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Main Article

Idiopathic sudden sensorineural hearing loss: prognostic factors

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Abstract

Objectives: Sudden sensorineural hearing loss (SSHL) remains a challenge for the clinician. In the majority of cases, no definite cause can be found and the prognosis is variable.

Methods: The present study assessed 114 patients suffering from idiopathic SSHL, with regard to the prognostic value of demographic, epidemiologic, neurotologic and audiometric factors. In addition, the relationship between the identification of wave V in auditory brainstem responses and the final hearing outcome was investigated. All patients received 75 mg/day intravenous prednisolone, divided into three daily doses, for 10 days, with gradual tapering of the dose over the next 10 days.

Results: The results (after one year follow up) revealed the following factors to be related to a better hearing outcome: younger age; male sex; less time elapsed between the onset of hearing loss and the beginning of treatment; and an upward-sloping or cupeloid audiogram contour. The detection of wave V early in recovery and within the first month of medical treatment might also constitute a significant favourable factor in respect to hearing recovery.

Conclusions: The present study revealed that there are certain factors that affect prognosis in idiopathic SSHL. This is very important in counselling patients and may affect current clinical practice.

Key words: Hearing Loss, Sensorineural; Hearing Loss, Sudden; Audiometry, Evoked Response; Prognosis

Introduction

Sudden sensorineural hearing loss (SSHL) remains a challenge for the clinician, despite the great advances in successful treatment of profound deafness made possible by cochlear implants. In the majority of cases, no definite cause can be found, the prognosis regarding final hearing outcome is quite variable, and treatment is mainly based on intravenous and/or intratympanic corticosteroid administration.[1]

The annual incidence of SSHL is estimated to be one case per 5000–10 000 population;[2,3] this number reflects patients seeking medical advice – the number of cases experiencing spontaneous recovery remain unknown. A minority of SSHL cases (10–15 per cent) are due to infectious, traumatic, neoplastic, immunologic, toxic, circulatory and neurologic causes.[4] However, the majority of cases (85–90 per cent) are characterized as 'idiopathic'; proposed aetiologic mechanisms include viral infection, vascular compromise, intracochlear membrane rupture and autoimmune inner-ear disease.[4,5]

Several studies have reported the prognostic value of such factors as age, tinnitus, electronystagmographic (ENG) findings, hearing level, shape of audiogram and time of initiation of treatment in predicting the final hearing outcome.[2,6–15]

Although brainstem evoked response (BSER) is a widely used electrophysiological method that assesses the functional integrity of the auditory system and may identify retrocochlear pathology,[16,17] its prognostic value in SSHL has not yet been studied.

The aim of the present study was to assess the prognostic value of demographic, epidemiologic, neurotologic and audiometric factors in predicting hearing recovery in idiopathic SSHL. The relationship between the identification of wave V in BSER audiometry and the final hearing outcome was also investigated.

Methods

Our research protocol received ethical approval from the scientific and ethical committee of the Hippokration General Hospital, Athens, Greece.

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Subjects

Patients presenting with idiopathic SSHL within 30 days from the onset of their symptoms and who were 15 years of age or older were included in this study. SSHL was defined as sensorineural hearing loss of at least 30 dB in three contiguous frequencies. The time elapsed between the onset of symptoms and the establishment of the actual hearing loss as perceived by the patient was no more than three days.[4,11] Any progressive hearing loss with a process lasting more than three days, was excluded.

A detailed history, a thorough head and neck clinical examination, laboratory examinations, and magnetic resonance imaging (MRI) of the cerebellopontine angle and the internal auditory canal with gadolinium enhancement were obtained for all individuals in order to exclude cases that could be related to infectious, traumatic, neoplastic, immunologic, toxic, circulatory, neurologic or metabolic causes.

Consequently, from the initial 124 patients, 10 (8 per cent) were excluded, comprising: two patients suffering from Ménière's disease; three patients in whom the MRI revealed the presence of an acoustic neuroma on the side of the affected ear; one patient with bilateral SSHL due to bacterial meningitis; one patient suffering from multiple sclerosis; one patient with a fluorescent treponemal antibody (FTA) positive serologic test; one patient with a known history of systemic lupus erythematosus; and one patient in whom Pierre-Marie syndrome was diagnosed. The rest of the cases were considered as idiopathic and were included in our study.

These cases comprised a total of 114 patients (62 (54.4 per cent) men and 52 (45.6 per cent) women). These patients' ages ranged from 15 to 79 years, with a mean age at time of presentation of 45.1 years (standard deviation (SD) = 15.2 years). Only one case (0.9 per cent) had bilateral involvement. The right ear was involved in 63 (55.3 per cent) patients and the left in 50 (43.8 per cent).

In the affected ear, pure tone average thresholds (PTAs) for the frequencies 500 Hz and 1, 2 and 4 KHz ranged from 30 to 105 dB at presentation (mean = 72.1 dB, SD = 23.0 dB). In the contralateral ear, PTAs for the frequencies 500 Hz and 1, 2 and 4 KHz ranged from 10 to 105 dB at presentation (mean = 23.1 dB, SD = 22.5 dB).

Patients presented to the investigators within a mean of 5.7 days (SD = 5.9 days) and a median of four days following onset of hearing loss, with a range of one to 30 days. Presentation was within a day in 23 patients (20.2 per cent) and within 10 days in 86.8 per cent of patients.

Treatment

All patients received 75 mg/day intravenous prednisolone, divided into three daily doses, for 10 days, with gradual tapering of the dose over the next 10 days. During steroid treatment, ranitidine was administered to all patients.

Procedures

At the time of presentation and before initiation of treatment, all individuals underwent audiometric testing.

Of the 114 patients, 78 (68 per cent) underwent BSER evaluation. The maximum click level was 121 dB peak equivalent sound pressure level, using an NBS type 9-A coupler (83 dB hearing loss) (*Bio-logic Systems Corp., Mundelein, Illinois, USA*). For the purposes of the study, we considered only the presence or absence of the waves I, III and V and not their latencies.

Most patients (85/114) also received ENG testing, including Hallpike caloric testing to measure lateral semicircular canal function (240 ml of cool (30°C) water over 40 seconds and 240 ml of warm (44°C) water over 40 seconds). During the caloric irrigation tests, a difference between the vestibular response of the two sides equal to or greater than 25 per cent was considered as hypoaesthesia of the lesser responding labyrinth.

Patients underwent treatment and close observation for 15 days. During this period, they underwent an audiogram and BSER testing every second day. Follow-up examination was performed one, three, six and 12 months following hospital discharge and included an audiogram and BSER.

Patients' symptoms of tinnitus and vertigo were recorded and a detailed history was taken with regard to possible contributing factors such as: recent viral infections, diving, noise exposure, hypertension, hypercholesterolaemia, diabetes mellitus, vasculitis, hypothyroidism, use of medications (including ototoxic drugs), history of previous hearing loss or chronic otitis media, neurologic diseases, autoimmune disorders, and emotional stress. Laboratory examinations included tests for: full blood count; erythrocyte sedimentation rate; prothrombin time; serum glucose, cholesterol, lipids, urea and creatinine concentrations; viral serologic for Epstein-Barr, herpes simplex type I and II and cytomegalovirus viruses; FTA test for *Treponema palladium* infection; serum thyroid hormones; and antigen-nonspecific serologic tests for autoimmune diseases [antinuclear antigen (ANA), anti-smooth muscle antibody (ASMA), anti-deoxyribonucleic acid antibody (anti-DNA), and rheumatoid factor (RF)].

The audiograms of the affected ears were classified as 'deaf', 'ascending', 'descending', 'flat' or 'cupeloid'. An audiogram was characterized as ascending when lower frequencies (250 and 500 Hz) were affected more than higher frequencies; descending when higher frequencies (4 and 8 KHz) were affected more than lower frequencies; cupeloid when the middle frequencies (1 and 2 KHz) were mostly affected; and flat when all frequencies were equally affected. When the hearing level was worse than 85 dB in all frequencies, the ear was characterized as deaf.

Hearing outcome measures included the final PTA, the difference between the initial and final PTAs, and the response to treatment, categorized

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as 'complete response' (CR), 'partial response' (PR), or 'no response' (NR). A CR was defined as a final PTA of ≤25 dB or a final PTA within 10 dB of that of the contralateral ear; a PR was defined as an improvement of more than 10 dB in three consecutive frequencies which did not reach the criteria for CR; and anything less than a PR, or deterioration of hearing level, was classified as NR.

Statistical analysis

Both parametric and non-parametric inferential statistics were performed. The non-parametric Spearman's Rho correlation was computed for correlation analyses, as some of the key variables were ordinal and others had limited values. For these analyses, patients in whom a BSER wave could not be indentified were arbitrarily assigned a high value (99) to rank them at the upper extreme. Response categories were assigned values of 0 (NR), one (PR) and two (CR), with a higher score representing a better response. Chi-square was used for examining relationships between categorical and/or ordinal variables. For analysis of mean change in PTA by groups, one-way analysis of variance was performed, using the Bonferroni method for post hoc multiple comparisons. Finally, a discriminate analysis was performed to determine which variables, if any, were most predictive of response category. The criterion for statistical significance was set at $p \leq 0.05$, two-tailed.

Results

Table I summarizes the audiologic characteristics of the patients at presentation. Nearly all subjects (91 per cent) reported tinnitus. More than half (57.6 per cent) had reduced vestibular response on ENG. The shape of the audiogram was quite varied, with nearly equal proportions of deaf (30 patients), ascending (29 patients) and descending (26 patients) patterns. Patients reported a recent history of viral illness in 26.9 per cent of cases and a recent history of hypertension, high cholesterol or stress in 24.4 per cent of cases.

TABLE I

AUDIOLOGICAL CHARACTERISTICS*

Characteristic	Incidence (%)
Vertigo	40.9
Tinnitus	90.9
ENG reduced	57.1
Audiogram contour	
Deaf	26.3
Ascending	25.4
Descending	22.8
Flat	13.2
Cupeloid	12.3
Predisposing factors	
Recent virus infection	26.9
Hypertension/cholesterol/stress	24.4

*For 114 sudden sensorineural hearing loss (SSHL) patients. ENG = electronystagmography

TABLE II

HEARING OUTCOMES BY RESPONSE CATEGORY*

Response	*n*	%	Final PTA (dB) (Mean (SD))	Improvement (dB) (Mean (SD))
CR	43	37.7	18.0 (7.9)	40.1 (16.9)
PR	34	29.8	49.9 (14.9)	29.7 (12.0)
NR	37	32.5	80.3 (25.0)	1.1 (2.9)

*For 114 sudden sensorineural hearing loss (SSHL) patients. PTA = pure tone average threshold; SD = standard deviation; CR = complete response; PR = partial response; NR = no response

The hearing outcomes are shown in Tables II and III. The final mean PTA was 47.7 dB, after a mean improvement of 24.3 dB. Thirty-two patients (28.1 per cent) experienced no change in hearing between presentation and post-treatment follow up, and another 15 patients (13.2 per cent) experienced an improvement of less than 20 dB. In the remainder (58.8 per cent), the PTA threshold improved by 20 dB or more. In eight cases (7 per cent), the PTA improvement was 60 dB or greater. Seventy-seven of the 114 patients (67.5 per cent) had a CR or PR. Both the final PTA and the amount of improvement in hearing differed significantly between all three response categories ($p \leq 0.001$). Of the 77 patients having a CR or PR, 73 experienced recovery within 30 days of onset of hearing loss. The remaining four patients had a delayed recovery commencing one month following onset of hearing loss.

Brainstem evoked response wave V was recorded in 60 (76.9 per cent) of the 78 patients with available follow-up BSER testing. During the one-year study period, wave V was identified within 30 days of the onset of treatment in all patients in whom it returned. In other words, in no patient was wave V identified later than one month following treatment, where it was present at all.

Factors related to hearing outcome

The relationship between the presence of waves I, III and V and the final hearing outcome was assessed. Table IV shows the non-parametric correlations between the time of identification of BSER waves (i.e. days from initial test) and the various outcomes. For all three waves, the time of wave identification was significantly related to response category, amount of improvement in hearing and final PTA. A shorter time to BSER waves identification was related to better response, greater amount of

TABLE III

HEARING OUTCOMES BY PTA MEAN AND RANGE*

PTA	Initial (dB)	Final (dB)	Improvement (dB)
Mean (SD)	72.1 (23.0)	47.7 (31.2)	24.3 (20.8)
Range	30–105	10–105	0–85

*For 114 sudden sensorineural hearing loss (SSHL) patients. PTA = pure tone average threshold; SD = standard deviation

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TABLE IV

CORRELATION BETWEEN BSER WAVES I, III, AND V AND HEARING OUTCOME*

BSER wave	Response	Hearing improvement	Final PTA	Time to presentation
I				
Rho†	−0.424	−0.315	0.572	0.437
p	0.001	0.005	0.000	0.000
III				
Rho	−0.443	−0.319	0.589	0.409
p	0.000	0.004	0.000	0.000
V				
Rho	−0.492	−0.379	0.610	0.308
p	0.000	0.001	0.000	0.006

*For 78 patients with brainstem evoked response (BSER) follow up. †Spearman's Rho correlation. PTA = pure tone average threshold

improvement and lower final PTA. A shorter time to BSER waves identification was also related to a shorter time between onset of hearing loss and presentation to our department.

With regard to wave V, comparisons were made between patients in whom the wave was recorded and those in whom it was not recorded within the one-year follow-up period (Table V). The identification of wave V was significantly related to hearing response; only 16.7 per cent of patients in whom wave V was recorded were categorized as NR, while 72.2 per cent of those in whom wave V was not identified were so categorized (chi-square = 20.9, $p \leq 0.001$). Patients with a recordable wave V showed significantly more improvement in hearing and a better final PTA threshold ($t = 3.6$, $p \leq 0.001$ and $t = -5.9$, $p \leq 0.001$, respectively) (t = t-student test). The mean time from hearing loss to presentation (and therefore to treatment) was significantly less in patients with a recordable wave V at presentation than in those in whom wave V could not be recorded ($t = -2.8$, $p \leq 0.011$). However, we must take into account the fact that these patients had less hearing loss to start with ($t = -4.8$, $p \leq 0.001$).

TABLE V

RELATIONSHIP OF BSER WAVE V TO HEARING OUTCOME AND OTHER VARIABLES*

Patient factor	Wave V		*p*
	Return ($n = 60$)	No return ($n = 18$)	
Hearing response			≤0.001
NR (%)	16.7	72.2	
PR (%)	33.3	16.7	
CR (%)	50.0	11.1	
PTA (mean (SD))			
Initial (dB)	64.5 (19.5)	90.0 (20.1)	≤0.001
Final (dB)	33.8 (19.6)	78.6 (30.5)	≤0.001
Improvement (dB)	30.7 (19.8)	11.4 (19.8)	≤0.001
Other factors			
Age (years)	42.7 (14.9)	41.3 (14.0)	NS
Days to presentation	4.9 (4.7)	10.2 (7.7)	≤0.011

*For 78 patients with brainstem evoked response (BSER) follow up. SD = standard deviation; NR = no response; PR = partial response; CR = complete response; PTA = pure tone average threshold; NS = not significant

TABLE VI

RESPONSE CATEGORY BY GENDER

Gender	NR	PR	CR	Total
Male				
n	15	17	30	62
% within gender	24.2	27.4	48.4	100.0
% within response	40.5	50.0	70.7	54.4
Female				
n	22	17	13	52
% within gender	42.3	32.7	25.0	100.0
% within response	59.5	50.0	30.2	45.6
Total				
n	37	34	43	114
% within gender	32.5	29.8	37.7	100.0
% within response	100.0	100.0	100.0	100.0

NR = no response; PR = partial response; CR = complete response

Other factors

Demographic variables, initial hearing, time to presentation and other factors were assessed in relationship to the final hearing outcome. Although the correlation coefficients were small, age was significantly related to final PTA (Rho = 0.229, $p \leq 0.014$), to amount of change (Rho = −0.241, $p \leq 0.010$) and to response (Rho = −0.232, $p \leq 0.013$), with older age being associated with a higher (i.e. poorer) final PTA, less improvement in hearing and a poorer response category. Men had a higher rate of CR than did women (48.4 *vs* 25.0 per cent) and a lower rate of NR (24.2 *vs* 42.3 per cent) (chi-square = 7.22, $p \leq 0.027$) (Table VI).

TABLE VII

RESPONSE CATEGORY BY AUDIOGRAM TYPE

Audiogram type	NR	PR	CR	Total
Deaf				
n	18	11	1	30
% within type	60	36.7	3.3	100.0
% within response	48.6	32.4	2.3	26.3
Ascending				
n	6	2	21	29
% within type	20.7	6.9	72.4	100.0
% within response	16.2	5.9	48.8	25.4
Descending				
n	8	13	5	26
% within type	30.8	50	19.2	100.0
% within response	21.6	38.2	11.6	22.8
Flat				
n	4	5	6	15
% within type	26.7	33.3	40	100.0
% within response	10.8	14.7	13.9	13.2
Cupeloid				
n	1	3	10	14
% within type	7.1	21.5	71.4	100.0
% within response	2.7	8.8	23.3	12.3
Total				
n	37	34	43	114
% within type	32.5	29.8	37.7	100.0
% within response	100.0	100.0	100.0	100.0

NR = no response; PR = partial response; CR = complete response

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Possible contributing factors (e.g. virus, hypertension, stress) did not appear to have a significant effect on the hearing outcome. Neither did the presence of vertigo or tinnitus at initial presentation. However, patients with a normal ENG had a higher rate of CR (55.6 per cent) than did those with a reduced ENG (CR in 22.9 per cent). There was no difference in the rate of NR between those with normal and abnormal ENG results.

As expected, initial PTA was highly related to final PTA (Rho = 0.713, $p \leq 0.001$) but was not significantly related to amount of improvement (Rho = −0.049, $p > 0.05$). Initial PTA, however, was related to response category (Rho = −0.428, $p < 0.001$), with higher initial thresholds related to poorer response. Poorer hearing results were correlated with worse initial thresholds at 4 KHz.

The shape of the initial audiogram was significantly related to response category (chi-square = 46.0, $p \leq 0.001$). Ascending and cupeloid types had higher rates of CR (72.4 per cent and 71.4 per cent, respectively) than did flat (40.0 per cent), descending (19.2 per cent) or deaf (3.3 per cent) types. Patients with a deaf classification had a higher rate of NR (60 per cent) than did those with other audiogram types (ranging from 30.8 to 7.1 per cent, for descending and cupeloid types, respectively) (Table VII).

Table VIII shows means for initial hearing, age and time to presentation, by response category. All of these factors were significantly related to response category. Patients who had a CR were found to have had better hearing at presentation, to be slightly younger and to have had a shorter time from onset of hearing loss to presentation than those who had NR. Patients who had a CR had better initial hearing, compared with those with a PR or NR.

Discussion

Despite the great advances in otology and skull base surgery over the last decades, idiopathic SSHL still remains a mystery. Most probably, we are not dealing with a specific disease but rather with the end result of various cochlear pathologies, such as viral infections, vascular disorders, rupture of intralabyrinthine membranes and inner-ear autoimmune diseases.[4,5] However, idiopathic SSHL is still a frightening experience for the patient and often a frustrating situation for the physician as he or she is forced to deal with the unknown, regarding not only aetiology, but also treatment and prognosis. Therefore, the identification of prognostic factors related to the final hearing outcome may offer tremendous help to both patients and ENT doctors.

The present paper assesses the prognostic value of demographic variables, level of initial hearing loss, time of presentation and other possible contributing factors (e.g. virus, hypertension, cholesterol, stress) in predicting final hearing outcome.

Increasing age was found to be related to a poor prognosis. The final hearing level, the amount of hearing improvement and the response to treatment were significantly related to age. These findings are consistent with those of other surveys.[3,11,14,15] In addition, in the present series, men showed a higher rate of CR and a lower rate of NR than women. Possible contributing factors, such as recent viral infection or hypertension, did not affect prognosis.

- **This paper presents demographic, epidemiologic, neurotologic and audiometric data from a series of 114 patients suffering from idiopathic sudden sensorineural hearing loss**
- **The paper concludes that the best prognosis was associated with youth, male sex, a shorter time lapse from the onset of hearing loss to the beginning of treatment, and upward-sloping or cupeloid audiogram contours**
- **The detection of BSER wave V within the first month of commencement of medical treatment might also be a significant favourable prognostic factor for hearing outcome**

Tinnitus was found to be the most frequent accompanying symptom; more than 90 per cent of the patients reported it. However, it did not have any prognostic significance regarding the final hearing outcome. This may very well be attributed to the small number of patients who did not have tinnitus (making meaningful comparisons difficult). In most studies, tinnitus is not reported to affect hearing

TABLE VIII

RESPONSE CATEGORY BY POSSIBLE PREDICTIVE FACTOR

Predictive factor	Response category (mean (SD))			*p*
	CR	PR	NR	
Initial PTA (dB)	58.1 (18.1)	79.6 (18.5)	81.4 (24.2)	≤0.001*
Initial threshold at 4 KHz (dB)	49.8 (22.4)	82.7 (19.9)	79.9 (27.1)	≤0.001*
Age (years)	41.3 (13.8)	45.0 (16.3)	49.7 (14.7)	≤0.047**
Days to presentation	3.9 (4.8)	5.2 (3.4)	8.2 (7.8)	≤0.003**

*Bonferroni post hoc comparisons: CR < PR; CR < NR. **Bonferroni post hoc comparisons: CR < NR. SD = standard deviation; CR = complete response; PR = partial response; NR = no response; PTA = pure tone average threshold

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outcome;[16] however, Danino *et al.* considered its presence an indication of surviving cell function and, consequently, a favourable prognostic sign.[9]

The prognostic significance of vertigo remains controversial. Although Danino *et al.*[9] and Ben-David *et al.*[7] considered vertigo an unfavourable prognostic factor, other studies failed to support these conclusions.[8,12,15] According to our data, the presence of vertigo had no prognostic significance in predicting hearing improvement. Interestingly however, in our report, individuals with normal ENG test results seem to have had a slightly better hearing outcome than those with a reduced vestibular response.

This study confirmed the idea that the sooner a patient is treated, the better the recovery.[3,4,15] The amount of time elapsed from onset of hearing loss to presentation and beginning of treatment appears to have significant prognostic value in predicting final hearing outcome. In addition, we noticed that the severity of hearing loss at the time of the initial evaluation was related to the occurrence of hearing improvement but not to the amount of hearing improvement.

Initial thresholds at 4 KHz were related to prognosis; this, to our knowledge, has not been previously reported.

Mattox and Simmons[13] and Mattox and Lyles[14] considered the shape of the audiogram to be the most useful predicting factor, with improved recovery being seen in patients with a low-frequency or mid-frequency audiogram contour. On the contrary, a downward-sloping audiogram was proposed to be an unfavourable indicator. In other words, high frequency thresholds (at 8 KHz) were considered to be good indicators of preservation of cochlear function at the basal turn and therefore of potential hearing recovery. Our outcome is consistent with these findings. Patients with ascending and cupeloid audiogram types had higher rates of CR (72.4 and 71.4 per cent, respectively) than did those with flat (40.0 per cent), descending (19.2 per cent) or deaf (3.3 per cent) types.

The patients in the present study underwent systematic BSER audiometry, as the recording of BSER waves is considered to reflect the functional integrity of the basal turn of the cochlea. It is known that the BSER at large stimulus intensities represents an onset phenomenon of the neural activity of the basal turn of the cochlea.[18] Accordingly, the identification of wave V was significantly related to hearing response, as was the type of audiogram, with only 16.7 per cent of patients with recordable wave V categorized as NR. On the other hand, 72.2 per cent of those with no identification of wave V were categorized as NR. Moreover, we noticed that the recording of wave V, in individuals in whom it returned at all, occurred within the first 30 days of treatment. During the one-year follow up, we did not record any 'delayed' (i.e. later than the 30th day) appearance of wave V.

Although a shorter time to wave V identification seemed to relate significantly to hearing improvement (Table IV), this result cannot be supported strongly by this study due to its design and to the subjective nature of the audiometry testing.

Consequently, we conclude that, in patients suffering from idiopathic SSHL, the earlier identification of BSER wave V during the first month of treatment might constitute a favourable prognostic factor regarding hearing recovery. In our experience, a shorter time to identification of wave V was related to better hearing recovery and lower final PTA. More targeted studies are need to confirm this finding.

Conclusion

We present demographic, epidemiologic, neurotologic and audiometric data from a series of 114 patients suffering from idiopathic SSHL who were treated in our department. After evaluating the prognostic significance of these factors, we conclude that younger age, male sex, shorter time elapsed from onset of hearing loss to beginning of treatment, and upward-sloping and cupeloid-type audiogram contours were related to better hearing outcome. We report that, during BSER audiometry, detection of wave V early in the first month following instigation of medical treatment might constitute a significant favourable factor regarding hearing recovery.

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