Case Report

Diagnostic Lacunae and Implications of an Automated Implantable Cardioverter Defibrillator Implantation in a Child with Type 3 Long QT (LQT3) Syndrome

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ABSTRACT

A diagnosis of congenital long QT interval syndrome based on history and electrocardiogram was made in a child in the absence of readily available genetic testing. A genotype 3 (LQT3) was suspected after exclusion of other variants as the child was non-responsive to beta-blocker and sodium channel blocker medication. As the child continues to show episodic bradycardia, polymorphic ventricular ectopy, and T-wave alternans, a single-chamber automated implantable cardioverter-defibrillator implantation was done successfully. This report highlights how the diagnosis of LQT3 was arrived at as well as the anesthetic challenges in the management of patients with LQTS.

Keywords: Anti-arrhythmia agents/administration and dosage, defibrillators, electrocardiography, implantable, long QT syndrome/diagnosis, long QT syndrome/drug therapy, voltage-gated sodium channel blockers/administration and dosage

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INTRODUCTION

Long QT syndrome (LQTS) may arise due to the involvement of genes encoding critical ion channels of the heart (congenital LQTS) or may be caused by metabolic abnormalities or drugs (acquired LQTS).[1] In patients with congenital LQTS, there is heterogeneity in the dispersion of the repolarization phase of the myocardial fast action potential between the epicardial, mid-myocardial, and endocardial layers of the myocardium.[2] This heterogeneity may be amplified by sympathetic activity, resulting in the formation of re-entry circuits predisposing to generation of premature ventricular contractions (PVCs) that may initiate Torsades de Pointes (TdP).[2] Genetic testing for LQTS has diagnostic, prognostic, and therapeutic implications and the absence of its ready availability poses a major limitation in the clinical management of patients with this condition. This case report describes the implantation of a single chamber automated implantable cardioverter defibrillator (AICD) in a child with a congenital Type 3 LQTS in whom the diagnosis was arrived at by an elimination process as genetic testing was not readily accessible. Informed consent from the parents of the child and institutional ethical committee approval (SRC#CR25/2020) were obtained for publishing this report.

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CASE HISTORY

A 3-year-old boy (weight: 11 kg; height: 102 cm) sustained a syncopal attack that lasted about 20 minutes while at play. After resuscitation, he was shifted to the authors' institution for further management. The child had delayed gross motor development and on a previous occasion had tonic and clonic seizures. There was no history of cardiac disease, syncope, or sudden death among the close family members. He has three siblings with normal electrocardiogram (ECG) readings and none had any history of syncopal episodes. The corrected QT intervals (QTc) on the ECGs of the father and mother were 450 ms and 420 ms, respectively. He was found to have macrocephaly with frontal bossing, exophthalmos, depressed nasal bridge, diffuse thyroid swelling, and short stubby fingers [Figure 1]. The ECG showed multiple PVCs, QT_C of 600 mc, and T-wave alternans [Figure 2a]. Transthoracic echocardiography revealed no structural cardiac anomaly. The child was diagnosed to have LQTS based on the European Society of Cardiology (2015) guidelines and Schwartz and colleagues' criteria.[3,4] Assuming that the child had the relatively common genotype 1 congenital LQTS which is a K^+ channelopathy, medical management was initiated with: syrup propranolol (1 mg initially increased to 7 mg TID), and syrup magnesium sulfate (200 mg BID). Serum K^+ levels were maintained within the normal limits. The response to these medications was poor as the QTc remained >500 ms, with the persistence of PVCs, frequent episodes of non-sustained ventricular tachycardiac (VT), and the abnormal T wave morphology (T-wave alternans). Hence, the possibility of a Type 3 LQTS (LQT3) was considered and tablet flecainide (40 mg BID) and syrup mexiletine (30 mg TID] were prescribed as additional medication. After 4 weeks of medical therapy, the frequency of non-sustained VTs decreased, however, there was no change in the QTc and the PVCs persisted [Figure 2b]. Therefore, a decision to implant an AICD was taken.

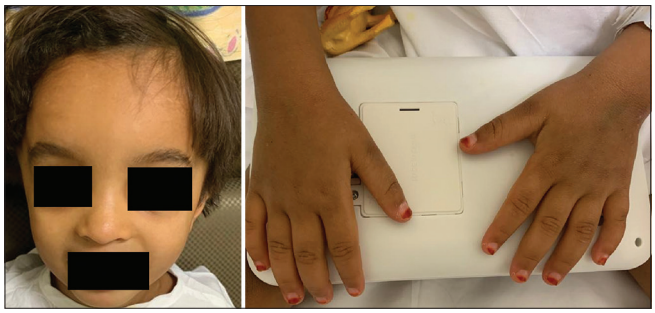

Figure 1: Picture of the child showing the frontal bossing, exophthalmos [eyes covered], depressed nasal bridge [left frame] and stumpy fingers [right frame]

Preoperatively, all the antiarrhythmic medications were continued. The child was premedicated with tablet midazolam (1 mg) in the operation holding area under ECG monitoring. In the operation room, transcutaneous pacemaker/defibrillator patches were applied before induction of anesthesia. Normothermia was maintained using a forced-air warming system (Bair Hugger system). General anesthesia was administered under standard ASA monitoring with fentanyl citrate 25 mcg IV, etomidate 30 mg IV, lignocaine 20 mg IV, and isoflurane (0.5 MAC). The trachea was intubated after administering inj cisatracurium besylate. Right radial artery and right internal jugular vein were cannulated under ultrasound guidance. Multi lead ECG monitoring was instituted to monitor the QTc values throughout the procedure [Figure 3]. Pressure control mode of mechanical ventilation was initiated to achieve normoxia and normocapnia while avoiding a sustained high intrathoracic pressure. Care was also taken to avoid electrolyte abnormalities. The epicardial leads were implanted through a lower mini-sternotomy and a single chamber AICD device (Mirro MRI Single Chamber, DVME3D1 [Medtronic Inc, Minneapolis, MN, USA]) was placed in a supra-diaphragmatic pouch. The AICD was interrogated by inducing a tachyarrhythmia followed by delivery of a shock to revert back to sinus rhythm [Video Clip 1]. The AICD was programmed for the ventricular tachycardia zone with anti-tachycardia pacing/burst pacing (ATP) (three attempts) followed by a shock (one 25 Joules shock and three 35 Joules shocks). For the ventricular fibrillation zone, the AICD was programmed to deliver shock/shocks (35 Joules six times) with ATP while charging. A backup rate of 60 beats/min in synchronous mode in case of bradycardia and 60 beats/min in asynchronous mode in case of a ventricular standstill following cardioversion was

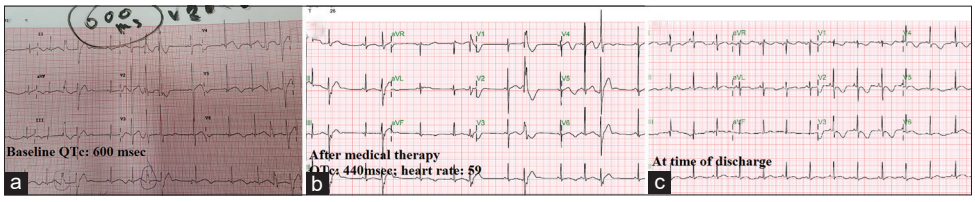

Figure 2: (a- c) The baseline electrocardiogram [a], the electrocardiogram after initiation of Beta and Na channel blockers showing the T wave alternans and persistent premature ventricular contractions [b] and electrocardiogram at discharge after AICD implantation [c]

Figure 3: Monitor displaying the hemodynamic parameters with QTc monitoring

ensured. Deep tracheal extubation was done with isoflurane anesthetic and the hemodynamic parameters were monitored for the next 24 hours. The child was discharged home on the 10th day with advice to continue the propranolol and flecainide medication. An ECG taken just prior to discharge showed no PVCs [Figure-2c]. On 6-month follow-up, though the QTc remained prolonged (500 ms), interrogation of the AICD revealed no episodes of nonsustained VTs and no shocks had been delivered. However, it has to be kept in mind that the device may not record only PVCs. In addition, the device may not record any short run of non-sustained VT if the cycle length is slow.

DISCUSSION

A QTc interval (Bazette formula) of >440 ms is considered prolonged and the current child as well as his parents had QTc prolongation by this criterion.[2] Medical therapy with beta-blockers, electrolyte supplementation, and sodium channel blockers were initiated with a poor response. The child had a successful AICD implantation subsequently. A Brugada syndrome was excluded as the child had prolonged QT interval.[5] An arrhythmogenic right ventricular dysplasia was excluded as the echocardiography showed no fatty or fibrous-fatty infiltrates of the right ventricle myocardium and the ECG did not display T wave inversion with prolonged S-wave upstroke in V_{1-3} electrocardiogram leads.[5]

Congenital LQTS is a rare inherited cardiac channelopathy with a prevalence of 1 per 1100–3000 of the population.[2] Though over 600 mutations in 14 susceptible genes have been identified, three genotypes, that is, LQT1, LQT2, and LQT3 account for 75% to 95% of congenital LQTS cases.[6] LQT1 is associated with mutations in *KCNQ1* gene with loss of function of the slow component of the delayed rectifier potassium current channel (I_{Ks}) which play a major role during the plateau of phase 2 action potential countering the depolarizing influence of the L-type Ca2+ currents.[2,6] The loss-of-function mutations in the KCNQ1 gene create a substrate in which the defective channel is unable to adapt to beta-adrenergic stimulation. Patients with LQT1 are very responsive to beta-blockade therapy.

LQT2 is caused by a mutation in the KCNH2 gene that is responsible for the rapidly activating component of the delayed rectifier potassium current [(Ikr) which assumes significance towards the end of the plateau phase (phase 2) of action potential.[2,6] Beta-blockade and potassium supplementation are the first line of therapy for these patients.[6] In the current child, beta-blockade and normalization of electrolytes did not abolish the PVCs suggesting that the child may not have a K^+ channelopathy.

LQT3 is caused by gain-of-function mutations in the cardiac sodium channel gene (SCN5A), resulting in the Na+ channel to open repetitively.[2,6] As in LQT1 and LQT2 patients, the first line of therapy in LQT3 patients is beta-blockade followed by sodium channel blockers with drugs like mexiletine and flecainide. The clinical effectiveness of these drugs in shortening the QT interval may not be uniformly successful as it may be mutation specific.[7,8] AICD with pacemaker capabilities may be indicated in patients with LQT3 to provide heart rate consistency, decrease repolarization heterogeneity, and avoid bradycardia that predisposes them to TdP.[9,10] In the current child medical management was not clinically effective necessitating an AICD implantation.

In conclusion, clinical manifestations, family history along with electrocardiogram features remain the mainstay of diagnosing of LQTS. Many anesthetic drugs and airway maneuvers in light planes of anesthesia have a tendency to prolong the QT interval predisposing the myocardium to malignant arrhythmias and anesthesiologists must be prepared to recognize and treat them promptly. Hence a working knowledge of the syndrome is important to anesthesiologists.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Kies SJ, Pabelick CM, Hurley HA, White RD, Ackerman MJ. Anesthesia for patients with congenital long QT syndrome. Anesthesiology

2005;102:204-10.
2. Booker PD, Whyte SD, Ladusans EJ. Long QT syndrome and anaesthesia. Br J Anaesth 2003;90:349-66.
3. Priori SG, Blomström-Lundqvist C, Mazzanti A, Blom N, Borggrefe M, Camm J, *et al.* 2015 ESC Guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death: The Task Force for the Management of Patients with Ventricular Arrhythmias and the Prevention of Sudden Cardiac Death of the European Society of Cardiology (ESC). Endorsed by: Association for European Paediatric and Congenital Cardiology (AEPC). Eur Heart J 2015;36:2793-867.
4. Schwartz PJ, Moss AJ, Vincent GM, Crampton RS. Diagnostic criteria for the long QT syndrome. An update. Circulation 1993;88:782-4.
5. Ferri FF. Ferri's Clinical Advisor: Elsevier,Inc Philadelphia, USA; 2017:p. 736.
6. Staudt GE, Watkins SC. Anesthetic considerations for pediatric patients with congenital long QT syndrome. J Cardiothorac Vasc Anesth 2019;33:2030-8.
7. Barsheshet A, Dotsenko O, Goldenberg I. Genotype-specific risk stratification and management of patients with long QT syndrome. Ann Noninvasive Electrocardiol 2013;18:499-509.
8. Ruan Y, Liu N, Napolitano C, Priori SG. Therapeutic strategies for long-QT syndrome: Does the molecular substrate matter? Circ Arrhythm Electrophysiol 2008;1:290-7.
9. Pérez-Riera AR, Barbosa-Barros R, Daminello Raimundo R, da Costa de Rezende Barbosa MP, Esposito Sorpreso IC, de Abreu LC. The congenital long QT syndrome type 3: An update. Indian Pacing Electrophysiol J 2018;18:25-35.
10. Schwartz PJ, Spazzolini C, Priori SG, Crotti L, Vicentini A, Landolina M, *et al.* Who are the long-QT syndrome patients who receive an implantable cardioverter-defibrillator and what happens to them? Data from the European long-QT syndrome implantable cardioverter-defibrillator (LQTS ICD) registry. Circulation 2010;122:1272-82.