SHORT COMMUNICATION

Long QT Syndrome in Children with Congenital Deafness

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The Jervell and Lange-Nielson syndrome(JLN) is an infrequent form of long QT syndrome (LQTS) in which prolonged QT interval and congenital deafness exist together. We attempted to identify patients with LQTS among 127 children (age 1.2-10 years) with congenital hearing loss. The corrected QT interval was measured from 12 lead electrocardiogram(ECG) , using Bazette's and Friedricia formulae.The QT interval was considered prolonged when it exceeded the upper limit of 440ms and 450ms, respectively. Ten children with congenital deafness had a corrected QT interval longer than 440ms. Although these children did not meet the definite criteria according to Schwartz parameters, all the 10 children could be defined as having intermediate probability of LQTS according to revised criteria. We advise that children with congenital deafness be screened for long QT syndrome .

Keywords : *Congenital deafness, ECG, QT interval*

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The long QT syndrome (LQTS) is an inherited arrhythmogenic disease. There is an increased susceptibility to life threatening ventricular arrhythmias. Two major forms of LQTS have been identified: (*i*) autosomal dominant Romanoward syndrome and (*ii*) autosomal recessive Jervell Lange-Nielson (JLN) syndrome with associated neuro-sensory deafness(1). The severity of clinical manifestations of LQTS is variable. At one end of the spectrum, there may be fullblown disease with markedly prolonged QT interval and recurrent syncope, and at the other end of the spectrum, there may be subclinical forms with borderline QT interval prolongation and no evidence of syncope or major arrhythmias. Thus, risk stratification becomes a crucial step for clinical management of these patients. We conducted this study to identify LQTS in a small population of children with congenital deafness.

METHODS

One hundred and twenty seven deaf children between the age group 1.2 and 10 years (64 boys) from a school for deaf children in Chennai were studied over a period of 6 months. Details were recorded with regard to history of seizures and syncope, family history, sibling history, developmen-tal milestones, and history of consanguineous parentage. All children were subjected to clinical examination by a cardiologist and electrocardiogra-phic assessment (ECG). A 12 lead ECG was recorded and the QT interval was measured in all children. Corrected QT interval was calculated by using the Bazette's(2) and Fridericia(3) formulae. Based on the ECG findings, clinical signs and symptoms, a modified Schwartz criteria(4) was employed to know the probability of LQTS in a given child.

RESULTS

Eighty children had normal QTc; 37 children had borderline prolonged QTc and 10 children had obviously prolonged QTc. The median age of the 10 children (6 males) was 4.1 year (range, 1.2–5.2) with QTc varying from 460 to 500 ms. The longest QTc recorded was 500ms. None of the 10 children with prolonged QTc had any symptoms, or family history

WHAT THIS STUDY ADDS ?

- Children with congenital deafness should be evaluated for presence of long QT syndrome.

of deafness or sudden death. However, one child aged 1.9 years, who did not have QT prolongation in ECG died suddenly while playing. There was history of consanguineous parentage in 40.9 % of children examined. Sibling history of deafness was present in 6.3 % of cases and parental deafness in 1.6 % of cases. Inverted 'T' waves were present in the ECG in 44% of cases. Broad based 'T' waves and increased QT dispersion were seen in 1.6% of cases each. Bifid 'T' wave was present in 4.7 % of cases.

DISCUSSION

LQTS associated with congenital deafness (JLN syndrome) is a heritable disorder of the heart and the hearing system(1). Symptoms are due to malignant ventricular arrhythmias and are associated with a propensity to syncope and sudden cardiac death. Consanguineous marriages are not uncommon in LQTS population especially in JLN syndrome. We also documented a high prevalence of consanguinity in our cases.

There are 2 cardinal manifestations of LQTS (*i*) syncopal episodes, and (*ii*) ECG abnormalities. The syncopal episodes are characteristically associated with sudden increase in sympathetic activity as in stress, emotional, and physical activity(5). In the present study, one child died while playing. The ECG changes include QT prolongation(6), broad based or notched or late onset 'T' waves, 'T' wave alternans *etc*(6,7). In the present study we found prolonged QTc in 7.9 % of cases. Molecular screening is required for a precise diagnosis. However we could not do it because of logistic and financial constraints.

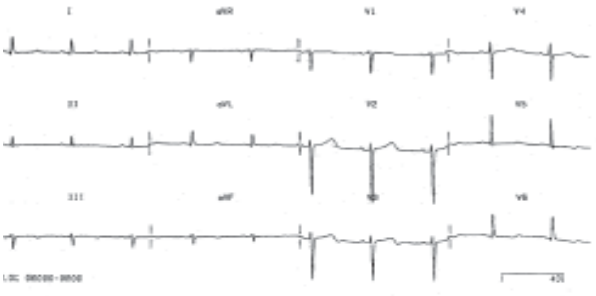

FIG. 1 *An example of child's ECG showing LQTS.*

We started these 10 children on prophylactic betablockers as also suggested earlier(8). Parents were alerted to avoid drugs that prolong QT interval, and to refrain these children from undue physical exertion, sudden exposure to cold, sudden awakening from sleep, fright and loud noise– the factors which could precipitate a sudden cardiac event in patients with LQTS.

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