# STUDY OF CARDIOVASCULAR AUTONOMIC FUNCTIONS IN CONGENITALLY DEAF CHILDREN

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**ABSTRACT: BACKGROUND:** Majority of congenitally deaf children are at risk of cardiac abnormalities in the form of long QT syndrome which could be due to an intracardiac abnormality or autonomic dysfunction. Altered sympathetic/parasympathetic balance as a result of the absence of auditory stimuli on the autonomic nervous system results in lower mean heart rate in congenitally deaf children. **OBJECTIVES:** To evaluate whether the cardiovascular autonomic functions are altered in congenitally deaf children. **MATERIALS AND METHODS:** 30 congenitally deaf children aged between 14-18 yrs and 30 age matched controls were included for the study. Parasympathetic activity was assessed by observing the heart rate changes to immediate standing from lying down position, heart rate changes during deep breathing and heart rate changes on immediate standing from lying down position and blood pressure changes during sustained hand grip. **RESULTS:** The results of the present study showed statistically significant decrease in the systolic blood pressure in response to immediate standing among congenitally deaf children suggestive of sympathetic imbalance and an early stage of autonomic dysfunction.

**KEYWORDS:** Congenital deafness, Autonomic function tests.

**INTRODUCTION:** The long QT syndrome has aroused considerable interest in the medical community since its first description by Jervell-Lange in congenitally deaf children and later independently by Romano and Ward in persons with normal hearing. Both variants are characterized by prolongation of QT interval on surface ECG, T wave abnormalities and stress evoked syncope due to ventricular arrhythmias sometimes leading to fatal ventricular fibrillations.<sup>(1)</sup> The association of cardiac defects with congenital deafness is also called cardio auditory syndrome.<sup>(2)</sup>

Deafness by itself has a marked impact on the development of the child which when associated with cardiac defect may adversely affect child's development by limiting his/her physical activity. Children who are born deaf also become deaf mute, thus the disability remains unrecognized unless it becomes severe enough and reach an irreversible stage. Genetic factors account for at least half of the cases of profound congenital deafness. Till date, 80 loci for non syndromic hearing loss have been mapped and more than 30 genes have been identified. These genes belong to a wide variety of protein classes such as ion channel and gap junction components, transcription factors, extracellular matrix proteins, and genes with an unknown function.<sup>(3)</sup>

The ion channel defect which would result in congenital deafness could be in the form of a K+ion channel defect which predisposes to congenital sensory neural hearing loss and cardiac repolarisation defects.<sup>(4)</sup> Studies have suggested that cochlear and vestibular hair cells are the primary regulators of autonomic response,<sup>(5)</sup> and decreased sympathetic tone as a result of the absence of auditory stimuli on the autonomic nervous system is responsible for lower mean heart rate in deaf mute children.<sup>(6)</sup> Since auditory system is said to be involved in the regulation of

cardiovascular autonomic response the present study was taken up to evaluate whether the activity of autonomic nervous system is altered in congenitally deaf children.

**MATERIALS AND METHODS:** The study was done on 30 congenitally deaf children aged between 14-18 yrs. residing at Victory boarding school for hearing impaired, Kuppam. Clearance from ethical committee of PES Medical College, Kuppam and informed consent from the director of the boarding school was obtained for the study. Both male and female children suffering from sensory neural hearing loss since birth were enrolled in the study group. Deaf children with conductive deafness and children having any other cardiovascular disease were excluded from the study group. 30 age matched controls were selected from a nearby school of the same locality.

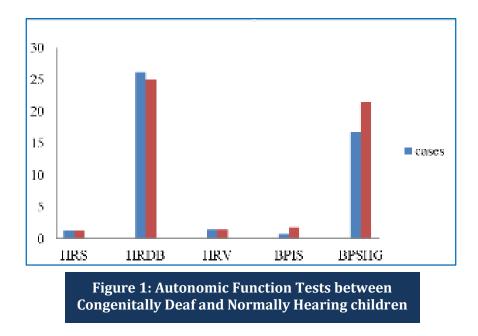
Five children were included for the study at each time. Height and weight was recorded and the BMI was calculated for each subject. After a brief general physical examination an ECG recording in lead II at rest was done, then the cardiovascular autonomic function tests were performed using Biopac student lab M P- 30 device. Due care was taken to remove the factors which could interfere with the results of the tests. Parasympathetic activity was assessed by observing the heart rate changes to immediate standing from lying down position, heart rate changes during deep breathing and heart rate changes during valsalva maneuver. Sympathetic activity was assessed by observing blood pressure changes on immediate standing from lying down position and blood pressure changes during sustained hand grip. Detailed instructions regarding the procedure which was employed for each test were given to the subjects through sign language by their class teacher for deaf children. The different maneuvers were demonstrated to the subjects and they were trained to perform the tests.

**RESULTS:** The results of the tests were expressed as means and differences between two groups. Statistical analysis was done by applying the unpaired student "t" test. P values <0.05 were considered to be statistically significant.

The mean values of the 30:15 ratio (the longest R-R occurring about 30 beats after standing divided by shortest R-R interval which occurs at about 15 beats after standing) which was used to assess the heart rate response to standing in the deaf were  $1.35\pm0.107$  and that in the normally hearing subjects were  $1.33\pm0.170$ , mean heart rate variation (the difference between the maximum and the minimum heart rates) during deep breathing in deaf were  $26.2\pm6.46$  and that in the normally hearing subjects were  $25.03\pm6.25$  and the mean values of valsalva ratio (ratio of longest R-R interval during phase IV to the shortest R-R interval during phase II) to assess the heart rate response to valsalva maneuver in the deaf were  $1.44\pm0.21$  and that in normally hearing were  $1.46\pm0.23$ . These findings suggests that the parasympathetic activity was not altered.

The mean increase in the diastolic blood pressure during sustained hand grip in deaf were  $16.76\pm7.56$  and in normally hearing subjects were  $21.35\pm10.71$  which is not statistically significant, whereas the mean reduction in the systolic blood pressure (mm Hg) in response to the immediate standing in deaf were  $0.73\pm1.52$  and that in normally hearing subjects were  $1.85\pm2.10$  which is statistically significant suggesting altered sympathetic activity.

Tests	Cases	Controls	P value
Heart rate response to standing (HRS)	$1.35 \pm 0.107$	$1.33 \pm 0.170$	0.58
Heart rate variation during deep breathing (HRDB)	26.2±6.46	25.03±6.25	0.58
Heart rate response to valsalva manuvre (HRV)	1.44±0.21	1.46±0.23	0.97
Blood pressure response to standing (BPIS)	0.73±1.52	$1.85 \pm 2.10$	0.02
Blood pressure response to sustained			
hand grip (BPSHG)	16.76 ± 7.56	$21.35 \pm 10.71$	0.09
Table1: Comparison of Autonomic Function Tests Between Congenitally Deaf and Normally Hearing Children			



**DISCUSSION:** Both deafness and cardiac arrhythmias occur as congenital problems either separately or together. Based on the study of two families discovered from an electrocardiographic survey of deaf children in Michigan, it is suggested that congenital deafness and cardiac arrhythmias may be closely related in inheritance but may occur separately within the siblings of a single family.<sup>(2)</sup>

The original description of severe congenital bilateral nerve deafness, associated with prolonged QT interval, syncope, and sudden death, has been followed by several reports.<sup>(7)</sup> Connexins and potassium channel proteins control key physiological processes in the heart and inner ear, demonstrating that similar electrically excitable tissues are present in both localizations,<sup>(8)</sup> which are considered to be the regulators of autonomic response.

The main finding of the present study was a statistically significant decrease in the systolic blood pressure in response to immediate standing suggestive of sympathetic imbalance. The findings of the present study is similar to Srivasthava et al which suggested a sympathetic imbalance hypothesis as well as a repolarisation defect in the deaf children's hearts.<sup>(1)</sup> Since sympathetic imbalance could lead to an early stage of autonomic dysfunction which in more severe form could pass into Jervel-Lange Neilsen variant of long QT syndrome and abnormal adrenergic neural control

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along with ion channelopathies are responsible for QT interval, ST segment and T wave changes found on ECG,<sup>(6)</sup> a battery of tests to detect autonomic dysfunction along with cardiovascular autonomic function tests and more detailed study of ion channelopathies predisposing to sensory neural hearing loss and cardiac repolarization defects can be performed which will lead to the deeper understanding of the disorder.

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