

A Healthy 64-Year-Old Man With Sudden Hearing Loss

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A 64-year-old man presents to his internist's office complaining of hearing difficulty of several days' duration. He believes it is the right ear that is affected. There had been a feeling of "fullness" in his ear for about a week. He noted having difficulty in following speech when watching the television. He also noted difficulty hearing people when speaking to them in person unless he was in a position to read their lips or by turning his head to direct the sounds to his left ear. The deficit of the right ear was continued when he used the telephone. He denies any vertigo or dizziness.

Otherwise, the patient's health seems excellent with only mild hypertension controlled by losartan. There has been no remote exposure to ototoxic medicines; he does not smoke; he has no history of exposure to excessive noise; and there is no history of recent head trauma.

Office examination revealed normal blood pressure (BP). Cranial nerves II-VII and IX-XII were normal. There was no nystagmus. The left and right ear canals had small amounts of cerumen: the right side

contained more cerumen than left, but the tympanic membrane was visible bilaterally. Tuning Fork examination revealed marked loss of hearing on the right side when a Rinne test was performed. The Weber test revealed distinct sound lateralization favoring the left ear.

Which of the following is the most appropriate next step in managing the presented patient?

- As an initial evaluation, obtain a comprehensive set of laboratory studies including CBC and metabolic.
- Send the patient electively for audiologic studies and evaluation for a hearing aid for suspected age-related hearing loss.
- Send the patient electively to an otolaryngologist for evaluation and manual extraction of presumed cerumen impaction.
- Send the patient urgently to an otolaryngologist and initiate a course of corticosteroids (prednisone 1 mg/kg) in the interim

Correct Answer: D

Send the patient urgently to an otolaryngologist and initiate a course of corticosteroids (prednisone 1 mg/kg) in the interim.

Hearing loss is a very common condition encountered in all populations and becoming even more common as the US population ages.¹ Final diagnosis and therapy should generally involve consultation with the otolaryngology specialist but often it will be an internist, family practice, or emergency room physician who will field the initial complaint.

The purpose of the present case is to offer a logical yet uncomplicated initial approach to hearing loss management by way of final, specific diagnosis and definitive therapy. The patient will notice the hearing deficit in a variety of ways depending on the degree of hearing loss. There is a correlation of symptoms reasonably related to the decibels in hearing level (dB HL).¹ Thus, patients with mild hearing loss (25-40 dB HL) have difficulty hearing speech with background noise. Those with moderate hearing loss (40-60 dB HL) have difficulty with normal speech and are unable to hear low or mumbled tones. Those with severe hearing loss (61-80 dB HL) require amplification via increased volume or closer proximity to the sound/speaker.¹ Thus, the presented patient's degree of hearing loss could be categorized as between moderate and severe.

Differential diagnosis

The next steps breakdown the potentially broad differential diagnosis into digestible subtypes. There are three interre-

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CITATION:

Rubin RN. A health 64-year-old man with sudden hearing loss. *Consultant*. 2022;62(5):e27-e29. doi:10.25270/con.2022.05.00002

DISCLOSURES:

The author reports no relevant financial relationships.

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lated subset areas that can and should be evaluated in light of available history, exam, and routine office maneuvers, eg, the Rinne and Weber tests, as will be discussed below.

First, by history: determine the onset of the hearing loss. Has the condition been slowly progressive, as seen with noise exposure patients, often preceded by or accompanied with tinnitus? Could it be age-related otosclerosis (hearing loss related to aging)? Or has it been sudden (usually defined as literally occurring over 2 days), which suggests a cerumen impaction. Additionally, the cause could be idiopathic sensorineural hearing loss, which will be discussed below.

Second, clinicians should ascertain if the hearing loss is an event isolated to the auditory function or associated with other findings in the patient, such as vertigo, nausea, emesis, medication exposures, or diabetes complications of which can be hearing loss.

Finally, and very importantly, it should be determined whether the hearing loss is conductive, in which sound is being blocked somewhere along the anatomy of the ear, (eg, ear canal due to cerumen, tympanic membrane due to fluid effusion, and ossicles in the middle ear due to age-related otosclerosis). Alternatively, the hearing loss could be sensorineural wherein the defect resides in the cochlea and the VIII nerve itself as with acute idiopathic sensorineural hearing loss.^{1,2}

Conductive vs sensorineural loss can be screened using some of the earliest diagnostic testing to which we were introduced in medical school. Clinicians can rub fingers together and whisper into patients' ears to get an idea of hearing loss severity and whether it is symmetrical/bilateral or unilateral. Then with a tuning fork perform the Rinne test, which demonstrates loss of air conducted hearing more so than mastoid bone conducted sound indicating conducting loss. Then perform the Weber test with the tuning fork placed mid-forehead, establishing laterality or lack thereof and conductive vs sensorineural type of loss. Careful otoscope

exam will visualize cerumen impaction, inflammation, and/or fluid pathology of the tympanic membrane.

As previously stated, most patients will find their way to otolaryngology offices for a more thorough ear examination and audiometry. But it needs to be noted that, at the primary care level we are describing, there is no role for either routine laboratory evaluation (unless there are obvious clues present such as fever, earache) or imaging studies which are best left to the specialist! This makes answer A an incorrect choice. Additionally, the findings on office hearing tests – profound loss of all hearing in the right ear and distinct lateralization on Weber testing favoring the left ear are consistent with sensorineural causation. Additionally, office ear exam seemed negative for excess cerumen and TM pathology. The former makes Answer C incorrect. And finally the onset in the presented patient was sudden—literally within days. All of these findings are consistent with the serious diagnosis of sudden sensorineural hearing loss.

Sudden hearing loss is considered an otologic emergency³ yet will almost always be first encountered in the primary care setting. The term “sudden hearing loss” implies an onset of 1-3 days and a sensorineural etiology involving the cochlea and auditory nerve. Recent 2019 society guidelines recommend referral for audiometry confirmation within 7 to 14 days and, importantly, the initiation of corticosteroid therapy within 14 days.³ Thus Answer B is incorrect on three counts – the patient findings are not consistent with age related (progressive, chronic) hearing loss, elective evaluation is too slow for evaluation/confirmation and initial therapy for acute sensorineural hearing loss and hearing aids do not work.

The Answer

The correct answer is D: send the patient urgently to an otolaryngologist and initiate a course of corticosteroids (prednisone 1 mg/kg) in the interim. Initiation of therapy should be as quickly as possible with a guideline window of 14 days from on-

set.³ In my professional experience, there is no reason for a primary care physician to defer starting steroids (eg, prednisone 1 mg/kg) pending the ENT visit and evaluation as any minimal risk of a few days on steroids is outweighed by the risk of permanent hearing loss if there is too much delay.

Management and Treatment

Some brief data on general therapeutics for hearing loss need be discussed. Therapeutics is contingent upon diagnosis to be sure. When conductive causation is involved, clinicians should first treat what is causing impaired conduction. A good place to start is removal of cerumen impaction. A variety of techniques are available including cerumen-dissolving drops. I would also recommend the patient should always have an audiometry exam after cerumen removal for completeness of diagnosis, and mechanical removal by the ENT specialist seems the safest and most effective means to do this. If tympanic membrane lesions (eg, fluid) are the cause, antibiotics +/- drainage, again by the otolaryngologist, is indicated. For otosclerosis and age-related hearing loss, the core therapeutics involve amplification (eg, hearing aids), which are now more effective and varied than ever.^{1,4} They amplify the sounds into the air to compensate for the dBHL of the patient. The technology is so advanced that, as with generic over-the-counter medications, the time may be near for certain patients to buy an amplification device at their local pharmacy.¹

As for sensorineural hearing loss, management after the initial oral steroids includes intratympanic steroid injection, which diffuses larger doses directly into the cochlear/tympanic auditory nerve area, and trials of immunosuppressive medications (eg, rituximab and cyclophosphamide) when initial therapeutics fail.³ A newer option is cochlear implantation, wherein an electronic device is surgically placed to partially restore hearing via sound signals directly to the auditory nerve, bypassing the hair cells of the cochlea.⁵ Although general anesthesia is

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required for procedure, particularly in a geriatric patient, the good news is that it is an outpatient procedure.⁵

Patient Follow-Up

The otolaryngologist saw the patient the following morning and confirmed unilateral hearing loss. Although the cerumen amount was not excessive, the ear canals were cleaned with good observation of both tympanic membranes without fluid or pathology. Audiometric exams performed revealed normal hearing in the left ear and moderate hearing loss (50 dB HL) in the right ear, which was confirmed as sensorineural in nature. Oral steroids were continued. For completeness, magnetic resonance imaging studies of the petrous bone, mastoid, and cochlear areas were performed and were normal.

The patient was reevaluated at 1 week, and symptoms and audiometry were stable. The treatment plan is to complete a steroid treatment then taper. There will be 10-day audiometry evaluations with either intratympanic steroid injection or secondary therapies (eg, immunosuppressives) if there is any deterioration or lack of improvement.

What's the Take Home?

Hearing loss is a common condition, and most cases will be initially seen in the setting of primary care. A battery of readily available details of history and an exam, coupled with basic office procedures can quickly ascertain key facts resulting in sub-classification of hearing loss. Although there are some rather simple and direct causes of hearing loss, eg, cerumen impactions, most patients should be referred to an otolaryngologists office for more comprehensive examination, testing, treatment. Sudden sensorineural hearing loss (SSNHL) occurs within 1 to 3 days and has a therapeutic window of approximately 14 days for corticosteroid initiation and thus is an urgent, specialty consultation. Therapeutics of hearing loss in general has shared in the great technological advances in recent medicine. The keystone to effectively manage

conductive hearing loss not amenable to local ENT maneuvers is amplification with a wide variety of choices of hearing aids now available for sensorineural causation. The technique of cochlear implantation devices to bypass the damages cochlear hair cells is also a good option for its convenience and relative effectiveness.

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