Ultra-High Frequency Sudden Sensorineural Hearing Loss

By Dennis Colucci, AuD, MA

Sudden sensorineural hearing loss (SSHL) is defined as a change in hearing of 30 dB or more in at least three consecutive frequencies within a 72-hour period. Clinically, a lower threshold is used, especially if there are known risk factors. The presenting audiometric findings are typically unilateral and characterized as a low, high, or flat frequency response using audiometric frequencies between 125-8,000 Hz. The uncomfortable loudness levels (ULL) may or may not demonstrate hyperacusis. Tinnitus matching is typically measured at or below 8,000 Hz, particularly for tinnitus caused by noise-induced hearing loss, although tinnitus may occur above this range (*Int Arch Otorhinolaryngol*. 2016;20[3]:248).

Remarkably, no systematic study using ultra-high frequency audiometry could be identified in the SSHL or tinnitus literature in the last five years. However, in the case presented herein, an extended frequency evaluation revealed an uncommon result worthy of examination. Although immediate air and bone conduction audiometric testing is paramount in the diagnosis of SSHL, it is also used to measure the outcome of medical treatments or spontaneous recovery, and in planning rehabilitation (*Hearing Journal*. 2013;66[4]:44). Ultra-high frequency testing may provide a wider range of understanding of patient complaints in the absence of hearing loss using standard audiometry.

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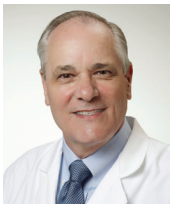
In some cases of SSHL, the cause can be determined and treated, but most cases are idiopathic. Identifiable causes include autoimmune disease, bacterial or viral infection, functional deficits, metabolic factors, neoplasms, neurologic factors, otologic factors, vascular disease, toxic exposure, and trauma (*Trends Amplif*. 2011;15[3]:91). In a small percentage of cases, the audiometric configuration may be useful in determining the potential causation (i.e., noise trauma, Meniere's disease), but this is not typical.



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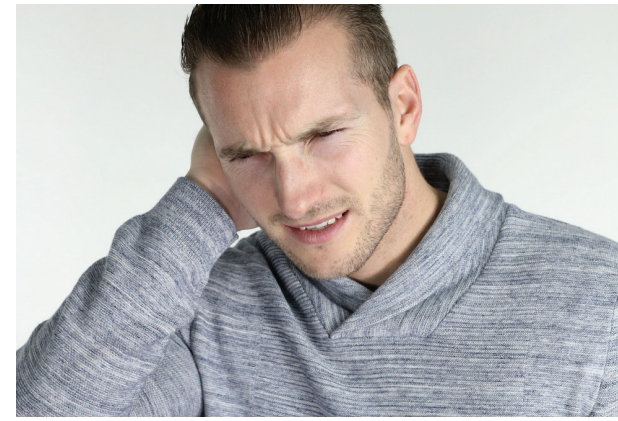

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SSHL SPECIFICS

Regardless of the audiometric configuration, SSHL may be accompanied by aural fullness, tinnitus, vertigo, pain, and hyperacusis, among others. Forty percent of tinnitus patients also complain of hyperacusis, while 85 percent of hyperacusis patients complain of tinnitus. In addition, approximately 76 percent of tinnitus patients complain of sleep deprivation, which is known to contribute to tinnitus severity and functional distress (British Tinnitus Association, 2016). In a study by Paulin et al, migraines were reported in 12.1 percent of patients with hyperacusis (*Noise Health*. 2016;18[83]:178).

Functionally, tinnitus patients complain of confusion, an inability to concentrate, communication failure, and the need to avoid social, family, and work situations. Depending on the severity of the conditions, there is a top-down impairment, as well as bottom-up deficits, that change the neurodynamics in the processing of incoming information, even in normal hearing subjects (*Hear Res*. 2016 Oct 8. pii:S0378).

Audiometric testing regularly measures frequencies between 125 Hz and 8,000 Hz. Typically, most of the hearing changes occur in this range, as does tinnitus. However, a case of unilateral ultra-high frequency SSHL suggests that a near normal or normal audiogram does not always mean a lack of significant hearing loss, and extended range testing may reveal valuable information.

CASE REPORT

The patient is a 23-year-old healthy, athletic male taking no daily medications, although occasionally uses ibuprofen and naproxen for muscle pains. The patient also uses Flonase and OTC medication for seasonal allergies. The patient denies acoustic trauma, barotrauma, or head or neck injury. There is

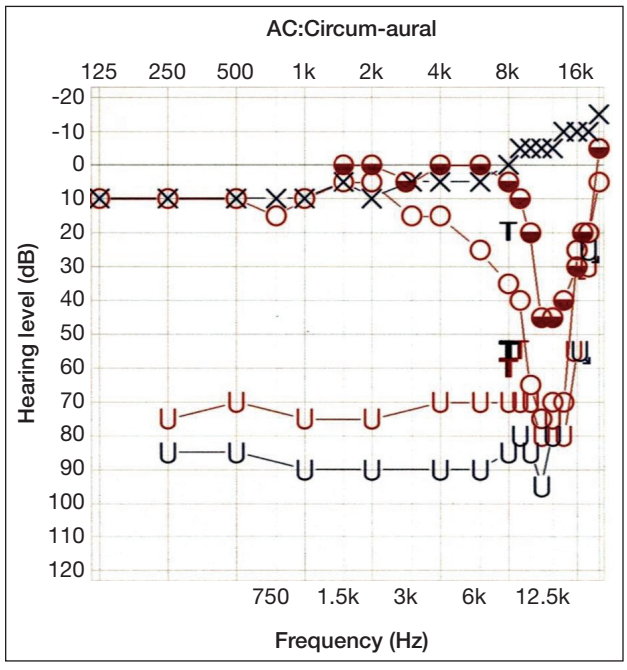

Figure 1. Ultra-High Frequency Sudden Sensorineural Hearing Loss. Open circles and X's represent air conduction findings for the right and left ears on the first day of testing. Half-filled circles are the findings at one-week post onset. Uncomfortable Loudness Levels are indicated by the "U" and the "T" is for tinnitus matchings on the two separate test dates.

no history of noise or toxic substance exposures, and there is a negative familial history of hearing loss. The remainder of the history is uneventful.

Initial symptoms were reported and informally scored for impingement on a 10-point scale by the patient. The symptoms started with are a minor sore throat (2/10) and moderate aural fullness (6/10) in the right ear upon awakening in the morning. Within two days, a mild to severe, dramatically sloping, ultra-high frequency hearing loss with tinnitus in the right ear was recorded (Fig. 1), which was accompanied by the onset of loud tinnitus 24/7 with sound sensitivity (8/10). The tinnitus was initially measured at 55 dB at 9,000 Hz in the right ear, and at 20 dB at 8,000 Hz in the left ear (Note: The patient only complains of tinnitus and sound sensitivity in the right ear; tests of the left ear are used to compare the two ears).

At two weeks, tinnitus at 8,000 Hz is measured at 60 dB in the right ear and balanced to 55 dB in the left. The ULL measurements between 250 Hz and 16,000 Hz confirmed the presence of hyperacusis. Evidence of asymmetry showed the right ear is more sensitive (by 10-15 dB) than the left ear, which also showed a decrease in tolerance.

Within days, the patient reported difficulty falling and staying asleep (10/10), difficulty understanding conversations, especially in the classroom (5/10), and increased anxiety (6/10). At six weeks post onset, the patient's Tinnitus Handicap Inventory score was profound (88 points, F=38/44, C=18/20, E=32/36) and the Modified Khalfa Hyperacusis Questionnaire total score was severe (89 points, F=30/35, S=27/30, E=32/35).

Following the initial findings, the patient was seen on day three by an ENT physician who diagnosed sudden hearing loss and prescribed oral corticosteroids and antiviral medication to be used for two weeks. At the end of the first week, the hearing loss reversed nearly in half and the aural fullness was resolved. However, there was no change thereafter, regardless of weekly transtympanic perfusions of Dexamethason for three weeks following the oral medications, and an eight-day course of hyperbaric oxygen treatments. At the end of all medical treatments, the tinnitus and hyperacusis symptoms and measurements remained unchanged. Based on the history, constellation of findings, and time course, a viral causation was suggested by the neurotologist and ENT physician.

In view of the functional complaints and the fact that auditory processing is altered by changes in the functional networks, both top-down and bottom-up CAP tests were conducted. Although the validity of these tests may be of question, certainly the functional changes are important to patient management. Using SCAN 3, RASP, Time Compressed Sentences, MLD, and QuiskSIN, the only abnormal findings were in the dichotic listening for words and sentences on the SCAN 3 (–3 sd) and a left ear advantage falling in the two percentile of prevalence. In a healthy individual, this would suggest difficulty related to focused attention or binaural separation, and divided attention or binaural integration.

SUMMARY

Regardless of causation, the level of recovery varies from "complete" to "no improvement" based on several factors, including the age of onset, degree and type of hearing loss, comorbidities, risk factors, and most importantly, the timeline between the onset of symptoms and treatment.

In this case, the initial audiogram reveals a slight asymmetry between the ears at 6,000-8,000 Hz, but by testing the ultra-high frequencies, the totality of the high frequency hearing loss is revealed as severe. Should the patient be evaluated after there is some time for hearing recovery, as seen in this case after only one week post-onset, the patient's hearing would have been reported as normal on standard audiometry, but abnormal when testing for loss in the ultra-high frequencies. Recognizing the hearing loss does not necessarily change the outcome or treatment. However, it does identify the loss as cochlear, providing a basis for measuring improvement from treatments or spontaneous recovery. It also provides the patient a confirmation of the site of lesion and supports the direction of therapy, especially when the psychopathology may be evolving.

The most important aspect of recovery following medical treatment is controlling cognitive distortions (i.e., life will never be the same, there is no hope), which may have adverse impact. Significant distress can become overwhelming when the patient has a negative evaluation of the tinnitus, along with a selective auditory attention that affects perception, central auditory processing complications, and other comorbidities (*BMC Ear Nose Throat Disord.* 2016;16:10). In this case, a psychiatrist may be consulted to control the medical aspects of comorbid insomnia, headaches and migraines, and anxiety and depression. A psychologist may also help in dealing with aberrational cognitive behaviors that lead to anxiety and stress, adjustment disorder, or PTSD.