Sudden deafness

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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 (Milikan et al 1959; Milikan 1964; Fisher 1967; Gussen 1976; Milikan and Futrell 1990a; Milikan 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
the morning (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 verteobasilar 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 two 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 hearing 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 (Vergheese 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 (Iwashaki 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 synthesises 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 neurolymphitis (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).

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**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

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