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The experience of a French specialized inherited arrhythmia center in the management of long QT syndrome patients

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Introduction: Congenital long QT syndrome (LQTS) is a hereditary disease characterized by prolonged QTc interval and risk of ventricular tachyarrhythmias, which may lead to syncope, cardiac arrest, or sudden death in young people.

Objective: The aim of this study is to report the experience of the French referral center of inherited arrhythmia of the University Hospital of Nantes.

Methods: Consecutive patients affected with long QT syndrome were recruited in a multicentric prospective registry in France (15 centers) between 1997 and 2018. Clinical data and 12-lead ECG were collected. Genetic screening was performed using dHPLC-DNA sequencing, HRM or targeted sequencing for at least KCNQ1, KCNH2 and SCN5A.

Results: In this study, we enrolled a total of 741 patients affected by LQTS according to the Schwartz score (447 (60%) females, 404 (55%) index cases). Mean age at diagnosis was 33±21 years. In this cohort, 343 patients (46%) were symptomatic: 66 patients experienced resuscitated sudden cardiac death (SCD, 9%), 211 (28%) syncope and 66 (9%) ventricular arrhythmias. One hundred and two patients (14%) had history of familial

SCD. At baseline, heart rate was 69±19 bpm, PR 149±39 ms, QRS 86±16 ms and QTc 479±62 ms.

Three hundred and eighty-six patients (52%) were treated with beta-blockers and 88 patients (12%) were implanted with an ICD.

Genetic screening was performed in 668 (90%) patients. Genetic screening for the 3 major genes of LQTS was positive for 411 patients (62%): 165 variants (25%) in KCNQ1, 178 variants (27%) in KCNH2, 68 variants (10%) in SCN5A. Moreover, we found variants in minor genes of LQTS for 22 patients (3%).

During a mean follow-up of 6.2±5.2 years, 64 patients (9%) underwent arrhythmic events (7 SCD (1%), 8 appropriate ICD therapy (1%) and 49 ventricular arrhythmias (7%). The rate of arrhythmic event was 1.39%/y. Mean age at the first event was 41.7±21.5 years. Sixteen patients (2%) died of non-arrhythmic causes.

Conclusion: Care in a specialized inherited arrhythmia center is associated with a low incidence of arrhythmic event (1.39%/y) in patients affected with LQTS.