each area; infants with QTc≥0.42 s were followed in the Kagoshima area. (QTc values by Bazett formula were expressed as $seconds^{0.5}$ for reference.) Genetic testing was performed on clinically diagnosed infants.
Results: Up to December 1, 2010, subjects were 2,019 infants (age 31±3 days). Mean QTc was 385±18 ms (412±19 $ms^{0.5}$). Among them, 20 (1.0 %) had a QTc of ≥0.42 s. In the Kagoshima area, 16 of 1,230 (1.3%) had the same values and all 16 infants had a QTc of <0.42 s a few weeks later. Six (0.3 %) had a QTc of ≥0.43 s; all were followed. Of the six, one (431 ms or 458 $ms^{0.5}$ at one month) was started on treatment at 51 days (500 ms or 535 $ms^{0.5}$ on the same day). Only one infant met the provisional criterion (463 ms or 492 $ms^{0.5}$ at one month) and treatment was started at 60 days (496 ms or 524 $ms^{0.5}$). Propranolol and mexiletine were administered. Genetic testing revealed a *KCNH2* mutation (3065T deletion) in the second case. The other is under investigation.
Conclusion The incidence of LQTS infants who are clinically diagnosed may be close to 1:1,000. ECG screening for LQTS in infants may be effective; cost-effectiveness of the screening should be further investigated.