The Classic ‘T on P’ Phenomenon

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Congenital long QT syndrome (LQTS) is a genetic channelopathy with variable penetrance characterised by abnormally prolonged ventricular repolarisation with an increased propensity to syncope and polymorphous ventricular tachycardia, which may lead to ventricular fibrillation and sudden cardiac death. Electrocardiography is the primary important step in the diagnosis of LQTS but electrocardiogram (ECG) findings may be easily ignored. We present an asymptomatic two-month-old infant with peculiar ECG features with ‘T on P’ phenomenon caused by a homozygous mutation in the KCNQ1 gene. Implantable cardioverter-defibrillator implantation is an effective therapy and can apply to small children successfully.

Keywords
‘T on P’ phenomenon, marked QT prolongation, LQTS syndrome, KCNQ1 mutation, epicardial implantable cardioverter-defibrillator

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Compliance with Ethics: All procedures were followed in accordance with the responsible committee on human experimentation and with the Helsinki Declaration of 1975 and subsequent revisions, and informed consent was received from the parents/guardian of the patient involved in this case study.

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Case report
A two-month-old infant referred to our hospital for innocent murmur. His parents had no complaints about their baby. Examination was normal except innocent murmur and echocardiography was unremarkable, which showed the prolonged corrected QT interval of 600 msec (see Figure 1A). Twenty-four hour Holter ECG was normal except prolongation of the corrected QT (QTc) interval with typical ‘T on P’ phenomenon (see Figure 1). The genetic analysis revealed a homozygous p.R366Q (c.1097>A) mutation in the KCNQ1 gene (LQT1). The hearing test was unrevealing.

Figure 2

Figure 1

The boy's post-operative course was uneventful, and he was discharged 3 days after the operation with propranolol 3 mg/kg/day. During the six-month follow-up period, the boy had no attacks of ventricular tachycardia or ventricular fibrillation.
In this article, we presented an ECG finding with classic ‘T on P’ phenomenon consisting of prolongation of Q and a delayed T wave, followed or overlapped by the succeeding P wave.\(^3\) It has been demonstrated that congenital LQTS with very long QT intervals can produce a functional block between the His bundle and ventricular muscle due to prolonged ventricular refractoriness that can lead to 2:1 atrioventricular block and severe bradycardia.\(^3\)

Genetic testing not only has significant diagnostic implications but also has prognostic and therapeutic implications in LQTS. For example, the underlying genetic basis heavily influences the response to standard LQTS pharmacotherapy (beta-blockers), because beta-blockers are extremely protective for patients with either LQT2 or LQT3.\(^4\) In addition, intra genotype risk stratification has been realised for LQT1 and LQT2 on the basis of mutation type, mutation location and cellular function.\(^4\) Therefore, genetic analysis of LQTS is important to identify the therapeutic option and risk stratification.\(^5\) In our patient, the genetic analysis identified a missense mutation (R366Q) in the \(\text{KCNQ1}\) gene. Propranolol therapy was useful in this patient because beta-blockers are extremely protective in LQTS1 patients. In the current patient, additional treatment with ICD was performed because the QTc interval was very long at 600 msn and male sex.

Implantation of ICDs in paediatric patients presents various difficulties:\(^6\) Despite the downsizing of pulse generators as well as improvements in lead design, paediatric patients present challenges in terms of patient size and growth. Relative to the venous diameter and thoracic anatomy of the infants, transvenous leads are large in calibre and length. Epicardial patches require sternotomy or thoracotomy, with higher rates of insulation and conductor fractures as well as the risk of the development of restrictive pericardial lesions due to the large surface area of the patches. Implantation techniques using a subcutaneous array and an abdominally placed ICD generator have been described previously.\(^6\) However, experience with infants younger than six months is limited. Despite low body weight and young age, ICD implantation applied to this child as a primary prophylaxis in LQTS. ICD implantation is an effective therapy and can be applied to small children successfully.