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ELECTROCARDIOGRAPHIC CHANGES FOLLOWING EXERCISE IN THE CONGENITALLY DEAF SCHOOL CHILDREN : RELATIONSHIP WITH JERVELL LANGE NEILSEN SYNDROME (THE LONG QT SYNDROME)

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Abstract : The present study was conducted to test the effects of exercise stress on the ECG of the congenitally deaf children from school for deaf, in view of the occurrence of the Jervell-Lange Neilsen (Surdo Cardiac) variant of the Long QT Syndrome (LQTS) in them. An ECG Lead II was recorded at rest and after two minutes of static jogging. For comparison, the same protocol was repeated in normal healthy children from another school. ECG were analysed for the calculation of corrected QT interval (QTc) by Bazett's equation $QTc = QT/\sqrt{R\text{–}R}$ and also for the evidence for other abnormalities. Both in the normal and deaf children, exercise did not produce significant ($P>0.05$) change in QTc from their resting values. However, when pre and post exercise QTc values of deaf children were compared with normal children, the female deaf had significantly longer QTc ($P<0.01$) both at rest and after exercise than normal female children.

Normal children did not show significant ECG abnormality either at rest or on exercise. On the contrary many of their counter part (deaf) exhibited 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.

These results lend credence to the hypothesis of sympathetic imbalance and repolarisation defects in deaf children's heart, which in more severe form could pass into frank Jervell - Lange Neilsen variant of the Long : QT Syndrome.

Key words : Long QT Syndrome (LQTS) ECG surdocardiac syndrome Jervell-Lange Neilsen syndrome congenital deafness

INTRODUCTION

The Long QT Syndrome has aroused considerable interest in the Medical Community of the west since its first description by Jervell-Lange Neilsen (1) in

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1957 in congenitally deaf children and later independently by Romano and colleagues (2) in 1963 and Ward (3) in 1964 in persons with normal hearing. Both variants are characterised by prolongation of QT interval on surface ECG> 440 msec, myriads of T wave abnormalities and stress evoked syncope due to ventricular arrhythmias sometimes culminating fatally (ventricular fibrillation). The spells of syncope are precipitated by those stresses which tend to augment sympathetic efferent traffic to the heart viz., fright, anger, quarrel, arousal by loud noise, exercise (especially swimming), menstruation and pregnancy etc.

These twin syndromes, therefore, form a unique link between electro-physiology, autonomic nervous system, cardiac arrhyhmias and noncoronary sudden death.

Although large body of work has been done in the west on both variants including their management to best of the author's knowledge, there are only two LQTS related studies (4, 5) from our country and no report on effects of exercise on ECG of congenitally deaf children, who may occasionally be victims of more sinister form of LQTS (Jervell-Lange Neilsen). These facts gave the impetus to undertake this study and also report any positive LQTS case if encountered to the international registry (6, 7).

METHODS

The present study was conducted in normal and deaf children in their respective schools. After obtaining consent from the Principals of the schools, a questionnaire was filled for deaf children with the assistance of the caretakers of the school for eliciting following informations :-

a) Exclusion of any intake of tricyclic antidepressants, antiarrhytmic agents or phenothiazene drugs.

b) History of syncope and its precipitating factors in the children and/or in their family members.

c) Incidence of congenital deafness and consanguinity among family members.

Any organic heart disease was ruled out by careful auscultation of the heart.

A resting ECG in Lead II (20 complexes) was recorded in deaf children after duly assuring them the noninvasive nature of the procedure and by demonstrating on a volunteer to allay any apprehension. While leads were still tied in place the children were asked to do static jogging for two minutes and a post exercise ECG was recorded as quickly as possible. Only ten children were screened per visit. The same protocol was repeated for the normal children.

All the children were in age group of 4-17 years; and were divided as normal male (n=15), and normal female (n=10) children and deaf male (n=50) and deaf female (n=27) children. Since pilot observations in normal children did not reveal appreciable ECG abnormality on exercise, the normal children group comprised of comparatively smaller numbers.

Analysis of the data : Electrocardiograms were analysed for the calculation of the pre and post exercise corrected QT interval (QTc) by applying Bazett's Formula (8) i.e. $QTc=QT/\sqrt{R-R}$ for normalisation of

frequency dependent variation of the QT interval. In each group, pre and post exercise mean QTc and its SE was calculated and inter group comparisons were made by Student's 't' test. The statistical significance was assigned at $P < 0.05$.

RESULTS

The results are summarised in the following histograms (Fig.1, 2) and (Table I). Ten congenitally deaf children had their brothers and/or sisters also deaf (14.3%).

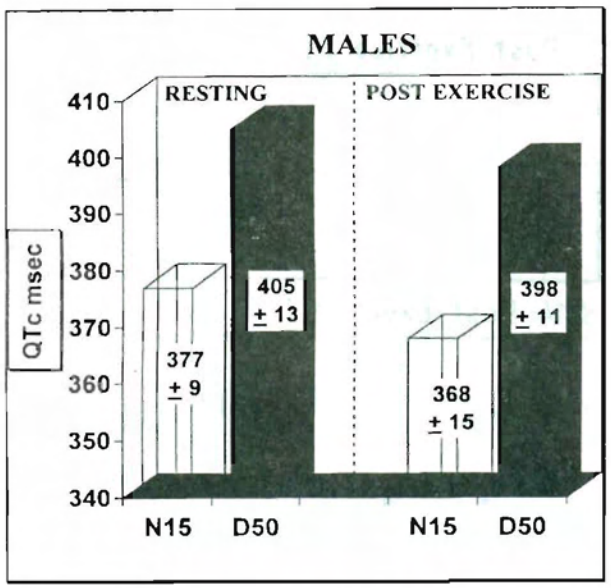

Fig.1: Showing mean QTc and its SE in normal (N) and deaf (D) male children at rest and after exercise.
N–Vesus – N – P > 0.05
D–Vrsus – D – P > 0.05

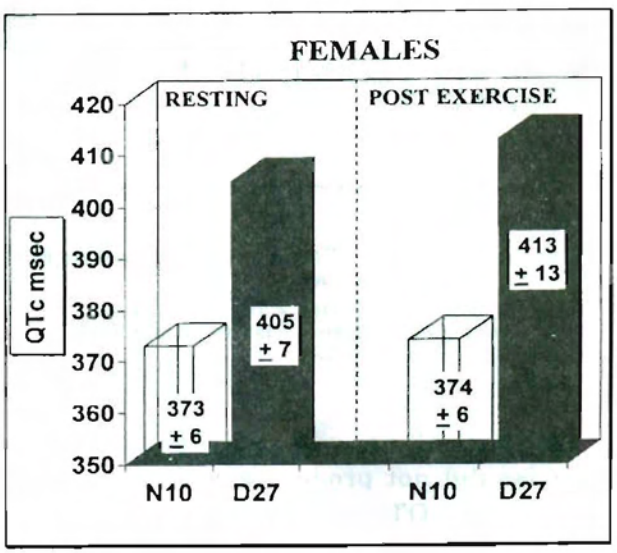

Fig. 2: Showing mean QTc and its SE in normal (N) and deaf (D) female children.
N – Versus – N – P > 0.05,
D – Versus – D – P > 0.05
Resting N – Versus – D – P < 0.01
Post exercise N – Versus – D – P < 0.01

TABLE I : Showing incidence and percentage occurrence of ECG abnormalities in deaf children following exercise.

ECG abnormalities	*Congenitally deaf*			
	Males		*Females*	
	No.	*%*	*No.*	*%*
Sinus arrhythmias	2	4	1	3.7
Sinus pauses	1	2	1	3.7
ST depression	2	4	2	7.4
T - inversion	1	2	2	7.4
Biphasic - T	3	6	–	–
Notched T	1	2	1	3.7
T Alternans	1	2	–	–
Nodal extrasystole	1	2	1	3.7
Junctional rhythm	2	4	–	–

Note :- Single child could exhibit more than one abnormality (Fig. 3).

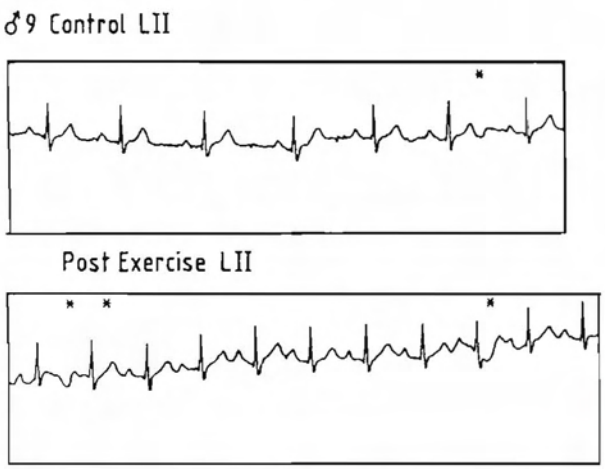

Fig. 3: Showing control (Resting) and post exercise ECG in L II in a deaf child.
Upper panel asterisk = Biphasic T wave
Lower panel asterisk – Ist and 2nd = T alternans
3rd = ST depression.

In normal as well as in deaf children exercise did not produce significant change ($P>0.05$) in QTc. However, when pre and post exercise QTc of deaf children was compared with normal children, the female deaf children showed a significantly longer QTc values ($P<0.01$) both at rest and after exercise as compared to their normal female counterparts.

Normal children did not show significant ECG abnormality either at rest or after exercise. On the contrary, many deaf children of both sexes exhibited occasional ECG abnormality at rest, but plethora of abnormalities on exercise viz., sinus arrhythmias, sinus pauses (Fig. 4), ST depression (Fig. 3), T-inversion, biphasic T (Figs. 3, 4), T-alternans (Fig. 3), notched T, nodal extrasystoles (Fig. 5) and junctional rhythm. Three girls had history of stress syncopes, however, their QTc was not prolonged.

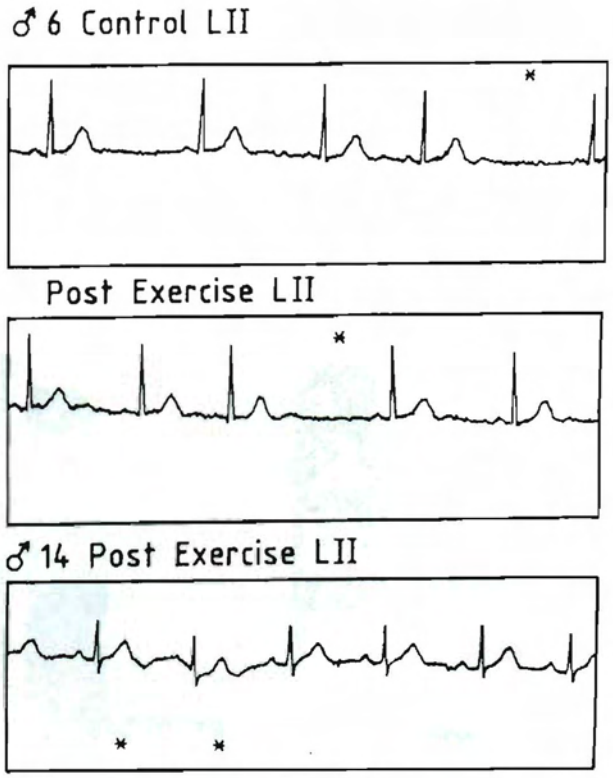

Fig. 4: Showing control (Resting) and post exercise ECG L II in deaf child 1 (upper and middle panels) and deaf child 2 (Lower panel).
Upper and middle panel asterisks = sinus pauses 1.24 and 1.2 msec respectively.
Lower panel asterisk – Ist = Biphasic T, 2nd = ST depression.
Many surdocardiac causes show sinus pauses > 1.2 seconds.

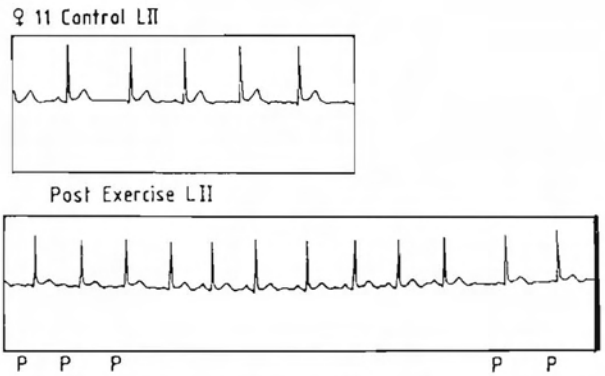

Fig. 5: Showing control (Resting) and post exercise ECG L II, in a deaf. In lower panel letters P = inverted P waves suggesting nodal extrasystoles.

DISCUSSION

For the quest of the surdocardiac syndrome surveys have been conducted in the schools for deaf in Ireland and England (9), USA (10), Spain (11) and Italy (12). Although, subject population in the present study is smaller, it is the only study from India to draw comparisons.

A longer QT interval in female deafs both on rest and exercise could in part account for the 69% occurrence of LQTS in female gender and also making it one of the most important determinant of cardiac events and sudden death (13). The incidence of ECG abnormality at rest was 2.1% in the surveys done by Cascos et al. (11) and by James (10). However, these investigators had not studied the effect of exercise on these children. In our study, the incidence of ECG abnormality was 6.5%. This variation could be due to smaller number in the present investigation. Following exercise the ECG abnormalities were present in 18% children. Non-availability of data on exercise evoked ECG changes in congenital deafness *per se*, makes comparison of our results rather difficult, nevertheless, Cascos et al. (11) observed, as we did that of 511 deaf screened by them, 12 exhibited certain features of the surdocardiac syndrome. Therefore, we shall compare the exercise evoked changes in congenital deafness *per se*, with studies reporting exercise or emotional stress evoked changes in well established surdocardiac cases. The ST segment depressions and sinus pauses are comparable to other studies on surdocardiac cases (14, 10, 13).

The constellation of T wave abnormalities witnessed in the present study is comparable to other reports on exercise or fright viz., notched T (9, 14, 15), T inversion (9, 11) biphasic T (9, 11, 15, 17) T alternans (9). Nodal ectopics and junctional rhythm observed in our study are again in conformity with earlier studies (10, 18, 19) and lend support to the opinion that atrial arrhythmias and conduction defects are not uncommon in surdocardiac syndrome (10, 18, 19).

The congenital deafness *per se* appeared to be of genetic nature as 10 subjects of our study (14.3) had their real brothers and/or sisters also deaf. Fraser et al. (9) has suggested an autosomal recessive inheritance for the congenital deafness. In the present study ECG abnormality both at rest and on exercise was more when the siblings were also deaf and often they manifested in both siblings.

Alongwith other surveys in the western countries, the results of present study warrant a national survey of all the schools for the deaf to identify children with stress evoked syncope and investigate them for positive surdocardiac cases, their management and enrollment in the international registry (20).

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