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Introduction

Sudden hearing loss (SHL) is a medical emergency for which definitive diagnosis and treatment is still largely unknown. It was first described in the literature by De Klevn in 1944. SHL generally a refers to hearing loss of sensorineural origin. It has been defined for research purposes and has been accepted by most authorities as 30 dB or more sensorineural hearing loss over at least three contiguous audiometric frequencies occurring within 3 days or less.

Estimates report approximately 15,000 reported cases of SHL per year worldwide with 4000 of those occurring in the United States. One in every 10,000 to 15,000 people will suffer from this condition, with the highest incidence occurs between 50 and 60 years of age. The lowest incidence is between 20 and 30 years of age. Of the patient suffering from SHL, 2% are bilateral. In most series, the incidence was nearly equal in men and women.

There are many potential causes of SHL, but despite extensive evaluation, the majority of cases elude definitive diagnosis and therefore, remain idiopathic. Reports estimate that the etiology of SHL is diagnosed in only 10% of cases. Suggested causes of idiopathic sudden sensorineural hearing loss (ISSNHL) include viral infections, immunologic, vascular compromise, and intracochlear membrane breaks. It is unlikely that any single one of these pathophysiologic processes explains all cases of ISSNHL. Treatment regimens aimed at addressing the underlying problem in each of these states have been suggested including decreasing cochlear inflammation, improving inner ear blood flow and oxygenation, and reestablishing the endocochlear potential.

Etiology

The etiology of SHL can be broken down into broad categories: (1) viral and infectious, (2) autoimmune, (3) labyrinthine membrane rupture/traumatic, (4) vascular, (5) neurologic, and (6) Neoplastic. There are multiple conditions within each of these categories that have been associated with sudden hearing loss. The following is a partial list of reported causes of SHL:

Infectious	Meningococcal meningitis
	Herpesvirus (simplex, zoster, varicella, cytomegalovirus0
	Mumps
	Human immunodeficiency virus
	Lassa fever
	Mycoplasma
	Cryptococcal meningitis
	Toxoplasmosis
	Syphilis
	Rubeola
	Rubella
	Human spumaretrovirus
Autoimmune	Autoimmune inner ear disease (AIED)
	Ulcerative colitis
	Relapsing polychondritis
	Lupus erythematosus
	Polyarteritis nodosa
	Cogan's syndrome
	Wegener's granulomatosis
Traumatic	Perilymph fistula
	Inner ear decompression sickness
	Temporal bone fracture
	Inner ear concussion
	Otologic surgery (stapedectomy)
	Surgical complication of nonotologic surgery
Vascular	Vascular disease/alteration of microcirculation
	Vascular disease associated with mitochondriopathy
	Vertebrobasilar insufficiency
	Red blood cell deformability
	Sickle cell disease
	Cardiopulmonary bypass
Neurologic	Multiple sclerosis
	Focal pontine ischemia

	Migraine
Neoplastic	Acoustic neuroma
	Leukemia
	Myeloma
	Metastasis to internal auditory canal
	Meningeal carcinomatosis
	Contralateral deafness after acoustic neuroma surgery

History and Physical

Evaluation and management of SHL should be considered medically urgent, if not an emergency. The primary goal is to rule out any treatable causes.

Diagnostic evaluation of the patient with sudden hearing loss begins with a thorough history and physical exam. Details of the circumstances surrounding the hearing loss and the time course of its onset should be elicited. Associated symptoms, such as tinnitus, vertigo or dizziness, and aural fullness should also be asked about. Clinical experience has shown that about one-third of patients note their hearing loss upon first awakening in the morning, and that about one- half the cases will have associated vertigo. Patients should also be questioned about previous otologic surgery, ototoxic drug use, and previous or concurrent viral or upper respiratory tract infections. Any history of trauma, straining, diving, flying, and intense noise exposure should be noted. Past medical history of other diseases associated with sudden hearing loss should also be obtained such as diabetes, autoimmune disorders, malignancies, neurologic conditions (multiple sclerosis), and hypercoagulable states. African-Americans should be asked about sickle cell disease.

A complete head and neck exam should be performed on all patients with sudden hearing loss. More often than not, the exam will be unremarkable, however, any processes such as middle ear effusions, infections, cholesteatoma, and cerumen impaction should be excluded. A thorough neurological exam including Weber and Rinne, cerebellar and vestibular testing should be performed.

Diagnostic Testing

An audiogram (pure tone, speech, tympanometry, including stapedial reflex testing) should be performed on all patients with sudden hearing loss. The audiogram is the foundation of the diagnosis and provides prognostic information. Serial testing provides documentation of the progression or resolution of the hearing loss and response to treatment. In addition it may help exclude patients with secondary gain or with pseudohypacusis.

The following is a list of laboratory studies that can be ordered. Initial screening tests should be directed based on history and suspected conditions.

1) Complete blood count (CBC)
2) Erythrocyte sedimentation rate (ESR)

3) Glucose
4) Cholesterol/triglycerides
5) T3, T4, TSH
6) PT, PTT
7) VDRL, RTA-ABS (MHA-TP)
8) HIV
9) Lyme titer

Magnetic resonance imaging (MRI) is recommended by the majority of authors for patients with asymmetric hearing loss. In one survey of 79 otolaryngologists, 38% would order imaging on the patient's initial visit. MRI is useful in evaluating for acoustic tumors, multiple sclerosis and cerebrovascular accidents. There are some proponents of following these patients and imaging only if asymmetric hearing persists. However, Berg *et al*., in a series of acoustic neuromas showed that 13% presented with sudden hearing loss, and of these 23% recovered auditory function.

Known Treatable Causes of Sudden Sensorineural Hearing Loss

Autoimmune

Autoimmune hearing loss may be associated with or part of systemic autoimmune diseases such as Cogan's syndrome, Wegener's granulomatosis, polyarteritis nodosa, temporal arteritis, Buerger's disease (thromboangitis obliterans), and systemic lupus erythomatosis, or may be primary to the inner ear. The pathogenesis of immune-mediated sensorineural deafness and vestibular dysfunction are unclear, but are presumed to include: vasculitis of vessels supplying the inner ear, autoantibodies directed against inner ear antigenic epitopes, or cross-reacting antibodies. Autoimmune hearing loss implies that inner ear proteins are recognized immunologically as foreign or non-self. Some authors contend that there is no such entity as autoimmune hearing loss as these putative inner ear antigens are as of yet unknown.

Cogan's syndrome (CS) is an autoimmune disease of the cornea and vestibuloauditory apparatus that was first described by Cogan, an ophthalmologist, in the 1940s. It occurs primarily in young adults (average age of onset 22-29 years) and typically presents with interstitial keratitis (IK) and Meniere's-like attacks of vertigo, ataxia, tinnitus, nausea, vomiting, and hearing loss which develop within several months of each other. CS may also be associated with other systemic manifestations of the inflammatory process such as Takayasu's-like or medium-sized vessel vasculitis. Approximately 10% of patients develop aortitis within weeks to years after the onset. Hearing fluctuation in CS coincides with disease exacerbations and remissions. Its course often culminates in deafness. One series reported that 12 out of 18 patients (67%) developed bilateral deafness.

The cause of CS is unknown. Microbial etiology has been suggested by some as URTI precede nearly 40% of patients who present with this disease. Clinical parallels between syphilis and CS have led some to belief that CS may be caused by Borrelia burgdorferi. However, evidence so far has been inconclusive. There have also been links to Chlamydia species with CS and is an area of ongoing investigation. Temporal bone

histopathologic studies done at autopsy of patients with CS are characterized by chronic inflammation including: infiltration of the spiral ligament with lymphocytes and plasma cells, endolymphatic hydrops, degenerative changes in the organ of Corti, and demyelination and atrophy of the vestibular and cochlear branches of the eight cranial nerve.

There is no criteria currently established for the diagnosis of CS. The general thinking is that the diagnosis requires clinical signs of both eye and inner ear inflammation. Work-up should include an audiogram and laboratory tests including CBC, ESR, and RPR. Imaging including MRI and/or CT should be done primarily to rule out cerebropontine angle tumors and other disorders. MRI may show enhancement of vestibular and cochlear structures with gadolinium.

The cornerstone of therapy is corticosteroids: topical for IK and oral for vestibuloauditory involvement. Most authors suggest using prednisone 1mg/kg for 2-4 weeks with a subsequent rapid taper for cases of complete resolution and slow taper for those with incomplete response. The best outcome is in patients in whom therapy begins shortly after the onset of symptoms. For patients who have failed steroid treatment and developed bilateral deafness, cochlear implantation should be considered and has been approved by the FDA in these patients.

Traumatic

Traumatic breaks in the membranous labyrinth are accepted causes of sudden hearing loss. Cochlear membrane breaks may be either intracochlear, as is thought to occur in Meniere's disease, or involve the labyrinthine oval and/or round windows with a resultant perilymph fistula. The patients history will usually elicit an inciting event such as a blow to the head, sneezing, bending over, lifting a heavy object, exposure to sudden changes in barometric pressure (such as during flying or diving), or exposure to a loud noise. Patients who are theoretically at high risk for fistulization are those who have undergone anatomical alterations such as stapedectomy patients and in children with inner ear anomalies, such as the Mondini malformation and large vestibular aqueducts where increases in CSF pressure may be transmitted to the inner ear more easily. In clinical practice, however, patients who have undergone stapedectomy who return to their normal lifestyles including scuba diving and flying have not resulted in an obvious increase in development of symptoms of perilymphatic fistulas.

Currently there is no definitive test to diagnose perilymph fistulae other than intraoperative observation of leakage of perilymph. Diagnosis is made by the clinical history of sudden or rapidly progressive hearing loss after one of the above listed inciting events. Inflammation, granuloma, or neoplasia disorders that can mimic a perilymph fistula should be ruled out (i.e. with MRI, MHATP, and ESR). Kohut proposes that two of the following vestibular symptoms are required for diagnosis: constant dysequilibrium, a positive fistula test (Hennebert's sign), and a positional nystagmus. Testing for Tullio's phenomenon should also be done.

Initial treatment should include 5 days of strict bed rest with the head of bed elevated thirty degrees. The patient should avoid straining or hard nose blowing. Stool softeners may be given. Some suggest daily audiograms. If the patient has improvement, 6 more weeks of modified physical activity should be followed. If no improvement is

seen after five days, surgical therapy including middle ear exploration with patching of the perilymphatic fistula should be performed.

Neoplastic

Acoustic neuromas are usually associated with gradually progressive hearing loss. However, the increasingly widespread use of CT and MRI imaging of patients has indicated that nearly 10% to 19% of patients with acoustic tumors may present with SHL. The rationale for imaging patients with unilateral hearing loss, accepted by most authorities is that, although uncommon, their symptoms may be due to neoplasm. It has been estimated that 1% of patients with asymmetric SHL will have an acoustic tumor. In addition, these imaging studies can pick other neurologic disorders such as multiple sclerosis and ischemic changes which may be associated with SHL.

Idiopathic Sudden Sensorineural Hearing loss (ISSNHL): *Etiologic Theories*

Viral

Although definitive proof remains to be established, the current belief is that viral cochleitis is the culprit in the majority of cases of ISSNHL. In 1983, serologic studies performed by Wilson and colleagues demonstrated a statistically significant increase in viral seroconversion in patients with ISSNHL compared with controls for CMV as well as influenza B, mumps, rubeola, and varicella zoster viruses. They looked at 122 patients over a 3 year period with SHL and found that 63% had documented viral titer seroconversion, compared with 40% of controls. Veltri and co-workers demonstrated seroconversion at a rate of 65% in 77 unmatched patients with SHL. Temporal bone histopathologic studies have demonstrated changes that are consistent with viral infection. Schuknecht and Donovan looked at the histopathology of twelve temporal bones from patients with ISSNHL and reviewed ten other cases from the literature. They compared these with cases from patients with known viral labyrinthitis and found similar histopathologic findings including atrophy of the organ of Corti, tectorial membrane, stria vascularis, cochlear nerve, and vestibular organ. This contrasts to the findings in animal models of hearing loss of vascular origin, which causes extensive cochlear fibrosis and ossification. In 1999, Albers and Schirm directly introduced HSV-1 into the round window of guinea pigs. Histopathologic examination of the cochlea revealed similar findings as those found by Schuknecht and Donovan.

The exact mechanism by which viruses cause hearing loss has not been definitively proven. However, studies suggest that there is a significant contribution of the host immune response to the pathologic changes in the membranous labyrinth and subsequent hearing loss of patients with ISSNHL. Harris *et al*. showed that in experimental guinea pigs with CMV-induced SSNHL, immunosuppressed animals shared a lesser degree of SNHL compared with immunocompetent controls. In addition, several studies in humans have shown beneficial effects of steroids on recovery from ISSNHL. Since the primary effect of steroids is anti-inflammatory, this lends additional support to host immune mediated cochleitis in response to a virus.

Direct identification of viruses or viral antigens from the perilymph of a affected patients provides some of the strongest evidence for the involvement of viruses in hearing loss. Also, animal studies that demonstrate the ability of viruses to infect the inner ear lend further support. Davis and Johnson demonstrated the ability of rubeola and mumps to infect the inner ears of animal models using immunofluorescent antigen studies. Westmore *et. al*. subsequently cultured the mumps virus from the perilymph of a patient suffering from SHL and in 1979, Davis and co-workers cultured CMV from the perilymph of infant with congenital CMV infection. In 1981, further studies using immunofluorescence showed CMV antigens on the stria vascularis, dark cells and Reissner's membrane of an infant who died of a CMV infection.

Autoimmune

McCabe first described autoimmune inner ear disease (AIED) in 1979. He reported on eighteen patients with bilateral rapidly progressive sensorineural hearing loss (SNHL) for which no identifiable cause could be found. Evidence of autoimmunity in these patients included a positive lymphocyte inhibition test. In this test, patient's serum was combined with antigens derived from the membranous labyrinth of patients who had undergone translabyrinthine resection of acoustic tumors. All patient's sera reacted to the antigens while controls did not. Additional evidence of autoimmune etiology is indicated by the fact that patients had substantial hearing improvement with steroid treatment. Furthermore, histopathologic examination of one patient's temporal bone revealed vasculitis. McCabe proposed that the diagnosis of AIED be based on positive immune laboratory tests and beneficial treatment response.

The clinical picture of AIED usually consists of rapidly progressive bilateral sensorineural hearing loss usually in the absence of other systemic manifestations distinguishing it from other known autoimmune disorders discussed previously. Approximately 50% of patients will complain of dizziness. Episodic light-headedness or mild ataxia is more common than true vertigo. Also, these episodes occur multiple times daily during active disease as opposed to two or three discrete episodes per week, as is seen in Meniere's disease. Occasionally symptoms of pressure and tinnitus can occur. The symptoms often progress over weeks or months but can also present as sudden hearing loss or protracted disease over many years. Most patients present with bilateral disease, and when dizziness is present, vestibular testing usually reveals bilateral reduced response. AIED has a slight predominance in middle-aged females, but can occur in both sexes and can begin in childhood.

The diagnosis of AIED is based for the most part on the presence of bilateral progressive sensorineural hearing loss and response to therapy. Hughes proposes that the two most clinically helpful tests for diagnosing AIED are the lymphocyte transformation test (LTT) and the Western blot immunoassay, however, these are rarely used by most practitioners because of availability. The sensitivity and specificity for LTT are estimated to be 50-80% and 93% respectively with positive predictive values ranging from 56-73% depending on the disease prevalence in the tested population. When applied to high risk populations (patients with bilateral rapidly-progressive SNHL), the Western blot has a sensitivity of 88%, a specificity of 80%, and an overall positive predictive value of 92%. The natural history of AIED is not known, however, clinical experience reveals that the disease waxes and wanes. Testing of patients should, therefore, be performed during periods of disease activity and before treatment is initiated.

Currently, tests for AIED are not routinely used except at certain centers (Cleveland Clinic and the Massachusetts Eye and Ear Infirmary) and in experimental trials. The major drawback to these studies is the lack of availability. If testing is desired, samples of whole blood from patients can be mailed to the Cleveland Clinic Foundation Regional Laboratory by overnight carrier for LTT. The test costs $120.00 and results take approximately seven days.

The best theoretical test for AIED would be a test for marker specific for AIED. Attempts have been made in this area and are promising. In 1990, Harris and colleagues published the results of studies which discovered, using Western blot, an anti-68kd autoantibody in the sera of patients with rapidly progressive SNHL. Since then, other studies have confirmed these findings. Overall 22% to 58% of sera of patients with rapidly progressive SNHL will contain this antibody. Harris has subsequently reported a 94% specificity for test correlating results with responsiveness to therapy and disease activity. Studies by Billings and Harris are now searching for the specific antigen involved in AIED. So far they have isolated a 68kd protein that is ubiquitous in the inner as well as other areas of the body, and have recently reported evidence that links the 68kd antigen with heat shock protein 70 (hsp 70), a highly inducible stress protein. Further research is needed in this are to determine the exact relationship of hsp 70 to AIED and whether it plays an important etiologic role or whether it is just a bi-product of the disease itself. Theories proposed are that (1) human hsp 70 may have a similar amino acid sequence to an infecting agent resulting in cross-reactivity or (2) that there may be a hsp 70 specific to the inner ear that is seen as foreign when it is over-expressed during times of chronic inflammation from an outside agent.

Treatment for AIED is controversial and widely varied from practitioner to practitioner. This is largely due to the lack of double-blind, prospective clinical trials on the matter. The general consensus is that steroids are effective and should be used. Most sources recommend prednisone 1mg/kg/day for 4 weeks followed by a slow taper if the patient responds. If the patient relapses on the taper, Harris recommends instituting high dose prednisone and if continued recurrence occurs with tapering, a cytotoxic agent such as methotrexate (MTX) at a dosage of 7.5-15 mg weekly with folic acid, or cyclophosphamide (Cytoxan) should be instituted. If MTX is used, the steroids should be continued after starting the MTX as it takes one to two months for the prednisone sparing affects of MTX to begin. Most physicians begin with MTX as it has fewer side-effects than Cytoxan. If both prednisone and MTX are ineffective, cytoxan should be used. It is important to monitor for side effects of both MTX and Cytoxan with routine monitoring of complete blood counts, platelets, LFTs, UA, and electrolytes. Those on Cytoxan should keep well-hydrated to prevent hemorrhagic cystitis.

Other authors such as McCabe are more in favor of starting cytotoxic drugs at the onset of the illness. He believes that Cytoxan is the preferred treatment of AIED rather than steroids because there is a higher response rate to this drug. Because the diagnosis of AIED is based partially on response to therapy, fewer patients with this diagnosis would be missed.

Vascular

It is not surprising that the cochleovestibular blood supply may be affected by circulatory disorders such as embolic phenomenon, thrombosis, vasospasm, and hypercoagulable or high viscosity states resulting in SSNHL. The underlying pathophysiology can be explained by the occurrence of sudden anoxic injury to the cochlea. The cochlea is extraordinarily intolerant of blood supply disruptions. Early studies by Kimura and Perlman in the1957 revealed that vascular occlusion of the labyrinthine artery in guinea pigs for greater than thirty minutes led to irreversible loss of

cochlear function. Suga and co-workers performed experimental embolizations of cochlear vessels and showed loss of cochlear action potentials within 60 seconds.

Much of the hard evidence for a vascular etiology of SHL comes from histopathologic comparison of a few human temporal bones with those of animal models. Belal, in 1980, looked at two temporal bones from patients suffering from SHL. He found similar histologic findings to those of animal models of vascular occlusion to the cochleovestibular apparatus including extensive fibrosis and new bone formation.

The blood supply of the membranous labyrinth is predominantly derived from he labyrinthine (internal artery) which is branch of the anterior inferior cerebellar artery. It may less commonly be a direct branch from the basilar artery. The labyrinthine artery enters the internal auditory canal and subsequently divides into the common cochlear artery and the anterior vestibular artery. The common cochlear artery divides to form the main cochlear and vestibulocochlear branches. Division of the vestibulocochlear artery results in the posterior vestibular artery and the cochlear ramus. The arterial supply to the cochlea is such that the basal turn is fed first by the main cochlear artery with the cochlear apex fed last. Based on this anatomy one would expect occlusion of the labyrinthine artery to cause both vestibular and auditory symptoms which is supported by histopathologic findings as describe above. In addition, one would expect temporary occlusion in blood flow to affect low frequency areas of the cochlea first as these areas are the most distal in terms of blood supply.

SHL can develop as a result of a number abnormal circulatory states. Patients with diseases such a sickle cell anemia and Waldenstrom's macroglobulinemia have been shown to have a higher risk of developing SHL than the normal population. Hearing loss in these individuals is usually reversible with treatment: Oxygen for sickle cell disease and plasmapheresis for Waldenstrom's. Strokes involving the anterior inferior cerebellar artery are associated with auditory and vestibular symptoms but often also affect cerebellar function. SHL following cardiopulmonary bypass has also been reported, most likely resulting from embolic phenomenon.

Treatment

The majority of cases of SHL treated by otolaryngologist are for those with no definable cause. The treatments for cases with known etiologies involve addressing the underlying condition (i.e. treatment of: acoustic neuroma with excision, ototoxicity with cessation of ototoxic drugs, multiple sclerosis with medical therapy, embolic disease with anticoagulants, sickle-cell crisis with oxygen, bacterial meningitis with antibiotics and so on). This discussion will be limited to the treatments which have been proposed for patients with no underlying cause for their hearing loss based on the previous mentioned theories addressing the etiology of ISSNHL.

Therapy for ISSNHL is a subject of controversy. The high spontaneous recovery rate of ISSNHL (47% to 63%) and its low incidence make validation of empirical treatment modalities difficult. Many treatment regimens have been proposed for ISSNHL. Below is a list of treatment modalities which have been used and some of which are currently used today for the treatment of ISSNHL:

Antiinflammatory/immunologic agents	Steroids
	Prostaglandin

	Cyclophosphamide Methotrexate
Diuretics	Hydochlorothiazide/triamterene Furosemide
Antiviral agents	Acyclovir Valacyclovir
Vasodilators	5% carbon dioxide with 95% oxygen (Carbogen) Papaverine Buphenine (nylidrin) Naftidrofuryl (nafronyl) Thymoxamine Prostacyclin Nicotinic acid Pentoxifylline
Volume expanders/hemodilutors	Hydroxyethyl starch Low-molecular-weight dextran
Defibrinogenators	Batroxobin
Calcium antagonists	Nifedipine
Other agents and procedures	Amidotrizoate Acupunture Iron Vitamins Procaine

A review of the literature confirms that the level of evidence for treating this condition is limited and consequently there is a wide disparity in consultant's practices.

In 1987, Wilkins and associates treated 109 with a "shotgun" regimen that included dextran, histamine, Hypaque, diuretics, steroids, vasodilators, and carbogen inhalation. They found no statistically significant difference in outcome between patients treated and those who were not treated. In this study there was no control group and treatment was defined as receiving the drug for at least three days. Prospective, randomized, double-blind studies have been done looking at the dextran 40, pentoxifylline, low-molecular-weight dextran, and intravenous procaine. None of these have shown significant differences in recovery compared to placebo.

Despite the disappointing results in early trials of steroid therapy in which low doses were used for short duration, later studies using higher doses for longer periods of time have been more promising in the treatment of ISSNHL. In 1980, Wilson and colleagues performed double-blind studies for the treatment of ISSNHL with oral

steroids. Their treatment consisted of a Decadron taper over 10-12 days. They stratified their patients based on audiogram and found that steroids had a significant effect on the recovery of hearing in patients with hearing loss between 40 and 90 db. Moskowitz *et al*. confirmed Wilson's findings in 1984. He demonstrated an overall 89% recovery rate for those treated with a twelve day Decadron taper compared with 44% recovery for those not treated with steroids. Again, these results were found in a patients with audiograms in the so called "steroid effective zone". In both of these studies, they found that patients with profound hearing loss did not benefit significantly from steroid use. They concluded that steroid appear to be of benefit only if the injury is partial and reversible. In a recent review by Hughes, he recommended treating with prednisone 1mg/kg/day for at least ten days and up to one month.

Hughes also advocates treatment with diuretics and low salt diet. Athough there is no data to support this treatment, his rationale is based on histopathologic studies of temporal bones in patients with autoimmune SHL (mostly Cogan's syndrome) which have shown endolymphatic hydrops. He prescribes Maxide (Hydrochlorthiazide 25mg and triamterene) once a day.

Carbogen, which is a combination of 95% oxygen and 5% carbon dioxide has been used, and is still used by some physicians in patients in whom a vascular etiology is suspected for their hearing loss. Studies have shown that carbogen increases the partial pressure of oxygen in perilymph. In addition carbon dioxide is a known potent vasodilator of the vestibulocochlear vasculature, resulting in increased blood flow. Administration of Carbogen must be done in-hospital over three days to monitor blood pressure which may increase. Insurance companies currently do not cover this treatment because it is considered experimental. No conclusive data has shown Carbogen inhalation to be of any benefit over spontaneous recovery .

Finally, antivirals have recently come into favor in the treatment of ISSNHL. Animal models of viral labyrinthitis treated with prednisone and acyclovir combined have shown significantly higher rates of hearing recovery compared to either drug alone. This combination therapy has already proven its effectiveness in Ramsay Hunt syndrome and herpes zoster oticus and has also been proposed for the treatment of Bell's palsy. Studies are now ongoing to look at the efficacy of acyclovir in the treatment of patients with ISSNHL. In general, many authorities treat patients empirically with acyclovir usually for ten days.

Recently, a survey was sent to 100 consultant otolaryngologists in the United Kingdom inquiring about their assessment and management of patients presenting with SHL. Results showed that 78% would perform routine blood tests including CBC, ESR, and syphilis serology. 38% would order an MRI at the first presentation. 98.5% of the consultants would treat with steroids alone or as part of a combination. 41% treat with Carbogen, 31% with acyclovir and 35% with betahistine.

Prognosis

Published series report spontaneous recovery rates for patients with SHL range from 47% to 63%. These reviews combined patients with partial and complete recovery and patients will all audiogram types. Four variables have been shown to affect recovery from ISSNHL: (1) time since onset, (2) audiogram type, (3) vertigo, and (4) age. In

1984, Byl published a prospective study conducted over 8 years that evaluated 225 patients with SHL. Factors evaluated included age, tinnitus, vertigo, audiogram pattern, time elapsed from onset of hearing loss to initial visit, and ESR level with respect to recovery. His findings were as follows:

1) **Time since onset** - His study confirmed that the sooner the patient was seen and therapy initiated, the better the recovery. 56% of patients presenting within the first seven days of their hearing loss recovered compared to 27% who presented thirty days or later. He noted that there is some self-selection bias whereby those that recover rapidly do not seek medical aid.
2) **Age** - The average age for those recovering totally was 41.8 years. Those under 15years and over 60 years had significantly poorer recovery rates.
3) **Vertigo** - Patients with severe vertigo had significantly worse outcomes than patients with no symptoms of vertigo. 29% of patients with vertigo recovered compared to 55% with no vertigo.
4) **Audiogram** - Patients with profound hearing loss significantly decreased recovery rates compared to all other groups (22% with complete recovery).

Other series have shown that patients with midfrequency hearing loss, particularly when hearing at 4000kHz was worse than 8000kHz, have an excellent prognosis. 100% of patients in Laird's series in 1983 recovered completely. The majority of studies confirm the findings that profound hearing loss is a poor prognostic sign indicating more severe injury.

Conclusion

Sudden hearing loss is a medical condition which can be particularly devastating to patients and frustrating for the otolaryngologist to diagnose and treat. Despite extensive investigation, only minimal data has been generated in the past thirty years to improve our understanding of the etiology and appropriate treatment of this disease. Most authorities agree that all patients should undergo audiometry, and imaging with MRI for those patients with asymmetric hearing loss, however the etiology for the majority of patients will go undiagnosed. Treatment is more controversial. Steroids have been shown to significantly improve hearing recovery in patients with moderate to severe hearing loss and seem to be favored for the treatment of autoimmune and idiopathic forms of SSNHL. The remainder of proposed treatments for this disease are based, for the most part, on theory and will require further investigation to confirm or disprove their efficacy.

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