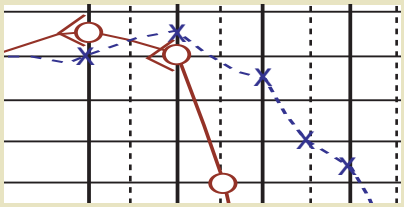
abstract



Hearing loss that occurs instantaneously or over a period of a few days without immediately apparent cause is called Idiopathic Sudden Sensorineural Hearing Loss (ISSNHL). In part 1 of this series, the diagnosis and initial treatment of this condition are described in relation to most patients' demands for active and aggressive intervention. Part 2 (to follow in the next issue) will address rehabilitation.

Key words: *audiology, deafness, diagnosis, hearing aids, idiopathic, otology, rehabilitation, unilateral and bilateral hearing loss, sensorineural*

Sudden Deafness, Part 1: Diagnosis and Treatment

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Introduction

Rush Limbaugh's severe-to-profound, bilateral, rapidly progressive hearing loss generated considerable public interest in sudden deafness. In his case, its cause was reportedly an autoimmune disease of the cochlea.[1] Diagnostically, however, many patients who suffer a sudden hearing loss are not as fortunate as the popular radio commentator. The cause of their sudden hearing loss remains unexplained.

Idiopathic sudden sensorineural hearing loss (ISSNHL) ranks high among the difficult clinical conundrums facing audiologists and otologists. Idiopathic refers to unknown etiology, and at least a third of cases of sudden hearing loss fall into this category.[2] Inherently, ISSNHL imposes difficult treatment decisions on the clinician.

Much of the literature urges prompt treatment. In the face of ISSNHL's mysterious onset and indeterminate prognosis, should rehabilitation proceed? The second instalment of this article addresses that question; here we describe ISSNHL, its incidence, diagnosis, and initial treatment.

It is estimated that between five and 25 per 100,000 persons in the U.S. will suffer ISSNHL in any given year.[1] Canadian experience with ISSNHL probably does not deviate significantly from this statistic,[3] although Canadians with impaired hearing differ in several demographic respects from their U.S. neighbours.[4] Reasons why reported incidence rates likely underestimate ISSNHL's occurrence in Canada are that available data are for reported cases; when hearing quickly recovers, the sudden loss is not reported; and lastly, many people do not seek assistance for mild losses, even though they are sudden and unexplained.

ISSNHL's incidence is far greater in adults than children. The average age at onset is 40–50 years, with the majority of the patients over 40 years old.[5] Another established relationship is that ISSNHL is 50 times more likely to be unilateral than bilateral.[6,7] Only the unilateral condition is considered in this paper because of the rarity of bilateral cases.

Again, the size of the available statistics must be viewed with caution for the reasons noted above. Nonetheless, the relationships within ISSNHL will probably remain as described.

Illustrative Case Histories

On 5 March 1999, Patient A, a 61-year-old surgeon, awoke with a severe hearing loss in his right ear. His general health was excellent, and he had no tinnitus or vertigo—hopeful diagnostic indicators. His audiologic findings appear in Figure 1. Hearing in the left ear was within normal limits. The right ear showed a severe, sensorineural hearing loss averaging 70 decibels (dB) hearing threshold level (HTL). The audiometric configuration was gently rising. His ability to understand speech was poor to absent—a prognostically bad sign.

He received a diagnosis of ISSNHL and was given a recommendation for a

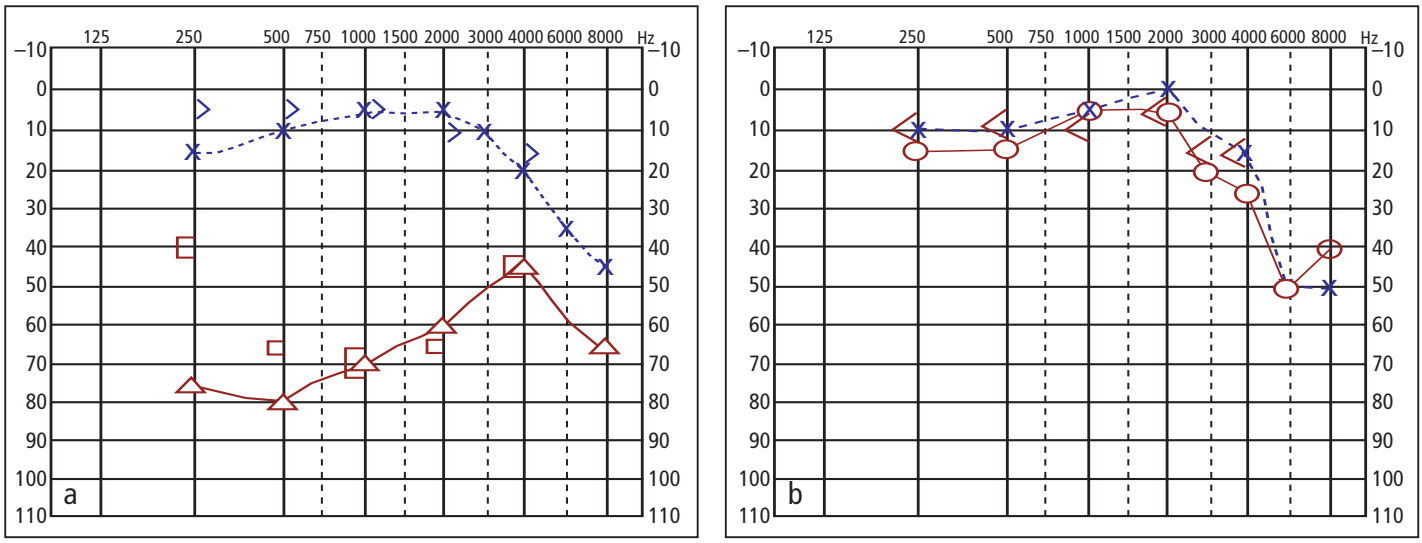

Figure 1: Idiopathic Sudden Sensorineural Hearing Loss (ISSNHL)

(a) Within 48 hours of onset, hearing within Patient A's left ear was within normal limits. His right ear (red) showed a severe hearing loss.

(b) Despite not filling his prescription of orally administered corticosteroids, Patient A's hearing returned to normal two weeks later.

short course of orally administered corticosteroids. He accepted the prescription but did not fill it! Two weeks later, without intervention, his hearing returned to normal.

His case illustrates complete spontaneous recovery of hearing. Had he followed the recommendation to take the corticosteroids, an incorrect causal relationship might have been drawn.

Patient B was also 61 years old. Upon return from a trip to Asia, he noticed a hearing loss in his right ear accompanied by "distortion and poor discrimination" and tinnitus, which he described as "aggressive." The pure-tone audiogram of 12 July 1999 (Figure 2) shows essentially normal hearing in the left ear. The right ear showed a mild sensorineural loss beginning at 1500 Hz and a moderate-to-severe loss in the 2,000–8,000 Hz range. His understanding of speech was moderately impaired.

He was started on oral prednisone (40mg/day). A week later, audiograms showed worsening of hearing in the right ear, with a flat configuration. It was decided to inject a corticosteroid directly into the middle ear for absorption into the cochlea. Ten to 20 minutes after the procedure, tinnitus was greatly reduced. Subsequently, audiometric testing showed significant improvement in hearing. He was fitted with a hearing aid on the right ear, which greatly improved his ability to communicate in challenging listening situations. The results of the procedures supported the efficacy of the surgically injected prednisone treatment to improve his hearing and relieve the tinnitus.

Diagnostic Factors

The interval between ISSNHL's onset and medical care appears critical to recovering hearing. Based on an eight-year prospective study of 225 patients, investigators found the proportion of cases enjoying spontaneous recovery declined after 30 days, and with little hope for regaining hearing spontaneously after six months.[6]

Further associations with spontaneous recovery are the degree of the presenting hearing loss and its audiometric shape. Although based on a small sample, evidence suggests that patients whose hearing losses following ISSNHL are less than 70dB HTL appear to have the best possibility for remission.[8] Cases with rising and mid-frequency audiometric curves predict spontaneous recovery and will occur more frequently than those with sloping or flat configurations.[5]

Medical/Surgical Treatment

Results of any treatment must be weighed against the possibility that hearing will be restored with no apparent intervention. Unfortunately, to date, no single treatment for ISSNHL appears to have achieved that level of success.

In a study of 266 patients with ISSNHL who were administered short-term oral steroid treatment and compared to a control group of 52 untreated patients, hearing improvement averaged 29dB HTL for the treated sample and 11dB HTL for the control group. Although both groups' word recognition improved, a significantly larger number of treated patients improved. However, 40% of steroid-treated cases did not recover at all, or their hearing worsened over the treatment period and their losses remained permanent.[9] Adding magnesium to the corticosteroid dosage improved results in another study.[10]

Mindful of the advisability in most diseases of prompt treatment, practitioners frequently prescribe steroids and vasodilators for ISSNHL before an etiology can be uncovered

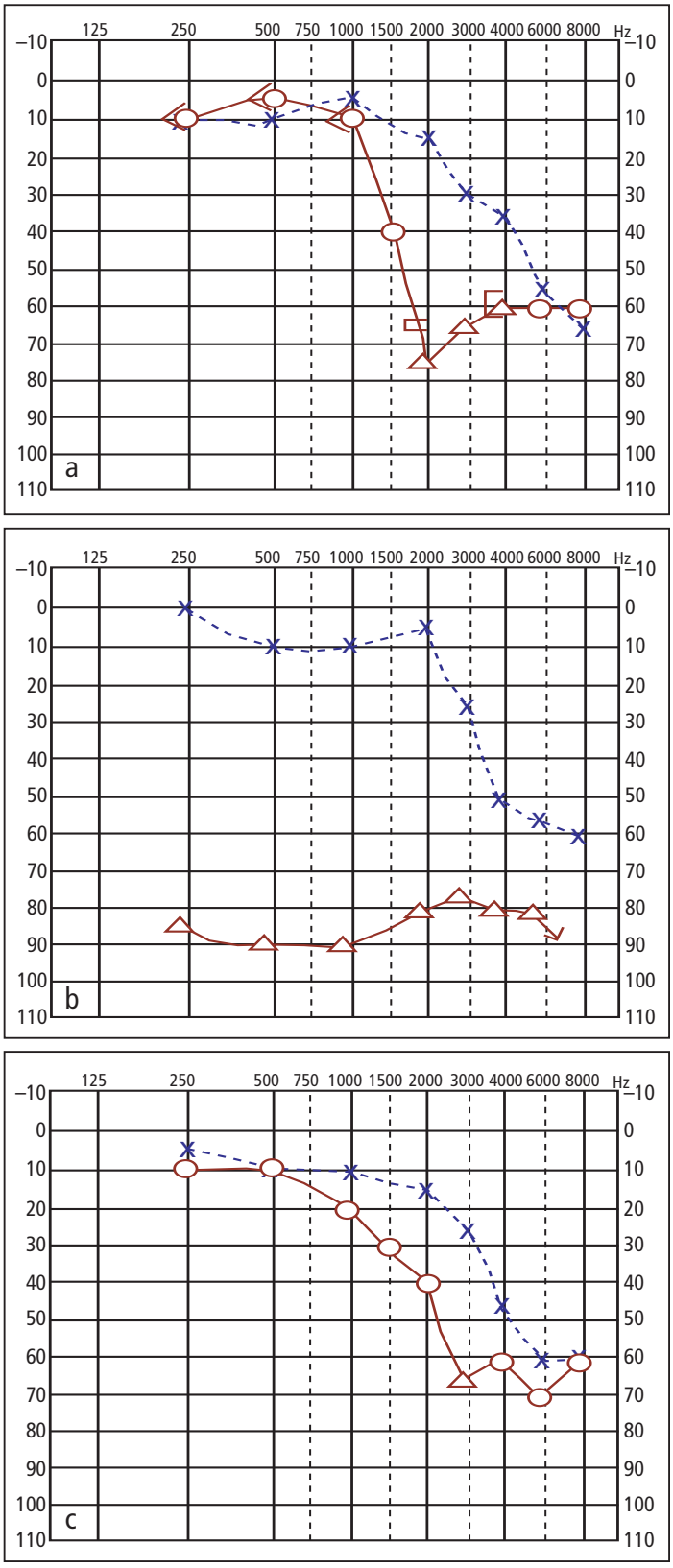

Figure 2: (a) Patient B noticed hearing loss in his right ear (red). He was started on oral prednisode (40mg/day). (b) A week later, audiograms showed a worsening of hearing in his right ear. It was decided to inject a corticosteroid directing into the middle ear. (c) 10 to 20 minutes later, tinnitus was greatly reduced; audiometric testing showed significant improvement in hearing.

(see case histories above). The rationale for using vasodilators is based on the theory that vascular occlusion may be implicated in some cases of ISSNHL.[7] The theory has been labelled "a neuro-otologic myth."[11] In a review of 41 ISSNHL patients' charts, researchers concluded, "There was no correlation of pre-existing signs, symptoms, or findings with hearing recovery [following use of vasodilators and steroids]."[12] Similar discrepant findings appear throughout the ISSNHL literature.[8,13–15] Yet, despite a recent study supporting their use for some patients,[9] many practitioners regard corticosteroids as the "gold standard" of treatment for ISSNHL.

Whether or not hearing can be recovered, early treatment should attempt to alleviate two conditions that accompany a large portion of ISSNHL cases: tinnitus in about 70% and vertigo in about 50%.[16] Some patients find these symptoms equally if not more bothersome than the loss of hearing. Vertigo frequently occurs concurrently with the ISSNHL but in some cases, may follow initial loss of hearing. Its presence is a poor indicator for recovery.

Whatever the degree of the impairment or the frequency of its occurrence, ISSNHL deserves attention. If it persists, it is worth attempting to manage because of the disruption it causes in the patient's life, and because it could accompany and reflect another serious condition.[17]

Summary

ISSNHL is a major challenge to patients who suffer from it and to the hearing professionals who attempt to manage it. Although it occurs infrequently, its effects are serious and debilitating. In addition to the loss of hearing, ISSNHL is often accompanied by tinnitus and vertigo. Older adults are more likely to suffer from it.

Diagnosis of sudden hearing loss can be difficult and, in about one of three cases, unavailing. Treatment must be considered in relation to the high rate of partial or complete spontaneous recovery. Recovery is related to audiologic characteristics and time between ISSNHL onset and treatment. Early evaluations by trained specialists and prompt initiation of steroid therapy, when indicated, can affect recovery.

If there is no return of hearing after a period of 60–90 days, with or without medical and/or surgical treatment, the audiologist should introduce amplification and supportive audiologic rehabilitation to the patient, which will be discussed in Part 2. ◆

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