A study of electrocardiographic changes in congenital deaf school children

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Abstract

Background There is evidence of cardiac abnormalities in congenital deaf school children, together called as Jervel Lange Nielsen syndrome or Long QT syndrome.

Aim The main aim was to study the electrocardiographic changes in congenital deaf children.

Materials and Methods Fifty congenital deaf children aged 6–18 years were selected. ECG was taken in lead II, at rest and after exercise, as some are known to exhibit the abnormality after exercise. The child was made to run on the Tread mill till exhaustion. Corrected QT interval (QTc) was calculated by Bazett's formula $QTc = QT / \sqrt{R-R}$. ECG was also analyzed for other abnormalities like Twave changes, ST depression, rhythm abnormalities etc.

Results Out of 50, 2 children showed resting QTc of 0.45 sec which is diagnostically high. Mean value of QTc in deaf children(Cases) before exercise was 0.4111 ± 0.0271 sec and in controls 0.379 ± 0.020 sec. Mean value of QTc after exercise in deaf(cases) was 0.403 ± 0.028 sec and in controls 0.376 ± 0.021 sec. Eight deaf children showed ST depression and 2 biphasic T and 2 notched T waves. Thirty-three deaf children's parents had consanguineous marriage.

Conclusion The results were explained on the basis of ion channelopathy in heart and inner ear which predisposes to sensorineural hearing loss and cardiac abnormality.

Keywords Jervel-Lange Nielsen syndrome · Long QT syndrome · Ion channelopathy · Congenital deafness · Consanguineous marriage

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Introduction

Children born deaf become deaf mute and they can not bring out their problems like other children, problem remains unrecognized, might go to a “no return stage”.

There are evidences that deaf children are more threatened than the general population by dangerous heart arrhythmias [1].

In 1957, Jervel and Lange Nielsen for the first time described four siblings in whom deaf mutism was associated with peculiar heart disease. Deaf mute children who otherwise seemed to be quite healthy suffered attacks of fainting, attacks were often provoked by exercise or fear. Three of the four children died suddenly [2]. This is named Jervel - Lange Nielsen Syndrome or Surdo cardiac Syndrome where there is prolongation of QTc on ECG.

Investigators have suggested the use of provocative tests such as isoproterenol infusion and Valsalva maneuver or exercise for diagnosis of long QT syndrome [3]. Surprisingly deaf children exhibit occasional ECG abnormality at rest but plethora of abnormalities after exercise viz, sinus arrhythmias, sinus pauses, ST depression, T inversion, biphasic-T, notched- T, T- alternans, nodal ectopics and junctional rhythm[4].

Consanguineous marriage of parents is known to be associated with cardiac abnormalities in the congenital deaf children. India is a country where consanguineous marriages are at high rate and it is advisable to perform an ECG at least once in deaf children, especially in countries where consanguineous marriages are widely practised [5].

As there are only few studies from our country on these deaf children in relation to their cardiac dysfunction, this study is taken up in order to give a better life to the deaf child by diagnosing its cardiac dysfunction at an early stage.

Materials and methods

Children who are deaf by birth and aged between 6–18 years (cases) were selected for the study from local deaf

and dumb school. They were certified as having congenital sensory neural hearing loss by the ENT Surgeon.

Both male and female children were enrolled in the study group, as it was difficult to stick on to either of the sex because of the scarcity of the availability of the congenitally deaf children. Deaf children with conductive deafness, children having any cardiovascular disease, children taking Tricyclic antidepressants, antiarrhythmic agents and phenothiazene drugs, as these drugs are known to cause prolongation of QTc on ECG were excluded from the study. Fifty matched controls were selected from local nearby school.

Study was conducted after bringing the children to the lab. Five children were brought from their respective schools at each time.

Along with deaf children, their respective class teachers were also brought because of language problem. Teacher used to convey to deaf children in sign language.

After bringing the children to the lab, they were made to relax for 5 minutes. Then, history was noted. In subjects, history regarding deafness, family history of deafness, history of consanguineous marriage, history of any cardiovascular disorder was elicited, followed by complete general and systemic examination.

Electrocardiogram was recorded at rest using cardiart 108T, J8A,14901 in lead II. About 20 complexes were recorded.

Children were made to run on the tread mill at the speed of 3 km per hour and at an inclination of 7° till they got exhausted. Once they got exhausted and said they can not run any more, they were immediately made to lie down on the couch and ECG was recorded as soon as possible. Recording was done in lead II and about 20 complexes were recorded.

QTc i, e interval between start of QRS complex and end of T wave was calculated in seconds. Since QTc interval changes with heart rate corrected QT interval [QTC] was calculated using Bazet's formula [6], $QTc = QT / \sqrt{R-R}$

Apart from QTc, other abnormalities like T alternans, biphasic T, T wave inversion, ST depression, sinus pauses, ectopic beats etc were also noted.

Results

Subjects selected for this study were of comparable ages and physical characteristics i.e. height, weight and BMI matched. Results were analyzed by applying paired and unpaired “t” test [7, 8].

The age (in years) of cases ranged from 10 to 18 years, the mean being 13.9 ± 2.15 and of controls ranged from 10 to 18 years, the mean being 13.8 ± 1.7 .

Out of the 50 cases and 50 controls, 39 were male and 11 were female.

The mean height (in mt) in cases was 1.43 ± 0.11 and in controls 1.42 ± 0.11 .

The mean weight (in kg) in cases was 34.8 ± 7.33 and in controls 34.82 ± 7.9 .

The mean BMI in cases was 16.82 ± 2.27 and in controls 16.86 ± 2.27 .

The mean body temperature (in degree celsius) in cases was 36.9 ± 0.9 and in controls 36.8 ± 0.7 .

The mean hemoglobin concentration (in g%) in cases was 11.8 ± 2.2 and in controls 12.0 ± 2.1 .

The above data of baseline characteristics is shown in Table 1.

Table 1 Data of baseline characteristics

Characteristic	Cases	Controls	Significance
Age (in years)	13.9 ± 2.15	13.8 ± 1.7	$t = 0.3590$ $p > 0.05$ (NS)
Males	39 (78%)	39 (78%)	NS
Females	11 (22%)	11 (22%)	NS
Height (in mt)	1.43 ± 0.11	1.42 ± 0.11	$t = 0.2783$ $p > 0.05$ (NS)
Weight (in kg)	34.8 ± 7.33	34.82 ± 7.9	$t = 0.01309$ $p > 0.05$ (NS)
Body mass index (BMI) (kg/m ²)	16.82 ± 2.27	16.86 ± 2.27	$t = 0.08784$ $p > 0.05$ (NS)
Body temperature(in °C)	36.9 ± 0.9	36.8 ± 0.7	$t = 0.0767$ $p > 0.05$ (NS)
Hemoglobin (in g%)	11.8 ± 2.2	12.0 ± 2.1	$t = 0.4691$ $p > 0.05$ (NS)

NS: Not significant

Before exercise

Two children showed resting QTc of 0.45 sec which is diagnostically high, according to Schwartz criteria. The mean values of QTc before exercise in deaf (cases) and control group were 0.4111 ± 0.0271 sec and 0.379 ± 0.020 sec respectively. Unpaired “t” test shows $p < 0.001$ and it is highly significant. Results are shown in Table 2 and Fig. 1.

After exercise

The mean values of QTc after exercise in deaf (cases) and control group were 0.403 ± 0.028 sec and 0.376 ± 0.021 sec respectively. Unpaired “t” test shows, $p < 0.001$ and it is highly significant. Results are shown in Table 2 and Fig. 2.

Table 2 Mean QTc values and their significance

	QTc – Before exercise (BE)	QTc – After exercise (AE)	Comparison between BE and AE
Cases n = 50	0.411 ± 0.0271 sec	0.403 ± 0.028 sec	$t = 0.65$ p = 0.05 (NS)
Controls n = 50	0.379 ± 0.020 sec	0.376 ± 0.021 sec	$t = 0.82$ p = 0.05 (NS)
Comparison between deaf and controls	$t = 6.67$ p < 0.001 (HS)	$t = 5.42$ p < 0.001 (HS)	

QTc = Corrected QT interval

NS = Not significant

HS = Highly significant

Fig. 1 Mean QTc values before exercise in cases and controls

Fig. 2 Mean QTc values after exercise in cases and controls

Effect of exercise

The mean values of QTc in deaf children (cases) before and after exercise were 0.411 ± 0.027 sec and 0.403 ± 0.028 sec respectively. Paired “t” test shows $p > 0.05$ and it is not significant. Results are shown in Table 2.

The mean values of QTc before and after exercise in controls were 0.379 ± 0.020 sec and 0.376 ± 0.021 sec respectively.

Paired “t” test shows, $p > 0.05$ and it is not significant.

Results are shown in Table 2.

Other abnormalities on ECG

Eight deaf children showed ST depression, 2 showed biphasic T wave and 2 showed notched T wave after exercise. Results are shown in Table 3. Control children did not exhibit any such changes.

Other associated findings

33(66%) deaf children had their parents with consanguineous marriage and 7(14%) control children had their parents with consanguineous marriage, the difference being statistically significant ($p < 0.001$). The results are shown in Table 4.

Table 3 Other ECG abnormalities

Parameter	Number	Percentage
ST segment depression	8	16%
Biphasic T wave	2	4%
Notched T wave	2	4%

Table 4 Proportion of consanguineous marriage of parents

	Number	Percentage
Cases (n = 50)	33	66%
Controls (n = 50)	07	14%
Significance	‘t’ = 6.54 p < 0.001 (Highly significant)	

Four deaf children had their sisters and brothers who were also deaf. Five deaf children had syncopal attacks with stress, i.e. while participating in sports events.

Deaf children (cases) have higher QTc values than the control group and the difference is statistically highly significant. Exercise did not produce any significant effect on the QTc interval in both deaf as well as control group. Exercise produced other abnormalities like ST Depression, Biphasic T, notched T in deaf children and such changes were not seen in control group.

Discussion

Present study on 50 congenitally deaf children showed that deaf children have longer QTc values both at rest and after exercise when compared to control group. Exercise has not produced any effect on QTc, but has produced other changes like ST depression (8 children), notched T waves (2 children) and biphasic T waves (2 children).

These results correlate with results of Rokicki et al. [1], Shrivasthava et al. [4], Kocak et al. [5] Habbal and Mahony [9], A Ilhan et al. [10] who showed that deaf children have longer QTc when compared to normal children. RD Shrivasthava et al. also showed that in both normal as well as deaf children, exercise does not produce any significant changes in QTc from their resting values.

The explanation of long QTc in congenitally deaf children by the above investigators is, the genetic defect causing abnormal potassium ion channels which are present in inner ear causing sensory neural hearing loss and in cardia causing prolonged QTc.

Prolongation of QTc and other ECG findings show the vulnerability of the heart during repolarization. In patients who have long QTc, there will be irregular cardiac repolarization and an increased incidence of ventricular arrhythmias and sudden death. The syndrome can be caused by number of different drugs, by electrolyte imbalance and by myocardial ischemia. It can also be congenital. Three documented causes of the congenital form are mutations in KVLQT-1, HERG, and minK, three genes that encode parts of K⁺ channels. Mutations in the KVLQT-1 also cause deafness because the resulting K⁺ channel abnormality causes abnormal K⁺ movement across the stria vascularis [11].

Other associated findings

Present study showed 66% association of congenital deafness with consanguineous marriage. Four deaf children had family history of congenital deafness, that is, four deaf children had their sisters and brothers who were also deaf. This explains the strong association of consanguineous marriage and genetic defects and their inheritance. Five deaf children had history of syncopal attack or fainting attacks while participating in sports events. Abrupt onset of syncope without any forewarning, especially that occurring in the setting of acute auditory, physical or emotional arousal is almost always due to a transient arrhythmia with cerebral hypoperfusion. These syncopal events usually clear rapidly when an adequate rhythm is spontaneously reestablished [12].

Taking into consideration, the prevalence of consanguineous marriage in India, the present study has successfully enabled us to identify the cardiac defect in these congenital deaf children at an early stage and counsel the children and their parents so that they take adequate precautions at the appropriate situations.

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