Electrocardiographic evidence of transient reverse remodelling of ventricular repolarization after prolonged recurrent episodes of torsade de pointes in a patient with congenital long QT syndrome

Antonio Sorgente1,2*, Gian-Battista Chierchia1, Andrea Sarkozy1, and Pedro Brugada1

1Heart Rhythm Management Center, UZ Brussel-VUB, Laarbeeklaan 101, 1090 Brussels, Belgium and 2University of L’Aquila, Piazza Salvatore Tommasi 1, 67010 Coppito, L’Aquila, Italy

* Corresponding author. Tel: +32 2 4763038, Fax: +32 2 4776840, Email: sorgente.antonio@gmail.com

A 15-year-old girl presented to the emergency room after a prolonged episode of seizures following a dental intervention. This episode had been preceded the same day by an episode of dizziness and pre-syncope during a small effort.

On admission, the electrocardiogram (ECG) showed sinus rhythm with a heart rate of 65 bpm, a short duration of the interval PR (102 ms), a QRS axis of 90°, and an important QT prolongation. The QT interval was 577 ms with a corrected QT interval (QTc) of 590 ms. Electrolytes were completely normal. The echocardiogram performed after the admission to the hospital revealed normal left ventricular function with no structural abnormalities. During the first night of hospitalization, the patient experienced three new episodes of loss of consciousness associated with myoclonus and generalized tonic–clonic seizures. The Holter ECG recording performed during these episodes showed an important prolongation of the QT interval and five episodes of polymorphic ventricular tachycardia (torsade de pointes), occasionally degenerating into ventricular fibrillation, with a maximal duration of 1 min and 40 s (Panel A). After the three most prolonged episodes of tachyarrhythmia, the patient converted spontaneously into sinus rhythm and showed repeatedly transient shortening of the QT interval: the shortest QT measured was of 400 ms with a QTc of 320 ms (Panel B). The QT interval prolonged again after about 2 h since the last episode of torsade de pointes, reaching a value of 620 ms (Panels C, D).

Congenital long QT syndrome was diagnosed. Genetic examination was only recently performed and the result is still on the way. The patient was promptly treated with an association of nadolol and mexiletin and finally underwent an implantation of a double chamber defibrillator. Familiar screening recognized the mother as a carrier of the long QT syndrome (LQTS). Her ECG showed spontaneously a marked QT prolongation (QTc 600 ms) in the absence of previous familiar or personal history of sudden death or syncope. The patient’s outcome has been uneventful. No new ventricular arrhythmias or electrical storm were documented in about 4 years of follow-up.

Our case report is remarkable for two reasons: (i) recurrent and prolonged episodes of torsade de pointes, all self-limiting, with a maximal duration of 1 min and 40 s in a single ECG Holter recording of a young patient affected by congenital LQTS and (ii) the electrocardiographic documentation of transient reverse remodelling of ventricular repolarization after restoration of sinus rhythm. The shortening of the QT interval after prolonged episodes of torsade de pointes in the context of a congenital LQTS is a rare finding and could be explained by a transient increase of intracellular potassium or by a transient reduction of intracellular calcium, as previously demonstrated in an animal model.

(A) ECG Holter recording report of the longest episode of torsade de pointes (total duration of 1 min and 40 s). (B) ECG Holter recording report of the QT interval shortening after the three most prolonged episodes of torsade de pointes (the shortest QT interval was equal to 400 ms). (C) ECG Holter registration showing a duration of the QT interval of 400 ms, immediately after the last episode of torsade de pointes. (D) ECG Holter registration showing a prolongation of the duration of the QT interval to 620 ms, 2 h after the last episode of torsade de pointes.

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