

## CLINICAL IMAGE

# Electrical storm in an acquired short QT syndrome successfully treated with quinidine

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## Abstract

Short QT syndrome (SQTS) is a malignant heart disorder defined by the presence of ventricular arrhythmias causing syncope and sudden cardiac arrest. The prevalence in the pediatric population is 0.05%. Quinidine is an established agent for pharmacological prophylaxis in SQTS patients, but can also terminate an electrical storm.

## KEYWORDS

arrhythmia, electrical storm, quinidine, short QT

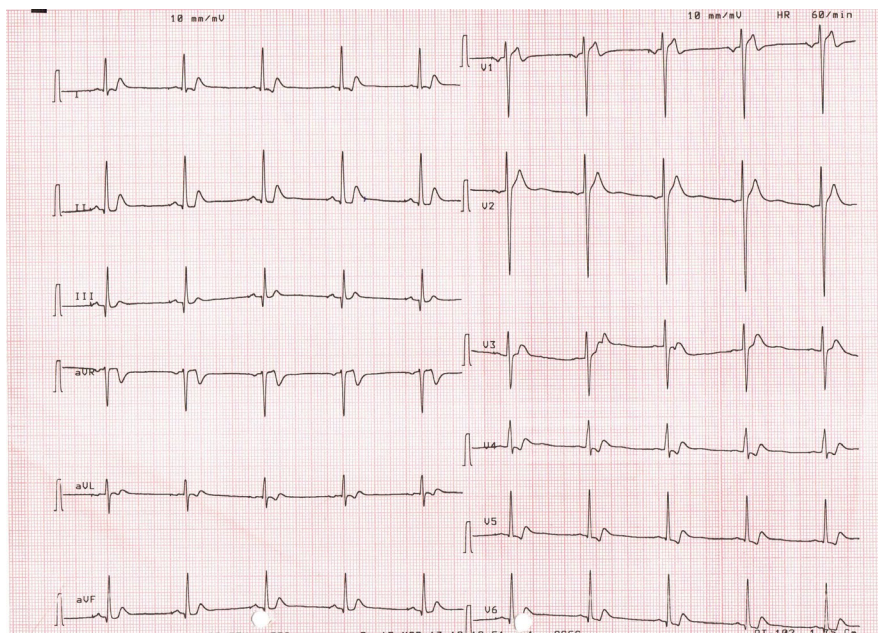
## 1 | CASE PRESENTATION

A 14-year-old male with no past medical history presented to our emergency department after an episode of loss of

consciousness in the classroom. Physical examination was unremarkable.

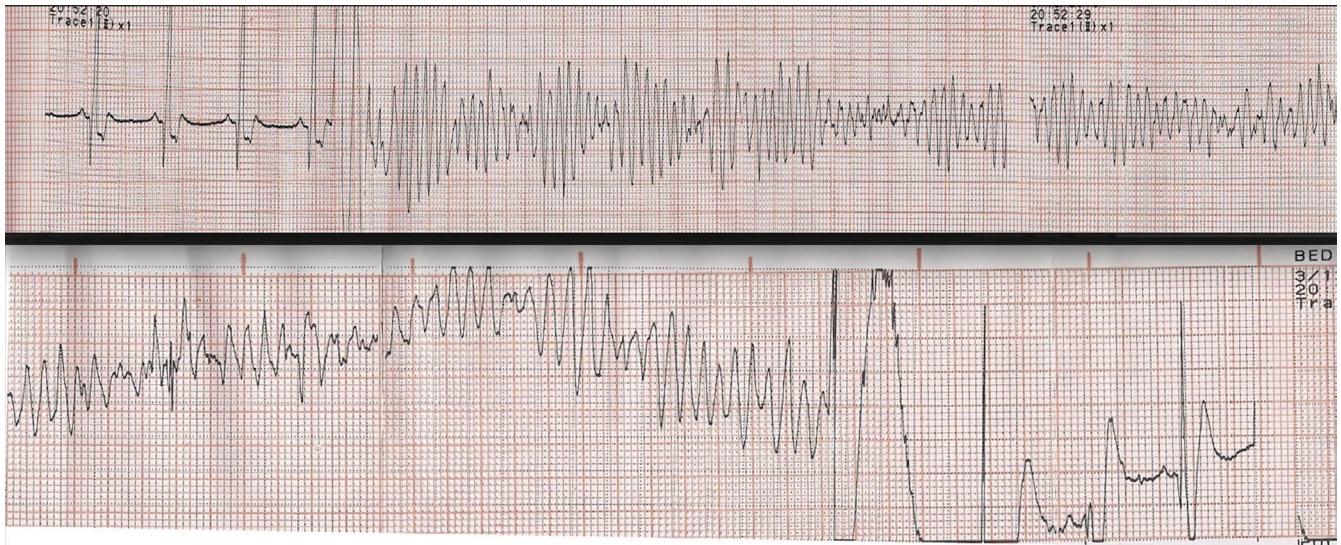
The electrocardiogram showed a QT of 320 ms and a QTc of 320 ms (Figure 1). Echocardiography, cardiac magnetic

**FIGURE 1** The 12-lead surface electrocardiogram (ECG) obtained on the admission showed sinus rhythm with a heart rate of 60 bpm, a QT of 320 ms and a QTc of 320 ms



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**FIGURE 2** ECG strip showing an episode of Torsade de pointes degenerate into ventricular fibrillation. The episode was induced after a premature ventricular beat due to R/T phenomenon

resonance imaging, and coronary angiography were normal. The patient went on to have a positive programmed ventricular stimulation for inducible ventricular tachycardia and underwent an implantable cardioverter defibrillator implantation. Genetic testing did not detect any mutations. A short QT interval is usually considered if QTc is  $\leq 340$  ms,<sup>1,2</sup> and a diagnosis of SQTS was established.

After 5 months, the patient presented with electrical storm with 16 successive episodes of Torsade de pointes that degenerate to ventricular fibrillation (Figure 2). The episodes were induced by a premature ventricular beat due to R/T phenomenon. Esmolol and amiodarone intravenous infusion were proven ineffective, as well as sotalol therapy. The patient received a 500 mg oral loading dose of quinidine and then 250 mg two times daily. Quinidine led to the rapid disappearance of premature beats and arrhythmia episodes.

Quinidine can normalize the QT interval, leading to significant QT prolongation, a longer ventricular effective refractory period, and is an established preventive therapy for SQTs.<sup>1,2</sup>

To the best of our knowledge, only a few cases of electrical storm in SQTs have been described in the pediatric population, and this is the first case demonstrating the beneficial effect of quinidine in the suppression of electrical storm in SQTs patients.<sup>1,2</sup>

## CONFLICT OF INTEREST

The authors report no financial relationships or conflicts of interest regarding the content herein.

## AUTHOR CONTRIBUTIONS

MS, AT: Conception and design of the research and writing of the manuscript. AT, MS: Acquisition of data. ES, MS: Analysis and interpretation of the data. EL: Critical revision of the manuscript for intellectual content.

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## REFERENCES

1. Pereira R, Campuzano O, Sarquella-Brugada G, et al. Short QT syndrome in pediatrics. *Clin Res Cardiol*. 2017;106(6):393-400.
2. Campuzano O, Sarquella-Brugada G, Cesar S, Arbelo E, Brugada J, Brugada R. Recent advances in short QT syndrome. *Front Cardiovasc Med*. 2018;5:149.

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