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Prevalence of cochlear dead regions in moderate to severe sensorineural hearing impaired children

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ABSTRACT

Objectives: To investigate the overall prevalence of cochlear dead regions in children with moderate to severe sensorineural hearing impairment.
Methods: Threshold-equalizing noise (or TEN) test was administered on thirty sensorineural hearing impaired children (8 girls, 22 boys), aged 5–14 years (mean ± SD ≡ 8.5 ± 2.8).
Results: Classifying by subject, 76% tested positive for cochlear dead regions in one or both ears at least at one frequency. Classifying by ears, 58.3% had dead regions at one frequency or more. Classifying by the number of frequencies were tested, 20% met the criteria for a dead region. The difference between mean absolute thresholds in two groups was statistically significant at 1000 Hz and below ($p < 0.05$).
Conclusion: The results indicated a relatively high prevalence of dead regions in children with sensory neural hearing impairment, especially for frequencies at which the hearing loss exceeds 70 dB HL.

1. Introduction

A dead region in the cochlea is referred to a region with no (or very limited) functioning inner hair cells and/or auditory neurons, so that a tone with frequency falling in that area may be detected at a different place in the cochlea, using off-place, or off-frequency listening [1]. The presence of cochlear dead regions can have considerable negative effects on speech understanding [1,2], and other auditory percepts including perception of puretones [3], growth of loudness [4], and pitch [5]. Considering the significance of diagnosing the dead regions for making clinical decisions such as choice of hearing aids, and optimizing the management of hearing loss in children, it is important to evaluate the overall prevalence of cochlear dead regions in young children with sensorineural hearing impairments.

Audiogram has historically been the main source of information about a patient's hearing loss, but several studies showed that the presence or absence of dead regions cannot be determined reliably from the audiogram [6,7]. In the last decade, a clinical test for discovering the cochlear dead regions has been introduced [7]. The test was based upon the detection of puretones in the presence of a broadband noise – called threshold equalizing noise (TEN) – designed to produce almost equal masked thresholds (in dB SPL) over a wide frequency range [7,8]. The noise was formed in such a way that the threshold for detecting a tone in the noise, specified in dB SPL, was nearly the same for all tone frequencies in the range 250 Hz–10 kHz for people with normal hearing. The masked threshold is approximately equal to the nominal level of the noise specified in dB SPL. When the puretone signal frequency falls in a dead region, the signal will only be detected when it produces sufficient basilar membrane vibration at a remote region in the cochlea where there are surviving IHCs and neurons. The amount of vibration produced by the tone at this remote region will be less than in the dead region, and so the noise will be very effective in masking it. Thus, the signal threshold is expected to be markedly higher than normal. A dead region at a particular frequency is indicated by a masked threshold that is at least 10 dB above the absolute threshold and 10 dB above the nominal noise level [1,7,8]. In a more recent version of the test, masked thresholds can be measured in dB HL [9]. The criteria for diagnosis of dead regions and validation of the test are done mainly in adult patients [1,6–8,10,11]. However, very little is known about the prevalence of cochlear dead regions in hearing impaired young children. Malicka et al. [2] investigated the presence of cochlear dead regions in a group of 12 hearing impaired children (21 ears), aged 7–13 years

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by using TEN test and psychophysical tuning curves (PTCs). Their results showed that all hearing-impaired children were able to perform the TEN test, and the results could be interpreted with the adult criteria. The results of the two tests were consistent in 17 of 21 ears (81%): eight ears did not show evidence of a dead region and nine ears did. In three ears, the criteria for a dead region were met on the TEN test, but there was no evidence of a dead regions on the PTC test. In one ear, the TEN test did not show evidence of dead regions at two frequencies, whereas PTCs did [2]. This agreement rate was consistent with other studies using adult listeners [7,12]. However, much uncertainty still exists about the overall prevalence of the cochlear dead regions in sensorineural hearing impaired children. Moreover, it is not known whether or not to expect different prevalence in larger sample size.

Therefore, current study attempts to show the prevalence of cochlear dead regions for all of the frequencies available in the TEN (HL) test in a larger number of children with moderate to severe sensorineural hearing impairment. Targeting different age group of people with sensorineural hearing loss can be informative if prevalence proves to be different in the different age groups.

2. Material and methods

2.1. Participants

Thirty hearing impaired children (8 girls, 22 boys), aged 5–14 years (mean $\pm$ SD $\equiv$ 8.5 $\pm$ 2.8), were selected through simple randomization by using an online random number generator from the Newsha hearing institute, Tehran, Iran. None of the subjects had a diagnosis of auditory neuropathy spectrum disorders (ANSD), auditory processing disorder (APD), attention deficit/hyperactivity disorder (ADHD), or learning disabilities (LD) based on their clinical records and parental reports. Exclusion of ANSD was based on normal findings in the tests of cochlear hair cell function (Otoacoustic emissions, or Cochlear Microphonics), and abnormal findings in Auditory brainstem response (ABR) to high-level click stimuli (80–90 dB nHL). Exclusion of APD was based on good performance in the Persian Version of the SCAN-C. All subjects had normal middle ear function supported by tympanometric findings. Audiometric thresholds between 45 and 85 dB HL for at least one frequency in the range of 0.25–4 kHz, and air-bone gap of less than 10 dB at all frequencies from 0.25 to 4 kHz were used as eligibility criteria required individuals to have moderate to severe sensorineural hearing loss. Subjects with more than a 35 dB difference in threshold between two ears were excluded from the study. All children and their parents gave their written consent to participate. All procedures were approved by Ethics Committee of the Iran University of Medical sciences.

2.2. Test procedures

In order to rule out the possibility of any external or middle ear condition, otoscopy, and middle ear analysis was done using the AT235 automatic middle ear analyzer (Interacoustics AS, Denmark). Audiometric thresholds (air and bone) were established in a sound-isolated booth according to American Speech-Language-Hearing Association guidelines for manual pure-tone threshold audiometry [13], using AC-40 audiometer (Interacoustics AS, Denmark) with Telephonics TDH39 earphones. The audiometer was calibrated just before starting the current project. The TEN(HL) test was carried out almost following to the protocol described by Moore et al. [9]. For administering the TEN(HL) test, signal tones (0.5, 0.75, 1, 1.5, 2, 3, and 4 kHz) and TEN were played through a CD player connected to the audiometer. For frequencies where hearing loss was up to 60 dB HL, the TEN level sets to 70 dB (unless subjects complained about the loudness of TEN) and for frequencies where hearing loss was 70 dB or more, the TEN level sets 10 dB above the audiometric threshold at that frequency. The level of the TEN was never more than 95 dB HL. The threshold for detecting a test signal in the TEN was determined using the same procedure as used for manual audiometry. At each specific frequency, when the threshold of the test tone in the TEN was 10 dB or more above the TEN level, and the threshold of the test tone in the TEN was 10 dB or more above the absolute threshold, a positive TEN test and diagnosis of a dead region at that particular frequency was indicated. In cases in which the TEN(HL) level could not be made high enough to elevate the absolute threshold by 10 dB or more, the results were considered as inconclusive [1,6,9]. This procedure was then repeated for the other ear. Both ears were tested in a single session.

2.3. Statistical analysis

To determine a significant difference between mean absolute threshold and pure tone average (PTA) for the ears with and without dead regions, and mean difference of signal-to-noise ratio at threshold (i.e., the signal level at threshold in dB HL minus the noise level in dB HL) which is denoted here SNR(T), independent t test was used. All statistical tests were considered significant at $p \leq 0.05$. Statistical analyses were performed using SPSS 16.0 (SPSS Inc., Chicago, USA).

3. Results

Classifying by subject, 23 of the 30 subjects (76%) tested positive for cochlear dead regions in one or both ears at least at one frequency. Twelve subjects met the criteria for a dead region in both ears, and 11 subjects tested positive for dead regions in one ear only. Five of the 30 subjects (17%) were not diagnosed as having a dead region, and in 2 subjects (7%), the result of the TEN-test was inconclusive at both ears. Classifying by ears, 35 of the 60 ears (58.3%) had dead regions at one frequency or more. Classifying by the number of frequencies were tested, 82 of the 420 (7 frequency $\times$ 2 ears $\times$ 30 subjects) frequencies (20%) met the criteria for a dead region. In a hundred of tested frequencies (24%), due to different reasons including insufficient tolerance of high TEN levels or having absolute thresholds of greater than 85 dB HL, results were inconclusive. The dead region criteria were not met in 238 tested frequencies (56%).

Table 1 provides the distribution of the number of ears as a function of the absolute threshold, for test frequencies ranging from 0.5 to 4 kHz, for ears with or without a dead region at any frequency. The number of ears with dead regions is specified in parentheses. Where no number in parentheses is given, there were no cases of dead regions. As shown in Table 1, the prevalence of dead regions increased significantly for hearing thresholds over 70 dB HL. For hearing losses of 75 dB or more (i.e., combining the data for hearing losses of 75, 80, and 85 dB), the percentage of ears with dead regions was 25.4, 45.7, and 44.4% for thresholds of 75, 80, and 85 dB, respectively.

The mean absolute threshold for the ears without dead regions was lower in all of the test frequencies compared to the ears with dead region. This difference in mean threshold was statistically significant at 0.5, 0.75, and 1 kHz, but did not reach to significance level at frequencies 1.5–4 kHz. In the dead region negative group, the pure tone average (PTA) from 0.5 to 2 kHz was 59.6 $\pm$ 5.4 dB. However, in the dead region positive group, the PTA from 0.5 to 2 kHz was 73.1 $\pm$ 11.6 dB and this difference was statistically significant ($p \equiv 0.02$). The value of signal-to-noise ratio at threshold was 2.19 $\pm$ 1.30 dB for the ears without dead regions and 12.22 $\pm$ 4.29 for the ears with dead regions. This difference was statistically significant ($p < 0.001$).