

## LONG-TERM FATE OF AN UNSELECTED COHORT OF CONGENITAL LONG QT SYNDROME PATIENTS DIAGNOSED IN CHILDHOOD

### Contact details:

Terézia Tavačová, MD  
Children's Heart Centre, Motol University Hospital  
V Úvalu 84, 150 00 Prague, Czech Republic  
T: +420 775 183 222  
e-mail: [terezia.tavacova@fnmotol.cz](mailto:terezia.tavacova@fnmotol.cz), [tereziatavacova@gmail.com](mailto:tereziatavacova@gmail.com)

Tavačová T. (1), Kubuš P. (1), Krebsová A. (2), Janoušek J. (1)

1. Children's Heart Centre, 2<sup>nd</sup> Faculty of Medicine, Charles University and Motol University Hospital, Prague, Czech Republic
2. Cardiology Department, Institute for Clinical and Experimental Medicine, Prague, Czech Republic

**OBJECTIVES:** Congenital long QT syndrome (LQTS) is genetically heterogeneous disorder with type-specific risks for major arrhythmic events (MAE). We aimed to perform a retrospective analysis of unselected cohort of LQTS patients diagnosed in childhood.

**METHODS:** All paediatric patients (N=224, female 119, 53 %) diagnosed with LQTS (Schwartz score  $\geq 1.5$  points and/or presence of a pathogenic or likely pathogenic variant) between July 1985 and December 2021 at median age (IQR) 11.7 (6.5-14.2) years were included. Data were retrieved from medical records and cross-mapped with the National Death Registry. Patients were followed-up for a median (IQR) of 8.8 (2.8-16.7) years.

**RESULTS:** Reasons for presentation were LQTS related symptoms (N=91, 40.6%), positive family history (N=66, 29.5%), incidental finding of prolonged QTc (N=37, 16.5%) and positive pre-participation screening (N=30, 13.4%). QTc interval and Schwartz score were median (IQR) 482 (460-516) ms and 4.0 (3.0-5.0) points, respectively. Likely pathogenic or pathogenic variants were found in 119/159 tested patients (74.8%). Betablockers (BB) were administered in 202 patients (90.2%) with the proportion of non-selective BB increasing from 16.7% to 58.5% comparing periods 1985-2015 and 2016-2021 ( $p < 0.001$ ). Twelve patients died from cardiovascular cause (5.4%) yielding a 5/10/20 years survival probability of 97.2/94.7/91.5%. Freedom from MAE defined as either sudden cardiac death/arrest or appropriate ICD therapy after diagnosis of LQTS was 92.9/87.7/83.5%. MAE was independently predicted by early presentation (HR 14.65,  $p = 0.0013$ ), Schwartz score (HR 1.77,  $p = 0.0022$ ), QTc (HR 1.018,  $p < 0.001$ ) and presence of LQTS3 (HR 34.54,  $p = 0.025$ ). MAE burden decreased significantly in patients on non-selective BB in comparison to selective BB regardless of other variables (gender: HR 0.15,  $p = 0.0121$ , early presentation: HR 0.11,  $p < 0.0001$ , Schwartz score: HR 0.16,  $p < 0.0001$ , LQTS3: HR 0.15,  $p = 0.0169$  and QTc: HR 0.14,  $p = 0.0096$ ).

**CONCLUSIONS:** Patients with LQTS diagnosed in childhood had a long-term survival probability of

91.5%. Early presentation, Schwartz score, genotype and QTc duration were major predictors of MAE. Genetic testing had a high diagnostic yield. Non-selective BB significantly decreased MAE burden regardless of other variables.

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