# Sudden deafness

By **Douglas J Lanska MD MS MSPH** (Dr. Lanska of the Great Lakes VA Healthcare System has no relevant financial relationships to disclose.)

*Originally released September 10, 2001; last updated May 22, 2016; expires May 22, 2019*

## Introduction

This article includes discussion of sudden deafness and sudden hearing loss. The foregoing terms may include synonyms, similar disorders, variations in usage, and abbreviations.

### Overview

The author explains the clinical presentation, pathophysiology, diagnostic work-up, and management of sudden deafness. "Sudden" deafness is defined as sensorineural hearing loss of 30 decibels or more in at least 3 contiguous frequencies occurring over less than 3 days. The pathophysiology of sudden deafness is poorly understood. Various theories have been proposed, including those attributing sudden deafness to vascular insults, infectious (especially viral) agents, autoimmune or inflammatory mechanisms, or disruption of labyrinthine membranes. Despite extensive investigation, most cases remain idiopathic. Systemic steroids, or a combination of systemic and intratympanic steroids, are commonly recommended, but some employ intratympanic steroid therapy as a first-line therapy because systemic and transtympanic administration of corticosteroids has been found to result in similar clinical outcomes. Intratympanic steroid perfusion should be offered in patients with incomplete recovery from idiopathic sudden sensorineural hearing loss after failure of initial management, and when used as salvage therapy, intratympanic steroids can result in significant gains in hearing. The overall prognosis depends on the underlying etiology, but a high rate of spontaneous resolution occurs overall (ie, about two thirds of cases). For those who do not recover from idiopathic sudden deafness in their only hearing ear (ie, producing bilateral deafness), cochlear implantation can be considered as early as 3 months after initiating treatment of sudden deafness.

### Key points

- "Sudden" deafness is defined as sensorineural hearing loss of 30 decibels or more in at least 3 contiguous frequencies, occurring over less than 3 days.
- In patients with sudden sensorineural hearing loss, tinnitus is associated with worse high-frequency hearing loss, whereas aural fullness and pressure sensations are typically associated with low-frequency hearing loss.
- The clinical manifestations of ischemia of the inner ear can include unilateral deafness and tinnitus as well as acute vertigo, nausea and vomiting, imbalance, and canal paresis.
- The spectrum of clinical presentation of anterior inferior cerebellar artery (AICA) infarction includes ipsilateral hearing loss with or without tinnitus as well as a range of labyrinthine, brainstem, and cerebellar symptoms and signs.
- The spectrum of clinical presentation of superior cerebellar artery syndrome includes ipsilateral Horner syndrome, ipsilateral limb ataxia, contralateral sensorineural hearing loss (due to involvement of the lateral lemniscus carrying decussated ascending auditory information), contralateral superficial sensory loss, vertigo, nystagmus, nausea, and vomiting.
- Acute bilateral hearing impairment suggests vertebrobasilar occlusive disease, but hearing loss associated with vertebrobasilar insufficiency is most frequently unilateral.
- The blood supply to the inner ear is via the internal auditory artery (also called the labyrinthine artery), which typically originates from the anterior inferior cerebellar artery (AICA).
- Patients with unilateral idiopathic sudden sensorineural hearing loss should be evaluated for retrocochlear pathology (eg, acoustic neuroma) using magnetic resonance imaging, brainstem auditory evoked potentials, or audiometric follow-up.
- The overall prognosis depends on the underlying etiology, but a high rate of spontaneous resolution occurs overall (ie, about two thirds of cases).

- Management is complicated, as the underlying etiology is not known in most patients. A presumptive approach is generally employed, but no consensus exists concerning the management of sudden hearing loss.
- Systematic syntheses and metaanalyses have failed to support the use of corticosteroids for sudden deafness and, instead, have concluded that “systemic or intratympanic steroid administration does not have a significant treatment effect.”
- For those who do not recover from idiopathic sudden deafness in their only hearing ear (ie, producing bilateral deafness), cochlear implantation can be considered as early as 3 months after initiating treatment of sudden deafness.

#### Historical note and terminology

"Sudden" deafness is typically defined as sensorineural hearing loss of 30 decibels or more in at least 3 contiguous frequencies, occurring over less than 3 days. Some (explicitly or implicitly) consider the syndrome to apply only to "idiopathic" monophasic cases, but in this article such restrictions are not employed.

## Clinical manifestations

#### Presentation and course

By definition, the principal manifestation of sudden hearing loss is sensorineural hearing loss occurring over less than 3 days (Lanska 2014). It is generally understood to be a monophasic illness, but recurrences can occur with some etiologies (Lanska 2014). Depending on the etiology and on damage to associated structures, associated manifestations may include aural fullness or pressure, tinnitus, vertigo, nausea and vomiting, and various brainstem and cerebellar signs (Lanska 2014). In patients with sudden sensorineural hearing loss, tinnitus is associated with worse high-frequency hearing loss, whereas aural fullness and pressure sensations are typically associated with low-frequency hearing loss (Sakata et al 2008). Tinnitus and aural fullness improve with improvements in hearing (Ishida et al 2008).

Men and women are generally affected with equal frequency, although one study found a higher frequency among females than males (Lanska 2014; Nakashima et al 2014). The condition occurs most commonly in the fifth and sixth decades. For unknown reasons, the left ear is affected slightly more often than the right ear (Reiss and Reiss 2014). The putative cause is identified in about 10% to 15% of cases, and the remainder is considered idiopathic after evaluation. Only a small minority (approximately 1%) has an identified retrocochlear cause (eg, vestibular schwannoma, demyelinating disease, stroke) (Rauch 2008; Hellmann et al 2011).

Putative risk factors for sudden sensorineural hearing loss include cardiovascular risk factors (smoking, increased alcohol consumption), obstructive sleep apnea (in men only), and recent subclinical viral or toxoplasmosis infections (Kikidis et al 2011; Lin et al 2012; Sheu et al 2012).

Sudden deafness may be caused by ischemia of the cochlea or eighth nerve (Lanska 2014). Cochlear ischemia may occur in isolation, in conjunction with labyrinthine ischemia, or in conjunction with brainstem and cerebellar ischemia as a result of involvement of the anterior inferior cerebellar artery or the vertebrobasilar system (Lanska 2014). The constellation of clinical manifestations depends on the extent and distribution of the ischemia. The clinical manifestations of ischemia of the inner ear can include unilateral deafness and tinnitus as well as acute vertigo, nausea and vomiting, imbalance, and canal paresis (Millikan et al 1959; Millikan 1964; Fisher 1967; Gussen 1976; Millikan and Futrell 1990a; Millikan and Futrell 1990b; Oas and Baloh 1992; Kim et al 1999; Strupp et al 2000; Yamasoba et al 2001; Rambold et al 2005; Lanska 2014). Patients with involvement of the common cochlear artery may present with deafness and vestibular involvement limited to paresis of the posterior semicircular duct (Rambold et al 2005). In those with predominant auditory dysfunction, patients may present with sudden deafness (Fisher 1967; Watanabe et al 1994), sometimes accompanied by transient dizziness and intermittent tinnitus (Fisher 1967; Gussen 1976). Mitral valve prolapse, mitral leaflet thickening, mitral regurgitation, and left atrial enlargement are risk factors for “idiopathic” sudden sensorineural hearing loss, and presumably these associations reflect an increased risk of cochlear or eighth nerve ischemia (Vazquez et al 2008).

The spectrum of clinical presentation of AICA infarction includes ipsilateral hearing loss with or without tinnitus as well as a range of labyrinthine, brainstem, and cerebellar symptoms and signs (Lanska 2014). Other manifestations include

ipsilateral Horner syndrome (rare), skew deviation (rare), nystagmus, ipsilateral facial numbness, ipsilateral facial paresis, vertigo, dysarthria, vomiting, unsteadiness, ipsilateral hemiataxia, and contralateral loss of pain and temperature sensation on the limbs and body (Amarenco and Hauw 1990; Hinojosa and Kohut 1990; Oas and Baloh 1992; Amarenco et al 1993; Lee et al 2004; Lanska 2014). Occasionally, isolated vertigo or isolated auditory disturbance may occur as transient ischemic attacks preceding AICA-territory infarction, or with partial infarcts (Oas and Baloh 1992; Amarenco et al 1993; Lee and Cho 2004). Bilateral sudden deafness may occur as a prodrome of anterior inferior cerebellar artery-territory infarction in the presence of severe vertebrobasilar occlusive disease (Lee et al 2001; Toyoda et al 2002). Rarely, the internal auditory artery branches off the PICA (rather than AICA) and sudden unilateral deafness may, therefore, result from PICA infarction (eg, with vertebral artery dissection) (Raupp et al 2004; Lee 2008).

The spectrum of clinical presentation of superior cerebellar artery syndrome includes ipsilateral Horner syndrome, ipsilateral limb ataxia, contralateral sensorineural hearing loss (due to involvement of the lateral lemniscus carrying decussated ascending auditory information), contralateral superficial sensory loss, vertigo, nystagmus, nausea, and vomiting (Murakami et al 2005).

Hearing loss occurs in about one fifth of patients with vertebrobasilar insufficiency and vertigo (Yamasoba et al 2001). Deafness associated with vertebrobasilar insufficiency mainly involves the cochlea, rather than central auditory pathways (Yamasoba et al 2001; Lee and Baloh 2005). Tinnitus and vertigo are frequent accompaniments, as are a wide a range of brainstem and cerebellar symptoms and signs (Lee et al 2003; Sauvaget et al 2004).

Ischemia may also occur with vascular obstruction in the venules and capillaries, draining the inner ear, as occurs most commonly with conditions that produce marked serum hyperviscosity. The "hyperviscosity syndrome" includes a number of diverse clinical manifestations including headache, fatigue, vertigo, nystagmus, sudden or progressive hearing loss, visual disturbances, and mucosal hemorrhages (Nomura et al 1982; Andrews et al 1988). Ophthalmoscopic findings include markedly distended and tortuous ("sausage-shaped") retinal veins and retinal hemorrhages, similar to the pattern seen in retinal vein occlusion.

Ramsay-Hunt syndrome (herpes zoster oticus) may be associated with vesicles in the external auditory canal, burning pain in the ear, unilateral Bell palsy, unilateral hearing loss, tinnitus, vertigo, and transient spontaneous nystagmus.

With viral neurolabyrinthitis, autoimmune hearing loss, and Ménière syndrome, the clinical manifestations are primarily otologic, whereas hearing loss, tinnitus, vertigo, and spontaneous nystagmus are the predominant manifestations. No neurologic manifestations are present, apart from those attributable to the labyrinth and eighth nerve.

Viral neurolabyrinthitis may be part of a systemic viral illness or it may be an isolated viral infection of the labyrinth and eighth nerve. Many patients report an upper respiratory illness within 1 week or 2 weeks prior to the onset of symptoms. The manifestations are unilateral, and may include clinically evident aural or vestibular symptoms, or both (Hyden 1996). When hearing loss is incomplete, it is usually most severe at high frequencies. Some cases may develop posterior semicircular canal benign paroxysmal positional vertigo with preservation of lateral semicircular canal function (Karlberg et al 2000).

Autoimmune hearing loss is often fluctuating, sometimes slowly progressive, and occasionally sudden. It may begin on 1 side, but invariably becomes bilateral. It may be associated with vertigo, if the involvement is sufficiently rapid and asymmetric. Often, a history of polyarteritis, rheumatoid arthritis, ulcerative colitis, Crohn disease, or other autoimmune-mediated conditions is present. Systemic manifestations may include interstitial keratitis, arthritis, rash, or gastrointestinal symptoms.

Ménière syndrome is associated with fluctuating sensorineural hearing loss, subjective tinnitus, aural fullness, episodic vertigo, and horizontal or horizontal-rotatory nystagmus. Onset may occur fairly suddenly over seconds, or it may develop over minutes or hours. The duration of hearing loss is variable among patients with some patients having this symptom for hours, others for days or weeks, and others permanently. Even with hearing recovery with continued episodes, recovery is often less complete, resulting in a progressive hearing loss. Involvement is typically unilateral at onset, but may become bilateral.

Sudden deafness can occur as a consequence of head trauma, but other manifestations of such injuries typically predominate, especially in the acute period. Rarely, sudden deafness can occur as a relatively isolated phenomena

following modest trauma. For example, Lee and colleagues reported a case of sudden, bilateral deafness associated with bilateral pneumolabyrinth, without temporal bone fracture, after a fall (Lee et al 2012).

Also rarely, cases of sudden deafness can be due to sudden-onset cortical deafness from bilateral temporal lobe infarcts (Bahls et al 1988; Murray and Fields 2001; Leussink et al 2005). The auditory cortex is located in the posterior superior aspect of both temporal lobes, with the primary auditory cortex located in the transverse temporal gyri of Heschl. Cortical deafness may evolve to auditory agnosia (ie, impairment of the ability to interpret both verbal and nonverbal sounds even though the patient can hear them) (Bahls et al 1988) or word deafness (ie, impairment of the ability to understand speech) (Murray and Fields 2001).

**Prognosis and complications**

The overall prognosis depends on the underlying etiology, but a high rate of spontaneous resolution occurs overall (ie, about two thirds of cases) (Eisenman and Arts 2000; Yimtae et al 2001; Penido Nde et al 2005; Stahl and Cohen 2006; Lanska 2014). The rate of complete recovery and the recovery rate from profound hearing loss are significantly higher in children than in adults (Na et al 2014). Most patients show either initial rapid recovery or a gradual and slow recovery (Harada 1996), but the spontaneous recovery that occurs typically is within the first 2 weeks after onset (Mattox and Simmons 1977). Improvement in hearing levels tends to occur mostly in the low to mid frequencies and is better in those with preserved otoacoustic emissions (Ishida et al 2008). Those with initial rapid recovery have the best prognosis, with a smaller degree of hearing loss at the first examination, greater degree of hearing improvement, and smaller degree of residual hearing loss once stable (Harada 1996). Patients with upsloping or with lower or middle frequency hearing loss generally have a better prognosis (Eisenman and Arts 2000; Zadeh et al 2003). Putative negative prognostic factors include longer time since onset of symptoms before treatment, more severe hearing loss, flat or downsloping audiograms, tinnitus, vertigo or evidence of vestibular dysfunction by neuro-otological studies (eg, vestibular evoked myogenic potentials and caloric testing), high signals in the affected inner ear on 3D-FLAIR MRI, very young or very old age, elevated sedimentation rate, and associated diabetes (Mosnier et al 1998; Eisenman and Arts 2000; Zadeh et al 2003; Weng et al 2005; Korres et al 2011; Ryu et al 2011; Suzuki et al 2011), but not all reports agree (Fetterman et al 1996; Zadeh et al 2003; Rauch 2008). Variation between reports depends in part on the method of outcome assessment, variation in patient characteristics including degree of hearing loss, and variation in adjustment for degree of hearing loss when considering recovery or improvement (Eisenman and Arts 2000). Audiovestibular residua or late effects can also include tinnitus, dysequilibrium, benign paroxysmal positioning vertigo, and Ménière syndrome (Yoon et al 1990; Hyden 1996; Karlberg et al 2000; Rauch 2008; Carlsson et al 2011). Annoying tinnitus and residual vertigo are the strongest predictors of the negative effects on quality of life in patients with sudden deafness (Carlsson et al 2011).

In isolated inner ear infarction, the vertigo, nystagmus, and autonomic manifestations resolve over days to weeks, but deafness and canal paresis typically remain (Millikan and Futrell 1990a; Millikan and Futrell 1990b; Watanabe et al 1994; Kim et al 1999). If no brainstem symptoms develop and brain imaging is normal, the risk of recurrence or subsequent stroke is rare (Futrell 1990b; Millikan and Futrell 1990a; Kim et al 1999). Patients with labyrinthine ischemia due to vertebrobasilar insufficiency can have an overall good prognosis with anticoagulation or antiplatelet therapy (Fife et al 1994) or, rarely, with surgical correction of a rotational vertebral artery syndrome (Strupp et al 2000). However, patients with inner ear infarction combined with brainstem or cerebellar infarcts have a worse prognosis (Gomez et al 1996), particularly if associated with occlusive disease of the basilar artery (Ferbert et al 1990; Huang et al 1993).

Sudden sensorineural hearing loss is associated with an increased risk of stroke within 5 years of onset; in a cohort study, 13% of patients with sensorineural hearing loss had a stroke within 5 years, compared to 8% in controls, and after adjusting for other risk factors, those with sensorineural hearing loss had a risk of stroke 1.6 times greater than controls (Lin et al 2008).

Sudden sensorineural hearing loss can also result in later development of secondary endolymphatic hydrops, with a mean interval of 8 years (Cho et al 2013).

**Clinical vignette**

A 66-year-old diabetic man developed bilateral deafness, right-sided tinnitus, and vertigo, which he noticed on rising in the morning (Lee et al 2001). The vertigo resolved over the next day, but the hearing loss persisted. He had no visual

field loss, diplopia, Horner syndrome, dysarthria, dysphagia, weakness, ataxia, or sensory loss. A week later he presented with worsened hearing loss in the right ear, right-sided tinnitus, vertigo, nausea, and incoordination. Examination demonstrated a spontaneous left-beating, horizontal-rotatory nystagmus; right facial hypesthesia; right peripheral facial palsy; and right-sided dysmetria. Audiometry showed moderate bilateral sensorineural hearing loss (55 dB on the right and 45 dB on the left) with 100% speech discrimination. Electronystagmography showed no response to caloric stimulation in the right ear. MRI demonstrated hyperintense foci on T2-weighted images involving the right dorsolateral pons and both middle cerebellar peduncles. MRA demonstrated moderately severe stenosis of the distal right vertebral artery and the middle third of the basilar artery. An electrocardiogram and a transthoracic echocardiogram were normal. The patient was anticoagulated. The vertigo and nausea improved over several days, and the right-sided incoordination and gait abnormalities improved over several weeks. A follow-up audiogram demonstrated profound hearing loss in the right ear and 30 dB loss in the left.

## Biological basis

### Anatomic localization

Sudden deafness may occur with interruption of peripheral or central structures involved with hearing. It most commonly occurs with damage to the cochlea or eighth nerve. Cochlear or eighth nerve infarction may occur in isolation or with concomitant infarction of the labyrinth, brainstem, and cerebellum (Lanska 2014). Acute bilateral hearing impairment suggests vertebrobasilar occlusive disease (Huang et al 1993; Lee et al 2001; Kim et al 2013), but hearing loss associated with vertebrobasilar insufficiency is most frequently unilateral (Yamasoba et al 2001). Viral infections, inflammatory conditions, or autoimmune disorders that produce sudden hearing loss generally involve the labyrinth or eighth nerve. Ménière syndrome involves the labyrinth. Tumors (eg, acoustic neuromas) and meningitis that produce sudden hearing loss generally involve the eighth nerve. The frequent spontaneous recovery of hearing loss, and improvement with steroid therapy, suggest that in many cases there is a potentially reversible metabolic inner ear process disrupting the endocochlear potential, rather than immediate hair-cell degeneration (Sismanis 2005).

### Pathophysiology

The pathophysiology of sudden deafness is poorly understood, and it is likely that a variety of pathophysiologies can all produce sudden deafness. Various theories have been proposed, including those attributing sudden deafness to vascular insults, infectious (especially viral) agents, autoimmune or inflammatory mechanisms, or disruption of labyrinthine membranes (Lanska 2014). Despite extensive investigation, most cases remain idiopathic. Of the non-idiopathic cases, vascular and infectious etiologies are probably the most common, and the pathophysiology of these is best understood.

In the 1950s, a series of important experimental studies in animals established that cochlear function is extremely sensitive to anoxia (Fernandez 1955; Kimura and Perlman 1956a; Kimura and Perlman 1956b; Kimura and Perlman 1958a; Kimura and Perlman 1958b; Perlman and Kimura 1957; Perlman and Fernandez 1959). Obstruction of either the inferior cochlear vein or the internal auditory artery produces rapid loss of function; electrical activity deteriorates within 60 seconds of interruption of blood flow. Cochlear function may return to normal if blood flow is restored within 8 minutes of complete obstruction, but not if blood flow is interrupted for more than 30 minutes. External hair cells and the ganglion cells of the cochlea are particularly vulnerable to arterial obstruction, whereas the vestibular end organs are relatively resistant.

Venous drainage of the cochlea is via the vein of the cochlear aqueduct, which empties into the bulb of the jugular vein (Axelsson 1968; Mazzoni 1990). Venous obstruction produces early epithelial edema, followed by hemorrhage into the epithelium and perilymphatic and endolymphatic spaces, hair cell damage with secondary ganglion cell degeneration and, later, fibrosis and ossification. Labyrinthine ischemia, attributed to impaired venous drainage, most commonly results from hyperviscosity syndromes (Morganstern and Manace 1969; Ruben et al 1969; Nomura et al 1982; Andrews et al 1988; Saadah 1993). Increased blood viscosity produces obstruction in the labyrinthine venules and capillaries with decreased blood flow and ischemia of the inner ear, subsequent hemorrhage and, later, fibrosis and ossification (Kimura and Perlman 1956a; Kimura and Perlman 1956b). Similar changes occur in the eye producing visual disturbances, markedly distended and tortuous ("sausage-shaped") retinal veins, and retinal hemorrhages.

Arterial obstruction produces more rapid and severe damage than venous obstruction, whereas arterial obstruction produces histologically evident changes in hair cells within 30 minutes, followed in a few hours by extensive necrosis

including the supporting cells without hemorrhage and, ultimately, severe fibrosis and ossification by 6 months. Several patterns of end organ involvement occur with arterial obstruction and correspond to involvement of different arterial distributions within the inner ear.

The blood supply to the inner ear is via the internal auditory artery (also called the labyrinthine artery), which typically originates from the AICA (Axelsson 1968; Mazzoni 1969; Mazzoni 1990). The internal auditory artery divides into 2 main branches within the internal auditory canal: (1) the common cochlear artery and (2) the anterior vestibular artery (Axelsson 1968; Mazzoni 1990). The common cochlear artery divides into the main cochlear artery and the vestibulocochlear artery, which together supply the cochlea (Axelsson 1968; Mazzoni 1990). The internal auditory artery and its branches are end arteries, so even transient ischemia can cause permanent inner ear damage. The organ of Corti is particularly sensitive to ischemia (Sando et al 1982).

The AICA supplies the lateral pons, the middle cerebellar peduncle, the flocculus, the anterior part of the cerebellar lobules, and the inner ear (Amarenco and Hauw 1990). In patients with AICA territory infarction, the most consistently involved areas are the lateral pons and the middle cerebellar peduncle (Amarenco and Hauw 1990). As a result of the sharp angulation of the AICA at its origin, it is rarely occluded by emboli (Watanabe et al 1994); rather, most occlusions are due to either basilar artery plaques extending into the AICA or microatheroma of its origin (Amarenco et al 1993). Whether isolated or in combination with other symptoms and signs, deafness and vertigo can occur in AICA-distribution infarctions due to involvement of several central and peripheral sites, which include the labyrinth, the eighth nerve, the vestibular nuclei, the vestibulocerebellum, or some combination.

Ramsay-Hunt syndrome (herpes zoster oticus) is caused by reactivation of herpes zoster virus that had been dormant in the seventh and eighth nerves following previous infection with chicken pox. Pathologic findings in Ramsay-Hunt syndrome include perivascular, perineural, and intraneural round-cell infiltration of the seventh and eighth nerves. A large number of other viruses have been associated with viral neurolabyrinthitis, but herpes simplex virus type 1 has been particularly associated with sudden sensorineural hearing loss (Wilson 1986; Rabinstein et al 2001). Pathologic studies in patients with viral neurolabyrinthitis and sudden deafness have shown evidence of viral damage to the cochlea and auditory nerve, similar to that seen in patients with well-documented viral disorders (eg, mumps). Experimental animal studies have also demonstrated that several viruses can selectively infect the labyrinth and eighth nerve.

## Differential diagnosis

Sudden deafness can be an isolated symptom or the presenting symptom of a systemic disease.

Sudden hearing loss can be caused by a variety of disorders, including inner ear or eighth nerve ischemia, viral infection of the labyrinth or cochlear nerve, Ménière disease, intralabyrinthine membrane rupture, and autoimmune or inflammatory causes (Stokroos and Albers 1996; Eisenman and Arts 2000; Tucci 2000; Berrocal and Ramirez-Camacho 2002; Maruyoshi et al 2005; Rauch 2008; Heywood et al 2013; Yin et al 2013; Lanska 2014). Uncommon causes include retrocochlear masses, demyelinating disease, syphilis, Lyme disease, *Rickettsia felis* infection, meningitis, carcinomatous meningitis, Takayasu arteritis, perilymph fistula, toxins, and pregnancy (Healy and Wood 2004; Hengstman et al 2004; Maruyoshi et al 2005; Jeffs et al 2006; Rauch 2008; Ohno et al 2010; Hou and Wang 2011; Kenny et al 2011; Peeters et al 2013; Cassilde et al 2014; Goel et al 2014; Lanska 2014; Leite et al 2014; Nilsson et al 2014). Barotrauma, head injury (especially with temporal bone fracture, but also with inner ear concussion), and otologic surgery can also produce sudden heading loss, but these are rare and fairly obvious causes (Rozsasi et al 2003; Lee et al 2012).

The putative cause is identified in about 10% to 15% of cases, and the remainder is almost always unilateral and considered idiopathic after evaluation (Rauch 2008; Greco et al 2011). Only a small minority (approximately 1%) has an identified retrocochlear cause (eg, vestibular schwannoma, demyelinating disease, stroke) (Rauch 2008; Hellmann et al 2011). Rare bilateral cases may be due to malingering, conversion disorders, and neurologic causes (eg, vertebrobasilar occlusive disease, carcinomatous meningitis, paraneoplastic syndromes, encephalitis, meningitis), and polysubstance abuse or overdose (Koda et al 2008; Rauch 2008; Schweitzer et al 2011; Song et al 2014).

A variety of conditions can cause inner ear ischemia, including thromboemboli of the posterior circulation (Gur et al 2006), migraine (Lee et al 2002; Piovesan et al 2003), fat emboli (Jaffe 1970), thromboangiitis obliterans (Kirikae et al 1962), hyperlipidemia (Saadah 1993), macroglobulinemia (Ruben et al 1969; Nomura et al 1982), sickle cell disease

(Morgenstern and Manace 1969; Andrews et al 1988), leukemia (Andrews et al 1988); polycythemia vera (Andrews et al 1988); other causes of hypercoagulation or hyperviscosity (Jaffe 1970; Andrews et al 1988), and hypotension in otherwise healthy young adults (Pirodda et al 2001). Inner ear infarction occurs most commonly in the setting of thromboembolic disease of the AICA or the basilar artery (Gussen 1976; Hinojosa and Kohut 1990; Oas and Baloh 1992; Huang et al 1993; Kim et al 1999; Verghese and Morocz 1999; Strupp et al 2000; Lee et al 2002). Sudden deafness in AICA infarction is often due to cochlea dysfunction from ischemia (Lee et al 2002), but mixed central and peripheral vestibular dysfunction also occurs, making recognition of the components difficult. AICA territory infarction can be confused with posterior inferior cerebellar artery territory infarction (Wallenberg syndrome), because of shared signs including Horner syndrome, facial sensory impairment, vestibular signs, dysmetria, and contralateral impairment of pain and temperature sensation; however, severe facial paresis, hearing loss, and tinnitus are atypical for PICA territory infarctions (Lee 2008), and their presence should alert the clinician to AICA territory infarction (Amarenco and Hauw 1990).

Various infectious disorders have been implicated in occasional cases of sudden hearing loss. A large number of viruses have been clinically, epidemiologically, or pathologically associated with hearing loss (Huang et al 2009), but proof of viral etiology in individual cases is difficult to establish, with the exception of Ramsay-Hunt syndrome where the clinical features are fairly obvious and characteristic. Furthermore, the Henle-Koch postulates have not been satisfied for establishing a viral causation for sudden sensorineural hearing loss (Merchant et al 2008). Bacterial meningitis, syphilis, Lyme disease, and *Rickettsia felis* infection are among other infectious etiologies implicated in sudden hearing loss (Peeters et al 2013; Nilsson et al 2014). *Rickettsia felis* is an emergent pathogen belonging to transitional group rickettsiae (Pérez-Osorio et al 2008). First described in 1990, *R felis* infections can present with clinical signs similar to those of murine typhus and other febrile illnesses such as dengue fever, but like Lyme disease *R felis* infections can result in peripheral facial palsy and sudden deafness (Peeters et al 2013; Nilsson et al 2014). Cat fleas appear to be the most common vectors of *R felis* infections (Pérez-Osorio et al 2008).

Although acoustic neuroma is a relatively rare cause of sudden hearing loss (less than 2% of patients with this problem) (Eisenman and Arts 2000), sudden hearing loss may be the presenting symptom in 10% of patients to 15% of patients with acoustic neuroma (Berenholz et al 1992; Eisenman and Arts 2000). In such patients, hearing may recover to normal levels with steroid therapy and may falsely suggest an inflammatory or immunologically mediated cause (Berenholz et al 1992; Gaffney and McShane 1996). Other tumors can sometimes present with sudden hearing loss, and anecdotal reports include sudden hearing loss from a cochlear schwannoma (Shin et al 2008). Sudden hearing loss may also complicate meningitis (Eden and Cummings 1978; Damodaran et al 1996), but it is rarely the presenting or only manifestation, except, in rare cases of chronic infectious, leukemic, or carcinomatous meningitis.

Sudden sensorineural hearing loss may also occur from medications, including nonsteroidal anti-inflammatory drugs (McKinnon and Lassen 1998), aminoglycosides, and phosphodiesterase inhibitors (Barreto and Bahmad 2013). Rapid ototoxic hearing loss is much more common in patients with poor renal function.

Sudden sensorineural hearing loss may be the initial presentation of Ménière disease, especially in patients with low-frequency hearing loss, or as a late manifestation years after onset (Rauch 2008). In those patients in whom sudden sensorineural hearing loss is the initial presentation, further fluctuation in hearing with attacks of vertigo are likely to develop within 3 years (Rauch 2008).

Psychogenic sudden deafness can be identified by a discrepancy between behavioral hearing thresholds (eg, pure tone audiometry) and objective electrophysiologic examinations (eg, impedance audiometry, otoacoustic emissions, and brainstem auditory evoked responses) (Ban and Jin 2006). Psychogenic sudden deafness can be unilateral or bilateral and generally ranges in severity from moderate hearing loss to profound hearing loss, with the majority having severe to profound hearing loss on pure tone audiometry (Ban and Jin 2006). It is most commonly reported in teenagers and young adults (Ban and Jin 2006). Many have preexisting psychiatric illnesses or readily identified psychosocial stresses (Ban and Jin 2006).

## Diagnostic workup

Initial evaluation of patients with presumptive sudden sensorineural hearing loss should include careful history and examination to identify bilateral sudden hearing loss, recurrent sudden hearing loss, or focal neurologic findings (Stachler et al 2012). In addition evaluation should identify likely toxic, otologic, or systemic causes, including evaluation of Lyme titers and syphilis serologies. Audiograms should also be obtained to demonstrate the pattern and

severity of hearing loss, which are helpful prognostically. It is important to distinguish sensorineural and conductive patterns of hearing loss in patients with sudden hearing loss (Stachler et al 2012). Audiograms should be obtained before and within 24 to 48 hours after initiation of treatment. Current clinical practice guidelines specify that follow-up audiometric evaluation should be obtained within 6 months of diagnosis (Stachler et al 2012), and others suggest serial audiograms over the course of a year (eg, at 2, 6, and 12 months after onset) (Rauch 2008).

Patients with unilateral idiopathic sudden sensorineural hearing loss should be evaluated for retrocochlear pathology (eg, acoustic neuroma) using magnetic resonance imaging, brainstem auditory evoked potentials, or audiometric follow-up (Stachler et al 2012), whether or not apparent improvement or recovery is taking place (with or without steroid therapy) (Seltzer and Mark 1991; Weber et al 1997; Aarnisalo et al 2004; Penido Nde et al 2005; Ramos et al 2005; Rauch 2008). Cranial imaging is also important to exclude brainstem or cerebellar lesions (Weber et al 1997; Aarnisalo et al 2004), and MRI may identify a number of other pathologies (eg, vascular abnormalities and demyelination) (Schick et al 2001; Aarnisalo et al 2004; Ramos et al 2005), but MRI does not visualize the inner ear well enough to reliably identify infarction and is insensitive for abnormalities (eg, enhancement) associated with cochleitis or labyrinthitis (Stokroos et al 1998b; Schick et al 2001). Approximately half of practicing otolaryngologists routinely utilize MRI in evaluating patients with sudden sensorineural hearing loss (Coelho et al 2011), and many neurologists routinely use this technology in the initial evaluation of patients with sudden sensorineural deafness, despite a low yield of identified retrocochlear pathology, because of medicolegal concerns (Jiang et al 2011). Current guidelines recommend *against* use of CT of the head and brain in the initial evaluation of patients with sudden sensorineural hearing loss (Stachler et al 2012). In patients who cannot have an MRI, CT and brainstem auditory evoked potential studies should be considered, though these are less sensitive than MRI for detection of retrocochlear pathology (Rauch 2008).

As a result of a high rate of spontaneous recovery (approximately two thirds of cases), and because a large proportion of cases are ultimately considered to be idiopathic even after extensive evaluation, some have advocated a staged approach to diagnostic testing (Cowan and Chow 1988; Lanska 2014). Patients with likely systemic causes or clinically evident neurologic abnormalities should have diagnostic testing without delay. In patients without other clinical findings, further diagnostic evaluation can possibly be delayed for a month to see if spontaneous improvement occurs. Note, though, that improvement with steroids (in the absence of MRI or brainstem auditory evoked responses) can result in failure to identify important clinical conditions, including acoustic neuroma. If improvement does not occur or if other symptoms or signs develop, more extensive diagnostic testing is indicated and should include cranial imaging with magnetic resonance imaging.

Additional diagnostic studies can include imaging of cerebral vessels, brainstem auditory evoked potentials, electronystagmography with bithermal caloric irrigation, vestibular-evoked myogenic potentials, lumbar puncture, and various blood studies. Brainstem auditory evoked potentials may show absence of wave I or all waveforms, but may also show absence of wave I with delay of wave III and wave V, if dysfunction is also occurring in the retrocochlear eighth nerve and brainstem auditory nuclei and pathways (Verghese and Morocz 1999). Electronystagmography or videonystagmography with bithermal caloric testing may demonstrate ipsilateral horizontal canal paresis. Vestibular-evoked myogenic potential studies may show an absence of response on the affected side, supporting labyrinthine damage, particularly in patients with associated vertigo (Iwasaki et al 2005; Rambold et al 2005). Lumbar puncture should be performed (after cranial imaging) in immunocompromised patients and those with suspected chronic meningitis. In cases of clinically suspected sudden hearing loss resulting from hyperviscosity, the following blood studies can be considered: serum viscosity determination, complete blood count, syphilis serologies, sedimentation rate, serum protein, serum protein electrophoresis, and lipid studies. In cases of clinically suspected autoimmune inner ear disease, the following blood studies can be considered: sedimentation rate, rheumatoid factor, antinuclear antibody assay, antineutrophil cytoplasmic antibody assay, circulating immune complex levels, and urinalysis. Moreover, a number of more sophisticated immunologic tests of serological or cell-mediated reactivity to homologous and heterologous inner ear antigen extracts may have some utility, but are not routinely available (Harris and O'Driscoll 1996). Antibodies to a 68 kD heat shock protein (anti-hsp70) are not helpful (Samuelsson et al 2003). No correlation has been demonstrated between antibodies to inner ear antigens in patients with presumed autoimmune hearing loss and cochlear enhancement on MRI (Zavod et al 2000).

## Management

Management is complicated as the underlying etiology is not known in most patients. A presumptive approach is

generally employed, but no consensus exists concerning the management of sudden hearing loss (Haberkamp and Tanyeri 1999; Coelho et al 2011). Because of the lack of consensus, significant differences exist across specialists in the treatment of sudden sensorineural hearing loss (Coelho et al 2011).

Systemic corticosteroids, or a combination of systemic and intratympanic steroids, have been considered the "current standard treatment" and had been thought to be modestly effective in treating idiopathic sudden hearing loss (Wilson et al 1980; Moskowitz et al 1984; Haberkamp and Tanyeri 1999; Eisenman and Arts 2000; Alexiou et al 2001; Marzo 2005; Sismanis 2005; Conlin and Parnes 2007a; Conlin and Parnes 2007b; Rauch 2008; Arsian et al 2011; Coelho et al 2011; Dispenze et al 2011; Park et al 2011; Rauch et al 2011; Spear and Schwartz 2011; Gundogan et al 2013). Steroids have also been used in patients with sudden hearing loss and known recent viral infections, autoimmune disease (eg, Crohn disease or ulcerative colitis), or meningitis (Eden and Cummings 1978; Bachmeyer et al 1998). Most of the reported benefit of steroids was within the first 1 to 2 weeks after onset, which is also the typical timeframe for spontaneous recovery, and little, if any, benefit could be expected if initiated 4 weeks or longer after onset (Rauch 2008). Unfortunately, available trials of corticosteroids for sudden hearing loss are generally of poor quality and have shown inconsistent and contradictory results (Wei et al 2013; Crane et al 2015). Systematic syntheses and metaanalyses have failed to support the use of corticosteroids for sudden deafness and, instead, have concluded that "systemic or intratympanic steroid administration does not have a significant treatment effect" (Wei et al 2013; Crane et al 2015; Hultcrantz and Nosrati-Zarenoe 2015).

Some authorities recommend intratympanic dexamethasone only for subsequent or salvage treatment of idiopathic sudden sensorineural hearing loss (Park et al 2011; Garavello et al 2012). Steroids for salvage treatment of patients failing traditional therapy may have a beneficial treatment effect in a metaanalysis of available trials, although this is only a tentative conclusion because of the poor quality of component trials (Crane et al 2015). If administered, intratympanic steroids should be administered within 6 weeks of onset of hearing loss and should be reserved for patients with at least severe sensorineural hearing loss (ie, pure-tone average greater than 50 dB and speech discrimination less than 50%) (Marzo 2005) or as "salvage therapy" for patients who do not improve with oral corticosteroids (Ahn et al 2008; Rauch 2008; Coelho et al 2011; Wu et al 2011). According to current clinical practice guidelines, intratympanic steroid perfusion should be offered in patients with incomplete recovery from idiopathic sudden sensorineural hearing loss after failure of initial management (Stachler et al 2012), and when used as salvage therapy, intratympanic steroids can result in significant gains in hearing (Wu et al 2011).

Adding low molecular weight dextran to oral corticosteroids is not associated with greater hearing gain or better hearing outcome in patients with idiopathic sudden sensorineural hearing loss (Wang et al 2012).

In preliminary studies, topically applied recombinant human insulin-like growth factor 1 (IGF1) using gelatin hydrogels was associated with a significant improvement in pure-tone thresholds in patients considered refractory to systemic steroids (Nakagawa et al 2012; Nakagawa et al 2014). The major effects of this salvage therapy are thought to occur in the first 4 weeks after treatment.

Hyperbaric oxygen therapy is an option within 3 months of diagnosis of idiopathic sudden sensorineural hearing loss (Alimoglu et al 2011; Holy et al 2011; Stachler et al 2012; Cvorovic et al 2013), but clinicians should *not* routinely prescribe antivirals, thrombolytics, vasodilators, vasoactive substances, or antioxidants to patients with idiopathic sudden sensorineural hearing loss (Stachler et al 2012). Consequently, former "shotgun" approaches employing a battery of simultaneously administered treatments directed at common potential causes of sudden hearing loss should be discouraged. The efficacy of antiviral agents, anticoagulants, vasodilators, rheologic agents, free radical scavengers, ginkgo products, and other drugs is unproved in patients with idiopathic sudden hearing loss (Kanzaki et al 2003; Conlin and Parnes 2007b; Rauch 2008; Stachler et al 2012); most studies have been uncontrolled trials, and results are not clearly different than the natural history of this condition. Surgery is rarely indicated, except possibly in cases where clear evidence of perilymphatic fistula exists or to manage associated problems (eg, facial palsy in Ramsay-Hunt syndrome).

Patients with identified etiologies for sudden sensorineural hearing loss may require targeted specific therapies. For example, patients with Ramsay-Hunt syndrome should be treated with acyclovir (1 gm daily for 10 days), but available data do not suggest a benefit of antiviral agents in clinically diagnosed viral neurolabyrinthitis (Stokroos et al 1998a). Anecdotal evidence suggests that infliximab may be helpful in some cases of sudden deafness due to autoimmune inner ear disease (Heywood et al 2013). In addition, psychotherapy has been employed successfully in some patients with psychogenic sudden deafness (Ban and Jin 2006).

Associated vertigo and the concomitant nausea and vomiting should be treated symptomatically with medications; vestibular rehabilitation should be begun early (Lanska 2005). For those who do not recover from idiopathic sudden deafness in their only hearing ear (ie, producing bilateral deafness), cochlear implantation can be considered as early as 3 months after initiating treatment of sudden deafness (Lee et al 2010). Cochlear implantation in unilateral sudden hearing loss with a normal functioning contralateral ear may also prove to be an effective therapy (Blasco and Redleaf 2014). Available data suggest that subjective tinnitus, speech discrimination, sound localization, and speech comprehension are improved by cochlear implantation in selected patients.

A multidisciplinary rehabilitation approach involving audiological may be necessary to help patients cope with the complex issues associated with sudden deafness (Carlsson et al 2011).

## References cited


Aarnisalo AA, Suoranta H, Ylikoski J. Magnetic resonance imaging findings in the auditory pathway of patients with sudden deafness. Otol Neurotol 2004;25:245-9. PMID 15129100

Ahn JH, Han MW, Kim JH, Chung JW, Yoon TH. Therapeutic effectiveness over time of intratympanic dexamethasone as salvage treatment of sudden deafness. Acta Otolaryngol 2008a;128:128-31. PMID 17851916

Alexiou C, Arnold W, Fausner C, et al. Sudden sensorineural hearing loss: does application of glucocorticoids make sense. Arch Otolaryngol Head Neck Surg 2001;127:253-8. PMID 11255468

Alimoglu Y, Inci E, Edizer DT, Ozdilek A, Aslan M. Efficacy comparison of oral steroid, intratympanic steroid, hyperbaric oxygen and oral steroid + hyperbaric oxygen treatments in idiopathic sudden sensorineural hearing loss cases. Eur Arch Otorhinolaryngol 2011;268(12):1735-41. PMID 21431435

Amarenco P, Hauw JJ. Cerebellar infarction in the territory of the anterior and inferior cerebellar artery: a clinicopathologic study of 20 cases. Brain 1990;113:139-55. PMID 2302529

Amarenco P, Rosengart A, DeWitt LD, Pessin MS, Caplan LR. Anterior inferior cerebellar artery territory infarcts: mechanisms and clinical features. Arch Neurol 1993;50:154-61. PMID 8431134

Andrews JC, Hoover LA, Lee RS, Honrubia V. Vertigo in the hyperviscosity syndrome. Otolaryngol Head Neck Surg 1988;98:144-9. PMID 3128757

Axelsson A. The vascular anatomy of the cochlea in the guinea pig and in man. Acta Otolaryngologica 1968;(Suppl):243:1-134.

Bachmeyer C, Leclerc-Landgraf N, Laurette F, et al. Acute autoimmune sensorineural hearing loss associated with Crohn's disease. Am J Gastroenterol 1998;93:2565-7. PMID 9860428

Bahls FH, Chatrian GE, Mesher RA, Sumi SM, Ruff RL. A case of persistent cortical deafness: clinical, neurophysiologic, and neuropathologic observations. Neurology 1988;38:1490-3. PMID 3412601

Ban JH, Jin SM. A clinical analysis of psychogenic sudden deafness. Otolaryngol Head Neck Surg 2006;134:970-4. PMID 16730540

Barreto MA, Bahmad F Jr. Phosphodiesterase type 5 inhibitors and sudden sensorineural hearing loss. Braz J Otorhinolaryngol 2013;79(6):727-33. PMID 24474485

Berenholz LP, Eriksen C, Hirsch FA. Recovery from repeated sudden hearing loss with corticosteroid use in the presence of an acoustic neuroma. Ann Otol Rhinol Laryngol 1992;101:827-31. PMID 1416637

Berrocal JR, Ramirez-Camacho R. Sudden sensorineural hearing loss: supporting the immunologic theory. Ann Otol Rhinol Laryngol 2002;111:989-97. PMID 12450172

Blasco MA, Redleaf MI. Cochlear implantation in unilateral sudden deafness improves tinnitus and speech comprehension: meta-analysis and systematic review. Otol Neurotol 2014;35(8):1426-32. PMID 24786540

Carlsson PI, Hall M, Lind KJ, Danermark B. Quality of life, psychosocial consequences, and audiological rehabilitation

after sudden sensorineural hearing loss. Int J Audiol 2011;50(2):139-44. PMID 21265640

Cassilde AL, Barnaud G, Baccar S, Mortier E. Sudden-onset bilateral deafness revealing early neurosyphilis. Eur Ann Otorhinolaryngol Head Neck Dis 2014;131(6):389-91. PMID 24880727

Cho TY, Cheng PW, Young YH. Secondary endolymphatic hydrops after sudden deafness. Acta Otolaryngol 2013;133(10):1040-6. PMID 24032569

Coelho DH, Thacker LR, Hsu DW. Variability in the management of idiopathic sudden sensorineural hearing loss. Otolaryngol Head Neck Surg 2011;145(5):813-7. PMID 21690271

Conlin AE, Parnes LS. Treatment of sudden sensorineural hearing loss: I. A systematic review. Arch Otolaryngol Head Neck Surg 2007a;133:573-81. PMID 17576908

Conlin AE, Parnes LS. Treatment of sudden sensorineural hearing loss: II. A meta-analysis. Arch Otolaryngol Head Neck Surg 2007b;133:582-6. PMID 17576909

Cowan PF, Chow JM. Sudden sensorineural hearing loss. Am Fam Physician 1988;37:207-10. PMID 3348121

Crane RA, Camilon M, Nguyen S, Meyer TA. Steroids for treatment of sudden sensorineural hearing loss: a meta-analysis of randomized controlled trials. Laryngoscope 2015;125(1):209-17. PMID 25045896

Cvorovic L, Jovanovic MB, Milutinovic Z, Arsovic N, Djeric D. Randomized prospective trial of hyperbaric oxygen therapy and intratympanic steroid injection as salvage treatment of sudden sensorineural hearing loss. Otol Neurotol 2013;34(6):1021-6. PMID 23820795

Damodaran A, Aneja S, Malhotra VL, et al. Sensorineural hearing loss following acute bacterial meningitis - a prospective evaluation. Indian Pediatr 1996;33:763-6. PMID 9057404

Eden AR, Cummings FR. Sudden bilateral hearing loss and meningitis in adults. J Otolaryngol 1978;7:304-9. PMID 691097

Eisenman DJ, Arts HA. Effectiveness of treatment for sudden sensorineural hearing loss. Arch Otolaryngol Head Neck Surg 2000;126:1161-4. PMID 10979137

Ferbert A, Bruckman H, Drummen R. Clinical features of proven basilar artery occlusion. Stroke 1990;21:1135-42. PMID 2389292

Fernandez C. The effect of oxygen lack on cochlear potentials. Ann Otol Rhinol Laryngol 1955;64:1193-203. PMID 13283493

Fetterman BL, Saunders JE, Luxford WM. Prognosis and treatment of sudden sensorineural hearing loss. Am J Otol 1996;17:529-36. PMID 8841697

Fife TD, Baloh RW, Duckwiler GR. Isolated dizziness in vertebrobasilar insufficiency: clinical features, angiography, and follow-up. J Stroke Cerebrovasc Dis 1994;4:4-12. PMID 26487528

Fisher CM. Vertigo in cerebrovascular disease. Arch Otolaryngol 1967;85:529-34. PMID 6023709

Gaffney RJ, McShane DP. Bilateral acoustic neurofibromatosis camouflaged by corticosteroid treatment of sudden sensorineural hearing loss. Ir J Med Sci 1996;165:151-2. PMID 8824013

Garavello W, Galluzzi F, Gaini RM, Zanetti D. Intratympanic steroid treatment for sudden deafness: a meta-analysis of randomized controlled trials. Otol Neurotol 2012;33(5):724-9. PMID 22699982

Goel A, Sharma S, Aneja S. Sudden onset permanent deafness as an early complication of bacterial meningitis. Indian J Pediatr 2014;81(6):625. PMID 23783768

Gomez CR, Cruz-Flores S, Malkoff MD, Sauer CM, Burch CM. Isolated vertigo as a manifestation of vertebrobasilar ischemia. Neurology 1996;47:94-7. PMID 8710132

Greco A, Fusconi M, Gallo A, Marinelli C, Macri GF, De Vincentiis M. Sudden sensorineural hearing loss: an autoimmune disease. Autoimmun Rev. 2011;10(12):756-61. PMID 21619944

Gundogan O, Pinar E, Imre A, Ozturkcan S, Cokmez O, Yigiter AC. Therapeutic efficacy of the combination of intratympanic methylprednisolone and oral steroid for idiopathic sudden deafness. Otolaryngol Head Neck Surg 2013;149(5):753-8. PMID 23959817

Gur C, Lalazar G, Raphaeli G, Gilon D, Ben-Chetrit E. Mitral stenosis presenting with acute hearing loss. PLoS Med 2006;3(6):e233. PMID 16768545

Gussen R. Sudden deafness of vascular origin: a human temporal bone study. Ann Otol 1976;85:94-100. PMID 1259320

Haberkamp TJ, Tanyeri HM. Management of idiopathic sudden sensorineural hearing loss. Am J Otol 1999;20:587-92. PMID 10503580

Harada T. Patterns of hearing recovery in idiopathic sudden sensorineural hearing loss. Br J Audiol 1996;30:363-7. PMID 8985560

Harris JP, O'Driscoll K. Autoimmune inner ear disease. In: Baloh RW, Halmagyi GM. Disorders of the vestibular system. New York: Oxford University Press, 1996:374-80.

Healy DG, Wood NW. Clinical picture of bilateral vestibular schwannomas, sudden bilateral hearing loss, and aviation. Neurology 2004;62:933. PMID 15365156

Hellmann MA, Steiner I, Mosberg-Galili R. Sudden sensorineural hearing loss in multiple sclerosis: clinical course and possible pathogenesis. Acta Neurol Scand 2011;124(4):245-9. PMID 21198448

Hengstman GJD, Schelhaas HJ, Zwarts MJ. Auditory dysfunction in chronic inflammatory demyelinating polyradiculopathy. Neurology 2004;62:1446-8. PMID 15111702

Heywood RL, Hadavi S, Donnelly S, Patel N. Infliximab for autoimmune inner ear disease: case report and literature review. J Laryngol Otol 2013;127(11):1145-7. PMID 24125068

Hinojosa R, Kohut RI. Clinical diagnosis of anterior inferior cerebellar artery thrombosis: autopsy and temporal bone histopathologic study. Ann Otol Rhinol Laryngol 1990;99:261-72. PMID 2327695

Holy R, Navara M, Dosel P, Fundova P, Prazenica P, Hahn A. Hyperbaric oxygen therapy in idiopathic sudden sensorineural hearing loss (ISSNHL) in association with combined treatment. Undersea Hyperb Med 2011;38(2):137-42. PMID 21510273

Hou ZQ, Wang QJ. A new disease: pregnancy-induced sudden sensorineural hearing loss. Acta Otolaryngol 2011;131(7):779-86. PMID 21426273

Huang CC, Lin WB, Chang PH, Chan KC, Lee TJ. Sudden deafness as a presenting symptom of chronic hepatitis B with acute exacerbation. Otolaryngol Head Neck Surg 2009;141(5):659-60. PMID 19861211

Huang MH, Huang CC, Ryu SJ, Chu NS. Sudden bilateral hearing impairment in vertebrobasilar occlusive disease. Stroke 1993;24:132-7. PMID 8418537

Hultcrantz E, Nosrati-Zarenoe R. Corticosteroid treatment of idiopathic sudden sensorineural hearing loss: analysis of an RCT and material drawn from the Swedish national database. Eur Arch Otorhinolaryngol 2015;272(11):3169-75. PMID 25351498

Hyden D. Mumps labyrinthitis, endolymphatic hydrops and sudden deafness in succession in the same ear. ORL J Otorhinolaryngol Relat Spec 1996;58:338-42. PMID 8958544

Ishida IM, Sugiura M, Teranishi M, Katayama N, Nakashima T. Otoacoustic emissions, ear fullness and tinnitus in the recovery course of sudden deafness. Auris Nasus Larynx 2008;35:41-6. PMID 17904320

Iwasaki S, Takai Y, Ozeki H, Ito K, Karino S, Murofushi T. Extent of lesions in idiopathic sudden hearing loss with vertigo:

study using click and galvanic vestibular evoked myogenic potentials. Arch Otolaryngol Head Neck Surg 2005;131:857-62. PMID 16230586

Jaffe BF. Sudden deafness - a local manifestation of systemic disorders: fat emboli, hypercoagulation and infections. Laryngoscope 1970;80:788-801. PMID 5452517

Jeffs GJ, Lee GY, Wong GT. Leptomeningeal carcinomatosis: an unusual cause of sudden onset bilateral sensorineural hearing loss. J Clin Neurosci 2006;13:116-8. PMID 16410210

Jiang ZY, Mhoon E, Saadia-Redleaf M. Medicolegal concerns among neurotologists in ordering MRIs for idiopathic sensorineural hearing loss and asymmetric sensorineural hearing loss. Otol Neurotol 2011;32(3):403-5. PMID 21358454

Kanzaki J, Inoue Y, Ogawa K, et al. Effect of single-drug treatment on idiopathic sudden sensorineural hearing loss. Auris Nasus Larynx 2003;30(2):123-7. PMID 12753981

Karlberg M, Halmagyi GM, Buttner U, Yavor RA. Sudden unilateral hearing loss with simultaneous ipsilateral posterior semicircular canal benign paroxysmal positional vertigo: a variant of vestibulo-cochlear neurolabyrinthitis. Arch Otolaryngol Head Neck Surg 2000;126:1024-9. PMID 10922239

Kenny R, Patil N, Considine N. Sudden (reversible) sensorineural hearing loss in pregnancy. Ir J Med Sci 2011;180(1):79-84. PMID 20665123

Kikidis D, Nikolopoulos TP, Kampessis G, Stamatiou G, Chrysovergis A. Sudden sensorineural hearing loss: subclinical viral and toxoplasmosis infections as aetiology and how they alter the clinical course. ORL J Otorhinolaryngol Relat Spec 2011;73(2):110-5. PMID 21389742

Kim JS, Lopez I, DiPatre PL, et al. Internal auditory artery infarction: clinicopathologic correlation. Neurology 1999;52:40-4. PMID 9921846

Kim E, Son MK, Kang CK, Lee YB. Vertebrobasilar occlusion presenting as sudden isolated bilateral sensorineural hearing loss: case report. J Cerebrovasc Endovasc Neurosurg 2013;15(3):225-8. PMID 24167804

Kimura R, Perlman HB. Extensive venous obstruction of the labyrinth. A. Cochlear changes. Ann Otol Rhinol Laryngol 1956a;61:332-51. PMID 13355160

Kimura R, Perlman HB. Extensive venous obstruction of the labyrinth. B. Vestibular changes. Ann Otol Rhinol Laryngol 1956b;61:332-51. PMID 13363222

Kimura R, Perlman HB. Arterial obstruction of the labyrinth: part I. cochlear changes. Ann Otol Rhinol Laryngol 1958a;67:5-24. PMID 13521620

Kimura R, Perlman HB. Arterial obstruction of the labyrinth: part II. vestibular changes. Ann Otol Rhinol Laryngol 1958b;67:25-40. PMID 13521621

Kirikae I, Nomura Y, Shitara T, Kobayashi T. Sudden deafness due to Buerger's disease. Arch Otolaryngol 1962;75:502-5. PMID 14456277

Koda H, Kimura Y, Iino Y, Eishi Y, Murakami Y, Kitamura K. Bilateral sudden deafness caused by diffuse metastatic leptomeningeal carcinomatosis. Otol Neurotol 2008;29:727-9. PMID 18451746

Korres S, Stamatiou GA, Gkoritsa E, Riga M, Xenelis J. Prognosis of patients with idiopathic sudden hearing loss: role of vestibular assessment. J Laryngol Otol 2011;125(3):251-7. PMID 21054906

Lanska DJ. Disorders of the special senses in the elderly. In: Nair A, Sabbagh M, editors. Geriatric neurology. Chichester, West Sussex, UK: John Wiley & Sons, 2014:396-459.

Lanska DJ. Vertigo and other forms of dizziness. In: Corey-Bloom J, editor. Adult neurology. 2nd edition. Oxford, England: Blackwell Publishing Ltd., 2005:85-101.**.

Lee H. Sudden deafness related to posterior circulation infarction in the territory of the nonanterior inferior cerebellar artery: frequency, origin, and vascular topographical pattern. Eur Neurol 2008;59:302-6. PMID 18408371

Lee H, Ahn BH, Baloh RW. Sudden deafness with vertigo as a sole manifestation of anterior inferior cerebellar artery infarction. J Neurol Sci 2004;222:105-7. PMID 15240204

Lee H, Baloh RW. Sudden deafness in vertebrobasilar ischemia: clinical features, vascular topographical patterns and long-term outcome. J Neurol Sci 2005;228:99-104. PMID 15607217

Lee H, Cho YW. Auditory disturbance as a prodrome of anterior inferior cerebellar artery infarction. J Neurol Neurosurg Psychiatry 2004;74:1644-8. PMID 14638883

Lee H, Sohn SI, Jung DK, et al. Sudden deafness and anterior inferior cerebellar artery infarction. Stroke 2002;33:2807-12. PMID 12468774

Lee H, Whitman GT, Lim JG, Lee SD, Park YC. Bilateral sudden deafness as a prodrome of anterior inferior cerebellar artery infarction. Arch Neurol 2001;58:1287-9. PMID 11493170

Lee EJ, Yang YS, Yoon YJ. Case of bilateral pneumolabyrinth presenting as sudden, bilateral deafness, without temporal bone fracture, after a fall. J Laryngol Otol 2012;126(7):717-20. PMID 22583781

Lee H, Yi HA, Baloh RW. Sudden bilateral simultaneous deafness with vertigo as a sole manifestation of vertebrobasilar insufficiency. J Neurol Neurosurg Psychiatry 2003;74:539-41. PMID 12640087

Lee SS, Cho HH, Jang CH, Cho YB. Fate of sudden deafness occurring in the only hearing ear: outcomes and timing to consider cochlear implantation. J Korean Med Sci 2010;25(2):283-6. PMID 20119584

Leite HF, Leite Jda C, Melo MH, Vasconcelos CC, Alvarenga RM. Deafness in patients with multiple sclerosis. Audiol Neurootol 2014;19(4):261-6. PMID 25170548

Leussink V, Andermann P, Reiners K, Shehata-Dieter W, Gunther-Lengsfeld T, Naumann M. Sudden deafness from stroke. Neurology 2005;64:1817-8. PMID 15911827

Lin HC, Chao PZ, Lee HC. Sudden sensorineural hearing loss increases the risk of stroke: a 5-year follow-up study. Stroke 2008;39:2744-8. PMID 18583554

Lin RJ, Krall R, Westerberg BD, Chadha NK, Chau JK. Systematic review and meta-analysis of the risk factors for sudden sensorineural hearing loss in adults. Laryngoscope 2012;122(3):624-35. PMID 22252719

Maruyoshi H, Toyama K, Kojima S, et al. Sensorineural hearing loss combined with Takayasu's arteritis. Intern Med 2005;44:124-8. PMID 15750272

Marzo SJ. Intratympanic steroid perfusion for sudden sensorineural hearing loss. Arch Otolaryngol Head Neck Surg 2005;131:730-2. PMID 16103308

Mattox DE, Simmons FB. Natural history of sudden sensorineural hearing loss. Ann Otol Rhinol Laryngol 1977;86:463-80. PMID 889223

Mazzoni A. Internal auditory canal arterial relations at the porus acusticus. Ann Otol Rhinol Laryngol 1969;78:797-814. PMID 5799404

Mazzoni A. The vascular anatomy of the vestibular labyrinth in man. Acta Otolaryngologica 1990;472:1-83. PMID 2239254

McKinnon BJ, Lassen LF. Naproxen-associated sudden sensorineural hearing loss. Mil Med 1998;163:792-3. PMID 9819544

Merchant SN, Durand ML, Adams JC. Sudden deafness: is it viral. ORL J Otorhinolaryngol Relat Spec 2008;70:52-60. PMID 18235206

Millikan C. Neurology: its role in ophthalmology and otolaryngology. Trans Am Acad Ophthalmol Otolaryngol 1964;68:208-17. PMID 14130928

Millikan C, Futrell N. Occlusion of the internal auditory artery. Ann Neurol 1990a;28:258.

Millikan C, Futrell N. Vertigo of vascular origin. Arch Neurol 1990b;47:12-3. PMID 2294887

Millikan CH, Siekert RG, Whisnant JP. The syndrome of occlusion of the labyrinthine division of the internal auditory artery. Trans Am Neurol Assoc 1959;11. PMID 14422630

Morganstern KM, Manace ED. Temporal bone histopathology in sickle cell disease. Laryngoscope 1969;79:2172-80. PMID 5362685

Moskowitz D, Lee KJ, Smith HW. Steroid use in idiopathic sudden sensorineural hearing loss. Laryngoscope 1984;94:664-6. PMID 6717224

Mosnier I, Bouccara D, Atassi-Dumont M, Sterkers O. Treatments of idiopathic sudden sensorineural hearing loss: retrospective study of 144 cases [Article in French]. Rev Laryngol Otol Rhinol (Bord) 1998;119(2):119-28. PMID 9770055

Murakami T, Ono Y, Akagi N, et al. A case of superior cerebellar artery syndrome with contralateral hearing loss at onset. J Neurol Neurosurg Psychiatry 2005;76:1744-5. PMID 16291913

Murray A, Fields MJ. Word deafness presenting as a sudden hearing loss. Int J Clin Pract 2001;55(6):420-1. PMID 11501237

Na SY, Kim MG, Hong SM, Chung JH, Kang HM, Yeo SG. Comparison of sudden deafness in adults and children. Clin Exp Otorhinolaryngol 2014;7(3):165-9. PMID 25177430

Nakagawa T, Kumakawa K, Usami S, et al. A randomized controlled clinical trial of topical insulin-like growth factor-1 therapy for sudden deafness refractory to systemic corticosteroid treatment. BMC Med 2014;12:219. PMID 25406953

Nakagawa T, Ogino-Nishimura E, Hiraumi H, Sakamoto T, Yamamoto N, Ito J. Audiometric outcomes of topical IGF1 treatment for sudden deafness refractory to systemic steroids. Otol Neurotol 2012;33(6):941-6. PMID 22772021

Nakashima T, Sato H, Gyo K, et al. Idiopathic sudden sensorineural hearing loss in Japan. Acta Otolaryngol 2014;134(11):1158-63. PMID 25315915

Nilsson K, Wallménius K, Hartwig S, Norlander T, Påhlson C. Bell's palsy and sudden deafness associated with Rickettsia spp. infection in Sweden. A retrospective and prospective serological survey including PCR findings. Eur J Neurol 2014;21(2):206-14. PMID 23790098

Nomura Y, Mori S, Tsuchida M, Sakurai T. Deafness in cryoglobulinemia. Ann Otol Rhinol Laryngol 1982;91:250-5. PMID 7092044

Oas JG, Baloh RW. Vertigo and the anterior inferior cerebellar artery syndrome. Neurology 1992;42:2274-9. PMID 1461378

Ohno T, Yokoyama Y, Aihara R, Mochiki E, Asao T, Kuwano H. Sudden bilateral sensorineural hearing loss as the presenting symptom of meningeal carcinomatosis of gastric cancer: report of a case. Surg Today 2010;40(6):561-5. PMID 20496139

Park MK, Lee CK, Park KH, Lee JD, Lee CG, Lee BD. Simultaneous versus subsequent intratympanic dexamethasone for idiopathic sudden sensorineural hearing loss. Otolaryngol Head Neck Surg 2011;145(6):1016-21. PMID 21817157

Peeters N, van der Kolk BY, Thijsen SF, Colnot DR. Lyme disease associated with sudden sensorineural hearing loss: case report and literature review. Otol Neurotol 2013;34(5):832-7. PMID 23303170

Penido Nde O, Ramos HV, Barros FA, Cruz OL, Toledo RN. Clinical, etiological and progression factors of hearing in sudden deafness. Braz J Otorhinolaryngol 2005;71:633-8. PMID 16612525

Pérez-Osorio CE, Zavala-Velázquez JE, Arias León JJ, Zavala-Castro JE. Rickettsia felis as emergent global threat for humans. Emerg Infect Dis 2008;14(7):1019-23. PMID 18598619

Perlman HB, Fernandez C. Experiments on temporary obstruction of the internal auditory artery. Laryngoscope 1959;69:591-613. PMID 13673604

Perlman HB, Kimura R. Experimental obstruction of the venous drainage and arterial supply of the inner ear. Ann Otol Rhinol Laryngol 1957;66:537-47. PMID 13459249

Piovesan EJ, Kowacs PA, Werneck LC, Siow C. Oscillucusis and sudden deafness in a migraine patient. Arq Neuropsiquiatr 2003;61(3B):848-50. PMID 14595494

Pirodda A, Ferri GG, Modungo GC, Borghi C. Systemic hypotension and the development of acute sensorineural hearing loss in young healthy subjects. Arch Otolaryngol Head Neck Surg 2001;127:1049-52. PMID 11556851

Rabinstein A, Jerry J, Saraf-Lavi E, Sklar E, Bradley WG. Sudden sensorineural hearing loss associated with Herpes simplex virus type 1 infection. Neurology 2001;56:571-2. PMID 11222814

Rambold H, Boenki G, Stritzke G, Wisst F, Neppert B, Helmchen C. Differential vestibular dysfunction in sudden unilateral heading loss. Neurology 2005;64:148-51. PMID 15642923

Ramos HV, Barros FA, Yamashita H, Penido Nde O, Souza AC, Yamaoka WY. Magnetic resonance imaging in sudden deafness. Braz J Otorhinolaryngol 2005;71(4):422-6. PMID 16446954

Rauch SD. Idiopathic sudden sensorineural hearing loss. N Engl J Med 2008;359:833-40. PMID 18716300

Rauch SD, Halpin CF, Antonelli PJ, Babu S, Carey JP, et al. Oral vs intratympanic corticosteroid therapy for idiopathic sudden sensorineural hearing loss: a randomized trial. JAMA 2011;305(20):2071-9. PMID 21610239

Raupp SF, Jellema K, Sluzewski M, de Kort PL, Visser LH. Sudden unilateral deafness due to a right vertebral artery dissection. Neurology 2004;62:1442. PMID 15111698

Reiss M, Reiss G. Laterality of sudden sensorineural hearing loss. Ear Nose Throat J 2014;93(8):318-20. PMID 25181661

Rozsasi A, Sigg O, Keck T. Persistent inner ear injury after diving. Otol Neurotol 2003;24(2):195-200. PMID 12621331

Ruben RJ, Distenfeld A, Berg P, Carr R. Sudden sequential deafness as the presenting symptom of macroglobulinemia. JAMA 1969;209:1364-5. PMID 4979452

Ryu IS, Yoon TH, Ahn JH, Kang WS, Choi BS, Lee JH, Shim MJ. Three-dimensional fluid-attenuated inversion recovery magnetic resonance imaging in sudden sensorineural hearing loss: correlations with audiologic and vestibular testing. Otol Neurotol 2011;32(8):1205-9. PMID 21921851

Saadah HA. Vestibular vertigo associated with hyperlipidemia: response to antilipemic therapy. Arch Intern Med 1993;153:1846, 9. PMID 8333820

Sakata T, Esaki Y, Yamano T, Sueta N, Nakagawa T. A comparison between the feeling of ear fullness and tinnitus in acute sensorineural hearing loss. Int J Audiol 2008;47:134-40. PMID 18307093

Samuelsson AK, Hyden D, Roberg M, Skogh T. Evaluation of anti-hsp70 antibody screening in sudden deafness. Ear Hear 2003;24(3):233-5. PMID 12799545

Sando I, Ogawa A, Jafek BW. Inner ear pathology following injury to the eighth cranial nerve and the labyrinthine artery. Ann Otol 1982;91:136-41. PMID 6979284

Sauvaget E, Kici S, Petelle B, et al. Vertebrobasilar occlusive disorders presenting as sudden sensorineural hearing loss. Laryngoscope 2004;114:327-32. PMID 14755213

Schick B, Brors D, Koch O, Schafers M, Kahle G. Magnetic resonance imaging in patients with sudden hearing loss, tinnitus and vertigo. Otol Neurotol 2001;22:808-12. PMID 11698800

Schweitzer VG, Darrat I, Stach BA, Gray E. Sudden bilateral sensorineural hearing loss following polysubstance narcotic overdose. J Am Acad Audiol 2011;22(4):208-14. PMID 21586255

Seltzer S, Mark AS. Contrast enhancement of the labyrinth on MR scans in patients with sudden hearing loss and vertigo: evidence of labyrinthine disease. AJNR 1991;12:13-6. PMID 1899498

Sheu JJ, Wu CS, Lin HC. Association between obstructive sleep apnea and sudden sensorineural hearing loss: a population-based case-control study. Arch Otolaryngol Head Neck Surg 2012;138(1):55-9. PMID 22249630

Shin SH, Chun YM, Lee HK. A cochlear schwannoma presenting with sudden hearing loss. Eur Arch Otorhinolaryngol 2008;265:839-42. PMID 18004581

Sismanis A. Diagnostic and management dilemma of sudden hearing loss. Arch Otolaryngol Head Neck Surg 2005;131:733-4. PMID 16103309

Song JJ, Mertens G, Deleye S, et al. Neural substrates of conversion deafness in a cochlear implant patient: a molecular imaging study using $H_2{}^{15}O$-PET. Otol Neurotol 2014;35(10):1780-4. PMID 25166017

Spear SA, Schwartz SR. Intratympanic steroids for sudden sensorineural hearing loss: a systematic review. Otolaryngol Head Neck Surg 2011;145(4):534-43. PMID 21873598

Stachler RJ, Chandrasekhar SS, Archer SM, et al. Clinical practice guideline: sudden hearing loss. Otolaryngol Head Neck Surg 2012;146(3 Suppl):S1-35. PMID 22383545

Stahl N, Cohen D. Idiopathic sudden sensorineural hearing loss in the only hearing ear: patient characteristics and hearing outcome. Arch Otolaryngol Head Neck Surg 2006;132:193-5. PMID 16490878

Stokroos RJ, Albers FW. The etiology of idiopathic sudden sensorineural hearing loss: a review of the literature. Acta Otorhinolaryngol Belg 1996;50:69-76. PMID 8669276

Stokroos RJ, Albers FW, Krikke AP, Casselman JW. Magnetic resonance imaging of the inner ear in patients with idiopathic sudden sensorineural hearing loss. Eur Arch Otorhinolaryngol 1998b;255:433-6. PMID 9833208

Stokroos RJ, Albers FW, Tenvergert EM. Antiviral treatment of idiopathic sudden sensorineural hearing loss: a prospective, randomized, double-blind clinical trial. Acta Otolaryngol 1998a;118:488-95. PMID 9726671

Strupp M, Planck JH, Arbusow V, et al. Rotational vertebral artery occlusion syndrome with vertigo due to "labyrinthine excitation." Neurology 2000;54:1376-9. PMID 10746615

Suzuki H, Mori T, Hashida K, Shibata M, Nguyen KH, Wakasugi T, Hohchi N. Prediction model for hearing outcome in patients with idiopathic sudden sensorineural hearing loss. Eur Arch Otorhinolaryngol 2011;268(4):497-500. PMID 21042804

Toyoda K, Hirano T, Kumai Y, et al. Bilateral deafness as a prodromal symptom of basilar artery occlusion. J Neurol Sci 2002;193:147-50. PMID 11790395

Tucci DL. Sudden sensorineural hearing loss: a viral etiology. Arch Otolaryngol Head Neck Surg 2000;126:1164-5. PMID 10979138

Vazquez R, Solanellas J, Alfageme I, et al. Mitral valve prolapse and sudden deafness. Int J Cardiol 2008;124:370-1. PMID 17363095

Verghese J, Morocz IA. Acute unilateral deafness. J Otolaryngol 1999;28:362-4. PMID 10604169

Wang CT, Chou HW, Fang KM, Lai MS, Cheng PW. Treatment outcome of additional dextran to corticosteroid therapy on sudden deafness: propensity score-matched cohort analysis. Otolaryngol Head Neck Surg 2012;147(6):1125-30. PMID 22910305

Watanabe Y, Ohi H, Shojaku H, Mizukoshi K. Sudden deafness from vertebrobasilar artery disorder. Am J Otol 1994;15:423-6. PMID 8579154

Weber PC, Zbar RI, Gantz BJ. Appropriateness of magnetic resonance imaging in sudden sensorineural hearing loss. Otolaryngol Head Neck Surg 1997;116:153-6. PMID 9051056

Wei BP, Stathopoulos D, O'Leary S. Steroids for idiopathic sudden sensorineural hearing loss. Cochrane Database Syst Rev 2013;7:CD003998. PMID 23818120

Weng SF, Chen YS, Hse CJ, Tseng FY. Clinical features of sudden sensorineural hearing loss in diabetic patients. Laryngoscope 2005;115:1676-80. PMID 16148716

Wilson WR. The relationship of the Herpesvirus family to sudden hearing loss: a prospective clinical study and literature review. Laryngoscope 1986;96:870-7. PMID 3016434

Wilson WR, Byl FM, Laird N. The efficacy of steroids in the treatment of idiopathic sudden hearing loss. Arch Otolaryngol 1980;106:772-6. PMID 7002129

Wu HP, Chou YF, Yu SH, Wang CP, Hsu CJ, Chen PR. Intratympanic steroid injections as a salvage treatment for sudden sensorineural hearing loss: a randomized, double-blind, placebo-controlled study. Otol Neurotol 2011;32(5):774-9. PMID 21646929

Yamasoba T, Kikuchi S, Higo R. Deafness associated with vertebrobasilar insufficiency. J Neurol Sci 2001;187:69-75. PMID 11440747

Yimtae K, Srirompotong S, Kraitrakul S. Idiopathic sudden sensorineural hearing loss. J Med Assoc Thai 2001;84:113-9. PMID 11281488

Yin T, Huang F, Ren J, et al. Bilateral sudden hearing loss following habitual abortion: a case report and review of literature. Int J Clin Exp Med 2013;6(8):720-3. PMID 24040484

Yoon TH, Paparella MM, Schachern PA, Alleva M. Histopathology of sudden hearing loss. Laryngoscope 1990;100:707-15. PMID 2362530

Zadeh MH, Storper IS, Spitzer JB. Diagnosis and treatment of sudden-onset sensorineural hearing loss: a study of 51 patients. Otolaryngol Head Neck Surg 2003;128:92-8. PMID 12574765

Zavod MB, Sataloff RT, Rao VM. Frequency of cochlear enhancement on magnetic resonance imaging in patients with autoimmune sensorineural hearing loss. Arch Otolaryngol Head Neck Surg 2000;126:969-71. PMID 10922229

**References especially recommended by the author or editor for general reading.

## Profile

### Age range of presentation

02-05 years
06-12 years
13-18 years
19-44 years
45-64 years
65+ years

## Differential diagnosis list

inner ear ischemia
eighth nerve ischemia
viral infection of the labyrinth or cochlear nerve
Ménière disease
intralabyrinthine membrane rupture
autoimmune or inflammatory causes
retrocochlear masses

demyelinating disease
syphilis
Lyme disease
*Rickettsia felis* infection
meningitis
carcinomatous meningitis
Takayasu arteritis
perilymph fistula
toxins
barotrauma
head injury
otologic surgery
vestibular schwannoma
demyelinating disease
stroke
vertebrobasilar occlusive disease
carcinomatous meningitis
paraneoplastic syndromes
encephalitis
meningitis
thromboemboli of the posterior circulation
migraine
fat emboli
thromboangiitis obliterans
hyperlipidemia
macroglobulinemia
sickle cell disease
leukemia
polycythemia vera
other causes of hypercoagulation or hyperviscosity
hypotension
cochlea dysfunction from ischemia
mixed central and peripheral vestibular dysfunction
posterior inferior cerebellar artery territory infarction (Wallenberg syndrome)
Ramsay-Hunt syndrome
acoustic neuroma
cochlear schwannoma
medication use, including nonsteroidal anti-inflammatory drugs, aminoglycosides, or phosphodiesterase inhibitors
psychogenic sudden deafness

## Other topics to consider

Autoimmune sensorineural hearing loss
Labyrinthine infarction
Sporadic schwannomas and neurofibromas
Susac syndrome
Vestibular schwannoma