

This review article discusses current and controversial issues related to the diagnosis, risk stratification, and management of patients with congenital long QT syndrome (LQTS). Recent data on the genetic architecture of LQTS are presented, a risk stratification model is analyzed, and new potential cardiovascular prognostic factors are characterized. Much attention is given to the description of genotype-phenotype correlations of LQTS and molecular genetic mechanisms of cardiac transmembrane ion channel abnormalities that are key in the arrhythmogenesis of LQTS. The main methods of management of patients with LQTS, especially those at high risk of cardiac events, including a genotype-specific approach to management, are also presented.