
Department of Clinical Sciences, Pediatrics (1); Heart Centre Umeå University Hospital, Department of Public Health and Clinical Medicine (2), both at Umeå University, Umeå, Sweden.

Background
In the congenital long QT syndrome (LQTS), mutations cause changes in the ion channels of the myocytes leading to severe arrhythmias like Torsades de Pointes. The symptoms vary from dizziness and syncope to aborted cardiac arrest (ACA) and sudden cardiac death (SCD). For patients with a high risk for ACA/SCD, implantable cardioverter defibrillator (ICD) is an option for treatment. This survey aimed to describe the pediatric LQTS-population with implanted ICDs in Sweden.

Methods
Patient data was extracted from the Swedish ICD- and Pacemaker Registry. The inclusion criteria were LQTS and implanted ICD, and 173 patients met the criteria. Informed consent was obtained from 138 patients. Medical records were reviewed and data concerning medical treatment with betablockers, indication for ICD and DNA analysis were collected. We compared the pediatric and the adult LQTS population.

Results
The number of ICD-implantations in patients diagnosed with LQTS in Sweden has increased exponentially since 1992. In the pediatric LQTS population, 22 patients received ICD in the age of 0-19 years. Three of these patients were found to have catecholaminergic polymorphic ventricular tachycardia (CPVT) when genetic testing was performed later. Boys were overrepresented until age 20 when it reversed. Based on medical records, the three most common indications for ICD-implantation were syncope, 55% vs 48% (age ≤19 vs >19); ACA, 14% vs 31%; and primary prophylactic treatment, 32% vs 10%. At the time for symptoms indicating ICD implantation, 73% of children/adolescents were prescribed betablockers compared to 48% of the adults. DNA analysis was performed in 82%. A mutation was found in 70%, the three most common being LQT2, 38%; LQT1, 29%; and LQT3, 15%.

Conclusions
ICD-implantations in LQTS-patients are increasing, which may be due to increased awareness of the diagnosis and the choice of primary prophylactic treatment. Primary prophylactic ICD seems to be more common in children/adolescents than in adults. In contrast to current recommendations, relatively few adult patients are on betablockers before receiving ICD. Most of the LQTS patients in the Swedish ICD registry have been tested genetically and this survey also indicates that comparatively many LQT1-patients receive ICDs.