A case report of Long QT presenting as seizures

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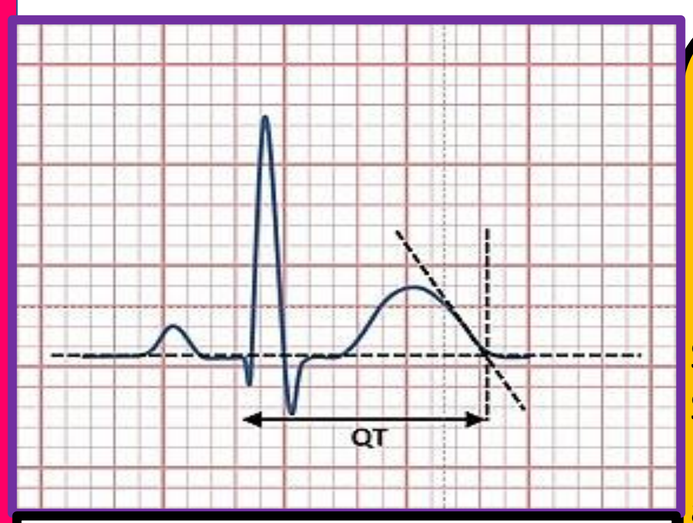
Introduction

Long QT syndrome is a genetically transmitted cardiac arrhythmia caused by ion channel protein abnormalities. It is characterized by ECG abnormalities and a high incidence of syncope and sudden cardiac death. Long QT syndrome can be mistaken for palpitations, neurocardiogenic syncope, and epilepsy. Long QT syndrome is responsible for approximately 1000 deaths each year in the United States. Almost 40% of people with long QT syndrome had a delayed diagnosis because the cause was labelled as fits. The prognosis for patients with long QT syndrome who have been treated with beta-blockers is satisfactory.



How do you measure QT interval?
measure from the onset of the Q wave 'to the intersection between the baseline and a tangent taken from the steepest part of down sweep of the T wave, not where the T wave reaches the baseline. Then correct for
heart rate with Bazett's formula:
corrected QT = QT/ √RR(in seconds)

Case Report

She was one of twin and born at 38 weeks with birth weight of 2.9kg. No neonatal problem. Sibling died as sudden unexpected death at 4 weeks.

At 16 months of age she presented with unresponsive episode and floppy, eyes rolled back and lips were blue. She was treated as suspected sepsis and febrile convulsion. Discharged home with plan of Outpatient MRI scan. She presented again within 2 days with another episode of floppy, became stiff, arched back, eyes rolled back and unresponsive for 2 minutes. It was taken as seizures, so urgent EEG and MRI arranged. During EEG, ECG was done noted to have prolonged QT. she was discharged with the plan of cardiology follow up.

In 2 weeks' time she had cardiac arrest due to ventricular Tachycardia while she is in the bus with her mum. Mum didn't recognize that child was unwell and she was resuscitated by bystander and taken to A&E . Epicardial pace maker was inserted next day.
Post cardiac arrest her development was regressed so neurorehabilitation follow up arranged and she was discharged home with propranolol and cardiology follow up.

Conclusion

This case illustrates

1. Importance of doing ECG in all the patients who is presenting with seizures.
2. Importance of interpreting ECG.
3. Appropriate early referral is needed when somebody diagnosed with Prolong QT syndrome.
4. Importance of training parents and carers how to recognise when the child is unwell.
5. Parents and carers of children with long QT syndrome need basic life support training
6. Significance of long QT in a sibling of sudden infant death syndrome.

References

1.Management of Patients with Long QT Syndrome
Yongkeun Cho, MD
Department of Internal Medicine, Kyungpook National University Hospital, Daegu, Korea
Korean Circ J 2016 Nov46 (6)747-752 published online October 20th 2016

2. The long QT syndrome in children.
An international study of 287 patients.
A GarsonJr , M Dick2nd , A Fournier , P C Gillette , R Hamilton , J D Kugler , G F van Hare3rd , V Vetter , and G W Vick3rd
1993https://doi.org/10.1161/01.CIR.87.6.1866Circulation. 1993;87:1866–1872