Chapter

Associated Health Issues of Patients with Acquired Unilateral Hearing Loss

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Abstract

Patients with unilateral hearing loss have impaired hearing of sounds coming from the affected side, decreased comprehension of speech in noisy environments, and lack of sound localization. There are many conditions that can induce unilateral hearing loss, but idiopathic sudden sensorineural hearing loss (ISSHL) is thought to be most notable because of its high incidence. Patients with ISSHL suddenly acquire unilateral hearing loss; therefore, there are additional significant health problems that complicate the abovementioned symptoms due to the characteristic clinical course of ISSHL including hearing-related discomfort, tinnitus, and anxiety. It has been reported that hearing-related discomfort is closely associated with patients' quality of life. In this chapter, the associated health issues of patients with ISSHL are described, and the interventions employed for patients with unilateral hearing loss are evaluated for their potential in improving the lives of ISSHL patients.

Keywords: idiopathic sudden sensorineural hearing loss, bone-anchored hearing aids, contralateral routing of signals, cochlear implant, hearing-related discomfort

1. Introduction

Because individuals with unilateral hearing loss have normal hearing in the opposite ear, impairment of auditory communication is less severe than in those with bilateral hearing loss. Therefore, interventions for these patients may not always be indicated. Unilateral hearing loss that is profound is known as single-sided deafness (SSD). Patients with SSD suffer from impaired hearing of sounds coming from the deaf side, lack of sound localization, and deteriorated comprehension of speech in noisy environments. These disorders and related problems can affect academic performance in children. Kuppler et al. reviewed that some children with SSD have significantly decreased self-esteem and increased level of exhaustion and stress because of the effort required to hear, and the tenfold increase of incidence (35%) of poor performer will be estimated [1]. It was also inferred that SSD have adverse effects on quality of life (QOL) and social life of both children and adults.

The causes of unilateral hearing loss include congenital and acquired diseases. Diseases inducing acquired unilateral hearing loss include idiopathic sudden sensorineural hearing loss (ISSHL), Meniere's disease, mumps, vestibular schwannoma, otosclerosis, otitis media with effusion, chronic otitis media, and cholesteatoma, among others. Hearing loss caused by otosclerosis, chronic otitis media, and

cholesteatoma can be improved by surgery, but inner ear damage resulting in SSD can rarely be reversed. Meniere's disease usually presents with mild-to-moderate hearing loss. Vestibular schwannoma presents with a wide range of hearing loss, from normal to total deafness, and if the tumor grows, surgical treatment is required, but the possibility of inducing SSD is relatively high after surgery. Though mumps is a disease that can acutely cause SSD, the incidence of hearing complications in mumps patients is quite low. ISSHL is quite a common condition, and it is estimated to produce the most patients with irreversible unilateral hearing loss.

Patients with ISSHL have an extremely dramatic clinical course in which unilateral hearing loss suddenly develops from normal hearing on both sides. In addition to the hearing-related problems that patients with congenital unilateral hearing loss or gradually worsening unilateral hearing loss face, there are other subsequent health issues that arise; therefore, ISSHL could be considered the most significant cause of acquired unilateral hearing loss. This chapter focuses on ISSHL as a representative condition that induces acquired unilateral hearing loss. First, the symptoms and health issues of patients with ISSHL based on the results of a nationwide survey of patients with ISSHL in Japan will be described. Next, the therapeutic interventions for patients with irreversible unilateral hearing loss caused by ISSHL will be considered.

2. Epidemiology and frequency of ISSHL

ISSHL is the sudden or acute onset of sensorineural hearing loss of unknown origin. The hearing loss is unilateral in most cases with bilateral involvement reported in <5% of cases [2]. The lesion is most often cochlear in origin, and less frequently retrocochlear. Although the cause of ISSHL has not been identified, several pathogenic possibilities have been proposed, such as vascular disorders, viral infections, and membrane breaks. Many treatment regimens have been investigated, including corticosteroids, vasoactive drugs, antiviral drugs, and hyperbaric oxygenation therapy, but none have proven effective. ISSHL is expected to improve on its own or with treatment; however, hearing levels become fixed ~2 months from the onset, and if it is not cured by then, permanent hearing loss remains.

The incidence of ISSHL is reported to be 3-30 per 100,000 population per year [2], but a recent report in Japan reported 60 per 100,000 population per year [3]. According to the population of Japan, about 78,000 new cases occur annually. For the hearing performance of 1113 patients with ISSHL in the author's hospital during the past 20 years during the persistent phase after treatment, 35% were completely cured, 27% had mild hearing loss, 26% had moderate hearing loss, 8% had severe hearing loss, and 4% had profound to total deafness. When this is considered with the above-estimated incidence, it is estimated that 21,000 patients with mild hearing loss, 20,000 patients with moderate hearing loss, 6200 patients with severe hearing loss, and 3100 patients with profound hearing loss develop unilateral hearing loss each year in Japan. Although vestibular schwannoma is also a relatively common disease inducing SSD, the number of surgeries performed annually in Japan to remove the tumors is estimated to be approximately 700 [4, 5], considerably less than the number of patients with SSD caused by ISSHL.

3. Symptoms and QOL in patients with ISSHL

Not many reports have investigated the symptoms or QOL in patients with ISSHL. Chiossoene-Kerdel et al. used the Hearing Handicap Inventory for Adults to

investigate the degree of handicap in patients with ISSHL [6], and most patients had a handicap associated with hearing loss and tinnitus. Carlsson et al. investigated QOL in patients with ISSHL using the EuroQoL-5D, Problems Impact Rating Scale, and the Hospital Anxiety and Depression Scale [7]. They reported that all three indicators were significantly influenced by the presence of tinnitus and vertigo. However, patients with Meniere's disease may have been included in that report, because the incidence of vertigo at the time of investigation was quite high (34%). Two main symptoms were reported to affect QOL in ISSHL patients with persistent hearing problems: difficulty in hearing and tinnitus [6, 7]. Unilateral hearing loss is a sudden change for patients who have never experienced hearing problems before the onset of ISSHL. Other problems, such as hearing-related discomfort and anxiety about recurrence, may also affect QOL. These problems may differ from those in patients with congenital SSD.

I and a few others conducted a multicenter clinical study by the Acute Profound Deafness Research Committee of the Ministry of Health, Labour and Welfare in Japan to investigate the symptoms and QOL in the patients with ISSHL in their persistent phase [8, 9]. The results from that study are described below.

A total of 140 patients with ISSHL (64 males, 76 females; mean age 59.1 years; range 21–85 years) and 24 patients with congenital SSD (13 males, 11 females; mean age 30.5 years; range 20–77 years) were investigated to determine their symptoms. In the patients with ISSHL, hearing levels of the affected ear were widely distributed from normal to profound, with a peak distribution of 70 dBHL. The distribution of time intervals from the onset of hearing loss varied widely from 30 days to 62 years (mean 5.5 years; median 2.7 years). The majority of patients with congenital SSD (71%) were between 20 and 29 years of age [8]. A symptom questionnaire was newly created to assess the patients' symptoms. In a previous investigation, information on symptoms was elicited from 104 patients with ISSHL using a freewriting method. These symptoms were rewritten and organized to create a new questionnaire comprising 17 questions that covered seven categories: hearing difficulty (three questions), spatial hearing (two questions), hearing-related discomfort (four questions), tinnitus (two questions), vertigo (one question), attitude to communications (two questions), and anxiety (three questions). We also asked patients with congenital SSD to answer the same questionnaire and compared the results with those patients with ISSHL [8]. The results of four major hearing-related symptoms, hearing difficulty, disability of spatial hearing, discomfort, and tinnitus, are shown in **Figure 1**. In response to questions regarding hearing difficulty, patients in both groups reported that they frequently had problems. With regard to the items "conversation with several people" and "conversation in noisy place," significantly more patients with ISSHL than with congenital SSD reported difficulty in hearing. In response to all questions regarding hearing-related discomfort, significantly more patients with ISSHL than with congenital SSD experienced symptoms. With respect to tinnitus, few patients with congenital SSD and many patients with ISSHL reported this symptom. In response to questions regarding spatial hearing, no difference between the groups was identified.

The health-related QOL in the patients with ISSHL and congenital SSD was investigated using the short-form health survey version 2 (SF-36). SF-36 provides scores for eight health-related QOL domains and two more comprehensive scores: the physical component summary (PCS) and the mental component summary (MCS). The scores for the eight domains and the two component summaries were standardized (norm-based scoring, Japanese average of 50, standard deviation of 10) for comparison with the scores of people in the general population or those reported in other studies. When the average scores for the two summary components in patients with ISSHL and those with congenital

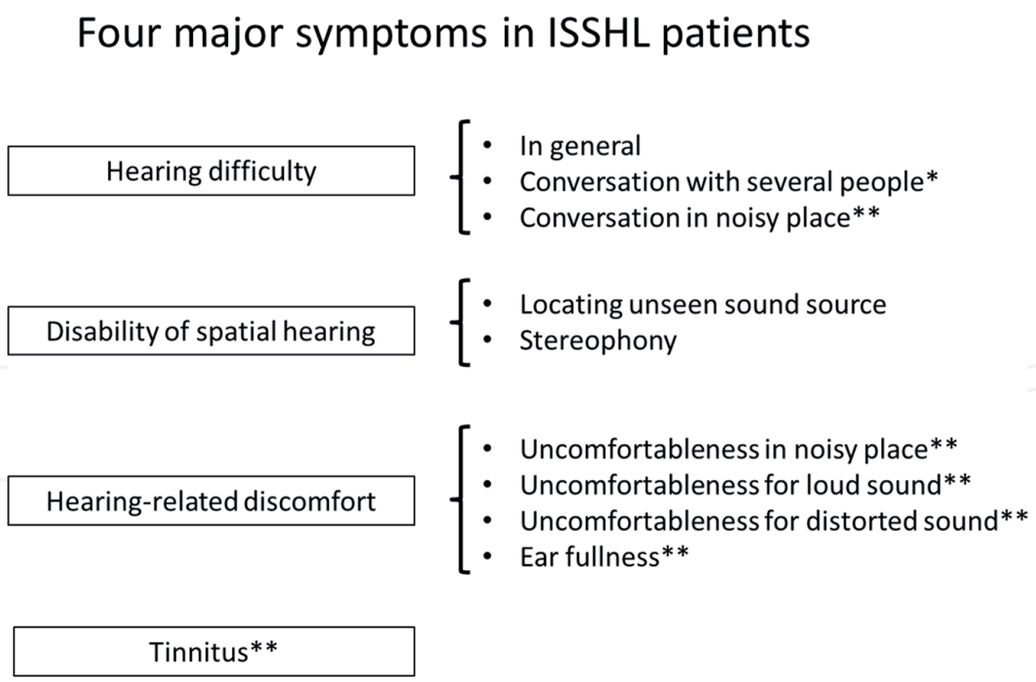

Figure 1.
Four major categories of symptoms reported by patients with ISSHL and congenital SSD. This figure was created from the results of Sano et al. [8]. "Hearing difficulty" consists of three items, "disability of spatial hearing" consists of two items, and "hearing-related discomfort" consists of four items in the questionnaire. P-values are presented as a comparison of the incidence between ISSHL and congenital SSD: $^{*}p < 0.05$, $^{**}p < 0.01$.

SSD were compared with average Japanese scores matched by decade of age, the PCS scores in all age groups for both ISSHL and congenital SSD patients were not significantly different from the Japanese average scores, and MCS scores for patients in their 20s for both ISSHL and congenital SSD and in their 30s for ISSHL were not significantly different from Japanese average scores. However, the MCS scores for patients with ISSHL in their 40–70s were significantly lower than the age-matched average Japanese scores (**Figure 2**).

Next, multiple linear regression analysis was used to investigate confounders influencing MCS scores in patients with ISSHL. The effects of age, hearing level at the time of investigation, time from onset, and responses to three items of the symptom questionnaire (hearing difficulty in general, discomfort in noisy places, and tinnitus) were evaluated. Unexpectedly, the results showed that the response to the item of discomfort in noisy places was the sole significant confounder.

"Hearing-related discomfort" was not a particularly notable symptom before this study; therefore, we further investigated this finding. When the relationship between the response to the item "discomfort in noisy places" and the hearing level in the affected ear was investigated, the response was not associated with hearing level [8]. A high incidence of this symptom was reported by ISSHL patients with moderate to profound hearing loss in the affected ear. We can infer that people with moderate hearing loss feel discomfort from noise because they hear noisy sounds in the affected ear, which can be too loud as a result of recruitment phenomenon and can be distorted by impairment of frequency selectivity. But why do patients with profound hearing loss in their affected ear feel this discomfort? In the direct expression of the discomfort by the patients, some of them described it as "It feels very noisy because the noise around me spreads all over the space around me." We proposed that the reason for this symptom in patients with unilateral profound hearing loss was sudden loss of the ability to localize the sounds coming from various directions, and we called this condition "collapse of spatial hearing perception."

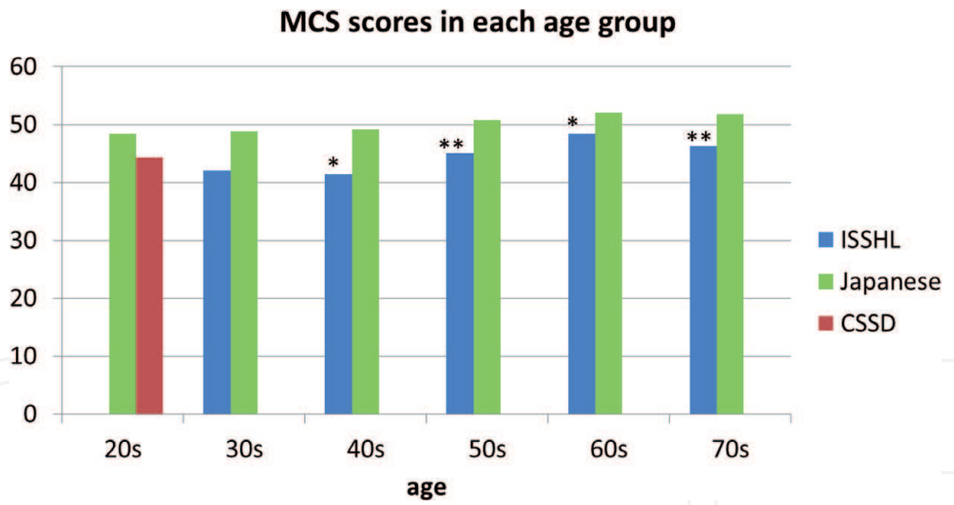

Figure 2.
*Mental component summary (MCS) scores of SF-36 in patients with ISSHL and congenital SSD. The graph was created using the data from Sano et al. [8]. "Japanese" indicates age-matched Japanese average scores. The results of 20s group of ISSHL and other than 20s groups of congenital SSD (CSSD) are not indicated because their numbers of patients were too small. P-values are presented as a comparison of scores between each age group of ISSHL or congenital SSD and the age-matched Japanese average: *p < 0.05, **p < 0.01.*

Individuals who had normal hearing in both ears before the onset of ISSHL suddenly lose the ability of spatial hearing, and they instantly begin and remain to feel discomfort in noisy environment. This symptom is important for understanding the QOL problems associated with ISSHL.

4. Therapeutic interventions for irreversible unilateral sensorineural hearing loss caused by ISSHL

As mentioned earlier, patients with ISSHL have several hearing-related symptoms, which can be divided into four categories: hearing difficulty, disability of spatial hearing, discomfort, and tinnitus. Hearing difficulty includes several situations, such as conversation with several people and speech perception in noise. The efficacy of therapeutic interventions for patients with ISSHL needs to be evaluated against those four categories. The available methods for evaluating those four categories are summarized in **Table 1**.

The degree of unilateral hearing loss caused by ISSHL varies from mild to profound, and the selection of interventions depends on the degree of hearing loss. The interventions for ISSHL patients with severe-to-profound hearing loss can be considered the same as those for SSD patients, which include cochlear implant (CI), bone-anchored hearing aids (BAHAs), and contralateral routing of signals (CROS)

	Objective evaluation	**Subjective evaluation**
Hearing difficulty	Speech discrimination test	SSQ/APHAB/GHABP
Spatial hearing	Sound localization	SSQ
Discomfort	Not available	Not available
Tinnitus	Not available	THI/VAS

SSQ, speech, spatial and qualities of hearing scale; APHAB, abbreviated profile of hearing aid benefit; GHABP, Glasgow hearing aid benefit profile; THI, tinnitus handicap inventory; VAS, visual analog scale.

Table 1.
Evaluation methods for four major problems in patients with ISSHL.

hearing aid. If hearing loss is mild to moderate, a conventional air- conducted hearing aid may be the primary treatment. These treatments are described below.

4.1 Therapeutic effects for patients with SSD

The methods for evaluating the therapeutic effects of interventions for patients with SSD have generally included sound localization test, speech comprehension in noise, and subjective evaluation. The effects of the BAHA, CROS hearing aid, and CI interventions are summarized below.

4.1.1 BAHA and CROS hearing aid

The BAHA and CROS hearing aid have essentially the same characteristics in that a microphone is placed on the affected ear side and the sound is heard in the normal ear. Whereas a BAHA transmits sound via bone conduction, a CROS hearing aid transmits sound to a receiver on the normal ear by wireless or wired transmission. The therapeutic effect of these hearing aids can be summarized from the results of two systematic reviews for adult-acquired SSD [10, 11].

For sound localization by BAHAs, Kim et al. [10] reported that the percentage of correct sound localization was 13–65.8% before BAHA implantation and 15–68.5% after implantation, showing no significant difference in six studies. In a recent study, Agtrberg et al. [12] reported that BAHA neither improved nor deteriorated the localization abilities of patients with SSD. Kitterick et al. [11] reported that sound localization was not changed by CROS hearing aids in five studies and a significant deficit was indicated in one study. Therefore, it can be concluded that there is no improvement in sound localization with BAHAs or CROS hearing aids.

Speech comprehension in noise improves under certain conditions. Kim et al. [10] reported that in the situation of speech coming from the front and noise coming from the normal ear side, speech discrimination was statistically significantly improved after BAHA implantation in four out of six studies. Kitterick et al. [11] conducted a meta-analysis using data from the Hearing in Noise Test. A significant benefit was identified in the situation of speech coming from the front and noise coming from the normal ear side for both BAHAs and CROS hearing aids; however, a significant deficit was identified for both devices in the situation of noise coming from the affected ear side. The effects of BAHAs and CROS hearing aids are generally similar, with the former thought to be slightly superior. For the subjective evaluation of the benefits and adverse effects of the interventions, the abbreviated profile of hearing aid benefit (APHAB) and the Glasgow hearing aid benefit profile (GHABP) have been widely used. Kitterick et al. conducted a meta-analysis using data from the APHAB before and after the use of BAHAs and CROS hearing aids. Significant benefits of the BAHA were found for three subscales, reverberation, ease of communication, and background noise, but not for aversion to loud sound. Significant benefits of the CROS hearing aid were also found for two subscales: background noise and reverberation.

4.1.2 Cochlear implant

CI is a method of treating the deaf ear itself so that it can regain hearing ability. Therefore, the treatment concept is essentially different from the BAHA and CROS hearing aid. A study of CI for adult-acquired SSD was reviewed. Two systematic reviews [13, 14] and several subsequent reports [15–18] yielded similar results. These are summarized as follows.

For sound localization, Kitterick et al. [11] reported that only one of the three studies showed statistically significant improvement after CI surgery. Although most of studies reported improvement of sound localization after CI, a meta-analysis could not be conducted because of heterogeneous methodologies.

For speech comprehension in noise, Blasco and Redleaf [12] conducted a meta-analysis and reported in the situation where both speech and noise were coming from the front; the signal-to-noise ratio for speech perception in noise was significantly improved following CI. However, in the situation where speech was coming from the front and noise was coming from the affected side, no improvement was observed.

For the subjective evaluation, Kitterick et al. [11] conducted a meta-analysis using data from the speech, spatial and qualities of hearing scale (SSQ) [19]. They found significant improvement for all three subscales: speech, spatial, and "other" qualities.

For the subjective evaluation of severity of tinnitus, Blasco and Redleaf [13] conducted a meta-analysis using a visual analog scale from three studies and found statistically significant improvement following CI implantation.

Overall, an important difference in the results of CI from those of the BAHA and CROS hearing aid is that there was a possibility of improvement for sound localization and spatial hearing ability. It is considered that these two factors are associated with each other. In addition, Legaris et al. reported that cortical reorganization and restoration of binaural function in the brain might be produced after 1 year of experience with CI in adult SSD patients by evaluation of cortical auditory evoked potential changes [20].

4.2 Potential of interventions for ISSHL patients with severe-to-profound hearing loss

BAHAs and CROS hearing aids improve hearing from the deaf side. Improvement in speech comprehension in noise can be expected when the speech comes from the deaf side or front and the noise comes from the normal ear side. However, the ability for sound localization cannot be expected, and it is not possible to restore spatial hearing function. Therefore, among the associated problems for patients with ISSHL with severe-to-profound hearing loss, both devices seem to give no benefit for spatial hearing, discomfort, or tinnitus.

A CI improves speech comprehension in noise at least as well as the BAHA or CROS hearing aid. A CI also seems to have potential to improve sound localization, which could lead to restoration of spatial hearing ability. The SSQ subjective evaluation contains many assessment items related to spatial hearing [19], and the scores of spatial hearing were reported to improve after cochlear implantation. Although there have been no reports directly evaluating "hearing-related discomfort" as an important symptom of ISSHL, it may be improved if spatial hearing ability can be restored. Direct evaluation of this symptom is needed in future assessments. Patients with tinnitus can also be expected to experience improvement with a CI [13, 15, 18]. Overall, although further investigation is needed, a CI has the potential to improve speech comprehension in noise, spatial hearing, and tinnitus and may also improve discomfort. The indication of CI in patients with ISSHL is the confirmation of cochlear pathogenesis, and relatively early surgery after ISSHL onset should be considered [18].

4.3 Potential of interventions for ISSHL patients with mild-to-moderate hearing loss

Conventional hearing aids are indicated for ISSHL patients with unilateral mild-to-moderate hearing loss. However, patients with mild-to-moderate hearing loss on

the affected ear and normal hearing on the opposite ear are less likely to realize the benefits of hearing aids on the affected ear and are often unable to wear them. Since some degree of auditory function remains in the affected ear, symptoms such as difficulty in hearing and impaired sound localization are milder than in patients with SSD, and, as a result, the beneficial effect of wearing a hearing aid seems to be difficult to perceive subjectively and to detect objectively. In addition, Kumpik et al. reviewed from several studies that horizontal localization by adult humans can adapt to varying degree to asymmetric hearing loss induced by occluding one ear [21]. Therefore, the abilities of sound localization and spatial hearing may be spontaneously restored in some degree in the patients with unilateral mild-to-moderate hearing loss. There do not seem to be any previous reports that examined the effect of hearing aids for hearing disability in patients with unilateral mild-to-moderate hearing loss.

Hearing-related discomfort is also common in ISSHL patients with unilateral moderate hearing loss, but the mechanism may differ from that in patients with unilateral severe-to-profound hearing loss. Patients with moderate hearing loss are more likely to have discomfort with sounds heard on the affected side, that is, increased loudness of noise caused by the recruitment phenomenon or distortion caused by the impairment of frequency selectivity function is unpleasant. Therefore, it is unlikely that a hearing aid will improve excessive loudness and distortion of sound.

On the other hand, tinnitus symptoms are more common in patients with mild-to-moderate hearing loss due to ISSHL. Tinnitus retraining therapy using a hearing aid as a means of sound therapy has been widely conducted. At present, there is no high-quality evidence from systematic reviews [22], but improvement in the Tinnitus Handicap Inventory or visual analog scales has been widely recognized. In the future, it will be necessary to establish evidence of tinnitus improvement and to evaluate speech comprehension in noise and sound localization as well as hearing-related discomfort in ISSHL patients with mild-to-moderate hearing loss.

5. Summary

ISSHL is an important cause of persistent unilateral sensorineural hearing loss that affects thousands of new patients annually in Japan. The problems caused by ISSHL can be categorized into four factors: hearing difficulty, deterioration of spatial hearing, hearing-related discomfort, and tinnitus. The interventions that have been used to treat patients with unilateral hearing loss can be adapted to patients with ISSHL. The expected benefits of interventions for ISSHL patients are shown in **Table 2**. Although there are presently no treatments that provide satisfactory outcomes, CI is possibly the current most effective means of restoring some

Grade of hearing loss	**Severe-deaf**		**Mild-moderate**
Intervention	**BAHA/CROS**	**CI**	**Conventional HA**
Speech in noise	Partially improved	Partially improved	?
Spatial hearing	→	Possibly improved	?
Discomfort	?	?	?
Tinnitus	→	Improved	Improved

BAHA, bone-anchored hearing aid; CROS, contralateral routing of signals; CI, cochlear implant; HA, hearing aid; →, not improved; ?, not available for applicable investigations.

Table 2.
Expected effect of interventions for patients with ISSHL.

of the lost binaural functions in patients with ISSHL who have severe-to-profound hearing loss. On the contrary, the effect of BAHA and CROS is quite restricted for such patients.

In the future, it will be necessary to unify evaluation methods for sound localization, speech comprehension in noise, and subjective questionnaires. Health-related QOL should be a component of the subjective assessments, and "hearing-related discomfort," which negatively impacts QOL for ISSHL patients, must be included as a subjective evaluation item.

Conflict of interest

The author has no conflict of interest to declare.

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