

Long QT syndrome, utility of bicameral pacemaker

Síndrome de QT largo, utilidad del marcapasos bicameral

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The use of implantable cardioverter defibrillator (ICD) in pediatric age represents a challenge, because of anatomic limitations, increased risk of lead fracture, T wave oversensing and inappropriate therapies¹. We present a case of a 4-year-old boy with no medical history of heart disease and no family history of sudden death, who was admitted to our institution with 1 month of recurrent syncope. He received treatment with lidocaine in other hospitals due to broad QRS complex tachycardia. The physical examination was unremarkable. The echocardiogram was normal. The electrocardiogram shows a long QTc of 510 ms and prolonged ST segment, suggesting long QT type III (LQT3) (Fig. 1).

During the 1st day of admission, he presented a polymorphic ventricular tachycardia with a heart rate of 220 bpm, preceded of premature ventricular contractions (Fig. 2).

He required electric cardioversión with 1 J/kg and 2 J/kg in three occasions, afterward treatment with intravenous magnesium and beta-blocker was started. Then, a endocardial dual-chamber pacemaker was implanted in the 3rd day since admission without complications (Fig. 3). The patient was discharged 2 days after.

In patients with LQT3 mutations, the ventricular arrhythmias occur more often during rest and bradycardia is an important triggering factor. The B-blockers may not be beneficial in this group, especially in symptomatic patients². So that, pacemakers have shown to be an effective therapy in preventing sudden death³. In addition, dual-chamber pacing permits A-V synchrony,

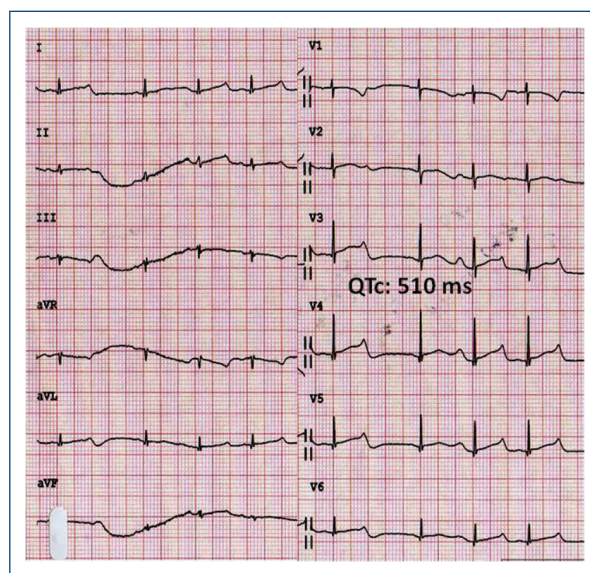


Figure 1. Electrocardiogram with long QT type III with corrected QT (QTc) of 510 ms and prolonged ST segment.

physiological heart rate, shortening of the QT interval and reduces the risk of R on T phenomenon⁴. Once the pacemaker has been implanted, the beta-blocker can be started to prevent ventricular arrhythmias. In symptomatic small patients with pause dependent ventricular arrhythmias, the dual-chamber pacemakers are an adequate alternative until they grow up and ICD can be implanted^{3,4}.

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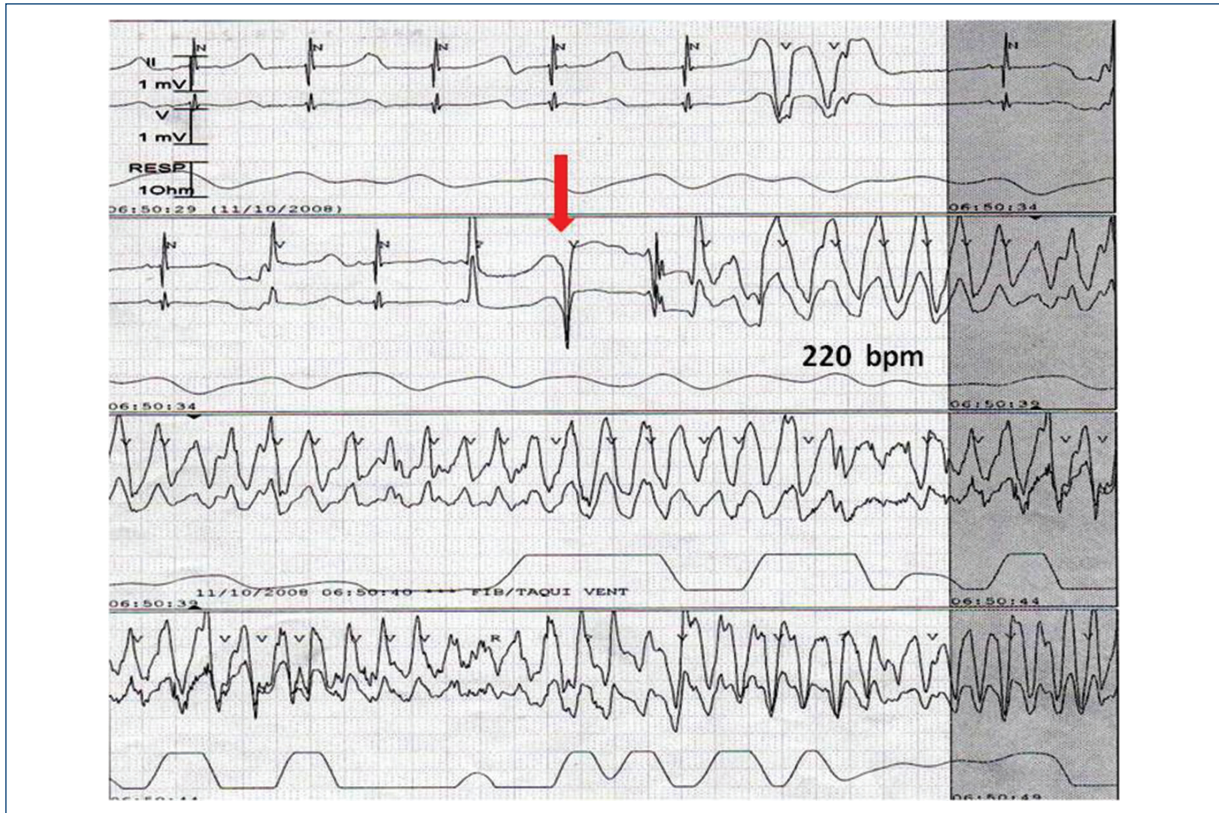


Figure 2. Monitor trace with a polymorphic ventricular tachycardia started with premature ventricular contraction with R on T phenomenon (red arrow).

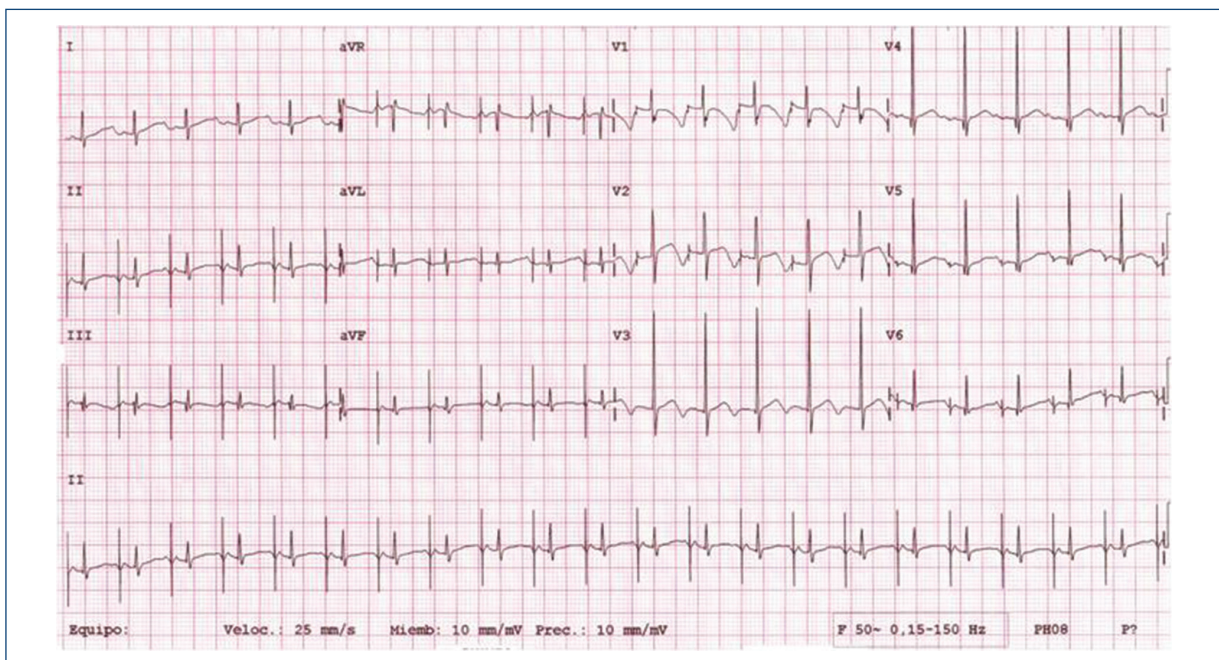


Figure 3. Electrocardiogram with pacemaker bicameral stimulation (DDDR mode), now with AAIR estimation and shortening of the QT to 460 ms with a heart rate of 125 bpm.

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Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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