Pediatr Cardiol (2008) 29:969–970
DOI 10.1007/s00246-007-9160-5

CASE REPORT

# Cardiac Arrest Secondary to Long $QT_C$ in a Child with Propionic Acidemia

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Received: 11 August 2007 / Accepted: 5 November 2007 / Published online: 5 December 2007


**Abstract** In 2006 a case report was published in this journal of a child with known propionic acidemia being diagnosed with the long $QT_C$ syndrome [2]. This following case documents the first reported case of a child with this condition suffering a life-threatening event from the associated long $QT_C$ syndrome.

**Keywords** Propionic academia · Long $QT_C$ syndrome

Propionic acidemia is an autosomal recessive disorder caused by a deficiency in propionyl-CoA carboxylase. The disorder usually presents with episodes of metabolic acidosis. Treatment consists of a protein-limited diet and l-carnitine. Cardiomyopathy is a well-recognized complication of this condition occurring mainly at times of metabolic decompensation. However, recent literature has also highlighted the association of propionic acidemia and long $QT_C$ (>440 ms) [1, 2]. Althought both these studies report the association, this case is the first report of a child with propionic acidemia developing life-threatening complications from long $QT_C$.

## Case Report

The patient is a 13-year-old girl who was diagnosed with propionic acidemia shortly after birth. She was otherwise well and her metabolic condition was well controlled on sodium benzoate and carnitine. She presented following an episode of collapse while playing netball at school. Cardiopulmonary resuscitation had been instigated immediately and was required for 7 min. She spent 48 h in the pediatric intensive care unit (PICU) where intravenous sodium benzoate, carnitine, and 10% dextrose were commenced. On discharge from the PICU she was referred to the cardiology team.

On further history-taking, she reported previous episodes of dizziness while playing netball, at times forcing her to sit down, but she had never collapsed. She denied any history of palpitations. Examination was normal as was an echo; however, a 24-h tape showed a prolonged $QT_C$ of 450 ms. This was confirmed on exercise testing when the $QT_C$ increased further to 490–508 ms in the recovery phase (Fig. 1). She was commenced on 40 mg nadalol twice a day and remains under cardiology review.

## Discussion

Kakavand et al. [2] were the first to publish a case demonstrating the association between propionic acidemia and long $QT_C$. They reported an incidental finding of a long $QT_C$ in a 7-year-old propionic acidemia patient who was undergoing routine cardiology assessment because of the known association with cardiomyopathy. This patient was subsequently commenced on atenolol.

Subsequently Baumgartner et al. [1] published a case series of 10 patients with propionic acidemia who under went cardiology investigation. They found a prolonged $QT_C$ >440 ms in 70% of the children after infancy and 60% had a markedly prolonged $QT_C$ >460 ms. This prolongation of the $QT_C$ was independent of their metabolic states.

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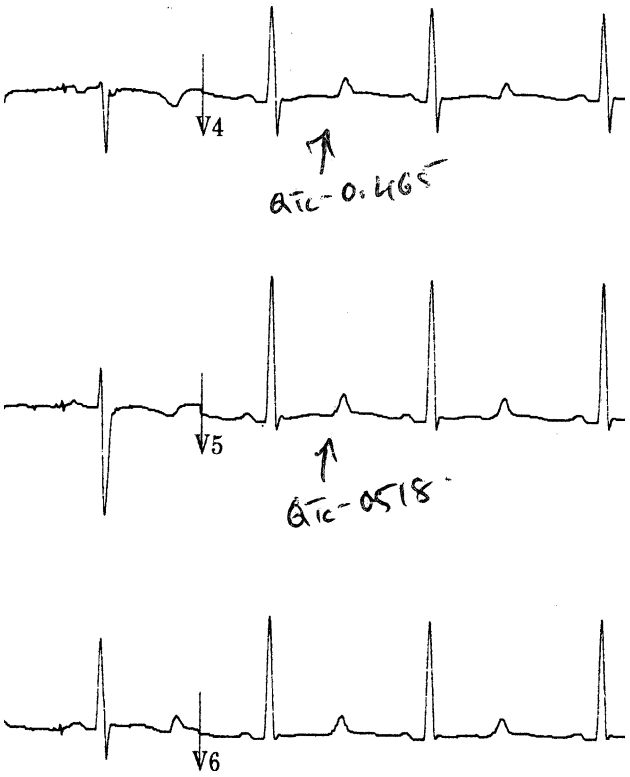


**Fig. 1** ECG trace showing prolongation of the QTC to 518 ms

Abnormal repolarization characterized by preterminal T-wave inversion on 12-lead ECG was present in 60%, and in each case, it was associated with a $QT_C$ >460 ms. Rhythm disturbances were seen in 20%, and 40% of those with $QT_C$ >450 ms had reduced left ventricular contractility.

Our report combined with the previous two articles on this subject should lead to the consideration that all children with propionic acidemia should receive regular cardiology follow-up with a 12-lead ECG, 24-h Holter monitoring, and exercise testing. In the case of the latter, it is important that the ECG recording continues for 10 min of the recovery phase; this is a common time for QT prolongation to appear. Care should also be taken when prescribing medications known to have associations with prolongation of $QT_C$ (e.g., erythromycin).

## References

1. Baumgartner D, et al. (2007) Prolonged $QT_C$ intervals and decreased left ventricular contractility in patients with propionic academia. J Paediatr 150:192–198
2. Kakavand B, Schroeder VA, Sessa Di (2006) Coincidence of long QT syndrome and propionic academia. Pediatric Cardiol 27: 160–161