The Long QT Syndrome (LQTS) was first identified as a clinical entity in 1957 and the genetic underpinning was established in the mid-1990s. The estimated frequency of LQTS is about 1 in 3,000 persons in the general population, and the disorder is characterized by QTc prolongation on the ECG, recurrent syncope due to ventricular tachyarrhythmias, and sudden cardiac death. To date, 13 different genes have been associated with LQTS involving hundreds of different mutations. Three forms of LQTS (LQT1, LQT2, and LQT3) make up over 90% of the reported patients with LQTS.

This presentation on LQTS will be drawn from our International Registry of LQTS involving 1,000 proband-identified families and over 2,000 genetically confirmed LQTS patients. The focus of the talk will be on genotype-phenotype aspects of this disorder, risk stratification, risk mechanisms, and established and potentially new therapies for the LQT1-3 forms of this disorder. The risk mechanisms for LQT1, LQT2, and LQT3 are quite different, and our latest patient-related insights and approaches to management regarding genotype risks, drug therapy, and surgical and device interventions will be presented and discussed.