

**Risk factors for major arrhythmic events in pediatric patients with long QT syndrome**

Burkard T.(1), Horndasch M.(1), Seidel H.(2), Hessling G.(1), Wolf C.M.(1)  
 Department of Pediatric Cardiology and Congenital Heart Disease, German Heart Center Munich, Technical University of Munich, Munich, Germany(1); Institute of Human Genetics, Klinikum Rechts der Isar, Technical University of Munich, Munich, Germany(2)

**Introduction/Objectives:**

Long QT-Syndrome (LQTS) is an inherited arrhythmic disorder associated with sudden cardiac death (SCD). The goal of this study was to define predictors for major arrhythmic events (MAEs) in pediatric LQTS patients.

**Methods:**

All patients aged ≤ 18 years with a clinical and molecular genetic diagnosis of LQTS type 1, 2 or 3 were included in this retrospective single-center study. Clinical data were recorded by medical chart review. MAEs were defined as the occurrence of SCD, aborted SCD, appropriate implantable cardioverter- defibrillator discharge or sustained ventricular tachycardia.

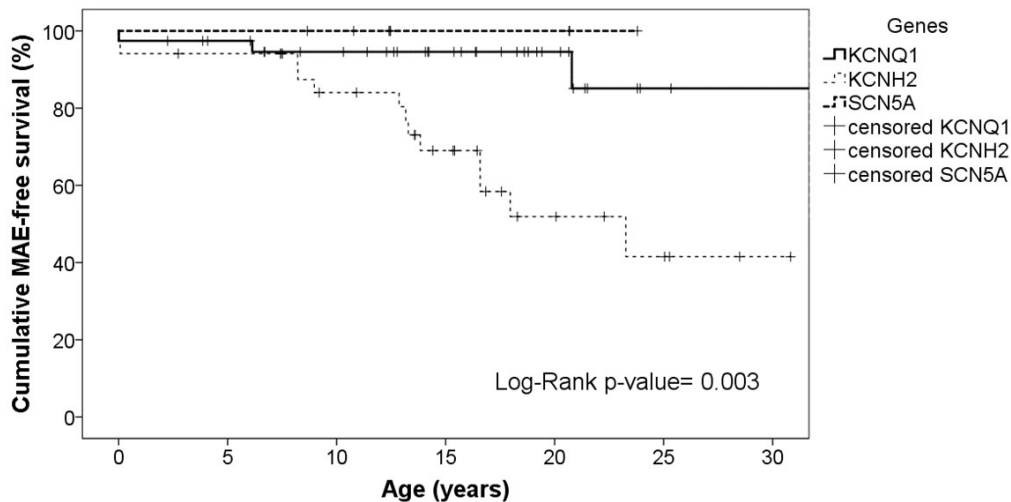
**Results:**

Childhood onset LQTS was diagnosed in 81 patients (46 males) from 60 families. Median age at diagnosis was 7.6 years (range 0.0 – 18.0 years). A pathogenic or likely pathogenic mutation in the *KCNQ1*, *KCNH2* and *SCN5A* gene was identified in 39, 34 and 8 patients, respectively. MAEs were documented in 16/81 patients (9 males), during a median follow-up time of 5.0 years (range 0.2 – 25.7 years). MAE were more likely in patients carrying a mutation in the *KCNH2* locus (13/16, 81.3 %) than in the *KCNQ1* (3/16, 18.8 %) or *SCN5A* locus (0/16) (see Figure). QTc-duration was longer in patients with MAE compared to patients without MAEs (570 ± 62 ms versus 497 ± 49 ms, p <0.001, independent t-test). Syncope occurred more often in patients with MAEs (9/16, 56.25%) than in patients without MAEs (11/65, 16.9 %) (p= 0.001, Chi-square-test).

**Conclusions:**

Risk factors for life-threatening events included mutations in the *KCNH2* locus, longer QTc-duration and a higher quantity of syncope in this pediatric LQTS cohort.

**Figure:**



No. at Risk							
	0	5	10	15	20	25	30
KCNQ1	39	30	12	3			
KCNH2	34	24	7	1			
SCN5A	8	7	3	0			