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Research Article

Hearing Loss - An Under Diagnosed Condition in Individuals with Hypertrophic Cardiomyopathy (HCM)

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Abstract

Hypertrophic Cardiomyopathy (HCM) is a condition characterized by the thickening of the heart muscle, potentially causing symptoms like fatigue, chest pain, and fainting. However, its relationship with hearing loss remains underexplored. The study titled "Hearing Loss - An Under diagnosed Condition in Individuals with Hypertrophic Cardiomyopathy (HCM)" investigates this connection by conducting detailed audiological assessments. The results revealed a significant increase in pure tone averages in HCM patients ($p < 0.05$), indicating higher levels of hearing loss. Moreover, speech recognition thresholds and discrimination scores were notably poorer in the HCM group. Tympanometry showed normal middle ear function, notably, otoacoustic emissions were absent, and the signal-to-noise ratios were much lower than those in healthy controls. These findings suggest that mild hearing loss, affecting the cochlea and brainstem, is common in individuals with HCM. The study highlights the need for further research to explore the mechanisms and clinical implications of this association.

Keywords: Hypertrophic Cardiomyopathy; Hearing Loss; Audiological Assessments; Tympanometry; Oto Acoustic Emissions

Abbreviations

HCM: Hypertrophic Cardiomyopathy

Introduction

Hypertrophic Cardiomyopathy (HCM) is a condition where the heart muscle thickens without a clear cause. Symptoms may include fatigue, shortness of breath, chest pain, fainting, and palpitations, which can worsen with dehydration. It can lead to reduced blood flow, causing exertional chest pain, dizziness, weakness, and even sudden cardiac death. Other complications may include heart failure, irregular heartbeats, and a lack of blood supply to the eyes, skin, and auditory system. The condition may develop gradually, progressively, or suddenly.

HCM affects 1 in 200 to 1 in 500 people globally, causing significant morbidity and mortality across all ages and genders. It is found in over 125 countries with diverse clinical presentations. Despite its genetic and clinical variation, HCM is now highly treatable, offering low morbidity, mortality, and the potential for normal life expectancy with personalized care.

In HCM, reduced blood supply to the cochlea can lead to cochlear degeneration and hearing impairment. El-Zarea., *et al.* [3] found that all patients with HCM experienced mild to severe bilateral sloping sensorineural hearing loss and study highlighted a significant link between hearing loss and the duration of the disease, emphasizing the importance of early detection and management of hearing impairments in HCM patients. Toth., *et al.* [2]. compared hearing functions in patients with HCM, DCM, and healthy controls. The study found that HCM patients had more frequent cochlear lesions than those with DCM, suggesting that abnormal myosin in the inner ear's muscular structures may contribute to hearing disorders in HCM.

Hidden hearing defects are found in about 50% of patients with HCM, significantly more often than in age- and clinical stage-matched patients with dilated cardiomyopathy (DCM) or those with valvular aortic stenosis. These defects are rarely seen in patients taking β-receptor blockers for conditions like hypertension or ischemic heart disease.

Materials and Methods

This prospective case-control study was conducted at the Department of Cardiology and ENT Hospital in Dehradun, Uttarakhand, India. It involved two groups: HCM patients (study group-HCM group comprised of 40 ears of 20 subjects (11 Male and 09 Female) in the age range of 30 to 50 years and a control group of healthy individuals. HCM patients were referred from the Cardiology Department for detailed audiological testing, and informed consent was obtained from all participants.

This study aimed to assess the audiological functions in patients with Hypertrophic Cardiomyopathy (HCM) through a multi-step evaluation process. The procedure involved three key components: cardiac evaluation, ENT examination, and audiological assessment.

Cardiac evaluation included personal and family history, physical examination, 12-lead ECG, and echocardiography to assess heart function. Electrocardiograms were performed using the BPL Smart Keep Cardiart 9108 ECG machine, while echo cardiographic imaging was conducted using the GE Vivid E9 Ultrasound Machine to evaluate the left ventricle's systolic function. The ENT examination focused on detecting abnormalities in the outer and middle ear.

Audiological testing included Pure Tone Audiometry, Speech Audiometry (Speech Recognition Threshold and Speech Discrimination Scores), Tympanometry, and Otoacoustic Emissions (TEOAE).

Pure tone audiometry was conducted at the conventional and extended high frequencies at octave frequencies (0.25 kHz to 8Hz) in all the enrolled participants in the study. The Hughson-Westlake Method (33) was used for hearing testing to measure the air conduction and bone conduction thresholds. The air conduction testing was evaluated in the frequency range of 250 Hz to 8 KHz while the bone conduction in the frequency ranges of 250 Hz to 4 KHz respectively. Three pure tone averages (PTAs) at three frequencies were computed, i.e., PTA for 500Hz, 1000Hz and 2000Hz frequencies for comparison between groups.

AC40 Inter acoustics clinical audiometer with TDH 39, HDA 300 headphones was used for conventional frequency audiometry respectively in a sound-treated room. Radio-ear B-71 bone vibrator was used to obtain the bone conduction threshold measurements.

Hearing was assessed using speech stimuli in both ears separately. Speech Recognition Threshold (SRT) and Speech Discrimination Scores (SDS) were obtained using Spondees and PB words respectively. The participants were instructed to repeat the spondee words. AC40 Interacoustics clinical audiometer was used to present the standardized spondee word list in Hindi, and the scores were calculated. The intensity level having 50% correct repetition of words was recorded as SRT. The Phonetically Balanced (PB) word list was presented to participants and administered through live monitored voice at 40 dB above the subject 's SRT. Out of 20 words, number of correctly repeated words was multiplied by 5 to get the percentage of speech discrimination scores.

Maico MI 34 middle ear analyzer was used to check the status of the middle ear. Tympanometric test was carried out using a probe tone frequency of 226 Hz at 85 dBSPL by varying pressure from +200 daPa to -400 daPa. Subjects having Type B tympanogram were excluded from the study. Tympanometry was performed to evaluate the ear canal volume, tympano-ossicular chain compliance value, and middle-ear pressure. According to Jerger classification the tympanograms were classified as type A, As, Ad, B, and C, in which A suggests a normal tympanogram. Tympanometry findings were considered as normal if the maximum middle ear pressure was between +/-100 daPa and the compliance was between 0.3 and 1.5 ml.

Transient - evoked otoacoustic emission (TEOAEs) were measured using Smart OAE version 1.0 of Intelligent Hearing Systems, USA. TEOAE: It was obtained using clicks stimuli at 85dBpeSPL at a rate of 19.3/s. Total sweeps of 1024 were with duration of 100 microseconds for a time window of 20 ms in both ears were tested separately, and signal to noise ratio (SNR) was measured. Signal to Noise ratio (SNR) values were recorded at 1000Hz, 1500Hz, 2000Hz, 3000Hz, and 4000Hz. TEOAEs were considered as present when the signal to noise ratio in three consecutive frequencies was above 3 dB.

All tests were carried out in a controlled environment, adhering to ANSI/ISO standards. The results from this comprehensive evaluation aimed to provide a detailed understanding of the cardiac and auditory impacts in patients with HCM.

Results and Discussion

The test results were analyzed using SPSS Statistics Version 20. The Shapiro-Wilk test confirmed normal distribution, and parametric independent t-test and non-parametric Mann-Whitney U test were used for data analysis. All the participants underwent detailed audiological test battery for assessing and profiling the auditory deficits. All the findings of the study are discussed below for different parameters.

Demographic profile

Table 1: Mean and standard deviation of age of HCM and control groups.

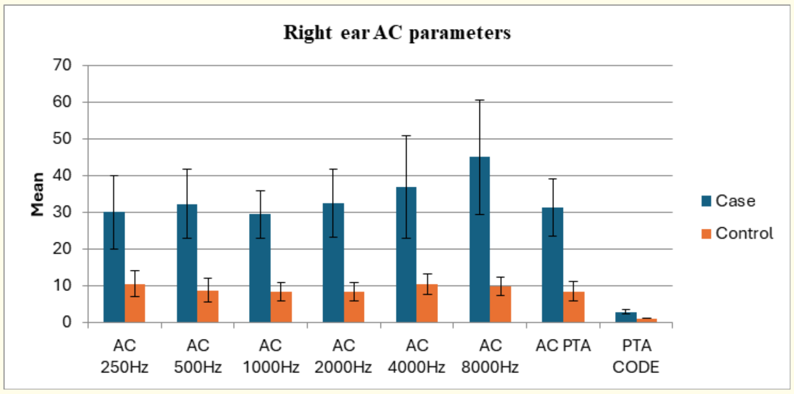
Groups	Number	Mean age (Years)	SD(±)
Study Group	20 (40 ears)	41.95	7.12
Control Group	20 (40 ears)	39.85	6.71

As shown in table 1, a total of 40 subjects were included in the study. Study group consisted of 20 subjects with Hypertrophic Cardiomyopathy (HCM)of (11 Males and 09 Females) with a mean age of 41.95(±7.12) years. The control group comprised of 20 Adults (12 Males and 08 Females) with a mean age of 39.85 (±6.71) years.

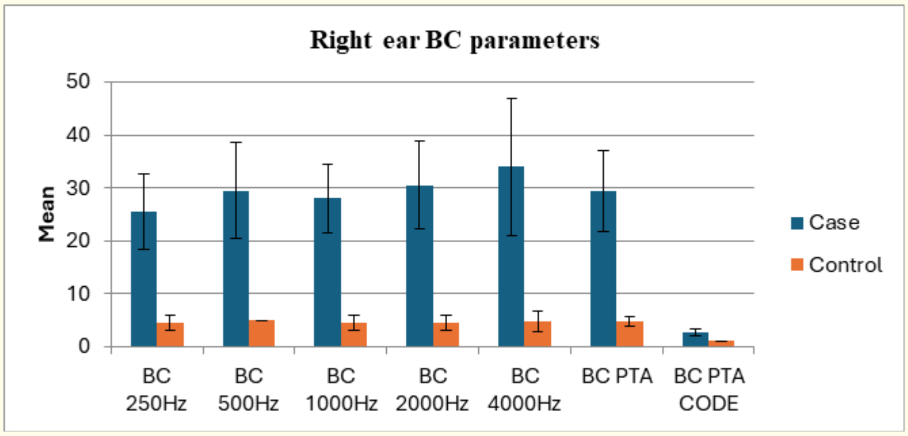
To assess the auditory function all the groups underwent an audiological test battery which included pure tone audiometry, speech audiometry, tympanometry, and OtoAcoustic Emission (OAE).

Pure tone audiometry

Pure tone audiometry was performed for frequency of 250 to 8 kHz in both right and left ears. Measures obtained and analyzed included the hearing thresholds at octave frequencies between 250-8kHz, in both ears.



Graph 1: Shows PTA thresholds (Air conduction) of right ears in study group (HCM) and control group.



Graph 2: Shows PTA thresholds (Bone conduction) of left ears in study group (HCM) and control group.

Graph 1 and 2 shows that normal hearing was observed in the control group across all the frequencies in right and left ears. In study group (HCM), mild hearing loss was present in the frequency range of 250-4 kHz and moderate hearing loss in the frequency of 8 kHz. A statistically significant difference ($p<0.05$) was observed between study group (HCM) and control group at frequencies 250Hz,500Hz, 1KHz, 2KHz, 4KHz, and 8KHz. The study group (HCM) has significantly higher air conduction thresholds on these frequencies as compared to the control group.

The patients with HCM were affected at all the frequencies in comparison to the control group. Thus, the finding in the present study suggested that people with HCM had hearing loss, as revealed in conventional audiometry (pure tone audiometry). These findings are indicative of abnormalities in the speech frequencies due to damage to the hair cells. Also the hearing deficit was more in HCM patients as compared to control subjects. Mild sensorineural hearing loss was observed across frequency range in HCM patients may be attributed to inner ear abnormalities.

The inner ear involvement may be attributed to an Autosomal dominant, myogenic abnormalities, degeneration in the stria vascularis, which affects the physical and chemical processes in the organ of corti, hence causing hearing impairment. Lack of blood supply in the cochlea, ventricular dysfunction produced progressive congestive heart failure and congenital deafness, which has been described in association with lentigines alone or with lentigines as part of the Leopard syndrome1, with mitral insufficiency, with associated skeletal abnormalities'201 and with pulmonary stenosis by M. Csanday., *et al*. [1]. This may lead to sensorineural hearing loss in HCM by Jost Schonberger., *et al*. [4] and Gehan Abd El-Rahman El-Zarea., *et al*. [3].

Similar findings in a study by Gehan Abd El-Rahman El-Zarea., *et al*. [3]. were that the affected adults had SNHL which affected frequencies to varying degrees with a variety of audiometric configurations, but the two youngest subjects had SNHL that affected only the low and middle frequencies. In a study by William R. Forney., *et al*. [6] pure tone audiometric test findings suggested that conductive hearing loss was observed in all 3 cases due to the malformation of the stapes, and osseous abnormalities consisting of fusion of cervical vertebrae and carpal bones in patients of short stature is reported. This was significant feature in this syndrome and rarely reported.

Pure tone averages were calculated in both the groups for right and left ear separately are shown in Table 2.

Frequency in Hz	Ear	Study Group (HCM)		Control Group		t-value	(p value)
		Mean (dB)	SD(±)	Mean (dB)	SD(±)		
PTA (500Hz, 1000Hz, 2000Hz)	Right	31.32	7.86	8.41	2.61	0.00	<.001
	Left	30.74	7.84	10.25	3.02	0.00	<.001
PTA Bone Conduction (500Hz, 1000Hz, 2000Hz)	Right	29.41	7.59	4.66	0.87	0.00	<.001
	Left	27.91	8.26	4.75	1.11	0.00	<.001

Table 2: Comparison of Pure Tone Averages (PTA) in both ears in Study and Control group.

Table 2 shows, PTA revealing a mild hearing loss on the comparison of air conduction average and bone conduction average in the study group (HCM). Normal hearing sensitivity was seen in the control group in both ears based on PTA. A statistically significant difference ($p<0.05$) was observed between study group (HCM) and control group at PTA. The study group (HCM) had significantly higher pure tone averages as compared to the control group.

Speech audiometry

Table 3 shows, statistically significant difference in both the ears in speech recognition thresholds as well as in speech discrimination scores among the two groups. Speech recognition and discrimination scores were fairly reduced in study group (HCM) as compared to controls.

As there was significant difference in pure tone average in the study groups compared to control groups for speech frequencies

Test	Ear	Study Group (HCM)		Control Group		t-value	(p value)
		Mean	SD(±)	Mean	SD(±)		
SRT	Right	34.25	8.15	11.50	2.35	0.00	<.001
	Left	34.00	7.88	1.00	.00	0.00	<.001
SDS (%)	Right	90.00	7.07	100.00	.00	40.00	<.001
	Left	90.50	6.26	100.00	.00	40.00	<.001

Table 3: Comparison of SRT and SDS between Study (HCM) and Control group.

in PTA (500 Hz, 1 kHz, 2 kHz), speech discrimination thresholds scores were also elevated in the study groups. The speech recognition scores in study and Control groups were between 90-100% which is similar to the findings obtained by Gehan Abd El-Rahman El-Zarea., *et al*. [3].

Impedance audiometry

Impedance audiometry was administered in study (HCM) and control groups. This test was performed to investigate the incidence of any middle ear pathology in both the ears separately. Measures included ear canal volume, static compliance, and middle ear pressure**.** Following results were observed.

Values	Ear	Study Group (HCM)		Control Group		t-value	(p value)
		Mean	SD(±)	Mean	SD(±)		
ECV	Right	1.11	0.31	1.34	0.51	153.00	0.20
	Left	1.19	0.25	2.00	1.02	-1.15	0.25
SC	Right	.52	0.20	.80	0.32	81.50	<.002
	Left	.61	0.25	1.33	0.48	-3.34	<.002
MEP	Right	-36.30	60.34	-13.83	16.09	159.0	0.26
	Left	-.25	53.66	.89	0.28	169.5	0.40

Table 4: Comparison of tympanometric measures in Study (HCM) and Control groups.

Type 'A' tympanogram was observed in both groups (study and controls). No statistically significant difference (<0.05) was observed in all Middle Ear Pressure (MEP) and Ear Canal Volume (ECV) values in both ears between the groups. It suggested that middle ear is functioning normally.

Type 'A' tympanogram was observed in both groups (study and controls). No statistically significant difference (<0.05) was observed in all Middle Ear Pressure (MEP) and Ear Canal Volume (ECV) values in both ears between the groups. It suggested that middle ear is functioning normally. In another similar study, type-A tympanograms were reported in Hypertrophic Cardiomyopathy (HCM) and normal healthy controls reflecting normal middle ear pressure by (Gehan Abd El-Rahman El-Zarea., *et al*. [3]). In the same study, they have found type C tympanogram in 3 cases due to Eustachian tube dysfunction due to common colds.

Oto acoustic emissions

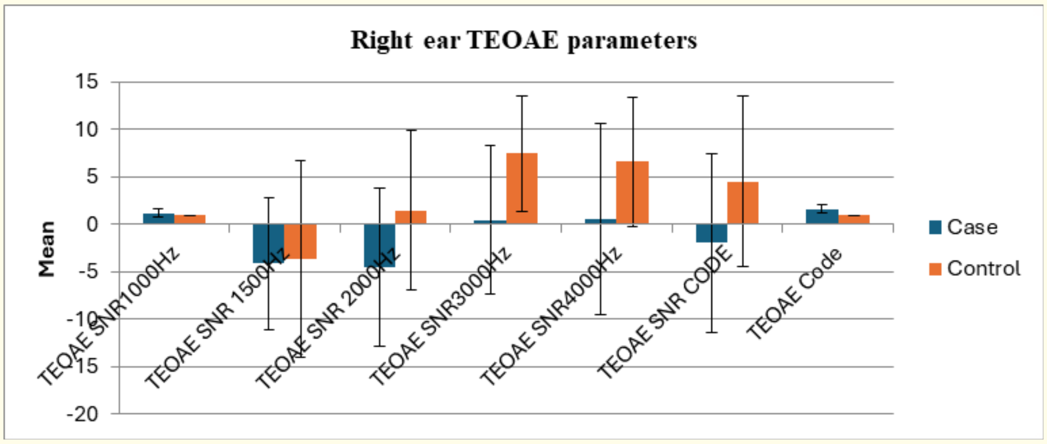
To compare the outer hair cell functioning in different groups, TEOAE tests, was administered on all the participants. Measures included SNR values at different frequencies in both the ears. Following findings were observed**.**

In this study, 35% (7 subjects) in the right ear and 40% (8 subjects) in left ear showed the presence of TEOAE's while 65% (13 subjects) in the right ear and 60% (12 subjects) in the left ear in the study (HCM) group showed the absence of TEOAEs. TEOAE were present in both the ears in the control group.

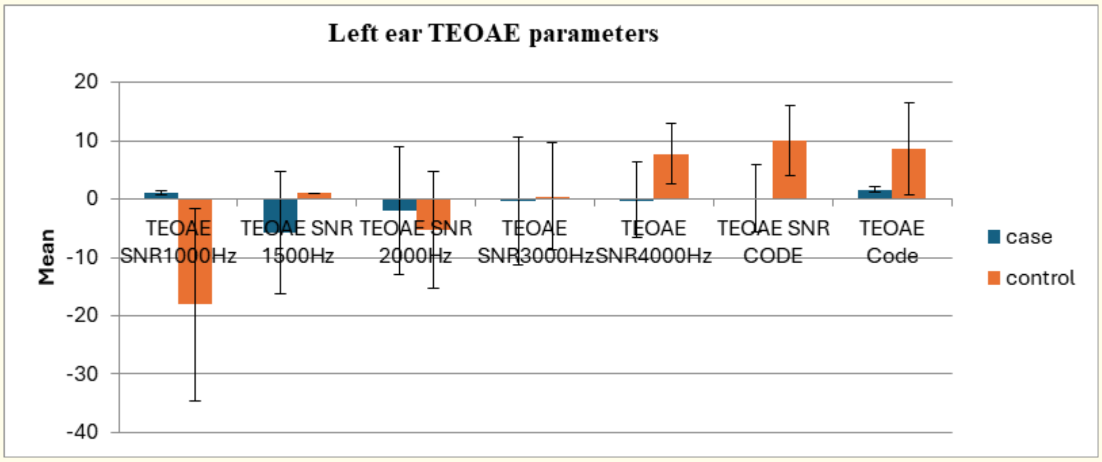
Significantly lower SNR (Graph 3 and 4) values were observed for TEOAE's at all frequencies in comparison to the control group except 1 kHz and 2 kHz in the left ear, and 3 kHz in both the ears.

Frequency in Hz	Ear	Study Group (HCM)		Control Group		t-value	(p value)
		Mean	SD(±)	Mean	SD(±)		
1000	Right	1.20	0.41	1.00	0.00	160.00	<0.03
	Left	1.10	0.30	-18.10	16.42	180.00	0.152
1500	Right	-4.14	6.97	-3.68	10.34	-0.16	0.87
	Left	-5.83	10.46	1.00	0.00	192.50	0.839
2000	Right	-4.51	8.35	1.48	8.40	-2.25	<.03
	Left	-2.00	10.96	-5.30	10.07	-0.77	0.44
3000	Right	0.47	7.88	7.46	6.07	-3.14	<0.003
	Left	-.37	10.99	.45	9.11	-2.97	<0.005
4000	Right	0.57	10.14	6.61	6.83	115.5-0	<0.022
	Left	-.05	6.45	7.72	5.18	-5.11	<.001

Table 5: Comparison of SNR on TEOAE in both ears in Study (HCM) and control groups.



Graph 3: Shows Comparison of SNR on TEOAE in right ears in Study (HCM) and control group's.



Graph 4: Shows Comparison of SNR on TEOAE in left ears in Study (HCM) and control group's.

In this study, 35% (7 subjects) in the right ear and 40% (8 subjects) in left ear showed the presence of TEOAE's while 65% (13 subjects) in the right ear and 60% (12 subjects) in the left ear in the study (HCM) group showed the absence of TEOAEs. TEOAE were present in both the ears in the control group.

Significantly lower SNR values were observed for TEOAE's at all frequencies in comparison to the control group except 1 kHz and 2kHz in the left ear, and 3 kHz in both the ears.

TEOAEs were present at all frequencies of SNR in both ears in comparison with study (HCM) and control groups. A significant difference was found in study and control groups for both the ears at 1000 Hz, and 2000Hz in right ears while left ears were insignificant. There was statistically significance observed on 3000 Hz and 4000 Hz in both the ears. At 1500 Hz, no statistically significant difference was found in both ears. The values were lower for study groups (HCM) in these frequencies. The reduction and lowering of SNR in Study group (HCM) may be due myogenic lesions reported by Miklós Csanády [2]. The reduction could be due to the fact that the basal region gets initially deteriorated in the early stage of various diseases. In a similar study, authors reported the lowering of TEOAE SNR values in study group (HCM) when compared to the control group suggested by Miklós Csanády [2]**.** In our study, SNR was significantly reduced as compared to the control group.

Conclusion

This study highlights a significant association between Hypertrophic Cardiomyopathy (HCM) and mild hearing loss, primarily affecting the cochlea and brainstem. The audiological evaluations, including pure tone audiometry, speech audiometry, tympanometry, and otoacoustic emissions, revealed notable differences in hearing function between HCM patients and healthy controls. Specifically, HCM patients demonstrated higher pure tone averages, reduced speech recognition thresholds, and absent otoacoustic emissions, suggesting an auditory impairment that may remain unrecognized in this group.

The results highlight the importance of heightened awareness and regular audiological evaluations in individuals with HCM to detect and manage hearing loss early. Further research is essential to explore the underlying mechanisms of this auditory dysfunction and its potential clinical significance in the context of HCM, paving the way for better patient care and management strategies.

There are multiple shortcomings in this study. First, the sample size was relatively small, which hampers the ability to generalize the results to the broader population. Additionally, certain audiological tests, such as Auditory Brainstem Response (ABR), Distortion Product Otoacoustic Emissions (DPOAE), Auditory Steady-State Response (ASSR), and tone burst ABR, were not conducted on the study participants, which could have provided a more detailed understanding of auditory function. Finally the study focused solely on individuals diagnosed with Hypertrophic Cardiomyopathy (HCM), emphasizing the need for future longitudinal follow-up studies to examine the long-term effects of hearing loss in this specific population.

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Conflict of Interest

Nil.

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