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Short-term hearing fluctuation in Meniere's disease

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Key Words

Meniere's disease
Hearing fluctuation
In-situ audiometry
Self-hearing test
Portable hearing aid programmer
Audiogram configuration

Short-term hearing fluctuation in Meniere's disease

Abstract

This study aimed to assess the extent and implications of short-term hearing fluctuation in Meniere's disease. Thirty-six subjects diagnosed with Meniere's were recruited to measure their own hearing using in-situ audiometry via a hearing aid (Widex Diva) and a portable programmer (SP3). Self-hearing tests measuring up to 14 frequency bands were conducted three times a day over eight weeks using the expanded SensogramTM. Twenty-three ears showed low frequency fluctuation while ten fluctuated in mid frequencies with some 'double peak' audiogram configurations. Eight ears in the later stages of Meniere's, contrary to expected, also recorded fluctuation across all frequencies. Self-hearing testing Meniere's ears over eight weeks revealed great hearing fluctuation with significant changes in audiogram configuration. It suggests that as endolymphatic hydrops progresses through the cochlea, low frequency fluctuation is followed by fluctuation in the mid frequencies, leading to fluctuation across all frequencies. Use of a self-hearing test may facilitate diagnosis and hearing aid fitting for this population, as clinical audiograms may not provide accurate information of hearing fluctuation.

Sumario

Este estudio intenta evaluar el grado y las implicaciones de las fluctuaciones de corto plazo en la enfermedad de Ménière. Se reclutaron treinta y seis sujetos diagnosticados con Ménière para medir su propia audición utilizando audiometría in-situ a través de un auxiliar auditivo (Widex Diva) y un programador portátil (SP3). Se condujo una prueba de auto-evaluación auditiva midiendo hasta 14 bandas de frecuencia, realizada tres veces al día durante ocho semanas usando el Sensogram expandido. Veintitrés oídos mostraron fluctuaciones en las frecuencias graves, mientras que diez fluctuaron en las frecuencias medias, con configuraciones audiométricas de "doble pico". Contrario a lo esperado, ocho oídos en etapas tardías de Ménière también registraron fluctuaciones en todas las frecuencias. El auto-examen auditivo en oídos con Ménière durante ocho semanas reveló grandes fluctuaciones auditivas con cambios significativos en la configuración del audiograma. Se sugiere que conforme el hidropo endolinfático progresa a lo largo de la cóclea, la fluctuación en los graves es seguida por fluctuación en las frecuencias medias, llevando a una fluctuación en todas las frecuencias. El uso de la prueba de auto-evaluación auditiva puede facilitar el diagnóstico y la adaptación de auxiliares auditivos para esta población, dado que los audiogramas clínicos pueden no aportar información precisa sobre la fluctuación auditiva.

Sensorineural hearing loss is one of the four symptoms characterizing Meniere's disease. Numerous studies have considered the shape or configuration of the pure-tone audiogram that occurs in Meniere's disease. Four main hearing loss configurations have been described in the literature: rising (low frequency loss), falling (high frequency loss), peak (low and high frequency loss with better hearing at 1000–2000 Hz), and flat (affecting all frequencies equally) (Savastano et al, 2006; Mateijnsen et al, 2001; Meyerhoff et al, 1981; Stahle et al, 1989; Ries et al, 1999). In the early stages of Meniere's disease there is predominantly a low frequency or peak hearing loss with a tendency to assume a flat configuration as the loss becomes more severe with disease progression. In older patients, the peak audiogram may be a combination of low frequency loss due to Meniere's, and high frequency loss due to presbycusis. This configuration, however, also occurs in young patients with Meniere's disease who have no evidence of a high frequency loss in the contralateral ear, leading to the theory that the high frequency damage is also due to Meniere's disease (Savastano et al, 2006; Ries et al, 1999).

Fluctuation is the main characteristic of hearing in Meniere's disease. Long-term studies have found this fluctuation to be present in the early stages in about 70% of patients through symptom assessment (Green et al, 1991) and audiometry (Savas-

tano et al, 2006), but becoming less prominent as the disease progresses. Only 9% of patients reported continued fluctuation after a minimum of nine years follow-up (Green et al, 1991).

Hearing fluctuation has mostly been associated with low frequency hearing loss, although it has been shown to affect high frequencies to a lesser extent (Savastano et al, 2006; Eggermont & Schmidt, 1985; Mateijnsen et al, 2001). The magnitude, frequency, and pattern of short-term hearing fluctuation however, are not well documented in the literature. Long-term studies inevitably suffer from a paucity of audiograms. Even where an audiogram could be performed every three months over a time span of 10 years, only 40 audiograms would be available.

Savastano et al (2006) recently described pure-tone audiometric fluctuation and the mean threshold shift in Meniere's patients in the early stages, but it was not clear how many audiograms were used to determine fluctuation. Other studies have analysed fluctuations but based only on the patients' subjective assessment (Green et al, 1991; Mateijnsen et al, 2001). Eggermont and Schmidt (1985) were able to present data on hearing fluctuation effectively in a long-term study using particular cases to illustrate observed trends.

The reliability of subjective assessment without documented audiogram, however, is highly questioned by the first author, as

patients are usually not able to detect fluctuations especially when best hearing thresholds are at levels below 50 dBHL. Clinical audiometry on the other hand cannot be relied upon to record hearing fluctuation, as they may not be performed frequently enough.

Demonstration of fluctuation requires a series of audiograms performed at regular intervals for a period of time. Audiograms performed several times a day would be the most reliable method to demonstrate hearing fluctuation patterns in an individual patient. Such procedures however are very time consuming and not viable in clinical settings.

Audiograms are traditionally performed using an audiometer. More recently audiologists have been able to perform in-situ audiograms in the clinic without an audiometer, but through hearing aid fitting software. This is a reliable way of assessing hearing threshold using a custom made ear mould instead of headphones with the test stimuli produced by the hearing aid.

The hearing instrument manufacturer Widex was the first to release a portable device which measures in-situ audiograms with the Senso Diva hearing aid. This procedure is trade marked as 'Sensogram' and allows the clinician to measure hearing thresholds in up to 14 sound frequency bands through the hearing aids, either connected to a computer using Hi-Pro or Noah Link, as well as via the SP3 which is Widex's portable hearing aid programmer.

Test-retest reliability with in-situ threshold measurements has been shown to be equivalent to that of currently accepted audiometric procedures. A previous study of in-situ hearing threshold measurement using the Widex Senso Diva and the SP3 (Smith-Olinde et al, 2006) found excellent test-retest reliability, which they considered equivalent to conventional audiometry. This study measured four frequencies (500, 1000, 2000, and 4000 Hz) finding that 98%, 100%, 100%, and 93% respectively of all patients fell between ± 5 dBHL with standard deviations of 3.14, 1.86, 1.97, and 4.02 dBHL. Furthermore, this hearing aid system enables performance of more detailed audiograms than some audiometers. It allows hearing threshold testing in 5-dB steps from 0 up to 100 dB at the frequencies of 250, 350, 500, 630, 800, 1000, 1250, 1600, 2000, 2500, 3200, 4000, 6000, and 8000 Hz, depending on the hearing aid model.

We used this system to assess the magnitude and pattern of short-term hearing fluctuation in Meniere's disease by asking a group of subjects to measure their own hearing thresholds several times a day over a period of time. It was our hope that these measurements would provide diagnostic confirmation and better insight into the audiological aspects of Meniere's disease.

Methods

Ethics and recruitment

Ethical approval for this study was obtained from Macquarie University Ethics Review Committee (Human Research). The study was advertised through a newsletter distributed by the Meniere's Support Group of New South Wales (Australia). Volunteers were recruited if they had a hearing loss in at least one ear due to Meniere's disease diagnosed by a specialist in otolaryngology. Diagnostic criteria was based on Gibson's 10 points scale and confirmed by trans-tympanic electrocochleography (ECoChG) (Conlon & Gibson, 1999). ECoChG was carried out using 1000-Hz, 100-dBHL stimulation with a 16-

ms tone burst (1-ms rise/fall and 14-ms plateau) at a rate of 30/s. The deflection was measured at approximately 10 ms. ECoChG was confirmative when a summing potential was more negative than $-6 \mu\text{V}$. Previous studies using this technique have determined that using this diagnostic threshold provides the optimum compromise between sensitivity and specificity (Gibson, 1996). Stage and duration of the disease, degree of hearing loss, and ear affected were not taken into account when recruiting for this study. We also did not consider any current or previous medical interventions that subjects may have undertaken along their disease process or during the study, such as diet, medication, and/or surgery. Subjects were expected to continue with their normal life styles during the study but needed to be conversant with technology and capable of testing their own hearing at home for a period of at least eight weeks.

Subjects

Thirty-six volunteers diagnosed with Meniere's disease, 20 females and 16 males aged from 33 to 78 years, were recruited and fitted with Widex Senso Diva hearing aids. Twenty-seven subjects were fitted monaurally and nine binaurally, according to the hearing loss, giving a total of 45 ears fitted with hearing aids. Four of the nine subjects with bilateral hearing loss had a contralateral sensorineural hearing loss unrelated to Meniere's, and five had bilateral disease.

Control group

A total of eight ears were used as control. One female subject with bilateral moderate mixed hearing loss due to otosclerosis and another with a congenital bilateral flat mild to moderate sensorineural hearing loss, measured their hearing in both ears twice a day for a period of two weeks. Four subjects of the study group were fitted with bilateral hearing aids in spite of having Meniere's disease in one ear only (as described before) and were also asked to measure their hearing bilaterally so that the contralateral ear could be used as a control.

Material and Procedure

Participants were fitted with Widex Senso Diva hearing aids with a custom mould to suit the hearing loss at the time of fitting. The fitting protocol was based on the Widex proprietor's procedure using the expanded Sensogram (Ludvigsen, 2001). The audiologist performed this via the Noah Link measuring the hearing thresholds at up to 14 frequency bands followed by the 'feedback test' as prescribed by the Widex fitting protocol. The Widex Compass software automatically programmed the hearing aids based on these two measurements. The 'expanded Sensogram' procedure was selected from the Compass menu (Ludvigsen, 2001). This procedure allows the measurement of hearing thresholds at 13 frequency bands from 250 to 6000 Hz in the behind-the-ear SD-9 and SD-19 models, and 14 frequency bands from 250 to 8000 Hz, in the custom devices SD-X ITC and SD-CIC models. A maximum stimulation level of 85 dB HL at 250 and 350 kHz, 90 dB at 500, 630, and 800 Hz, 95 dB at 1000 and 1250 Hz, and 100 dB at 2000, 3200, 4000, and 6000 Hz can be achieved using the SD19 model, while the custom aids produce frequency stimulation up to 8000 Hz at 80 dB with the ITC SD-X, and 75 dB with the SD-CIC models.

The participants were then taught how to connect the SP3 device to their hearing aid and to perform their own expanded Sensogram. Their ability to carry out this procedure was checked in the clinic, by asking them to repeat the measurements made by the audiologist earlier in the session. They were provided with an SP3 to take home and were requested to perform their Sensograms three times per day over a period of at least eight weeks and record these on a spreadsheet. They were instructed to perform these tests in as quiet an environment as possible while plugging the contralateral ear with their index finger. Whenever possible, tests were to be performed in the same room every time. They were also requested to inspect the earmould prior to testing to exclude the presence of any obstruction, which would interfere with sound delivery. A follow-up visit was carried out two weeks after the initial fitting to ensure Sensograms could be performed with ease. Telephone follow-ups were also conducted as needed.

Prior to measuring their hearing, subjects were also asked to rate their symptoms of ear fullness, tinnitus, and vertigo on a scale 1–5, to be recorded on the spreadsheet along with their hearing thresholds at each tested frequency.

Analysis

Including the control group, a total of 49 ears were included in the study. Every Sensogram obtained from each individual participant was entered in an Excel spreadsheet. These data were plotted in one single audiogram format graph, making it easier to visualize the hearing fluctuation for each subject (example in Figure 1). These graphs were visually inspected by three audiologists independently and classified according to the fluctuation into predominantly low frequency, mid frequency, all frequencies, or no fluctuation.

The magnitude and configuration of hearing fluctuation of each participant was also plotted using the standard deviation from the mean at each frequency measured (example in Figure 2). This allowed easier comparison of the pattern in hearing fluctuation between participants. Considering that test-retest reliability of psychophysical hearing assessment in subjects with sensorineural hearing loss is 10 dBHL (± 5 dBHL), a standard deviation of up to 2.5 dBHL of the mean audiometric threshold was assumed as normal. Therefore only threshold fluctuations of greater than 2.5 dB were considered as significant.

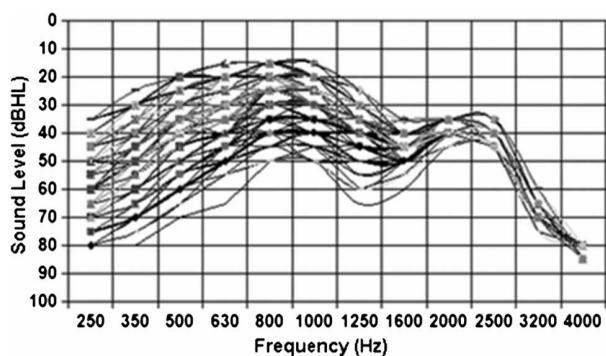


Figure 1. Example of a low frequency fluctuating hearing loss showing data from 247 audiograms where the greatest changes in hearing thresholds were measured at 250–500 Hz.

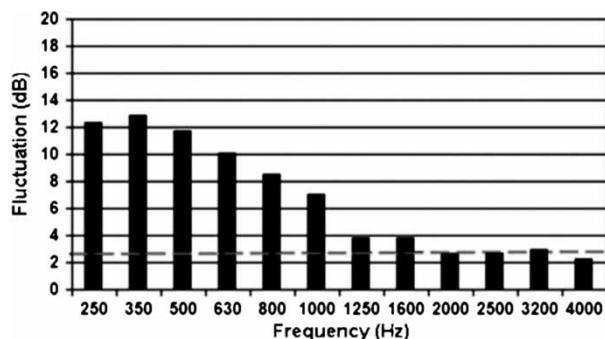


Figure 2. Example from the same patient as Figure 1 where the hearing fluctuation is displayed as the standard deviation of the mean change in hearing at each frequency. The dashed line represents the expected fluctuation from conventional test-retest measurements.

Results

All thirty-six participants with Meniere's disease plus two others, with hearing losses from causes other than endolymphatic hydrops, complied with the task of measuring their own hearing on a regular basis. The minimum number of Sensograms provided by a participant was 25 and the maximum was 381, giving a total of 5316 audiograms. These records provided an equivalent mean of 118 audiograms per ear over a three month period.

Visual classification of audiograms resulted in 23 ears defined as having a low frequency hearing fluctuation, 10 ears with a mid frequency fluctuation, eight ears with an all frequency fluctuation, and eight ears presented no fluctuation. Inter-observer agreement was achieved by all three audiologists for 47 ears, with the remaining two classified by majority decision.

Figure 1 is an example of one subject with a low frequency fluctuating hearing loss showing 247 audiograms where the greatest changes in hearing thresholds were measured at 250–500 Hz. Figure 3 is an example of a mid-frequency fluctuating hearing loss showing 110 audiograms of another subject where the greatest fluctuation peaked at any one frequency around 800–3000 Hz. Figure 4 exemplifies a case of an all frequency fluctuating hearing loss showing 188 audiograms where the fluctuation appeared similar for the majority of frequencies.

The standard deviation of the mean change in hearing for these three different patients is respectively demonstrated in Figure 2, 5, and Figure 6. The area below the dashed line represents the expected fluctuation in hearing thresholds for subjects with normal hearing or a stable sensorineural hearing loss. Figure 7 shows the group data of the 23 ears with a low frequency fluctuating loss, Figure 8 shows 10 ears with mid frequency fluctuation, and Figure 9 displays the eight ears with all frequency fluctuation.

Three ears were found to return to normal thresholds when their hearing was at its best (two with low frequency, and one with all frequency hearing loss). When the shape of the audiogram was considered, it was clear that the same one patient could change between different configurations (e.g. peak to flat or falling to flat), depending on the site of fluctuation, at potentially daily intervals. Figure 10 is an example of the fluctuation that may

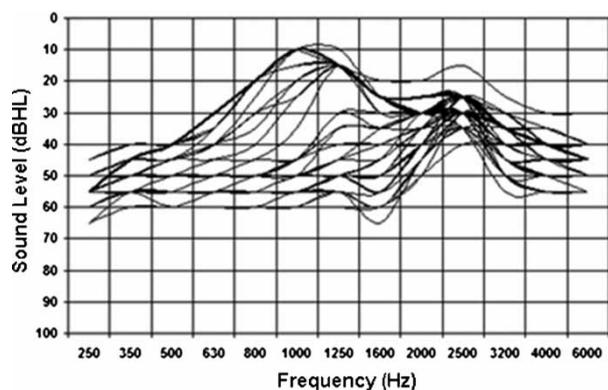


Figure 3. Example of a mid-frequency fluctuating hearing loss showing data from 110 audiograms where the greatest fluctuation peaked at any one frequency around 800–3000 Hz.

occur for a single patient within a period of 24 hours. When the best thresholds obtainable at each frequency (not necessarily from the same point in time) were examined together, 16 ears showed a ‘double peak’ audiogram, with better hearing peaks at 800–1000 Hz and 2000–2500 Hz, and a dip at 1600 Hz (examples in Figures 1 and 3).

No significant fluctuation, (greater than ± 5 dB) were recorded by the control group at any of the tested frequencies, suggesting good test/retest reliability. This included the four participants who had unilateral Meniere’s disease and recorded significant fluctuation in the contralateral ear. These participants recorded bilateral tests performed at the same time and so would have had similar test conditions. This suggests that the change in measured thresholds in the active ear reflects reliable hearing threshold fluctuations, at least for these patients.

Methodological bias

There are several potential sources of bias with regard to our methodology as subjects tested their own hearing in a non-controlled environment. The previous study assessing test/retest reliability of the Sensogram was performed in a soundproof booth (Smith-Olinde et al, 2006) whereas our participants were instructed to conduct their hearing measurements in a ‘quiet’ room at home or at work.

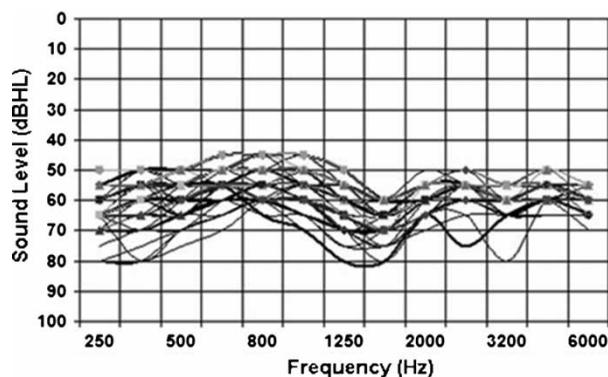


Figure 4. Example of an all-frequency fluctuating hearing loss showing data from 188 audiograms where the fluctuation appeared similar for the majority of frequencies.

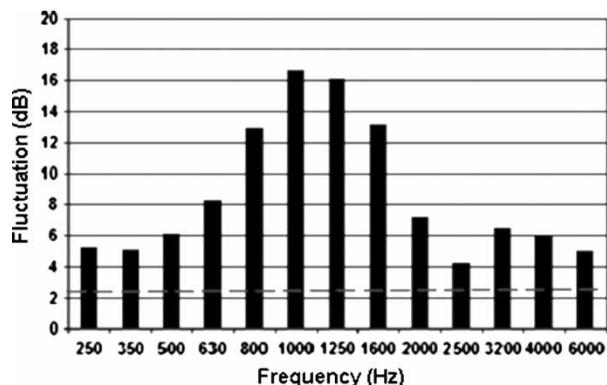


Figure 5. Example from the same patient as Figure 3 where the hearing fluctuation is displayed as the standard deviation of the mean change in hearing at each frequency. The dashed line represents the expected fluctuation from conventional test-retest measurements.

A large study has compared hearing thresholds in a sound-proofed environment with those from the same participants in the workplace with a background noise of up to 68 dB (Wong et al, 2003). This found that standard deviations of the test differences were 8–11 dB, suggesting background noise has a significant effect on test/retest reliability. It seems however unlikely that the background noise at the test environment of our participants was as great as the one in that study but this effect cannot be discounted.

The participants were also not blinded to their test results, as they would be during conventional testing. However, this was pointed out as a positive factor by many participants suggesting that self-testing may elicit far less false positive responses. This factor nevertheless needs researching.

Given the large number of audiograms performed by each participant in this study, it seems unlikely that bias had an effect in the overall results, however further investigation is needed to confirm reliability of self-hearing tests performed in a non controlled environment.

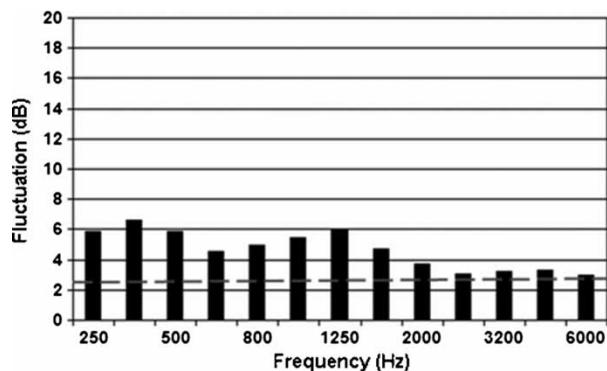


Figure 6. Example from the same patient as Figure 4 where the hearing fluctuation is displayed as the standard deviation of the mean change in hearing at each frequency. The dashed line represents the expected fluctuation from conventional test-retest measurements.

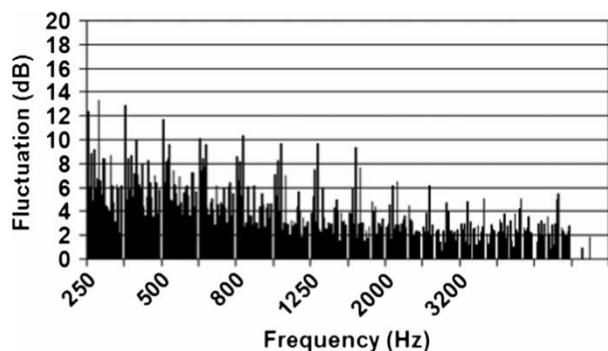


Figure 7. The group data of the 23 ears with low frequency fluctuating hearing loss showing standard deviation of the mean change in hearing at each frequency.

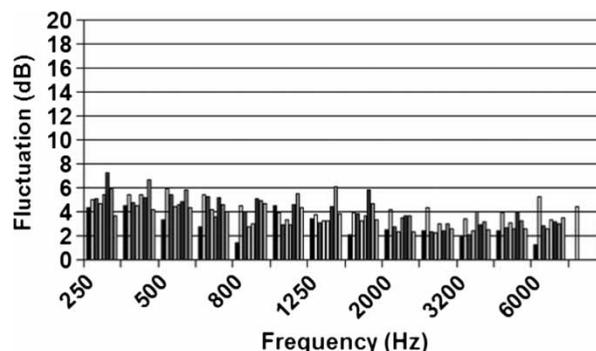


Figure 9. The group data of the eight ears with all frequency fluctuating hearing loss standard deviation of the mean change in hearing at each frequency.

Discussion

To the best of our knowledge this is the first study to examine hearing fluctuation in Meniere's disease on a daily basis and with the addition of intermediate frequencies. It was found that there was a wide variety of audiogram shapes and hearing fluctuation, with some fluctuating as much as 60 dBHL. Although the low frequencies were the commonest site of fluctuation, a significant proportion had greatest variation at the middle frequencies. The same subjects could change between different audiogram shapes (e.g. peak to flat or falling to flat), depending on the frequencies where fluctuation occurred, at potentially daily intervals. These shapes also included a 'double peak' audiogram with better hearing thresholds at 800–1000 Hz and 2000–2500 Hz. In general the high frequencies (4000–8000 Hz) fluctuated less than the low and mid frequencies, but there were still fluctuations in a minority of subjects.

High frequency losses were also noted in some younger patients with normal hearing in the contralateral ear. As the mean hearing loss became more severe, hearing tended to fluctuate less and to assume a flat audiogram. However even with this group there could still be a significant fluctuation of up to 30 dBHL.

This study allowed for a more detailed assessment of hearing fluctuation that may occur over a short period of time, which

would not have been possible using routine audiological assessments in the clinic. The extent of hearing fluctuation found in our results was more than we would have predicted based on current literature. Furthermore hearing fluctuation seems to occur independently of the other symptoms of Meniere's disease and was mostly not associated with vestibular episodes according to our preliminary findings.

The relationship between hearing fluctuation and vertigo attacks, although of great interest, is beyond the scope of this paper. Further analysis looking into correlations of fluctuation amongst the symptoms of Meniere's disease is currently underway as part of a larger study.

Some of our findings are in agreement with those of Eggermont and Schmidt (1985) as they also demonstrated fluctuation to be most prominent at the low frequencies and least at the high frequencies. Our results also show that as hearing loss progresses to more severe it does become flatter in configuration as in other reports, but in our group fluctuation, although reduced, still continued to occur.

A proportion of our subjects had greatest fluctuation at the mid frequencies with relatively little at the lows, which as far as we are aware, has not been previously described in the literature. The peak audiogram is a prominent feature of Meniere's disease, particularly in the early stage, albeit with significant discrepancy between studies as to its prevalence (Savastano et al, 2006; Mateijns et al, 2001; Meyerhoff et al, 1981; Stahle et al, 1989; Ries et al, 1999). The variability in audiogram configuration for each individual found in this study confirmed previous findings (McNeill, 2005). It shows that a single patient can have significant changes in audiogram configurations within a relatively short space of time, particularly in those presenting with mid frequency fluctuation. This may be why different studies have found discrepancy in the prevalence of the peak audiogram. Previous studies have also variously found that this peak occurs at either 1000 or 2000 Hz (Meyerhoff et al, 1981; Ries et al, 1999). With the addition of intermediate frequency evaluation in this study, we have consistently found two separate peaks with an intervening dip at 1600 Hz, showing that the same patient may have peaks at different frequencies at different times.

It is however possible that this double peak finding is an artefact, secondary to an insertion loss effect due to the presence of a hearing aid mould in the ear canal during in-situ threshold

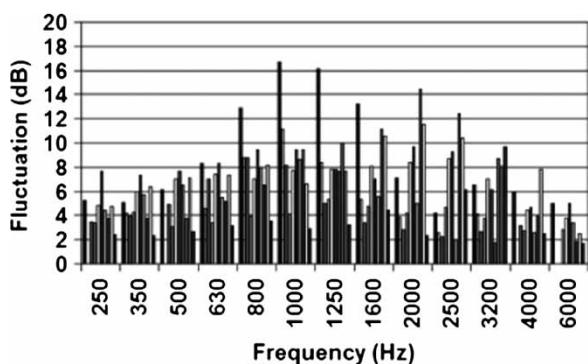


Figure 8. The group data of the 10 ears with mid frequency fluctuating hearing loss standard deviation of the mean change in hearing at each frequency.

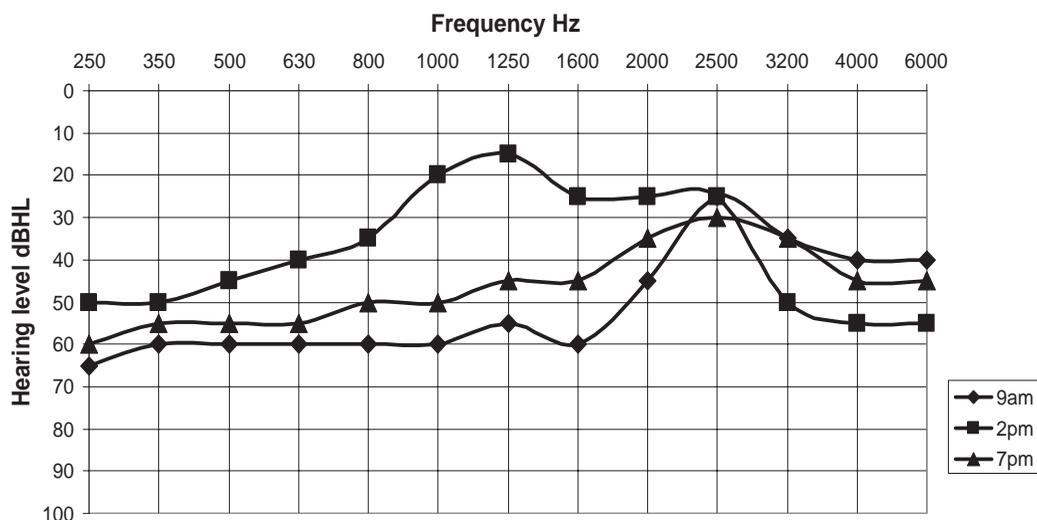


Figure 10. Example from the same patient as Figures 3 and 5 showing the daily hearing fluctuation from three audiograms during the same 24 hour period.

measurement. This hypothesis however needs to be further verified in future studies.

These results have different implications in the diagnosis and treatment of Meniere's disease. A self-testing protocol to assess hearing fluctuation may be a useful tool in the diagnosis of endolymphatic hydrops. The difficulty in establishing a firm diagnosis is in many instances imposed by the fact that when the patient reaches the clinic the symptoms may have subsided. A self-hearing assessment will allow the patients to provide the clinician with a much more accurate picture of the cochlea status.

Our findings also help to explain the difficulties in satisfactorily fitting hearing aids to patients with Meniere's disease. As previously reported (McNeill, 2005), these patients usually leave the audiology clinic satisfied with a new fitting only to return a few days or even hours later, complaining that the sound through the hearing aid is either too booming, too muffled, or distorted. It has been suggested that such poor outcomes are associated with low tolerance to amplification due to recruitment and diplacusis (Valente et al, 2006). Our conclusion however, is that the difficulties associated with fitting hearing aids to this population is most likely due to the erratic fluctuation in the hearing levels. Hearing fluctuation leading to changes in audiogram configuration, as found in this study, explains why neither a volume control nor even a sophisticated multi-program hearing aid has been able to effectively improve hearing in so many patients with Meniere's disease.

In keeping with current theories (Braun, 1996; Xenellis et al, 2004), our findings suggest that the hearing loss of Meniere's disease is due to a combination of endolymphatic fluid changes with basilar membrane displacement and outer hair cell dysfunction, leading to temporary threshold shifts, and hair cell damage, which will ultimately turn into permanent threshold shift.

All the participants in our group had reduced low frequency hearing at some stage, with mean thresholds usually at 40–60 dBHL. The pattern found in the 'all frequency' fluctuating group seems to be predominantly of those who are in the so-called 'burnt out' stage of Meniere's disease (Gibson, 1999). In this group the

mean hearing thresholds and magnitude of fluctuation is reduced across all frequencies most likely due to progressive hair cell damage. One exception in this group was one patient with better hearing. We speculate that hydrops in this individual may have spread beyond the apex without concurrent permanent damage, allowing hearing to return to normal at times. Damage to the high frequencies of the basal turn seems to be less related to fluctuation. Either this may be a different pathological process or these hair cells are more readily damaged by fluid changes. Patterns of hearing fluctuation therefore may be a clear indication as to which part of the cochlea may be affected by fluid changes and which part is due to hair cell damage.

Based on our findings we hypothesize that the pattern of low frequency fluctuation may be due to hydrops in the cochlear apical region. The mid-frequency fluctuation may occur as there is already permanent damage in the apex so that the middle cochlear regions become more affected by endolymphatic hydrops. In the late stages of Meniere's disease the whole cochlea has been affected so that the hearing becomes more impaired with the audiogram assuming a flatter configuration but small fluctuations in hearing thresholds may still continue to occur with eventual changes in endolymphatic pressure. The precise correlation of stage and duration of disease with the degree of fluctuation of hearing loss requires further study.

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