Long-term Prognosis of Asian Patients with Short QT Syndrome

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Introduction: Short QT syndrome (SQTS) is a rare, life-threatening, inherited heart disease presenting as sudden cardiac death (SCD). The characteristics and prognosis of SQTS have not been known in Asian patients.

Methods: We consecutively included patients who were diagnosed with SQTS in South Korea. SQTS was defined as corrected QT interval \( \leq 340 \) ms in serial electrocardiograms. Patients without SQTS and overt cardiovascular disease were included by 1:4 age- and sex-matching. Electrocardiogram characteristics and cardiovascular events were compared between patients with and without SQTS.

Result: Thirty-four patients [age, 23.5 (21–30.5) years; male, 31] were followed up for 4.8 (2.0–7.7) years. In this SQTS cohort, young (<40 years) male were dominant. Symptoms included palpitation (n=4, 11.8%), loss of consciousness (n=3, 8.8%), and chest pain (n=3, 8.8%). Early repolarization, tall T wave, J wave, and U wave were more frequent in patients with SQTS than the patients without SQTS. QT dispersion [44.0 (28.0–73.0) vs 20.0 ms, \( p < 0.001 \)] was significantly higher and heart rate [52.0 (17.0–58.0) vs 70.0 (62.0–84.0) /min, \( p = 0.001 \)] was significantly slower in patients with SQTS than patients without SQTS. Atrial fibrillation (11.8% vs 2.9%, \( p = 0.030 \)) and ventricular tachyarrhythmia (5.8% vs 0%, \( p = 0.005 \)) were significantly more frequent in patients with SQTS than patients without SQTS.

Conclusion: In Asian patients, SQTS is associated with atrial and ventricular arrhythmias.