

Case Report

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Auditory processing disorders associated with a case of Kartagner's syndrome

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Summary

Kartagner's syndrome is a rare autosomal recessive disorder characterized by sinusitis, bronchiectasis and situs inversus. Otitis media is seen in 95% of the individuals with this syndrome due to recurrent respiratory infections and dysfunctional cilia in the middle ear. Earlier research reported the presence of structural and functional deficits in the auditory brainstem following long standing otitis media. However, no such findings have been reported in individuals with this syndrome. Thus, the present case report highlights the results of various audiological tests with special emphasis on investigating the auditory processing abilities in a known case of Kartagner's syndrome. In order to accomplish the aim, the audiological test battery was carried out on a 42 year old male patient diagnosed as having Kartagner's syndrome. The basic audiological tests, including immittance audiometry, pure tone audiometry, otoacoustic emission and auditory brainstem response (using click stimulus) results indicated the presence of mild to moderate mixed hearing loss in both ears. However, results of the auditory brainstem response (using speech stimulus) pointed toward abnormal speech processing skills. Thus, the behavioral test battery approach (including speech perception in noise test, gap detection test, temporal modulation transfer function test and duration pattern test) was followed and the findings suggested presence of auditory closure and temporal processing deficit. The outcome of the case study recommends that a complete test battery approach involving psychoacoustic tests should be used to assess such cases and auditory rehabilitation should be suggested accordingly.

Keywords: Kartagner's syndrome, primary ciliary dyskinesia, auditory processing, psychoacoustics

1. Introduction

Kartagner's syndrome, also known as Primary Ciliary Dyskinesia (PCD) is a rare congenital disorder with an occurrence of approximately 1 in 30,000 live births (1,2) in the Western population. Although the true prevalence of the disease is unknown, the global prevalence data it is found to be 5 in 100,000 live births (3). The statistically extrapolated data from the global prevalence indicate that approximately 60,000 individuals in India are suffering from the disorder. The disorder was first described by Siewart (4), although Kartagner (5) was the first to describe the characteristics of the disease. The incidence of the disease was found to be more in males in comparison to females (6).

Kartagner's syndrome is an autosomal recessive disorder (7) characterized by sinusitis, bronchiectasis and situs inversus (8). The main symptoms of the disease include chronic cough, chronic rhinitis, mucopurulent sputum, agenesis of the frontal sinus and other respiratory infections occurring during early childhood (9). Recurrent respiratory infections (10) and infertility (11) is commonly seen in adulthood. Otitis media is also commonly seen in this disorder due to recurrent respiratory infection and dysfunctional cilia in the middle ear (12).

Recurrent otitis media was reported in 95% of the individuals with PCD (13). Afzelius (9) also reported the presence of recurrent otitis media in these individuals. Long standing otitis media may result in structural and functional deficits in the auditory nervous system (14). However, no anatomical defects of the nervous system have been reported to be associated with Kartagner's syndrome. Yamashita *et al.* (15) reported the presence of amyotrophic lateral sclerosis

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in one of the patients suffering from Kartagener's syndrome. Olbrich *et al.* (16) also reported right-left asymmetry in such cases. In a review on the evaluation and management of PCD, Rossman and Newhouse (17) indicated that meningitis or brain abscess may occur as a consequence of sinusitis and bronchiectasis. The literature indicates that some form of anatomical and physiological deficits may be associated with Kartagener's syndrome, although, the investigation of auditory processing abilities has not been carried out in these individuals, as per the knowledge of the investigator. Thus, the present case report highlights the results of various audiological tests with special emphasis on investigating the auditory processing abilities in a known case of Kartagener's syndrome.

2. Case Report

A case aged 42 years/male reported to the department of audiology with a complaint of decreased hearing sensitivity in both ears. The case was referred from the department of otology and was diagnosed as having Kartagener's syndrome. Available reports of the otomicroscopic examination revealed subtotal tympanic membrane perforation and the presence of thick mucosa in the middle ear in both ears. Ear discharge was not present at the time of investigation. The case was diagnosed as having chronic suppurative otitis media with hereditary spherocytosis with Kartagener's syndrome. Available reports of Hematology showed the presence of spherocytes, polychromasia and anisocytosis in the smear. No other significant abnormality was noted and the features were consistent with hereditary spherocytosis. The x-ray investigation of the para nasal sinus revealed bilateral haziness of

both maxillary sinuses, hypoplastic left frontal sinus and deviated nasal septum to the left side (Figure 1A). The ultrasonography test report for the abdomen revealed situs invertus and the chest x-ray revealed dextrocardia (Figure 1B). The report of the chest x-ray also showed abnormal lung thickening and thickened airway walls. The available result confirms the presence of Kartagener's triad (sinusitis, bronchiectasis and situs inversus) and hence the diagnosis.

A written consent was obtained from the patient in his native language (Kannada) prior to the evaluations. Detailed case history information obtained from the case reported the complaint of hearing loss in both the ears for the last 25 years. The case was an auto driver by profession and reported more difficulty in hearing while driving. The loss was slowly progressive in nature. The case history also pointed towards the complaint of ear discharge since childhood, which was mild, serous and intermittent in both ears. The frequent attacks of upper respiratory tract infection were also noted in the case history. There was no other significant history of ear pain, tinnitus or vertigo.

Acoustic Immitance measurements were carried out using an Interacoustics MT10 Immitance meter to investigate the presence of middle ear pathology and the results indicated a bilateral "B" type tympanogram, because of the subtotal perforation in both tympanic membranes. Distortion product and transient evoked oto-acoustic emission (Oto-read, Interacoustics) were also absent (SNR < 6) within the frequency range of 700 Hz to 12 KHz. An audiometric measurement using a Maico MA53 dual channel diagnostic audiometer was carried out and the results indicated the presence of mild to moderate sloping mixed hearing loss (Table 1). Word recognition scores and speech identification

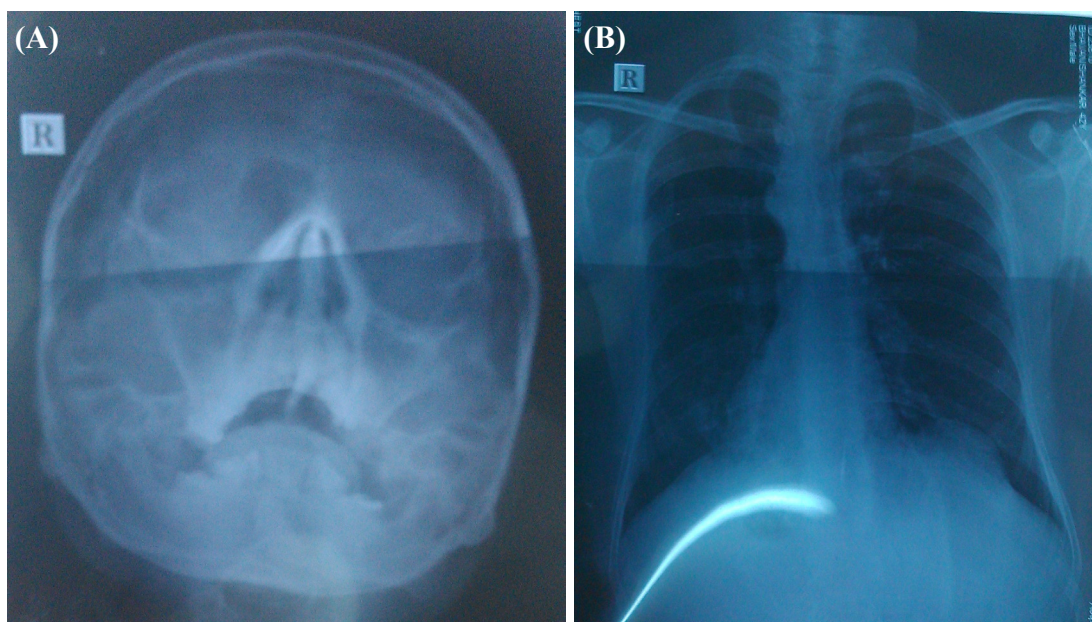


Figure 1. The X-Ray representation of (A) the para nasal sinuses; (B): the chest showing dextrocardia.

scores were consistent with the pure tone average.

The results were confirmed with the help of auditory brainstem responses (ABR). Click evoked ABR at the rate of 33.1/sec revealed presence of V peak at 55 dBnHL in right ear and 60d BnHL in left ear. Prolonged latencies of the III and V peak and delayed for I-III and I-V inter wave latency intervals were observed in both ears. The inter peak latency for the V peak was also found to be within normal range for different rate of stimulus presentation viz. 11.1/sec and 90.1/sec. The absolute latency and the inter wave latency difference of different ABR peaks at different rates of stimulus presentation is illustrated in Table 2.

The presence of ABR waveform indicated fair integrity of the auditory nerve and the lower auditory brainstem. In order to assess the integrity of the higher brainstem and the lower auditory cortex, middle latency response (MLR) and late latency responses (LLR) were measured using IHS Smart EP. The MLR was measured using click stimulus at the rate of 5.1/sec and the LLR was measured using click stimulus at the rate of 1.1/sec at a presentation level of 99 dBnHL. The results of MLR indicated a robust amplitude of the Na-Pa complex within normal absolute latency limits (Figure 2). The results of LLR also indicated the presence of the N1-P2 wave complex within normal absolute latency limits and good amplitude (Figure 3).

Vestibular evoked myogenic potentials (VEMP) were also administered using IHS Smart EP. Although VEMP is a neurophysiological technique to assess the functioning of the vestibular apparatus, the organs

which are responsible for maintaining balance and equilibrium of the body in space, and the patient did not complain of any balance related problem, it is assessed because of the closed synchrony of the vestibular system with the hearing system. The results of the VEMP test indicated a prominent P13-N23 wave complex in both ears at an intensity of 107 dBnHL (Figure 4).

Speech evoked ABR using standardized "da" stimulus at an intensity of 95 dBnHL and a rate of 3.1/sec was also administered in the audiological test battery. The results of the speech evoked ABR were interesting with the presence of a V-A complex in

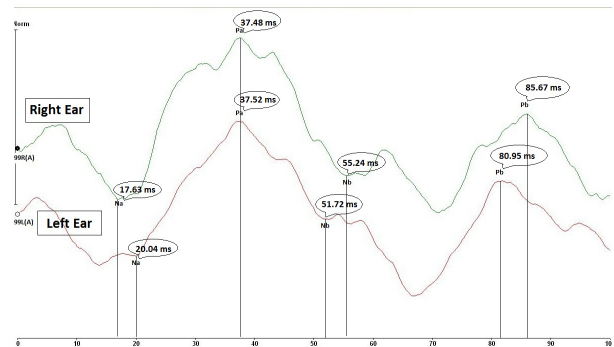


Figure 2. Middle Latency Responses. The waveform showing Na-Pa peak complex for the right and left ear.

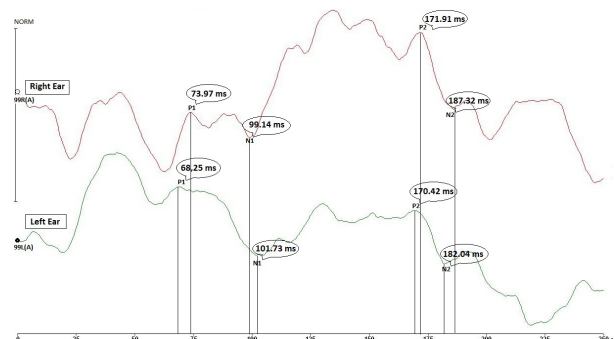


Figure 3. Late Latency Response. The waveform showing N1-P2 peak complex for the right and left ear.

Table 1. Audiometric findings for right and left ear for pure tone and speech

Procedure	Right Ear	Left Ear
PTA	33.3 dB	35 dB
SRT	35 dB	40 dB
SIS	90%	90%
MCL	85 dB	85 dB
UCL	>100 dB	>100 dB

Table 2. Latencies estimated from ABR test for both ears using various protocols

Parameters		Right Ear					Left Ear				
		Absolute Latency (in ms)			Inter-wave Latency Difference (in ms)		Absolute Latency (in ms)			Inter-wave Latency Difference (in ms)	
Intensity	Rate	I	III	V	III-V	I-V	I	III	V	III-V	I-V
90 dB	33.1/sec	1.37	3.75	5.89	2.14	4.52	1.44	3.78	6.09	2.31	4.65
80 dB	33.1/sec	1.56	3.94	6.52	2.58	4.96	1.69	3.86	6.38	2.52	4.69
70 dB	33.1/sec	NP	4.18	6.89	2.71	-	1.98	4.09	6.86	2.77	4.88
60 dB	33.1/sec	NP	NP	7.15	-	-	NP	NP	7.16	-	-
55 dB	33.1/sec	NP	NP	7.38	-	-	NP	NP	7.54	-	-
50 dB	33.1/sec	NP	NP	7.82	-	-	NP	NP	NP	-	-
45 dB	33.1/sec	NP	NP	NP	-	-	NP	NP	NP	-	-
90 dB	11.1/sec	1.29	3.62	5.77	2.15	4.48	1.39	3.56	5.98	2.42	4.59
90 dB	90.1/sec	NP	4.03	6.14	2.11	-	NP	NP	6.43	-	-

*NP: No identifiable peak observed.

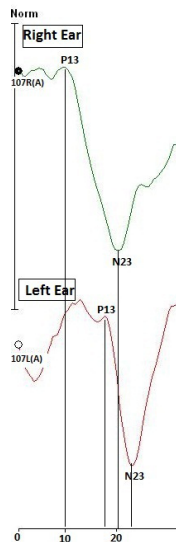


Figure 4. Vestibular Evoked Myogenic Potentials. The waveform is showing P13-N23 peak complex for the right and left ear.

both ears but absence of late peaks. These results were consistent across waveform replicability with the signal to noise ratio greater than '1' and the residual noise level less than $0.08 \mu\text{V}$. These results were inconsistent with the other neurophysiological tests, however, but consistent with the patient's complaint of inability to understand speech clearly. Abnormal speech evoked ABR waveform suggests abnormal speech processing and the finding with a normal click evoked ABR is suggestive of possible auditory processing deficit (18). Thus, a detailed investigation of processing abilities was warranted to confirm the findings.

In order to investigate the presence of auditory processing abilities, a test battery approach was followed and tests like speech perception in noise (SPIN), gap detection thresholds (GDT), temporal modulation transfer function (TMTF) using 8 Hz modulation, 60 Hz modulation and 200 Hz modulation rates, and a duration pattern test (DPT) was administered.

SPIN was measured using standardized Kannada paired words (19) and monosyllables using a Maico MA53 dual channel diagnostic audiometer. The stimulus was presented using monitored live voice and the intensity level was kept constant at 10 dB above the most comfortable level in both the ears and the intensity of the speech noise was varied in such a way that the SNR was +10 dB, +5 dB, 0 dB and -5 dB. The results indicated more than 50% scores for both paired words and monosyllables at the SNR level of +10 dB and +5 dB, however, less than 50% scores were obtained for the SNR of 0 dB and -5 dB. The results for the paired words were consistently better than the monosyllables. The poor scores on the SPIN are indicative of abnormal auditory closure ability.

GDT was measured using a maximum likelihood procedure (MLP, a MATLAB based toolbox for

psychoacoustic experiments) (20). The stimulus was presented using the personal computer routed *via* a Maico MA53 audiometer equipped with TDH 39 headphones. The intensity was monitored at the most comfortable level in both ears. The results indicated poor gap detection scores (9.13 ms) in comparison to normal (5 ms) (21) individuals. The poor scores on the GDT indicated abnormal temporal resolution abilities. The abnormal performance on the temporal resolution skills were also indicated by poor TMTF scores for 8 Hz, 60 Hz and 200 Hz. TMTF was also measured using MLP routed *via* audiometer with the stimulus presented at the most comfortable level in both ears. Results indicated -8 dB modulation detection thresholds at 8 Hz, -6 dB thresholds at 60 Hz and -2 dB thresholds at 200 Hz. These results were far poorer in comparison to normal individuals (-13 dB for 8 Hz, -9 dB for 60 Hz, and -4 Hz for 200 Hz) (21).

Finally, the DPT was also administered using MLP. A total of 30 stimuli using long and short stimulus in three tone patterns was presented. The patient's task was to indicate the correct sequence of stimulus presentation. Results of the test indicated very poor scores (16 correct pattern identification) in comparison to age matched normal individuals (28 correct pattern identification) (21). Poor scores on this test indicate abnormal temporal sequencing abilities.

Results of various auditory processing tests confirm the presence of abnormal auditory processing ability in the individual with Kartagner's syndrome.

3. Discussion

Kartagner's syndrome is a rare genetic disorder due to deficits in ciliary motion (15). Immobile ciliary action in Kartagner's syndrome is mainly manifested in the respiratory tract and the fallopian tube (22). As a result sinusitis and bronchiectasis are associated with the disorder. Defective ciliary rotation in these cases results in situs inversus (23,24). Recurrent otitis media are commonly seen in such individuals (25).

As Webster *et al.* (14) reported that long standing otitis media may result in structural and functional deficits in the brainstem and higher cortical structures, there is a need to investigate the presence of auditory processing disorders in individuals with Kartagner's syndrome. Moreover, significant hearing loss is also associated with recurrent otitis media (26). Thus, the present study aimed at investigating the audiological characteristics of a known case of Kartagner's syndrome with special emphasis on auditory processing disorders.

A test battery approach was used to assess the auditory functioning in the client. Acoustic Immitance measurement revealed a flat tympanogram with normal ear canal volume. Such findings are suggestive of a perforated tympanic membrane with active middle ear pathology (27). Acoustic reflex and otoacoustic

emissions were not obtained probably because of the perforated tympanic membrane (28,29). Audiometry reports indicated bilateral mild to moderate sloping mixed hearing loss with correlating speech scores. These findings were consistent with long standing otitis media (30).

ABR to click stimuli also indicated the presence of middle ear pathology in terms of prolonged absolute latencies and delayed inter wave intervals. Prolonged latencies of wave III and wave V was also reported by Folsom, Weber and Thompson (31) in individuals with recurrent otitis media. Gunnarson and Finitzo (32) and Hall and Grose (33) reported delayed interwave intervals for I-V and III-V wave associated with persistent recurrent otitis media. Thus, the findings for click evoked ABR was consistent with the literature findings. MLR and LLR were also administered and the findings pointed towards normal integrity of the auditory brainstem and lower auditory cortex. The presence of MLR and LLR peaks within normal absolute latency range in both ears (without delay caused due to the presence of middle ear pathology) may be due to the high level of stimulus presentation. Both MLR and LLR were administered at 99 dBnHL, which is approximately 60 dB above the pure tone average. With such a high intensity of stimulus presentation, attenuation caused due to the conductive component may be compensated.

The basic audiological findings suggested the presence of mixed hearing loss in the present individual. However, the results of speech evoked ABR were not consistent with the non speech findings. The results signify the presence of an auditory processing disorder and a detailed investigation was carried out using a set of psychoacoustic tests. The findings on the SPIN indicated the presence of auditory closure deficits, on GDT and TMTF pointed towards the presence of temporal resolution deficits and poor scores on the DPT showed existence of temporal sequencing deficits. These results confirmed the presence of an auditory processing disorder in the individual with Kartagener's syndrome, and thus, the detailed investigation should be carried out with each such individual and management considering the processing deficit should be provided.

4. Conclusion

Kartagener's syndrome is a congenital disorder characterized by the presence of sinusitis, bronchiectasis and situs invertus. Recurrent otitis media is common in these individuals and hence, it is likely that auditory processing disorders may be present in such individuals. The present study aimed at investigating audiological characteristics with special emphasis on auditory processing disorders in a known case of Kartagener's syndrome. A test battery approach was applied and results of basic audiological tests pointed towards the

presence of bilateral mild to moderate mixed hearing loss. In order to assess auditory processing ability, a set of psychoacoustic tests were administered and the results showed the presence of auditory closure and temporal processing deficits.

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