Overt long QT syndrome in children presenting with seizure

Faisal Zafar, Fawad Saleem, Safdar Hussain, Nuzhat Noreen, Naveed Anjum, Zia ul Rehman

Department of Pediatric Neurology, The Children's Hospital and The Institute of Child Health, Multan, Pakistan

18TH ICNC 2024 Footprints to the future
INTERNATIONAL CHILD NEUROLOGY CONGRESS
Cape Town, South Africa I May 6-10, 2024

Introduction

Long QT syndrome (LQTS) indeed falls within the category of rare inherited disorders, with an estimated prevalence ranging from approximately 1 in 2,500 to 1 in 10,000 individuals. LQTS is characterized by an abnormality in the heart's electrical system, leading to a lengthening of the QT interval on an electrocardiogram (ECG). This prolonged QT interval is associated with an elevated risk of arrhythmogenic syncope, where individuals may experience fainting or loss of consciousness due to irregular heart rhythms. LQTS can predispose affected individuals to polymorphous ventricular tachycardia, a type of abnormal rapid heart rhythm that can be life-threatening if not managed or treated promptly. Sudden cardiac death is a severe complication linked to LQTS, particularly in cases where arrhythmias provoke a dangerously irregular heartbeat, leading to an abrupt cessation of cardiac function. Clinically, children with LQTS typically have seizures, syncope, or sudden cardiac death syndrome, which is often attributable to its distinctive ventricular tachyarrhythmia, such as torsades de pointes (TdP).6 Regrettably, individuals with LQTS sometimes receive incorrect diagnoses as having epilepsy or seizure disorders and subsequently antiepileptic drugs (AEDs) are given to them.7 It is noteworthy that the literature has documented around 11% of patients with LQTS to have experienced seizures or episodes resembling seizures, with 1.6% having an erroneous epilepsy diagnosis.

Objectives

To evaluate the clinical spectrum and relevant risk factors of long QT syndrome (LQTS) among children with seizure disorders presenting at a leading tertiary childcare hospital of South Punjab, Pakistan.

Material and Methods

A total of 310 children of either gender, aged up to 16 years, who had been diagnosed with seizures previously were analyzed. Diagnostic workup for seizures done in form of electroencephalogram, an ECG, and Holter monitoring as per indication. The LQTS was diagnosed including a corrected QT (QTc) interval above 440 msec in either lead II or V5 and an absolute QT interval > 98th percentile for heart rate in lead V5, in combination with clinical parameters.

Results

In a total of 310 children with seizure disorders, 172 (55.5%) were boys. The mean age was 5.4±3.6 years. LQTS was observed in 6 (1.9%) children. Age (5.8±2.8 years vs. 5.3±3.6, $p< 0.0001$), corrected QT intervals (531.4±25.6 ms vs. 384.3±14.3 ms, $p<0.0001$), Schwartz scores > 3.5 ($p<0.0001$), and presentation with syncope (33.3% vs. 3.6%, $p=0.0003$) were significantly associated with LQTC. Family history of seizures ($p=0.0144$), deafness ($p<0.0001$), sudden unexplained death ($p=0.0164$), and abnormal neurological examination ($p=0.0036$) were linked with LQTS.

Conclusion

Burden of LQTS was relatively low in children presenting with seizure disorders. The identification of gender, age, ECG parameters, Schwartz scores, syncope, and familial history as associated factors underscores the multifaceted nature of LQTS diagnosis.

References

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Acknowledgement and Contact

Department of Pediatric Neurology, The Children's Hospital and The Institute of Child Health, Multan, Pakistan.

Email : drfaisal867@yahoo.com

Contact #: +92-300-7195420