Case Reports

Incorporating an electrocardiogram in the preoperative evaluation of sensorineural hearing loss: a necessity

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Introduction

Jervell and Lange-Nielsen syndrome (JLNS) is a form of long QT syndrome (LQTS) associated with severe, bilateral sensorineural (SN) deafness. It was first described by Anton Jervell and Fred Lange-Nielsen in 1957[1]. Cochlear implant is commonly performed for profound SN deafness but a preoperative ECG is not insisted upon in many cases especially if child is not symptomatic. We describe two cases of SN deafness who underwent cochlear implant in early childhood and were diagnosed to have LQTS on follow up.

Case 1

A 7 year old boy, who was congenitally profoundly deaf, had a cochlear implant inserted at the age of 2 years. At 6 years of age he had two attacks of loss of consciousness within a one month period while he was at school. The first episode was diagnosed as syncope following exertion and the second episode was reported as an episode of atonic seizure. The episodes were initially thought to be neurological. Magnetic resonance imaging (MRI) of his brain was not done due to the presence of the cochlear implant. The electroencephalogram (EEG) was reported to be normal. After the second episode the child was started on levetiracetam. Child was also noticed to have bradycardia for which a cardiologist was consulted and an electrocardiogram (ECG) was done which revealed a QTc of 0.5 seconds (upper limit of normal 0.44 seconds), with a heart rate of 75 beats per minute (Figure 1).

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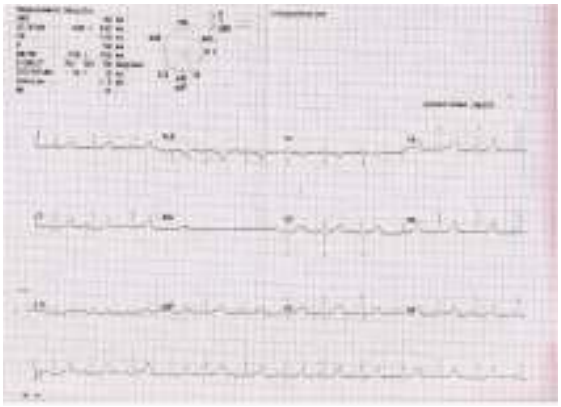

Figure 1: ***Electrocardiogram of patient***

Child was started on beta–blockers (propranolol) and anti-epileptics were gradually tapered and stopped. Child had no repeat episode of unconsciousness for the past one year. Genetic testing showed homozygous mutation in the KCNQ1 gene (p.S253C:758C>G in exon 5) (Figure 2). S253C mutation identified in this patient has not been reported in the literature.

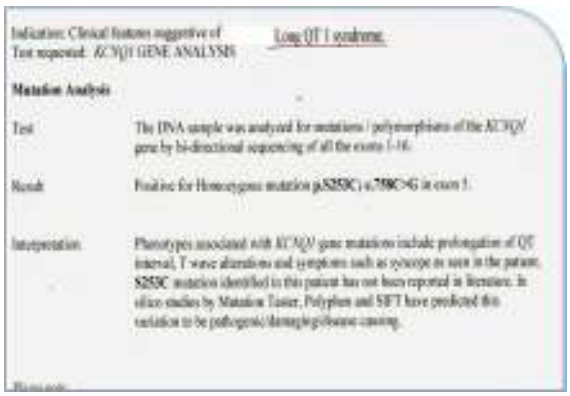

Figure 2: ***Genetic testing***

Case 2

A 13 year old boy presented to emergency with a history of fainting while playing cricket in school one hour back. At the time of presentation to the emergency room, child was drowsy and had good volume pulses. Heart rate was found to be 200 beats per minute and an ECG was done which showed a ventricular tachycardia. Child was given injection amiodarone after which he reverted back to sinus rhythm. Repeat ECG showed increased QTc interval

of 0.62 seconds. Child also had a history of severe sensorineural deafness since birth for which he had undergone cochlear implantation at the age of one year. Child was started on propranolol and was advised to have genetic testing but they refused due to financial constraints.

Discussion

The 7 year old boy suffered from syncopal attacks on exertion, and was initially treated with anticonvulsant therapy. However, the seizures are now thought to have represented the cerebral effect of transient ischaemia occurring during episodes of arrhythmias. The 13 year old boy too had syncopal attacks and arrhythmias. The severity of the arrhythmias and their clinical presentation are known to vary in JLNS.

A QTc interval of 0.44 seconds or more in males and 0.46 seconds in females is diagnostic of LQTS. ECG abnormalities in JLNS, in addition to a prolonged QTc, can include abnormal T wave morphology: biphasic, inverted or bifid[2,3]. Multifocal ventricular ectopic beats, ventricular tachycardia, ventricular fibrillation and Torsade des Pointes can occur and are associated with a poor prognosis. Arrhythmias cause syncopal attacks and may lead to sudden death if not treated. The attacks can be precipitated by a sudden increase in autonomic activity, for example during exercise, or following emotional stress and loud sounds.

Diagnosis of JLNS is confirmed by detecting homozygous mutations in KCNQ1 and KCNE1 genes[4]. JLNS is the most severe variant of LQTS. By the time the child is 3 years old, half of them are symptomatic, and by the time they are 18 years of age, 90% have symptoms[4]. More than 50% of untreated children with JLNS die before 15 years of age[5,6]. The parents of both cases were found to have normal QTc. The importance of highlighting the sub-clinical cases in the family should be stressed, as severe symptoms or even sudden death can be triggered by drugs such as sotalol, amiodarone, quinidine, procainamide, erythromycin, terfenadine, fexofendine[7], haloperidol some tricyclic anti-depressants and fluoroquinolones, all of which may affect cardiac repolarisation[8,9].

The most important aims in treatment of LQTS are prevention of syncope, cardiac arrest and sudden death. According to Schwartz *et al* the effectiveness of beta-blocker therapy is only partial[5]. Goldenberg *et al* showed a 35% mortality rate for individuals receiving beta blockers exclusively[10]. Implantable cardioverter defibrillators (ICDs) should be considered in individuals with a history of cardiac arrest or failure to respond to other treatment[5].

Cardiac investigation should form part of the diagnostic work up of all patients with congenital or early onset hearing loss. JLNS may present in a potentially fatal manner but can be treatable. There are numerous reports in the literature of undiagnosed cases of JLNS with unpredictable preventable cardiac arrhythmias during surgery causing serious complications. Adu-Gyamfi *et al.*[11] reported a case of a child with congenital profound deafness, who was misdiagnosed as having epilepsy. While undergoing a CT scan of the brain under general anaesthesia he experienced polymorphous ventricular tachycardia on induction with halothane. In retrospect he was found to have JLNS.

In cases when the diagnosis of JLNS has already been made, pre-operative beta-blockade, with optimisation of the dose, should be commenced prior to the induction of anaesthesia, as anaesthesia increases the risk of ventricular arrhythmias. Drugs such as halothane, which sensitise the myocardium to the effect of catecholamines, should be avoided. Effective preoperative sedation, oximetry and avoidance of drugs which might further prolong the QT interval, ensuring a normal serum potassium and electrolytes and, above all, continuous ECG monitoring from the time the patient enters the anaesthetic room are all important aspects of management.

ECG is a simple and easy test to diagnose children with LQTS who have sensorineural hearing loss. Thus, all children with severe congenital sensorineural hearing loss having unexplained syncopal attacks or convulsions must be screened for JLNS with a 12 lead ECG.

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