Idiopathic Sudden Sensorineural Hearing Loss: Speech Intelligibility Deficits Following Threshold Recovery

Masahiro Okada,1 Aravindakshan Parthasarathy,2,3 D. Bradley Welling,3,4 M. Charles Liberman,2,3,4 and Stéphane F. Maison2,3,4

OBJECTIVES: This retrospective study tests the hypothesis that patients who have recovered from idiopathic sudden sensorineural hearing loss (SSNHL) show deficits in word recognition tasks that cannot be entirely explained by a loss in audibility.

DESIGN: We reviewed the audiologic profile of 166 patients presenting with a unilateral SSNHL. Hearing loss severity, degree of threshold recovery, residual hearing loss, and word recognition performance were considered as outcome variables. Age, route of treatment, delay between SSNHL onset and treatment, and audiogram configuration were considered as predictor variables.

RESULTS: Severity, residual hearing loss, and recovery were highly variable across patients. While age and onset-treatment delay could not account for the severity, residual hearing loss and recovery in thresholds, configuration of the SSNHL and overall inner ear status as measured by thresholds on the contralateral ear were predictive of threshold recovery. Speech recognition performance was significantly poorer than predicted by the speech intelligibility curve derived from the patient's audiogram.

CONCLUSIONS: SSNHL is associated with (1) changes in thresholds that are consistent with ischemia and (2) speech intelligibility deficits that cannot be entirely explained by a change in hearing sensitivity.

KEY WORDS: Auditory nerve, Cochlea, Cochlear synaptopathy, Hearing in noise, Hidden hearing loss, Speech intelligibility, Sudden sensorineural hearing loss.

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INTRODUCTION

Idiopathic sudden sensorineural hearing loss (SSNHL) is characterized by a rapid loss of hearing (≤3 days), nearly always unilateral, which presents with varying levels of severity from mild to profound. SSNHL often occurs with other symptoms, including tinnitus, hyperacusis, and vertigo (Kuhn et al. 2011; Chung et al. 2015). With a low incidence of 5 to 30 cases per 100,000 patients per year (Schreiber et al. 2010), it is nonetheless one of the most common emergencies in otolaryngology practice (Marx et al. 2018). Partial or complete recovery in hearing may occur either spontaneously or following therapies including systemic and/or trans-tympanic corticosteroids to reduce inflammation (Wei et al. 2006; Conlin & Parnes 2007a,b) or vasoactive agents such as prostaglandins to enhance cochlear blood flow (Agarwal & Pothier 2009). With a high rate of spontaneous remission, ranging from ~30 to ~60% across studies, treatment efficacy is difficult to gauge (Mattoo 1980; Byl 1984; Huy & Sauvaget 2005; Nosrati-Zarenoe et al. 2007). The clinical guidelines of the American Academy of Otolaryngology—Head & Neck Surgery (AAO-HNS) conclude that systemic steroids are not proven as either effective or ineffective, and no recommendation has been made for or against their usage.

Sudden deafness is regarded as idiopathic despite the emergence of several pathophysiological hypotheses, including viral infection, intracochlear membrane rupture, vascular compromise, and activation of cellular stress pathways within the cochlea (Saunders & Lippy 1959; Schuknecht et al. 1962; Simmons 1968, 1979; Fisch et al. 1984; Harris 1984; Fetterman et al. 1996; Suckfüll et al. 2002; Merchant et al. 2005; Okada et al. 2017). An interruption of the vascular supply will impair oxygen delivery to the inner ear (Kim et al. 1999) causing ATP depletion (Thalmann et al. 1972) and compromised cochlear function. The metabolically active stria vascularis is a target of vascular compromise, as shown by the rapid decline of the endocochlear potential following ischemia onset (Bosher 1979; Konishi 1979; Morizane et al. 2005). The dendrites of cochlear afferent neurons are also sensitive to ischemia, with damage often preceding morphological changes in hair cells (Tabuchi et al. 2002). Cochlear ischemia leads to high levels of glutamate in the perilymph, presumably because supporting-cell uptake from the synaptic cleft is inadequate (Hakuba et al. 1997, 2000), resulting in damage to afferent dendrites (Puel et al. 1994).

While a decline in endocochlear potential, which directly affects the cochlear amplifier, results in threshold elevation, damage to the synapses between inner hair cells and primary afferent neurons will impact thresholds only if many of the low-threshold fibers, with high spontaneous rate (SR), are impacted. Aging, overexposure to noise, or ototoxic agents on the other hand appears to preferentially damage the cochlear neurons with high thresholds and low SRs (Schmiedt et al. 1996; Kujawa & Liberman 2009; Sergeyenko et al. 2013; Furman et al. 2013), which are key to the coding of transient stimuli in the presence of continuous background noise (Costalupes et al. 1984; Schmiedt et al. 1996; Furman et al. 2013). While synaptopathy will not elevate behavioral or electrophysiological thresholds, loss of high thresholds neurons could be a major contributor to difficulties in speech discrimination in noisy environments (Alvord 1983; Dubno et al. 1984; Rajan & Cainer 2008; Kujawa & Liberman 2015).

The present retrospective study aims to test the hypothesis that patients who have partially or fully recovered from idiopathic SSNHL show deficits in word recognition (WR) tasks that cannot be entirely explained by a loss in audibility and that are possibly consistent with cochlear synaptopathy.

MATERIALS AND METHODS

We collected audiological data from patients seen at Massachusetts Eye and Ear from 2007 to 2018 for otologic evaluation. We included patients, age 18 to 89, who underwent

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A comprehensive hearing evaluation and presented with an idiopathic unilateral SSNHL. All patients presenting a difference >10 dB between the mean threshold for air conduction (AC) and mean threshold for bone conduction (BC) (air-bone gap) were excluded. Audiometric records spanning less than a month were not considered. This study was reviewed and approved by the Institutional Review Board of Partners HealthCare.

Audiometric thresholds were obtained using a number of different audiometers including Grason-Stadler (GS-10, GS-16), Interacoustics AC-30, Virtual 320 and Interacoustics Equinox, running under the same Harvard Audiometer Operating System (Thornton et al. 1994). Pure-tone AC thresholds were measured at standard audiometric frequencies from 0.25 to 8 kHz, in octave steps using TDH39 headphones or ER-3A insert earphones. Bone conduction thresholds were acquired from 250 Hz to 4000 Hz with a Radioear B-71 vibrator over the mastoid. The pure-tone average (PTA) was defined as the average threshold at 500, 1000, and 2000 Hz. Hearing loss configurations were divided into three groups: (1) low-frequency (LF) dominated SSNHL where mean AC thresholds measured between 250 and 1000 Hz were on average 15 dB poorer than mean AC-thresholds measured between 2 and 8 kHz; (2) high-frequency (HF) dominated SSNHL where mean AC thresholds measured between 250 and 1000 Hz were on average 15 dB better than mean AC thresholds measured between 2 and 8 kHz; and (3) flat SSNHL when there were no more than 15 dB average difference between LF (250 Hz to 1 kHz) and HF (2 to 8 kHz) audiometric frequencies.

Speech recognition performance was assessed using a recorded Central Institute for the Deaf (CID) W-22 phonetically balanced test, consisting of 50 CNC word lists presented with a contralateral masker (speech-shaped noise). The Articulation Index (AI) was used to predict the speech intelligibility curve (SIC), a performance/intensity function for speech (Pavlovic et al. 1986; Wilde & Humes 1990) based on the audiogram, using a transfer function for CID W-22 (Sherbecoe & Studebaker 1990; Studebaker & Sherbecoe 1991). This procedure was automatically generated by the Harvard Audiometer Operating System software as described in Halpin et al. (1994). The level at which maximal intelligibility was predicted was chosen as presentation level. If this value, however, fell below 70 dB HL, presentation level remained at 70 dB HL. All WR scores were obtained from native speakers of English.

Statistical analyses included univariate linear regressions to determine relationship between independent and dependent variables, adding age and sex as possible confounders. The Mann–Whitney U test was performed to examine the significance of the differences across groups. Finally, the relationship between WR score and predicted WR score based on the SIC was tested using a Wilcoxon signed-rank test. The threshold for statistical significance was p = 0.05.

RESULTS

One hundred sixty-six patients (91 male, 75 female) presenting with a unilateral SSNHL met our inclusion criteria. The average age was 59.2 ± 1.1 years, and the observation span ranged from 1 month to ~9 years between the first and last hearing evaluation. As shown in Figure 1A, (1) the severity of the hearing loss was defined as the interaural difference in AC thresholds at the first visit; (2) the recovery was defined as the difference in ipsilateral thresholds between the first and last visit; and (3) the residual hearing loss was defined as the interaural threshold difference at the last visit. The contralateral side was chosen as the reference, as no baseline before SSNHL onset was available and because contralateral thresholds were stable between the first and last visits (Z = −0.12, p = 0.905).

Overall, the severity of the SSNHL was similar across test frequencies (~30 dB; Fig. 1A), and the magnitude of the interaural threshold difference was significant (Z = 16.70, p < 0.001). On the last visit, there was a significant ~15-dB mean residual hearing loss remaining (Z = 9.49, p < 0.001) and a ~15-dB mean...

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**Fig. 1.** Profile of cohort study. A–C, Mean hearing sensitivity as assessed via air conduction (AC) thresholds. Arrows in (A) define the severity of the condition (1) as the interaural difference in AC thresholds measured at the initial visit; the recovery (2) as the ipsilateral difference in AC thresholds between the initial and final visits; and the residual hearing loss (3) as the interaural difference in AC thresholds measured at the final visit. B, Box and whiskers plots of individual AC thresholds C, Mean word recognition scores obtained on the CID W-22 phonetically balanced test. Error bars in (A and C) are for SEMs. Key in (B) applies to all plots. Statistical significance is indicated (***p < 0.001).
threshold recovery over the audiometric frequency range ($Z = 6.40, p < 0.001$). As shown in Figure 1B, individual threshold elevations were highly variable across subjects.

While mean WR scores remained stable on the contralateral side between the first and last visits ($Z = -0.45, p = 0.653$), they were greatly reduced on the ipsilateral side (Fig. 1C). This asymmetry in WR performance was significant: $59.0 \pm 2.7\%$ versus $95.4 \pm 0.6\%$ of words correctly repeated on the first visit ($Z = -13.07, p < 0.001$). Despite a substantial improvement in ipsilateral word scores measured at the last visit ($78.2 \pm 2.2\%$, $Z = -5.52, p < 0.001$), scores remained significantly poorer than the contralateral side ($95.8 \pm 0.5\%, Z = -7.88, p < 0.001$).

Of the 94% of patients receiving corticosteroid treatment (Table 1), ~58% received oral prednisone (1 mg/kg/day for 2 to 3 weeks followed by a taper) and ~40% also received trans-tympanic injections (TTIs) of dexamethasone (~3 to 5 mg per injection). A few patients received only the TTI of dexamethasone ($n = 4$). The severity of the condition was not associated with age or sex and, except for the four patients who only received TTI of steroids, the combination of oral and TTI steroids was chosen for the most severe cases (Fig. 2A; stepwise regression: $F = 5.20, p = 0.002$ with choice of treatment, $p < 0.001$; age, $p = 0.164$; sex, $p = 0.570$). On the other hand, neither the residual hearing loss nor the recovery were statistically associated with interaural difference in AC thresholds, $p = 0.269$ and residual hearing loss (stepwise regression, $F = 24.77, p < 0.001$ with interaural difference in AC thresholds, $p < 0.001$; age, $p = 0.917$ and sex, $p = 0.837$) independently from the age or sex of patients.

Likewise, using stepwise regression including age and sex as possible confounders, we found no statistically significant effect of treatment delay on severity, residual hearing loss or recovery (Fig. 3; severity: $F = 1.39, p = 0.249$; residual: $F = 1.40, p = 0.246$; recovery: $F = 0.68, p = 0.565$). In contrast, using the same statistical approach, the degree of recovery from SSNHL was associated with its audiometric pattern (stepwise regression, $F = 3.45, p = 0.018$ with configuration of hearing loss, $p = 0.002$; age, $p = 0.509$; sex, $p = 0.972$). On average, ~54% of the threshold shift recovered in patients with LF SSNHL (Fig. 4A), while only ~28% recovered from HF SSNHL (Fig. 4C). Those with a flat SSNHL saw a ~49% improvement (Fig. 4B).

Interestingly, these SSNHL audiometric configurations had different patterns of contralateral hearing loss: patients with HF SSNHL had near-normal thresholds contralaterally and were younger than those with an LF SSNHL who presented downward sloping moderate SNHL contralaterally (Fig. 4A–C). Remarkably, the interaural difference in AC thresholds measured from 2 to 8 kHz was predictive of both recovery (stepwise regression, $F = 9.88, p < 0.001$ with interaural difference in AC thresholds, $p < 0.001$; age, $p = 0.081$; and sex, $p = 0.269$) and residual hearing loss (stepwise regression, $F = 24.77, p < 0.001$ with interaural difference in AC thresholds, $p < 0.001$; age, $p = 0.917$ and sex, $p = 0.837$) independently from the age or sex of patients. Likewise, severity was associated with this interaural difference in the AC threshold and predictively, and age was a significant confounding variable (stepwise regression, $F = 158.29, p < 0.001$ with interaural difference in AC thresholds, $p < 0.001$; age, $p = 0.002$ and sex, $p = 0.024$). Delay between the SSNHL onset and treatment was similar across groups (Table 2).

### Table 1. Study population groups (mean ± SEM): effect of corticosteroid treatments

<table>
<thead>
<tr>
<th></th>
<th>No Steroid</th>
<th>Oral</th>
<th>TTI</th>
<th>Oral + TTI</th>
</tr>
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<tbody>
<tr>
<td>n</td>
<td>11</td>
<td>90</td>
<td>4</td>
<td>62</td>
</tr>
<tr>
<td>Age (yrs)</td>
<td>62.1 ± 3.8</td>
<td>58.2 ± 1.6</td>
<td>65.0 ± 7.9</td>
<td>59.7 ± 1.8</td>
</tr>
<tr>
<td>Sex (male vs. female)</td>
<td>6–5</td>
<td>51–39</td>
<td>0–4</td>
<td>34–27</td>
</tr>
<tr>
<td>Observation span (mo)</td>
<td>6.8 ± 1.6</td>
<td>14.4 ± 2.6</td>
<td>4.7 ± 1.6</td>
<td>9.9 ± 1.5</td>
</tr>
<tr>
<td>Onset—therapy delay (d)</td>
<td>N/A</td>
<td>12.3 ± 1.0</td>
<td>12.0 ± 6.2</td>
<td>11.0 ± 0.9</td>
</tr>
<tr>
<td>Severity (dB)</td>
<td>19.0 ± 3.2</td>
<td>27.3 ± 1.9</td>
<td>36.0 ± 8.0</td>
<td>36.9 ± 2.7</td>
</tr>
<tr>
<td>Recovery (dB)</td>
<td>3.3 ± 2.9</td>
<td>13.3 ± 2.1</td>
<td>14.0 ± 3.6</td>
<td>16.4 ± 2.3</td>
</tr>
<tr>
<td>Residual loss (dB)</td>
<td>15.7 ± 3.4</td>
<td>13.5 ± 1.8</td>
<td>23.3 ± 11.8</td>
<td>20.1 ± 2.4</td>
</tr>
</tbody>
</table>

TTI indicates trans-tympanic injection.
WR scores for these three groups followed hearing thresholds as predicted by the audibility index (Fig. 4D–F); that is, losses near 2 kHz had the most impact on speech intelligibility. While a majority of word scores fell within the range predicted by the SIC, many patients scored in the poor or very poor range (<64% of words correctly repeated; Fig. 5A). These SICs are derived from the articulation index for CID W-22 word lists (see Materials and Methods), which predicts word scores as a function of presentation level based on simply filtering the speech according to the audiometric losses. Deviation between measured and predicted is indicative of a deficit in speech intelligibility. When this deviation is normalized to the contra-lateral side (Fig. 5C), the mismatch was as large as ~70 dB and increased with PTA (Wilcoxon signed-rank test of measured vs. predicted: ipsilateral, \( V = -5.15, p < 0.001 \); contralateral: \( V = -0.49, p = 0.624 \)). These results support the idea that intelligibility deficits in SSNHL cannot be entirely explained by a deficit in audibility.

We then examined the word scores in patients whose thresholds presumably returned to preonset levels; that is, losses near 2 kHz had the most impact on speech intelligibility. While a majority of word scores fell within the range predicted by the SIC, many patients scored in the poor or very poor range (<64% of words correctly repeated; Fig. 5A). These SICs are derived from the articulation index for CID W-22 word lists (see Materials and Methods), which predicts word scores as a function of presentation level based on simply filtering the speech according to the audiometric losses. Deviation between measured and predicted is indicative of a deficit in speech intelligibility. When this deviation is normalized to the contra-lateral side (Fig. 5C), the mismatch was as large as ~70 dB and increased with PTA (Wilcoxon signed-rank test of measured vs. predicted: ipsilateral, \( V = -5.15, p < 0.001 \); contralateral: \( V = -0.49, p = 0.624 \)). These results support the idea that intelligibility deficits in SSNHL cannot be entirely explained by a deficit in audibility.

We examined the word scores in patients whose thresholds presumably returned to preonset levels; that is, with no significant residual hearing loss (Table 3). The chart in Figure 6 details how patients were selected. One hundred eight out of one hundred sixty-six patients remained with an interaural asymmetry in hearing thresholds (>5 dB) at their last visit. Among the 58 patients with symmetrical hearing at the last visit, 50 patients completely recovered from a significant interaural asymmetry in hearing threshold and word score by the last visit (Fig. 7A; interaural word score difference at the first visit: \( Z = -7.27, p < 0.001 \) vs. final visit: \( Z = 0.20, p = 0.843 \)); and eight patients had a significant asymmetry in word scores at the last visit despite having hearing thresholds returning to pre-onset levels (Fig. 7B; interaural word score difference at the first visit: \( Z = -2.41, p = 0.016 \) vs. final visit: \( Z = -4.23, p < 0.001 \)). The latter observations further support the existence of speech intelligibility deficits unrelated to hearing sensitivity.

**DISCUSSION**

**Steroid Treatment Efficacy**

Within the therapeutic armamentarium, corticosteroids are a favored option for a wide variety of indications (Marx et al. 2018). In the absence of clear evidence for benefit, the use of systemic corticosteroids as a first-line of treatment for SSNHL arises because the side effects are typically mild (Alexander et al. 2009; Rauch et al. 2011), yet hearing loss can significantly impact quality of life, with degraded speech recognition,
Fig. 4. Hearing loss configuration impact on recovery from SSNHL. Audiometric thresholds, speech intelligibility curves and word recognition scores are plotted as a function of the SSNHL configuration. Hearing loss configurations were divided into three groups: (A) low-frequency (LF) hearing loss when mean AC thresholds from 250 to 1000 Hz were 15 dB poorer than those from 2 to 8 kHz; (C) high-frequency (HF) hearing loss when mean AC thresholds from 250 to 1000 Hz were 15 dB better than those from 2 to 8 kHz; and (B) flat hearing loss when there was no more than 15 dB difference between LF (250 to 1000 Hz) and HF (2 to 8 kHz). Mean delay between reported SSNHL onset and treatment (D) and mean group age (A) are indicated on each audiogram. Keys in (A) apply to all plots. Error bars in (A–C) are for SEMs. Error bars in (D–F) indicate the 95% confidence interval. SSNHL indicates sudden sensorineural hearing loss.
particularly in noisy environments and impaired sound localization ability (Mattox & Lyles 1989; Huy & Sauvaget 2005). TTI of corticosteroids can also be a primary therapy (Rauch et al. 2011), as higher concentrations of corticosteroids reach the inner ear and systemic effects are avoided (Chandrasekhar 2001; Bird et al. 2007).

TTI of corticosteroids appears to be associated with greater chances of hearing improvement than no therapy or placebo (Plontke 2017). As observed here (Table 1), TTI is often combined with oral therapy (Dispensa et al. 2011), particularly for the most severe hearing losses (Plontke et al. 2009) or after failure or insufficient recovery using systemic therapies (Plontke 2017). It is not clear if TTI produces better outcomes than systemic administration: two meta-analyses showed no significant differences between systemic and trans-tympanic steroids, as a first treatment (Crane et al. 2015; El Sabbagh et al. 2017). On the other hand, a third meta-analysis found better recovery for TTI in patients with mild to moderate hearing loss (Qiang et al. 2017). While the association between choice of treatment and residual hearing loss or recovery failed to reach the statistical significance, our retrospective study does not allow any strong conclusions re the efficacy of corticosteroids as (1) we cannot determine the extent to which steroids accessed the inner ear fluids when applied to the middle ear space and (2) the high rate of spontaneous recovery, estimated to be 1/3 to 2/3 of patients with SSNHL (Mattox & Simmons 1977; Byl 1984; Nosrati-Zarenoe et al. 2007), further complicates the interpretation of treatment efficacy. Finally, our study lacks an equal-size control group as only ~7% of our patients (n = 11) did not receive any kind of treatment.

Age was correlated with residual hearing loss (Fig. 3), with older patients showing less residual hearing: likely a ceiling effect, as older patients start with less to lose at SSNHL onset. When considered as a possible confounding variable in linear regression analyses, age along with sex did not contribute to residual hearing loss or recovery from SSNHL. This result echoes the findings of a nationwide survey in Japan (Okada et al. 2017). As for the effects of treatment delay, there is again no consensus due to the high rate of spontaneous recovery: while some studies argue that each additional day following SSNHL onset will negatively impact recovery (Attanasio et al. 2018), others estimate that excellent outcomes are still achieved for delays of up to 2 weeks (Mattox & Simmons 1977; Edizer et al. 2015; Amarillo et al. 2019) as observed in the present study (Fig. 3F).

**TABLE 2. Study population groups (mean ± SEM): effect of hearing loss configuration**

<table>
<thead>
<tr>
<th></th>
<th>Low Frequency</th>
<th>Flat</th>
<th>High Frequency</th>
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</thead>
<tbody>
<tr>
<td>N</td>
<td>47</td>
<td>71</td>
<td>48</td>
</tr>
<tr>
<td>Age (yrs)</td>
<td>64.1 ± 2.2</td>
<td>60.7 ± 1.5</td>
<td>52.3 ± 2.1</td>
</tr>
<tr>
<td>Sex (male vs. female)</td>
<td>34–13</td>
<td>32–39</td>
<td>25–23</td>
</tr>
<tr>
<td>Observation span (mo)</td>
<td>12.7 ± 3.0</td>
<td>14.1 ± 2.9</td>
<td>8.4 ± 1.4</td>
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<tr>
<td>Onset—therapy delay (d)</td>
<td>10.1 ± 1.4</td>
<td>13.1 ± 1.1</td>
<td>11.5 ± 1.1</td>
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<tr>
<td>Severity (dB)</td>
<td>30.0 ± 2.5</td>
<td>33.6 ± 2.7</td>
<td>26.3 ± 2.0</td>
</tr>
<tr>
<td>Recovery (dB)</td>
<td>16.2 ± 2.7</td>
<td>16.6 ± 2.4</td>
<td>7.3 ± 1.9</td>
</tr>
<tr>
<td>Residual loss (dB)</td>
<td>13.6 ± 2.0</td>
<td>16.8 ± 2.5</td>
<td>19.0 ± 2.4</td>
</tr>
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</table>

**Fig. 5.** Patients with SSNHL show speech intelligibility deficits unrelated to threshold elevation. Speech intelligibility (SI) curves were derived from each audiogram obtained on the SSNHL side (A) or contralateral to that ear (B). Differences between measured and predicted speech recognition scores from the same patients were computed and normalized to the differences between measured and predicted scores on the contralateral ear (C). Key in (B) applies to all plots. Error bars indicate the 95% confidence interval. SSNHL indicates sudden sensorineural hearing loss.
Hearing Loss Configuration and Etiology

As seen in prior reports (Mattox & Simmons 1977; Zadeh et al. 2003), the frequency pattern of SSNHL is a strong predictor of outcome. For LF SSNHL, more than half of the initial threshold shift recovered by the last visit, compared with less than a third in patients with an HF loss (Fig. 4). Interestingly, while these groups had similar onset-treatment delays, age and audiogram configurations on the contralateral side were different. Patients with the LF SSNHL pattern were older than those with HF SSNHL and showed classic high-tone presbyacusis on the contralateral side (Fig. 4). The presumed pre-existence of a high-tone loss would limit the additional shift from SSNHL, such that the transient pathology of the “LF” and “Flat” groups might actually be quite similar in the apical regions.

There is no consensus on the etiology of SSNHL, but hypotheses include viral infection, intracochlear membrane rupture, vascular compromise, and activation of cellular stress pathways within the cochlea (Saunders & Lippy 1959; Schuknecht et al. 1962; Simmons 1968; Fisch et al. 1984; Harris 1984; Fetterman et al. 1996; Suckfüll et al. 2002; Mierzwa et al. 2004; Merchant et al. 2005; Lee et al. 2015). Several studies examined the histopathological correlates of irreversible SSNHL (e.g., Schuknecht et al. 1962, 1973; Schuknecht & Donovan 1986; Merchant et al. 2005; 2008). There was no histological evidence of membrane breaks; however, most ears showed damage similar to that observed in viral infections (e.g., mumps, rubella; Merchant & Nadol 2010), including atrophy of the organ of Corti and stria vascularis in the basal turn and loss of auditory nerve fibers (Bordley & Kapur 1977; Gussen 1981; Davis & Johnsson 1983). The basal turn focus of these permanent lesions is consistent with the HF bias of the irreversible cases of SSNHL. Further support for the viral hypothesis include the observations that ~50% of these cases had SSNHL onset during an upper respiratory infection (Merchant & Nadol 2010) and that other studies found evidence of viral infection in serologic samples from SSNHL patients (van Dishoeck &

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TABLE 3. Study population groups (mean ± SEM) with threshold recovery

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<th>Thr. and WRS Recovery</th>
<th>Thr. Recovery Only</th>
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<tr>
<td>N</td>
<td>50</td>
<td>8</td>
</tr>
<tr>
<td>Age (yrs)</td>
<td>62.2 ± 2.0</td>
<td>60.5 ± 3.8</td>
</tr>
<tr>
<td>Sex (male vs. female)</td>
<td>29–21</td>
<td>4–4</td>
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<tr>
<td>Observation span (mo)</td>
<td>17.0 ± 3.3</td>
<td>32.5 ± 16.2</td>
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<tr>
<td>Onset—therapy delay (d)</td>
<td>12.0 ± 1.2</td>
<td>10.3 ± 4.0</td>
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<tr>
<td>Severity (dB)</td>
<td>28.2 ± 2.4</td>
<td>10.7 ± 5.3</td>
</tr>
<tr>
<td>Recovery (dB)</td>
<td>26.9 ± 2.5</td>
<td>7.5 ± 5.2</td>
</tr>
<tr>
<td>Residual loss (dB)</td>
<td>1.2 ± 0.4</td>
<td>-1.0 ± 1.5</td>
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WRS indicates word recognition score.

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Fig. 6. Flowchart of the decision tree for diagnosis group classification. IA indicates interaural; SSNHL, sudden sensorineural hearing loss; WRS, word recognition score.

The LF hearing loss in SSNHL could be due to vascular compromise. Axelsson studies of cochlear arterial supply in human show a prominent branch point where the common cochlear artery splits into a vestibulo-cochlear artery that supplies the whole cochlea, and a spiral modiolar artery that supplies only the apical half (Axelsson 1988). Given such an architecture, an upstream occlusion might affect all frequencies, whereas a more downstream occlusion might affect predominantly LFs. Animal experiments in which the cochlear arterial supply was either permanently compromised or transiently occluded often found hair
cell lesions preferentially in the middle or apical turns (Schuknecht & Bernstein 1967). Cochlear ischemia tends to impact the inner hair cells more dramatically than outer hair cells (Hakaba et al. 2003), possibly due to the interactions with dendritic damage to auditory nerve fibers elicited by glutamate excitotoxicity.

Impact on Speech Intelligibility

This study shows that, after recovery from an idiopathic SSNHL, patients have poorer WR scores than predicted by the residual loss of audibility. Because word scores in this study were obtained in quiet, the real-world handicap of affected patients is likely underestimated.

As shown in Figures 4 and 7, there were minimal mismatches between predicted and measured word scores for the ear contralateral to the SSNHL, suggesting that cognitive factors, such as deficits in attention, working memory or linguistic abilities, cannot be the cause of speech intelligibility deficits for the ear recovered from SSNHL. A lack of sensory input has profound effects on neuronal organization and sensory maps of the central auditory system during developmental critical periods but also in the mature brain, particularly following hearing loss (for reviews, see Persie et al. 2020; Pienkowski 2017). For example, acoustic overexposures causing mild to moderate HF hearing loss lead to profound reorganization of the cortical tonotopic map in cats (Eggermont & Komiya 2000). Likewise, even exposures that do not cause a permanent change in thresholds can alter the neural activity in the auditory cortex (Pienkowski & Eggermont 2012; Pienkowski 2017). These lines of research suggest that a permanent threshold elevation is not a prerequisite for reorganization of the central auditory pathways, and therefore, speech recognition deficits, in the absence or presence of permanent threshold elevation, may arise from a reorganization of the central auditory system following a cochlear insult.

We were inspired to look for speech intelligibility deficits that could not be explained by changes in hearing sensitivity in patients with SSNHL, because recent animal research showed peripheral plasticity and cochlear synaptopathy in adult mice following a prolonged unilateral hearing loss (Liberman et al. 2015). Indeed, similar deficits in intelligibility were observed in patients presenting with a chronic conductive hearing loss and normal bone conduction thresholds (Okada et al. 2020). While we cannot provide definitive evidence of cochlear synaptopathy in these patients, or in the recovered patients from the present study, poor word scores have been noted in prior human histopathological studies of cases with high levels of cochlear neuronal degeneration (Felder & Schrott-Fischer 1995). A loss of speech discrimination ability without significant audiometric shift is consistent with loss of inner hair cells or auditory nerve fibers, because loss of these elements must be >80% before it begins to affect threshold sensitivity (Lobarinas et al. 2013), whereas this degree of loss should significantly affect speech discrimination. Loss of cochlear neurons is one of the most common histopathological features of SSNHL in human temporal bones (Merchant & Nadol 2010). Both viral infection and ischemia can lead to the permanent damage of afferent neurons (Lindsay 1973; Linthicum 1978; Sawada 1979; Puel et al. 1994) and chronic strial dysfunction leads to hypnotrophy of auditory nerve cell bodies and peripheral axons (Suryadevara et al. 2001).

An additional contributor to this residual impairment could be hair cell damage in the cochlear apex, which constitutes another form of “hidden hearing loss.” A recent temporal bone study revealed that the apical cochlea is a major focus of inner and outer hair cell loss in the aging ear. This loss is undetected by standard audiometry (Wu et al. 2020), because the apical 20% of the human cochlea is tuned to frequencies below 250 Hz (Greenwood 1990). In patients with SSNHL, the LF region is clearly a major focus of damage. An incomplete recovery (or eventual death) of inner hair cells in this region could silence large numbers of auditory nerve fibers that normally carry information rich in temporal fine-structure cues. Their loss from the ascending neural signal could also contribute to the WR deficits in these patients.

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