

Original Article

# Speech Recognition During Follow-Up of Patients with Ménière's Disease: What Are We Missing?

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**BACKGROUND:** Hearing loss causes a significant reduction in the quality of life of patients with Ménière's disease. Although speech recognition is also affected, it has not been extensively studied. The objective of the study was to describe speech recognition behavior during a prolonged period in patients with unilateral Ménière's disease.

**METHODS:** A prospective case-control study was performed. The case group included patients with defined unilateral Ménière's disease and the control group included patients with progressive non-fluctuating hearing loss. Patients underwent an auditory evaluation periodically. Pure-tone audiometry and speech recognition tests—speech recognition threshold and speech discrimination score—were administered. The dissociation between pure-tone audiometry and speech recognition was assessed through a linear regression analysis. During follow-up, Ménière's disease patients were subdivided into a stable and fluctuating subgroup (a change of >20% in the speech discrimination score with a change no greater than 15 dB in pure-tone audiometry).

**RESULTS:** The average follow-up time was 79.9 months. Fifty-seven patients were included (30 cases, 27 controls). Dissociation between pure-tone audiometry and speech recognition threshold began to appear in the case group after 21 months, and it was statistically significant at 108 months. Duration of the disease was the only variable studied that influenced the dissociation. The fluctuation subgroup included 56.6% of the cases.

**CONCLUSION:** We described 2 audiological peculiarities in Ménière's disease patients: dissociation between pure-tone audiometry and speech recognition during the evolution of the disease and the fluctuation of speech recognition regardless of the change in pure-tone audiometry. Our results highlight the importance of performing speech recognition tests during follow-up in patients with Ménière's disease.

**KEYWORDS:** Hearing loss, ménière disease, pure-tone audiometry, speech discrimination, speech recognition

## INTRODUCTION

Ménière's disease (MD) is characterized by recurrent fluctuating sensorineural hearing loss (HL), tinnitus, and aural fullness that occur during the episodes of vertigo. Currently, idiopathic endolymphatic hydrops is considered the major pathophysiological event and histologic marker of this disease.<sup>1,2</sup>

Even though HL is not the most disabling symptom, it causes a significant reduction in the quality of life in these patients.<sup>3</sup> Since the very first guidelines for MD diagnosis published in 1985,<sup>4</sup> HL has been only addressed using mean pure-tone audiometry (PTA). The way HL evolves during follow-up of patients with MD has been the subject of several papers. Most authors agree that there is a low frequency sensorineural HL that fluctuates and deteriorates until a moderate or severe HL with a "flattened" audiogram is reached.<sup>3</sup> Although the audiogram shape does not depend on the disease duration, the progression in HL in the first 2 years, and in particular when it occurs in middle and high frequencies, is related with a poor hearing prognosis.<sup>5</sup>

Speech recognition (SR), its progression or fluctuation, has not been extensively studied. In general, dissociation between PTA and SR has been described.<sup>6</sup> Mateijsen<sup>7</sup> reported a correlation between the speech reception threshold (SRT) shift and maximum speech

discrimination (MSD) with the PTA in the affected ear of patients with unilateral MD. The MSD was 100% for PTA of up to 40 dB, above which there was a drastic reduction, showing a wide distribution of scores, from a normal range to very low scores in MSD percentage.

The objectives of this study included: (1) describing the audiometric evolution during a prolonged period, through hearing thresholds and SR tests, in patients with unilateral MD and in a control group of patients with a progressive non-fluctuating HL without MD; (2) determining the existence of dissociation between PTA and SR in patients with MD and the possible variables related to it; and (3) determining if MD patients are able to present a fluctuation in SR, without necessarily having a fluctuation in the PTA.

## METHODS

A prospective case-control study was performed. Patients included in both groups underwent an auditory evaluation periodically, which consisted of a PTA and the assessment of SR, measured by SRT and speech discrimination score (SDS). Ethical committee approval was received from the ethical committee of the Clínica Universidad de Navarra (2020/047, 14/02/2020). Informed consent was obtained from all participants who participated in this study.

## Subjects

Case group included patients with unilateral MD that accomplished "definite" MD criteria according to guidelines published in 1995<sup>8</sup> and its amendment in 2015.<sup>9</sup> Control group included patients with a known progressive, non-fluctuating, and unilateral or bilateral HL. For bilateral HL cases, the most affected ear was the one included. The duration of disease was more than 6 months for both groups. Demographic and clinical variables such as cardiovascular risk factors (hypercholesterolemia, high blood pressure, smoking, and diabetes), age, and duration of disease were compiled. Depending on the presence or absence of vertigo spells in the last 6 months previous to visit, cases were considered as "active" or "quiescent." The patients in the case group were treated during hearing fluctuations (with or without accompanying vertigo spells) with steroids for a short period of time (intramuscular 1-day treatment or oral 21-day treatment). In between, they followed different medications. Exclusion criteria included bilateral MD patients, patients treated surgically, and those treated with intratympanic steroids or gentamicin.

## Data Collection and Audiological Evaluation

Standard pure-tone air-conduction audiometry with headphones was used in all patients (Audiotest, Equinox IEC 645-1/ANSI S3.6-1996 type I, IEC 645-2/ANSI S3.6-1996 type B, Denmark). Pure-tone average threshold (PTAt) data were calculated from 5 frequencies (0.25, 0.5, 1, 2, and 4).

Speech reception threshold is defined as the intensity (dB) at which the subject correctly identifies 50% of words, and SDS as the percentage (%) of words correctly identified, under headphones from a standardized list that is recognized and repeated by the subject. Both tests were performed with the disyllabic phonetically balanced word identification test of Cárdenas-Marrero.<sup>10</sup> This test was performed in a soundproof booth, with the patient located at 1 m from each speaker, at a 45° angle. The intensity of stimulation was 65 dB HL. The speech-audiometric materials were presented once on

a compact disc recording. The item could not be repeated. The lists for adults included 20 groups of 25 meaningful, phonetically balanced, disyllabic words. Two groups of words were presented in each session. The patients answered correctly when they repeated the same word, without changing any phoneme. The results were presented as a percentage of correct answers. Both hearing tests were performed masking the other ear using the Goldstein method.<sup>11</sup> All visits during the follow-up period were scheduled according to the criteria of the treating physician or by the patient's demand, without any specific time pattern. In the case of patients with MD, they could also occur due to relapsing vertigo spells. All subjects included fulfilled informed consent.

In order to define the dissociation between PTA and SR, a linear regression analysis was carried out with the differences between PTAt and SRT. Dissociation was considered when the 95% CI of the line generated did not include 0 between its limits.

In this study, a fluctuation in SR was defined as a change of >20% in the SDS at 65 dB HL with a change no greater than 15 dB in PTAt during a follow-up visit. We chose to fix the intensity at 65 dB because it is a comfortable level for listening to speech in quiet or low levels of background noise and because it corresponded to the level of normal conversation speech in the Spanish language at 1 m.<sup>12,13</sup> The MD group was subdivided into 2 groups (fluctuating subgroup and stable subgroup), considering the presence of fluctuations of SR during the follow-up visits and regardless of the activity of the disease. Only those patients who did not present any fluctuations of SR during the follow-up visits were considered within the stable group.

## Statistical Analysis

The influence of the variables included on the dissociation between PTAt and SRT in both groups was assessed by an analysis of covariance (ANCOVA). In order to be able to evaluate if the activity of the disease was related to a fluctuation in SR at each visit within the group of patients with MD, all visits were considered independently and not by each patient independently.

To assess the evolution of PTAt and SRT throughout the duration of disease for each group, a regression analysis with fractional polynomials was performed, due to the non-linearity of the results at each group. Time 0 was considered as the first audiological evaluation and not as the beginning of the disease.

Once the regression equation was obtained, crossing points between both measures for each group were obtained by iteration using the package Solver in Excel. With the aim of establishing statistical differences in each group, a regression analysis with fractional polynomials was performed to measure hearing thresholds and SRT along duration of disease.

A *P*-value less than .05 was considered as statistically significant. Quantitative data were shown as mean (standard deviation) if they were normally distributed or median (p25; p75) if not. Qualitative data were represented as *n* (%). All statistical analysis was carried out using StataIC 12 (StataCorp. 2011. Stata Statistical Software: Release 12. College Station, TX: StataCorp LP).

**Table 1.** Demographic and Clinical Variables

	MD Group (n = 30)	Control Group (n = 27)
Gender (Male:Female)	19:11	10:17
Age	58 ( $\pm 2SD$ )	67 ( $\pm 2SD$ )
Duration Disease (Months)	48 (48)	132.9 (151.6)
Cardiovascular Risk Factors	3 (9.6%)	13 (48.1%)

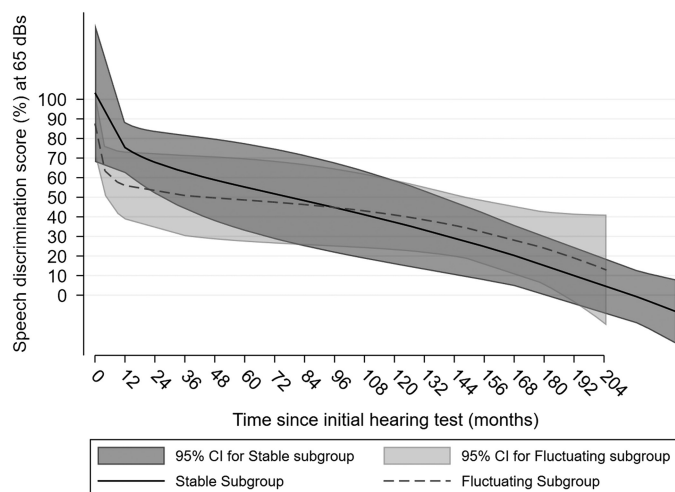
MD: Meniere Disease, SD: Standard Deviation.

**RESULTS**

Fifty-seven patients were included in the study. Demographic and clinical variables are shown in Table 1. A total of 27 patients were included in the control group after excluding 3 because the exact timing of the disease was unknown. The average follow-up time for all patients was 79.9 months ( $SD=46.2$ ). During this period, between 2 and 4 follow-ups took place per patient. Regarding the cases, the average follow-up time was 76.4 months ( $SD=56.1$ ), with an average difference between the first and second follow-up visit of 25.9 months ( $SD=20.6$ ), between the second and third 25.5 months ( $SD=37.1$ ), and between the third and fourth 26.9 months ( $SD=37.1$ ). For controls, the average follow-up time was 83.4 months ( $SD=35.4$ ), with an average difference between the first and second follow-up visit of 42.4 months ( $SD=38.4$ ), between the second and third 25.3 months ( $SD=20.7$ ), and between the third and fourth 23.1 months ( $SD=15.6$ ).

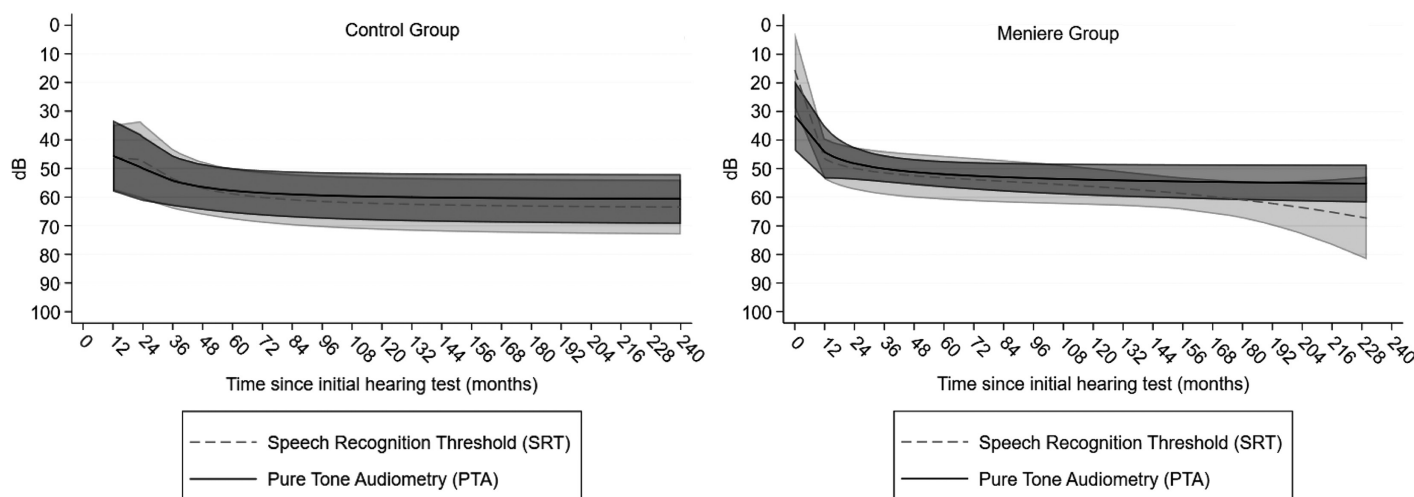
In Figure 1, the PTAt and SRT results along follow-up are shown. In both groups, as time passes, the results reflect a more severe auditory deficit. In patients diagnosed with MD, during the first audiometric studies, PTAt and SRT showed a parallel behavior, both increasing progressively. At 21 months, dissociation between PTAt and SRT was seen, as SRT was higher than PTAt, and at 9 years (108 months), the difference was statistically significant ( $P < .05$ ). Duration of disease was the only variable studied that may influence this dissociation. Contrarily, in the control group, both PTAt and SRT didn't show dissociation in successive follow-up visits, and both values became increasingly higher without any significant difference ( $P > .05$ ).

In the MD group, 56.6% (17/30) of patients presented a fluctuation in SR (fluctuation subgroup) and this was not related to the duration of

**Figure 2.** Speech discrimination score at 65 dB in fluctuating subgroup and stable subgroup. Shade areas indicate the 95% CI for the mean data.

the disease, presence of cardiovascular risk factors, sex, age, and headache/migraine. A variation in SDS at 65 dB was seen in this subgroup, for better or for worse and independently of the PTAt. The variation of the SDS at 65 dB during each visit was 10.8 % ( $SD=21.2$ ) in the fluctuation subgroup and 7.3% ( $SD=16.5$ ) in the stable subgroup. Up to approximately the first 2 years, SDS at 65 dB was different between the 2 groups. The 95% CI of the 2 subgroups included the average of the other subgroup between their boundaries (Figure 2). Pure-tone average threshold and SDS at 65 db throughout the follow-up time of each patient in the MD group is represented in Figure 3. Neither of the audiometric studies followed the same pattern in all the patients, and interestingly, SDS at 65 dB might improve or get worse regardless of the changes in PTA thresholds.

Eighty-two follow-up visits were analyzed in the MD group. In 29 (35.3%), the disease was active and in 63 (76.7%), it was quiescent. Active cases had a variation of 23.0% in the SDS at 65 dB and 14.1 dB in PTAt during each follow-up visit, compared to a variation of 11.3% and  $-1.23$  dB in quiescent cases. The activity of the disease was not significantly related to changes in the SDS at 65 dB ( $P=.168$ ); however, it was significantly related to the changes in PTAt ( $P=.001$ ).

**Figure 1.** Pure-tone audiometry and speech recognition thresholds through time. Gray areas indicate the 95% CI for the mean data.

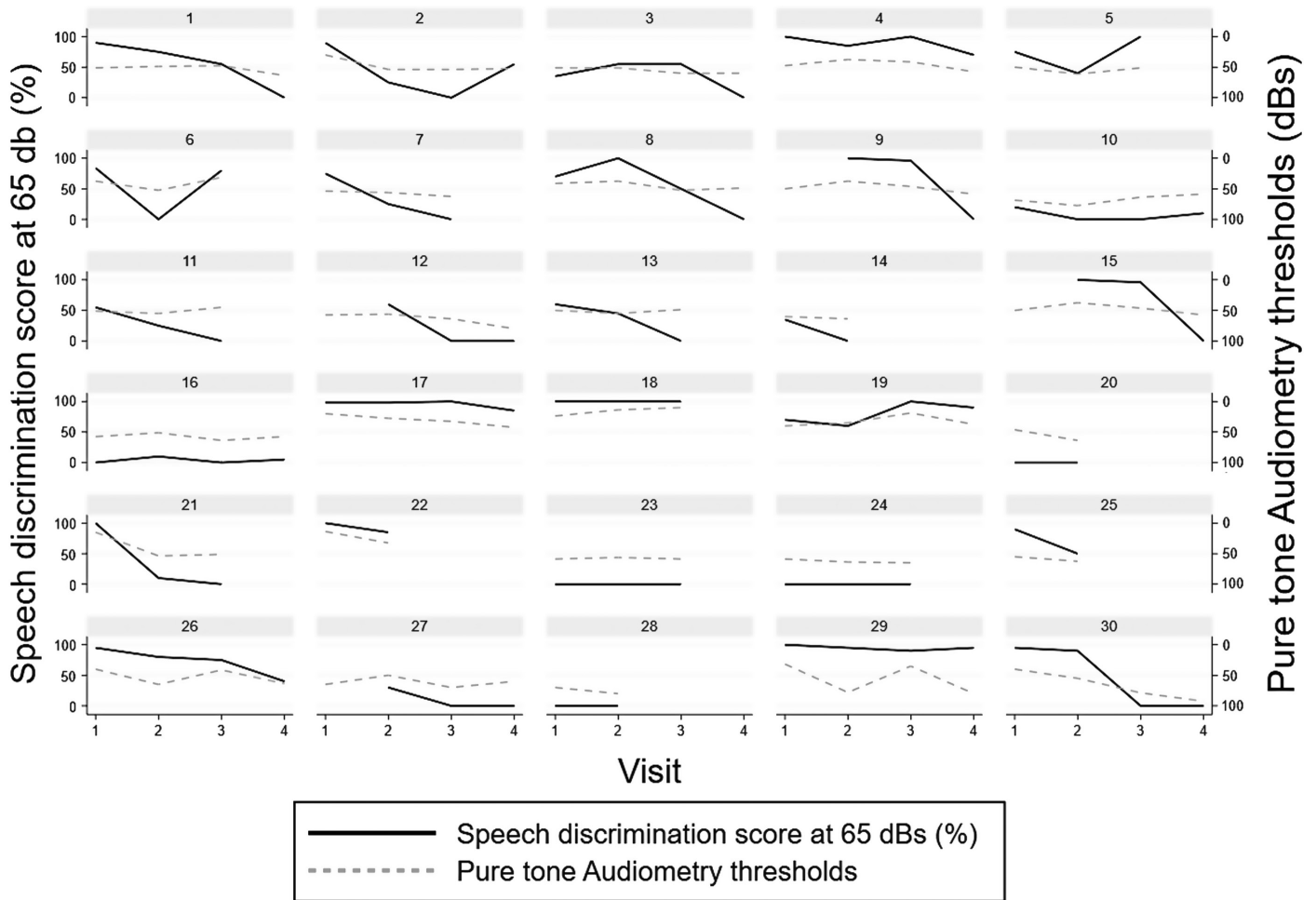


Figure 3. Pure-tone audiometry thresholds and speech discrimination score throughout the time of each patient in the Meniere's disease group.

## DISCUSSION

According to our follow-up findings in patients with MD, during its clinical progression, a moment comes when SRT decreases significantly compared to reduction in mean PTA. This occurs regardless of the age of the patient and most of the other clinical variables included, highlighting only the role of the duration of the disease on this audiological phenomenon.

If we consider MD as a peripheral lesion, the ability to understand speech can be the effect of loudness recruitment.<sup>14,15</sup> The exact mechanism of this perceptual phenomenon of sounds becoming rapidly louder with increasing sound level is still unknown. The most accepted theory is the loss of the compressive non-linearity of the basilar membrane (BM), which can be explained in MD by the presence of an endolymphatic hydrops (EH), and/or outer hair cells (OHC) damage.<sup>16-18</sup> Although in this study we did not assess the presence of loudness recruitment, some authors describe that this phenomenon is not always present in MD,<sup>15,19</sup> and also it may change from over-recruitment to under-recruitment.<sup>20</sup>

The dissociation between PTA and SR in MD during its clinical progression, in which recruitment can no longer be the sole cause, poses the question of a possible retrocochlear disorder. Even though Prosper Meniere first described this syndrome as an inner ear dysfunction and not a neurological disorder,<sup>21</sup> we now know that MD also affects

structures beyond the cochlear labyrinth. Histological studies have described a greater focal loss of neurons and a ganglion degeneration in the apical and upper middle turns of the cochlea.<sup>22-24</sup> It is possible that excitotoxicity (a form of neurotoxicity) occurs in the spiral ganglion (SG).<sup>25</sup> The hydropic condition of the cochlea would lead to an altered regional expression of glutamate aspartate transporter. The mechanism of cell death in the SG appears to be a progressive programmed cell death. This progressive apoptosis follows a topographic organization with cell loss at the apex preceding loss at the basal turn.<sup>26</sup>

Along with this SG condition, some reports previously described hair cell (HC) degeneration to be more severe in OHC.<sup>23</sup> Kimura<sup>27</sup> found that the stereocilia of OHC were lower in number and had fused together. Horner<sup>28</sup> described an atrophy of the short and middle stereocilia on OHC while the inner HC stereocilia did not have such alteration. The relevance of these findings to our work rests in the fact that OHC function is required for optimum SR.<sup>29</sup> Outer hair cells impairment can result in an amplification loss of around 40 dB, which might affect SR in MD.<sup>7</sup> Due to the fact that HL does not correlate with the reduction of the cell population in the SG<sup>23,24,30</sup> and that ganglion cell loss precedes the loss of HC,<sup>25,31,32</sup> we believe that both mechanisms, a probable ganglionopathy followed by an OHC damage, might explain this audiological phenomenon.

Fluctuating HL is typical and expected in MD. Hoa<sup>33</sup> described changes in low- and high-frequency thresholds and an evident fluctuation of more than 10 dB in 27% of patients with MD during audiometric follow-up. Although the underlying pathophysiological mechanism of this fluctuation is not yet fully understood, a mechanical model based on an EH that disturbs sound transmission as a result of the bulging of the BM is one of the most accepted theories.<sup>31,34</sup>

We have also found that SR in MD fluctuates, regardless of the PTAt, until eventually it starts to decrease. Johnson<sup>35</sup> observed the fluctuation pattern of speech results in a group of patients with MD. In their sample, 42% remained with the same SRT while 27% improved by 10 dB or more and 31% worsened by 10 dB or more. They concluded that speech results followed the same general fluctuating pattern of pure tone results, which coincides in a certain way with our results. In order to explain how SR fluctuates independently of PTA fluctuation, 2 different hypotheses could be proposed: a disturbance of wave transmission throughout the BM in the context of a fluctuant hydropic distension of the endolymphatic compartment or local potassium intoxication.<sup>28</sup> Apical EH, which is less severe than basal hydrops,<sup>24</sup> would be not enough to bulge the BM and affect PTA but may be sufficient to cause a K<sup>+</sup> intoxication. The leakage of potassium-rich endolymph into the perilymph via gaps of the reticular lamina or Reissner's membrane might paralyze the sensory HC affecting sound amplification and eventually SR.

We consider that awareness of the dissociation between PTA and SR and fluctuations in SR independently of PTA are essential for the follow-up. If we do not perform SR tests periodically in patients with MD, we might miss the presence of fluctuations in SR and wrongly assume that the patient's HL is stable. Furthermore, it is important to take this dissociation into account for adequate audiological care of patients with MD. Nowadays, fitting of hearing aids in MD patients is still a great challenge. This is mainly due to the fluctuating, unilateral, or asymmetrical HL, the re-activation of the disease whenever an intense sound pressure is maintained in the external ear,<sup>36</sup> and the poor SR scores,<sup>7,35</sup> which according to our results, may become progressively worse over time, regardless of the evolution of the PTA. Different strategies of treatment have been proposed<sup>37,38</sup>; however, when poor SR is present, the audiologist might run out of tools and a different approach may be needed. Cochlear implantation in MD patients has shown promising outcomes,<sup>39,40</sup> some studies have described substantial hearing benefits compared with hearing aids and a significant improvement in SR.<sup>41,42</sup> We believe that this audiological amelioration with cochlear implantation is consistent with a possible ganglionopathy and HC damage because the auditory nerve is being directly stimulated, bypassing the inner ear—an impossibility with hearing aids.

A controversial aspect of this study might be the assessment of SDS at 65 dB HL. This intensity corresponds to the level of normal conversation speech in the Spanish language at 1 m and also it is a comfortable level for listening to speech in quiet or low levels of background noise.<sup>12,13</sup> This intensity is also the level of speech required for 50% intelligibility in a flat moderate HL, and it's the average level of speech required for 50% intelligibility in flat severe HL and HL increasing with frequency.<sup>14</sup> The reason why we decided not to use the MSD or the word recognition test (35 dB above the SRT) is because even though those values are most commonly used in some clinics, they

do not reflect the discrimination at 65 dB, which is fundamental in the Spanish language. The main limitations of this study were: the follow-up time and the time when the periodic follow-ups were carried out was not the same for all the patients. Dietary changes, stress reduction techniques, and additional medical treatments were not included in the analysis. Hearing thresholds were analyzed as PTAt, and this average could mask the impact of the individual-frequencies information, especially low frequencies where there is room to fluctuate. Future studies in patients who speak other languages could reduce the limitations of the Spanish language assessment for non-Spanish speaking patients.

## CONCLUSION

Ménière's disease patients present a dissociation between PTA and SR during the evolution of the disease. This dissociation tends to increase along the time of follow-up and is statistically related to the duration of disease. In addition, patients with MD may present fluctuations in SR without a change in PTA during the follow-up. The presence of vertigo spells in the last 6 months is not related to changes in SDS at 65 dB, although it is related to changes in PTAt. The dissociation between PTA and SR might be explained by our hypothesis that introduces the concept of a ganglionopathy followed by disturbance of the OHC in the context of the hydrops and at the end, probably independent of it.

**Ethics Committee Approval:** Ethical committee approval was received from the ethical committee of the Clínica Universidad de Navarra (2020/047, 14/02/2020).

**Informed Consent:** Informed consent was obtained from all participants who participated in this study.

**Peer-review:** Externally peer-reviewed.

**Author Contributions:** Concept – N.P., O.G., R.M., L.G., D.C.; Design – N.P., O.G., R.M., L.G., D.C.; Supervision – N.P., R.M., L.G.; Resources – O.G., D.C.; Materials – O.G., R.M.; Data Collection and/or Processing – O.G., D.C.; Analysis and/or Interpretation – N.P., O.G., R.M., L.G., D.C.; Literature Search – N.P., O.G., R.M.; Writing Manuscript – O.G., N.P., R.M.; Critical Review N.P., O.G., R.M., L.G., D.C.

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