**Underprivileged hearing impaired cohort deprived from concession/benefit in hearing disability Act**

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**Abstract**

Four clients had visited to clinic seeking hearing disability certificate. All the clients reported as having reduced hearing sensitivity in both ears. Audiological evaluation and hearing aid trial were performed and hearing handicap checklist was administered on them. Based on findings of audiological evaluation, it was confirmed that two of them had bilateral symmetrical sensorineural hearing loss (SNHL) with moderate and moderately severe degree, respectively. Whereas other two clients were diagnosed as bilateral symmetrical auditory neuropathy spectrum disorder (ANSD) with mild and moderately severe degree, respectively. In addition, severity of hearing handicap was mild to moderate for SNHL clients and severe in ANSD clients. Further, hearing aid was found to be beneficial to those clients with SNHL. Unlike SNHL, ANSD clients are not benefitted with hearing aid. Irrespective of site of lesion the clients who had moderately severe hearing loss (≥ 60 dB HL) can avail concessions/ benefits for their issued disability certificate (RPWD, 2016). Though individual with ANSD had mild degree of hearing loss their speech perception was found severely impaired, hearing aid was not beneficial and severely affected in hearing handicap. In spite of these problems the client with ANSD is not an eligible candidate to avail any concessions/ benefits from hearing disability certificate if issued. The probable reasons for devoid of any concessions/ benefits in such clients is discussed in the present study.

**Key-Words:** Disability, hearing loss, certificate.

**Introduction**

Auditory Neuropathy Spectrum Disorder (ANSD) is a form of sensori-neural hearing loss (SNHL) in which functioning of outer hair cells is preserved but neural mechanism is disrupted. The diagnosis of ANSD with respect to audiological test results is based on 1. Presence of oto-acoustic emissions 2. Absence or severe abnormality of auditory brainstem responses. 3 Absence or marked elevation of acoustic middle ear reflexes and 4. Impaired speech perception. In case of cochlear pathology, the severity of hearing loss is positively correlated with speech recognition scores. To be specific as the degree of hearing loss increases their speech recognition score reduces (negative correlation). In contrast, individuals with ANSD show severe speech understanding problems irrespective of their degree of hearing loss (no agreement). In India, 63 million people (6.3%) suffers from significant hearing loss. Four in every 1000 children suffer from severe to profound hearing loss. The prevalence of ANSD varies from 0.53 to 2.27 (1) and is often noticed at late adolescence (2).

To evaluate the degree of handicap in individual with hearing impairment experiences in the real world, communication handicap checklists are necessary. Hearing loss affects everyday communication and it varies from person to person. The self- assessment data provide insight on individual’s response to hearing impairment, which cannot be gleaned from audiometric data alone. In SNHL, percentage of communication handicap depends on their pure-tone average on or speech identification scores (SIS) (3). If these individuals are rehabilitated with a suitable hearing aid, their speech perception may improve. The percentage of communication handicap in them reduces after fitting the hearing aid, due to which their quality of life improves (4). In ANSD, hearing aid benefit was accounted to 27 % and 5 % for those who had good and poor speech recognition scores, respectively (5). In addition, severe communication handicap remains or even becomes worse after fitted with hearing aid (6).

In India, disability certificate for individuals with hearing impairment is issued based on degree of hearing loss (RPWD Act, 2016). The present criteria to issue disability certificate are most suitable for individuals with cochlear pathology. In contrast, individuals with ANSD show severe speech understanding problems irrespective of their degree of hearing loss. Individuals with ANSD though have mild hearing loss, it was observed a severe handicap on communication and no benefit from hearing aid. The present study highlights on how RPWD (2016) preclude from availing benefits to a client with hearing impairment who have genuine speech perception problem with mild hearing loss and a severe hearing handicap.

**Case report**

**Clinical History**

A total of four clients (AB AC AD & AE) arrived with a compliant of reduced hearing sensitivity in both ears and their age range from 10 to 12 years (mean age = 11.5 years). To be specific, clients AB and AD reported difficult to listen speech at a soft level. Clients AC and AE reported normal hearing till 10.5 years and encountered a sudden hearing loss in both ears. They claim ‘can hears a speech but unable to understand it’. They even reported difficult to follow speech in noise. None of the clients had ear infection and dizziness when they reported to the clinic. A detailed audiological testing, hearing aid trial was performed and hearing handicap questionnaire was administered on all the clients.

**Audiological Testing**

After case history and ontological examination, a detailed audiological evaluation was completed. Conventional pure-tone audiometry revealed a bilateral moderate hearing loss in client AB and mild hearing loss in client AC and moderately severe hearing loss in AD and AE clients. Speech audiometry was performed in each ear for all the clients. In client AB, the speech recognition threshold (SRT) was 55 and 88 % in speech identification score (SIS) on the right ear. Whereas on left ear SRT was 60 dB and SIS was 88 %. The SRT for client AC were 70 dB and 80 dB in the right and left ears, respectively and SIS was 28 % in each ear. In both AD and AE clients, the SRT in each ear was 60 dB and 90 dB, respectively. In client AD, speech recognition thresholds were 80 % and 84 % in right and left ear, respectively. Whereas in client AE, SRT (s) were 32% and 36 % in right and left ear, respectively. Speech in noise (SPIN) test was conducted on both ears of clients’ AC and AE. In client AC the SPIN scores were12 % and 8 % in right and left ear, respectively. In client AE there was no measurable score obtained in each ear on SPIN test. Tympanometric evaluation revealed ‘A’ type on both ears, indicative of normal middle ear status. This was true for all the clients. Ipsilateral and contralateral acoustic reflexes at 500 Hz to 4000 Hz (in octaves) were absent in each ear in all the clients except AB. In client AB, measurable Ipsi- and Contra- reflexes were present at 100 dB. Otoacoustic emissions (OAE) testing documented the presence of transient OAE in each ear for all the client except AD. Auditory brainstem response (ABR) was administered on each client. A click stimulus of 1500 sweeps were presented through insert receiver earphones at initial presentation level of 90 dBnHL at 11.1 /sec repetition rate. The intensity was reduced till the level where no longer identifiable V peak was observed. ABR was absent in AC and AE clients. In client AB, V peak latency was identified till 50 dBnHL in each ear, which is correlated to their behavioural threshold. Whereas, in client AD, an identifiable peak was observed only at 90 dBnHL. From the detailed audiological evaluation provisional diagnosis was made as bilateral moderate SNHL in client AB and moderately severe SNHL in AD client. In addition, AC was diagnosed as bilateral mild SNHL with ANSD and client AE was diagnosed as moderately severe SNHL with ANSD. The audiological findings of each client are tabulated in Table-1.

Table-1. Detail report of audiological findings on each client.

|  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- |
| Clients | AB | | AC | | AD | | AE | |
| EAR | Right | Left | Right | Left | Right | Left | Right | Left |
| PTA | 42.5 | 46.25 | 38.75 | 31.25 | 61.5 | 61.5 | 61.25 | 61.25 |
| SRT | 55 | 60 | 70 | 80 | 60 | 60 | 90 | 90 |
| SIS | 88% | 88% | 28% | 24% | 80% @ 85 dBHL | 84% @ 85 dBHL | 32% | 36% |
| SPIN | --- | --- | 12% | 8% | -- | -- | 0 | 0 |
| UCL | >100 | >100 | >100 | >100 | 90 | 90 | >100 | >100 |
| Tympanometry | A | A | A | A | A | A | A | A |
| Reflexes | P | P | A | A | A | A | A | A |
| ABR | V peak present till 50 dB | V peak present till 50 dB | NR | NR | -- | -- | A | A |
| OAE | A | A | P | P | --- | --- | P | P |
| Diagnosis | Moderate SNHL | Moderate SNHL | Mild SNHL with ANSD | Mild SNHL with ANSD | Moderately severe SNHL | Moderately severe SNHL | Moderately severe SNHL with ANSD | Moderately severe SNHL with ANSD |

*Note – P- Present; A – Absent*

**Hearing handicap**

The Kannada version of Hearing Handicap Inventory for Adults (HHIA)(7) was administered in each client and or informants. There are 25 questions in total. It includes 13 and 12 questions in the emotional and social sub sections, respectively. The scores on the degree of handicap in the emotional and social life of an individual were obtained. Each question was rated on a three-point rating scale ‘yes’ as 4, ‘sometimes’ as 2, and ‘no’ as 0. The maximum overall score is 100. The maximum number of points for social and emotional subsections is 48 and 52, respectively. A score of 0 implies no handicap while a score of 100 implies total handicap. A score ranging from 0-16% indicates no handicap, a score of 18-42% indicates Mild-Moderate Handicap. A score of above 44% indicates a significant Handicap. The score on hearing handicap for AB and AD were 34 % and 40 %, respectively, indicative of mild to moderate hearing handicap. Clients AC (90 %) and AD (92%) were significantly handicap on the hearing handicap questionnaire.

**Hearing aid trial**

Hearing aid trial was performed in all the individuals. Hearing aids were chosen based on the degree of hearing loss and financial affordability of the individual. Aided performance was examined separately for each ear using a five standardized phrases (8) and 25 phonemically and phonetically balanced words (9). Aided performance in each hearing aid was obtained using phrases and words, which were presented at 65 dB SPL delivered from a loudspeaker positioned at 00 azimuth from a distance of 1 meter. A paired comparison method was used to select the appropriate hearing aids. The result of hearing aid trial of prescribed hearing aid on each client is tabulated in Table-2. It was observed that AB and AC are benefited with the hearing aid. However, AC and AE clients reported as able to hear but difficult to follow speech even after fitted with hearing aid. Thus, scores on phrases and word recognition were poor in both AC and AE clients. Hearing aid was proscribed to them for protection from hazardous situation and insisted to wear hearing aid and speech read.

Table-2. Hearing aid trial results of prescribed hearing aid on each client.

|  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- |
|  |  | ***Right ear*** | | ***Left ear*** | |
| ***Clients*** | ***Hearing aid*** | ***Phrases (5)*** | ***Words (25) - %*** | ***Phrases (5)*** | ***Words (25) - %*** |
| ***AB*** | Win 112 | 4/5 | 22/25 – 88% | 5/5 | 22/25 – 88% |
| ***AC*** | Ino P | 2/5 | 6/25 – 20% | 2/5 | 9/25 – 36% |
| ***AD*** | RiA pro Mini RITE | 5/5 | 17/25 -68% | 5/5 | 18/25 – 72 % |
| ***AE*** | Logar 290 | 2/5 | 5/25 – 20% | 1/5 | 3/25 – 12 % |

**Discussion**

The clients ‘AD’ (moderately severe SNHL PTA: 61.5 dB) and ‘AE’ (moderately severe ANSD; PTA: 61.25) were eligible candidate to avail concession/ benefit for their 43.2 % hearing disability. This is because a minimum degree of hearing loss should be 60 dB HL accounts to 40% hearing disability in order become an eligible for any concessions/ benefits (RPWD, 2016). The client ‘AB’ who was diagnosed as mild SNHL (PTA 42.25), which accounts to 10 % hearing disability. Whereas, client ‘AC’ was diagnosed as mild ANSD (PTA better ear: 31.25) accounts to 1%. According to RPWD, 2016 these two clients ‘AB’ and ‘AC’ are not eligible to avail any concession.

The severity of handicap is mild to moderate in clients ‘AB’ and ‘AD’ who had sensorineural hearing loss and both of them were benefitted with the hearing aids. Client ‘AE’ (ANSD) had a significant hearing handicap due to a problem at retro cochlear level. Despite with the hearing aid a benefit was limited to an awareness. In client ‘AC’ (ANSD), though their hearing loss was 31.25 dB in better ear, speech recognition score was severely impaired. In addition, severe hearing handicap was found with no benefit from hearing aid.

The client ‘AC’ had 1 % disability based ready reckoned degree of hearing loss and disability table of RPWD (2016) but their speech impairment is severely impaired. Moreover, client showed no benefit with hearing aid. The disability of such cohort population is underestimated in both PWD (2002) and RPWD (2016). PWD 2002 considered degree of hearing loss and speech identification score for issuing disability certificate. This was considered because if degree of loss increases then speech identification score reduces but this agreement fails in case of ANSD. Whereas, In RPWD 2016 disability certificate are issued to irrespective of degree of hearing loss but to avail the monitory/ financial benefit or reservation for job under quota one should have 60 dB hearing loss corresponds to 40 % hearing disability.

In the present study client ‘AC’ has a genuine problem in understanding speech though having mild hearing loss due to retrocochlear pathology. Whereas, this kind of disagreement is less likely in individuals who give false negative response. This is because they will have loudness yardstick to response to tone and as well to speech. So they would be responding worse on both tone and speech stimuli. Still it is unfortunate that a ANSD who have their degree of hearing loss less than 60 dB HL to avail concession/ benefit.

There is a practical reason for considering only pure tone average for calculating the disability percentage. Some individuals fake their hearing ability or they do not speak at all for monitory benefit and or concessions. In such scenario, using auditory brainstem response for click or tone burst stimulus an audiologist can assess the actual hearing ability though they fake it. Unlike hearing ability, speech perception ability cannot be assessed objectively till date. Unfortunately, none of the audiological test objectively quantifies speech understanding ability. In addition, a case profile like ‘AC’ is less in number compared to overall hearing impairment. That is the prevalence of ANSD is limited to 0.53 to 2.27 [1]. In addition, a few ANSD clients with mild to moderate hearing loss has 70 to 80 % speech identification scores. However, their speech perception score reduces dramatically in 0 dB SNR. Further, hearing handicap questionnaire may not be sensitive because even with the minimum degree of hearing loss client response to have severe degree of handicap.

Exclusively for ANSD patients with mild degree the following criteria may be consider in availing concession/ benefit in near future. To induct criteria of disagreement between PTA and SIS into RPWD act to calculate hearing disability, requires a test which should determines client’s speech perception ability objectively until then it is hard to include. Else, identifying the nodal center to accurately diagnose the ANSD client and administer speech in noise test to confirm hearing disability. Considering the speech identification score in 0 dB SNR is utmost important factor as it is a sensitive test for retrocochlear cases. The above statements are just the author opinion and it is beyond the scope of this present study.

**Conclusion**

Client AC who was diagnosed as SNHL with auditory neuropathy spectrum disorder showed severe speech perception problem with a hearing threshold 31.25 dB HL. Despite with advanced digital hearing aid their speech perception did not improve. In addition, severe hearing handicap was observed. Client ‘AC’ precluded to receive any concession or benefit as the hearing threshold is 31.25 dB in better ear which accounts to 1% disability.

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