A case of Long QT syndrome

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Introduction

Long QT syndrome (LQTS) is a group of genetically distinct arrhythmogenic disorders resulting in abnormal cardiac sodium and potassium ion channels causing delayed repolarization of the heart[1]. This syndrome can present clinically as syncope, seizures, or sudden cardiac death secondary to its characteristic ventricular arrhythmia of torsades de pointes[2,3]. The clinical and electrocardiographic description of long QT syndrome was first reported in 1957 by Anton Jervell and Fred Lange Nielsen[4]. Disease prevalence is estimated to be from 1 in 5, 000[5] to 1 in 20, 000[6]. Schwartz *et al*[7] proposed some major and minor criteria which have been modified for children (Table 1). According to these criteria, scores of 1 or less are designated as low probability, 2-3 as medium probability and 4 or more as high probability.

Table 1: ***Diagnostic criteria for Long QT syndrome***[7]

Diagnostic criterion	Points
ECG findings	
Corrected QT interval	
≥480 milliseconds	3
460–470 milliseconds	2
450 milliseconds (in males)	1
Torsade de pointes	2
T-wave alternans	1
Notched T wave in 3 derivation	1
Low heart rate for age	0.5
Clinical history	
Syncope	
With stress	2
Without stress	1
Congenital deafness	0.5
Family history	
Siblings with LQTS	1
History of sudden death before 30 years of age	0.5

Case report

A 5 year old boy presented to us with an episode of loss of consciousness while playing which lasted for 10 minutes. There was no pallor or cyanosis, no generalized tonic movements, no urinary or faecal incontinence and no post-ictal drowsiness. He has had 4 episodes of simple febrile convulsions since 9 months of age, the last being at 4 years of age. He was born following a normal vaginal delivery without any antenatal or postnatal complications. His mother had febrile convulsions in childhood. He had no history of familial unexplained sudden deaths at younger ages or syncopal attacks or congenital deafness.

On examination, his pulse rate was 96 beats per minute and the pulse was regular and of good volume. His blood pressure was 90/60 mmHg. Precordial auscultation was unremarkable with dual rhythm without any murmurs or fixed split of heart sounds. Full blood count, total and ionized calcium levels, serum electrolytes and random blood sugar were within normal limits. The electroencephalogram (EEG) was normal. His electrocardiogram (ECG) revealed sinus rhythm, a heart rate of 100 per minute, a normal axis, QRS duration of 40 milliseconds and corrected QT (QT_c) interval of 480 milliseconds (prolonged for his age), and no T-wave abnormalities or dysrhythmia (Figure 1).

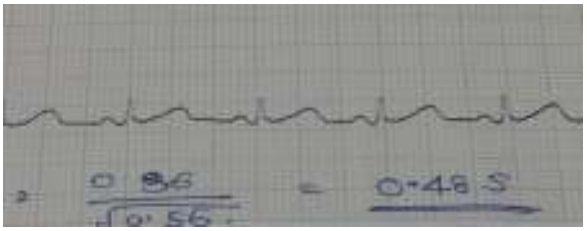

Figure 1: ***ECG of the patient***

Due to the possibility of the seizure being due to cardiac syncope, child was referred to the Cardiac Electrophysiology Unit of the National Hospital of

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Sri Lanka where long QT was confirmed and he was started on beta blockers.

Discussion

Although a rare congenital disorder, LQTS can have devastating consequences, ranging from syncope to sudden cardiac death[8]. Torsades de pointes and ventricular fibrillation triggered by LQTS can cause syncope, seizures, or sudden death depending on the duration of the ventricular arrhythmia[8]. The clinical manifestation of LQTS in children is most often a syncope brought about by exercise or fright; some events occur during sleep[9]. The diagnosis is based on ECG and clinical criteria (Table 1). Characteristic ECG findings are long QTc interval for age, T wave abnormalities and torsade de pointes-type ventricular tachyarrhythmias induced by exercise[3,4]. In our patient using the modified Schwartz criteria[7], the overall score was 5 (QTc 480 milliseconds - 3 points and syncope with stress - 2 points) which makes the diagnosis of LQTS highly probable. Treatment of a patient with LQTS includes medical management (usually ß-blocker therapy), left cardiac sympathetic denervation and occasionally, implantable cardioverter-defibrillator (ICD) placement[10]. Beta-blockers are the first-line treatment for LQTS and all patients should receive them as the initial therapy[10]. They provide a reduction in the risk of cardiovascular events of up to 64%[8]. We wish to emphasize that LQTS may present with stereotypical clinical events associated with seizure activity which need appropriate evaluation.

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