CASE REPORT

Sudden Paradoxical QT-Interval Prolongation Exacerbating T-Wave Alternans in a Patient with Type 3 Long QT Syndrome

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We report a case with type 3 congenital long QT syndrome, who exhibited a sudden paradoxical QT-interval prolongation during a progressive increase in heart rate, which exacerbated T-wave alternans.

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congenital long QT syndrome; QT interval; RR interval; T-wave alternans

CLINICAL CASE

An 8-year-old boy received an implantable cardioverter defibrillator (ICD) after suffering cardiopulmonary arrest. He had been diagnosed genetically with long QT syndrome (LQTS) type 3 (Ala1186Thr) at the age of 2 years. He was on medications including mexiletine 280 mg and propranolol 7 mg daily. Appropriate ICD shock has occurred once a year. Twenty-four-hour continuous electrocardiography showed a total of 112 episodes of T-wave alternans (TWA) with beat-to-beat alternating polarity of the T wave, that is, T-wave polarity alternans (TWPA). The RR and QT intervals before and during the episodes of TWPA were evaluated. Twenty-nine episodes of TWPA initiated by progressive shortening of the RR interval were analyzed. The other 83 episodes were not eligible for this analysis because of irregular or sudden RR interval shortening or could not be analyzed due to noise/artifact. In all 29 episodes, an abrupt prolongation of the QT interval

(by 46.5 ± 26.9 milliseconds, mean $\pm$ SD) occurred in spite of consistently increasing heart rate preceding the onset of TWPA. A representative episode is shown (Fig. 1). Figure 1A demonstrates the onset of TWA (beat 10~), TWPA (beat 18~), and QRST-wave alternans (QRSTWA) (beat 22~) with abrupt paradoxical QT-interval prolongation (beat 17). The RR interval became consistently shorter across beats 7–22. The QT interval became progressively shorter in beats 7–16; however, abrupt prolongation of the QT interval by 63 milliseconds occurred at beat 17. After beat 17, the QT interval was gradually prolonged and fluctuated alternately along with the appearance of TWPA. TWA magnitude was amplified and degenerated into QRSTWA with the gradual shortening of the TQ interval. The relationship between QT interval and preceding RR interval was suddenly inverted after beat 17 (Fig. 1B). QRSTWA changed into TWA after the prolongation of RR interval resulting in TQ interval >0 millisecond, and then TWA was attenuated and finally disappeared with the decrease in heart rate.

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Figure 1. (A) Representative example of onset of T-wave alternans, T-wave polarity alternans, and QRST-wave alternans with abrupt QT-interval prolongation during a progressive increase in heart rate in a patient with type 3 long QT syndrome. Numbers in italics indicate beat numbers. Red and blue numbers represent RR and QT intervals, respectively. (B) QT interval and the preceding RR interval of each beat. QT intervals were measured manually with the electronic calipers. RR intervals were automatically generated.

Rosenbaum and Acunzo demonstrated that a shortening of the TQ interval plays a major role in the initiation and maintenance of TWA in patients with LQTS.¹ Thus, the inverse QT/RR relationship may trigger and worsen TWA. Although the exact mechanism of the paradoxical QT-interval prolongation is unknown, our case suggests that the failure of the QT interval to adapt to the increase in heart rate may play a role in exacerbating TWA in patients with type 3 LQTS. This report is a follow-up on the same patient as previously presented.^{2,3}

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