Original Article

A Study of Cardiovascular Autonomic Functions in Congenitally Deaf Children with a Long QTC Interval

Physiology Section

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ABSTRACT

Background: Congenitally deaf children are at a risk of cardiac abnormalities in the form of the Long QT syndrome. It can be because of an intracardiac abnormality or autonomic dysfunction.

Aim: To study the cardiovascular autonomic functions in congenitally deaf children with a prolonged QTc interval.

Material and Methods: Congenitally deaf children who were aged between 6-18 years and having a prolonged QTc interval on ECG were selected as the cases and were compared with matched controls. Children with conductive deafness or those having any cardiovascular disorder were excluded. The tests which were done were Heart rate variation during deep breathing Heart rate response to standing, blood pressure response to standing and blood pressure response to a sustained hand grip. Statistical analysis was done by using the unpaired t test.

Results: No abnormal response was recorded in the cases or in the controls with respect to the above tests. The autonomic functions were normal.

Conclusion: Since there are no abnormalities in the cardiovascular autonomic functions in the congenitally deaf children, the autonomic imbalance theory for the Long QT syndrome stands invalid. The aetiology goes more in favour of intracardiac abnormality which predisposes to the Long QT syndrome.

Key Words: Autonomic function tests, Long QT syndrome, Congenital deafness; Jervel Lange Nielsen Syndrome; QTC interval

INTRODUCTION

Thomas Carlyle's statement **"Our main business is not to see what lies dimly at a distance but to do what lies clearly at hand"**, was an integral part of William Osler's Physiology of life, which he discussed in his 1913 address, **'A Way Of Life'**. In the genetic disorders of the heart, such as inherited long QT syndrome or hypertrophic cardiomyopathy, what lies clearly at hand has been clouded by a reduced penetrance and a variable expression of the phenotype. The more obvious and severe cases of these disorders were of course, identified and they provided the basis for the subsequent diagnosis and for the assessment of the clinical course; yet this phenotype was only the tip of the iceberg, representing only a small proportion of the patients. The broader spectrum of the disease which was manifested by a larger proportion of the patients at the base of the iceberg was not recognized until the diagnosis could be made by genetic analysis [1].

As we know, **'Today's children are tomorrow's citizens of the country'.** But for no fault of theirs, some children are born deaf and nature has been still harsh on them by not bestowing them with good hearts. There is some evidence that deaf children are more threatened than the general population by dangerous heart arrhythmias [2].

Children who are born deaf become deaf mute. Deaf mute children cannot bring out their problems like other children and the problems remain unrecognized unless they become severe enough to express themselves but by then, it would have reached a 'no return stage'.

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, which were often provoked by exercise or fear. Three of the four children died suddenly at the ages of 4, 5 and 9 years [3]. This condition was named as the Jervel – Lange Nielsen Syndrome or the Surdo cardiac Syndrome, where there was a prolongation of the QT interval on ECG.

The Long QT Syndrome represents the intriguing link between cardiac electrophysiology, the autonomic nervous system and lethal arrhythmias [4].

The aetiology of the Long QT syndrome is either an ion channellopathy or an autonomic imbalance. The ion channellopathy could be in the form of a K^+ ion channel defect which predisposes to congenital sensory neural hearing loss (the function of which is much required at the stria vascularis in the inner ear) and cardiac repolarisation defects in the form of a prolonged QT interval. The autonomic imbalance in these congenitally deaf children could also cause a prolonged QT interval [5]. So, the present study was taken up to know the status of the autonomic nervous system in these congenitally deaf children with a prolongation of the QT interval on ECG.

MATERIAL AND METHODS

This study was done on 20 congenitally deaf children with a prolongation of the QTc interval on ECG and those who were aged between 6–18 years. An equal number of healthy matched controls was included for the study.

Clearance from the ethical committee of the S. N. Medical College, Bagalkot, Karnataka, India, was obtained for this study. Informed consent was obtained from the guardians of each subject.

Children who were deaf by birth, who had a prolonged QTc interval (>0.43 seconds) on ECG and who were aged between 6–18

years were selected for the study. They were certified as having congenital sensory neural hearing loss by the ENT surgeon and the prolongation of the QTc interval on ECG was confirmed by the physician.

Both male and female children were enrolled in the study group, as it was difficult to stick on to either of the sexes because of the scarce availability of the cases. Deaf children with conductive deafness and children having any cardiovascular disease were excluded from the study group. Twenty matched controls were selected from a nearby local school.

Five children were brought from their respective schools at each time. Along with the deaf children, their respective class teachers were also brought because of a language problem. The teachers used to convey the messages to the deaf children in sign language.

After bringing the children to the laboratory, they were made to relax for five minutes. Then, their brief history was noted. After noting the history, their height and weight were noted and their body mass index was calculated.

Blood pressure in mm of mercury, pulse rate in beats per minute, respiratory ratein breaths per minute and body temperature in degree Celsius, were recorded subsequently. This was followed by a routine clinical examination of all the systems namely, the cardiovascular system the respiratory system, the central nervous system, and per abdominal examination. This was followed by an ECG recording in lead II at rest and after exercise (the child was made to run on the tread mill at an inclination of 7 degrees and at 3 km per secs of speed till exhaustion).

Once the ECG recording was over, child was made to relax for fifteen minutes. During this time, the QT interval i.e., the interval between the start of the QRS complex and the end of the T wave was calculated in seconds. Since the QT interval changes with the heart rate, the corrected QT interval [QT_C] was calculated by using Bazzet's formula [6], $QT_C = QT / \sqrt{R\text{-}R}$

For example, QT interval = 0.32 sec and R-R interval = 0.8 sec, QTc = $0.32/\sqrt{0.8} = 0.357$ seconds or 357 milliseconds.

Then, cardiovascular autonomic function tests were done.

AUTONOMIC FUNCTION TESTS

Four standard cardiovascular autonomic function tests were selected. The equipments which were required, included- a sphygmomanometer, an ECG machine, a hand – grip dynamometer, a couch and a chair. Due care was taken to remove the factors which could interfere with the results of the tests.

Methods of Assessment

Detailed instructions regarding the procedure which was employed for each test were given to the subjects. The different maneouvers were demonstrated to the subjects and they were trained to perform the tests. Actual recordings were made only after they were able to perform the tests satisfactorily. When the tests were performed by one subject, the others were asked to observe him/her performing the tests.

The following cardiovascular autonomic function tests were performed [7,8,9]:

1. Heart – rate variation during deep breathing.
2. Heart – rate response to standing {30: 15 ratio}.
3. Blood pressure response to standing.
4. Blood pressure response to a sustained hand grip.

The results of the tests were expressed as ratios and differences and they were analyzed by applying the unpaired "t" test [10, 11]. P values <0.05 were considered to be statistically significant.

RESULTS

The cardiovascular autonomic function tests of 20 congenitally deaf children with a prolongation of the QTc interval on ECG and those of an equal number of children as matched controls were studied.

The cases and the controls who were selected for this study were of comparable ages and physical characteristics i.e., height, weight and body mass index. The ages (in years) of the cases ranged from 10 to 18 years, their mean age being 13.9 ± 2.15 and those of the controls ranged from 10 to 18 years, their mean age being 13.8 ± 1.7.

Out of the 20 cases and 20 controls, 13 were males and 07 were females in each group.

The mean height (in meters) of the cases was 1.43 ± 0.11 and that of the controls was 1.42 ± 0.11.

The mean weight (in kg) of the cases was 34.8 ± 7.33 and that of the controls was 34.82 ± 7.9. The mean BMI (in kg/sq.m.) of the cases was 16.82 ± 2.27 and that of the controls was 16.86 ± 2.27. The mean body temperature (in degree Celsius) of the cases was 36.9 ± 0.9 and that of the controls was 36.8 ± 0.7.

The mean haemoglobin concentration (in gram %) of the cases was 11.8 ± 2.2 and that of the controls was 12.0 ± 2.1.

The above data of the baseline characteristics is shown in [Table/Fig-1].

TESTS FOR THE PARASYMPATHETIC SYSTEM

Heart Rate Variation during Deep Breathing (HRDB)

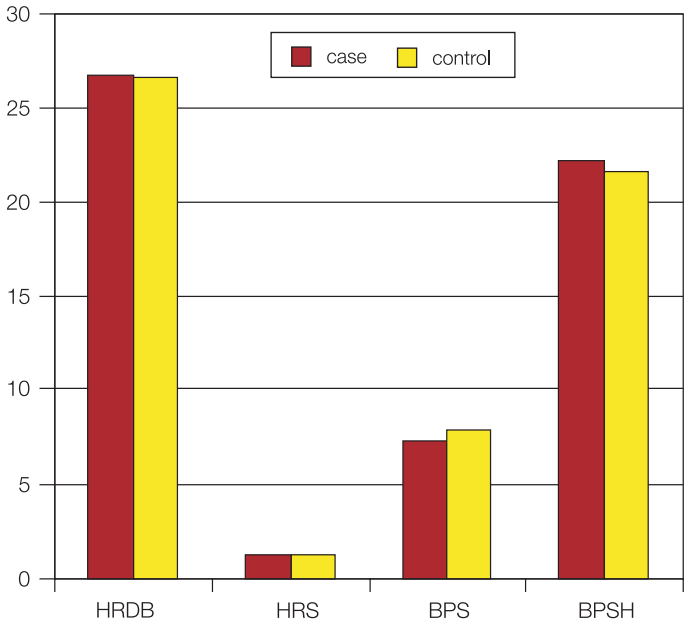
The mean heart rate variation (beats per minute) i.e., the difference between the maximum and the minimum heart rates during deep breathing in the cases was 26.76 ± 8.11 and that in the controls was 26.7 ± 6.25. The difference between the two groups was not statistically significant

Characteristic	Cases	Controls	Significance
Age (in years)	13.9 ± 2.15	13.8±1.7	t = 0.3590 p > 0.05(NS)
Males	13 (65%)	13 (65%)	NS
Females	07 (35%)	07 (35%)	NS
Height (in meters)	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 ^0C)	36.9 ± 0.9	36.8 ± 0.7	t = 0.0767 p > 0.05 (NS)
Hemoglobin (in gm %)	11.8 ± 2.2	12.0 ± 2.1	t = 0.4691 p > 0.05 (NS)

[Table/Fig-1]: Data of baseline characteristics
NS: Not significant

Test	Cases	Controls	Significance
Heart rate variation during deep breathing (beats/min) (HRDB)	26.76 ± 8.11	26.7 ± 6.25	t = 0.041 p > 0.05 (NS)
Heart rate response to standing (30:15 ratio) (HRS)	1.22 ± 0.07	1.21 ± 0.05	t = 0.136 p > 0.05 (NS)
Blood pressure response to standing (mmHg) (BPS)	7.32 ± 2.28	7.84 ± 1.86	t = 1.246 p > 0.05 (NS)
Blood pressure response to sustained hand grip (mmHg) (BPSH)	22.16 ± 4.37	21.64 ± 4.43	t = 0.590 p > 0.05 (NS)

[Table/Fig-2]: Autonomic function tests and their significance
NS: Not significant



[Table/Fig-3]: Results of autonomic function tests
Note: HRDB in beats/min, BPS and BPSH in mm of Hg

Heart Rate Response to Standing (HRS)

The mean value of the 30:15 ratio which was used to assess the heart rate response to standing, in the cases was 1.22±0.07 and that in the controls was 1.21±0.05. The difference between the two groups was not statistically significant [Table/Fig-2 & 3].

TESTS FOR THE SYMPATHETIC SYSTEM

Blood Pressure Response to Standing (BPS)

The mean reduction in the systolic blood pressure (mmHg) in the cases was 7.32±2.28 and that in the controls was 7.84±1.86. The difference between the two groups was not statistically significant [Table/Fig-2 & 3].

Blood Pressure Response to a Sustained Hand Grip (BPSH)

The mean increase in the diastolic blood pressure (mmHg) in the cases was 22.16 ± 4.37 and that in the controls was 21.64 ± 4.43. The difference between the two groups was not statistically significant [Table/Fig-2 & 3].

The tests for the sympathetic and the parasympathetic systems did not reveal any abnormal response in both the cases and the controls.

DISCUSSION

After screening about 100 congenitally deaf children by ECG for the QT interval, we got about 20 congenitally deaf children with a long QT interval and these cases were selected for the study. The main aim of the study was to know the aetiology of the Long QT syndrome.

We knew that the aetiology could be K^+ ion channellopathy which predisposed to sensory neural hearing loss because of the malfunctioning of the stria vascularis and similar K^+ ion channels in the heart, thus causing the repolarisation defects.

But, the Long QT Syndrome or the Jervel-Lange Nielsen syndrome as it was previously called, is the one that shows an intriguing link between the autonomic nervous system and cardiac electrophysiology [4]. Srivasthava et al [5] suggested a sympathetic imbalance hypothesis as well as a repolarisation defect in the deaf children's hearts. An abnormal adrenergic neural control, along with ion channellopathies are responsible for the QT interval, the ST segment and the T wave changes which are found on the electrocardiogram [12].

Hence, we decided to investigate the aetiology of this syndrome – whether it was because of ion channellopathy or because of autonomic imbalance or whether both of these existed together in congenitally deaf children? So, we decided to study the autonomic function tests in these children, as studying the ion channels and their defects was beyond the scope of our set up.

In our study, none of the children exhibited defects in the autonomic function tests. The results of the tests were same in both the controls and in the congenitally deaf children.

Therefore, according to our study, the aetiology of the Long QT Syndrome is more in favour of ion channellopathy which predisposes to sensory neural hearing loss and cardiac repolarisation defects. The autonomic imbalance theory, as the aetiology of the Long QT syndrome can be stated as invalid from the present study.

A study on these K+ ion channels and their typing, as to which type of K+ channels and what type of defect-whether the opening or the closing of these ion channels is responsible for this syndrome, would lead to a deeper understanding of this disorder.

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