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. STQS was defined as corrected QT interval ≤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.